Pediatric Cataract Surgery and IOL Implantation

A Case-Based Guide

Courtney L. Kraus
Editor

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Foreword

Cataracts in children are common enough so that every pediatric ophthalmologist sees them and yet they are rare enough to create anxiety for the clinician who is trying to stay up on all of the latest evidence-based trends. Properly timed and expertly performed cataract surgery can bring sight to a child who would otherwise be blind. When the number of blind-years prevented is taken into account, pediatric cataract surgery may be the most cost-effective procedure in all of ophthalmology. However, unlike cataract surgery in an elderly adult, lens replacement in childhood removes youthful accommodation and derails emmetropization by removing the eye's natural offset to axial growth of the globe. As we commonly say: "Kids are not just small adults."

In this important book, Dr. Courtney Kraus has assembled an impressive group of experts and has chosen a case-based approach as an efficient and effective way to transfer knowledge. It should be on the required reading list of every pediatric ophthalmology fellowship program and every clinician who operates on children.

In Part I: Approach to Lens Opacities, the readers will be able to refresh their knowledge about how to think simultaneously like a developmentalist, a geneticist, and a surgeon. Cataracts can appear at any age and in an endless variety of forms. For some visually insignificant opacities, surgery is not recommended but change is always possible as the child grows. Illustrative cases are used to guide our recognition of random unilateral developmental missteps, familial bilateral progressive opacities, and cataracts secondary to systemic diseases and disorders. Pattern recognition is emphasized and a directed specific workup is preferred over a "shotgun" approach.

In Part II: Surgery, the readers will enjoy the practical step-by-step guidance that is provided for the surgical procedure, the implant power decisions, and the care of the child's eye during healing. Surgeons who operate on children and adults will be able to fully understand the stark differences between the needed steps in children compared to what is commonly done in adults. In Part III: Correcting Surgically-Induced Aphakia, 15 chapters are presented to guide surgeons through complex settings and associations. These chapters describe the pros and cons of the many

options we have when faced with a decision about what is best for an individual child or an unusual situation.

I congratulate Dr. Kraus for assembling this guide that so successfully presents a complex topic in a format that is so easy to read and understand. The children are the future and the future for children with cataracts is brighter when we are all informed and when we adopt new innovations while still remaining cautious and careful. In short, we treat our patients the way we would want our own children to be treated.

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Preface

The presence of a lens opacification in childhood requires special considerations and nuanced care. For those ophthalmic surgeons working to identify and treat pediatric cataracts, surgical technique has progressed dramatically over the last decade with advancements in surgical tools and diagnostic devices. Through this text, the latest technology in lens implantation will be presented, but it remains screening, prompt identification, and consistent follow-up and amblyopia management that are the real determinants of visual acuity outcomes.

The goal of this textbook is to give the reader an overview of pediatric cataract surgery, with a focus on the use of intraocular lenses. By using the format of a case-based guide, it is hoped that the reader will consult each chapter as those unique and complex patients present themselves. Furthermore, for those still in training or looking to complement their education, the systematic progression through case scenarios will ideally form a strong curriculum with which to supplement those cases encountered in clinic and the operating room.

Baltimore, MD, USA — Courtney L. Kraus

Acknowledgments

This book represents the culmination of the collective efforts of 47 authors, some at the very beginning of their careers in ophthalmology and others with decades of experience. They span a multitude of institutions, academic and private practice, four countries, and three continents. I consider many dear friends and all remarkable authorities on the topics presented. To each individual, who took time out of a busy practice or training experience, time away from family and friends, and instead focused on authoring a carefully researched and thoughtfully presented chapter, I am tremendously grateful. Because of the great variety in backgrounds, quality of content, and clarity in the provided expertise, I truly believe this book can sit alongside many of the great texts in pediatric ophthalmology. It will hopefully offer trainee and established surgeon pearls, tips, and additional insight.

As I now have the privileged position to provide advice and guidance to my own trainees, I think my most oft-repeated advice when asked about fellowship is to seek out your mentor. Surgical fellowship is one of the last true apprenticeships, where master craftsman imparts upon novice the subtleties and wisdom that sets his/her practice apart from the next. In doing so, friendship and respect form, and for many, a lasting bond. It has been a great privilege to call Dr. M. Edward Wilson my mentor. In my time in fellowship, and through continued advice and guidance in the years that have followed, I have truly benefited from his knowledge, skill, mastery, care, and respect. For this I am tremendously grateful. I must also extend my thanks to the others that mentored me in my career in pediatric ophthalmology: Drs. Michael Repka, David Guyton, Edward Cheeseman, Richard Saunders, M. Millicent Peterseim, Lawrence Tychsen, Gregg Lueder, and especially Susan Culican.

I would also like to specifically thank Drs. Brita Rook and Jennifer Davidson. Co-fellows are often very special partners in crime while in training, but these two have become two of my greatest friends, and trusted advisors, in all things eye- and non-eye-related. There are too many friends along my career, be it in undergraduate, medical school, residency, fellowship, or beyond to call out by name. But to each, I promise to find and share a hug of appreciation and celebration the next time we see each other at a meeting.

And finally, to the people without whom I could never be in the fortunate position I am today – my family. To my newest family, the Desais, may we all be so fortunate to have in-laws as caring and full of love as you. You celebrated my work on this book and its completion. To my parents – where to begin – I really cannot imagine two more supportive people. Your love, understanding, and unconditional support started from the beginning and never wavered. Mom, you reviewed every paper I wrote for years, and for that, I spared you having to proofread these 25 chapters. And Dad, your hard work and attention to detail are attributes I am still trying to master, but I called on both many a time as this project neared completion. And to my brother Alex, thanks for keeping me in the loop on Duke basketball scores when time would not allow me to catch games in full.

To my person, my husband, Shaun Desai, thank you for being you. You are my biggest cheerleader and best friend. You know more about pediatric cataracts than some who may consider reading this book. And this is because you know to love me means to love what I love. You never questioned that this project was within my reach and pushed and supported me right to the end. Thank you for walking me outside of my comfort zone and holding my hand the entire way.

Finally, the preliminary discussions outlining this book first came about when my daughter was 10 weeks old. She is now 19 months. To Isla, your first year and half of life was delightful, joyful, funny, and all too quick. I will treasure this book as the other creation that grew to life over the course of the last year.

Thank you everyone.

Courtney L. Kraus

Overview of Structure

This book is divided into four parts.

- *Part I: Approach to Lens Opacities* provides an overview of lens opacities in childhood, providing a framework for evaluation, nonsurgical management, and preoperative planning. Here, the basics differentiating unilateral from bilateral cataracts, as well as epidemiology and genetics, are reviewed. Preoperative examination and counseling is reviewed.
- *Part II: Surgery* gives the reader an overview of surgical steps, IOL calculations, and postoperative considerations. Case-based presentations assist in illustrating surgical techniques and IOL selection.
- *Part III: Correcting Surgically Induced Aphakia* remains critical for visual rehabilitation following cataract surgery. Options include aphakic glasses, contact lenses, and intraocular lens implantation. The later chapters in this section present unique approaches and in-depth discussion of IOL implantation. Less common lens selections, such as multifocal and accommodative IOLs, and techniques, such as poly-pseudophakia and IOL exchange, are presented.
- *Part IV: Special Considerations in IOL Implantation* concludes the textbook. Within this section, seven situations requiring careful preoperative planning, innovative surgical technique, and special postoperative care are reviewed. Standing alone, this section provides a quick consultation for challenging cases.

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Part I
Approach to Lens Opacities

Chapter 1
Congenital and Hereditary Cataracts: Epidemiology and Genetics

Nadav Shoshany, Fielding Hejtmancik, Alan Shiels, and Manuel B. Datiles III

The crystalline lens is a unique structure specialized in transmitting and focusing light onto the retina. Transparency is crucial for proper transmission of light and has to be preserved throughout life to ensure sustainability of visual function. Lens transparency occurs with the appropriate architecture of lens cells and tight packing of their proteins, resulting in a constant refractive index over distances approximating the wavelength of light [1, 2]. The refractive index of the human lens rises gradually from the cortex (1.38) to the nucleus (1.41), where there is an enrichment of tightly packed γ-crystallins.

Cataracts, which have multiple causes, are often associated with breakdown in the lens's microarchitecture [3, 4], vacuole formation, and resultant fluctuations in density causing scattering of light. A compromise in the short-range ordered packing of crystallins and a disturbance in their homogenous phase impair transparency and cause opacification. Opacification also accompanies the formation of high molecular weight aggregates, sized 1000 Å or more [5, 6].

Cataracts in the pediatric population raise a particular concern. Unlike age-related cataract, which, once treated, generally allows prompt visual rehabilitation, deferred removal of vision-impairing opacities during the first years of life causes

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amblyopia and interferes with normal cortical visual development, thereby limiting the visual potential of the involved eye. Given the genetic background of many pediatric cataracts, certain cases can be anticipated and diagnosed early to maximize a young individual's visual potential.

Epidemiology

Hereditary cataracts are estimated to account for between 8.3% and 25% of congenital cataracts, depending on the population and study [7–9]. Developing nations, with higher frequencies of environmental and infectious etiologies, naturally attribute a lower fraction of cataracts to inheritance, despite relatively constant mutation rate.

Inheritance patterns also vary due to marriage patterns in specific populations. While about 85% of inherited cataracts worldwide are autosomal dominant (see below), in Pakistan, which has a high rate of consanguineous marriages, about 87% of genetic cataracts are inherited as an autosomal recessive trait [10]. Similarly, it has been estimated that 71% of inherited congenital cataracts in Saudi Arabia are autosomal recessive [11].

Clinical Features and Classification of Congenital Cataracts

Human cataracts can be classified using a variety of characteristics such as their age of onset, etiology, location in the lens, size, pattern or shape, density, and rate of progression.

When classified by *age of onset*, cataracts visible within the first year of life are considered infantile or congenital, and later-onset (within the first decade) cataracts can be classified as juvenile. With congenital opacities, early onset generally implies greater amblyogenic risk and poorer visual prognosis, unless treated promptly. Occasionally, asymptomatic congenital opacities might be overlooked for years, thus deferring the age of diagnosis and obscuring the correct classification.

Etiology-based classification yields varying proportions of contributing factors. About 30% of congenital cataracts in developed countries have a genetic etiology, while many of the remainder are idiopathic. Intrauterine infections and trauma account for a small percentage [9], which increases considerably in less developed nations [12]. Congenital cataracts can be isolated (Table 1.1) or appear in conjunction with other ocular or systemic conditions, including craniofacial, renal, and musculoskeletal syndromes and metabolic diseases. With systemic disorders, bilateral cataracts are expected, although in many cases asymmetric progression can be observed.

Table 1.1 Loci, genes, and phenotypes for non-syndromic cataract

Gene	Inheritance	Associated extra-lenticular phenotypes	MIM no.	Gene/locus MIM no.	Locus
1. *Transcription and developmental factors*					
PITX3	AD	Anterior segment mesenchymal dysgenesis, microphthalmia, neurodevelopmental abnormalities	610,623	602,669	10q24.32
EPHA2	AD/AR	Susceptibility to age-related cortical cataract	116,600	176,946	1p36.13
HSF4	AD/AR		116,800	602,438	16q21
MAF	AD	With or without microcornea	610,202	177,075	16q22-q23
SIPA1L3	AR		616,851	616,655	19q13.1-13.2
NHS	X-linked	Nance-Horan (cataract-dental) syndrome	302,200	300,457	Xp22.13
2. *Lens crystallins*					
CRYGB	AD		615,188	123,670	2q34
CRYBA2	AD		115,900	600,836	2q34
CRYGC	AD	With or without microcornea	604,307	123,680	2q33.3
CRYGD	AD	With or without microcornea	115,700	123,690	2q33.3
CRYGS	AD		116,100	123,730	3q27.3
CRYAB	AD/AR	Myopathy, multiple types	613,763	123,590	11q22.3
CRYBA1	AD		600,881	123,610	17q11.2
CRYAA	AD/AR	With or without microcornea, susceptibility to age-related nuclear cataract	604,219	123,580	21q22.3
CRYBB2	AD	With or without microcornea	601,547	123,620	22q11.23
CRYBB3	AD/AR		609,741	123,630	22q11.23
CRYBB1	AD/AR		611,544	6,009,291	22q12.1
CRYBA4	AD		610,425	123,631	22q12.1
3. *Gap junction proteins (connexins)*					
GJA8	AD/AR	With or without microcornea	116,200	600,897	1q21.1
GJA3	AD		601,885	121,015	13q12.1
4. *Membranes and their proteins*					
WFS1	AD	Wolfram syndrome (DIDMOAD)	116,400	606,201	4p16.1
LEMD2	AR		212,500	616,312	6p21.31
AGK	AR	Sengers syndrome	614,691	610,345	7q34
MIP	AD		615,274	154,050	12q13.3
LIM2	AR		615,277	154,045	19q13.41
LSS	AR		616,509	600,909	21q22.3
5. *Beaded filament and other intermediate filament proteins*					
BFSP2	AD	Myopia	611,597	603,212	3q22.1
VIM	AD		116,300	193,060	10p13

(continued)

Table 1.1 (continued)

Gene	Inheritance	Associated extra-lenticular phenotypes	MIM no.	Gene/locus MIM no.	Locus
BFSP1	AR		611,391	603,307	20p12.1
6. *Chaperones and protein degradation*					
FYCO1	AR		610,019	607,182	3p21.31
UNC45B	AD		616,279	611,220	17q12
CHMP4B	AD		605,387	610,897	20q11.21
7. *Other genes and pathways*					
TDRD7	AR		613,887	611,258	9q22.33
GCNT2	AR	Adult i blood group phenotype	110,800	600,429	6p24
8. *Unknown loci*					
?	AD		115,665	NA	1pter-p36.13
?	AR	With or without microcornea	612,968	NA	1p34.3-p32.2
?	AD		115,800	NA	2pter-p24
?	AD		607,304	NA	2p12
?	?	Susceptibility to age-related cortical cataract	609,026	NA	6p12-q12
?	AR		605,749	NA	9q13-q22
?	AD		614,422	NA	12q24.2-q24.3
?	AD		115,650%	NA	14q22-q23
?	AD		605,728	NA	15q21-q22
?	AD		601,202	NA	17p13
?	AD		115,660	NA	17q24
?	AR		609,376	NA	19q13

Further information and references can be found at Cat-Map: https://cat-map.wustl.edu/

Perhaps most usefully, cataracts can be classified based on their *appearance and anatomic location in the lens*. Based on lens development, the location of a lens opacity can suggest the time at which the pathology initiated and, at times, suggest the genetic cause of the cataract. The most commonly used system is that described by Merin [13], in which the cataract is classified as zonular (indicating zones or locations in the lens, including nuclear, lamellar, and sutural), polar (including anterior or posterior), capsular or membranous, and total (mature or complete).

Consistent with ocular embryonic development, *nuclear opacities* can be localized to the embryonic (months 1–3), fetal (months 3–9), or infantile (postnatal) nucleus (Fig. 1.1a, b) and are likely to result from mutations in genes active during these periods. The opacifications can vary in severity – from fine, pulverulent opacities with minimal visual impact to large, dense, vision blocking ones that require prompt surgical removal. *Lamellar* cataracts (Fig. 1.1c, d) affect concurrently formed lens fibers, resulting in a shell-like opacity. They are the most common type of congenital cataract and can be caused by a wide variety of genes (Table 1.2). Some have associated arcuate opacities within the cortex called cortical riders (Fig. 1.1d).

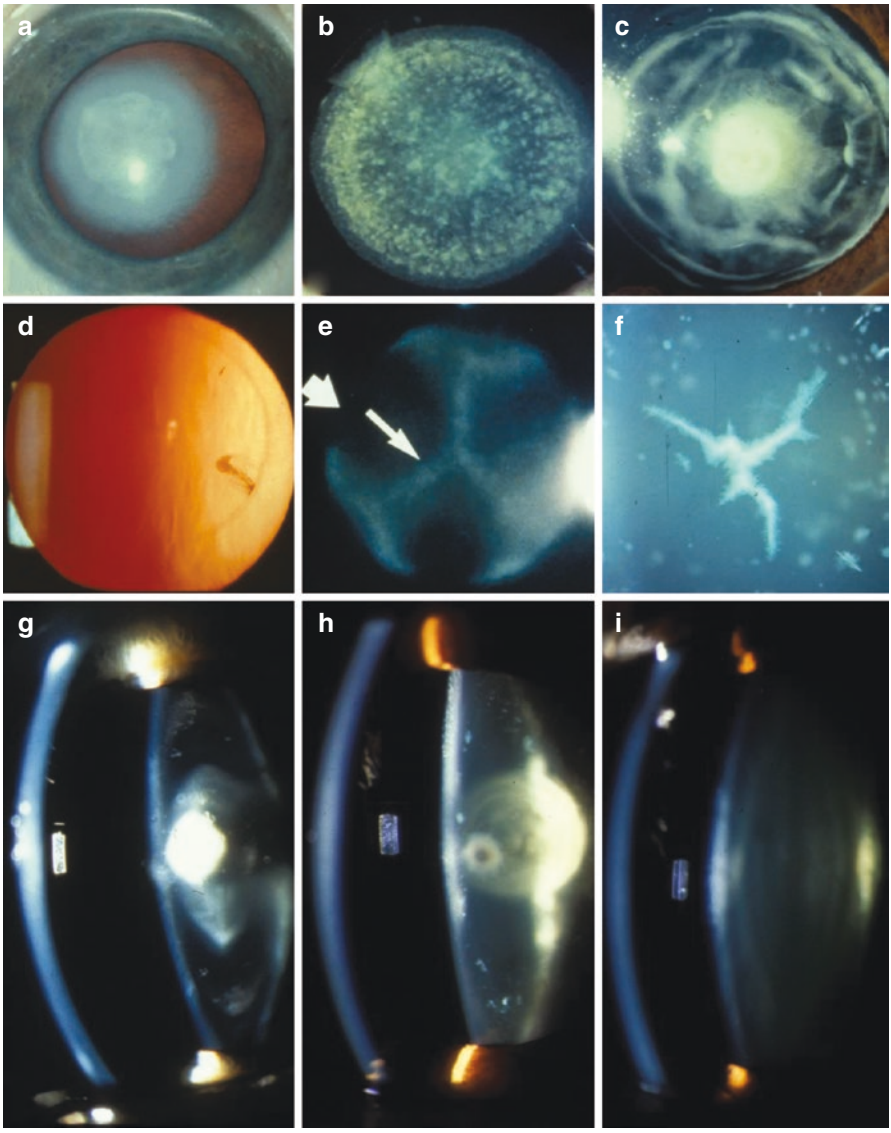

Fig. 1.1 Examples of cataract morphologies. (**a**) Dense nuclear cataract. The macula and optic nerve are obscured by this cataract. (**b**) Punctate nuclear cataract. (**c**) Multi-lamellar cataract with an anterior polar component. (**d**) Very fine nuclear lamellar pulverulent cataract, demonstrated by retroillumination, with a cortical rider at 10 o'clock. (**e**) Sutural cataract with a nuclear lamellar component. (**f**) Sutural cataract with a cortical cerulean or blue dot component. (**g**) Dense anterior polar cataract visible on slit-lamp examination. Some opacification of the lens nucleus is also visible. (**h**) Dense posterior polar cataract visible on slit-lamp examination. A smaller anterior polar cataract is also visible so that this would be termed a bipolar cataract. (**i**) Posterior subcapsular cataract

Table 1.2 Fractions of cataract types caused by specific genes

	Nuclear	Lamellar / Zonular	Sutural	Cortical	Post. Polar	Ant. Polar	Corraliform	Cerulean	PSC
GJA8	0.09	0.12	0.05	0.00	0.06	0.00	0.00	0.00	0.29
GJA3	0.10	0.16	0.05	0.02	0.06	0.00	0.16	0.00	0.00
CRYAA	0.12	0.12	0.03	0.02	0.06	0.40	0.00	0.00	0.00
CRYAB	0.02	0.03	0.00	0.00	0.13	0.00	0.00	0.00	0.00
CRYBB1	0.06	0.00	0.03	0.05	0.03	0.00	0.00	0.00	0.00
CRYBB2	0.04	0.05	0.03	0.07	0.00	0.00	0.05	0.43	0.14
CRYBB3	0.01	0.00	0.00	0.05	0.00	0.00	0.00	0.00	0.00
CRYBA3	0.08	0.10	0.19	0.07	0.03	0.00	0.00	0.00	0.00
CRYBA4	0.01	0.03	0.00	0.00	0.00	0.10	0.00	0.00	0.00
CRYGC	0.08	0.06	0.00	0.00	0.00	0.00	0.00	0.00	0.00
CRYGD	0.09	0.04	0.03	0.02	0.06	0.10	0.74	0.21	0.00
CRYGS	0.00	0.04	0.05	0.07	0.00	0.00	0.00	0.00	0.00
NHS	0.05	0.00	0.30	0.12	0.00	0.00	0.05	0.00	0.00
HSF4	0.02	0.12	0.03	0.10	0.00	0.10	0.00	0.00	0.00
EPHA2	0.05	0.01	0.00	0.14	0.06	0.10	0.00	0.00	0.14
FOXE3	0.01	0.00	0.00	0.07	0.00	0.00	0.00	0.21	0.00
MAF	0.02	0.04	0.00	0.00	0.09	0.10	0.00	0.07	0.00
PITX3	0.00	0.00	0.00	0.02	0.38	0.00	0.00	0.00	0.43
EYA1	0.01	0.01	0.00	0.02	0.00	0.00	0.00	0.00	0.00
BFSP2	0.01	0.03	0.14	0.07	0.00	0.00	0.00	0.00	0.00
AQP0	0.04	0.04	0.08	0.07	0.03	0.00	0.00	0.07	0.00
CHMP4B	0.01	0.01	0.00	0.00	0.00	0.10	0.00	0.00	0.00
FYCO1	0.05	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00

Frequencies are calculated from CAT-MAP

Sutural or stellate cataracts (Fig. 1.1e, f) affect the region of convergence of lens fibers in the fetal nucleus (Y sutures). The sutures are visible even in normal lenses by slit-lamp biomicroscopy as an upright Y anteriorly and an inverted Y posteriorly. About 30% of sutural cataracts result from mutations in *NHS*, 19% in CRYBA3, and 14% in *BFSP2*, and the remainder are caused by multiple additional genes (Table 1.2).

Cerulean or *blue dot* cataracts are characterized by numerous small bluish opacities in the cortical and nuclear areas of the lens (Fig. 1.1f). About 43% of cerulean cataracts are caused by *CRYBB2* mutations, while *CRYGD* and *FOXE3* mutations account for 21% each.

Coralliform cataracts can be described as dispersed popcorn or coral-like opacities, primarily in the nuclear area (Fig. 1.1g), with 74% caused by *CRYGD* mutations and 16% by mutations in GJA3 (Table 1.2).

Polar opacities may involve the anterior (Fig. 1.1h), posterior (Fig. 1.1i), or both poles of the lens (bipolar). Anterior polar cataracts are often bilateral and minor in size and visual impact and tend not to progress. They can be associated with microphthalmos, persistent pupillary membrane, or anterior lenticonus. *CRYAA* mutations account for 40% of isolated anterior polar cataracts. Posterior polar opacities generally imply a significant visual threat, regardless of size. They can be isolated or appear in association with other abnormalities such as lentiglobus, lenticonus, or remnants of

the tunica vasculosa lentis. Involvement of the posterior capsule may include capsular fragility, which complicates surgical interventions. Although usually stable over time, some cases may progress. Thirty percent of isolated inherited cases are caused by mutations in *PITX3*. Involvement of the posterior subcapsular lens cortex and capsule, although frequently acquired and associated with exogenous insults such as steroids or radiation, occasionally accompanies posterior polar opacities.

As lens fiber cells continue to be laid down throughout life, cataracts developing postnatally tend to present as cortical or, occasionally, posterior subcapsular opacification. *Posterior subcapsular* cataracts (PSCs) have been classically associated with proliferation of Wedl cells (dysplastic bladderlike fiber cells); however, they can also be secondary to abnormalities of the posterior fiber ends. Forty-three percent of genetic PSCs are caused by *PITX3* mutations and 29% by mutations in *GJA8* (Table 1.2).

Other varieties of cataract can usually be described through a combination of the above terms, although some cases have unique appearance, such as ant's egg cataract, in which a mutation in connexin 46 (*GJA3)* causes formation of beaded structures resembling ant's eggs [14, 15].

Membranous cataracts result from resorption of lens proteins, often from a traumatized lens, with resulting fusion of the anterior and posterior lens capsules to form a dense white plaque. They usually cause severe loss of vision.

Mature or total or complete cataracts may represent a late stage of any of the above types of cataract, in which the entire lens is opacified. Visualization of the posterior lens capsule is not possible, vision is obscured, and deep amblyopia can be expected in early, asymmetric unilateral cases. Mature cataracts present a special challenge in surgical removal due to associated liquefied contents, a weak friable capsule and zonules, and high risk of vitreous loss and warrant staining of the anterior capsule and usage of specialized surgical techniques.

Etiology

Inheritance and Genetic Architecture

In contrast to age-related cataracts, which have a strong environmental component, hereditary congenital cataracts are almost completely determined by germline mutations, which may present as autosomal dominant (most frequent), autosomal recessive, or X-linked traits. Although involvement of specific genes can be implied by the location and appearance of the opacity, clinically identical cataracts can result from different mutations and even separate genes and be inherited in different patterns. Conversely, morphologically distinct cataracts can result from a single mutant gene in a single large family [16]. The number of known cataract loci has increased dramatically in the last few years to well over 60 loci at which mutations in over 40 genes have been demonstrated to cause inherited human cataracts, with the best indications being that approximately 40% of cataract loci have been identified. Obviously, much remains to be learned about the genetic contributions to inherited congenital cataracts.

The genetic architecture of Mendelian cataracts largely comprises a limited number of functional groups making up biological pathways or processes critical for lens development, homeostasis, and transparency (Table 1.1). About a third of cataracts result from mutations in lens crystallins, about a quarter result from mutations in transcription or growth factors, slightly less than one-seventh result from mutations in connexins, about one-tenth result from mutations in membrane proteins or components, somewhat less than 5% show mutations in chaperone or protein degradation components each, and about 2% result from mutations in a mixed group of other genes, while the genes of about 3% of cataract loci have not been identified yet (Fig. 1.2). A more complete list with detailed descriptions and references can be found in Cat-Map [17].

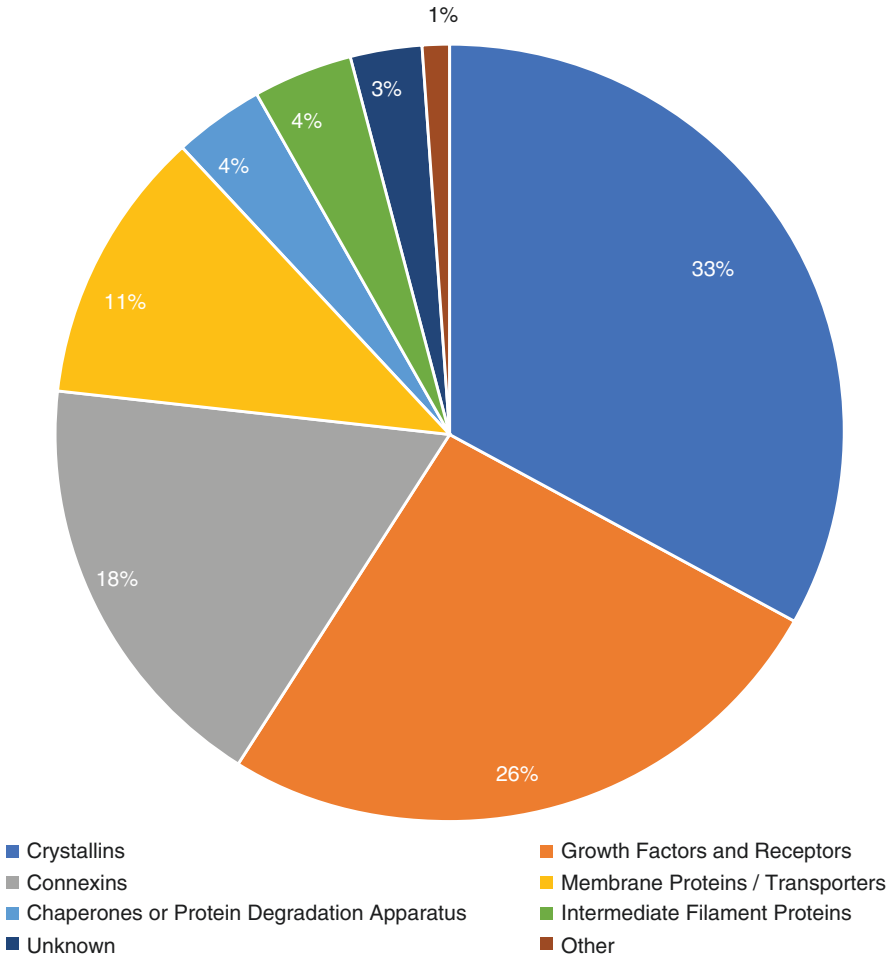

Fig. 1.2 Fraction of cataract families with mutations in genes belonging to specific pathways, processes, or protein families. Crystallins are the most commonly mutated genes in congenital cataract, followed closely by growth factors, connexins, and then membrane proteins. The remainder are caused by additional groups of genes important in a variety of metabolic and functional processes in the lens

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The lens has a single layer of anterior epithelial cells present under the anterior lens capsule, overlaying the fiber cells wrapped onion-like around the lens nucleus [18]. Cell division occurs mainly in the germinative zone just anterior to the equator (bow region of the lens). The cells then move laterally toward the equator, where the anterior epithelial cells undergo mitosis and then differentiate, migrating inward toward the lens nucleus and elongating to form the secondary lens fibers [19].

The organelle-rich anterior epithelial cells beneath the lens capsule control movements of substances into and out of the lens and are connected by gap junctions [20], which facilitate exchange of ions and other low molecular weight metabolites, but tend to lack tight junctions, which would seal the extracellular spaces to these molecules [21]. Differentiating lens fiber cells move toward the nucleus and lose their organelles, including the mitochondria, Golgi bodies, and both rough and smooth endoplasmic reticulum (ER). Fiber cells, located in the cortical area of the lens, have many interdigitations with minimal extracellular space [22] and are joined by frequent junctional complexes, allowing for intercellular transfer of metabolites [23]. Both the anterior epithelial cells and especially the fiber cells contain large amounts of crystallins, as well as cytoskeletal proteins. The complex process of lens differentiation with its changing protein components is largely under transcriptional factor control.

Transcription and Developmental Factors

Although the process and mechanisms of lens development are still being elucidated, a number of transcription and developmental factors including *PAX6*, *RAX*, *VSX2*, *MAF*, *FOXE3*, *EYA1*, and *PITX3* are critical for lens development [24–29]. Mutations in *PAX6*, which is expressed in the entire developing eye, are associated with aniridia, which is often accompanied by cataracts [30]. Mutations in *PITX3* often cause posterior polar cataracts (70%) and can be associated with anterior segment mesenchymal dysgenesis (ASMD or ASD). Mutations in *NHS* are associated with the Nance-Horan syndrome (NHS), which includes cataracts, facial dysmorphism, dental abnormalities, and developmental delay [31]. The cataract in NHS is typically nuclear (39%) or sutural (39%). In contrast, although it is expressed across most ocular tissues, mutations in *HSF4* (heat shock factor 4) tend to cause isolated nuclear or lamellar cataracts [32], as do mutations in *SIPAIL3*, which functions in epithelial cell morphogenesis and polarity [33].

Overall, most mutations in transcription and developmental factors tend to result in autosomal dominant cataracts with a ratio of about 2.5:1. Mutations in *TDRD7*, a widely expressed Tudor domain RNA-binding protein of RNA granules that interact with STAU-1 ribonucleoproteins, also cause cataract, probably related to the high levels of mRNA synthesis required during lens differentiation [34]. Similarly

included in this group is the ephrin receptor EPHA2, which, while not actually a transcription factor, plays a major role in developmental processes in the eye and nervous system. Mutations in *EPHA2* can cause both dominant and recessive congenital cataracts, as well as contributing to age-related cataract [35–40].

Lens Crystallins

Crystallins are the most highly expressed proteins in the lens, comprising about 90% of the soluble protein. Their physical properties, specifically close packing and stability, are critical for lens transparency. Both characteristics are probably responsible for the crystallins being the most commonly mutated genes implicated in human congenital cataracts.

The three classes of crystallins in humans are encoded by multiple genes. α-Crystallins are large proteins with chaperone-like activity, able to bind partially denatured proteins and prevent aggregation (especially relevant to age-related cataracts). The β- and γ-crystallins, comprising most of the water-soluble mass of the lens, are part of a large gene superfamily and are present in extraocular tissues. As damaged or mutant β- and γ-crystallins start to form irreversible aggregates that eventually precipitate out of solution, they are bound by α-crystallins and held in soluble aggregates. However, if the mutation is severe enough to result in rapid denaturation without an intermediate molten globule state, they can escape binding by α-crystallins and other chaperones in the lens, causing direct damage to lens cells or initiating cellular processes such as the unfolded protein response (UPR) and apoptosis [41]. Similarly, although most pertinent to age-related cataract, denaturation and binding of large amounts of crystallins can lead to high molecular weight aggregates large enough to scatter light themselves and eventually overwhelm the α-crystallin chaperone system causing cataract [42]. Thus, denatured crystallins can lead to cataract directly by scattering light or more catastrophically by toxic effects on the lens cells and microarchitecture perhaps inducing the UPR and/or apoptosis [43].

Most cataracts resulting from mutations in crystallins are autosomal dominant, with a ratio of about 12:1 dominant to recessive. This finding is consistent with a deleterious gain of function manifested by denaturation and precipitation of protein aggregates, with toxic effects on lens cells and induction of the UPR. Crystallin-related cataracts are heavily biased toward nuclear or lamellar cataracts, although 40% of *CRYAB* cataracts are posterior polar and 50% of *CRYBB3* cataracts are cortical (Table 1.3a).

Some crystallin mutations cause autosomal recessive cataracts. These include *CRYAA* (3 of 41), *CRYAB* (5 of 16), *CRYBB1* (6 of 19), and *CRYBA4* (1 of 5), suggesting that these crystallins might have additional functions in the lens other than solely structural roles. The α-crystallins are well known to function as molecular chaperones, but additional functions for the β-crystallins remain to be identified, and no recessive mutations have been identified for any γ-crystallin. Alternatively, mere haplo-insufficiency in crystallin genes causing autosomal recessive cataracts might be sufficient to impair lens transparency and function.

Table 1.3 Clinical characteristics of cataracts by their genetic cause

	GJA8	GJA3	CRYAA	CRYAB	CRYBB1	CRYBB2	CRYBB3	CRYBA3	CRYBA4	CRYGC	CRYGD	CRYGS
Nuclear	0.56	0.51	0.59	0.40	0.75	0.33	0.50	0.47	0.50	0.76	0.43	0.11
Lamellar	0.26	0.29	0.22	0.20	0.00	0.17	0.00	0.22	0.33	0.24	0.07	0.33
Sutural	0.06	0.05	0.02	0.00	0.06	0.04	0.00	0.19	0.00	0.00	0.02	0.22
Cortical	0.00	0.02	0.02	0.00	0.13	0.13	0.50	0.08	0.00	0.00	0.02	0.33
Post. Polar	0.06	0.05	0.05	0.40	0.06	0.00	0.00	0.03	0.00	0.00	0.05	0.00
Ant. Polar	0.00	0.00	0.10	0.00	0.00	0.00	0.00	0.00	0.17	0.00	0.02	0.00
Corralliform	0.00	0.07	0.00	0.00	0.00	0.04	0.00	0.00	0.00	0.00	0.32	0.00
Cerulean	0.00	0.00	0.00	0.00	0.00	0.25	0.00	0.00	0.00	0.00	0.07	0.00
PSC	0.06	0.00	0.00	0.00	0.00	0.04	0.00	0.00	0.00	0.00	0.00	0.00
% defined	0.58	0.77	0.89	0.56	0.55	0.65	0.50	0.91	0.75	0.62	0.79	1.13
% other	0.42	0.23	0.11	0.44	0.45	0.35	0.50	0.00	0.25	0.38	0.21	0.00

	NHS	HSF4	EPHA2	FOXE3	MAF	PITX3	BFSP1	BFSP2	AQP0	GCNT2	FYCO1
Nuclear	0.39	0.25	0.50	0.25	0.33	0.06	0.50	0.17	0.45	0.50	1.00
Lamellar	0.00	0.45	0.05	0.00	0.25	0.00	0.25	0.17	0.15	0.25	0.00
Sutural	0.39	0.05	0.00	0.00	0.00	0.00	0.00	0.42	0.15	0.00	0.00
Cortical	0.18	0.20	0.27	0.38	0.00	0.06	0.25	0.25	0.15	0.00	0.00
Post. Polar	0.00	0.00	0.09	0.00	0.25	0.71	0.00	0.00	0.05	0.00	0.00
Ant. Polar	0.00	0.05	0.05	0.00	0.08	0.00	0.00	0.00	0.00	0.25	0.00
Corralliform	0.04	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00	0.00
Cerulean	0.00	0.00	0.00	0.38	0.08	0.00	0.00	0.00	0.05	0.00	0.00
PSC	0.00	0.00	0.05	0.00	0.00	0.18	0.00	0.00	0.00	0.00	0.00
% defined	0.50	0.71	0.96	0.42	0.63	0.61	0.57	1.00	0.61	0.27	0.67
% other	0.50	0.29	0.04	0.58	0.37	0.39	0.43	0.00	0.39	0.73	0.33

	GJA8	GJA3	CRYAA	CRYAB	CRYBB1	CRYBB2	CRYBB3	CRYBA3	CRYBA4	CRYGC	CRYGD	CRYGS
AD	49	44	38	11	13	28	3	26	4	30	51	8
AR	4	1	3	5	6	0	2	0	1	0	0	0
AD/AR	12.25	44.0	12.67	2.20	2.17	N/A	1.50	N/A	4.00	N/A	N/A	N/A
Group ratio	18.6		12.47									
% AD	0.92	0.98	0.93	0.69	0.68	1.00	0.60	1.00	0.80	1.00	1.00	1.00

	NHS	HSF4	EPHA2	FOXE3	MAF	PITX3	BFSP1	BFSP2	AQP0	GCNT2	FYCO1
AD	0	16	18	6	10	27	2	8	29	0	0
AR	0	7	5	11	0	1	2	2	1	12	14
AD/AR	N/A	2.29	3.60	0.55	N/A	27.00	1.00	4.00	29.00	0.00	0.00
Group ratio	2.54						2.50		Varied		
% AD	na	0.70	0.78	0.35	1.00	0.96	0.50	0.80	0.97	0.00	0.00

	Total
AD	421
AR	77
AD/AR	5.47
Group ratio	
% AD	0.85

Gap Junction Proteins (Connexins)

Lacking blood vessels, the lens is dependent on gap junctions and intercellular channels composed of hexameric hemichannels from two adjacent cells joined to allow communication and transfer of nutrients, especially between fiber cells. Lens junctions contain *GJA3* (encoding connexin 46) and *GJA8* (encoding connexin 50) [44, 45]. Ninety-two percent of mutations in *GJA3* and 98% in *GJA8* have been implicated in autosomal dominant human cataract with a few autosomal recessive families reported for each. They also usually cause nuclear or lamellar cataracts (Table 1.3a). Because of their multimeric nature, some missense mutations in connexins can have a dominant negative effect on gap junction function as exemplified by the p.P88S change in *GJA8* [46]. The mutant protein is incorporated into the gap junction structure and inactivates the entire junction [47]. Similarly, autosomal dominant p.E134G and p.T138R mutations inhibit normal trafficking of aquaporin 0 (AQP0) to the plasma membrane [48] and also interfere with water channel activity by normal AQP0, consistent with a dominant negative mechanism. Thus, when the mutant AQP0 is inserted into the channel, it adversely affects channel function, even in the presence of wild-type molecules in the same channel.

Some gap junction mutations causing retention in the endoplasmic reticulum can induce the UPR [49], and conversely, mutations causing enhanced hemichannel function also can lead to cell death and cataract [50]. *GJA8* mutant cataracts have also been associated with microcornea with or without myopia and occasionally with microphthalmia, while *GJA3* mutations are usually isolated.

Membranes and Their Proteins

In addition to the gap junction proteins, lens epithelia require large amounts of membranes when they elongate to form fiber cells and must synthesize the lipids making up their membranes. They are also required as the protein components for circulation of water and small molecules critical for lens fiber cell homeostasis and function. Mutations in *SLC16A12*, a transmembrane protein functioning in creatine transport, can cause dominant cataracts, sometimes accompanied by microcornea or renal glycosuria.

Aquaporins are integral membrane proteins that generally act as water channels. Mutations in *AQP0*, also known as major intrinsic protein, *MIP*, are also a major contributor to inherited nuclear congenital cataracts, although some lamellar, sutural, or cortical cataracts may also form (Table 1.3a). Similar to some gap junction mutations, autosomal dominant p.E134G and p.T138R mutations inhibit normal trafficking of AQP0 to the plasma membrane [51] and also interfere with water channel activity by normal AQP0, consistent with a dominant negative mechanism. *LIM2* is also required for cell junctions in lens fiber cells, and autosomal recessive cataracts have been associated with its mutated form [52–54].

TMEM114, a transmembrane glycoprotein member of a group of calcium channel gamma subunits, can also cause cataracts when mutated. While mutations in the wolframin ER transmembrane glycoprotein (*WFS1*) can cause Wolfram syndrome, they have also been described in a family with isolated cataracts [55, 56]. Mutations in *LEMD2*, an important signaling and organization protein in the nuclear membrane, have been associated with autosomal recessive cataracts [57], as well as mutations in acylglycerol kinase (*AGK*), a mitochondrial membrane lipid kinase required for synthesis of phosphatidic and lysophosphatidic acids [58], and mutations in lanosterol synthase (*LSS*), which is required for synthesis of cholesterol. These are possibly related to the large amounts of membrane components required to be synthesized during fiber cell differentiation, although lanosterol has been also shown to act as a chaperone for denatured crystallins [59].

Beaded Filament and Other Intermediate Filament Proteins

Intermediate filaments are cytoskeletal proteins with an average diameter of around 10 nm. In the lens, these include vimentin filaments, which are present in the anterior epithelial cells but are replaced by lens-specific beaded filaments as the cells differentiate into fiber cells.

Beaded filaments are composed of BFSP1 (CP115, filensin) and BFSP2 (CP49, phakinin), both highly divergent members of the intermediate filament protein family. About 50% of mutations in *BFSP1* cause nuclear cataracts [60], while about 42% of mutations in BFSP2 cause sutural cataracts [61] (Table 1.3a). BFSP mutations can be either dominant or recessive, with missense mutations tending to cause dominant cataracts while nonsense and frameshift mutations causing deletions leading to recessive cataracts.

Mutations in vimentin can cause autosomal dominant cataracts. Mutations in *COL4A1* can cause dominant cataracts [62], and mutations in prolyl 3-hydroxylase 2 (*P3H2*, also known as *LEPREL1*) which is active in collagen chain cross-linking, can cause cataracts, sometimes accompanied by ectopia lentis and high myopia.

Chaperones and Protein Degradation

Lens fiber cells lack nuclei, and therefore, the stability and longevity of their proteins must suffice for the lifetime of an individual. To facilitate this, the lens contains high levels of chaperones such as the α-crystallins. In this light, a mutation in *UNC45B*, a co-chaperone for HSP90, has been implicated in juvenile cataract [63].

Conversely, lens fiber cell differentiation also requires elimination of all organelles and their associated proteins, requiring highly active protein degradation systems. Mutations in *CHMP4B*, part of the endosomal-sorting complex required for transport and autophagy, have been shown to cause autosomal dominant posterior

polar or subcapsular cataract [64]. Mutations in Ras-related GTP-binding protein A (*RRAGA*), a component of the mTORC signaling cascade controlling protein synthesis, have been implicated in autosomal dominant cataracts [65]. Mutations in the mitochondrial chaperone and protein degradation in protease lon peptidase 1 (*LONP1*) can also cause recessive cataracts, emphasizing the importance of mitochondrial function in the lens epithelia for lens transparency. *FYCO1* is a scaffolding protein active in microtubule transport of lysosomes including autophagic vesicles [66]. Mutations in *FYCO1* can cause autosomal recessive cataracts [67], consistent with an important role for autophagic vesicles in organelle degradation as equatorial epithelia differentiate into lens fiber cells. Interestingly, all cataracts resulting from *FYCO1* are nuclear. Finally, mutations in *EPG5*, a key regulator of autophagy that is active in autolysosome formation, while not shown to cause isolated cataracts, do cause Vici syndrome, which includes cataracts [68].

Other Genes and Pathways

GCNT2 encodes the I-branching enzyme for poly-N-acetyllactosaminoglycans. In addition to determining the i (predominantly fetal and neonatal) and I (predominantly adult) antigens of the I blood groups, it influences the epithelial to mesenchymal transition and cell migration and can cause autosomal recessive cataracts when mutated [69]. About 50% of these cataracts are nuclear, 25% are lamellar, and another 25% are anterior polar.

Mutations in *TAPT1*, which can disrupt Golgi structure and trafficking, can cause autosomal recessive cataracts, as can mutations in aldo-keto reductase family 1 member E2 (*AKR1E2*) and renalase (*RNLS*, FAD-dependent amine oxidase).

Interestingly, mutations in the iron-responsive element of ferritin L (light chain, *FTL*) cause the hyperferritinemia-cataract syndrome in which loss of translational control results in massive overexpression of FTL that crystallizes in the lens and gives granular opacities in the nucleus and cortex [70, 71]. This example of an extraneous protein expressed at high levels in the lens emphasizes the requirement that crystallins or other proteins must be exceptionally soluble and stable to be expressed at crystallin-like levels without causing dysfunction.

Finally, *TDRD7*, a widely expressed Tudor domain RNA-binding and RNA-processing protein of RNA granules, also causes cataract when mutated, presumably secondary to high levels of unbound mRNA during lens differentiation [34, 72, 73].

Pathology

The many etiologies described above are consistent with diverse pathological findings. Basically, the pathological characteristics of cataracts can be grouped into two broad categories, based on the condition of the lens microarchitecture*:* those

causing rapid gross structural changes and those preserving microarchitecture initially, slowly inducing change over time.

Some congenital cataracts result from mutations with catastrophic effects on the protein, causing gross structural changes and precipitation of similar impact in other lens components. The denatured proteins either escape or overwhelm binding by α-crystallin or other lens chaperones and are toxic to lens cells, interfering with their proper differentiation. This leads to death and degeneration, often through UPR and apoptosis. These mutations are often associated with breakdown of lens microarchitecture, including degeneration (and possible calcification) of lens fiber cells, eventually forming large lacunae filled with proteinaceous debris, rupturing the lens capsule in the most severe cases. The resulting large fluctuations in optical density cause light scattering and are the best studied animal models of inherited congenital cataracts. One example is a c.215 + 1G > A splice mutation in *CRYBA1*, causing a p.Ile33_Ala119del mutant βA3/A1-crystallin protein [74], and many other well-studied changes [75–78].

The mechanisms described above are not the exclusive cause of congenital lens opacities, as potentially toxic high molecular weight protein aggregates can form when the lens cell α-crystallin becomes saturated with denatured crystallins, resulting in damage to lens cells.

Genetic Aspects/Inheritance Patterns of Congenital Cataracts

About 85% of inherited congenital cataracts show an autosomal dominant inheritance pattern, although this varies significantly depending on the population and study (Table 1.3b). In addition, there is significant variation in inheritance pattern among the various genes. All cataracts caused by *CRYBB2*, *CRYBA3*, *CRYGC*, *CRYGD*, *CRYGS*, and *MAF* are dominant, which suggests that there might be redundant biological systems for these proteins in the lens so that their absence by itself would not disrupt lens biology and transparency.

In contrast, the presence of autosomal recessive inheritance patterns of cataracts caused by *CRYBB3* and *CRYBA4* suggests that they might have an irreplaceable role in lens biology in addition to that of structural lens crystallins. The absence of autosomal dominantly inherited cataracts resulting from *GCNT2* and *FYCO1* suggests that these cataracts all result from the absence of the functional protein, implying a unique and necessary role for these genes in the lens.

Inherited congenital cataracts affect all populations throughout the world and without early diagnosis and prompt treatment are a significant cause of blindness in infants. While clinically identical cataracts can be caused by mutations in different genes and identical mutations in the same gene can cause clinically different cataracts, it is possible to identify general correlations between some of the causative genes and specific cataract morphologies, which might be useful in guiding genetic diagnosis. Genes associated with congenital cataracts tend to belong to molecular or biochemical pathways important for lens development and homeostasis. While

many genes have been identified, there remains much work to be done both in identifying the remaining causative genes and understanding the molecular pathologies that lead to the common endpoint of lens opacity or cataract.

Case A 2-month-old baby boy presented with concern for dull red reflex in both eyes. He had an otherwise unremarkable birth history. He was diagnosed with congenital cataracts, the right greater than the left. He underwent prompt surgical removal of these opacities, with lensectomy, anterior vitrectomy, and intentional aphakia. The right denser lens was removed first, and the left lens was removed 1 week later. Following surgery, he recovered well, using aphakic glasses consistently. He had further evaluation with genetics to guide systemic workup.

A complete multigenerational family history was obtained from the parents. No family history of early onset cataracts, glaucoma, or other vision-threatening condition was found on either maternal or paternal side. Insurance pre-authorization allowed for skeletal survey, urine glycosaminoglycans, and oligosaccharides screening, as well as a whole blood sample to be transmitted to Invitae© Genetics Lab (Invitae Corporation, USA) for a 37 gene cataracts panel. Targeted sequencing for genes related to congenital cataract was performed, including sequence analysis and deletion/duplication analysis.

Testing revealed the child had two gene variants identified on the gene panel for cataracts: the c.390 + 1G > A variant in the AGK gene and the c.551 T > G variant in the *CRYBB2* gene.

The c.390 + 1G > A variant in the *AGK* gene has been seen previously in other patients, and this variant is known to affect gene function; however, all previously reported patients who have cataracts due to AGK have two gene variants present, as this is a recessive condition, and also have other medical problems such as cardiomyopathy, low muscle tone, and developmental delay.

To help clarify if the *CRYBB2* gene variant was a spontaneous DNA change in this child or if it was a benign DNA change inherited from a parent without cataracts, parental testing for this variant was conducted. Parents completed this testing and neither had this variant. Therefore, since this was a de novo DNA variant, it was felt to be the likely the cause of his cataracts.

Comment The presenting child was otherwise healthy and had an unremarkable birth history and no family history of inherited lens opacities. A cataract panel was sent revealing two gene variants, the c.390 + 1G > A variant in the *AGK* gene and the c.551 T > G variant in the *CRYBB2* gene. Since the patient was healthy aside from his cataracts and because he had only one *AGK* gene variant present, this was not felt to be the underlying cause for his cataracts. To confirm lack of significant cardiomyopathy, which is associated with *AGK* mutations, an echocardiogram was recommended. The c.551 T > G variant in the *CRYBB2* gene is considered a "variant of uncertain significance" because it has not been published in the literature and the effects of this variant on the *CRYBB2* gene function are currently unclear. It is thought that patients who have congenital cataracts due to a known variant in the *CRYBB2* gene have a dominant cause

to their cataracts, and there are no additional associated medical problems caused by the *CRYBB2* gene. This was felt to be most consistent with the child's presentation.

Another important consideration was to ensure that the patient did not have a storage disorder such as a mucopolysaccharidosis or oligosaccharidosis. These conditions are not characteristically known to present with cataracts at birth, but are important to rule out because effective treatment must be initiated before 2 years of age. Reassuring aspects against these diagnoses include the fact that his physical exam was normal; specifically, he had no organomegaly or spinal deformity. Laboratory testing and skeletal survey confirmed no concern for storage disorder.

To date, numerous genes have been identified to cause congenital cataracts, and genetic testing for several of these genes is available commercially. Sometimes, testing for mutations in all of these known genes is entirely normal, indicating that some cases of congenital cataracts are attributed to other, yet undiscovered, genes.

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Chapter 2
Nonsurgical Management of Infantile and Juvenile Cataract

Michael X. Repka

Visually significant or total cataracts in infants and children, both unilateral and bilateral, typically involve straight-forward decision-making in terms of moving expeditiously to clearing the visual axis soon after discovery. However, partial opacities present more difficult management decisions, especially in young children, at the time of presentation. In addition, that decision-making process continues well into the future based on patient factors, cataract stability/progression, and visual needs. The decision to treat or monitor will also be influenced by laterality, type of lens opacity, age of patient, developmental outlook, visual prognosis, and family history. Bilateral partial cataracts allow more time to monitor the child's course than unilateral partial cataracts. Early treatment may prevent amblyopia but raises the risk of glaucoma [1, 2], retinal detachment, the need for additional surgery, and other less common complications. The outcomes for monitoring (and not operating) are not reported and likely quite heterogenous. The clinical tools to assist with informing those decisions are limited by age, measurement variability, and cooperation.

Crucial to decision-making are the ability of the clinician to examine the child, to be able to see the retina in spite of the partial cataract, measure the refractive error, and vision when it can be measured. The surgeon is trying to balance amblyopia development and visual impairment level with the issues associated with surgery, including the need for one or more future surgeries, as well as other complications.

Tools to Assess

The ophthalmologist needs access to a retinoscope, direct ophthalmoscope, indirect ophthalmoscope, and handheld slit lamp. All of the instruments are used to

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develop an overall judgment of the impact of the cataract on vision, hopefully as part of in office testing. On occasion, an exam under anesthesia may be needed for some infants and children, but contemporary pediatric care has moved to perform these exams as infrequently as possible. There has been recent concern about the impact of multiple anesthesia events on brain development, so the ophthalmologist needs to balance the needs of the eye exam with developmental concerns and neurological toxicity [3]. This area of concern needs to be monitored by the surgeon and discussed with parents if necessary.

Using each instrument, the surgeon assesses the quality of the red reflex and how much of the pupillary aperture is occluded. The most useful for monitoring the visual impact of the cataract is probably the retinoscope. Although a 3 mm size is often stated as a maximum guideline, it remains a consensus recommendation and the decision customized to the patient. The assessment of size is best done with a direct ophthalmoscope and distortion of the retinal reflex with the retinoscope. This size threshold for significance also assumes there is no disruption of retinoscopy beyond the opacity. If the retinoscopy uncovers significant distortions in the red reflex beyond the central opacity, the size including the area of distortion needs to be considered as the true size of the cataract. Try to be certain that the retinoscopic reflex distortion is from the lens and not the cornea.

The lens opacity for some children may be eccentric, often in the area of prior attachment of the hyaloid vessels (slightly nasal and inferior to the visual axis), and thus less damaging to the retinoscopy. This may require review at a subsequent visit to confirm if the central axis is indeed clear of the lens abnormality or not. The timing of that visit will depend on the overall level of concern. The use of a handlight to examine the eye can be misleading in some cases. Because the pupil will be constricted from the bright light, it might seem that the child cannot see around the opacity. For this evaluation, often the retinoscope on dim can be very useful.

The slit lamp is used primarily to determine the depth of the cataract. It too will constrict the undilated pupil. Posterior and posterior subcapsular lesions are generally more of a problem for vision development than anterior capsular and anterior polar lesions. In many cases, that is because they do not distort the retinoscopy and thus the image viewed as much as posterior lesions.

Visual acuity should be measured with optotypes whenever possible to guide therapy. Lea symbol charts and HOTV single surrounded optotypes can be introduced in the second or third year of life. Preferential looking techniques or fixation preference assessments are done in children unable to perform optotypes. These are sensitive only to large differences in acuity. Whenever possible, a child with a lens opacity should have their visual acuity measured a second time if a treatment decision is anticipated, to be certain they understood the test on the first occasion, performed their best, and it was not simply a poor test. This is usually simple to accomplish for those children with unilateral partial cataract as they will attempt a course of amblyopia therapy (e.g., occlusion) if there is an open optical pathway before moving on to surgery.

Modifying Factors

Bilateral cataracts are much less amblyogenic than unilateral cataract and, generally, the surgeon can wait longer to pursue surgery. Bilateral cataracts are not always symmetric, so the surgeon should carefully consider if both eyes are affected sufficiently and do both require surgery. Guidelines for visual acuity thresholds should be considered consensus-based and subject to individualized decision-making. For unilateral surgery, the outcomes for younger children are much poorer than for older children and poorer for those with bilateral cataracts. Thus, for younger children with measurable accurate visual acuity of better than 20/80 with a unilateral cataract, it is probably best to monitor. For older children, the threshold to perform surgery may be 20/50 or worse or if the opacity is judged to cause at least three lines of vision impairment. For bilateral cataracts, vision of 20/60 or better is typically monitored until the children are older or begin to have functional deficits. As children age, needs will likely become more visually demanding, and surgery may become necessary. This is a very common situation with central and lamellar cataracts.

It is much harder to decide on treatment timing for children unable to perform optotype acuities. For those children, the decision is largely driven by how poor vision appears to be while wearing an occlusive patch over an unaffected eye and clinical judgment about the degree of severity of the opacity.

Anterior polar cataracts are typically much less of a problem than central or posterior polar cataracts. Lamellar cataracts are also usually associated with much better vision and frequently bilateral, allowing surgery to be delayed until significant decline in ability to function. Lamellar cataracts look far worse than their impact on visual acuity.

Family history is increasingly important. In the developed world, cataracts are increasingly familial or inherited (with reduction in the number related to infectious disease). Thus, the family will often have specific concerns about parental and sibling experiences, which will affect the discussion about when to do surgery.

Expected development of the child also plays a role. If there is substantial developmental delay, yet the cataract is significant, then surgery should be done sooner just as in children with no developmental delay. However, if the cataract is mild and the child is able to achieve daily goals, then a delay in treatment would be reasonable.

Management of Cataract

If the patient can be adequately refracted and the decision is to initially manage without surgery, correct the refractive error and have the child get used to wearing glasses full time. This is often the most important part of the treatment. The child should return in a few weeks for reassessment of the vision. If a deficit remains and there is a reasonable visual axis, then an attempt with occlusion therapy for as many hours per day is possible for the parents. Half of waking hours is a useful guideline

to maximize the chance for improvement. In some cases, atropine penalization of the fellow eye is offered with unilateral cataract, but there are no data to affirm its efficacy.

Pupillary dilation may also be used for very central opacities with good peripheral clarity and normal retinoscopy. Topical phenylephrine 2.5% eye drops are often a first choice with partial opacities, but the surgeon needs to verify the adequacy of the dilation to accomplish the goal of providing an optical pathway. In some cases, topical atropine 1.0% to the affected eye can be used for its more potent mydriasis, but the patient will then need to wear appropriate glasses to correct for the cycloplegia. For a child under 30 months, single vision at an intermediate distance works well along with occlusion therapy, whereas for older children bifocals would be needed.

For some cataracts, a delay in surgery will be for a lifetime because of relative advantages and disadvantages of surgery for that child. However, in many instances, surgery is simply deferred until the child is older. The advantages of older age will include a better measured IOL power, easier posterior capsule management, possibly less risk for glaucoma, and perhaps better technology will be available for surgery, the implant, and subsequent care.

Case 1

A 32-month-old boy was found by his pediatrician to have central crystalline lens opacities. His vision was 20/40 in each eye with symbols with +2.50 D refractive error and no heterotropia. No prescription and 6-month follow-up exams planned; this continued for the next 13 years. Over that time, the nuclear cataracts remained unchanged, retinoscopy was crisp, and the hyperopia decreased. At the most recent exam, he noted classroom seating accommodation in the front half of the room worked, although there was some glare when he was outdoors. Visual acuity was 20/40–2 in each eye, normal stereo, and low hyperopic refractive error. There was some parental concern about driving and glare, but the decision to follow was reaffirmed (Fig. 2.1).

Comment In this case, the child's ability to perform a visual acuity at an early age along with the crisp and symmetrical retinoscopy helped drive the decision to delay surgery. In addition, classroom accommodations and his excellent grades in school may have allayed any parental concerns. The issue of driving is important as this issue, especially with nuclear and lamellar cataracts, serves as the impetus for teenagers to undergo surgery.

Case 2

A 33-month-old girl was seen with a history of bilateral asymmetric cataracts. She had a lensectomy performed in the left eye as an infant, which was subsequently treated with an extended wear contact lens. The right eye's mild anterior polar

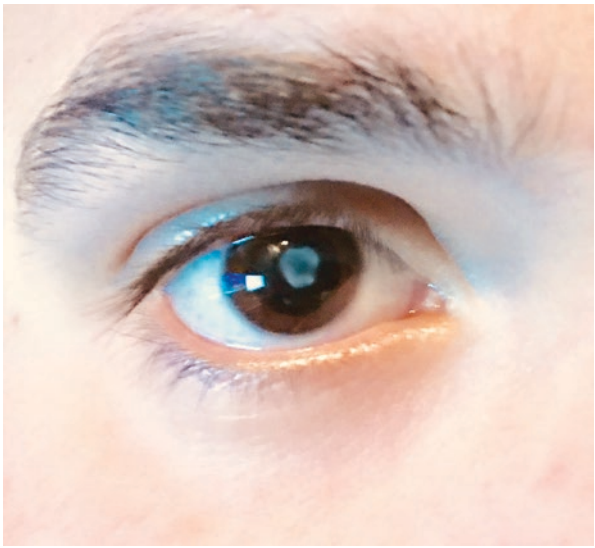

Fig. 2.1 Prominent central nuclear cataract. Central opacity was present and unchanged for many years. The retinoscopy through the lens around the opacity remains normal, and the retinoscopy reflex was crisp

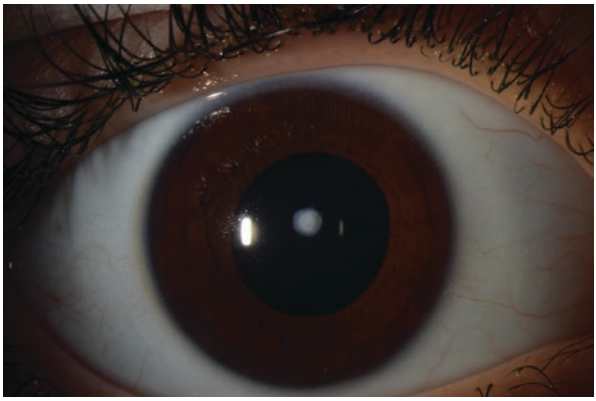

Fig. 2.2 Anterior polar cataract about 1.5 mm in diameter with good vision was not disruptive of the visual axis or the retinoscopy

cataract was monitored (Fig. 2.2). She had a secondary intraocular lens placed at 16 months of age in the left eye with a resultant refractive error of +1.50 D. Amblyopia therapy was performed by underplussing the more hyperopic, phakic right eye (+4.50 D) along with administration of atropine penalization. At 33 months of age, she was 20/25 in the right and 20/20 in the left. The cataract in the right eye was a 2 mm anterior polar opacity with no disruption of the retinoscopy. The optical axis of the left eye was clear. There was no strabismus with positive response to the Fly stereo test. She was to return in 4 months and would have follow-up visits about twice per year.

