

# Requirement of Optometric Services in Children with Low Vision Due to **Congenital Eye Anomalies**



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# **bstract**

Purpose: Purpose of this study was to evaluate specific needs of patients with low vision due to congenital eye anomalies

Introduction: Severe visual impairment (SVI) and blindness in infants must be detected as early as possible to initiate immediate treatment to prevent deep amblyopia. Problems in the embryonic developmental process can lead to congenital eye malformations, such as albinism, amblyopia, high congenital refractive errors, strabismus, retinopathy of prematurity, nystagmus, anophthalmia, microphthalmia, coloboma and aniridia. Symptoms include an abnormal-looking eye and reduced eye vision. After medical and surgical assistance, a child may require optometric services e.g. spectacles, low vision aids, contact lens, rehabilitation techniques and counseling.

Material & methods: Sixty five children visually impaired but still having some functional vision with congenital eye anomalies were included in the study. All those children were. All cases were checked, diagnosed, treated (medically and surgically) by ophthalmologists before being referred to the optometrist

Results: Twenty two children had myopic shift, Sixteen children showed hypermetropic shift and Fifteen children showed astigmatism with all types. Twelve children were having no refractive error or a little amount of refractive error which was not considered significant. These, were directly treated with low vision aids.

Forty (61%) children having Visual Acuity equivalent to or better than 6/18, showed improvement of visual acuity of one or two lines with refraction or contact lens.

Twenty two (33%) cases, having visual acuity less than 6/18, were managed with distance low vision aids. Three (4%) out of 65 children were having PL visual acuity. All those were treated with non optical low vision devices and counseling

49 (75%) children could read 12M on Near Light House Visual Acuity chart at 40 cm unaided or with distance refraction. 13 (20%) children were provided with magnifiers to reach this level. Three (4.6%) children having PL distance VA showed no results on near VA chart unaided, aided or with distance correction



#### INTRODUCTION

Approximately 1 in 250 (0.4%) children are estimated to suffer from visual impairment as a direct consequence to eye involvement. Data regarding visually impaired children can be obtained from a variety of sources such as eve departments of hospitals & clinics, school screening programmes, community based rehabilitation (CBR) programmes or special schools for the visually impaired.1

Children are said to have 'low vision' or 'partial sight' when they have: (a) a corrected visual acuity in the better eve of <6/18 to 'perception of light' (or a visual field of less than 10 degrees); and (b) the ability to use their residual vision to orientate themselves or to perform tasks.2

A congenital eye disease is a disease of the eye that is present at birth and usually patients may develop an illness during pregnancy that passes to the fetus resulting in birth defects. However, some congenital eye diseases do not originate from an underlying defect and may not be suspected until after the baby is born.

Symptoms include an abnormal-looking eye and reduced eye vision. The problems are usually apparent in an eye examination. A patient may occasionally require further systemic tests, such as an MRI, CT scan or ultrasound. After medical and surgical assistance, a child may require optometric services e.g. spectacles, low vision aids, contact lens, rehabilitation techniques and counseling.

Albinism is a hereditary condition characterized by a variable lack of pigment in the eyes, skin, or hair. People with albinism are sensitive to bright light and glare and may wear tinted eyeglasses. Bifocals, magnifiers, and other optical devices can help people with albinism.3In amblyopia early treatment is advisable, because if left untreated, this condition may lead to permanent vision problems. Treatment options include vision therapy exercises or prescription eyeglasses and patching.4

Partial or complete absence of the iris of the eye (aniridia) is usually present at birth. People with aniridia may benefit from optometric services such as wearing tinted contact lenses or sunglasses, using magnifiers, and avoiding intense or glaring light.5

High refractive errors including astigmatism, usually present since birth. Astigmatism can be corrected with eveglasses or contact lenses. Corrective surgery is another option.6

Children whose vision is impaired by coloboma may benefit from using reading materials that have large black print and well-spaced letters and words. They may also find it helpful to read one line at a time with the aid of a cutout reading window.7

Nystagmus may be present at birth or may occur. later on, from disease or injury. Children with nystagmus may frequently lose their place when reading. Placing a cutout reading window over words or using a card to "underline" text can be helpful.8

