LIGHT UP

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LVPEI

World Health Organization
Collaborating Centre for
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Meera & L.B. Deshpande Centre for Sight Enhancement

Dr. P.R.K. Prasad Centre for Rehabilitation of Blind and Visually Impaired

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Cataracts are a major cause of vision problems in young children, throwing up challenges in terms of surgery and medical management, and accommodation of the resulting low vision. This issue of* Light Up *explores congenital or pediatric cataract from three angles-from the viewpoint of surgical management, low vision care, and the underlying genetics of the problem.

Management of Pediatric Cataract

Introduction

The majority of congenital cataracts are of hereditary or idiopathic origin. The past few years have seen a dramatic evolution in the management of pediatric cataract. Intraocular lens implantation, which was considered radical and dangerous less than a decade ago, is now accepted by many surgeons as the treatment of choice for congenital and traumatic cataracts in children 2 years of age or older.

Evaluation

Most children with cataracts are presented to the clinician with a white pupillary reflex or leukokoria. Occasionally, strabismus, nystagmus and /or poor visual function may be a presenting feature. Increasing awareness among pediatricians and regular vision screenings at school have contributed to early diagnosis and referral of childhood cataracts.

Assessing the nature of fixation is the simplest and the most practical test for visual assessment in children less than one year of age. Preferential looking or visual evoked potential recordings also provide an accurate assessment of infant visual acuity. Older children may be tested reliably with Snellen optotype equivalents. Ocular examination should include measurement of corneal diameter (horizontal and vertical), record of intraocular pressure (with a Perkin's tonometer), extent of lens opacity, and status of posterior segment (with an indirect ophthalmoscope or B-scan ultrasonography), and measurements for IOL power calculation, including axial length and keratometry.

Surgical Decision

Unilateral and dense bilateral cataracts can be taken up for surgery within the first few weeks after birth. In bilateral, symmetrically dense cataracts, the two eyes should be operated within a short interval of 1 - 2 weeks.

Aphakic Correction

A variety of options are available for aphakic correction, including spectacles, epikeratophakia, contact lenses and IOLs.

Spectacles

Spectacle correction continues to be an important modality for treating bilateral aphakia. Spectacles are affordable and allow frequent correction of changing refractive error. However, spectacles are not

suitable for monocular aphakic correction, are cosmetically not pleasing, uncomfortable for wear, and prone to scratches or damage. Moreover, they provide an image of suboptimal quality.

Contact Lenses

Aphakic correction is perhaps the most important indication for fitting contact lenses in children. Contact lenses are the most logical choice for correcting monocular infantile aphakia. However, high cost, limited availability and the need for frequent replacement are the major limiting factors. Contact lens fitting should be performed as early in the postoperative period as possible. An addition of +1.00 to +3.00D to the emmetropic correction is necessary for the infant to appreciate its immediate surroundings.

Intraocular Lenses

Intraocular lenses (IOLs) have established their safety and efficacy in correcting aphakia in the adult population. Now, improved surgical techniques and availability of superior IOL designs and materials facilitate safe implantation of IOL's in children.

Murali K Aasuri, MD
Consultant -Cornea & Anterior Segment Services
L V Prasad Eye Institute

Low Vision Care in Management of Childhood Cataract

Of the estimated 1.5 million blind children worldwide 1 million live in Asia. Blind school and population based surveys performed in south India have shown that childhood cataract is a significant cause of blindness and severe visual impairment, accounting for up to 12-15% of all childhood blindness

Despite using the various modalities described above for aphakic correction and management of amblyopia, a large number of children continue to have low vision. Poor management of low vision in children with congenital cataract adds to social, economic and psychological problems for both the family and the community. The cumulative loss from childhood blindness in India for 0.25 million childhood blind and for an estimated 33 working years of life is Rupees 801 billion (US $ 22.2 billion) which is 28.7% of the cumulative GNP loss due to all blindness.

Management approaches for low vision due to congenital cataract include refraction (Bifocals), prescription of Low Vision Devices (LVDs) and low vision training, assessment of vision in young children, treatment of amblyopia and visual stimulation. A telescope could be prescribed for board work and high plus spectacle devices for reading tasks. Unlike other low vision children these children require an addition for near vision tasks, as they do not have the ability to bring the book close to their eyes due to loss of accommodation. These children also need good contrast, proper lightning and counseling for educational and vocational needs. Comprehensive and timely low vision care of children with paediatric cataract and associated decreased vision could minimize long-term permanent visual disability and reduce or prevent the economic loss from blind years.

Sarfaraz A Khan, MD
Director - Vision Rehabilitation Centres
L V Prasad Eye Institute

Genetics of Congenital Cataracts

Congenital cataracts can arise due to hereditary or non-hereditary causes. Some common non-hereditary causes include prenatal infections such as rubella, toxoplasma, cytomegalovirus, herpes simplex and herpes zoster. Cataracts can be also associated with developmental abnormalities such as prematurity with low birth weight, birth anoxia, central nervous system involvement leading to seizures, and cerebral palsy.

