

Pediatric Cataract Surgery

Techniques, Complications, and Management



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Second Edition

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Preface

This second edition textbook on pediatric cataract surgery is almost a complete rewrite. Many significant surgical advances for cataract management in children have occurred since the first edition of this book was published in 2005. We were surprised, when we embarked on the first edition of this book, that there existed no single reference textbook on pediatric cataract surgery. As with the first edition, our goal with this updated text is to provide an overview of state-of-the-art cataract management for children. We hope that it is both practical and scholarly, both easy to read and comprehensive. We have tried to cover all the necessary details on the detection, analysis, and management of pediatric cataracts.

Pediatric cataracts are common and represent one of the most treatable causes of lifelong visual impairment in this population. The global estimate of 1.5 million severely visually impaired and blind children is relatively low compared to the 17 million adults who are blind owing to cataracts. However, the burden of disability in terms of “blind-years” is approximately 75 million (1.5 million children \times 50 years of age) because of the child’s life expectancy after developing the visual disability.

Presently, the only known treatment for a cataract is the surgical removal of the opaque lens. This is often followed by implantation of an intraocular lens (IOL), either at the time of surgery or as a secondary procedure later in childhood. Management of congenital and childhood cataracts is more challenging than cataract management in adults. Increased intraoperative difficulties, a propensity for increased postoperative inflammation, and the changing refractive state of the eye can make perioperative decision-making more complex. More common postoperative complications, such as capsule opacification, secondary membranes, and postoperative glaucoma, and the tendency to develop amblyopia all add to the difficulty of achieving a good outcome. Cataract surgeons worldwide face the unique challenges posed by children’s eyes, often without knowing how to alter the familiar adult approach to maximize the chance for a successful outcome. This book is directed to both the pediatric ophthalmologist and the adult cataract surgeon. Childhood cataracts will be seen in nearly all ophthalmic practices, but not often enough for most surgeons to feel maximally comfortable with the treatment options.

Dr. Edward Epstein and Professor D. Peter Choyce (UK) performed the first intraocular lens (IOL) implantations in children in the late 1950s. Pediatric cataract

surgery, at that time, was completely nonautomated. In the 1970s, surgery for congenital cataracts was revolutionized by the popularization of lensectomy, posterior capsulectomy, and anterior vitrectomy. Dr. Marshall Parks and Professor David Taylor deserve much credit for carrying that torch and demonstrating that outcomes could be improved dramatically. However, ophthalmologists continued to be hesitant to implant IOLs in the pediatric population because of the unknown long-term effect of synthetic material, the changing refractive status of the developing eye, and the increased inflammatory response that occurs in pediatric eyes. Our good friend, Dr. Bob Sinskey provided guidance and inspiration when we first began to implant IOLs routinely in children 25 years ago. As time passed and technology improved, publications appeared supporting the safety and effectiveness of IOLs in children of nearly all ages. Advances in microsurgical techniques and the availability of better viscoelastic materials and appropriately sized and styled implants, suitable for small eyes, have greatly increased the success of pediatric cataract–IOL surgery worldwide.

Pediatric cataract management has become a rapidly changing field. The pediatric surgeon, to assure that the treatment for cataracts in children continues to evolve at its current rapid rate, must master the balancing act of being careful and conservative and yet innovative and bold. Not only do we recommend that pediatric surgeons read textbooks and study surgical videos in the spirit of lifelong learning, we also endorse what we refer to as “innovation trips.” These are visits to the operating room of a colleague who has mastered a technique or is practiced with a new technology. The surgeon who travels only to medical meetings and not to observe innovators in action, misses many opportunities to accelerate his or her own surgical evolution.

We hope that this book will also be a catalyst for discovery and progress. The text and accompanying videos are meant to do more than just increase knowledge; they are also designed to stimulate conversation, encourage observation of others in action, and develop hands-on expertise. The reader is then able to go beyond the state-of-the-art that was present at the time the words were placed on the page. If we are able to speed the process of innovation and shorten the learning curve to mastery, we will have succeeded. Many of you will see, in these pages, pearls that you may have passed to us and now we are passing on to others. We will continue to collect your

comments and ideas on how we can make this text even better. In addition, innovative peer-reviewed science will continue to be published and technology will continue to be improved.

We are committed to continuous learning and progress so that we can be ready when another edition is needed.

We believe that this second edition will be useful for ophthalmic surgeons at all levels, from the novice or occasional pediatric surgeon to the leaders in the field.

M. Edward Wilson, MD
Rupal H. Trivedi, MD, MSCR



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Genetics of Congenital Cataracts

Lucy T. Xu and Elias I. Traboulsi

THE DEVELOPING LENS

Embryology of the Lens

The eye starts developing on the 22nd day of gestation.¹ Shallow grooves first form on the opposite sides of the forebrain, eventually developing into outpockets called optic vesicles.¹ The vesicle comes into contact with the surface ectoderm and induces the differentiation of ectodermal cells into the lens placode.¹ The optic vesicle then invaginates and forms the optic cup and stalk.¹ The lens placodes invaginate as well to form the lens vesicles on both sides.¹ By the 5th week, the lens vesicle loses contact with the surface ectoderm and is located in the front part of the optic cup (Fig. 1.1).^{1,2}

Within the lens vesicle, the anterior cells give rise to a sheet of cuboidal epithelial cells.¹ The posterior vesicle cells elongate to reach the anterior wall by the end of week 7.¹ These long cells form the primary lens fibers filled with lens-specific proteins such as crystallins.^{1,3}

New secondary fibers are continuously added from the germinative zone—the section of epithelium just above the equator.⁴ Proliferation occurs rapidly here, with daughter cells migrating below the equator where they differentiate and elongate into secondary fiber cells.⁴ These new crescent-shaped cells organize in concentric circles over older cells while synthesizing crystallins.⁴ As these secondary fiber cells migrate through successive zones, they develop membrane undulations and eventually lose their cellular organelles to become compacted containers of protein solution.⁴ Eventually, the mature secondary fiber cells overlap in specific arc segments known as sutures.⁴ At birth, the suture pattern consists

of three branches shaped as a Y.⁴ As the lens grows, the sutures become more complex, eventually forming six, nine, and then twelve branch sutures.⁴ The mature lens consists of an ordered organization of epithelial and fiber cells, with peripheral fiber cells meeting at the sutures.⁵

Regulation of Lens Development

Lens differentiation is regulated by many transcription factors. Fibroblast growth factor (FGF) induces cell proliferation in low doses, whereas it induces cell migration and fiber cell differentiation at higher doses.⁶ Thus, the polarity of the lens may be due to the varying concentrations of FGF in the ocular media, with more FGF found in the vitreous than the aqueous.⁶ Bone morphogenetic protein (BMP) has also been shown to interact with FGF during early lens induction.⁶ For proper lens induction to occur, the canonical Wnt/ β -catenin pathway must be down-regulated, while the antagonistic noncanonical Wnt pathway must be up-regulated.⁶ The Wnt/ β -catenin pathway has also been shown to play a role in beta crystallin production.⁶ Finally, paired box 6 (*PAX6*) is crucial for the formation of the lens placode and the expression of crystallin genes.⁷ Crystallins are essential components of the lens, and mutations in crystallin genes account for many of the inherited nonsyndromic congenital cataracts.

CRYSTALLINS

Crystallins are major protein components of the lens. They constitute over 90% of the proteins in each fiber cell.⁸ They are crucial in maintaining lens transparency.⁹ Mutations in crystallin genes represent a large portion

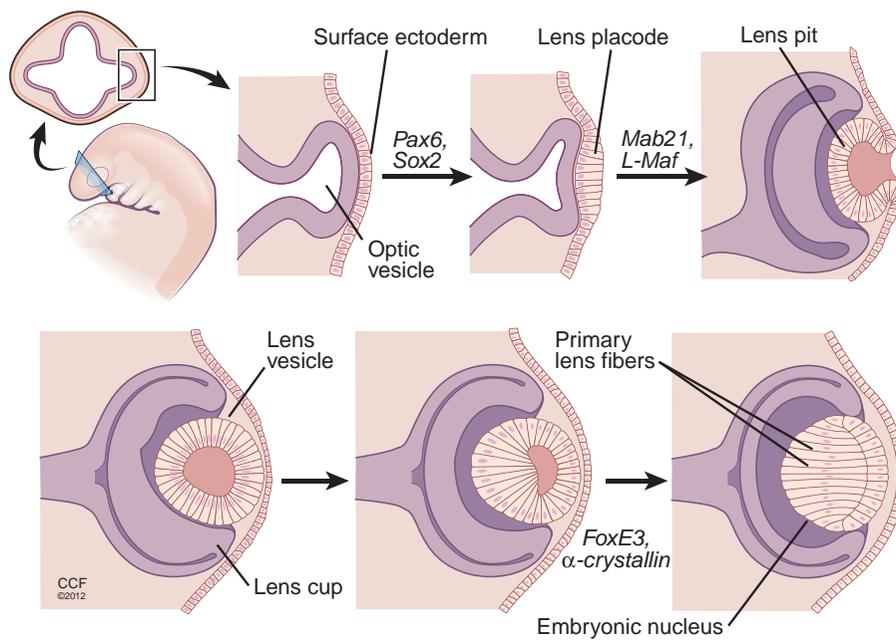


Figure 1.1. In the development of the eye, grooves in the forebrain develop into optic vesicles. The vesicle comes into contact with the surface ectoderm and induces the differentiation of ectodermal cells into the lens placode. This process is regulated by *Pax6* and *Sox2*. The lens placode invaginates to form the lens pit as regulated by *Mab21* and *L-Maf*. The lens pit will eventually detach from the surface ectoderm to produce the lens vesicle. The formation of primary lens fibers (PLF) and the embryonic nucleus is regulated by *FoxE3* and α -crystallin genes. The role of the genes is in part derived from animal experiments.²

of autosomal dominant congenital cataracts.¹⁰ Mutations have been identified in α A-crystallin (*CRYAA*), α B-crystallin (*CRYAB*), β A1/A3-crystallin (*CRYBA1/A3*), β B1-crystallin (*CRYBB1*), β B2-crystallin (*CRYBB2*), γ C-crystallin (*CRYGC*), and γ D-crystallin (*CRYGD*).¹⁰ The β - and γ -crystallins form a superfamily called the $\beta\gamma$ -crystallins that share a common polypeptide chain fold.¹¹ The α -crystallins form a separate family due to their relation to small heat shock proteins.¹¹

α -Crystallins

α -Crystallins constitute approximately 35% of the lens.¹² They are small heat shock proteins that contribute to the refractive power of the lens, and act as a molecular chaperone.¹² Specifically, α -crystallins bind unfolded and denatured proteins in the lens to prevent aggregation.¹² Aggregated proteins will interfere with the refractive properties of the lens, leading to the formation of cataracts.¹²

Two types of α -crystallins exist in the lens: α A-crystallins and α B-crystallins. The α A-crystallin gene (*CRYAA*) is on chromosome 21 and encodes a 173 amino acid protein.¹² The α B-crystallin gene (*CRYAB*) is found on chromosome 11 and encodes a 175 amino acid protein.¹² The two α -crystallins share a 57% homology.¹² α A-crystallin is expressed solely in the lens while α B-crystallin is

expressed systemically. The ratio of α A-crystallin to α B-crystallin in the lens is 3:1.¹²

Although α -crystallins are generally stable enough to tolerate many substitutions in its primary structure, certain mutations have been implicated in the formation of congenital cataracts.¹² These mutations alter the tertiary and quaternary structure of the protein, impinging on its ability to bind substrate proteins.¹⁰ Abnormal oligomerization of mutated α -crystallins have been found in congenital cataracts.¹⁰ These changes decrease the stability of the protein and reduce its chaperone ability.¹⁰ Mutations have also affected posttranslational modifications that lead to misfolded proteins that form aggregates and scatter light.¹³

An amino acid substitution mutation in the *CRYAB* gene leads to congenital cataracts but also systemic manifestations, since α B-crystallin is expressed ubiquitously.¹⁴ An A>G transition at nucleotide 3787 in the genomic sequence translates to the replacement of arginine at position 120 by glycine.¹⁴ The p.R120G mutation causes desmin-related myopathy, which is an autosomal dominant disease characterized by muscle weakness, cardiomyopathy, and congenital cataracts.¹⁴ Muscle biopsies show aggregates of desmin possibly due to altered interactions between desmin and the mutated α B-crystallin.¹⁴

A deletion in exon 3 (450delA) of the *CRYAB* gene leads to a frameshift mutation that manifests clinically as an isolated congenital cataract, inherited in an autosomal dominant pattern.¹³ The cataracts are described as bilateral posterior polar.¹³ Because the opacity is in the back of the lens and thus close to the optical center of the eye, visual acuity is severely affected.¹³ The opacity was either present at birth or developed within the first few months after birth but did not progress to any other parts of the lens.¹³

β-Crystallins

β-Crystallins are the most abundant water-soluble proteins in the lens, and their specific, closely packed arrangements play an important role in maintaining lens transparency.⁹ β- and γ-crystallins are very similar in structure. They both have two domains connected by an 8–10-amino acid peptide.¹¹ Each domain is composed of an identical polypeptide chain fold in a Greek key motif: a β-sandwich of two antiparallel β-sheets. Normally, βA3- and βB2-crystallins associate reversibly into homo- or heterodimers.¹¹ These dimers may either dissociate into monomers or further associate into multimeric complexes.¹¹ It is thought that the multimeric complexes are essential in maintaining lens transparency.¹¹ Mutations that disrupt the protein structure or change its ability to associate can cause nonspecific aggregates of these crystallins that precipitate cataract formation.

Padma et al. described a family with isolated congenital cataracts due to a mutation in the *CRYBA1* gene that encodes βA3/A1-crystallin.¹⁵ Like most isolated congenital cataracts, it is inherited in an autosomal dominant fashion.¹⁵ All affected members exhibited both zonular and sutural cataracts.¹⁵ The fetal and embryonic nuclei in the zonular cataract were clear, while the sutural cataract involved an anterior erect Y-shape and a posterior inverted Y-shape.¹⁵

A chain termination mutation in the *CRYBB2* gene that encodes for the β-2-crystallin protein has been implicated in a cerulean-type cataract.^{16,17} The cataracts manifest as blue flakes and spoke-like central opacities with only mild reductions in visual acuity.¹⁶

An autosomal recessive mutation in the *CRYBB3* gene has been shown to cause a nuclear cataract with cortical riders.⁹ While crystallin-causing cataracts often follow an autosomal dominant inheritance pattern due to the structural function of the proteins, this autosomal recessive inheritance suggests that βB3-crystallin may play a role beyond that of structure, such as an enzyme or developmental protein.⁹

γ-Crystallins

The γ-crystallins are part of the βγ-crystallin superfamily and like the β-crystallins, they form a Greek key motif with four β-sheets. It is thought that the γ-crystallins also

contribute to the maintenance of lens transparency in a similar way to that of the β-crystallins. The γ-crystallins gene cluster, (*CRYGA-F*) is located in the loci 2q33–35.¹⁸ Mutations that cause cataracts have been identified mainly in the *CRYGC* and *CRYGD* genes, which also happen to the most expressed γ-crystallin genes in humans.¹⁸ The *γS-crystallin* gene, formerly known as *βS-crystallin*, is distinct from other γ-crystallins.¹⁹ *γS-crystallin* is found on chromosome 3, and the protein is structurally similar to the other γ-crystallins except in that it has an additional α-helix between the third and fourth β-sheets.^{11,19} It has been shown that there is a calcium binding site in the Greek key motif shared by the βγ-crystallin superfamily, at the residue next to the conserved amino acid sequence Y/F/WXXXXXXXXG near the end of the first β-sheet.²⁰ Glycine is irreplaceable in this sequence as it is necessary to form a dihedral angle.¹⁹ A family with autosomal dominant progressive cortical cataracts was found to have a missense mutation at the glycine codon, possibly leading to a change in calcium binding and storage in the lens.¹⁹ A change in the calcium homeostasis may have triggered the lens opacity.¹⁹

In addition, mutations in the γD-crystallin gene (*CRYGD*) can cause an autosomal dominant aculeiform cataract, described as needle-like opacifications that involve the superficial layer of the fetal and embryonic nuclei.^{21,22}

MEMBRANE TRANSPORT PROTEINS

Because the lens is avascular, membrane transport proteins are critical in the transport of nutrients and removal of waste in the lens fiber cells, especially the cells in the center of the lens where passive diffusion is not adequate.²³ Precise control of hydration and volume of the cells will maintain the architecture necessary to establish lens transparency.²³ Four main channels exist in the fiber cells: major intrinsic proteins, connexins, Na⁺/K⁺ pumps, and glucose transporters.²⁴ However, only the first two have been implicated in the development of congenital cataracts.²⁴

Major Intrinsic Protein (Aquaporin-0)

Major intrinsic protein (MIP) is also called aquaporin-0 due to its function as a voltage-dependent water channel and adhesion molecule.²³ It is only expressed in terminally differentiated fiber cells and is the most abundant integral membrane protein in the lens.

MIP is located on chromosome 12 and encodes a transmembrane protein that has two tandem repeats, with each repeat consisting of three transmembrane α-helices and a hydrophobic loop with a conserved asparagine–proline–alanine motif.²³

Studies have revealed a point mutation, E134G, which is inherited in an autosomal dominant fashion and causes

unilamellar cataracts that do not progress after birth.²⁵ Another point mutation, T138R, causes multifocal opacities that do progress.²⁵ Both these mutations impair trafficking of MIP to the membrane surface, thus causing a loss of protein function.²⁵

Connexins

Connexins form low-resistance gap junctions between lens fiber cells.²³ They contain four transmembrane domains with two extracellular loops and one intracellular loop.²³ The transmembrane domains and the extracellular loops are highly conserved and join the lens fiber cells to each other and the epithelial cells to maintain functional ionic concentrations by mediating the transport of small molecules such as ions, metabolites, and second messengers.²³

Three connexins have been found in the lens: connexin 43, connexin 46 (encoded by *GJA3* on chromosome 13), and connexin 50 (encoded by *GJA8* on chromosome 1).²³ Mutations in *GJA8* and *GJA3* have been implicated in the development of zonular pulverulent 1 and zonular pulverulent 3 congenital cataracts, respectively. ZP1 cataracts are described to be lenticular opacities in the fetal nucleus with scattered, fine cortical opacities and incomplete cortical ridges. ZP3 cataracts are characterized by a central pulverulent opacity surrounded by snowflake-like opacities in anterior and posterior cortical regions of the lens. In addition, missense mutations in the *GJA8* gene have been shown to be responsible for congenital cataracts with microcornea.²⁶

CYTOSKELETAL PROTEINS

Cytoskeletal proteins interact with crystallins in the lens cells to define the structural framework of the cell.²⁴ Cytoskeletal proteins include microfilaments, microtubules, and intermediate filaments.²⁴ Microfilaments and microtubules form the cytoskeleton, contribute to cell-to-cell contacts and define membrane compartments.²⁴ Intermediate filaments help the lens cells overcome physical strain such as changes in temperature and during lens accommodation.²⁴

Beaded Filament Structural Protein 2

Beaded filaments are specific to lens fiber cells.²⁴ These filaments are composed of two intermediate filament proteins (beaded filament structural protein 2 and filensin) interacting with α -crystallin.²⁴ The *BFSP2* gene is found on chromosome 3, and mutations in this gene have been linked to hereditary cataracts.²⁴ One missense mutation is responsible for autosomal dominant juvenile-onset lamellar cataracts.²⁷ Another deletion mutation in *BFSP2* causes an autosomal dominant form of congenital cataracts characterized by a variety of morphologies including nuclear, sutural, and stellate or spoke-like cortical cataracts.²⁸

DEVELOPMENTAL REGULATORS

The development of the lens in embryo is directed by the spatial and temporal interaction of several genes that encode growth factors and transcription factors. Depending on the genetic defect involved, multiple abnormalities can evolve in the development of the eye.²⁹ Congenital cataracts have been linked to mutations to three of the genes: *PITX3*, *MAF*, and *HSF4*.²⁹ *PITX3* is a member of the paired-like class of homeodomain proteins located on chromosome 10.²⁴ For example, a S13N substitution mutation in a conserved region of the *PITX3* gene leads to total cataracts.²⁹ *MAF* is a basic region leucine zipper (bZIP) transcription factor found on chromosome 16 and expressed following contact between the lens placode and the optic vesicle.²⁴ The *MAF* protein has been found to bind to *MAF* response elements (MAREs) found in the promoters of crystalline genes and *PITX3*, indicating that *MAF* may direct the transcription of these genes.²⁴ One family with an R288P substitution mutation in the *MAF* gene presented with autosomal dominant purulent cataracts. However, some members of the family also had microcornea and iris coloboma, suggesting that *MAF* may have a wider role in ocular development. Heat shock transcription factor 4 (*HSF4*) regulates small and large heat shock proteins.²⁴ In contrast to small heat shock proteins such as α -crystallins, which have chaperone activity and are not ATP dependent, large heat shock proteins are ATP dependent and are also capable of refolding denatured proteins and protecting newly synthesized proteins from misfolding.²⁴ Various substitution mutations in the *HSF4* gene located on chromosome 16 can cause lamellar or total mutations.²⁹

GENETICS OF HUMAN CATARACTS

Hereditary cataracts account for 8% to 25% of all congenital cataracts.²⁹ Noninherited cataracts can result from intrauterine infections, exposure to other intrauterine insults, or indeterminate causes. Congenital cataracts can be isolated (without any accompanying systemic abnormalities), can occur in conjunction with other ocular abnormalities or in association with multisystem genetic disorders.

Isolated congenital cataracts account for approximately 70% of all congenital cataracts.⁵ Most isolated congenital cataracts are inherited in an autosomal dominant fashion,⁵ as a result of mutations in the crystallin genes, membrane transport proteins, and cytoskeletal proteins.⁵ Autosomal recessive and X-linked modes of inheritance have also been described (Table 1.1). Isolated congenital cataracts often manifest bilaterally and are often classified by their morphology based on anatomic location, size, density, and opacity.⁵ Polar opacities can involve either the anterior or posterior pole of the lens, or both poles.⁵

Table 1.1 COMMON CAUSES OF CONGENITAL OR EARLY ACQUIRED CATARACTS

Disease	Location	Gene	Phenotype	OMIM number
Autosomal Dominant				
Hyperferritinemia–cataract syndrome	19q13.33	<i>FTL</i>	Congenital nuclear cataract and elevated serum ferritin	600886
Coppock-like cataracts	2q33.3	<i>CRYGC</i>	Dusty opacity of the fetal nucleus with frequent involvement of the zonular lens	604307
	22q11.23	<i>CRYBB2</i>		
Volkman-type congenital cataract	1p36	Unknown	Central and zonular cataract	115665
Zonular with sutural opacities	17q11.2	<i>CRYBA1</i>	Zonular cataracts with sutural opacities	600881
Posterior polar 1 (CTPP1)	1p36.13	<i>EPHA2</i>	Opacity located at back of lens	116600
Posterior polar 2 (CTPP2)	11q23.1	<i>CRYAB</i>	Single well-defined plaque in posterior pole of lens; bilateral	613763
Posterior polar 3 (CTPP3)	20q11.22	<i>CHMP4B</i>	Progressive, disc-shaped, posterior subcapsular opacity	605387
Posterior pole 4 (CTPP4)	10q24.32	<i>PITX3</i>	Single well-defined plaque in posterior pole of lens	610623
Posterior pole 5 (CTPP5)	14q22-q23	Unknown	Mat reflex of posterior capsule that progresses into well-demarcated disc in posterior pole, forming opaque plaque	610634
Zonular pulverulent 1 (CZP1)	1q21.2	<i>GJA8</i>	Lenticular opacities located in the fetal nucleus with scattered, fine, diffuse cortical opacities and incomplete cortical “riders”	116200
Zonular pulverulent 3 (CZP3)	13q12.11	<i>GJA3</i>	Central pulverulent opacity surrounded by snowflake-like opacities in anterior and posterior cortical regions of the lens	601885
Anterior polar cataract 1	14q24-qter	Unknown	Small opacities on anterior surface of lens	115650
Anterior polar cataract 2	17q13	Unknown	Small opacities on anterior surface of lens	601202
Cerulean type 1 (CCA1)	17q24	Unknown	Peripheral blue and white opacities in concentric circles	115660
Cerulean type 2 (CCA2)	22q11.23	<i>CRYBB2</i>	Numerous peripheral blue flakes and occasional spoke-like central opacities	601547
Cerulean type 3 (CCA3)	2q33.3	<i>CRYGD</i>	Progressive blue dot opacities	608983
Crystalline aculeiform cataract	2q33.3	<i>CRYGD</i>	Needle-like crystals projecting in different directions, through or close to the axial region of the lens	115700
Nonnuclear polymorphic congenital cataract	2q33.3	<i>CRYGD</i>	Opacities between the fetal nucleus and the cortex of the lens	601286
Sutural cataract with punctate and cerulean opacities	22q11.23	<i>CRYBB2</i>	Dense, white opacification around the anterior and posterior Y sutures, oval punctate and cerulean opacities of various sizes arranged in lamellar form	607133
Myotonic dystrophy 1 (DM1)	19q13.32	<i>DMPK</i>	Myotonia, muscular dystrophy, cataracts, hypogonadism, frontal balding, and ECG changes	160900
Polymorphic and lamellar cataracts	12q13.3	<i>MIP</i>	Lamellar, sutural, polar and cortical opacities	604219
Cataract, autosomal dominant, multiple types 1	3q22.1	<i>BFSP2</i>	Nuclear and sutural opacities	611597
Autosomal Recessive				
Congenital cataracts, facial dysmorphism, and neuropathy (CCFDN)	18q23	<i>CTDP1</i>	Congenital cataracts, facial dysmorphism, neuropathy, delayed psychomotor development, skeletal anomalies, microcornea, and hypogonadism	604168

(Continued)

Table 1.1 COMMON CAUSES OF CONGENITAL OR EARLY ACQUIRED CATARACTS (*Cont'd*)

Disease	Location	Gene	Phenotype	OMIM number
Marinesco-Sjogren syndrome	5q31.2	<i>SIL1</i>	Congenital cataracts, cerebellar ataxia, muscle weakness, delayed psychomotor development, short stature, hypergonadotrophic hypogonadism, and skeletal deformities	248800
Warburg micro syndrome 1	2q21.3	<i>RAB3GAP1</i>	Microcephaly, microphthalmia, microcornea, optic atrophy, cortical dysplasia, in particular corpus callosum hypoplasia, severe mental retardation, spastic diplegia, and hypogonadism	600118
Warburg micro syndrome 2	1q41	<i>RAB3GAP2</i>		614225
Warburg micro syndrome 3	10p12.1	<i>RAB18</i>		614222
Martsolf syndrome	1q41	<i>RAB3GAP2</i>	Mental retardation, hypogonadism, microcephaly	212720
Hallermann-Streiff syndrome (Francois dyscephalic syndrome)	6q22.31	<i>GJA1</i>	Brachycephaly, hypotrichosis, microphthalmia, beaked nose, skin atrophy, dental anomalies, short stature	234100
Rothmund-Thomson syndrome	8q24.3	<i>RECQL4</i>	Skin atrophy, telangiectasia, hyper- and hypopigmentation, congenital skeletal abnormalities, premature aging, increased risk of malignant disease	268400
Smith-Lemli-Opitz syndrome	11q13.4	<i>DHCR7</i>	Microcephaly, mental retardation, hypotonia, micrognathia, polydactyly, cleft palate	270400
Congenital nuclear cataracts 2	22q11.23	<i>CRYBB3</i>	Nuclear cataract with cortical riders	609741
X-Linked				
Norrie disease	Xp11.3	<i>NDP</i>	Early childhood blindness, mental disorder, sensorineural deafness	310600
Nance-Horan syndrome	Xp22.13	<i>NHS</i>	Males have dense nuclear cataracts, microcornea, dental abnormalities, and developmental delay. Carrier females have posterior Y-sutural cataracts with small corneas	302350

From OMIM—Online mendelian inheritance in man. <http://www.omim.org>. Ref. 30.

Zonular cataracts involve specific regions of the lens and are also referred to as lamellar cataracts.⁵ Nuclear cataracts develop from abnormalities in the fetal or embryonic lens nucleus, while lamellar cataracts affect the lens fibers that result in a “shell-like opacity.”⁵ Zonular cataracts can also be described as dense or pulverulent (dusty).⁵ Sutural or stellate cataracts reflect abnormalities in the fetal nucleus where the lens fiber cells converge to form the suture regions.⁵ Cerulean cataracts have small blue opacities in the nucleus and cortex regions of the lens, and are thus also fittingly called blue dot cataracts.⁵ Membranous or capsular cataracts are the result of lens proteins resorption after capsular rupture caused by trauma or severe lens malformation.⁵ They can also occur in Lowe syndrome.

Congenital cataracts can also be found in conjunction with other ocular abnormalities such as Peters anomaly and aniridia.³¹ Numerous multisystem genetic disorders also present with congenital or developmental cataracts.

These include metabolic diseases such as galactosemia and Wilson disease.³¹ Patients with chromosomal abnormalities can also present with congenital cataracts. These disorders include Down syndrome and cri-du-chat syndrome, as well as numerous other trisomies, duplications, and deletions.³¹

GALACTOSEMIA

Galactosemia is defined as an abnormally high serum level of galactose. Genetically, it is due to mutations in galactokinase (*GALK1*), galactose-1-phosphate uridylyltransferase or uridine diphosphate 1-4 epimerase.⁸ In these conditions, galactitol accumulation occurs and osmotic swelling damages the lens, causing cataracts.⁸ While galactokinase deficiency is associated with isolated congenital cataracts, deficiencies in the latter two enzymes also have systemic manifestations such as vomiting, failure to thrive, liver disease and mental retardation.⁸

Galactosemia cataracts often present as nuclear cataracts, but they can also be anterior or posterior subcapsular in morphology.³² Because of the increased refractive power of the nucleus, cataracts due to galactosemia are often described as an “oil droplet.”³² These changes are initially reversible by eliminating galactose from the diet, but the anterior or posterior subcapsular component of the cataract usually remains.³²

WILSON DISEASE

Wilson disease is an autosomal recessive disorder characterized by hepatic and cerebellar dysfunction due to an excess in serum free copper. Cataracts due to Wilson disease are usually anterior subcapsular with a classic “sunflower” appearance. The cataracts can resolve with penicillamine or trientine treatment.³²

NORRIE DISEASE

Norrie disease is an X-linked recessive disorder that manifests with many congenital ocular abnormalities, including dense congenital cataracts.³³ These other ocular findings include retinal folds, retinal detachment, vitreous hemorrhage, and bilateral retrolental masses consisting of hemorrhagic vascular and glial tissue.³³ Patients are often blind at birth or by early infancy with a third of patients also progressing to sensorineural deafness.³³ Developmental delay is also seen in approximately a quarter of the patients.³³ The disease is caused by mutations in the *NDP* gene located on the X chromosome. It has been hypothesized that the Norrie protein is involved in retinal vascular development. Mutations in *NDP* have also been linked to X-linked familial exudative vitreoretinopathy (FEVR), retinopathy of prematurity (ROP), and Coats disease (retinal telangiectasis).³³

PETERS ANOMALY

Peters anomaly was first described in 1906 by Albert Peters as an ocular abnormality characterized by shallow anterior chamber, synechia between the iris and cornea, central corneal leukoma, and a defect in the Descemet membrane.³⁴ Peters plus syndrome refers to cases in which Peters anomaly occurs in association with multi-systemic malformations such as cleft lip/palate, short stature, abnormal ears, and mental retardation.³⁴ Mutations within the *PAX6* gene, *PITX2* gene, and *FOXC1* gene have all been implicated in Peters anomaly.³⁴ It is thought that mutations in these homeodomain genes control the differentiation of primordial cells and are responsible for abnormal neural crest cell migration to the posterior cornea.³⁴ The majority of cases are sporadic; however, autosomal recessive and dominant patterns of inheritance have been found in consanguineous mates, and specific

chromosomal abnormalities have also been found to be responsible for Peters anomaly. Peters plus syndrome is characterized by Peters anomaly plus other signs and symptoms including short stature, shortened upper limbs (rhizomelia) and shortened toes and fingers (brachydactyly).³⁵ Peters plus syndrome is an autosomal recessive disorder caused by mutations β -glycosyltransferase-like in the β -1,3-galactosyltransferase-like gene (*B3GALT*). β -1,3-glycosyltransferase is believed to be involved in the glycosylation of certain proteins containing the thrombospondin type 1 repeats.³⁶ Missense mutations such as the 660+1G-A mutation have been associated with some Peters plus syndrome cases.³⁷

NANCE-HORAN SYNDROME

Nance-Horan syndrome (NHS), first described by Walter Nance and Margaret Horan, is an X-linked recessive disorder characterized by central congenital cataracts, microcornea, antverted and simplex pinnae, and dental abnormalities.³⁸ For this reason, it is also known as cataract–oto–dental syndrome, X-linked cataract–dental syndrome, and X-linked congenital cataracts and microcornea.³⁸ The *NHS* gene is located on the Xp22.31–p22.13b locus.³⁸ Affected males have severe bilateral dense stellate or nuclear cataracts and usually require surgery at an early age.³⁸ Female carriers may also have mild lens opacities in the form of fine, punctate opacities aligning with or outlining the posterior Y-suture, sometimes with a dense, solid organization.³⁸ Female carriers can also have small cornea diameter and presenile cataracts.

SUMMARY

Hereditary cataracts account for a large portion of congenital cataracts. The development of the human lens requires a tightly regulated sequence of events and the interplay of many genes. Mutations in these genes negatively affect the structure and transparency of the lens, leading to cataracts. While most isolated congenital cataracts are inherited in an autosomal dominant fashion, autosomal recessive and X-linked inheritance have also been described, particularly for congenital cataracts associated with other ocular abnormalities and systemic disorders.

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2

Etiology and Morphology of Pediatric Cataracts

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Childhood cataracts can be classified as congenital, infantile, or juvenile, depending on the age at onset. Congenital cataracts are present at birth but may go unnoticed until an effect on the child's visual function is noticed or a white pupil reflex develops. Infantile cataracts develop in the first 2 years of life, and juvenile cataracts have an onset within the first decade of life.^{1,2} The term *presenile* cataract is sometimes seen in the literature. This term refers to cataracts with an onset prior to 45 years of age. *Age-related* or what has historically been called "senile" cataracts occur at or after age 45 years. Childhood cataracts can also be classified according to etiology (e.g., hereditary cataract, traumatic cataract) or morphology.³⁻⁷ Childhood cataract and lens-related pathology can have widely different appearance (Figs. 2.1 through 2.40).

ETIOLOGY OF PEDIATRIC CATARACTS

It is important to consider the origin of a cataract. The common teaching for many years has been that roughly one-third of childhood cataracts are inherited, one-third are associated with other diseases or syndromes (Figs. 2.6 and 2.40), and the remaining one-third are

idiopathic. However, the rate of idiopathic congenital cataracts has been reported as high as two-thirds.⁸ The etiology of pediatric cataracts has been reviewed by several authors, and several classifications have been proposed.^{2,3,9} The etiology of pediatric cataracts can be broadly classified and summarized in the following subgroups.

Hereditary Cataracts

Hereditary cataracts are passed from one generation to the next. Autosomal dominant transmission is responsible for 75% of congenital hereditary cataracts. Affected individuals are usually otherwise perfectly well and have no associated systemic illness. Less commonly, the inheritance may be autosomal recessive. These cataracts are bilateral but may be asymmetric. Also, marked variability can be seen between affected family members. Some cataracts are so mild that family members do not know they have them.

There are also a number of rare hereditary syndromes in which the affected individual not only has cataracts but also has an associated systemic illness. Cataracts may be associated with renal and cerebral disease as in the X-linked oculocerebrorenal syndrome of Lowe.



Figure 2.1. Anterior polar cataract in the right eye of a 12-year-old child.

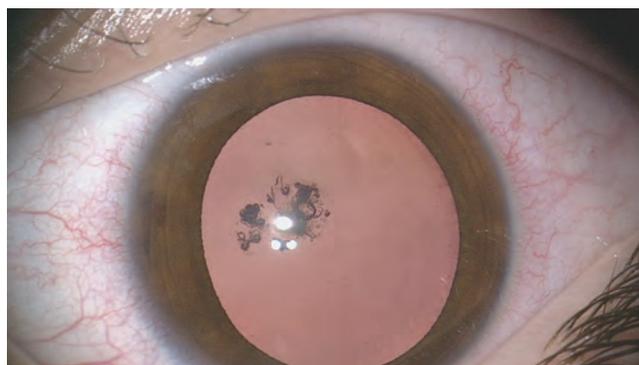


Figure 2.2. Mild posterior capsular cataract in a 9-year-old child.



Figure 2.3. Visually significant anterior polar cataract in a 2-year-old child.

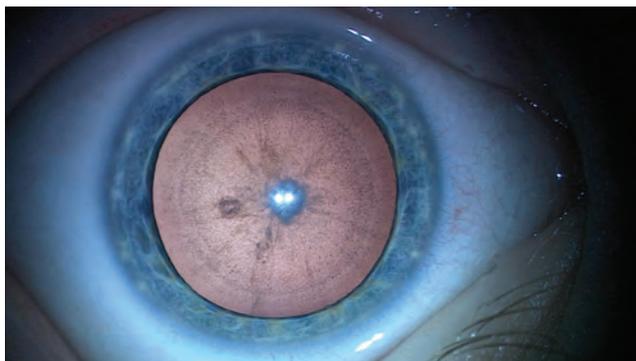


Figure 2.6. Cataract in an 18-year-old child with Down syndrome.

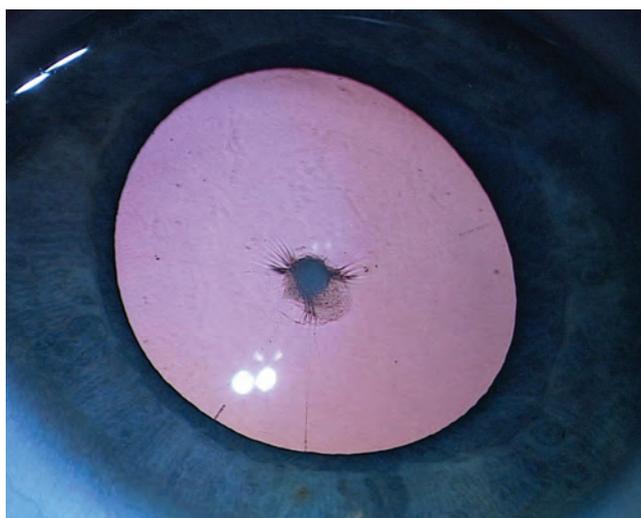


Figure 2.4. Anterior polar cataract in a 2-year-old child. Cataract had progressed to involve a portion of the surrounding cortex.



Figure 2.7. Lamellar cataract.



Figure 2.5. Slowly progressing cataract in a 13-year-old child.



Figure 2.8. Nuclear and cortical cataract.

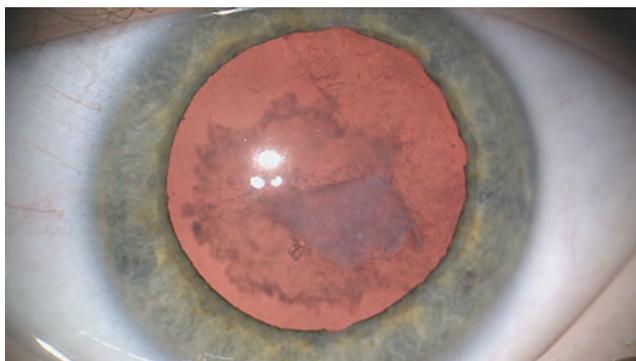


Figure 2.9. Traumatic cataract in a 6-year-old boy. History of blunt trauma 2 months ago.

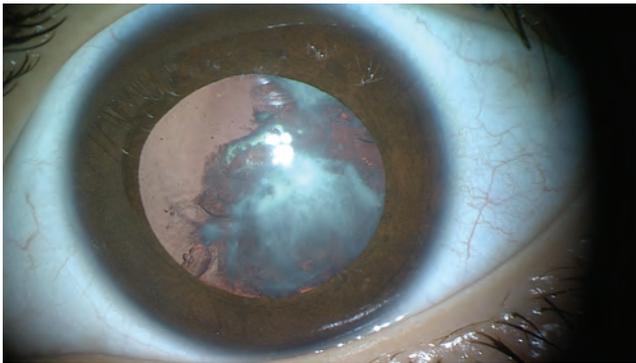


Figure 2.10. An 8-year-old child with cataract and fibrotic posterior capsule.

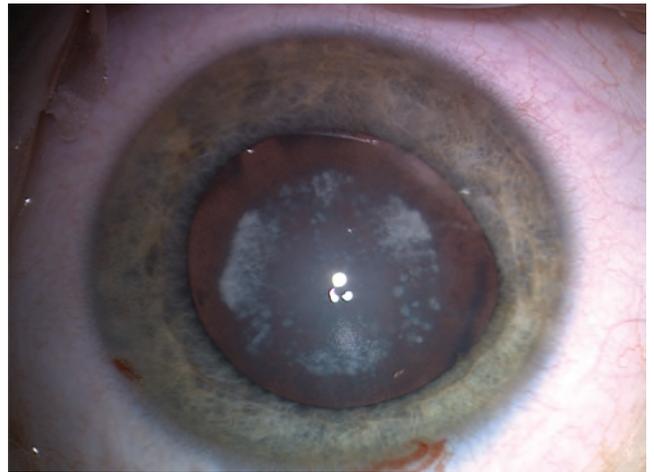


Figure 2.13. Four-year-old child with history of bilateral cataract.

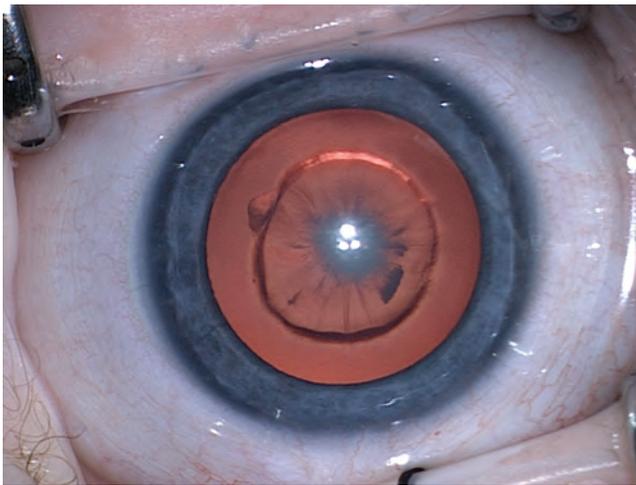


Figure 2.11. Cataract in a 1-month-old child. Posterior capsule plaque was noted at the time of cataract surgery.

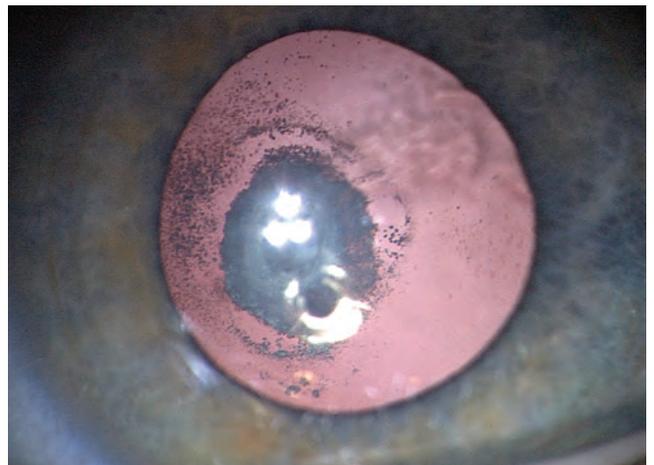


Figure 2.14. Visually significant cataract in the right eye of a 6-year-old girl. Cataract was diagnosed after failed vision screening in the school. The cataract involved at the posterior capsule and that lens material was in the vitreous.

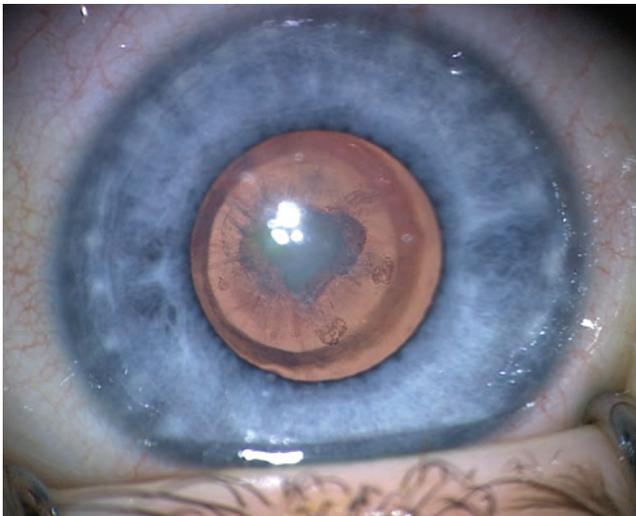


Figure 2.12. Congenital nuclear cataract associated with posterior capsule plaque in a 4-month-old child.

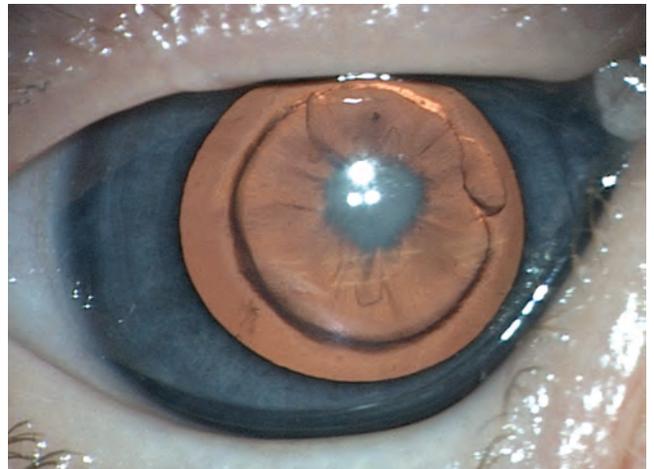


Figure 2.15. Visually significant unilateral cataract in the left eye of a 6-week-old infant. The child was noted to have a posterior cortical opacity with a posterior capsule plaque and an evolving fetal nuclear opacity as well.

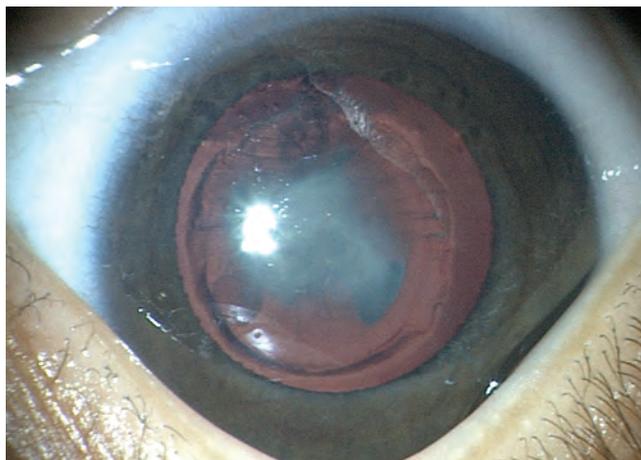


Figure 2.16. Visually significant cataract in a 1-month-old child. The cataract was nuclear and with advancing cortical spread and posterior capsule plaque.

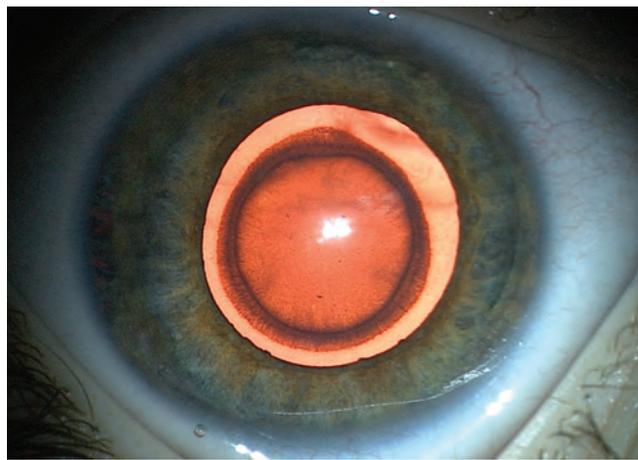


Figure 2.19. Lamellar cataract in a 3-year-old child with ocular albinism.

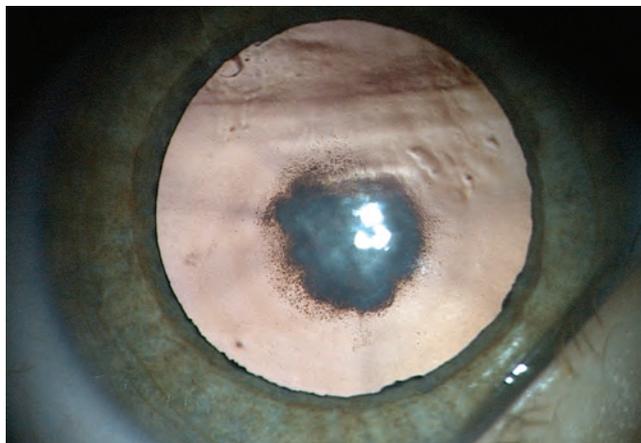


Figure 2.17. Slowly progressive posterior cataract in the left eye of a 3-year-old child.

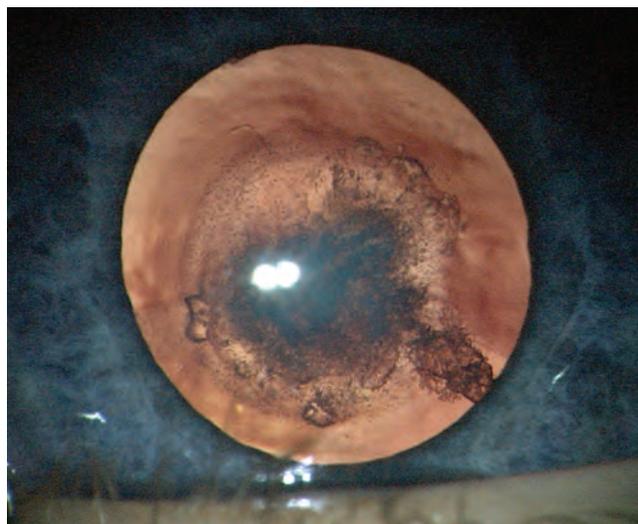


Figure 2.20. Congenital capsular and subcapsular cortical cataract in right eye of a 14-month-old child.

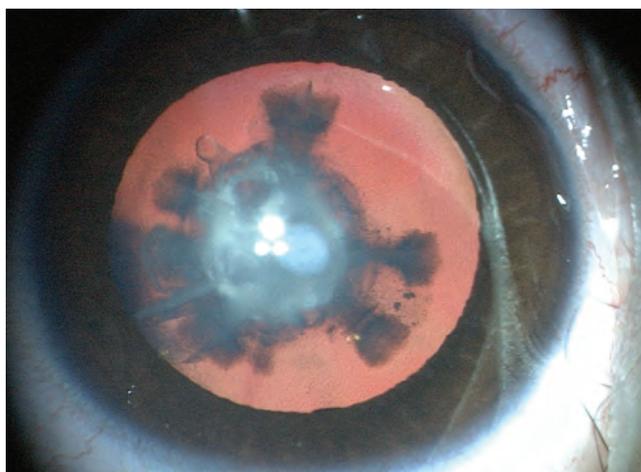


Figure 2.18. Dense nuclear and cortical cataract in a 2-year-old child.



Figure 2.21. Cataract in the left eye of a 10-month-old child.

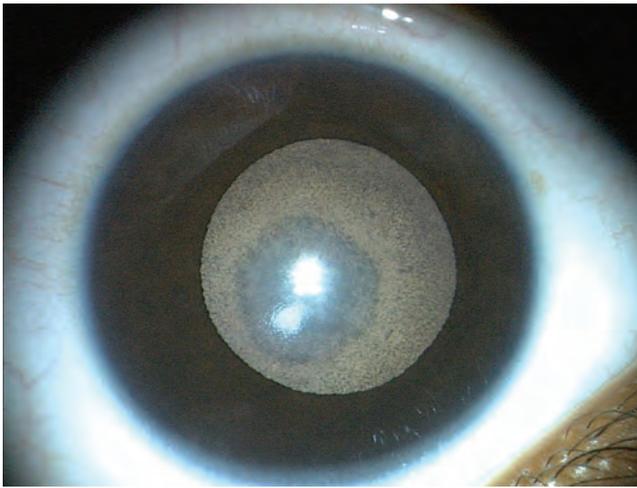


Figure 2.22. Cataract in an eye with semidilated pupil.

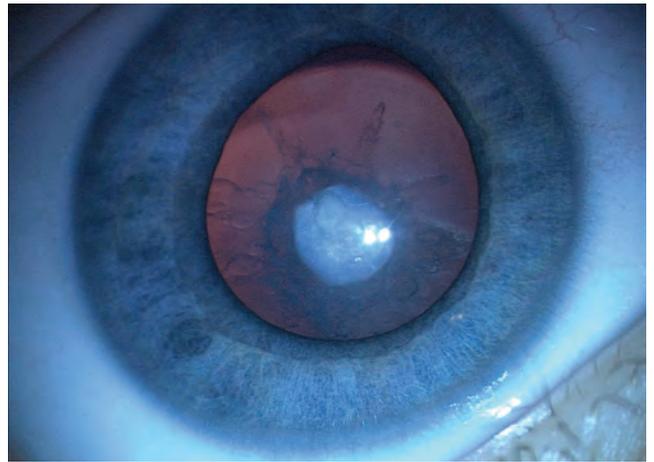


Figure 2.25. Cataract in 1 year in a 10-month-old child. The child had anterior polar cataract in both eyes; however, in this left eye, cataract has grown and spread throughout the cortex.

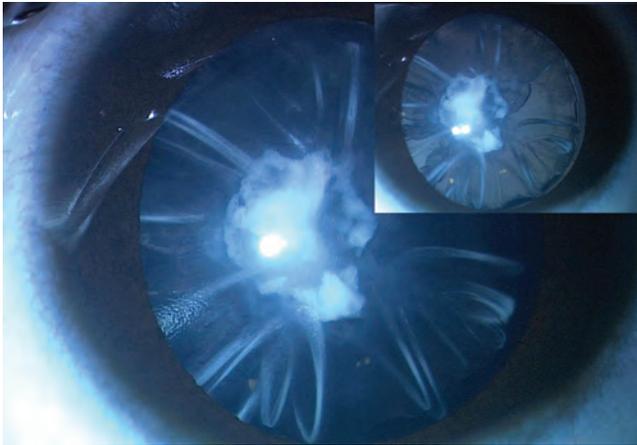


Figure 2.23. Dense cataract in a 23-month-old child.

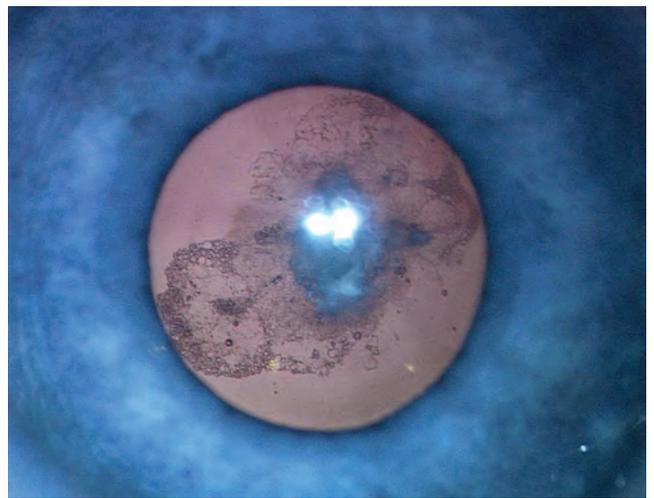


Figure 2.26. Cataract in a 6-month-old child. Cataract was posteriorly located with spread in the cortex.

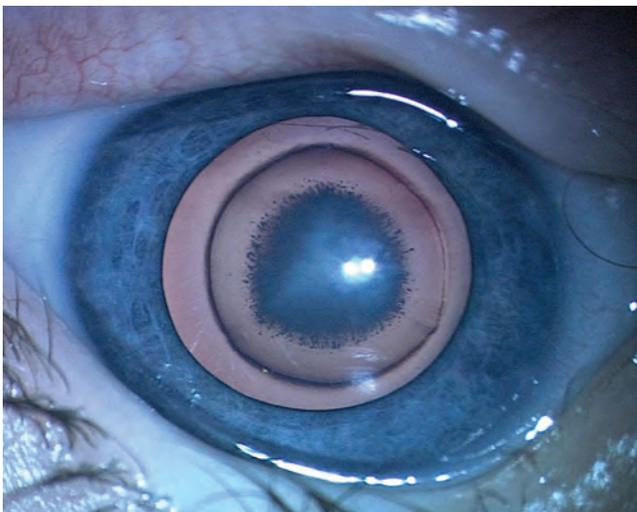


Figure 2.24. Central nuclear cataract in a 6-week-old child.

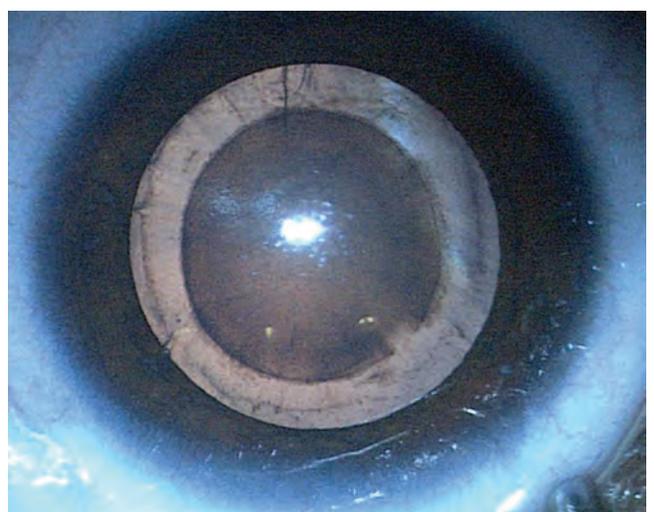


Figure 2.27. Lamellar cataract in a 2-year-old child.

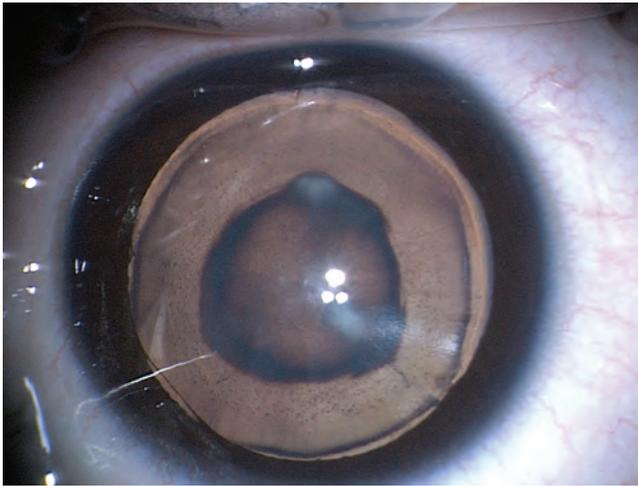


Figure 2.28. Nuclear cataract in 3-year-old child.



Figure 2.31. Anterior capsule plaque (courtesy of Dr. Abhay Vasavada, India).



Figure 2.29. Posterior lentiginous in the left eye of a 9-year-old girl. Vision is reduced to 20/300.

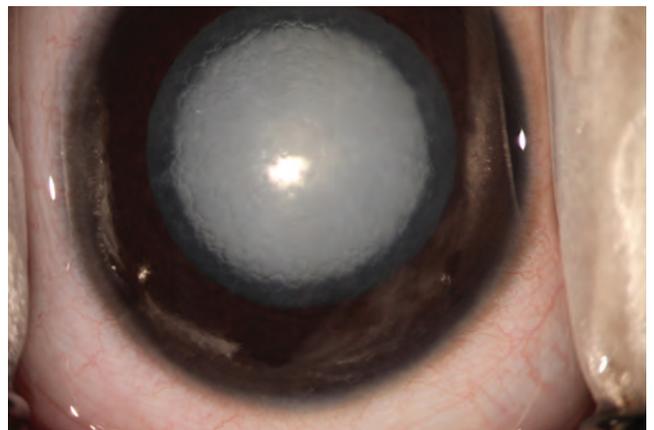


Figure 2.32. Total cataract (courtesy of Dr. Abhay Vasavada, India).

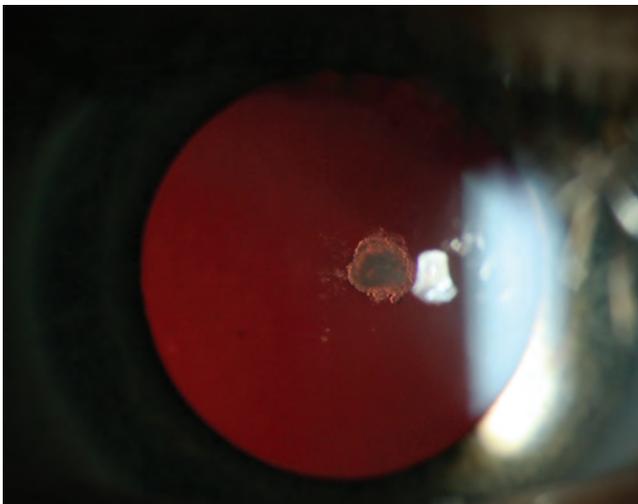


Figure 2.30. Posterior polar cataract in the left eye of an 11-year-old child. Visual acuity was 20/30; the child did not undergo cataract surgery.

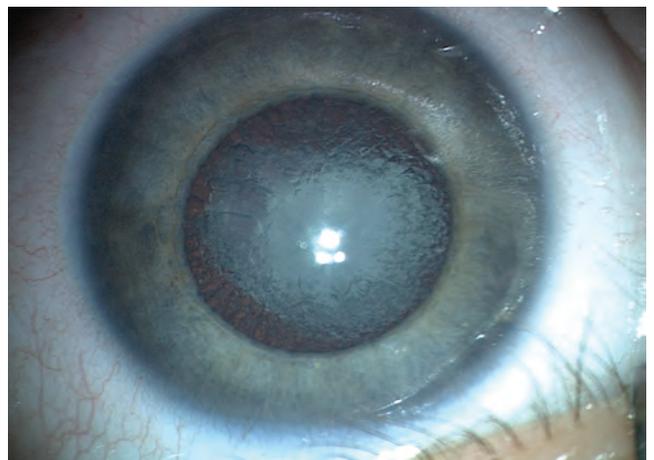


Figure 2.33. Dense cataract in a 2-month-old child.



Figure 2.34. Traumatic cataract in the left eye of a 6-year-old boy.

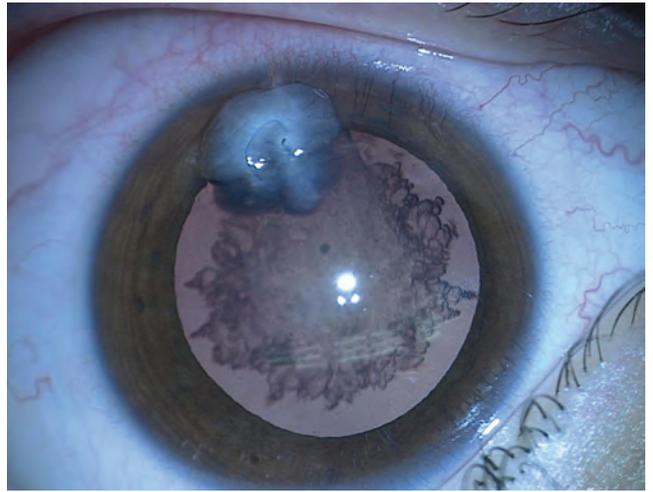


Figure 2.37. Traumatic cataract. Note corneal suture.

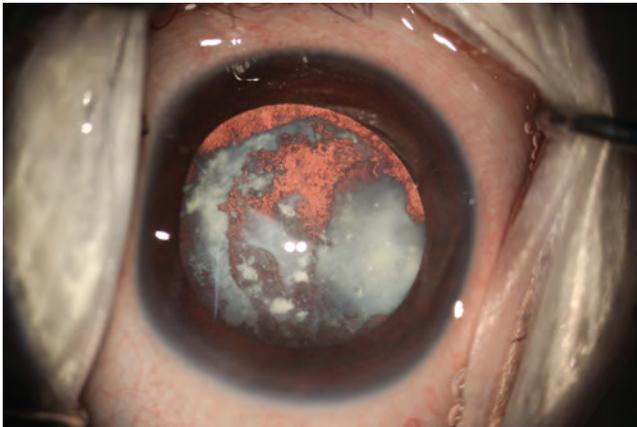


Figure 2.35. Membranous cataract (courtesy of Dr. Abhay Vasavada, India).



Figure 2.38. Traumatic cataract. Note synechia.

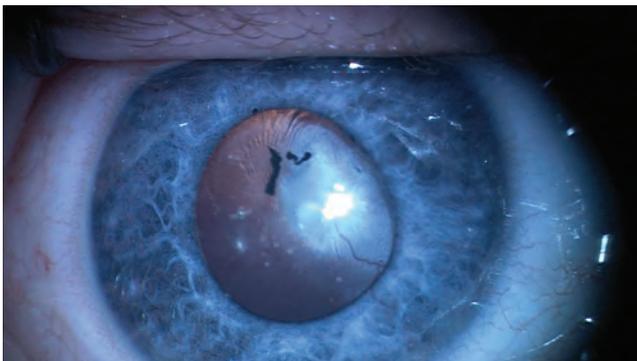


Figure 2.36. Persistent fetal vasculature and cataract in a 4-month-old child.

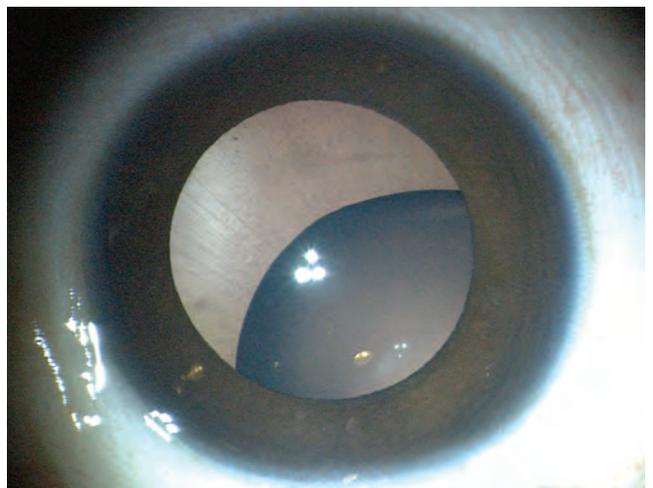


Figure 2.39. Ectopia lentis in a 5-year-old child with Marfan syndrome.

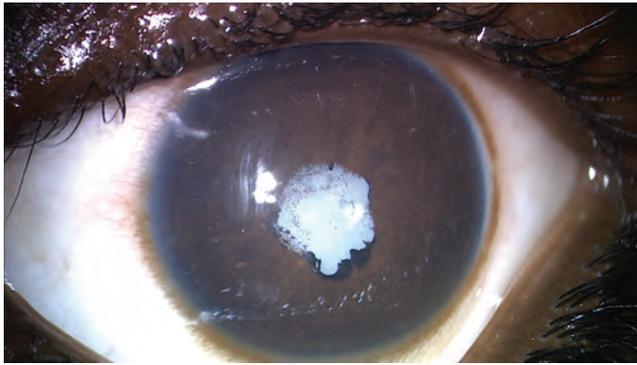


Figure 2.40. Cataract in a 12-year-old girl with a history of fetal alcohol syndrome.

An examination by a developmental pediatrician and/or a clinical geneticist is important for children with bilateral cataracts to help detect any associated systemic disorder. Genetic disorders linked to cataracts are Norrie disease, Peters anomaly, etc. (see Chapter 1).

Metabolic Cataracts

Congenital, infantile, or juvenile lens opacities may have an underlying metabolic cause. Galactosemia, for example, is a metabolic disorder in which the child's body cannot metabolize galactose, a major component of milk and milk products. The baby will have vomiting and diarrhea and may develop "oil droplet" cataracts. It is thought that 10% to 30% of newborns with classic galactosemia develop cataracts in the first few days or weeks of life. Once a newborn is put on a galactose-restricted diet, cataracts usually clear. Surgery is sometimes necessary when dietary treatment is delayed. Many galactosemia patients have eye examinations to check for the presence of cataracts on a regular basis. These examinations are required more frequently during the 1st year of life (e.g., every 3–4 months) but less often (e.g., one or two times a year) in older children. It is a good idea to have an eye exam if galactose-1-phosphate levels are observed to rise above a "target" range.

Glucose 6-phosphatase dehydrogenase deficiency is an X-linked disorder and therefore affects mainly males. These babies present with jaundice and hemolytic anemia and may also develop infantile cataracts. Infection, acute illness, and ingestion of fava beans will precipitate an attack of hemolysis in these children. Death may result unless the condition is diagnosed and treated with an urgent blood transfusion.

Hypoglycemia of whatever cause may give rise to lens opacities in children. Babies with profound hypoglycemia also have convulsions and may develop permanent brain damage. Hypocalcemia may result in cataracts, although these are usually functionally less significant than are cataracts resulting from hypoglycemia.

Traumatic Cataracts

Traumatic cataracts are a common cause of unilateral loss of vision in children (Figs. 2.9, 2.34, 2.37, and 2.38). Penetrating injuries (Fig. 2.37) are usually more common than blunt injuries (Figs. 2.9 and 2.34). Most of the injuries that result in a traumatic cataract occur in children while they are playing or involved in a sport-related activity. Thorns, firecrackers, sticks, arrows, darts, BB pellets, and automobile airbags are other items that are often implicated as a cause for traumatic cataracts. Cataracts caused by blunt trauma classically form stellate- or rosette-shaped posterior axial opacities that may be stable or progressive. Penetrating trauma often leads to cortical lens changes that may remain focal if small or may progress rapidly to total cortical opacification if the lens capsule has been ruptured.

Secondary Cataracts

The most common type of secondary cataract is a result of uveitis occurring in conjunction with arthritis (juvenile idiopathic arthritis) or as a result of intermediate or posterior uveitis of any cause. The cataract may be a direct result of inflammation within the eye or can result from the steroids used to treat the condition. Cataracts caused by steroid ingestion are usually posterior subcapsular.

Less frequently, a cataract may be seen secondary to an intraocular tumor, a foreign body, or a chronic retinal detachment. Cataracts have also been reported after laser treatment for threshold retinopathy of prematurity.

Cataracts Secondary to Maternal Infection During Pregnancy

The most common maternal infection leading to congenital cataracts in children is rubella, also known as German measles. Maternal infection with the rubella virus, an RNA toga virus, can cause fetal damage, especially if the infection occurs during the first trimester of pregnancy. Systemic manifestations of congenital rubella infection include cardiac defects, deafness, and mental retardation.

Cataracts resulting from congenital rubella syndrome are characterized by pearly white nuclear opacity. Sometimes the entire lens is opacified (complete cataract) and the cortex may liquefy. Histologically, lens fiber nuclei are retained deep within the lens substance. Cataract removal may be complicated by excessive postoperative inflammation caused by release of these virus particles. Congenital rubella syndrome, which occurred in epidemic proportions in the United States in the early 1960s, is manifest in its complete form by several systemic findings. These include failure to thrive, deafness, cardiac anomalies, patent ductus arteriosus, and thrombocytopenic rash. Congenital rubella remains an important cause of congenital cataracts in some parts of the developing world. Johar et al. quantified this in their prospective,

observational study of 172 children with cataracts from 10 days to 15 years old. They found 4.6% of nontraumatic cataracts occurred in children with congenital rubella syndrome.¹⁰ However, rubella is practically nonexistent in the United States because of widespread vaccination of the population.

Other infectious diseases that may have affected the mother during pregnancy, such as toxoplasmosis, toxocariasis, and cytomegalovirus, can also cause congenital cataracts along with systemic illness in the newborn baby. The data published by Rahi and the British Congenital Cataract Interest Group demonstrated prenatal infections and systemic factors were present in 2% of unilateral and 6% of bilateral cataracts among the 243 children with congenital or infantile cataracts.¹¹

Iatrogenic Cataracts

Iatrogenic cataracts are most commonly seen in children who have had total-body irradiation for leukemia and in children who have had organ transplants and are on long-term systemic steroid therapy. These are usually older children who do very well after cataract surgery. As stated above, cataracts have been reported after laser treatment for threshold retinopathy of prematurity.¹² Also, cataracts may develop after vitrectomy to remove a vitreous hemorrhage from birth trauma or to treat retinal detachment. Sometimes the cataract develops almost immediately after vitrectomy. In these instances, the lens capsule has usually been inadvertently opened at surgery. Infant eyes are small and soft, and vitrectomy without lens damage is difficult for all but the most experienced pediatric vitreo-retinal surgeons.

Syndromes and Congenital Cataracts

There is a large variety of chromosomal and dysmorphic syndromes in which the child will have a high risk of having congenital cataracts. It is important to notice any abnormal features in children presenting with cataracts, such as unusual facial features, extra digits, unusual skin, short stature, developmental delay, and microcephaly or hydrocephaly, as it is essential that a diagnosis is made such that the child receives any necessary treatment and the parents receive genetic counseling about the possible risks of producing other offspring with similar problems.

Cataracts of Unknown Etiology (Idiopathic or Sporadic)

The vast majority of nontraumatic unilateral cataracts fall into this category. Many surgeons would say that this is the most common single category. It is, of course, a diagnosis of exclusion. Unilateral cataracts will not be metabolic or hereditary. A search for signs of trauma or eye inflammation is warranted. Bilateral cataracts may also be of unknown etiology. However, the hereditary

nature of some cataracts will not be evident from the history alone. A careful examination of the parents may reveal visually insignificant cataracts that a parent did not know were present. In the context of the overall medical and developmental history and the age at onset of the cataract, a metabolic and genetic workup may be indicated before an idiopathic etiology can be declared. These workups should be customized with the assistance of a developmental pediatrician so that every test ordered is done so with a real suspicion for a positive finding. Shotgun workups directed by the ophthalmologist should be discouraged. Hargard et al. published their findings that bilateral idiopathic congenital or infantile cataracts were noted more often in low birth weight (<2 kg), but not significantly in unilateral idiopathic congenital or infantile cataracts.¹³ A subsequent study by Prakalapakorn et al.¹⁴ in the National Birth Defects Prevention Study, corroborated the Hargard finding that bilateral cataracts occurred significantly more in the 1.5 to 2.0 kg birth weight (low birth weight), and found that unilateral cataracts were significantly more likely in primigravid women than those with two or more pregnancies. However, Prakalapakorn found that among the very low birth weight cases (<1.5 kg), both unilateral and bilateral cataracts occurred with statistical significance. Other risk factors were assessed and fell short of statistical significance, but cataracts did occur more frequently with those factors. Unilateral cataracts occurred more frequently among mothers with substance abuse during pregnancy. Bilateral cases were more likely among mothers using aspirin during pregnancy and contracting urinary tract infections during pregnancy. This study offers both insight into findings associated with cataracts of unknown etiology and the residual need for further investigation to clarify known and novel predictors.

MORPHOLOGY OF PEDIATRIC CATARACTS

Congenital, developmental, and traumatic cataracts can have different morphologic characteristics. For surgeons, a classification based on morphologic type is most useful. The classification recommended below is in common clinical use and emphasizes those cataract types most likely to be visually significant. It is a modified (added-to) form of a clinical descriptive classification published in 1982¹⁵ and affirmed in a follow-up article in 1993.¹⁶ These simple labels lend themselves to easy stratification of a pediatric surgical case series but do not include some of the myriad rare visually insignificant variations and patterns that have been described by genetic researchers. Terms such as punctate, pulverulent, coralliform, coronary, floriform, retrodot, sunflower, blue-dot, and sutural, while possibly important to etiology, are not included here since they are used to describe lens changes that are nearly always

static and visually insignificant. The “oil droplet” type of cataract is also excluded here even though it has a classic appearance, since it either resolves when the galactosemia is treated or becomes a “lamellar” cataract. At the time of surgery, this cataract would be grouped with other lamellar opacities, rather than carrying a label based on its earlier appearance.

In this section, we have chosen to emphasize childhood cataract types based on their anatomical location within the lens and their characteristic natural history. They are all surgical or potentially surgical cataract types. Visual outcome and complications, too, can sometimes be predicted based on these lens types. Slowly progressing cataract at older age are less amblyogenic (Fig. 2.5). Posteriorly located cataract (Fig. 2.17) is more amblyogenic as compared to anteriorly located cataract. Nondilating or semidilated pupil (Fig. 2.22) not only makes cataract removal difficult but is also associated with poor postoperative outcome. This can help guide the follow-up as well as the informed consent leading to surgery.

Morphologic Classification and Characteristics of Pediatric Cataracts

The categories below are each discussed separately.

1. Diffuse/total
2. Anterior polar
3. Cortical lamellar
4. Fetal nuclear
5. Posterior polar
6. Posterior lentiglobus
7. Posterior (and anterior) subcapsular
8. Persistent fetal vasculature (Persistent hyperplastic primary vitreous)
9. Traumatic

Diffuse/Total Cataracts

Diffuse or “total” cataracts are seen most commonly after trauma. However, since trauma is a separate category, only nontraumatic total cataracts are discussed here (Figs. 2.32 and 2.33). In the United States, nontraumatic total cataracts are not common. In a previous pediatric cataract series from the United States, only 4 of 199 eyes were classified as total. However, in the developing world, total cataracts are commonly seen in children. This difference probably relates to the timing of detection of the cataracts in the developing world. Many cataract types, if left untreated, will slowly become diffuse, total cataracts. This is especially true of lamellar cataracts, posterior lentiglobus, and fetal nuclear opacities. Some total cataracts represent spontaneously ruptured posterior lentiglobus, neglected lamellar or nuclear opacities, or occult trauma. A B-scan ultrasound is indicated whenever the retina cannot be visualized using an ophthalmoscope. Surgery on total cataracts may reveal watery or partially absorbed

lens material within the capsular bag. Occasionally, the total cataract will be little more than a white membrane (Fig. 2.35) representing a fused and fibrotic anterior and posterior lens capsule (Fig. 2.10).

Anterior Polar Cataracts

Anterior polar cataracts (Figs. 2.4 and 2.25) are often bilateral, hereditary, and visually insignificant. However, notable exceptions occur. If neither parent has anterior polar cataracts, the child may have a sporadic (nonhereditary) form or may have a new mutation and thus pass on some risk of recurrence to the next generation. The most common type of anterior polar cataract presents as a tiny white dot in the center of the anterior capsule. These cataracts are usually bilateral but may be unilateral and probably represent a mild abnormality of lens vesicle detachment. They are usually 1 mm or less in diameter and almost never progress. Corneal astigmatism may be present, however, and can cause amblyopia.⁶

Pyramidal cataracts are a distinct and more severe form of anterior polar opacity, named because the shape of the anterior opacity resembles a pyramid. A more accurate and modern description is a likeness to the shape of the chocolate candy called the Hershey’s Kiss. The tips of these opacities extend into the anterior chamber and rarely have even been known to be fused with the cornea.¹⁷ They are fibrous and may be associated with an underlying cataract in the anterior subcapsular cortex that, when present, often progresses to become visually significant. At surgery, the fibrous “Hershey’s Kiss” is not easily removed with the vitreous cutter. After it is detached from the anterior capsule, it usually spins around the anterior chamber and has to be delivered through the incision using forceps. These pyramidal cataracts are almost always bilaterally symmetric and may be dominantly inherited.

Anterior lenticonus is less common than the posterior variety and is usually associated with Alport syndrome. Cataracts are a late finding in anterior lenticonus, with proper slit-lamp photography assisting to assess the lens curvature before cataracts diagnosed.¹⁸ However, the secondary refractive error may cause enough visual symptoms to require clear lens extraction and intraocular lens placement. Rarely, the lens will spontaneously rupture, causing lens hydration and total cataract. See Chapter 41 for more on Alport syndrome and anterior lenticonus. Early anterior lenticonus is most easily detected using the retinoscope and can be easily missed at the slit lamp.

Cortical Lamellar Cataracts

Lamellar cataracts (Figs. 2.7, 2.19, and 2.27) are usually acquired (rather than congenital) and involve a layer (lamellae) of cortex surrounding the fetal nucleus, peripheral to the “Y” sutures. They are almost always bilateral

but are commonly asymmetric. Microphthalmia is not usually associated with this cataract type, and the risk of secondary aphakic/pseudophakic glaucoma is much lower than with fetal nuclear opacities. Lamellar cataracts are often hereditary, following an autosomal dominant transmission pattern. The visual prognosis is usually better with lamellar cataracts (even when surgery is delayed) than with cataract types that are densely opaque at birth such as fetal nuclear opacities (discussed below). This improved prognosis is related to the later development of the cataract such that during the critical period of visual development, the cataract did not preclude normal visual development. Lamellar cataracts are characteristically mild initially and slowly worsen with time. Nystagmus does not often develop as a result of these cataracts. Remarkably, children can sometimes function quite well visually even when the lamellar opacity blackens the retinoscopic reflection completely. These cataracts are usually about 5 to 6 mm in diameter and characteristically have a thin layer of clear cortex external to the opacity. The nucleus, internal to the cataract, is also characteristically clear. At surgery, the lamellar opacity will sometimes pop out of the capsular bag as soon as the anterior capsule is opened and the cortex is hydrated. Care must be taken to avoid an extension of the anterior capsulotomy edge when this happens. Despite this tendency to pop out of the bag, lamellar cataracts are soft and can be aspirated easily.

Fetal Nuclear Cataracts

The most common congenital cataract is the fetal nuclear cataract. In the Infant Aphakia Treatment Study (IATS), cataract morphology was categorized carefully for each case.¹⁹ In the IATS cohort, 54% of analyzed surgical videos (84 available of 114 cases) demonstrated a nuclear cataract. The IATS morphology review also revealed that all fetal nuclear cataracts demonstrated a posterior capsule plaque (Figs. 2.11, 2.12, 2.15, and 2.16). Nuclear cataracts (Figs. 2.20, 2.21, and 2.26) usually present with a white central opacity (unilateral or bilateral) about 3.5 mm in diameter (between the “Y” sutures) surrounded by mostly clear cortex. As time passes from birth, the surrounding cortex may become more opaque diffusely or in radial extensions called “riders.” Microphthalmia and microcornea are usually present. The iris often dilates poorly and appears immature, with few crypts, a poorly formed pupillary ruff, and little or no collarette. These cataracts can be unilateral or bilateral and can lead to deprivation amblyopia if not removed early in life (before 6 weeks of age if unilateral or before nystagmus appears if bilateral). According to Forster et al.,²⁰ nuclear cataracts are the most severe type for visual impairment, based on their subjective scale. In their study, they combined a subjective score and analytic intensity profiles of light transmission based on photo-slit-lamp studies. Nuclear cataracts, combined with persistence of fetal vasculature (PFV) and posterior

lenticonus, constitute the types in their study requiring early surgery to avoid visual deprivation. Sometimes a hyaloid artery remnant is also present in nuclear cataracts, but unlike classic forms of severe PFV, the cataract is predominantly nuclear and no ciliary process traction is seen. Less common nuclear opacities include the varieties of pulverulent (pulverized tiny dots) cataract. These, as stated above, are usually static and do not often need surgery.

Posterior Polar Cataracts

Posterior polar cataracts (Fig. 2.30) are usually sporadic cortical opacities with a propensity for spontaneous posterior capsule rupture (Fig. 2.14). These cataracts can be unilateral or bilateral, mild (Fig. 2.2) or severe. Caution is in order when doing surgery for posterior polar opacities since the posterior capsule may already be ruptured or honeycombed into a weakened meshwork. Aggressive hydrodissection is discouraged when any posterior polar opacity is seen, since it may result in a large uncontrolled posterior capsular tear, with cortical lens material pushed back into the vitreous gel. Common posterior polar opacities are distinct from posterior lentiglobus but in some cases there can be overlap. A posterior polar opacity with a bulging posterior capsule can be seen. In posterior lentiglobus (see Posterior Lentiglobus section below) the bulge begins first before any opacity forms. With posterior polar, the opacity is present first. When detected late, both a bulge and a polar opacity may already be present.

Posterior Lentiglobus

Posterior lentiglobus (Fig. 2.29) is mostly unilateral and not associated with microphthalmia. It represents the most common type of developmental cataract in a normal-sized eye. Most forms are sporadic, but occasionally an autosomal dominant inherited bilateral form will be encountered. The lens changes begin in the posterior capsule, possibly secondary to a weakness in the area of prior contact with the hyaloid artery. The bulge in the posterior capsule is usually not present at birth but becomes more exaggerated as the intralenticular pressure increases with age. Although some publications refer to this lens capsule bulge as a posterior “lenticonus,” the term *lentiglobus* better reflects the globular, not conical, bowing of the capsule. The disorder is progressive and usually requires surgery. Cataractous changes occur in the posterior cortical layers as the lentiglobus bulge worsens. Spontaneous rupture may occur, leading to a total white cortical cataract. In these cases, the true nature of the lentiglobus may not be discovered until after the lens is entered during surgery. However, posterior capsule rupture can be diagnosed preoperatively even in a total white cataract by careful immersion A-scan ultrasound. The visual prognosis, on average, is good since the condition tends to progress slowly over time and is not as

likely as fetal nuclear opacities to cause a severe deprivation amblyopia.

Posterior (and Anterior) Subcapsular Cataracts

Anterior subcapsular cataracts are often associated with trauma, radiation, or acquired diseases such as uveitis, Alport syndrome (cataracts associated with anterior lenticonus), and atopic skin disease (shield-like anterior subcapsular cataracts are classic). Anterior subcapsular opacities may also be part of a more widespread multi-layer cataract.

Posterior subcapsular cataracts are often “crystalline” in appearance, resembling, at times, the look of frosted glass. This type of cataract is seen in association with inflammatory conditions, especially after steroid use or after radiation. It can also be idiopathic. Unlike posterior cortical or posterior polar opacities, posterior subcapsular cataracts are not associated with defects in the posterior capsule. Most are progressive and reduce visual acuity early in the course of progression, especially in bright light.

Persistent Fetal Vasculature (Previously Termed Persistence and Hyperplasia of the Primary Vitreous)

An important and varied type of capsular opacity is associated with PFV. This new term, suggested by Goldberg²¹ in 1997, has replaced the older term, persistence and hyperplasia of the primary vitreous (Fig. 2.36). The cardinal features of this spectrum include a membrane behind, but inseparable from, the posterior lens capsule with blood vessels coursing through it. The membrane is often attached to the ciliary processes, pulling them in toward the center of the pupillary space. A persistent hyaloid artery is present that can, in rare cases, have enough persistent blood flow to cause hemorrhage when cut by the vitrector handpiece at surgery. Posterior retinal involvement portends a poor prognosis for visual outcome. Surgical aggressiveness is usually reserved for those cases with little or no posterior involvement. Mild cases of PFV are also seen, with only a few, less visually significant, findings. Readers should refer to Chapter 34 for more information on PFV.

Traumatic Cataracts

Since trauma is an etiology rather than a morphology, this type of cataract is discussed above in the section on etiology. However, it is also included here since it does not fit into any one anatomical category (Figs. 2.9, 2.34, 2.37, and 2.38). Penetrating trauma most often causes a total cataract since the lens is usually ruptured. Blunt traumas cause cataracts that are variable in severity but often appear plaque-like (Fig. 2.31) and fibrotic, with anterior and posterior subcapsular opacity as well. Zonular disruption is common, as is anterior chamber angle recession. Chapter 35 covers traumatic cataracts in more detail.

Selected Lens Morphology

Congenital Aphakia

Congenital aphakia occurs when the crystalline lens is absent at birth. This finding occurs via primary or secondary paths. A total absence of lens or lens primordia is the mark of the primary manifestation of congenital aphakia. In addition, primary cases also lack an iris and anterior chamber.²² Conversely, if lens development begins in gestation and later becomes disturbed and lens material is obliterated, then secondary congenital aphakia appears. Disturbances such as the rubella virus can cause secondary cases. Stickler syndrome type I also has congenital aphakia in approximately 50% of cases. While only in a minority of cases, Marfan syndrome also carries the possibility of an aphakic phenotype. Further work is ongoing to illuminate the entire spectrum of underlying genetic and molecular factors impacting congenital aphakia.

Microphakia/Spherophakia

When the zonular attachments fail to properly form in development, the lens does not assume its proper shape. A properly formed physiologic lens occupies a generally ellipsoid shape. Part of the formation of this ellipse is the equatorial tension provided by the support structure (the zonule). Without this component, the lens develops into a spherical shape, thus *spherophakia*. Presenting as a singular finding, spherophakia is rare, but it occurs more frequently with other conditions. As Nirankari and Maudgal²³ noted, secondary glaucoma would not be unusual for spherophakic eyes due to a “ball-valve” interaction with the iris to prevent aqueous humor from escaping into the anterior chamber.

Form Abnormalities

Coloboma of the lens occurs also as a result of zonular malformation. It is reported that unilateral cases occur more frequently than bilateral. In a case report by Goel et al.²⁴ phacoemulsification is outlined as a more effective method for managing lensectomy in adult lens coloboma patients with subluxated lenses than manual small incision cataract surgery.

Anterior lenticonus is a form anomaly demonstrated by a conical protrusion from the anterior lens cortex. Often it is associated with Alport syndrome. This collection of findings all relate to a defect in type IV collagen.²⁵ Posterior lentiglobus, as discussed above in the cataract morphology, is a defect on the posterior lens face. Instead of a conical protrusion, as with anterior lenticonus, posterior lentiglobus has more of a bulge than a cone. This pathology presents more often in a unilateral fashion without significant evidence for predictive familial component. Often a weakness in the central posterior capsule is present, and represents one theory for the mechanism for posterior lentiglobus formation.²⁶

Congenital Lens Ectopia

A malpositioned crystalline lens can have several causes. Naturally, it can occur as a singular finding, simple ectopia lentis (Fig. 2.39), but there are several systemic conditions that facilitate an ectopic lens. Marfan syndrome has a constellation of systemic findings in addition to lens subluxation (60%–80% of cases) from zonular weakness. Homocysteinuria, a defect in methionine metabolism with significant systemic complications, has approximately 90% ectopia lentis among untreated individuals due to zonular breaks.²⁷ Ectopia lentis et pupillae is a condition that occurs typically without systemic findings, while other ocular pathology is not unusual. This often-bilateral condition normally presents with pupils and lenses displaced in opposite directions. Genetically, ectopia lentis et pupillae exhibits a strong familial link, though significant variation in expression exists.²⁸

SUMMARY

Congenital, infantile, and juvenile onset cataracts represent important causes of visual impairment in childhood. A thorough ocular and systemic examination will often be needed to uncover valuable information needed to assign the appropriate etiology and develop the best treatment plan for the child. The morphologic features of childhood cataracts are varied. A classification system based on categories commonly used by surgeons and in clinics is presented and recommended. Segregating childhood cataracts in this way may also help predict the prognosis and the risk for later complications.

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3

Epidemiology of Pediatric Cataracts and Associated Blindness

Rupal H. Trivedi and M. Edward Wilson

Cataract is numerically the largest cause of preventable and treatable visual handicap in childhood.¹ Children who are born blind or who become blind after birth have a lifetime of blindness ahead of them that includes all the associated socioeconomic impacts to the child, the family, and the society. Visual defect in childhood is numerically small when compared with blindness at other ages, but since the survivors are likely to live for a long time, when expressed in blindness-years, child blindness becomes very significant. The control of blindness in children is one of the main priorities of Vision 2020: The Right to Sight, the global initiative of the World Health Organization (WHO) and the International Agency for the Prevention of Blindness.²

The major preventable causes of blindness in children (e.g., those causing corneal opacification) are declining in poor countries as a result of large-scale public health interventions, and cataract is becoming a relatively more important avoidable cause.³ Using a standardized classification and coding system, Gilbert et al. evaluated 9,293 children in 40 countries and reported that the lens is responsible for 12% (range, 7%–20%) of anatomical abnormalities in children. Over 200,000 children are blind from disorders of the lens, principally due to unoperated cataract, but also as a result of dense amblyopia following delayed surgery, complications of surgery, or from associated ocular abnormalities.³ Because of the high incidence and treatable nature of the condition, it is reasonable to think that an improved approach to the management of childhood cataracts would have a large impact on childhood blindness as a whole. In this chapter, we describe the epidemiology of childhood cataracts and the blindness associated with them.

PREVALENCE AND INCIDENCE

Prevalence can be defined as the total number of cases of a disease in a given population at a specific time, while the incidence is the number of new cases that develop in

a population during a specified time interval. Several articles have been published that attempt to quantify blindness from childhood cataract. Such studies either describe the frequency of lens opacity irrespective of its impact on vision or report the prevalence of blindness due to lens opacity. The reliability of these studies and their usefulness for comparison are uncertain for various reasons, which include the following:

1. *Noncataractous reasons for blindness.* Most of the studies define blindness as being caused by pediatric cataracts. However, besides cataracts, there are many conditions that can prevent the achievement of normal visual acuity (e.g., amblyopia, secondary opacification of the visual axis, residual refractive error). It should also be noted that lesser degrees of visual loss, visual field defects, or unilateral blindness could also result in significant visual disability in children, which has not been accounted for in most studies.
2. *Varying standards between studies.* Standards for “visual impairment” and “age at childhood cataract” vary between studies. This makes comparison of data difficult. WHO defines the visually impaired child as having a corrected visual acuity of <20/60 in the better eye, severe visual impairment as having a corrected visual acuity of <20/200, and blindness as having a corrected visual acuity of <20/400. Gilbert et al.⁴ tried to improve the overall collection of data by publishing standardized severe visual impairment forms for childhood data collection. Widespread use of these standardized forms will hopefully lead to more meaningful data collection that will allow comparison between studies.
3. *Different methods for collecting data.* Registration data, population surveys, and studies of children in schools for the blind have been found to vary widely in the methods used for data collection. Differences may underestimate or overestimate the severity of disease. Comparisons between these studies are therefore unreliable.

4. *Difficulty in assessing childhood visual function.* Visual function can be difficult to assess accurately in children even under ideal conditions. This can lead to variations between examinations of the same children and unreliable visual function reporting.

Childhood Cataracts

The prevalence of childhood cataracts has been reported as 1 to 15 per 10,000 children.⁵ The wide range is because of the reasons described above: variety of methods, different age groups, and varying case definitions used in the studies, as well as true differences between populations.⁵ Birth prevalence of congenital bilateral cataracts in industrialized countries is 1 to 3 per 10,000 children. Foster, Gilbert and Rahi have calculated that approximately 4 children per million total population per year will be born with bilateral cataracts in industrialized countries, and the figure from developing countries is likely to be 10 children per million total population per year.⁵ The birth defect monitoring program (BDMP) in the United States reported the prevalence of congenital cataracts as 0.8/10,000 births for 1970 to 1987.⁶ A cluster of areas with a significantly high prevalence was found in Michigan in a geographic band from the southwestern to the east-central section of the state. Edmonds and James,⁷ examining the BDMP data, have shown that from 1979/1980 to 1988/1989 there was a 6.8% rate increase (range, 0.7–1.3 per 10,000 births). James⁶ reported the 1988/1989 BDMP/Commission on Professional and Hospital Activities prevalence of 1.2/10,000 births, with the highest rates found in the Northeast (1.7/10,000). The Metropolitan Atlanta Congenital Defects Program surveillance reported a congenital cataract prevalence of 2.1/10,000 live births from 1988 to 1991.⁸ Screening of 2,447 four-year-olds yielded a rate of 7.7 cataracts/10,000 live births,⁹ while two other cohort birth studies suggested a prevalence of 5.3 and 4.4 cataracts/10,000 live births.^{10,11} A prospective collaborative perinatal project conducted by 12 U.S. universities reported the prevalence of infantile cataract as 13.6/10,000 infants.¹² The report further noted that isolated infantile cataracts occurred 3.8 times as often among infants born at weight $\leq 2,500$ g than among those born at $>2,500$ g. In 2003 Holmes et al.¹³ reported, in a retrospective population-based medical record retrieval in the U.S. population, that the birth prevalence of visually significant cataracts was 3.0/10,000 (infantile cataract) to 4.5/10,000 (possible infantile cataract, defined as a cataract diagnosed after the 1st year for which there is no evidence of an acquired etiology). The authors estimated a total of 1,774 cases a year, with a prevalence rate of 4.5/10,000 live births. The Nordic registers of the blind suggest that the prevalence of visual impairment as a result of cataract is 0.6/10,000 children aged 0 to 17 years.¹⁴

The incidence of pediatric cataracts is difficult to ascertain. Wirth et al.¹⁵ have indirectly calculated it for

Australia during their 25-year study (1975–2000). Over these 25 years, 421 index cases with congenital and pediatric cataracts were identified (16.8% a year), giving an incidence of pediatric cataracts in Australia of 2.2/10,000 (i.e., 1 in 4,500). Rahi and Dezateux¹⁶ reported that the adjusted annual age-specific incidence of new diagnosis of congenital and infantile cataract was highest in the first year of life, being 2.49 per 10,000 children. Adjusted cumulative incidence at 5 years was 3.18 per 10,000, increasing to 3.46 per 10,000 by 15 years. A Danish study reported overall cumulative risk of childhood cataract as 10.84 per 10,000 children.¹⁷

Blindness Associated with Childhood Cataracts

Cataract-associated vision loss in these children may be caused by a combination of the lens opacity and other associated findings (e.g., amblyopia, secondary opacification of the visual axis, residual refractive error). Limburg et al. reported that among children with disorders of the lens, almost half had unoperated cataract, whereas the remainder had undergone cataract surgery and had visual loss from amblyopia, the complications of surgery, or other associated ocular abnormalities.¹⁸ The prevalence of blindness from cataracts in children in developing countries is probably 1 to 4/10,000, compared with approximately 0.1 to 0.4/10,000 children in the industrialized world.

Scenario in the Developing World

The higher prevalence of cataract blindness in children of the developing world may be because there is a higher prevalence of cataracts attributed to causes such as congenital rubella syndrome (CRS), consanguinity, and poor nutrition. Limburg et al.¹⁸ noted that the mortality rate of those younger than 5 years declined in recent years. Improved control of measles and vitamin A deficiency improved the preventable blindness due to corneal disease. With that said, eye care services for children should now focus on refractive error, cataract, and control of retinopathy of prematurity.¹⁸ Additionally, clinical services to restore sight or prevent blindness are less well developed in poor countries.

Children with cataract in the developing world also typically present late. Reasons for late presentation include lack of awareness on the part of parents that their child has a treatable condition, and in some countries there is a belief that children who are born blind cannot have their sight restored. General physicians or community health workers, who are often the first point of contact, may tell parents that their child is too young for surgery, or that the cataract needs to mature. Delays from disease recognition to disease presentation are common in the developing world. Mean delays of 39 and 44 months were reported in two studies from Tanzania.^{19,20}

Similar delays have been reported from East Africa.²¹ A study from China reported on 196 children (309 eyes) with congenital or developmental cataracts. The mean delay from cataract recognition to presentation for surgery was 49.6 months.²² The authors reported that most infants with dense bilateral congenital cataracts were not referred to them until after 6 months of age. However, during the 10 years of the study, the delay in presentation showed a steady decline as caregivers began to recognize the benefits of early referral. Even with delayed presentation (defined as >18 months between detection and presentation for surgery), surgery helps to regain functional vision, which can be used for navigation and low vision aids.²³ In the developing world setting, barriers to the uptake of surgical care for childhood cataract include fear of surgery or of a poor outcome, beliefs that congenital blindness cannot be cured, lack of information on available services, and poorly educated mothers—all of which need to be addressed through counseling of parents. More training of health worker is also needed, as they often do not recognize the urgency of cataract in children and give parents the wrong advice.²⁴

In many poorly developed countries, boys present for cataract more frequently than girls,^{3,25} despite there being no evidence that there are significant gender differences in the incidence.¹⁶ Poor parents are more willing to use their scarce resources for health care for their sons rather than for their daughters. Gender inequity, documented in the baseline data, was eliminated in the postintervention period by counseling and tracking system that recorded each child's contact information (including cell phone number) for parents or neighbor.²⁶

CONTROL OF CATARACT BLINDNESS IN CHILDREN

Pediatric cataract-associated blindness can be avoided or treated using a combination of preventive services at the community level, specialized surgical services in pediatric ophthalmic units, and low-vision devices and services. Primary prevention is currently limited to avoidance, where possible, of known teratogens. CRS is the most important preventable cause of congenital cataracts. In 2003, one article estimated that at least 100,000 infants are born each year with CRS.²⁷ This occurs in spite of the fact that CRS has virtually been eliminated in the United States by childhood immunization programs. Rubella infection (German measles) is usually a mild illness characterized by rash, fever, and lymphadenopathy and it primarily affects children. The immunoglobulin G (IgG) antibody response confers protection against subsequent infection. Immunization for measles, mumps, and rubella (MMR) are now usually given to young children in the developed world to better assure population immunity. Although the rubella

infection is mild, a woman who becomes infected during her first 10 weeks of pregnancy has a 90% likelihood that the fetus will become infected, leading to premature fetal death or CRS. Infants born with CRS may have microphthalmia with cataracts and glaucoma. In addition, deafness, cardiovascular disease, microcephaly, and developmental delay are common.

Immunization strategies for the developing world included selective targeting of sero-negative women early in their childbearing age or the strategy of immunizing all male and female infants (to give herd immunity). This is a more complicated decision that might be evident on the surface. When infant immunization programs begin, coverage is usually enough to prevent the cyclical epidemics (about every 4 years) which produce much of the natural acquired immunity in children. However, if the immunization coverage is not complete, the proportion of women of childbearing age who are susceptible can actually increase and the incidence of CRS can go up. Success can only be assured in the face of ongoing monitoring for vaccine coverage and surveillance for rubella infection and CRS.²⁸

Early detection of cataract (regardless of cause) is essential in ensuring that treatment, together with parental advice and support, can be provided promptly. Consequently, in many industrial countries routine examination of the red-reflex of newborn children is an established practice. Most developing nations provide health delivery services through a tiered system, with central hospitals supporting smaller rural hospitals and health delivery centers. In Africa and Asia, for example, many countries have established a three-tiered system, consisting of primary, secondary, and tertiary levels. The goal is to restore sight by early detection and evaluation of blind children and early surgical referral of children with cataract. For those detected early, well equipped pediatric operating facilities are needed to assure good-quality cataract surgery followed by appropriate optical correction and regular, long-term follow-up care.

Primary Level of Care

Primary eye care includes services provided by trained community health workers. Their main responsibility is to prevent blindness from occurring. Personnel trained in primary eye care are essential for the control of blindness in children. Primary care providers should

- provide ophthalmoscopic screening of neonates and identify patients who need referrals;
- refer such patients for ophthalmologic assessment and treatment;
- encourage and motivate parents and/or children regarding patching, when suggested by secondary and/or tertiary eye care providers;
- provide services for immunization;

- encourage long-term ocular follow-up;
- provide counseling to avoid consensual marriages between families with histories of childhood cataracts;
- educate people to use preventive measures to avoid sports-related and other ocular trauma.

Lack of primary level of eye care in developing world means that children with cataracts are not being detected early and visual outcomes are being compromised as a consequence.²⁴

Secondary Level of Care

At the secondary level, the main responsibility is to maintain or improve functional visual outcome. The eye surgeon should

- be able to carry out a full eye examination and assessment and make a provisional diagnosis;
- arrange surgery for the affected eye at the tertiary center;
- follow up postoperative eyes to detect secondary opacities;
- work as a joining link between primary-level care and tertiary centers;
- help patients to follow patching instructions;
- effectively communicate with parents to ensure their involvement;
- arrange screening for children in their schools.

Tertiary Level of Care

Tertiary care services should include facilities for the trained ophthalmologist, optometrist, anesthetist, pediatrician, and neonatologist. Their main responsibility is to restore vision to patients blinded by anatomical and/or functional causes. The central hospital, usually attached to a medical school, is the tertiary resource. There may be several tertiary hospitals in larger countries serving large geographic regions. This facility usually is a large general hospital and offers a wide range of specialty services. Although it consumes a high proportion of the health budget, it often is overwhelmed by the demand for services. The tertiary eye center provides more sophisticated eye care than that available at the provincial hospital, and ophthalmic subspecialists.

Tertiary care centers should

- be able to provide surgical services within well-equipped centers containing vitrectomy instruments, high-viscosity viscoelastic agents, and high-quality intraocular lens material;
- organize and provide low-vision services;
- responsibly participate in research;

- train the faculty of primary- and secondary-level programs;
- support, supervise, motivate, and provide feedback to staff at secondary-level centers;
- correct residual refractive error when needed;
- provide long-term regular follow-up with assessment and treatment of posterior capsule opacification, glaucoma, refractive error, and amblyopia;
- provide guidance to improve the infrastructure and technology that will ensure the development of low-cost, high-quality, low-vision devices.

SUMMARY

Globally, an estimated 200,000 children are bilaterally blind from cataract. Many more suffer from partial cataracts that progress and cause increasing visual difficulty as the child ages. Exact epidemiologic data for children of all ages are difficult to gather. However, because of the number of blind years presented, treatment of childhood cataracts is among the most cost-effective intervention in all of ophthalmology. Management of pediatric cataracts is often difficult and tedious, and it requires a dedicated team effort by the parents, pediatrician, surgeon, anesthesiologist, orthoptist, and community health worker. We should concentrate on not only improving but also maintaining both anatomical and functional outcomes in eyes with pediatric cataracts and prepare for lifelong follow-up.

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4

Role of Vision Screening in the Detection of Congenital or Early Acquired Cataracts

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Pediatric cataract remains an important form of treatable childhood blindness and lifelong visual impairment. There are an estimated 200,000 children blind from cataract worldwide.¹ Thus congenital cataract, as a treatable cause of visual handicap in childhood, is a priority of the global Vision 2020 initiative.² Successful outcomes require the early recognition of infantile and congenital cataracts. Children are a vulnerable population because, unlike adults who note a loss of vision, children are usually unaware of and do not complain of their deficit. Detection of pediatric cataracts is carried out by caretakers and primary care providers as part of a general childhood health screen. Tools used by primary care givers range from observation of the eyes and visual function to acuity measurement. By necessity, these tools change as the child grows and develops. This chapter reviews the importance of screening and useful techniques in detection of childhood cataracts.

DETECTION IN INFANTS

Of primary concern is the detection of cataracts in infants because of the risk of devastating amblyopia resulting in a lifetime of visual impairment. Successful surgical repair and rehabilitation must be instituted early to allow development of normal vision. Significant lens opacity during the critical period of visual development results in poor vision and, in bilateral cases, sensory nystagmus that usually persists even after excellent surgical and restorative care. In visually significant newborn cases, cataract surgery within 6 weeks of birth produces the best outcome. Sensory nystagmus will develop if dense bilateral cataracts are not cleared before at least 3 months of age (see Chapter 6). Therefore, screening for pediatric cataract is imperative as part of the initial newborn as well as subsequent well-baby exams.

A Swedish study found that 80% of congenital cataracts were detected when screening was performed in the hospital maternity ward, compared with 67% of cases when screening was deferred to the first well-baby check. Three children per year were operated on too late and thus missed the opportunity for good visual development in the future.³ Thus, it is important that newborns undergo screening for congenital cataracts within the first few days of life with subsequent screenings at a few weeks of age.

THE RED REFLEX TEST

The American Academy of Pediatrics (AAP) states that “red reflex testing is an essential component of the neonatal, infant, and child physical examination” and has published the practice pattern guidelines and techniques of this test. The AAP recommends all children undergo a red reflex assessment as a component of the eye evaluation in the newborn nursery and during all subsequent routine health supervision visits.⁴ The test should be performed in a darkened room (to maximize pupil dilation). The direct ophthalmoscope is focused on each pupil individually approximately 12 to 18 inches away from the eyes. As a second stage, both eyes are viewed simultaneously approximately 3 feet away with the child focusing on the ophthalmoscope light (Fig. 4.1). The red reflex seen in each eye individually should be bright reddish-yellow in lightly pigmented eyes or grey in darkly pigmented, brown-eyed patients, and identical in both eyes. Dark spots in the red reflex that do not move with a blink, a blunted dull red reflex, lack of a red reflex, a white reflex, or asymmetry of the reflexes (Bruckner test) are all indications for referral (Fig. 4.2). The *See Red* card is available to provide reference for this test (Fig. 4.3).⁵



Figure 4.1. Symmetric red reflexes in an infant.

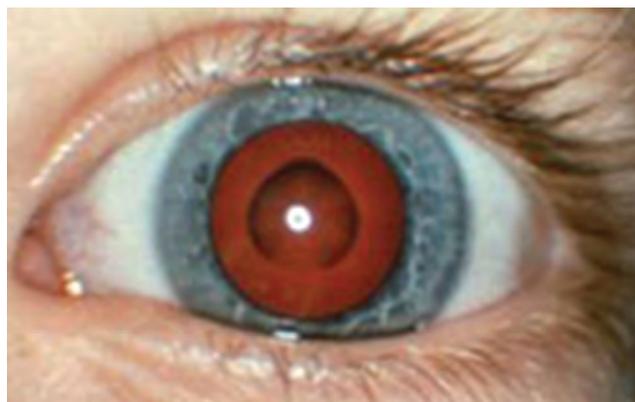


Figure 4.2. Cataract opacity visible in red reflex.

Any question as to the red reflex test findings should warrant expedited referral. Historically, poor communication between eye care providers and primary care doctors has hampered the effectiveness of vision screening.⁶ Effective communication between the primary care giver and ophthalmic health professionals is critical to the success of this approach and the care of the child.

Medical training and public policy initiatives designed to decrease childhood blindness need to continue to emphasize the importance of the simple red reflex test at the routine newborn and 6- to 8-week examinations. Instruction to primary care givers should include this screening tool in both the developed and developing world. In Britain, where the cataract incidence by age 1 year was found to be 2.49/10,000, only 47% of infantile

cataracts were detected through screening examinations in the first 3 months of life.^{7,8} In Israel, where less than half the neonatology departments performed the red reflex test routinely, it was found that the addition of the “efficient” red reflex test increased the detection of infantile cataracts.⁹ In developing nations, where cataracts account for 10% to 30% of childhood blindness,^{10,11} detection with an inexpensive ophthalmoscope in the hands of a properly trained primary care giver, combined with access to care, will allow more children to see and become fully functional members of society.

Red reflex examination.

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See
RED

Red reflexes from the retina can be used by the physician to assess ocular health. The location and shape of the red reflex, the equality of the red reflexes on the maculae with the red reflexes in various positions. The white dots represent corneal light reflexes.

Technique: Both the ophthalmologist and the parent stand in front of the child with the child seated in the parent's lap. Ask the child to look at the light source. Encourage the child to look at the light source. Compare the red reflex from each eye. Both red reflexes should be viewed simultaneously and symmetrically. An asymmetric reflex is the problem of the white reflex, the corneal light reflex.

The beauty of this test is that it can be done with a “hands-off” approach. It can be done with an ophthalmoscope or a screening device. It is very easy to do. The technique is to work with the technique. It is useful for detecting all other ocular conditions such as strabismus, nystagmus, and eyelid anomalies.

- ↓ **NORMAL**—Child looks at light. Both red reflexes are equal.
- ↓ **UNEQUAL REFRACTION**—One red reflex is brighter than the other.
- ↓ **NO REFLEX (CATARACT)**—The presence of lens or other media opacities blocks the red reflex or diminishes it.
- ↓ **FOREIGN BODY ABRASION (LEFT CORNEA)**—The red reflex from the pupil will be both light colored defects or foreign bodies. Movement of the examiner's head in one direction will appear to move the corneal defects in the opposite direction. (Frankel) 2004
- ↓ **STRABISMUS**—The red reflex is more intense from the deviated eye.

et al. *Pediatrics* 2008; 122: 1401-1404

DETECTION IN CHILDHOOD

Cataracts may develop as a child grows due to family history, metabolic disease, trauma, and other causes. Assessment for eye pathology, including the red reflex test, and visual function should occur at all subsequent routine health supervision visits in early childhood.⁵ The American Academy of Ophthalmology has developed guidelines to be used by physicians, nurses, educational institutions, public health departments, and other professionals who perform vision evaluation services.¹² By age 4 months, a cooperative child should fixate a visual target and follow it smoothly. The assessment should be performed binocularly and then monocularly. If poor visual fixation and following is noted binocularly after 3 months of age, referral is needed.

Visual acuity testing may be possible as early as age 2 or 3 years old. Acuity tests should be repeated at age 3, 4, and 5 and repeated every 1 to 2 years thereafter. AAP recommends these screenings at well-child visits.⁴ Screenings are also carried out in preschools and schools, and community events by nurses and trained laypersons. Picture tests, such as LEA symbols[®] and Allen cards, or the HOTV test (a letter-matching test involving these four letters) can be used for children 2 to 4 years of age. Tests for children older than 4 years include wall charts containing Snellen letters or numbers. Monocular cataracts often have no signs, as the child seems to see normally. Therefore, visual acuity testing must be carried out with good monocular occlusion in a cooperative child.

AUTOMATED SCREENERS

Increasingly, automated screeners are allowing early detection of children's ocular pathology. Pediatric lens pathology has been documented and monitored using the MTI Polaroid photoscreener.¹³ Over the last decade, new autorefractors and photoscreeners have been developed and marketed as pediatric vision screeners.¹⁴ The computerized optics frequently allow quick assessment of ocular media, gaze fixation, ocular alignment, and refractive error, facilitating earlier detection of visual problems in children (Fig. 4.4). Screenings performed with the MTI photoscreener in statewide preschool screenings were able to detect cataracts in 0.7% (in Alaska) and 0.2% (in Tennessee) of screened children.¹⁵ Typically, these machines will be unable to "read" from an eye with a moderate or large lens opacity. Thus, any child unable to test with an automated screener is at risk for having a cataract and should be referred for evaluation.

REFERRAL FOR HIGH-RISK CHILDREN

While red reflex and pediatric vision screenings are essential for detecting cataracts in the general population, children at high risk for cataracts should undergo



Figure 4.4. Screening with an automated screener.

ophthalmologic evaluation regardless of the status of the red reflex.⁴ Prematurity, growth retardation, neurologic disorders, or craniofacial abnormalities may be associated with lens opacities and these children should be referred by the primary care provider for an eye examination. Infants who fail newborn screenings for systemic diseases should be evaluated as part of their workup. For instance, those children found to have galactokinase deficiency will respond better to dietary treatment if instituted early.¹⁶ One percent of infants diagnosed with congenital toxoplasmosis were found to have cataracts.¹⁷ As well, any child with a family history of cataracts at an early age should undergo a comprehensive examination expeditiously by an ophthalmologist experienced in treating children. Genetic markers are now available for some heritable forms of cataract.¹⁸ Systemic diseases associated with eye abnormalities, such as juvenile idiopathic arthritis, or chronic steroid use warrant a full examination based upon recommended criteria.¹⁹

SUMMARY

Through careful history and multiple age-appropriate vision screenings, including a properly performed red reflex test, pediatric cataracts may be detected earlier leading to improved visual outcomes.

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5

Preoperative Issues

Rupal H. Trivedi and M. Edward Wilson

A thorough preoperative evaluation sets the stage for the decision making that precedes surgery.¹ A comprehensive history and an ocular and systemic examination help the care team to plan the overall management of the child with a cataract. The goals of the preoperative assessment include deciding whether surgery is needed, and if needed, what the appropriate timing of the surgery is. In addition, the characteristics of the cataract are documented and the postoperative visual prognosis estimated. During the assessment, we also often gain insight into whether the child and the family will comply with the postoperative correction of residual refractive error and amblyopia treatment. These data, together, help us decide whether it is best to implant an intraocular lens (IOL) or not, and if implanted, what postoperative refractive error should be aimed for when selecting an IOL power.

IMPORTANT DECISIONS

During history taking and examination, the physician needs to make several important decisions. Among these are whether surgery is indicated or not, and if indicated, how to handle aphakic rehabilitation, etc.

Indication for Surgery

The surgeon should be careful to operate on only those cataracts where the visual disturbance is severe enough to justify sacrificing normal youthful accommodation. Indications for cataract surgery include a cataract that obstructs the examiner's view of the fundus of the nondilated pupil or a blackened retinoscopic reflex preventing refraction of the patient. Deciding when to remove a partial cataract can be difficult. Nonverbal children add more difficulties to this decision. In each individual case, the ophthalmologist needs to use his or her best judgment about whether a partial cataract is interfering with visual functioning enough to warrant removal. Partial cataracts can be amblyogenic and may disturb emmetropization leading to abnormal axial elongation. For verbal children, cataract surgery is contemplated if Snellen visual acuity (VA) is 20/50 or worse or

if the child is intolerant to glare or resistant to amblyopia therapy with documented deteriorating visual function. Since a subjective VA cannot be obtained in infants with cataracts, greater reliance is placed on the morphology of the cataract, other associated ocular findings, and the visual behavior of the child, in order to ascertain whether the cataract is visually significant or not. The degree of visual impairment induced by a lens opacity differs markedly depending on the location of the opacity. Generally, the more posterior and the more central the opacity, the more amblyogenic it is. Generally speaking, a cataract that blackens the retinoscopic reflex for 3 mm or more in the center of the pupil is considered visually significant.

If a partial cataract is being treated conservatively, it is important to carefully follow these children. Conservative treatment using mydriatic drops necessitates the patient's wearing glasses for reading if any cycloplegic effect is induced. This has not found widespread acceptance. Associated glare and loss of accommodation are the most common obstacles. Visual outcome has also been unimpressive. Despite these limitations, the use of mydriatic drops may be kept in reserve in eyes with slowly progressive cataracts or paracentral cataracts <3 mm and, especially, in patients for whom cataract surgery needs to be deferred for any reason—be it medical (high risk for anesthesia), social, or economical.

Timing of Surgery

Deciding on the appropriate timing of surgery is most critical during early infancy. In the case of a unilateral dense cataract diagnosed at birth, the surgeon can wait until 4 to 6 weeks of age. Waiting until 30 days of age or more decreases the anesthesia-related risks and often allows term infants to be healthy enough for discharge to home after surgery. Premature babies or term infants operated before 30 days of age are usually kept overnight for observation since the incidence of apnea after anesthesia is higher. For dense unilateral cataracts documented to be visually significant at birth, waiting beyond 6 weeks may adversely affect visual outcome.^{2,3} In the case

of a bilateral cataract diagnosed at birth, a good visual outcome can be achieved if the child is operated before 10 weeks of age.⁴ The first eye surgery can be offered at 4 to 6 weeks of age, and the second eye surgery after another 1 to 2 weeks' time. It is important to keep the time interval to a minimum between the two eye surgeries. Some surgeons advise patching the first operated eye until the second has had surgery, to prevent amblyopia in the second operated eye.⁵ This type of occlusion is not commonly done but undue delays between surgeries should be avoided in infants. For older children, the timing of surgery is not as crucial. In children beyond the amblyopic age, surgery can often be decided based on convenience and other logistical issues.

Sequential cataract surgery, more popularly known as immediately sequential bilateral cataract surgery (ISBCS), remains controversial (see Chapter 9). Almost every discussion on ISBCS either starts or ends with a comment on the disagreement surrounding its use. *The important question is not "can it be done?" but, more properly, "should it be done?"* Even conservative surgeons, who vote against the routine use of ISBCS in children, are more likely to use this approach when anesthesia poses more than average risks or if the patient lives far away and a visit for surgery on the second eye would be difficult.

Timing of surgery in children with traumatic cataract, uveitis, and retinoblastoma is discussed in appropriate chapters (see Chapters 35, 38, and 40).

Aphakic Rehabilitation

IOL implantation in children has the benefit of reducing dependency on compliance with other external optical devices (aphakic glasses and contact lenses) and providing at least a partial optical correction constantly. These are important advantages to the visual development in amblyopia-prone eyes. However, concerns about primary IOL implantation are the technical difficulties of implanting an IOL in the eyes of small children, selecting an appropriate IOL power, and the risk of visual axis opacification (VAO) after implantation in the very young.⁶ On the other hand, although it is possible for an eye with a unilateral infantile cataract to achieve a good visual outcome following contact lens correction, it requires cooperation from children. Both IOLs and aphakic contact lenses may support similar VA after surgery for unilateral cataract in the presence of good compliance with contact lens. However, IOLs support better VA when compliance with contact lens wear is moderate or poor.⁷ For bilateral cataracts, aphakic glasses or contact lens use are reasonable options. Infant aphakia treatment study⁶ concluded that until longer-term follow-up data are available, caution should be exercised when performing primary IOL implantation in children aged 7 months or younger given the higher incidence of adverse events and the absence of short-term visual outcome compared with the contact

lens use. For children beyond infancy, IOL implantation is less controversial and more commonly employed.

HISTORY

Parent/caregivers may present with a chief complaint of noticing a white spot in the child's eye (Fig. 5.1A–C), visual inattentiveness, nystagmus, strabismus, asymmetry of one eye relative to the other (e.g., microphthalmos), photophobia, ocular injury, or simply because of referral from other physician who has identified a possible lens opacity. At times, the evaluation is scheduled because of a family history of childhood cataracts or because the child has one of a growing number of systemic conditions or syndromes that can be associated with cataracts. Specific information is gathered on gender, ethnicity, and date of birth; birth weight; evidence of maternal infection (especially the TORCH infections), rash or febrile illness during pregnancy (may be suggestive of intrauterine infection), any other prenatal and perinatal history that may be pertinent (e.g., alcohol, tobacco, drug use, ionizing radiation during pregnancy), history of ocular trauma (unless cataract appears to be purely nontraumatic), age at onset of visual symptoms, ocular status on previous eye examinations (can be helpful in assessing visual prognosis after treatment), and history of corticosteroid therapy (especially in posterior subcapsular cataract [PSC]).¹ Simple questions can help in determining the surgical need, the timing or urgency of surgery, and the visual prognosis after cataract removal (e.g., Does your child appear to see well? Do your child's eyes look straight or do they seem to cross or drift or seem lazy? How long have you noticed a change in your child's visual function?). Frequently, even with poor vision, a child may be functioning reasonably well in a familiar environment. The child will be reluctant, however, to explore an unfamiliar area. Ask parents how well their child functions in a new environment, which is a useful indicator of vision. Infants with complete bilateral congenital cataracts usually demonstrate decreased visual interest and experience delayed milestones.

Approximately one-third of cataracts are inherited, so family history of childhood cataract should be evaluated (especially in bilateral cataract). Mention to the parents that you may want to look at their eyes (Fig. 5.2). Explain that a finding in the eyes of the parents may help streamline the evaluation and workup of the child, thus preventing unnecessary tests.

EXAMINATION

Art of Examining Children

A friendly manner, a little trickery, and a lot of praise are essential elements in the art of examining children.⁸ Pediatric exam is not as methodical and sequenced as the adult exam. Since children are not merely small adults,



Figure 5.1 A-C. Cataract presented as white reflex. (Reprinted from Trivedi RH, Wilson ME. Pediatric cataract: preoperative issues and considerations. In: Wilson ME, Saunders RA, Trivedi RH, eds. *Pediatric Ophthalmology: Current Thought and a Practical Guide*. Berlin, Heidelberg: Springer, 2009, with permission.)

the temptation to proceed methodically and sequentially through each portion of the complete eye exam in each patient needs to be resisted. Remember that the doctor does not decide when the exam is over, the child does. Use the limited attention span and cooperation of the child to perform the investigations most essential. If child is not cooperative in office, most examinations can be performed during examinations under anesthesia (EUA) before surgery. The doctor's initial behavior should be aimed at establishing trust. The doctor should be seated, so as not

to stand over the child. Invite the child to sit in the BIG chair on a parent's lap or alone. Raise the chair quickly so that the child is at least at eye level with everyone in the room. Don't surprise the child. Talk directly to the child. For older children, comment on his or her clothing or ask a question you know he/she can answer, such as: How old are you? What grade are you in? What are you doing this summer? When a child begins to speak, his/her anxiety level drops dramatically. It is also helpful to show the child a toy and let the child hold it. Tell the child what to do. Be animated. Have colorful toys. Whistle, make noises, and call the child by name. Use an age-appropriate vocabulary (e.g., Phoropter = elephant glasses). During the course of the exam, pause whenever you need a break or the child "demands" one. Remember, the child and parent will detect any hint of frustration in your voice. Praise children for being "grown-up," for doing great, and for having amazing eyes. Throw a few "almost done" comments to keep them cooperative. More praise can be done at the end, even if they did not do as well as you hoped. Patients with pediatric cataract require repeated follow-up examinations. It is important to have the child feel reasonably good about the clinic visit, which may help during the next clinic visit.

Children dislike the drops and they can get cranky after waiting for dilated eye examination. We do dilated eye examination during EUA just before surgery. If pupil dilation is needed in the clinic, have someone other than the examiner place the drops and make sure they are preceded with a topical anesthetic. This too can be done

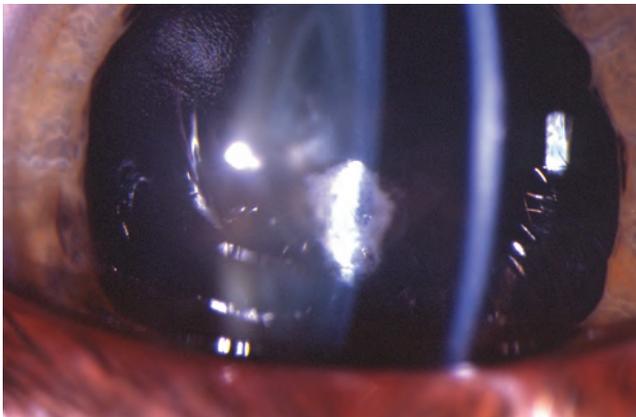


Figure 5.2. Unoperated cataract observed in mother's eye. The child was operated for cataract. (Reprinted from Trivedi RH, Wilson ME. Pediatric cataract: preoperative issues and considerations. In: Wilson ME, Saunders RA, Trivedi RH, eds. *Pediatric Ophthalmology: Current Thought and a Practical Guide*. Berlin, Heidelberg: Springer, 2009, with permission.)

quickly so the child can be consoled by the parent and can retreat to the comfort of a playroom or a toy-filled sub-waiting area. If fundus examination is done in the clinic, it helps to convey to children that you are putting on a “strange hat” that can look all the way into their “brains.” Then as you view the optic nerve and macula, praise the children for being smart since they have “lot of brains.” Be efficient by getting the look you need as quickly as possible. Pediatric eye exams are done as a team. The need to see more patients in less time has eliminated the luxury of having the pediatric ophthalmologist perform the entire exam him/herself. Pediatric ophthalmology is no longer a solo sport; it is a team game. Train the team so that everyone adopts the same child-friendly habits.

Assessment of Visual Function

The method of evaluating visual function will vary according to the age of the child and the level of cooperation. A communication barrier exists between the ophthalmologist and very young preverbal or preliterate patients. Parents and caregivers can help to break this communication barrier by working together to encourage the child to perform functional testing in the office that can help quantify the VA of the child with cataract as precisely as possible. Documentation of the child’s level of cooperation with the examination can be useful in interpreting the results and in making comparisons among the examinations over time. When results are equivocal, repeated office visits may be needed. We use an occlusive patch to isolate one eye for VA testing. Children are experts at peaking around an occluder. We tell the child that they can take the patch off as soon as the testing is completed. That seems to comfort them somewhat. They know the patch is temporary.

Infant and Preverbal Child

The assessment strategy is to determine whether each eye can fixate on an object, maintain fixation, and then follow the object into all directions. The assessment should be performed binocularly and then monocularly. This can be done by drawing the child’s attention to the examiner’s or family member’s face (infants under 3 months) or a hand-held toy either at 1/3 m or any other measured distance (1 m, 3 m/10 feet, 6 m/20 feet). The force with which the child objects to alternate occlusion of the eyes is useful to judge the relative vision in each eye. Fixation behavior is recorded for each eye as “fixates, follows, maintains.” Some prefer the terms “central, steady, and maintained” to describe the fixation observed. In an awake and alert child, if poor fixation and following are noted binocularly after 3 to 4 months of age, a significant visual loss is suspected, and searched for.

For strabismic children, an assessment of binocular fixation pattern is performed in which the examiner determines the length of time that the nonpreferred eye can

hold fixation. It can be reported as *will not hold fixation with nonpreferred eye, holds fixation briefly with nonpreferred eye, or no fixation preference*. With a straight-eyed child and those with small angle deviation, the base-down prism-induced tropia fixation test can be used to optically separate the two eyes. A 20 prism-diopter base-down prism is our preferred instrument since it displaces the pupillary reflex and the image sufficient to detect which eye is being used for fixation at any time during testing (Fig. 5.3A–C). The prism is placed before one eye at a time for approximately 2 seconds and the fixation response is described. A scoring system can be used where the responses to the base-down prism are noted using a –2 to +2 scale. In this system, the right eye receives the prism first. A score of –2 means that the child fixates with the left eye only, –1 is when the child alternates at times but prefers OS, 0 indicates alternate fixation without preference, +1 means that the child alternates at times but prefers OD, and +2 indicates that the child fixates with the right eye only. The prism is then placed over the left eye, and it is scored using the same scale. The total induced tropia test (ITT) score is the sum of the grades of the right eye and the left eye. Scores of ± 3 or ± 4 indicate a strong fixation preference and probable poor vision, whereas scores of 0 or ± 1 indicate little or no fixation preference and probably better vision. The results of the ITT can be recorded simply as alternates or the preferred eye is the right/left and nonpreferred eye holds well, holds briefly, or shows no hold. However, the quantitative scoring system is helpful to monitor early postoperative surgical outcome and in subsequent amblyopia management.⁹

In preverbal children with partial cataracts or lamellar cataracts, we also use foot-pedal-operated “barking dogs” or other noise-making and moving toys at the distance within the examination room. Each toy needs to be mounted on a shelf and displaced vertically from one another (Fig. 5.4A–C). As the examiner activates one toy and then another using the foot-pedal switch, the quality of the saccade from the child is noted. At 20 feet, a brisk saccade to the activated toy from the one simultaneously inactivated toy represents sufficient visual function in that eye to place into question the need for surgery on a partial cataract.

Other quantitative methods such as preferential looking techniques (Teller acuity cards or Lea paddles) or Cardiff cards can be used. The Hiding Heidi test by Lea is also useful to detect loss of vision in the lower contrast setting. It works on the preferential looking principle. A sweep visual evoked response can also quantify VA, but it may not always be readily available. Candy sprinkles can be used on a tray to get additional information about the child’s functional vision. If the child can easily find a small candy sprinkle, this illustrates an ability to localize a small object. Watching the child struggle or fail this task can also serve to help verify the significance of the lens opacity.



Figure 5.3 A-C. Base-down prism test. (Reprinted from Trivedi RH, Wilson ME. Pediatric cataract: preoperative issues and considerations. In: Wilson ME, Saunders RA, Trivedi RH, eds. *Pediatric Ophthalmology: Current Thought and a Practical Guide*. Berlin, Heidelberg: Springer, 2009, with permission.)

Verbal Child

Quantitative VA assessment in cooperative verbal children can be assessed using optotype VA testing (identifying or matching symbols or letters), allowing quantification of

VA on a Snellen or preferably, a logMAR scale. Distance VA should be determined monocularly whenever possible. The fellow eye should be completely covered (with adhesive occluder to prevent peeking). The test should

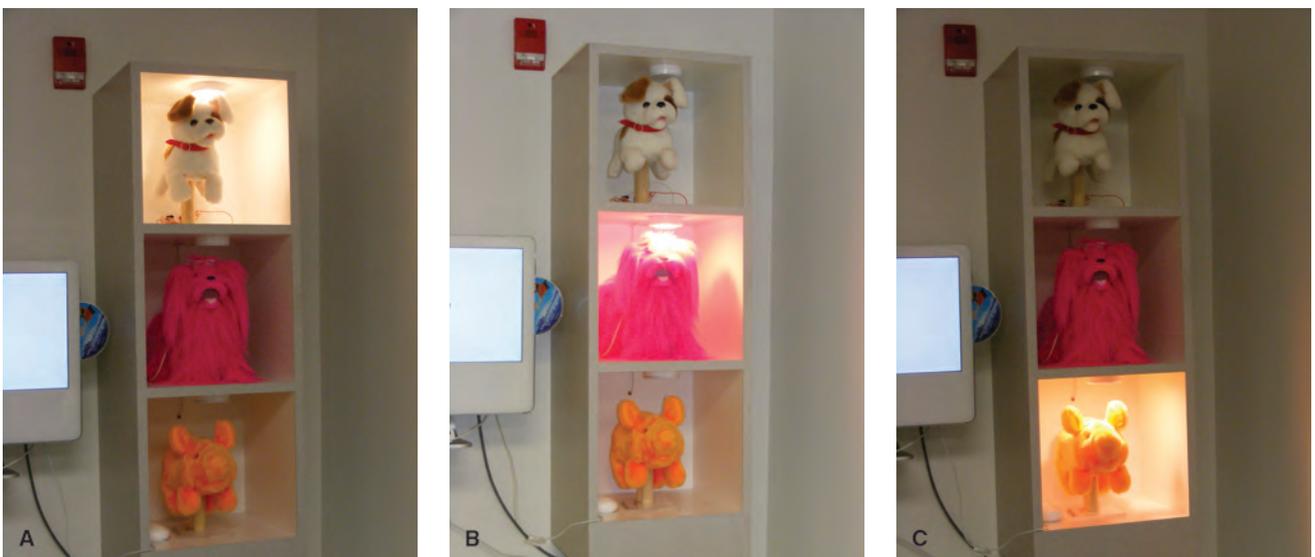


Figure 5.4. **A:** The upper foot-pedal-operated animal is activated. **B:** The middle foot-pedal-operated animal is activated. **C:** The lower foot-pedal-operated animal is activated. (Reprinted from Wilson ME. The art and science of examining a child. In: Wilson ME, Saunders RA, Trivedi RH, eds. *Pediatric Ophthalmology: Current Thought and a Practical Guide*. Berlin, Heidelberg: Springer, 2009, with permission.)

be performed at a distance of 10 to 20 feet with the eye chart calibrated to the exact measured distance from the examination chair to the chart. Children should be tested using a linear display of letters if possible. Young children may be easier to test if isolated optotypes with crowding bars are used. We like the HOTV or the Lea symbols matching game since the letters/symbols can be called by name or identified by pointing to the matching letter/symbol on the lap card.¹ The letters HOTV are chosen because they avoid right to left confusion by being mirror-image letters. In addition to VA, the testing distance, type of optotype, whether the optotype is presented a line at a time or isolated, and cooperation level of the child should be documented in the medical record. If the child returns for repeat testing, the weaker eye should be tested first. Young children have short attention spans and may perform better on the first eye tested and lose interest more quickly on the second eye. Also, when testing VA at near, the testing distance must be controlled precisely. We prefer the HOTV or Lea near chart with a rope attached so that the testing distance can be easily verified and maintained. Other near cards may give falsely good VA measurements when the child leans in and reduces the testing distance to a fraction of the distance to which the chart was calibrated.

Fusion and stereoacuity testing at distance as well as near may also be helpful when deciding how much visual dysfunction is present in a cataract patient. In children with PSCs who complain of intolerable glare, but have good Snellen VA, glare testing should be performed to evaluate the need for surgery.

VA is a useful means of assessing one aspect of visual function, but it provides a very limited indication of the patient's visual perception. Most objects in the visual scene are not at the high black–white contrasts employed in VA testing. Assessment of the patient's ability to detect stimuli at lower contrasts is very useful. Tests such as low-contrast test pictures in infancy and childhood provide important information about the distance at which the child can see facial features. Contrast sensitivity measurements can first be performed using the enhancement game, that is, single low-contrast symbols.¹⁰ Low-contrast optotype tests are either VA charts at low-contrast or tests with one symbol size at different contrast levels. Individuals with equal VA at high contrast may have different low contrast VA.

Red Reflex Test

The red reflex test can be used to detect the density and extent of the opacity in the visual axis. The retinoscope is a very useful instrument for viewing the red reflex within the pupillary space to see how much of the reflex is blackened by the cataract. In addition, the direct ophthalmoscope can be used to perform the Bruchner red reflex test where both eyes are viewed together and the red reflexes compared. When both eyes are viewed simultaneously, potentially amblyogenic conditions, such as anisometropia,

strabismus, and asymmetric cataracts can be identified. The direct ophthalmoscope is focused on both eyes simultaneously at approximately 3 feet away.

Ocular Alignment and Motility

Details on strabismus and nystagmus mainly help when explaining prognosis to the parents and to prepare them for patching or further surgeries. Ocular alignment is assessed by comparing the corneal light reflection of one eye with the other, looking for asymmetry between the eyes using the binocular red reflex test, and by the cover/uncover and alternate cover tests. Cover/uncover and alternate cover tests are performed in primary gaze at distance and at near. Detailed targets that require precise accommodation are utilized when feasible. These tests require the patient's cooperation and interaction with the examiner in addition to sufficient vision to fixate on the target. Earlier-onset unilateral cataracts have the highest risk for strabismus and late-onset bilateral cataracts have the least risk. Also, as a general rule, patients with partial cataracts and relatively good preoperative VA have less strabismus. Strabismus at presentation is often an indication that the cataract is long standing and that significant amblyopia is likely to be present. Infants with profound bilateral dense cataracts develop nystagmus at approximately 3 months of age because the fixation reflex that normally appears by that time is prevented from developing. Once nystagmus has developed, it is likely to persist even if the cataracts are subsequently removed. On occasion, nystagmus will disappear when surgery is performed within weeks to months after onset of the nystagmus. However, this is the exception rather than the rule. When manifest nystagmus does develop and persist, the visual prognosis is worse. VA in eyes with nystagmus and infantile cataracts is rarely better than 20/100 after cataract surgery.

EXTERNAL EXAMINATION, ANTERIOR SEGMENT EVALUATION, AND OTHER INVESTIGATIONS

External examination of the eye with a suspected cataract usually consists of a penlight evaluation of eyelids, eyelashes, conjunctiva, sclera, cornea, and iris. Evidence of blepharitis (Fig. 5.5) or any discharge or tearing should be evaluated and if applicable, treatment should be advised prior to the proposed surgery date. For pupil—size, shape, symmetry, and reaction to light should be noted. Microphthalmia and poorly dilating pupils are indicators of arrested development and increase the risk of a poor anatomical and functional outcome after cataract surgery. It has been our impression that poorly dilating pupils indicate an overall immaturity of the anterior segment and may be a marker for increased risk of glaucoma after cataract surgery.



Figure 5.5. Blepharitis in a 6-year-old child scheduled for intraocular surgery.

After dilation, a slit-lamp evaluation should be carried out if the child is old enough to be cooperative. The slit-lamp examination findings can help with the search for a cause of the cataract, help establish a prognosis, and help plan the surgical strategy. The morphology of the cataract may affect prognosis and give a clue to the etiology. Unilateral PSC should prompt a careful search for evidence of trauma. Bilateral PSC cataract may result from chronic uveitis, prolonged corticosteroid treatment for chronic disease, radiation treatment for malignancy, or nonaccidental injury (child abuse). Children with juvenile idiopathic arthritis (JIA) may have associated band-shaped keratopathy and posterior synechia. Lens subluxation, iridodonesis, and aniridia should be looked for. Total cataract involving the whole lens can occur in Down syndrome, type 1 diabetes mellitus, congenital rubella, and posterior lentiglobus. In cases of unilateral cataract, examination of the fellow eye after pupil dilation is essential to rule out asymmetric bilateral findings. Anterior lenticonus is most often associated with Alport syndrome and should be investigated accordingly. A sudden onset of total cataract may be an indication of unsuspected trauma, diabetic cataract, or preexisting ruptured anterior (reported in anterior lenticonus) or posterior capsule (reported in posterior lentiglobus). If the anterior vitreous can be visualized, the “fish-tail” sign suggests that the posterior capsule is incompetent or grossly ruptured. Fish-tail refers to the to-and-fro movement of the lens material in the vitreous as the eye gazes slightly right and left.

For children above about 5 to 6 years of age, the ability of the child to cooperate for slit-lamp examination is also an indirect indicator that the child will cooperate for Nd-YAG laser capsulotomy if needed. In children above 5 to 6 years of age with an intact posterior capsule and an AcrySof® IOL implantation, visually significant posterior capsule opacification (PCO) is known to develop most commonly at 18 to 24 months after surgery. If a child in this age range seems to be cooperative for slit-lamp examination during the preoperative evaluation, the surgeon may decide to leave behind an intact

posterior capsule (assuming high odds of getting the child’s cooperation for YAG-laser if needed).

A slit-lamp examination of both parents, if possible, helps to establish the presence of familial cataracts and cataract-associated conditions. These findings can be subtle and the parents may not have been told that they have any pathology at all. Variability of the severity of cataracts within the same family is common.

OCT can be performed preoperatively if the lens is clear enough and the child is able to cooperate (see Chapter 21). Baseline specular microscopy is advised by some; however, it is more commonly used for secondary IOL implantation or anterior chamber IOL implantation.

Axial Length Measurement and Keratometry

For older children, axial length (AL) measurement can be obtained in office using ultrasound or optical biometry. This is especially important if A-scan instrument is not available in the operating room. Similarly, keratometry can be performed in the clinic.

Ultrasound Biometry

The ultrasound probe is placed into the solution and positioned parallel to the axis of the eye. Axiality is judged by watching for the correct spike patterns on the oscilloscope screen as the probe position is adjusted. The examiner should be familiar with the characteristics of a good A-scan tracing with a spike from each layer of the eye. When the probe is aligned with the optical axis of the eye and the ultrasound beam is perpendicular to the retina, the retinal spike is displayed as a straight, steeply rising echo spike. When the probe is not properly aligned with the optical axis of the eye, the ultrasound beam is not perpendicular to the retinal surface and the retinal spike is displayed as a jagged, slow-rising echo spike. Repeated measurements are taken until a few equal measurements are obtained with sharp retinal spikes.

Ultrasound can be done with either contact or immersion methods. In the contact method, the probe touches the cornea and may result in corneal compression and a shorter AL. Corneal compression is more likely in pediatric eyes because of low corneal and sclera rigidity. Using the immersion technique, the ultrasound probe does not come into direct contact with the cornea, but instead uses a coupling fluid between the cornea and probe preventing corneal indentation. Immersion A-scan has been shown to be superior to contact biometry in children.^{11,12} If contact A-scan is used, it is important to make sure that the tip does not indent the cornea. Pediatric cataract surgeons use the contact technique more frequently when measuring the AL of pediatric eyes at the time of cataract surgery. This statement is based on 2009 e-mail survey sent to pediatric ophthalmologists, in which 173 (82.4%) surgeons reported using contact A-scan compared with

37 (17.6%) who reported using the immersion technique.¹² Because of a lack of cooperation in the clinic setting, AL measurements in young children often must be obtained in the operating room under general anesthesia. In the operating room setting, an experienced ultrasonographer may not be available. Contact A-scan measurements are easier for the surgeon or an operating room technician to perform. Immersion A-scan requires more experience and practice and is best performed by an experienced ultrasonographer. In a prospective clinical trial, we compared AL measurements by contact and immersion techniques in pediatric cataractous eyes ($n = 50$ eyes of 50 children).¹² AL was measured by both contact and immersion techniques for all eyes, randomized as to which to perform first to avoid measurement bias. AL measurement by contact technique was significantly shorter as compared with immersion technique (21.36 ± 3.04 mm and 21.63 ± 3.09 mm, respectively; $P < 0.001$). AL measurements using the contact technique were on an average 0.27 mm shorter than those obtained using the immersion technique. Forty-two eyes (84%) had shorter AL when measured using the contact technique as compared with the immersion technique. Lens thickness (LT) measurements by contact technique was not significantly different from that of immersion technique (3.61 ± 0.74 and 3.60 ± 0.67 mm, respectively; $P = 0.673$). Anterior chamber depth (ACD) measurements were significantly more shallow with the contact technique (3.39 ± 0.59 mm and 3.69 ± 0.54 mm, respectively; $P < 0.001$). As can be seen here, shorter AL in contact group was mainly as a result of ACD value rather than LT value. IOL power needed for emmetropia was significantly different (28.68 diopters [D] versus 27.63 D; $P < 0.001$). During IOL power calculation, if AL measured by contact technique is used, it will result in the use of an average 1-D stronger IOL power than is actually required. This can lead to induced myopia in the postoperative refraction. A consistent error could be compensated for by the addition of a constant or by formula personalization; however, this is not possible because the compression error varies from eye to eye.

In a subsequent study, we compared prediction error (PE) and absolute prediction error (APE) using contact and immersion techniques.¹¹ The contact and immersion A-scan biometry techniques had been performed in each eye and PE using each technique was compared. There was a significant difference in PE between contact and immersion A-scan biometry in children. The mean PE was $+0.4 \pm 0.7$ D in the contact group and -0.4 ± 0.8 D in the immersion group ($P < 0.001$) and the mean APE was 0.7 ± 0.4 D and 0.7 ± 0.6 D, respectively ($P = 0.694$). The APE was <0.5 D in 5 eyes (23%) using the contact technique and in 11 eyes (50%) using the immersion technique. The mean postoperative spherical equivalent was $+2.9 \pm 2.5$ D, which was significantly different from

the mean predicted refraction for contact A-scan (3.3 ± 2.8 D; $P = 0.010$) but not immersion A-scan (2.5 ± 2.5 D; $P = 0.065$). Ben-Zion et al.¹³ compared PEs of 138 pediatric eyes measured by the contact A-scan technique with a later group of 65 children measured with the immersion technique. They found no significant difference in APE (1.11 and 1.03 D, respectively) and noted PE of $+0.23$ and -0.32 D with the contact technique and immersion technique, respectively.

Optical Biometry

Optical biometry is based on partial coherence interferometry (PCI)—IOLMaster (Carl Zeiss Meditec) or LenStar (Haag Streit). LenStar allows higher resolution compared with the IOLMaster. The measurement includes corneal thickness, ACD, LT, AL, keratometry, white-to-white distance, pupillometry, eccentricity of the visual axis, and retinal thickness at the point of fixation. It can also be used to access the horizontal iris width, pupil diameter, eccentricity of the visual axis, and retinal thickness. PCI has been used in cooperative children with reliability and accuracy. PCI requires patient cooperation and thus may not be a viable option in infants and young children. Claimed improvements over conventional ultrasound techniques include high reproducibility, contact-free measurements, and observer independence of the measurements. Lenhart et al.¹⁴ reported PE for AL measurements obtained using PCI versus immersion ultrasonography in children. AL measurements in the operative eye were obtained using PCI at the preoperative clinic visit and then using immersion US in the operating room before surgery. The data were compared to determine the degree of agreement. The charts of 18 patients (27 eyes) were reviewed. Preoperative AL measurements by PCI were obtained in 21 eyes (78%). On average, the PCI-measured ALs were 0.1 mm less than the immersion US values (95% confidence interval, -0.2 to -0.1 ; $P = 0.002$). All eyes with an AL of 23.5 mm or less had lower PCI values than immersion US values. There was no systematic pattern of 1 measurement being greater or lesser than the other in eyes with an AL longer than 23.5 mm. The authors concluded that there was a systematic difference in AL measurement between PCI and immersion technique, with PCI tending to give lower values, particularly in eyes with an AL of 23.5 mm or less. Gursoy et al.¹⁵ compared AL, ACD, and LT measured with LenStar with those obtained with A-scan contact technique. Right eyes of 565 school children were included (mean age 10.5 years). The mean difference between contact ultrasound and LenStar was -0.72 , -0.27 , and $+0.24$ mm for AL, ACD, and LT, respectively. PCI technology is not able to obtain measurements in eyes with dense cataract and in those that cannot fixate on the red light of the instrument because of inadequate vision. In children, the cataracts are often dense and fixation may be inadequate.

Biometry Values in Eyes with Pediatric Cataract

We reported biometry data of pediatric cataractous eyes (randomly selected single eye in bilateral cases; cataractous eye in unilateral cases).¹⁶ Three hundred ten eyes were analyzed, with a mean age at cataract surgery of 45.30 ± 48.1 months (median, 27.50; range, 0.23–203.08); mean AL of 20.52 ± 2.87 mm (range, 14.19–29.10); ACD of 3.29 ± 0.60 mm (range, 1.48–4.35); and LT of 3.62 ± 0.86 mm (range, 0.61–6.35). Table 5.1 shows the mean AL per age group. In Table 5.2, the first 2 years of life are divided into age groups, showing the mean AL per age group.

The overall mean AL of pediatric cataractous eyes (20.5 ± 2.9 mm) in our series was significantly different ($P < 0.001$) than the overall mean AL of pediatric non-cataractous eyes described in a Gordon and Donzis series (21.9 ± 1.6 mm).¹⁷ More important, the standard deviation was nearly two times more in eyes with cataract than in those without (± 2.9 mm versus ± 1.6 mm). This difference is a very important factor to keep in mind; that is, these cataractous eyes are abnormal to begin with, which may also lead to variations in postoperative growth. Eyes with cataract showed a shorter AL in the first 12 months of life (cataractous, 17.9 ± 2.0 mm; noncataractous, 19.2 ± 0.7 mm). In the first 12 months of life, the standard

Table 5.1 AL MEASUREMENTS IN UNILATERAL AND RANDOMLY SELECTED SINGLE EYES OF PEDIATRIC PATIENTS WITH BILATERAL CATARACTS, CATEGORIZED BY AGE

Age ^a (y)	n	Length (Mean \pm SD)	95% CI	Range
<1	119	17.67 \pm 1.88	17.33–18.02	14.19–22.62
1–2	30	21.54 \pm 1.34	21.04–22.04	19.09–25.86
2–3	19	21.84 \pm 1.68	21.03–22.65	18.10–24.56
3–4	22	22.37 \pm 1.92	21.52–23.22	18.91–26.26
4–5	20	21.83 \pm 1.07	21.33–22.34	19.68–23.63
5–6	18	22.02 \pm 1.89	21.08–22.96	18.37–27.59
6–7	15	22.37 \pm 1.74	21.41–23.34	19.18–26.37
7–8	16	22.58 \pm 1.62	21.72–23.45	20.68–26.73
8–9	13	23.43 \pm 2.59	21.86–25.00	20.61–29.10
9–10	12	22.25 \pm 1.15	21.52–22.98	20.72–24.44
10–18	26	23.20 \pm 1.54	22.58–23.83	20.85–27.99
Total	310	20.52 \pm 2.87	20.20–20.84	14.19–29.10

Data are expressed as mean millimeters \pm SD.

^aPost hoc analysis (Tukey test) revealed that AL at <1 year was significantly different from that in all other age groups ($P < 0.001$). AL of 1- to 2-year-old children was significantly different from that in children older than 10 years ($P = 0.02$). No other pair reached significance.

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Table 5.2 AL MEASUREMENTS DURING FIRST 2 YEARS OF LIFE — AL MEASUREMENTS IN UNILATERAL AND RANDOMLY SELECTED SINGLE EYES OF PEDIATRIC PATIENTS WITH BILATERAL CATARACTS

Age (mo)	n	Length (Mean \pm SD)	95% CI	Range
<1	24	16.01 \pm 1.17	15.51–16.51	14.19–18.13
1–2	36	16.78 \pm 1.15	16.39–17.17	14.33–20.44
2–3	13	17.30 \pm 1.42	16.44–18.16	15.64–20.57
3–6	21	18.63 \pm 1.35	18.01–19.24	16.13–21.11
6–12	25	19.95 \pm 1.17	19.47–20.44	17.09–22.62
12–18	15	21.73 \pm 0.98	21.18–22.27	20.04–23.65
18–24	15	21.35 \pm 1.64	20.44–22.26	19.09–25.86
Total	149	18.45 \pm 2.37	18.07–18.84	14.19–25.86

Data are expressed in millimeters.

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deviation was almost three times that of eyes without cataract (± 2.0 mm versus ± 0.7 mm).

Logarithmic transformation of age explained more variation in AL than did age (70% when log of age is used and 41% with age). Linear regression analysis revealed that during the first 6 months of life, AL increased by 0.62 mm per month. From 6 to 18 months of age, it increased by 0.19 mm per month and after 18 months, by 0.01 mm per month or 0.12 mm per year. The mean AL during the first month of age was 16.01 mm, which increased to 23.20 mm in children 10 to 18 years of age, showing 7.19 mm in axial growth. Our finding suggested a *rapid, postnatal* phase from birth to 6 months of age, followed by a *slower, infantile* phase from 6 to 18 months of age, and finally a *slow, juvenile* phase from 18 months forward. At the younger ages, eyes with cataract had a shorter AL compared with their normal fellow eyes or data of the pediatric population without cataract. With advancing age, eyes with cataract had a longer AL than did their normal fellow eyes.

Table 5.3 shows descriptive statistics of mean AL as they relate to gender, race, and laterality of cataract per the indicated age groups. Girls had a shorter AL than did the boys (20.23 mm versus 20.78 mm, $P = 0.09$).¹⁶ Significantly longer eyes were found in African American patients than in Caucasian patients (21.66 mm versus 20.14 mm, $P < 0.001$). We found that eyes with unilateral cataracts overall had a shorter mean AL than those with bilateral cataracts (20.15 mm versus 21.10 mm, $P = 0.003$).¹⁶ This effect was seen until 60 months of age. Eyes with cataract may also be associated with ocular anomalies (e.g., microphthalmos) leading to shorter AL. However, in children beyond 60 months of age, unilateral

Table 5.3 AL MEASUREMENTS IN UNILATERAL AND RANDOMLY SELECTED SINGLE EYES OF PEDIATRIC PATIENTS WITH BILATERAL CATARACTS, IN REFERENCE TO GENDER, RACE, AND LATERALITY OF CATARACT

	Birth–6	6–18	18–60	60–200	All
Gender					
F	17.12 ± 1.48 (n = 49)	20.32 ± 1.17 (n = 20)	21.55 ± 1.76 (n = 32)	22.65 ± 1.53 (n = 45)	20.23 ± 2.78 (n = 146)
M	17.01 ± 1.62 (n = 45)	20.92 ± 1.57 (n = 20)	22.15 ± 1.48 (n = 44)	22.71 ± 2.02 (n = 55)	20.78 ± 2.93 (n = 164)
P	0.75	0.18	0.11	0.87	0.09
Race					
Caucasian	17.01 ± 1.55 (n = 75)	20.51 ± 1.33 (n = 34)	21.65 ± 1.49 (n = 53)	22.16 ± 1.26 (n = 70)	20.14 ± 2.63 (n = 232)
African American	17.28 ± 1.54 (n = 19)	21.25 ± 1.74 (n = 6)	22.46 ± 1.80 (n = 23)	23.90 ± 2.28 (n = 30)	21.66 ± 3.24 (n = 78)
P	0.50	0.24	0.043^a	<0.001^a	<0.001^a
Laterality					
Unilateral	17.02 ± 1.56 (n = 73)	20.38 ± 1.45 (n = 23)	21.85 ± 1.65 (n = 41)	23.06 ± 2.04 (n = 53)	20.15 ± 3.11 (n = 190)
Bilateral	17.24 ± 1.50 (n = 21)	20.94 ± 1.30 (n = 17)	21.95 ± 1.61 (n = 35)	22.25 ± 1.41 (n = 47)	21.10 ± 2.34 (n = 120)
P	0.57	0.21	0.80	0.02^a	0.003^a
Total	17.07 ± 1.54 (n = 94)	20.62 ± 1.40 (n = 40)	21.90 ± 1.62 (n = 76)	22.68 ± 1.81 (n = 100)	20.52 ± 2.87 (n = 310)

Data are expressed as mean millimeters ± SD. Age ranges are in months.

^aSignificant at $P < 0.05$.

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cataractous eyes were longer than bilateral cataractous eyes (23.06 mm versus 22.25 mm) perhaps due to elongation caused by deprivation amblyopia. Table 5.4 illustrates AL in unilateral cataractous pediatric eyes compared with those in fellow eye with clear lens. In young eyes with unilateral cataract, the cataractous eyes had a shorter mean AL than did their fellow eyes. As age advances, eyes with unilateral cataract have a mean AL longer than their fellow eyes with clear lens.

Nearly half of the subjects reported to have ≥ 0.5 mm interocular AL difference (difference of AL between

Table 5.4 ALs IN UNILATERAL CATARACTOUS PEDIATRIC EYES COMPARED WITH THOSE IN FELLOW EYES WITH CLEAR LENSES

Age (mo)	n	Cataract Eye	Fellow Eye	P
0–6	72	16.98 ± 1.54	17.50 ± 1.48	0.001^a
6–18	23	20.38 ± 1.45	20.52 ± 0.93	0.534
18–60	38	21.75 ± 1.65	21.58 ± 1.07	0.513
60–200	51	23.11 ± 2.07	23.04 ± 1.39	0.735
Total	184^b	20.09 ± 3.13	20.26 ± 2.69	0.095

Data are expressed as mean millimeters ± SD.

^aSignificant at $P < 0.05$.

^bFellow eye AL data were not available for six eyes.

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operated eye and the fellow eye).¹⁸ Forty-one patients (24.0%) had operated (cataractous) eyes that were ≥ 0.5 mm shorter than the fellow eye, and 41 patients (24.0%) had operated eyes that were ≥ 0.5 mm longer than the fellow eye. If absolute values were used, mean interocular AL difference (IALD) was 0.76 ± 0.86 mm (for unilateral and bilateral cases) as it was 0.98 ± 0.90 and 0.40 ± 0.65 in unilateral and bilateral cases, respectively [$P < 0.001$].

Tables 5.5 and 5.6 show descriptive statistics of mean ACD and LT, respectively, as they relate to gender, race, and cataract laterality, per the indicated age groups. Tables 5.7 and 5.8 report ACD and LT in eyes of pediatric unilateral cataract compared with their corresponding fellow eye with clear lens.

Keratometry in Eyes with Pediatric Cataract

We reported preoperative keratometry in children with cataract.¹⁹ Of the 299 eyes analyzed in our study, the average (SD) keratometry value was 45.39 ± 3.08 D (range 39.25–63.5) (Table 5.9). Age and AL demonstrated a significant linear relationship with K values ($P < 0.001$, R^2 : log of age, 0.31; AL 0.32). Keratometry values of younger children (aged 0–6 months) were significantly different from those of older children ($P < 0.001$). Girls had steeper corneas when compared with boys ($P = 0.03$). The values of eyes with cataract in monocular cases were steeper than that of bilateral cases ($P = 0.07$). For unilateral cataract, the eye with the cataract had a significantly steeper cornea than the fellow eye (Table 5.10) ($P = 0.02$).

Table 5.5 ACD MEASUREMENTS IN UNILATERAL AND RANDOMLY SELECTED SINGLE EYES OF PEDIATRIC PATIENTS WITH BILATERAL CATARACTS, IN REFERENCE TO GENDER, RACE, AND LATERALITY OF CATARACT

	Birth–6	6–18	18–60	60–200	All
Gender					
F	2.67 ± 0.39 (n = 33)	3.29 ± 0.45 (n = 18)	3.44 ± 0.37 (n = 21)	3.56 ± 0.48 (n = 30)	3.20 ± 0.56 (n = 102)
M	2.68 ± 0.57 (n = 23)	3.42 ± 0.50 (n = 15)	3.56 ± 0.51 (n = 30)	3.63 ± 0.45 (n = 40)	3.38 ± 0.62 (n = 108)
P	0.94	0.43	0.34	0.51	0.026^a
Race					
Caucasian	2.62 ± 0.45 (n = 43)	3.42 ± 0.44 (n = 27)	3.50 ± 0.43 (n = 38)	3.65 ± 0.42 (n = 52)	3.30 ± 0.60 (n = 160)
African American	2.84 ± 0.50 (n = 13)	3.00 ± 0.48 (n = 6)	3.56 ± 0.53 (n = 13)	3.48 ± 0.55 (n = 18)	3.28 ± 0.59 (n = 50)
P	0.14	0.046^a	0.68	0.25	0.82
Laterality					
Unilateral	2.60 ± 0.46 (n = 42)	3.41 ± 0.42 (n = 18)	3.53 ± 0.40 (n = 28)	3.49 ± 0.52 (n = 35)	3.18 ± 0.6 (n = 123)
Bilateral	2.88 ± 0.43 (n = 14)	3.26 ± 0.52 (n = 15)	3.49 ± 0.52 (n = 23)	3.72 ± 0.36 (n = 35)	3.44 ± 0.53 (n = 87)
P	0.054	0.37	0.77	0.033^a	0.001^a
Total	2.67 ± 0.47 (n = 56)	3.35 ± 0.47 (n = 33)	3.51 ± 0.45 (n = 51)	3.60 ± 0.46 (n = 70)	3.29 ± 0.60 (n = 210^b)

Data are expressed as mean millimeters ± SD. Age ranges are expressed in months.

^aSignificant at $P < 0.05$.

^bACD data were not available for 100/310 (32.3%) eyes.

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Table 5.6 LT MEASUREMENTS IN UNILATERAL AND RANDOMLY SELECTED SINGLE EYES OF PEDIATRIC PATIENTS WITH BILATERAL CATARACTS, IN REFERENCE TO GENDER, RACE, AND LATERALITY OF CATARACT

	Birth–6	6–18	18–60	60–200	All
Gender					
F	3.10 ± 0.76 (n = 32)	3.91 ± 0.96 (n = 17)	3.71 ± 0.56 (n = 20)	3.86 ± 0.97 (n = 30)	3.59 ± 0.89 (n = 99)
M	3.19 ± 0.97 (n = 22)	3.84 ± 0.92 (n = 15)	3.82 ± 0.77 (n = 29)	3.70 ± 0.66 (n = 40)	3.64 ± 0.83 (n = 106)
P	0.689	0.84	0.59	0.42	0.67
Race					
Caucasian	3.22 ± 0.70 (n = 41)	3.86 ± 0.79 (n = 26)	3.83 ± 0.75 (n = 37)	3.70 ± 0.73 (n = 52)	3.63 ± 0.77 (n = 156)
African American	2.87 ± 1.20 (n = 13)	3.95 ± 1.49 (n = 6)	3.60 ± 0.43 (n = 12)	3.97 ± 0.98 (n = 18)	3.59 ± 1.09 (n = 49)
P	0.34	0.83	0.32	0.29	0.79
Laterality					
Unilateral	3.15 ± 0.79 (n = 40)	3.73 ± 0.88 (n = 18)	3.76 ± 0.32 (n = 28)	4.03 ± 0.92 (n = 35)	3.63 ± 0.84 (n = 121)
Bilateral	3.09 ± 1.02 (n = 14)	4.08 ± 0.99 (n = 14)	3.79 ± 1.00 (n = 21)	3.50 ± 0.81 (n = 70)	3.60 ± 0.89 (n = 84)
P	0.81	0.30	0.91	0.005^a	0.79
Total	3.14 ± 0.84 (n = 54)	3.88 ± 0.93 (n = 32)	3.77 ± 0.69 (n = 49)	3.77 ± 0.81 (n = 70)	3.62 ± 0.86 (n = 205^b)

Data are expressed as mean millimeters ± SD. Age ranges are expressed in months.

^aSignificant at $P < 0.05$.

^bLT data were not available for 105/310 (33.9%) eyes.

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Table 5.7 ACDs IN UNILATERAL CATARACTOUS PEDIATRIC EYES COMPARED WITH THOSE IN FELLOW EYES WITH CLEAR LENSES

Age (mo)	n	Cataract Eye	Fellow Eye	P
0–6	41	2.60 ± 0.47	2.74 ± 0.46	0.047^a
6–18	18	3.41 ± 0.42	3.51 ± 0.30	0.353
18–60	27	3.58 ± 0.32	3.53 ± 0.27	0.381
60–200	32	3.50 ± 0.52	3.65 ± 0.33	0.072
Total	118^b	3.19 ± 0.62	3.29 ± 0.54	0.013^a

Data are expressed as mean millimeters ± SD.
^aSignificant at $P < 0.05$.
^bFellow eye ACD data were not available for 65 eyes.
 (Reprinted from Trivedi RH, Wilson ME. Biometry data from Caucasian and African-American cataractous pediatric eyes. *Invest Ophthalmol Vis Sci* 2007;48:4671–4678 (p. 4677), with permission.

Table 5.8 LTs IN UNILATERAL CATARACTOUS PEDIATRIC EYES COMPARED WITH THOSE IN FELLOW EYES WITH CLEAR LENSES

Age (mo)	n	Cataract Eye	Fellow Eye	P
0–6	39	3.18 ± 0.79	3.65 ± 0.49	0.001^a
6–18	18	3.73 ± 0.88	3.73 ± 0.23	0.968
18–60	27	3.74 ± 0.31	3.81 ± 0.26	0.369
60–200	32	4.07 ± 0.95	3.74 ± 0.34	0.067
Total	116^b	3.64 ± 0.84	3.73 ± 0.37	0.294

Data are expressed as mean millimeters ± SD.
^aSignificant at $P < 0.05$.
^bFellow eye lens thickness data were not available for five eyes.
 (Reprinted from Trivedi RH, Wilson ME. Biometry data from Caucasian and African-American cataractous pediatric eyes. *Invest Ophthalmol Vis Sci* 2007;48:4671–4678 (p. 4677), with permission.

PARENTAL COUNSELING AND INFORMED CONSENT

As soon as surgery is proposed for a cataract in a child, the informed consent process begins. Informed consent is an essential part of a surgical practice. As with many other aspects of medicine, this process is both a *science* and an *art*. It is a fundamental duty and responsibility that is recognized by virtually every professional medical organization.²⁰ The

ophthalmologist must be able to explain to the patient’s parents or legal guardian (and to the child as well, if appropriate) the nature of the problem and why surgery is being recommended to alleviate it. It is the physician’s ethical responsibility to be honest with the patient. It is the parents’ right to make decisions regarding the destiny of their children. These guardians are not in a position to do this without appropriate knowledge and the physician is obligated to educate them and obtain an informed consent.

Table 5.9 KERATOMETRY MEASUREMENTS IN UNILATERAL AND RANDOMLY SELECTED SINGLE EYES OF PEDIATRIC PATIENTS WITH BILATERAL CATARACTS, IN REFERENCE TO GENDER, RACE, AND LATERALITY OF CATARACT

	Birth–6	6–18	18–60	60–200	All
Gender					
Female	48.27 ± 4.19 (n = 48)	44.63 ± 2.24 (n = 19)	44.17 ± 2.11 (n = 30)	44.72 ± 1.94 (n = 43)	45.81 ± 3.45 (n = 140)
Male	47.68 ± 2.82 (n = 42)	44.77 ± 1.88 (n = 20)	43.83 ± 1.81 (n = 43)	43.98 ± 1.82 (n = 54)	45.02 ± 2.67 (n = 159)
P	0.44	0.84	0.470	0.056	0.03 ^a
Race					
White	48.01 ± 3.23 (n = 71)	44.70 ± 2.03 (n = 33)	43.90 ± 1.86 (n = 51)	44.37 ± 1.80 (n = 68)	45.47 ± 2.95 (n = 223)
African American	47.93 ± 4.90 (n = 19)	44.73 ± 2.26 (n = 6)	44.13 ± 2.12 (n = 22)	44.16 ± 2.14 (n = 29)	45.14 ± 3.42 (n = 76)
P	0.93	0.98	0.64	0.62	0.42
Laterality					
Unilateral cataract	48.37 ± 3.72 (n = 70)	44.29 ± 2.09 (n = 22)	43.78 ± 1.76 (n = 40)	43.87 ± 1.97 (n = 51)	45.62 ± 3.49 (n = 183)
Bilateral cataract	46.69 ± 2.90 (n = 20)	45.25 ± 1.89 (n = 17)	44.20 ± 2.13 (n = 33)	44.79 ± 1.70 (n = 46)	45.01 ± 2.23 (n = 116)
P	0.07	0.15	0.37	0.02 ^a	0.07
Total	47.99 ± 3.61 (n = 90)	44.70 ± 2.04 (n = 39)	43.97 ± 1.93 (n = 73)	44.30 ± 1.90 (n = 97)	45.39 ± 3.08 (n = 299)

Data are expressed as mean diopters ± SD. Age ranges are in months.
^aSignificant at $P < 0.05$.
 (Reprinted from Trivedi RH, Wilson ME. Keratometry in pediatric eyes with cataract. *Arch Ophthalmol* 2008;126:38–42, with permission.)

Table 5.10 KERATOMETRY IN UNILATERAL CATARACTOUS PEDIATRIC EYES COMPARED WITH THOSE IN FELLOW EYES WITH CLEAR LENSES

Age (mo)	n	Cataract Eye	Fellow Eye	P
0–6	57	48.73 ± 3.81	47.56 ± 2.96	0.01 ^a
6–18	18	44.08 ± 2.14	43.99 ± 1.97	0.79
18–60	32	43.50 ± 1.63	43.39 ± 1.20	0.53
60–200	43	43.57 ± 1.91	43.64 ± 1.98	0.25
Total	150^b	45.58 ± 3.70	45.11 ± 2.97	0.02^a

Data are expressed as mean diopters ± SD.

^aSignificant at $P < 0.05$.

^bFellow eye keratometry data were missing in 33 eyes. Thus, out of 183 eyes with unilateral cataract, only 150 eyes were available for comparative analyses.

(Reprinted from Trivedi RH, Wilson ME. Keratometry in pediatric eyes with cataract. *Arch Ophthalmol* 2008;126:38–42, with permission.)

Appropriate available options for managing the cataract and the optical rehabilitation should be explained in terms that the family can understand. In this electronic age, with nearly everyone having access to Internet information, families often arrive in the office with much more knowledge about their child's condition than would have been true only a few years ago. While not all of the information available on the Internet is scientifically based, overall the availability of information has led to more knowledgeable and inquisitive parents and patients. Surgeons who perform pediatric cataract surgery should be prepared for lengthy discussions with this new breed of parents. The extra time is not wasted, however, because a better-informed family is much more likely to comply with the frequent follow-ups, medications, patching, glasses wear, etc., that are so essential to an eventual quality VA outcome from the surgery.

Spaeth has written that “informed consent is, in many ways, at the heart of the American system of medical and surgical practice.”²¹ When informed consent is done properly, the parents and the physician become partners with the common goal of doing what is best for the patient's health and well being. Time spent establishing this partnership prior to surgery will help assure compliance with the difficult aftercare and follow-up. Parents have a right to understand the treatment that is being recommended for their child. The surgeon must use the language that best accomplishes that goal. While the signing of the surgical consent form can sometimes be delegated to an assistant, the surgeon should be the one who informs parents about the treatment details, alternatives, and meaningful risks of the proposed surgery. A request for a second opinion may come up during discussion, and

the surgeon should be open to facilitating the request. Surgeons should also be prepared for the common question, “What would you do if this were your child?”

Some surgeons view the informed consent process as primarily a way to help prevent malpractice lawsuits. However, the signed consent document does little more than prevent accusations of battery. The family may state at a later time that they did not understand the nature or the risks of the surgery and that they signed the form because they were told to sign it. An informed consent form in itself will not effectively prevent the filing of malpractice lawsuits. On the other hand, a meaningful exchange of information that leads to the partnership mentioned earlier will significantly decrease the risk of a lawsuit even if the surgery has a poor outcome. A family that agrees wholeheartedly with the decision to pursue a particular surgical option is much less likely to file a lawsuit when a complication occurs, even if that particular complication was not specifically discussed preoperatively. Lack of adequate information about a particular complication is not often the basis for a malpractice claim. In one study, 2.5% of malpractice claims were based on the failure to obtain adequate informed consent.²²

Negligence is a more common claim by a prosecuting attorney. When families feel that the surgical procedure adopted was not indicated or understood, they are less accepting when a complication occurs. Plaintiffs' lawyers plead lack of informed consent only as a last resort, and not usually as the primary charge against the surgeon.

In addition to the commonsense considerations discussed above, specific professional responsibilities also govern the discussion of risks, benefits, and alternatives when medical or surgical treatments are proposed. The following advisory opinion, based on the American Academy of Ophthalmology (AAO)'s Code of Ethics, is reprinted from the AAO Web site (<http://www.aao.org/about/ethics/upload/Informed-Consent-2008.pdf>):

When medical and surgical procedures are proposed, both ethical principles and the law require discussion of significant associated risk. Clearly, an ophthalmologist must understand and conform to the minimum required by applicable law. In some states, a community standard is used by which a physician must disclose any information about risks and other factors that the average prudent physician in the community would disclose. Most other states set a higher standard, requiring disclosure of all information possessed by the doctor that a reasonable patient would find significant in deciding whether or not to undergo a procedure. Although legal requirements are an important benchmark on which to build, they should be regarded as a minimum standard that is routinely exceeded by practice of good professional ethics.

From the ethical perspective, any risks or potential complications that are sufficiently common or significant that they might reasonably influence the patient's judgment to

accept the proposed treatment must be disclosed. Exclusions may include very minor, rare, or inconsequential risks. Similarly, if a risk is readily apparent to people of common sense, then discussion can reasonably be excluded unless the physician has reason to believe that such a disclosure is necessary or appropriate in view of the unique needs of a particular patient. Essentially, the physician must explain the rationale for the treatment, significant benefits, risks, and reasonable alternatives to the treatment proposed in language that the patient can understand.

When a patient is too young to legally consent to treatment, or when a patient lacks the capacity to comprehend and decide independently, the informed consent must be obtained from a surrogate who is legally entitled to provide consent on the patient's behalf. The same procedure for explaining the rationale, risks, benefits, and alternatives should be followed with a guardian or surrogate.

In the specific instance of pediatric cataract surgery, the informed consent discussion should include the criteria used by the surgeon to decide that the cataract is visually significant, the details of the surgical procedure being proposed (i.e., whether a posterior capsulectomy and anterior vitrectomy will be used and whether an IOL will be placed), whether the surgery will result in the postoperative need for glasses, contact lens, and/or patching for amblyopia, and the importance of postoperative follow-up and postoperative medications as well as the importance of avoiding postoperative trauma to the recently operated eye. A general discussion of operative and postoperative risks should include a discussion of VAO, IOL malposition, abnormalities of the size and shape of the pupil, and variability in the postoperative residual refractive error. Endophthalmitis and retinal detachment are rare after pediatric cataract surgery but are of such significance to vision that they should be mentioned as part of the informed consent. Complicating conditions that may occur unrelated to the surgical event itself are nonetheless very important as part of the informed consent process. The partnership being developed with the parents is meant to help assure appropriate follow-up and appropriate attention to complicating conditions that may not appear for years after the surgery. Aphakic/pseudophakic glaucoma, deprivation amblyopia, strabismus, and changing refractive error are conditions in this category and are ideally discussed as part of the preoperative knowledge exchange.

The informed consent discussion should include relevant issues related to the placement of an IOL. While IOL implantation has become the most common method used to correct aphakia in children overall, it is still considered "off label," or as a "physician-directed" indication. This designation means that the IOLs implanted in children were tested as part of their FDA market approval process only in adults. It does not mean that the FDA has disallowed their use in children. It implies only that the device is being used for a purpose or in a patient population that

is different from the one for which it was tested as part of the market approval process. The following paragraph is taken directly from the FDA Web site and serves to give surgeons some guidance on IOL use in children and whether institutional review board oversight is indicated when these devices are implanted in children.²³

Good medical practice in the best interests of the patient requires that physicians use legally available drugs, biologics and devices according to their best knowledge and judgment. If physicians use a product for an indication not in the approved labeling, they have the responsibility to be well informed about the product, to base its use on a firm scientific rationale and on sound medical evidence, and to maintain records of the products use and effects. Use of a marketed product in this manner when the intent is the "practice of medicine" does not require the submission of an investigational new drug application (IND), investigational device exemption (IDE) or review by an institutional review board (IRB). However, the institution at which the product will be used may, under its own authority, require IRB review or other institutional oversight.

Parents should be made aware that IOL implantation in children is considered an "off label" or "physician-directed" use. However, they should also be made aware that IOL implantation at the time of cataract surgery has become the treatment of choice for a large majority of surgeons when the recipient is beyond infancy. As Levin²⁴ has stated, patients and their families come to us "with a trust that we cannot betray." The foundation of that trust is that we will act in their best interests and that we will tell them the truth at all times. We must openly and honestly discuss with our patients the innovative nature of proposed treatments, such as IOL implantation and its variations, including the degree of newness, the past experience of others and ourselves, and even the disclosure that "I have never done this before." Conversations are also needed about potential conflicts of interest, academic, financial, or otherwise.

There is no doubt that the informed consent process surrounding pediatric cataract surgery is more time-consuming and in many ways more complex than for most other ophthalmic procedures. Truth telling and the artful transfer of knowledge in terms appropriate for the family will help assure that a physician-patient relationship built on trust will develop.

A coordinated plan of action can best be developed when the parents understand the reasons for, goals of, and the advantages and potential complications of cataract surgery. The parents/caregivers play a critical role in the postoperative care of the eye and treatment of amblyopia. They must understand that a successful visual outcome also depends on their ability to maintain adequate aphakic correction and follow through with amblyopia

therapy. Before moving forward with IOL implantation, it is important to discuss the major pros and cons of the available options with the parents/legal guardian.

Erraguntla et al.²⁰ prospectively investigated the effectiveness of information transfer by the pediatric cataract surgeon (Dr. Alex Levin) to the parents or guardian of children during informed-consent process.²⁰ Of 31 parents, 58% overestimated their understanding of the informed-consent discussion. Parents scored well on the questions about the nature of the disease and the postoperative follow-up, but scored lower on questions regarding surgical risks and outcomes. Parents identified several barriers to understanding, including the large amount of information, stress, and preoccupation with the child.

Parents should be made aware that surgery is only one component of the treatment. A child operated for cataract requires regular scheduled care for the first decade of life, and then every 1 to 2 years throughout life. However, to achieve the best visual outcome for the child, a long-term commitment from the parents is required. Visual prognosis can be explained to the parents based on the preoperative evaluation. Longer duration between onset of cataract and cataract surgery, unilateral cataract, asymmetrical bilateral cataract, presence of strabismus/nystagmus, poor preoperative VA, longer preoperative interocular AL difference, and associated ocular anomalies (such as JIA) are indicators for poor visual prognosis postoperatively.

The changing refraction will require frequent follow-up examinations. Glaucoma is known to develop even years after cataract surgery. Parents need to understand that their child may need serial EUA until the child is cooperative enough to get examined in the office. The treatment of VAO, strabismus, glaucoma, and rarely, decentered IOL, and synechiolysis, or removal of a loose stitch should be discussed with the parents. For eyes operated during early infancy, parents should be made aware that the first 6 months of follow-up is very crucial. Despite performing primary posterior capsulectomy and vitrectomy, many infant eyes develop VAO, and most eyes that develop VAO develop it in the first 6 postoperative months. Earlier detection (and treatment if needed) can help to achieve a better visual outcome. For eyes operated with an intact posterior capsule, parents should be made aware that the child may require a secondary procedure for PCO. Parents of children with lens implants are also made aware that glasses will likely still be needed postoperatively even when an IOL is implanted. In addition, glasses power may need to be changed frequently after surgery, because of changing refraction.

NEXT STEP

The surgeon may elect to prepare for surgery but delay the final decision on surgery until the time of the EUA. We routinely perform an EUA during the same session as

the cataract surgery. However, to do an EUA as a separate session is also an acceptable approach. As preoperative preparation for surgery we use dilating drops (ped combo, 2 mL 2% cyclopentolate, 0.5 mL 10% phenylephrine, 0.5 mL 1% tropicamide) every 5 minutes, three times. We give dilating drops to both eyes, as we also perform dilated examination of the fellow eye.

LABORATORY INVESTIGATION TO DETECT THE CAUSE OF CATARACT

As compared to unilateral cataract, laboratory investigation of bilateral cases is more rewarding. Exhaustive lists of possible laboratory investigations for a child with cataract can be found in several text books; however, in an otherwise healthy child, most physicians do not advise extensive laboratory and genetic investigations. After detailed evaluation, 86% of unilateral and 68% of bilateral cataracts have no discernible cause.²⁵ Based on history and examination, customized laboratory investigations can be advised. While recommending laboratory investigation, it is important to keep in mind that the common causes of cataract in children include intrauterine infections, metabolic disorders, and genetically transmitted syndromes. Since cataracts can be the presenting sign of diabetes, children with acquired cataracts of unknown etiology should be questioned about classic symptoms of diabetes, and evaluation for hyperglycemia should be performed. Children with Lowe syndrome have hypotonia, mental retardation, aminoaciduria, and an abnormal facial appearance with frontal bossing and chubby cheeks. The lens typically has a reduced anterior–posterior diameter.²⁶ In addition, these eyes have frequent association with glaucoma. If Lowe syndrome is suspected, the urine should be screened for amino acids. If there is history of maternal rash, fever, flu-like symptoms, or neonatal physical signs of intrauterine infection, then acute and convalescent TORCH titers should be obtained. Developmental pediatricians and clinical geneticist are experts in selective investigation based on characteristics of the child. These specialists are invaluable and should be consulted when appropriate.

EXAMINATION UNDER ANESTHESIA

Intraocular pressure should be checked as soon as possible after induction of anesthesia. Although we routinely use the tonopen in the operating room, if in doubt, we recheck IOP using the Perkins tonometer. In addition to high IOP, a difference of IOP between the two eyes is alarming. Cataracts and glaucoma can occur with congenital rubella and Lowe syndrome. The next step for us is to take keratometry measurements to avoid the problems associated with corneal drying. We use the Nidek handheld keratometer (Fig. 5.6). However, many other centers use an autorefractokeratometer for this purpose.



Figure 5.6. Handheld keratometry in operating room during examination under anesthesia. (Reprinted from Trivedi RH, Wilson ME. Pediatric cataract: preoperative issues and considerations. In: Wilson ME, Saunders RA, Trivedi RH, eds. *Pediatric Ophthalmology: Current Thought and a Practical Guide*. Berlin, Heidelberg: Springer, 2009, with permission.)

The automated readings should be recorded in diopters (D), and average K-value should be calculated for analysis of IOL power calculation. The accuracy of the handheld keratometer for cylinder axis measurement is reported to be less reliable than manual keratometer. However, as we are concerned with power and not axis for IOL power calculation, we think it is reasonable to use the handheld keratometer under EUA when the child is not cooperative for manual keratometry during office visit. The remaining examinations can be performed in any chronology: examine the eye using the operating microscope, immersion A-scan ultrasound for globe AL, horizontal corneal diameter, retinoscopy (if possible), pachymetry, and a retinal fundus examination. Some physicians use a slit-lamp attachment to operating microscope for evaluating location of the cataract.

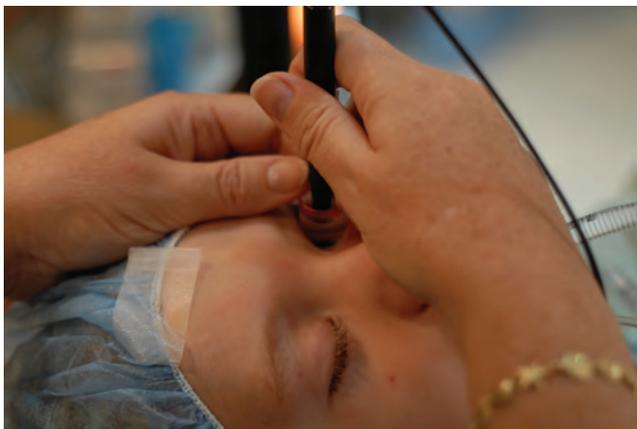


Figure 5.7. Immersion A-scan during examination under anesthesia. (Reprinted from Trivedi RH, Wilson ME. Pediatric cataract: preoperative issues and considerations. In: Wilson ME, Saunders RA, Trivedi RH, eds. *Pediatric Ophthalmology: Current Thought and a Practical Guide*. Berlin, Heidelberg: Springer, 2009, with permission.)

Immersion A-scan (Fig. 5.7) performed by a skilled ultrasonographer helps to reduce the incidence of postoperative refractive surprises. A shorter or longer AL in the eye with the cataract can be a sign of poor prognosis. Many A-scan instruments are available, and ensuring that the unit you used has been calibrated and is capable of accurate measurement is important. Readers are urged to refer to the specific technical instructions of the machine they are using. The instrument should have an oscilloscope screen such that true echo spikes are observed in determining axiality. The surgeon may need to transport the A-scan unit from the clinic to the operating room. The lack of A-scan ultrasound in operating room setting increases the difficulty of calculating the IOL power to use for pediatric cataract surgery. Measuring the AL of the eye using an A-scan is dependent upon the sound velocity the instrument is set at for the measurement.²⁷ Some instruments use an average velocity for the entire eye while others use individual velocities for each part of the eye. Because of the inversely proportional change in the axial ratio of solid to liquid as the eye increases in length, the average phakic velocity of a short 20-mm eye is 1,561 m/s (instead of the 1,555 m/s for 23.5 mm for AL). This factor amounts to only a small (0.25 D) error in extremes of AL.

When possible, at least the posterior pole of the fundus should be assessed by indirect ophthalmoscopy, looking particularly for underdevelopment or malformation of the disc or macula and the presence of abnormal pigmentation. When there is no view of the retina, a B-scan ultrasound (Fig. 5.8) should ideally be performed by the surgeon, not technician (see Chapter 21). B-scan ultrasound examinations are dynamic, and the surgeon must observe the study in motion rather than merely look at the selected freeze-frame images saved by the technician.

Although it is often not possible to do a preoperative cycloplegic refraction of an eye with a dense cataract, an uninvolved or less involved fellow eye should be refracted. The presence of refractive error in the fellow eye may help

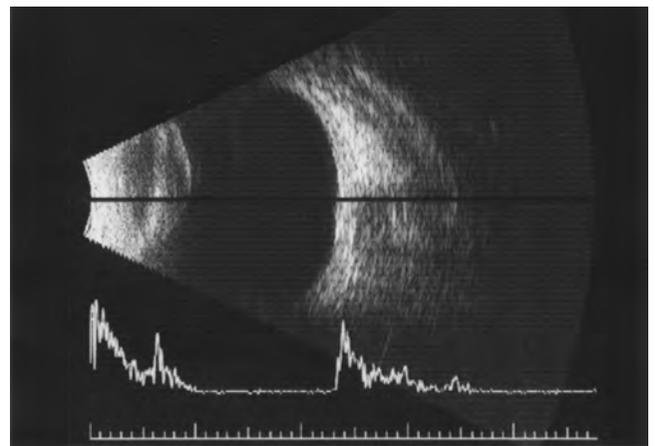


Figure 5.8. Preoperative B-scan in a 10-year-old boy with total cataract and severe developmental delay.

when deciding on an IOL power. In case of congenital anterior lens opacity (CALO), the associated anisometropia has been reported to be more amblyogenic than the lens opacity itself.²⁸ Patients with CALOs who have anisometropia of 1 D or more are 6.5 times more likely to develop amblyopia.