Retinitis pigmentosa is a rare, inherited disease for which there is as yet no treatment or cure. Early diagnosis enables a person with the disease to plan and prepare for its progression. In addition, depending on the degree of vision loss, electronic magnifiers, night-vision scopes, and other such special devices for impaired vision can provide some benefit for people with the disease.9

Retinopathy of prematurity can lead to reduced vision or blindness. Children who experience minor effects may benefit from the use of devices for low vision as they get older.10

Stargardt's Disease causes gradual degeneration of the macula. There is no cure or treatment for Stargardt's Disease, but such devices as magnifying screens and binocular lenses can help people cope with vision limitations.<sup>11</sup> Strabismus is most commonly an inherited condition, but may also be caused by disease or injury. The condition may be treated with corrective eyeglasses, eye-muscle exercises, surgery, or a combination of these approaches. Young children with this condition may need to wear an eve patch over their stronger eye to force their weaker eye to function correctly. Children whose strabismus is not corrected may develop amblyopia.12

Congenital eye defects can impair vision or even cause blindness. Undetected, such problems can lead to difficulties which can persist into adulthood and result in lack of self-confidence, together with lack of educational attainment and job opportunities.13

The major barrier to accessing eye care services for infants is the lack of trained personnel who can diagnose a problem early. Pediatric eye care centers are rare in developing countries like Pakistan. There are financial and geographic barriers for many parents from poorer communities. Diagnostic, curative, and rehabilitative services are not available in many regions. Another significant barrier is the lack of knowledge on the part of healthcare providers.<sup>14</sup>

Even when the infant has been referred for treatment appropriately, many parents believe that their infant is too small to undergo surgery or wear spectacles.

However, a child who cannot be helped by medicine or surgery may still benefit from use of spectacles and/or low



vision aids. Completely and irreversibly blind children can benefit by rehabilitation and special school education. This should be emphasized during parent counseling.

Blindness and severe visual impairment in infants is not that difficult to detect and diagnose. With proper care. most of these infants can be helped and formation of dense amblyopia prevented. Even if the ophthalmologist may not be able to help medically or surgically, optical aids and rehabilitation can help children reach their full capacity.

#### Purpose:

The aim of the study was

- (1) To determine the need for spectacles and contact lenses in children with congenital eye anomalies
- (2) To describe their refractive errors, assess their needs for low vision aids (LVAs) and rehabilitation services.

#### Inclusion criteria:

- 1. Children with congenital eye abnormalities.
- Verbal children were included.
- 3. Those children were included who have done with all medical and surgical intervention.

#### **Exclusion criteria:**

- 1. Children aged from 0-4 years were excluded
- 2. Mentally retarded children
- 3. Those who are under medical or surgical treatment

#### Methods:

Children with congenital eye anomalies were recruited from special education for the blind and hospital records in Lahore Pakistan. Those whose distance vision in their better eye improved with refraction were prescribed spectacles.

Those unable to read 12M on The Light House Near Visual Acuity Chart were assessed for LVAs for near. Those with distance visual acuity of < 6/18 in the better eye were assessed for telescopes to aid distance vision.

Sixty five children with congenital eye anomalies were included in the study. All those children were visually impaired but still having some functional vision. All cases were checked, diagnosed, treated (medically and surgically) by ophthalmologists before visiting optometrist.

Many physicians limit "optometric services" to "refraction" only. They are unaware of use and advantages of contact lens, proper dispensing benefits, low vision aids and rehabilitation services. They consequently don't refer their patients and patients go untreated.

In this study, patients with congenital eye defects

were provided with these optometric services:

- 1. Refraction, dispensing and selection of appropriate frame and lenses
- 2. Contact lenses in cases where needed either to improve VA or to decrease photophobia
- 3. Low vision aids were provided to those who could not get benefit from spectacles and contact lenses.
- 4. Scanning and tracking techniques were given to the children for fluent reading.
- Training of low vision aids and counseling was done to maximize the results.
- Students were trained to use their residual vision as per maximum. They were taught and trained "how to live independently by using their remaining vision"

#### Results:

Congenital cataract	4
Retinopathy of prematurity	6
Albinism, nystagmus and aniridia	6
High refractive errors	10
Retinitis pigmentosa	16
Optic atrophy/ macular degenerations	4
Microphthalmos	4
Amblyopia	6
Others	9

#### Distribution according to Refractive error:

Out of total sixty five children, 22 children had myopic shift, 16 showed hypermetropic shift.