Congenital cataracts that are hereditary, i.e., which arise due to genetic factors, usually show Mendelian inheritance. This means that the disease is caused by a single genetic alteration. Examination of the family history of patients with hereditary cataracts reveals either autosomal dominant, autosomal recessive or X-linked modes of inheritance. If one looks at the pattern of disease within a family, the dominant mode of inheritance is characterised by the presence of disease in every generation. In the recessive form, the parents or other individuals in earlier generations may not be affected, but more than one offspring in the same generation may be affected. We do not have accurate data about the prevalence of hereditary cataracts in general or of any particular subtype in India. In south India, the autosomal recessive form may be more common than elsewhere due to the presence of consanguinity. Recessive disease is often (though not always) associated with consanguinity. X-linked types of cataracts are not very common and should be suspected if disease segregates only among male members even though it is transmitted through females. Thus a careful evaluation of family history including examination, if required, of parents and siblings of patients with congenital cataracts can reveal a hereditary

cause. Although the presence of a family history does point to a genetic origin of disease, sporadic or isolated cases of congenital cataract may also be genetic. Recessive cataracts can present as isolated cases in which case, the presence of consanguinity in the parents can further point to a recessive inheritance. However, possible non-genetic causes such as those mentioned above need to be evaluated before making a conclusion.

Hereditary cataracts may constitute up to about 20% of cataracts [1] in children under 1 yr of age. Although estimates of the prevalence of hereditary cataracts are made on the basis of a family history, there are a large number of sporadic cases of undetermined etiology some of which may also be genetic. Hereditary cataracts can occur in isolation or in association with other ocular or systemic abnormalities. Ocular disorders associated with cataracts include anterior segment dysgenesis, microcornea, microphthalmia, and retinitis pigmentosa. Systemic diseases due to inborn errors of metabolism can be associated with cataracts. Examples are galactosemia and congenital lactose intolerance, where early diagnosis and appropriate dietary restrictions can reverse the formation of cataracts. These can be recognised by means of blood and urine tests and by the presence of other signs and symptoms.

The underlying genetic causes of hereditary cataracts have been defined for the various dominant cataracts as well as some recessive cataracts. The genes causing congenital cataracts are several and differ from one family to another. These genes encode proteins that maintain lens structure, homeostasis and regulate its development. The phenotypes (manifestations) of hereditary cataract both in terms of the severity and the morphology, also vary a great deal even among affected members of the same family. These factors should be considered when determining the possible mode of inheritance, since there may be mildly affected individuals whose phenotype may be identifiable only upon ophthalmic examination. Knowledge of the inheritance pattern and the presence of other associated abnormalities is important for counseling of patients.

Dr. Chitra Kannabiran
Scientist - L V Prasad Eye Institute

ref. [1]. Eckstein M., Vijayalakshmi P., Killedar M., Gilbert C., Foster A. Aetiology of childhood cataract in south India. BJO 1996; 80: 628-632.

Case Study

Mast. Ajit Kumar

Master Ajit Kumar had been operated for bilateral congenital cataract in LVPEI at the age of 2 months followed by prescription of glasses for his aphakia. A functional vision assessment done at the age of three at the children's rehabilitation centre showed that he had good residual vision and was able to identify pictures, pegs, beads follow and locate the moving ball at 10-15 meters; and move around independently in familiar and unfamiliar areas in good illumination. Parental counseling and educational guidance was provided. The child was admitted in a normal preparatory school. At the Center for Sight Enhancement the child had an appropriate and timely follow-up program to check his glasses and the importance of using glasses was explained to the parents. A letter was given to his teacher requested that the child be provided a in the front row during board work. Thanks to the early surgical interventions followed by optical correction of aphakia, counseling and environmental modification allowing him to gain access to the board work and book print, hence improving the quality of education and employment opportunities.

Education

Optometrists Santosh Kumar Tripati from Kanpur and Sadhucharan Barik from Cuttack has completed the Short term fellowship program in low vision care from 1st May - 31st July, 2002.

NEWS

Dr. S A Khan

"GOLDEN EYE 2002" Golden Jubilee Conference of Tamilnadu Ophthalmic Association, Tiruchirappalli, Tamilnadu, August 2-4, 2002. Delivered a guest lecture on "Current concepts in the management of Low Vision and conducted a course on Low Vision Care."

Attended the **7th International Conference on Low Vision, Goteborg, Sweden,** July 18-25, 2002. Conducted pre-congress workshop entitled "Low Vision Service model for developing world".

Presented free papers on "Low vision service in developing countries ", and: "Management of patients with Age Related Macular Degeneration at a Tertiary Eye Care Centre in South India".

Invited faculty member at the **Lighthouse International workshop** on "Low Vision Centres of Excellence: A Comprehensive Approach"

IAPB Low Vision Working Group Meeting, Goteborg, Sweden, July 20, 2002. Attended as a regional representative for South East Asia

Ms. Beula Christy

Atttended the **7th International Conference on Low Vision, Gotenburg, Sweden** from 21 to 25, July 2002. Made several poster presentations on:

1. Creating Barrier free work environment for visually impaired persons.
2. Addressing phychological issues of Elderly Visually impaired persons
3. Perceptions and attitudes towards blindness in Urban South India - A population based study.
4. Way finding techniques for persons with low vision.

Also made oral presentations on "Integration of rehabilitation services in clinical low vision", "Early intervention for visually impaired children with additional disabilities", "Study on self-perception of visually impaired children".

Also attended the **ICEVI World Conference, Netherlands;** 27 July to 2 August 2002. On the impact of technology in meeting the educational needs of visually impaired children.

Award

The 2002 Constance W. Atwell award for Excellence in Low Vision Research was presented to Vijaya K Gothwal on 9th May 2002 during Association for Research in Vision & Ophthalmology at Fort Lauderdale, FL, USA.

Low Vision Awareness Program

September 27 to 29, 2002
March 14 to 16, 2003
September 26 to 28, 2003

Short term fellowship program in Low Vision Care

Three month program for ophthalmologists and optometrists

For more details contact to:

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