IN THIS CHAPTER

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Pediatric Cataract Surgery

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5.13

Definition: Cataracts occurring in the pediatric age group, arbitrarily defined as birth to adolescence.

Key features

- Two main approaches are used to remove cataracts in children: pars plana and limbal.
- Intraocular lenses, contact lenses, and spectacles, are the most readily available means to correct aphakia in children.
- Posterior chamber intraocular lenses supplemented by spectacles are the best option for correction of aphakia in children, because most of the correction is permanently situated inside the eye globe.

INTRODUCTION

Cataracts in childhood not only reduce vision but also interfere with normal visual development.[1–3] The management of pediatric cataracts is far more complex than the management of cataracts in adults. The timing of surgery, the surgical technique, the choice of the aphakic correction, and the amblyopia management are of utmost importance in achieving good and long-lasting results in children.[4–10] Children's eyes are not only smaller than adults' eyes, but their tissues are also much softer. The inflammatory response to surgical insult seems more pronounced in children, often because of iatrogenic damage to the iris.[11] During the past two decades, the refinements that have occurred in adult cataract surgery have contributed to the further development of pediatric cataract surgery (PCS).[2,4–8] Certain adaptations and modifications in surgical technique are required to achieve results similar to those achieved in adults.[2–8] Furthermore, postoperative amblyopia management forms an integral part of visual rehabilitation in children.[1–10]

HISTORICAL REVIEW

Discission of soft cataracts was first described by Aurelius Cornelius Celsius, a Roman physician who lived 2000 years ago. Because of its simplicity, discission remained the method of choice until the middle of the twentieth century. The technique consisted of lacerating the anterior capsule and exposing the lens material to the aqueous humor for resorption and/or secondary washout. Repeated discissions were often required to manage the inevitable secondary cataracts.[2,11] Many early complications, e.g., plastic iritis, glaucoma, and retinal detachments were associated with these early techniques.[2,11] With the advent of vitrectomy machines and viscoelastic substances, as well as the refinements in cataract surgery, these complications have been reduced markedly over the past two decades.[2–11]

PREOPERATIVE EVALUATION AND DIAGNOSTIC APPROACH

A careful history assists the clinician in selecting the investigations needed for determining the cataract's etiology.[2] Problems during pregnancy (e.g., infections, rashes or febrile illnesses, exposures to drugs, toxins, or ionizing radiation) should be elicited. Family history of cataracts in childhood or other ocular abnormalities can be relevant. Both parents and all siblings should be examined with a slit lamp to determine any lens abnormalities. When family history is positive, consultation with a geneticist is recommended. A thorough examination by a pediatrician to assess the child's general health and elicit information about other congenital abnormalities is mandatory.

Laboratory tests in children who have bilateral cataracts in nonhereditary cases are listed in Box 5-13-1. Most unilateral pediatric cataracts are idiopathic and do not warrant exhaustive laboratory tests.

The ophthalmologic part of the evaluation starts with a complete ocular examination, which includes an assessment of visual acuity, pupillary response, and ocular motility. Biomicroscopy follows and might necessitate sedation or even general anesthesia in very young patients. Indirect fundus examination with dilated pupils is made unless the cataract is complete. A- and B-scan ultrasonography is carried out in both eyes to compare axial lengths and to discover any posterior segment abnormalities. Earlier photographs should be examined for the quality of the pupil's red reflexes. This might help to date the onset of the cataracts.

ALTERNATIVES TO SURGERY

The development of metabolic cataracts, such as those found in galactosemia, can be reversed if they are discovered in the early phases. With the elimination of galactose from the diet, the early changes in the lens, which resemble an oil droplet in the center of the lens, can be reversed.[12] Later on, lamellar or total cataracts develop, which require surgery.

When lens opacities are confined to the center of the anterior capsule or the anterior cortex, mild dilatation of the pupils with homatropine 2% twice daily can improve vision and postpone the need for surgery. Photophobia and partial loss of accommodation are side-effects of this measure. This temporary management should be implemented only in bilateral cataracts in which vision is equal in both eyes and better than 20/60.

