Paediatric Cataract Surgery

Charlotta Zetterström

Core Messages

- It is important to detect cataract within the first weeks of life
- Life-long control is mandatory if operated on early, high risk for development of glaucoma
- Loss of accommodation, important to inform before surgery
- IOL implantation is safe and generally accepted over the age of 1 year
- After-cataract is problematic in children
- For lamellar cataract, surgery is often indicated before school start

9.1 Introduction

Bilateral congenital cataract is the most common cause of treatable childhood blindness. Unilateral congenital cataract is an important cause of amblyopia and strabismus.

Cataract surgery in children has changed and improved dramatically in recent decades. This is mainly a result of modern surgical techniques and improved intraocular lenses. Also, better knowledge of irreversible deprivation amblyopia and how to treat this has made an important contribution [13, 33].

9.1.1 Aetiology

In the developed world, the cause of most cases of bilateral congenital cataracts is idiopathic. About one third are hereditary without systemic disease. These are mostly autosomal dominant but autosomal recessive and X-linked traits occur. Rare causes of childhood cataracts are metabolic disorders such as galactosemia and hypocalcemia. Congenital cataracts can be combined with systemic abnormalities such as trisomy 21 and Turner's syndrome. Mental retardation is common in series of bilateral congenital cataract and there is a multitude of inherited syndromes with this combination associated with other abnormalities such as craniofacial or skeletal deformities, myopathy, spasticity or other neurological disturbances.

A number of intrauterine infections (toxoplasmosis, rubella, cytomegalic inclusion disease, herpes infection, varicella, and syphilis) may cause congenital cataracts. Of these, rubella is the most important. The rubella cataract is usually bilateral but may be unilateral.

Ocular conditions such as aniridia (Fig. 9.1) and iris coloboma (Fig. 9.2) are often seen together with cataract.

Unilateral congenital cataract is, as a rule, not associated with systemic disease, is rarely inherited and the cause is in the majority of cases idiopathic. About 10 % of cases are associated with lenticonus/lentiglobus and persistent foetal vasculature. It may also be masked bilateral cataract because of asymmetric lens involvement.

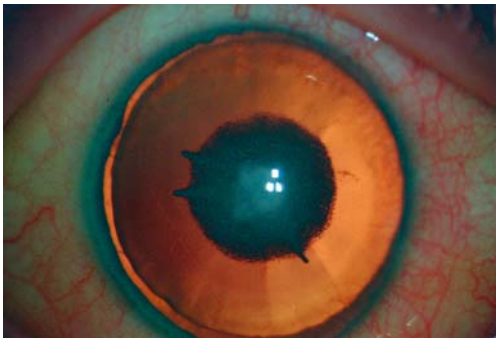

Fig. 9.1. Child with aniridia and complete nuclear cataract

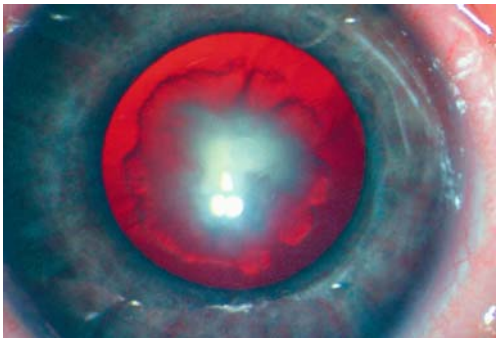

Fig. 9.3. New-born child with nuclear cataract dense in the centre

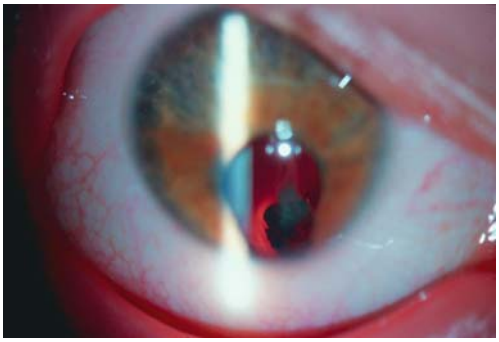

Fig. 9.2. Child with coloboma and cataract

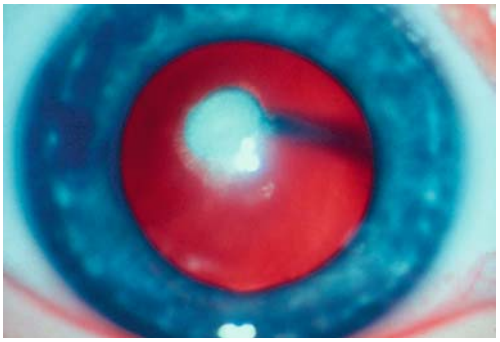

Fig. 9.4. Posterior cataract with persistent foetal vasculature

Summary for the Clinician

- **In a clinically healthy child with one parent with the disease, and also in unilateral cases, an extensive pre-operative investigation to establish a cause for the cataract is not necessary**

9.1.2 Morphology

Nuclear cataract is usually present at birth and is non-progressive [30]. Dense cataracts present at birth, where early surgery is mandatory, are in most cases of nuclear type (Fig. 9.3). The opacification is located to the embryonic and foetal nuclei between the anterior and posterior Y sutures and is usually very dense in the centre. The eyes are almost always smaller than normal eyes [18]. The cataract is bilateral in about 80 % of cases. In cases with bilateral congenital nuclear cataract, inheritance can be demonstrated in 30 %–50 %. The inheritance is in most cases autosomal dominant.

Posterior unilateral cataracts in infants and children are in most cases associated with persistent foetal vasculature (PFV) and the affected eye is usually small (Fig. 9.4). The retrolental vascular structure in contact with the lens capsule may give way to blood vessels encircling the lens causing haemorrhage, particularly during surgery (Fig. 9.5). The fibrovascular stalk may cause tractional retinal detachment. After early surgery, secondary glaucoma is unfortunately a common complication in these eyes [26].

