EXTENDED REPORT

Causes of severe visual impairment and blindness in schools for visually handicapped children in Iran

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Aims: This survey was conducted on children in schools for the blind in Tehran (from 2002 to 2003) to determine the causes of severe visual impairment and blindness and to identify preventable and treatable conditions.

Methods: The study was performed on 362 students at different grades in three schools for the blind. Patient sex, age, family history of blindness or low vision, visual acuity, causes of blindness, and treatable and preventable conditions were studied.

Results: Of the 362 cases, 210 (58%) were boys and 152 (42%) were girls. Mean age was 13.5 (SD 4) years. Severe visual loss was seen in 80.9%. Retinal diseases were the most common cause for low vision (51%); cataract, optic nerve atrophy, corneal and anterior segment diseases, glaucoma, anophthalmia, and globe malformations were other major causes of blindness. Treatable aetiologies and positive family history of blindness were seen in 25.7% and 36% of the patients, respectively. The incidence of preventable diseases, excluding familial disorders, was low.

Conclusion: In addition to the prevention and treatment of some conditions, premarital genetic counselling and family planning control in families with inherited diseases could decrease the number of blind children in the future in Iran.

Childhood blindness is one of the most important priorities in the World Health Organization (WHO) vision 2020¹ programme. There are approximately 1.4 million blind children all over the world, two thirds of whom live in developing countries.²

There are few published studies on the causes of blindness in Iran. The diagnosis of diseases, which affect vision in children, is important because some of these diseases are preventable and some are treatable. The prevalence of diseases leading to blindness is different in various populations, depending on geographical location, culture, race, socioeconomic status, etc.

Although not a major problem in terms of absolute number, childhood blindness accounts for a significant number of years with blindness. In planning to deal with causes of blindness in children in future, public health services have to be strengthened at all levels and collaboration with the government should be improved.

It is essential to study the epidemiological status of diseases in order to decide on important health programmes.

Diseases that lead to blindness are dependent on inheritance and environmental conditions. Some of them are preventable, some others are treatable,^{3–6} and some are untreatable. Currently, the way to deal with such problems is to improve the tactile and hearing senses in patients at low vision schools. This rehabilitation allows a return to normal life; however, if the causes of blindness remain unknown and no programme for their prevention is designed, the current facilities are not enough for such a large population.

Related studies have been performed in many countries.^{3–14} The prevalence of diseases, which affect vision and are unpreventable, is unknown in Iran, but one related study showed that retinal diseases were the most common cause of visual loss.¹⁵ There are treatable diseases that would not lead to blindness if some measures are taken. The pattern of the prevalence of these diseases at a young age would be helped by implementing a health programme and primary treatment.

MATERIALS AND METHODS

This study was carried out via interview and examination in three blind educational centres in Tehran, Iran, from 2002 to 2003.

All students at Narjes educational centre and half of the students at Mohebbi educational centre (low vision schools) were examined at the eye clinic of the Labbafinejad Medical Centre. The rest of the students, together with students from Khazaeli, were examined at their schools. History taking and eye examination including visual acuity (VA), refraction if possible, anterior segment examination by biomicroscope, fundus examination by indirect ophthalmoscope, and IOP measurement were performed. Data were recorded on special forms, and causes leading to blindness were highlighted regarding the possibility of prevention and treatment. Findings were statistically analysed by software programs. Loss of vision in students was defined according to the blind educational classifications of the special education organisation: visual acuity between 20/200 and 20/80 in the better eye, so that the patient could read large optotypes with maximum correction and could educate at ordinary schools, was defined as mild visual loss. Visual acuity between 10/200 and 20/200 in the better eye, so that the patients could walk but not read large optotypes and should learn the Braille system, was defined as moderate loss of vision. Visual acuity between no light perception and 10/200 in the better eye, so that the patient is absolutely blind, was defined as severe visual loss. These patients must learn the Braille system and other skills to increase individual abilities for orientation and learning.

