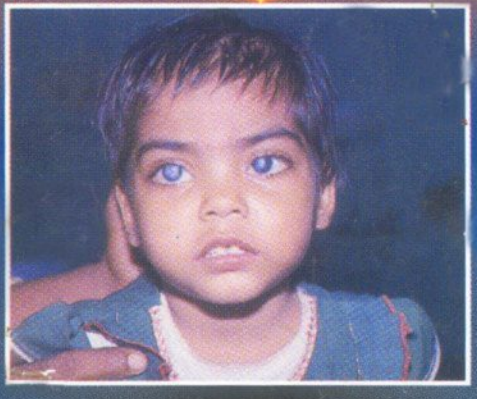
CME SERIES

1998

PAEDIATRIC CATARACT



ALL INDIA OPHTHALMOLOGICAL SOCIETY

By

Dr. Daljit Singh
57, Joshi Colony,
The Mall,
Amritsar - 143 001

Published by :

ALL INDIA OPHTHALMOLOGICAL SOCIETY
Dr. Rajendra Prasad Centre for Ophthalmic Sciences,
ALL INDIA INSTITUTE OF MEDICAL SCIENCES,
Ansari Nagar, New Delhi - 110 029

(as a part of the CME Programme)

For any suggestions, please write to :

DR. H.K. TEWARI
Hony. General secretary

CONGENITAL AND INFATILE CATARACTS

Dr. Daljit Singh

The opacification of any part of the crystalline lens is termed cataract. It may be unilateral or bilateral. The degree of opacification and the number and size of the opacities may vary. The consistency of the cataract varies from fluid to stony hard (if the lens is calcified). Some of the common forms of congenital cataract are central, total, zonular, cerulian, sutural, anterior or/and posterior subcapsular, posterior polar, axial (corraliform) and anterior pyramidal. Rare anomalies of the lens include anterior lenticonus and posterior lenticonus. The lens size varies from less than 6 mm to 8 mm. Persistent hyperplastic primary vitreous may be seen on the back of the crystalline lens. The posterior polar cataract may be thick and may have blood vessels that are derived from the hyaloid canal or from the ciliary processes. Long ciliary processes on the back of the lens carry the blood vessels.

Many congenital cataracts show a pre-existing opening in the posterior capsule. The appearances of the affected lens varies. There is variable amount of absorption of the lens. The anterior chamber may be deeper than normal. One may find a totally absorbed cataract with a few remaining shreds of the capsule. There may be a dense white central depressed cataract surrounded by a ring of the cortex (dumbell shaped cataract). The cataract may be in the form of a translucent thin flat bag containing some opalescent fluid, or it may be filled with milky fluid. Some cataracts show a dense white thick anterior capsule, while the lens matter behind it may have any consistency ranging from milky fluid to hard opaque material. In some cases the cataract is partly absorbed and partly mixed in the anterior vitreous, where it shows movement with the movement of the eyeball. Dense central cum posterior cataract might show dense white spots at or near the edge of the opacity.

Chalky white spots, usually arranged as a cluster or in a roughly circular form, may be seen at the level of the posterior capsule, before surgery or, during the course of operation and indicate the presence of a pre-existing posterior capsule opening (Singh's sign).

The congenital cataract may be accompanied by ocular abnormalities like microcornea, megalo-cornea, coloboma of the iris, aniridia and zonular dehiscence etc. Catarract may form a part of a wider syndrome.

Traumatic cataract has variable presentations, depending upon the nature and severity of the injury and the length of time that has elapsed between the injury and the time of examination. Rossette cataract is a well known result of blunt injury. The anterior and/or the posterior capsule may be ruptured. There may be subluxation of the cataractous lens, with disturbance of the vitreous. There may be an avulsion of thei iris, iridodialysis, traumatic mydriasis with rupture of the sphincter muscle, lens-iris adhesions, formation of a cornea-iris-lens complex etc. The blood vessels may invade the cataract from the uveal tissues. Glaucoma and chorio-vitreo-retinal problems may coexist in these patients. Signs of uveitis may also be present.

Etiology/Incidence

In most cases of congenital cataract, no cause can be found. In others, it may be associated with :

- Systemic syndromes
- Dermatological disorders
- Other ocular abnormalities
- Metabolic disorders
- Chromosomal disorders
- Intrauterine viral infections especially rubella
- Injuries
- Long term steriod use.

