

## INTRODUCTION

Bilateral congenital cataract is the most common cause of treatable childhood blindness, accounting for 5% to 20% of blindness in children worldwide. In the developing world, the occurrence of loss of sight from cataract is higher, about 1 to 4 per 10 000 children. (*Foster et al., 1997*)

Childhood cataract blindness presents an enormous challenge to developing countries in terms of human morbidity, economic loss, and social liability. Managing cataracts in children remains a difficult task, with a treatment plan that is often demanding and requires a collaborative team work. To assure the best long term outcome for children with cataract appropriate pediatric surgical techniques need to be defined and adopted by ophthalmic surgeons of developing countries and proper assessment and follow up need to be undertaken in order to achieve best results of visual outcome. (*Wilson et al., 2003*)

Increased intraoperative difficulties, inclination for augmented postoperative inflammation, changing refractive state of the eye, and a tendency to develop amblyopia, all add to the difficulty in achieving a good visual outcome in the pediatric patient. Adaptation of techniques for cataract surgery specific to children is necessary owing to low scleral rigidity, increased elasticity of the anterior capsule, and high vitreous pressure. Also, microphthalmia and pupillary miosis often add to the surgical complexity. Finally, surgical timing and adequate visual rehabilitation are vital, to avoid irrevocable visual damage secondary to amblyopia. *(Taylor et al., 1999)*

In unilateral cases, posterior capsular opacification (PCO) if the capsule remains intact, occurs in 51% to 100% of cases in pediatric cataract surgery and prevents visual rehabilitation. *(Dahan et al., 1990)*

## **AIM OF THE STUDY**

To evaluate and compare the results of 2 intraocular lens (IOL) implantation techniques in pediatric cataract surgery after performing anterior and posterior capsulorhexis and anterior vitrectomy, namely optic capture of the implanted three piece acrylic intraocular lens in both anterior and posterior capsulorhexis in one group, to implanting the same IOL in the capsular bag in the other group.

# **Chapter (1)**

## **Congenital cataract**

### **Etiology**

In developed countries, the leading reason of congenital cataracts is usually idiopathic. Some of the presentation might be due to hereditary disease, with no associated systemic condition. (*Bardelli et al., 1989*)

Cataracts due to hereditary causes are different in their genetic categorization. Autosomal dominance is common in lots of cases, but autosomal recessive and X-linked cases may be present as well. (*Wright et al., 1995*)

On the other hand metabolic disease are a much less common reason for the occurrence of cataracts. Causes such as galactosemia and hypocalcemia are less commonly encountered. (*Beigi et al., 1993*)

Cataract that is congenital can be associated with conditions such as trisomy 21 and Turner's syndrome. It is not uncommon to have cases presenting with mental retardation and congenital cataract. Syndromes such as craniofacial or skeletal deformities, myopathy, spasticity or

other neurological disturbances may also be encountered with inherited cataracts. (*Lambert et al., 1997*)

Genes involved in cataractogenesis have been commonly found in research. Infections such as toxoplasmosis, rubella, herpes infection and syphilis can all be the reason for a number of congenital cataracts. (*Lambert et al., 1989*)

Rubella is by far the most significant cause of cataract, that is mostly affecting both eyes rather than one. Unilateral congenital cataracts are more commonly unrelated to the occurrence of systemic disease and are commonly not inherited. Lenticonus/lentiglobus and persistent fetal vasculature (PFV) are not uncommonly encountered. (*wright et al., 1995*)

## **Morphology**

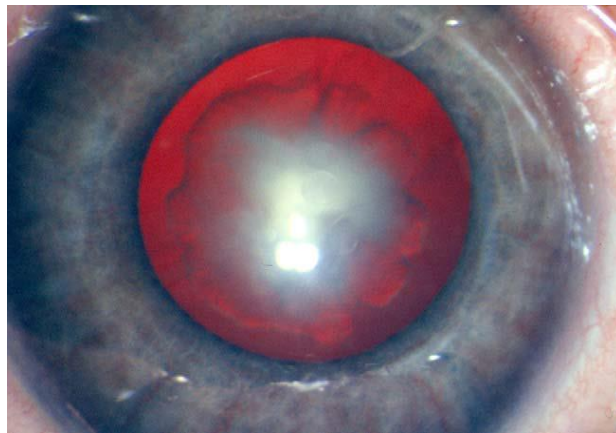
### **Nuclear cataract**

Children with this kind of cataract usually present with it early on in life. In cases that present with cataract dense enough to affect visual development, delay of surgery is avoided. (*Parks et al., 1993*)

The position of the opacity is usually between Y sutures and usually located more in the center; thus affecting vision

markedly (figure 1). Microphthalmia is not uncommon in such children. (*Kugelberg et al., 1996*)

In up to 50% of cases where inheritance is the case, bilateral nuclear cataract is observed and autosomal dominance is present.