At 10 years of age, visual acuity was 20/25 OD and 20/20 OS with normal stereo and no strabismus. The refractive error remained anisometropic with +4.50 OD and − 0.50 OS (pseudophakic), but the intraocular pressure was in the mid-20s to upper 20s in the left eye, while the phakic eye was normal.

Comment It is common for children who have frequent eyecare visits to be able to perform optotype visual acuity at a young age easing some decision-making. The localization of the polar cataracts to anterior or posterior lens surface can be difficult with the retinoscope and direct ophthalmoscope in children, but the absence of retinoscopy distortion suggests anterior capsule. This unoperated anterior polar cataract has had a very good outcome, but that can be a tough decision early on in a patient with bilateral cataracts in whom one eye was operated. Early surgery is known to be associated with sustained increased intraocular pressure in the mid-20s and is of increasing concern in this child. Lastly, bilateral cataracts do not always require surgery in both eyes. That decision has to be made considering the nature of the cataract in each eye and the impact of each on vision.

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Chapter 3
Unilateral Congenital Cataracts

Crystal S. Y. Cheung and Bharti R. Nihalani

Unilateral congenital cataracts present unique challenges in diagnosis and management. The age at diagnosis can be delayed due to good vision in the unaffected eye. Most patients with unilateral cataract have deprivation amblyopia at the time of diagnosis owing to perpetual competition from the dominant normal eye. The critical period of visual development is shorter for unilateral compared to bilateral cataracts, making early diagnosis and management necessary. Amblyopia treatment plays a key role in the management of unilateral cataracts. Visual outcomes depend on the age at diagnosis, compliance to optical correction, and amblyopia management.

Epidemiology of Unilateral Congenital Cataracts

The overall prevalence of pediatric cataracts ranges from 0.01% to 0.15% [1], with unilateral cataracts being less common than bilateral cataracts. In a nationwide Danish study of 1027 congenital cataracts, 36% were unilateral cataracts compared to 64% bilateral cataracts [2]. In contrast, a collaborative study by 12 university medical centers in the United States reported that the prevalence of unilateral cataracts was 7.1 per 10,000 cases compared to 6.5 per 10,000 cases for bilateral cataracts [3].

Unilateral congenital cataracts are believed to be the result of localized ocular dysgenesis. Eighty to 90% of unilateral cataracts are idiopathic [2, 4]. The British Congenital Cataract Interest Group (BCCIG) found hereditary disease was associated with only 6% of unilateral cases, compared to 56% of bilateral cases [4].

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Prenatal infections contributed to 2% of unilateral cases compared to 6% of bilateral cases [4]. Microphthalmos and persistent fetal vasculature were the most common associated ocular disorders for unilateral cataracts [4].

Morphology of Unilateral Congenital Cataracts

Nuclear Cataract

Nuclear cataract is the most frequently observed morphology found in pediatric unilateral cataracts. It accounted for 34% of unilateral cataracts across all major ethical groups in the Danish study [2]. This is similar to the findings reported by Infant Aphakia Treatment Study (IATS) where nuclear cataracts accounted for 54% of the cohort [5].

Cortical Cataract

Anterior and posterior cortical cataract, not involving the nucleus, were noted in 25% eyes in IATS [5].

Posterior Capsular Plaque

Posterior capsular plaque was observed in 88% eyes in IATS.; 7.2% had isolated posterior capsule plaque, and it was noted in all nuclear cataracts [5]. Plaques are also common in total cataracts [6] (Fig. 3.1).

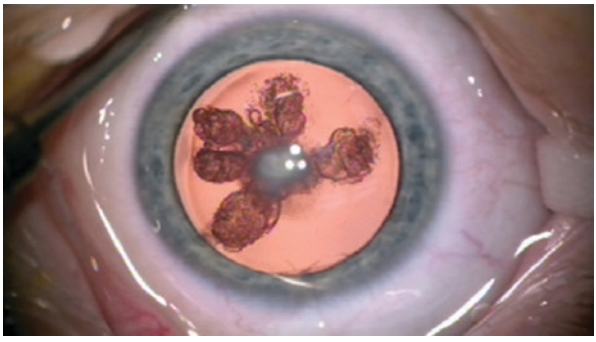

Fig. 3.1 Unilateral cataract with combination of nuclear and cortical cataract with central posterior capsule plaque

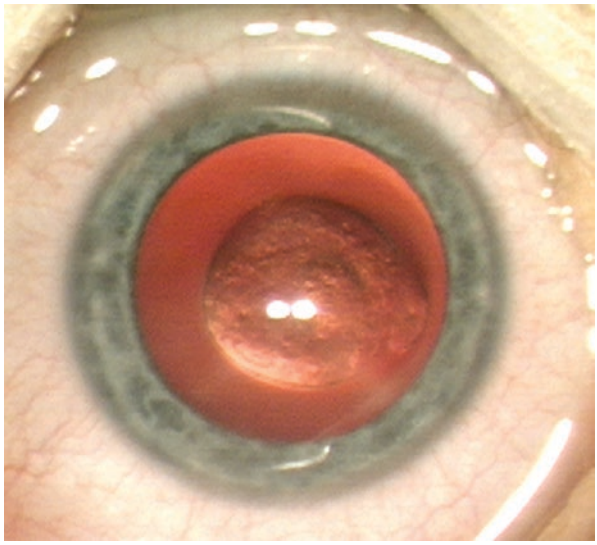

Fig. 3.2 Posterior lenticonus (Image courtesy: Dr. Deborah VanderVeen)

Posterior Lenticonus

Posterior lenticonus was noted in 5–7% of unilateral pediatric cataracts [2, 5]. The progression of posterior lenticonus to cataract involving the cortex and nucleus is variable. During infancy, the cause of vision loss from posterior lenticonus may be due to anisometropia, posterior oil droplet-induced optical distortion [7], or amblyopia [8, 9] (Fig. 3.2).

Preexisting Posterior Capsule Defect (PCD)

PCD is believed to result from progression of posterior lenticonus. The prevalence of PCD varies from 2.2% to 6.75% [6, 10]. The cataract progresses rapidly once the PCD develops. In a classic case, PCD is hidden behind a total white cataract when viewed through an undilated, normal-sized pupil. Preoperative evaluation of such a cataract under maximum dilation is mandatory to unveil the important diagnostic signs such as well-demarcated thick defect margins, white dots on the posterior capsule (Fig. 3.3), and white dots in the anterior vitreous that move with the degenerated vitreous like a fish tail (fish-tail sign) [6].

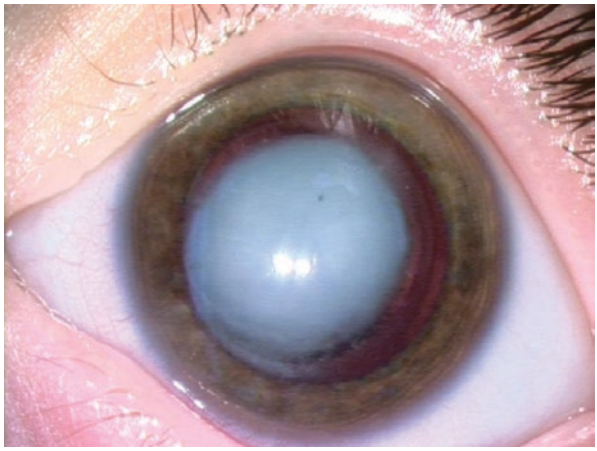

Fig. 3.3 White cataract with preexisting posterior capsule defect indicated by white granules from 6 to 8 o'clock position in this eye (Image courtesy: Dr. Deborah VanderVeen)

Persistent Fetal Vasculature (PFV)

PFV contributes to 15–30% of unilateral congenital cataracts [2, 5]. PFV is typically unilateral, but bilateral PFV has been described in 10–15% of cases [11]. Persistence of some or significant portion of fetal vasculature leads to broad spectrum of clinical manifestations of PFV ranging from iridohyaloid vasculature, posterior fibrovascular sheath of the lens (retrolenticular membrane), persistent hyaloid artery, and Bergmeister papilla to severe retinal folds or detachment. PFV is often associated with microphthalmos [11]. The anterior segment vascular remnants are less common in unilateral compared to bilateral PFV [12].

The most common clinical presentation is a faint, small, vascular remnant within the Cloquet's canal, attached to the posterior lens capsule. A less common presentation is the persistence of the entire hyaloid artery with varied amounts of perfusion from the optic nerve to the posterior lens surface. Rarely, it may also be attached to the optic nerve with its anterior end floating freely in the anterior vitreous [13]. Bergmeister papilla represents the remnant of the posterior portion of the hyaloid artery, causing primary congenital malformation of the optic nerve head. Figure 3.4 shows an eye with mature cataract with posterior fibrovascular sheath of lens with prominent ciliary processes.

Anterior Polar Cataract

Anterior polar cataract can be highly amblyogenic, not because it obscures the visual axis but because of induced refractive error, which is most often hyperopic anisometropia and astigmatism. They can be associated with reduced axial length [14].

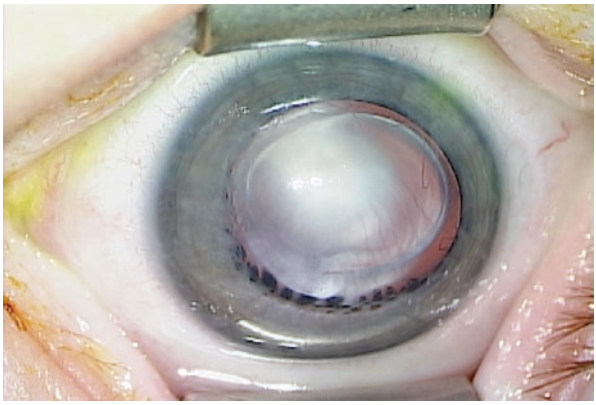

Fig. 3.4 Persistent fetal vasculature (PFV) – Mature cataract with posterior fibrovascular membrane with visible blood vessels and prominent ciliary processes (Image courtesy: Dr. Deborah VanderVeen)

Workup of Unilateral Congenital Cataracts

A thorough ocular exam of both eyes is recommended. The assessment should include age-appropriate visual acuity assessment; note the presence of strabismus, nystagmus, microcornea, intraocular pressure (IOP), refraction, and posterior segment evaluation with fully dilated pupils to look for PCD or PFV. B-scan ultrasonography should be performed if there is a poor view to the posterior segment. No systemic workup is typically recommended for a unilateral cataract.

Eyes with unilateral congenital cataract often have greater central corneal thickness, higher average keratometry values, and smaller corneal diameters. [15]

Outcomes of Pediatric Unilateral Cataracts

IATS is a landmark study in the management of congenital cataract in infants and young children. IATS randomized children 1–7 months of age at 12 sites across the United States to unilateral cataract surgery with or without intraocular lens (IOL) implantation [19]. The outcomes of this study are applicable to unilateral as well as bilateral congenital cataract surgery. Visual acuity, strabismus, stereopsis, and glaucoma outcomes were identical statistically between the IOL and contact lens (CL) group. The infants undergoing IOL implantation proved to have a more complicated course, including higher rates of additional intraocular surgery (72%) and adverse events such as lens proliferation into visual axis (40%), pupillary membrane (28%), corectopia (28%), glaucoma (19%), and glaucoma suspect (9%) [20]. At the end of 5-year follow-up, the study did not demonstrate any visual benefit of implanting an IOL at the time of unilateral cataract surgery in infants younger than 7 months of age [21]. About 50% of treated eyes in both groups had visual acuity of 20/200 or worse owing to deprivational amblyopia [23]. The data led the authors to conclude IOL

implantation should be delayed in infants younger than 7 months at the time of surgery [19–24].

Young age at surgery is a risk factor for the development of glaucoma despite deferring cataract surgery for the first 4 weeks of life. There is 15–25% chance of developing glaucoma after congenital cataract surgery [23–25].

Early cataract extraction and successful optical rehabilitation are important to treat visual deprivation in children with congenital cataract to reduce the incidence of strabismus and nystagmus [18, 26]. The percentage of patients demonstrating strabismus over time increased from 24.6% at baseline to 70.4% by 12 months after cataract surgery. [27]

The same surgeons from IATS sites evaluated the outcomes of unilateral cataract surgery in 56 children aged 7–24 months operated in the same study period. Ninety-two percent received a primary IOL implantation in this group [26, 27]. The incidence of complications, reoperations, and glaucoma was low supporting the relatively safe use of IOLs in children older than 7 months of age [28]. The visual acuity was 20/40 or better only in 11% eyes in this cohort and 20/200 or worse in 44% eyes at 5 years of age owing to deprivation amblyopia [28].

Anterior PFV poses additional challenge owing to its association with microphthalmia and lifelong risk of developing glaucoma and retinal detachment [11, 29–31].

Case 1

A 6-day-old baby girl was urgently referred for absent red reflex in the left eye. She was otherwise healthy and born full term via vaginal delivery. No family history of childhood cataracts. On exam, her vision was blink to light in both eyes. Retinoscopy revealed a refraction of +4.00 +0.50 at 90 degrees in the right eye, but could not be performed on the left due to absent retinoscopic reflex. Corneal light reflexes were centered with Krimsky. Her anterior segment exam was unremarkable in the right and notable for symmetric corneal diameters and an opacity of the left lens. Eye pressures were 7 and 8 mmHg, respectively, with Tonopen. Fundus exam of the right eye was normal and not possible due to lens opacity in the left. B-scan was reassuringly unremarkable.

This girl underwent an uneventful surgery at 4 weeks of age and was left aphakic. She started CL correction and patching treatment on day 6 after the surgery. The visual acuity in the left eye was 20/125 with correction compared to 20/20 in the right eye at 5-year follow-up visit. She has 30–35 prism diopters of intermittent exotropia. She tolerates CL well at the time of writing, but she can be considered for secondary IOL placement if she becomes intolerant to contact lenses or desires functional vision without correction. She might subsequently need surgery for strabismus correction.

Comment This fits into a classic presentation of unilateral congenital cataract. It is critical to diagnose unilateral cataracts early. The eye with unilateral cataract has

deprivation amblyopia from day 1 after birth owing to competition from the healthy contralateral eye. The critical period for unilateral congenital cataract surgery is before 6 weeks of age, compared to 12–14 weeks for bilateral cataracts [16, 17]. Cataract surgery before 4 weeks is associated with greater prevalence of secondary membrane formation and glaucoma [16]. This child should be operated between 4 and 6 weeks of age, ideally at age 4 weeks.

It is important to counsel the family that cataract surgery is the beginning of treatment. Preoperative counseling should stress the importance of full-time optical correction and amblyopia management after surgery. Intraocular lens (IOL) implantation is not recommended in the first 7 months of life even in unilateral cataracts. IOL implantation can be offered in children older than 7 months at the time of surgery. When the child is left aphakic at the time of primary surgery, CL fitting should be performed within the first week after surgery. Extended wear silicone CL (Silsoft, Bausch and Lomb, Bridgewater, NJ) is preferred in this age group. It is important to remember that infants with aphakia are unable to accommodate and should be overcorrected by +2 or + 3 Diopters (D) to focus vision at a near viewing point. The families are advised to remove the CL at least once a week; however, it is recommended that the parents should try to manipulate CL every 2–3 days so that the parents as well as the child are used to the manipulation. The aphakic glasses are not a good option for optical correction in unilateral cataracts given the large amount of anisometropia. Amblyopia treatment should be initiated right after the CL fitting within the first week after surgery.

It is important to mention to the family that the eyes with unilateral congenital cataract are always left with some residual deprivation amblyopia despite early surgical intervention, full-time optical correction, and amblyopia management. The presence of strabismus or nystagmus in preoperative evaluation is an indicator of severe deprivation amblyopia and is more common in children who are diagnosed late [18].

Case 2

A 7-year-old girl was found to have unilateral cataract in her left eye at her annual physical exam. She presented to an outside ophthalmologist who gave her glasses correction for full-time use. She was recommended to undergo cataract surgery in the left eye at a follow-up visit 8 weeks later. The parents were conservative and wanted to pursue surgery only if it was really necessary, so they brought her for a second opinion. She was otherwise healthy except for asthma, for which she uses nebulizer as needed. There is no family history of childhood cataract.

On exam, her visual acuity was 20/20 in the right eye and 20/40 + 1 in the left with correction. Her current glasses were +0.75 D in the right eye and +1.25 D in the left eye. Her cycloplegic refraction showed more hyperopia: right eye, +3.25 D; left eye, +4.00 D. Sensorimotor exam did not demonstrate any strabismus or nystagmus. She had 60 arc seconds of stereoacuity. Anterior segment exam of the right eye

Fig. 3.5 Patient at age 9 wearing glasses well, having never undergone surgery for lens opacity in the left eye

was unremarkable; the left eye had a 2 mm posterior capsule plaque cataract, temporal to the central axis with a clear zone nasally, superiorly, and inferiorly allowing refraction and fundus examination. Fundus examination was unremarkable in both eyes.

This girl likely had congenital or developmental unilateral cataract but was diagnosed late at age 7 years. Given the presence of cataract and anisometropia, she had deprivation amblyopia in the left eye that had become established over the years, though it was mild. Her vision was 20/40 with correction in the left eye, but she had good stereopsis with no strabismus. Given that she had established deprivation amblyopia, she may not significantly benefit from cataract surgery. The cataract was not associated with strabismus and allowed refraction and fundus evaluation, which indicated that it was not visually significant. The decision was made to just monitor her closely for cataract progression. She did patching treatment for 6 months (2 to 4 hours per day), but it did not improve her vision. Her cataract and vision have remained stable for 1.5 years without surgical intervention until the last follow-up visit (Fig. 3.5).

Comment This case illustrates that visually insignificant unilateral cataracts that are not progressive can be monitored safely. They can be treated with pupillary dilation, refractive correction, and patching in early stages to promote visual development.

Early diagnosis and prompt treatment are important factors for optimizing visual outcomes for visually significant unilateral congenital cataracts. The compliance and cost of optical correction with contact lenses stay a challenge in unilateral congenital cataracts operated in the first 7 months of life. IOL implantation is a safe option in older children. The amblyopia management plays a key role in improving visual prognosis. The visual outcomes are limited by early onset deprivation amblyopia in unilateral congenital cataracts.

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Chapter 4
Bilateral Congenital Cataracts

Anna G. Escuder and Deborah K. VanderVeen

Congenital cataract is the leading cause of preventable childhood blindness [1]. The incidence of congenital cataract is 1–6 cases per 10,000 live births in developed countries [2] and 5–15 cases per 10,000 in the developing countries [3]. Globally, an estimated 200,000 children are bilaterally blind from cataracts [4]. Due to the visual deprivation associated with complete or central opacities, successful management requires early detection and treatment, since the critical period of visual development lasts until about 4 months of age for bilateral cataracts and 2 months of age for unilateral cataracts [5]. For bilateral congenital cataracts, the best visual acuity results are typically associated with surgery prior to 14 weeks, during which time there is a trend for better visual acuity with earlier surgery [6]. With regard to classification and management, pediatric cataracts are typically categorized as bilateral or unilateral and into the following groups:

1. Isolated congenital cataracts (hereditary or sporadic)
2. Cataracts associated with ocular developmental anomalies
3. Cataracts that are part of multisystem genetic or metabolic disease [7]

Examination

Prior to examination, a detailed prenatal, birth, medical, and family history should be obtained. A thorough ocular examination is an important part of the workup of congenital cataracts. Visual acuity testing is typically not possible in young infants, who may only demonstrate light responses. Older infants should be able to fix and

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follow with each eye, in a central, steady, maintained fashion, with no nystagmus. External features should be noted as typical or dysmorphic. A portable slit lamp can be used in infants to assess any corneal opacities, iris vascular anomalies, undilated pupil size and shape, and cataract morphology. The red reflex and view to the fundus should be assessed prior to dilation. Indirect ophthalmoscopy should be used to evaluate the fundus, with special attention to persistent fetal vasculature, optic nerve, and other retinal anomalies. If the cataract is too dense to obtain a view to the posterior pole, a B-scan ultrasound can be performed. Finally, review of family photos in an older infant may help elicit the timing of cataract onset. Examination of family members may assist in determining familial etiology. Central cataracts >3 mm in diameter are generally visually significant.

Classification of cataract morphology can sometimes help suggest an identifiable hereditary or genetic cause. Central cataracts include nuclear, lamellar, cortical, sutural, pulverulent, and cerulean. A detailed summary of morphologies can be found in Trumler and Krishnamurthy's reviews [8, 9]. Polar cataracts can be either anterior (anterior polar, anterior pyramidal, and anterior subcapsular) or posterior (posterior subcapsular, posterior lenticonus, posterior fetal vascular [PFV]) [10]. Nuclear cataracts are bilateral in up to 80% of cases, and many affected eyes are microphthalmic [11]. Bilateral nuclear cataracts represent the most frequent morphology for autosomal dominant inherited cataracts. Phenotype alone can occasionally suggest a specific genetic cause; however, in most cases, cataract structure is insufficient to predict a specific gene mutation, since mutations at different loci within a gene can result in different phenotypes, and different gene mutations can result in similar phenotypes [12].

Inherited Bilateral Cataracts

Hereditary cataracts are typically isolated and inherited in an autosomal dominant pattern with a high degree of penetrance [10]. History is very important to determine any family history of congenital cataract, as there will often be several family members with a similar condition, and parents and/or siblings of the affected infant can be examined. Most mutations are in genes for lens crystallins and connexins [13]. Autosomal recessive and X-linked inheritance patterns have been described, but these are less common. Hereditary cataracts account for 12–30% of all congenital cataracts [14, 15–17]. Gene panel testing can also be offered to identify not only mutations in a particular family but also pathogenic mutations in genes that cause cataract for sporadic bilateral cases, some of which may be passed on to future generations.

Non-inherited Bilateral Cataracts

In patients where there is no known family history, it is important to establish whether the cataracts are isolated and, if not, determine an identifiable cause. Isolated cataracts could be due to a sporadic gene mutation but can also be associated with other ocular abnormalities or systemic or metabolic syndromes. In an

infant with bilateral cataracts, note in the history any prior retinopathy of prematurity treated with laser, radiation exposure, steroid use, or trauma, which are each secondary causes of cataract. Cataracts should only be considered "idiopathic" if other causes are ruled out. Older studies have estimated that idiopathic cataracts comprise about 50% of congenital/infantile cataracts [14, 15].

Cataracts associated with ocular abnormalities can be seen in microcornea or microphthalmos, aniridia (evaluate for Wilms tumor, especially in sporadic aniridia), Peters anomaly, or other types of anterior segment dysgenesis. It is also crucial to rule out intraocular tumors. These abnormalities should be apparent during the ophthalmologic evaluation, and ultrasound testing should be done if there is no view of the posterior segment. Some forms of anterior segment dysgenesis with cataract are associated with genetic causes (e.g., *PAX6* mutations) with most findings limited to the eye. However, others may have systemic implications (e.g., *B3GLCT* gene mutation, Peters plus syndrome with cleft lip/palate, short stature, abnormal ears, and mental retardation) [18, 19].

Careful physical examination of the child will provide clues into any genetic or systemic conditions associated with congenital cataracts, as well as tailor the systemic workup. Abnormal facial and orbital features, as well as dermatologic/hair, skeletal, genitourinary, and gastrointestinal (failure to thrive or vomiting), may have a correlation and are worth noting. With assistance from the pediatrician or geneticist, noting the head circumference (presence of hydrocephalus or encephalocele), ear appearance, presence of hearing loss, syndactyly/polydactyly, nasal appearance, presence of cleft lip/palate, or dental abnormalities is valuable when seen in conjunction with cataracts. A highly extensive list of multisystemic associations with syndromic cataract can be found in Trumler's 2011 review [9]. A large Danish observational study showed cataracts with systemic abnormalities are bilateral in 89% of cases [14]. Down syndrome comprised almost a third of cases, the majority of which (72.2% [13/18]) had bilateral cataracts. Patau syndrome (trisomy 13) and Edwards syndrome (trisomy 18) also commonly have the feature of cataract [5]. These syndromes are typically diagnosed by characteristic phenotypical features and confirmed by karyotype testing.

Extensive systemic evaluations cannot feasibly be performed on every child with non-hereditary bilateral cataracts, and assessments are highly unlikely to yield abnormal results in a well, non-dysmorphic infant [20]. In a retrospective study of 421 cases of pediatric cataract in Australia, no child who was otherwise well and had cataract was found to have an associated syndrome on further investigations [21]. However, it is prudent to enlist the assistance of the pediatrician when considering a need for further metabolic and genetic testing, tailored to the medical and developmental history of the child. In the United States, routine newborn screening often includes evaluation for infectious exposures and some metabolic conditions. The US Department of Health and Human Services provides recommendations of core conditions that each state may include in newborn screening panels, and this information is available online or can be confirmed in the child's health record [22].

In the case where newborn laboratory testing has not been performed, it is crucial to elicit any history of intrauterine infections or exposures in utero. Routine testing of all bilateral cataracts for TORCHS (toxoplasmosis, rubella, cytomegalovirus, herpes simplex, and syphilis) infection should be performed. A history of maternal fever/rash during pregnancy or systemic clinical indicators in the child such as

microcephaly, hearing loss, developmental delay, thrombocytopenia, hepatosplenomegaly, or skin abnormalities should alert the clinician to possible infectious etiology [9]. Maternal and fetal rubella infection usually occurs in the first trimester and can manifest with growth deficiency, microcephaly, cardiac abnormalities, deafness, and ocular manifestations (cataract, glaucoma, retinopathy) [5].

Metabolic disorders that have not yet manifested systemic symptoms can be found in infants with bilateral cataract. Urine amino acids and serum electrolytes can be checked especially in males with cataracts, hypotonia, poor weight gain, and mental retardation with concern for Lowe oculocerebrorenal syndrome [10]. Lowe syndrome is an X-linked recessive syndrome, which is also frequently associated with glaucoma and corneal keloids [7]. Galactosemia is an autosomal recessive condition caused by mutations in galactokinase (*GALK1*), galactose-1-phosphate uridylyltransferase (*GALT*), or uridine disphosphate galactose-4-epimerase *(GALE)*, which result in high-serum galactose (which can be measured with urine galactitol). In the more common transferase deficiency, symptoms may become apparent when the child starts drinking whole milk and can present with vomiting, failure to thrive, liver disease, and lethargy [23, 9, 5]. Generally, these patients are diagnosed by systemic symptoms prior to development of cataract. These patients should be screened for urine-reducing substances. Erythrocyte galactokinase can also identify the less common *GALK1* deficiency [5]. Sengers syndrome is a metabolic disorder associated with cardiomyopathy (mutation in the *acylglycerol kinase* gene, chromosome 7q34); asymptomatic and undiagnosed cases can be detected by cardiomegaly on chest X-ray. Spoke-like cortical cataracts can also be seen in lysosomal storage diseases (alpha-mannosidosis and the X-linked Fabry disease), though these are typically not seen in infants.

Cat-Map (http://cat-map.wustl.edu/) is an online chromosome map and reference database for inherited and age-related forms of cataracts in humans and other selected animals [24]. Hejtmancik also summarizes the different types of genes in detail associated with congenital cataracts [13]. In the future, it may be possible to find the genetic cause for more sporadic cataracts using next-generation sequencing [20].

Case 1

An 11-day-old full-term baby girl presented to the pediatric ophthalmologist after referral from the pediatrician for an abnormal red reflex in both eyes on newborn screening. Her sister, father, and paternal grandmother had a history of congenital cataracts, for which they underwent surgery at young ages. On examination, she was a well-appearing full-term baby, who opened her eyes spontaneously and blinked to light in both eyes. Anterior segment exam with portable slit lamp showed a central 4 mm nuclear cataract in each eye (Fig. 4.1), with a clear peripheral lens. The corneas were clear and estimated to be of normal diameter (about 10 mm). The conjunctiva was white and quiet bilaterally. The pupils were normally reactive without afferent pupillary defect. The iris was blue, and the pupils were round without any synechiae or persistent iris fetal vasculature. Intraocular pressures were soft to tactile palpation bilaterally. The dilated fundus exam showed normal optic nerve without evidence of persistent stalk, normal macula, vessels, and periphery in each eye.

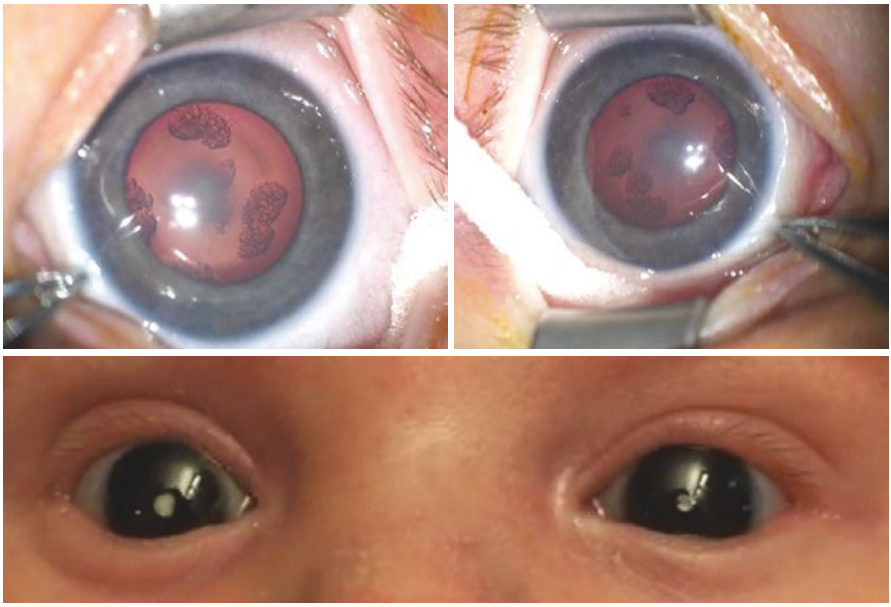

Fig. 4.1 Intraoperative photos of bilateral hereditary cataracts of Case 1, the right eye and the left eye, demonstrating central nuclear opacities with peripheral vacuoles

No additional workup was done due to the strong family ocular history of hereditary cataract in a well child. The patient underwent bilateral (immediate sequential) 23-gauge cataract extraction with primary posterior capsulectomy and anterior mechanical vitrectomy at week 4 of life. Surgery was uneventful and the patient was left aphakic. She was fitted with aphakic soft contact lenses at postoperative day 5 (Silsoft base curve 7.5, diameter 11.3, power +29.00 diopters in each eye).

Comment In this case, the strong family history likely contributed to the early identification of bilateral cataracts by the child's pediatrician and prompt referral. Workup was not necessary with such a strong family history, but genetic testing could be offered if desired.

Case 2

A 13-day-old full-term baby girl was seen after ophthalmology was consulted to evaluate for leukocoria. Her parents reported "white pupils" in both eyes since birth but stated that she responds to light. There is no family history of childhood cataracts.

Her pregnancy was uncomplicated with no history of intrauterine infections. She was born at an outside hospital by spontaneous vaginal delivery, with heart rate deceleration and cooling protocol initiation given concern for hypoxic encephalopathy. She completed a sepsis rule out and was monitored for supraventricular tachycardia (SVT) by cardiology. An echocardiogram done on day 4 of life showed a patent foramen ovale with normal anatomy and systolic function. The SVT was successfully treated with Sotalol. On examination, she had a non-dysmorphic and symmetric

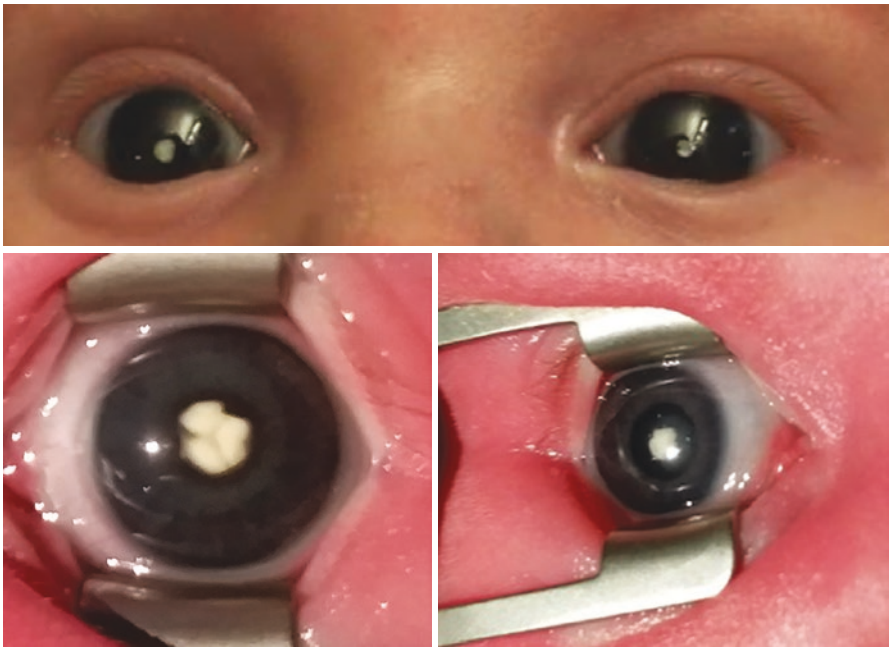

Fig. 4.2 Top photo shows preoperative miotic pupils with dense white central cataracts of patient no. 2. Bottom photos of the same patient show intraoperative dense white chalky cataracts, the left greater than the right, with poor pupillary dilation

facial appearance. On ocular exam, she unreliably blinked to light in both eyes, pupils were very small and poorly reactive, and there was no red reflex bilaterally. Hand light examination showed clear corneas (9 mm) and normal anterior segments, with central dense chalky white cataracts and poor dilation bilaterally (Fig. 4.2). There was no view to the posterior pole; B-scan ultrasound showed attached retina, clear vitreous, and no masses. Given the lack of family history for childhood cataracts, a systemic infectious, metabolic, and genetic workup was performed.

The genetics and metabolism teams examined the baby. She underwent evaluation for metabolic causes of cataract not included in her state newborn screening, such as galactosemia (urine galactitol) and Lowe syndrome (urine and plasma amino acids, serum electrolytes). There are 32 disorders included in required newborn screening in Massachusetts, which can be found in detail through the New England Newborn Screening Program [25]. The genetics team did not find any additional dysmorphisms and ordered genetic testing using a Cataract Panel (GeneDx, https://www.genedx.com/test-catalog/available-tests/cataract-panel/). The methods utilized by this panel are expected to detect over 99% of sequencing variants present in the covered regions of the majority of genes known to be associated with cataract. This patient had a heterogeneous mutation in the major intrinsic protein (*MIP*) gene, which was reported as a "likely pathogenic variant – most likely consistent with diagnosis of autosomal dominant MIP-related cataract." The *MIP* gene encodes the major intrinsic protein of the ocular lens fiber membrane, also

referred to as aquaporin-0. MIP belongs to the aquaporin family of water channels. It is involved with water transport across lens cortical fiber cell membranes and may be involved in fiber-fiber adhesions, functions which are important in keeping the lens transparent [26]. The genetics team determined that this mutation was the most likely etiology of her cataracts, but not as a cohesive unifying diagnosis for her history of SVT and perinatal stress requiring protocol cooling. They recommended monitoring head circumference, and if there were out of proportion growth or craniosynostosis, they would re-evaluate for other genetic causes of her constellation of symptoms.

At 34 days of life, this patient underwent bilateral (immediate sequential) and uncomplicated 20-gauge cataract extraction with primary posterior capsulectomy and anterior vitrectomy. She was left aphakic and fitted for aphakic contact lenses in each eye the following week (Silsoft base curve 7.5, diameter 11.3 mm, power +32.00 diopters in each eye).

Comment Once an infant has been diagnosed with bilateral cataract that is nonfamilial, prior to proceeding with surgery, we recommend confirmation from the

Table 4.1 Stepwise approach to ancillary testing for bilateral infantile cataracts

History	Inquire about pre- or postnatal exposures, infections, trauma	
	Examine parents and siblings to determine hereditary etiology	
	Communicate with pediatrician regarding what testing was done in newborn screen and if any concerns for associated medical conditions	
Physical exam and systemic associations	Dysmorphic facies or other organ systems	
	Failure to thrive	
	Developmental delay	
Laboratory testing[a]	Infectious	Titers for toxoplasmosis, rubella, cytomegalovirus, herpes simplex, syphilis (VDRL)
	Metabolic	Galactosemia: urine-reducing substances; erythrocyte galactokinase (less common *GALK1* deficiency)
		Lowe syndrome: urine and plasma amino acids; serum electrolytes
		Others: glucose, calcium, phosphorus
		Chest X-ray: cardiomyopathy (Sengers syndrome)
Genetic testing	Next-generation sequencing[b] or	
	GeneDX Cataract Panel	

Reprinted from Vanderveen [5]. With permission from Elsevier
[a]When not included in newborn screening
[b]Musleh et al. [20]

pediatrician that the child is well and non-dysmorphic. If there is any concern for a syndromic or metabolic diagnosis, referral to a geneticist is recommended and further workup tailored to the findings. If the child is well, confirmation that the newborn screening tests have ruled out conditions that may require intervention or add risk to the anesthetic or surgical exposure should also be performed. Table 4.1 lists the classic and more common conditions that can be identified in these patients.

Pediatric cataracts occur in 1–15 per 10,000 births, with 60% being bilateral [9]. A detailed family history (possibly with examination of family members) and a prenatal history and current health history are important. Collaboration with the pediatrician to determine whether further testing for metabolic, genetic, or syndromic disease is needed is paramount to successful management. Completion of a thorough ocular and physical examination can provide helpful clues in identifying cataract cause and targeting the workup. Care is required to identify and treat underlying systemic conditions, when present. Genetic testing is available and may help identify causes for "idiopathic" cases.

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Chapter 5
Secondary and Acquired Cataracts

David George Morrison and Allison Carol Umfress

General Workup for Acquired Cataracts

Cataract is the most common cause of treatable visual disability in children worldwide and affects approximately 500 children annually in the United States. The prevalence has been reported to range between 1 and 15 per 10,000 children [1]. When evaluating a child with acquired cataract, several factors need to be considered. As with all medical issues, history is paramount. Cataracts can be genetic, associated with other diseases, or idiopathic in nature. Although many hereditary cataracts present at birth, lens opacities such as posterior lenticonus and lamellar opacities may have a relatively clear visual axis and avoid diagnosis until worsening later in childhood. Family history of these types of lens opacities may allow the clinician to avoid extensive workup for other diseases when present.

Other syndromic diseases such as trisomy 21, Lowe syndrome, and myotonic dystrophy can have associated cataract formation later in life. Careful history and evaluation for syndromic morphology are important when evaluating a child with developmental cataract.

Numerous systemic diseases can be associated with the development of cataract. Galactosemia is the prototypical systemic disease associated with cataract development. Due to the lack of enzymes necessary for the metabolism of the galactose sugar molecule, metabolites collect in the lens, and "oil droplet" cataract formation results. Galactosemia is part of normal newborn nursery testing but may be missed if the infant has not received a sufficient milk feed prior to discharge. Type 1 diabetes mellitus (T1DM) is another systemic disease where cataract formation can occur.

Appropriate laboratory workup is needed for children with acquired bilateral cataracts of unknown origin. At a minimum, a comprehensive metabolic panel (CMP) with fasting blood glucose and red blood cell galactose-1-phosphate is

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necessary. Depending on other suspected diagnoses, a number of additional laboratory tests including fatty acid and peroxisomal profile, very-long-chain fatty acids (VLCFA), plasma amino acid profile, plasma and serum cholestanol levels, and genetic testing may be warranted. Rare disorders of peroxisomal metabolism including Zellweger syndrome, Refsum disease, or X-linked adrenoleukodystrophy can be diagnosed with elevated serum levels of VLCFA. Refsum disease is a progressive neurologic disease caused by deficient alpha-oxidation of phytanic acid, leading to accumulation of this substance in peroxisomes and throughout body tissues, including the lens, retina, and vitreous. The condition can be confirmed with elevated serum phytanic acid levels. In severe cases, these syndromes can present with intellectual disability, hypotonia, and seizures but in the early stages may present only with cataract or subtle signs of chondrodysplasia or craniofacial abnormalities. Lowe syndrome (oculocerebrorenal syndrome) is a rare X-linked recessive disorder associated with cataracts, hypotonia, intellectual disability, proximal tubular acidosis, aminoaciduria, and proteinuria that can be evaluated by checking the urine or plasma amino acid levels.

Finally, many cataracts may be associated with the presence of other ophthalmic diseases. The presence of other anterior or posterior segment findings is critical on exam to best determine the origin of the cataract and necessary treatment. Uveitis associated with juvenile idiopathic arthritis (JIA) is the most common cause of cataract formation associated with anterior segment inflammation. However, any form of ocular inflammation due to infectious or noninfectious etiology may cause cataract formation. Drug exposure, especially to systemic steroids often used to treat these conditions, can cause posterior subcapsular cataracts.

Cataracts with Associated Anterior Segment Abnormalities

When evaluating children with acquired cataract, a careful examination for associated anterior segment findings can give insight into the etiology of the cataract. The most common association seen with secondary cataract in children is intraocular inflammation, most often related to juvenile idiopathic arthritis (JIA). Up to 70% of children with chronic uveitis develop cataracts [2]. In children with uveitis, cataract can be secondary to the inflammation itself, iatrogenically induced from steroid use, or a combination of both. However, the popularization of steroid-sparing agents and earlier diagnosis of uveitis through routine screening for those children with JIA has lessened the severity and prevalence of secondary cataract formation in this population.

Cataracts caused primarily by steroid use are typically posterior subcapsular. Associated findings on exam that suggest a history of inflammation include keratic precipitates, anterior chamber cell and flare, posterior synechiae, iris atrophy, vitreous cell or haze, snowballs and snowbanking, vasculitis, or chorioretinal lesions. These associated ocular abnormalities often make the cataract surgery more technically challenging, requiring synechiolysis, and these patients are at higher risk for

postoperative complications including flare of intraocular inflammation, glaucoma, and retinal detachment [3–6].

Traumatic injuries are the most common cause of unilateral vision loss in children. Boys are more commonly affected by trauma than girls. These injuries commonly occur during sports activities but may also be secondary to environmental hazards such as firecracker injuries, BB pellets, and airbags and glass from motor vehicle accidents. Both blunt and penetrating trauma can lead to cataract formation. Blunt trauma classically leads to stellate-shaped posterior opacities, which can be stable or progressive. Penetrating trauma leads to more focal lens changes that can rapidly opacify in the setting of lens capsule violation. Other signs of prior ocular trauma include corneal or scleral lacerations, zonular dehiscence, angle recession, hyphema, corneal blood staining, and peripheral anterior synechiae (Fig. 5.1). Careful ocular examination is also essential in these cases to rule out a retained ocular or orbital foreign body or open globe injury.

The management of traumatic cataract in children is variable and depends on the age of the child at the time of injury, development of amblyopia, duration of cataract

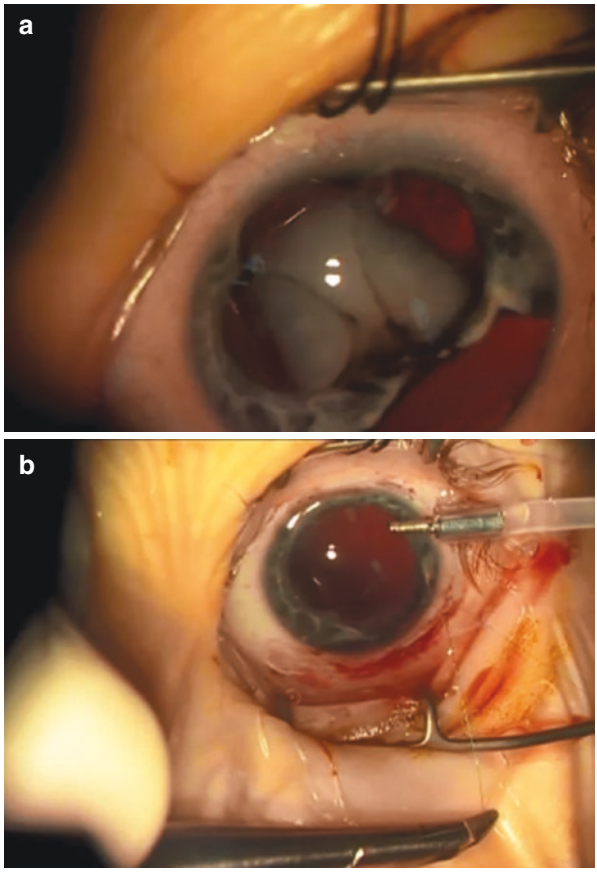

Fig. 5.1 Traumatic cataract with iridodialysis before (**a**) and after (**b**) repair with lensectomy and McCannel suture

presence, and the associated ocular injuries and the state of the posterior capsule, angle structures, and zonules.

Other rare secondary causes of cataract in children include occult ocular disease, such as intraocular tumor or chronic retinal detachment, and congenital diseases in which lens opacity manifests later in the disease course. Neurofibromatosis type 2 is commonly associated with posterior subcapsular cataract. If previously undiagnosed, Lisch nodules or café au lait spots can assist with diagnosis. Wilson's disease can present with sunflower cataract and associated Kayser-Fleischer ring. Atopic dermatitis can be associated with characteristic shield-shaped anterior subcapsular cataracts, periocular dermatitis, conjunctivitis, and keratitis. Children with radiation-induced cataract often have severe ocular surface disease and may be less likely to tolerate a contact lens, making intraocular lens insertion a higher consideration.

Case 1

An 11-year-old female presented in referral from the retina service for bilateral cataracts. She had a history of HLA-B27-associated chronic bilateral anterior uveitis. She had been previously treated with topical corticosteroid eyedrops, oral prednisone, and an intravitreal steroid injection in the right eye. She ultimately required systemic immunosuppression with methotrexate and adalimumab for control of her disease. At the time of presentation for cataract evaluation, her intraocular inflammation had been controlled for 6 months. Medical history was otherwise unremarkable. Rheumatologic evaluation was negative for any systemic disease.

Visual acuity was 20/200 in the right eye and 20/20 in the left eye. She was orthophoric with full ocular motility. Pupils were equal, reactive, and without afferent pupillary defect. Anterior segment examination demonstrated deep and quiet anterior chamber with bilateral posterior synechiae and nuclear sclerotic cataracts, the right eye greater than the left eye (Fig. 5.2).

The decision was made to proceed with cataract extraction with intraocular lens placement in the right eye. The patient was given a course of perioperative oral prednisone and IV dexamethasone on the day of surgery. Cataract extraction and synechiolysis was performed without complication, and an intraocular lens was placed in the capsular bag. Postoperatively, she recovered well but developed posterior capsular opacification requiring YAG capsulotomy, which she underwent 6 months after cataract surgery. At her follow-up visit after YAG capsulotomy, her visual acuity had improved to 20/25.

Comment This case describes the most common type of secondary cataract in children – occurring as a result of uveitis, usually in association with arthritis (JIA) or any etiology of intermediate or posterior uveitis. In the setting of known uveitis, it is recommended that cataract surgery be deferred until the inflammation has been controlled for at least 3 months, and perioperative steroids should be considered [3, 7]. In the described case, inflammation had been controlled for over 6 months.

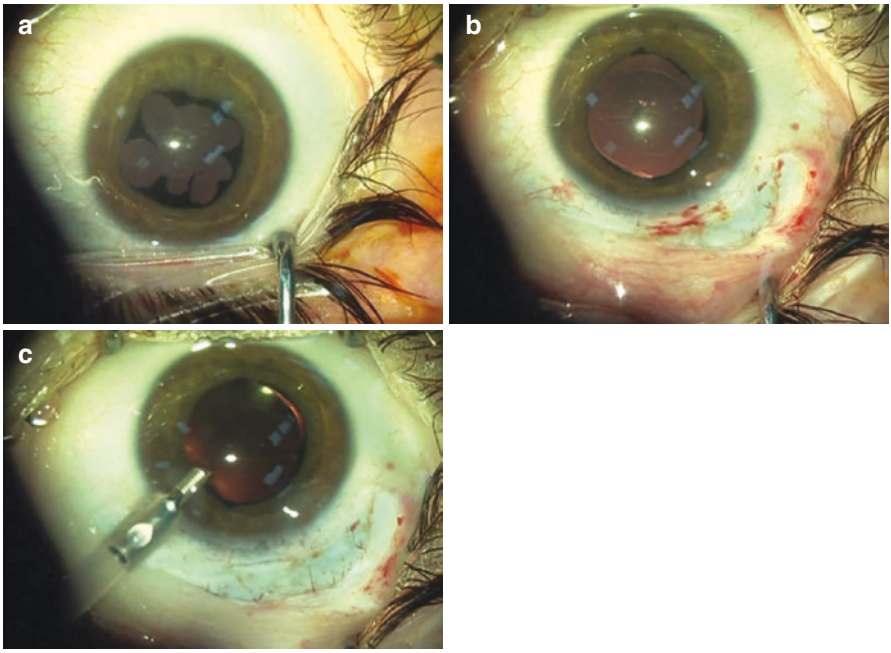

Fig. 5.2 Posterior synechiae after dilation (**a**), after synechialysis and capsulorhexis (**b**), and after IOL implantation (**c**)

Several studies have demonstrated improved postoperative visual acuity outcomes with lower rates of complication with perioperative corticosteroids and systemic immunosuppression [8, 9]. Several authors have described their approach to perioperative steroid treatment, which generally consists of oral prednisone 2–4 days preoperatively followed by a taper 1–4 weeks postoperatively [10–15]. Traditionally, IOL implantation was avoided in patients with uveitis due to the risk of cyclitic membrane formation [16]. However, with improved perioperative control of inflammation, intraocular lens implantation is now performed regularly with success (for more information, see Chap. 21: IOL Placement in the Uveitic Patient) [8, 9, 13, 17–19]. The patient described has been followed for 5 years and subsequently underwent pars plana vitrectomy for removal of vitreous opacities and dense posterior capsule opacification (PCO) but has maintained best-corrected vision of 20/25 without further issues arising from IOL implantation.

Case 2

A 3-year-old otherwise healthy boy presented for evaluation in referral from his optometrist for bilateral lens opacities. His parents reported that he had been having difficulty seeing the television compared to other family members for the last several months.

Vision was central, steady, and unmaintained in the right eye and central, steady, and maintained in the left eye. No red reflex was visible in either eye. Slit lamp exam revealed bilateral white posterior lens opacities greater in the right eye than the left. B-scan showed attached retina with no masses.

Decision was made to proceed with cataract extraction, and this was scheduled approximately 1 month after presentation. Intraoperatively, after the lens was removed, the patient was noted to have dense vitreous haze obscuring the red reflex. Laboratory workup including complete blood count and comprehensive metabolic panel as well as testing for toxoplasma titers, toxocara titers, syphilis, Lyme titers, Quantiferon Gold, and a chest X-ray was obtained intraoperatively for evaluation. Postoperatively, he recovered well and was referred to the retina service for further management.

Despite an extensive workup, no etiology was identified. The patient was ultimately diagnosed with chronic bilateral panuveitis. He later required pars plana vitrectomy in the right eye and cataract extraction with pars plana vitrectomy in the left and was initiated on methotrexate for control of his intraocular inflammation under the direction of the retina and rheumatology services.

Comment In this case, cataract was the first presenting sign of ongoing intraocular inflammation that had been previously undiagnosed. In children with acquired cataract, evaluation for uveitis should be included as part of the initial workup through a detailed medical history and slit lamp examination. JIA is the most common form of chronic uveitis in children and typically presents with anterior uveitis. However, posterior uveitis accounts for a greater proportion of uveitis cases among children than among adults [20], especially in young children [21]. In atypical cases, alternate diagnoses including toxocariasis, toxoplasmosis, sarcoidosis, tuberculosis, tubulointerstitial nephritis and uveitis, syphilis, and herpes simplex should be considered. In particular, sarcoidosis is a much more common cause of ocular inflammation in young children than among older children or adults, and anterior uveitis is the most common intraocular manifestation of sarcoidosis in children [22].

Cataracts Without Associated Anterior Segment Abnormalities

Acquired cataracts without associated ocular abnormalities are less common in children. In these cases, one must often rely on the clinical history and evaluation for systemic disease to ascertain the underlying etiology.

In children, systemic metabolic diseases must be considered in cases of acquired cataract. Diabetes mellitus is one of the most common metabolic etiologies of acquired cataract in children, but other rarer metabolic diseases should be considered on the differential. Next-generation sequencing has allowed for precise diagnosis of rare systemic metabolic disorders such as disorders of lipid and peroxisome

metabolism, Lowe syndrome, and Hallermann-Streiff syndrome if the classic associated systemic signs and symptoms are not clinically apparent.

Other etiologies of acquired cataract in otherwise normal eyes include iatrogenic sources such as systemic steroid use or post-vitrectomy cataract. Systemic steroids rarely lead to the development of cataract in children and are generally only seen with high doses of oral corticosteroids used for treatment of uveitis or autoimmune disease and are not associated with the use of inhaled corticosteroids for asthma in children [23]. Development of cataract after vitrectomy occurs in as many as 60% of children [24]. Other systemic medications may rarely be associated with cataract development in children including topical anticholinesterases and systemic phenothiazines.

Case 3

A 3-year-old girl presented as a referral from her pediatrician for an abnormal red reflex in the right eye for 2–3 weeks. Her medical history was significant for sensorineural hearing loss and congenital hypotonia. Cochlear implant surgery had been performed. She had seen genetics and had a negative genetic workup associated with the hearing loss.

Visual acuity was reaction to light in the right eye but no fix or follow. The left eye was central, steady, and maintained with good fix and follow. Slit lamp examination revealed lens opacities in both eyes, the right much greater than the left. Dilated fundus examination was unable to be performed in the right eye, but the left eye had a normal optic nerve and fundus exam. B-scan showed attached retina and no retinal masses.

She was scheduled for cataract extraction with intraocular lens placement on the right eye first. At the time of surgery, labs included a comprehensive metabolic panel (CMP), red blood cell galactose-1-phosphate, peroxisomal profile, fatty acid profile, VLCFA and phytanic acid, plasma amino acid profile, and plasma acylcarnitine. All labs were negative; however, her fasting blood sugar on CMP was 203. She was diagnosed with juvenile-onset type 1 diabetes mellitus. Her cataracts were the result of lens hydration due to the osmotic gradient from uncontrolled diabetes.

Comment In this case, cataract formation was the first presenting sign of type 1 diabetes mellitus. Approximately half a million children worldwide are affected by type 1 diabetes mellitus (T1DM) [25]. Diabetic cataract has been shown to occur in 0.7–3.4% of pediatric diabetic patients, classically the bilateral snowflake cortical deposits [25, 26]. In most pediatric patients, diabetic cataract either is the presenting sign of T1DM or occurs within the first 6 months of diagnosis [25]. This is in contrast to other ocular findings associated with diabetes mellitus such as diabetic retinopathy, which typically does not present before puberty, and for which screening is generally not recommended before 10 years of age [27, 28].

Case 4

A 5-year-old male was referred for evaluation of leukocoria noticed by his mother for approximately 2 months in addition to some right eye crossing. He had no past ocular history, and mom reported that he had always passed his vision screening tests in school and with his pediatrician. He was seen by his local optometrist with concern for bilateral cataracts and referred for surgical evaluation.

On examination, visual acuity was hand motion in the right eye and 20/60 in the left. He had a variable small-angle right esotropia. Slit lamp examination showed a dense white cortical cataract on the right. The left eye had a cortical opacity with central sparing. There was no red reflex or view to the posterior pole on the right. B-scan ultrasound showed attached retina with no masses. Dilated examination on the left was unremarkable.

The decision was made to proceed with cataract extraction with intraocular lens placement in the right eye. He recovered well postoperatively, and at his 6-month follow-up, vision had improved to 20/60 in the right eye and 20/40 in the left eye, and he was treated with patching for amblyopia. At follow-up visit approximately 2 years later, at age 8, he presented with decreased vision to 20/250 in the left eye with worsening of the cataract in this eye. He was scheduled for cataract extraction of the left eye. Intraoperatively, he was noted to have scarring of the posterior capsule with cortical and posterior capsular opacification consistent with posterior lenticonus (Fig. 5.3).

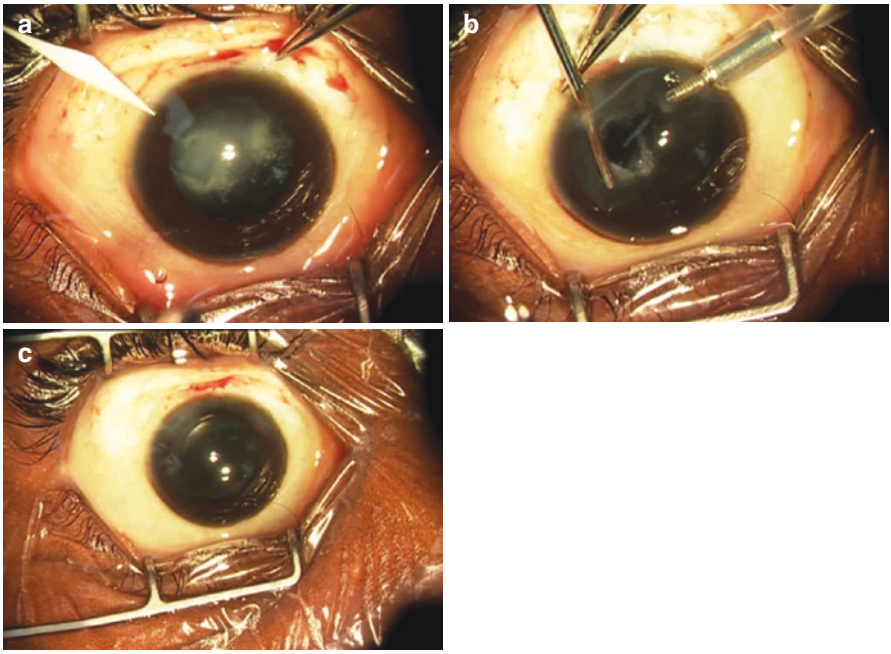

Fig. 5.3 Posterior lenticonus cataract prior to surgery (**a**), with lens removed and posterior capsular defect shown (**b**), and after posterior capsulotomy and sulcus-placed three-piece IOL (**c**)

Comment Posterior lenticonus is a rare condition caused by a protrusion in the posterior capsule of the lens, occurring in approximately 1 in every 100,000 children [29–31]. It is usually unilateral and sporadic, but familial and bilateral cases have been reported [32]. Initially, this condition can be detected by the oil droplet reflex seen on retinoscopy, with the central area of protrusion resulting in a focal area of extreme myopia. Cataract is usually not present at birth but develops later as the intralenticular pressure increases with age. This can progress slowly and the lens may remain relatively clear and patient asymptomatic, or spontaneous rupture of the lens capsule can lead to rapid total lens opacification. In these cases of rapid opacification, the underlying diagnosis may be unknown until the time of surgery.

Acquired cataracts are those that develop later in life, as opposed to congenital cataracts, which are present at birth. These can be secondary to medication or toxic exposures, other ocular diseases or injuries, or systemic diseases with lens opacification as a manifestation. The approach to acquired cataracts should first include an investigation into the underlying etiology. Often, the presence of associated anterior segment signs can lead to the underlying diagnosis. In the presence of an otherwise normal ocular exam, the clinical history, past medical history, and history of medication and toxic exposures are essential to make the diagnosis. It is standard practice for most children over the age of one to have intraocular lens implantation at the time of cataract surgery when able, and good visual outcomes have been demonstrated in children with acquired cataracts with timely surgical intervention and treatment of associated amblyopia.

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Chapter 6
Preoperative Examination

Brita S. Rook and Scott A. Davis

Proper preoperative counseling, planning, and management strategies require attention to a multitude of factors surrounding a pediatric patient presenting with a cataract. A thoughtful approach to these patients considers all of the possible medical and surgical needs of the patient. The timing of surgery, discussion of the surgical plan, preoperative measurements with biometry, and addressing postoperative expectations (aphakic contact lens versus spectacles) are important factors to consider. It all begins with the preoperative visit.

History

First and foremost, obtaining a thorough patient history is essential to understanding possible etiologies and the visual impact of the cataract. Pertinent information from the history includes prenatal and perinatal history, ethnicity, gender, history of maternal infection (TORCH infections), history of ocular trauma, and family history of childhood cataracts. History of juvenile idiopathic arthritis (JIA) can indicate that a cataract is secondary to uveitis and/or long-term corticosteroid use. In addition, the age of onset of visual symptoms and change from previous eye examinations should be ascertained. It is important to question parents about the child's perceived vision, as this can provide clues about the duration of the cataract and its visual significance. Nearly one-third of cataracts are inherited, so an examination of the eyes of the parents may be helpful and spare unnecessary testing associated with a workup [1].

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Examination

Immediately upon entering the room with a pediatric patient, one can begin gathering information about visual behavior and ocular alignment. These observations are easily made without even coming close to the patient.

One of the most useful skills includes the use of the direct ophthalmoscope to visualize the red reflex. It is a fairly quick test that does not require much cooperation from the patient but can provide a lot of information regarding the presence of a media opacity. The Bruckner test may be employed to compare one reflex to the other, allowing the provider to estimate the amount of visual significance produced by the cataract. Determining the location of the cataract and whether or not it is directly in the visual axis can also be noted while observing the red reflex.

Although red reflex testing can be a quick clue to the presence of a lenticular opacity, further testing is necessary to determine the visual significance of a cataract. Obtaining an accurate assessment of visual function in a young child can present a major challenge. Typically, the older and more cooperative the patient, the more likely the physician will succeed at obtaining an accurate visual acuity. Even though it may be difficult, it is not impossible to achieve a good examination in a young child or infant. Allow the child to participate in the exam. Use toys and other attention-grabbing props such as the "barking dog" to help assess the visual function. Encouragement from the physician and from family members can promote better cooperation and thus better exam results. Noting the patient's level of participation during an exam is also an important consideration when interpreting your exam findings.

