

Causes of Childhood Blindness among Students of Blinds' School in Shiraz, Iran

M. Mehdizadeh, M. Afarid, A. Attarzadeh

Abstract

Background: Causes of blindness in children vary according to the region and socioeconomic development. Within a given country these causes vary with passage of time. This reflects different levels of socioeconomic development and provision of healthcare services. This cross-sectional study was undertaken to estimate the major causes of severe visual impairment in children and specially preventable and curable blindness in southern Iran.

Methods: 145 students of Shiraz Blinds' School, Shiraz, Iran, interviewed, examined and their medical documents reviewed. Causes of visual loss were analyzed according to involved anatomic area.

Results: 145 students (290 eyes) were examined (53.1% girls and 46.9% boys), among which 42.1% had vision of no light perception. Eye dysgenesis was found in 21.4%, glaucoma in 15.9%, lens associated visual loss in 6.2%, optic nerve lesions in 1.4%, retinal problems in 54.48% and corneal problems in less than 1%. Sixty five students had retinitis pigmentosa, which was the single commonest cause of visual loss.

Conclusion: The major cause of childhood blindness in this region is retinitis pigmentosa, followed by microphthalmia, eye dysgenesis and congenital glaucoma. Large proportion of these blind children had a positive history of consanguineous marriage in their parents and this seemed to be the most probable cause of increased frequency of genetic causes of childhood blindness.

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Keywords • Childhood Blindness • retinitis pigmentosa • consanguinity

Introduction

Blindness is defined by World Health Organization as visual acuity in the better eye of less than 20/400 (3/60).¹ Approximately 1.5 million children are blind worldwide and about 48% of them live in Asia and 24% in Africa. It is estimated that up to 400,000 children in Africa, 270,000 in India, and 200,000 in China are blind. About half of the world cases of avoidable pediatric blindness occur in India and Africa.² The three main causes of pediatric blindness are retinal diseases (29%), corneal scarring (21%), and disorders of entire globe (14%). The prevalence of childhood blindness varies from estimates of 100 per 100,000 in Kenya to 9 per

Department of Ophthalmology,
Shiraz University of Medical Sciences,
Shiraz, Iran.

Correspondence:

Morteza Mehdizadeh MD,
Department of Ophthalmology,
Shiraz University of Medical Sciences,
Shiraz, Iran.

Tel/Fax: +98 711 6279373

E-mail: mehdizadeh@sums.ac.ir

100,000 in United Kingdom and the United States.²

Although blindness is more frequent among adults worldwide, the total number of years of blindness in all blind children is reflected in the total number of years of blindness in all adults.^{3,4} Unlike most adults, blind children often have other disabilities too. It is some what comforting to know that about half of all blind children can have their vision, including the ability to read normal-sized print, improved with proper optical means.^{3,4}

Several studies have been conducted in different countries about the causes of childhood blindness and resulted in various main causes, so there is a need to identify the causes of childhood blindness in our area. To decrease the incidence of pediatric blindness in Iran the causes of blindness in children are investigated.

Patients and methods

This study was performed on all students at Shourideh Educational Center located in Shiraz, Iran during 2002-2003. Shourideh Educational Center is the only educational center for blinds in South of Iran, and has students at different levels including preschool, elementary school, junior high school and high school.

The study was performed in two stages. In the first stage, participants demographic characteristics and the history of their blindness including age, sex as well as the history of surgery, eye trauma, prematurity, familial comorbidity were taken from their health profiles stored in the center. In the second stage, examination of the eyes of the participant was carried out using WHO eye examination record for children with blindness and low vision.¹

Monocular and binocular visual acuity (VA) was measured using the VA Snellen's chart and if VA was less than 3/60, each eye was tested for ability to perceive light. Pinhole VA was assessed and those cases who improved by pinhole were refracted. Anterior segment examination was done with slit lamp. The pupils were dilated, and direct and indirect ophthalmoscopy were done except where inappropriate (for example large central corneal opacity and phthisis bulbi). The results were expressed as descriptive study regarding the causes of loss of vision.

Results

Total of 145 students including 77 girls (53.1%) and 68 boys (46.9%) participated in the study with mean age of 11.8±1.0 years (range 6-14 years). 42.2% of the students (n=61) were total blind with vision of no light perception (NLP). The causes of blindness in each individual

were similar in two eyes. One student had good vision with VA of 6/10 which was inappropriately admitted to this school and was excluded from the study group. The distribution of visual acuities is summarized in Table 1.

Table1: Categories of visual acuity (VA) among students according to WHO criteria.

No eyes (%)	Level of VA
4.83 (14)	6/18-6/60
17.24 (50)	6/60-3/60
35.86 (104)	3/60-LP
42.06 (122)	NLP

LP= Light perception, NLP= No light perception

Among all the eyes which examined, 130 eyes (44.8%) had retinitis pigmentosa, which was the commonest single cause of visual loss. 62 eyes (21.4%) had whole globe lesion. Glaucoma/buphthalmus accounted for 46 eyes (15.86%). Two eyes (<1%) had corneal opacity and scarring accounted of visual loss. 18 eyes (6.2%) had visual loss due to cataract and 158 eyes (54.5%) had retinal problems. Four eyes (1.4%) had optic nerve atrophy and 16 (1.4%) had retinopathy of prematurity. The causes of visual loss are summarized in Table 2.

