

Congenital Cataract Screening

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Abstract

Congenital cataract is a leading cause of visual deprivation which can damage the developing visual system of a child; therefore early diagnosis, management and long-term follow-up are essential. It is recommended that all neonates be screened by red reflex examination at birth and suspected cases be referred to ophthalmic centers. Early surgery (<6 weeks of age, based on general neonatal health) is important for achieving the best visual outcome particularly in unilateral cases. In bilateral cases, surgery is highly recommended before appearance of strabismus or nystagmus (<10 weeks of age) with no longer than a one-week interval between the fellow eyes. Parents should be informed that surgery is a starting point and not the endpoint of treatment. Appropriate postoperative management including immediate optical correction in the form of aphakic glasses or contact lenses, or intraocular lens (IOL) implantation at the appropriate age (>1 year) is highly recommended. After surgery, amblyopia treatment and periodic follow-up examinations should be started as soon as possible to achieve a satisfactory visual outcome. Practitioners should consider the possibility of posterior capsular opacity, elevated intraocular pressure and amblyopia during follow-up, especially in eyes with microphthalmia and/or associated congenital anomalies. All strabismic children should undergo slit lamp examination prior to strabismus surgery to rule out congenital lens opacities. From a social point of view, equal and fair medical care should be provided to all children regardless of gender.

Keywords: Congenital Cataract; Screening; Management

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Congenital cataracts and other disorders of visual deprivation can damage the developing visual system of a child, and permanently reduce central and peripheral vision. Therefore early diagnosis and surgery, appropriate refractive error correction, amblyopia therapy and long-term follow-up are essential.^[1]

It is well known that recovery of normal visual function after cataract surgery is more probable in

adults as compared to children due to impairment of the developing visual system.^[2,3]

The prevalence of congenital cataract has been reported from 1 to 15 per 10,000 children worldwide, whereas it ranges from 1 to 3 per 10,000 births in developing countries.^[4] The number of blind children due to congenital cataracts globally and in developing countries are 200,000 and 133,000, respectively.^[5]

Red reflex examination at birth is an easy method to screen for congenital cataracts leading to early diagnosis and timely surgery.^[6] The sensitivity and specificity of red reflex examination with no pupil dilation immediately after birth have been reported as 85 and 38.50%, respectively for detection of all types

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of congenital ocular diseases,^[7] thus pediatricians should be aware of the possibility of false positive results.^[8]

Accompanying abnormalities such as ocular hypoplasia and early or late surgical complications such as inflammation and glaucoma may adversely affect visual outcomes in children undergoing congenital cataract surgery.^[1]

Birch et al showed that late surgery may be considered as one of the leading causes of decreased best-corrected visual acuity (BCVA), and only 53% of cases with history of late congenital cataract surgery achieved BCVA of 0.60 LogMAR or better.^[9]

According to the literature, the best visual outcomes may be achieved when surgery is performed during the first 6 weeks of age in unilateral cases and within 5 to 8 weeks of age in bilateral cases.^[10] Furthermore, age at surgery and time interval between affected fellow eyes in developed countries have been reported less than one year and one week, respectively.^[11-14] However, in a recent study by the authors of the current perspective, these figures were 3 years and 3 months,^[1] respectively. This difference may be explained by lack of routine early screening for media opacities in our neonates resulting in late diagnosis and late surgery, poor economic status, or a combination of the above-mentioned causes.

Based on the literature, mean BCVA is 0.9 (range, 0.2-1.4) LogMAR in unilateral and 0.57 (range, 0.2-1.0) LogMAR in bilateral congenital cataract subjects.^[10,15,16] Our results were consistent with these findings (0.90 LogMAR in unilateral and 0.35 LogMAR in bilateral cases).^[1] Since the prognosis for good VA in unilateral cases is disappointing as compared to bilateral cases, earlier diagnosis, surgery, and regular follow-up are mandatory.

Amblyopia is the major cause of visual impairment following congenital cataract surgery.^[10,11] It was the reason for decreased vision in 76% of cases in the study by Ledoux et al^[17] and was also observed in 56% of our cases^[1] which can be due to unilateral cataract, late surgery, longer time interval between fellow eyes and lack of compliance with amblyopia therapy.

Strabismus following congenital cataract has been reported from 13% to 86% in the literature,^[16,17] and was observed in 47.6% of our cases.^[1] Ocular deviation in these children represents unequal reduced VA in their both eyes due to unilateral or asymmetric bilateral lens opacities.

Non-desirable surgical outcomes have been reported even in subjects with early cataract surgery, wearing appropriate glasses, contact lenses or implantation of intraocular lenses (IOL), which has been due to the lack of long-term follow-up (at least up to the age of 10 years) stressing the fundamental role of follow-up visits for management of possible complications in children with congenital cataracts.^[9]

In the study by Rajavi et al,^[1] risk factors for lower postoperative visual acuity included unilateral cataract, nystagmus, female gender (due to discrimination), strabismus and all types of congenital cataract except the zonular type. Furthermore, lack of parental compliance for treatment of amblyopia, longer surgical interval, and delayed surgery were other risk factors for not gaining an acceptable VA after the operation.^[9,15,18,19]

Female gender is still considered as the second-grade sibling in the traditional families with the excuse of sons' family responsibility in the future;^[1] therefore, parents are recommended to concern over the health of their children regardless of their gender.

Posterior capsular opacity (PCO) and glaucoma are major complications of cataract surgery in children.^[10,17] Although the possibility of PCO is reduced by posterior capsulotomy and anterior vitrectomy during cataract surgery, glaucoma is still considered as a significant late complication in these cases requiring specific attention.