In most cases, the etiology of paediatric cataract remains undiscovered, but heredity is important. The inheritence may be autosomal dominant, autosomal recessive, X-linked or sporadic. The incidence is higher in the developing countries, due to high incidence of consanguinity, large families and lack of genetic counselling. Maternal rubella in the first trimester, complications of pregnancy, ingestion of drugs, exposure to X-rays and malnutrition are also

important factors predisposing to the occurence of cataract in neonates and infants. Injuries from poorly designed toys, especially bow and arrow, fire crackers and unprotected sports are important causes of traumatic cataract in our country.

Course/Prognosis

Many forms of congenital cataract remain stationary for years and decades. If macula does not receive sufficient stimulus, amblyopia, strabismus and nystagmus may develop. Amblyopia develops more rapidly in complete unilateral cataract. Congenital or traumatic dislocated crystalline lenses often become opaque. Traumatic cataracts especially after perforating injury generally progress rapidly. Glaucoma and iridocyclitis are common associated features. Many untreated or poorly managed perforating injuries with vitreous and ciliary body involvement lead to rapid hypotony and phthisis bulbi. An unattended retained foreign body worsens the prognosis. Timely pars plana lensectomy, vitrectomy and foreign body removal coupled with lens implantation provides the best hope for such cases.

If the visual axis is cleared early enough, by an appropriate atraumatic surgical technique, there ought to be more or less improvement in every case. If an optical correction like aphakic glasses or contact lens is accepted, the improvement is better. However, a successful intraocular lens implant provides the best vision, central as well as peripheral. The surgically treated unilateral congenital cataract cases improve, if timely and prolonged amblyopia exercises are given. Bilateral cases do better, provided the surgery is not much delayed. Traumatic cataract cases recover good sight if the transparency and the curvature of the central cornea is preserved and the macula has escaped injury. Good eyesight may be restored in many cases of traumatic endophthalmitis who undergo pars plana vitrectomy and lensectomy. Secondary lens implantation after a suitable waiting period is usually successful.

Delayed milestones and mental retardation cases may not show a palpable appreciation of visual improvement after cataract surgery.

Diagnosis/Lab Findings

The clinical diagnosis is clinched by:

- Oblique illumination
- Direct ophthalmoscopy
- Slit lamp examination
- B-scan in a case of dense cataract helps in the diagnosis of a break in the posterior capsule and presence of a lens-vitreous mix. The presence of vitreous exudates, hemorrhage, a foreign body, choroidal detachment, retinal detachment or a retinoblastoma are also diagnosed by ultrasonography.
- X-ray examination is important in many cases of perforating injuries.

If the anterior part of the lens is opaque, the parents notice it. At other times, lack of fixation to light, nystagmus, squint and failure to notice toys and parents become the cause of concern. Cases of mild or moderate zonular cataract might remain unsuspected for many years. The only disability in these patients may be photophobia to bright light.

The list of possible associations or causes of infant cataract is long. It is therefore not practical to order extensive laboratory investigations to cover every possibility. In a child with cataract, who is otherwise healthy, two tests seem important:

- Galactokinase
- Test for rubella

The first test is helpful. In early positive cases, elimination of galactose from the diet can cause a regression of cataract as well as the systemic manifestations. A positive test for rubella fore-warns about the possibility of slightly increased postoperative reaction.

Differential Diagnosis

The usual differential diagnosis in a patient with paediatric cataract is :

- Remains of pupillary membrane
- Exudates in the pupillary area

- Persistent hyperplastic primary vitreous
- Retinoblastoma
- Endophthalmitis
- Retinal detachment

Prophylaxis

Can paediatric cataract be prevented? The question assumes importance in view of the complexities involved in success rehabilitation of a child with cataract. Some measures which could undoubtedly bring down the incidence of this problem include :

- Compulsory rubella vaccination for all girls after the age of 12-a country wide campaign is needed;
- Protection of children from fire crackers and projectile throwing toys;
- Eye protection devices should be used in all sports that can cause accidental injury;
- The children should be taught to handle sharp and pointed writing and other tools in their armoury very carefully;
- Avoid prolonged use of local steroids for treating such conditions as spring catarrh;
- Consaguinous marriages should be discouraged;
- Genetic counselling in the event of even single case of congenital cataract in a family. When more than one case of congenital cataract occurs, molecular genetic study should be undertaken. However precise localisation of the genes for most forms of congenital cataract and their linkage to specific candidate genes continues to be elusive. Unless these genes are characterised at molecular level or an extremely strong linkage established with some molecular markers, it would not be possible to prevent the recurrence of congenital cataract or to undertake its prenatal diagnosis.

