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Overview

Childhood Cataracts: Aetiology and Management

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Courtesy - Dr. K K Srivastava

Childhood Cataracts

Childhood cataracts are responsible for 5% to 20% of blindness in children worldwide and for an even higher percentage of childhood visual impairment in developing countries.¹ The prevalence of childhood cataract varies from 1.2 to 6.0 cases per 10,000 infants.² Cataracts in children not only blur the retinal image but also disrupt the development of the immature visual pathways in the central nervous system. Hence timely removal of cataract followed by prompt visual rehabilitation is of utmost importance in children.

Aetiology

Approximately half of the bilateral cataracts and majority of the unilateral cataracts in children are idiopathic in nature. Bilateral infantile cataracts are more common with systemic diseases and more likely to be inherited, whereas

unilateral cataracts are commonly associated with other ocular abnormalities. Important causes of childhood cataracts include: genetic disorders, intrauterine infection, metabolic disorders, drug induced, trauma and other ocular disorders like aniridia, microphthalmia, persistent hypertrophic primary vitreous, and anterior segment cleavage syndrome. In developed countries, hereditary cataracts are the most common type of congenital cataract.³ In some developing countries, approximately 25% of infantile cataracts are due to congenital rubella infection.⁴

Work-up

A careful history should be taken focusing on prenatal and postnatal events that might suggest aetiology for cataract. A family history is helpful in determining whether cataract may be hereditary. A complete ocular examination including slit-lamp-biomicroscopy and indirect ophthalmoscopy should be performed. Parents and siblings should also be examined to determine if they have lens opacities. In bilateral cataracts if a hereditary basis cannot be established, laboratory investigations for fasting blood sugar, plasma calcium and phosphate, TORCH titre and urine reducing substances after milk feed should be performed. These tests should be tailored for each patient. A paediatric consultation is also required as the child may require medical intervention to treat systemic conditions that may pose an additional risk during general anaesthesia.

Management of Childhood Cataract

Cataract extraction is now the preferred treatment for visually significant cataracts. Mydriatics, which may improve vision in central cataracts, are rarely practised.

Indications for cataract surgery

Since a subjective assessment of visual acuity cannot be obtained in very young children, greater reliance must be placed on morphology of cataract, other associated ocular findings and the visual behaviour of the child, in order to ascertain whether or not the cataract is visually significant. The degree of visual impairment induced by lens opacity differs markedly depending on the location of the opacity. Generally the more central and posterior the opacity, the more significant the cataract. Dense central opacity larger than 3 mm in diameter usually warrants surgical removal.² In partial cataracts surgery is indicated when the visual acuity is less than 6/18 or in preverbal children when fixation is poor. If the cataract is not visually significant, observation alone may be sufficient. However, a careful follow up



Courtesy - Dr. K K Srivastava

is mandatory in these children to ensure that cataract does not progress and to monitor refractive error and amblyopia.

Timing of surgery

A visually significant cataract should be removed as soon as possible. Prompt treatment of total cataracts during the first 6 weeks of life usually results in good visual outcome and prevents the development of nystagmus.⁵ In bilateral cataracts the second eye should be operated within a short time. Unless the risk of a second anaesthesia is too high, simultaneous cataract surgery in both eyes should be avoided.

Type of surgery

- A. Children < 2 years: Extracapsular cataract extraction (ECCE) with primary posterior capsulotomy (PPC) and anterior vitrectomy (AV). IOL implantation should be considered in unilateral cases where contact lens is not feasible.
- B. Children between 2-5 years: ECCE with PPC with anterior vitrectomy and IOL implantation. We do not prefer optic capture through posterior capsulorhexis because unless this is combined with anterior vitrectomy, it does not prevent secondary membrane formation.⁶
- C. Children > 5 years: ECCE with IOL lens implantation. A PPC should be considered in children who are not expected to be candidates for ND: YAG capsulotomy, for instance, children with nystagmus or with mental retardation.

Postoperative care

Topical steroid (Prednisolone acetate 1% drops), antibiotic (Tobramycin 0.3% eye drops) and cycloplegic (Cyclopentolate 1%) are given in tapering doses over 6 weeks. A short course of systemic corticosteroid (Prednisolone 1mg / kg body weight / day) for 10-15 days is given to patients with a fibrinous reaction. Children should be examined daily till discharge, at one-month post-op and every

3 to 6 months thereafter. Visual acuity estimation, refraction, intraocular pressure measurement and fundus examination should be done at each visit. Special attention should be paid to clarity of visual axis and development of posterior capsular opacification.

Postoperative visual rehabilitation

The real challenge starts after the cataract surgery as the child is left with a gross refractive error, which can lead to development of amblyopia if not corrected in time. Achieving a good visual outcome following cataract surgery in children remains difficult, requiring extra effort and patience on part of the ophthalmologist and good compliance from the parents.