To assess visual function in a preverbal child, check fix and follow behavior of each eye. Patch occlusion of a preferred eye during the exam may illicit a strong negative response from the child, signaling poor vision in that eye. This of course assumes minimal objection to occlusion of the non-preferred eye. The induced tropia test is also useful in a non-strabismic patient. This is done by placing a 20 prism-diopter base-down prism over an eye, one at a time, and allowing a couple of seconds to determine whether the child fixates on the second image. This prism amount is sufficient to provide two images for the child and two corneal light reflexes for the examiner to allow detection of which eye is being used for fixation [2]. In the strabismic patient, fixation preference can be determined by quantifying how long the non-preferred eye is able to maintain fixation. This may mean that the non-preferred eye immediately switches fixation back to the preferred eye when the cover is removed, indicating a very strong preference. Sometimes, the non-preferred eye can maintain fixation for several seconds after the cover is removed or will maintain fixation up to a blink, still indicating a preference for the other eye but less so than not maintaining fixation at all. Preferential looking techniques, such as Teller acuity cards, can also be used with good reliability in this age group.

In the verbal child, optotype visual acuity testing with HOTV matching, LEA symbols, or Snellen visual acuity testing can be employed. Each eye should be

occluded with a patch and separately tested for distance acuity with one of the aforementioned modalities. If it is determined that one eye is testing better than the other and you are unsure of the reliability of this result, the weaker eye should be tested first on a return office visit. Glare testing may also be useful in determining the visual significance of the cataract.

As mentioned previously, the assessment of the ocular alignment can be helpful in determining the amblyogenic effect of a unilateral cataract. The presence of strabismus typically indicates that the cataract is long-standing. The presence of nystagmus portends a poorer visual outcome, as it indicates visual deprivation from bilateral cataracts beginning in infancy. Sensory nystagmus usually develops around 3 months of age secondary to inhibited development of the fixation reflex that is normally formed by this age [3]. These eyes typically will not see better than 20/100 even after removal of the cataracts [1].

To test ocular alignment, the Hirschberg test is a good place to start and is done by assessing the position of the corneal light reflex. Cover-uncover and alternate cover testing at distance and near are superior for assessing ocular alignment and the degree of strabismic deviation. Overall, gathering information on the presence of strabismus or nystagmus may be useful for providing parents with predictive information regarding potential visual acuity [3]. It is important to counsel parents that the nystagmus is likely to persist after cataract surgery.

The anterior segment examination follows the assessment of visual function and motility. Penlight or portable slit lamp examination of the anterior structures of the eye includes examination of the eyelids and eyelashes, conjunctiva, sclera, cornea, anterior chamber, and iris. In the event that abnormal findings such a blepharitis or nasolacrimal duct obstruction are found, treatment should be initiated prior to cataract surgery. When possible, corneal diameter should be assessed. The pupil examination is important both before and after dilation and can offer further prognostic indicators. Aniridia should be noted as there is a known correlation between this condition and infantile cataracts and glaucoma. Poorly dilating pupils may reflect immaturity of the anterior segment and place the eye at an increased risk of glaucoma after cataract surgery [1].

The slit lamp examination is important to help further classify the morphology, location, and possible etiology of the cataract. Anterior chamber depth should be assessed when possible. Signs of inflammation such as cell, flare, or posterior synechiae indicate an inflammatory etiology. The presence of lens subluxation, iridodonesis, or phacodonesis could indicate prior trauma or a genetic condition as a cause of the cataract [3].

Those children who are able to sit for a slit lamp examination may be able to cooperate for a neodymium-doped yttrium aluninum garnet (Nd:YAG) laser session postsurgically. This bit of information is crucial for surgical planning, as it can help the surgeon to decide whether or not to perform a primary posterior capsulotomy with anterior vitrectomy at the time of surgery.

If there is an adequate view through the cataract, a fundoscopic evaluation of the posterior pole and retinoscopy should be performed. The health of the optic nerve and fovea are important to note and assist in predicting visual outcome

postoperatively. Determining the refractive error of both eyes, if possible, and noting the presence of anisometropia are important in surgical planning. If there is no view through the lens, then B-scan ultrasonography can be done in the clinic or at the time of surgery to rule out posterior segment abnormalities such as a retinal detachment or persistent fetal vasculature.

Some of these exam components will not be possible in the clinic due to the patient's age and level of cooperation. These exam details can be obtained during an examination under anesthesia (EUA) before or at the time of surgery. Intraocular pressure and pachymetry measurements are both important data points to gather but will likely be more accurate in pediatric patients while under anesthesia. Note that it is best to check the intraocular pressure at the time of induction of anesthesia, as general anesthesia can artificially lower eye pressure.

Preoperative Testing

Biometry and Keratometry

Biometry and keratometry are both required for surgical planning regardless of a plan for IOL implantation or contact lens use. Biometry may be obtained successfully in the outpatient setting in children who are cooperative enough to sit for measurements. Infants and younger children who cannot sit for biometry typically have measurements conducted during an EUA preceding their cataract surgery [3]. In the clinical setting, choices for biometry include optical biometry with the IOLMaster (Carl Zeiss Meditec AG) or LenStar (Haag-Streit Diagnostics, USA) or via ultrasound biometry with immersion or contact techniques. In the operating room, measurements are obtained with ultrasound biometry. Keratometry can be performed manually in the cooperative patient; however, various handheld automated keratometers are available for use in clinic or the operating room. It should be noted that the cylinder axis measurement may not be as reliable with the handheld keratometer [3].

Laboratory Investigation

Studies show that nearly 86% of unilateral and 68% of bilateral cataracts have no apparent cause [4]. Therefore, most surgeons do not advocate laboratory workup for congenital cataracts unless the patient's history is concerning. The differential diagnosis of bilateral congenital cataracts includes genetic, metabolic, infectious, inflammatory, and idiopathic causes. The recommended workup is tailored based upon the clinical history and can include TORCH titers to rule out infectious causes such as toxoplasma, rubella, cytomegalovirus (CMV), herpes, syphilis, varicella-zoster virus, and parvovirus B19. Urine amino acid testing can be recommended to

rule out genetic causes such as Lowe or Alport syndrome. Blood glucose testing should be completed to rule out diabetes.

Case 1

A 4-day-old male infant in the neonatal intensive care unit (NICU) presented with a poor red reflex in both eyes. The baby was a term infant but had been sent to the NICU for persistent hypotonia, difficulty feeding, and hyperbilirubinemia.

On exam, he was noted to have blink to light vision in both eyes. His intraocular pressures were normal. Pupils were equal in size and slowly reactive to light. There was no evidence of a relative afferent pupillary defect. On slit lamp examination, the anterior segment was normal with the exception of dense nuclear and cortical lenticular opacities in both eyes (Fig. 6.1). The pupils dilated poorly after installation of mydriatic drops. There was no view of the posterior pole secondary the lenticular opacities. B-scan ultrasonography was conducted and found to be normal, without evidence of retinal detachment, mass, or persistent fetal vasculature.

Extensive laboratory workup was conducted, and the patient was found to have Trisomy 21 on genetic testing. Additional workup was performed by the NICU team to rule out other comorbidities commonly seen in patients with Trisomy 21, but the remainder of systemic workup was normal. There is a known correlation between Trisomy 21 and congenital cataracts; thus, this was the presumed etiology for this patient's cataracts.

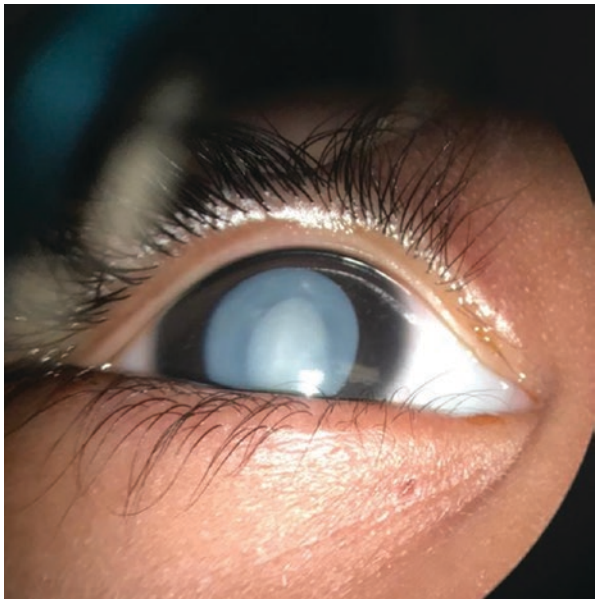

Fig. 6.1 Total lenticular opacity involving the nucleus and surrounding cortex of the lens. Also note the incompletely dilated pupil despite multiple rounds of mydriatic drops. A similar cataract was present in the fellow eye

The patient underwent immediate sequential bilateral cataract surgery at 6 weeks of age. The decision for immediate sequential surgery was made given the patient's age and risk of anesthesia-related adverse events. A complete examination under anesthesia and contact ultrasound biometry and automated keratometry measurements were done in the operating room. He was left aphakic, and refractive correction was planned with contact lenses.

Comment This case illustrates the importance of an infectious, genetic, and metabolic workup in infants with bilateral congenital cataracts and other concerning findings in their history and/or physical exam. As mentioned previously, in an otherwise healthy patient, a cause is not often found; thus, workup is not recommended [4]. It is imperative that any systemic condition that could lead to additional morbidity or mortality for the patient be detected and treated appropriately. It also illustrates the importance of discussing the timing of surgery, as well as the decision for immediate sequential cataract extraction in infants. The risk of anesthesia was felt to be greater due to this patient's Trisomy 21, and therefore, immediate sequential cataract extraction was performed. In an otherwise healthy baby, this same decision may not be pursued.

Case 2

A healthy 4-year-old girl presented to clinic as a referral for cataract evaluation. She did not have any medical conditions with known cataract associations nor did she have a history of trauma to either eye. She did not take any medications. Her first eye exam occurred approximately 1 month prior to this clinic visit. Her uncorrected Snellen visual acuity was 20/250 in the right eye and 20/30 in the left eye. Her pupillary examination was normal. Intraocular pressures, motility, and muscle balance testing were all normal, with the exception of reduced stereo acuity. Her slit lamp examination revealed a posterior subcapsular cataract in the superior temporal quadrant of the right eye. The cataract was mostly outside the visual axis when looking through the un-dilated pupil. After dilation, the opacity was noted to be largely occupying the superior temporal quadrant of the lens but again was mostly outside of the visual axis. The remainder of the slit lamp and fundoscopic exam was normal in both eyes. Cycloplegic retinoscopy revealed a refraction of +5.00 sphere in the right eye and +2.00 + 0.75 × 180 in the left eye. Because of significant anisometropia, the patient was prescribed glasses for full-time wear with the following prescription: +3.00 sphere in the right eye and plano+0.75 × 180 in the left eye. She was also instructed to start patching the left eye 3 h daily until her follow-up visit.

Three months later, the patient returned to clinic for follow-up. Her vision with correction was 20/150 and 20/30 in the right and left eyes, respectively. Mom reports she patched well for the first 2 months but had not been as compliant in the past few weeks. The remainder of her exam appeared stable; thus, the patient was instructed to continue full-time glasses wear, and the importance of patching was reinforced.

She returned several months later, and her vision with correction was 20/200 in the right eye and 20/30 in the left eye. Her mother admits that they have not been compliant with the glasses or patching. On slit lamp examination, the posterior subcapsular cataract appeared to be occupying more of the visual axis compared to her two previous examinations. The decision was made to proceed with cataract extraction with intraocular lens implantation. This patient underwent keratometry and contact ultrasound biometry in the operating room. Postoperative refraction of +2.00 was chosen in order to match her fellow eye and to allow for emmetropization with axial elongation.

Comment This case illustrates the importance of considering the etiology of the amblyopia. Just because a cataract is present, it is not always surgical. The leading reasons for amblyopia in this case include deprivation amblyopia from the cataract and refractive amblyopia from her uncorrected anisometropia. In this case, reinforcing the importance of a thoughtful and attentive preoperative exam, the cataract was not fully obstructing the visual axis, and there was a significant amount of anisometropia present. Treating her with refractive correction and patching first allowed the surgeon to be certain that deprivation amblyopia was causing a significant amount of her amblyopia. It is also important to note that the posterior subcapsular cataract progressed during this time. Sometimes, it is necessary to examine the patient several times in the clinic or perform an EUA prior to making a decision regarding surgery. This was evident not only on slit lamp exam, but the patient's vision also worsened compared to the previous visit.

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Chapter 7
Preoperative Counseling

Jana Bregman, Janet Alexander, and Moran Levin

Preoperative discussions with caregivers and patients are an essential component of the early phases of care related to childhood cataracts. These conversations are detailed and complex, requiring the physician to set aside appropriate time and participate as both an active educator and listener. The physician will also need to coordinate the interdisciplinary needs of each unique patient. Appropriate expectations should be set, terminology should be introduced, and empathy should be used, particularly in addressing the concerns and needs of each family. The postoperative course after childhood cataract surgery often lasts for decades, so the family must be prepared for long-term care and follow-up. The family should understand that the physician-family-patient relationship will endure both triumphs and challenges requiring short- and long-term support from the medical team. Successful outcomes often require years of individualized vision rehabilitation and lifelong monitoring for complications.

Etiology

Often, the first question families ask is, "Why did my child get a cataract?" Physicians and parents alike have an interest in the answer to this important question. In some situations, the answer is known at the time of diagnosis (e.g., if a child had a family history of childhood cataracts, a congenital infection, or trisomy 21). However, in many instances, there is either no identifiable cause or testing will be

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recommended to help arrive at a conclusive answer. In the early phases of the physician-family relationship, it is important to outline for the family the broad categories of cataract etiology including idiopathic, inherited, secondary to other intraocular pathology, and secondary to a systemic condition. At this point, the physician may offer the most likely explanation for the cataract based on the history and exam findings. In cases where additional lab work is warranted, we recommend describing the occurrence of cataracts as an "important clue" that may help the ophthalmologist identify an underlying systemic issue that would benefit from early treatment. Chapter 1 (Cataract Epidemiology and Genetics) provides a comprehensive discussion of the specific causative examples of congenital cataracts. Table 7.1 highlights the steps in preoperative assessment for cataract causality.

The etiology of congenital cataract can be narrowed according to key clinical features: laterality, family history, syndromic systemic features, exposure history, and associated ocular pathology. A decision tree illustrating how the clinical features and history aid in determining the cause of congenital cataracts is shown in Fig. 7.1. The family, pediatrician, and other subspecialists, such as geneticists, are key participants in this investigation preoperatively. Often, targeted testing at the time of surgery can be arranged by thoughtful collaboration with a multidisciplinary team.

Table 7.1 Preoperative assessment for cataract causality

Physical and ocular exam in clinic	Systemic testing	Interdisciplinary consults
History – family and patient history may reveal the cause of childhood cataract(s)	Blood tests – TORCH infection workup, complete blood count (CBC), chemistry panel (glucose, creatinine, blood urea nitrogen (BUN), phosphorus, calcium), ferritin, red blood cell galactokinase (GALK) activity, other galactosemia tests[a]	Pediatrician
		Geneticist
	Urine tests – reducing substances, amino acids, galactosemia enzymes	Genetic counsellor
Exam – information about laterality, corneal diameter, presence of persistent fetal vasculature or Peters anomaly, B-scan for possible tumor or retinal pathology, signs of trauma	Genetic tests – specific test based upon presumed diagnosis, such as genetic panel (for non-syndromic bilateral congenital cataract), karyotype (for highly recognized chromosomal syndromes), or chromosomal microarray (for bilateral cataracts with less specific syndromic features) [1, 2][b]	Anesthesiologist
Exam of parents, siblings – if positive may help refine a presumed genetic diagnosis or identify siblings with cataracts or other ocular anomalies		Other subspecialists as needed
Continued surveillance: At least once per year, patients with a history of idiopathic bilateral cataract should be re-evaluated for any change or update to the review of systems.		

[a]Classic galactosemia test is part of routine newborn screening
[b]Performed under the guidance of a trained geneticist and genetics counsellor

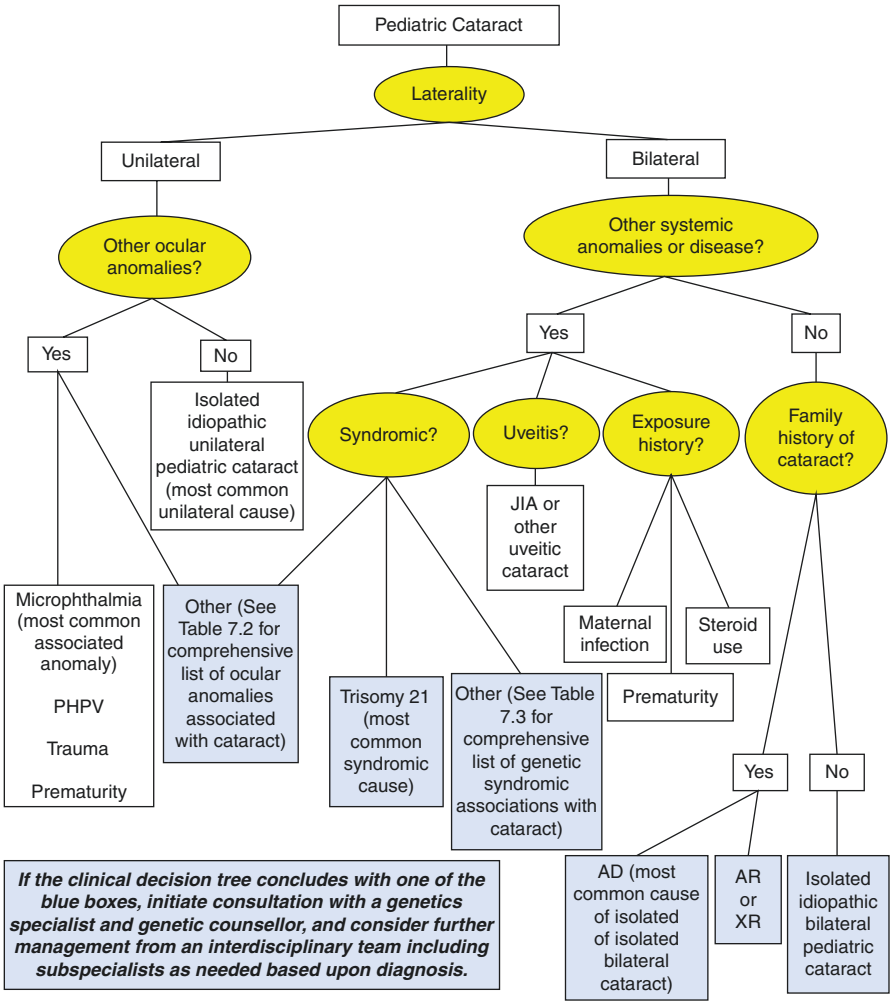

Fig. 7.1 Decision tree pediatric cataracts

Following a careful assessment of cataract causality with the assistance of genetics specialists, a patient-specific multidisciplinary team should be assembled as an essential part of appropriate care for a child with cataracts. Table 7.2 outlines cataract etiology according to the associated systemic manifestation with the greatest morbidity. This organization provides an initial framework for the early stages of developing a medical team that will help care for the child. Ophthalmologists should have a low threshold to refer to colleagues with expertise in managing these complex conditions, thereby ensuring that cataract management does not overshadow the child's global progress.

Table 7.2 Genetic syndromic associations with cataract, grouped according to primary systemic findings[a] [3, 4]

Trisomy	Metabolic	Craniofacial	Dermatologic
21 (Down) 13 (Patau) 14 (mosaic) 15q 18 (Edwards) 10q 20p	Galactosemia Hypoparathyroidism Pseudohypoparathyroidism Diabetes Refsum Hypoglycemia Mannosidosis Wilson's disease Sulfatase deficiency Fabry Glucose-6-phosphatase deficiency	Hallermann-Streiff Rubinstein-Taybi Smith-Lemli-Opitz Pierre Robin Oxycephaly Crouzon Apert	Cockayne Poikiloderma Atrophicans Incontinentia pigmenti Congenital ichthyosis Atopic dermatitis Ectodermal dysplasia Progeria

Central nervous system	Renal	Musculoskeletal	Gastrointestinal	Multisystem
Laurence-Moon Sjogren-Larsson Peroxisomal Zellweger Cerebral gigantism Batten disease	Lowe Alport WAGR Aniridia	Chondrodysplasia Myotonic dystrophy Albright osteodystrophy Potter Chondrodystrophic myotonia Spondylo-ocular syndrome	Cerebrotendinous xanthomatosis (CTX)	Nance-Horan Cockayne Marfan Turner NF2

[a]Note each disease grouped according to most relevant category; many diseases mentioned above have multi-organ involvement

Timing of Pediatric Cataract Surgery

Preoperative discussion regarding the timing of congenital cataract surgery should focus on weighing the risks and benefits of early versus delayed surgical intervention. The optimal age for pediatric cataract surgery is that which reduces the risk of deprivation amblyopia without significantly increasing the risk of secondary postoperative complications. Families should understand that in general, the younger the child is at the time of cataract surgery, the greater the incidence of associated complications. However, delaying surgery increases the risk and severity of amblyopia.

Retrospective and prospective surgical outcome data have helped define a latent period for pediatric cataract surgery beyond which the risk of amblyopia rises significantly [5–11]. There is consensus that surgery for unilateral congenital cataract should be performed when the child is 4–6 weeks old [10]. Beyond this latent period, visual outcomes and rates of amblyopia have been shown to steadily worsen over time [6, 7, 9, 10]. Surgery at an earlier age significantly increases the risk of secondary glaucoma, which carries severe ocular morbidity.

The surgical timing for children with bilateral cataracts is less well defined. Families should be counseled that poorer visual outcomes have been demonstrated in children who have surgery after 8–10 weeks of age [5, 10, 11]. Surgeries for the two eyes are ideally separated by approximately 1–2 weeks. A longer period

between surgeries increases the child's risk of developing amblyopia in the eye undergoing later surgery. Immediate sequential bilateral cataract surgery should be reserved for cases where general anesthesia exposure must be limited due to patient comorbidities [12]. In these cases, families must fully understand that this approach has the theoretical increased risk of bilateral endophthalmitis and is therefore not the preferred method for otherwise healthy children. In cases where a child is systemically unstable, surgery may need to be delayed until the child can safely undergo anesthesia.

Risks of Pediatric Cataract Surgery

Informed consent for pediatric cataract surgery should be discussed on multiple occasions prior to the procedure. Some families are referred to the ophthalmologist with a known diagnosis of cataract and the expectation of surgery, while for others, a cataract is a brand new diagnosis. In either scenario, all families will need a great deal of additional information to be adequately informed. Discussion of the potential benefits of cataract surgery is likely to be readily accepted and understood. However, for children who present at a later age, especially those with unilateral cataract, the risk of limited visual recovery and guarded visual prognosis should be explained in detail. Advantages of lens removal for such patients include enhanced contrast and color, improved peripheral vision, prevention of complications related to end-stage cataracts (phacomorphic glaucoma or phacolytic glaucoma), and improved view of the optic nerve and retina for both ocular health monitoring and cycloplegic refraction. For patients who present in a timely fashion, the potential benefits are even greater, but the risks of perioperative and postoperative complications for all children cannot be understated. This part of the discussion with families typically requires more time as it raises additional concerns and questions that need to be addressed. Table 7.3 outlines the many risks that the pediatric ophthalmologist must anticipate at the time of surgery and at every postoperative visit. While this list is too long to discuss each item in detail during the informed consent process, parents and families should be aware of and understand the very common risks of glaucoma, strabismus, refractive error, and amblyopia following pediatric cataract extraction. The rare but devastating risks of infection, retinal detachment, and anesthesia-related surgical risks must also be described in sufficient detail. Table 7.4 highlights the risk of significant complications related to pediatric cataract surgery.

Case 1

A 4-month-old boy presents to the emergency department for shaking eyes and failure to thrive. His ocular exam is notable for blink to light visual acuity, dense bilateral lamellar cataracts (Fig. 7.2), and an otherwise normal anterior segment.

Table 7.3 Operative risks related to pediatric cataract surgery

Immediate operative risks (intraoperative to week 1 following surgery)	Early risks (week 1 to year 1)	Late risks (after 1 year)
Anesthesia risk (postoperative apnea risk is greatest for patients below 45-week post-menstrual age) Discomfort (dryness and light sensitivity) Damage to nearby ocular structures (lids and lashes) Corneal abrasion Corneal scarring (expected at incision sites) Corneal clouding Wound leak Endothelial toxicity (toxic anterior segment syndrome or TASS) Subconjunctival hyphema or conjunctival injection Retained cortex Hyphema Iris prolapse Iris damage Iris sphincterotomy IOL malposition Vitreous prolapse Lens fragment in vitreous Ruptured posterior capsule Aggressive inflammatory response Infection	Amblyopia Strabismus Glaucoma Endothelial decompensation Iris heterochromia Refractive error Contact lens-related infection Membrane formation Cortical reproliferation Cystoid macular edema Iris synechiae formation Membrane formation or capsular scarring (phimosis or posterior capsular opacification)	Amblyopia Strabismus Glaucoma Retinal detachment Refractive error Potential need for secondary IOL or IOL exchange Contact lens-related infection

Fundus examination could be performed through a crescent of clear red reflex around the cataract with a widely dilated pupil and was normal.

Comment At this time, discussion with our patient's parents centered on the causation of his cataracts and the possibility that they represented an "important clue" into his failure to thrive. A genetics and comprehensive inpatient pediatric admission were recommended to assist in his workup. We discussed the timing and risks of surgery, as discussed above. Due to his presenting nystagmus, visual prognosis was felt to be more uncertain. Nonetheless, it was advised to proceed with surgery. Because of his overall good state of health, surgeries were performed 1 week apart.

Two weeks after presentation, the patient successfully underwent bilateral lensectomy and posterior capsulotomy with anterior vitrectomy. He was given aphakic contact lenses. There was resolution of the nystagmus, and he appeared to intermittently fix and follow with each eye. His eye pressure was 11 in each eye with iCare tonometry, fundus examination was normal, and cycloplegic refraction was +20.0 in each eye. At this point, his systemic workup was unrevealing, and he began gaining weight appropriately.

Table 7.4 Rates of significant complications associated with pediatric cataract surgery

Complication[a]	Risk	Notes
Death	<1–10 deaths per 10,000 anesthetics [13–16]	Studies cited are not specific to ophthalmic procedures, which may have lower risk given the short case duration and hemodynamic stability of eye surgery compared to other organ surgeries. Incidence of anesthesia-related pulmonary and cardiac arrests is highest in neonates [17, 18]
Cardiac arrest	~500 in 1 million anesthetics [19]	Studies cited are not specific to ophthalmic procedures [19–22]
Anaphylaxis	~20 in 160,000 anesthetics [20]	
Laryngospasm/bronchospasm	~5 in 100 anesthetics [21]	
Malignant hyperthermia	~1 in 100,000 anesthetics [22]	
Infection	0.71% [3]	Most commonly diagnosed 48–96 hours after surgery. Gram-positive species are the most common organisms [3]
Retinal detachment (RD)	2.5%, 5-year incidence [23]	Median time to RD 70 months, 5.5% cumulative risk at 10 years [23]
Corneal changes (requiring surgical intervention)	<1% [24]	According to the Infant Aphakia Treatment Study (IATS)
Glaucoma suspect	31% at age ~5 years [25]	
Glaucoma	17% at age ~5 years [25, 26]	Variable rates reported in the literature
Need for additional surgery related to cataract surgery	13% of aphakic patients, 63% of pseudophakic patients [27]	Percentage of patients requiring additional intraocular surgery in the first 12 months. Most of such surgeries are for the purpose of clearing the visual axis [27]
Secondary membrane or proliferation of lens material	80% in pseudophakes less than age 1 year (percentage is lower in older children or children without IOL) [28]	Most frequent long-term post-op complication which can be amblyogenic if visually significant. Risk increases for traumatic cataracts, young children [28–30]

[a]Listed in order of severity

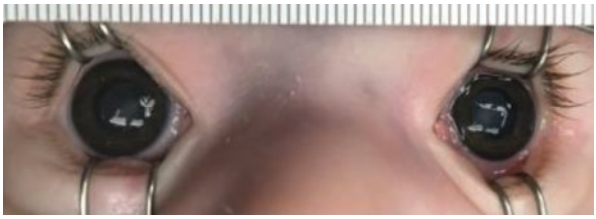

Fig. 7.2 External photo with ruler. Preoperative corneal assessment during examination under anesthesia demonstrates corneal diameters of 11 mm and 10 mm, respectively

Two years later, the patient was able to reliably read single Allen pictures in aphakic contact lenses, and his vision was 20/30 in each eye. Unfortunately, he had developed elevated eye pressures, enlarged corneal diameters, corneal clouding, and a myopic shift bilaterally but worse in the right eye, leading to the diagnosis of glaucoma following cataract surgery. He required surgical treatment for his right eye and topical antihypertensives in his left. He also underwent two strabismus surgeries for esotropia.

Comment The patient did well with improved early visual functioning and excellent acuity 2 years after his surgery. This case demonstrates how good visual outcomes can be achieved with prompt intervention, even if referral is delayed and the patient has signs of damage to the visual pathways (nystagmus) prior to lensectomy. Ongoing discussions with his parents continued to focus on the multiple nuances of recovery and care following cataract surgery.

We had decided on aphakic correction with contact lenses (see below for discussion of intraocular lens (IOL) implantation). The needs for long-term care included a discussion of the risks of amblyopia, strabismus, and glaucoma, among others. While patient age at the time of cataract surgery is a key risk factor for development of secondary glaucoma, delayed surgery (even a significant delay of 3 months) does not universally prevent secondary glaucoma. Due to excellent, consistent follow-up, we were able to diagnosis elevated eye pressure early, optimize our patient's outcome, and minimize risks of vision-threatening complications.

This patient's care was enhanced by consistent communication and collaboration with his pediatrician and our affiliated genetics team. Genetic testing identified a rare deletion previously reported in other individuals with congenital cataracts. This genetic finding offered a presumed genetic diagnosis but did not confirm the association with and etiology for his cataracts. Continued multidisciplinary involvement, including genetics subspecialists and genetic counselors, is an essential component of his ongoing ophthalmic care.

Primary Pseudophakia Versus Aphakia

The discussion of intraocular lens (IOL) implantation should review the risks and benefits specific to this unique surgical situation. Although IOLs have been found to be safe and effective in children, the Infant Aphakia Treatment Study (IATS) did not demonstrate a visual benefit to primary IOL implantation over primary aphakia

Table 7.5 Risks specific to intraocular lens (IOL) implantation

Postoperative complications	
Posterior capsule opacification Anterior capsular phimosis Secondary membrane Pupillary abnormalities	May require surgery to clear visual axis
Lens reproliferation causing IOL decentration Primary decentration IOL repositioning Anterior IOL malposition with or without iris capture exchange Refractive surprise Myopic shift with axial growth	May require or IOL
Inflammation Cystoid macular edema	May require medical management.

with contact lens placement in children with unilateral congenital cataracts. In the IATS, there was an increased rate of adverse events and higher rate of additional surgeries in the IOL compared with the aphakic group [31–33]. Disadvantages of primary IOL implantation that should be discussed include increased ocular inflammation, scarring, and the potential need for additional surgery including lens power changes [28, 32, 34]. Table 7.5 details specific risks related to IOL implantation in children.

It should be explained that the majority of the growth of the eye occurs during the first 2 years of life; however, the eye continues to grow and elongate throughout the second decade. This makes selecting an IOL power to minimize refractive error in young adulthood a challenge due to our limited accuracy in predicting the growth trajectory of each child's eye [34, 35]. Various refractive options should be discussed including aiming for a hyperopic correction to avoid a large amount of myopia later in life versus aiming for emmetropia with the use of a piggyback lens or refractive surgery when the axial length has stabilized [36]. Multifocal IOLs may be considered for older children [35, 37].

If the patient is to be placed in aphakic contact lenses, the management of contact lenses should be discussed in detail preoperatively. Active involvement and participation from the patient's caretaker are critical for success with contact lenses [38]. The caretaker will need to be comfortable with insertion, removal, and cleaning of contact lenses. Families should be counseled preoperatively that adherence to patching and contact lenses is essential for prevention of amblyopia. Disadvantages of contact lenses include the cost associated with frequent replacement due to mucous buildup and change in refractive power [39]. If the physician or family suspects that there are barriers to successful management of aphakic contact lenses that put the patient at risk for poor vision, the physician may in turn select IOL implantation or aphakic spectacles [40].

Long-Term Management

It is important to counsel families that regular ophthalmologic care will extend far beyond the time of the initial cataract surgery and the immediate postoperative period. Preparing families for long-term surveillance allows the ophthalmologist to follow closely for pathology associated with pediatric cataracts and cataract surgery. Amblyopia, secondary glaucoma, and strabismus are three of the most common long-term complications associated with pediatric cataracts. A complete and thorough discussion of these risks should take place. Families should understand that frequent follow-up appointments for patching, refraction, adjustment of spectacles and/or contact lenses, and monitoring of intraocular pressure are an important part of the long-term management and treatment of pediatric cataracts.

Amblyopia

Families should be aware that cataract extraction does not guarantee full visual recovery. Children with cataracts are at high risk for amblyopia [41]. Early cataract surgery reduces the risk of amblyopia; however, postoperative amblyopia therapy is equally as important. Families should be counseled that patching and optical correction, including glasses and aphakic contact lenses, will be needed, with frequent adjustment to account for refractive shifts as the eye grows. The IATS demonstrated that higher socioeconomic status, lower parental stress, and younger age of the child were associated with improved adherence to postoperative patching regimens [42]. These potential factors may help identify parents who require additional guidance in the postoperative period to optimize long-term patient outcomes. It is also important to utilize social support or additional services when needed to help families adhere fully to the prescribed visual rehabilitation regimen.

Glaucoma

Secondary glaucoma is a potential and serious complication that should be discussed with families prior to pediatric cataract surgery. Glaucoma occurs in 15–30% of cases and may be detected as early as 2 weeks or as late as 5 years following cataract surgery [29, 43–46]. It is often insidious in onset without the typical hallmark findings of primary congenital glaucoma (buphthalmos, epiphora, corneal clouding). Therefore, close intraocular pressure and optic nerve monitoring by the ophthalmologist are required for detection of glaucoma long after the initial surgery and at every clinical visit postoperatively [46]. Glaucoma after cataract surgery is more common in children with microphthalmia and microcornea and in those who

undergo cataract surgery at a younger age, although it may occur in children without these risk factors. Additional risk factors for developing glaucoma include persistent fetal vasculature, prolonged postoperative inflammation, and the need for multiple operations [46, 47]. As previously mentioned, families should be counseled that the most readily modifiable risk factor for developing secondary glaucoma following cataract extraction is the age at which the child undergoes cataract surgery. Delaying cataract surgery – especially unilateral cataract surgery – significantly increases the risk of amblyopia. Therefore, a degree of postoperative glaucoma risk is most often accepted by the surgeon in order to reduce the risk of amblyopia development.

Strabismus

The majority of children who undergo cataract surgery will develop strabismus, and it is often present at the time of cataract diagnosis [48]. In some instances, parents may have noticed their child's ocular misalignment prior to ophthalmologic evaluation. It is important to counsel parents that almost half of children with pediatric cataracts will require strabismus surgery following cataract extraction [49]. Families should be counseled that success with strabismus surgery in this population is lower compared to children without cataracts and the rate of reoperation may be higher. This is due to amblyopia disrupting binocular functioning required to help drive ocular alignment.

Long-Term Visual Prognosis

Parents will want to know what can be done to ensure that their child has the best possible outcome following cataract surgery. A host of factors impact a child's long-term visual prognosis including, but not limited to, the age of cataract onset and timing of cataract extraction, the density and laterality of the cataract (unilateral versus bilateral), associated ocular and systemic pathology, and the quality of postoperative visual rehabilitation. Children who are of kindergarten age or older when they develop cataracts, and infants who have early surgery for bilateral cataracts, have a superior visual prognosis compared to those with early-onset unilateral cataracts. Major studies such as the IATS can be referenced to explain that almost half the children in the latter group achieved vision better than 20/200 following cataract surgery and the majority acquired some depth perception [6–8]. Close surveillance, possible chronic intraocular pressure-lowering therapy, and, often, additional eye surgeries are often required. Therefore, long-term treatment and support must be offered well after the initial cataract surgery is completed in order to optimize and preserve the child's visual functioning.

Health Literacy and Informed Consent

It is important for physicians to gauge the family's health literacy in order to guide preoperative discussions and to target patient information materials and written consent forms appropriately. Families with limited education levels or literacy skills pose a challenge in preoperative counseling and obtaining informed consent for pediatric cataract surgery. Written and verbal discussions with families should take into account that 50% of the US population cannot read above an eighth-grade level. Even patients with the highest levels of education may have limited health literacy, hindering their ability to understand health information and make appropriate health-related decisions. The National Institutes of Health and American Medical Association recommend that patient educational resources be written at or below an eighth-grade reading level so that the information is accessible to most patients. While the risks and nuances of long-term management of pediatric cataracts may be complex, the surgeon should make every effort to use clear, concise, and simple language in counseling patients and their families [50, 51].

Appendix 1 contains a sample consent form that can be used during the preoperative discussion with families in planning for pediatric cataract surgery. This form is modified from the adult cataract consent form available on the Ophthalmic Mutual Insurance Company web page and is validated through readability indexes to be written at or below an eighth-grade level.

Case 2

An 11-month-old baby boy with trisomy 21 was referred by his pediatrician for dull red reflexes. He presented to the ophthalmology clinic with his parents, who live in a rural area 2 hours away. His mother and father are 17 years old, did not attend high school, and have a history of substance abuse. The patient was found to have visually significant cataracts in both eyes (Fig. 7.3). After thorough discussion with the family regarding the risks and benefits of surgery, the decision was made to pursue bilateral sequential lensectomy, without implantation of an intraocular lens.

Immediate postoperative appointments were uneventful, and the postsurgical course was without complications. Two weeks later, the patient failed to return for follow-up appointments. A pediatric clinical social worker was then recruited to assist in contacting social services, and transportation was provided for appointments. The patient's grandmother was noted to be an involved caretaker, and she was encouraged to attend all clinic appointments along with the parents. The family was educated on the importance of timely follow-up and the risks of vision loss. Given the complex social dynamic and inconsistent follow-up history, aphakic spectacles were prescribed instead of contact lenses.

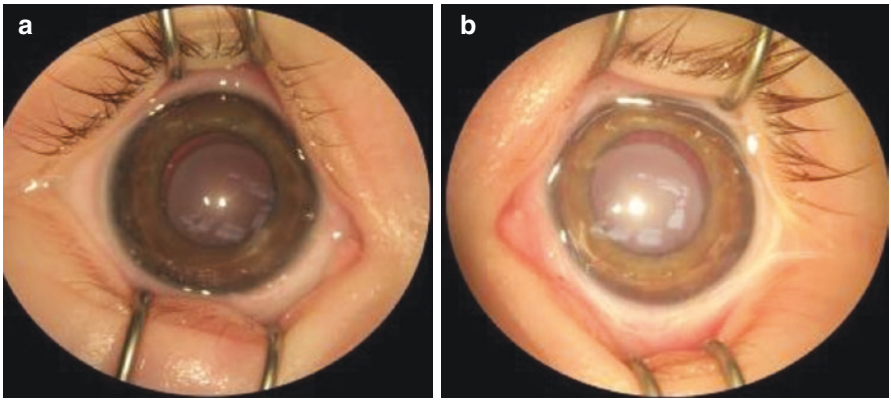

Fig. 7.3 Microscope view bilaterally. Preoperative lens assessment during examination under anesthesia demonstrates bilateral dense lamellar cataracts

After 3 years of follow-up, the patient was 4 years old and demonstrated very good vision. He continued to return for follow-up appointments with the assistance of social services. He is doing well with aphakic spectacles and may be a good candidate for secondary IOL implantation in the future.

Comment The surgeon should identify any potential barriers to postoperative management and timely follow-up prior to cataract surgery. This may include difficulty obtaining medications, contact lenses, glasses, or attending postoperative appointments. Early involvement of social support services should not be overlooked in its ability to assist in these vital components of post-cataract surgery rehabilitation. For situations where primary caregivers are unable to adequately absorb all duties of the postoperative care, other family members can be recruited. Understanding family dynamics and identifying risk factors can aid in the decision to use aphakic spectacles versus contact lenses or to implant a secondary intraocular lens.

Appendix 1: Pediatric Cataract Surgery Consent Form

What Is a Cataract?

The lens is the clear part of your eye that helps focus images. *A cataract* happens when the lens of the eye becomes cloudy. Cataracts can cause blurry vision or blindness in children. If a cataract is not removed early in life, a child may develop permanent vision loss. This is a serious condition, since vision is important for the brain to develop.

What Causes Cataracts?

Some children are born with cataracts. Other children develop cataracts from medications, infections, eye injuries, or medical diseases. Some cataracts are genetic (can run in families).

How Are Cataracts Treated?

Surgery is the only way to remove a cataract. During cataract surgery, an eye surgeon will remove the cloudy lens. The surgeon may replace the cloudy lens with an "IOL" (intraocular lens, a clear artificial lens). The surgeon may choose to leave your child without a lens (aphakia). In this case, the child may need glasses and/or contact lenses following surgery. In some cases, an IOL can be placed in the eye years after the cataract is removed.

What Are the Major Risks of Cataract Surgery?

Cataract surgery is usually safe and successful. There are risks (problems that can happen) with cataract surgery. While the eye surgeon cannot tell you about every risk, here are some of the common or serious risks:

- *Risks from cataract surgery* include vision loss, blindness, or negative results. Bleeding, damage to parts of the eye, infection, and inflammation can happen. The retina at the back of the eye can pull away from where it is attached (detached retina).
- Your child may need another surgery to take out pieces of the cataract that were not removed.
- *Glaucoma (high eye pressure)* can happen after cataract surgery. Children with glaucoma may need eye drops or glaucoma surgery. Glaucoma can happen whether or not your child has an IOL.
- *It is common for part of the lens to grow back.* This can form a lens membrane or "after cataract." Your child may need a laser procedure or another surgery to remove a lens membrane.
- *Most children will need glasses and/or a contact lens* after cataract surgery.
- *Lazy eye (amblyopia)* is common, and children may need eye patches or drops after cataract surgery.
- *Misaligned eyes (strabismus) and poor depth perception (stereopsis)* can happen, and children may need eye muscle surgery.
- Your child may need frequent eye surgery or exams that require anesthesia.
- *Anesthesia* can cause heart and breathing problems. Very rarely, it can cause death.

- *Other risks.* Cataract surgery only corrects vision problems caused by cataracts. This surgery cannot correct vision problems caused by problems in other parts of the eye. There is no guarantee that cataract surgery will improve your child's vision. It is possible that vision loss, blindness, or even the loss of an eye can occur. These problems can appear weeks, months, or even years after surgery.

Risks from an IOL

Adults with cataracts almost always have an IOL put in their eye. In children, this part of surgery is slightly more risky, so for children with cataracts, an IOL is optional. The good part of an IOL is that it decreases the need for strong glasses or contact lenses. The bad part of an IOL is the slightly increased risk of needing additional future surgery. Most children will be good candidates for putting a lens implant in at a later surgery, when it might be safer or more accurate. As your child's eye grows, the power of the eye changes. The IOL may be too weak or too strong. The eye surgeon may need to replace your child's IOL or make changes to glasses months or years after surgery. An IOL can cause inflammation, glaucoma, scarring, a lens membrane, or other surgery complications.

Acceptance of Risks

I understand that it is impossible for the doctor to inform me of every possible complication that may occur. By signing below, I agree that I have read this form or someone has read it to me, that my doctor has answered all of my questions, and that I understand and accept the risks, benefits, and alternatives of cataract surgery.

I consent for my child to have cataract surgery with an intraocular lens (IOL) in the _____ (state right eye, left eye, or both eyes).

_____ _____
Person authorized to sign for the patient Date

_____ _____
Surgeon Date

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Part II
Surgery

Chapter 8
Surgical Steps

Angeline Nguyen and Courtney L. Kraus

Many steps of cataract surgery must be approached differently in the pediatric population compared to the adult population. Anatomical features of younger eyes such as the elastic anterior capsule, soft nucleus, decreased scleral rigidity, and adherent vitreous base are just a few factors that necessitate a much altered approach to surgery from that in an adult. The propensity for inflammation, infection, visual axis opacification, and glaucoma in this age group also necessitates tackling certain precautions to minimize potential adverse outcomes. This chapter will provide tips for cataract surgeons to aid in the preparation for tackling the distinct technical aspects of managing the pediatric cataract.

Preoperative Steps

To achieve adequate dilation by the time the patient enters the operating room, the patient should undergo preoperative administration of three rounds of dilating drops while in the preoperative suite. We use a combination of a cycloplegic (cyclopentolate 1%) and a sympathomimetic agent (phenylephrine 2.5%) in our practice. Phenylephrine 10% should be avoided, as it has been associated with tachyarrhythmia and hypertension [1].

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Anesthesia

As with most other pediatric surgeries, pediatric cataract surgery is performed under general anesthesia. Due to the risk of systemic toxicity from excessive absorption of phenylephrine drops, IV access should be obtained promptly to allow for administration of vasodilators if necessary [2]. More modern inhalational anesthetics such as desflurane and sevoflurane have been found to be associated with a greater chance for emergence delirium [3, 4]. To reduce this risk, some have tried newer intravenous anesthetics, such as dexmedetomidine, which has anxiolytic, sedative, and analgesic properties. It should be noted that this agent would not be as ideal for strabismus cases, as it can worsen bradycardia from the oculocardiac reflex [5, 6]. For management of the airway, intubation with paralysis may be preferred, as supraglottic devices such as the laryngeal mask airway (LMA) may increase the risk of laryngospasm [7]. Adjunctive topical or subtenon anesthesia has been found to reduce postoperative pain and better satisfaction scores among parents [8, 9].

Equipment, Fluids, and Injectables

Prior to having the patient enter the room, all machinery should be confirmed to be in working order and properly primed. The decision should be made about what irrigation fluid to run through the machine. Balanced salt solution mixed with 0.5 mL of epinephrine 1:1000 to each 500 mL bottle of irrigating solute is preferred for maintenance of mydriasis intraoperatively. All instruments, injections, lens calculations, and the selected intraocular lens (IOL) should be readily available once the case is underway in order to avoid unnecessary delays while the patient is under anesthesia.

Surgical Site Sterilization and Draping

Proper sterilization of the eye surface is important for reducing the risk of infection. Due to lack of evidence that preoperative antibiotics decrease rate of endophthalmitis, this is not recommended as part of the sterilization procedure. Placement of a 5% povidone-iodine drop is recommended due to the higher level of evidence that it reduces conjunctival flora [10]. Following sterilization of the ocular surface with 5% povidone-iodine, the lashes and skin surrounding the eye should be sterilized with 10% povidone-iodine and allowed to fully dry prior to placing the drape. The eye should be draped in a manner that keeps the eyelashes away from the operative field as to reduce the risk of contamination.

Intraoperative Steps

1. *Stabilize the eye*: The position of the eye should be in a stable, primary position. If a patient is not paralyzed by anesthesia, the patient's eye may supraduct under light anesthesia due to Bell's phenomenon. One method for stabilization of the eye position is with the use of a traction suture placed underneath the superior rectus muscle with 4-0 silk. It has been suggested that a potential downside of this technique is violation of the superior conjunctiva, which may jeopardize the success of potential glaucoma filtering surgeries in that area [11, 12]. However, traction sutures are placed quite posterior to the location of an ideal bleb, and no study has focused on the impact of a traction suture on filtering surgeries. In our own experience, the presence of former traction suture has not hindered the success of subsequent trabeculotomies or glaucoma drainage device placement.
2. *Paracenteses*: The paracenteses (side ports) may be made in the clear cornea, limbus, or sclera. To allow for as much intact anterior conjunctiva as possible, the authors prefer clear cornea incisions. Positioning the incisions superotemporally and superonasally allows for them to be protected from trauma and environmental sources of infection by the upper eyelid. The paracenteses should ideally be equally sized and spaced at least 4 clock hours apart for better access to the lens capsule and cortex in subsequent steps. The incisions should be made just large enough to allow passage of instruments to guard against anterior chamber collapse, which is more common in pediatric eyes than in adult eyes. If 20-gauge instruments are to be used, then the paracenteses should be created with a 23-gauge microvitreoretinal (MVR) blade. Alternatively, a 15° or 1.0 Sideport blade can be used to create the paracenteses. The depth of entry of the 15° blade can be titrated to the preferred size. Since the phacoemulsification handpiece or similar such large instrument is not required in pediatric surgery, the incisions can be kept small until the point at which the intraocular lens (IOL) needs to be inserted. At this later point, one of the paracenteses can be enlarged, as discussed below.
3. *Trypan blue (optional)*: After construction of the paracenteses, the decision should be made about whether trypan blue is needed for improved visualization of the anterior capsule. Cases have been reported in adults of inadvertent posterior capsule or vitreous staining with trypan in cases of zonular weakness, which can lead to obscuration of the red reflex. Therefore, caution should be taken to not overfill with trypan in cases of suspected zonular weakness or after use of iris hooks [13] (Fig. 8.1).
4. *Ophthalmic viscosurgical device (OVD) injection*: High molecular weight (cohesive) OVD is preferred to low molecular weight (dispersive) OVD, as it can more effectively deepen the anterior chamber and flatten the anterior capsule during anterior capsulorrhexis formation, thus reducing the risk of an anterior capsule radial tear. High molecular weight OVD is therefore injected prior to the creation

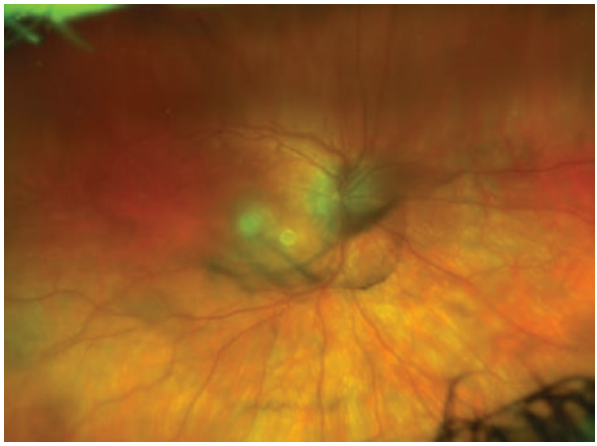

Fig. 8.1 Trypan blue staining of the vitreous on postoperative day 1 following lensectomy, IOL insertion, and anterior vitrectomy

of the anterior capsulorhexis, especially when a manual capsulorhexis is planned. The high-viscosity cohesive agent we use is Healon GV (AMO, USA).

5. *Anterior capsulorhexis*: The proper creation of the capsulorhexis is a critical step to ensuring a well-centered and stable IOL. In adults, a capsulorhexis that is continuous and curvilinear (continuous curvilinear capsulorhexis, CCC) is considered ideal for resisting radial tears. The same principles apply in children: the anterior capsulorhexis should be continuous, round, and centered in order to withstand the stress of IOL implantation and to allow for a well-positioned IOL:

   (a) *Manual capsulorhexis*: The performance of a manual capsulorhexis in children has unique challenges related to the elastic nature of the anterior capsule in young eyes, which increases the tendency for a radial tearing force. The technique for a capsulorhexis involves creating a small tear in the center of the anterior capsule and using microincision capsulorhexis forceps to grasp the leading edge of the flap and pull toward the center of the capsule. The leading edge should be picked up with frequent small grasps with a pulling force toward the center of the pupil to prevent a tear or enlargement of the anterior capsule. The start of the capsulorhexis should be smaller in diameter than the target diameter of the capsulorhexis, as the tendency is for the resulting capsulorhexis to be larger than originally intended and stretch. High molecular weight OVD can be injected frequently into the anterior chamber to flatten the anterior capsule and aid against forces that contribute to a radial tear of the anterior capsule.

   (b) *Vitrectorhexis*: While manual capsulorhexis is considered the gold standard for capsulorhexis formation due to its resistance to capsule tearing [14], the manual technique is highly challenging and requires microincision forceps, which are not always readily available. An alternative to manual capsulorhexis was described by Wilson et al. and involves the use of the vitrectomy cutter to create the anterior capsular opening, a technique referred to as

vitrectorhexis. One advantage of vitrectorhexis in pediatric eyes is that both anterior capsulorhexis and lens aspiration can be performed sequentially with the same instrument, thus minimizing the number of entries into the eye.

The vitrector should be ideally supported by a Venturi pump, as a peristaltic pump will not cut the anterior capsule as easily. The vitrectorhexis involves a bimanual technique through the paracenteses. The settings of the vitrector should be set to a low cutting rate (150–300 cuts per minute) and a high aspiration rate (300 mmHg). An initial tear in the anterior capsule is not required for the vitrectorhexis technique. Rather, the vitrector can be positioned with its cutting port facing posteriorly at the center of the anterior capsule. The vitrector is used to engage the anterior capsule to initiate a small opening in the center of the anterior capsule which can then be radially enlarged to the desired diameter of the capsulorhexis with care to maintain as rounded edges as possible. Even if the capsulorhexis is initiated manually, the vitrector can be used to rescue the capsulorhexis if a radial tear is made during manual formation of the capsulorhexis. Despite the relative ease and convenience of vitrectorhexis, we prefer a manual capsulorrhexis whenever possible, with the exception of in young infants (<7 month old). In these young patients in whom an IOL will not be placed, the concern for a radial tear is greater due to the greater elasticity of the anterior capsule. Also, since such infants will likely not receive an IOL, there will be less stress made on the capsulorrhexis due to IOL insertion.

(c) *New techniques for capsulorhexis*: An example of a newer manual technique for creating the CCC in pediatric cataract surgery is the two-incision push-pull technique (TIPP), described by Nischal [12]. In this technique, after the anterior chamber is inflated with OVD (without hyper-inflating), the anterior capsule is punctured at the superior and inferior ends of the CCC with either an MVR blade or cystotome. The superior flap is then pushed inferiorly, and the inferior flap is pulled superiorly with microincision forceps until the tearing edges meet each other centrally.

(d) *New devices for capsulorhexis*: Certain new devices may reduce zonular tension and decrease the risk of capsulotomy extension. Examples of devices include the Kloti diathermy unit (Oertli Instruments, Berneck, Switzerland), Diacapsutom (ERBE Elektromedizin GmbH, Tuebingen, Germany), PEAK-fc probe (pulsed electron avalanche knife; Carl Zeiss Meditec, Jena, Germany), and the Fugo blade (Medisurg Ltd., Norristown, Pennsylvania, USA), which have been suggested to be favorable for use in adult cases. Examples of assistive technology include femtosecond laser-assisted cataract surgery for creating the CCC in cooperative children under topical anesthesia [15].

6. *Avoid hydrodissection*: In adults, hydrodissection is generally performed in order to help with cortical cleanup. However, hydrodissection should be avoided in children, as it may cause rupture of the posterior capsule if there is a preexisting defect in the posterior capsule that is not visualized preoperatively.

7. *Lens aspiration*: Lens removal in the pediatric population rarely requires phacoemulsification due to the softer consistency of the pediatric lenses, which can be readily aspirated. As with adults, the lens can be removed by coaxial (through a single side port) or bimanual techniques (through separate irrigation and aspiration through two side ports). However, single port irrigation/aspiration (IA) can make subincisional cortical removal highly difficult and requires a larger incision. Therefore, a bimanual approach is preferred, which allows for better anterior chamber stability as well as more flexibility in switching the handpieces between the two side ports to access all quadrants. The bimanual technique can be performed either with separate irrigation and aspiration handpieces (Fig. 8.2a) or with a vitrectomy handpiece along with an anterior chamber maintainer, such as the Lewicky AC maintainer (Bausch & Lomb) (Fig. 8.2b). Regardless of which technique is used, it is important to aspirate as much OVD from the anterior chamber as possible prior to aspirating the lens in order to avoid impaction of the OVD onto the trabecular meshwork [12]. One advantage of an aspirator for lens removal is the smooth, rapid removal of lens material and fairly low risk of damage to the lens capsule.

When using the vitrector for lens aspiration, the same instrument can also be used to perform vitrectorhexis, irrigation/aspiration, posterior capsulorhexis, and anterior vitrectomy, thus minimizing having to switch instruments and repeatedly enter and exit the eye with instruments. The settings of the vitrector should be changed for the individual steps according to the manufacturer guide. The surgeon can elect to use an irrigator handpiece through the second incision or the anterior chamber maintainer. Since the Lewicky is a stationary device, the dominant hand can be used to hold the vitrector while the nondominant hand stabilizes the eye with forceps. In situations in which there are significant densities in the lens, the bimanual IA technique (Fig. 8.2a) offers an advantage over the Lewicky-vitrector technique (Fig. 8.2b), since the irrigation can be directly manipulated inside the eye such that both instruments can work together to mechanically break down the cataract between the two tips.

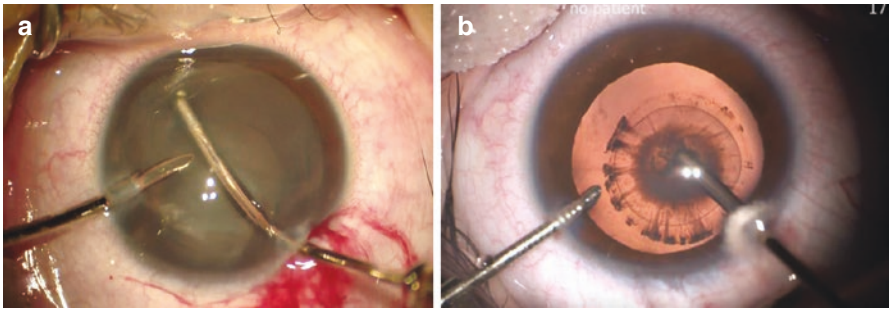

Fig. 8.2 (**a**) Bimanual technique for lens removal with a separate irrigation handpiece (left) and aspiration handpiece (right). (**b**) Bimanual technique for lens removal with a separate Lewicky anterior chamber maintainer (left) and vitrectomy handpiece (right)

Regardless of which bimanual technique is used, it is important to check that the irrigation cannula is patent by turning on infusion prior to entry into the anterior chamber. Having the irrigation cannula turned on during entry into the anterior chamber also prevents the OVD inside the anterior chamber from refluxing into the irrigation cannula and clogging its opening. Starting the irrigation rate at 30 generally allows for acceptable maintenance of chamber depth, but the rate may need to be increased depending on aspiration rate or chamber depth.

Our technique for lens aspiration using the vitrector or aspirator handpiece involves placing the handpiece under the edge of the capsulorhexis into the capsular fornix and increasing the aspiration until the cortex enters the port. For Venturi pump machines, complete occlusion of the tip is not needed to bring the cortex into the tip. Conversely, the peristaltic pump machines require that the surgeon occludes the aspiration tip to allow for an increase in the vacuum to the maximum level that is preset on the machine. Once the cortex is engaged, the tip is then moved slowly toward the center of the bag to detach the cortex from the capsule. We prefer to remove as much cortex from the periphery in as many quadrants as possible before removing the central nucleus in case there is a posterior capsular defect centrally. When only the subincisional cortex remains, it may be helpful to switch the handpiece to the opposite side port to achieve an optimized angle for removing the remaining cortex.

One of the key features to remember during pediatric cataract surgery is that complete removal of all cortical material is significantly more critical in children than in adults, as the cells that are left behind can be highly mitotically active. These cells have a high tendency to proliferate and cause visual axis opacification that may even require a subsequent operation to remove. To prevent this common complication, extra care should be taken to remove all visible lens material.

8. *Posterior capsulorhexis*: In young children undergoing cataract surgery, visual axis opacification is nearly guaranteed to occur if the posterior capsule and anterior vitreous are left intact. The anterior vitreous is believed to act as a scaffold for lens epithelial cells to migrate and form a secondary membrane. Therefore, to further prevent visual axis opacification, the center of the posterior capsule and the anterior vitreous should be removed at the time of cataract surgery in younger children. If posterior capsulorhexis is not performed at the time of cataract surgery, then a neodymium-yttrium-aluminum garnet (Nd:YAG) laser posterior capsulotomy could be performed in the clinic setting once posterior capsular opacification occurs, which happens usually 18 months to 2 years after initial surgery. If performed at the time of surgery, the posterior capsulorhexis should be made about 1 mm smaller in diameter than the anterior capsulorhexis in order to ensure centration and stabilization of an IOL, if placement of an IOL is planned.

   In determining whether to perform primary posterior capsulectomy versus Nd:YAG laser at a later date, the patient's age, cooperation of the child in sitting for laser, and availability of Nd:YAG laser are taken into consideration. The Nd:YAG laser is more challenging in children than in adults due to their level of

cooperation as well as the relatively greater thickness and robust formation of the posterior capsular opacification that occurs in children. Thus, higher amounts of energy are required for Nd:YAG in a child, and there may be a need to repeat the procedure due to subsequent closure of the posterior capsule opening. Khokhar and colleagues recommend performing primary posterior capsulotomy in patients up to 8 years old and anterior vitrectomy in patients up to 6 years old [16] to prevent visual axis opacification.

Similar to the anterior capsulorhexis, the posterior capsulectomy may be performed either manually or by vitrectomy. The manual technique confers similar advantages to that of the anterior CCC in terms of resistance against a capsular tear:

(a) *Manual posterior capsulorhexis*: The technique is similar to the anterior capsulorhexis in that a cystotome is used to puncture the posterior capsule and capsulorhexis forceps are used to grasp and pull the posterior capsule in a circular motion to create a capsulorhexis of the desired size. However, maximizing safety of the manual posterior capsulorhexis differs from the anterior capsulorhexis in that underfill (as opposed to overfill) of the eye with OVD is preferred in order prevent forces that would cause the posterior capsulorhexis to tear outward. Additionally, to protect the anterior vitreous face during capsulorhexis, OVD can be injected into Berger's space after the initial puncture of the posterior capsule with the cystotome [12].