Treatment

Options available for management of a case of paediatric cataract include :

- Mydriatics
- Optical iridectomy
- Cataract surgery

For most cases of paediatric cataract, surgery remains the only treatment. The desirability of intraocular lens implantation is being increasingly appreciated. However, the selection of lens design and the mode of application varies due to personal reasons and the nature of the laws in a country. In general, however, an ideal intraocular lens is one which is :

- Well tolerated by the ocular tissues
- Should be suitable for implanation in the diverse paediatric situations
- Once in place, it should not change its position
- The lens and the surrounding tissues should be available for inspection
- It should not come in the way of managing behind the iris developments like secondary cataract and thick membrane formation
- Should not get entangled in the tissue reactions
- In the event of the development of gross refractive error, subluxation or dislocation or a chronic inflammation, it should be possible to explant or exchange the intraocular lens with minimal trauma. In short it should be a fail-safe lens.

The eye of a new born differs from the adult eye in many respects. The eyeball length is only 16.5 mm (adult 23.5 mm) and the volume is 2.8 ml (adult 7.1 ml). The ciliary body is markedly cellular. The ciliary processes touch the iris. The ciliary body is small in size, an so is the pars plana. The ora serrata develops by the seventh year. The lens diameter is about 6 mm (adult 9.1 mm). By 20th year the lens diameter is 8 mm. The corneal diameter at birth is 10 mm (adult 12 mm). It is obvious that the tissues in the neonates, meant to receive posterior chamber lenses are still not developed in size and structure. No suitable size and design of posterior chamber lens is available for use in these eyes. The use of adult size intraocular lenses in anatomically immature tissues is less than safe. The white

to white diameter of the cornea is also not final. For this reason, no design of angle supported lens is suitable for implantation in infants.

In the new born, the anterior chamber depth is about 2.5 mm (adult 3.2 mm). When the cataract is removed, the chamber depth becomes comparable to the adults. The iris and pupil are well developed. The sphincter muscle is fully developed, the dilator less so. An intraocular lens attached to the anterior surface of the iris, as does an iris-claw lens, overcomes the physical problems of size and design for infant eyes. The lens is suitable for use even in the absence of the posterior capsule and in cases of megalocornea. Small lenses can be used in cases of microcornea.

From the point of view of tissue reaction, there are only two kinds of intraocular lenses-those inside the bag and those outside. All those lenses that are outside the bag are uvea supported. Scleral spur support in the angle is a myth, since only a microscopic tip of the scleral spur is visible in the angle on gonioscopy, that too covered by the corneo-scleral trabeculae. The angle ends of the ciliary body, where most if not all the flexible loop anterior chamber lenses will impinge and erode. The "sulcus" supported posterior chamber lenses will likewise erode the ciliary body. If uveal tissues were non-responsive, these erosions would not matter. Unfortunately the uveal tissues respond. However the response is different when iris and ciliary body are compared. The response is practically nil from the iris, but is clinically important from the ciliary body. They ciliary vasculature is designed to leak. The capillaries are almost venular in width. They are 15-30 microns in diameter, fenestrated (30-100 mm) and permeable to plasma proteins and blood borne tracer materials. Injury and erosion of the epithelium of the ciliary body produces rapid breakdown of blood-aqueous barrier. Erosion, inflammation and scarring in the area of the ciliary body can produce changes in the position of the intraocular lens. In contrast, the iris vasculature is designed to resist leakage. The endothelial cells of the capillaries are non-fenestrated. Their cell junctions and the gap junctions are tight and there is a thick multilayered basal lamina, outside which are numerous round and oval bodies. All arterioles and venules have a tube in a tube structure, the vessels being surrounded by elastic, fibrous and muscular tissue.

While the capsule of the crystalline lens is inert, the same can not be said about the germinal cells at the equator and the cells of the anterior capsule. Inflammatory, proliferative and fibrotic reactions occur inside the capsular bag. This may be compounded by any retained lens matter. The changes in the capsular bag cause decentration of the optic, or its exteriorisation from the bag and deposition of inflammatory cells on the optic.

