

WORLD VIEW

Causes of blindness at the "Wiyata Guna" School for the Blind, Indonesia

R Sitorus, M Preising, B Lorenz

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S RuitSee end of article for
authors' affiliationsCorrespondence to:
Rita S Sitorus, MD,
Department of
Ophthalmology, School of
Medicine, University of
Indonesia, Salemba 6,
Jakarta-10430, Indonesia;
ritass@hotmail.comAccepted for publication
6 February 2003**Aim:** To determine the anatomical site and patterns of cause of blindness in one of the biggest schools for the blind in Indonesia with a view to determining potentially preventable and treatable causes.**Methods:** 165 students in one school for the blind in Bandung, Indonesia, were examined clinically and data reported using the WHO/PBL childhood blindness assessment form.**Results:** Most of the students (96.4%) were blind (BL); 3% were severely visually impaired (SVI). The major causes of SVI/BL in this study were: (1) corneal staphyloma, corneal scar, and phthisis bulbi (mainly attributed to infection) in 29.7%; (2) retinal dystrophies (mainly Leber congenital amaurosis, early onset retinitis pigmentosa) in 20.6%; (3) congenital and familial cataract (13.3%); (4) microphthalmus, anophthalmus (10.9%). The whole globe was the major anatomical site of visual loss (32.7%), followed by the retina (26.0%), cornea (17.6%), lens (13.3%), optic nerve (6.1%), and uvea (4.3%).**Conclusions:** This is a small study in a selected population and the results should be interpreted with caution. This blind school study, adopting the WHO/PBL eye form for data analysing, is the first reported for Indonesia. Hereditary disease and infective causes of blindness are the predominant causes of blindness, accounting for 42.4% and 29.7%, respectively. This pattern of causes is a mixed pattern which lies in an intermediate position between the patterns seen in developing countries and those seen in developed countries. The importance both of preventive public health strategies and of specialist paediatric ophthalmic and optical services in the management of childhood blindness in Indonesia are therefore strongly suggested to cover the problems that exist.

Childhood blindness has been identified as a priority in the World Health Organization's global initiative for the elimination of avoidable blindness by year 2020.¹⁻³

The prevalence and major causes of blindness in children vary widely between countries and over times. Currently, by World Health Organization criteria, it is estimated that there are 1.5 million blind children in the world, of whom 1 million live in Asia, 0.3 million in Africa, 0.1 million in Latin America, and 0.1 million in the rest of the world. The recorded prevalence of SVI/BL in European countries varies from 0.1 to 0.41 per 1000 children.^{4,5}

Indonesia has a population of 206 million, who are spread over 25 provinces (Center of Statistic Office/Biro Pusat Statistik, Indonesia: survey of population, 2000), of which 61 250 199 are under 14 years of age and 82 399 716 are under 19 years of age (Survey of Indonesian Population, 2000)

According to the latest available data of blindness (Indonesian Ministry of Health), in the year 2000 the rate of blindness in Indonesia was 1.5%, and is the highest rate among South East Asian countries; rates in Bangladesh are 1.0%, in India 0.7%, and in Thailand 0.3%. But, there are still no exact data on low vision and blindness in children in Indonesia

The aim of this study was to determine the pattern of the causes of visual loss, with an emphasis on preventable and treatable causes.

PATIENTS AND METHODS

The Wiyata Guna Blind School or Wiyata Guna Social Home for Blind-Indonesia is one of the biggest institutions for the blind in Indonesia. The school caters for both residential (mostly) and day students, who mainly come from east Java.

During the academic year 2000-2, 165 students were examined on the days of the visit by three ophthalmologists (BL, RS, SS). Information was gathered using interviews with students, parents (if possible), school staff, and by consulting

hospital records. A history of the age at onset of visual loss, family history, history of consanguinity, and place of residence (village, town, or city) was taken whenever possible. Binocular and monocular visual acuity were measured. Simple tests of functional vision were used, such as the ability to walk around and the ability to recognise faces and/or objects. Anterior segment examination was performed using a slit lamp or flashlight and magnifying lens. Posterior segment examination was performed by indirect or direct ophthalmoscopy after mydriasis. Photodocumentation was performed whenever possible using a hand held fundus camera (Kowa Genesis, Tokyo, Japan).

To analyse the data including the anatomical and aetiological classification the WHO/PBL eye examination records were adapted.² Included were all students with an onset of blindness during infancy/childhood or before the age of 16 years (determined from medical records or a history from adults with knowledge of the child's past medical history, or by the presence of nystagmus). Non-compliant students were excluded.