At the end of the EUA and before beginning the surgery, we visit with the parents to explain the findings of the EUA in brief. We explain what findings were expected and those that were not expected. We discuss any changes in the surgical plan that might be appropriate given the findings at EUA. This also gives the parents a good sense of how long the exam took and how long the surgery will take.

SELECTION OF IOL POWER

Implantation of a fixed-power IOL into an eye that is still growing makes it difficult to choose the IOL power to implant (see Chapter 7). Several nomograms have been published in the literature. However, we do not recommend the use of any published table alone for deciding IOL power. We typically do less under-correction compared to published tables, which are mainly to minimize myopia. These tables are only meant to help as a starting point toward appropriate IOL power selection, which is a multifactorial decision customized for each child based on many variables (age, laterality [one eye or both], amblyopia status [dense or mild], likely compliance with glasses, and family history of myopia).

SUMMARY

Pediatric cataract care is complex. The surgery itself is but one-step among many aimed at achieving normal visual function over a long life span. A comprehensive history and an ocular and systemic examination are important for preoperative decision-making in children with cataracts. Lengthy discussions between the parents/caregivers and the surgeon are common. The outcome is often better when the parents/caregivers are informed and committed partners with the ophthalmic team. Preoperative EUA is necessary for most children undergoing cataract surgery. It is important to make decisions in partnership with the parents. Taking the extra time to help parents understand the implications of the cataracts their child has and the options for treatment will save time later and will promote better compliance with medications, glasses, contact lenses, and occlusion therapy. An informed parent is usually a willing participant in the treatment of his or her child. The more they understand and accept the necessary steps, the better partners they will become in the battle for good visual function.

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6

Critical Periods for Visual Development and the Timing of Congenital Cataract Surgery

Eileen E. Birch and Anna R. O'Connor

CRITICAL PERIODS

The influence of early experience on development has been and continues to be a dominant theme in developmental neurobiology. The explosion of interest in visual system development was kindled in the early 1960s by the investigations of David Hubel and Torsten Wiesel into the complex organization of the primary visual cortex where, among other features, they were the first to describe the organization of monocular connections between the lateral geniculate nucleus and layer 4C of the visual cortex into ocular dominance columns and the higher-level binocular connections that provide sensitivity to binocular disparity.^{1,2} Following this extremely important initial work, they turned to the question of neurodevelopment, investigating how the highly specific response properties of cortical cells emerged during postnatal development.^{1,2}

Their research led to three influential ideas. *First*, there is a critical period for visual development that requires normal postnatal visual experience. Their earliest investigations looked at the effects of depriving one eye of vision during the first weeks of life. In the cat, normal visual experience leads to a visual cortex that has a normal distribution of ocular dominance, some left eye driven cells, some driven by the right eye, and most binocular cells. However, if one eye is deprived of vision during the critical early weeks, the visual cortex develops only cells connected to the nondeprived eye. If this same deprivation is performed at an older age, the critical period for visual development has already ended and there is no effect on ocular dominance. *Second*, not only is it possible to induce cortical changes by visual deprivation during the critical period, but there is also a critical period during which these changes can be reversed in whole or in part by forcing use of the previously deprived eye. Moreover, the critical period for rehabilitation may extend beyond the

critical period for visual development. *Third*, areas that are normally controlled by the deprived eye are not just empty areas, which lack connections due to disuse. Instead, the areas connected to the deprived eye actually shrink relative to the areas controlled by the normal fellow eye; in other words, there is competition between the eyes for cortical space during the critical period for visual development.

Although ophthalmology was developing an awareness of these important discoveries in neurobiology, there was a long delay before they were translated into clinical practice, particularly for the treatment of unilateral cataracts. As recently as 1980, papers in pediatric ophthalmology stated that there is “virtually no chance of achieving a good visual result” in children with congenital unilateral cataracts.³ In fact, it was not until the publication of two seminal papers in 1981 that pediatric ophthalmology took notice of the importance of critical periods for visual development and rehabilitation in the treatment of congenital unilateral cataracts. The first paper was the result of a collaboration between Creig Hoyt and Elwin Marg in which eight children had dense unilateral congenital cataracts removed at 7 hours to 41 days of age and all achieved visual acuity of 20/80 or better (≤ 0.6 logMAR), including two children who astoundingly achieved 20/20 (0.0 logMAR) vision.⁴ This paper provided the impetus for a major shift in thinking. The critical period paradigm altered the perception of congenital cataract from an irremediable cause of visual impairment to a leading *preventable* cause of visual impairment in children.

The second paper came from Richard Held's laboratory, using the newly developed forced-choice preferential looking technique to monitor visual acuity development in a few individual children following early surgery for dense congenital unilateral cataract.⁵ Despite an initially large visual acuity deficit, we could see for the first time the recovery of visual acuity with occlusion of the fellow

eye, its setbacks during periods of noncompliance, and continuation of recovery once occlusion therapy was restarted.

The dramatic change in clinical practice that quickly followed the publication of the seminal papers is illustrated in Figure 6.1. Prior to 1980, the median visual acuity outcome category following surgery for congenital unilateral cataracts was >20/1,200-light perception (>1.8 logMAR)^{3,6-11} and, for congenital bilateral cataracts, was >20/200 to 20/400 (1.0–1.3 logMAR).^{9,11} With the new emphasis on surgery during the first weeks of life to minimize the effects of visual deprivation on the developing visual system and careful monitoring of occlusion therapy, the median outcome category changed dramatically to 20/15 to 20/70 (–0.1 to 0.55 logMAR) for both unilateral and bilateral congenital cataracts.¹²⁻⁴⁰

MODELS OF THE CRITICAL PERIOD FOR VISUAL DEVELOPMENT: VISUAL DEPRIVATION

Numerous studies since 1981 have demonstrated that earlier treatment of dense congenital cataracts is associated with better visual acuity outcomes. More recent studies have explored the nature of the relationship between the timing of treatment and the long-term visual outcome.^{12,14} Three alternative models of the critical period have been evaluated:

- *Linear*: The earlier the surgery, the better the long-term visual acuity outcome.
- *Bilinear/Early Window*: There exists an early window of time during which the optimum long-term visual

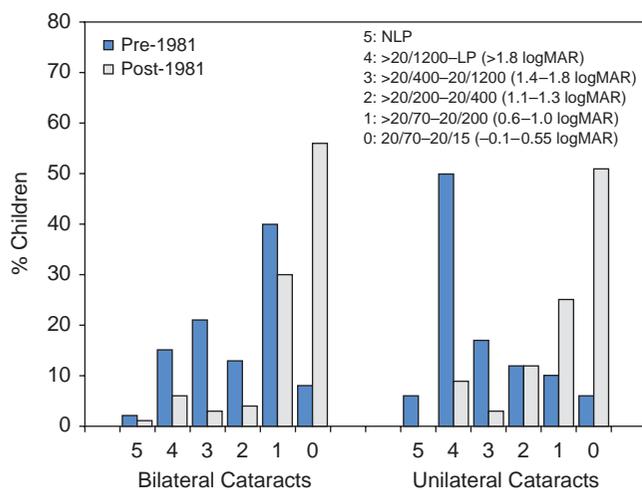


Figure 6.1. Visual acuity (logMAR) outcomes following surgery for dense congenital unilateral or bilateral cataracts from manuscripts published pre- and post-1981. Only children with age of onset of dense cataracts documented during the 1st week of life are included. Visual acuity categories correspond to the current World Health Organization International Statistical Classification of Diseases for Visual Impairment. NLP, no light perception; LP, light perception.

acuity outcome can be obtained; beyond this window, visual acuity outcome declines linearly with further delay in treatment.

- *Bilinear/Plateau*: Delay in treatment degrades long-term visual acuity outcome only during the first weeks or months of life, after which the outcome changes little with further delay.

Using a likelihood ratio test to determine which model best fit the data from children treated for dense congenital unilateral and bilateral cataracts, the slope (rate of change in outcome with delay in treatment) and critical ages were determined.^{12,14} The best-fit model for the critical period for visual deprivation due to dense congenital unilateral cataracts is shown in Figure 6.2. The *Bilinear/Early Window* model provides the best fit. During the first 6 weeks of life, there is little effect of age at surgery and long-term visual acuity outcome with a mean visual acuity outcome of 0.3 logMAR (20/40). After the critical age of 6 weeks, there is a steep decline in visual acuity outcome with further delay in treatment; each doubling in age at surgery (e.g., surgery at 10 weeks versus surgery at 20 weeks) is accompanied by a 0.3 logMAR drop in visual acuity (e.g., 20/80 versus 20/160).

The best-fit model for visual deprivation due to dense congenital bilateral cataracts is shown in Figure 6.3. The *Bilinear/Plateau* model provides the best fit. During the first 14 weeks of life, mean visual acuity decreased by one line with each 3 weeks’ delay in surgery, from

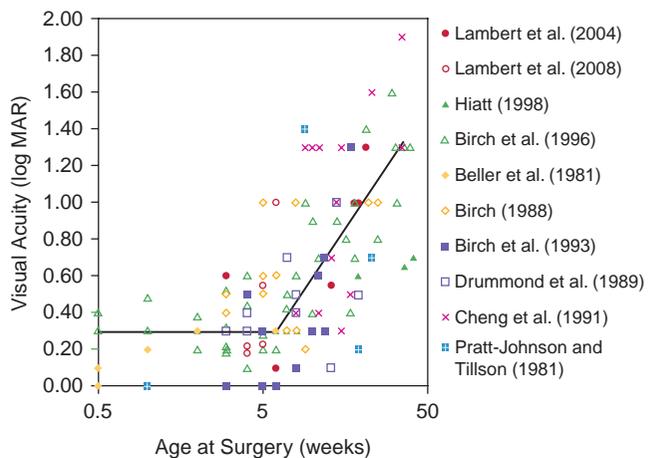


Figure 6.2. Best-fit model of the critical period for development with visual deprivation due to congenital unilateral cataract based on the data from Birch and Stager.¹⁴ The bilinear model exhibits a “latent period” that lasts up to 6 weeks of age during which the optimum long-term visual acuity outcome can be obtained; beyond 6 weeks, visual acuity outcome declines linearly with further delay in surgery. Data from other studies that evaluated the effect of age at surgery on long-term visual outcome have been added to the figure.^{4,13,16-18,24,28,30,36} Only children with age of onset of dense cataracts documented during the first week of life are included. Lambert et al. (2004)²⁸; Lambert et al. (2008)³⁰; Hiatt (1998)²⁴; Birch et al. (1996)¹⁴; Beller et al. (1981)⁴; Birch (1988)¹³; Birch et al. (1993)¹⁶; Drummond et al. (1989)¹⁸; Cheng et al. (1991)¹⁷; Pratt-Johnson and Tillson (1981).³⁶

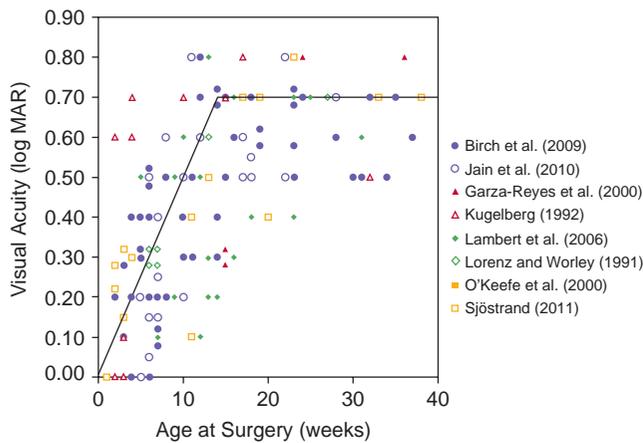


Figure 6.3. Best-fit model of the critical period for development with visual deprivation due to congenital bilateral cataract based on the data from Birch et al.¹² Delay in treatment degrades long-term visual acuity outcome only during the first 14 weeks, decreasing by one line with each 3 weeks' delay in surgery; after the critical age of 14 weeks, there is little change in visual acuity outcome with further delay in treatment up to at least 40 weeks. Data from other studies that evaluated the effect of age at surgery on long-term visual outcome have been added to the figure.^{11,20,27,29,34,35,39} Only children with age of onset of dense cataracts documented during the first week of life are included. Birch et al. (2009)¹²; Jain et al. (2010)²⁹; Garza-Reyes et al. (2000)²⁰; Kugelberg (1992)²⁷; Lambert et al. (2006)²⁹; Lorenz and Würle (1991)³³; O'Keefe et al. (2000)³⁵; Sjöstrand et al. (2011).³⁹

0.2 logMAR (20/30) at 1 week to 0.6 logMAR (20/80) at 14 weeks. After the age of 14 weeks, there is little change in visual acuity outcome with further delay in treatment, at least up to 40 weeks; mean visual acuity outcome for surgery delayed until 14 to 40 weeks is 0.6 logMAR (20/80).

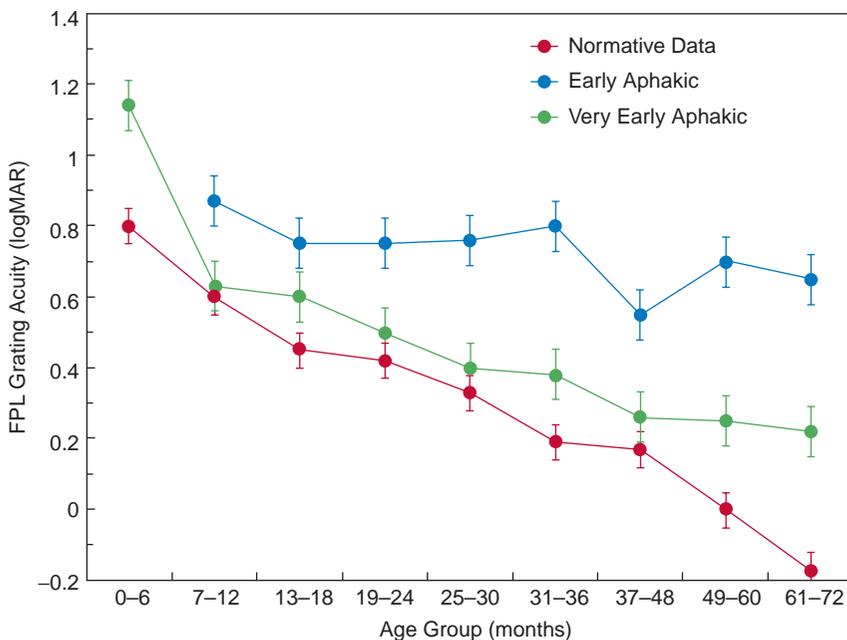


Figure 6.4. Grating acuity development during the first 6 years of life assessed by forced-choice preferential looking for children who had surgery for dense congenital unilateral cataracts at 1 to 6 weeks (very early) or 2 to 8 months (early). Also shown are normative data.⁴⁸ (From Birch EE, Hale LA. Criteria for monocular acuity deficit in infancy and early childhood. *Invest Ophthalmol Vis Sci* 1988;29:636-643.)

CRITICAL PERIOD FOR REHABILITATION

Following the influential studies by Held et al.,^{5,41,42} there were a large number of studies using the forced-choice preferential looking or visual evoked potential technique that showed that these visual acuity assessments were useful in guiding occlusion therapy, providing regular feedback on the progress of visual rehabilitation after each change in treatment protocol and/or a proxy measure for compliance with the prescribed occlusion therapy regimen.^{15,16,34,43-47} The ability to quantify visual acuity during the first years of life gave us a new means of understanding the critical period. The effect of age at surgery on long-term visual acuity outcome described in the section above helped us to define the critical period from the perspective of sensitivity to abnormal visual experience. On the other hand, evaluation of the development of visual acuity during infancy and early childhood allowed us to examine the critical period from the perspective of response to attempts to reverse the effects of visual deprivation through optical correction and occlusion therapy. In the example shown in Figure 6.4, visual acuity development of children with dense congenital unilateral cataracts who had “very early surgery” (by 6 weeks of age) was compared with the maturation of children who had surgery at 2 to 8 months.¹⁶ When tested within a few weeks of surgery, both groups of children initially had a visual acuity deficit of approximately 0.3 logMAR (three lines) relative to normal infants; that is, the effect of visual deprivation on visual acuity development was about the same. When optical correction and occlusion therapy were initiated at 2 to 7 weeks, the very early group quickly recovered from much of the initial deficit and achieved a mean final visual acuity within 0.3 logMAR of normal controls. In

contrast, the children who had begun optical correction and occlusion treatment at 2 to 8 months showed a slower and diminished response and had a final acuity that was 0.8 logMAR below normal controls. Thus, even though the visual acuity deficit that resulted from early deprivation was similar in magnitude, the later start to optical correction and occlusion therapy resulted in less response to treatment. As an additional practical benefit, these studies were the first to demonstrate that the visual acuity deficits detected by forced-choice preferential looking as early as 2 years of age were predictive of long-term logMAR letter acuity outcome at 5 years of age.

CRITICAL PERIOD FOR VISUAL DEVELOPMENT: INTEROCULAR COMPETITION

Hubel and Wiesel clearly demonstrated that monocular visual deprivation during the critical period results not simply in a scarcity of cortical connections from the deprived eye, leaving areas of the visual cortex devoid of input, but shrinkage relative to the areas controlled by the normal fellow eye. This shrinkage is the result of competition between the eyes for cortical connectivity during the critical period. In fact, an hypothesis that seeks to explain the 6-week “latent period” for the treatment of dense congenital unilateral cataracts is that the interocular competition is not yet active in these first few weeks of life; the only effect of the cataract on the developing visual cortex is the effect of deprivation/disuse. One straightforward prediction of this hypothesis is that we would expect similar visual acuity deficits in children with unilateral cataracts and bilateral cataracts if we measure visual acuity immediately after surgery and optical correction as long as surgery occurs during the first 6 weeks. Beyond the age of 6 weeks, we would expect the effect of interocular competition to result in more

severe postoperative visual acuity deficits in children with unilateral cataracts than bilateral cataracts. This prediction fits the published data⁴⁹; forced-choice preferential looking grating acuity deficits in the immediate post-treatment period were similar for unilateral and bilateral cataracts with surgery during the first few weeks of life but, if surgery was delayed beyond the latent period, children treated for unilateral cataract had significantly larger post-treatment grating acuity deficits than children with bilateral cataracts.

Contrast sensitivity data also support the hypothesis that interocular competition is not present during the first weeks of life.⁴⁹ Children treated for bilateral cataracts experience only the effect of visual deprivation and show virtually no contrast sensitivity deficit for low spatial frequencies, but a significant deficit for higher spatial frequencies.^{49,50} Children who have surgery for unilateral cataracts during the first few weeks of life show a remarkably similar pattern of contrast sensitivity deficits, suggesting that they too are experiencing only the effect of visual deprivation.⁴⁹ With delay in surgery for unilateral cataract, however, the added effect of interocular competition results in deeper and nearly uniform contrast sensitivity deficits at all spatial frequencies.^{49,50}

BEYOND VISUAL ACUITY

Most studies of outcome following treatment of dense congenital or infantile cataracts have evaluated the effects of age at surgery, duration of visual deprivation, and laterality (unilateral versus bilateral) on long-term visual acuity outcome. However, visual deprivation during infancy can also alter the maturational course of binocular vision, ocular motor function, reading ability, and fine motor skills.

Stereoacuity

Studies that have documented age at surgery and stereoacuity outcome in children treated for dense congenital cataracts are summarized in Table 6.1.

Table 6.1 PREVALENCE OF GOOD STEREOACUITY AND OCULAR MOTOR OUTCOMES FOLLOWING SURGERY FOR DENSE CONGENITAL CATARACTS^a

	Stereoacuity	Strabismus	Nystagmus
Unilateral Congenital Cataracts			
Surgery ≤6 wk	10/24 (42%) ^{25,40,51–54}	43/62 (69%) ^{16–18,25,37,40,45,52,53,55,56}	6/16 (38%) ^{45,55,56}
Surgery 7–12 wk	2/11 (18%) ^{25,40,51,54,57}	35/41 (85%) ^{16–18,37,38,40,45,56,58}	5/7 (71%) ^{45,56}
Surgery >12 wk	3/27 (11%) ^{25,57,59}	75/82 (91%) ^{7,10,17,18,24,37,38,56,58,59}	2/2 (100%) ⁵⁶
Bilateral Congenital Cataracts			
Surgery ≤6 wk	4/9 (44%) ⁶⁰	12/18 (67%) ^{12,55,56,60}	4/4 (100%) ^{55,56}
Surgery 7–12 wk	0/3 (0%) ⁶⁰	17/22 (77%) ^{12,56,60}	6/6 (100%) ⁵⁶
Surgery >12 wk	0/2 (0%) ^{59,60}	10/12 (83%) ^{10,12,59}	4/4 (100%) ⁵⁶

^aOnly children who were documented to have dense cataracts present by the 1st week of life were included. Nystagmus data were included only if the waveform was assessed. Stereoacuity data were included only if they were obtained with standardized clinical test. Nil stereoacuity was classified as a poor stereoacuity outcome. INS and FMNS were considered poor ocular motor outcomes.

For both unilateral and bilateral congenital cataracts, surgery prior to 6 weeks is associated with a higher prevalence of measurable stereoacuity than later surgery.^{25,40,51–54,57,59,60} In addition, there is preliminary evidence that a reduced or progressive protocol for occlusion therapy following surgery for unilateral cataracts may be of benefit for the development of stereoacuity.^{40,51,52,54}

Ocular Motor Function

Little is known about the critical period for ocular motor outcomes. Nystagmus has been reported to be more prevalent among children with bilateral than unilateral cataracts.^{22,61,62} In addition to laterality, both age at surgery and duration of visual deprivation have been proposed as risk factors for development of nystagmus.^{12,14,25,49,58,63} The majority of studies do not report the incidence or prevalence of strabismus. Even when the incidence or prevalence of strabismus is addressed, there are inconsistencies among studies in methods of assessment and classification of strabismus and in whether they report the initial deviation or the deviation at the most recent visit. Even fewer studies used eye movement recording systems to assess fixation, or the presence of associated ocular oscillations, relying on clinical observation despite the inadequacy of this approach for determining the presence or nature of any involuntary fixation instability or nystagmus.⁵⁸ Thus, the critical period for adverse ocular motor outcomes remains ill defined.

Two recent studies have begun to address these gaps in our evidence base.^{55,56} What is clear from these studies is that normal visual experience during the first weeks of life is a key factor in ensuring normal ocular alignment and stable fixation. As summarized in Table 6.1, children who had dense cataracts for even a very brief period during infancy had a high probability of developing strabismus and nystagmus. Overall, strabismus has been reported to occur in >75% of children treated for congenital cataracts.^{7,10,12,13,16–18,24,25,37,38,40,45,52,53,55,56,58–60} Children with congenital cataracts who are treated by 6 weeks of age have reduced risk for strabismus than those treated later.⁵⁶ Following cataract surgery during the first 6 weeks of life, esotropia was much more frequently observed than exotropia, with onset of esotropia most often occurring during the first year of life.^{45,55,56} Exotropia, when it occurred, was almost invariably associated with a poor visual acuity outcome, due to late surgery and/or poor compliance with postoperative treatment.^{45,55,56}

Most children treated for congenital or infantile cataracts develop nystagmus with the infantile nystagmus syndrome (INS, previously termed “congenital nystagmus”) or fusion maldevelopment nystagmus syndrome waveform (FMNS, previously termed “manifest latent nystagmus”).^{55,56} INS and FMNS are always associated with strabismus.^{55,56} For congenital cataract, reduced risk for nystagmus is associated with surgery by 6 weeks of age.⁵⁶ Nystagmus

was always bilateral, although significant asymmetries in waveform and intensity were present, particularly in children who had been treated for unilateral congenital cataracts after 6 weeks of age.^{45,55,56} The asymmetry may cause the nystagmus to appear “clinically silent” in one of the two eyes, especially when there is a large interocular difference in visual acuity. However, ocular motor recordings in patients with clinically unilateral oscillations usually reveal the binocular, but asymmetric, presence of nystagmus.^{55,56}

Reading

Reading may be an important measure of the success of rehabilitation of eyes treated for congenital cataract because it is the foundation for academic success and there is a clear link between academic success in childhood and long-term social and economic outcomes.⁶⁴ To date, there is only a single study of reading in children treated for congenital unilateral cataracts, in which reading rate, accuracy, and comprehension were evaluated for the affected eye with the fellow eye occluded.⁶⁵ When forced to rely on the affected eye alone to read grade-level text, children treated for congenital unilateral cataract as a group read more slowly, had poorer reading accuracy, and poorer comprehension than their peers. The slower reading rate and reduced accuracy may result when the visual acuity of the affected eye is close to the critical reading print size for grade level.⁶⁶ Similarly, the lower comprehension score may result because reduced visual acuity causes the child to spend more time visually decoding graphemes⁶⁷ and, therefore, being less attentive to meaning. On the other hand, when the reading ability of a subgroup that had surgery by 6 weeks of age and visual acuity outcomes of 0.1 to 0.3 logMAR were tested, their reading rate, accuracy, and comprehension did not differ significantly from their peers. Thus, the 6-week critical period for treatment of congenital unilateral cataract, at least indirectly through its influence on visual acuity outcome, can have long-term consequences for vision-related activities.

No studies yet have evaluated binocular reading performance of children affected by unilateral or bilateral cataracts, that is, as they would normally read in school. Because reading requires the interaction of visual and ocular motor function, we might expect that binocular reading performance will be degraded by the bilateral nystagmus that develops in most children with congenital or infantile cataracts.^{55,56} Two recent studies have demonstrated that adults with INS read more slowly than normal controls, regardless of their visual acuity.^{68,69} However, reading speed was higher than expected, because the adults with INS utilized a variety of adaptive strategies to enhance their reading performance, which allowed them to adapt their nystagmus to achieve the desired gaze patterns, for example, to allow involuntary slow-phase drift to run across lines of text.⁶⁸ The reduced

risk for nystagmus associated with cataract surgery during the critical first 6 weeks of life could potentially also support better long-term reading speed outcomes.⁵⁶

Fine Motor Skills

No studies yet have evaluated fine motor skills of children affected by unilateral or bilateral cataracts. There have been a number of recent studies that have explored the impact of amblyopia (etiology either unspecified or various etiologies mixed together within the cohort) and reduced or nil stereoacuity on fine motor skills. Fine motor skills performance (cutting, drawing, copying, sorting) of children with amblyopia was poorer than age-matched control children, more so in tasks that required speed and dexterity rather than tasks that required accuracy and control.⁷⁰ In reaching and grasping, amblyopic children program slower, more cautious movements and spend almost twice as long in the final approach before closing in and applying their grip.⁷¹ This latter deficit appears to be a consequence of nil stereoacuity rather than the amblyopia per se.⁷¹ Amblyopic children with nil stereoacuity also make more spatial errors in initially positioning their grip of static objects⁷¹ and in timing their grasp of moving objects.⁷² Although severity of amblyopia and severity of stereoacuity deficit often go hand-in-hand, two recent studies reported analyses that were able to demonstrate that binocular performance on fine motor tasks deteriorated in association with reduced or nil stereoacuity, but not with visual acuity of the amblyopic eye.^{73,74} Moreover, there is some evidence that, although fine motor skill performance is best with normal stereoacuity, performance with reduced stereoacuity is better than with nil stereoacuity.⁷⁴ Thus, the rehabilitation of at least reduced stereoacuity that is possible when surgery occurs during the critical first 6 weeks of life^{25,40,51-54,57,59,60} and a part-time or progressive postoperative occlusion therapy protocol for unilateral cataracts^{40,51,52,54} also may be of benefit for fine motor skills.

SUMMARY

Congenital and infantile cataracts are an important cause of lifelong visual impairment, ocular motor dysfunction, and decreased ability to perform common visually guided tasks. The developing visual system is profoundly affected by visual deprivation and, in unilateral cases, by interocular competition. Successful rehabilitation requires prompt surgical intervention and closely monitored optical correction and occlusion therapy.

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7

Calculation and Selection of Intraocular Lens Power for Children

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Choosing an intraocular lens (IOL) power in an adult is driven by the desire for the immediate outcome of emmetropia. Choosing an IOL power in children is also driven by the postoperative goal refraction. But in contrast to adults, the initial refractive goal for a child is driven by long-term outcome, and we think that it is best to start with this goal in mind: good vision when the child matures. This outcome goal can be divided into three parts: emmetropia in adulthood, good visual acuity as an adult, and a manageable course of refraction between IOL implantation and adulthood. Incorporated in these goals is the need to treat amblyopia and management of the changing refractive error as the child's eye grows. Thus, selection of an IOL power is directly related to the ophthalmologist's plan for managing amblyopia and the child's initial and future refractive error.

In this chapter, we review the elements that are crucial in selecting an initial postoperative refractive goal: IOL calculation, amblyopia management, anisometropia management, and the logarithmic growth of the eye. Finally, we suggest a strategy that takes into account age and whether the cataract surgery is uni- or bilateral.

IOL CALCULATION

Calculating an IOL power in an adult is simple: the surgeon chooses a power to give emmetropia based on measurements of axial length (AL) and cornea power (K) using one of the formulas designed for adult eyes, such as the Hoffer Q, Haigis, Holladay, or SRK-T formula; in published series they show a mean absolute error of <0.6 D (diopter).¹ In contrast, the calculation of IOL power in children is complicated by less accurate biometry, the larger effect of measurement error in small eyes, and possibly by the lack of a published IOL formula specifically for children.

Studies of IOL implants in children demonstrate larger formula prediction errors than are found when the same formulas are used in adults. Mezer et al.² found

that the mean difference between the predicted and the actual postoperative refractions was slightly more accurate using theoretic formulas (1.06 D versus 1.22 D with regression formulas). Andreo et al.³ studied 47 pediatric patients, age 0.25 to 16 years. They measured the initial postoperative pseudophakic refractions and compared them to those predicted by four formulas (SRK-II, SRK-T, Holladay, and Hoffer Q). They found that the average initial postoperative refractive error was between 1.2 and 1.4 D. Moore et al.⁴ performed a retrospective review of 50 pediatric eyes undergoing secondary IOL implantation, and found a mean absolute value prediction error of 1.64 D.

Biometry (measurement of AL and K) in young children is typically performed in the operating room with the child asleep under general anesthesia, and is usually less precise than in adults. The nonstandard conditions for biometry limit the tools to those that are portable to the operating room; some instruments used for biometry in adults such as the IOLMaster cannot be used. The AL may be difficult to measure accurately because the anesthetized child cannot voluntarily align his or her line of sight with the axis of measurement; in addition, a small amount of pressure from the ultrasonic A-scan transducer can easily deform a child's soft cornea. The cornea power is also difficult to measure accurately: artifacts induced by drying or by pressure on the eye from a speculum can distort the cornea curvature.

Errors of biometry in small eyes lead to larger errors in IOL power calculation than do the same errors in adults. This is because the effect of these errors is inversely related to the size of the eye: a 0.1-mm error of measurement in a 24-mm eye is an error of 0.4%; the same error in a 16-mm eye is an error of 0.6%. We calculate that this 0.1 mm error would result in an absolute refractive error of 0.23 D in the 24-mm eye and 0.57 D in the 16-mm eye (Table 7.1). Thus biometric errors are propagated by IOL calculation formulas and magnified in small eyes.

Table 7.1

CALCULATION OF THE REFRACTIVE ERROR INDUCED BY A 0.1 MM ERROR IN THE MEASUREMENT OF AL IN TWO EYES, A 16 MM INFANT EYE VERSUS A 24 MM ADULT EYE

AL (mm)	16.0	16.1	24.0	24.1
K (D)	49.2	49.2	43.9	43.9
IOL power (D)	53.0	53.0	18.0	18.0
Refraction (D)	0.00	-0.57	0.00	-0.23
Induced error (D)		-0.57		-0.23

All modern IOL calculation formulas are based on adult eyes. They assume that the IOL will rest in its usual position in the adult-sized eye, in relation to AL, cornea curvature, and (in some formulas) other parameters. In addition to their basis in optics theory, some of the formulas are empirically modified to optimize results in adult eyes. It is unknown precisely how much children's eyes vary from adult proportions. Some of the observed errors in the calculation of initial postoperative refractions after pediatric IOL implantation may be due to incorrect assumptions in these formulas or to unusual proportions in pediatric eyes with some types of cataract. However, most children's eyes eventually grow to adult proportions, so adult formulas may be better in the end than a formula designed for pediatric eyes.

The combination of increased errors of biometry, the greater effect of these errors on IOL calculations, and the lack of an IOL formula for children may be the primary cause of the observed lesser accuracy of IOL calculations in children than in adults. In addition, postoperative refractions are more difficult to obtain precisely in children than in adults, and there may be a small growth of the eye between IOL implantation and the first refraction; these additional reasons may help to explain why published studies find a greater error in pediatric eyes. Despite these causes, a carefully done recent study by Trivedi et al.⁵ of 45 eyes with surgery at a mean age of 3.56 years found a remarkably low mean absolute error of 0.68 to 0.84 D, using four theoretic IOL formulas; the Holladay 2 formula gave slightly better predictions than the others. The authors of this study clearly used careful measurement techniques including immersion ultrasound for AL; these results may be close to the best possible in theory.

AMBLYOPIA MANAGEMENT

High refractive error in unilateral aphakia makes it very difficult to obtain good vision in the long term due to amblyopia. Children with unilateral aphakia are generally treated with contact lenses that are not always worn

continually; the contact lens can also cause blur when it is displaced to the edge of the pupil in daily life. Even short intervals of uncorrected aphakia can cause dense and long-term amblyopia.⁶ The hope of IOL implantation is that a constantly sharp retinal image will simplify amblyopia management and result in better vision. A large long-term retrospective study found a better mean visual acuity for pseudophakic eyes than aphakic eyes in all age groups (Table 2 in McClatchey et al.⁷). A small study of infants who received a primary IOL after extraction of dense congenital unilateral cataract showed improvement from a mean visual acuity of 20/170 at 6 months to 20/85 at 12 months and 20/54 at 4 years. Visual acuity in the IOL group at 4 years was similar to that of children who had good-to-excellent contact lens wear compliance and better than that of children who had moderate-to-poor compliance.⁸

A survey of the American Association for Pediatric Ophthalmology and Strabismus (AAPOS) in 2001 found that most members preferred to leave infants aphakic after cataract surgery and use contact lenses for initial correction.⁹ It is certainly possible to achieve good vision with this strategy: two of our patients with unilateral cataract surgery who were left aphakic have achieved 20/25 vision, thanks to constant use of contact lenses and consistent patching.

The Infantile Aphakia Treatment Study is a randomized, multicenter clinical trial of 114 infants with a unilateral congenital cataract comparing IOL implantation at cataract surgery to leaving the infant aphakic. Preliminary results of logMAR grating visual acuity at 1 year of age failed to show a significant difference between the treated eyes in the two groups (contact lens group, 0.80; IOL group, 0.97; $P = 0.19$).¹⁰ In addition, there were more patients with intraoperative complications (28% versus 11%; $P = 0.03$), adverse events (77% versus 25%; $P < 0.0001$), and additional intraocular surgeries (63% versus 12%; $P < 0.0001$) in the IOL group than the contact lens group.¹¹

Thus the evidence on whether IOL implantation improves ultimate amblyopia management of children with unilateral cataract surgery is incomplete, and age may be a factor. Certainly IOL implantation is not sufficient by itself in most children, and the surgeon must plan to manage amblyopia using standard treatment techniques: patching and correction of residual refractive error. Frequent and diligent follow-up must be vigorously pursued, and parents must be taught the importance of patching, often repeatedly.

ANISOMETROPIA MANAGEMENT

Anisometropia is of great concern for children who have unilateral cataract surgery; bilateral cataract surgery generally results in nearly equal refractions both initially and

with increasing age, though the two eyes of a child may grow at different rates in some. The problems due to the often substantial anisometropia in children with unilateral pseudophakia are best broken down into several categories:

1. *Aniseikonia*, a significant difference in the image size between the eyes. Due to Knapp Law, aniseikonia is not a significant issue in axial anisometropia, but instead occurs when there is spectacle correction of anisometropia after cataract surgery. This artifact is due to the difference in magnification from spectacle lens power disparity between the eyes; it generally causes adults to have asthenopia when wearing spectacles to correct ≥ 3.5 D of anisometropia after cataract surgery. Contact lenses or corneal refractive surgery greatly reduce the aniseikonia and eliminate symptoms.
2. *Anisophoria*, the smoothly varying prismatic effect of off-axis gaze through spectacle lenses of different powers due to Prentice Law.¹² This is not a great problem for children with poor binocularity (e.g., many of those with anisometropic hyperopia and amblyopia), but some children with unilateral pseudophakia and spectacle correction of significant anisometropia will notice diplopia when looking off-axis. We postulate that this is most bothersome for many when the spectacle-induced misalignment puts the second image outside of Panum fusional space: this corresponds to about 3 degrees of misalignment, and would be induced by 4 D of anisometropia when gazing through the spectacles 1.5 cm off-axis. In contrast, contact lenses move with the eye, thus completely eliminating anisophoria.
3. *Amblyopia management* is more difficult when the amount of anisometropia is greater. We think it best to estimate the future compliance with amblyopia management (including both glasses and patching or penalization) when choosing how much anisometropia to induce initially. Of note, in children whose normal eye is moderately hyperopic (+1 to +4), the amount of anisometropia found on cycloplegic exam underestimates the amount present when a child takes off the glasses and accommodates to see with the normal eye.
4. *Changes in anisometropia with age* are inexorable but difficult to predict. A young child who initially has 3 D more hyperopia in the pseudophakic eye may eventually develop significant myopia in that eye, resulting in greater anisometropia due to the myopia than he or she had from the initial hyperopia. One strategy is to use the graph output from an IOL calculator^{13,14} to choose the age at which the refraction is likely to cross from hyperopia to myopia.

Therefore, the clinician who manages children with unilateral pseudophakia must be prepared to manage these several aspects of anisometropia. In the case of a child who wears spectacles, this becomes significant when the

anisometropia is greater than about 4 D. Depending on the age, cooperation with treatment, and ease of contact wear, this can be managed by switching from spectacles to contact lenses. In some, reducing the power of the minus spectacle lens in the pseudophakic eye by about a diopter will allow good near vision with that eye while reducing the aniseikonia and anisophoria to acceptable levels.

THE LOGARITHMIC GROWTH OF THE EYE

Gordon and Donzis¹⁵ published a cross-sectional study of biometry of normal children, and calculated the IOL power required for emmetropia for average eyes across a range of ages. They found a rapid reduction in lens power in infancy, followed by a less rapid reduction in early childhood. McClatchey and Parks¹⁶ studied a large number of aphakic eyes with long follow-up, and found that the mean refraction followed a logarithmic curve from infancy through age 20 years, with a very high correlation ($r^2 = 0.97$). These data were shown to closely match the curve predicted by the earlier Gordon and Donzis data, indicating that cataract extraction without IOL implantation does not affect the growth of the eye in childhood.¹⁷

The slope of the line in a graph of aphakic refraction versus log of age is a straight line. The slope in the logarithmic model can be calculated for pseudophakic eyes by mathematically removing the optical effect of the IOL.^{18,19} The slope is not a direct measure of myopic shift, so it was named rate of refractive growth (RRG), because it is a metric indicating how fast the refraction changes due to the growth of the eye. A study of pooled long-term data from eyes of pseudophakic children from around the world found that RRG was slightly lesser in pseudophakic eyes than in aphakic eyes.²⁰ Calculation of RRG eliminates the confounding factors of nonlinear growth of children's eyes and the variables of age at surgery, length of follow-up, and variations in IOL power.

In order to eliminate the artifact inherent in a logarithmic model of the asymptote at zero (age 0 year [yr]), McClatchey and Hofmeister²¹ shifted the age frame to account for the growth of the eye in utero. This revised model is called RRG2, defined by this formula:

$$\text{RRG2} = (\text{Refraction}_2 - \text{Refraction}_1) / (\log(\text{Age}_2 + 0.6 \text{ yr}) - \log(\text{Age}_1 + 0.6 \text{ yr}))$$

where log is base 10, refractions are measured at the spectacle plane, Refraction_1 and Age_1 are at the younger age and Refraction_2 and Age_2 are at the older age. Because subtraction of the logarithms of two numbers is mathematically equivalent to taking the logarithm of the ratio of the two numbers, RRG2 has units of diopters.

When the ratio of adjusted final to initial ages ($(\text{Age}_2 + 0.6 \text{ yr})/(\text{Age}_1 + 0.6 \text{ yr}) = 10$), then the denominator in RRG2 is $\log(10) = 1.0$. One way to understand the logarithmic model is to consider that when an aphakic child's adjusted age increases 10-fold, his or her refraction will change by the quantity given by the RRG2 value. Since the mean RRG2 for pseudophakic eyes was about -6.7 D , these eyes had a mean myopic shift of 6.7 D when the adjusted ages compared were 10-fold different, for example, from age 1.4 years to age 19.4 years (where the adjusted ages would be 2 and 20 years, respectively).

A study of 45 patients with unilateral cataract surgery (with or without IOL implantation) failed to find a statistically significant difference in AL growth between the operated eyes and fellow nonoperated eyes.²²

The logarithmic model is useful in research and practice. By converting an inherently nonlinear system into a linear transform it allows a researcher to compare groups of dissimilar eyes, just as the use of logMAR visual acuity allows a researcher to calculate the mean visual acuity of a group of eyes. It allows the clinician to predict the future refractions of a child, within the limits imposed by the natural variance in RRG2. Two computer programs have been published that allow surgeons to predict the likely refractive trajectory of a child who has an IOL implant.^{23,24}

RRG2 has not yet been fully validated, especially in the youngest infants and in older pseudophakic children. The validity of this model in pseudophakic infants with surgery before 3 months has not been tested, and will likely remain unknown until the Infantile Aphakia Treatment Study 5-year results are published. The validity of this model in older children is controversial: Superstein et al.²⁵ studied 92 pseudophakic children with a mean age at surgery of 7.3 years and found a relatively small myopic shift over a relatively short period of follow-up. They found that their patients' refractive trajectories did not match that predicted by RRG; however, the length of follow-up was not sufficient to allow proper calculation of RRG for most of these eyes.

Another factor that must be considered is the large variance between eyes: some eyes grow faster than others, and we do not yet know why. The quantity of the standard deviation in RRG2 is approximately 45% of the value of RRG2. The practical effect of this is that the prediction of future refractions cannot be precise.

MAGNIFICATION OF MYOPIC SHIFT DUE TO THE OPTICS OF THE IOL IN A GROWING EYE

As a pseudophakic eye grows, the distance from the lens to the retina increases, moving the focal point of the IOL anterior in the eye. This magnifies the total myopic shift of the eye, when compared to an otherwise identical eye without an IOL. This magnification of myopic shift is

analogous to the refractive difference between contact lenses and spectacles: the vertex distance of the spectacle lens has a large optical effect for high-power lenses.

For example, an aphakic child's eye that is corrected with a $+15 \text{ D}$ lens at the spectacle plane would require a contact lens of $+18.3 \text{ D}$. The need for an increased power at the cornea plane is due to the optical effect of vertex distance: for example, an eye that required a $+24 \text{ D}$ spectacle lens would require a $+33.7 \text{ D}$ contact lens.

Just as in these contact lens examples, where vertex distance has a greater effect at high refractive power lenses, high-power IOLs result in a greater myopic shift for the same increase in AL as the eye grows. We calculated what the myopic shift would be if a child was left aphakic or given one of three IOL powers, assuming that the physical growth of the eye was unaffected by the presence of an IOL or its power. The results (Table 7.2) show that using a high-power IOL can result in a significantly larger myopic shift than using a lower-power IOL, for optical reasons alone.

INITIAL POSTOPERATIVE GOAL REFRACTION

Published preferences of initial postoperative goal refraction range from myopia to hyperopia that varies with age. There is no evidence that any particular strategy results in better long-term visual outcomes. Early in the experience of IOLs in children, some surgeons chose to use an adult-power IOL.^{26,27} Some cataract surgeons who operate primarily on adults prefer initial emmetropia or myopia,²⁸ at least after infancy,²⁹ but most pediatric ophthalmologists prefer hyperopia that varies with age.

Rapid growth of infant eyes after cataract surgery results in large myopic shift in these eyes. Lambert et al. did cataract surgery and IOL implantation on 11 infants with unilateral congenital cataracts who had a mean age of 10 weeks; they found a mean myopic shift of 5.5 D a year after surgery.³⁰ In a study of 16 children with bilateral cataract surgery and IOL implantation in the first year of life, Wilson et al. found a mean myopic shift of 6.2 D after

Table 7.2

THE OPTICAL EFFECT OF IOL POWER ON MYOPIC SHIFT, BASED ON A HYPOTHETICAL EYE THAT HAS CATARACT SURGERY AT AGE 6 MONTHS^a

Age (yr)	IOL Power (D) in Top Row, Refractions Below		
	0 (Aphakic)	38	20
0.5	+18.8	-2.7	+9.5
20	+10.6	-17.4	-1.0
Total myopic shift	-8.2	-14.7	-10.5

^aRefractions are measured at the spectacle plane.

a mean follow-up of 22 months.³¹ Plager et al.³² recommended initial postoperative hyperopia ranging from +5.0 D at age 3 years down to plano at age 13. They suggested that some children with unexpectedly larger myopic shift after IOL implantation may have a genetic predisposition to myopia. Eibschitz-Tsimhoni et al. noted that myopic shift after IOL implantation is greatest in the youngest patients, and there is a marked variability in postoperative refraction.³³ They note that there is no study demonstrating a visual advantage of any particular refractive strategy.

RECOMMENDATION TABLES

Many recommendation tables have been published over the last decade by pediatric ophthalmologists, representing years of experience in managing these children. Although none has data to demonstrate better long-term results than any other, we think that these tables may be a good way to choose an initial postoperative refraction in children (Table 7.3).

Because children with bilateral cataract surgery are not subject to anisometropia, it may be justifiable to leave them with a greater initial hyperopia than children with unilateral cataracts. To illustrate this we have developed two tables with suggested initial postoperative goals based on age, one for unilateral cataracts and one for bilateral cataracts (Tables 7.4 and 7.5). Because IOL implantation has greater risks in infants, the authors of this chapter would choose to leave infants with bilateral cataracts aphakic up through age 1 or 2 years. Also, because high-power IOLs magnify the myopic shift due to the growth of the eye, the authors would hesitate to implant single IOLs with powers >30 D.

Table 7.4 MANAGEMENT OF UNILATERAL CATARACTS

Age at Surgery (yr)	Management	Initial Refractive Goal If IOL Is Implanted (D)	Likely Refraction At Age 20 yr (D)
<0.5	Aphakia	—	+11
0.5 to 1	Aphakia or pseudophakia	+9 to +6	-3.4 to -4.9
1	IOL	+5	-4.9
2	IOL	+4	-3.4
4	IOL	+2.5	-2.7
8	IOL	+1.25	-1.5

Note, these goals are for illustration only; we do not have science to suggest that one goal is better than another. “Likely refraction at age 20 yrs” is the mean based on the RRG2 model of refractive growth; many children will have a greater or lesser refractive error at this age.

We do not recommend using tables exclusively to guide IOL power choice. There are many additional factors to consider in each situation, such as family circumstances, ability to follow up, and possibly using other approaches such as temporary polypseudophakia or planning for implantation of a secondary IOL in aphakic eyes. In addition, the rapidly advancing field of refractive surgery will allow correction for many of the children with adverse ultimate refractive outcomes. The surgeon who implants IOLs in young children should be aware of these factors and account for them in his or her planning.

Table 7.3 RECOMMENDED POSTOPERATIVE REFRACTIVE GOALS FOR CHILDREN AGED 1 TO 8 YEARS, FROM VARIOUS AUTHORS

Age at Surgery (yr)	Postoperative Goal (D)		
	Enyedi et al. (1998) ³⁴	Plager et al. (2002) ³⁵	Trivedi and Wilson (2009) ³⁶
1	+6		+6
2	+5		+5
3	+4	+5	
4	+3	+4	+4
5	+2	+3	+3
6	+1	+2.25	+2
7	0	+1.5	+1.5
8	-1 to -2	+1	+1
10		+0.5	+0.5
>14			0

Table 7.5 MANAGEMENT OF BILATERAL CATARACTS

Age at Surgery (yr)	Management	Initial Refractive Goal If IOL Is Implanted (D)	Likely Refraction At Age 20 yr (D)
<0.5	Aphakia	—	+11
0.5 to 1	Aphakia	—	+11
1	Aphakia or pseudophakia	+7	-3.0
2	IOL	+5	-2.5
4	IOL	+3.5	-1.7
8	IOL	+2	-0.8

Note, these goals are for illustration only; we do not have science to suggest that one goal is better than another. The *lifelong* safety of bilateral pseudophakia after cataract surgery in childhood is not yet determined; bilateral aphakia is generally well tolerated. “Likely refraction at age 20 yr” is the mean based on the RRG2 model of refractive growth; many children will have a greater or lesser refractive error at this age.

DISCLAIMER

The views expressed in this article are those of the authors and do not necessarily reflect the official policy or position of the Department of the Navy, Department of Defense, or the U.S. Government.

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8

Planning Pediatric Cataract Surgery: Diverse Issues

Rupal H. Trivedi and M. Edward Wilson

Children are not miniature adults; they have unique anatomy, physiology, psychology, and social status. Not only are children's eyes smaller than adults', but also their tissues are much softer. A cataract in an adult reduces visual acuity, while in a child it may also interfere with normal visual (brain) development. In a young child, a cataract blurs the image received by the retina and disrupts the development of the visual pathways in the central nervous system. The *timing of surgery, the surgical technique, the choice of aphakic correction, and the amblyopia management* are of utmost importance in achieving good and long-lasting results in children.

The management of pediatric cataracts is far more complex than the management of cataracts in adults. Differences and difficulties encountered during the preoperative, intraoperative, and postoperative periods are listed in Table 8.1. In this chapter, we cover some of the diverse issues that are not covered as individual chapters in the book.

WHO SHOULD PERFORM PEDIATRIC CATARACT SURGERY: PEDIATRIC OPHTHALMOLOGISTS OR ADULT CATARACT SURGEONS?

The incidence of pediatric cataracts is not high enough to allow many surgeons to devote their entire careers to *pediatric cataract surgery*. The result of our worldwide survey revealed that more than 71.5% of American Society of Cataract and Refractive Surgeon respondents performed <10 pediatric cataract surgeries per year, while the majority of American Association of Pediatric Ophthalmologist and Strabismus (AAPOS) respondents (85%) indicated <20 pediatric cataract surgeries performed per year.¹ In most cases, either pediatric ophthalmologists or cataract surgeons (primarily performing adult cataract surgery) cultivate the interest in pediatric cataract surgery. In 2000, Wood and Ogawa² wrote, "Given the overall paucity

of clinical experience in pediatric patients and its hazardous nature, who should be performing pediatric cataract surgery in the first place? Is this the realm of the pediatric ophthalmologist, the adult cataract surgeon, or perhaps both?"

The question of who is best suited to perform pediatric cataract surgery is not easily answered. Is it best performed by pediatric ophthalmologists, who deal with children exclusively, or cataract surgeons, who frequently perform adult cataract surgery?³ We raised this question in our survey and found that 77.4% of ophthalmologists not performing pediatric cataract surgery referred patients to a pediatric ophthalmologist.¹ Among ophthalmologists performing pediatric cataract surgery, more than half (52.6%) stated that either a pediatric ophthalmologist or an adult cataract surgeon should perform this procedure.¹

This issue may depend on "local" situations and there is no "must" here. The ophthalmic surgeon with the most experience and interest in pediatric cataract surgery should be sent the surgical cases from the locale.⁴ Co-management among ophthalmologists works well in this setting. In the United States, pediatric ophthalmologists are more likely to be the most experienced, however, outside the United States, adult cataract surgeons generally lead the field. Pediatric ophthalmologists are much more aware of the anatomy and functional parameters of pediatric eyes. They have much to teach adult cataract surgeons about operating on the infant eye and various functional issues in postoperative management. Conversely, cataract surgeons (primarily performing adult cataract surgery) are much more experienced in surgical technique such as capsulorhexis and innovations in intraocular lens (IOL)-related technology, and they also have much to teach pediatric ophthalmologists about adult surgical advances that should be applied to children.⁴

In our opinion, the surgeon must have enough experience to feel comfortable with the specific difficulties of the pediatric eye. Children with cataracts often have other associated health problems that increase anesthesia risks.

Table 8.1 HOW DOES PEDIATRIC CATARACT SURGERY DIFFER FROM ADULT CATARACT SURGERY?**Preoperative Period**

1. Difficult and often delayed diagnosis
2. Timing of surgery: In sharp contrast to the treatment of adult cataracts, the timing of cataract surgery in children is of paramount importance. It affects the visual result to a much greater extent than does the surgical technique or method of postoperative optical correction used by the surgeon.
3. High incidence of associated ocular and systemic anomalies and prematurity
4. Setup for pediatric general anesthesia a prerequisite
5. Apprehension about general anesthesia
6. Examination under anesthesia: necessary sometimes even to diagnose cataract and for preoperative assessment
7. Need for automated keratometer and A-scan in operating room
8. Difficulty in calculating IOL power
9. Psychologic issues and preoperative counseling of parents

Intraoperative Period

1. Risks of general anesthesia
2. Smaller size of the eye
3. Poor dilation of pupil more often associated with pediatric eyes
4. Low scleral rigidity
5. Relative size of the pars plana: The pars plana region in the infant eye is incompletely developed, so the anterior retina lies just behind the pars plicata.
6. Incision and suturing: As opposed to adult eyes, a superior tunnel is preferable in pediatric eyes (as it provides better protection and, in general, children do not have deep-seated eyes, which would require temporal incision). It is preferable to suture even a “self-seal” tunnel incision in children as opposed to adults.
7. Need for high-viscosity viscoelastic for capsular management
8. Difficulty in performing an anterior capsulorhexis associated with a highly elastic anterior capsule and increased intralenticular and intravitreal pressure
9. Densely formed vitreous and scleral collapse contributing to vitreous upthrust giving rise to raised intravitreal and lenticular pressure, making anterior and posterior capsular management difficult
10. Removal of lens substance rarely requires phacoemulsification, but the cortex is stickier and gummier than in adults
11. Posterior capsule plaques are common and may require intraocular scissors in addition to the vitrector
12. Need for primary posterior capsule management to prevent dense and thick PCO in those too young to apply an awake YAG laser
13. Need for vitrectomy instrumentation—preference for Venturi pump, which requires a nitrogen tank or wall hook up
14. Difficult IOL implantation

Postoperative Period

1. Higher risk for opacification of the visual axis
2. Propensity for increased postoperative inflammation
3. Compliance with the use of topical postoperative medications difficult
4. Requirement for frequent correction of residual refractive error, as it is constantly changing due to growth of the eye
5. Difficulty in documenting anatomic, refractive, and visual acuity changes due to poor compliance—also, young children do not know their letters, so alternative testing methods may be needed
6. Examination possibly requiring repeated brief anesthesia due to lack of cooperation with office exam
7. Tendency to develop amblyopia and need for patching
8. Long-term follow-up important but not always easily achieved

They require close monitoring pre- and postoperatively by staff with expertise in pediatric anesthesia and recovery. It is mandatory to have appropriate backup in case of intraoperative or postoperative anesthetic complications, especially since childhood cataracts can be associated with a wide range of systemic and metabolic abnormalities. These cases are best handled at a pediatric care center or an operating room facility offering experienced pediatric anesthesiologists. Whoever performs pediatric cataract surgery must understand the importance of *teamwork*. A pediatric ophthalmologist should generally be following these eyes during the postoperative course for strabismus, amblyopia, and other functionally related

issues. However, for each region a solution has to be individually tailored based on the resources available and the willingness of pediatric and adult cataract surgeons to continue to learn from each other and to work together to provide good care.

DOES CONSERVATIVE MANAGEMENT HAVE A PLACE IN PEDIATRIC CATARACT SURGERY?

Poor anatomical and functional outcomes of cataract surgery in children have prompted many surgeons to try conservative treatment such as the use of mydriatic drops.⁵⁻⁸

Chandler^{5,6} noted in the 24th Edward Jackson Memorial Lecture that “the fact that so many eyes are lost after surgery for congenital cataract is the reason why operation is not advised unless the vision is quite low.” DeVoe⁸ stated that “it is better to have 20/50 vision with accommodation than 20/20 vision without accommodation.” A conservative approach is most commonly indicated for a unilateral partial cataract. However, many conservative surgeons have also stated that bilateral partial cataracts should not be extracted if the visual acuity is better than 20/50 to 20/70.^{5,6} But it is not possible to measure visual acuity in infants with these partial cataracts. Difficulties in documenting visual acuity in infants with incomplete, but visually significant, cataracts may unnecessarily defer surgery in these patients, leading to irreversible damage because of amblyopia. In earlier years, it was believed that no surgery should be considered if the fundus can be viewed around that cataract, and therefore, conservative treatment should be used in eyes with a central opacity but relative peripheral sparing⁷ (Figs. 8.1 and 8.2). When treating eyes with a conservative approach, it is important to prescribe appropriate amblyopia therapy. Faye noted the choice of mydriasis for conservative management.⁹ The weakest mydriatics should be used at first, for example, 0.5% or 1% Tropicamide once or twice daily, increasing the dosage as needed. When the iris becomes refractory to one drug, one drop of Homatropine 2%, cyclopentolate 0.5% or 1%, or scopolamine 0.25% can be added as reinforcement or a substitute in case of allergy. The angle and intraocular pressure should be checked frequently at first, and then at least once a year. Use of mydriatic drops necessitates the patient’s wearing glasses for reading if any cycloplegic effect is induced. This has not found widespread acceptance. Associated glare and

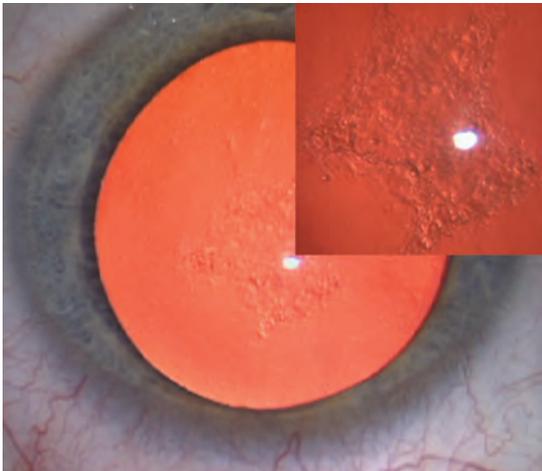


Figure 8.1. Posterior subcapsular cataract in an 11-year-old girl who received a bone marrow transplantation for multiple histiocytic cysts. Vision is reduced only to 20/40, but glare is a problem in some lighting conditions. Deciding between surgery and observation can be difficult in cases like this.

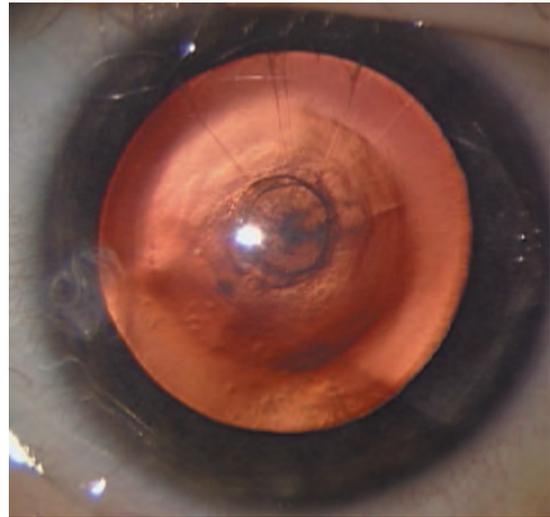


Figure 8.2. Central unilateral lens opacity in a 7-week-old child. Should surgery be performed without delay or is a conservative approach indicated? These decisions can be very difficult. This opacity worsened slowly over time and was eventually operated. Patching and observation worked well initially, however.

loss of accommodation are the most common obstacles. Visual outcomes have also been unimpressive. Despite these limitations, the use of mydriatics drops can be kept in reserve in eyes with slowly progressive cataracts (e.g., steroid-induced cataracts) or central cataracts and, especially, in patients for whom cataract surgery needs to be deferred for any reason—be it medical (high risk for anesthesia), social, or economic.

ANTERIOR POLAR CATARACT: HOW TO MANAGE?

Congenital anterior polar cataracts are small anterior axial lens opacities (Fig. 8.3A–D), often bilateral and symmetric in the two eyes. Because of their anterior location, they are often noted soon after birth by parents or pediatricians. They are generally considered to have little or no visual significance. They occasionally project forward into the anterior chamber, forming a pyramidal cataract. They usually remain stationary and traditionally have been considered to have little or no effect on vision. In 1984, Jaafar and Robb reviewed the cases of 63 patients with congenital anterior polar cataracts and found an unexpectedly high incidence of amblyopia and strabismus.¹⁰ In 1985, Nelson and colleagues reported five patients with anterior polar cataract. The authors noted that all five children ultimately required surgery.¹¹ In two children, the progressive opacification occurred in the posterior portion of the lens and was inapparent to the parents. The authors recommended a regular and careful follow-up of all children with anterior polar cataract.

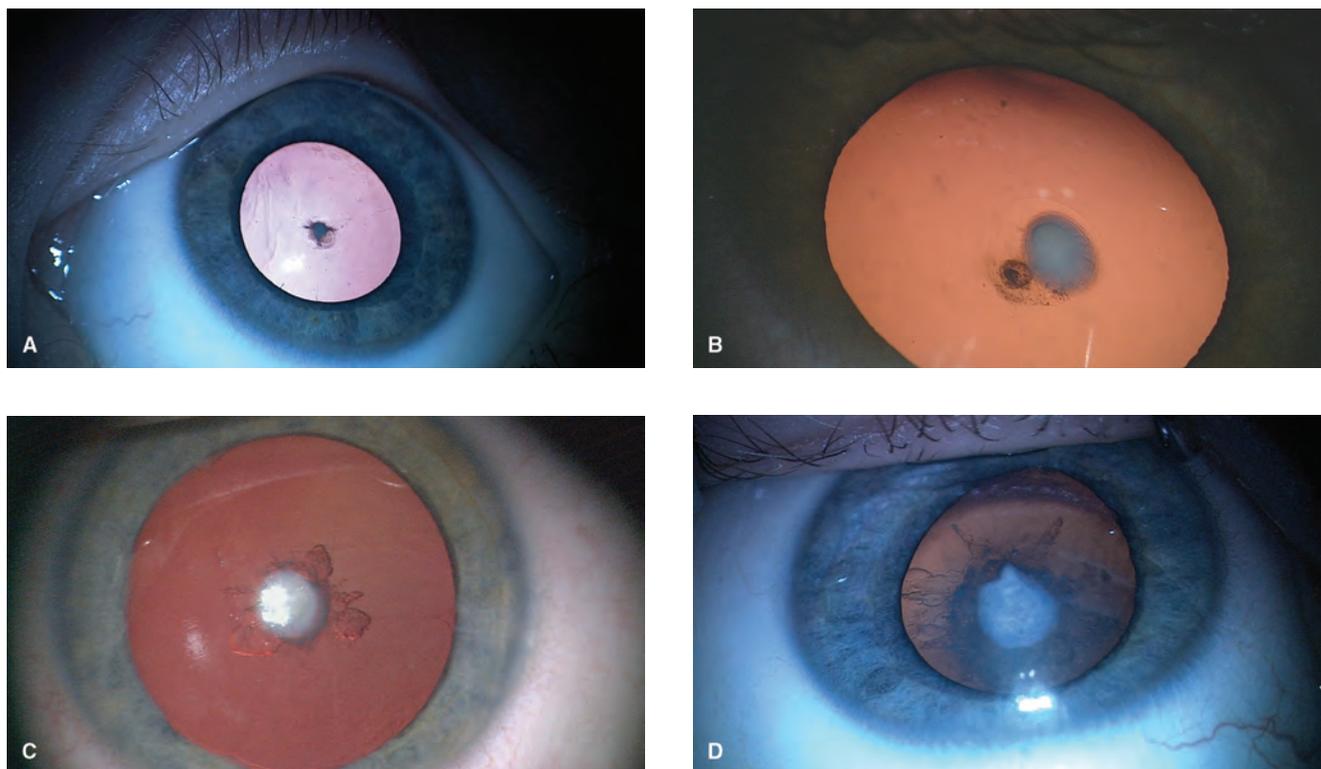


Figure 8.3 A-D: Anterior polar cataract.

In 1999, Wheeler and colleagues reported the clinical features and management of children with pyramidal anterior polar cataract.¹² Nine children had bilateral and six had unilateral pyramidal cataracts (24 eyes). Twenty of twenty-four eyes developed cortical opacification that extended significantly beyond the base of the pyramidal lesion. Nineteen eyes required cataract surgery. Amblyopia was present or developed in all six patients with unilateral cataract and in eight of nine patients with bilateral cataract. The incidence of amblyopia in this series was relatively higher than other reviews of anterior polar cataracts, perhaps because this series evaluated only the pyramidal cataract subset. Pyramidal cataract may represent a more severe variant of anterior polar cataract and therefore is associated with a higher incidence of amblyopia and cataract progression into the lens cortex. Amblyopia is related to the size of the opacity, the asymmetry of opacities, and the superimposition of cortical changes (astigmatism is commonly associated with these cataracts). Visual acuity in many eyes remained poor despite amblyopia therapy. The pyramidal opacities consisted of hyperplastic lens epithelium, which exhibited a loss of polarity and was surrounded by a collagenous matrix. Whenever possible, conservative treatment involving refractive correction, pupillary dilation, and occlusion therapy for amblyopia was used. If the visual axis was occluded by a lens opacity of 3 mm or more in diameter

or significant refractory amblyopia was present, or in case of both, cataract surgery was performed.

In 2005, Ceyhan and colleagues reported risk factors for amblyopia in congenital anterior lens opacity.¹³ The authors sought to examine several variables. These included cataract subtype (polar, subcapsular, or pyramidal), location (central, paracentral, peripheral), diameter, presence of adherent iris tissue, anisometropia (spherical equivalent), and unilateral versus bilateral. Amblyopia was present in 17 of 59 patients (28.8%). Patients with amblyopia had a mean anisometropia of 1.23 diopters. The only variable associated with increased relative risk for amblyopia in this group of patients was anisometropia. Patients with congenital anterior lens opacity who have anisometropia of 1 diopter or greater are 6.5 times more likely to develop amblyopia. Note that only one patient in this series had a pyramidal cataract.

In summary, close follow-up is essential in patients with anterior polar cataract. We advise follow-up every 3 months within 1st year of life and to gradually decrease it to yearly follow-up. Patients with unilateral cataract and progressive lesion require closer follow-up. Astigmatism and anisometropia need to be suspected even when the cataracts are very small. The pyramidal cataracts spread into the underlying cortex more often and can become visually significant over time, requiring surgical removal.

SIMULTANEOUS INTRAOCULAR AND STRABISMUS SURGERY

Simultaneous intraocular and strabismus surgery may be considered by the surgeon or requested by the parents. Combining cataract extraction with strabismus surgery is done uncommonly in children. After removal of a visually disabling cataract or other media opacity, sensory strabismus, particularly exotropia, may improve, either spontaneously or with nonsurgical therapy. Therefore, in certain patients with cataract and strabismus, concurrent eye muscle surgery may be unnecessary and could lead to an overcorrection. In other patients, the strabismus is stable, constant, and longstanding. In these patients, combined surgery can be considered. It is best to perform the strabismus procedure on the opposite eye from where the cataract is being removed. However, a sensory strabismus may be chronic and associated with contracted muscles (such as a tight lateral rectus in a chronic exotropia). In these settings we have performed both surgeries on the same eye. Since this may create a slightly greater risk of endophthalmitis, we often defer the strabismus procedure until there has been full healing from the cataract procedure.

Combining other lens procedures (e.g., posterior capsule opacification [PCO] removal or secondary IOL implantation) with a strabismus procedure has been discussed more frequently. Combining these two surgeries reduces the number of surgical and anesthetic procedures, speeds rehabilitation, and offers financial benefit to the patient and third-party payors.¹⁴ However, concern may still arise regarding a potential increased risk of infection, anterior segment ischemia, or excessive discomfort to the patient. Determining the ocular alignment in eyes with poor vision may be difficult. After the eye is visually rehabilitated, a more accurate measurement may be possible. Eye muscle surgery can alter refraction and corneal topography, thus there is the potential for an undesirable postoperative refraction when combining strabismus surgery and IOL implantation in the same eye.

In eyes undergoing incisional intraocular surgery combined with strabismus surgery under the same anesthesia, the muscle procedure generally precedes the lens surgery to avoid disrupting or opening the corneal incision during extraocular muscle manipulation. For the patients undergoing combined YAG laser posterior capsulotomy and strabismus surgery, the laser procedure was performed first. Ticho and colleagues¹⁴ reported outcome of 12 eyes of 11 children undergoing combined strabismus and lens surgery. Five of these children underwent combined strabismus and unilateral cataract surgery with lens implantation (one underwent subsequent contralateral lens and combined strabismus surgery), two underwent combined strabismus and unilateral cataract extraction (without lens implantation), two underwent combined strabismus

surgery with unilateral secondary IOL implantation, and two underwent combined strabismus surgery with unilateral YAG laser posterior capsulotomy, for a total of 12 cases. A limbal strabismus incision was used for two patients, whereas a fornix approach was used for all other muscle procedures. In 1986, Maltzman et al.¹⁵ reported the results of simultaneous exotropia and extracapsular cataract surgery (four patients) or secondary IOL placement (six patients). A unilateral recession resection procedure was performed on all patients. Surgical amounts were not specified. With an average follow-up of 15 months, they reported excellent postoperative alignment in all patients: one patient with a distance residual exodeviation of 16 prism diopters (PD), and all other patients with distance deviations of 12 PD or less. Zwaan and al-Sadhan¹⁶ in 1998 reported 10 patients with simultaneous cataract and strabismus surgery, and 4 additional patients with strabismus correction combined with glaucoma, keratoplasty, pupilloplasty, or YAG capsulotomy procedures. Eleven patients had recession–resection procedures ipsilateral to the intraocular surgery, and two patients had bilateral strabismus surgery. Orthophoria was achieved in 11 patients at 2 months postoperatively. Two patients with esotropia and one patient with exotropia were overcorrected by more than 10 PD. An adjustable suture was used in only one patient in this series (on the eye contralateral to cataract surgery). We avoided using an adjustable suture during combined surgery on an eye undergoing incisional intraocular surgery out of concern that pulling on the muscle during the adjustment could disturb the cataract incision or IOL position. We perform eye muscle surgery first if the same eye is to have incisional lens surgery, again to avoid traction on the globe incision or IOL.

PHAKIC IOL IMPLANTATION AND CLEAR LENS EXTRACTION

Corneal refractive surgery is less invasive than intraocular surgery; however, it is not suitable for corrective extreme refractive errors. In eyes with these errors, phakic IOL implantation or refractive lens exchange with IOL implantation can be useful alternatives. Although intraocular surgery is a viable alternative when treating such eyes, care should be taken to document the best corrected visual acuity and discuss the options with the patient or parents. In the rare case of a severe complication (e.g., endophthalmitis) it may be difficult to justify the indication for clear lens extraction. Documenting the decision-making process is very important in these cases. Another challenging aspect of refractive lens exchange in an individual is maintenance of clear visual axis. PCO is one of the most challenging aspect of lens surgery in these patients. We have reported a patient with Angelman syndrome who underwent refractive lens exchange with IOL implantation at 22 years of age.¹⁷

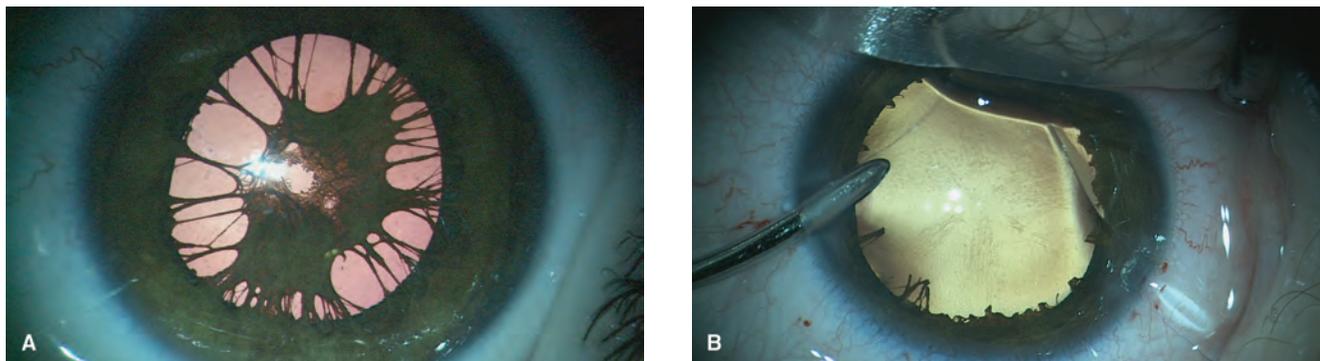


Figure 8.4 A, B: Persistent pupillary membrane (PPM) in a 2-year-old child. PPM was present in both eyes. **A:** Preoperative view. **B:** After removal of pupillary membrane.

PERSISTENT PUPILLARY MEMBRANE

Persistent pupillary membrane (PPM) is a congenital, incomplete, involution of the tunica vasculosa lentis (Fig. 8.4A).¹⁸ This membrane is usually tenacious, with fine fibrils attached to the iris collarette on one side, with the other end either free floating or attached to the lens or iris focally on the opposite side. This condition is thought to represent ectopic iris tissue on the lens with abnormal iris stroma caused by aberrant involutinal changes in the primitive embryologic vascular system of anterior segment. PPM was observed in 5% of children with Aicardi syndrome.¹⁹

Visually asymptomatic PPM can be conservatively managed. Amblyopia treatment is very important with the patient treated by conservative approach. Dense and thick membrane, particularly within the pupil, may result in deprivation amblyopia. Symptomatic cases can be treated with a laser or surgery. Lens-preserving excision of congenital hyperplastic pupillary membrane has been reported¹⁸ (Fig. 8.4B). Secondary cataract may occur in children treated by this approach. PPM is occasionally associated with cataract. Such cases can either be treated by surgery alone or combined sequential argon-YAG laser membranectomy and surgical removal of cataract.²⁰

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9

Immediately Sequential Bilateral Pediatric Cataract Surgery

Rupal H. Trivedi and M. Edward Wilson

Surgery on both eyes during one trip to the operating room has been performed on muscles, on lids, and in refractive laser surgery. However, surgical management of bilateral intraocular pathology (e.g., cataract, glaucoma) has generally been performed during separate sessions at different intervals depending on surgeon's preference, patient's/parent's preference, and the outcome of the first procedure. The terminology used to describe the practice of bilateral cataract surgery done on the same day has been confusing. It has been referred to as same-day sequential, immediately consecutive, simultaneous bilateral, or immediately sequential bilateral surgery.¹ "Immediately sequential bilateral cataract surgery" (ISBCS) is the currently preferred title, contrasting it with "delayed sequential bilateral cataract surgeries" (DSBCS), where the two procedures are performed as totally separate operations on different days.²

By any name, bilateral intraocular surgery not performed as totally separate operations on different days remains controversial. The risks of bilateral infection or toxic anterior segment syndrome (TASS) are weighed against the benefits of surgery under a single general anesthesia (GA). The latter reduces the travel time for the family and eliminates the risk of amblyopia due to delay between surgery on the first eye and the second eye. Cost considerations may favor the immediate sequential approach, but as Kushner³ warned in his 2010 editorial, allowing those in charge of health care dollars to influence practice patterns may introduce devastating potential risks. As with many debates in health care, the best management of risk and resources requires an assessment of the surgical setting and the health of the patient.

The use of ISBCS has been reported at least as far back as the early 1950s.^{4,5} An international society of bilateral cataract surgeons has now been founded (www.isbcs.org). However, ISBCS remains a hotly debated topic where almost every discussion either starts or ends with a comment on the disagreement surrounding its

use. Peer-reviewed publications on ISBCS often have an accompanying editorial or letter to the editor reflecting the controversial nature of the subject.^{3,6-21}

The historical reluctance to perform ISBCS stems, in major part, from the potentially devastating consequences of a severe complication when it occurs bilaterally and the difficulty we have in quantitating how often such a rare complication would affect both eyes simultaneously if both eyes were put at risk at the same time. Can the benefits of ISBCS justify even the smallest risk of bilateral blindness? *The important question here is not "Can it be done?" but, more properly, "Should it be done?"* Although ISBCS remains controversial, for children with health problems, the risks of GA should be weighed against the risks of ISBCS. Many surgeons agree that there is a place for the ISBCS approach in patients for whom GA poses more than average risks.²²⁻²⁵ Several authors have proposed that ISBCS is a feasible and recommendable approach in selected pediatric cataract patients.^{22-24,26,27} We have used this approach in about 5% of infants operated for bilateral visually significant cataract, mainly because of higher risk of anesthesia, however, we have also used it (rarely) when we knew that the family would have difficulty getting back to us for the surgery on the second eye.

PROS

Medical

It may be logical to believe that performing ISBCS in the very young reduces the risk to the patient incurred from GA by exposing them to only one extended anesthesia episode rather than two separate episodes. Although we are not aware of a published study comparing the risk of more than one GA with that of prolonged anesthesia time, it is commonly accepted that induction, tracheal intubation, and emergence are high-risk periods of anesthesia.²⁸ However, a longer procedure may also tend to increase the risk of anesthetic mortality in infants, and one should

weigh the risk of 1 longer operation versus 2 shorter ones.³ Anesthetic agents could adversely affect neurologic, cognitive, and social development of neonates and young children. One cohort study found an association between the development of reading, written language, and mathematics learning disabilities and two or more anesthesia episodes as well as longer cumulative duration of anesthesia in children younger than the age of 4 years.²⁹ Later, Rappaport et al.³⁰ noted that there is not enough information to draw any firm conclusions regarding an association between anesthetic exposure and subsequent learning disabilities. Nallasamy et al.²⁸ recommended ISBCS in infants with bilateral congenital cataracts who are rated with an American Society of Anesthesiologists physical status >2 or generally those who are at higher risk for anesthesia-related complications. The systemic diseases associated with congenital/infantile cataracts in which anesthetic difficulties are higher than average are described by Zwaan²⁴: rubella syndrome (congenital heart defects), Lowe syndrome (hypocalcemia, acidosis, renal failure), homocystinuria (thromboembolic episodes), prematurity (respiratory problems), Marfan syndrome (cardiovascular and respiratory problems), and craniosynostoses (difficult intubation, increased intracranial pressure, associated heart defects, respiratory problems), among others. Although Marfan syndrome and homocystinuria are included in the above list, eyes with subluxated lens are complicated to operate, and most physicians would avoid performing ISBCS for such cases. Also, for each child, the risk of anesthesia is best determined in consultation with an experienced pediatric anesthesiologist and with the physicians caring week to week for the child.

Social

Social benefits of ISBCS include less anxiety for parents because their infant will undergo GA only once. This saves time for the caregiver who may have to travel far distances to the hospital and may have to arrange for the care of other children in the family. One of the relative indications of ISBCS is when follow-up care and travel are significant hardships for the parents. Sarikkola et al.³¹ reported ISBCS in adults was positively received by patients. It improved patient satisfaction, and 91% of patients who underwent ISBCS reported that they would recommend it to relatives or friends. Chung et al.³² also noted a high degree of patient satisfaction in a prospective controlled trial. These adult studies may lead us to think that ISBCS could produce higher satisfaction for the parents of our pediatric patients, too.

Ocular

It has also been argued that it may be advantageous to remove bilateral cataracts in one session to minimize the risk of monocular stimulus deprivation amblyopia.²⁸ However, as long as surgery for the second eye is performed in a timely manner, the impact of this factor appears to be

minimal. Often surgery on the second eye in young children is planned 1 week or less after the first eye. However, when children get upper respiratory infections or other minor illnesses, and they often do, the second anesthesia may need to be postponed and the planned short interval between surgeries for the two eyes can become extended and the risks of amblyopia increased.

Economical

O'Brien et al.² noted that average hospital costs were significantly reduced when performing ISBCS (32% reduction) in adults. Dave et al.³³ reported ISBCS is associated with a 22% reduction in medical payments for congenital cataract surgery. They analyzed medical payments to their hospital as a surrogate for health care costs in general. As is discussed below, physicians working in a fee-for-service model may see these reduced payments as a disincentive.

Developing World

ISBCS may be a reasonable option for pediatric cataract surgery in many developing world settings where the availability of GA is limited. Travel and housing logistics may make it difficult to arrange for two separate operations on 2 separate days for each bilaterally blind child who is in the amblyopic years of life. In these settings, ISBCS can help to manage the backlog of cataract blindness.¹⁴ In many parts of the world, it is not unusual for patients from remote and rural areas to have surgery in one eye, regain good vision, and get lost to follow-up. Such a scenario can be prevented by ISBCS. As early as 1956, in Delhi, India, ISBCS was used as an eye camp in Darbhanga (Patna), in approximately 30% of patients.²² Dr. Nagpal (Ahmedabad, India) has stated that he would consider ISBCS in eye camps, when operating adult patients with bilateral hypermature cataract.²² To avoid the risks and costs of a second anesthesia and to make maximal use of the balanced salt solution bottles and vitrector tubing, ISBCS is being utilized or is being given consideration in many developing world pediatric surgery centers.³⁴

CONS

Complications

The main cause of concern in patients undergoing ISBCS is the possibility of visual impairment due to serious complications affecting both eyes. *Fear of endophthalmitis occurring in both eyes, leading to blindness, is the most important factor deterring ophthalmologists from performing ISBCS.*²³ Uniocular¹⁹ as well as bilateral³⁵⁻³⁸ endophthalmitis after bilateral surgery has been reported. Arshinoff³⁹ remarked on 10 cases of unilateral endophthalmitis in 33,000 ISBCS cases. There have been four reported cases of bilateral endophthalmitis following ISBCS, but only one of these occurred in non-at-risk patients or where

adequate precautions had been taken.^{38,39} Small sample size and infrequency of this complication prevent us from drawing any meaningful conclusion regarding its absence.

An increased risk of endophthalmitis in adult cases was reported when an anterior vitrectomy is performed.^{40,41} These data, however, were collected in adults with inadvertent rupture of the posterior capsule, leading to a longer surgery time and often other complications. Whether pediatric posterior capsulectomy and anterior vitrectomy increase the risk of endophthalmitis is not known.

Although potential problem that is most frequently highlighted in the literature is the risk of bilateral endophthalmitis, other complications are also rarely reported (bilateral expulsive hemorrhage,⁵ visual impairment due to bilateral corneal endothelia failure,⁷ etc.). Olson¹² noted that bilateral TASS is more likely than that of bilateral endophthalmitis.

Legal Implications

Complications do occur, and although having a serious complication in one eye is undesirable, having the complications in both eyes is a disaster.²⁰ If complications arise during ISBCS that result in complete blindness, the potential settlement would likely be much higher than for loss of the use of one eye alone. This could lead to increased malpractice costs for all cataract surgeons.²⁰

Economical Considerations

ISBCS is associated with decreased reimbursement (compared to DSBCS) in many countries, including the United States.¹² Arshinoff and Chen⁴² showed that discounting second-eye cataract surgery in ISBCS was a financial deterrent. Although increased efficiency was a slight incentive to ophthalmologists and surgical centers, anesthesiologists experienced significant financial disincentives. The United Kingdom was the only jurisdiction they evaluated where the increased efficiency of ISBCS offset the second-eye payment reduction and may actually encourage ISBCS.

PREPARATION AND PRECAUTION WHEN USING ISBCS

Patient selection for ISBCS: Eyes with a higher risk of infection, for example, nasolacrimal duct obstruction, iodine allergy, etc., present a relative contraindication to ISBCS, as do eyes with associated ocular conditions that may complicate the surgery, for example, subluxation, uveitis, etc. Before proceeding with ISBCS, one should consider the questions listed in Table 9.1. Some reports in the adult literature have quoted a higher incidence of *endophthalmitis* in secondary intraocular lens (IOL) patients compared to primary cataract surgery with IOL placement.⁴³ With that said, immediately sequential

Table 9.1

QUESTIONS THAT NEED TO BE ANSWERED BEFORE PROCEEDING WITH IMMEDIATELY SEQUENTIAL BILATERAL CATARACT SURGERY

- Do both eyes have a visually significant cataract?
- What is the risk of endophthalmitis?
- What do we achieve by doing immediately sequential surgery?
- What are the risks of anesthesia, in particular, those of two separate inductions of GA within a period of a few days or weeks, compared with lengthening the duration of one anesthesia?
- What are the chances that the patient will not come for follow-up?
- Has there been a careful checkup for evidence of conjunctivitis, nasolacrimal duct obstruction, or upper respiratory tract infection?
- Is there a history of intake of immunosuppressive agents?
- Have the parents or caregiver received appropriate information and have they provided informed consent?

surgery may not be recommended for secondary IOL implantation. We cared for an older child who underwent DSBCS for bilateral secondary IOL implantation and developed unilateral endophthalmitis after swimming in canal water on the second post-operative day after his second eye was operated (and 1 month after his first eye had been operated).⁴⁴ Had this patient undergone ISBCS, his risk-laden behavior may have resulted in bilateral blindness.

Informed consent and documentations: If ISBCS is considered as an option, the parents/caregivers should be carefully informed of the potential advantages and disadvantages. Parent should be free to choose ISBCS or DSBCS. The ophthalmologists considering ISBCS should carefully obtain informed consent regarding all issues of potential patient concern. The ophthalmologist should be prepared to justify a decision to perform bilateral cataract surgery. Written documentation is important.

Surgical preparation: Once the decision is made to move forward with this option, techniques should be chosen throughout to minimize the associated risk, especially infection (see Table 9.2).

PRACTICE GUIDELINES

American Academy of Ophthalmology

The 2011 American Academy of Ophthalmology (AAO) preferred practice pattern (PPP®) (<http://one.aao.org/CE/PracticeGuidelines/PPP.aspx>) stated that if ISBCS is being considered the patient must be carefully informed of potential disadvantages, and still did not support routine ISBCS. The AAO PPPs do not include relative

Table 9.2 PRECAUTIONS FOR SIMULTANEOUS BILATERAL CATARACT SURGERY

- Nothing in physical contact of first-eye surgery should be used for the second.²¹ Consider treating the second eye as a separate case (scrubbing in between the surgeries, using a separate instrument set, reprepping of the second eye with povidone–iodine, redraping of the second eye, and using separate intraocular medications and irrigating fluid) but recognize that doing so has not been proven to reduce the risk of bilateral endophthalmitis.
- The separate instrument trays for the two eyes should go through complete and separate sterilization cycles with indicators.²¹
- Efforts should be made to minimize right–left eye error.
- Different ophthalmic viscosurgical devices (OVDs), and different manufacturer lots of surgical supplies should be used, whenever reasonable and possible for the right and left eye.²¹ Arshinoff¹⁵ noted that as OVD carries the highest bioburden (and therefore risk of contamination), use of different companies' OVD for two eyes has been suggested when doing ISBCS.
- If complications occur with the first eye, careful consideration should be given before proceeding with surgery on the second eye. Even though the same complication is not likely to be repeated in the other eye, severe bilateral complications may be difficult to defend with patients, families, and litigators.¹²
- Instructions should be given on using separate drop bottles for each eye postoperatively and washing hands before instilling eye drops (separately for both eyes).
- Wounds should be well sealed and reinforced with sutures whenever appropriate as protection against postoperative trauma.
- Covering the eyes with protective shields that allow the child some vision through them will reduce the disorientation the child might have if he/she wakes up blind from having both eyes bandaged. If bilateral bandages are necessary, have the parents present in the recovery room to comfort the child immediately upon awakening. For cataract surgery with IOL implantation, we do patch them with clear shield for at least 4 h postoperatively so they can see their parents. Drops are started when patches come off. For cataract surgery without IOL implantation, we do give contact lens on table, and do not give any patch.
- Intracameral antibiotics are recommended.²¹

indications for ISBCS, although they do list indications reported in the literature: *need for general anesthesia in the presence of bilateral visually significant cataracts, rare occasions where travel for surgery and follow-up care is a significant hardship for the patient, and when the health of the patient may limit surgery to one surgical encounter.*

Royal College of Ophthalmologists

The 2010 Royal College of Ophthalmologists (RCO) Cataract Surgery Guidelines (www.rcophth.ac.uk/core/core_picker/download.asp?id=544) include indications for ISBCS. The guidelines do not contain an argument against ISBCS; they merely urge that strict aseptic precautions must be taken when choosing ISBCS.¹

SUMMARY

ISBCS for children should be a joint decision among the anesthetist, the ophthalmic surgeon, and the parents. The concept of ISBCS is still controversial. Although the number of surgeons who report performing this procedure has increased, most appear to oppose the concept except in exceptional circumstances. This technique should be reserved for confident, experienced surgeons who have studied their complication rates and are certain that complications occur extremely rarely in their hands. We consider ISBCS for children who are at a higher-than-normal risk for anesthesia-related complications. The ISBCS approach is also discussed when a visit for surgery on the second eye would be difficult. However, the decision to perform ISBCS is never taken lightly. The ophthalmologist should be prepared to justify a decision to perform ISBCS. It is critical to carefully weigh the risks and benefits of ISBCS for a patient. These decisions are made on a case-by-case basis, weighing the risks and benefits of each unique situation. It must be undertaken with great care, and performed under complete aseptic technique, with strict separation of right and left eye surgeries. In other words, the second eye should be treated like the eye of a different patient. Although most pediatric studies reported no complications in their series, data in the current literature on endophthalmitis after cataract surgery and on the risks of repetitive anesthesia are inadequate to weigh the risk of bilateral endophthalmitis against the reduced risk of one anesthetic versus two. Until such information becomes available, simultaneous removal of bilateral infantile cataracts should probably be reserved for selected cases where the anesthetic risk is higher than average and risk of endophthalmitis is not higher than average.

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10

Cost Analyses for the Treatment of Unilateral Childhood Cataracts

David Stager Jr and Joost Felius

Medical decision and policy making are increasingly driven by cost-effectiveness data. Ideally, for decisions on resource allocation, a comprehensive societal perspective is taken that includes all costs and benefits, regardless of who incurs the costs and who reaps the benefits.¹ However, calculation of the cost-effectiveness from the societal perspective is complicated and depends on numerous unknown variables. A third-party payer perspective is typically more constrained and thus more straightforward to calculate, and the results are useful for the evaluation of specific procedures or for comparing alternative treatment approaches. As a worked-out example of establishing a cost-utility framework for pediatric cataract care, we present a third-party payer perspective cost assessment for the treatment and care of congenital unilateral cataracts.

EXAMPLE

A spreadsheet (Microsoft® Excel, Redmond, WA) was constructed to calculate the costs associated with a basic scenario as well as selected complications. The basic scenario (Fig. 10.1) consisted of extraction of a congenital unilateral cataract with anterior vitrectomy during the first few months of life, followed by contact lens optical correction until placement of a secondary intraocular lens (IOL) at age 6 years and spectacle correction thereafter. The scenario also included a regimen of alternate-day patching of the fellow eye from the time of cataract removal until age 8 years, as well as follow-up care until age 12 years. Two comprehensive and two intermediate office examinations annually as well as one additional contact lens visit each year were included. Exams under anesthesia were assumed to take place annually during the first 4 years and once in the year of IOL placement. The corresponding direct costs were based on 2011 Current Procedural Terminology (CPT) codes and Medicare

payment information² for procedures and examinations (including anesthesia and facility fees; Table 10.1) and on costs and quantities of medications, optical aids, and patches, thus taking a third-party payer perspective.

In addition to the basic scenario, known sequelae and complications of cataract extraction in infants were included and weighed by published rates of incidence (Table 10.1). Lens re proliferation or secondary membranes were assumed to occur in 20% of cases.^{3,4} Management requires membrane removal in the operating room and one follow-up examination in the office (in addition to follow-up included in the basic scenario). Clinically significant strabismus was assumed to occur in 70% of cases.⁵ In an estimated 80% of those cases, management requires one extraocular muscle surgery, whereas in the remaining 20% of the strabismus cases a second extraocular muscle surgery is needed for ocular alignment. Again, one extra office examination following each surgical procedure was included. Glaucoma was assumed to occur in 15% of cases,⁶⁻¹² half of which are treated with drops (estimated one 5-mL bottle of generic timolol per month, at \$17 each) while the other half receives a glaucoma implant. We assumed that glaucoma occurs around age 6 years. All patients with glaucoma have a follow-up schedule of six examinations in the year of occurrence and three examinations in the following years. Retinal detachment was assumed to occur in 1% of cases, with management consisting of a scleral buckling procedure. Endophthalmitis after pediatric cataract surgery is rare and was modeled at 0.1% incidence,^{13,14} with usual treatment consisting of a vitrectomy and antibiotics. The management of retinal detachment and endophthalmitis also included one extra office examination following each surgical procedure.

This example yielded the following cost estimates. The costs associated with the basic scenario (\$20,091 per patient) and with the typical complications (\$2,485) are

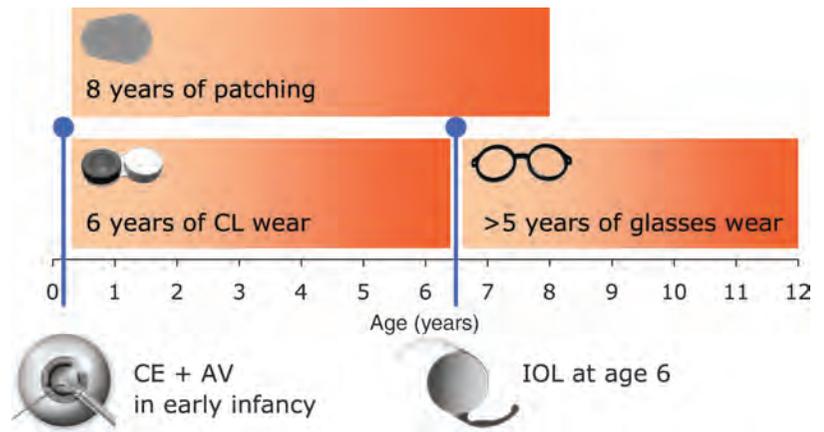


Figure 10.1. Schematic overview of the basic scenario, including lens removal (CE + AV) in the first year of life, placement of a secondary IOL at age 6 years, and follow-up until age 12 years. Bars indicate regimens of optical correction and patching.

Table 10.1 COST ELEMENTS FOR THE BASIC SCENARIO AND TYPICAL COMPLICATIONS

Cost Element	CPT Code	Reimbursement/ Cost per Unit (USD) ^a	Anesthesia (USD)	Facility Fees (USD) ^b	Number of Occurrences
Basic Scenario					
CE + AV	66982, 67010	1,589	253	837	1
Secondary IOL implantation	66985	759	211	962	1
Initial office exam	92004	146	—	—	1
Follow-up exam (intermediate)	92012	83	—	—	14
Follow-up exam (comprehensive)	92014	121	—	—	16
Exam under anesthesia	92018	140	84	1,031	5
Contact lens visit	92310	95	—	—	6
Patches	—	0.17	—	—	4,384
Contact lenses	—	116	—	—	18
Spectacles	—	384	—	—	6
IOL	—	150	—	—	1
Drops/postsurgery meds	—	50	—	—	2
Lens Reproliferation or Secondary Membrane					
Membrane removal	66830	687	211	736	0.2
Follow-up exam	92012	83	—	—	0.2
Drops/postsurgery meds	—	50	—	—	0.2
Strabismus					
First procedure	67312	721	190	736	0.7
Second procedure	67311, 67332	933	190	736	0.14
Follow-up exam	92012	83	—	—	0.84
Drops/postsurgery meds	—	50	—	—	0.84
Glaucoma					
Filter implant	66180	1,150	211	837	0.075
Glaucoma drops	—	17	—	—	6.3
Follow-up exam	92012	83	—	—	3.6
Drops/postsurgery meds	—	50	—	—	0.075

Table 10.1 COST ELEMENTS FOR THE BASIC SCENARIO AND TYPICAL COMPLICATIONS (Cont'd)

Cost Element	CPT Code	Reimbursement/ Cost per Unit (USD) ^a	Anesthesia (USD)	Facility Fees (USD) ^b	Number of Occurrences
Retinal Detachment					
Scleral buckle	67112	1,312	211	1,163	0.01
Follow-up exam	92012	83	—	—	0.01
Drops/postsurgery meds	—	50	—	—	0.01
Endophthalmitis					
Vitrectomy	67036	949	211	730	0.001
Follow-up exam	92012	83	—	—	0.001
Drops/postsurgery meds	—	164	—	—	0.001

CE + AV, cataract extraction with anterior vitrectomy; CPT, current procedural terminology; IOL, intraocular lens; USD, United States dollars.

^aListed are reimbursement numbers for examinations and procedures, and cost estimates for aids and supplies. Reimbursement figures reflect Medicare payment information.

American Medical Association. CPT/RVU Search. Available at <https://ocm.ama-assn.org/OCM/CPTRelativeValueSearch.do>. Accessed on January 20, 2012.

^bAmbulatory Surgery Center.

presented in Table 10.2. The total estimated costs in this model (sum of basic scenario and typical complications) are then \$12,824 per patient for operating room procedures (including surgical procedures and examination under anesthesia, facility fees, anesthesia), \$4,202 for office examinations (comprehensive and intermediate exams, contact lens checkups), \$5,287 for optical aids (spectacles, contact lenses) and patches, and \$263 for medications (glaucoma drops). These numbers combined yielded \$22,576 per patient for a point estimate of the total cost associated with this particular management approach of a congenital unilateral cataract.

In order to appreciate the relative influence of various elements in these calculations and to arrive as a plausible *range* of actual cost values, univariate sensitivity analysis was performed on the following parameters around the

base estimates. The complication rates were reduced to zero, thus reducing the model to the basic scenario (i.e., \$20,091 per case). Alternatively, complication rates were set to “high values” by doubling the incidences of lens reproliferation and glaucoma, assuming 100% incidence of strabismus, and a 10-fold increase of the rarer complications. Under these assumptions, the total estimated cost would increase to \$24,432 per patient. Similarly, varying the costs of optical aids and patches by $\pm 50\%$ yielded a variation between \$19,933 and \$25,220 for the total costs per patient. Both operating room costs and office exam costs were varied by $\pm 50\%$, resulting in range estimates of \$16,164 to \$28,988 and \$20,475 to \$24,677, respectively. Varying the timing of the secondary IOL between age 4 and age 10 years resulted in \$23,854 and \$21,435, respectively (Fig. 10.2).

COMMENT

Attitudes toward surgical treatment of congenital unilateral cataract have undergone drastic change over the last half century. While Costenbader and Albert in 1957 “would unequivocally advise against surgery in unilateral congenital cataracts,”¹⁵ the turning point came with promising results in the early 1980s.¹⁶ Nowadays, surgical removal of congenital unilateral cataract is standard practice in the United States, and it is generally accepted that treatment at an early age improves visual outcome.^{17,18} Results of a National Eye Institute-sponsored randomized clinical trial have been published comparing the safety and efficacy of the management of unilateral

Table 10.2 COST MODEL RESULTS IN US DOLLARS

Cost Element Category	Basic Scenario	Typical Complications	Total
Operating room procedures	10,890	1,934	12,824
Office exams	3,814	388	4,202
Optical aids and patches	5,287	0	5,287
Medications	100	163	263
Total	20,091	2,485	22,576

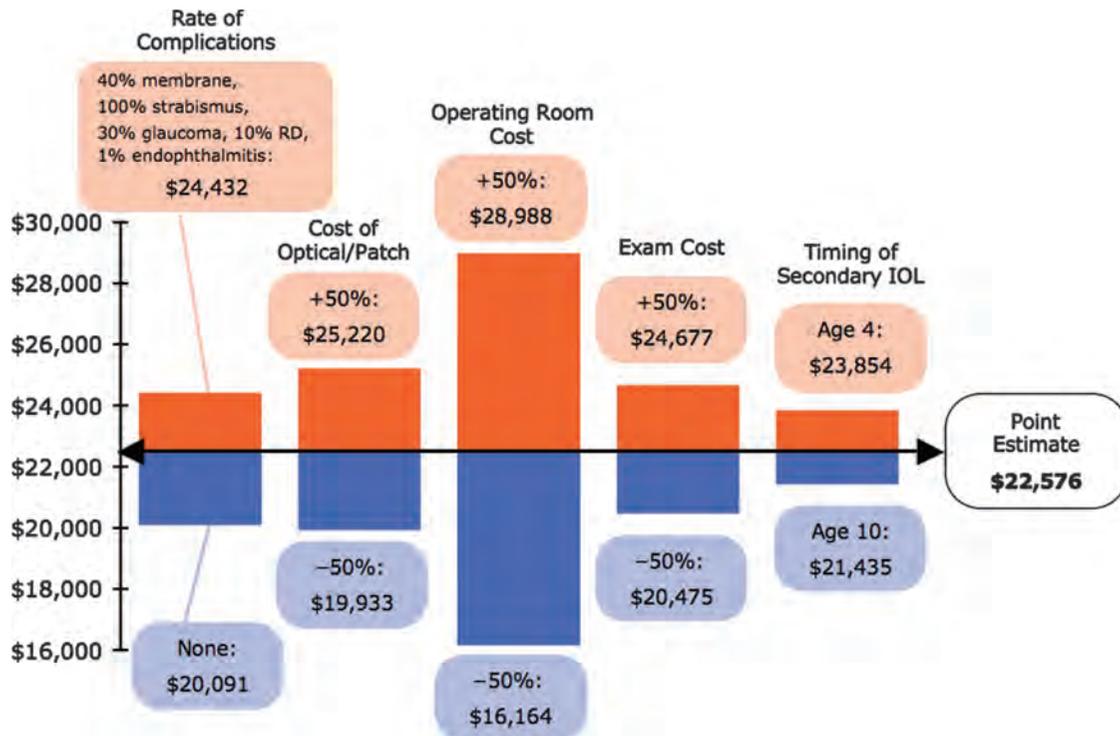


Figure 10.2. Univariate sensitivity analysis around the point estimate of \$24,650 total cost per patient. Bars indicate the range around the point estimate resulting from variation in each of the cost elements.

aphakia in infants with IOLs versus the traditional common approach of contact lens treatment followed by a secondary IOL later in childhood (Infant Aphakia Treatment Study, IATS).¹⁹

The cost analysis presented here was modeled after the “standard” treatment with aphakic contact lenses as outlined in the IATS trial, and resulted in a point estimate of \$22,576 total cost per patient through age 12 years. It should be kept in mind that a third-payer perspective was taken, based on Medicare reimbursement data, and that actual costs will likely be higher. The model can be adapted to different scenarios (cataract types and treatment approaches) and used for cost-effectiveness analyses (CEAs). For example, assuming a mean visual outcome in the treated eye of 20/70,²⁰ an associated quality-of-life (utility) gain of 0.1,²¹ and typical values for life expectancy and discounting, treatment in the above scenario leads to 2.9 quality-adjusted life years (QALYs). These results thus correspond to a cost-effectiveness ratio of approximately 7,800 US\$/QALY, comparable to the cost-effectiveness of laser photocoagulation therapy for subfoveal choroidal neovascularization,²² and should be considered highly cost-effective.²³ For comparison, estimates of the cost-effectiveness of routine cataract surgery in adults in developed countries range from 2,600 to 9,400 US\$/QALY.^{24,25}

CEAs have been carried out for a great number of conditions and their treatments, and attempts have been made to catalogue the wealth of results.²⁶ However, it

should be kept in mind that methodic differences across CEA have large effects on the results—both in terms of cost and in terms of QALYs. Also, there is no unanimous agreement on the oft-cited number of 50,000 US\$/QALY as a cutoff, below which a treatment should be deemed cost-effective.²³ A typical example of an “expensive” treatment is the liver transplant. Depending on methodology, cost-effectiveness estimates of liver transplants range from 36,000 to 100,000 US\$/QALY,^{26–28} and thus may or may not be cost-effective. Indeed, fair comparisons across treatment options from an economic standpoint require comparable health payer perspectives as well as comparable methods to elicit patient preferences. Within-study comparisons may be highly preferable over across-study comparisons. A within-study CEA comparing treatment options for congenital unilateral cataract in the IATS¹⁹ has recently been published.²⁹

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11

Historical Overview

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This chapter highlights the historical aspects of cataract management in children. We also take an opportunity to give credit to the innovators whose efforts have helped us reach the current level of success in the management of pediatric cataracts. In the past, poor outcomes from pediatric cataract surgery prompted many surgeons to use a *conservative approach* (described in Chapter 8). Since then, many surgical strategies have come and gone. They have, however, led us to where we are today. Our current “modern” techniques will also, someday, be included in the historical approach category. We need to study the past so the lessons learned do not have to be learned again. Also, one must not forget the giants of thought and skill on whose shoulders we now stand. They led the way to our current understanding and our current technology.

IS SURGERY RECOMMENDED FOR VISUALLY SIGNIFICANT BILATERAL CONGENITAL CATARACTS? IF YES, AT WHAT AGE?

Thick secondary membranes, glaucoma, and corneal decompensation occurred so frequently after pediatric cataract surgery that surgical aggressiveness seemed pointless to many surgeons.

In 1805, James Ware made the statement, “Operation is not so certain a cure for children as in persons of more advanced age.”¹ He further noted, “Children born with cataracts, are in general so unmanageable that the operation becomes almost impracticable. For these reasons it is advisable to

postpone it until they arrive at the age of reason and reflection, and feel by experience the necessity of submitting to it” (as cited in Ref.²). There has been a good deal of variance regarding the age at which congenital cataracts should be operated upon.

By 1830, Mackenzie questioned (as cited in Ref.²), “In cases of congenital cataract, ought the operation to be delayed till the patient has attained an age sufficient to enable him to give his assent, or ought to be practiced during infancy?” He then answered: “The answer decidedly is to operate in infancy—about the age of from 18 months to 2 years.”

Gurhrie in 1830 stated (as cited in Ref.²): “some surgeons recommending 6 months, others 20 months to 3 years. If the child is healthy at 3, 4 and certainly 6 months, the operation ought to be performed.”

In 1882, Critchett³ noted that “a considerable diversity of opinion still exists as to the period of life when operative measures become not only justifiable, but expedient, and also as to the form of operation which should be adopted in any given case.”

In 1908, Treacher Collins recommended: “to wait until a child is 10 months old before operating. Before that the amount of aqueous humour is so small that it does not suffice for solution of the liberated lens substance” (cited in Ref.⁴).

In 1936, Kirby (as cited in Ref.²) stated: “if the operation is decided upon, it may well be performed early, as good results are obtained with careful needling.”

In 1948, Cordes recommended that for bilateral dense infantile cataract, the first eye could be operated at 6 months of age, while second eye surgery could wait until 2 to 3 years of age.²

It was reported in the older literature that eyes operated for congenital cataract tended to develop retinal detachments (RDs), irrespective of what age the cataract surgery is undertaken. Unilateral surgery for bilateral congenital cataracts has been advocated in the past because of the reported high incidence of subsequent RD, bilaterality of complication, and poor outlook for successful RD repair.⁵ When the inevitable RD occurred approximately 25 years later, the cataract in the remaining eye could then be removed with the hope of restoring useful vision and function for another 25 years, until this eye in turn sustained a RD.

Current recommendation of timing of cataract surgery is described in Chapter 6.

SHOULD SURGERY BE DONE FOR UNILATERAL CONGENITAL CATARACT? IS THERE EVER A GOOD VISUAL OUTCOME?

Historically, the management of dense unilateral cataract was very controversial since dense amblyopia was always present and was often severe and unrelenting. Dense unilateral congenital cataract was once regarded as having a dismal visual prognosis. Even today, children with unilateral congenital cataracts remain challenging, and their visual outcomes are still often disappointing.

In 1957, Costenbader and Albert⁶ stated that they had not seen a single child with a monocular congenital cataract who had benefited from surgical removal of the lens opacity. They noted that it was best not to operate on eyes with a congenital monocular cataract because of morbidity and a poor visual result. They⁶ further noted that “...since visual acuity is not improved, strabismus is not favorable influenced, and photophobia is not alleviated, we would unequivocally advise against surgery in unilateral congenital cataract unless the cataract is becoming hypermature. The appearance of the eye is usually not improved; function has not been helped, and the eye has been jeopardized if surgery is performed...” (p. 428)

Frey and coworkers,⁷ in 1973, report that “...some children with monocular cataract can achieve useful central vision” and were the first to advocate that “the dictum of extreme conservatism in the management of monocular cataracts in children needs to be re-evaluated” (p. 388).

Even as late as 1979, Francois⁸ wrote that “everyone knows the uselessness of operating on unilateral congenital cataracts, as the functional result is always very bad.”

Subsequently, in a landmark paper, Bellar, Hoyt, and coworkers⁹ in 1981, demonstrated that excellent visual results could be obtained in selected children with monocular congenital cataracts. They reported early treatment and compliant contact lens wear and patching.

They emphasized the importance of detection and treatment during the sensitive period of vision development. They further noted,⁹ “... we believe that surgery during the neonatal period is not only justified but probably essential in any successful treatment of monocular congenital cataract...” (p. 564).

A review of the literature published from 1980 to 1993 revealed that 37% of infants with early surgery achieved a visual acuity outcome of 20/80 or better after surgery for dense unilateral congenital cataract.¹⁰ Birch and Stager¹¹ have reported that there exists a 6-week window of time, beginning at birth, during which treatment of a dense congenital unilateral cataract is maximally effective. If treatment is initiated during this period and the child is compliant with contact lens wear and occlusion therapy, an excellent visual acuity outcome can be obtained. Early treatment with good compliance is also associated with a lower prevalence of strabismus and a higher prevalence of fusion and stereopsis.

Taylor,¹² in his Doyne Memorial Lecture published in 1998, points out that unilateral congenital cataract is not a socially significant disease since it must be a coincidence of considerable rarity when a person who has such a cataract treated successfully (creating a spare eye) blinds the phakic eye. He points out that if days off work, long-distance travel, repeated clinic visits, fees, and all other disturbances are taken into account, the costs and the disturbance to the daily lives of patients and their parents are very substantial.¹² He points out that even at major referral centers, a good functional result (e.g., driving vision) is unlikely to be achieved in more than 50% of patients presenting with unilateral congenital cataract. Taylor¹² acknowledged, however, that an increasing number of cases with some degree of binocular vision were being reported; this has functional benefits of its own and, also, reduces the normally very high incidence of strabismus.

In 2001, Wright stated, “Not all children treated aggressively will obtain that lofty goal of good visual acuity and binocular vision, but it is guaranteed that without aggressive treatment virtually all children with a visually significant cataract at birth will end up with a blind eye and strabismus...” (p. 1122).¹³ We concur with Wright’s view in our practice and continue treating these eyes surgically as soon after 30 days of life as possible.¹³ In recent years, dense monocular infantile cataract has changed from a “hopeless” disease to a treatable, although demanding and sometimes frustrating, condition. If treatment is initiated in the ideal 4- to 6-week age (allowing the newborn to “feed and grow” for 30 days but still removing the unilateral cataract during the 6 weeks window), and the child is compliant with optical correction and occlusion therapy, an excellent visual acuity outcome can be obtained.¹¹ In the past few years, advances in surgical

techniques and aphakic correction have occurred but amblyopia is still the major determinant of the ultimate visual outcome of pediatric cataract surgery. Gradually, the evolution of cataract surgical techniques combined with early detection and surgery and the ability to provide early, effective, and constant optical rehabilitation led to better outcomes.

OPERATIVE TECHNIQUES

In 1951 Cordes described that the ideal operation for congenital cataract should meet the following conditions.¹⁴

- It would be a single procedure providing for the removal of the major portion of the lens, so that the rest could absorb.
- There would be no interference with the pupil, and the danger of incarceration or adherence of the iris to the wound would be minimal.
- There would be no interference with the posterior capsule and the vitreous.
- The danger of glaucoma would be minimized.

Unfortunately, this ideal operation did not exist then and still does not exist today. Pediatric cataract surgery involves higher rate of intraoperative and postoperative complications, and we have learned that dealing safely with the necessity of “interference” with the posterior capsule and the vitreous is a must for the young child. Innovations in surgical techniques and technology have allowed us to achieve improved anatomical and visual outcomes from our pediatric cataract surgeries.

Optical Iridectomy

For visually significant congenital cataracts, some surgeons resorted to an optical iridectomy to avoid the inflammatory response and membranes that developed when the lens was entered (Fig. 11.1). Optical iridectomy was useless when total cataracts were present. However, it was recommended in cases of fetal nuclear opacity, zonular

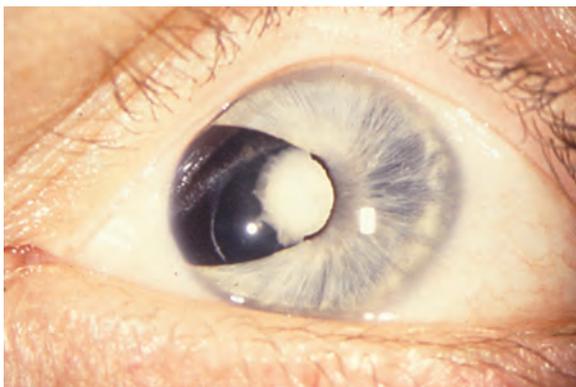


Figure 11.1. Optical iridectomy in an eye with congenital nuclear cataract.

or polar cataracts in which dilation of the pupil improved vision.¹⁴ Optical iridectomy was often performed for the purpose of helping near vision, in the inferior nasal sector between 4 and 5 o'clock in the right and 7 and 8 o'clock in the left eye. Some surgeons preferred the 12 o'clock position for cases where the development of the cataract would later make a more radical intervention necessary. However, the upper eye lid covered the upper part of the cornea and reduced, to some extent, the benefit of the intervention.¹⁵ This optical iridectomy technique preserved accommodation and avoided secondary membranes.⁶ In multi-handicapped children with bilateral central fetal nuclear cataracts, this technique was advocated also to avoid contact lens and spectacle problems. However, the visual results were not satisfactory. Despite bilateral surgery, nystagmus nearly always developed and vision was rarely better than 20/200 even when a clear zone existed peripheral to the fetal nuclear opacity.¹⁶

Discission/Needling

Although couching for cataract was practiced in the Hellenistic period, its use in children was not ideal: an already tricky operation was turned into an exceptionally difficult one by the child's anatomy and the lack of an anesthetic (cited in Ref.¹²). Needling/discission is probably the oldest of all surgeries for removal of congenital cataracts. Aurelius Cornelius, a Roman physician who lived 2,000 years ago (quoted in Ref.¹⁷), first described discission of soft cataracts. The technique used by the ancients was credited to Potts in 1775 (quoted in Ref.¹⁶). Because of its simplicity, discission remained the method of choice until the middle of the 20th century.

The discission was developed because of surgeons' experiences with the couching operation. It is based on the observation that the lens material of small children is comprised of soluble protein, which is often absorbed over weeks or months following an incision of the anterior lens capsule. The discission involves lacerating the anterior capsule of the lens (in one of several shapes and sizes) and allowing the lens material to be absorbed by the aqueous. Most practitioners used an anterior approach, but Saunders advocated a posterior approach, because he considered the dispersion of the lens material into the vitreous to be more effective (cited in Ref.¹²).

In 1968, Chandler^{18,19} reported the outcome of discission surgery without iridectomy in 33 eyes. Nearly one-fifth of the eyes were eventually lost, mostly secondary to complications related to chronic inflammation, scarring, and pupillary block. A thick capsular membrane developed in nearly all eyes undergoing this procedure and secondary glaucoma also occurred frequently. One of the disadvantages was that the procedure needed to be repeated a number of times, as many as 17 or 18 times before a good opening or good visual results were obtained.

Derby stressed the importance of a widely dilated pupil (quoted in Ref.¹⁶). He felt that the degree of pupillary dilation should be determined before deciding on the type of surgery to be performed. If the pupil failed to dilate, he advised preliminary iridectomy 3 or 4 weeks before doing a discission. Scheie¹⁶ noted, in 1960, that this rule is equally valid for all types of operations for congenital cataract. Even as of today, in 2013, we cannot stress enough how important the pupil size is. Failure to dilate the pupil reflects iris hypoplasia, and outcome, in general, is not as desirable.

Through-and-Through Discission

This variation of the discission procedure was described by Ziegler.²⁰ He incised the lens through both its anterior and its posterior capsule so as to disperse the lens proteins into both the aqueous and the vitreous humor in hopes of promoting complete absorption. The procedure did not find widespread acceptance.

Linear Extraction

For many children, the needling procedure alone proved to be inadequate. The remaining lens material did not absorb completely and often produced an inflammatory reaction. The removal of lens substance with irrigation was sometimes performed at the same time as the needling procedure (one-stage procedure) or a week or more later (two-stage procedure). In children <6 years old, the needling and removal of lens substance could be performed as a one-stage procedure. However, in older children a preliminary needling was preferred, to allow greater mixing of the lens cortex and aqueous. This permitted the lens cortex to fluff up and become flocculent, thus easing the irrigation of lens material from the anterior chamber out, through a wound made at the limbus.

In the first half of the 20th century, this needling plus irrigation procedure was the most common procedure for surgery on congenital cataracts.²¹ Gibson is credited for the technique based on a description in 1811. It was subsequently popularized as linear extraction (cited in Ref.¹⁶). Gibson's original technique involved using a couching needle to rupture the anterior capsule. The eye was left to settle over a period of 2 or 3 weeks. An incision was then made *"by a corneal knife of the largest size"* and was used to extract the cataract; as the knife was withdrawn, aqueous and some of the lens tissue evacuated spontaneously or was helped by a curette (cited in Ref.¹²). Many modifications of Gibson's technique were used. A capsule forceps was sometimes introduced into the anterior chamber to express dense opaque lens material. A hook or curette was also sometimes used to massage the cornea to express this material without the introduction of an instrument into the anterior chamber. An irrigation needle was often inserted into the anterior chamber, and

the saline irrigation flushed the remaining fragments of the lens from the chamber through the corneal wound.

In 1965, Ryan et al.²² reported better results with linear extraction compared to repeated discissions. Chandler¹⁸ reported that the results of linear extraction and iridectomy were poor if the operation was done in infancy. An updrawn pupil and dense secondary membranes formed. Most of the eyes were lost.

Aspiration of Cataracts

In the early 1960s, Scheie popularized the aspiration procedure. With the widespread use of the operating microscope, the Scheie procedure became the accepted technique for extracting cataracts in infants and children. In 1977, Scheie and Ewing¹⁷ described the history of the technique. Rhazes, a Persian physician and philosopher, mentioned aspiration, as did Antyllus (a contemporary of Galen), in the fourth century AD. Aspiration was also practiced in Japan in the 15th century, in Italy in 1829, and by Laugier in 1847. Although described as early as the fourth century, Scheie's report in 1960¹⁶ reawakened ophthalmologists' interest in the aspiration technique and repopularized it.^{22,23}

Scheie^{16,17,24,25} reported that the aspiration technique could be performed as a one-stage procedure in eyes with complete cataract or as a two-stage procedure in eyes with partial cataract. The first step was to open the anterior capsule widely and the second step was to aspirate the lens material via the same puncture site using a large-bore, 19-gauge needle and a 2-mL syringe. Many ophthalmologists later reported that the two-stage procedure was unnecessary and also dangerous, since it led to a higher incidence of glaucoma.²⁶

Parks²⁶ reported modifications of the aspiration technique. His technique differed from the classic method in that it aspirated the lens cortex while leaving the anterior lens capsule intact except for a 2-mm opening near the superior pupillary margin. The coaptation of the collapsed anterior and posterior lens capsule provided a framework for the regrowth of a sheet of new lens fiber, almost always needing secondary postcataract surgery within a few months. The secondary surgery could be avoided by creating a 5-mm central opening in the coapted anterior and posterior lens capsule after the aspiration of the cortex was completed.

The Scheie aspiration technique was simple and safe. However, since the cortex removal was often incomplete and the posterior capsule was left intact by most surgeons, secondary opacification of the visual axis occurred frequently and multiple additional procedures were often required.

Irrigation-Aspiration Technique

In the mid-1960s, a double-barreled cannula was introduced, with one barrel for aspiration and one for irrigation.

The dual irrigation–aspiration technique enabled the ophthalmologist to maintain the anterior chamber depth during cataract aspiration.

Intracapsular Extraction

Classic intracapsular surgery found disfavor among most ophthalmologists for children because of the higher risk of vitreous loss caused by the forces required to rupture the tough zonule in young patients.^{16,22} A large incision was also needed, which led to wound-related complications. Results using chymotrypsin were not encouraging in congenital cataract surgery.

Phacoemulsification

In the 1970s, phacoemulsification was used in pediatric cataract surgery for the first time.²⁷ At most, only short bursts of ultrasound power were used in pediatric eyes. However, even without ultrasound power, the phacoemulsification handpiece was an effective irrigation/aspiration device. If any hard material was encountered, ultrasound power was available to help remove it. Hiles et al.²⁸ and Callahan²⁹ recommended phacoemulsification as a useful extension of the aspiration technique in children since the softness of the lens material can vary greatly from case to case.

Automated vitrector

In the first two-thirds of the 20th century, pediatric cataract surgery was completely nonautomated. The advent of vitreous suction cutting devices in the mid-1970s revolutionized pediatric cataract surgery.^{30–34}

In 1975, Calhoun and Harley described their initial experience using a new automated cutting instrument known as a roto-extractor. The cataract procedure was performed under an operating microscope through a 2.4-mm incision made 1 mm posterior to the limbus.³⁵ They operated 66 eyes of 57 patients beginning in the spring of 1974 when they purchased the instrument, which had been designed by Douvas and introduced at the American Academy of Ophthalmology meeting in late 1972. Unlike the early vitreous infusion suction cutter (VISC) designed and introduced in 1972 by Machemer, the roto-extractor was a side-cutting instrument. The instrument had self-contained irrigation, aspiration, and cutting. The fluid was gravity fed, and the aspiration was via a syringe. The cutting blade could be either rotated or oscillated by a motor that was contained in the handpiece and controlled by the surgeon via a foot pedal. They continued a sequence of positioning of the tip, application of suction, and activation of the cutter until the pupillary aperture was clear. The authors noted that they removed a portion of the anterior vitreous and the posterior capsule in nearly all cases.³⁵ Usually, the anterior capsule and cortex of the lens were removed first. Removing the posterior capsule

was usually performed near the end of the procedure when a better view of this structure could be obtained. This visualization was aided by coaxial illumination. The most frequent complications were unplanned defects in the pupil margin (an iridectomy or sphincterectomy that was grossly visible) and persistent inflammation (pain, photophobia, ciliary injection, and anterior and vitreous inflammation lasting longer than a week). Each of these occurred in 9 of 66 eyes. With short follow-up, the visual axis was clear in all but 3 of the 66 eyes.

In 1977, Peyman described a pars plicata approach for removal of congenital cataracts using a cutting device (cited in Ref.³⁶). In 1978, Peyman and Goldberg and coworkers published results of lensectomy and pars plicata vitrectomy in children.³⁶ They noted that the pars plicata approach avoided direct manipulation of the corneal endothelium and eliminated the unplanned iris sphincterectomies. In 1978, Kanski reported using vitrectomy techniques in congenital cataract removal.³⁷ Kanski noted that the technique has all the features of an infusion–aspiration system, with the added advantage of a cutting capability for the removal of anterior vitreous, posterior capsule, and any calcified lenticular plaques that may be difficult to remove by simple aspiration. Using this new technology, Taylor³⁸ and Parks³⁹ began cautiously removing the center of the posterior capsule and a portion of the anterior vitreous during the initial surgery in all young children needing cataract surgery. Removal of all but 2 mm of the peripheral posterior lens capsule with a vitreous suction cutter was recommended, in addition to a generous anterior vitrectomy. In 1981, Taylor³⁸ reported needing no reoperations in 23 infantile eyes operated with lensectomy using the vitrectomy machine to remove the whole lens including both the anterior and posterior capsule. In contrast, of 28 age-matched eyes (collected from just prior to beginning the new procedure) in which the posterior capsule was left intact, 19 eyes required a total of 32 reoperations to keep the visual axis clear (29 needlings and 3 were capsulotomies using the new vitrectomy machine). Taylor noted that vitrectomy has two major advantages: first it is a good technique for dealing with other ocular anomalies associated with congenital cataract and second, a clear pupillary axis can be guaranteed in the crucial first 18 months after surgery during which the child's developing vision is most sensitive to the effects of a degraded visual input. On the other hand, anticipated disadvantages were the increased risk of RD and cystoid macular edema.

In addition to its use as a cutting device, the various vitreous suction cutting instruments were used for automated irrigation/aspiration of lens cortex and nucleus. The often thick and gummy lens material found in children was more easily and efficiently aspirated using this instrument as opposed to the slow and incomplete manual irrigation and aspiration. Also, bursts of cutting could be

used for any lens material that was initially resistant to aspiration. After their introduction, manufacturers made advances quickly and the instruments became smaller and more efficient. The O'Malley Ocutome became a favorite of many pediatric surgeons in the early and mid-1980s when it was introduced with a safer and more effective guillotine-like cutting mechanism rather than a rotary or oscillating blade. It had a vacuum pump to produce suction force and could be easily activated or interrupted by the surgeon. Bimanual techniques gradually became the standard as manufacturers developed instruments that allowed the separation of irrigation and aspiration/cutting. This separation, while not as important when aspirating lens cortex, was recommended whenever a vitrectomy was needed. Later, these vitrector machines were also fit with separate (noncutting) irrigation/aspiration handpieces designed specifically for removal of the lens cortex.

Intraocular Lens Implantation

After the successful implantation of an intraocular lens (IOL) in an adult by Sir Harold Ridley in 1949,^{40,41} Epstein placed an IOL in the eye of a child as early as 1951 (cited in Ref.⁴²). Peter Choyce⁴³ reported the implantation of an IOL in a 10-year-old child in 1955. Binkhorst and coworkers^{44,45} implanted iridocapsular fixated IOLs in children in 1959. Hiles advocated IOL implantation in children and published many articles related to his experience.⁴⁶⁻⁵² Sinskey and Patel,⁵³ BenEzra and Paez,⁵⁴ and Dahan and Salmenson⁵⁵ were also early advocates of IOL implantation in children.

However, early attempts at IOL implantation in children resulted in frequent complications secondary to poor lens design and the greater inflammatory response that occurred in these implanted eyes after intraocular surgery. As a result, IOL implantation at the time of cataract surgery in children did not become common practice until the 1990s, 20 years after the adoption of automated aspiration and cutting instruments for capsulectomy and vitrectomy in children with cataracts.

SUMMARY

Automation and IOLs have brought pediatric cataract surgery into the modern age and the resulting outcomes are much improved. The evolution of this surgery continues, with many adult cataract surgery techniques being applied to older children with only minor technical adjustments. In addition, new techniques designed specifically for children have emerged. In very young children, automated posterior capsulotomy and anterior vitrectomy continues to provide the best long-term outcomes, with fewer reoperations and

complications compared to older methods described in this chapter. For pediatric cataract surgery, the unique needs of the pediatric cataract surgeon were met by incorporating automatic irrigation/aspiration and cutting instrumentation. This technology, developed initially to provide retinal surgeons a way to remove diabetic vitreous hemorrhage, was adapted and adopted by innovative pediatric eye surgeons soon after its introduction. These trailblazers revolutionized outcomes in congenital cataract surgery at a time when the status quo was clearly not satisfactory.

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Endophthalmitis Prophylaxis

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Post-cataract surgery endophthalmitis represents an infrequent but devastating complication of cataract surgery. In a systematic review of 215 studies reporting on 3,140,650 adult cataract extractions, the endophthalmitis rate was 0.128%.¹ Swedish national cataract registry of 1 million cataract surgeries between 1998 and 2009 reported endophthalmitis has decreased from 0.1% to below 0.04%.² Analysis of Medicare database reported national rate in the United States as 0.133% in 2003 and 0.111% in 2004 for adult cataract surgery.³ Good et al.⁴ reported, in 1990, an incidence of 0.45% in children. Wheeler et al.⁵ surveyed 500 pediatric ophthalmologists and reported endophthalmitis incidence of 0.071% following pediatric intraocular surgery for cataracts and congenital glaucoma. Although endophthalmitis occurs infrequently, the visual morbidity is high even with appropriate treatment, and consequently, ophthalmic surgeons have adopted a variety of prophylactic techniques.⁶ Prophylaxis of endophthalmitis in children typically mirrored that in adult endophthalmitis.

SOURCES OF CONTAMINATION

While infections may be induced by contaminated surgical products or contaminated operating room environments, it has been established that the patient's periocular flora is the source of the microbes responsible for most cases of sporadic postoperative infection.⁷ Organisms isolated from the vitreous were indistinguishable from those recovered from the eyelids, conjunctiva, or nose in 14/17 (82%) cases of endophthalmitis.⁷ The most common organisms isolated were gram-positive coagulase-negative cocci, including *Staphylococcus epidermidis* and *Staphylococcus aureus*.⁷ These organisms are the most common organisms recovered from the eyelids.^{6,7} In the Endophthalmitis Vitrectomy Study, where analysis was possible, postoperatively cultured eyelid isolates were indistinguishable from intraocular isolates in 67.7% (71/105).⁸

CAUSATIVE ORGANISMS

Numerous bacterial and fungal organisms have been found to cause postoperative endophthalmitis, but gram-positive organisms are the most common cause of endophthalmitis after cataract surgery. Gram-positive, coagulase-negative bacteria caused endophthalmitis in 70% of cases in the Endophthalmitis Vitrectomy Study.⁹ This same study found that 9.9% of culture-proven cases of endophthalmitis were caused by *S. aureus*, 9.0% by *Streptococcus* spp., and 2.2% by *Enterococcus* spp. Various gram-negative organisms accounted for 5.9% of isolates.⁹ Wejde et al.,¹⁰ in a study of over 188,000 patients in Sweden from 1999 to 2001, found a predominance of gram-positive organisms as well, with 84.6% of their culture-proven cases revealing gram-positive organisms. Interestingly, they noted that there was a relative increase in the number of cases of endophthalmitis caused by *Enterococcus* spp. when compared with a study that had reviewed the same population in 1998. The incidence of methicillin-resistant *Staphylococcus aureus* (MRSA) infections is rising.¹¹

RISK FACTORS FOR ENDOPHTHALMITIS

Wound Construction

Since surgical incisions likely provide the entry site for infecting organisms, wound construction can play a role in the incidence of endophthalmitis. The intraocular concentration of organisms is highest at the time of surgery, after which it usually declines. If there is ingress of fluid from the ocular surface after cataract surgery, bacteria can be introduced at that time, increasing the likelihood of the development of endophthalmitis. An increased rate of post-cataract surgery infections has been reported during the last decade.^{1,12,13} It has been proposed that the increased infection rates correspond to the increased use of clear corneal incisions for cataract surgery, because improperly constructed clear corneal cataract incisions are more prone to postoperative instability, leakage, and a potential influx of microbes than are sclerocorneal incisions.¹⁴

Few series found no greater likelihood of infection with corneal versus other types of incisions.^{15,16} Nevertheless, careful watertight incision construction and closure (with or without sutures) is mandatory, irrespective of surgical style, because the incidence of infection increases when there is a wound leak.¹³ Pediatric cataract surgeons tend to suture the wound more often than do adult cataract surgeons.

Surgical Complications and Techniques

Surgical complications prolong the time spent in surgery and thus the exposure to potential infectious agents. Iris prolapse during the procedure, inadvertent rupture of the posterior capsule, and wound leaks are all potential complications that may increase the risk of postoperative endophthalmitis. Techniques that require multiple entries and exits of instruments into and from the eye may introduce more bacteria at the time of surgery. Any break in the posterior capsule that usually separates the anterior from the posterior chamber may lead to higher prevalence of endophthalmitis. In pediatric surgeries, the necessary removal of the posterior capsule in very young patients to prevent reopacification theoretically gives bacteria easier access to the vitreous cavity.

Other Factors

Immunodeficiency, active blepharitis, and lacrimal duct obstruction may increase the risk of infection. Good et al.⁴ noted the role of coexisting upper respiratory infection in postoperative endophthalmitis for children. The authors noted that upper respiratory infection is so common and repetitive in young children that their existence can be overlooked. All three cases reported by Good et al. received clearance for anesthesia from their pediatricians. Endophthalmitis is usually the result of introduction of bacteria into the eye at the time of surgery. However, Good et al.⁴ noted that they cannot exclude the possibility that the fitting of a contact lens in one of their case was the source of bacterial introduction, especially since infection occurred 5 days postoperatively, 2 days after contact lens fitting. Eyes with traumatic cataract and those receiving secondary intraocular lens (IOL) implantation may be at higher risk for developing infection. Although prevention of endophthalmitis is essential for every cataract surgery, eyes with traumatic cataract, reoperation (e.g., secondary IOL), and immediately sequential bilateral cataract surgery are especially more important to keep in mind. Eyes receiving immediately sequential bilateral cataract surgery require that surgeon treat both eye surgeries as completely separate procedures. Details are discussed in Chapter 9.

PROPHYLAXIS

The risk for endophthalmitis may be lessened by reducing the number of microbes on the ocular surface, by reducing the opportunity for microbes to reach the intraocular

environment during or after surgery, or by eliminating those organisms that may have reached the eye intra- or postoperatively. Given the ability of surface flora to enter the eye during surgery, many of the prophylactic techniques to decrease the risk of endophthalmitis aim to suppress their number and to limit the growth of those organisms that do enter the eye before the development of frank endophthalmitis.¹⁰ Although some recently developed oral quinolones may achieve significant intraocular levels, many systemically administered antistaphylococcal antibiotics have not demonstrated good intraocular penetration. Consequently, various routes of administration have evolved including preoperative topical, intraoperative infusion, and postoperative subconjunctival routes.¹⁰ Numerous factors may affect the surgeon's choice regarding the type and route of antibacterial administration, including the spectrum of bacterial coverage, potential adverse effects, efficacy, ease of application, cost, local standards of care, and personal experience. Our current (2013) practice is listed in Table 12.1.

Preoperative Prophylaxis

In addition to measures for hand disinfection, preoperative sterile preparation of the surgical site, and proper draping methods, the following measures have been reported.

Preoperative Topical Drops

Topical application of antibiotics appears rational to reduce the number of bacterial organisms in the cul-de-sac. Use of preoperative, patient-administered topical antibiotics in routine intraocular surgery received a level C clinical rating, as possibly relevant but not definitely related to clinical outcome.⁶ The use of preoperative antibiotic drops is still debated. Several studies have been cited as evidence of the lack of efficacy of topical antibiotics in reducing the rate of contamination of the anterior chamber at the time of surgery or in reducing the

Table 12.1

CURRENT (2013) ANTI-INFECTIVE PROTOCOL FOR PEDIATRIC CATARACT SURGERY AT STORM EYE INSTITUTE

- Fresh bottle of moxifloxacin (Vigamox) is opened when the patient arrives in the operating room
- Antibiotics are dripped into a sterile tray on the operating sterile field where the scrub nurse will draw up the intracameral dose (0.1 mL of 50% dilution, 250 µg). Note that each milliliter of Vigamox solution contains 5 mg (5,000 µg) moxifloxacin base.
- Before prep, Vigamox and 5% povidone-iodine are given
- Intracameral Vigamox is given at the conclusion of surgery
- The 5% povidone-iodine and Vigamox drops are given at the conclusion of surgery
- Same Vigamox bottle is sent home with parent for use q.i.d. for 1 week.

rate of endophthalmitis.^{17–20} Nevertheless, preoperative antibiotics are commonly used. Fourth-generation fluoroquinolones have emerged as the most commonly prescribed topical prophylactic therapeutics because of their broad spectrum activity and superior ocular penetration.¹¹

The optimal timing and frequency of topical antibiotic prophylaxis has also been the subject of debate. Inoue et al.²¹ assessed the optimal duration of the preoperative application of 0.5% levofloxacin ophthalmic solution in a prospective randomized multicenter study. Two hundred and seventy-two elderly cataract patients who had undergone phacoemulsification with IOL insertion at 12 clinical facilities in Japan were randomized into three groups: 3-day, 1-day, and 1-hour preoperative application of 0.5% levofloxacin. The 3-day application of levofloxacin had a significantly higher disinfection rate than the 1-day or 1-hour applications ($P < 0.05$). Based on pharmacokinetics of these drugs, starting 1 to 3 days before surgery may be advantageous.¹¹ However, there is also evidence that frequent instillation of topical fluoroquinolones immediately before surgery may increase the concentration of drug in the anterior chamber and decrease the bacterial load.¹¹ In children, with questionable compliance, a common practice is to start topical antibiotic drops on the same day as surgery, either as soon as possible after patient check-in for surgery or when the patient arrives in the surgical suite prior to the prep.

Povidone–Iodine

With regard to clinical practice recommendations, povidone–iodine preparations received a level B clinical rating (highest), because it was deemed moderately important to clinical outcome.⁶ The strength of the data supporting this recommendation is relatively high compared with other prophylactic techniques in that the literature shows fewer conflicting studies with regard to its usefulness. There are convincing data suggesting that povidone–iodine decreases the conjunctival flora.²² Several studies have evaluated its use in a prospective manner. One group performed a prospective study in 4,110 patients with random patient allocation to 2.5% povidone–iodine application to skin and conjunctiva compared with application to skin only.²³ There was no difference in postoperative endophthalmitis incidence, but infected cases were not culture proven.²³ Another group of investigators conducted a large open-label nonrandomized parallel trial during an 11-month period in which topical 5% PI was applied preoperatively in one set of five operating rooms, whereas silver protein solution was applied in another set of five rooms.²² In all cases, surgeons continued to use their customary prophylactic antibiotics. The povidone–iodine group showed a significantly ($P < 0.03$) lower incidence of culture-positive endophthalmitis (2 of 3,489; 0.06%) compared with the silver protein group (11 of 4,594; 0.24%). There were no adverse reactions to

povidone–iodine.²² Inoue et al.²¹ compared disinfection by povidone–iodine solution with that by polyvinyl alcohol–iodine (PAI) solution in a preoperative eyewash in a prospective randomized multicenter study. The disinfection rate of povidone–iodine was 78.0%, and that of PAI was 79.4%; PAI was not inferior to povidone–iodine as a preoperative disinfectant. For *skin antisepsis*, a 10% povidone–iodine solution is widely used. In the periorbital region with its many sebaceous glands, the antiseptic should be administered about 10 minutes before surgery to act sufficiently. For *antisepsis of the conjunctiva*, povidone–iodine may also be used. As little as 1% of the solution reduces conjunctival contamination.

Saline Irrigation

Saline irrigation is commonly performed before intraocular surgery. However, this intervention has not been shown to reduce ocular surface flora or aqueous contamination. Saline irrigation received a level C clinical rating, because it cannot be definitely related to clinical outcome.⁶

INTRAOPERATIVE PROPHYLAXIS

Irrigating Solution

According to surveys carried out in various countries, antibiotics in the irrigation solution during phacoemulsification are used in varying percentages by the responding eye surgeons. In several publications and letters to the editor, it has been suggested that the addition of antibiotics to the irrigation solution should have a protective effect, but this has not been confirmed yet by any prospective study, nor has it reduced the incidence of endophthalmitis. In addition, these suggestions have been based on retrospective data or on studies of antibiotic use without control groups. The available evidence suggests that antibiotics placed in the infusion fluid do not help prevent endophthalmitis.^{20,24,25,26}

Intracameral

Intraocular penetration of topically applied antibacterials can vary significantly. A direct injection of an antibiotic into the anterior chamber following surgery should theoretically be the most efficient method for delivering therapeutic antibacterial concentrations to the eye at the end of surgery, eliminating susceptible bacteria introduced during the operation.²⁷ There has been concern about antibiotic toxicity, especially given the possibility of dilution errors when large numbers of operative eyes receive prophylactic antibiotics intracamerally. Aminoglycosides demonstrate significant retinal toxicity, including devastating macular infarction, in cases of dilution error, and their use should be avoided.

The European Society of Cataract and Refractive Surgeons (ESCRS) multicenter randomized prospective

study of the prophylactic effect of intracameral cefuroxime injection at the conclusion of the procedure and/or perioperative levofloxacin eye drops on the incidence of endophthalmitis after phacoemulsification was halted early because of results of a beneficial effect of intracameral cefuroxime. With data from 13,698 patients with complete follow-up records, investigators found that the odds ratio for developing endophthalmitis was 4.59 (95% CI, 1.74–12.08; $P = 0.002$) in the group not receiving intracameral cefuroxime injection. An earlier retrospective study in Sweden reported efficacy of intracameral cefuroxime, a second-generation cephalosporin, in reducing postcataract endophthalmitis. Two retrospective studies in Spain have reported that intracameral injection of cefazolin, a first-generation cephalosporin, reduced postcataract endophthalmitis. In 2012, a prospective French study of 5,115 consecutive adult cataract surgeries was reported. In 2,289 surgeries, cefuroxime was given intracamerally and in 2,826 surgeries it was not. All other aspects of the preoperative and postoperative care of the patients were identical. The incidence of endophthalmitis was 35 (1.238%) of 2,826 patients without cefuroxime and 1 (0.044%) of 2,289 patients with cefuroxime ($P < 0.0001$). While univariate analysis found the absence of cefuroxime ($P < 0.0001$), male sex ($P < 0.05$), and surgeon in training ($P = 0.0676$) to be associated with an increase in endophthalmitis, only the absence of cefuroxime was found to be significant on multivariate analysis. Importantly, no cases of toxicity were reported and no dilution errors were detected in the French study. The method recommended for preparation of the intracameral injection is worthy of mention. Cefuroxime was prepared extemporaneously on the operating table at the end of cataract surgery by the operating nurse. A 15-mL solution of cefuroxime containing 1,500 mg of cefuroxime was transferred into a sterile cup on the operating table. The surgeon then aspirated 0.1 mL (containing 10 mg of cefuroxime) of the solution into a 1.0-mL syringe. The solution was further diluted 10 to 1 using balanced salt solution, and 0.1 mL or 1.0 mg of cefuroxime was injected into the anterior chamber at the end of surgery after the surgeon verified that the corneal wounds were sealed.²⁸

Although the intracameral delivery of antibacterials has been shown to be effective in preventing endophthalmitis, the optimal choice of drugs has yet to be determined. Intracameral cefuroxime and moxifloxacin are currently most commonly used drugs. As a result of the ESCRS study, cefuroxime is the most commonly used intracameral antibiotic in Europe. It remains the only intracameral antibiotic shown by a randomized prospective trial to reduce the incidence of endophthalmitis. Vancomycin is also commonly used as an intracameral antibiotic. The Centers for Disease Control and Prevention (CDC) has limited routine application of vancomycin because

of rising concerns over resistant organisms. However, intracameral use in ophthalmology carries a very low risk for producing resistance.

Of the late-generation topical fluoroquinolones, moxifloxacin has been the most studied, in part because one of its commercially available topical formulations (Vigamox) is already preservative free and seems to have no obvious toxic effects if injected into the anterior chamber.²⁹ If given topically, four drops per day produces moxifloxacin therapeutic levels only up to 1.9 $\mu\text{g}/\text{mL}$ in the anterior chamber. If the dosage is increased to one drop every 10 minutes to 2 hours, the levels in the anterior chamber go up to 2.3 $\mu\text{g}/\text{mL}$. However, an intracameral injection of 250 μg dose of moxifloxacin produces aqueous humor concentration of 710 to 1,250 $\mu\text{g}/\text{mL}$ (250 $\mu\text{g}/0.2\text{--}0.35\text{ mL}$) with less systemic effects and less resistance.³⁰ Since aqueous humor is formed at a rate of 2 to 3 $\mu\text{g}/\text{min}$, the high aqueous humor concentration should be maintained for 1.5 to 2 hours after surgery.^{29,31–33} Aprokam[®] (Laboratoires Théa), premixed intracameral formulation, is now available in Europe as powder solution of Cefuroxime 50mg. This can be mixed with 5 ml of sterile sodium chloride injection, resulting in desired dose of 1 mg in 0.1 ml.

There is concern that late-generation fluoroquinolones may be less effective in preventing endophthalmitis caused by MRSA.²⁹ While intracameral cefuroxime has proven to be efficacious in preventing postoperative endophthalmitis overall, its activity against enterococci is poor, which accounts for a significant number of endophthalmitis cases in eyes receiving intracameral cefuroxime. Enterococcal endophthalmitis is associated with poor visual outcome, so this limitation of intracameral cefuroxime is of particular concern.²⁷ Cefuroxime concentrations higher than 2.75 mg/mL and vancomycin concentrations higher than 5.0 mg/mL led to significant reduction in cell viability.³⁴ This study reported dose-dependent toxicity of cefuroxime and vancomycin on human endothelial cell in vitro with a narrow range of safety and concluded that although the clinically used concentrations seem to be safe, slightly higher concentrations might induce irreversible cell death and thus should be avoided.³⁴ Commonly used dosage for intracameral use in adult eyes undergoing cataract surgery is listed in Table 12.2.

Table 12.2 INTRACAMERAL DOSE

- Cefuroxime: 1 mg in 0.1 mL saline (0.9%).
- Vancomycin: 1 mg of vancomycin in 0.1 mL of balanced salt solution
- Moxifloxacin: 250 μg of moxifloxacin (Vigamox brand has no preservative) given as 0.05 mL of undiluted³⁵ or 0.01 mL if diluted 1 to 1 with balanced salt solution. Prepare solution from a newly opened bottle using a tuberculin syringe.

POSTOPERATIVE PROPHYLAXIS

The very high concentration of antibiotic achieved from intracameral injection will be reduced 1.5 to 2 hours after surgery. Therefore, intracameral antibiotics alone offer no protection if there were bacterial ingress after the completion of the surgery, as might be expected in the case of a wound leak following surgery.²⁷

Subconjunctival

Evidence of the benefit of injecting subconjunctival antibiotics at the close of surgery is inconclusive and is associated with risks that include intraocular toxicity with the potential for macular infarction when aminoglycosides are used. Available literature indicates that it may be relevant but cannot be definitely related to clinical outcome (C level).⁶ Authors noted that the data supporting the use of subconjunctival antibiotics are somewhat stronger than the data supporting the use of the other prophylactic interventions, which also received clinical recommendation of “C” (including preoperative lash trimming, preoperative saline irrigation, preoperative topical antibiotics, irrigating solution containing antibiotics, and the use of heparin).⁶

Topical

Postoperatively, the application of 1.25% povidone-iodine leads to a significant reduction in conjunctival contamination. On the other hand, there are no studies confirming any benefit of postoperative antibiotics, especially after 24 hours, although most surgeons use an antibiotic ointment or solution at the end of the procedure and for several days postoperatively. Topical antibacterials are commonly used for the prophylaxis of endophthalmitis following cataract surgery, despite the fact that the clinical evidence for their efficacy is limited. In a survey of members of the American Society of Cataract and Refractive Surgery (ASCRS), 91% of respondents stated that they used topical antibacterials prophylaxis at the time of cataract surgery and 98% used topical antibacterials postoperatively.³⁶ Intraocular penetration of antibacterials varies greatly among topical antibacterials. There is evidence to suggest that late-generation fluoroquinolones penetrate the cornea well and achieve measurable concentrations in the anterior chamber aqueous fluid.

Drug Delivery Systems

Collagen shields presoaked in antibiotic solutions have been explored as a method of drug delivery to the eye.³⁷ The utility of this approach compared with the topical instillation is questionable, especially when the role of topical drops itself is questionable. Potential use of polymeric devices to achieve sustained release of antibiotics post cataract surgery may have several advantages including better compliance.³⁸

Regional Difference in Prophylaxis

Since there are numerous strategies one can pursue in the prophylaxis of postoperative endophthalmitis, there is significant practice variance, which may be related to regional and cultural influences.²⁷ The survey of the members of the ASCRS showed that only 30% of respondents used intracameral antibacterials. Of those who used intracameral antibacterials, 61% used vancomycin and 23% used a cephalosporin. The vast majority of respondents reported the routine use of topical fourth-generation fluoroquinolones as their antibacterial of choice.³⁶ In Europe, however, the publication of the ESCRS study of intracameral cefuroxime led to a change in practice. A recent survey in the United Kingdom showed that 63% of respondents used intracameral antibacterials. The majority of surgeons in that study reported using intracameral cefuroxime, and 48% of these had switched after the publication of the trial.³⁹ Although the reasons for such practice variance in prophylactic strategies are not entirely clear, factors that can influence regional differences may include historical practice, ease of application, commercial availability, medical-legal concerns, safety track record, experimental rationale, industry marketing, cost, and clinical evidence of effectiveness.²⁷

SUMMARY

- Incidence is reported as approximately 1 into 1,000;
- Povidone-iodine application prior to cataract surgery remains a universal practice and is backed by good-quality evidence.
- Given the absence of clear evidence about the benefit of other prophylactic measures, it is up to the ophthalmologist to decide on the use of any particular strategy in addition to povidone-iodine in the perioperative period.
- It is incumbent on the surgeon to assure that all incisions are closed in a watertight fashion at the end of the procedure.
- Several studies have reported advantage of intracameral antibiotics.

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13

Anesthetic Management of the Pediatric Patient with Cataracts

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Anesthesia for the pediatric ophthalmic patient presents many special challenges. These include control of the intraocular pressure (IOP), awareness of the oculocardiac reflex, thorough knowledge of drug interactions, smooth anesthetic maintenance and emergence, and control or prevention of postoperative nausea and vomiting (PONV).

PERIOPERATIVE ROUTINE AND SELECTION OF ANESTHETIC AGENTS

Stringent fasting guidelines and separation from parents make the perioperative period an anxious time for small children. Although the standard period of 8 hours of NPO status (fasting) for solid food and nonclear liquids is required, children may receive clear liquids 2 to 3 hours before surgery without increasing the risk of aspiration.¹ Adequate preoperative hydration with clear liquids decreases the incidence of intraoperative hypoglycemia, hypovolemia, and PONV. Parental separation and fears of pain and blindness after surgery can be effectively addressed with the preoperative interview and appropriate anxiolysis. Many institutions routinely use premedication for young children (between 10 months and 12 years of age) before ophthalmic procedures. Midazolam, 0.5 mg/kg (up to a maximum of 10 mg regardless of the patient's actual weight), is given as an oral premedication 10 to 20 minutes prior to surgery to alleviate separation anxiety and ease the induction of general anesthesia. It may be given with apple juice, lemon-lime soda, and grape syrup or combined with an acetaminophen elixir or ibuprofen elixir.

Infants and children may be induced by either intravenous or inhalational techniques, but most general anesthetics in young children undergoing cataract surgery are done with an inhalational induction having the IV placed after the child is anesthetized. The important exception to this is the pediatric patient with a personal or family history of malignant hyperthermia where inhalational agents are contraindicated.

Sevoflurane is the induction agent of choice in children, because it possesses a minimally offensive odor and is less stimulating to the airway. General anesthesia may be maintained with sevoflurane, isoflurane, or desflurane in air and oxygen. Desflurane has been associated with increased airway irritability when used for induction and is used mainly for maintenance of general anesthesia. Nitrous oxide, traditionally used with volatile anesthetics, may increase PONV. The cause-and-effect relationship between nausea/vomiting and nitrous oxide remains controversial. Since ophthalmic procedures increase the risk of PONV, a mixture of air/oxygen or 100% oxygen can be used in place of nitrous oxide. Nitrous oxide may be used in conjunction with sevoflurane to speed the induction of general anesthesia and then be turned off once the child is past the excitement stage of induction.

Propofol is the intravenous induction agent of choice for cataract surgery in pediatric patients with preoperative intravenous access. Propofol is an isopropyl phenol that has a rapid onset and offset of action. Intravenous injection of a therapeutic dose of propofol produces hypnosis rapidly with minimal excitation and postoperative residual effects. It is also believed to have antiemetic properties. A small intravenous bolus dose of propofol is often used in older children prior to placing an airway device, either an endotracheal tube (ETT) or laryngeal mask airway (LMA), as it puts the child into a deeper plane of anesthesia.

Ketamine hydrochloride, a dissociative anesthetic that can be used for the induction and maintenance of anesthesia, is occasionally utilized for pediatric patients undergoing short procedures, such as an examination under anesthesia.² An intramuscular injection of 5 to 7 mg/kg will provide approximately 30 minutes of anesthesia. Additional doses at one-half of the initial dose can be given intravenously. There is a prolongation of emergence with repeated doses. Some of the principal adverse reactions include tachycardia, hypertension,

laryngospasm, tonic-clonic movements, hypersalivation, nausea and vomiting, diplopia, nystagmus, and an elevation of IOP (which should be remembered when following pediatric patients with glaucoma). It is a respiratory stimulant that can become a respiratory depressant if an overdose is given.

During anesthesia, even in patients whose IOP is usually normal, an increase in pressure can produce permanent visual loss. If penetration of the globe occurs when the IOP is excessively high, blood vessel rupture with subsequent hemorrhage may transpire. The IOP becomes atmospheric when the eye cavity has been entered, and any sudden increase in pressure may lead to prolapse of the iris and lens and loss of vitreous. Since proper control of IOP is critical during such delicate intraocular procedures as in pediatric cataract surgery, ketamine is not the agent of choice.

However, in some developing world settings where inhalation agents are not available, ketamine is used for pediatric cataract surgery. After ketamine induction, a peribulbar injection of lidocaine is usually given followed by ocular massage prior to surgical entry into the eye.³

Bell phenomenon can be seen at any time during anesthesia when the patient's depth of anesthesia has changed. It manifests as an upward (or sometimes downward) movement of the eye. It is usually a sign of the patient being in a lighter stage of anesthesia but not awake. This phenomenon can make exams under anesthesia, corneal measurements, and A and B scans difficult or impossible to perform. The anesthesia provider should be aware of this reaction and be prepared to "deepen" the anesthesia to facilitate these procedures.

Phenylephrine drops are commonly used to produce pupillary dilation in order to facilitate surgical access.⁴ As an alpha agonist mydriatic agent, some dramatic side effects may occur, ranging from transient hypertension with reflex bradycardia, ventricular dysrhythmias, and even pulmonary edema. For this reason, a 2.5% solution rather than a 10% solution of phenylephrine is recommended. Furthermore, absorption via the nasal mucosa may be reduced by occlusion of the nasal puncta via pressure on the inner canthus to prevent unintentional routing of the drug through the nasolacrimal duct. Treatment of the iatrogenic hypertension with beta-blockers is contraindicated since it can produce unopposed alpha-adrenergic stimulation. The anesthesia provider should be informed at the time of drop instillation so that he or she may monitor for a hypertensive response.

OCULOCARDIAC REFLEX

The oculocardiac reflex is triggered by pressure on the globe and by traction on the extraocular muscles, the conjunctiva, or the orbital structures.⁵ The afferent limb is trigeminal, and the efferent limb is vagal. Although

the oculocardiac reflex is commonly associated with bradycardia, virtually any dysrhythmia, including ventricular tachycardia and asystole, may be seen.^{6,7} Children have increased vagal tone and, therefore, are more apt to manifest this problem. If a dysrhythmia appears, the initial treatment is to ask the surgeon to cease the manipulation. At this point, the anesthesia provider should determine that the depth of anesthesia and ventilatory status are adequate. Commonly, the heart rate and rhythm return to baseline within 20 seconds. Usually, the reflex fatigues with repeated manipulations, and if it does occur again, it is not as dramatic. If the reflex becomes problematic, then atropine or glycopyrrolate should be given intravenously.

PREVENTION AND TREATMENT OF INTRAOPERATIVE AND POSTOPERATIVE PAIN

The most commonly used medications for intraoperative analgesia are acetaminophen, ketorolac, narcotics (morphine and fentanyl), and dexmedetomidine.^{8,9} In infants, an acetaminophen suppository (20–40 mg/kg) may be placed after induction. Once intravenous access is established, ketorolac (0.5 mg/kg) may be used. If necessary, narcotics (morphine 0.1 mg/kg or fentanyl 1–2 µg/kg) can also be administered in divided doses. Narcotics in very young infants are avoided if possible to prevent delayed emergence and postoperative respiratory depression. Likewise, dexmedetomidine is also avoided in this patient population to avoid delayed emergence.

In older patients, liquid acetaminophen (10–20 mg/kg) or ibuprofen (10 mg/kg) may be mixed with the midazolam preoperative medication. Alternatively, ketorolac may be given intravenously in lieu of ibuprofen. Ketorolac is a nonsteroidal anti-inflammatory drug that interferes with prostaglandin production by blocking the cyclooxygenase pathway. It is effective within approximately 45 minutes when administered intravenously and has an analgesic half-life of approximately 6 hours. Morphine and fentanyl may be added as needed for the surgical procedure. Dexmedetomidine may also be used in addition to the previously mentioned analgesics.¹⁰ As an α_2 -adrenoceptor agonist, it functions as an analgesic and anxiolytic without the respiratory depression of narcotics and is useful in the prevention and treatment of emergence delirium. In our institution, we often give older pediatric patients and teenagers an intraoperative dose of 0.25 µg/kg and if needed, additional divided doses up to 1 µg/kg in the postanesthesia care unit (PACU) to quell emergence delirium.

Excessive postoperative pain causes postoperative nausea, emotional distress, and delays in PACU discharge. Although narcotics have traditionally been used for the prevention and treatment of pain, they can cause

excessive sedation, respiratory depression, and nausea and vomiting. Judicious use of narcotics in combination with non-narcotic analgesics (acetaminophen, ketorolac, dexmedetomidine) may be used to prevent/treat pain while allowing expeditious discharge from the PACU.

EFFICIENCY ISSUES IN OPERATING ROOM TURNOVER

Multiple factors contribute to the efficiency of operating room turnover. Many people and departments are involved in the process. Ideally, the history, physical examinations, and informed consent should be done before the patient's arrival at the hospital for surgery. If there is a preoperative clinic, the anesthesia workup and any additional examinations/investigations may be completed prior to surgery. Incomplete paperwork can slow down an otherwise efficient process.

If the surgeon can give a 10-minute notification of completion of the surgical or examination process, then the anesthesia provider can get the patient back to spontaneous respirations, discontinue the air warming unit, tally the intravenous fluids, and administer any antiemetics not already given. This would also be the appropriate time frame in which to premedicate the next patient. If there are no anesthesia technicians working in the department, the anesthesia provider should prepare for the next patient by drawing up medications, preparing an ETT or LMA, and setting up intravenous fluids. Another major factor affecting turnover time involves the housekeeping department. By having enough housekeeping staff to expedite room cleaning, the operating room staff can quickly prepare the room for the next surgery. Also, having a second operating room in which to alternate cases is ideal in reducing if not eliminating turnover time. As the surgical procedure is ending in the first operating room, the anesthetist assigned to the second room may begin the induction of the next patient. While the surgeon speaks with the family members of the first patient, the second patient can be positioned and prepped for the upcoming operation.

In our institution, we utilize LMAs in patients over 1 year of age or 9 to 10 kg if a surgical procedure is involved. This allows us to remove the airway device earlier than an ETT, which can also reduce turnover time. In infants under 9 kg, we use an ETT to provide a secure airway. When an LMA is used, some ophthalmologists may prefer the flexible version because of the long malleable tube on the device. This allows the microscope to be closer to the field without interfering with airway management.

The need for a rapid operating room turnover time should never jeopardize patient safety. Patient safety is paramount. Sometimes a patient will require additional attention from the anesthesia provider in the PACU

following surgery. All patients should have stable vital signs, be comfortable, and be left with a competent RN to monitor their continuing emergence from the effects of general anesthesia.

PREVENTION AND TREATMENT OF POSTOPERATIVE NAUSEA AND VOMITING

The incidence of PONV over all patient populations is 10% to 20% and is influenced by many factors, including patient age, gender, surgical procedure, anesthetic technique, and underlying diseases.¹¹ Multiple factors increase the incidence of PONV in the ophthalmic patient, including an alteration in visual perception, anesthetic technique, and the oculocardiac reflex.³ The most common antiemetic used to prevent PONV is ondansetron. Ondansetron is a centrally acting serotonin antagonist at the 5-hydroxytryptamine₃ (5-HT₃) receptor. Effective intravenous doses are 0.1 to 0.15 mg/kg but not to exceed 4 mg.^{12,13} Its use is associated with minimal side effects, and it lacks the sedative and extrapyramidal side effects of other antiemetics such as droperidol and metoclopramide.¹⁴⁻¹⁷ Dexamethasone (0.25 mg/kg), a synthetic glucocorticosteroid, may also be used for its antiemetic effect.¹⁸ Droperidol is a butyrophenone that possesses antiemetic activity as a result of its central antagonism of dopamine receptors in the chemoreceptor trigger zone.¹⁴ Effective intravenous doses of droperidol range from 50 to 75 µg/kg. Potential side effects from the use of droperidol include sedation and delayed emergence from anesthesia, extrapyramidal side effects, restlessness, and anxiety.¹⁵ Metoclopramide, a benzamide, acts as a central dopaminergic antagonist and acts peripherally to increase lower esophageal sphincter tone and increase gastric motility.¹⁶ Conventional antiemetic doses range from 0.1 to 0.2 mg/kg. It may cause sedation and extrapyramidal side effects.

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14

Principles of Incision Construction, Location, and Closure in Pediatric Cataract Surgery

Rupal H. Trivedi and M. Edward Wilson

Principles of incision construction in pediatric cataract surgery generally follow the techniques and innovations developed for adult cataract surgery.¹ As in adults, pediatric cataract incisions are getting smaller. Intraocular lens (IOL) insertion devices are being constantly redesigned to allow the IOL to be inserted through ever-smaller openings. Bimanual surgical tools are now available in very small gauge sizes giving the surgeon more options than ever before. Unlike in adults, the wound is more likely to be traumatized by the child during the postoperative period. Children are more likely to rub the eyes and fight the installation of the postoperative drops.

The most immediate goal when planning the incision strategy is to aid intraoperative maneuvering. Consideration must be given to the intraocular maneuvers that are anticipated for the surgery being planned. Where do the incisions need to be made and how many incisions will be needed in order to easily accomplish the goals of surgery? Will membranes need to be cut? Will synechia need to be severed? Will a small pupil need to be enlarged? Will the lens equator need to be directly assessed from a specific angle? Are zonular fibers missing or loose in one quadrant or another? What IOL will be used and what size incision will be required to implant it? Will the incisions be sutured? Will the sutures need to be removed or will they be absorbable? Each of these considerations is important to the incision planning. Each of the 360 degrees of the eye's circumference is available for incision planning. Proper consideration and selection among all of the options helps make even the most difficult intraocular gymnastics more safe and effective.

Tunnel incisions have replaced limbal incisions made with corneoscleral scissors in children, just as in adults.² As incision construction has gotten better and surgical

entry wounds have gotten smaller, worries about induced astigmatism have diminished. However, surgical incisions can still create unwanted astigmatism and can also be used (in a limited way) to reduce preexisting astigmatism as astigmatism may complicate the management of amblyopia.³ These considerations are covered in more detail later in the chapter.

Before moving forward, it is important to remember the applied anatomy of the child's eye and how it is different from the adult eye in relation to the construction of the incision. The cornea in premature and full-term babies is thick and reaches adult levels within the first 2 to 4 years of life.⁴ The sclera has a low rigidity and is not as thick. The anterior chamber is shallower in infant eyes compared to adult eyes. It is now generally agreed that there is a higher prevalence of against-the-rule astigmatism during infancy, which decreases with increasing age, while the prevalence of with-the-rule astigmatism increases.^{3,5,6}

TUNNEL INCISION ARCHITECTURE

Location

Superior/Temporal/Meridian of Steepest Curvature

Most adult cataract surgeons have moved toward the temporal side as the "preferred location" for a cataract surgical incision. The temporal wound presents similar advantages in children as it does in adults. However, the superior approach allows the wound to be protected by the brow and Bell phenomenon in the trauma-prone childhood years. Both scleral and corneal tunnels can be easily made from a superior approach since children rarely have deep-set orbits or overhanging brows.

There is very little emphasis given in reducing, minimizing, or altering surgically induced astigmatism. An incision at the steepest meridian would help decrease surgically induced astigmatism.^{7,8} However, in eyes with pediatric cataracts, it may be difficult to accurately document the steep corneal axis. Refraction by retinoscopy is difficult when a cataract is present. The reliability of handheld autokeratometers, while reasonably good for corneal curvature measurement, may be dubious for the purpose of axis measurement.⁹ Corneal topography requires cooperation and fixation. Also, pediatric eyes undergo changes in astigmatism with growth. Since it is likely that children will wear glasses after cataract surgery to correct for residual refractive error or at least for reading, less emphasis has been given in children to surgical maneuvers designed to treat preexisting astigmatism.

Scleral/Corneal

Pros and cons of a corneal tunnel are listed in Table 14.1. Eyes operated on for cataracts during infancy are at increased risk for glaucoma. A scleral incision involving the conjunctiva increases the number of conjunctival fibroblasts and inflammatory cells. This may account for the increased risk of trabeculectomy failure.¹⁰ Today,

Table 14.1 CORNEAL TUNNEL INCISION

Pros

- Avoids conjunctival peritomy and subsequent cauterization
- Avoids occasional hyphema and conjunctival ballooning encountered with scleral tunnel
- Ease of intraoperative maneuvering
- Cosmetically better
- Future filtration surgery—better outcome with untouched conjunctiva¹⁰
- Indicated in eyes undergoing coagulant therapy and patients with insufficient platelet count
- May decrease early postoperative breakdown in blood–aqueous barrier¹¹
- May reduce the risk of inadvertent conjunctival implantation of viable tumor cells and may allow for direct inspection of the incision site for tumor recurrence¹²

Cons

- Higher rate of surgically induced astigmatism
- Poor stability, especially if larger incision required
- Increased risk of endophthalmitis if left unsutured^{13,14}
- Higher rate of endothelial cell loss¹⁵
- Because an avascular structure, healing possibly delayed compared to that with a vascular scleral tunnel

angle surgery or seton implantation would likely be chosen over a trabeculectomy in an aphakic or pseudophakic child, making the state of the conjunctiva less important. Our survey of preferences back in 2001 indicated that only 38% of American Society of Cataract and Refractive Surgery (ASCRS) respondents and 27% of American Association of Pediatric Ophthalmology and Strabismus (AAPOS) respondents preferred a corneal tunnel.¹⁶ It is likely that the use of corneal tunnels in children is now much higher. Over the last decade, the adult literature has contained many reports indicating that unsutured clear corneal incisions are a statistically significant risk factor for acute post–cataract surgery endophthalmitis when compared to scleral tunnel incisions.¹³ Most of the adult corneal tunnels were left unsutured, but pediatric corneal tunnels are nearly always sutured. The additional risk of endophthalmitis may diminish or disappear when suture closure is added to the corneal incision location.

When inserting a foldable IOL, we generally prefer a corneal tunnel in children. However, a scleral tunnel is still sometimes chosen in infants. Infant corneal tissue opacifies easily at the sight of the tunnel and can appear unsightly after healing.¹⁷ Scleral tunnels heal in a more invisible way in very young infants. We also use a scleral tunnel when it is not possible to implant a foldable lens and a rigid one has to be used; the increased width of the incision—generally between 5 and 7 mm—favors a scleral location. In addition, when we are not certain which IOL we are going to implant—foldable or polymethyl methacrylate (PMMA)—we prefer a scleral tunnel, as it is easy to enlarge if the need arises. This is mostly observed in eyes with traumatic cataract or in a secondary IOL when little capsular support is present. Some authors also recommend using a scleral tunnel when absorbable sutures are not available or not preferred.¹⁷ Nylon sutures are thus covered by conjunctiva and are placed further from the cornea. Exposed nylon sutures usually require removal, but when used in association with a scleral tunnel, removal is more easily avoided.

Shape of the Incision: Straight/Frown/Circumlimbal

Straight or circumlimbal incisions are widely used in children. All of the shapes listed above are acceptable and all have their proponents.

ARCHITECTURE OF A PARACENTESIS

Preparation of paracentesis incisions is very important in pediatric cataract surgery. If IOL implantation is not planned, the pediatric cataract can be aspirated through two paracentesis incisions. Paracentesis incisions are useful for

1. performing bimanual maneuvering (irrigation/cutting/aspiration);
2. injecting intraocular solutions and ophthalmic viscosurgical devices (OVDs)—without losing the chamber;
3. stabilizing the globe;
4. controlling the movements of instruments;
5. facilitating the implantation of the lens; and
6. facilitating the use of iris retractors if needed.

TECHNIQUE

Diamond/Steel Knife

Steel blades have been considered the standard instrument in the construction of incisions. Diamond knives have several advantages over steel knives. However, several disadvantages of diamond knives have led many physicians to use disposable steel knives instead (Table 14.2). The need for instruments with perfect cutting edges becomes more obvious when enlarging an incision (for IOL placement) in soft vitrectomized eyes.

Surgical Technique

Conjunctival Opening

For a scleral tunnel, a limbal conjunctival flap of approximately 3 clock hours is made with Westcott scissors and 0.3-mm toothed forceps. Vertical relaxing incisions can be placed in the conjunctiva and Tenon fascia, at both ends, if needed. The subtenon space is bluntly dissected with a scissors.

Cauterization

For scleral tunnel incision, after the conjunctival peritomy is performed, cauterization can be done for hemostasis. Mild bipolar cautery (using an eraser tip) is usually performed near the limbus. Posteriorly, however, heavier cautery may be needed when a large scleral tunnel is being made.¹ The large vessels emanating from the rectus mus-

cles and perforating the sclera between the muscle and the beginning of the tunnels can be cauterized directly if necessary so that the area of the scleral tunnel is dry.¹

Groove

A groove is made for scleral tunnels and can even be made for corneal tunnels if desired. The desired width (depends mainly on the type of IOL—rigid or flexible—and size of the optic) of the incision is marked (stamped at the tip with a marking pen) using a caliper: for a scleral tunnel, 1.5 mm from the insertion of the conjunctiva, and for a corneal tunnel—clear corneal, limbal, or sclerocorneal, depending on the preference of the surgeon. The globe is fixed with a forceps, and the sclera/cornea is cut perpendicularly to make a groove.

Dissection

Starting above the base of the groove and remaining at half of the scleral/corneal thickness with a bevel-up blade (crescent knife), the tunnel should be dissected. Right-to-left lateral movement should be performed while progressing forward. Superficial incisions tend to be fragile and may tear during the surgery. If the dissection is too deep, the anterior chamber may be entered prematurely. As the limbus is crossed, the plane of the dissection is slightly anterior so that the appropriate corneal plane is maintained. If the scleral dissection plane is maintained across the limbus, the anterior chamber will be entered or the internal corneal lip will be thin.

Anterior Chamber Entry

Anterior chamber entry is made at an angle of 45 degrees with a “dimple-down” maneuver. After entering the anterior chamber, the direction of the knife should be changed and moved to the iris plane to avoid injuring the iris or the capsule. Remember that the anterior chamber is shallower in infant eyes compared to adult eyes. OVD should be injected into anterior chamber.

Paracentesis

Paracentesis is performed in front of the limbal vascular arcade, about 70 degrees to the left of the primary incision (for the right-handed surgeon and vice versa). Two paracentesis incisions are sometimes made at the 2 o'clock and the 10 o'clock positions to help assist bimanual maneuvering. The paracentesis on the surgeon's dominant side can then be enlarged later for IOL entry using a keratome. As the corneal tissue is entered, it may prove useful to exert light counterpressure on the opposite side with forceps. The paracentesis should measure approximately 1 mm and run parallel to the iris plane, to facilitate sealing at the end of surgery. For instance, a microvitreo-retinal (MVR) blade with a 20-gauge (0.9-mm) opening that provides a precise incision for insertion of a 0.9-mm (20-gauge) cannula is ideal for a 20-gauge vitrector/

Table 14.2 PROS AND CONS OF DIAMOND KNIFE COMPARED TO METAL KNIFE

PROS AND CONS OF DIAMOND KNIFE COMPARED TO METAL KNIFE	
Pros	
Great cutting precision	
Less pressure required while cutting	
Less friction on tissue being cut	
Cutting edge remains sharp after repeated use	
Cons	
Expensive	
Extreme attention required for cleaning and repositioning by operating assistants	
Lower tendency to forgive a surgeon's mistake	

aspirator to enter the anterior chamber. A 20-gauge, blunt-tipped irrigating cannula can also be used through a separate MVR blade stab incision. If the instrument positions need to be reversed, the snug fit is maintained. We generally use a MVR blade that matches the gauge of the instruments being used. This gives a tight incision for the irrigation, aspiration, and cutter handpieces. A second sideport incision, as described above, is used, particularly when aspiration of the cortical material is performed with two separate handpieces. Although these incisions are referred to as “stab” incisions, they take on a tunnel shape. If a tunnel is not formed, leakage is immediate when the instruments are removed, even in older children. Floppy iris, low scleral rigidity, and increased intravitreal pressure can contribute to iris prolapse (Fig. 14.1). However, if the tunnel is too long, the instruments cannot be maneuvered in the anterior chamber well because of “oar locking.”

Enlargement of the Incision (Internal Entry) for IOL Implantation

The internal entry needs to be enlarged for smooth maneuvering in IOL implantation. The size of the internal entry selected depends on the type of IOL. Keratomes commonly used are from 2.2 to 2.75 mm.

Pars Plana Incision

When pars plana/plicata posterior capsulectomy and vitrectomy is planned, our current approach is to first remove OVD from anterior chamber using bimanual irrigation and aspiration. The corneal tunnel has a stitch already placed to hold it closed when the aspiration cannula is withdrawn. The irrigation cannula remains in the anterior chamber to keep the chamber full when the aspiration cannula is removed. The MVR blade is used to create pars plana/plicata stab wound, with a direction

slightly anterior to the center of the eye. This is done while the irrigation remains in the anterior chamber. The pars plana/plicata entry is created at a preplaced mark 2 mm posterior to the limbus in patients younger than 1 year old, 2.5 mm posterior in patients 1 to 4 years old, and 3 mm posterior in patients older than 4 years old. After the MVR blade has made the sclerotomy, the vitrector is inserted through the pars plana wound to create the posterior capsulectomy. It is important that the irrigation into the anterior chamber be continued throughout this step. From the beginning of the OVD removal to the completion of the posterior capsulectomy and vitrectomy, the irrigation cannula remains in place through the paracentesis. When the aspiration cannula is withdrawn after OVD removal, the Venturi-pump machine is switched from I/A mode to vitrectomy mode while the MVR is making the sclerotomy. Care must be taken to quickly restart the irrigation fluid on the machine as it is switched from I/A to vitrectomy.

Suturing

Unlike in adults, tunnel incisions do not often self-seal in children. Our survey indicated that only 20% of ASCRS respondees left both (tunnel and paracentesis) incisions unsutured. The figure for AAPOS respondees was 3%.¹⁶ It is important to suture the incision during the trauma-prone years of childhood (Fig. 14.2).^{18,19} The elasticity of the pediatric eye wall tends to promote leakage from corneal or scleral tunnel incisions that would not leak in adults.

1. Self-sealing wounds failed to remain watertight in children <11 years of age, especially when an anterior vitrectomy was combined with cataract extraction.¹⁸ Vitrectomy collapses the sclera, making the already relatively elastic sclera of children even less rigid. Low scleral rigidity results in fish-mouthing of the wound, leading to poor approximation of the internal corneal valve to the overlying stroma.

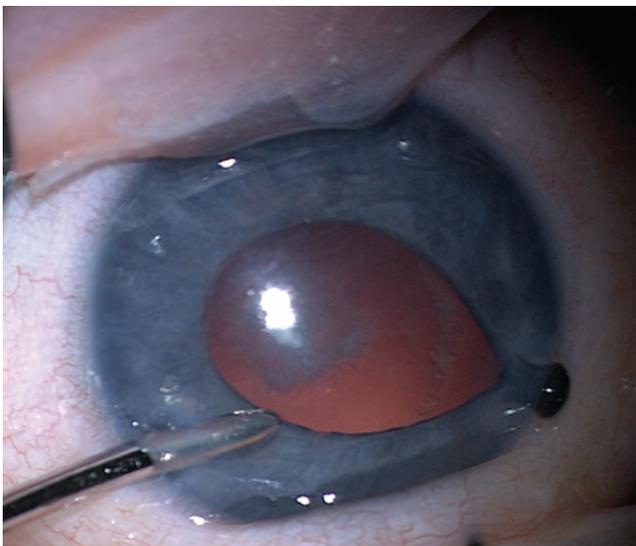


Figure 14.1. Iris prolapse in the right eye of a 4-week-old infant.



Figure 14.2. Ocular trauma in an eye 1 week postoperatively.

- Suture closure is usually needed since children rub or traumatize the eye after surgery more often than do adults.
- A higher dose of steroid drops is often used for cataract surgery in children compared to adults. This may be another reason to suture the wounds since the steroids can theoretically postpone the healing.
- It can be difficult to perform detailed examinations in the immediate postoperative period, making it difficult to identify a small leak, a slightly shallow anterior chamber, or a subnormal intraocular pressure. These are additional reasons to prefer the security of a well-sutured incision in children. A child with bilateral cataract often undergoes cataract surgery in the second eye within 1 to 7 days after the first eye surgery. Examination of the previously operated eye should be performed while child is under anesthesia for second eye surgery (Fig. 14.3).

Table 14.3 summarizes basic surgical principles for suturing the incision. We prefer synthetic absorbable 10-0 sutures rather than nonabsorbable sutures that would need to be removed later. Corneal vascularization requiring suture removal was noted in 18% of the eyes in a 10-0 polyester (Mersilene) group and none in a Vicryl group.²⁰ These authors also reported one case of endophthalmitis after suture removal.²⁰ The use of nonabsorbable sutures (e.g., nylon) calls for an examination under anesthesia (EUA) for suture removal with all the inherent risks of anesthesia. We use 10/0 Vicryl (polyglactin acid suture) absorbable suture (synthetic braided), which takes 60 to 90 days to completely absorb. In vivo half-life tensile strength of polyglactin is 2 weeks. Ethicon is also manufacturing Vicryl plus antibacterial suture, designed to reduce bacterial colonization on

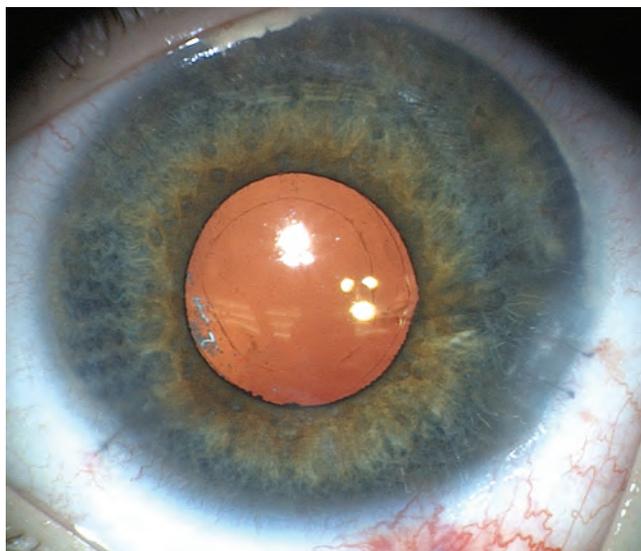


Figure 14.3. Two-day postoperative examination of the right eye of a 3-year-old boy. This photo was taken on a day of the left eye cataract surgery.

Table 14.3 BASIC GENERAL SURGICAL PRINCIPLES TO KEEP IN MIND WHILE PERFORMING SUTURING

Lower microscope magnification
Equidistant needle entries from wound edge
Uniform closure tension
Placed at same depth
Knots should be embedded or at least tucked snugly on the scleral side

the suture. However, according to the Ethicon Web site, it is not indicated for ophthalmic procedures (<http://www.ethicon360.com/products/coated-vicryl-polyglactin-910-suture>). Dr. Vanderveen prefers to use 10/0 polyglycolic acid suture instead (Sinusorb, braided and monofilament). It handles more like nylon and appears to be less inflammatory than Vicryl. The tensile strength is significantly reduced by 3 to 4 weeks, allowing retinoscopy and prescribing glasses (personal communication).

If a pars plana entry is created, we close the sclerotomy wound with an 8-0 Vicryl suture. A 9-0 or 10-0 Vicryl suture can be used for smaller-gauge sclerotomies. We generally close conjunctiva overlying the sclerotomy with a single buried stitch of the same material just used to close the sclerotomy. However, the conjunctiva can also be closed using cautery. For typical cases, no subconjunctival injections of antibiotics or steroids are needed.

SUMMARY

Adult cataract surgeons have developed wound construction techniques that require no or minimal suturing and induce little astigmatism in the immediate postoperative period. Although appealing, these techniques are not without added risk when applied to pediatric eyes, which are more elastic than adult eyes. A return to the operating room may be required to detect and treat a leaking wound. Surgeons desiring uncomplicated postoperative wound management may prefer to suture the wounds in children.

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Anti-Inflammatory Effect of Heparin in Pediatric Cataract Surgery

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Surgical management of pediatric cataract has shown marked development. The higher risk of postoperative complications after pediatric cataract surgery is attributed mainly to the greater inflammatory response after surgery. This reaction is associated with younger age and may be influenced by the surgical technique, intraoperative injury to adjacent structures such as the iris, and presence of antecedent ocular infection (e.g., rubella). Although cataract surgical techniques have seen a plethora of improvements, postoperative inflammation continues to be a significant factor compromising the outcome of pediatric cataract surgery.¹⁻⁴ Certain discernible signs of intraocular inflammation are increased number of cells and flare, inflammatory precipitates on the intraocular lens (IOL), formation of synechia, and inflammatory cyclitic membranes. Some authors have also suggested that increased postoperative inflammation may contribute to posterior capsule opacification (PCO).⁴ Therefore, techniques that might prevent or at least lower the inflammatory response may prove to be beneficial.

HEPARIN AND ITS ANTI-INFLAMMATORY PROPERTIES

Proteoglycan heparin is a long-chain polysaccharide composed of repeating units of hexosamine and either iduronic acid or glucuronic acid. It is an intracellular component of mast cells.

The effects of heparin as an anticoagulative agent are well known. It acts indirectly at multiple sites in both the intrinsic and extrinsic blood-clotting systems, where it potentiates the inhibitory action of antithrombin II on several activated coagulation factors, such as IX, XI, and XII. Through these mechanisms, heparin acts to inhibit formation of a fibrin scaffold. Storage of heparin in mast cell granules, which are released by several inflammatory effect or mechanisms, suggests that endogenous heparin has a role in inflammation.⁵ Experimental studies have

demonstrated anti-inflammatory and antiproliferative properties of heparin.⁵⁻⁷ It may decrease the postoperative intraocular inflammatory response by deactivation of the coagulation pathways and the formation of fibrin. Other mechanisms through which heparin may inhibit inflammation include inhibition of fibroblast proliferation,⁶ interference with lymphocyte recirculation,⁷ induction of apoptosis in human peripheral blood neutrophils,⁸ and inhibition of complementary activation.

The use of heparin in ophthalmic surgery was first described by Johnson and colleagues,⁹ who found that supplementing heparin in the infusion solution or giving a single intravenous injection resulted in a significant reduction in postoperative intraocular fibrin formation after vitrectomy and cyclocryotherapy in rabbits. Subsequently, experimental studies^{10,11} showed that infusion of heparin or low molecular weight heparin inhibits the formation of fibrin during lensectomy and vitrectomy. The anti-inflammatory properties of heparin have been used during or following cataract surgery in the following forms:

1. Surface coating of IOLs with heparin—Heparin Surface-modified IOLs:

With polymethyl methacrylate (PMMA) IOLs, there were concerns of a greater inflammatory response, particularly in children. Heparin surface-modified (HSM) IOLs were designed to improve the biocompatibility of IOLs. It has been suggested that an HSM IOL may reduce the incidence of inflammatory precipitates on its surface.¹² The use of HSM IOL in conjunction with posterior capsulorhexis and intracamerular heparin decreased PCO in pediatric cataract surgery without significant side effects.¹³ However, heparin-coated IOLs did not gain widespread popularity because of their cost and the need for a large incision. Subsequently, PMMA was replaced by newer IOL materials with enhanced biocompatibility. The current day foldable acrylic lenses might result in less

IOL precipitation as a result of lens hydrophobicity and smaller surgical wound.¹⁴⁻¹⁶

2. Heparin drops:

Topical administration of heparin drops following cataract surgery has also been tried, and there are very few reports that suggest that this can diminish the occurrence of precipitates on PMMA IOLs.¹⁷ As a result, this modality has not gained popularity.

3. Heparin added to the infusion solution:

Studies carried out on animal eyes revealed that supplementing the irrigating solution with heparin caused a lower level of disturbance in the blood-aqueous barrier.^{18,19} Subsequently, studies carried out in adult eyes^{20,21} reported that infusion of heparin may reduce the postoperative inflammatory reaction following small incision cataract surgery with IOL implantation. The use of heparin during pediatric cataract surgery has also been reported²²⁻²⁶ and is discussed in more detail below.

Potential Concerns with Intraocular Use of Heparin

Although intraocular bleeding is a potential risk of heparin supplementation, bleeding of iris or intraocular structures was not reported^{20,21} in standard cataract procedures involving both scleral and clear corneal tunnel incisions.

Low Molecular Weight Heparin

In the last few years, low molecular weight heparin derivatives have increasingly been used for systemic administration. Enoxaparin, a low molecular weight heparin (molecular weight 4,500 Dalton),^{11,12} is most frequently used as a parenteral injection to prevent deep venous thrombosis after a major surgery. Enoxaparin has anti-inflammatory mechanisms similar to heparin but poses a lower risk of incurring complications such as major bleeding. In a nonrandomized, unmasked series,²⁴ authors showed that intraoperative irrigation of enoxaparin was effective in reducing postoperative inflammation following pediatric cataract surgery. Subsequently, a randomized clinical trial of an intraocular infusion of enoxaparin in children after cataract surgery with IOL implantation was conducted (see below).

Results of a Randomized Clinical Trial Evaluating Anti-Inflammatory Effect of Low Molecular Weight Heparin in Pediatric Cataract Surgery

At our center, we conducted a randomized, controlled, clinical trial to evaluate whether an intraocular infusion of enoxaparin helps in reducing the inflammatory response following bilateral cataract surgery and IOL implantation in children. The study population comprised 20 children between birth and 15 years of age.²⁶

Clinical Evaluation and Procedures

A thorough preoperative evaluation was performed, under anesthesia, when required. Patients were randomized to either receive 40 mg/500 mL enoxaparin in the intraocular infusion fluid (Group I) or not to receive enoxaparin—(Group II). A single surgeon carried out all the surgical procedures using a standardized technique. Posterior continuous curvilinear capsulorhexis was performed in children below 6 years of age and anterior vitrectomy in children below 2 years of age. A single-piece hydrophobic acrylic aspheric IOL (SN60WF—Alcon laboratories, USA) was implanted in the capsular bag in all eyes. The postoperative treatment regime was also standardized for both groups. The follow-up visits were in the first week and subsequently at 1 and 3 months. Postoperative assessment included anterior chamber inflammation (cells and flare), cell deposits on IOL (small and large), as well as the presence or absence of posterior synechia.

