

9 CHILDHOOD BLINDNESS WORLDWIDE

9.1 Global Prospective

9.1.1 Introduction

There are an estimated 1.5 million blind children worldwide ¹⁸¹ with an additional 5 million visually disabled ²⁵³. Among those, 90% live in the developing countries ⁷⁵, with some 1.3 million living in Asia and Africa ⁶⁵ and some 72,000 of them in Europe, the USA and Japan ⁷⁸.

Approximately one child goes blind every minute ¹⁸¹ in the world, making a total of 500,000 children who go blind each year in the world, of whom 60-80% die in the subsequent 1-2 years from the disease that contributed to their blindness, or from neglect consequent upon being blind ^{75, 138}. In the UK, this figure is 10% ⁸⁰. In Africa, the prevalence of infant and childhood mortality (and also childhood blindness) is the highest in the world ¹³⁸.

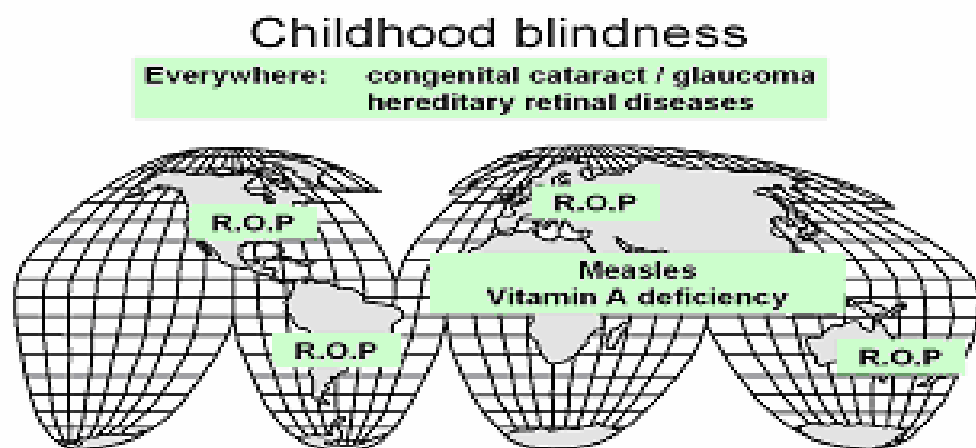


Figure 9-1: Childhood blindness worldwide

Adopted from WHO. Blindness: Vision 2020 - control of major blinding diseases and disorders. The Global Initiative for the Elimination of Avoidable Blindness. Fact sheet No. 214, reviewed February 2000. <http://www.who.int/mediacentre/factsheets/fs214/en/>

9.1.2 Regional Variations

There are variations in the causes of blindness across countries and the pattern of blindness changes with time ⁴⁸⁴. The prevalence of blindness in children is closely related to socio-economic development and the availability of health care in terms of nutrition, immunisation etc, and also to childhood survival and under five mortality rates. Those in excess of 170/1000 are likely to have a rate of childhood blindness in excess of 1/1000, whilst those with less than 30/1000 have rates between 0.2-0.5/1000 ^{65, 612}. More recently the rates in low-income countries have been estimated at a much higher level of 1.5/1000 ^{77, 99} (Figure 9-1, Table 9-1).

One of the most noticeable effects of an increased standard of living and improved health and sanitation, is the shift in the causation of blindness; from that due to nutritional deficiencies and infections in 'developing' countries to a hereditary causation in 'developed' countries. Whilst some of the 'developing' countries now show a similar pattern to technically 'developed' countries in the causes of blindness, many others are still held in the grip of blindness caused by poverty, poor nutrition and poor hygiene ^{65, 610}. Gilbert et al ¹⁷¹ noted that retinal dystrophies were the commonest form of genetic eye disease (49-80%) in all countries apart from Thailand and the Philippines).