(b) *Posterior vitrectorhexis (anterior vs posterior approach)*: Many surgeons choose to perform vitrectorhexis rather than the manual method for posterior capsulorhexis, especially if there is a plan to perform anterior vitrectomy. Settings for the posterior vitrectorhexis and subsequent anterior vitrectomy should be performed with the vitrector set to a high cut rate (>2500 cuts/minute) and low vacuum (less than 150 mm Hg). The posterior vitrectorhexis and anterior vitrectomy can be performed through the anterior approach (through the original limbal or clear corneal incision) or through a posterior approach (pars plana). Surgeons who are accustomed to performing anterior segment surgery often opt for the limbal approach due to greater familiarity with this approach. Also, the limbal approach spares the conjunctiva from manipulation and possible scarring. However, a disadvantage of the limbal approach is the potential for greater traction on the vitreous base compared to the pars plana approach. If the surgeon decides on performing the limbal approach, then the IOL insertion must occur after the capsulorhexis in order to allow space for the instruments to reach the posterior capsule through the anterior chamber without interference from the IOL:

    (i) *Limbal approach*: The vitrector should be placed with the cutter facing down in order to engage the posterior capsule centrally before moving outward in a circular motion. The posterior vitrectorhexis through the limbal approach must be performed prior to IOL insertion in order to gain access to cut the posterior capsule from the limbus.

(ii) *Pars plana approach*: In the pars plana approach, the IOL should be inserted first into the capsular bag. The OVD that was used to fill the bag for IOL insertion should then be removed prior to the posterior vitrectorhexis. In order to access the pars plana, an MVR blade is used to make an incision about 2–3 mm posterior to the limbus (2 mm for patients less than 1 year old, 2.5 mm for patients 1–4 years old, and 3 mm in patients over 4 years old) [17], while the anterior chamber maintainer or irrigation cannula remains in the anterior chamber. Creating the incision for the vitrector can either involve using a trocar or an MVR blade. The trocar or MVR blade is placed obliquely through the conjunctiva and sclera after the conjunctiva is displaced; the trajectory for insertion is as if aiming toward the optic nerve and then flattening to progress into the mid-anterior vitreous cavity. If using a trocar, the microcannula is left behind as the trocar is removed. The vitrector is then placed into the incision with cutter facing up in order to engage the posterior capsule.

9. *Anterior vitrectomy*: Once the posterior capsulorhexis is completed, removal of the anterior vitreous should again be performed with the vitrector set to a high cut rate and low vacuum. The anterior vitrectomy is performed immediately following the posterior capsulorrhexis without having to remove the instruments. In order to achieve adequate anterior vitrectomy, the surgeon should ensure that the tip of the vitrector is swept around the underside of the posterior capsule, reaching the entire circumference of the capsule with the cutter facing downward. Once the anterior vitrectomy is completed, it is important to remove the vitrector only after the irrigation has been turned off. Doing so ensures that the irrigation does not push hydrated vitreous out the same tract as the vitrector as it is withdrawn from the eye. If one is unsure that all vitreous strands are removed from the anterior chamber, intracameral triamcinolone (Triesence®) can be injected into the anterior chamber, staining vitreous strands that would otherwise be difficult to detect. If the sclerotomy sites show signs of leakage after removing the cannula, then the sclerotomy should be closed with a buried 8-0 vicryl suture.
10. *Main wound*: In preparation for IOL insertion, one of the paracenteses needs to be enlarged – most preferably the one on the side of the surgeon's dominant hand. Surgically induced astigmatism has been found to be similar between clear corneal and scleral incisions [18]. Enlargement of the paracentesis can be achieved with a microkeratome entering through the paracentesis in the plane of the iris. If a single-piece foldable IOL is to be inserted with a cartridge, the size of wound needs to range from 2.2 to 3.0 mm, depending on the IOL and cartridge design. The microkeratome used may either be sufficient for the wound size with a single pass, or additional enlargement on the sides of the wound may be necessary to allow for insertion of the cartridge being used.

11. *IOL insertion*: If one chooses the limbal approach for the posterior capsulorrhexis and anterior vitrectomy, then the IOL still needs to be inserted into the capsular bag. Once the vitrector and irrigation are safely withdrawn from the eye, high molecular weight OVD should be injected into the space between the anterior and posterior capsules in order to inflate the peripheral remains of the capsular bag. The OVD should be injected into multiple quadrants to separate the anterior from the posterior capsule and allow for ample room for the IOL to be inserted. A single-piece foldable hydrophobic IOL (SN60WF, Alcon Laboratories) is the most commonly used lens by the authors. After filling the cartridge for the injector system with high molecular weight OVD, the IOL is loaded into the cartridge. Once loaded, the cartridge passes through the main wound with the bevel down while the surgeon's other hand stabilizes the eye with forceps. The plunger is twisted to allow the leading haptics to be guided into the inflated peripheral capsular bag. It is important to keep the IOL as level with the plane of the capsular bag as possible as to not allow the IOL to be inadvertently injected through the posterior capsular opening. The IOL should be injected slowly and with a trajectory toward the anterior capsule and away from the posterior capsule as much as possible to achieve proper placement. Once the IOL has been completely injected, a Sinskey hook can be used to dial the trailing haptics into the bag if it is in the anterior chamber. Alternatively, the IOL can be injected into the sulcus, and it can then be dialed slowly into the capsular bag in a controlled manner using the Sinskey hook.

    For surgeons who prefer pars plan posterior vitrectorhexis, the IOL should be inserted first into the bag. The bag must again be filled fully with high molecular weight OVD prior to lens injection into the bag in order to not allow the IOL to cause a defect of the posterior capsule as it is being injected. The OVD can then be aspirated with the posterior capsule still intact and therefore without fear of engaging the vitreous:

    (a) *Optic capture*: As discussed in Chap. 13: Primary Lens Placement, posterior optic capture has been found to reduce opacification of the visual axis and lens decentration in children, although this may not necessarily result in improved best-corrected visual acuity (VA) [19]. To perform optic capture, the IOL is implanted into the capsular bag as described above. The edge of the optic is then slid behind the posterior capsulorhexis using a Leister hook circumferentially [20]. A change from round- to elliptical-shaped posterior capsule opening indicates successful capture. It should be kept in mind that an appropriate posterior capsule for optic capture is one that is continuous (therefore, it was ideally created manually), and it should also be well-centered and optimally sized (1 mm smaller than the IOL optic). Alternative means of optic capture include (1) capturing the IOL through both the anterior and posterior cap-

sulorhexis with the haptics in the ciliary sulcus and (2) reverse optic capture with the haptics in the bag and optic in the sulcus. The former is performed when the anterior capsulorhexis is compromised and is therefore inadequate for placement of the haptics in the capsular bag. The latter is useful when a tear exists in the posterior capsulorhexis, which makes the bag inadequate for placement of the IOL within the capsular bag.

(b) Alternatives to in-the-bag IOL: In-the-bag IOL insertion is preferred to other locations of IOL fixation due to decreased risk of glaucoma, damage to endothelial cells, and risk of cystoid macular edema. However, there are times in which in-the-bag insertion is not possible, and the lens must be placed in the sulcus (when there is sufficient capsular rim), or the IOL must be fixated to the iris or sclera.

12. *OVD removal and wound closure*: OVD can be removed using bimanular IA handpieces or a single-port technique with the double-barrel Simcoe cannula. Prior to introduction of either of these devices, the anterior chamber should be stabilized as much as possible with closure of the paracentesis and wound. With the OVD still in the eye, the main wound should be closed with three interrupted 10-0 vicryl sutures or a "figure 8" cross-stitch. The final interrupted suture in the wound is left untied until after all of the OVD has been removed. If the single-port technique with the Simcoe is to be used, then the paracentesis should be closed with a single interrupted 10-0 vicryl suture as well. Through the open part of the main wound, the Simcoe cannula is introduced with low aspiration (rate of 20) for removal of OVD from the bag with care as to not flip or destabilize the IOL. After a satisfactory amount of OVD has been removed, the third interrupted suture in the wound is finally tied, and knots can be buried.
13. *Injectables*: In order to limit the incidence of endophthalmitis following cataract surgery, some surgeons have adopted the use of intracameral perioperative antibiotics, either as an additive to the irrigating solution throughout the case or as an injection at the end of the case. Use of intracameral antibiotics is preferred by the authors, as there has been substantial evidence supporting its effectiveness in preventing endophthalmitis in adult cataract surgeries [21]. The use of intracameral moxifloxacin is convenient due to its availability as a preservative-free eye-drop solution (Vigamox®), which can be readily diluted 1:1 with balanced salt solution for injection of 250 μg in 0.01 mL solution [22]. Other commonly injected intracameral antibiotics include cefuroxime and vancomycin.

    To prevent postoperative inflammation, intracameral steroid (Triesence®) or subconjunctival steroid (dexamethasone) should be delivered prior to the end of the case.

Postoperative Steps

Following the conclusion of a case, some surgeons opt to place an additional drop of 5% povidone-iodine onto the surface eye for endophthalmitis prophylaxis. Finally, an antibiotic and steroid ointment is placed into the eye before patching the eye until the following day.

Case 1

A 13-year-old male patient developed a hyphema and, eventually, a traumatic cataract of the right eye following blunt injury from a rock 1.5 months prior to presentation. Presenting visual acuity (VA) to our service was 20/200, and examination revealed traumatic mydriasis due to multiple sphincter tears and a dense stellate anterior subcapsular and posterior subcapsular cataract. B-scan and fundus examination revealed no pathology. The decision was made to proceed with cataract extraction and placement of an IOL as well as pupilloplasty. The surgical approach was as described above. Trypan blue was injected into the anterior chamber, which led to appropriate staining of the anterior capsule but also obscuration of the red reflex due to presumed posterior migration of trypan blue. Despite difficulty with the view, the cataractous lens was removed and an IOL placed safely in the bag. Pupilloplasty was then successfully performed. Figure 8.1 depicts an Optos photograph of the patient's fundus 1 day after surgery, with trypan blue still staining the vitreous following lensectomy, IOL insertion, and anterior vitrectomy. The trypan staining resolved by postoperative week 1.

Comment This case serves to caution the reader about the potential hazard of the use of trypan blue in cases of weak zonules as discussed above. Situations of which to be aware of this possible complication include cataracts following trauma or prior intraocular surgery. Slow and deliberate injection can prevent overfill of the chamber and avoid posterior migration of the stain.

Case 2

A 4-year-old boy presented after a failed school vision screen. His family history was notable for a younger brother who had undergone bilateral cataract surgery at ages 4 and 6 weeks. On exam, vision was 20/50 uncorrected in the right eye and 20/70 uncorrected in the left. Slit lamp exam revealed mild lamellar lens opacities,

the left greater than the right. Cycloplegic refraction was −1.50 diopters (D) in the right eye and −2.25 D in the left eye. Dilated exam was within normal limits. The child was prescribed glasses and returned for follow-up 3 months later. VA at that time of follow-up was 20/25 OD and 20/30 OS. Several months later, he returned with complaints of decreased vision and photosensitivity, and VA was 20/40 and 20/70 after an updated cycloplegic refraction. The decision was made to proceed with cataract extraction and IOL implantation of the left eye.

The surgical approach in this case was similar to that described above. Following instillation of trypan blue, the anterior chamber was inflated with OVD. Using a cystotome needle, the anterior capsule was pierced, and a small flap was created. The microincision forceps were able to be passed through a 1.0 mm paracentesis, and a continuous curvilinear capsulorrhexis was created. The lens was aspirated, and a single-piece SA60AT 15.5 D lens, target plano, was placed in the bag. A posterior capsular opening was created through a pars plana approach.

Comment Manual creation of the capsulorhexis is technically more challenging but, when executed well, is more resistant to radial tears. It should be considered in older children, where larger palpebral fissures, improved control, and less elasticity of the capsule support this technique. Use of microincision forceps (as opposed to Utrata forceps) is essential to allow for maneuverability through a small paracentesis while maintaining chamber stability.

Case 3

A 4-year-old boy with history of posterior lentiglobus of the left eye diagnosed 2 years prior presented for follow-up with progressively worsening vision of his affected eye. He had not undergone surgery previously due to parental concern about general anesthesia, but had close follow-up since diagnosis. His parents reported excellent compliance with patching of the right eye 5 hours a day, 7 days a week. On exam, his vision was 20/25 in the right eye and 20/125 in the left eye. Slit lamp exam of the left eye revealed a spherical protrusion of the posterior lens surface with a speckled opacity in the region of the outpouched lens. Due to continued vision decline, his parents agreed to cataract extraction with intraocular lens implantation of the left eye.

The surgical approach included the creation of a manual CCC followed by lens aspiration through the clear cornea. Due to concern for the presence of a preexisting defect in the posterior capsule, which is common in posterior lentiglobus, the lens material was first removed from the periphery and then in the central area of the lenticular outpouching. After complete lens aspiration, there was a suspicious area that appeared to be an opening in the posterior capsule. As such, the vitrector

was kept inside the eye from the limbal incision to initiate creation of a round posterior vitrectorhexis from the edges of the posterior capsule defect and to perform an anterior vitrectomy. An SN60WF 21.0 diopter lens was carefully placed in the bag.

Comment This case illustrates the use of a limbal approach to the creation of a posterior vitrectorhexis. This technique reduced the need to remove instruments from the eye in order to create a posterior capsulorrhexis from the pars plana approach, as there was already a defect in the posterior capsule, and the edges of that defect could simply be used as the start of a posterior vitrectorhexis from the limbal approach. Delaying the creation of the posterior capsulorrhexis in order to switch to the pars plana approach might allow time for anterior vitreous to migrate anteriorly and thus cause traction on the posterior vitreous in a scenario where a posterior capsular defect is already noted at the time of lens aspiration.

Case 4

A 4-year-old boy with history of unoperated persistence of fetal vasculature (PFV) of the left eye presented with a dense white cataract. He had been previously evaluated, but had not undergone cataract surgery. Parents report occasional attempts at patching, with glasses worn full time for protection. Vision was 4/600. This was a significant decline from visual acuity obtained 1 year prior. Dilated exam revealed a dense white cataract with no view of the posterior pole. B-scan confirmed a thin, avascular stalk, with no clear traction on the retina. Decision was made to operate. Extensive discussion of risks and potential limited visual acuity improvement was undertaken.

Approach to the lens was through a limbal clear corneal incision. The lens was easily aspirated using bimanual irrigation and aspiration handpieces. The main incision is enlarged, and an SA60AT 26.5 lens was placed in the bag. The eye was then infraducted by placing traction on the superior rectus stay suture, and a small 2 clock hour peritomy was created from 1:00 to 11:00. Cautery was applied. Three mm from the limbus, a 23G MVR blade was used to enter the pars plana. The 25G vitrector was introduced. The avascular stalk was severed at its attachment to the posterior lens and removed to the midvitreous. This was accomplished without trauma or bleeding and followed by a limited anterior vitrectomy. Then, the vitrector port was turned skyward, and the posterior capsule was engaged. A posterior capsule opening was created 1 mm smaller than the anterior capsule opening.

On follow-up 1 month after surgery, the child had a clear visual axis, well-centered IOL, and a best-corrected visual acuity that had improved to 20/400. Parents reported they had begun to have more success with patching.

Comment This case illustrates the use of a pars plana approach to create a posterior vitrectorhexis. This technique allowed for creation of a posterior capsular opening and removal of stalk in a controlled fashion. By approaching from the pars

plana, the risk of anterior vitreous traction was minimized. Approaching the stalk from the pars plana and avoiding having to initially open the posterior capsule as with a limbal approach decreased likelihood of tension on the capsule and allowed uncomplicated removal of the stalk without tearing the posterior capsule.

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Chapter 9
Calculation of Intraocular Lens Power

Stacey J. Kruger

The selection of intraocular lens (IOL) power in children is decidedly different and more complex than when making similar choice in adult patients. The choice of IOL power is driven by many factors such as the age of the patient at surgery, the presence of unilateral or bilateral cataracts, the refractive error of the fellow eye, the physical properties of the eye including corneal size and axial length, and the presence and density of preexisting amblyopia. It is also important to consider the growing/changing eye in childhood and how these changes will translate into challenges managing resultant amblyopia.

Biometry and Keratometry

Technically, IOL calculation is performed in infants and most children during an eye exam under anesthesia (EUA) due to the limited cooperation one usually encounters in the office setting, rendering these tests unfeasible without sedation. The instrumentation that is used therefore needs to be portable for ease of manipulation and transportation to the operating room (OR). In this setting, the results of biometry are often less accurate as the asleep child cannot voluntarily fixate their gaze with the axis of measurement. It is often difficult and time-consuming to obtain keratometry readings, but it can be worthwhile to obtain three or more measurements per eye. The surgeon should select several readings that appear mathematically similar to the average of collected list of numbers. The accuracy of axial length determination is critical as even small errors in measurements can lead to large discrepancies in the postoperative refractive state as they are magnified by the

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various IOL calculation formulas. For determination of axial length, immersion A-scan measurements are preferred as they limit compression errors induced by contact A-scan.

IOL Formulas

There are numerous formulae for IOL calculation in use today, which are based on use in adult eyes. There has been significant research seeking to validate the use of one formula over another in the pediatric population. One such paper by Vanderveen et al. evaluated Hoffer Q, Holladay 1, Holladay 2, Sanders-Retzlaff-Kraff (SRK), and Sanders-Retzlaff-Kraff theoretic (SRK/T) formulae in infants that received IOL implantation at age 7 months or younger in the pseudophakic arm of the Infant Aphakia Treatment Study [1]. In this report, 43 eyes were studied with a mean axial length of 18.1 ± 1.1 mm. This study found that the Holladay 1 formula showed the lowest median absolute prediction error, while a paired comparison of medians showed similar results between Holladay 1 and SRK/T. The study is most applicable to infants undergoing primary IOL implantation, as the mean age of the study group was 2.5 ± 1.5 months. Another study by Trivedi et al. evaluated 45 eyes of children who underwent IOL implantation at a mean age of 3.56 years [2]. In this study there was a low mean absolute error of 0.68–0.84 D with the Holladay 2 formula giving slightly better predictions.

Axial Elongation and Myopic Shift

Another parameter that makes selecting an IOL for a pediatric patient challenging is the anticipation that the eye will grow during the child's lifetime. The human eye usually undergoes 3–4 mm of axial elongation in the first year of life, as well as corneal flattening and a reduction in lens power. Axial elongation was also studied in the Infant Aphakia Treatment Study. Axial length was measured before cataract surgery and again at ages 12 months and 5 years [3, 4]. In the first year, the rate of axial elongation was found to be nearly constant at a rate of 0.17 mm/month in the aphakic arm ($n = 57$), while in the pseudophakic arm ($n = 57$), the rate of elongation was found to be 0.24 mm/month [3]. In both groups, this rate was independent of age at surgery. In contrast, the rate of growth of normal fellow eyes decreased with older age at surgery. It is important to note that eyes with cataracts were shorter than fellow eyes at the time of surgery. Patients with glaucoma or suspected glaucoma were excluded, as this condition in infants is known to cause axial elongation. The same groups were then again reanalyzed at 5 years. Axial length was significantly different between treated and fellow eyes preoperatively (18.1 vs. 18.7 mm, $P < 0.0001$) and at 5 years follow-up (21.5 vs. 22.1 mm, $P = 0.0004$) [4]. The difference in axial length growth between treated and fellow eyes was not significant.

The change in axial length between the two arms (CL and IOL) was not significant between treatments. It is therefore important to remember when selecting an IOL for a patient with a monocular cataract that, although the rates of growth may be similar, a preoperative difference in axial length may persist throughout childhood.

Another important consideration is that as a pseudophakic eye elongates, myopia becomes magnified due to the optics of the IOL. As the eye grows, the focal point of the IOL moves forward, and as a result of the increased distance between the lens and the retina, the eye grows more myopic.

Secondary IOL

In children undergoing secondary IOL placement, special consideration should also be given to the calculation of lens power. Although most children undergoing this procedure are older, measurements are frequently still taken in the OR on the day of surgery with the patient under general anesthesia as described earlier. Moore et al. reviewed 50 consecutive eyes undergoing secondary IOL implantation at a single institution [5]. IOL calculations were made assuming "in the bag" positioning and then reduced by 0.5 D if placement in the ciliary sulcus was required. Despite the uniformity of EUA and IOL calculation procedures, patients still showed variability in predicted versus actual postoperative outcomes. In this study, the mean patient age at surgery was 6.5 years (range 0.6–15.0). The predicted postoperative refraction was +1.69 ± 1.85 D, whereas the actual postoperative refraction was +1.23 ± 1.25 D with a mean absolute value of prediction error of 1.64 ± 1.58 D. This resulted in a difference of 1.5 D in actual versus predicted postoperative refraction.

Lastly, a decision for placement "in the bag" versus the ciliary sulcus should be considered when placing a secondary IOL. This will likely be based on the status of the capsular bag. If the edge of the anterior capsule is not well visualized for 360° or the size of the anterior and posterior capsulotomies at the time of the original surgery were large, sulcus placement is recommended. A large retrospective review of secondary in the bag lens implantation was performed by Wilson et al. in which 10 years of data at a single institution were analyzed [6]. Patients receiving sulcus placed secondary IOLs during the same time frame were also analyzed. The mean pre- and postoperative spherical equivalents were not statistically significant when analyzed in patients with at least 6 months of postoperative follow-up.

Case Report

A 3-year-old girl presented to the office with a family history of bilateral congenital cataracts in her older brother, now age 8 years. She had been seen elsewhere 1 year prior and found to have high myopia for which she was prescribed glasses that she never wore. At the time of the exam, the patient's vision was 20/30 OD and 20/40

OS tested using Allen pictures. The patient was found to have bilateral lamellar cataracts that were clear in appearance. There was a good view of both fundi, which appeared normal. Refraction was found to be +1.50 + 0.50 × 90 OD and +1.50 + 1.00 × 90 OS. It was decided to follow the patient closely for changes in her vision and/or refractive state. The patient's exam was the same 3 months later, but after another 3 months, her vision had decreased to 20/80 and 20/150 in the right and left eyes, respectively. The appearance of the lenses had changed, the lamella now significantly opacified OS>OD. An EUA was performed, and the cataract was removed from the left, poorer seeing eye, first. A decision to place an IOL with the family had been made. The patient's K's were 44.00 and 46.50 in the right eye and 43.50 and 46.00 in the left eye. The axial lengths measured 21.02 mm and 20.41 mm in the right and left eyes, respectively. IOL calculations were made using the Holladay I formula since the patient's measurements were fairly average. A +27 D lens was placed in the bag during surgery. The patient's postoperative refraction approximately 2 months later was +1.50 + 1.50 × 105.

Comment In this case, the patient was intentionally left with a postoperative target refraction of approximately +3.00 D. Placing a +30 D lens would have left the patient +1.15 D, somewhat closer to emmetropia and with a relatively symmetric refraction compared with the fellow eye. However, the second eye surgery had already been planned for a future date at the time of the first surgery, and it was therefore felt it would be easy to match the +3.00 target refraction in the fellow eye. As discussed earlier, higher-powered IOLs, especially those over +30 D, can magnify the myopic shift as the patient grows. The parents were counseled regarding the myopic shift and were fortunately very knowledgeable about this since their older son, who had surgery early in life, now wore glasses to correct his moderate myopia.

The patient's postoperative actual refraction was somewhat less than targeted. It is possible that the "surgeon factor" was off in this case, as this was the first case by this author with a new contact A-scan device versus an immersion A-scan ultrasound that was used at a prior institution. The postoperative cylinder is felt related to the persistence of an absorbable suture remaining at the wound and may flatten over time.

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Chapter 10
Postoperative Care

Laura S. Kueny and Heather C. de Beaufort

Postoperative Medication Regimen

Much of the postoperative management in the setting of pediatric cataract surgery is extrapolated from the practice patterns of cataract surgery in adults. The American Academy of Ophthalmology Preferred Practice Guidelines do not specify a particular regimen for postoperative topical agents, as this varies by surgeon and there are no controlled trials establishing guidelines for postoperative topical agents [1]. Most adult patients are treated with a topical antibiotic, corticosteroid, and NSAID following cataract surgery. The postoperative eyedrop regimen used in the Infant Aphakia Treatment Study consisted of topical prednisolone acetate 1% 4 times per day for 1 month, a topical antibiotic 3–4 times per day for 1 week, and atropine 2 times per day for 2–4 weeks following surgery [2]. When an intraocular lens (IOL) is placed after pediatric cataract surgery, atropine and other cycloplegics are not generally advised due to the risk of IOL pupillary capture, especially with sulcus lens placement. In addition, topical NSAIDs are typically not necessary due to the lower risk of cystoid macular edema (CME) in pediatric patients and the lower incidence of comorbid risk factors such as diabetes. Medication side effects such as intraocular hypertension with corticosteroid use and allergic reactions to topical antibiotic use should be discussed with the patient or patient's family prior to use [1].

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Postoperative Complications

Endophthalmitis

Endophthalmitis is a severe and devastating complication following cataract surgery. This complication typically presents 3–5 days after surgery and presents as blurry vision, eye pain, and light sensitivity. In pediatric patients, the most common presenting symptoms are photophobia (50%) and pain (40.9%), and the most common clinical signs are conjunctival injection (36.4%) and hypopyon (31.8%) of the operated eye [3]. Other symptoms reported include eyelid swelling, fever, and lethargy. The pediatric patient may not be able to verbalize eye pain or a change in vision, and conjunctival injection and other clinical signs may not be immediately apparent; therefore close vigilance postoperatively is essential.

In 2007 the European Society of Cataract and Refractive Surgeons published a large prospective study that demonstrated a 4.92-fold increase in postoperative endophthalmitis in the absence of prophylactic intracameral antibiotics [4]. However, the use of intracameral antibiotics does not completely prevent endophthalmitis. In one series of pediatric postoperative endophthalmitis cases, 68.2% had received prophylactic intracameral antibiotics, and therefore, the use of postoperative topical antibiotics to prevent endophthalmitis and close postoperative monitoring to identify infection are necessary [3].

The management of endophthalmitis in the acute setting relies on early diagnosis and prompt treatment to prevent permanent vision loss. To ensure proper diagnosis, the operative surgeon or practice must be easy to contact, the patient or patient's family must be warned of the postoperative signs of infection, and the family must be instructed to contact immediately at the first signs of clinical worsening. The physician must then respond early with the correct diagnosis, which in the pediatric population may require an examination under anesthesia for definitive diagnosis. The physician must then treat immediately once the diagnosis is suspected.

Treatment consists of obtaining a specimen for gram stain and culture via vitreous or anterior chamber tap, with a 25- or 23-gauge needle, or 30-gauge needle, respectively. Intravitreal antibiotics, prepared by the pharmacy, are then injected. These include vancomycin (1 mg/0.1 cc) for gram-positive coverage and ceftazidime (2.25 mg/0.1 cc) for gram-negative coverage. The concurrent use of systemic antibiotics does not improve visual outcomes [5]. In adult cases of severe endophthalmitis, vitrectomy in addition to intravitreal antibiotics is the standard of care, though its use in pediatrics is variable. There is some evidence that vitrectomy in pediatric endophthalmitis also improves visual outcomes [6].

Case 1

A 15-month-old female presented with a progressive anterior pyramidal cataract of the left eye who underwent uncomplicated cataract extraction and posterior chamber intraocular lens placement followed by pars plana anterior vitrectomy and

posterior vitrectorrhexis. She received prophylactic intracameral moxifloxacin after implantation of the lens, and she was treated postoperatively with moxifloxacin and prednisolone 4 times daily and tobramycin/dexamethasone ointment at bedtime.

The patient presented for urgent follow-up on postoperative day 4 for redness and swelling of the left eyelid that had acutely worsened over the 24 hours prior to presentation. She had been compliant with her postoperative eyedrop regimen. On presentation, she was light sensitive; however, she was cooperative with the examination and did not appear to be in significant pain.

On examination, the vision in her left eye was difficult to elicit due to photophobia. Her left upper eyelid demonstrated 2+ edema with mild erythema, and her left eye had 2–3+ diffusely injected conjunctiva with horizontal fibrinous/plasmoid strands in the anterior chamber tracking to the paracentesis ports with no hypopyon.

Due to a concern for endophthalmitis, a retina specialist was consulted, and she was immediately taken to the operating room and examined under anesthesia. Endophthalmitis was suspected based on her exam findings, and an intravitreal tap was attempted 3 mm posterior to the limbus; due to her formed vitreous, no fluid could be aspirated. An anterior chamber tap was performed, and fluid was sent for bacterial and fungal culture. 2.25 mg of ceftazidime and 1.0 mg of vancomycin was then injected into the vitreous cavity of the left eye.

Postoperatively, she was placed on moxifloxacin and prednisolone drops every 2 hours while awake followed by atropine ointment and tobradex ointment at bedtime in the left eye. Her anterior chamber fluid was tested for gram stain and cultured for bacteria and fungi, all of which returned negative. She responded well to the intravitreal antibiotics and her clinical picture significantly improved over the following week. She is now doing well with preserved vision and no permanent structural damage to her left eye.

Comment In this case of suspected postoperative infectious endophthalmitis immediate diagnosis and treatment resulted in preserved vision in the operated eye. Often the presenting signs can be subtle and overlap with typical healing. This case demonstrates the importance of early identification and treatment of endophthalmitis in the pediatric population. The parents' early observation of eyelid swelling followed by immediate evaluation by her surgeon and treatment by the retina specialist resulted in preservation of her vision. In addition, while always recommended, cultures may not be diagnostic.

Intraocular Hypertension

Intraocular pressure (IOP) elevation following cataract surgery can occur acutely due to retained viscoelastic or within several days to weeks from steroid response due to the use of topical, intracameral, or intravitreal corticosteroids. In the pediatric population, this steroid response often occurs earlier and to a greater degree and frequency than in adults [7]. The use of Icare tonometry has made IOP monitoring much easier, especially in younger patients who do not tolerate applanation.

Dexamethasone and prednisolone cause a greater increase in IOP than fluorometholone, medrysone, rimexolone, and loteprednol. If a patient exhibits a steroid response, tapering off corticosteroids or transitioning to a topical steroid with less effect on IOP may be necessary [7]. For milder elevations of eye pressure, topical glaucoma medications should be initiated. First-line treatment is dorzolamide (1 drop twice daily), which is safe and effective in all pediatric age groups. Timolol (0.5% solution twice daily or 0.25% gel once daily) is also effective, but should be used cautiously in young children and asthmatics. Alpha-agonists, such as brimonidine 0.2% (1 drop twice daily), are effective but must be avoided in children under 6 years of age due to risk of CNS depression [8]. The prostaglandin analogue latanoprost (1 drop once daily) is safe, and it has a better IOP-lowering effect in older children [9]. It takes 2 weeks to reach a therapeutic effect, so latanoprost should not be selected to lower pressure acutely. For very elevated IOP, oral acetazolamide (15–30 mg/kg divided TID or QID) is sometimes necessary. Unlike in adults, acetazolamide can have an additive IOP-lowering effect when used concurrently with dorzolamide drops [10]. Acetazolamide is generally well tolerated, though there is a risk of metabolic acidosis, often presenting with tachypnea and light-headedness, which may require bicarbonate supplementation if the bicarbonate level is found to be low. Some pharmacies are able to compound acetazolamide in an oral solution for those patients who cannot swallow pills.

Case 2

A 7-year-old male presented for a secondary IOL implantation in both eyes. His past ocular history was significant for bilateral congenital cataracts that were removed at 3 months of age. His right eye underwent an anterior vitrectomy under visualization with intraoperative Triesence® with sulcus lens placement. His postoperative course was complicated by an IOP spike of 50 mmHg by Icare tonometry on postoperative day 1. This was accompanied by nausea and vomiting. His IOP was reduced in the office with topical dorzolamide/timolol and brimonidine and 250 mg of oral acetazolamide. He was sent home on 250 mg of acetazolamide 4×/day, Trusopt/timolol 2×/day, and brimonidine 2×/day. On postoperative day 4 he returned with an IOP of 10 mmHg in the right eye and was only taking acetazolamide at that time. He then returned on postoperative week 2, noncompliant with all topical glaucoma medications, and his IOP had increased to 42 mmHg in the right eye. It was again brought down with dorzolamide/timolol and brimonidine in the office. On subsequent visits his IOP returned to normal after tapering off topical corticosteroids, and he was weaned off topical glaucoma medications over several weeks. He had a similar course of steroid response glaucoma after secondary intraocular lens placement in his left eye.

Comment This case highlights the potential for IOP elevation following surgery after the use of intraocular Triesence and postoperative topical corticosteroids and the ability of the resulting pressure spike to be managed medically.

Visual Axis Opacification

In compliance with the official American Association of Pediatric Ophthalmology and Strabismus policy all children under the age of 9 should receive a posterior capsulotomy and anterior vitrectomy following cataract extraction to prevent visual axis opacification [11]. Surgeon discretion can be used in children >6 years of age, as some may be able to sit for a neodymium-yttrium-aluminum-garnet (Nd:YAG) capsulotomy. Visual axis opacification occurs in almost all pediatric patients that undergo cataract extraction without posterior capsulotomy and anterior vitrectomy and in up to 37% of patients who undergo posterior capsulotomy with anterior vitrectomy [12]. Pediatric patients who are unable to tolerate YAG capsulotomy in the office require repeat intraocular surgery with membranectomy to restore vision.

Glasses and Bifocal Use

Glasses are necessary after pediatric cataract surgery because the postoperative IOL target is typically hyperopic to adjust for eye growth over the first two decades of life. Generally, if vicryl sutures are used to close the corneal wounds, they dissolve by 1–2 months postoperatively, at which point a more stable final refraction can be obtained and glasses prescribed. Patients with IOL implants should receive spectacle correction for hyperopia >1 D, myopia >3 D, or astigmatism >1.5 D [2]. Patients should be left slightly myopic until 2–3 years of age at which point they can be transitioned into bifocals. Initially, the bifocal should be executive style with the line bisecting the pupil. Once a patient is well-adjusted to using the bifocal, they can be transitioned to a progressive bifocal if they prefer a less obvious bifocal appearance. If a patient has unilateral pseudophakia, the bifocal can be placed bilaterally and atropine used in the phakic eye to help them learn to use the bifocal segment well.

Amblyopia Treatment

Patients with unilateral or bilateral infantile cataracts develop dense amblyopia that must be aggressively treated following cataract surgery. Patching should be initiated 1–2 weeks postoperatively in the phakic eye in patients with unilateral cataracts. Recommendations on patching regimens vary. The Infant Aphakia Treatment Study patched patients for 1 hour per day per month of age until 8 months of age after which the phakic eye was patched for all hours the child was awake every other day or 1/2 of the child's waking hours per day [2]. There is now evidence that severe amblyopia may be successfully treated with patching the contralateral eye 6 hours per day [13]. Parents must be educated to only count patching hours when the

patient is awake and to provide full optical correction at all times. If the dominant eye is hyperopic, atropine penalization is as effective at treating amblyopia as occlusive patching and is a good alternative in patients who demonstrate difficulty tolerating patching [14].

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Part III
Correcting Surgically-Induced Aphakia

Chapter 11
Pediatric Aphakic Contact Lenses

James Hoekel

Uncorrected aphakia is as apt as the original lens opacity to limit visual potential. There exist situations where the best and safest option for a patient is to not place an intraocular lens (IOL). In these circumstances, aphakic contact lens fitting after lensectomy can be the preferred means of optical correction. With contact lens correction of aphakia, the image projected on the retina is of normal size and lacks the aberrations and distortion seen with high plus aphakic glasses. Additionally, high plus spectacles can induce a prismatic effect, decrease peripheral vision, and cause anisometropic diplopia. The weight and the thickness of the lenses can render aphakic glasses challenging for babies or children with flat nasal bridges to wear comfortably. With the advent of more materials, better education, and scientific studies, the outcomes with contact lenses are more predictable than a few decades prior. Another advantage of aphakic contact lenses is ease of replacement, especially when needing to adjust the power. Especially in a young eye with rapid axial length elongation, contact lens power adjustments can be made far more readily than new aphakic spectacles. Aphakic rehabilitation requires that the physician, the caregiver, and the patient work together to overcome hurdles leading to amblyopia. The ultimate goal is to provide the child with usable vision and prevent or limit amblyopia by eliminating sight-threatening blur.

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Fitting Aphakic Contact Lenses

Exam

The fitting process begins with a conversation between the eye care team and the child's family. Just as the surgical team discusses the risks, benefits, and alternatives to performing cataract surgery, the same discussion is necessary before fitting a child with a contact lens. The first decisions will be daily wear versus extended wear and soft lens versus rigid gas permeable (RGP). The parents' own history with contact lens wear may influence the choice for their child. For instance, if a parent is an RGP wearer, then they may be more comfortable with their child being fit with an RGP contact lens.

The timing of the initial fit is up to the team involved in caring for the child. The currently available literature supports fitting the child 1–27 days after surgery. In the author's practice, there are multiple surgeons with individual preferences for the timing of the initial fit. Fitting the child as early as possible is advisable, but can be complicated by the drops and ointment used during the postoperative period. The use of steroid antibiotic ointment is not compatible with contact lens wear, so it is recommended to use only steroid antibiotic drop after surgery or wait until the ointment is finished. The commonly prescribed prednisolone acetate is a suspension that can ruin the surface of silicone elastomer lenses.

Determining Diameter, Base Curve, and Power

The next step to proper fitting is determining necessary diameter, power, and base curve. If the infant is quite young, these values may be difficult to acquire. Many contact lens fitters utilize keratometry values, corneal diameter, and pachymetry from the examination at the time of the lensectomy. While this information is certainly useful, it is possible to achieve a good contact lens fit and prescription without any prior information. In young infants, the cornea tends to be steep, especially in eyes with persistent fetal vasculature (PFV).

At the fitting appointment, this author frequently has several new contact lenses for diagnostic purposes (Fig. 11.1). By having multiple lens choices, one can typically improve the likelihood of achieving an ideal fit. For use in infancy, many providers start with a SilSoft® Super Plus, which is a silicone elastomer lens developed by Bausch & Lomb. This silicone contact lens is made from a rubberlike material that is highly oxygen permeable and has been utilized for extended wear for over three decades since being introduced by Dow Corning Ophthalmics in 1983. The SilSoft® Super Plus comes in three base curves (7.5, 7.7, 7.9) in the 11.3 diameter. The power choices are +23.00 to +32.00 in three diopter steps. There is a larger diameter SilSoft® that has a 12.5 mm diameter, but it is only available in power choices from +12.00 to +20.00. The initial diameter is chosen based on the corneal

Fig. 11.1 Examples of aphakic contact lenses

diameter or what many surgeons refer to as the "white to white" measurement. In soft lens fitting, utilizing a larger diameter provides increased stability, but the lens becomes more difficult to insert and remove. If the edge of the soft contact lens does not completely cover the limbus, then the child may experience redness, discomfort, lens awareness, and punctate keratopathy.

The initial contact lens power is determined based on retinoscopic findings and then prescribed near addition. This author typically refracts twice in the fitting appointment to confirm correct power selection. The cycloplegic refraction is performed utilizing loose lenses and then the child is over-refracted with the contact lens in place. This adds time to the fitting appointment but it also serves as confirmation of the proper lens power.

Contact Lens Fit

The contact lens should move some, but not too much. It should conform to the cornea, but if too tight it can cause limbal impingement and if too loose it is uncomfortable and fails to provide sharp vision. Ideally, the lens should have 0.5 mm of movement and be easy to move with the finger. If at first the lens moves too much, allow the patient to wait 15 minutes before changing to a different lens, as the lens tightens after the initial tearing subsides. A child rubbing excessively is frequently displaying signs of poor comfort or fit.

At birth, the horizontal corneal diameter is about 10 mm and increases in the first 18 months of life [1]. As the cornea becomes flatter, the initial CTL may become too steep and have bubbles or tighten excessively and "pop off" of the eye. When this occurs and a SilSoft® lens is still desired, the 7.5 initial base curve should be changed to a 7.7 base curve. Small diameter corneas, such as in persistent fetal vasculature or microcornea/microphthalmia, need the smallest lens diameter available or a lens that is custom manufactured to an overall diameter between 11 and 12 mm.

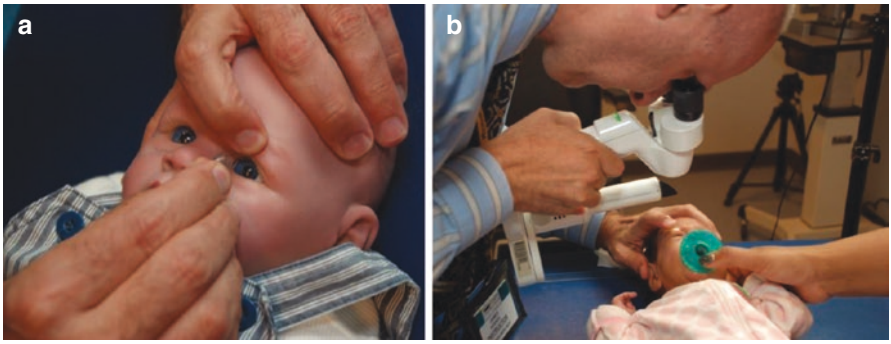

Fig. 11.2 (**a**) Insertion of a SilSoft contact lens. (**b**) Portable slit lamp exam for centration and fit

Case 1

AH was diagnosed with a congenital cataract on day of life one. Lensectomy without IOL implantation was performed at 4 weeks of age and contact lens fitting was scheduled 1 week postoperatively. The child's keratometry values were 48.75/49.00 in her aphakic eye. Her axial length was 17.1 mm. Her refraction was +21.50. This vertexes to +29.12 in contact lens power and 3.00 diopters were added to focus the child for near distances. A SilSoft +32.00 contact lens was chosen. Initially, this lens was a little flat for the steep infant cornea, but it was selected due to the ease of insertion (Fig. 11.2), good centration, and clear optics. The lens was dispensed on a 30-day extended wear basis. Since this was her first contact lens, the author evaluated the child at 1 week to ensure no extended wear complications occurred. A multipurpose contact lens solution was provided for cleaning and storage and also in case of contact lens coming out of the eye.

Comment This case describes the typical approach to an aphakic infant. A common frustration expressed by parents is when the lens is seen to float off of the cornea. In this case, the parent would diligently look and share with the office how many times per month the lens moved out of position or came off the eye. This information is useful for determining need to change lens parameters. The author recommends a 1-week follow-up to measure fixation and perform portable slit lamp biomicroscopy and over-refraction with trial lenses. The author also removes the lens to look carefully at the cornea, limbus, and anterior chamber, measures IOP, and inspects the lens under the microscope. If the lens is too cloudy or has already accumulated deposits, then a new contact lens is inserted. Multiple studies have established that there are inherent risks with contact lens wear, especially on an extended wear basis, but with diligence and good fitting lenses, infantile wear is quite safe [2–10].

Selection of Aphakic Contact Lenses

When the eye is open, the cornea receives oxygen from the atmosphere, the limbal vasculature, and the aqueous humor. Contact lenses sit on the eye, obstructing the complete transmission of oxygen to the eye. Each lens material has intrinsic oxygen permeability (Dk). The contact lens manufacturers publish the Dk for each contact lens and when fitting an aphakic child, the thickness of the lens should also be taken into consideration. This measurement is called critical oxygen transmissibility (Dk/t), with 't' being the thickness of the lens. The Dk/t of the lens should be 125×10^{-9} for extended wear and 35×10^{-9} for daily wear [11]. The published oxygen permeability of silicone elastomer lenses is $340 \times 10^{-11,}$ but there is some debate as the reports of oxygen transmissibility through thick lenses vary in the literature [11]. Currently with RGPs, there are a number of lenses with known Dk values greater than 100. Ophthalmologic and optometric research have established how much oxygen is required for safe contact lens wear and how much oxygen permeability is tolerable before one may experience pathologic changes in corneal morphology [12].

Custom Contact Lenses

Custom contact lenses are an indispensable tool, freeing the provider from being limited by only a few base curves and a couple diameters. The ability to design, order, and dispense a contact lens that will fit a child exactly confers a higher success rate of both comfort and visual clarity. In the early days of contact lenses, poly-2-hydroxyethyl methacrylate (HEMA) could be made in a variety of specifications, but the lenses were limited in their use in high plus pediatric correction due to reduced oxygen permeability. The advent of silicone hydrogels introduced a lens that delivered greater oxygen permeability with less fragility of silicone elastomer. Silicone hydrogels are different than silicone elastomer lenses.

One such silicone hydrogel material (Definitive®) can be lathe cut by manufacturers of specialty soft contact lenses. Currently there are several major contact lens manufacturers that have higher Dk silicone hydrogels, but at the time of this publication the lens powers are not available in high plus for aphakia. The Definitive® material typically provides robust oxygen permeability, good comfort, and quality optics (Dk/t up to 60). A contact lens lab may lathe this material into the curve, power, and diameter desired to provide an ideal fit. The ability to manufacture this in high plus with a variety of diameters and base curves gives the pediatric specialist a myriad of choices. Unfortunately, these lenses are not easy to insert in an infant, but should be considered if non-custom lenses are too tight, too loose, or coat and film too quickly. Significant chair time may be necessary to get an ideal fit as the stiffness of this lens renders it challenging to achieve this on the first try.

Some manufacturers will have a guaranteed fit period in which exchanges and reorders may be included, but each custom lens manufacturer has their own unique fitting policies. This lens has FDA approval for daily wear, but many providers prescribe 7-day 6-night off-label, extended wear.

When fitting a child with a custom contact lens look for a fit that does not compress the apex of the cornea. If a lens fit is too steep, it can create central corneal issues, peripheral tightening, and vascularization. While too steep of lens typically leaves a small space over the apex of the cornea, it is too tight in the periphery or mid-periphery causing discomfort, redness, limbal impingement, or improper tear exchange. Too loose of contact lens will display excessive movement and migrate temporally or superiorly. The silicone hydrogel must be fit appropriately; otherwise mechanical complications may arise such as contact lens-related papillary conjunctivitis and superior epithelial splits [11, 13–21]. Some practitioners utilize Fluresoft to evaluate contact lens staining patterns, although it can stain the lens. It is possible to evaluate limbal coverage, soft lens movement, and pooling or bearing with a portable slit lamp or handheld Burton lamp. Follow-up slit lamp examinations during the fitting or refitting may alert the provider to the need to replace on a more frequent basis especially in the setting of papillary conjunctivitis, corneal staining, microcysts, mucin balls, edema or polymegathism.

Case 2

MB suffered from bilateral congenital cataracts. She underwent lensectomy and vitrectomy of her right eye at 6 weeks of age. The left eye underwent the same successful lensectomy and vitrectomy at 7 weeks. She did well in the immediate postoperative period and, at 24 months of life, underwent implantation of an MA60 IOL in her right eye. At 25 months of life an MA60 IOL was implanted in her left eye.

Postoperative day one and week one visits were unremarkable. At the 1-month post-op visit, the left IOL appeared subluxed below the visual axis. The child underwent an examination under anesthesia with attempt at lens repositioning, but this was not successful as there was not enough capsule to support an IOL so she was referred for aphakic contact lens fitting.

As a 25-month-old infant her corneal diameter was 10.5 mm, her keratometry values in the aphakic eye were 42.62/43.00, and her axial length was 20.79. Her refraction at this time was +14.50. At this time Ciba made a custom silicone hydrogel lens called O2 Optix. This silicone hydrogel innovation had a Dk/t of 117 at −3.00. The patient was fit with a +18.50 contact lens in the 8.00 base curve radius (BCR) with a 13.2 overall diameter (OAD). Over the following 6 months there were no significant changes in refractive power but the lens became too small and failed to provide stable corneal coverage. At 31 months of life she was refit into the 8.4 BCR with a 14.0 OAD. This larger lens had better centration and provided less movement off her eye.

Ciba vision discontinued manufacturing her contact lens when she was around 50 months of age. The child had enjoyed silicone hydrogel's comfort and

replaceability since the lens came in a 4 pack; we wanted to continue in that same modality. Xcel contact lenses made the same parameter lens in a multipack utilizing Definitive® silicone hydrogel material. Over the following 6 years her visual acuity in the contact lens has been equal to the visual acuity in her pseudophakic eye although over time this eye has been become dramatically more myopic, whereas the aphakic eye has minimally changed refraction. As the child grew older, insertion became much easier and the child switched from extended wear to daily wear.

Comment This case reviews the process of initial contact lens selection and the need for reevaluation in the care of children with aphakic contact lenses. The author selected a silicone hydrogel lens for this unilaterally aphakic child as the Dk was 175×10^{-9} and the lens came in multipack options. This case further highlights how eye growth can lead to changes in type and parameters of contact lenses. In addition, factors such as manufacturer availability can have an impact on lens selection. It is encouraging to note that in this case of bilateral cataracts, the aphakic and pseudophakic eyes developed equal visual acuity.

Gas Permeable Lenses (Rigid Gas Permeable)

Gas permeable (GP) lenses for pediatric aphakia have advantages and disadvantages (Table 11.1). They are useful for the refractive treatment of traumatic aphakia and post pediatric penetrating keratoplasty. Each manufacturer has their own fitting guidelines and typically provides consultation lines to assist in achieving an ideal fit. Sitting for corneal topography is helpful when the child is old enough to do so.

The high plus GP has increased thickness; in addition, because its center of gravity is more anterior, its biggest challenge is centration. Many providers will utilize a lenticular design or a corneoscleral design in order to achieve stability.

Table 11.1 Comparison of Soft versus Gas Permeable contact lenses

Soft contact lens	Rigid gas permeable
Advantages	*Advantages*
Good initial comfort	Can mask or correct corneal astigmatism
Relatively uncomplicated fitting	Can be easy to insert and remove
Reduced provider time	Durable and long lasting
Less parental apprehension	Can be manufactured for high oxygen permeability
Easily replaceable	
Disadvantages	*Disadvantages*
Microbial keratitis	May have parental apprehension
Potential hypoxia concerns	Injection and redness
Relatively sparse approval for extended wear in high powers	Keratopathy or abrasion
	Initial discomfort
	Increased provider time (chair time)
	Relatively few choices in hyper Dk material

If the lens decenters inferiorly, this can cause corneal staining, contact lens discomfort, conjunctival injection, and unstable acuity. If the high plus lens is manufactured with a minus lenticular, then this may assist the upper lid in holding the contact lens in position.

Currently, there are numerous hyper Dk lenses that provide adequate oxygen permeability and safe extended wear. For the successful fitting of an infant, it is recommended to use a diagnostic fitting set. Often these fitting sets are loaned from the lab of choice or purchased from the manufacturer, distributor, or contact lens supplier. The initial lens may be chosen based upon the age/table/previous keratometry value of fitting experience. It is most important to be precise in base curve choice as to not create irritation and corneal harm. For a neonate the initial base curve of choice will be 7.00 mm with a +29.00 as a diagnostic lens. After allowing the contact lens to settle the power can be checked with an over-refraction, fluorescein staining can be evaluated to see if the lens is bearing or vaulting, and the overall diameter of the lens can be assessed to determine its stability and movement. At this stage, one should also evaluate the GP for edge lift and centration.

Caring and Wearing Aphakic Lenses

Insertion and Removal

Many parents will initially struggle with the insertion and removal of pediatric aphakic contact lenses. There are a number of contact lens providers that will insert the lenses for the parents on a weekly or monthly basis, although this practice is time intensive for both the family and provider. This author always tries to identify those families that struggle with insertion and provide "anytime" service as most children can have their lens inserted by an experienced practitioner rather quickly. Lie the child flat with a staff member holding the head and parent holding the hands of the child (Fig. 11.3). Right-handed providers may find it easier to stand on the child's right side in order to insert. The size of the lens and the amount of squeezing and the depth of the orbit and the vertical distance between the lid margins all factor into the challenges and ease of insertion.

Upkeep and Wear

Silicone elastomer lenses like the SilSoft® are prone to film development due to the proteinaceous nature of tears. This renders it necessary to replace the lens anywhere from monthly to every 6 months. Handling of this lens requires diligent and careful precautions. The use of soaps, lotions, creams, ointments, perfumes, and deodorants may cause irritation or damage to the lens. Customer service representatives

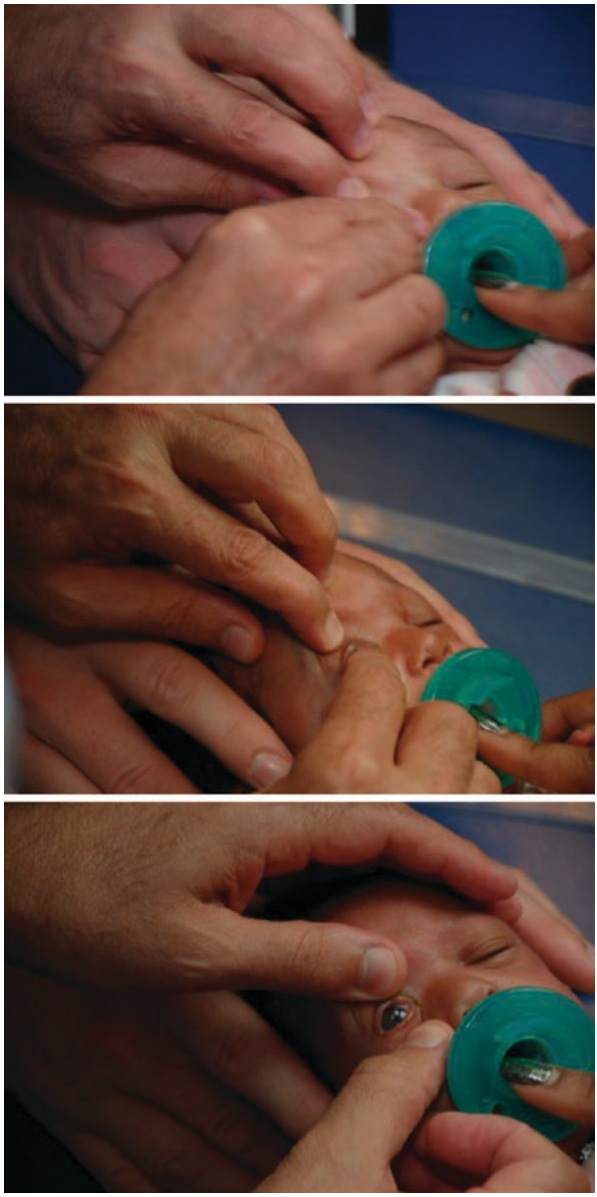

Fig. 11.3 Insertion of contact lens in a young infant by a right-handed author

frequently recommend non-scented glycerin soaps to avoid contamination and filming of the lens. Many hospital-based offices will have antibacterial soaps with lotion additives that may film and ruin the surface of the lens. Parents should be informed of the need to rinse hands thoroughly before handling the lens to avoid film buildup. Over the last two decades, contact lens solutions have improved with additives that clean the lens better and help keep the surface moist. However, these solutions can

interact poorly with silicone and silicone hydrogel lenses. This author recommends the use of BioTrue® for cleaning and storage but there are a large number of multipurpose solutions to choose from.

Complications

There are reassuringly few complications associated with SilSoft® lenses although the greatest concern is microbial keratitis and corneal ulcers. The incidence of microbial keratitis in extended wear is variable in infants and toddlers [9]. The rate in infants may be lower due to the robust tear film and diligence of newborn parents. Many fitters worry that the higher rate of community-acquired conjunctivitis in this young population places children at higher risk, but a well-fitting lens is the best protection.

Central scarring, neovascularization, and central or peripheral infiltrates are also devastating adverse events for contact lens wearers. There are also minor and temporary complications such as keratopathy, edema, injection, and lid swelling. However, the most frequent complication is lens loss. Compliance is variable in children and when contact lens compliance is poor, the child is certainly more at risk for underuse or misuse. Many of these contact lenses are quite expensive and some insurers will not cover their cost even when medically indicated. Some children will become intolerant of contact lenses during the 18–48-month age even in the setting of an ideal fit and good tear surface.

Continuous wear lenses (formerly referred to as extended wear) require vigilance as wear on an extended wear basis places a child at risk of hypoxic-related events including edema, infection, neovascularization, scarring, keratitis, endothelial polymegathism, pleomorphism, epithelial microcysts, and epithelial thinning [22]. When selecting such a lens, the goal is to have a lens that provides enough oxygen to the cornea when the child is awake and especially when the eyes are closed during sleep. It has been exciting to watch the contact lens industry improve oxygen transmission through improved materials.

The risk of potential harm to a child's cornea by a contact lens is outweighed by the visually devastating condition of uncorrected aphakia. Most contact lens experts advocate that extended wear is far riskier to corneal health than daily wear, but in a young child who struggles with insertion, it frequently becomes more practical to leave the lens in place. As the SilSoft® and some hyperDk GP materials may see the lens surface degrade with excessive handling, it may be beneficial to use extended wear. The alternative option is the use of daily wear. Like many duties of the parent, daily insertion and removal of a lens is initially a chore that is met with mixed emotions, but eventually becomes a routine and less of a hassle. It is with great hope that polymer chemistry and improved engineering will create more lens choices with thinner centers, multipack options, increased wettability, and increased oxygen transmission to deliver the best visual treatments for those that need it at the most critical time in visual development.

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Chapter 12
Aphakic Glasses

Christina M. Twardowski

Optical correction for aphakic patients is uniquely challenging secondary to age. The pediatric eye is undergoing anatomical evolution and the visual system is immature; due to the former, IOLs are not the first line in infants <7 months of age, and due to the latter, optimized optical correction is necessary [1]. With the evolution of contact lenses, many providers are gravitating toward them as a primary method of treatment. But glasses should not be forgotten, as their simplicity can lend to many benefits for the patient and practitioner.

Some may view aphakic glasses as a last resort due to their cosmetic appearance and cumbersome weight, but with careful consideration and appropriate use this refractive correction has many advantages. Glasses provide the highest ease of use for parents and patients, which can help reduce frequent follow-ups and ease parental stress during an otherwise difficult time. In addition, many insurance companies cover at least a portion of the cost of glasses. The medical coverage for contact lenses varies greatly by insurance company, with no guarantee of specific reimbursement for patients even when advocated as "medically necessary." In addition, contact lenses need to be frequently replaced due to patient loss (rubbing, falling out when sleeping, etc.) and/or FDA contact lens replacement hygiene recommendations. As a result, the cost of contact lenses typically far surpasses glasses.

Timing of Correction

It is critical to provide optical correction for aphakic patients promptly following surgery to optimize visual development. There are some cases where congenital cataracts are detected late (after 4 months of age), but an aggressive postoperative visual

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rehabilitation plan can make the difference in visual outcome [2]. Due to the high-power optical design of aphakic glasses, the time required for the manufacturer to produce these types of lenses usually takes an extra 2–3 weeks compared to a standard prescription. As a result, short-term contact lens wear may be considered in the interim in order to achieve optical clarity as early as possible. Assuming no complications arise after the lensectomy procedure, a contact lens may be inserted within 1 week postoperatively. This contact lens can stay in place until the glasses have been received, at which point the contact lens is removed and the patient is able to proceed with spectacles.

Optics of Refraction

Performing accurate retinoscopy is imperative for aphakic patients, as it is the foundation of their visual rehabilitation. When performing this technique postoperatively, most patients are still using atropine and although the cycloplegic effect is not necessary as the patient can no longer accommodate, the enlarged pupil is particularly helpful. There are times after the postoperative period where retinoscopy will need to be performed but the pupil of the patient may be quite small. In these situations it may be beneficial to add a dilating drop to increase the diameter of the pupil and improve retinoscopy precision.

A critical optical component to remember when performing retinoscopy on the aphakic patient is vertex distance. Vertex distance is the measurement between the back surface of corrective lens and the front surface of the cornea. This becomes important when the doctor is holding the trial lens during the neutralization of the retinoscopy reflex. The key is to place the neutralizing lens at the same location the corrective lenses will be positioned (Fig. 12.1). Many times the doctor holds the

Fig. 12.1 Shows the correct placement of the neutralizing lens to ensure the focal length of the correcting lens and the far point of the eye correspond

neutralizing trial lens further from the patient, which optically leads to an inadequate amount of prescribed hyperopia. Remember the goal of the refractive correction is to have the focal point of the corrective lens and the far point of the eye correspond. It is important to keep in mind that the focal length of a lens remains constant, which means if the neutralizing lens is placed further away from the patient's face, compared to the position of the corrective lens, the focal length will no longer match with the patient's far point.

Prescribing Considerations

Overcorrection

When prescribing optical correction for a pediatric aphakic patient, overcorrection is a necessity. Overcorrection is the process of adding additional plus power to the retinoscopy results to compensate for the lack of accommodation in aphakic patients. This is especially important for infants as they are highly stimulated by near objects and depend on clarity of their near world to make critical visual connections. Depending upon the age, the amount of overcorrection will vary, as the older a child becomes the more their visual world transitions from all near objects to a combination of distance and near stimuli. For example, a 6-month-old aphakic patient whose refraction is found to be +20.00 DS should be given a spectacle prescription for +23.00 DS. This additional plus power will set the focal length for the patient at 33 cm, which is the arm's length of an infant. The focal length is important as it should be the distance an infant is able to hold and view objects with their glasses in place. In general, when any concerns arise with overcorrection it will optically be in the patient's best interest to prescribe more plus power as the patient can always move the object closer to adjust for the focal point, but cannot necessarily extend the object further if an inadequate amount of power is given.

A general guideline for overcorrection can be seen in Table 12.1.

Table 12.1 Refractive overcorrection for aphakic patients

Infant – 1 y.o.	Three diopters
1 y.o. – 2 y.o.	Two diopters
2 y.o. – 3 y.o.	One diopter
3 y.o. – 4 y.o.	Bifocal

Bifocal Lenses

The decision to switch over to bifocal lenses versus continuing with overcorrection comes between the second and fourth year of life. The appreciation for distance clarity begins to develop during this age and the concept of using a bifocal will begin to be understood.

Bilateral Versus Unilateral Aphakic Patients

In bilaterally aphakic patients, glasses can be offered to all patients as large amounts of anisometropia rarely occur. This allows for easy prescribing and minimal concern for a cosmetically unequal appearance. On the other hand, in the unilateral aphakic patient anisometropia and asymmetric lens thickness and weight result in an unsatisfactory cosmetic appearance and challenge amblyopia treatment.

One strategy for dealing with the unbalanced appearance is to prescribe a balance lens over the sound eye. This lens would be similar in power to the aphakic eye, providing an equalized cosmetic look that helps stabilize the frame on the patient's face. In addition, a balance lens would fog the image quality of the sound eye acting as a form of amblyopia treatment. The balanced lens concept for unilateral aphakic patients is best used during the first 1–2 years of life. After that time conventional glasses tend to work well as the patient's growth leads to a decrease in refractive error, improved neck/head control, and increased head size. Together these lead to improved compliance and fewer complications related to spectacle wear.