ANESTHESIA

General anesthesia is presently the only anesthetic option in PCS. It is extremely important to request deep anesthesia throughout the procedure in order to minimize iatrogenic damage to iris and cornea.[5,7,8]

BOX 5-13-1 LABORATORY TESTS FOR BILATERAL NONHEREDITARY PEDIATRIC CATARACTS

Full blood count
Random blood sugar
Plasma calcium and phosphorus
Urine assay for reducing substances after milk feeding
Red blood cell transferase and galactokinase levels
If Lowe's syndrome is suspected, screening for amino acids in urine
Toxoplasmosis titer
Rubella titer
Cytomegalovirus titer
Herpes simplex titer

Children's scleras and corneas are particularly soft, therefore, any tension on the extraocular muscles results in loss of anterior chamber depth and increased intraocular pressure. A useful marker for anesthesia depth is the position of the eye during surgery. If the cornea moves upwards, the anesthesia is too light and should be deepened. When this advice is followed, surgery is easier to perform and iatrogenic damage to the iris and cornea is diminished.

GENERAL TECHNIQUES

Unlike in adults, pediatric cataracts are soft. Their lens material can be aspirated through incisions that are 1–1.5 mm long at the limbus or can be subjected to lensectomy through pars plana. When intraocular lens (IOL) implantation is intended, a larger limbal wound is needed to introduce the IOL. A scleral tunnel is safer than a clear corneal incision. Unlike in adults, the wound should be securely sutured to prevent wound dehiscence with iris incarceration – a common complication in children.[2,4,5,7,8,10]

SPECIFIC TECHNIQUES

See clip: 5.13.01

Two main approaches exist for the removal of cataracts in children: the pars plana approach and the limbal approach.

Both techniques have advantages and disadvantages. The pars plana approach was developed with the advent of vitrectomy machines in the late 1970s;[13,14] it was intended to deal mainly with very young infants in whom surgery is more difficult. With the continuing refinements in cataract and implant surgery in adults, the pars plana approach is being abandoned gradually in favor of the limbal approach, because the latter allows better preservation of the capsular bag for in-the-bag IOL placement.[2,5,7,8]

Pars Plana Approach

The pars plana approach is indicated mainly for neonates and infants under 2 years of age, particularly for those who have bilateral congenital cataracts for whom immediate IOL implantation is not intended.[2] The technique requires a guillotine-type vitrectome and balanced salt solution containing epinephrine (adrenaline) 1 : 500 000. The location of the pars plana in infants can be 1.5–3.5 mm from the limbus. In the last decade surgeons have largely abandoned the 20G vitrectomy apparatus in favor of the 23G or the 25G version. A lensectomy-anterior vitrectomy is completed, sparing a 2–3 mm peripheral rim of anterior and posterior capsule. These capsule remnants are used to create a shelf to support a posterior chamber IOL that may be implanted later on in life.[15] It is important to avoid vitreous incarceration in the wounds by turning off the infusion before withdrawing the vitrectomy cutter from the eye. This precaution reduces the chances of suffering retinal traction and detachment later in life.

This technique is rapid and allows a permanently clear visual axis. The postoperative course is normally less complicated than that after the limbal approach, because fewer maneuvers occur in the anterior chamber. Consequently, the iris and the corneal endothelium suffer less iatrogenic damage. In neonates who have bilateral cataracts, for whom the anesthetic risk is great, the two eyes can be operated on at the same sitting using different sets of instruments.[2,6] Simultaneous surgery also reduces the risk of relative amblyopia, which can occur when two operations are undertaken a few days apart.[2,6]

A possible occurrence of the pars plana approach is the incarceration of vitreous in the scleral incisions. Subsequent vitreous traction may lead to retinal breaks and/or detachments.[2,16] Another hindrance with the pars plana approach arises when the pupil is dilated insufficiently; the lensectomy has to be performed under partially 'blind' conditions, which means either leaving too much lens material in the periphery or too little peripheral capsular support for future posterior chamber IOL implantation.[15]