Opacification of the posterior capsule is a matter of serious concern in every child undergoing cataract and implant surgery. The onset and severity of the secondary cataract formation is related to the size of the anterior capsule retained. The secondary membrane may be so thick that it can not be cut with YAG laser. Aggressive YAG laser treatment only increases the number of pits on the optic of the lens. Vacuum cleaning of the anterior capsule during the surgery perhaps reduces the problem. On the table posterior capsulorrhexis, anterior vitrectomy and pushing the optic backwards, through the posterior capsular opening, is being tried to overcome some of the in-the-bag lens problems. Subtotal anterior capsulectomy and sulculs supported or iris-claw lenses minimise and delay opacification. In angle support and iris-claw lens cases, there is a space between the lens optic and the secondary cataract. That makes it easy to do Nd: YAG capsulotomy by setting the focus on the capsule and without ever hitting the lens optic. Manual cutting of a thick membrane behind the lens is also easier than with the posterior chamber lenses.

The intraocular lenses may also be known by the way they get fixation. Except an iris claw lens, all lenses are oversized for the site meant for fixation. The lens is pushed into the desired place, so that the eye tissues catch the lens. An iris-claw lens, with a maximum width of 6 mm to 7 mm and an optic of 4 mm, is much smaller than the space where it is fixed. This lens has to be positively fixed to the iris by the surgeon himself. It is away from the angle of the anterior chamber, away from the endothelium and is totally free from the pupil.

All the benefits of the intraocular lens are derived from the optics. All the problems arise from the haptics. The iris-claw lens has the

smallest haptics. Micro and macro trauma in the daily life of children can cause friction and erosion at haptic-tissue junction in the angle supported and the sulcus supported intraocular lenses. A severe injury can open the claw and produce dislocation of an iris-claw lens into the anterior chamber. In such an eventuality, the lens can be explanted or exchanged without difficulty.

The possibility of a life long observation of the intraocular lens and the surrounding tissues is important. In case a problem arises that necessitates explantation, it should be possible to do the same atraumatically. One such situation may be a chronic inflammation, that refuses to go. Or else a gross change in refraction occurs, so that lens exchange becomes necessary. It is obvious that it will be "fficult to manage intraocular lenses in the sulcus or in the bag in ıis manner.

A large number of paediatric aphakes need secondary lens implants. The posterior capsule may or may not be present. A lens implant system should solve this problem with minimum possible trauma. The integrity of the vitreous and the retina should not be jeopardized. An iris claw lens comes handy in all situations. Scleral fixation of a posterior chamber is more traumatic. It also introduces a long term uncertainty of biodegradability of the holding nylon or polypropylene sutures, that are used.

Cases with traumatic cataract are often a great challenge to the ingenuity of the surgeon, in overcoming the tissue adhesions, damage, distortion and tissue losses, and yet being able to provide a corrective optic. Needless to say, the surgeon should be able to pick a lens design that suits the specific situation.

Surgical Management Techniques

ı nere are three ways to operate paediatric cataract.

- **Anterior approach :** The entry into the anterior chamber is made through pocket sections at the cornea, limbus or the sclera. The corneal endothelium is protected and the anterior chamber deepened by viscoelastic materials. Anterior capsulectomy techniques take into account the age of the patient, the type and consistency of cataract, and the type of intraocular lens to

be implanted. It may vary from a 4 mm capsulorrhexis to a subtotal anterior capsulectomy. The cataract may be removed by one port or two port irrigation/aspiration. Harder lenses may be removed either by phakoemulsification or are delivered manually through enlarged incisions. A posterior chamber intraocular lens may be implanted in the bag or in the ciliary sulcus. For implantation in the anterior chamber, there are two choices - an angle supported lens and an iris claw lens. The iris claw lens grips the anterior surface of the iris at two points 180 degrees apart, beyond the collarette. Another choice, sensible but sadly rarely used, is Binkhorst lens in which the loops of the lens are lodged in the capsular bag, while the optic lies in the anterior chamber.

