

DSP Fan 范舒屏  
 TYY Lai 賴旭佑  
 EYY Cheung 張欣欣  
 DSC Lam 林順潮

# Causes of childhood blindness in a school for the visually impaired in Hong Kong

## 香港一家視障學校學童致盲的原因

**Objective.** To identify the causes of blindness in children attending a school for the blind in Hong Kong.

**Design.** Cross-sectional observational study.

**Setting.** School for blind children in Hong Kong.

**Participants.** Eighty-two blind students at the Ebenezer School and Home for the Visually Impaired were examined between December 1998 and August 1999.

**Main outcome measures.** Demographic data were obtained from students and a questionnaire assessment made of their medical and ocular history. Visual acuity was assessed and visual loss classified according to the World Health Organization classification of visual impairment. Complete ophthalmic assessments were performed in all students including slit-lamp examination and dilated binocular indirect ophthalmoscopy.

**Results.** The mean age of the students was 12.2 years. Ten (12.2%) had a family history of eye disease. Major past medical illnesses were reported in 50% with prematurity and diseases of the central nervous system found in 26.8% and 11.0% of students, respectively. The most common anatomical site for visual impairment was the retina (47.6%), followed by diseases of the optic nerve (14.6%), and diseases of the anterior segment and the lens (14.6%).

**Conclusions.** The pattern of childhood blindness in Hong Kong is similar to that seen in other developed countries. Preventable causes of childhood blindness, such as prematurity and birth asphyxia, were responsible for a large proportion of cases. Early diagnosis and treatment of such conditions may reduce the incidence of childhood blindness in Hong Kong.

### Key words:

Blindness;  
 Cataract;  
 Child;  
 Optic nerve diseases;  
 Retinopathy of prematurity

### 關鍵詞：

失明；  
 白內障；  
 兒童；  
 視神經疾病；  
 早產新生兒視網膜病

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Department of Ophthalmology and Visual Sciences, The Chinese University of Hong Kong, Hong Kong Eye Hospital, 147K Argyle Street, Hong Kong

DSP Fan, FRCS (Edin), FHKAM (Ophthalmology)  
 TYY Lai, MMedSc, MRCS  
 EYY Cheung, BSc, MMedSc  
 DSC Lam, MD, FHKAM (Ophthalmology)

Correspondence to: Prof DSC Lam  
 (e-mail: dennislam\_pub@cuhk.edu.hk)

**目的：**確定香港一家盲人學校學童的致盲原因。

**設計：**橫斷面觀察研究。

**安排：**盲童學校，香港。

**參與者：**1998年12月至1999年8月，就讀心光盲人院暨學校並接受檢測的82位學童。

**主要結果測量：**研究人員從學童取得有關人口資料，並以問卷評估其病歷和眼疾的病史。研究亦評估學童的視力，以及按照世界衛生組織的視障分類標準劃分失明程度。所有學童均接受全面的眼科檢驗，包括裂隙燈檢查和擴張雙目間接檢眼鏡檢查。

**結果：**學童平均年齡為12.2歲，當中10位(12.2%)有家族成員曾患眼疾，50%受訪學童曾患重病，其中包括不足月出生(26.8%)和中樞神經系統疾病(11.0%)。視網膜問題(47.6%)、視神經疾病(14.6%)，以及眼球前部和晶狀體疾病(14.6%)是導致視障的主因。

**結論：**香港兒童失明的情況與其他已發展國家相似。早產新生兒視網膜病和新生嬰兒窒息等可預防的致盲因素，是造成不少兒童失明的主要成因。及早診斷及治療有助減低兒童致盲的機會。

## Introduction

Childhood blindness is a major problem throughout the world and results in a substantial life-long burden of disability. There are approximately 1.5 million blind children worldwide.<sup>1</sup> Childhood blindness has been identified as a priority in the World Health Organization's (WHO's) global initiative to eliminate avoidable blindness by the year 2020.<sup>2</sup> The prevalence of blindness in children is estimated to range from 0.2 to 0.3 per 1000 children in developed countries and 1.0 to 1.5 per 1000 children in developing areas.<sup>3</sup> In 2003, the mid-year population aged 15 or under in Hong Kong was about 1.1 million.<sup>4</sup> Extrapolation of these data produces a figure of approximately 400 to 1000 blind children in Hong Kong.