**Figure (1):** Nuclear cataract (*Kugleburg et al., 1996*)



**Figure (2):** Posterior polar cataract (*Vasada et al., 2004*)

## **Posterior Cataract**

Children with persistent fetal vasculature more recently referred to as persistent hyperplastic primary vitreous and who commonly have microphthalmia. These present with cataract that is more posterior in location and thus more vigorously affecting vision. It is quite a challenge to operate such cases because weakness of the posterior capsule can cause intraoperative complications and should be considered before surgery. (*Vasavada et al., 2004*)

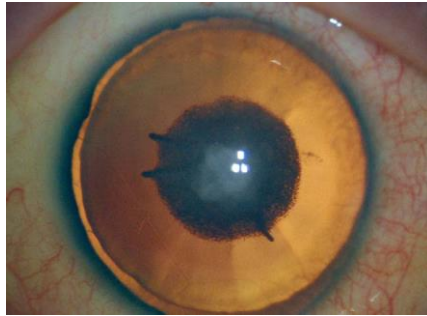
## **Lamellar Cataract**

A progressive subtype that appears later in life with opacification occurring around the Y sutures of the lens. This type is not associated with microphthalmia, has a high rate of bilaterality and usually of autosomal dominant inheritance. (*Park et al., 1993*)

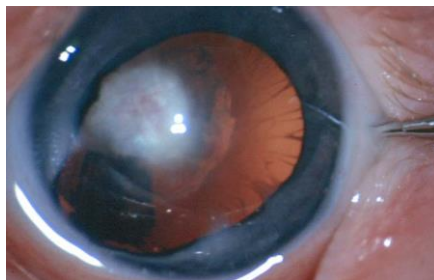
## **Other Morphological Types of Cataract**

Those are usually because of defects in the development of the lens that were present earlier in life and less commonly have an effect on visual acuity and are less liable to progress. Those include sutural and anterior polar cataracts. (*Arkin et al., 1992*)

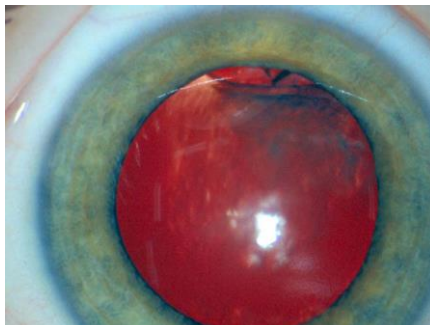
Ocular conditions such as aniridia (Figure 3), iris coloboma (Figure 4), and lens coloboma (Figure 5) are often seen with cataract. (*Nelson et al., 1984*)



**Figure (3):** nuclear cataract in a case of aniridia. (*Charlotta et al., 2005*)



**Figure (4):** A case of iris coloboma associated with cataract. (*Charlotta et al., 2005*)



**Figure (5):** A case of lens coloboma associated with cataract. (*Charlotta et al., 2005*)



## **Chapter (2)**

# **Management of Childhood Cataract**

### **Amblyopia in childhood cataract:**

One of the biggest challenges in children with cataract is the management of Amblyopia; a neurological functional developmental abnormality. The possibility of this change is related to how well developed the visual system was at the start of the deprivation process. This is most evident when deprivation occurs in the most critical age of visual development, in the first 2 years of the child's life. Early intervention is therefore of the utmost importance at a very early age in congenital cases in order to avoid the development of nystagmus and permanent amblyopia. (*Dutton et al., 1990*).

The more dense and more posteriorly located the opacification the more visually significant the affection and the more the importance of its management. The density of the cataract is assessed in term of clarity of large vessels in the fundus as an indicator of its relevance.

In cases of subclinical cataract the visual development and the affection of binocular vision is used to assess which cases should be conservative and which ones should be operated. Weighing the risk and benefit of surgery should be well calculated.

The clinical evaluation should entail evaluation of visual behavior including fixation patterns. (*Charlotta et al., 2005*)

## **Treatment plan**

A treatment plan based on surgery within 2 months of birth combined with prompt optical correction of the aphakia and aggressive occlusion therapy with frequent follow-up has been successful in unilateral and bilateral cases. Both anterior and posterior capsulorhexes are performed in most children. Intraocular lens implantation can be executed safely in children older than 1 year. Anterior dry vitrectomy is recommended in preschool children to avoid posterior capsular opacification. (*Zetterstro et al., 2005*)

At one point, lensectomy was the procedure most commonly executed in treatment of such cases. Nonetheless, in recent times, small incisions, anterior capsulorhexis, bimanual irrigation/aspiration, and primary posterior

capsulectomy and vitrectomy have become standard treatment options for infantile cataract. IOL implantation has become the standard procedure of the optical rehabilitation of children with cataract, from the toddler age group and up. (*Vasavada et al., 2012*)