Results

The mean age at the time of cataract surgery was 40.8 ± 24.8 months. None of the eyes in either group developed intraoperative or postoperative hyphema, subconjunctival hemorrhage, or intraocular hemorrhage. Anterior chamber cells and flare of more than grade 2 were not detected in any eye at any follow-up. In the first postoperative week, anterior chamber cells rated at grade 2 were more common in the no-enoxaparin group compared to when enoxaparin was used (80% versus 40%, $P = 0.009$). However, at 3 months after surgery, there were no detectable cells and flare in any of the eyes. Fibrinous exudates or inflammatory membranes were not detected in any of the eyes.

In the first week, a higher proportion of eyes in the enoxaparin group (20%) had >10 small cells on the IOL surface when compared with eyes in the no-enoxaparin group (0) ($P = 0.005$). However, at the ensuing follow-up visits, no detectable difference was noted between the two groups. Similarly, the number of large cell deposits on the IOL surface was comparable between the two groups at all the follow-up visits. At 1 month postoperatively, posterior synechia were seen in two (10%) eyes when enoxaparin was used versus none when enoxaparin was not used ($P = 0.14$).

Our prospective, randomized study failed to show a significant anti-inflammatory benefit from the addition of low molecular weight heparin to the infusion in pediatric eyes undergoing bilateral cataract surgery with IOL implantation. However, we did not have any cases in our series with fibrinous exudates or inflammatory membranes, even in the group where enoxaparin was not used. Various factors such as the use of the closed chamber technique, modest bottle height and aspiration flow rate, high molecular weight ophthalmic viscosurgical devices, minimal tissue

injury, thorough cortical cleanup, hydrophobic acrylic in-the-bag IOL implantation, and surgeon's experience may be responsible for reducing postoperative inflammation even in these young eyes. The role of heparin and similar adjuncts need to be further evaluated in those eyes that are prone to higher inflammatory response, such as eyes with coexisting uveitis and in eyes undergoing cataract surgery during early infancy. Although we intended to include children from birth to 15 years of age, our study did not have any children younger than 1 year of age.

SUMMARY

Perhaps, the advent of newer techniques, cutting-edge technology and superior IOL materials may have reduced the occurrence of postoperative inflammation in these young eyes without the need for any adjunctive agents. While not needed in every case, intraocular heparin may be beneficial in rare cases when a marked inflammatory response is expected. The application of intraocular heparin needs to be further studied in those special circumstances.

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Ophthalmic Viscosurgical Devices

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The aim of surgery using *viscoelastic agents*, or *viscosurgery*, is to provide greater protection to intraocular tissue from mechanical damage, while increasing the space available for surgical manipulation. The use of viscoelastic substances has become an integral part of ophthalmic surgery, particularly anterior segment surgery. Although the term *viscoelastics* has made its way into the ophthalmic lexicon, viscous products are not always elastic. The term *ophthalmic viscosurgical devices* (OVDs) was introduced in 2000.¹ OVD more clearly implies the intended surgical role of these substances, which is viscosurgery. In common clinical conversations, “viscoelastic” and “OVD” terms are used interchangeably. For published medical literature, most editors will require that the OVD term be used. OVDs are classified as devices because they remain in an unaltered state when removed from the eye after intraocular usage. Many OVDs are now available, each possessing specific chemical and physical properties and leading to different intraoperative behaviors. Better understanding of this advancing area of technology can help pediatric cataract surgeons optimize the use of these devices to improve outcomes.

A GLOSSARY OF OVD TERMS AND PROPERTIES

Viscosity is defined as the internal friction caused by molecular attraction that leads to a solution’s resistance to flow. Viscosity denotes the protective and lubricating property of the material. Increasing either the concentration or the molecular weight of a solution can increase the viscosity and make the material more resistant to flow. Viscosity depends on the degree of molecular movement within a solution (also known as the *shear rate*), and it varies inversely with temperature. This is the reason why it is essential to state the temperature at which viscosity is being measured. Fluids that have the same viscosity at low shear rates and at high shear rates (i.e., when the viscosity is independent of shear rates) are referred to as *newtonian fluids*. Chondroitin sulfate is such an example.

Fluids exhibiting a decrease in viscosity at high shear rates are referred to as *non-newtonian fluids*. Sodium hyaluronate (Na–Ha) is one such example. A fluid that is highly viscous will require greater force for its injection into the eye than would a less viscous fluid injected through a same-sized cannula, and the greater the force required, the less able the surgeon is to judge either the required force or the sufficiency of the injection.²

Very low shear rates are present when the ocular structures are stationary and an OVD is simply expanding space during the surgical procedure. *Zero-shear viscosity* correlates with the molecular weight of a rheologically active OVD and can be used to rank and classify these agents. *High viscosity at low shear rates* maintains space and protects intraocular tissues.

Medium shear rates correspond to the velocities at which the surgeon moves objects through the eye, such as during an intraocular lens (IOL) implantation. *Moderate viscosity at medium shear rates* allows movement of surgical instruments, thus assisting IOL implantation.

Very high shear rates are operative when an OVD is being injected through a cannula. *Low viscosity at high shear rates* allows for easy introduction into the eye through a small cannula. *Elasticity* is the tendency of a material to return to its original size and shape after having been deformed (i.e., stretched or compressed). Long-chain molecules such as Na–Ha tend to be more elastic than do short-chain molecules.

Plasticity can be defined as the initial resistance to flow. The resistance decreases once a fluid begins to move. The more plastic a substance, the less force is required to initiate this movement.

Pseudoplasticity refers to a solution’s ability to transform when under pressure from a gel-like state to a more liquid state. Pseudoplastic fluids, which all of our OVDs are, exhibit decreasing viscosity as the external stress is increased but, unlike plastics, possess a limiting viscosity as the stress is reduced, and always remain fluid.²

Cohesiveness is the degree to which a material adheres to itself. It is a function of molecular weight and elasticity.

Long-stranded OVDs with a high molecular weight become entangled and tend to remain as a single mass. To date, all cohesive OVDs are of higher zero-shear viscosity, and all dispersive OVDs are of lesser zero-shear viscosity.² *Superviscous cohesive* OVDs have zero-shear viscosities exceeding 1,000,000 mPs (millipascal seconds), while *viscous cohesive* agents have zero-shear viscosities between 100,000 and 1,000,000 mPs. Cohesion is actually more complicated than this, and all OVDs are really cohesive at rest, but lower molecular weight OVDs behave in a dispersive fashion under the low vacuum stress imposed by irrigation and aspiration encountered during cataract surgery. Generally, given two viscoelastic substances of the same chemical family and concentration, the greater the mean molecular weight (i.e., the greater the polymeric chain length) of a substance, the greater its cohesion and zero-shear viscosity.²

Dispersiveness is the tendency of a material to disperse when it is injected into the anterior chamber. *Dispersive* agents commonly have low molecular weights and shorter molecular chains. In agents that have lower zero-shear viscosities, molecular chain entanglements become far less important and cohesion tends to be significantly weaker, resulting in a tendency to disperse when injected into the anterior chamber. *Medium-viscosity dispersive* OVDs possess zero-shear viscosities between 10,000 and 100,000 mPs. *Very-low-viscosity dispersive* agents include all of the unmodified hydroxypropyl methylcellulose (HPMC) agents.

Coatability refers to the ability of an OVD to adhere to the surface of tissues, instruments, and implants. A lower surface tension and a lower contact angle indicate a better ability to coat. In addition, the molecular charge of the viscoelastic substance may influence its coating ability.

Arshinoff described desirable characteristics of an OVD as (1) low viscosity during its injection into the operative space, to facilitate rapid movement through a small-bore cannula while preserving the surgeon's tactile feedback sensitivity; (2) high viscosity when stationary, to create and maintain surgical spaces; and (3) intermediate viscosity at intermediate shear rates, to allow the passage of an IOL or a surgical instrument as required during the operation.²

AVAILABLE MATERIALS

Balanced salt solutions (BSSs) and *air*, while not technically viscosurgical materials, were the first substances used as protective agents. Both are readily lost once the cornea is retracted or during difficult surgical manipulations in the anterior chamber.

HPMC is synthesized from methylcellulose as a raw wood pulp product of medical grade. Methylcellulose, in a 1% solution, was used to coat IOLs prior to implantation, and later 2% methylcellulose was used to

maintain the anterior chamber.³ HPMC (2%) consists of a long chain of glucose molecules with replacement of the hydroxy groups by methoxypropyl and hydroxypropyl side chains. This polymeric backbone is cellulose, a carbohydrate that is not a natural component of animals or humans, and so its fate in the eye remains unknown. The physical properties of HPMC require that a large-bore cannula with increased infusion pressure be used for injection. It is relatively difficult to completely remove HPMC from the eye. The primary advantages of HPMC are its ability to coat, availability, ease of preparation, room temperature storage, ability to withstand autoclaving, and low cost compared with other OVDs.⁴ Its safety and efficacy in intraocular surgery have been reported.^{3,5} Cellugel, OcuCoat, Visilon, and I-Cel are some of the commercially available HPMC OVDs.

Sodium hyaluronate (Na-Ha), a viscous substance, was used in animal implant experiments as early as 1977 and in human implant experiments beginning in 1979.⁶ It is a naturally occurring lubricant and shock absorber present in nearly all vertebrate connective tissue matrices. In the eye, Na-Ha is found at high concentrations in the vitreous and connective tissue of the trabecular angle and at low concentrations in the aqueous humor and covering the corneal endothelium.⁷ Importantly, the use of this product in surgical situations does not represent the introduction of a "foreign" material. All Na-Ha products require refrigeration, with subsequent acclimation to room temperature prior to use. The prime advantages of Na-Ha products are its creation and maintenance of space in the anterior chamber, its ease of insertion and removal, and the fact that it is a natural product of the eye. The disadvantages are its poor coating ability, its removal as a mass during high-turbulence situations, and the necessity to refrigerate it. Manufacturers emphasize the importance of the product's purity, with various proprietary methods used to ensure this quality. It has been extracted from a variety of sources, including the dermis of rooster combs, umbilical cords, and cultures of streptococci.⁴ Although highly purified Na-Ha from each of these sources has the same structure, the molecular weight can vary. Healon, Healon GV, Healon5, Amvisc, Amvisc Plus, and Biolon are some of the available Na-Ha products.

Chondroitin sulfate at low concentrations is useful for coating tissue but poor for maintaining space because of its low viscosity. Viscoat is a 1:3 mixture of 4% chondroitin sulfate and 3% Na-Ha. The Na-Ha in Viscoat is produced by bacterial fermentation through genetic engineering techniques, and the chondroitin sulfate is obtained from shark fin cartilage. The combination of two biologic polymers creates a unique chemical structure with a relatively high viscosity and perhaps increases its coating ability and cell protection, because of the additional presence of a negative charge. Viscoat requires refrigeration, with subsequent acclimation to room

temperature prior to use. Ocugel is a combination of chondroitin sulfate and HPMC.

*Polyacrylamide*⁸ and *collagen*⁹ are also described in the literature as OVDs.

HIGH-VISCOSITY COHESIVE VISCOELASTIC AGENTS

Cohesive viscoelastic agents with high zero-shear viscosities are better for creating space compared to dispersive OVDs. This is especially important when expanding a shallow anterior chamber in an infantile eye. They are also useful when it is desirable to pressurize the anterior chamber to a level equal to the posterior pressure. This pressure equalization during cataract surgery is especially helpful for capsulorhexis, because it flattens the lens capsule. Cohesive OVDs can also be used to enlarge a small pupil, to dissect adhesions, and to aid IOL implantation. The high cohesiveness of viscous and superviscous material results in easy removal at the end of the surgical procedure. However, because of this same cohesive behavior, these agents also rapidly leave the anterior chamber during surgery.

Some surgeons find that the extremely high zero-shear viscosity of superviscous cohesive materials make them initially somewhat difficult to work with. When using these agents, surgeons should be more precise in their movements, as things moved into the wrong place tend to stay there (e.g., folds of the capsular flap when creating a capsulorhexis). However, with practice, the benefits of these materials quickly become apparent to the surgeon, and many have come to prefer them as their primary viscoelastic agents in pediatric cataract surgery, unless a dispersive material is surgically indicated.

Superviscous cohesive agents include I-Visc Plus (I-MED Pharma), Healon GV (AMO), and Eyefill C. (Croma Pharma). Viscous cohesive includes I-Visc (I-MED Pharma), Healon (AMO), Provisc (Alcon), Amvisc Plus (B & L), Amvisc (B & L).

LOWER-VISCOSITY DISPERSIVE VISCOELASTIC AGENTS

The most useful properties of dispersive OVDs are their resistance to aspiration and their ability to partition spaces. Their dispersive nature, a negative electrical charge, and the presence of hyaluronic acid that can bind to specific binding sites on the corneal endothelium improves the retention of these agents within the anterior chamber throughout the surgery. Thus, these agents are capable of partitioning the anterior chamber into an OVD-occupied space and a surgical zone in which irrigation/aspiration can be continued, without the two areas mixing. This is referred to as surgical

compartmentalization. Therefore, their use is even more beneficial in eyes in which a compromised endothelium is suspected. Dispersive OVDs can also selectively move or isolate a single intraocular structure within the anterior chamber (e.g., holding back vitreous at an area of zonule disinsertion or at a small hole in the posterior capsule).

Prevention of posterior capsule opacification remains an important goal in cataract surgery, especially pediatric cataract surgery. Posterior capsule opacification is caused mainly by the proliferation of lens epithelial cells. Budo et al.¹⁰ investigated the morphologic effects of Viscoat on lens epithelial cells. They conclude that light microscopy and transmission electron microscopy of human lens capsule suggest that Viscoat induces significant morphologic changes in lens epithelial cells during cataract surgery. The changes may underlie the improved visualization of these cells that has been reported during cataract surgery. Studies in a rabbit model suggest that the hyperosmolarity of Viscoat may play a partial role in the lens epithelial cell changes.

The major drawback of lower-viscosity dispersive OVDs is their relatively low viscosity and elasticity, which do not allow them to maintain or stabilize spaces as well as higher-viscosity cohesive OVDs (e.g., in the performance of capsulorhexis or IOL implantation). They tend to be aspirated in small fragments during irrigation/aspiration, which leads to an irregular viscoelastic–aqueous interface that partially obscures the surgeon's view of the posterior capsule during surgery. The microbubbles that can form during surgery also tend to become trapped at this irregular interface, further obscuring visibility and making surgical maneuvers in the posterior chamber even more difficult. Because of low cohesion, lower-viscosity dispersive OVDs are more difficult to remove at the end of surgery.

As mentioned earlier, cohesive agents are best at creating and preserving space, while lower-viscosity dispersive agents are retained better in the anterior chamber and are capable of partitioning spaces. High zero-shear viscosity agents tend to leave the anterior chamber rapidly during the turbulent surgical outflow. Although zero-shear low-viscosity OVDs remain in the anterior chamber during surgery, they are not of use in pressure-equalized surgery, and they are more difficult to remove. The so-called *soft-shell technique* has been described in the literature; it maximizes the advantages and minimizes the disadvantages of both cohesive and dispersive OVDs by using them together.¹¹ Healon5 is a product attempting to combine the best of the cohesive and dispersive agents into a single agent.^{12,13}

The only viscous dispersive agent available is DisCo-Visc. It has nearly the same zero-shear viscosity as the viscous cohesive described above. Lower-viscosity dispersives available are Viscoat and Cellugel, and very-low-viscosity dispersives are Ocucoat (B & L), Visilon

(Shah & Shah), and I-Cel (I-MED Pharma) among others. Duovisc is a combined packet of Viscoat and Provisc.

SOFT-SHELL TECHNIQUE

A lower-viscosity dispersive agent is first injected into the anterior chamber. Then a high-viscosity cohesive agent is injected into the posterior center of the dispersive agent. This combination of agents gives corneal endothelial cell protection throughout most of the lens removal, since the dispersive agent retains its attachment to the cornea even at higher turbulence.¹¹

After the lens substance has been removed, these same two OVDs are injected, but in a reversed manner. That is, the high-viscosity cohesive agent is injected first, and then the lower-viscosity dispersive agent is injected into the center of the cohesive agent. This allows free movement of the incoming IOL through the dispersive agent, with better stabilization of the surrounding iris and capsular bag by the higher-viscosity cohesive agent. Removal of OVDs at the end of surgery can be easily accomplished, since the lower-viscosity dispersive OVD (more difficult to remove) can be aspirated from the central anterior segment first, followed by the easier-to-remove high-viscosity cohesive agent.

VISCOADAPTIVE AGENTS

Healon5 is the first OVD to be referred to as *viscoadaptive*, denoting its adaptability to the surgical environment. This adaptability was accomplished by modifying the Na–Ha molecule to a log unit higher viscosity than most viscous products available to date. Theoretically, such a cohesive substance will become dispersive by fracturing under turbulent energetic conditions while, at the same time, remaining as a shield to protect the corneal endothelium. This ability to alter its viscosity during various manipulations is an important property of viscoadaptive agents. Healon5 has a molecular weight of 4 million daltons, which is similar to that of Healon. However, the concentration of Na–Ha in Healon5 is 23 mg/mL, compared to 14 mg/mL in Healon GV and 12 mg/mL in Healon. Healon5 has a higher viscosity at the zero-shear rate than any currently available OVD. In the midshear range (e.g., when being manipulated with instruments), it behaves very similarly to Healon GV. At high shear rates (e.g., during injection), it flows very easily through a 25- to 27-gauge cannula, thus allowing surgeons to use their fingertips to sense the inflation pressure in the eye, rather than the resistance in the cannula.

The rheologic features of Healon5 are both cohesive and dispersive. At low shear rates, it has the properties of a cohesive viscoelastic, while at high shear rates, because it can be fractured, it acts like a dispersive agent. Thus, the agent can adapt to different surgical needs and tur-

bulence. The adaptive character of Healon5 may be the result of its molecular structure. At low turbulence, long molecular chains become entangled and maintain space (as cohesive OVDs do). Thus, Healon5 stays in the anterior chamber during capsulorhexis (a nonturbulent situation). During lens substance removal, and the accompanying increase in turbulence at that time, the agent fractures into smaller pieces and starts behaving as a dispersive OVD. During lens substance removal, Healon5 partially remains in the anterior chamber, whereas a cohesive OVD such as Healon GV has a tendency to leave the eye immediately. The cohesive qualities of Healon5 allow expansion of the operating space and protection of the intraocular tissue, yet its dispersive qualities allow surgeons to easily remove the product upon completion of the procedure. During IOL implantation, Healon5 minimizes the problems that can be caused by positive vitreous pressure. It is easy to inject, like a cohesive agent, and it remains in the anterior chamber during surgery, like a dispersive agent. It flattens the anterior lens capsule for controlled tearing, and it reduces the risk of peripheral extension. However, like any other new aid, it has its own learning curve. Other available viscoadaptive OVDs are IVisc Phaco and BD MultiVisc.

*Ultimate soft-shell technique*²: In this technique, the anterior chamber is first filled (about 70%) with viscoadaptive through the main incision. Make sure to block the incision by injecting OVD as the cannula is withdrawn. BSS is then injected through the same incision, with the tip of the cannula remote from the incision and slightly indenting the anterior lens capsule, until the eye begins to become firm. The capsulorhexis is performed with the resistance of water, but the anterior chamber is pressurized as if only viscoadaptive was being used. After completion of the capsulorhexis, the BSS cannula is again introduced, but this time wiggled as the eye is entered, slowly injecting BSS all the time, which breaks out the small piece of OVD, permitting free circulation of BSS under the viscoadaptive protective dome. When capsular dye is used, it is gently painted over the capsular surface, before the BSS injection step. Very little trypan blue is required, and the subsequent injection of the BSS visually clears the field, making capsulorhexis easy in these otherwise difficult cases. Before IOL implantation, viscoadaptive OVD is injected into the eye, across the capsulorhexis, not into it. When the anterior chamber begins to fill, OVD is seen to begin to enter the capsular bag. At that point, injection stops, and the BSS cannula is retrieved. BSS is injected into the capsular bag, by placing the tip of the cannula under the capsulorhexis edge, in the same manner as is done with hydrodissection. As BSS fills the capsular bag, the bag is seen to distend, and the OVD moves upward out of the bag toward the incision. As the eye begins to become firm, stop injecting. When an injector is used, the leading IOL haptic is observed to open as it enters the BSS-filled capsular bag. The trailing haptic,

however, remains folded, as it remains in viscoadaptive OVD. When the I/A instruments are reinserted to begin OVD removal, the trailing haptic is observed to fall into the bag and open as soon as irrigation is begun, thus pressurizing the eye.² All of the viscoadaptive is now in front of the IOL and can be removed with the so-called “Rock ‘n’ Roll” technique.²

APPLIED ANATOMY—USING OVDs IN PEDIATRIC SURGERY

With this understanding of the physiochemical characteristics of OVDs in mind, a discussion of how children’s eyes differ anatomically from adult eyes and the role of specific OVDs at different stages of pediatric cataract surgery follows.

Small Pupil

In the newborn or infant, the pupils are miotic and remain that way through the 1st year of life. The pupil also often dilates poorly in infantile eyes. At birth, the dilator muscle is poorly developed, which explains the difficulty in obtaining good mydriasis with sympathomimetic agents.¹⁴ Achieving adequate mydriasis is generally difficult in infants particularly in children <6 months of age due to iris hypoplasia. Superviscous and viscous cohesive OVDs can help to dilate the pupil as an adjunct to mydriatic agents. Jhanji et al.¹⁵ reported the use of Healon5 to manage intraoperative miosis during pediatric cataract surgery.

Corneal Endothelium

An obligatory level of endothelial cell loss occurs during growth and with aging.¹⁶ In addition, operated eyes continue to lose endothelial cells at a higher rate (2.5% per year) for several years after cataract surgery.¹⁷ Since the eyes of a child will need to function for many more years after surgery compared to those of an adult, it is very important to protect the corneal endothelium in these small eyes. OVDs play an important role in this regard.

Shallow Anterior Chamber

The depth of the anterior chamber is at its minimum in the newborn eye. The anterior chamber depth shows a rapid increase early in life, reaching its final adult depth at between 8 and 12 years. However, the depth will decrease in the presence of advanced intumescent infantile or childhood cataracts. OVDs can deepen the anterior chamber in these small eyes, allowing safer surgical maneuvering within the anterior segment.

Anterior Lens Capsule

The pediatric anterior lens capsule is more elastic than that in adults. The anterior lens capsule is thin in

pediatric eyes, which also adds to the difficulty encountered during capsulorhexis. Capsulorhexis is made easier by using OVDs to fill the anterior lens capsule and flatten the anterior lens capsule surface. Vitreous upthrust from low scleral rigidity promotes a taut anterior lens capsule surface. This taut surface encourages radial tearing and the so-called *runaway rbexis*.¹⁸ OVDs can flatten the anterior lens capsule surface and create a less taut surface for a more controlled capsulorhexis.

High-viscosity viscoelastic is most useful during manual capsulorhexis for pediatric cataract surgery (Fig. 16.1). In general, we do not need any OVD prior to the vitrectorhexis technique. However, if the anterior chamber collapses (due to aqueous outflow, subsequent to entry in the anterior chamber), we use a high-viscosity OVD to maintain the anterior chamber depth. Jeng et al.¹⁹ reported that the Healon5 may improve the continuous curvilinear capsulorhexis (CCC) completion rate in children. A round CCC was successfully performed in 7/15 (46.7%) eyes using Healon or Viscoat and in 9/10 eyes (90%) using the Healon5.¹⁹ The mean age of patients in the Healon/Viscoat group was 7.6 years and in the Healon5 group was 6.4 years ($P = 0.23$).¹⁹ Gibbon and Quinn reported that manual CCC was performed successfully without radial tears or the need to convert to an alternative capsulotomy method in 21/22 (95%) eyes (median age 9 months).²⁰

Low Scleral Rigidity

Low scleral rigidity increases fluctuations of the anterior chamber during high surgical turbulence. Maintaining a tight fit when instruments enter the eye, as well as using OVDs, promotes anterior chamber stability and avoids collapse during surgical manipulation.

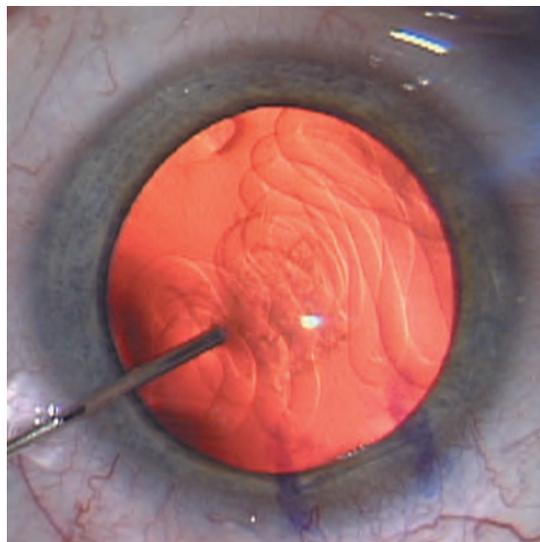


Figure 16.1. Injection of a high-viscosity viscoelastic before anterior capsulorhexis.

Posterior Capsule

Management of the posterior capsule is very important in children.²¹ Integrity of the posterior capsule opening is essential in long-term stability of the IOL. OVDs can stabilize the posterior chamber and push back the vitreous face during posterior capsulorhexis. Dholakia et al.²² reported completion rate of posterior continuous curvilinear capsulorhexis (PCCC) and the frequency of vitreous disturbance during pediatric cataract surgery.²² PCCC was performed under high-viscosity sodium hyaluronate (Healon GV). The PCCC was initiated with a 26-gauge cystitome and completed with Kraff-Utrata forceps by frequent grasping and regrasping the flap. Additional OVD was injected in the area surrounding the puncture to make the posterior capsule flat or concave. Authors suggested that OVD should not be injected through the puncture toward the vitreous face. Excessive injection through the central puncture may make the posterior capsule convex, and this can direct the tear peripherally. Arshinoff has recommended that posterior capsulorhexis be completed under OVD, using the soft-shell technique, by first covering the area with lower-viscosity dispersive OVD and then filling the anterior chamber with a higher-viscosity cohesive OVD without pressure (pressure could extend the tear).²

Increased Intraocular Pressure

Lifelong changes are also a characteristic of the vitreous. Although the vitreous probably has a very small liquid component at birth, the liquid fraction is measurable by the age of 3 years and increases steadily in volume thereafter, eventually making up just over half of the vitreous volume. As the liquid fraction increases, the collagens in the gel apparently aggregate. This solid vitreous, combined with the low scleral rigidity, increases the vitreous pressure and adds to the difficulties encountered during anterior capsulorhexis, posterior capsulorhexis, and also IOL implantation. OVDs allow surgeons to neutralize the posterior vitreous pressure during pediatric cataract surgery.

SPECIAL CONSIDERATIONS

Poor Corneal Visibility

Poor corneal visibility during surgery is more common in pediatric cataract surgery than in its adult counterpart. Often in young children, a complete examination under anesthesia, as well as keratometry and “A-scan” ultrasound, is performed before the surgery begins. Corneal drying and epithelial damage can occur during these examinations. Exposure keratopathy during the induction of general anesthesia may aggravate this situation. Visualization through the cornea may deteriorate further as the surgery progresses. Sometimes it is helpful

to cover the cornea with dispersive lubricating OVDs. The lubricating effect of the OVD helps to improve clarity by maintaining corneal moisture throughout the surgery.

Hydrodissection

Hydrodissection is often performed in older children just prior to lens aspiration. Care should be taken not to inject too much of the highly viscous OVD before hydrodissection. The OVD may not evacuate from the anterior chamber during hydrodissection. The injected fluid, combined with an already overfilled anterior chamber, can markedly raise the intraocular pressure and increase the risk of posterior capsule rupture during hydrodissection.

Posterior Capsule Rupture with Vitreous Herniation

To avoid widening the posterior capsular opening, OVDs can be injected into the anterior chamber and the capsular bag as soon as a posterior capsule rupture is detected. Using low-flow aspiration, the remaining cortex can be carefully removed. A vitrectomy can be done under dry conditions beneath the OVD, thus avoiding an uncontrolled extension of the posterior capsular tear.

Preexisting Posterior Capsule Defect

A preexisting posterior capsule defect may be present in children presenting with a white, complete cataract (see Chapter 33). This is seen most often with posterior lentiglobus or trauma. Care must be taken to avoid an uncontrolled extension of any posterior capsule defect at the end of the lens aspiration. The anterior chamber may collapse during the withdrawal of the aspiration probe, allowing the vitreous face to bulge forward, extending the posterior capsular defect. To prevent this, a high-viscosity OVD is injected through the paracentesis incision. The irrigation probe is pulled out only when the majority of the chamber is filled with viscoelastic. This aids in keeping the vitreous in the posterior segment and may prevent the defect from extending further. The surgeon may need to reenter the eye for further cortex removal or for vitrectomy. However, every time the ports are exchanged, it is necessary to repeat the procedure described above to maintain the anterior chamber. With the help of OVDs, posterior capsule support can usually be maintained, thus facilitating an in-the-bag placement of the IOL.

Posterior Capsule Plaque

It is not uncommon to observe a plaque on the posterior capsule. If the opacity cannot be removed completely during lens aspiration, a posterior capsulorhexis will be needed. In older children, OVDs can assist in

the performance of a manual posterior capsulorhexis to remove the plaque without disturbing the vitreous face. A cystitome is used to puncture the posterior capsule, peripheral to the plaque. A viscous OVD is injected slowly through the puncture to push the intact vitreous face posteriorly. Capsulorhexis forceps can then be used to complete a posterior capsulorhexis that incorporates the plaque and removes it completely. Although the OVD is removed from the anterior segment and from the capsular bag after IOL insertion, OVD in the vitreous cavity can be left in place although some risk of a postoperative intraocular pressure spike is still present when higher-viscosity OVDs are used. Using a lower-viscosity cohesive OVD may reduce this risk. The point here is that the use of OVDs can help avoid a vitrectomy in older children, even when a posterior capsulorhexis is performed.

IOL Implantation

When using OVDs before IOL implantation, it is important to remember the site of fixation. *State-of-the-art* “in-the-bag” fixation requires injection of OVDs into the capsular fornices (between the anterior capsule and the posterior capsule) (Fig. 16.2A and B). The OVD should be injected directly into multiple quadrants of the capsular equator to separate the anterior capsule from the posterior capsule. In children, this maneuver needs to be more deliberate than in adults. If OVD is merely placed into the anterior chamber, the capsular equator may remain closed. Since the pediatric posterior capsule may be abnormal in cases of plaque or

lentiginosus, filling the capsular bag with OVD helps to assure that the IOL can be injected into the capsular bag and rotated if necessary without touching the posterior capsule. A safe cushion of OVD anterior to and posterior to the IOL at insertion is the safest way to position the IOL. Some single-piece acrylic IOLs are tacky on their surface, and posterior capsule traction can occur if these IOLs are rotated when in contact with the posterior capsule. Placing highly viscous OVD posterior to the IOL avoids that potential traction on the posterior capsule. Removal of the posterior OVD may be challenging, however. It is recommended that the aspiration handpiece be placed under the optic when removing OVD to assure that none remains between the IOL and the posterior capsule.

Congenital Subluxated Crystalline Lens

A lower-viscosity dispersive OVD can be placed over the area of the weakened or absent zonular fibers to tamponade the vitreous. A high-viscosity cohesive OVD can then be placed over the crystalline lens to help when performing the capsulorhexis. Low-flow aspiration of the lens contents can be accomplished with retention of the dispersive OVD placed over the exposed vitreous.

Uveitis

In eyes with uveitis, the surgeon often needs to deal with a miotic pupil associated with synechia. OVDs can facilitate synechiolysis and add to dilation of the pupil (Fig. 16.3).

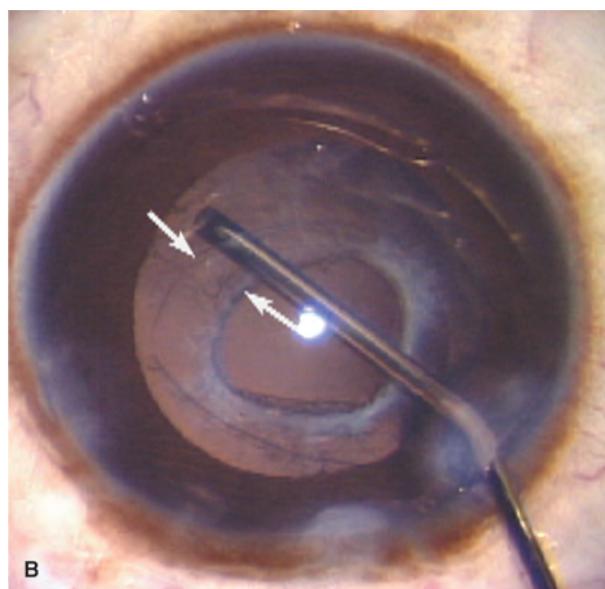
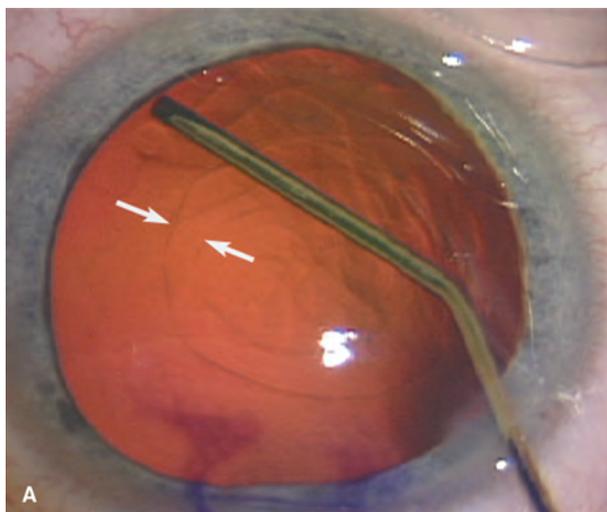


Figure 16.2. Injection of OVDs into the capsular fornices (between the anterior and the posterior capsules).
A: Primary IOL implantation. **B:** Secondary IOL implantation.

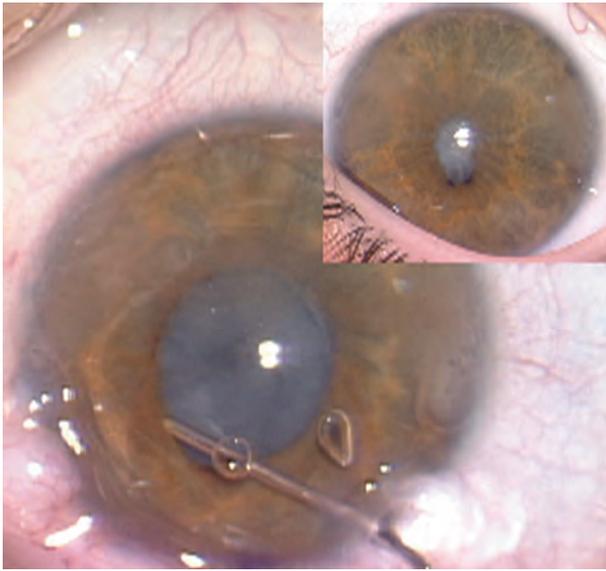


Figure 16.3. An eye with uveitis in a 7-year-old child with juvenile rheumatoid arthritis. **Inset:** Preoperative appearance.

Secondary IOL Implantation

OVDs can assist with a more atraumatic lysis of posterior synechia. Viscodissection, combined with gentle instrument dissection, can clear the ciliary sulcus in preparation for placement of an IOL, thus avoiding lens implant decentration that can result from incomplete synechiolysis. When attempting secondary IOL implantation in-the-bag, again it is important to inject OVDs into the capsular fornices (between the anterior capsule and the posterior capsule) (see Fig. 16.2B).

REMOVAL OF OVDs

As in adults, retained viscous OVDs can cause a marked postoperative intraocular pressure elevation after surgery for childhood cataracts. Englert and Wilson²³ have

suggested the need for more meticulous removal of OVDs. We reported high incidence of symptomatic early IOP spike in patients with aphakic glaucoma undergoing secondary IOL implantation and warranted meticulous OVD removal, considering the use of topical and/or systemic glaucoma medication, and monitoring during the early postoperative period.²⁴ Cohesive OVDs, while responsible for most intraocular pressure spikes, are easier to remove than are dispersive OVDs. As discussed previously, most high-viscosity cohesive OVDs are made from Na–Ha, which is not metabolized in the eye. It passes through the trabecular meshwork as a voluminous molecule, such that anterior chamber clearance depends mostly on the viscosity and the amount of OVD injected. In contrast, HPMC, found mostly in dispersive OVDs, may be partially metabolized prior to its exit from the eye.

Na–Ha stays in the vitreous much longer than in the aqueous, and its residence in the vitreous depends on the concentration, the viscosity, the volume, the presence of a lens capsule, and inflammation of the vitreous and on the thickness, density, and structural integrity of the cortical gel. The vitreous may not tolerate HPMC as well as it tolerates Na–Ha, however. With the former, white precipitates, vitreous bands, and inflammation have been reported in rabbit eyes.²⁵

OVDs can be removed with irrigation/aspiration using a small bimanual handpiece or the vitrector handpiece. Although posterior capsular striae have been reported as a sign of complete removal of the OVD in adult eyes, striae may not appear in the young eye, even with an intact posterior capsule.^{26,27} It is relatively easy to remove OVDs in an eye with an intact capsule (Fig. 16.4A and B). When planning to do posterior capsulectomy and anterior vitrectomy, from the *OVD removal* point of view, the pars plana approach (after OVD removal) is better. With the limbal approach, it is sometimes difficult to achieve complete removal of OVDs (see Chapter 20). Dholakia et al.²² reported Healon GV to be removed using two paracentesis incision with bimanual limbal vitrectomy.

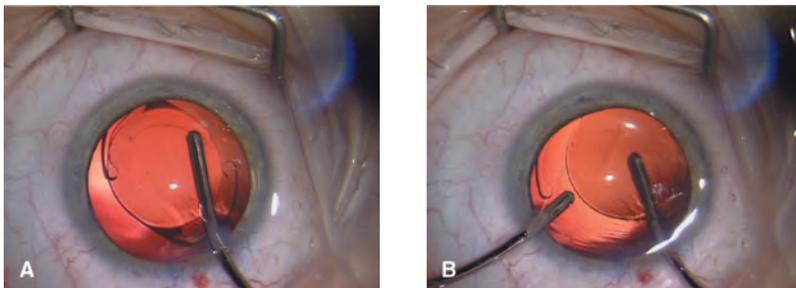


Figure 16.4. Removal of OVDs (A,B). Note that sometimes it is also necessary to go behind the IOL optic for thorough removal of OVDs (B).

SUMMARY

Advances in OVDs hold the promise of making surgical maneuvers easier within the small, often unstable anterior chamber of young children. Using high-viscosity cohesive OVDs is a *necessity rather than a choice* during pediatric cataract surgery. The lower-viscosity dispersive OVD is used to isolate and protect an area of concern, whereas the higher-viscosity cohesive is used to create and preserve spaces and to pressurize. The cohesive will rapidly be aspirated out while the dispersive will largely remain where it was placed.² We recommend using viscoadaptive agents or superviscous OVDs during pediatric cataract surgery to facilitate the difficult intraocular manipulations. These cohesive OVDs help maintain space, promote anterior chamber stability, and offset somewhat the low scleral rigidity and increased vitreous upthrust found in pediatric eyes. Although superviscous and viscous cohesive OVDs are costly, which may prevent their use in developing countries, we recommend their use whenever possible. In special situations (e.g., compromised corneal endothelium), it is advantageous to use a lower-viscosity dispersive agent in combination with cohesive OVDs as discussed in more detail under “soft-shell technique.” Also, during subluxated lens removal, it is advisable to use dispersive OVDs to place over the area of the weakened zonular fibers to tamponade the vitreous.

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Anterior Capsule Management

M. Edward Wilson and Rupal H. Trivedi

Anterior capsulotomy, specifically a continuous curvilinear capsulorhexis (CCC), is notoriously difficult in infants and young children because of the extreme elasticity of the anterior capsule, positive vitreous pressure, and at times, poor dilatation of the pupil.^{1,2} The availability of better operating microscopes and microsurgical instruments combined with higher-viscosity ophthalmic viscosurgical devices (OVDs) is helpful, but the creation of an intact capsulorhexis in infants remains a challenge. Unique characteristics of the anterior capsule in children have created the need for variations in techniques used for anterior capsulotomy in adult eyes. A surgeon who applies adult anterior capsule maneuvers to the pediatric anterior capsule may be surprised and disappointed at the outcomes. This is especially true in infant eyes. This unpredictability has led researchers and surgeons to use alternative methods to open the anterior capsule in children.³⁻¹¹ The desired end result of any alternative is the consistent achievement of a continuous curvilinear anterior capsulotomy of a desired size and shape, centered on the visual axis. In addition, the capsule opening should be smooth and strong with no tags, tears, or right-angled edges. Prior to the late 1980s, the strength and size of the capsulotomy was of little concern in pediatric eyes. In fact, when the vitrector was introduced as an instrument to remove pediatric cataracts, the entire lens capsule was often removed—even the capsular equator—to assure that secondary closure of the capsulotomy did not occur.

The current widespread acceptance of intraocular lens (IOL) implantation in the pediatric eye has created the need for an opening in the anterior capsule that can be made to a specific size and shape and with an edge that can withstand the stress of implantation maneuvers. An intact anterior capsulotomy supports all the subsequent steps of lens aspiration and IOL implantation. Anterior capsulotomy techniques have evolved in pediatric cataract surgery. Adult techniques have been modified, and some uniquely pediatric techniques have been developed. For surgeons experienced in adult procedures, the pediatric anterior capsulotomy is often mentioned as the

operative step requiring the most adjustment from the adult routine. Wilson detailed the evolution of pediatric capsulotomy and compared various capsulotomy options (using clinical, cadaveric, and porcine data and including scanning electron microscopy [SEM]) in a thesis submitted as partial requirement for membership in the American Ophthalmological Society.²

CHARACTERISTICS OF THE PEDIATRIC ANTERIOR CAPSULE

The anterior capsule is thinnest at birth, increasing in thickness with age until approximately age 75 years.¹² Seland¹³ described the neonatal anterior lens capsule as uniform and approximately 4 mm in thickness. By old age, the capsule had increased to 20 to 25 mm centrally and 30 mm near the lens equator.¹³ Krag et al.¹² analyzed 67 human eyes from age 7 months to 98 years and found that anterior capsule thickness increased from 11 to 33 mm. The association of thickness with age fit a straight line from birth to age 75 years, after which it changed slope and thinned slightly.¹² In conjunction with these anatomic changes, the anterior capsule's biomechanical properties are also altered with age.

The young anterior lens capsule is strong and very elastic. The elderly anterior lens capsule is, by comparison, weak and inelastic. Krag et al.¹² found that anterior lens capsule extensibility was maximal in infancy and decreased about 0.5% per year throughout life (measured range, 108% to 40%). Aging of the human anterior lens capsule leads to a progressive loss of mechanical strength. Overall tensile strength decreases by a factor of five during the life span, and the extensibility decreases by at least a factor of two. In surgery, the young capsule is highly elastic and difficult to puncture. Much more force is required before tearing begins. In contrast, the capsule of the elderly is much less extensible, easier to open, and tears with much less force. These differences between the adult and child must be understood and accounted for when planning for surgery on the pediatric lens.

ANTERIOR CAPSULOTOMY TECHNIQUES

The first anterior capsulotomy technique associated with an IOL implantation, performed by Sir Harold Ridley in the 1950s, was an uncontrolled tear using forceps without the aid of microscope magnification.¹⁴ It served only as a means to gain access to the lens contents. Complete removal of the anterior capsule was desired. If remnants or tags were seen after nucleus expression, they were torn with smooth-bladed capsule forceps. This maneuver at times resulted in the entire posterior capsule's also being delivered from the eye, effecting an unplanned intracapsular surgery.

Can-Opener Capsulotomy

To reduce stress placed on the zonular fibers and produce a more rounded capsulotomy with fewer tags, the multi-puncture or can-opener capsulotomy came into common use. A capsulotomy was made by punching a 27-gauge bent-needle cystitome repeatedly through the anterior capsule in a postage stamp or can-opener fashion and connecting the punches to form a circular opening. A jagged but circular opening in the capsule was formed. Frequently, however, radial extensions of the capsulotomy occurred during lens removal or IOL insertion. These tears resulted in uncertainty regarding the stability of the capsular bag and the position of the IOL after implantation. The can-opener technique puts more stress on the zonular fibers and is more likely to leave capsule tags or strands that can interfere with lens aspiration. Pediatric surgeons seldom use the can-opener capsulotomy method, but when needed, it is safer and more effective in children than in adults. The elastic nature of the pediatric capsule causes each can-opener puncture to convert to a small arc tear similar to a mini manual CCC.

Manual CCC

The technique known today as manual CCC was developed simultaneously in North America by Gimbel and in Europe by Neuhann.¹⁵ The CCC method clearly creates the “gold standard” capsulotomy edge. While a manual CCC is ideal for adults, it is more difficult to perform in infantile eyes with cataracts. However, it is still the gold standard since it resists tearing once completed successfully. Because of the increased elasticity of the pediatric anterior capsule, more force is required when pulling on the capsular flap before tearing begins. Control of the capsulectomy and prevention of extensions out toward the lens equator are inversely related to the force needed to generate the tear. As a result, inadvertent extensions out to the lens equator (known as the “runaway” rhexis) are common in children. In addition, reduced scleral rigidity in children produces what has been called posterior

vitreous “pressure” when the eye is entered. The vitreous in children is formed and less liquefied. Scleral collapse causes an “upthrust” from the formed vitreous that pushes the lens anteriorly and keeps the anterior capsule domed, convex, and taut. This results in the tendency for the capsulorhexis to spiral outward.

When performing a manual CCC capsulotomy in children, the following caveats are offered²:

1. Use a high molecular weight OVD to flatten the anterior capsule and deepen the anterior chamber. This will create laxity in the anterior capsule and combat the effects of low scleral rigidity.
2. Aim for a slightly smaller-than-desired capsulotomy. With the stretch in the anterior capsule, the opening is usually larger at completion than it appears to be during the active tearing.
3. When creating the manual CCC capsulotomy, frequently release the capsular flap and inspect the size, shape, and direction of the tear. Regrasp near the site of the continuous tear and readjust the direction of pull if needed to keep the capsulotomy on the planned course. Pulling the leading edge along the path of the tear is called “shearing,” and this is the predominant force used in adult capsulotomy. In children, more pull is often needed toward the center of the pupil (90 degrees to the path of the tear) to avoid an extension of the manual CCC out toward the lens equator. This is referred to as “tearing.”

Additional OVD should be added as needed to keep the capsule lax during the tearing. Use of capsular dye can be beneficial (Fig. 17.1). To rescue a manual CCC that begins to extend peripherally, regrasp near the leading edge of the capsulotomy and redirect the capsular tear by pulling back in the direction from which the tear was originating (so-called “pull-back” technique). Rarely, the

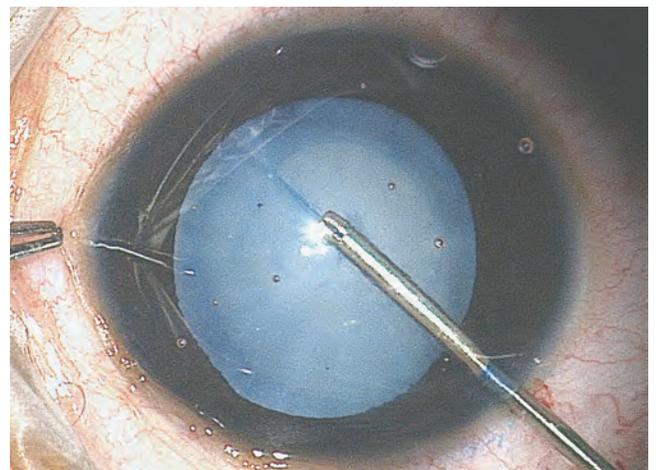


Figure 17.1. Manual CCC using trypan blue in a 2-year-old child with total cataract.

vitrector or the Kloti radiofrequency diathermy device is used to rescue the errant capsular tear and round out the capsulorhexis.

Some surgeons have altered the adult technique even more substantially in order to facilitate CCC in elastic capsules. Auffarth et al.⁴ developed a modified CCC technique for use in experiments on eyes of young albino rabbits and suggested that it be used for young human capsules as well. The technique begins with a puncture of the lens capsule at the superior border of the intended capsulotomy using a 27-gauge needle. Capsulorhexis forceps are then used to grasp the anterior capsule centrally. The capsular flap is torn toward the 6 o'clock position until a half-circle is completed. The force is then reversed toward 12 o'clock position, pulling with equal force to both tearing edges. The authors reported a radial tear in only 2 of the 32 rabbit eyes.

Nischal⁸ described a modification of the Auffarth technique in which two stab incisions are made in the anterior capsule, outlining the desired diameter of the capsulorhexis (two-incision push-pull, or TIPP). Capsulorhexis forceps are used to grasp one end of the distal edge of the proximal anterior capsule stab incision. The grasped edge is gently pushed toward the corresponding point of the distal stab incision until the edge reaches halfway to the distal stab. The corresponding end of the proximal edge of the distal stab incision is similarly grasped but with the capsule pulled gently toward the proximal stab incision. The two tears meet to form the CCC. This is repeated for the other end of each stab incision to complete the entire CCC. The tearing force, using this technique, is always directed toward the center of the pupil.¹⁶

Vitrectorhexis

Mechanical suction and cutting instruments began to be utilized for pediatric cataract surgery in the late 1970s and early 1980s. Parks¹⁷ and Taylor¹⁸ were among the first to advocate for the performance of a primary mechanized posterior capsulectomy and anterior vitrectomy during pediatric cataract surgery. Taylor also described removal of the anterior capsule mechanically.¹⁸ A mechanized (vitrector-cut) anterior capsulotomy technique combined with IOL insertion in children was first described, studied, and popularized by Wilson et al. in the mid-1990s.^{3,19} It rapidly replaced the can-opener and even the manual CCC for infants and toddlers having cataract surgery, whether or not an IOL was implanted primarily.

The commonly used term “vitrectorhexis” emphasizes the fact that it is a substitute for capsulorhexis performed using the vitrector. It is, in reality, a misnomer, because “rhexis” means to tear rather than to cut. The original name, “mechanized anterior capsulectomy,” emphasized the mechanized nature of the vitrector handpiece and the fact that a portion of the capsule was removed (capsulectomy) rather than merely opened (capsulotomy).^{3,19}

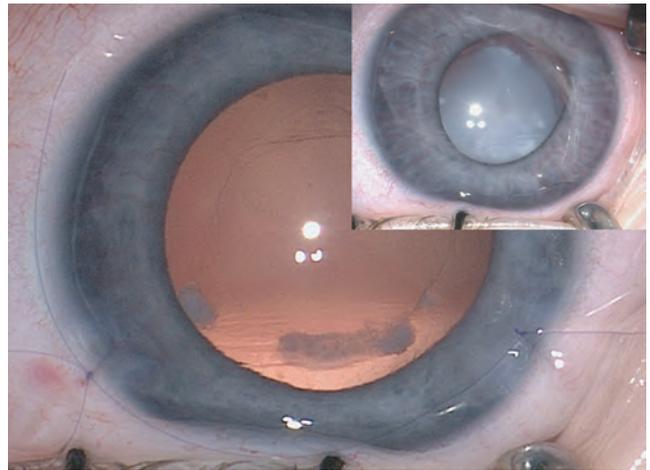


Figure 17.2. Vitrectorhexis in a 4-month-old child. **Inset:** Preoperative appearance.

When performing a vitrectorhexis anterior capsulotomy (Fig. 17.2), the following caveats are offered²:

1. Use a vitrector supported by a Venturi pump. Peristaltic pump systems will not cut the anterior capsule as easily.
2. Use a separate infusion cannula that matches the gauge of the vitrector handpiece, thus assuring that the incisions into the eye provide for a tight fit while the instruments are in the eye. The anterior chamber of these soft eyes will collapse readily if leakage occurs around the instruments.
3. Separate the incisions for the infusion and vitrector handpieces by at least 4 clock hours. This will enable easy access to lens capsule and lens cortex throughout the anterior chamber. If the infusion cannula and the vitrector handpiece are the same gauge, the instrument positions can be switched to gain access to subincisional lens cortex during aspiration without producing leakage at either wound site.
4. Do not begin the capsulotomy with a bent-needle cystitome. The increased intralenticular pressure and vitreous upthrust of the pediatric eye may cause lens material to spontaneously prolapse through this initial opening and create a radial tear. Instead, place the vitrector, with its cutting port positioned posteriorly, in contact with the intact anterior capsule. Turn the cutter on and increase the suction using the foot pedal until the capsule is engaged and opened. A slow cutting rate of 150 cuts per minute with high infusion rate (raise the bottle to the maximum height for gravity-fed infusion or use a fluid setting of 50 mm Hg for active fluid pump systems) is recommended when cutting the anterior capsule (as opposed to the high cutting rate with lower infusion used to cut the vitreous). With the cutting port facing down against the capsule, enlarge the round capsulotomy in a spiral fashion until the desired size and shape are achieved.

5. Any lens cortex that escapes into the anterior chamber during the capsulotomy can be easily aspirated without interfering with the capsulotomy technique.
6. Care should be taken to avoid right-angled edges, which are predisposed to radial tear formation. With experience, a more rounded capsulotomy is made and fewer angled edges are formed. If a right-angled edge is seen during the capsulotomy, it should be rounded out using the vitrector before completion of the capsulotomy. If the capsulotomy needs to be enlarged, the vitrector can be used for this purpose even after the IOL is in place (often coincident with removal of the OVD).
7. The vitrectorhexis technique is not recommended for older children (above age 8 years) unless the anterior capsule is fibrotic or already ruptured preoperatively. The manual CCC is easier to control and complete in older children as opposed to the very young. The manual CCC is the “gold standard” capsulotomy and produces the most stable edge. It is recommended for children above the age of 2 years who will have the posterior capsule left intact and will undergo no anterior vitrectomy. This transition from vitrectorhexis anterior capsulotomy, lens aspiration, IOL implantation, vitrectorhexis posterior capsulotomy, and anterior vitrectomy to manual CCC, lens aspiration, IOL implantation, and an intact posterior capsule will occur with patients aged 6 to 8 years for many surgeons but will occur at an earlier age for others.

The advantage of the vitrectorhexis when operating on small eyes is that the anterior capsulotomy and the lens aspiration can be done sequentially without taking the instruments from the eye. Since the vitrector is most efficient when placed through a tight-fitting wound, small-incision capsulorhexis forceps are recommended for manual CCC in young eyes. The smaller wound (20 gauge or smaller) allows easy conversion to a vitrectorhexis if the manual CCC begins to tear peripherally.

Radiofrequency Diathermy Capsulotomy

Developed by Kloti et al., radiofrequency diathermy has been used as an alternative to CCC for cataract surgery in children (Fig. 17.3). The Kloti device (Oertli Instruments, Berneck, Switzerland) consists of a curved cannula housing an active electrode tip. It cuts the anterior capsule with a platinum-alloy-tipped probe using high-frequency current (500 kHz). The probe tip is heated to about 160°C and produces a thermal capsulotomy as it is moved in a circular path across the anterior capsule. The Kloti radiofrequency diathermy handpiece is reusable but must be placed in fluid immediately after use to avoid coagulated capsule remnants from interfering with the function of the tip on the next application. Gentle but consistent contact with the capsule must be maintained

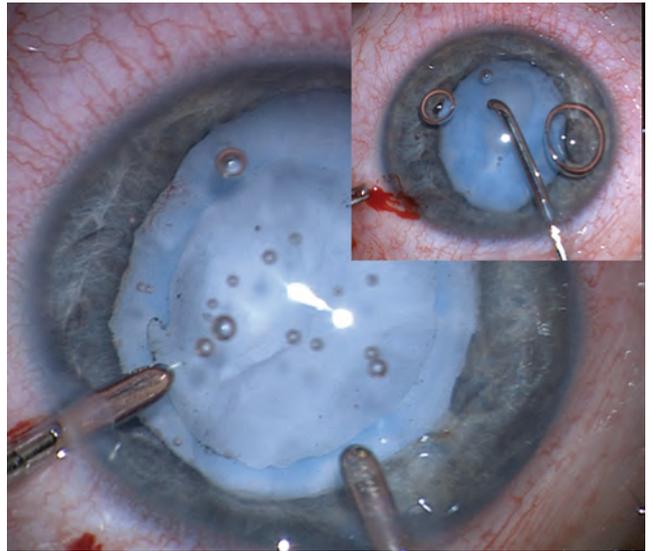


Figure 17.3. Diathermy was used to perform anterior capsulotomy in a 6-year-old child with traumatic cataract. Note that capsulorhexis was initially started with forceps and later converted to diathermy.

when using the Kloti radiofrequency diathermy handpiece. If contact is too light or movement is too fast, skipped areas will result. If contact is too firm or movement too slow, the tip will burn through the capsule and enter the lens cortex. Subsequent tip movement drags the capsulotomy edge rather than cutting it, which can cause radial tearing.

Small gas bubbles are formed while the tip is active. Even when performed perfectly, a diathermy-cut capsulotomy can be seen to have coagulated capsular debris along the circular edge. In addition, this edge has been shown experimentally to be less elastic than a comparable CCC edge.^{20,21} Since the stretching force needed to break the edge of a diathermy-cut capsulotomy is reduced compared to a CCC edge, surgical manipulations needed to remove a cataract and place an IOL may result in more radial tears when the diathermy is used. Several advantages were listed for the diathermy capsulotomy over other techniques, including a precisely controlled capsulotomy size and a well-defined grayish coagulation line that helps define the border of the capsulotomy (making implantation easier). Kloti²² recommended using a viscosurgical device rather than continuous irrigation to ensure that the anterior chamber was maintained and that the corneal endothelium was protected from heat.

Fugo

The Fugo blade unit is a portable electronic system that operates on rechargeable batteries. It requires no red reflex for visualization of the capsulotomy edge during the cutting procedure. The Fugo plasma blade was recently developed and has been approved by the Food and Drug Administration for capsulotomy use. The Fugo blade

uses plasma technology to create a nearly resistance-free incision into the anterior capsule. The anterior capsule is gently applanated with the tip activated, and a continuous circular movement is made to create the capsulotomy. Alternatively, several arcuate incisions on the capsule can be made and connected to form a circular capsulotomy. The capsulotomy is performed under an OVD. Cavitation bubbles are created along the path of the capsulotomy as the Fugo blade cuts it.

LABORATORY STUDIES

Wilson et al.³ performed a laboratory comparison of vitrectorhexis and manual CCC. A mechanized anterior capsulectomy was performed on one eye of each pair, and a CCC was performed on the fellow eye. The integrity of the anterior capsulotomy edge was assessed after capsulotomy completion, after lens removal, and again after IOL implantation. Radial tears were noted and described. A radial tear developed in only one of the 18 pediatric eyes in which a mechanized anterior capsulectomy was performed. This occurred in one of the oldest pediatric eyes (age 16 years), where a single radial tear extended from an angled capsulectomy edge during IOL insertion. In contrast, no radial tears occurred in any of the fellow eyes in which a manual CCC was performed. However, in six of the fellow eyes, the manual CCC edge extended out to the lens equator rather than continuing in a circular fashion. All six of these errant capsulotomies were from eyes of children younger than 5 years of age.

Subsequently, an additional comparative study between manual CCC and vitrectorhexis using adult pig eyes was performed.²³ To validate the use of the pig eyes, CCC and vitrectorhexis made in pig eyes were also compared to those made in pediatric human eyes. The mean percentage stretch at the break point for human infant eyes aged 1 to 8 months was 165% for CCC and 138% for vitrectorhexis, which correlated well with the data from the adult porcine model (mean stretch = 157% for CCC and 135% for vitrectorhexis). The percentage of stretch prior to rupture was higher for CCC than for vitrectorhexis ($P < 0.001$), but all capsules stretched adequately for IOL insertion. The porcine capsule has been shown to be a valid model for the pediatric human capsule. As in human pediatric eyes, the capsule of the pig is very elastic and difficult to puncture.

Luck et al.²¹ noted that on SEM analysis, the diathermy caused a loss of the normal lamellar architecture of the capsule with distorted and unrecognizable collagen fibrils and adherent lens fibers. In contrast, the manual CCC edge was completely smooth and free from irregularity with a preserved lamellar organization. Morgan et al.²⁰ found that the mean increase in edge length before breakage was 53% using the manual CCC and 18% in the diathermy group. Krag et al.¹² found that the mean

extensibility (percentage elongation above the resting circumference at break point) of the diathermy capsulotomy was markedly greater in the porcine eyes (62%) than in the human adult eyes (38%). Also, the difference in mean extensibility between the manual CCC and the diathermy capsulotomy (68% versus 38%, $P < 0.001$) in human adult eyes was not present in the porcine eyes (62% versus 62%, $P > 0.05$). This implies that the diathermy capsulotomy may have a maximum extensibility more like a manual CCC when performed on an elastic capsule like the pediatric human lens capsule. Interestingly, Krag et al.¹² found that the force required to reach the maximum extensibility (the break point) was much less in the diathermy capsulotomy compared to the manual CCC in both the adult human and the porcine eyes ($P < 0.001$). This implies that the heat from the diathermy denatures the collagen and causes it to lose its stiffness. Using a porcine model, Wood and Schelonka²⁴ compared can-opener capsulotomy to manual CCC capsulotomies. One radial tear was noted in 27 consecutive can-opener capsulotomies (2.1%). In contrast, the overall failure rate for manual CCC in their study was 22.5%. The relative risk of an uncontrolled radial tear during the manual CCC was 10.58, as compared to the can-opener capsulotomy. Even more surprising was the finding that the mean maximum strain (stretch to rupture) of the can-opener capsulotomies (46.7%) was not statistically different from the mean maximum strain of the manual CCC (47.7%) in their porcine model.²⁴

We compared the extensibility and SEM of five currently used pediatric anterior capsulotomy techniques: vitrectorhexis, manual CCC, can-opener, radiofrequency diathermy, and plasma blade, in a porcine model.²⁵ Applicability of these results assumes that the greater the stretching force needed to break the edge of a capsulotomy and the smoother the capsule edge, the less the risk for anticipated radial tear during surgical manipulations to remove a cataract and implant an IOL. To determine extensibility, we stretched each capsulotomy until it ruptured and calculated the mean stretch-to-rupture circumference of each capsulotomy (20 eyes per technique) as a percentage of its baseline circumference. The mean extensibility of each technique tested (vitrectorhexis 161%, CCC 185%, can-opener 149%, radiofrequency 145%, and plasma blade 170%) was significantly different ($P < 0.001$, one-way analysis of variance).

The SEM examination (Fig. 17.4) found that the vitrectorhexis had a scalloped edge with the whole edge rolled over, presenting a smooth surface toward the inside of the capsulotomy; the manual CCC produced the smoothest edge, with no irregularities noted; the can-opener edge was irregular, showing each puncture of the needle had created a small arc, with occasional regions of the edge rolled over in a “hit-and-miss” fashion; the radiofrequency diathermy capsulotomy edge was ragged,

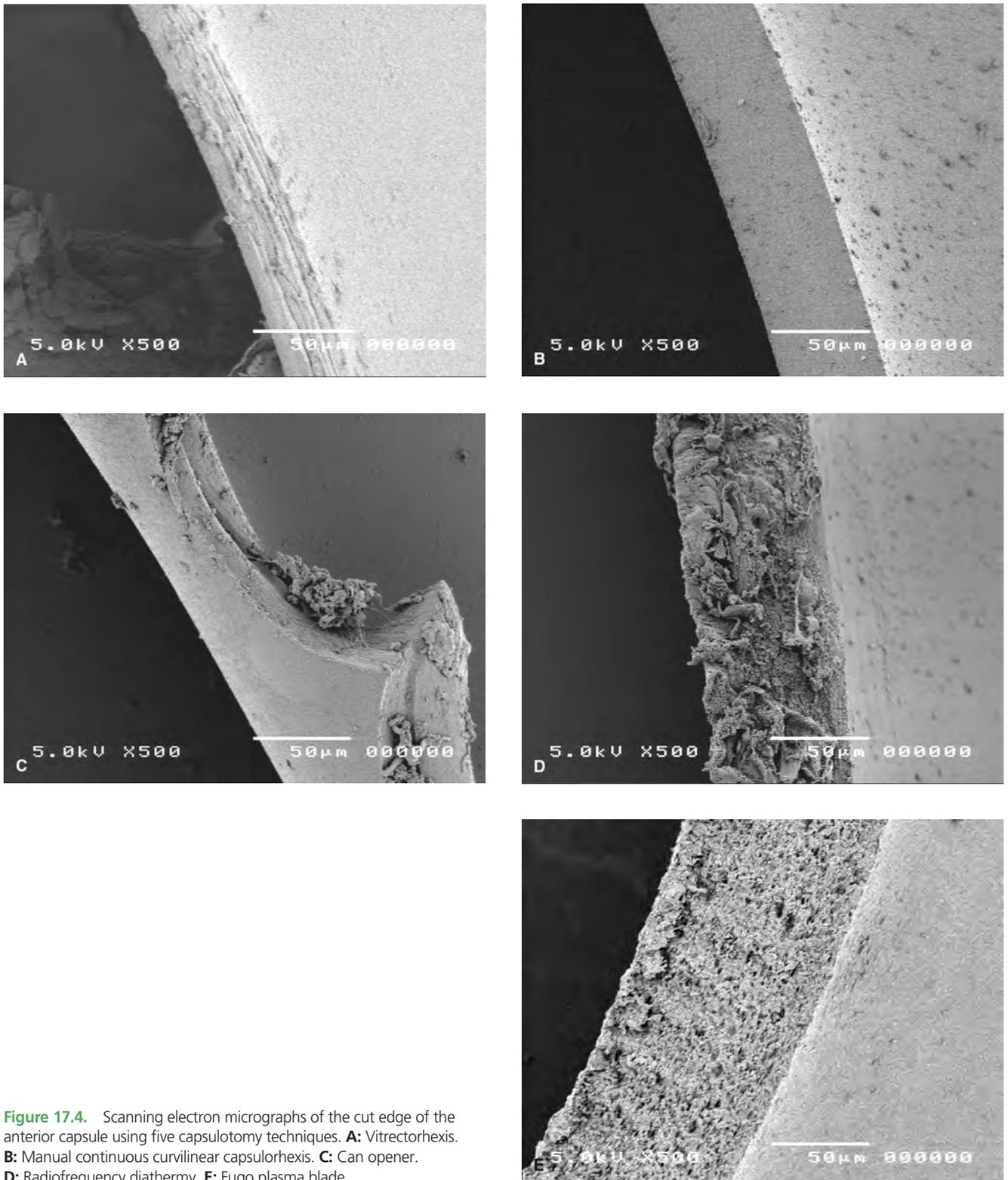


Figure 17.4. Scanning electron micrographs of the cut edge of the anterior capsule using five capsulotomy techniques. **A:** Vitreorhexis. **B:** Manual continuous curvilinear capsulorhexis. **C:** Can opener. **D:** Radiofrequency diathermy. **E:** Fugo plasma blade.

rough, and irregular; and the plasma blade capsulotomy edge was rougher than the manual CCC, but there were fewer irregularities than the radiofrequency diathermy edge had. We concluded that the manual CCC technique produced the most extensible porcine capsulotomy,

followed by the plasma blade, vitreorhexis, can-opener, and radiofrequency techniques, in a porcine model. The manual CCC technique also produced the smoothest anterior capsulotomy edge according to SEM evaluation. We found that the can-opener capsulotomy stretched to

a mean diameter of 149% of the unstretched diameter before breaking. This is very similar to the findings of Wood and Schelonka²⁴ discussed above. Extensibility in the current study is reported as the percentage of stretched compared to unstretched capsulotomy circumference. Wood and Schelonka²⁴ reported the percentage of stretch beyond the baseline resting circumference. Thus, the two figures, 149% and 46.7%, represent very similar stretch characteristics. The manual CCC capsulotomy in our porcine study stretched to a mean of 185% compared to the unstretched circumference. This is in contrast to the 47.7% mean stretch-to-rupture reported by Wood and Schelonka.²⁴

Techniques described below have not been reported for pediatric clinical use. Palanker et al.²⁵ evaluated pulsed electron avalanche knife (PEAK) for creating anterior capsulotomy. The study included 50 bovine eyes and 10 rabbit eyes. Authors reported that histology and SEM showed that the PEAK produced smooth capsule cutting without radial tears. This electrosurgical device was introduced for precise cutting of ocular tissue without thermal damage. It produces a sharp and clean-edged capsulotomy, without radial nicks and tears. However, it is still a manual procedure, with the associated problem of centration and sizing of the capsulotomy. Femtosecond laser-assisted anterior capsulotomy has been reported to overcome this problem.^{26,27} Femtosecond laser produced continuous anterior capsule incisions, which were twice as strong and more than five times as precise in size and shape than manual CCC.²⁶

Clinical Outcome

We conducted a retrospective analysis of 358 consecutive pediatric cataract surgeries performed between January 1, 1994, and December 31, 2003.² The mean age of all patients was 53.5 months \pm 50.4 (birth to 234 months). These cases included 74 eyes left aphakic and 284 pseudophakic eyes. The analysis revealed that the anterior capsulotomy withstood the stresses of surgical manipulation without tearing in 92% (329 of 358) of cases. Twenty-nine anterior capsular tears were recorded during the 284 pediatric cataract surgeries where an IOL was implanted. The surgical step during which each tear occurred is as follows: 4 (13.8%) of 29 tears occurred during the capsulotomy, 7 (24.1%) during cataract removal, 13 (44.8%) during IOL insertion, 4 (13.8%) during hydrodissection, and 1 (3.4%) during OVD removal. With current techniques, the tear rate has dropped even further.

Can-Opener Capsulotomy

A meta-analysis of published cases from 1983 to 1995 revealed a total of 509 can-opener capsulotomies in children with follow-up ranging from 4 to 18 years.²⁴ A reoperation was documented in seven eyes (1.4%). None of

the repeated surgeries were attributed to defects or tears in the capsulotomy. Basti et al.²⁸ reported a total of 169 operated eyes with all “except the last few” having had a can-opener capsulotomy. A radial tear was reported in only four eyes, and in each of these, the IOL was still placed easily into the capsular bag. These rather dramatic differences between the pediatric and adult can-opener capsulotomy make the technique more recommendable for children than for adults. However, it appears that pediatric surgeons no longer use the can-opener technique except in very rare instances.

Manual CCC and Vitrectorhexis

We compared the rate of inadvertent anterior lens capsular tears with CCC or vitrectorhexis in pediatric cataract and IOL implantation surgery.⁶ Of the 339 eyes, 19 eyes (5.6%) were noted to develop an anterior capsule tear (vitrectorhexis, 12 of 226 eyes, 5.3%; CCC, 7 of 113, 6.2%). These tears occurred during anterior capsulotomy in seven eyes, hydrodissection in one, cataract removal in three, and IOL insertion/manipulation in eight. In eyes operated for cataract at or before 72 months of age, the manual CCC technique was more likely to develop a tear (relative risk, 3.09) compared with eyes of older children (>72 months of age), where the vitrectorhexis technique was more likely to develop a tear (relative risk, 3.14).

Kloti Radiofrequency Diathermy

Rate of radial tear is reported as 0% to 21%.^{2,7}

Fugo Plasma Blade

We reported five capsule tears (62.5%) in eight cases using the Fugo plasma blade capsulotomy technique. The tears occurred during hydrodissection (two eyes), IOL insertion (one eye), or cataract removal (two eyes).

WORLDWIDE ANTERIOR CAPSULOTOMY SURVEY OF PEDIATRIC SURGEONS

We compared the pediatric anterior capsulotomy preferences of members of the American Society of Cataract and Refractive Surgery (ASCRS) and the American Association of Pediatric Ophthalmology and Strabismus (AAPOS) reported in three surveys (1993, 2001, and 2003).^{2,29} Vitrectorhexis and manual capsulotomies were the most popular choices, where preference appears to inversely relate to patient age, particularly during the first 6 years of life, between these two techniques. In 1993 and 2001, more than 50% of ASCRS respondents preferred manual anterior capsulotomy techniques; in 2001 and 2003, AAPOS respondents preferred manual and vitrector techniques. The ASCRS preferences

remained unchanged when subdivided into domestic and international, as did AAPOS domestic preferences; however, more than 50% of AAPOS international preferences changed from manual alone in 2001 to a manual–vitrector combination in 2003. In 2003, more than 50% of AAPOS respondents worldwide preferred this combination: the vitrector for the very young patient and the manual anterior capsulotomy for the older child. In a 1994 survey, only 15.0% of the responding surgeons reported using a can-opener capsulotomy when operating on children. The 2003 surgeon preference survey included in this thesis showed an even lower utilization rate of 1.9% to 5.2%, depending on the age of the child. For U.S. surgeons, the can-opener capsulotomy technique utilization rate ranged from a high of 4.5% of surgeons when operating on patients <1 year of age to 1.9% of surgeons when operating on children older than 6 to 8 years of age. Multiple responses were allowed, so this represents the total number of responding surgeons who reported the technique as one of their preferred options utilized at the indicated ages between <1 and 18 years. The manual CCC was the most commonly utilized anterior capsulotomy technique when operating on children aged 2 years and older based on the survey of the AAPOS membership.

SPECIFIC ISSUES

Dye Enhanced

Saini and colleagues compared the clinical efficacy of trypan blue 0.1% (Bluehex) in creating anterior CCC during pediatric cataract surgery. In a prospective randomized study, an anterior CCC was performed with or without trypan blue. The authors reported 21/23 (91.3%) eyes had complete CCC when dye was used compared to 14/19 (73.5%) when dye was not used.

White Cataract

As emphasized earlier, a manual CCC creates an opening in the anterior capsule of the crystalline lens that is resistant to tear during cataract removal and ensures in-the-bag implantation of an IOL. The retroillumination produced by the operating microscope is important in visualizing the anterior capsule while performing CCC. It is difficult to distinguish the anterior capsule from the underlying cortex in total, advanced/white cataracts and in patients with pigmented fundi or vitreous disease, owing to the absence of the red reflex. Poor visualization of the capsule results in an inadequate CCC, with a high risk of radial tears toward or beyond the lens equator and associated complications, such as zonular and posterior capsular rupture, vitreous loss, and IOL displacement. Methods used to enhance the visualization of the anterior capsule during CCC in white cataracts include capsular staining, side illumination with an endoilluminator,

injection of an air bubble in the anterior chamber, hemo-coloration of the capsule with the patient's autologous blood. A two-step CCC method, which involves creating a small CCC followed by a second CCC to enlarge the initial capsular opening, is another option. Other techniques to facilitate performing CCC in the aforementioned cases include dimming the room lights, increasing the optical magnification of the operating microscope, using a high-frequency diathermy probe, using high-density viscoelastic agents, performing a vitrectorhexis, and aspirating cortical material in liquefied cataracts.

Traumatic Cataract

Visualization of the anterior lens capsule in pediatric traumatic cataract cases may be difficult due to a ruptured lens capsule with flocculent lens matter in the anterior chamber. Creation of an intact capsulorhexis may be difficult in such a situation. A vitrectorhexis is a good alternative to manual capsulorhexis when traumatic anterior capsule fibrosis is present. Staining of the anterior lens capsule is also usually helpful to enhance visibility in these eyes with a “torn anterior capsule” or “white cataract.”

Anterior Capsule Plaque

Anterior capsule plaques can be better removed by vitrector or with the help of intraocular scissors. Intraocular forceps can also be used with scissors for a bimanual technique. This allows for counter traction and helps the scissors work more efficiently.

Aphakia

Many of the children left aphakic in infancy have now presented for secondary IOL implantation as older children or young adults. Time and again, the vitrectorhexis remains as a round, intact, smooth anterior capsular edge, providing circumferential support for the secondary implant. Unfortunately, in an attempt to decrease inflammation and the incidence of synechia between the capsule remnants and the iris, some pediatric surgeons advocated mechanized removal of as much of the equatorial capsular remnants as possible. This procedure eliminated posterior synechia and reduced re-proliferation of cortex but left no support for a secondary implant. During the late 1980s and the early 1990s, nonimplanting pediatric surgeons nearly uniformly recognized the need for a capsular rim to support an IOL later. Mechanized, anterior capsular openings evolved to a size small enough to leave an adequate capsular ring to support a secondary IOL, yet large enough to prevent frequent closure of the opening by secondary membrane formation. At the time of secondary IOL implantation, the anterior capsular opening is characteristically round, smooth edged, and fused to the posterior capsule.

SUMMARY

The development of capsulotomy techniques that consistently preserve the structure and stability of the remaining lens capsule has helped lead to safer pediatric cataract surgery and the emergence of IOL implantation for children of all ages. The thin, strong, and elastic anterior capsule of children requires a unique approach to the anterior capsulotomy. Manual CCC produces the most stable capsulotomy edge and should be utilized whenever possible, even in the very young. However, the risk of peripheral extensions is greater when manual CCC is attempted in the very young child. The vitrectorhexis technique has become popular anterior capsulotomy for children in the first 1 to 2 years of life and remains a popular choice for some surgeons when operating on patients up through age 6 to 8 years. The vitrectorhexis technique is ideally suited to the very young child with a highly elastic anterior capsule. In these children, the manual CCC is more difficult to complete even with liberal use of a high molecular weight OVD. The vitrectorhexis technique is recommended as the anterior capsulotomy of choice in the first year of life. Manual CCC is recommended for children operated when older than 1 or 2 years of age. For those transitioning from a vitrectorhexis to a manual CCC, the TIPP technique popularized by Nischal is a good option. For surgeons experienced in adult manual CCC, the standard CCC technique will feel most comfortable. The challenge is learning to perform more tearing and pull-back maneuvers when performing a manual CCC on a child from 0-8 years old. The Kloti radiofrequency diathermy and Fugo plasma blade techniques are recommended when fibrotic capsules are encountered. Case selection is an additional factor that has led to a reduction in radial tears in the last 2 years.

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18

Hydrodissection

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and Mamidipudi R. Praveen*

Cortical-cleaving hydrodissection is recognized as an important surgical step for adult cataract surgery. Several authorities have advocated various forms of interlenticular injection of fluid in adult cataract surgery.¹⁻⁶ Faust coined the term *hydrodissection* in 1984.¹ He described it as an injection of fluid designed to separate the lens nucleus from the cortex. In 1992, Fine² published his classic description of the “cortical-cleaving hydrodissection” technique. This highly effective procedure enhances the separation of the capsule from the cortex and is widely accepted as a routine surgical step for adult cataract surgery.

Pediatric cataract surgeons have been less inclined, however, to follow the adult trend for hydrodissection. Hydrodissection is barely mentioned or is glossed over in the management of pediatric cataract surgery. As the main function of hydrodissection is to release the nucleus to facilitate phacoemulsification, which is not usually an issue for pediatric cataract surgeons, the value of hydrodissection in pediatric cataract surgery has been questioned. Since pre-existing posterior capsule defects or weaknesses occur commonly in pediatric cataract patients, the fear of disrupting an already incompetent posterior capsule has made some pediatric surgeons reluctant to use the technique at all.

Numerous advantages of cortical-cleaving hydrodissection (e.g., ease, safety, and efficacy of nuclear emulsification of adult cataracts) are well established in the literature.⁶ Apple et al.⁵ reported that the major advantage of hydrodissection lies in the shearing effect of the fluid wave. Direct injection of fluid under the anterior capsule is particularly useful in helping to remove equatorial lens epithelial cells, thereby reducing the incidence of posterior capsule opacification (PCO). When surgeons leave cortical material, which clinically resembles a strand of fiber, they actually leave large numbers of mitotically active cells from the equatorial lens bow. These cells have the potential to grow across the visual axis, especially the pearl form. The best means of reducing the incidence of this complication is to remove as many of these cells

as possible at the outset. Since PCO is one of the most frequent and severe complications in pediatric cataract surgery, reports that hydrodissection may decrease the incidence of PCO prompted us to apply this inexpensive, practical, immediately implementable procedure to pediatric cataract surgery.⁷

TECHNIQUE

The technique for performing cortical-cleaving hydrodissection has been well described in the literature. Hydrodissection is an absolute contraindication in eyes with a suspected preexisting posterior capsular defect, so case selection is key. Table 18.1 describes the technique of multiple quadrant hydrodissection (Fig. 18.1). A J-shaped Binkhorst cannula can be useful for injection at the subincision site.⁸ Signs of successful hydrodissection are listed in Table 18.2. Like any other technique, hydrodissection has its own learning curve. During the learning phase, it may not be possible to consistently achieve thorough hydrodissection.

Capsulorhexis size is critical in hydrodissection. A small capsulorhexis provides a broader area of anterior capsule into which the cannula can be inserted (up to an optimal length). However, this may make removal of the lens substance more difficult. Conversely, a large capsulorhexis may not prove to have a sufficient area of anterior capsule under which the cannula can be inserted, and it may hamper endocapsular surgery.

COMPLICATIONS

1. *Anterior capsule tear.* We believe that hydrodissection should not be performed with other forms of capsulotomy (vitrectorhexis, Fugo capsulotomy, radiofrequency diathermy, multipuncture capsulotomy). The edge of the other capsulotomy openings may not be strong enough and may tear during the hydrodissection procedure. Wilson noted that 4/29 (14%) anterior capsule tear occurred during hydrodissection

Table 18.1 MULTIQUADRANT HYDRODISSECTION TECHNIQUE FOR PEDIATRIC CATARACT SURGERY

- Instrument: A 27-gauge modified bent cannula attached to a 5-mL syringe filled with balanced salt solution. A J-shaped Binkhorst cannula can be useful for injection at the subincision site.⁸
 - Technique
1. After performing capsulorhexis, the tip of the cannula is inserted under the anterior capsule for a distance of approximately 1 or 2 mm.
 2. Fluid is gently injected after careful tenting of the anterior capsule. Decompress the nucleus after each wave.
 3. Multiple quadrant hydrodissection, of at least three quadrants, is performed, repeating the technique described above.

(4 during capsulotomy, 7 during lens substance removal, 13 during intraocular lens [IOL] insertion, and 1 during ophthalmic viscosurgical device [OVD] removal).⁹ Of four tears that occurred during hydrodissection, one occurred in vitrectorhexis group, one in Kloti group, and two in Fugo blade group (none in manual capsulorhexis group).⁹ Not all eyes reported in this series received hydrodissection. However, the data suggest that hydrodissection should not be attempted with capsulotomy techniques other than manual continuous curvilinear capsulorhexis.

2. *Viscoelastic escape.* Cohesive OVDs may escape as a bolus if forceful hydrodissection is performed. It is crucial to do a *gentle* hydrodissection when using such OVDs. With any OVD, the surgeon should gently depress the posterior lip of the incision between every one or two injections to permit excess fluid to escape and to allow equilibration of the pressure between the anterior and the posterior chambers.
3. *Iris prolapse.* It is important to remember that excessive hydrodissection from a paracentesis opening can cause increased fluid pressure behind the iris. This increased pressure can lead to iris prolapse through the incisions.

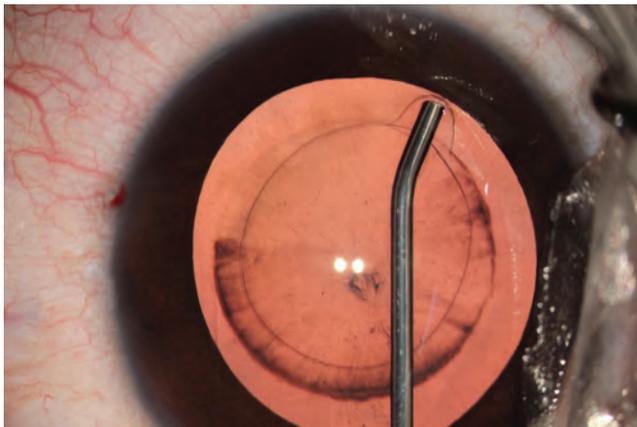


Figure 18.1. Hydrodissection with 27-gauge modified bent cannula.

Table 18.2 SIGNS OF SUCCESSFUL HYDRODISSECTION

- Forward bulge of the nucleus
- Visible presence of a fluid wave: This is considered a definitive sign of successful hydrodissection, but it may not always be visible in pediatric eyes.
- Prominence of the capsulorhexis edge
- Release of trapped fluid from the rhexis margin following decompression of the nucleus

4. *Nucleus prolapse in the anterior chamber.* An unwanted consequence of continuous and forceful hydrodissection is nucleus prolapse into the anterior chamber. Strong corticocapsular adhesions in children require excessive force to cleave apart from the capsule. This results in prolapse of the nucleus more frequently in eyes with pediatric cataract. Immediate decompression of the nucleus after the injection of fluid prevents this complication.
5. *Posterior capsule rupture.* In the absence of a preexisting defect, this is a very rare complication. *Hydrodissection is an absolute contraindication in eyes with a suspected preexisting posterior capsular defect.* Hydrodissection in a total cataract with poor visibility should be done with special caution because the opaque lens substance may camouflage a preexisting posterior capsular defect.

OUTCOME OF HYDRODISSECTION IN EYES WITH PEDIATRIC CATARACTS

We have published results of a prospective, randomized, multicenter clinical trial to evaluate the impact of hydrodissection on intraoperative performance for pediatric cataract surgery. Multiquadrant cortical-cleaving hydrodissection decreases the lens substance removal time, lessens the fluid volume used for lens substance removal, and facilitates lens substance removal in pediatric cataract surgery.⁷ At the end of lens substance removal, residual cortical fibers on the posterior capsule were noted in 12.5% of the hydrodissection group and 22.5% of the no-hydrodissection group. Although this difference was not statistically significant, we believe that it may eventually affect the postoperative outcome with regard to PCO.

In a prospective, randomized trial of adult eyes, we evaluated the effect of multiquadrant cortical cleaving hydrodissection on the development of PCO.¹⁰ We observed that in eyes with multiquadrant cortical cleaving hydrodissection, a significantly lower proportion of the area of the central posterior capsule was involved in the development of PCO as compared to eyes that did not undergo hydrodissection.¹⁰

We found statistically insignificant difference in the incidence of PCO between pediatric eyes that received hydrodissection and those that did not.¹¹ (Although the difference was not statistically significant, we do believe that clinically this difference in PCO is important). There

was also a delay in performing secondary procedures in the hydrodissection group. The lower incidence of PCO and delayed need for secondary surgery in the hydrodissection group indicate that cortical cleaving hydrodissection may postpone but not eliminate PCO development.

SUMMARY

Hydrodissection is a simple and inexpensive procedure that allows fast and easy removal of lens substance during pediatric cataract surgery. Multiple-quadrant hydrodissection is an important component of the removal and washout of equatorial lens epithelial cells. Due to better intraoperative performance, a trend toward lower incidence of PCO, and delay in developing visually significant PCO, we recommend multi-quadrant hydrodissection during pediatric cataract surgery, unless contraindicated due to questionable posterior capsule integrity.

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19

Lens Substance Aspiration

Rupal H. Trivedi and M. Edward Wilson

Lens substance aspiration has probably received the least interest in the discussion of management of pediatric cataracts. Pediatric cataracts are generally soft and can be aspirated easily. However, our target should be not only to *aspirate the lens substance, but to aspirate it thoroughly*. If lens material is left behind, even if it clinically resembles a harmless strand or fiber, it may leave a large number of mitotically active cells.^{1,2} These cells have the potential to grow across the visual axis and cause the proliferative form of visual axis opacification (VAO). Since VAO is one of the most frequent and severe complications in pediatric cataract surgery, meticulous removal of lens substance is a crucial step in the management of pediatric cataracts. The best means of reducing the incidence of this complication is to remove as many of these cells as possible at the time of cataract removal.^{1,2} Amaya and colleagues recommend using the term *phacoaspiration* for children as in most eyes, the lens is not hard enough to require any phacoemulsification power.³ However, the cortex is often gummy and is occasionally difficult to aspirate.

SINGLE-PORT VERSUS BIMANUAL APPROACH

The lens substance can be aspirated through a single port or using a bimanual approach. Single-port irrigation/aspiration (I/A) is an awkward maneuver for subincisional cortex removal.⁴ The technique of bimanual I/A was developed to make cortical removal easier. The technique uses two separate cannulas inserted through two sideport incisions. Anterior chamber (AC) stability during bimanual I/A requires the irrigation system to have a flow resistance lower than that of the aspiration system, which can be accomplished by using cannulas with larger lumen diameters and shorter lengths.⁵ A J-shaped cannula also helps us to approach the subincisional cortex more easily.⁶ Pediatric eyes are especially appropriate candidates for bimanual I/A because

- Separate irrigation and aspiration help to maintain the AC and decrease the fluctuation of the AC. This is especially advantageous in pediatric eyes with low scleral rigidity.
- The bimanual approach helps to achieve thorough removal of lens substance (especially subincisional), which, again, is very crucial when performing pediatric cataract surgery.

MANUAL VERSUS AUTOMATED APPROACH

Pediatric cataracts can be aspirated using the manual or the automated approach. In automated removal, the I/A mode is selected on either a phacoemulsification machine or a vitrector machine.

Machemer et al.^{7,8} developed the vitreous infusion suction cutter in 1970, primarily for removal of the vitreous and vitreous membranes. Pediatric cataracts can be aspirated using the cutter-off position of the vitrectomy machine. The advantage of using the vitrector is that it is possible to perform vitrectorhexis, I/A, posterior capsulectomy, and vitrectomy—all with one instrument (the setting needs to be changed) (see Fig. 19.1). This avoids extra manipulation and repeated entry to and exit from the eye. We use either the Accurus (Alcon Surgical) or the Constellation (Alcon Surgical) for this purpose. We recommend referring to the instruction manual provided by the manufacturer to learn the mechanics of the specific machine.

In cases in which intraocular lens implantation is not intended to be the primary procedure, pediatric cataracts can be removed through one or two very small incisions—wide enough to permit entry of the I/A probe only. Incision width is very important—it should be loose enough to insert the instrument but tight enough to avoid any leakage of fluid. This helps to prevent AC fluctuation.

Maintenance of the AC is critical when removing lens substances. Aspiration of fluid from the AC must be

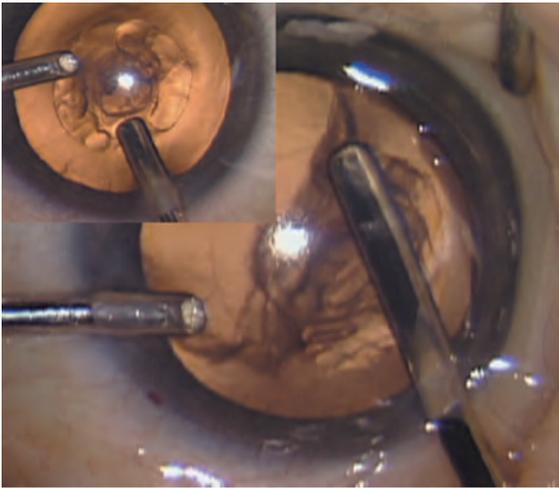


Figure 19.1. After vitreorhexis (inset), without exiting an eye, irrigation/aspiration is being performed with a Nichamin (20 g) cannula and the vitrectomy handpiece set to the cutter-off position.

balanced by adequate infusion. Either a vitrector probe (cutter-off position) or an aspiration probe (e.g., aspiration handpiece Alcon/Grieshaber 170-02; Fig. 19.2) can be used for aspiration maneuvering. We use the irrigation handpiece of a bimanual I/A set. These bimanual I/A handpieces come as reusable or disposable sets. Our preferred gauge for bimanual I/A is still 20 gauge, although we are often using small gauge instruments for vitrectomy (23, 25, 27 gauge). With the 20-gauge instruments, speed and efficiency of bimanual I/A is better compared with the smaller gauges, especially when gummy cortex is encountered. The infusion requirements are usually supplied by a Venturi pump positive-pressure irrigation system. The gravity-fed irrigation systems found on peristaltic pump phacoemulsification machines do not work as well to keep the AC stable when operating soft

pediatric eyes. We highly recommend selecting a Venturi pump machine for this reason.

The addition of 0.5 mL of adrenaline to the infusion bottle (1:1,000 for cardiac use) helps to maintain mydriasis. We have reported a case of bilateral cataract when epinephrine was added to the irrigation fluid in one eye but was inadvertently omitted in the fellow eye. Although the pupil was initially well dilated in both eyes, signs of intraoperative floppy iris syndrome (e.g., iris floppiness, iris prolapse to the incision, and progressive miosis) was observed in the eye not receiving adrenaline in the infusion fluid. We described it as pediatric intraoperative floppy iris syndrome (PIFIS) (see Chapter 49). If the pupil is not dilated well at the beginning of the surgery, aspirate easily reached cortex and give the epinephrine in the irrigating fluid enough time to further enlarge the pupil. Pupil expanders (iris hooks or and iris ring) can be used to assure thorough cortical aspiration if the pupil is not dilated. The possible role of heparin in the irrigating solution is described in Chapter 15.

Before introducing the probe into the AC, it is a good rule of thumb to examine the patency of the cannula and to check that the level of irrigation is adequate. While entering the AC, the irrigation must be on. We prefer to set irrigation to the *irrigation continuous* mode, and the fluid is initially set at 50. This can be adjusted up or down depending on how much leak is present around the instruments and how much aspiration is required to remove the lens cortex and nucleus. The goal is to have a very stable AC without fluctuation. This AC stability reduces iris “bounce,” which then minimizes inflammatory mediators. Iris movement and trauma account for most of the inflammation seen after pediatric cataract surgery. For cortex removal, we place the aspiration handpiece just under the edge of the capsulorhexis and



Figure 19.2. Irrigation/aspiration handpiece (Alcon/Grieshaber).

increase the aspiration until cortex enters the aspiration port. Venturi pump machines allow the aspiration vacuum to be increased using the foot pedal without the need for tip occlusion. The advantage of this approach is that the tip can bring the cortex toward and into the tip even without occlusion. When using a peristaltic pump machine, the pedal is moved to position 2 when the tip is in contact with the cortical material. With peristaltic pump machines, the surgeon must occlude the aspirating orifices to allow an increase in the vacuum level to the maximum preset level. The maximum preset vacuum value is reached with these machines only when the orifice has been occluded.

Insert the aspirating probe with the aspiration port aimed anteriorly. Place the tip of the instrument into the capsular fornix while keeping close to the posterior capsule. This allows the cortex in the capsular fornix to occlude the opening as aspiration begins. Once the cortical material is engaged, the tip is moved slowly toward the center of the pupil to detach the lens substance progressively from the periphery to the center. At times, in pediatric eyes, the cortex does not strip from the lens periphery easily. In this situation, the aspirating tip is turned 90 degrees so that the aspiration port faces the lens equator. It is held in the lens periphery while the aspiration vacuum is increased until the cortex is removed. The aspirating orifice of the instrument must always be under the visual control of the operator. This prevents snagging unwanted tissue (anterior capsule, iris, posterior capsule). First, aspirate free-floating material (if any), then remove stratified material, and, finally, remove material adhering to the posterior capsule (if any). In general, it is easiest to aspirate more accessible sectors first, that is, the temporal, nasal, and inferior sectors, and then proceed to those that are more difficult to reach, that is, subincisional. The subincisional cortex may aspirate more easily if the surgeon switches hands and places the aspiration handpiece through the nondominant sideport incision. If the external diameters of the aspiration and the irrigation handpieces are equal, this reversal of hand positions can be accomplished without creating leakage of fluid around the instruments. An alternative to switching hands is to use the irrigation

handpiece to hydrodissect the stubborn cortex from the lens equator into the center of the pupillary space. This then allows the aspirating handpiece to reach it. Many pediatric surgeons prefer the use of an AC maintainer, such as the Lewicky (Bausch & Lomb E4981 20 gauge and E4984 23 gauge) instrument, rather than a handheld aspiration cannula. These surgeons use their nondominant hand to stabilize the globe with toothed forceps or place both hands on the aspiration cannula. Many of the maneuvers described above required two hands and two instruments working in unison to help open the capsular equator or push gummy cortex into the aspiration or cutting port. This is a matter of surgeon preference.

SUMMARY

We cannot overstate the importance of bimanual I/A technology for lens substance aspiration in pediatric eyes. It is crucial to achieve thorough removal of lens substance using closed chamber technique. Ensuring a tight fit around the instruments as they enter the eye will allow a more atraumatic yet thorough removal of lens cortex and nucleus. Avoiding AC shallowing is essential since this creates iris “bounce” and increases inflammation.

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Posterior Capsulectomy and Anterior Vitrectomy for the Management of Pediatric Cataracts

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Management of the posterior capsule greatly influences the outcome of pediatric cataract surgery (Figs. 20.1 to 20.9). Visual axis opacification (VAO) is rapid and virtually inevitable in young children when adult-style cataract surgery, leaving the posterior capsule intact, is performed. The younger the child, the more acute the problem, because VAO is faster and the amblyogenic effect is greater.

The anterior vitreous face (AVF) is closely linked to the posterior lens capsule and is more “reactive” in infants and young children (Fig. 20.2A and B). The AVF can also serve as a scaffold, not only for lens epithelial cell proliferation but also for metaplastic pigment epithelial cells, exudates, and cells that result from a break in the blood–aqueous barrier. The inflammatory response in small children may be severe, and fibrous membranes may form on an intact AVF, resulting in VAO. The need for posterior capsulectomy and vitrectomy (PCV) has been well recognized in the literature.^{1–6} With the advent of automated vitrectomy techniques in the late 1970s, many pediatric cataract surgeons began to routinely perform PCV with a vitrector.^{1,7–10} As of today, a primary PCV are considered “routine surgical steps,” especially in younger children.

In earlier years, surgeons recommended removing all but the peripheral 2 mm of the posterior capsule to minimize postoperative inflammation and VAO when primary intraocular lens (IOL) is not implanted. There was no consideration given for capsular support of a secondary IOL. However, many of these aphakic children may need secondary IOL implantation later. Thus the modern-day challenge is to leave as much capsule as possible in anticipation for future secondary IOL implantation but not so much that the incidence of VAO is too high. Leaving more capsule remnant at the conclusion of cataract surgery provides a more stable

platform for a sulcus IOL implantation later, and it also increases the likelihood that secondary in-the-bag IOL placement can be accomplished. However, a larger capsule remnant in an infant will also increase the risk of posterior synechia. With older techniques, there was very little capsule remnant for the iris to adhere to. Also, smaller anterior and posterior capsulectomies make thorough cleaning of cortex from the lens equator more of a challenge. If cortex is not cleaned thoroughly, VAO incidence may increase. For an infant, we recommend a 4 to 4.5 mm round vitrectorhexis (or manual continuous curvilinear capsulorhexis [CCC]) of the anterior and posterior capsules combined with thorough cortical cleanup and a vitrectomy. Matching the anterior and posterior capsulectomy size and shape promote a sealed bag that traps any Soemmering ring formation and enhances the chances for in-the-bag secondary IOL placement years later.

Modern high-quality vitrectomy machines, and the improved visualization provided by the optics of the operating microscopes, have allowed surgeons to perform PCV more safely. In addition, better understanding of the importance of tight fitting wounds around instruments that enter the eye of a child has helped reduce the anterior chamber instability that can occur during these surgical maneuvers in young eyes. Despite this, various questions for the surgeon remain. *When should the posterior capsule be opened and when can it be left intact? When does an anterior vitrectomy need to be added to the posterior capsulectomy and when can manipulation of the vitreous be avoided? Is it best to perform posterior capsulectomy and anterior vitrectomy before or after an IOL is implanted? Is the anterior (limbal) or the posterior (pars plana/pars plicata) approach preferred for PCV?* In this chapter we address some of these controversial issues.

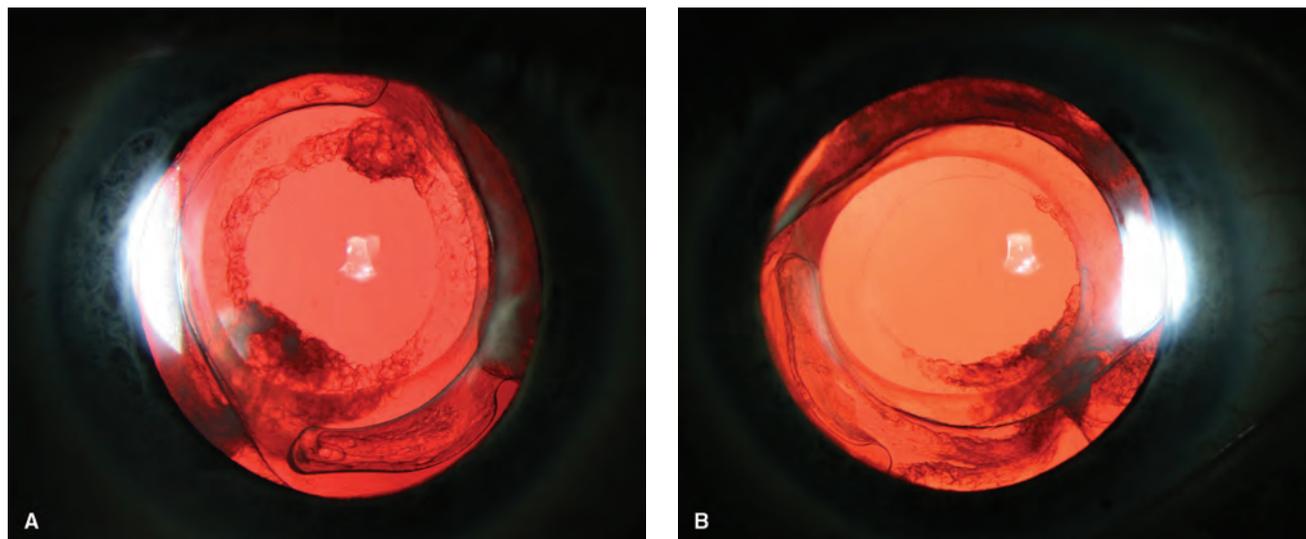


Figure 20.1. Eight-year postoperative picture of a child operated for cataract surgery with vitrectorhexis, posterior capsulectomy/vitrectomy, and SA60AT IOL. **A:** Right eye. **B:** left eye.

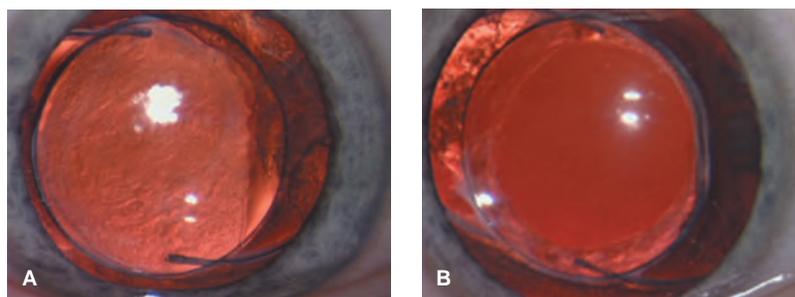


Figure 20.2. A five-and-a-half-year-old child operated for bilateral cataract and IOL implantation. The right eye received posterior capsulectomy (but no vitrectomy), and the left eye received PCV. At 18 months postoperatively, the right eye was noted to have translucent opacification of the visual axis (**A**) and the left eye was clear (**B**).

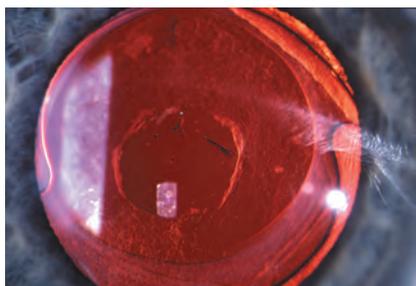


Figure 20.3. Nd:YAG laser has been successfully accomplished in this child's eye returning the visual acuity to 20/20.

Figure 20.4. A 3-month-old child operated for cataract surgery. **A:** Preoperative appearance. **B:** Surgical photos. Note dense plaque on posterior capsule (**inset**). As IOL implantation was not the intended management for this eye, the limbal approach was used. A large posterior capsulectomy was required to encompass the plaque. If IOL implantation had been intended, we would have used the pars plicata approach in this eye (after IOL implantation).

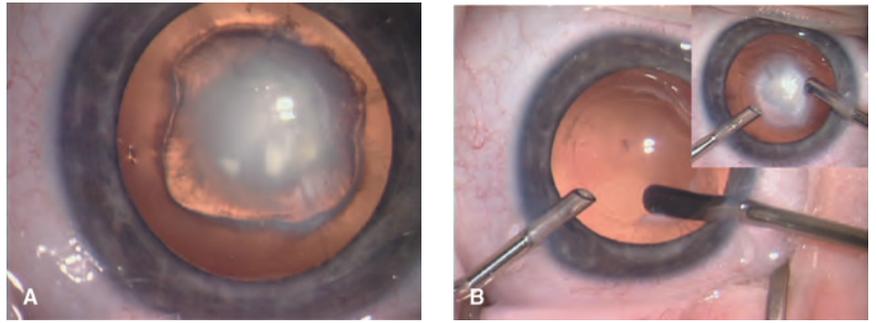


Figure 20.5. Limbal-approach PCV. **A:** Note smaller opening of posterior capsule (than anterior capsule) and **B:** well-centered IOL.

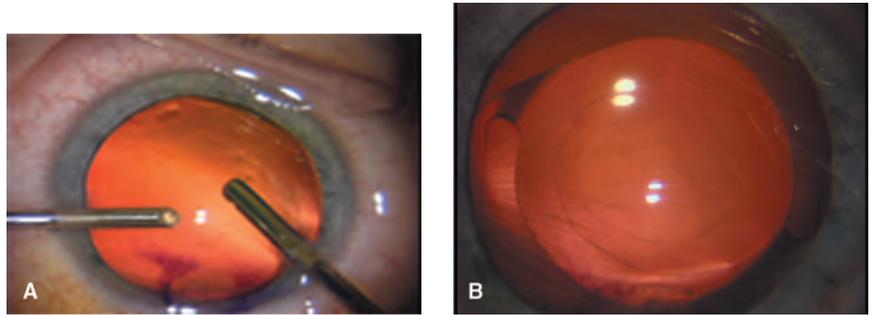
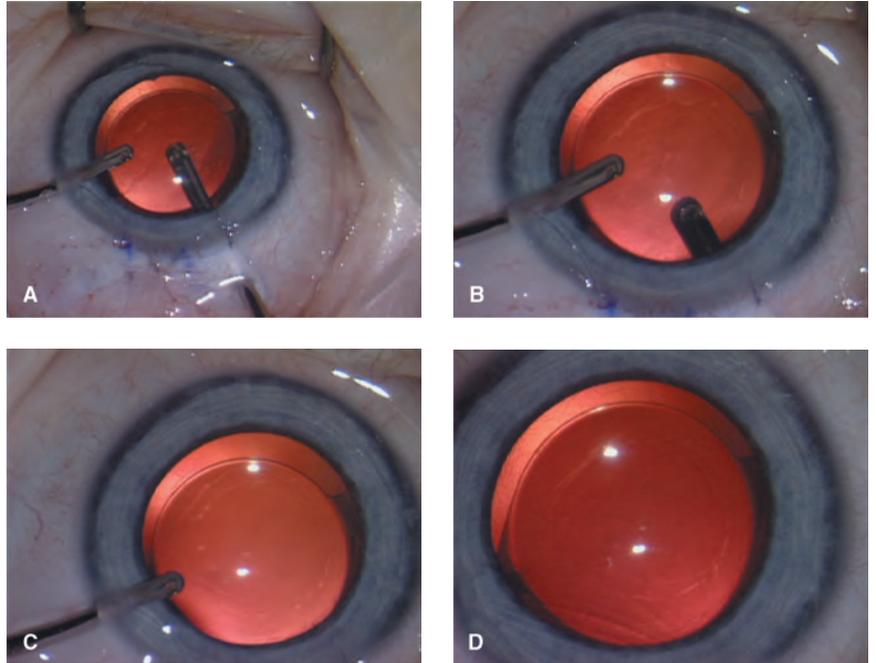


Figure 20.6. Pars plana PCV (after IOL implantation). **A:** The vitrector has been placed 2.5 mm posterior to the limbus. **B:** A round posterior capsulectomy is made with cutting port facing anterior. **C:** Irrigation is from an anterior approach. **D:** The completed surgery reveals a well-centered posterior capsulectomy and a capsular-fixated IOL.



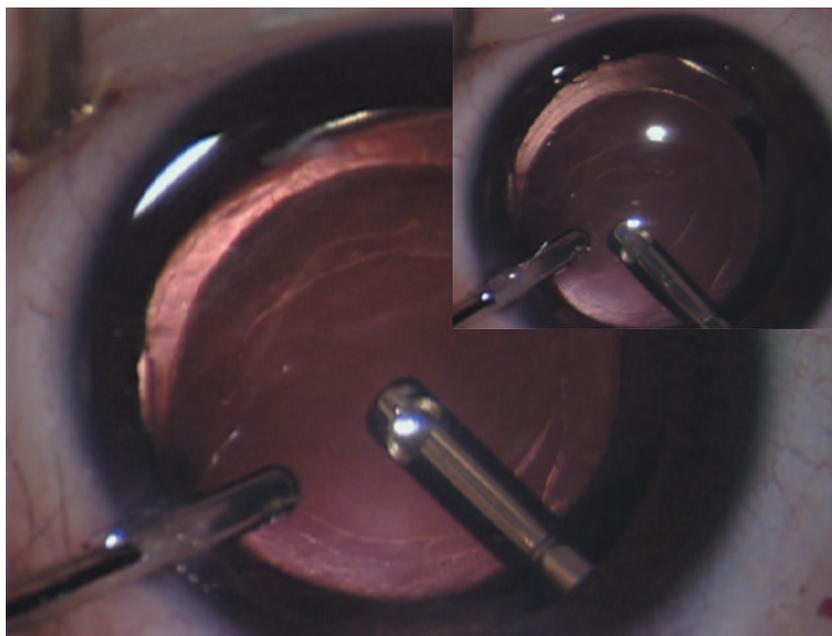


Figure 20.7. Pars plana posterior capsulectomy and anterior vitrectomy after IOL implantation. (Main figure photo shows vitrectomy being performed after completion of posterior capsulectomy) (inset shows capsulectomy being enlarged in a circular fashion by the cutter in the vitrector handpiece).

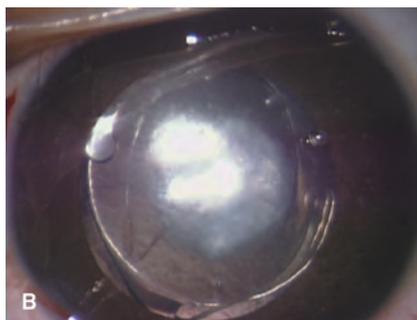


Figure 20.8. In the presence of a dense plaque (A), it is better to perform the PCV after IOL placement (B).

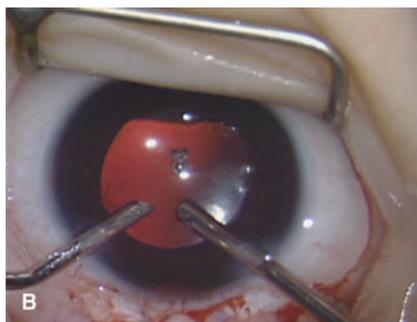
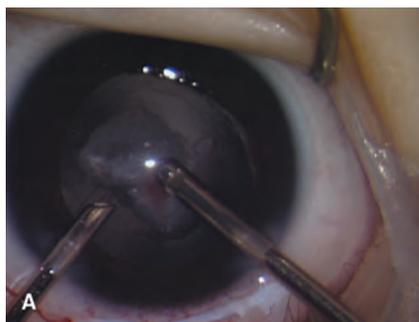


Figure 20.9. Vascularized plaque in an eye with persistent fetal vasculature (A). In this case, the IOL was placed in the ciliary sulcus with the optic captured in the anterior capsulectomy, since most of the posterior capsule had to be removed to clear the plaque. Bleeding from the fetal vessels cleared spontaneously in the first postoperative week (B).

WHEN SHOULD THE POSTERIOR CAPSULE BE LEFT INTACT?

This is perhaps one of the most common questions. The answer depends mainly on the age at cataract surgery but also on the condition of the posterior capsule and the child's presumed cooperation for YAG laser capsulotomy in the future. Proponents of primary PCV argue that performance of these steps is a *necessity* in all children within the amblyopic ages to prevent VAO and worsening amblyopia. However, opponents argue that it is not a safe *choice*. The posterior capsule is a physiologic barrier between the anterior and the posterior segment of the eye and should not be removed. It prevents the vitreous from entering the anterior chamber, and theoretically preserves the ocular anatomic relationship and the blood–aqueous barrier after cataract surgery. Despite this controversy, PCV are viewed by most pediatric surgeons as necessary steps for the best outcome from cataract surgery in young children. Using currently available IOLs, visually significant posterior capsule opacification (PCO) develops most commonly about 18 months to 2 years after surgery when the posterior capsule is left intact in children older than 5 years of age. Therefore the posterior capsule can be left unopened if it is anticipated that the child is cooperative for slit-lamp examination before cataract surgery, which may be an indication that the child would be cooperative for a YAG laser capsulotomy 18 months to 2 years after surgery. One-third or more YAG laser capsulotomies close spontaneously when performed in young children (<6 years of age or so). The practice of leaving the posterior capsule intact in all children at the time of surgery and bringing them back to the operating room 6 weeks later for a YAG laser capsulotomy has also been described, but did not find widespread acceptance because of the recurrence of VAO. Vertically mounted YAG lasers designed for use in the operating room are now difficult to find because of their infrequent use.

Generally speaking, the posterior capsule can be left intact when children present for cataract surgery at age 8 years or older. A primary PCV should be performed even for older children if YAG laser availability is in question, a posterior capsule anomaly (plaque, defect, etc.) is present, or the child is developmentally delayed or uncooperative for YAG laser capsulotomy.

DO I NEED TO PERFORM AN ANTERIOR VITRECTOMY WHENEVER I PERFORM A POSTERIOR CAPSULECTOMY?

In general, in patients up to 5 years of age, this is an *essential* step, since vitreous face opacification is likely to occur if an anterior vitrectomy is not performed. However, in children between 5 and 8 years of age, it is an *optional* step since the chances are better that a posterior capsulotomy

(posterior capsulorhexis) alone may result in a long-term clear visual axis. Nonetheless, surgical or YAG laser may be needed secondary to vitreous face opacification.

HOW SHOULD I TREAT THE POSTERIOR CAPSULE? SO MANY OPTIONS: WHICH TO CHOOSE?

Various options are available for PCV in children. Selection among the various technical alternatives is probably best left to the individual surgeon's preference based on the individual case, available facilities, individual circumstances, and personal experience.

Primary Capsulectomy Versus Secondary Capsulectomy

Posterior capsulectomy may be performed at the time of cataract removal or secondarily. As mentioned previously, most pediatric cataract surgeons now prefer to manage the posterior capsule at the time of cataract surgery. Parks^{1,10} was an early proponent of primary PCV. He stated, “*It should be abundantly clear that the best visual acuity in patients with intractable amblyopia is usually far worse than the best acuity in recovered clinically significant cystoid macular oedema (CME).*” He further asked, “*How can invasion of the vitreous during a secondary procedure to open the translucent membranous posterior lens capsule be less likely to cause CME than opening the posterior capsule during the primary lens surgery?*” CME turned out to occur much less frequently in children after PCV than originally feared. Remarkably, retinal detachment rates have also been very low after this procedure, owing in part to the formed vitreous of the child's eye. With the advent of higher-quality vitrectomy machines and sharp, high-speed cutting handpieces, the procedure is even safer than when Parks and others popularized it. We recommend continuing to treat the posterior capsule as a primary procedure in young children until a method is developed to predictably kill residual lens epithelial cells and reduce the likelihood of postoperative VAO.

Surgical Capsulectomy Versus YAG Laser Capsulotomy

Nd:YAG laser capsulotomy can be used as a primary or secondary posterior capsulotomy (Fig. 20.3). Although YAG lasers were most commonly used to perform secondary capsulotomy, it became possible to perform primary capsulectomy with the advent of YAG lasers mounted vertically and used with patients in the supine position. These instruments are rarely used today and are manufactured in small numbers if at all. Table 20.1 lists the pros and cons of both techniques.

In 1985 Maltzman et al.¹¹ described the outcome of YAG laser capsulotomy for secondary membranes in the

Table 20.1 SURGICAL VERSUS Nd:YAG LASER POSTERIOR CAPSULOTOMY

Surgical Capsulectomy	Nd-YAG Laser Capsulotomy
Reopacification rate is lower because it is possible to treat the AVF also.	Reopacification rate is high because it is not possible to treat a scaffold of the AVF.
Routinely used by most surgeons for primary capsulectomy	Reopacification rate, cost, and availability of YAG laser mounted on operative microscope preclude its routine use in the very young.
If used as a treatment of visually significant PCO—anesthesia consideration should be kept in mind	If used as a treatment of visually significant PCO—anesthesia can be avoided

pediatric population by using a standard office-based laser delivery system. However, this approach cannot be used for young or uncooperative children. A headrest was designed by Kaufman (cited in Ref.¹¹) for patients placed in the lateral decubitus position. This headrest allowed for treatment of anesthetized children, but it has not come into wide use. The Microruptor III laser system allows for controlled laser procedures in supine anesthetized patients and offers the advantage of being familiar to most surgeons.

Atkinson and Hiles,¹² in 1994, reported leaving the posterior capsule intact and performing Nd:YAG capsulotomy under a second general anesthesia in the early postoperative period. Hutcheson et al.¹³ examined the clarity of the visual axis after Nd:YAG laser capsulotomy following cataract extraction and primary IOL implantation in a pediatric population. One eye (3%) in primary surgical PCV experienced postoperative VAO. Thirteen (57%) of 23 eyes in Nd:YAG laser capsulotomy group experienced reopacification, requiring retreatment. Four eyes (17%) treated with Nd:YAG laser required a third treatment. Stager et al.¹⁴ reported rates of PCO following foldable acrylic IOL implantation. The authors noted that 60% of the eyes developed recurrent opacification following Nd:YAG laser treatment in the younger age group (<4 years), which suggested that surgical PCV, rather than YAG laser capsulotomy, may be needed in these young children to keep the visual axis clear.

A transient rise in intraocular pressure has been described after use of a Nd:YAG laser in pediatric patients also.¹¹ The cost and nonavailability of the instrument in children's hospitals or pediatric ophthalmology offices may be additional barriers to the use of the Nd:YAG laser. Many surgeons prefer to perform a surgical procedure initially that will, hopefully, preclude the need for laser capsulotomy.¹⁵

Type of Surgical Opening: Capsulorhexis or Capsulectomy?

Before discussing surgical techniques for opening the posterior capsule, a review of the relevant anatomy and physiology of the posterior lens capsule is appropriate. Mechanical properties of the human posterior capsule differ in several aspects from those of the anterior capsule.¹⁶ Krag and Andreassen reported that the thickness of the posterior capsule ranged from 4 to 9 μm and showed no significant change with age.¹⁶ The posterior capsule was three to five times thinner than the anterior capsule. The mechanical strength of the posterior lens capsule (ultimate strain, ultimate load, ultimate elastic stiffness, ultimate stress, and ultimate elastic modulus) was found to decrease markedly with age.¹⁶ The age-related loss of mechanical strength seemed to begin earlier in the posterior lens capsule than in the anterior lens capsule. Extensibility (ultimate strain) of the posterior lens capsule decreased by a factor of two during the life span, and the forces required to break the posterior lens capsule were found to decrease by a factor of five.¹⁶

The posterior capsule opening can be performed with the manual technique (capsulorhexis) or with a vitrector. Primary posterior capsulorhexis makes it possible to achieve an opening with a strong margin that resists tearing and holds the vitreous in place. It allows safe anterior vitrectomy and prevents uncontrolled widening of the opening. The IOL can be supported over the capsule, and if desired, optic capture can be obtained through the opening. When vitrectomy has been planned in addition to posterior capsulotomy, many surgeons prefer to use the vitrector to cut an opening instead of performing a manual posterior continuous curvilinear capsulorhexis (PCCC). The use of radiofrequency diathermy for this purpose has also been reported in the literature.¹⁷ The radiofrequency bipolar unit is not easily manipulated beneath an IOL and is therefore usually performed on the posterior capsule from an anterior approach prior to IOL insertion. Use of femtosecond laser for posterior capsule opening in children is another viable alternative that is being explored.

Limbal Versus Pars Plicata Approach

Deciding between a limbal and a pars plana/pars plicata approach depends on several factors—condition of posterior capsule (plaque, defect, vascularization), whether or not IOL implantation is intended, etc.^{7–9,18,19} Pros and cons of these techniques are listed in Table 20.2. Inadequate size of the posterior capsule opening (<3 mm) was three times more commonly reported with the limbal route than the pars plana route.²⁰ Fibrin formation was not significantly different between pars plana and limbal route (26 versus 21%; $P > 0.05$).

Our current strategy when we decide to open the posterior capsule is to perform PCV via the pars plicata

Table 20.2 LIMBAL VERSUS PARS PLANA PCV

	Limbal	Pars Plana
Cauterization	Can be avoided	Required
Size	Adequate size may not be obtained since IOL is yet to be implanted (usually)	Larger opening is possible since IOL is already in place
IOL implantation	Relatively difficult. It is common practice to perform limbal vitrectomy before IOL implantation. It is sometimes difficult to implant in soft vitrectomized eyes of young children	Relatively easy to implant in nonvitrectomized eyes with posterior capsule still intact, as these are generally performed after IOL implantation when using this technique
Surgeon comfort and familiarity	Most anterior segment surgeons are more accustomed to limbal approach	Most anterior segment surgeons are not routinely accustomed to pars plana approach
Viscoelastic removal	More difficult since it is done after the posterior capsule is opened	Easier since the removal is usually done after IOL insertion and before posterior capsulectomy
Posterior capsule plaque	Easy but posterior opening may need to be large and this would complicate in-the-bag insertion of IOL	Easy to insert IOL with plaque still present, and removing posterior capsule through the pars plana—plaque makes posterior capsule easy to see under the IOL and opening can be safely made larger
Traction of vitreous base	If a vitreous strand remains attached to anterior wound, traction on retina is increased	Less likely to result in vitreous attached to anterior wound

(irrigation via the limbal paracentesis and vitrector through the pars plana) after IOL implantation whenever an IOL is being implanted. A pars plana approach to anterior vitrectomy minimizes vitreoretinal traction, reduces the need to manipulate the iris, and produces less corneal endothelial damage.

If surgical strategy is to open posterior capsule without vitrectomy, a limbal approach manual PCCC before IOL implantation is preferred. In eyes with preexisting posterior capsule defect, the limbal approach has been used more often. A limbal approach is used in children who are being left aphakic (Fig. 20.4). A limbal approach is also used when the visualization of intraocular contents is anticipated to be marginal. This occurs when operating in remote areas, when the operating microscope lighting is less than ideal or when corneal clouding is present. In these instances, the pars plicata approach after IOL implantation may be more difficult since it requires ideal lighting and a clear cornea. In some developing world setting, some surgeons use limbal approach if the IOLs available are large and stiff and the microscope lighting marginal. IOL implanted in the sulcus (with or without optic capture) may avoid the problem of implanting it in the bag in these soft vitrectomized eyes. Miochol at the end of the procedure may help prevent pupil capture when using this technique.

Before Versus After IOL Implantation

The IOL can be implanted before or after PCV.^{6,21} The advantage of the latter approach is that the IOL can be more safely fixed in the desired plane compared to placing an IOL into a soft vitrectomized eye with an already

opened posterior capsule (Table 20.3). In infantile eyes, the rigidity of the sclera is very low. IOL implantation is technically difficult in these eyes with low scleral rigidity. The thickness of the infantile sclera is 0.4 times, and the coefficient of stretching 0.6 times, that of the adult.²² Additional vitrectomy adds to the difficulty as eye becomes more softer. The common practice is to do the posterior capsulectomy before IOL implantation if the limbal approach has been used (Fig. 20.5). However, if the pars plana/plicata approach is used, in general, the PCV are performed after IOL implantation (Fig. 20.6 to 20.8). A limbal-approach retropseudophakic vitrectomy has also been reported.²³ As mentioned before, our current most commonly used approach is to implant the

Table 20.3 POSTERIOR CAPSULECTOMY BEFORE OR AFTER IOL IMPLANTATION?

Before IOL Implantation	After IOL Implantation
If capsulectomy extends during procedure, it may hamper achieving successful in-the-bag fixation.	Stable IOL fixation can be achieved if anterior capsule is intact.
IOL implantation becomes difficult in soft nonvitrectomized eyes.	IOL implantation is relatively easy.
PCV can be achieved under better visibility.	Visibility may be poor.
Generally aimed at a smaller opening to facilitate IOL implantation.	A wide posterior capsule opening can be achieved.

IOL first. We then either leave the capsule intact or we perform PCV. There are times when we perform a manual PCCC without vitrectomy in an older child prior to IOL implantation. In these cases, the PCCC is smaller (3.5–4.0 mm) than the typical pars plana capsulectomy, which makes it easier and safer to implant the IOL after the capsule is opened. In the presence of a large posterior capsule plaque, large posterior capsule opening is needed to encompass the plaque, which may hamper secure in-the-bag placement of IOL, if IOL is to be implanted after PCV. Even for surgeons performing IOL implantation after PCV as a routine, it is advisable to implant an IOL before opening the posterior capsule when such a large opening in the posterior capsule is anticipated (see Fig. 20.8).

Architecture of the Posterior Capsule Opening: Size, Centricity, and Shape

During the early 1990s, many pediatric ophthalmologists recommended removing all but 1 or 2 mm of the peripheral posterior lens capsule during pediatric cataract surgery. Caputo et al.²⁴ described a method of using a small central posterior capsulectomy for pediatric cataracts that is designed to eliminate VAO and to keep open the option of later secondary implantation of a posterior chamber IOL. However, a small opening increases the chances of closure. Ideally one should aim to achieve an optimum-size (ca. 1–1.5 mm smaller than the IOL optic) centric, circular opening in the posterior capsule. The size of the PCCC is very important when attempting to capture the optic of the IOL. The optimum size (neither too small, which makes it difficult to capture the optic, nor too large, in which case the optic may not remain captured) is a prerequisite for ideal and stable capture.

Does No-Suture Vitrectomy Technology Have a Role?

Sutureless pars plana vitrectomy through self-sealing sclerotomies has been reported in the literature.^{25–27} Kwok et al.²⁸ described the ultrabiomicroscopy of conventional and sutureless pars plana sclerotomies. They concluded that ultrabiomicroscopy showed no difference in the amount of visible vitreous incarceration in conventionally sutured versus sutureless sclerotomies. However, several complications have been reported with this technique, including wound leakage, extension, dehiscence, hemorrhage, vitreous and/or retinal incarceration, retinal tear, and retinal dialysis. Difficulty with passage of instruments has also been observed when tunnels are used. In addition, the scleral tunnel technique still requires conjunctival dissection, which often requires suturing. Fujii et al.²⁷ reported a 25-gauge vitrectomy system that allows for completely sutureless vitrectomy surgery, which can be performed without scleral or conjunctival suture.

Lam et al.²⁹ evaluated sutureless pars plana anterior vitrectomy through self-sealing sclerotomies in children with thick posterior pseudophakic membranes (secondary capsulectomy and vitrectomy). They commented that although the scleral rigidity in children is lower, the self-sealing effect of this technique is good, with the integrity of the eyes well maintained, and that this technique can be considered in selected cases. Our approach has been to suture every sclerotomy in a pediatric eye. We have witnessed significant gape of unsutured scleral wounds in children. See Chapter 21 for more advice about vitrectomy specifics for the pediatric eye.

Are Special Aids or Techniques for Visualization Needed?

Use of dye for posterior capsulorhexis has been described in the literature.^{30–32} However, dyes have been used less frequently for the posterior than for the anterior capsule. In the experimental studies, Pandey and associates demonstrated that indocyanine green (ICG) and trypan blue dyes can be successfully used to stain the posterior lens capsule to enhance visualization while learning and performing posterior capsulorhexis.³⁰ Posterior capsule staining can be done by instilling one drop of the dye solution into the capsular bag, after cortical cleanup. After 60 to 90 seconds, the excessive dye is washed out and posterior capsule can be opened. Intraoperative use of trypan blue to stain lens epithelial cell during pediatric cataract surgery has also been reported.³³

Triamcinolone injection into the anterior chamber provides the anterior segment surgeon a means for localizing and identifying vitreous gel (Chapter 22).³⁴ Clear visualization of the vitreous gel allows thorough removal of the prolapsed vitreous and alerts surgeons to residual strands of vitreous that might have gone unnoticed. It also allows surgeons to observe vitreous behavior so that they can avoid maneuvers that increase vitreous traction or prolapse.

How Is the End Point of the Vitrectomy Defined? How Much Vitreous Should Be Removed?

These questions have not been answered in a scientific manner. Sufficient vitreous should be removed centrally so that the lens epithelial cells cannot use the vitreous face as a scaffolding to create VAO. Any vitreous that tracks forward past the plane of the posterior capsulectomy needs to be removed. VAO after PCV is often blamed on an inadequate posterior capsule opening or an inadequate vitrectomy. These assertions have not been verified scientifically. In babies we aim to remove approximately one-third of the vitreous volume. In older children, less vitreous removal may be adequate. Most surgeons advise a “generous” anterior vitrectomy without placing the vitrector so deep that visualization would require a

posterior vitrectomy viewing attachment to the operating microscope. As mentioned before, triamcinolone can be injected into the anterior chamber to aid in visualization and thorough removal.

TECHNIQUE

Posterior Capsulorhexis

After aspiration of the lens matter with two-port automated irrigation/aspiration, the capsule bag and anterior chamber are filled with high-viscosity sodium hyaluronate (Healon GV[®] or Healon5[®]) and posterior capsulorhexis is initiated. The central posterior capsule is thinnest. Grasping the capsule is difficult at the initial stage. Therefore, initiation of the puncture and flap is better done with the help of the vertical element of the cystitome. The 26-gauge cystitome needle descends on the capsule at a slant. It engages the central capsule, lifts it up toward the surgeon, and, at the same time, initiates the puncture. A small flap is created by pushing the margin of the puncture inferiorly. High-viscosity sodium hyaluronate is injected through the puncture between the capsule and the vitreous face. This should be done slowly. Forceful injection through the tear can extend it toward the periphery. Then additional viscoelastic should be placed over the posterior capsule surrounding the puncture. This flattens the central posterior capsule and reduces potential peripheral extension. The flap is then held with capsulorhexis forceps, and posterior capsulorhexis is accomplished. Frequent grasping and viscomanipulation of the flap is often helpful. As mentioned before, posterior capsulorhexis is usually performed prior to IOL implantation but can also be performed after placement of an IOL. This after-implantation option ensures IOL fixation in the desired plane. However, performing PCCC can be technically more challenging when done from an anterior retro-pseudophakic approach, especially if the anterior CCC is somewhat small.

Posterior Capsulectomy and Vitrectomy

Machemer et al.³⁵ developed the vitreous infusion suction cutter in the early 1970s. The advent of the vitrectomy machine reduced the rate of retinal detachment as a late complication of pediatric cataract surgery. Similar to an anterior vitrectorhexis, a Venturi pump system is recommended, as it cuts vitreous and the posterior capsule more safely and efficiently than a peristaltic pump machine. We recommend that readers follow the instruction manual of the manufacturer for using a specific machine and setting.

The unique surgical anatomy of infant eyes requires modification of standard vitreous surgery techniques. In addition to its small size, the infant eye differs from that of the adult with respect to the spatial relationship of various ocular structures. A major anatomic constraint is

imposed by the relative size of the pars plana. In the newborn eye, the pars plana region is incompletely developed so that the anterior retina lies just behind the pars plicata. To avoid iatrogenic retinal breaks, entry incisions for vitreoretinal surgery, therefore, are made through or anterior to the region of the pars plicata.³⁶ We recommend an entry 2.0 mm posterior to the limbus in the first year of life, but we do not perform surgery prior to 30 days of age for term infants. Premature infants are operated slightly later and have anterior limbal-approach surgical wounds. Transillumination can aid in identifying the location of the ciliary body.

Aiello et al.³⁷ noted that the temporal ciliary body is longer than the nasal ciliary body in the pediatric age groups as well as in adults. These authors noted that the pars plana was 2.2 and 2.5 for the nasal and temporal aspect at <6 months of age, 2.7 and 3 at 6 to 12 months, 3.0 and 3.1 at 1 to 2 years of age, and 3.2 and 3.8 at 2 to 6 years of age. They estimated that the dimensions of the ciliary body in the vertical meridian fall between the measurements of the nasal and the temporal ciliary body, as observed in adults.

Good success has been reported with two-port phakic vitrectomies in infants when the sclerotomies were made through the iris root at 0.5 mm posterior to the limbus.³⁶ The most rapid phase of pars plana growth occurs between 26 and 35 weeks of gestation. At full term, the mean pars plana width is 1.87 mm (range, 0.9–2.8 mm). Not until 62 weeks after conception does the pars plana attain a width >3 mm. Therefore, anatomically, it may be that a pars plana vitrectomy can be safely performed only after 62 weeks postconception. Prior to that, pars plicata or iris root sclerotomy is done.

Reporting the outcome of vitrectomy for infantile vitreous hemorrhage, Ferrone and de Juan³⁸ noted that two eyes had a retinal dialysis. The authors hypothesized that a potential reason for these dialyses was that the sclerotomies were 2 mm posterior to the limbus. This may be too far posterior in these young, developing eyes. In very young patients, it may also be that the pars plana is not wide enough to perform a pars plana vitrectomy safely. The two eyes with dialyses in this series were only 44 and 47 weeks postconceptional age. Hairston et al.³⁹ reported that the dimension of the pars plana is correlated closely with axial length and postconceptional age. A linear relationship between pars plana width and axial length existed once the axial length reached 12 mm. In cataractous eyes, the axial length may be even shorter, so we can assume that the pars plana is shorter also.

Considering the above, our current approach is to enter 2 mm (depending on the axial length of the eye) posterior to the limbus in patients <1 year old, 2.5 mm posterior in patients 1 to 4 years old, and 3 mm posterior in patients >4 years old. The entry angles should be toward the center of the vitreous when entering at 3 mm;

however, it should be adjusted toward the optic disc when a more anterior entry site is used to avoid the equator of the capsular bag. Meier et al.⁴⁰ have recommended distances of 1.5, 2.0, and 2.5 mm at <3, 3 to 6, and >6 months of age, respectively. As stated above, in the eye of the mature neonate, the pars plicata of the ciliary body is almost fully developed, whereas the pars plana is hardly established. In the absence of the pars plana, ideally we should use the term pars plicata PCV in infants instead of the so-called pars plana used in literature.

While performing vitrectomy during pediatric cataract surgery, the aim is to remove the central anterior vitreous without attempting to remove all of the peripheral or posterior vitreous. For this limited vitrectomy, we perform the procedure through two ports. Separating the irrigation from the cutting and aspiration reduces hydration of the vitreous.⁴⁵ Modern vitrectors have higher cut rates, and most now recommend that the vitreous be cut at the highest cut rate available on the model being used. On the Accurus machine, we have traditionally used an irrigation rate of 30 to 50 and a cutting rate of ≥ 800 cuts/min. With newer machines like the Constellation, much higher cutting rates (7500 cuts/min) are used. Retinal traction at the time of vitrectomy is reduced by high cutting rates. In children, even small sclerotomy incisions are best closed with 8.0 or 9.0 vicryl suture. Buckley et al.²¹ have mentioned a preplaced 8-0 Vicryl suture passed across the sclerotomy site to facilitate closure after vitreous removal.

Posterior Capsule Plaque

It is not uncommon to observe a plaque on the posterior capsule.^{41,42} This may be peeled free with a cystitome under OVD. It is also possible to do a large PCCC to encompass the plaque or to do a mechanized PCV large enough to fully remove the plaque. Other pathology associated with the posterior capsule such as a preexisting defect or persistent fetal vasculature (see Fig. 20.9) is discussed in other chapters.

Secondary Surgical Posterior Capsulectomy

Removal of the center of the posterior capsule does not guarantee that secondary cataracts will not regrow, especially in young infants. When this occurs, an irrigation cannula is usually placed into the anterior chamber through a paracentesis incision. The PCV are usually done through a pars plana entry if an IOL is present and is within the capsular bag. Sometimes, the opacification can be easily reached from the limbal position by placing the vitrector posterior to the edge of the anterior capsulorhexis and then posterior to the IOL optic. Occasionally, a combined approach (anterior and posterior) may be needed to remove reepithelialized cortex from both sides of the IOL. When the IOL has been placed in the ciliary sulcus, recurrent opacities can usually be managed from the limbal approach since placing

the vitrector posterior to the IOL optic is more easily done than when in-the-bag fixation is present. In contrast, the pars plana approach is recommended for posterior opacities when an IOL is in the capsular bag to avoid any risk of displacing it from its often “shrink-wrapped” location within the capsular leaflets.

SUMMARY

Posterior capsulectomy and anterior vitrectomy are essential surgical steps in the management of pediatric cataract surgery. Treatment of the posterior capsule often determines the ultimate outcome of pediatric cataract surgery. Various alternatives have been suggested in the literature to achieve this goal. We need to choose the way in which the posterior capsule is managed for each patient based on the multiple factors discussed in this chapter.

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Current Vitrectomy Concepts and Techniques for the Pediatric Anterior Segment Surgeon

Steve Charles

Amblyopia and aphakic correction are long recognized^{1,2} as the major issues in pediatric cataract surgery. Fortunately, vitreoretinal complications are uncommon after cataract surgery in children. However, when these complications occur, visual consequences can be devastating. Attention to modern vitreoretinal surgical principles can help the pediatric cataract surgeon minimize risks to the retina.

Endophthalmitis, while catastrophic, appears to be extremely uncommon³⁻⁷ in pediatric cataract surgery, with the best current estimate being <0.02%.⁶ When endophthalmitis is suspected, treatment decisions mirror those made in adult endophthalmitis, and there is no apparent etiologic disparity between children and adults compared to the endophthalmitis vitrectomy study.^{6,8}

Cystoid macular edema (CME) occurs with unknown frequency after pediatric cataract extraction, in part due to the difficulty in detecting CME in the pediatric patient because of the challenges of performing macular examination, the inability to visualize CME with the indirect ophthalmoscope or RetCam, the sedation issues associated with fluorescein angiography, and the inability of children to position for ocular coherence tomography (OCT). Reports attempting to assess CME⁹⁻¹³ have concluded that its occurrence in the pediatric population appears to be infrequent. If detected and visually significant, the treatment should therefore parallel guidelines for adult pseudophakic CME,^{14,15} including topical corticosteroids and nonsteroidal anti-inflammatory medications.

Retinal detachment can occur decades after what is thought to be uneventful cataract surgery.¹⁶⁻²¹ The frequency of retinal detachment following pediatric cataract surgery varies among reports between 0.5% and 5%^{1,2,21-25}; however, there is a paucity of recent prospectively analyzed data. Nevertheless, choices made in the

intraoperative management of vitreous may alter the likelihood of late retinal detachment. This chapter guides the surgeon toward appropriate techniques to help minimize the risk of intraoperative retinal injury and aids in the timely and appropriate diagnosis of posterior segment pathology following pediatric cataract surgery.

THE IMPORTANCE OF INTRAOPERATIVE VITREORETINAL TRACTION

Many surgeons believe that as long as there is no vitreous in the wound at the end of the case, vitreoretinal traction is not a problem; to the contrary, acute intraoperative traction is a major cause of retinal breaks and therefore retinal detachment. Anterior vitrectomy is performed close to the vitreous base, the region of the strongest vitreoretinal adherence^{26,27} (estimates indicate retinal tensile strength is 1/100 that of central retina, with no data on pediatric population).

Using the highest possible cutting rate reduces pulsatile vitreoretinal traction by reducing pulse flow.²⁶ Always use proportional (linear) vacuum, not 3-D or dual linear control in order to maximize the cutting rate. Using the lowest effective vacuum or aspiration flow rate reduces acute vitreoretinal traction. Venturi pumps are better than peristaltic pumps for anterior vitrectomy because they do not produce pulsatile vacuum. Never withdraw the vitreous cutter while aspirating; pullback force is additive to aspiration force and causes acute vitreoretinal traction as well as chamber shallowing.²⁶ There should be a tight fit around the infusion and cutter port wounds to avoid chamber instability and shallowing, which occur in these soft eyes when aspiration is increased.

Bimanual infusion is preferable to so-called “dry vitrectomy”; vitreous removal is more complete, and miosis as well as striate keratopathy associated with a soft eye

is avoided. An anterior chamber maintainer can be very effective but care must be taken to avoid contact with the endothelium. An angulated, blunt 23-gauge infusion cannula is preferable because it permits tool exchange.

The surgeon should never attempt to aspirate liquid vitreous with the irrigation/aspiration (I/A) tip or any other aspiration tool; excessive vitreoretinal traction will always occur. Similarly, a surgeon should never use the I/A in vitreous. Although phacoemulsification is rarely used in children, it is important to be aware that aggressive aspiration as well as ultrasonic energy emulsifies hyaluronan (hyaluronic acid) giving the false appearance of vitrectomy but does not shear vitreous collagen. I/A is best done with the vitreous cutter; if vitreous is encountered, it will be sheared thereby greatly reducing vitreoretinal traction. Always inject an ophthalmic viscosurgical device (OVD) before withdrawing a phacoemulsification probe to avoid fluid surge from the wounds and acute vitreoretinal traction.

Cellulose sponges should never be used for removal of vitreous or testing for vitreous; wicking as well as lifting to cut with scissors produces marked intraoperative vitreoretinal traction. Using preservative-free triamcinolone (Triesence, Alcon, Ft Worth, TX) for particulate vitreous marking enables more complete vitrectomy without using cellulose sponges to test for vitreous. Postop vitreous wick should be removed immediately; it may lead to endophthalmitis, retinal detachment, and CME. Wound sweeping should likewise be avoided; although this technique can remove vitreous to the wound, it may also induce acute vitreoretinal traction.

Bimanual technique with separate cutter and infusion is far better than using the coaxial infusion sleeve because turbulent flow prolongs vitrectomy time, increases fluid throughput, and damages the corneal endothelium.

The vitreous cutter should never be used through a limbal wound >1 mm diameter; it increases the chances of iris and vitreous prolapse; infusion should be performed with an angulated cannula through one sideport incision and the vitreous cutter in a second sideport or the pars plana. Bimanual vitrectomy has the same advantages as has bimanual I/A; less turbulence results when using widely separated infusion and aspiration,²⁶ and the tools can be alternated between ports for better access to lens material and vitreous.

A pars plana approach to anterior vitrectomy eliminates all vitreoretinal traction without the very unsafe use of cellulose sponges or sweeping the wound. It allows removal of residual lens cortex without vitreoretinal traction, reduces the need to manipulate the iris, and produces less corneal endothelial damage.

Postoperative CME (Irvine-Gass) is an anterior segment inflammatory process mostly caused by iris trauma, not vitreous removal; total posterior vitrectomy rarely if ever causes CME. Although it is argued by some that

pseudophakic CME does not occur as often in children as adults,⁹⁻¹³ there are no accurate, current data on this subject. Cellulose sponges traumatize the iris because they swell when they imbibe liquid vitreous. Anteroposterior vitreous motion during repeated chamber collapse, residual lens material, possibly agents added to the infusion fluid, and, theoretically, some OVDs may contribute to inflammation and therefore increase the risk of CME.

Coaxial I/A should always be used, and the surgeon should avoid push-pull I/A (stepwise irrigation followed by aspiration) because anterior chamber shallowing causes acute vitreoretinal traction. Anterior chamber stability is crucial in I/A; children seldom have a posterior vitreous detachment, and anterior movement of the lens-iris diaphragm results in anterior movement of the vitreous and acute vitreoretinal traction. Children have less scleral rigidity, which results in greater anterior chamber collapse when the chamber shallows or an intraocular lens (IOL) is inserted. OVDs can help maintain the anterior chamber and reduce anterior movement of the vitreous.

POSTOPERATIVE VITREORETINAL TRACTION

Retained lens cortex can result in fibrous proliferation of lens epithelial cells, a Soemmering ring, and posterior synechia. Adherence of this fibrous complex to peripheral vitreous, which is attached in turn to the vitreous base where the retina has 1/100 the tensile strength of central retina, predisposes to late retinal detachment from subsequent surgical procedures such as discission of the capsule and IOL insertion as well as seemingly inconsequential blunt trauma. Careful removal of as much lens cortex as possible reduces inflammation and fibrous proliferation.

RETINAL EXAMINATION

The retina should always be examined with a 20-diopter lens and the indirect ophthalmoscope before pediatric cataract surgery; sedation is not required in most instances. The indirect ophthalmoscope should be used on the operating table to examine the fundus if the red reflex is abnormal at the time of surgery, if intraoperative complications occur, or anytime the fundus cannot be seen preoperatively, but preferably in all cases.

B-scan ultrasound should ideally be performed by the surgeon, not a technician, if the fundus cannot be seen preoperatively. B-scan ultrasound examinations are dynamic, and the surgeon must observe the study in motion rather than merely looking at selected freeze-frame images saved by the technician. OCT should be performed preoperatively if the lens is clear enough and the child is able to cooperate.

Yearly examination under anesthesia should include not just keratometry, axial length, and intraocular pressure

(IOP) but also careful dilated retinal examination using indirect ophthalmoscopy and scleral depression. If identified, retinal breaks can be treated with the laser indirect ophthalmoscope (LIO).

RETINAL DETACHMENT REPAIR—VITRECTOMY TECHNIQUES

In the author's opinion, scleral buckling procedures for pediatric retinal detachment repair should be avoided due to significant damage to the extraocular muscles and induced axial myopia. These features may be particularly important considering methods for following the evolution of pediatric glaucoma via axial length measurement, the preservation of tissue for the potential need for filtering surgery, and amblyopia management. Twenty-seven- and twenty-five-gauge transconjunctival vitrectomy cause much less postoperative discomfort and conjunctival damage than do 20-gauge or even 23-gauge technology. Many of the pediatric cataract patients will already have or will later develop glaucoma, and some will require filtering procedures; preservation of as much conjunctiva as possible is essential. Sutured-on fundus contact lenses should never be used because of conjunctival damage. Although silicone oil is said to reduce postoperative positioning problems in children, silicone oil floats superiorly and is therefore ineffective for inferior retinal breaks. Silicone oil can be left in place in older adults if an in-the-bag IOL has been implanted that may prevent silicone oil from entering the anterior chamber. Unfortunately, small droplets of emulsified silicone oil will still reach the trabecular meshwork over extended periods of time causing emulsification glaucoma, which is very difficult to manage. Using SF₆ gas for inferior breaks requires prone positioning that can be accomplished with education of the parents; children sleep face down better than do adults.

TRANSLIMBAL LENSECTOMY—VITRECTOMY

Indications for translimbal lensectomy–vitrectomy include traumatic cataract, abnormal pars plana anatomy, persistent fetal vasculature (PFV), scleral laceration in ciliary body region, pars plana region, and severe pars planitis. The advantages of a translimbal approach are prevention of inadvertent suprachoroidal infusion, subretinal infusion, and introduction of instruments into the subretinal space. The disadvantages include corneal striae and poor visualization of retro-iris anatomy and peripheral retina because iris contact causes miosis.

Using the vitreous cutter for I/A is particularly important in traumatic cataracts because of the high likelihood of encountering vitreous due to defects in the lens capsule. It is crucial to avoid vitreoretinal traction in the setting of

trauma because traumatic retinal injury and retinal dialysis may be present and undetected because of the cataract.

LENS MANAGEMENT IN PEDIATRIC RETINAL DISEASES

Retinopathy of Prematurity

Although many surgeons advocate lens-sparing vitrectomy for stage 4A retinopathy of prematurity (ROP) (peripheral detachment only), the author disagrees with this approach and emphasizes that there is no randomized trial evidence to support this indication. Some stage 4B (macula detached, subtotal detachments) can be operated with lens-sparing technique. Recent stage 5 wide funnel cases not requiring epiretinal membrane surgery can be successfully operated, but lens sparing is not always possible. If the lensectomy is performed, it should be done through the pars plicata and all capsule removed with forceps to prevent fibrous scarring on the posterior surface of the iris, ciliary body, and pars plana.

Persistent Fetal Vasculature

Elongated ciliary processes, smaller corneal diameter, and persistent elements of the hyaloid artery system are key elements of the diagnosis of PFV, formerly known as PHPV (persistent hyperplastic primary vitreous). These cases are usually unilateral. Mild PFV cases, without stretched ciliary processes, can be approached much like other infantile cataracts. However, most PFV cases require pars plicata lensectomy that should be combined with removal of the entire capsule and scissors sectioning between each ciliary process; this technique was developed by the author to facilitate ocular growth and prevent phthisis. If a peripapillary traction retinal detachment is present, scissors delamination of tissue extending from the stalk onto the retinal surface is necessary, not just sectioning of the stalk.

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Intraocular Triamcinolone in Pediatric Cataract Surgery

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Posterior capsulectomy with or without vitrectomy is an essential surgical step in the management of pediatric cataract surgery. Pediatric cataract surgeons are more likely to encounter vitreous as compared with adult cataract surgeons. When the surgical plan is manual primary posterior continuous curvilinear capsulorhexis (PCCC) without vitrectomy, it is important to detect if the anterior vitreous face (AVF) is intact or disturbed. If the AVF is disturbed, an anterior vitrectomy is a necessary surgical step. When the surgical strategy is posterior capsulectomy with vitrectomy, it is vital to visualize vitreous to perform a thorough vitrectomy and to identify residual vitreous fibers. Any residual vitreous strands need to be removed as they can lead to retinal traction as well as irregularities in the pupil, intraocular lens (IOL) displacement, and visual axis opacification (VAO). This may lead to complications such as glaucoma, and retinal detachment. Thus, identification of vitreous plays a crucial role in ensuring the successful outcome of pediatric cataract surgery.

Although partial removal of the posterior capsule and anterior vitreous is commonly performed during pediatric cataract surgery, surgeons often find it difficult to visualize vitreous and define the end point of vitrectomy. Vitreous gel, transparent by design, is virtually invisible under the operating microscope. Vitreoretinal surgeons have used intravitreal injections of triamcinolone acetonide (TA) to visualize the vitreous. TA is a synthetic insoluble corticosteroid. In ocular surgery, TA functions like a dye to stain the vitreous, mainly because of crystal deposition.¹ During pediatric cataract surgery, intracameral TA can be used to highlight vitreous known to be present (e.g., ectopia lentis), to check for suspected vitreous (e.g., ectopia lentis, history of ocular trauma, AVF disturbance), to confirm that all the vitreous has been cleared (e.g., after vitrectomy) from the anterior chamber, or as

therapeutic agent to decrease inflammation. We have reported TA-assisted pediatric cataract surgery describing TA intraoperatively for better visualization of vitreous^{2,3} and suggesting its beneficial effect on postoperative inflammation.⁴ In addition, use of intracameral TA is also reported for both adult⁵ and pediatric cataract surgery to help decrease postoperative inflammation.⁶

TECHNIQUE

TA is commercially available under various brand names—for example, Triesence® (Alcon Labs, Fort Worth, TX, 40 mg/mL); Kenalog (Bristol-Myers-Squibb, Peapack, NJ, 40 mg/mL); Trivaris (Allergan, Irvine, CA, 80 mg/mL); Aurocort (AuroLab, India, 40 mg/mL), etc.^{1,2} Triesence® is US FDA approved for intraocular use. It is indicated for the treatment of uveitis and ocular inflammatory conditions unresponsive to topical corticosteroids. Triesence® suspension is also indicated for visualization during vitrectomy (*package insert*, Triesence®). It is preservative free (does not contain benzyl alcohol). Recommended dose for visualization is 1 to 4 mg (25–100 µL of 40 mg/mL suspension) administered intravitreally. It is provided as a single-use 1-mL vial containing 40 mg/mL of TA suspension.

We recommend using preservative-free TA suspension. If preservative-free suspension is not available, a method of removing the preservative has been reported in the literature.⁷ However, it is a tedious technique. Alternatively, some surgeons simply dilute 40 mg/mL TA 1:10 with balanced salt solution (BSS). However, the final product will contain 0.01% of benzyl alcohol preservative. Other surgeons prefer a sedimentation–resuspension–dilution technique. The technique involves leaving a Kenalog 40 mg/mL vial sitting undisturbed in the operating room. When TA is needed, the supernatant

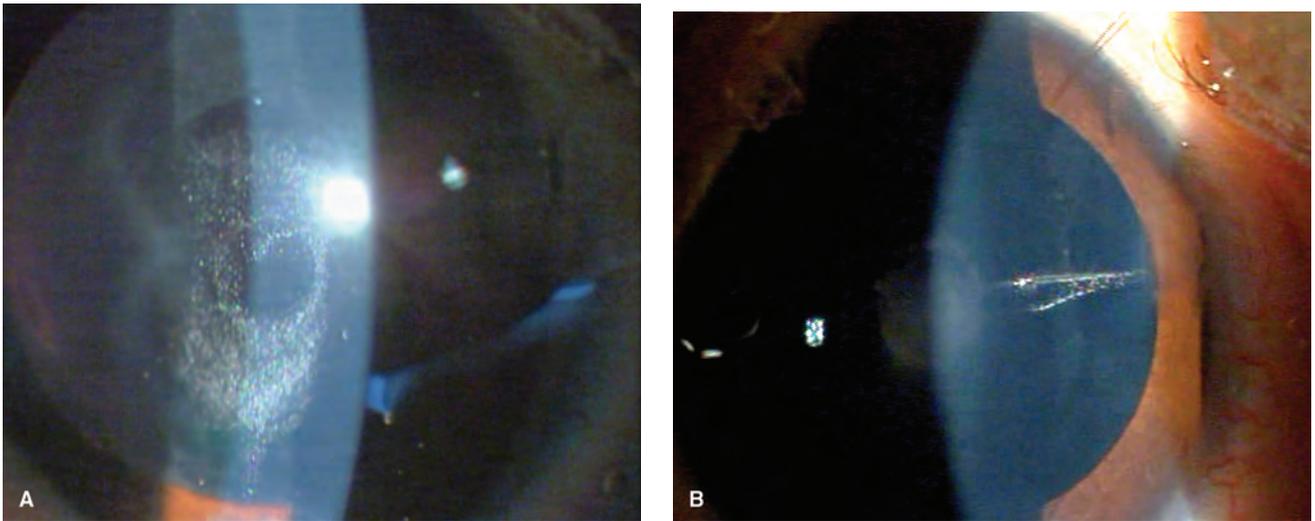


Figure 22.1. **A:** Disruption in the AVF, clearly visualized by the triamcinolone staining. **B:** Vitreous strands extending toward the incision.

is drawn off and replaced with an equal volume of BSS. The TA is typically then diluted 1:10 in BSS. Assuming supernatant removal of 90% or greater, the final product will contain 0.001% benzyl alcohol or less.⁸

In the United States, most operating rooms now prefer to stock Triesence since it is approved for intraocular use and does not contain preservative. The lead authors of this chapter (ARV, SKS, MRP) use preservative-free Aurocort (AuroLab), which is similar to Triesence and available in India.²⁻⁴ These authors describe their use of TA during pediatric surgery below.

From a well-shaken injection of preservative-free TA (40 mg/mL), 0.5 mL is withdrawn in a 5.0-mL syringe containing 4.5 mL of BSS to achieve the desired concentration of 4.0 mg/mL. Next, 1.0 mL of this preservative-free TA suspension is drawn into a tuberculin syringe, and 0.2 mL is injected twice during the surgery. Extreme care to avoid clumping is taken by shaking thoroughly to ensure a uniform suspension before the drug is drawn into the syringe. The suspension is immediately injected into the anterior chamber to avoid precipitation of the TA particles in the syringe.² During each application, 0.1 to 0.2 mL (4 mg/mL) is injected into the anterior chamber. It is important to remember that TA can sometimes stain the ophthalmic viscosurgical device (OVD) in the anterior chamber. However, there are a few differences in the staining patterns. Generally, OVD forms a diffuse, uniform pattern of staining and is aspirated very fast in a bolus, whereas with vitreous disturbance, the staining pattern is localized to the vitreous strands.

1. The first injection is given on completion of the PCCC, to visualize the AVF. In the presence of AVF disturbance, TA particles tend to become entrapped and impregnated in the vitreous gel making it clearly visible (Fig. 22.1A and B). When AVF is intact, it

appears as a convex, bulging structure (Fig. 22.2), and TA particles tend to swirl freely when injected within the rhexis margin. Subsequently, a two-port limbal anterior vitrectomy is performed if vitrectomy is the planned surgical procedure or if AVF is disturbed. The triamcinolone-stained vitreous gel is used as a guide during the procedure.

2. The second injection is given on completion of two-port anterior vitrectomy, to detect the presence of any residual vitreous strands. Additional anterior vitrectomy is performed if vitreous strands are identified in the anterior chamber.
3. The third application is done after the residual OVD is removed from the eye, to detect any residual vitreous strands. Additional anterior vitrectomy is performed if vitreous strands are identified in the anterior chamber.

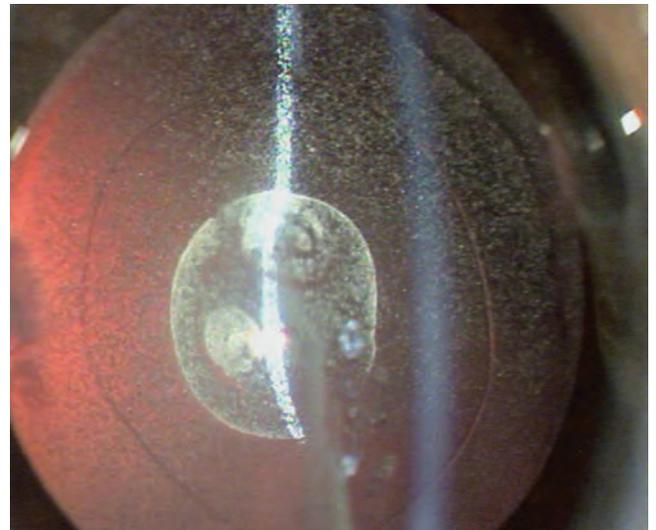


Figure 22.2. Intact AVF, seen as a convex, bulging structure highlighted by triamcinolone.

BENEFITS

Visualization of Vitreous

The intracameral use of TA has been reported to aid visualization of the presence of vitreous in the anterior chamber after posterior capsule rupture in adults.⁹ TA helps to identify vitreous in pediatric cataract surgery also.^{2-4,6} Using this procedure, surgeons can detect the presence of AVF disturbance and perform vitrectomy in such eyes. Some surgeons might argue that certain clinical signs may be sufficient in detecting residual vitreous strands. However, at times, it is difficult to detect the vitreous strands due to the transparent nature of the vitreous and retroillumination of the microscope. While dealing with young children <2 years of age, the presence of a disturbance in the AVF may not matter because vitrectomy will be performed in any case. However, for children between 2 and 6 years of age, in whom PCCC without vitrectomy may be planned, using TA to render the vitreous visible has tremendous value. Visualization of the vitreous would allow surgeons to be more precise and thorough in removing vitreous that may have remained in the anterior segment.

Anti-inflammatory Effect

Steroids have traditionally been used topically and subconjunctivally to control postoperative inflammation. While topical steroid drops are effective in controlling inflammation following cataract surgery, they have several disadvantages. One of them is the compliance issue—which is more important in children. Missed doses and noncompliance lead to subtherapeutic administration of topical anti-inflammatory medication.⁶ Possible significant advantages of intracameral TA include ease of administration at the end of surgery because the crystals persist for several days in the anterior chamber, providing a prolonged anti-inflammatory effect.⁶ In a retrospective, age-matched case-control study, we observed that in eyes with TA, it was easier to control postoperative anterior segment inflammation.⁴ The visual axis was not obscured in any eye in TA group, while 9 eyes (10.8%) in the control group developed an obscured visual axis ($P < 0.029$). Six eyes (7.2%) in the control group required secondary membranectomy with pars plana vitrectomy. There was a statistically significant difference between the two groups in posterior synechia and cell deposits. Posterior synechia were observed in 4 eyes (9.8%) in the TA group and 21 eyes (25.3%) in the control group. Cell deposits were observed in 4 eyes (9.8%) in the TA group and 21 eyes (25.3%) in the control group.

After intravitreal injection, crystals of TA have been detected as late as 40 days postsurgery.¹⁰ After single intravitreal injection of TA, the mean elimination half-life was 18.6 days in nonvitrectomized eyes, while

it was 3.2 days in vitrectomized eyes.¹¹ Such pharmacokinetic data are not available after use of intracameral TA. Even though, in our study, the majority of TA was removed, we observed less inflammation, suggesting that the steroid effect may have stabilized the blood-aqueous barrier and minimized postoperative inflammation in these complicated anterior segment surgeries. We propose that very low doses of TA injected directly into the anterior chamber or vitreous could help reduce postoperative inflammation after cataract surgery, allowing these patients to be less drop dependent. While using intracameral TA, an attempt should be made to place the solution in the bag behind the IOL to reduce the amount of medication against the corneal endothelium.⁶ Intravitreal TA has also been reported efficacious in the treatment of uveitic cystoid macular edema in children.¹² In children with uveitis cataract, the use of TA is even more beneficial.

CONCERNS

Although TA helped in detecting the vitreous strands in the anterior chamber and in reducing postoperative inflammation, there are potential concerns about its usage. The preserved form of commercial TA suspension caused damage to human and rabbit retinal pigment epithelial cells, to rabbit corneal endothelial cells, and to proliferating cells of retinal origin in vitro at doses normally used in clinical practice. Vehicle-free TA suspensions do not seem to damage human retinal pigment epithelial cells. Benzyl alcohol, a vehicle of the commercial TA, and TA in the concentration of 40 mg/mL, with or without the vehicle, were shown to be toxic to corneal endothelium. The use of a more dilute 4 mg/mL vehicle removed TA remains useful for anterior vitreous staining and may present less risk to the corneal endothelium.

In the adult literature, the major concerns with the use of TA include the occurrence of glaucoma⁹ and sterile endophthalmitis. One of six eyes had an increase in intraocular pressure (IOP).⁹ Because this patient was not a corticosteroid responder, authors believe that this was unlikely to be due to corticosteroid-induced glaucoma. Authors further noted that “it is possible that TA granules plug up the trabecular meshwork and inhibit aqueous outflow, because the IOP was normalized soon by additional washing.” Because no apparent increase of IOP was found in the other eyes, intracameral TA might not necessarily induce the increase of IOP when TA granules were sufficiently removed. The risk of steroid-induced glaucoma seems to be minimal because only a small amount of TA is used and the majority of it is removed along with the vitreous gel. In our case-control study described above,⁴ we observed no significant difference in preoperative or postoperative

IOP ($P = 0.29$ and $P = 0.50$, respectively). The mean preoperative IOP was 12.64 ± 2.15 mm Hg in the TA group and 14.29 ± 3.24 mm Hg in the control group. The mean postoperative IOPs were 13.60 ± 3.24 mm Hg and 12.40 ± 2.04 mm Hg, respectively, at 1 month and 14.02 ± 2.69 mm Hg and 14.75 ± 5.16 mm Hg, respectively, at 1 year. There was no statistically significant difference between groups in preoperative IOP or postoperative IOP ($P = 0.29$ and $P = 0.50$, respectively). No patient developed glaucoma postoperatively. Secondly, postoperative endophthalmitis is one of the serious complications that may occur after administering intravitreal TA. Endophthalmitis was not observed in our series.⁴

It is important to distinguish intravitreal from intracameral administration of TA. The worrisome incidence of sustained IOP elevation that has been reported in children when TA is placed in the vitreous of nonvitrectomized eyes has not been reported when TA is left in the anterior chamber at the time of IOL implantation. One of the authors (MEW) now has several years of experience leaving 2 mg of Triamcinolone intracamerally (without removal) during pediatric cataract and IOL surgery. It appears that this use of TA is safe and effective although further analysis is ongoing. Cleary et al.⁶ reported their experience placing TA at the conclusion of every pediatric cataract/IOL surgery. Good anti-inflammatory control was seen without significant IOP elevations.

SUMMARY

Preservative-free TA helps in visualizing the vitreous and decreasing postoperative inflammation.

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Lensectomy and Anterior Vitrectomy When an Intraocular Lens Is Not Being Implanted

M. Edward Wilson and Suresh K. Pandey

Intraocular lens (IOL) implantation at the time of cataract surgery is commonplace in older children. However, infants are often left aphakic. Optical rehabilitation is accomplished with contact lenses or aphakic glasses during the rapid years of eye growth. Later in childhood, secondary IOL implantation is usually performed. For infants, there are some significant advantages to the lensectomy and vitrectomy procedure compared to the lensectomy, vitrectomy, and IOL implantation procedure.

1. Lensectomy and vitrectomy can be performed through two small corneal stab incisions, ≤ 20 gauge. This causes less surgical trauma for the infant eye compared to when an IOL is inserted. The avoidance of a larger incision makes a patch and shield unnecessary. We usually place a contact lens on the eye at the conclusion of surgery and leave the eye unpatched. Post-operative drops can be started immediately instead of waiting until the patch and shield are removed the day after surgery. Having the baby return after surgery with no eye bandage is a great psychological boost for the parents. The baby can “start seeing” right away.
2. The risk of recurrent visual axis opacification after infant surgery is much lower when an IOL is not placed primarily. The anterior and posterior capsule remnants seal to each other more securely when an IOL is not placed between them. If a Soemmering ring forms, it is more likely to remain peripheral to the visual axis. In contrast, infants with an IOL placed primarily are at higher risk of visual axis opacification from cortex that escapes the Soemmering ring and reaches the visual axis.
3. Aphakic contact lenses can be changed whenever the eye grows, allowing precise optical rehabilitation. When an IOL is placed in infancy, glasses must be worn for the residual hyperopia. Later in childhood,

myopic glasses are often needed. Some parents find that contact lens wear (especially with the easy-to-handle extended-wear silicone contact lenses designed especially for infants) is easier to manage than glasses, at least in infancy. Later, if contact lens wear becomes more difficult, the eye has grown enough that the glasses (as an adjunct to an IOL) are less thick and less necessary for prevention or treatment of amblyopia.

The disadvantages of aphakia in infancy with optical replacement using contact lenses include worsening amblyopia whenever a contact lens is lost and the risk of corneal ulcers owing to the extended wear of the lenses during the day and night. These issues are also covered in other chapters in this book.

SURGICAL APPROACHES FOR LENSECTOMY AND ANTERIOR VITRECTOMY

Two main approaches exist for lensectomy and anterior vitrectomy in children: the pars plana/pars plicata approach and the limbal/corneal approach.¹⁻⁸

The Pars Plana Approach for Lensectomy and Anterior Vitrectomy

The pars plana/pars plicata approach is not commonly used today for primary removal of a congenital cataract unless vitreoretinal pathology is also being addressed. When retina surgeons perform a lensectomy combined with a posterior vitrectomy and retinal repair in an infant, a pars plana approach is often preferred. Pediatric anterior segment surgeons are more likely to prefer a limbal approach. Pars plana posterior capsulectomy after limbal-approach lensectomy and IOL placement is covered elsewhere in this book.

Surgical Technique

Pars plana/pars plicata lensectomy requires a guillotine-type vitrectome. Epinephrine (adrenaline), 1:500,000, is mixed in balanced salt solution (BSS; Alcon Laboratories, Fort Worth, TX) to avoid intraoperative miosis and to avoid occurrence of pediatric intraoperative floppy-iris syndrome.⁹ The conjunctiva is opened at the 10 o'clock and 2 o'clock positions to expose the sclera at the level of the pars plana. Two scleral perforations are made using a microvitoretinal (MVR) knife that matches the gauge of instruments to be used (20, 23, 25, 27 gauge) or a Stiletto knife (DORC, the Netherlands) at the pars plana/pars plicata level: one for the vitrectomy probe and the second for the infusion cannula. Stab incision placement recommendations are as follows: 2 mm posterior to the limbus in an infant, 2.5 mm in a toddler, and 3 mm in a school-aged child. A lensectomy–anterior vitrectomy is completed, sparing a peripheral rim of the capsular sac including the anterior, equatorial, and posterior capsule. These capsule remnants are used to create a shelf to support a posterior chamber IOL that may be implanted later in life. It is important to avoid vitreous incarceration in the wounds by turning off the infusion before withdrawing the vitrectome from the eye. This precaution reduces the chances of inducing retinal traction and retinal detachment later in life. The pars plana/pars plicata scleral incisions are usually closed with 8-0, 9-0, or 10-0 synthetic absorbable suture. Recently, sutureless pars plana anterior vitrectomy through self-sealing sclerotomies in children has also been reported.¹⁰

Advantages and Disadvantages of the Pars Plana Approach

This approach minimizes the possibility of surgical trauma to the iris and the corneal endothelium because fewer maneuvers occur in the anterior chamber. However, there is a greater likelihood that inadequate capsule will remain for support of a secondary sulcus-fixated IOL using this approach.

The Limbal Approach for Lensectomy and Anterior Vitrectomy

Pediatric surgeons, when operating on infants, use the limbal/corneal approach most often. Most use a bimanual technique, but some use a single incision with an infusion sleeve attached to the vitrector handpiece. We recommend separating the infusion and the vitrector rather than using an infusion sleeve.

Surgical Technique

Figure 23.1 illustrates our technique for pediatric cataract surgery with the limbal/corneal approach to lensectomy and anterior vitrectomy. Under general anesthesia,

two corneal stab incisions are recommended, using a 20-gauge MVR knife (smaller 23-, 25-, and 27-gauge instruments will have a corresponding smaller-gauge MVR knife) at the 10 o'clock and 2 o'clock positions: one for an irrigation cannula (connected to a BSS) and the other one for an aspiration/cutting handpiece. The stab incisions should be located at the terminal end of the limbal blood vessels as they reach the clear cornea. If the incisions are made too close to the conjunctiva, ballooning of the conjunctiva will occur from fluid that tracks back into the subconjunctival space during lensectomy. Care should be taken when making the incisions to assure a tight fit with the instruments to be inserted into the anterior chamber. The MVR knife is initially aimed posteriorly but is flattened out into the iris plane after the tip enters the anterior chamber. This maneuver will produce a tunneled incision about 1 mm wide and about 1 mm long. With the soft eyes (low tissue rigidity) of babies, an iris plane incision from the point of entry will result in a longer tunnel than expected. The anterior chamber will be entered closer to the visual axis than intended. Maneuvering the instruments will also be more difficult with this long tunnel due to the “oar locking” phenomenon. Also, the MVR knife needs to be backed out precisely along the path of entry. Beginning surgeons have a tendency to retract the knife along a slightly different path than the entry. This causes the soft corneal tissue of the infant to slice open wider than the blade width. Leakage around the instruments will result during surgery, leading to anterior chamber instability. Finally, the nondominant-side incision should be made first (left-hand incision for a right-handed surgeon). Usually, the incision can be made without resulting in a flat anterior chamber. In that circumstance, the second incision can be made without placing an ophthalmic viscosurgical device (OVD) into the eye. If the chamber flattens after the nondominant incision, the irrigating cannula can be placed in the eye through the initial opening, and the second incision can be made with a fluid-filled chamber. No OVD is needed when an IOL is not being implanted.

Our preferred infusion instruments are a 20-gauge blunt-tip, angled, beveled irrigating cannula (Grieshaber 170.01, Alcon or Nichamin sideport, Bausch & Lomb Storz) or a 25 gauge anterior chamber maintainer (Lewicky, Bausch & Lomb Storz). Aspiration and cutting is provided by a 20-gauge (more efficient when removing gummy cortex) or a 25 gauge vitrectomy handpiece (Accurus or Constellation, Alcon). The aspiration handpiece (Grieshaber 170.02, Alcon) that matches the bimanual irrigation cannula is also placed on the surgical table for use during cortex aspiration if needed. However, using the vitrector handpiece for both the cutting steps and the aspiration steps reduces the need to enter and exit the eye multiple times.

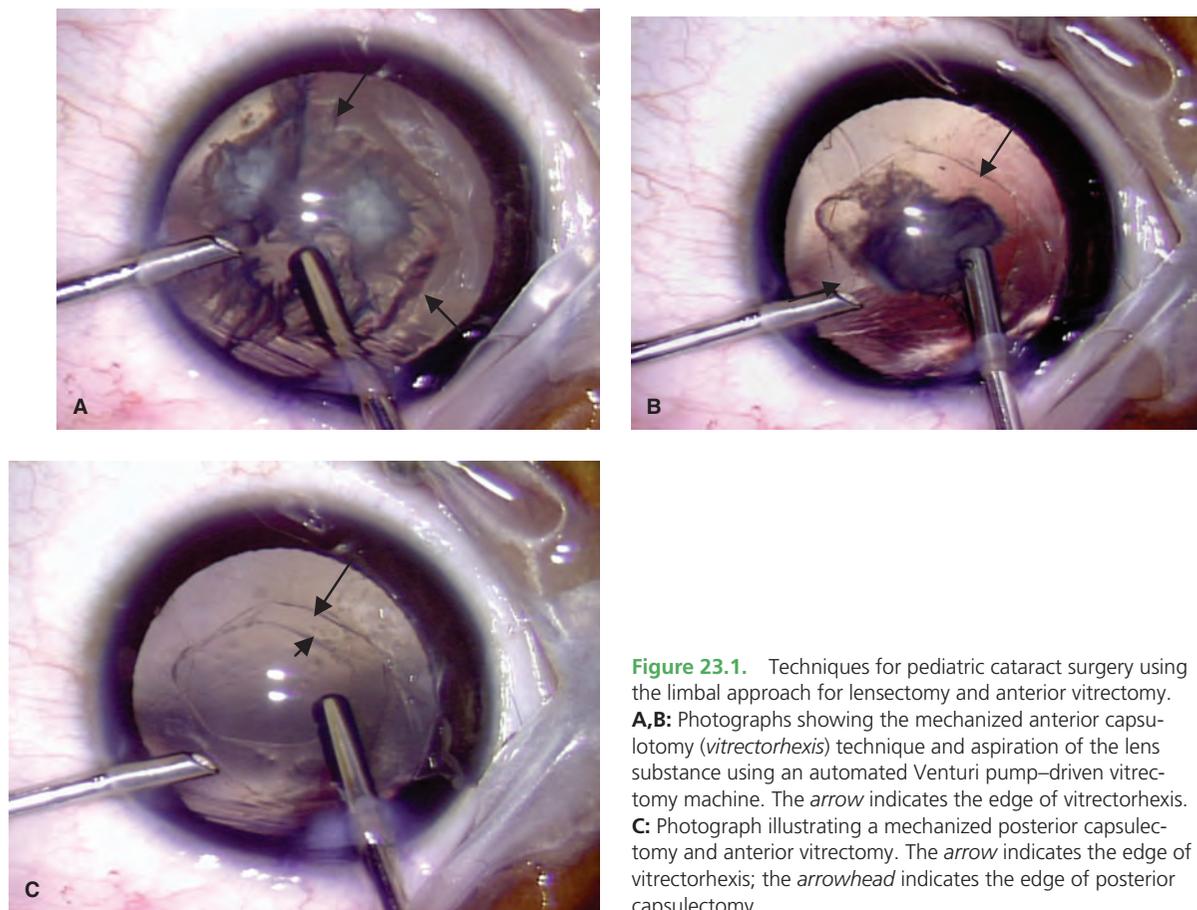


Figure 23.1. Techniques for pediatric cataract surgery using the limbal approach for lensectomy and anterior vitrectomy. **A,B:** Photographs showing the mechanized anterior capsulotomy (*vitrectorhexis*) technique and aspiration of the lens substance using an automated Venturi pump-driven vitrectomy machine. The *arrow* indicates the edge of vitrectorhexis. **C:** Photograph illustrating a mechanized posterior capsulectomy and anterior vitrectomy. The *arrow* indicates the edge of vitrectorhexis; the *arrowhead* indicates the edge of posterior capsulectomy.

It is important that the instruments be equal in size. Subincisional cortex removal may require that the right- and left-hand instruments be switched. If the incisions and the instruments are of identical size, the tight fit can be maintained, which will keep the anterior chamber depth stable. Epinephrine (1:500,000) is usually added in the BSS to maintain the pupillary mydriasis. Although Ringer lactate solution works well for irrigation associated with adult extracapsular cataract surgery, temporary corneal clouding can occur when it is used in the high-flow pediatric cataract surgery described here.

The anterior chamber is entered first (with the surgeon's nondominant hand using the nondominant-side incision) with the irrigating cannula. Care is taken to place the beveled tip in along the plane of the tunneled incision. A slight swirling motion often facilitates entry. The vitrectomy handpiece is more difficult to place because it is not beveled. Twirling movements aid entry. Take care not to aim too superficially within the incision. This will result in the instrument's missing the internal lip of the wound and tracking into the soft infantile corneal stroma. If this happens, back the instrument out and reenter with more posterior pressure on the tip of the instrument as it enters the wound. Occasionally, it will be

necessary to have an assistant lift up on the incision's outer lip to facilitate entry. Resist the temptation to enlarge the wound. Operating on very young infants requires a stable anterior chamber and a leaking wound will not facilitate chamber stability.

With both instruments in the anterior chamber, the vitrectorhexis can be performed. Our preferred settings on the Accurus machine are a fluid pressure of 50, aspiration of 200, and a cutting rate of 150. The vitrector is placed just anterior to the center of the anterior capsule, with the cutting port facing directly posterior. The aspiration foot pedal is depressed until the anterior capsule enters the cutting port and is opened. The surgeon should stay slightly anterior to the plane of the anterior capsule and let the capsule come to the cutter rather than chasing the capsule with the cutter and inadvertently reaching the posterior capsule prematurely. The anterior capsulotomy should be enlarged to the desired size using a gentle spiraling movement from the center outward. If the instrument slips posterior to the capsule edge, it should be pulled back anterior to the capsule where the cutter can be redirected. Care should be taken to avoid creating right-angled edges in the capsulotomy. The ideal size is 4.5 to 5 mm.

Without removing the instruments from the eye, the cutter is then turned off, leaving the instrument in the aspiration-only mode. The vitrector tip (now functioning as an aspiration handpiece) is directed under the anterior capsulotomy into the equator of the lens, with the port facing the equator of the lens and turned toward the surgeon. Peripheral cortex is aspirated first. Rather than pulling the instrument toward the center of the capsule, the aspiration foot pedal should be depressed (increasing the aspiration toward the maximum setting of 200) while the instrument is still near the lens equator. As the lens cortex begins to disappear into the aspiration port, some stripping toward the center is appropriate, but not to the degree usually used in adult surgery. The center of the lens is often aspirated last. This keeps the posterior capsule back until the equatorial cortex has been removed. (Note: If needed, the vitrector handpiece can be removed and replaced by the bimanual aspiration handpiece. The advantage is that this handpiece is angled and tapered. It reaches equatorial cortex easier, and it is a more efficient aspiration device compared to the vitrector. However, the disadvantage is that it must be removed after lens aspiration and again replaced by the vitrector for capsulectomy/vitrectomy. The surgeon can weigh these advantages and disadvantages. We change to the aspiration bimanual cannula in babies only when the vitrector is not able to remove all lens cortex safely and efficiently.)

When the capsular bag has been cleaned of all lens material, the cutter is again turned on, and the cutting port is positioned centrally, just anterior to the posterior capsule. The surgeon then aspirates the capsule up into the cutting port. Often, the posterior capsule is engaged with this technique without simultaneously engaging the vitreous face. Nonetheless, the cutter speed is increased to 800+ cuts/min, and the irrigation is decreased to 30, in case vitreous is encountered. As with the anterior capsule, enlargement of the posterior capsulotomy is done in a slow, spiraling manner. A central vitrectomy is performed (aiming to remove approximately one-third of the vitreous volume) after completion of the posterior vitrectorhexis. The irrigation cannula should be placed so as to direct fluid away from the vitrector tip.

At the end of the vitrectomy, the instruments are removed from the eye at nearly the same time. The irrigation cannula should exit just before (and not after) the removal of the vitrector tip. If a vitreous strand to either of the exit wounds is suspected, a miotic can be irrigated into the anterior chamber to constrict the pupil. If peaking of the pupil is seen, the vitrector is placed back into the anterior chamber until removal is complete. Rarely, an OVD will be needed along with a sweep of a vitreous strand from the wound using an iris spatula. Amazingly, vitreous rarely comes to the wound using this technique with infants. The formed vitreous of the baby combined with a generous central anterior vitrectomy results in an

anterior segment devoid of vitreous nearly every time. A recent study confirmed benefits of preservative-free triamcinolone acetonide to improve visualization of the vitreous during pediatric cataract surgery, thereby ensuring thorough and complete anterior vitrectomy.¹¹ The authors of this study revealed that IOP was not affected, and no adverse postoperative results were observed.¹¹

The stab incisions are closed with one interrupted 10-0 Vicryl suture each. A drop of 5% povidone-iodine along with atropine and steroid/antibiotic drops is placed on the eye at the end of the surgery. An appropriate Silsoft aphakic contact lens (usually a 7.5-base curve, 29- or 32-diopter power for neonates) is placed on the eye as well (see Chapter 45). The child is then awakened and recovered.

Advantages and Disadvantages of the Limbal/Corneal Approach

The limbal/corneal approach is more familiar to the anterior segment surgeon than the pars plana/pars plicata approach. The conjunctiva need not be disturbed, and an adequate (for secondary IOL implantation later) capsular rim is more likely to remain when this approach is used. However, there is a greater risk of iris manipulation or iris damage by the cutting port when this approach is used. To avoid inadvertent iris cutting, the surgeon should take care to keep the vitrector cutting port facing away from the iris. If not, a sudden shallowing of the anterior chamber from fluid leaking around the instruments can result in the iris entering the cutting port.

SUMMARY

Lensectomy and anterior vitrectomy without an IOL constitute a widely used surgical technique for the removal of visually significant congenital cataracts in the 1st year of life. The technique, as described here, allows the anterior capsulotomy, lens aspiration, posterior capsulectomy, and anterior vitrectomy all to be done without ever having to take the instruments out of the eye. The procedure is fast and efficient, and it induces a minimal amount of surgical trauma. No conjunctival incision is needed. Closure is with two interrupted 10-0 synthetic absorbable sutures. The aphakic contact lens can be placed on the eye at the conclusion of surgery. No bandage or shield is necessary.

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Intraocular Lens: Experimental Studies and Clinical Applications

Suresh K. Pandey and M. Edward Wilson

Intraocular lens (IOL) implantation at the time of cataract surgery has become the most common means of optical correction for children beyond infancy. A growing number of case series and a survey of practice styles and preferences of American Society of Cataract and Refractive Surgery (ASCRS) and American Association for Pediatric Ophthalmology and Strabismus (AAPOS) members support the safety and effectiveness of IOLs for children.¹⁻¹¹ An IOL can provide a full-time correction with optics that closely simulate those of the crystalline lens, with the exception of accommodation. However, some concern still remains about the unknown long-term risks of an IOL over the lifespan of a child. In addition, predicting the refractive change of even older children is difficult since individual variation in eye growth is common.

An IOL provides permanent continuous correction as opposed to aphakic glasses or aphakic contact lenses that can easily be removed by the child. This may be important in preventing amblyopia and encouraging normal visual development. Although glasses are usually necessary to obtain the best vision, uncorrected pseudophakic vision is probably less amblyogenic than is uncorrected aphakic vision.

IOL IMPLANTATION IN PEDIATRIC CATARACT SURGERY

Intraocular lenses manufactured from various rigid and foldable biomaterials have been used for pediatric cataract surgery for decades. Rigid implants manufactured from polymethyl methacrylate (PMMA) have been implanted in humans (adults and children) beginning more than 50 years ago. An IOL implanted in a child's eye must stay there for several decades without biodegrading. PMMA has the longest safety record in both adults and children.

From the 1990s on, IOL implantation has been the standard method for correcting aphakia beyond infancy

and when there is sufficient capsular support. The three most important reasons for increased use of IOLs in children are as follows.

1. Appropriately sized and more flexible implants can be inserted much more easily into the capsular bag of the child. Despite their increased flexibility, newer lens designs retain enough "memory" to resist the intense equatorial capsular fibrosis seen in children after implantation. PMMA as an implant material proved to be very biocompatible. In addition, copolymerization of different acrylate and methacrylate acids resulted in foldable lenses with many of the same biocompatibility features as PMMA. As an example, foldable hydrophobic acrylic lenses, such as the AcrySof® (Alcon, Inc., Fort Worth, TX), are being implanted in the eyes of children of all ages. The biocompatibility of the hydrophobic acrylic lenses may equal or exceed that of the tried and true PMMA lenses. The foldable acrylic lenses are easier to insert in a small eye, and the squared edge of many of the optic designs now on the market may result in delayed posterior capsule opacification in young eyes. Pavlovic et al.⁵ reported that foldable silicone IOL implantation in children is also a safe procedure with stable short-term anatomic results.
2. Refined surgical techniques can more predictably ensure capsular fixation of the IOL. Surgeons have gained more confidence in the safety of "in-the-bag" implantation even over the extended lifespan of a child. Capsular fixation provides sequestration of the implant away from vascularized tissues. Although ciliary sulcus fixation of the IOL may also be safe, uveal contact for a lifetime is not as desirable. In addition, complications such as pupillary capture and IOL decentration are more common with ciliary sulcus fixation. The confidence in the safety of capsular fixation has resulted in more IOLs being implanted in children at the time of cataract extraction.

- Finally, customized management of the anterior and posterior capsules for pediatric eyes at the time of implantation has improved outcomes and decreased complications. These improvements have provided for long-term centration of the IOL and a reduction in opacification of the visual axis after implantation.

IOL SIZING FOR PEDIATRIC CATARACT SURGERY

The mean axial length of a newborn's eye is 17.0 mm compared to 23 to 24 mm in an adult. The pediatric eye, especially in the first 1 to 3 years of life, is significantly smaller than the adult eye.¹² This has led to concerns about implantation of adult-sized IOLs in these eyes. In an effort to determine the size of the pediatric lens, Bluestein et al.¹² examined 50 fresh, nonpreserved autopsy eyes from patients ranging in age from 1 day to 16 years. A variety of measurements were made, including the anterior–posterior, vertical, and horizontal lengths of the globe, corneal diameter, lens diameter, and diameter of the zonular free zone. The most rapid growth of the globe, the lens (Fig. 24.1), and the capsular bag occurred from birth to 2 years of age.