Cataract associated with posterior lenticonus or posterior lentiglobus usually develops

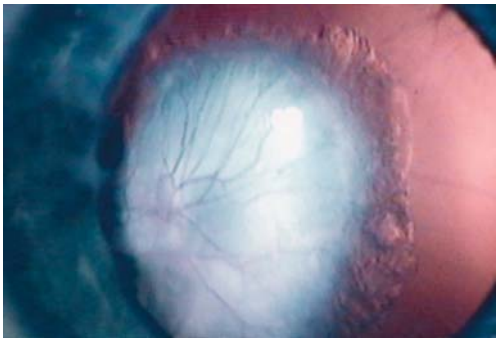

Fig. 9.5. Posterior persistent foetal vasculature

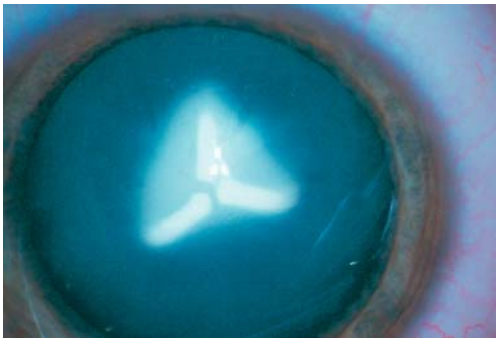

Fig. 9.7. Sutural cataract with little influence on vision

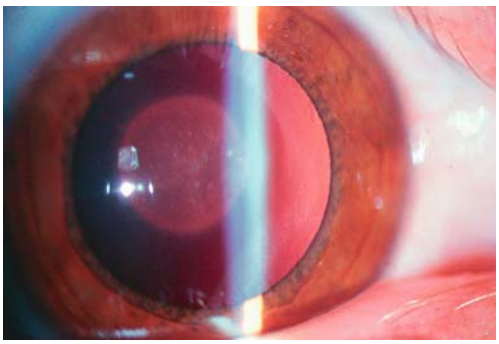

Fig. 9.6. Lamellar cataract in a 4-year-old child

after the critical period of visual development. It is mostly unilateral and occurs sporadically. The change in the lens develops as a small defect in the posterior lens capsule, which exhibits a progressive bowing resulting in a posterior bulging and disorganisation of the subcapsular lamellae and opacification. It is important to be aware of the weakness of the posterior capsule in these eyes during surgery and, if possible, avoid hydrodissection.

Lamellar cataract usually develops after establishment of fixation (Fig. 9.6). It is usually progressive and surgery is often performed before school age, but the cataract can remain subclinical for many years. The cataract involves the lamellae surrounding the foetal nucleus peripheral to the Y sutures [30]. Eyes with lamellar cataracts are usually of normal size with a normal-sized cornea. It is uniformly bilateral and has commonly an autosomal dominant pattern of inheritance.

There are some other morphological types of congenital cataracts. Some are due to lenticular developmental defects present at birth. These may have only little influence on vision. Such defects are sutural cataract (Fig. 9.7), and anterior polar cataract, which usually do not progress.

Summary for the Clinician

- **In the case of nuclear cataract, early surgery is often needed**
- **For lamellar cataract, surgery is often indicated before school start**

9.1.3 Amblyopia and Congenital Cataracts

Amblyopia is caused by abnormal structural and functional evolution of the lateral geniculate nucleus and striate cortex due to the abnormal visual stimulation during the sensitive period of visual development.

Reversibility of amblyopia depends on the stage of maturity of the visual system at which abnormal visual experience began, the duration of deprivation and the age at which therapy was instituted. The most critical period is probably between 1 week and 2 months [40]. Disruption of vision during this period usually causes severe and permanent visual loss and permanent nystagmus if not managed. If visual deprivation

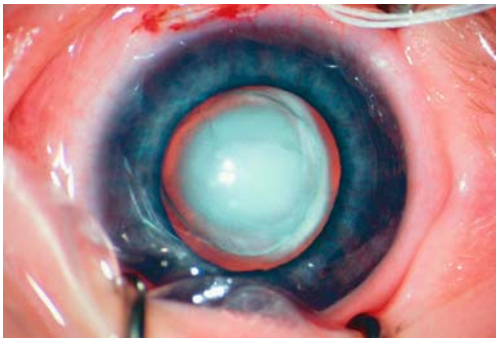

Fig. 9.8. Dense congenital cataract

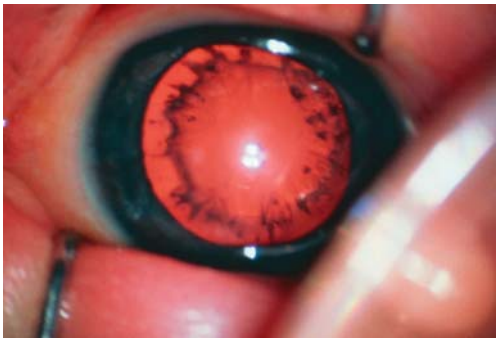

Fig. 9.9. Incomplete congenital cataract

occurs after the age of 2–3 months, the amblyopia is usually reversible. The sensitivity to amblyopia gradually decreases until the age of 6 or 7 years when the visual maturation is complete and the retinocortical pathway and the visual centres become immune to abnormal visual input [37]. It is thus essential that early treatment of dense congenital cataract is instituted in order to avoid irreversible amblyopia [9, 16, 31].

Visual loss and the development of amblyopia depend on the size and location of the cataract and particularly on the density. If the opacities are large enough to obscure the fundus view through an undilated pupil, amblyopia development can be expected (Fig. 9.8). If retinal details such as the larger vessels can be distinguished through the central portion of the cataract, conservative treatment can be considered (Fig. 9.9). Some infants with partial subclinical congenital cataract develop sufficient binocular interaction and form vision to allow a normal maturity of the visual system. Thus, amblyopia might not be a problem for some children with partial congenital cataract. If surgery is considered in children with partial cataract it should, if possible, be postponed until the age at which the risk of post-operative complications diminishes. Children with partial cataract treated conservatively must be followed closely. Occlusion therapy is necessary in unilateral cases to prevent amblyopia. The clinical evaluation should entail evaluation of visual behaviour including monocular and binocular fixation patterns.