Region	Population <15	Prevalence Per 1000	Estimated Nos. (millions)
Africa	240	1.1	264,000
Asia	1200	0.9	1,080,000
Latin America	130	0.6	78,000
Europe/USA/Japan	240	0.3	72,000
Total			1,494,000

Table 9-1: Estimated prevalence and magnitude of childhood blindness, by region (WHO, 1992)

(Adopted from Gilbert, Epidemiology of Eye Disease 1998) ⁷⁹.

9.2 Impact of Childhood Blindness (Table 9-2)

The impact of blindness in childhood is enormous. This is summed up in the following Table 9-2.

Obviously childhood blindness is recorded much earlier and, therefore is of longer duration. When “total person-years” of blindness are calculated, the impact of monogenic blindness is much greater in this population at 6,849 person-years, compared to 270 for diabetes and 430 person-year for ARMD^{22, 99}.

Education

<10% of blind children in many developing countries have access to special education.

Economic

In India the cumulative economic lifetime loss for individuals blind in childhood is estimated to be US \$ 22 billion.

Quality of life

Requires further study

Table 9-2: The impact of blindness on childhood

Adopted from Gilbert: Eliminating of Childhood Blindness⁶¹¹

9.3 Causes of Childhood Blindness

Childhood blindness in poorer countries is caused mainly by VAD, measles, conjunctivitis in the newborn and congenital cataract. Nutritional blindness is common in almost all the developing countries with an estimated 350,000 cases of xerophthalmia annually, although this is no longer considered a problem in Sri Lanka, Myanmar, and Korea³²³. ROP was an established cause in developed countries which is now subsiding as a cause of blindness; this as a result of the concerted efforts made in recent years by prompt screening of premature babies and treatment of early retinopathy. Instead, blindness from the neurological sequences of prematurity is emerging as the main cause. In middle-income countries, however, ROP is beginning to emerge as an important cause⁴⁸⁴. Ophthalmia neonatorum, and less

frequently seen ocular diseases such as HSV infections and vernal keratoconjunctivitis, are other causes^{253, 322, 323}. Other causes of childhood blindness that are congenital or genetically determined, do not generally lend themselves easily to preventive strategies at present.

Foster and Gilbert⁶⁵ concluded that at least 50%, and probably 75%, of childhood blindness in the developing world is potentially preventable or curable with today's level of medical knowledge and expertise.

Risk factors contributing to childhood blindness are: poverty, uneducated mothers, cultural practices, poor primary health and eye care, poor surgery and follow up, poor optical services, and lack of special education or rehabilitation⁶¹¹ and inbreeding^{19, 23, 33, 40, 88}.

9.4 Data Collection

Data collection is also problematical^{78, 79} in respect of childhood blindness and this subject is addressed in section 5.3. A relevant point in evaluating CB is the observed possible improvement of visual functions in visually disabled children with increasing age. This was reported in a study of 123 Swedish patients born between 1962-1976 who had their medical records reviewed, were interviewed and re-examined when necessary. In 59% of them vision was still impaired, while in 23% the vision had improved to more or equal 0.3. This gain was significantly higher for patients without additional impairments. Conditions that showed improved visual development included albinism and congenital nystagmus, while retinal diseases showed poorer results. An increase in visual function was seen even in the initially older age groups, indicating a maturation of visual function beyond what is usually considered the limit of plasticity of the visual system^{470, 646}.

9.5 Gender Differences

Male dominance in childhood blindness in various proportions has been described in several studies, particularly those based on school surveys and blind registers^{33, 43, 86, 96, 97, 141, 330}. This phenomenon however was not evident or significant in other studies, particularly those obtained via population studies and on some occasions the picture was different^{80, 120, 121, 129, 177, 326, 653}.

Male predominance has been attributed to several factors, (Table 9-3), although in many instances it remains inexplicable or has not been addressed by the authors of these studies^{33, 43, 86, 96, 97, 141, 330}. Social factors are important in developing countries, as a result of a bias in the admission to blind schools; this however appears to be on the decline as seen by comparing India's figures in 1968¹⁴³, when the M/F ratio was 14-fold higher in boys (12.8:1), to the current ratio of 1.7:1¹⁴¹.