The causes of childhood blindness have been studied in various countries including Mainland China, and vary between undeveloped, developing, and developed countries.<sup>5-16</sup> Corneal scarring secondary to vitamin A deficiency, measles infection, and ophthalmia neonatorum are the main causes of childhood blindness in developing countries, whereas in developed countries, retinal diseases and lesions of the central nervous system (CNS) are more common.<sup>16,17</sup> Genetic and hereditary components also play an important role in these ocular disorders.

Determining the causes of childhood blindness is important because the epidemiological data may be used to determine how resources are allocated for the prevention and treatment of eye disease.<sup>15</sup> These data are nonetheless not readily available in many developed countries, including Hong Kong.<sup>2</sup> This study aimed to identify the causes of blindness among children attending the only school for visually impaired in Hong Kong.

## Methods

Students from the Ebenezer School and Home for the Visually Impaired were recruited between December 1998 and August 1999 for detailed ophthalmic examination. The school can accommodate 150 students, with space for 120 residential pupils.<sup>18</sup> The admission criteria for the school are: (i) Hong Kong resident, (ii) aged 4 to 18 years, (iii) visual acuity  $\leq 6/18$ , or visual field  $<20^\circ$  (Education Department criteria), and (iv) capable of benefiting from a regular education curriculum.<sup>18</sup> Inclusion criteria for the study included all students enrolled at the school for the academic year 1998/99. Students whose informed consent could not be obtained from their family members

**Table 1. World Health Organization (WHO) category of visual loss**

| WHO category             | Visual acuity                  | Students No. (%) |
|--------------------------|--------------------------------|------------------|
| No impairment            | 6/6 to 6/18                    | 4 (4.9)          |
| Visual impairment        | $<6/18$ to 6/60                | 7 (8.5)          |
| Severe visual impairment | $<6/60$ to 3/60                | 16 (19.5)        |
| Blind                    | $<3/60$ to no light perception | 55 (67.1)        |

were excluded. Initial ocular examination was carried out at the school by an ophthalmologist. At the time of examination, the children's parents or carers were asked to provide details of the children's general medical and eye condition including age of onset of the eye condition, perinatal period, previous eye surgery, and family history. Family members were also examined if the cause of visual impairment was hereditary or if ocular examination was requested. The main purpose of examining family members was to provide them with access to specialist ophthalmologist care. The students' best-corrected visual acuity was assessed, wherever possible, using a standard Snellen chart. Visual loss was classified according to the WHO classification for visual impairment (Table 1). Slit-lamp examination of the anterior segment was performed. Fundi were examined by binocular indirect ophthalmoscopy after pupillary dilatation using 0.5% phenylephrine and 0.5% tropicamide eye drops (Mydrin-P; Santen, Osaka, Japan). Some students required further ophthalmic investigations including ocular examination under sedation, fundus fluorescein angiography, ocular ultrasonography, automated visual field testing, and electrophysiological studies. These were performed at the Department of Ophthalmology and Visual Sciences, Prince of Wales Hospital, Hong Kong. The study protocol was approved by the ethics committee of the Chinese University of Hong Kong.

## Results

### *Demographics of participants*

A total of 82 students underwent ocular examination. This represented a recruitment rate of 78.8% of students. Ocular examination was not performed in 21.2% of students because written consent could not be obtained. The mean age (standard deviation) of the 82 students was 12.2 (4.3) years, with a range of 4 to 23 years.

### *Family and medical history*

A family history of eye disease was present in 10 (12.2%) students. Three had a family history of

**Table 2. Significant medical history**

| Medical history                        | Students<br>No. (%) |
|----------------------------------------|---------------------|
| Prematurity                            | 22 (26.8)           |
| Diseases of the central nervous system | 9 (11.0)            |
| Congenital heart disease               | 2 (2.4)             |
| Congenital toxoplasmosis               | 1 (1.2)             |
| Albinism                               | 1 (1.2)             |
| Kidney disease                         | 1 (1.2)             |
| Neurofibromatosis                      | 1 (1.2)             |
| Measles                                | 1 (1.2)             |
| Mild mental handicap                   | 1 (1.2)             |
| Musculoskeletal disorder               | 1 (1.2)             |
| Ectodermal dysplasia                   | 1 (1.2)             |
| None                                   | 41 (50.0)           |
| <b>Total</b>                           | <b>82 (100.0)</b>   |

congenital cataract. Other significant family histories included glaucoma, optic atrophy, corneal opacity, retinal dystrophy, retinopathy of prematurity (ROP), and high myopia.