Unilateral congenital cataract, dense from birth, causes amblyopia with loss of binocular function and the development of secondary strabismus. In cases of dense bilateral congenital cataract, bilateral amblyopia and nystagmus will occur.

Summary for the Clinician

- **To prevent irreversible amblyopia in infants with dense cataract from birth, the cataract extraction must be performed early**
- **Children with partial cataract must be followed closely**

9.2 Congenital Dense Cataract

9.2.1 Pre-operative Examination

A careful pre-operative examination of the eyes is essential, The red reflex should first be assessed by direct ophthalmoscopy with the pupil undilated. The cataract is often most dense in the central part of the lens, after dilatation it seems to be less significant. It is important to examine both eyes and establish whether the cataract is bi- or unilateral. Unilateral congenital cataract presents a challenging problem, since even a mild cataract will cause irreversible deep amblyopia in one eye if not treated. In these children, vision in the affected eye is prevented from developing through active suppression by the non-affected eye [6]. While the newborn child is awake it is also important to assess visual function, if possible, with a clinical pref-

erential looking grating acuity (Teller acuity card). It is also important to look at the ability to fix and follow and ask the parents if they have had any visual interaction with the child. Children with significant bilateral congenital cataracts may seem to have delayed development as well as obviously impaired visual behaviour. In contrast, children with monocular cataracts often present with strabismus, which may not develop until irreparable visual loss has occurred. Since their visual behaviour may be unaffected, children with monocular cataract are almost always detected much later than cases with bilateral cataract. Moreover, in most cases with monocular cataract, they have no family history and are otherwise healthy. The presence of manifest nystagmus at the age of 2–3 months or more generally indicates a poor visual prognosis. Complete examinations of infants often require sedation or general anaesthesia and can often be performed during the same anaesthesia as the surgery. Both eyes should be examined with dilated pupils because malformations in the non-cataractous eye are commonly found [24]. Anterior segment examination is carried out with measurements of the corneal diameter and intraocular pressure (IOP) by Tonopen or handheld Perkins tonometer. The IOP in the new-born is much lower than in the adult, often below 10 mmHg. If the clarity of the media permits so, indirect ophthalmoscopy may reveal persistent foetal vessels or any other posterior segment abnormalities that may have an impact on the visual outcome. Surgery for visually significant cataract should be carried out as soon as possible, preferably within the first weeks of life. If the cataract is unilateral it is even more important with early surgery to obtain some useful visual acuity in the affected eye. A treatment regime based on surgery within 2 months of life, prompt optical correction of the aphakia and aggressive occlusion therapy with frequent follow-up have been successful in several series [5, 7]. If the cataract is incomplete and not interfering with normal visual development, it is better to postpone surgery until the child is older with less post-operative complications. In these cases it is mandatory with close follow-ups by a well-trained paediatric ophthalmologist.

Summary for the Clinician

- **An undilated pupil and no red reflex are indications for immediate surgery**

9.2.2 Surgical Technique in Infants

In infants with bilateral cataracts it is advantageous to perform surgery in both eyes at the same surgical intervention. Most of the children are new-born, only a few weeks old, and these small children are extremely sensitive with regard to developing amblyopia. Surgery in one week on one eye while the other eye remains with dense cataract can cause irreversible amblyopia in the non-operated eye. If both eyes are operated at the same time sterility must be maintained during the whole procedure and changing all instruments and sterile clothing of the surgeon and nurse are advisable between the eyes.

Axial length and corneal curvature are essential measurements for contact lens fitting and IOL power calculation and could advantageously be performed immediately before surgery during the same anaesthesia. Eyes of infants with congenital cataract are shorter and have a smaller corneal diameter compared to controls.

Before surgery the pupil is dilated with a combination of 1.5% phenylephrine and 0.85% cyclopentolate. Rinsing of the conjunctiva with chlorhexidine solution 0.05% is performed 5 min before surgery.

The surgical intervention should be performed by a well-trained anterior segment surgeon to obtain the most advantageous outcome. Consequently most of the procedures are done with the anterior approach starting with a sclerocorneal tunnel, which ought to be rather long to minimise the risk for iris prolapse. High-viscosity ocular viscosurgical device (OVD) is used because the anterior chamber is shallow and a high vitreous pressure is found in these small eyes. If the pupil is small, which is rather common in eyes with congenital cataract, flexible iris retractors can be very helpful and four of them are placed in the pupil before the continu-

ous anterior capsulorhexis is performed. There is no reason to open the pupil with the iris retractors more than necessary, since a damaged iris will cause more post-operative inflammation. If the cataract is very dense and grey, staining of the anterior capsule with dye makes the anterior capsulorhexis much easier and also safer to perform [32]. The dye can be administered with a blunt syringe under the OVD just above the anterior capsule and painted on the capsule with the end of the syringe. In this way a very small amount of the dye is needed and just the capsule and not the whole anterior segment is dyed. If an intraocular lens (IOL) is implanted the anterior capsulorhexis ought to be smaller than the optic, round and placed in the centre. Since the capsule is elastic and thick in the new-born it is important to re-grasp continuously when performing the rhexis. In contrast to the adult eye, one ought to aim for a small rhexis to achieve a good size. A complete rhexis without any tears is essential in cases when IOL implantation is planned.

Hydrodissection ought to be carried out with caution and sometimes avoided. In some eyes, often with very dense cataract, a defect in the posterior capsule, most often formed like an almond, could be found. Hydrodissection in these cases is of course not recommended because the high risk of loosing lens material into the vitreous during the procedure. These cases also have opacification with cells in the anterior part of the vitreous, looking like a fishtail in the anterior vitreous when moving the eye. The posterior capsule is thin and fragile in eyes with posterior lenticonus and hydrodissection ought to be performed carefully.