RESULTS

A total of 362 patients with mean age of 13.5 (SD 4) years (range 5–25 years) were included in this survey, 210 (58%) were male and 152 (42%) were female. Family history of blindness was reported in 131 (36%), which included 38.2%

Abbreviations: FEVR, familial exudative vitreoretinopathy; LCA, Leber's congenital amaurosis

Figure 1 Frequency of mild, moderate, and severe visual loss in 362 children from schools for the blind in Tehran 2002–3.

of the girls and 34.8% of the boys. Figure 1 shows the cause of decreased vision in the studied cases: more than 80% of patients had severe visual loss.

Figure 2 shows the prevalence of causes of blindness. Retinal diseases were the most common cause of blindness, followed by cataract, optic nerve atrophy, and glaucoma. Phthisis bulbi in two, trauma in three, neoplasm in one, and uveal coloboma in six cases had a minimal role in causing blindness (fig 2).

Table 1 indicates the distribution of diseases leading to blindness. Leber's congenital amaurosis (LCA) was the most common disease, present in 160 cases (44.2%) and is untreatable and unpreventable. The most common preventable disease was secondary atrophy of the optic nerve with a prevalence of 5.8%. The most common treatable disorder was congenital cataract, with a prevalence of 13.5%, followed by congenital glaucoma with a prevalence of 7.5%. Retinal detachment, familial exudative vitreoretinopathy (FEVR), anophthalmia, cortical blindness and phthisis bulbi, congenital retinal detachment, Usher syndrome and corneal endothelial dystrophy, Ehler-Danlos syndrome, and retinoblastoma were other causes of blindness.

One hundred eighty five patients (51%) had retinal diseases; the most common disorder in this group was LCA with 160 patients (86.5%).

There are several causes for severe visual impairment in children and some of them are preventable.^{13–14} Secondary atrophy of the optic nerve, congenital toxoplasmosis, and cataract secondary to intrauterine infection were detected in 15 patients and comprised 4.1% of preventable causes of blindness. If inherited diseases are added to acquired diseases

Figure 2 Frequency of causes of blindness in 362 children of schools for the blind in Tehran 2002–3.

more frequency will be obtained; however, since the determination of inherited diseases needs a study with better facilities, this group of diseases was not included in the preventable diseases group. Therefore, most disorders were categorised as unpreventable. Ninety three patients (25.7%) had treatable disorders, which mostly included cataract, glaucoma, anterior segment diseases, secondary atrophy of the optic nerve and, rarely, retinal diseases.

Retinal diseases were the most common untreatable diseases (68% of cases) (table 2).

DISCUSSION

The prevalence of childhood blindness was reported as 0.17% in southern India.¹³ Blindness was caused by treatable refractive errors (33.3%) followed by preventable causes (16.6% the result of vitamin A deficiency and 8.3% the result of amblyopia after cataract surgery). The major remaining causes of blindness were congenital eye anomalies (16.7%) and retinal degeneration (16.7%). The authors concluded that the priorities for action to reduce childhood blindness in India are refractive error, cataract related amblyopia, and corneal diseases. We did not evaluate overall causes of childhood blindness in our population but only the prevalence of causes of blindness in pupils in schools for the blind. Regarding frequency of retinal diseases, LCA, with 160 patients (86.5%), was the most frequent disease; other retinal diseases were less frequent as indicated in table 2. Seventy seven (48.1%) patients with LCA had a positive family history of blindness.

Kello and Gilbert¹⁴ studied causes of severe visual impairment and blindness in Ethiopia and concluded that vitamin A deficiency and measles were the major causes of severe visual impairment/blindness in children in schools for the blind. Most causes are preventable during childhood through provision of basic primary healthcare services. This shows the role of socioeconomic conditions in epidemiology of blindness in underdeveloped countries.

We did not have any proved case of vitamin A deficiency in our series.

