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Editorial

The challenge of childhood cataract blindness

Childhood blindness remains a cause of great concern globally despite large advances in the field of ophthalmology in the past few decades. Childhood visual impairment and blindness are due to preventable causes in most cases[1]. With over 1.4 million children blind throughout the world, low vision that can significantly hinder activities of daily living is present in over 17.5 million[1]. Cataract forms a large share of the preventable blindness with the latest WHO estimates (2010) ranging above 200,000 cases world over[2]. Developing countries like India and China form the epicenter of the problem with more than 20 per cent of their population visually impaired[1]. High percentages are also reported from Africa (15%) and Eastern Mediterranean Region (EMR)[1, 2]. These figures are likely underestimating the actual numbers because many children with multiple disabilities may go unreported. Economic burden and years of productive life lost due to untreated paediatric cataract clearly favours urgent attention to this challenge[1-3].

The best way to address a problem is to delve into the root cause: There are numerous lacunae in the understanding of aetio-pathogenesis of infantile/ developmental cataract[3,4]. While certain causes of cataract in children are well-documented, including perinatal and intrauterine infections, many cases may go without a definite aetiological diagnosis[3]. Childhood cataract is associated with genetic[5,6], metabolic[7] and acquired causes, such as trauma[8], retinopathy of prematurity (ROP)[4], enzyme defects[9] and idiopathic. Drug-induced cataracts due to steroids are also common in this age group. Certain genetic defects are associated with abnormal lenticular shape/position needing surgical management including entities like spherophakia[10] and lenticonus[11]. Mutations in crystallin and connexin genes have been identified as putative causes of cataract in children. Genetic polymorphisms may be associated with increased incidence of cataract, some of which are yet to be identified[4,6].

The patients are hidden in the community - we need to identify them: There is often a delay in the diagnosis of paediatric cataract that compounds the problem[2]. Delayed treatment is associated with permanent visual impairment even after adequate specialized care due to development of severe amblyopia. Vigorous case-search and prompt referral are areas that need to be strengthened if we desire to tackle the problem successfully[1,12]. Thorough visual acuity assessment using specialized charts, pupillary reaction, strabismus and ocular motility must be done. Slit-lamp biomicroscopy is the best method to evaluate lenticular opacities and study the morphology of cataract[4]. Intraocular pressure measurement is a must to rule out congenital glaucoma which can frequently co-exist. Fundus examination is performed by indirect ophthalmoscopy after pupillary dilation to rule out posterior segment pathology. Detailed evaluation of family history, features of congenital rubella syndrome, metabolic diseases and survey of enzyme defects must follow if clinical symptoms suggest.

The biggest challenge in paediatric cataract surgery is gearing up for the next challenge: Though there have been tremendous changes with the ushering of technology in the field of ophthalmology but the task of managing childhood cataract remains difficult, fraught with potential complications at every stage[3,13]. Presently, the modern technique of phacoaspiration with posterior chamber intraocular lens implantation in the capsular bag is the preferred technique to manage paediatric cataract[3,4,13-15]. This has replaced older techniques of lensectomy and extracapsular cataract surgery. Ultrasound B-scan rules out posterior segment pathology in children with dense cataracts. Calculation of intraocular lens (IOL) power is a subject

in itself with more than a dozen guidelines available in literature[3,13-16]. Popular guidelines include those laid down by Dahan and Drusedau[16], where an under-correction is performed keeping in mind the future growth of the eye.

A child's eye is different from those of an adult in many more ways than one: Unlike for adult cataracts, there is a sense of urgency in the management of childhood cataracts, especially unilateral cases given the poor prognosis associated with delayed surgery[3,4,14,15]. Certain attributes such as low scleral rigidity, highly elastic anterior capsule and increased vitreous pressure contribute to surgical difficulty. Hence, interventions may be limited to dedicated, specialized centers[17]. Phacoaspiration in children is performed using automated systems under controlled parameters. Specialized Ophthalmic Viscosurgical Devices (OVDs) are used, *viz*. 1.4 per cent sodium hyaluronate to maintain the integrity of anterior chamber intraoperatively. After continuous curvilinear capsulorhexis (CCC), cortical aspiration is performed[14,15]. In young children aged 1 month to 6 years, posterior capsulotomy is done as a primary procedure with limited anterior vitrectomy to minimize the risk of posterior capsular opacification (PCO) and further interventions[18]. The current preferred practice to manage surgical aphakia is to place posterior chamber IOL in children to achieve optimal visual rehabilitation[3,4,14-16,19]. Extensive studies have proven that hydrophobic acrylic IOLs perform the best with lesser complication rates compared to older IOLs[3,14,15,20].

Cornerstone of successful paediatric cataract surgery is the post-operative management: Post-operative care in children is highly specialized as paediatric eyes are susceptible to high levels of inflammation[18]. High dose steroids are required topically combined with antibiotic therapy. Complicated cases of uveitis and trauma may benefit from intraocular depot steroids like dexamethasone implants[21]. Visual axis obscuration (VAO) can occur in the post-operative period due to formation of PCO or other IOL-related complications like tilt, decentration or subluxation/dislocation[18]. Maintaining a clear visual axis by managing VAO and prompt institution of occlusion therapy are *sine-quo-none* for achieving optimal amblyopia therapy.

The future holds promise: Futuristic approach includes surgery with newer IOL designs including multifocal IOLs in children[22], bag-in-the-lens IOL[23], use of newer synthetic suture materials, better vitrectomy devices, innovative instruments like plasma blade, maneuvers like optic capture and array of more efficacious drugs to control inflammation[3,4,8,13,15,19]. Benchmarks for post-operative outcomes after paediatric cataract surgery have risen and presently, the focus is on achieving highest grades of binocularity, stereopsis and contrast sensitivity along with emmetropia[22].