It is almost always possible to remove the nucleus and cortex with irrigation and aspiration. However, sometimes in very dense nuclear cataracts, and with white calcified parts in the nucleus, ultrasound has to be used. The new technique AquaLase liquefaction, which uses a warm-water stream, may prove very useful when removing these dense cataracts. It is important to remove all lens material to minimise post-operative inflammation, which is very pronounced in young patients. To reduce opacification of the visual axis removal of most lens epithelial cells is important; however, it is almost impossible with the technique used routinely today. The most important cells concerning after-cataract are located at the lens equator and are impossible to see during surgery. When the capsular bag is empty of all lens material high-viscosity OVD is injected to fill the capsular bag and a posterior continuous capsulorhexis is performed: slightly smaller than the anterior rhexis. The posterior capsule is thinner than the anterior and not so elastic. Sometimes fibrotic parts are found in the posterior capsule which makes tearing impossible and scissors have to be used. It is wise to look for persistent foetal vasculatures, particularly in unilateral cases with posterior cataract. Persistent hyaloid artery is adherent to the posterior aspect of the lens and the optic disc. If present it ought to be cut with fine scissors; sometimes the vessel contains blood, but cautery is seldom indicated. Using this method it is possible to implant an IOL in the capsular bag during primary surgery or in the ciliary sulcus if a secondary implantation is scheduled for the future. Capsular fixation is preferred over ciliary sulcus placement because such complications such as pupillary capture and IOL decentration are more common with ciliary sulcus fixation [29].

Primary IOL implantation in the new-born eye is still debated [22, 39]. In unilateral cases the amblyopia is more severe, the occlusion therapy is very hard and the child has an otherwise healthy eye. Therefore it is easier to accept an IOL implantation in theses eyes today, even though there is no available IOL that really fits the small new-born eye. The bilateral cases are often easier concerning contact-lens wear and treatment of the amblyopia. If an IOL is to be implanted it ought to be in the bag with complete anterior and posterior capsulorhexes. A foldable one-piece hydrophobic IOL could be implanted with an injector through a 2.75-mm incision also in this age group. With high-viscosity OVD remaining in the anterior chamber and the IOL implanted in the bag, a dry anterior vitrectomy could safely be performed through the pupil and the two capsulorhexes. The OVD ought to be removed to avoid elevated IOP after surgery. The pupil is closed with acetylcholine (Miochol), and it is wise to ensure that no vitreous is present in the anterior part of the eye. The

sclera is soft and elastic in children and it is hard to achieve a self-sealing incision in most cases. Thus the incision should be closed with a running or horizontal 10–0 nylon suture. Anterior synechia formation to the wound is rather often seen in the youngest. It is important at the end of the procedure to look for this and to have a stable and good anterior chamber. Iridectomy is not necessary in these eyes; this is true also if the eye is left aphakic. However, the lens capsule has to remain in the eye, otherwise it is wise to perform an iridectomy to avoid intra-ocular pressure spikes.

Endophthalmitis is one of the most serious complications after intraocular surgery and prophylactic antibiotics are recommended. Perioperatively, at the end of surgery, injection of 1 mg of cefuroxime (Zinacef) in 0.1 ml saline 0.9 % into the anterior chamber is an effective and safe method [27, 28] to avoid infection. In the new-born eye 0.5 mg of cefuroxime seems to be sufficient. This regime effectively prevents gram-positive bacteria species which is by far the most common bacteria. Prophylactic vancomycin in the irrigating solution during cataract surgery is not routinely recommended by the authors because of possible increased incidence of cystoid macular oedema [4] and the risk of emerging resistance to the antibiotic. The anti-inflammatory treatment should start early after surgery and a perioperative subconjunctival injection of 2 mg of steroids (Betapred) is recommended at the end of the surgical procedure. We have not used protective patches for any children after surgery for many years and have not seen any disadvantages with this regime. On the contrary, the child starts the amblyopia treatment immediately and the parents are very pleased when they are able to establish visual interaction with the child for the first time soon after surgery.

Summary for the Clinician

- **Surgery ought to be centralised**
- **No patching after surgery**
- **Prophylactic antibiotic treatment should take the following forms:**
  - **Rinsing of conjunctiva with 0.05 % chlorhexidine solution or 5 % povidone iodine before surgery**
  - **0.5–1.0 mg cefuroxime intracameral at the end of surgery**

9.3 Cataracts in Older Children

9.3.1 Pre-operative Examination

If the cataract was incomplete at birth close follow-up by a paediatric ophthalmologist is advised. Visual acuity ought to be followed, if possible, with clinical preferential looking grating acuity (Teller acuity card). It is also important to look at the ability to fix and follow and ask the parents if they have visual interaction with the child. Examination of strabismus and binocular functions are also important. In older children visual acuity could be measured with greater reliability. Above the age of 4 years most children could be examined with letters and monocularly. In children not only the visual acuity has to be considered but also the development of amblyopia. If a child has unilateral cataract or a more dense cataract in one eye, occlusion therapy has to be considered. Most of the children with congenital cataract have small eyes and hyperopic glasses should be prescribed if needed.

Nowadays, surgery in children has almost the same indications as in grown-ups with one important exception, if the cataract is too dense, children below the age of 7 will develop amblyopia, which means it is not advisable to postpone the surgery.

In older children, both eyes could be operated during the same surgical intervention or in two sessions, while the development of amblyopia is not as quick as in the youngest. To wait 1–2 months between the eyes is in most cases acceptable. The child and the parents have to be informed before the intervention that accommodation is going to be lost after surgery and spectacles, often bifocal, are needed.

Summary for the Clinician

- **Amblyopia can be avoided by performing surgery**
- **After surgery loss of accommodation is found and glasses are needed**

9.3.2 Surgical Technique

The surgical technique used in children above the age of 1 year does not differ greatly from the technique used in the infant eye. The incision can be sclerocorneal or clear corneal and 12 o'-clock or temporally if preferred.