Male favouring in blind registration was put forward by Schappert-Kimmijser²²³ in the Netherlands, but this has been questioned in recent studies from the same country⁶⁷⁵ and has been discarded in other countries⁹⁷.

Gender differences have been thoroughly addressed in the Nordic studies, where a tendency towards male preponderance was found in all these countries. This was more significant in the Danish and Finnish register^{96, 135} in both the genetic and non-genetic cases¹³⁵. This predominance was also found in all the diagnostic groups except the mixed group, in particular the congenital dysfunction group (1.9:1) and the neuro-ophthalmic group (1.32:1). The M/F ratio in the general population in the Nordic countries is 1.045:1.

The dominance of males in genetic cases can be easily explained on the basis of the X-linked conditions; however, it is difficult to account for it in the perinatal conditions. The commonest X-linked conditions contributing to this predominance are albinism, choroïdæmia, X-linked juvenile retinosis, Lowe syndrome and X-linked cataract. Genetic factors (X-linked) are found especially in the Danish and Finnish groups (table 9-4).

Causes of Male Predominance in Childhood Blindness

- Social factors favouring boys in
 - School admission
 - Inclusion on the register²²³
- X-linked conditions^{90, 96, 135}
- Perinatal affections^{90, 96, 135, 669}
- Cataract associated with perinatal difficulties⁹⁰
- Optic atrophy^{96, 135}
- Buphthalmos⁹⁰
- Unexplainable^{97, 669}

Table 9-3: Male predominance in childhood

Study	Genetic	Non-genetic	Isolated vi	Multiple
Danish	1.42:1	1.35:1	1.32	1.43:1
Finnish	2.42:1	1.38:1	1.82	1.55:1

Table 9-4: Male predominance in relation to aetiological categories ¹³⁵

The significant preponderance of males found in the prenatal asphyxia and prematurity has been attributed to genetic factors predisposing to these conditions; this is supported by the evidence that male babies deliver at earlier gestation than females ¹³⁵. The same applies to optic atrophy and perinatal lesions, which may be due to X-linked disorders inducing pre-term birth and perinatal asphyxia ⁶⁶⁹.

The remainder of this chapter covers a search of literature on the prevalence, incidence and causes of childhood blindness in the various regions and countries worldwide.

9.6 Childhood Blindness in the EME and Europe

9.6.1. Introduction

Europe differs in many aspects, such as political, socioeconomic, and geographical, and there is a noticeable demarcation between EME and PSE countries of Eastern Europe. After substantial political changes at the beginning of the 1990s, the majority of central and eastern European countries started to rebuild their healthcare systems. It is apparent that eastern Europe represents a highly diverse region where the difference among countries broadens year after year ⁷⁶.

9.6.2 Historical Aspects

One of the earliest statistics on childhood blindness quoted in the literature came from the UK on the causes of blindness among the inmates of the Liverpool School for the Indigent Blind between 1791 and 1872. This revealed that nearly half of the pupils had lost their sight from inflammations and infections and, of these, one third were as a result of smallpox. By the end of

the 19th century, ophthalmia neonatorum was the leading cause of blindness in schoolchildren, being responsible for approximately 30% of cases ^{70, 82}.

