

# CAUSES OF SEVERE VISUAL IMPAIRMENT AND BLINDNESS AMONG CHILDREN IN VISUALLY HANDICAPPED SCHOOLS IN JORDAN

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## ABSTRACT

**Objectives:** To identify the major causes of severe visual impairment and blindness among children in visually handicapped schools in Amman-Jordan with a view to determine potentially preventable and treatable causes.

**Methods:** This study was conducted in two schools for visually handicapped children in Amman. The medical records of 160 students were reviewed during the period between August and October 2005.

**Results:** A total of one hundred and sixty students (85 males and 75 females, aged 6-18 years) were enrolled in the study. Twenty-eight children (17.5%) had severe visual impairment and 132 (82.5%) were blind. Retinal abnormalities were the most common pathology (41.9%). Glaucoma, primary optic nerve pathology and cataract constituted 18.8%, 13.1% and 11.3% respectively. According to timing of insult, the major cause of visual impairment was hereditary diseases (50%), followed by abnormalities of unknown timing of insult (30%). It was estimated that 71 children (44.4%) suffer from visual impairment caused by potentially preventable or treatable conditions.

**Conclusions:** The major causes of severe visual impairment and blindness among children in visually handicapped schools were glaucoma, primary optic nerve pathology and cataract.

**Key words:** Severe visual impairment, Blindness, Visually handicapped

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## Introduction

The World Health Organization defines severe visual impairment as visual acuity less than 6/60 up to 3/60 in the better eye with the best correction and blindness as visual acuity of less than 3/60 in the better eye with the best correction.<sup>(1)</sup> For a child who is born blind or who becomes blind the total number of years of disability are greater than for a person who becomes blind later in life.<sup>(2)</sup> Currently it is estimated that there are 1.5 million blind children in the world, of whom one million live in Asia.<sup>(3)</sup> The prevalence of blindness in children ranges from

approximately 0.3/1000 children in affluent regions to 1.5/1000 in the poorest communities.<sup>(4)</sup> Childhood blindness had been identified as a priority by the World Health Organization and the International Agency for the Prevention of Blindness. The goal is to eliminate avoidable causes of blindness by year 2020 as stated in "Vision 2020-the right to sight" programme.<sup>(5)</sup> At least half and possibly up to three quarters of childhood blindness are avoidable.<sup>(6)</sup>

In our review of published data the prevalence of blindness among Jordanian children could not be found. The prevalence of visual acuity deficit and

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ocular disorders in Jordanian schools was reported to range from 2% to 4%.<sup>(7-8)</sup> Genetically determined causes were the most common etiological factors in two different generations in Jordan according to Al-Salem *et al.*<sup>(9)</sup> In general, genetic causation of childhood blindness is frequent in developed countries, whereas nutritional and infectious factors are more common causes of childhood blindness in developing countries.<sup>(10)</sup>

The aim of the study was to identify the major causes of severe visual impairment and blindness among children in visually handicapped schools in Amman with a view to determine potentially preventable and treatable causes.

## Methods

A retrospective study was conducted in two schools for visually handicapped children in Amman (Abdallah Ben Maktoom School and Zahran School). Medical records were reviewed for 160 students. Each student's record included a medical report signed from a competent authority showing his best corrected visual acuity and diagnosis. These authorities included either the Royal Medical Services, or Ministry of Health, or Jordan University Hospital. Students' selection for joining these schools followed the World Health Organization criteria for definitions of severe visual impairment and blindness. Causes of severe visual impairment and blindness were classified anatomically and aetiologically according to timing of insult. Avoidable causes were identified.