Table 1 WHO categories of visual impairment. Distribution of visual acuity

WHO category	Level of visual acuity (better eye)	No	%
Blind	NPL	6	3.7
Blind	<3/60-PL	153	92.7
Severe visual impairment	<6/60 to 3/60	6	3.6
Visual impairment	<6/18 to 6/60	0	0
No impairment	6/18 or better	0	0
Total		165	100.0

NPL = no perception of light; PL = perception of light.

Table 2 Anatomical classification of the causes of visual impairment

Anatomical site	No	%
Whole globe	54	32.7
Retina	43	26.0
Cornea	29	17.6
Lens	22	13.3
Optic nerve/CNS	10	6.1
Uvea	7	4.3
Total	165	100.0

Table 3 Aetiological classification

Aetiology	No	%
Hereditary disease	70	42.4
Postnatal/infancy/childhood	51	30.9
Perinatal/neonatal	3	1.8
Intrauterine	1	0.6
Unknown	40	24.3
Total	165	100.0

A genetic disease was diagnosed if there was either a definite family history of the same condition, or if the condition was the result of a well recognised or proved genetic abnormality in the absence of a family history. Electrodiagnostic tests were not undertaken as facilities were not available.

RESULTS

In all, 167 students participated, two were excluded because of non-compliance; 95 were males and 70 were females; 101 (91.5%) of them were aged 11 years or over, 12 (7.3%) were aged 5–10 years, and two (1.2%) were less than 5 years. The distribution of visual acuity is given in Table 1.

Of the students, 3.6% were severely visually impaired (SVI), and 96.4% were blind (BL), with vision mostly between hand movement and light perception only.

Anatomical cause of visual loss

The anatomical sites of abnormality leading to SVI/BL are shown in Table 2.

The major anatomical site of the causes of visual impairment was the whole globe (32.7%). Phthisis bulbi caused by the infection was responsible for 12.1% of the cases. Other abnormalities in this category were microphthalmos or anophthalmos in 11.5% of the cases, and buphthalmos in 9.1% of the cases.

Retinal causes were responsible for SVI/BL in 43 students (26.0%). The majority were retinal dystrophies accounting for 20.6% (mainly Leber congenital amaurosis or early onset retinitis pigmentosa as the most likely diagnoses based on history and clinical examination). Retinopathy of prematurity (ROP) was diagnosed in three cases (1.8%). Other retinal disorders, including retinal detachment, intraocular tumour, and other retinopathies accounted for the remainder (3.6%).

Corneal disease was seen in 29 cases (17.6%), presenting as corneal staphyloma and corneal scar caused by previous eye infections.

Congenital cataract was the underlying cause in 22 cases (13.3%). Of these, nine cases were familial. Deprivation amblyopia was probably the leading cause of visual impairment in these patients, the result of delayed cataract surgery in all instances.

Aetiology of visual loss

The aetiology of SVI/BL is shown in Table 3. The major aetiological category of visual loss was hereditary (42.4%), followed by postnatal (30.9%), perinatal/neonatal (1.8%) and intrauterine (0.6%); the categories remained unknown or undetermined in 24.3%.

In the unknown aetiology group, 40 of 165 cases had congenital anomalies which had been present since birth, such as microphthalmos, cataract, anterior segment dysgenesis, which could not be definitely classified as being caused by genetic factors or events occurring during the intrauterine period. This was the case, in particular, in students without any detailed information of family history.

In the hereditary disease category we included cases of buphthalmos, Leber congenital amaurosis (LCA), and early onset retinitis pigmentosa (RP), which are usually known to be inherited in an autosomal recessive pattern. In a separate study we identified mutations in the spectrum of genes related to the diseases in some of the patients⁶ (Sitorus *et al*, unpublished data).

Postnatal factors were the second commonest cause of blindness, accounting for 30.9% of the cases. The predominant cause of the postnatal factors were corneal staphyloma, corneal scars, and phthisis bulbi, all attributed to eye infection.

Perinatal factors were identified in only three cases (1.8%), whereas intrauterine factors such as congenital toxoplasmosis, were present in one case only (0.6%).

Parental consanguinity was identified in 11 cases (6.7%); no indication of consanguinity was reported in 92 cases (55.8%); and 62 cases (37.5%) were unknown because of lack of information.

The major causes of SVI/BL in this study were (1) corneal staphyloma, corneal scar, and phthisis bulbi (mainly attributed to infection) in 29.7%; (2) retinal dystrophies (mainly LCA, early onset RP) in 20.6%; (3) congenital or familial cataract in 13.3%, and (4) microphthalmos, anophthalmos in 10.9%. The remainder were buphthalmos/PCG in 9.1%, optic atrophy in 5.5%, anterior segment dysgenesis in 2.4%, others (ROP, congenital toxoplasmosis, intraocular tumour, etc) in 8.4%.