Sorsby analysed the causes of blindness between 1948 and 1968 and highlighted the increasing trend towards genetically determined causes of blindness, and the almost 50% decline in blindness between 1923 and 1943. He found that in the age group 0-4 years and 5-14 years, congenital, hereditary and developmental defects accounted for 60-62.5% of all cases. However, in the 15-29 years, although the same group of causes predominated, its percentage was much less at 38.7% and other infective aetiologies such as syphilis and ophthalmia neonatorum contributed to 13.7% and 12.2% respectively ⁷¹⁻⁷². This trend was confirmed by a later DHSS study based on blind and partially sighted registrations of children under the age of 16 years between the years 1976-85. This study included 2,000 blind and a further 2,400 partially sighted children. Of note was the fact that there were disproportionately more boys than girls. Analysis of the causes is limited by the fact that only 60% of registrations had accompanying blind Registration forms. The main cause found for both sexes was optic atrophy (30%), followed by congenital anomalies (20%), and retinal conditions (20%). The only new finding in this study, in comparison to reports covering the period 1969-1976, is the drop in the incidence of cataract from 16% to 8% ^{83, 225}.

In main land Europe, and as late as the 1970s prenatal and Perinatal factors mainly rubella, toxoplasmosis, dysoxygenation, other affections during pregnancy, prematurity and low birth weight and birth injuries such as asphyxia were considered important contributors ²¹¹, according to a survey of 4306 children in five countries (Belgium, Denmark, Sweden, Norway and the Netherlands), although hereditary conditions were the prime aetiology of blindness ²¹¹.

9.6.3 Changing Patterns

Data on new registrations (ie 1968) had shown no substantial decline in blind registrations despite the end of registrations for those with ROP. Much of the changes and increase in those registered was due to administrative changes and changes in the age structure of the population.

Fraser and Friedman in 1967⁹⁰ undertook a study of 776 children on the blind register in the UK, almost one quarter of the total number of children registered with severe visual handicap. These were from a variety of special schools that included blind children with additional handicaps. This study went further than previous ones in that all those included underwent ophthalmic and systemic examination and investigations, in addition to detailed histories being obtained from the families. The study revealed that the three commonest causes of blindness, accounting for more than 50% of all cases, were retinopathy of prematurity, chorioretinal degenerations (RP, LCA, and related conditions), and cataract. About 42% of the disorders were genetically determined with 20% of all cases being AD, 17% AR and 5% x-linked; 8% were thought to be multifactorial.

In the main land Europe, many differences in childhood blindness were demonstrated. In the PSE, the prevalence of between 0.01/1000 and 0.04/1000 with the main anatomical causes being retinal disorders and optic atrophy. This is in contrast to the highly industrialised countries of Europe, where the leading causes of childhood serious visual loss are CNS lesions, congenital anomalies and retinal disorders. This is a trend similar to that reported in England and Scotland. In the middle-income countries of Europe, congenital cataract, glaucoma and, mainly, retinopathy of prematurity are highly expressed⁷⁶.

9.7 Childhood Blindness in the UK and Ireland

9.7.1 The prevalence

The prevalence of childhood blindness in the UK was assessed by Stewart-Brown and Haslum in 1980, in a nationally representative sample of 15,000 10-year-old children. The prevalence of blindness was between 3.4 and 4.0/10,000, and partial sightedness (SVI) between 5.4 and 8.7/10,000⁶⁵⁴.

9.7.2 Causes and Aetiologies

Genetically determined disorders accounted for 51% of 386 subjects studied in Northern Ireland, by Bryars and Archer in 1977⁸⁹ which was considered to be in excess of any other survey at the time, with the exception

of those undertaken in Cyprus²³ and Lebanon⁸⁶. These were followed in importance by pre- and perinatal complications such as pre-eclamptic tox-aemia, dysmaturity, prematurity etc. These findings demonstrated significant differences from those reported 10 years earlier in a study of children from selected schools by Fraser and Friedman⁹⁰. The rate of cataract in the Northern Ireland study had risen to from 15% to 32% and ROP had diminished from 20% to 12%. The former was also the most common cause of blindness or partial sight in 1980 in the UK followed by CG⁶⁵⁴.