IOL implantation within the capsular bag is important to decrease after-cataract formation [42] and inflammation. Posterior capsulorhexis is regularly performed until at least the age of 15 years otherwise after-cataract will develop within a short time. Anterior dry vitrectomy ought to be performed at the primary surgery in pre-school children to avoid early after-cataract formation [15, 17, 35]. The authors prefer to perform the vitrectomy through the limbal incision, but others advocate a pars plana approach. With the pars plana approach, separate irrigation is provided anteriorly through a limbal paracentesis incision.

In children with cataract after trauma both the anterior and posterior capsule could be damaged. In cases with relatively immediate cataract development after a corneal wound the anterior capsule is often broken. The condition of the posterior capsule is unknown in most cases, and it is important to avoid hydrodissection, or performing it very carefully to avoid losing lens material into the vitreous. In some cases with blunt trauma, no perforation of the globe and rapid development of dense cataract one ought to consider a damaged posterior capsule. While the posterior capsule is much thinner and more fragile than the anterior one it could also break without perforation of the globe.

In cases with an incomplete anterior capsulorhexis as after trauma, and a broken anterior or posterior capsule, optic capture could be a good option to avoid late decentration of the IOL [10]. The optic is then pushed behind the posterior capsulorhexis, the haptics remaining in the bag. The capsulorhexis has to be in the centre and smaller than the optic, otherwise the IOL optic will escape from the capture. In these cases the lens epithelial cells may grow on the IOL surface forming after-cataract [36]. If performing optic capture in the posterior or anterior rhexis is considered, a three-piece IOL should be chosen rather than a one-piece design. The one-piece acrylic IOL cannot be pushed behind the capsulorhexis since the haptics and optic have to be in the same plane. Also, most of the three-piece lenses are posteriorly angulated and therefore suitable for this technique.

Summary for the Clinician

- **Anterior and posterior capsulorhexis in all children**
- **Anterior dry vitrectomy in younger children below the age of 7 years**
- **IOL implantation is safe, including bilaterally, in children over the age of 1 year**

9.4 IOL Power and Model

These days it is perfectly safe and acceptable to do a primary implant from the age of 1 year, even when both eyes are operated on [11, 41]. Primary implants in younger children is still controversial. In the unilateral cases operated on early, i.e. at only a few weeks of age, a possible option is a primary implant. It has been found that an implant in the capsular bag would decrease the total amount of proliferating lens epithelial cells [42]. The formation of after-cataract is particularly problematic in the youngest patients.

At least in the unilateral cases, implantation of an IOL could be an improvement in the treatment of aphakia, amblyopia, after-cataract and maybe also secondary glaucoma. In this age group the deprivation amblyopia is by far the greatest problem. Aiming for emmetropia at surgery is therefore most appropriate in this age group. Sometimes the desired IOL power is not available because of high hyperopia, in these cases a supplement of a contact lens for some time could be an option. The refraction will change considerably during the subsequent years and the eye will become highly myopic but hopefully not highly amblyopic. Corneal refractive surgery, piggyback implantation or implanted contact lenses are all different options in the future for correction of the myopia.

In bilateral cases the best solution is probably aiming for emmetropia when the child becomes an adult. Depending on the age at surgery the amount of hyperopia will differ. It is important to inform the child and parents before the surgery that the child will probably need bifocal glasses for the rest of his or her life.

Accurate axial length and corneal curvature measurements before surgery are necessary for IOL power calculation. In small children this could be performed during the same anaesthesia as the surgery and in older children it could be performed before the surgery. Eyes with congenital cataract are often shorter than normal eyes and most of the children do need higher IOL power than the average child of the same age.

The inflammatory reaction is more pronounced in children and it is very important to use an IOL with high biocompatibility and accurate clinical documentation.

Capsular bag growth does not continue after lensectomy, which is important when selecting lens implants [38].

A foldable acrylic lens with a sharp edge and an optic diameter of 6 mm is advantageous in decreasing or delaying after-cataract formation in the visual axis and minimising the incision size [12], which is advantageous because a smaller incision results in less post-operative inflammation [23]. By using a single-piece acrylic IOL and the Monarch injector, the incision size could be minimised to 2.75 mm [34]. This is also a very soft IOL and therefore probably suitable for the small eyes of the new-born baby when implanted in the capsular bag [20]. Problems such as breakage of the haptics and retarded ocular growth have been encountered with earlier generation IOLs implanted in the monkey and rabbit eye [19, 21]. The single-piece AcrySof is not recommended for sulcus fixation. Unlike the three-piece design, the haptics are thick and are not posteriorly angulated. Years after surgery decentration and iris chafing can occur. An IOL containing a filter removing the harmful blue light is most probably advantageous for the maculae in these eyes, which will have implants for many years.

Multifocal IOLs should not be considered in young children with a growing eye. The axial length increases during life from a mean of approximately 16.8 mm at birth to 23.6 mm in the adult with a very rapid growth during the first 18 months. The mean refractive power of the cornea decreases from about 51 dioptres at birth to 45 dioptres at 6 months of age and to 43.5 dioptres in adults. Consequently, the refraction will change dramatically during childhood.

Summary for the Clinician

- **In unilateral cases aim for emmetropia at surgery**
- **In bilateral cases aim for emmetropia in adulthood**

9.4.1 Post-operative Treatment

Post-operatively, the eye of a child will tend to react with much more inflammation than the adult eye, particularly in darkly pigmented eyes. Systemic treatment with glucocorticoids is most often not indicated, an exception to this rule being children with uveitis. It is however important to start the topical treatment with dexamethasone 0.1% or another potent topical corticosteroid immediately after surgery. The drops are tapered over 1 month starting with four to five times per day in the elderly. In the new-born the treatment has to be more intense starting with eight to ten times daily and tapered over 2–3 months. In the youngest eyes and

in eyes with dark brown irides, mydriatic drops (tropicamide or cyclopentolate) are administered for several weeks after surgery. Following this treatment regime, synechia formation could more easily be avoided and retinoscopy would be easier to perform before glasses are prescribed.