Table2: The causes of visual loss among students.

Disease	Sex		no. of eyes (%)
	F	M	
RP&MD	36	29	130 (44.8)
Gl & B	11	12	46 ((15.9)
Cat	6	3	18 (6.2)
M & ED	13	15	56 (19.3)
ROP	4	4	16 (5.5)
Alb	3	0	6 (2.1)
H & RD	2	0	4 (1.4)
ONA	1	1	4 (1.4)
PB ?	0	3	6 (2.1)
Rb	0	1	2 (0.7)
SJS	1	0	2 (0.7)

RP & MD= Retinitis pigmentosa & Macular dystrophy; Gl & B= Congenital glaucoma& buphthalmus; Cat= Congenital cataract; M & ED= Microphthalmia & eye dysgenesis; ROP= Retinopathy of prematurity; Alb= Albinism; H & RD= Hyaloidoretinopathy & Retinal detachment; ONA= Optic Nerve Atrophy; PB= Phthisis Bulbi (unknown cause);

Some systemic diseases were also found in these blind children. Deafness were found in two males, diabetes mellitus in two males, convulsion in two males, skeletal anomaly in two females and facial anomaly in one male and one female. 47.5% of the children had the history of parental consanguinity which was evidenced by first cousins marriage and also 58% of parents of children with retinitis pigmentosa had the history of consanguineous marriage.

Discussion

Causes of childhood blindness vary greatly throughout the world. The differences in the causes of blindness between developed and developing countries are stark.¹ Childhood blindness in developed countries usually arise from problems such as retinopathy of prematurity, genetic diseases like cataract, retinal dystrophies, nystagmus, *etc.*² In developing countries the major causes of blindness include corneal scarring resulting from vitamin A deficiency, often is associated with measles, and other illnesses such as trachoma, cataract and ophthalmia neonatorum.⁵ Countries such as India have etiological encompassing parts of both lists.⁶

In our study retinopathy of prematurity (ROP) was the cause of visual loss in eight eyes (4 patients). It is interesting to mention that three patients were less than eight years old, so one might predict a trend toward more ROP in future. Surveys conducted in Africa have indicated a zero ROP frequency because few low-birth weight neonates survive.⁷ The prevalence of ROP has increased as African countries more developed, because they have got the technology to save the live of low birth weight premature infants.⁸ In rural areas of the Philippine, 49% of blindness was found to be due to corneal disease caused by vitamin A deficiency combined with infections such as measles.⁹ In urban Manila, the same study revealed that ROP was still the major causes of blindness.⁹ In developed countries, such as Sweden, the incidence of blindness from ROP has declined due to better screening and treatment methods.¹⁰

In the present study, the most common causes of childhood blindness were retinitis pigmentosa, hereditary retinal dystrophy, cataract, glaucoma and congenital malformations (microphthalmia and eye dysgenesis). About 47.5% of parents of these children had positive history of consanguineous marriage. The most frequency of consanguinity was related to retinitis pigmentosa, congenital glaucoma and eye dysgenesis and microphthalmia. The parents of about 58% of children with retinitis pigmentosa had history of consanguinity marriage and an avoidance of consanguineous marriage may reduce the number of blindness caused by retinitis pigmentosa.

The practice of consanguineous marriages between first cousins has increased the frequency of genetic causes of blindness.¹ In Sri Lanka, where about 25% of marriages are consanguineous, hereditary diseases such as retinal dystrophies Leber's amaurosis and Usher syndrome, cataract, optic atrophy and

microphthalmia account for at least 35% of pediatric blindness.¹¹ Similarly, in the West Bank and Gaza, 62% of childhood blindness seems to due to consanguinity and a positive family history was found in 57% of such cases.¹² The familial causes of blindness were reported to be Leber's amaurosis, cone dystrophy, retinitis pigments, and albinism in the latter study.¹²

Regarding the scarring corneal causes of blindness except for three cases of undetermined cause we could not find any infection or nutritional source for corneal blindness. So our result is not in accord with the reports from other developing countries,^{5,7} which might be due to actual low incidence of scarring corneal causes. We also found a case of good vision with VA of 6/10 in this school-a case of spasmus nutans-who had a previous history of nystagmus and head nudding which had been improved,¹³ So periodic examination may be required for detection of similar cases.

Evaluation and more reliable data collection from a large population of a blind children is cumbersome and very expensive.^{1,2} Most investigations obtained data by surveying local schools for blind children. However, this approach has two major problems; that is only school age children are considered, and in some cultures some of them may not participate to these schools. Despite all these reservations, school surveys have provided most of the available information in the literature.

Conclusion

This study demonstrated that hereditary retinal degeneration is the most common causes of blindness in children of the school for blind in southern Iran. The consanguineous marriage, as found in their family history, is also responsible for the most of this hereditary disease.

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