Requirements for optical services in children with microphthalmos, coloboma and microcornea in Southern India

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CLINICAL STUDY

Abstract

Purpose **The aim of the study was (1) to determine the need for spectacles in children in Southern India with coloboma, microphthalmos and microcornea, (2) to describe their refractive errors and (3) to assess their needs for low vision aids (LVAs).**
Methods **Children with congenital eye anomalies were recruited from special education for the blind, schools for the mentally handicapped, community-based rehabilitation programmes and hospital records in Andhra Pradesh, India. All those with at least light perception vision (PL) in one eye and who had navigational vision were refracted. Those whose distance vision in their better eye improved with refraction were prescribed spectacles. Those unable to read N10 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.**
Results **Ninety-nine children with coloboma, microcornea or microphthalmos had functional vision. Eight unilateral cases were excluded. Ninety-one bilateral cases were refracted and assessed for LVAs. The vision in 52 children (57%) improved in the better-seeing eye by 1 or more lines of Snellen acuity with spectacles. Spectacles were prescribed most frequently for myopia associated with choroidal coloboma. After refraction, all 19 children with a visual acuity of 6/18 or better could read N10, and 43 of the 72 children (60%) with a visual acuity of <6/18 to PL with functional vision could read N10 unaided, or with distance correction. A further 6 (8%) reached this level with magnifiers. Thirteen children (18%) were given telescopes.**
Conclusion **Children with congenital anomalies of the eye and functional vision benefit from refraction and low vision services.**

Key words Child, Coloboma, Microphthalmos, Refraction, Low vision aid

Data from blind school studies suggest that microphthalmos and coloboma are important causes of severe visual loss in children, particularly in India and Sri Lanka.[1,2] A community-based rehabilitation programme as part of a comprehensive eye care (CBR/CEC) in a rural population in West Godavari district of Coastal Andhra Pradesh, India, found 25% of visual loss in children under 16 years was caused by congenital anomalies of the globe.[3]

Congenital anomalies of the eye are localised structural defects resulting from disruption of embryonic eye development. They occur in all races and can cause major visual disability. There are a wide range of congenital anomalies which may affect the whole eye (as in microphthalmia) or specific eye structures (e.g. congenital cataract, Peter's anomaly). However, the present study includes only coloboma, microphthalmos and microcornea, which form a spectrum of disorders (of which anophthalmia is an extreme form) and have many possible causes, both genetic and environmental.[4] They are a heterogeneous group, can be unilateral or bilateral, and asymmetry is very common. Most children with these anomalies present in infancy and many have strabismus and nystagmus.

There is no universally agreed definition of microphthalmos. Recently several investigators have adopted a definition for microphthalmos using an axial length below the 5th percentile, adjusted for age.[5,6] Warburg[4] described a phenotypic classification for microphthalmos, which does not include axial length and not all categories are mutually exclusive. In the past microcornea has been used as a marker for microphthalmos as they are often, but not invariably, associated. The normal range for horizontal corneal diameter at birth is 9.8 ±

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0.33 mm in boys, 10.1 ± 0.33 mm for girls, and by the age of 7 years the adult value of 11.7 mm is reached.[7] Microcornea is said to be present if the corneal diameter is less than 10 mm (or less than 9 mm in the newborn).[8]

For the purposes of this study a case was defined as any child with microphthalmos, microcornea or coloboma in either eye. Microcornea was defined as a horizontal corneal diameter of 10 mm or less, measured with corneal callipers. Microphthalmos was diagnosed on the basis of axial length measured by B-scan ultrasound (where possible) as an axial length < 2 standard deviations below the mean adjusted for age (i.e. < 21 mm for those over 40 months).[5,6] A coloboma was defined as a developmental defect of any uveal tissue in a typical site consistent with abnormal closure of the embryonic fissure and present since birth.[9] Eyes with coloboma were classified as whether this was associated with microcornea and/or microphthalmos. As the majority of children with these eye anomalies do not have potential for binocular vision the need for spectacles was defined as improvement of at least 1 line of Snellen acuity in the better eye.

Low vision has traditionally been defined by the World Health Organization (WHO) as a corrected visual acuity of < 6/18–3/60 in the better eye. However, individuals who have a corrected visual acuity in the better eye of less than 3/60 may have useful residual vision and benefit from low vision services.[10] A revised 'working definition' was recently agreed, which includes people with visual acuities of less than 6/18 down to and including light perception, provided they have functional vision for navigation.[11] Near vision, also not part of the standard WHO definition of blindness, is included in the revised definition. Isolated case reports have shown that some children with microphthalmos and coloboma affecting the macula have surprisingly good navigational and near vision,[12] but there are currently no data on refraction or low vision assessments in a large series of children with microphthalmos and coloboma. As options for prevention or treatment for children with these conditions are limited it is important to know how many can be helped by spectacles and low vision aids.