## Results

A total of 160 students (85 males and 75 females, aged 6-18 years) were included in the study (Table I). There was no significant difference between males and females according to the anatomical site of pathology. Twenty-eight (17.5%) children had severe visual impairment and 132 were blind with five of them having no light perception (Table II). Anatomically, the most common affected site was the retina (41.9%) (Table III). Retinal abnormalities included hereditary diseases (31.3%) such as retinitis pigmentosa and albinism, retinopathy of prematurity (9.4%), and retinoblastoma (1.3%). Optic nerve pathology included optic nerve hypoplasia (6.9%), primary optic atrophy (6.3%),

and secondary optic atrophy due to cortical problem (1.3%). Whole globe was affected in 9.4% (phthisis 5.6% and buphthalmous 3.8%). Corneal pathology included dystrophies (1.9%) and scarring (1.3%). Table IV shows aetiological classification according to timing of insult; the major cause of visual impairment was hereditary disease (50%), followed by abnormalities of unknown timing of insult (30%). Hereditary causes comprised retinal pathology (31.3%), glaucoma/buphthalmous (10%), cataract (either aphakic or pseudophakic or not operated (6.9%), and corneal dystrophy (1.9%). Unknown timing of insult included optic nerve pathology (13.1%), glaucoma (8.8%), cataract (3.1%), phthisis (3.1%), retinoblastoma (1.3%), and uveal coloboma (0.6%). Retinopathy of prematurity (9.4%) and secondary optic atrophy (1.3%) occurred in neonatal period. Postnatal disorders included infantile glaucoma (3.8%), cataract (1.3%), phthisis (either microphthalmia or anophthalmia) (1.3%), uveitis (0.6%), and corneal scarring (0.6%). Intrauterine causes were phthisis (1.3%) and corneal scarring (0.6%). Visual impairment due to potentially preventable or treatable conditions was estimated in 71 (44.4%) children (Table V). These included glaucoma, cataract, retinopathy of prematurity, phthisis, corneal scarring, secondary optic atrophy, and uveitis. The first three causes comprised 88.7% of the preventable or treatable causes.

Retinoblastoma was seen in two children. Both of them were bilateral having severe visual impairment. Secondary optic atrophy, a preventable cause of blindness, was found in two patients. Both occurred in infancy period; one due to head trauma and the other following meningitis. Glaucoma was seen in 36 children (16 were hereditary, 14 with unknown timing and 6 had infantile onset of glaucoma).

Thirty were categorized as glaucoma and six had buphthalmous and were categorized as whole globe affection. All of these cases presented at late stage of disease. Cataract related problems were seen in 18 patients; 11 were hereditary, five with unknown timing, and two occurred in the postnatal period.

Nine patients had phthisis bulbi; five of unknown timing, two related to intrauterine infections, and two to postnatal infections. Five of these children had bilateral no light perception.

**Table I.** Age and sex distribution and its relation to anatomical site of pathology

Age and sex (years)		Whole globe	Cornea	Lens	Uvea	Retina	Glaucoma	Optic nerve	Total
6-8	Male	2	1	2	0	6	2	2	15
	Female	1	1	3	0	3	2	0	10
8-10	Male	1	1	1	0	4	4	3	14
	Female	1	0	1	0	6	2	2	12
10-12	Male	2	1	2	1	9	3	0	18
	Female	2	0	0	0	6	3	2	13
12-14	Male	1	0	2	0	5	3	1	12
	Female	1	0	1	0	5	2	2	11
14-16	Male	1	0	3	0	5	2	3	14
	Female	2	1	1	1	6	2	2	15
16-18	Male	1	0	0	0	7	1	3	12
	Female	0	0	2	0	5	4	3	14
<b>Totals</b>	Male	8	3	10	1	36	15	12	85
	Female	7	2	8	1	31	15	11	75

**Table II.** Distribution of visual acuity in the better eye

WHO category	Visual acuity	Number	Percentage
Blind	No PL*	5	3.1
Blind	3/60-PL	127	79.4
Severe visual impairment	<6/60 to 3/60	28	17.5

\* Perception of light

**Table III.** Causes of severe visual impairment and blindness according to anatomical site

Site	Number	Percentage
Whole globe	15	9.4
Cornea	5	3.1
Lens	18	11.3
Uvea	2	1.3
Retina	67	41.9
Glaucoma	30	18.8
Optic nerve/neurological	23	14.4
Total	160	100