- **Pars plana/plicata approach :** This approach is used, when no intraocular lens implantation is intended. An attempt is made to remove the entire cataract and the adjacent vitreous with a vitreous cutter.
- **Combined anterior and pars plana/plicata approach :** The aim is to implant a posterior chamber lens. This is done in two steps. By the anterior route, the intraocular lens is placed in the sulcus, without dealing with the cataract. The incision line is closed. The pars plana/plicata route is then used to remove anterior vitreous, the posterior capsule, the cataract and the central part of the anterior capsule.

Dislocated lenses, which are usually congenital, but also sometimes traumatic, are treated by removal in toto, followed by such lens implantation as is not dependent upon an intact posterior capsule. This includes options like a scleral fixated posterior chamber lens, iris-claw lens or an angle supported lens.

For traumatic cataract following blunt or perforating injuries, the surgical management has many facets. The aim is best restoration of the anatomy, free flow of aqueous humour, removal of cataract, clearing of the visual axis and the fixation of such intraocular lens, as is suitable for the individual case.

Complications

The paediatric eyeball is elastic, which predisposes it to many intraoperative problems. Small port anterior route cataract extraction is safe in these patients. Larger incisions allow rapid collapse of the anterior chamber, forward push of the iris, lens and vitreous. intraoperative iris prolapse, fibrin formation, bleeding from the root of the iris, rupture of the posterior capsule and lens-vitreous mix. These problems are minimised by making a 4 mm or larger pocket section, deepening the anterior chamber with viscoelastic and by using a low vacuum for aspiration. A collapsed anterior chamber during the operation encourages the formation of fibrin. Pars plana approach avoids anterior chamber problems, but the chances of complete cataract removal, lens-vitreous mix and vitreous heamorrhage are increased.

Early and late postoperative complications are related to surgical trauma, the retained anterior lens capsule lens matter, lens-vitreous mix, blood and fibrin released during surgery and the type and position of the implanted intraocular lens.

A rise of the intraocular pressure in the post operative period common in cases with a lens-vitreous mix. Postoperative inflammation is likely in many traumatic cases. Exudates may form in the pupil, in front and behind the intraocular lens. PC IOL-iris adhesions produce iris bombe, if a patent peripheral iridectomy does not exist.

In well operated cases, late problems are connected with inflammatory, proliferative and fibrotic reactions. Secondary cataract formation is common. There may be partial or complete pupil capture, and/or decentration or dislocation of a posterior chamber lens. Posterior synechiae and iris-bombe may also occur.

Postoperative Management

In the postoperative period, it is important to prevent and keep under check any tendency for the occurence of an inflammatory reaction. Prolonged use of local steroids, non-steroidal anti-inflammatory agents and atropine are best for this purpose. Oral NSAID and sometimes even steroids may be prescribed as needed.

The eye needs to be watched regularly for the development of a secondary cataract or any other early or delayed peoblem. For the prevention of amblyopia, active vision therapy needs to be started early and pursued vigorously. Intraocular lens implantation without active vision therapy is work half done. It goes without saying that life long careful follow up is essential in all operated cases of paediatric cataract, especially those with intraocular lenses.

Conclusion

Compared to adult cataract surgery, the surgical challange in the congenital and infantile cataract is far more complex. Every patient needs a personalised appropriate management, be it in the manner of cataract surgery, the selection of the intraocular lens of its actual application, early and late postoperative follow up and management of various expected and unexpected problems and the steps that are taken to prevent or to treat amblyopia. If the available technology is applied diligently, the results are extremely gratifying.

Suggested Reading

- *Born A. J., Tripathi R.C. and Tripati B.J: Wolff's Anatomy of the Eye Orbit. Chapman & Hall, London, 1997; 217-220, 280 and 398-405.*
- *D Singh D., Worst J., Singh R. And Singh I.R.: Cataract and IOL, Jaypee Brothers Medical Publishers, New Delhi,; 1993; 160-168, 189-200.*
- *Foster A., Gilbert C., and Rahi J.: Epidemiology of cataract in childgood: A global perspective. J. Cataract Refract Surg. 1997; 23: 601-604.*
- *Gimbel H.V.: Posterior continous curvilinear capsulorhexis and optic capture of the intraocular lens to prevent secondary opacification in paediatric cataract surgery. J. Cataract Refract Surg. 1997. 23: 652-656.*
- *Verma A. and Singh D.: Active vision therapy for pseudophakic amblyopia. J. Cataract Refract Surg - 1997; 23: 1089-1094.*