Currently available adult-sized IOLs are slightly oversized in relation to capsular bag measurements but may actually fit into eyes in the first 2 years of life, although possibly not into very small infantile eyes. The small capsular bag may become misshaped into an oval with these adult-sized IOLs. The ovality may vary from minimal to severe, depending on the design, type, and overall diameter of the implanted IOL (Fig. 24.2). There are several possible consequences of implantation of adult-sized IOLs into the relatively small capsular bag of infants and young children. First, in contrast to adult cataract surgery, dialing of the IOL haptics into the capsular bag can be difficult in infants and children. The combination of a small soft eye with vitreous upthrust and an oversized IOL will increase the risk of asymmetric (bag–sulcus) IOL fixation, which can lead to decentration of the IOL. Second, implantation of an oversized IOL in the

capsular bag of a child may cause marked capsular bag stretching, resulting in posterior capsular folds and striae. The lens epithelial cells may migrate toward the visual axis, through the capsular folds, leading to opacification of the posterior capsule. Third, implantation of an oversized IOL in the capsular bag of a small child may cause zonular stress in the direction parallel to the IOL haptics. The long-term sequelae of the capsular bag stretching (and also the zonular stress) on the axial growth of the globe remains to be further investigated.

Pandey et al.¹³ compared the amount of capsulorhexis ovaling and capsular bag stretch produced by various rigid and foldable IOLs when implanted into pediatric human eyes obtained postmortem. In this study, 16 pediatric human eyes obtained postmortem were divided into two groups: Eight eyes were obtained from children <2 years of age (group A) and eight eyes were obtained from children >2 years of age (group B). All eyes were prepared according to the Miyake–Apple posterior video technique.^{14,15}

Six IOL types (listed below), manufactured from rigid and foldable biomaterials, were implanted to determine the IOL design best suited for implantation in pediatric eyes.

1. Single-piece hydrophobic acrylic IOL (AcrySof®, SA30AL; 5.5-mm optic, 12.5-mm overall diameter; Alcon Inc., Fort Worth, TX).
2. Three-piece hydrophobic acrylic optic–PMMA haptic (AcrySof®, MA60BM; 6-mm optic, 13-mm overall diameter; Alcon Inc.)
3. Three-piece silicone optic–PMMA haptic (SI40NB; 6-mm optic, 13-mm overall diameter; AMO Inc., Santa Ana, CA).
4. Three-piece silicone optic–polyimide haptic (elastimide; 6-mm optic, 12.5-mm overall diameter; Staar Surgical Co., Monrovia, CA).
5. One-piece silicone plate IOL (AA-4203VF; 6-mm optic, 10.5-mm overall diameter; Staar Surgical Co.).
6. One-piece PMMA optic–PMMA haptic (809 P; 5-mm optic, 12-mm overall diameter; Pfizer Ophthalmics [formerly Pharmacia Inc.], New York).

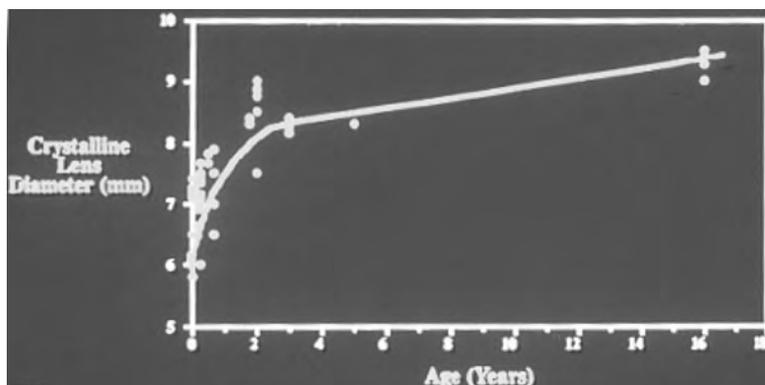


Figure 24.1. Graph showing the growth of the crystalline lens. The most rapid growth occurs from birth to 2 years of age, and the growth of the crystalline lens is almost completed by the age of 2 years. (Reprinted from Bluestein EC, Wilson ME, Wang XH, et al. Dimensions of the pediatric crystalline lens: implications for intraocular lenses in children. *J Pediatr Ophthalmol Strabismus* 1996;33:18–20, with permission.)

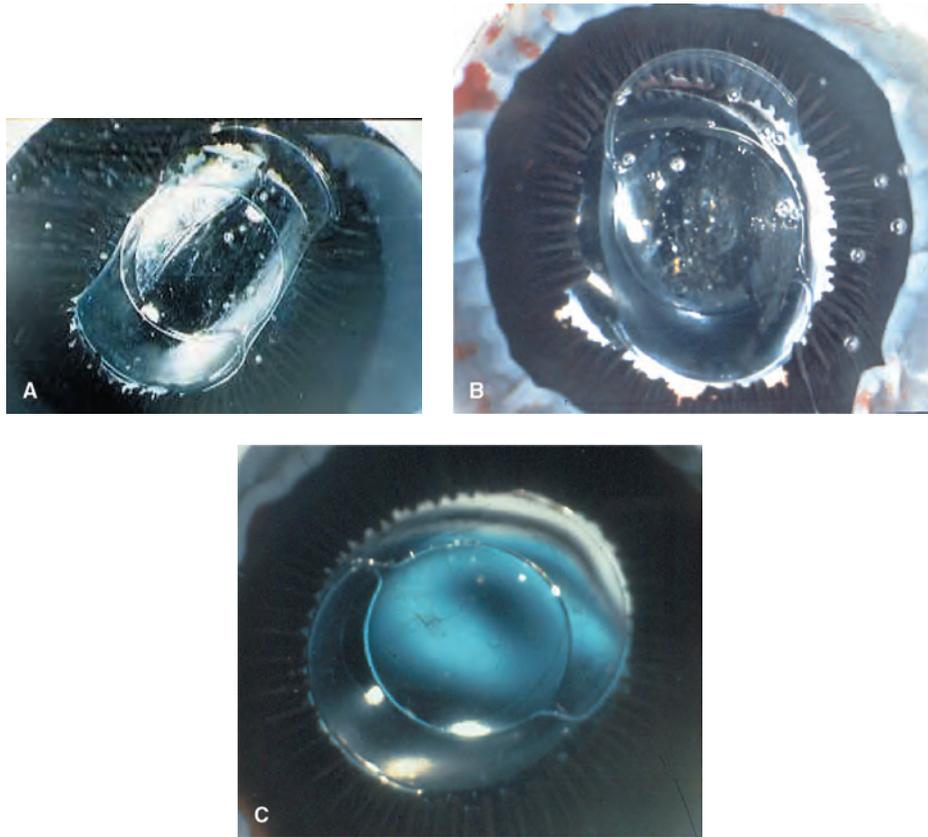


Figure 24.2. The small capsular bag may ovalize with these adult-sized IOLs; the ovality may vary from minimal to severe, depending on the design, type, and overall diameter of the implanted IOL. **A:** Miyake-Apple view after in-the-bag implantation of a one-piece, flexible open-loop, all-PMMA IOL, 13.75 mm in overall length, into an eye obtained postmortem from a 2-year-old child. Note the severe capsular bag stretch and ovaling between 1 o'clock and 7 o'clock. These indicate marked oversizing of the IOL in relation to the size of the capsular bag. **B:** Miyake-Apple view after in-the-bag implantation of a one-piece PMMA, flexible open-loop, modified C-loop capsular IOL, 12.5 mm in diameter, into an eye obtained postmortem from a 5-year-old child. Note the marked ovaling and elongation of the bag between 12 o'clock and 6 o'clock, indicating the slightly oversized IOL. **C:** Miyake-Apple posterior photographic view showing a one-piece, all-PMMA, 12.0-mm-diameter, modified C-loop IOL experimentally placed into the capsular bag in a cadaver eye of a 2-year-old child. Note the very optimum "fit" of the implanted IOL. (Reprinted from Wilson ME, Bluestein EC, Wang XH. Current trends in the use of intraocular lenses in children. *J Cataract Refract Surg* 1994;20:579–583, with permission.)

The capsulorhexis opening and the capsular bag diameters were measured before IOL implantation and subsequently after in-the-bag IOL fixation with the haptics (or the main axis) of the lens at the 3 o'clock to 9 o'clock meridian. The percentage ovaling of the capsulorhexis opening was calculated by noting the difference in its horizontal diameter before and after IOL implantation. The percentage of capsular bag stretch was also calculated by noting the difference in the horizontal capsular bag diameter before and after IOL implantation.

All of the IOLs produced ovaling of the capsulorhexis opening, and stretching of the capsular bag, parallel to the IOL haptics. Figures 24.3 and 24.4 illustrate ovaling of the capsulorhexis opening and capsular bag stretch after implantation of rigid and foldable lenses in postmortem human eyes obtained from both group A and group B. Comparison of all six lens types within each group of eyes

revealed significant differences in capsulorhexis ovaling and capsular bag stretch ($P < 0.001$, analysis of variance). However, the postcomparison difference was found to be significant only between the single-piece hydrophobic acrylic (AcrySof®) lens and the other lenses. The single-piece hydrophobic acrylic lens was associated with significantly less capsulorhexis ovaling and capsular bag stretch in both group A and group B ($12.06\% \pm 0.59\%$ and $7.6\% \pm 1.47\%$, respectively).

Our study using postmortem human eyes suggests that IOLs, designed for the adult population, can be implanted in the capsular bag of infants and children. However, a variable degree of ovaling of the capsulorhexis opening and capsular bag stretch was seen after implantation of the adult-sized IOLs. Miyake-Apple posterior video technique confirmed the well-maintained configuration of the capsular bag (with minimal ovaling) after implantation of

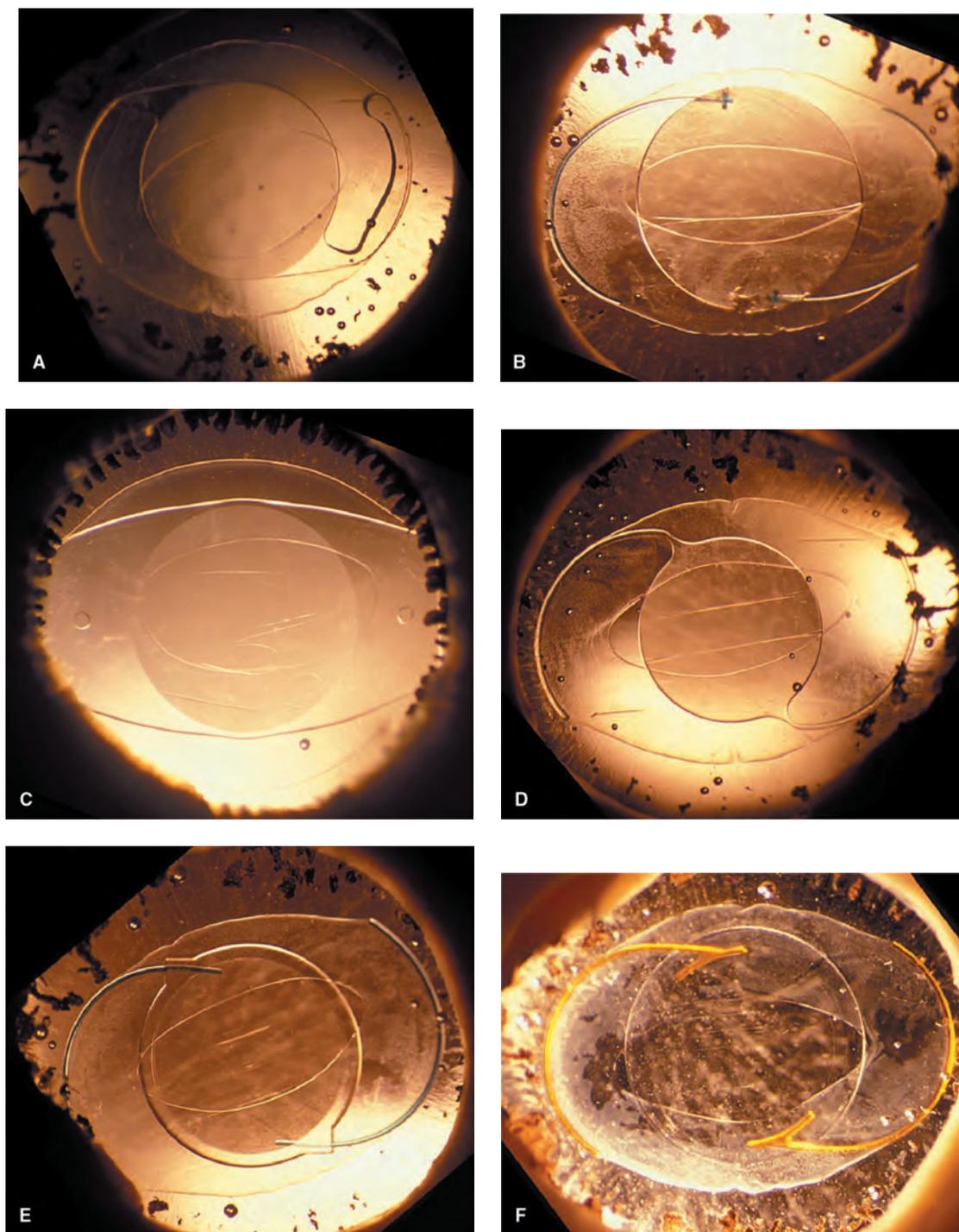


Figure 24.3. Gross photographs from the left eye of a 5-month-old child (anterior view with retroillumination). Note the variable degree of capsulorhexis ovaling and capsular bag stretch after implantation of the following rigid and foldable lens designs. Minimal capsulorhexis ovaling and capsular bag stretch were documented after implantation of a single-piece hydrophobic acrylic lens design. **A:** Single-piece hydrophobic acrylic (AcrySof®) IOL. **B:** Three-piece hydrophobic acrylic optic-PMMA haptic IOL. **C:** One-piece silicone plate IOL. **D:** One-piece PMMA optic-PMMA haptic IOL. **E:** Three-piece silicone optic-PMMA haptic IOL. **F:** Three-piece silicone optic-polyimide haptic IOL.

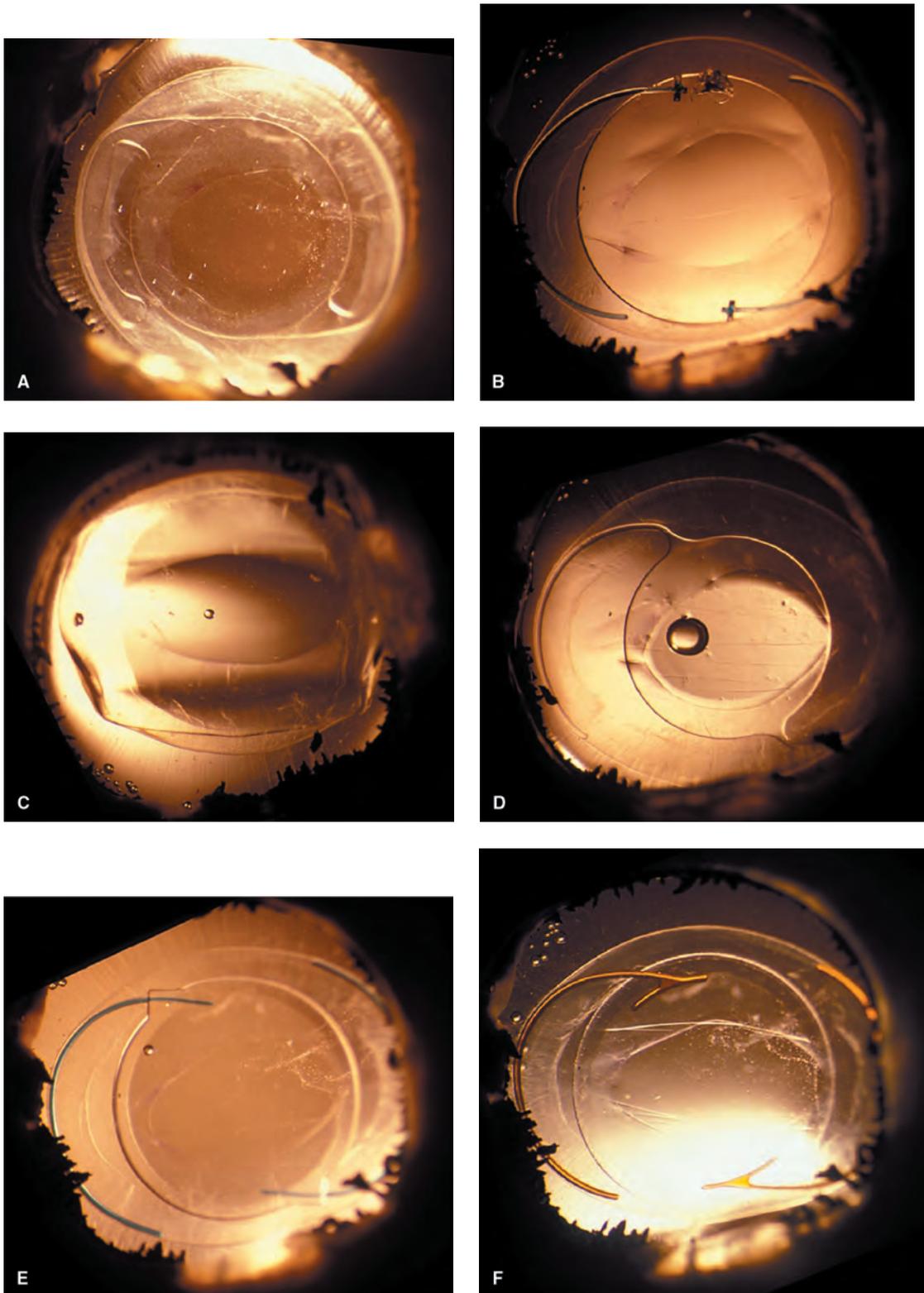


Figure 24.4. Gross photographs from the left eye of a 4-year-old child (anterior view with retroillumination). Note the variable degree of capsulorhexis ovaling and capsular bag stretch after implantation of the following rigid and foldable lens designs. Minimal capsulorhexis ovaling and capsular bag stretch were documented after implantation of a single-piece hydrophobic acrylic lens design. **A:** Single-piece hydrophobic acrylic (AcrySof®) IOL. **B:** Three-piece hydrophobic acrylic optic-PMMA haptic IOL. **C:** One-piece silicone plate IOL. **D:** One-piece PMMA optic-PMMA haptic IOL. **E:** Three-piece silicone optic-PMMA haptic IOL. **F:** Three-piece silicone optic-polyimide haptic IOL.

the single-piece hydrophobic acrylic lens in both groups due to its flexible haptic design.

The reason for this minimal ovaling of the capsulorhexis opening and capsular bag stretch is probably the result of the unique optic/haptic design. The memory and flexibility of the hydrophobic acrylic biomaterial allow the haptics of the single-piece design to bend and twist during entry and then conform to any size of capsular bag to a much greater degree than the other lenses tested. The modified “L”-loop haptics are described as Stable-Force haptics and are made of the same hydrophobic acrylic biomaterial used for the optic of the lens.¹⁶ The loop memory of the Stable-Force haptics has not been addressed in the previous laboratory studies.^{17,18} However, these haptics appear to have very good memory (reexpansion) characteristics despite being more flexible than PMMA or prolene haptics. The single-piece AcrySof[®] seems to have an ideal combination of softness and flexibility during implantation, causing minimal ovaling of the capsule, and enough memory to resist the equatorial fibrosis of the capsular bag known to occur in children. Our clinical follow-up has verified this fact. We have not seen any examples of the soft single-piece AcrySof[®] haptics being compressed over the optic by the equatorial capsular fibrosis. Besides the IOL haptic design and memory, the stickiness of the hydrophobic acrylic biomaterial may also play some role in reducing the extent of capsulorhexis and capsular bag distortion in young eyes.

Wilson et al.¹⁹ conducted an experimental IOL implantation study in 50 pediatric eyes obtained postmortem to determine the biomaterials, designs, and sizes that may be appropriate for pediatric implantation. Based on this study, using the Miyake-Apple posterior video technique, these researchers recommended downsizing PMMA IOLs to approximately 10.0-mm diameter if they were to be used for children <2 years of age. Capsular IOLs were defined as flexible open-loop, one-piece, all-PMMA, modified C-loop designs made especially for in-the-bag placement. With the advent of single-piece acrylic foldable IOLs, the recommended downsizing did not need to occur. The newer lenses adapted much better to the smaller capsular bag of young children, allowing the pediatric surgeon to use the adult-sized IOLs more safely.^{3,10,20}

AcrySof[®] IOL remains most commonly implanted IOL in children, and it is now possible to implant AcrySof[®] IOL using a 2.2-mm incision. Introduction of newer IOL designs, for example, Tecnis one-piece IOL (Abbott Medical Optics, Santa Ana, CA) and Microincision IOL (MIL) (Bausch & Lomb, Rochester, NY), can also be successfully implanted in smaller capsular bag of children and infants, using 2.2- and 1.8-mm incisions, respectively. AcrySof[®] Toric IOL and AcrySof[®] Multifocal Toric IOLs (Alcon Lab., Fort Worth, TX) are now available to correct preexisting corneal astigmatism of 1.0 diopter (D) or more. Clinical studies have now been

published to evaluate efficacy, safety, and feasibility of implantation of a bag-in-the-lens (BIL) in children and babies.²¹ First reported by Marie-Jose Tassignon, the BIL technique may be helpful to maintain a clear visual axis in infantile cataract surgery.

CURRENTLY AVAILABLE IOLS AND THEIR CLINICAL APPLICATION

Since experience with newly available implants in children is very limited, a significant amount of the information provided in this chapter on the various designs/biomaterials is derived from cataract-IOL surgery in adults.

Aspherical Implants

Three aspheric and/or aberration-free IOLs are currently FDA approved: the AcrySof[®] (aspheric optic SN60WF, Alcon), the Tecnis (AMO), and the SofPort AO (Bausch & Lomb).

AcrySof[®] IOL Model SN60WF

AcrySof[®] IOL Model SN60WF has a similar design and biomaterial characteristics as the Single-Piece AcrySof[®] Natural except it has a posterior aspheric surface. The AcrySof[®] IOL Model SN60WF compensates for spherical aberration (SA) by addressing the effects of overrefraction at the periphery. This is accomplished without increasing edge thickness. The IOL asphericity is designed to counteract the “normal” human corneal asphericity. The one-piece AcrySof[®] IOL remains one of the most preferred IOLs for implantation during pediatric cataract surgery due to its satisfactory visual axis clarity, acceptable inflammatory response, and centration in pediatric eyes. However, a recent study²² reported more early transient inflammation with implantation of AcrySof[®] Natural (yellow) when compared to nontinted (clear) IOLs in pediatric cataract surgery, but long-term inflammatory sequelae were found to be equal, as was the rate of posterior capsule opacification.

The Tecnis Z9000 Monofocal IOL

The Tecnis Z9000 (AMO Inc., Santa Ana, CA) IOL is designed to reduce SA. Investigators have demonstrated improvement on the optical bench in contrast sensitivity under mesopic and photopic conditions with this technology. Recent advances in wave front measurement of total ocular aberrations and corneal aberrations have demonstrated that the asphericity of the cornea remains constant throughout life, while the refractive gradient of the lens changes and produces increasing SA.

The Tecnis IOL is designed to reproduce the compensatory negative SA of the youthful crystalline lens and increases pseudophakic contrast sensitivity. Clinical trials in adults have shown that this modified prolate IOL

improves functional vision under most scenarios; however, decentration >0.5 mm will decrease the functional vision when compared to spherical IOLs.²²⁻²⁴

Aberration-Free Aspheric IOLs

The most recently introduced aspheric optic designs are the aberration-free optics introduced on the Akreos® Advanced Optic IOL and Sofport® Advanced Optic IOL, both from Bausch & Lomb. These lenses have aspheric posterior and anterior surfaces designed to have uniform centre-to-edge power and no SA. Asphericity here relates to the IOL itself, not to the IOL being part of the human eye lens system.

The aberration-free aspheric IOLs are expected to offer several benefits:

1. Being aberration free, they reduce the overall SA compared to a conventional spherical IOL, thereby improving quality of vision and contrast sensitivity.
2. By leaving the pseudophakic eye with its natural degree of corneal positive SA, the aberration-free lens allows for greater depth of field compared to IOLs with negative SA.
3. They should not add to or have any deleterious effect on the aberration profile of the eye, making them suitable for all patients, regardless of corneal shape.
4. The uniform centre-to-edge power of the optics makes them less susceptible to the effects of decentration or misalignment with the visual axis.

To fully appreciate this last point, it is important to consider the imperfect alignment of the natural phakic eye. The visual axis is oblique and not aligned to the optical axis; the pupil is not centered on the optical or visual axis, and even a lens that is centered in the capsular bag is likely to be misaligned with the visual axis. The specific benefits and disadvantages of the three principle optic types (those with positive SA, negative SA, and zero SA) need to be considered in terms of their effect on contrast sensitivity, effect on total higher-order aberration (HOA) profile, depth of field, and the effects of misalignment or decentration.

Most standard IOLs have biconvex spherical surfaces that create positive SA. These lenses have increasing centre-to-edge power, so peripheral rays come to a shorter focus than do paraxial or central rays. These standard IOLs provide good quality of vision when the pupil diameter is small, and the optic is perfectly aligned with the visual axis. However, the image blurring caused by SA will reduce the image quality in larger or dilated pupils. Furthermore, lens decentration or misalignment with the visual axis will induce asymmetrical HOAs such as coma. Studies have shown that pseudophakic patients implanted with standard IOLs have increased SA and loss of contrast sensitivity compared to phakic patients of the same age.²⁵

The aberration-free aspheric IOLs are suitable for all patients regardless of corneal aberration profile, pupil size, or angle kappa. By leaving the pseudophakic eye with its natural degree of corneal positive SA, they allow for depth of field; however, having zero SA, they reduce the total amount of SA thereby enhancing quality of vision and contrast sensitivity compared to a conventional spherical IOL. The aberration neutral design and uniform power, center-to-edge, means that they will not add HOAs to the pseudophakic eye, and unlike positive and negative SA IOLs, they minimize the optical effects of misalignment with the visual axis.

Toric IOLs

One of the great challenges in cataract surgery is the management of preexisting astigmatism. Use of the toric IOL (Staar Surgical, Monrovia, CA) has been aided by the acceptance of small-incision cataract surgery, as well as the availability of new foldable toric IOLs (e.g., AcrySof® Single-Piece Toric lens, Alcon Inc., Fort Worth, TX) that can be inserted through small clear corneal incisions using an injector. Different models (T3–T9) of AcrySof® Toric IOL can correct significant amounts of astigmatism. This lens also is available in aspheric versions for crisper vision. The AcrySof® Toric IOL is based on the AcrySof® Natural single-piece platform (Alcon Laboratories, Inc.) and is a foldable lens with a fully functional, 6-mm toric optic and Stableforce haptics (Alcon Laboratories, Inc.). The lens' acrylic material is highly biocompatible and has adhesive properties that, along with the haptic design, help to prevent rotation of the IOL after its implantation in the capsular bag. The posterior surface of the lens has added cylindrical power and axis markings to help the surgeon align the IOL after implanting it in the capsular bag. Recently, the *AcrySof® IQ ReSTOR Toric* IOL has also become available. The AcrySof® IQ ReSTOR® Multifocal Toric IOL combines the technologies of AcrySof® IQ ReSTOR® +3 add multifocal IOL and the AcrySof® Toric IOL. The AcrySof® IQ ReSTOR® Multifocal Toric IOL has the ability to deliver an excellent visual outcome with independence from glasses in many adult cases, but experience in children is lacking at the time of this writing. The toric IOL is particularly interesting to the cataract surgeon, as it does not require the alteration of current surgical technique or the acquisition of new instrumentation or skills for successful outcomes.

Multifocal Implants

The IOLs commonly in use are monofocal. It is not possible to see near and distant objects clearly with the same lens without correction, and thus, patients are at least potentially dependent on spectacles. Over the past decade, a variety of multifocal intraocular lenses (MIOLs) have been introduced and have enjoyed widespread clinical use. Both refractive and diffractive models have been shown to

be effective in allowing each eye to achieve quality, uncorrected distance, and near visual acuity after cataract surgery. The major concerns with the use of these lenses are the loss of contrast sensitivity and the inducement of glare and halos from light sources during night vision, which more commonly occurs with refractive designs. All multifocal lenses require careful attention to IOL power calculations and minimal refractive error after cataract surgery. Jacobi et al.²⁶ evaluated implantation of a zonal-progressive MIOL in children in a prospective, noncomparative, interventional case series. Thirty-five eyes of 26 pediatric patients aged 2 to 14 years with MIOL implantation at one institution with more than 1 year of follow-up were included in the study. Standard surgical procedure comprised an anterior capsulorhexis, lens material aspiration via two side-port incisions, temporal tunnel incision, and MIOL (SA40-N; Allergan, Irvine, CA) implantation in all eyes. At last follow-up, best-corrected distance visual acuity improved significantly in 71% of eyes with a visual acuity of 20/40 or better and 31% of eyes with a visual acuity of 20/25 or better. According to the authors, MIOL implantation is a viable alternative to monofocal pseudophakia in this age group.

AcrySof® ReSTOR® Apodized Diffractive Optic IOL

The AcrySof® ReSTOR® Apodized Diffractive Optic IOL design (MA60D3 or SA60D3) was found (during FDA trials) to provide patients with excellent near visual acuity without compromising distance vision. Good functional intermediate vision has also been demonstrated during the early clinical studies. No significant reduction in contrast sensitivity was found, and the incidence of severe visual disturbances was not increased compared with monofocal controls. This IOL has an anterior conventional refractive surface on the peripheral optic that provides the distant viewing power as well as an anterior concentric diffractive plate. This apodization feature in the diffractive zone combines with the refractive periphery to provide a full range of functional vision under all lighting conditions, while reducing the potential for halos and glare. The AcrySof® ReSTOR® Apodized Diffractive lens was approved by the U.S. FDA in early 2005.

The Tecnis Multifocal Lens

The Tecnis Multifocal lens (Abbot Medical Optics, Inc., Santa Ana, CA) has enhanced the ophthalmologists' IOL options. Not only does this lens perform as a foldable, diffractive, MIOL with an improved near addition, its optic also incorporates “intelligent asphericity” to counteract most corneal SAs.

The Tecnis Multifocal IOL is based on the aspheric optic design of the Tecnis IOL (Abbott Medical Optics, Inc.), which was engineered to reduce the SA of an average cornea. Based on the principle of diffraction—as is the AcrySof® Restor IOL (Alcon Laboratories, Inc., Fort

Worth, TX)—the Tecnis Multifocal IOL features diffractive rings on its posterior surface. The lens provides patients with a near and a distant focus, each of which is distinct. The rings start very close to the optic's center and then continue out toward the periphery, usually with an increasing distance between the rings. As a result, the multifocal effect has less dependence on the pupil size. Even in the presence of relatively small pupils, this IOL provides excellent near vision.

The innovation of the Tecnis Multifocal IOL lies in its combination of two optical principles: multifocality and an “intelligent” prolate, anterior, optical surface. Whereas MIOLs have been known for reducing patients' contrast sensitivity, a loss inherent in the optical principle of multifocality, the Tecnis IOL improves contrast by reducing higher-order aberrations compared with a normal monofocal optic. One can therefore anticipate that the multifocal version of this lens will also offer benefits with regard to contrast sensitivity.

The amount of near addition that can be incorporated into either a refractive or diffractive lens appears to be limited. A refractive multifocal lens such as the ReZoom (Advanced Medical Optics, Inc.) has 3.50 D at the lens level, approximately 2.80 D on the corneal plane, and about 2.40 D on the spectacle plane. Diffractive lenses such as the AcrySof® ReSTOR and the Tecnis Multifocal are 4.00 D on the IOL plane, an amount that is normally equivalent to 3.00 D at the spectacle plane. Both IOLs therefore provide a superior near focus compared to that of refractive multifocal lenses; even if the final IOL power is a little off (e.g., +0.25 or even +0.50 D), these diffractive MIOLs still work efficiently.

Accommodating Implants

The excellent results of modern small-incision cataract-IOL surgery have provided motivation to cataract surgeons to restore accommodation. Several manufacturers, ophthalmologists, and vision research scientists are in the process of designing and evaluating accommodative IOLs for placement in the capsular bag of patients to assess restoration of functional accommodation (as opposed to pseudo-accommodation provided by MIOLs) after cataract surgery. Another possible approach that has been experimentally explored in animals is injection of a polymer lens into the capsular bag through a small capsulorhexis after cataract removal.

Based on the design concepts, the accommodating IOLs can be broadly classified in the following three categories.

Accommodating IOL with Single Fixed Optic, Flexible Haptic Support System

In accordance with the Helmholtz theory, one concept for an accommodating lens design (and the simplest) would be a fixed-power single optic with a dynamic, flexible haptic

support system. Some of the accommodative lens designs undergoing clinical trial in adult cataract surgery utilize the concept of a single fixed optic, with a flexible haptic support system. These include the *CrystaLens* 5.0 (Bausch & Lomb), which is a modified plate haptic lens with a 5-mm optic, and the *Akkomodative* 1CU (HumanOptics, Erlangen, Germany), which is a hydrophilic acrylic IOL with a 5.5-mm optic. These IOLs use the anterior movement of the optic to improve near vision. Issues needing further study include dysphotopsia with these small optics, correlation of objective with subjective results, whether the aging eye loses pseudophakic accommodative amplitude and the diminished result expected with low dioptric power IOLs, as well as issues related to postoperative opacification of the capsular bag. In pediatric patients, the postoperative capsular fibrosis may have an effect on the IOL movement with these designs.

Accommodating IOL with Dynamic Optic

Another accommodative IOL concept is a dynamic optic that increases power but maintains its position with accommodation. This design requires flexibility of the optic. One example, the SmartIOL (Medennium, Inc., Irvine, CA), consists only of a full-sized, 9.5-mm diameter by 3.5-mm optic made of a thermodynamic, hydrophobic acrylic material. This IOL is designed to fill the entire capsular bag. Accommodative forces are transmitted to the capsular bag from the ciliary body and, in turn, to the pliable IOL. With accommodative effort, the lens increases its anterior/posterior dimension, thus increasing optical power. The SmartIOL requires continued flexibility of the capsular bag. The lens' hydrophobic acrylic material, its tacky surface, and the fact that it fills the capsular bag had led to predictions that the amount of posterior capsular opacification and capsular fibrosis will be limited. Another feature of this lens, given its flexibility and thermodynamic properties, is that it will be implantable through a 3-mm incision, although the IOL is equivalent to the size of the normal human lens.

A second dynamic optic lens, the Power Vision IOL (Power Vision, Santa Barbara, CA), does not change position within the eye during accommodation. Based upon the use of applied microfluidics, this lens—theoretical at this time—has a peripheral fluid reservoir. Upon accommodative stimulation, an actuator triggers microscopic pumps to move fluid from the periphery to the center of the lens, thereby increasing its anterior/posterior dimension and, hence, its optical power. As accommodation relaxes and the stimulus to near vision decreases, fluid is pumped back from the central to the peripheral aspect of the IOL, thus altering its optical power to the distance mode.

Accommodating IOL with Dual-Optic Design

Visiogen, Inc. (Irvine, CA, now acquired by AMO) has developed a dual-optic, silicone (refractive index 1.43),

single-piece, foldable, accommodating lens called Synchrony™. The IOL features two optics and haptics that have a spring-like action. The haptics separate a high-plus anterior lens from a posterior minus lens. The optical power of the anterior optic is 30.00 to 35.00 D, and the posterior optic is assigned a variable diverging power in order to produce emmetropia for a given eye. The Synchrony™ accommodating IOL system is designed to work in the capsular bag, according to the traditional Helmholtz's theory of accommodation. The distance between the lens optics is minimal in the unaccommodated state and maximum in the accommodated state. Minimal movement results in large refractive changes due to the dual lens design. This lens has been implanted in approximately 120 eyes outside the United States and appears to generate 2.50 D of accommodation, as measured by distance defocus curves.²⁷ Early clinical results are reported to be promising, but only time will tell if capsular contraction decreases the accommodation amplitude over time.

Light Adjustable Lens

The Light Adjustable Lens (LAL, Calhoun Vision, Pasadena, CA) is currently undergoing in vivo study in rabbit eyes as well as preliminary clinical study in human eyes. LAL consists of a silicone matrix into which smaller, photosensitive molecules are embedded.^{28,29} Shining a low-level UV light onto the center of the lens polymerizes the photosensitive molecules, which creates a concentration gradient between the irradiated region where the photosensitive silicone molecules are polymerized and the rest of the optic. Over a 12-hour period, the photosensitive molecules migrate from the untreated areas, down the concentration gradient, and into the irradiated region until there is no concentration gradient. This movement causes the irradiated region to swell and thereby increases the lens power. If instead, we treat the edges of the lens, the photosensitive molecules will migrate outward from the central, untreated region, which will flatten the center of the lens and reduce its dioptric power. Treatment may also be directed along a specific meridian or region of the lens in order to correct astigmatic errors and high-order aberrations.

The amount of energy applied to the LAL determines the degree to which the lens power changes. This dose response is highly reproducible, and in vitro studies demonstrated power adjustments accurate to within <0.25 D. One day after the power adjustment, the patient returns to check to see if the lens has the desired power. If satisfactory results are obtained, the entire lens is irradiated, which polymerizes all of the remaining photosensitive silicone molecules and locks the lens. After this lock-in step, the LAL is essentially a standard silicone IOL. Before lock-in, the LAL can be readjusted, a fact that will make it possible for the patient to try monovision or multifocality and then have that modality removed if it

proves undesirable. Theoretically, lock-in may be avoided and the lens adjusted over time as changes in refractive precision occur that would be perfect for pediatric cases.

SUMMARY

The success and excellent results of current small-incision cataract-IOL surgery have provided motivation to manufacturers, ophthalmologists, and vision research scientists to design and evaluate multifocal, accommodative, and other new technology IOLs to potentially restore accommodation and improve the quality of vision. Visual rehabilitation in children in the industrialized world is now being revolutionized by the implantation of the IOL. Highly refined and perfected microsurgical techniques have now propelled the cataract-IOL procedure in adults toward being one of the most successful surgical techniques in history. Application of this procedure to children is now improving the treatment of pediatric cataract. Surgeons are now applying the best tried and true techniques perfected over the years for adult cataract surgery. They are modifying these to meet the specific characteristics of the infantile eye.

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Primary Intraocular Lens Implantation in Children

Rupal H. Trivedi and M. Edward Wilson

Primary intraocular lens (IOL) implantation has become the most common method used to correct aphakia in children beyond their first birthday (Figs. 25.1 through 25.4). However, it is still considered an “off-label” (or “physician directed”) use by the U.S. Food and Drug Administration (FDA). It provides the benefit of at least a *partial but uninterrupted optical correction*, which is an important advantage to the visual development in amblyopia-prone eyes. Uncorrected pseudophakic vision is probably less amblyogenic than uncorrected aphakic vision. While there is certainly a benefit to IOL implantation by reducing the dependency on compliance with other external optical treatments (aphakic glasses and contact lenses), it was slow to catch on in children as compared with adults. This is because many early attempts at IOL implantation in children resulted in severe complications. Lack of awareness of the need for primary posterior capsulectomy and vitrectomy, poor IOL design, and the inherent propensity for an increased inflammatory response that occurs in a child’s eye after intraocular surgery resulted in this higher rate of postoperative complications. IOL implantation in children began to increase steadily only after 1990, even though its first reported use was in 1952 as a secondary IOL in a 12-year-old aphakic child.¹ The use of mechanized vitrectomy instrumentation to perform a primary posterior capsulectomy and vitrectomy combined with implantation of better-quality IOLs resulted in fewer complications in children. This encouraged more and more surgeons to use IOLs for children and to gradually decrease the lower limit for age of implantation. Binkhorst and Hiles pioneered the use of IOLs in children in the 1970s and 1980s.^{2,3} Surgeons were initially hesitant to implant adult-size IOLs (even high-quality IOLs) in the pediatric age group until a number of studies began to show their safety and efficacy in children.⁴⁻⁷ Although IOL implantation in children was mostly investigational before the mid-1990s, since that time, its use has risen dramatically among pediatric cataract surgeons. In 1996 editorial, Wilson⁸ stated that IOL implantation had become the standard of care for uncomplicated cataract surgeries

in children beyond infancy. The dramatic increase in acceptance of IOL implantation in children was noticeable when comparing our 1993 practice preferences survey data with that of our 2001 practice preferences survey. We showed a nearly fivefold increase in the number of respondents from the American Society of Cataract and Refractive Surgeons (ASCRS) and more than a 13-fold increase in the number of respondents from the American Association for Pediatric Ophthalmology and Strabismus (AAPOS) implanting IOLs in children ≤ 2 years.⁹

IOL IMPLANTATION IN INFANTS

As primary IOL implantation has become more broadly used, the age in which the procedure is acceptable and appropriate has steadily decreased. IOL implantation is common for children undergoing cataract surgery beyond infancy (Fig. 25.5). For bilateral cataract during infancy, aphakic glasses and/or contact lens use may be a reasonable option; however, for unilateral cataract, we were equipoised between whether or not to offer primary IOL implantation at the time of infantile cataract surgery. The Infant Aphakia Treatment Study (IATS) was a multicenter, randomized, controlled clinical trial comparing the outcomes of contact lens versus IOL correction for monocular aphakia during infancy. The 1-year results were published in 2010 and the 5-year results are being published in 2014.¹⁰ In the 1-year outcome study report, the authors advised that until longer-term follow-up data are available, caution should be exercised when performing IOL implantation in children aged 6 months or younger given the higher incidence of adverse events and the absence of an improved short-term visual outcome compared with contact lens use.¹⁰

Table 25.1 describes the arguments for and against primary IOL implantation in infantile cataract surgery. The important concerns about primary IOL implantation during infancy are *the technical difficulties of implanting an IOL and selecting an IOL power, and the higher rate of visual axis opacification (VAO)*. Despite performing primary posterior capsulectomy and vitrectomy, the rate of

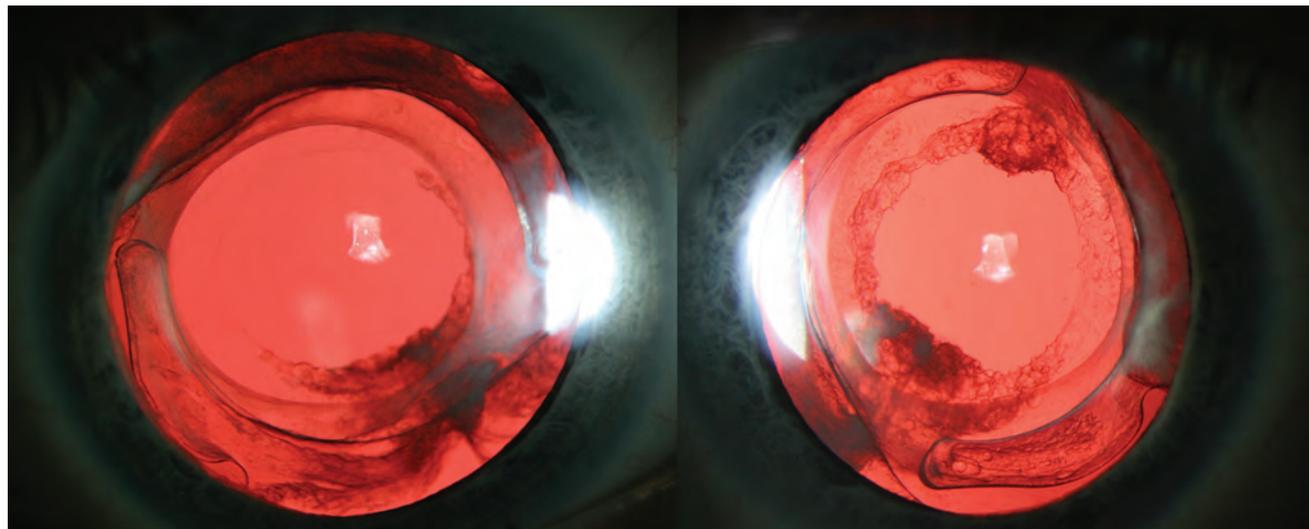


Figure 25.1. Eight-year follow-up of a child operated for bilateral cataract surgery using vitrectorhexis, posterior capsulectomy, vitrectomy, and in-the-bag AcrySof® SA60AT IOL implantation at 4 years of age.

VAO is higher in pseudophakic infantile eyes as compared with aphakic infantile eyes.^{11,12} The IATS reported that at the 1-year visit, more patients in the IOL group had undergone one or more additional intraocular operations than did patients in the contact lens group (63% vs. 12%; $P < 0.001$). Most of these additional operations were performed to clear lens re proliferation and pupillary membranes from the visual axis.¹⁰ For IATS patients up to 12 months of age, cataract surgery coupled with IOL implantation and spectacle correction was 37.5%

(approximately \$4,000) more expensive than was cataract surgery coupled with contact lens correction.¹³

The main concerns of contact lens correction are *poor compliance, high lens loss rate, high cost, and keratitis* (Fig. 25.6). These challenges make contact lenses impractical in many children especially in most parts of the developing world. It is certainly possible for an eye with a unilateral infantile cataract to achieve a good visual outcome following contact lens correction. However, it has continued to be the exception rather than the rule when contact lens

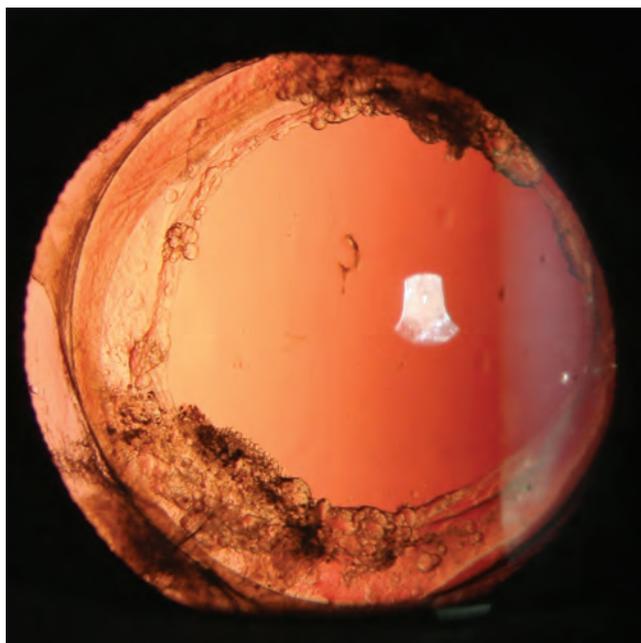


Figure 25.2. Ten-year follow-up of a child operated for posterior polar cataract in his left eye at 5 years of age. Surgical technique included Kloti bipolar anterior capsulotomy and posterior capsulectomy, vitrectomy, and in-the-bag implantation of AcrySof® IOL.

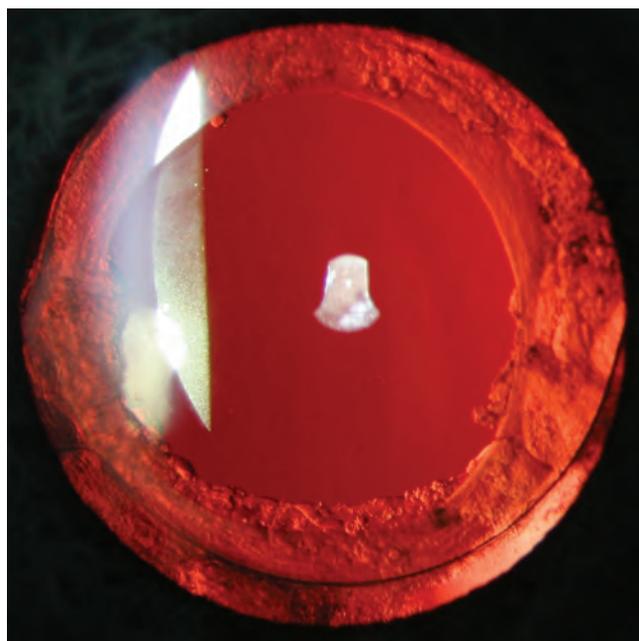


Figure 25.3. Six-year follow-up of a girl operated for cataract in her left eye at 2.5 years of age. Surgical technique included vitrectorhexis, posterior capsulectomy, vitrectomy, and in-the-bag IOL implantation.

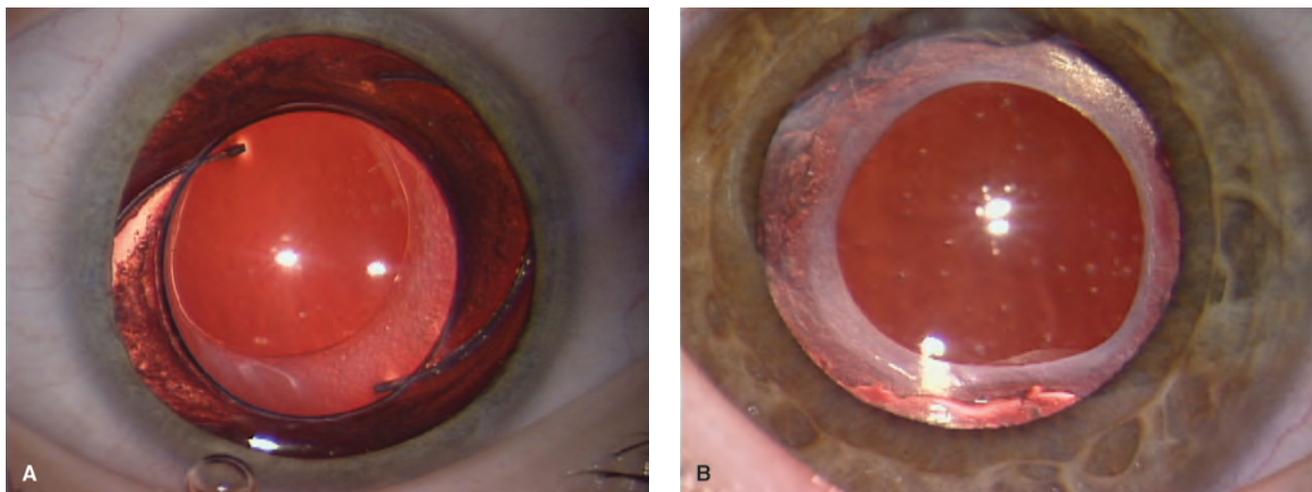


Figure 25.4. Postoperative follow-up showing a clear visual axis. **A:** Note haptic compression and peripheral capsular changes. **B:** Note well-centered IOL and anterior capsule opacification.

losses are high. Both IOLs and aphakic contact lenses may support similar visual acuity after surgery for unilateral cataract in the presence of good compliance and a low loss of contact lenses. However, IOLs support better visual acuity in those situations when compliance with contact lens wear is moderate or poor or when contact lens replacements/refittings are a hardship because of cost or transportation difficulties.¹⁴

Contact lens correction is the most commonly chosen option for aphakic correction in early infancy in our practice, especially in bilateral cataract patients. In unilateral cases, primary IOL implantation is more often chosen when compared to bilateral cases. This is especially true in families in which contact lens care is predicted to be difficult. In our state, Medicaid does not pay for contact lenses even when we write letters stating that the contact lenses are medically necessary. We prefer silicone SilSoft contact lenses; however, these lenses cost more than U.S. \$150 each and are often lost or in need of frequent replacement. Many poor families will not be able to afford this expense. In these families, IOL implantation with glasses for residual hyperopia is given more consideration since it may be the only viable option when the cataract is unilateral. To its credit, Bausch & Lomb SilSoft Contact Lens Patient Assistance Program (<http://www.bausch.com/Search%20Results?searchtext=Silsoft+Contact+Lens+Patient+Assistance+Program>) has worked well for us. Typically when patients lose a lens, they are told to call our central scheduling number during working hours, and then one of our pediatric ophthalmology clinic technicians return the call and determine if they have the means to purchase a replacement contact lens. If so, we send a script and inform them of the online ordering sites, such as Walgreens.com. If we determine that they cannot afford

to replace the lens, we order one for them through the Bausch & Lomb Patient Assistance Program by filling out an online form. We explain to the family that if they qualify for the Bausch & Lomb program, they can receive up to one free SilSoft lens order (maximum two lenses per order) every 3 months but that a new form must be filled out for each new order. The free lenses are shipped to us, and we mail them to the family or insert them at the next office visit.

WHAT IOL TO IMPLANT?

Type of IOL

Since posterior capsule opacification (PCO) is one of the most frequent and severe complications in pediatric cataract surgery, an IOL that decreases the incidence of PCO is more often used in pediatric cataract surgery.^{15,16} The choice of an IOL material and type for children is a modified version of what has evolved for adults. A biocompatible material that could be inserted via a smaller incision and fit the smallest capsular bag without excessive stretching is preferred.¹⁷ Our 2001 survey of ASCRS and AAPOS members reported that 66.8% of ASCRS respondees and 71.7% of AAPOS respondees preferred hydrophobic acrylic IOLs when implanting lenses in children.⁹ A later survey of AAPOS members conducted in 2006 showed that for in-the-bag fixation, hydrophobic acrylic IOLs were preferred by 93.3% of the respondents.¹⁸ AcrySof[®] hydrophobic acrylic lenses were preferred by 90.2% of respondents. Among AcrySof[®] users, more physicians preferred the single-piece AcrySof[®] lens for bag fixation of an IOL. For ciliary sulcus fixation of an IOL, hydrophobic acrylic IOLs were preferred by 69.9% of the respondents, and the AcrySof[®] hydrophobic acrylic IOL was preferred by 65.3% of overall respondents.

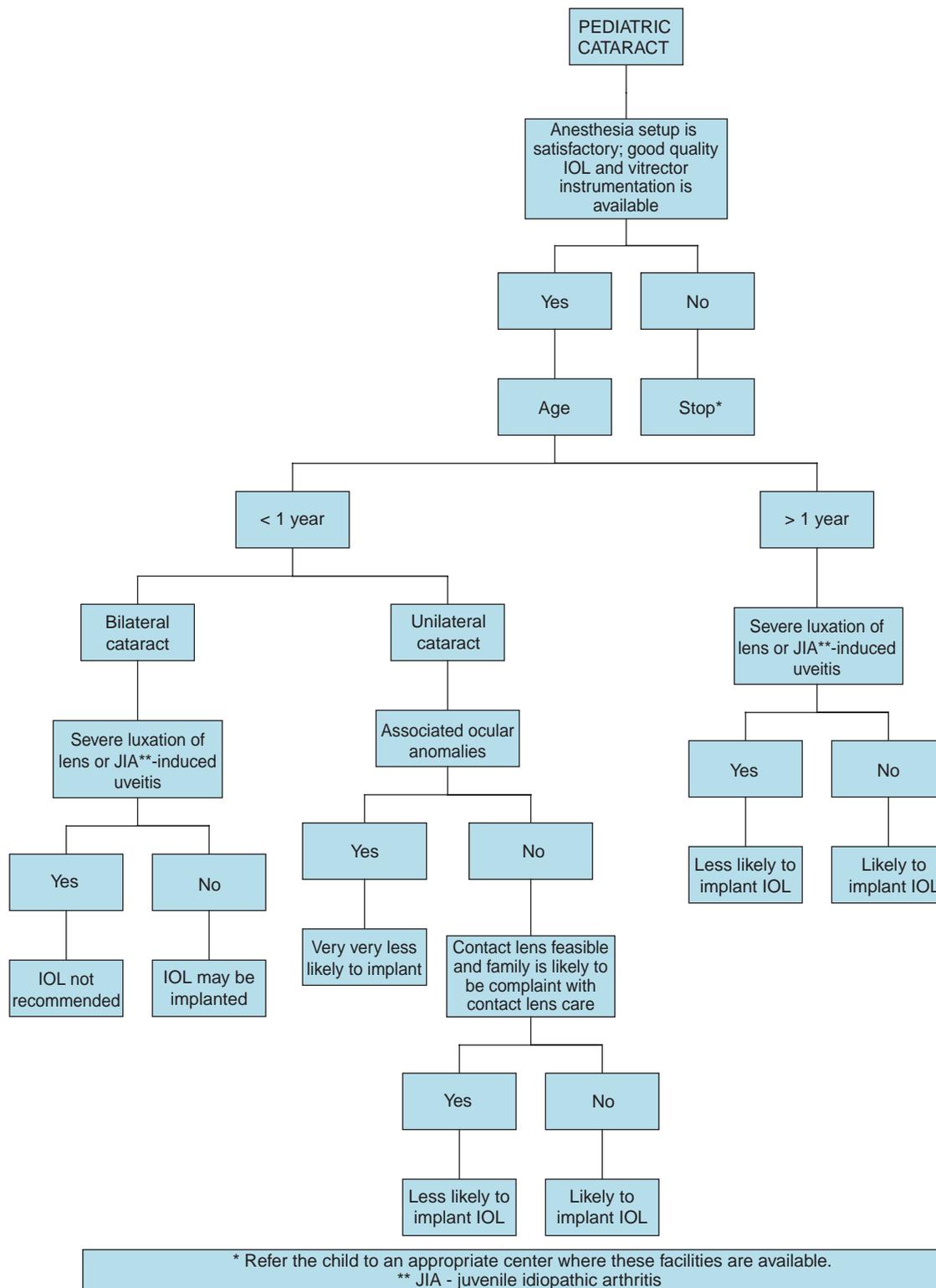


Figure 25.5. Flow diagram of our current considerations when determining whether or not to offer an IOL as treatment for a child with a cataract.

Table 25.1 ARGUMENTS FOR AND AGAINST PRIMARY IOL IMPLANTATION IN INFANTILE CATARACT SURGERY

Proponents of Contact Lens	Proponents of IOL Implantation
1. The power of a contact lens can be easily changed to compensate for the rapid myopic shift that occurs during the first 2 years of life. An IOL can be implanted in these eyes when these children are older and their refractive errors have stabilized.	1. Compliance with contact lens wear is a major concern. Noncompliance may result in dense amblyopia by the time their refractive error stabilizes.
2. Implanting an adult-size IOL in the small capsular bag of an infant is technically difficult.	2. With the availability of foldable IOLs and injectors, the success of implanting in-the-bag is relatively high.
3. IOLs in infantile eyes lead to a high rate of reoperations for VAO.	3. Eyes with aphakia more often need other surgery, for example, strabismus or secondary IOL. If we consider total reoperations, the reoperation rate may be similar in both the groups.
4. A good visual outcome is possible if a child is compliant with contact lens wear and occlusion therapy.	4. Compliance with contact lens wear is a major hurdle. Uncorrected pseudophakic vision is probably less amblyogenic than uncorrected aphakic vision.
5. The long-term safety of an artificial lens is not known.	5. Contact lens wear, long-term, can lead to corneal vascularization, and a risk for corneal ulceration and scarring.

Among those implanting AcrySof[®], most preferred the three-piece AcrySof[®] IOL for sulcus fixation. There are several published case series detailing the use of hydrophobic acrylic IOLs in children.^{9,12,15,17,19–40} Wilson et al.¹⁵ reported that the YAG laser capsulotomy rate was 45.4% for AcrySof[®] and 50% for PMMA IOL. None of the patients with AcrySof[®] IOLs have needed more than one YAG capsulotomy. Five of the patients with PMMA IOLs have needed multiple YAG laser treatments for recurrent opacification ($P \leq 0.05$).

Vasavada et al.³³ evaluated 103 consecutive eyes of 72 children with pediatric cataracts. Two groups were formed based on age at surgery: group 1, younger than 2 years, and group 2, older than 2 years. All eyes in group 1 ($n = 37$) had primary posterior continuous curvilinear capsulorhexis (PCCC) with anterior vitrectomy. In group 2 ($n = 66$), management of the posterior capsule was assigned randomly to no PCCC (group 2A; $N = 37$) or PCCC (group 2B; $N = 29$). The PCCC group was further randomized into two subgroups: no vitrectomy (group 2BN; $N = 14$) or vitrectomy (group 2BV; $N = 15$). The primary outcome measures were

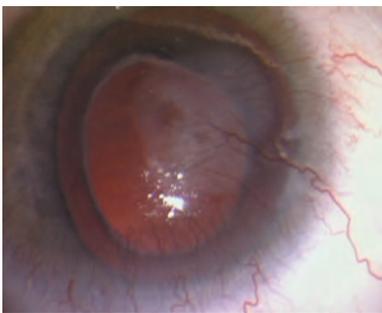


Figure 25.6. A healed corneal ulcer in an eye followed by the use of contact lens for correction for pediatric aphakia. Note the corneal vascularization.

VAO and the resulting need for a secondary procedure. The mean age of the patients was 5.2 ± 5.0 years (range, 0.2–16.0 years), and the mean follow-up was 2.3 ± 0.9 years (range, 1.0–4.0 years). Overall, 41 eyes (39.8%) developed VAO and 14 (13.6%) eyes required secondary intervention. In group 1, four eyes (10.8%) developed VAO and three (8.1%) eyes had a secondary pars plana vitrectomy. In group 2A, 31 eyes (83.8%) developed PCO and 10 eyes (27.7%) had secondary intervention. The incidence of PCO was highest between 2 and 3 years postoperatively. In group 2BN, five eyes (37.5%) had opacification of the anterior vitreous face, one of which required a secondary procedure 2.7 years after cataract surgery. In group 2BV, 1 (6.7%) of 15 eyes had VAO with a fibrous band across the visual axis in front of the IOL, which did not require a secondary procedure as of the last follow-up.

IOL Power

With a growing eye prone to develop a myopic shift of refraction after cataract removal, the surgeon faces the decision of what refraction is the immediate postoperative target.^{41–44} *Implantation of a fixed-power IOL into an eye that is still growing makes it difficult to choose the IOL power to implant. IOL implantation at the calculated emmetropic power helps to fight amblyopia during childhood but risks significant myopia at ocular maturity. An ideal IOL power would balance the best help to amblyopia management in childhood with the least possible refractive error in adulthood.* Chapter 7 provides details on IOL power calculation. Several nomograms have been published in the literature. However, we do not recommend relying solely on any published guideline table when deciding IOL power. These tables are only meant to help as a starting point toward appropriate IOL power selection, which is a multifactorial decision customized for each child based on many variables

(especially age, laterality [one eye or both], amblyopia status [dense or mild], compliance with glasses, and family history of myopia). When implanting children with bilateral cataracts and no amblyopia, leaving mild, moderate, or even a marked amount of hypermetropia is reasonable. However, in unilateral cataracts with dense amblyopia, less early dependence on glasses may help the amblyopia treatment. The late myopia, even if marked, may be an acceptable trade for better visual outcome from amblyopia treatment. Refractive surgery or IOL exchange may be needed in these eyes at ocular maturity. As IOL selection and refractive shift management evolves, we predict that a multiple procedure approach will become more common. Minimizing refractive error early and continuing to have a minimal refractive error later will be the goal. To achieve this in the young patient, either a temporary piggyback primarily, a secondary piggyback later, or a planned IOL exchange may be needed.

PREOPERATIVE COUNSELING

Before moving forward with lens implantation, it is important to discuss with the parents/legal guardian the major pros and cons of the available treatment options. The lion's share of this discussion of pediatric cataract surgery will be regarding issues related to the IOL. Surgeons should be prepared for the common question "Would you implant an IOL if this were your child?" A coordinated plan of action is best developed when the parents understand the reasons for, goals of, and the advantages and potential complications of cataract surgery with implantation. When properly informed preoperatively, the parents and the physician become partners with the common goal of doing what is best for the child, given the age, level of maturity, family situation, travel distance to the doctor's office, and many other unique considerations. Time spent establishing this partnership is not wasted, because a better-informed family is much more likely to comply with the frequent follow-ups, medications, patching, glasses wear, etc., that are so essential to the eventual visual outcome. A child operated for cataract (with or without IOL implantation) requires regular scheduled care for the first decade of life, and then every 1 to 2 years throughout life. For eyes operated during early infancy, parents should be made aware that the first 6-month follow-up is very crucial. Most eyes that develop VAO do so in the first 6 postoperative months (Fig. 25.7). Earlier detection (and treatment if needed) can help to achieve a better visual outcome. For eyes operated with an intact posterior capsule, parents should be made aware that the child is likely to require a secondary procedure for PCO 1 to 2 years after the surgery. Parents of children with lens implants are also made aware that glasses will likely still be needed postoperatively even when an IOL is implanted. In addition, glasses power may need to

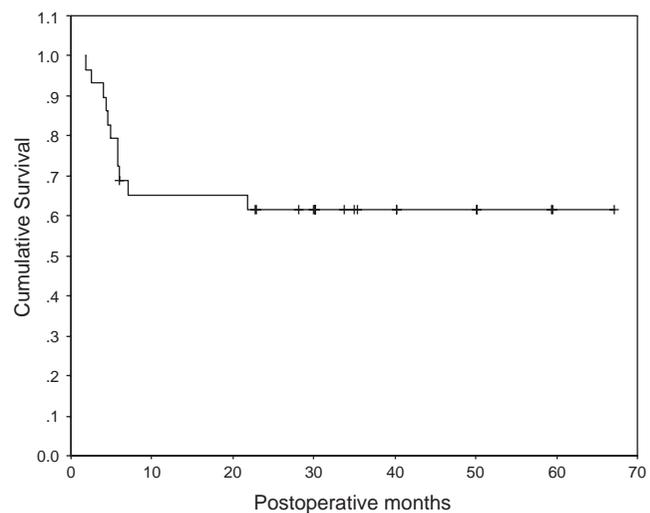


Figure 25.7. Kaplan-Meier curve representing the secondary surgery-free survival of eyes without VAO following primary cataract surgery at <1 year of age and showing excellent maintenance of visual axis clarity after 6 postoperative months. (Reprinted from Trivedi RH, Wilson ME Jr, Bartholomew LR, et al. Opacification of the visual axis after cataract surgery and single acrylic intraocular lens implantation in the first year-of-life. *J AAPOS* 2004;8:156–164, with permission.)

be changed frequently after surgery because of changing refraction.

In the United States, parents should be made aware that while IOL implantation has become the most common method used to correct aphakia in children overall, it is still considered “off label” by the FDA. This designation means that the IOLs implanted in children were tested as part of their FDA market approval process, but only in adults. This does not mean that the FDA has disallowed their use in children. It only implies that the device is being used for a purpose or in a patient population that is different from the one in which it was tested as part of the market approval process.⁴⁵ The term “physician directed” is sometimes substituted for “off label” to better explain that the devices are market approved but are being used in a physician-directed manner that has been documented in many peer-reviewed scientific papers but has not been formally reviewed or tested by the FDA.

INTRAOPERATIVE CONSIDERATIONS

Surgical Technique

General principles of pediatric cataract surgery discussed throughout the book should be used. In the following section, we have paid particular attention to issues focused on IOL implantation.

Incision

A soft cataract in children can be easily aspirated through a sub-1-mm incision when an IOL is not being implanted. However, IOL implantation calls for

a relatively large wound, for example, 2.2 to 3.0 mm depending on the IOL design.

Anterior Capsulorhexis

The anterior capsule in children is highly elastic and poses challenges in the creation of the capsulotomy. While a CCC is a reasonable option at any age, CCC will be difficult to control for even the experienced surgeon when attempted on the elastic capsule of an infant eye. The vitrectorhexis is an alternative; however, care should be taken to avoid leaving any right-angle edges, which could predispose to radial tear formation during IOL implantation. When primary IOL implantation is the surgical preference, a CCC is the better option. However, when IOL is not going to be implanted, vitrectorhexis can be chosen as a procedure of choice. The size of the completed anterior capsulotomy should be 1 mm smaller than the size of the IOL optic being implanted. If an IOL is not going to be implanted, a slight larger capsulotomy is acceptable. We should leave sufficient capsular support in anticipation of future secondary IOL placement.

Posterior Capsule Management

When an IOL is not intending to be implanted, surgery can be simplified by performing irrigation/aspiration with the vitrector handpiece (cutter off) and then posterior capsulectomy/vitrectomy (cutter on) through a limbal approach without ever having to remove the instruments from the eye. When an IOL is to be implanted, we most often use a pars plicata/plana approach to perform a posterior capsulectomy after IOL implantation in the bag. This approach allows the IOL to be inserted while the posterior capsule is still intact. The OVD is removed, and then the pars plicata/plana approach can be used for opening the posterior capsule to remove capsule plaque or to perform a standard posterior vitrectorhexis (Fig. 25.8). Preexisting posterior capsule defects can be converted to a posterior vitrectorhexis (Fig. 25.9). This technique helps assure that the IOL is verified to be completely within the capsular bag and allows for the posterior vitrectorhexis with minimal displacement of the implanted IOL.

IOL Implantation

In addition to the smaller eyes, decreased scleral rigidity makes IOL implantation more challenging in infants compared to older children. Also, infantile cataracts are often associated with microphthalmos and anterior segment anomalies. We use single-piece acrylic IOLs. Single-piece IOL implantation with an injector has markedly reduced implantation-related issues in children, even in microphthalmic eyes. These soft IOLs can be injected precisely into the capsular bag and can also be more easily repositioned (compared to older rigid IOLs or multipiece IOLs) in the eye if necessary. When placing an IOL in a child's eye, in-the-bag implantation is strongly recommended.

When capsular fixation is not possible, sulcus placement of an IOL in a child is acceptable for us; however, we know that some surgeons would argue differently. We use a three-piece AcrySof[®] IOL or Rayner c-flex IOL for sulcus fixation. Optic capture of an IOL maintained better IOL centration but was reported to predispose to an increased inflammatory response.⁴⁶ Care should be taken to avoid asymmetrical fixation with one haptic in the capsular bag and the other in the ciliary sulcus.

INTRAOPERATIVE COMPLICATIONS

Primary IOL implantation does make surgery in young patients more difficult as compared to no IOL surgery. Details of intraoperative complications have been discussed in Chapter 49. Herein, we briefly describe issues pertinent to IOL implantation. An intact anterior capsulotomy may tear during IOL implantation, especially if it is too small and especially if it is a vitrectorhexis. To avoid this complication, aim for a 5-mm capsulorhexis using a manual tear technique when possible. If a vitrectorhexis is used, avoid leaving any right-angle edges and minimize stress and stretch of the capsulotomy edge when inserting and positioning the IOL. When using a vitrectorhexis, an anterior capsule tear may develop if the chamber temporarily flattens at the end of the case when irrigation is withdrawn before the suture can be tied securely. To avoid this problem, an air bubble can be used to hold the IOL back and stabilize the anterior chamber while tying the final stitch.

IOL implantation may at times become difficult. The IOL may dislocate into the vitreous cavity. This complication is minimized if the IOL is inserted before the posterior capsulectomy and vitrectomy are performed. The occurrence of iris prolapse is more frequent when an IOL is inserted since a larger wound is needed. This complication results from a tunnel wound that is not extended far enough into clear cornea before entry. The placement of the internal lip of the wound is the key to preventing iris prolapse. In addition, young children have a naturally floppy iris that can prolapse into even properly placed entry wounds. Placing epinephrine in the irrigation fluid will help minimize the consequences of the floppy iris.

POSTOPERATIVE MEDICATIONS

In our practice, babies who are left aphakic do not receive the ointment, patch, and shield that are placed when an IOL is inserted. We use topical drops for these eyes, and rather than patching the eye, we apply a SilSoft contact lens at the end of the surgery. The parents can then begin the drops right away. In contrast, eyes receiving a primary IOL implantation receive an antibiotic-steroid ointment at the end of the surgery followed by a patch and

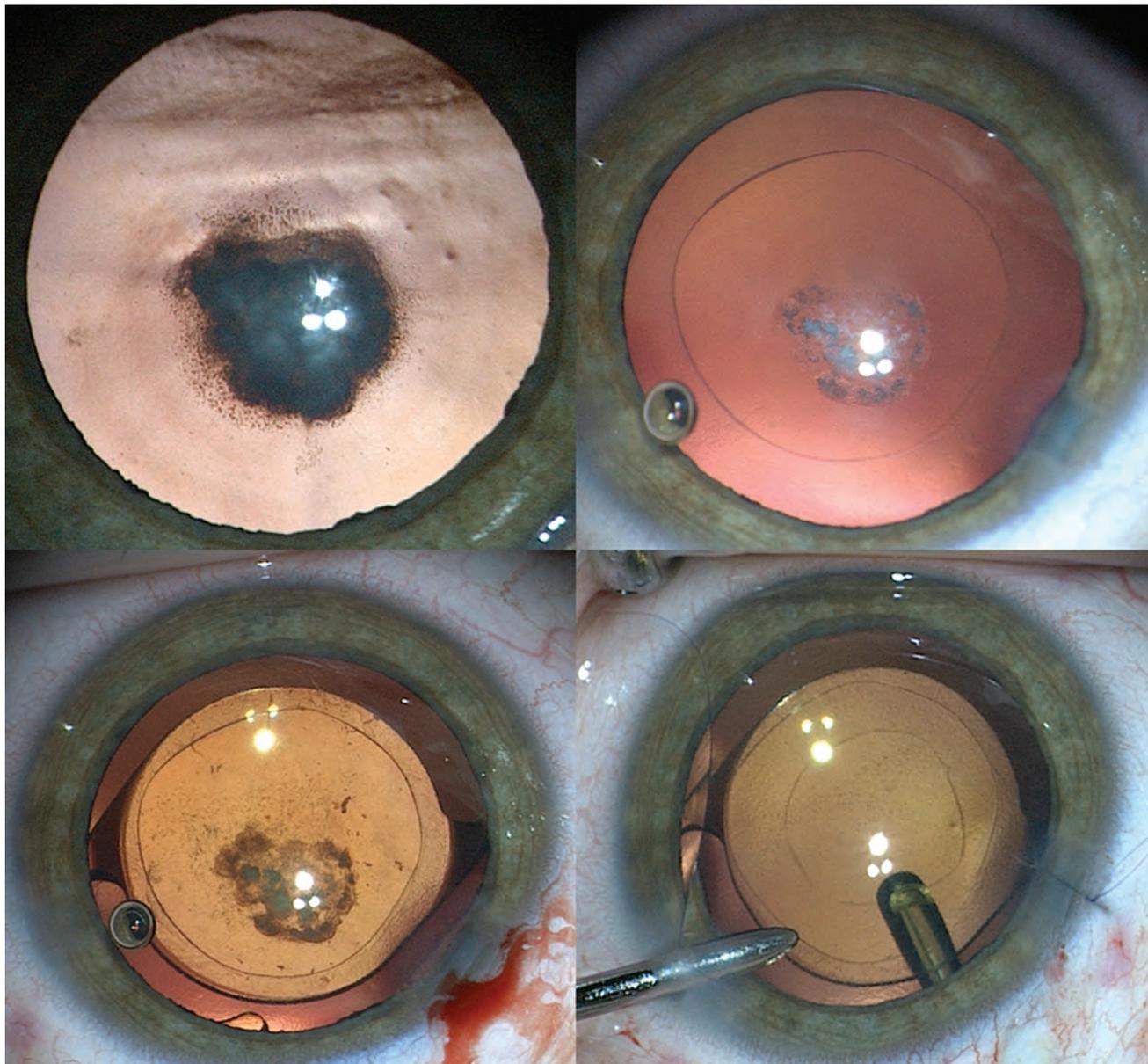


Figure 25.8. Left eye cataract removal in a 3-year-old child. Note posterior capsule plaque.

shield. Atropine ointment is added for children aged 2 and below (older children often do not receive atropine). Dilute (5%) povidone-iodine is placed on the eye after intracameral antibiotics in our cases whether an IOL is placed or not.

POSTOPERATIVE ISSUES

Postoperative complications are discussed elsewhere in this book. Herein, we described it briefly.

Visual Axis Opacification

As discussed above, eyes receiving an IOL implantation are at higher risk of postoperative VAO. The reason for

this increased risk is now clear. We have learned that when surgery is done very early in life, lens epithelial cells remain in the lens equator despite very precise and complete cortical cleanup during lens aspiration. These cells are in an active growth phase and continue to produce lens proteins after surgery. This cortex repopulation varies somewhat, patient to patient, and usually stays trapped in what becomes a Soemmering ring. In eyes with an IOL, the capsule may not seal as well to the IOL as it can to itself. This may allow some of the new lens cortex to reach the central visual axis. Also, it has been hypothesized that the IOL may act as scaffolding to which fibrous tissue can attach. In our experience, VAO in infants does not involve attachment of

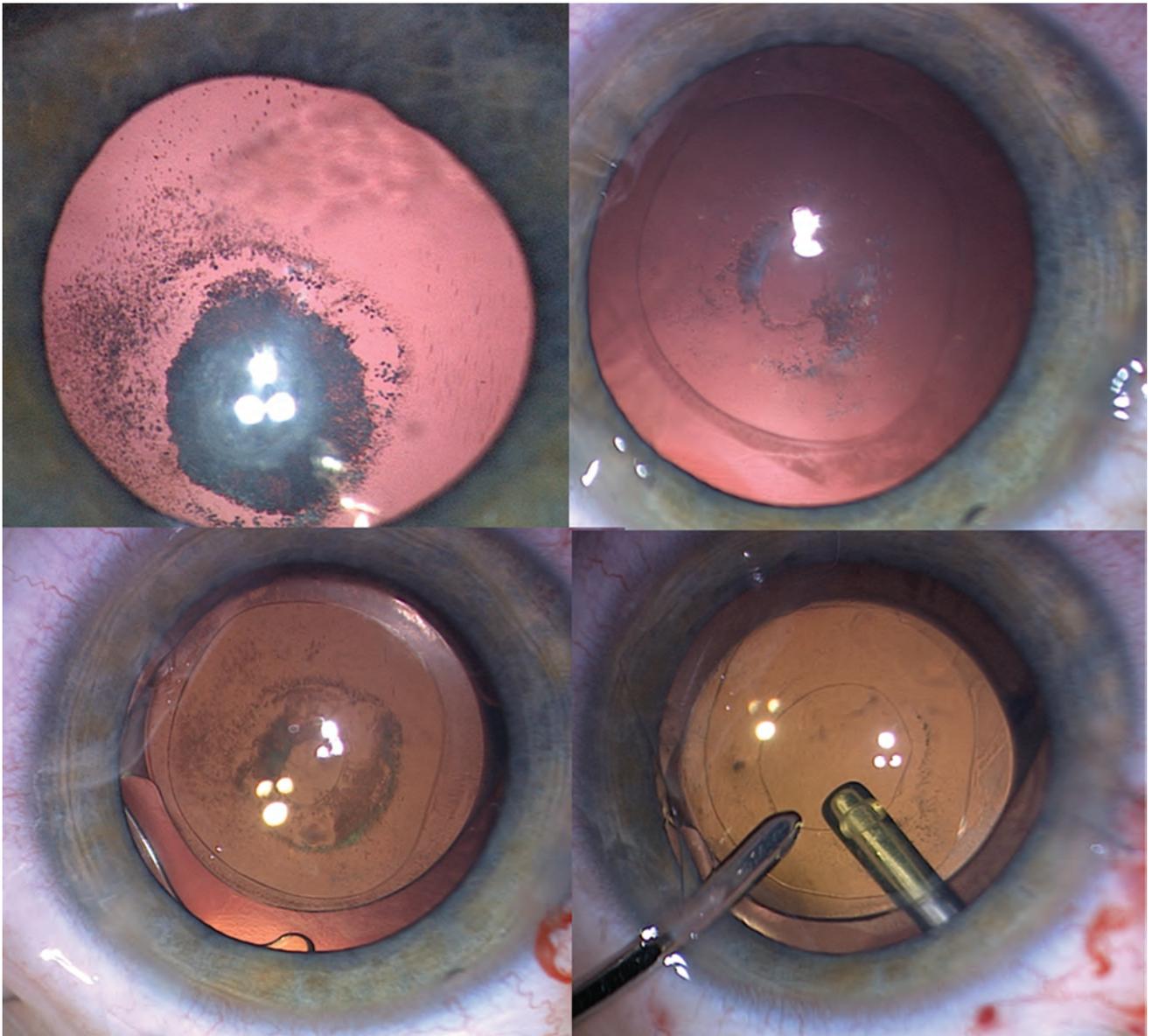


Figure 25.9. Surgery of a 6-year-old girl operated for cataract. Note preexisting posterior capsule defect.

cells or membranes to the IOL itself. VAO after AcrySof® implantation with an intact posterior capsule is more “proliferative,” as opposed to the “fibrous” reaction commonly seen in conjunction with PMMA IOLs. The proliferative VAO may progress more slowly, be less visually significant, and require secondary intervention less often. After a primary posterior capsulotomy and an anterior vitrectomy, VAO is rare in children operated over age 1. When it does occur, it is usually in a baby operated on in the 1st year of life and tends to occur within the first 6 months after surgery (if it is going to occur). The AcrySof® IOL has been documented to stimulate what has been called an “anterior vitreous reticular response” when exposed to the vitreous face after pediatric cataract surgery and posterior capsulotomy but no vitrectomy.

This finding on the anterior vitreous face was usually not visually significant.

Glaucoma

Cataract surgery in the 1st year of life has been the most consistent risk factor for glaucoma development. Earlier studies reported a low incidence of glaucoma in pseudophakic eyes, the implication being that pseudophakia in children somehow protects against glaucoma. However, this was perhaps due to selection bias.⁴⁷ We reported that patients undergoing cataract surgery at an early age are at high risk for the development of glaucoma with or without glaucoma.⁴⁷ The IATS also noted that of the 57 patients who underwent lensectomy and anterior vitrectomy, 5 (9%) developed a glaucoma-related

adverse event; of the 57 patients who underwent an IOL implant, 9 (16%) developed a glaucoma-related adverse event.⁴⁸

Axial Elongation

The IATS reported that the change in axial length/month was smaller in operated eyes treated with a contact lens than in operated eyes treated with an IOL (0.17 vs. 0.24 mm, $P < 0.0001$).⁴⁹ This topic is more fully explored in Chapter 52.

Adherence to Occlusion Therapy during the Early Postoperative Period

The IATS reported that the type of correction (IOL vs. contact lens) was not associated with the amount of patching achieved, whereas family socioeconomic status and maternal stress appeared to play a role.⁵⁰

Visual Outcome

As mentioned earlier, an IOL provides the benefit of at least a *partial but uninterrupted optical correction*, which is an important advantage to the visual development in amblyopia-prone eyes. However, the 1-year IATS reported that the median logMAR visual acuity was not significantly different between the treated eyes in the two groups (contact lens group, 0.80; IOL group, 0.97; $P = 0.19$).¹⁰ We await the long-term visual outcome of this cohort, when children are cooperative for Snellen visual acuity measurement.

SPECIAL SITUATIONS

Developing World Setting

Primary implantation of an IOL during cataract surgery in the developing world seems to be the practical option of choice, while contact lenses are less suitable in most of these settings. Cost is the major issue of using hydrophobic acrylic foldable IOLs. In addition, the limited availability of modern-day OVDs makes implantation of acrylic IOLs into the capsular bag of small eyes more difficult. When using stiffer IOLs, less viscous OVDs, less precise biometry, and lower-quality microscopes, ciliary sulcus implantation of an IOL may be advisable. The sulcus IOL placement is easier when visualization is poor. It allows the capsular bag leaflets to seal, trapping reepithelialized cortex (if any is produced—more likely with young patients or when cortex aspiration is incomplete) in the Soemmering ring. Therefore, the reoperation for VAO may be less. With imprecise biometry, IOL exchange at a later age is much easier with sulcus IOLs than with capsular fixated IOLs. Care must be taken to avoid IOL pupil capture. Also, only IOLs appropriate for sulcus placement should be used if this technique is chosen.

Absent Capsular Support

In cases of poor capsular support, such as in ectopia lentis or traumatic zonular instability, options include placement of an angle-supported anterior chamber IOL, a scleral-fixated IOL, or an iris-fixated IOL (Chapter 32).

Bag-in-the-Lens Implantation

Tassignon et al.⁵¹ reported the outcome of a surgical procedure they called “bag-in-the-lens” in pediatric cataract patients. In this technique (see Chapter 27), the anterior and posterior capsules are placed in the groove of a specially designed IOL after a capsulorhexis of the same size is created in both capsules.^{51,52} The authors reported a clear visual axis in all pediatric patients with an average follow-up of 17 months.

Multifocal and Accommodating IOL Implantation

See Chapter 29. Although multifocal IOLs are being used commonly in adults, they have been mostly restricted to older end (late teens) of the pediatric age group. These lenses are not often utilized in young children because of the myopic shift of the pediatric eye over time and the potential consequences of contrast sensitivity loss and photopsia on the ongoing amblyopia treatment. We predict that multifocal IOLs will disappear from the market when truly accommodating IOL are developed that have a large range of predictable accommodation. Multifocality (eventually) will be remembered historically as an intermediate stage between monofocal implants and those that produce smooth and more natural accommodation.

Adjustable IOLs

The ongoing development in adjustable IOL technology may prove very useful in the future of pediatric cataract surgical management.⁵³ The possibility of a lens that could be adjusted to counter the myopia induced by ocular growth is exciting. Several designs of such a lens have already been described: (1) by changing the actual refractive power of the IOL, such as the light-adjustable Werblin lens system and (2) by changing the position of the optic of the IOL with respect to the point of focus, such as a screw-based optic lens or a piston-based movable optic lens. Owing to its ability to provide changes in IOL power,^{54,55} the Light Adjustable™ lens (Calhoun Vision, Pasadena, CA) may have potential applicability for pediatric eyes. Early in 2007, the light-adjustable lens was approved for use in Europe with the full CE mark. Clinical studies in the United States are ongoing. Jahn and Schopfer⁵⁶ described a mechanically adjustable PMMA IOL that necessitates intraocular manipulation via two paracenteses. Follow-up extending to 18 months in adult patients shows encouraging results; however, the

authors note a higher rate of PCO in the study group, as compared to the control group. An ideal adjustable IOL implant should be biocompatible; allow for safe repeatable adjustment procedures performed at any time after cataract surgery; be precise, with stable correction of myopic, hyperopic, and astigmatic refractive error; and have an adequate refractive error range. As of today, the ideal adjustable IOL does not exist; however, the concept of such an IOL is being developed.

SUMMARY

IOL implantation has become the standard of care for the optical rehabilitation of children beyond their infancy. IOLs are a viable alternative even for young infants, but aphakia with plans for a secondary IOL after some years of eye growth and development will likely remain the preferred option when it is feasible. IOL implantation has the benefit of providing at least a partial optical correction at all times and a potentially improved functional outcome, but this gain must be balanced against the high reoperation rate for VAO and the higher rate of IOL exchange later in childhood when the large myopic shift has occurred. With all that said, we must still remember that an IOL implanted in a child's eye must be stable enough to stay there for several decades, perhaps 70 years or more, without biodegrading. To date, modern acrylic IOLs have been found to be efficacious in providing good short- to intermediate-term results after implantation in congenital, juvenile, and traumatic cataract surgery. Long-term outcome will remain an open question for the years to come. In children beyond infancy, combined posterior capsulectomy, vitrectomy, and modern IOL implantation eliminates the need for a secondary intervention in most eyes. Slowly, IOL implantation guidelines are being cautiously updated for the more complicated pediatric cases, including those with compromised capsular support, complex microphthalmia, persistent fetal vasculature, and a history of uveitis, etc.

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Role of Intraocular Lens Optic Capture in Pediatric Cataract Surgery

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Improvements in surgical techniques and technologies have helped us to provide better surgical outcomes for cataract surgery in children. However, visual axis obscuration (VAO) remains one of the major hurdles that impede successful visual rehabilitation following pediatric cataract surgery. Posterior capsulorhexis and anterior vitrectomy are considered standard surgical steps for infants and young children undergoing cataract surgery to prevent VAO. In 1994, Gimbel and DeBroff described a posterior “optic capture” technique for pediatric cataract surgery that they proposed would maintain a clear visual axis and eliminate the need for vitrectomy.¹

The concept of capturing an intraocular lens (IOL) optic through an anterior capsulorhexis was first suggested in a case of posterior capsule tear in an adult eye. The IOL haptics were placed in the ciliary sulcus, and the IOL optic was placed through the anterior capsulorhexis to “capture” the optic and ensure stable optic fixation. The concept was subsequently described as “optic capture,”^{1–15} “optic entrapment,”¹⁵ or “optic buttonholing.”^{16–23}

Optic capture fuses the anterior and posterior leaflets of the capsular bag almost completely in a 360-degree position, except at the haptic–optic junction. Theoretically, capsular fusion anterior to the IOL optic reduces central lens epithelial cell migration or at least directs the cell movement anteriorly over the lens optic, which is presumably an unsuitable substrate for lens survival. Thus, capsular fusion may help in decreasing VAO. Another advantage of performing optic capture is its ability to achieve a well-centered IOL.^{1,7}

There are several possible sites for optic capture (Fig. 26.1). *Conventional (or posterior) optic capture* techniques utilize the *posterior capture* technique. Here, the optic is captured through the anterior capsulorhexis (haptics in the ciliary sulcus, optic in the bag), through the posterior capsulorhexis (haptics in the bag, optic in the

vitreous), or through both the rhexes (haptics in the ciliary sulcus, optic in the vitreous). Optic capture can be helpful for both primary and secondary IOL implantation.^{7,24}

CAPTURE THROUGH POSTERIOR CAPSULORHEXIS (HAPTICS IN THE BAG, OPTICS BEHIND THE POSTERIOR CAPSULE)

Before moving forward, it is worth noting that IOL power may need to be adjusted 0.5 to 1 diopter higher than for bag-fixed IOLs. The success of optic capture technique may depend on the IOL haptic–optic junction design.¹³ For example, a haptic with a right angle at the haptic–optic junction is preferred for optic capture as it helps in achieving complete capsule closure.

Technique

Following manual anterior continuous curvilinear capsulorhexis and irrigation/aspiration of lens material, a continuous manual posterior continuous curvilinear capsulorhexis (PCCC) is performed (Fig. 26.2). Under a high-viscosity ophthalmic viscosurgical device (OVD), the IOL is implanted in the capsular bag. Thereafter, the IOL optic is gently pressed backward and is captured through the posterior capsulorhexis using a spatula/Leister hook (Fig. 26.3). The haptics are placed in the capsular bag fornix. If the round opening is stretched into an elliptical one, it indicates complete capture (Fig. 26.4).

Optic capture is a technically challenging procedure. The key point is to achieve a “capturable” PCCC. Although there are several options for performing PCCC, a manual PCCC is a prerequisite for optic capture of the IOL through the PCCC. The opening in the posterior capsule should be not only continuous

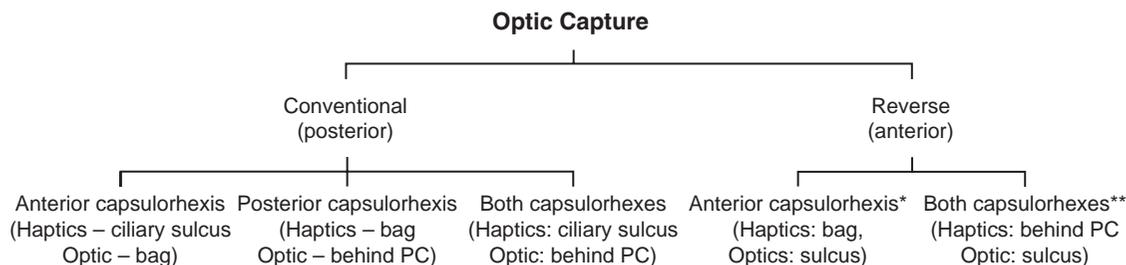


Figure 26.1. Schematic diagram showing different sites for capturing the IOL optic. *In the case of a large posterior capsule tear noticed after IOL implantation and in-the-bag fixation; not considered stable. **In the case of posterior dislocation (pars plana repositioning of an IOL).

but also well centered and of an optimum size. If the opening is too small, it is difficult to capture the optic, and there is the possibility of tearing the posterior capsule due to excessive stress to the PCCC edge. On the other hand, if it is too large, the optic may not remain captured. Thus, creating an opening of an optimum size is a prerequisite for successful optic capture. The diameter of the PCCC opening should be approximately 1.0 mm smaller than the IOL optic. The principle behind the use of this technique is to avoid the need for vitrectomy. However, occasionally, the vitreous face is disturbed while performing primary PCCC or capturing the IOL optic, and an unplanned vitrectomy may be required.

Removal of the OVD may sometimes cause the optic to vault forward. This vaulting can undo the optic capture.

IOL CAPTURE THROUGH BOTH CAPSULORHEXIS (HAPTICS IN THE CILIARY SULCUS, OPTIC BEHIND THE POSTERIOR CAPSULE)

The surgeon may consider this technique as an alternative to sulcus IOL placement in those cases in which the anterior capsulorhexis has more than one large

tear to the equator of the capsule that compromises the secure placement of the haptics in the capsular bag. In addition, an intact PCCC must be present. In such a situation, the surgeon may position the IOL in the ciliary sulcus and capture the optic posteriorly through the anterior and posterior capsulorhexis openings. This approach maintains IOL centration and prevents Elschnig pearl formation posterior to the IOL. IOL decentration is the most common complication in eyes with secondary IOL implantation. Optic capture through fused anterior and posterior capsulorhexis (haptics in the ciliary sulcus) may help to avoid IOL decentration postoperatively.²⁴

REVERSE OPTIC CAPTURE (HAPTICS IN THE BAG, OPTICS IN THE SULCUS)

Bringing the optic of the IOL anteriorly through the anterior capsulorhexis opening effectively achieves a reverse capsulorhexis fixation. This technique is useful when a radial tear of the PCCC occurs that renders the IOL unstable within the capsular bag. The surgeon may then bring the optic of the IOL forward and capture it through the anterior continuous curvilinear capsulorhexis, thus creating IOL stability and ensuring lens

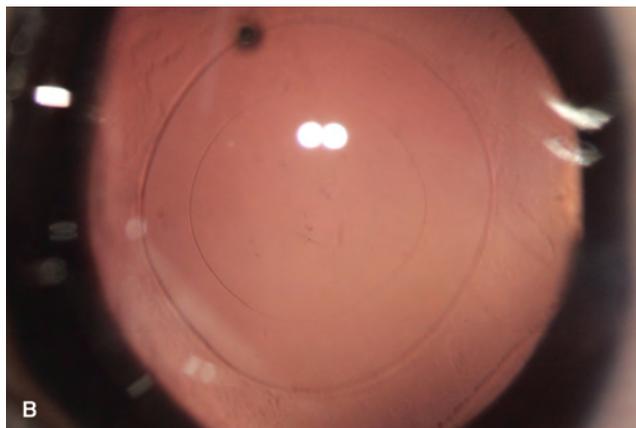
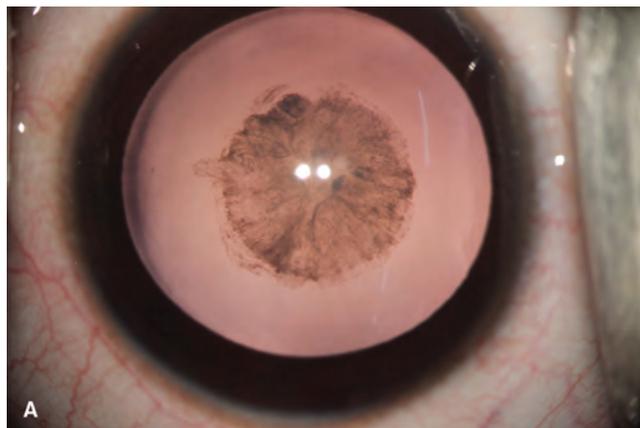


Figure 26.2. **A:** Lamellar cataract. **B:** Optimum-sized anterior continuous curvilinear capsulorhexis and PCCC.

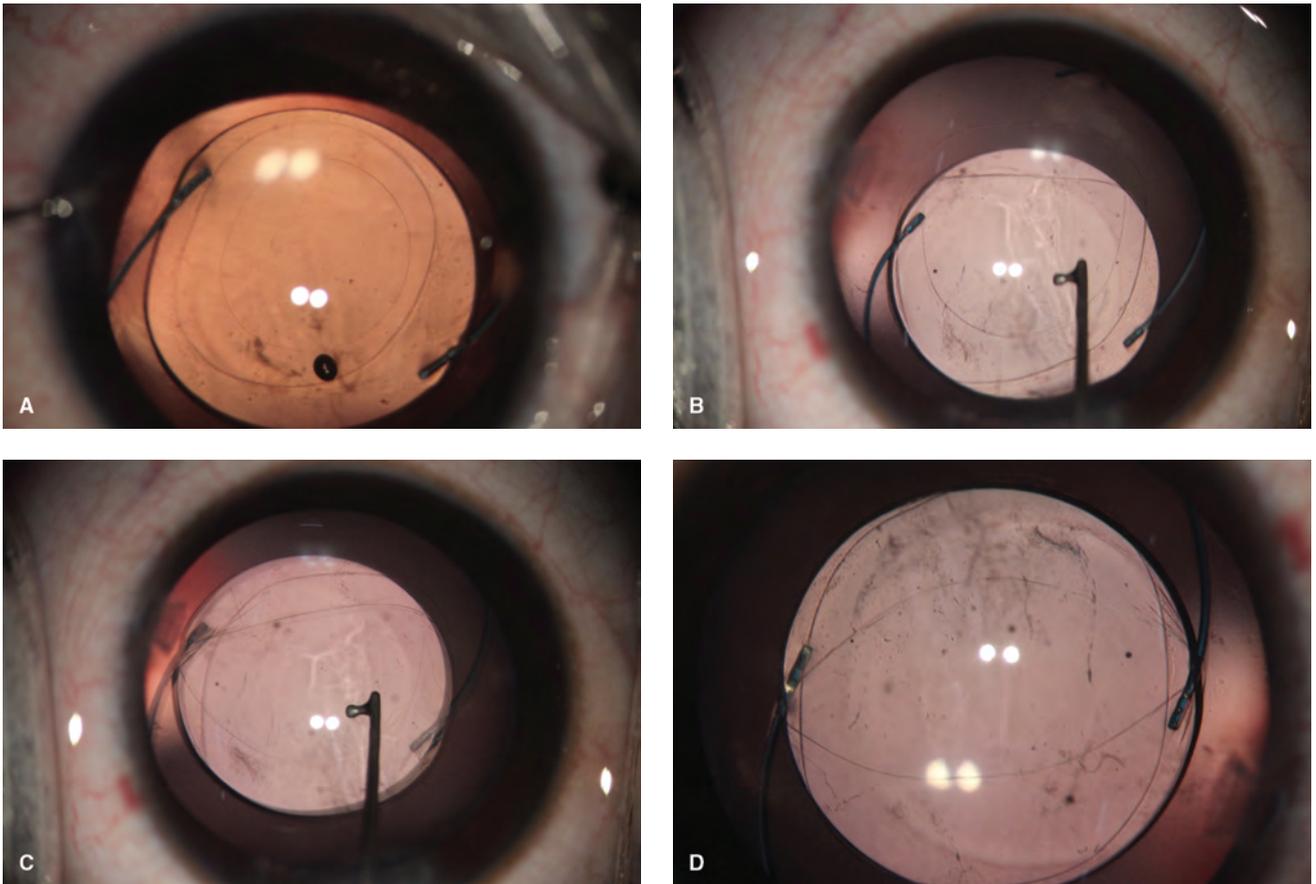


Figure 26.3. **A:** A three-piece hydrophobic acrylic IOL placed in the capsular bag. **B and C:** Using Leister hook, the IOL is pressed backward and captured through the PCCC. **D:** Optic capture of IOL through the PCCC.

centration. This technique is reported to be useful to resolve pseudophakic negative dysphotopsia in the adult population.²⁵ This technique can also be useful for piggyback IOL implantation. In this technique, haptics of both piggyback IOLs remain in the bag. The anterior IOL

optic is pulled out of the bag for reverse optic capture (the posterior IOL optic remains in the bag). This approach may reduce the accumulation of pearls in the interface between the two IOLs (interlenticular opacification), while maintaining bag fixation of the second IOL.

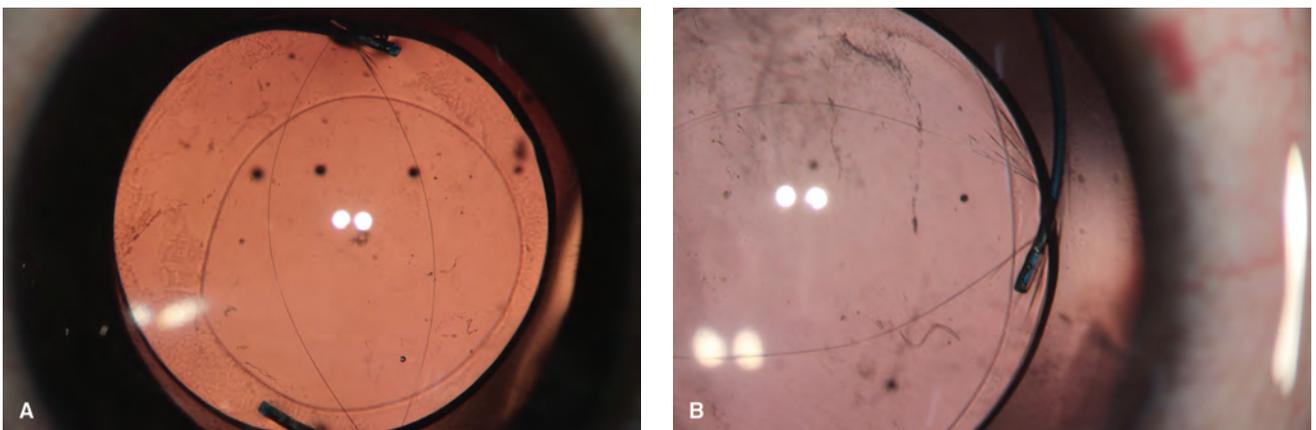


Figure 26.4. **A:** Successful optic capture of IOL indicated by elliptical shape of the PCCC. **B:** Striations at the optic-haptic junction.

PERIOPERATIVE OUTCOMES OF OPTIC CAPTURE OF THE IOL

Spontaneous Release of Optic Capture

Spontaneous release of the optic capture through the posterior capsulorhexis has been reported.⁷ The optic did not remain captured in 4.9% of the eyes in older children and in 30.0% of the eyes in younger children.⁷ This phenomenon is observed more frequently in eyes with a large posterior capsulorhexis.⁷

Visual Axis Obscuration

The role of optic capture in preventing VAO in the absence of vitrectomy is controversial. Combining primary posterior capsulorhexis with primary optic buttonholing has proven to be successful in several clinical series in adult cataract surgery.^{16,17} In a study of children younger than 5 years, it is reported that optic capture without anterior vitrectomy did not always ensure a clear visual axis.⁷ In another prospective, randomized, controlled study, the authors concluded that anterior vitrectomy is a necessary step while performing optic capture in children with congenital cataract.⁹ In this study of children over 5 years, even when optic capture was performed, the authors found a significant difference in visual axis clarity between eyes that had undergone vitrectomy and those that had not.⁹ Eyes with VAO had reticular fibrosis of the anterior vitreous face in the first 2 months after surgery. Hence, vitreous opacification

could be a primary response of the anterior vitreous face when it occurs with the IOL optic rather than a secondary scaffold response caused by proliferating lens epithelial cells, inflammatory cells, and deposits. Data summarizing the incidence of VAO in the presence of optic capture with and without vitrectomy are presented in Tables 26.1 and 26.2.^{2,4,5,7-10,12,14,15}

Inflammation

The incidence of posterior synechia and cell deposits on the IOL was reported to be significantly higher in the optic-capture group compared to the no-capture group in a pediatric study.⁷ An adult study reported that cataract surgery with combined primary PCCC and optic buttonholing led to significantly lower postoperative anterior chamber reaction than conventional in-the-bag fixation (also with primary PCCC).²⁰

Centration of an IOL

In our opinion, the most important benefit of optic capture is that the continuous capsular margin locks the IOL optic and prevents it from decentering.⁷

Intraocular Pressure Spike

A prospective, randomized adult study has reported that postoperative intraocular pressure (IOP) spikes (during first 24 hours) after cataract surgery and PCCC can be effectively prevented by buttonholing the IOL through the PCCC.¹⁹

Table 26.1 PREVALENCE OF VAO AFTER OPTIC CAPTURE WITH PCCC WITHOUT VITRECTOMY

Author	Year of Publication	Age (Months)	Total (Follow-up) (Months)	Optic Capture (N = Eyes)	VAO	At Which Follow-up (Months)	Type
Gimbel ¹	1996	69.6 ^b	19 ^b	13	0		
Koch and Kohnen ^{4,5}	1997	18–144 ^a	24 ^b	5	80%	6 ^b	Not given
Vasavada and Trivedi ⁷	2000	26.08 ^b	16.53 ^b	14	0%		Membrane in front of IOL-1 eye, which required secondary procedure
Argento et al. ⁸	2001	24–96 ^a	28.9 ± 5.3 ^c	8	0%		
Vasavada et al. ⁹	2001	83.57 ^b	21.04 ^b	20	70%		Anterior vitreous fibrosis
Raina et al. ¹⁰	2002	18–144 ^a	17.5 ^b	16	0%		
Mullner-Eidenbock et al. ¹²	2003	72–180 ^a	20.73 ^b	7	0%		
Grieshaber et al. ²⁶	2009	49 ^b	48.02 ^b	47	0%		

^aRange.

^bMean.

^cMean and SD.

VAO, visual axis obscuration.

Table 26.2 PREVALENCE OF VAO AFTER OPTIC CAPTURE WITH PCCC AND VITRECTOMY

Author ^{reference}	Year of Publication	Age (Months)	Total Follow-up (Months)	Optic Capture ± Vitrectomy N = Eyes	VAO
Retrospective Study					
Koch and Kohnen ^{4,5}	1997	18–144 ^a	24 ^b	3	0%
Prospective Studies					
Vasavada et al. ⁹	2001	83.57 ^b	21.04 ^b	21	0%
Mullner-Eidenbock et al. ¹²	2003	24–69 ^a	20.73 ^b	8	0%
Raina et al. ¹⁴	2004	78 ^b	13 ^b	6	0%
^a Range					
^b Mean					
VAO, visual axis opacification					

SUMMARY

The role of optic capture in preventing VAO in the absence of vitrectomy is controversial. However, optic capture helps in achieving a well-centered IOL.

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Bag-in-the-Lens

Marie-José Tassignon

Modern-day cataract surgeons have struggled to address the most common postoperative complication, posterior capsule opacification (PCO). In cases where the posterior capsule has been kept intact, the term PCO is used. However, in cases where a primary posterior continuous curvilinear capsulorhexis (PPCCC) has been performed (which is considered standard in cataract surgery of babies and infants), the term visual axis opacification (VAO) or visual axis re proliferation (VAR) is used. Even after PPCCC, the incidence of VAO has been reported ranging from 8% to 80%.^{1,2} The highest incidences occur when the child presents with associated ocular anomalies and is operated at <6 months of age. VAO occurs very quickly postoperatively, within the first 6 months, however the incidence of VAO does not increase very much after the first postoperative year.

Traditionally, the intraocular lens (IOL) is placed within the capsular bag. This approach can be thought of as a lens-in-the-bag (LIB) implantation. A new concept of lens implantation was introduced in the early 2000s, called the bag-in-the-lens (BIL) implantation technique (Fig. 27.1A; Table 27.1). The BIL technique has been routinely implanted by the author in adults since 2004. The BIL technique requires a slightly adapted surgical approach. Compared to the traditional LIB technique, all surgical steps in the BIL procedure need more precision, particularly sizing of the anterior and posterior capsulorhexis. This requirement for precision is the reason that from its conception, some surgeons have felt that it was too difficult in theory to perform. Now that results of BIL have been published, this stigma is decreasing. The outcome results of BIL show it to be superior to other techniques in preventing VAO, which are as low as 0% in adult eyes^{3,4} provided the lens could be implanted in the appropriate way. Pediatric ophthalmologists were less resistant to the BIL technique because they commonly perform PPCCC, a procedure that is done routinely by only a few high-volume adult cataract surgeons worldwide.

The BIL involves the use of a twin capsulorhexis IOL design (see Fig. 27.1B and C) requiring anterior and posterior capsulorhexes that are matched in size. The term *matching* is used rather than *congruent* on purpose, since it is more important to have similar capsulorhexis sizes than shapes. The idea behind the BIL technique is that if both capsules are stretched around the 5-mm central optic diameter as currently proposed (though larger diameters are available if needed), the lens epithelial cells (LECs) will be captured within the remaining peripheral space of the capsular bag, and their proliferation will be limited to this space, so that the visual axis will remain clear.⁵⁻⁹

The BIL is manufactured by the Morcher, Germany, under references 89 A/D/F (see Table 27.1). It is made of hydrophilic Coacryl biomaterial with 28% water content. The 89A reference is the standard type of BIL with an overall length of 7.5 mm and a biconvex optic of 5.0 mm. The company proposes to use the A-constant of 118.2. Based on our clinical experience, I prefer to use 118.0 for the A-constant. The 89D type of BIL is mainly used for very small eyes. It has the same design but with 4.5 mm optic. The 89F type is an adaptation of the BIL at the request of Claus Eckhart from Germany, who desired a larger anterior haptic with an overall diameter of 8.5 mm to be used in his combined cataract-vitreotomy surgeries with gas or silicone tamponade. This larger haptic design precludes the incidence of iris capture postoperatively.

Since the anterior capsule in children is highly elastic, an anterior continuous curvilinear capsulorhexis (ACCC) is more challenging to perform even more so in babies. Different solutions have been proposed in the literature to increase the performance of this surgical step.¹⁰⁻¹² Our approach was to design a ring caliper made of polymethyl methacrylate, which can be cut in a ring of 0.2 mm of cross-sectional thickness and variable inner overall diameter matching the size of the desired capsulorhexis. This ring caliper presents properties of flexibility and memory that are ideal for the intended purpose

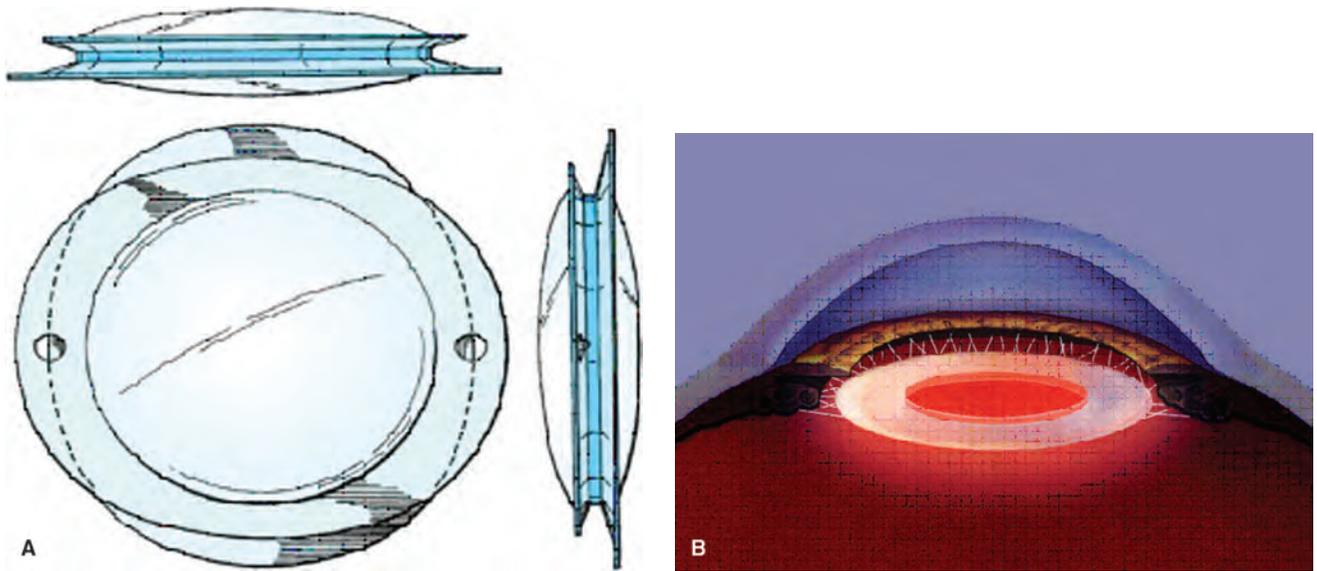


Figure 27.1. Bag-in-the-lens. **A:** Front and side view of the technical drawing of the BIL. **B:** Schematic drawing of the position of the capsular bag after having done a sized and centered anterior and posterior capsulorhexis. **C:** Schematic drawing of the BIL in the proper position within the anterior chamber.

(Table 27.2; Fig. 27.2). The caliper ring was introduced in 2006.¹³ The device can be inserted into the anterior chamber through a 1.2-mm corneoscleral opening by means of a caliper ring positioner (see Table 27.2) and is stabilized on top of the anterior capsule by means of a 1% sodium hyaluronate solution with high viscosity. This ophthalmic viscosurgical device (OVD) has the

advantage of flattening the anterior capsule, making it easier for the surgeon to perform an ACCC in a reproducible and controlled fashion. I personally use Healon® GV for this purpose (Advanced Medical Optics [AMO], Santa Ana, CA). Capsular staining by means of trypan blue 0.1% (VisionBlue, Dutch Ophthalmic Research Center)¹⁴ is in our hands only used for treating white cataracts or congenital anterior capsule disorders. In the latter cases, the staining will enable careful dissection of the embryologic remnants of the anterior part of the tunica vasculosa from the underlying anterior capsule (Fig. 27.3).

In children, a caliper ring of 4.5-mm internal diameter (4 L Tassignon ring caliper, Morcher, Germany) is preferred instead of a 5-mm internal caliper (5 Tassignon caliper ring, Morcher, Germany) as used in adults (see Table 27.2). By using slightly smaller caliper rings in babies and children, the surgeon includes a safety margin of 0.5 mm for the ACCC to compensate for the highly elastic properties of the capsular bag. Before starting the puncture of the anterior capsule, the ring

Table 27.1 TECHNICAL PROPERTIES OF THE BIL

Manufacturer	Morcher
Design	89 A/D/F
Material	Hydrophilic Coacryl 28
Overall length	A: 7.5; D: 6.5; F: 8.5 mm
Optic	Biconvex. Size: A, F 5.0; D: 4.5 mm
Power range	Standard 10–30 D/other on request (up to 39 D)
Estimated A constant	118.2 (My personal A-constant is 118.0.)

Table 27.2 INSTRUMENTATION LIST

No	Description	Comments	Type No.	Manufacturer
1	"Bag-in-the-lens" foldable IOL	28% hydrophilic acrylic	89A-F	Morcher
2	Ring caliper (4.5–5)	To caliper the size and position of the anterior capsulorhexis	Type 4 L/type 5	Morcher
3	Tassignon caliper ring positioner	To position the ring caliper	sh-7017	EyeTech
4	Ikeda angled 30-degree capsulorhexis 23 g forceps	To perform anterior and posterior capsulorhexis	Fr 2268	EyeTech
5	Straight scissors in curved shaft	To adjust the capsulorhexis if needed	Fr 2295c	EyeTech
6	Naviject injector atraumatic/ Naviglide			Medical
	Cartridge 2.5-IP injector set foldable	Up to +20.0 diopters	Lp 604420	
	Cartridge 2.8-IP injector set foldable	For all diopters	Lp 604410	
7	Rycroft/Helsinki hydrodissection needle 27 gauge	To inject dispersive viscoelastic behind the posterior capsule	1273E	Steriseal
8	41-gauge needle (same type from two different manufacturers)	Idem then 7 but to be used in babies and children	E7370 1270.0.100	Bausch & Lomb Dorc

caliper is centered on top of the anterior capsule using the first and fourth Purkinje reflex images (Fig. 27.4) of the light of our microscope (we use the Zeiss Microscope S8, Germany) present in the anterior segment of the eye (Purkinje I reflex corresponds to the anterior face of the cornea, and Purkinje IV reflex comes from the back surface of the lens). The pupillary center can also be used for centering when the iris is optimally dilated and regular in shape, though this is not always the case, especially not in eyes with associated

anomalies. The anterior capsule puncture is performed preferably with a 30-gauge needle in very young children. In children older than 3 years, the anterior capsule can be punctured by means of the angulated tip of a microforceps. The microforceps I preferably use are the Ikeda capsulorhexis microforceps with 30-degree angulation of the grasping tips (FR-2268, EyeTech, UK) (see Table 27.2). The anterior capsulorhexis is performed by frequent grasping and regrasping and by following the internal border of the caliper ring as a guide

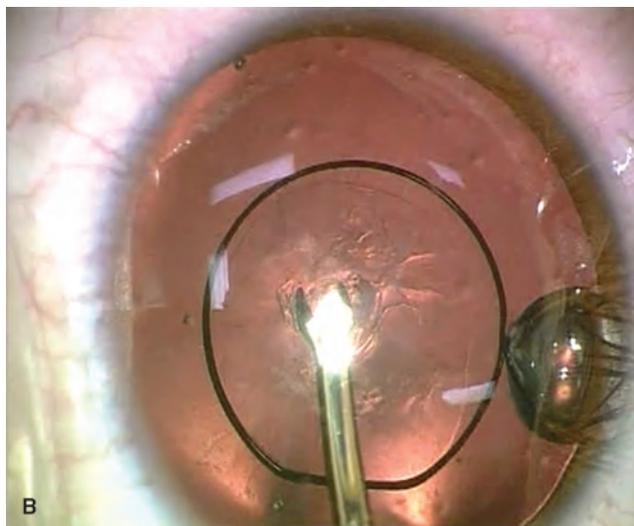
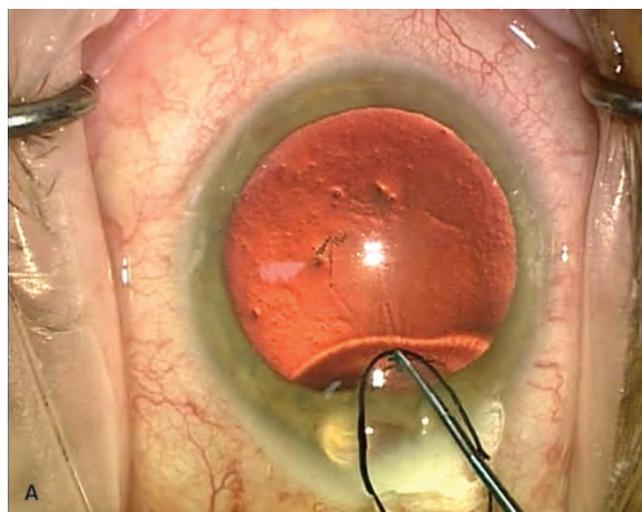


Figure 27.2. Ring caliper. **A:** The ring caliper is introduced into the anterior chamber. **B:** The ring caliper is positioned on top of the anterior lens capsule. The anterior capsulorhexis is completed.



Figure 27.3. Example of white cataract needing the use of vision blue to perform an accurate anterior capsulorhexis.

(see Fig. 27.2B). This maneuver may take some minutes because this step is one of the most important of the BIL technique.

The next step is to clean the lens content from the capsular bag. Cataracts in children and babies are typically very soft and as a result very easy to take out by simple irrigation–aspiration, provided that the ideal balance between irrigation and aspiration forces is respected. Hydrodissection, though highly recommended in adults, is less reliable in children and should be tried, though without persisting.

The next step consists of creating a PPCCC of similar size than the ACCC.

Remember that the posterior capsule is one-third in thickness of the anterior capsule and less elastic

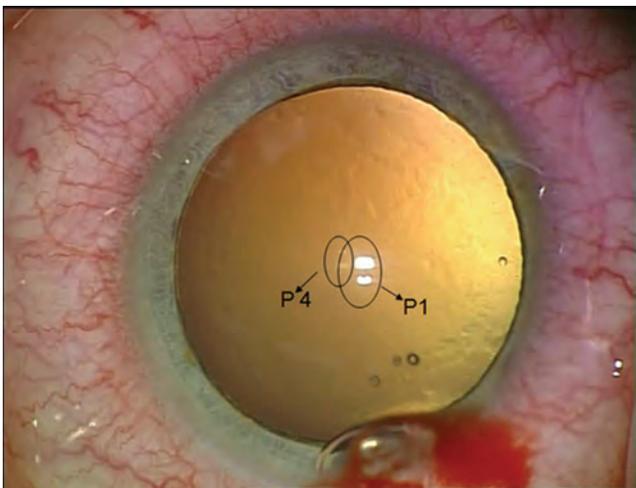


Figure 27.4. Surgical view of the P1 (Purkinje 1) and P4 (Purkinje 4) reflexes on the light sources of the microscope on the cornea (P1) and posterior face of the crystalline lens (P4).

than the anterior capsule, in children. The same capsulorhexis microforceps as used for the anterior capsulorhexis are used to perform the PPCCC. It is important to position the posterior capsule in the most ideal position to facilitate the surgical act of tearing the posterior capsule in a controlled manner. Because the anterior capsulorhexis will serve as a guide for the posterior capsulorhexis, it is important that the anterior capsulorhexis is very close to the posterior capsule. To reach this goal, heavy chain OVD is injected on top of the ACCC, which will then be pushed back close to the posterior capsule until the anterior and posterior capsule complex becomes closely positioned within the anterior chamber (Fig. 27.5). The posterior capsule puncture can be performed by means of a 30-gauge needle.

Separating the posterior capsule from the anterior hyaloid is the next step. This is performed by introducing the OVD needle into the posterior capsule puncture and injecting the OVD until a very well-defined OVD blister is formed, slightly larger in size than the ACCC. The contour of the blister will be defined by the ligament of Wiegert, which corresponds to the attachment of the anterior hyaloid to the posterior capsule. The blister shape is most often not regular which explains the bridges present between the posterior capsule and the anterior hyaloid. Those bridges are disrupted by injecting OVD into the Berger space. A low-viscosity sodium hyaluronate 1% is used for the injection behind the posterior capsule. My preference goes to regular Healon (AMO, USA). In order to ensure an appropriate positioning of the BIL and more precisely of the posterior haptic in the Berger space, it is important to inject a little more of the regular OVD behind the posterior capsule, using a curved needle allowing an accurate separation of the anterior hyaloid from the posterior capsule over 360 degrees. Omitting doing this maneuver may jeopardize the clinical outcome by increasing the risk of VAO due to retraction of the posterior capsule, out of the lens groove, and then allowing the growth of the LECs toward the visual axis along the anterior hyaloid. This complication was found in three eyes (Fig. 27.6A–C) treated with the BIL technique following an otherwise uneventful surgical procedure. Since then, I control the presence of an accurate separation between the posterior capsule and the anterior hyaloid systematically.

The BIL surgical technique described above should be altered somewhat in infants and eyes with associated ocular anomalies. In babies, the initiation of the dissection of the anterior vitreous face from the posterior capsule is done by means of a 41-gauge needle borrowed from the vitreoretinal surgeons (Bausch & Lomb E7370, Dorc 1270.0.100) (see Table 27.2). The blister formation can be accomplished in a more controlled way once

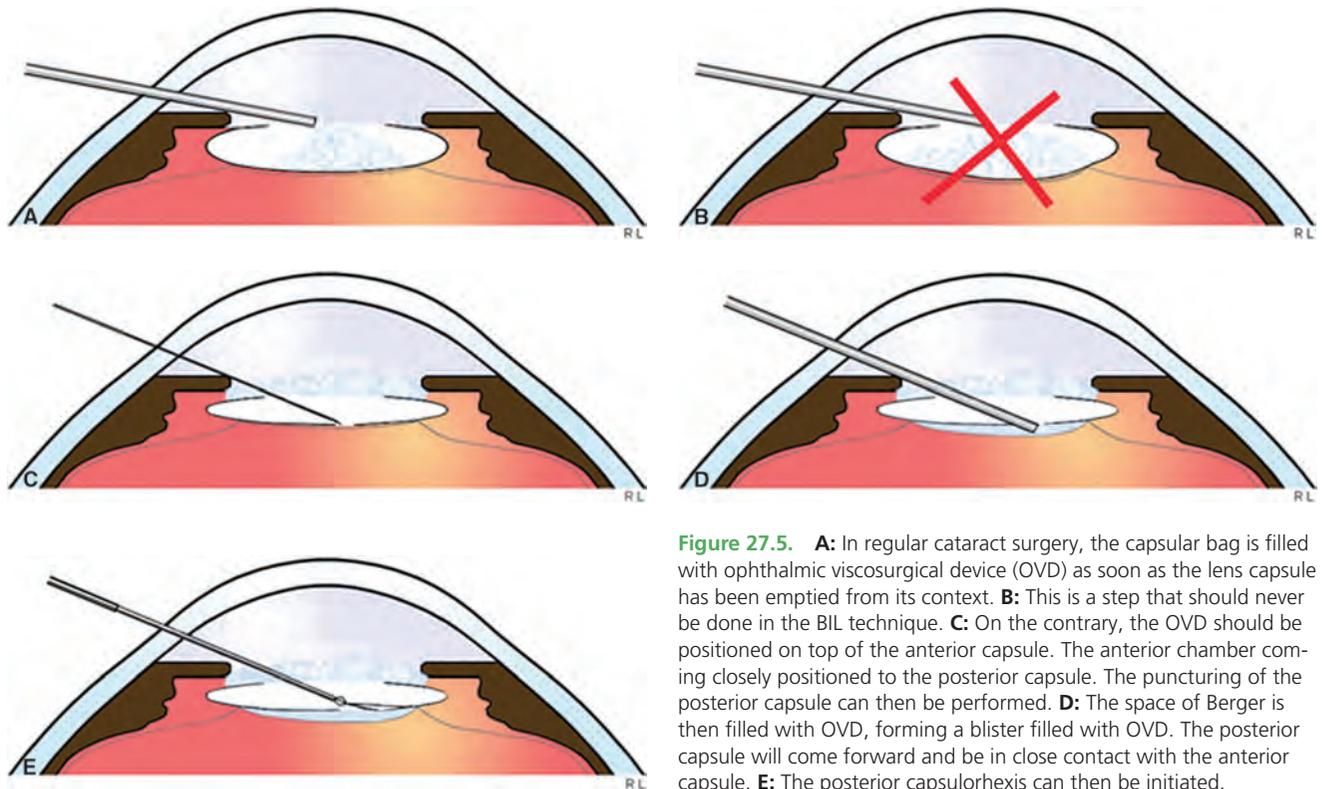


Figure 27.5. **A:** In regular cataract surgery, the capsular bag is filled with ophthalmic viscosurgical device (OVD) as soon as the lens capsule has been emptied from its context. **B:** This is a step that should never be done in the BIL technique. **C:** On the contrary, the OVD should be positioned on top of the anterior capsule. The anterior chamber coming closely positioned to the posterior capsule. The puncturing of the posterior capsule can then be performed. **D:** The space of Berger is then filled with OVD, forming a blister filled with OVD. The posterior capsule will come forward and be in close contact with the anterior capsule. **E:** The posterior capsulorhexis can then be initiated.

the separation of the anterior hyaloid has been initiated. The residual blister can be enlarged using the regular OVD needle. Because the 41-gauge fine needle is mounted on a solid metallic needle, the corneoscleral incision needs to be enlarged at this stage. This enlargement is better done right away at 2.8 mm, which is the size needed to inject the high dioptric power BIL by means of an appropriate injector (powers up to 39 D are available on demand).

At this stage, the BIL can be implanted by injecting it into the anterior chamber, using an adequate cartridge (Medicel Lp 604420, Lp 604410) (see Table 27.2 and Fig. 27.7). A preloaded injection system is currently in development but not yet commercially available. Once the lens is unfolded in the anterior chamber, it is pushed back in close contact with and in front of the ACCC where the optical part takes its final position, taking care that the largest diameter of the posterior haptic part is located at the surgical 3 o'clock to 9 o'clock position. This lens positioning in the anterior chamber of the eye is done by using regular OVD. The BIL is then glided slightly sideways to the surgical 9 o'clock position so that the posterior haptic can be glided behind the posterior capsule at the 3 o'clock position. By pushing softly on the lens optic, the capsules are progressively engaged in the lens groove. Once approximately 180 degrees of

the capsule's circumference is correctly positioned in the lens groove, the OVD needle is positioned between both haptics at the 9 o'clock position and the lens is pushed slightly backward allowing the remainder of the capsule rim to glide properly and completely into the lens groove. This ends the insertion maneuver. The BIL can also be inserted with the longest axis of the posterior haptic in the surgical 6 o'clock to 12 o'clock position. In this case, the insertion of the BIL will be done starting by gliding the posterior haptic behind the posterior capsule at the surgeon's 6 o'clock position and ending the insertion at the surgeon's 12 o'clock position.

The iris is constricted after injection of a solution of Miochol, diluted five times with balanced salt solution (BSS), and the corneoscleral wounds are sutured. A cefuroxime solution is then injected in the anterior chamber and corneal tissue (Table 27.3).

All babies and children operated in our department for cataract since 2006 have been treated using the BIL technique.^{15,16} Thirty-four eyes of 22 children had implantation of BIL (age at surgery: 2 months to 14 years). Postoperative follow-up was 17.45 ± 17.12 (4–68) months. In three eyes, the IOL could not be properly implanted. In these eyes, only the anterior capsule was inserted in the IOL groove. Secondary intervention was necessary in those cases to treat early PCO.

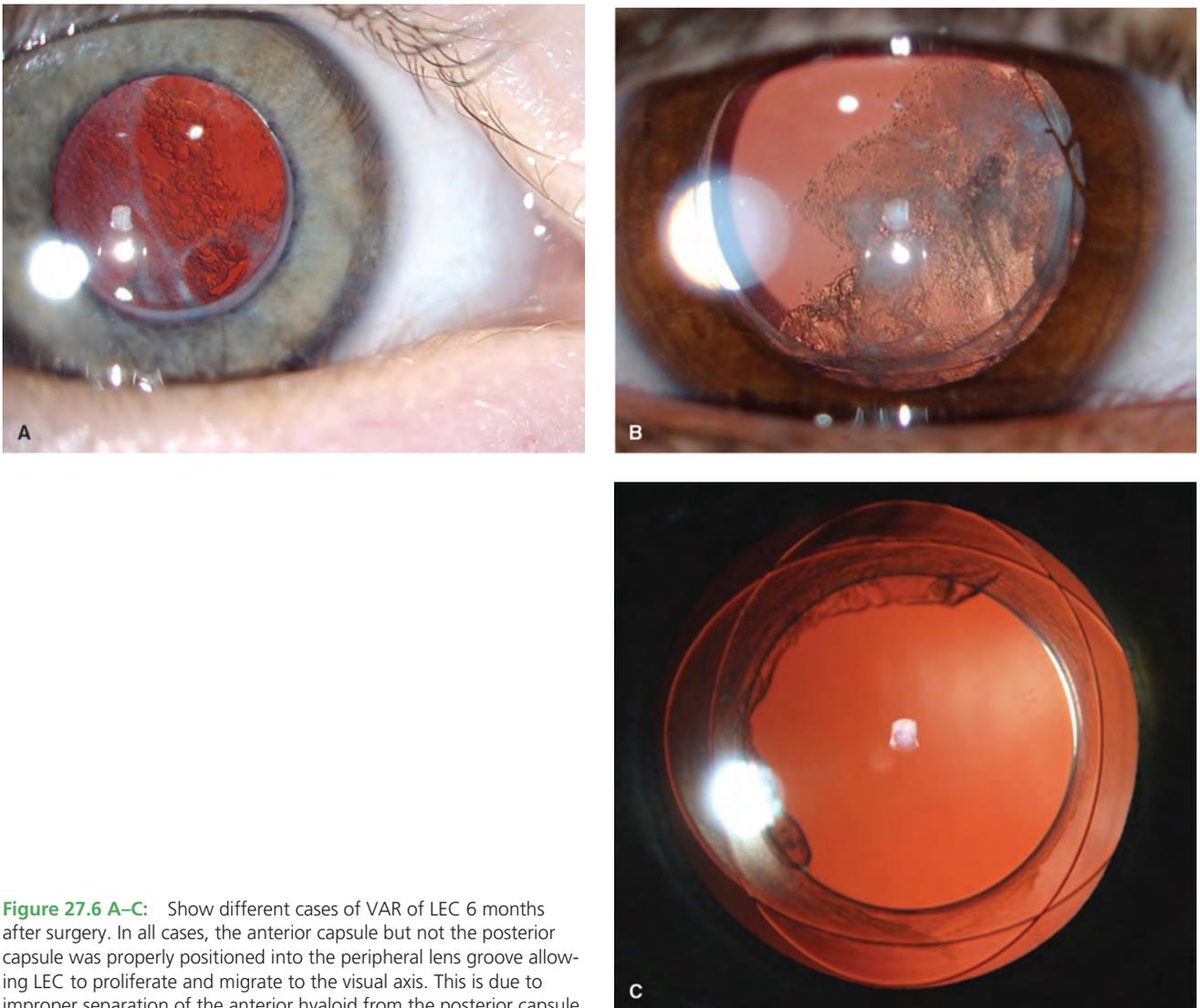


Figure 27.6 A-C: Show different cases of VAR of LEC 6 months after surgery. In all cases, the anterior capsule but not the posterior capsule was properly positioned into the peripheral lens groove allowing LEC to proliferate and migrate to the visual axis. This is due to improper separation of the anterior hyaloid from the posterior capsule.



Figure 27.7. The BIL is positioned in the cartridge of the injector (see Table 27.2).

Table 27.3 PROCEDURE CEFUROXIME (ZINACEF) SOLUTION

- Take 2.5 mL NaCl in the 10-mL syringe.
 - Inject these 2.5 mL NaCl into the bottle filled with cefuroxime 250-mg powder.
 - Shake thoroughly until the cefuroxime powder is properly diluted.
 - Using the 10-mL syringe, take 1 mL out of this solution.
 - Fill the additional 9 mL of the syringe with NaCl.
- NaCl, sodium chloride.

The optical axis remained clear during the follow-up period in all eyes that had successful IOL implantation. The results motivated us to adjust the calculation method of the BIL power and adapting our strategy for visual rehabilitation.

Because of our increasing experience in dissecting and freeing the anterior interface, we are interested in classifying the anatomical variations and analyzing histologically the dissected structures. This is an ongoing study and the topic of a departmental PhD thesis project. This topic is not further developed since it is outside the scope of this chapter.

In developmental cataract, there is more at issue than simple opacification of the lens only. The anterior vitreolenticular interface or space plays an important role in the surgical approach, which is especially so in the BIL technique. As explained earlier, it is in this space that the posterior haptic of the BIL will be positioned. It is therefore important to define this space very accurately during surgery. Not taking the anatomical variations of the vitreolenticular space into consideration will increase the complication rate of VAO dramatically

as shown in Figure 27.6. The posterior capsule is in close anatomical contact with the anterior vitreous hyaloid, and this explains why the posterior capsule can be retracted out of the lens groove in case it has not been freed from the vitreous body. We currently lack medical devices allowing us to visualize the anterior interface prior to surgery. The preoperative clinical appearance of a congenital cataract does not disclose all anomalies we may encounter preoperatively at the level of the anterior interface. The anterior vitreolenticular interface is not well understood, except for the fact that there is a large anatomical variety in what is commonly called the space of Berger. In an attempt to reduce the need of anterior vitrectomy, our practical approach is to try to dissect manually all embryologic remnants present in the pupillary area or they may interfere with the transparency of the visual axis (Fig. 27.8).

The toric BIL has been developed recently and implanted in a few eyes of children.¹⁷ The technical properties of this one-sided intraocular toric IOL have been published in detail, including the calculation methodology used.¹⁸ The particularity of our calculation approach is that the IOL calculation formula SRKT has been applied for each meridian of K1 and K2 instead of the mean of K1 and K2 as done in most toric lens calculations. We presume that this approach more accurately reflects the refraction of the incoming light at both meridians.

Because some eyes present with very unstable capsule support, for example, in Marfan syndrome or in case of capsule contraction after primary cataract surgery as shown in Figure 27.9A, we developed in collaboration with the Morcher (Germany) new auxiliary haptic devices that fit the lens groove of the BIL and can be positioned in the ciliary sulcus (Fig. 27.10). Because of their particular shape, these devices are called bean haptics.

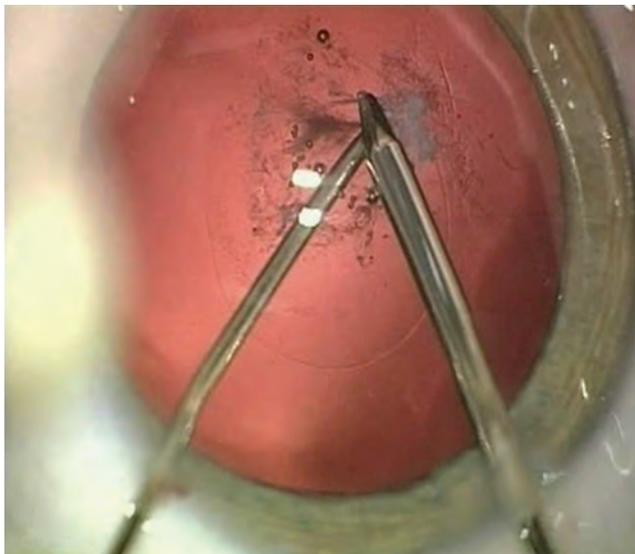


Figure 27.8. Bimanual dissection of the posterior hyaloid and remnants of incomplete regression of the embryonal tunica vasculosa.

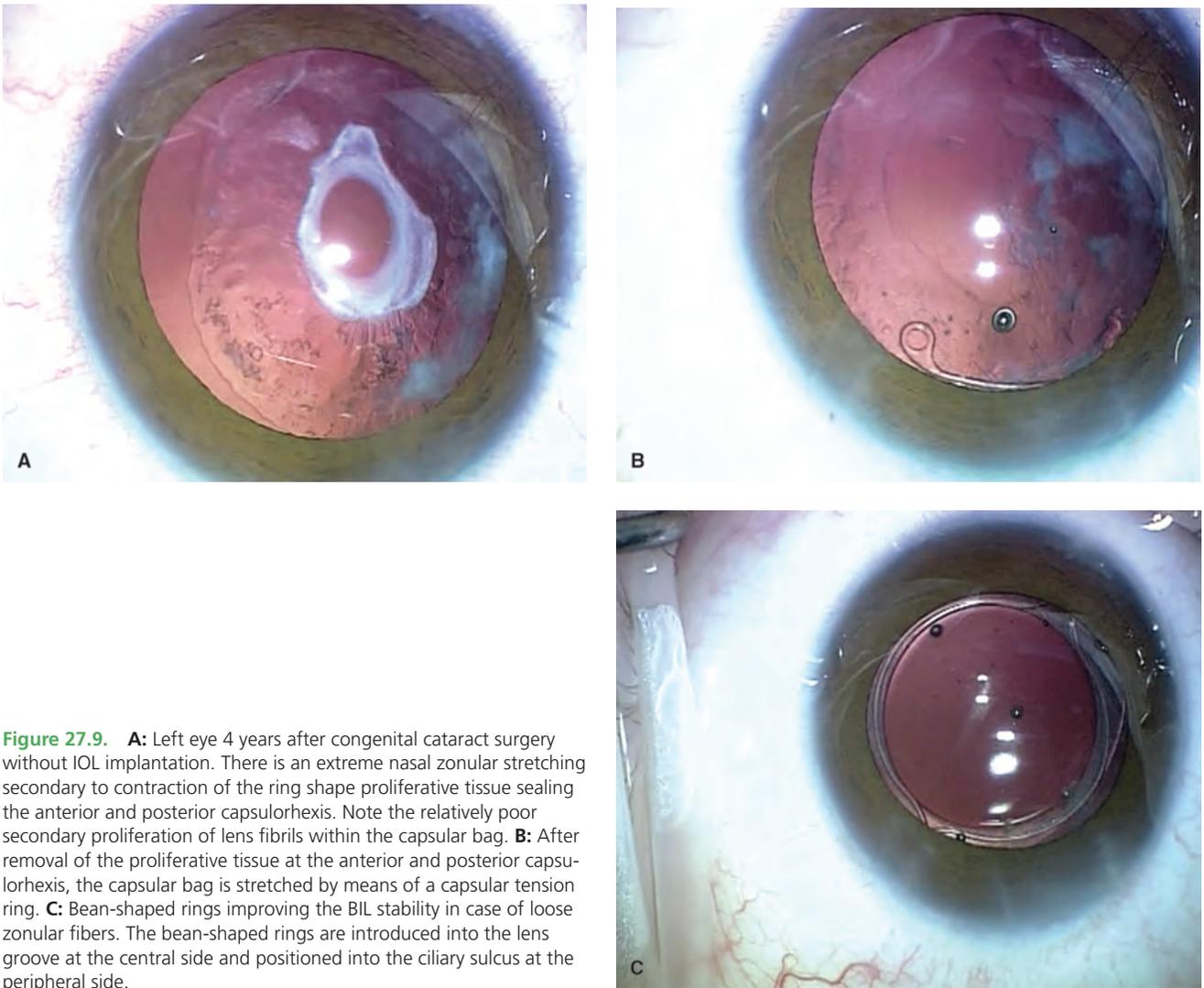


Figure 27.9. **A:** Left eye 4 years after congenital cataract surgery without IOL implantation. There is an extreme nasal zonular stretching secondary to contraction of the ring shape proliferative tissue sealing the anterior and posterior capsulorhexis. Note the relatively poor secondary proliferation of lens fibrils within the capsular bag. **B:** After removal of the proliferative tissue at the anterior and posterior capsulorhexis, the capsular bag is stretched by means of a capsular tension ring. **C:** Bean-shaped rings improving the BIL stability in case of loose zonular fibers. The bean-shaped rings are introduced into the lens groove at the central side and positioned into the ciliary sulcus at the peripheral side.

Their internal diameter fits the external diameter of the lens optic and will typically have 5-mm curvature at the lens side. At the ciliary sulcus side, the diameter will vary according to the white-to-white diameter as measured with the Lenstar (Haag-Streit, Switzerland) or any other device capable of measuring this parameter. As a consequence, this external diameter of the additional bean-shaped haptic is available between 11 and 14 mm in increments of 1 mm.

As illustration, a clinical case is shown in Figure 27.9. This 3-year-old boy who was treated for congenital cataract at 6 months of age. He was referred to our department with a request for secondary IOL implantation. Figure 27.9A shows the condition of the eye at the time of surgical repair. It shows capsular phimosia and subsequent overstretching of the nasal zonular fibers. After peeling of the fibrotic tissue lining the internal border

of both the anterior and posterior capsulorhexes, the capsular bag was stretched by means of a capsular tension ring (see Fig. 27.9B). The capsular bag was then cleaned from any proliferation of lens material present at the capsular bag equator. The BIL IOL was then positioned as were two bean-shaped rings. The loops of the bean-shaped rings are visible within the groove of the BIL (see Fig. 27.9C). The peripheral loop of the bean ring has a diameter of 13 mm and is positioned into the ciliary sulcus. As a result, the BIL is firmly grasped by both bean haptics, preventing the BIL from wiggling when the eye is rotating.

FINANCIAL DISCLOSURE

M.J. Tassignon has an intellectual property in the bag-in-the-lens licensed to Morcher, Germany.

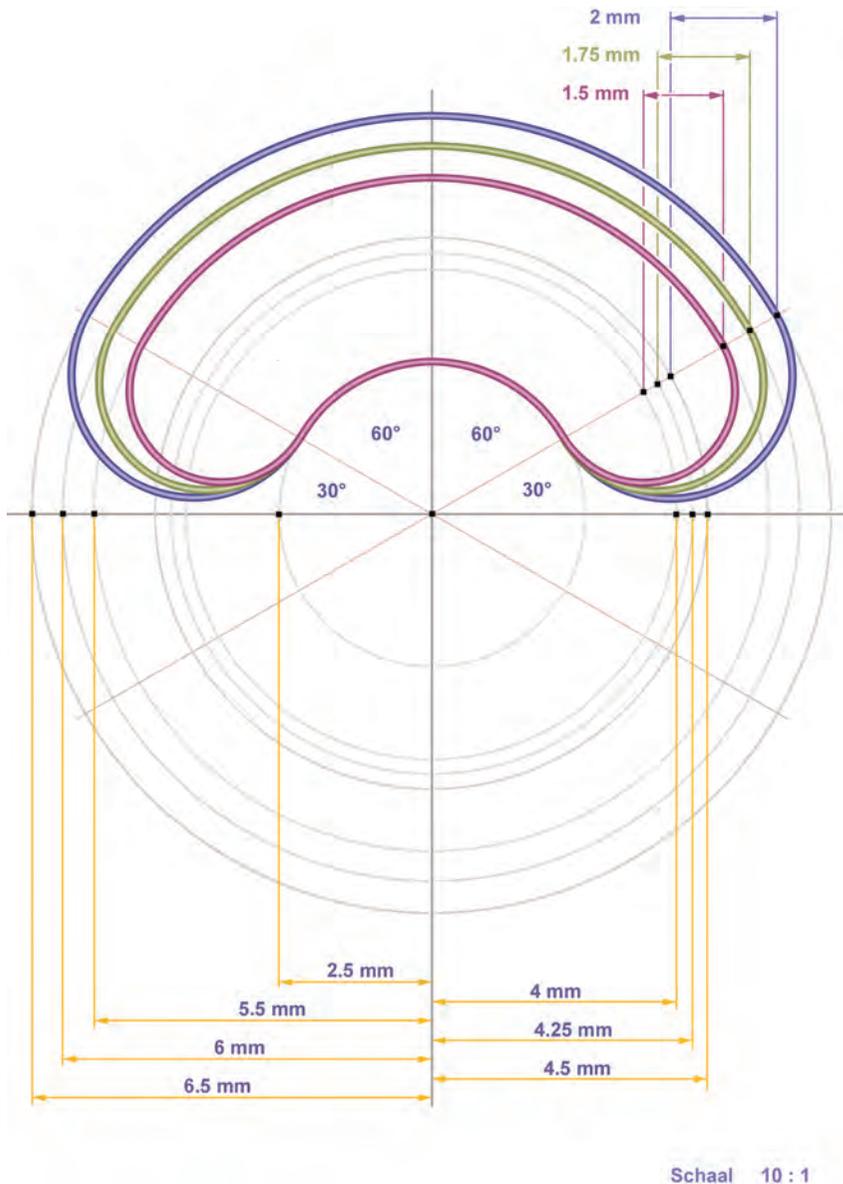


Figure 27.10. The bean-shaped rings are available with internal diameter of 5 mm but external diameters varying from 11 to 14 mm depending of the white-to-white diameter of the eye.

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Polypseudophakia (Piggyback IOLs)

M. Edward Wilson and Rupal H. Trivedi

Implantation of multiple intraocular lenses (IOLs; polypseudophakia or piggyback IOLs) has been described as a solution to the problem of providing adequate IOL power to adult patients with microphthalmos and extreme hyperopia.¹⁻⁴ We have used this concept in children, when indicated, since the mid-1990s (Figs. 28.1 to 28.3). Children with nanophthalmos can require very high IOL powers. Initially, when standard IOL powers extended only to a maximum of 30 diopters (D), primary piggyback IOLs were needed to attain the required power. More recently, IOL power ranges have extended to 40 D, reducing the need for this type of IOL piggybacking. We also sometimes use piggyback secondary IOL implantation in aphakic patients who become contact lens intolerant and are nearly full-grown but still have microphthalmia and require a high IOL power. Finally, a piggyback IOL is sometimes used instead of an IOL exchange when residual refractive error is present or develops over time. In each of these situations, the piggyback IOLs are expected to remain in the eye from that point forward.

In contrast, we introduced and developed (in the mid-1990s) the concept and technique of piggybacking IOLs to develop the technique of *temporary polypseudophakia* for infantile eyes.⁵⁻⁷ By mid-1999, one of us (M.E.W.) had implanted 13 eyes (10 of them infant eyes) with this temporary piggyback IOL technique. These were unilateral cataract patients who were deemed unlikely to comply with contact lenses for aphakia or thick hyperopic glasses for the residual hyperopia after a single IOL implantation. In a report published in 2000, we reported on bilateral piggyback implantation as well.⁵ In 2001, we reported on the short-term outcome of pseudophakia and polypseudophakia in the 1st year of life.⁶ This 2001 report included outcomes for 15 eyes of 11 infants who received piggyback IOLs in the first year of life (from 16 days to 6.8 months of age) and were followed for up to 22 months.

In the temporary piggyback technique, the posterior IOL is implanted in the capsular bag (permanent) and the anterior IOL is placed in the ciliary sulcus (temporary). This approach may help in the prevention and treatment

of amblyopia by eliminating residual hyperopia in small children after IOL implantation. The combination of a permanent IOL sequestered in the capsular bag and a temporary IOL placed in the ciliary sulcus—a location from which it can be easily removed later—makes this *temporary polypseudophakia* option attractive for the correction of aphakia in infancy, especially when compliance with glasses or contacts is expected to be poor. This technique minimizes the residual refractive error during very early growth and development (to minimize amblyopia) and yet allows the expected high myopia that occurs with growth to be treated with a planned removal of the lower power sulcus-fixated IOL after sufficient eye growth has occurred. As is discussed more later in the chapter, the technique is not intended or recommended for every infant and toddler needing IOL surgery. For those families who can comply with glasses or contact lenses, the effects of residual refractive error can be managed without needing to place multiple IOLs in the eye. The surgery can be technically challenging and requires a reoperation some years later to remove the sulcus-placed IOL. The placement of piggyback IOLs is a surgically aggressive technique that should be used when needed but avoided in favor of a less traumatic surgery when it is not needed.

Additional discussion for the rationale behind piggyback IOLs may be useful at this point in the chapter. Although uncorrected aphakia can be eliminated by implanting a single IOL in infancy, undercorrected aphakia may remain a frequent occurrence. Because the eye grows an average of 4.5 mm in axial length in the first 2 years of life, IOLs implanted in infancy are usually selected to produce a 20% or more undercorrection. This approach often produces from 6 to 18 D of residual hyperopia. If compliance with glasses or contact lens is poor, an amblyogenic hyperopic refractive error will be present despite the implantation of an IOL. However, if the combined IOL power implanted could be enough for early emmetropia, the amblyopia risk from residual hyperopia could be eliminated. If a single IOL is implanted early in life with enough power to achieve

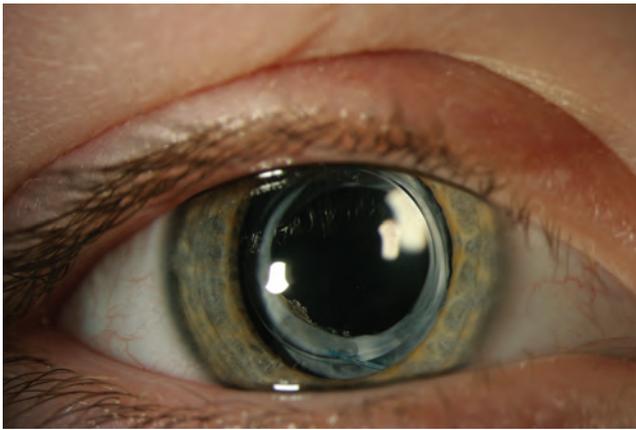


Figure 28.1. Nine-year postoperative appearance of an eye with piggyback IOL implantation. Note that surgery was done at 6 months of age for unilateral cataract using AcrySof® SA60 27 D IOL in the bag and 8 D MA60 IOL in the sulcus. Even at 9-year postoperative visit, the child still has both IOLs in the eye (refraction -1.25 D sphere, 0.5 D cyl at 85 degrees).

emmetropia, a high degree of myopia would undoubtedly occur later. In an effort to address this problem, we developed the piggyback IOL technique. A permanent IOL is placed in the capsular bag intended to be retained by the infant throughout life. An additional IOL is placed in a piggyback fashion in the ciliary sulcus.

As the eye grows, myopia develops and increases. Because it is well known that myopia is much less amblyogenic than is hyperopia, we have hopes that ultimately the visual acuity will be improved and amblyopia more easily treated. After the myopia increased beyond 4 D, glasses should be prescribed. However, if compliance with glasses is poor, the child can still attain a sharp focus with the polypseudophakic eye during occlusion of the normal eye by holding the object of interest close.

It must be noted, however, that high myopia can also be amblyogenic, and a second planned surgery will be

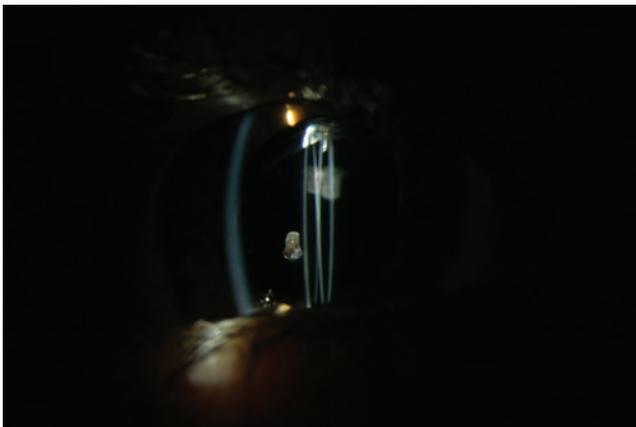


Figure 28.2. Twelve-year postoperative appearance of right eye with piggyback IOL implantation. Note that bilateral piggyback IOL implantation for nanophthalmos was done at 5 years of age at 1-week interval. Right eye has both IOLs 22 D.

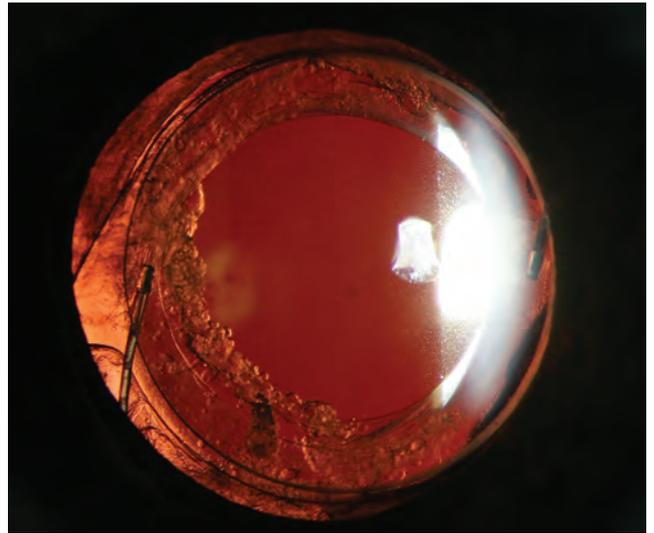


Figure 28.3. Five-year postoperative appearance of an eye implanted with piggyback IOL at 2 years of age. Note that refraction at this visit was -4.5 D sphere, 0.75 D cyl at 150 degrees. Removal of $+6$ D MA60 IOL was advised.

needed to remove the sulcus-fixated IOL. The best time to explant the anterior IOL is when biometry predicts a refractive error near plano when calculated using only the posterior IOL power. We perform serial biometry, yearly, to help determine when this planned explantation should occur. The biometry (done as part of an exam under anesthesia in young children) is also a recheck on the cycloplegic refraction since the two methods of predicting the refractive error of the growing polypseudophakic eye should agree within a small measurement error. If the measurements do not agree, we recheck them both to determine if our retinoscopy or the axial length measurement is the source of the error. Care must be taken when performing retinoscopy under anesthesia to measure the working distance and calculate the net retinoscopy accordingly. Often, working distances are shorter when refracting a supine patient in the operating room compared to the clinic setting.

Boisvert et al.⁸ have published their thoughts on choosing piggyback IOL powers. They suggest that the anterior IOL can be removed when the child's myopia equals half the anterior IOL power. IOLs placed in the ciliary sulcus do not scar in place, and they can be easily rotated, exchanged, and removed even several years after implantation. This has been a consistent finding for us over many years.

Choosing the optimal mix of IOL powers can be difficult to determine in advance. When implanting piggyback lenses in the first few weeks of life, we keep a target of $+2$ or $+3$ D because this mild hyperopia would likely disappear in the first 4 to 6 weeks. After 2 months of age, we keep the target postoperative refraction as plano since the eye will growth into mild myopia relatively quickly.

Glasses may not need to be prescribed if a low residual refractive error is present. The power of the temporary anteriormost IOL is chosen based on how much refractive change is anticipated during growth and development. Historically, we often amounted this to approximately one-third of the total IOL power. Boisvert et al.⁸ developed a theoretical strategy for choosing IOL power combination for temporary pseudophakia. They found that optimal results were obtained when the initial postoperative refractive goal refraction with polypseudophakia was moderate hyperopia, and the anterior IOL had approximately 20% of the total required power. With a higher anterior IOL power (30%), the final average refraction is closer to emmetropia, but considering the high variability of the rate of myopic shift, this strategy is more likely to yield a hyperopic refraction in adulthood. When choosing 10% of the total IOL power, the average refraction is too myopic. The curve was more balanced when selecting 20% as an initial strategy. This theoretical optimal strategy can be used by surgeons considering what piggyback IOL powers to implant in a child's eye. This may be used as a starting point and can be adjusted for every patient.

Our operative technique of piggyback IOL implantation is identical to that used for single IOL insertion except that an additional IOL is implanted in the ciliary sulcus immediately after the initial IOL is placed in the capsular bag. A single-piece AcrySof[®] IOL is used most often for placement within the capsular bag. For sulcus fixation, we use either the three-piece AcrySof[®] IOL or the Rayner C-flex IOL. Interlenticular opacification (ILO), a complication of capsule-fixed piggyback IOLs in adults, is avoided in our patients because one of the IOLs is placed in the ciliary sulcus.⁹ ILO seemed to be related to two posterior capsule IOLs being implanted in the capsular bag through a small capsulorhexis, with its margins overlapping the optic edge of the anterior IOL for 360 degrees. Analyses of the cases of ILO concluded that the opacification within the interlenticular space is derived from retained/regenerative cortex and pearls.

Pupillary capture of an IOL optic was noted in one of our patients with a sulcus-fixed IOL 1 day after the procedure and required reoperation.⁶ We now constrict the pupil with an intracameral miotic at the end of surgery. Gayton et al.¹⁰ have also noted pupillary capture of secondary piggyback IOL implantation.

Of note, we have also used the concept of polypseudophakia to treat pseudophakic refractive error, to avoid the risks associated with lens exchange. The additional manipulation required for the removal process of an IOL, particularly if the IOL is strongly fixed,

increases the risk of complications. A second IOL can be implanted anteriorly to the primary IOL. The power of a secondary piggyback IOL implant is also more predictable than that of an IOL exchange. For this secondary piggyback, we used the sulcus-preferred lenses mentioned above. In Europe, the Rayner Sulcoflex IOL is designed for this type of piggyback implantation. As of this writing, the Rayner Sulcoflex is not available in the United States.

SUMMARY

Infants and toddlers who are anticipated to have difficulty complying with contact lens wear and amblyopia therapy can be candidates for piggyback IOL implantation. It must be noted that not all infant eyes are suitable for piggyback lens placement; piggyback IOL implantation should be confined to eyes in which the benefit of this implantation exceeds the potential risks. With marked microphthalmia, we have sometimes elected not to place a second IOL because of insufficient space in the eye. Although we are waiting for long-term results in terms of the safety of this procedure and its impact on rapid myopization and visual outcome, it appears to be a useful option for addressing some of the questions associated with infantile IOL implantation.

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Multifocal and Accommodative Intraocular Lens Implantation in Children

M. Edward Wilson, Muralidhar Ramappa, and Rupal H. Trivedi

One of the most intriguing challenges of modern cataract surgery is the restoration of the accommodative ability in a pseudophakic eye. Although monofocal intraocular lenses (IOLs) offer excellent visual function, they do not restore any of the remarkable accommodation that is lost when the crystalline lens is removed from a child. With that said, subjects with small pupil size, myopic astigmatism, corneal aberrations, and corneal multifocality and subjects with good visual perception can have an increased range of what has been called “pseudo-accommodation” with monofocal IOLs. Monovision techniques (one eye corrected for distance and the other for near) have been used as a partial substitute for the absence of accommodation, but it may be at the cost of binocularity. Typically, if a monofocal IOL power is selected for distance correction, the family is informed that glasses will be needed for near and intermediate vision. However, it is remarkable how many times parents report that their bilaterally pseudophakic child functions well at near even when looking over the bifocals or when they are not being worn at all. While this is not universal for all children, it is a common occurrence that cannot be explained simply by depth of focus with the pupillary miosis that occurs with attempted accommodation. The importance of this finding is that any study of pseudo-accommodation in children implanted with multifocal IOLs needs a comparison group of age-matched children with similar residual refractive errors who were implanted with monofocal IOLs.

Newer IOLs designs have enabled adult cataract surgeons to both neutralize corneal astigmatism and reestablish the range of near and far vision without spectacles. The success of multifocal IOLs in the adult population has led to a growing interest in implanting them in children.¹ Unlike most adults who have cataract surgery with IOL

placement, children have excellent accommodation, so multifocal IOLs are often viewed as an attractive way to mediate the loss of the ability to zoom in and out at will. There are children who do not wear spectacles or contact lenses secondary to behavioral issues related to syndromes, neurologic issues, or other medical issues. Multifocal IOLs have been suggested for these children (or young adults) with special needs who develop cataracts requiring surgical intervention. There is a dearth of literature in children after multifocal IOLs with long-term outcomes.^{2,3} This chapter focuses on the multifocal and accommodative IOL designs available in the United States and Europe and those under investigation at this time.

MULTIFOCAL IOLS

Multifocal IOL designs are based on the concept of simultaneous vision. These IOLs focus light toward distant, near, and sometimes intermediate focal points to provide the patient near and distant targets.⁴ Visual disturbances can arise from light focused in multiple areas. The quality of the vision produced by a multifocal IOL is based on several factors, that is, pupil size, shape of the IOL, and the IOL's refractive or diffractive characteristics. Haloes around lights, glare, and decreased contrast sensitivity are potential problems with multifocal IOLs.⁴ For example, when attention is to light rays focused for the distant focal point, the retina still receives light for the near focal point, and these out-of-focus light rays may be interpreted as glare and lower contrast sensitivity. Currently, many multifocal IOLs have adopted aspheric designs to improve contrast sensitivity.⁴ Advantages are that many designs are similar to current foldable monofocal IOLs implanted in routine cataract surgery; minimal modification of surgical technique is required. Multifocal IOLs

currently give more predictable results for near vision when compared with single optic accommodating IOLs.⁵

IOL DESIGN VARIABLES

Different areas of the IOL have different focal planes, usually for near and distant vision. At any given time, one image is in focus at the retina, and the second image is highly defocused with very little structure. Distant objects are focused by the distance power of the lens and defocused by the near power. For near objects, the opposite is true; near objects are focused by the near power of the lens and defocused by the distance power.

OPTICS

Multifocal IOLs can be broadly categorized as diffractive or refractive.

Refractive IOLs

Refractive multifocal IOLs direct the light at different focal points using concentric zones of varying dioptric power within the optic. The principle is similar to the bifocal spectacle. These are also referred to as multizonal refractive IOLs.⁶ As the pupil size changes, the number of zones in use varies, and eventually, the relative proportion of light directed to the near and distant focal points changes as well.⁶ Thus, image quality can vary depending on pupil size. The ReZoom lens (Abbott Medical Optics, Santa Ana, CA) is an example of a refractive multifocal IOL.

Diffractive IOLs

Diffractive IOLs have closely arranged concentric rings on one of the surfaces of the lens to divide incoming light into multiple beams; they add together in phase at a predetermined point on the optical axis for near focus, while the overall curvature of the lens provides the distance focus. The number of the rings, spacing, and step heights vary by IOL design and manufacturer. The apodized design consists of concentric rings showing a decrease in height from the taller central diffractive steps to the shorter outer steps.⁶ The perceived advantages of apodization include the fact that the gradual change in step height decreases sudden shifts in optical boundaries, reducing distracting out-of-focus light rays that produce glare and haloes when viewing distant objects through a large pupil. As the pupil size increases, more light is focused to the distant focal point. The rationale for this design is that in mesopic conditions with dim light when the pupils are large, such as driving at night, distance vision is the priority. In most situations, humans rely on near vision in well-lit environments, such as reading under artificial light or in daylight, when the pupil is constricted. In these situations, apodization directs equal

light to both focal points.⁶ The ReSTOR IOL (Alcon Laboratories, Fort Worth, TX) is an example of an apodized diffractive multifocal IOL. Nonapodized diffractive multifocal IOLs have diffractive steps with fixed height from the center to the periphery. Thus, these lenses distribute light to the near and distant focal points in constant proportions, regardless of the pupil size.⁴ Current examples of nonapodized diffractive multifocal IOLs are the TECNIS Multifocal IOL (Abbott Medical Optics) and the AT LISA 809 IOL (Carl Zeiss Meditec Company, Hennigsdorf, Germany).

ACCOMMODATIVE IOLS

In accommodative IOLs, a shift in the focal length of the IOL-eye optical system is induced by a change in the ciliary muscle tension. In contrast to multifocal IOLs, accommodative IOLs are free from glare and haloes and result in improved contrast sensitivity since these lenses are designed to focus light at the desired focal point.⁷ Accommodating IOLs are the most diverse in terms of design.

SINGLE-OPTIC ACCOMMODATING IOLS

The presumed mechanism of accommodation is a forward axial shift in the optic of the IOL and changes in the lens architecture as a result of contraction of the ciliary muscle.⁷ Theoretically, the accommodation is achieved by combined mechanisms. It is believed that pseudoaccommodation might play a role in addition to changes in lens position and lens architecture.⁸ In the case of the Crystalens (Bausch & Lomb, Rochester, NY), the optic may curve anteriorly, which changes the radius of curvature of the anterior surface of the optic, resulting in more near vision.^{4,8} The Crystalens HD variant has a small refractive number add centered in the optic to improve depth of focus for intermediate and near distances.^{8,9} This essentially adds a little refractive multifocal feature to the accommodative lens design. The Crystalens AO is an aspheric version. One study comparing the Crystalens HD to a monofocal IOL found a mean accommodating range of 1.5 ± 0.0 D in the Crystalens HD group and 1.0 ± 0.0 D in the monofocal IOL group.⁹ The Tetraflex (Lenstec, St. Petersburg, FL) is another single-optic accommodating IOL (not yet approved in the United States). Vitreous pressure and possibly ciliary muscle activity change the shape of the Tetraflex lens to increase higher-order aberrations such as coma, trefoil, and spherical aberration, which augment the depth of field.¹⁰

DUAL-OPTIC ACCOMMODATING IOLS

To maximize the amplitude of accommodation, the dual-optic IOL design was developed.⁹ The degree of accommodation depends on two decisive factors, that is, the

range of axial displacement of the IOL optic and the power of the displaced IOL.^{11,12} For example, a +19 D lens in the bag with 1 mm of axial displacement yields a +1.2 D change in accommodative power. However, a +32 D lens in the bag with the same displacement gives in +2.6 D of accommodative change.¹¹ The same model suggests limited accommodation (0.3–1.9 D) for 1 mm of axial displacement for IOL powers in the +15 to +25 D range. In a dual-optic system, moving a strong plus power lens coupled to a stationary negative minus lens should result in a greater accommodative change compared with a single-optic system.^{11,12} The Synchrony (Abbott Medical Optics) is an example of a dual-optic accommodating IOL. This dual lens system rather resembles a Galilean telescope, with an anterior convex lens connected to the posterior concave lens.⁷ However, there are some notable distinctions between a dual-lens accommodative IOL and a Galilean telescope. The dual-lens accommodative IOL is designed to provide vergence in the range of an IOL (+15 to +30 D), whereas a Galilean telescope produces 0 vergence.¹² A telescope magnifies an image, which could result in uncomfortable aniseikonia in a patient who has a telescope implanted in one eye and a single-optic IOL in the other. Image magnification in the Synchrony is limited to 2.5%, which is within the reported 8% tolerance before patients notice aniseikonia.¹² When the IOL is in the bag, tension from the capsule compresses the optics closer together. When accommodation is attempted, the ciliary muscle contracts, relaxing tension in the bag. The dual IOL optics separate due to the stored energy in the bridging haptics. Pressure from the vitreous face supports the posterior optic so the anterior optic moves forward during accommodation.¹² The haptics are designed to allow 1.5 mm of optic movement. In one study, the mean accommodative range of the Synchrony was 3.22 ± 0.88 D compared with a monofocal IOL, which measured 1.65 ± 0.58 D.⁷ A disadvantage of the Synchrony is a larger 3.8-mm incision required for implantation.⁷ In postvitrectomy eyes, it is unclear how well dual-optic accommodating systems perform.

CLINICAL STUDIES

Wilson et al. have published their understanding about refractive change during eye growth as well as amblyopia secondary to the loss of contrast sensitivity associated with IOL implantation.^{1,13} As recommended by their study, these issues can be partially dealt with by restricting the use of multifocal lens to children older than 10 years of age (Fig. 29.1). As 80% to 90% of the eye growth occurs in the first 2 years of life, some surgeons argue that multifocal IOL implantation could be offered in a younger population.¹⁴ The multifocal IOL requires precise measurements, calculations, and positioning for satisfactory performance. For the optimal performance of multifocal

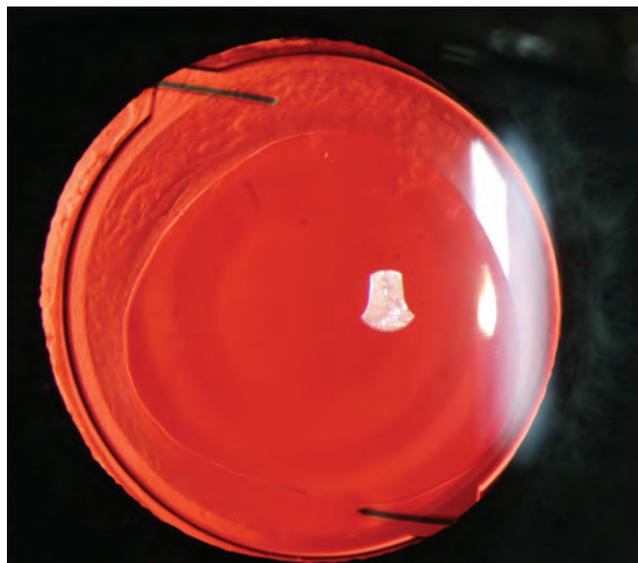


Figure 29.1. Twelve-year follow-up after array cataract surgery and multifocal IOL implantation at 17 years of age. She wears glasses for fine reading (distant uncorrected 20/15, near uncorrected 20/30 both eyes).

IOLs, it has been found that a residual hyperopia up to 0.50 D helps in minimizing halos and to maintain distance and near visual acuity.¹⁵ In addition, postoperative residual refractive error plays a critical role in enhancing patient satisfaction.¹⁵ The multifocal IOL designs do not function as well when the eye grows and becomes myopic. Ironically, an eye with a multifocal IOL may actually be more spectacle dependent than a monofocal IOL eye when a myopic shift occurs. With multifocal IOLs in mind, Wilson et al.¹³ looked at axial length changes in the second decade of life and noted an average axial length of 23.36 mm at 11.5 years of age and 23.86 mm at 15.2 years of age. Average growth during this time frame was 0.53 mm. However, there was variable growth throughout the second decade of life. Their data showed 57.1% of the eyes grew 0 to 0.5 mm, 37.8% grew 0.5 to 1.5 mm, and 5.1% grew more than 1.5 mm. The authors calculated the dioptric change if a patient followed the average growth and found it to be 4 D between ages 10 and 20 years. This study suggests caution in the use of multifocal IOLs during the second decade of life.

Jacobi et al.² performed a prospective case study on 35 eyes in children aged 2 to 14 years implanted with zonal-progressive multifocal IOLs. The average age at implantation was 6.1 ± 3.4 years with an average follow-up of 27.4 ± 12.7 months. All patients showed improved vision, with 71% having 20/40 vision or better and 31% having 20/25 vision or better. The average distance vision was 20/35, and the average near vision was 20/55. The near vision improved to 20/35 with the use of additional plus lenses. In the cases with bilateral multifocal IOL implantation, spectacle dependence was reported in 67%

of patients. Reported complications include posterior synechia (54%), visual axis opacification (46%), moderate to severe fibrin reaction (34%), and visually significant IOL decentration (11%). Of nine children who underwent bilateral implantation, six (67%) reported wearing glasses throughout the day, two with bifocals and four with distance-only glasses. Among those older than 6 years of age at last follow-up, three children (16%) reported glare and halos. The optical performance of a multifocal IOL is highly sensitive to lens tilt and decentration. Seventeen percent of patients in the study by Jacobi et al.² required surgical intervention to treat IOL decentration. The pediatric capsule is well known to experience aggressive fibrosis and opacification after cataract surgery. Any shift in IOL position, that is, tilt and decentration, can potentially deteriorate the optical performance of these IOLs.

To the best of our knowledge at this point in time, there are no publications on accommodating IOL usage in children for the treatment of aphakia in available databases (PubMed, Cochrane Library, and Google Scholar).

OPTIMIZING THE RESULTS

In the editorial that accompanied the Jacobi et al. article, Hunter urged caution, especially when implanting these IOLs in younger children when surgical complications may be more common. Decentration of the IOL could disrupt the multifocality of the IOL, and posterior synechia could cover up the IOL zones and functionally convert it to a monofocal lens. He was particularly cautious about using these lenses in children at risk for amblyopia. He points out that the reduction in contrast sensitivity from the multifocal lens design may not be large, but it may nevertheless induce a corresponding degree of amblyopia. Tychsen, however, expressed a different view.^{13,14} The minimally degraded contrast sensitivity of a multifocal IOL, according to Tyschen, is not an important disadvantage. Rather, the multifocal IOL may give the child a zone of some 3½ D of optical “play,” and this may aid in their amblyopia therapy. Managing expectations is important. Children who are in their growing years are not likely to be spectacle free long-term. Tychsen recommends using the multifocal IOL to provide the growing child with a broader band of potentially clear, less amblyogenic vision with and without glasses.

Rychwalski,³ in his editorial on the utility of multifocal IOLs in the pediatric age group, has raised several pertinent issues relevant for determining the suitability of primary multifocal IOL implantation in pediatric patients. Most importantly, there is a dearth of published data regarding pediatric multifocal IOL use. Without data, it is difficult to make proper evidence-based assessments of a therapy. Postoperative healing following primary implantation in children will often include posterior opacification related to the posterior capsule

or its remnants, synechia, or fibrotic changes anteriorly. While a component of this correlates positively with age at the time of surgery, proper surgical technique including posterior capsule management (i.e., polishing, posterior capsulorhexis with or without anterior vitrectomy) at the time of surgery can decrease the observed complication rate of lens decentration, posterior synechia, and posterior opacification following surgery. The trade-off for slightly greater glare disability and decreased contrast sensitivity is the increased depth of focus provided by multifocal IOLs.^{16,17}

TO SUMMARIZE

Ultimately, it is our belief that accommodating IOLs will surpass multifocal IOLs, and eventually, multifocal IOLs will be remembered as a historical stepping-stone prior to the development of high-performing accommodating IOLs. The concept of simultaneous images and the splitting of available light with its loss of contrast sensitivity and potential for photopsias will last in the marketplace so long as a better alternative is not yet available. However, but at this time, multifocality is the more successful and popular option for correcting presbyopia in adults. For children, the fact that these lenses have a long track record in adults makes them a viable consideration for certain children. Pediatric surgeons must carefully consider the rather vast literature now available from adult use and the small amount of pediatric data when deciding whether their patient is a good candidate for multifocal IOL implantation. Proper preoperative evaluation, lens selection, astigmatism control, and postoperative care are paramount when considering the multifocal IOL implantation. An accurate biometry is essential in obviating the need for spectacles dependency after multifocal IOL implantation.¹⁸ Status of the fellow eye should be considered to avoid conditions that may preclude optimal results, such as extreme aniseikonia. Unilateral multifocal IOL implantation in a child who has normal accommodation in a noncataractous fellow eye is questionable since the accommodating eye will always be used at near preferentially. Multifocality, if it has a role to play in children, will be useful mainly in bilateral implantations. In eyes with a small, eccentric, or nonreacting pupil,^{19,20} it is prudent to choose a lens that gives optimal performance irrespective of pupillary size. The architecture of corneal incision, location, final wound size, and technique of wound closure can have direct influence of on induced astigmatism. Astigmatism-induced deterioration of both distance and intermediate visual acuity was significantly worse with multifocal IOLs than with monofocal IOLs.²¹ Therefore, care should be taken to avoid the extent of surgically induced astigmatism by appropriately designing the wound architecture or choosing the correct toric multifocal IOL.^{22–24} Finally, knowledge of the myopic

shift that may occur after implantation much be taken into account and planned for. Appropriate expectations for long-term spectacle use need to be emphasized to avoid patient and family disappointment.

SUMMARY

Several inherent limitations of multifocal IOLs have, so far, restricted their use in the pediatric population. As IOL designs continue to improve, the newer-generation multifocal IOLs seem to be addressing the problems of glare, halos, optical aberrations, and poor intermediate vision. Accommodating IOL designs will also improve and will eventually command a larger share of the IOL market. The multifocal IOL may play a pivotal part for selected pediatric patients, especially if surgeon and parent expectations are clearly identified and understood. The pediatric cataract surgeon is obligated to proceed cautiously in the young amblyopia patient as the decision of which IOL to implant will affect not only visual acuity but also general visual maturation. Further study is needed before these IOLs can be recommended for routine use in the pediatric patient.

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Astigmatism Correction in Children with Cataracts

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Corneal astigmatism is often documented during preoperative biometry measurements in children and adults. In infancy, there is a high prevalence ($\geq 42\%$) of astigmatism (≥ 1.00 D).¹⁻¹³ A reduction in this infantile astigmatism occurs in the first years of life because of a natural decrease in toricity of the cornea and the anterior lens, along with a decrease in the variation of the cornea and lenticular surfaces.¹⁴ Hispanic and African-American children are more likely to have astigmatism than are non-Hispanic white children, and this association remains even after correcting for the presence of spherical equivalent refractive error (myopia or hyperopia).¹⁵ In adults, residual postoperative corneal astigmatism, if uncorrected, has been noted to reduce uncorrected visual acuity and induce symptomatic blur, ghosting of images, and halos.¹⁶ Children with astigmatism > 2.00 D in 3-year-olds or > 1.50 D in 4-year-olds have been shown to have visual motor integration deficits.¹⁷ Amblyopia treatment seems to be less favorable in patients with either hyperopic or myopic against-the-rule (ATR) astigmatism.¹⁸

To meet parents' expectations for good visual acuity after cataract surgery and to help amblyopic management, it is important to address both sphere and cylinder errors that may affect final refractive outcomes. Parents want to know how clear the vision will be when glasses are removed, even if it has been explained that the child's best visual acuity will always be with bifocal glasses. Preoperative assessment for cataract surgery should be followed by comprehensive counseling that focuses on a patient's individual vision needs. The refractive part of the discussion should include a discussion about the refractive aim for immediately after surgery and also what the refraction is expected to be years later. In a young child, this may include a discussion of far-sighted bifocal glasses after surgery with a gradual reduction in the strength of the glasses as the eye grows. If the child is older than 3 years and has preoperative astigmatism, parents are told that the astigmatism will likely be unchanged after surgery and will be corrected with glasses. For older children, a review of surgical options for reducing astigmatism is becoming

more common. These options include astigmatism management with peripheral corneal-relaxing incisions (PCRIs), toric intraocular lens (IOL) (Fig. 30.1), and the possibility of postoperative laser refractive surgery. It is important to estimate the likelihood of less spectacle dependence based on the age that can realistically be achieved with a given surgical option. Videos or computer-based illustrations may be a useful and effective way of explaining the different approaches. Spherical refractive errors are managed by accurate biometry and IOL power calculations. The target refraction should be based on age at surgery and other factors (see Chapter 7). Relying solely on manual keratometry to characterize a patient's corneal curvature is commonly done during standard biometry for IOL power calculations. However, if the surgeon desires to surgically manage astigmatism, manual keratometry may be insufficient. Significant cylindrical errors may exist inside and outside the central 3.2-mm optical zone measured by keratometry. In addition, meridian changes over the entrance pupil and irregular astigmatism may be missed. Using the IOL Master® (Carl Zeiss Meditec, CA, USA) for corneal curvature measurements can result in similar mistakes.¹⁹ For these reasons, computerized corneal topography is the current standard of care when astigmatism correction is being considered. Topography accurately measures the global corneal astigmatism, quantifies the nature of the astigmatism (i.e., symmetric bow tie versus asymmetric or irregular astigmatism), and identifies the steep meridian of astigmatism. Patients implanted with monofocal IOLs usually achieve better-uncorrected visual acuity if the spherical error approaches emmetropia and residual manifest astigmatism is less than 1 D. For patients implanted with multifocal IOLs, the postoperative refractive goal is to achieve < 0.75 D of residual corneal astigmatism. However, as mentioned above, children have a targeted refraction that depends on age at surgery. Final results are also dependent on the pupil size and the amount of residual spherical error. Overcorrections and large rotations in the axis of residual astigmatism should be avoided.

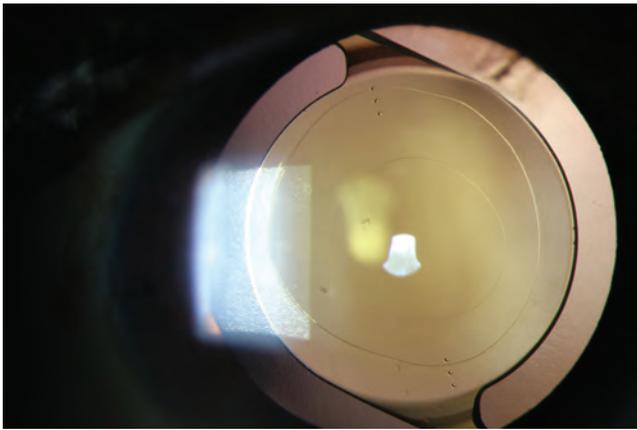


Figure 30.1. Six-month follow-up of toric IOL implantation in a child.

Astigmatism correction at the time of cataract surgery or in aphakic/pseudophakic eyes of children is still uncommon and is only now beginning to be reported.^{20,21} However, witnessing impressive outcomes in adults, pediatric cataract surgeons have started discussing correction of astigmatism in children. In the following sections of this chapter, we review the literature on astigmatism correction in adult eyes. We anticipate a more conservative approach in the growing eyes of children.

STEPLADDER APPROACH TO ASTIGMATISM MANAGEMENT

Subjects with symmetrical corneal astigmatism can be placed into one of four categories.²² These categories are as shown in Table 30.1. Cases with <1 D of corneal astigmatism can be neutralized by designing the surgical incision on the steep corneal meridian. For 1 to 3 D

of corneal astigmatism, single or paired PCRI's may be fashioned. Toric IOLs may be implanted in eyes with 1 to 4 D of corneal astigmatism. Toric IOLs and PCRI's may be combined for eyes with 4.5 to 7 D of corneal astigmatism. Finally, astigmatism of >4.5 to 6 D may warrant the use of high-power toric IOLs that are available in Europe and elsewhere.

Miller PCRI Nomogram

For symmetrical bow tie corneal astigmatism on corneal topography, clear corneal-paired incisions are placed in the steeper axis, just inside the limbus. The incisions are 500 to 550 microns in depth and as long in clock hours as the cornea is steep in diopters by simulated keratometry. For asymmetric bow tie astigmatism, in which the axis of astigmatism is constant across the cornea, longer incisions are made on the side of the cornea that is steeper. The astigmatic correction in diopters equals the sum of the lengths of the incisions in clock hours divided by 2. For asymmetric bow tie astigmatism in which the axis of astigmatism changes across the cornea, the incisions are rotated to coincide with the location where the steep axis intersects the limbus. Surgical management options in cases of very high postoperative residual astigmatism include piggyback IOL implantation, IOL exchange, toric IOL rotation, photorefractive keratectomy (PRK), and laser-assisted in situ keratomileusis (LASIK).

Concepts in Applying the Stepladder Approach

Surgical incisions placed on the steep meridian and PCRI's do not alter the spherical equivalent power of the cornea enough to alter IOL power calculations. The location and architecture of the corneal incision are the most important surgical variables. Any clear corneal incision (CCI) flattens the meridian in which it is placed and steepens the perpendicular meridian with a coupling effect.²³ For a 3.2-mm wide incision, the resultant surgically induced astigmatism (SIA) is approximately 0.5 D (95% CI: 0.4–0.6 D).²⁴ Incisions <2.4 mm wide may induce smaller degrees of SIA, but not in a linear logarithmic manner and not significantly <0.5 D, possibly due to surgical manipulation while IOL implantation. Consequently, when placing a cataract incision on the steep meridian, the surgeon can predict an approximate 0.5-D correction of preexisting corneal astigmatism, which is ideal for patients with <1 D of preexisting corneal astigmatism. To accurately calculate surgeon-specific SIA, software is available.²⁵ Preferably, it is better to place a CCI inside a convenient PCRI if a relaxing incision approach to astigmatism management is planned. If a toric IOL is planned, the surgeon must determine the vector sum of the preexisting corneal astigmatism and SIA before selecting the appropriate IOL to be implanted.^{23,26} Most of the toric IOL manufacturers

Table 30.1 STEPLADDER APPROACH TO THE MANAGEMENT OF CORNEAL ASTIGMATISM AT THE TIME OF ADULT CATARACT SURGERY

Magnitude of Corneal Astigmatism	Treatment Approach
<1 D	CCI on the steep axis of corneal astigmatism
1–3 D (if a toric IOL option is unavailable)	PCRI's
1–4 D	Toric IOL implantation
4.5–7 D	Toric IOL implantation plus PCRI's
>6 D	Obtain a compassionate use exemption and local Institutional Review Board approval to implant a high-power toric IOL. These devices not currently U.S. FDA approved

Table 30.2 ONLINE TORIC IOL CALCULATORS

Company	Web site
Alcon Laboratories (TX, USA)	www.acrysoftoriccalculator.com
Carl Zeiss Meditec (Jena, Germany)	www.meditec.zeiss.com/iolmaster-online
Rayner (East Sussex, UK)	www.rayner.com/raytrace
STAAR Surgical (CA, USA)	www.staartoric.com

(Alcon Laboratories, STAAR Surgical, Rayner, and Carl Zeiss Meditec) have toric IOL calculator software that performs this function (Table 30.2). These calculators use subject's corneal topography measurement and SIA to select an appropriate toric IOL model and determine orientation IOL in the capsular bag.