The WHO definition of childhood is up to 16 years of age, but as older students were examined in the schools and all the eye conditions were congenital, individuals up to the age of 20 years have been included in the present study.

The aims of the study were (1) to determine the need for spectacles in children with coloboma, microphthalmos and microcornea who had functional vision, (2) to describe their refractive errors and (3) to assess their needs for low vision aids (LVAs) for near and for distance.

Materials and methods

Cases were recruited from six special schools for the blind, three integrated schools, three schools for the mentally handicapped, from hospital records and from a community-based rehabilitation programme as part of a comprehensive eye care (CBR/CEC). The study, which was undertaken in the State of Andhra Pradesh in Southern India, took place between January 1998 and January 1999. Every child was examined by one ophthalmologist using standard methods, and refractions and low vision assessments were performed by one optometrist who had received appropriate training at the low vision clinic in L.V.P. Eye Institute, Hyderabad. Data were recorded on a standard proforma, and entered into a database in Epi-Info 6.04.

Distance visual acuity was assessed using a Snellen E illiterate acuity chart. If the child was unable to cooperate the ability to fix and follow a light was assessed. Near vision was measured using a chart with logMar discontinuous text in English. If unfamiliar with English letters the child was asked to copy the shape of a particular letter. N10 was chosen as the cut-off for low visual aid assessment as this is the size of print used in books in primary schools in India. Functional vision was assessed by determining whether the child could navigate by vision alone around two chairs placed 1 m apart in a well-lit classroom.

Where appropriate every child was refracted under cycloplegia. The visual acuity was rechecked on a different day and the child assigned a WHO category according to the best corrected visual acuity in the better eye.

Children with visual acuity greater than light perception and who had functional vision were assessed for LVAs for near. The exceptions to this were those who were mentally handicapped and those who were able to read N10 easily unaided. A trial set of stand magnifiers of increasing strength was used. The LVA assessments were frequently performed on a different day from the rest of the examination when the effects of cycloplegia had worn off.

Children were assigned to a phenotypic category based on examination of the eye, in conjunction with B-scan ultrasound to measure the axial length. The horizontal corneal diameter was measured using callipers. In the present study only those children who had functional vision were included.

Results

A total of 168 children with congenital eye anomalies were recruited to a larger clinical and epidemiological study. For the present study, those with very poor distance visual acuity who did not have functional vision were excluded (n = 51), as were those who could not be formally tested (n = 18), and those with unilateral anomalies (n= 8) (Fig. 1). Ninety-one children with bilateral pathology (i.e. coloboma, microphthalmos and/or microcornea) with functional vision were included. There were 49 males (54%) and 42 females (46%) with ages ranging from 4 to 18 years (mean 11.9 years). Before refraction 10 of these children (11%) had a distance visual acuity of 6/18 or better, and 81 (89%) had a visual acuity

