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The causes of visual impairment in children in a school for the blind in Johannesburg

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Abstract

Background: More than 1.4 million children are blind worldwide. The significance lies in the social, emotional and economic implications of childhood visual impairment, which are endured throughout a lifetime of 'blind years'. Information regarding the epidemiology and risk factors relating to childhood visual impairment is essential for the development and implementation of targeted interventions.

Objectives: To identify the causes of childhood blindness in a school for the blind in Johannesburg, South Africa, as a representation of trends in our urban population.

Methods: All learners attending a school for the blind in Johannesburg were evaluated. Information obtained was recorded using the World Health Organization's programme for the prevention of blindness (WHO/PBL) method and reporting form.

Results: A total of 189 learners were examined, of which 110 (59%) had severe visual impairment or blindness. The major affected anatomical sites were the retina (42%), whole globe (16%), optic nerve (10%), cornea (8%), uvea (6%) and lens (5%). Retinopathy of prematurity was the most common retinal condition (n=26, 14%). Hereditary conditions were responsible for 28% of visual impairment in learners. Aetiology was indeterminate in 41% of learners. Avoidable causes accounted for 29% of learners.

Conclusion: There has been a change in the disease pattern of childhood blindness in this study population, when compared to the findings of a nationwide study conducted in 1996. While many advances have been made regarding immunisation, vitamin supplementation and ophthalmic management of paediatric cataracts, the implementation of further measures are still required in order to overcome avoidable causes of childhood visual impairment.

Keywords: ophthalmology, paediatrics, South Africa, prevention, public health

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Introduction

It has been estimated that 1.4 million children are blind globally, three-quarters of whom are from developing countries.[1] The World Health Organization's (WHO) Vision 20/20 – The Right to Sight Programme has highlighted the importance of the management and control of visual impairment in childhood. This is for a number of a reasons: 1) individuals who are born blind or become blind during their childhood endure a lifetime of 'blind years' with their accompanying social, emotional and economic implications; 2) many causes of visual impairment in childhood are preventable or curable, particularly in developing countries where over 30% of childhood blindness has been found to be avoidable; and 3) several of these causes also lead to child mortality and therefore their management is associated with improved child survival.[2]

In order for the implementation of

targeted interventions, more information is needed regarding the epidemiology and risk factors relating to childhood visual impairment.[3] These are affected by a variety of factors including, age, geographic location, level of socio-economic development and health care accessibility.[4] Genetic or hereditary diseases are the main cause of childhood blindness in developed countries.[5] In developing countries, the main aetiologies have been found to be related to infections (congenital rubella or acquired measles infection) or nutritional deficiencies (vitamin A deficiency).[5]

South Africa (SA) has been classified as an upper middle-income economy, the largest in Africa.[6] Despite this, poverty and inequality continue to soar.[6] Since the establishment of democracy, unemployment has been exceptionally high, worsening between 1994 and 2003, thereby altering the population's access to health care.[6] Despite the disparity in access to health care in SA, there have been major improvements in the last 21 years with regard to measles immunisation, vitamin A supplementation, and antenatal and neonatal care.

The most recent data available for the causes of childhood blindness in SA were collected in a nationwide study of blind schools in 1996, which found that 39% of childhood visual impairment was avoidable.[7] Retinal disease featured prominently, with dystrophies and albinism accounting for the majority of cases in this disease spectrum. Retinopathy of prematurity (ROP) was found to be an emerging and preventable aetiology of blindness in white and Indian communities.[7] Twelve per cent of the children included in this study had normal vision or mild visual impairment and the need for possible integration of these children into mainstream schooling was identified.[7]

Objectives

The aim of this study was to identify the causes of childhood blindness in a school for the blind in Johannesburg, South Africa, as a representation of trends in our urban population, in order to inform health policy decision-making.

Methods

Patient selection

This was a cross-sectional study of all 189 learners attending Sibonile School in the south of Johannesburg, South Africa, that caters for the education of blind and partially sighted learners, including those with multiple disabilities.