With increasing evidence of genetic polymorphisms responsible for cataract, genetic counselling is probably going to play an important role in the management of these patients in the future[24]. Identification of mutations responsible for early cataractogenesis can help formulate guidelines regarding genetic counselling and prenatal diagnosis in the future[24]. It may become possible to identify cataract very early and intervene before damage is done.

To conclude, the future of paediatric cataract management is exciting. It is the need of the hour to take cognizance of this challenge and the rewards will be truly gratifying.

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References

1. Pascolini D, Mariotti SP. Global estimates of visual impairment: 2010. *Br J Ophthalmol* 2012; *96* : 614-8.
2. Open Database: World Health Organization. Visual impairment and blindess. Factsheet N°282 updated August 2014. Available from: *http://www.who.int/mediacentre/factsheets/fs282/en/*, accessed on October 8, 2014.
3. Wilson ME, Pandey SK, Thakur J. Paediatric cataract blindness in the developing world: surgical techniques and intraocular lenses in the new millennium. *Br J Ophthalmol* 2003; *87* : 14-9.
4. Lambert SR, Drack AV. Infantile cataracts. *Surv Ophthalmol* 1996; *40* : 427-58.
5. Mostafa MSE, Temtamy S, El-Gammal MY, Sayed SI, Abdel-Salam M, El-Baroudy R. Genetic studies of congenital cataract. *Metab Pediatr Ophthalmol* 1981; *5* : 233- 42.
6. Devi RR, Yao W, Vijayalakshmi P, Sergeev YV, Sundaresan P, Hejtmancik JF. Crystallin gene mutations in Indian families with inherited pediatric cataract. *Mol Vis* 2008; *14* : 1157-70.
7. Stambolian D. Galactose and cataract. *Surv Ophthalmol* 1988; *132* : 333-49.

8. Sukhija J, Ram J. Pediatric traumatic cataract: maximizing the surgical outcome. *J Cataract Refract Surg* 2012; *38* : 2210-1.

9. Singh R, Ram J, Kaur G, Prasad R. Galactokinase deficiency induced cataracts in Indian infants: identification of 4 novel mutations in *GALK* gene. *Curr Eye Res* 2012; *37* : 949-54.

10. Ram J, Gupta N. Bilateral spontaneous anterior dislocation of crystalline lens in an infant. *Lancet* 2011; *378* : 1501.

11. Cheng KP, Hiles DA, Biglan AW, Pettapiece MC. Management of posterior lenticonus. *J Pediatr Ophthalmol Strabismus* 1991; *28* : 143-9.

12. Courtright P. Childhood cataract in sub-Saharan Africa. *Saudi J Ophthalmol* 2012; *26* : 3-6.

13. Apple DJ, Ram J, Foster A, Peng Q. Elimination of cataract blindness: A global perspective entering new Millennium. *Survey Ophthalmol* 2000; *45* (Suppl): 1-196.

14. Ram J, Gupta N, Sukhija JS, Chaudhary M, Verma N, Kumar S, *et al*. Outcome of cataract surgery with primary intraocular lens implantation in children. *Br J Ophthalmol* 2011; *95* : 1086-90.

15. Vasavada AR, Nihalani BR. Paediatric cataract surgery. *Curr Opin Ophthalmol* 2006; *17* : 54-61.

16. Dahan E, Drusedau MU. Choice of lens and dioptric power in pediatric pseudophakia. *J Cataract Refract Surg* 1997; *23* : 618-23.

17. Ram J, Sukhija J, Thapa BR, Arya VK. Comparison of hospital versus rural eye camp based pediatric cataract surgery. *Middle East Afr J Ophthalmol* 2012; *19* : 141-6.

18. Ram J, Brar GS, Kaushik S, Gupta A, Gupta A. Role of posterior capsulotomy with vitrectomy and intraocular lens design and material in reducing posterior capsule opacification after pediatric cataract surgery. *J Cataract Refract Surg* 2003; *29* : 1579-84.

19. Fan DS, Yip WW, Yu CB, Rao SK, Lam DS. Updates on the surgical management of paediatric cataract with primary intraocular lens implantation. *Ann Acad Med Singapore* 2006; *35* : 564-70.

20. Ram J, Apple D, Peng Q, Visessook N, Auffarth GU, Schoderbek RJ Jr, *et al*. Update on fixation of rigid and foldable posterior chamber intraocular lenses. Part II: Choosing the correct haptic fixation and intraocular lens design to help eradicate posterior capsule opacification. *Ophthalmology* 1999; *106* : 891-900.

21. Gupta A, Ram J, Gupta A, Gupta V. Intraoperative dexamethasone implant in uveitis patients with cataract undergoing phacoemulsification. *Ocul Immunol Inflamm* 2013; *21* : 462-7.

22. Ram J, Agarwal A, Kumar J, Gupta A. Bilateral implantation of multifocal versus monofocal intraocular lens in children above 5 years of age. *Graefes Arch Clin Exp Ophthalmol* 2014; *252* : 441-7.

23. Tassignon MJ, De Veuster I, Godts D, Kosec D, Van den Dooren K, Gobin L. Bag-in-the-lens intraocular lens implantation in the pediatric eye. *J Cataract Refract Surg* 2007; *33* : 611-7.

24. Deng H, Yuan L. Molecular genetics of congenital nuclear cataract. *Eur J Med Genet* 2014; *57* : 113-22.