**Table IV.** Aetiological classification according to timing of insult

Aetiology	Number	Percentage
Hereditary disease	80	50
Intrauterine	3	1.9
Perinatal/neonatal	17	10.6
Unknown timing	48	30
Postnatal/infancy/childhood	12	7.5
Total	160	100

**Table V.** Preventable/treatable causes of blindness

Cause	Number	Percentage
Glaucoma	30	42.3
Cataract	18	25.4
ROP*	15	21.1
Phthisis	3	4.2
CNS problem	2	2.8
Corneal scar	2	2.8
Uveitis	1	1.4
Total	71	100

\* Retinopathy of prematurity

## Discussion

The study was conducted in two schools for the blind in Amman. Blind school studies are relatively inexpensive and provide an indication of relative importance of the different causes of blindness.<sup>(6)</sup>

Hereditary disease is a major cause of severe visual impairment and blindness. It is associated with a high rate of consanguineous marriages<sup>(11-12)</sup> and could be prevented at least in part by genetic counselling.<sup>(3)</sup> In industrialised nations, hereditary disease accounts for up to half of cases of childhood blindness.<sup>(2)</sup> In comparison, studies using the same methodology in other countries of Asia found genetic disease to range from 16.8% to 35%.<sup>(13-14)</sup>

A study of 260 Jordanians who became blind or visually impaired before the age of 15 years showed the dominant effects of genetically determined causes in two generations.<sup>(9)</sup> Hereditary causes were found in 50% of the patients in our study, and comprised around three quarters of retinal abnormalities.

The second retinal pathology was retinopathy of prematurity. In urban settings, retinopathy of prematurity is reaching almost epidemic proportions with between a quarter and a half of all childhood blindness.<sup>(15)</sup> There is no national plan for the prevention of retinopathy of prematurity in Jordan. This has to be developed and implemented. It is vitally important that programs for screening and treating babies are established in all units where preterm babies weighing less than 1500 g survive to six weeks and more.<sup>(16)</sup>

Optic nerve pathology comprised optic nerve hypoplasia and optic atrophy. According to timing of insult, optic nerve hypoplasia and primary optic atrophy are considered to have unknown timing of insult.<sup>(17)</sup> Other hypoxic insults can cause loss of vision secondary to maldevelopment involving the geniculostriate pathways such as encephalitis, hydrocephalus, or metabolic derangements.<sup>(18)</sup>

Cataract is a potentially treatable disease. This ought to be avoidable by early detection followed by appropriate surgical techniques and postoperative rehabilitation. Early diagnosis followed by appropriate treatments including good surgery, genetic counselling for the families with hereditary disease would improve the frequency of cases which are potentially treatable such as cataract and congenital glaucoma.<sup>(3)</sup> In a study conducted at King Hussein Medical Centre, congenital glaucoma and cataract were the most common causes of

blindness in children.<sup>(19)</sup> Cataract is also one of the common treatable causes of blindness in adults. In a hospital-based study done at Jordan University Hospital and Princess Basma Teaching Hospital in Jordan, cataract was the second commonest cause of blindness among adult Jordanians.<sup>(20)</sup>

Corneal pathology was seen in five patients: three with hereditary dystrophies, one with corneal scar attributed to intrauterine rubella infection, and one had postnatal infection. In industrialised countries corneal disease is responsible for less than 2% of blindness in children while in the poorest areas of Africa and Asia corneal scarring accounts for 25-50%. The major cause is vitamin A deficiency often precipitated by measles or gastroenteritis in children aged typically six months to four years.<sup>(16)</sup> The low figures in our study could be attributed to appropriate immunization programs and satisfactory vitamin and nutrition supplements.

One of the main priorities of Vision 2020 is elimination of corneal scarring caused by vitamin A deficiency, measles, or ophthalmia neonatorum. This could be achieved through promotion of maternal and child health care, measles immunisation, vitamin A supplementation, and nutrition education.<sup>(3, 5, 21)</sup>

Other risk factors such as maltreatments, parental consanguinity, parental education level, and other socioeconomic factors were not listed in the medical records; this drawback precluded the use of further statistical analysis.