Forty-one (50.0%) students had significant past medical illnesses other than ocular disease (Table 2). The two most common, prematurity and diseases of the CNS, were found in 22 (26.8%) and nine (11.0%) students, respectively. One student had a history suggestive of intrauterine infection causing congenital toxoplasmosis. Other significant medical history included congenital heart disease, albinism, kidney disease, neurofibromatosis, measles, mild mental handicap, musculoskeletal disorder, and ectodermal dysplasia.

#### **Previous ocular surgery**

Thirty-two (39.0%) students had undergone previous ophthalmic surgery on at least one occasion (Table 3). The most common surgery was cataract extraction (eight cases), four of which were performed for congenital cataract, three were performed in association with retinal detachment surgery, and one with penetrating keratoplasty. The majority of cataract surgery did not result in good visual outcome due to other ocular co-morbidities. Enucleation operations were performed in three students with a history of retinoblastoma and two who had end-stage glaucoma. Other types of ophthalmic surgery were for retinal detachment and glaucoma.

#### **Extent of visual impairment**

Using the WHO classification, 55 (67.1%) students were categorised as blind. Severe visual impairment was present in 16 (19.5%). The best-corrected visual acuity of the better eye is shown in Table 1. Four students with visual acuity between 6/6 and 6/18 attended the school because their visual field constriction due to glaucoma or retinal dystrophy was  $<20^\circ$ .

**Table 3. Type of ocular surgery among the 32 students who had previous ophthalmic surgery**

| Previous ocular surgery                    | No. of<br>students* |
|--------------------------------------------|---------------------|
| Cataract surgery                           | 8                   |
| Enucleation                                | 5                   |
| Glaucoma surgery                           | 5                   |
| Retinal detachment surgery                 | 5                   |
| Cryotherapy for retinopathy of prematurity | 4                   |
| Examination under anaesthesia              | 3                   |
| Penetrating keratoplasty                   | 2                   |
| Strabismus surgery                         | 1                   |
| Unknown                                    | 1                   |

\* Two students had more than one type of previous surgery

#### **Causes of blindness and onset of eye disease**

The anatomical sites of visual impairment are listed in Table 4. The most common was the retina with 39 (47.6%) cases. Major retinal causes included 21 students with ROP, seven with retinal dystrophies, four with retinal detachment, and three with retinoblastoma. Other causes included myopic and chorioretinal degeneration.

Optic nerve and CNS diseases due to optic atrophy or cortical blindness were the second most common anatomical sites for visual impairment with 12 (14.6%) cases and two (2.4%) cases, respectively. The known causes of optic atrophy included intracranial tumours (two cases), cerebral asphyxia (two cases), intracranial haemorrhage (two cases), and hydrocephalus (one case).

Disorders of the anterior segment, including diseases of the cornea and the lens, were also significant causes of visual impairment. Six (7.3%) students had corneal opacity, four (4.9%) had cataract, and two (2.4%) had anterior segment dysgenesis (aniridia).

Glaucoma with buphthalmos was diagnosed in 10 (12.2%) cases. Anatomical abnormality of the whole globe contributed to two (2.4%) cases. Vitreous disorders caused three (3.7%) cases of visual impairment which were due to persistent foetal vasculature.

In 50 (58.5%) students, the onset of eye disease was at birth and included ROP (21 cases, 42%), glaucoma (seven cases, 14%), and optic atrophy (five cases, 10%). Disease onset occurred in the first year of life in a further 11 (13.4%) students.