Table I: Distribution of visual impairment		
WHO category	**Level of vision**	**n=184 (%)**
0: Normal vision or mild impairment	Equal to 6/18 or better	27 (15)
1: Moderate visual impairment	6/18–6/60	47 (26)
2: Severe visual impairment	6/60–3/60	0
3: Blindness	Counting fingers at 1 m	26 (14)
4: Blindness	Light perception	54 (29)
5: Blindness	No light perception	30 (16)

WHO: World Health Organization

Data acquisition

The 189 learners enrolled in the school were examined by an ophthalmic registrar at St John Eye Hospital from 1 September 2017 to 15 September 2017, with assessments confirmed by two ophthalmic consultants.

Each learner was examined in the presence of his or her teacher. Data were extracted from previous medical records, where available. Uni-ocular visual acuity was measured, where possible, using Snellen charts and Sheridan-Gardner test cards. A torch or slit lamp were used to examine the anterior segment. An indirect ophthalmoscope was used for posterior segment evaluation, following mydriasis. B-scan ultrasonography was performed when indicated. Examinations were conducted using the World Health Organization's programme for the prevention of blindness (WHO/PBL) method and reporting form, a standardised assessment tool for reporting of the causes of visual loss.[8]

Visual impairment was documented according to the category of visual impairment, graded 0 to 5, and using the anatomical and aetiological classifications in the form.

Where indicated, learners were scheduled for further assessment, surgery and refraction.

Ethical approval was obtained from the University of the Witwatersrand Human Research Ethics Committee (clearance number M170211) and from the both the Hospital Management and Department of Education.

Results were entered onto an Excel spreadsheet (2011, Microsoft, USA). With the consult of a biostatistician, data analysis was carried out using STATA® (v 12.0). These results were documented and displayed graphically by means of tables.

Results

A total of 189 learners were included in the study. There were 121 (64%) males, and 68 (36%) females, all of black ethnicity. The mean age of the learners was 11.3 years (standard deviation 3.2 years) with a range of 5–20 years. Additional disabilities, including mental retardation, physical handicap and epilepsy were found in 37 (20%) cases.

Categories of visual impairment

The distribution of visual impairment according to the WHO categories are shown in *Table I*. Twenty-seven learners (15%) were found to have mild or no visual impairment (equal to 6/18 or better), 47 (26%) moderate visual impairment (6/18– 6/60) and 110 (59%) learners were blind (less than 3/60). Learners who were classified blind had 3/60 to light perception in 43% (n=80) and no light perception in 16% (n=30). Due to multiple disabilities, the degree of visual impairment could not be assessed in five learners.

Anatomical site of visual abnormality

The anatomical sites of abnormality in the learners are shown in *Table II*.

Table II: Anatomical sites of abnormality resulting in visual impairment	
Site	**n (%)**
Retina	79 (42)
Whole globe	30 (16)
Normal globe	25 (13)
Optic nerve	18 (10)
Cornea	16 (8)
Uvea	12 (6)
Lens	9 (5)

Retinal conditions were the most common cause of visual impairment. Of the 79 (42%) learners with retinal pathologies, 28 (15%) had oculocutaneous albinism (OCA), while ROP and retinal detachment accounted for 14% (n=26) and 3% (n=6) respectively. Seven learners had retinal dystrophies including retinitis pigmentosa (n=3), Bardet-Biedl (n=3)

and Best's disease (n=1). Three learners had a history of retinoblastoma, all of whom had previously undergone bilateral enucleation.

Abnormalities of the whole globe were the second most frequent anatomical site, with glaucoma present in 7% (n=14) and microphthalmos in 5% (n=10). All four learners that had one eye removed had glaucoma as their underlying pathology.

Twenty-five learners had normal ocular examinations and included visual impairment attributed to cortical abnormalities in 9% (n=17), amblyopia in 3% (n=5) and refractive errors in 2% (n=3).