Lenses

When choosing lens material options for these high-power prescriptions, aspheric designs and high index of refraction lenses should be considered. Aspheric polycarbonate lenses are an excellent option for kids as they are cost effective and safe. The aspheric design flattens the front surface of the lens, reducing thickness/weight of the lens and maintaining consistent magnification of the image across the lens surface. The optics behind high index lenses renders them better able to bend light rays to provide a clear image. This means the edges of the lens are thinner, which requires less material and ultimately decreases the overall weight of the lens. A standard plastic lens typically has a 1.50 index of refraction vs 1.74, which is the highest index lens that is available at this time.

Fig. 12.2 Miraflex® frame with color spool options

Frame

Frame considerations should be discussed with the family when dispensing a spectacle prescription as an inappropriately fitting frame can lead to poor compliance and optical degradation. In infants who require glasses, a soft plastic frame with a head strap is ideal (Fig. 12.2). The plastic material and strap component help keep the glasses in place, ensuring that the patient is looking through the optical centers. Metal frames tend to move more frequently, sliding down the patient's nose and breaking more easily. In addition, the high-powered lenses sit better in a plastic frame compared to a metal frame. This is due to the thicker frame edge that can accommodate the bulkier high-powered lens. Another recommendation is erring toward selecting a smaller, rounded frame. This design helps to decrease the weight of the lenses and optical aberrations in the periphery, as there is less lens to be affected by these distortions.

Case

A 7-week-old infant presents to clinic for a 1-week postoperative appointment following a lensectomy and anterior vitrectomy procedure with intentional aphakia in the right eye. Exam is unremarkable for any surgical complications. Retinoscopy was performed in the aphakic right eye and found to be +21.00 DS. At this time a spectacle prescription is written, right eye: +24.00 D; left eye: balance.

Since it will take approximately 3–4 weeks before the glasses are complete, a +32.00 Silsoft contact lens is inserted in the office. The power of the contact lens is determined by first using the effective power equation (see Fig. 12.3); this formula will calculate the modified lens power that is needed due to the alteration in vertex distance. The placement of the contact lens is located directly on the eye, compared to spectacle lens, which is located at a specific vertex distance in front of the eye. Once the appropriate contact lens power has been determined, the additional near power must be added. The effective power equation determines the patient's +21.00 D refractive power is equal +28.88 D. Then +3.00 needs to be added to focus the

Fig. 12.3 Effective power equation

$$F_{new} = F_{current}/(1 - dF_{current})$$

child at near. The resulting power is +31.88 D, which equates to the +32.00 D Silsoft contact lens that is being used. The patient is instructed to continue postoperative eye drops with the contact lens in place and begin occlusion treatment of the left eye for 6 waking hours of the day.

A follow-up examination is conducted in 3 weeks. At this time the patient has completed all postoperative eye drops and the contact lens is removed since the spectacle correction is now available. The patient begins wearing her glasses prescription 6 waking hours per day and follow-up examinations every 3–4 months are recommended to monitor the patient's vision, refraction, and eye pressure.

When the patient turns 2 years old the balance lens of the left eye is removed due to the presence of +3.50 D astigmatic refractive correction. The vision is 20/40 in both eyes using isolated picture testing. The patient now wears her glasses all day with aphakic refractive power in the right eye and astigmatic correction in the left eye; this spectacle change will ensure proper visual development of the left eye. In addition, the patient continues part time occlusion of 6 h/day of the left eye to optimize visual improvement of the right eye.

Comment The ability to switch from one refractive modality to another is easy and can be guided by the needs and wants of the family. It is crucial to remember that the refractive modality alone does not determine the visual acuity outcome, but rather it is the compliance of treatment. In addition, it is important to monitor the vision and refractive status of both eyes, even in patients with unilateral aphakia, and continue to tailor the treatment for the requirements of the patient's evolving visual system.

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Chapter 13
Primary Intraocular Lens Implantation

Natalie C. Weil and Scott R. Lambert

Primary intraocular lens (IOL) implantation is performed when a cataract is removed and an IOL is implanted at the time of cataract surgery. Cataract surgery is recommended for any visually significant lens opacity in a child. It should be performed at 4–8 weeks of age for infants with a unilateral visually significant cataract and at 6–8 weeks of age for infants with bilateral visually significant cataracts. While cataract surgery at even earlier ages may result in improved visual results, it is generally not recommended because of the increased risk of adverse events associated with cataract surgery before 6 weeks of age [1–3]. When cataracts are diagnosed after age 8 weeks, cataract surgery should be performed as soon as possible because of the risk of amblyopia worsening with a delay in treatment. Bilateral immediate sequential cataract surgery is advocated in many situations for young children to expedite their visual rehabilitation and to reduce their exposure to general anesthesia, with appropriate precautions taken to minimize the risk of bilateral endophthalmitis and toxic anterior segment syndrome [4].

Primary IOL Versus Aphakia

Primary IOL implantation is generally performed at the time of cataract surgery for children 1 year of age or older [5]; however, there remains controversy as to whether IOL implantation in children less than 1 year of age is the best surgical decision.

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The Infant Aphakia Treatment Study (IATS) found that infants less than 7 months of age who underwent primary IOL implantation experienced more adverse events and required more additional intraocular surgeries than children left aphakic and corrected with contact lenses. Most pediatric cataract surgeons in high income countries do not perform primary IOL implantation in infants less than 7 months of age unless there are social factors that would make contact lens correction difficult. In low income countries, primary IOL implantation is more commonly performed during infancy because of the limited availability of aphakic contact lenses.

In addition to social factors, the size of the eye must also be taken into consideration when deciding whether primary IOL implantation should be performed. Eyes with corneal diameters less than 9 mm should be left aphakic until the eye grows to an adequate size to accommodate an IOL since most commercially available are not sized appropriately for small eyes.

Lens Power Selection and Refractive Goal

One of the most challenging facets of primary IOL implantation in children is determining the most appropriate lens power to implant. Since the eyes of young children are rapidly growing, implanting an IOL that targets emmetropia at the time of cataract surgery may result in a large myopic refractive error only a few years later. For this reason, most pediatric cataract surgeons target hyperopia (undercorrection) for the eyes of children at the time of cataract surgery [6]. A variety of recommendations for IOL powers in children have been made based on estimated ocular growth with a goal of emmetropia between 7 and 10 years of age [7–9] (Table 13.1). Table 13.2 represents authors' preference.

When patients are undercorrected at the time of primary IOL implantation, they should have their residual refractive error corrected as soon as it can be accurately determined. Children who are <2 years of age should be given single vision glasses with a near-point correction. Children older than 2 years of age should be corrected

Table 13.1 Postoperative target refraction tables for residual hyperopia in children

Age at surgery (years)	Enyedi 1998 [7]	Crouch 2002 [8]	Plager 2002 [9]
<1			
1	+6	+4.0	
2	+5	+3.5	
3	+4	+2.5	+5
4	+3	+2.5	+4
5	+2	+2.0	+3
6	+1	+2.0	+2.25
7	Plano	+1.0	+1.5
8		+1.0	+1.0
10		Plano	+0.5[a] + 0.5%

[a]Emmetropia or slightly myopia after age 10

Table 13.2 Authors' target refractions in pediatric cataract surgery

Age at surgery	Target refraction for residual hyperopia
1 year	+5
2 years	+4
3 years	+3.50
4 years	+3
5 years	+2
6 years	+1.50
7 years	Plano

for emmetropia and given a bifocal segment for near correction. Children that have unilateral aphakia can be corrected with a contact lens using the same age guidelines; children who are <2 years of age should be corrected with a contact lens for near-point correction and those older than 2 years of age should be corrected for emmetropia with a contact lens and given a bifocal segment for near correction.

Placement of Intraocular Lens

Intraocular lenses can be placed in the capsular bag and sulcus or fixated to the fused anterior and posterior capsular remnants. Capsular bag fixation is most commonly used. One of the advantages of primary IOL implantation is that it facilitates implantation of the IOL in the capsular bag (Case 1). Implantation of an IOL in the capsular bag generally results in excellent long-term centration. Either a one-piece or three-piece IOL can be used for intracapsular IOL implantation. The anterior capsulorhexis should be made only slightly smaller than the diameter of the lens optic to facilitate placement of the haptics into the capsular bag. Primary IOL implantation can also be performed in the ciliary sulcus. If there is adequate capsular support, the haptics of the IOL can be positioned anterior to the capsular ring. If the capsule has been torn or there is zonular dehiscence, it may be necessary to fixate the haptics with sutures or scleral fixation. Only three-piece IOLs with posterior angulation should be used for sulcus fixation because of the risk of inducing the uveitis-glaucoma-hyphema (UGH) syndrome from chaffing of the iris by the haptics of a one-piece IOL. A sulcus fixated IOL can either be left anterior to the capsular bag or captured behind the capsular bag. In optic capture the anterior and posterior leaflets of the capsule are sealed anterior to the optic, except for the area around the optic-haptic junction. This technique prevents migration of lens epithelial cells along the vitreous face. Successful capture can be demonstrated when the lens optic is beyond the posterior capsule and an ellipsoid shape of the posterior continuous curvilinear capsulorhexis (PCCC) is noted (Case 2) [10]. Limitations to this procedure include a PCCC that is too small, leading to posterior capsular tear or a PCCC that is too large and unable to capture the optic [11]. A third option is placing the remnants of the capsular bag in a groove on a specially designed IOL ("bag in the lens" technique) [12]. This involves placing the capsular ring in a groove on the lens that allows the anterior and posterior capsular bag remnants to fuse together, thereby reducing the risk of reproliferating lens material growing into the pupillary space.

Posterior Capsule Management

One of the challenges of pediatric cataract surgery is that if the posterior capsule is left intact posterior capsular opacification (PCO) universally occurs. In some instances preserving the posterior capsule may be indicated, such as in older age groups, history of posterior segment pathology, or ocular inflammation. Since treatment of PCO generally requires a reoperation in young children, a posterior capsulotomy should be created at the time of primary IOL implantation. Various techniques may be used to create a posterior capsulotomy, either with a vitrector or manually. One commonly used approach is to use a vitrector after aspirating lens cortex. The posterior capsule opening should be well centered, concentric, and smaller than the anterior capsulorhexis (4 mm). A central anterior vitrectomy can then be created. A limbal or pars plana approach can be used to create a posterior capsulotomy with a vitrector. The advantage of a limbal approach is that the same incisions used to remove the cataract can be used to perform the posterior capsulotomy and anterior vitrectomy. With the pars plana approach, the location of the entry site should be based on the age of the child. In patients <1 year the incision should be made 2 mm posterior to limbus. In patients 1–4 years the incision should be made 2.5 mm posterior to the limbus, and in patients >4 years the incision should be 3 mm posterior to the limbus [13, 14]. The pars plana approach has the advantage that the IOL is implanted with the posterior capsule intact [15]. However, it has the disadvantage that a pars plana incision has to be created and there are concerns about the long-term safety of this technique.

A posterior capsular opening can also be performed using a manual PCCC technique. Because the posterior lens capsule is 3–5 times thinner than the anterior capsule, a manual PCCC is technically challenging [16]. The use of an ophthalmic viscoelastic device (OVD) is necessary to flatten the capsule while creating a manual PCCC. Some surgeons use the technique of making a small hole in the posterior capsule and injecting OVD to displace the vitreous face posteriorly in order to prevent it from being opened when the posterior capsulotomy is being performed [17]. In addition, by displacing the anterior hyaloid membrane posteriorly, some have hypothesized that this reduces the likelihood that the anterior hyaloid can act as a scaffold for future lens epithelial cell migration [18–20]. Performing a PCCC without a vitrectomy frequently results in opacification of the anterior hyaloid membrane. However, using this approach makes it more difficult to implant the IOL in the capsular bag (Case 1). When implanting an IOL in an eye with a posterior capsulotomy, it is very important to keep the lens flat while injecting it into the capsular bag, since the lens can easily be injected into the posterior segment of the eye through the posterior capsular opening. Once the anterior portion of the lens is in the correct plane the lens can easily be rotated into place using an IOL manipulator.

If the posterior capsule is left intact at the time of cataract surgery, then an in office neodymium:YAG capsulotomy can be performed for cooperative children when the posterior capsule opacifies. In young children, PCO can occur within a few months, but in an older child PCO usually develops 1–2 years after cataract surgery. Multiple YAG laser treatments or even a surgical posterior capsulotomy may be necessary to clear the visual axis [21]. If it is deemed likely that the child will not cooperate with an office YAG capsulotomy, a posterior capsulotomy should

be performed at the time of cataract surgery. While a YAG laser is available in some operating rooms, performing this procedure in the operating room exposes a child to general anesthesia and in most cases the procedure must be performed with the child in a sitting position, which is difficult with an intubated child.

Wound Construction in Primary IOLs

The most common options for the primary wound in pediatric cataract surgery are either a scleral tunnel or a corneal incision. The advantage of a scleral tunnel wound is that there is a lower risk of wound leak and there is no visible corneal scar. However, this technique does require that the conjunctiva be disturbed, thus resulting in more postoperative discomfort and making it more difficult to perform glaucoma procedures in the future. A scleral tunnel is constructed using a beveled ophthalmic blade starting 2–3 mm posterior to the limbus. The initial grove should be the width of the IOL injector that will be used. The tunnel should extend into the cornea, but not enter the anterior chamber. Entry into the anterior chamber is completed using a smaller blade, with the incision size depending on the size of the instrument used to aspirate the lens. The wound is later enlarged for IOL insertion with a keratome blade.

The advantage of a corneal incision is the conjunctiva is not disturbed and iris prolapse is less likely to occur since the wound is more anterior. However, in younger children it can result in a visible corneal scar. An initial small incision is made for cataract removal and vitrectomy and the wound is then enlarged with a keratome for lens insertion (Fig. 13.1). Starting with a small incision for capsulorhexis and lensectomy prior to IOL insertion allows for better anterior chamber fluidics and less anterior chamber collapse.

For either the scleral tunnel or corneal incision, the wound is usually made superiorly so that it will be protected by the upper eyelid. Most pediatric cataract surgeons suture the wound with absorbable polyglactin suture (10–0 for clear corneal incisions, 9–0 for scleral tunneled incisions).

Case 1

A 4-year-old male presented with bilateral congenital cataracts. His medical history was significant for being born at 37 gestational weeks with jaundice and pulmonary edema requiring a 1-month hospitalization. He was diagnosed with bilateral congenital cataracts at age 3 years. The first surgeon who evaluated him did not recommend cataract surgery. The child's parents had not noted a problem with his vision, but did note increased photophobia. There was no family history of congenital cataracts. Slit lamp examination was significant for bilateral lamellar cataracts with best corrected visual acuity of 20/40 OD and 20/60 OS. Due to decreased vision and increased light sensitivity secondary to cataracts, he underwent bilateral cataract surgery, first on the left eye as shown in Fig. 13.1.

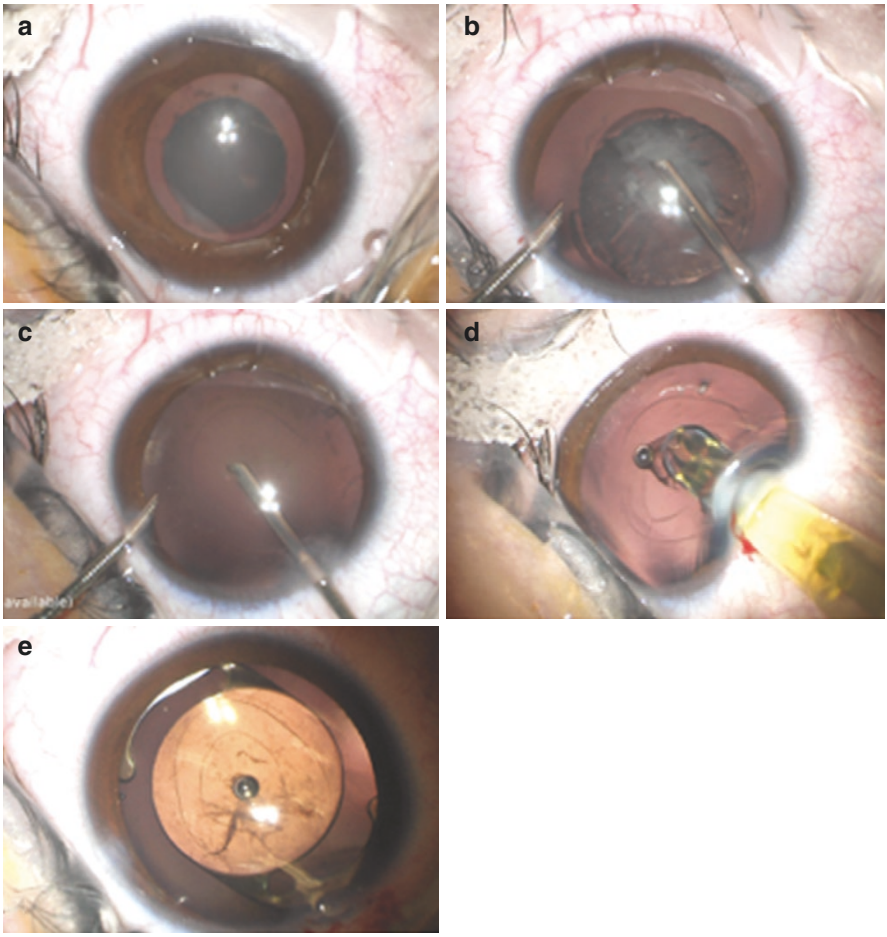

Fig. 13.1 (**a**) Lamellar unilateral cataract of the left eye. (**b**) A 5 mm anterior capsulorhexis has been made. The lens cortex and nucleus are being aspirated with a vitrector using a corneal incision. A Lewicky cannula is being used to maintain the anterior chamber. (**c**) A posterior capsulotomy is created through the corneal incision using the vitrector on shave mode. It is smaller than the anterior capsulotomy. (**d**) The capsular bag has been filled with an OVD, and the one-piece IOL is being injected into the capsular bag after enlarging the corneal incision to 3 mm with a keratome blade. Note the flat delivery of the leading edge of the lens into the capsular bag to ensure that the leading haptic is positioned in the capsular bag inferiorly. (**e**) The IOL is shown after being positioned in the lens capsular bag. The larger anterior capsulotomy and smaller posterior capsulotomy are visible. The wound is then sutured closed with 10.0 absorbable polyglactin suture and the remaining OVD is aspirated with a Simcoe cannula

Comment This case demonstrates primary IOL implantation in a 4-year-old. His age made the likelihood of posterior capsule opacification higher and he was judged to be too young to sit successfully for a postoperative YAG capsulotomy. Therefore, a posterior capsulotomy was created, in this circumstance, using the anterior surgical incision. Wounds were closed using 10–0 absorbable sutures.

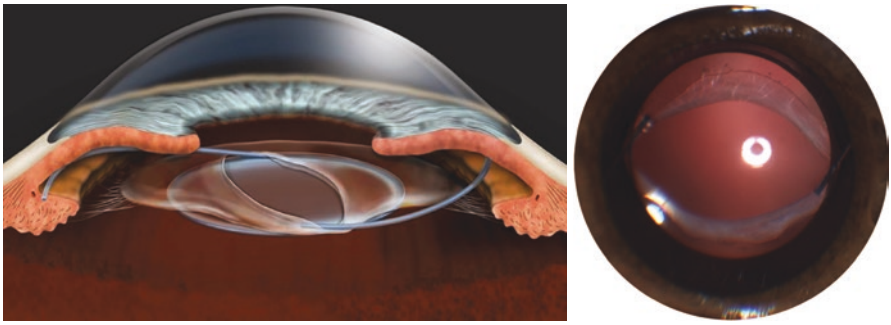

Fig. 13.2 Intraocular lens capture. (Images provided by Stephen Lipsky MD)

Case 2

A 2-year-old male was referred by his pediatrician for right eye drifting over the past 3 months. His family stated that he had a normal birth history and denied any significant medical problems. On exam, he had poor fixation with the right eye and a 20 prism diopter intermittent exotropia. On slit lamp exam a dense lamellar cataract was present in the right eye and the left lens was clear. B-scan of the right eye demonstrated no signs of retinal detachment or stalk associated with persistent fetal vasculature. His family agreed to proceed with cataract removal and IOL placement. Based on his age, a three-piece lens was selected with a target of +4.00 intentional hyperopia.

An anterior manual continuous curvilinear capsulorhexis 5 mm in diameter is created and the cataract is removed in its entirety. A 4 mm posterior capsulotomy is then made concentric to the anterior capsulorhexis. The lens is injected into the lens sulcus and optic carefully pushed behind the anterior and posterior capsule, creating an ellipsoid shape of the capsule (Fig. 13.2).

Comment In this case, a three-piece IOL was selected and optic capture was performed to enhance lens stability. The resulting fusion of the anterior and posterior leaflets of the capsule anterior to the optic prevents migration of lens epithelial cells along the vitreous face. This technique reduces the likelihood of lens reproliferation into the visual axis in this young child.

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Chapter 14
Refractive Targets

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Choosing the initial postoperative refractive target for an infant or child who will be undergoing cataract extraction with intraocular lens (IOL) implantation is perhaps the area in which pediatric IOL surgery differs most from the procedure in adults. It is also the area where there is the most nuance in the surgeon's approach and decisions.

The mature eye's refractive state is not expected to change much during the years following surgery, whereas the very young eye is highly likely to undergo considerable change as part of normal growth and development. This creates a situation where the final postoperative refractive error can be unexpected and unsatisfactory. While myopic shifts are anticipated, how much and how quickly are tremendously variable. Some patients do not reach emmetropia at all, remaining hyperopic. Others quickly become myopic and may even require an IOL exchange (see Chap. 18: IOL exchange).

In a Delphi process, pediatric cataract surgeons reached a consensus on the following targeted postoperative refractions according to age: <6 months, +6-10D; 6–12 months, +4-6D; 1–3 years, +4D; 3–4 years, +3D; 4–6 years, +2-3D; 6–8 years, +1-2D; and >8 years, 0-1D [1]. In the Infant Aphakia Treatment Study, targeted hyperopia for infants (+8 for those 4–6 weeks of age, +6 for those 6 weeks to 6 months) was recommended [2]. Following these guidelines, most pediatric cataract surgeons elect for varying degrees of hyperopia for the pseudophakic child. The goal is gradual progress toward emmetropization, with refractive correction in the form of glasses and/or contact lenses along the way.

However, while adults may bristle at a need to wear spectacle correction after surgery, compliance is rarely a real problem. In early childhood, simply keeping glasses in place can be challenging or even impossible. This makes an unexpected or large refractive error more challenging to treat. A high degree of refractive error very early in life may contribute to amblyopia or impede its treatment, with a

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potentially major impact on the eye's ultimate vision. The young pseudophakic eye's lack of accommodation greatly amplifies the cost of uncorrected hyperopic refractive error, particularly in unilateral cases with a normally accommodating fellow eye. For this reason, when targeting hyperopia, the importance of spectacle compliance must be emphasized with caregivers and, when possible, the patient.

Another unique consideration when determining the refractive target for a child is intraocular lens availability. It is an infrequent occurrence for adults to require an IOL outside of the normal range available in most surgery centers. However, when preoperative lens calculations suggest a high plus or a low plus (or even a myopic) implant is required, this can be obtained prior to surgery date. However, the IOL power required to achieve emmetropia or desired hyperopia in very young eyes may be over +30 and unavailable in some implant models. Even if commercially available, a particular lens power may not be stocked in a surgery center and since lens calculations are frequently performed during an exam under anesthesia, the need would not be known.

One advantage of cataract surgery in childhood is neural plasticity. The adult patient's capacity to adapt to an abrupt change in refractive status is much less than a child's, the former situation creating potential for significant patient dissatisfaction with an outcome that differs significantly from preoperative refractive status, even if such a change could be viewed as advantageous; no such concern exists before maturity.

Because an important goal of most pediatric IOL implantations is optimizing adult refractive status, surgeons, starting during the first years of the procedure in the 1990s, have focused on choosing a lens power that will result in the most desirable refraction in maturity. Generally, this is thought to be emmetropia or low minus power requirement, with less concern about refraction during the short- and mid-term postoperative periods. Ample evidence of overall trend in the myopic direction (Table 14.1) [3–8] led to the establishment of a recommendation for targeting an early refraction on the plus side, more so in younger patients (Table 14.2).

Table 14.1 Refractive change after IOL surgery

Author (year)	Number eyes	Age years (mean or range)	F/U years (mean)	Shift diopters/ year (mean)	Shift diopters total (range)	Shift SD/ mean
Brady (1995)	45	7.2	1.5	−0.45	+4.25 to −4.00	
Hutchinson (1997)	21	6.3	3.2	−0.31	+0.38 to −3.25	
Dahan (1997)	68	0–1.5	6.9	−0.92		
	36	1.5–3	3.5	−0.79		
	52	3–8	3.8	−0.68		
Enyedi (1998)	12	0–2	2.5	−0.9	+0 to −10	0.9
	23	2–4	2.2	−1.8	+5 to −10	1.8
	16	4–6	1.9	−1.5	+1 to −5	1.5
	9	6–8	3.0	−1.6	+2 to −6	1.6

F/U follow-up, *SD* standard deviation

Table 14.2 Target refraction recommendations (Enyedi, 1998)

Age years	Target diopters
1	+6
2	+5
3	+4
4	+3
5	+2
6	+1

This approach does have two potential drawbacks. The lack of pseudophakic accommodation makes it imperative that refractive correction, generally in the form of spectacles, be provided immediately and consistently from the earliest postoperative days. Failure to consistently wear glasses results in poorly focused images, especially for viewing at near, where the young child's vision is mostly directed. This creates an amblyogenic situation, more harmful with younger age and higher degrees of "pseudohyperopia." This is especially true if one eye remains phakic.

"Pseudomyopia," on the other hand, like naturally occurring myopia, is less likely to contribute as significantly to amblyopia, particularly if it develops, as is more likely, later in childhood [9]. While refractive shift in the direction of myopia occurs most often after pediatric cataract surgery, variability among eyes is very large, as indicated by Table 14.1. Some progress to substantial myopia even from a starting point well on the plus side, while others remain stubbornly pseudohyperopic or even shift in the plus direction, necessitating lifelong low plus correction that may result in significant dissatisfaction for the family and ultimately the patient.

Based on these considerations, it has been the author's practice to target early post-op emmetropia for most eyes undergoing IOL surgery in childhood, regardless of age [10]. Inevitably this results in many eyes that become significantly myopic by maturity. The impact of such acquired myopia is mitigated by a number of circumstances. The uncorrected refractive state of the involved eye continuously permits exposure to optically sharp images at some distance that is easily achievable in the real world, minimizing amblyogenic stress. Very young children with uncorrected myopia, even bilateral, are generally not particularly bothered by the condition. They typically pay limited attention to distant parts of the world and are usually happy to approach any object of interest for close inspection. Most often by the time myopia develops after IOL surgery (typically years), the child is at an age when spectacle wear is reasonably well tolerated and considered socially acceptable. Contact lens correction for myopia of any degree poses fewer problems than in cases of low plus power requirement and can usually be achieved without much difficulty by the age at which high minus power may be needed. Finally, keratorefractive surgery is a very reasonable solution for the young adult who desires permanent correction. However, it is important to consider that the axial length elongation in the first 24 months can give rise to rapid myopia if emmetropia is targeted in this young age range.

A recent retrospective comparative clinical study from two institutions using different targeting strategies, with surgery performed age 2–6 years and mean follow-up 6 years, showed no significant difference in final best corrected visual acuity between two groups of 12 patients each with mean initial refractions of −0.1 D and + 3.3 D [11]. Final refraction ranged from −4.5 to +1.1 D (mean − 2.0, standard

deviation 1.7) in the near-emmetropia targeted group and −1.8 to +3.5 D (mean + 1.3, standard deviation 1.6) in the plus targeted group.

The counter perspective, held by many surgeons, is to prefer a refractive target other than emmetropia [12]. A number of tables exist to assist the surgeon is selecting targeted hyperopia (see Chap. 13: Primary Intraocular Lens Placement). If the fellow eye is myopic and either already pseudophakic or unlikely to require lens surgery, it may be appropriate to aim for an early refraction within about 3D of that eye and similar or less net refractive error if possible. When surgery is planned for both eyes at the outset, risk of amblyopia from symmetrical bilateral pseudohyperopia is much less than in the unilateral case, and use of glasses at a young age is likely to be less problematic than if only one eye is affected. In these situations, starting more hyperopic in hope of reducing the ultimate degree of myopia is reasonable.

With surgery in infancy, particularly before age 6 months, considerable myopic shift is highly probable, but ability to predict ultimate refraction is also very poor [13]. Infants who qualify for an implant are likely to lose most of their pseudohyperopia within a few months if less than a +6 to +8 is set as the target.

Case 1

A healthy 12-month-old girl with no significant family ocular history was referred by her pediatrician for "dull reflex right eye"; previous evaluations by the same doctor had shown no abnormality, and the parents had noted no disturbance of vision or eye appearance. Eye examination showed good fixation with each eye, but a left eye (OS) preference. Grating acuity measured with Teller cards was markedly reduced in the right eye (OD). Pupils, alignment, and motility were normal. Retinoscopy reflex OD was poor secondary to a posterior cortical lens opacity; no significant refractive error was noted OS.

Anterior segment findings under general anesthesia 2 weeks later included normal symmetrical corneal diameter (11.0–11.5 mm both eyes) and keratometry (mean 46.0 D both eyes). Intraocular pressure was normal bilaterally. Findings with handheld slit lamp included normal left eye and normal anterior segment in the right except for the lens, which showed dense opacification of the central 2.5 mm of posterior cortex and partial nuclear opacification with diameter 4–5 mm; no retrolental plaque or vessels were present. Fundus appearance was normal and symmetric in both eyes.

Retinoscopy with full cycloplegia was estimated to be -12D OD, plano OS. Axial length measured by A-scan biometry was 22.1 mm OD, 19.2 mm OS. Intraocular lens power calculation for emmetropia OD was 21.0 (SRK-II formula) to 21.5 (SRK-T formula).

Lensectomy was performed in standard fashion, including removal of the central 2.5 mm of posterior capsule, which remained opaque but was otherwise unremarkable after cortical aspiration, and limited anterior vitrectomy. A +21 power one-piece PMMA lens was placed in the capsular bag.

One month after surgery, refraction in the pseudophakic eye was +1.00 + 1.00 x 90. Visual fixation was good, and alignment normal. Teller card grating acuity was improved but remained considerably reduced. Five months after surgery (age 17 months) refraction was unchanged. Single vision glasses were prescribed to correct the full astigmatic error in both eyes and full hyperopic error in the right. Compliance with glasses and patching (up to 6–8 hours/day) was excellent.

At age 3 years, refraction remained the same in both eyes. Visual acuity measured 20/60 OD, 20/25 OS with best distance correction. A 25 prism diopter intermittent exotropia was present, with only small exophoria and fusion for near. At age 5 years, refraction was −1.00 + 1.00 × 90 OD, plano +1.50 × 90 OS; corrected VA 20/30 OD, 20/20 OS; and motility unchanged, with stereo 200 seconds. First bifocal lens was prescribed for the right eye, with +2.50 add.

At age 12 years, both eyes had undergone myopic shifts and corrected to 20/25 OD, 20/20 OS. In 2019, at age 21 years, refraction was −5.00 + 2.00 × 65 OD, −5.50 + 1.50 × 90 OS; VA with glasses was 20/20- OD, 20/20 OS. The lens implant was well positioned, with a small central posterior capsular opening. IOP and fundus appearance were normal and similar in both eyes. Having had no further procedures since her original surgery, the patient was orthophoric for distance and about to enter law school.

Comment The above case is from the author's personal experience, chosen due to 20 years of continuous follow-up. Though not necessarily typical, this case provides an example of how targeting near emmetropia can succeed over the long term. When diagnosed with unilateral cataract secondary to congenital central posterior capsule abnormality, this patient already had unilateral axial myopia, attributed to the effect of visual deprivation. Her unaffected eye was plano, notably not hyperopic as expected in this age range. Lensectomy and IOL implantation at age 12 months resulted in low hyperopia for her pseudophakic eye; then refraction did not budge for nearly 4 years. With conscientious refractive correction and patching for amblyopia from infancy, she achieved a remarkably good visual outcome. Theoretically if an early postoperative refraction of +6.00 had been targeted, her refractive journey may have included far more anisometropia, which complicates amblyopia treatment.

Case 2

A 15-month-old girl was referred for an intermittent exotropia of her right eye. On exam, she was central, steady, unmaintained in the right eye, and central, steady, maintained in the left. She had a constant exotropia of 30 prism diopters at distance and an intermittent deviation of 15 at near. Exam was notable for a patchy posterior cortical opacity obscuring 4 mm of the red reflex OD. Anterior segment exam was otherwise normal in both eyes. It was possible to view the posterior pole in the right eye, which appeared grossly normal but view was poor. Cycloplegic refraction was challenging in the right and +1.50 sphere OS.

Parents had initiated patching for 2 hours/day OS prior to consultation. They were having moderate success but were highly motivated. After extensive conversation, including need for amblyopia treatment and glasses use after surgery, cataract extraction with IOL placement and anterior vitrectomy was planned. Target for the surgical eye was +5.0.

Exam under anesthesia confirmed normal intraocular pressures in both eyes. Portable slit lamp evaluation confirmed the right lens had a 4 mm posterior cortical opacification and a dense posterior plaque measuring 2 mm centrally. Axial length measurements were 19.55 mm OD and 20.12 mm OS. Cataract extraction, implantation of a SA60AT 26.0 diopter lens (Alcon, USA), and pars plana posterior capsulotomy and vitrectomy were performed.

The child did well in the immediate postoperative period. She obtained glasses at postoperative week 3, when refraction was judged to be stable from previous week. Her refraction at that time was +5.00 + 0.75 × 90. She was given glasses with a prescription of +7.00 + 0.75 × 90 OD and plano OS. She was tolerating 4 hours of patching a day.

She did well over the ensuing 6 months, tolerating glasses and patching. Distance exotropia remained constant and eye preference testing improved to intermittent maintain on the right. She received updated glasses 12 months following surgery. At this point, her cycloplegic refraction was +4.50 + 0.75 × 90 OD and + 1.00 OS. Her glasses prescription was +4.50 + 0.75 × 90 OD, plano OS. Bilateral bifocal add of +3.00 was introduced.

Six months later, her visual acuity could be tested using HOTV matching. She was 20/150 OD and 20/25 OS. Parents continued to patch 4 hours a day. Distance exotropia had improved with the glasses change.

At last follow-up at age 4, she was 20/80 best-corrected OD and 20/20 OS with a cycloplegic refraction of +3.25 + 1.25 × 80 OD and + 0.25 + 0.25 × 110 OS. She was wearing glasses full time, with full cycloplegic refraction on the right, plano on the left, and a bifocal add.

Comment This case is from the editor's practice, illustrating the clinical course of a patient with intentional hyperopic postoperative refractive error and the necessary resultant spectacle dependency. In this example, the young girl did very well with glasses correction. She was initially prescribed glasses with overcorrection to focus her world at near. As she aged, her glasses were changed to bifocals. Of note, she was given a bifocal add in her phakic eye as well. This encourages the child to engage the near add in both eyes and facilitates the use of the amblyopic eye at near. With time, she has undergone an expected reduction in her hyperopic refractive error. Her visual acuity likely reflects the later presentation with a unilateral cataract, but diligent patching history.

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Chapter 15
Multifocal and Accommodating Intraocular Lenses

Basak Can Ermerak, William Johnson, and Erin Stahl

Loss of accommodation is one of the main challenges faced by pediatric cataract surgeons during postoperative management and visual rehabilitation of their patients. Without accommodation, dependence on corrective aids to provide image clarity over multiple distances becomes necessary. With continued innovation in surgical techniques and intraocular lens (IOL) design, enhanced visual and refractive outcomes have become possible for the adult population. Multifocal, extended depth of focus (EDOF), and accommodating IOLs, sometimes termed "premium IOLs," have allowed modern cataract surgery to provide spectacle-free, clear vision at nearly all distances for select patients. These options have extensively reduced patients' overall postoperative spectacle dependence and improved their quality of life, though evidence demonstrates careful patient selection remains a key component when considering presbyopia-managing IOLs.

For pediatric patients, loss of accommodation after cataract surgery is a well-published contributor to amblyopia, with subsequent disruption of binocular vision. Thus, early rehabilitation of near vision after lens extraction has utmost importance in this at-risk group [1–3]. The ability to have a range of functional vision has become an exciting topic for pediatric cataract surgeons given presbyopia-managing IOL technology has been steadily evolving since its introduction in the 1990s [4]. As these novel applications are being investigated and widely adopted in adults, the same question arises for many pediatric ophthalmologists: Would premium IOLs be an adequate and safe alternative to conventional monofocal options without sacrificing image quality, degrading contrast sensitivity, or introducing dysphotopsias?

In practice, pediatric cataract surgeons likely experience preoperative questions influenced by the senile cataract experience, such as, "Will my child need to wear glasses/contact lenses after her/his cataract surgery?" From the lay perspective, the

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opportunity or expectation of spectacle independence is commonplace, due to awareness of adult cataract surgery from direct to consumer advertising, interaction with family and friends who have experienced lens-based procedures, or confusion with other ophthalmic surgery, such as laser-based keratorefractive procedures. Of course, all of these assumptions are false. Nonetheless, they remain prevalent, likely to be an enduring feature of the collective outlook of patients' parents and grandparents due to the success of adult cataract surgery. Furthermore, if a patient's family is aware of "premium" options from advertising or personal experience, they may be confused or suspicious if they do not hear at least a discussion of the technology. It would be natural for a family to want to make every effort to optimize the child's long-term spectacle independence, though the durability of such an expectation remains tenuous given the myriad of variables including age of the patient, visual potential of the eye, etiology of lens abnormality, possible surgical complications, and amount of remaining axial growth.

Varying degrees of near spectacle mitigating strategies are available with a monofocal IOL in place. Classic monovision provided by disparate IOL powers is not typically feasible in pediatric patients due to the unknown remaining refractive shift. However, future intervention to allow monovision such as photorefractive keratectomy (PRK), laser-assisted in situ keratomileusis (LASIK), or contact lenses could provide simultaneous near and distance vision. Often monovision techniques remain limited in providing adequate depth of vision or a true level of near. Multifocal contact lenses are also an option, though the quality of vision may be worse than a similarly designed IOL and it exposes the patient to the risks of contact lens wear. The recent suggestion of multifocal contact lenses delaying the progression of myopia in pediatric patients [5, 6] raises the question of whether presbyopia-managing intraocular lenses may affect the level of postoperative refractive myopic shift in patients following cataract extraction.

A feature that has been described in pediatric patients with monofocal intraocular lenses is pseudo-accommodation. A study conducted in France showed 90% of pediatric patients demonstrated pseudo-accommodation compared to 7% of adult patients [7]. The threshold for pseudo-accommodation was 20/50 near vision in the presence of 20/25 corrected distance vision or better. All patients were able to achieve 20/25 near with appropriate diopter near add. The presence of higher order corneal aberrations, higher IOL power, and spherical equivalent were positively correlated with the presence of pseudo-accommodation in both adult and pediatric age groups. Shift in IOL position and pupil aperture size were not correlated with pseudo-accommodation. This suggests pediatric patients with monofocal IOLs may perform better than adult patients with a similar implant unaided at near, though this may decrease with age and a 20/50 definition of pseudo-accommodation is unlikely to be universally suitable for near tasks in pediatric or adult patients.

Neural plasticity has been described as an important component in adequately tolerating an IOL designed to provide depth of vision. Dysphotopsias are common side effects of presbyopia-correcting IOLs, typically described as halos or starbursts. An improvement in the degree patients are bothered by these dysphotopsias over the first several months postoperatively, in the absence of posterior capsule

opacification or other optically significant pathology, has been attributed to neuroadaptation. Functional magnetic resonance imaging collected by Rosa and colleagues provided evidence for this process [8]. A study of a trifocal diffractive IOL in 2016 showed improvement in near acuity from the 1-month to 3-month postoperative evaluations, also suggestive of this process [9]. These findings have been described in the adult population, but the pediatric age group would theoretically harbor a larger degree of neural adaptability than patients in their seventh decade and beyond, potentially representing a positive attribute in the use of presbyopia-correcting IOLs in the pediatric population.

Lens Discussion

Multifocal intraocular lenses (MFIOLs) are designed to provide adequate functional outcomes at all distances unaided. Although different IOL technologies use various strategies to achieve this goal, the main principle is to divide incoming light into different foci and simultaneously create near and distance images on the retina. This multifocal optical design is based on dispersion of incoming rays, requiring a neuroadaptive process to accurately distinguish these multiple images [10]. Furthermore, MFIOLs consist of two or more focal points, each serving fixed working distances to deliver a sharp image to the retina [11]. When an object is viewed at a certain distance, the unwanted effect of simultaneously redirected light on the out-of-focus image may lead to reduction in contrast of the in-focus image. This design may also lead to the perception of photic phenomenon such as glare and halos [10–12].

Multifocal IOLs can be classified as refractive, diffractive, or a combination. The number of focal points embedded in the lens is another defining property of MFIOLs. Starting with the early generations of MFIOLs, a commonly implemented design was a bifocal. More recently, intermediate vision has also become important to daily routines due to increased utilization of computers, smartphones, and other electronic devices. To address the need for intermediate vision, low-add MFIOLs, extended depth of focus (EDOF) lenses with echelette design, trifocal MFIOLs, and "mix-and-match" strategies have emerged to optimize distance, intermediate, and near vision. The pediatric population has also experienced this dramatic increase in intermediate vision demands, though younger children likely maintain the need for mostly near with reading and electronic tasks given their shorter arm length and positioning from electronic screen stimuli.

Refractive IOLs consist of annular optical zones of differing, or sometimes alternating, dioptric powers for distance and near foci, in order to create simultaneous images. The main limitation of this design is dependence on pupil size and sensitivity for decentration [10, 13]. This can be explained by varying pupil dynamics under photopic and scotopic conditions, affecting the balance of distance and near foci.

The AMO Array lens (Allergan, Irvine, CA) was the first refractive MFIOL to become United States Food and Drug Administration (FDA) approved in 1997. This foldable, silicone lens provided better results for uncorrected and corrected distance

visual acuity along with better distance-corrected near visual acuity over monofocal lenses [14–17]. Overall spectacle dependence was lower in multifocal groups; however, increased rates of halos and glare along with some loss of contrast sensitivity were reported [15–18]. This lens was upgraded to the ReZoom IOL (Advanced Medical Optics, Santa Ana, CA), which was approved by the FDA in 2005. The ReZoom is a three-piece, hydrophobic acrylic lens with a distance-dominant center and five alternating concentric zones with aspheric transitions, theoretically providing increased image quality at distance and intermediate focal lengths [19]. These modifications led to decreased complaints of glare and halos, along with favorable distance, intermediate, and near visual acuity results [20, 21].

Another hydrophobic acrylic, refractive MFIOL commercially available in Europe is the Lentis Mplus (Oculentis, Berlin, Germany). Instead of using symmetric concentric multifocal rings, this lens has a rotationally asymmetric segmental, bifocal design with a surface-embedded near section. It offers good uncorrected and distance-corrected near visual acuity with high contrast sensitivity, although image quality continues to be affected by IOL tilt and decentration [22, 23]. A newer generation is now available in the European market, offering a toric option, the Lentis Mplus Toric (Oculentis, Berlin, Germany) for eyes with more than 1.50 diopters (D) of preexisting corneal astigmatism [24].

Diffractive MFIOLs use diffractive micro zones, or steps, on the posterior lens surface to direct light evenly between distant and near focal points. The 3M 815LE Lens (3M Corp, St Paul, Minnesota, USA) was one of the first generation diffractive MFIOLs that was then purchased by Alcon Laboratories and renamed ReSTOR (Alcon, Fort Worth, Texas, USA). It is a single-piece, hydrophobic acrylic, bifocal lens initially with a +4.0 D additional power for near vision in the IOL plane (approximately +3.2 D at the spectacle plane). It consists of 12 concentric diffractive rings utilizing an apodization principle and was the first diffractive MFIOL to obtain FDA approval in 2005 [25]. An apodized, diffractive MFIOL is one with steps that grow closer peripherally, helping to smooth the diffractive characteristic. If the central zone is devoted to a near focal point, then under gradually lower lighting, more of the light is then devoted to the distance focus in an effort to decrease glare and halos in mesopic conditions. Early studies provided results with good uncorrected visual performance at distance and near with less helpful intermediate range performance, due to the relatively high add [26–29]. Halos and glare were the most commonly reported visual phenomena for this type of lens [26–29]. While maintaining its apodized structure, the next-generation ReSTOR AcrySof IQ (Alcon, Fort Worth, Texas, USA) was designed as a hybrid diffractive and refractive lens and was granted with FDA approval in 2007. The addition of asphericity was intended to reduce the visual phenomena, with the ability of increased range of focus and improved image quality. The aspheric design further reduced the positive spherical aberration of the cornea to improve contrast sensitivity [10, 11]. Early results for this lens also showed good visual acuities at distance and near and maintained contrast sensitivity. The intermediate acuity was acceptable [30, 31]. The AcrySof IQ ReSTOR +3.00 D was later introduced to improve uncorrected intermediate range vision while maintaining optimal near and distance visual acuity results

[32]. This model provided a +3.00 D correction in the IOL plane or +2.6 D in the spectacle plane. Comparative studies between the +3.00 D and +4.00 D models showed improved intermediate vision with the +3.00 D model without meaningful loss of distance or near acuities [33–35]. Despite favorable contrast sensitivity outcomes, glares and halos continued to be reported [34, 35]. The AcrySof MFIOL line received FDA approval for a +2.50 D model, as well as toric versions in 2014 [36]. After FDA approval in late 2019, the most recent addition to the AcrySof family is the Panoptix IOL (Alcon, Fort Worth, TX, USA), a hydrophobic acrylic lens consisting of 15 diffractive rings along with a refractive only outer annulus zone [37]. Its trifocal optics provide approximate focal points of 60 cm for intermediate focus and 40 cm for near focus [38]. Early results demonstrated good visual outcomes for corrected and uncorrected distance, near, and intermediate distances [39].

The Tecnis multifocal IOL (Johnson & Johnson Surgical Vision, Santa Ana, CA) was approved by the FDA in 2010, as a single-piece acrylic diffractive following its earlier generations of Array and ReZoom lenses. This lens has an aspheric anterior surface with a bifocal add of +4.0 D on the IOL plane. The Tecnis multifocal IOL also has a fully diffractive posterior surface as opposed to the apodized diffractive design of the ReSTOR MFIOL. Overall, this pattern splits the light so that it is distributed evenly between the near and distant foci to mitigate the dependence on pupil size [40–42]. Early results demonstrated better uncorrected and distance-corrected near vision over the monofocal group with good uncorrected and corrected distance visual acuity [43, 44]. Furthermore, high patient satisfaction was achieved despite a slight loss of contrast sensitivity and reported glare and halos [43, 44]. The FDA later approved two models with less add power, +3.25 D and +2.75 D, in 2015 to better address intermediated vision. In a large, prospective, comparative case series between the three Tecnis MFIOLs (+4.00 D, +3.25 D and +2.75 D), the low-add options provided less spectacle dependence for near and distance vision along with higher patient satisfaction compared to previous high-add model. Contrast sensitivity was similar between the groups, and more than one-third of subjects reported experiencing glares or halos in each group [45].

In comparative studies between different designs of diffractive and refractive MFIOLs, all groups demonstrated high distance acuity, but diffractive designs provided better corrected and uncorrected near visual acuities [46–48]. Patient satisfaction was high in all groups despite a similar level of diminished contrast sensitivity and the presence of glare and halos [46, 47]. Additionally, a randomized, prospective, double-masked trial examined the ReSTOR +3.00 D and +4.00 D with the Tecnis MFIOL suggested that newer-generation aspheric, especially low-add hybrid apodized or full diffractive lenses are well-tolerated for working-age cataractous patients in visual outcomes, reading performance, and quality of life results. The ReStOR +4.00 D model, not surprisingly, showed the lowest distance-corrected intermediate visual acuity (DCIVA) and uncorrected intermediate visual acuity (UIVA) [49].

Other examples of diffractive MFIOLs not commercially available in the US market include the AT LISA (formerly known as the Acri.Lisa; Carl Zeiss Meditec, Dublin, CA) and FineVision (PhysIOL, Liège, Belgium). The AT LISA was

introduced as an aspheric, single-piece, bifocal MFIOL with a near add of +3.75 D at the lens plane. Its hybrid refractive-diffractive MFIOL design divides usable light 65% for distance and 35% for near [50]. Good outcomes of corrected and uncorrected distance and near vision has been reported along with similar drawbacks on diminished intermediate image [50–52]. AT LISA tri (Carl Zeiss Meditec, Germany) was then brought to market as a trifocal with a four-point haptic design to provide improved intermediate range of focus and better contrast sensitivity outcomes despite some visual phenomena [53, 54]. A toric version of this lens, AT LISA tri toric IOL (Carl Zeiss Meditec, Germany), has become available in Europe with promising clinical outcomes [55]. The FineVision lens is a relatively new, hydrophilic acrylic, full diffractive lens with double loop haptics. Its trifocal design with add powers of 1.75 D and 3.5 D reflects focal points of 40 cm for near and 80 cm for intermediate ranges [38]. Studies on the FineVision lens have shown good visual outcomes for all distances [56, 57].

Extended range of vision (EROV) or extended depth of focus (EDOF) IOLs is a relatively new concept proposed to enhance the focal range with improved image quality via an echelette design. Their mechanism works by focusing incoming light on an elongated longitudinal plane. Their unique diffractive pattern eliminates spherical aberrations within the IOLs allowing corneal spherical aberrations to create an extended depth of focus [10, 58, 59]. This elongated focus plane diminishes overlapping of near and far images to eliminate the common halo effect of traditional MFIOLs, though a "starburst" photopsia is more characteristic of this design. The Tecnis Symfony IOL (Johnson and Johnson Surgical Vision, Santa Ana, CA) is the only FDA-approved EDOF IOL, certified in 2016. The Symfony is also available as a toric lens. Recent adult studies report good uncorrected visual results on a wide focal range with few optical phenomena [59, 60].

By definition, accommodative intraocular lenses (AIOLs) are designed to produce a dynamic increase in the dioptric power of the eye with efforts to bring focus to near or intermediate range from a distance target [61, 62]. However, it may be helpful to first distinguish real accommodation and pseudo-accommodation. Pseudophakic accommodation is the ability of true dynamic refractive variations during near and intermediate vision to produce a clear image, whereas pseudo-accommodation includes increased depth of focus along with multifocality or aberrations and subjective adaption to defocus during near tasks [63]. Given the difficulty in consistently distinguishing these two categories, the American Academy of Ophthalmology (AAO) recently published a task force statement suggesting related clinical studies should use objective instrumentation and methodology to obtain accurate accommodation measurements [64]. The task of accommodation via an IOL may be achieved through alternations of the axial position, curvature, or refractive index [61, 62]. By initiation of accommodative effort, the ciliary muscle contracts along with a shift of the zonular diagram that subsequently leads to increased vitreous cavity pressure and forward movement of the lens complex, which can be adapted, in part or whole, to allow AIOLs to increase total refractive power of the eye. The Crystalens (Bausch and Lomb, Inc., Rochester, NY) is the only accommodative IOL currently approved by the FDA. The Crystalens design is a

monofocal, biconvex, silicone lens with relatively rigid haptics driving anterior displacement of the optic due to anterior-posterior hinges of the haptic with ciliary contraction, producing accommodation. Although there are some controversial results, accommodative IOLs provide similar results to monofocal IOLs regarding distance visual acuity and contrast sensitivity with better results in terms of near vision than monofocal IOLs [65, 66]. The Crystalens tends to have higher rates of posterior capsule opacification (PCO) [66]. A rare but visually significant complication of the Crystalens is the "Z syndrome," which is described as decentration and tilting of the lens secondary to capsular fibrosis resulting in one haptic hinge anteriorly displaced and one posteriorly displaced. This condition can usually be managed by neodymium-yttrium-aluminum garnet (Nd:YAG) laser [67, 68].

Toric presbyopia-correcting IOLs are available in many of the above lens designs, including ReSTOR Toric, Panoptix Toric, Tecnis Multifocal Toric, Tecnis Symfony Toric, Trulign (Crystalens Toric), and AT Lisa tri Toric. In certain situations, pediatric patients may have visually significant cylinder that may warrant treatment. However, in many cases, the expected shift in corneal astigmatism from "with-the-rule" to "against-the-rule" in adulthood may warrant a conservative approach rather than a more aggressive approach to treating corneal astigmatism, though, if necessary, IOLs with combined capability are now widely available.

Several new lens technologies are under development with potential benefit for presbyopia correction and use in pediatric cataract surgery specifically. Numerous additional emerging technologies are being investigated that may play a role in adjustable IOL power, but these are not specifically applicable for postoperative presbyopia management. One that may affect both monofocal adjustment and presbyopia-correcting adjustments would be in the style of the PreciSight Lens (InfiniteVision Optics, Strasbourg, France). This is a multicomponent IOL (MCIOL) that has an intracapsular base with a central portion to hold an exchangeable optic, theoretically exchangeable anytime postoperatively to optimize the desired optical correction [69]. A more novel mechanism would be in the style of the Perfect Lens (Perfect Lens, LLC, Irvine, CA), a femtosecond laser adjustment of an IOL that can adjust monofocal power, switch between monocular and multifocal, and can be adjusted repeatably without degradation of material [70]. Work on this technology is ongoing, but certainly represents an exciting frontier in presbyopia-managing IOLs for the pediatric population.

Premium IOLs have managed to provide encouragingly high rates of patient satisfaction over the years. Despite notable improvements, the major drawbacks of the multifocal intraocular lenses include visual symptoms (halos, glares), varying stages of diminished contrast sensitivity, and mesopic visual function [10, 11]. For optimal visual cortex function, clear image focus onto the retina is essential. Although these postoperative visual functions have largely been studied and described in the adult population, pediatric patients may be more or less vulnerable depending on the level of neuroadaptation.

Early visual near rehabilitation after cataract extraction has paramount importance to restore visual function and prevent amblyopia. Contrary to adults; children may not be able to perceive visual disturbances caused by light dispersion of

multifocal intraocular lens designs. Similarly, we cannot estimate the effect of having an out-of-focus image on final image quality or the ability of pediatric patients to adapt to this. Reports of decreased contrast sensitivity have been published for amblyopic patients [71, 72]. This could lead to an underestimation of diminished image quality after pediatric cataract surgery within the amblyogenic years [73, 74]. Currently there are two reports on amblyopia and MFIOL implantation, both in anisometropic adults without strabismus, which suggest improvements in visual acuity levels and binocular function [75, 76]. However, it would be premature to claim similar results for a pediatric population.

Challenges of Implementation

Postoperative refractive status is a complex, often changing, landscape in pediatric patients. The typically shorter axial length (AL) and higher keratometry (K) values change during the early years of life, leading to subsequent shifts in refractive error. Given this compensatory change (increasing AL and decreasing mean Ks) is even more brisk within the first 2 years, a greater degree of uncertainty is expected when surgery is performed at younger ages [77–81]. These features are important to selection of proper IOL power. Current common practice is to leave younger children with age-dependent amounts of hyperopic refractive error to allow myopic shift [14]. No specifically pediatric IOL calculation formulae currently exist (for further discussion see Chap. 9). Thus, after a desired postoperative refraction is selected, an IOL power selection can be made using an adult formula, although no consensus exists for which formula is best suited to this population [77–84].

Refractive outcomes for presbyopia-correcting IOLs remain challenging in the pediatric population compared to adults given axial growth prediction error, keratometry changes, difficulty in obtaining accurate biometry, and determination of the optimal preoperative IOL calculation. If a presbyopia-correcting IOL is implanted during childhood, the lens can only achieve the desired refraction for a limited time given the myopic shift with continued growth. However, from a theoretical perspective, targeting an immediate postoperative refraction of +3.00 D in a child could allow for immediate emmetropia to a degree via the near focus if a +4.00 multifocal IOL were used. Axial growth may continue during the second decade of life [85]. A large change would cause dependence on corrective aids or further surgery performed in early adulthood to address their refractive error [73, 85, 86].

The surgeon should always consider intraoperative challenges of pediatric cataract surgery over standard phacoemulsification. Proper implantation along with centration is a key factor affecting successful function of presbyopia-correcting IOLs [87]. Besides a well-centered anterior capsulotomy, a continuous and strong posterior capsulotomy is needed to prevent additional lens tilt and decentration that may eventually lead to diminished image quality. A thorough anterior vitrectomy is often employed in pediatric surgery to minimalize the risk of visual axis opacification postoperatively, which can also decrease risk of lens decentration due to lens

epithelial proliferation. The often-opened posterior capsule can make subsequent IOL exchange or explantation technically more challenging than those performed in adults with an intact posterior capsule. This can be a factor when considering a secondary presbyopia-correcting IOL exchanged for a primary monofocal or in the setting of primary aphakia, given all modern presbyopia-correcting IOLs are designed for implantation within the capsular bag.

Case-Based Application

Because consideration of a presbyopia-correcting IOL is unique to each patient, and discussed more often than utilized in our experience, the following case-based discussion will focus on three age ranges to help in applying the above principles, technology, and techniques into a clinical application, rather than recounting individual cases.

Case 1

A 4-month-old with unilateral fetal nuclear cataract.

Comment Infants would be unideal presbyopia-correcting IOL candidates. Based on the Infant Aphakia Treatment Study, children developed fewer complications, mainly visual axis opacification, when left aphakic. There remain certain instances that favor primary IOL implantation, such as demonstrated poor compliance with aphakic correction [88]. Additional factors not favoring presbyopia-correcting IOLs would be the need to overcorrect by up to 10 diopters to account for later growth. The ability of an infant brain to differentiate between multiple images remains to be demonstrated, and the amblyogenicity of unilateral cataracts does not favor this type of implant. The incidence of reoperation in this age group for obscuration of the visual axis, glaucoma, and other intraocular surgery indications would add a risk of lens decentration or damage. Perhaps most significant about this age group of cataractous eyes is the likelihood they have additional pathology likely to produce a subnormal visual potential even with aggressive occlusion therapy and refractive correction and would likely not be able to gain all the benefit of the presbyopia-correcting IOL technology.

Case 2

A 6-year-old patient with bilateral posterior subcapsular cataracts due to systemic corticosteroid therapy for unrelated systemic illness. Eyes are otherwise healthy with normal visual development. Normal fundus exam in both eyes.

Comment The older, though still amblyogenic, age group of 2–9-year-old patients represent a distinct scenario from infants <1 year of age. Eyes in this range have grown and matured enough to allow a technically easier surgery compared to infants. However, the still robust postoperative lens epithelial response can lead to visual axis opacification and lens tilt/decentration due to Soemmering's ring mass, the latter capable of skewing the optical properties of a presbyopia-correcting IOL. Brain development is still occurring with imperfectly understood impact from presbyopia-correcting IOLs, and the level of residual refractive shift makes power selection difficult. However, as eluded to above, there could be theoretical utility with an extended range of vision provided by an IOL if patients have targeted initial hyperopia. With regard to amblyopia, the decline in contrast sensitivity from presbyopia-correcting IOLs could negatively impact visual development. The case reports of amblyopic patients implanted as adults showed a favorable outcome with acuity and binocularity, but this is difficult to generalize to patients implanted in childhood. Without exception, this vignette is not complete without a thorough conversation with the patient's family and the patient and an evaluation of their visual needs and desires postoperatively. A strong desire for presbyopia management with an IOL would have to be expressed and justified. Given the need for planned hyperopia, a period of spectacle or contact lens correction would be necessary, but time-limited if the predicted shift with continued growth is accurate. Alternatively, the patient may elect to have keratorefractive surgery or an additional lens-based refractive surgery without explant of the initial presbyopia-correcting IOL. Overall, given present technology, the applicability is likely relatively unusual in this age group, though more likely than in younger children.

Case 3

A 13-year-old teenager developed bilateral posterior subcapsular cataracts. There was family history of similar cataract development in her 17-year-old sister. The sister had undergone uncomplicated cataract extraction but did not enjoy wearing reading glasses. The patient had an otherwise unremarkable eye exam.

Comment Children beyond the amblyogenic age range, approximately 9–18 years old, are the best candidates for presbyopia-correcting IOLs. Continued axial growth is still common, but these patients would be closest to the age of eligibility for keratorefractive treatment of their resultant refractive error. Patients in this age group receiving presbyopia-correcting IOLs will have a myopic shift in refraction, but the magnitude of this and the degree to which the patients would need to embrace spectacle correction would be variable based on tasks and personal preferences.

While current literature is limited regarding presbyopia-correcting IOL implantation in the pediatric population, there are a handful of studies on multifocal IOLs in the literature. Jacobi et al. published the first study in 2001 using the AMO Array Lens in 35 eyes of 26 patients between 2 and 14 years [89]. This study had an

average follow-up of 27.4 ± 12.7 months and reported a statistically improved best-corrected distance visual acuity, stereopsis, and spectacle independency at the last visit. Only 18% of patients had postoperative complaints of visual phenomena. In 2010, Cristóbal et al. published their results on 3M Lens implantation in five patients with unilateral cataract [90]. These patients were 4–6 years of age and the follow-up period of the study was 21 months. Five patients demonstrated improved visual acuity and four had improved stereoacuity. No patients reported glare or halos. Another retrospective study included 26 pediatric patients (34 eyes) aged between 2 and 15 years who underwent ReSTOR Lens implantation with a +4.00 D model [91]. At a mean follow-up of 25.73 ± 10.5 months, patients showed good results for distance and near vision with improvement in stereopsis. It should be noted these studies have self-reported data on visual disturbances and relatively short follow-up periods for postoperative pediatric cataract results. Wilson et al. published the first case report with long-term follow-up [92]. This report included three siblings with bilateral posterior subcapsular cataracts who underwent lens extraction and implantation of Array Multifocal IOL at 16, 19, and 16 years of age. A recall examination was performed at 12, 11, and 9 years, respectively. Four out of six eyes were noted to have a minimal refractive shift of ≤0.50 D. The third and oldest patient had 0.75 D of myopia in both eyes. Two patients denied glare, whereas one did not drive at night because of glare she experienced. The sibling experiencing glare was not interested in an IOL exchange. Another individual case report described a 7-year-old patient implanted with an apodized, diffractive, multifocal IOL [93]. At a 7-year-follow-up the patient had good visual acuity outcomes. A case report on bilateral implantation of AcrySof IQ PanOptix Lens in a 9-year-old boy was the first report on trifocal lens implantation in a pediatric patient. He had very good distance, intermediate, and near uncorrected visual outcomes [94]. The first comparative study between monofocal and multifocal intraocular lenses in a pediatric population was published in 2014 by Ram et al. [95]. This was a prospective, nonrandomized clinical trial of 42 eyes of 21 children who received a monofocal IOL or diffractive and refractive multifocal IOL. The mean age in both groups was approximately 7 years old with 1 year of follow-up. Corrected distance visual acuity levels were comparable for both groups, whereas the multifocal group showed significantly better distance-corrected near visual acuity results without decrease in contrast sensitivity. Prospective randomized trials with longer follow-up periods should be directed toward evaluation of presbyopia-correcting IOL implantation profiles in pediatric eyes in order to assess data and safety monitoring.