Optical rehabilitation

Aphakic glasses are the safest mode of visual rehabilitation for bilateral aphakia and their power can be readily changed to compensate for ocular growth. They are not suitable for unilateral aphakia because of high degree of aniseikonia they induce. An overcorrection of 2-3 D for children below 3 years and bifocals with 3D add for older children are prescribed for clear near vision. Plastic glasses with plastic frames are the best as they are lightweight, have a high impact resistance and do not break on trauma. However, they offer poor cosmesis and centration, get scratched easily, and can lead to peripheral aberrations and poor compliance.

Contact lenses are suitable for bilateral as well as unilateral aphakia. They have certain advantages over spectacles such as less aniseikonia, no aberration and better cosmesis. The power can be adjusted to compensate for ocular growth. Of the three main types of contact lenses available for the paediatric age group (hydrophilic, silicon and rigid gas permeable lenses), silicon lenses are the most preferred. An overcorrection of 2-3 D for young children and bifocals for children over 3 years of age is prescribed. The potential disadvantages are poor compliance, high cost, difficulties in maintenance problems arising from poor hygiene and frequent lens loss.

Primary IOL implantation has now become the standard optical treatment in children (other than infants below three months) following cataract surgery, except in cases of uveitis, microphthalmos, chronic glaucoma. We implant an IOL in children older than 3 months with a unilateral cataract and in children older than 2 years with bilateral cataracts. IOLs provide excellent optical correction, independent of patient / parent compliance. The choice of lens power poses a challenge and commonly the eye is undercorrected based on the child's age. Secondary IOL implantation can be done successfully in most of the cases following cataract surgery. Anterior chamber IOL implantation is generally not recommended in children.

Amblyopia therapy

Amblyopia therapy is the most important and also most neglected aspect of visual rehabilitation following cataract surgery in children. This should start as early as possible following cataract surgery. The schedule of occlusion is based on the age of the child.

Outcome

The visual outcome following cataract surgery depends upon the age of onset, type of cataract, laterality, method of optical rehabilitation, amblyopia therapy, associated ocular disorder and post-operative complications.

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Dealing with Paediatric Cataract at Drashti Netralaya - Our Experience

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There are few reports of prevalence and causes for blindness among children on a global or regional basis. A study from Andhra Pradesh¹ suggested that congenital cataracts account for up to 11% of blindness among children. Appropriate and timely treatment can help restore sight in cases of congenital cataract. Surgical techniques have evolved over the years with intraocular lens (IOL) implants now the treatment of choice for congenital and traumatic cataract in children over the age of 2.² However, there are conflicting opinions of whether intraocular lens implants are safe for children below two years. Determining the target postoperative refraction³ and the complexity of surgical procedures⁴ are additional concerns related to implanting intraocular lenses in small children. The impact of inflammation, amblyopia and posterior capsule opacification on postoperative results also has to be kept in mind. We present the results of a retrospective analysis evaluating the outcome and factors affecting outcome of cataract surgery done in the paediatric age group at our hospital during 2003.

We reviewed charts of all cases of surgery performed for cataract due to any cause among children below the age of 16 during the period January to December 2003. Evaluation included visual assessment, and anterior and posterior segment examinations. Younger children (aged below 4 years) were operated under



Courtesy - Dr. Mehul Shah

general anaesthesia, and older children were operated under peri-bulbar block with sedation if the child cooperated.

Surgical technique

Wound construction was done using self-sealing suture less wound in most cases.⁵ Capsular management was obtained in most cases through a central capsulorhexis measuring around 4 mm. Lensectomy and Anterior vitrectomy were done in children under 2 years and in traumatic cases where posterior capsule was suspected to be ruptured, a limbal approach was taken for lensectomy and anterior vitrectomy.⁶ A primary posterior capsulotomy with or without vitrectomy was performed. A PMMA lens (12 mm) was implanted in the bag for children below 2 years.⁷ Intraocular lenses were not implanted for children aged less than 2 years. Children who did not receive IOL implants were rehabilitated postoperatively using spectacles or contact lenses. Patching was done if the cataract was unilateral and amblyopia was present.



Courtesy - Dr. Mehul Shah

Results

A total of 41 (males 22, females 19) paediatric cataract cases were seen and managed in the year 2003. Twenty of the 41 cases were developmental or congenital cataract, 20 had cataracts due to trauma, and 1 case was diagnosed as complicated cataract. 30 cases underwent Extra Capsular Cataract Extraction (ECCE) with IOL implants and 11 cases had lensectomy and vitrectomy procedures performed. 21 children had bilateral cataracts. Major causes of trauma included wooden stick (n=9), firecrackers (n=3), and thorns (n=3). Six children were injured while engaged in subsistence labour, while 14 were injured during play.

Thirty-four children had vision less than or equal to 3/60 pre-operatively (vision could not be assessed for 7 children) in the affected eye. Postoperatively, vision improved to 6/18 or better in the operated eye for 21 children. Only 3 still had poor vision of 3/60 or less while 9 had moderate vision impairment and would benefit from further optical correction. Vision could not be assessed for 8 children postoperatively. Half were lost to follow-up after a month of surgery while another 30% were followed up for more than 2 months.

