Congenital Cataract

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ABSTRACT

Congenital cataract is a leading cause of childhood blindness. Early diagnosis and treatment are important because of the high risk of amblyopia. Newborn babies should be screened early. Often the cataract is regarded as idiopathic; it can be genetic but is frequently associated with other systemic or ocular abnormalities which can influence the final visual outcome. Visual rehabilitation of congenital cataract includes optical correction of aphakia and amblyopia treatment.

Keywords: Congenital cataract, aphakia, ambliyopia.

Congenital Cataract (CC) is defined as any opacity of the crystalline lens of the eye at birth or detected within the first year of life . CC one of the leading causes of preventable blindness in pediatric ages and are responsible for 5% to 20% of blindness in children worldwide.[1-5] Incidence varies from country to country. Incidence is 1-5/10,000 in economically developed countries and 5-15/10,000 in developing world. Highest prevalence observed in Asia.[2]

Congenital cataract impedes the image clarity in the critical time causing reduced visual acuity and impaired contrast sensitivity and caused amblyopia. Visual acuity in children may be impaired as a result of amblyopia due to closure of the optic axis, anisometropia and related amblyopia may develop in some cases due to high degree of astigmatism.[3-4-6]

CCs can be seen unilateral or bilateral and different from senile cataracts in terms of their etiologic, clinic and morphologic characteristics.

Clinical Presentation

Early diagnosis and surgery-if needed, appropriate refractive error correction, amblyopia therapy and long-term follow-up are important for CC.

Cataract may become obvious to a parent or a pediatrician as a white reflex in the pupil (leukocoria). Red reflex examination at birth is an easy method to screen for congenital cataracts[7]. Sometimes parents may notice that their young child shows lack of visual attention to the environment. Another important presenting symptom may be nystagmus of the searching or wandering type, that is the nystagmus of the blind and strabismus.[8]

All newborns should screening for CC and have an eye examination performed by pediatrician/ophthalmologist including evaluation of red reflex. Retinoscopy is useful for early detection of axial lens opacities.[7]

After the detection of CC clinician should ask family history, newborns growth and in addition clinician should investigate the newborn for systemic and metabolic diseases that may be associated.[8]

Etiology and Genetics

Idiopathic congenital cataracts are the most common and their diagnosis based on exclusion. Unilateral CCs are generally idiopathic and they are not associated with any systemic or genetic disorders.[5] However, the majority of bilateral CCs result from several genetic and systemic conditions. As Rahi reported in a UK CC case review study, unilateral CC constitutes 56% of idiopathic CC but just 6% of hereditary CC.[9] As a general admission one-third of CC cases are hereditary, one-third are associated with risk factors and the remaining one-third are idiopathic. Related

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factors are chromosomal abnormalities, systemic and metabolic disorders and congenital infections.[10] Congenital cataracts also combined with ocular disorders including anterior segment dysgenesis, aniridia, micropthalmia, persistent fetal vasculature (PHPV) and other retinal disease. PHPV is most concomitant oculopathies which was consistent with results from previous studies, such as 2.0% reported by Toshiyuki et al. in Japan.[11] The second most common concomitant oculopathy was retinochoroidal coloboma.[12]

About 50% of bilateral CC cases have a genetic basis**.** Congenital cataract is both clinically and genetically heterogeneous; isolated congenital cataract is usually inherited as an autosomal dominant trait although autosomal recessive and X linked inheritance are seen less commonly.[13] Congenital cataracts also seen a part of a number of syndromes and chromozomal trisomies: trisomy 21 (Down syndrome), 13, 18 are is the most common trisomies and Cri du chat syndrome and Turner's syndrome can associated CC.[13]

Related systemic disease are metabolic disorders such as galactosemia, Wilson disease, hypocalcemia and hypoglisemia .Galactosemia develops a typical oil droplet cataract and easily seen by red reflex. CC regressed with diet in some metabolic diseases. CC may also be present as part of a broader spectrum of syndromic multisystem disease: Lowe syndrome, Alport syndrome, Hallerman- Streiff-Francois syndrome, myotonic dystrophy, incontinentia pigmenti and Stickler.[14]

Some intrauterine infections cause the development of congenital cataracts and this group termed as TORCH-associated and including CMV, rubella, toxoplasmosis, varicella and syphilis. These infections may be acquired in-utero or during delivery and may present clinically during the neonatal period. Rubella being the most common type of maternal infection related CC. Congenital rubella syndrome has a triad of cataract, sensorineural hearing loss, and patent ductus arteriosus. TORCH related cataracts usually bilateral central and dense.The cataracts caused by rubella may be present at birth or develop several months later[15].