### **Development of technique of cataract surgery**

Over the past years, improvements in technology and adjustments in surgical procedures have brought pediatric cataract surgery to a new horizon. Automation and the use of intraocular lenses (IOLs) have enhancement in the anatomical and functional outcomes in children. (*Vasavada et al., 2012*)

The mid-1970s foreshown a revolution in pediatric cataract surgery with the institution of vitreous suction-cutting devices. By enabling primary posterior capsulotomy and anterior vitrectomy at the time of primary surgery, these devices allowed surgeons to overcome the difficulties of cataract and management of vitreous in the anterior chamber associated with lens aspiration and needling. For nearly three decades, the 20-gauge vitrectomy system has been the primary weapon in the pediatric cataract surgeon's armamentarium. (*Chee et al., 2009*)

As mentioned before the most common complication to be addressed in pediatric cataract surgery is PCO thus neodymium: yttrium aluminum garnet (Nd:YAG) capsulotomy was previously used to deal with it however it does not provide a long-lasting clear visual axis because lens epithelial cells (LECs) find their way to the posterior surface of the lens, not to mention the technical difficulty in children. (*Nishi, 1988*)

The transformation of residual LECs results in dense membranes on the anterior hyaloid surface, visual axis re-occlusion, haptic displacement, and iris capture. (*Hiles, 1990*)

To provide better conditions for visual restoration, primary posterior capsulotomy and anterior vitrectomy have been endorsed. (*Ravishankar et al., 1996*)

There are 2 approaches to primary posterior capsulectomy and anterior vitrectomy: through the limbus and through the pars plana. Even though more recent studies have shown promising results of pediatric IOL implantation, caution in selecting patients for this procedure is critical, especially in very young children where incidence of acquired glaucoma is very significant. Visual rehabilitation of unilateral aphakic

children is more challenging than bilateral cases. Thus, the minimum age for which IOL implantation should be considered in unilateral cases of pediatric cataract is lower than that for bilateral cases and that is also related to the practical difficulty of the use of contact lenses in young children which is not always very applicable. (*Wilson et al., 1994*)

In a survey, the mean age for IOL implantation in children was 3 years in unilateral cases and 5 years in bilateral cases .In calculating IOL power, the goal is to achieve emmetropia and reducing anisometropia. This is a significant issue in visual rehabilitation in children. The correct IOL power leads to more successful amblyopia treatment and increases the chance of binocularity in this critical age period. However, different approaches have been considered in IOL power calculation. In one study, under correction was the objective in patients younger than 9 years and yielded good results owing to the avoidance of the highly expected myopic shift especially in younger age group. (*Dahan and salmenson, 1990*)

Some opinions consider this a slightly extreme measure and choose to use under correction only in children 4 years or younger. The under correction was in an effort to counteract

the myopic shifts same as in the work of Dahan et al but in a different protocol. Other authors believe that achievement of emmetropia is vital in patients with a pliable visual system and that its benefits overshadows the consequence myopia. The growth of the globe is almost complete at 2 years of age. Thus, the myopic shift in children 3 years and older would be estimated to be reduced. (*Gimbel, 1993*)

To provide clear media for visual rehabilitation, primary posterior capsulotomy has been instituted. Several approaches to prevent or delay PCO have been studied. Primary posterior capsulotomy with a needle or a vitrectomy cutting device, a limbal or pars plana approach anterior vitrectomy, and primary posterior continuous curvilinear capsulorhexis (PCCC) are some techniques that have been evaluated. (*Zetterstro et al., 1994*)

The capsulotomy edge produced by continuous curvilinear capsulorhexis is smooth. However, performing PCCC is technically difficult with highly elastic capsule with a high tendency of extension; a procedure in need of experience in that field. Capsulorhexis of the anterior capsule in young patients is even more challenging than PCCC. (*Koch et al., 1997*)

There is no agreement whether the IOL should be implanted before or after the primary posterior capsulectomy and anterior vitrectomy. Some support removing the posterior capsule and anterior vitreous before IOL implantation. (*Basti et al., 1996*)

When the anterior vitrectomy is done, in-the-bag IOL implantation is easily achieved. Dahan and Salmenson found no grave complications using this technique in 80 children. Other surgeons have a preference to have the IOL in place before removing the posterior capsule to allow for an adequate posterior capsulectomy and a better anterior vitrectomy. The advantage of implanting the IOL before the posterior capsulectomy is that the IOL can be safely situated in the desired plane. The risk of the IOL being pushed out of the capsular bag may be minimized by keeping the infusion in the anterior chamber. (*Buckley et al., 1993*)

## **Historical Perspective of IOL Capture Technique**

With pediatric IOL implantation surgery, there is a high incidence of posterior capsule opacification and retro-pseudophakic membrane formation. Lens epithelial cells proliferate, form Elschnig pearls, and use the posterior capsule, anterior vitreous face, and face of the IOL as a