DISCUSSION

Marked differences in the causes of childhood blindness have been reported in the past for different countries worldwide, apparently based on socioeconomic factors. In high income countries, lesions of the optic nerve and higher visual pathways predominate as the causes of blindness,^{1,7,8} while corneal scarring from measles, vitamin A deficiency, the use of harmful traditional eye remedies, and ophthalmia neonatorum are the major causes in low income countries.^{1,9} Other significant causes in all countries are cataract, congenital cataract, and hereditary retinal dystrophies.¹

In our study, almost all students (96.4%) were blind (BL) with vision mostly between hand movement and light perception only. These findings were much poorer compared to other reported studies.^{8,10,11}

Hereditary diseases and postnatal infectious eye diseases were the predominant causes of blindness, accounting for 42.4% and 29.7% respectively. The relative frequencies of genetic disorders and of infectious and nutritional conditions lie in an intermediate position between the patterns seen in developing countries and those seen in developed countries, which is similar to a study reported from India.⁹ This result, however, is in contrast with studies from other Asian countries such as Malaysia,¹² Sri Lanka,¹³ and China,¹⁰ which show a mixed pattern between hereditary and unknown aetiologies (a pattern similar to that in industrialised countries).

Of the total cases in our blind school study, genetic eye disease was predominantly responsible for 70 cases (42.4%) of

Table 4 Predominant causes of blindness attributed to genetic/hereditary disease

	Europe			Eastern Mediterranean		Asia				
	UK	Denmark	Iceland	Saudi Arabia	Cyprus	Srilanka	Thailand and Philippines	China	India	Indonesia
Number with genetic cases (%)	26	35	35	84	80	35	16.8	30.7	22.9	42.4
Retinal dystrophies (%)	43*	48*	0*	32*	39*	56.5*	41.5*	most*, †	79.9*	48.6*
Cataract (%)	0	7	14	34	18	16	16.9	11.8	4.3	13.3
Buphthalmos, PCG (%)	2	2	0	17	6	6	7.3	9	0	9.1
Reference	11	23	23	16	17	13	23	10	9	Present study

*Of the hereditary cases; without * = of the total cases; †no detailed data.
PCG = primary congenital glaucoma.

SVI/BL. In comparison, studies in other Asian countries found genetic eye disease to be responsible for 35.0% of SVI/BL in Sri Lanka,¹³ 16.8% in Thailand/Philippines,¹⁴ 30.7% in China,¹⁰ 23.0% in India,⁹ and 29.5% in Malaysia.¹² This is in contrast with studies reported from developed countries such as the United Kingdom¹¹ and the United States,⁷ where blindness related to perinatal problems including prematurity was the largest aetiological category group of SVI/BL. In addition to ROP,⁷ the leading causes of paediatric blindness in the United States are cortical visual impairment⁷ and optic nerve disorders.^{7, 8} In our study, perinatally related diseases, such as ROP, were identified in only three (1.8%) cases. The low incidence of ROP found in our study is probably the result of the much higher mortality of premature children in Indonesia, in particular in rural areas, compared to developed countries. Most students in the blind schools were born and spent their childhood in rural areas. In the future, ROP is likely to become a much bigger problem in Indonesia, as neonatal care services are bound to expand, particularly in urban areas.

Of the 70 hereditary cases, retinal dystrophies were the predominant causes in 34 (48.6%), followed by buphthalmos/primary congenital glaucoma in 22 (21.4%), and cataract in nine (12.9%). In the retinal dystrophies group, we included conditions where retinal dystrophy was very probably based on history and ophthalmoscopic findings but not confirmed by electrodiagnostic tests because of unavailability. However, the diagnosis was confirmed in many instances by molecular genetics in a separate study by identifying the RPE65, RetGC1, and AIPL1 genes, the genes responsible for LCA and early onset RP (Sitorus *et al*, unpublished data).

Retinal dystrophies were also the most common form of genetic eye diseases (49–80%) in all other countries except Thailand and the Philippines¹⁴ where cataract was the most common cause (43.9%) (Table 4). Autosomal-recessive inheritance was the most common mode of transmission, accounting for 77.1% of the hereditary group in our study, followed by autosomal dominant in 7.1%, X linked in 2.9%, and undetermined in 12.9%. This high proportion of autosomal recessive eye diseases has been attributed to a high level of consanguineous marriage in this specific ethnic group from the west Java region. Unfortunately, we could identify consanguinity in only 6.7% of the cases in our study. The other 37.5% were unknown because of the difficulties in obtaining detailed information of the family history.