The Steep Meridian Clear Corneal Incision

A 3.2-mm CCI results in an SIA of 0.5 D (95% CI: 0.4–0.6 D).²⁴ Hill suggests making incisions <2.4mm wide does not reduce SIA much below 0.5 D, perhaps because these incisions stretch during IOL implantation.²³ Accordingly, for patients with <1 D of preexisting corneal astigmatism, placing the CCIs on the steep meridian of corneal astigmatism is the ideal approach. The surgeon must be capable of moving the CCIs to different meridians and comfortably operating at different positions around the patient's head. Some right-handed surgeons may prefer to place superotemporal main incisions in right eyes and superonasal main incisions in left eyes.²⁷ Although the mean SIA by vector addition may not markedly differ between the two groups, individual patient outcomes may vary significantly if the surgeon consistently places the incision in the same location without accounting for preexisting corneal astigmatism.

Peripheral Corneal-relaxing Incisions

PCRI can be used to manage corneal astigmatism up to 3 D. The risks associated with PCRI include postoperative regression and wound dehiscence. PCRI are a reasonable option in markets where toric IOLs are unavailable and in eyes with up to 3.0 D of preexisting corneal astigmatism.²⁸ Numerous PCRI nomograms are available, differing by whether they specify incision length in millimeters, degrees, or clock hours and whether or not they compensate for age.^{22,29–34} Degree and clock-hour nomograms compensate for corneal diameter, whereas millimeter nomograms do not. One particular advantage of a clock-hour nomogram is that it permits simple mental visualization of incision length, sparing the need for incision-length markers.

For regular corneal astigmatism, the paired incisions with 90% in depth should be placed in the steep corneal

meridian. The incisions are as long in clock hours as the cornea is steep in diopters as measured by simulated keratometry.³² For example, paired incisions that are each 2.2 clock hours in length would be made in the peripheral cornea to treat 2.2 D of regular corneal astigmatism. The corneal tunnel incision is placed through one of the PCRI, usually the one closest to the temporal or superior cornea. Corrections of up to 3 D can be achieved using this nomogram. Because of the availability of toric IOLs, we typically limit our use of PCRI to treat between 1 and 1.5 D of corneal astigmatism in adults.

For irregular corneal astigmatism, in which the steep meridian is constant across the cornea, longer incisions should be made on the side of the cornea that is steeper. The more general nomogram applies here, that is, the sum of the lengths of the incisions divided by 2 gives the astigmatic correction in D. Supposing, if a patient has 2 D of asymmetric corneal astigmatism, a 3–clock-hour relaxing incision on the steep side of the steep meridian might be paired with a 1–clock-hour relaxing incision on the flatter side of the steep meridian. In this case, $(3 + 1)/2 = 2$ D of correction. If a patient has 1 D of asymmetric corneal astigmatism, a single 2–clock-hour relaxing incision might be made on the steep side of the steep meridian. The corneal tunnel incision is placed through the center of this incision. In this case, $(2 + 0)/2 = 1$ D of correction. For asymmetric bow tie astigmatism, in which the meridian changes across the entrance pupil, the incisions should be rotated into the locations where the steep meridians intersect the limbus. The standard incision length rule applies.

It is important to note that the clock-hour nomogram undercorrects younger patients. Longer and/or deeper incisions need to be made in younger eyes to achieve the same benefit. PCRI should not be fashioned if there is evidence of corneal ectasia. PCRI should be made before any other incisions. Doing so ensures consistent intraocular pressure and blade depth, thereby reducing the likelihood of undercorrection. Before making PCRI, the cornea should be dry so to recognize any inadvertent entry into the anterior chamber with the keratotomy blade. If aqueous leak is encountered, it can be sealed by stromal hydration or wound suturing. Some surgeons like to measure corneal thickness at the sites where they will make their PCRI. However, pachymetry can usually be deferred when using a blade depth setting of 500 to 550 μm if the topography and slit-lamp examination preclude peripheral corneal pathology.

TORIC IOL IMPLANTATION

Toric IOLs can be used to treat 1 to 12 D of cylinder in the IOL plane, but their availability depends on the local markets and regulatory approvals. A variety of toric IOL models are available worldwide at the time of this writing

Table 30.3 COMMERCIALY AVAILABLE TORIC IOLS

Manufacturer	Model	Toric Correction in the IOL Plane (D)
Alcon Laboratories	SN60T3, SN6AT3	1.5
	SN60T4, SN6AT4	2.25
	SN60T5, SN6AT5	3
	SN60T6, SN6AT6	3.75
	SN60T7, SN6AT7	4.5
	SN60T8, SN6AT8	5.25
HumanOptics	SN60T9, SN6AT9	6
	Torica-s	2–12 in 1-D increments
	Torica-sY	2–12 in 1-D increments
	Torica-sS	2–12 in 1-D increments
	Torica-sSY	2–12 in 1-D increments
	Torica-sPB	1–6 in 1-D increments
Rayner	Torica-sPYB	1–6 in 1-D increments
	T-flex Aspheric Toric IOLs	1–11 in 0.25-D increments
	573T	1–11 in 0.25-D increments
STAAR Surgical	623T	1–11 in 0.25-D increments
	AA4203TF	2 and 3.5
Carl Zeiss Meditec	AA4203TL	2 and 3.5
	AT LISA toric	1–12 in 0.5-D increments
	909M/Acri.LISA toric 466TD	1–12 in 0.5-D increments
	AT TORBI 709M/Acri.Comfort 646TLC	1–12 in 0.5-D increments

(Table 30.3). In the United States, we have access to toric IOLs to treat 1 to 4 D of astigmatism in the corneal plane. Since their first reported use by Shimizu et al.,³⁰ numerous clinical studies have shown good postoperative refractive outcomes.^{35–43} Unlike PCRI, which function by correcting preexisting corneal astigmatism at the source, toric IOLs compensate for corneal astigmatism in the IOL plane. As the source of astigmatism is separated a small distance from the toric correction in the IOL plane, toric IOLs have the potential to induce image distortion.

The surgeon must take into account the vector sum of the preexisting corneal astigmatism, age-related astigmatism changes in children, and the SIA when determining the power and meridian of a toric IOL to implant.^{20,22,23} By using manufacturer-specific toric IOL calculators (see Table 30.2), a surgeon can readily determine the appropriate lens power and orientation for neutralization of corneal astigmatism.

The important factors, which determine a predictable outcome when implanting toric IOLs, are axis misalignment at the time of implantation and axis rotation during the early postoperative period.^{30,44} Theoretical calculations demonstrate approximately a 3.3% loss in compensatory astigmatic correction for every degree of IOL off-axis and a 33% loss of astigmatic correction if the optic rotates 10 degrees.⁴⁵ With a greater amount of

misalignment, a greater loss of astigmatic compensation occurs. Approximately two-thirds of the effect is lost with a 20-degree rotation, and a net increase in astigmatism results when the lens is off-axis more than 30 degrees.^{34,46} Thus, correct intraoperative alignment of the IOL and good rotational stability are critical factors for achieving the desired astigmatic correction. To assure proper alignment, the surgeon should make a reference mark at the limbus (typically at 6 o'clock or the 12 o'clock and 6 o'clock hour positions) with the patient in the upright, seated position before surgery commences. The reference mark is important since ocular rotation can occur when the patient lies supine under anesthesia. The reference mark can be placed in the preoperative holding area, at a slit-lamp biomicroscope with the chin and brow fixed in a centered position, or at a slit-lamp camera that combines with a topographic analyzer.⁴⁷ Thereafter, a Mendez Gauge, Dell Marker, or other axis marker can be used under the microscope to mark the appropriate axis of IOL alignment. All viscoelastic material must be completely out of the anterior chamber and capsular bag before final IOL positioning.

Correction of Residual Astigmatism Following Cataract Surgery

As stated previously, patients implanted with monofocal IOLs can achieve 20/20 uncorrected visual acuity if the spherical equivalent error is approximately emmetropic and the residual manifest astigmatism is <1 D in the absence of preexisting amblyopia. Rarely, it is necessary to perform additional surgery to control postoperative astigmatism. However, to correct residual astigmatism after cataract surgery, there are several surgical options, including piggyback IOL implantation, IOL exchange, PRK, and LASIK. As a piggyback option, the Rayner Sulcoflex[®] toric IOL is capable of correcting –3 D to +3 D of spherical error in 1 D toric increments from 1 to 3 D. For patients with multifocal IOLs, good unaided visual acuity can be achieved with <0.75 D of residual corneal astigmatism. Finally, a reliable contingency plan should be part of every pediatric cataract surgeon's practice to manage suboptimal refractive surprises, whether this plan involves management within the practice or referral to a refractive surgeon outside the practice.

SUMMARY

This simple stepladder approach can be followed to guide astigmatism management at the time of cataract surgery. Doing so will minimize a patient's chance of a significant postoperative cylinder error, thereby minimizing the spectacle dependence and risk of meridional amblyopia. Preexisting corneal astigmatism is a significant component of preoperative ametropia. To

minimize the risk of a child developing meridional amblyopia, it is important to correct preexisting spherical–cylindrical errors by accurate biometry and IOL power calculation and to manage preoperative corneal cylindrical errors by a suitable method. The technologic advancements enable pediatric cataract surgeon to effectively tackle preexisting corneal astigmatism at the time of cataract surgery. Corneal astigmatism can be placed into a variety of dioptric categories guided by corneal topography. The degree of refractive error guides the optimum approach to surgical correction. For patients with <1 D of corneal astigmatism, the preferred approach is to construct the surgical incision on the steep meridian. For 1 to 3 D of corneal astigmatism, single or paired PCRI may be used. Alternately, for astigmatic errors of 1 to 4 D, toric IOLs can be implanted. Astigmatism of 4.5 to 7 D may be addressed using a combined approach of toric IOL implantation and PCRI placement in the steep meridian of corneal cylinder. Moreover, high-power toric IOLs are currently available in select markets and not currently approved by the U.S. Food and Drug Administration. If visually significant refractive errors are present after cataract surgery, surgical management options include piggyback IOL implantation, IOL exchange, toric IOL rotation, PRK, and LASIK. Of these options, PRK and LASIK usually provide the simplest solution to manage suboptimal refractive outcomes. This simple stepladder approach to astigmatism management, which is based on preoperative corneal topography, can help pediatric cataract surgeons minimize refractive error, thus reducing the risk of amblyopia.

FUTURE

Newer technologic advancements in the field of refractive cataract surgery are expected to change considerably over the next few years. Femtosecond lasers will be used in combination with preoperative photos and iris registration to enable intrastromal PCRI. The Calhoun Vision (Calhoun Vision, Inc., CA, USA) light-adjustable lens is capable of postimplantation refinement in spherical power in the range of -2 to $+2$ D and astigmatic corrections in the range of -3 D to $+3$ D.^{48–50} New high-power toric IOLs^{42,46,51,52} are currently available in select markets and are expected to come into widespread use. Intraoperative wavefront aberrometer (WaveTec Vision's ORange) which lets the surgeon fine-tune spherical and astigmatic lens choices by measuring the refraction when the eye is aphakic and checking the refraction after the new lens is implanted.^{30,53–55} It is important for the surgeon to constantly keep current with technologic advances.

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Secondary Intraocular Lens Implantation in Children: In-the-Bag and Ciliary Sulcus Fixation

Rupal H. Trivedi and M. Edward Wilson

The use of primary intraocular lenses (IOLs) for pediatric aphakia has become increasingly common in recent years and is now a well-accepted approach for children beyond infancy. Nevertheless, the implantation of an IOL at the time of cataract surgery in children during infancy remains controversial.¹ The Infant Aphakia Treatment Study (IATS) was a multicenter randomized controlled trial comparing (1) primary IOL implantation (with glasses for residual hyperopia) with (2) primary aphakia with contact lens correction for unilateral cataracts operated from 1 through 7 months of age. In the report detailing the 1-year IATS results, the authors stated that caution should be exercised when performing IOL implantation in children aged 7 months or younger given the higher incidence of adverse events and the absence of an improved short-term visual outcome compared with contact lens use.¹ Many of these aphakic eyes will later require secondary IOL implantation. Secondary implantation of an IOL is generally recommended when traditional spectacle or contact lens correction of aphakia is unsuccessful. In addition, many parents electively choose secondary IOL implantation for their aphakic children when eye growth slows after age 4 years. Silsoft contact lenses are well tolerated in very young children, but the material is less well tolerated as children get older. The silicone contact lenses develop more deposits and the lens coating breaks down more quickly when worn in older children. Rather than change to a nonsilicone contact lens material, some parents will opt to have a secondary IOL implanted at that point.

To the best of our knowledge, the first IOL implantation in a child was performed as a secondary implantation in 1952. Dr. Edward Epstein performed this secondary IOL implantation for a traumatic cataract in a 12-year-old girl. A cruciate needling was performed on a cataractous lens on April 2, 1952. The lens became hydrated, and

much of the cortex was absorbed. On June 26, 1952, the residual cortex was washed out and a Ridley IOL was inserted. Forty-six years later, the best-corrected visual acuity was 20/20, and the IOL remained centered and optically clear (cited in Ref.²). While proceeding with secondary IOL implantation, surgeons face some important questions. Should I implant in this eye? If yes, what is the best site for fixation and what type of lens material should I use? This chapter reviews the literature on bag-fixated and sulcus-fixated secondary IOL implantation. In the absence of sufficient capsular support, anterior chamber, sutured, or iris-fixated IOL may be used (see Chapter 32).

AGE AT IMPLANTATION

In an article published by us in 2005, the average age at secondary implantation was 7.8 ± 5.0 years with median of 7.2 and range of 0.5 to 18.9 years ($n = 77$).³ We are now implanting secondary IOLs at relatively younger ages. Most commonly, secondary IOL implantation in our patients was performed between the ages of 2 and 4 years, a period when contact lens compliance can be difficult to achieve, and yet, there is still hope to reverse amblyopia. Due to the relatively slower growth of eyes after this age, IOL power calculations are also more predictable. Many of these children receive secondary IOL implantation before they enter school. Another smaller peak was observed in our group of patients who were 12 to 14 years of age. Most of these were bilaterally aphakic patients who requested IOL implantation for convenience (eliminating contact lens care) or for an improvement on the poor appearance and optical disadvantages of aphakic spectacles. Other series have noted mean ages of 8 years (range 2–16 years),⁴ 10.3 years (1–22 years),⁵ 7.4 years (1.1–15.4 years),⁶ and 2.1 years (1.5–2.5 years).⁷ If sufficient capsular support for bag or sulcus fixation is present,

we now offer secondary IOL implantation soon after we notice that contact lens/spectacle wear is becoming difficult or fails. However, when inadequate capsular support is present for sulcus fixation in a child, implantation of an IOL is not recommended unless every contact lens and spectacle option has been fully explored.

EXAMINATION

A complete ophthalmic examination including visual acuity, slit-lamp biomicroscopy, and fundus examination after mydriasis should be performed. In children who do not permit a reliable office examination, an examination under anesthesia should be performed.

It is important to document central corneal thickness, horizontal corneal diameter, keratometry, globe axial length, intraocular pressure (IOP), and a thorough evaluation of the status of the optic nerve, macula, and retinal periphery. The shape and size of the pupil, any transillumination defects or damage to the iris, the presence of posterior synechia, any vitreous in the anterior chamber, and any other associated ocular anomalies should be documented. Ultrasound biomicroscopy (UBM) may help to detect residual capsular support and image the ciliary sulcus when viewing it directly is difficult (Fig. 31.1).

The technical success of secondary implantation depends mainly on how much capsular support was left behind at the time of primary cataract surgery (Fig. 31.2). Three hundred sixty-degree visibility of the fused edge of the posterior and anterior capsulectomy from the previous surgery increases the chances of achieving successful posterior segment implantation of an IOL. In the office, if the posterior capsule is not initially visible at the slit lamp, having the patient look in extremes of gaze

while maintaining the slit-lamp chin position may allow capsular remnant to be seen under the iris. Sometimes, examination through a peripheral iridectomy (if present) is useful to look for capsular remnants. The presence of posterior synechia is evidence of at least some capsular support since adhesion between iris and capsule is what creates the synechia. There are times, however, when uncertainty about how much capsular support is present remains until surgery when a push-pull instrument or hook (Fig. 31.3) can be used to look directly under the iris in all quadrants.

SITE OF FIXATION

Most aphakic children have had a primary posterior capsulectomy and vitrectomy. It is rare to find an aphakic child with an intact posterior capsule. Aphakic children with residual capsule often have synechia between the anterior and posterior capsule. The surgeon has the option of placing an IOL in the ciliary sulcus after lysing iridocapsular adhesions or attempting capsular bag placement after reopening the capsular bag. If capsular support is available, the IOL should be placed in the posterior segment either in-the-bag or in the ciliary sulcus. The most desirable position for the IOL is within the reopened capsular bag. However, if the capsular leaflets are sealed to one another without reepithelialized cortex (Soemmerring ring), ciliary sulcus fixation is the better choice.⁸ The technique of in-the-bag secondary IOL implantation in children was reported in 1999 by one of us (MEW).⁹ Our updated experience with this technique was published in 2005 and 2011.^{3,10} Grewal and Basti¹¹ reported a modification of the technique, where the authors suggested initiating a new anterior capsulectomy using a

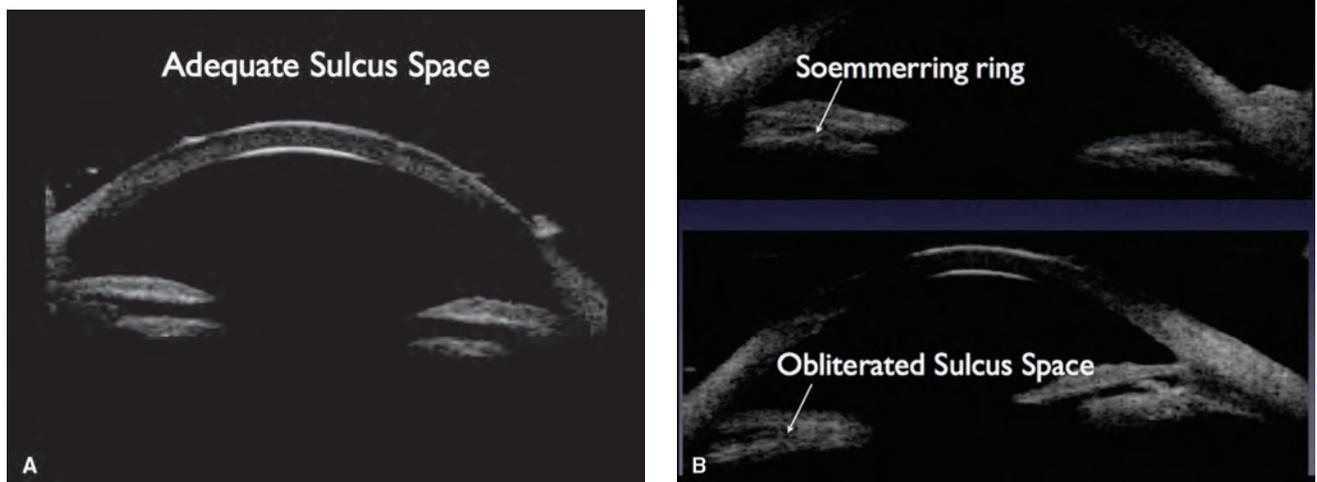


Figure 31.1 A and B: Preoperative UBM. (Courtesy Dr. Abhay R. Vasavada and Dr. Sajani Shah, Ahmedabad, India.)

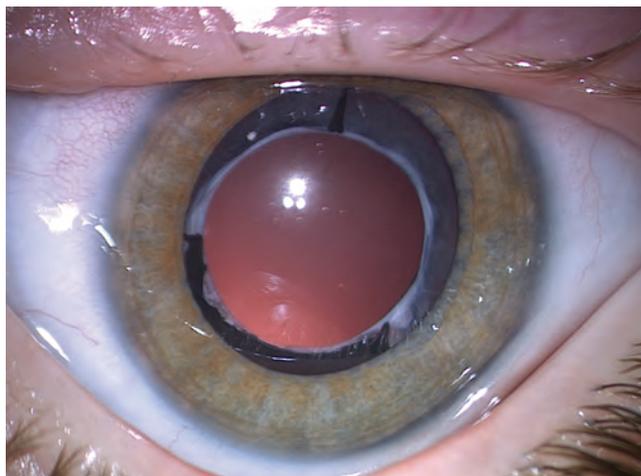


Figure 31.2. Preoperative view of a 5-year-old child scheduled for secondary IOL implantation. Note posterior synechia and pigmented attachment, fused anterior and posterior capsulotomies with clear central opening, and large Soemmering ring.

cystitome to incise the anterior capsule close to its attachment to the posterior capsule. A pair of curved microscissors is used to cut circumferentially, completing the capsulectomy, and a dispersive ophthalmic viscosurgical device (OVD) is used to viscoexpress Soemmering ring material from the capsular bag. A two-handed maneuver was used to manually divide the Soemmering ring. Later, slow-motion phacoemulsification was used to emulsify and remove the pieces. Viscoexpression of the fragments of Soemmering ring was done. The residual capsular bag was filled with OVD, and a three-piece IOL was injected and dialed in. The authors concluded that this technique allows complete evacuation of Soemmering ring and placement of secondary IOL in the bag.¹¹

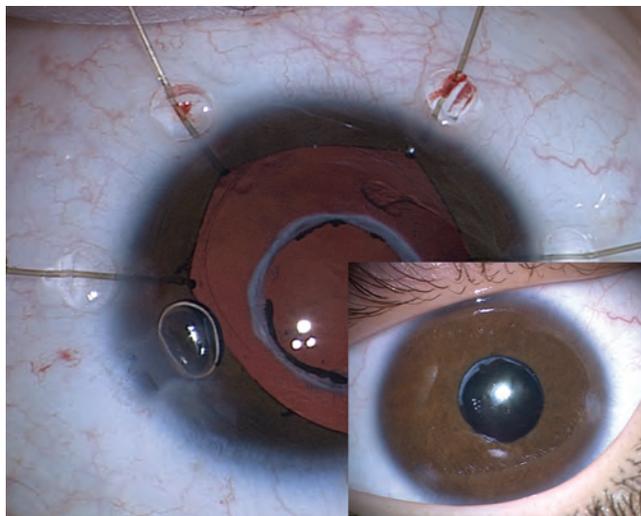


Figure 31.3. Cataract surgery was performed at 4 months of age. The pupil would not dilate well at the time of secondary IOL implantation (**inset**). Use of iris hooks allowed better visualization and opening of space between anterior and posterior capsule.

In-the-bag fixation is more consistently achieved in eyes made primarily aphakic during early infancy. This age at surgery relates to the tendency for a more exuberant Soemmering ring formation in those operated in the first 6 months of life. The Soemmering ring lens cortex fills the capsular bag equator and prevents the anterior and posterior capsule remnants from sealing to one another and closing the capsular bag remnant. Removing the contents of the Soemmering ring and replacing them with OVD allows in-the-bag IOL placement to occur more predictably. An odds ratio analysis revealed that within our study group, eyes originally operated before 6 months of age were 8.7 times more likely to receive a secondary in-the-bag than those of patients who were rendered aphakic after the age of 6 months.³ In a subsequent study, we noticed that all of the eyes that received in-the-bag IOL insertion had a cataract extraction within the first 4 months of life.¹⁰ We hypothesized, as stated above, that young children have much more equatorial epithelial cell proliferation after surgery, even when cortical cleanup is felt to be complete. The few remaining lens epithelial cells are able to produce a dense ring of cortex that becomes trapped between the lens equator and fused anterior and posterior capsulectomy edges; a potential space for in-the-bag placement of an IOL is thus maintained.³ In eyes undergoing cataract surgery later in life, perhaps less dense Soemmering ring leads to fusion of anterior and posterior capsular leaflets. This scarred fibrous fusion may make the anterior and posterior capsule remnants inseparable, and the in-the-bag option may not be feasible.³ At times, these fused leaflets can be viscodissected apart, but attempting this degree of dissection can risk disruption of what would otherwise be a stable platform for ciliary sulcus IOL fixation.

Ciliary sulcus fixation is the best second alternative site for fixation of the IOL in the presence of available capsular support.^{3,5,8-10,12,13} Several case series have been reported, indicating that a lens placed in the ciliary sulcus is reasonably safe and effective over the short term. Awad et al.⁴ performed ultrabiomicroscopy on the ciliary sulcus in 10 eyes after secondary IOL implantation. No gross haptic erosion into the sclera, ciliary body, or sulcus was seen. The structure of the sulcus in the implanted eye appeared similar to the sulcus in the contralateral normal eye. Ciliary sulcus–fixated IOLs are more prone to develop pupillary capture, pigment dispersion, ciliary body erosion, decentration, unstable loop fixation, and lens tilt than IOL with bag fixation.¹⁴ A study of adult eyes reported that even more than 2 years after cataract surgery, anterior chamber flare counts in eyes with sulcus-to-sulcus IOL fixation were significantly higher than in eyes with in-the-bag fixation.¹⁵ Vasavada et al.¹⁶ evaluated the anatomic location of the haptic and optic of single-piece sulcus-fixated IOLs and their proximity to the iris and ciliary body using UBM. Both IOL haptics were in the sulcus in 7/10 eyes, 1 haptic was abutting the

iris and the other was in the sulcus in 2 eyes, and both haptics were embedded in the ciliary body in 1 eye. These authors concluded that after intended sulcus implantation of a single-piece nonangulated acrylic IOL, both haptics were not always actually in the ciliary sulcus. In the studied eyes, the haptic and optic remained in close proximity to the posterior iris surface. The IOL utilized in the study has a square-edged optic and haptics and no posterior angulation. Alcon has not recommended implanting single-piece AcrySof® IOL in the sulcus because of the unique configuration of the haptics, optic, and a zero angulation of the haptic–optic junction.¹⁷ Micheli et al.¹⁷ noted that surgeons should avoid sulcus placement of the single-piece AcrySof® because of the lens' potential to induce haptic-related pigmentary glaucoma. In contrast, the three-piece AcrySof® IOL is posterior angulated and does not share the same haptic configuration as the single-piece AcrySof®. The three-piece AcrySof® is appropriate for sulcus fixation as well as in-the-bag positioning. It does not have a propensity to cause pigment dispersion when placed in the ciliary sulcus.

Surgeons need not avoid implanting all nonangulated single-piece acrylic IOLs in the ciliary sulcus. For instance, we have found that the Rayner *C-flex* (570C) IOL is well tolerated by pediatric eyes when it is placed in either the ciliary sulcus or the capsular bag. It is not tacky on its surface like the single-piece AcrySof®, and it does not have the same shaped haptics as the AcrySof®. We have not observed pigment dispersion or chronic inflammation in eyes implanted with this lens. The Rayacryl hydrophilic acrylic material has high biocompatibility and excellent handling characteristics. We have also found that it remains centered in the sulcus better than the three-piece AcrySof® in large eyes. Optic capture through fused anterior and posterior capsule and haptics in the ciliary sulcus is another viable option for keeping the three-piece AcrySof® IOL centered and stable. At the time of this writing, the Rayner *C-flex* (570C) is our preferred IOL for sulcus placement. In the United States, we do not have access to the Rayner Sulcoflex IOL, which has been designed specifically for sulcus placement as a supplementary piggyback IOL.

In our series, eyes operated for cataracts before 1991 were more likely to have an absence of adequate capsular support. More of these eyes required scleral-fixated or anterior chamber IOLs. After 1991, enough capsular support was more routinely left at the time of primary surgery, in anticipation of IOL implantation later. In the absence of sufficient lens capsule support, choosing the best site for the IOL is more difficult and controversial. Sutured posterior chamber IOLs, an open-loop flexible anterior chamber intraocular lens (ACIOL) or the Artisan (Ophtec) iris claw lens are available options. In the absence of capsular support and the presence of disrupted iris, sutured transscleral posterior chamber IOL is the only viable option. Various options and outcomes have been described in Chapter 32.

IOL POWER CALCULATION

While secondary IOL power can be estimated using the aphakic refraction,^{18,19} the estimate is less accurate than biometry.²⁰ If biometry cannot be obtained in the clinic due to poor cooperation and if biometry is not available in the operating room, aphakic refraction can be used to estimate the IOL power. Change of fixation site may require adjusting the IOL power. If decision is made to place the IOL in the ciliary sulcus, rather than the capsular bag, a decrease in lens power is often necessary. This is due to the fact that as the optic is shifted more anterior (moved closer to the cornea), its “effective power” increases. The amount of this change is dependent on the “base power” of the IOL. The greater the power, the greater the difference. If IOL power calculated for bag needs to be changed to sulcus, subtract 1.5 D for ≥ 28.5 D, 1 D for 28.0 to 17.5 D, and 0.5 D for 17.0 to 9.5 D. No subtraction is required if power at capsular bag is < 9 D (<http://www.doctor-hill.com/iol-main/bag-sulcus.htm>).

COMBINED STRABISMUS SURGERY

Simultaneous eye muscle and IOL implantation surgery in patients with strabismus and aphakia has been reported.²¹ Combining IOL implantation with an extraocular muscle procedure may reduce the number of surgical and anesthetic procedures, speed rehabilitation, and offer financial benefit to the patient and third-party payers. However, concerns may arise regarding a potential increased risk of postoperative infection, anterior segment ischemia, or excessive discomfort for the patient. Determining the ocular alignment in eyes with poor vision may be difficult.²¹ We typically offer secondary IOL implantation surgery first (if indicated), wait for vision to recover, start amblyopia treatment, evaluate for strabismus, and offer strabismus surgery as a separate procedure.

OUTCOME OF SECONDARY IOL IMPLANTATION

Anatomical Outcome

Peaked Pupil

A peaked pupil may rarely occur after secondary IOL implantation. This is usually due to a vitreous strand and should be repaired during relatively early postoperative period (Figs. 31.4 and 31.5).

Visual Axis Opacification

In our series, visual axis opacification (VAO) was noted in 4 (5.2%) eyes.³ Crnic et al. reported VAO in 9% of their sulcus positioned IOLs. In our 2011 series, 2 eyes (12.5%) implanted in the capsular bag developed VAO, whereas none of the eyes implanted in the ciliary sulcus developed VAO.¹⁰ Opening the capsular bag space may allow for the escape of residual cells with proliferative capabilities, causing the later

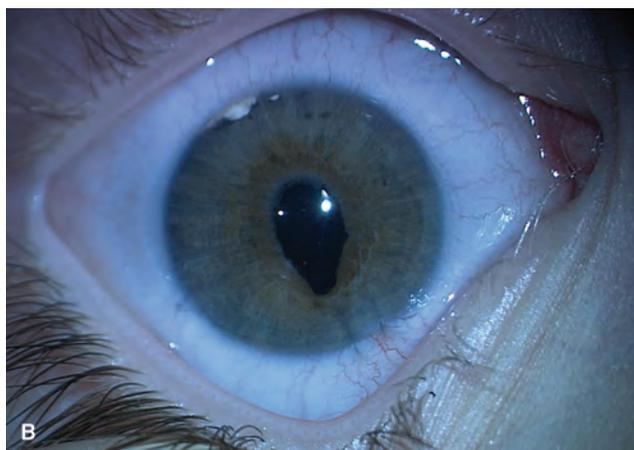
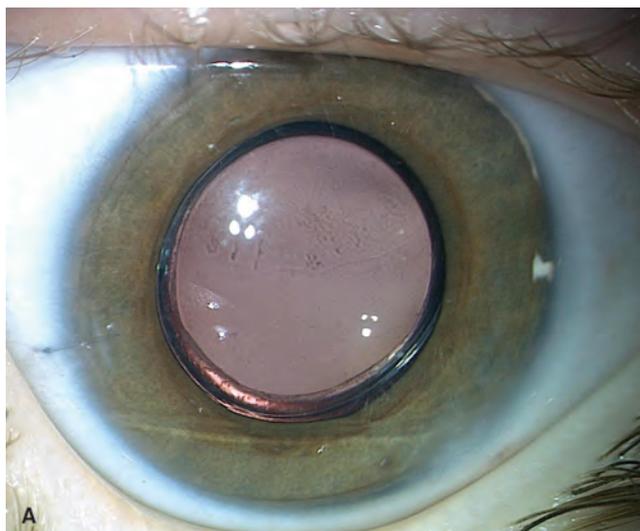


Figure 31.4. **A:** Sulcus-fixated Rayner C-flex secondary IOL implantation in the left eye of a 4-year-old boy. **B:** Vitreous strand noted at 2 weeks after surgery. The photograph was taken 1 month after secondary IOL implantation when surgery was performed to remove vitreous strand. Vitreous strand appeared to be attached to the internal lip of the tunnel incision.

development of VAO. Theoretically, opening the bag gives the cortical material in the Soemmering ring access to the eye with a possible higher risk of postoperative inflammation; however, we have not observed it in our series (Fig. 31.6).¹⁰

IOP Spike

Biglan et al.⁵ noted that two eyes had elevated IOP following secondary implant. One was transient and was considered to be related to use of OVD.⁵ We report that eyes with preexisting glaucoma were more likely to develop IOP spike during early postoperative period.²² We recommend topical antiglaucoma medication or Diamox for subjects undergoing secondary IOL implantation with preoperative diagnosis of aphakic glaucoma.

Glaucoma

Biglan et al.⁵ noted that one eye developed glaucoma (in addition to one mentioned above with IOP spike). We

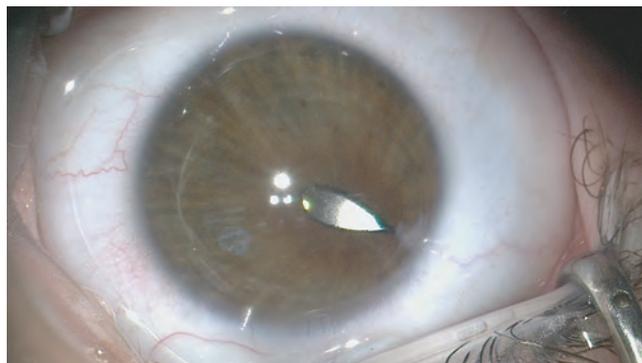


Figure 31.5. Peaked pupil noted after 8 months of secondary in-the-bag IOL implantation. Note that the child was examined in the clinic at 1 month and 3 months after surgery; however, peaked pupil was documented at 8-month postoperative visit. Synechiolysis was performed.

noted postoperative glaucoma in 1 (6.3%) bag-fixated eye and 1 (6.7%) eye in the sulcus fixation group.¹⁰

Decentration and Dislocation of IOL

In our series, complications included clinically significant decentration, 4 (5.2%) eyes, and dislocation of the IOL, 2 (2.6%) eyes; and pupillary capture requiring repositioning of IOL, 1 (1.3%) eye. *Decentration of the IOL* was the most common complication (Figs. 31.7 and 31.8). Clinically significant decentration requiring surgical intervention was noted only in eyes with sulcus-fixated foldable IOLs (28.6%; 4/14). None of the 29 eyes with sulcus-fixated polymethyl methacrylate (PMMA) IOL implantation developed decentration. Perhaps the rigidity of PMMA IOLs helped to avoid decentration. Foldable

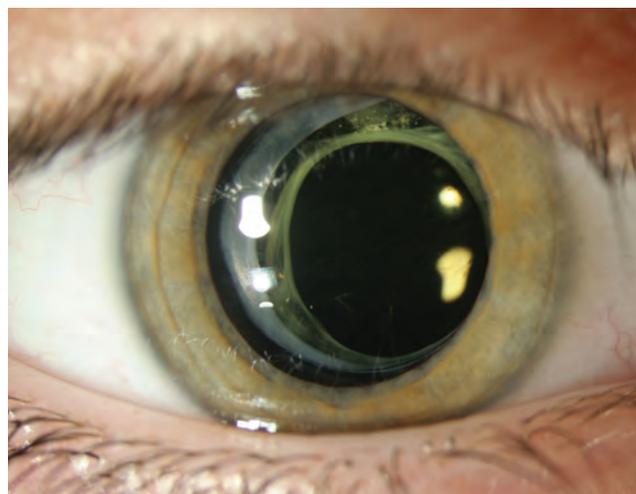


Figure 31.6. Six-year follow-up of secondary SN60 IOL in the bag. Cataract surgery was performed at 2 weeks of age, and secondary IOL was implanted at 4 years of age.

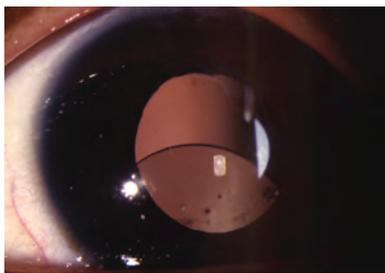


Figure 31.7. Decentration of AcrySof® IOL 3 weeks postoperatively in a 13.8-year-old male.

IOLs in the sulcus have been noted to be at risk for decentration and dislocation.²³ Decentration/dislocation was responsible for 21% of explanted three-piece hydrophobic acrylic IOLs in a 2001 survey.²⁴ All the decentrations were in an inferior direction and occurred in eyes of male patients.³ Perhaps this is due to the higher incidence of trauma among males. Another possible reason is that male eyes have been noted to have a longer axial length than do female eyes. Eyes with an axial length of >23 mm were four times more likely to develop decentration if implanted with a sulcus-fixated foldable IOL when compared with eyes measuring <23 mm. We hypothesized that longer eyes may also have a “wider” anterior segment. A wider sulcus-to-sulcus distance may promote IOL decentration.³ Another recent study has reported decentration in 6% of eyes after secondary placement of foldable AcrySof® IOLs in the ciliary sulcus.⁶ Jacobi et al.²⁵ noted decentration of a scleral fixation of a secondary foldable monofocal or multifocal IOL implant in 19.2% of eyes of children and young adults. We have now begun to use the Rayner C-flex (570C) IOL in the ciliary sulcus more often than the AcrySof® MA-60 IOL, especially in eyes longer than 23 mm in globe axial length. Hopefully, this change will result in a lower incidence of IOL decentration when sulcus implantation of the IOL is required.

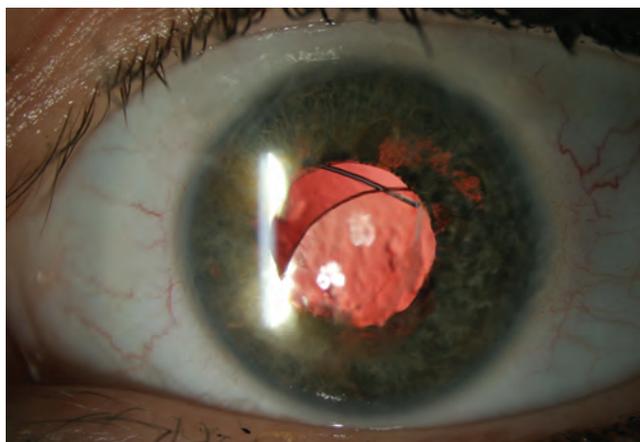


Figure 31.8. Decentration of secondary sulcus-fixated three-piece AcrySof® MA60 IOL at 7-year follow-up visit. IOL in this eye was replaced with PMMA MC60 IOL in the sulcus as patient was complaining of double vision.

Uveitis

Bilateral uveitis (possibly sympathetic ophthalmia) resulting in blindness in a child following secondary iris supported IOL implantation following extraction of a unilateral congenital cataract has been reported in 1991.²⁶ In September 1984, the patient underwent an aspiration of a cataract in the right eye. In April 1985, a secondary IOL was implanted (Binkhorst iris clip). Prior to each of these procedures, the fellow eye was completely healthy. Eight months later, the child developed a bilateral plastic uveitis which, in spite of intensive treatment, resulted in no perception of light in the implanted eye and 2/60 vision in the fellow eye. The authors concluded that sympathetic ophthalmia possibly played a role in the pathogenesis of the uveitis.

Cystoid Macular Edema

A higher incidence of *cystoid macular edema* with secondary IOL implantation in adults in whom vitreous loss occurred at the time of initial cataract surgery has been reported. Other studies, however, reported excellent results when a careful and controlled vitrectomy was performed with secondary IOL implantation. Although we did not perform angiography, we did not observe clinically significant cystoid macular edema after secondary implantation in our published series of pediatric eyes.³

Endophthalmitis

Some reports in the adult literature have found a higher incidence of *endophthalmitis* in secondary IOL patients compared to primary cataract surgery with IOL placement.²⁷ Kattan et al.²⁷ noted an incidence of culture-proven endophthalmitis as 0.07% (17/23, 625) after cataract surgery with or without IOL implantation and 0.3% (3/988) after secondary IOL implantation. We cared for an older child who underwent bilateral secondary IOL implantation and developed unilateral endophthalmitis after swimming in canal water on the second postoperative day after his second eye was operated (and 1 month after his first eye had been operated).¹⁰

Visual Outcome

In 1996, Biglan et al.⁵ noted that 20 of 28 (71.4%) eyes had a measurable improvement in visual acuity when a secondary IOL was implanted. One eye had a decrease in visual acuity of two lines. DeVaro et al.⁸ reported, in 1997, that visual acuity of 20/40 or better was achieved with secondary IOL implantation and overrefraction in three of 11 (27%) eyes operated for nontraumatic cataract. A 1998 series reported that 42% of patients implanted with an IOL secondarily had a best-corrected visual acuity of 20/40 or better and 78% better than 20/80.⁴ In 2005, we reported the results of 77 eyes implanted with a secondary IOL at an average age of 7.8 ± 5.0 years with an average follow-up of 2.7 ± 1.9 years (minimum follow-up of 6 months). The postoperative geometric mean visual

acuity was significantly better than the preoperative visual acuity ($P < 0.001$). Best-corrected visual acuity improved after implantation in 72%, remained unchanged in 10%, and decreased in 18%. The nine eyes with one or two lines of decreased vision at last follow-up were analyzed more closely. Four eyes had glaucoma, one eye was from a Down syndrome patient known to have variable coopera-

tion for visual acuity testing, and the other four eyes had amblyopia. It is uncertain in these four eyes if the amblyopia worsened or if the surgery itself was responsible for the vision loss. No patient lost more than two lines of visual acuity.³ In a 2011 series of bag- and sulcus-fixated secondary IOL surgeries, we reported an overall median visual acuity after implantation of 20/40.¹⁰

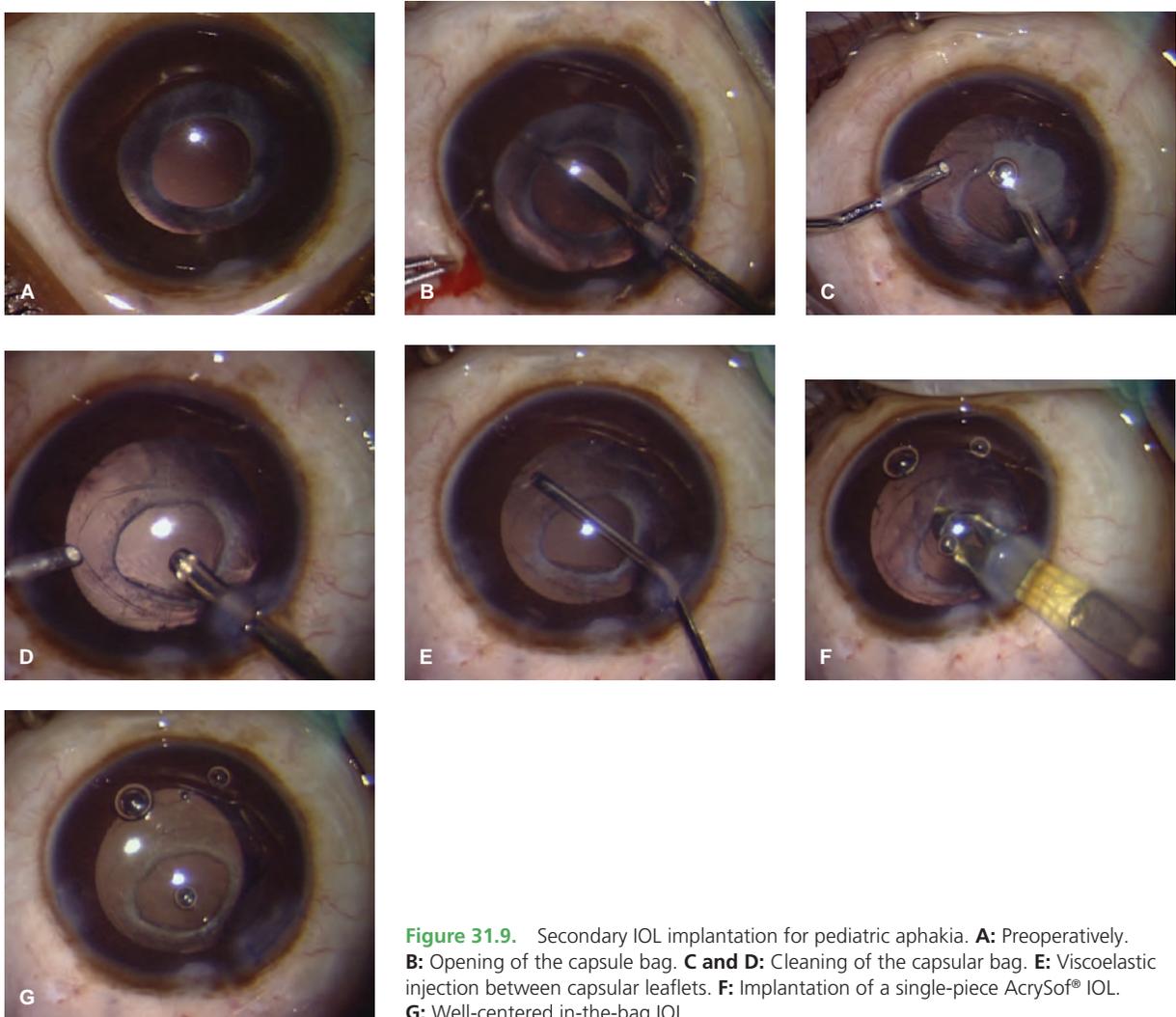


Figure 31.9. Secondary IOL implantation for pediatric aphakia. **A:** Preoperatively. **B:** Opening of the capsule bag. **C and D:** Cleaning of the capsular bag. **E:** Viscoelastic injection between capsular leaflets. **F:** Implantation of a single-piece AcrySof® IOL. **G:** Well-centered in-the-bag IOL.

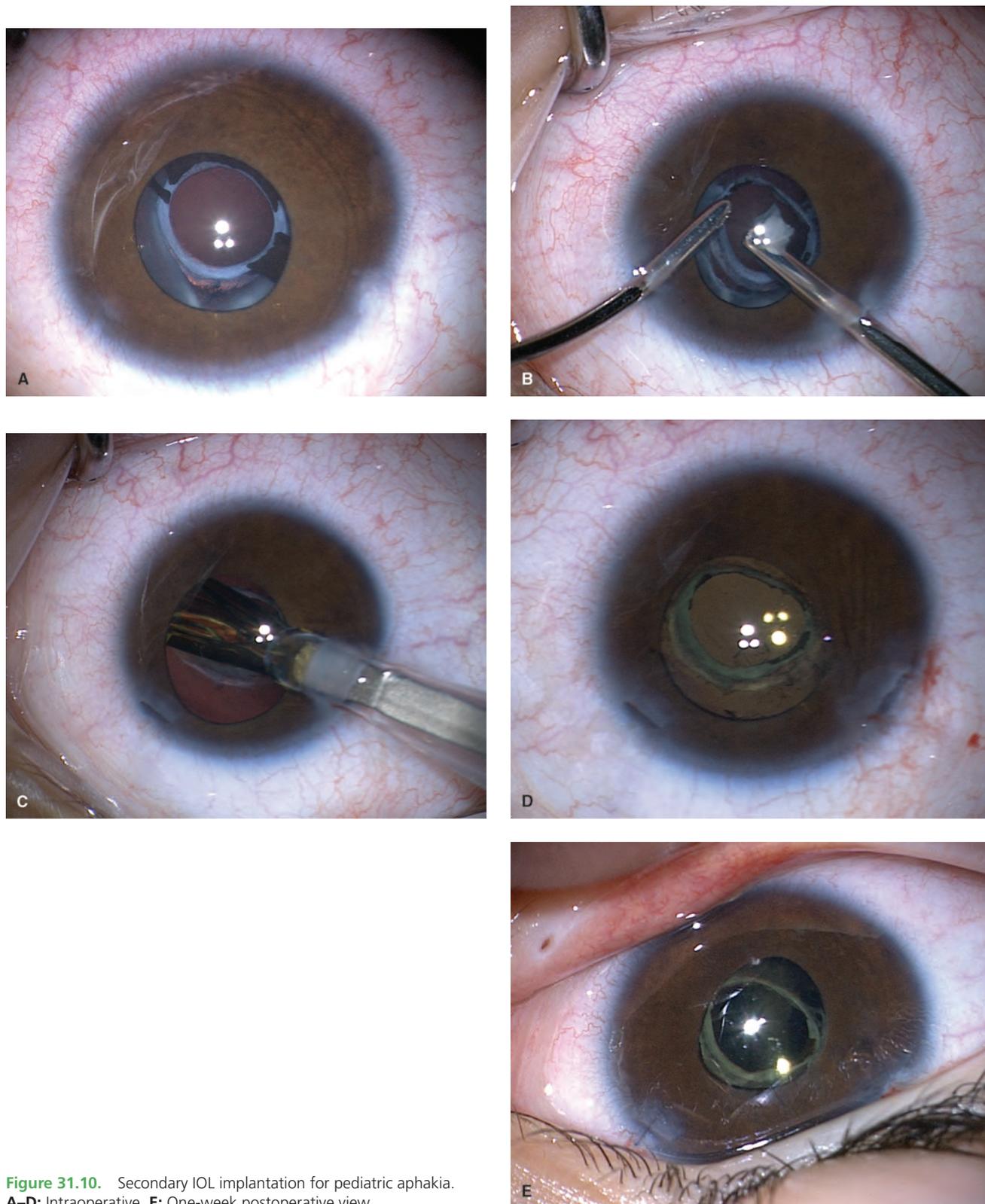


Figure 31.10. Secondary IOL implantation for pediatric aphakia. **A–D:** Intraoperative. **E:** One-week postoperative view.

SUMMARY

- When performing infantile cataract surgery without primary IOL implantation, leave an adequate capsular rim for subsequent IOL placement. A 4.5-mm central posterior and anterior capsulectomy is usually adequate to prevent opacification of the visual axis but assures an adequate rim of support when secondary IOL implantation is elected.
- In a patient with bilateral aphakia, implant the eye with the worst capsular support first. If it is not feasible to safely achieve implantation, bilateral aphakia can still be chosen. This approach can help avoid an IOL in one eye and aphakia in the other.
- While corneal tunnel incisions are usually utilized for secondary IOL implantation, a scleral tunnel incision should be considered when the capsular support is limited. After the posterior synechia are severed, a change to a PMMA IOL may be warranted to provide a stable bridge across a large posterior capsule opening. This change can be accomplished more easily from a scleral incision.
- Change of fixation site may require adjusting the IOL power. Note that higher IOL powers require higher adjustments.
- Assess whether it is possible to reopen the capsular bag leaflets. Figures 31.9 and 31.10 describe the surgical steps for in-the-bag secondary IOL implantation. The key is to locate one area in which the anterior capsule edge is not strongly adherent to the posterior capsule.²⁸ Using the entry point, viscoelastic agents can be very useful in the separation of the capsular layers. A combination of dissection techniques using intraocular scissors, a microvitrectomy knife, and the vitrector handpiece are often used to create a new anterior capsule edge and reopen the capsular bag for 360 degrees. Remove the Soemmering ring by bimanual irrigation/aspiration and place a secondary IOL into the capsular bag whenever possible.
- When the ciliary sulcus is the intended placement site for a secondary IOL, at present we use a Rayner C-flex (570C) (Rayner IOL Ltd, Hove, East Sussex, UK) IOL or a three-piece AcrySof[®] MA-60.
- In the absence of available capsular support, sutured IOLs, iris claw lenses, or ACIOLs can be used, depending on the surgeon's preference, the ocular environment, and IOL availability (see Chapter 32). Each of these options requires careful follow-up over an extended period of time.

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Intraocular Lens Implantation in the Absence of Capsular Support

M. Edward Wilson, Muralidhar Ramappa, and Rupal H. Trivedi

When planning an intraocular lens (IOL) implantation in the absence of capsular support, key questions immediately come to mind. *Are anterior chamber (AC) IOLs (angle-supported or iris claw), or posterior chamber IOLs (scleral or iris sutured or "glued") safe and effective in children's eyes with inadequate capsular support for in the capsular bag or ciliary sulcus fixation of IOLs? Is there an optimal choice of IOL design or material, method, and site of fixation in eyes with inadequate capsular support?*

In this chapter, we explore these controversial issues and provide information that will help the pediatric cataract surgeon make decisions that provide the best possible visual outcome. By their very nature, pediatric surgeons are conservative while still being innovative. Risk must be minimized, but amblyopia must not be left untreated or undertreated. Nowhere is that balance more difficult than when there is inadequate capsular support in the face of contact lens and aphakic spectacle intolerance.

Some key points from the discussion that follows are included in Table 32.1.

BACKGROUND

IOL implantation in the absence of adequate capsular support is a challenging situation to manage, particularly in a pediatric eye. The long-term implications of these surgical interventions, when performed on children, are not well documented, and the optimal lens placement techniques for the pediatric eye are uncertain. Poor capsular support is often seen in pediatric aphakic eye, particularly after removal of congenital cataracts, traumatic cataract, or ectopia lentis. After extensive removal of the lens capsule, either by a surgeon at the primary lensectomy or at subsequent membranectomy in cases of reocclusion of the visual axis, there may not be enough of a

capsular shelf to safely implant a secondary IOL in the ciliary sulcus. Also, in cases where extensive adhesion between capsular remnants and uveal tissues are broad and fibrotic, reestablishment of the ciliary sulcus may not be possible. In eyes with sufficient capsular rim, a posterior chamber IOL (PCIOL) can be safely implanted in the ciliary sulcus or in the re-opened capsular bag remnant. However, in eyes with poor capsular support, the possible locations to fixate the IOL include angle-supported anterior chamber intraocular lens (ACIOL), iris claw IOL, or transscleral or iris-fixated PCIOL. In eyes with either a partially subluxated or dislocated IOL, it can be repositioned by securing it to the sclera or iris in a minimally invasive or less traumatic procedure than IOL exchange. However, IOL fixation elsewhere poses several intraoperative and postoperative surgical challenges. ACIOL implantation is contraindicated in the presence of extensive damage to the iris and AC angle, preexisting glaucoma, peripheral anterior synechia (PAS), low endothelial cell count, and shallow AC.¹ Iris claw IOLs, which are lenses placed in the AC and attached to the iris with small claw-like haptics, have been described to correct aphakia in adults,² and children,³ with good visual results. These lenses have been available in most parts of the world for several years and are now being implanted in 20 sites in the United States under a compassionate-use FDA investigational device exemption. PCIOL offers several advantages, and many authors recommend them even in eyes lacking capsular or zonular support.⁴ Scleral fixation of PCIOL is a method to overcome the lack of capsular support but is technically more difficult and time consuming compared to ACIOL implantation. Due to its anatomic location in the eye, PCIOL is more appropriate for patients with glaucoma, low endothelial cell counts, PAS, and cystoid macular edema (CME).⁵

Table 32.1 KEY POINTS IN THE DISCUSSION OF IOL IMPLANTATION WHEN THERE IS AN ABSENCE OF CAPSULAR SUPPORT

- Surgical aphakia in children is extremely difficult to handle; timely refractive correction is necessary in this vulnerable population to preserve monocular vision and binocularity and to protect against stimulus deprivation amblyopia
- Conservative management may include correction with glasses or contact lenses, although these measures are not equally effective in all patients. Aphakic spectacles are an option for bilateral aphakic children but often undesirable for social and optical reasons
- The possible locations to fixate the IOL in the absence of adequate capsular support include angle-supported AC, iris- (sutured or claw fixation) supported AC, or scleral (transscleral sutured or externalized haptics with glued flap)
- Angle-supported ACIOL implantation has been linked in past literature with higher risks of iritis, pigment dispersion, corectopia, glaucoma escalation, and corneal endothelial attrition leading to corneal decompensation
- A review of angle-supported ACIOLs and transscleral-sutured IOLs by the American Academy of Ophthalmology concluded that there was insufficient evidence to prove superiority of one lens type or fixation site.

Suitable candidates for IOL fixation elsewhere include those aphakic children who meet the following criteria: (1) unilateral or bilateral aphakia in children intolerant of or not suitable for contact lens wear; (2) extensive capsular deficiency precluding safety of stable in the bag or sulcus placement of an IOL; (3) absence of uncontrolled glaucoma, uveitis, or endothelial dysfunction; (4) absence of retinal hole/fluid, extensive lattice degeneration, or major retinal disease; and (5) motivated child's parent(s), who acknowledges the risks/alternatives and the importance of follow-up examinations. Comprehensive preoperative assessments are needed, including examination under general anesthesia if complete office examination was not possible. This assessment includes age-appropriate testing of corrected distant visual acuity, refraction, sensorimotor examination of eye alignment/eye movement, measurement of intraocular pressure (IOP), slit-lamp examination, gonioscopy, and funduscopy.

TRANSSCLERAL-SUTURED IOL FIXATION

Transscleral-sutured IOL fixation can be considered in cases with inadequate capsular support or in cases with compromised AC structure. The ciliary sulcus is located 0.83 mm posterior to the surgical limbus in the vertical meridian, whereas in the horizontal, it is 0.46 mm from surgical limbus.^{6,7} Duffey et al.⁸ passed the needle perpendicular to the sclera 1, 2, and 3 mm posterior to the surgical limbus and found that the needle exited internally at the ciliary sulcus, pars plicata, and pars plana, respectively. Scleral suturing of a PCIOL requires that the needle pass through vascular uveal tissue, with the attendant risk of bleeding. In many cases, this is minor and resolves spontaneously.⁹ Keeping the suture anteriorly (0.5–1 mm behind the surgical limbus) and avoiding the 3 o'clock and 9 o'clock positions may reduce the risk of bleeding by avoiding vascular ciliary body tissue and the long posterior ciliary arteries.¹⁰ In addition, taking care to maintain a pressurized globe and to minimize tissues traversed

during transscleral needle passes will reduce the risk of intraoperative intraocular hemorrhage.¹¹

Scleral fixation too close to either iris or pars plicata can have long-term protracted complications. Scleral fixation can be performed by either *ab externo* (passing needle from outside to inside) or *ab interno* approach (inside to out). The *ab externo* approach is most preferred. Adequate anterior vitrectomy is mandatory before placement of sutures and the IOL to avoid complications related to the vitreous base.

Surgical Technique

Ab Interno Approach

In this technique, two partial-thickness scleral flaps are made, 180 degrees apart. A right-handed surgeon should create the superior flap toward 1 o'clock and the inferior flap toward 7 o'clock for ease of maneuverability. The Ethicon CIF-4 long tapered needle can be used to pass a polypropylene suture through the inferior scleral flap, and the Ethicon TG 160-6 small curved needle can be used to pass a polypropylene suture through the superior scleral flap. The sutures can be looped through the eyelets of the haptics or tied to the haptic, or a girth hitch suture can be used. These sutures can be used for two-point or four-point fixation of the IOL. After passing out the polypropylene sutures through the scleral flaps, the ends of the sutures are tied in a 3-1-1 knot. The ends of the knot are cut 1 mm long. The scleral flaps are tied with 6-0 Vicryl. The corneoscleral wound is closed with 10-0 monofilament nylon or Vicryl, and the overlying conjunctival wound is closed with 8-0 Vicryl sutures or sealed with fibrin glue.

Ab Externo Approach

By this approach, the IOL can be placed more accurately in the ciliary sulcus. After designing the partial-thickness scleral flaps, the long straight needle is passed approximately 1 mm from the surgical limbus through one side; on the other side, a 27-gauge hypodermic needle should be passed. The straight needle is then negotiated through the hollow hypodermic needle. The hypodermic needle should be withdrawn with the straight solid needle inside it. The AC is

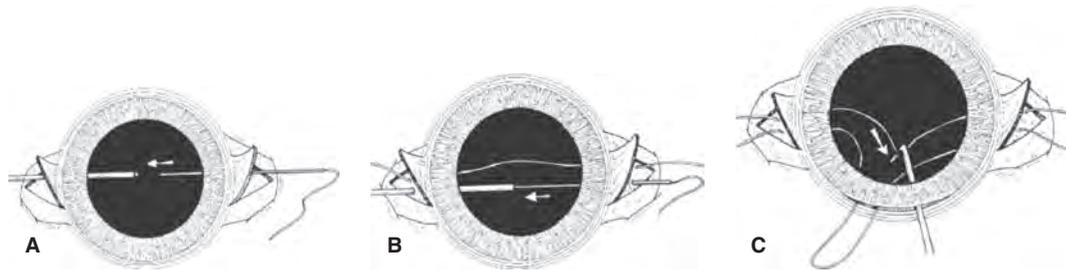


Figure 32.1. **A:** The double-suture variant of the Lewis ab externo technique begins similarly to the single-suture technique, except that the suture entry point under the scleral flap is displaced to one side. **B:** The second suture is passed parallel to the first, with 1 to 1.5 mm between the two sutures. **C:** Care must be taken to keep the sutures taut, to avoid crossing them or confusing which suture originates from each sclera site, while a Kuglen hook or similar instrument withdraws the suture loop through the previously prepared principal incision. (Reprinted from Steinert RF, Arkin MS. Secondary intraocular lenses. In: Stienert RF, ed. *Cataract Surgery: Techniques, Complications, Management*. Philadelphia, PA: Saunders, 2004:429–441, with permission.)

entered, and the suture is taken out with a hook. The suture is cut, and the end is tied to the eyelet of the IOL haptic. If the surgeon desires four-point fixation of the IOL, the above steps are repeated (Figs. 32.1 and 32.2). It should be noted that the main advantage of the ab externo approach is its greater precision in the location of scleral sutures.

The polypropylene suture knot should not be kept exposed, with attendant risk of suture-related irritation or suture-wick endophthalmitis. Scleral flaps are recommended to cover it. If possible, the knot should be buried in the deep in the scleral pocket. Alternatively, a four-point fixation technique can be achieved where a loop of suture is created and the knot can be rotated into the sclera, thus avoiding the need for a scleral flap (Fig. 32.3). Another technique, known as a Hoffman pocket, creates a scleral pocket dissected from a corneal groove outward to the sclera. This is discussed more fully below. Some surgeons prefer leaving the polypropylene suture ends long and tucking them under the conjunctiva toward the fornix. It was believed that short ends of suture tend to extrude, whereas long ends tend to lie down against the sclera and are well tolerated. We have seen exposure and one infection from this tucking of the long suture ends method, and we no longer advocate it. The PCIOL used for scleral fixation should ideally have large diameter

optics (6.5–7 mm), and haptics should ideally have large and well-polished eyelets.

Complications After Transscleral-Sutured IOLs

Suture Breakage

A disturbing late complication of transscleral fixation of IOLs is the spontaneous breakage of the polypropylene suture leading to IOL displacement, especially in young patients.^{12,13} Vote et al.¹² reported late breakage of polypropylene sutures in 16 of 61 eyes (26.2%) of mostly adult patients occurring about 50 ± 28 months after IOL fixation. Walter et al.¹⁴ had only 2.2% suture breakage in 89 eyes of adult patients during a mean follow-up of 24 months after penetrating keratoplasty and scleral-fixated PCIOL (SF-PCIOL) implantation. Buckley¹⁵ reviewed the literature on transscleral-sutured IOLs in children. He reported outcomes of 33 eyes with an average follow-up of 61 months (range 9–200 months). Twenty one of the eyes had >3 years of follow-up and 14 eyes (42%) had more than 5 years of follow-up. Four patients had spontaneous suture breakage at 38, 66, 96, and 107 months after implantation. An additional 13 cases of 10-0 Prolene suture breakage in children were uncovered by a survey of pediatric ophthalmologists. As

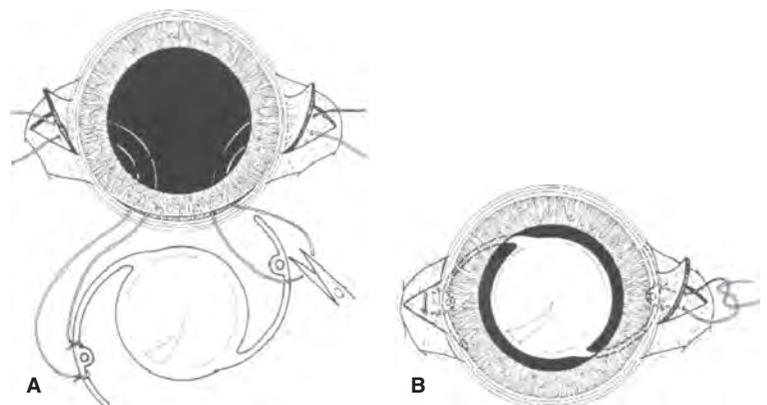


Figure 32.2. **A:** To achieve four-point stable fixation, the cut sutures are tied to the haptic on either side of the eyelet. **B:** The IOL is placed in the posterior chamber, keeping the sutures taut to avoid enlargement. The ends are then tied under the scleral flaps, and the conjunctiva is closed over the flaps. (Reprinted from Steinert RF, Arkin MS. Secondary intraocular lenses. In: Stienert RF, ed. *Cataract Surgery: Techniques, Complications, Management*. Philadelphia, PA: Saunders, 2004:429–441, with permission.)

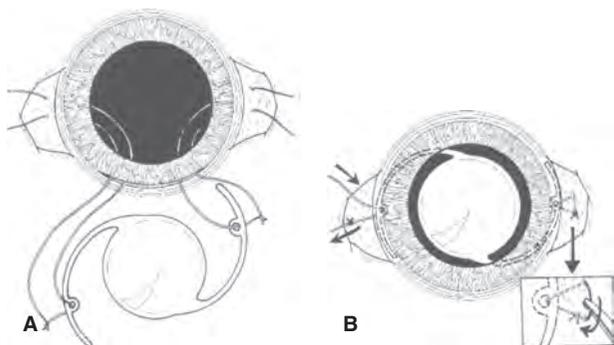


Figure 32.3. In this variant of the double-suture ab externo technique, the goal is to achieve a loop of suture where the knot can be rotated beneath the sclera, avoiding the necessity for a sclera flap. **A:** Cut ends of each suture are passed through the haptic positioning hole and tied. **B:** As the IOL is positioned in the posterior chamber, one end of the suture on each side is pulled, so it is cut off. The remaining suture ends are then tied together, and the knot is rotated beneath the sclera (**inset**), achieving the same result as illustrated in Figure 32.2. (Reprinted from Steinert RF, Arkin MS. Secondary intraocular lenses. In: Stienert RF, ed. *Cataract Surgery: Techniques, Complications, Management*. Philadelphia, PA: Saunders, 2004:429–441, with permission.)

a conclusion of his Costenbader lecture, Buckley¹⁵ urged caution in the use of 10-0 polypropylene suture to fixate an IOL to the sclera in children. He recommended using an alternative material or an alternative size (9-0 polypropylene instead of 10-0). Appropriate ophthalmic needles are now available on the 9-0 Prolene suture. When necessary, however, the 9-0 Prolene suture can be tied to the end of a 10-0 Prolene and pulled through the eye using the 10-0 Prolene needle.

Long-term IOL stability depends on ongoing suture fixation, sulcus positioning, and residual capsule, if present.¹⁶ In addition to slow biodegradation of the suture, there are other factors that may contribute to the higher probability of suture breakage in children. These include globe enlargement with age, continuous eye rubbing, and higher probability of eye trauma due to more active lifestyle. To reduce the chance of suture breakage, it is important to consider the use of multiple sutures on each haptic, thicker sutures¹⁷ (such as the 9-0 polypropylene mentioned above) or a different suture material such as GORE-TEX (W. L. Gore & Associates, Newark, DE).

Other Complications

Postoperatively, mild vitreous hemorrhage is a commonly encountered complication following scleral-fixed IOL, but it is likely to spontaneously resolve with no subsequent sequelae. As discussed earlier, by avoiding 3 o'clock and 9 o'clock positions and, in addition, by tamponading internally, this complication could be minimized. Late endophthalmitis after SF-PCIOL implantation has been reported in the literature and remains a rare but sight-threatening complication after this procedure.^{18–22} Parents must, therefore, be warned about the signs and symptoms of this complication. It seems that in cases

of late endophthalmitis after IOL fixation, the infecting organism gains access to the eye via exposed sutures. Although the scleral flaps reduce the risk of suture exposure, in the long term, the suture ends can erode through partial-thickness scleral flaps and conjunctiva. This illustrates the importance of avoiding exposed suture ends. Methods of preventing this may include leaving the suture ends long, rotating knots into the sclera, or tying the knot in the depths of a partial-thickness scleral incision.¹¹ A unique and innovative technique for burying the suture knots is by using a scleral pocket initiated through a clear corneal incision dissected posteriorly (away from the center of the cornea) into the sclera. The technique is known as a Hoffman pocket,²³ and it avoids the need for conjunctival dissection, scleral cauterization, or suture closure of the flap over the suture knots. To begin, two partial-thickness (300–400 μm) 1–clock-hour groove incisions are made 180 degrees apart in clear cornea just anterior to the conjunctival insertion at the limbus. Two scleral pocket incisions are then dissected approximately 3 mm posterior from the clear corneal incision. A paracentesis is made adjacent to each of the corneal incisions. A 27-gauge needle is passed ab externo through the conjunctiva and the full thickness of the scleral pocket. The 9-0 Prolene is passed through the opposing paracentesis across the AC to dock into the barrel of the 27-gauge needle. The needle is withdrawn and the Prolene with it. A Sinsky hook can then be used to retrieve the suture by sweeping into the pocket from the corneal side. This maneuver is repeated to create four-point fixation of the IOL as shown in Figures 32.1 through 32.3. The major difference from the figure is that with the Hoffman pocket, the knot slides under the roof of the scleral pocket as it is tied. No scleral flap closure is needed, and no conjunctival dissection and closure have been necessary. The Prolene knots are well covered and less likely to erode over time.

Retinal detachment (RD) has been reported after implantation of an SF-PCIOL.^{18,24} In a series of 122 eyes, Lee et al.²⁵ found RD in 4.9% of adult cases during a follow-up of up to 42 months. In most reported cases, the location of the retinal tear corresponded to the IOL axis near the fixation sutures, suggesting the probable disturbance of the vitreous base by passing needles or lens haptics. This complication may be reduced by meticulous removal of the vitreous before IOL implantation.²⁶ Other less known complications such as IOL tilting, decentration, suprachoroidal hemorrhage, glaucoma, iris capture, chronic inflammation, and filtering bleb at the site of suture fixation have been reported in the literature.^{18,27–30}

ANTERIOR CHAMBER (ANGLE-SUPPORTED) IOL IMPLANTATION

The implantation of an open-loop ACIOL is the simplest surgical procedure for surgical aphakic correction

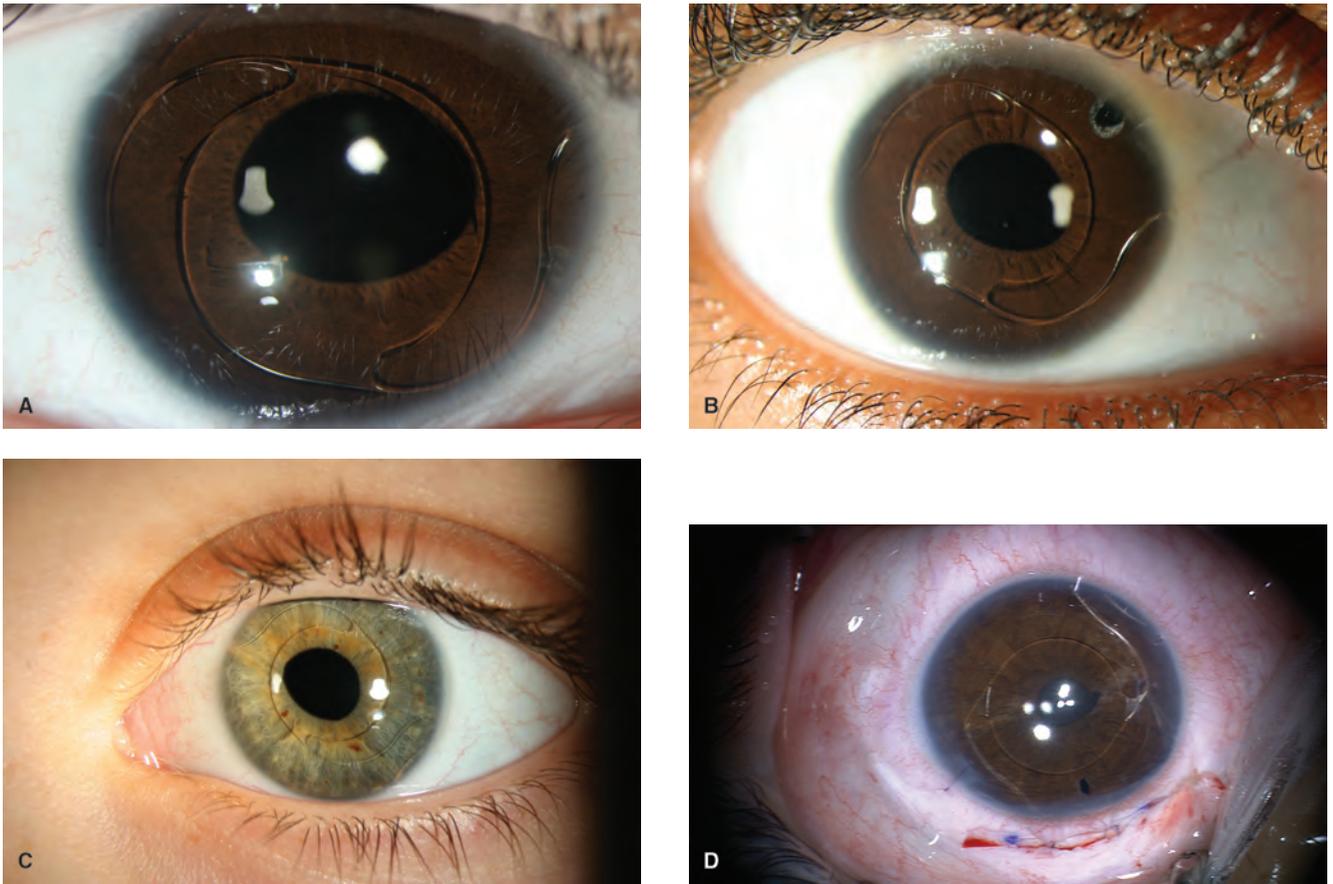


Figure 32.4 A–D: ACIOL in eyes with pediatric aphakia.

(Fig. 32.4A–D). Modern, flexible ACIOLs have a much lower incidence of complications compared to the poorly tolerated rigid closed-loop ACIOLs of the past. The current designs of ACIOLs remain a viable alternative to sutured IOLs when there is an absence of capsular support. In fact, no study has demonstrated a clear advantage of sutured IOLs when compared to ACIOLs. With that said, ACIOLs are used with caution in children. The most common complication seen in children relates to sizing of an angle-supported ACIOL. Lens rotation over time (lens is too short) can cause ovaling of the pupil and iris entrapment. A lens that is too long can cause corneal contact and iris atrophy. Correct sizing for AC angle width is critical to prevent IOL rotation and/or corneal contact or iris entrapment and chronic inflammation. Traditionally, surgeons have used the corneal white-to-white measurement +1 mm as a guide for correct ACIOL sizing. However, recent imaging studies with high-speed optical coherence tomography have found that this method is relatively inaccurate and has a lack of correlation, thus creating uncertainty with ACIOL implantation.²³ Furthermore, a relatively large incision of at least 6 mm is required for currently available ACIOLs.

Surgical Technique

A 6-mm scleral tunnel incision is made above the superior surgical limbus. The pupil is pharmacologically constricted and the AC inflated with a viscoelastic device. An open-loop AC polymethyl methacrylate (PMMA) IOL (5.5-mm optic and a total length of 12 or 13 mm depending on the white-to-white corneal measurement) is meticulously positioned (with or without the aid of a Sheets glide). The pupil should be completely round when the lens is properly inserted. A surgical peripheral iridectomy should be performed superiorly. The ophthalmic viscosurgical device (OVD) is removed after complete suture closure of the scleral tunnel with 10-0 or 9-0 suture (usually Vicryl for children). The conjunctiva can be closed with Vicryl suture, plain gut suture, or sealed with fibrin glue. Very few previous studies investigated ACIOL use in children. Morrison et al.³¹ reported short-term encouraging results in their series of eight eyes of five patients with ACIOLs in children with Marfan syndrome. Epley et al.³² reported their experience with ACIOLs versus sulcus-sutured PCIOLs in 28 pediatric eyes. Of the 18 eyes with sulcus-sutured IOLs, two had corectopia and one had a visually insignificant vitreous hemorrhage that resolved

spontaneously. In the 10 eyes with ACIOLs, an undefined number had visually insignificant pigment deposits on the lens surface, two patients had corectopia, and one patient had lens haptic migration requiring explanation. This same eye developed glaucoma necessitating an Ahmed shunt placement. No patient in either group had evidence of chronic inflammation, CME, or corneal decompensation. The implantation of the original, rigid closed-loop ACIOL in adults resulted in bullous keratopathy, glaucoma, hyphema, iris damage, and liberation of inflammatory mediators that potentiated CME.³³ Flexible open-loop ACIOLs are now used more commonly in adults and have a much safer profile. Several studies in adults have demonstrated comparable complication rates and final visual acuities in patients receiving ACIOLs when compared with those receiving sulcus-sutured PCIOLs.^{34,35} A recent meta-analysis performed as part of the OTAC series revealed that insufficient evidence exists to demonstrate the superiority of a posterior versus an ACIOL in adults.³⁶

THE ARTISAN (IRIS CLAW) ANTERIOR CHAMBER IOL

The aphakic Artisan IOL is a peripheral “iris bridge” supported lens (Fig. 32.5). The standard aphakic Artisan IOL is made of Perspex CQ UV (a type of PMMA) and is 8.5 mm in overall diameter. The optic size is 5.0 mm, and the overall body diameter is 5.4 mm. Implantation requires a 5.4-mm incision. Two smaller IOL sizes for microphthalmic eyes are manufactured at 6.5 and 7.5 mm in overall diameter with optic sizes of 4.4 mm. The fixation points for all of the Artisan IOLs are in the immobile peripheral iris. The IOL has a central oval-shaped optic having an anterior vault (minimizes iris damage) and two fixation arms (haptics) that have a cut in them that provides a claw-grasping mechanism. The “iris bridges”

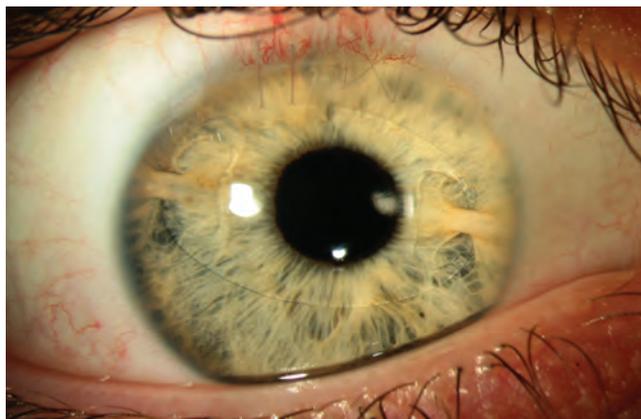


Figure 32.5. Postoperative view of an Artisan IOL implantation in eye with pediatric aphakia.

Table 32.2 IDEAL METHOD OF IRIS CLAW LENS FIXATION

- Optic is centrally placed over the pupil.
- An adequate amount of iris tissue is passed through the claws of the lens.
- The iris root is not pulled in the process.
- The sphincter muscle/pupil margin doesn't get drawn toward the claw.
- There should be no injury to the corneal endothelium either by the implant or by the instruments used to manipulate it.

within the grasping haptics are said to protect the cornea from touching the PMMA haptics of the IOL. In addition, the fixation arms attach to an area of the midperipheral iris that is virtually immobile, thus allowing full dilation and constriction of the pupil. It has universal applications, provided that adequate iris tissue is available for secure fixation. The method of iris claw fixation is described in Table 32.2, while Table 32.3 describes advantages and disadvantages.

Surgical Technique

The pupil is constricted prior to implantation. Calipers are used to mark the 5.5-mm incision width at the 12 o'clock position. Make a half depth corneal or corneal-scleral incision. Make two paracenteses (one at the 2 o'clock and the other at the 10 o'clock positions). If

Table 32.3 ADVANTAGES AND DISADVANTAGES OF ARTISAN IOL IMPLANTATION

Advantages of Artisan IOL Implantation

- The iris bridge protects the endothelium from touching the PMMA
- There are no restrictions to pupil dilation or constriction
- Excellent centration stability once fixated
- The IOL has maximal visibility, accessibility, and controllability
- Virtually cosmetically invisible
- Easy to reposition and is reversible and exchangeable
- No interference with vascular iris physiology
- No sizing is normally needed—one size fits all

Disadvantages of Artisan IOL Implantation

- Requires surgical skill to position properly and create the ideal iris bridge, but the learning curve is short.
- Requires an incision of 5.4 mm ideally located at the limbus since the IOL is grasped through the incision when the iris bridge fixations are created. Early astigmatism is high but fades with healing. Wounds are sutured.

the enclavation needle is to be used, then the paracentesis openings are aimed toward the proposed site of enclavation. If the enclavation forceps are going to be used, then the paracenteses will need to be at 3 o'clock and 9 o'clock positions and are directed toward the pupil. Inject an OVD to fill the AC. OVD should not be placed into the pupillary space, and the AC should not be overfilled. The iris should be flat or slightly concave. Also, cover the conjunctiva with OVD at the wound to reduce contamination of the IOL as it is inserted. Open the tunnel incision with a keratome. The Artisan IOL is inserted in a vertical position at the 12 o'clock position. The IOL is then rotated to the desired position (haptics should be at 3 o'clock and 9 o'clock positions) using the Artisan lens manipulator. Perform the first enclavation with the nondominant hand. Insert the enclavation needle through one of the paracentesis openings (there is a right and a left enclavation needle) and steady the IOL, so it can be easily grasped through the tunnel wound. Insert the implantation forceps through the corneal tunnel and firmly grasp the lens at the optic edge. While securely holding the lens body with one hand, use the other hand to create a knuckle of iris tissue using the enclavation needle. Make a "snow-ploughing" movement at the desired fixation site. Hold the knuckle of iris with the needle while gently pressing the slotted center of the lens haptic over the knuckle, thus fixating the iris tissue in the slot. A significant fold of iris tissue must be delivered through the haptic slot to ensure lens stability. The pupil, however, should remain perfectly round. Repeat the enclavation steps if needed until adequate. Pushing down through the haptic slot from the AC toward the iris will release the enclavation so it can be repeated. Transfer the instruments to the opposite hands and repeat the fixation steps for the second haptic. Enclavate the second haptic with the dominant hand. Assure at the end that the IOL is centered over the pupil and that each iris bridge is adequate, and yet the pupil is still round. A small peripheral iridectomy is highly recommended despite the fact that the IOL is vaulted. The OVD is then removed, and the wound is closed with 2-4 sutures. We use 10-0 Vicryl, but some surgeons use 10-0 nylon.

The Artisan IOL can also be fixated in a retropupillary manner if desired. To accomplish this, the pupil is not initially constricted. The IOL is inserted with the convex side down (upside down) and held behind the pupil with the IOL implantation forceps through the corneal tunnel. As the lens is being inserted behind the pupil, a miotic should be injected into the AC to constrict the pupil. The lens is lifted and tilted slightly to show the contour of the claws through the iris stroma. A fms spatula is inserted through a paracentesis to exert gentle pressure on the slotted center of the lens haptic to perform the enclavation. The same maneuver is repeated on the other side.

Postoperative Complications

Late dislocation of the lens can occur when one of the enclavation sites becomes dislodged, most often due to trauma. Prevention of this complication involves being meticulous about getting the proper amount of iris enclavated into each claw of the IOL. Late endothelial decompensation is expected to be very rare with the Artisan lens, but ongoing long-term monitoring continues. Odenthal et al.³⁷ measured corneal endothelial cell density in three patients with congenital cataracts who were implanted with an Artisan lens at an average age of 2.7 years. After a mean follow-up of 9.5 years (range, 4.7–14.7 years), there was no statistical difference in central endothelial cell density between the operated/implanted eyes ($3,323 \pm 410$ cells/mm²) and the unoperated fellow eyes ($3,165 \pm 205$ cell/mm²).

IRIS-FIXATED POSTERIOR CHAMBER INTRAOCULAR LENSES

In adult eyes, the iris-sutured IOLs have been used successfully to correct aphakia. The application of these techniques in the pediatric age group has been limited. The iris-sutured IOL fixation is preferred by some surgeons over the transcleral IOL fixation technique because it is technically less demanding^{38,39} and requires a shorter operative time. It also believed that the risk for CME, IOL tilt, and late suture breakage is lower in iris-fixated IOLs.^{40,41} Another potential advantage of iris suturing is a lower risk of suture erosion and diminished risk of suture-wick endophthalmitis.^{16,18,32,42,43} Condon et al.⁴⁰ suggested that the lower risk of late suture breakage may be the result of the fact that the elasticity of the peripheral iris provides a more forgiving suspension system than the sclera, which, in a transsclerally sutured IOL, is fixated to a rigid IOL spanning the posterior segment.

Surgical Technique

The pupil is constricted at the beginning of the surgery to facilitate a temporary pupil capture of the IOL optic during the iris suturing. A multipiece acrylic IOL is often used. The lens is injected or delivered into the eye. The three-piece AcrySof® (Alcon, Fort Worth, TX) can be placed into the pupil using the moustache fold technique. The haptics are allowed to unfold into the posterior segment, but the IOL optic is captured by the pupil so that it remains in the AC. A 10-0 Prolene suture on a long curved needle is passed through peripheral clear cornea, under the IOL haptic and back out at another clear corneal location. The suture is tied by using the Siepser "slip-knot" technique. The key to keeping the pupil from becoming distorted is to keep the iris suture path length as short as possible and its location as peripheral as possible. Long suture passes will bunch up the iris tissue, and passes that are not peripheral enough will restrict the

movement of the pupil. The final step in this procedure is to prolapse the optic back into the posterior chamber once the iris sutures have been tied securely around the haptics—fixing them to the peripheral iris. This technique can be done to one haptic rather than to both if there is sulcus support for the other haptic.

Kaiura et al.³⁹ suggested that placement of the iris fixation sutures at the 3 o'clock and 9 o'clock horizontal meridians may prevent slippage related to equal gravitational pull on each haptic. They further recommended placement of the suture at the midperipheral iris rather than far periphery and positioning of the sutures over the proximal straight portion of the haptics.³⁹ Ensuring that the iris-to-haptic suture is tight enough also may prevent IOL dislocation or slippage.^{40,44} The use of a Siesper sliding knot, as mentioned above, to position the knot in the AC may allow tighter fixation of the haptic to the iris compared to an external knot tied through a large paracentesis at the clock hour of the haptic.⁴⁰

Iris fixation of a PCIOL is an alternative for secondary IOL placement in pediatric aphakic patients without capsular support in the short term but should be approached with caution. Overall, in PCIOL suture techniques, the complication rate seems to be decreasing over the years, probably due to the increasing experience of surgeons in these difficult techniques.

SUMMARY

Several concerns still exist about the AC placement or suture fixation of IOLs in children. While implantation of IOLs in the absence of capsular support may be visually rewarding in well-selected children and also may improve their quality of life, there is a high incidence of complications in the long term. This is especially true in children, for whom a long follow-up with a more active lifestyle is expected. On the other hand, if a child with unilateral aphakia could not tolerate a contact lens and there is no adequate capsular support for PCIOL implantation, surgical options for optical correction are limited. Therefore, until a safe method for fixation of an IOL to the sclera or iris or until some other keratorefractive surgery becomes available, implantation of an AC, iris-sutured, or scleral-sutured IOL are viable options to avoid amblyopia in these cases. We believe that these surgeries should be performed only after the surgeon and parents are convinced that visual rehabilitation of the patient cannot be achieved with a contact lens or a safer surgical procedure. Because of the possibility of late complications such as dislocation of IOL in these children, they should be observed regularly. Further research is needed to find the safest option of optical correction for unilateral aphakia in children who have contact lens intolerance and inadequate capsular support.

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MANAGEMENT OF CATARACT ASSOCIATED WITH OCULAR AND SYSTEMIC COMORBIDITY

33

Preexisting Posterior Capsule Defects

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and Mamidipudi R. Praveen*

Posterior capsule defect (PCD) is associated quite frequently in cases of traumatic cataract in both adults and children.^{1,2} Occurrences of preexisting PCD have also been well documented in adults with posterior polar cataract, and such cases are easily recognized owing to their characteristic appearance.^{3,4} Preexisting PCD also manifests in infants and small children in association with cataract. In children, the presence of preexisting PCD is not easily recognized, because the defect is hidden behind a dense central nuclear or combined nuclear/cortical cataract when viewed through a normal-sized, undilated pupil. This difficulty in recognizing the presence of preexisting PCD in pediatric eyes can pose a challenge to pediatric cataract surgeons.

INCIDENCE

While evaluating 400 consecutive eyes with pediatric cataract surgery at our centre, we observed that 27 eyes (20 children) had preexisting PCD. Thus, the incidence of preexisting PCD was 6.75%, which is significant for pediatric cataract surgeons.⁵ Singh and coauthors⁶ found chalky white spots in a cluster or a rough circle at the level of the posterior capsule before or during surgery. Vajpayee and Sandramouli⁷ observed preexisting PCD in an eye of a 2-year-old patient with congenital cataract. Wilson and Trivedi⁸ reported preexisting PCD in 8/364 (2.2%) eyes implanted with an intraocular lens (IOL) at

the time of surgery for nontraumatic cataracts. Six of the eight eyes presented with leukocoria.

LATERALITY

Preexisting PCD is reported in both unilateral and bilateral cases; however, a higher incidence of unilateral preexisting PCD is reported, which perhaps begins as posterior lentiglobus.⁹⁻¹²

THEORIES AND HYPOTHESIS

Based on our clinical experience, we speculate that the defect perhaps begins at birth as a posterior lentiglobus with herniation of the lens substance through an intact, but developmentally weak, posterior capsule. Many theories have been proposed to explain the development of posterior lentiglobus. These include subepithelial capsule hyperplasia, embryologic hyaloid artery traction,¹¹ inherent weakness of the posterior capsule wall, and the excessive strain of accommodation, which results in posterior herniation of the lens.^{9,10} Franceschetti and Rickli¹² also noted overgrowth or aberrant hypertrophy of the posterior lens cortex. This overgrowth of lens fibers forces backward displacement of a thin and defective posterior capsule.

When the barrier of the posterior capsule is breached, the elements of the crystalline lens are exposed to outside

fluids. The ingress of even scanty vitreous fluid into the crystalline lens triggers a chain of events. There is local hydration, opacification, liquefaction, absorption, and posterior migration of the lens material toward the Berger space. There may also be changes in the rest of the lens and in the anterior capsule. Thus, a break in the posterior capsule has the potential to trigger myriad lenticular changes. Unlike classical posterior lentiglobus, PCD develops at an accelerated pace until a fully thickened defect is seen.⁶

CLINICAL PRESENTATION

In classic cases of preexisting PCD, the defect is hidden behind a seemingly routine pediatric cataract when viewed through a normal-sized, undilated pupil. The defect looks like a total or subtotal white cataract. A preoperative evaluation of such a cataract under maximum dilation is mandatory to unveil the important diagnostic signs. Preexisting PCDs are visibly demarcated by their thick margins due to capsule fibrosis. When the globe is moved with forceps, the degenerated vitreous with white granules moves like a “fish tail” as a result of a long-standing or extensive PCD.

DIAGNOSTIC SIGNS OF PREEXISTING PCD

The following are the characteristic diagnostic signs of preexisting PCD (Fig. 33.1):

- The presence of well-demarcated, thickened margins of the PCD
- White dots on the posterior capsule and in the anterior vitreous
- A “fish-tail sign”—pathognomonic of preexisting PCD



Figure 33.1. The characteristic diagnostic signs of preexisting PCD.

VARIED PRESENTATIONS

The varied forms in which preexisting PCD presents itself are described below:

- The lens is totally white but with a deeper-than-normal anterior chamber and a slight concavity to the anterior capsule rather than the normal convexity of the hydrated but intact white childhood cataract.
- The lens looks membranous, sometimes even transparent, while most of the lens material lies outside the posterior capsule, on the anterior vitreous. There is a clearly visible opening in the posterior capsule (Fig. 33.2A and B).
- A cataractous posterior lentiglobus may show localized opacification (Fig. 33.3).
- A posterior polar or subcapsular cataract may be attached to a persistent hyaloid artery.

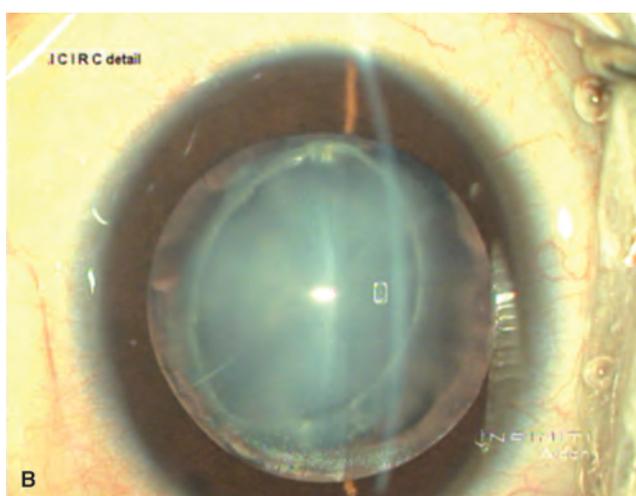
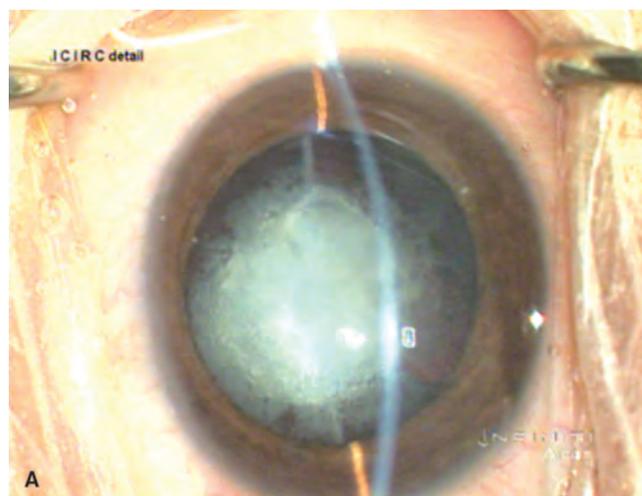


Figure 33.2 A and B: Membranous and transparent cataract with most of the lens material lies outside the posterior capsule, on the anterior vitreous.

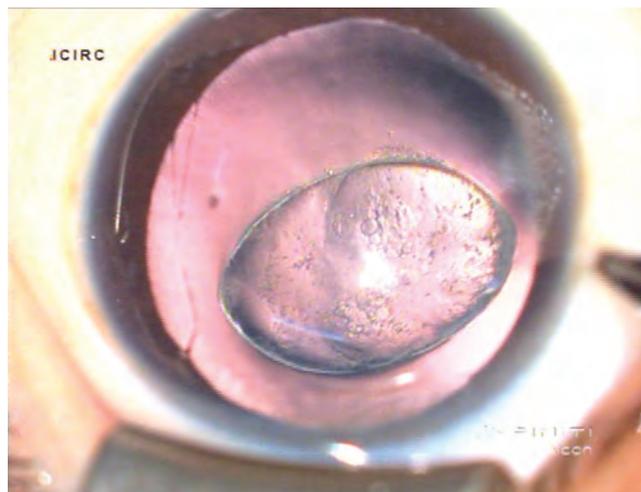


Figure 33.3. A cataractous posterior lentiglobus with preexisting PCD.

When cut, this artery may bleed profusely. Hafez-Abdel and colleagues reported rare occurrence of progression of a unilateral posterior lentiglobus associated with persistent fetal vasculature (PFV).¹³ An 8-month-old boy was found to have unilateral PFV and a small visually insignificant lens opacity. No changes were noted in the appearance of the eye on examinations up to age 3 years. At the age of 4 years, a posterior lentiglobus requiring surgical intervention developed. The thinned and bulging posterior capsule, although intact, was ruptured at surgery.

A peculiar dumbbell-shaped cataract may occur. The periphery is like a Soemmering ring, while the central one-third portion is thick and membranous (Fig. 33.4).

- Posterior polar cataract may occur in the shape of an onion ring.

INVESTIGATIONS

A B-scan ultrasound is necessary when a thorough slit-lamp examination cannot be performed due to the

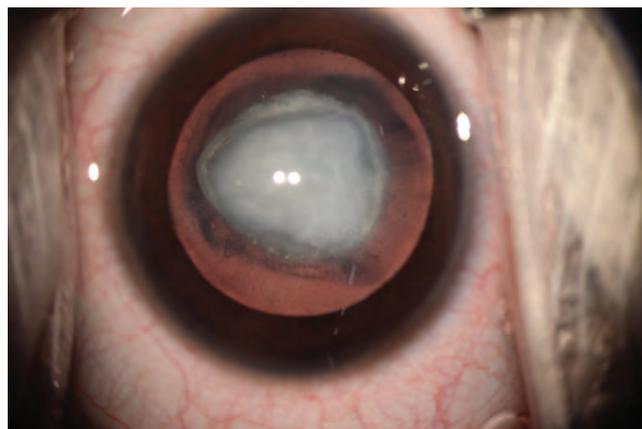


Figure 33.4. A visible opening in the posterior capsule with total cataract.



Figure 33.5. UBM picture suggestive of defect in posterior capsule.

presence of a dense opacity. If the posterior edge of the lens is not sharp and there are new shadows in the adjacent vitreous space, it is suggestive of a possible break in the posterior capsule.

Ultrasound Biomicroscopy

Ultrasound biomicroscopy (UBM) with 25- and 35-MHz probe is an extremely useful tool to detect presence of PCD preoperatively (Fig. 33.5).

SURGICAL MANAGEMENT

Pars Plana Approach

Using pars plana approach in eyes with white mature cataract, the surgeon cannot see where the probe is. If there is visibility, then putting the IOL in the emptied bag and cleaning up the edge of the PCD and cleaning out the vitreous granules of lens material via the pars plana works well. During earlier years, it was noted that with this approach, there are possibilities of traction on vitreous base and the creation of lens vitreous mix, which may give rise to vitreoretinal problems, glaucoma, and inflammation.¹⁴

Limbus Approach

The surgeon should strictly adhere to the principles of the closed chamber technique, such as valvular incision, injection of ophthalmic viscosurgical devices (OVDs) before removing any instrument from the eye, and bimanual irrigation/aspiration. Low flow is maintained since a sudden influx of irrigation fluid may extend the preexisting PCD. Initially, two paracentesis incisions of 1 mm each are made in the clear cornea. High-viscosity OVD sodium hyaluronate is injected thorough the paracentesis incision. Trypan blue (0.0125%) is very useful to stain the anterior capsule in cases in which anterior capsule visibility is poor. The anterior capsulorhexis is

initiated through the same incision. Microrhexis forceps are used to make a nick, and thereafter, capsulorhexis is completed by repeatedly grasping the flap. We believe that manual anterior capsulorhexis is the gold standard in terms of maintaining the integrity of the capsular edge. A Fugo Blade also provides traction-free incisions while performing anterior capsulorhexis.⁶

Performing hydrodissection in cataractous eyes with preexisting PCD is a contraindication. It leads to a sudden buildup of hydraulic pressure, which can enlarge the defect and threaten the stability of the capsular bag. The fluid passes through the defect, entering the posterior compartment and hydrating the vitreous. While some surgeons recommend gentle hydrodelineation in adult posterior polar cataracts, this maneuver is not necessary in children.

Performing bimanual irrigation and aspiration helps to maintain the stability of the anterior chamber as it reduces fluctuations in the iris–capsule diaphragm and minimizes zonular stress. Before removing the instruments from the eye, a high-viscosity OVD is injected through the paracentesis incision to avoid fluctuations of the iris–capsule diaphragm, thereby reducing any peripheral extension of the PCD.

Managing Posterior Capsule in Preexisting PCD

An attempt should be made to convert the PCD into a posterior continuous curvilinear capsulorhexis (PCCC). Converting it to a PCCC with strong, continuous margins allows stable fixation of IOL within the capsular bag and also prevents further extension of the defect during subsequent maneuvers. A small, centric PCCC should be aimed for. However, this is not always possible. A high-viscosity OVD is injected on and around the area of PCD to flatten it. Avoid overinjection of OVD beneath the PCD opening, as it may lead to peripheral extension. Microcapsulorhexis forceps are used to create a nick and then grasp the posterior capsule. The force applied should be centripetal and upward toward the corneal endothelium. When necessary, the virector handpiece can be used to round out the posterior capsule edge while performing a vitrectomy.

Vitreous Management

This depends on the age at surgery and the presence or absence of vitreous prolapse. The use of preservative-free triamcinolone acetonide is invaluable in identifying the presence of vitreous strands in the anterior chamber. A triamcinolone-assisted bimanual limbal vitrectomy is performed.

IOL Implantation

The capsular bag is the most desirable location for placement of the IOL. A single-piece hydrophobic acrylic IOL is implanted inside the capsular bag if possible. However,

a three-piece hydrophobic acrylic IOL is implanted in the sulcus if the PCD is large.

OUTCOMES IN EYES WITH PREEXISTING PCD

As noted above, when we evaluated the diagnostic signs, intraoperative performance, and postoperative outcomes in children with congenital cataract, preexisting PCD was observed in 27 out of 400 eyes (20 children). Seven children had bilateral defects. The mean age of the 16 boys and 4 girls with PCD was 21.98 to 33.33 months. Nineteen eyes (70.3%) had a total white mature cataract. In seven eyes (25.92%), the preexisting PCD was converted into a posterior capsulorhexis. In 20 eyes (74.07%), an AcrySof® MA30BA IOL was implanted in the bag, and in 4 eyes (14.81%), it was implanted in the sulcus. Three eyes (11.11%) were left aphakic. The visual axis remained clear in all eyes, and the IOL was well centered in 24 eyes (88.88%).⁵

We also reported a case of two children with total cataract in one eye and posterior subcapsular cataract in the other. Later in subsequent follow-up visits, we found rapid progression of the posterior subcapsular cataract into a preexisting PCD and subsequently into a white, mature cataract. We propose that early intervention should be considered in cases showing any posterior subcapsular changes (no matter how subtle) and a history of total cataract in the fellow eye, especially if lack of follow-up is likely to occur. In the event that surgery is not advised, parents should be warned about possible cataract progression and the importance of regular follow-up examinations.¹⁵

SUMMARY

The preoperative diagnostic signs of preexisting PCD under maximum pupil dilation include well-demarcated, thick defect margins, white dots on the posterior capsule, and white dots in the anterior vitreous that move with the degenerated vitreous like a fish tail (fish-tail sign).

We recommend that in eyes with preexisting PCD, surgeons should adhere to the principles of the closed chamber technique and use a low flow rate and low bottle height to reduce the possibility of uncontrolled peripheral extension of the PCD.

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Persistent Fetal Vasculature

Irene Anteby and David Morrison

Persistent fetal vasculature (PFV) is one of the most common congenital malformations of the human eye. It includes a complex spectrum of clinical manifestations, which develop due to the abnormal persistence of fetal vasculature. The term PFV, coined by Goldberg¹ in his 1997 Jackson Memorial Lecture, replaced the more commonly used persistent hyperplastic primary vitreous (PHPV).² The substitution of the term PFV reflects Goldberg's more accurate description of anatomic and pathologic features of this disease. In PFV, some, or all, components of the fetal intraocular vasculature remain after birth. This malformation may affect the anterior, retrolental, and/or posterior parts of the infant eye. The extent of the vascular anomaly directly influences both the prognosis for and the therapeutic approach to the PFV eye.

CLINICAL MANIFESTATIONS

It is important to recognize the range of clinical manifestations in PFV. Knowledge of the embryologic milestones responsible for the development of fetal vasculature is crucial for the understanding of the diversity of symptoms and signs appearing in PFV. During fetal development, transient sets of proliferating blood vessels extend throughout the posterior and anterior poles of the eye. The vessels anastomose freely, creating a rich network by the equator and thus connecting all compartments of the eye. These fetal vessels start to grow during the 1st month of gestation, reach their maximum proliferative activity by the 2nd to 3rd month, and begin to involute at 4 months' gestation. Normally, these fetal vessels disappear by birth.^{1,3} In eyes with PFV, the process of fetal vascular regression is arrested. Persistence of some or all fetal vasculature may have profound morphologic consequences. Although individual components of the fetal vasculature often persist in combination with others, any one of the vascular remnants either may predominate in such combinations or may occur alone. Therefore, PFV may cause any of several clinical variants.¹

- *Persistent pupillary membrane.* When the tunica vasculosa lentis fails to properly regress, thread-like vessels or pigmented strands may be seen on the lens surface or arising from the iris collarette and attaching to the anterior lens capsule. The pupil may be deformed by these vessels. In rare instances, the entire pupillary axis is obstructed. A thin fibrous sheet may appear, and congenital iris ectropion (entropion uveae) may also occur. Vision may be unaffected or reduced, depending on the extent of pupillary occlusion. The presence of a pupillary membrane may aid in the diagnosis of PFV in an eye with total cataract or whitish retrolental mass.
- *Iridobyaloid blood vessels.* These fetal vessels appear as radial, short, and parallel vessels by the equator of the lens. They constitute a vascular connection between the posterior and the anterior tunica vasculosa lentis. When these vessels do not regress by the second trimester of gestation, they contribute to the appearance of radial superficial vessels in iris stroma. Often, white limbal connective tissue malformation may be seen in the same meridian. When the vessels reach the pupil, they make hairpin loops, inducing a small pupillary notch.
- *Posterior fibrovascular sheath of the lens.* Persistence of the posterior tunica vasculosa lentis may cause the appearance of a fibrovascular mass behind the lens. Reese² described this as the hallmark of PHPV syndrome. The retrolental membrane may be small or may cover the entire posterior capsule of the lens. It may be associated with a clear lens or cause variable degrees of lenticular opacification. Typically, the retrolental membrane is white or pink in color, differentiating it from the yellow tissue seen in Coats disease or the snow-white tissue typical of calcified retinoblastoma. Formation of a retrolental membrane in PFV is often accompanied by traction on and elongation of the ciliary processes, which may become visible as the pupil is dilated. Although prominent and centrally displaced ciliary processes were once considered pathognomonic for

PHPV, they are also seen in retinopathy of prematurity stage V, Norrie disease, trisomy 13, and congenital subluxated lenses.¹

- *Posterior capsular plaque.* The association of posterior capsular plaque and PFV has only recently been described. Mullner-Eidenbock et al.⁴ first reported that a high proportion of unilateral cataracts had associated findings such as posterior capsular plaque that could represent a subtle form of PFV. The association of posterior capsular plaque and unilateral cataract was later confirmed by the Infant Aphakia Treatment Study (IATS).⁵ In this multicenter study of primary intraocular lens (IOL) placement versus contact lens use at the time of surgical removal of unilateral congenital cataract, 88% of all children with cataract and 100% of infants with unilateral nuclear cataract had an associated posterior capsular plaque. In the IATS manuscript, plaque was hypothesized to be formed by fetal vessels perforating the lens capsule during lens development. Mullner-Eidenbock et al.⁴ theorized that these perforating fetal vessels create an abnormally strong adherence of the lens to the posterior capsule. The vessels subsequently resolve, but the lens opacity and posterior capsular plaque remain (Fig. 34.1).
- *Mittendorf dot.* This small white dot on the posterior surface of the lens is typically found 0.5 mm to the nasal side of the center of the posterior pole and designates the point of incomplete regression of the hyaloid artery, where it attaches to the posterior surface of the lens. It is normally found in 0.7% to 2.0% of the population and rarely causes any visual disturbance.³
- *Persistent hyaloid artery.* The fetal hyaloid artery lies within the Cloquet canal and normally loses perfusion

around the 7th month of gestation. When this vessel persists, it extends from the optic nerve to the lens. It may be filled with blood but is usually bloodless.

- *Bergmeister papilla.* This term is used to describe a benign remnant of the posterior part of the hyaloid artery that can be seen as an epipapillary vascular tissue. Its effect on vision depends on the presence of other associated optic nerve abnormalities.
- *Congenital tent-shaped retinal detachment.* Congenitally detached retina can result from PFV traction on the retina. It typically has the shape of a traction retinal detachment, and it adheres to the posterior surface of the lens, ciliary body, or both. The detachment may progress, and it has grave visual consequences.
- *Macular abnormalities.* Various dysplastic and hypoplastic abnormalities of the macula may occur in PFV, and these will inevitably affect vision.
- *Optic nerve abnormalities.* Both primary and secondary abnormalities of the optic nerve, including optic disc hypoplasia, may be seen in PFV.
- *Microphthalmos.* Retention of fetal vasculature may be accompanied by an arrest in the growth of the eye globe. Typically, eyes with severe forms of PFV have some degree of microphthalmos. Additional changes include a decreased corneal diameter and distortion of the configuration of globe wall, with colobomatous microphthalmos as a result.

ADJUNCTIVE TOOLS FOR DIAGNOSIS

Despite the wealth of clinical manifestations, diagnosing PFV may sometimes be challenging. Any child with a cataract, unilateral or bilateral, especially when associated with

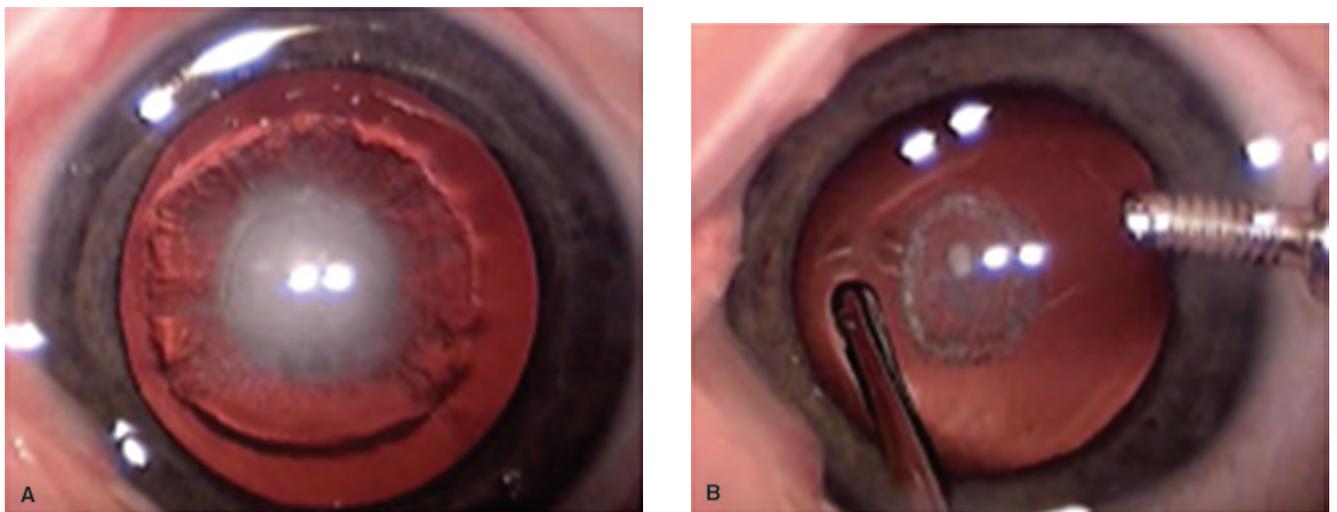


Figure 34.1. Intraoperative photograph of posterior capsular plaque viewed before (A) and after (B) lens removal. Some authors have hypothesized that plaques may be associated with a variant of PFV. (Reprinted from Wilson ME, Trivedi RH, Morrison DG, et al. The Infant Aphakia Treatment Study: evaluation of cataract morphology in eyes with monocular cataracts. *J AAPOS* 2011;15:421–426, with permission from Elsevier.)

a microphthalmic globe, should be suspected of having PFV. When PFV is associated with cataract, the differential diagnosis includes diseases causing leukocoria. When clinical signs are nonconclusive, adjunctive imaging may aid in making the correct diagnosis. The most helpful and noninvasive tool is echography. Both posterior segment echography and ultrasound biomicroscopy of the anterior segment⁶ are valuable. Posterior segment echography typically shows a small globe with a retrolental membrane and a vitreous band extending from the posterior lens capsule to the disc area.⁷ It can also reveal whether a retinal detachment is present, which may influence the choice of surgical technique used to remove the cataract. High-frequency ultrasonography may demonstrate an anteriorly placed and swollen lens with a resultant shallow anterior chamber, centrally dragged ciliary processes, and thickened anterior vitreous face appearing as a double linear echo near the pars plana or pars plicata.⁶ In addition, color Doppler imaging of the persistent hyaloid artery may detect blood flow within the stalk (Fig. 34.2). Computerized tomography and magnetic resonance imaging have also been reported as excellent adjunctive devices in the evaluation of PFV.⁸⁻¹⁰ The demonstration of calcifications within the globe is suggestive of retinoblastoma, which is the most important differential diagnosis to rule out in eyes with leukocoria. The rare occurrence of retinoblastoma in an eye with PHPV has been reported.¹¹

PFV AND ASSOCIATED ANOMALIES

Although PFV mostly appears as a single anomaly, sometimes it may be associated with other ocular abnormalities such as Peters anomaly,¹² Rieger anomaly,¹³ and morning glory syndrome.^{14,15} Only 5% to 10% of children with PFV have binocular involvement. Bilaterality represents a more widespread degree of abnormal embryologic development. Associated systemic anomalies may occur, especially neurologic abnormalities.¹⁶ Haddad et al.¹⁷ reported systemic abnormalities including cleft palate and lip, polydactyly, and microcephaly in association with



Figure 34.2. A 6-month-old boy with PFV in the right eye. Progressive swelling of the lens has induced shallowing of the anterior segment, with resultant angle-closure glaucoma. The boy suffers from marked epiphora from the microphthalmic and leukocoric eye. In addition, his increased irritability prompted his parents to bring him in for examination.

bilateral PHPV. Goldberg reported on the association of PFV with trisomy 13.¹ A few pedigrees with familial PFV have been described,^{18,19} suggesting the possibility of an autosomal recessive²⁰ or autosomal dominant²¹ inheritance pattern in selected cases. In animal models, the presence of PFV has been associated with Arf tumor suppressor gene deficiency,²² angiopoietin-2 deficiency,²³ abnormalities of macrophage-induced programmed cell death,²⁴ and abnormalities of astrocyte cell migration.²⁵

MANAGEMENT

Historically, eyes with PFV, especially when accompanied by dense cataracts hampering ocular stimuli during the critical period of visual development, were doomed to be blind.^{26,27} Surgery was indicated only to avoid or treat complications such as angle-closure glaucoma, vitreous hemorrhage, progressive retinal detachment, and phthisis.² Many eyes eventually required enucleation, with a resultant poor cosmetic outcome.²⁸ Since the advent of closed-system vitrectomy instrumentation, removal of the cataract, retrolental mass, and persistent hyaloid stalk has been made possible. By surgical release of the traction on the ciliary body in eyes with PFV, the eye is allowed to grow and acceptable cosmetic improvement achieved.²⁹ Even though initial surgical goals in eyes with PFV were mainly to avoid the complications of the disease and improve cosmesis, reports of useful postoperative vision following microsurgical vitrectomy techniques began to appear in the 1980s.³⁰⁻³³ Successful visual rehabilitation has been reported for the most part in PFV with anterior presentation, that is, without disc or macular involvement.³⁴

Today, the introduction of sophisticated microsurgical techniques, in combination with aggressive amblyopia therapy, has resulted in more favorable visual outcomes for eyes with PFV.³⁵⁻³⁸ Therapeutic goals should therefore be expanded to include saving useful vision. Several surgical approaches removing the cataract and the retrolenticular fibrovascular membrane in PFV have been described. During the past 15 years, we have applied a modified surgical approach in the treatment of PFV, similar to that previously described for uncomplicated cataracts.^{39,40}

Surgical Methods

Two techniques are described briefly: the pars plana (posterior) approach and the limbal (anterior) approach. In both, the cataract is removed by a lensectomy in combination with an anterior vitrectomy and removal of any retrolenticular membranes or fibrovascular tissue associated with persistence of the hyaloid system.

Posterior Approach

Using the posterior approach, a microvitreoretinal (MVR) blade (20 gauge) is used to perform a sclerotomy

at the 10 o'clock position, 1.5 to 2.0 mm from the limbus. The MVR blade is then pierced through the lens by the equator, leaving an opening in the anterior capsule. A cannula with irrigation fluid is introduced through a similar sclerotomy site at the 2 o'clock position. A vitrectomy handpiece is then introduced through the sclerotomy site at the 10 o'clock position and inserted in the opened bag, and the lens material is aspirated within the bag. After removal of all lens material, the lens capsule and adjacent retrolental membranes are removed by the vitrector. When the retrolental membrane is too thick to be cut with a vitrector alone, intraocular scissors may help to segmentally cut the membrane into fragments small enough for the vitrector to remove.⁴¹ An anterior vitrectomy with removal of the anterior part of the hyaloid stalk is then performed. Possible bleeding from the patent hyaloid artery can usually be controlled by raising the infusion set or by applying diathermy to the bleeding stump. Since all lens capsule material is removed when using this posterior approach, the eye remains aphakic. When posterior abnormalities are present, a complete posterior vitrectomy is suggested, with peeling of membranes to release retinal traction and folds. In some patients, an air-fluid exchange may be indicated.

Anterior Approach

A second approach to cataract removal in PFV is the anterior (limbal or corneal) approach. The main advantage of this method is the feasibility of IOL implantation, enabling better visual rehabilitation and final cosmetic outcome.³⁵ In addition, the anterior approach facilitates surgery by avoiding the peripheral retina, which might be attached to the ciliary body with an absent pars plana.¹⁷ Briefly, in the anterior approach, an MVR blade or 15-degree super sharp blade is used to create two paracentesis incisions at the limbus or in the peripheral

cornea near the limbus: one for the vitrectomy handpiece and a second for the anterior chamber maintainer (irrigation set). These incisions can be made in the superior nasal and superior temporal locations (about 100 degrees apart) or placed superotemporally for the vitrector and inferotemporally for the infusion (Fig. 34.3A,B). A mechanized anterior capsulotomy is performed. The lens is aspirated within the bag. A posterior capsulotomy and removal of retrolental membranes are performed using the vitrector. In cases where the membranes are thick and stiffened, scissors may be used to fragment the membrane before final removal with the vitrector (Fig. 34.3C). Through the opening in the posterior capsule, the anterior vitreous and anterior portions of the persistent hyaloid artery are removed using the vitrectomy-cutting instrument. An IOL can then be inserted either in the bag or in the sulcus, similarly to uncomplicated childhood cataract. Figure 34.3 demonstrates this technique in an eye that was left aphakic.

VISUAL OUTCOME

To achieve favorable vision in children with PFV, irrespective of surgical approach, early intervention followed by aggressive antiamblyopic patching therapy is indicated.³⁵⁻³⁸ Anteby et al.³⁵ reported the visual outcome in 89 eyes with unilateral PFV, comparing 60 operated eyes (using the above-mentioned surgical techniques) to 29 nonoperated eyes. In this large series, a final visual acuity of 20/200 or better in 25% of 60 operated eyes was achieved. Mitra et al.³⁸ reported even more encouraging results of 14 eyes managed by surgery and aggressive antiamblyopic therapy, with 66% achieving 20/100 or better vision. However, in this series, no long-term follow-up was available, and possible late complications affecting vision were not taken into account. Alexandrakis et al.,³⁶

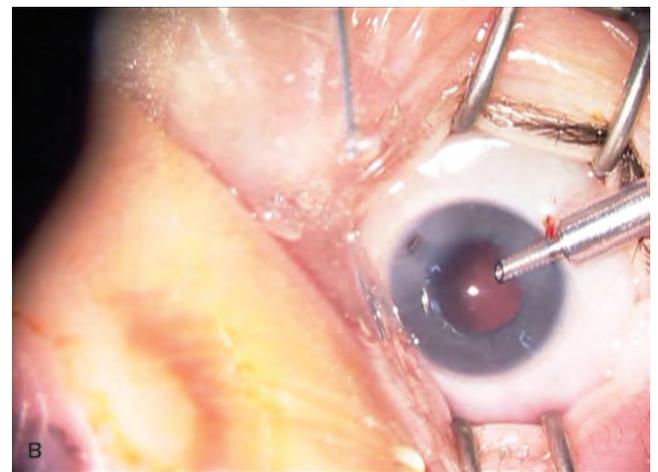
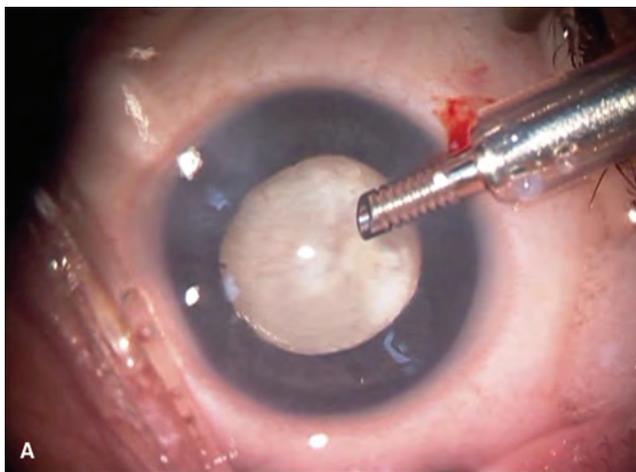


Figure 34.3. PFV cataract with hairpin loop iris vessels and retrolental vascular membrane viewed prior to lensectomy (A), aphakic eye with clear visual axis (B), with incision in dense retrolental membrane using 23-gauge scissors and the vitrectomy handpiece

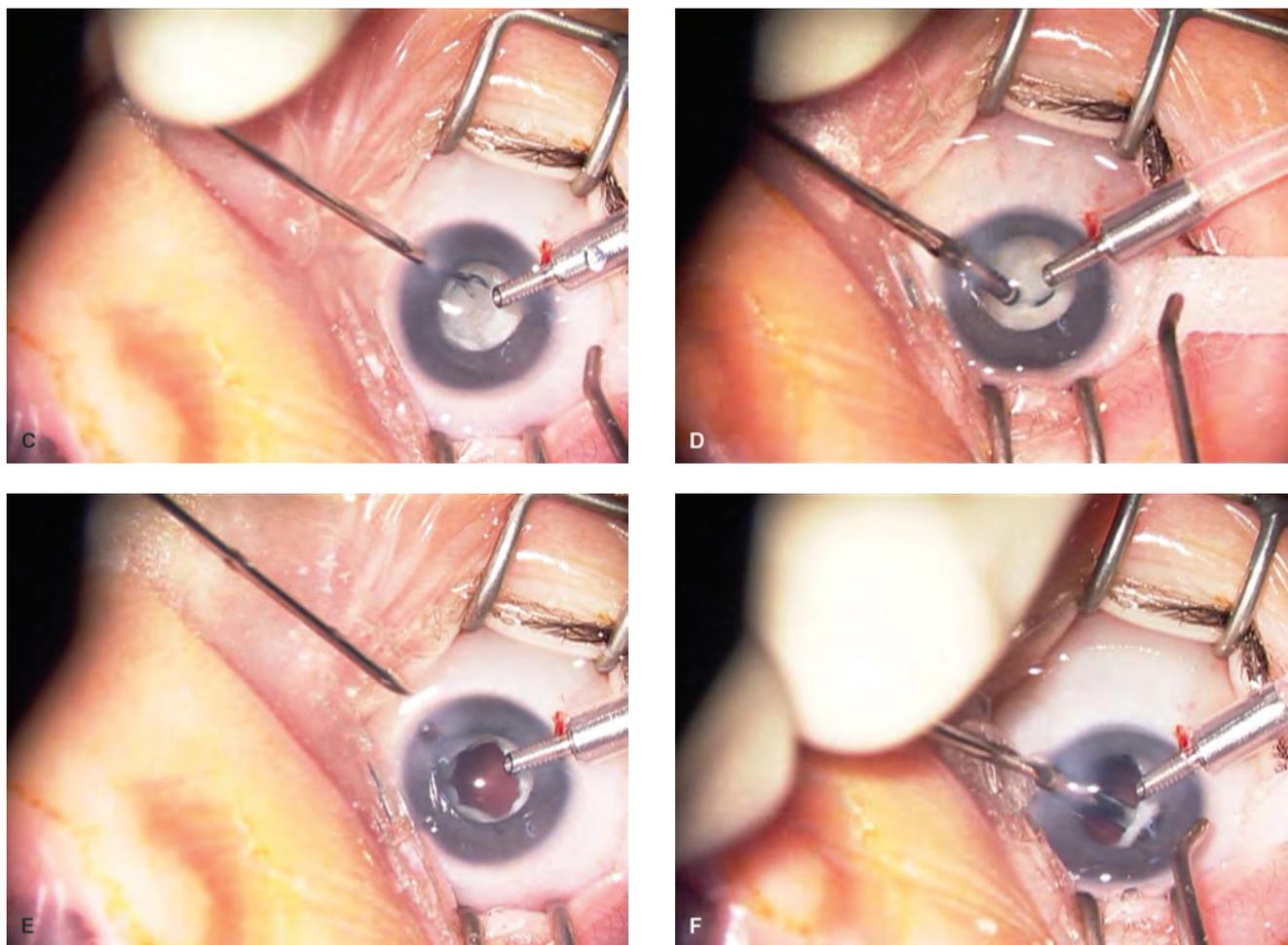


Figure 34.3. (Continued) (C–E), and removal of membrane from stretched ciliary processes (F).

in a study of 30 eyes managed by surgery, reported 47% of the operated eyes achieving 20/400 or better, compared to 12% in nonoperated eyes. Sisk et al.⁴² reported that 49/70 eyes had the presence of form vision after surgery defined as counting fingers or better. In all of these studies, more severe, posterior forms of PFV were associated with a poorer final visual and structural outcome.^{37,42} However, successful visual results have been reported after surgery with PFV of the anterior type.^{28,34}

Traditionally, operated PFV eyes remained aphakic. With an anterior surgical approach, the insertion of an IOL during the initial surgery is feasible. IOLs facilitate postoperative care by avoiding the need for contact lenses. Due to the microphthalmos often encountered in PFV, contact lenses need a high refractive power and can be difficult to fit on a small cornea. Recent reports have included attempts to rehabilitate vision in PFV eyes by IOL implantation. Anteby et al.³⁵ inserted IOLs in 30 eyes with unilateral PFV over the past 15 years. A good visual acuity, 20/50 or better, was seen in 20% of these eyes, and a fair visual acuity, 20/200 or better, was

obtained in 33.3%. Mitra et al.³⁸ reported on the use of IOL in two eyes with PFV with a resultant acceptable visual outcome. Although the IATS was not powered to detect differences in IOL versus contact lens use in children with PFV, results for children with PFV from this study have been reported separately.⁴³ The IATS had specific inclusion criteria for enrollment regarding PFV. Infants had to have a unilateral lens opacity present before 7 months of age with a corneal diameter of 9 mm or greater.⁴⁴ An absence of posterior findings such as ciliary process stretching and retinal traction was also mandated. In the IATS, 18 infants were determined to have evidence of persistent hyaloid remnant or retrolental vascular membrane. Eleven children were randomized to receive aphakic treatment with contact lenses, and seven received a primary IOL. Median logMAR visual acuity at 1 year was 0.88 for patients with PFV and 0.80 for patients without PFV. This difference was not significant. One or more adverse events during the first year after surgery occurred in 67% of infants with PFV and 46% of infants without PFV.

POSTOPERATIVE COMPLICATIONS

The main postoperative complications in PFV eyes include glaucoma, secondary membrane formation, vitreous hemorrhage, retinal detachment, and strabismus. Dass and Trese³⁷ reported a general reoperation rate of 32% in 27 eyes with PFV.

The rate of glaucoma in eyes with PFV varies from series to series. Anteby et al.³⁵ reported a 15% overall rate of glaucoma in 89 PFV eyes. Glaucoma developed twice as often in eyes with aphakia (22%) as in nonoperated eyes (11%). The rate of glaucoma in PFV eyes with pseudophakia was only 8%.³⁵ This suggests that IOL implantation in these eyes does not increase the risk for glaucoma, although eyes selected for IOL implantation may have less severe forms of PFV. Others reported glaucoma to occur in up to 30% of the eyes operated for PFV.⁴⁵ Glaucoma associated with PFV is often diagnosed within the first year after lensectomy but may also develop several years after surgery.⁴⁵

Despite the performance of a relatively large posterior capsulotomy in eyes with PFV, the rate of secondary cataract and membrane formation necessitating further surgery is high—up to 30%.^{35,45} Possibly, this high rate of secondary cataract can be attributed to the microphthalmos itself, as typical postoperative complications have been found to be less common in PFV eyes that are myopic.⁴⁶ It is thought that a more complete relief of circumferential traction on the ciliary processes will reduce reoperation rates. This is done by cutting the retrolental membrane between each stretched ciliary process until they all fall back into a more normal position. The reoperation rate seems unaffected by insertion of an IOL during the initial lensectomy.³⁵ In the IATS, adverse events in the first year after surgery were significantly higher in patients with PFV compared to patients without PFV in the contact lens group (55% versus 20%) but not in the IOL group (86% versus 71%), possibly because all children receiving IOLs had higher rates of adverse events when compared to aphakic children (73% versus 29%) in the study.^{43,44}

COSMETIC OUTCOME

One of the major goals of surgical treatment of eyes with PFV should include the achievement of good cosmetic rehabilitation. Complications such as glaucoma, vitreous hemorrhage, corneal opacification, and phthisis have been attributed mainly to eyes with PFV that are not receiving surgical intervention. These complications often leave the globe gravely disfigured. In addition, as no significant vision is achieved, sensory strabismus develops, further blemishing the child (Fig. 34.4). The quality of life for children with PFV is severely affected by the development of phthisis and deformed eye or the need for a

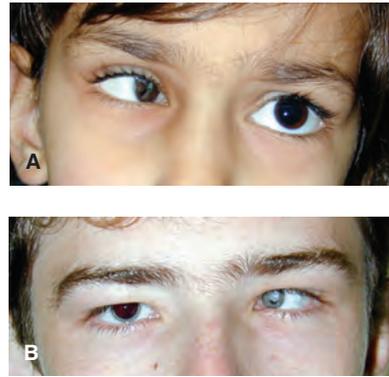


Figure 34.4. When left untreated, eyes with PFV become a cosmetic blemish. These young children have a microphthalmic PFV eye (**A and B**), with leukocoria (**B**), ectropion uvea (**B**), and large-angle esotropia (**A and B**).

cosmetic shell or prosthesis. Anteby et al. reported 30% of PFV eyes developing a visible cosmetic blemish due to advanced microphthalmos, buphthalmos, extensive corneal leukoma, or total phthisis. Interestingly, also operated eyes may become cosmetically blemished. Anteby et al.³⁵ showed that 25% of aphakic eyes and 7.1% of nonoperated eyes needed a prosthesis or cosmetic shell during the years of follow-up, whereas none of the eyes with pseudophakia needed this type of rehabilitation.³⁵ Pollard²⁸ reported that only 2 of 83 eyes with PFV required cosmetic shell. None of the patients needed enucleation. In Scott and coworkers³³ series, the rate of enucleation was also low: 8% in nonoperated and 4% in aphakic PFV eyes.

SUMMARY

The key to success in managing the child with PFV is early diagnosis of the disease. In eyes where vision is gravely impaired by optic axis occlusion owing to cataract and retrolental membranes, early surgery should be attempted to enable visual and cosmetic rehabilitation. During surgery, whether using an anterior or a posterior approach, lensectomy, anterior vitrectomy, release of ciliary body traction, and removal of the anterior portion of the hyaloid stalk should be performed. The rehabilitation of vision may be further facilitated using an IOL when technically possible. Final visual outcome depends not only on the extent of the disease but also on the promptness of treatment, including aggressive antiamblyopic therapy after surgery (see Fig. 34.4). Children with PFV need to be carefully monitored for years, for the possible development of common postoperative complications such as glaucoma and secondary cataract formation. To assure a good quality of life for children with PFV, the goals of therapy should include saving useful vision and achieving a good cosmetic outcome.

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Traumatic Cataracts in Children

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Ocular trauma is a leading cause of unilateral blindness. Children are particularly vulnerable to ocular injury, especially sports-related ocular injury. Prevention of eye injuries is of utmost importance and is the team responsibility of parents, teachers, coaches, ophthalmologists, pediatricians, and optometrists. The American Academy of Pediatrics and the American Academy of Ophthalmology have published a joint statement recommending types of protective lenses and frames for specific sports.¹

CLASSIFICATION OF OCULAR TRAUMA

The Ocular Trauma Classification Group has designed a standardized system for reporting mechanical injury associated with open- and closed-globe trauma. The system is based on the injury type, grade (based on visual acuity), zone, and presence or absence of an afferent pupillary defect.^{2,3} We provide a short summary of the reporting system below. However, interested readers are referred to the detailed description published by Kuhn² and Pieramici³ and to the American Society of Ocular Trauma Web site (<http://www.asotonline.org>).

Closed-Globe Injury

A closed-globe injury is when the eye wall (sclera and cornea) does not have a full-thickness wound. Mechanisms of injury include (1) contusion (blunt force), (2) superficial foreign body, (3) partial-thickness sharp force (lamellar laceration), or (4) a combination of the above.

Open-Globe Injury

An open-globe injury is when the eye wall has a full-thickness wound. These injuries and the mechanisms of injury are further subdivided as outlined below:

1. *Rupture*. Full-thickness wound of the eye wall caused by a blunt object. The impact results in a momentary increase in intraocular pressure (IOP) and an inside-out injury mechanism.
2. *Laceration*. Full-thickness corneal and/or scleral wound caused by a sharp object. This is an outside-in injury mechanism.
3. *Penetrating injury*. Single full-thickness laceration of the eye wall usually caused by a sharp object, with no exit wound.
4. *Intraocular foreign body (IOFB)*. Retained foreign object(s) causing an entrance laceration(s).
5. *Perforating injury*. Two full-thickness lacerations are present with an entrance and exit wound caused by the same agent.

OCULAR TRAUMA AND CATARACT

The crystalline lens may be involved in any case of ocular trauma. Traumatic cataract may be an immediate, early, or late sequel of any ocular trauma. Trauma has been reported to be responsible for up to 29% of all childhood cataracts.⁴ At the Storm Eye Institute, our database includes more than 100 cases of traumatic cataract. We analyzed 47 consecutive eyes of 47 children operated for traumatic cataract.⁵ These consecutive trauma cases represented 10.5% of the cataracts we operated during that time interval. Commonly implicated objects leading to the traumatic event include knives, BB guns, firecrackers, sticks, thorns, rocks, pencils, arrows, airbags, paintballs, and toys.

Blunt trauma is responsible for coup and countercoup ocular injury.⁶ *Coup* is the mechanism of direct impact. It is responsible for the Vossius ring (imprinted iris pigment) sometimes found on the anterior lens capsule following blunt injury. *Countercoup* refers to distant injury caused by shockwaves traveling along the line of concussion. When the anterior surface of the eye is struck bluntly, there is a rapid anterior-posterior shortening accompanied by *equatorial expansion*. This equatorial stretching can disrupt the lens capsule, zonule, or both. Combinations of coup, countercoup, and equatorial expansion are responsible for the formation of traumatic cataracts following blunt ocular injury. *Penetrating trauma* that directly compromises the lens capsule often leads to

cortical opacification at the site of injury. If the rent is sufficiently large, the entire lens rapidly opacifies. When the capsular rent is small, however, the capsule may seal, and the cortical cataract may remain localized.

Cataracts caused by blunt trauma classically form stellate or rosette-shaped posterior axial opacities that may be stable or progressive. At times, an anterior capsular scar (white fibrous capsule) can be seen at the sight of direct impact, especially in children whose eyes are more elastic than those of adults. The pupillary sphincter also often shows multiple small ruptures from blunt trauma, and these may cause a traumatic mydriasis even when the pupil shape is relatively normal.

In penetrating trauma, disruption of the lens capsule often occurs leading to cortical opacification. These cataracts may remain focal if small or may progress rapidly to total cortical opacification. Anterior and/or posterior capsule defect, intralenticular foreign body, partial/total zonular loss, dislocation, and lens subluxation are often found in combination with traumatic cataract. Other associated complications include glaucoma (phacolytic, phacomorphic, pupillary block, and angle recession), phacoanaphylactic uveitis, retinal detachment, choroidal rupture, hyphema, retrobulbar hemorrhage, traumatic optic neuropathy, and globe rupture. Anterior capsule rupture (with flocculent lens matter in the anterior chamber) is usually well tolerated in children. Although an increased IOP and/or lens-induced uveitis (e.g., phacoanaphylactic and phacotoxic endophthalmitis) are known complications of lens rupture, these rarely occur in children. We often cautiously wait 2 to 3 weeks for the eye to heal from the traumatic event before proceeding with cataract surgery (Fig. 35.1), even when flocculent material from lens rupture is seen in the anterior chamber. Outcome of cataract surgery is better when the traumatic iritis and hyphema have subsided.

In the setting of traumatic cataract, the ophthalmologist must first “take a step back” and examine other ocular injuries in detail.⁷ The cataract surgeon should be suspicious of injury to other ocular structures. Management

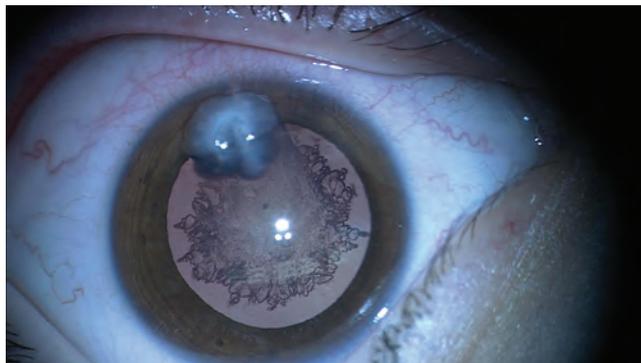


Figure 35.1. A 6-year-old child with traumatic cataract. History of corneal laceration by pencil 2 months ago.

depends on the degree and type of injury. Localized traumatic cataracts (especially if not in the visual axis) may be managed conservatively, while more significant lens opacities generally require cataract extraction. Similarly, capsular perforation may be managed with observation if small and not centrally located. Frequently, such injuries will develop only very localized opacification of the underlying cortex, without progression to generalized cataracts.

The initial patient evaluation is one of the most important critical steps in the management of traumatic cataract. Data gathered during this examination, to a large extent, direct further investigations and establish immediate priorities. One of the most important aspects of this first examination is the description of the exact circumstances of the injury. This facilitates the development of risk estimates for occult injuries, such as IOFB, chemical exposure, and posterior rupture of the globe.

Examination

Before Dilatation

1. Best-corrected visual acuity (BCVA).
2. Fixation preference.
3. Pupillary reflex: The presence of an afferent pupillary defect may be indicative of traumatic optic neuropathy.
4. IOP (if there is no evidence of ocular rupture).
5. Iris: The clinical evaluation should also include a careful predilatation examination of the iris for transillumination defects or tears in the pupillary sphincter. If present, it should be documented, and following dilation, the underlying lens surface should also be inspected for an anterior capsular defect that indicates a penetrating injury or IOFB.
6. Zonule: Although detection of zonular loss is not always possible prior to pupil dilation, suggestive findings include irido- or phacodonesis, an increase in myopic refractive error, abnormal peripheral lens curvature in one or more quadrants, an abnormal light reflex on retinoscopy, a visible lens equator, or vitreous in the anterior chamber.

After Dilatation

1. *Slit-lamp examination* (after pupillary dilation) is recommended if feasible. The slit-lamp examination helps to identify and document the type of cataract, position and stability of the lens, integrity of the lens capsule, and status of the anterior segment. When slit-lamp examination is not possible in the awake state, it can be done using a portable instrument in the operating room in conjunction with the examination using the operating microscope.
2. A *posterior segment examination*, including examination of the retinal periphery, should be carried out in detail if the view through the lens allows. Otherwise B-scan ultrasonography is advisable in all eyes preoperatively.

3. *Gonioscopy* may be helpful for evaluating angle structures and for recognizing vitreous at the lens equator or areas of loss of zonular support.
4. If intraocular lens (IOL) implantation is planned, all eyes need to have keratometry and immersion A-scan ultrasound for globe axial length measurement. Even when corneal scarring is present, keratometry of the injured eye should be attempted. Changes in corneal curvature as the result of an injury will change the IOL power needed to achieve the refractive goal. At times, the keratometry readings of the fellow eye are used, but this can compromise the accuracy of the postoperative refraction in relation to the postoperative goal.

A *guarded prognosis* for anatomical and functional outcome is to be thoroughly explained to the patient and the patient's relatives. The full extent of the eye injuries is not always known prior to cataract surgery. The patient and relative must understand that IOL implantation is not always possible at the initial surgery. It is also important to explain about the possible need for additional surgeries depending on the type of injury (retinal detachment, keratoplasty for dense corneal scar obstructing visual axis, etc.). The use of an IOL in traumatic cataracts is well accepted (Fig. 35.2A and B). It offers a constant, maintenance-free optical correction and, as such, helps in the prevention of amblyopia. Malplacement of the IOL is more common in traumatic cataracts since damage to the capsular bag, zonules, and iris may predispose to decentration and pupil capture. In addition, contact lens wear may be helpful after ocular trauma to help compensate for an irregular corneal curvature caused by a healed corneal wound. However, children often have difficulty wearing these lenses due to discomfort and poor motivation to wear the lens.

With this background, we now describe some of our experience with IOL implantation in eyes with traumatic cataract at Storm Eye Institute. Then, we discuss some of the relevant issues related to pediatric traumatic cataracts.⁸⁻¹⁶ These include (1) timing of surgery for pediatric traumatic cataracts, (2) cataract surgery and IOL implantation, (3) postoperative complications, and (4) visual results.

Analyses of Eyes Operated on for Traumatic Cataracts at Storm Eye Institute

We analyzed 23 consecutive eyes with traumatic cataract.⁵ Ages at IOL implantation ranged from 3.5 to 13.8 years (mean, 6.9 ± 2.5 years). Only 5 (21.0%) of the total 23 patients were females. All 23 children had unilateral cataracts. Nine eyes (39.0%) suffered blunt injury and 14 (60.0%) had penetrating injuries with prior repair of a corneoscleral laceration. The injury preceded cataract surgery by 1 day to 7 years. Nineteen patients (82.6%) had cataract surgery within 3 months of the initial injury. Treatment prior to IOL implantation included repair of the corneoscleral laceration secondary to penetrating trauma in 13 children (56.5%) and repair of a limbal rupture after blunt injury in 1 case (4.3%). Six patients (26.0%) had undergone amblyopia therapy prior to cataract surgery. Preoperative BCVA ranged from 20/40 to possible light perception.

An IOL implantation was performed at the time of cataract removal in 20 eyes (87.0%), while 3 eyes (13.0%) underwent secondary IOL implantation. Two of these patients (who underwent secondary IOL implantation) demonstrated amblyopia preoperatively (vision, 20/50 and 20/100 with poor fusion or strabismus). Traumatic cataracts were noted to be “dense,” “white,” or “total” in 14 of the patients (60.0%), “cortical” in 4 (17.3%), and posterior subcapsular in 2 patients (8.6%), who underwent a primary IOL implantation procedure. Ten patients (43.4%) had an anterior capsule rupture due to the ocular injury noted at presentation or at the time of surgery (Fig. 35.3A and B). Thirteen cases (56.5%) had a mechanical vitrector anterior capsulotomy, and in seven children (30.4%), the anterior capsulotomy was completed manually. An IOL was placed in the capsular bag in 13 (56.2%) eyes and in the ciliary sulcus in 9 eyes (39.1%), and in 1 eye (4.3%), the IOL was placed in the anterior chamber. Six foldable lens designs manufactured with hydrophobic acrylic biomaterial were used, and 14 eyes received rigid polymethyl methacrylate (PMMA) lenses. Fourteen patients (60.8%) (age range, 3.5–10 years) received a primary posterior capsulotomy and anterior vitrectomy. This was done in one case of traumatic posterior capsule

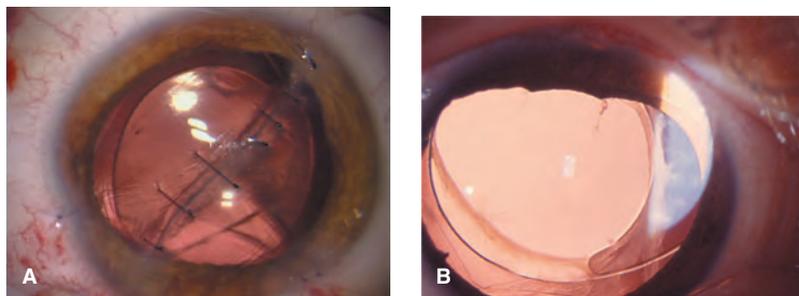


Figure 35.2. **A:** A 7-year-old boy presented with a history of trauma with knife. A heparin-surface-modified (Pharmacia 722C) IOL implanted. **B:** Two-month postoperative photo in a 14-year-old child.

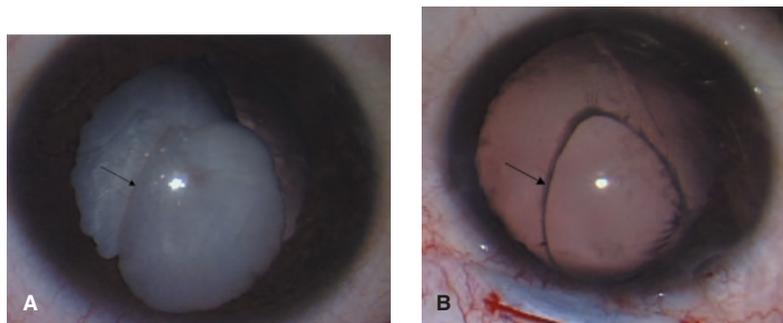


Figure 35.3. **A:** Traumatic cataract associated with ruptured anterior capsule (*black arrow*) and zonular loss in a 5-year-old boy. **B:** After irrigation/aspiration.

rupture and in all patients <6 years old. Overall, the BCVA in 18 patients (78.2%) was 20/20 to 20/40 at the last follow-up visit. Three patients (13.0%) attained 20/50 to 20/80 vision and two patients (8.6%), 20/140 or less. The mean follow-up was 113 ± 99 weeks (range, 4–403 weeks). The most common postoperative complication was the development of visually significant posterior capsular opacification (PCO) in five eyes (21.7%). The posterior capsule was left intact in six eyes (26%) during the cataract surgery and IOL implantation procedure. Of the six children with an intact posterior capsule, five cases (83.0%) developed PCO and required Nd:YAG laser posterior capsulotomy. In addition, two patients (8.6%) also required anterior vitrectomy and posterior capsulotomy, as Nd:YAG laser posterior capsulotomy was not successful in improving the visual acuity. One of these patients suffered a sharp penetrating injury at the age of 5 years, had undergone multiple Nd:YAG capsulotomies, and required surgical posterior membranectomy and anterior vitrectomy 14 months following IOL placement. At the 7-year follow-up examination, the patient was able to achieve a BCVA of 20/20. We have noticed IOL dislocation and pupillary capture in one (4.3%) and two (8.6%) cases, respectively. Dislocation of the IOL was seen in a 4-year-old child who had suffered zonular loss and lens subluxation at the time of injury. A PMMA IOL (Pharmacia Inc., model 815A) was implanted in the ciliary sulcus. This child did well for 19 months of follow-up but then presented with poor vision. The IOL was dislocated into the vitreous. Of two eyes with pupillary capture, the IOL was fixated in the capsular bag in one case and within the ciliary sulcus in another. Both had central corneal scars, anterior capsule rupture, and iris damage. An attempt to correct the pupillary capture was made in both cases. In the latter case (7-year-old), no recurrence of the pupil capture has been seen. In the former (3.5-year-old), the pupil capture was corrected 3 weeks postoperatively. However, the IOL optic recaptured when iris-to-posterior capsule adhesion reformed. No further intervention was attempted, and no persistent inflammation was noted. The BCVA in this case was 20/140 at 66-week follow-up due to a central corneal scar and poor compliance with glasses or contact lenses for corneal astigmatism and amblyopia

therapy. Occurrence of amblyopia in six patients (26.0%) was also associated with poor visual outcome. Three patients (13.0%) developed strabismus postoperatively requiring strabismus surgery.

Timing of Surgery for Pediatric Traumatic Cataract

The timing of traumatic cataract surgery in children is important. Some authors have reported IOL implantation at the time of primary repair.⁹ While the development of amblyopia in children necessitates prompt removal of a cataract when it develops, in our experience, cataract surgery is not necessarily required at the time of initial repair even when anterior capsular rupture is present. This was mentioned in a previous paragraph in this chapter. We prefer to defer cataract surgery and IOL implantation while the inflammatory response is treated with topical steroids. Ten of our patients (43.4%) had anterior capsule rupture and crystalline lens involvement at the time of injury and had their cataract surgery deferred for times ranging between 2 days and 6 months (average, 20 days). Cataract surgery with IOL implantation can often be safely delayed to allow a complete evaluation of damage to intraocular structures. It is important to rule out associated injury (e.g., posterior capsule rupture, vitreous hemorrhage, and retinal detachment) using ancillary methods such as B-scan ultrasonography and to allow healing after the primary repair. However, medically uncontrolled ocular hypertension may occasionally necessitate earlier cataract surgery. Cataract surgery and IOL implantation (combined during the primary repair of ocular trauma) may be considered in younger children predisposed to amblyopia.

Cataract Surgery and IOL Implantation

General principles of pediatric cataract surgery and IOL implantation described elsewhere in this book should be followed. Specific differences are described below.

- *Anesthesia.* General anesthesia is preferable even in older children who might otherwise be cooperative for local anesthesia.

- **Incision.** Although corneal tunnel incisions have become routine, even in traumatic cataract cases, we still perform a scleral tunnel incision on occasion when operating on a traumatic cataract. The integrity of the capsular bag and the zonular support is often in question. If we suspect that conversion from a planned foldable IOL insertion to a rigid PMMA IOL insertion may be needed, small scleral tunnel wounds are more easily enlarged to the 6- to 7-mm length that may be required.
- **Synechiolysis.** Traumatic cataracts are often associated with posterior synechia, and it is necessary to perform synechiolysis in these eyes. Iris reposer instruments or high-viscosity viscoelastic material can be used for this purpose.
- **Anterior capsule management.** Management of the lens capsule in traumatic cataract cases may be difficult due to a ruptured lens capsule with flocculent lens matter in the anterior chamber. Creation of an intact capsulorhexis may be difficult in such a situation. Intraocular microscissors must be on hand for these situations. A vitrectorhexis is also a good alternative to manual capsulorhexis, especially when the anterior capsule is fibrotic and scarred. Thirteen eyes in our series (56.5%) had a mechanical vitrector anterior capsulotomy, and in seven eyes (30.4%), the anterior capsulotomy was completed manually. Very dense fibrous capsule can be removed with intraocular microscissors, radiofrequency diathermy, or Fugo plasma blade. Staining of the anterior lens capsule may be helpful to enhance visibility in these eyes with a “torn anterior capsule” or “white cataract.” Anterior capsule staining can be successfully done using a nontoxic capsular dye such as 0.5% indocyanine green or 0.1% trypan blue.
- **Hydrodissection.** Avoid doing *hydrodissection* in such cases if the integrity of the posterior capsule is unknown.
- **Posterior capsule and vitreous management.** Management of the posterior capsule depends on the age of the patient and status of the posterior capsule (intact versus torn). In our results, a primary posterior capsulotomy was performed in 14 eyes (60.8%); 9 of these patients (39.1%) underwent a pars plana posterior capsulotomy and 5 patients (21.7%) had the posterior capsulotomy performed using an anterior approach. In young patients (too young to sit still at the Nd:YAG laser) with complex trauma, proper placement of the IOL is paramount. If this can be accomplished best by leaving the posterior capsule intact at the initial cataract surgery, plan a secondary surgery to remove the center of the posterior capsule after the IOL has become healed into the lens capsule. This staged approach may be better for surgeons unaccustomed to operating on young children. PCO occurs quickly in most cases of complex traumatic cataract surgery. Therefore, prepare the family that the best vision will likely come after this

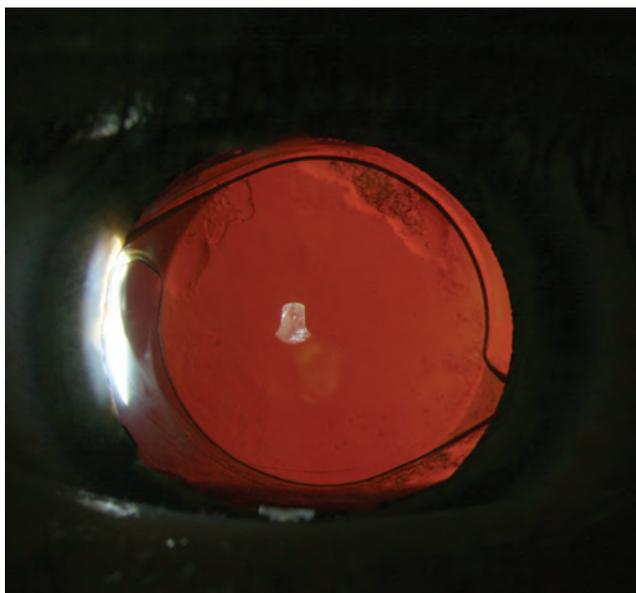


Figure 35.4 Two-year postoperative examination of a child operated for blunt cataract at 6 years of age. Note that primary posterior capsulotomy, vitrectomy, and in-the-bag SN60WF IOL implantation were performed at the time of cataract surgery.

planned second surgery. Primary posterior capsulotomy and vitrectomy increase the chances of a clear visual axis (Fig. 35.4). Repair of iris defects or other more elective surgical maneuvers can also be done during this secondary procedure, which is often done 4 to 8 weeks after the initial cataract removal.

- **IOL Implantation.** Continued advances in IOL designs, biomaterial, and power calculations are making this decision easier in the pediatric population. BenEzra and associates¹⁰ had reported a better visual acuity and less strabismus in children with traumatic cataract after implanting an IOL, compared to those wearing contact lenses. In-the-bag placement of the IOL haptics improves implant stability and minimizes uveitis and pupillary capture.¹⁷ Fifteen of our twenty-one eyes with primary implantation had the IOL placed in the capsular bag. In-the-bag fixation is believed by most to be the best site for IOL implantation, as it sequesters the implant from uveal structures, reduces the chance of lens decentration, and delays PCO formation.¹⁸ However, traumatic cataract cases often present unique challenges, as it is not always possible to fixate the IOL haptics in the capsular bag due to anterior and/or posterior capsule tears from trauma or the difficult surgical procedure. If the IOL must be placed in the ciliary sulcus, as with extensive traumatic posterior capsule rupture, try to capture the IOL optic through the anterior capsulotomy. If this can be done, it will eliminate the risk of pupil capture. Since the iris sphincter is often damaged in trauma, pupil capture of sulcus-fixated IOLs is higher in traumatic as opposed to nontraumatic IOL sulcus fixation.

A technique of glued intrascleral fixation of posterior chamber IOL (glued IOL) has also been reported in children without adequate capsular support.¹⁹

- **Iris suture.** Traumatic iris injury may require suturing. Iridodialysis defects are usually repaired at the time of IOL implantation using a series of double-armed 10/0 Prolene sutures on a long, straight STC-6 needle. A small paracentesis is made 180 degrees away from the iridodialysis. Both needles of the double-armed Prolene are passed through the paracentesis (one at a time) and across the anterior chamber. The needle is allowed to pick up the peripheral detached edge of the iris base and then exits the sclera as close to where that iris segment should naturally attach as possible. Each double-armed Prolene is passed in a mattress fashion and is tied external to the sclera. A Hoffman pocket is a nice alternative to the standard scleral flap for burying the Prolene knot.²⁰ Cuts and tears in the pupil margin are also often closed with the same type of suture material. This can be done at the initial surgery (Fig. 35.5A–D) but is often easier when done as a secondary procedure in a well-healed pseudophakic eye (Fig. 35.6).
- **Removal of corneal suture.** Do not forget to remove corneal sutures from the original traumatic globe rupture repair if they are present and wound healing has been completed.
- **Postoperative medication.** Depending on the case, we may sometime increase the frequency of steroid drops. Also, a short course of systemic steroids may be indicated. If IOP control had been a problem after the original trauma, perhaps during hyphema resolution, it is likely



Figure 35.6. An 11-year-old girl with a history of traumatic cataract removal and secondary IOL implantation. Iris repaired (10/0 Prolene) after 2.5 years of IOL implantation. Photo taken 2 months after iris repair.

that elevated IOP will be seen transiently after cataract surgery. Prophylactic oral Diamox is recommended during the early healing phase in such cases.

Postoperative Complications

The main postoperative complications following pediatric cataract surgery with IOL implantation include PCO and/or secondary membrane formation, pupillary capture, IOL precipitates, and decentration/dislocation of the implant. Complications in our series included visually significant PCO in five cases (21.7%), pupillary capture in two cases (8.6%), and IOL dislocation in one case (4.34%). Posterior capsule opacification developed in five of the six children when the posterior capsule was left intact. We continue to recommend planned primary posterior capsulotomy in children too young to undergo an awake Nd:YAG laser capsulotomy. The occurrence of

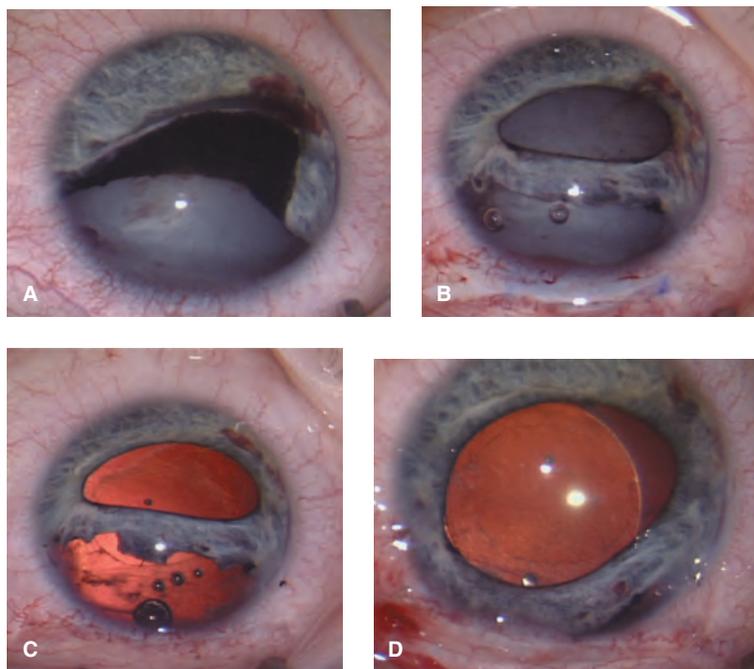


Figure 35.5. A 12-year-old boy presented with a history of trauma 2 months previously. **A:** Traumatic cataract was associated with iridodialysis and loss of zonules in superior 5 clock hours. Iris was folded. **B:** Reposition of folded iris. **C:** After irrigation/aspiration. **D:** Iris was sutured with 10/0 Prolene as the primary procedure and an AcrySof® (SN60, Alcon Laboratory) IOL implanted.

pupillary capture can be reduced after precise fixation of the IOL within the capsular bag. The one eye that developed pupillary capture after in-the-bag IOL placement had a ruptured anterior capsule and iris damage prior to IOL implantation. Had it been possible to maintain an anterior capsular opening smaller than the IOL optic, pupillary capture could have been avoided. Iris-to-posterior capsule adhesions re-formed in this patient, causing recurrence of the pupillary capture. Decentration/dislocation of an IOL can occur because of traumatic zonular loss and/or inadequate capsular support. Capsular bag placement of the IOL combined with a capsular tension ring may be helpful in the presence of zonular dehiscence/loss. Capsular tension rings or segments are not recommended when the integrity of the posterior capsule has been breached. Posterior capture of the IOL optic may be useful, at times, to obtain better centration of the implanted IOL. Asymmetric IOL fixation, with one of the haptics in the capsular bag and the other in the ciliary sulcus, can also lead to decentration and should, therefore, be avoided. Complete IOL dislocation can occur after trauma. Explantation or repositioning of the IOL may be necessary in some cases presenting with significant decentration/dislocation.

Besides PCO, pupillary capture, and implant decentration, poor visual outcome was also associated with amblyopia in six (26%) cases and retinal scar in one (4.3%) case. Three of our patients with significant amblyopia had marked traumatic astigmatism and were poorly compliant with glasses or contact lens wear. Poor compliance with occlusion therapy was also a factor in poor visual outcome. All of the patients with resultant vision $\leq 20/80$ were at an age to be at risk for amblyopia. Amblyopia can develop preoperatively with cataract or aphakia, owing to corneal astigmatism, PCO, or loss of accommodation

with an IOL. Compliance with amblyopia therapy is necessary in younger children to get maximum visual outcome, even following an excellent surgical result.

Visual Results

The prognosis for retention of good vision in pediatric eyes suffering traumatic cataracts has greatly improved over the last few decades. Binkhorst and Gobin,²¹ in 1964, reviewed a case series originally published by McKimura in 1961. Twenty-six children with unilateral traumatic cataracts had been treated at McGill University Hospital and the University of California Medical Center in San Francisco. Despite treatment, most of the patient's visual acuity was in the range of counting fingers; only one child retained a visual acuity better than 20/200. Binkhorst and Gobin²¹ argued that the visual prognosis for young children after treatment for a traumatic cataract need not be so poor. They recommended the use of IOLs in this situation and suggested that this treatment would improve the visual outcomes in children with lenticular opacity. Several surgeons from throughout the world, often in countries with high traumatic cataract rates and conditions prohibitive of contact lens wear, have reported successful IOL implantation in injured children.^{9-11,13,15,16} Our results (as well as the experience of several other authors) confirm that good visual outcome is frequently possible following IOL implantation in children (Table 35.1). In our patients, 78% achieved a BCVA of 20/40 or better after a mean follow-up of 2.3 years. In a comparable series, Koenig et al.¹⁶ reported 20/40 or better visual acuity in 87% (seven of eight) of eyes undergoing IOL implantation for pediatric traumatic cataracts. The average follow-up in their series was 10 months. Gupta et al.¹⁵ reported that 9 (50%) of 18 children with unilateral traumatic cataracts achieved 20/40 (or better) visual acuity after IOL

Table 35.1 STUDIES OF POSTERIOR CHAMBER IOL IMPLANTATION IN CHILDREN WITH TRAUMATIC CATARACTS

Study (Reference Number)	Number of Patients	Age Range (yr)	Mean Follow-up (yr)	BCVA ($\geq 6/12$) (%)	Complications		
					Fibrous Anterior Uveitis	Pupillary Capture	PCO
Anwar et al. ⁹	15	3–8	3.2	73.3	NR	NR	40
Bienfait et al. ¹¹	23	0.4–11	6.5	70.1	0	9	83
Eckstein et al. ⁴	52	2–10	2.9	67	19	41	92
Gupta et al. ^{15,a}	22	3–11	0.9	45	81.8	9	27
Koenig et al. ¹⁶	8	4–17	0.8	87	NR	NR	37
Pandey et al. ¹⁷	20	4–10	2.5	85	45	20	60
BenEzra and Hemo ²⁴	23	2–13	6.2	65.2	NR	26	100
Bustos et al. ²²	19	3–15	0.7	79	26	10.5	21

BCVA, best-corrected visual acuity; PCO, posterior capsule opacification; NR, not reported.

^aFour patients had an anterior chamber IOL.

implantation, with an average follow-up of 12 months. In many cases, corneal leukomata contributed to decreased postoperative visual acuity. Similarly, Anwar et al.,⁹ Bustos et al.,²² BenEzra et al.,¹⁰ Eckstein et al.,⁴ Pandey et al.,¹⁷ and Brar et al.²³ reported visual acuity of 20/40 or better in 73.3%, 79.0%, 65.2%, 67.0%, 85%, and 62% of cases, respectively, after traumatic cataract surgery with IOL implantation in children (see Table 35.1).

SUMMARY

Traumatic cataract can present many medical and surgical challenges to the ophthalmologist. It adds to the challenges presented by childhood cataract. Comprehensive examinations, careful planning for surgical management, and close follow-up are necessary for a favorable outcome in these cases. We support the continued use of IOLs in children in eyes with traumatic cataracts. Further prospective studies are probably needed to specifically address the optimum timing of cataract-IOL surgery in cases of pediatric traumatic cataract. However, based on our experience, we suggest primary repair of the injury first and cataract-IOL surgery after 2 to 4 weeks of topical steroid and atropine treatment. This delay may be helpful in achieving the optimum surgical outcome by reducing the postoperative inflammation in these eyes and allowing healing to occur. Long delays before cataract removal must be avoided during the amblyopia-prone years, which extend to approximate the age of 8 years.

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Cataract can be seen in eyes with familial or traumatic aniridia. This chapter discusses mainly the management of familial aniridia; however, some of the principles are useful for traumatic causes of iris loss as well. Cataracts develop in 50% to 85% of patients with familial aniridia, usually during the first two decades of life (Fig. 36.1).¹ Netland et al.² evaluated the prevalence of ocular and systemic abnormalities associated with aniridia in members of Aniridia Foundation International in a survey. Cataract was documented in 71% of respondents. Frequently, small anterior and posterior lens opacities are noted at birth, but these do not usually cause significant visual difficulty. Aniridia was present at birth in all subjects but not diagnosed until a median age of 1.5 months (range, 0–528 months).² Cortical, subcapsular, and lamellar opacities often develop by the teenage years and may require lens extraction. In those families whose members maintain good vision throughout life, there appears to be a lower incidence of visually significant lens opacities.¹ Yoshikawa et al.³ reported a membranous cataract in association with aniridia.

Other clinical features of familial aniridia include the following:

Decreased vision: Even in the absence of cataract, vision is usually in the 20/100 to 20/200 range in patients with aniridia secondary to macular hypoplasia and optic nerve hypoplasia. Although iris hypoplasia is the most common ophthalmic manifestation of the aniridic eye, it is usually not the major determinant of visual function. Poor visual acuity appears to be correlated with absence of the macular reflex, optic nerve hypoplasia, and the development of cataracts, glaucoma, and corneal opacification.

Photophobia: Photophobia may occur secondary to excessive light stimulation because of poor pupillary constriction. A characteristic facial expression in many children consists of narrowing of the palpebral fissures and furrowing of the brow.

Ectopia lentis: Partial dislocation of the lens, usually due to “weak” zonular fibers (due to a molecular defect of the zonule), is more common in aniridic patients. Ectopia lentis has been reported in 0% to 56% of patients with aniridia.^{1,4}

Glaucoma: Glaucoma in infants with aniridia is rare, although it is relatively common later in childhood. Glaucoma was reported in 46% of respondents.² The median age at diagnosis of glaucoma was 8.5 years (range, 0–58 years).¹ Routine gonioscopic examination is important to detect anatomical changes in angle structure that may progress to angle closure. A small stump of iris can gradually produce angle closure by mechanically covering the trabecular meshwork. Patients with more residual iris (often up to where the collarette would be) seem to be at lower risk of glaucoma from this mechanism.

Corneal involvement: Changes in the corneal epithelium occur in all patients with aniridia over time. A superficial, slightly elevated, faint gray pannus with fine radially oriented blood vessels that stain positive with fluorescein is characteristic. Defects appear in the corneal periphery and progress to the center with age. Corneal erosions and frank ulceration occur in some cases. These lesions may progress to end-stage corneal scarring involving all layers. Dry eye was reported to be present in 53% and keratopathy in 45% of respondents.²

Brandt et al.⁵ reported a markedly increased central corneal thickness in eyes with aniridia. The authors noted an average central corneal thickness at least 100 μm greater than literature-derived normal values.⁵ This may lead to incorrect estimates of intraocular pressure (IOP) by applanation techniques and highlights the importance of monitoring patients with aniridia for the development of glaucoma by gonioscopy and optic nerve examination. Limbal stem cell transplantation may be considered to help prevent or treat corneal blindness in aniridia.

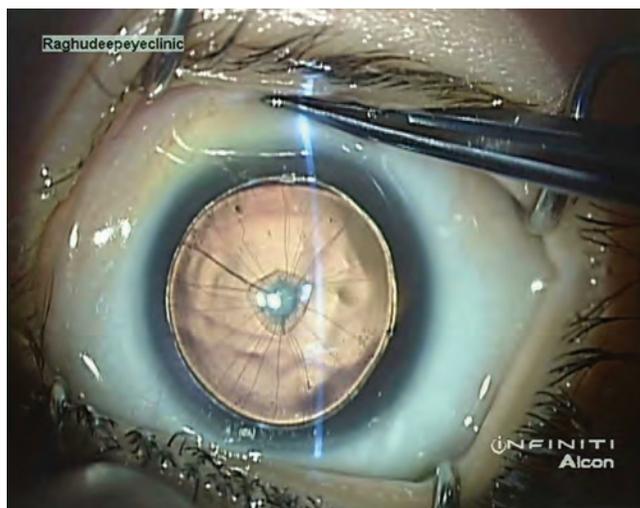


Figure 36.1. Aniridia with anterior polar cataract in a 2-month-old child. (Courtesy Dr. Abhay R. Vasavada and Dr. Sajani Shah, Ahmedabad, India.)

Optic nerve hypoplasia, strabismus, and nystagmus: Macular hypoplasia is usually accompanied by noticeable optic nerve hypoplasia. Foveal hypoplasia was documented to be present in 41% of respondents and absent in 14%, while 45% were not sure.²

Nystagmus and strabismus are common as well. Nystagmus was reported to be present in 83% of respondents, while strabismus in 31%.²

HypHEMA: Theobald et al.⁶ have published a case report of a 2-month-old infant with known hemophilia A and aniridia who presented with spontaneous hypHEMA and severe IOP elevation. IOP remained uncontrolled with medical intervention. Anterior chamber washout was performed; at that time, an extensive pupillary plexus was noted over the anterior lens of both eyes, consistent with persistent iris structures associated with aniridia.

Type 2 aniridia with preserved ocular function: One of us (M.E.W.) has reported two cases of so-called “type 2 aniridia with preserved ocular function.”⁷

Duane retraction syndrome: Association of bilateral Duane syndrome with bilateral aniridia has been recently reported.⁸

Computed tomography (CT) scan: Mehta et al.⁹ noted an abnormal lens shape on CT in a patient with aniridia and cataract. The lens shape appeared reversed; that is, the anterior lens surface was more convex and the posterior lens surface less convex.⁹ A B-mode ultrasound scan showed that the lens capsule shape was normal. In addition, it showed that the cataract was the same shape as the CT image. The CT had imaged the cataract of the patient as opposed to the lens capsule, hence giving the abnormal appearance. The authors cautioned clinicians about misinterpretation when assessing patients with aniridia and cataract by CT scan.⁹

MANAGEMENT OF ANIRIDIA AND CATARACTS

Detailed preoperative assessment of ocular structures including careful evaluation of the zonular apparatus is essential. In the preoperative assessment of the aniridic patient with a cataract, it is important to attempt to ascertain whether the progressing visual loss arises from increasing lens opacification and not from other factors, such as worsening glaucoma or corneal opacification.¹⁰ The guarded prognosis for visual outcome needs to be explained to the patients/parents/legal guardian.

One aspect of an aniridic eye that can be an advantage when performing cataract surgery is that the absence of iris tissue gives better visualization while performing cataract and intraocular lens (IOL) surgery. There are literature reports of thinning of the anterior capsule in association with congenital aniridia.¹¹ However, caution is required, as all aniridic eyes with a thin anterior capsule were in younger patients in the reported series compared with the control group. It is not clear whether younger age or aniridia led to the thinner capsules. Uka et al.¹² reported endoscopic-aided cataract surgery in a patient with corneal opacity associated with aniridia. Many attempts have been made historically to compensate for lack of iris diaphragm, including eyelid surgery, use of colored contact lenses, coloring of the cornea, and the implantation of iris prosthetic devices. The optical correction of aphakia in aniridic patients is difficult because concomitant corneal pannus may be a relative contraindication to contact lenses, and nystagmus may exacerbate the optical aberration of aphakic spectacles. Conventional IOL implantation in eyes with congenital aniridia has been reported in several studies.^{10,13} The lack of a normal iris presents difficulties in placement of an IOL. All attempts should be made to achieve capsular fixation of the posterior chamber IOL. Accurate positioning of an angle-fixated anterior chamber IOL would be problematic, and the lens might be at higher risk of dislocation. In the absence of an iris, an iris-fixated lens is out of the question. Ciliary sulcus fixation of an IOL is potentially unstable.¹⁰ Segal and Li¹⁴ reported a case with a successful outcome of transscleral ciliary sulcus fixation of a posterior chamber lens in an eye with congenital aniridia.

PROSTHETIC DEVICES

A number of prosthetic iris devices have been developed. Iris prosthetic devices are generally used in combination with a posterior chamber IOL (for refractive correction, optical correction is available from 10 to 30 D) and a capsular tension ring (for stabilizing the capsular bag) in eyes with aniridia. Most studies have described the use of prosthetic devices in adult or adolescent populations. Thus, pertinent discussion of the subject will include adult literature as well. We recommend that interested readers visit manufacturers' Web sites to see the available designs and their dimensions

(www.morcher.com and www.ophtec.com). Iris reconstruction implants from Ophtec (Boca Raton, FL, USA) are available in black, brown, green, and blue. Black implants are made of polycarbonate. Brown, green, and blue implants are made of molecularly bound pigment in polymethyl methacrylate (PMMA). The anterior segment reconstruction prostheses are only available in the four standard colors mentioned above, and therefore, perfect color match with the fellow eye is not possible. After implantation in the eye, the color of the prosthesis might look lighter. This is more of an issue with traumatic aniridia, which is more likely to be unilateral. Familial aniridia is more likely to be bilateral, and thus, bilateral asymmetry would not be much of a concern for these eyes. For special situations, Ophtec also offers a custom-designed iris prosthetic system (IPS).

Iris Diaphragm Lenses

Choyce¹⁵ reported the implantation of colored diaphragm IOLs more than 40 years ago. Sundmacher et al.^{16,17} introduced a modern artificial iris implant in Europe in 1994. He reported using the black diaphragm IOL in aniridia. It consists of a plate of black PMMA with a plate diameter of 10 mm, curved haptics, with or without fixation loops. It provides a central opening of variable diameter. Iris diaphragm lenses have their main indication in cases of total to subtotal aniridia, especially when sulcus fixation is mandated. Since its first description, different models have been developed. Morcher (Stuttgart, Germany) single-piece iris diaphragm IOL styles 67F and 67G are currently the most commonly used designs, which have a full iris diaphragm of 10.0 mm in diameter surrounding a central optic 5.0 mm in diameter. They differ only in overall diameter (67F, 13.5 mm; 67G, 12.5 mm) and are inserted through an incision at least 10 mm long. The Artisan (Ophtec, Boca Raton, FL) iris reconstruction lens is a custom-made PMMA IOL with a PMMA–iris diaphragm, containing two or three “lobster claw” fixation haptics devised for fixation at residual iris. It is therefore only suitable for cases of partial aniridia with enough iris tissue left for enclavation of the haptic portions.

Segmental Iris Prosthetic Devices

One drawback of placing the optic within an iris diaphragm is that a relatively large incision is required to obtain a full iris diaphragm. To avoid the drawbacks of large incisions, Hermeking, of Germany, proposed a system in which the optic portion and the iris diaphragm are inserted separately and then assembled in the eye. His Iris Prosthetic System (IPS, Ophtec) consists of different standard combinable elements and is used for artificial iris reconstruction. It provides occlusion of partial or total iris defects and creates a new iris diaphragm (Fig. 36.2). Capsular tension rings with integrated tinted sector shields have been developed to compensate for aniridia (Morcher type 50C).¹⁸ Two rings are implanted. When adequately positioned,

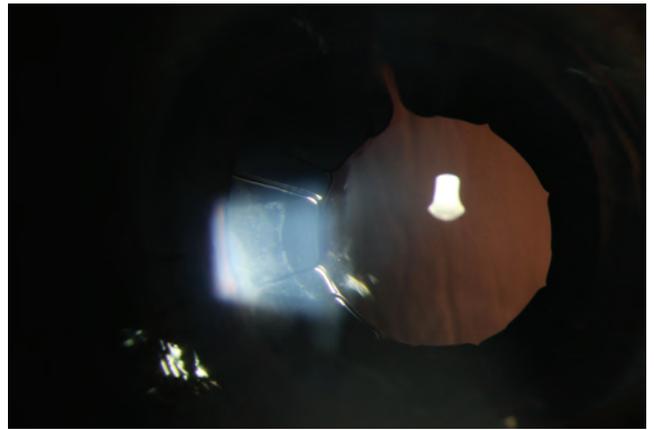


Figure 36.2. Use of ring in an eye with aniridia. (Courtesy Dr. Abhay R. Vasavada and Dr. Sajani Shah, Ahmedabad, India.)

the interspaces of the first ring are covered by the sector shields of the second, forming a contiguous artificial iris. This approach offers the advantages of a full iris diaphragm and separate optical system, both of which may be inserted through a small incision. The iris diaphragm produces a pupil size of approximately 6.0 mm, which is compatible with excellent fundus viewing. The disadvantage of the 50C is that the device is brittle and susceptible to fracture.¹⁹ Segmental iris prosthetic devices are best used when there is an intact capsular bag into which they can be implanted. Their drawback is possible dislocation in the bag, resulting in only partial overlap of the elements with each other or with the iris defect. If this occurs, repositioning may be needed but is relatively easy to accomplish. These segmental iris devices should be used when the capsular bag is perfectly intact and when there is segmental aniridia and a small incision is of high priority. With friable capsules, such as in many cases of congenital aniridia, manipulations with the implantation may cause a capsular break, necessitating a change of the surgical plan.²⁰

The Artificial Iris

A recent addition to management of aniridia is an iris prosthesis made from Gore-Tex material by Schmidt.²⁰ It exhibits a surface structure resembling that of a natural iris and can be custom made to closely match the patient's remaining iris. It is thick but can be rolled up and implanted through a small incision of 3 to 4 mm. It can be used as a complete iris prosthesis, implanted in the capsular bag or suture-fixed in the ciliary sulcus.²⁰ It can also be used to close partial aniridic defects, by cutting out an appropriate segment from the artificial iris and suturing it to the edges of the defect in the patient's iris.

LITERATURE REVIEW

Encouraging short-term results for iris diaphragm lens were reported in 1994.^{16,17} Reinhard et al.²¹ reported long-term follow-up of eyes with black iris diaphragm IOLs.

Almost 75% (14/19) of patients with congenital aniridia and cataracts had increased visual acuity after implantation of the black diaphragm aniridia IOL. Eleven of 14 patients (79%) reported reduced glare. However, these authors feel that this functional improvement in vision was achieved at the (presumed) risk of a variety of complications in some patients.

Osher and Burk¹⁹ reported seven eyes of six patients with congenital aniridia, traumatic iris loss, or chronic mydriasis. All patients noted a marked reduction of glare symptoms and qualitative improvement in their vision after implantation of these prosthetic devices.

Burk et al.²² have reported their experience with prosthetic iris implantation. Glare disability was assessed by questioning patients directly and recording their subjective appraisal of the preoperative and postoperative impairment in bright light and high-contrast settings. The subjective glare disability was reduced from a mean of 2.8 preoperatively to 1.3 postoperatively in eyes with congenital aniridia. The authors recommend the single-piece iris diaphragm IOL (type 67) when the capsular bag is absent or damaged. Of the two models, the 67G is more readily available and well suited for suture fixation. For implantation in the ciliary sulcus, the authors prefer the greater overall diameter of the 67F. The endocapsular devices were generally implanted at the time of cataract surgery. When a full iris diaphragm was needed, two multiple-fin rings (type 50C) were inserted and rotated within the capsular bag until the fins interdigitated, creating a confluent diaphragm.

Pozdeyeva et al.²³ reported that 15/20 (75%) eyes experienced improvement in corrected visual acuity after implantation of iris–lens diaphragms. All patients were satisfied with the cosmetic results of the surgery and reported a decrease in glare and photophobia.

Sminia et al.²⁴ published a series of five pediatric trauma cases using the Artisan iris reconstruction lens. In all of them, implantation was secondary after previous trauma repair and extraction of a traumatic cataract. Cosmetic and functional results were within the limits of the preexisting abnormality, and complications could not be specifically attributed to the implant.

POSTOPERATIVE COMPLICATIONS

Glaucoma: Reinhard et al.²¹ reported that the main postoperative problem noted was chronic IOP elevation, in 42% (8/19). In four of these eyes, chronic glaucoma had been diagnosed and well controlled preoperatively. In the other 4 eyes (29%) of the 14 eyes without evident preoperative glaucoma, chronic glaucoma became evident after implantation of the black diaphragm aniridia IOL. In two, glaucoma could only be controlled by

trabeculectomy, cyclodestruction, and explantation of the aniridia IOL. Although glaucoma is a common complication of congenital aniridia, the blood–aqueous barrier may be altered by the black diaphragm aniridia IOL, accelerating glaucoma progression. The authors hypothesized several reasons for chronic alteration of the blood–aqueous barrier. These include the following: (1) The IOL is too large to be implanted in the capsular bag. Thus, the haptics and the diaphragm are in direct contact with uveal remnants in front of the capsular bag and may cause continuous irritation. (2) Proper placement of an IOL is difficult in an aniridic patient. Improper fixation of haptics may have accelerated the problem. (3) The black diaphragm IOL might have greater mobility than do conventional IOLs after sulcus implantation behind the intact iris of “normal” eyes. (4) The blood–aqueous barrier in eyes with congenital aniridia may be much more vulnerable to all types of trauma than that in normal eyes. The authors recommended caution when using the aniridia IOL in eyes with preexisting glaucoma.

Fibrosis: Patients with congenital aniridia should be monitored carefully for the development of intraocular fibrosis after intraocular surgery, because the risk of aniridic fibrosis syndrome likely increases with increasing intraocular hardware and/or procedures.²⁵

Corneal opacity: Aniridia is a profibrotic disorder and thus has a high rate of failure of surgical interventions including penetrating keratoplasty and trabeculectomy.²⁶

CATARACT, ANIRIDIA AND GLAUCOMA

Congenital glaucoma in patients with aniridia can occur but is rare; the risk for glaucoma increases with age. Angle closure as well as an open-angle mechanism can be causative in these eyes. Cataract may develop after glaucoma surgery (Fig. 36.3). Khaw²⁷ has raised a question in the 2002 issue of *Journal of glaucoma*: “If the patient presents at 29 years of age, not only with raised IOP (28 mm Hg), but also a dense cortical lens opacity, how should he be managed?” Although the patient described there was 29 years of age, we thought it would be interesting for the reader of this text on pediatric surgery. David S. Walton replied, “I should attempt medical therapy and, if necessary, surgery to control the glaucoma. A combined procedure at 29 years of age would seem reasonable.” Comments by Eugenio Maul suggested, “This combination is best treated by a procedure combining an Ahmed valve with phacoemulsification and a foldable acrylic intraocular lens.” The authors further noted, “Glare and edge effects may be reduced by the anterior capsule remaining peripheral to the capsulorhexis, which generally opacify soon after surgery.” Khaw noted, “I carry out drainage tube surgery with Mitomycin-C. I personally

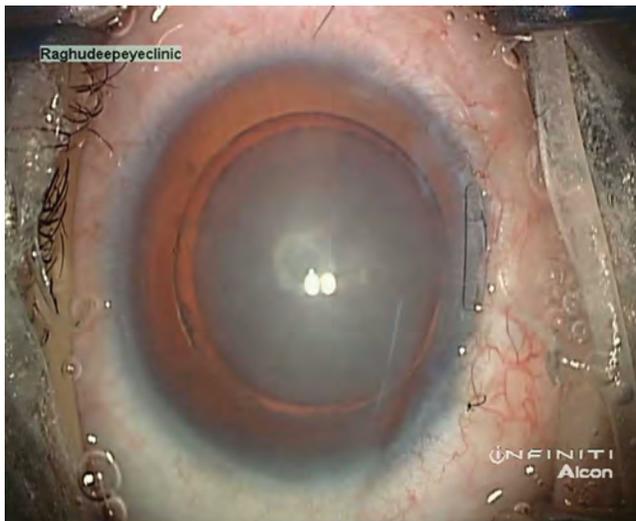


Figure 36.3. Total cataract after trabeculectomy in eye with aniridia in a 5-month-old child. (Courtesy Dr. Abhay R. Vasavada and Dr. Sajani Shah, Ahmedabad, India.)

try not to carry out simultaneous surgery and usually do the tube implant first unless the lens is hypermature and swollen, when I will do the cataract alone if the glaucoma can be medically controlled.” *The authors prefer Baerveldt with Mitomycin-C (MMC).*

Arroyave et al.²⁸ have reported that glaucoma drainage device placement for glaucoma associated with aniridia achieves IOP control and vision preservation in most patients. Most series have reported a significant improvement in visual acuity after cataract surgery in aniridic patients, even though it is subnormal compared to that of patients with cataract without aniridia. Despite a successful outcome of cataract surgery, poor vision in an aniridic child is correlated with absence of the macular reflex, optic nerve hypoplasia, development of glaucoma, and corneal opacification.¹ When cataract surgery and IOL implantation are needed in young children with aniridia, it has been our impression that glaucoma occurs earlier in life in those eyes compared to the average for aniridia patients in general, even when artificial iris devices are not implanted.

SUMMARY

Cataract surgery with aniridia is complicated by pathologic alternations due to the underlying cause of aniridia. Challenges include corneal opacification, younger age at surgery, and reconstruction of iris and pupil. Due to heterogeneity of the underlying conditions and associated abnormalities, a customized approach is needed and careful long-term follow-up is indispensable. Under such circumstances, however, good to excellent results can be obtained in these

usually badly compromised eyes. Implantation of an artificial iris device appears to be a reasonably effective method for reducing the subjective perception of glare resulting from the iris deficiency. As we move forward with the development of artificial irises, the design, availability, and flexibility, as well as insertion techniques, will likely continue to improve and provide better benefits to these eyes.