Table 1 Frequency of causes of blindness in schools for the blind in Tehran in 2002–3

Diseases	No of cases	%
Leber's congenital amaurosis (LCA)	160	44.2
Congenital cataract	49	13.5
Congenital glaucoma	27	7.5
Secondary optic atrophy	21	5.8
Primary optic atrophy	16	4.4
Peter's anomaly	12	3.3
Anophthalmia	11	3.0
Anterior segment dysgenesis	8	2.2
Microcornea	6	1.7
Uveal coloboma	6	1.7
Rod monochromatism	5	1.4
Congenital toxoplasmosis	5	1.4
Bardet-Biedel syndrome	5	1.4
Sclerocornea	5	1.4
Albinism	4	1.1
Microphthalmia	4	1.1
Trauma	3	0.8
Others	15	4.1
Total	362	100

Table 2 Frequency of retinal diseases among students in Tehran low vision schools 2002–3

Diseases	No of cases	% distribution
LCA	160	86.5
Rod monochromatism	5	2.7
Retinal detachment	2	1.1
Albinism	4	2.2
Congenital toxoplasmosis	5	2.7
FEVR	2	1.1
Congenital retinal detachment	1	0.5
Bardet-Biedel syndrome	5	2.7
Usher syndrome	1	0.5
Total	185	100

About 36.2% of the students had a positive history of blindness in their families and relatives. No significant difference was seen in family history of blindness in both sexes. To our knowledge, this variable has not been studied in other reports to date.

Severe visual loss was the most frequent type. There was no significant difference in loss of vision between boys and girls; in other words, boys and girls were distributed equally in the three groups of visual loss (mild, moderate, and severe).

Retinal disorders, with a frequency of 51.1%, were the most common cause of blindness: LCA is transferred in an autosomal recessive pattern. Amini¹⁵ reported that the most common diseases leading to blindness were retinal dystrophy followed by optic nerve atrophy and cataract.

In 1988, Schwab⁴ performed a study in two schools for the blind in Zimbabwe, which indicated that the cause of blindness in 75% of students was bilateral corneal opacity. In a study carried out in 1992 by Antonowicz,¹⁰ 100 patients aged 3–18 years were studied in a blind centre in a Polish city. The causes of blindness or low vision in order of frequency were as follows: refractive errors, optic nerve atrophy, congenital cataract, retinal and choroidal degeneration, neoplasia, anophthalmia or globe malformations, iris or choroidal coloboma, retinoblastoma, ocular trauma, and congenital glaucoma.

In 1992 a study was performed at six schools for the blind in Ethiopia by Gebriel.¹¹ Causes of blindness were corneal opacity or phthisis bulbi in 70% and cataract in 14%. In another survey conducted in southern Australia by Newland,¹² data from 3520 patients under 6 years of age were collected and the cause of blindness was cataract in 85% of cases. A study carried out by Cook³ indicated that senile cataract and chronic glaucoma were the main causes of blindness.

Because of the low incidence of some diseases in some subgroups, it was not possible to determine the significance of the difference in both sexes; however, the sexual dominancy could be determined in any disease. Preventable causes of blindness were as follows: secondary atrophy of the optic nerve, cortical blindness due to trauma, congenital toxoplasmosis, retinal detachment, and cataract secondary to intrauterine infections.

Forty per cent of preventable causes were seen in males and 60% in females; thus the frequency of preventable causes was different in the two sexes. The frequency of treatable diseases was 25.7%, based on primary aetiology of blindness. Retinal diseases, with 185 patients (68%), are the most common untreatable cause of blindness. Cataract, glaucoma, corneal and anterior segment diseases, secondary atrophy of the optic nerve, and rare retinal diseases were among the treatable diseases. In Gilbert's study,⁶ treatable and preventable disorders in Chile, South Africa, and India were 54%, 70%, and 48%, respectively. The prevalence of treatable and preventable diseases in students from Tehran blind schools was 30.1%, which is lower than in Gilbert's study.⁶ Because

retinal diseases are the most common untreatable diseases, particularly LCA, it seems that despite measures taken to treat and prevent such disorders, a considerable number of children had (about 70%) developed untreatable and unpreventable diseases.

In conclusion, severe loss of vision was the most common type of visual loss in schools for the blind in Tehran. Retinal diseases, mostly Leber's congenital amaurosis, are the most common disorders leading to blindness. These diseases mostly have an autosomal recessive pattern and parents transfer recessive genes to their offspring usually by consanguineous marriages. Other diseases such as congenital cataract, optic nerve atrophy, anterior segment and corneal diseases, and glaucoma had lower frequencies.

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