International Clinical and Medical Case Reports Journal
Review Article (ISSN: 2832-5788)

The Worldwide Burden of Paediatric Cataract: A Review

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Citation: *Ashraf S, Bhasin P, Dhage A, Parveen S. The Worldwide Burden of Paediatric Cataract: A Review. Int Clinc Med Case Rep Jour. 2022;1(2):1-7.*

Received Date: 24 June, 2022; **Accepted Date:** 04 July, 2022; **Published Date:** 06 July, 2022

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ABSTRACT

Introduction: This article reviews the causes of childhood blindness, the legal interpretation of blindness and highlights the role of surgery in the management of these children.

Background: The term blindness is a general term that can include those with legal blindness and reduced vision. The global initiative by the World Health Organization (WHO) for elimination of avoidable and preventable blindness by year 2020 has given highest priority to the control of childhood blindness. Globally, around 285 million people are visually impaired, out of which the number of blind people is an estimated 39 million.

Blindness prevalence among children in affluent regions is about 0.3/1000 children, and in the poor communities it is approximately 1.5/1000.

Methods: E literature on Paediatric Cataract was retrieved from PubMed, Google Scholar and Cochrane database. Keywords and phrases used during the search included “aphakia,” “paediatric,” “cataract” and “blindness”.

Conclusion: A vast number of factors are needed to be considered when deciding on the treatment of choice for paediatric cataract. The decision whether to implant an IOL or leaving the child aphakic, needs compliance and planning and compliance along with family support. Adequate rehabilitation of neuro-ophthalmic axis is extremely important for better long-term visual outcomes.

Keywords: Paediatrics; Blindness; Surgery; Children

INTRODUCTION

The term blindness is a general term that can include those with legal blindness and reduced vision. The global initiative by the World Health Organization (WHO) for elimination of avoidable and preventable blindness by year 2020[1] has given highest priority to the control of childhood blindness. At present, cataract is one of the most important causes of treatable blindness, especially in children.[2] Cataracts if not treated in children can lead to immense economical, emotional and social burden to the society, family and the child himself. With proper identification and treatment, paediatric cataract can be easily managed. Most of the cases are diagnosed on routine screening and some are diagnosed after parents notice strabismus or leukocoria in the child.

Visual impairment is defined on the basis of function, instead of visual field cutoff values or visual acuity. This article reviews the causes of childhood blindness, the legal interpretation of blindness and highlights the role of surgery in the management of these children.[3]

PREVALENCE

Globally, around 285 million people are visually impaired, out of which the number of blind people is an estimated 39 million.[2]

Blindness prevalence among children in affluent regions is about 0.3/1000 children, and in the poor communities it is approximately 1.5/1000. Globally, there are estimated 1.4 million blind children, 3/4th of which reside in developing countries. India, being one of the largest south asian countries with a population of around 1.03 billion as in 2001, it has been found that roughly 420 million were kids under 16 yrs of age (40.8%).[4]

Almost 70 million blind- person-years are due to childhood blindness, out of which around 10 million blind-person-years (14%) are caused by paediatric cataract. India has a huge burden of 280,000–320,000 visually impaired children, giving rise to an estimated loss of US $3,500 million.[5]

ETIOPATHOGENESIS

Paediatric cataracts are often categorised as:

- Congenital cataracts – cataracts present since birth
- Developmental, juvenile or infantile cataracts – cataracts identified in adolescents, older children or babies.[6]

The causes and types of cataracts in children can vary- it can be unilateral or bilateral; can appear in various parts of lens- ranging from tiny dots to dense clouds. It can also be caused due to trauma, metabolic disorders- like diabetes or galactossemia. Most of the times, the exact cause of cataracts in children is unknown. In fact, it is rare to find cataracts in children and babies. In the United Kingdom, about 3 to 4 in every 10,000 babies are born with cataracts.[7]

Congenital paediatric cataracts can often occur as part of numerous birth defects like, Congenital rubella syndrome, Chondrodysplasia syndrome, Down syndrome (trisomy 21), Lowe syndrome, Pierre-Robin syndrome and Trisomy 13.