Limbal Approach

With the proper precautions, the limbal approach is the most versatile technique for PCS.[2,4,5,7,8] Many surgeons have not yet recognized the importance of the anterior chamber maintainer (ACM) when operating on eyes in young patients. Although it is possible to use an aspiration-irrigation device or a vitrectome with an irrigation sleeve in order to remove a soft cataract, the use of an ACM makes the surgery safer. Moreover, although viscoelastic materials maintain space, the ACM provides, in addition, a steady intraoperative intraocular pressure (IOP) with continuous washout of blood, pigment, and prostaglandins that may be released during surgery. The ACM also helps to keep the pupil well dilated throughout the procedure because of the positive hydrostatic pressure. It prevents collapse of the globe when the instruments are withdrawn from the eye and thus helps to reduce damage to the iris and corneal endothelium. This feature of the ACM allows leaving a clear and 'clean' media with minimal occurrence of postoperative anterior chamber (A/C) fibrinous reaction.

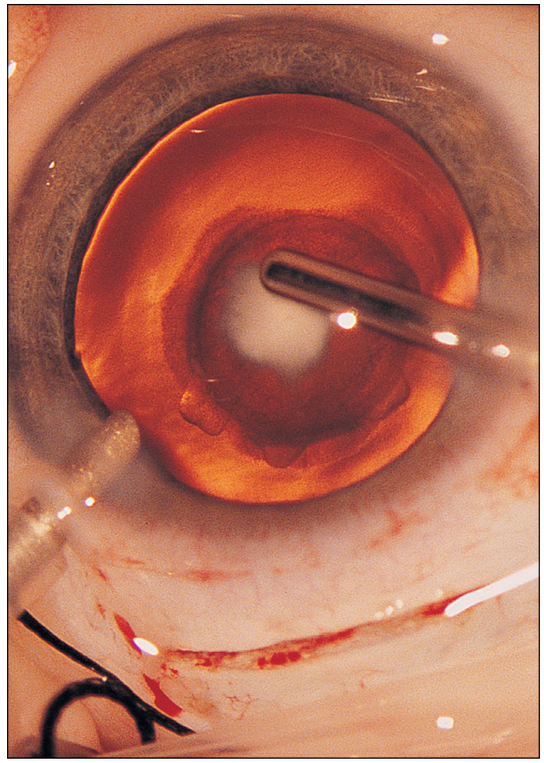

Fig. 5-13-1 Anterior capsulectomy performed using a vitrectomy probe in a congenital cataract. Note the use of the anterior chamber maintainer for a deep anterior chamber and a well-dilated pupil.

Two limbal incisions are made with a 20G stiletto knife; one for the ACM (connected to a balanced salt solution with epinephrine 1 : 500 000) and the other one for the aspiration cannula according to the surgeon's preference positions.

Various techniques have been described by which to open the anterior capsule.[2,4,13,14] Capsulorrhexis can be carried out with the help of high-viscosity viscoelastics; however, the younger the child is, the more difficult it is to perform a capsulorrhexis. Infants have a very elastic anterior capsule, which easily tears toward the periphery. A practical alternative to manual capsulorrhexis is to use a vitrectomy probe to create a small central opening in the anterior capsule (Fig. 5-13-1). This initial hole can be enlarged gradually by 'biting' into the anterior capsule with the vitrectome until the desired 4–5 mm opening is achieved. The lens material can be aspirated manually or with an automatic aspiration device.

Once the capsular bag is empty, the decision has to be made as to the management of the posterior capsule. Most surgeons agree that infants under 2 years of age should receive an elective posterior capsulectomy-anterior vitrectomy.[2,4–8,13,14] Posterior capsulorrhexis can be carried out either manually or with the vitrectome, as described for the anterior capsule.[2,4–8,13,14,17] The posterior capsulorrhexis diameter must be at least 4 mm. One third of the anterior vitreous must be removed to ensure a permanently clear visual axis (Fig. 5-13-2).