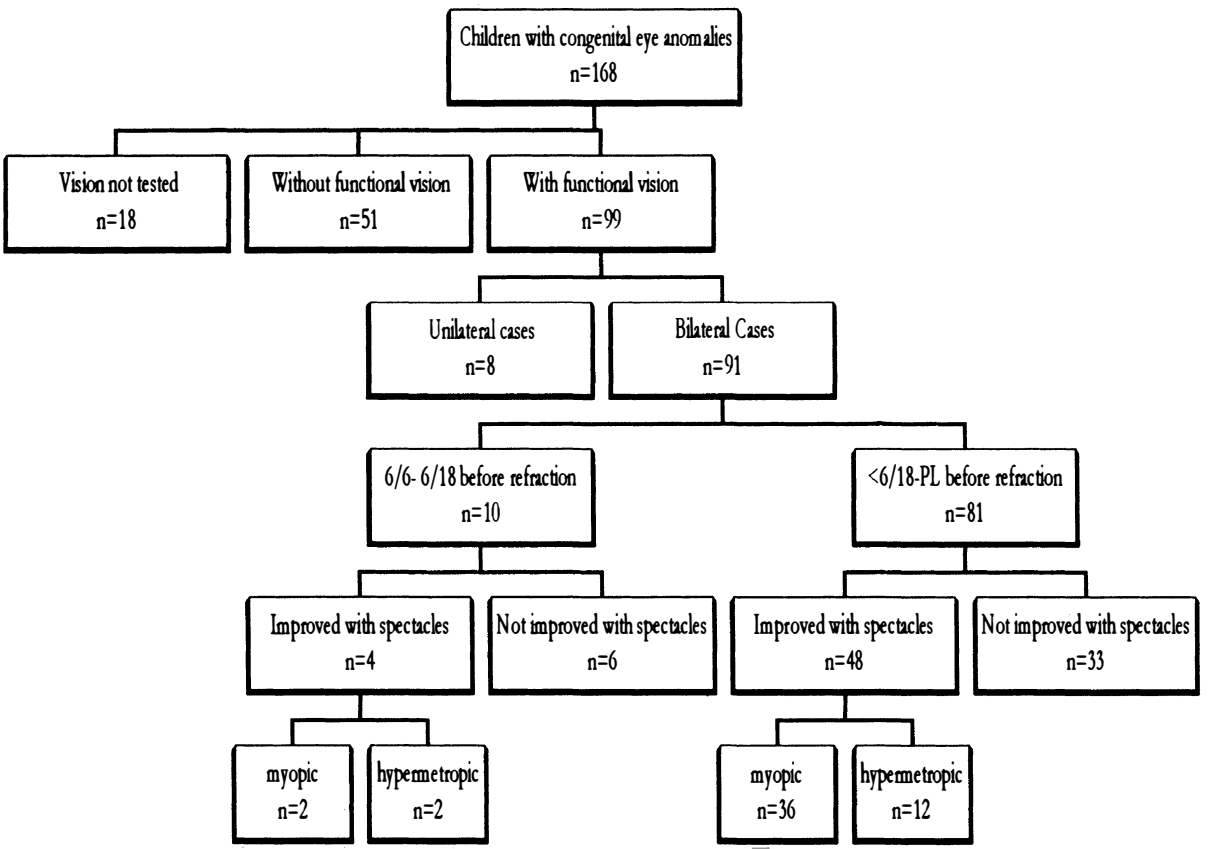

Fig. 1. *Recruitment of cases and the need for spectacles.*

of less than 6/18 down to and including light perception (PL) (i.e. low vision according to the revised working definition).

The need for spectacles

After refraction 19 children achieved a distance visual acuity of 6/18 or better, and 72 children had visual acuity of < 6/18 to PL, i.e. fulfilled the revised definition of functional low vision. Nine children moved from the functional low vision category to the no impairment group after refraction. Fifty-two children were prescribed spectacles (58%).

Distance visual acuities before and after refraction, grouped by WHO categories of visual impairment, are shown in Table 1. Thirty-three (36%) children improved by at least one WHO category after refraction. If the 10 who were in the no impairment category before refraction are excluded, the proportion of children who improved by a WHO category after refraction was 41%.

Types of refractive error

The refractive error in the better seeing eye was myopic in 38 of the 52 children needing spectacles (73%) and hypermetropic in 14 (27%) (Fig. 1). The mean spherical equivalent in the better eye was −2.0 D (range −14 D to +16 D ± 6 D). In children needing a myopic correction the mean spherical equivalent was −5.0 D ± 3.4 D, and in those needing a hypermetropic correction the mean spherical equivalent was +6.2 Ds ± 4.6 D.

The refractive error varied according to the phenotype of the better eye. Eyes with colobomatous microphthalmos were frequently hypermetropic, whereas those with coloboma and microcornea without microphthalmos, and those with simple coloboma, were all myopic except one. The number of children with each phenotype and the mean, standard deviation and range of refractive errors by phenotype are shown in Table 2.

The need for low vision aids

After refraction all 19 children with a corrected visual acuity of 6/6–6/18 could read N10 unaided or with distance spectacles and were not assessed for LVAs (Fig. 2). Of the 72 children with corrected acuities of < 6/18 to PL with 'functional low vision', 43 (60%) could read N10 unaided or with distance spectacles and 29 (40%) could not. Those who could not read N10 were assessed for LVAs for near. Six children (8%) improved to at least N10 with magnifiers. Three magnifiers (+8 D, +10 D and +30 D) were given to children

Table 1. *The WHO categories of distance visual acuity before and after refraction in 91 children with bilateral congenital eye anomalies with functional vision*