HISTORY AND PHYSICAL EXAMINATION

It is essential to collect a thorough case history followed by physical examination. The history should comprise of :[6]

- Child's ocular and visual history
- Family ocular and visual history
- Child's history of any medications
- Medical history of the family
- Social history
- Vocational, hobby and educational history

Following history, a visual and ocular examination is imperative to assess the current status of vision and the functioning of the eyes.[6]

- Near and distant visual acuity
- Refraction
- Motility and size of the pupils, binocular vision
- Visual field
- Testing of glare, color vision, contrast sensitivity
- Ocular health examination with a dilated fundus examination

EVALUATION

Examination, diagnosis and treatment of paediatric cataract are the very important first steps of a multi-faceted journey towards visual rehabilitation. A close collaboration between the patients, the parents and the ophthalmic team is extremely crucial for achieving the best visual prognosis.

It is vital to conduct a thorough evaluation, along with taking a good history which should include signs of failure to thriving and any delayed milestones.[8] Eye exam comprises of measurement of anterior segment, intraocular pressure, as well as the posterior segment evaluation. Supplemental testing like formal visual field examination, tests of colour vision, contrast sensitivity testing, visual evoked potentials, electrooculography, electroretinography, optical coherence tomography, enetic testing and fluorescein angiography may be important to determine the etiology of the cataract, monitor the vision and appropriately manage the condition.[8]

Cataracts present in each child in a different way, depending upon the timing of presentation, the amount of visual loss, and the density of cataract. Infants may be referred to the ophthalmologist after incidental finding of any abnormalities at the newborn baby check for example, a recent onset squint, but most often it is due to an absent red reflex.[9]

Older children and adolescents may present after they are unable to pass school visual screening test, or after parents raise concern saying that the child bumps into things while walking. Children with systemic disease are referred to the ophthalmology department from the paediatrician.

It is necessary to consider the early inputs from paediatrician to facilitate the general assessment of the child for potential systemic disease.[10] Hence, it is vital to ask about the mother's gestational and pre-natal health to rule out intrauterine TORCH infections and drug use.[10]

Prior to eye examination, a general inspection of the child's/ adolescents face must be performed, to look for any dysmorphia in the face. Various syndromes associated with characteristic facial features with congenital cataracts are listed in Table 1.[5]

Table 1: Congenital syndromes associated with characteristic facial features with congenital cataracts.[5]

Associated syndromes	Notable facial / ocular features
Lowe Syndrome	Sunken eyes
	Chubby cheeks
	Frontal prominence
Down Syndrome	Upward slanting palpebral fissures
	Flattened occiput & midface
	Small ears
	Epicanthic folds
Edward Syndrome	Low-set ears
	Webbed neck
	Mcirognathia
Cri-Du-Chat Syndrome	Down-sloping palpebral fissures
	Low-set ears
	Microcephaly
TORCH Infections	Microphthalmos
	Microcephaly

MANAGEMENT AND CHALLENGES

A number of important factors are needed to be considered in management of infant, newborn and adolescent cataracts. These involve age at presentation, laterality and type of cataract, changes of eye during the critical developmental period, and the risk of any complications or surgery or interventions which might be invasive in nature. Therefore, early diagnosis is the key in helping the ophthalmology team to develop an individualised management plan and to limit further amblyopia, nystagmus or visual loss. Also it is extremely important to involve the parents and/or guardian in the decision making process, as further visual rehabilitation, management and postoperative care relies heavily and solely on the cooperation of the family.[11]

CONSERVATIVE

There's a proportion of childhood cataracts that does not require immediate urgent intervention and hence can be effectively observed periodically to take note of any discrepancy of the visual axis. It needs regular objective assessment with particular emphasis on the present morphology. These types of cataracts are the ones which are less than 3mm in diameter, in total anterior or peripheral opacity. Small punctate zones in anterior cataracts can also be managed conservatively.[12]