Irradiation of the pregnant woman, medication during pregnancy are other maternal factors of caused CC.

Morphology

CC can be classified based on the place of opacity in the lens and morphology. CCs are very different morphological subtypes and differences from senile nuclear cataracts. Morphology of CC effects visual prognosis, like duration and reason of cataract.

Considering the lesion location, morphological types of congenital cataract can be classified in: nuclear, cortical, lamellar, anterior polar, posterior polar, pulverulent, total, etc. Long et al. reported a prevalence of 32.9% of total cataracts, 30.4% - nuclear cataracts, anterior polar - 8%, posterior polar - 13.5% and lamellar 9.6%.[16]

Lamellar Cataracts:

Lamellar congenital cataract is most common type of CC. Lamellar cataracts are characterized by opacification of the lamellae surrounding the fetal nucleus. Affected lenses have a clear center and discrete lamellar opacity. Their effect of visual prognosis varies with size and density of the opacity. This type usually carry a better prognosis than others. They are characteristically bilateral, symmetric, disc-like and static, which affects certain layers of the lens. These cataracts are often visually unsignificant and surgery in infancy is only necessary in some cases.[17]

Polar Cataracts:

Anterior polar cataract is an opacity at the anterior capsule of the lens. The majority of anterior polar cataracts are <3mm. Surgery is generally not necessary if the opacities need to be removed with surgery and will not harm a child's vision. Anterior polar cataract does not affect visual acuity to any significant extent but some studies suggest that surgery because of these cataract type can be associated with amblyopia and this strabismus due to anisometropia[18]. Follow up and refraction are important in unilateral cases.

Posterior polar cataract presents as a distinctive discoid lens opacity situated posteriorly, adjacent to the posterior capsule. Posterior polar cataract posterior capsule is extreme thin and fragile with adherence of the acellular opacity to the capsule. In approximately 20% of the cases, an association with a congenital defect in the posterior capsule has been reported. Posterior polar cataracts have a high incidence of posterior capsule rupture during the surgery[19].

Sutural Cataracts:

Sutural cataracts are located at the anterior and posterior Y-sutures of the embryonic nucleus. Occurring during the lens formative stage, sutural cataracts are static and present as white or blue band-shaped opacities. They are usually confined to the Y-suture and may be combined with cerulean cataract, coronary cataract, or other forms of cataracts[20].

Total Cataracts:

A total cataract represents a general opacity of all the lens fibers. Some lenses are completely opaque when first

diagnosed. In other cases, they develop from lamellar or nuclear cataracts. They are frequently bilateral and may progress. Cataracts involving the whole lens occur in Down syndrome, acute metabolic cataracts, congenital rubella and can also be seen in familial or sporadic cases as well as in some rare syndrome.[5-18-20]

Congenital Morgagnian Cataracts:

These are uncommon, total, dense cataracts, named after Giovanni Morgagni who described them in 1762; the outer zones of the lens become liquefied, while the nucleus remains intact.[21]

Treatment

When to operate?

Small cataracts can be observed, whereas dense cataracts that obstruct vision should be excised at an early age to prevent amblyopia. In infants, if the cataract is not removed during the critical period, the vision will never be fully regained after surgery. In unilateral cataract, clinical studies have revealed that surgery by 6-8 weeks has a better visual outcome as compared to later intervention. This may also be the critical period for bilateral disease.[28] Additionally optimal timing for surgery is difficult to establish due to the association of aphakic glaucoma with very early surgery. Compared to adults, decision for surgery is more difficult as subjective visual assessment in children cannot be obtained, and surgeons rely largely on the morphology and location of the cataract and behavior of the child.[22]

Ophthalmologists should preoperatively examine under operating microscope or slit lamp biomicroscope to assess the cataract and reveal potential challenges to surgery including; iris deformities, synechiae, zonulolysis, posterior lentiglobus, intumescent cataract, anterior and posterior capsule plaques.The extent and location of the cataract must be evaluated.[23]

CC surgery has many differences and challenges. The paediatric capsule is very elastic and requires the application of more force before tearing begins. When performing a CCC, control of the capsulectomy and prevention of extensions out towards the lens equator are inversely related to the force needed to generate the tear. A practical alternative to manual CCC is to create a small central opening in the anterior capsule using a vitrector probe. As another difference, posterior capsulotomy and anterior vitrectomy should be performed to prevent posterior capsule opacification (PCO) formation. Posterior capsulotomy can be carried out easily with the vitrector from an anterior incision or via the pars-plana. The posterior capsulotomy should be four to five mm in diameter and approximately one third of the anterior vitreous should be removed to ensure a permanently clear visual axis. The anterior vitreous forward after posterior capsulotomy.[24-25] New surgical improvements have contributed to a decrease in the incidence of postoperative complications such as posterior capsule opacification (PCO), postoperative inflammation and glaucoma.[24]