Similarly, but to a lesser degree, a study by Phillips in 1987 of 99 children at the Edinburgh Royal Blind School⁹¹, in a comparison to a study done in Japan by Tsukahara⁹², confirmed hereditary causes as responsible for between 48-58 of the 99 pupils. The major causes of blindness were optic atrophy (15 cases), congenital cataract (12 cases), retinal aplasia (LCA) (12 cases), often with other handicaps, and ROP (11 cases)⁹¹.

More recently, 15 years from the last study a repeat study at the same blind school in 2002 by Alagaratnam demonstrated a new trend in childhood blindness⁹³, away from hereditary causes and towards perinatal causes (40%), with prematurity forming the largest single group, followed by hereditary factors (26%); most commonly these were optic atrophy or retinal dystrophy. The majority of the latter were thought to be AR in origin. Disease or malformation of the CNS or optic nerve was the commonest cause of visual impairment and was present in 50% of the total examined. It is also interesting to note that although ROP accounted for 18%, the youngest child with ROP was 9 years old, highlighting a decreasing incidence. A similar trend was demonstrated by Schwarz et al in the City of Bradford, England in the same year⁹⁴. Bamashmus et al confirmed that congenital glaucoma, cataract and corneal infections were no longer causes of registration in children; instead registration caused by brain damage has become a major contributor¹⁸⁹.

9.7.3 Mental Retardation in the UK Cohorts

Mental retardation was the most common defined additional impairment, being present in 29 (27%) of those examined and many of the children had multiple impairments due to brain injury eg cerebral palsy, mental retard-

ation and epilepsy. However, a caution in interpreting the results was raised given the small size of the study in a selected population. It was put forward that additional fields for more detailed reporting of cerebral visual impairment and associated handicaps were needed to increase the usefulness of the WHO/PBL form for population based studies and also for use in developed countries ⁸⁰.

9.7.4 British Paediatric Surveillance Study

A UK study of particular interest is that of Rahi and Cable of 439 children, newly diagnosed during the year 2000, with severe visual impairment and blindness. The study identified eligible children through both the British Ophthalmological and British Paediatric Surveillance Units ⁸⁰. It was pointed out that notification by ophthalmologists and paediatricians had not been totally independent, and that a child was less likely to be reported by one specialist if the other was involved in the care and that communications existed between specialists. However, a simulated analysis of all children being treated by both specialists (376) suggested that 99% of eligible cases had been ascertained. The study included only children with permanent SVI/BL at one year of follow-up, ie it excluded those whose vision was better after treatment or spontaneously. The author found that SVI/BL occurred more commonly in the context of complex non-ophthalmic impairments and had greater associated mortality than previously assumed. The study found 336 (76.5%) with additional non-ophthalmic disorders, and 10% of all children studied (439) died from the additional disorder within 1 year of the diagnosis of blindness. Similar mortality rates were reported in Swedish children ²⁰¹ (13%) the majority of whom died from respiratory causes.

Rahi' et al study ⁸⁰ concluded that children of low birth-weight, and from ethnic minorities, had the highest rates of SVI/BL, with those from the most deprived socio-economic groups being over-represented. Three-quarters of those studied had disorders that were neither preventable nor treatable; the major anatomical site affected being the cerebral/visual pathways (77%). These were more frequent in those with SVI/BL plus, than in those with isolated SVI/BL. Conversely, retinal disorders, (particularly retinal dystrophies and albinism) and glaucoma were more important in those with

isolated visual loss than in those with SVI/BL plus. Hereditary disorders were found to affect 46% (n=32) of south Asian children (Indian, Pakistani or Bangladeshi) compared with 29% (n=86) of white, 33% (n=6) of black, and 33% (n=9) of other ethnic groups. It was also found that the prevalence of blindness in this study was greater than anticipated from published yearly rates of notification to the British registers of blindness over the previous two decades. The trends reflected the changing picture of chronic disease and disability in childhood more broadly, in particular the associations with low birth-weight, ethnic origin and socio-economic deprivation. The author concluded that the scope for intervention against blindness in children in the UK, and other industrialised countries, was very limited.