Visual impairment in 12 learners (6%) was due to optic nerve atrophy. The causes of optic atrophy included hydrocephalus in 0.05% (n=1), neoplasm 2% (n=4) (craniopharyngiomas n=2, Burkitt's lymphoma n=1, medulloblastoma n=1) and of indeterminate cause in 4% (n=7). Six learners (3%) had optic nerve hypoplasia.

Corneal scarring accounted for 8% (n=16) of visual impairment. Causative factors identified include measles keratitis in one case and ophthalmia neonatorum in one other. Uveal conditions were found in 12 (6%) learners, seven (4%) of whom had previous uveitis and four (2%) aniridia. Cataract was the current cause for visual impairment in 5% of learners (n=9); however, 17 learners (9%) had already undergone cataract surgery at the time of the study.

Aetiology of visual loss

Fifty-two learners (28%) were found to have visual impairment due to hereditary conditions, with more than half being autosomal recessive in pattern. Perinatal causes (predominantly due to cerebral hypoxia) accounted for 22% (n=42) of visual impairment, while neonatal causes (ROP) accounted for 14% (n=26). Childhood causes were found in 14 learners (7%) and were mostly due to childhood infections and neoplasm. The majority (n=78) of learners were found to have visual impairment secondary to causes that could not be determined (*Table III*).

Table III: Aetiological causes of childhood visual impairment

Aetiology	n (%)
Hereditary	52 (28)
Intrauterine	3 (2)
Perinatal	42 (22)
Postnatal	14 (7)
Indeterminate	78 (41)

Avoidable causes

Visual impairment that would have been amenable to prevention or treatment are shown in *Table IV*. Fifty-five learners (29%) had underlying causes of visual impairment that could have been avoided. This includes 4% (n=7) of learners that had conditions amenable to primary prevention and 25% (n=48) secondary to treatable conditions (secondary prevention).

Table IV: Avoidable causes of childhood visual impairment

Causes	n (%)
Avoidable	55 (29)
• Preventable	7 (4)
○ Congenital rubella	3 (2)
○ Ophthalmia neonatorum	1 (1)
○ Measles keratitis	1 (1)
○ Trauma	2 (1)
• Treatable	48 (25)
○ ROP	26 (14)
○ Cataract	6 (3)
○ Glaucoma	14 (7)

Discussion

Our study suggests a change in disease pattern, when considered in comparison to the findings of O'Sullivan *et al*.[7] We found pathology of the whole globe and retina to be significantly more prevalent in our population, whereas pathologies of the optic nerve and retina had previously predominated. The number of learners with visual impairment due to cortical conditions has also increased. Our population reveals a reduction in visual impairment due to avoidable causes. This applies specifically to visual impairment that could have been prevented.

The male-to-female ratio in this study was 1.8:1. This is similar to other studies conducted within Africa, in countries such as Nigeria, where the male-to-female ratio has been found to be as high as 2:1.[9] This may be as a result of a gender-related bias, whereby the education of male learners may be of a higher priority to a family than if the learners were female.

Visual impairment due to indeterminate causes accounts for a large proportion of the selected population, mirroring previous findings. This could possibly be reduced with extensive genetic testing but would be of limited clinical significance. Learners who were found to have retinal degenerations that may have been caused by genetic mutations were identified and registered with Retina SA.

While the retina remains the most common site of visual impairment, the distribution of retinal pathology, within our study population has changed substantially when compared to the findings of 1996. An overall increase in ROP was found. In 1996 the most common retinal conditions were retinal dystrophies and albinism. ROP was found in 11% of learners, of whom only 1% were black. This can possibly be attributed to the infant mortality rate, which was noted to be up to 54.3/1 000 births in the black population at that time.[7] When considering only black learners in the study by O'Sullivan *et al*.,[7] we have found an increase in visual impairment due to ROP, consistent with trends mirrored in other developing countries in Eastern Europe and Latin America.[10] This is in contrast to highly developed countries in which ROP is now an uncommon cause of visual impairment.[11] The increased rate of ROP in our population is the result of a high rate of preterm birth and improved infant survival (infant mortality rate now 32/1 000 live births[12]), coupled with suboptimal oxygen monitoring in neonatal facilities and poorly established ROP screening protocols. While 85% of learners with ROP had severe visual impairment (equal to 3/60 or less), none of them had gone on to require enucleation. Of note, one-third of learners with ROP were aged between 5 and 9 years, while the remaining two-thirds were 10 years or older. Since the learners included in this study were mainly from urban areas, this may reflect the improvements in and success of ROP prevention, screening and treatment programmes in these areas over the last ten years.