Fifteen children were having astigmatism with all types.

Twelve patients had no refractive error or a little amount of refractive error which was not considered significant. These patients were directly treated with low vision aids.

### Results after optometric services:

After every child's careful optometric assessment and management, 40 (61%) children having Visual Acuity equivalent to or better than 6/18, showed improvement of visual acuity of one or two lines on ETDRS Distance visual acuity chart at 4 meters with refraction or contact lens.

Twenty two (33%) cases were managed with distance low vision aids (telescope) having less visual acuity than 6/18 in better-seeing eye with best possible refractive correction.

Three (4%) out of 65 children were having PL visual acuity. All those were treated with non optical low vision devices and other rehabilitative measures.

Forty nine (75%) patients among these 65 patients could read 12M on near light house visual acuity chart at 40



cm unaided or with distance refraction. Thirteen (20%) patients were provided with magnifiers to reach this level. Three (4.6%) patients having PL distance VA showed no results on near VA chart unaided, aided or with distance correction.

#### Conclusion:

Children with congenital anomalies of the eve and functional vision benefit from refraction and low vision services.

#### Discussion:

Children with aphakia, coloboma, refractive errors and microphthalmos benefit from spectacles. According to Pal, out of 124 children with low vision but having useful residual vision, 51 (41.1%) were able to read N-10 unaided or with distance spectacles and 30 children (22.6%) improved to N-10 with spectacle magnifiers and were prescribed the same. He concluded that visually impaired children with aphakia and congenital anomalies of the eye benefit from refraction and low vision services. 15

Medical therapy or surgery are rarely effective in congenital cases; however, refraction and low vision aids can help many of these cases.<sup>16</sup> Many children with congenital defects can be taught to read print with low vision aids instead of Braille.

It is important to ensure that children with low vision are refracted and provided with any spectacles they require. Work in West Africa indicates that at least 30% of children with low vision need spectacles.17 Refraction should always be carried out before a magnification assessment.

In the majority of cases, even with in-depth molecular genetic testing and detailed investigation, no cause can be identified.18 This is because these anomalies are likely to be due to genes controlling eye development (which are largely unknown) and possibly, gene-environment interactions, reflecting similar processes elucidated for folic acid and spina bifida.1

A study related to ourstudy describes and gives same results. Retinal dystrophies have variable genetic inheritance and are common in communities with high rates of consanguinity. Although the dystrophy may only give visual symptoms later in life, this is a common cause of blindness in children worldwide, particularly in Middle Eastern countries. Low vision aids are effective for navigation and reading for individuals affected with retinal dystrophies. Genetic counseling is required if more than one sibling is involved.<sup>20</sup> The birth prevalence of cataract is 3-5/1,000 live births.<sup>21</sup> while monogenetic abnormalities, syndromes, metabolic disorders, genetic disorders, and congenital rubella syndrome are

recognized causes of this condition, in the majority, the cause is unknown.22

The importance of congenital cataract is increasing as a cause of blindness among children in developing countries, with other causes declining. There can be 2 years' delay from cataract diagnosis to presentation for surgery.<sup>23</sup> Early recognition and referral is essential to prevent development of dense amblyopia. However, the timing of intraocular lens (IOL) implantation remains controversial.<sup>24</sup> Generally speaking, the target postoperative refraction is emmetropia by the age of 5-6 years. Aphakic glasses have to be corrected for near vision.

The improvement in neonatal care can lead to an increase in survival rates of premature infants in middleincome countries and major cities of even some not so affluent countries. Premature infants are at increased risk of developing refractive errors, strabismus, amblyopia, and low vision; hence, long-term follow-up is recommended.

Low vision aids and rehabilitation are often the only recourse for optic nerve lesions and cerebral visual impairment. Often, such children are severely handicapped, which makes assessment and management even more challenging.