A high proportion of autosomal recessive diseases were also found in other countries with high levels of consanguineous marriage such as India,⁹ Sri Lanka,¹³ and eastern Mediterranean countries.^{15–18}

Interestingly, Leber congenital amaurosis (LCA) and early onset retinitis pigmentosa were the most common retinal dystrophies identified in our study. LCA had not been well recognised in Indonesian children; no cases have been reported before in the Indonesian population. On the other hand, as the knowledge of the molecular basis of inherited

retinal diseases is constantly increasing; the development of effective treatment strategies such as gene therapy becomes possible. LCA patients carrying defects in the RPE65 gene will probably be the first candidates to be potentially treatable in the future.¹⁹ With this perspective the need for paediatric ophthalmologists or ophthalmologists who are capable of identifying LCA in general, and its specific genotypes in particular, in blind Indonesian children is evident, in order to determine an appropriate treatment.

Congenital ocular anomalies such as microphthalmos and anophthalmos accounted for 10.9% of SVI/BL. The definite aetiology of this phenotype, whether genetically or prenatally acquired (teratological agents and intrauterine deformations),²⁰ is still unknown. This finding, however, was in accordance with those from blind school surveys done in Japan (11.2%)²¹ and Chile (7%).^{22, 23}

As the Wiyata Guna School for the Blind does not generally accept students with multiple handicaps, except if the additional handicaps are mild, most cases (98.2%) identified in this study had no disability apart from blindness. This finding is in accordance with the school for blind in China, but is in contrast with those in the United Kingdom and United States, where a high proportion of children have additional disabilities often associated with cortical visual impairments. The commonest associated disability in the present study was hearing loss in 1.2% and polydactyly associated with obesity in 0.6%.

Treatable and preventable cases

Corneal staphyloma, corneal scars, and phthisis bulbi were responsible for 29.7% of SVI/BL cases. Those condition were most likely attributed to infections and malnutrition in the postnatal/infancy or childhood period—that is, they are potentially preventable causes of blindness. Vitamin A deficiency could be related to the cause of visual morbidity. The incidence of morbidity and mortality rate related to vitamin A deficiency in Indonesia has decreased to 30–50%, since the high dose vitamin A supplementary campaign has been performed through *Posyandu*, an integrated primary health-care programme in selected urban slums and rural areas all over Indonesia. The vitamin A capsules are consumed by babies aged 6–11 months once yearly and children aged 1–5 years twice yearly in selected urban slums and rural area (Indonesian Ministry of Health; <http://www.gizi.net/pedoman-gizi/suplementasi-vit-a.shtml>, 13 January 2003). Health education, improvement of nutrition, and measles immunisation should become the strategies to prevent blindness caused by infection in the community.

Cataract, which was responsible for 13.3% of SVI/BL, is also a potentially treatable disease. Severe amblyopia occurred after cataract surgery in all cases, and was probably a consequence of a delay in surgery which was performed after the age of 3. This ought to be avoidable by early detection followed by appropriate surgical techniques and postoperative

rehabilitation. Similarly, most of the buphthalmos/primary congenital glaucoma cases found in this study (9.1% of SVI/BL) were in the late stage of disease at the time of presentation. At least the progression of this disease ought to be slowed down by early detection and appropriate glaucoma treatment in order to achieve better preservation of vision or restore sight.

Hereditary disease, a major cause of SVI/BL, in particular with autosomal recessive inheritance associated with a high rate of consanguineous marriages, could be prevented at least in part by genetic counselling. As mentioned earlier, in the upcoming years, LCA is expected to be a candidate eye disease for gene therapy. Therefore, much attention should be paid to identifying this disease among Indonesian children at an early age.

CONCLUSIONS

The relative frequency of genetic disorders and of infectious and nutritional conditions in our study lies in an intermediate position between the patterns seen in developing countries and those seen in developed countries.

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 primary congenital glaucoma.

The causes and problems identified indicate the importance of both preventive public health strategies and of specialist paediatric ophthalmic, genetic and optical services in the management of childhood blindness in Indonesia. A comprehensive primary eye care programme, paediatric ophthalmic units, and low vision services need to be established to face up to the problems of combatting childhood blindness in the context of Vision 2020: the right to sight. As this study provides information on the causes in a selected population (one single school for the blind), the findings as to absolute numbers have to be interpreted with caution. Population based studies are therefore necessary in order to obtain more appropriate epidemiological information on childhood blindness in Indonesia.

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Authors' affiliations

R Sitorus, M Preising, B Lorenz, Department of Paediatric Ophthalmology, Strabismology and Ophthalmogenetics, University of Regensburg, Germany
R Sitorus, Department of Ophthalmology, Faculty of Medicine, University of Indonesia

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