WHO category	Level of visual acuity in better eye	Before refraction		After refraction	
		n	%	*n*	%
No impairment	≥6/18	10	11	19	21
Visual impairment	<6/18–6/60	21	23	31	34
Severe visual impairment	<6/60–3/60	28	31	18	20
Blindness	<3/60–PL	32	35	23	25
Total		91	100	91	100

Table 2. *Type of refractive error in the better eye by phenotype for those children prescribed spectacles*

Phenotype	*n*	Myopic	Hyperopic	Mean spherical equivalent	Standard deviation	Range of spherical equivalent (D)
Coloboma + microphthalmos	10	2	8	2.9	4.9	−8 to +9
Microcornea + coloboma	26	26	0	−5.3	3.7	−14 to −0.75
Simple coloboma	8	7	1	−2.3	3.7	−5.5 to +4.5
Isolated microcornea	4	2	2	−2.4	5.2	−7.5 to +3.25
Other	4	1	3	3.8	5.6	−6.5 to +16
Total	52	38	14	−2.0	6.2	−14 to +16

Other = simple microphthalmos (1), microphthalmos and aphakia (1), microphthalmos and retinal scarring (1), microcornea and aphakia but not microphthalmos (1).

with microcornea and coloboma (without microphthalmos). Two magnifiers (+60 D and +17 D) were given to children with colobomatous microphthalmos. A +28 D magnifier was given to a child with microphthalmos and persistent hyperplastic primary vitreous. The lowest distance visual acuity of a child prescribed a LVA was 1/60.

Thirteen children (18% of the low vision group) were given hand-held monocular telescopes (× 4) to aid their distance vision. These children had the following phenotypes in the better eye: microcornea and coloboma without microphthalmos (9 children), colobomatous microphthalmos (2 children), simple coloboma (1 child) and microphthalmos with anterior chamber malformation (1 child). After refraction the visual acuities of this group were 3/60 in 6 children, 6/60 in 3, 6/36 in 3, 6/24 in 1 child. Distance visual acuities improved with the telescopes in 6/6 to 6/24.

Discussion

In this study attempts were made to recruit all children with congenital anomalies of the eye from the population living in two districts of Andhra Pradesh. However, due to the methods of ascertainment the sample of children included in this study may not represent all children with these anomalies in the population, tending to be biased in favour of those with more severe visual impairment.

Refraction techniques

Retinoscopy in children with colobomas and/or microphthalmos is difficult as they frequently have nystagmus, strabismus and eccentric fixation. It is important to refract these children along their habitual fixating gaze without correcting their abnormal head posture. Full-aperture trial lenses are recommended as conventional reduced-aperture lenses can restrict the patient's field of gaze, already limited by a coloboma. In the presence of a choroidal coloboma the retinoscopic reflex appears white and since many of these eyes have high refractive errors in addition to being small, the retinoscopic reflex is difficult to appreciate. For this reason it is recommended that the refractionist begin with a high convex or concave lens. In cases where the retinoscopic reflex is difficult to interpret, subjective refraction is worthwhile.