Intraocular lens (IOL) implantation has been advocated in children two years and above, due to problems arising due to IOL power, size, availability, material, refraction change and long term IOL safety. However, some studies have a succesful outcomes, they implant IOL in younger age groups like one year.[26]

The indication and selection of IOL continues to be a controversial subject. The Infant Aphakia Treatment Study (IATS) compared the treatment of unilateral cataract in infants aged 1-6 months with primary IOL implantation vs aphakia with contact lens (CL) correction. Recent reports showed no significant difference between the median BCVA of operated eyes in children who underwent primary IOL implantation and those left aphakic. However, higher rates of complications, adverse events and additional intraocular surgeries were noted in the primary IOL group.[27]

Postoperative complications in congenital cataract surgery include visual axis opacification, secondary glaucoma, fibrinoid reactions, decentralization of the pupil, retinal detachment and endophthalmitis.[24-25]

Table 1: *Morphologic types of Congenital Cataracts.*

WHOLE LENS	CENTRAL	ANTERIOR	POSTERIOR	MISCELLANEOUS
Total	Lamellar	Anterior polar	Mittendorf dots	Punctate lens opacities
Congenital Morgagnian	Central pulverulent	Dot like	Posterior cortical	Sutural
Membranous	Ant egg	Plaque like	Posterior	Coralliform
	Nuclear	Anterior pyramidal	subcapsular	Wedge shaped
	Oil droplet	Anterior subcapsular	Posterior lenticonus	Persistent hyperplastic
	Cortical	Anterior lenticonus		primary vitreous
	Coronary			

Complications of cataract

1- Amblyopia

Obstruction of vision by media opacities may cause serious damage to the immature visual system. Total cataracts, dense axial (nuclear or polar) opacities more than about 3 mm in diameter, and other comparable disturbances of media clarity can produce bilateral or unilateral amblyopia on the basis of form vision deprivation.[28]

Deprivation, refractive and strabismic amblyopia are the major causes of visual defect in cataractous or aphakic children. Patients with monocular cataracts also have amblyopia of binocular interaction secondary to their anisometropia and aniseikonia.[29] The age of the patient, the length of time the cataract is present, and the interval until successful optical rehabilitation of the eye is achieved determine the depth of amblyopia and influence its response to occlusion therapy.

Causes of amblyopia in paediatric cataractous patients:
1) Pre- operative
A- Deprivation amblyopia caused by the lens opacity.
B- Strabismus (commonly associated with infantile cataracts and frequently requires corrective surgery)
2) Post-operative
A- Absence or non-compliance of aphakic collection causes anisometropic amblyopia.
B- Amblyopia induced by aphakic correction modalities

2- Strabismus

Strabismus is common in CC and be present before cataract surgery or afterwards. Robb and Peterson reported strabismus in 68% of children with bilateral congenital cataracts.[30] In studies, strabismus has been reported more frequently in unilateral cataracts than in bilateral cases.[31] Exotropia was more common than esotropia.

Parks and Hiles reported that there was a strong correlation between cataract type and the risk of strabismus; however, there was not a significant association observed between cataract density and strabismus in some other studies.[32])

3- Glaucoma

Apart from visual loss, the main indication for cataract extraction, the major complication of cataract formation is some form of glaucoma.

After surgery most frequent long-term complication of aphakic eyes in connection with posterior capsulorhexis and anterior vitrectomy is aphakic glaucoma, which in individual cases can lead to substantial impairment of vision. In individual treatment planning it has to be considered that although younger age at the time of cataract removal can provide better prerequisites for prophylaxis of amblyopia, it also confers a higher risk of development of aphakic glaucoma.[31]

Many factors have been reported to increase the risk of postoperative glaucoma, including microphthalmos, fetal nuclear cataract, conspicuous family history and associated ocular malformations, such as PFV.

CONCLUSIONS

Treatment and management of congenital cataract remains a challenge. It is resource demanding, difficult and requires a dedicated joint effort by parents and the medical profession.The optimal time for surgical intervention in congenital cataract is still a matter of controversy. Improved treatment strategies and surgical methods diminished the long term complications.

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