All practitioners, caregivers and parents should be aware of the following points:

1. Screening for red reflex opacities in all neonates is highly recommended, and suspected cases should be referred to a pediatric ophthalmologist for further examination. The importance and simplicity of performing this test makes it a feasible screening method.
2. Parents should be informed about the long therapeutic process and aware that cataract surgery is a starting and not the ending point of treatment for a child with congenital cataract.
3. Early cataract surgery, particularly in unilateral cases, is recommended (<6 weeks, based on the general neonatal health).^[10] Contact lens fitting rapidly after surgery and IOL implantation at the appropriate age is also recommended.
4. Bilateral congenital cataracts should be operated before strabismus or nystagmus appears (<10 weeks)^[10] with no longer than one-week interval between fellow eyes. Refractive error correction by aphakic glasses or contact lenses is recommended. IOL implantation at the appropriate age (>1 year) is suggested.
5. Due to the possibility of ocular deviation secondary to congenital lens opacity, all strabismic children should have slit lamp examination prior to strabismus surgery.
6. After congenital cataract surgery, amblyopia treatment should be started as soon as possible together with periodic follow-up examinations in order to achieve a satisfactory visual outcome.
7. The practitioners should consider the possibility of PCO, elevated IOP and amblyopia at each follow-up visit especially in cases with concurrent microphthalmia and/or associated congenital anomalies.

8. It is expected that equal and fair medical care should be provided to all children regardless of gender.

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Conflicts of Interest

There are no conflicts of interest.

REFERENCES

- Rajavi Z, Mokhtari S, Sabbaghi H, Yaseri M. Long-term visual outcome of congenital cataract at a Tertiary Referral Center from 2004 to 2014. *J Curr Ophthalmol* 2016;27:103-109.
- Lai FH, Lok JY, Chow PP, Young AL. Clinical outcomes of cataract surgery in very elderly adults. *J Am Geriatr Soc* 2014;62:165-170.
- Nagamoto T, Oshika T, Fujikado T, Ishibashi T, Sato M, Kondo M, et al. Surgical outcomes of congenital and developmental cataracts in Japan. *Jpn J Ophthalmol* 2016;60:127-134.
- Lin H, Yang Y, Chen J, Zhong X, Liu Z, Lin Z, et al. Congenital cataract: Prevalence and surgery age at Zhongshan Ophthalmic Center (ZOC). *PLoS One* 2014;9:e101781.
- Tomkins O, Ben-Zion I, Moore DB, Helveston EE. Outcomes of pediatric cataract surgery at a tertiary care center in rural southern Ethiopia. *Arch Ophthalmol* 2011;129:1293-1297.
- Eventov-Friedman S, Leiba H, Flidel-Rimon O, Juster-Reicher A, Shinwell ES. The red reflex examination in neonates: An efficient tool for early diagnosis of congenital ocular diseases. *Isr Med Assoc J* 2010;12:259-261.
- Mussavi M, Asadollahi K, Janbaz F, Mansoori E, Abbasi N. The evaluation of red reflex sensitivity and specificity test among neonates in different conditions. *Iran J Pediatr* 2014;24:697-702.
- American Academy of Pediatrics; Section on Ophthalmology; American Association for Pediatric Ophthalmology And Strabismus; American Academy of Ophthalmology; American Association of Certified Orthoptists. Red reflex examination in neonates, infants, and children. *Pediatrics* 2008;122:1401-1404.
- Birch EE, Wang J, Felius J, Stager DR Jr., Hertle RW. Fixation control and eye alignment in children treated for dense congenital or developmental cataracts. *J AAPOS* 2012;16:156-160.
- Lambert SR, Lynn MJ, Reeves R, Plager DA, Buckley EG, Wilson ME. Is there a latent period for the surgical treatment of children with dense bilateral congenital cataracts? *J AAPOS* 2006;10:30-36.
- Birch EE, Cheng C, Stager DR Jr., Weakley DR Jr., Stager DR Sr. The critical period for surgical treatment of dense congenital bilateral cataracts. *J AAPOS* 2009;13:67-71.
- Jain S, Ashworth J, Biswas S, Lloyd IC. Duration of form deprivation and visual outcome in infants with bilateral congenital cataracts. *J AAPOS* 2010;14:31-34.
- Chak M, Wade A, Rahi JS; British Congenital Cataract Interest Group. Long-term visual acuity and its predictors after surgery for congenital cataract: Findings of the British congenital cataract study. *Invest Ophthalmol Vis Sci* 2006;47:4262-4269.
- Magnusson G, Bizjajeva S, Haargaard B, Lundström M, Nyström A, Tornqvist K. Congenital cataract screening in maternity wards is effective: Evaluation of the Paediatric Cataract Register of Sweden. *Acta Paediatr* 2013;102:263-267.
- Lundvall A, Zetterström C. Primary intraocular lens implantation in infants: Complications and visual results. *J Cataract Refract Surg* 2006;32:1672-1677.
- Hussin HM, Markham R. Long-term visual function outcomes of congenital cataract surgery with intraocular lens implantation in children under 5 years of age. *Eur J Ophthalmol* 2009;19:754-761.
- Ledoux DM, Trivedi RH, Wilson ME Jr., Payne JF. Pediatric cataract extraction with intraocular lens implantation: Visual acuity outcome when measured at age four years and older. *J AAPOS* 2007;11:218-224.
- Eriksen JR, Bronsard A, Mosha M, Carmichael D, Hall A, Courtright P. Predictors of poor follow-up in children that had cataract surgery. *Ophthalmic Epidemiol* 2006;13:237-243.
- Lloyd IC, Ashworth J, Biswas S, Abadi RV. Advances in the management of congenital and infantile cataract. *Eye (Lond)* 2007;21:1301-1309.