The unifying goal of pediatric cataract surgery remains preservation/development of the best visual acuity over as many focal points as early as possible for as long as possible from the fewest interventions with the fewest complications and with the lowest possible spectacle burden. Presbyopia-correcting IOLs potentially relate to each element of this mantra, though the details of their application remain incompletely understood. Many elements of preoperative planning can be difficult for the pediatric cataract surgeon, and each patient brings unique elements to consider a customized treatment, produced through counseling conversations with the patient and family as well as careful planning of the optimal surgical approach in

order to maximize each element of the goal previously stated. As with many elements in medicine, the optimal solution for each patient is likely to be a balance of priorities tailored individually instead of a truly perfect solution across all facets. Evaluation of new technology is an important component of advancing a discipline. Without studies and experience, the improvement of the technique and outcomes would have a tendency to remain stagnant, a result no advocate of the pediatric cataract patient would endorse as acceptable.

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Chapter 16
Temporary Polypseudophakia (Piggyback IOLs)

M. Edward Wilson and Rupal H. Trivedi

When a cataract is removed from a child's eye, the critical offset to axial eye growth is also removed. The process of emmetropization requires a crystalline lens to change its focusing power to match axial eye growth in an attempt to keep the image focused on the retina. We currently do not have any artificial intraocular lens (IOL) implants that automatically change as the eye grows. If the surgeon aims for emmetropia at the time of IOL insertion in a growing eye, myopia will develop over time. When surgery for cataracts is done early in life, this change in refractive error can create high degrees of myopia in a few years or even months. Surgery to exchange an IOL that is firmly "shrink-wrapped" into the capsular bag can be challenging. For these reasons, pediatric eye surgeons often aim for hyperopia immediately after cataract and IOL surgery when operating on young children and rely on glasses to correct the residual refractive error during eye growth. However, poor compliance with glasses can worsen amblyopia despite the presence of the IOL. A toddler may require 5 or 6 diopters (D) of intentional residual hyperopia after surgery if the goal is to achieve emmetropia at maturity and throughout adulthood. This approach reduces the chances that an IOL exchange will be needed when growth is complete. However, the downside is that if the glasses are not worn full time, the uncorrected residual hyperopia is amblyogenic, especially for children whose visual world is mostly at near.

With these concerns in mind, one of the authors (MEW) introduced the concept of temporary multiple IOLs (or polypseudophakia or piggyback IOL) [1]. Primary implantation of multiple IOLs has been described to provide adequate IOL power to adult patients when sufficient power of a single IOL was not available [2]. In contrast to adults where primary piggyback IOL is intended to stay permanently, in children with temporary polypseudophakia, the posterior IOL is implanted in the capsular bag (permanent) and the anterior IOL is placed in the ciliary sulcus

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(temporary), a location from which it may be easily removed at a later point. This concept reduces the amount of hyperopia during infancy and myopia during adulthood by removing the anterior IOL when the eye becomes sufficiently myopic. It is well known that hyperopia is more amblyogenic than myopia. With this technique, during the critical period of visual development, patients are able to minimize the need to wear thick spectacles or contact lenses. We are following 40 children (51 eyes) implanted with primary temporary polypseudophakia and 44 of those eyes now have more than 5 years of follow-up. The median follow-up of those 44 eyes is 12.24 years. Planned piggyback IOL removal has occurred at a median of 3.24 years after implantation. Only four eyes underwent unplanned, early piggyback IOL removal, one each for IOL tilt, pupillary block, pupillary capture, and a pupillary membrane. Each of those complications occurred when the piggyback technique was used on infants younger than 7 months of age.

Case Report

A 22-month-old female child presented with a diagnosis of bilateral anterior polar cataracts since birth. The cataract in the left eye had gradually progressed over the preceding few months and was visually significant. A sensory exotropia had developed and the child strongly objected to having the right eye covered. The cataract in the left eye was a pyramidal anterior polar opacity with extension into the underlying cortex and nucleus, while a very small anterior polar cataract in the fellow right eye was visually nonsignificant. Examination under anesthesia was scheduled along with surgery on the left eye. The globe axial length (AL) of the left eye was 0.61 mm longer than the right eye, further suggesting the presence of deprivation amblyopia (Table 16.1). After thoroughly discussing options with the parents, piggyback IOLs were selected as it was felt to be the best way to visually rehabilitate the eye. The child was nearing her second birthday and the parents acknowledged that full-time spectacles would be a burden and they were fearful that they would not be able to comply. Biometry predicted an IOL power for emmetropia of 27.76 D (calculated

Table 16.1 Preoperative parameters

	RE	LE
IOP (mmHg)	10	9
Keratometry (D)	42.25/45.25	43.50/49.50
Axial length (mm)	20.23	20.84
Anterior chamber depth (mm)	3.33	3.26
Lens thickness (mm)	3.67	4.21
Corneal diameter (mm)	11.5	11
Corneal thickness (μm)	573 ± 2.3	547 ± 4.3
Refraction (D)	+3 sph +0.5 cyl @100	Not possible

D diopter, *mm* millimeter, *μm* micrometer, *sph* sphere, *cyl* cylinder, @ axis

Table 16.2 Left eye refraction

Age (yrs)	Follow-up duration	Sph	Cyl	Axis	SE
1.94	1 week	−1.5	+2.5	90	−0.25
2.05	6 weeks	+0.75	+1.00	75	+1.25
3.55	1.6 yrs	−0.50	+0.50	90	−0.25
3.76	1.8 yrs	−0.75	+0.50	90	−0.50
5.08	3.2 yrs	−1.75	+0.75	85	−1.38
6.54	4.6 yrs	−2.50	+0.75	85	−2.13
7.46	5.5 yrs	−5.25	+1.00	90	−4.75
8.21	6.29 yrs (PB IOL explantation)	−6.00	+2.00	90	−5.00
8.36	6.44 yrs (2 months post explantation)	−1.25	+1.25	75	−0.63

PB IOL piggyback IOL, *Yrs* years, *Sph* sphere, *Cyl* cylinder, *SE* spherical equivalent

using Holladay 1 formula). With a +21 D IOL chosen for implantation into the capsular bag, the predicted residual refraction in the spectacle plane was +4.01 D. It was decided to implant a +21 D AcrySof® SN60WF into the capsular bag and a +6 D AcrySof® MA60 into the ciliary sulcus. Biometry had predicted that if a total of +27 D of IOL power was implanted, the predicted refraction immediately post-op would be +0.52 D. Since eye growth is very active at age 2, it was predicted that she would become emmetropic and even mildly myopic within months of surgery. Surgery was uneventful. Both of the IOLs were well centered and we did not prescribe glasses after surgery. At 1 week after surgery, the refraction was −0.25 SE (Table 16.2).

Globe AL measurements were done at 3.8 years of age (1.8 years after cataract surgery). The globe AL was 21.20 and 21.36 mm for right and left eye, respectively. Biometry predicted a refraction of +4.80 D with only a +21.0 D IOL (if the 6 D is explanted). Corneal thickness was 559 ± 1.5 and 582 ± 2.1 μm in the right and left eye, respectively. Refraction at this visit was −0.5 D SE (Table 16.2). At age 5 years, her refraction was −1.50 D SE and she was wearing glasses sparingly. By 8.2 years of age (6.3 years after cataract surgery), her refraction was −5 D SE (wearing glasses part time) and we decided to proceed with piggyback IOL removal. Axial length was 22.34 and 22.75 mm, respectively. Sulcus IOL explantation was uneventful (Fig. 16.1a, b). At the most recent visit, the child was 8.4 years old. Her best-corrected visual acuity was 20/20 in the right eye and 20/25 in the left eye. Her refraction in the pseudophakic left eye was −1.25+1.25X75. Unlike when she was age 2, at age 8 she is comfortable putting on glasses when needed. The IOP was under control (16 and 20 mmHg in the right and left eye, respectively). The very small anterior polar cataract in the fellow right eye cataract was still visually nonsignificant.

Comment For piggyback IOL power calculation, it is recommended that the surgeon decide on the lens power for the anterior, temporary IOL first. The power of this IOL is chosen based on how much refractive change is anticipated during growth and development. A worldwide opinion paper utilizing the Delphi method reported a consensus among surgeons for immediate postoperative target refractions (based on expected refractive change during growth) as follows: age at surgery

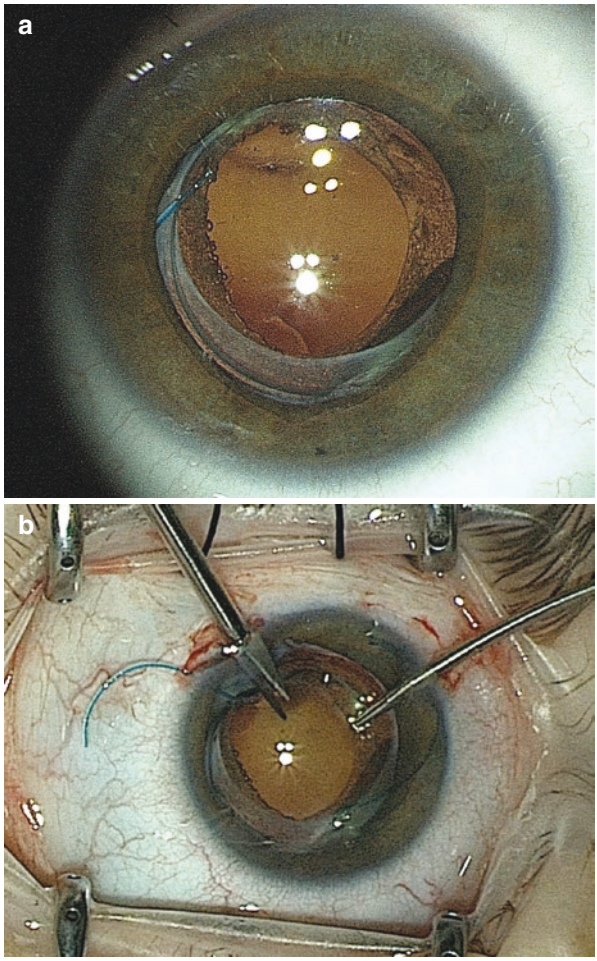

Fig. 16.1 (**a**, **b**) Postoperative view at the time of planned explantation (2-A) and piggyback IOL explantation in progress (2-B)

<6 months, +6–10 D; 6–12 months, +4–6 D; and 1–3 years, +4 D [3]. One method for estimating the needed piggyback IOL power is to multiply the targeted hyperopic postoperative refraction by 1.5. This method was developed for adults to calculate the anterior bag-fixated IOL power. Since the temporary IOL is placed in the ciliary sulcus, a minor adjustment in power is needed if the anterior lens is >+8.5 D. The adjustment of power from capsular bag to sulcus position is reducing by 0.5 D lenses from +8.5 D to +15 D and reducing by 1 D for lens from +15.50 D to +25 D [4]. In the case example, if a single IOL was selected, the immediate postoperative refractive target would have been +4. Based on this, a +6 piggyback IOL was selected for the sulcus-placed lens. Another suggested calculation is to place 20% of the total power for emmetropia in the piggyback IOL [5]. In the above case, 20% of the power needed for emmetropia would have been 5.6 D, which also concurs closely with our choice of +6 D of IOL power.

The second step is to calculate the power of the IOL that will be placed in the capsular bag. For the patient described, the total power needed for emmetropia was +27.76 D. and a +21 D was selected for the capsular bag-fixated IOL to be combined with the +6 selected above. We used the Holladay 1 formula for the calculations. However, the Holladay 2 formula can also be used for selecting a power for the posterior lens, once the sulcus IOL power is determined. In this case, with a +6 D anterior lens, the Holladay 2 formula predicted a need for a +23.5 D IOL for immediate postoperative emmetropia.

The operative technique of piggyback IOL implantation is similar 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. It is very important to remove the ophthalmic viscosurgical device (OVD) from the capsular bag after the first IOL is placed and then fill the ciliary sulcus with additional OVD. A common mistake is to inset the piggyback IOL without first emptying the bag of OVD. Doing that will make the implantation of the second IOL more traumatic for the eye, resulting in more iris trauma and more postoperative inflammation. A single-piece acrylic IOL is used most often for placement within the capsular bag. For sulcus fixation, we recommend a three-piece acrylic IOL.

Interlenticular opacification (ILO), a complication of piggyback IOLs in adults, is avoided in pediatric patients because one of the IOLs is placed in the ciliary sulcus [6]. Interlenticular opacification seems to be related to two IOLs being both implanted in the capsular bag through a small capsulorhexis, with the rhexis margin overlapping the optic edge of the anterior IOL for 360°. Analyses of cases of ILO concluded that the opacification within the interlenticular space is derived from retained/regenerative cortex and pearls from the capsular bag equator growing between the 2 optics within the confined space of the capsular bag. In the case example, an Alcon AcrySof SN60WF® was chosen for the permanent capsule-fixated IOL. Alternatively, the Alcon MA50BM® IOL, which has the majority of its power on the posterior surface, is recommended by some adult surgery websites, such as Warren Hill's doctor-hill.com site [4], when piggyback IOLs are planned in adults. The design of this lens allows for the lowest possible profile at the level of anterior lens capsule.

It has been our hope that visual acuity outcomes would be better after piggyback IOL implantation compared to when uncorrected hyperopia occurs after single IOL placement. Theoretically, slowing increasing myopia after IOL surgery in young children is less amblyogenic than initially high hyperopia that slowly decreases. This seems logical given that hyperopic error is highest in early childhood when amblyopia risk is the highest. This superior benefit would only be realized for those children who comply poorly with wearing spectacles after surgery. In the sample case, the visual acuity outcome has been excellent, despite poor glasses compliance and many cancelled appointments over time. We have no data that indicate this outcome would have been worse had we not used piggyback IOLs. In fact, we have a marked negative selection bias, meaning that we often select this technique in settings of delayed presentation or when poor compliance with glasses or patching is anticipated. That makes any comparison of visual outcomes in piggyback IOLs to

single IOLs meaningless in our cohort. Instead, we have concentrated on a comparative analysis of safety. It would take a randomized trial to adequately compare outcomes. Since the technique is surgically aggressive, we do not often choose piggyback IOLs for children predicted to be excellent at wearing glasses and patching.

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. Boisvert, Beverly, and McClatchey have published their thoughts on choosing piggyback IOL powers [5]. 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. We have not had any difficulty performing any of the planned IOL removals.

The final question remains whether temporary polypseudophakia is recommended for children. Hwang and colleagues reported that compared with the primary single IOL implantation, their temporary piggyback IOL implantation group had higher complications [7]. In the authors' cohort, we have not noted more inflammation, glaucoma, or visual axis opacification compared to age-matched single IOL surgery. Each of our four unplanned early IOL removals came after the technique was used in the first 7 months of life. Now, in our practice, these children are most often left aphakic based on the recommendations of the Infant Aphakia Treatment Study [8]. For surgery on children ages 7 months–5 years, the piggyback technique is employed when compliance with postoperative spectacles is expected to be poor. We believe that this technique can be beneficial in select children. It 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 surgical approach that should be used when needed but avoided in favor of a less traumatic surgery whenever possible. To summarize, infants and toddlers who are anticipated to have difficulty complying with contact lens wear and amblyopia therapy can be considered candidates for piggyback IOL implantation.

Authors do not have any financial interest.

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Chapter 17
Secondary Intraocular Lens Placement

Kamiar Mireskandari

The Infant Aphakia Treatment Study (IATS) reported that infants undergoing cataract surgery prior to 7 months of age had more complications and no added visual benefit when they underwent primary intraocular lens (IOL) implantation [2, 3]. Therefore, most surgeons perform lensectomy, posterior capsulotomy, and anterior vitrectomy in this population, managing resultant refractive error with glasses and/or contact lenses. Ideally, the peripheral capsular bag is preserved in anticipation of these children undergoing secondary IOL implantation when older [1, 4]. The natural healing response in children is for the capsular bag to fuse and lens epithelial cells to proliferate inside the bag with the formation of a Soemmering ring. When implanting an IOL secondarily in children, the surgeon aims to visualize and open the Soemmering ring, aspirate proliferating lens matter, and implant the IOL with optic capture.

Step-by-Step Guide for Surgery

Preoperative Considerations

Refractive targets and preoperative counseling have been discussed in earlier chapters. When the decision is made to leave a child aphakic, the surgeon anticipates eventually returning to the eye to place an IOL secondarily. In surgical preparation,

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consideration must also be given to whether secondary IOL implantation with optic capture would be a difficult or a poor surgical choice for a particular patient.

If not the operating surgeon for the original cataract surgery, one must establish if and how much capsule is present. Most surgeons perform a capsular opening of between 5 and 6 mm, which is ideal for secondary IOLs. However, some surgeons perform a much larger capsulotomy or inadvertently enlarge the capsular opening during vitrectomy. It is important to know if capsular support is suboptimal and alternative options need to be considered.

The surgeon will want to consider whether the correct IOL is in stock for the anticipated surgical plan and refractive target. Some facilities may only stock single-piece IOLs or a limited selection of low- and high-power lenses. Since opening a fibrosed capsular bag may not allow for total "in-the-bag" implantation, a three-piece IOL can be a safer option for sulcus IOL placement with optic capture. In these circumstances, a one-piece IOL is known to have high complications including subluxation, iris chaffing, and uveitis-glaucoma-hyphema (UGH) syndrome. The strength of IOL may not be known prior to surgery because preoperative biometry is often not possible in young children, especially if they have poor vision or nystagmus. Therefore, parents must know that intraoperative biometry will be performed and a high-powered IOL may be required. If the eye has not "grown enough" to be implanted with a standard "off-the-shelf" IOL power, the surgery may be postponed for ordering of the appropriate custom lens.

Informed consent should include the usual explanation of risks and benefits of any intraocular surgery. However, the author finds some features worth mentioning above and beyond the routine discussion. Depending on the age at the time of secondary IOL, the target refraction is calculated based on the future "potential growth of the child/eye" and hence we usually aim for a degree of hyperopia. This, together with the fact that the eye cannot accommodate, means that *all* children will need glasses postoperatively. The author finds this point extremely important as some parents erroneously assume an IOL means no glasses will be required. After all, many have a relative who had cataract surgery at an old age and "never wore glasses again." Glasses must also be worn for protection if the eye is amblyopic. And amblyopia management must continue postoperatively. As with disappointment over ongoing need for glasses, some parents assume that an IOL implant "fixes everything" and may be frustrated to learn that patching therapy is still required.

Operative Steps

The specific steps important to secondary IOL placement in an eye with Soemmering ring are as follows:

1. Two paracenteses are required approximately 150–180° apart to allow the future steps of opening the Soemmering ring and aspirating the contents easier. These should be positioned in a way to allow good hand position when ports are accessed (Fig. 17.1).
2. Viscoelastic can be injected over the iris and under the incisions to prevent vitreous prolapse as required.

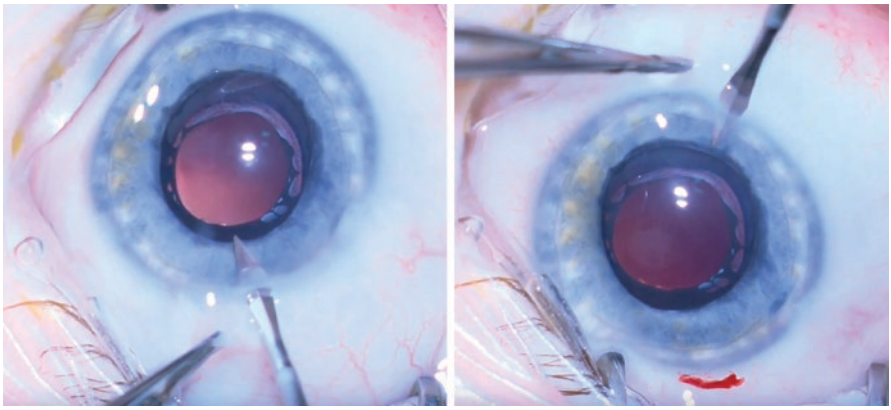

Fig. 17.1 Demonstrating two paracentesis placed approximately 150° apart

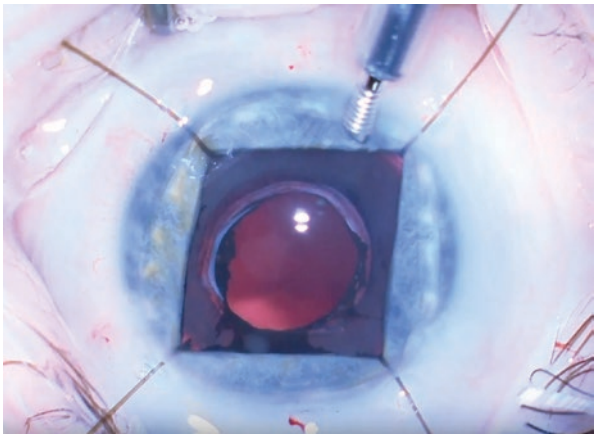

Fig. 17.2 Iris hooks inserted to allow enhanced visualization of Soemmering ring

3. An anterior chamber (AC) maintainer should be inserted to prevent hypotony and AC shallowing during the procedure. Alternatively, the bimanual irrigation handpiece can be employed in conjunction with the aspirator or vitrector.
4. Any iris adhesions to the capsular bag should be divided.
5. A repeat anterior vitrectomy is often required at the beginning of the case since many children develop a degree of vitreous syneresis and prolapse into the AC or pupillary plane. Even if not present at the outset, vitreous prolapse is possible throughout the case and must be dealt with to avoid traction on the retina.
6. Since visualization of the peripheral capsular bag and Soemmering ring is vital for good cortical clean up, placement of iris hooks is an important step in this procedure (Fig. 17.2). The author recommends this even if the pupillary dilation appears adequate at the start of surgery. The accompanying video illustrates several circumstances when this extra dilation proves helpful. Furthermore, since aspiration of the Soemmering content occurs in close

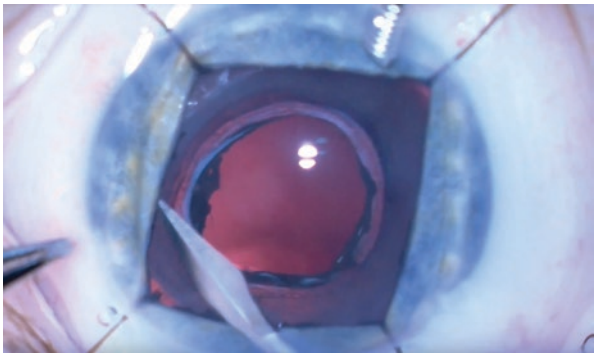

Fig. 17.3 An MVR blade in inserted parallel to the iris plane separates the anterior and posterior capsule

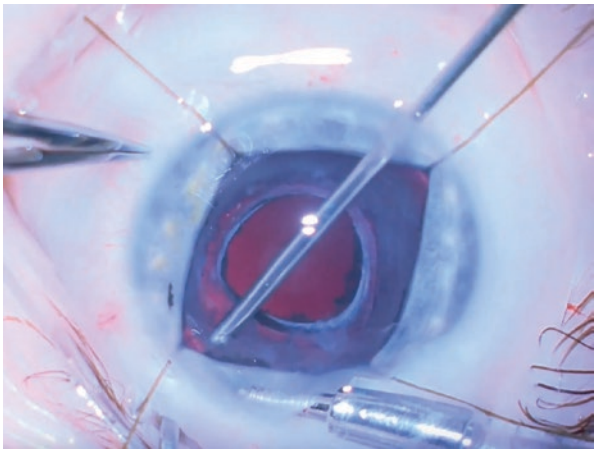

Fig. 17.4 The vitrector is placed on aspirator mode and the port turned sideways. This is used to aspirate the contents of the Soemmering ring

proximity of the iris, hooks provide mechanical support to prevent inadvertent iris grab.

7. An MVR blade is then inserted through one incision and in an orientation parallel to the iris plane. It is used to incise the Soemmering ring just anterior to the fibrous ring where the anterior and posterior capsules have fused (Fig. 17.3). The flat orientation of the blade is important to avoid cutting the "posterior" capsular element of the ring. If this happens, the structural integrity of the central capsule can be compromised. Through this incision, approximately 180° of the Soemmering ring is opened.
8. The AC maintainer is then switched to the opposite paracentesis and the MVR blade is similarly used to open up the remaining Soemmering ring.
9. It is important that the bag is opened 360° to allow easy access to the contents.
10. The vitrector handpiece is then inserted through the paracentesis and a limited vitrectomy is again performed to ensure no vitreous has prolapsed during the preceding maneuvers.
11. The vitrector is then turned off. With the port facing horizontally, the capsular bag is entered and contents removed using aspiration only (Fig. 17.4). The iris hooks are critical to fully visualize and remove the equatorial content.

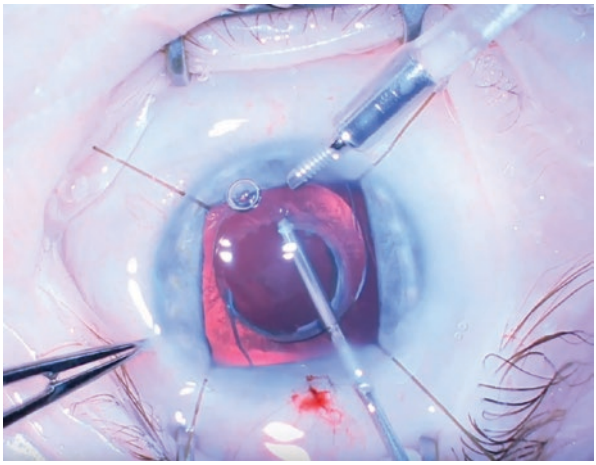

Fig. 17.5 The vitrector handpiece is passed under the IOL and used to enlarge the capsular opening

12. The AC maintainer and the vitrector ports are then switched and the opposite 180° of the bag is emptied. It is important that the central fibrous ring is not interrupted during lens aspiration.
13. The eye is then made ready for IOL insertion. Viscoelastic is injected into the sulcus and the incision enlarged to appropriate size for the IOL inserter.
14. During IOL insertion, care must be taken to ensure the leading haptic does not enter the vitreous cavity and the IOL stays in front of the capsular bag. If the bag is secure enough for the haptic placement, this should be achieved. Similarly, sulcus placement and optic capture will provide a secure IOL placement.
15. The main incision is then secured with sutures
16. With the IOL in front of the capsular bag, it is easy to gauge how much the fibrous ring should be enlarged to posterior capture the optic. The vitrector handpiece is then passed under the IOL and used to enlarge the capsular opening to just 0.5–1.0 mm smaller than the optic diameter (Fig. 17.5).
17. The optic is then posteriorly captured by gently pressing down and flipping the edge of the optic under the capsule.
18. Iris hooks are then removed and Miochol is used to bring down the pupil.
19. Each port is then sutured and checked for a leak. In pediatric eyes, even a needle track for iris hooks may require sutures.

Case

A 3-year-old girl with trisomy 21 was referred for secondary IOL implantation. Her history was significant for right cataract surgery at the age of 6 weeks with lensectomy, posterior capsulotomy, and anterior vitrectomy with preservation of capsular bag peripherally. She had initially worn contact lenses successfully; however, she had recently increased frequency of lens loss and eye rubbing despite good contact

lens fit. Her contact lens and amblyopia management were complicated further by her decreased cooperation with treatment. She also had multiple systemic comorbidities and parents were interested in ways to improve her quality of life by avoiding regular struggle to insert a contact lens in her eye.

Her visual acuity was CSUM in the right eye and CSM in the left. She had normal intraocular pressures and fundus examinations throughout her clinical course. Important positive findings on examination were presence of mild apical corneal scar, vitreous in the pupillary plane, and pupils that dilated to a maximum of 5 mm. Following informed consent surgery was performed as discussed above and demonstrated in the video.

Comment In this case, the decision to proceed with secondary IOL implantation was supported by the patient's increasing difficulty with her contact lens. Age and presence of amblyopia also factored into parental and surgeon decision-making. Surgery was performed using the author's preferred technique of opening the Soemmering ring and placing a three-piece lens in the bag with optic capture. Visualization of the entirety of the capsular bag and ensuring adequate remaining support are key. Use of iris hooks even in cases of good initial dilation can be beneficial.

Conflicts of Interest No financial disclosures or conflicts of interest

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Chapter 18
Intraocular Lens Exchange

Angela Zhu and Courtney L. Kraus

Background

Primary intraocular lens (IOL) implantation after lensectomy was established as an acceptable alternative to aphakia in pediatric cataract surgery after the Infant Aphakia Treatment Study (IATS) showed no significant difference in visual outcomes at age 5 years with those implanted with an IOL in infancy and those left aphakic with contact lens correction [1]. Especially in patients where contact lens use may be challenging for various ophthalmologic, behavioral, or socioeconomic reasons, early IOL implantation may be the preferred alternative to aphakia in facilitating compliance with refractive amblyopia management. While optimal targets for postoperative refraction after cataract extraction have been discussed in previous chapters, there is a known increase in axial elongation acutely after pseudophakia resulting in varying degrees of myopic shift reported in literature [2, 3]. Given the difficulty in predicting the precise amount of myopic shift after primary IOL implantation, it is not uncommon for these children to have significant anisometropia, especially in cases of unilateral cataracts. As surgical techniques and medical technology have advanced, attention has shifted from simply determining when pediatric cataract surgery should be performed and more toward optimizing refractive outcomes. A retrospective case series of 15 eyes undergoing IOL exchange for refractive indications demonstrated successful visual rehabilitation with predictable

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targeted postoperative refractions after this technique [4]. IOL exchange has therefore become an increasingly popular option for managing severe anisometropia and associated intolerable aniseikonia in pseudophakic children, which can further facilitate amblyopia management.

Case 1

An 8-year-old girl with history of infantile unilateral cataract presented for consideration of refractive options for aniseikonia. She had initially undergone lensectomy with posterior capsulotomy, anterior vitrectomy, and primary IOL implantation at age 1.2 months, with placement of an intracapsular MA60MA IOL with initial targeted postoperative refraction of +6.00 D. She had initially worn contact lenses successfully and was compliant with penalizing amblyopia therapy. At time of presentation, her best corrected visual acuity was 20/30 in this eye with a manifest refraction of −8.50 +0.25 ×170 (spherical equivalent −8.5 D) due to axial length elongation of 6.3 mm; visual acuity was 20/20 in the contralateral, unaffected eye with a manifest refraction of −1.75 +0.25 ×085 (spherical equivalent −1.50 D). However, she had grown increasingly intolerant to contact lens wear with frequent eye rubbing and suffered from significant aniseikonia with spectacle wear, so her parents were interested in surgical options to correct the high myopic shift in this eye given her excellent corrected visual acuity.

Clinical examination of the affected eye was significant for a three-piece IOL placed within the capsular bag, with fusion of the peripheral anterior and posterior capsules but a clear visual axis through the anterior/posterior capsulotomies. She had normal intraocular pressures and fundus examination throughout her clinical course. After informed consent was obtained, the patient and parents elected for intracapsular intraocular lens exchange, with surgery performed as discussed below. A replacement three-piece +10.5 D IOL was placed in the ciliary sulcus using posterior optic capture with a target refraction of −3.00 D. There were no intraoperative or postoperative complications. Manifest refraction at 1 month postoperatively was −4.50 +1.50 ×015 (spherical equivalent −3.75 D) and at 6 months postoperatively was −4.00 +0.75 ×040 (spherical equivalent −3.50 D), and the patient was able to tolerate spectacle wear at this time.

Comment In this case, the patient's decision to proceed with IOL exchange was a largely refractive decision that was supported by her excellent visual potential due to compliance with amblyopia therapy, relatively young age, increasing contact lens intolerance, and aniseikonia with spectacles. There were no instigating factors such as IOL dislocation or subluxation that would necessitate more prompt IOL removal; an intracapsular three-piece IOL was easily exchanged for another three-piece IOL placed in the ciliary sulcus with posterior optic capture following the steps as outlined below.

Step-by-Step Guide for Surgery

Preoperative Considerations

Specifics of the original cataract surgery are crucial in the planning of IOL exchange. If the original cataract surgery was performed by another surgeon, it is best to obtain operative note records in order to know type and strength of IOL implanted. Careful slit lamp biomicroscopy examination and/or high-resolution ultrasound biomicroscopy is incredibly helpful in determining preoperative positioning of the haptics and how much capsular bag support remains (both anterior and posterior capsule). Furthermore, if the original cataract surgery was a significant amount of time prior to planned IOL exchange, the peripheral capsular bag may be fibrotic or phimotic with the anterior and posterior capsules tightly fused; this can increase the difficulty of completing the IOL extraction without damaging the remaining capsular bag. Presence of a Soemmering ring can further complicate IOL exchange and make intracapsular placement of a new IOL very difficult. If any doubts exist regarding the integrity of the capsular bag for intracapsular IOL placement, it may be safer to plan for sulcus placement of a three-piece IOL with posterior optic capture, if possible, for refractive stabilization.

Since IOL exchange is often done for refractive purposes assuming there are no abnormalities with the original IOL (e.g., IOL dislocation or subluxation) that would necessitate more prompt IOL removal, special consideration must be given to the refractive target of the new implant. The targeted postoperative refractive can vary depending on the patient's age, refractive/visual status of the other eye, and indication for IOL exchange (e.g., debilitating aniseikonia vs sizable myopic shift). For example, a younger child within the amblyogenic age may benefit from a target closer to emmetropia or to the contralateral eye refraction, but one must account for whether further axial elongation may still occur and how much anisometropia can be tolerated. Biometry may also be difficult or imprecise in these cases, especially if unilateral pseudophakia or other ocular comorbidities triggered an axial myopic shift. In these cases, intraoperative biometry may be used to confirm appropriate IOL selection.

During any informed consent discussion of the risks, benefits, and alternatives to IOL exchange surgery, the necessity of postoperative refractive correction must be mentioned. As one primary indication for IOL exchange surgery is intolerance to contact lenses and/or aniseikonia or significant anisometropia with glasses, many patients and parents may believe that no glasses or contact lenses will be necessary postoperatively. Furthermore, it must be mentioned that amblyopia management will continue postoperatively, but that the goals of IOL exchange are often more to facilitate tolerance of refractive correction and amblyopia therapy. Depending on biometry and patient factors, the patient may be a candidate for other refractive options, including piggyback IOL or laser refractive surgery, which can also be discussed with the patient and family (as described in other chapters).

Operative Steps

The specific steps important to intracapsular exchange of an acrylic IOL implant are as follows:

1. One or two paracentesis incisions (approximately 120–180° apart) should be positioned in a way to allow access to both IOL haptics with good hand position (each generally 60–90° from a haptic).
2. Viscoelastic should first be injected into the anterior chamber to maintain the anterior chamber and prevent any IOL or vitreous prolapse.
3. Since visualization of the peripheral capsular bag and positioning of the haptics are crucial for IOL removal while preserving the integrity of the capsular bag, consider placement of iris hooks if the pupil is poorly dilated. An iris manipulating instrument (e.g., Kuglen hook) may also be used to temporarily visualize the peripheral capsular bag and haptic placement.
4. Using viscoelastic on a 27G needle, slide the needle with bevel side up underneath the anterior lens capsule edge at the haptic-optic junctions. Inject viscoelastic gradually to gently dissect the anterior capsular edge off of the IOL. Switching to viscoelastic on a blunt cannula, attempt to further inject viscoelastic both anterior and posterior to the haptic in order to dissect the capsular bag away from the haptic.
5. Using a lens manipulating instrument (e.g., Kuglen or Sinskey hook) placed at the haptic-optic junction, gently rotate each haptic out of the capsular bag and into the anterior chamber. Inject viscoelastic anterior/posterior to the IOL centrally as well as further peripherally within the capsular bag to release the haptics as needed during this process. If the initial IOL was placed in the ciliary sulcus, copious viscoelastic can be used to inflate the sulcus to prolapse the IOL into the anterior chamber for extraction.
6. A clear corneal incision should be created approximately 90–120° away from a paracentesis, generally close to the width of the radius of the prior IOL (usually 2.4–2.8 mm depending on how flexible the IOL material is) (Fig. 18.1). If a rigid IOL that cannot be cut with intraocular scissors was initially placed, consider a large superior scleral tunnel incision in order to remove the IOL in one piece while also minimizing incision-induced astigmatism.
7. Using lens-holding intraocular forceps (e.g., GRIESHABER® Maxgrip® forceps, Alcon, USA) through a paracentesis and lens-cutting intraocular scissors (e.g., Packer/Chang IOL cutters, MicroSurgical Technology, USA) through the corneal incision, cut the IOL into two halves within the anterior chamber (Fig. 18.2).
8. Gently extract the two halves of the IOL through the main incision using the lens-holding forceps and/or fine-tipped forceps, being careful to rotate each half through the incision following the curve of the IOL optic/haptic. Avoid grabbing the end of the haptic in order to prevent pieces from breaking prior to extraction of the entire IOL (Fig. 18.3).

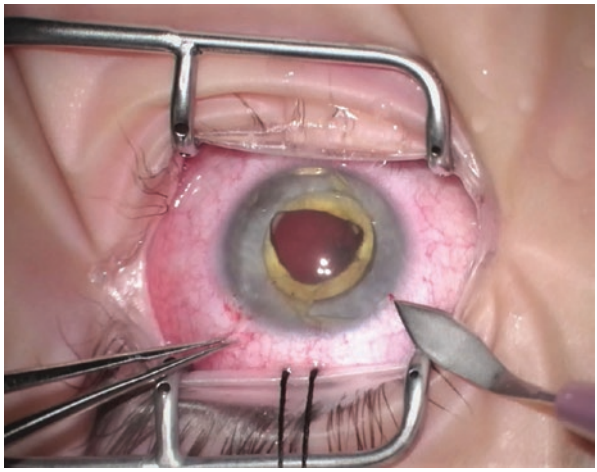

Fig. 18.1 Creation of a clear corneal incision approximately 120° away from a paracentesis incision after the IOL has been prolapsed into the anterior chamber

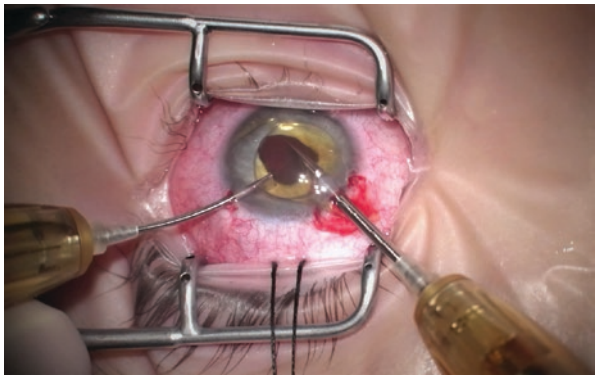

Fig. 18.2 Cutting of the IOL optic into two halves (each containing a haptic) using lens-holding intraocular forceps in the left hand through a paracentesis and lens-cutting intraocular scissors in the right hand through the corneal incision

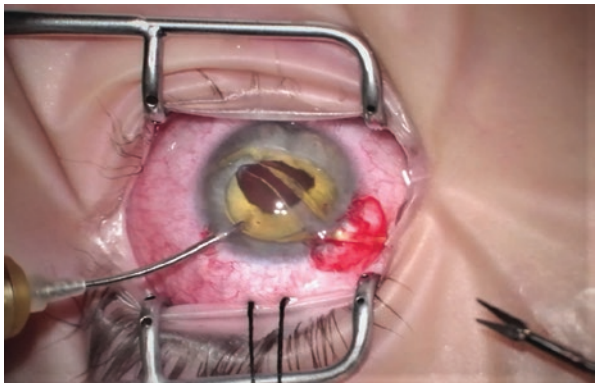

Fig. 18.3 Prolapse of IOL haptic out of main corneal incision using lens-holding intraocular forceps for subsequent extraction of each half of the IOL; at this point, fine-tipped IOL forceps can be used to grasp the optic-haptic junction external to the corneal incision to rotate each half through the incision following the curve of the IOL optic/haptic

9. At this point, inspect the globe for any signs of vitreous prolapse. Perform anterior vitrectomy if needed.
10. The globe is then prepared for IOL insertion by injecting viscoelastic into the sulcus. The incision may need to be enlarged to appropriate size for the IOL inserter.
11. During IOL insertion, care must be taken to ensure the leading haptic does not enter the vitreous cavity and the IOL stays in front of the capsular bag. If the peripheral capsular bag is secure enough for the haptic placement, intracapsular placement of the new IOL can be achieved. However, sulcus IOL placement with posterior optic capture may be more secure.
12. Further irrigation/aspiration and/or vitrectomy may be performed to remove residual viscoelastic or vitreous as needed.
13. All incisions (both paracentesis and larger corneal incisions) should be sutured and checked for a leak. In pediatric eyes, even small paracentesis incisions require sutures.

Postoperative Considerations

In the initial postoperative period, it again should be stressed that refractive correction and amblyopia management must resume as soon as possible. While the refractive outcomes may not be stabilized until a few weeks postoperatively, the overall recovery period should be shorter than the original cataract surgery, so it will be possible to resume spectacle wear with an updated refraction fairly promptly. While children are often highly immunogenic with profound inflammatory responses to any surgery, the postoperative inflammation is again expected to be overall less than for many other intraocular surgeries. However, careful follow-up evaluation for rebound iridocyclitis and cystoid macular edema should be performed if any decline in vision does occur. There has also been evidence showing longer-term endothelial cell loss and subsequent corneal decompensation (>5 years) after IOL reposition or exchange surgeries in pediatric patients [5]. While this risk can be mitigated with judicious use of dispersive viscoelastic intraoperatively, it is worthwhile to discuss this as a risk with the family and continue following these patients to ensure no late corneal complications occur or require further surgical intervention.

Financial Disclosures There are no financial disclosures or conflicts of interest.

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Part IV
Special Considerations in IOL Implantation

Chapter 19
IOL Implantation in the Absence of Capsular Support

Secondary IOL After Initial Aphakia: Ciliary Sulcus Fixation and IOL Placement in Absence of Capsular Support

Jan Tjeerd de Faber and Martha Tjon-Fo-Sang

The use of an intraocular lens (IOL) in pediatric cataract surgery became common practice in the 1990s. Refined surgical techniques, technology, and IOL material advancements together decreased complication rates making IOL placement a more viable option. However, in very young children, primary implantation of an IOL remains controversial because of the ongoing growth of the eye, increased rate of complications, adverse events, and additional associated surgeries [1, 2]. Thus, the clinical decision is often made to keep the eye aphakic after lensectomy and to correct the aphakia with either contact lenses or spectacles. Secondary implantation can then ideally be considered after the age of 2 or at any moment when contact lens intolerance occurs. In patients with adequate capsular support, the IOL may be placed in the bag or the sulcus, in front of the remnants of the anterior and posterior capsule. In cases of trauma or inadequate capsular or zonular support (e.g., Marfan's syndrome), IOL implantation outside the bag may be necessary.

Artisan® Iris-Fixated Anterior Chamber IOL

When there is inadequate capsular support for an IOL, options include the use of the Artisan® (Ophtec, Groningen, the Netherlands) iris-fixated IOL in the anterior chamber (AC) [3] and scleral-sutured or scleral-glued posterior chamber IOLs. The Artisan® iris-fixated IOL was first designed by Jan Worst in 1979 based on a

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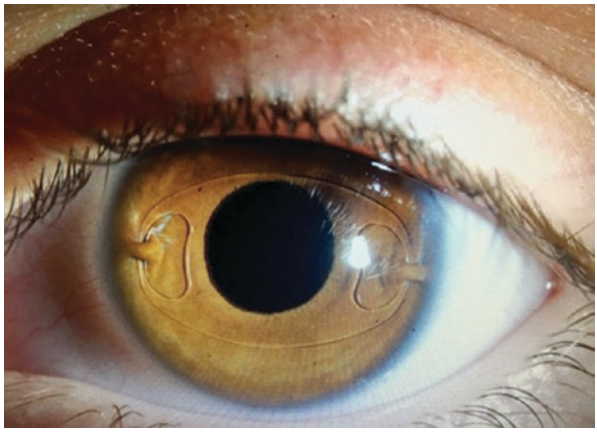

Fig. 19.1 Artisan Aphakic IOL with good centration and adequate enclavation of nasal and temporal peripheral iris 12 years after implantation

concept that the peripheral iris stroma is almost immobile and therefore could serve as a fixing point in the eye. The haptics of the Artisan® IOL are shaped like a ring with an incision site. The lens is enclavated by passing the iris through this break in the haptics. Attached to the peripheral iris, the lens is far away from the angle of the anterior chamber and the corneal endothelium, reducing problems associated with the chamber angle and limiting contact with endothelial cells. A major advantage of the Artisan® IOL over a scleral-sutured or scleral-glued PC IOL is that it can be easily and safely explanted or exchanged should there be a large myopic shift due to excessive axial elongation (Fig. 19.1).

Before implanting an Artisan® IOL, the surgeon should take a moment and anticipate the enclavation sites of the haptics. This assists the surgeon in selecting the site of the paracenteses, which in these cases are made obliquely to allow access to the peripheral iris. A 5.6 mm main incision is created, which most often is created superiorly and tunneled as the IOL is rigid and unable to be folded to allow passage through a smaller incision. The IOL is inserted through the main incision and while one hand stabilizes the IOL over the center of the pupil, the other uses enclavation forceps or needle to pass the midperipheral iris tissue through the slit of the haptics, making sure that an adequate amount of iris tissue is enclavated (Fig. 19.2).

It is the authors' preference to fixate the lens on the anterior surface of the iris because this enables easier monitoring of the IOL's stability and fixation during slit lamp examination. Additionally, if the IOL was to dislocate it is easier to locate and retrieve in the anterior chamber than in the vitreous. Some surgeons prefer to fixate the Artisan® on the posterior surface of the iris arguing that the IOL is then physically farther from the cornea, creating less stress on the corneal endothelium. If fixating on the anterior surface of the iris, the convex side of the lens must face up and the haptics angulate posteriorly to prevent pseudophakic glaucoma. When using it on the posterior surface of the iris, the convex side should be facing the vitreous (Fig. 19.3). This is only feasible when a full vitrectomy has been performed, because a posterior fixation will interfere with the vitreous base and the peripheral retina risking retinal tears or detachments.

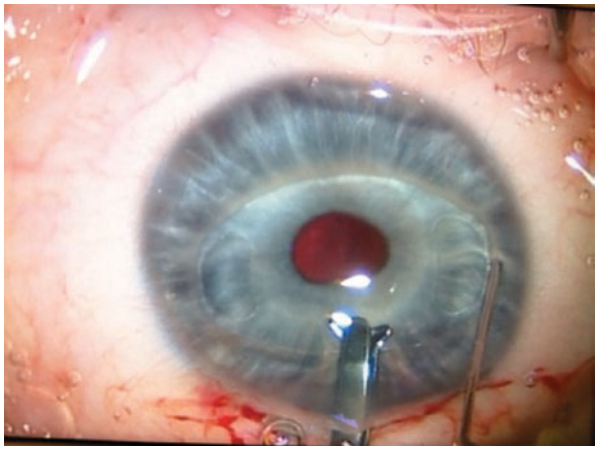

Fig. 19.2 While one hand holds the Artisan over the central pupil, the other hand enclavates iris tissue between the haptics. One should select the desired spot on the iris before starting the enclavation movement

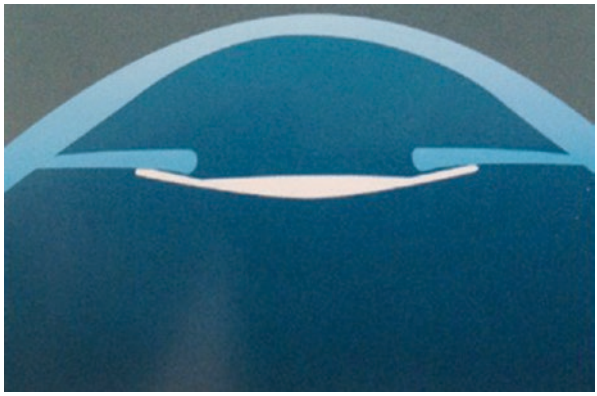

Fig. 19.3 In posterior iris fixation, concave angulation facilitates enclavation of posterior iris tissue

In extremely difficult cases where the iris tissue is stiff and so flat that grasping it for enclavation presents a significant challenge, the manufacturer designed the VacuFix enclavation system. This consists of two handles, which are connected to the phaco-machine, and when the VacuFix contacts the iris, the suction allows for an easier grasp of iris tissue and lift of the fold of iris through the slot of the haptic. When the VacuFix system is not available the enclavation needle attached to a 1 or 3 cc syringe with extension tubing (Fig. 19.4) can be employed. During enclavation, an assistant is instructed to pull back on the plunger of the syringe, creating a vacuum, enabling the surgeon to aspirate strands of iris tissue to facilitate enclavation.

For all cases where an iris claw lens is used, an iridotomy or iridectomy is essential in order to prevent pseudophakic pupillary block glaucoma, especially in eyes that have only had a core vitrectomy. Iridotomies can be performed during surgery or with a neodymium-yttrium-aluminum-garnet (Nd:YAG) laser prior to surgery.

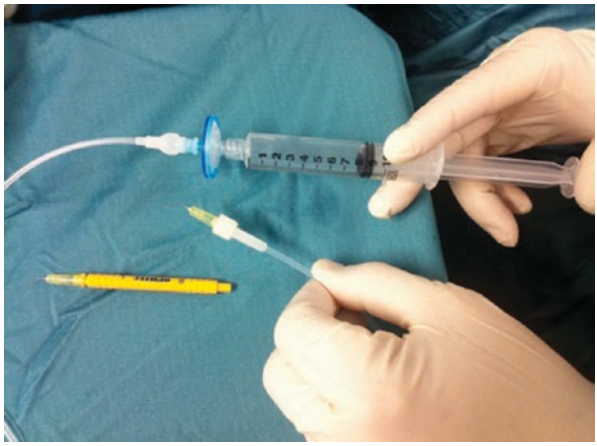

Fig. 19.4 Connecting the enclavation needle with an extension tube to a syringe will add a suction function to the needle to facilitate enclavation

Case 1

A 12-year-old male presented 1 week following a trauma in the left eye related to a firework injury. Presenting visual acuity (VA) was 20/800 in the affected eye and 20/20 in the unaffected. A red reflex was visualized, but view to the fundus was difficult; B-scan showed an attached retina with some blood in the vitreous cavity. IOP was around 20 mmHg. Due to the blast the entire lens had luxated into the anterior chamber. Additionally, the iris showed iridodialysis over 3'clock hours (Fig. 19.5). There were no corneal lacerations or scleral breaks due to trauma. Medical history of the child was unremarkable.

In this case, initial management consisted of aspiration of the lens followed by an anterior vitrectomy. Subsequently, the pupil was contracted with an intracameral miotic agent and the AC again deepened with a cohesive viscoelastic device. The ruptured iris was reattached with two Prolene 10.0 sutures to the iris root in the angle of the anterior chamber. Despite iris trauma, an Artisan® lens was able to be enclavated horizontally.

Following the procedure, the child did well with a postoperative month one VA of 20/200 due to a macular scar caused by a choroidal rupture. No further complications or surgeries were required.

Comment The Artisan® IOL can be successfully implanted in cases with limited available iris tissue, even after trauma. The Artisan® needs very little support as long as there is a large enough iris bite through the fibrous strands of the iris and the iris is not tremulous. After a trauma the iris can be damaged or a traumatic mydriasis can make enclavation challenging but not impossible as seen in Fig. 19.6. It is also possible to enclavate the lens during primary lensectomy, allowing immediate best possible refractive correction and potential acuity.

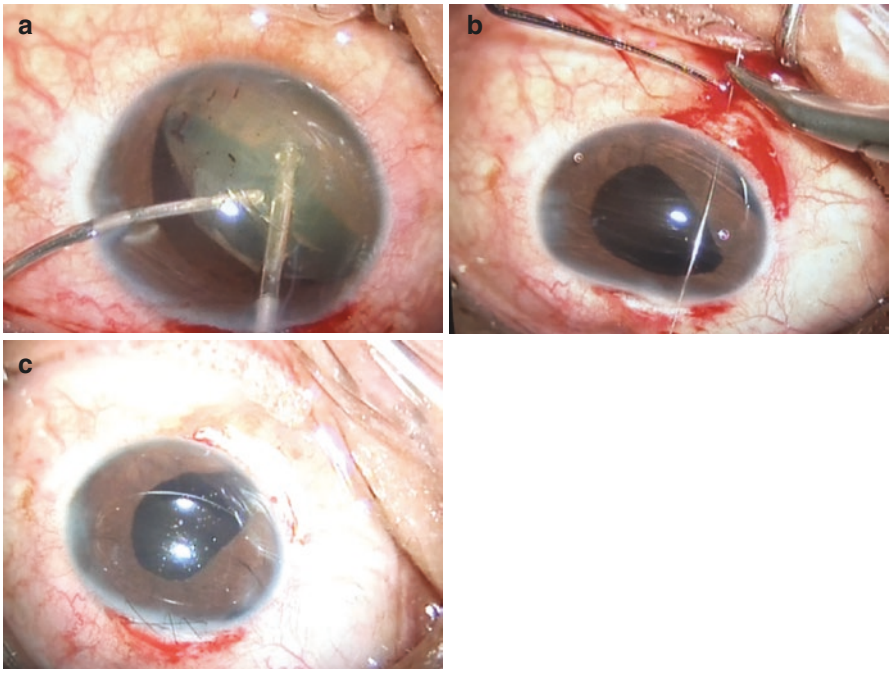

Fig. 19.5 The crystalline lens is luxated into the anterior chamber following blast injury with fireworks. It is aspirated (**a**). Iris dialysis of 3'clock hours is repaired using 2 10-0 Prolene sutures to the iris root (**b**). An iris claw lens is enclavated to the remaining iris and the wound closed with running 10-0 nylon (**c**)

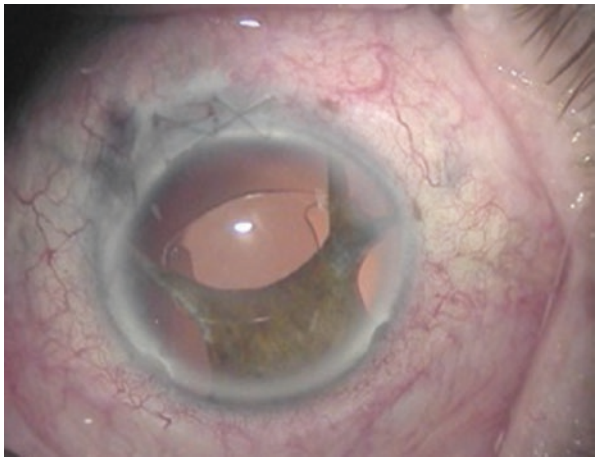

Fig. 19.6 Traumatic cataract and mydriasis after firework injury follow-up picture after 8 years, note how little enclavation room there is used to bridge the traumatic wide pupil

Artisan® IOL in Cases of Subluxated Lenses Due to Marfan's Disease

In pediatric cases with lens subluxation the decision for surgery is weighed against the risk of amblyopia. When the lenticular astigmatism can no longer be corrected by spectacles and the VA regresses despite occlusion therapy, the risk of amblyopia is high and surgery may be recommended. At this stage, patients are most often older than two and IOL implantation can be pursued instead of the use of aphakic contact lenses postoperatively.

During surgery, the amount of zonular support can be assessed by using a blunt instrument, such as a sweep, to press down on the anterior capsule. When there is lack of zonular support despite the use of a viscoelastic device, radial folds will appear on the anterior capsule. This is pathognomonic for weak zonular fibers (Fig. 19.7).

Opening the anterior capsule is more difficult due to the loose zonular fibers. Inserting an iris or capsular retractor in the rhexis helps to pull the lens toward the loose zonular fibers, stabilizing the lens and facilitating a properly sized capsulorhexis so the lens material can safely be aspirated (Fig. 19.8).

Once the bag is empty it can be carefully removed using smooth forceps ensuring the breakage of the intact zonular fibers without rupturing the capsule or disrupting the anterior hyaloid membrane. In the event of vitreous loss, an anterior vitrectomy should be performed; however, in our series we were able to spare the anterior hyaloid membrane in 72% of the cases, precluding the need for an anterior vitrectomy (Fig. 19.9).

Next, the pupil is contracted with a miotic agent (e.g., pilocarpine or Miochol) and an Artisan® IOL can be inserted through the main wound and placed horizontally on the iris.

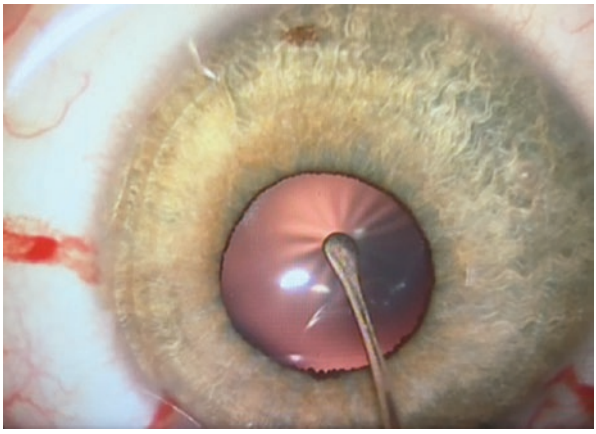

Fig. 19.7 Pushing with a blunt instrument on the anterior capsule shows radial folds indicating weak zonular fibers in a 3-year-old child with lens subluxation due to Marfan's disease

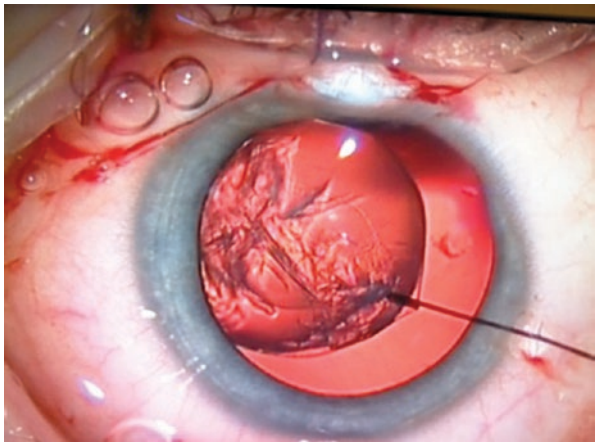

Fig. 19.8 An iris hook is used to pull the rhexis more to the center of the pupil. This facilitates the aspiration of lens cortex behind the iris

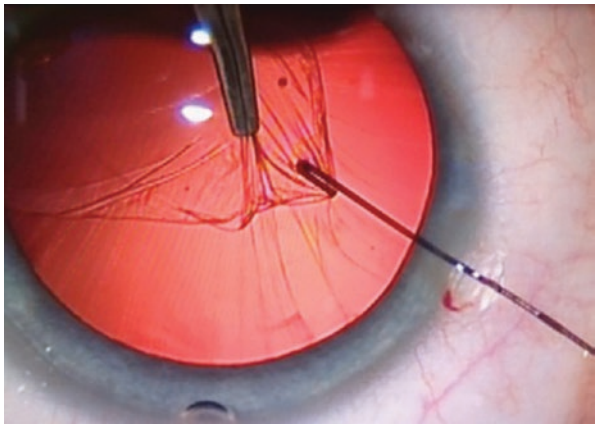

Fig. 19.9 With a forceps the rhexis of the bag is gently pulled to the wound giving the remaining zonular fibers time to break without rupturing the hyaloid membrane. The intact bag is pulled out of the eye through the main wound

Long-Term Results

There are numerous studies on the long-term effect of the Artisan® Aphakia IOL on the endothelial cell density (ECD) in children. In a retrospective study, it was found that the mean endothelial cell counts after 10 years of follow-up was comparable to mean normal ECD in a same age group of children reported in literature [4]. Our own patient population who have received Artisan® IOLs more than 25 years ago maintain excellent ECD counts and are now requesting the same procedure for their own children with Marfan's disease (Fig. 19.10).

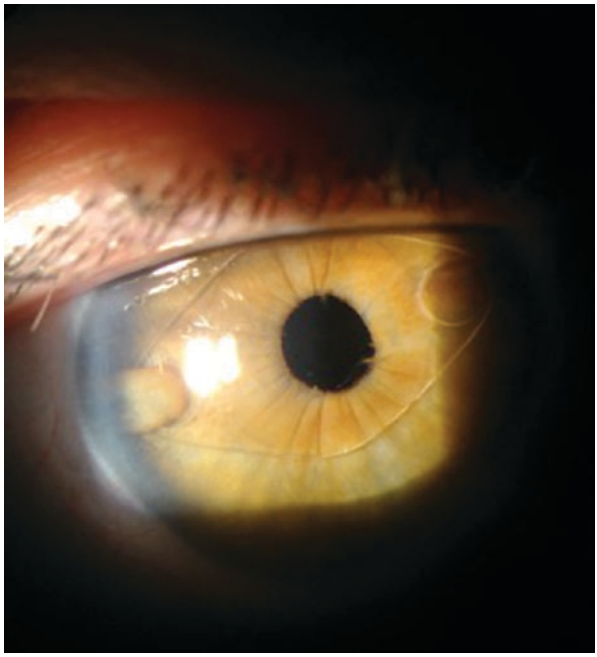

Fig. 19.10 25 years after implantation of first type of Artisan Aphakic IOL

Complications of Artisan IOL

Complications of the Artisan® aphakia IOL include those that occur in the immediate perioperative period, as well as longer postoperative period. Decentration can occur early or late following surgery. Careful enclavation is the key to prevent decentration of the IOL. Haptics may dislocate with blunt trauma and require urgent re-enclavation. Fortunately in approximately 95% of the cases only one haptic is involved and the patient almost universally notices an immediate drop in VA as well as discomfort when the cornea is intermittently touched by the loose haptic.