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Ectopia Lentis

Rupal H. Trivedi and M. Edward Wilson

Ectopia lentis (EL) refers to the displacement of the lens away from its position in the center of the visual axis (Figs. 37.1 through 37.3). When the lens is displaced but remains attached to the ciliary processes by some portion of the zonule, it is referred to as being subluxated (Figs. 37.1 through 37.3). The term subluxated implies a partial or incomplete dislocation and is the preferred term rather than “subluxed.” The term “dislocation” is reserved for situations where there is complete disruption of all zonular attachments and free movement of the lens in the eye. In some patients, the zonule loosens in all quadrants equally, producing high myopia and phacodonesis without the edge of the lens coming into view. This phenomenon is usually also included in the concept of subluxation.

Even without development of cataract, EL often induces large refractive errors and anisometropia. In addition, movement of the dislocated lens can cause an intermittent phakic or aphakic visual axis, leading to marked visual disturbances. Such visual disturbances in a child undergoing visual development can result in amblyopia. Close follow-up is needed when young children have significant EL so that amblyopia can be prevented with glasses correction or amblyopia therapy can be started. Many of these lenses may remain centered long enough to allow the child to develop and maintain normal vision well beyond the amblyogenic years when timing of intervention is less critical.

EL can be hereditary or acquired, and trauma is the most common cause of acquired EL. A Danish national study on nontraumatic EL reported an estimated prevalence rate of 6.4/100,000. In 69% of the cases, nosologic classification was possible; identified etiologies and associations were 68.2% Marfan syndrome (MFS), 21.2% EL et pupillae, 8% simple dominant EL, 1.1% homocystinuria, 0.7% sulfite oxidase deficiency, and 0.7% Weill-Marchesani syndrome (WMS).¹

The zonular fibers connect the ciliary process to the equatorial region of the lens and suspend the lens in a centered position behind the iris and pupil. Zonular fibers (all fibers together are referred to as the zonule)

are rich in fibrillin, which in turn is rich in cysteine and extensively disulfide bonded. The zonule is hence disrupted in MFS, where abnormalities of fibrillin-1 cause the clinical manifestations of the MFS as well as diseases of sulfate metabolism such as homocystinuria and sulfite oxidase deficiency.² During aging, zonular fibers become more fragile, and concomitantly, an increased risk for ocular pathologies is observed. The difference between young and old zonules is likely due to modifications in the structure of microfibrils. Older eyes have fewer fibers than do young ones, suggesting slow turnover, and these fibers are also more brittle.³

Management of EL is challenging. In addition to the optical and visual challenges presented by subluxation of the lens, the potential associated systemic conditions are of major concern, the diagnosis of which may be lifesaving.⁴

NONTRAUMATIC ECTOPIA LENTIS

A. Genetic EL without systemic manifestation

Simple EL: Simple EL occurs either as a congenital disorder or as a spontaneous disorder of late onset. Both are inherited in the majority of cases as autosomal dominant conditions without associated systemic abnormalities. Recessive inheritance is rare, usually occurring in families in which consanguinity has been documented. The ocular anomaly in simple EL is usually manifested as a bilateral, symmetric, upward, and temporal displacement of the lens.⁵

EL et pupillae: EL et pupillae is an autosomal recessive condition that is not associated with cardiovascular or skeletal features of MFS. This purely ocular disease is characterized by combined anomalies of the lens with pupillary displacement. The condition is usually bilateral, commonly not symmetrical but with the lenses and pupils displaced in opposite directions from each other. The pupils are characteristically oval or slit shaped and ectopic, and they may dilate poorly.

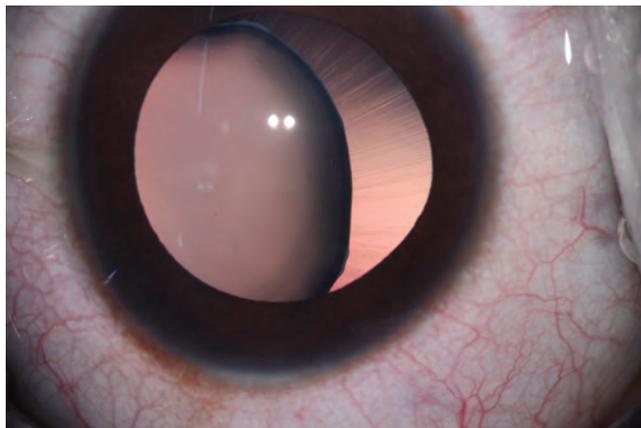


Figure 37.1. Subluxated lens. (Courtesy Dr. Abhay Vasavada, Ahmedabad, India.)



Figure 37.2. Subluxated lens in a 14-year-old child.

There are usually remnants of the pupillary membrane and subtle signs of anterior segment dysgenesis. In EL et pupillae, the lenses can be displaced in any direction, and zonules are generally stretched but may be disrupted. Glaucoma and retinal detachment (RD) are not uncommon and can occur spontaneously or following lens extraction.

B. Systemic disorders commonly associated with EL:

Marfan syndrome: MFS occurs in approximately 4 to 6 per 100,000 births.⁴ It is an autosomal dominant disorder without apparent racial or ethnic predisposition. Mutations of fibrillin gene FBN1 on chromosome 15 have been isolated, producing skeletal, cardiac, and ocular abnormalities.⁴ The recently revised Ghent criteria for MFS puts more weight on EL as a cardinal clinical feature. In the absence of any family history, the presence of EL and aortic root aneurysm is sufficient for unequivocal diagnosis of MFS.⁶ The most prominent ocular features of MFS are myopia and EL. However, since myopia is quite a common finding in the general population, the Ghent criteria have given less importance to it in the systemic score of MFS.⁶

EL occurs in 50% to 80% of patients with MFS. It is almost always bilateral and symmetrical. The amount of dislocation can be stable from early childhood, or it may worsen gradually. Dislocation of the lens in MFS is most typically upward and temporal, but deviation in any direction may occur. Increased globe length and corneal flattening are seen in MFS, but they have unclear specificity.

Weill-Marchesani syndrome: WMS is a connective tissue disorder characterized by abnormalities of the lens of the eye, proportionate short stature, brachydactyly, and joint stiffness. The ocular problems, typically recognized in childhood, include microspherophakia (small spherical lens), myopia secondary to the abnormal shape of the lens, EL, and glaucoma (due to tendency of lens to move into the pupillary space, leading to pupillary block). Although lenticular abnormalities may be absent in MFS, and homocystinuria, the presence of microspherophakia is considered a prerequisite for the diagnosis of WMS. Total dislocation into the anterior chamber is uncommon but may occur.



Figure 37.3. Bilateral subluxation in a 5-year-old child with MFS.



Homocystinuria: Affecting approximately 1 per 200,000 births, this inborn error of amino acid metabolism is more common in some ethnic groups (e.g., Ireland).⁶ Untreated, EL develops in 90% of affected individuals, most commonly in later childhood or early adulthood. The disorder is characterized by variable intellectual disability, EL and/or severe myopia, skeletal abnormalities, and a tendency for thromboembolic events. Overlap with MFS can be extensive and includes an asthenic (long and lean) body habitus, pectus deformity, scoliosis, mitral valve prolapse, highly arched palate, hernia, and EL. The appearance of zonular remnants may help differentiate patients with homocystinuria from those with MFS. Unlike simply elongated zonules of MFS, the lens zonules in homocystinuria are markedly abnormal and broken. Subluxation is typically inferiorly or nasally (57%), and complete dislocation into the anterior chamber or vitreous cavity can occur in up to 32% of cases in some series. The lens in homocystinuria is much more mobile than in the MFS. Typical treatment consists of a low-methionine and high-cysteine diet with supplemental pyridoxine (vitamin B₆), which may prevent or delay mental retardation and lens subluxation if initiated early in life.⁴ Patients with homocystinuria have an increased risk of life-threatening thromboembolic events, especially with general anesthesia.

Other disorders: EL has also been described in hyperlysinemia, sulfite oxidase deficiency, Ehlers-Danlos syndrome, Crouzon syndrome, Refsum syndrome, and scleroderma.⁵

TRAUMATIC ECTOPIA LENTIS

Traumatic EL can occur following blunt trauma to the head, orbit, or eye. Frequently, a high-energy projectile such as a BB gun, golf ball, or baseball is involved. Associated injuries are common and may include iris trauma, sphincter tears, angle recession, hyphema, vitreous hemorrhage, and choroidal rupture. Lens dislocation from apparently minor trauma should raise the question of possible underlying systemic disorders.

PRESENTATION, EXAMINATION AND MANAGEMENT OF ECTOPIA LENTIS

Presentation

EL may cause marked reduction in visual acuity (VA), which varies with the type and degree of dislocation and presence of other ocular abnormalities.⁵ The major symptom of EL is decreased VA because the progressive subluxation of the lens can cause frequent refractive changes and highly irregular astigmatism. Intermittent phakic and aphakic visual axes due to movement of the subluxated lens can also occur. Thus, the optical defocus caused by

severe astigmatism and frequent aphakic condition can cause amblyopia. Monocular diplopia also can occur. Displacement of the lens into the anterior chamber can induce acute angle-closure glaucoma. Displacement into the vitreous cavity may cause lens protein-induced uveitis, vitreoretinal traction, and RD. At times, a dislocated lens can remain for years without causing any complications.

Examination

Each affected individual should have a complete ophthalmologic evaluation of both eyes and an ophthalmologic evaluation of other family members. The majority of affected persons have congenital EL as a manifestation of a systemic disease. It is therefore essential to evaluate EL patients systemically with a general physical examination, a metabolic screening, and an echocardiography as a minimum, in order to make a nosologic diagnosis and to prevent potentially life-threatening systemic complications.¹ Annual ophthalmologic evaluation for the detection of EL, cataract, glaucoma, and RD is essential.

Retinoscopy and Refraction

Retinoscopy in children with EL may reveal a significant refractive error, usually myopia and astigmatism. High myopia may be caused by the higher refractive power of the periphery of the lens or increased curvature of the lens due to the absence of a portion of the zonule. High astigmatism may be due to the periphery of the lens being situated in midpupil, lens tilts, or irregularity of the lens shape because of partial loss of zonular tension. At times, an accurate refraction is difficult because of tilting or dislocation of the lens. If there are both phakic and aphakic parts of the pupil, the aphakic part is usually better for refraction because it is associated with less astigmatism. Careful and repeated phakic or aphakic refractions are necessary to achieve the best possible vision.

Slit-Lamp Examination

The diagnosis of EL usually requires a thorough evaluation with wide dilation of the pupil. However, phacodonesis is more noticeable and dramatic before dilation because dilation often stabilizes the ciliary body and iris, damping any iris or lens movement.⁷ The lens may be minimally subluxated, markedly subluxated and bisecting the pupil, totally dislocated into the anterior chamber,⁸ or free floating in the vitreous.⁵ When the entire zonule is disrupted, as in untreated homocystinuria, the lens becomes globular, its diameter is reduced, and high myopia develops. This is called spherophakia. Spherophakia may also be present in MFS, WMS, or EL et pupillae. The direction of lens subluxation or dislocation and the appearance of the zonular fibers can provide a clue to the systemic diagnosis. In MFS, the lens most often moves superotemporally, and stretched zonular fibers may be visible before or after pupillary dilation. In untreated homocystinuria, the

lenses become dislocated after 4 or 5 years of age; they move initially in an inferior direction behind the iris but may later occlude the pupil or dislocate into the anterior chamber. The latter is almost pathognomonic of homocystinuria. Another differentiating clinical finding is the appearance of the zonular fibers, which are nearly absent in homocystinuria (the equatorial area of the lens has only a fuzz of zonular remnants), as opposed to the elongated and sometimes rarefied zonule of patients with MFS.

When a patient is evaluated for the presence of lens subluxation to rule out MFS, he or she is asked to look down while positioned at the slit lamp. Using retroillumination, the examiner looks for evidence of posterior and superior displacement of the lens by observing the inferior portion of the lens behind the iris. If the lens is in its physiologic position, there is no separation between the pupillary margin and the lens, and the equator of the lens is not visible. If the equatorial region is seen, the lens is considered subluxated. The mere visualization of the zonule on downgaze is not sufficient to diagnose lens subluxation (can be seen in high myopia).

A proposed grading scheme for subluxation of the lens (assuming that the pupil dilates to at least 7 mm) is² as follows:

- Minimal subluxation (the equator of the lens is seen only in downgaze)
- Mild subluxation (the equatorial edge of the lens is visible in primary gaze but only through a dilated pupil)
- Moderate subluxation (the equatorial portion of the lens is visible through the undilated pupil)
- Dislocation (the lens has lost all zonular attachments and moves freely behind or in front of the iris)

Axial Length Measurements

The first histopathologic report on the eyes of a patient with the MFS demonstrated extreme size of the globe.⁵ Further pathologic and clinical reports have confirmed this observation. The mean AL measurement for MFS patients without dislocation was 23.39 mm and for those with dislocation, 25.96 mm. The mean AL in patients 12 years and older with an RD was 28.47 mm versus a mean of 24.90 mm for 40 eyes of patients 15 years and older with the MFS but without an RD.⁹

Keratometry and Pachymetry

MFS is known to be associated with a flattened cornea. These data, known for many years, are included in the Berlin criteria and in the revised diagnostic criteria for MFS as a minor sign. Flat keratometry values may reflect part of the emmetropization process—to compensate for increasing AL, the keratometry value may decrease. Sultan et al. investigated corneal thickness, curvature, and morphology in patients with MFS in a prospective, comparative case series.¹⁰ A significant decrease of the mean

keratometry measurement appeared in the MFS group compared with the control group. Pachymetry in the MFS groups was also significantly decreased compared with the control group. Heur et al. also noted that Marfan patients had significantly lower keratometry and central corneal thickness value than did controls. A highly significant difference in keratometry values between Marfan and control patients was observed, and values <42 D could be used as a clinical diagnostic criterion for MFS.¹¹

Management

Medical

Rapid and adequate correction of refractive errors can prevent amblyopia during the amblyogenic period. In cases where the lens is displaced to only a small degree and is fairly stable, the refractive error may be only slight and easily corrected by glasses or contact lens. Because accommodation is good, bifocals are not normally needed. The patient with moderate or moderately severe lens subluxation presents the greatest challenge. Large amounts of myopic astigmatism can be induced, and the lens edge may bisect the pupillary axis, interfering with any form of optical correction. If the edge of the subluxated lens crosses the pupil, a patient may use the aphakic part with correction for distance and the phakic portion for near vision.⁵

If VA fails to improve immediately, occlusion therapy should be initiated. If good vision can be achieved and there is no evidence of complications, it is not necessary to hurry the surgical correction of EL. The importance of careful refraction through both the phakic and aphakic portions of the visual axis before concluding that optical correction is inadequate cannot be overstated.⁴

Surgical

As discussed before, although many children with EL can be managed conservatively, increasing subluxation results in anisometropia or refractive errors that cannot be corrected with spectacles or contact lenses, leading to poor VA and possible amblyopia. Surgery is also indicated when the displacement of the lens causes secondary glaucoma, corneal endothelial cell damage, or other imminent complications.

In the past, surgical attempts to treat EL were considered dangerous and led to high rates of intraoperative and postoperative complications and poor visual outcomes. Thus, the tendency was to delay surgical treatment unless severe complications were observed. Several noninvasive techniques have been tried in an effort to avoid or postpone surgical lensectomy. These include chronic pupillary dilation to either expose more of the crystalline lens or to allow a sufficient aphakic portion of the pupil for refraction and correction, laser pupilloplasty, laser zonulolysis and optical iridectomy. Although these interventions may

offer some visual improvement in selected individuals, for most severely affected patients, surgical lensectomy is required.⁴ With the advent of ophthalmic microsurgical instrumentation and methods minimizing vitreoretinal traction, lensectomy of the subluxated lens has become a much safer surgical procedure. The methods of visual rehabilitation after surgery include glasses, contact lenses, and intraocular lenses (IOLs), details of which are discussed later in the chapter.

Timing of Surgery

Romano advocated relatively early lensectomy. He stated that it is not necessary, and in fact probably inappropriate, to wait until the pupil is actually bisected by the lens.¹² According to Romano, one need wait only until the edge of the lens is near the center of the pupil; that is, approaching within 2 mm. Once the edge of the lens reaches this point, it is already interfering with vision enough that surgery is indicated. By the time the lens edge reaches the center of the pupil and bisects it, the most amblyogenic period is, in fact, already past. Corrected aphakia is probably less amblyogenic than the partly aphakic/partly phakic situation.¹² The author also hypothesized that anything that blurs vision may stimulate axial growth and produce axial high myopia. This has also been our impression. Those with early lensectomy seem to have less abnormal axial growth over time compared to those without lensectomy. This may be especially true in patients who have an inherent collagen deficiency such as MFS. Therefore, earlier surgical intervention in progressive genetic EL may prevent both amblyopia and high myopia, reducing the risk of RD that is directly related to axial high myopia.¹²

Simultaneous bilateral lensectomy is chosen by some in patients with homocystinuria on the theory that the risks of performing bilateral surgery (bilateral infection) are outweighed by the risks and costs of two general anesthetics in this population.⁴ When choosing this route, totally separate surgical preps, scrubs, and instrument sets are used for the two eyes (see Chapter 9).

Surgical Technique

Before surgery, the surgeon should characterize the areas of zonular weakness in terms of degrees of loss, location of the defect, presence or absence of vitreous prolapse, and the presence or absence of phacodonesis.⁷ The surgeon should be wary of the inferiorly subluxated lens, especially if congenital. Inferior displacement of a congenitally subluxated lens may indicate 360 degrees of very significant zonular weakness combined with the effect of gravity.⁷ Such significant generalized zonular weakness makes it unlikely that the surgeon will be able to remove the lens while maintaining the capsular bag for PC IOL support. Lensectomy should be considered in these eyes. On the other hand, if the etiology of the inferior

subluxation is trauma, there will often be strong inferior zonules remaining, making the chance for successful implantation of an in-the-bag PC IOL more likely.⁷

The ideal position from which the surgeon approaches a subluxated lens is arguable. Frequently, the standard position above the patient's head is not the best one.⁴ However, the surgeon should not compromise his or her surgical abilities by operating at a meridian that is uncomfortable.⁴ For superiorly subluxated lenses, Neely et al. recommended positioning the surgeon at the side of the patient and placing the entrance of the vitrectomy handpiece through the inferior cornea to permit better access to the peripheral lens.⁴ Vasavada et al.¹³ reported using a temporal approach irrespective of the location of subluxation. Cionni⁷ notes that the surgeon should attempt to make the incision away from the area of zonular weakness. This will help reduce the stress placed on the existing zonular fibers. We prefer to approach a subluxated lens from an incision placed 180 degrees from the direction of the subluxation. This approach makes it easier to aspirate the peripheral lens cortex in the part of the lens that is far under the iris. With both the irrigation and the aspiration handpieces inside the capsular bag, one can often keep the capsular bag more centered during surgery than it was preoperatively. Some patients may have generalized zonular weakness, and we have found that the above technique where both instruments are inside the capsule assists in keeping the loose capsular bag centered during aspiration. Some have even suggested that the surgeon place the incision over the quadrant of subluxation because the zonules in the opposite quadrant have proven to be the weakest.

Because the pars plana is relatively underdeveloped in young eyes, some authors prefer the limbal technique. Most anterior segment surgeons are more comfortable with the limbal approach. Limbal approaches are necessary when a lens dislocates into the anterior chamber. Some prefer complete lensectomy with vitrectomy. Others favor the preservation of the posterior capsule with anterior lensectomy techniques.

The surgeon should always work through the smallest incision possible without compromising his or her ability to perform the necessary maneuvers.⁷ Doing so will minimize fluid egress through the incision and, therefore, will help to limit anterior chamber collapse or instability during surgery. The initial anterior chamber entry should be made just large enough to insert the viscoelastic cannula and place a generous amount of a dispersive viscoelastic (e.g., Viscoat, see Chapter 16) over the area of zonular dialysis to tamponade vitreous. Next, a cohesive viscoelastic is injected to maintain a deep anterior chamber.⁷

Anterior continuous curvilinear capsulorhexis (CCC) is a challenge in young eyes with significant subluxation. The anterior chamber should preferably be filled with a highly viscous ophthalmic viscoelastic device (OVD). The

surgeon should start the capsulorhexis in an area remote from the dialysis and use the countertraction provided by the remaining healthy zonular fibers. The capsulorhexis can be made “off-center” as bag recentration with a capsular tension ring (CTR) or modified-CTR (MCTR) will change what appears to be the center of the anterior capsule.⁷ It is often difficult even to penetrate the anterior lens capsule with a regular cystitome. A slit knife can be used for the initial cut. The CCC can then be completed using capsule forceps by pulling the capsule anteriorly, thus reducing the tension on the zonular fibers. When the lens is decentered, it might be very difficult to create a central capsulorhexis of a desired diameter (5.0 to 5.5 mm), since some of the capsule is then hidden behind the iris. If the initial CCC is not wide enough, it can be enlarged later, after lens removal or even at the end of the procedure after IOL implantation.¹⁴ Stabilization of the capsular bag by grasping the CCC edge with iris retractor is recommended.¹³ If a complete and intact capsulorhexis is not obtained, CTR or MCTR placement should not be attempted as the expansile force of these rings will likely induce complete bag rupture.⁷ If the pediatric lens capsule is contracted from chronic subluxation, it may not stretch enough to allow a full CTR to be inserted. For these capsules, we revert to a complete lensectomy and capsulectomy.

Hydrodissection should be done carefully, by repeated injection of a small amount of fluid. Even though it might be quite difficult to achieve a satisfactory hydrodissection when the lens is unstable, this procedure should be attempted.¹⁴ Separation of the lens material from the capsule allows manipulation of lens material with minimal trauma to the zonular fibers. Injection of fluid at various locations, seeing that the nucleus moves anteriorly, followed by gentle pushing of the nucleus backward, indicate that the hydrodissection is complete. Details of CTR usage are discussed below in a separate section.

Lens removal is usually carried out in a routine, yet very careful, manner. The basic rule of surgery in cases of a loose or torn zonule is to minimize the tension over the diseased zonular fibers. The instinct of the beginner surgeon is to work away from the affected area and pull the lens material toward the opposite side. This may stretch the weakened area of the zonule or further unzip the remaining fibers. Therefore, lens material should first be carefully separated from the lens capsule and only then removed with minimal tension. To reduce zonular stress during surgery, always pull toward, not away from, weakened zonules.¹⁴

Several articles have been published on complete lensectomy (including complete anterior/posterior capsule removal) in EL patients. Bimanual irrigation/aspiration techniques are also described (using low aspiration parameters)¹³; however, manual primary posterior CCC is not commonly described in published series of EL.

Menapase noted that a CTR evenly stretches out the posterior capsule, which enhances the control of both central puncturing and tearing of the posterior capsule.¹⁵ He argues that as the capsule is brought under tension, it is moved forward, thereby increasing its distance to the anterior vitreous face. When tangentially punctured by a needle, the central posterior capsule is easily perforated and the risk of compromising the vitreous face is decreased.¹⁵

Residual refractive error can be corrected using glasses or contact lenses. Some authors feel that aphakia means there is nothing to dislodge during soccer practice, no polypropylene sutures to erode or break over the course of decades, no need for secondary procedures to open the posterior capsule, and an optimum view of the peripheral retina. Patients with MFS tend to have longer AL and flatter cornea than do those with normal eyes. In combination, this can result in an aphakic refractive error of $<+5$ D, and in some cases, a negative-power IOL is required to achieve a plano result. Intraocular acrobatics are not justified in these patients as they see quite well with ordinary spectacles that incorporate a progressive bifocal.¹⁶

Despite this controversy, an increasing number of surgeons have attempted IOL implantation after lensectomy in patients with EL. The preferred location of the implanted lens is still controversial. IOLs can be implanted in the anterior chamber (open-loop anterior chamber IOLs or Artisan IOLs enclaved onto the iris) or in the posterior chamber using transscleral fixation or iris suture fixation (Figs. 37.4 through 37.7). Implantation of an IOL should be done using a cohesive OVD. Whereas most surgeons prefer in-the-bag fixation, in some cases, the loose fibers may eventually break, even many years later, and the entire IOL capsule complex may subluxate or dislocate. The preferred direction of the lens axis, relative to the area of missing zonules, is also controversial. Some surgeons prefer to place the lens axis in the

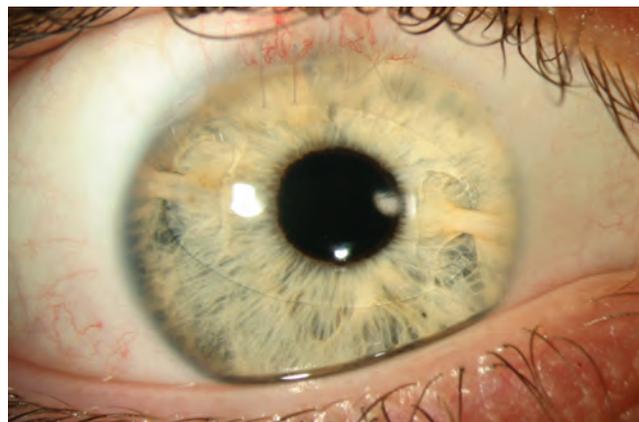


Figure 37.4. One-month follow-up of an Artisan IOL implantation in a child with bilateral idiopathic ectopia lentis. Lensectomy was done at 8 years of age, and Artisan IOL was implanted at 15 years of age.

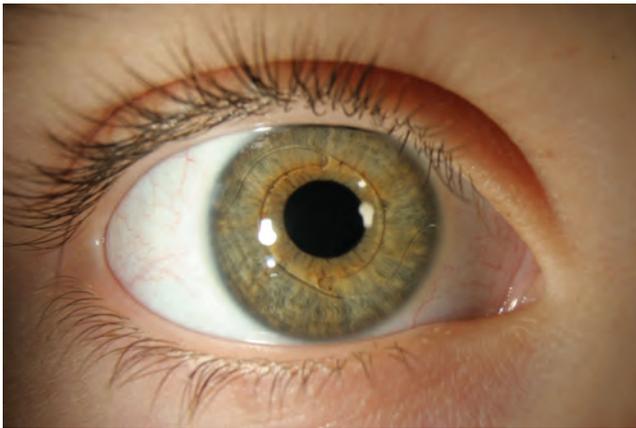


Figure 37.5. Five-year follow-up of a child operated for lensectomy and anterior chamber IOL implantation in both eyes for ectopia lentis.



Figure 37.7. Two-year postoperative view of a child operated for bilateral EL and anterior chamber IOL implantation at 3.5 years of age.

direction of the dialysis so that the IOL haptic will push away the capsular equator.¹⁴ Others advocate placing the haptic perpendicular to the missing zonules to achieve maximal lens support; however, the IOL may then be slightly decentered.

Cleary et al. reported a favorable outcome using Artisan iris-claw lens implants in eight eyes of five children with aphakia following lensectomy for EL.¹⁷ The authors noted that a significant advantage of iris-claw lenses, particularly in patients with MFS who are at higher risk of RD, is that no manipulation of the posterior segment or vitreous is required. This contrasts favorably with the drawbacks associated with scleral-fixated posterior chamber implants.¹⁷

Olsen and Pribila reported a technique that uses a pars plana approach, endoscopic ab interno suture placement, buried knots, substantial scleral imbrication, and a large optic lens while specifically avoiding scleral flaps and exposed suture knots. The IOL was sutured into the posterior chamber.¹⁸

Babu et al. reported a technique using 23-gauge two-port transconjunctival pars plana lensectomy and anterior vitrectomy for the management of EL in children.¹⁹ The sclerotomies were made at inferotemporal and superotemporal sites, 3 mm from the limbus.

Capsular Tension Ring

The introduction of CTRs in 1993 revolutionized lens removal and IOL implantation in eyes with loose zonules (Figs. 37.8 and 37.9). The CTR not only helps to support the IOL postoperatively but is also used as an important surgical tool to allow safe removal of the crystalline lens. The contact of CTR elements on the lens equator, at the location of the epithelial germinal cells, may potentially reduce cell proliferation and posterior capsular opacification (PCO). Using a CTR evenly distributes the forces around the capsular equator, making the IOL position less significant; however, it also adds weight to the compromised capsular bag. The CTR is ideal for traumatic lens subluxation where the zonular disruption is not progressive.

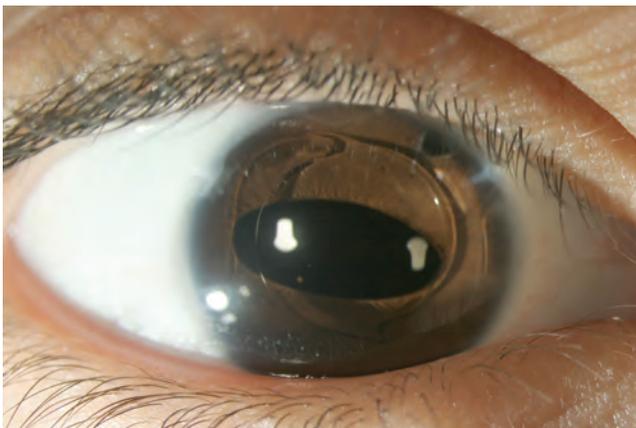


Figure 37.6. Three-year postoperative view of a child operated for bilateral familial EL at 6 years of age.

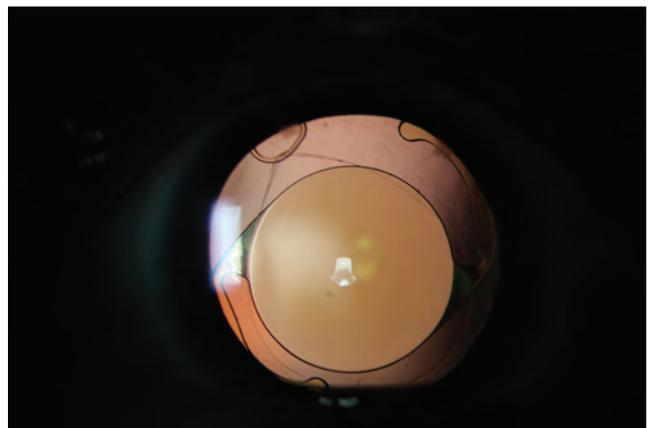


Figure 37.8. Six-month follow-up of Cionni ring in-the-bag IOL implantation. (Courtesy Dr. Abhay Vasavada, Ahmedabad, India.)

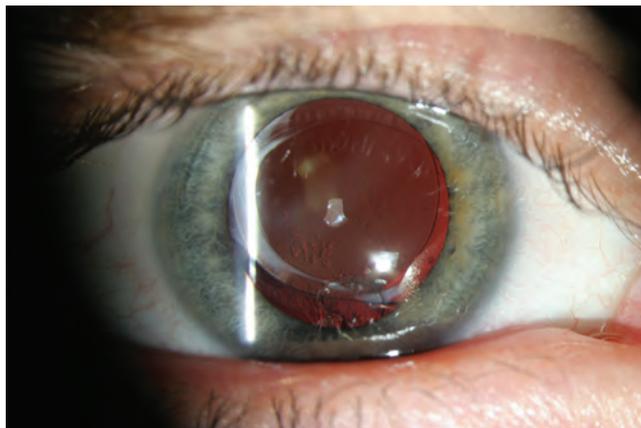


Figure 37.9. Two-year follow-up of a child operated at 14 years of age in both eyes using modified Cionni ring.

Common practice is that if the dialysis is <90 degrees—CTR is an option, not a necessity; 90 to 150 degrees—CTR is required to assure capsular stability and IOL centration. Dialysis of 150 to 210 degrees—CTR can be used but may not be sufficient. The lens or the ring should also be sutured to adjacent structures. Dialysis of more than 210 degrees—usually requires complete lens removal and implantation of an anterior chamber IOL (angle or iris supported) or PC IOL sutured to the sclera and/or iris.¹⁴

There are several models and sizes of CTRs ranging between 12 and 14 mm in the open configuration and 10 to 12 mm when the ring is compressed. In case of small zonular defects, many surgeons prefer inserting the CTR after lens substance aspiration. A large zonular defect, especially in the inferior half, may make lens aspiration a very complex procedure as the loose capsular equator tends to be sucked into the aspiration probe at any attempt to aspirate the lens material. Insertion of a CTR before lens aspiration stabilizes the lens equator and maintains the posterior capsular diaphragm in a taut and backward position.⁷ Before inserting a CTR or an MCTR with an added eyelet for transscleral suturing, the surgeon should place viscoelastic just under the surface of the residual anterior capsular rim to create a space for the ring and to dissect residual cortex away from the peripheral capsule, making cortical entrapment by the ring less likely. The CTR can be inserted manually by using forceps and lens hooks, or it can be inserted using an injector. If inserted manually, it is safer and easier to insert the ring through the side-port paracentesis, rather than through the main incision, as the narrow paracentesis eliminates the side-to-side movements of the ring and allows a smoother insertion. A CTR with an additional positioning hole in the center may further assist ring manipulation and direction.¹⁴ An easier technique is to utilize a spring-loaded injector. The injector is introduced through a corneal tunnel (the paracentesis is too small), and the ring is slowly inserted in a controlled manner and released only when its correct

position has been established. If the ring is misplaced during implantation, the CTR can be easily retracted into the barrel in a reverse motion and reinjected in the proper direction.¹⁴ The loading of the ring determines the direction of insertion. Using the “left” eyelet would load the ring in a counter-clockwise direction, thus releasing it into the eye in a clockwise direction. The opposite occurs when the “right” eyelet is used. The ring should first be directed toward the areas of the loose or missing zonules to minimize the stress on the fibers adjacent to the defect. Since the capsular equator is loose in this area, entanglement of the leading eyelet in the capsule may push the capsule rather than advance the ring. The ring should then be slightly redrawn into the injector and redirected after the bag is refilled with a highly viscous OVD. Before releasing of the second eyelet, it should be assured that the ring edge is posterior and lateral to the edge of the anterior CCC, otherwise the loop might be released into the anterior chamber, over the iris. Retrieval and redirecting the ring into the capsular bag is then a risky maneuver that may damage the angle and cause bleeding. Trapping of cortical fibers between the ring and the capsule often occurs when the CTR is inserted prior to lens removal. A thorough cortical cleavage hydrodissection performed prior to CTR insertion may facilitate cortical fiber aspiration. Removal of the fibers should not be done by pulling them in the regular manner toward the center, as this may inflict stress on the remaining zonules. Preferably, the cortical fibers should be pulled side-to-side in a circumferential manner until they are liberated. When using an MCTR, one should preplace a double-armed suture through the eyelet of the fixation hook before inserting the ring into the capsular bag. Alternatively, the suture can be single armed and the free end of the suture tied to the fixation hook eyelet. Cases of late suture breakage when using 10.0 Prolene suture for this purpose has led to the current recommendation of 9.0 Prolene or 8.0 Gore-Tex suture for MCTR fixation.⁷

Outcome of Pediatric Clinical Series

Retinal Detachment

The cumulative incidence of RD after lensectomy for EL in pediatric eyes has been reported to be as low as 2/342 eyes (0.58%).²⁰ RD can be seen several years after surgery. Neely and Plager reported that two patients developed RD, one patient 5 years after operation at age 8 and one patient 14 years after operation at age 21.⁴ These eyes did not have RD when they were included in a series published in 1992.²¹

Pupillary Capture of the IOL Optic

Lam et al.²² noted that pupillary capture of an IOL occurred after 6 weeks in 2 eyes and after 12 months in 1 eye. All were asymptomatic. It is important to warn parents of this potential complication since repositioning

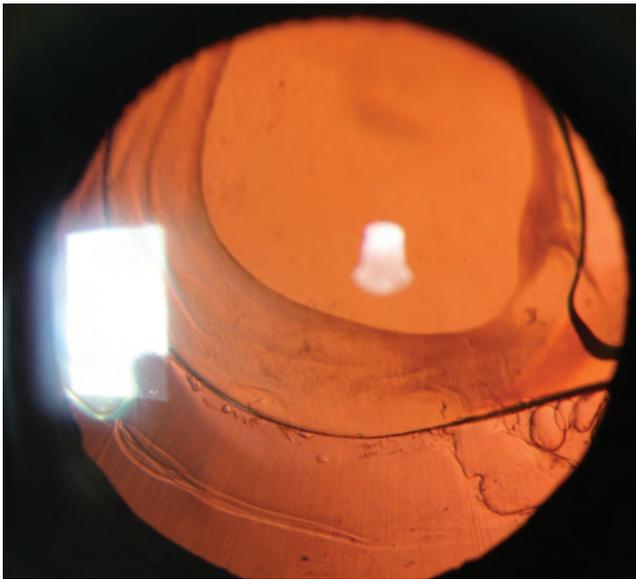


Figure 37.10. IOL decentration in an eye with CTR. (Courtesy Dr. Abhay Vasavada, Ahmedabad, India.)

soon after onset of the pupillary capture is recommended. Regular long-term follow-up is advisable. Reversal of the pupillary capture has also been reported by pupil dilation with the patient in a supine position.²²

IOL Dislocation/Decentration

This is a common complication (Fig. 37.10). Logically, if the crystalline lens was decentered prior to surgery, one may expect that an artificial lens, placed within the same bag, will also be displaced. Kopel et al.²⁰ reported IOL dislocation in 4/12 (33%) eyes in their IOL group.

Visual Outcome

Plager et al. reported 29 eyes of 15 patients undergoing lensectomy at a mean age of 5.8 years (range, 3 to 11 years).²¹ VA improved in all eyes, and 27 of 29 were 20/40 or better postoperatively. In the other two patients, vision was limited by preexisting amblyopia; they were aged 10 years and 11 years at the time of lensectomy.

Shortt et al.²³ reported outcomes of pars plana lensectomy for the management of EL in 24 eyes of 13 children. These authors reported that postoperative VA was 6/9 or better in 17 of 22 eyes and 6/12 or better in 19 of 22 eyes.

Kim et al. reported the results of lensectomy in children with EL.²⁴ The mean age at diagnosis was 4.6 years, mean age at surgery was 5.7 years, and follow-up was an average of 7.1 years. Of 42 patients exhibiting bilateral EL, 6 received monocular surgery. The position of the displaced lens seemed to correlate more closely with the degree of amblyopia than any other factor, including the cause of EL, preoperative VA, or age at the time of surgery.

Kopel et al.²⁰ compared the outcome and complications of pars plana lensectomy–vitrectomy for the management of EL in children with and without a foldable iris-sutured IOL. Visual outcome was not significantly different between the two groups.²⁰

Seetner and Crawford reported the outcome of nontraumatic EL. VA improved in 29/30 eyes and was 20/60 or better in 24 (80%).²⁵

PARTICIPATION IN SPORT

Decisions regarding exercise restriction should be made on an individual basis. Recommendations from the National Marfan Foundation (<http://www.marfan.org>) and guidelines from the American Heart Association/American College of Cardiology task forces are useful resources. In general, patients with MFS should avoid contact sports, exercise to exhaustion, or isometric activities involving a Valsalva maneuver. Most patients can and should participate in aerobic activities performed in moderation.⁶

SUMMARY

MFS, homocystinuria, trauma, and simple EL are the most common causes of pediatric lens subluxation. Many patients are best treated with a careful refraction of the phakic or aphakic pupillary axis. For those patients not achieving satisfactory refractive results, surgical intervention is required. Many advances have been made in the ability to surgically treat patients with a weak zonule or missing zonular fibers. With the introduction of small-incision cataract surgery and vitreous cutting devices, the success rate has of these procedures dramatically increased. Pars plana vitrectomy and lensectomy, combined with aphakic contact lens wear or IOL implantation, are viable surgical options. CTRs, MCTR, and CTSs have provided the opportunity to perform small-incision lens aspiration and in-the-bag implantation of a PC IOL. Combined with appropriate amblyopia therapy and good surgical technique, the prognosis for significant and safe visual rehabilitation of the child with EL is satisfactory. Annual ophthalmologic evaluation for the progression of EL, cataract, glaucoma, and RD is essential.

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Pediatric Cataract Surgery in Eyes with Uveitis

Rupal H. Trivedi and M. Edward Wilson

Cataract is a frequent complication of childhood uveitis.¹ It may develop secondary to long-standing inflammation or corticosteroid treatment. Pediatric cataract surgery (even for nonuveitic cataract) is surgically challenging and associated with a higher risk of postoperative inflammation and complications in comparison to routine adult cataract surgery. Cataract surgery in eyes with uveitis poses additional surgical challenges because of preexisting intraocular inflammation, band keratopathy, anterior and posterior synechia, and a small pupil. Despite our best effort, uveitis patients are also at a much higher risk for many adverse events that occur after the surgery is completed. These include recurrent band keratopathy, uncontrolled glaucoma, recurrent inflammatory membranes, hypotony, and cystoid macular edema (CME).

INCIDENCE OF CATARACT

An early tabulation of complications in pediatric uveitis, from 1965, found cataract in 40% (26/65).² The cataract risk has been reported to be highest for panuveitis followed by anterior, intermediate, and posterior uveitis (risk ratio, 1.06, 1.00 reference category, 0.99, and 0.27, respectively, $P = 0.007$).³ In a study of 51 juvenile rheumatoid arthritis (JRA)-associated iridocyclitis patients,⁴ cataract was noted in 46%. Of note, the term juvenile idiopathic arthritis (JIA) is preferred over JRA.⁵⁻⁹ Herein, we use the term JIA preferentially even if the article quoted used the older JRA term in the published manuscript. Foster and Barrett¹⁰ reported that even with aggressive therapy to control inflammation, 18% of patients with JIA-associated anterior uveitis developed cataracts. Narayana et al.¹¹ studied the patterns of uveitis in children at a referral eye care center and reported that approximately 25% (8/31) developed cataracts. de Boer et al.¹² reported cataract in 35% (43/123) of children with uveitis. In eyes with Behçet disease, a 47% incidence of cataract in the involved eye has been reported.¹³ Cataract was reported in 53% of 148 pediatric patients from the uveitis service of a university referral center.³ On the

basis of the number of events and total follow-up in the cohort, cataracts would be predicted to occur at a rate of 0.16 events per patient-year of follow-up.³ It was further noted that cataract was diagnosed in 11%, 15%, 24%, 39%, 44%, 56%, and 68% of patients after 3, 6, 12, 24, 36, 60, and 120 months, respectively, of the diagnosis of uveitis. Kalinina Ayuso et al.¹⁴ reported that cataract was present at initial presentation in 3% (1/35) of the eyes with intermediate uveitis, while during follow-up, cataract surgery was required in 26% (9/35) of the eyes. The mean time to the development of cataract was 2.6 years. Cataract surgery was required more frequently if the age of uveitis onset was younger than 7 years as compared to older than 7 years (44% versus 11%). Overall, cataract or cataract surgery has been reported in 18% to 68% of pediatric patients with uveitis.

TYPE OF UVEITIS

Tugal-Tutkun et al.¹⁵ reviewed the records of 130 consecutive children with uveitis and noted that the cataract was most common in JIA-associated uveitis with an incidence of 71%, followed by 50% in pars planitis, 34% in idiopathic uveitis, and 28.5% in the remaining types of uveitis. BenEzra and Cohen¹⁶ noted that of 17 children with uveitis, eight (all girls) had chronic uveitis associated with JIA, one girl had *Toxocara*, and in eight children (five boys and three girls) no systemic manifestations or definite cause was detected. The children with JIA-associated uveitis had an earlier and more severe ocular disease, leading to the need for cataract surgery when 3 to 8 years old (mean age, 5 years).¹⁶ Children with idiopathic uveitis had a more chronic and less severe type of ocular disease, leading to the need for surgery when older (mean age, 12.8 years).¹⁶ Paikos et al.¹⁷ noted that five of seven pediatric uveitis patients with cataract had JIA and the other two had chronic anterior uveitis of unknown etiology. Quinones et al.¹⁸ noted 21/34 children had JIA, seven had pars planitis, and six had other conditions (herpes zoster virus uveitis, idiopathic anterior uveitis, idiopathic

panuveitis, or sarcoid panuveitis). Terrada et al.¹⁹ reported that uveitis was JIA associated in 9/16 children (56%); idiopathic in four (25%); and associated with Behçet disease, sarcoidosis, and varicella zoster in one patient each.

As seen above, JIA-associated uveitis is the most common cause of cataract in pediatric uveitis. It has also been observed that children with JIA-associated uveitis are younger, demonstrate an active intraocular inflammation for an extended period after surgery, and tend to develop secondary membranes postoperatively, necessitating additional surgical interventions.¹⁶ The relative risk for the development of band keratopathy, cataract, and posterior synechia was highest for JIA.³ Children in whom the diagnosis of uveitis was the initial manifestation of JIA had a significantly shorter mean uveitis to cataract interval than did children in whom arthritis preceded uveitis (3.5 versus 6.6 years).²⁰ Nemet et al.²¹ reported that children with JIA-associated uveitis were seen and underwent cataract surgery at an earlier age, and had a lower preoperative visual acuity and more severe uveitic complications when first seen as well as after surgery, than those with non-JIA-associated uveitis.

TYPE AND LATERALITY OF CATARACT

Terrada et al.¹⁹ noted that 21/22 eyes with uveitic cataract had moderate to severe posterior subcapsular cataract (PSC) and one eye had a total cataract. Although uveitis is usually a bilateral disease, the severity of inflammation and cataract is often asymmetrical. The percentage of cases with cataract that were bilateral at diagnosis was 41%.³ Lundvall and Zetterstrom²² reported four patients who underwent unilateral surgery and three who had bilateral surgery. Nemet et al.²¹ reported that only 1/18 patient required bilateral cataract surgery during follow-up. The median time from involvement of the first eye to involvement of the second eye cataract was 81 months in cases of bilateral pediatric uveitis.³

CAUSE OF CATARACT

As mentioned previously, recurrent long-standing inflammation and corticosteroid use are the most common causes of cataract in eyes with pediatric uveitis. Sallam et al.²³ reported on the use of intravitreal triamcinolone acetonide for treating pediatric CME secondary to non-infectious uveitis and concluded that steroid-induced cataract occurred in 6/199 phakic eyes (55%). Thorne et al.²⁴ (while reporting a series of JIA-associated uveitis) reported that topical corticosteroid use was associated with an increased risk of cataract formation independent of active uveitis or the presence of posterior synechia. The incidence of new-onset cataract was 0.04/eye-year. There was a dose-dependent increase in the rate of cataract development among eyes receiving topical

corticosteroids. The incidence of cataract was 0.01/eye-year for eyes treated with ≤ 3 drops daily and 0.16/eye-year ($P = 0.0006$) for eyes treated with > 3 drops daily. Among eyes receiving ≤ 2 drops daily, the incidence of cataract was 0/eye-year. The presence of posterior synechia, active uveitis, and use of topical corticosteroids at presentation were significantly associated with cataract development after controlling for confounding variables. As stated above, the use of topical corticosteroids was associated with cataract formation independent of uveitis activity. Using longitudinal data analysis and controlling for the duration of uveitis, the presence and degree of active uveitis, and the concomitant use of other forms of corticosteroids in a time-updated fashion, treatment with ≤ 3 drops daily of topical corticosteroid was associated with an 87% lower risk of cataract formation compared with eyes treated with > 3 drops daily (relative risk, 0.13; $P = 0.02$).

ASSOCIATED OCULAR PATHOLOGY

BenEzra and Cohen¹⁶ reported that four of the nine eyes in children with JIA and cataract had a high intraocular pressure (IOP), and all had corneal involvement (mostly band keratopathy), extensive posterior synechia, a poor fixation pattern, and strabismus. Although band keratopathy can occur at any age, it is a hallmark of childhood uveitis. If it is present in a child with cataracts who is not a candidate for IOL implantation, it may preclude successful aphakic contact lens wear. Lundvall and Zetterstrom²² reported preoperative glaucoma in 3/10 eyes. Petric and colleagues noted glaucoma in 3/7 eyes before cataract surgery. All three eyes underwent combined cataract and trabeculectomy with mitomycin C.²⁵ Terrada et al.¹⁹ noted band keratopathy in 9/16 cases (41%) and posterior synechia in 18 cases (82%). Trabeculectomy was performed in 1/22 (4.5%) eyes before cataract surgery.¹⁹ Quinones et al.¹⁸ noted that 17/41 (42%) eyes had intraoperative posterior segment pathology—vitreous hemorrhage (1), vitreous debris or inflammation (5), fibrovascular membrane at the pars plana or retina (7), optic disc pallor/atrophy (1), papillitis (5), epiretinal membrane formation (4), CME (2), macular pseudohole (1), foveal scarring (1), and retinal vasculitis (6). Sijssens et al.²⁰ reported that the presence of adherent posterior synechia at the time of the diagnosis of uveitis is strongly associated with the early development of cataract requiring surgery in JIA-associated uveitis. More intensive screening in the early phase of JIA may result in less frequent development of posterior synechia and therefore less early development of cataract.

TIMING OF SURGERY

Cataract extraction and other elective surgeries should be deferred until inflammation is at its best level of control for at least 3 to 4 months consecutively.²⁶ Some parents

may request that surgery be scheduled to coincide with summer vacations or holidays, to minimize the child's time away from school.²⁶ Such accommodations should be made only if inflammation is controlled at that point in time. Deferral of surgery may require that children have special educational arrangements, such as low vision aids or tutors, until surgery can be performed. The medical advantages of deferral should be weighed against the risks of amblyopia in very young patients with unilateral cataracts.²⁶

It may not be possible to completely inhibit all uveitis activity. However, every effort should be made to reduce inflammation prior to surgery. Inflammation needs to be in remission, for example, 10 cells or less in the slit-lamp high-power field in the anterior chamber (or 1+), or the flare value obtained with laser photometry under maximal topical and/or systemic therapy must be at the minimum before cataract surgery can be planned. Low IOP, cells in the vitreous body, and thickening of the choroid may also represent signs of inflammation.

Treatment with methotrexate in the first year after the diagnosis of uveitis is associated with a delay in cataract surgery.²⁰ The presence of posterior synechia may be an indication for early treatment with methotrexate because this drug has been associated with a delay in the development of visually significant cataracts requiring surgery.

PREOPERATIVE ANTI-INFLAMMATORY MEDICATION

Anti-inflammatory medication is commonly instituted prior to surgical intervention. Usually, the application of topical corticosteroids, such as prednisolone acetate 1% or dexamethasone 0.1%, five times daily for 1 week in addition to the individual treatment regimen is generally sufficient. A systemic corticosteroid application is indicated in selected cases with previous or current CME, with intermediate or posterior uveitis or with known attacks of severe inflammation after previous intraocular surgery. Although the optimal preoperative dosages are not well defined, prednisone 1 mg/kg body weight given for 3 days or intravenous methyl prednisone injections on the day before surgery may be effective. Patients with severe chronic uveitis exhibiting vision threatening complications should receive immunosuppressants in advance of surgery. This should be comanaged with a pediatric rheumatologist. The value of topical and/or systemic nonsteroidal anti-inflammatory drugs (NSAIDs) for the preoperative management of uveitic inflammation is not well known. IOP should be stabilized before surgery. However, miotic drugs before surgery are discouraged because of their tendency to disrupt the blood-aqueous barrier resulting in an increased rate of postoperative fibrin formation and because of the small pupil size that may make surgery more difficult and that may increase

the bleeding rate. Since many of the uveitis patients are under long-term immunosuppressive treatment, additional measures to prevent postcataract surgery infections should also be considered. A reactivation rate of 36% for ocular toxoplasmosis within 4 months of cataract surgery has been reported, and thus, prophylactic treatment with antiparasitic drugs may be considered for patients with ocular toxoplasmosis. The frequency and intensity of uveitic recurrences prior to cataract surgery help predict the likely postoperative course.

SURGICAL TECHNIQUE

The current trend is to perform lens aspiration with posterior capsulectomy plus anterior vitrectomy (with or without IOL implantation).¹⁷⁻²⁷ The most important general principle for the surgery is to minimize intraoperative trauma. The implantation of IOLs in children with uveitis has been controversial, with valid arguments both for and against their use.²⁶ Visual rehabilitation with aphakic contact lenses can be difficult, especially in young children, because of poor compliance or intolerance. Contact lens wear may be impossible in patients with severe band keratopathy, and there may be an increased risk of contact lens-related infections in patients who require chronic topical corticosteroid therapy. In the past, most uveitis specialist considered IOL implantation to be contraindicated in children with JIA-associated uveitis because IOLs were often seen to serve as scaffolding upon which inflammatory membranes form and re-form. Most pediatric cataract surgeons remain cautious about implanting IOLs in children with JIA-associated uveitis. IOL implantation in children with non-JIA uveitis characteristically has better outcomes than in those with JIA.

Despite continued caution, increasing evidence is now available that the appropriate use of IOLs in carefully selected uveitis patients may be helpful for better visual rehabilitation even when associated with JIA or when simultaneous trabeculectomy is done.²⁸ BenEzra and Cohen¹⁶ evaluated the use of IOLs in children with both JIA-associated and non-JIA-associated uveitis. The authors concluded that IOL implantation seems preferable to correction with contact lenses in young children needing surgery in one eye. They found that their tolerance of contact lenses tended to be poor and resulted in a high rate of strabismus on long-term follow-up.

Sijssens et al.²⁹ reported ocular complications in aphakic and pseudophakic eyes of children with JIA-associated uveitis. There were some differences in presurgical characteristics between the groups. The pseudophakic group had a significantly longer time interval between the diagnosis of uveitis and cataract extraction (3 versus 1.6 years), had more ocular hypertension (66% versus 26%) and secondary glaucoma, more intraocular surgeries, and significantly more children were treated with

methotrexate. The number of complications after cataract extraction, including new onset of ocular hypertension and secondary glaucoma, CME, and optic disc swelling, did not differ between aphakic and pseudophakic eyes. Moreover, no hypotony, perilenticular membranes, or phthisis was encountered in the pseudophakic group. Sijssens et al.²⁹ noted that relative contraindications for IOL implantation in patients with JIA are age <4 years, hypotony, IOL-related complications in the fellow eye, and shallow anterior chamber. The American Academy of Ophthalmology's preferred practice pattern (for adult eyes) document noted that leaving the eye aphakic may be considered in severely damaged uveitis eyes with extensive pupillary or ciliary membrane formation or signs of intractable inflammation such as hypotony and severe flare.³⁰

There is also controversy over the best type of IOL in cases of uveitis. IOL material does not seem to be a major influence on the course of postoperative inflammation.³⁰ Foldable lenses have been evaluated in adult patients and found to be safe, but the authors concluded that the optimal biomaterial for these uveitic eyes has yet to be found.³¹ The use of heparin surface–modified (HSM) polymethyl methacrylate (PMMA) IOLs has also been advocated in these high-risk cases. It has been suggested that implantation of HSM IOLs in cataract patients with chronic uveitis may decrease the number and severity of deposits on the surface of the IOL. However, in a clinical study on adult patients with inactive uveitis or diabetes,³² no statistically significant difference was found between HSM and uncoated PMMA IOLs in the number of cellular deposits found on the anterior IOL surface, the number of adhesions between the iris and the IOL, or the incidence of capsular opacification. Similar results were seen in another study of similar design comparing HSM and uncoated PMMA IOLs in eyes with uveitis.³³ However, these results may not be strictly applicable to children with uveitis where the inflammation is much more severe. Rauz et al.,³¹ while reporting a series of adult patient to evaluate various foldable IOLs (acrylic,

silicone hydrogel) in patients with uveitis, reported that there was no association between posterior capsule opacification (PCO) and the various lens biomaterials. Another study of adult patients³⁴ compared hydrophilic acrylic (Hydroview, Bausch & Lomb), hydrophobic acrylic (AcrySof® MA60BM, Alcon Inc.), silicone (CeeOn 911, Abbott Medical Optics, Inc.), hydrophilic acrylic (Injectacryl F3000, OphthalMed), and hydrophilic acrylic with heparin surface modification (BioVue 3, Ophthalmic Innovations International, Inc.). There were no significant differences in anterior chamber flare results between the five groups. The Hydroview group had the highest grade and the CeeOn IOL and AcrySof® groups had the lowest grade of PCO. The BioVue3 and Injectacryl IOLs had good uveal biocompatibility.

INTRAOPERATIVE CONSIDERATIONS

There are many challenges during pediatric cataract surgery in eyes with uveitis. First, associated ocular complications such as synechia may make the surgery difficult and the pupil may not dilate well (Figs. 38.1 to 38.4). Similarly, the associated band keratopathy may impair visualization (Fig. 38.3A and 38.4). There is also a greater tendency toward bleeding during surgery. Fan et al.³⁵ noted that although band keratopathy can be removed surgically using a 10% solution of ethylenediaminetetraacetic acid (EDTA), excimer phototherapeutic keratectomy also has the added value of improving the stability of the tear film and the ocular surface. Use of the excimer laser requires that cataract surgery be deferred until corneal surface healing is complete.³⁵

Adjunctive corticosteroids at the time of surgery (intravenous, periocular, or intraocular) may be considered. Corneal incisions are advantageous in these eyes as they may require glaucoma surgery in the future. Although the pupil may dilate poorly in eyes with uveitis, excessive iris manipulation should be avoided so as not to exacerbate inflammation and stimulate new posterior synechia

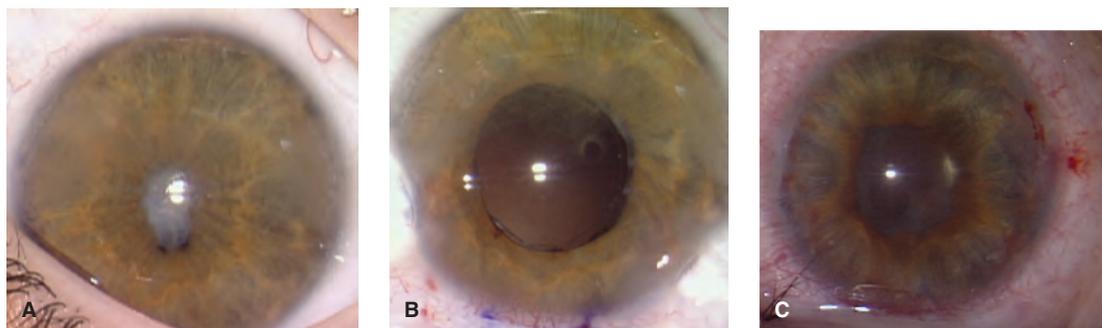


Figure 38.1. Cataract secondary to JIA-related uveitis in a 7-year-old child. Band keratopathy removed 2 months before cataract surgery. **A:** Preoperative photograph showing nondilated oval pupil secondary to iridolenticular synechia. **B:** In-the-bag fixation of a single-piece AcrySof® IOL. **C:** Iris bombe and recurrence of posterior synechia; photograph taken 6 months after cataract surgery.

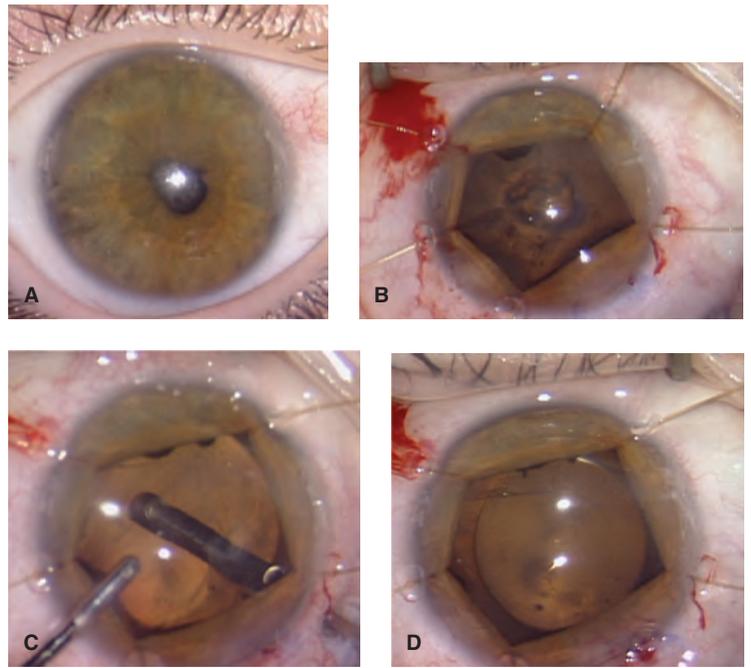


Figure 38.2. **A:** Cataract secondary to uveitis associated with a small pupil. The pupil failed to dilate after synechiolysis. **B–D:** Iris retractors were used to facilitate visualization during surgery.

formation.³⁰ The least iris manipulation consistent with adequate pupillary access should be the aim. Blunt dissection with ophthalmic viscosurgical device (OVD) is preferred for synechiolysis, and sharp dissection should be minimized. More adherent posterior synechia can be

eliminated using an iris spatula under OVD. Occasionally, the firmly fixed adhesions may need to be dissected with fine scissors, however. In more severe cases, better mydriasis can be obtained using iris hooks placed in each quadrant through small corneal incisions (see Fig. 38.2B–D)

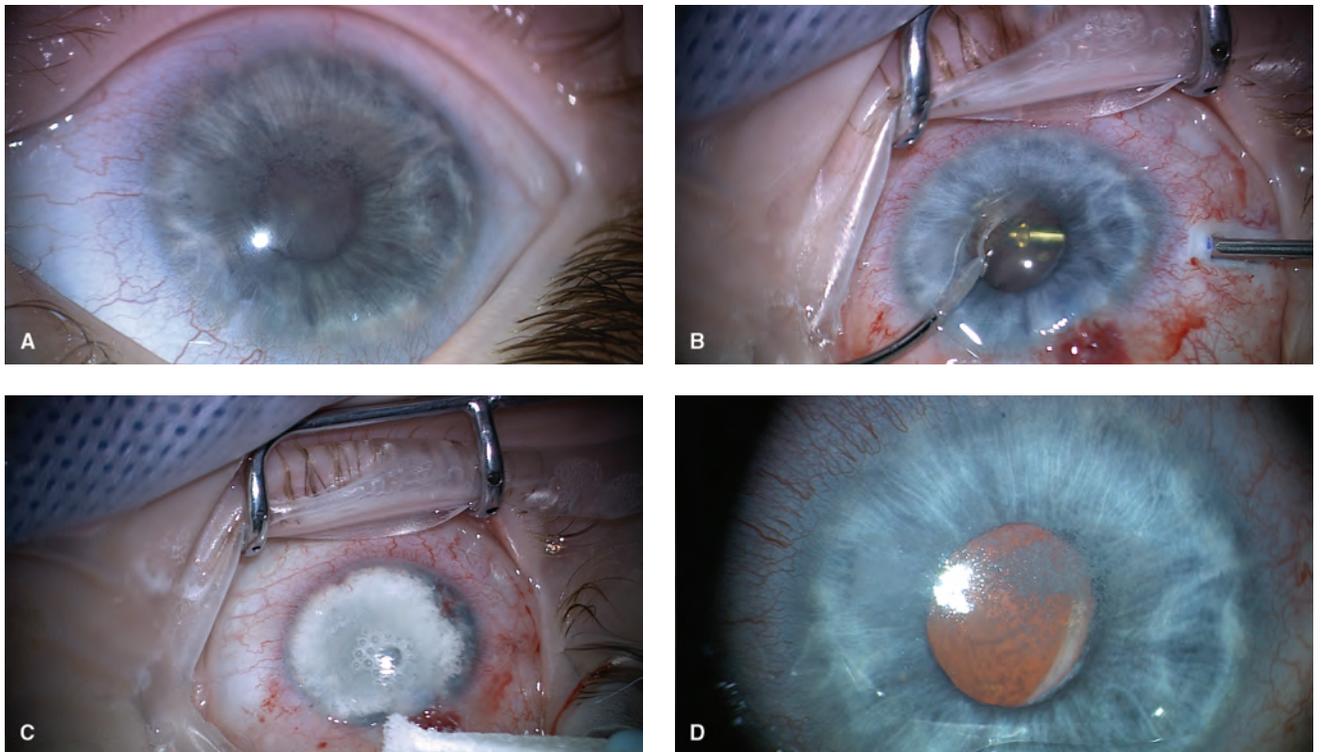


Figure 38.3. **A:** Preoperative view of right eye in child aged 4 years. **B:** Pars plicata posterior capsulectomy and vitrectomy after IOL implantation. **C:** Intracameral Triescence. **D:** Three months after cataract surgery. Note that chelation with EDTA was done at this visit.

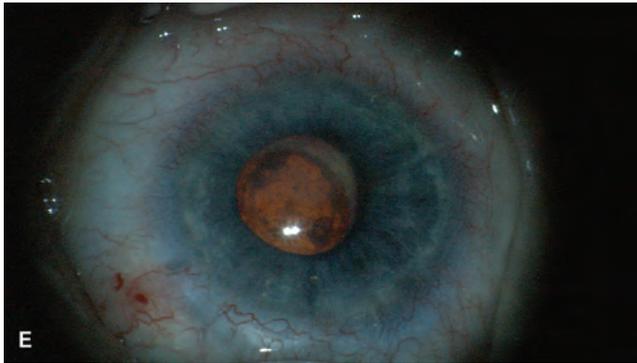


Figure 38.3. (Continued) **E:** Surgical removal of visual axis opacification was required 1 year after cataract surgery.

or by stretching maneuvers with surgical hooks under OVD. Intraoperative hyphema may occur but can be managed with the injection of OVD. Rarely, wet-field cautery is required. A large capsulorhexis reduces the risk of synechia formation between the iris and the anterior capsular margin.

Alkawas et al.³⁶ reported that intraoperative intravitreal injection of triamcinolone acetonide was effective in controlling postoperative inflammation for adult cataract extraction in patients with uveitis. Li et al.³⁷ reported on the outcomes of cataract surgery in children with JIA. Triamcinolone acetonide was injected into the anterior chamber at the end of surgery in one group, while the other group received an intraoperative intravenous injection of methylprednisolone and postoperative oral prednisolone. The authors reported that fibrin formation was not seen after surgery in the triamcinolone acetonide group but occurred in five patients in the systemic treatment group. Additional systemic corticosteroids were not required in the triamcinolone acetonide group. No increase in IOP was noted after the intracameral triamcinolone acetonide injections. The authors concluded that an intraoperative intracameral injection of



Figure 38.4. Left eye of the same child reported in Figure 38.3. The child presented with uveitis glaucoma. Cataract surgery with pars plana posterior capsulectomy and vitrectomy was performed.

4 mg of triamcinolone acetonide may be more effective than intraoperative intravenous methylprednisolone and additional postoperative short-term oral prednisolone in preventing postoperative fibrin formation after cataract surgery in patients with JIA and uveitis.

We routinely inject 0.1 cc (4 mg) of Triescence (preservative-free triamcinolone, Alcon, Fort Worth, TX) into the anterior chamber at the conclusion of cataract surgery in children with uveitis (Fig. 38.3C).

In most cases, placement of the IOL haptics into the capsular bag is preferred; however, sulcus fixation of the haptics may allow the IOL to block the formation of iridocapsular adhesions in high-risk eyes (e.g., extensive iris damage or preoperative posterior synechia). This technique does not seem to increase postoperative inflammation. Anterior chamber IOLs may stimulate more inflammation and may be problematic if the angle anatomy is compromised.³⁰

POSTOPERATIVE TREATMENT

The postoperative anti-inflammatory treatment should be adjusted according to the surgical maneuvers and the observed inflammatory activity seen postoperatively. The gold standard is to increase the corticosteroid dosage postoperatively compared to the treatment level that the patients had before surgery. The dosages must be adapted to the inflammation. It is especially important that the increased dose be continued for 8 to 12 weeks after surgery. Tapering off the dosages too early is followed by an increased risk for hypotony, posterior synechia, IOL cell deposits, or CME. Some authors consider giving systemic steroid postoperatively.²¹ Postoperative use of short-acting topical mydriatics agents may help to prevent postoperative synechia formation; however, fixed dilation with long-acting cycloplegic agents such as atropine may lead to formation of posterior synechia in the dilated state.³⁰

POSTOPERATIVE COMPLICATIONS

Various postoperative complications may occur, such as recurrence of uveitis, which may even be of the severe fibrinous type, macular edema, and hypotony, even leading to hypotonus maculopathy. In a series of seven patients with JIA who underwent cataract extraction and IOL implantation, Probst and Holland³⁸ described less favorable results among children compared with adults. The major complications included membrane and synechia formation as well as elevated IOP. Quinones et al.¹⁸ reported hyphema, vitreous hemorrhage, iris bombe, cell deposits on the IOL, hypotony, and reactivation of uveitis during the early postoperative period. Reactivation of uveitis was the most frequent early complication, followed by hyphema and hypotony. They further

reported new-onset glaucoma, PCO, retinal detachment, CME, and pupillary membrane during the later postoperative period. PCO (Fig. 38.3E) and CME were the most frequent complications followed by new-onset glaucoma and retinal detachment. Nemet et al.²¹ reported that there was no significant difference in postoperative course or complication in eyes with JIA-associated uveitis to those without JIA-associated uveitis.

BenEzra and Cohen¹⁶ noted persistent intraocular inflammation after surgery in 2/20 eyes. Five eyes (17%) experienced postoperative reactive uveitis, which was controlled by modifying their current regimen.³⁹ Fibrinous uveitis should initially be treated with intensive topical steroids, but if it persists, an intracameral injection of recombinant tissue plasminogen activator (rtPA) should be considered. It has been suggested that intracameral rtPA may be used for fibrinolysis after cataract surgery in children when severe fibrin formation is seen despite intensive topical steroid therapy.⁴⁰ Ten micrograms of rtPA was applied 7.18 ± 2.04 days after intraocular surgery. Complete resolution of fibrin formation occurred in 90% of the eyes, and in these cases, no recurrent fibrinous reaction or adverse effects were noted. However, fibrin clot dissolution was incomplete in two eyes, in the same patient, with a history of JIA and chronic uveitis.

The rate of visual axis opacification requiring a secondary procedure has been reported as 100% by Probst and Holland,³⁸ 83% by Lam et al.,²⁸ 70% (7/10) by Lundvall and Zetterstrom,²² 80% by BenEzra and Cohen,¹⁶ and 56% by Urban and Bakunowicz-Lazarczyk⁴¹. Nemet et al.²¹ reported anterior and posterior capsular opacification, which occurred in 11 (85%) of the 13 patients in whom the posterior capsule was left intact after surgery. Six of these children required Nd:YAG laser capsulotomy, and three others later needed surgical posterior capsulectomy. Of the six patients who underwent anterior vitrectomy and posterior capsule removal at cataract extraction, none required further capsulotomy; in two children, however, pupillary membranes developed. Of the six eyes that underwent anterior vitrectomy during cataract surgery, three continued to have significant inflammation compared with four of the 13 eyes that did not undergo anterior vitrectomy. However, the authors attributed this to a more active disease in the affected patients who required primary capsulectomy and vitrectomy, rather than an adverse effect of the procedure. Quinones et al.¹⁸ reported PCO requiring YAG in 4/41 eyes (with or without IOL). Terrada et al.¹⁹ noted PCO requiring laser capsulotomy in 4/22 eyes, the mean time between cataract surgery and laser treatment was 14 months. One eye required a second Nd:YAG laser treatment. In many uveitis patients, multiple Nd:YAG laser capsulotomy treatments are necessary.³⁹ Lundvall and Zetterstrom²² reported one patient where five reoperations were required.

Posterior synechia developed in 100% and 66%, respectively, of eyes with uveitis and IOL in the studies by BenEzra and Cohen¹⁶ and Probst and Holland.³⁸ In contrast, Lam et al.²⁸ did not find any posterior synechia. Terrada et al.¹⁹ noted deposits on the IOL in 3/22 eyes.

The incidence of glaucoma has been reported as 56%,¹⁶ 60%,²² 83%,²⁸ 100%,³⁸ and 23%.¹⁹ Terrada et al.¹⁹ noted glaucoma in 3/22 eyes severe enough to require trabeculectomy. In another study, 10% of the eyes developed glaucoma that required the insertion of an Ahmed valve at an average of 28.3 ± 32.6 months after surgery.³⁹

Severe band keratopathy can be treated with chelating agents (Fig. 38.3D) or the excimer laser can be used to clear the cornea. However, if any residual inflammation is present, the deposition of calcium tends to recur.

Terrada et al.¹⁹ noted CME/macular dysfunction in 3/22 eyes. One case of Behçet-associated uveitis and cataract surgery had a poor visual outcome secondary to preoperative retinitis scars. Another patient with idiopathic uveitis developed a retinal detachment related to inflammation and ultimately ended up with a poor visual outcome.

Kanski (in a series of combined adult and pediatric patients with juvenile chronic iridocyclitis, average age 12.7 years, range 3 to 40 years) noted that the incidence of postoperative phthisis was related to the level of preoperative IOP. IOP <10 mm was noted in 29 eyes. Seven of 29 (24%) eyes became phthisical, usually within a few weeks of surgery. Seven of 158 (4%) eyes that were not hypotonous at the time of surgery subsequently developed phthisis. The overall incidence of phthisis was 14/187 (8%).⁴²

IOL explantation is occasionally necessary in children with persistent uveitis. The use of IOLs in uveitis may, in some cases, lead to increased inflammation compared to preoperative levels, and the IOL may need to be explanted. In these cases, IOL explantation may salvage or stabilize the vision, provided that inflammation has not resulted in irreversible damage to the macula or optic nerve prior to explantation.⁴³ Adan et al.⁴⁴ reported two cases of explantation of an IOL in children with JIA-associated uveitis. The preoperative conditions in the two patients were similar: lack of control of inflammation before cataract surgery, oligoarticular ANA positive subgroup of JIA, and younger than 10 years.

VISUAL OUTCOME

Many children do not attain good vision after surgery owing to complications such as glaucoma and macular disease (chronic CME, macular hole, hypotony maculopathy, and recurrent macular pucker)⁴⁵ and other serious problems such as severe postoperative uveitis and phthisis bulbi.²⁷ However, Lam et al.²⁸ reported that postoperative Snellen visual acuity of 20/40 or better was achievable. Nemet et al.²¹ reported that visual acuity improved by two

or more lines in all patients, and in 13/19 eyes, the final visual acuity was 20/40 or better. Quinones et al.¹⁸ noted that most patients (88%) who received immunomodulatory therapy attained better vision, but this was not statistically significant compared with those who did not ($P = 0.47$). Terrada et al.¹⁹ noted that at the 5-year visit, the corrected distant visual acuity remained better than preoperatively in 19/22 eyes, had stabilized to preoperative levels in two eyes, and was worse than preoperatively in one eye. Postoperative visual acuity in JIA-associated uveitis was 20/90 at 1 year after surgery. The majority of patients with poor postoperative visual acuity had preexisting retinal or optic nerve pathology. Ninety-three percent (28/30) of eyes showed improvement or maintenance of visual acuity, whereas 7% (2/30) of the eyes worsened.³⁹

SUMMARY

The treatment of pediatric patients with chronic uveitis requires a multidisciplinary approach. Ophthalmologists should work with pediatric rheumatologists to exclude or manage systemic rheumatologic disease and to ensure the safe use of systemic immunomodulation in these children. Surgical treatment should be considered only when the uveitis is controlled medically. Multistage surgery may be necessary to address each complication of the uveitis using diverse strategies and appropriate surgical techniques.

Pediatric cataract surgery in eyes with uveitis may be associated with increased ocular morbidity owing to the greater technical difficulty during surgery and increased intraocular inflammation in the postoperative period. It is advisable to refer such complicated patients to an experienced pediatric cataract surgeon. It is important to adopt strategies before, during, and after such surgery to minimize the complications. Standard surgery with IOL implantation may be feasible if preoperative anti-inflammatory medications are used and the inflammatory condition is inactive. Postoperative control of inflammation and long-term follow-up is mandatory.

Pharmacologic agents intraoperatively and postoperatively must be customized to the individual patient and titrated based on frequent postoperative examinations. Intracameral nonpreserved triamcinolone should be strongly considered in these patients. We recommend caution when implanting IOLs in children with JIA-associated cataracts unless the inflammation is very well controlled preoperatively. Cataract associated with non-JIA uveitis in children can usually be managed with IOL implantation without the frequent complications seen in JIA.

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Retinopathy of Prematurity

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Retinopathy of prematurity (ROP), formerly known as retrolental fibroplasia, is a proliferative disease of premature infants characterized by abnormal blood vessel development in the retina. With the continued improvement in survival of low birth weight and early gestational age infants, ROP remains a significant cause of childhood blindness. Cataract can occur as a complication of the disease or the treatment. Cataract can be an associated ocular pathology in eyes with ROP, as low birth weight and prematurity are risk factors for both entities.^{1,2} Alden et al.³ reported transient lens opacities in 3% of low-birth weight infants. These opacities, which are reversible in nature, are characterized by clear fluid vacuoles just anterior to the posterior capsule of the lens. Marcus et al.⁴ reported hereditary cataract developing in a premature infant after birth. The authors noted that very early cataract extraction may be necessary in premature infants to allow ROP evaluation. However, the occurrence of secondary cataract (secondary to treatment of ROP) has been reported more frequently in the literature and is our focus in this chapter.

Mild ROP has a negligible incidence of cataract. In more advanced disease, cataract is a known association. The Early Treatment for ROP study reported that for prethreshold ROP patients 2 years of age, cataract or aphakia was found in 4% of the treated eyes (with peripheral retinal ablation applied at prethreshold) and 6% of the conventionally managed (observed at prethreshold and only treated if threshold was reached) eyes.⁵ Patients with retinal detachment (RD) associated with ROP, that is, stage 5 ROP, have as high as a 50% risk of developing cataracts.⁶

Laser photocoagulation has long ago supplanted cryotherapy as the standard treatment for threshold ROP. Compared with cryotherapy, laser is associated with a lower rate of RD and less myopia. However, laser retinal photocoagulation in eyes with ROP may be associated with an increased risk of cataract formation. Laser therapy in ROP patients can be applied with an argon, diode, or frequency-doubled YAG laser. Argon laser-treated eyes for ROP are at a somewhat higher risk of

developing secondary cataract; however, the occurrence of cataract has also been reported in diode laser-treated eyes and less so with cryotherapy. A survey performed by Gold⁷ reported that 62% of the reported cataracts were associated with argon laser, 31% with diode laser, and 7% with cryotherapy. Sanghi et al.⁸ noted no cataracts in eyes treated with either frequency-doubled Nd:YAG laser or diode laser. Salgado et al.⁹ reported three eyes with cataracts (two requiring surgery) in a consecutive series of 259 eyes of 184 children treated for threshold or prethreshold ROP with transpupillary diode laser photocoagulation. A study of Bevacizumab in the treatment of patients with prethreshold ROP study reported cataract development in one eye (1%).¹⁰

TYPES OF SECONDARY CATARACTS

Transient

Focal opacities (either punctate or vacuolated) may occur at the capsular or subcapsular level. These are generally visually insignificant and often resolve spontaneously.¹¹

Progressive and Visually Significant Lens Opacity without Retinal Detachment

Progressive lens opacification generally leads to total cataract and complete obstruction of the visual axis.¹² Most of these eyes have had transpupillary laser treatment or “lens-sparing” vitrectomy. The latent period before cataract develops is reported as 1 to 4 weeks after laser treatment, but it can be as long as 6 months.¹³ Associated findings may include corneal edema, shallow anterior chamber, pupillary membrane, pigment on the anterior lens surface, iris atrophy, hyphema, posterior synechia, and iris neovascularization.¹³

Secondary to Retinal Detachment

A cataract develops frequently in eyes with ROP stage 4 or 5 (i.e., partial or total RD). These cataracts differ from the two types of cataracts described above in that their

onset is later. They occur months or years after RD or vitreoretinal surgery, rather than in the early posttreatment time frame. Approximately 15% of children treated by lens-sparing vitrectomy for ROP-related RD develop this type of delayed-onset cataract.¹⁴ Associated findings may include angle-closure glaucoma, shallow anterior chamber, corneal opacification, and posterior synechia.¹³ Knight-Nanan et al.⁶ reported cataracts in 55% of eyes treated for advanced cicatricial ROP.

ETIOPATHOGENESIS

Cataract formation after laser photocoagulation has been noted immediately, during the laser treatment, to as long as 99 days after the laser treatment, with most occurring in the first few postoperative weeks.

Anterior Segment Ischemia

Associated clinical findings such as iris and ciliary process atrophy^{13,15} suggest an ischemic etiology. Lambert et al.¹³ suggested that thermal injury to the long posterior ciliary arteries had resulted in anterior segment ischemia. This led to total cataract formation and often to eventual phthisis bulbi. We have seen children after photocoagulation for ROP where the entire eye appeared to have suffered an ischemic event. These eyes presented with corneal edema, iris ischemia, and cataract within 24 hours of laser therapy. In our experience, these rare cases are not associated with overly heavy treatment. They occur without warning in eyes with reportedly uneventful laser sessions. The cornea often improves in clarity but remains thickened. The cataract may also appear improved somewhat in appearance. In a few of these eyes, however, severe corneal thickening and opacity, total cataract, hypotony, and retinal ischemia have persisted. In these cases, the prognosis for visual recovery after cataract surgery is poor.

Thermal Injury

Premature infants have much of the tunica vasculosa lentis intact, which may allow for absorption of energy on the lens surface. Hazy vitreous and miosis in these infants may also necessitate higher-power settings. Paysse et al.¹⁶ theorized that postlaser cataracts are, in most cases, the result of thermal injury. This results from absorption of laser energy by lens proteins or hemoglobin in the blood circulating through a persistent anterior tunica vasculosa lentis. Thermal cataracts, for the most part, occur in the first postoperative weeks after laser treatment. This hypothesis is further supported by the lower incidence of cataract formation with diode laser compare to argon laser, as this phenomenon of laser energy absorption is likely to be less frequent because of the reduced absorption of diode laser energy by hemoglobin.

Uveal Effusion

Diode laser application in nanophthalmic eyes has been reported to cause uveal effusion that resulted in anterior rotation of the ciliary body and shallowing of the already narrow anterior chamber. The resulting corneolenticular apposition can lead to cataract formation in these eyes.¹⁷

Vitreoretinal Pathology

Cataract may be associated with chronic RD even when no intraocular surgery has been performed. In other eyes, the posterior lens capsule may have been breached at the time of vitreoretinal surgery. This can lead to lens hydration and complete cataract.

Phacoantigenic Uveitis

Lambert et al.¹³ noted iridocyclitis and posterior synechia in eyes with cataract after laser photocoagulation for ROP and theorized that phacoantigenic uveitis could have caused the cataract. They reported one patient with a rent in the posterior lens capsule at the time of cataract surgery when the only prior treatment had been laser therapy. The authors noted that the lens material was liquefied and theorized that microperforations may occur in some eyes during laser therapy.

RISK FACTORS

Argon laser-treated eyes for ROP may be at higher risk of developing secondary cataract compared with diode laser-treated eyes.

Transpupillary laser administration of laser may increase the cataract risk as opposed to a transscleral approach.

Prominent anterior tunica vasculosa lentis may increase the cataract risk. Sufficient laser energy may be absorbed by the persistent lens vasculature to cause thermal injury to the lens.

Inadvertent burns placed on the iris. Energy absorption caused by the iris pigment epithelium can cause heating of the anterior lens.

Confluent laser therapy or heavy laser treatment over the posterior ciliary vessels. A confluent laser pattern may have a higher success rate for regression of ROP than a less confluent laser pattern. However, cataract has been identified as a possible complication of confluent treatment.¹⁵ Also, extensive use of laser energy over the posterior ciliary vessels at the 3 o'clock and 9 o'clock positions may add to the risk of anterior segment ischemia and secondary cataract formation.

Vitreoretinal surgery. Cataracts occur more commonly after intraocular surgery.

REVIEW OF THE LITERATURE

Argon Laser Therapy

1. 1992. Drack et al.¹¹ reported transient punctate and comma-shaped opacities at the level of the anterior cortex/lens capsule, associated with a few posterior synechia.
2. 1995. Christiansen and Bradford¹⁸ reported a 6% incidence of visually significant cataracts and a 1% incidence of transient cataracts. The lens opacities were diagnosed between 19 and 99 days (median, 20 days) after treatment. All eyes with permanent cataract were noted to have persistent tunica vasculosa lentis at the time of treatment. After laser therapy, these eyes developed hyphema, shallowing of the anterior chamber, corneal edema, and progressive opacification of the lens. The number of laser burns was higher in eyes developing a subsequent cataract compared to eyes that did not develop a cataract (average, 1,320 and 1,126, respectively).
3. 1997. A survey conducted by Gold⁷ revealed a total of 42 cataracts in eyes treated by argon laser—22 visually significant and 20 nonsignificant.
4. 1998. O'Neil et al.¹⁹ reported 4/374 (1%) eyes with cataracts. In two of the four eyes, it was judged that the cataract was related to the laser treatment. The incidence of cataract formation in eyes with persistent tunica vasculosa lentis was not significantly higher than in eyes without it ($P = 0.057$).
5. 2000. Lambert et al.¹³ reported eight infants (10 eyes) who developed visually significant cataracts after bilateral transpupillary laser photocoagulation. Five eyes were treated with diode laser, and five eyes were treated with argon laser. Nine of the ten eyes eventually progressed to phthisis bulbi and no light perception.

Diode Laser Therapy

1. 1994. Capone and Drack²⁰ reported transient cavitory lenticular changes in two infants.
2. 1994. Pogrebniak et al.¹² published a case report of a visually significant cataract.
3. 1995. Seiberth et al.²¹ noted that no cataract formation was observed in eyes with tunica vasculosa lentis. The number of burns was $1,556 \pm 315$ in their series.
4. 1995. Campolattaro and Lueder¹⁷ reported development of a cataract in an eye with nanophthalmos; the fellow eye remained clear.
5. 1997. A survey conducted by Gold⁷ revealed a total of 21 cataracts in eyes treated by diode laser—9 were visually significant and 12 were not.

6. 1997. Christiansen and Bradford²² reported a case of bilateral visually significant cataracts that developed in a premature infant treated for threshold ROP in one eye with persistent tunica vasculosa lentis in both eyes. A total of 1,529 burns were delivered to the right eye and 1,259 to the left eye.
7. 2000. Lambert et al.¹³ reported five eyes that developed visually significant cataracts a mean of 9.6 weeks after laser treatment.
8. 2001. Kaiser and Trese¹⁵ reported seven eyes that developed cataract following confluent treatment of threshold ROP.
9. 2002. Fallaha et al.²³ reported the clinical outcome of confluent diode laser photoablation for ROP. The authors noticed postoperative cataracts in 5% of eyes. Two infants were noted to develop bilateral cataracts. One patient developed visually insignificant peripheral cortical lens opacities 6 months after laser treatment. The second infant developed dense central lens opacities 5 months after laser photoablation and underwent bilateral cataract surgery. This infant had received 1,500 laser burns in both eyes.
10. 2002. Paysse et al.¹⁶ reported a low incidence of acquired cataracts following transpupillary diode laser photocoagulation for threshold ROP in a large group of treated eyes. Only one eye (0.003%) developed small nonprogressive peripheral cortical lenticular opacities that were noted shortly after treatment. The advantage of diode over argon may be most visible in infants with significant persistent anterior tunica vasculosa lentis.
11. 2006. Kieselbach et al.²⁴ noted cataract formation in 1/37 (3%) eyes, which had received transpupillary laser coagulation.
12. 2010. Salgado et al.⁹ reported that 3 of 120 eyes treated at prethreshold were found to have cataract (two eyes of one patient require surgery) versus 0 of 139 treated at threshold.

Cryotherapy

1. 1997. A survey conducted by Gold⁷ revealed a total of five cataracts in eyes treated with cryotherapy—three were visually significant and two were not.
2. 1998. Repka et al.² reported that cataract surgery was required in 2% of the eyes with threshold ROP and was performed equally often in treated (4/235) and control (3/231) eyes. An association with cryotherapy was not found, as control eyes were as likely as treated eyes to have cataracts develop of sufficient severity to warrant surgery.
3. 2001. Shalev et al.²⁵ reported a randomized comparison of cryotherapy versus diode laser and reported no cataract in either group at 7 years of follow-up.

MANAGEMENT

Preventive

Use of longer-wavelength laser energy, such as the diode laser (810 nm), which is minimally absorbed by hemoglobin, could decrease the risk of cataract formation. This is especially true for eyes with persistent tunica vasculosa lentis.

Transscleral administration offers the advantage of decreasing the risk of cataract formation by circumventing the lens as compared with the transpupillary approach.

A near-confluent pattern of treatment may be equally effective at preventing RD (compared to confluent treatment) and less likely to result in the formation of a cataract for an infant with threshold ROP.²⁶

Lightly treating the 3 o'clock and 9 o'clock meridians with laser photocoagulation or using cryotherapy for these meridians may reduce the chances for thermal injury to the posterior ciliary vessels and lower the risk for anterior segment ischemia.¹¹

Therapeutic

A thorough retinal examination is necessary. If the retina cannot be visualized, a B-scan ultrasound is recommended (Fig. 39.1). Lambert et al.¹³ recommended a delay in cataract surgery if there are objective signs of anterior segment ischemia, because cataract surgery may accelerate the process of these eyes becoming phthisical. A visually significant cataract after laser treatment or vitrectomy for ROP is approached much like childhood cataracts in children without ROP. Central corneal thickness and intraocular pressure should be monitored since a persistently thickened cornea and hypotony may be signs of ischemia. RD can occur before or after cataract surgery in eyes with signs of ischemia. Careful serial examinations of the retina are indicated.

Intraocular lens (IOL) can be implanted unless the child is in the early months of life and has microphthalmia. For a child with ROP and cataract, slightly higher hypermetropia may be considered in anticipation of developing

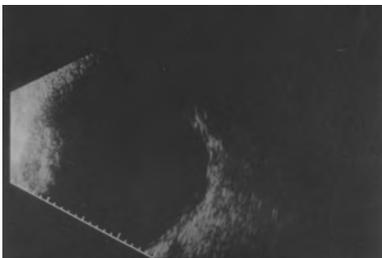


Figure 39.1. Preoperative B-scan in the left eye of a 44-month-old girl with a secondary cataract. Same child as described in the legend of Figure 39.2.

more myopia, especially if treated with cryotherapy.²⁷ The rate of myopia was 91% in a series by Smith and Tasman²⁸ evaluating adults with ROP. In these reports, globe axial length is often not reported. At times, the myopia in ROP is lenticular rather than axial. Prevalence of astigmatism at 6 years of age was reported to be higher in children who participated in the Early Treatment of ROP study. Long-term follow-up of the prevalence of surgically-induced and age-related changes in astigmatism in ROP-treated eyes is needed.

The general principles of pediatric cataract surgery described in this book should be applied to ROP-treated eyes (Figs. 39.2A–D and 39.3A–C). However, eyes with ROP may also be associated with microphthalmos, iris atrophy, glaucoma, shallow anterior chamber, and band-shaped keratopathy, which further complicates already complicated infantile cataract surgery. We have observed that these eyes are more often associated with fibrous changes in the anterior capsule (see Figs. 39.2 and 39.3). When performing anterior capsulotomy in eyes with a fibrous capsule, we try to avoid the fibrous area if possible. If that is not possible, we have found a Klotz diathermy unit or Fugo unit useful to make an opening through a fibrous capsule. It is also important to remember that sometimes the posterior capsule may have been compromised during previous surgery.¹³ Hydrodissection should be avoided if the integrity of the posterior capsule is compromised or in cases with a total cataract when the posterior capsule cannot be visualized. Extensive retinal–lenticular adhesions may be present as well.²⁹ Postoperative complications can also be treated in a similar manner as those in non-ROP children operated for cataract (Fig. 39.4).

SPECIAL SITUATIONS

Lensectomy for Angle-Closure Glaucoma

Angle-closure glaucoma is known to be associated with RD and ROP. When iridectomy and medical management fail, surgical management remains the next step. Lensectomy, which can effectively treat both pupillary and ciliary block glaucoma by lessening the anterior displacement of the iris and reducing angle closure, remains an effective means of controlling acute angle-closure glaucoma in ROP. Although visual outcome is poor, due to RD and other associated condition, lensectomy may be helpful in controlling pain and lowering IOP in eyes with a history of ROP and RD.

Cataract Surgery in Adult Eyes

There are ranges of residual ophthalmic manifestations that make it important to monitor all premature patients for a lifetime. Suspected complications of ROP that develop in adulthood include early cataract formation,

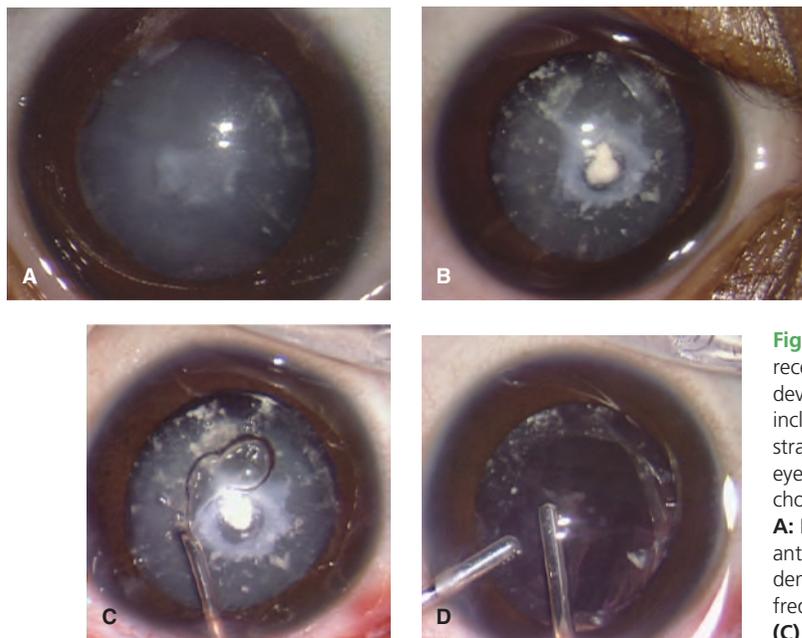


Figure 39.2. A 44-month-old girl with ROP had received laser treatment during infancy. Gradually, she developed cataracts in both eyes. Associated findings include degenerative myopia (axial length, 26 mm OU) and strabismus. The right eye also has a macular scar. Right eye fundus examination 1 week postoperatively revealed chorioretinal scarring almost up to the border of zone 1. **A:** Right eye with a total cataract. Note changes in the anterior capsule. **B–D:** Left eye with a total cataract. Note dense fibrous changes in anterior capsule (**B**). A Klotz radio-frequency diathermy unit was used to create the capsulotomy (**C**). Note changes in the anterior and posterior capsules.

early onset of glaucoma, high myopia, retinal tears, and RD.^{28,30} Kaiser et al.³⁰ reported that in patients with a history of premature birth, cataract surgery tends to be performed at a young age (median age, 40 years), has a mixed range of visual results, and can be associated with a high rate of retinal complications. Some authors have reported favorable outcomes of cataract surgery in adult eyes with ROP.^{31,32} Smith and Tasman²⁸ reported late complications of ROP in patients aged 45 to 56 years. Fourteen of the 86 eyes (16%) had a clear natural lens, and the remaining 72 eyes (84%) had a cataract or implant or were aphakic. Stated another way, 75% had cataract surgery on one or both eyes, whereas 25% retained their natural lenses. Nuclear sclerosis was the most common type of cataract, occurring in 45% of eyes that did not undergo cataract extraction. The authors concluded that there are significant late complications of ROP underscoring the importance of lifelong follow-up.

SUMMARY

Cataracts can occur with ROP as a complication of the disease or the treatment. The risk of secondary cataract was highest in argon laser-treated eyes, followed by diode laser and cryotherapy, respectively. Eyes with significant tunica vasculosa lentis undergoing laser therapy are at a higher risk for development of cataracts. Cataract formation after laser treatment has been noted most commonly in the first few weeks after laser. Preventive measures play a significant role in minimizing the risk of cataract formation in eyes with ROP. For cataract surgery and IOL implantation, the general principles of pediatric cataract surgery should be followed. However, while selecting IOL power, slightly higher hypermetropia may be considered in anticipation of developing more myopia, especially if treated with cryotherapy.

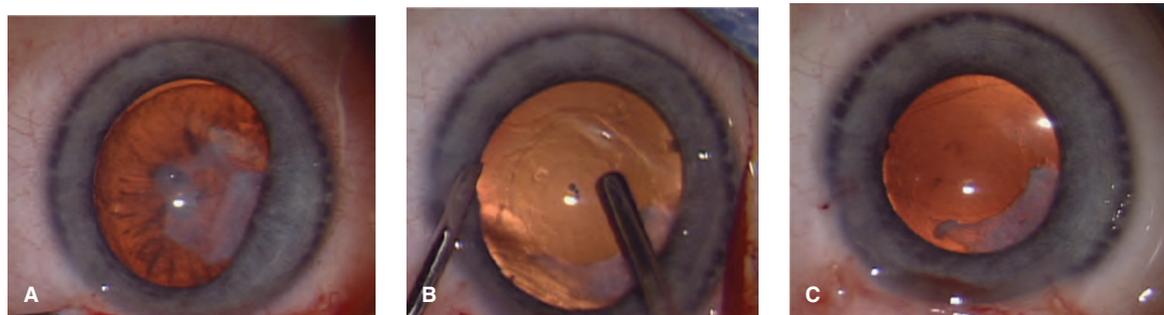


Figure 39.3 A–C: A 3.5-year-old girl who was born premature and developed ROP. The right eye developed inoperable RD and became phthisical. The left eye underwent laser therapy and developed a visually significant cataract. Note fibrous changes in anterior capsule. This eye received a 14.5-diopter SA60 (Alcon, AcrySof®) IOL implantation.

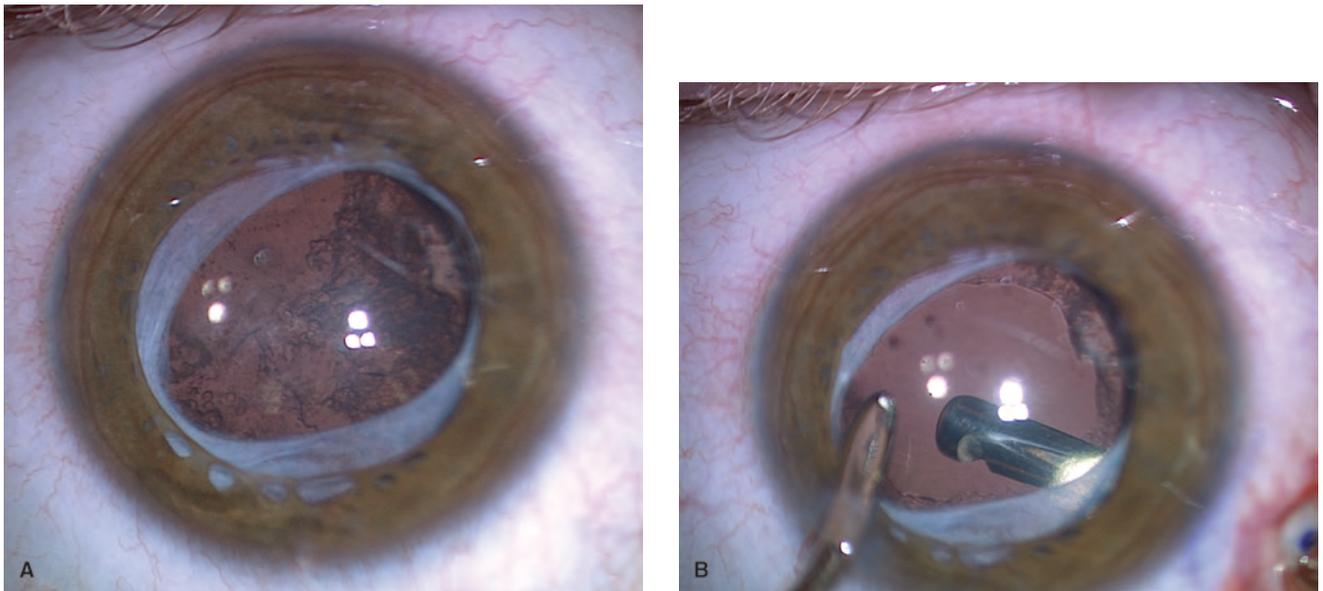


Figure 39.4. A 11-year-old girl with a history of treatment of ROP underwent both eye cataract surgery with primary in-the-bag Rayner C-flex IOL implantation. Posterior capsule was left intact at the time of cataract surgery. Iris abnormalities were noted in both eyes at the time of cataract surgery. At 1-year postoperative period, both eyes developed visually significant cataract. The child was not cooperative for YAG laser capsulotomy and needed surgical posterior capsulectomy and vitrectomy in both eyes. **A:** Left eye visually significant cataract. **B:** Removal of visually significant cataract using pars plicata approach.

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Pediatric Radiation-Induced Cataracts in Retinoblastoma

Rupal H. Trivedi and M. Edward Wilson

Retinoblastoma (RB) represents the most common primary intraocular malignancy in children. Historically, bilateral disease was managed by enucleation of the more involved eye and globe-conserving therapy for the less involved eye. Unilateral tumors generally were treated with enucleation. Treatment has evolved to globe-conserving therapies for both eyes in the setting of bilateral disease and in select cases of unilateral disease. Chemoreduction in combination with local ablative therapies has become the standard of care in lieu of external beam radiotherapy (EBRT) because of radiation-related complications (e.g., cataract). However, EBRT continues to be applied as “salvage” treatment for eyes failing chemoreduction therapy or in the setting of extraocular extension and orbital disease.

The major ocular complication of EBRT is radiation-induced cataracts. The crystalline lens is the most radiosensitive structure in the eye, with doses <2 Gy causing cataract formation. Recent studies indicate that the threshold for cataract development is less than was previously estimated and is approximately 0.5 Gy.¹ The treatment of RB requires doses in excess of 40 Gy, so cataract formation is an expected side effect.² Cataracts impair both the visual development of the child and the ability of the ophthalmologist to examine the eye. Radiation-induced cataracts can be formed as these rays penetrate the cornea and interact with the molecular components of the lens. Kase et al.³ showed that growth factors produced by RB cells may lead to cataract formation. The RB cells lack cohesion. Therefore, detached fragments of tumor are easily separated from the main mass, spread throughout the globe, and seed themselves onto other internal structures including the lens capsule, zonule, ciliary body, iris, and cornea. The tumor deposits in certain parts of the globe influence the clinical behavior. Tumor cells on the lens capsule may lead to secondary cataract.⁴ Radiotherapy-induced cataracts occur as a result of damage to the germinative zone of the lens

epithelium, leading to cell death, compensatory mitosis, and generation of “Wedl” cells. The morphology of radiation-induced cataract was described by Cogan and Donaldson.⁵ Following exposure of the lens to radiation, there is a latent period of variable length, depending largely on the radiation dose and the age of the patient. The initial changes are seen as vacuoles at the posterior pole, followed by involvement of the cortex. Posterior subcapsular cataract is the most common morphology seen in such eyes. Transforming growth factor- β is known to potentiate radiation effects.

The incidence of radiation-induced cataracts has decreased dramatically because of the growing popularity of chemotherapy rather than radiation and lens-sparing radiotherapy techniques. This may be achieved simply by irradiating the posterior aspect of the globe. When more focused methods are employed or the target is prone to motion, fixation of the globe is required. Such immobilization is available at highly specialized centers of ocular oncology.⁶ A 1990 study reported that 2 of 23 (9%) eyes developed cataracts after episcleral plaque therapy, 4 of 21 (19%) developed cataracts after EBRT, and 9 of 29 (31%) after combined episcleral plaque and EBRT therapy.⁷ A 1995 study reported that cataracts occurred in 9/11 (82%) eyes with anterior field techniques and 1/8 (12%) eyes treated with lens-sparing techniques.⁸ A 1999 study reported radiation-induced cataracts in approximately 20% of eyes over 36 months of follow-up, even when relative lens-sparing radiation protocols are used.⁹ In 2001, a series was reported where cataract surgery after RB treatment was needed in 45 of 900 (5%) eyes.¹⁰ In 2009, Chodick et al.¹¹ reported risk of cataract extraction among adult RB survivors. Authors evaluated 828 eyes for an average of 32 years of follow-up. During this period, 51 cataract extractions were reported. One extraction was reported in an eye with no radiotherapy compared with 36 extractions in 306 eyes with one course of radiotherapy and 14 among 38 eyes with two or three treatments.

Eyes exposed to 5 Gy or more had a sixfold increased risk of cataract extraction compared with eyes exposed to 2.5 Gy or less.

The interval from EBRT to documentation of a radiation-induced cataract ranged from 9 to 48 months (median, 20 months).¹² Portellos and Buckley¹³ performed surgery for cataract a mean of 54 months after EBRT. In a series by Honavar et al.,¹⁰ the mean time interval for cataract surgery was 26 months after the final treatment for RB. As the average age at diagnosis of RB is 18 months, it is reasonable to assume that most of the eyes developing cataracts are still within the amblyogenic age range. However, lifelong follow-up is required as mentioned above. In adult RB survivors, the average time interval to cataract extraction in irradiated eyes was 51 years following one treatment and 32 years after two or three treatments.¹¹

Secondary cataract formation complicates the management of RB by precluding visualization of the tumor and may necessitate enucleation if there is suspicion of tumor recurrence. Intraocular surgery in these eyes raises genuine concerns about the patient's systemic outcome because of

the risks for viable tumor seeding.^{10,14,15} However, surgical intervention for cataracts is justified in certain clinical settings, especially if the tumor is judged to be clinically stable and in regression.¹⁰ A few case reports have documented simultaneous association of RB with cataract.¹⁶⁻¹⁸ Our focus in this chapter is to review the literature and provide guidelines regarding management of secondary cataracts in eyes with RB (Figs. 40.1 and 40.2).

In 1939, Reese¹⁹ first reported operative treatment of radiation cataracts. Of the 112 eyes with cataracts, 25 eyes were operated on; intracapsular extraction was done on 16, extracapsular extraction on 6, and linear extraction on 3. Reese¹⁹ noted that in cataract caused by irradiation, there is a tendency toward a proliferation of the epithelium under the anterior capsule into a metaplastic fibrous layer. This strengthens the anterior capsule and makes this type of cataract particularly suitable for intracapsular extraction. Extracapsular extraction in such cases is contraindicated because the lens epithelium remaining after the nucleus is extracted may continue to proliferate and form dense fibrous tissue, which tends

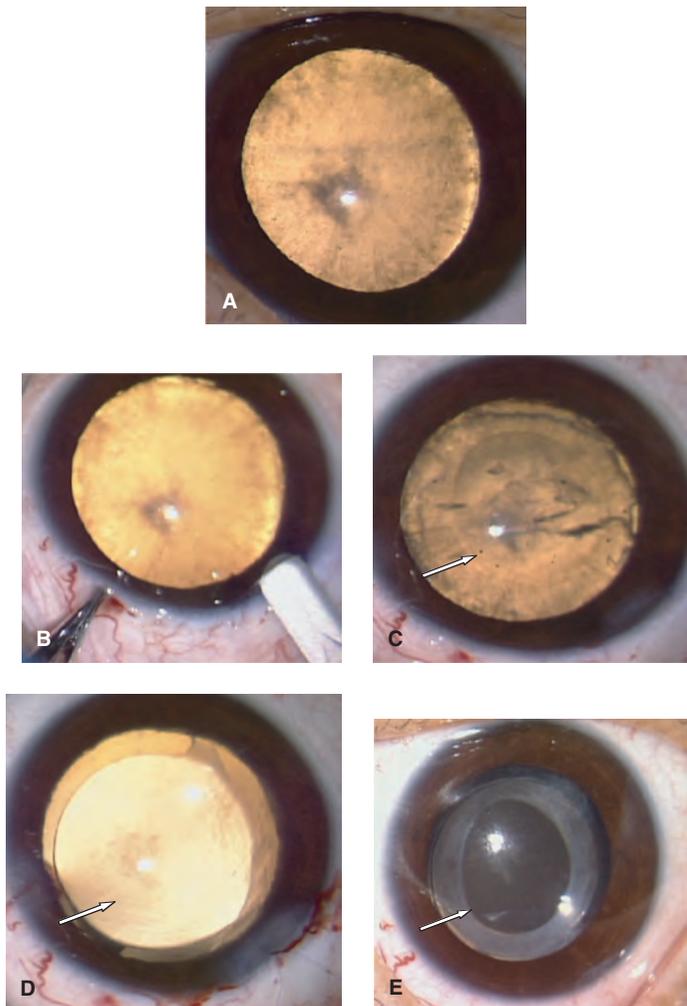


Figure 40.1. **A:** An eye with a radiation-induced cataract (OD) in a 4-year-old child. RB was treated with radiotherapy and chemotherapy. **B:** Corneal incision. **C:** Anterior capsulotomy (arrow shows the edge of the capsulotomy). **D:** AcrySof® IOL implantation in-the-bag. Note the intact posterior capsule; the arrow shows the edge of the anterior capsule. **E:** Two months after cataract surgery. The arrow shows the edge of the anterior capsule.

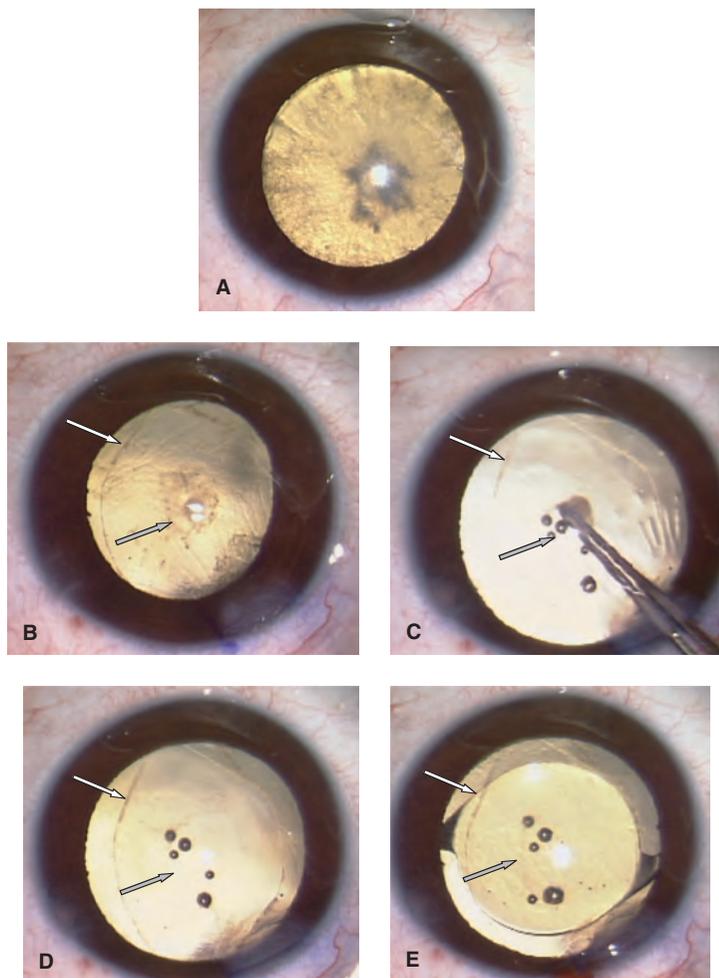


Figure 40.2. **A:** Left eye of the child described in the legend of Figure 40.1. **B:** A Fugo plasma blade was used to perform anterior capsulotomy. During hydrodissection, a radial tear developed. The *white arrow* shows the edge of the anterior capsule. Note posterior capsule plaque (*black arrow*). **C:** Manual posterior capsulorhexis was performed to remove the posterior capsule plaque. Care was taken to avoid disturbing the vitreous face. **D:** Posterior capsulorhexis. **E:** A single-piece AcrySof IOL was implanted in the capsular bag. Haptics were positioned away from the areas of anterior capsule tear.

to produce iridocyclitis and secondary glaucoma. Since then, numerous other innovations in pediatric cataract surgery have evolved. During the past six decades, there have been several reports of successful and innovative surgical procedures in patients treated for RB.^{12,13,20,21} In 2000, Honavar et al.¹⁰ published an excellent article analyzing the results of intraocular surgery in patients treated for RB. There is still concern regarding the safety of performing any intraocular surgery in such eyes, mainly because of the risk for tumor dissemination and systemic metastasis after open globe manipulation of eyes with RB.^{10,14,15}

Following are some practical considerations one should keep in mind when treating such eyes.

1. *Consultation with an experienced RB specialist* is prudent during preoperative and postoperative management of such eyes.
2. The *optimum interval* between completion of treatment of RB and intraocular surgery is not clearly established. Intraocular surgery should be withheld if the tumor is viable or if there is uncertainty about

its activity. Even in patients with documented tumor regression, it may be worthwhile to allow observation for at least 6 to 12 months before attempting cataract removal.¹⁰ Miller et al.²² reported that they observe for a minimum 18-month period after the conclusion of all tumor treatments before considering intraocular surgical management of radiation-induced cataract. If vitreous or subretinal seeding was noted at initial presentation or at any time throughout the treatment course, the cataract was observed for 28 months before surgical intervention.¹⁹ Tumor status and risk of surgery should be assessed individually and discussed in detail with the family and physician treating RB before arriving at a decision about cataract removal. Recurrence rate varied from 10% for Brooks et al.,¹² 31% for Honavar et al.,¹⁰ and 0% for Miller et al.²² who applied the longest interval, suggesting a correlation between the longer interval and the lower rate of recurrence. In Osman's series, one recurrent case was operated 37 months after RB treatment completion and only 3 and 5 months elapsed for the two other cases for the sake of proper monitoring of the tumor.²³

This confirms the importance of adequate tumor control before any surgical intervention and the difficulty of weighing the risk of surgery to the risk of delay of diagnosing a recurrence due to an opaque lens with insufficient tumor visualization. While the optimal quiescent interval is not known, several studies have shown no tumor recurrences when the quiescent interval was at least 16 months.²⁴

3. Dissemination of RB cells through the cataract surgery *incision* has been reported.¹² The clear corneal incision may reduce the risk of inadvertent conjunctival implantation of viable tumor cells and may allow for direct inspection of the incision site for tumor recurrence (unlike the limbal or scleral incision, which may be obscured by the overlying conjunctival flap)¹⁰ (see Fig. 40.1B).
4. The presence of a *posterior capsule* opening theoretically increases the risk of dissemination of viable RB cells to the anterior chamber and extraocular extension through the incision site. In a multicenter trial published in 1990,¹² major complications occurred only in those eyes that underwent pars plana or pars plicata lensectomy. One eye that underwent pars plana lensectomy developed a retinal detachment. The authors recommended avoidance of posterior capsulectomy in eyes with persistent vitreous haze. If RB regression has been deemed stable for at least 6 to 12 months after cataract surgery, Nd:YAG laser posterior capsulotomy may be cautiously performed where required. However, Portellos and Buckley¹³ later reported the safety of extracapsular cataract extraction and posterior chamber intraocular lens (IOL) implantation in combination with pars plana posterior capsulectomy and anterior vitrectomy in a series of eight patients (11 eyes) with radiation-induced cataracts after RB treatment. We prefer to avoid opening the posterior capsule if the

posterior capsule is not associated with plaque and age limits permit (see Fig. 40.1D). However, more often these eyes are associated with posterior capsule plaque (see Fig. 40.2B) or defect. In this case, it may become necessary to perform posterior capsulectomy. However, we still try to achieve manual posterior capsulorhexis (with intact anterior vitreous face), to avoid vitreous face disturbance and subsequent vitrectomy (Fig. 40.2C and D). Primary posterior capsulectomy with or without vitrectomy helps decrease visual axis opacification (Figs. 40.3 and 40.4).

5. Implantation of an *IOL* by itself may not increase the risk of recurrence of RB or systemic metastasis and can be considered for providing optimal visual rehabilitation after cataract surgery.^{10,23} It is important to remember that these eyes are *poor candidates for contact lens wear due to dry eyes associated with decreased tear production from lacrimal gland irradiation*. Thus, in unilateral cases, IOL implantation is the only reasonable option for the correction of aphakia.
6. *Cytologic examination* of vitrectomy fluid can provide direct intraoperative evidence of viable RB. Prompt enucleation and adjuvant chemotherapy with or without orbital radiotherapy may be considered in such situations.
7. *Close long-term follow-up* (Fig. 40.5) is warranted, for several years, to detect possible tumor recurrence and systemic metastasis. Recurrence of RB after intraocular surgery is a potentially serious problem. Tumor recurrence has been reported to range from 0% to 45% after various intraocular procedures. One study¹² reported a recurrence of RB in three eyes (8%), necessitating enucleation of two eyes. Orbital exenteration was required in one case for subconjunctival RB recurrence that developed at the site of the cataract incision. RB recurrence was confined primarily to eyes with

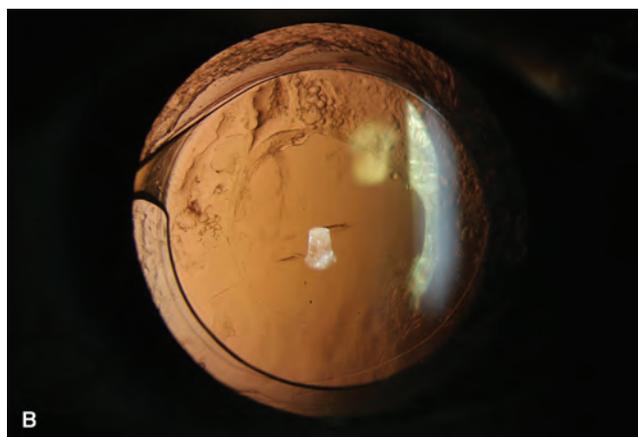
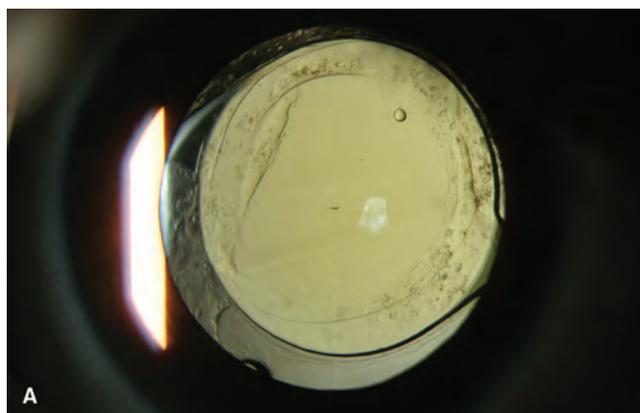


Figure 40.3. A and B: Three-year follow-up of a child operated for bilateral radiation-induced cataract for RB. Cataract surgery was performed at 7 years of age using manual posterior continuous curvilinear capsulorhexis and in-the-bag AcrySof® SN60WF IOL. Note that vitrectomy was not performed. Left eye underwent YAG laser capsulotomy 2 years postoperatively (**B**).

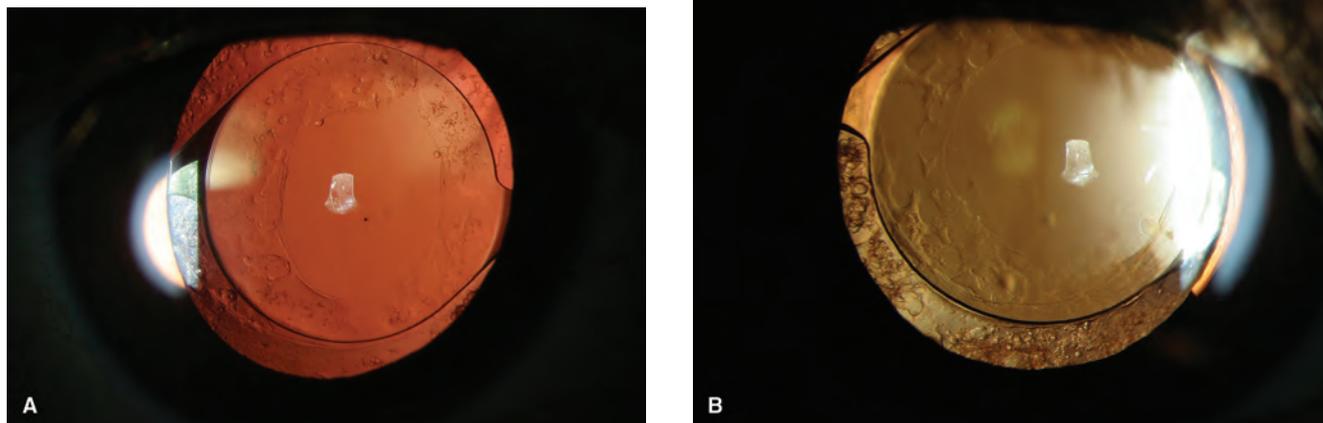


Figure 40.4. Three-year postoperative follow-up of a child operated at 5 years of age. Right eye underwent posterior capsulectomy and vitrectomy (A), while left eye received manual primary continuous curvilinear capsulorhexis without vitrectomy (B).

persistent vitreous haze or vitreous hemorrhage at the time of surgery. There was no systemic metastasis. Use of the limbal approach and avoidance of primary posterior capsulectomy–vitrectomy and avoidance of scleral incision were among the recommendations. Honavar et al.¹⁰ reported recurrence of RB in 21% (7/34) of eyes undergoing cataract surgery, all of which underwent subsequent enucleation. None of the patients who underwent cataract surgery developed metastasis. Most recurrence occurred within the first year, with the longest interval being 19 months in that series. Patients needing a scleral buckling procedure or pars plana vitrectomy seemed to be at greater risk for RB recurrence compared with those needing cataract surgery.¹⁰ RB continued to regress in 31 eyes (69%) after pars plana vitrectomy. A viable tumor was detected by cytologic examination of the vitrectomy fluid in two patients (4%) in whom vitreous hemorrhage had precluded visualization of the tumor immediately before

intraocular surgery. Both of these patients underwent immediate enucleation. Long-term follow-up of these eyes is helpful to see whether surgery in eyes harboring regressed RB allows for a reasonable visual outcome, as opposed to recurrence of RB necessitating enucleation or systemic metastasis.

Postoperative complications in Miller's series included cystoid macular edema in 5 of 16 eyes (31%) and the development of iridocyclitis in 3 of 16 eyes (19%). These complications were transient and responded to topical anti-inflammatory therapy. No local tumor recurrence, orbital tumors, or metastatic disease was detected during the follow-up period. No lens displacement, persistent inflammation, radiation vasculopathy and optic neuropathy, or retinal detachment was seen in any of the patients.

VISUAL REHABILITATION

Due to radiation or early enucleation, the child may have a fellow eye that is significantly enophthalmic in appearance. If this is the case, consider prescribing a +6.00 or +7.00 sphere for an already poorly seeing eye or an anophthalmic socket with a prosthesis.²⁵ This will give the optical illusion of a larger and therefore more symmetric-appearing eye. When proposing patching or atropine penalization to the patient with RB, consider the overall clinical picture. Is the family overwhelmed? Is the retina so distorted from residual tumor and treatment scars that vision may not improve? Remember that these are not “normal” amblyopic eyes. They often have extensive pathology that precludes improvement in vision even with the most rigorous patching regimen. Sometimes, especially in eyes that have received EBRT in very young children, sensitivity to the patch adhesive can occur.

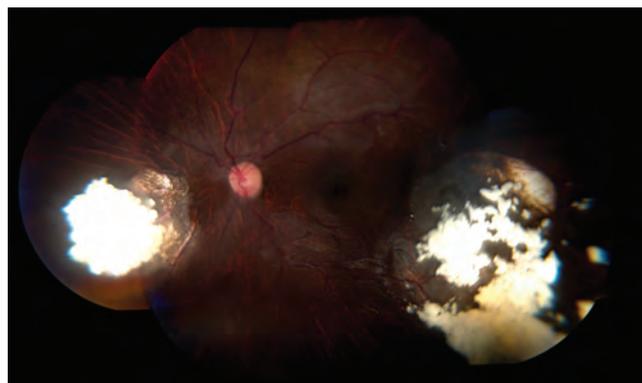


Figure 40.5. Fundus photograph of the left eye of an 18-year-old girl who was operated for radiation-induced cataract at 3 years of age. Note regressed tumor with intralésional calcification.

VISUAL OUTCOME

Even after a successful surgical outcome, the final *visual acuity* depends on several factors. In addition to clarity of the visual axis, other factors such as amblyopia, refractive error, macular tumors,²⁶ radiation complications (keratopathy and/or retinopathy), optic atrophy, and chronic retinal detachment can affect visual outcome. Final visual acuity outcomes generally correlated with the extent of preoperative macular involvement.²² The visual prognosis is limited by initial tumor involvement of the macula and by corneal complications of radiotherapy.

SUMMARY

It is important to weigh the expected benefit of visual rehabilitation against the risk of tumor recurrence and metastasis and to discuss it with the family before proceeding to cataract surgery.¹⁰ Surgery of radiation-induced cataracts in children with RB is a challenge as early intervention is weighted against the need to delay surgery until complete tumor control is obtained. Current techniques for pediatric cataract and IOL surgery can be applicable for radiation-induced cataracts after complete regression of RB. However, considering the risk of tumor recurrence, it is advisable to take a cautious approach, including clear corneal incision and preservation of the posterior capsule if possible. Whatever method is chosen, a discussion with the family regarding realistic outcomes is necessary. As stated previously, the retinal pathology may be so extensive that there may not be an improvement in the vision. While rare, reactivation has been well documented in the literature and should be included in a proper informed consent.

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Pediatric Cataract Associated with Ocular and Systemic Anomalies

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As ophthalmic surgeons are well aware, surgery in pediatric eyes can be challenging. Pediatric cataract surgery in eyes with additional ocular anomalies can be even more demanding. In addition, concomitant systemic disease may put a child at higher anesthesia risk (for anesthesia during surgery as well as any future examinations under anesthesia). Visual acuity outcomes may be reduced when the cataract is part of a systemic disease compared with children without a systemic syndrome.¹ In this chapter, we discuss pediatric cataracts associated with ocular and systemic anomalies. We do not provide an exhaustive list of anomalies; instead, we review those anomalies where management or outcomes may differ from what would otherwise be expected and if they are not discussed as an individual chapter elsewhere in this book.

ALPORT SYNDROME

Alport syndrome is a hereditary nephritis accompanied by high-tone sensorineural deafness and distinctive ocular signs. It was first reported in the early 1900s (quoted in Ref.²). Guthrie described several cases of familial idiopathic hematuria and suggested maternal genetic transmittance.² In 1927, Cecil A. Alport described three generations of a family with a combination of progressive hereditary nephritis and deafness. He linked the hematuria with the auditory defects and noted that the severity of the disease corresponded to gender.³ Many families were subsequently described, and the eponym Alport syndrome was coined in 1961. Anomalous basement membranes in the ocular, auditory, and renal systems cause the characteristic triad of abnormalities in patients with Alport syndrome (i.e., ocular signs, sensorineural deafness, and hereditary nephritis).

The ocular signs were initially discussed by Sohar.⁴ Anterior lenticonus is a distinctive feature, and its presence in any individual is highly suggestive of Alport syndrome. *Anterior lenticonus* is a rare bilateral condition

wherein the anterior surface of the lens protrudes to assume a conical form. Usually, the raised portion consists of clear cortex, while the lens nucleus remains intact and undistorted. Therefore, the deformity is thought to originate in late intrauterine or postnatal life. It is less common than posterior lentiglobus.² Anterior lenticonus is an important indicator of a poor systemic prognosis because of renal disease in Alport syndrome patients. Anterior lenticonus is more common in male than female patients with Alport syndrome.⁵ The inheritance is predominantly X linked (85%), although it can be autosomal recessive (10%) or autosomal dominant (5%).⁶ In addition to Alport syndrome, isolated cases of anterior lenticonus have also been reported, as well as a rare association with Lowe syndrome and Waardenburg syndrome.⁷

Clinical Features

When using a parallelepiped or optic section during slit-lamp biomicroscopy, the lenticonus is seen as an axial protrusion, often conical, within the pupillary zone of the lens. Minor degrees of lenticonus are difficult to detect but are suggested by a distinctive “oil drop” appearance (effect produced by *oil globules in water*) of the red reflex on slit-lamp examination. This is due to the fact that none of the rays from the fundus reaches the observer’s eye owing to prismatic reflection in the axial region.¹ Examination with a retinoscope can sometimes detect anterior lenticonus even when it is difficult to see with the slit lamp.

Associated Ocular Findings

Refractive Error

Slowly progressing myopia and astigmatism may occur. Literature has reported the use of wavefront sensing to evaluate lenticular irregular astigmatism in eyes with lenticonus.⁸ The authors noted that irregular astigmatism induced by lenticonus is a relatively symmetrical, spherical-like aberration, in contrast to irregular astigmatism in typical keratoconus, which is an asymmetrical, comma-like aberration.⁸

Corneal Abnormalities

Posterior polymorphous dystrophy (PPMD) and arcus juveniles are frequently encountered. Thickening of Descemet layer with later endothelial cell changes can lead to PPMD. It should be noted that certain corneal abnormalities can be observed in renal failure patients regardless of etiology. These include a white limbal girdle of Vogt and band keratopathy.² Care must be taken to ensure a complete differential diagnosis of the etiology of the patient's renal disease.

Glaucoma

Iridocorneal adhesions and transparent membranes owing to PPMD result in an increased risk for glaucoma in these eyes.

Cataract

Certain lens opacities are observed in patients with Alport syndrome. First, *anterior subcapsular cortical cataracts* can occur secondarily to lens capsule rupture of the anterior lenticonus. Spontaneous rupture can lead to a complete white total cataract. On careful inspection, these ruptures are usually preceded by small cracks and splits in the capsule visible at the slit lamp. We now recognize these changes as an impending lens rupture. Second, *posterior subcapsular cataracts* (PSCs) may appear because of steroid use with post-renal transplant therapy. Third, internal lenticonus may be seen as a *posterior lamellar opacity* with a posterior projection along the visual axis.⁵ Combined anterior lenticonus and posterior lentiginosus have been reported.⁹ Rarely, lens coloboma has been reported associated with Alport syndrome.¹⁰

Fundus

Yellow-white to silver flecks within the macular and mid-peripheral regions of the retina can be seen.

Anterior Capsule and Lenticonus

Alport syndrome is caused by a genetic defect within one of the α chains of type IV collagen, a major constituent of basement membranes throughout the body.¹¹ In the eye, it mainly affects the anterior capsule of the lens. Streeten et al.¹² identified specific histologic structures that are affected in the crystalline lens capsule. They inferred that the appearance of the lens capsule lesion was similar to the Bowman capsule basement membrane defect in the renal system of Alport syndrome patients. The anterior lens capsule was noted to be one-third the normal thickness centrally and to be more fibrillar than usual, as well as to be associated with large numbers of partial capsular dehiscences containing fibrillar material and vacuoles. The pathologic thinning in eyes with lenticonus as well as the abnormal epithelial cells and fibers may allow bulging of the anterior capsule. Kato et al.¹¹ also noted that

the thickness of the anterior lens capsule was decreased, and there were many vertical capsular dehiscences localized in the inner part of the lens capsule. Besides the anatomical problems with the capsule, manipulation of the lens because of accommodation and normal growth causes added stress on the already weakened structure. Abnormal composition of α (IV) chains in the anterior lens capsule of a patient with anterior lenticonus caused by a mutation in the COL4A5 gene has been reported.¹³

The weakness in the anterior capsule can cause the capsule to rupture, with subsequent formation of an anterior subcapsular cortical cataract or a total white cataract. Traumatic and nontraumatic rupture of the lens capsule has been reported in the literature.¹⁴ In contrast, some reports have noted that the anterior capsule is not so fragile (when performing capsulectomy) in these eyes.^{15,16} These authors theorize that fragility and thinness may present in some patients only in the advanced stages of the disease.^{15,16}

Treatment

Our recommendation for treating an eye with anterior lenticonus is described in Figure 41.1.

Conservative Management

Even if there are no lens opacities, associated refractive errors may affect vision significantly. Glasses and/or contact lenses should be the first line of treatment in such cases. Patients with Alport syndrome should be warned about the possible complication of spontaneous traumatic or nontraumatic rupture of the anterior capsule, leading to total cataract requiring surgery, and informed of the need for prompt evaluation if any sudden change in vision occurs. Careful slit-lamp examinations are needed periodically to look for progression of lenticonus including signs of impending capsule rupture.

Optionally, reduced vision secondary to lenticular changes can be treated with topical mydriatics. Topical phenylephrine can be administered if the patient has axial opacities. If the patient has systemic hypertension, care must be taken when prescribing topical phenylephrine drops, and a diluted concentration can be a viable option.

Surgical Approach

Despite all efforts, conservative management may fail to improve vision satisfactorily. In such cases, clear lens extraction with intraocular lens (IOL) implantation is the reasonable option.^{15,17} Occasional reports of traumatic and nontraumatic rupture of the anterior capsule have prompted some surgeons to treat this disease more aggressively.⁹ When signs of impending rupture are present (newly formed cracks and microbreaks in the capsule near the tip of the lenticonus), a clear lens extraction with IOL should be considered. Rupture of the anterior

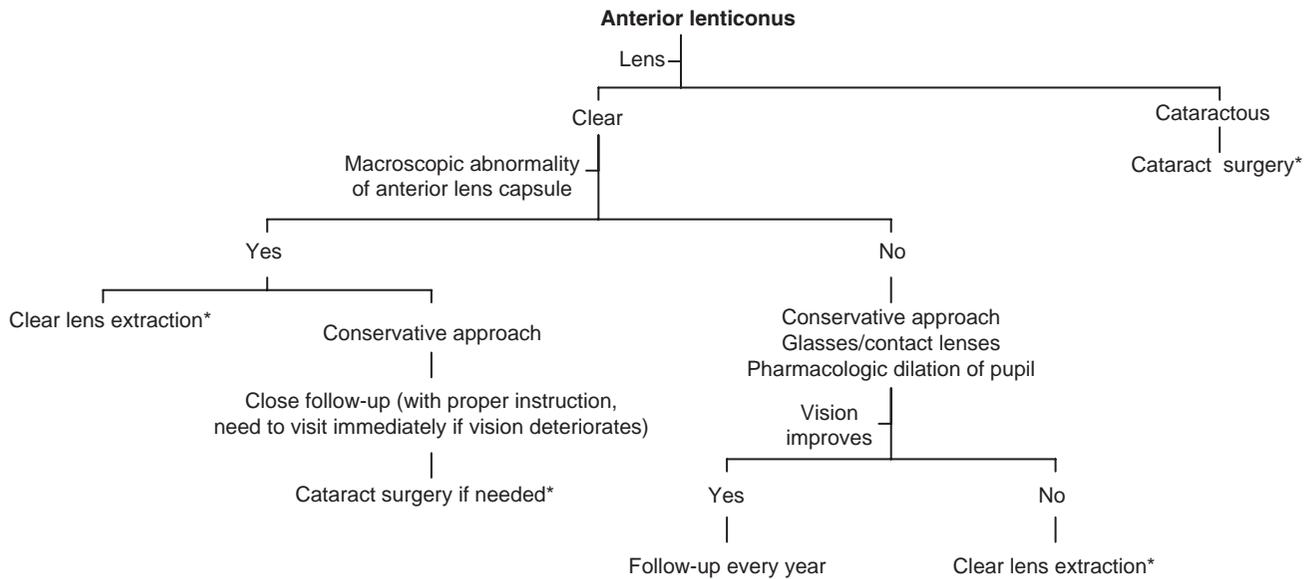


Figure 41.1. Flow diagram showing our current recommendations for treating an eye with lenticonus.

*The other eye generally requires surgery within a few weeks.

capsule may require more urgent intervention, and the uncontrolled spontaneously ruptured anterior capsule (often torn from equator to equator) adds intraoperative difficulties.¹⁸

Although clear lens extraction may be a viable alternative when treating such eyes, care should be taken to document the corrected distant visual acuity and discuss the options with the patient or parents. As with any elective lens surgery, a rare but severe complication (e.g., endophthalmitis) can occur. Documenting the decision-making process is very important in these cases.

Whether operating on a clear lens or a cataractous one, extra care is needed when performing surgery. Herwig et al.¹⁹ reported histologic corneal findings in patients with Alport syndrome. Histology revealed marked irregular thickening of the epithelial basement membrane and of Bowman layer and endothelial changes. This suggests that extra care should be taken to protect corneal endothelium during surgery. Extra care is also needed when performing anterior capsulectomy, as the anterior capsule has been noted to be fragile. Some authors, however, have reported that they did not notice any extra difficulty when performing anterior capsulectomy in such cases.²⁰ Probably, anterior capsule fragility is a concern in advanced cases of anterior lenticonus but not when early intervention is attempted. In our experience, the anterior capsulorhexis is not distinguishable from that of other patients of a similar age. Undoubtedly, the center of the capsule is fragile, but if care is taken to control the capsulorhexis peripheral to the fragile center (using ample viscosurgical agent), a strong capsulectomy edge can be created. For the remaining surgical steps, general surgical principles (as outlined elsewhere in this book) should be followed.

Once one eye is operated on, the other eye will likely require surgery to achieve better binocular vision and often to prevent the crisis of a ruptured anterior capsule in the fellow eye.

Supportive Treatment

Appropriate genetic counseling is essential for the management of Alport syndrome. Due to the high risk for developmental delay and decreased social integration, management requires a team effort from medical, behavioral, psychosocial, and educational specialists. Patients with Alport syndrome should also consider the use of protective lenses during participation in contact sports (Figs. 41.2 through 41.4).

Conclusion

To summarize, Alport syndrome offers many challenges to the ophthalmologist. Patients will present with the characteristic triad of hereditary nephritis, hearing loss, and ocular manifestations. A thorough investigation of the hereditary nature of this syndrome within a family is essential. A multidisciplinary approach in the management of these patients, including assistance with developmental and social deficiencies, is necessary to minimize detrimental effects on their quality of life and improve management outcomes. Close follow-up by an ophthalmologist is essential in patients with Alport syndrome. If signs of early or impending anterior lens capsule rupture are observed, clear lens extraction may be considered to avoid an uncontrolled rupture of the anterior lens capsule and subsequent cataract formation necessitating urgent intervention.



Figure 41.2. Right eye of a 12-year-old African American boy with anterior lenticonus secondary to Alport syndrome. On slit-lamp examination, the paracentral anterior capsule was noted to have findings of early spontaneous rupture. **A:** Direct split illumination. **B:** Retroillumination. **C:** High magnification of spontaneous rupture.

BONE MARROW TRANSPLANTATION

Cataract is a common late side effect of the radiation and steroid treatments used in bone marrow transplantation (BMT).²¹ Total dose and duration of corticosteroid therapy are the most important risk factors for development of cataract.²² Total-body irradiation has also been extensively studied as a cataract risk factor.²² Holmstrom et al.²³ compared the frequency of cataract development in bone marrow–transplanted children who have been given either total-body irradiation or busulphan as conditioning treatment before BMT. The study confirms total-body irradiation as an etiologic factor for cataract (95%) and suggests that busulphan is also, but less frequently (21%), related to cataract development. The study also shows that cataracts in total-body irradiation–treated children developed earlier after BMT than in busulphan-treated children.²³ PSC is the most common type of cataract. PSC typically appears within the first few years after BMT. Frisk et al.²⁴ reported cataract after autologous BMT in children. The authors noted that all children who received total-body irradiation developed PSC ($n = 29$). Six patients (10 eyes) needed cataract surgery (median time 5 years after BMT, range, 4–9 years). We follow 3 children (6 eyes) in this group. Visual outcome is good in this cohort, as cataract typically develops slowly, bilaterally, and at a relatively older age.

CONGENITAL RUBELLA SYNDROME

Congenital rubella syndrome (CRS) remains a prevalent preventable cause of congenital cataract in many parts of the world (Fig. 41.5). The cataracts are usually bilateral, and the children often have associated deafness. Acquired rubella infection is generally a self-limited, relatively benign viral disease in children or adults, but it has the potential to cause serious complications in the developing fetus if the mother contracts rubella within the first 3 months of pregnancy. It is estimated that 25% of the 50,000 children born blind with congenital cataracts in India were born of mothers who contracted rubella during pregnancy.²⁵ In industrial countries, rubella is rare but remains an ever-present public health risk, particularly in immigrant populations.²⁶

Norman Gregg²⁷ noted that “In the first half of the year 1941, an unusual number of cases of congenital cataract made their appearance in Sydney. Cases of similar type, which appeared during the same period, have since been reported from widely separated parts of Australia. Their frequency, unusual characteristics, and wide distribution warranted close investigations.” He contacted colleagues around Australia and eventually recorded a total of 78 cases. He linked maternal German measles (rubella) infection in early pregnancy as the cause of cataract.²⁷

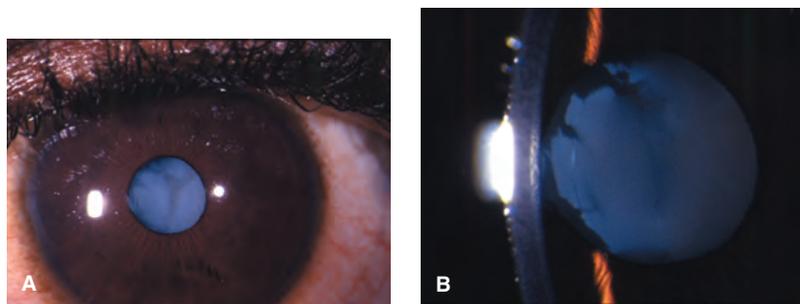


Figure 41.3. The same eye as described in Figure 41.2. Note the total cataract following nontraumatic rupture of the anterior capsule. **A:** Low magnification. **B:** Higher magnification.

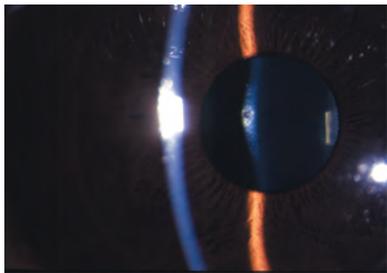


Figure 41.4. Left eye of the same child described in Figure 41.2.

The cataracts were unusual, involving all but the outermost layers of the lens. Gregg suggested that this implied that the cataractous process had begun early in the life of the embryo. A characteristic central or nuclear cataract was the unique symptom observed by Gregg.²⁷ There is potential for a partial cataract to progress to full cataract and further become membranous because of partial absorption of lens matter.²⁸ Nuclear cataract was seen in 90% (36/40) of the eyes; 2 eyes had total cataract and 2 eyes had membranous cataract.²⁹ Cataract is likely to be associated with microphthalmia and poor pupillary dilation. Postoperative glaucoma is more frequent.

Scheie et al.³⁰ reported the outcomes of 49 congenital rubella cataracts. Intraocular specimens from 16 of the congenital rubella eyes were cultured, and the rubella virus was grown from 7. The oldest infant yielding the virus was 18 months. Negative cultures from the nose and throat were found in patients with positive intraocular cultures, indicating the virus may persist in the eye longer than in other tissues. The presence of virus within the eye may be one of the reasons for the high rate of complications following surgery on congenital rubella cataracts.

Vijayalakshmi et al. reported that the visual acuity at final follow-up was 6/24 or better in 6 (15.0%) eyes, and 22 (55.0%) eyes had visual acuity <3/60. Postoperative complications included transient corneal edema in 18

(45.0%) eyes, glaucoma in 5 (12.5%) eyes, after cataract in 1 (2.5%) eye, and hyphema in 1 (2.5%) eye. As mentioned above, the most common postoperative complication in their series was transient corneal edema.²⁹ It is believed that this edema is related to the virus invading the corneal endothelium and reflects the surgical insult on an already-compromised corneal endothelium.²⁹

DIABETES MELLITUS

Cataract formation as a complication of type I diabetes mellitus in young children and adolescents is rare (ca. 1%).^{31–34} Cataracts in patients with insulin-dependent diabetes mellitus (IDDM) were first documented by John Rollo in 1798 (cited in Ref.³²). In 1934, O'Brien and Malsberry studied 126 cases of diabetic patients between 2 and 33 years of age and found cataracts in 16% (cited in Ref.³²).

Cataracts in younger patients with diabetes are usually bilateral and can be mild or severe (total) (Fig. 41.6). Unilateral cataracts do occur in diabetes, but close follow-up is needed since a cataract is likely to develop in the fellow eye. Diabetic cataracts are unique to younger patients with diabetes who have a long duration of symptoms or a history of poorly controlled disease. Acute cataracts have even been described in young people as a presenting feature of their diabetes. This should prompt a detailed history and appropriate lab testing. For more slowly forming cataracts, one study showed that good metabolic control did not protect against cataract formation.³³ However, most studies, including the one just mentioned, noted that metabolic control of these patients is generally poor.

Cataracts associated with diabetes in childhood typically have a band of anterior or posterior subcapsular vacuoles or dense white cortical “snowflake” opacities.^{31–33} The cataracts may arrest, partially regress, or progress with the appropriate treatment of diabetes. Early cataracts have been shown to resolve with metabolic control and normalization of fluid and electrolyte status but are permanent once lens protein coagulation occurs.³⁴

The mechanism of cataract formation from diabetes appears to be multifactorial since all children with diabetes do not form cataracts.³¹ Local factors, genetic predisposition, and nutritional status, combined with other potential known causes such as steroid use, may play a role in the development of diabetic cataracts.³² The osmotic hypothesis, with aldose reductase playing a key role in hyperglycemic-associated cataracts, has been

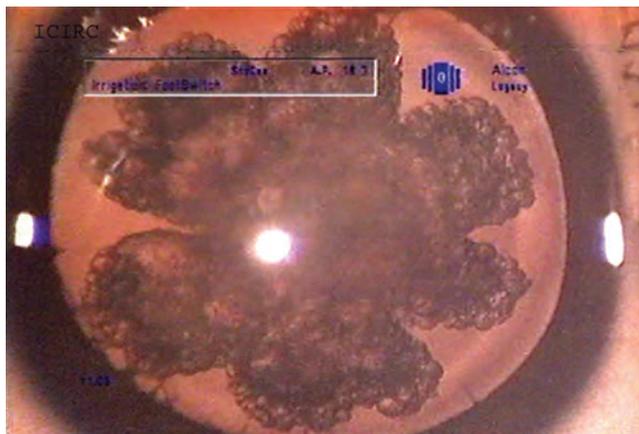


Figure 41.5. Rubella cataract. (Courtesy of Dr. Abhay R. Vasavada, Ahmedabad, India.)



Figure 41.6. Bilateral total cataracts in a diabetic patient.

supported by studies using cell cultures, galactose-fed rats, and aldose reductase inhibitors in rats.³⁵ Dog studies have shown a similar mechanism involving aldose reductase. Its activity in an adult dog lens is similar to that in a human.³⁶ The polyol pathway involves intracellular excess glucose being reduced to sorbitol by aldose reductase. Sorbitol is then reduced by sorbitol dehydrogenase to fructose, which can penetrate the cell membrane. The increase in intracellular sorbitol causes an osmotic gradient leading to swelling of lens fibers and subsequent alterations of membrane permeability. There is a resultant loss of potassium ions and amino acids, with a rise in sodium ions and a cessation of lens protein production. Continued lens hydration and electrolyte disturbances result in lenticular opacification.³⁵

Ehrlich et al.³⁷ reported 10 cases of cataracts in children with IDDM in a 10-year period. IDDM in these patients had been diagnosed at age 3 weeks to 14 years, with the cataracts diagnosed at age 6 to 16 years. Metabolic control was considered average to poor in these 10 patients. Montgomery and Batch³² have reported nine cases of diabetic cataracts over 16 years. The average age of the diabetic cataract patient was 10.1 years. Two of nine patients had cataracts at the time of diagnosis of IDDM, one within 3 weeks of diagnosis, and the other six patients developed cataracts 1.7 to 13.0 years after diagnosis. The authors noted that metabolic control of the nine patients was generally poor, with only one patient achieving a satisfactory average hemoglobin A_{1c}. Falck and Laatikainen³³ retrospectively analyzed the occurrence and possible predisposing factors of diabetic cataracts in a population-based series of some 600 children. Six patients (1%) needed cataract surgery. At the diagnosis of cataracts, they were 9.1 to 17.5 years old, and the duration of diabetes was between 0 months and 3 years 11 months. The type of cataract was similar in all patients, characterized by bilateral snowflake-type cortical deposits and PSC. Four of the six patients had at least a 6-month history of diabetic symptoms before treatment was started, and five patients had ketoacidosis at initial admission to hospital. In 1 of the 11 operated eyes, diabetic retinopathy was observed immediately after surgery. Three patients developed proliferative retinopathy within 7 to 10 months after the operation, after 6.3 to 11.8 years of diabetes.

We have reported the outcomes of 11 cases (22 eyes) from the accumulated data of six pediatric ophthalmic practices.³⁸ The mean age at the time of diagnosis of diabetes was 9.6 years (range, 6 months to 14 years), while the mean age at the time of cataract diagnosis was 10.7 years (5.0–15.5 years). In two patients, the diagnosis of cataract preceded the diagnosis of IDDM, while in three patients, the diagnoses were simultaneous. In some patients, the cataracts were bilateral at presentation, and in others, a unilateral cataract was present, with subsequent cataract

development in the other eye. Various studies have reported a preponderance of girls.^{31,32} Our results were similar, with 8 of 11 patients being female. The description of cataracts in our patients included posterior subcapsular, lamellar, flake-like opacities throughout the nucleus and cortex, and dense white milky cataracts. Nine of the eleven patients (17 eyes) underwent cataract extraction, with a postoperative visual acuity 20/40 or better in 15 eyes. In 13 of these eyes, an IOL was implanted. In 6 eyes, a primary posterior capsulectomy was performed, while in 11 eyes, the posterior capsule was left intact. The literature on adults has reported that diabetic patients developed significantly more posterior capsule opacification after cataract surgery than did nondiabetic patients.³⁹ Pediatric eyes are at high risk for posterior capsule opacification if the posterior capsule is left intact. Eyes with diabetic cataracts may be at higher risk (Fig. 41.7). Eight of these eleven eyes eventually required Nd:YAG laser capsulotomy. Two patients developed diabetic retinopathy postoperatively in our series. It has been reported that cataract surgery increases the risk of diabetic retinopathy.⁴⁰ However, to the best of our knowledge, an influence of primary posterior capsulectomy and vitrectomy on diabetic retinopathy has not been reported. Theoretically, these eyes may be at higher risk for diabetic retinopathy. Long-term follow-up is required (Fig. 41.8).

In summary, since cataracts can be a presenting symptom of diabetes, children with acquired cataracts of unknown etiology should be questioned about classic symptoms of diabetes. Evaluation for hyperglycemia should be performed. Early diagnosis and initiation of metabolic control are important since cataract formation

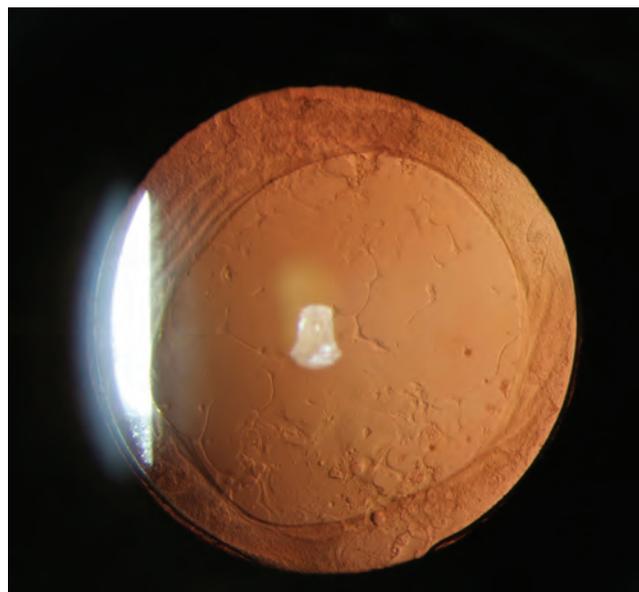


Figure 41.7. Two years postoperative. The child was operated for cataract at 12 years of age. Posterior capsule was left intact. AcrySof® IOL was implanted in the bag.

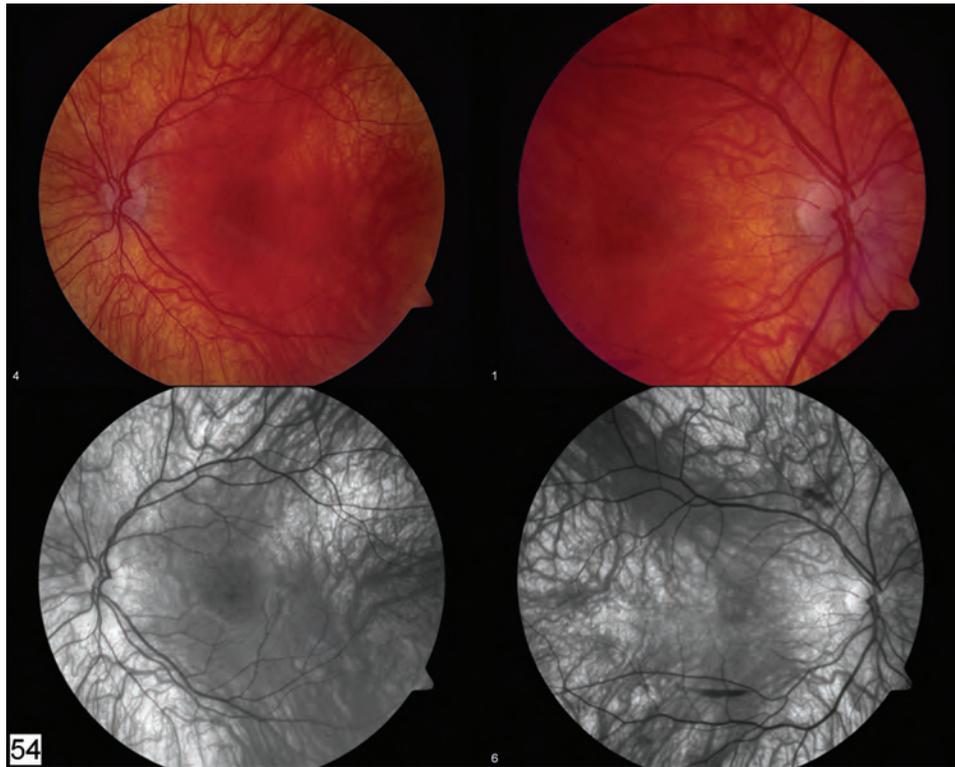


Figure 41.8. Sixteen-year follow-up after cataract surgery in a diabetic girl at 13 years of age. Patient was c/o floaters in the right eye. Preretinal hemorrhage and vitreous hemorrhage were seen in the right eye. The nature of mild nonproliferative diabetic retinopathy was discussed with the patient, and emphasis was placed on tight glucose, blood pressure, and serum lipid control. Avoidance of smoking was emphasized. Maintenance of normal body weight was emphasized.

may be influenced by the duration of symptoms prior to treatment. Early detection of cataracts, determination of etiology with subsequent glycemic control, and evaluation by an ophthalmologist for surgical removal are essential in the treatment of diabetic cataracts in children. Modern cataract surgery techniques can be performed safely when visual acuity is diminished provided that there is close observation to monitor the potential onset of diabetic retinopathy.

DOWN SYNDROME

Lenticular opacity is frequently observed in patients with Down syndrome (Figs. 41.9 and 41.10). In 1910, Pearce et al.⁴¹ reported that among 28 cases of Down syndrome, 19 have lens opacities in some form or other. A 4.6% incidence of cataract in children between 3 months and 19 years old with Down syndrome in Ireland has been reported.⁴² A population-based study of Down syndrome and early cataract (congenital and developmental from 0 to 17 years) from Denmark found an overall frequency of lens opacities of 1.4% (visually significant, 1%).⁴³ Gardiner et al.⁴⁴ reported that surgery was needed for lens opacities in Down syndrome in 33 of 467 eyes (7%).

Other studies showed that Down syndrome was present in 5.4% (13 patients of 243) in the United Kingdom⁴⁵ and 2.8% (29 patients of 1,027) in Denmark⁴³ of patients with early cataract. Lim et al.⁴⁶ noted that among syndrome-associated pediatric cataract, Down syndrome represented one-third of the cases ($n = 18$). A third had dense total or near-total cataracts, followed next by posterior subcapsular and nuclear cataracts. The median age at presentation

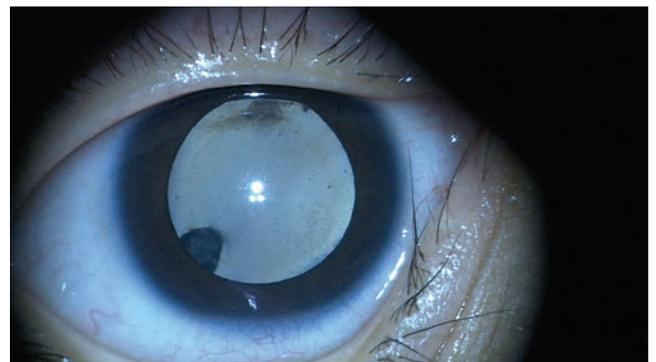


Figure 41.9. Visually nonsignificant cataract noted at the time of probing for congenital lacrimal duct obstruction in a 9-month-old child with Down syndrome.

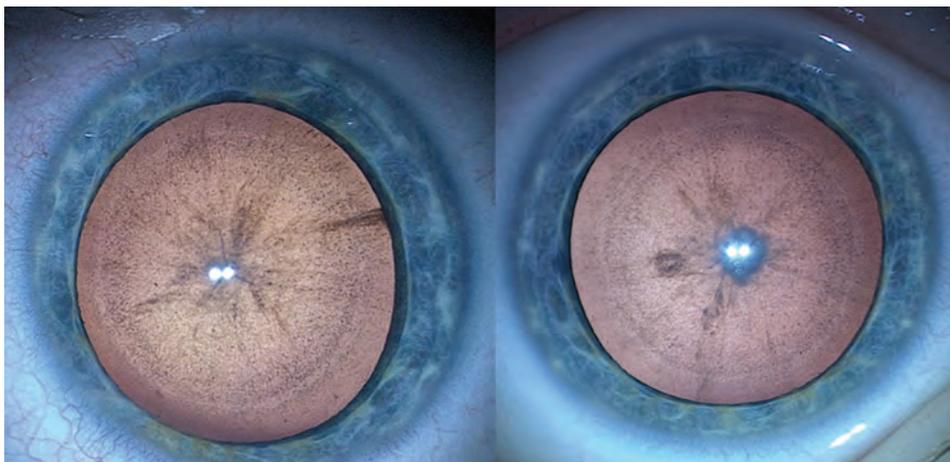


Figure 41.10. Bilateral visually significant cataract in an 18-year-old girl with Down syndrome. Vision in right eye 20/100, left eye 20/200.

in Lim's series was 2 months. The majority had bilateral cataract. Sixteen of the eighteen patients required surgery, with a median age at surgery of 3.3 months. Ten patients had strabismus.

Median age at surgery was 2 years in the Haargaard and Fledelius study.⁴³ Patients with Down syndrome are better suited to IOL implantation because of difficulties with contact lenses or spectacle wear.⁴⁴ The high incidence of myopia and the large myopic shift in Down syndrome patients are well known and should be taken into account when selecting IOL power. The incidence of RD in Down syndrome patients has been reported to be higher (6%)⁴⁴ than would be expected in the general population. This may be attributed to the higher degree of myopia. Hiles et al.⁴⁷ reported that three eyes were lost by self-injury in children with Down syndrome. We found that IOL reposition/dislocated was noted in 4/14 (28.5%) eyes with primary IOL implantation, attributed to self-injury.⁴⁸

GLAUCOMA-ASSOCIATED CATARACT

Glaucoma is a known postoperative complication of cataract surgery. However, glaucoma may also present along with cataract or a cataract can develop in eyes operated for glaucoma. When cataract and glaucoma present simultaneously, an assessment is needed as to which one of the two vision-threatening processes needs the more urgent treatment. If the glaucoma can be managed medically, the cataract is operated first. If the cataract is partial but the glaucoma is an infantile glaucoma, then surgery on the drainage angle is done first. Cataract surgery and goniotomy or trabeculotomy can be performed together if needed. Surgery to place a glaucoma seton is normally done separate from the cataract procedure but could also be done together if indicated. al-Hazmi et al.⁴⁹

documented cataract in 2/180 eyes after glaucoma surgery using mitomycin C. In eyes with buphthalmos, corneal decompensation has been reported, and thus, extra precautions are advisable in eyes undergoing cataract surgery.⁵⁰

HALLERMANN-STREIFF CATARACTS

Hallermann-Streiff syndrome is an isolated disorder with a characteristic facial appearance (brachycephaly, micrognathia, and thin, pointed small nose), as well as short stature, skin abnormalities, and variable developmental delay. The cataract has a tendency to resorb, but even when it does, surgery is still required to remove the opaque capsules. Such membranous cataracts are thin fibrotic lenses caused by the reabsorption of lens proteins. The anterior and posterior lens capsules fuse forming a dense white membrane. This occurs most typically in Hallermann-Streiff syndrome but has also been observed in congenital rubella and Lowe syndrome.

LOWE SYNDROME

Lowe syndrome (the oculocerebrorenal syndrome of Lowe) is an X-linked recessive hereditary disorder. The syndrome was first described by Lowe in 1952 and is characterized by mental retardation, Fanconi syndrome of the proximal renal tubules, and congenital cataract.⁵¹ Other findings include glaucoma, corneal opacity (keloid), enophthalmos, hypotonia, metabolic acidosis, proteinuria, and amino aciduria. Patients with Lowe syndrome often exhibit typical facial features that include frontal bossing, deep-set eyes, chubby cheeks, a fair complexion, and blonde hair (Fig. 41.11).^{52,53}

Lowe syndrome patients have bilateral cataracts that are usually deemed visually significant at or near birth. Cataract was observed in 10/12 Lowe syndrome cases in



Figure 41.11. An 8-year-old patient who demonstrates the typical facial features of Lowe syndrome. (Reprinted from Kruger CJ, Wilson ME Jr, Hutchinson AK, et al. Cataracts and glaucoma in patients with oculocerebrorenal syndrome. *Arch Ophthalmol* 2003;121(9):1234–1237, with permission.)

one cohort, either total or nuclear and often combined with a posterior polar plaque opacity (cited in Ref.⁵⁴). The characteristic appearance of the lens includes multiple gray-white opacities present in all the layers of the cortex, often in wedge-shaped segments.⁵⁵ Tripathi et al.⁵⁶ suggested that the characteristic lens opacities in Lowe syndrome result from a genetic defect in the lens cells. This defect manifests early in embryogenesis, and the progression of the lens opacities is related to both the inherent genetic abnormality and the prevailing extralenticular environment. A flattened, discoid, or ring-shaped cataract can be caused by defective formation and subsequent degeneration of the primary posterior lens fibers. The other findings, such as anterior polar cataract, subcapsular fibrous plaque, capsular excrescences, bladder cells, and posterior lenticonus, are not necessarily specific for Lowe syndrome. The authors further noted that the pathogenesis of Lowe syndrome cataract can be explained by the Lyon hypothesis, which implies that, very early in embryogenesis (at the stage of the primitive streak), one of the two X chromosomes in females

is deactivated. They considered the high incidence of lens opacities in female carriers to be due to this random deactivation. In male probands, since there is no normal X chromosome to nullify the effect of the Lowe gene, all lens cells are affected. Female carriers can be clinically identified solely on the basis of lens examination. The gene responsible for this syndrome has been localized to the Xq25 region using restriction fragment length polymorphism analysis.⁵⁷

Lowe syndrome cataract surgery outcomes from here at Storm Eye Institute were retrospectively reviewed and published by Kruger et al.⁵³ Seven patients (14 eyes) with bilateral cataracts associated with Lowe syndrome were identified and reported. The mean age at cataract surgery was 1.2 months. All but one patient underwent cataract surgery in the first 2 months of life. IOL placement was used less frequently. Age at surgery, glaucoma propensity in these eyes, and poor pupil dilation were recorded as reasons the patients did not get IOL implantation at the time of surgery. Spierer and Desatnik⁵⁸ have noted spontaneous intracameral bleeding at the end of cataract surgery in both eyes of two patients with Lowe syndrome. It is very crucial to follow these eyes for glaucoma. Glaucoma may develop before cataract, present along with cataract, or develop anytime after cataract surgery (Fig. 41.12A and B). Many of the eyes in the Storm Eye series were diagnosed with glaucoma at the time of cataract surgery when they underwent examination under anesthesia. Patients should be monitored closely for changes in IOP, optic nerve cupping, and refractive error, so that glaucoma can be detected and treated promptly.

It is difficult to assess the visual outcome in this group of patients, as many of them are severely developmentally delayed. In 1986, Tripathi et al.⁵⁶ reported that the best visual outcome that can be expected is in the range of 20/100. Even with early surgery and early optical replacement of the crystalline lens, visual acuity is not likely to be normal in these patients, and nystagmus usually develops.

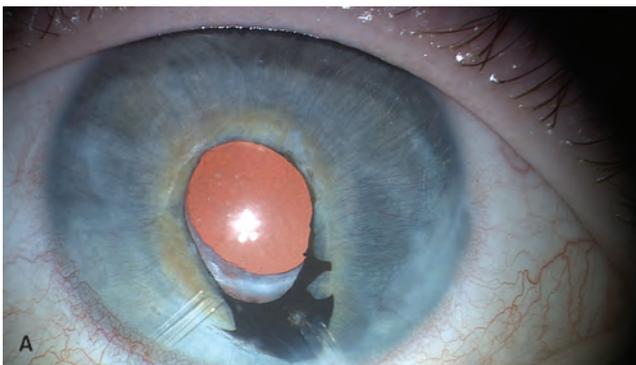


Figure 41.12 A and B: Both eyes in a boy with Lowe syndrome operated for cataract during infancy. Secondary IOL implantation was implanted in both eyes. Right eye underwent glaucoma seton surgery twice.

STICKLER SYNDROME

Stickler syndrome is an autosomal dominant, inherited disorder with several ocular and systemic manifestations including myopia, strabismus, glaucoma, vitreoretinal degenerative changes with retinal detachment, and cataract (cited in Ref.⁵⁹). Various types of lens opacities have been observed in patients with Stickler syndrome. Seery et al. evaluated characteristics of cataract in 133 patients with Stickler syndrome. Cataracts of various types or aphakia were found in 115 of 231 eyes (49.8%).⁵⁹ The most frequent and distinctive lesions, described as wedge and fleck cataracts, accounted for 40 of the 93 (43%) cataracts observed. These distinctive opacities may serve as a clinical marker for Stickler syndrome and may facilitate early diagnosis.⁵⁹ Cataract may also develop after retinal detachment surgery. Visual outcome is generally poor.

FETAL ALCOHOL SYNDROME

Maternal alcohol abuse during pregnancy causes malformations of the eyes with serious consequences to the vision of the affected children.⁶⁰ A high percentage (up to 90%) of children suffering from fetal alcohol syndrome (FAS) have eye abnormalities, including malformation in the outer eye region, disorder of motility, and defects of different intraocular structures. Hypoplasia of optic nerve head and increased tortuosity of retinal arteries are commonly observed. Stromland⁶⁰ noted that 13% of FAS children displayed abnormalities of the anterior segment and media, for example, microcornea, iris defects, cataract, and persistent hyaloid. We care for one child with FAS (Fig. 41.13). Cataract was unilateral and was removed at 12 years of age. One-month postoperative corrected distant visual acuity was 20/150.

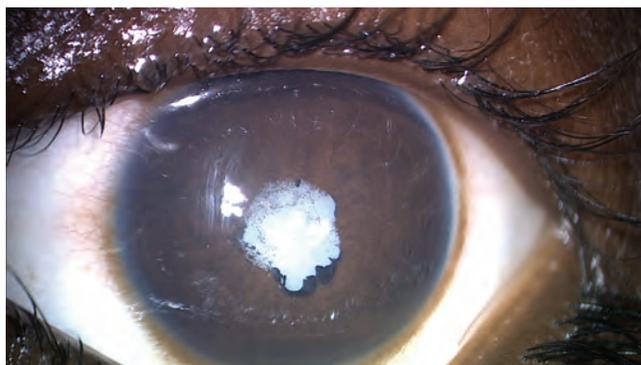


Figure 41.13. Dense cataract in a child with FAS.

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Pediatric Cataracts in Developing-World Settings

M. Edward Wilson, Rupal H. Trivedi, and Parikshit Gogate

Worldwide, an estimated 1.4 million children are blind, of whom approximately 190,000 (14%) are blind due to bilateral unoperated cataract, complications of cataract surgery, amblyopia due to delayed cataract surgery, or the presence of other cataract-associated anomalies. Pediatric cataract blindness presents an enormous problem to developing countries in terms of human morbidity, economic loss, and social burden.¹ Managing cataracts in children remains a challenge, even in the industrialized world. Treatment is often difficult and tedious and requires a dedicated team effort. Since the children are the future of any society, it is time to give childhood cataract treatment in the developing world the attention that adult cataract treatment has received for many years. Blindness or visual impairment in children often results in more than just loss of vision. Their overall development, educational progress, and vocational opportunities can be dramatically changed. To assure the best long-term outcome for cataract-blind children, appropriate pediatric surgical techniques need to be defined and adopted by ophthalmic surgeons in developing countries. This chapter focuses on practical guidelines and recommendations for ophthalmic surgeons and health planners dealing with childhood cataract in the developing world.

Of the 1.4 million blind children in the world, approximately 90% live in Asia and Africa, and 75% of all causes are preventable or curable.¹ The prevalence of blindness varies according to the socioeconomic development of the country. In developing countries, the rate of blindness can be as high as 1.5 per 1,000 population. Compared to industrialized countries, this figure is 10 times higher.¹ Due to lifetime of blindness ahead, the burden of disability in terms of “blind-years” is huge. The child who goes blind today is likely to remain with us into 2050.² Restoring the sight of one child blind from cataracts may be equivalent to restoring the sight of 10 elderly adults in terms of blind-person years prevented.² Irrespective of the cause, childhood blindness has far-reaching effects on

the child and his/her family throughout life. It profoundly influences educational, employment, personal, and social prospects for the affected child. The control of childhood blindness has been identified as a priority of the World Health Organization’s (WHO’s) global initiative for the elimination of avoidable blindness by the year 2020.³

An uneven distribution of ophthalmologists, pediatricians, and anesthesiologists creates unique challenges in developing-world settings. In general, health services are concentrated in the larger cities, and people living in rural areas often live beyond the reach of the services provided by health care delivery teams.

GUIDELINES AND RECOMMENDATIONS FOR HEALTH PLANNERS AND SURGEONS

Delayed presentation and late surgical treatment are the major causes of severe visual impairment and blindness in children with cataract.⁴ It is important to *improve the early identification and referral* of children with cataracts by educating and training pediatricians, rural health clinic personnel, midwives, and eye-camp workers to screen for loss of the eye’s red reflex and poor visual functioning in newborn, toddlers, and school-aged children. Although early surgical intervention and prompt optical rehabilitation are mandatory to prevent irreversible deprivational amblyopia, surgery for cataract with delayed presentation can also help to regain functional vision.⁵ Delivering good pediatric eye care is a team approach, and ophthalmologists need to work in tandem with other health care and education personnel to best help the affected children.

The primary health care worker can also be taught to identify cataracts. Ophthalmic assistants, working with primary health workers, can maximize the efficiency of a limited eye care system by screening and referring only patients requiring surgery or other specialized attention. It is also important to emphasize

that all types of eye staff need to have the stimulus of continuing education and eye seminars to provide the necessary incentive and encouragement and to update their knowledge and skills.

Most developing and developed nations provide health delivery services through a tiered system, with central hospitals supporting smaller and rural health delivery centers. In Africa and Asia, for example, many countries have established a three-tiered system, consisting of primary, secondary, and tertiary levels. The primary health care worker can diagnose and treat the most prevalent diseases and refer complicated cases to treatment facilities. The usual referral resource for the primary health care worker is the secondary facility. Provincial, district, and subdistrict hospitals and health centers serve as secondary medical units. Ophthalmologists are assigned to provincial and sometimes to district hospitals. The central urban hospital, usually attached to a medical school, is the tertiary resource. There may be several tertiary hospitals in larger states, provinces, or countries serving large geographic regions. This facility usually is a large general hospital and offers a wide range of specialty services. We believe that the treatment of cataract-blind children should be done in *specialized, well-equipped, pediatric eye care centers that are piggy-backed on comprehensive eye surgical centers*, where cataract operations on children are done on a regular basis.

There are many persons who have close contact with the community, especially at the village level. These persons, by use of appropriate knowledge, could help in the prevention of blindness. These may include, the school teacher, mukhia (leader of the village), religious leader, traditional birth attendants, and others have great influence in rural regions of most developing countries. The key informant approach used this premise to identify blind children in Bangladesh and Malawi to great success.^{6,7}

Pediatric eye care is a teamwork of primary health and education service facilities that help in identifying children with visual impairment and pediatric eye care units based at tertiary centers where medical and surgical service is imparted. ORBIS International has used this approach as have other nongovernmental organizations working in the area of childhood blindness prevention. The referral is not just from primary to tertiary centers but also in the reverse direction, after the surgery, for better compliance with refractive correction and amblyopia treatment and to promote sensitivity to the child's education needs. *Publicity* may be accomplished through a variety of mechanisms at several levels using health care personnel, radio, television, and other media. Service organizations (e.g., Lions Club, Rotary Club) are often involved in promotional activities.

The harvest season may be a time of reduced surgery since travel to a centralized treatment facility may not be possible without loss of the family livelihood. Many

parents of school-age children prefer to have elective surgery done during the annual school vacation.

A significant influence in persuading parents to allow their child to undergo cataract surgery is likely to be the example of other children in the community who have had sight restored by such an operation. A reputation for good results from surgery is the major influence in the decision-making process for parents. Thus, in the initial phase, "*patient selection*" is very important. Patients with good visual potential (bilateral dense cataracts without nystagmus or microphthalmia in children who are progressing developmentally) should be operated on first to assure that parents and community leaders will trust and believe in the surgery being offered to the blind children. After establishing some initial success, cases with a more guarded prognosis can be operated. There are many reports of children with dense cataract operated late who nonetheless improve markedly on their preoperative visual acuity.

In some locations the facilities are in place but are *underutilized*. Valuable resources of trained staff have often been wasted, or at best poorly used, because they have not been given even the basic equipment to carry out their work—even though the equipment is often inexpensive and simple. Basic instruments to diagnose ophthalmic disorders should be provided (visual acuity charts, torch, direct ophthalmoscope, etc.) to such a setup. Measuring a child's visual acuity before and after the intervention can help to gauge the outcome of the treatment. Children as young as 8 to 9 months can have their visual acuity measured by preferential looking charts.

Power fluctuations and outages are additional challenges in the developing-world setting when surgical procedures, like pediatric cataract surgery, depend on automated machinery. The development of better battery-operated vitrectomy cutting instruments would be helpful in these settings.

It could be argued that all that has to be done is to transfer the well-proved methods and instrumentation from the West to all areas of the world and all problems would be solved!⁸ Even if such a transfer were possible, nothing could be farther from the truth. For example, no matter how many vitrector machines were made available, this would not solve the problem. The machines would break down, and the cost of a vitrector handpiece would prevent its routine use. We have witnessed a setup where a machine was available and the surgeons were proficient enough to use this technology, but because the tubing and handpieces were not available the machine was not being used! In general, it is advisable to choose instruments that have reusable tubing sets and handpieces whenever possible for use in developing-world settings. Machines that reboot quickly when power outages occur, work well in a dusty environment, have modular design for easy repair, and continue to cut when

a nitrogen or compressed air supply is exhausted (combined venturi and peristaltic pump systems) are ideal. In addition, investments in biomedical maintenance and repair training will pay off almost as well as the surgical training itself.

For surgical methods to be appropriate for developing world settings, they must be cost-effective, time effective, and sight effective. Surgical techniques described in other chapters in this book can be followed when treating children with pediatric cataract. However, in developing-world settings where communication and follow-up are difficult, a “*once and for all*” rather than a staged procedure may be chosen more often than in the developed world. For this reason, it may be better to do a more comprehensive procedure rather than trying a method that may need to be repeated. Primary posterior capsulectomy and anterior vitrectomy need to be done up to an older age, perhaps into the second decade of life, when compliance to follow-up is uncertain.

SIMULTANEOUS BILATERAL PEDIATRIC CATARACT SURGERY OR IMMEDIATELY SEQUENTIAL CATARACT SURGERY

Some authors have proposed simultaneous pediatric cataract surgery to manage the backlog of cataract blindness in the developing world (see Chapter 9). The fears of bilateral blinding endophthalmitis or bilateral post-operative wound rupture have made unilateral surgery the normal procedure in the industrialized world. In the amblyopic years, surgery in the second eye is usually done within a few days or weeks of the initial surgery. In the developing world, this cautious approach may not be practical. To avoid the risks and costs of a second anesthesia and to make maximal use of the vitrector tubing and cutter, simultaneous bilateral surgery on children may be given consideration, especially for very young children or in those locations with rudimentary anesthesia facilities. Wherever possible, however, the second eye should be operated upon by a sterile different set of instruments and consumables to minimize the chances of infection.¹

GENERAL ANESTHESIA VIA ENDOTRACHEAL INTUBATION OR LARYNGEAL MASK AIRWAY

A detailed review of the techniques of general anesthesia at ophthalmic care centers in the developing world is beyond the scope of this chapter. Cost and effectiveness are both very important when choosing anesthetic agents in a developing-world setting. We recommend that pediatric cataract surgery be performed under general anesthesia with constant monitoring of the vital signs.

Ophthalmic surgeons in Guatemala and India have utilized modern inhalation agents via laryngeal mask airway (LMA) combined with peribulbar lidocaine and marcaine in children to facilitate maintenance of anesthesia on as little inhalation agent as possible.¹ If necessary, ophthalmic surgeons should be given a special course in general anesthesia for eye surgery.

Considering the limited resources and availability of anesthetists in remote areas of the developing world, use of ketamine (2 mg/kg body weight for induction) has been proposed. Ketamine combines analgesic and sleep-producing effects without significant cardiovascular and respiratory depression. Ketamine can also be used intramuscularly when a vein is not accessible in a young child, and the resultant sedation may later be used to gain intravenous access. The most common side effects are the so-called “emergence phenomenon,” which includes disorientation, vivid dreams, and sensory or perceptual illusion. Literature has reported the use of ketamine anesthesia in combination with peribulbar lidocaine for pediatric cataract surgery.⁹ While an anesthetist is not always available, the anesthesia is always administered by a person trained in pediatric airway management and resuscitation. A pulse oximeter is used as part of the vital sign monitoring. This technique has the advantage of using ketamine as the dissociative anesthesia and peribulbar lidocaine (combined with ocular massage) as the local anesthetic agent to anesthetize the ocular tissues and to counter the effect of increased intraocular pressure caused by the ketamine. The authors concluded that ketamine may be useful in a simple ophthalmic setup in the developing world. Hennig, from Lahan, and Ruit, from Kathmandu (both in Nepal), have used intravenous ketamine with good results. Hennig added a small Pulseox/ECG for monitoring and an oxygen concentrator. He has also found that most of the children in Nepal older than 5 years can be operated on using local anesthesia (Albrecht Hennig, *e-mail communication*). Surgeons from Bangladesh and India have also used this approach.

SURGICAL INSTRUMENT AND STERILIZATION

It is much easier to ensure that a few instruments are maintained in good condition at all times rather than a large and complicated set. Simpler methods are generally used for sterilization of the instruments. Sharp instruments are simply sterilized by immersion in an efficient antiseptic solution such as 2% Savlon (a mixture of hibitane and cetrimide) together with an antirust agent such as sodium nitrite. This method is simple and inexpensive and does not depend on electricity or the integrity of a sterilizer. All other nonsharp surgical instruments can be sterilized by boiling (protecting the points with a rubber or silicone

tube [2-ram sheath]). When instruments are protected in this way, the use of trays and racks is unnecessary. In all cases, soft or rain water must be used for boiling to prevent damaging the instruments with deposits from hard water. Again, this method is simple and inexpensive, and almost universally applicable, even in very simple and rural situations. But, as always, autoclaving of instruments is the gold standard and should be used whenever possible. There are autoclaves available that can function without electric supply by using cooking gas or stoves.

PEDIATRIC CATARACT SURGERY IN THE DEVELOPING WORLD

Pediatric cataract surgery is much costlier than is adult cataract surgery in developing countries.¹⁰ The cost of adult cataract surgery has been estimated to range from \$15 to \$42, while pediatric cataract surgery cost was estimated to range from \$121 to \$475.¹⁰ A cataract-blind adult gains an average of 5 blindness-free years when operated. In contrast, cataract surgery may give a cataract-blind child 50 or more blindness-free years.¹⁰ Several authors have reported the outcome of cataract surgery in children from developing-world settings.^{4,11-14}

Due to poor compliance and the difficulty in following pediatric surgical patients in many areas of the developing world, we usually suggest a primary posterior capsulotomy followed by an anterior vitrectomy for children up to at least 8 years of age and perhaps even older, depending on the availability of a Nd:YAG laser.

Aphakic glasses, contact lenses, and intraocular lenses (IOLs) are each proven methods of visual rehabilitation after pediatric cataract surgery. Pros and cons of these options have been described elsewhere in this book. Implantation of an IOL during cataract surgery in the developing world seems to be a practical option, while other methods of visual rehabilitation (aphakic glasses and contact lenses) are less suitable in these settings. It is difficult to replace spectacles once broken or damaged, due to the expense and unavailability. Contact lenses are impractical for most patients in the developing world because of environmental and hygienic problems as well as initial cost and the cost of replacement when lost. Regular follow-up visits to eye care clinics are problematic due to the cost and distance of travel. Contact lenses are expensive and easily lost. An IOL can provide a full-time correction with optics that closely simulate those of the crystalline lens. In the industrialized world, IOL implantation at the time of cataract surgery is the most common means of optical correction for children beyond infancy. In a study done in Africa, Yorston and coworkers¹² also recommend IOL implantation as the treatment of choice for most children with cataracts in the developing world. We agree with this assessment. In addition, Yorston has proposed that the IOL in young children be placed in the

ciliary sulcus, with the optic captured through an anterior and a posterior vitrectorhexis. We recommend this technique whenever polymethylmethacrylate (PMMA) IOLs are utilized in young children in the developing world. These IOLs are inexpensive but often not flexible enough to safely be placed into the capsular bag of a young child. We recommend that a vitrector be used to remove the cataract and to make an identically sized anterior and posterior capsular opening. These openings should be 1 to 2 mm smaller than the IOL optic. A generous vitrectomy should also be performed if possible. The IOL is then placed into the ciliary sulcus and, depending on the design of the IOL, captured posterior to the dual capsule openings. Recurrent visual axis opacification may be low using this technique since the capsulotomy edges will seal to create a closed Soemmering ring. The additional advantage of sulcus fixation over capsular fixation when a posterior capsulectomy is being done in the developing world is the ease of IOL exchange later and the ability to safely implant when microscope visualization is not ideal.

There are numerous instances of manufacturing of high-quality PMMA IOLs inexpensively in the developing world. Financial self-sufficiency can be attained by the physician as cataract surgery programs are able to recover costs from user fees. The IOL-producing facilities in Madurai, India (AuroLab of Aravind Eye Hospital), and in Nepal and Eritrea (Fred Hollows Foundation facilities) are successful examples. Foldable lenses made from hydrophobic acrylic biomaterials (AcrySof®; Alcon, Fort Worth, TX, USA) have become the most commonly implanted IOLs for children in the United States, but are prohibitively expensive for use in many developing-world settings. Numerous manufacturers based in India/Asia now offer foldable acrylic IOLs at around \$20.

Selecting the best IOL power to implant in a growing child presents unique challenges even in industrial countries. The lack of instrumentation in the developing-world operating-room setting, such as the handheld keratometer and the A-scan ultrasound, increases the difficulty of selecting the appropriate IOL power to use for pediatric cataract surgery. To minimize the need to exchange IOLs later in life when a large myopic shift occurs, it has been advised to leave children hyperopic after IOL implantation with an IOL so that they can grow into emmetropia or mild myopia in adult life. However, if glasses are difficult to get and will introduce delays in achieving the best visual functioning, the initial IOL in the developing world may be selected aimed at emmetropia or even mild myopia at the time of surgery. This makes immediate postoperative amblyopia treatment easier and also gives the child good unaided vision for near viewing, which is important for their scholastic tasks. IOL exchange, however, will be needed more often using this approach as the child gets older since some of these patients will become very highly myopic over time. The sulcus IOL option

discussed earlier is not often chosen in the developed world but it does facilitate the easy IOL exchange when eye growth has occurred.

POSTOPERATIVE CARE AND REFRACTION

Many surgical facilities and parents erroneously believe that a good pediatric cataract surgery is the answer to all of the child's problems. Actually pediatric cataract surgery is just an important step in a series of steps needed to rehabilitate a child's vision. Proper postoperative medication and refraction with spectacle wear are a must. Most unilaterally affected and many bilaterally affected children will also need some kind of amblyopia treatment. A regular follow-up is needed to achieve this. Follow up can best be improved by using a systematic approach which links individual children, their families, and the hospital.¹⁵ Many children improve over months as the surgery and optical correction help the visual system to mature. The ophthalmologists have to guard against postoperative inflammation, intraocular pressure rise, and visual axis opacification. A good distance visual acuity is no guarantee that a child will be able to read his/her textbook, unless bifocals are prescribed. Congdon and colleagues reported that provision of postoperative spectacles was one of the factors predicting better visual acuity.¹⁶ A counselor/social worker/coordinator can help deliver yeoman care by repeatedly contacting parents and ensuring a regular follow-up. Pediatric cataract surgery is not a 100-m sprint but rather a marathon that can tire many stakeholders (parents, children, and doctors).

SUMMARY

The acceptability, accessibility, and affordability of cataract surgical services must each be carefully addressed to improve efficiency. In some locations the facilities are in place but underutilized, because there is a lack of knowledge, monetary constraints, or a negative public perception of the surgery owing to poor results using inadequate or poorly timed treatment. Inadequate ophthalmic and anesthesiology staff, lack of ophthalmic surgical instruments, and poor equipment maintenance are also widespread in developing countries. Other problems include the logistic complexities of identifying the children who will benefit most from surgery and arranging reliable transportation to the treatment center.

We recommend the following steps to improve the long-term visual outcome of children with cataracts, regardless of the cause of those cataracts. These recommendations are in addition to the many ongoing efforts aimed at the eradication of childhood cataracts through such programs as rubella vaccination and nutritional improvements. The steps should be funded by nongovernment organizations and regional eye centers through donations and cost recovery plans. Our recommendations to facilitate the application of these surgical steps are as follows. (1) Improve the early identification and referral of children. (2) Designate regional centers for the treatment of pediatric cataracts. (3) Set up a twinning relationship between each regional developing-world pediatric cataract center and an industrialized world center that has experience with pediatric cataract surgery.