Of 1 911 new-borns screened at St John Eye Hospital by Kana *et al.* from 2013–2015, 1% required treatment.[13] A similar incidence of new-borns with ROP requiring treatment has been reported in high-income countries such as Switzerland.[14] However, according to the experience of the senior staff at St John Eye Hospital, ROP continues to be a major problem in outlying areas in SA, where neonatal high care facilities are available in the absence of an on-site ophthalmology department. Therefore, emphasis needs to be placed on improving oxygen saturation monitoring of neonates receiving supplemental oxygen in these facilities as well as formalisation of a referral protocol into the bigger centres for ROP screening and management.

The reduction in childhood visual

impairment secondary to avoidable causes reflects the successful implementation of large-scale primary health care campaigns, that have resulted in the wide-scale distribution of measles and rubella immunisation through the Expanded Programme on Immunisation in SA (EPI-SA), initiated in 1995, as well as the administration of new-born ocular prophylaxis and vitamin A supplementation.[15] However, due to lack of immunisation stock or of awareness and/or attendance by mothers, the level of immunisation coverage, particularly in rural areas, has been noted to be considerably lower than government targets.[16,17] Further improvements are thus required in order to ensure adequate and consistent provision of basic primary health care in SA.[17]

In this study, oculocutaneous albinism (OCA) was found in 15% of learners (n=28). Of these, 27 learners had mild or no visual impairment. In 1996, 9% of black learners attending blind schools were reported to have OCA, while it was found in only 1% of white learners.[7] This is likely a reflection of social and cultural attitudes towards albinism in the community, founded on myth, superstition and fear.[18] As a result of this, acceptance and integration remain problematic and continued efforts to counter such attitudes, in the form of education and destigmatisation, are still required to allow individuals with OCA to succeed and contribute to their communities.

O' Sullivan *et al.*[7] reported that 12% of the learners attending blind schools in SA had a visual acuity of 6/18 or better in their best eye. This was likely due to poor home social circumstances, in which the board and lodging as well as care and concern provided by schools for the blind may be taken advantage of. Our study revealed that this finding has increased over the last 20 years. This figure remains unacceptably high and calls for major improvement in and regulation of blind school admission assessments as well as dialogue with both teachers and parents as to whether these learners could be integrated into the mainstream schooling system.

This is a relatively small study, in a chosen population and is therefore subject to selection bias. The results obtained from one school have been considered in comparison to another study, previously conducted on a national level, with differing methodology. This therefore limits the validity of the conclusions that can be drawn from such a comparison.

All learners assessed in this study are from an urban area. The pattern of childhood visual impairment within SA may vary according to region, especially in rural areas, and therefore the findings of this study may not reflect the overall trend of childhood visual impairment in SA as a whole.

Advantages of this study include many learners being examined for causes of childhood visual impairment, by few examiners, employing standard methodology.

Conclusion

This study has suggested a change in the disease pattern of childhood blindness, in the population examined, over the last 21 years. Preventable blindness has been reduced due to improved immunisation and vitamin supplementation programmes. ROP remains a major cause of childhood visual impairment. Emphasis should be placed on improving prevention, screening and management programmes in peripheral facilities where its incidence is still high.

Fifteen per cent of learners had normal or mild visual impairment. Their placement in a special needs school is most commonly due to the social stigma surrounding OCA.

Education of the community as well as improved admission processes are required in order to maximise mainstream level education where possible and reduce the burden on the blind school system.

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