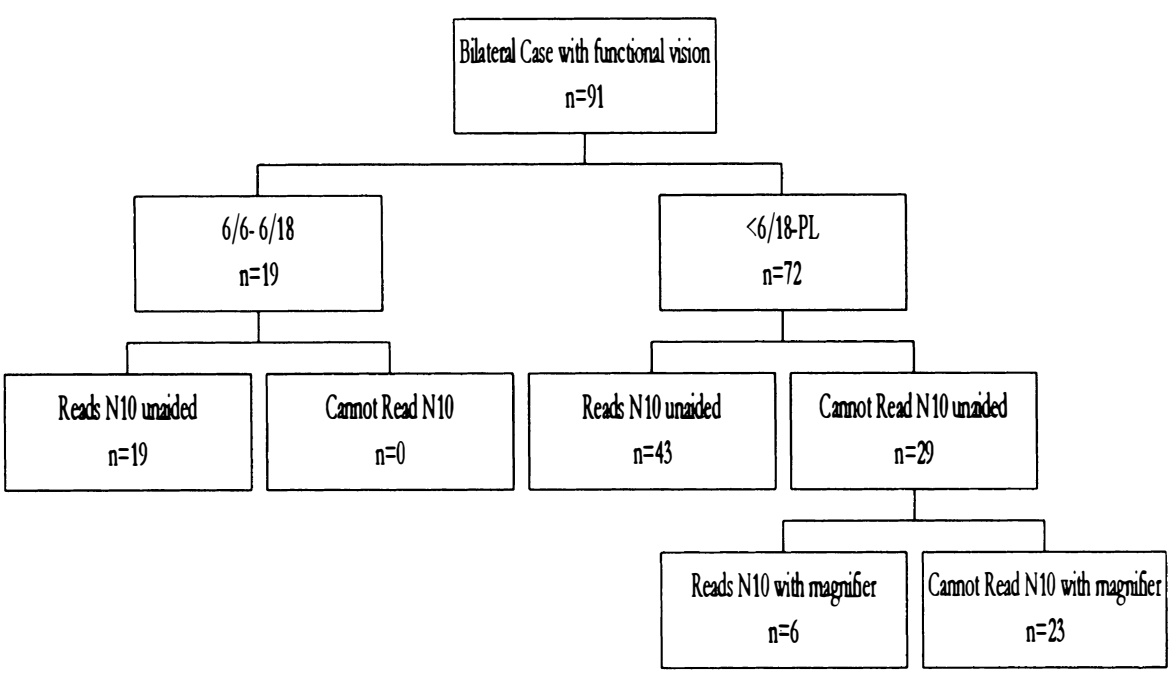

Fig. 2. *The need for low vision aids for near (visual acuity in the better eye after refraction).*

The need for spectacles

After refraction the distance visual acuity improved by at least 1 line of Snellen acuity in the better seeing eye in 57% of children, and in 41% improvement was by at least one WHO category. Eyes with colobomata were found to benefit more frequently from spectacles than other phenotypes, and myopia was the most common refractive error. The type of refractive error varied with the phenotype of the better eye. Eyes with colobomatous microphthalmos tended to be hypermetropic (mean spherical equivalent +2.9 D) whereas those with coloboma and microcornea without microphthalmos tended to be myopic (mean spherical equivalent − 5.3 D). Unfortunately a high myopic correction makes the appearance of microcornea worse. Eyes with simple coloboma also tended to be myopic (mean spherical equivalent −2.3 D) but less so than when coloboma was associated with microcornea. With isolated microcornea refractive errors were equally divided between hypermetropic and myopic errors, but numbers were small.

Low vision assessment

The prescription of low vision aids depends on the assessment of many factors involving the child, their motivation and educational requirements, the environment and, in the case of children, the awareness and motivation of the parents and carers. Fewer LVAs were prescribed in the present study than probably would be the case in industrialised countries where educational and home circumstances might be more favourable. As in other studies, LVAs were found not to be of benefit to children without form vision (i.e. less than 1/60).

Low vision aids

The most frequently prescribed LVA was a monocular hand-held ×4 telescope for distance vision (n = 13, 18%). A ×4 telescope was chosen as it offers a reasonable compromise between magnification and field of view. The criteria for dispensing telescopes in the present study were good manual dexterity; adequate intellectual skills; impossibility of making environmental modifications in the classroom to manage blackboard work; no peripheral field restriction; starting visual acuity not less than 3/60; acceptability to parents and child. Only a minority of children were prescribed LVAs for near vision, and stand magnifiers were the type most commonly prescribed. Children needed time to become familiar with the use of LVAs and this required several training sessions in the classroom.[13]

Non-optical interventions

Since the majority of children with colobomas were myopic they had good near visual acuity but were limited by a short working distance. Non-optical interventions, such as increased illumination, reading stands, reduced working distance, large print and high-contrast reading material,[14] were found to be more useful for these children than magnifiers. Reassurance that a short working distance is not harmful was useful for teachers and parents, and advice was given that the child should sit as near to the blackboard as possible, preferably near a window or with a reading lamp.

Affordability of low vision aids

All LVAs for the present study were provided free of cost. Spectacles and magnifiers up to +24 D are available at relatively low cost in India, but are still beyond the means of many families.[15] Telescopes and high-power stand magnifiers have to be imported and are expensive. Non-optical interventions which are cheaper and can be made locally have received little attention to date in many developing countries. In this study these approaches offered the most practical benefit to children with microphthalmos and coloboma.

Conclusions

All children with microphthalmos, microcornea or coloboma who can use vision for navigation should be refracted, as a high proportion benefit from spectacle correction. The present study suggests that children with a distance visual acuity of < 6/18 to 1/60 should also be assessed for low vision aids. Non-optical interventions are of particular benefit to children with these anomalies, as they often have reasonably good near vision, allowing them access to print and hence better educational and employment opportunities.

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