Recommendations

- Concerted efforts must be made to educate people about the prevention of blindness due to cataract in all age groups in general and the importance of continued follow up in the paediatric age group in particular.
- Apart from school screening programmes, training programmes must be conducted to equip teachers to identify vision problems and to sensitise them about the significance of early reporting.
- The facilities at primary health care institutes need to be upgraded and the staff trained to provide postoperative follow up care to paediatric patients.

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VISION 2020 - The Right to Sight-India Forum

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The VISION 2020 - The Right to Sight: India forum was registered as a not-for-profit society under "The Societies Act of India" on 26 May 2004 (Regn. No: 48). It is a national confederate body that aims to strengthen the implementation of VISION 2020 activities in alignment with national objectives and targets and thus contributes to the global elimination of avoidable blindness. It is poised to develop into a "National Entity for Transformation, Human Resource Development, Research, and Advocacy" - NETHRA (or "eye" in Sanskrit) for eye care in India.

The national secretariat is located at Lions Aravind Institute of Community Ophthalmology of the Aravind Eye Care System in Madurai. Aravind, one of the founder members of VISION 2020 has offered to provide initial support and guidance to the national secretariat of VISION 2020 India with systems, logistics, and access to its excellent professional systems and its extensive network.

Mr. P. Kamala Manohar Swamy joined as Executive Director on 18 May 2004. He has been working as a development professional in the non-governmental sector for the past 25 years and has also received a national award for his work in

rural development. He has several years of experience in the development sector in institution building, project management, capacity building, networking and resource mobilisation from government and non-governmental resources.

VISION 2020 India has started its own institutional development initiatives and is developing a series of Thrust Area Programs (TAP). ORBIS International, one of founder members, has already given a grant to support VISION 2020 India to initiate activities such as state launches, sensitisation workshops, operational research, resource material development, publication of the Indian supplement to the Journal of Community Eye Health and capacity building programmes for stakeholders across the country.

The other founder members are CBM, Sight Savers, Operation Eyesight Universal, Seva Foundation, Lions Clubs International Foundation, Dr. R. P. Centre for Ophthalmic Sciences (AIIMS, New Delhi), LV Prasad Eye Institute (Hyderabad) and the Aravind Eye Care System. These organisations have joined ORBIS to extend their partnership support to VISION 2020 India to pursue the global objectives of VISION 2020. They currently constitute the management board of this new organisation.

The management board has met for the first time since the constitution of this organisation on July 9 to define the

activities of VISION 2020. A detailed report with clear articulation of the deliverables at the national level will be presented in the next issue. With the support of LVPEI, VISION 2020 India has been facilitating the publication of the Indian supplement of the Journal of CEH. VISION 2020 India will evolve strategies to work closely with the Government of India and the state governments so that it brings a synergy to the eye care activities of its members in a manner that aligns with national and global objectives. VISION 2020 India was born out of the recognition of the significant contribution to eye care by the non-governmental sector. This should evolve into a platform for closer cooperation and collaboration within the NGO sector, both International and National towards making the VISION 2020 National Plan of Action a reality.

The plan for next Quarter:

- Inviting membership from eye care organizations - INGOs, NNGOs, teaching hospitals and the corporate sector in the country
- Preparation for celebration of World Sight Day on 14 October
- Appreciation workshop for the stakeholders on VISION 2020
- Initiating a series of activities in the areas of advocacy, human resource development, operational research, etc., through partner organisations to develop eye care protocols, best practices, curricula and other resources

We request the eye care fraternity in India to join us in the eradication of avoidable blindness in our country.

Abstract

Aetiology of childhood cataract in south India

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AIM: To identify the causes of childhood cataract in south India with emphasis on factors that might be potentially preventable. **METHODS:** A total of 514 consecutive children with cataract attending an eye hospital outpatient clinic were examined and their parents interviewed by a trained interviewer using

a standardised questionnaire in the local language. Serology was performed on children under 1 year of age to detect congenital rubella syndrome (CRS). Other investigations were performed as clinically indicated. **RESULTS:** Of the 366 children with non-traumatic cataract 25% were hereditary, 15% were due to congenital rubella syndrome, and 51% were undetermined. In children under 1 year of age 25% were due to rubella and cataract of nuclear morphology had a 75% positive predictive value for CRS. Mothers of children in the undetermined group were more likely to have taken abortifacients than a group of age matched controls ($p=0.1$) but use of other medications in

pregnancy was similar in both groups. Of the 148 (29%) children with traumatic cataracts three quarters were over the age of 6 years. Stick injuries were responsible for 28%, thorn injuries for 21%, and firecrackers for 5%. **CONCLUSION:** Nearly half of non-traumatic cataract in south India is due to potentially preventable causes (CRS and autosomal dominant disease). There is need for further work to identify the factors leading to childhood cataract in at least half of the cases for which no definite cause can as yet be determined.

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