Hyphemas occur intraoperatively or in the immediate postoperative period from both manual separation of adhesions before implantation or inadvertent tearing of the iris. Pigmentary dispersion can result from multiple attempts to grasp the iris. If the iridotomy or iridectomy is inadequate, pupillary block leads to acute glaucoma. If this occurs, additional peripheral iridectomies with a YAG laser should be attempted. When dealing with a shallow anterior chamber, an optimal location to place the iridectomy would be just between the claw and the optic where the iris is stretched and not in contact with corneal endothelium.

Angle Supported Anterior Chamber IOLs (ACIOL)

The placement of ACIOL haptics in the angle of the anterior chamber (Fig. 19.11) creates a risk of trabecular meshwork damage, angle fibrosis, and even peripheral anterior synechiae formation [5]. This creates an obstruction to aqueous outflow, whereby IOP can increase resulting in secondary glaucoma. Therefore, it is the authors' recommendation that angle-supported anterior chamber lenses not be used as a secondary implant in children.

Posterior Chamber Intraocular Lenses

Iris-Sutured Lenses

Several surgeons have described techniques to suture posterior chamber IOLs to the iris. However, one of the reported complications is chafing of the iris. Placing sutures within iris tissue holding an IOL can trigger chronic inflammation because

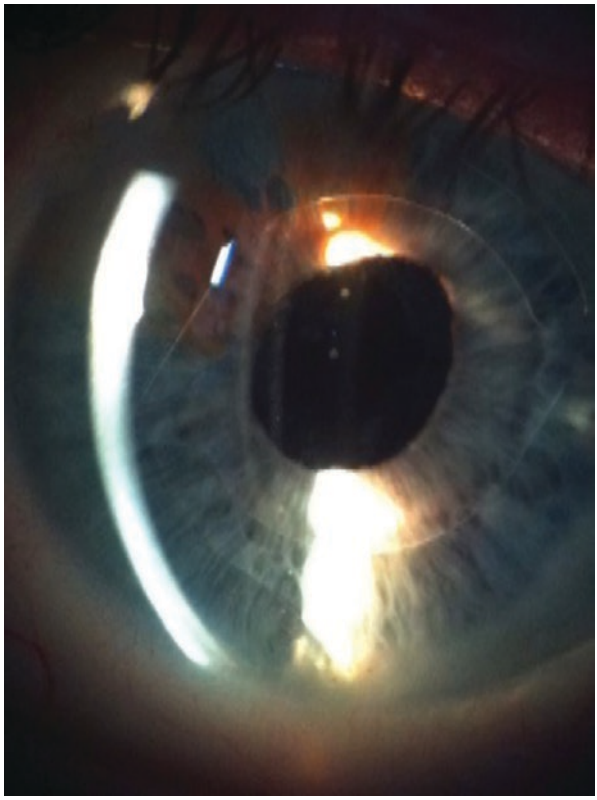

Fig. 19.11 Anterior chamber angle-supported IOL, note slightly distorted pupil and loss of iris pigment

of the mobility of iris tissue. The location and the tightness of the sutures are two important considerations that can increase the likelihood of chafing. The central iris is the most mobile – the more central the suture placement, the more inflammation is to be expected. It can also create an irregular pupil with peaking at the sites of suturing. Excessively tight sutures or excessively large bites of iris can cause peaking of the pupil or bunching of the iris, resulting in increased contact of iris and IOL and, thus, increased chafing.

Scleral-Sutured Lenses

Scleral-sutured PC IOLs are also an option to correct aphakia in cases with absent capsular support. However, IOL tilt or displacement into the anterior vitreous has been reported as a result of suture loosening or breakage. In the past, polypropylene sutures have been used, but these are prone to biodegradation after 7–10 years [6, 7]. Gore-Tex sutures seem to have a longer lifetime and are therefore now widely used [8].

Fibrin-Glued PC IOL

This is a technique using fibrin glue to embed the flexible prolene haptics of a multipiece IOL under two scleral flaps. After the scleral flaps have been made, the haptics of the PC IOL are externalized and attached to the scleral bed with glue. Then, the flap and the conjunctiva are also closed with fibrin glue. Adopters of this technique argue that the fibrin glue provides good flap closure and IOL centration and stability without suture-related complications [9]. We have no experience with this technique in children.

Flanged Intrascleral PC IOL Fixation

In 2017 Yamane [10] described fixation of a PC IOL by making a flange at the end of the haptic. After two scleral tunnels have been made, the prolene haptics of a multipiece IOL are externalized and cauterized to make a flange of the haptics (Fig. 19.12). The flanges of the haptics are subsequently pushed back where they become embedded in the scleral tunnel due to their increased thickness. This allows fixation and centration of the PC IOL without the use of glue or sutures. This procedure can only be performed in an eye that has undergone vitrectomy combined with preventive laser coagulation of the peripheral retina.

Implanting an IOL has become the standard of care in many cases of pediatric cataract surgery. Although there have been many technical advancements since Sir Harold Ridley first implanted his lens, we still prefer to place the lens as he did, in

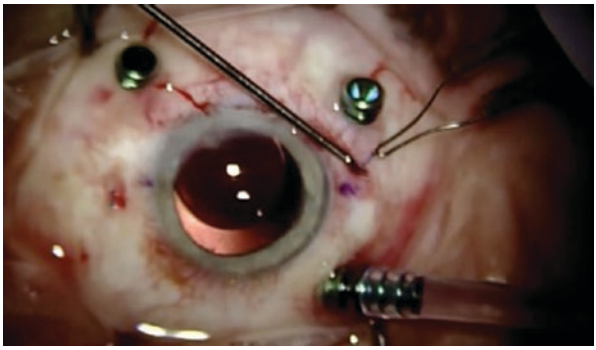

Fig. 19.12 The tunneled prolene haptic is heated at the end. This will create a flange which will hold the IOL at the entrance of the tunnels on opposing sides of the eye

the capsular bag. Unfortunately, it is not always possible to follow this approach due to either preexisting conditions or surgical difficulties. Many procedures have been described to allow IOL implantation in an eye without capsular support. Each of these techniques differs with regard to technical difficulty, potential postoperative problems, and long-term complications. Improvements to implant design and material development have made anterior chamber lenses a more attractive and feasible alternative.

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Chapter 20
IOL Placement in the Setting of Trauma

Jennifer Dudney Davidson and Sydney Michelle Mohr

It has been well established that the least amount of time between injury and surgical repair results in better postoperative outcomes for adults following ocular trauma [1]. The same is true in children. Traumatic cataracts in children result from both perforating eye injuries "open globe injuries" and non-perforating "blunt trauma" [2]. Ruptured globes in children, like adults, are repaired as soon as possible following the injury. When lens material extends through a ruptured anterior capsule, a surgeon may choose to wait for inflammation to decrease before proceeding with cataract extraction. However, for a child in the amblyogenic period, surgery should be performed as soon as safely possible and amblyopia treatment initiated immediately after. Trauma that involves the capsular support system of the lens requires additional techniques and appropriate lens selection to ensure stability of the intraocular lens (IOL) for the lifetime of the child.

Preoperative Assessment and Surgical Planning

Much like adults, when a child presents following trauma, a brief examination seeks to determine if a penetrating injury, foreign body, cataract, or retinal damage exists. Often exams in children are limited by cooperation and may require sedation in an emergency room setting. When there is a corneal or scleral penetration, the eye

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should be patched with a hard shield and standard ruptured globe protocol initiated. If a cataract is noted at the time of the injury, one will need to weigh the pros and cons of lensectomy at the time of primary globe repair.

Lens Selection and Timing of Lensectomy

A salient consideration is what type of lens should be selected based on whether or not the anterior capsule, posterior capsule, or zonule have been injured. When only the anterior capsule has been violated, with little lens material in the anterior chamber, the best option is a stepwise approach. First, close any corneal and scleral lacerations, place intracameral antibiotics and steroids, then allow the eye to heal. Most surgeons wait a minimum of 2–6 weeks before removing corneal sutures and cataract extraction [2]. This time period will give the eye time to quiet and provides a more predictable surgical environment.

If there is a large opening in the anterior capsule, lens material may need to be removed in order to close the wound. Even in this unique situation, a surgeon should prepare for complete lens removal and intraocular lens placement at a later date. Delaying surgery allows for better visualization following corneal repair and management of inflammation. In the instance of an unaffected lens capsule and zonule, a single-piece acrylic lens should be preferentially placed in the capsular bag. If there is any concern for possible zonular weakness, a three-piece lens in the capsular bag will allow for securing the lens to the sclera if necessary, in the future. Sulcus lenses should always be selected in advance in case capsular support is found to be inadequate for an in-the-bag lens placement. Three-piece acrylic lenses or a Rayner (Rayner Intraocular Lenses Limited, USA) single-piece acrylic lens is a good option for the sulcus (with or without enclavation) [2].

Biometry Measurements and Lens Calculations

Biometry measurements may be performed in older children while awake; however, most small children require intraoperative biometry. Obtaining the most accurate lens calculations is another reason it is best for cataract surgery to follow initial ruptured globe repair. Biometry measurements for the contralateral eye may be helpful for lens selection when there has been extensive globe trauma causing keratometry and axial length measurements to be unreliable. Lens power selection in the setting of trauma should follow the same recommendations for inherited cataracts in children to minimize late myopia [2].

Special Techniques

Secondary surgery after globe repair often involves cataract extraction, placement of an intraocular lens, possible posterior capsulotomy, and then removal of any corneal sutures. Visualization may be limited by a corneal scar or edema, but management of inflammation and intraocular pressure between the time of trauma and secondary cataract extraction will help to clear the cornea.

Limbal corneal incisions should be strategically placed for best visualization and easiest access to the involved lens or capsular violation. Bimanual techniques with an irrigating handpiece or an anterior chamber maintainer may be used. When there is extensive trauma, the anterior chamber maintainer allows for both hands to be used and helps to prevent chamber collapse when removing an instrument from the eye. Although an aspirating handpiece may be appropriate for a standard cataract extraction in a child, the primary use of the vitrector is prudent in case any vitreous is encountered in the anterior chamber. Before the insertion of irrigation, viscoelastic and trypan blue help with visualization of the anterior capsule. Lens material should first be removed from the anterior chamber. If the capsular opening is small, it may be enlarged with a vitrectorhexis or manual technique. The lens material should carefully be removed from within the capsular bag, seeking to maintain the integrity of the posterior capsule with the least amount of zonular traction. Once the lens nucleus and cortex have been completely removed, the surgeon may better assess the integrity of the posterior capsule and zonule prior to inserting a lens.

Lens Location

If possible, a lens should be placed in the capsular bag. When there is zonular absence or weakness, capsular tension rings and segments are useful in children just as they are in adults. Requesting these specialized instruments during surgical planning may be necessary when operating in a children's hospital where they may not be readily available. Iris hooks are also a valuable tool when operating on pediatric cataracts following trauma. They may be used to retract torn or displaced iris tissue or help stabilize the anterior capsule/lens complex when there has been extensive zonular loss (Case 3).

Once a lens is placed in the capsular bag, the next decision involves creating or enlarging a posterior capsulectomy. Posterior capsular opacification (PCO) is said to occur in 17–100% of patients with intact posterior capsules at the time of primary surgery [3]. A capsulectomy can often be created with the vitrector from an anterior approach by gently moving the optic aside and enlarging an already present capsular opening. This same technique is also ideal to enlarge a posterior capsular

opening behind a sulcus placed lens. When the posterior capsule is intact, the same decision making applies as when deciding whether a child will be cooperative for a neodymium-yttrium-aluminum-garnet (Nd:YAG) capsulotomy in the future or if a pars plana approach should be utilized to create a posterior capsulectomy at the time of lens placement.

When posterior capsular support is minimal, a lens may be placed in the ciliary sulcus if at least half of the anterior capsule is present and stable [2]. Viscoelastic may be used to open the sulcus to make placing the leading haptic easier. Whether inserting a three-piece hydrophobic acrylic lens or a single piece that can be placed in the sulcus, leaving the trailing haptic anterior to the iris may at first be necessary. Once the lens is in the anterior chamber with one haptic in the sulcus and one haptic anterior to the iris, the enlarged corneal wound may be partially closed with 10-0 vicryl suture. This will allow for greater stability of the anterior chamber once the viscoelastic is removed. If not already present, an additional paracentesis should be made 180° from the trailing haptic and a small incision forceps (often found on an MST set) used to grasp and place the trailing haptic behind the iris with minimal lens manipulation in an otherwise weak zonular system. A forceps often provides better control and placement of a haptic than a traditional second instrument for both three-piece and single-piece lenses.

Postoperative Aphakia

Placement of an intraocular lens has been shown to result in better visual outcomes than aphakia, but of course there are times when the placement of a posterior chamber lens is not possible [2, 3] (see Chapter 32: Intraocular lens implantation in the absence of capsular support). Children tolerate contact lenses surprisingly well in the setting of unilateral aphakia. Corneal scarring after trauma may decrease visual acuity even after cataract removal. A specialty contact lens may help minimize irregular astigmatism and give a child their best-corrected vision, whether or not a secondary intraocular lens has been placed.

Postoperative Care

Even with the best surgical and medical management of traumatic cataracts in children, complications may arise. Aggressive refractive rehabilitation and amblyopia treatment should be initiated after surgery. Children should also be monitored closely for increases in intraocular pressure, recurrent inflammation, lens dislocation, or retinal detachment. Repeat examinations under anesthesia may be required for children who are difficult to examine in the office. Secondary surgical procedures are more common after traumatic cataracts and parents should be counseled

appropriately. If significant unilateral vision loss results from trauma, children may need full-time spectacle wear for protection.

Case 1

An 8-year-old male was fishing when he was hit in the eye with a fishing weight resulting in a complete hyphema and white cataract formation. Once the hyphema cleared, there was notable phacodonesis and 180° of zonular weakness with vitreous in the anterior chamber. The retina visible around the lens was normal and a B-scan ultrasound showed an attached retina and clear vitreous.

Two peripheral paracentesis wounds were made. An anterior chamber maintainer was used. Vitreous was removed from the anterior chamber with a 25-gauge vitrector on cut mode. A small opening was made with the vitrector in the center of the anterior capsule. Iris hooks were placed in the anterior capsular opening and used to stabilize and centralize the lens complex. The lens material was removed from within the capsular bag with the vitrector on I/A mode. An 11 mm capsular tension ring was inserted into the capsular bag followed by a three-piece MA60AC lens. A capsular tension segment was available, but not inserted because the lens centered well and was stable. A posterior capsulotomy was not performed.

The patient has a stable and well-centered lens 18 months following the injury; however, his best-corrected vision is 20/400 due to a macular scar. He wears glasses for protection.

Comment Surgical management of this traumatic cataract was greatly facilitated by the pediatric cataract surgeon's familiarity with the vitrector and anterior chamber maintainer. Similarly, management of vitreous in the anterior chamber can be handled without introducing new instrumentation to the surgeon's repertoire. However, use of a capsular tension ring, a tool many adult surgeons are familiar with, may be less commonly employed in pediatric cataract surgery. Overall, thorough surgical planning for all possible scenarios at the time of lensectomy will allow a surgeon to be prepared for anything that globe trauma in a child may bring.

Case 2

An 18-month-old female was noted to have a red eye with white material adherent to the cornea, 2 days after her brother poked her in the eye with a fork. Her mother did not witness the event and was unaware of the severity of the injury. When she presented, she was noted to have lens material plugging a full thickness corneal laceration (Fig. 20.1a). Vannas scissors were used to remove the lens material and the corneal wound was closed (Fig. 20.1b). She received

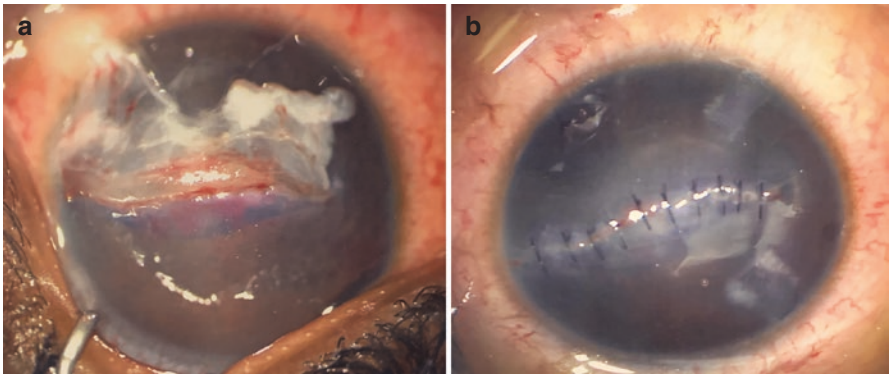

Fig. 20.1 (**a**) Lens material is extending through a full-thickness corneal laceration. (**b**) Corneal sutures at the conclusion of the corneal laceration repair and primary lensectomy

subconjunctival antibiotic and steroid injections and was placed on topical moxifloxacin four times per day for 1 week and prednisolone acetate four times per day for 1 month.

Four weeks following the primary injury, the patient returned to the operating room. The patient was unable to be thoroughly examined in the office due to age and cooperation. The cornea was significantly clearer, but a red reflex was absent. A B-scan ultrasound revealed vitreous hemorrhage with an attached retina with an area concerning for retinal traction. The patient was also examined by a retina specialist during this anesthesia. Residual lens material was removed and an anterior vitrectomy was performed. There was an absence of capsular support and a lens was not placed. The corneal sutures were removed.

The patient was initially followed by both a retina specialist and pediatric ophthalmologist. She had a corneal scar, but vitreous hemorrhage resolved and her retina remained attached. She did not keep her appointment for a contact lens fitting. When she returned for a follow-up exam 1 year after her initial injury, after missing several visits in a row, she had a central corneal scar, was aphakic and not wearing glasses or a contact lens, and had a closed and inoperable retinal detachment. She has been prescribed full-time glasses wear for protection of the uninvolved eye.

Comment The decision to leave this child aphakic was made due to absence of capsular support with the intention of contact lens management. Lack of follow-up and failure to obtain aphakic correction contributed to a poor visual outcome, made even more unfortunate by the total retinal detachment. Ultimately in the setting of trauma, even with timely repair of globe injury and removal of lens opacity, worsening corneal scarring, retinal scarring, retinal detachment, glaucoma, and hyphema, among other complications, can compromise visual acuity.

Case 3

A 6-year-old male was cutting with a pocket knife when his hand slipped and he sustained a self-inflicted penetrating injury to the cornea, lens, and sclera with uveal loss. He had a ruptured globe repair within 24 hours of the initial injury. Visual acuity was hand motion only.

The patient returned to the operating room 3.5 weeks following the initial injury. A B-scan ultrasound confirmed an attached retina and clear vitreous. Posterior iris synechia existed between the superior iris and the anterior capsule tear (Fig. 20.2a). Viscoelastic was used to gently separate the synechia from the anterior capsule. A 25-gauge vitrector was placed on aspirate mode to remove the lens material through the preexisting capsular opening (Fig. 20.2b). Hydrodissection was not

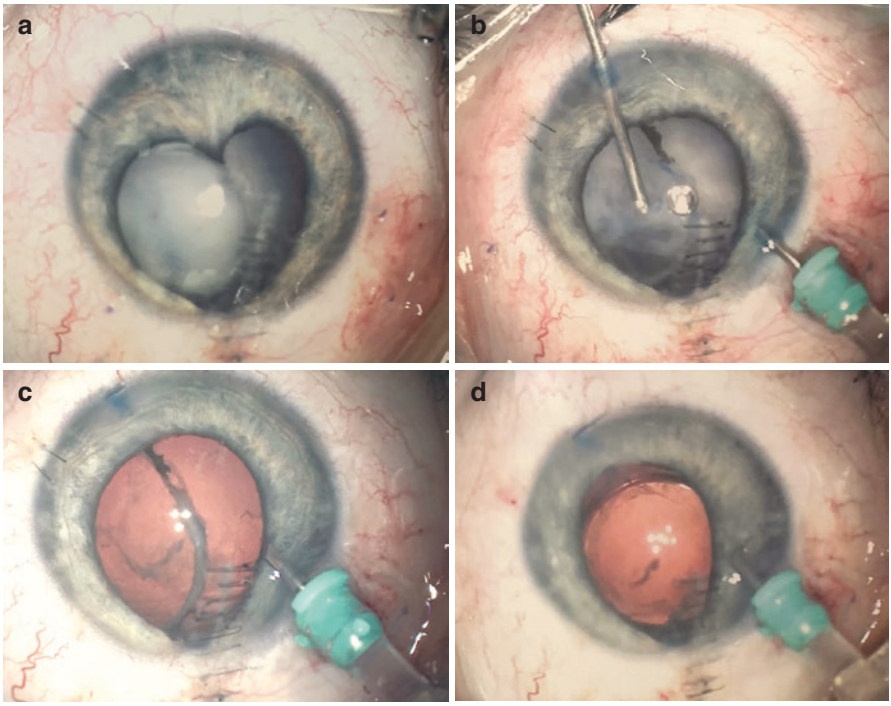

Fig. 20.2 (**a**) Eye following corneoscleral laceration repair with resulting cataract formation. Iris is adherent to the anterior capsule at the site of anterior capsular violation. (**b**) An anterior chamber maintainer is placed at 4'oclock and a paracentesis at 11 o'clock. A 25-gauge vitrector is used to remove lens material through a preexisting opening in the anterior capsule. (**c**) All lens material has been removed from within the capsule, but a dense fibrotic band remained in the anterior capsule. (**d**) The anterior capsulotomy was enlarged prior to IOL insertion

performed in case there was an opening in the posterior capsule due to the nature of the injury. Once the lens material was completely removed, an opening was found in the posterior capsule with residual fibrosis of the anterior capsule (Fig. 20.2c). Both the anterior and posterior capsular openings were enlarged and an anterior vitrectomy performed. A Rayner single-piece foldable acrylic intraocular lens is placed in the ciliary sulcus (Fig. 20.2d).

Two weeks following surgery the patient's visual acuity improved to 20/50 uncorrected and 20/25+ with a soft contact lens.

Comment Use of preexisting capsular opening is a safe technique for removing lens material, especially when the status of the posterior capsule is in question. Similarly, use of hydrodissection should be avoided. In this child, the surgical approach was in many ways similar to that in an adult, but additional attention to IOL selection and placement is required when considering the active lifestyle of a child and the increased lifetime of an IOL in a child.

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Chapter 21
Cataract Surgery and IOL Implantation in Children with Uveitis

Sydni Coleman, Karen R. Armbrust, and Raymond G. Areaux Jr.

Cataract formation is a common complication of pediatric uveitis due to chronic inflammation and corticosteroid use. Although the safety of cataract extraction with IOL implantation has been validated in children, perioperative management and even IOL implantation itself in children with uveitis is controversial [10]. Current literature is sparse and limited in power, consisting of small retrospective studies.

The most common cause of uveitis in childhood is juvenile idiopathic arthritis (JIA) associated, especially patients that are antinuclear antibody (ANA) positive with oligoarticular arthritis. Less common causes include pars planitis, sarcoidosis, toxoplasmosis, toxocariasis, and herpetic infections [1–3]. Additionally, many cases of uveitis are idiopathic [2, 4, 5]. Complications of chronic inflammation include posterior synechiae, ciliary body inflammation, cyclitic membranes which can be thick and robust in children with uveitis, band keratopathy, uveitic glaucoma, hypotony, and cataracts. The diagnosis and timely management of cataracts is especially important in children given their risk for developing amblyopia, but it is critical to recall that perioperative protocols for cataract surgery in children cannot be directly applied to children with uveitis. Similarly, protocols for cataract surgery in the setting of adult uveitis cannot be directly applied to children.

Determining the optimal timing for cataract surgery in pediatric uveitis requires balancing the competing interests of delaying surgery until inflammation is adequately controlled to reduce the unique risks of intraoperative and postoperative complications, with the increased risk of developing amblyopia in the setting of surgical delay. Timing considerations should include etiology of the uveitis, severity

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of inflammation, efficacy of therapy, secondary complications of uveitis, visual potential of the eye, and risk of amblyopia conferred by the morphology and density of the cataract in light of the patient's age. Pediatric uveitis with visually significant cataract formation typically requires systemic corticosteroid-sparing immunomodulatory therapy (IMT) in consultation with a pediatric rheumatologist. The definition of sufficient uveitis control varies by etiology and expert opinion but most experts recommend a minimum of 3 months of uveitis quiescence prior to surgery [13, 14], and some physicians advocate for longer periods of sufficient inflammatory control prior to cataract surgery [4, 6]. Particularly in the pediatric population, cooperation with slit lamp and fundus examination and ancillary testing may limit the data available for clinical decision making.

Experts agree that limiting the inflammation caused by cataract surgery is critical and requires additional intensive perioperative topical and systemic corticosteroids, even with excellent preoperative inflammatory control [11, 12]. Our practice generally is to use oral prednisone 1 mg/kg/day for 2 days prior to surgery and an intraoperative dose of intravenous systemic corticosteroid (methylprednisolone or dexamethasone). Depending on uveitis severity, postoperative corticosteroid treatment may be solely topical or a combination of systemic and topical corticosteroids. In pediatric uveitis patients, postoperative corticosteroids are tapered over a longer period and are more frequently adjusted based on clinical examination than in non-uveitic patients.

In the past many pediatric uveitic cataract patients were left aphakic after cataract extraction. However, in 1996 Probst and Holland reported successful IOL implantation in a small population of patients with JIA-associated uveitis [7]. Additional studies have validated this observation in small cohorts with varying caveats. Contact lens intolerance in the context of a need for ongoing topical corticosteroids, irregular ocular surface, and/or glaucoma drainage device associated bleb is a potential reason to favor IOL implantation, at least in certain cases. If an IOL is implanted, IOL biocompatibility is important to reduce inflammation. Heparin sulfate modified (HSM) PMMA and acrylic lenses are preferred over non-HSM PMMA and silicone lenses due to greater biocompatibility [8, 9]. Acrylic lenses typically are favored since they are less expensive, require a smaller wound, and are more widely available than HSM-PMMA IOLs.

Each case will be presented and followed by commentary from Dr. Areaux (regarding surgical and amblyopia management) and Dr. Armbrust (regarding uveitis control and appropriateness for IOL implantation).

Case 1

BI is a 12-year-old African female who presented with bilateral granulomatous anterior uveitis, active on topical corticosteroids, with secondary findings of bilateral posterior subcapsular cataracts, extensive bilateral posterior synechiae, and bilateral optic disc edema. Visual acuity with glasses was 20/25 right eye and 20/60

pinhole 20/25 left eye. Her history of joint pains in her back and hands raised suspicion for JIA, although examination with a pediatric rheumatologist was unremarkable. Infectious and inflammatory workup was normal including negative Quantiferon gold, RPR, Treponema antibody, Lyme Ab, Bartonella Ab, rheumatoid factor (RF), ACE, HLA-B27, anti-CCP, anti-SSA (Ro), CRP, and urinary beta-2 microglobulin. ESR was elevated (41, normal 0–15) and ANA was mildly elevated (1.8, normal <1.0). The patient was diagnosed with idiopathic chronic anterior uveitis.

The patient's age and her good best-corrected visual acuity alleviated concern for amblyopia development, and total control of uveitis was prioritized over cataract surgery. Oral corticosteroids and methotrexate were added to the topical corticosteroids, and adalimumab was added when uveitis recurred during systemic corticosteroid taper. Uveitis control and topical atropine were unsuccessful at lysing posterior synechiae. Additionally, the patient's clinical course was complicated by uveitic glaucoma with steroid-induced ocular hypertension, which was treated with maximal topical therapy of latanoprost, dorzolamide, timolol, and brimonidine, but elevated intraocular pressures persisted. Diamox was added for pressure control, and adalimumab was increased to weekly dosing in order to taper off oral prednisone. The uveitis was finally controlled (zero to trace cell in both eyes) off corticosteroids on weekly adalimumab and subcutaneous methotrexate 15 mg per week. The optic disc edema slowly resolved over the year following initiation of systemic uveitis treatment.

At this point, significantly reduced vision from cataracts (BCVA 20/100 OD, 20/70 OS) was impacting the patient's school performance and daily life. Combined cataract and glaucoma surgery were performed after inflammation had been controlled for 4 months. Posterior synechiolysis, cataract extraction, acrylic IOL (Alcon SN60WF) implantation, viscocanaloplasty, and circumferential trabeculotomy ab interno was performed on each eye 1 week apart. Inflammation was well-controlled with the following corticosteroid regimen: 1 mg/kg/day oral prednisone for 2 days prior to surgery, Solumedrol 1 mg/kg intraoperatively, a postoperative taper of oral prednisone over 12 days, and postoperative topical prednisolone acetate. After oral prednisone taper, anterior chamber inflammation was controlled on topical prednisolone acetate TID, weekly adalimumab, and 17.5 mg/week methotrexate. Although there was RNFL thickening as measured by OCT in the 3 months after surgery, the thickening was mild compared to presentation and it slowly returned to the patient's baseline. IOP normalized without medical therapy, despite chronic topical prednisolone acetate. BCVA was 20/20 in both eyes 7 months after surgery.

Comment (RGA) This case was challenging due to the dual threat of both cataracts and glaucoma in the setting of uveitis. Although the patient was not at risk for amblyopia, the need for maximal topical therapy and oral acetazolamide for IOP control raised the urgency. Gonioscopy was obviously key preoperatively and revealed angles with early uveitic changes but still amenable to surgery in both eyes by Spaeth grading: right eye was B-C 20f 1-2+PTM with fine scattered PAS and left eye was B 20f 1-2+PTM with fine scattered PAS. The option of simultaneous

glaucoma and cataract surgery was preferred by the patient and her family to optimize recovery time, but we discussed the possibility that excessive hemorrhaging during synechiolysis or trabeculotomy might obscure the view for additional surgery and require staging the procedures. Fortunately, this did not occur. In prepping the operating team for the case, iris hooks, intraocular scissors, and trypan blue were requested in addition to our usual equipment for lensectomy, anterior vitrectomy, and IOL implantation in a child. The patient and family were very reliable with excellent uveitis control preoperatively for 4 months, and sufficient support for an IOL was anticipated given that there had not been a prolonged course of uveitis. As such, an IOL implant was planned with the caveat to the family that intraoperative complications might preclude the placement of a lens. Direct gonioscopy lenses and the OMNI® (Sight Sciences, USA) combined viscocanaloplasty and trabeculotomy *ab interno* device was also requested. Despite posterior synechiae, the pupil was fairly mobile so preoperative dilation was planned, followed by cataract extraction and IOL implantation. Then Miochol was used to constrict the pupil and expose the angle, followed by *ab interno* angle surgery.

Intraoperatively through a clear corneal approach, trypan blue was used to improve visualization of the lens capsule at the start of the case and Healon GV was used to stabilize the lens for anterior capsulotomy. A cyclodialysis spatula was used to lyse numerous posterior synechiae (Fig. 21.1). In a bimanual approach, 20-gauge vitrectomy was used to fashion a 5 mm anterior capsulotomy. Irrigation and aspiration were used to remove the entirety of the cataractous lens without complication. Healon was used to fill the capsular bag and an SN60WF Alcon lens was injected into the bag. Healon was exchanged for BSS and the wounds were closed with 10-0 vicryl, leaving one untied for subsequent angle surgery (Fig. 21.2). Healon GV was used to deepen the nasal angle and central anterior chamber. The head was turned 45° in the opposite direction of the eye and the scope tilted 45° to align the plane of sight with the ipsilateral temple. A Swan-Jacobs lens was placed on the eye and the nasal angle was visualized clearly (Fig. 21.3). The OMNI® device needle was introduced into the anterior chamber and the nasal angle was engaged. A 360-degree viscocanaloplasty and subsequent trabeculotomy *ab interno* was completed. The hope (though there is no evidence currently) with this technique is that viscoelastic

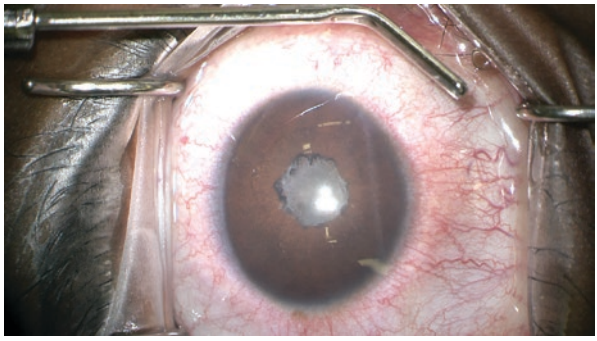

Fig. 21.1 Numerous posterior synechiae prior to lysis with a cyclodialysis spatula

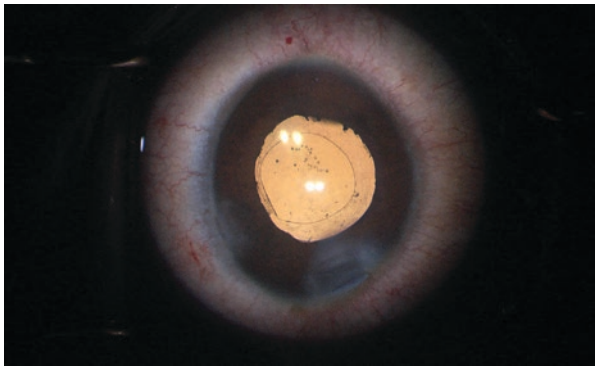

Fig. 21.2 Clear central axis after cataract extraction and IOL implantation

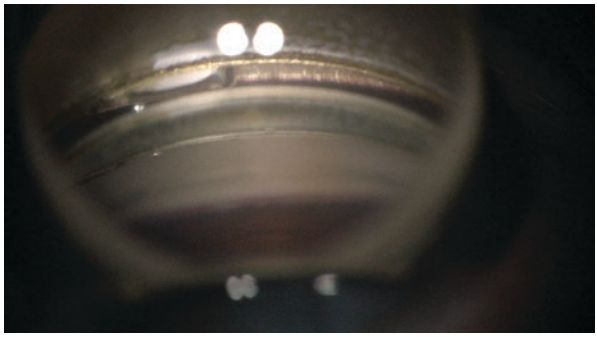

Fig. 21.3 Clear view of nasal angle as visualized using a Swan-Jacobs lens

may be advanced into the collector channels beyond the canal of Schlemm and improve outcomes compared to trabeculotomy alone in eyes with uveitic glaucoma by releasing scar tissue downstream of the trabecular meshwork similar to the way that goniosynechialysis works to release scarring in the angle in these patients. Healon GV was exchanged for BSS and the final wound was closed. 0.05 mL of Triesence followed by 0.1 mL of 50:50 Vigamox/BSS was injected into the anterior chamber at the end of the case. Maxitrol ointment and 1% atropine were placed on the eye followed by a light pressure patch and a shield. Postoperatively the IOP normalized without medications and the vision has been excellent. Corectopia, an artifact of preoperative uveitic iris ischemia, is visible superiorly in the slit lamp photo at 9 months postoperatively (Fig. 21.4).

Comment (KRA) This case illustrates the need for cooperation between ophthalmology and rheumatology to achieve sustained uveitis quiescence, which is the cornerstone for a successful ophthalmic outcome in severe, chronic, noninfectious uveitis. The ophthalmologist may start topical corticosteroids prior to obtaining results from an infectious evaluation, as long as the patient's presentation is more consistent with a noninfectious etiology. In this case, with bilateral granulomatous anterior uveitis and optic disc edema, the infectious workup should include testing for syphilis, tuberculosis, Lyme (in an endemic area or with suspected exposure),

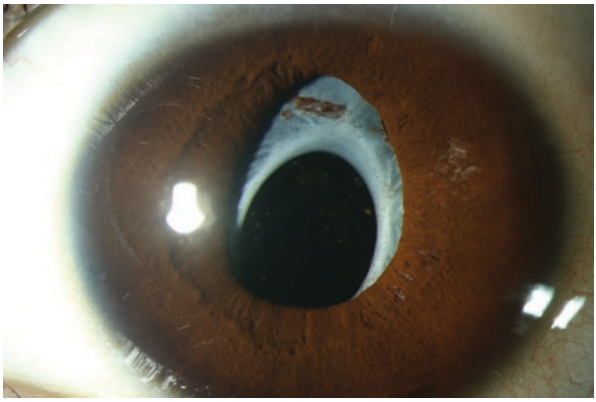

Fig. 21.4 Slit lamp photo showing corectopia superiorly at 9 months postoperatively

and bartonella. Then, as long as the infectious evaluation is negative, the ophthalmologist may add systemic corticosteroids depending on the severity of the uveitis and refer to a pediatric rheumatologist for management of systemic corticosteroid-sparing IMT. Historically, methotrexate has been the first-line IMT agent for many types of pediatric uveitis and remains an excellent choice for many pediatric patients. The newer anti-TNFα agents also are efficacious for uveitis and typically well-tolerated, but may require additional time and effort on the part of the rheumatologist for insurance approval. In a pediatric patient with severe noninfectious uveitis, letting the rheumatologist know at the time of initial consultation that starting methotrexate with likely need for addition of an anti-TNFα agent may facilitate timely approval of medications.

It is instructive to examine the response of posterior synechiae and uveitic optic disc edema to treatment in this case. Although posterior synechiae may lyse with uveitis control and cycloplegia, this type of treatment is more successful with newly formed synechiae. Chronic posterior synechiae often require surgical intervention, as in this case. Improvement and resolution of uveitic optic disc edema typically lags behind improvement in uveitis, and here the RNFL thickening on OCT continued to improve 6 months after the anterior uveitis became quiescent.

The relatively older age, good preoperative uveitis control without corticosteroids, and excellent patient and family compliance make IOL implantation in this case a good decision. IOL calculations are more accurate in older children, so delaying cataract surgery if feasible can be advantageous purely on a refractive basis. More importantly, if there is genuine concern that uveitis control will be difficult in the postoperative period, aphakia is preferred over IOL implantation. Therefore, the quality and duration of uveitis control, need for corticosteroids for uveitis control, and patient compliance all are important factors when considering IOL implantation.

This case is a good illustration of the inflammatory burden of surgery in pediatric patients with uveitis. In the early postoperative period it is critical to follow uveitis patients closely and promptly increase anti-inflammatory medications with any sign of inflammation, such as anterior chamber cell, posterior synechiae to the intraocular lens, optic disc edema, and cystoid macular edema. It also is important to expand

our surveillance later after surgery. In this case, despite excellent preoperative inflammatory control on systemic steroid-sparing immunosuppression for 4 months prior to surgery, increased uveitis persisted even after the typical postoperative period of 3 months. Seven months postoperatively, uveitis quiescence required increased methotrexate and addition of topical corticosteroids as compared to the patient's preoperative regimen. With close monitoring and adjustment of medications as needed, excellent uveitic and surgical outcome was achieved in this case.

Case 2

AL is a 9-year-old Asian male who presented with bilateral anterior and intermediate uveitis. Infectious workup was negative including Quantiferon gold, RPR, Treponema antibody, and Lyme antibodies. Inflammatory labs including CRP, ANA, cANCA, pANCA, ACE, and HLA-B27 were all normal. ESR was elevated. Evaluation by rheumatology revealed evidence of inflammatory arthritis involving the knee and TMJ (confirmed on MRI). The patient was diagnosed with JIA-associated anterior and intermediate uveitis. Treatment was initiated with topical corticosteroids (prednisolone acetate, then difluprednate), high-dose oral prednisone (started at 1 mg/kg/day), and methotrexate, and then adalimumab (every 2 weeks) was added 3 weeks later for persistent anterior chamber cell and vitreous haze despite high-dose systemic corticosteroid treatment. Methotrexate was slowly tapered upward to 20 mg/week, and the combination of methotrexate and adalimumab allowed total inflammation control by 6 months with successful wean off oral corticosteroids at 7 months and topical corticosteroids at 12 months. This patient's course was complicated by ocular hypertension prior to corticosteroid therapy as well as steroid-induced ocular hypertension that was treated with brinzolamide, timolol, latanoprost, and brimonidine.

At presentation there was mild nuclear sclerotic cataract of the left eye; 1 month later, posterior subcapsular cataract (PSC) of the left eye developed centrally, but uncorrected visual acuity remained excellent at 20/25+2 when all corticosteroids were stopped and uveitis was quiescent. Over the next 8 months, visual acuity remained stable, the uveitis was quiescent, and no corticosteroids were prescribed. However, in 2 months, the PSC rapidly became visually significant, with BCVA 20/200. The patient and family elected to proceed and cataract extraction with intraocular lens implant along with posterior synechiolysis. Perioperative steroid management included oral prednisone (0.5 mg/kg/day) 2 days prior to surgery, dexamethasone 4 mg intraoperatively, and a short oral prednisone taper postoperatively (0.5 mg/kg/day for 2 days, then tapered over 4 days).

After instillation of Healon GV, lysis of fairly extensive posterior synechiae was completed using an iris sweep and the significantly fibrotic pupil was stretched with a Kuglen hook (Fig. 21.5). Trypan blue was then instilled beneath the Healon GV and over the anterior capsule. Additional Healon GV was used to visco-dilate the pupil and stabilize the capsule. A cystotome was used to initiate and

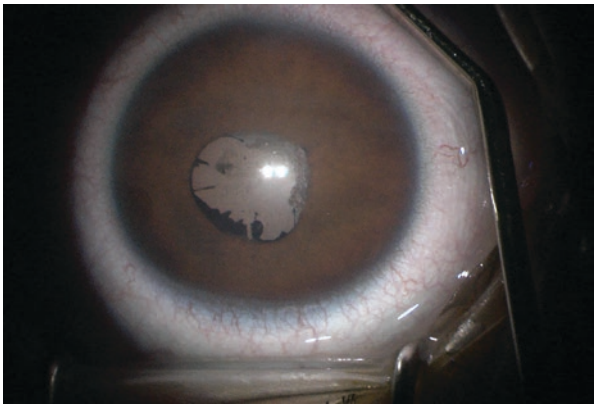

Fig. 21.5 Extensive posterior synechiae and PSC cataract

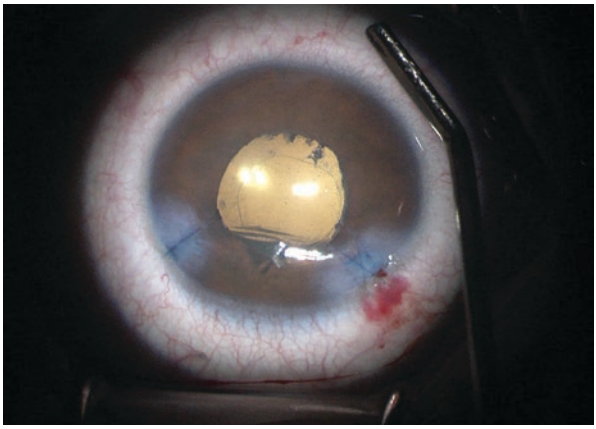

Fig. 21.6 Clear central axis after cataract extraction and IOL implantation

micro-capsulorhexis forceps were used to complete a continuous curvilinear capsulorhexis of 4.5 millimeters. In a bimanual approach, an irrigation handpiece and vitrector handpiece were used to remove the entirety of the cataractous lens without complication. Healon was used to fill the capsular bag, and an SN60WF Alcon lens was injected into the bag using the Monarch® delivery system. Healon was exchanged for BSS and the wounds were closed with 10-0 vicryl (Fig. 21.6). 0.05 milliliters of Triesence and 0.1 milliliters of Vigamox that had been diluted with sterile balanced salt solution in a 1:1 ratio were instilled into the anterior chamber at the end of the case. 1 drop of 1% atropine, apraclonidine, and timolol were placed on the eye followed by Maxitrol ophthalmic ointment, a light patch, and a shield.

Topical prednisone was increased to QID postoperatively and slowly tapered over 6 months based on anterior chamber cells. Six months postoperatively, the BCVA was 20/25 and the patient remained on topical prednisolone acetate 1% daily along with timolol and brinzolamide for IOP control. Six months later, despite aggressive utilization of perioperative systemic and topical steroids and a very slow ongoing taper of topical steroids, a posterior synechia that formed in the immediate postoperative period can be seen just below 9:00 (Fig. 21.7).

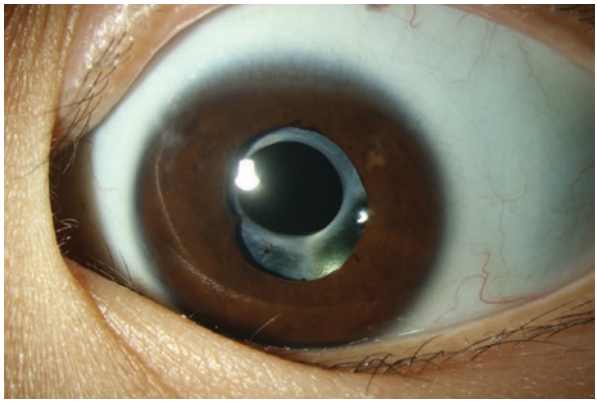

Fig. 21.7 A posterior synechia can be seen just below 9:00

Comment (*RGA)* Preoperatively there was debate as to whether simultaneous angle surgery was warranted to control IOP since we would already be in the eye and the higher efficacy of angle surgery in uveitic glaucoma early in the disease process. The IOP had spiked markedly with topical steroids implying a significant steroid response component and the family wished to minimize surgical risk. As a result, we agreed to exclusively cataract surgery with subsequent angle surgery if the IOP remained uncontrolled. Subsequent to surgery, the patient has been weaned to timolol daily and brinzolamide BID with excellent tolerance and IOP control in the teens. Further weaning of ocular hypotensives is planned. Thus, glaucoma surgery is not planned at this time.

The timeline to inflammation control merits discussion. It took 6 months to achieve control (zero to trace cell) of the uveitis. In a 9-year-old, the risk of amblyopia was essentially zero and the cataract did not become visually significant until 1 year after initial presentation. I have faced similar scenarios in younger patients, 4–5 years old, where aggressive topical and systemic immunomodulatory therapy for months result in improved but incompletely controlled inflammation and inadequate control for IOL implantation (at least 1+ active cell). In these scenarios, I have elected to intervene to remove the cataract with a posterior capsulotomy and anterior vitrectomy and leave the child aphakic with contact lens correction initiated within 1 week postoperatively. Aggressive amblyopia rehab with patching and bifocals can then be advanced while inflammation is further controlled. Secondary IOLs can be considered later with caution in the context of the level of inflammation control and the optimized visual potential.

Comment (KRA) The etiologic evaluation for anterior versus intermediate uveitis has substantial overlap but differs slightly. The first priority in any type of uveitis is to evaluate for infection. Syphilis should be ruled out in any type of uveitis. Lyme disease should be ruled out in both anterior and intermediate uveitis cases that are bilateral in an endemic area. Tuberculosis should be ruled out in granulomatous anterior uveitis, in any intermediate uveitis, and prior to systemic immunosuppression. In unilateral intermediate uveitis toxoplasmosis and toxocariasis should be

considered; these typically are diagnosed clinically but may require examination under anesthesia in the pediatric population. An elevated ESR is nonspecific but is helpful in raising suspicion for systemic inflammatory syndromes, such as JIA, TINU, and sarcoidosis. JIA-associated uveitis is typically anterior, but as in this case intermediate JIA-associated uveitis can occur. TINU also is more typical in anterior than in intermediate uveitis. Sarcoidosis should be a consideration with any type of uveitis. Intermediate uveitis has been associated with multiple sclerosis in studies of adult patients; however, multiple sclerosis-associated uveitis does not appear to be common in the pediatric population since no cases of multiple sclerosis were found in a series of pediatric intermediate uveitis patients [15]. A directed review of systems can help rule out multiple sclerosis symptoms, with a neurologic consultation and/or brain MRI to evaluate for white matter lesions if the review of systems is suspicious. Brain MRI to screen for multiple sclerosis in pediatric intermediate uveitis patients without neurological symptoms is not routinely performed. Again, this case illustrates the need for active collaboration between ophthalmology and rheumatology for appropriate systemic treatment, and in this case, rheumatologic evaluation also revealed the underlying etiology of JIA.

Uveitis and steroid-induced cataracts are significant complications in children with uveitis. Cataract surgery in pediatric patients with uveitis poses unique challenges. Excellent visual outcomes are possible with proper timing, perioperative optimization of inflammation control, and with the cooperation of a multidisciplinary team of pediatric ophthalmologists and rheumatologists. Published data on this topic is limited prompting the need for further research.

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Chapter 22
Intraocular Lens Placement in the Setting of Glaucoma

Emily M. Zepeda and Brenda L. Bohnsack

Glaucoma in the Setting of Aphakia

The most common clinical scenarios in which glaucoma complicates aphakia are glaucoma following cataract surgery and uveitic glaucoma. Intraocular pressure (IOP) management must take precedence as substantial vision loss due to glaucoma negates the importance of intraocular lens (IOL) placement for visual rehabilitation. In both glaucoma following cataract surgery and uveitic glaucoma, topical antihypertensive medications are the first-line treatment for increased intraocular pressures [1, 2]. In eyes refractory to medications, the choice of traditional surgical options including goniotomy, trabeculotomy, trabeculectomy, glaucoma drainage devices, and cycloablation (transscleral and endoscopic) is dependent on factors such as angle configuration, corneal clarity, eye size, and previous eye surgeries [3–11]. Glaucoma surgery that is required to obtain IOP control should be done prior to secondary IOL placement.

Evidence of good IOP control includes serial pressure measurements, reversal of optic nerve cupping, resolution of corneal edema, and stabilization of corneal diameter and axial length. While axial lengths measured via A-scan ultrasound or optical biometry systems (i.e., Lenstar®, IOL-Master®) are most accurate, they may not be plausible in-clinic options for young children. In this case, stabilization of the refraction may be an acceptable substitute. For the purpose of IOL selection, it is important to note that in glaucomatous eyes with buphthalmos, rapid intraocular pressure control obtained through glaucoma surgery often causes a sudden decrease

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in axial length followed by a slower re-expansion of the globe. The final length may fall between the immediate postoperative and the maximum buphthalmic eye lengths [12, 13]. Thus, selecting a lens based on a buphthalmic measurement can result in a hyperopic shift, while not waiting until axial length stabilization following glaucoma surgery may cause a more myopic end refraction [14, 15].

Intraocular Lens Placement in Glaucoma Following Cataract Surgery

Following congenital cataract extraction, it is well-known that the newly rendered aphakic eye is at high risk for developing glaucoma, with an incidence ranging from 15% to 50% [16, 17]. In these patients, elevated IOP is often observed within the first few years after surgery but can occur years later, highlighting the importance of diligent, lifelong surveillance.

The majority of cases of glaucoma following cataract surgery have an open-angle configuration and arise directly because of the aphakic status [16–19]. In these cases, the angle typically has a normal appearance both prior to and following cataract surgery making it unclear as to the pathogenesis of this form of glaucoma [20, 21]. Since the most notable risk factor is age at the time of cataract surgery, with infants under the age of 2 months having the highest rate of glaucoma, the mechanistic theories revolve around the lens, mechanically (tension from the lens on the ciliary body) and/or molecularly (secreted factors), being required for angle and aqueous outflow channel development [16, 19, 22]. Additional theories suggest that post-surgical inflammation or vitreous factors released after breaking the anterior hyaloid damage the aqueous channels [23]. In these cases, antihypertensive medication is the first-line treatment; however, up to 50% of eyes require glaucoma surgery for IOP control [24, 25]. Angle surgery (goniotomy or trabeculotomy) is typically the primary glaucoma surgery and has a 50–70% success rate [26–30]. If angle surgery fails to control IOP, many patients will have placement of a glaucoma drainage device [31, 32]. In eyes that are aphakic, placement of the tube in the pars plana with a concurrent vitrectomy should be considered. This is especially true if a secondary IOL will be implanted in the future [33–35]. Additional patients may undergo trabeculectomy, although filtering surgeries with adjunctive anti-fibrotics have become less popular in the pediatric population due to the lifelong risk of endophthalmitis [36, 37]. Another option is cyclophotocoagulation, which can be successfully used to control IOP either alone or in conjunction with glaucoma drainage devices [38].

In some cases of glaucoma following cataract surgery, there is a narrow-angle or a closed-angle configuration. In these eyes, there are often preexisting pathologic states such as microphthalmia and/or microcornea [16, 18]. Elevated IOP may develop early during childhood due to the congenital anatomically shallow anterior chambers. In addition, glaucoma may be diagnosed in late childhood to adulthood

due to further crowding of the anterior chamber from the Soemmering ring, which gradually increases in diameter due to proliferation of the lens epithelial cells and cortical fibers within the posterior and anterior capsules [39–41]. In these situations, angle surgery often fails, and glaucoma drainage device implantation is typically used to obtain IOP control [33]. However, given the shallow anterior chamber, the safest place for the tube is posteriorly placed within the pars plana.

Case 1

The patient is a 9-year-old girl with a history of congenital cataracts with microphthalmia of both eyes. She underwent cataract extraction of the right eye at 6 weeks of age and the left eye at 7 weeks of age. Her vision was corrected with contact lenses, which she tolerated well. She was diagnosed with glaucoma following cataract surgery in both eyes at 7 years of age and underwent combined 180-degree trabeculotomy with trabeculectomy at 8 years of age. However, her intraocular pressure in her right eye remained uncontrolled, and she was referred for further treatment.

At the time of presentation, the patient's best-corrected visual acuity with +20.0 SilSoft® contact lenses was 20/40 in the right eye and 20/25 in the left eye. The intraocular pressures by Goldmann tomometry tonometry were 30 mmHg in the right eye and 17 mmHg in the left eye on timolol, dorzolamide, brimonidine, and latanoprost in both eyes. Slit lamp examination showed that both corneas were thin and clear (Fig. 22.1a). The right cornea was 9 mm in diameter, and the left cornea was 10 mm in diameter. The anterior chambers were shallow with approximately 2.5 mm depth centrally and 0.5 mm depth peripherally. Both eyes were aphakic and had Soemmering rings. Fundoscopic examination showed a dysplastic optic nerve with a cup to disc ratio of 0.8 in the right eye. The left eye had a cup to disc ratio of 0.1. The macula, vessels, and retinal periphery of both eyes were normal.

For glaucoma control, the patient underwent pars plana placement of a Baerveldt® 350 glaucoma drainage device with concurrent vitrectomy in the right eye at 9 years of age. This was followed by pars plana placement of a Baerveldt® 350 glaucoma drainage device with concurrent pars plana vitrectomy in the left eye at 10 years of age.

At 12 years of age, the patient desired placement of IOLs. At that time, her best-corrected visual acuity was 20/20 in each eye. Slit lamp examination showed blebs over the superotemporal Baerveldt plates in both eyes. Intraocular pressures by Goldmann applanation tonometry were 15 mmHg in both eyes on no glaucoma medications. The right optic nerve showed reversal of cupping with a color cup to disc ratio of 0.3 (Fig. 22.1b). Axial lengths measured 20.4 mm in the right eye and 19.73 mm in the left eye, and keratometry was 42.39 × 44.64 @ 98 in the right eye and 44.57 × 45.82 @ 94 in the left eye. The patient underwent serial placement of a three-piece acrylic lens in the sulcus in both eyes.

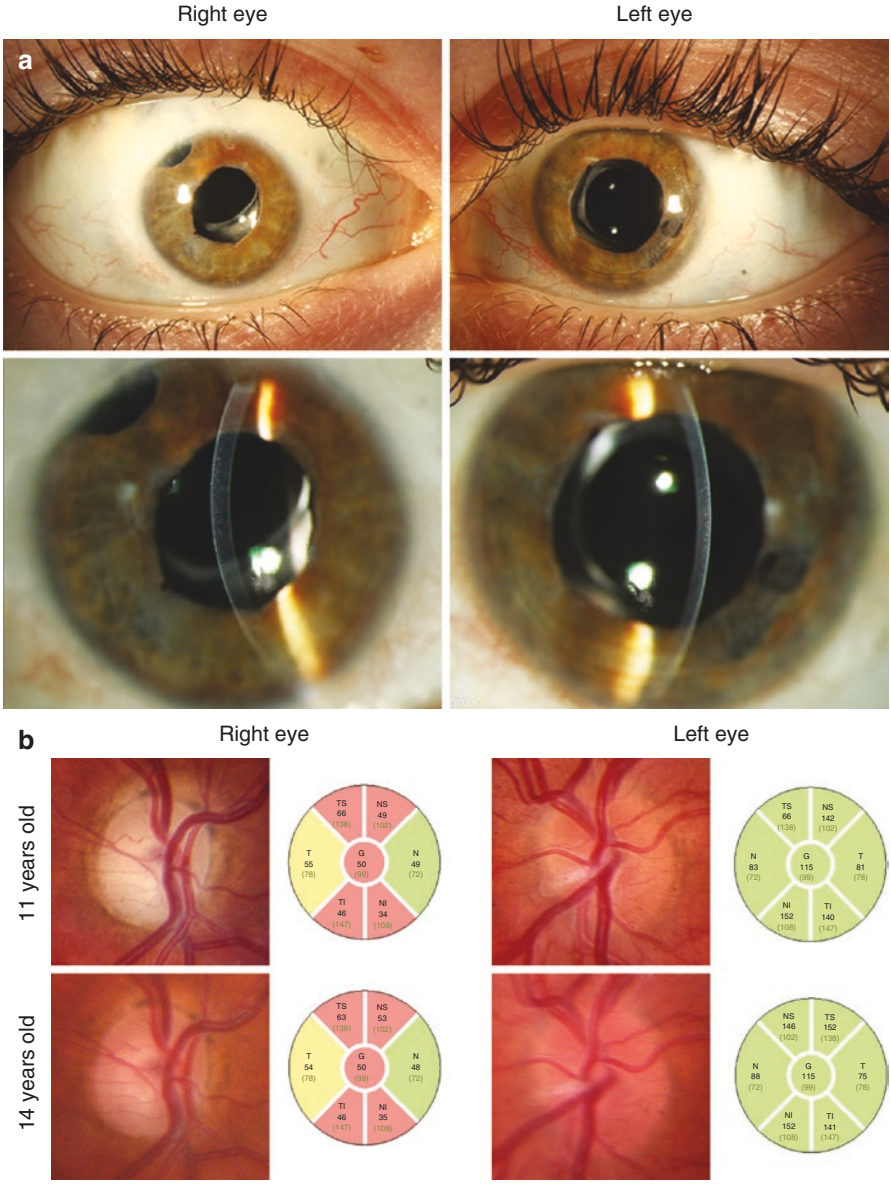

Fig. 22.1 Case 1. (**a**) External and slit lamp photographs of the right and left eyes at time of final follow-up in Case 1 demonstrated bilateral microcornea with shallow anterior chambers. Intraocular lenses were in good position, anterior to the Soemmering rings in both eyes. The superotemporal scleral patch graft was seen in the right eye, but was covered by the upper lid in the left eye. The tubes in both eyes were in the pars plana and thus not evident in the photographs. (**b**) Optic nerve photographs and optical coherence testing prior to intraocular lens placement (11 years of age) and at final follow-up (14 years of age) showed no glaucomatous progression. (**c**) Humphrey visual field testing demonstrated a superior arcuate and an early inferior arcuate defect in the right eye that correlated with the retinal nerve fiber layer thickness. Visual field in the left eye was full

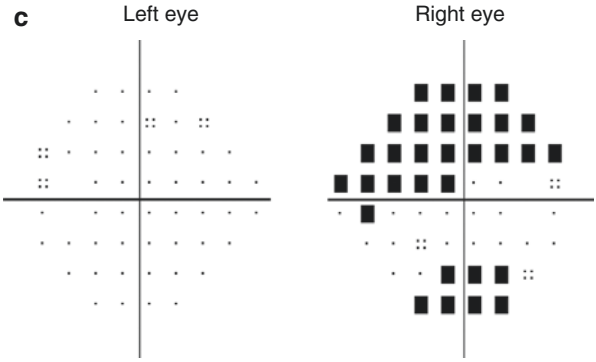

Fig. 22.1 (continued)

At final follow-up at 14 years of age, the patient's uncorrected visual acuity was 20/20 in each eye. Her intraocular pressures by Goldmann applanation tonometry were 16 mmHg in the right eye and 15 mmHg in the left eye on no glaucoma medications. She had superotemporal blebs over the Baerveldt plates, thin and clear corneas, shallow anterior chambers, and sulcus IOLs in both eyes. The optic nerves were stable in appearance (Fig. 22.1b) as was retinal nerve fiber layer thickness and visual field testing (Fig. 22.1c) compared to before IOL placement.

Comment Placement of a secondary IOL in an eye with glaucoma following cataract surgery requires IOP control and recognition of the angle and anterior chamber anatomy. Special consideration should be given to the anatomical features of the aphakic eye with glaucoma when deciding whether to place the lens in the sulcus or the capsular bag [42, 43].

Secondary IOLs are typically placed within the sulcus, using the Soemmering ring as a scaffold [44]. However, in the setting of glaucoma, placement of the IOL within the sulcus may further impede outflow through the trabecular meshwork especially in eyes with shallow anterior chambers due to microphthalmia or microcornea. Furthermore, in eyes that have previously undergone angle surgery, gonioscopy should be done prior to IOL placement to evaluate patency of the cleft and the amount of space within the peripheral anterior chamber. Another consideration for the surgeon is that commercially available one-piece acrylic lenses (Alcon® SA60AT) come in powers up to 40 diopters; the three-piece acrylic lenses (Alcon® MA60AC) only have a maximum power of 30 diopters. Especially in small, microphthalmic eyes, placement of a high-power one-piece lens within the capsule allows for greater accuracy in achieving the target refraction.