Immediate correction of the aphakia is mandatory for the best possible visual outcome. There are different options and these days most of the children are implanted with an IOL during primary surgery. Some new-borns are implanted with an IOL; however, in most cases the IOL is not strong enough, leaving some uncorrected hyperopia in the beginning. In theses cases a contact lens is the best option, giving the opportunity to change the strength over time.

If no IOL is implanted contact lenses are best fitted already in the operating theatre for immediate optical correction, to prevent otherwise irreversible deprivation amblyopia. Several types of lenses are available. Rigid gas permeable lenses have a wide range of available strengths and have a great ability to correct large astigmatic errors. They are easy to insert and remove, but cause more foreign body sensation than soft lenses. The two major soft lenses are silicone and soft hydrogel lenses. Both are suitable but soft hydrogel lenses are less expensive which is an important consideration due to frequent lens loss. Loss of lenses and fast eye growth during infancy necessitate frequent lens replacements, especially during the first 2 years of life. Frequent retinoscopy must be performed to decide the power of the lens. Most authorities recommend an overcorrection of +2.0 to +3.0 D until bifocals can be tolerated, which occurs between the age of 2 and 3 years. The child should be provided with aphakic spectacles as an option if contact lenses are unsuitable.

In older children implanted with an IOL, bifocal glasses are prescribed after retinoscopy and the remaining hyperopia is corrected. This is often done at 1 month after surgery at the latest.

Occlusion therapy is started in unilateral cases as soon as the media is clear and the aphakia is corrected, and the therapy needs to be aggressive. Virtually all children with unilateral congenital cataract develop strabismus. In bilateral cases occlusion therapy is sometimes useful if one eye is more amblyopic than the other eye. The occlusion therapy in cases with bilateral cataract usually do not need to be as aggressive as in the unilateral cases.

Close follow-up by a paediatric ophthalmologist is mandatory until the patient is 7 years of age, while untreated amblyopia is soon irreversible. When surgery is performed during the first months of life, life-long follow-up is essential because of the high risk of developing secondary glaucoma.

Summary for the Clinician

- **Intense topical treatment with a strong topical corticosteroid, e.g. dexamethasone, particularly in infants below age 2 years, is recommended**
- **Mydriatic drops in very young patients**

9.4.2 Post-operative Complications

9.4.2.1 After-Cataract

Opacification of the visual axis is the most common complication found after cataract surgery, particularly in the youngest. Even when a posterior capsulorhexis has been performed, growth of lens epithelial cells on the vitreous surface or on the back of the optic can be found some months after surgery (Fig. 9.10). Performing posterior capsulorhexis and dry anterior vitrectomy seems to be one way to decrease opacifica-

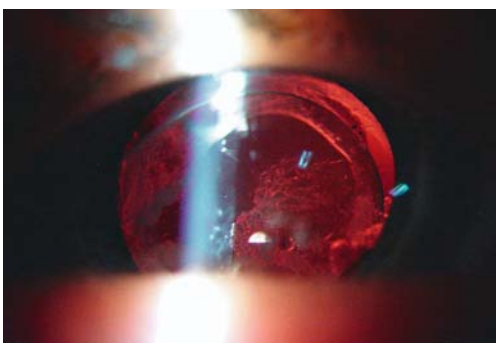

Fig. 9.10. After-cataract 2 years after surgery

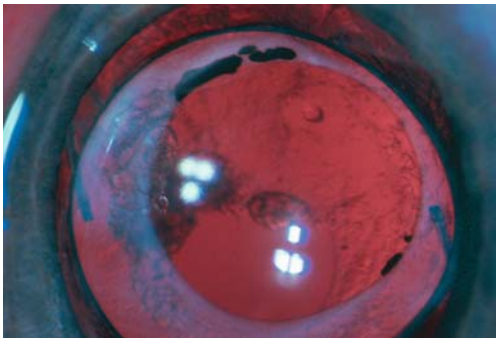

Fig. 9.11. Decentration of the posterior capsulorhexis

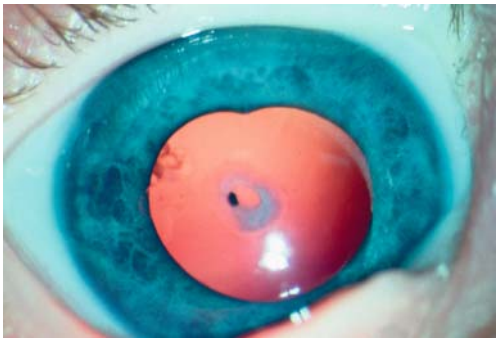

Fig. 9.12. Shrinkage of the capsule with phimosis of the rhexis

tion in the post-operative time [17]. In the very young patients, implanted during their first weeks of life, surgical intervention could be necessary several times, while the lens epithelial cells are growing over and over again on the back of the optic. An IOL implanted in the bag will decrease or prevent formation of Soemmering's ring and the epithelial cells can easier migrate from the periphery to the centre of the pupil. If opacification occurs in the pupil Nd:YAG laser treatment could be tried; however, this is often inadequate in children since reformation of the opacification can be found some months later in these highly reactive eyes. Surgical intervention is often necessary under general anaesthesia and has to be performed promptly to avoid amblyopia. Through a small limbal incision high-viscosity OVD is injected into the anterior chamber to keep it stable during the procedure. An incision in the pars plana and a sharp thin knife is inserted behind the iris and IOL and the membrane is divided. With dry anterior vitrectomy the lens epithelial cells growing in the pupil and also the anterior part of vitreous are removed. At the end of the surgery the pars plana incision is closed with a running suture and the OVD is removed from the anterior chamber to avoid a pressure peak postoperatively.