Smaller posterior capsulectomies with shallow anterior vitrectomies tend to close down, especially in neonates.[18] Posterior capsulectomy, either alone or when combined with a shallow anterior vitrectomy, does not guarantee a permanently clear visual axis, because vitreous remnants serve as a scaffold for the lens epithelium to grow on, which results in the formation of new opaque membranes. Furthermore, the immediate postoperative iritis seems markedly reduced when a generous anterior vitrectomy has been performed.[2,4,5,7,8,14] Management

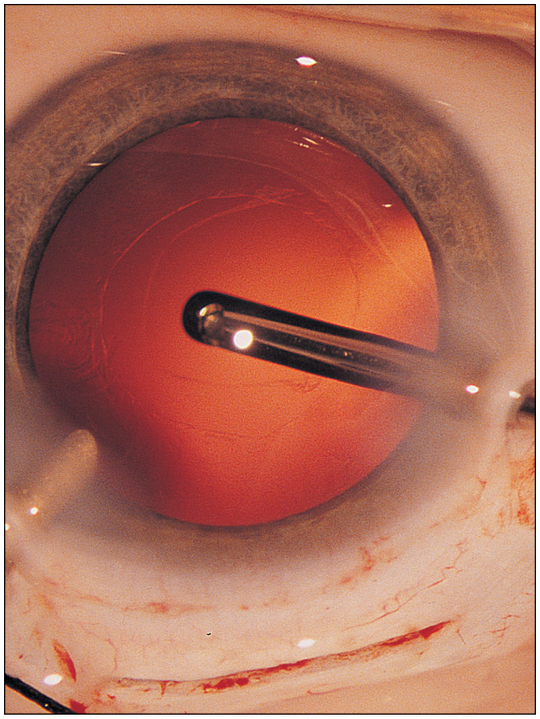

Fig. 5-13-2 Elective posterior capsulectomy and a deep anterior vitrectomy. This is performed using a vitrectomy probe, after all the lens material has been aspirated within the capsular bag.

of the posterior capsule in children older than 2 years remains controversial. Some authors prefer to leave it intact until opacification occurs; others perform an yttrium–aluminum–garnet (YAG) laser capsulectomy immediately after surgery. Experienced pediatric cataract surgeons choose to perform an elective posterior capsulectomy–anterior vitrectomy, routinely, in every child under 8 years of age in order to provide a one-stop treatment in this age group wherein amblyopia is still a risk. This alternative is logical when attentive follow-up is uncertain.[4–10,13,14]

CHOICE OF APHAKIC CORRECTION IN CHILDREN

Spectacles, contact lenses, and IOLs are the most readily available means to correct aphakia in children.

Spectacles

Aphakic spectacles provide a satisfactory correction only in cases of bilateral aphakia in which anisometropia does not represent a problem.[2] Most of these patients develop good visual acuity with spectacles, provided the eyes are not excessively microphthalmic.[2] The disadvantages of spectacles are cosmetic blemish and the poor optical quality of high-plus lenses.

Contact Lenses

During the 1970s and 1980s, contact lenses were described as the method of choice to correct unilateral and bilateral aphakia in childhood.[2,9,10] Contact lenses provide a better optical correction than spectacles, and their dioptric power can be adjusted throughout life. However, the management of contact lenses in children can be very difficult and costly. Frequent loss of lenses, recurrent infections, and poor follow-up turn this theoretically ideal choice into the most impractical option. Most ophthalmologists, therefore, now recommend the use of IOLs supplemented by spectacles in children rather than contact lenses.[2,4,5,7,8,10,13]

Intraocular Lenses

The IOL option was originally advocated in cases of unilateral pediatric cataracts because it facilitates amblyopia management by providing a more permanent correction.[2,4,5,7,8,10,13] Implanting an IOL in a growing eye is not an ideal solution, but it is currently the most practical one. The aim in the IOL option, unlike in the contact lens alternative, is to correct most, but not all, of the aphakia; the residual refractive error has to be corrected using spectacles, which can be adjusted throughout life.