In order to minimize anterior chamber crowding, maintain patency of the angle, and place the IOL within the capsule, the Soemmering ring needs to be opened and debulked [43]. In this procedure, a 4–5-clock-hour peritomy is created. A 3 mm scleral tunnel is centered within the incision, and a stab incision at the limbus is made on each side of the tunnel. An anterior chamber maintainer is placed through one of the stab incisions. A MVR blade, placed through the other stab incision, is used to

separate the anterior and posterior capsules where they are fused centrally. Care should be taken to open the ring 360 degrees for maximal removal of lens material. The vitrector is then used to remove the proliferated cortical fibers and lens epithelial cells within the capsule. Dense Soemmering rings with hardened, calcified cortical fibers may not be easily removed with the vitrector. Viscoelastic can be injected into the anterior chamber, and the calcified fibers can be prolapsed out of the capsule in the anterior chamber. The scleral tunnel is then opened with a keratome, and the calcified fibers can be removed "extracapsular" style through the scleral tunnel. Following removal of the remnant lens fibers, the anterior and posterior capsules are inspected. If there is adequate support, then the IOL can be carefully placed within the capsule. If the posterior capsule does not have enough support, the entire IOL can be placed in the sulcus, or only the haptics can be placed in the sulcus with optic capture. A third option for older teenagers (>16 years of age) and adults is complete removal of the Soemmering ring and capsule and placement of a scleral-fixated IOL.

In microphthalmic eyes with glaucoma, if the aqueous outflow is not dependent on the angle, a secondary lens can be placed in the sulcus without debulking the Soemmering ring as described in the case above. In this example, the patient's intraocular pressures were controlled by glaucoma drainage devices allowing the lenses to be placed in the sulcus and the Soemmering ring left intact. It is important to note that in both eyes the tube portion of the glaucoma drainage device was preemptively placed in the pars plana in a combined procedure with a vitrectomy [33]. Posterior placement of the tube serves two purposes: (1) long-term prevention of corneal decompensation in a shallow anterior chamber and (2) better positioning of the tube for later placement of a secondary IOL [33, 34, 45]. In aphakic eyes with glaucoma drainage devices previously placed within the anterior chamber, consideration should be given to moving the tube to the pars plana either in conjunction with or prior to placement of a secondary IOL. The disadvantage to posterior placement of glaucoma drainage devices is the need for concurrent vitrectomy. Coordination of surgery between retina and glaucoma specialists can be difficult but decreases the need for multiple surgeries and improves communication to ensure adequate vitreous removal in the area of the tube. The increased risk of retinal detachment due to vitrectomy is low and outweighs the risk of corneal decompensation, especially in eyes with crowded anterior chambers [33–35, 45]. With pre-planning and appropriate tube placement, Case 1 demonstrates how sulcus placement of a secondary IOL achieves excellent uncorrected visual acuity and maintenance of intraocular pressure control.

Thus, when placing an IOL in an eye with glaucoma following cataract surgery, attention should be paid to the size of the anterior chamber and the previous glaucoma surgeries. Care must be taken to prevent exacerbation of intraocular pressures in eyes that are prone for glaucoma due to the aphakic status and inherent ocular anatomy.

Intraocular Lens Placement in Uveitic Glaucoma

Uveitis causes significant visual impairment as chronic inflammation leads to cataracts, glaucoma, band keratopathy, synechiae, and macular edema [46–48]. Treatment focuses on suppression of inflammation in order to prevent long-term damage first with local and systemic steroids and second with systemic

steroid-sparing therapy. However, both the inflammation and steroids contribute to cataract formation and increased intraocular pressures [49, 50]. The removal of uveitic cataracts should only be undertaken after preoperative inflammation control has been achieved. In general, there should be at least 3 months without uveitic activity prior to cataract extraction. However, whether an IOL can be placed in uveitis remains unsettled. Although many surgeons now elect to place standard acrylic IOLs, there remains a possibility that the IOL could exacerbate the inflammation inciting further complications. Thus, it is an acceptable practice to leave the eye aphakic, especially in situations of tenuous uveitis control [51–53].

Increased IOP in uveitis is due to multiple mechanisms. Uveitic debris collects within the trabecular meshwork and angle leading to decreased aqueous humor outflow [54–56]. In this situation, a membrane is often removed during angle surgery, rendering goniotomy and trabeculotomy highly successful [3–6]. Chronic inflammation also causes synechiae formation and a closed-angle configuration. In some cases, goniosynechiolysis may be employed to reopen the angle, but in others, either glaucoma drainage device implantation or trabeculectomy is a better option to obtain IOP control [57–66]. It is also important to remember that local steroids needed for inflammation control can raise eye pressure [67–69]. Aggressive use of systemic steroid-sparing therapy in order to taper off of local steroids may be required. Regardless of the mechanism, control of the uveitis is paramount for treating the glaucoma. Any consideration of IOL implantation should occur only following both inflammation and IOP control.

Case 2

The patient is a 20-year-old woman with a history of idiopathic uveitis diagnosed at 4 years of age. Her uveitis was controlled with topical steroids, and she did not require steroid-sparing therapy. She was successfully tapered off of topical steroids at 15 years of age without uveitis recurrence. The patient underwent cataract extraction without IOL placement in both eyes at 5 years of age, and glaucoma was diagnosed at 8 years of age. At 11 years of age, the right eye underwent a trabeculectomy with mitomycin C, which was complicated by a postoperative suprachoroidal hemorrhage. She then underwent two goniotomies of the left eye at 16 and 18 years of age. Her intraocular pressures in her left eye remained uncontrolled, and she was referred for further treatment.

At the time of presentation, the patient's best-corrected visual acuity was 20/100 in the right eye and 20/40 in the left eye. Her aphakic correction was +10.75 in the right eye and +10.00 in the left eye. The intraocular pressures by Goldmann applanation tonometry were 8 mmHg in the right eye and 20 mmHg in the left eye. The patient was on timolol, dorzolamide, brimonidine, and bimatoprost in the left eye and oral acetazolamide. Slit lamp examination showed an avascular, thin, and cystic bleb at the superonasal limbus in the right eye. Both corneas were thin and clear. The anterior chambers were deep and quiet. The eyes were aphakic with synechiae between the iris and the Soemmering ring. Fundoscopic examination showed pale optic nerves with a cup to disc ratio of 0.95 in both eyes. The macula in the right eye lacked a foveal light reflex. The macula, vessels, and retinal periphery of the left eye were normal.

For the increased IOP in the left eye, the patient underwent pars plana placement of a Baerveldt® 350 glaucoma drainage device with concurrent vitrectomy. Following surgery, the patient's intraocular pressure in the left eye ranged from 6 to 10 mmHg off of all glaucoma medications.

At 25 years of age, the patient desired placement of IOLs. Her visual acuity had remained stable. Slit lamp examination showed a stable trabeculectomy bleb superonasally in the right eye and a large bleb over the superotemporal Baerveldt® plate in the left eye. Intraocular pressures by Goldmann applanation tonometry were 12 mmHg in the right eye and 7 mmHg in the left eye on no glaucoma medications. Axial lengths measured 23.6 mm in the right eye and 23.4 mm in the left eye, and keratometry was 42.15 × 44.91 @ 38 in the right eye and 42.74 × 44.40 @ 87 in the left eye. The patient desired a mild myopic target. In the right eye, the Soemmering ring was opened and debulked, and a three-piece acrylic lens was placed in the capsular bag. In addition, a subconjunctival injection of mitomycin C (0.2 mg/ml) was administered posterior to the trabeculectomy bleb, and the bleb was needled to remove surrounding Tenon's encapsulation. In the left eye, the Soemmering ring was opened and debulked, and a three-piece acrylic lens was placed in the capsule.

At final follow-up at 27 years of age, the patient's best-corrected visual acuity was 20/125 in the right eye and 20/40 in the left eye. Her refraction was −2.00 in the right eye and −3.50+1.50 × 105 in the left eye. Intraocular pressures by Goldmann applanation tonometry were 10 mmHg in the right eye and 8 mmHg in the left eye off of all glaucoma medications. Slit lamp examination showed a diffuse, mildly elevated trabeculectomy bleb at the superonasal limbus of the right eye and an elevated bleb over the Baerveldt® plate superotemporally in the left eye. The corneas were clear, and the anterior chambers were deep with no evidence of uveitis. In both eyes, the IOLs were in good position within the capsules (Fig. 22.2a). The optic nerves, retinal nerve fiber layer thicknesses (Fig. 22.2b), and visual fields were stable (Fig. 22.2c).

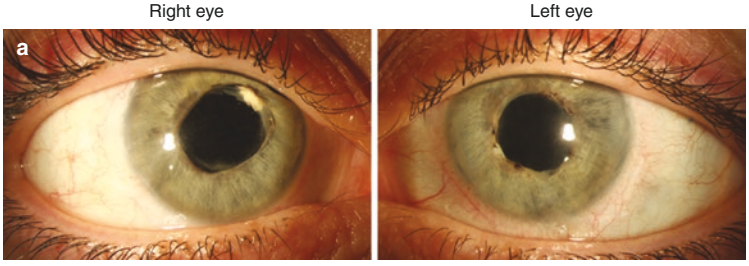

Fig. 22.2 Case 2. (**a**) External photographs of the right and left eyes presented in Case 2 demonstrated IOLs placed within the capsule. There was evidence of synechiae that were lysed with debulking of the Soemmering ring. The superonasal trabeculectomy bleb in the right eye and the superotemporal patch graft in the left eye were covered by the upper lids. The tube in the left eye was in the pars plana and so not evident in the photograph. (**b**) Optic nerve photographs demonstrated pale optic nerves with 0.9 cup to disc ratio in both eyes. Optical coherence testing showed that the retinal nerve fiber layer thickness was stable before and after IOL placement. (**c**) Goldmann visual field testing demonstrated a temporal island with loss of central fixation in the right eye and an inferior arcuate defect in the left eye

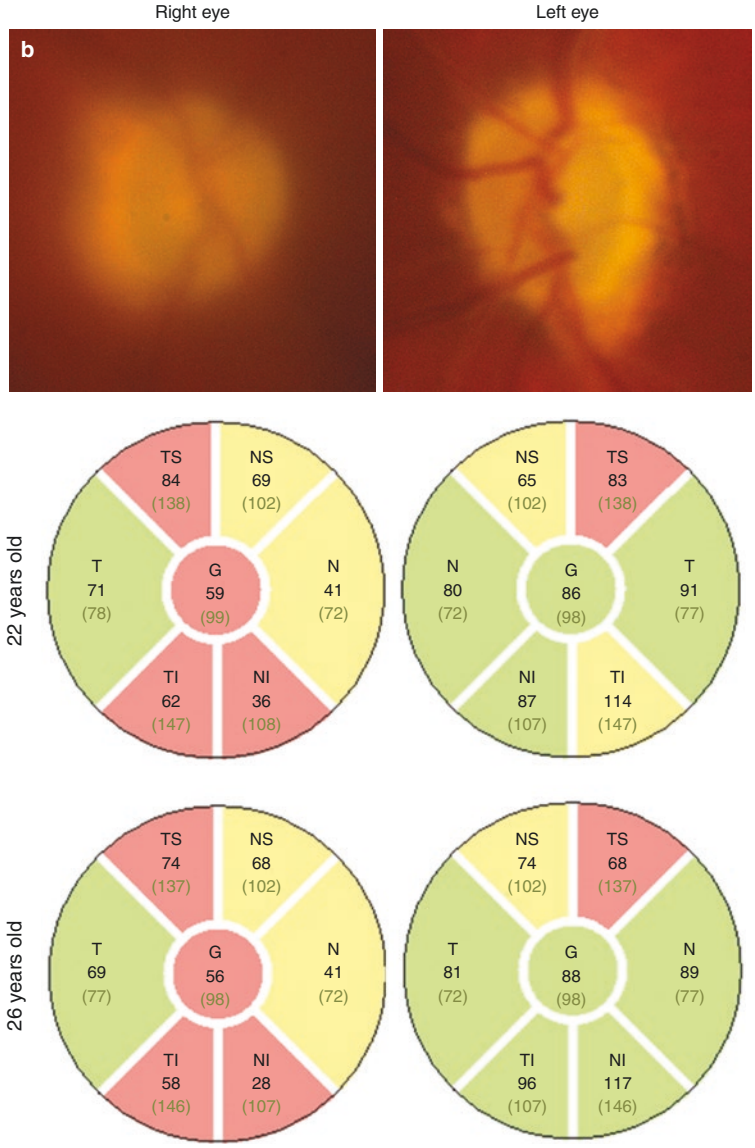

Fig. 22.2 (continued)

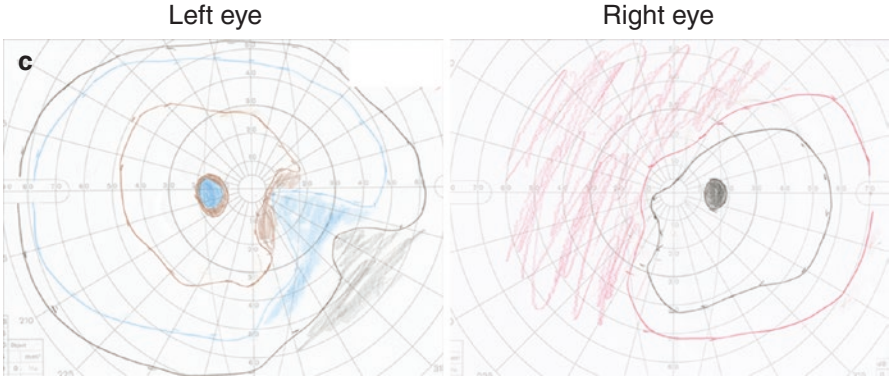

Fig. 22.2 (continued)

Comment Placement of a secondary IOL in an eye with uveitic glaucoma requires inflammation and IOP control. Unlike cataract surgery, which has to be done to improve vision, secondary IOL implantation is elective and should only be pursued if the uveitis has become quiescent [70]. In pediatric cases of uveitis, this typically occurs after puberty and is established when the patient has been successfully tapered off of all steroid and steroid-sparing therapies. Even though the uveitis has been inactive, the patient should be treated with oral steroids for 3–5 days prior to and following surgery to suppress the anticipated higher inflammatory response.

There are additional considerations in uveitic eyes that have undergone glaucoma surgery. Similar to eyes with glaucoma following cataract surgery, in eyes that have undergone angle surgery for uveitic glaucoma, attention should be paid to maintaining the open-angle configuration and cleft patency [3–6]. This includes placement of the IOL within the capsule and treatment with oral and topical steroids to minimize the postoperative inflammation and synechiae formation. The surgeon should be aware that the robust postoperative inflammatory response can result in hypotony, bleb flattening, and subsequent bleb failure. In eyes that have previously undergone trabeculectomy, the incisions for the IOL placement should be placed temporally to avoid injury to the superior bleb. Case 2 illustrated how a subconjunctival injection of mitomycin C posterior to the existing bleb and removal of Tenon's encapsulation tissue are important in preventing postoperative bleb failure. The bleb should be monitored carefully, and additional subconjunctival antifibrotic injections and bleb revisions may be needed to salvage flow through the trabeculectomy and maintain bleb morphology [71, 72]. As stated, in eyes with glaucoma drainage devices, the ideal position for the tube is within the pars plana as placement of the IOL will not interfere with tube function. A previously placed tube within the anterior chamber may be moved posteriorly concurrent with vitrectomy and IOL placement [33]. Although less important in uveitis compared to microphthalmia, posterior placement of the tube prevents corneal decompensation [33–35]. Thus, in uveitic glaucoma, IOLs should be placed within the capsule to

prevent reactivation of inflammation. Additional procedures may be required in eyes, which have had glaucoma surgery, especially trabeculectomies and glaucoma drainage devices.

Intraocular lenses can be safely placed in the setting of glaucoma, but attention needs to be paid to a number of special considerations. The intraocular pressure must be well-controlled prior to IOL placement; this is for preserving vision as well as for accurate lens calculations in young children. In eyes with glaucoma following cataract surgery, the angle and size of the anterior chamber should be evaluated. In uveitic glaucoma, the IOL is ideally placed within the capsule to prevent reactivation of the inflammation. In both of these clinical scenarios, prior glaucoma surgeries may dictate the safest location for the incision for IOL placement, and additional procedures may be required to maintain pressure control. While the goal of lens placement is to improve visual function without contacts or glasses, it is essential that intraocular pressure control not be compromised.

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Chapter 23
Pediatric Cataract Surgery in the Abnormal Anterior Segment

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An abnormal anterior segment can create many difficulties in pediatric cataract surgery. Issues most commonly encountered include corneal opacities, iris abnormalities, large or small anterior segments, and eyes with prior corneal or glaucoma surgery. This chapter will describe techniques to address the most common scenarios in the above categories.

Corneal Opacities

Corneal opacities seen in association with a number of conditions pose a challenge to the pediatric cataract surgeon. Causes of corneal opacification that are common in the context of pediatric cataract include acquired causes such as traumatic or infectious corneal scars, failed corneal grafts, and congenital causes such as Peters anomaly and endothelial corneal dystrophies. It may be beneficial to perform keratoplasty prior to cataract surgery or perform the procedures at the same time (triple procedure). Such procedures will typically be performed by a corneal surgeon, and the details are out of the scope of this discussion. However there may be situations where it is desirable to avoid keratoplasty due to a high risk of graft rejection/failure or when it is prudent to observe the visual results of cataract surgery first. In such situations, it is helpful to have techniques to improve visualization in the anterior chamber.

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Adjusting the patient positioning and the tilt of the operating microscope before the case may improve the surgical view in many of these cases. Some operating microscopes also allow adjustment of the intensity of oblique field and coaxial illumination in order to improve visualization during different surgical steps. This feature may also be useful when there is an underlying poor red reflex from vitreous abnormalities, such as a tumor and vitreous hemorrhage, or in a vitrectomized eye. With focal opacities or scars, simple rotation of the eye with grasping forceps at the limbus can improve visualization of instruments in the anterior chamber. Parallax induced by such movements also helps in the judgment of the depth of an otherwise obscured feature in the anterior chamber. Trypan blue dye improves visualization of the capsule but may also worsen the view in the context of a poor corneal endothelium as it stains damaged endothelial cells [1]. It should be used judiciously, with the smallest volume needed to produce good contrast. Staining of the endothelium is minimized with the use of an injection of an air bubble or viscoelastic first, but in the latter technique, care needs to be taken to make sure that trypan blue contacts the lens capsule directly or else there will be poor staining. Triamcinolone is used to visualize any vitreous in the anterior chamber, but its use should also be judicious as a large volume of particulate matter can worsen the surgeon's view. During capsulorrhexis, the oblique field illumination may be decreased to improve the red reflex (especially in eyes with prior vitrectomy or compromised zonules).

A very helpful adjunct in pediatric cataract cases with corneal opacities is the use of an endoilluminator, which is a tool commonly used in vitreoretinal surgery (see Case 2). These endoilluminators can come standard in vitrector sets and are especially useful for junior surgeons. They can provide oblique illumination if held near the limbus outside the eye or more effectively can be used inside the eye to tangentially illuminate the field, forming shadows that enhance depth perception [2]. This may require an additional limbal incision. It is helpful to decrease the illumination of the operating microscope when the endoilluminator is used to reduce the light scatter off the opacity back to the surgeon.

Case 1: Pediatric Cataract Surgery Following Penetrating Keratoplasty

A 15-year-old girl with a history of bilateral Peters anomaly with bilateral sequential penetrating keratoplasties at age 2-3 months presented with decreased vision in her right eye. Preoperative examination indicated visual acuity of hand motions in the right eye, horizontal nystagmus, normal intraocular pressure in both eyes, clear corneal grafts, and a white cataract in the right eye. A preoperative B-scan ultrasound demonstrated an attached retina in the right eye. The corneal diameter was 10.5 mm. The family and patient wished to proceed with cataract surgery in the right eye. During the surgery, microscissors were first used to cut iridocorneal adhesions, and then the synechiae between the iris and lens were carefully dissected using micrograspers (Fig. 23.1). In addition to trypan blue for improved visualization, microscissors were needed to complete the capsulorrhexis due to capsular

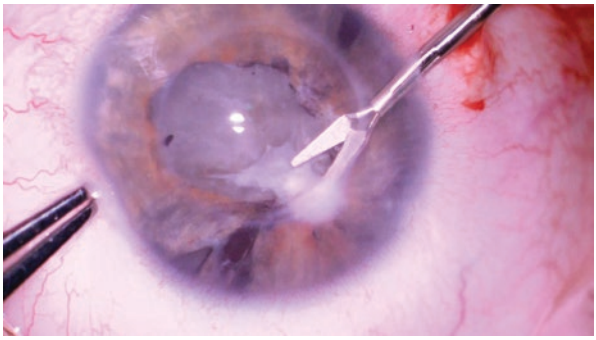

Fig. 23.1 Use of microscissors to cut iridocorneal adhesions during cataract surgery

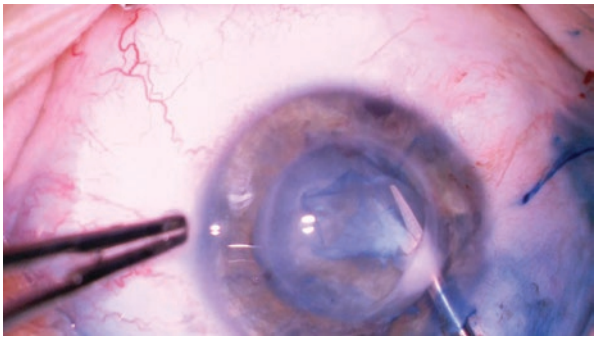

Fig. 23.2 Microscissors for completing capsulorrhexis in case of capsular fibrosis

fibrosis (Fig. 23.2). A one-piece intraocular lens was placed safely in the capsular bag, and the patient had a postoperative visual acuity of 20/250.

Comment The above case illustrates a case of cataract following a penetrating keratoplasty. Careful surgical technique with judicious use of trypan blue allowed for successful creation of a capsulorrhexis. The surgeon was attentive to not using too much trypan, which would compromise the integrity of the corneal graft. Use of microscissors, an instrument not conventionally used for creation of a rhexis, allowed for an opening to be created in the setting of fibrosis of the anterior capsule. Successful in-the-bag implantation of an IOL was possible, and postoperative visual acuity was significantly improved.

Iris Abnormalities

In the case of a cataract following a corneal laceration, there is often an organized membrane from the iris to the old corneal wound, and this scenario may require use of an OVD or microscissors to sever the adhesions and revise the wound prior to addressing the cataract [3, 4]. Similarly, OVD and microscissors are used to break lens-iris adhesions in Peters anomaly. Peripheral anterior synechiae in uveitic or glaucoma patients may also require synechiolysis with OVD during cataract

surgery. Only judicious pushes of OVD are required to achieve maximum force in breaking these adhesions. The use of additional OVD will not increase efficiency and may inadvertently induce reverse pupillary block, increase IOP, or cause iris prolapse. If OVD is unsuccessful, then mechanical means including a cyclodialysis spatula or microscissors are indicated.

The pupil may be persistently miotic in certain scenarios, such as trauma, Marfan syndrome, uveitis or in association with congenital cataract. In these cases, mydriasis may be achieved by using the same array of tools as in adult cataract surgery including iris hooks, ring devices, OVDs, and preservative-free epinephrine in irrigation fluid [4–6]. Care should be taken with the use of ring devices in an eye that is microphthalmic due to size or where a posterior capsulotomy and anterior vitrectomy are to be performed due to risk of posterior dislocation.

Hypoplastic iris tissue that is floppy and easy to tear can be encountered in congenital cataract cases especially in anterior dysgenesis syndromes like Axenfeld-Rieger and congenital rubella [7, 8]. Care must be taken in these cases to disturb the iris tissue as little as possible to avoid pigment loss and intraoperative iris prolapse. The most common cause of iris prolapse in these patients is overfilling the anterior chamber with OVD when not taking into consideration the smaller volume of the pediatric anterior chamber.

Case 2: Traumatic Cataract Following Open-Globe Injury

A 9-year-old boy presented 6 months after injury to the right eye with a pipe and primary corneal laceration repair done overseas. Visual acuity was count fingers at 2 feet. There was a corneal scar with neovascularization and a loose exposed suture, iris-cornea touch, retained cilia in the anterior chamber, inferotemporal iridodialysis, and a mixed anterior capsular and nuclear sclerotic cataract. In the operating room, the loose suture and intraocular cilia were first removed with forceps (Fig. 23.3). Iris tissue adherent to the corneal scar was dissected using OVD and microscissors. An endoilluminator and movement of globe to induce parallax were used to improve the view of the anterior chamber and facilitate cataract removal

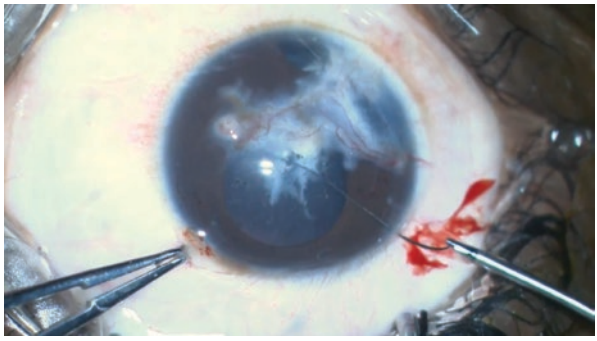

Fig. 23.3 Removal of intraocular cilia during surgery for traumatic cataract

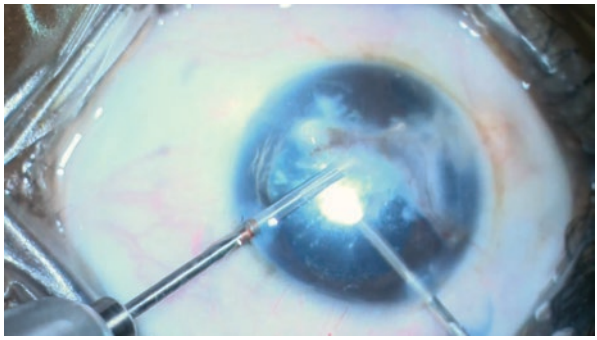

Fig. 23.4 Endoilluminator for improved view during lensectomy for traumatic cataract with corneal scar

(Fig. 23.4). An intraocular lens was placed in the sulcus with posterior optic capture with a power based on intraoperative A-scan of the operated eye and the keratometry values from the fellow eye (the corneal scar precluded keratometry in the operated eye). Postoperatively, the manifest refraction in the operated eye was $-1.00 + 1.50 \times 050$ with a visual acuity of 20/32–2 despite the corneal scar at 1-year follow-up.

Comment Globe trauma and the resultant cataracts can have very poor visual prognosis. Corneal scars and zonular weakness are just two of many reasons the surgical removal of lens opacities can be complex and challenging. That said, when they occur in an older child outside of the amblyogenic age range, the surgeon has the potential to greatly improve vision. Appropriate preoperative planning and strategic intraoperative maneuvers are important. This case introduces the reader to the use of an endoilluminator for improved visualization when corneal scar prevents clear visualization of all lens quadrants.

Abnormally Large or Small Anterior Segment

An abnormal anterior segment may involve an anterior chamber deeper or shallower than the cataract surgeon is accustomed to. In small[1] eyes, attention should be given to identifying the limbus carefully as limbal incisions tend to be positioned too

[1] The classification of small eyes is confusing [9]. Simple microphthalmia is defined as having a short axial length less than 2 standard deviations below the normal for age, typically 17.8 mm in infants and 20.5 mm in adults with no other malformations. Complex microphthalmia indicates the presence of microphthalmia with additional malformations such as iris coloboma, chorioretinal coloboma, persistent fetal vasculature, or retinal dysplasia. Relative anterior microphthalmos is a normal length eye with an abnormally small anterior segment [9, 10]. Microcornea is defined as a corneal diameter of less than 9 mm in an infant and 10 mm in an adult. Microcornea may be a feature of microphthalmic eyes, nanophthalmic eyes, and sometimes even long, myopic eyes [11]. Nanophthalmia is a short eye with a small anterior segment and thick sclera and choroid; there is no agreed-on axial length cutoff for nanophthalmic eyes [10].

anteriorly, increasing visible scarring and making access to the lens more challenging. Sometimes transillumination with an endoilluminator or the use of ultrasound biomicroscopy (UBM) may be helpful to mark the limbus when it is indeterminate. In eyes with a small anterior chamber, the corneal thickness is often greater, and limbal incision with a typical entry angle will form an incision that is too long [10]. A scleral tunnel may be used as an alternative to a limbal incision if an IOL is to be placed. When considering an IOL in a small eye, the surgeon should consider whether the eye is large enough to safely fit the IOL, whether an IOL with sufficient power is available, and the reduced accuracy of IOL calculations for these cases [10]. Leaving the child aphakic and using an aphakic contact lens or spectacles is the preferred approach if there is any question as to whether the small eye can fit an IOL. Corneal diameters may be measured using the traditional white-to-white measurement with calipers, optical biometry, or UBM [12, 13]. Postoperative care is frequently complicated by glaucoma in these cases.

In microspherophakia, weak zonules induce an increased curvature of the lens and a challenge for the cataract surgeon. The increased lens curvature causes a shallow anterior chamber with associated risk of pupillary block as well as high lenticular refractive power [14–16]. In microspherophakia, additional viscous OVD will be necessary to deepen the shallow anterior chamber. A capsular or iris hook at the capsulorrhexis margin may assist in performing the rhexis as the capsule has little zonular support [17]. Capsular tension rings or segment may be used to stabilize the lens and capsule, although the lens diameter is often too small to fit these devices and the IOL [18]. In some cases, it may be best to remove both the lens and the capsule completely if the zonules are so loose as to make the capsule useless as a method of supporting an IOL. IOL calculations in microspherophakia may be inaccurate because of postoperative changes in effective lens position, and the small size of the capsular bag may make it difficult to place an IOL within the bag [14].

In a large eye with a deep anterior chamber, the IOL may become easily decentered due to the large capsular bag and ciliary ring. An anterior capture of a three-piece IOL with the optic located inside of the capsular bag and the haptics located in the sulcus can provide the best centration of the optic [19]. The haptics in the sulcus stabilize the optic rather than relying on zonules that may be stretched and weak to support an in-the-bag IOL. Axial myopia also increases the uncertainty of IOL power calculation and increases risk of a hyperopic surprise, but formulas such as the Barrett Universal II and Haigis have shown excellent performance in eyes with axial length greater than 26 mm [20].

Eyes with Prior Glaucoma/Corneal Surgery

In cataract cases in which the eye already has a previously placed glaucoma drainage device, a few techniques may be used to avoid complications. The tube and patch graft area should be avoided when making the cataract surgery incisions. At the end of the case, the tube should be primed with BSS to ensure no OVD remains inside the tube. A low-flow technique should be used with the vitrector to avoid

massive subconjunctival chemosis from the bleb which can obscure the view. Some surgeons advocate tying off the tube, but in our experience, with low-flow techniques, this is not needed.

In the context of a graft or cornea with limited endothelial cells, reducing the intraocular pressure and total amount of fluid irrigation through the eye is also prudent to minimize further endothelial damage. A dispersive OVD should be used to protect the endothelium during cataract removal, and a cohesive OVD should be used during IOL implantation for its ease of removal. Minimizing wound leaks around incisions decreases the total volume of fluid used.

In summary, an eye with an abnormal anterior segment certainly makes pediatric cataract surgery more complex, but with appropriate planning and creativity, these challenges can be met. This chapter has presented what in our experience are the most common issues we have faced in these eyes and the techniques we have found most useful in addressing them.

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Chapter 24
IOL Placement in Developmentally Delayed Patients

Andrew Robert Lee

Introduction

Although cataracts in the pediatric population are typically idiopathic, they can also often occur in association with systemic disease. Common systemic diseases and syndromes associated with cataracts include Down syndrome, Lowe syndrome, galactosemia, intrauterine infection including TORCHS infections, Sturge-Weber syndrome, and Nance-Horan syndrome [1, 2]. Children with these disorders commonly have developmental delay, and management of patients with cataracts and developmental delay poses unique challenges at all phases of care. Preoperative assessment is frequently limited, and patients with systemic abnormalities or developmental delay have higher reported rates of complications after cataract surgery [3] and poorer visual outcomes [4].

Preoperative Assessment

The preoperative assessment of patients with developmental delay and cataracts is uniquely challenging. Assessment of visual acuity is often limited by nonverbal status and lack of sustained attention or cooperation. When optotype visual acuity cannot be obtained, preferential-looking tests such as Teller or Cardiff acuity testing should be used to assess visual acuity [5–7]. If cooperation is inadequate to perform a preferential-looking test, then assessment of fixation preference using the induced tropia test can assess for amblyopia [8, 9]. Pattern visual evoked potential can also

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assess for abnormal vision in patients with congenital cataracts [10]. Other clinical findings such as nystagmus or monocular strabismus in the cataractous eye may also provide evidence for the visual significance of a cataract.

In patients where a cataract is suspected, examination of the lens using an upright or handheld slit lamp and a retinoscope should be performed with careful attention to the location and size of the opacity. A central opacity of greater than 3 mm places the patient at higher risk for visual impairment [11, 12]. Cycloplegic refraction should be performed given the high incidence of astigmatism and anisometropia associated with lenticular abnormalities, particularly anterior polar cataracts [13]. In some cases, examination under anesthesia may be required in order to determine if a cataract is visually significant enough to warrant extraction.

Non-surgical Management

The etiology of visual impairment in children with cataracts may have multiple components including direct obstruction of the visual axis, refractive error, and amblyopia, which may be deprivational, anisometropic, refractive, and/or strabismic. In cases of partial cataract where the visual significance is unclear, non-surgical management including spectacle correction, part-time occlusion, and pharmacologic dilation should be attempted [11, 14, 15]. Close follow-up with frequent reassessment of visual function is necessary. For developmentally delayed patients, tolerance of glasses or patching should be assessed preoperatively as this will play a role in intraocular lens (IOL) selection. In cases where there is poor adherence to prescribed treatment, the surgeon may consider adjusting the postoperative refractive target in order to minimize initial refractive error, which may be amblyogenic if uncorrected.

Surgical Planning: Placement of an Intraocular Lens

If the decision is made to perform cataract extraction in a patient with developmental delay, several surgical considerations should be reviewed prior to surgery and discussed with the patient's parents or legal guardian.

One of the most important decisions when planning pediatric cataract extraction is whether to leave the eye aphakic, requiring contact lens correction (see Chap. 11 "Contact Lenses") or aphakic glasses (see Chap. 12 "Aphakic Glasses"), or to implant an IOL. The Infant Aphakia Treatment Study found similar visual acuities, but higher rates of adverse events and additional surgeries in infants aged 0–6 months who received an IOL as compared to those who were left aphakic [16]. Thus the recommendation from the study was that primary IOL implantation in infants <7 months of age should only be performed when contact lens wear might be especially challenging, risking periods of uncorrected aphakia [12]. Patients with developmental delay frequently fall into this category, and thus IOL implantation may be considered in this population at a younger age. Primary IOL implantation in

children has increasingly become more common in children over 1 year old, and for children who were initially left aphakic, secondary IOL implantation can be considered as early as 2–4 years of age when the rate of growth of the eye has slowed or aphakic contact lens or spectacle correction has become more difficult [12].

If an IOL is implanted in a child with developmental delay, postoperative refractive target must be carefully considered. Because growth of the eye during childhood typically results in myopic shift, many established recommendations target hyperopia in order to avoid significant myopia in the long term [17, 18]. However, in patients with poor compliance with spectacle correction, initial high hyperopia creates the risk of uncorrected refractive error and, in cases of unilateral pseudophakia, anisometropia. If there is poor compliance with spectacle correction and amblyopia treatment, then refractory or worsened amblyopia may result. Poor compliance with patching has been reported as the factor most strongly associated with poor visual acuity 7 years following cataract surgery in children [19]. Although published studies vary as to how important postoperative refraction is to visual outcomes [20, 21], if the patient's parents or guardians have significant concerns about compliance with spectacle correction or patching prior to surgery, then a refractive error closer to emmetropia should be considered in order to minimize the risk of amblyopia. If significant myopia or anisometropia develops later, then possible strategies may include contact lens correction, IOL exchange [22], corneal refractive surgery, piggyback intraocular lens, or implantable contact lens [23].

Intraoperative Technique

Patients with developmental delay undergoing cataract surgery are at higher risk for postoperative complications [3] and worse visual outcomes [4]. Published rates of surgical complications in children with Down syndrome are higher in non-ophthalmic surgery as well [24, 25]. Furthermore, self-injurious behavior in developmentally delayed patients also poses risks of ophthalmic injuries [26]. Administration of postoperative topical eye drops can be more difficult for caregivers of patients with developmental delay. Certain intraoperative techniques can help to minimize the risk of postoperative complications.

Scleral tunnel incisions should be considered particularly if a larger incision is required, as in the case of intraocular lens implantation (Fig. 24.1). With a scleral tunnel incision, if there is wound leak or dehiscence, there will not be direct communication between the anterior segment and the external environment. Scleral tunnel incisions have been found to have a lower risk of endophthalmitis than corneal wounds following adult cataract surgery [27, 28]. All wounds, including paracenteses, in pediatric cataract surgery should be sutured to minimize risk of wound leak [29]. Scleral wounds, if covered by conjunctiva, may be sutured with non-absorbable 10-0 Nylon suture or absorbable 8-0 or 9-0 Vicryl® (polyglactin, Ethicon, Johnson & Johnson, USA) (Fig. 24.1D). Corneal wounds should be closed with absorbable suture such as 10-0 Vicryl® in order to avoid the need for suture removal under anesthesia postoperatively (Fig. 24.2).

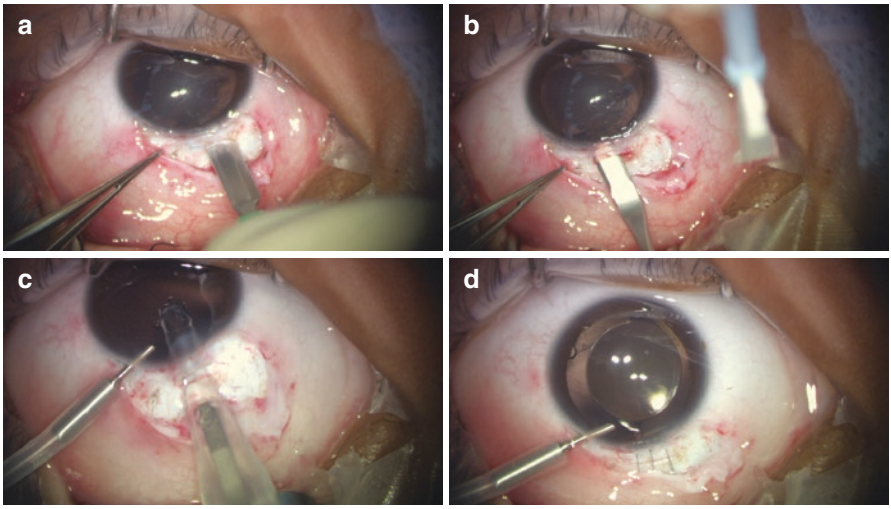

Fig. 24.1 Scleral tunnel incision. In a patient undergoing cataract extraction and IOL implantation, a 6 mm conjunctival peritomy is made (not pictured). A rounded tip blade is then used to make a 4-mm-wide partial-thickness incision into the sclera (**a**), after which a tunneling blade is used to tunnel into the peripheral cornea (**b**). After removal of the cataract, a keratome is used to open the wound into the anterior chamber (not pictured) and the IOL is injected into the anterior chamber (**c**). The wound is sutured with three interrupted 10-0 Nylon sutures (**d**)

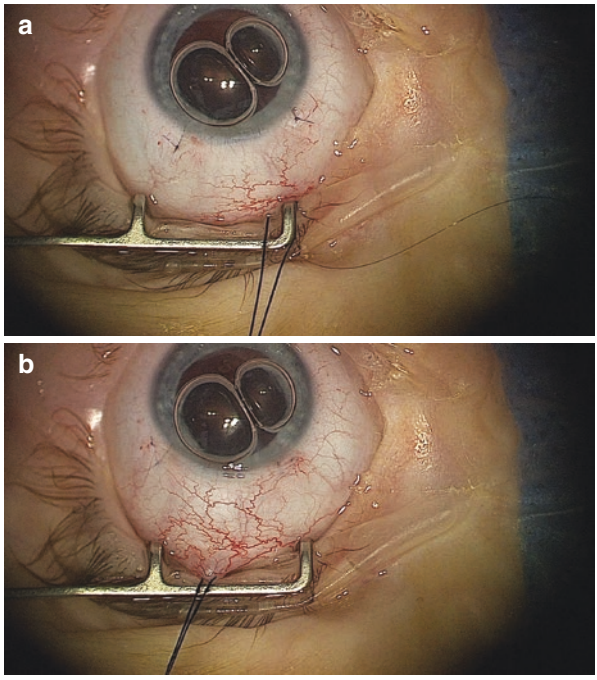

Fig. 24.2 Sutured limbal incisions. In a patient left aphakic, the limbal wounds are closed with 9-0 Vicryl sutures (**a**). Small conjunctival peritomies are closed over the wounds to minimize suture and wound exposure and improve comfort postoperatively (**b**). A filtered air bubble in the anterior chamber is used to improve anterior chamber stability and facilitate postoperative examination (**a**, **b**)

Intracameral or periocular medication delivery is especially valuable in developmentally delayed patients who may have difficulty with topical eye drop administration postoperatively. Intracameral triamcinolone in pediatric cataract surgery has been reported to reduce postoperative inflammation and visual axis obscuration without any increased rate of complication or intraocular pressure increase [30–34]. Intracameral cefuroxime [35, 36] and moxifloxacin [37, 38] have both been reported to be safe and effective in the reduction of post-cataract endophthalmitis in adults and, depending on availability, should be strongly considered in developmentally delayed children undergoing cataract surgery. Though data is limited regarding rates of endophthalmitis in patients with developmental delay, endophthalmitis remains a concern especially in patients who may be prone to eye rubbing or other high-risk behaviors. Postoperative endophthalmitis, while rare following intraocular surgery in children [39], can be a devastating, vision-threatening complication.

A small bubble of filtered air injected with a cannula into the anterior chamber at the end of surgery helps to promote anterior chamber stability (Fig. 24.2) [40]. Additionally, in patients who are uncooperative with examination on the first postoperative day, an air bubble is an easily visible indicator that the anterior chamber is formed and that the surgical wounds are not leaking significantly.

Postoperative Care

In the postoperative period, efforts should be directed toward avoiding trauma to the eye, such as rubbing and self-injurious behavior. In children with self-injurious behavior, a multidisciplinary approach is necessary in order to minimize the risk of postoperative injury or complication. Strategies can include behavioral modification strategies, as well as safety devices such as soft goggles and elbow restraints [26, 41]. Severely delayed or self-injurious children may require inpatient admission for sedation and observation during the critical few days immediately following surgery.

Immediately Sequential Bilateral Cataract Surgery

For some patients with bilateral cataracts and developmental delay, there may be significant concern about the medical risks of two episodes of general anesthesia within a short time frame. If such patients are at higher risk of anesthesia-related complications due to their comorbid systemic conditions, immediate sequential bilateral cataract surgery (ISBCS) can be considered [12, 42, 43]. It should be noted that there is a lack of consensus regarding the risks, benefits, and role for ISBCS. In particular, risks of potentially blinding bilateral complications such as endophthalmitis, toxic anterior segment syndrome, and expulsive hemorrhage must be considered. Thus, an extensive discussion with the anesthesiologist and caregivers is of

critical importance. If ISBCS is performed, extensive precautions in aseptic technique, including separate instrumentation, surgical prep, and surgeon sterility, should be implemented [12, 44].

Case 1

A 15-month-old male presented with intermittent crossing of the right eye for 1 month. His past medical history was significant for autism and developmental delay. He was nonverbal. On exam, he was able to perform Cardiff preferential-looking test, and his right eye has an acuity of 20/160 and his left eye 20/40. He has an intermittent right esotropia of 16 prism diopters with a strong left eye fixation preference. Portable slit lamp examination revealed a posterior capsular speckled opacity with mild obscuration of red reflex in the central 3 mm of the right eye. Left lens was clear. Cycloplegic refraction was +2.00 sphere in the right eye and +2.50 sphere in the left.

Due to the multiple possible mechanisms for amblyopia and esotropia, after discussion with the parents, the decision was made to attempt non-surgical management initially with patching of the left eye 4 hours per day. At 6-week follow-up, Cardiff visual acuity was unchanged. The right esotropia was now constant. The central red reflex in the right eye was more dull than it was previously. The decision was made to proceed with cataract extraction in the right eye. The parents expressed concern about contact lens placement due to the patient's developmental delay and intolerance of patching, and they elected to have an IOL implanted.

Cataract extraction with IOL implantation was performed on the right eye. A hydrophobic single-piece acrylic IOL was inserted through a superior scleral tunnel incision into the capsular bag with initial postoperative refractive target of +2.00. Posterior capsulotomy and anterior vitrectomy were performed via a pars plana incision. Prior to closing, cefuroxime and triamcinolone were injected intracamerally. The scleral tunnel was sutured with 10-0 Nylon, and the remaining incisions were closed with 9-0 Vicryl®.

Comment This case illustrates a number of the difficulties in managing cataracts in children with developmental delay. At the patient's initial visit, the patient had a lenticular opacity of borderline size for visual significance, and it was not immediately apparent whether the strabismus was secondary to the cataract or an independent contributory factor to the patient's amblyopia. Therefore, non-surgical management was initially prescribed. On follow-up, progression of the cataract, worsening of the control of the esotropia, and lack of improvement in vision all provided evidence for the visual significance of the cataract, and thus cataract extraction was recommended to the parents.

A thorough discussion with the patient's parents was essential to surgical planning. Because of concerns about aphakic contact lens insertion, the decision was made to implant an IOL. A postoperative refractive target of +2.00 was chosen in

order to minimize anisometropic amblyopia if the patient did not tolerate glasses or patching, with the parents understanding that he may become significantly myopic in the future.

Intraoperatively, the decisions to use a scleral tunnel (as opposed to a clear corneal incision), suture all wounds, and inject intracameral cefuroxime were intended to minimize risk of endophthalmitis, and intracameral triamcinolone was used to help control intraocular inflammation given concerns about compliance with drop administration postoperatively in a developmentally delayed child.

Case 2

A 17-year-old female with CHARGE syndrome presents for her first eye examination in several years. Her past ocular history includes myopia, bilateral iris and inferior retinal colobomas, and chronic total retinal detachment left eye previously deemed inoperable. Her medical history is notable for CHARGE syndrome, cardiac valvular abnormalities, and developmental delay. She is nonverbal.

She was able to do a Teller preferential-looking test and her right eye was 20/100. The left eye had no light perception. On slit lamp and fundus examination, her right eye had an inferior iris coloboma and 3+ posterior subcapsular cataract. There was an inferior retinal coloboma not involving the macula, with no retinal detachment. The left eye had an inferior iris coloboma; fundus exam confirmed total retinal detachment. Cycloplegic refraction in the right eye was −15.00 sphere; the left eye had a poor red reflex.

Cataract extraction with IOL implantation was recommended to the family because of the visually significant cataract in the right eye. At home, the patient does not wear glasses and frequently uses an electronic tablet for communication. Outside the home, she does wear glasses and enjoys looking at distant objects. Therefore, it was decided to target a postoperative refraction of −2.00. Cataract extraction was performed in similar fashion as in Case 1. However, because of her history of retinal detachment in the fellow eye, the posterior capsule was left intact, and no vitrectomy was performed.

Postoperatively, refraction in the right eye was −2.25. Teller visual acuity was 20/40. The parents found the child to have improved visual alertness around the house.

Comment In this case, the visual significance of the cataract in the right eye was clear. Due to the patient's monocular status and developmental delay, an extensive discussion with the patient's parents regarding the risks, benefits, and alternatives of surgery was essential. As in Case 1, a clear understanding of the patient's activities of daily living and tolerance of glasses assisted in the preoperative planning. Because the patient did not wear glasses at home and performed frequent near work with an electronic tablet, the postoperative refractive target was −2.00, with plans to prescribe myopic spectacle correction for distance when the patient is outside the

house. Because of the patient's older age, concern for postoperative visual axis obscuration was lower. Therefore, in light of the patient's inferior retinal coloboma and history of retinal detachment in the fellow eye, the decision was made to not perform posterior capsulotomy or anterior vitrectomy in order to minimize any potential risk of retinal detachment in the patient's only seeing eye.

The management of cataracts and IOL placement in children with developmental delay poses numerous difficulties. Preoperatively, cooperation and examination are frequently limited; postoperatively, complication rates may be higher, and adherence to treatment may be poor. Thorough and careful examination, extensive discussion with the patient's parents or guardians, thoughtful preoperative planning, and the adoption of certain intraoperative techniques can increase the likelihood of positive outcomes in this challenging patient population.

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Chapter 25
Pediatric Cataract Surgery in the Developing World

Lee M. Woodward and Amadou Alfa Bio Issifou

Recent public health initiatives have decreased the number of children who become blind from measles and vitamin A deficiency [1–3]. As a result, cataracts have become a leading cause of childhood blindness in the developing world. It is estimated that cataracts are responsible for 5–20% of pediatric blindness worldwide, with the prevalence of blindness due to childhood cataract being as much as 10 times higher in low-income economies compared with high-income economies [4–6]. Cataracts are a potentially curable form of blindness with timely identification, prompt surgery, and proper post-operative treatment of refractive error and amblyopia. However, limitations to obtaining timely surgery make cataracts a cause of irreversible blindness in children in the developing world.

The management of pediatric cataracts in the developing world has many unique considerations and challenges. The infrastructure needed to overcome these challenges is often very different from the developed world. Collaborative efforts from health-care providers and public health workers are needed to promote early detection, obtain cost-effective resources, and ensure proper post-operative follow-up. Surgical technique is often modified to adapt for limited surgical equipment and supplies.

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Delayed Presentation

Early detection and timely surgery are critical for successful outcomes in childhood cataract surgery. An unobstructed visual axis is needed to stimulate visual development. A delay in clearing the visual axis can lead to untreatable amblyopia. Long travel distances and improper awareness of the urgency of a lens opacity create delays in developing countries. Mwende and colleagues found the mean time between recognition of the cataract by the caregiver and presentation to the hospital was 34 months in Tanzania [7]. Similar studies in India and China found a mean age at surgery for congenital cataract of 27.6 months and 48.2 months, respectively [8–9]. Long delays in presentation were found to be associated with having progressive cataracts, living far from the hospital, and low socio-educational status of the mother [7]. Congenital bilateral cataracts present with less delay as they tend to be more severe, and the associated nystagmus alerts the caregiver to the presence of a vision-related problem.

Red reflex screening programs for early cataract detection are not as widespread or reliable in developing countries. The cost and training of primary health workers make such programs difficult to implement and sustain. Key informants have been used as a cost-effective way to promote awareness and improve early detection. Key informants are respected members of a community trained to detect vision loss in children in their local population. They have proven effective in different areas of the world in identifying and referring children to appropriate surgical centers [10–12]. Despite efforts made in community awareness, delayed presentation continues to be an ongoing struggle preventing timely cataract surgery in children.

Facilities and Personnel

To address the problem of childhood blindness in the developing world, the World Health Organization and the International Agency for the Prevention of Blindness recommend that there be one Child Eye Health Tertiary Facility (CEHTF) per ten million people [13]. These facilities should be capable of treating complex pediatric eye conditions, including cataracts. Ideally, they should provide optical, low-vision, and pediatric anesthesia services. It is estimated that only about 28 of these facilities exist in sub-Saharan Africa, which has a total population of just over 1 billion [14].

Children typically travel long distances to reach these tertiary centers. As travel back and forth between the home and center is often not practical, children are typically admitted to an inpatient ward during the entire surgical process, including immediate post-operative care. The wards are staffed with ophthalmic nurses trained in pediatric eye care. Trained pediatric ophthalmologists perform surgeries while utilizing pediatric anesthesia services. Surgical staff with knowledge of assisting in pediatric surgery, sterilization procedures, and equipment maintenance are critically important. Ophthalmic assistants and low-vision specialists provide pre-operative screening and post-operative care including refractions (see Fig. 25.1). Childhood

Fig. 25.1 Low-vision specialist performing post-operative refraction

Table 25.1 CEHTF personnel

Pediatric ophthalmologist
Pediatric anesthetist
Low-vision specialist/optometrist
Ophthalmic nurse
Surgical assistant
Childhood blindness coordinator and counselor

blindness coordinators are helpful with pre-operative counseling, tracking patient demographic data, facilitating future travel for follow-up care, and assisting with obtaining special educational needs. Table 25.1 lists the team members at a typical CEHTF in the developing world.

Surgical Equipment and Supplies

Economic constraints limit the availability of surgical equipment and supplies in the developing world. The cost to maintain or update equipment and replenish consumables is often too high to sustain pediatric cataract services. A cost analysis at two CEHTFs in Malawi and Zambia found the equipment costs at $178,121 and $179,832 [15]. Taking into consideration labor, consumables, and medications, the total cost for pediatric cataract surgery was $689 per child in Malawi and $763 per child in Zambia. While these costs are low compared to higher-income countries such as the United States [16], funding is largely dependent on donors who are often limited and inconsistent. Furthermore, the cost related to surgical equipment and

Table 25.2 Surgical equipment and consumables

Equipment and instruments	Consumables
Vitron 2020 (Geuder) pneumatic vitrectomy unit including tubing, handpiece, and AC maintainer	Intraocular lens – single-piece foldable acrylic or PMMA (Aurolab)
Scan Optics SO-161-R operating microscope	9-0 nylon suture
15 blade	Hydroxypropyl methylcellulose
Keratome	Triamcinolone and intracameral cefuroxime
MVR blade	

supplies can be substantially higher for pediatric surgery compared to that of adult surgery. Specifically, the costs of a vitrector, anesthesia machine, and foldable intraocular lenses are higher than instrumentation used for adults but are essential for pediatric cataract surgery [17].

To perform surgery within the economic constraints, older models and second-hand equipment are often used. Adaptations in surgical technique can be made to work with limited supplies and consumables. Table 25.2 lists suggested equipment and supplies.

Post-operative Management

Surgery is only the first step in a long process of restoring sight to a child with cataracts. After the natural lens is removed, the image a child sees must be focused on the retina to stimulate visual development in the brain. In an ideal setting, this is achieved with spectacles or contact lenses. However, these are not practical options in much of the developing world. The costs of continuously updating glasses or contact lens strength as the eye grows and replacing broken or lost ones make them an unreliable option. Poor hygienic living conditions and lack of freshwater to properly care for contact lenses also make them a high-risk option. This is why placing an intraocular lens, whenever possible, is critically important in children undergoing cataract surgery in the developing world. Uncorrected aphakia in this setting is often no better than the cataract itself in regard to visual outcomes.

Continued post-operative care with follow-up examinations is critical for maintaining updated refractions and to monitor for associated complications, such as glaucoma, strabismus, and visual axis opacifications. Long travel distances and the cost of travel make follow-up care challenging in the developing world. A childhood blindness coordinator can promote good follow-up through the use of cell phone reminders, patient tracking, and reimbursement for transport [18].

All the above limitations in the developing world affect surgical technique and decision-making. Preferred surgical methods are influenced by cost and possibilities of follow-up. We provide an extreme example that is influenced by cost-effectiveness. Our method may vary depending on location and availability of resources.

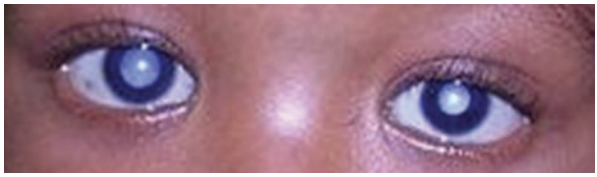

Fig. 25.2 An 8-month-old with bilateral cataracts

Case 1

An 8-month-old child presents with bilateral congenital cataracts (see Fig. 25.2). The mother noted nystagmus around 3 months of age. A key informant in the community alerted the mother of a potentially serious vision problem with her child that required medical attention. The mother brought the child to a local health worker where immunizations were provided. This led to referral to a pediatric surgical outreach in Mwanza, Tanzania. The outreach team was composed of members from two different CEHTFs in Tanzania. The mother and child traveled for 10 hours by bus to reach the center.

On examination, both lenses were white and opaque with no view to the fundus. The child appeared to have light perception vision in each eye, but no fixation or tracking of objects. The child otherwise appeared healthy and well-nourished.

Comment After examination, the mother was sent for counseling with the childhood blindness coordinator. The mother was educated on risks and benefits of the surgery, including the urgency of the situation in order to best stimulate the child's visual development. She expressed understanding of the logistics of and rationale for the surgery, including the importance of follow-up examinations and optical rehabilitation. The child was then admitted to pediatric eye ward and put on the list for next available surgery, which was anticipated to be 3–4 days later. Prior to surgery, the child was evaluated by the anesthesia team and deemed healthy enough for general anesthesia.

Immediate sequential bilateral cataract surgery was planned. Limited availability of anesthesia and a list of over 100 children waiting for surgery make sequential bilateral surgery a good option in this situation. However, if there are concerns about the sterilization process of instruments at a facility or wound security due to lack of suture, performing unilateral cataract surgery might be preferred.

Anesthesia

General anesthesia is administered by a nurse anesthetist with pediatric experience. Halothane gas is used as the anesthetic agent. Halothane is less expensive than newer agents such as sevoflurane but can give more post-operative nausea and

prolonged somnolence. Alternatively, if an anesthetist and/or anesthesia machine is not available, intravenous ketamine is an inexpensive alternative that can produce sleep-inducing and analgesic effects. It typically is combined with a periocular local anesthetic injection. Ketamine is a safe alternative without the respiratory or cardiovascular suppression effects of inhaled agents but does create a less controlled anesthetic experience with potentially vivid dreams and illusions for the patient.

Equipment

The Vitron 2020 (Geuder, Germany) pneumatic vitrectomy unit was used for the case. It comes with a 20-gauge cutting probe and an anterior chamber (AC) maintainer. It has the advantages of being significantly less expensive and easier to maintain compared to other vitrectors. It is also compact and relatively lightweight making it a good portable option for the outreach setting. It utilizes a manual syringe for its suction mechanism. This mechanism can be more cumbersome and provide less controlled suction if the surgeon is not experienced with the device. It also has a limited cut speed of only up to 800 cuts per minute, which can cause unwanted vitreoretinal traction during anterior vitrectomy.

Scan Optics SO-161-R operating microscope was used for the case. It has the advantages of being cost-effective, easy to maintain, and portable. The optical clarity is not as good when compared to more expensive and modern operating microscopes. It is also less user-friendly as it has only a manual focus knob and no zoom capability.

Wound Construction

A superior scleral tunnel is constructed with a 15 blade. This wound is later extended through the cornea into the anterior chamber to facilitate IOL insertion. The scleral flap can later be closed with non-absorbable suture that is covered by the conjunctiva. Two 20-gauge stab incisions are made at the limbus, one for the vitrector probe and the other for the AC maintainer. The location of each incision may vary based on surgeon's preference. These incisions and the entry incision into the anterior chamber can be approximated to size with the tip of the 15 blade if a 20-gauge MVR blade and keratome are not available.

Cataract Removal

Using the vitrector probe at a cut rate of 200–300 cuts/minute, an anterior capsulectomy, or vitrectorhexis, is made. The lens is aspirated in its entirety using the manual suction action of the syringe attached to the vitrector probe.

IOL Decision-Making and Implantation

For this patient, there is no keratometer or A-scan ultrasound available for IOL calculations. IOL power selection is made based on the patient age (see Table 25.3). These are suggested guidelines for bilateral cases when biometry is not available. If A-scan ultrasound were available, estimations could be made based on axial length (see Table 25.4). A target post-operative refraction of emmetropia is preferred as post-operative spectacle correction is not a reliable option. With this refractive target, the child is likely to become fairly myopic as they age, but treating amblyopia is of much greater concern in this setting. Young children have a limited window to stimulate their visual pathway, whereas progressive myopia can be corrected at any age. We implant IOLs whenever possible in the developing world. Reasons for not implanting IOLs include microphthalmia and corneal diameter less than 9 mm. Caution is used when implanting IOLs with corneal diameters less than 10 mm.

Hydroxypropyl methylcellulose is used as an inexpensive viscoelastic for filling the capsular bag. A foldable, single-piece acrylic IOL is selected for capsular bag placement. Aurolab in India provides these IOLs at a low cost. PMMA can be used as a less expensive option, but these lenses require larger wounds. They can also cause significant post-operative fibrinous uveitis, especially in very young children with strong immune systems. For this reason, we try to avoid PMMA lenses in children less than 5. The Aurolab foldable lens comes with a disposable injector that uses a syringe-like plunger to implant the IOL into the bag. Alternatively, the Monarch® injector system (Alcon, USA) has a reusable handpiece and cartridge that can provide a more controlled insertion with plunger that is guided by a screw mechanism. While this device is designed for Alcon IOLs, we have found it to be interchangeable with IOLs from other manufacturers.

Table 25.3 IOL power selection based on age

Age	Power
<6 months	27–30
6–12 months	26
1 year	25
2 years	24
3 years	23
4 years	22
5 years and older	20–22

Table 25.4 IOL power selection based on axial length

Axial length	Power
17 mm	28–30
18 mm	27
19 mm	26
20 mm	25
21 mm	23
22 mm	22

Posterior Capsulotomy and Anterior Vitrectomy

A pars plana posterior capsulotomy and anterior vitrectomy are performed after the IOL is inserted into the bag. Alternatively, this could be performed anteriorly through the limbal incision by lifting up and going under the IOL with the vitrector probe. Pars plana has the advantages of less IOL disruption in smaller eyes and less vitreoretinal traction. As follow-up care and availability of a YAG laser are uncertain, primary posterior capsulotomy is performed in all children less than 10 years old.

Wound Closure and Intraoperative Medication

The scleral incision and pars plana sclerotomy are closed with a single cross-stitch 9-0 nylon. The limbal incisions are hydrated. An anterior chamber air bubble can provide additional temporary tamponade to the limbal incisions. Given concerns for sanitation in the developing world, intracameral antibiotic (cefuroxime) is used. The superior conjunctiva incision site is hydrated with triamcinolone, which allows it to cover the scleral sutures. The triamcinolone stays deposited on the ocular surface longer compared to dexamethasone. This can be beneficial if compliance with post-operative eye drops is a concern.

Post-operative Care

The child stays overnight in the hospital ward. After patch removal the following day, atropine 1% drops and a combination of chloramphenicol 0.5% + dexamethasone 0.1% drops are started. On post-operative day 2, the child is refracted. By post-operative day 3, the child has received spectacles and is discharged home after the eye is cleared of any evidence of endophthalmitis. The child is counseled by the childhood blindness counselor with emphasis on the need to return for future post-operative examinations.

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