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Postoperative Medications and Follow-up

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Regular postoperative examinations and appropriate medical treatment are crucial for a successful outcome in all surgical interventions. There are certain aspects in pediatric cataract surgery that make the issue of follow-up more important. Children, especially infants, are unable to express complaints and depend on their parents or a caretaker for the use of medications; furthermore, ophthalmologic examinations including assessment of visual acuity, slit-lamp examination, and intraocular pressure (IOP) measurement are tedious or impossible in some cases. Certain postoperative complications such as inflammation, synechia formation, glaucoma, anterior capsule contracture, posterior capsular opacification, and membrane formation are encountered more frequently in pediatric cataract surgery. Cataract surgery in a child is also different in the sense that the operation aims to provide lifelong useful vision in an individual in whom amblyopia threatens a successful outcome.

The key to a successful outcome in pediatric cataract surgery lies in continuous and periodic reevaluation with the meticulous attention of the surgeon, pediatric ophthalmologist, and the parents or caretaker. An uneventful operation is simply the first step toward achieving the main goals; only by careful observation for expected conditions, and also the unexpected, does one reach the ultimate objectives. The surgeon should inform the parents of the need for continuous follow-up prior to undertaking surgery and stress the importance of effort and dedication for the long period of observation.

The postoperative care of pediatric cataract surgery consists of medical treatment and periodic examinations.

POSTOPERATIVE MEDICATIONS

Medical treatment is of prime importance following pediatric cataract surgery. The bases for postoperative medical therapy in children are studies performed on adults' and surgeons' experience. Although guidelines are often stated, the postoperative regimen should be tailored individually. There are certain points to be considered in small children. Due to their lower body weight, one main consideration in the application of eye drops in children is the possibility of systemic absorption and related side effects. Most of postoperative medications for pediatric cataract surgery are prescribed for the first time; therefore, attention to systemic side effects is mandatory. Measures to decrease systemic absorption, such as digital punctal occlusion and eyelid closure, are therefore more important in children.

Medications prescribed in the postoperative care of pediatric cataract surgery fall into two main categories. First are those routinely prescribed in uncomplicated cases and, second, drugs used under certain circumstances and for specific indications. This chapter focuses on the first group of medications, which includes antibiotics, steroids, nonsteroidal anti-inflammatory drugs (NSAIDs), cycloplegics, mydriatics, and topical anesthetics.

Antibiotics

The use of perioperative antibiotics for prevention of infection and postoperative endophthalmitis remains a controversial issue. Postoperative endophthalmitis following pediatric cataract surgery is very uncommon, and the overall incidence has been reported to be 7 per 10,000 operations.¹ Due to the severity of the condition and the potential of visual loss, surgeons generally take preventive measures to reduce its incidence. Prophylaxis of endophthalmitis is discussed in Chapter 12. Povidone-iodine preparations are widely used for conjunctival antisepsis immediately before and at the conclusion of surgery.² The use of intracameral antibiotics is discussed in Chapter 12. Another preventive measure against endophthalmitis and wound infection is the use of perioperative topical antibiotics including gentamicin, neomycin, tobramycin, and polymyxin B.³⁻⁵ Many of these medications have been abandoned due to local and systemic side effects; instead fluoroquinolones are commonly used for this purpose. Fluoroquinolones were first introduced for the treatment of corneal and conjunctival infections, but ophthalmic surgeons currently use them before intraocular surgery to prevent bacterial endophthalmitis. These drugs have certain attractive characteristics for prophylactic use. They exhibit good activity against gram-negative and gram-positive organisms that are the main causative agents of postoperative endophthalmitis. These also have minimal epithelial toxicity and when used frequently can attain effective concentrations in the corneal stroma and the anterior chamber. Among topical fluoroquinolones, ofloxacin has the best transepithelial penetration. Ciprofloxacin is also effective against the major organisms producing endophthalmitis and also shows good coverage for gram-negative microorganisms. Perioperative use of both ciprofloxacin and ofloxacin has reduced bacterial colony counts of external ocular flora. Norfloxacin is less suitable for prophylactic use in ophthalmic practice since it is less effective against gram-positive organisms and penetrates the epithelium less readily.⁶

Fourth-generation fluoroquinolones such as gatifloxacin and moxifloxacin offer a broader spectrum of coverage than do previous generations of fluoroquinolones. These potent topical bactericidal agents promise to have greater antibacterial properties against gram-positive organisms and atypical mycobacteria. Recent studies showed that both of these fourth-generation fluoroquinolones achieved greater aqueous humor concentration after four times daily dosing relative to prior-generation fluoroquinolones. Moxifloxacin 0.5% ophthalmic solution achieved a significantly higher aqueous humor concentration than gatifloxacin 0.3% ophthalmic solution. The superior penetration of moxifloxacin into the aqueous humor may be attributed partially to its high degree of lipophilicity, greater solubility at neutral pH, and higher

concentration in the commercial formulation.⁷⁻⁹ It has been suggested that moxifloxacin 0.5% used in a normal clinical setting is unlikely to cause any obvious adverse effects on the normal human cornea.¹⁰

Unlike intracameral antibiotics, topical antibiotics have not been shown to decrease the incidence of endophthalmitis in a randomized clinical trial. The best ocular penetration from eye drops still falls far below that achieved with intracameral use. The study by Espiritu et al.¹¹ showed that intracameral moxifloxacin 0.5 mg/mL appeared to be nontoxic in terms of visual rehabilitation, anterior chamber reaction, pachymetry, and corneal endothelial cell density. Another study showed no untoward effects after intracameral injection of moxifloxacin 0.5% ophthalmic solution diluted to a 0.1% concentration at the conclusion of routine cataract surgery.¹² There was also no increased risk associated with a 250 µg/0.050 mL intracameral injection of moxifloxacin for prophylaxis of endophthalmitis after cataract surgery.¹³

One regimen for prophylactic use of fluoroquinolones is preoperative frequent instillation, every 5 to 15 minutes, 1 to 1.5 hours before surgery and postoperative use every 6 hours for 5 to 7 days.⁶ However, a study by Ta et al.¹⁴ indicated that preoperative ofloxacin started 3 days prior to surgery was more effective than instillation 1 hour before the operation in reducing colony counts. All available fluoroquinolones are well tolerated by the corneal epithelium. Fluoroquinolones may cause damage to cartilage and result in arthropathy; therefore, systemic administration should be avoided in children. Topical use in children entails no risk of arthropathy.⁶

Steroids

The inflammatory response following cataract surgery is more intense in children, which increases the risk of some postoperative complications such as fibrinous membrane formation, pupillary block, pigmentary and cellular deposits on the intraocular lens (IOL), posterior synechia, IOL capture, posterior capsule opacity, and cystoid macular edema (CME). Frequent administration of topical and sometimes regional and systemic steroids is needed to reduce the risk of these complications. *Corticosteroids* are relatively potent anti-inflammatory agents with multimodal mechanisms of action. Despite the efficacy of steroids in reducing postoperative inflammation, their long-term use is associated with multiple adverse systemic and ocular effects. Systemic complications of long-term steroid use in children include growth retardation, affective disorders, Cushing syndrome, skin atrophy, hirsutism, acne, osteoporosis, femoral head avascular necrosis, myopathy, water and electrolyte imbalance, hypertension, duodenal ulcer, immunosuppression, and delayed wound healing. Intracranial hypertension (pseudotumor cerebri) has also been associated with discontinuation of steroids.¹⁵

Ocular complications of long-term steroid use are ocular hypertension, glaucoma, predisposition to infections, and initiation or progression of cataract in the fellow eye.¹⁵ Ocular side effects are related to the potency of the steroid and individual susceptibility. Newer corticosteroids such as rimexolone and loteprednol have been shown to decrease the frequency of IOP elevation during long-term treatment (4 weeks or more) but have shown to be less effective in reducing inflammation.¹⁶ Difluprednate ophthalmic emulsion 0.05% (Durezol[®]),^{17,18} which has been shown to be effective in the treatment of postoperative inflammation and pain, is being used more and more in children.

Following pediatric cataract surgery corticosteroids may be administered either regionally or systemically. Regional steroids may be used topically or locally in the form of eye drops, ointments, and subconjunctival or subtenon injections. Preservative-free triamcinolone acetonide (Triessence, Alcon Labs, and Fort Worth) improves visualization of the vitreous during pediatric cataract surgery, thereby ensuring a complete anterior vitrectomy without adverse postoperative effects.¹⁹ When used for this purpose, it is removed, at least in part, before the surgery is completed. It can also be added intracamerally and not removed (2–4 mg in children) in hopes that it will reduce the dependence on parental compliance with postoperative topical corticosteroids. In the anterior chamber, the triamcinolone is visible for 1 to 2 weeks but does not stay in the eye for a longer period of time like when it is injected into the vitreous cavity. This topic is explored in more detail in Chapter 22.

Common steroids for subconjunctival use include betamethasone and dexamethasone, 2 to 4 mg, injected into the inferior bulbar conjunctiva at the conclusion of surgery.^{5,20–25} The authors usually inject betamethasone, 2 mg in patients under the age of 1 year and 4 mg in older patients. Steroids used for subtenon injection include methylprednisolone acetate and triamcinolone acetonide, 20 to 40 mg, depending on the age of the patient.^{24,26} This form of periocular steroid has been associated with precipitous elevations of IOP.²⁷ Some pediatric surgeons no longer use any subconjunctival steroid in routine cases, relying instead on topical or intracameral administration (M. Edward Wilson, *e-mail communication*).

Steroid eye drops are generally prescribed from the first postoperative day. Steroid drops are an easy, safe, effective, and inexpensive method of drug delivery and usually do not entail systemic effects. Children are usually defensive against eye drop instillation, so one must take time and be patient. Topical steroid eye drops used in the postoperative care of pediatric cataract surgery include prednisolone acetate, betamethasone, dexamethasone, and difluprednate. Most authors recommend 1% prednisolone acetate eye drops for routine use^{3–5,20,28–31}; our experience has shown 0.1% betamethasone to be an acceptable alternative.²⁶ In uncomplicated cases with

minimal postoperative inflammation, 1% prednisolone acetate administered every 4 to 6 hours should suffice. With more severe inflammation, sterile uveitis, or pre-existing intraocular inflammation, more frequent dosage (e.g., every 1–2 hours) may be required. The duration of topical steroid therapy is 4 to 12 weeks, depending on the clinical course. Steroid ointments are limited to bedtime use and are not recommended during waking hours because of the possibility of blurred vision.

Systemic steroid use is uncommon in adult cataract surgery. However owing to the more severe inflammatory response, more extensive surgical manipulation (removal of the posterior capsule, anterior vitrectomy), and lack of adequate cooperation for eye drop instillation, systemic steroids may be needed for pediatric cataract surgery. The usual dose is 1 to 2 mg/kg oral Prednisolone for 1 week, which is tapered and discontinued by 1 to 2 weeks.^{3,20,24,25} No randomized clinical trials have been done to evaluate the efficacy of systemic steroids compared to topical alone or topical and intracameral use. However, for patients with a history of uveitis or trauma or in whom extrasurgical manipulation of the iris is needed, oral steroids are considered, empirically, to help treat the excessive inflammation that is anticipated. These decisions may need to be made on a case-by-case basis.

Nonsteroidal Anti-Inflammatory Drugs

This class of medications reduces inflammation through inhibition of prostaglandin synthesis. The possible advantage of these medications is the potential for reduction of steroid-related complications such as increased IOP, risk of infections, and delayed wound healing. The anti-inflammatory effect of NSAIDs including 1% indomethacin, 0.03% flurbiprofen, 0.5% ketorolac, 0.1% diclofenac, 0.1% nepafenac, and 0.09% bromfenac has been proven. Four of these drugs (0.5% ketorolac, 0.1% diclofenac, 0.1% nepafenac, and 0.09% bromfenac) are FDA approved for this purpose.³² Randomized double-blind clinical trials have shown the anti-inflammatory effect of 0.5% ketorolac and 0.01, 0.1, and 0.5% diclofenac to be equal to that of 0.1% dexamethasone and 1% prednisolone, respectively.³³ A recent study by Miyake et al.³⁴ suggested that nepafenac is more effective than fluorometholone in preventing angiographic CME and blood aqueous barrier disruption, leading to more rapid visual recovery. CME is rarely seen in children, and, therefore, NSAIDs are rarely part of the routine postoperative regimen after pediatric cataract surgery.

When used, NSAIDs are usually administered three or four times daily for 4 to 6 weeks following cataract surgery. These medications may cause ocular irritation and, by interfering with platelet function, predispose to hemorrhage from the surgical wound. Therefore, NSAIDs should be used with caution in patients receiving systemic anticoagulants or with hemorrhagic predisposition.

Another contraindication for the use of topical NSAIDs is herpes simplex keratitis; however, experimental evidence suggests that flurbiprofen may be beneficial in some instances.¹⁵ The above-mentioned studies have been performed in adults; at present, clinical experience with NSAID use after pediatric cataract surgery is rather limited.^{4,22,25,35,36} However, no specific side effects have been reported in children.¹⁵

Cycloplegics and Mydriatics

Mydriatics and cycloplegics diminish ciliary spasm, stabilize the blood–aqueous barrier, and dilate the pupil, thereby reducing pain, inflammation, and the risk of pupillary block caused by synechia or fibrinous membranes. The pupil-dilating effect of these medications may remain for a few hours to a few days, depending on the drug potency and individual susceptibility. The latter depends on iris pigmentation; dark irides tend to respond more slowly to these drugs and return to normal much sooner.³⁷ Commonly available cycloplegic eye drops include atropine, homatropine, cyclopentolate, and tropicamide.

Phenylephrine hydrochloride (PEH) is a potent mydriatic with no effect on accommodation and may be used separately or in combination with cycloplegics. PEH is available at 1%, 2.5%, 5%, and 10% concentrations. The 10% solution should be avoided in children because of possible cardiovascular side effects.³⁸ PEH at 2.5% is preferred in children; however, neonates and premature infants should receive the 1% solution.³⁸

There is no uniformity in the postoperative use of cycloplegics in pediatric cataract surgery; some surgeons prescribe these medications routinely for infants (once per day for 4 weeks) but not older children, some for all patients one to three times daily for 1 to 4 weeks^{5,22,29,36,39,40}; others avoid the routine use of these drugs altogether.^{3,4,28} Use of cycloplegics, especially long-acting ones, in cases where the posterior capsule is intact and anterior vitrectomy is not performed may predispose to IOL pupillary capture if the IOL has been implanted in the ciliary sulcus. However, when a continuous curvilinear capsulorhexis has been performed and the implant is well positioned in the capsular bag with anterior capsule covering the optic edge 360 degrees, the risk of pupillary capture is minimal.

Due to the insidious onset of pupillary capture of IOL, in our practice, we refrain from routine prescription of cycloplegics when the pupillary reflex is fair and insignificant postoperative inflammation is present. We prefer in-the-office instillation of cycloplegics only when postoperative examination reveals an unresponsive pupil or a pupillary membrane is forming. After instillation of a single drop of 0.5% proparacaine, we use one drop of 2.5% phenylephrine followed by one drop of cyclopentolate or

tropicamide. The patient is reexamined after 30 minutes, and if visible signs of drug effect are not noted, the preceding routine is repeated one more time. Some surgeons prefer to use atropine once at bed time to reduce the risk of synechia formation, lessen photophobia, and facilitate examinations in infants less than the 2 years of age (M. Edward Wilson, *personal communication*).

Topical Anesthetics

Topical ocular anesthetics commonly used in pediatric patients are 0.5% atropine, 0.5% proparacaine and 0.5% tetracaine. Proparacaine hydrochloride causes anesthesia in a few seconds; however, the anesthesia wears off after 11 minutes.⁴¹ Many ophthalmologists routinely administer 0.5% proparacaine hydrochloride with cycloplegics and mydriatics; the epithelial changes induced by proparacaine facilitate corneal penetration of cycloplegics and mydriatics, thereby enhancing their effect.⁴¹ Additionally, the corneal and conjunctival anesthesia provided by proparacaine reduces reflex tearing upon instillation of the other drops, increasing patient comfort and promoting more effective contact of the medication with the ocular surface. Adverse effects of proparacaine include contact dermatitis, epithelial keratitis, pupillary dilation, and seizures.⁴¹ Tetracaine is not as well tolerated as proparacaine; it is more irritating and the duration of its effect is shorter (<10 minutes).⁴¹

FOLLOW-UP

As previously stated, lifelong follow-up of children undergoing cataract surgery is crucial for a successful outcome. Surgery alone will have limited value if postoperative follow-up is inappropriate.⁴² Young children are unable to report symptoms; therefore, regular follow-up and detailed examination are mandatory for early recognition of complications and to initiate timely treatment. The first postoperative month and the 1st week, in particular, are the period when complications such as wound problems, endophthalmitis, and sterile uveitis may occur. Furthermore, the refractive status of the eye undergoes constant change requiring regular refraction and necessary changes to obtain optimal vision.⁴³ Prevention and treatment of amblyopia by timely occlusion is another principal issue in the postoperative course.⁴⁴ Such extensive efforts call for cooperation among the surgeon, the pediatric ophthalmologist, and the parents.

Perioperative Considerations

The overall atmosphere in the hospital, particularly in the operating room, causes uneasiness or frank fear in most children. Therefore, it is preferable not to admit children if at all possible. Children are intolerant of being kept hungry and thirsty; therefore, the operation time should

be scheduled precisely to avoid undue expectation or delay. The parents, in particular, undergo a considerable amount of stress and anxiety and are generally easily irritated by signs of inattention.

Postoperative visits should be scheduled well in advance, so that both child and parents can be prepared for full cooperation. These examinations are best scheduled for the early hours of clinic or office activity, when the examiner can deal with the child with more energy and patience. One should avoid disturbing the child's sleeping schedule and avoid unnecessary delay. Taking a few moments before starting the examination to establish rapport or communication, either verbal or physical, will facilitate the process. It is better to start the examination with less invasive procedures such as evaluation of the red reflex; and more invasive or painful procedures such as instillation of topical anesthetics and IOP measurement should be performed toward the end.

An examination of a child after cataract surgery on the first postoperative day is highly recommended. Confirmation that the anterior chamber is formed and a good red reflex is visible may be all that is possible on this visit.⁴⁵ Subsequent visits are scheduled according to the patient's age, the presence of complications and the condition of the operated eye at the first postoperative visit, and the surgeon's experience. Some authors recommend visits on day 1, week 1, and months 1, 3, and 6, followed by examinations every 6 months thereafter.⁵ Others have suggested monthly visits for 1 year; with subsequent visits at 3- to 6-month intervals. Our routine for postoperative visits in uncomplicated cases is daily examinations for the first 3 days, weekly examinations for a month, and monthly visits for 3 months, followed by examinations every 3 months for 1 year and every 6 months thereafter.

At each postoperative visit, visual acuity, slit-lamp examination, and IOP (if feasible) should be assessed. A dilated fundus examination is recommended in the first week after the operation and repeated at 3 months, and every 6 months in uncomplicated cases. On slit-lamp examination, attention must be paid to corneal clarity, wound stability and sutures, anterior chamber depth and reaction, iris, pupil shape and reaction, and IOL position, including centration and distance from the pupil margin. In cases where slit-lamp examination is impossible, penlight-assisted examination with a magnifier (+20 diopter lens) may be adequate. In these cases, a detailed examination under anesthesia 1 to 2 months after the operation is mandatory. If the penlight examination causes suspicion of any complication a detailed examination under sedation or anesthesia becomes necessary. Examination under sedation is another alternative in uncooperative children. Miyahara et al.⁴⁶ have reported the use of oral triclofos sodium, 8 mg/kg, for IOP measurement in children younger than 3 years. A yearly examination under anesthesia in young children is recommended if a detailed

examination is not possible in the office and if sedation is not feasible or practical.

Suture Removal

Wound healing occurs more rapidly in children; therefore, sutures may become loose sooner than in adults. In addition to causing irritation and discomfort, loose sutures predispose to suture abscess and possible intraocular infection. Synthetic absorbable sutures (10-0 Vicryl) are used in children. If permanent nylon sutures are used, removal of loose sutures is necessary at the slit-lamp examination; if that procedure is not possible, suture removal under anesthesia in the operating room should be considered.

Surgically induced or preexisting astigmatism can be managed by timely removal of nonabsorbable sutures. Some surgeons use absorbable sutures, for example, 10-0 polyglactin (Vicryl®) with small incisions during cataract surgery. No induction of astigmatism occurs with this technique.⁴⁷ Postoperative corneal astigmatism may be assessed by keratometry or topography in cooperative children; in other cases, retinoscopy may be a rough guide to corneal astigmatism. Factors such as the type (corneal versus scleral), size and location of the incision, patient's age, degree of scar formation, and required steroid dose affect the timing for suture removal. During the era of larger incisions for polymethylmethacrylate (PMMA) IOLs, selective suture removal was considered in the presence of more than 2 diopters of suture-induced astigmatism 6 weeks after surgery. With the newer technique of foldable IOL implantation, sutures are used to provide secure water-tight wound closure and can be removed within 2 to 3 weeks.

Restrictions

Eye protection is of major concern for prevention of trauma to the operated eye, especially in the trauma-prone period of childhood. Eye shields or protective glasses may be used during waking hours and a shield may be applied during the nighttime. Owing to the rapid wound healing in childhood, use of eye protection is required for only 3 weeks during waking hours and 1 to 2 weeks during sleep. Because of the potential for amblyopia, protective glasses are preferred over eye shields during periods of activity. Strenuous activity including gym classes, contact sports, and swimming should be avoided for 3 weeks.⁴⁸

Correction of Refractive Errors

An excellent surgical outcome and clear optical media are only prerequisites for useful vision after pediatric cataract surgery. Achievement of a successful visual outcome depends on timely correction of residual refractive errors, particularly aphakia. Bilateral and unilateral refractive errors, whether aphakic or pseudophakic, should

be corrected at the first possible opportunity. Another important point is periodic recheck of refractive errors, especially during the first few years of life, when globe growth causes a myopic shift.²⁹ Some studies have shown a mean decrease of 9 diopters in contact lens power during the first 4 years of life.⁴⁹ Eyes with shorter axial length than the fellow eye have a higher rate of growth in comparison to eyes with longer axial length.⁴⁹ Details on options for correction of refractive errors after cataract surgery are described in Chapter 45.

Amblyopia Therapy

Amblyopia and its treatment, particularly in unilateral cases, are one of the formidable obstacles in the postoperative course of pediatric cataract surgery. Eyes with unilateral cataracts requiring surgery have variable degrees of amblyopia.⁵⁰ Furthermore, the unilateral aphakic or pseudophakic eye, even with appropriate correction, can never compete with the normal fellow eye with natural accommodation. Bilateral cases are less prone to develop amblyopia. Amblyopia therapy should be considered in bilateral cases when fixation preference is present in one eye or when discrepancy in visual acuity between the eyes exceeds two Snellen lines.^{5,29,51}

One of the cornerstones of successful amblyopia therapy is cooperation and compliance of the patient and family. The vital importance of proper refractive correction and adherence to the amblyopia therapy schedule should be clearly explained to the parents; their low compliance will lead to suboptimal results.^{52,53} According to the study by Zwaan et al.,⁵⁴ only half of the operated patients were compliant with the treatment regimen; of those compliant with treatment, 70% achieved a visual acuity of 20/80 or better, while the corresponding figure for those with poor compliance was only 38%. Good binocular performance and stereopsis are closely related to good visual acuity in each eye.^{55,56} Conditions related to amblyopia and low vision include strabismus and nystagmus, the management of which requires close and active collaboration with a pediatric ophthalmologist. Additional information on amblyopia can be found in Chapter 47.

Postoperative Complications

Postoperative complications and their management have been discussed in detail in other chapters of this book. Herein, we briefly review the most important complications.

Postoperative sterile uveitis is common after pediatric cataract surgery. The inflammatory reaction may be severe, but hypopyon formation is rare.⁴⁵ Toxic anterior segment syndrome has been reported after pediatric cataract surgery.⁵⁷ Mydriatics are used to prevent synechia formation. Intracameral tissue-plasminogen activator has been used intraoperatively to prevent fibrin formation.⁵⁸ It has also been used after pediatric cataract surgery for

lysis of formed fibrin.⁵⁹ Infectious endophthalmitis must always be considered and ruled out. The incidence of endophthalmitis following pediatric cataract surgery is comparable to that of adults.⁴⁵ The most common isolated organisms are gram-positive bacteria. In cases that fail to respond to medical treatment, vitrectomy must be considered.⁶⁰

Development of glaucoma is possible any time, from months to decades, after pediatric cataract surgery.⁶¹ Due to difficulties in measurement of IOP and lack of adequate patient cooperation, delayed diagnosis is common. Risk factors for development of glaucoma after pediatric cataract surgery include younger age (<9 months) at the time of surgery,⁶² family history of aphakic glaucoma, nuclear cataract morphology, persistent fetal vasculature syndrome,⁶³ microcornea,⁶⁴ significant residual lens material,⁶⁵ and surgery for secondary cataracts.^{61,66} Previously, retinal detachment was a common complication following lens aspiration for pediatric cataracts. This complication may occur after primary surgery or secondary procedures for management of dense and fibrotic membranes. Early diagnosis should prompt surgical intervention.⁶⁷

SUMMARY

Postoperative care, including follow-up examinations and medical treatment, following cataract surgery is a delicate and sophisticated issue in children compared to adults. The follow-up schedule should be individualized as dictated by the clinical course. However, closely spaced examination in the early postoperative period and lifelong observation are mandatory for all children undergoing cataract extraction. Attention to postoperative inflammation and its control are also of utmost importance. Topical steroids and cycloplegics are the cornerstones of treatment for children; however, systemic steroids may be required in certain cases. Periodic assessment of the operated eye is necessary to detect media opacification, glaucoma, or any other complication. Performing an uncomplicated pediatric cataract extraction is only the first step toward visual rehabilitation. Timely correction of refractive errors, amblyopia therapy, and early recognition of other complications are critical for a successful outcome. Interaction among the surgeon, the pediatric ophthalmologist, and the parents is critical in achieving this goal.

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Measuring and Managing Residual Refractive Error After Intraocular Lens Implantation

Scott K. McClatchey

Choosing an intraocular lens (IOL) power in children is driven by the postoperative refraction goal. Managing the residual refractive error in turn depends on the refractive trajectory until the relative stability of adult life. In contrast to adults, the initial refractive goal for a child is driven by long-term outcome, and we think that it is best to start with this goal in mind: good vision when the child grows up into an adult. The same goal applies to the clinician who manages residual refractive error after IOL implantation. This outcome goal can be divided into three parts: emmetropia in adulthood, good visual acuity as an adult, and a manageable course of refraction between IOL implantation and adulthood; this includes the need to treat amblyopia and management of the changing refractive error. Because the surgeon's choice of initial postoperative refraction determines much of the future management of the child, this chapter builds on the discussion in Chapter 7 of this book, "Calculation and Selection of Intraocular Lens Power for Children."

MEASUREMENT

Measurement of refractive error is well known to all ophthalmologists who examine children. Simple objective retinoscopy with or without cycloplegic drops can give an accurate assessment of the refractive error in children after cataract surgery. On the other hand, subjective refractions are difficult to perform in young children and are more prone to error in amblyopic eyes, since the child will have difficulty distinguishing the difference between alternative lenses. I do not recommend using subjective techniques until about age 7 years; even then, I prefer retinoscopy. High-power spherical IOLs will cause a greater spherical aberration than will standard-power IOLs. This can be easily seen with the retinoscope as a refraction at the peripheral pupil of a well-dilated eye

that is more myopic than the central refraction; it can be measured with a wavefront machine. In comparison with retinoscopy, automated refraction can give more repeatable results.¹

Refractive error can also be estimated from biometry in aphakic eyes, though this is less accurate than direct retinoscopy. This can be a useful adjunct in the few children who are strongly or violently resistant to exam in clinic; however, biometry would require anesthesia in these children, and retinoscopy will measure the refractive error directly.

The clinician should wait for 2 to 4 weeks after surgery to obtain an accurate refraction. In the early postoperative period, there are several temporary changes that resolve quickly. The young child's eye is soft and malleable. Surgeons usually use a fine absorbable suture to close the wound in young children: this can cause several diopters (D) of astigmatism that resolves more quickly than in adults as the suture breaks or erodes through the soft corneal tissue. Residual viscoelastic, especially if it remains between the IOL and an intact posterior capsule, can displace the IOL anteriorly in the eye in the early postoperative period, giving in a brief period of myopia (or lesser hyperopia) that resolves as the viscoelastic gradually diffuses away.

The position of a corrective lens can have large optical effects when the lens power is large: the vertex distance must be precisely correct to avoid induced error. For high refractive errors (greater than about ± 4 D), refraction should be measured with corrective lenses in place whenever possible. When the child is wearing his or her own glasses (or contact lenses), this corrective lens is already at the proper position with respect to the cornea; a low-power trial lens held over the glasses will allow the retinoscopist to be more precise in measurement. Because the trial lens held to do a refraction over glasses has a

lesser power, vertex distance errors will cause a much smaller error in refraction than a high-power lens.

Residual refractive error in aphakic and pseudophakic eyes will vary with age, and can change significantly within a few months during the 1st year of life. Because of this, it is useful to time routine follow-up examinations every 3 months up to about age 3 years, and to gradually lengthen this period up to yearly in teenage years and beyond.

MANAGEMENT

The residual error should be managed with the goal in mind: good vision in adult life. Amblyopia management is paramount to this goal in unilateral cataract patients; for all, management of refractive error should minimize the duration of blur in young children and should minimize the untoward effects of anisometropia.

If two IOLs were implanted in an eye at cataract surgery (temporary polypseudophakia²), the anterior IOL should have between 20% and 25% of the total IOL power.³ When the myopia in the polypseudophakic eye becomes excessive, the anterior IOL can be removed: this will result in moderate hyperopia that must be managed with glasses. The child's eye may continue to grow at the same rate, resulting in reduction of hyperopia or even myopia with age.

When cataract surgery is performed in the 1st year of life, many surgeons prefer to leave these infants aphakic. Contact lens management can be very successful even with unilateral cataracts in some cases; two of my patients achieved 20/25 vision with this strategy. However, many children become intolerant of contact lenses in early childhood. For children with bilateral aphakia, this is easily managed by glasses. For unilateral aphakia, the correction with glasses is usually intolerable: the heavy, high-power lens on one side results in aniseikonia, anisophoria, and a lopsided weight on the child's nose; the good vision present in the normal eye compounds the difficulty because the child perceives no direct benefit from the glasses. If the child is left uncorrected with unilateral aphakia in early childhood (before about age 6), this can result in rapid worsening of vision and ultimately a deep, intractable amblyopia.

When a unilaterally aphakic child fails contact lens wear, another optical correction must be attempted; implantation of a secondary IOL may be the best option. Fortunately, the remaining capsular bag usually provides a platform: the growing equatorial lens cells result in a Soemmering ring that separates the anterior and posterior capsule ring-shaped leaflets; when this is opened and the lens cells aspirated, an IOL can be placed in the bag, or in some cases in the sulcus with optic capture.⁴

Secondary IOLs have additional risks due to the second intraocular surgery, but they have a few advantages over primary IOL implantation. Since much of the postoperative ocular growth has already occurred, the myopic shift will not be as great. It is possible that waiting to implant an IOL until the child is 2 to 6 years of age may lessen some of the complications of very early IOL implantation such as fibrosis of the vitreous face. It has also been hypothesized that the rate of refractive growth (RRG2) remains constant throughout childhood, and a measurement RRG2 prior to implantation of a secondary IOL could (hypothetically) be used to more precisely predict the future refractive trajectory of that eye.⁵

Exchange of the IOL to compensate for progressive myopia has been proposed (K. Hoffer, *personal communication*) and reported.⁶ Dahan and Salmenson⁷ stated that because of changes in ocular axial length in the growing eye, an IOL exchange might become necessary to avoid anisometropia.

Anisometropia over 4 D corrected by glasses can give significant aniseikonia and anisophoria in those with good binocularity and can be managed in several ways. Substituting contact lenses for the glasses will practically eliminate the symptoms. Changing the amount of correction in the normal eye can reduce anisometropia, within limits. Dahan and Salmenson⁷ mistakenly undercorrected one patient by 8 D because of a calculation error. Since the cycloplegic refraction of the other eye was +6.0 D, they managed the potential anisometropia with spectacle correction of both eyes, which was well tolerated. However, undercorrecting hyperopia in the normal eye may induce accommodative esotropia in some. Another approach is to undercorrect the pseudophakic eye by 1 to 3 D when it becomes excessively myopic; this allows that eye to continue to be used at near. Some surgeons aim to correct the child's eye to focus at near (equivalent to myopia of -2.0 or -3.0 D), even before age 2 years.⁸

If a child has reasonably good vision in his or her pseudophakic eye, a bifocal segment should be considered when a child needs good vision both at distance and near, perhaps at 2 to 3 years of age.⁹ My personal preference is to wait till school age, as the expense of bifocals can be excessive for parents who must change them frequently if a child breaks the glasses and as the eye grows.

In addition to bifocals, a small amount of myopic astigmatism can give good vision at distance and near.¹⁰ Thus, undercorrecting astigmatism in the pseudophakic eye will give a useful depth of field for many. I do this personally for my own presbyopic eyes.

Eventually the child will grow up, and the refraction will become reasonably stable. Of note, I do not know if (on average) the eyes ever stop growing. I think they may

continue to have an increased myopia on average through at least age 25. In fact, many otherwise normal adults who have refractive surgery eventually require a “touch-up” for recurrent myopia.

Once the patient is at least 18 years of age and the refraction has remained stable for a year, I think it is reasonable to consider refractive surgery in coordination with a thoughtful refractive surgeon. As long as there is not a contraindication, small errors (perhaps +3 to -8 D) can be corrected with corneal refractive surgery including laser-assisted in situ keratomileusis (LASIK) and photorefractive keratectomy (PRK). In patients in whom the refractive error is too great for LASIK or PRK to safely correct, exchanging the IOL for one of lesser power may be a better option. Unilateral refractive surgery may not be well tolerated in normal adults, but after childhood cataract the binocularity is reduced and the difference in image quality will not be noticed. There are few reports of this; my unilaterally pseudophakic son had correction of -8 D unilateral myopia with PRK and regained stereopsis as a result. My preferred goal for most such patients is emmetropia.

Hsuan et al.¹¹ reported implantation of Staar Collamer lenses in six pseudophakic eyes to correct anisometropia: all patients had a reduction in anisometropia to asymptomatic levels. However, the follow-up period was <1 year, too short to demonstrate the long-term safety needed for pediatric eyes. Although the anterior segment of the eye reaches close to adult proportions before age 5, implantation of these lenses in children has not been reported to my knowledge.

Always remember this goal: the child has good vision as an adult. In addition to refractive correction, the key to good vision is amblyopia treatment! In a study of 122 children who had cataract surgery, at 7 years' follow-up, poor compliance with occlusion was the factor

most strongly associated with poorer acuity. The odds of worse vision in unilateral cataract were 7.9 times greater when compliance was <50% than when compliance was complete.¹²

DISCLAIMER

The views expressed in this article are those of the author and do not necessarily reflect the official policy or position of the Department of the Navy, Department of Defense, or the U.S. Government.

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Correction of Aphakia: Glasses and Contact Lens

Rupal H. Trivedi and M. Edward Wilson

Primary intraocular lens (IOL) implantation has become increasingly common and is now the standard of care for older children undergoing cataract surgery. Although more and more surgeons implant IOLs in older children, the use of an IOL still remains controversial in infants and young children.¹ The Infant Aphakia Treatment Study (IATS) reported that there was no statistically significant difference in grating visual acuity at age 1 year between the IOL and contact lens (CL) groups in eyes operated for unilateral cataract; however, additional intraocular operations were performed more frequently in the IOL group.² Until longer-term follow-up data are available, caution should be exercised when performing IOL implantation in children aged 6 months or younger, given the higher incidence of adverse events and the absence of an improved short-term visual outcome compared with CL use.¹ Many very young children presenting with congenital cataracts are left aphakic (Fig. 45.1). A secondary IOL can be implanted later if needed.

Situations where an IOL implant may not be the primary surgical procedure (relative contraindication to IOL implantation), necessitating aphakic correction, are listed in Table 45.1. Because the resulting residual refractive error is amblyogenic, it is crucial to correct this aphakia.

Various modalities can help to correct pediatric aphakia. Table 45.2 describes the arguments *for and against* these approaches. For completeness, this table includes details for IOL implantation as well. Although many of the problems inherent in the use of aphakic spectacles can be overcome by the use of CLs, preference for one modality over the other depends on several considerations that are listed in Table 45.3.

APHAKIC GLASSES

Aphakic glasses are often used for the correction of bilateral aphakia in children (Fig. 45.2). Pros and cons of various forms of aphakic correction are listed in Table 45.2. Since IOL implants are commonly used for aphakic correction in adults, the availability of and advancement in

technology for high-power plus lenses (generally over 10 D [diopters]) have declined. Therefore, options are very limited for the child requiring prescription eyewear following cataract surgery. Details of aphakic eyeglass design and optics can be found in basic textbooks on optic and refraction, and are beyond the scope of this book.³ Instead, we furnish information designed to provide guidelines for parents and practicing clinicians published on the Pediatric Glaucoma and Cataract Family Association (PGCFA) Web site.⁴ In general, three primary types of high-power plus lenses are in use. Ultraviolet protection should be added to any lenses used.

- *Lenticular lenses.* These lenses have the prescribed power at the center of the lens surrounded by a “ring” of little or no power. Although these lenses are inferior to other types, they may be the only lenses that can be found for powers greater than +20 D.
- *Aspheric lenticular lenses.* These have a nonspherical (aspheric) central area surrounded by a ring with little or no power. Power ranges for the aspheric lenticular lens are commonly provided in the range of +10 to +20 D. The optical properties of this lens type are superior to those of the lenticular lens.
- *Multidrop lenses.* These lenses have a spherical central zone that flattens into an aspheric zone and then is blended into an area of lesser power. The lens resembles the aspheric lenticular lens without the noticeable “ring.” It is far superior to other lens designs. However, it is commonly available only in the range of +10 to +16 D.

Frame Selection

It is important that the line of vision rests as close as possible to the vertical center of the frame. Since the central portion of the lens has the greatest thickness and curvature, the depth of the frame should be sufficient to prevent exposure of the lens thickness along the top of the frame. Frame selection is therefore very important. When it can be used, multidrop lenses positioned

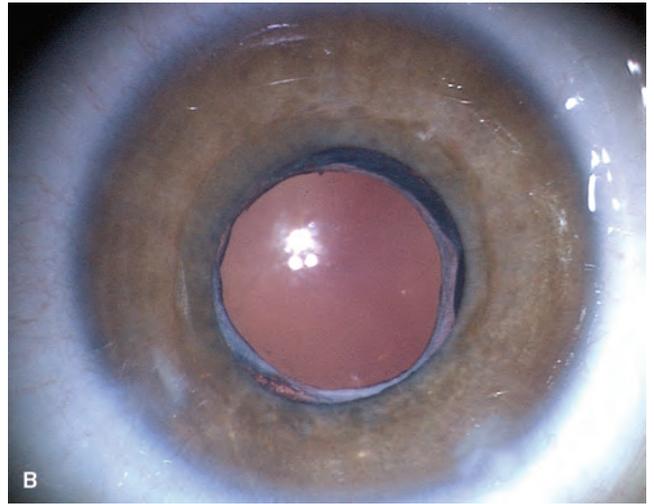
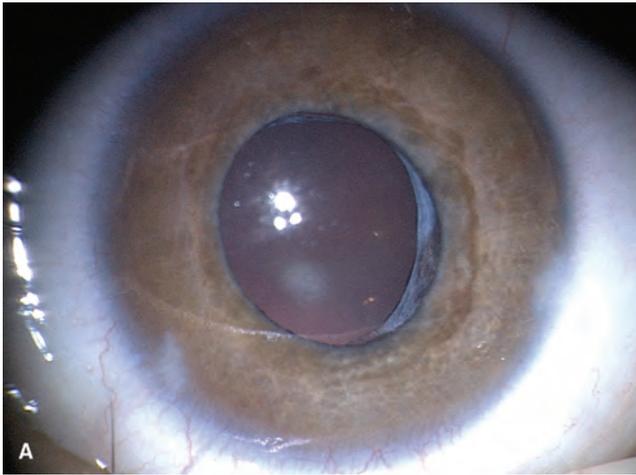


Figure 45.1 A and B: Three year follow-up of both eyes of a child operated for bilateral cataract at 1 month of age.

correctly in a small frame will offer the child the best optics and the best appearance. Some additional considerations for frame selection are listed in Table 45.4. Whenever possible, it is important to involve the child in the decision-making process.

CONTACT LENS

Pediatric aphakia can be successfully treated with CL (Figs. 45.3 to 45.7). The development of binocular vision and stereopsis has been reported after early removal of infantile cataracts in patients who show excellent compliance with CL use and occlusion regimens.⁵ A major advantage of CL use is that the power can be easily adjusted as the child's eye grows. Such lens wear can be both safe and effective with appropriate lens fit, good lens care, and compliance with proper hygiene. Otherwise, it may become hazardous. *Compliance* and *complications* are major factors that may limit the routine use of CL for pediatric aphakia in some settings.

Table 45.1

RELATIVE CONTRAINDICATIONS TO IOL IMPLANTATION

Institutional factor: Nonavailability of an IOL

Surgical factor: Surgeon prefers not to implant a lens in the patient.

Patient factors: Minimum age at surgery for an IOL implantation varies from surgeon to surgeon and varies between unilateral and bilateral cataracts.

Ocular factors: Vary from surgeon to surgeon-associated uveitis, severe microphthalmia such that IOL size is not feasible to implant, persistent fetal vasculature, inadequate anterior and/or posterior capsular support, etc.

Parental factor: Permission/consent denied

How Pediatric CL Practice Differs from That in the Adult

Pediatric CL practice is quite challenging as well as different from adult lens practice.

Repeated insertion and removal of a CL can be psychologically traumatic to a child as well as to the parents. The insertion and removal of the lenses on a daily-wear basis may be stressful. Extended wear is a popular feature for many parents.

Measuring the eye parameters of such young children is not easy, and assessment of the fit on the eye with a fixed or portable slit lamp may be challenging.

The conjunctival fornix is shallower and the globe is smaller than in the adult eye. The infant cornea is smaller and steeper and the vault is less than in the adult cornea.

The continuously changing refraction in pediatric eyes is primarily related to the increasing axial length (AL), but the progressive corneal flattening that occurs in the first few years of life also contributes. Corneal flattening has a larger effect on the fit of the CL.

Although there are differences between pediatric and adult CL practice, there are also some similarities between the two. The similarities include the basic fitting principles used to obtain well-fitted lenses and the physiologic requirements of the cornea and anterior segment. The optical lens characteristics and materials are essentially the same regardless of the age of the person wearing the lens. Lens-care techniques are identical for children and adults, although the method of handling the lens is modified for the pediatric patient.

Contact Lens Choices

There are three main types of CL: the hard lenses (including polymethylmethacrylate [PMMA] lens and rigid gas-permeable [RGP] lens), the hydrogel extended-wear

Table 45.2 ARGUMENTS FOR AND AGAINST DIFFERENT MODALITIES TO CORRECT APHAKIA

	Arguments For	Arguments Against
Aphakic glasses	<ul style="list-style-type: none"> • Power can be easily adjusted • Safest approach 	<ul style="list-style-type: none"> • Restriction of visual field to approximately 30 degrees • Because of marked retinal size disparity (approximately 30% magnification), not suitable for long-term monocular aphakia • Distortion of image and prismatic effect • Heavy weight of the glasses may be cumbersome • May be debilitating visually, cosmetically, and psychologically
Contact lenses	<ul style="list-style-type: none"> • Power can be easily adjusted • Suitable for unilateral aphakia • Image-size differences lessened compared to glasses • Field of vision better than with glasses 	<ul style="list-style-type: none"> • Noncompliance of both parents and patients • Difficulty of insertion • Corneal complication • Frequency of lens loss • Cost • Psychologically traumatic • 5%–9% magnification
Intraocular lens	<ul style="list-style-type: none"> • Immediate and constant visual input • Maximum compliance with amblyopia therapy • Minimum anisometropia, which is especially important in unilateral cataract 	<ul style="list-style-type: none"> • Frequent changes of residual refractive error • Correct sizing still a problem in severe microphthalmic eyes • Concern for long-term safety of IOL • High incidence of visual axis opacification in infantile eyes

or daily-wear lenses, and the silicone lenses. Advantages and disadvantages of various CL materials are listed in Table 45.5. Some commercially available, flexible CLs for aphakia are listed in Tables 45.6A and B. The silicone CL combines the best features of hard and soft lenses. The silicone CL can mask up to 2 D of astigmatism. Aasuri et al.⁶ reported that these lenses are safe, provide satisfactory optical correction, and are easy to handle. The limited availability of CL and the financial cost (ca. US \$170/unit) associated with frequent lens replacement are limitations to CL use in developing countries. The average annual CL replacement costs ranged from \$1,275 for RGP wearer to \$1,925 for silicone, which represents approximately 17 lenses per year for RGP and 11 lenses per year for silicone lenses.⁷ A disadvantage of silicone CL in older children and adults is that they may be uncomfortable initially. Other disadvantages are their hydrophobicity and adhesion effects. Since infants have more watery tear layers and produce less mucus, the drying and discomfort of the silicone elastomer lens, as noted with

adult wearers, are not as common in younger children. Silicone lenses also have a tendency to acquire deposits when drying, which can alter the lens surface characteristics. The most important feature of silicone elastomer is its high oxygen permeability. If a SilSoft CL cannot be worn by a child, a RGP lens can be tried. The aphakic eye is potentially more vulnerable to retina changes related to ultraviolet exposure because the crystalline lens filters UV light. Silicone CLs do not provide UV protection, whereas RGP CLs may be ordered with UV blockers. This is a theoretical advantage for RGP lenses over silicone lenses.⁸ Saltarelli⁹ reported on outcomes of the use of a hyper-oxygen-permeable rigid CL material, Menicon Z, for the fitting of aphakic infant eyes. The authors noted that the material had excellent fit characteristics, was not damaging to the ocular structures, was relatively

Table 45.3 FACTORS TO CONSIDER WHEN DECIDING BETWEEN APHAKIC EYEGLASSES AND CONTACT LENSES

Unilateral aphakia versus bilateral aphakia factors: Aphakic spectacles are not appropriate for monocular aphakia because of relative magnification differences; if possible, CLs should be tried in such a situation. Eyeglasses can be used for bilateral aphakia with reasonable success.

Institutional and compliance factors: When good CL care is not available, eyeglasses can be offered.

Cost factors: Available silicone CLs are expensive. The need for repeated lens purchase because of changing refractive error or lost lenses makes the cost even higher.



Figure 45.2. This 3-year-old boy is bilaterally aphakic and has aphakic glaucoma controlled with topical medications. He is shown wearing aphakic spectacles.

Table 45.4 FACTORS TO CONSIDER WHEN SELECTING A FRAME FOR PEDIATRIC EYES

- The *smallest* frame is the best choice for children.
- A frame with *strong color* will help direct attention to the frames and away from the lens.
- The *bridge of the frame* should carefully conform in shape to that of the child. If the bridge of the frame is too high, the child's nose will appear longer. A lower bridge will make the child's nose appear shorter.
- *Cable temples* (earpieces) that wrap around the back of the ear are recommended. This will help to keep heavier lenses positioned correctly in front of the child's eyes. Temples should not extend past the ear lobe.
- *Spring hinges* will absorb a lot of abuse and are considered a must-have for children.

easy to manipulate for caregivers, and maintained its integrity well throughout the course of the study.

Daily-Wear Versus Extended-Wear Lenses

Extended-wear CLs are, in many ways, ideal for children with aphakia. However, owing to the increased incidence of severe complications (e.g., acute red eye reaction, giant papillary conjunctivitis, neovascularization, abrasion, infective keratitis) associated with the use of some extended-wear designs and materials, daily wear is recommended by some experts for all lens types. The majority of pediatric ophthalmologists in the United States use SilSoft extended-wear lenses for young aphakic children. These lenses are designed and approved for continuous use up to a maximum of 30 days between removals for cleaning and disinfecting (U.S. FDA approved up to 30 days extended wear). However, the risk of ulcerative keratitis is greater with extended-wear CLs compared to daily-wear CLs. The risk among extended-wear lens users increases with the number of consecutive days that the



Figure 45.3. A 6-month-old child with a unilateral cataract operated on at 2 weeks of age. Patching (OD) and a SilSoft CL (OS) are tolerated well.

lenses are worn between removals. We typically advised removal and cleaning every week, even though they can be used for continuous use up to a maximum of 30 days.

Bausch & Lomb SilSoft Pediatric Patient Assistance Program: The Bausch & Lomb (B&L) Web site contains information on a patient assist program for SilSoft users. An application can be downloaded from the site. The B&L Patient Assistance Program is designed to assist pediatric patients and their families who have no medical device coverage through private insurance or from public assistance (i.e., Medicaid, Medicare, or charitable organizations) and fall below the U.S. Health and Human services 2004 poverty guidelines, (family of four with an annual income of <\$18,850 and for each additional person, add \$3,180). Requests for free SilSoft lenses will be accepted once per calendar quarter per patient. A maximum of two lenses is allowed per patient per order.

Initial Fitting: Infants who are aphakic should be fitted with a CL as soon as possible (Fig. 45.4A–C), preferably immediately after the cataract surgery. The need for high plus refractive power leads to lenses with remarkable center thickness. To avoid decentration (especially low riding) or lens loss, the lens diameter must often be as large as the cornea, and a relatively steep base curve is indicated. The SilSoft lens can be fitted near the flat keratometry reading. Although keratometry can be revealing in many cases, it usually is not essential for fitting silicone CLs. Final CL power for the infant and young child is most easily determined by retinoscopy over a trial CL of an estimated power. A lens with a back optical zone radius (BOZR) of 7.5 mm, a total diameter of 11.3 mm, and 32 D is generally inserted as first choice in small infants. However, even the steepest available BOZR (7.5 mm) is often too flat for aphakic infants, and the highest available plus power (32 D) can be below the required aphakic prescription during early infancy. These lens power suggestions are a starting point before the necessary retinoscopic overrefraction. Martin et al.¹⁰ provided initial fitting guidelines based on keratometry readings. 45 to 46 D, 7.3 base curve, 11.3 mm diameter; 43 to 45 D, 7.7 or 7.5 base curve, 12.5 or 11.3 diameter; 41 to 43 D, 7.9 or 8.1 base curve, 12.5 diameter; <41 D, 8.1 or 8.3 base curve, 12.5 mm diameter. The SilSoft SuperPlus pediatric CLs are manufactured in only one diameter (11.3 mm) and in only three different base curves (7.5, 7.7, and 7.9). The dioptric powers available are +32, +29, +26, and +23. The aphakic series contains powers from +11.5 to +20 D in 0.5 D steps. In these lower powers, base curves of 7.5, 7.7, 7.9, 8.1, and 8.3 are available along with 2 diameters (11.3 and 12.5 mm).

Lightman and Marshall¹¹ published their recommended base curves and powers for an initial lens choice in the absence of keratometry readings (0–6 months, +29 D; 7–17 months, +26 D; 18–28 months, +23 D; 29–34 months, +18 D). In a prospective study, de Brabander et al.¹² noted that the BOZR remained stable at 7.5 mm, up to the age of 1.5 years. At the age of 4 years, almost all

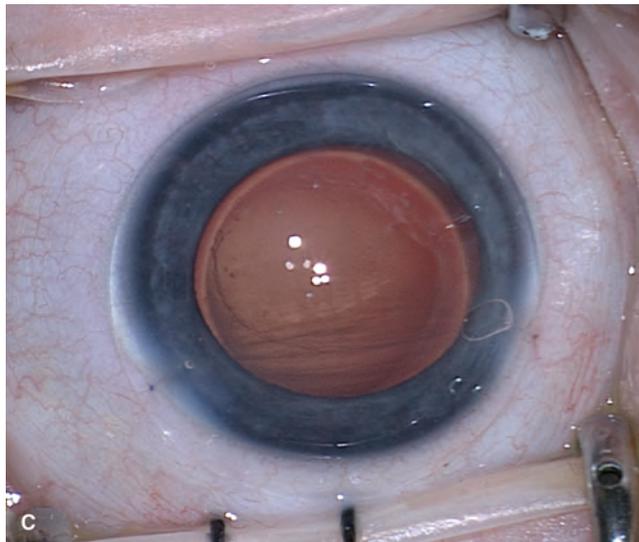
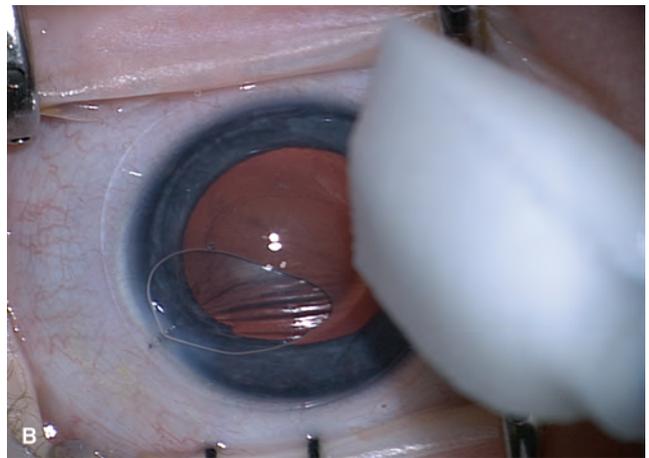
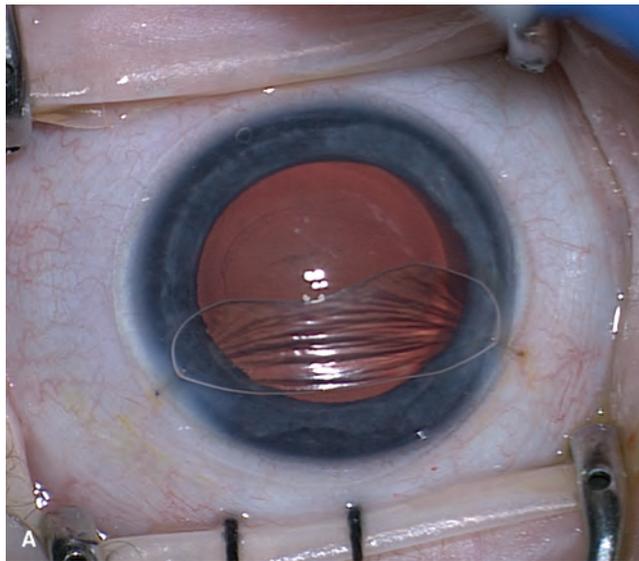


Figure 45.4 A–C: CL applied on table at the end of cataract surgery.

eyes needed a BOZR of 7.9 mm. CL power was $+25.47 \pm 4.0$ D (range $+32.00$ to $+20.00$ D) at 3 months of age and 17.94 ± 3.8 D (range $+29.00$ to $+15.00$ D) at 3 years of age. Regression analysis revealed that CL power = $84.4 - 3.2 \times \text{AL}$ ($R^2 = 0.82$; $P < 0.001$).¹³ CL power can also be estimated using an A-constant of 112.176 in IOL power calculation formula. In the absence of refraction, a 32 D CL has been used by some as the initial CL power. However, we noted that if 32 D CL had been used, 44% eyes would have needed a replacement of CL. Refraction over CL in our series ranged from -13 to $+11$ D, and based on this wide range CL replacement was needed within a week after initial insertion. Considering an approximate overcorrection of $+2$ D and the availability of SilSoft CL powers, we recommend a 32 D CL when the preoperative AL is < 17 mm; 29 D when the preoperative AL is between 17 and 18.5 mm; 26 D when the preoperative AL is 18.5 to 19.5; 23 D for 19.5 to 20 mm (21 mm); and

a 20 D CL for an AL of 20 to 21 mm (20 D for > 21 mm). These power suggestions are a starting point before the necessary retinoscopic overrefraction. When the CL power falls between available powers (SilSoft superplus lenses are available in 3 D steps), a customized decision should be made by the physician. For example, if an eye requires 30.5 D, a $+32$ D can be chosen since the infant's visual needs are mostly at near. However, a $+29$ D can be chosen if replacing a CL in the near future will be a burden, as it is likely that the eye will grow very fast in this age group and soon this eye will require $+29$ D.

Using a penlight, a magnifying lens, and an ophthalmoscope or a handheld slit lamp, the movement and centering of the lens can be evaluated a few minutes after insertion. Lens movement with blinking is the most critical and important factor for evaluating the CL fit. If the lens moves too much or decenters, the total diameter can be increased to 12.5 mm. When there is

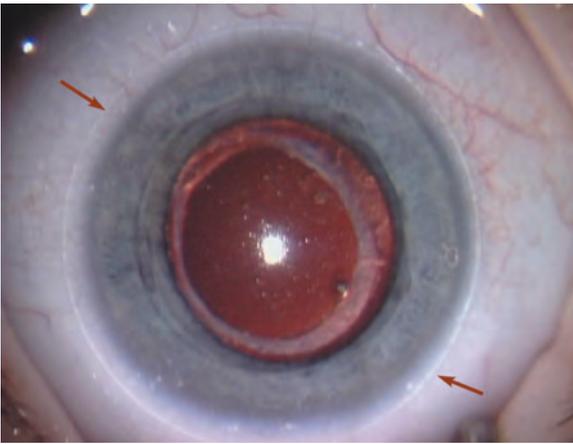


Figure 45.5. A silicone CL in pediatric aphakia.

insufficient movement, a lens with a flatter BOZR should be used and evaluated again. A fluorescein pattern may be used during the fitting sequence as needed. Lens power can be calculated from retinoscopy. An addition of 4 D at birth, 3 D at 1 year, 2 D at 2 years, and 1 D at 3 years can be given to assure optimal focusing for near.

For RGP lens fitting, a base curve should be selected 1 to 1.5 mm steeper than the flattest keratometry reading. For hard lens fitting, a set of trial lenses (all 10 mm in diameter) with base curves varying from 7.2 to 8.2 mm at 0.1-mm increments is often used. For hard lenses, a study of the fluorescein pattern is made and the lenses changed if indicated. Fluorescein evaluation with these template lenses in situ allows an experienced observer to determine the lens that best aligns with the underlying cornea. A “steep” lens produces distinct central pooling of fluorescent tears, perhaps with bubbles, a “flat” lens produces an absence of fluorescence centrally, and a lens in alignment with the corneal curvature produces a uniform distribution of the dye. The major problem with fitting young children is stability. To overcome displacement due to the small size of the eye and high tonicity of the

lids, it is necessary to fit a lens that is both tight and relatively large in comparison with the fitting criteria for an adult eye. In one report, it was found necessary to fit as steep as 0.1 mm flatter than the flattest corneal reading.¹⁴

For a routine comprehensive follow-up examination, we recommend an examination under anesthesia one year after cataract surgery in an infant. However, for the CL fitting, there may be significant differences in the fitting characteristics of infants when examined under anesthesia compared to when assessment is performed while they are awake.^{15,16} Thus, CL fitting in the examination room is preferred. In the awake state, the centration of the CL and the evaluation of the lens movement are more reliable.^{15,16}

Contact Lens Insertion and Removal

The success of the CL fitting, and ultimately of the child’s vision correction, often depends on the training and understanding of the parents or caregivers who will be responsible for the child’s eyes and lenses. The parent who is going to take the most responsibility for the lens-wearing schedule and the insertion (Fig. 45.6), removal (Fig. 45.7), storage, and disinfections of the lens should be encouraged to attend the clinics as much as possible to learn these techniques and to develop a relationship with the practitioner. This aids in communication, should any CL problems arise in the future. From the outset, parents should always be present and included in such procedures as insertion and removal of a CL. In this way, they will learn about the handling of the CL and their responsibilities toward the success of their child’s CL treatment. Each step should be practiced in the office, under supervision. Parents/caregiver should not only learn how to apply, remove, and care for the lenses but also how to make these activities a simple daily or weekly routine. Written instructions regarding the fundamentals of hygiene, handling, insertion, and removal of the lenses should be given to reinforce oral instructions. The clinician should tell the



Figure 45.6. Insertion of CL. **A:** Teaching mom to insert a silicone CL. **B:** Parent successfully inserts a silicone CL in the office.



Figure 45.7. Removing a silicone CL. **A:** The eyelids are gently pulled apart. **B:** Pressure is applied to the superior and inferior lens edge as the thumbs are brought together. The lens lifts out onto the skin.

parents what to observe in terms of a steep (nonmoving) lens, which may induce injection or a sectorial or complete compression “ring” indentation in the sclera (e.g., when using the hydrogel lens); or a loose lens, which can produce some edge lift, increased lens excursions, and poor centering. The parents should also be told that the child’s eye should “quiet” quickly after CL insertion; but if the child continues to cry for unusually longer duration, the lens may be torn, chipped, cracked, or inside-out, or there may be a foreign body underneath it. If the child continues to be uncomfortable, the lens should be removed and inspected. Parents should be instructed to routinely check the eyes of their child. They should be instructed to look for a clear reflex (a shiny luster) from the CL front surface because this verifies the wettability of the lens. If dull areas are observed, parents can apply

a saline solution containing no preservative. Parents can also routinely insert one drop of saline (no preservatives) when the child awakens. Parents should be taught how to remove, clean, and insert the lens and should be advised that cleaning should only be done when re-wetting fails. If the lens is dull even after cleaning, the hydrophilic coating on the lens is likely worn off and the lens should be discarded and replaced. As long as the surface is wet and the eye remains clear, the lenses can be worn for the full week in between cleanings. If the eyes became red or there was any doubt about the lens or eye condition, the parents should be instructed to consult the clinic immediately.

When the clinician is convinced that the parents can adequately care for the lenses, they can be given a lens-wearing schedule for the child and a return appointment. Wearing time for daily-wear lenses is rapidly

Table 45.5 ARGUMENTS FOR AND AGAINST VARIOUS CL MATERIAL

	Arguments For	Arguments Against
PMMA	<ul style="list-style-type: none"> • Available in a wide range of prescriptions • Can be customized to power and base curve • Comparatively inexpensive • Good optical performance (in most cases neutralizing astigmatic and spherical components of refractive error) • More durable and easy to handle. 	<ul style="list-style-type: none"> • Must be removed daily • Some initial discomfort • Occasional lens breakage and loss
Soft material	<ul style="list-style-type: none"> • Comfort 	<ul style="list-style-type: none"> • Frequent and rapid lens loss • Poor correction of residual refractive astigmatism • Difficulty in lens insertion
Silicone	<ul style="list-style-type: none"> • Superior corneal oxygenation • Easy to handle, durable • Relatively low loss rate • Can be fitted using either ocular measurements or trial techniques. 	<ul style="list-style-type: none"> • High cost • Availability may vary in certain parts of the world • Inability to obtain full optical correction because available in limited powers • In some children, can be easily ejected and possibly lost • Surface may be damaged by handling. Once it is damaged, it attracts protein and mucus and lens becomes unwearable

PMMA, polymethylmethacrylate.

Table 45.6 A. HIGH-POWER CONTACT LENS, >20 D

Manufacturer	Lens Name	Parameters Availability
Bausch & Lomb	SilSoft Super Plus pediatric lenses	+20 to +32 (3 D step) BC: 7.5, 7.7, 7.9 Diameter: 11.3 Optical zone: 7 mm
Flexlens	Flexlens Aphakic	0 to +50 D in 0.25 D step BC: 5–11 mm in 0.1 mm steps Diameter: 8–16 mm 0.1 mm steps Anterior optic zone: 6.5 mm (pediatric), 8 mm (adult) Daily wear

BC, base curve; D, diopter.

increased to all waking hours if no particular problems are encountered. Parents should be certain that the lenses are always adequately cleaned and disinfected before reinsertion. For silicone lenses, weekly removal for cleaning is usually recommended. Strict hygiene is important for any CL use, but it is especially important with recent surgery.

Topical anesthetics are never used to facilitate either the evaluation of the fitting of CL or the teaching of insertion and removal techniques. The greatest obstacle to the insertion of a lens is the child's fear. Topical anesthetics have little effect on this critical issue, and they also give a false

Table 45.6 B. HIGH-POWER CONTACT LENS, 10–20 D

Manufacturer	Lens Name	Parameters Availability
Bausch & Lomb	SilSoft aphakic—adult lenses	+11.5 to +20 D (0.5 D steps) BC: 7.5, 7.7, 7.9, 8.1, 8.3 mm Diameter: 11.3, 12.5 mm Optical zone: 7 mm
Proclear	Proclear Compatibles	–20 to +20 D
Cooper Vision	Hydrasoft	+10.5 to +20 D BC: 8.3, 8.6, 8.9, 9.2 Diameter: 14.2, 15 mm
Flexlens	Flexlens Aphakic	0 to +50 D in 0.25 D step BC: 5–11 mm in 0.1 mm steps Diameter: 8–16 mm 0.1 mm steps Anterior optic zone: 6.5 mm (pediatric), 8 mm (adult)
Menicon	Menicon Z	Power: –20 to +20 D Diameter: 8.8–9.6 mm

impression as to what the child will experience when the lenses are inserted at home. Some suggestions follow.

- *When the patient is <2 years of age.* Insertion in this age group is more easily managed than in the older toddlers. The practitioner should pull up the baby's upper lid and insert the lens under it, then pull the lower lid over the lower edge of the lens. The lens should then be checked to ensure that it has not folded during insertion.
- To remove a silicone or soft CL in a child, the lids are pulled apart as much as possible and gentle pressure is put on the superior and inferior lens edges; this produces an interruption of the suction and allows the lens to be lifted out by the lids. If there is struggling, the lids may be squeezed tightly shut and this may actually help ejection of the lens as the practitioner pulls the lids apart. Once the child is old enough, the lens can be pinched off in the conventional way.
- *When the patient is between 2 and 5 years of age.* Insertion may be easier if the child is laid on a bed. With time, no holding is required, and eventually the lenses can be inserted while the child is sitting in the chair. To remove the lenses, the same method as described for younger children can be used, although sometimes holding the head may be necessary.
- *When the patient is >5 years old.* It may be possible to encourage the children to begin to manage insertion and removal themselves. Insertion is not always easily achieved at this age, so initially help is required from parents to hold the lids or guide the child's finger. Once children have gained confidence and appreciate the advantages of being independent, their insertion technique improves, enabling them to handle the lenses without any assistance. To remove the lenses, children may pinch off soft CL.

Contact Lens Follow-Up

Care of the patient after the CL is fitted is a shared responsibility between the parent or caregiver and the practitioner. Follow-up visits are generally scheduled at 1 day, 1 week, 1 month, and every 3 months thereafter. Questions at each visit may include the following: Have there been any difficulties handling the lens since the last visit? Have any lenses been lost? Has there been any irritation of the eyes? Was any visual progress observed? Excessive blinking, photophobia, tearing, conjunctival injection, or discharge may indicate the possible presence of conjunctival or corneal pathology. Careful slit-lamp examination after CL removal, with and without fluorescein staining, is done as needed. Parents should be queried regarding any untoward reaction. Lens-wearing time is gradually increased, starting with a few hours at first. Follow-up keratometry is recommended. The corneal radius and diameter change over the first few years. The possibility therefore exists that if an extended-wear lens is deliberately fitted tight and is left in situ for too long,

the cornea will grow beneath it and the fit will become tighter. This can lead to complications. Such a tight lens will cause corneal edema and conjunctival chemosis and, in turn, cause embarrassment of the limbal capillaries. If a regular follow-up pattern is maintained, little difficulty will arise. During follow-up visits, an ophthalmologic examination should be performed that includes an assessment of movement, centration, and condition of the lens. An overrefraction should be done at each visit, measuring any residual refractive error that remains with the CL in place. At times, the lens is removed and the refraction is repeated at the spectacle plane. For bilateral cataract patients, an up-to-date aphakic spectacle refraction will allow glasses to be ordered as a backup. If CLs are lost or if the eye is red, glasses can be used until the CL is refitted. Vertex distance is very important in CL fitting and refracting for aphakic infants. If the spherical equivalent is +23 in the spectacle plane and if a 10-mm vertex distance is assumed, the CL power would be 29.9 D. However, if a 12-mm vertex distance is assumed, the CL power needed increases to 31.8 D. Small differences in vertex distance can make a significant difference in the measured refractions.¹⁷ The examiner should select a CL with power within several diopters of the calculated power, and the refraction should be repeated and refined with the CL in place. Some authors routinely exchanged the CL every 3 months. We prefer to exchange the CL only when it is required. If possible, the parents should maintain a spare lens, and spare lenses should be stocked in the clinic if possible. At a minimum, the clinic should maintain enough lenses to allow a CL overrefraction when a baby comes in after a lens has been lost. This avoids the hazards of refracting the eye in the spectacle plane and trying to accurately calculate the CL power that needs to be ordered.

As the eye grows, the aphakic power requirements will decrease. Also, the cornea changes rapidly in young children, increasing in overall diameter and decreasing in radius of curvature. Thus the importance of frequent early visits cannot be overemphasized. We continue to see aphakic CL patients every 3 months until we are sure that the follow-up frequency can safely be lengthened.

In children with bilateral aphakia who wear CL and have unilateral amblyopia, the CL of the better eye can be removed for a few weeks if needed for amblyopia therapy. Since the fellow eye is aphakic (or rarely pseudophakic), this form of optical penalization can be very effective.¹⁸

Considerations and Complications of Contact Lens

- *Lens loss.* Frequent CL loss is commonly observed in infancy and even more so in the toddler years. Babies only a few months of age can easily rub the lenses out during sleep. Jacobs¹⁹ reported that one mother even attempted to recover a lens from an infant who was seen to remove it and then swallow it. Her efforts were unrewarded. For continued optical correction to prevent amblyopia, a spare set of lenses should always be available through the clinic, and ideally, with the parents. An up-to-date pair of spectacles must be available in case of CL loss or any other CL problems. Once on a daily-wear schedule, the problem of loss decreases, and eventually lens deterioration becomes the major cause of lens replacement, occurring approximately every 6 months. de Brabander et al.¹² reported lens usage as 5.6 lenses/eye/year (including the regular exchange every 3 months).
- *Noncompliance.* Noncompliance of both the parents and the child is the major obstacle in CL practice. Loss of CL, conjunctival erythema, and poor lens fit are reasons for noncompliance in pediatric patients. Poor compliance in children is multifactorial, but the main contributing factors are difficulty in insertion/removal and high cost. For unilateral aphakia, good vision in the fellow eye tends to promote noncompliance. When CL wear fails, it does so between age 2 and 4 years most commonly.
- *Infection.* Minor infections can occur from time to time, especially if the child is using soft extended-wear contacts. The parents must always be carefully alerted to the removal of a lens if they see the slightest redness of the bulbar conjunctiva. If at this stage they are unable to handle the lens, then they must get ophthalmic help. If the infant is bilaterally aphakic, removal of both lenses is advisable, as amblyopia can quickly develop in the eye without the lens. Aphakic spectacles should then be worn until refitting of the CL.
- *Corneal vascularization.* Due to various degrees of anoxia in soft CL wearers, corneal vascularization can occasionally be observed with CL use. In such a situation, a child using extended-wear lenses must be refitted with daily-wear lenses, or a child using daily-wear soft lenses, with RGP lenses. Otherwise, spectacles can be used for bilateral aphakia and even, at times, for unilateral aphakia when all else fails. Glasses may also be needed, at least temporarily, for CL-induced giant papillary conjunctivitis. Some CL problems are more common in the rural poor in the United States or in the developing world. These include infectious keratitis, corneal vascularization, hypoxic corneal ulceration, and red eye without ulcerations.
- *Power changes.* Frequent follow-up is needed, as the power changes occur very rapidly. The younger the child, the more frequent the follow-up. de Brabander et al.¹² reported average decrease of 0.23 D/month. The power decrease in unilateral cases was significantly lower ($P < 0.01$) than in bilateral aphakia. The rate of power change in the de Brabander study was 0.32 D/month and for Lightman's study it was 0.35 D.¹¹

- **Deposits.** de Brabander et al.¹² reported a positive correlation ($r = 0.89$) between age and deposit buildup. These authors reported that from the age of 3 years on, deposit formation was the determining factor for refitting out of silicone lenses and into high-water content soft or RGP lenses.
- **Parental stress.** Psychological stress (to parents, caregivers, or patient) can be an obstacle when prescribing CL to children. However, Ma et al.⁸ reported that CL seemed to be well tolerated by most patients, as assessed by caregivers. Although initial resistance to CL use is high, this often decreases with time.
- **Swimming.** A question that is often asked is whether or not it is OK to wear contacts while swimming. Swimming in contacts does increase the potential for the lens to be exposed to bacteria and other organisms (especially when swimming in fresh water). Such infections can be potentially blinding. The potential of a lens being lost during swimming increases only slightly. Fresh water has the effect of making the lens adhere or stick to the eye more tightly. Well-fitting swim goggles should be worn. Thorough cleaning and disinfection of the lens and case are extremely important to reduce the risk of contamination. Diligence in the care and cleaning are even more important.

SUMMARY

In cases in which it is not feasible to implant an IOL, CL can be the first line of treatment, at least in unilateral cases. However, in bilateral cases, aphakic glasses are still a reasonable option. When patients are not compliant with either aphakic eyeglasses or CL, it may be necessary to implant a secondary IOL. Silicone CLs are easy to fit and very well tolerated in infants and toddlers. From the age of 4 years on, as tear composition changes, silicone lenses may become less tolerated and may need to be replaced by daily-wear high-water content soft lenses, or, at a later age, replaced with RGP lenses. It is common in our practice to use silicone CLs until the preschool ages and then proceed to a secondary IOL. Some patients tolerate silicone lenses even into the second decade and do

not need secondary IOLs. However, cleanings need to be more frequent and lens replacements for deposits and nonwettability are more common. Refitting to a nonsilicone CL material or changing to aphakic spectacles may be needed in older children if IOL implantation is not advisable.

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Visual Outcome after Pediatric Cataract Surgery

Rupal H. Trivedi and M. Edward Wilson

Despite the continuing threat of amblyopia, the prognosis for a good visual outcome after cataract surgery in children has improved over the last few decades. Not only has there been a refinement of the surgical techniques and technology, there has also been an improvement of our understanding of the sensitive periods for the development and reversal of amblyopia. Age at onset, laterality (unilateral versus bilateral), and associated ocular and systemic diseases are important nonmodifiable factors influencing visual outcomes. On the other hand, duration of deprivation, correction of residual refraction, and amblyopia therapy are, at least in part, modifiable factors influencing visual outcome. Table 46.1 describes factors associated with visual outcome after pediatric cataract surgery.

The literature on visual outcomes in pediatric cataracts presents a wide spectrum of results. The heterogeneous nature of the data makes comparison of the studies very difficult. For example, it is difficult to compare two studies when (1) Study A's inclusion criteria are age at surgery <1 years, while study B states <18 years; (2) Study A's inclusion criteria are age at last follow-up >5 years, while study B includes patients at any age; (3) Study A involves the majority of unilateral cataract cases in contrast to study B that has a majority of bilateral cataracts, and the laterality of the cataract is not accounted for in the analysis of either study; (4) Study A includes pediatric traumatic and other complicated cataract, while study B lists them as an exclusion criteria; (5) Study A is reporting visual outcome of only those with quantifiable visual acuity (VA), while study B has reported overall median visual outcome; (6) Study A has follow-up range from 1 month to 1 year, while study B includes those who have minimum follow-up of 1 year; and (7) Study A uses an ETDRS chart, while study B uses an HOTV chart. To make matters worse, for bilateral disease, some studies include data from both eyes, without considering the correlation effect in statistical

analysis, while others have randomly selected one eye of bilateral cataract patients for analysis. Some studies have reported the mean of LogMAR VA in contrast to others reporting an arithmetic mean of denominator of Snellen VA or of decimal VA. As seen above, readers should pay particular attention to methodology including the inclusion–exclusion criteria.

We reported the visual outcome (measured at age 4 and older) of pediatric eyes that underwent cataract extraction with primary intraocular lens (IOL) implantation.¹ One-hundred and thirty nine eyes met inclusion criteria. Median age at surgery was 5.1 years (range, 0.03–16.9); the median age at last follow-up was 9.0 years, and the median follow-up was 3.6 years. The median corrected distant VA was 20/30. Forty-five patients had a final VA worse than 20/40. Of these, 34 (76%) had a diagnosis of amblyopia as the sole cause. Lim and colleagues reported the outcome of 778 patients (1,122 eyes) diagnosed with cataract over 10 years. Including only patients with quantifiable VA, the authors found that a final VA of 20/30 or better was observed in 39%.²

Age at cataract development is significantly associated with adverse visual outcome. Age at the cataract diagnosis is not necessarily the age at which the cataract developed. Screening and detection of cataracts at the earliest is very useful. Birch and Stager reported that when cataracts are dense at birth, there exists only a 6-week window of time beginning at birth, during which treatment of a dense congenital unilateral cataract is maximally effective.³ However, even late operated cataracts get some functional visual outcome. In 1842, Stafford reported a case of congenital cataract where sight was acquired by operation at the age of 23 years.⁴ The authors reported that the acquisition of sight was very gradual. At first, all was confusion. The 3rd week the patient began to distinguish objects and to be conscious of the differences of one thing from another. The patient was aware, when a piece of rag was waved before her eye, that something

Table 46.1 FACTORS ASSOCIATED WITH VISUAL OUTCOME IN CHILDREN WITH CATARACT**Preoperative**

- Age:
 - Development of cataract
 - Surgery
- Duration between diagnosis of cataract and cataract surgery
- Cataract:
 - Unilateral versus bilateral cataract
 - If bilateral: symmetrical/asymmetrical
 - Type
 - Density of cataract
- Visual function:
 - Poor VA
 - Strabismus
 - Nystagmus
 - Dense amblyopia
- Axial length
- Interocular axial length difference
- Associated ocular anomalies
- Associated systemic syndrome
- Targeted refraction if IOL implantation is planned

Intraoperative

- Major complication
- IOL implantation: yes/no

Postoperative

- Refraction
- Complications: Untreated posterior capsule opacification (PCO)/glaucoma/retinal detachment (RD)/cystoid macular edema (CME)/endophthalmitis
- Noncompliance to amblyopia therapy/correction of residual VA

was moving backward and forward. In a month, she differentiated black and white from color. Her knowledge of form and color rapidly improved.

The eyes of patients who have bilateral cataracts have a better prognosis after surgery than do the eyes of those affected by unilateral cataracts. In 1956,⁵ Crawford reported, “*I have today seen a patient who makes one feel more cheerful about the results of operation for congenital cataract. Both eyes were operated on at about the age of 15, at Moorfields, by Mr. Adams. The patient is now aged 90 and with glasses can see 6/9 with the right eye and 6/6 with the left, and can read the smallest print with comfort. He has sought advice only because one of these venerable spectacles has been broken.*” The presence of a sound, healthy eye frequently causes a delay in presentation, diagnosis, and treatment. Patients who have unilateral cataracts also have a higher incidence of coexisting ocular defects. In the past, some argued whether it is worth operating eyes with unilateral cataract as most children were not benefited from surgery.

Now, we know that although treatment is frequently frustrating, functional visual outcome may be achieved in some children with monocular cataract. Chak and colleagues⁶ reported a median postoperative VA of 6/60 in unilateral and 6/18 in bilateral disease. Median age at surgery was 2.99 months in unilateral and 4.6 months in bilateral cases. We reported median VA of unilateral and bilateral cases being 20/40 and 20/25, respectively.¹ Hussin and Markham evaluated the visual outcome of IOL implantation after cataract surgery in children. The examination for the prospective part of this study included the recording of VA. Mean final VA for the unilateral group was 0.91 logMAR, and for the bilateral group, it was 0.57 logMAR (Snellen equivalent 20/162 and 20/74, respectively).⁷

Even during the first few months of age, a cataract located anteriorly is less amblyogenic as compared with a cataract located posteriorly. Similarly, at the same age, and similar laterality, dense cataracts are more amblyogenic as compared with mild cataracts. Bilateral symmetrical cataracts would be less amblyogenic as compared to bilateral visually significant asymmetrical cataracts.

Preoperative interocular axial length difference helps to predict the postoperative visual prognosis. Eyes with longer axial length as compared to fellow eyes may imply a greater severity of amblyopia. Gochnauer et al.⁸ reviewed records from 64 children with unilateral cataract and found that deprivation amblyopia is likely severe when the interocular axial length difference exceeds 0.5 mm.

Children who underwent congenital cataract surgery developed poorer VA if the cataract was a part of a systemic disease as compared with children without a systemic syndrome.⁹ Francis and colleagues reported visual outcome in patients with isolated autosomal dominant congenital cataract.¹⁰ Almost half of the patients achieved a VA of 20/40 or better. Patients with isolated inherited congenital cataract have a better visual and surgical outcome than those with coexisting ocular and systemic abnormalities. The improved prognosis is related in part to the lack of other developmental abnormalities of the eye, and because inherited cataracts are often partial at birth. If partial, surgery may be delayed to later infancy and childhood when there is a lower incidence of surgical complications and refractive correction is easier. Chak and colleagues,⁶ using multivariate analysis, reported that in bilateral disease, the odds of worse vision were increased by the presence of additional medical conditions (3.53, 95% CI, 1.08–11.44) and the presence of postoperative ocular complications (2.94, 95% CI, 1.38–6.51).⁶ The cortical visual pathways may also have coexisting defects that influence the visual outcome. Decreased function of the visual pathways associated with global developmental delay may adversely affect the visual outcome.

The presence of preoperative strabismus and nystagmus predicts worse postoperative visual outcomes. For

a more thorough discussion of strabismus in cataract surgery patients, go to Chapter 48. Nystagmus is discussed here. Rabiah et al.¹¹ reviewed 95 cases of bilateral congenital cataracts presenting with sensory nystagmus at the time of their cataract surgery. Nystagmus was reduced or eliminated postoperatively in 40%. Mild, as opposed to severe, preoperative nystagmus was predictive of both a better VA outcome and reduced or eliminated nystagmus postoperatively. The authors concluded that good or even excellent VAs were, on occasion, possible after cataract surgery in children with bilateral cataract and sensory nystagmus. In these children, the nystagmus improved or disappeared postoperatively. In another study, the visual results of 13 children (26 eyes) with bilateral congenital cataracts associated with preoperative nystagmus were reported. All patients had at least 5 years of follow-up after surgery.¹² Only five patients (38%) had 20/80 or better best-corrected visual acuity (BCVA), and four patients (30%) had VA \leq 20/400. Contrast sensitivity (CS) and stereopsis were severely affected in all children.

The method of optical correction of aphakia is also reported to influence visual outcome. Retrospective observational studies have suggested that pseudophakia may result in better final VA in congenital cataracts depending on the timing of surgery. The Infant Aphakia Treatment Study (IATS) randomized infants 1 to 7 months of age with unilateral cataracts to IOL implantation versus aphakia and found no significant difference in grating acuity at 1 year.¹³ Five year follow-up results for infants in this study are being published in 2014.

The targeted refraction in pseudophakia may influence visual outcome.¹⁴ Lambert and colleagues¹⁵ reviewed 24 cases of children with unilateral pseudophakia and compared outcomes between those initially left undercorrected by 2 or more diopters and those close to emmetropia immediately after implantation (-1.00 to $+1.00$ D). They found no significant difference in final VA.

Associated ocular conditions or postoperative complications also influence visual outcome. In traumatic cases, it is often the accompanying ocular defects that limit the visual outcome. For example, corneal opacification or scarring, retinal detachment, traumatic maculopathy, or optic nerve injury may influence the visual result more than the cataract itself. Eyes with cataracts associated with juvenile idiopathic arthritis are known to have poor anatomical and visual outcomes. Eyes operated during early infancy are at a higher risk for the development of postoperative glaucoma, which contributes to the poor visual outcome.

Compliance with optical correction of aphakia and management of amblyopia ultimately becomes a determining factor. The earlier that the treatment of amblyopia is initiated, the more satisfying are the results. Chak and colleagues noted that in multivariate analysis, poor compliance with occlusion was the factor most strongly associated with poorer acuity in both unilateral and

bilateral disease: the odds of worse vision in unilateral cataract were eight times greater with 50% versus 100% compliance.⁶

POSTOPERATIVE STRABISMUS

See Chapter 48.

POSTOPERATIVE NYSTAGMUS

Birch and colleagues prospectively examined children who had undergone extraction of dense cataracts by the age of 5 years.¹⁶ Fixation stability and associated ocular oscillations were determined from eye movement recordings at \geq 5 years old. A total of 41 children were included. Of these, 29 (71%) developed nystagmus (18 predominantly infantile nystagmus syndrome [INS] and 11 predominantly fusion maldevelopment nystagmus syndrome [FMNS]). None of the children had “latent nystagmus vera,” that is, eyes electrophysiologically quiet under binocular conditions and oscillations present under monocular conditions. Both INS and FMNS were statistically significantly associated with strabismus. Significant risk for nystagmus was associated with infantile (\leq 12 months) onset (OR, 13.7; 95% CI, 1.6–302) and with visual deprivation $>$ 6-week duration (OR, 46.2; 95% CI, 6.0–1,004). Neither congenital onset nor laterality (unilateral versus bilateral) was associated with significant risk for nystagmus. Fourteen children (34%) had asymmetric fixation instability: nine had interocular differences in amplitude, four had FMNS when fixing with one eye but both FMNS and INS when fixing with the other eye, and one had INS only when fixing with one eye but not when fixing with the fellow eye. Significant risk for interocular difference in type or amplitude of nystagmus was associated with unilateral cataract (OR, 58.9; 95% CI, 5.1–2,318) and with congenital onset (OR, 25.0; 95% CI, 2.6–649). Hussin and Markham reported that nystagmus was recorded in 56% of the bilateral group and 23% of the unilateral group.⁷

BINOCULAR FUNCTION

Ing studied binocular function in bilateral pseudophakic children. The author visited four different medical centers and examined 21 patients. The mean patient age at surgery performed on the first eye was 6 years 4 months. The mean age at the date of the author’s examination was 16 years 5 months, and the mean length of follow-up was 10 years 4 months. All but two patients had motor alignment within 8 prism diopters of orthotropia at near. Fusion and some stereopsis were found to be present in 15 patients, but only four of these patients demonstrated fine (60 seconds of arc or better) stereoacuity. Patients with fine versus gross stereoacuity were compared and found to be similar in type of cataract, age at first surgery, interval between surgeries, and

length of follow-up and refraction but to differ in the quality of BCVA. The author concluded that although satisfactory motor alignment, fusion, and some stereopsis are present in the majority of patients, fine stereoacuity is uncommon in pseudophakic children.¹⁷

Kim and Plager¹⁸ reported stereopsis in children with unilateral pseudophakia. 21/38 patients had stereopsis better than 400 seconds of arc.

CONTRAST SENSITIVITY

CS testing has become an important clinical tool in the battery of tests used to characterize patients' vision. VA is only one measure of visual function, and it is correlated with how well an observer can detect and process high spatial frequencies. CS measures how much contrast is required to detect a particular spatial frequency. Vasavada et al. studied a vitrectomy and a no-vitrectomy group of children operated at 5 to 12 years of age.¹⁹ The authors concluded that although high-contrast VA was not significantly different between the groups, postoperative low CS was significantly better in vitrectomy group.¹⁹

VISUAL FIELD

Martin and colleagues reported that dense cataract causes a persistent impairment of spatial vision, both in the fovea and the visual field. However, the effect on the visual field is less pronounced than on VA.²⁰

QUALITY OF LIFE

See Chapter 53.

ORAL READING

Reading is a critical measure of the potential utility of the affected eye. Monocular oral reading ability and comprehension with eyes previously treated for dense congenital unilateral cataract, when measured with an abbreviated version of the Gray oral reading test, were worse compared with fellow eyes or normal control eyes.²¹

SUMMARY

Early surgery and compliance with patching are important factors to achieve good visual outcomes after cataract surgery in children. Unilateral cataracts, particularly when congenital, present the greatest challenge for visual rehabilitation. We await the long-term results of the IATS to evaluate the role of IOL implantation in early onset monocular cataract. Continued outcome analysis of older-onset unilateral cataract and bilateral cataract patients before and after surgery are encouraged.

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Amblyopia Management in the Pediatric Cataract Patient

Ronald G.W. Teed and David K. Wallace

INTRODUCTION

Amblyopia is often the principal barrier to a successful visual outcome in the management of pediatric cataracts. Leinfelder¹ described a condition of “congenital amblyopia” in children with congenital cataracts and poor visual acuity (VA) results following lensectomy. Indeed, it was once debatable if surgical management of unilateral congenital cataracts was advisable.² Advances in the understanding of the pathophysiology of amblyopia³ supported more aggressive therapy. Frey et al.⁴ suggested, in the 1970s, that treatment of monocular cataracts in children may be worthwhile. Over the past several decades, the understanding of deprivation amblyopia physiology and treatment has continued to evolve. This, coupled with ongoing refinement of surgical timing and technique, has resulted in even better VA outcomes in pediatric cataracts. Treatment of childhood cataracts is now commonplace worldwide, and surgical correction of even unilateral congenital cataracts is standard of care.⁵ This chapter reviews the visual outcomes in children with unilateral and bilateral cataracts, discusses the mechanisms and treatment of deprivation amblyopia caused by cataracts, considers the optimal timing of surgical treatment, suggests postoperative amblyopia therapy regimens, and explores the relationship between cataract-related amblyopia and binocular vision.

VISUAL ACUITY OUTCOMES IN CHILDHOOD CATARACTS

A review of VA outcomes will frame the approaches and expectations of amblyopia therapy. These data represent the full range of pediatric cataracts—unilateral and bilateral, congenital and acquired—and varied surgical and amblyopia management approaches. As expected, unilateral cataracts result in worse VA compared to bilateral cataracts, and age of surgery is strongly correlated with final vision.

In one of the largest reviews of childhood cataracts, Zwann et al.⁶ reported that 44% of 306 pseudophakic eyes achieved vision of 20/40 or better, with amblyopia as the major factor in acuity outcome. This result was similar to the mean acuity outcome of 20/55 in 100 pseudophakic eyes described by McClatchey et al.⁷ Another large study of 139 mixed childhood cataracts treated with intraocular lens (IOL) implantation had a median final VA of 20/30. Forty-five eyes in this study had a final VA of worse than 20/40; amblyopia was the cause of poor vision in 34.⁸

Within these large studies, we find poorer outcomes among cases of unilateral cataracts. Ledoux et al.⁸ reported a median final VA of 20/40 in these patients, compared to 20/25 in bilateral cases. Other series of unilateral cases confirm this finding and often find much worse final acuity. Hosal et al.⁹ reviewed 74 unilateral cataracts and found that only 37% had a final acuity of 20/40 or better. A series of unilateral traumatic cataracts in children had a mean final VA of 20/50, with amblyopia affecting 75% of children under 6 years of age.¹⁰ When only unilateral congenital cataracts are studied, the outcomes may be even worse.¹¹ In a review of 62 such patients, with a mean age of lensectomy of 5 weeks, only 32% had a final VA of 20/80 or better.¹² Early surgery was correlated with better final acuity.

It has been suggested that the method of optical correction (IOL implantation versus contact lens or spectacle correction) is an important factor in final VA. Some studies found no significant overall difference in VA between pseudophakia and aphakia,^{13,14} and others found that the difference depended on whether the child had unilateral or bilateral cataracts.¹⁵ Overall, though, these early studies did not study IOL implantation in children under 2 years of age. The Infant Aphakia Treatment Study (IATS) randomized infants 1 to 7 months of age with unilateral cataracts to IOL implantation versus aphakia and found no significant difference in grating acuity at 1 year.¹⁶ Follow-up is ongoing for infants in this study and

is planned until 5 years of age. Observational studies have suggested that pseudophakia may result in better final VA in congenital cataracts depending on the timing of surgery.^{14,17} This was also shown with traumatic cataracts.^{10,18}

The literature on final VA outcomes in pediatric cataracts presents a wide spectrum of pathology and results. The heterogeneous nature of these data makes conclusions difficult, but general trends are evident. Certainly, early surgery is preferred. The use of IOLs may not significantly improve final VA; however, pseudophakia may have other advantages to visual function, such as improved binocularity. Unilateral cataracts, particularly when congenital, present the greatest challenge for visual rehabilitation. The reasons behind these findings, and their practical implications, are the focus of the remainder of this chapter.

MECHANISMS OF DEPRIVATION AMBLYOPIA AND “THE CRITICAL PERIOD”

The relatively worse outcomes in unilateral cataracts, and those treated later in life, stem directly from the pathophysiology of deprivation amblyopia. Early visual experience is required for normal development of the visual cortex.³ Following a latent period during which vision is mediated via subcortical pathways,^{19,20} the visual system is highly dependent on robust visual input. Inducing monocular deprivation in animals (usually by suturing of eyelids) during this “sensitive period” causes a loss of normal cortical cell response to the deprived eye.³ Cells in the ocular dominance columns of the brain’s striate visual cortex that are normally attributed to the deprived eye switch their response to the functioning eye, resulting in corresponding shrinkage and growth of the columns.^{21–23} This cortical brain correlate of amblyopia can be partially reversed by preventing continued deprivation.²² Within this sensitive period, there is a “critical period” when early visual exposure is essential; during this time, absence of equal quality input to each eye can lead to irreversibly poor vision.

Management of amblyopia has been well described and is generally quite successful for strabismic and anisometropic amblyopia.^{24–27} Deprivation amblyopia, representing approximately 3% of all cases of amblyopia, is the most refractory to treatment.²⁸ The most common cause of deprivation amblyopia is unilateral congenital cataract, and bilateral congenital cataracts are a common cause as well.²⁹ Failure to promptly remove congenital cataracts, especially unilateral ones, can result in dense amblyopia refractory to management. The literature on VA outcomes consistently documents a universal age-related effect. Early surgery has the best chance of a good visual outcome, but what is early enough? Gregg and Parks³⁰ described a child with a unilateral congenital cataract who underwent lensectomy at 1 day of life and had a final acuity of 20/25 at age 8.

Further studies have attempted to determine the critical period during which cataract surgery should be conducted for congenital cataracts. Birch et al.³¹ found that very early surgery for unilateral congenital cataracts resulted in better VA. Subsequent analysis led to the conclusion that acuity outcomes drastically declined if surgery was conducted after 6 weeks.³² There was not a significant benefit of earlier surgery within these first 6 weeks (see Chapter 6).

Similarly, a critical period may exist for bilateral congenital cataracts. Lambert et al.³³ reviewed VA outcomes in 43 children and found that poor vision outcomes increased if surgery was conducted after 10 weeks of age. No eyes operated before 10 weeks of age had worse than 20/80 VA, though both groups (before and after 10 weeks) had an equal amount of children with 20/40 or better VA. Birch et al.³⁴ did not find a specific break point in 37 infants with bilateral congenital cataracts but did provide evidence that surgery before 14 weeks of age resulted in a better outcome. Surgery prior to 14 weeks did result in improved final outcome in a linear fashion. A smaller series of 13 children suggested that the decline was exponential, and no specific breakpoint was identified.³⁵ Given that bilateral deprivation causes less specific effects on the visual cortex,^{36,37} it is not surprising that a reproducible critical period was not identified in these infants. Overall, we can conclude that early surgery is also better in bilateral cataracts, though it may be delayed relative to unilateral cases.

Surgical correction of bilateral cataracts should occur in close succession, if the cataracts are symmetrical. A prolonged interval between cases could result in dense deprivation amblyopia in the untreated eye. Some have advocated for full-time occlusion of the treated eye until the second eye is operated on, though most surgeons attempt to remove the fellow cataract within 2 weeks.^{33–35} Limiting the delay between surgical treatment of bilateral cataracts is essential; indeed, immediate sequential same-day surgery has been advocated in select cases.

Asymmetrical bilateral cataracts that require surgical removal present a unique challenge, as deprivation amblyopia may be present in the worse eye. A novel approach to these cases has been suggested by Yu and Dahan³⁸: both eyes were operated simultaneously on the same day and only the worse eye received an IOL. The better eye was left aphakic; a secondary IOL was implanted once the amblyopic eye has improved. A similar approach has been described in bilaterally aphakic eyes: the contact lens is periodically removed from the better eye. Another approach is to operate on the worse eye and delay surgery on the fellow eye until VA is approximately equal.

Overall, a critical period likely exists for congenital cataracts. Unilateral cases should be operated on within 6 weeks to maximize VA; even earlier surgery may be beneficial for motor and sensory outcomes.^{39,40} Bilateral cases should be treated early in life as well, although the critical period likely extends to a few months of age.

PREDICTORS OF AMBLYOPIA SUCCESS

Because early visual experience is essential for a good visual outcome, congenital cataracts must be removed quickly. Acquired cataracts have a better outcome as patient age increases, since the visual experience is usually normal before acquiring the cataract.^{8,10} Similarly, better outcomes are correlated with a shorter duration from cataract onset to removal.¹⁰

The development of nystagmus predicts worse VA outcomes. This sensory-type nystagmus tends to develop by 13 weeks of age when congenital cataracts are left untreated,⁴¹ suggesting that a critical period of fixation stability exists.⁴⁰ Nystagmus therefore likely represents a surrogate for insufficient early visual exposure *and* has a direct effect on VA. Indeed, the presence of nystagmus was more strongly correlated with poor visual outcome than was late surgery in a series of 43 children with bilateral congenital cataracts.³³ Nonetheless, other series^{41,42} have reported good outcomes in these children. Rabiah et al.⁴¹ reviewed 95 cases of bilateral congenital cataracts presenting with sensory nystagmus at the time of their cataract surgery. They found 46% with 20/60 or better vision in the better eye. Interestingly, they reported that the nystagmus was eliminated or reduced in 40% of these children following treatment.

Another surrogate for amblyopia depth, specifically in unilateral cases, may be interocular axial length difference. It appears that deprivation amblyopia influences eye growth and leads to axial myopia in animals^{43,44} and humans.^{45,46} Thus, a longer axial length in a unilateral cataractous eye may imply a greater severity of amblyopia. Conversely, an eye with a *shorter* than normal axial length may also be at risk for a worse outcome, given associated ocular abnormalities found in microphthalmos. Gochbauer et al.⁴⁷ reviewed records of 64 children with unilateral cataract and found that deprivation amblyopia is likely severe when the interocular axial length difference exceeds 0.5 mm.

AMBLYOPIA AND OPERATIVE PLANNING

Once the decision is made to proceed with cataract surgery, the surgeon must decide on the type and power of optical correction. When choosing between IOL implantation and contact lens correction, there does not appear to be a significant difference in final VA between these two modalities, though pseudophakia may have advantages for binocular vision. This advantage may be greater for unilateral cases, as the high degree of anisometropia in aphakia would be quite amblyopiogenic if spectacles or a contact lens were not tolerated.⁴⁰ Bilaterally aphakic children tend to tolerate spectacle correction well.

The target postoperative refraction serves to maximize the child's visual needs and increase the likelihood of amblyopia treatment success. In congenital cataracts treated with lensectomy, most clinicians prescribe spectacle or contact lens correction to target a small amount of myopia, providing a near point of focus.⁴⁸ Correction is gradually updated over time, bifocals are initiated when they become useful to the child (e.g., age 2–4 years), and secondary IOL implantation is considered in the future.

The targeted refraction for pseudophakic correction is more challenging. As the child ages, there is a *somewhat* predictable myopic shift and axial lengthening^{7,49–51} that extends into the second decade of life.⁵² Thus, the targeted refraction is hypermetropic, and spectacle or contact lens correction is required postoperatively. The degree of hypermetropia depends on the age of the patient: an undercorrection of as much as +8.00 diopters (D) is considered in infants under 6 weeks of age,¹⁶ and the targeted hypermetropic refraction decreases with age. Even when the postoperative refraction is appropriately targeted, there remains a substantial prediction error in children.^{53,54} Moore et al.⁵⁴ found a mean prediction error of 1.08 diopters in a group of 203 consecutive eyes.

The targeted refraction in pseudophakia may influence the success of amblyopia therapy. In general, the child is prescribed spectacles to correct the residual hypermetropia, either to emmetropia or an overcorrection to induce myopia and allow for a near point of focus. In unilateral cases, though, poor compliance with spectacles would result in anisometropia and amplify amblyopia. Therefore, some clinicians have suggested that targeting the refraction to emmetropia or low hypermetropia is a better strategy. This approach potentially trades improved amblyopia therapy and better final visual outcome for more myopia in the future. Lowery et al.⁵⁵ suggested that a low hypermetropic target (+1.75 to +5.00 D) resulted in a better final VA outcome in children with unilateral pseudophakia. Interestingly, the children with a small initial postoperative refractive error (–1.00 to +1.00 D) did not have a better VA outcome, though the heterogeneous nature of the group and late age of operation make conclusions difficult. Lambert et al.⁵⁶ reviewed 24 cases of children with unilateral pseudophakia and compared outcomes between those initially left undercorrected by 2 or more diopters and those close to emmetropia (–1.00 to +1.00 D). They found no significant difference in final VA or rate of myopic shift. Perhaps the initial target refraction matters less than compliance with occlusion therapy and spectacle wear.

AMBLYOPIA THERAPY REGIMENS

Regardless of the postoperative refraction, amblyopia therapy remains critical in the rehabilitation of these eyes. Amblyopia is the principle cause of vision loss in

unilateral cataracts, and all cases require some degree of treatment. Unlike strabismic and anisometric amblyopia,²⁴⁻²⁷ there are no evidence-based guidelines for the treatment of deprivation amblyopia.²⁹ Occlusion of the sound eye remains the mainstay of amblyopia therapy, though optical blur has been used in select cases.³⁸ Atropine penalization is unlikely to be effective in deprivation amblyopia in pseudophakic eyes. Deprivation amblyopia from unilateral congenital cataract is likely the most refractory to treatment; it has therefore been assumed that substantial amounts of occlusion are required for a good visual outcome. Some studies suggested intensive occlusion of 6 to 8^{31,40} or 8 to 10⁹ hours per day, gradually reducing daily amounts as the child ages. Lloyd et al.⁵⁷ started with full-time occlusion in the 1st week following lensectomy and then changed the duration of occlusion to between 50% and 100% of waking hours based on the interocular VA difference as determined by a preferential looking test.

An alternate approach was developed that gradually increased the amount of occlusion over time, as some evidence suggests that binocular visual outcomes are improved with early binocular experiences.⁵⁸ Several series supported this “progressive occlusion” regimen that started with 1 hour/day of occlusion per month of age, increasing to 50% of waking hours by 4 to 8 months of age.^{12,30,58,59} Jeffrey et al.⁵⁸ reviewed several studies of progressive occlusion and found that 26 of 35 total patients had a final VA of 20/80 or greater. The IATS has used a progressive occlusion regimen of 1 hour/day/month until 8 months of age and then 50% of waking hours (either full time every other day or half time daily).¹⁶

It is assumed that occlusion therapy must continue throughout childhood,¹² although no data exist on the rates of amblyopia recurrence in these patients. Once the amblyopia therapy has reached an end point, maintenance treatment of 1 to 4 hours/day has been suggested.⁹ Alternatively, amblyopia therapy may be stopped once VA has reached a plateau and restarted only if acuity regresses.

CATARACT-RELATED AMBLYOPIA AND BINOCULARITY

Poor binocular vision outcomes are common in children treated for cataracts, even when VA is good in both eyes. Strabismus is common in these patients, with most large series reporting over 50%.^{8,9,13,15,31,49,58} Sensory fusion and stereopsis are similarly impaired in these patients, even in the absence of strabismus.^{9,58} Unilateral congenital cataracts, whether treated with IOL implantation or contact lens correction, frequently demonstrate higher rates of sensorimotor defects.

Impaired binocularity in unilateral cataracts results from abnormal input to the binocular cells outside of layer 4 of the visual cortex. With deprivation, these cells begin to respond only to the sound eye. With rehabilitation, the

amblyopic eye begins to reestablish its ocular dominance column and VA improves; however, the binocular cells fail to regain bilateral responsiveness.²² Normal binocularity is thus disrupted by the original deprivation. In addition, anisometropia and/or strabismus, which are common in these patients, will continue to facilitate poor binocularity. It has been proposed that binocular vision may have an even shorter critical period than does monocular acuity.^{39,40}

Nonetheless, there are scattered reports of good binocular outcomes in children with cataracts. Greenwald and Glaser¹³ reported that 58% of children with unilateral cataracts had at least 400 seconds of arc stereopsis. In a similar group, Hosal et al.⁹ found that 11 of 74 patients (15%) had at least 100 seconds of arc stereopsis. Unilateral congenital cataracts tend to have worse outcomes; in the series by Hosal et al.,⁹ only 2 of 19 (11%) demonstrated at least 100 seconds of arc, though one patient demonstrated 40 seconds of arc. Additional series have reported mostly poor levels of stereopsis in these patients, though scattered cases of good, and even excellent, binocularity can be found.^{15,30,31,59}

Pseudophakia may provide an advantage to binocularity in unilateral cataracts. Greenwald and Glaser¹³ reported gross stereopsis in 90% of 20 children with pseudophakia, compared to only 39% of 31 aphakes. Yamamoto et al.¹⁵ found at least gross stereopsis in 63% of 8 patients treated with an IOL for unilateral congenital cataract, compared to 9% of 34 treated with contact lens correction. Similarly, the incidence of strabismus appears to be less in IOL-managed infants.¹⁴ In the IATS, the incidence of strabismus at 1-year follow-up was lower for the IOL group (42% compared to 62%; $P = 0.5$).¹⁶

Early binocular experience may also improve ultimate binocular outcomes. Several of the early reports of good sensory results followed a progressive occlusion regimen.^{30,59,60} Similarly, the incidence of strabismus appears to be reduced when a progressive occlusion regimen is used.^{58,61} It has been proposed that allowing for more binocular exposure early, even with an amblyopic eye, may preserve binocular cell responsiveness. Jeffrey et al.⁵⁸ compared 29 cases of unilateral congenital cataract that underwent intensive amblyopia therapy to eight cases treated with a progressive regimen. Fifty percent of the progressive occlusion group demonstrated stereoacuity or fusion, compared to only 14% of the intensive group. Strabismus was less common in the progressive group (63%–90%). This support for a progressive regimen was tempered by the significant difference in age of cataract surgery (20 days in the progressive group, compared to 33 days in the intensive group).

Overall, good binocular outcomes in pediatric cataracts remain uncommon, particularly for unilateral congenital cases. Pseudophakia and a progressive occlusion regimen may maximize outcomes; however, early surgery and diligent amblyopia compliance likely play major roles in success.

AMBLYOPIA THERAPY COMPLIANCE

Adherence to amblyopia therapy remains challenging, even when the level of visual impairment is only moderate.^{62,63} Recommended occlusion regimens for deprivation amblyopia are much more stringent, and these children likely require maintenance occlusion throughout childhood. Unilateral congenital cataracts likely require the most therapeutic compliance to achieve a good visual outcome and are likely the most difficult to maintain. Allen et al.¹² reported that 31% of unilateral congenital cases had abandoned therapy by age 4. Lloyd et al.⁵⁷ found that percentage compliance (actual occlusion divided by target occlusion) was over 79% in 9 of 10 cases: a patching diary was kept to record and encourage adherence. In the IATS, over 70% of caregivers in both the IOL group and the contact lens group reported good compliance with therapy (at least 75% of prescribed hours) at 2 months following surgery.¹⁶ This gradually decreased to 50% to 60% by 6 months, and there was a trend toward better compliance in the aphakia group.

If compliance among participants in the highly controlled setting of a clinical trial is suboptimal, then it is likely even worse in routine clinical practice. Studies of other types of amblyopia measured compliance and found concordance with patching between 30% and 80%, with better compliance in the first few weeks of therapy.^{62,64} Compliance may be more challenging in amblyopia after cataracts because of worse VA and an inability to accommodate. Collaboration between the ophthalmic team and the caregivers must occur early in the course of management and realistic expectations of occlusion therapy and compliance must be agreed upon. The caregivers should be counseled on the primary importance of amblyopia management, perhaps even before surgery. One approach is to counsel parents that “surgery removes the cataract, but it is the patching that will treat the amblyopia and allow the eye to see.”

PHAKIC AMBLYOPIA

This chapter has focused on the development and management of amblyopia associated with cataracts that required surgical removal. We now briefly discuss the management of small cataracts that are not severe enough to warrant removal.

As a general rule, a lenticular opacity of <3 mm is observed, though the type and location of the cataract influence the amblyopiogenic risk. Occlusion therapy in small cataracts can improve vision.⁶⁵ Ceyhan et al.⁶⁶ reviewed 59 patients with congenital anterior lens opacities and reported that 29% were diagnosed with amblyopia. Interestingly, the type of cataract (anterior subcapsular, polar, or pyramidal) and size of opacity were not associated with a diagnosis of amblyopia. Anisometropia was associated with amblyopia in this series. Wheeler et al.⁶⁷ reported amblyopia in 14 of 15 patients (93%)

with an anterior pyramidal cataract. Travi et al.⁶⁸ reported amblyopia in 48 of 53 children (91%) with posterior lens changes (posterior lenticonus, posterior subcapsular cataract, and anterior persistent fetal vasculature [PFV]).

Amblyopia treatment is successful in many of these cases. Ceyhan et al.⁶⁶ reported that 12 of the 17 children diagnosed with amblyopia had a successful result, though final visual acuities were not reported. Travi et al.⁶⁸ reported that 67% of patients diagnosed with amblyopia and with recordable VA improved by 0.3 logMAR units or attained 20/20 vision in the affected eye. A poor visual outcome was more likely among the children with posterior lenticonus; anisometropia did not increase the risk of a poor outcome. Of note, six children in this series progressed to cataract surgery.

The mechanisms and management of small cataract-related amblyopia remain unclear. It is likely that both deprivation and anisometropia play a role in the development of amblyopia, depending on the location and type of opacity. No specific guidelines exist for amblyopia therapy: these opacities have been variably treated with part-time occlusion, full-time occlusion, and atropine penalization.⁶⁸

Perhaps most challenging is the decision to stop amblyopia therapy and recommend cataract surgery. Failure of amblyopia therapy and increasing opacity size may encourage cataract removal, though it is difficult to predict if VA will improve after surgery. Factors suggesting that cataract removal may be warranted include poor low contrast sensitivity vision, glare symptoms, and axial elongation; however, no set guidelines exist for these patients.

SUMMARY

Visual outcomes in pediatric cataract surgery are strongly influenced by the success of therapy for deprivation amblyopia. A critical period exists for congenital cataracts, and unilateral congenital cataracts should be removed by 6 weeks of age. As a general rule, earlier surgery results in better visual outcomes, though very early surgery may increase the risk of other complications including glaucoma and uveitis.⁶⁹ Pseudophakia may improve binocular visual outcomes in congenital cataracts, though this approach involves more challenging refractive management and may increase the risk of intraoperative and postoperative complications.⁷⁰ In visually significant acquired pediatric cataracts, rapid removal and rehabilitation is recommended, especially for younger children. Whether the cataract is congenital or acquired, corrected with pseudophakia or aphakia, and treated with intensive or progressive occlusion, the greatest challenge for many families is amblyopia treatment compliance.

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Strabismus in Pediatric Aphakia and Pseudophakia

M. Edward Wilson and Rupal H. Trivedi

Children with cataracts in one or both eyes may experience enough disruption in binocular sensory and motor fusion mechanisms to lose proper ocular alignment. The frequent association of strabismus and cataracts in children is well known.¹⁻¹² It appears to present in all age groups and at a greater incidence than in the general population. Strabismus is common when deprivation amblyopia is present but may occur even in the absence of amblyopia, as in dense bilateral lens opacities. Furthermore, strabismus can also have its onset after the cataracts have been removed, especially if the proper optical correction is not worn or if occlusion therapy leaves little time for binocular viewing. Earlier-onset unilateral cataracts have the greatest risk for strabismus, and late-onset bilateral cataracts have the least risk. Also, as a general rule, patients with partial cataracts and relatively good preoperative visual acuity have less strabismus. Long delays in detection and treatment of visually significant cataracts increase the risk of strabismus, even when the cataracts are incomplete. Strabismus at presentation is often an indication that the cataract is long-standing and that significant amblyopia is likely to be present. In review of the literature, strabismus was present in 33.3% of patients preoperatively and in 78.1% of patients postoperatively.⁶ The higher incidence of strabismus after treatment than before may reflect the intensity of the occlusion prescribed, the ongoing susceptibility of the infantile eye to amblyopia owing to an uncorrected refractive error, or the easier detection of strabismus in older children.

Below we review literature on strabismus associated with cataract in children. Comparing the literature, we remind the reader that it is difficult to compare studies because of the variations among the inclusion criteria, exclusion criteria, prescription of occlusion, and compliance to occlusion; however, certain general conclusions can be drawn.

Parks and Hiles¹ had found a strong correlation between cataract type and risk for strabismus. Strabismus

was found in 100% of persistent fetal vasculature cataracts and 65.5% of nuclear cataract patients but in only 48.4% with posterior lentiglobus and 21.8% in the cortical lamellar group ($P = 0.0001$). Ocular misalignment was found in 66.7% of patients with unilateral cataracts but only 37.9% of patients with bilateral cataracts ($P = 0.0018$). France and Frank³ found that strabismus was present in 40% of their patients with cataracts or dislocated lenses at the time of initial diagnosis. At last follow-up, 86% of the congenital cataract patients and 61% of the acquired cataract patients had strabismus. Unilateral congenital cataract patients had esotropia or exotropia at about equal proportions. However, bilateral congenital cataract patients, when strabismic, were all esotropic. The acquired cataract patients had mostly exotropia. In 2003, we reported strabismus in 50% of our patients preoperatively, while it occurred in 45% of patients after cataract surgery in eyes with monocular cataract.⁶

Use of intraocular lens (IOL) implantation and the trend toward earlier surgery for partial or complete cataracts in children had brought with it the hope for a decrease in the incidence of strabismus. BenEzra and Cohen⁴ noted that only 9% of children with unilateral pseudophakia developed strabismus compared to 71% of children with unilateral aphakia treated with contact lenses (CLs). The reduced incidence of strabismus in children with unilateral pseudophakia compared to unilateral aphakia is probably because of the constancy of the optical correction and the improved visual outcome in pseudophakic children. Lambert et al.⁵ reported that 9 of the 12 patients in the IOL group had strabismus (4 esotropia, 4 exotropia, and 1 hypertropia) compared with 12 of 13 in the CL group (5 esotropia, 5 exotropia, and 2 hypertropia; $P = 0.24$). Results of the Infant Aphakia Treatment Study found that approximately two-thirds of the patients in both the CL and IOL groups were orthotropic at the 1-month examination. However,

at the 12-month examination, there was a trend for more of the patients in the IOL group (58%) to be orthotropic compared with patients in the CL group (38%; $P = 0.05$).⁹

Spanou et al.¹¹ reported that strabismus was present in 199/842 (24.2%) of children. Esotropia was more common in congenital cataract, while exotropia was more common in acquired cataract.¹¹ Birch et al. evaluated children who had undergone extraction of dense cataracts by the age of 5 years. Sixty-six percent of children developed strabismus; esotropia was most common. Visual deprivation >6-week duration was associated with significant risk of strabismus.¹²

Strabismus surgery is often delayed until after the initial amblyopia treatment has improved the visual acuity somewhat. Occasionally, an intermittent strabismus will resolve as the visual acuity improves. However, this is the exception rather than the rule. Strabismus associated with cataracts in children usually requires surgery. At times, strabismus surgery is performed prior to cataract surgery. However, it has been our experience that stability of ocular alignment is not often achieved when strabismus surgery precedes cataract surgery. The preferred route is to rehabilitate the optical system of the eye or eyes and strive for the best possible vision prior to strabismus correction. That said, strabismus correction can still be done during, rather than at the completion of, amblyopia treatment. Normally, the strabismus is measured several times during the postoperative follow-up after cataract surgery. If the strabismus angle and pattern are stable, correction is planned at an elective time such as during a scheduled examination under anesthesia where intraocular pressure, retinal examination, and serial axial length measurements are planned.

Stability of alignment after strabismus surgery in aphakic or pseudophakic children is difficult to predict. Generally, children with better visual outcomes and better

binocularity have better stability of ocular alignment. Dense deprivation amblyopia lessens the chance of long-term correction of strabismus, but despite this, a cosmetically acceptable alignment is often maintained for many years. Overall, the success of strabismus surgery is higher for strabismus that develops after cataract surgery compared to strabismus that is present preoperatively, at initial presentation. Although occlusion therapy for amblyopia must usually be continued after surgical realignment of the eyes, full-time occlusion is not recommended since it carries a high risk for recurrence of strabismus.

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Intraoperative and Postoperative Complications

Rupal H. Trivedi and M. Edward Wilson

Cataract surgery complications in children are higher as compared to adults. Intraoperative complications are higher as a result of the smaller size of the eye, sometimes poorly dilating pupil, highly elastic anterior capsule, low scleral rigidity, and increased intralenticular/intravitreal “pressure.” Postoperative complications are higher due to an increased inflammatory response. The rate of intraoperative complications is inversely proportion to the age at surgery. In addition, failure to achieve pupil dilation is an important risk factor. Detection and diagnosis of the complications can be challenging because of the difficulties encountered in performing detailed postoperative examinations in children, who may be uncooperative.

INTRAOPERATIVE COMPLICATIONS

Closed chamber surgical techniques using tunnel incisions and bimanual irrigation/aspiration as well as the use of high-viscosity ophthalmic viscosurgical devices (OVDs) have helped us to minimize iris touch and improve intraoperative performance. Primary intraocular lens (IOL) implantation may increase the risk of intraoperative complications. Results of the Infant Aphakia Treatment Study (IATS) reported that intraoperative complications were observed in 11% (6/57) of the eyes of the *no-IOL group* versus 28% (16/57) of the eyes of the *IOL group*.¹

Iris and Pupil

Iris Prolapse

Iris prolapse is more frequently observed during pediatric cataract surgery as compared to adult (Fig. 49.1A and B). This results from the less rigid, softer corneal tissue and the very floppy iris tissue of the infant or young child. Iris may come into even a well-constructed corneal tunnel or even a paracentesis with an adequate internal lip. Overfilling the anterior segment with OVD may contribute to the iris prolapse. Iris entering the internal opening of a well-constructed wound is more common in infancy and

almost never occurs in older children. Care must be taken to not traumatize the iris when this occurs as instruments enter and leave the eye. Incisions should be constructed to provide a snug fit for the instruments that pass into the anterior chamber (AC). Smaller wounds and tunnel configurations decrease the incidence of iris prolapse during surgery. Iris prolapse during surgery was observed in 4% (2/57) of the eyes of the *no-IOL group* and 21% (12/57) of the eyes of the *IOL group* ($P = 0.008$) in IATS.¹ The authors attributed this fivefold increase in the IOL group to a larger wound size and the greater intraocular manipulation required to implant an IOL. Iris damage and postoperative iris transillumination defects may be observed in eyes with iris prolapse, especially in blue-eyed babies.

Intraoperative Miotic Pupil

A pupil may fail to dilate preoperatively due to anterior segment dysgenesis/iris hypoplasia, or a well-dilated pupil may constrict due to surgical manipulation/iris touch. Use of 0.5 mL of 1:1,000 adrenaline (epinephrine) in 500 mL of irrigating fluid is essential to maintain pupillary dilation throughout the surgery. Use of OVD can help to improve or maintain pupil dilation as well.

Since the management of pediatric surgery in the presence of a small pupil (Fig. 49.2) is not discussed elsewhere in this book, we include here a brief summary of options available to remove a cataract in the presence of a small pupil (as cited in Ref.²). The solution may be as simple as manipulating the pupil edge using a second instrument such as an aspiration handpiece or an iris repositor or iris hook. As each quadrant is operated, the second instrument is used to expose that quadrant by holding back the iris and working under it. Iris hooks (1–5) can also be placed through small self-sealing corneal openings to pull back the iris in all quadrants or just in those where the second instrument method does not work. These are easy to insert and can be selectively tightened or loosened to allow visualization of the peripheral lens material

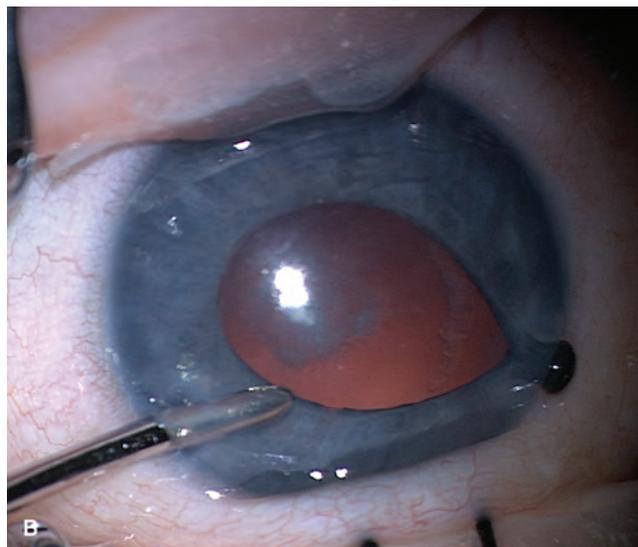
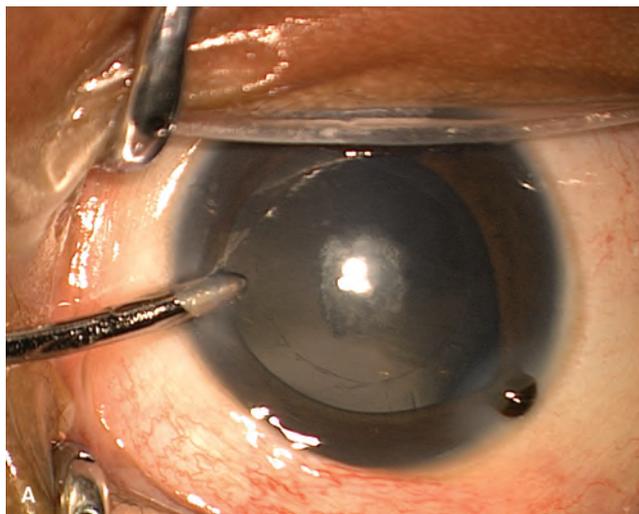


Figure 49.1 A and B: Iris prolapse during cataract surgery.

while not overly traumatizing the iris. Iris hooks come in a packet of five disposable and adjustable hooks. The intent of the manufacturers is for the surgeon to use four to hold the pupil open in a square shape. There is one additional hook included in the package as an extra. Some surgeons use all five and create the shape of a “home-plate” in baseball. Even in babies, these wounds do not need suture closer when made properly. It is our recommendation that surgeons consider using these iris hooks more often when pupils are small. The extra time to place the hooks is well worth it to insure that all peripheral lens cortex is visualized and removed completely.

Pupil expander rings represent another option in the surgical armamentarium for small pupil cases. The Hydroview Iris Protector Ring (Grieshaber) forms a compressed oval in its dehydrated state. It can then be placed in the AC and inserted into the small pupil. This hydrogel

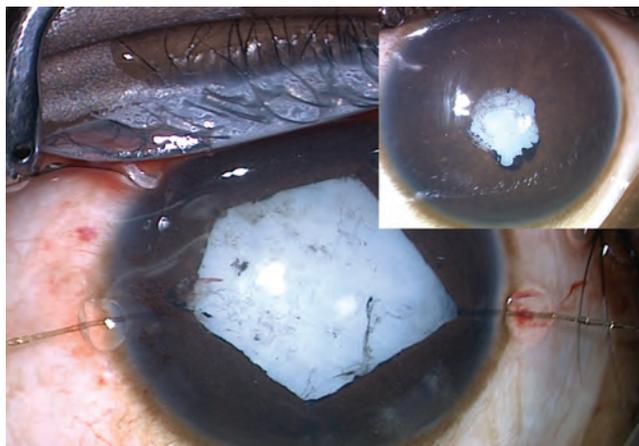


Figure 49.2. Use of iris hook in a 12-year-old child operated for cataract with small pupil. **Inset** shows preoperative view.

device expands with hydration and captures the pupillary margin by means of flanges. The ring can be manipulated to expand the pupil as it hydrates. The device can then be removed at the end of surgery through the same small incision. The Morcher Pupil Expander Ring Type 5S is a solid polymethylmethacrylate (PMMA) ring that is placed at the pupillary margin and expands the pupil. It reduces the likelihood of iris sphincter tears and postoperative pupillary deformity. Following implantation of the IOL, the ring is removed. An injection device is also available for the ring from Geuder. The Perfect Pupil (Becton, Dickinson) represents another effective option for both maintaining mydriasis and protecting the pupillary margin during surgery. This polyurethane device features a 7-mm internal diameter and an available injection device. Another helpful and easy-to-use expansion ring is the Malyugin Ring (MicroSurgical Technology [MST]). Its use has also been reported for pediatric cataract surgery.³ This ring is supplied with a disposable injector that compresses the ring to allow its insertion and then its controlled expansion within the eye. The smaller version of the Malyugin Ring is recommended when operating on children. The full 7-mm size can be overly traumatic for a small soft eye. Alternatively, the Beehler pupil dilator (Moria #19009) stretches the pupil to 6 to 7 mm while usually creating tiny microsphincterotomies circumferentially around the pupil. The pupil can then be mechanically reduced at the end of the procedure with an intraocular miotic agent.

Lastly, some miotic pupils in children are fibrotic and return to their sub-2-mm size after stretching. Reoperations for these miotic and nonreactive pupils require larger radial sphincterotomies or a pupilloplasty using the vitrector handpiece to remove a portion of the fibrotic pupillary edge. Care must be taken to not remove too much iris. If done properly, the cosmetic appearance of

the pupil can be surprisingly good although reactivity to light is still absent.

Pediatric Intraoperative Floppy Iris Syndrome

The intraoperative floppy iris syndrome (IFIS) includes a triad of intraoperative signs: *iris billowing and floppiness, iris prolapse, and progressive miosis*. We reported pediatric IFIS in a 4-month-old child with bilateral cataract.⁴ Surgery in the right eye was uneventful (Fig. 49.3A). During removal of the cataract in the left eye, signs of IFIS were observed (Fig. 49.3B). The surgical technique was identical in both eyes except that epinephrine was added to the irrigating solution in the right eye but inadvertently omitted in the left eye surgery. Our case highlighted the importance of epinephrine in pediatric cataract surgery. Of note, pediatric IFIS was also reported in the case of a 1-month-old girl with bilateral persistent pupillary membranes without cataracts.⁵ It is our contention that many, if not all, infants have a floppy iris and that preoperative dilation with strong mydriatics and epinephrine placed in the irrigation solutions are necessary steps to reduce the effects of the naturally floppy iris. Without the intraocular epinephrine, iris prolapse into even the smallest wound would be even more common with infant surgery.

Collapse of Anterior Chamber

Fluctuation of AC depth occurs frequently in soft pediatric eyes. Techniques mentioned to avoid iris prolapse (tight fit wounds) also help to maintain a stable and deep AC. Bimanual irrigation/aspiration approaches help to avoid AC fluctuation. Some surgeons prefer to use an AC maintainer rather than holding both instruments as in bimanual surgery. An unstable AC usually results from too much leak around the instruments that are placed in the AC. Venturi pump machines often have a fluid pump that can be increased to offset the leak. Gravity-fed irrigation systems are not as effective. The surgeon must also learn to balance the aspiration with the irrigation in a way that does not repeatedly shallow the AC during surgery. Repeated shallowing or flattening and then deepening of the chamber during surgery acts like extensive iris manipulation. It releases inflammatory mediators and results in a marked increase in postoperative inflammation as well as intraoperative miosis.

Anterior Capsule Tear

It is no longer a secret that anterior capsulectomy is notoriously difficult in infants and young children because of the extreme elasticity of the anterior capsule, scleral collapse causing positive vitreous pressure, and, at times, poor dilatation of the pupil. Rarely, an anterior capsule tear may be observed as soon as the AC is opened, caused by the sharp tip of the entrance knife, especially when the AC is very shallow. The availability of better operating microscopes

and microsurgical instruments combined with higher-viscosity OVDs are helpful; however, “runaway rhexis” or tear extending out toward the equator is still frequently encountered (Fig. 49.4). For manual continuous curvilinear capsulorhexis (CCC), it occurs mainly during anterior capsulectomy. Once a manual CCC is achieved, the edge is very strong and will withstand intraocular gymnastics very efficiently. When a tear occurs during manual CCC, the surgeon should stop tearing, place more OVD, regrasp close to the tear edge, and pull toward the center of the pupil. If this does not recover the capsulectomy easily, conversion to a vitrectorhexis or to a Kloti diathermy capsulectomy has been successful for us. For vitrectorhexis and Kloti diathermy, a tear is more likely to occur during IOL implantation or irrigation/aspiration or after OVD removal. Care should be taken to avoid right-angled edges, which is a weak point and likely to tear during intraoperative maneuvering. If a right-angled edge is seen during the capsulectomy, it should be rounded out using the vitrector before completion of the capsulectomy. The sudden flat AC that can occur after the OVD is removed before the wounds are sutured can cause the IOL to move anteriorly and place stress on the capsulectomy edge. Every effort should be made to sew the wounds prior to complete removal of the OVD if possible. A temporary air bubble can be used at the end of the procedure as another way to avoid shallowing of the chamber as the wound suture is being tied. Chapter 17 describes studies reporting the intraoperative tear rate using different techniques and other specific issues related to anterior capsule management.

Inappropriate size and shape are also complications of the anterior capsulectomy that are seen more often with children than with adults. Surgeons are urged to go slow and pay particular attention to size, shape, and centration each time the capsulectomy edge is released. If the capsulectomy opening is too small, it can be enlarged after the IOL is inserted. If it is too large or poorly shaped, take care to get the haptics of the IOL under the capsulorhexis edge and place the haptics where the capsulectomy edges can be most easily seen. Every attempt should be made to cover the edge of the IOL optic with the capsulorhexis edge when possible.

Positive Vitreous Pressure

A high intraoperative vitreous pressure is produced as a result of scleral collapse due to low scleral rigidity, which results in forward movements of the iris–lens diaphragm. In pediatric eyes, the posterior capsule is often convex rather than concave from the surgeon’s viewpoint. We often remove the peripheral lens cortex first, leaving the central nucleus of the lens until last since it holds the posterior capsule back in a more flat or concave configuration. Use of a high-viscosity OVD (e.g., Healon GV) helps to maintain a deep AC and keep the iris–lens diaphragm back.

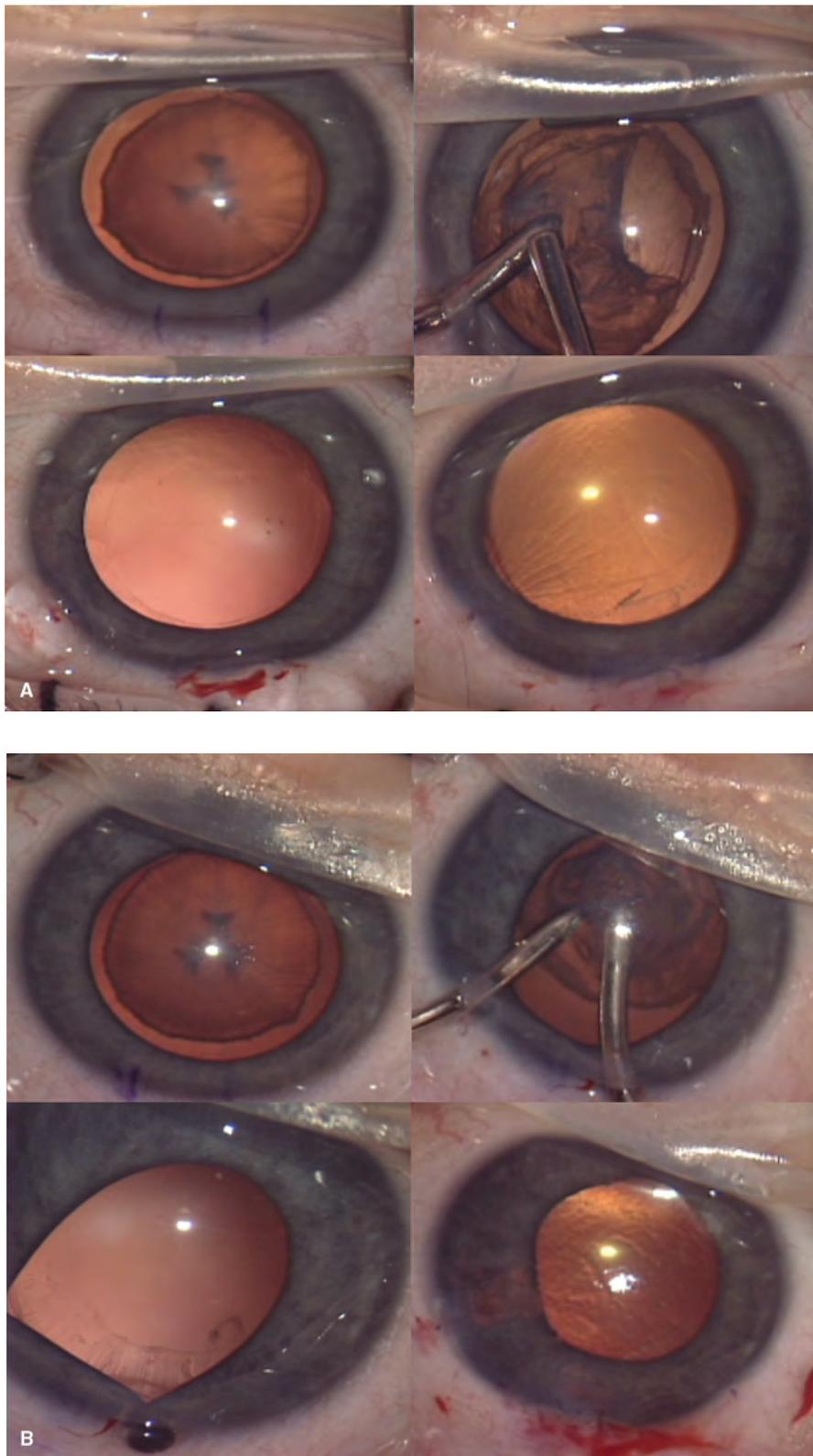


Figure 49.3 A and B: Pediatric intraoperative floppy iris syndrome.

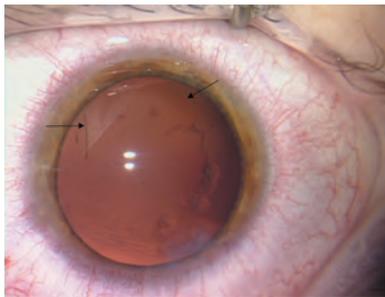


Figure 49.4. Photograph illustrating peripheral extension of the anterior capsule tear (“runaway rhexis” shown by arrows) during the vitrectorhexis procedure. This is not uncommon during pediatric cataract surgery owing to the elasticity of the lens capsule.

Posterior Capsule Tear

A posterior capsule tear is not as devastating to a pediatric cataract surgeon as it is to an adult cataract surgeon, because pediatric cataract removal often includes posterior capsulectomy and vitrectomy. However, an uncontrolled posterior capsule tear may still compromise the ability of the surgeon to safely place an IOL into the capsule. Posterior capsule tears may occur due to preexisting posterior capsule defects (Fig. 49.5). The surgeon can recognize a *torn posterior capsule* during lens aspiration by signs such as a sudden deepening of the AC. This occurs instantaneously as a rent appears in the capsule. As this occurs, the pupil will dilate in response to the deepening AC. Finally, during aspiration, lenticular particles/residual cortical matter falls away from, and will not come toward, the tip of the aspirator. This occurs because the tear in the posterior capsule alters the flow dynamics in the AC. Sometimes the presence of vitreous in the AC may also indicate a torn posterior capsule. The posterior capsule can tear during hydrodissection, irrigation and aspiration, capsular polishing, lens insertion, and OVD

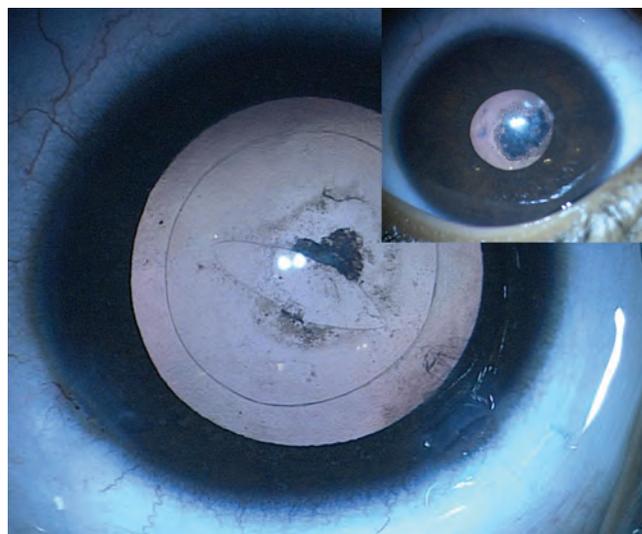


Figure 49.5. Posterior capsule defect.

removal. Posterior capsule tear can also occur during incision enlargement with the sharp tip of the keratome.

In all cases, if the posterior capsular tear is entirely within view, a posterior capsulorhexis can be attempted. If the vitreous face is intact, this is done by placing OVD above and below the tear, pushing the vitreous face back, and allowing room to grasp the torn capsule. If the vitreous face is already broken, OVD is necessary only to stabilize the AC and make room for the forceps. Using a capsulorhexis forceps, the capsule is then gently torn to create a 360-degree posterior capsulorhexis. Failed attempts may result in extension of the tear and, if not already present, vitreous loss.

Alternatively, the vitrector handpiece can be used to round out the posterior capsule tear, remove residual cortex, and remove prolapsed vitreous. Pediatric surgeons are more likely to use this approach since the use of the vitrector is often more familiar than the use of a capsulorhexis forceps on the posterior capsule. Care should be taken to begin with a low flow so that the AC dynamics are not changed drastically when the instruments are placed in the eye. This could lead to an extension of the posterior capsule tear. Once rounded out, the posterior vitrectorhexis can be quite stable and resistant to further tearing. The advantage of the vitrector handpiece is that it can safely remove cortex when it is mixed with vitreous. It can also cut the capsule and vitreous simultaneously without undue traction of the vitreous base or retina. A Venturi pump–driven vitrector handpiece works best when used in this way.

Anterior Vitreous Face Disturbance

Anterior vitreous face (AVF) disturbance during posterior capsule rupture is well recognized in adult cataract surgery. However, in children, this is less of a problem because anterior vitrectomy has become an integral part of the surgical strategy. However, some surgeons prefer to err on the conservative side and avoid anterior vitrectomy in children 2 to 8 years of age. When performing a posterior continuous curvilinear capsulorhexis (PCCC) without vitrectomy, surgeons should watch for signs for AVF disturbance, and if seen, a vitrectomy should be performed. The subtle signs are a vitreous strand in the AC, vitreous strands attached to the capsule flap, and distortion of the anterior and posterior capsulorhexis. The latter is considered a pathognomonic sign of AVF disturbance.⁶ Signs of an intact AVF include its bulging structure and its characteristic triamcinolone staining patterns recently documented by Sudhalkar et al.⁷ These include the homogenous staining pattern of three buttonholes, giving the impression of a tabletop configuration with three subramifications.

Residual Vitreous in the Anterior Chamber

Vitreous strands can remain in the AC. When this occurs, it should be removed using vitrector. A second instrument may be needed, through a paracentesis, to sweep the

vitreous strand away from the wound so that the vitrector handpiece can remove it. A taut strand of vitreous will not easily enter the vitreous cutter until its connection to the wound is broken. A pars plana approach to the anterior vitrectomy avoids this possible complication. Some surgeons use triamcinolone to identify any residual vitreous, while some choose to use miotics to look for peaking of the pupil.

HypHEMA and Intraoperative Vitreous Hemorrhage

Intraoperative hypHEMA may occur and may or may not need to be cleared before wound closure.⁸ A small hypHEMA will clear spontaneously. HypHEMA was noted in approximately 5% of eyes in the IATS.¹ Vascularized plaques (Fig. 49.6) or patent hyaloid artery remnants may lead to intraoperative vitreous hemorrhage.

Complications of Pars Plicata Vitrectomy

Laceration of the equator of the capsular bag with the microvitrectomy (MVR) blade is a possible complication of a pars plana incision. The entry with the MVR blade should be aimed toward the center of the vitreous cavity to avoid hitting the capsular bag. The capsule may still be distended with an OVD or irrigation fluid. Surgeons like to see the tip of the MVR blade in the pupillary space through the operating microscope before withdrawing it. The tendency is to advance the MVR blade too anteriorly. In very young infants, the entry is even closer to the limbus. In these cases, the MVR blade must be aimed at the optic nerve to avoid hitting the capsular bag. Another less commonly encountered complication is bleeding into the vitreous cavity. The vitrector cutter must be turned off before the handpiece is withdrawn. If not, the cutter may engage the tip on a ciliary process during handpiece withdrawal, causing profuse bleeding. If this occurs, intraocular cautery will likely not be needed but a full core vitrectomy will be needed to clear the blood. A retinal specialist will need to be consulted unless the surgeon is experienced in core vitrectomy techniques.

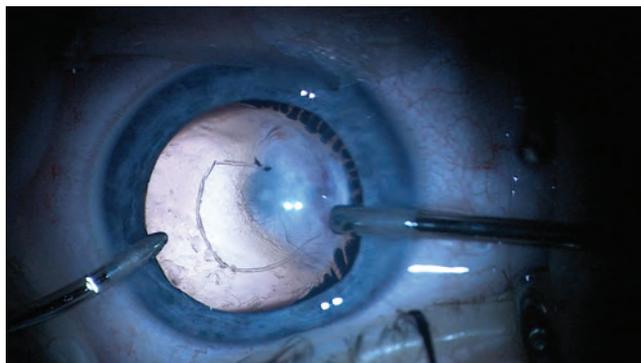


Figure 49.6. Vascularized plaque and stretched ciliary processes in a 4-month-old child.

Intraocular Lens Complications

The most common intraoperative complication related to the IOL is malplacement or malpositioning of the implant on entry into the eye (Fig. 49.7). When PMMA IOLs were used, asymmetrical fixation was common, especially in infants—the leading haptic into the capsular bag but unintended placement of the trailing haptic in the ciliary sulcus. Placing an oversized rigid IOL into a small soft eye was a real challenge. After the leading haptic entered the capsular bag, the OVD often exited the eye through the large wound, the pupil became miotic, and the surgeon was pleased just to get the trailing haptic somewhere posterior to the iris. Posterior vitreous upthrust made dialing the lens into the capsular bag more difficult. The use of foldable IOLs has made this complication less frequent. The trailing haptic of the single-piece AcrySof® IOL can be manually placed under the capsulectomy edge using a push-pull instrument. A second hook can be used to pull the iris edge back for better visualization if needed. Care should be taken to place the haptics into the capsular bag before they have unfolded completely. Once the single-piece AcrySof® haptics have unfolded outside the capsular bag, they are more difficult to place into proper position manually. If this happens, the IOL optic should be displaced eccentrically within the capsular bag as much as possible before an attempt is made to pull the haptic out of the ciliary sulcus. The single-piece AcrySof® IOL does not dial easily.

Many pediatric surgeons choose to perform a primary posterior capsulectomy and an anterior vitrectomy prior to implantation of an IOL. An OVD is then used to inflate the remaining capsular “tire.” The IOL must be carefully aimed on entry so that it enters the capsular bag. Fearing the deep entry, some surgeons aim too anteriorly, and the IOL enters the ciliary sulcus. Unlike the situation mentioned earlier, where IOLs dial up into the ciliary sulcus due to vitreous upthrust, IOLs often dial through the posterior capsulectomy when a vitrectomy has already been performed. Even gentle dialing and gentle posterior force on the optic will send a soft foldable IOL through an even modestly sized posterior capsulectomy. In this

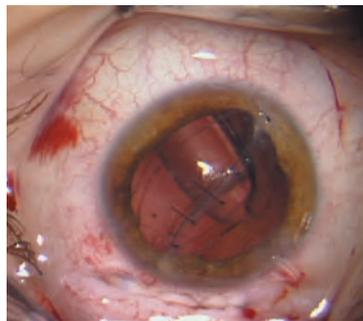


Figure 49.7. Photograph illustrating intraoperative decentration/misplacement of the IOL within the capsular bag in a case of traumatic cataract with repaired corneal laceration.

situation, the IOL is best removed and reinserted, rather than dialed. To avoid this complication and make IOL insertion easier, we recommend IOL insertion into an intact capsular bag prior to the posterior capsulectomy. The IOL haptics should be oriented 90 degrees away from the wound. This allows the vitrector to be placed under the IOL optic more easily for OVD removal or for primary posterior capsulectomy and anterior vitrectomy. To avoid the possibility of dragging a strand of vitreous back to the wound, we recommend removing the OVD after the IOL is in place but performing the primary posterior capsulectomy and anterior vitrectomy through the pars plana, leaving the irrigation cannula in the AC.

Zonular Dialysis

It should be noted that zonular dialysis greatly increases the risk of both vitreous loss and lenticular material sinking into the vitreous. Obviously, IOL implantation may be more difficult. The surgeon needs to evaluate the number of clock hours involved with the zonular dialysis and the surgeon's ability to perform lens aspiration in this setting. When inserted into the capsular sac, an endocapsular ring provides a circumferential expansile force to the capsular equator. However, the endocapsular ring does not always stabilize the lens position and often entraps the lens cortex between itself and the lens capsule. Removal of the trapped cortex can be difficult, and in fact, attempts to do so can cause further zonule dehiscence. Nonetheless, expansion of the capsular sac is often desirable either during or after lens removal, and these devices enhance implant centration and reduce postoperative pseudophakodonesis. Intracapsular rings (e.g., Cionni ring and capsular tension ring segments) are available and are used frequently in adult cataract surgery.

The relatively short length of the iris retractor hook and the single-plane design may cause them to slip off the capsule easily during manipulation of the nucleus. In addition, short iris retractors do not extend into the capsular fornix and, therefore, do not offer support to this region. Because of some of these disadvantages, Mackool designed *capsule retraction devices* specifically shaped for the purpose of retracting the anterior capsule. According to experience during adult cataract surgery, multiple retractors can be placed at 45-degree intervals to provide reliable support to the capsule and the enclosed lens. The elongated return of the retractor extends into the capsular fornix and therefore functions to prevent attraction of the equatorial capsule to the tip of the aspiration handpiece. After lenticular material removal is complete, an endocapsular ring (standard or Cionni design) can be inserted prior to removal of the retractors and insertion of an IOL.

Other Rare Complications

Cloudy cornea, iris sphincterotomy, retained cortex, and lens fragments in the vitreous were reported as intraoperative complications in the IATS cohort.¹

POSTOPERATIVE COMPLICATIONS

When comparing literature reports of postoperative outcomes, a cautionary note should be kept in mind. Results from different studies are difficult to directly compare because of common inconsistencies that include variations in age at surgery, surgical technique, and associated ocular pathology. Major factors to keep in mind when interpreting these results are as follows.

- *Age at surgery.* Children who are the youngest at the time of cataract surgery may also be the ones most prone to subsequent complications. Some studies include only older children, while others have a predominance of infants.
- *Associated ocular anomalies.* Eyes with associated ocular anomalies (such as “complex microphthalmia”) are at high risk for postoperative complications.
- *Etiology of cataracts.* In general, eyes with traumatic cataracts develop a more severe inflammatory response after IOL implantation compared with eyes of similar-aged children with nontraumatic cataracts.
- *Posterior capsule and anterior vitreous management.* Posterior capsule and anterior vitreous management greatly influences the ultimate maintenance of visual axis clarity and ultimate visual outcome in children.
- *Follow-up.* Studies with longer follow-ups will have high rates of opacification. Eyes with an intact posterior capsule tend to opacify between 18 months and 3 years after surgery. Studies that include eyes with <3 years of postsurgical follow-up probably underestimate the incidence of posterior capsule opacification (PCO) in the study population. We have reported that eyes that undergo cataract removal in infancy and receive a primary posterior capsulectomy and vitrectomy (and AcrySof® IOL implantation) tend to be at risk for recurrent opacification mainly during the first 6 months after surgery.⁹ Therefore, studies of this population will predict long-term VAO accurately as long as 6 months of postsurgical follow-up is included.

With this background, we focus on the postoperative outcome of cataract surgery in children.

Visual Axis Opacification

Visual axis opacification (VAO) has been discussed elsewhere in this book. PCO or VAO is most common complication after cataract surgery in children (Figs. 49.8 through 49.15). It is important to note that VAO may occur as closure of a posterior capsule opening, PCO in eyes with intact posterior capsule, lens repopulation into the visual axis, pupillary membrane, reticular fibrosis of the AVF, or corneal edema/opacity. VAO may occur anytime after cataract surgery. For example, a case of soft lens matter recurrence causing VAO 17 years after the

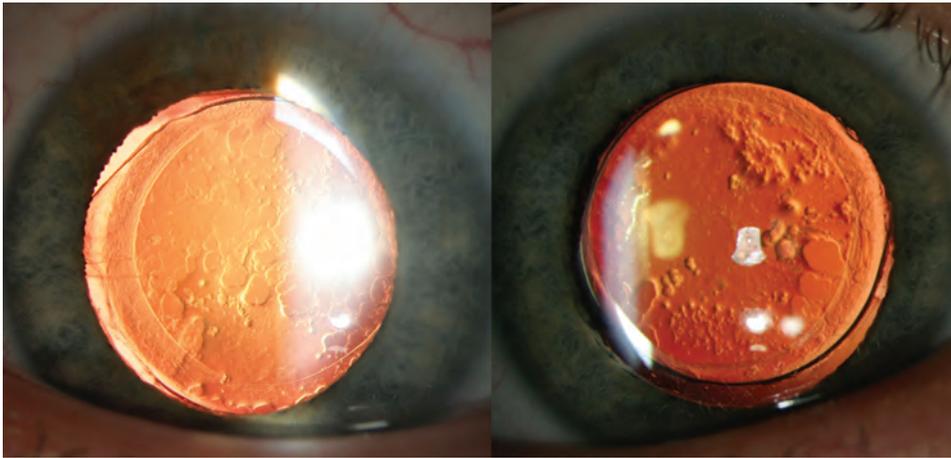


Figure 49.8. Fifteen-month postoperative photo. Note that in both eyes cataract surgery was performed at 6 years of age (at 1-week intervals). Posterior capsule was left intact in both eyes, and 38-D SN60AT IOL was implanted in-the-bag. YAG laser capsulotomy was performed in both eyes. Six months after YAG, the right eye was noted to have closure of YAG opening requiring surgical removal of VAO.

surgical removal of congenital cataract was reported.¹⁰ As discussed throughout the book, primary posterior capsulectomy and vitrectomy are considered essential steps to prevent VAO in the management of pediatric cataract. Researchers are working on other options to prevent this complication (e.g., sealed capsule irrigation, see Chapter 50) (Fig. 49.16).

Glaucoma

Glaucoma can occur during the early or late postoperative period. Although open-angle glaucoma is more common, angle-closure glaucoma is rarely observed (Fig. 49.17) (see Chapter 51).

Toxic anterior segment syndrome (TASS) is a rare inflammatory condition usually observed within the first 2 days after anterior segment surgery.^{11–14} The most common finding is a diffuse corneal edema secondary to damage of the endothelial cell layer. Widespread breakdown of the blood–aqueous barrier is also characteristic, with fibrin formation in the AC and increased AC inflammation, often resulting in a sterile hypopyon. In severe cases, damage to the iris and trabecular meshwork might occur, resulting in glaucoma that can be medically refractory. With intense topical corticosteroid treatment, most cases resolve over a period of weeks to months, with the cornea eventually clearing and the inflammation subsiding. However, severe

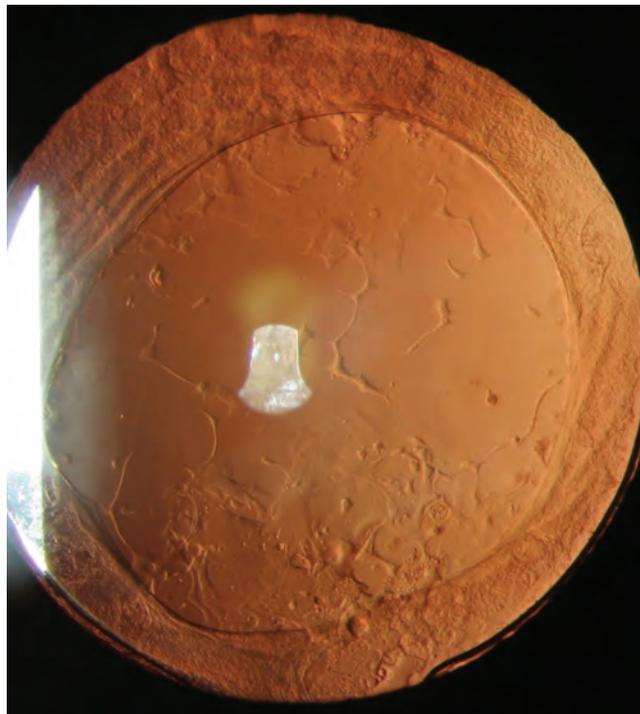


Figure 49.9. PCO documented 19 months after cataract surgery (posterior capsule was left intact, 26-D AcrySof® SN60WF IOL was implanted in-the-bag).

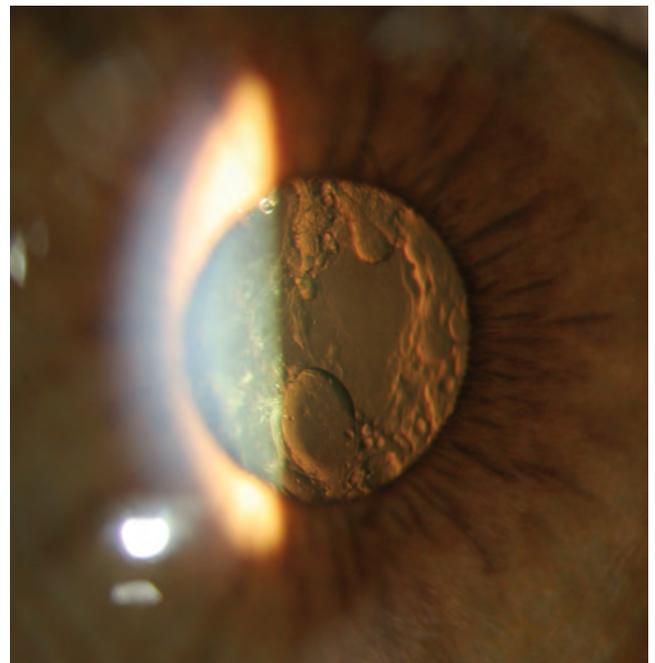


Figure 49.10. Six-year postoperative view of a child operated for cataract surgery at 15 years of age (posterior capsule was left intact at the time of cataract surgery, 21-D AcrySof® SN60WF IOL was implanted in-the-bag).

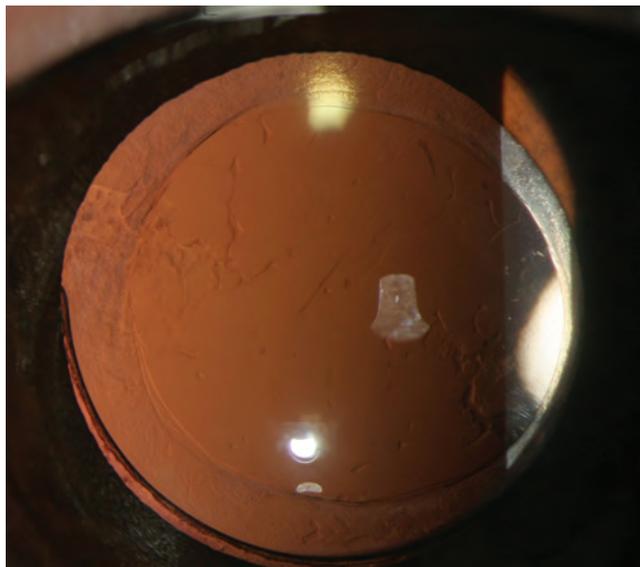


Figure 49.11. Early PCO documented 21 months after cataract surgery (posterior capsule was left intact, 26-D AcrySof® SN60WF IOL was implanted in-the-bag).

cases may result in permanent sequelae, such as persistent corneal edema, iris thinning, a permanently dilated or irregular pupil, peripheral anterior synechia, and glaucoma.

Many substances can cause TASS if they gain access to the AC during surgery or in the immediate postoperative period. Some materials shown to cause TASS include endotoxin; denatured OVDs; preservative and stabilizing agents; heavy metals; residue left behind by substances used during the cleaning and sterilization of instruments; intraocular medications at toxic doses; irrigating solutions with incorrect pH, osmolarity, or ionic composition; and retained OVD or lens cortical material.

Huang et al.¹⁵ report a case of TASS in an 8-year-old child after cataract surgery. Three months after surgery, the central corneal thickness in the operated eye was 831 μ m. Although the visual acuity eventually improved to 20/25, the endothelial cell count was reduced to 851 from

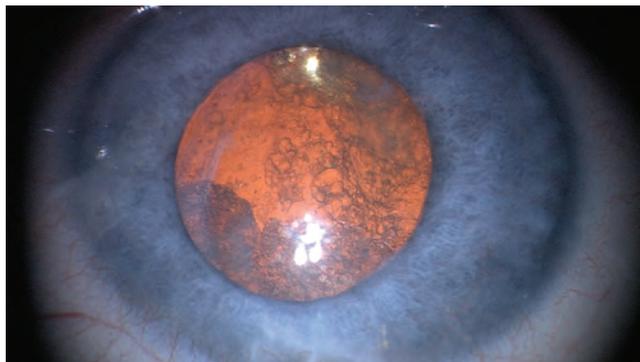


Figure 49.12. Twenty-three-week postoperative view of a child operated for monocular cataract at 9 weeks of age. Pars plicata posterior capsulectomy and vitrectomy were performed along with 35-D AcrySof® SN60WF IOL in-the-bag.



Figure 49.13. One-year postoperative photo of a child operated for cataract surgery at 12 years of age (posterior capsule was left intact, Rayner 570C IOL was implanted in-the-bag).

a preoperative value of almost 3,000. The authors attribute TASS in this child to inadequate rinsing of glutaraldehyde off of the vitreous cutting instrument before its reuse. Ari et al.¹⁶ evaluated 893 eyes of patients undergoing pediatric cataract surgery. TASS was observed in 19 eyes. In all TASS cases, it was noted that ethylene oxide-sterilized vitrectomy packs were used for anterior vitrectomy. After the abolition of use of this material, the authors did not have any new TASS cases.¹⁶

A task force of the American Society of Cataract and Refractive Surgery (ASCRS) has made a number of recommendations for cleaning and sterilizing intraocular surgical instruments, including the following¹¹: (1) keep instruments moist until they are cleaned to avoid debris and viscoelastic agents drying on the instruments; (2) rinse reusable instruments and cannulas with copious volumes of water, as specified by the manufacturer; (3) use disposable cannulas and tubing whenever possible; (4) do not reuse devices labeled for single use; and (5) do not use glutaraldehyde to sterilize intraocular instruments. This has resulted in increased awareness of the causes of TASS and resulted in a greater proportion of surgical centers implementing improved instrument flushing and rinsing protocols to reduce their risk of TASS.¹⁷ It is imperative that pediatric ophthalmologists be familiar with the recommendations of the ASCRS Ad Hoc Task Force on Cleaning and Sterilization of Intraocular Instruments.¹⁸ It may be helpful for pediatric ophthalmologists to provide formal instruction to the operating room and central processing staff at their own hospitals to ensure that the recommendations of the ASCRS Ad Hoc Task Force on Cleaning and Sterilization of Intraocular Instruments are being followed. This is especially important

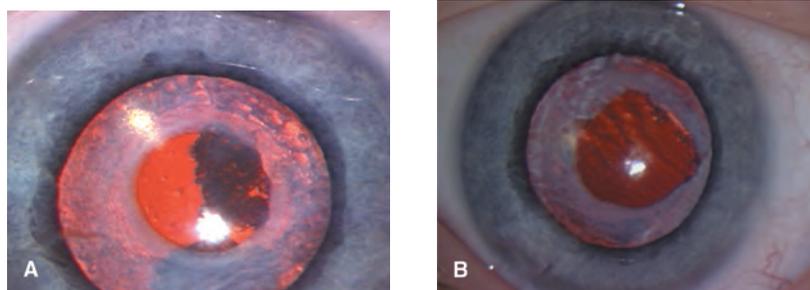


Figure 49.14. **A:** Cortex after proliferations and anterior capsule fibrosis. **B:** After removal of opacification. Proliferated cortex was easy to wash out; however, dense anterior capsule fibrosis was difficult to remove.

if pediatric ophthalmologists are operating in a setup where adult cataract surgeries are not being performed.

Endophthalmitis

See Chapter 12.

Incision and Suture-related Complications

See Chapter 14.

Cornea

Edema

Corneal edema, either focal or diffuse, may be observed after cataract surgery. The IATS defined >30 days of corneal edema as an adverse postoperative complication, and this was observed in 1 of 57 eyes in the IOL group.¹ Reduction in intraocular inflammation and controlling intraocular pressure (IOP) may benefit some cases.

Contact Lens–related Problems

Contact lens–associated corneal problems have been reported in aphakic patients using contact lens correction of their aphakia. Contact lens–associated bacterial keratitis, corneal opacity due to tight contact lenses, and corneal vascularization are the most common among the contact lens–related postoperative complications reported.

Corneal Abrasion

Corneal abrasions are not uncommon in young patients, either from the struggle of contact lens insertion and

removal or from unrelated trauma. The IATS reported a corneal abrasion in 1 eye of the 57 in the contact lens group.

Endothelial Cell Loss Documented by Specular Microscopy

Many factors influence endothelial cell loss after cataract surgery. These include cataract type, axial length, AC depth, surgical technique, surgical experience, size and shape of the tunnel, OVD, irrigation fluids, irrigation volume, type of IOL implanted, intracameral injections, duration of surgery, postoperative inflammation, and IOP. Specular microscopy can provide a noninvasive morphologic analysis of the corneal endothelial cells. Vasavada et al.¹⁹ compared preoperative and 3-month postoperative specular microscopy in children. Comparison of preoperative and postoperative specular microscopy is given here: endothelial cell density, $3,225.1 \pm 346.8$ cells/mm² versus $3,057.7 \pm 330.1$ cells/mm² ($P < 0.001$); coefficient of variation, 27.5 ± 10.6 versus 37.7 ± 16.3 ($P < 0.001$); and percentage of hexagonality, 58.1 ± 15.3 versus 48.6 ± 13.4 ($P < 0.001$). There was a 5.1% decrease in mean endothelial cell loss at 3 months after surgery. No statistically significant difference was noted in the percentage decrease in mean endothelial cell density between eyes undergoing cataract surgery with intact posterior capsules, eyes undergoing manual posterior capsulorhexis without anterior limbal vitrectomy, and eyes undergoing anterior limbal vitrectomy ($P = 0.543$). Nilforushan et al.²⁰ reported that there was no significant difference in the corneal



Figure 49.15. **A:** Fibrous opacification of visual axis in an eye operated on for cataract at 7 months of age. **B:** Kloti radiofrequency diathermy was used to remove the fibrous opacification. **C:** After removal of opacification.

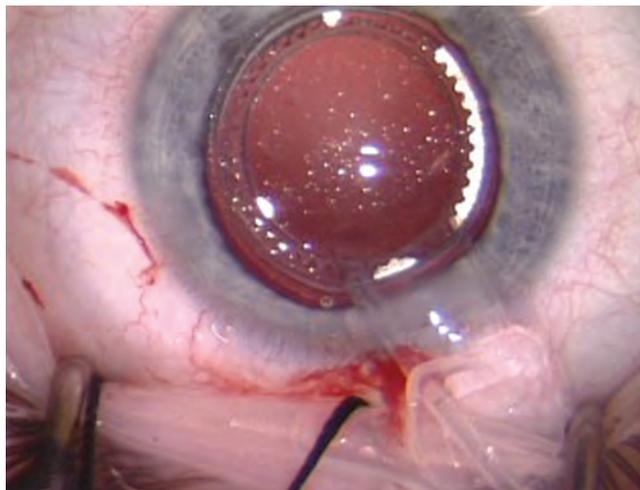


Figure 49.16. Sealed capsule irrigation in pediatric cataract surgery.

endothelial cell count, coefficient of variation, and mean cell area of endothelial cells between operated eyes and the control group. We typically do not perform specular microscopy in all children. Our indications are secondary IOL implantation, Artisan iris-claw IOL implantation, and angle-supported AC IOL implantation. It is important to note that age-related changes occur in children, even without cataract surgery. For example, the number of cells decreases with advanced age. Polymegathism and pleomorphism also increased with age, measured as an increase in the coefficient of variation of cell area and a decrease in the percentage of hexagonal cells with age.

Central Corneal Thickness Changes as Measured by Pachymetry

See Chapter 51.

Anterior Chamber

Shallow Anterior Chamber

A shallow or flattened AC may be observed during the early postoperative period, and a peaked pupil may accompany this finding. This finding is usually from

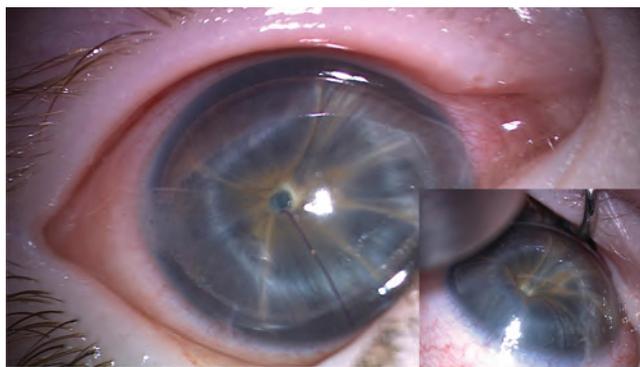


Figure 49.17. Acute angle-closure glaucoma in a child operated for cataract 4 years ago at age 6 months.

a wound leak caused by the child rubbing the eye and forcing fluid out of the sutured wound. The chamber will usually need to be reformed, and the wound may need to be resutured. Although the chamber will usually deepen on its own, failure to surgically reform the chamber and reposition the iris may lead to a peaked pupil that chronically sticks in place and is not easily repaired later.

Fibrinous Uveitis

Fibrinous uveitis due to increased tissue reactivity is a common complication during the early postoperative period in eyes undergoing pediatric cataract surgery (Fig. 49.18). However, modern surgical techniques that limit iris manipulation and ensure capsular bag fixation of the IOL have resulted in less postoperative inflammation/fibrinous uveitis even in small children.

Kuchle et al.²¹ reported that postoperative fibrin formation was less frequent in eyes with an AcrySof® IOL compared to eyes with a PMMA IOL (1 of 10 in the acrylic group versus 9 of 20 in the PMMA group). Uveitis results in fibrinous membrane formation, pigment deposits on the IOL, and posterior synechia formation. Frequent topical steroids and even systemic steroids may be needed in selected cases to reduce uveitis-related complications. Sanghi and Jain²² reported single-bolus intravenous hydrocortisone and dexamethasone for minimizing inflammatory reaction in pediatric cataract and IOL surgery. The use of heparin is described elsewhere in this book (see Chapter 15). Mullaney et al.²³ reported dissolution of pseudophakic fibrinous exudates with the use of intraocular streptokinase (500–1,000 IU) without any adverse effect. Similarly, Klais et al.²⁴ performed fibrinolysis in 11 eyes of 10 children who developed severe fibrin formation despite intensive topical steroid therapy. A complete resolution of fibrin formation was seen in 90% of the children after using 10 µg of recombinant tissue plasminogen activator (rtPA). Besides



Figure 49.18. AC inflammation during early postoperative period.

incomplete resolution and recurrence of membranes, other complications of rtPA use include hyphema, dysfunction of corneal endothelial cells, and corneal band keratopathy. Other possibilities for treating fibrin formation after pediatric IOL surgery are Nd:YAG laser discission, simple mechanical discission, and an intraocular steroid (e.g., dexamethasone) delivery system.

Postoperative Hyphema

A new hyphema occurring postoperatively (as opposed to residual hyphema from an intraoperative bleed) is uncommon, but it can occur. Hiles and Watson⁸ reported that one patient in their series had a total hyphema on the second postoperative day, which cleared spontaneously within 2 weeks. A lightly pigmented fibrinous membrane remained, covering the IOL surface. Recurrent hyphema in an aphakic child was reported.²⁵ Lin and colleagues reported uveitis–glaucoma–hyphema (UGH) syndrome caused by a posterior chamber IOL. A 15-year-old girl underwent pediatric cataract surgery in the both eyes 10 years prior. The pathogenesis of the UGH syndrome was attributed to chafing of the iris by the haptic/optic of the IOL as a result of poor fit, poor design, or malpositioning. It was commonly encountered during AC IOL implantation; however, it was also reported after early-generation posterior chamber IOLs migrated anteriorly.²⁶

Residual Triamcinolone in Anterior Chamber

As discussed in Chapter 22, surgeons are using triamcinolone during cataract surgery. Triamcinolone disappears from AC in most eyes within days after surgery; however, it may persist rarely (Fig. 49.19).

Iris and Pupil

Corectopia and Pupil Shape Abnormalities

Postoperative iris and pupil abnormalities are observed after cataract surgery, more commonly after AC IOL implantation (Figs. 49.20 through 49.23). Corectopia was



Figure 49.19. Residual triamcinolone after cataract surgery. Note that the child underwent cataract surgery 2 weeks ago.

reported in 2% of the eyes in the no-IOL group versus 19% of the eyes in the IOL group in the IATS.¹ Sharma et al.²⁷ reported that an updrawn pupil was observed in 38.5% of the eyes with pediatric cataract surgery and IOL implantation. Dislocated or poorly sized angle-supported AC IOLs cause peaked pupils more frequently compared to posterior chamber IOLs.

Iris Erosion and Atrophy

Iris erosion and atrophy are more likely to develop after implanting iris-supported lens. Intraoperative iris trauma may contribute to this postoperative complication.

Heterochromia

Heterochromia iridis (Fig. 49.24) is an asymmetry of iris color in 1 eye in relation to the other. Summers and Letson reported it in 9% of patients.²⁸ Cataract surgery stimulates a prostaglandin release resulting in the darkening of iris color, which may occur through the same or a similar mechanism by which latanoprost causes darkening of iris color. The darkening effect occurs more often



Figure 49.20. Postoperative view of bilateral secondary AC IOL implantation.

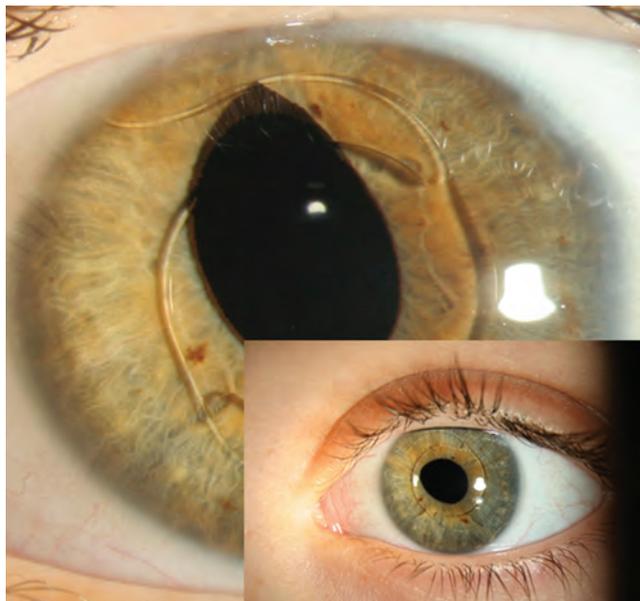


Figure 49.21. Iris abnormality in a child with AC IOL. Inset shows the same eye 1 year ago.

when surgery is done in infancy. Lenart and colleagues evaluated 15 children.²⁹ Photographs were taken of both eyes. Masked examiners reviewed the photographs and compared, in each patient, the iris color of the eye that was operated and the eye that was not operated. Thirteen of fifteen children had darker iris color in the operated eye relative to the nonoperated eye.

Intraocular Pressure Related

IOP Spike

As in adults, retained viscous OVDs can cause a marked postoperative IOP elevation after surgery for childhood cataracts. Englert and Wilson³⁰ have suggested the need for more meticulous removal of OVDs. We reported a high incidence of symptomatic early IOP spikes in

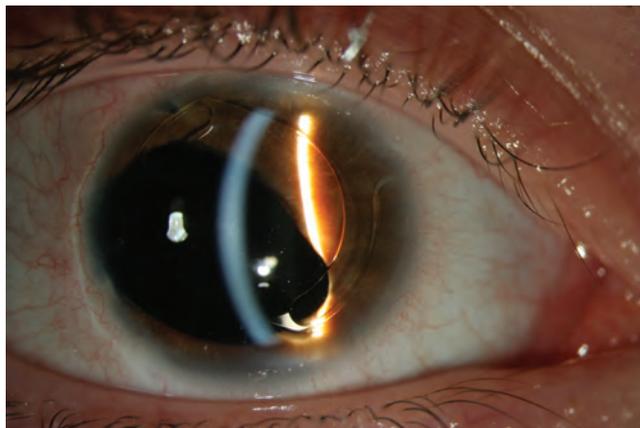


Figure 49.22. AC IOL in a 19-year-old child. IOL was implanted at 6 years of age.

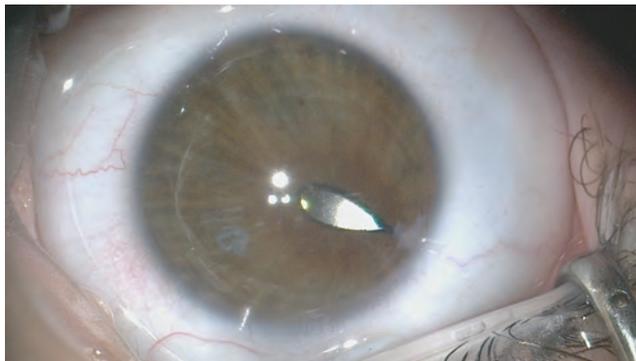


Figure 49.23. Peaked pupil after 8 months after secondary IOL implantation.

patients with aphakic glaucoma undergoing secondary IOL implantation, and we recommended the use of topical and/or systemic glaucoma medications to help prevent or minimize the IOP spike. Monitoring during the early postoperative period was also suggested in these higher-risk cases.³¹

Postoperative Hypotony

Early hypotony may indicate a wound leak. Once this is ruled out, sustained hypotony may accompany a postoperative retinal detachment (RD). In glaucoma and uveitis patients, sustained hypotony can be an impending sign of phthisis bulbi. Hypotonus maculopathy may also be present. In patients with nanophthalmos, mild hypotony may be associated with large choroidal effusions. These usually resolve with time.

Anterior Capsule Fibrosis

Excessive anterior capsule fibrosis and shrinkage of the CCC opening can lead to difficulty with retinoscopy and with examining the retinal periphery and, occasionally, decentration of the IOL.

Intraocular Lens

Deposits on the IOL Surface

Precipitates composed of pigment, inflammatory cells, fibrin, blood breakdown products, and other elements are often seen during postoperative period on the surface of an IOL optic implanted in a child (Figs. 49.25 and 49.26).



Figure 49.24. Heterochromia iridis.



Figure 49.25. Deposits on IOL surface. (Courtesy Dr. Abhay R. Vasavada, Ahmedabad, India.)

The deposits can be pigmented or nonpigmented but are usually not visually significant. They occur much more commonly in children with a dark iris and when compliance with postoperative medications has been poor. The site of IOL implantation can also influence the formation of deposits. For example, sulcus-fixated IOL would likely have more deposits compared to bag-fixated IOL. In addition, Vasavada and Trivedi³² found that the incidence of deposits was higher in eyes with the IOL optic captured through the PCCC in comparison with in-the-bag fixated IOLs. The type of IOL also influences IOL deposits. Wilson et al.³³ reported that IOL cell deposits were seen in 7 of 110 (6.4%) hydrophobic acrylic lenses, compared with 26 of 120 (21.75%) PMMA lenses. In addition, the incidence of deposits is inversely proportional to age at surgery.

Posterior Synechia

Younger age at the time of cataract surgery increases the risk for synechia formation. Wilson et al.³³ noted posterior synechia in 5 of 110 AcrySof® lenses (4.5%), compared with 23 of 120 PMMA lenses (19.2%). Evaluating

single-piece IOLs in children, we noted synechia in 5 eyes (11.9%).³⁴ None produced enough corectopia to cause a noticeable cosmetic deformity. In most cases, the synechia were pinpoint adhesions of the iris to the anterior capsulectomy edge. No adhesions were seen between the iris and the IOL. Vasavada et al.³⁵ noted posterior synechia in 13.6%. The incidence of synechia formation was significantly higher when operated at younger age. Trivedi et al.⁹ noted that synechia were seen in 31% of eyes in the first year of life.

IOL Malposition

In a recent review from Toronto, IOL repositioning was required in 4/55 eyes.³⁶

Pupillary Capture

Placing the IOL in the capsular bag with an anterior capsulectomy smaller than the IOL optic helps to prevent pupillary capture, a complication that is much more common in children than in adults (Fig. 49.27). It often occurs in association with posterior synechia formation and PCO. Pupillary capture occurs most often in children <2 years of age, when an optic size <6 mm is used, and the lens is placed in the ciliary sulcus. Pupillary capture can be left untreated if it is not associated with decreased visual acuity or glaucoma. However, surgical repair recreates a more round pupil shape and IOL centration. Fixation of posterior chamber intraocular lenses (PCIOLs) in the capsular bag (whenever possible) is recommended to decrease the incidence of this complication. Also, the anterior capsulectomy should be smaller than the IOL optic if possible. Prolapsing the optic of a secondary sulcus-fixated IOL through the anterior capsulorhexis opening can also prevent pupillary capture.

Decentration/Dislocation

Excessive capsular fibrosis and asymmetric IOL fixation are the most common causes leading to decentration of an IOL. Decentration of an IOL can also occur because of traumatic zonular loss and/or inadequate capsular

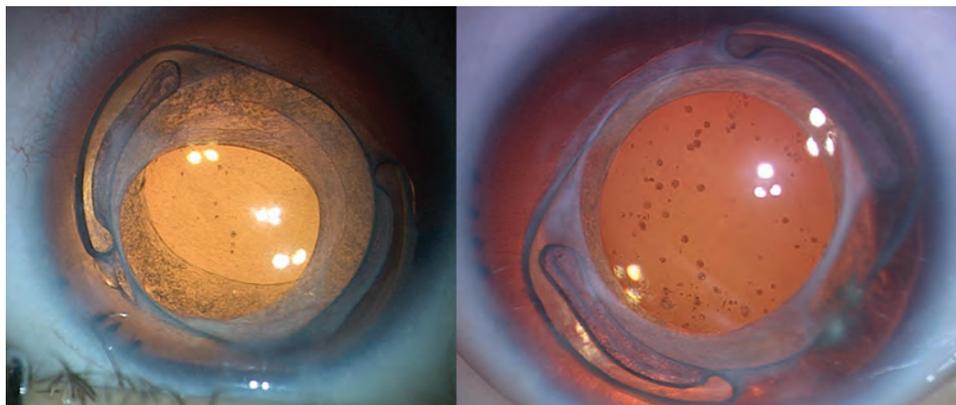


Figure 49.26. Deposits on IOL.



Figure 49.27. A 10-year-old child having cataract surgery with IOL implantation surgery done at 10 months of age. Child presented to us with IOL captured by the pupil and high myopia. IOL was exchanged.

support (Fig. 49.28). Capsular bag placement of the IOL is the most successful way to reduce this complication. Posterior capture of the IOL optic also resulted in better centration of the implanted IOL.³² Explantation or repositioning of the IOL may be necessary in some cases presenting with significant decentration/dislocation.

Delayed Postoperative Opacification of Foldable IOLs

Late postoperative opacification of some specific models of hydrophilic IOLs in adults has been reported widely in the literature. In these cases, the opacification was severe enough to require the IOL to be explanted. The majority of reports concern adult cases and are reported from Asia, Australia, Canada, Europe, Latin America, and South Africa. One report included opacification of hydrophilic acrylic IOLs in children and suggested that there may be a special pattern of dystrophic calcification in this population. Kleinmann et al.³⁷ reported the clinicopathologic and ultrastructural features of three hydrophilic acrylic IOLs, manufactured from two different biomaterials,

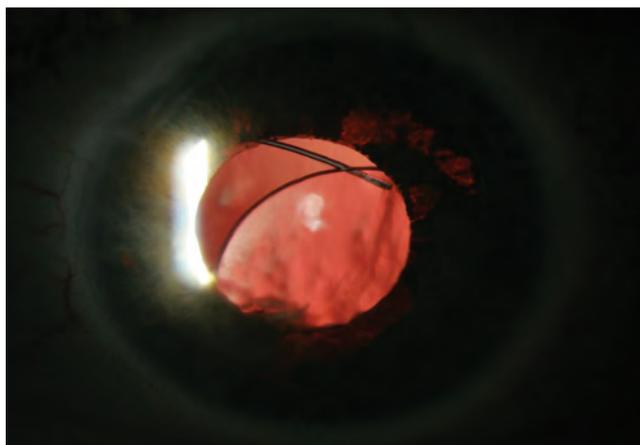


Figure 49.28. IOL decentration.

explanted from children who had visual disturbances caused by progressive postoperative opacification of the lenses' optic component. These lenses were explanted at 20, 22, and 25 months postoperatively, from children aged 10, 36, and 20 months, respectively, at lens implantation. Pehere et al.³⁸ reported that the deposits were found to be composed of calcium, phosphate, and silicone.

Glistening

Mullner-Eidenbock et al.³⁹ noted glistening in both eyes of one patient 1 week postoperatively. The glistenings increased during the first 2 postoperative years to a degree of 3+ and then remained stable until the last follow-up at 40 months. Glistenings are very commonly observed on postoperative slit-lamp examination when a single-piece AcrySof® IOL has been implanted. They appear to be visually insignificant, but some surgeons argue that they may have some yet unproven deleterious affect on visual performance.

Retina, Choroid, and Macula

Retinal Detachment

RD is a serious, often delayed complication of pediatric cataract surgery. In the past, unilateral surgery for bilateral congenital cataracts was advocated because of the reported high incidence of subsequent RD and the poor outlook for successful RD repair.⁴⁰ When the inevitable RD occurred approximately 25 years later, the cataract in the remaining eye could then be removed with the hope of restoring useful vision and function for another 25 years, until this eye in turn sustained a RD. Previous studies reporting this complication relate to the older techniques of cataract surgery and may not be strictly applicable to the currently used techniques. The incidence of RD following pediatric cataract surgery appears to have decreased markedly as surgical techniques have advanced and evolved. The use of a modern high-speed vitreous cutter reduces the traction on the retina. The increased use of a pars plana approach to the anterior vitreous (with anterior irrigation) has also decreased vitreous prolapse or incarceration into an AC wound, thus further decreasing the rate and risk of late RD.

A long-term follow-up is critical in children, as RD can occur many years after cataract surgery. A detailed retinal examination is recommended after cataract surgery at least yearly. This is especially important for those eyes at higher risk for RD by virtue of a long axial length for age, persistent fetal vasculature, diagnosis of a syndrome known to be associated with liquefaction of the vitreous, or multiple surgeries.

In 1957, Cordes reviewed 56 globes enucleated following congenital cataract surgery. He noted 23 had RD, more than half of which occurred following multiple needling

procedures. In 1974, Taylor and Tasman⁴¹ reported RD as a complication of pediatric cataract surgery—the patients were predominantly males and most were in the second and fifth decade of life. In 1980, Toyofuku et al.⁴² reported that RD was typified by a high incidence of men, myopia, preference for the second and fourth decades of life, and a fairly long interval after cataract surgery. In 2005, Rabiah reported a multivariable Cox proportional hazards regression analysis with adjustment for intrasubject correlation. An aphakic refractive error that was more myopic than the age-adjusted aphakic norm and a history of a post-ataract surgery wound dehiscence were found as predictors of RD. A primary posterior capsulectomy/anterior vitrectomy procedure was not predictive of RD.⁴³ High myopia, peripheral retinal degenerations, and vitreous loss have also been found to be risk factors for the development of RD.

RD developed from 10 days to 52 years after cataract surgery, with an average time lapsed of 22.8 years.⁴² RD occurred within the first year after cataract surgery in 9.6% of these cases; RD developed in 72% after an interval of more than 10 years. Kanski and others found an average development period of 33 years. RD was diagnosed at a mean of 6.8 ± 4.4 years after cataract surgery (range 0.4–14.8 years).⁴³ In 1974, Taylor and Tasman⁴¹ reported that the interval between cataract surgery and RD varied between 21 days and 51 years, with median interval of 21 years. The average interval between cataract surgery and subsequent development of a detachment in that eye was 33.3 years in the Kanski et al.⁴⁴ report. In 1983, it was noted that the interval between the initial cataract surgery and first presentation with detachment was 28 years (range 1–51 years).⁴⁵

Patients may present with loss of central vision, seeing a veil, floaters, or flashes or may be asymptomatic. Patients whose vision had always been poor might have easily overlooked the symptoms of RD. Choices made in the intraoperative management of vitreous may alter the likelihood of late RD. Chapter 21 will guide the surgeon toward appropriate techniques to help minimize the risk of intraoperative retinal injury and aid in the timely and appropriate diagnosis of posterior segment pathology following pediatric cataract surgery.

If a vitreous wick is seen attached to one of the anterior cataract wound (corneal tunnel or paracentesis) during a postoperative examination, it should be removed as soon as possible to relieve any traction on the retina at the vitreous base. During surgery, wound sweeping (Weck-Cel vitrectomy) should likewise be avoided. This technique can remove vitreous from the wound, but it invariably induces acute vitreoretinal traction. Bimanual technique with separate cutter and infusion is far better than using the coaxial infusion sleeve. Taylor and Tasman⁴¹ reported surgery RDs after congenital cataract surgery results in reattachment of the retina in 76% of the eyes.

RD as a postoperative complication is not very commonly seen with techniques used during the last decade. Eyes with traumatic cataract, ectopia lentis, and Stickler syndrome are at higher risk for the development of this complication. As RD may develop many years after surgery, long-term follow-up is warranted.

Hemorrhagic Retinopathy

This complication may occur following infantile cataract surgery in up to one-third of eyes.^{46,47} It presents with flame-shaped retinal hemorrhages and may be associated with concurrent vitreous hemorrhage. The hemorrhages develop during the first 24 hours following surgery, are nonprogressive, and typically resolve without sequelae within a few weeks.

Cystoid macular edema (CME)

Cystoid macular edema (CME) is a rare complication following pediatric cataract surgery, probably because of the healthy retinal vasculature and formed vitreous in children. It occurs with unknown frequency after pediatric cataract extraction, in part due to the difficulty in detecting CME in the pediatric patient because of the challenges of performing macular examination, inability to visualize CME with the indirect ophthalmoscope or RetCam, the sedation issues associated with fluorescein angiography, and the inability of children to position for ocular coherence tomography (OCT). Because of the difficulty of performing fluorescein angiography in young patients, surgeons seldom evaluate children for this complication.