The implantation of anterior chamber IOLs in children was discontinued in the mid-1980s. Devastating complications, such as secondary glaucoma and corneal decompensation, were attributed to anterior chamber IOLs, especially in younger patients.[19] Posterior chamber IOL implantation represents by far the better method for the correction of aphakia in adults, and the same applies in children.

Selection of intraocular lenses

The choice of the dioptric power of IOL to implant in young children is the main difficulty that faces the ophthalmologist.[2] Pediatric IOLs are not yet readily available,[20,21] and the rapid growth of the eye during the first 2 years of life makes an effective choice difficult.[2,4,7,8,22–25] Nevertheless, in the 1990s increasingly positive reports were published on the use of posterior chamber IOLs in children and even in neonates.

The material from which the IOL is made must have a long track record of safety. Polymethyl methacrylate (PMMA) IOLs have been in use for more than 50 years; PMMA is probably the safest material to be used for children, until a similar follow-up is obtained for other biomaterials.[20] Nevertheless, during the last decade many surgeons have switched to the use of foldable hydrophilic and hydrophobic IOLs in children. The actual size of the capsular bag and the ciliary sulcus in children has been ascertained by the work of Bluestein and coworkers.[21] Posterior chamber IOLs, which were originally oversized, have been reduced from 13–14 mm to 12–12.5 mm in diameter in most modern models. In children it is even more important to implant an IOL of the correct size.[21] Oversized IOLs act like loaded springs in the eye and can dislocate, especially when a child rubs the pseudophakic eye causing damage to intraocular structures. Pediatric IOLs should not exceed 12 mm overall diameter because the average adult ciliary sulcus diameter rarely exceeds 11.5 mm. Ideally, the pediatric IOL should be available in diameters of the range 10.5–12 mm.[21] The choice of IOL size is determined mainly by the site of implantation (i.e., in-the-bag or ciliary sulcus).

Both the biometry and the age of the child determine the choice of the IOL dioptric power. Two main age groups exist in PCS: patients younger than 2 years and patients between 2 and 8 years. In the first group, the axial length and the keratometric (K) readings change rapidly, whereas in the second group the changes are slower and more moderate.[22–25] In order to minimize the need to exchange IOLs later in life, when a large myopic shift occurs, it is advisable to undercorrect children with IOLs so that they can grow into emmetropia or mild myopia in adult life.[22–25]

Those who are under 2 years of age should receive 80% of the power needed for emmetropia at the time of surgery. Since the K readings also change rapidly during the first 18 months of life, it is practical to rely on the axial length only when the IOL dioptric power is chosen for infants (Box 5-13-2). The postoperative residual refractive error is corrected by spectacles, which can be adjusted at will as the child grows. Infants and toddlers can tolerate up to 6 D of anisometropia, which gradually disappears within 2–3 years.[24] Most of the infants who have unilateral pseudophakia need a patch over the sound eye for half their waking hours until 4–5 years of age. Patches alleviate the symptoms of anisometropia but at the same time affect the chances for good binocularity to develop.[26]

BOX 5-13-2 GUIDELINES FOR THE CHOICE OF INTRAOCULAR LENS DIOPTRIC POWER

Children Less Than 2 Years Old

- Do biometry and undercorrect by 20%, or
- Use axial length measurements only:
  - Axial length IOL dioptic power
  - 17 mm, 25 D
  - 18 mm, 24 D
  - 19 mm, 23 D
  - 20 mm, 21 D
  - 21 mm, 19 D

Children between 2 and 8 Years Old

- Do biometry and undercorrect by 10%

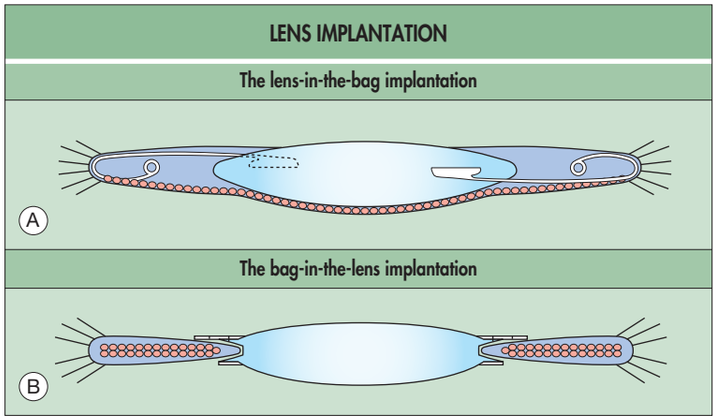

Fig. 5-13-3 Schematic drawing of the lens-in-the-bag implantation (A) and the bag-in-the-lens implantation (B).