#### **Discussion**

The pattern of major causes of childhood visual impairment and blindness in Hong Kong is similar to that

**Table 4. Anatomical sites and causes of visual impairment in 82 students**

| Anatomical site                        | Cause                                | No. of students | Subtotal No. (%) |
|----------------------------------------|--------------------------------------|-----------------|------------------|
| Whole globe                            | Microphthalmia                       | 2               | 2 (2.4)          |
| Cornea                                 | Corneal opacity                      | 6               | 6 (7.3)          |
| Lens                                   | Cataract                             | 4               | 4 (4.9)          |
| Retina                                 |                                      |                 | 39 (47.6)        |
|                                        | Retinal dystrophies                  | 7               |                  |
|                                        | Retinal detachment                   | 4               |                  |
|                                        | Retinopathy of prematurity           | 21              |                  |
|                                        | Chorioretinal degeneration           | 3               |                  |
|                                        | Retinoblastoma                       | 3               |                  |
|                                        | Myopic degeneration                  | 1               |                  |
| Uvea                                   | Anterior segment dysgenesis          | 2               | 2 (2.4)          |
| Vitreous                               | Persistent foetal vasculature        | 3               | 3 (3.7)          |
| Optic nerve and central nervous system | Optic atrophy and cortical blindness | 14              | 14 (17.1)        |
| Glaucoma                               | Glaucoma                             | 10              | 10 (12.2)        |
| Others                                 |                                      |                 | 2 (2.4)          |
|                                        | Albinism                             | 1               |                  |
|                                        | Unknown cause                        | 1               |                  |
| <b>Total</b>                           |                                      |                 | <b>82 (100)</b>  |

seen in other developed countries. Optic atrophy, cataract, retinal dystrophies, and ROP are responsible for a large proportion of cases.<sup>3,15,16</sup>

Perinatal factors, including prematurity and birth asphyxia, accounted for approximately 10% of cases and resulted in ROP and optic atrophy. Intrauterine infections or drugs ingested during early pregnancy may also have caused significant visual impairment in these children. These causes of visual impairment are potentially preventable through better antenatal and perinatal care: high-risk neonates can be identified to allow treatment commenced early on in the disease process.

Congenital cataract, glaucoma, ROP, and retinal detachment are potentially preventable and treatable and accounted for one third of cases in this study. This finding emphasises the importance of early detection and treatment of paediatric eye diseases by screening (eg screening for ROP). Primary preventative measures should also be carried out where appropriate. Rubella immunisation in pubertal girls and non-immune women contemplating pregnancy can prevent congenital cataract. An epidemic of rubella in the United Kingdom in 1996 caused two cases of congenital cataract.<sup>19</sup> Advances in surgical techniques for various paediatric eye conditions, for example, paediatric cataract surgery with contact lens or intra-ocular lens implantation combined with aggressive treatment of amblyopia, may reduce the severity of childhood visual impairment.<sup>20</sup>

A large percentage of children in this study were blind at birth, and genetic and prenatal factors were important aetiological factors. Ophthalmic examination of parents and family members of the affected children would enable the provision of informed genetic counselling. Research is being carried out in order to build up an extensive database for genetic eye diseases and gene therapy for the eye is a future possibility.<sup>21</sup>

One of the weaknesses of the current study is the small sample size. Nevertheless, this study recruited over 75% of students attending the blind school and thus the results should be representative of the school population. Another limitation of this study is the selection bias that arises because a school for the blind, rather than the general population, was used for subject recruitment. Blind students not attending the school are thus excluded. Because a substantial proportion of children with other disabilities including deafness and mental handicap also have visual impairment, students in special schools for the deaf or mentally handicapped were also excluded from this study. Despite these disadvantages, blind school studies are widely accepted because a large number of children with visual loss can be examined in a short period of time.<sup>7,10,13</sup>

In conclusion, the causes of childhood blindness in Hong Kong are similar to those identified in other developed countries. Retinopathy of prematurity, birth asphyxia, and potentially treatable causes of visual impairment in children, such as cataract and retinal detachment, accounted for a substantial proportion of

cases. Because the risk of amblyopia is high in visually impaired children, early detection and diagnosis is of paramount importance to reduce the severity of visual impairment. Genetic studies and counselling will also play a significant role in the future when more become known about hereditary eye disease.

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