#### References:

- 1. Ager L. Optical services for visually impaired children. J Comm Eye Health 1998;11(27): 38-40.
- 2. The management of low vision in children. Proceedings of WHO/PBL consultation, Bangkok, July 1992. WHO, 1993.
- 3. Low Vision Aids. [Serial online] [Accessed March 2012]. Available from URL: http:// www.albinism.org/publications/low-vision-aids.html
- 4. Prevent Blindness America. Amblyopia (Lazy Eye). [Serial online][Accessed February 2012]. Available from URL: http://www.preventblindness.org/children/ amblyopiaFAQ.html
- 5. American Foundation for the Blind. Aniridia. [Serial online][Accessed April 2012]. Available from URL: http://www.afb.org/section.aspx?SectionID=74&SubT opicID=112&DocumentID=3833www.nei.nih.gov/heal th/cornealdisease/index.asp
- 6. American Optometric Association. Astigmatism [serial



- online][Accessed April 2012]. Available from URL: http://www.aoa.org/Astigmatism.xml
- 7. Bishop VE. Selected Anomalies and Diseases of the Eye [serial online] [Accessed April 2012]. Available from URL: http://www.tsbvi.edu/attachments/091 eve.rtf
- 8. American Nystagmus Network. About nystagmus. [Serial online] [accessed April 2012]. Available from URL: http://www.nystagmus.org/new.aboutn.php
- 9. Prevent Blindness America. Retinitis Pigmentosa. [Serial online][Accessed February 2012]. Available from URL: http://www.preventblindness.org/retinitispigmentosa
- 10. National Eye Institute. Retinopathy of Prematurity. [Serial online] [Accessed April 2012]. Available from URL: http://www.nei.nih.gov/health/rop/index.asp
- 11. www.mdsupport.org/library/stargrdt.html
- www.cnib.ca/eng/eye\_con/cospubs/strabsms.htm 12.
- Davidson S, Quinn GE. The impact of pediatric vision 13. disorders in adulthood. Pediatrics. 2011 Feb;127(2):334-9. Epub 2011 Jan 3.
- Severe Visual Impairment and Blindness in Infants: 14. Causes and Opportunities for Control. Parikshit Gogate, 1.2 Clare Gilbert, 3 and Andrea Zin4
- 15. Pal N, Titival JS, Tandon R, Vajpayee RB, Gupta S, Murthy GV. Need for optical and low vision services for children in schools for the blind in North India. Indian J Ophthalmol. 2006 Sep;54(3):189-93.
- 16. Hornby SJ, Adolph S, Gothwal VK, Gilbert CE, Dandona L, Foster A. Requirements for optical services in children with microphthalmos, coloboma and microcornea in southern India. Eve. 2000; 14:219-24.
- 17. Ager LR. Annual report of low vision services, Ghana National Eye Care Programme, 1996.
- 18. Shah SP, Taylor AE, Sowden JC, Ragge NK, Russell-Eggitt I, Rahi JS, et al. Anophthalmos, micophthalmos and typical coloboma in the United Kingdom: A prospective study of incidence and risk. Invest

- Ophthalmol Vis Sci. 2011;52:558-64.
- 19. Hornby SJ, Ward SJ, Gilbert CE. Hypothesis: Eye birth defects in humans may be caused by a recessivelyinherited genetic predisposition to the effects of maternal vitamin A deficiency during pregnancy. Med Sci Monit. 2003:9:23-6.
- 20. Kotb AA, Hammouda EF, Tabbara KF. Childhood blindness at a school for the blind in Riyadh, Saudi Arabia. Ophthalmic Epidemiol. 2006;13:1-5.
- 21. Rahi JS, Dezateux C. British Congenital Cataract Interest Group. Measuring and interpreting the incidence of congenital ocular anomalies: Lessons from a national study of congenital cataract in the UK. Invest Ophthalmol Vis Sci. 2001; 42:1444-8.
- 22. Rahi JS, Dezateux C. Congenital and infantile cataract in the United Kingdom: Underlying or associated factors. British Congenital Cataract Interest Group. Invest Ophthalmol Vis Sci. 2000;41:2108-14.
- 23. Mwende J, Bronsard A, Mosha M, Bowman R, Geneau R, Courtright P. Delay in presentation to hospital for surgery for congenital and developmental cataract in Tanzania. Br J Ophthalmol. 2005;89:1478–82.
- 24. Wilson ME, Pandev 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.