For the age range 2–8 years, the IOL dioptric power should be 90% of that needed for emmetropia at the time of surgery (see Box 5-13-2). The induced anisometropia is moderate and lessens with the expected myopic shift that occurs in adolescence.[22–25]

The need for spectacles after IOL implantation in PCS has some positive aspects:

- More dependency on the ophthalmologist is needed because spectacles have to be taken care of, adjusted, and repaired periodically; this increases the chances of attentive follow-up.
- The pseudophakic eye is protected from direct trauma by the spectacles.
- Spectacles can be used as an adjunct to amblyopia therapy by atropine penalization of the sound eye and alteration of the dioptric power of its lens.

Implantation in children under 2 years of age

In unilateral cases, primary implantation is indicated as soon as the patient is fit for anesthesia, ideally between 2 and 3 months of age. The earlier the surgery is done, the better the chance that deep amblyopia can be overcome.

After the cataract has been aspirated, an elective posterior capsulectomy–anterior vitrectomy is performed. The posterior chamber IOL is inserted through a scleral tunnel, which is prepared in advance. The surgeon has to choose between ciliary sulcus and the bag implantation according to his/her surgical experience. Sulcus implantation is easier and also allows an easier explantation in cases where IOL exchange will be needed later in life.[24] This option is indicated in neonates and infants less than 1 year of age. The in-the-bag placement is more physiological, but more difficult technically. An in-the-bag IOL is more difficult to explant; this option should be chosen for infants above 1 year of age because they are less likely to need an IOL exchange, provided they are undercorrected by 20%.[27]

Implantation in children above 2 years of age

For children older than 2 years, the IOL should be inserted in the bag because the eye has reached nearly the adult size, although its sclera is much softer. Gimbel[17] has described a special IOL implantation for this group of patients. The technique requires extreme dexterity as both anterior and posterior capsulorrhexises are performed. The IOL haptics are placed in the bag fornices, while the optic is protruded through both capsulorrhexises to be captured beneath the posterior capsule remnants. Tassignon has recently developed a new technique for a special IOL called bag-in-the-lens.[20] The technique consists of creating an anterior and posterior capsulorrhexis. The specially designed IOL has, at its periphery, a groove that contains both anterior and posterior capsule rims (Fig. 5-13-3). Although technically demanding, promising early results indicate that this technique might do away with the need for elective anterior vitrectomy (Fig. 5-13-4).[19]

Postoperative treatment

Topical medications are sufficient when surgery has not been excessively traumatic. A combination of antibiotic–corticosteroid drops every 2 hours with a mild mydriatic agent twice daily is given for the first week. Thereafter, the medications are tapered off during the next 3 weeks. Some authors have used systemic corticosteroids to overcome the intense inflammatory response in young children's eyes.