After the procedure, topical dexamethasone is needed for some weeks. Problems with the lens capsule after surgery, such as decentration (Fig. 9.11) of the anterior or posterior capsulorhexis or shrinkage of the capsular bag with phimosis (Fig. 9.12) of the capsulorhexis could make a secondary intervention necessary. Capsule contraction is more often found in eyes implanted with a silicone IOL [8].

9.4.2.2 Secondary Glaucoma

Secondary glaucoma is unfortunately a common complication and by far the most sight-threatening [2, 25]. The highest incidence is found when the surgery has been done early, that is below the age of 2 months, and a much lower incidence is found when surgery has been performed over the age of 1 year [25]. Eyes with small corneal size, nuclear cataract and persistent foetal vasculature are at greatest risk [30]. Implantation of an IOL into the capsular bag seems to inhibit the development of secondary glaucoma [3].

In the new-born eye a rise in the intraocular pressure will cause epithelial oedema of the cornea, a poor red reflex, photophobia and fast regression of hyperopia or growing eye. These children must be examined promptly under general anaesthesia. Corneal oedema in children wearing contact lenses could be due to hypoxia, and the contact lens must be removed immediately. The anaesthesia of choice is ketamine hydrochloride since it does not lower the IOP which most of the other anaesthetics do. During the evaluation under anaesthesia, IOP, corneal diameter, and axial length are measured and examination of the optic disc and retino-

scopy are performed. Acute glaucoma may develop in cases with excessive inflammation leading to pupillary block and iris bombé. A peripheral iridectomy and an anterior vitrectomy are often sufficient to solve the problem. Later on, a more chronic type of glaucoma could develop probably due to the heavy inflammatory response after surgery leading to synechia formation in the chamber angle and a slow rise in IOP over time [14]. Some of the eyes with secondary glaucoma could be controlled with topical medication but many will need a trabeculectomy with mitomycin C. In some cases also a glaucoma shunt is required for pressure control.

It is important to remember that when cataract surgery has been performed during the first months of life the IOP and optic nerve have to be controlled life-long [1].

9.4.2.3 Fibrinoid Reaction

Because of the high degree of inflammation in children, fibrin in the pupil can be found even when an IOL with high biocompatibility has been implanted. Frequently administered topical steroids and mydriatics are helpful in these cases. In a few cases Nd:YAG laser treatment could be indicated to clear the visual axis. Posterior synechiae formation in the post-operative period is common, especially in the new-born when no IOL has been implanted.

9.4.2.4 Decentration of the Pupil

Incarceration of the iris in the wound is sometimes encountered. To avoid this complication a rather long tunnel is recommended during cataract extraction and suture ought to be used to close the wound. Careful surgery is also helpful leaving the iris without trauma and atrophy. If the visual axis is covered with iris it is important to promptly reposition the iris or make a new central pupil with surgical intervention or Nd:YAG laser treatment.

Summary for the Clinician

- **Good red reflex is essential for favourable development of vision**
- **Rapid growth of the eye should arouse suspicion of secondary glaucoma**
- **Life-long follow-up in cases with surgery within the first months of life because of secondary glaucoma**

9.5 Current Clinical Recommendations

Summarising the previously given information, the following recommendations can be made:

- Prompt surgery is performed in cases with dense congenital cataract – if nystagmus has developed, the amblyopia is irreversible
- Post-operative complications such as high rate of after-cataract and secondary glaucoma are matters of concern in the new-born and life-long follow-up is essential in these cases
- Occlusion therapy ought to be initiated if amblyopia is present in one eye
- IOL implantation could safely be performed above the age of one year
- Anterior dry vitrectomy ought to be performed in pre-school children to avoid after-cataract

References

1. Asrani SG, Wilensky JT (1995) Glaucoma after congenital cataract surgery. Ophthalmology 102:863–867
2. Asrani SG, Wilensky JT (1995) Glaucoma after congenital cataract surgery. Ophthalmology 102:863–867
3. Asrani, S, Freedman S, Hasselblad V, Buckley EG, Egbert J, Dahan E, Gimbel H, Johnson D, McClatchey S, Parks M, Plager D, Maselli E (2000) Does primary intraouclar lens implantation prevent "aphakic" glaucoma in children? J AAPOS 4:33–39
4. Axer-Siegel R, Stiebel-Kalish H, Rosenblatt I, Strassman E, Yassur Y, Weinberger D (1999) Cystoid macular edema after cataract surgery with intraocular vancomycin. Ophthalmology 106:1660–1664