Reports attempting to assess CME have concluded that its occurrence in the pediatric population appears to be infrequent. If detected and visually significant, the treatment should therefore parallel guidelines for adult pseudophakic CME, including topical corticosteroids and nonsteroidal anti-inflammatory medications.

Early reports of cataract surgery in children indicated that the incidence of CME may be lower in these eyes. Hoyt and Nickel,⁴⁸ however, reported a CME incidence of 37% in children undergoing lensectomy and vitrectomy. Ahmadiet al.⁴⁹ evaluated 45 eyes of 31 children undergoing cataract surgery and IOL implantation, using intravenous fluorescein and fundus fluorescein angiography, and did not detect CME in any eye at 6 weeks after surgery. Rao et al.⁵⁰ performed a similar evaluation 4 to 6 weeks after surgery, using oral fluorescein and angiography, in 25 eyes of children and did not detect CME in any of them. The prevalence of CME is higher in children with uveitis. The use of nonsteroidal anti-inflammatory drugs is uncommon after pediatric cataract surgery but may be efficacious in high-risk groups or when CME is documented.

As seen above, despite the need to violate the vitreous in young children, it appears that such a procedure, using modern technology and techniques, does not predispose

the child to an increased risk of CME. The reasons for this are not clear, since similar procedures in adults tend to show a higher occurrence of CME. It may be that the pediatric eye is structurally more resistant to the effects of anterior segment manipulations. The youthful structure of the vitreous gel is likely the key difference. Improved understanding of the pathophysiology of intraocular inflammation and its role in the causation of CME has resulted in better treatment approaches in the postoperative period. This has led to improved outcomes of cataract surgery in children. Despite these advances, there are still many unanswered questions. These include the possibility of late-onset changes in the macula of these eyes, sequelae of posterior vitreous detachments as these children grow older, and the relevance of some of the newly described methods to treat CME, such as intravitreal triamcinolone injections and pars plana vitrectomy, in children.

Of note, macular hypoplasia was reported to be associated with familial cataract.⁵¹ This condition may be more common than suspected and most of the time identified when visual outcomes do not match clinical expectations and the investigator searches for a cause.

Refractive Error

Surgically Induced Astigmatism

To increase safety during wound healing, pediatric cataract surgeons typically place sutures. Since children rub their eyes after surgery and the wounds are prone to leaking, the sutures are placed and tied tight. This can induce large amounts of surgically induced astigmatism in the immediate postoperative period. However, relaxation of a large amount of this suture-induced astigmatism occurs in children having cataract surgery when the wound heals and the Vicryl suture dissolves.^{52–60} Spierer and Nahum⁵⁴ reported that mean astigmatism 1 week postoperatively was 5.8 ± 3.6 D (range 3.0–14.0 D). Thereafter, the astigmatic component of the refractive error underwent a spontaneous steady decline, reaching a mean value of 2.1 ± 1.3 D (range 1–4.0 D) 5 months after surgery. Bradfield et al.⁶¹ reported that the mean postoperative retinoscopic cylinder in all children was 0.63 D (range 0.0–4.50 D) at 1 month, 0.40 D (range 0.0–1.75 D) at 6 months, and 0.51 D (range 0.0–2.50 D) at 1 year. Less astigmatism was observed in children having surgery before they were 36 months old. Bothun et al.⁶² reported that the mean cylindrical correction on the first postoperative day was 6.6 D, which dropped to 1.9 D between 2 and 4 weeks and 1.2 D by 3-month postoperative visit. Gupta et al.⁶⁰ reported that the mean postoperative astigmatism in eyes with a scleral incision was 1.3, 1.4, and 1.4 D at 1, 3, and 6 months, respectively. The mean astigmatism in eyes with a clear corneal incision was 1.3, 1.1, and 1.0 D at 1, 3, and 6 months, respectively.

Myopic Shift

See Chapter 52. High postoperative myopia can be found, and optical rehabilitation with contact lens, spectacles, or IOL exchange will be necessary. Alternatively, secondary implantation of piggyback lens in the ciliary sulcus or corneal refractive surgery can be performed. Secondary Artisan phakic IOL for correction of progressive high myopia in a pseudophakic child has also been reported.⁶³

Postoperative Ptosis

Sharma and colleagues²⁷ reported ptosis in 1/39 (2.6%) children. We have observed this in a few children operated for cataract surgery. None have required surgery for the eyelid ptosis.

Phthisis Bulbi

In 1985, Gieser et al.⁶⁴ reported phthisis bulbi after an iris-supported Binkhorst two-loop IOL. The child was operated for cataract at 4 years of age. Four years later, the implant dislocated anteriorly, and it was removed. Eleven years postoperatively, the eye was blind, painful, and shrunken. The ongoing IATS reported phthisis bulbi in 1 eye of the 57 who did not receive an IOL.¹

SUMMARY

Postoperative complications may develop in the early postoperative period or after many years. Therefore, it is crucial to follow children closely on a long-term basis after pediatric cataract surgery.

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Opacification of the Ocular Media

Mohammad Ali Javadi, Hamid Ahmadi, and Sepehr Feizi

Media opacification after cataract surgery is more common in children than in adults. It interferes with the main goals of the operation and is a potential cause of amblyopia. Media opacification may develop rapidly or insidiously. Examples of sudden-onset media opacification include corneal edema, inflammatory pupillary membrane, and vitreous hemorrhage; these events are usually identified promptly, sometimes by the parents or caretaker, and usually resolve with appropriate treatment and with time. In contrast, some types of media opacity are delayed and tend to be progressive, including cellular, fibrinous, or pigmentary deposits on the intraocular lens (IOL) and posterior capsule opacification (PCO) or hyaloid face opacification. Since children may be unable to report decreased vision, these may go unnoticed and lead to irreversible visual damage.

Posterior capsular opacification remains the most common complication of pediatric cataract surgery¹⁻³ (Fig. 50.1). PCO can be treated using Nd:YAG laser capsulotomy or surgical posterior capsulectomy. Nd:YAG laser technique involves clearing the visual axis by creating a central opening in the opacified posterior capsule.^{1,4} Although this procedure is easy and quick, there are complications, including retinal detachment, damage to the IOL, cystoid macular edema (CME), an increase in intraocular pressure (IOP), iris hemorrhage, corneal edema, IOL subluxation, and exacerbation of localized endophthalmitis.^{1,4,5}

Posterior capsular opacification is more threatening in young adults and children, with a higher incidence, quicker onset, and greater amblyogenic effect. Additionally, PCO in young children is often dense (see Fig. 50.1) and may need to be removed with additional surgery, which carries more risks of potential complications.^{6,7} This chapter focuses on PCO and its incidence, predisposing factors including patient characteristics, IOL features, surgical techniques, preventive measures such as primary posterior capsulectomy accompanied by anterior vitrectomy, and intraoperative cytotoxic agents. Additionally, other less common causes of media opacification are reviewed.

POSTERIOR CAPSULE OPACIFICATION

Until the late 1960s, the preferred method of pediatric cataract surgery was lens aspiration, leaving the posterior capsule intact as popularized by Scheie.⁸ With this method, remaining lens epithelial cells (LECs) proliferate and migrate on the intact posterior capsule, leading to opacification of the once-clear capsule. In some cases, the ensuing opacity causes more visual deterioration than the original cataract itself.⁹ After the advent of automated posterior capsulectomy and vitrectomy in 1976, pediatric cataract surgeons began to perform lensectomy with anterior vitrectomy through a large opening in the posterior capsule, leaving only a 1- to 2-mm rim behind.¹⁰ This new trend quickly caught on and was used by the majority of pediatric cataract surgeons by the mid-1980s. It resulted in superior media clarity.¹⁰ Further refinements of surgical technique, improved IOL quality, and favorable results of IOL implantation in adults started a trend toward IOL implantation in children in the early 1990s. However, opacification of the retained lens capsule still led to poor vision and amblyopia.^{10,11} The secondary opacification and fibrosis may be severe enough to cause IOL decentration and even break the optic-haptic junction.¹² Even in the era of modern cataract surgery, PCO still poses a challenge for ophthalmologists.¹³⁻¹⁵

Mechanism of PCO Development

LECs left behind in the capsular bag after any type of extracapsular cataract surgery are mainly responsible for PCO development.⁹ Proliferation, migration, epithelial-to-mesenchymal transition (EMT), collagen deposition, and lens fiber regeneration of LECs are the main causes of opacification. It appears that cataract surgery induces a wound healing response in the lens, and retained LECs proliferate and migrate across the posterior capsule and undergo lens fiber regeneration and EMT.¹⁶⁻¹⁸ In vitro studies and animal models of PCO suggest that several cytokines and growth factors play a major role in the pathogenesis of PCO.^{19,20} Studies show that levels of



Figure 50.1. A dense posterior capsular opacification develops after lensectomy without posterior capsulectomy for congenital cataract.

several cytokines and growth factors increase in aqueous humor and influence the behavior of the remaining LECs after cataract surgery. These factors include transforming growth factor β (TGF- β), fibroblast growth factor 2 (FGF-2), hepatocyte growth factor, interleukins 1 and 6 (IL-1 and IL-6), and epithelial growth factor.^{19,21} TGF- β plays a central role in the cell biology of PCO. Clinically, there are two morphologic types of PCO, the fibrosis type and the pearl type. Fibrosis-type PCO is caused by the proliferation and migration of LECs, which undergo EMT, resulting in fibrous metaplasia and leading to significant visual loss by producing folds and wrinkles in the posterior capsule.¹⁶ Pearl-type PCO is caused by the LECs located at the equatorial lens region (lens bow) causing regeneration of crystallin-expressing lenticular fibers and forming Elschnig pearls and Soemmering ring, responsible for most cases of PCO-related visual loss.^{2,22}

Incidence and Impact of PCO

PCO has been reported in 39% to 100% of pediatric cases in which the capsule is left intact.^{7,9,13,14,23–46} Taylor⁴⁷ reported 32 reoperations for capsular opacity in 28 pediatric eyes with an intact posterior capsule following lens aspiration. The incidence of PCO depends on the age at surgery, the type of cataract (congenital, developmental, or traumatic), aphakia versus pseudophakia, ciliary sulcus versus capsular bag fixation, associated ocular abnormalities (e.g., persistent fetal vasculature [PFV], microcornea), and preexisting systemic conditions such as juvenile rheumatoid arthritis. Additionally, the duration of follow-up affects the incidence of PCO; the longer the follow-up, the more PCO is manifested.

The burden of PCO is reflected in numerous reports. Basti et al.⁴³ reported the mean time for PCO appearance

to be 3 months after surgery. Jensen et al.³⁷ observed significant PCO 1 to 26 months after cataract surgery in children with a mean age of 7.3 years. The peak incidence of PCO was 18 months after surgery; thereafter, the curve flattened considerably. PCO was more common in younger children: 64% of children 1 to 6 years old versus of those 6 to 13 years old developed PCO.³⁷ Gimbel et al.⁴² reported a cumulative rate for Nd:YAG laser capsulotomy of 17%, 42%, 52%, and 59% at 1, 2, 3, and 4 years after surgery, respectively. Plager et al.¹⁴ observed a mean duration of 2 years for PCO development. Plager et al.¹⁴ reported PCO in 90% of 71 eyes with a retained posterior capsule by 3.5 years in children 10 months to 17 years of age. According to Crouch et al.,²⁹ PCO developed in 72% of 35 eyes of children 5 to 18 years of age undergoing cataract surgery and IOL implantation with retention of the posterior capsule. Müllner-Eidenböck et al.⁴⁸ observed PCO in 9 out of 15 eyes (60%) of children 6 to 16 years of age with an intact posterior capsule and acrylic IOL implant. Hosal and Biglan³⁶ observed secondary capsular membranes in 78.6% of operated eyes 3 weeks to 53 months after surgery; they reported a 10.7-fold risk of media opacity with posterior capsule retention. In the same study, eyes with a primary posterior capsulectomy but no vitrectomy developed secondary media opacity 42.9% of the time compared to 22.5% when both a posterior capsulectomy and an anterior vitrectomy were done at the time of the cataract aspiration. Age <1 year at surgery was also an independent risk factor with a 4.7-fold added risk of secondary media opacity.

Recent modifications in IOL design (i.e., square edge profile) and improved biocompatibility (e.g., acrylic) have significantly reduced the incidence of PCO in adults; however, there are insufficient data to know whether these new lenses result in a commensurate reduction in children.^{48,49} Therefore, current options for prevention of PCO include primary posterior capsulectomy (with or without vitrectomy) or postoperative Nd:YAG laser disruption of the posterior capsule before opacification.⁵⁰ Disadvantages of the latter method include the lack of sufficient cooperation by children and possible need for a second anesthesia, need for a special Nd:YAG laser capable of functioning with the patient in the supine position, risk of IOL damage, and, above all, risk of recurrent media opacity on the anterior vitreous or hyaloid face. A limitation of the Nd:YAG laser capsulotomy is that it does not address the problem of the residual intact anterior vitreous face that provides a scaffold on which residual lens fibers grow to create secondary opaque membranes. The anterior hyaloid face not only serves as a scaffold for migration and metaplasia of LECs but also acts as a surface for deposition of inflammatory cells, debris, and pigment.⁵¹ This procedure is not advisable in very young children. Atkinson and Hiles⁷ reported on 32 eyes of 28 children undergoing cataract surgery with or

without IOL implantation who received postoperative Nd:YAG laser capsulotomy with the Microruptor III, which is capable of rotating 90 degrees. In 16 eyes in which laser capsulotomy was performed earlier than 4 weeks, the mean energy required was 114 mJ; however, in the remaining eyes, the procedure was performed later than 4 weeks, and the mean energy required was 324 mJ. Overall, the rate of membrane reformation was 25% in this series. Hutcheson et al.³⁸ compared the incidence of media opacity between pediatric cataract extractions with primary posterior capsulectomy (33 eyes) and those with postoperative laser disruption (23 eyes); 3% versus 57% of the eyes, respectively, developed media opacity needing further treatment. Furthermore, in the second group, 17% required repeat laser treatment after the second laser session. Koch and Kohnen⁵² noted that all 5 eyes with intact posterior capsules required Nd:YAG laser capsulotomies within 18 months of the initial surgery and 3 eyes required a second laser treatment within 9 months of the first one. In a study of PCO rates after foldable acrylic IOL implantation, Müllner-Eidenböck et al.⁴⁸ observed that 60% of the eyes belonging to the younger age group (<4 years) developed recurrent opacification following Nd:YAG laser treatment. Based on this observation, the authors concluded that surgical capsulectomy combined with anterior vitrectomy rather than Nd:YAG laser capsulotomy may be needed in young children to maintain a clear visual axis. Laser capsulotomy is often ineffective in the presence of fibrous PCO. When a dense retrolental membrane develops, a more invasive approach, using the vitrectomy handpiece, is required.

Primary posterior capsulectomy with or without anterior vitrectomy decreases the chance for PCO, and it is believed to be safe (Fig. 50.2). Dahan and Salmenson⁵³ suggested removal of the center of the posterior capsule with a vitrector in association with IOL implantation. Buckley et al.⁵⁴ observed no media opacity by 13 months in 20 eyes undergoing IOL implantation followed by primary posterior capsulectomy and anterior vitrectomy through a pars plana approach. Luo et al.⁵⁵ observed that the rate of fibrous exudates in the anterior chamber and the rate of IOL subluxation or capture were significantly lower when posterior continuous curvilinear capsulorhexis (PCCC) and anterior vitrectomy were performed in children aged between 2 and 5 years. They concluded that primary PCCC with anterior vitrectomy is an effective procedure with a low PCO rate in pediatric cataract. However, opponents of this approach believe that retention of the posterior capsule without concomitant vitrectomy may decrease the risk of CME, retinal detachment, and vitreous incarceration into the surgical wound, facilitate in-the-bag IOL implantation, and provide long-term IOL stability.⁵⁰

Opinions differ regarding the effect of primary posterior capsulectomy on CME. Hoyt and Nickel⁵⁶ operated

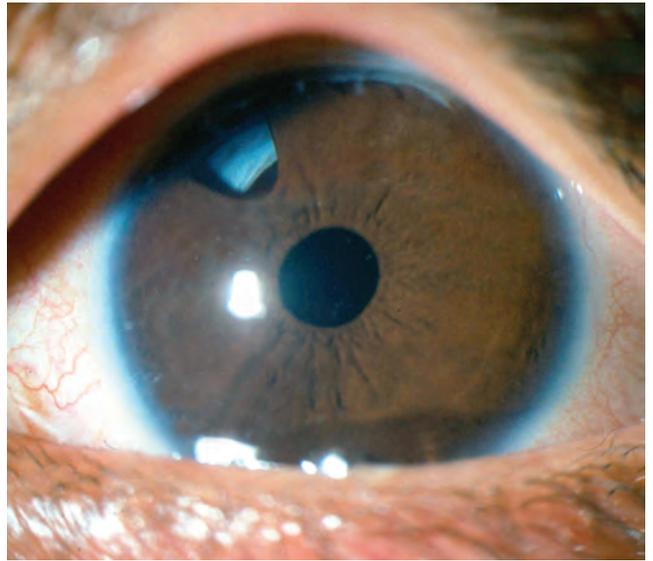


Figure 50.2. A clear media after lensectomy, primary posterior capsulectomy, vitrectomy, and in-the-bag IOL implantation.

on 27 children with bilateral cataracts: one eye of each patient underwent lensectomy and vitrectomy; the fellow eye was treated with lens aspiration and discission of the posterior capsule. In the first group, 10 eyes developed CME, while no instances were noted in the fellow eyes. On the other hand, other investigators have reported different results. Gilbard et al.⁵⁷ reported only one case of suspected CME on angiographic and angioscopic examination of 25 eyes of 17 patients undergoing lensectomy and anterior vitrectomy. Green et al.⁵⁸ observed no cases of CME in 52 eyes undergoing pars plicata lensectomy and vitrectomy. In a series of 24 children undergoing bilateral cataract surgery and IOL implantation, Gimbel et al.⁴⁶ reported only one case of bilateral CME in a patient with retinitis pigmentosa. Pinchoff et al.⁵⁹ reported on lensectomy and vitrectomy (12 eyes), extracapsular cataract extraction and discission (5 eyes), extracapsular cataract extraction (3 eyes), and secondary discission (3 eyes); in none of the eyes, CME was detectable by angiography. Ahmadiéh et al.⁶⁰ also reported no case of CME on the angioscopic examination of 38 eyes undergoing limbal or pars plana lensectomy, anterior vitrectomy, and posterior chamber IOL implantation. It is generally difficult to diagnose and document CME in children as it is a rare event. Even more notable is the low retinal detachment rate after this procedure.⁵⁶ This can be partly attributed to the presence of a well-formed vitreous in the child's eye.

Overall, current evidence supports the idea that primary posterior capsulectomy is a safe procedure in children and is effective in the prevention of media opacity. Primary posterior capsulectomy may be performed with a vitrectomy probe via the limbus, pars plicata, or pars plana; another method is to perform a PCCC with

forceps. Following posterior capsulectomy, the posterior chamber IOL may be implanted into the capsular bag or ciliary sulcus. In either case, capture of the optic into the posterior capsular opening may be performed in hopes of providing better lens centration or further reduction of the risk of media opacity.

Despite removal of the posterior capsule, numerous studies have shown that secondary membranes and media opacity can still develop.^{9,15,25,27,28,30,32,40,41,49,58,61,62} An intact hyaloid face can serve as a scaffold for migration of LECs and their subsequent proliferation and transformation.^{30,49,62,63} This phenomenon is further enhanced in children by the more intense inflammatory response following surgery.⁶⁴ O'Keefe et al.²⁵ reported media opacity in 33.3% of cases following primary posterior capsulectomy when anterior vitrectomy was not performed. Metge et al.⁶⁵ also showed that primary posterior capsulectomy alone cannot prevent formation of secondary membranes. Cassidy et al.²⁸ reported on three patients in whom posterior capsulorhexis had been performed; in all patients, PCO and anterior hyaloid opacification occurred, and two of them eventually required surgical membranectomy due to ineffective Nd:YAG laser treatment. Raina et al.⁶⁶ reported a 44.4% incidence of significant media opacity in 18 eyes of children 1.5 to 12 years of age in whom posterior capsulorhexis without anterior vitrectomy and in-the-bag implantation of a polymethylmethacrylate (PMMA) lens had been performed.

It seems that removal of the posterior capsule by itself is not sufficient to prevent secondary opacification of the visual axis. One alternative proposed for this problem is posterior capsulorhexis with optic capture without anterior vitrectomy. It has been hypothesized that obliteration of the capsular bag and the posterior location of the IOL optic prevent LEC migration along the vitreous face. Gimbel⁶⁷ performed posterior capsulorhexis with optic capture without anterior vitrectomy in 16 eyes of patients 2.5 to 12 years of age and observed no cases of media opacity by 35.5 months. In this study, heparin surface-modified (HSM) PMMA lenses were used. The same author reported use of this technique in a 2.5-year-old patient, which resulted in a clear media up to 5 months after surgery.⁶⁸ Raina et al.⁶⁶ also reported no media opacity in 16 eyes of children 1.5 to 12 years of age for up to 17.5 months with the same technique. Müllner-Eidenböck et al.⁴⁸ performed the same technique but using hydrophobic IOLs in seven patients older than 6 years and observed no media opacity by 20.7 months. Argento et al.⁴⁹ also implanted hydrophobic acrylic IOLs using the same technique and observed no secondary opacity in 8 eyes of children 2 to 8 years old by 18 to 34 months.

Despite the favorable results described above, a number of investigators have reported conflicting results. In a study by Vasavada and Desai,³⁰ posterior capsulorhexis and optic capture were performed in children

<5 years old, and subsequently 62.5% underwent pars plana membranectomy. In the same study, the incidence of secondary opacification in eyes that underwent concomitant anterior vitrectomy was only 10%, and none required additional surgery. Vasavada et al.⁶¹ compared posterior capsulectomy and optic capture with and without vitrectomy in 41 eyes of 25 patients aged 5 to 12 years. After a mean follow-up of 21 months, 70% showed media opacity of a reticular pattern on the vitreous if vitrectomy was not performed along with primary capsulectomy and optic capture. In contrast, all eyes in the primary capsulectomy, vitrectomy, and optic capture maintained clear visual axis. Koch and Kohlen³⁴ reported visual axis opacity by 6 months in 4 of 5 eyes after posterior capsulorhexis and optic capture without vitrectomy. Vasavada and Trivedi⁶⁹ evaluated the role of optic capture in eyes having cataract surgery at average age of 26 months. All eyes received primary posterior capsulectomy and vitrectomy; however, performance of optic capture was assigned randomly. One eye in the optic capture group developed a membrane in front of the IOL that required a secondary procedure.

Based on the above studies, posterior capsulorhexis and optic capture without vitrectomy may not eliminate secondary opacification, especially in patients <5 years old. Further studies with acrylic IOLs may improve the results with this technique.

Another option is bag-in-the-lens implantation (see Chapter 27). The technique was performed in 54 eyes of 37 children (aged 4–68 months). After a mean follow-up of 17.5 months, there was no reopacification of the visual axis in 93.8% of eyes and 100% of children older than 1 year of age.⁷⁰ The procedure is more technically demanding than is standard IOL implantation and is especially challenging to use in very young children. Reporting the outcomes of a bag-in-the-lens IOL implantation in 34 eyes of 22 children, Tassignon et al.⁷⁰ noted that the IOL could not be properly implanted in 3 eyes, necessitating a secondary intervention.

Another alternative to overcome the problem of secondary opacification following removal of the posterior capsule is anterior vitrectomy, which may be performed via the limbus after posterior capsulectomy or trans pars plana following IOL implantation.^{10,30,43,60,68} Buckley et al.⁵⁴ reported no case of secondary opacification in 20 cases of lensectomy, posterior capsulectomy, and anterior vitrectomy. Awner et al.²⁶ also reported no opacification in 21 patients <4 years of age who underwent the same procedure together with posterior chamber IOL implantation. Vasavada et al.⁶¹ observed no media opacification after performing optic capture in addition to the above-mentioned procedure in 21 eyes of children 5 to 12 years old. Vasavada and Desai³⁰ reported no media opacification in 10 eyes undergoing anterior vitrectomy in addition to primary posterior capsulectomy. Green et al.⁵⁸

performed pars plana lensectomy and vitrectomy in 52 eyes of patients 2 weeks to 4.5 years of age and observed secondary membranes in 6 eyes, all of which had an axial length of <17.4 mm and a corneal diameter of <9.5 mm. Koch and Kohner³⁴ performed lensectomy and anterior vitrectomy with or without optic capture in 6 eyes of children aged 1.5 to 2 years with no secondary opacification. Chrousos et al.⁷¹ reported no opacification after lensectomy, posterior capsulectomy, and vitrectomy in 54 eyes of children 3 months to 17 years of age. Vasavada and Trivedi⁶⁹ performed lensectomy, posterior capsulectomy, and anterior vitrectomy in 40 eyes of patients 4 to 55 months of age; optic capture was performed in 14 cases. After a mean follow-up of 16.5 months, only one case of secondary membrane formation was noted anterior to the IOL optic. In a report by Zubcov et al.⁴⁰ on 12 eyes in patients 3 to 12 years of age, the incidence of clinically significant opacity was 25% by 17 months. Ahmadiéh et al.⁶⁰ reported four cases of opacification in 38 eyes of pediatric patients (mean age, 6.3 years) who had undergone limbal or pars plana operation with IOL implantation; however, only one was significant. Basti et al.⁴³ compared cataract extraction and posterior chamber IOL implantation with (82 eyes) and without (87 eyes) anterior vitrectomy in children 2 to 8 years of age; secondary opacification occurred in 3.6% and 43.7% of the eyes, respectively, by 11 months.

Considering these results, primary surgical removal of the posterior capsule should now be considered the gold standard in pediatric cataract surgery, especially under age 7, when the risk of PCO is higher and the potential for amblyopia still exists. Our routine for pediatric cataract surgery is primary posterior capsulectomy together with anterior vitrectomy up to the age of 9 years.⁶⁰ Furthermore, we perform and recommend the same procedure under certain circumstances in older patients; factors such as type of cataract, overall ocular condition, and socioeconomic status may influence such a decision. For instance, in a 14-year-old patient with traumatic cataract and an open anterior capsule with no possibility for in-the-bag IOL implantation, the risk for PCO is high enough to warrant posterior capsulectomy and anterior vitrectomy. A family with suboptimal resources and a low income who has a long distance to travel may not be compliant with appropriate follow-up visits; in such a case, it is best to remove the posterior capsule and perform anterior vitrectomy rather than accept the risk of PCO and amblyopia.

PREDISPOSING FACTORS FOR RECURRENT OPACIFICATION

Despite posterior capsulectomy and anterior vitrectomy, secondary opacification may occur in the visual axis. Predisposing factors for recurrent opacification include the following: small-sized capsulectomy (<4 mm), young age,

traumatic or uveitic type of cataract, IOL in the ciliary sulcus instead of in the capsular bag, and the presence of associated ocular anomalies or systemic inflammatory conditions.

Age at Surgery

The younger the age at surgery, the more intense the postoperative inflammation and risk for subsequent opacification. Morgan and Karcioğlu¹⁵ reported significant opacification in 3 of 4 eyes in infants 2 weeks to 2 months of age despite a capsulectomy size of at least 5 mm and anterior vitrectomy. Alexandrakis et al.¹¹ reported 7 cases of opacification among 66 eyes of 61 children (median age, 21 months); 6 of these cases occurred in children <6 months old. Peterseim and Wilson³⁹ also observed media opacification to occur much more frequently in children <2 months old compared to older children (4 of 8 cases versus 1 of 24). Lambert et al.⁷² observed a significant correlation between age at surgery and rate of complications. In a study by Hosal and Biglan,³⁶ relative risk for development of opacification was 4.7 times higher in children <1 year of age compared to older children. The rate of media opacification necessitating a secondary surgery can be as high as 23.6% in children <1 year in spite of primary posterior capsulectomy, vitrectomy, and in-the-bag hydrophobic acrylic IOL implantation.⁷³

Type of Cataract

Some investigators have reported the results for congenital, developmental, and traumatic cataracts to be different. Gimbel et al.⁴² calculated the cumulative incidence of need for capsulotomy to be 41% for congenital cataracts and 66% for traumatic cataracts over 2 years. Gupta et al.⁷⁴ operated on 22 eyes with traumatic cataracts in children 3 to 11 years old. A posterior chamber IOL was implanted in 18 eyes, however, whether sulcus or bag fixation was used was not stated. After 6 to 15 months, the most common postoperative complications included fibrinous uveitis, in 81.8% and synechia, in 54.4%. Kora et al.⁴⁴ compared the results of cataract surgery and IOL implantation among congenital, developmental, and traumatic cataracts. PCO was noted to occur more commonly and earlier in congenital cases than in the other two categories. However, patients were not age-matched, which may explain the higher incidence of PCO in congenital cases. Ahmadiéh et al.⁶⁰ also reported no difference between traumatic and developmental cataracts. The apparently increased incidence of PCO in traumatic cataracts may be due to lower possibility of in-the-bag IOL implantation and more severe inflammation following trauma.

Associated Ocular and Systemic Conditions

Anomalies such as PFV or microcornea; ocular conditions such as rubella syndrome, toxocariasis, toxoplasmosis, and

pars planitis; and systemic disease such as juvenile rheumatoid arthritis are associated with a higher incidence of complications and capsular opacity.^{9,23,45,58,62,72} Green et al.⁵⁸ performed pars plana lensectomy and anterior vitrectomy in 52 cases and observed secondary opacification in 6 eyes, all of which had an axial length of <17.4 mm and a corneal diameter of <9.5 mm. According to Lambert et al.,⁷² of eight complications, four were related to eyes with PFV. BenEzra and Cohen⁴⁵ observed retrolental membranes in 80% of eyes with juvenile rheumatoid arthritis despite posterior capsulectomy and vitrectomy, all of which required a second surgical intervention. Lundvall and Zetterstrom⁷⁵ reported on cataract surgery with IOL implantation in 10 eyes with juvenile rheumatoid arthritis; in 5 eyes, anterior vitrectomy was also performed. After a mean follow-up of 28 months, media opacification occurred in 7 eyes (70%).

Capsulectomy Size

Chrousos et al.⁷¹ reported opacification in 12% of cases in which the posterior capsule was minimally opened; however, when the capsulectomy size was adequate, no instance of opacification was reported. All cases had undergone vitrectomy; however, the authors failed to define what size capsulectomy was considered adequate. Ahmadieh et al.⁶⁰ reported four cases of opacity, all in which the capsulectomy size was <3 mm; however, only 1 eye required surgical intervention.

Sulcus Versus Bag Fixation

In 1985, Apple et al.⁷⁶ described the advantages of capsular bag fixation in adults, including less risk of pupillary capture, reduced decentration, less trauma to pigmented uveal tissue, decreased disruption of the blood–aqueous barrier, and less inflammation. Pandey et al.³¹ operated on 20 eyes with traumatic cataracts. Lens aspiration was performed, followed by IOL implantation either in the capsular bag or in the ciliary sulcus. Complications including postoperative uveitis, PCO, pupillary capture, and lens decentration occurred more frequently in the sulcus fixation group. However, Jensen et al.³⁷ found no significant difference in the rate of PCO in relation to IOL position.

INFLAMMATORY MEMBRANES

Another cause of media opacification after cataract surgery is formation of inflammatory membranes in the pupil, anterior and/or posterior to the IOL surface, together with synechia formation and pigment deposition. Iris manipulation, blood–aqueous barrier disruption, reaction to IOL material, younger age at surgery, and associated ocular or systemic inflammation predispose to greater postoperative inflammation. Fibrinous uveitis

following pediatric cataract surgery has been reported in some series as high as 57.5%²³ to 81.8%³¹ of cases.

These membranes may be transient, with varying degrees of resorption in several days or a few weeks. Some cases may persist, necessitating a second surgical intervention. Due to the more intense inflammatory reaction in children and the potential for amblyopia, this issue is of greater concern in children. Attempts may be directed toward prevention or immediate medical or surgical treatment once the membrane has developed.

The risk of inflammatory membranes may be decreased by frequent use of topical and systemic steroids before and following cataract surgery. In cases of uveitic cataracts, preexisting intraocular inflammation must be controlled with topical and systemic medications prior to surgery.⁷⁵ Postoperatively, high-dose topical and, if needed, systemic steroids must be continued and slowly tapered off. Intraoperative use of low molecular weight heparin in irrigation fluids may decrease the severity of postoperative inflammation.^{12,77} HSM-PMMA IOLs may also be useful in reducing inflammatory membranes.^{67,68,78} However, recent studies have shown no significant difference between PMMA and HSM-PMMA IOLs in quiescent uveitis.^{79,80} HSM-PMMA IOLs are no longer easily available. Hydrophobic acrylic IOLs may reduce LEC migration,⁸¹ PCO, and giant cell deposits.⁸² In a study by Gatinel et al.,⁷⁹ no significant difference was observed between HSM-PMMA and hydrophobic acrylic IOLs in diabetic patients. It is noteworthy that the above-mentioned studies were performed in adults, and generalization of the results to pediatric cases is uncertain.

Once inflammatory membranes have developed, topical and systemic steroids may be necessary for improvement. Tissue plasminogen activator (TPA), first used in 1988,⁸³ has recently been used for dissolution of fibrin clots after pediatric cataract surgery. TPA is normally produced by vascular endothelial cells, corneal epithelium and endothelium, and the trabecular meshwork.⁸⁴ Significant reduction of TPA activity has been noted after cataract surgery, which, in combination with blood–aqueous barrier disruption, leads to fibrin deposition.⁸⁵ Recombinant TPA is a fibrinolytic serine protease produced genetically by cloning and expression of recombinant DNA.⁸⁶ Plasminogen activator causes fibrin lysis by localized transformation of plasminogen to plasmin in the presence of fibrin. The half-life of the intravenous TPA is 7 minutes.⁸⁷ No systemic effects have been noted with injection into the anterior chamber.⁸⁸ Recombinant TPA has no deleterious effect on corneal endothelium⁸⁸ and has been used for dissolution of fibrin clots after penetrating keratoplasty,⁸⁷ vitrectomy, and glaucoma surgery.⁸⁹ Mean time for effect is 3.3 hours. Complications of intraocular TPA are hyphema and increased IOP; therefore, its use is not recommended earlier than 3 days after surgery.⁹⁰

The optimal dosing for TPA remains unsettled. Klais et al.⁹¹ used 10 µg/100 µL TPA in the anterior chamber of 11 eyes of children aged 3 to 13 years who developed a severe fibrinous reaction unresponsive to conventional treatment. Dissolution of the fibrin and resolution of synechia were observed in 9 eyes by 6 days; no complications or recurrence of fibrin deposition was observed. Mehta and Adams⁹² reported a 25-µg dose without any complications. TPA seems to be effective only up to 3 weeks after cataract surgery.⁹³

Dense fibrin membranes may require surgical intervention. In the pupillary space, the membrane may be retracted with a sharp needle⁹⁴ or by Nd:YAG laser application in cooperative cases; however, dense membranes anterior or posterior to the IOL require membranectomy.

OTHER CAUSES OF MEDIA OPACITY

Vitreous condensation and opacification may follow excessive iris manipulation, inadequate vitrectomy, vitreous incarceration into the surgical wound, and admixture of lens material with vitreous. To prevent the latter complication, the surgeon should try to avoid opening the posterior capsule prior to complete removal of lens material. In cases of uveitic cataracts such as juvenile rheumatoid arthritis, pars planitis, and rubella syndrome, complete anterior vitrectomy is recommended to clear organized vitreous and to remove any remaining strands capable of causing secondary opacities. In some instances, hemorrhage may occur when performing peripheral iridectomy. It is best to avoid excessive stretch on the iris root to prevent hemorrhage and its seepage into the vitreous cavity. Vitreous hemorrhage after cataract surgery may also occur in cases with PFV; use of intraocular cautery may be useful in preventing this complication.⁹

Corneal edema is another cause of media opacification, which may be caused by irrigation fluids, toxic substances, excessive intraocular manipulations, IOL touch during implantation, or late corneal touch. In cases with microcornea, there is a higher risk of corneal edema with limbal approaches. Hiles²⁴ reported the overall incidence of corneal edema to be 4% in congenital and traumatic cataracts with posterior chamber, anterior chamber, and iris-fixated IOLs.

Perhaps the most serious condition that can lead to media opacity is glaucoma. This is a well-known complication of pediatric cataract surgery that has been reported by many investigators.^{33,71,77,95,96} There seem to be multiple mechanisms involved including surgical trauma, inflammation, obliteration of angle structures by synechia, and predisposing conditions. Media opacification caused by glaucoma is an end-stage condition indicating a poor visual prognosis.

PROCEDURES PREVENTING MEDIA OPACITY

Primary Posterior Capsulectomy with or without Anterior Vitrectomy

See Chapter 20.

Sealed-Capsule Irrigation

Because PCO is predominantly caused by residual LECs in the capsular bag after cataract surgery,^{2,20,97} several surgical techniques have been attempted for the removal of these LECs at the time of lens extraction. These techniques include aspiration of the anterior capsule using an extensive irrigation/aspiration system during cataract surgery,^{98,99} manual polishing of the anterior and/or posterior capsule,^{99,100} as well as pharmacologic dispersion and aspiration of the anterior capsule.^{101–104} Maloof et al.¹⁰⁵ designed a sealed-capsule irrigation device that can help in selectively irrigating the capsular bag.¹⁰⁶ The device is a sterile single-use ophthalmic instrument to perform sealed irrigation of the capsular bag during cataract surgery; this prevents the irrigation fluid from coming into contact with the other ocular structures.

In studies in rabbit eyes, 5-fluorouracil (5-FU) was effective in preventing PCO, whereas the use of distilled water did not prevent PCO.¹⁰⁷ Similarly, in human eyes, distilled water alone, however, did not significantly reduce PCO.¹⁰⁸

Other pharmacologic agents such as thermosetting plastic prepared with phenol formaldehyde resins (Catalin), methotrexate, mitomycin, and daunomycin have been shown to be effective in preventing PCO in vitro,^{20,109} but in vivo studies have shown their toxicity to corneal endothelial cells, iris, ciliary body epithelial cells, and retina.¹¹⁰ These studies showed promise for finding medical treatment of PCO by targeting the survival, adhesion, proliferation, migration, and transdifferentiation of residual LECs, but the risk of their toxic effects on surrounding intraocular tissues has restricted their clinical use. A recent study by Kim et al.¹¹¹ showed that in rabbit eyes, mitomycin is more effective than distilled water for reducing PCO and that the sealed capsule irrigation device protected the surrounding tissue from mitomycin toxicity.

Other Preventive Measures

Hydrodissection: see Chapter 18

One study showed that ultrasonic vacuuming during cataract surgery reduced the number of patients requiring laser capsulotomy.⁹⁸ Another study showed that capsule vacuuming reduced but did not eliminate PCO.¹¹² Khalifa¹⁰⁰ determined that vacuuming the posterior

capsule had no effect on the long-term development of PCO. Therefore, vacuuming or polishing the capsule may delay the onset of PCO, but the long-term benefit is limited because PCO is mainly caused by germinative LECs in the equatorial region rather than the displaced metaplastic LECs already on the posterior capsule.¹¹³ Also, equatorial capsule vacuuming has been found to be associated with additional surgery time and trauma and risk of capsule tears, damaging the capsular support of the IOL implant.¹¹⁴ Hara et al.¹¹⁵ reported the advantages of a closed endocapsular ring to prevent PCO. This approach was shown to be promising in PCO prevention in the eyes of adults and children.¹¹⁶

IOL Materials and Designs

There are two main types of materials used for IOL manufacturing: acrylic and silicone. Acrylic lenses are rigid (made from PMMA) or foldable (made from hydrophobic or hydrophilic materials).¹¹⁷ Although it is well recognized that a hydrophilic acrylic material is more biocompatible,¹¹⁸ IOLs made of this material have been shown to support LEC adhesion, migration, and proliferation and thus PCO development¹¹⁹ compared with an IOL made of PMMA or hydrophobic acrylic materials.¹²⁰ Surface modifications of PMMA IOLs by carbon and titanium,¹²¹ heparin,¹²² and polytetrafluoroethylene¹²³ and of silicon IOLs by oxygen and carbon dioxide plasma¹²⁴ or a sulfonate and carboxylate group containing polymer¹²⁵ have been reported to have higher biocompatibility and effectiveness in prevention of PCO. Recently, IOL surface modification by gas plasma¹²⁶ and polyethylene glycol¹²⁷ has been shown to influence LEC behavior and to prevent PCO.

There is strong evidence that a truncated edge is the key factor for the prevention of PCO, regardless of the IOL material. A higher PCO inhibitor effect has been observed with IOLs that provide a mechanical barrier effect on the posterior lens capsule.^{2,20} Nishi et al.¹²⁸ demonstrated that the sharp-edge optic IOL and the formation of a capsular bend are highly effective in reducing PCO. Both the sharpness of the bend and the duration of its formation are important factors. The differences in speed may be influenced not only by reflex differences in design but also by the IOL material.

SUMMARY

The main goal of cataract surgery in children is relief of media opacity and proper correction of refractive errors with concomitant amblyopia therapy. All these depend on maintenance of clear ocular media. Primary posterior capsulectomy and vitrectomy may help prevent or delay PCO.

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Postoperative Glaucoma

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Postoperative glaucoma after childhood cataract surgery remains a major concern despite all of the improvements in technology and the increased use of intraocular lenses (IOLs). To date, scientists cannot submit an etiology that is accepted by all clinicians, clinicians cannot offer a treatment that is effective for all patients, and the onset of glaucoma can range from the immediate postoperative period to many years later. To make matters worse, children do not always cooperate for intraocular pressure (IOP) measurements, optic disc evaluation, or visual field documentations. Thus, defining when glaucoma is present and when a treatment is effective at halting the progression of damage is unusually challenging. The common factor for all of these patients is the fact that cataract surgery has been performed. Even with severe microphthalmia, glaucoma does not seem to occur in the absence of the cataract procedure. In the predisposed eye, surgery to remove the cataract may trigger a cascade of events that can lead to elevated IOP and/or glaucoma early on or even 5 to 15 years later. Ophthalmologists must be vigilant about assessing for postoperative glaucoma even after uncomplicated cataract removal in young children.

How often does late postoperative glaucoma occur? Why does it occur? Which patients are more likely to develop glaucoma after surgery? What therapy is appropriate once this is recognized? These questions are discussed in this chapter.

ESTIMATE OF FREQUENCY OF POSTCATARACT GLAUCOMA IN CHILDREN

The incidence of glaucoma following pediatric cataract removal has been reported to be as low as 5% and as high as 41%.¹⁻⁶ In a review of 13 studies from the international literature on congenital cataract surgery in an earlier era, Francois³ reported that delayed glaucoma following cataract surgery in children occurred from 0% to 14% of cases. The majority of these data are available in reports from the 1940s or 1950s, when the linear aspiration tech-

nique was used. This older method resulted in lens swelling and, at times, a prolonged flat anterior chamber. Acute angle-closure glaucoma and excessive inflammation often ensued. Scheie⁷ introduced an aspiration technique in 1960 that may have reduced the incidence of complications compared to the linear extraction method. In 1984, Chrousos et al.² reported their 15-year experience with pediatric cataract surgery; the surgeons utilized Scheie's manual aspiration technique as well as automated styles of cataract removal. The standard needle-and-syringe technique was performed in 304 eyes, rotoextraction with a small opening in the posterior capsule was performed in 34 eyes, and Ocutome vitrector aspiration with wide posterior capsular excision was performed in 54 eyes. All patients had at least 3 months of follow-up. Overall, chronic glaucoma was found in 6.1% of the eyes; no patients with Ocutome aspiration developed glaucoma. Interestingly, the 6.1% overall rate of glaucoma reported by Chrousos et al.² was similar to the 5% rate achieved by Francois³ before aspiration techniques were used. The authors of the 1984 study admitted that a limitation of the study is the length of follow-up of some patients, which could have resulted in underreporting glaucoma. While the aspiration-only group was followed for a mean of 6.3 years, the Ocutome group was followed for only 2 years on average.

Making comparison between studies is rather unreasonable for many reasons, including the variety of definitions of glaucoma that authors have used through the ages and the variety of instruments used to measure IOP. However, perhaps the most important factor in studies aiming to report the frequency of glaucoma in a postoperative population is the length of follow-up, as a study by Simon and associates addresses below.

Simon et al.⁵ invited their patients who had undergone pediatric cataract surgery back to their office to check for asymptomatic glaucoma, which was defined as an IOP ≥ 26 . While almost a quarter of the patients examined had glaucoma, the interesting data involved length of follow-up. While only 1 of 14 eyes (7%) with ≤ 60 months of

follow-up since lensectomy developed glaucoma, 7 of 17 eyes (41%) followed for >5 years had glaucoma. This suggests that reports with <5 years of follow-up may significantly underestimate the frequency with which glaucoma develops after pediatric cataract surgery. Chrousos et al.² and Simon et al.⁵ both pointed out an important finding in asymptomatic postoperative glaucoma—that although the onset is typically delayed until years after the surgery, the diagnosis may be made within the first year of surgery in some cases and surveillance of IOP must be initiated early.

Simon et al.⁵ did not choose objective signs of glaucoma such as visual field loss or optic nerve head damage in their definition of glaucoma. Therefore, it is likely that some of his patients actually had ocular hypertension (OHTN), and “true” glaucoma probably occurred less frequently than was reported. In adults, the Ocular Hypertension Treatment Study (OHTS)⁸ recognized that only a small percentage of patients with an IOP >21 mm Hg will progress to develop glaucoma after several years.

A prospective, nonrandomized study provides some valuable data on the rates of pediatric glaucoma and OHTN after automated lensectomy and vitrectomy. Egbert et al.⁹ reviewed records of patients with 5 years of follow-up after lensectomy. Of 159 patients, 52 were excluded because of trauma, microphthalmos, uveitis, or a similar complicating factor. Sixty-two of 107 patients (58%) participated, and participating and nonparticipating patients had no differences in average corneal diameter or age at the time of surgery. Glaucoma was defined as an IOP >21 with a cup/disk ratio >0.5 or a cup/disk asymmetry ≥ 0.2 . OHTN was defined as an IOP >21 without the aforementioned optic nerve parameters. Six of 40 (15%) patients had glaucoma in their aphakic eye. OHTN was even more common. Thirteen of forty (32.5%) patients had OHTN in their unilaterally aphakic eye, while 10 of 22 (45%) patients who previously had bilateral cataracts had OHTN in one (four patients) or both (six patients) eyes. Overall, 23 of 62 (37%) patients had aphakic OHTN when examined ≥ 5 years after surgery. This report demonstrates that OHTN is a common long-term finding in patients with pediatric aphakia and glaucoma is less common but not rare event. With time, more patients with a normal IOP or with OHTN may develop glaucoma.

In an additional study by Egbert et al.,¹⁰ OHTN and glaucoma rates were examined for patients 5 and 10 years after undergoing automated lensectomy and vitrectomy. Their data showed a significant trend of glaucoma development with additional years after surgery. They showed a 23% conversion rate from OHTN to glaucoma, with similar definitions for the respective findings as their earlier paper. This occurred during a mean observational period of 7.2 years. Also shown were the rates of glaucoma in the 63-patient cohort at 5 (7/63; 11%)

and 10 years (5/59 remaining; 9%) after surgery. These findings suggest that the risk of aphakic patients developing glaucoma does not dramatically change in the first 10 years postoperatively, demonstrating the necessity of continued follow-up. So far, evidence has not been offered to suggest that the risk of glaucoma in aphakic eyes ever subsides; thus follow-up should be pursued indefinitely. Swamy et al.¹¹ also noted the lifelong risk of developing secondary glaucoma in their 2007 review of 423 eyes over 20 years that received cataract removal with or without primary IOL. Their data demonstrated a range of time to glaucoma onset postoperatively of 2 weeks to 16.8 years. These two studies by Egbert and one by Swamy demonstrate the necessity of continual follow-up with patients after surgery.

We believe that it is vital to examine patients on at least a yearly basis after uneventful pediatric cataract surgery with or without IOL implantation. Egbert et al.⁹ concluded that good data can be obtained from patients >5 years of age in the clinic. We schedule examinations under anesthesia (EUA) whenever clinic examinations with IOP measurements, refractions, serial axial length measurements, and optic nerve evaluations cannot be done awake in the clinic. Baseline optic disc documentation should be performed (Fig. 51.1). Early in childhood, EUA is often done yearly, but by age 5 to 6, the examination in the office is more reliable (except in those children with

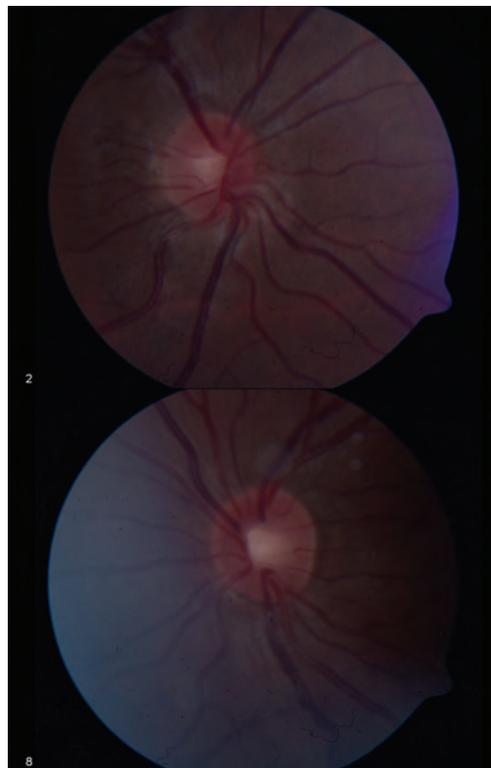


Figure 51.1. Disc photo of 11-year-old pseudophakic child diagnosed as glaucoma suspect due to high IOP.

developmental delay). As discussed below, the Icare (Icare USA, Raleigh, NC) rebound tonometer has reduced the need for EUA in some children due to its ease of use and the fact that no topical anesthetic needs to be instilled.

EXAMINATION UNDER ANESTHESIA

When evaluating under anesthesia, the method of anesthetizing must be considered due to the interaction between anesthetic agents and IOP. In the literature, there are many resources describing the effect of anesthesia methods on IOP. In their book series, *The Glaucomas*, Sampaolesi et al.¹² provided an effective overview of the impact of different medications potentially used during an EUA. They reviewed the effects of the more historical agent, ether, with its increase in IOP from the Valsalva-like response, and then discussed differences among more modern agents including barbiturates, fluorinated inhalation agents, etc. Though all general anesthetic agents affect IOP, Sampaolesi and associates point out that the biometric measurement of axial length is not affected by IOP fluctuations. This allows axial length to be assessed as another interpretable finding independent of anesthesia for evaluating glaucoma beyond just an IOP measurement. As outlined by Sampaolesi and associates, barbiturate dosage is difficult to accomplish accurately. Too little and the patient begins to regain consciousness and struggle, raising IOP, and too much will decrease IOP due to blood pressure decrease from respiratory depression. Conversely, if succinylcholine is used to paralyze respiratory movements, the blood pressure spike facilitates an increased IOP. The inhalation agents halothane (Fluothane), methoxyflurane (Penthrane), and sevoflurane (Sevorane) categorically lower IOP. However, halothane and methoxyflurane represent older generation agents largely eclipsed by sevoflurane in modern settings. In addition to practical advantages such as being well tolerated and having a quick recovery, sevoflurane maintains the effect of its predecessor halothane in maintaining the eyes in a straight ahead position while deeply anesthetized, helping facilitate accurate assessment of clinical parameters during an EUA. In a setting with controlled ventilation and normocapnia, sevoflurane causes a proportional decrease in IOP compared to depth of anesthesia. A proportional relationship offers the ability to interpret the IOP figures for a nonanesthetized state when evaluating for glaucoma.

Nagdeve et al. demonstrated that ketamine in a low dose (3 mg/kg) does not affect IOP. This was shown to contrast with an induction dose of ketamine, which raised IOP after administration.¹³ Additionally, Jones et al.¹⁴ demonstrated the effect of sequential treatment of pediatric patients with ketamine and sevoflurane. They showed that IOP measurements while anesthesia was maintained using sevoflurane were significantly lower than measure-

ments taken after ketamine was administered. This effect of sevoflurane remains consistent with the effect of an earlier inhalation agent, halothane. While the Jones study conceded their small, retrospective sample size (16 eyes from 8 patients) was a limitation, components from their study are demonstrated independently elsewhere. These studies taken together offer the opportunity to use a non-induction dose of a dissociative anesthetic, ketamine, for collecting accurate IOP measurements, with sevoflurane utilized for the balance of the EUA, offering its many advantages over ketamine for anesthesia.

Recently, a new instrument has become available for the evaluation of IOP, the Icare rebound tonometer (Icare TAO1i) (Icare USA, Raleigh, NC). This instrument allows for IOP evaluation without the need for a topical anesthetic. In a study by Lundvall et al.¹⁵ in 2011, 46 infants were recruited into this evaluation of the Icare tonometer while awake in the clinic. Only seven refused to cooperate. The remaining 39 were evaluated successfully. This new technology allows for the possibility of avoiding the operating room for EUAs, if the only indication is an inability to acquire an accurate IOP measurement. Even if the operating room cannot be avoided for other parts of an EUA, the Icare allows for more frequent evaluation of IOP outside the operating room in the infant population. Until recently, the Icare was limited by its mechanism requiring the patient to be upright to allow for proper measurement; however, the latest model (Icare Pro) allows for supine patients, providing additional flexibility.

Central Corneal Thickness

Central corneal thickness (CCT) has been more recently studied with respect to pediatric cataract surgery patients. Goldmann applanation tonometry has been the gold standard for IOP measurement, but variability in CCT affects applanation tonometry measurement. Simsek et al.¹⁶ published a study in 2006 outlining the involvement of CCT in pediatric cataract surgery. They noted CCT of aphakic and pseudophakic patients to be significantly higher than age-matched controls. Muir et al.¹⁷ compared the CCT of normal children (controls) and in those with cataract and those operated for cataract. In the absence of factors known to affect CCT (Down syndrome, Marfan syndrome, and aniridia), CCT is similar in eyes with pediatric cataracts and normal control and increases after cataract surgery. Further data from Muir lend corroboration to this theory. Unoperated unilateral cataracts ($n = 9$) in their study were shown to correlate strongly ($r^2 = 0.7$) with a nonzero slope ($P = 0.005$) to the CCT of the fellow noncataractous eye. Time after surgery is shown to be a positive predictor for CCT. The CCT of aphakic eyes without glaucoma in this study were assessed against postoperative years. The slope offered a statistically nonzero P -value (<0.001) and a positive correlation

between increasing CCT and years after lensectomy ($r^2 = 0.6$). The data analyzed for these results even included developmental, persistent fetal vasculature (PFV), and microphthalmic cataracts because their CCT values were not significantly different from the congenital cataract CCT cohort. A difference was noticed in the comparison of aphakic eyes with glaucoma and those aphakic eyes without glaucoma, although the authors postulated that perhaps there was an unintentional selection bias in those eyes because of a predisposition for thicker corneas and glaucoma or that the glaucoma contributed to increased CCT via subclinical edema.

In addition to the data comparisons, Muir et al.¹⁷ noted a potential disparity between the findings of CCT in postoperative cataract children and the CCT findings detailed in the OHTS. OHTS work yielded a model describing increasing CCT as a protective measure against glaucoma development. Conversely, this study demonstrated that CCT among aphakic eyes with glaucoma was further increased beyond the typical nonglaucoma postoperative increase in CCT. Muir pointed out the potential inconsistency in applying the OHTS results from adults to aphakic children. If increasing CCT protects against glaucoma, why does a significant minority of patients develop glaucoma after surgery to remove a cataractous lens? These findings highlight a potential dichotomy in the interpretation of CCT changes in aphakic pediatric eyes compared with natural adult eyes. Endothelial corneal damage or an inherent response to aphakia/pseudophakia was offered as potential explanations of this finding of increased CCT postoperatively. Further, for those patients developing glaucoma postoperatively, it was “tempting” for the authors to interpret the additional CCT increase as the result of increased IOP, leading to added corneal endothelial injury and subclinical stromal edema.

PROPOSED MECHANISMS FOR THE DEVELOPMENT OF GLAUCOMA AFTER PEDIATRIC CATARACT SURGERY

The causes of pediatric aphakic open-angle glaucoma are as elusive today as they were years ago. In 1977, Phelps and Arafat¹⁸ brought this to the attention of ophthalmologists as a “warning.” They were surprised to diagnose an insidious, asymptomatic type of glaucoma in 18 patients who had undergone congenital cataract removal years (6–56 years) before the patients manifested an elevated IOP. They expressed concern that although those patients had cataracts removed by simple needling, linear extraction, or intracapsular extraction, more modern techniques (such as needling and aspiration, phacoemulsification, and rotoextraction) may also produce glaucoma.

The authors discussed several of the mechanisms that could be at fault. Could an undescribed ocular syndrome involving both cataract and glaucoma be causal? Does early surgery promote such significant inflammation or expose the fledgling trabeculum to so much lens protein that it is irrevocably damaged? Is a vitreous component toxic to the trabeculum? Even today we do not know the answer to these questions or how much these factors contribute to the development of glaucoma, if at all.

Late-onset open-angle glaucoma occurs much more commonly than angle closure after pediatric cataract surgery.^{1,4-6} Of the 11 eyes with glaucoma for which gonioscopic data were available, all had open angles.² Open-angle glaucoma can occur without the presence of any symptoms or gross changes in the appearance of the eye and has been described both in normal-appearing open angles and in angles that have undergone change. Acute angle closure following pediatric cataract surgery with modern techniques is relatively uncommon, and peripheral iridectomies are performed less commonly today than in years past.^{1,4-6} Kang et al.¹⁹ discussed their findings of a bimodal distribution. Early onset was found to favor angle closure, while late onset was found to occur more frequently in an open-angle fashion. In 1986, Walton²⁰ discussed pupillary block and chronic angle closure from peripheral anterior synechia as the typical mechanism following cataract removal by the “aspiration” mechanism. A decade later, Walton’s American Ophthalmological Society thesis⁶ concluded that the asymptomatic, postoperative glaucoma in aphakic patients was actually an open-angle mechanism. Walton⁶ studied the angle structure of 65 aphakic children with glaucoma. Vitrectomy techniques were utilized in the majority (80%) of cases. Preoperatively, the majority of patients with available gonioscopy (19/29 eyes) had no angle abnormalities, while 10 patients did have “anomalous attachments from the iris root to Schwalbe line and the trabecular meshwork.” Postoperatively, the angles were open in 79 of 80 eyes, but in 76 of 79 (96%) eyes, “circumferential repositioning of the iris insertion anteriorly at the level of the posterior or mid-trabecular meshwork with resultant loss to view of the ciliary body band and scleral spur” occurred. Windows of visible scleral spur or ciliary body were noted in these eyes, confirming open angles. Phelps and Arafat¹⁸ had described similar changes in the anterior chamber angles in their patients. Walton⁶ observed scattered pigment deposits in the exposed anterior trabecular meshwork and, less frequently, white crystalline deposits suggestive of lens protein.

The mechanism does not appear to be related to clinically identifiable *late* postoperative inflammation. Walton⁶ reported on slit-lamp examinations of 19 children performed after lensectomy and before glaucoma was diagnosed. In none of those patients was an anterior chamber cell or band keratopathy present; these patients had a

different appearance than those with chronic inflammation. In addition, none of the patients examined after the diagnosis of glaucoma had evidence of active intraocular inflammation.

The absence of active intraocular inflammation years after surgery does not negate the possibility of acute postoperative inflammation causing significant, immediate damage to the trabecular meshwork that develops into a chronic, secondary open-angle glaucoma. Several authors have reported cases of bilateral cataracts that had subtle, bilateral angle abnormalities but developed glaucoma only in the operated eye.^{6,21} We are curious whether the presence of “anomalous attachments from the iris root to Schwalbe line and the trabecular meshwork” documented in 10 of Walton’s⁶ patients (but absent in 19 patients) before surgery implies that an abnormal process was already occurring subclinically in all eyes. Perhaps cataract surgery simply amplified the process such that “circumferential repositioning of the iris insertion anteriorly” was observed at a later time. Phelps and Arafat¹⁸ observing similar gonioscopic findings in patients after surgery implied that the uniformity of the angle findings “throughout its circumference” instead suggested that these findings were congenital and not related to the cataract surgery. There is no way to prove, however, that those angle findings were not indicative of subclinical dysfunction.

Walton⁶ himself, in the discussion following his study, lamented that despite his careful attention to gonioscopic detail in his patients, the cause of the glaucoma could not be inferred. Whether or not the angle is described as open or closed is probably irrelevant if we cannot correlate microscopic, ultrastructural changes (by light microscopy or electron microscopy) in the trabecular meshwork with changes in aqueous outflow and increases in IOP in eyes of children with and without glaucoma who had cataract surgery in one or both eyes. Such a study may lead to the elucidation of the mechanism of the glaucoma and, ultimately, its true cause.

It is possible that cataract extraction may indeed damage a growing, vulnerable (“immature” or “developmentally arrested”) anterior chamber angle in an eye with a subclinically imperfect trabecular outflow in a way that creates high IOP years later.⁴ This may be why patients with a preexisting ocular abnormality (such as trauma, dislocated lens, chronic uveitis, or anterior segment dysgenesis) may be at higher risk for postoperative glaucoma.⁶

If such a theory were correct, and if surgery were to be performed satisfactorily with modern techniques, the most important step in preventing glaucoma for these patients would be to minimize acute postoperative inflammation by applying various forms of anti-inflammatory preoperative and postoperative medications. The results of a randomized, prospective trial addressing this issue would be most helpful. Until then, we operate on eyes with

significant cataracts using the least traumatic techniques available and hope we are doing more good than harm. Then we treat glaucoma as we detect it.

RISK FACTORS HISTORICALLY ASSOCIATED WITH POSTCATARACT, PEDIATRIC GLAUCOMA

Clinicians can care for their patients better by identifying those who appear to be at risk for developing the disorder. A number of reports have discussed the following risk factors associated with postoperative glaucoma in children with cataracts: microcornea, poorly dilating pupils, surgery at <1 year of age, the presence of other ocular disease (e.g., congenital rubella syndrome), nuclear cataract, PFV, and performance of a posterior capsulorhexis. Much disagreement accompanies these risk factors, and some authors found no association among glaucoma, age at surgery, microphthalmos, and surgical complications.⁵

Age at Surgery

Mills and Robb⁴ reported risk factors for childhood glaucoma: cataract surgery at an age of <1 year (relative risk [RR] = 9.9; $P \leq 0.001$), microcornea (RR = 4.4; $P \leq 0.001$), poor pupillary dilation (RR = 5.2; $P \leq 0.001$), and congenital rubella syndrome (RR = 5.8; $P \leq 0.001$). The RR was notably high for patients undergoing surgery before the age of 6 months (RR = 5.4; $P \leq 0.001$) and 1 year (9.9; $P \leq 0.001$). No patient who had surgery after 1.25 years of age developed chronic open- or closed-angle glaucoma. The authors state,

The time at surgery may not be independent of other pathologic factors... [as] a disproportionate share of those patients who had early cataract surgery had other ocular abnormalities (congenital rubella syndrome (10.1% of 79 eyes operated on before 1 year of age), poorly dilating pupils (22.0%), microcornea (10.1%), or persistent fetal vasculature (6.3%)) ... or more complete lens opacity.

The majority of Walton’s patients (77%) were also operated on at <1 year of age. The author suggests that performing surgery on “small eyes with small corneas and often poorly dilating pupils must be considered a risk factor for the development of glaucoma.” The author implies that cataract surgery is difficult to perform adequately on these eyes and cites residual lens tissue behind the iris in 78% of patients and a prominent need for secondary lens surgery (in 47% of patients) as evidence of technical inadequacy of the surgery in these patients. Walton⁶ argues that the angle abnormalities are the result of cataract surgery—especially early surgery. The immature trabecular meshwork of patients undergoing cataract surgery at a very young age was exposed to inflammation or direct surgical trauma and led to glaucoma.

Magnusson et al.²² prospectively followed a cohort of 137 patients in Sweden for an average of 9 years and concluded that cataract extraction in children <10 days old is associated with double the frequency of glaucoma. Twenty-nine percent (4/14) of patients operated on before the age of 10 days developed glaucoma; operations performed after 10 days of life had half the frequency of glaucoma. A 2004 article by Peter Rabiah²³ has made it more difficult to retain the belief that age at surgery is unrelated to the development of glaucoma. Five hundred seventy eyes of 322 patients who underwent limbal approach surgery without IOL implantation in Saudi Arabia were analyzed. Patients were excluded if follow-up was for <5 years or if ocular trauma, PFV, prior eye surgery, or rubella or Lowe syndrome was present. Microcornea was not an exclusion criterion. Glaucoma was defined as an IOP ≥ 26 mm Hg on two occasions. Potential predictors of risk were entered into a univariate and multivariate model. Glaucoma was diagnosed in 118 of 570 eyes (21%) at a mean age of 5.4 years after surgery (range 2 weeks–15.6 years); average total follow-up for eyes with and without glaucoma was 8.5 and 10.9 years, respectively. The vast majority (86%) of the glaucoma was diagnosed in patients who underwent surgery at or before 9 months of age. Of patients with cataracts in one or both eyes, *no unoperated fellow eye* developed glaucoma. The significant predictors of glaucoma in the multivariable analysis included microcornea, primary posterior capsulotomy/anterior vitrectomy, secondary membrane surgery, and surgery at ≤ 9 months of age. The risk appeared substantially lower in children operated on after 3 years of age. The significantly lower rate of glaucoma with late surgery in these patients lends credence to early age at cataract surgery as an additional, independent risk factor for postsurgical glaucoma.

Chen et al.²⁴ offered similar conclusions in their 2006 article. They retrospectively reviewed pediatric glaucoma patients with previous cataract surgery in their center from 1970 to 2003. This produced a cohort of 368 eyes of 258 patients to analyze for aphakic glaucoma risk factors. Two hundred and sixteen eyes (58.7%) of 150 patients in this group developed aphakic glaucoma. They assessed significant risk factors, and the most prominent ($P < 0.001$) were cataract removal before 1 year of age and postoperative complications. We noted that all eyes in which glaucoma developed had cataract surgery before 4.5 months of age when eyes with known glaucoma risk factors were excluded.²⁵ In addition, all eyes that developed glaucoma had cataract diagnosed within the first month of life. Michaelides et al.²⁶ also noted the significance of age as an aphakic glaucoma risk factor. In their review of 71 eyes with at least 5 years of follow-up, the average age at surgery for the eyes developing glaucoma was 1.6 months compared to 28.7 months old at surgery on average for the eyes not developing aphakic glaucoma.

Among patients who undergo surgery before a year of age (specifically before 10 months of age in this study), Khan and Al-Dahmash²⁷ noted the lowest risk for developing aphakic glaucoma when surgery was performed between 3 and 4 months of age. The authors cautioned against waiting much beyond about 4 to 6 weeks of life to remove infantile cataracts due to the amblyopia risk. Wong et al.²⁸ noted this link to surgical trauma as well, though other age risk factors were not identically noted. In the Wong study, 4 eyes underwent explantation of the IOL during the procedure because the IOL began to dislocate posteriorly. The authors noted the significant iris manipulation necessary to remove the IOL, which they correlated with the observation that all four of these explanted cases developed glaucoma. In 2010, Al-Dahmash and Khan²⁹ demonstrated a greater than double risk factor for eyes operated on before 10 months of age (26.2% versus 11.9%) for developing aphakic glaucoma.

The Infant Aphakia Treatment Study (IATS) 1-year data showed that a patient 1 month younger than another carried a 1.6 times higher risk for a glaucoma-related adverse event.³⁰ It is important to note that one of the strengths of this study is the standardized clinical definition of glaucoma and glaucoma suspect (Table 51.1). Because this was a prospective randomized clinical trial conducted across 12 clinical centers, the standardized definitions were important to promote uniformity in reporting the results. The ethics of performing a randomized, prospective trial involving a delay of surgery beyond 9 months for visually significant cataracts seems questionable.

Table 51.1 DEFINITION OF GLAUCOMA AND GLAUCOMA SUSPECT IN IATS

1. Glaucoma: IOP >21 mm Hg with one or more of the following anatomical changes:
 - a. Corneal enlargement;
 - b. Asymmetrical progressive myopic shift coupled with enlargement of the corneal diameter and/or axial length;
 - c. Increased optic nerve cupping defined as an increase of 0.2 or more in the cup-to-disc ratio; or
 - d. The use of a surgical procedure for IOP control.
2. Glaucoma suspect:
 - a. If two consecutive IOP reading above 21 mm Hg on different dates after topical corticosteroid had been discontinued without any of the anatomical changes listed above or if the patient had received glaucoma medication to control IOP without any of the anatomical changes listed above or
 - b. Took glaucoma medications to control IOP without experiencing any of the anatomical changes listed previously

From Beck AD, Freedman SF, Lynn MJ, et al. Glaucoma-related adverse events in the Infant Aphakia Treatment Study: 1-year results. *Arch Ophthalmol* 2012;130:300–305.

A few authors^{31,32} have discussed the critical period for binocular development beginning at about the 5th or 6th week of life. At our current level of knowledge, we feel it is reasonable to postpone surgeries until 5 to 6 weeks of age. This sentiment was shared in Biglan's 2006 Costenbader lecture³³ where he concluded that waiting until after 3 to 4 weeks of age before performing cataract surgery was appropriate to avoid the highest risk of postoperative glaucoma. It also allows the surgeon to operate on a firmer, more developed eye, which is technically easier to operate on.

To us, age at diagnosis of cataract is more important as compared to age at surgery. All 10 eyes that developed glaucoma in our series had cataract diagnosed within the first month of age, although for medical reasons, surgery was delayed in some. Cataract developing at an early age may be more often associated with other ocular anomalies (especially angle structure anomalies or incomplete "arrested" development), and, therefore, these patients may be at a higher risk of developing glaucoma, irrespective of their age at surgery.²⁵ Perhaps a gene that causes these eyes to have cataract at early age may also lead to increased propensity to develop glaucoma from a structural or biochemical abnormality.

Microcornea

Simon et al.⁵ found no association between microcornea and the development of glaucoma in their patients. *Microcornea* did appear to be a risk factor for glaucoma in Parks et al.'s study.³⁴ Glaucoma in aphakic eyes occurred much more frequently in patients with microcornea (23/72 eyes = 31.9%) than in patients with normal corneal diameters (3/102 eyes = 2.9%). Parks et al. noted that of the patients who developed glaucoma in aphakic eyes, all those with PFV cataracts (5/5 eyes) and all but one with nuclear cataracts (15/16 eyes) had microcornea. However, all the remaining nuclear cataract patients (35/51 eyes = 69%) and PFV patients (13/18 eyes = 72%) had microcornea but did not develop glaucoma. Development of glaucoma in aphakia is related to the two cataract types that are related to small cornea size. Keech, in an editorial immediately following the Parks et al. study,³⁴ discusses the limitations of retrospective analyses.³⁵ He also indicated that it may be difficult to identify independent influences on outcome if two factors are closely linked. He cautioned the reader to recognize that other variables, while not found to be *independently* significant in this study, may also be important factors in achieving good visual acuity (VA). Although he discussed this in reference to VA, it is also relevant for postoperative complication, for example, glaucoma.

Wallace and Plager³⁶ evaluated corneal diameter as a potential risk factor in childhood aphakic glaucoma, which had been suggested by earlier reports.^{4,34} In a retrospective analysis of all patients treated for glaucoma in

aphakic eyes within a 5-year period, 48 eyes of 29 patients were identified. The authors defined glaucoma as an IOP of >21 with an increased cup-to-disk ratio or unexpected increases in axial length or decreases in hyperopia. Prior to this report, the authors had measured the corneal diameter of 200 patients of many ages to establish an age-related curve. Microcornea was defined as a corneal diameter smaller for that eye than the diameter established by the authors' age-related curve. Almost all of the children (45/48 eyes = 94%) with glaucoma in aphakic eyes had microcornea. The frequency of microcornea in their surgical population in general was unavailable. During our initial analysis (including patients of all age), we also observed that eyes that developed glaucoma had a significantly smaller corneal diameter than did eyes that did not develop glaucoma. However, when we focused our analysis on eyes in children who underwent surgery at an early age (<4.5 months of age), we did not find a significant difference in corneal diameter between those that developed glaucoma and those that did not (pseudophakic eyes, $P = 0.860$; aphakic eyes, $P = 0.254$). Different approaches when comparing corneal diameter could have resulted in different conclusions between our and other studies. Wallace and Plager³⁶ defined microcornea as any corneal diameter smaller than that established by the authors' age-related curve. They compared corneal diameter in aphakic eyes that developed glaucoma with that of normal corneal diameter. It may be possible that all or most of the aphakic eyes had microcornea (even those eyes that did not develop glaucoma). The study did not have a control group of aphakic eyes that did not develop glaucoma. In contrast to the postoperative corneal diameter in the Wallace and Plager series, we compared preoperative corneal diameter of eyes that developed glaucoma with those eyes that did not develop glaucoma and treated corneal diameter as a continuous variable. In addition, our analysis included only eyes of children who underwent surgery in the first 4.5 months of life.²⁵ Our study was limited by lack of comparative age-related data.

The IATS 1-year results offered a variation from previous studies linking microcornea and postoperative glaucoma development.³⁰ Based on the data available from the first year of follow-up, the corneal diameter was not statistically significant to the development of glaucoma. However, the IATS group conceded that the lack of significance could be caused by the small range of corneal diameters in the study cohort and the influence of age on corneal diameter, since all patients were operated on between 1 and 6 months of age. Nishina et al.³⁷ also offered some encouraging results with their 2007 work examining bilateral congenital cataract patients with microcornea. They found that even with an average age at surgery of 7.7 weeks and an average follow-up of 115 months, 55% (6/11 patients) achieved VA between 20/20

and 20/40. Two of the remaining patients were between 20/100 and 20/200, and the final three patients were 20/100 or worse, and these three had poorly controlled postoperative glaucoma. In all, 9 eyes developed aphakic glaucoma, and all of these had corneal diameters between 7.5 and 8.5 mm. In addition, glaucoma occurred more commonly when surgery was done by 8 weeks of life (64%) compared to the overall cohort (41%). While the study only enrolled 22 eyes, the authors noted the possibility of favorable VA outcomes in this relatively high at-risk group. This was corroborated by Vasavada's study in 2009³⁸ that demonstrated the potential for good visual outcomes (visual improvement in 90.5%) in the 42 eyes of 21 patients enrolled, though this study used both limbal approach and pars plicata approaches based on pupil dilation, anterior chamber depth, and dense plaque.

Type of Cataract

Cataract types such as nuclear and PFV imply abnormal anterior segment development, and these defective angles may lead to glaucoma.⁶ Wallace and Plager³⁶ argued that certain cataract types (nuclear and PFV) cause early visual axis opacification and necessitate early surgery, and these cataract types are in turn associated with glaucoma; microcornea is the unifying sign that anterior segment dysgenesis is present (including angle anomalies), which may lead to glaucoma. Wallace and Plager's work corroborates the conclusions of earlier work by Parks et al.³⁴ A persistent fetal vasculature-type cataract was associated with a 3.1 times higher likelihood of developing a glaucoma-related adverse event in IATS 1-year outcome report.³⁰

Aphakia Versus Primary Implantation

In years past, pediatric patients were left aphakic after cataract extraction. Today, IOL implantation is a reasonable alternative, and IOLs are now sometimes implanted even in those eyes with mild-to-moderate microcornea. There have been many reports of pediatric patients developing glaucoma in aphakic eyes but fewer reports of glaucoma in pseudophakic eyes. Asrani et al.³⁹ noted striking difference in the incidence of postoperative glaucoma between aphakic and primary IOL implantation patients. They offered possible explanations including mechanical support to the trabecular meshwork and providing a protective barrier for the meshwork against invading substances from the vitreous chamber. Astle et al.⁴⁰ also demonstrated this phenomenon in their 2009 study in a cohort of 150 eyes over 24 years with primary IOL implantation from 1 month old to 18 years old at the time of surgery. In this group, not a single eye developed secondary glaucoma postoperatively. Though there was a tremendous spread in the patient age range, the average age at surgery was 66.4 months. Astle offered a recommendation, corroborated in a 2004 work by Vishwanath et al.,⁴¹ to delay surgery until after 4 weeks

(1 month) old to avoid the highest risk for postoperative glaucoma. With a similar result, Khan and Al-Dahmash⁴² reported in 2010 on 36 eyes of 31 patients without any developing postoperative glaucoma with mean follow-up of 67.1 months (60–84 range). This study was conducted only on eyes between 5 and 10 months old (mean 7.1 months). An additional study⁴³ by these authors was carried out retrospectively on patients between 5 and 141 months old at the time of surgery to include eyes beyond the infantile group. Of 489 eyes, without additional abnormalities beyond a cataractous lens, having surgery with a primary implant, only eight developed glaucoma (1.6%). The authors concluded that pseudophakic glaucoma in the pediatric subset (instead of purely infantile) does not follow a strict risk profile, in addition to being quite rare. However, the authors conceded that their data did demonstrate a clear selection bias against primary implantation before 9 months old. Beyond a postulated protective mechanism inherent to an IOL, the implanted population tended to be both older and carry fewer known risk factors for glaucoma.

We²⁵ noted in a subsequent retrospective study a similar trend overall. The patients receiving a primary IOL were less likely to develop glaucoma (10/266 3.8% pseudophakia versus 8/47 17% aphakia) but tended to be older. Beyond broad analysis, our study offered an age-matched cohort below 4.5 months age at surgery. In this group, a statistically significant difference in glaucoma was not found between aphakia (19%; 8/42) and primary IOL implantation (24.4%; 10/41). Thus, our study demonstrated that placing an IOL was not protective against glaucoma. In our study, there was less of a selection bias since we had microphthalmic eyes at high risk for glaucoma in both the aphakic group and the IOL group. This was corroborated by Kirwan et al. in 2010⁴⁴ when they retrospectively reported a statistically significantly higher age of eyes with primary IOL implantation. However, they further analyzed their cases performed below 2.5 months of age for both categories (36 aphakic and 33 pseudophakic) and found that within this subset, microcornea was not more severe in the glaucoma patients and the IOL was not protective against glaucoma. They concluded that early age at surgery (<2.5 months of age) carried a high risk of glaucoma regardless of whether an IOL was implanted. The IATS provided 1-year results that offer a similar evidence regarding age at the time of surgery being more of a risk factor than whether an IOL was placed or not.³⁰ These IATS 1-year data do not have a significant difference of age at surgery between the aphakic and pseudophakic cohorts since the patients were assigned in a randomized manner. The incidence of glaucoma in aphakic eyes has been reported to be higher when children are followed for a longer period after cataract surgery. In many of these studies, the IOL patients have been followed for a shorter period of time.

We might expect the same trend with pseudophakic eyes, and the incidence may be higher as we have longer-term follow-up.

TREATMENT

The treatment for a disease is aimed at altering the known pathophysiology of the disease by medical or surgical therapy. At present, it is unknown what causes the delayed-onset postsurgical glaucoma in children with or without IOL implantation. Although pupillary block was once the leading cause of postoperative glaucoma in aphakic eyes,⁴⁵ acute angle closure rarely occurs in aphakic eyes after modern surgery⁴; open-angle glaucoma occurs more frequently.⁴⁻⁶ A surgical or laser peripheral iridectomy is standard treatment once pupillary block is recognized. Pupillary block glaucoma may occur more commonly in pseudophakic rather than aphakic patients, especially in patients with significant amounts of inflammation or those with prior trauma or poor lens placement.^{46,47} Posterior synechia may attach to the lens capsule and promote acute angle closure. Although some surgeons perform peripheral iridectomy in every case,⁴ we reserve this step for some patients with preexisting synechia or with prior trauma in whom we are implanting an IOL, especially if the IOL is placed in the anterior chamber.

The typical approach for treating open-angle glaucoma differs for adult and pediatric patients. Treatment of glaucoma in adults is typically first managed medically, and only when maximal medical management is exhausted is a surgical remedy attempted. Management of congenital glaucoma in children often demands a primary surgical treatment, as in goniotomy or trabeculotomy, followed by the addition of topical medications for refractory cases.

The management of children with glaucoma in aphakic or pseudophakic eyes differs from that of congenital glaucoma. There is a paucity of literature available on PubMed on children with glaucoma in aphakic eyes treated with angle surgery in the face of myriad publications of congenital glaucoma treated with angle surgery. Although the number of patients treated was small, Walton's success rates for postoperative glaucoma were significantly lower after goniotomy (2/13 = 15%) than for trabeculectomy (9/14 = 64%) or for seton placement (3/6 = 50%).⁶ It has not been demonstrated whether, for cataractous children thought to be at high risk of glaucoma, performing prophylactic angle surgery (or, more interestingly, endocyclophotocoagulation) at the time of cataract surgery would forestall glaucoma in aphakic eyes without introducing other complications.

Medical management is often initiated after glaucoma is diagnosed in aphakic or pseudophakic eyes. One retrospective study of 64 eyes of 38 patients with glaucoma reported that medications alone controlled IOP in

63.6% (21/33 eyes) of patients, almost all of whom had open-angle glaucoma.¹ Timolol solution 0.25% twice daily (a beta-adrenergic antagonist, also referred to as a “beta-blocker”) is often the first medication started in aphakic or pseudophakic glaucoma. Timolol reduces aqueous production and is well tolerated by most children. However, timolol should be avoided in children with asthma or cardiac problems. When indicated, timolol should be used at the 0.25% strength twice per day or the 0.25% gel-forming solution once per day. Rarely does the 0.5% timolol produce any additional IOP lowering in children compared to the 0.25%. Additional side effect, however, may occur with the higher concentration. The relative beta-1-selective betaxolol is also used at the 0.25% strength twice daily and may offer less pulmonary risks in children compared with timolol.

Topical carbonic anhydrase inhibitors (CAIs) also reduce aqueous production. These CAIs (dorzolamide 2%/Trusopt or brinzolamide 1%/Azopt—these are taken two or three times daily) are often used as primary treatment when beta-blockers are contraindicated or as a second-line treatment when beta-blockers are ineffective alone (timolol combined with dorzolamide is available as Cosopt). The topical CAIs have minimal systemic side effects and are well tolerated by children. They are not as effective as oral CAIs (acetazolamide/Diamox) but are safer. When oral CAIs are used, it is usually as a temporary measure to get the IOP down while surgery is being planned. At times, oral CAIs are used at 10 to 15 mg/kg/d more chronically, but our preferred use is during a short transition to other medications or while scheduling a surgical intervention. Oral CAIs can produce metabolic acidosis. Side effects include tingling of the fingers and around the mouth, diminished energy levels, loss of appetite, and diarrhea.

Phospholine Iodide (PI) (Pfizer—formerly Wyeth) is echothiophate iodide for ophthalmic use. It is made as a 0.125% solution and is most often used twice daily for the treatment of aphakic glaucoma. Rarely is it used for any other type of glaucoma as will be explained below.

Wyeth (now a part of Pfizer) is the only manufacturer of the echothiophate powder used to create PI ophthalmic drops. It is often in short supply due to raw material shortage. PI is a strong miotic that is rarely used except in aphakic glaucoma. For adult glaucoma, miotic drugs (cholinergic stimulators) are rarely used. PI has been associated with cataract formation and, therefore, is not usually recommended at all for phakic patients. However, it is an effective IOP lowering medication in aphakic glaucoma in children. We use it in aphakic or pseudophakic children with glaucoma who are not controlled on Cosopt. Other miotics do not work well, but PI can produce impressive IOP lowering in otherwise medically unresponsive aphakic glaucoma patients. Many surgeries for glaucoma in these children have been avoided because

this medication is available. Pediatric ophthalmologists who treat children with aphakic glaucoma are the major lobbyists for the manufacturer to continue the limited production of PI. Currently, sales of PI are limited to one bottle per patient per month. Shipments are on-demand and are restricted to pharmacies or directly to physicians. The manufacturer does not accept stocking orders from pharmacies.

Systemic side effects from PI ophthalmic drops are rare but include gastrointestinal upset, increased salivation, and bradycardia. PI is a potent and relatively irreversible inhibitor of cholinesterase. PI may prolong the effect of succinylcholine if used during anesthesia for surgery. If surgery is needed for a patient who is taking PI, the anesthesiologist will avoid using succinylcholine (see chapter on anesthesia elsewhere in this book).

The prostaglandin analogues have become a first-line therapy for adult glaucoma. However, this class of ophthalmic topical medications is relatively less effective in aphakic glaucoma and tends to be added in children as a secondary or tertiary treatment, if used at all. They are effective in some cases of juvenile-onset open-angle glaucoma and may work well in selected children with aphakic glaucoma but not in the majority. These medications enhance uveoscleral outflow in adults with a smaller secondary effect on trabecular outflow. These positive effects on outflow are diminished in typical cases of aphakic or pseudophakic glaucoma. Latanoprost 0.005%, travoprost 0.004%, and bimatoprost 0.03% have the advantage of once-daily administration and an excellent systemic safety profile. However, they can lead to elongation and thickening of the eyelashes and change in eye color. Hyperemia is often reported as well.

The topical alpha-2 agonist, brimonidine, is used in older children and adults, but it is also relatively less effective in aphakic glaucoma. More importantly, brimonidine can produce life-threatening systemic side effects in infants (bradycardia, hypotonia, hypothermia, hypotension, and apnea) and severe somnolence in toddlers.⁴⁸ These side effects are associated with the lipophilic brimonidine much more than the more hydrophilic drugs like apraclonidine 0.5% (Iopidine), which are less likely to cross the blood–brain barrier. Iopidine is sometimes utilized at the time of angle surgery to minimize intraoperative hyphema.

For those children with aphakic glaucoma who are not controlled with medical treatment alone, surgical treatments are added (Fig. 51.2). A study by Bhola et al.⁴⁹ demonstrated the potential efficacy of medical management combined with surgical intervention as necessary for aphakic glaucoma patients to achieve a good visual outcome, 54.5% with 20/40 or better (30/55 eyes). In their study, only 15 required a surgical intervention, but one-third of the 55 eyes required six or more medication changes. These data offer encouragement about the

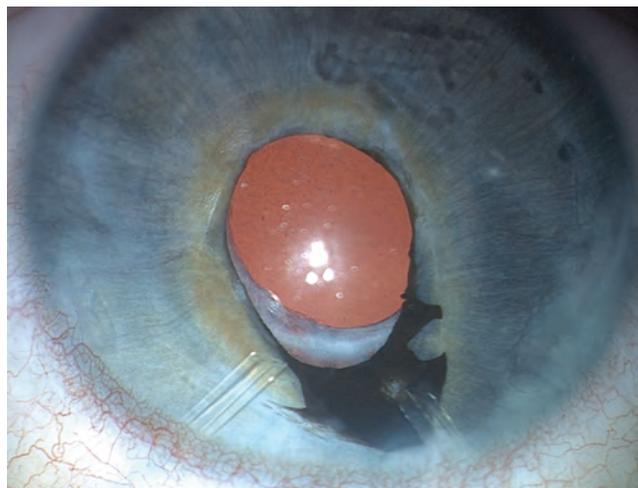


Figure 51.2. An 8-year-old boy with a diagnosis of Lowe syndrome was operated for cataract during early infancy and secondary IOL at 6 years of age. Repeated glaucoma surgery was needed. Note two intraocular tube drainage devices.

potential for good visual outcomes but underscore the necessity of close monitoring to achieve an ideal result.

When medical management fails, realistic surgical options include seton implantation, trabeculectomy, trabeculotomy, goniotomy, and cyclodestructive procedures. Aphakia has been previously reported as a significant risk factor for failure of trabeculectomy with mitomycin C (TMMC) not only in adult patients^{50,51} but also in patients <1 year of age^{52,53}; the latter studies are discussed below.

Freedman et al.⁵³ retrospectively evaluated results for 17 consecutive children (21 eyes) <17 years of age (median age, 2.6 years) who had failed maximum medical therapy, prior angle or filtration surgery (goniotomy, trabeculotomy, trabeculectomy), or both. TMMC with or without postoperative 5-fluorouracil (5-FU) or laser suture lysis or both was performed. Aphakic patients performed worse than phakic patients whether TMMC was performed before or after 1 year of age. Success was poor in all patients <1 year old, whether phakic (3/8 eyes; 38%) or aphakic (0/2 eyes). Success rates were higher in patients ≥1 year of age, for both the phakic (6/6 eyes) and the aphakic (2/5 eyes; 40%) groups. The authors contend that laser suture lysis or 5-FU augmentation of TMMC did not improve success in younger, aphakic children and may have increased the complication rate. The study is limited by the small number of patients in each subgroup and limited (median, 23-month) follow-up of successful cases. Mandal et al.⁵⁴ also reported high success rates in older, phakic patients without identifying success in the subgroup of patients who were younger or aphakic.

Beck et al.⁵² provided another report of failure of TMMC in aphakic patients <1 year of age. Records of 49 patients (60 eyes) ≤17 years of age (mean age, 7.6 years) who had undergone TMMC for various etiologies were retrospectively reviewed. Success (IOP ≤22 without

glaucoma progression or visually devastating complications) rates were 67% at 1 year and 59% at 2 years. Young age (≤ 1 year) and aphakic status were statistically significant risk factors in multivariate analysis. Failure occurred in 60% of 20 aphakic eyes and in 24% of 29 phakic eyes. Failure occurred in 7 of 8 eyes of children < 1 year old and in 29% of 41 eyes of patients 1 to 17 years of age. Late-onset, bleb-related endophthalmitis occurred in 5 of 60 (8%) eyes. Although TMMC demonstrated considerable efficacy in phakic patients > 1 year old in this and other reports,⁵⁴ the authors express concern about the “substantial” risk of infection with TMMC in aphakic infants.

Age < 1 year and aphakia are risk factors for failure of TMMC in the two aforementioned retrospective studies. How do aqueous shunt devices compare? Beck et al.,⁵⁵ in 2003, reported greater efficacy of aqueous shunt devices over TMMC for children 2 years and younger. A minority of the studied patients were aphakic or pseudophakic. In this retrospective, age-matched comparison of aqueous shunt devices and TMMC, Beck et al.⁵⁵ determined the likelihood of maintaining an IOP of < 23 mm Hg in 46 eyes of 32 patients < 2 years of age. According to the authors, pressure below this level provides clinical stability in very young patients with glaucoma. For the 46 eyes receiving aqueous shunts, 16 eyes (34.8%) were aphakic or pseudophakic, compared to 3 of 24 eyes (12.5%) in the TMMC group.

Beck et al.⁵⁵ utilized Baerveldt implants for 32 eyes and Ahmed valves for 14 eyes. After the aforementioned procedures, success achieved was 87% and 53% in the aqueous shunt device group at 1 and 3 years, respectively, compared to 36% and 19% in the TMMC group at the same intervals. Interestingly, although the seton implantation group was composed of more high-risk patients (16 of 46 [34.8%] eyes aphakic or pseudophakic) than was the TMMC group (3 of 24 [12.5%] eyes), the seton group overall (no separate success rates were reported for aphakic and pseudophakic patients) fared better (72% versus 21% success in the TMMC group) and had no infections (versus 8.3% in the TMMC group). Infection is an even larger concern for contact lens-wearing aphakic patients.⁵⁶

The poor success rates and potential for infection with TMMC for young, aphakic patients in the retrospective studies discussed previously corroborate the results of Beck and associates' work discussed above. For the first surgical procedure for aphakic or pseudophakic patients on maximal medical therapy, seton implantation appears more likely to succeed in controlling IOP than TMMC, especially in infants.

Further work on alternatives to setons and TMMC has been published recently by Bothun and colleagues. They retrospectively reviewed 16 years worth of data to evaluate the efficacy of goniotomy and trabeculotomy angle surgery procedures. The study only included 14

aphakic eyes undergoing trabeculotomy (9 eyes) or goniotomy (5 with 1 eye having a trabeculotomy reoperation), but the results were still encouraging. Success was defined as an IOP of 24 mm Hg or below (with or without topical medication), lack of a sight-threatening complication, and avoidance of trabeculectomy or tube shunt. With a median follow-up of 4.2 years and the above parameters for success, 8 (57.1%) qualified as treatment successes overall. Treatment success after a single operation was achieved in 42.9% of cases, all occurring in cases of initial trabeculotomy. The authors noted this reported success rate was much higher than a previous study³⁷ of goniotomy and trabeculotomy in aphakic patients. Though the success criteria for IOP were stricter (21 mm Hg or lower) in this previous study, the success rate was still only 16.0% compared to 42.9% by Bothun, if the more strict standards were applied uniformly. The only complication noted in Bothun and associates' work was the subretinal passage of a polypropylene suture resulting in a linear retinal pigmented epithelium disturbance. Though there are potential complications and the sample size is small, the observed complications in this study were less severe than those reported for TMMC, implanted drainage devices, or cyclodestructive procedures.

An additional development for trabeculotomy was the result outlined in the work by Beck et al.⁵⁸ in their 2011 publication regarding 360-degree suture trabeculotomy in difficult cases of pediatric glaucoma. This approach offers an alternative compared to the established use of the trabeculotome for trabeculotomy. The procedure uses 6-0 polypropylene suture to cannulate Schlemm canal 360 degrees. The suture is then pulled from both ends for a cheesewire-type effect that opens the canal, as described in Beck's earlier 1995 work.⁵⁹ Beck and colleagues conducted their 2011 study on 45 eyes of 33 patients categorized into five groups, one of which was “infantile-onset glaucoma following congenital cataract surgery.” Although this group only represented 4 eyes of the total, it allowed a comparison of how aphakic glaucoma cases respond compared to other types of childhood glaucoma. Success in this study was defined as IOP below 22 mm Hg (with medical therapy allowed), stable cup:disk ratio, and no additional surgery recommended or performed. Using these metrics, 3 of the 4 aphakic eyes with glaucoma were considered successful (75%). The primary congenital from birth group had 100% (3/3) success, followed by 67% (4/6 eyes) in the congenital category with onset after 1 year of age. Primary congenital glaucoma with failed goniotomy surgery shared the identical 75% success (3/4) with the postcongenital cataract surgery group. The infantile onset with associated anomalies group (16 eyes) had a 44% (7/16) success rate. This work demonstrated the potential benefits of applying this procedure to aphakic glaucoma and offered

a treatment outcome comparison of aphakic glaucoma against pediatric glaucomas of other etiologies.

Beyond the efficacy shown with 360-degree suture trabeculotomy in the aphakic glaucoma population, the use of a microcatheter offers the potential for greater precision in conducting this procedure. In a 2010 report,⁶⁰ Sarkisian offered a retrospective study on the use of an illuminated microcatheter (iTrack 250A, iScience Interventional) in 360-degree trabeculotomies for congenital glaucoma. Although the study only enrolled 16 consecutive eyes of 10 patients, the results offer support for the benefits of this device. Sarkisian points out that the design includes an atraumatic tip, which can also be used to inject viscoelastic in an effort to overcome resistance, and its transscleral light emitting diode (LED) to demonstrate its location. These combine to offer several advantages over using a 360-degree suture trabeculotomy. Complications in this study included 43.8% (7/16) with transient hyphema. All resolved within a week postoperatively and four resolved within the first postoperative day. Secondary surgical procedures were performed in only 12.5% (2/16 eyes). Of the 16 eyes operated on, 12 (75%) received a full 360-degree procedure, with the remaining 4 having a mixture of procedures after a 360-degree microcatheter application was unsuccessful. The 6-month postoperative IOP of all 16 eyes demonstrated a 47.0% reduction, and a statistically significant decline in IOP was reported at postoperative months 1, 3, and 6.

Seton implantation size and type are additional surgical considerations when these devices are chosen for children. Higher success rates have been reported for Ahmed glaucoma valve implants, Baerveldt implants, and double-plate Molteno implants than for single-plate Molteno implants.^{61–68}

An attractive feature of Ahmed valve implants is the immediate pressure-lowering effect delivered to the glaucomatous eye without a high risk of hypotony; the non-valved Baerveldt implants will not fully lower the pressure until the temporary tube occluder is removed at least 1 month after the original surgery. Without this temporary tube occlusion, the Baerveldt implant would produce marked early hypotony. Rapid IOP reduction may be less crucial in glaucoma in aphakia than in patients with congenital glaucoma who fail angle surgery—the latter patients depend on rapid clearing of the visual axis from lower pressure. Patients with glaucoma in aphakic eyes typically have clear corneas. It is easier to implant an Ahmed glaucoma valve than a Baerveldt in an infant due to eye and orbit size, but glaucoma in aphakia is most commonly diagnosed 4 to 5 years after the cataract surgery is performed. The valve in the Ahmed implant may fail,⁶⁹ and there is a greater probability of having a hypertensive phase in an Ahmed valve than in a Baerveldt in pediatric patients. The hypertensive phase tends to peak at 1 month and resolve by 6 months after Ahmed implantation in adults.⁷⁰

In children, success has been reported with Molteno implants,^{67,71–73} Ahmed valve glaucoma implants,^{61,62,64} and Baerveldt implants.^{65,74,75} Since all types of glaucoma implants will demonstrate a decline in success rates over time,^{63,75,76} the ideal seton implant for the aphakic or pseudophakic child with glaucoma is not currently agreed on.

Cycloablative techniques have generally been reserved for refractory cases of glaucoma in children.^{77–80} Reported success rates have been low when these techniques are used as the initial surgical option.⁸¹ Cyclocryotherapy and laser cyclophotocoagulation in children may result in severe complications in some patients. Reported complications include retinal detachment, sympathetic ophthalmia, and phthisis.^{80,82,83} Surgical revision or addition of a second tube implant can also be associated with high rates of complications such as new corneal edema.⁸⁴ Supplemental transcleral laser photocoagulation is a viable alternative for children suffering tube failure.⁸⁵ Several laser treatments may be required to achieve long-term control.

Endocyclophotocoagulation is a relatively recently applied technique⁸⁶ that has demonstrated some promise in treating refractory glaucoma in children and adults. Would this be applicable to children with glaucoma in aphakic eyes?

Neely and Plager⁷⁹ reported on 51 endoscopic diode laser cyclophotocoagulation procedures performed on 36 eyes of 29 pediatric patients. The cumulative success rate after all procedures at a mean of 19 months of follow-up was 43%, which is similar to the 50% success rates achieved by Phelan and Higginbotham⁸³ and Bock et al.⁸² with forms of transcleral Nd:YAG or diode laser. Severe visual complication rates were lowest with endocyclophotocoagulation (11% with the endoscope versus 50% or 19% of patients with transcleral Nd:YAG or diode, respectively). In fact, the authors point out that when combining their study with other studies of this procedure,^{87–90} only 1 of 123 diode endolaser-treated eyes progressed to phthisis. In contrast, cyclocryotherapy is historically associated with more morbidity, as 12% to 34% of patients treated with cyclocryotherapy progressed to phthisis in past reports.^{91–98} Nonetheless, for aphakic patients especially, Neely and Plager⁷⁹ report that endocyclophotocoagulation is not undertaken without risk, as retinal detachment, hypotony, and decreased vision all have occurred. Long-term results are not available, so this procedure should still be used with caution in children with refractory glaucoma in aphakic eyes.

The methods we use to evaluate IOP and other relevant data points in children may be inadequate. The Tonopen, which is the easiest to use and the most portable of the devices commonly used, may be less valid than the pneumotonometer.⁹⁶ The Icare rebound tonometer has added another useful but imperfect device. We remeasure IOP using a different device (as a verification) whenever the first device renders an IOP that is a surprise based

on what was expected. An ideal instrument for a child's eye would instantly, easily, and accurately measure IOP (and anterior chamber angle and optic nerve health, for that matter) and obviate diagnostic visits to the operating room. Ideally, we would want an instrument to determine whether removing a child's cataract will result in good vision and whether or not glaucoma will occur. Until the days of such magic wands arrive, surgeons must remove cataracts from the eyes of children according to available data and the principles of the Hippocratic Oath, search relentlessly for glaucoma based on clinical suspicion and knowledge of risk factors, and treat whatever glaucoma is found quickly and effectively.

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Myopic Shift in Pseudophakic and Aphakic Eyes of Children

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Pediatric eyes have a rapidly developing visual system.¹⁻⁴ The growth of different components of the eye is regulated by a process known as *emmetropization*.⁵ Alterations of the process of emmetropization after cataract surgery in children are complex and poorly understood. Both active and passive components of the emmetropization process may become altered in these eyes compared with phakic eyes. Increasing axial length (AL) can lead to a myopic shift of refraction in both phakic eyes and eyes operated for cataract surgery. However, in phakic eyes, compensatory changes occur in the optical system making large refractive errors uncommon. For example, to compensate power changes due to increasing AL, lens power declines from 34.4 to 18.8 diopters (D).² After cataract surgery, in the absence of the crystalline lens, these eyes are not able to compensate through change in lens power, and thus, a large myopic shift of refraction is observed.⁶⁻⁹ Note that this is a myopic shift of refraction, not necessarily myopia. Postoperative refraction is attributed to the refraction immediately after cataract surgery and the amount of myopic shift since surgery (postoperative refraction = initial refraction + shift of refraction, which is myopic). If the initial postoperative refraction is “0” and the shift of refraction is -5 D, the postoperative refraction would be -5 D. However, if the initial refraction is “+10” and the shift of refraction is -5, the postoperative refraction would be +5. In this case, although shift of refraction is in the myopic direction, the actual postoperative refraction is hyperopic.

A tendency toward *axial elongation and a myopic change of refraction* after pediatric cataract surgery has been reported.^{6,10-34} Studies on animals and humans have focused on various factors influencing axial growth and refractive change of the eye after cataract surgery. A cautionary note is needed at this point. Studies describing the growth of an eye can be problematic to interpret. Results from different studies are difficult to compare because of inconsistencies in inclusion and exclusion criteria, surgical technique, and length of follow-up. The impact of

length of follow-up can be slightly minimized if refractive shift per year is reported as compared with total refractive shift.³⁵ Some studies report growth as axial growth²⁴ and others, as refractive change.^{15,24-28} Some studies have reported growth as an absolute value (AL in millimeters or refraction in diopters), while others have reported rate of growth (depending on refractive or axial rate of growth—[final refraction or AL minus initial refraction or AL] divided by log of ratio of age at which initial and final refraction or AL are observed).

A word of caution is also needed here, as many of the factors described below may not occur individually. For example, when aphakic and pseudophakic eyes are compared, several factors may confound the interpretation of results. Aphakic eyes may have been operated at a younger age, may have more eyes with bilateral etiology, may have more eyes with poor visual outcome, etc. Some of these factors may have been responsible for the appearance of retardation of growth in pseudophakic eyes. When comparing unilateral and bilateral cataract, again, unilateral cataractous eyes may have been operated at an earlier age (more elongation) and may be more likely to be amblyopic (more elongation), to receive intraocular lens (IOL) implantation, and to have more initial interocular axial length difference (IALD)³⁶ between eyes, etc. These confounders may alter the results when true differences in growth between unilateral and bilateral cataractous eyes are being sought in a retrospective analysis.

It is important to differentiate “refractive change” and “axial growth.” Refraction determines the final optical correction the child needs to use. However, as it reflects not only axial changes but also changes produced by corneal curvature and IOL-related factors (more refractive change per millimeter of growth when a higher power of IOL is implanted), it is not the best way to document and report the growth of the eye. Thus, to understand the growth pattern of an eye, the reporting of axial growth is recommended.

Table 52.1 FACTORS THAT MAY AFFECT GROWTH OF APHAKIC AND PSEUDOPHAKIC EYES

I. General
1. Age of the child at the time of cataract diagnosis and cataract surgery
2. Gender
3. Ethnicity
4. Heredity
II. Ocular and related factors
A. Preoperative
1. Laterality
2. Type of cataract
3. AL, keratometry, IALD
B. Intraoperative
1. Vitrectomy
2. Aphakia versus pseudophakia
• If aphakia—optical correction with glasses, CLs, or secondary IOLs?
• If pseudophakia—primary versus secondary IOL, size of IOL, and IOL power
C. Postoperative
1. Duration of follow-up
2. Glaucoma
3. Visual axis opacification
D. Functional issues
1. Visual deprivation; density of amblyopia and compliance with amblyopia therapy; compliance with residual refractive error
2. Excessive near work and optical correction of refractive error

Table 52.1 lists factors that may affect the growth of the eye. Some of them are identified to affect growth after cataract surgery in children, while others have been reported in the literature to affect the growth of phakic eyes, which might affect aphakic/pseudophakic eyes. In this chapter, we discuss the available literature on myopic shift in aphakic and pseudophakic eyes.

GENERAL FACTORS

Age of the Child at the Time of Cataract Diagnosis and Cataract Surgery

It is well documented in the literature that the normal phakic human eye undergoes extensive growth in the postnatal period.¹⁻³ Larsen¹ reported a *rapid postnatal growth phase*, with an increase in AL of 3.7 to 3.8 mm in the 1st year and a half, followed by a *slower infantile growth phase* from the 2nd to the 5th year of life, with an increase in AL of 1.1 to 1.2 mm, and, finally, by a *slow juvenile growth phase* lasting until the age of 13 years, with an increase in AL of 1.3 to 1.4 mm. Longitudinal growth is reported to be minimal after this age. Gordon and Donzis² noted that the AL increases from an average of 16.8 mm at birth to 23.6 mm in adult life. Although most

studies have observed rapid axial growth in infantile eyes, there is no sharp cutoff point when axial growth stabilizes.

It is reasonable to believe that eyes with cataracts follow a similar triphasic curve—before surgery as well as after surgery. However, we have noted that the mean AL of our patients' cataractous eyes is different (20.52 ± 2.87 mm) from that of the noncataractous eyes in the Gordon and Donzis² data (21.9 ± 1.6 mm).³⁷ Not only did the mean values differ, but more importantly, the standard deviation was double that of the normal population. Also, the younger the age at the time of measurement, the more the variability of the AL.³⁷

The age at onset and the duration of cataract-related visual deprivation also influence axial growth. Lambert¹⁷ reported that age at the time of lensectomy appears to be a critical factor in determining subsequent axial growth in monkeys. When surgery was done at a very early age, retardation of axial growth occurred compared to the normal eye. However, this retardation was not seen with lensectomy in slightly older monkeys. Below we review the available literature on the influence of age on the axial growth of aphakic/pseudophakic eyes.

Axial Length Studies

- Flitcroft et al.¹⁴ reported a mean increase in AL of 3.41 mm in congenital cataracts (<1 year) versus 0.36 mm in developmental cataracts (>1 year) at mean follow-up of 2.7 and 2.86 years, respectively.
- Vasavada et al.³⁸ noted that the rate of axial growth (RAG) in children when operated at ≤ 1 year of age (23.5%) was significantly higher than in those operated between 1 and 3 years (4.8%; $P = 0.0001$) and 3 to 10 years old (4.3%; $P = 0.0001$). In children operated at ≤ 1 year of age, the temporal profile of RAG was higher in the first 2 years after surgery. Temporal profile of RAG was the difference between two consecutive mean ALs with respect to the previous reading.
- Fan et al.³⁹ reported that although eyes operated before 6 months of age have shorter preoperative AL compared with eyes operated between 7 and 12 months of age (18.9 mm versus 20.3 mm), 3-year follow-up AL was longer in infants operated before 6 months of age (22.7 mm versus 21.2 mm).
- We⁴⁰ reported AL elongation even during the second decade of life. Globe AL was 23.4 mm (mean age 11.5 years) at the initial measurement and 23.9 mm at the last measurement (mean age 15.2 years).

Refractive Error Studies

- Moore¹⁵ noted that the refractive error of the aphakic eye of patients treated for a unilateral congenital cataract decreases most rapidly during infancy and less rapidly during the next few years of childhood.

- McClatchey and Parks⁶ reported that the average refraction tended to follow a logarithmic decline with age. The average rate of myopic shift was -5.5 D. A stepwise regression analysis showed that age at surgery had a small but significant effect on rate of the growth. Much of the observed myopic shift in aphakic eyes is due to normal growth of the eye.
- McClatchey and Parks⁷ used aphakic refraction at last follow-up to calculate the final pseudophakic refraction, and these values were compared with the prediction of a logarithmic model of myopic shift. They reported a median calculated pseudophakic refraction at last follow-up of -6.6 D, with a range of -36.3 to $+2.9$ D. Children who underwent surgery in the first 2 years of life had a substantially greater myopic shift (11.9 D) than did older children (4.7 D) and a larger variance in this myopic shift. The logarithmic model accurately predicted the final refraction within 3 D in 24% of eyes undergoing surgery before 2 years of age and in 77% of eyes undergoing surgery after this age.
- Dahan and Drusedau¹³ reported an average elongation of 19% for age <18 months and 3.4% for age >18 months.
- Enyedi et al.²⁹ reported that children operated on at ages 0 to 2, 2 to 6, 6 to 8, and >8 years had refractive shifts of -3.0 , -1.5 , -1.8 , and -0.38 D, respectively (2.5, 2.5, 3.0, and 1.8 years postoperatively). The authors noted a statistically significant difference in the average total change in refraction between the youngest age group (0–2 years) and the oldest age group (>8 years).
- Plager et al.²⁶ reported that children operated on at ages 2 to 3, 6 to 7, 8 to 9, and 10 to 15 years had mean myopic shifts of -4.60 , -2.68 , -1.25 , and -0.61 D, respectively (5.8, 5.3, 6.8, and 5.7 years postoperatively).
- Crouch et al.¹⁰ reported that children operated on at ages 1 to 3, 3 to 4, 5 to 6, 7 to 8, 9 to 10, 11 to 14, and 15 to 18 years had mean myopic shifts of -5.96 , -3.66 , -3.40 , -2.03 , -1.88 , -0.97 , and -0.38 D, respectively, with an average follow-up of 5.45 years.
- Ashworth et al.⁴¹ reported refractive outcomes after primary IOL implantation in infants. Mean age at surgery was 18 weeks (range, 1–51 weeks), and mean follow-up was 44 (SD 31) months. The amount of myopic shift was greater in the first 12 months after surgery than was in the 3rd year (5.43 D versus 0.87 D). The amount of myopic shift in the first 12 months after surgery performed at <10 weeks of age was significantly greater than that in the later surgery group (6.26 D versus 2.33 D).⁴¹ The mean myopic shift was 5.43 D (SD 3.7 D) in the first 12 months after surgery and was significantly greater when surgery was performed at <10 weeks of age.
- Astle et al.⁴² reported mean rate of change per year of -2.05 , -1.85 , -1.10 , -0.64 , and -0.30 D for patients operated at <1 , <2 , 2 to 4, 4 to 7, and 7 to 18 years of age, respectively.
- Hoevenaars et al.⁴³ reported average change of refraction as -3.18 , -1.06 , and -0.06 for patients operated before the first birthday, 1 to 7 years, and 7 to 18 years of age, respectively. Rate of change (D/years) was -1.05 , -0.40 , and -0.05 D, respectively.
- Lu et al.⁴⁴ reported refractive shift for infantile cataract surgery (age at surgery 6–12 months). The mean value of myopic shift were 5.15 ± 2.08 , 6.46 ± 2.13 , 7.54 ± 3.16 , 7.92 ± 3.40 , 8.25 ± 3.57 , and 8.67 ± 3.62 D at 1, 2, 3, 4, 5, and 6 years after surgery, respectively. As seen, the greatest annual myopic change was observed in the first 12 months after surgery.
- Nystrom et al.⁴⁵ reported that the decrease of aphakic hyperopic refraction at the corneal plane followed a logarithmic trend. Aphakic refraction at the corneal plane equals $37.2 - 10.6 \times \log_{10}(x)$, where x = patient age in months ($R^2 = 0.95$).
- We reported mean myopic shift of refraction per year as -1.13 D in the second decade of life.³⁵ Note that we evaluated all refractions performed during the second decade of life.

Gender

Sex-linked differences in AL have been reported in the literature on normal phakic eyes.^{1,46} In eyes with cataracts, we have noted that, on average, girls have a shorter AL than do boys (20.23 mm versus 20.78 mm, $P = 0.09$).³⁷ McClatchey and Parks⁶ noted that gender had no effect on the rate of myopic shift in pediatric aphakic eyes. Further prospective studies of large sample size are required to answer the question if myopic shift of refraction is influenced by gender.

Ethnicity

Several studies in normal phakic eyes have reported racial differences in relation to axial and refractive status. The prevalence of myopia is 37% among Chinese school children^{47–48} versus only 7.5% among American school children. Gwiazda et al.⁴⁶ noted that they did not find a difference in axial dimensions in different ethnic groups. In our cataractous population, we noted significantly longer eyes in African American patients than in Caucasian patients (21.66 mm versus 20.14 mm, $P < 0.001$).³⁷

Heredity

Parental refractive error has also been shown to be an important predictor for the refractive errors of their children. For example, if both parents are myopic, 30% to 40% of their children become myopic, whereas if only one parent is myopic, 20% to 25% of their children will become myopic. If neither parent is myopic, fewer than

10% of their children will become myopic (cited in Ref.⁴⁹). Moore¹⁵ noted that 2 of 42 patients were highly myopic in their phakic eyes and were less hyperopic in their aphakic eyes than the group means. Both of these patients had a father with high bilateral myopia and can be presumed to have a hereditary form of myopia superimposed on their unilateral congenital cataract. Plager et al.²⁶ noted an unexpected larger myopic shift in a genetically predisposed patient (moderate myopia in both parents).

Down Syndrome

Eyes with Down syndrome have more myopic shift after cataract surgery.

OCULAR AND RELATED FACTORS

Preoperative

Laterality

We reported that eyes with unilateral cataracts had shorter preoperative ALs than those with bilateral cataracts before 5 years of age but had longer ALs than the eyes with bilateral cataracts when the preoperative measurement was after 5 years of age.³⁷ A way to interpret these data is that unilateral cataracts operated before age 5 were more likely microphthalmic than were age-matched bilateral cataract eyes. Unilateral cataracts operated after age 5 were more likely than were bilateral cataract eyes to have increased axial elongation from form vision deprivation.

- Rasooly and BenEzra¹⁸ reported that in cases of unilateral aphakia, the aphakic eye was consistently longer than was the normal fellow eye.
- Lorenz et al.¹⁹ reported that the mean decrease in refraction was 15 D in unilateral cataracts and 10 D in bilateral cataracts.
- In older children, Kora et al.³² reported a longer AL in operated eyes than in normal fellow eyes. These were unilateral cataracts and likely had form vision deprivation that caused the increased axial elongation.
- McClatchey and Parks⁶ reported that eyes with unilateral cataracts tended to have a higher rate of myopic shift than did eyes with bilateral cataracts; this effect was statistically significant among eyes with cataract removal after the age of 6 months.
- Results of Hutchinson et al.³¹ in a series of children <2 years old with IOL implantation showed that in eyes with a unilateral cataract, the operated eye grew an average of 0.35 mm more than the fellow noncataractous eye. (Calculation was done from data in Ref.³³; only 9 eyes with a unilateral cataract were included here for analysis.) Operated eyes grew 1.49 mm (SD, 0.74 mm) on average, while unoperated eyes grew 1.14 mm (SD, 0.88 mm).

- Griener et al.¹⁶ concluded, in their retrospective study, that there was a reduction in axial growth in unilateral pseudophakic eyes compared to fellow normal eyes. The authors noted that in seven patients receiving unilateral IOL implantation at between 2 and 4 months of age, the mean AL was 0.46 mm less in the pseudophakic eye than in the fellow eye (range, 0.15–0.70 mm).
- Crouch et al.¹⁰ noted that there was a minimal myopic shift difference in patients with binocular implants, suggesting that both eyes grew similarly. In the unilateral case, they noted that the pseudophakic eye showed (not surprisingly) a larger myopic shift than that of the unoperated fellow eye.
- Weakley et al.³⁰ noted that the difference in refractive rate of growth between good- and poor-seeing eyes was less in eyes with bilateral cataracts than in eyes with unilateral cataracts. This again shows that the effect of form vision deprivation on normal axial growth is greater in unilateral than in bilateral cataract eyes.
- Vasavada et al.³⁸ noted that in children operated at ≤1 year of age, RAG in unilateral pseudophakia was 25.5% compared with 18.5% in bilateral pseudophakia ($P = 0.001$, CI -13.00 to -0.20). In children operated at >1 year of age, the corresponding RAG was 4.2% and 4.5% ($P = 0.8$, CI -2.6 to -3.3). In children >1 year of age, laterality had no significant effect on RAG.
- Barry et al.⁵⁰ reported that there was a consistent pattern in each infant who underwent bilateral surgery, with both eyes following a similar pattern of refractive change with time.
- Leiba et al.⁵¹ reported that mean AL growth was greater for the operated eyes (1.72 mm) than for the nonoperated eyes (1.21 mm) but the difference was not statistically significant.
- Gouws et al.⁵² reported significantly greater myopic shift in the unilateral cases compared to bilateral cases (-5.53 D compared to -2.77 D).
- Walker and Romanchuk⁵³ reported that eyes with unilateral cataract surgery had a slightly faster rate of refractive change, and therefore a higher myopic final refraction, than that of eyes with bilateral surgery.
- Astle et al.⁴² reported a mean myopic shift of 1.34 D/year compared with the fellow phakic eye, which had a mean myopic shift of 0.38 D/year ($P = 0.0034$).
- Sminia et al.⁵⁴ reported that for patients operated at a younger age (<18 months), growth of the operated eyes was less than that in the fellow nonoperated eyes ($P = 0.049$). However, a large myopic shift was found in the operated eye. For patients operated after 18 months of age, no significant difference in eye growth over time was found between operated eyes and nonoperated eyes.

- Hussin and Markham⁵⁵ reported AL growth after pediatric cataract surgery with IOL implantation in children younger than 5 years. The authors found more mean AL growth for operated eyes than for nonoperated fellow eyes (4.83 mm versus 4.49 mm, $P = 0.067$). In addition, the growth in operated eyes in the unilateral cataract group was greater than the growth seen in right eyes of the bilateral cataract group (4.83 versus 4.28, $P = 0.26$). But again, this difference did not reach statistical significance. There was not a statistically significant difference in AL growth between the right and left eyes in the bilateral group (4.28 versus 4.38, $P = 0.37$).
- Hoevenaars et al.⁴³ reported that patients who underwent a unilateral phacoaspiration had a significantly higher rate of change in refraction per year than did patients who underwent bilateral phacoaspiration (-0.61 versus -0.29 D/year, $P = 0.045$).
- In a study of measurements in the second decade of life, we³⁵ reported that in unilateral cataract patients, the mean refractive shift per year was -0.19 D in the operated eye and -0.22 D in the fellow eye ($P = 0.67$).

Type of Cataract

The type of cataract does not seem to influence the growth of aphakic eyes.⁷

Axial Length, Keratometry, and Interocular Axial Length Difference

There are insufficient data in the literature to conclude with any certainty whether preoperative AL and keratometry values affect subsequent growth rates in aphakic and pseudophakic eyes. Ashworth et al.⁴¹ reported that the myopic shift in the first 12 months was significantly more in patients with an AL <20 mm (5.3 D versus 1.75 D). Studies aimed at solving the mysteries of growth of an eye after pediatric cataract surgery may need to consider IALD as a covariate. Animal studies have shown that making the eyes of chicks functionally myopic with positive spectacle lenses or functionally hyperopic with negative spectacle lenses results in a compensatory change in the growth of the eye.⁵⁶ Troilo and Wallman⁵⁷ noted that growth stopped in eyes compensating for myopia and continued in eyes recovering from hyperopia. We have reported the influence of AL of the fellow eye on the growth of an eye.⁵⁸ IALD was defined as the AL of the study eye minus the AL of the fellow eye. Three groups were formed based on the preoperative IALD: <-0.2 , group 1; ≥ -0.2 , and ≤ 0.2 , group 2; and >0.2 , group 3. The change in IALD (postoperative IALD minus preoperative IALD) was significant between the three groups (0.3, 0.2, and -0.4 mm, respectively; $P = 0.02$). The average rate of AL growth was significantly different between three groups (3.7, 2.4, and 2.5 mm, respectively; $P = 0.03$). We concluded that eyes with a shorter AL than the fellow

eye showed a postoperative RAG that exceeded the growth rate of eyes with a longer IALD.

Intraoperative Vitrectomy

Sorkin and Lambert²⁴ noted that while, in theory, vitrectomy may affect axial elongation, they did not find a correlation between axial elongation and vitreous management at the time of cataract surgery. It is difficult to design a study evaluating the effect of vitrectomy, as age would be a strong confounder. All patients operated at a younger age would have received a vitrectomy, while older children may not have received a vitrectomy.

Aphakia Versus Pseudophakia

Several studies have reported that aphakic eyes grow more than pseudophakic eyes²⁰; however, analysis of the 1-year data from the Infant Aphakia Treatment Study (IATS) revealed a retardation of growth in the aphakic eyes compared to the pseudophakic eyes.⁵⁹ For the operated eyes, the mean change in AL per month was smaller in the contact lens (CL) group (0.17 mm/month) than in the IOL group (0.24 mm/month) ($P = 0.0006$) and was independent of age at surgery ($P = 0.19$). In contrast, the change in AL per month for fellow eyes decreased with older age at surgery ($P < 0.0001$). At age 1 year, operated eyes treated with a CL were 0.6 mm shorter on average than operated eyes treated with an IOL ($P = 0.009$). These data are from a randomized control trial, and therefore, confounders are not as likely to have influenced the result. Longer follow-up will be needed to verify this growth difference over the long term. Several factors may have contributed to these conflicting conclusions with regard to a retardation of growth in pseudophakic eyes in some studies and in aphakic eyes in the IATS. Aphakic eyes in the nonrandomized reports may have a poorer visual outcome (more amblyopia) compared to pseudophakic eyes, which in turn leads to axial elongation. This could have occurred if the surgeon's criteria for implanting an IOL were different than for leaving an eye aphakic. Aphakic eyes may be shorter to start with (more microphthalmia), so to "catch up," they grow more. Most published series have a longer follow-up in aphakic eyes compared to pseudophakic eyes.

- Sinskey et al.²⁰ reported the case of an 18-year-old patient who had developmental cataracts treated at 7 years of age with bilateral cataract extraction. One eye was implanted with an IOL, and the other eye was fit with an aphakic CL. The compliance with CL wear was poor, resulting in more form vision deprivation in that eye as compared with the pseudophakic eye. At the time of secondary IOL implantation at age 18, the AL growth was seen to be excessive in the aphakic eye and normal for age in the pseudophakic eye. This occurs

despite the eyes having the same best-corrected visual acuity. This case illustrates that abnormal axial elongation can occur secondary to form vision deprivation even in the absence of amblyopia.

- Both aphakic and pseudophakic eyes have a slower rate of refractive growth after cataract surgery from 3 to 6 months of age. Overall, pseudophakic eyes showed a lower rate of refractive growth than did aphakic eyes (-4.6 D versus -5.7 D; $P = 0.03$).⁹
- Superstein et al.³³ concluded that pseudophakic eyes show less postoperative myopic shift than do aphakic eyes.
- Pan and Tang³⁴ analyzed 65 eyes of 65 patients from 5 to 12 years of age who underwent IOL implantation. Sixty-five healthy eyes were in the control group. The mean preoperative AL of the surgical group was 22.48 ± 0.44 mm and that of the control group was 22.43 ± 0.41 mm. There was no statistically significant difference in AL after 10 years in either group: 23.45 ± 0.53 mm in the surgical group and 23.41 ± 0.50 mm in the control group. All the eyes showed a myopic drift, of 3.29 D in the surgical group and 1.75 D in the control group.

If Aphakic—Optical Correction With Glasses or Contact Lenses?

To the best of our knowledge, no studies have analyzed this issue.

If Pseudophakia—Primary Versus Secondary IOL, Size of IOL, and IOL Power

To the best of our knowledge, the effect of primary versus secondary IOL implantation has not been reported in the literature. Animal studies have shown that an inappropriately sized IOL can adversely influence ocular growth.⁶⁰ Implanting a regular-sized IOL in a newborn rabbit eye retards eye growth.⁶⁰ The mathematical analysis by McClatchey⁸ opined that choosing an IOL to initially give moderate hyperopia results in less myopic shift as the eye grows than choosing an IOL to initially give emmetropia, for optical reasons alone. High-power IOLs result in a greater myopic shift per millimeter of globe axial growth. Myopic shift is greater in eyes with high-power IOL due to an optical phenomenon analogous to the effect of vertex distance.⁶¹ For example, an aphakic patient who is corrected with a +12 D lens at a vertex distance of 12 mm would require a CL power of +14 D, while a patient who requires +20 D spectacle lens would require a CL power of 26.3 D.⁶¹

Postoperative

Duration of Follow-up

Studies reporting a longer follow-up will show more myopic shift than do studies reporting a shorter follow-up. Crouch et al. observed a myopic shift that ranged

from plano to -2.25 D. With a longer follow-up of 5.45 years, the same authors observed a more significant myopic shift.¹⁰ Including eyes with insufficient follow-up time may lead to erroneous conclusions.

Glaucoma

Excessive eye elongation may be a presenting sign of aphakic glaucoma.^{62,63} However, controlled glaucoma has been shown to have no effect on the rate of growth.⁶

Visual Axis Opacification

It is reasonable to assume that, in the amblyogenic age group, if visual axis opacification exists for a sufficient period of time, the eye may become elongated. It is difficult to document the exact onset of visual axis opacification. When it is noticed, the tendency is to clear it as soon as possible. Thus, it is not surprising that many studies did not find this to be a significant factor.

Functional Issues

Visual Deprivation, Density of Amblyopia and Compliance With Amblyopia Therapy, and Compliance With Residual Refractive Error

Emmetropization of the eye may be affected by visual experience. Excessive eye elongation may be induced in experimental animals by lid suturing, corneal opacification, or opaque CLs. These treatments resulted in axial elongation, whereas lesser degrees of visual deprivation were shown to retard axial growth.⁶⁴⁻⁶⁷ Poor vision seems to influence the evolution of ocular growth away from emmetropization.⁶⁸ In humans, however, this is not always predictable.^{69,70} In the case of cataracts in children, two main factors predict the pre-cataract surgery AL. Developmental anomalies may lead to microphthalmos and a shorter AL, while deprivation amblyopia may lead to axial elongation. In a case report of identical twins, Johnson et al.⁷¹ reported that the AL of the visually deprived eye was 2 mm longer. As compared to the amblyopic eye, the fixating eye showed increased AL in hypermetropia and less increased AL in myopia.⁶⁸ Authors concluded that good vision seems to influence the evolution of ocular growth toward emmetropization.

Several investigators have examined the association between visual acuity outcome and myopic shift or axial elongation in both aphakic and pseudophakic eyes. McClatchey and Parks⁶ noted that best-corrected visual acuity had no significant effect on rate of myopic shift. Weakley et al.³⁰ noted that the rate of growth was significantly lower in eyes with better acuity.

Excessive Near Work and Optical Correction of Refractive Error During Childhood

Near work may increase the risk of myopia. Optical correction of refractive error during childhood may disturb

the process of emmetropization. A higher percentage of children with moderate or high hyperopia remain hyperopic if their hyperopia is corrected with spectacles during infancy.⁷² In the process of emmetropization, the rate of change of refractive error has been shown to be greater in the presence of higher initial refractive errors.⁷³ Thus, if we leave behind high residual refractive error, these eyes may grow more. However, a recent study found no difference in the amount of myopic shift between age-matched groups, who were either corrected near emmetropia with an IOL or left with significant hyperopia at the time of implantation.⁷⁴

PREVENTION

Eyes with cataract surgery without IOL implantation will result in refractive shift based on causes that are generally not modifiable. However, eyes with cataract surgery and IOL implantation will result in refractive error that is based on residual refractive error (in general, hypermetropia) at the time of that surgery. This residual refractive error is based on selection of an IOL power. Historically, three major approaches have been used for IOL power calculation in children—initial hyperopia, initial emmetropia, or a customized approach. Each approach has its pros and cons, which has been described in the chapter on IOL power calculation (Chapter 7). Initial high hyperopia helps prevent high myopia later; however, it may hamper initial amblyopia management. Initial management with temporary pseudophakia (piggyback IOLs), with the anterior IOL of a low power for possible removal if the eye becomes overly myopic, may be a helpful strategy (see Chapter 28).⁶¹

TREATMENT

As time passes, more and more children operated for cataract will present for treatment with significant myopia. Fortunately, there are many options for these patients, including glasses, CLs, photorefractive keratectomy (PRK) or laser in-situ keratomileusis (LASIK), and implantable collamer lenses. For aphakic eyes, if secondary IOL implantation is considered, choosing IOL power helps to achieve near-emmetropic refraction. For pseudophakic eyes, choices include IOL exchange, implantation of a piggyback IOL, or corneal refractive surgery. Each of the currently available refractive procedures has advantages and disadvantages when used on pediatric patients. LASEK allows faster visual rehabilitation and needs a shorter course of postoperative medications. The corneal flap, however, may be vulnerable to childhood trauma. PRK would hold up much better to trauma but has a slower healing and needs a longer course of postoperative medications. Laser-assisted sub-epithelial keratectomy (LASEK) has been used in children as a variation in the PRK procedure, but more data are needed before its

role in pediatric refractive surgery can be defined. Each of these procedures could be used for the moderate degree of myopia that will develop in most pseudophakic children and young adults. If high myopia develops, an IOL exchange would likely be chosen over laser refractive surgery. As more data are collected, nomograms designed specifically for children, perhaps even specific to pseudophakic children, may be developed. After cataract surgery, children will most likely need laser refractive surgery at the end of their growing years. At that age, predictability and stability may approach that found in adults.

SUMMARY

Predicting ocular growth and postoperative refractive changes to appropriately calculate IOL power has become one of the major challenges in pediatric cataract management. The surgeon who performs pediatric cataract surgery in young children must be prepared for a wide variation in long-term myopic shift. Both the magnitude of and the variance in this myopic shift are likely to be greater among children who undergo cataract surgery in the first few years of life.

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Quality of Life in Pediatric Cataracts

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In 1946, the Constitution of the World Health Organization defined health as “a state of complete physical, mental, and social well-being and not merely the absence of disease or infirmity.” Yet, until recently, medical care and health research have focused almost exclusively on physical disease-related outcomes. As medical innovations led to better treatment outcomes, the importance of assessing treatment protocols not only in terms of their physical effects but also in terms of the impact on quality of life (QOL) became apparent. The value of determining QOL of patients is now well recognized within health care practice and health research as beneficial in addition to standard clinical measures. For example, in adults with cataracts, QOL measures correlate more highly with patient-perceived disability than do standard clinical tests like visual acuity,¹⁻³ and so are being implemented in both clinical trials and clinical care.

Most patient reported outcome measures (PROMs), such as those available in the Patient Reported Outcomes Measurement Information System (PROMIS®) developed by the US-based National Institutes of Health, are intended for use as primary or secondary end points in clinical studies of the effectiveness of treatment across a range of medical conditions.⁴ In addition, PROMs can be used on an individual level to facilitate the design of treatment plans, and aid communication and management of chronic diseases. While the PROMIS® instruments assess dimensions common to many acute and chronic diseases, including emotional distress, pain, physical function, and social roles/activities, they were not designed to be sensitive to the effects of ophthalmic conditions. Most PROMs are questionnaires composed of fixed sets of items, but some now use item banks and computer-adaptive testing^{5,6} to guide item selection based on the patient's responses to prior questions, yielding more rapid and precise assessment.

Numerous questionnaires have been developed to evaluate the impact of specific ophthalmic conditions in adults. Adult onset cataracts in particular have been extensively researched, with at least 16 different PROMs available.⁷ However, PROMs designed for adults treated for cataracts include activities that are not applicable to the pediatric

population, including driving and work-related activities, while ignoring important pediatric domains such as development milestones, education, and bullying. Moreover, the intraoperative and postoperative adverse events, postoperative management challenges, and severity of visual impairment differ from those experienced by adults.

Many of the pediatric PROMs are generic tools designed to assess and compare health status among children with different diseases, among children undergoing alternative treatments for the same disease, or between sick and healthy children.^{8,9} Such PROMs are important to gain a more complete understanding of the burden of pediatric eye disease within the context of other pediatric diseases. On the other hand, generic pediatric PROMs may lack sensitivity to detect small but clinically significant differences in vision-related QOL over time or due to treatment in the context of clinical trials. Both pediatric vision-related PROMs^{10,11} and pediatric ophthalmic condition-specific PROMs^{12,13} have been developed, to evaluate vision-related activities, and mental and social well-being, outcomes that are difficult to quantify and are consistently underreported by physicians.¹⁴

Proxy reporting by a parent provides an opportunity to evaluate QOL when a child is too young or cognitively unable to complete a questionnaire or interview. However, the perception of the proxy is influenced by many factors, including whether the mother or father completes the questionnaire,¹⁴ the age of parents,¹⁵ if any siblings are disabled,¹⁵ and parental QOL.¹⁶ Parents consistently reported a lower QOL in their child than did the child in a recent study of low vision¹⁷ and in a study of children undergoing cancer treatment.¹⁸ For these reasons, the U.S. Food and Drug Administration currently discourages proxy-reported outcome measures.¹⁹ The creation of pediatric self-reporting instruments which involve children in the development with the aim of capturing the child's concerns and experiences directly has become a priority.²⁰⁻²²

There are currently a number of PROMs available that could be, or have been, used to assess children with cataracts, as shown in Table 53.1. None were specifically

Table 53.1

PROMS THAT HAVE BEEN USED TO EVALUATE THE EFFECTS OF CATARACTS, ASSOCIATED OCULAR MORBIDITIES, AND TREATMENTS ON QOL AS REPORTED BY CHILDREN, THEIR PROXIES (PARENTS ANSWERING ON BEHALF OF THEIR CHILD), OR THEIR PARENTS (REPORTING HOW THE CONDITION AFFECTS THEM, NOT THEIR CHILD)

Instrument	Developer	Subscales	Intended Age Range	Intended Mode of Administration	Relevant Studies
Overall QOL					
PedsQL™	Varni et al. ²³	Physical function Psychological function Social function Symptoms Treatment	2–5 y 5–18 y	Child and proxy	Children with cataracts ²⁴ Children with amblyopia and/or strabismus ²⁵ Children with refractive error ^{26,27} Children with uveitis ²⁸
HUI3-HRQOL	Furlong et al. ²⁹	Vision Hearing Speech Ambulation Dexterity Emotion Cognition Pain	5+ y	Child if possible	Visually impaired children ages 3–8 y ³⁰
PSI	Abidin ³¹	<i>Child domain:</i> Distractibility/ hyperactivity Adaptability Reinforces parent Demandingness Mood Acceptability <i>Parent domain:</i> Competence Isolation Attachment Health Role restriction Depression Spouse	3 mo–10 y	Proxy	Children with infantile cataracts ³² Children with amblyopia ³³
Pediatric Vision–Related QOL					
CVFQ	Felius et al. ³⁴	General health General vision Competence Personality Family impact Treatment difficulty	0–7 y	Proxy	Children with congenital cataracts ^{35,36}
Cardiff Visual Ability Questionnaire for Children	Khadka et al. ¹¹	Education Near vision Distance vision Getting around Social interaction Entertainment Sports	5–18 y without additional disability	Child	Visually impaired children ages 5–19 y ¹¹
IVI_C	Cochrane et al. ³⁷	None	8–18 y without additional disability	Child	Visually impaired children ages 8–18 y ³⁷
LV Prasad-Functional Vision Questionnaire	Gothwal et al. ³⁸	None	8–18 y without intellectual impairment ³⁹	Child	Visually impaired children ages 8–18 y ³⁸

Table 53.1

PROMS THAT HAVE BEEN USED TO EVALUATE THE EFFECTS OF CATARACTS, ASSOCIATED OCULAR MORBIDITIES, AND TREATMENTS ON QOL AS REPORTED BY CHILDREN, THEIR PROXIES (PARENTS ANSWERING ON BEHALF OF THEIR CHILD), OR THEIR PARENTS (REPORTING HOW THE CONDITION AFFECTS THEM, NOT THEIR CHILD) (Cont'd)

Instrument	Developer	Subscales	Intended Age Range	Intended Mode of Administration	Relevant Studies
EYE-Q	Angeles-Han et al. ²⁸	Photosensitivity Night vision Visual aids Visual function Competence Sports	8–18 y	Child	Children with uveitis ²⁸
Vision-Related QOL (Not Pediatric)					
NEI-VFQ-25	Mangione et al. ⁴⁰	General health General vision Ophthalmologic pain Near vision Distance vision Social functions Mental problems Social role Dependency Driving Color vision Peripheral vision	21 y or older	Adult	Children with strabismus ⁴¹ Adolescents and young adults with a history of congenital cataracts ²
VF-14	Steinberg et al. ¹	None	21 y or older	Adult	Children with nystagmus ⁴² Children with amblyopia and/or strabismus ⁴³
Hospital Anxiety and Depression Scale	Zigmond and Snaith ⁴⁴	Hospital anxiety Depression	21 y or older	Adult	Children with strabismus ⁴¹
LVQOL	Wolffsohn and Cochrane ⁴⁵	Distance vision, mobility, and lighting Adjustment Reading and fine work Activities of daily living	21 y or older	Adult	Children with visual impairment ⁴⁶
Psychological/Social Effects on the Child and Treatment Difficulty					
Minnesota Child Development Inventory^a	Ireton ⁴⁷	General development Gross motor Fine motor Expressive language Comprehension/conceptual Situation comprehension Self-help Personal/social	1–6 y	Proxy	Children with congenital cataracts ⁴⁸
Child Behavior Checklist	Achenbach and Edelbrock ⁴⁹	Internalizing Externalizing	2–16 y	Proxy	Children with congenital cataracts ⁴⁸
Perceived Psychosocial Questionnaire	Lukman ^{33,50}	None	No lower limit specified	Proxy	Children with amblyopia ³³
Self-perception Profile for Children	Harter ⁵¹	Scholastic Social Athletic Physical appearance Behavioral Global self-esteem	8–16 y	Child	Children with amblyopia ⁵² Children with myopia ⁵³

(Continued)

Table 53.1 PROMS THAT HAVE BEEN USED TO EVALUATE THE EFFECTS OF CATARACTS, ASSOCIATED OCULAR MORBIDITIES, AND TREATMENTS ON QOL AS REPORTED BY CHILDREN, THEIR PROXIES (PARENTS ANSWERING ON BEHALF OF THEIR CHILD), OR THEIR PARENTS (REPORTING HOW THE CONDITION AFFECTS THEM, NOT THEIR CHILD) (Cont'd)

Instrument	Developer	Subscales	Intended Age Range	Intended Mode of Administration	Relevant Studies
Revised Rutter Parent Scale for Preschool Children	Rutter ⁵⁴	Emotional Conduct	Up to 5 y	Proxy	Children with amblyopia ⁵⁵
Ocular Treatment Index	Drewe et al. ³²	None	Young children— no set limits	Proxy	In use with children with cataracts but results not yet published ⁵⁶
ATI	Cole et al. ⁵⁷	Adverse effects Compliance Social stigma	3–6 y	Proxy	Children with amblyopia ⁵⁸
Strabismus, Nystagmus, and Refraction					
ASQ	Felius et al. ¹⁰	Fear of losing better eye Distance estimation Visual disorientation Double vision Social contact and appearance	18+ y	Adult	None published yet
IQTQ	Hatt et al. ¹²	Three questionnaires for child, proxy, and parent Subscales: Psychosocial effects Function Eye muscle surgery	2–17 y	Child (age 5+) Proxy Parent	Parents of children with intermittent exotropia ⁵⁹ Spectacle wear in children ²⁷
Pediatric Refractive Error Profile	Walline et al. ¹³	Overall vision Near vision Far vision Symptoms Appearance Satisfaction Activities Academics Handling Peer perceptions	8–17 y	Child	CL wear in children ^{60,61}

^aThis has been replaced by the Child Development Inventory.

designed for children with cataracts, but they do address potential functional deficits due to the visual impairment or common comorbidities, such as amblyopia or strabismus. The table highlights the age range for which each PROM was designed, its subscales, and whether it is administered to the child or a proxy.

PEDIATRIC HEALTH-RELATED PROMs

PROMs developed to evaluate pediatric health-related QOL have demonstrated the impact of visual impairment, both in isolation and in conjunction with other deficits.³⁰ Tools such as the Health Utilities Index have the advantage of covering a wide range of domains; however, this

particular tool is better suited to studying outcomes of a large population than for identifying deficits in individuals, due to low levels of agreement with standardized measures of ability.⁶² The Pediatric QOL Inventory (PedsQL™) has been utilized in numerous studies, evaluating the impact of many pediatric conditions, including visual impairment. Figure 53.1 summarizes data from different ocular conditions; children with retinal disorders have the lowest mean score,⁶³ but children with congenital cataracts have lower scores than do those with uveitis,²⁸ visual acuity >0.3 logMAR,²⁶ strabismus,³⁰ and amblyopia.²⁵

In pediatric cataracts, postoperative treatment is crucial in determining the long-term outcome, with parents bearing responsibility for optical correction and amblyopia

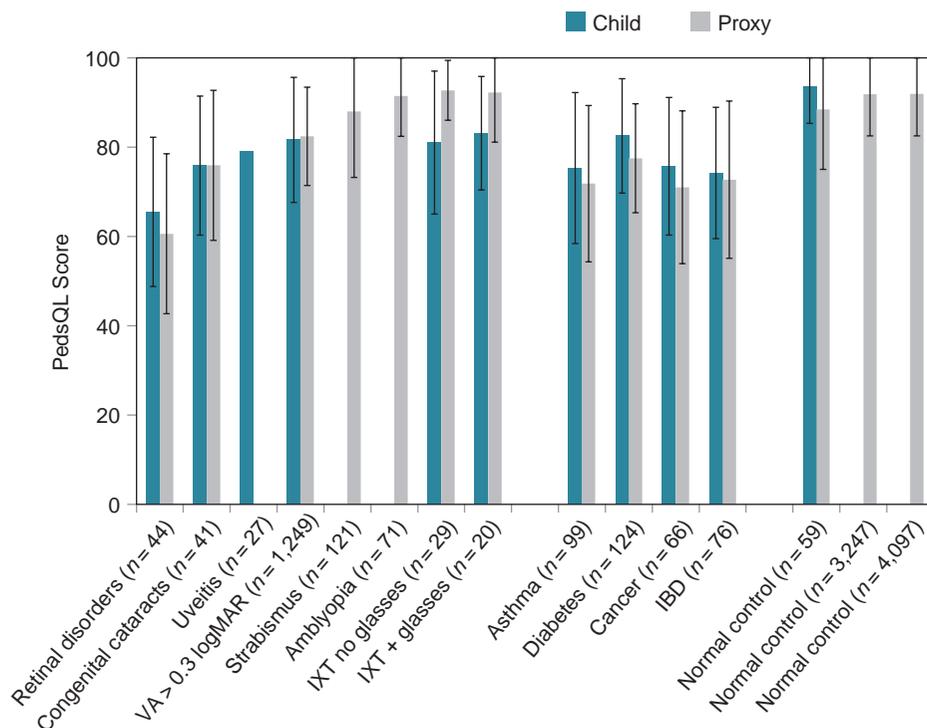


Figure 53.1. PedsQL child and proxy scores (mean ± SD) for children with vision disorders,^{24–28,59,63} general health disorders,⁶⁴ and normative data.^{25,27}

treatment. Despite the demands of treatment, parents of children with congenital cataracts reported normal parenting stress levels on Parenting Stress Index (PSI)³² but with a trend for parents of children with unilateral congenital cataracts to report higher levels of stress than did parents of children with bilateral congenital cataracts, particularly parents of children who wore aphakic contact lenses (CLs).³²

QOL in children with cataracts may also be diminished by the effects of anesthesia on the growing brain.⁶⁵ The risk of developmental and behavioral disorders increases in relation to the number of surgeries, with a hazard ratio of 1.1 (95% CI: 0.8, 1.4) for 1 exposure to anesthesia, 2.9 (94% CI: 2.5, 3.1) for 2 exposures, and 4.0 (95% CI: 3.5, 4.5) for ≥3 exposures.⁶⁶ The number of exposures to anesthesia during infancy is higher in children with primary intraocular lens (IOL) implantation than for children treated with aphakic CLs (63% versus 12% required at least one additional surgery).⁶⁷ Additional surgeries (e.g., secondary membrane removal, strabismus correction, glaucoma surgery) and multiple examinations under anesthesia also may increase this risk.

PEDIATRIC VISION-RELATED PROMs

Pediatric Cataracts

The first report of a condition-specific instrument to evaluate QOL in pediatric cataracts was by Hiatt in 1998.⁶⁸ The instrument, which was not validated, assessed

psychosocial function, parental concerns about vision, and the parents' opinion about how the visual impairment affected the child's education and work. Lower psychosocial scores were found for children with bilateral cataracts compared to those with unilateral cataracts. Later research, using the validated Children's Visual Function Questionnaire (CVFQ), differentiated among different aspects of the QOL, including competence, personality, family impact, and treatment difficulty.³⁵ Children with bilateral cataracts had lower competence subscale scores in two studies^{35,36}; on the other hand, those with unilateral cataracts had lower scores on the family impact and treatment difficulty subscales.

Other pediatric vision-related PROMs, such as the Cardiff Visual Ability Questionnaire for Children (CVAQC),¹¹ were designed for children whose visual impairment is their only disability. The etiology of pediatric cataracts is variable but has been reported in association with syndromes and systemic disease in 16%⁶⁹; therefore, PROMs like the CVAQC are not appropriate for a population-based study of pediatric cataracts.

VISION-RELATED PROMs, NOT CHILD SPECIFIC

There are a range of validated vision-related PROMs, initially developed for use with adults, which have been applied to pediatric cohorts, as shown in Table 53.1.

Overall, pediatric cataracts have been shown to have a significant impact on QOL using the instruments developed for adults, but large study differences are present. These differences may be attributable to the different populations assessed, as the mean control group scores also differ significantly among studies. Decreasing QOL with increasing age in children with bilateral visual impairment has been reported,⁴⁶ as with increasing age, even when visual acuity remains constant, visual tasks may become more demanding (e.g., smaller textbook print).

ADDITIONAL OCULAR CONDITIONS

The ophthalmic consequences of pediatric cataract can include amblyopia, strabismus, refractive errors, and nystagmus, all of which may impact the patient and family.

Amblyopia

Stimulus deprivation amblyopia has been reported to have the same mean social acceptance scores as strabismic amblyopia, which was statistically significantly worse than the control group.⁵² Note, however, the small sample size of the stimulus deprivation group and the larger standard deviation than in the strabismic amblyopia group, which may have contributed to the failure to detect a significant difference. Amblyopia treatment may also have an effect on QOL.⁷⁰ Irrespective of the condition-specific PROM utilized, occlusion therapy had a negative impact on the child.^{52,55,58} The negative impact of occlusion treatment did not reach statistical significance in all cases,³³ and reassuringly, the child's global well-being and behavior were not affected during or after treatment.⁵⁵

Strabismus

The incidence of strabismus can be as high as 85% in cases of unilateral congenital cataract.⁷¹ Strabismus has a significant impact in social interaction from a young age⁷² with potential consequences in adulthood on job prospects and finding a partner.⁷³ Reduced or nil stereoacuity is associated with strabismus, and very few children with congenital cataracts develop normal stereoacuity.^{74,75} The presence of stereoacuity provides quantifiable benefits in the speed and accuracy in performing fine motor skills,⁷⁶ and a lack of stereoacuity is also strongly associated with reduced vision-related QOL.⁷⁷

Refractive Errors

Whether the child is left aphakic or pseudophakic after cataract removal, further refractive correction will be required. Instruments such as the CVFQ include subscales on treatment difficulty, addressing discomfort, difficulty with activities, and the parent's experience. Treatment difficulty scores are worse when treated with an aphakic CL than with an IOL.³⁵ Using a PROM designed

specifically to evaluate the efficacy of CL treatment for pediatric cataracts, including the perceptions and experiences during treatment, Ma et al.⁷⁸ found that in the early stages of treatment, the resistance to treatment score was high. This did reduce with time, but, even after 3 years of CL wear, 18% reported high levels of resistance. These reports of difficulty with CL treatment are in young children, where the responsibility lies with the parent, but in the long term, different factors may come into play. The perceived benefits of CLs compared to glasses, in myopic children, include benefits when performing certain activities, such as sports, and their appearance with the optical correction.⁶⁰

Nystagmus

About 65% of children treated for dense congenital or infantile cataracts develop nystagmus,^{79,80} primarily fusion maldevelopment nystagmus syndrome (previously "manifest latent nystagmus"), but infantile nystagmus syndrome develops in about 25%. Both forms of nystagmus are present during binocular viewing and may significantly impact on QOL, on both functional ability and appearance.^{42,81} Although nystagmus has similar characteristics to other types of visual impairment, the appearance of the condition is unique to nystagmus, and as yet, there is no condition-specific PROM for nystagmus.

SUMMARY

Data from children demonstrate a significant impact on QOL in many domains, affecting both the child and parent. At present, there is no single validated PROM specifically designed for children with cataracts. Even with a pediatric cataract-specific PROM, a strong correlation between severity of visual impairment and QOL may not be present because access to support for treatment, educational accommodations, family socioeconomic status, and family and peer relationships may modify the effects of cataracts on the child's QOL. What is clear is that children and their families are broadly impacted by pediatric cataracts, so assessment of QOL is of benefit in both research and practice settings.

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Low Vision Rehabilitation

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CORE MESSAGE

- Service models and providers for the pediatric low vision population need to work together to identify and provide appropriate intervention on a timely basis.
- Thorough evaluations need to be completed, and effective intervention needs to be addressed in the medical plan of care as well as the developmental intervention plan and the mandated educational plan.
- Appropriate low vision therapeutic intervention is imperative for optimal outcomes to address the child's visual challenges and to achieve the desired functional skills.

INTRODUCTION

Partial vision loss that cannot be corrected causes a vision impairment known as low vision.¹ “Low vision” is a general term for people with visual impairments that interfere with their activities of daily living (ADLs) and quality of life. Low vision care is not a cure. It is about rehabilitation, finding new ways to accomplish the tasks of daily life (e.g., helping children in school and play).

Pediatric cataract surgery in childhood is often complicated by preexisting amblyopia, sensory nystagmus, strabismus, and abnormal binocularity. In infancy, the eye may be microphthalmic with maldevelopment of both the anterior and posterior structures in the eye. Low vision of at least a moderate degree is very common in this population. Even when the resulting postsurgical visual acuity is normal, these eyes often have residual refractive error and are all rendered presbyopic by the surgery. There is a high potential for interference with ADLs in every child operated for cataracts. A customized rehabilitation plan can be developed for each aphakic or pseudophakic child. Surgery often produces dramatic improvements in best-corrected visual acuity. However, the loss of accommodation that accompanies removal of the crystalline lens and the changing refractive error that accompanies eye growth can challenge even the child with a 20/20

outcome. In addition, many children have moderate or even severe low vision after surgery as a result of delays in presentation for surgery or anatomic abnormalities that cannot be normalized with surgery. The low vision occupational therapist and the low vision optometrist or ophthalmologist can assist the pediatric ophthalmic team in many ways. Vision rehabilitation services are underutilized for aphakic and pseudophakic children. Children presenting with impaired visual function after corrective surgery will benefit from low vision rehabilitation services that help optimize visual functioning with regard to the execution of ADLs. Many, if not most, of these children qualify and would benefit from visual rehabilitation. In addition, legislation designed for all forms of childhood visual impairment apply to this special group of patients.

FEDERAL LEGISLATION AS IT APPLIES TO VISUALLY IMPAIRED

Landmark federal mandates have had a significant influence with regard to early intervention services in the United States. In nearly every instance, children with visually significant congenital cataracts qualify for these early intervention services. Children may present for surgery at any age, and the law now provides for services beginning even in infancy. The pediatric ophthalmic team should enroll these children into the early intervention system based on the high risk for visual developmental delay and permanent visual impairment.

In 1990, the Education for All Handicapped Children Act, which had mandated free and appropriate public education to all children with disabilities 3 to 21 years of age, was reauthorized in P.L. 101-476² and renamed the Individuals with Disabilities Education Act (IDEA). In 1997, IDEA amendments under P.L. 105-17³ specified that services were to be delivered to infants from birth to 36 months under Part C, which became law in 1998. With the implementation of these laws, the federal law requires that eligible infants receive comprehensive, multidisciplinary evaluations and that an Individualized Family Service Plan

(IFSP) be developed for the infants in conjunction with their family. The primary intent of the law was to enhance the development of handicapped infants and toddlers and to minimize their potential for developmental delays; to enhance the capacity of families to meet the special needs of their infants and toddlers with handicaps; to provide assistance to states to develop and implement a statewide, comprehensive, coordinated, multidisciplinary, inter-agency program of early intervention services for infants, toddlers, and their families; and to expand and improve existing early intervention services being provided to handicapped infants, toddlers, and their families.²

Eligibility for IDEA identifies three criteria:

- A developmental delay in one or more of the following areas: physical development, adaptive development, cognitive, and/or communication and/or social-emotional development
- An established risk or diagnosed condition that has a high probability of resulting in delay, including visual impairments (and other diagnoses)
- A biologic or environmental risk, such as medical or home conditions that may significantly compromise a child's health and development²

SERVICE MODELS AND SERVICE PROVIDERS FOR VISUALLY IMPAIRED

The IDEA identifies a variety of services that may be provided through early intervention when indicated. To meet the infant's needs as specified on the IFSP, services specific to the visually impaired and/or multiple handicapped may include the following: medical services for diagnosis and evaluation, nursing services, nutritional services, occupational services, physical therapy services, audiology services, speech and language services, social work service, special education services, vision services, assistive technology devices, family training, counseling and home visits, and coordination of services. The IDEA mandates a multidisciplinary service system and multidisciplinary assessments for eligible infants, which has been interpreted as three models for teaming that include multidisciplinary, interdisciplinary, and transdisciplinary.

The medical model for the visually impaired would be considered to be a multidisciplinary approach in that each professional conducts his or her specific assessments and develops and implements resulting interventions. This information is shared mainly through written reports. The medical model would include an ophthalmologist (MD) to diagnose and treat the medical needs as related to the eye and vision. A referral would then be made to the low vision optometrist (OD) or ophthalmologist for a functional visual assessment and to determine appropriate magnification and select any devices (optical, nonoptical, and electronic) or techniques that may be indicated to enhance

optimal vision performance. The MD or OD would then make a referral to the low-vision occupational therapist who would address the child's functional skills as they relate to all aspects of life including the academic requirements. Depending on the child's diagnoses and identified needs, the combined findings of the ophthalmologist, the low vision optometrist/ophthalmologist, and the low vision occupational therapist may include recommendations for other key service providers for the visually impaired.

The combined efforts of the medical and educational representatives would represent the desired interdisciplinary model. This service delivery model is used to determine appropriate interventions in a coordinated approach to low vision services. It requires a concerted effort to share key providers' assessment results in order to collaboratively determine the optimal action. Once the detailed plan of actions is determined, frequent communication among key providers (Table 54.1) is needed to modify the actions in response to changes in the child's growth and development, and his or her academic and social environment. A completely new action plan may be needed if there are significant improvements or deteriorations in visual functioning.

Table 54.1 KEY SERVICE PROVIDERS FOR THE VISUALLY IMPAIRED WITH CONSIDERATION OF THE MULTIPLY HANDICAPPED

Pediatrician (MD)
Ophthalmologist (MD)
Ophthalmologist Specialist (Pediatric) (MD)
Optometrist Specialist (Low Vision) (OD)
Certified Ophthalmic Technician (COT)
Occupational Therapist (OTR) a specialty certification in Low Vision (SCLV)
Certified Low Vision Therapist (rehab professional certified in low vision [CLVT])
Case Manager (State Early Intervention Program and State Commission for the Blind)
Certified Teacher of the Visually Impaired (TVI)
Early Interventionist (EI)
Certified Orientation and Mobility Specialist (COMS)
Physical Therapist (PT)
Audiologist (CCC-A)
Rehabilitation Teacher (RT)
Speech and Language Therapist (CCC-SLP)
Social Worker (SW)
Nurse (RN)

Note that there are other professionals not listed who may also be key service providers.

Educational and medical services providing for infants/toddlers/children/adolescents with low vision are charged with caring for the sight and vision of this visually impaired population. This includes the health of the eye, medical treatments, prescription of lenses, assessments for functional use of vision, and work toward improvement of functional vision with and without prescribed low vision devices (optical, nonoptical, and electronic).⁴ These professionals are trained to extend the functional vision for the visually impaired child to become as visually independent as possible and to gain better control over their environment. The low vision rehabilitation specialists assist in the appropriate selection and integration of accommodations, adaptations, and assistance. The framework of treatment intervention addresses compensatory skills, orientation and mobility, ADLs (developmentally age appropriate level), sensory efficiency, academic/career education, recreation and leisure skills, social interaction, and self-esteem and determination. It is important to understand that these services are needed if the child is a print reader, a dual media reader (Braille and print), or a Braille reader. With the proper assembly of services, the visually challenged child and his/her family will be able to make good choices, based on functionality, whether to use vision and/or to choose nonvisual approaches.

It is important for the key providers of low vision services to coordinate services and resources and to make referrals to appropriate state and federal agencies, special schools, and local educational schools districts. This coordination implies that the medical model providers refer to educational providers and vice versa. It is imperative that the educators of the visually impaired advocate for medical–clinical low vision evaluations of the ophthalmologist, low vision optometrist, and occupational therapist. It is also imperative that the medical providers advocate and refer for the appropriate services for the visually impaired in the educational system. To better ensure comprehensive low vision care for children, consideration should address the following areas⁴: sociology, public policy, medicine, education, economics, and research.

- *Sociology*: It is important not to restrict medical and education services due to predetermined or limited expectations about a child presenting with low vision. Proactive and supportive referrals between and among the professionals in the medical and educational models will enhance low vision services and promote optimal outcomes.
- *Public Policy*: It is important for the medical/educational/rehabilitation needs of children with visual challenges be addressed in public state and federal policies. This will help to ensure that barriers for the low vision between the ages of 0 and 21 years are addressed and hopefully eliminated.

- *Medical*: It is important for the pediatrician to refer infants/children/adolescents to the ophthalmologist when there is a question regarding vision. In the specific case of a child with aphakia or pseudophakia, the pediatrician can help assure that medical follow-up and rehabilitation are not being ignored. In addition, changes in visual functioning or loss of optical correction may come to the attention of the pediatrician during visits for other acute illnesses or well-child visits. It is also essential that the pediatric ophthalmic team refer aphakic and pseudophakic children to any or all appropriate key service providers for the visually impaired. This action will help to initiate and support ongoing dialogue and documentation among and between the medical and educational providers and service delivery models.
- *Education*: It is important that children receive clinical low vision evaluations along with instruction in visual efficiency with the prescribed low vision devices. This will help to determine the most beneficial and practical low vision devices and interventions that should be provided and implemented in the educational setting.
- *Economics*: It is important that the local, state, and federal funding sources address the needs of the 0 to 18 years of age low vision population. To emphasize the importance of the prescribed low vision devices (optical and nonoptical) and recommended intervention, these will need to be listed on the IFSP and the Individualized and Educational Program (IEP) so that they are provided the prescribed devices and recommended services.
- *Research*: It is important that research studies be conducted that address the effectiveness of practice protocols for children with low vision. Coordination of the educational and medical models will help to achieve optimal outcomes for children with low vision.

An example of such a coordinated research effort is the Project Magnify vision outreach program, which is sponsored by the South Carolina School for the Deaf and Blind and the South Carolina Department of Education Office of Exceptional Children. This program has been carried out in cooperation with the Storm Eye Institute at the Medical University of South Carolina. The Storm Eye Institute Low Vision Rehabilitation team provides services designed to increase visual reading skills through intensive training of prescribed low vision devices.

The goals of Project Magnify are to

- Increase student reading performance
- Increase access to appropriate optical aids
- Increase instructors' competencies in teaching the use of low vision aids
- Determine students' best reading method and reading media

The process of Project Magnify is to

- Conduct reading performance assessments by the educational professionals (teacher of the visually impaired [TVI], rehabilitation teacher [RT], and certification for low vision therapist [CLVT])
- Provide clinical low vision evaluations by the medical professionals (low vision specialists, optometrists and occupational therapist)
- Provide prescribed low vision aids
- Document reading progress using the prescribed low vision aids

Outcomes of Project Magnify: Students made excellent documented improvement in their reading abilities using the prescribed low vision devices along with low vision rehabilitation. In addition to improved literacy, students gained a greater independence and confidence in school, home, and community activities. The expertise of these educational and medical models ensured the successful outcomes of Project Magnify, which has led to the extension of funding for this low vision collaborative intervention model.

THE LOW VISION REHABILITATION TEAM—MEDICAL MODEL

The rehabilitation process works best when the treating ophthalmologist, the low vision optometrist, and the vision rehabilitation occupational therapist function as a coordinated team.⁵

- The *ophthalmologist* specializes in the medical and surgical care of the eyes and visual systems. In the pediatric cataract population, this care will address not only the removal of the cataract and placement of an intraocular lens when appropriate but also all comorbidities (microphthalmia, sensory nystagmus, strabismus, glaucoma, retinal detachment, high myopia) or systemic/metabolic conditions that may affect the ultimate visual outcome of cataract surgery. The ophthalmologist is critical in the determination and assignment of a medical diagnosis. The medical diagnosis is critical not only for medical insurance coverage but also for the justification of support services in the age appropriate educational setting (early intervention: 0–3 years; preschool: 3–5 years; school age: 5–18 years; transitions programs: 16–21 years).
- The *optometrist* with a specialty in low vision rehabilitation specializes in the assessment of the child's ability to focus and coordinate eyes for near, intermediate, and distant viewing. They prescribe eyeglasses, contacts, low vision devices, and low vision rehabilitation therapy.
- The *occupational therapist* is a rehabilitation professional who administers functional assessments and

provides appropriate intervention strategies and bills the appropriate insurances for the rendered low vision rehabilitation intervention. The specialty area of low vision has been emerging for the field of occupational therapy. The American Occupational Therapy Association offers a Specialty Certification for Low Vision (SCLV). The Academy for Certification of Vision Rehabilitation and Education Professionals also offers a CLVT, but it does not require that this be a registered licensed occupational therapist. At this point in time, only the registered, licensed occupational therapist can bill insurances (Medicaid, private insurances, Medicare, etc.) for low vision rehabilitation services.

OCCUPATIONAL THERAPY LOW VISION REHABILITATION

A registered and licensed occupational therapist (OTR or OTR/L) will generally receive a referral from the ophthalmologist or low vision optometrist to perform a low vision assessment and provide occupational therapy services as indicated. Once the OTR has completed the assessment, the OTR will develop a plan of care, which addresses the patient's goals, and this will be reviewed and signed by the referring doctor. Any changes to the plan of care must be reviewed by the referring doctor. Treatment sessions are provided in a certification period, which is typically up to 90 days. Ongoing progress notes are maintained by the OTR that address (SOAP)

- **Subjective:** response of the patient
- **Objective:** overview/summary of the session's therapeutic intervention strategies
- **Assessment:** functional outcomes of the session and progress toward patient goals
- **Plan:** actions to be considered prior to or for the next treatment session

If there is good rehabilitation potential and the patient has made good progress on the initial goals and additional or upgraded goals, the OTR can submit a recertification plan of care, which documents the justification for continuation of services. This recertification must be reviewed and signed by the referring doctor. Once the patient meets the established goals, the OTR will write a discharge summary, which is also reviewed and signed off by the referring doctor if in agreement with the discharge. Note that a patient may be discharged at any time if the patient is noncompliant or is not benefitting from the intervention. For insurance billing purposes, the occupational therapist will code using the visual impairment and medical diagnoses, as determined by the ophthalmologist and/or low vision optometrist. The visual impairment is based on the combined acuity of both eyes. Per the ICD-9, to qualify for low vision, the patient's vision must

correspond to visual impairment codes ranging from total impairment (no light perception [NLP]) to moderate impairment (20/70, incorporating contrast sensitivity or <20 degrees of visual field). Disorders of contrast sensitivity can be incorporated and may allow patients with better visual acuity than 20/70 to qualify for rehabilitation. In addition, those young or developmentally delayed children with uncertain or untestable visual acuity may qualify when the ophthalmologist states that the child “functions at the level of blindness” or is at a high risk of becoming legally blind. The occupational therapist will bill for therapeutic services using the following current procedural terminology (CPT) codes: Occupational Therapy Evaluation (97003), Therapeutic Exercises (97110), Therapeutic Activities (97530), Developmental Cognitive Skills (97532), Sensory Integrative Techniques (97533), Self-Care Management (97535), and Community Re-integration (97537).

THE OCCUPATIONAL THERAPY LOW VISION PEDIATRIC EVALUATION

The OTR will need to take into account the findings of the ophthalmologist and low vision optometrist. These findings may include but are not limited to visual acuity (near, intermediate, distant), contrast sensitivity, light/dark adaptability, photosensitivity, glare interference, night blindness, color perception, visual field loss, binocularity, stereopsis, strabismus measurements, and level of amblyopia. Oculomotor behaviors to be evaluated by the low vision occupational therapist would include fixation, convergence and divergence, tracing tracking, shift of gaze, and scanning. Other visual processing skills to be assessed would include visual perception, depth perception, figure-ground perception, spatial orientation visual sequencing, and visual closure. If reading is an expectation, print size, reading rate, and comprehension will need to be accessed. Also, handwriting and keyboard skills will need to be observed.

With consideration of these findings, the OTR will gather additional information from the child, parents, caregivers, and the key service providers to determine the child's strengths, limitations, interests, motivating triggers, requirements (academic), and goals. Depending on the age of the infant/child/adolescent and his/her cognitive status, the following information should be included: background information including developmental history, medical history, musculoskeletal, neurological, sensory processing, and integration. This would include assessment of the senses (tactile, proprioceptive, vestibular, auditory, gustatory, olfactory) as well as fine and gross motor skills, play and/or academic skills, ADLs (as relate to the child's developmental age), and behavior.

It is important to determine the rehabilitation potential for this infant/child/adolescent with regard to the low vision rehabilitation intervention in order to set appropriate

and realistic goals. It is imperative for the infant/child/adolescent that their support system be involved in the determination of the goals since they will play a critical role in helping to achieve the goals. The therapeutic goals may be related to or incorporated into those that appear on the infant's IFSP or the child/adolescent's IEP.

The plan of care developed by the low vision occupational therapist needs to include the recommended low vision devices (optical and nonoptical), assistive technology, positioning equipment, and compensatory measures that would benefit the infant/child/adolescent in the home, school, and community settings. Depending on the age of the child and funding sources, different low vision devices and assistive technology will need to be considered. The low vision devices may include but are not limited to prescription glasses or contacts, handheld magnifiers, stand magnifiers, telescopes, electronic magnifiers, computer adaptations, and other electronic accessible devices and absorptive filters addressing light sensitivities (Fig. 54.1).

The assistive technology can include more advanced SMART technology that provides text to voice and voice to text technology. With the importance and emphasis on accessibility, there is exciting new technology being developed or improved upon, so it is important to continually explore and consider the benefits of this new technology. Positioning equipment is important for an infant/child/adolescent that is multiply handicapped who



Figure 54.1. Boy with magnifier quoting “I can see the words better now!”

has musculoskeletal and/or postural challenges. Without proper body positioning for optimal eye alignment, visual tasks and outcomes will be compromised. Other compensatory measures to take into account could include such things as seat location in the classroom, using specific font and size for printed material, recommending specific lighting for optimal performance, closing the blinds to reduce glare, and the use of good contrast colors and materials (Figs. 54.2 and 54.3).

OCCUPATIONAL THERAPY LOW VISION PEDIATRIC INTERVENTION

Occupational therapy is a profession concerned with promoting health and well-being through engagement in occupation as defined by the World Federation of Occupational Therapists.⁶ It is a discipline aimed at promoting health and well-being by enabling the individual's performance necessary for meaningful and purposeful activities as defined by the American Occupational Therapy Association.⁷ The occupational therapist will perform a task analysis of the functional ADLs and instrumental ADLs for age/developmentally appropriate activities with consideration not only for low vision but also for physical, environmental, psychosocial, mental, spiritual, and cultural factors that may create barriers.



Figure 54.2. Use of contrast in daily activity. Milk in white versus green cup.



Figure 54.3. Providing difference in contrast: stuffed white kitty on beige chair (A) versus red chair (B).

With regard to low vision, it is imperative to have an awareness of normal visual development.

The scale presented in Table 54.2 provides a guide for age-appropriate visual development.⁸

Low vision rehabilitation emphasizes functional age/developmentally appropriate visual skills. To accomplish this, emphasis should be placed on visual stimulation, visual efficiency, and visual utilization.⁹

- Visual stimulation encourages visual skills by attending to light sources, single object, rudimentary tracking, shifting gaze, and reaching for an object.

- Visual efficiency helps the child to discriminate and differentiate lines, patterns, intensity, and the transition from concrete forms to symbolic forms.
- Visual utilization teaches the child to accommodate and/or modify the environment, which may include the incorporation of low vision devices and assistive technology. Literacy is emphasized in low vision rehabilitation since it is a critical and necessary developmental skill needed to be able to perform many ADLs. Literacy has three levels⁹: emergent literacy, academic literacy, and functional literacy.

Table 54.2 VISUAL DEVELOPMENTAL SCALE⁸

Age	Visual Characteristic or Skills	Age	Visual Characteristic or Skills
Birth	Can see patterns of light and dark, but specific objects blurry Has some degree of fixation	9–10 months	Imitates expressions Looks around corners Spills, to watch liquid spill Is visually alert to new things Plays games Develops object permanence
1 month	Can focus eyes at 1½ inches Displays beginning of conjugate following movements (binocular) Follows slow moving objects Follows horizontal movement of objects to the midline	1 year	Has distance and near visual acuity Displays improved binocular vision Displays improved accommodation
2 months	Displays developmental protective blink reflex Prefers faces to complex patterns Follows vertical movements of objects	1½ years	Displays vertical orientation: builds 2–3 block towers Matches identical objects: two spoons, two blocks, etc. Points to pictures in book Imitates vertical and horizontal strokes
3 months	Displays smoother eye movements Smiles at visual stimuli Displays improving visual acuity Displays improving binocular vision Notices gross color differences Seems aware of objects only when manipulating them Anticipates feeding by visual stimuli	2 years	Inspects objects with eyes alone Imitates movements Visually seeks missing objects or person Has increased color vision Has increased visual memory
4 months	Displays accommodative flexibility (shift focus) Displays improved eye–hand coordination Shows interest in small, bright objects Attempts to move toward objects in visual field Recognizes familiar faces Visually explores new environments Follows objects across the middling movements Shows horizontal, vertical, and circular eye movements Makes unsuccessful attempts at reaching mouth and looks at objects in hand	3 years	Matches simple forms: simple form board but relies on tactile cues Pretends to pick up objects from the page of a book Able to draw a crude circle
5 months	Develops eye–hand coordination Grasps objects successfully Looks intently at objects held close to eyes Examines objects with the eyes, rather than using objects for play	4 years	Able to accurately discriminate sizes Has accurate depth perception Displays automatic eye–hand coordination Discriminates length regardless of orientation
6 months	Shifts visual attention from one object to another in a field of vision Recognizes faces up to 6 feet away Retrieves toys dropped within reach Turns objects in hand and explores visually Has capacity of both eyes holding fixation and convergence equally	5 years	Displays mature coordination: precise pick up and release Colors, cuts, and pastes Demonstrates knowledge of concept and muscle control for assembling Able to draw a square
		6 years	Handles and attempts to use tools and materials Prints capital letters but reversals are commonplace Able to draw a triangle Begins to read
		7–9 years	Prints sentences Has speed and smoothness of eye–hand coordination Includes details in drawings

- Emergent literacy refers to the young child's developing ability for reading and writing activities.
- Academic literacy refers to the type of reading and writing mastery skills that are learned in the formal school years.
- Functional literacy relates to reading activities for practical tasks such as reading a road sign or menu.

Appropriate exposure to stimuli and activities with the introduction and training of prescribed optical devices and/or other low vision devices is essential for optimal literacy development. Interventions may include helping the child adjust to lenses that may include prisms prescribed by the low vision optometrist. Performing functional tasks with occlusion of vision is a common practice for the pediatric low vision population. Amblyopia may occur in developmental disabilities and is not an uncommon occurrence after pediatric cataract surgery. Traditional amblyopia therapy intervention requires the patching of the good eye for a specified period of time each day in order to force the weaker eye to be used.¹⁰ Determination of appropriate filters for indoor and outdoor use that address light sensitivity and glare interference need to be identified for specific functional tasks. Fine motor and ocular motor skills are independent but are also interdependent skill sets. Ocular motor intervention emphasizes visual attention, saccadic fixations, smooth scanning, and efficient pursuits. Typical therapeutic activities that should be provided for the visually impaired pediatric population include fixation, location, eye tracking, smooth pursuits, scanning, peripheral awareness, contrast awareness, eccentric viewing, and management of their visual field.

Therapeutic intervention often includes teaching the effective implementation of low vision devices including but not limited to nonoptical devices, magnifying spectacles, prisms, minus lenses, telescopes, reverse telescopes, microscopes, head loops, hand magnifiers, stand magnifiers (including dome magnifiers), electronic–optical magnifiers (CCTV), computer magnification, touch-screen tablet computer devices, and new/emerging technical devices.

SUMMARY

- It is important for the ophthalmologist to advocate for optimal services and low vision devices and technology in order for the child to attain optimal outcomes.
- It is essential for the ophthalmologist to be proactive in making referrals to the low vision rehabilitation team in order to better identify beneficial devices and intervention.

Table 54.3 LOW VISION REHABILITATION PEDIATRIC RESOURCES

Journals

Journal of Visual Impairment and Blindness

AccessWorld: Journal of Technology for Consumers with Visual Impairments

Infant–Toddler Intervention: A Transdisciplinary Journal

Infant and Young Children: A Transdisciplinary Journal of Special Care Practices

Journal of Early Intervention

Young Exceptional Children

Zero to Three Bulletin

Low Vision Device and Assistive Technology Resources^a

American Printing House for the Blind (APH) www.aph.org

LS & S Group www.lssgroup.com

Maxi-Aids www.maxiaids.com

Enhanced Vision Systems www.enhancedvision.com

Eschenbach Optik of America www.eschenbach.com

Innovations www.magnicam.com

Optelec USA www.optelec.com

Information and Support Services

NICHCY: National Dissemination Center for Children and Disabilities <http://nichcy.org/>

National Association for Parents of Children with Visual Impairments (NAPVI) www.spedex.com/napvi/

Low Vision Information Center www.lowvisioninfo.org

The Low Vision Network <http://www.lowvision-project.org>

National Eye Institute www.nei.nih.gov/

National Organization for Albinism or Hypo pigmentation (NOAH) www.albinism.org

National Organization of Parents of Blind Children (NOPBC) <https://www.nfb.org/>

American Foundation for the Blind www.afb.org

Council for Exceptional Children (CEC) www.cec.sped.org

Professional Organizations

American Academy of Ophthalmology www.aao.org and www.eyenet.org

American Academy of Pediatric Ophthalmology and Strabismus www.aapos.org

Association for Education and Rehabilitation of the Blind and Visually Impaired (AER) www.aerbvi.org

American Occupational Therapy Association (AOTA) www.aota.org

Academy for Certification of Vision Rehabilitation and Education Professionals (ACVREP) www.acvrep.org

^aAll Web sites valid as of January 4, 2013.

- It is imperative for the ophthalmologist and the other professionals to communicate with the other key service providers of the medical and educational model teams.
- The ophthalmologist treating visually impaired children should have ongoing documented communication with the child's family and/or caregivers as well as the key service providers.

Table 54.3 contains resources for low vision rehabilitation from the educational (journals) and technologic realms, as well as governmental and professional information and support services

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Future Direction

M. Edward Wilson and Rupal H. Trivedi

In the years to come, pediatric cataract surgery will evolve along its own unique path. Surgeons who operate on children are, by their very nature, more conservative since they know that children will endure the consequences of their treatments over a long life. However, innovation still occurs, often in response to the unique difficulties a surgeon faces when operating on a growing eye attached to a less than fully cooperative and energetic child. Pediatric surgeons strive to get the best results with the least surgical trauma and the fewest trips to the operating room. This continues to be a challenge. A more vigorous inflammatory response combined with variable postoperative medication compliance will continue to push us into the more frequent use of intracameral agents at the time of surgery. In the future, we will move as close as we can to eliminating the need for postoperative medications. Intracameral antibiotics will become routine for every intraocular case, and the use of intracameral dexamethasone or triamcinolone will be commonplace. Povidone-iodine will continue to be placed on the eye before and after every surgery but in a much more diluted form than commonly used today. Evidence is mounting that the effectiveness of povidone-iodine is not diminished when it is diluted down to 2.5% or even lower but the risk of chemical conjunctivitis as a complication is reduced markedly.

When vitrectomy and posterior capsulectomy were developed in the 1970s, it rapidly became a standard for infantile cataract surgery. Surgeons feared that cystoid macular edema and retinal detachment rates would soar, but outcomes without posterior capsulectomy and vitrectomy were so poor that taking those risks was warranted.

This was a uniquely pediatric problem, and it was met with a uniquely pediatric innovation. Today, the advances in vitreoretinal surgery have also benefited the pediatric cataract surgeon. Venturi pump technology has made vitrectorhexis and pediatric lens aspiration easier. High-speed cutters have made vitrectomy safer with less risk of traction on the retina. We will all move to >1,000 cuts per minute and smaller gauge handpieces. These smaller handpieces are already improved over the initial models and are now more capable of the unusual tasks we ask of these instruments (membrane and plaque removal, etc.). While manual posterior capsulorhexis without vitrectomy will gain in popularity for children above age 4, vitrectomy and posterior capsulectomy will remain the standard for babies and toddlers. In the future, more surgeons will adopt the pars plana/plicata approach when a vitrectomy is chosen and an intraocular lens (IOL) is being placed.

For babies operated in the first 6 months of life, the least traumatic surgery to clear the visual axis will continue to be a bimanual, closed chamber, microsurgical approach that includes a primary posterior capsulectomy and anterior vitrectomy and no IOL. The Infant Aphakia Treatment Study's 5-year results will be available soon, and we expect that these reports will echo what the 1-year outcomes found for visual acuity. We predict that the contact lens and IOL treatment groups will continue to show no significant differences in best-corrected visual acuity. However, in these 5-year results, we will see very clearly how poor we are at predicting the growth rate and refractive change in these eyes when they are implanted primarily. There will be many IOL exchange

surgeries needed, as the children get older. Also, the reoperation rate after cataract surgery in the first 6 months of life is too high. This is especially true after primary IOL implantation, but, to a lesser degree, it is still true when no IOL is placed. Amazingly, when surgery for a childhood cataract is needed after the first birthday, these reoperations are very uncommon even when an IOL is implanted. The reepithelialization of lens cortex and the formation of pupillary membranes are particularly prevalent when surgery is done on microphthalmic eyes in the first 6 months of life. Newer approaches will be used in the future. This is another situation where innovation is needed to help solve a uniquely pediatric problem. We can start by using more pupil expanders such as iris hooks and pupil expansion rings when operating on babies whose eyes do not dilate well. Complete visualization of the lens equator will help the surgeon remove the lens cortical cells more completely. Small gauge, tapered, and curved bimanual instruments fit into tight-fit incisions will help. Having the ability to switch hands allows the aspiration handpiece to access the lens equator from two nearly opposite entry points. In these small babies, rapid lens growth means that the lens cells are very actively producing lens fibers. Residual cells left behind at surgery have more of a negative consequence than they do when surgery is performed in older children. Therefore, we need to get better at removing them. In these small babies with poorly dilating pupils, cortex removal can be difficult. The elastic capsular bag equator does not resist the aspiration force as well as the older pediatric capsule does, making striping of the equatorial fibers less complete unless the material is actively aspirated by placing the aspiration handpiece further into the capsular equator. In the future, along with pupil expanders like hooks and rings, we will get better at using sequential hydrodissection and viscodissection along with improved aspiration techniques to raise the standard for the completeness of lens fiber and lens cell removal at surgery. Efficient fluid pumps available on Venturi pump machines will aid in this transition by better stabilizing the anterior chamber of these soft eyes compared with gravity-fed irrigation systems that are inadequate for these higher standards. More complete cortical cleanup must be accomplished without adding trauma to the procedure. These eyes need to remain as uninflamed after surgery as possible, equal to what we can achieve now when the pupil dilates well and iris manipulation is avoided.

In the future, IOL implantation will become even less common in the first 6 months than it is now. This is ironic since IOL implantation as a primary procedure is uniformly accepted after the first or at least after the second birthday. The problems with IOL implantation in the first 6 months include more surgical trauma, more reoperations, and the impossible task of selecting an IOL power. Even if power could be selected properly, the need for

high hypermetropic glasses or temporary piggybacking of IOLs presents more complexity. While we are getting much more precise at predicting eye growth and calculating what the refraction will be when we operate later in childhood, these predictions are woefully inaccurate in early infancy. Not every family in the United States can manage or afford contact lenses for aphakia in infancy. For now, IOL implantation (single lens or piggyback) may appear to be the only viable option, especially when the cataract is unilateral. In the future, we need to make aphakic contact lenses (silicone or rigid gas permeable) more affordable and more available, even in remote rural areas of the United States and in the developing world. This may seem an unrealistic amount of change for this small population of patients, but it can be done. With secondary IOL implantation at an age older than 2 years, we have a fighting chance when it comes to the task of selecting an IOL power that will not leave huge amounts of hypermetropia at the time of implantation and will not lead to more than a modest amount of myopia at age 20 years. These children need to be able to function, even when their glasses are broken, as successful schoolchildren when they are young and as working adults later—a daunting task indeed. To that point, we have been surprised and impressed with how well children function (and read the eye chart) when they have mild to moderate (1–4 D) residual uncorrected hypermetropia after IOL implantation. No one would predict that they would see and function so well. It is only partially due to depth of focus from a small pupil aperture. Some additional form of pseudoaccommodation must be in play. It is ideal if we can keep all of our IOL-implanted patients (toddlers to teenagers) in a residual refractive error zone from +3 to –3 diopters.

In the early days of infantile posterior capsulectomy and vitrectomy, the goal was to remove as much as possible of the capsular bag. I (M.E.W.) recall, during my fellowship, watching Marshall Parks even use scleral depression during vitrector-assisted cataract surgery in order to see and thus remove as much of the capsular bag equator as possible. Reoperations, ironically, were lower than they are now. Removal of all of the lens equatorial epithelial cells eliminates the risk of reepithelialized lens cortex causing visual axis reopacification. Removal of all capsular bag remnants also prevents synechia formation by removing the structure that the iris sticks too when synechia form. The surgeons in those days were not preparing for secondary IOL implantation later. Lifelong aphakia was the accepted norm for these babies. Keeping a clear visual axis was the goal. We now attempt to leave enough residual lens capsule so that secondary IOL implantation can be accomplished easily. We prefer that the matching anterior and posterior capsulectomy openings to be no more than 4.5 to 5 mm so that in-the-bag IOL implantation can be accomplished. However, leaving this much capsule means that there is a much higher chance of synechia formation

and secondary visual axis opacification. In the future, this need for more capsular remnant needs to be accompanied (at least in infancy) with a higher standard of complete lens cortex removal (as discussed above). We have moved almost full circle from high reoperation rates in babies operated before the routine use of the vitrector to very low reoperation rates after the vitrector became the standard and now back to higher reoperation rates again with the advent of residual capsule and primary IOL implantation for infants operated very early in life. In the future, when babies are operated in the first half of the first year of life, we will perform better cortical cleanup and utilize fewer in-the-bag primary IOLs. When IOLs are necessary in early infancy, we predict that sulcus placement will become more popular and accepted than it is today since it allows the capsular bag to seal like it does in the aphakic child, and it places the IOL in a location where the inevitable IOL exchange will be more easily accomplished. We also predict that well-designed lenses selected specifically for sulcus use will not cause iris chafing and will not scar in place or cause any increased inflammation.

For children beyond the toddler ages, we will all become more adept at performing high-quality cataract surgery, including a predictably sized posterior capsulorhexis, without disturbing the hyaloid face of the vitreous body. Low flow and slow motion techniques with refined uses of ophthalmic viscosurgical devices and staining techniques will help make this possible. The formed vitreous and the relatively resistant vitreous face improve the likelihood that this procedure can be accomplished predictably. Again, meticulous cortical lens epithelial cell removal is essential since the vitreous face can serve as a scaffold across which proliferating cells can migrate. Innovative designs like the bag-in-the-lens IOL will gain popularity in children beyond the toddler age since these innovative designs help to avoid any contact of lens epithelial cells with the intact vitreous face.

Another future advancement will be in IOL technology. Truly accommodative IOLs will eventually become popular for pediatric implantation but only when they are able to achieve significant ranges of accommodation. Multifocal IOLs will remain mostly as a niche

premium product used in adults and some teenagers. The multifocal simultaneous vision concept has too many flaws to become a preferred product for the growing eye. As the eye of a child grows, a myopic shift is inevitable, and multifocal IOLs do not perform well when the eye becomes myopic. We predict that historically, multifocality will be remembered as a temporary stop on the road to truly accommodating IOLs.

Predicting axial growth, and the refractive changes that accompany it, is another of the major remaining challenges for the long-term care of children after cataract surgery. In the coming years, we will have a better understanding of eye growth after cataract surgery, which can help us to develop formulas specifically designed for pediatric eyes. We may also have a better understanding of amblyopia management in aphakic/pseudophakic eyes to help us provide better functional outcome to these children. In addition to further knowledge about how much and how often patching needs to be done, ways to expand the plasticity of the visual system will be further defined. Successful treatment of amblyopia well beyond the age of “visual maturation” may require complex pharmacotherapy. Initial attempts have provided proof of concept, and promising progress is expected in coming years.

Overall, we will continue to benefit from the constant advances in the field of adult cataract surgery by adopting those innovations that have applicability for children. As discussed above, we will also change what we do in response to our specific pediatric needs. Better outcomes are a mandate. We must innovate without denying our cautious and conservative nature. Our patients are different than adults, more vulnerable and ever changing as they grow and develop. We cannot be quick to adopt untested or unproven methods, but we also cannot accept a status quo that produces a less than optimal outcome. This balance is uniquely complex for us as pediatric surgeons. As history has shown us, we get it right more often than not, and now that worldwide communication and consensus building among thought leaders in this field are at an all-time high, our prospects for future progress are excellent.



Note: Page numbers followed by “P” denotes figures and those followed by “t” denotes tables.

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