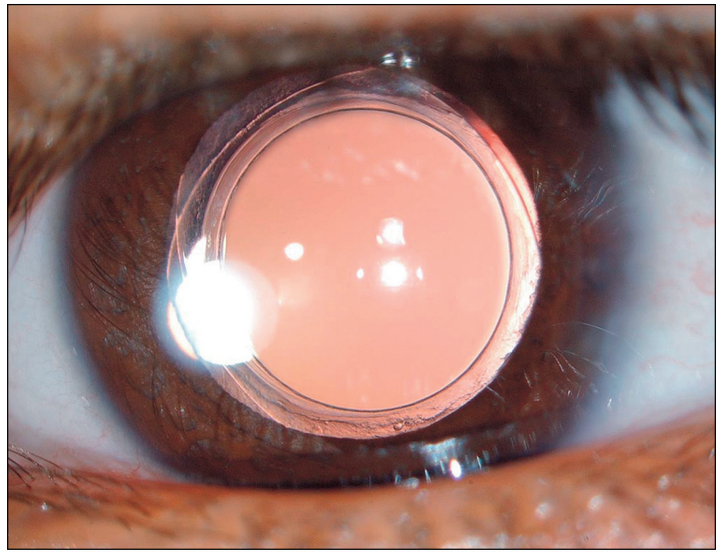

Fig. 5-13-4 Bag-in-the-lens IOL implanted in a 5-year-old child at 41 months follow-up. Note the perfectly clear visual axis and the capsular rims contained in the IOL peripheral groove. (Reproduced with permission of MJ Tassignon.)

COMPLICATIONS

Intraoperative complications are usually related to the surgeon's unfamiliarity with the child's soft ocular tissues. The anterior chamber tends to collapse, the iris can protrude through the surgical wounds, and the pupil constricts on injury to the iris. These events can be avoided by operating under deep anesthesia and by using an ACM.

Immediate postoperative complications include anterior plastic uveitis, high IOP, incarceration of iris tissue in the wound, and endophthalmitis. Atraumatic surgery, use of an ACM during surgery, thorough removal of viscoelastics at completion of surgery, and meticulous closure of the wound reduce the occurrence of these complications.

Late complications include dislocation of the IOL, chronic iritis, glaucoma, and retinal detachment. Close follow-up enables detection of these complications at an early stage. Their treatment is similar to that for the same occurrences in adults.

Amblyopia Management

The child's parents must understand that visual rehabilitation only starts with surgery and must be continued throughout childhood.

The unilateral cases are the most difficult to manage.[2,4,5,7–10] Amblyopia treatment starts soon after surgery, after postoperative inflammation subsides and the media becomes clear. The initial treatment must be aggressive in order to boost vision in the deprived eye. Full-time occlusion of the sound eye is carried out for a few days – 1 day per month of age. For example, a 3-month-old neonate should be subjected to occlusion for 3 consecutive days, a 4-month-old infant for 4 days, etc. Thereafter, occlusion is reduced to half the waking hours. The younger the infant, the easier it is to comply with the patch regimen. Autorefractometers, especially portable ones, help to determine the residual refractive error; retinoscopy is often difficult in pseudophakic children. Spectacles are prescribed from the age of 4 months onward. A bifocal lens with an add of +3.00 is prescribed in the pseudophakic eye from the age of 3 years, when the child becomes verbal. Unilateral pseudophakes should continue with half-day patches until 4–5 years of age. Thereafter, the patch time can be reduced gradually, but should not be abandoned until 10–12 years of age. After that age, amblyopia management is practically superfluous.

Cases of bilateral pseudophakia should be followed closely to detect and treat relative amblyopia.

Intraocular Lens Exchange and Alternative Options

Exchange of IOLs should be considered when a great myopic shift has occurred.[22–25] When the pseudophakic eye becomes 7 D more myopic

than the sound eye, the IOL should be exchanged, unless contact lens wear is a viable option. Refractive surgery in children is not yet an acceptable option. An experienced anterior segment surgeon who is familiar with IOL exchange should perform the procedure. An alternative to IOL exchange is to implant, preferably in the posterior chamber, an additional negative dioptric power IOL to correct the myopia. This procedure is easily performed when the primary IOL was inserted in the bag.

OUTCOME

The visual outcome depends largely on the type of cataract the laterality of the pathology, the timing of intervention, the quality of surgery, and, above all, the amblyopia management. It is possible to achieve nearly normal vision even in unilateral congenital cataracts, provided amblyopia management is aggressive.[2–10,24] Binocularity is usually poor in these cases, but some gross stereopsis can be expected.[26] Aphakic and pseudophakic children certainly should be followed-up throughout childhood and preferably throughout life.[28]

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