5. Birch EE, Swanson WH, Stager DR, Woddy M, Everett M (1993) Outcome after very early treatment of dense congenital unilateral cataract. Invest Ophthalmol Vis Sci 34:3687–3699
6. Birch EE, Stager D, Leffler J, Weakley D (1998) Early treatment of congenital unilateral cataract minimizes unequal competition. Invest Ophthalmol Vis Sci 39:1560–1566
7. Cheng KP, Hiles DA, Biglan AW, Pettapiece MC (1991) Visual results after early surgical treatment of unilateral congenital cataracts. Ophthalmology 98:903–910
8. Cochener B, Jacq PL, Colin J (1999) Capsule contraction after continuous curvilinear capsulorhexis: Poly (methylmethacrylate) versus silicone intraocular lenses. J Cataract Refract Surg 25:1362–1369
9. Dutton JJ, Baker JD, Hiles DA et al (1990) Viewpoints: visual rehabilitation in aphakic children. Surv Ophthalmol 34:365–384
10. Gimbel HV (1997) Posterior continuous curvilinear capsulorhexis and optic capture of the intraocular lens to prevent secondary opacification in pediatric cataract surgery. J Cataract Refract Surg 23:652–656
11. Gimbel HV, Basti S, Ferensowicz M, DeBroff BM (1997) Results of bilateral cataract extraction with posterior chamber intraocular lens implantation in children. Ophthalmology 104:1737–1743
12. Hollick EJ, Spalton DJ, Ursell PG, Pande MV (1998) Lens epithelial cell regression on the posterior capsule with different intraocular lens materials. Br J Ophthalmol 82:1182–1188
13. Jacobson SG, Mohindra, Held R (1981) Development of visual acuity in infants with congenital cataracts. Br J Ophthalmol 65: 727–735
14. Keech RV, Tongue AC, Scott WE (1989) Complications after surgery for congenital and infantile cataracts. Am J Ophthalmol 108: 136–141
15. Koch DD, Kohnen T (1997) Retrospective comparison of techniques to prevent secondary cataract formation after posterior chamber intraocular lens implantation in infants and children. J Cataract Refract Surg 23: 657–663
16. Kugelberg U (1992) Visual acuity following treatment of bilateral congenital cataracts. Doc Ophthalmol 82: 211–215
17. Kugelberg M, Zetterström C (2002) Pediatric cataract surgery with or without anterior vitrectomy. J Cataract Refract Surg 28: 1770–1773
18. Kugelberg U, Zetterström C, Syren-Nordqvist S (1996) Ocular axial length in children with unilateral congenital cataract. Acta Ophthalmol Scand 74: 220–223
19. Kugelberg U, Zetterström C, Lundgren B, Syrén-Nordqvist S (1996) Eye growth in the aphakic newborn rabbit. J Cataract Refract Surg 22: 337–341
20. Kugelberg M, Shafiei K, Zetterström C (2004) One-piece AcrySof in the newborn rabbit eye. J Cataract Refract Surg, in press
21. Lambert SR, Fernandes A, Grossniklaus HE (1995) Haptic breakage following neonatal IOL implantation in a non-human primate model. J Pediatr Ophthalmol Strabismus 32:219–224
22. Lambert SR, Buckley EG, Plager DA, Medow NB, Wilson ME (1999) Unilateral intraocular lens implantation during the first six months of life. J AAPOS 3:344–349
23. Laurell CG, Zetterström C, Lundgren B, Torngren L, Andersson K (1997) Inflammatory response in the rabbit after phacoemulsification and intraocular lens implantation using a 5.2 or 11.0 mm incision. J Cataract Refract Surg 23:126–131
24. Lewis TL, Maurer D, Tytla ME, Bowering ER, Brent HP (1992) Vision in the "good" eye of children for unilateral congenital cataract. Ophthalmology 99:1013–1017
25. Lundvall A, Zetterström C (1999) Complications after early surgery for congenital cataracts. Acta Ophthalmol Scand 77:677–680
26. Lundvall A, Kugelberg U (2002) Outcome after treatment of congenital unilateral cataract. Acta Ophthalmol Scand 80:588–592
27. Montan PG, Wejde G, Setterquist H, Rylander M, Setterström C (2002) Prophylactic intracameral cefuroxime. Evaluation of safety and kinetics in cataract surgery. J Cataract Refract Surg 28:982–987
28. Montan PG, Wejde G, Koranyi G, Rylander M (2002) Prophylactic intracameral cefuroxime. Efficacy in preventing endophthalmitis after cataract surgery. J Cataract Refract Surg 28:977–981
29. Pandey SK, Wilson ME, Trivedi RH, Izak AM, Macky TA, Werner L, Apple DJ (2001) Pediatric cataract surgery and intraocular lens implantation: current techniques, complications, and management. Int Ophthalmol Clin 41:175–196
30. Parks MM, Johnson DA, Reed GW (1993) Long-term visual results and complications in children with aphakia. A function of cataract type. Ophthalmology 100:826–841
31. Rogers GL, Tishler CL, Tsou BH, Hertle RW, Fellows RR (1981) Visual acuity in infants with congenital cataracts operated on prior to 6 months of age. Arch Ophthalmol 99:999–1003

32. Saini JS, Jain AK, Sukhija J, Gupta P, Saroha V (2003) Anterior and posterior capsulorhexis in pediatric cataract surgery with or without trypan blue dye: randomized prospective clinical study. J Cataract Refr Surg 29:1733–1737
33. Taylor D, Vaegan X, Morris JA, Rodgers JE, Warland J (1979) Amblyopia in bilateral infantile and juvenile cataract. Relationship to timing of treatment. Trans Ophthalmol Soc UK 99:170–175
34. Trivedi RH, Wilson EM (2003) Single-piece acrylic intraocular lens implantation in children 29:1738–1743
35. Vasavada A, Desai J (1997) Primary posterior capsulorhexis with and without anterior vitrectomy in congenital cataracts. J Cataract Refract Surg 23:645–651
36. Vasavada AR, Trivedi RH, Singh R (2001) Necessity of vitrectomy when optic capture is performed in children older than 5 years. J Cataract Refract Surg 27:1185–1193
37. von Noorden G (1978) Klinische Aspekte der Deprivationsamblyopie. Klin Mbl Augenheilk 173: 464–469
38. Wilson ME, Apple PJ, Bluestein EC, Wang XH (1994) Intraocular lenses for pediatric implantation: biomaterials, designs, and sizing. J Cataract Refract Surg 20:584–591
39. Wilson ME, Peterseim MW, Englert JA, Lall-Trail JK, Elliot LA (2001) Pseudophakia and polypseudophakia in the first year of life. J AAPOS 5:238–245
40. Wright K (1995) Visual development, amblyopia, and sensory adaptations. In: Wright KW (ed) Pediatric ophthalmology and strabismus. Mosby Year Book, St Louis
41. Zetterström C, Kugelberg U, Oskarsson C (1994) Cataract surgery in children with capsulorhexis of anterior and posterior capsules and heparin-surface-modified intraocular lenses. J Cataract Refract Surg 20:599–601
42. Zetterström C, Kugelberg U, Lundgren B, Syrén-Nordqvist S (1996) After-cataract formation in newborn rabbits implanted with intraocular lenses. J Cataract Refract Surg 22:85–88