Table VI shows the main causes of blindness in children in different countries. In the Mediterranean countries, genetic abnormalities including retinal degeneration, congenital cataract and glaucoma were the leading causes of blindness. Retinopathy of prematurity is the commonest cause of blindness in many countries in Europe, the Americas and the Caribbean. Whole globe affection was frequently seen in China and India. In Africa, corneal scarring is an important cause for childhood blindness.

Understanding the causes of childhood blindness, particularly the preventable and the treatable causes, is important in decreasing the prevalence of blindness in children. This requires genetic counselling for families with hereditary diseases, early diagnosis and management for potentially treatable conditions such as congenital cataract and glaucoma, establishing retinopathy of prematurity screening and treatment programs, appropriate immunisation programs, promotion of breast

**Table VI.** Main causes of blindness in children in different countries

Country	Causes of blindness		
	First	Second	Third
Argentina <sup>(23)</sup>	ROP*	Retinal dystrophy	Optic nerve pathology
Bolivia <sup>(23)</sup>	Corneal pathology	Retina dystrophy	cataract
Brazil <sup>(23)</sup>	Glaucoma	chorioretinitis	Optic atrophy
Britain <sup>(24)</sup>	Congenital cataract	Cortical impairment	Optic atrophy
Chile <sup>(23)</sup>	Retinal dystrophy	ROP	Optic nerve pathology
China <sup>(2)</sup>	Whole globe	Retinal disorders	Lens abnormalities
Colombia <sup>(23)</sup>	ROP	-	-
Cuba <sup>(23)</sup>	ROP	-	-
Czech <sup>(17)</sup>	ROP	Optic nerve pathology	Retinal dystrophy
Dominican <sup>(23)</sup>	Cataract	Corneal pathology	Glaucoma
Ecuador <sup>(23)</sup>	ROP	-	-
Ethiopia <sup>(25)</sup>	Corneal scar	Phthisis	Optic nerve pathology
Guatemala <sup>(23)</sup>	ROP	-	-
Hungary <sup>(24)</sup>	Congenital cataract	Congenital anomalies	Myopia
India <sup>(6)</sup>	Whole globe	Corneal pathology	Retinal dystrophy
Iran <sup>(26)</sup>	Retinal disorders	Cataract	Optic atrophy
Jamaica <sup>(23)</sup>	Cataract	Optic nerve pathology	Glaucoma
Mediterranean <sup>(10)</sup>	Retinal degeneration	Congenital cataract	Congenital glaucoma
Nigeria <sup>(27)</sup>	Lens abnormalities	Corneal pathology	Whole globe
Paraguay <sup>(23)</sup>	ROP	-	-
Peru <sup>(23)</sup>	Corneal pathology	Glaucoma	Cataract
Scandinavia <sup>(24)</sup>	Cerebral amblyopia	Optic atrophy	Congenital anomalies
Uruguay <sup>(23)</sup>	Cataract	Retinal dystrophy	Optic nerve pathology
USA <sup>(23)</sup>	ROP	Optic atrophy	Retinal dystrophy
Uzbekistan <sup>(24)</sup>	Congenital cataract	Retinal disorders	Microphthalmos

\* Retinopathy of prematurity.

feeding, health and nutrition education, and targeting vitamin A supplementation programs at preschool children in areas where vitamin A deficiency is endemic.<sup>(22)</sup>

## Conclusion

The major causes of severe visual impairment and blindness among children in visually handicapped schools were glaucoma, primary optic nerve pathology and cataract.

## Recommendation

It is recommended to develop a paediatric ophthalmology centre in Jordan to improve the diagnosis and management of potentially treatable conditions. Screening and early detection are the responsibility of the whole community. A national programme has to be developed and implemented in order to prevent childhood blindness.

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