On the other hand, posterior + central cataracts and confluent cataracts larger than 4mm are best treated surgically because of their high likelihood for visual disruption and the child later presenting with more than one visual complication at diagnosis. Although lamellar cataracts are a more central type of cataract, they tend to be formed partially at birth and they develop gradually, that allows for adequate and early visual development. Hence, these can be managed initially by observation.[13] In borderline cases, pupillary dilatation is useful to delay surgery by instilling 2.5% phenylephrine hydrochloride to the cataractous eye and part-time occlusion of the better eye. Cycloplegic agent like atropine when used daily should, however, be deferred due to the likelihood of amblyopia. Such patients should be closely monitored and if amblyopia develops, surgical treatment will be required.[14]

Children having amblyopia but visually non-significant cataracts can be managed successfully with eye-patching and glasses. The treatment of amblyopia is very important post-operatively and the patching results depend on compliance, age and the visual acuity level before patching.[15]

SURGICAL

Surgical management is necessary in patients with significantly large cataracts that affect the visual axis, and timing is extremely crucial to preserve the visual acuity during the essential period of eye development that ranges from 8 weeks to 6 months of age. As a rule of thumb, congenital unilateral cataracts must be operated as early as possible, prior to 6 weeks of age, and bilateral cataracts prior to 8 weeks of age.[14] If the surgery is delayed beyond 2-3 months of age, studies have shown that it potentially increases the risk of nystagmus and strabismus. Naturally since

birth, the neonatal eye is hypermetropic and the globe is much shorter that of an adult, and it has a more plastic cornea and sclera. It means that any eye surgery in this age group has a greater chance of causing an inflammatory response, which can also be due to an immature blood-to-aqueous barrier.[15]

The drawbacks and benefits of IOL implantation versus aphakia should also be discussed with the family. In an aphakic child, the use of glasses or contact lenses is necessary to achieve desired visual acuity, as well as it accounts for accommodative loss. Non-compliance with the glasses may cause significant amblyopia. Long-term study outcomes have depicted that in unilateral cataract, the visual acuity at 1,4 and 10 years is almost the same irrespective of treatment with aphakia or primary IOL or with contact lenses.[16]

It is essential to conduct preoperative biometry to obtain ocular measurements and to evaluate strength of IOL as well as potential myopic shift. This is performed under (GA) General Anaesthesia before planning surgery. Under correction of the eyes is usually accepted, which renders the child moderately hypermetropic, with the use of contact lenses or glasses or contact lenses in the interim period so as to avoid undesired myopia in adulthood. However, some doctors might aim for an emmetropic target as they feel that it can enhance visual development during childhood while also accepting the need for a myopic correction in the future.[17]

Primary Intra-Ocular Lens implantation is mostly reserved for visually significant paediatric cataract in adolescents and older children,[16] whereas in younger children and infants, it is often not recommended due to its many associated complications. Developmental cataracts fare far better in prognosis as compared to congenital cataracts, since they are less likely to have visual disruption during the critical early period.[15]

SUMMARY

Around 70 million blind- person-years are due to childhood blindness, out of which approximately 10 million blind-person-years (14%) are because of paediatric cataract. This means that a huge chunk of population is affected by this ailment, and it has even worse prognosis if it occurs in childhood, as development of a child is hampered.

A vast number of factors are needed to be considered when deciding on the treatment of choice for paediatric cataract- whether it is unilateral or bilateral; whether it is congenital or developmental. Whether the cataract is hampering the child's visual development? Surgery can be beneficial if initiated early, but it is associated with the risk of secondary postoperative complications.

The decision whether to implant an IOL or leaving the child aphakic, needs compliance and planning and compliance along with family support. Adequate rehabilitation of neuro-ophthalmic axis is extremely important for better long-term visual outcomes.

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