9.8 Childhood Blindness in the Republic of Ireland

A survey in the Republic of Ireland of 172 children (93 males and 79 females) between 1989 and 1990, showed that 56% of the causes were congenital. Of these, 40% were due to prenatal factors and 16% genetic. The commonest perinatal conditions were birth asphyxia in 11% of cases and ROP in 11% of cases⁶⁵³. Aetiological factors are shown in Table 22-6.

9.9 Childhood Blindness in North European Countries

9.9.1 Prevalence

Significant differences in the prevalence of blindness were reported in Denmark, Iceland, Finland, Norway and Southern Sweden in 1993 despite identical classification criteria. The reported number was 2527 visually impaired children from the total <17 years child population of 3,818,000 which formed 22% of the total population of these countries. The prevalence rate in these countries varied from 0.15/1000 to 0.41/1000 (blindness) and 0.48/1000 to 1.05/1000 (visual impairment). This variation was attributed mainly to under-registration in the countries with the lowest rates. Visual impairment had been reported before the age of 3 years in 50% of cases^{95, 96, 135-137, 222}.

9.9.2 Causes of Childhood Blindness

Prenatal factors, including genetic conditions, accounted for 66% of the cases in the Nordic Countries but in children without additional impair-

ment the percentage was 74%. Genetic factors accounted for a little over half of prenatal cases and in 40% of cases of prenatal aetiology the causes were unknown. Compared with a previous prevalence study in 1992, the relative impact of ROP had dropped from the third most frequent cause (10%) in the prevalence study, to seventh (4%) in the incidence study^{96, 708}.

The predominant causes in the Nordic countries were congenital malformations, neuro-ophthalmic disorders and retinal conditions. Optic atrophy was the single most common cause. ROP was the principle cause for severe visual impairment. Congenital cataract was also an important cause. Visual impairment due to brain damage, which was referred to as cerebral amblyopia, accounted for 23% of cases affected, with different proportions in the 5 countries, with a range between 36% in Sweden to only 10% in Finland. The authors noted a trend towards an increase in multiply impaired children from 30-50% in the earlier study, to 61% in this study^{137, 223}.

Other studies in Sweden, comparing the current trend in childhood blindness to that in 1980, also reached the same conclusions of the presence of a shift from prenatal to perinatal causation noted above²⁰¹⁻²⁰³.

9.9.3 Associated Findings in the Nordic Study

Two thirds of the children in the Nordic countries who suffered from the most severe visual impairments, also had additional impairments, mainly secondary to brain disorders. The proportion of visually impaired with an additional morbidity, hearing or mental impairment, were between one-third and one-half of the national cohorts, indicating the need for interdisciplinary tracing of, and care of, the visually impaired child^{96, 468}.

9.9.4 Greenland

Data from Greenland⁹⁵ compares to the findings in the Nordic countries^{95, 96, 135-137}. Only 38 cases of congenital visual impairment were reported in the Inuit population of Greenland over a period of 40 years (1950-1989) representing an incidence of 0.86/1000 live born children. The two most common causes of blindness were optic atrophy and cerebral visual impairment due to brain disorders of various aetiologies. ROP and congenital cataract were rare causes. Fifteen out of the 38 cases were of unknown aetio-

logy. Genetic disorders accounted for 18.4% of cases, significantly lower than the 66% found in the Nordic countries^{96, 463}. The number of cases of probable genetic visual impairment was 25 patients. The causation of the isolated cases of aniridia, lens ectopia, and Down syndrome, was attributed to new mutations, while inbreeding was a possible contributory factor in a few AR conditions⁴⁶³.

9.9.5 The Netherlands

The figures date back to 1980 when Van Der Pol reported in a study of 1334 pupils in 11 Dutch institutions for visually handicapped children that the pattern of blindness had not changed in ten years, and that congenital cataracts, tapetoretinal degenerations and disorders of the optic nerve were still the three most common causes of childhood blindness in the country⁶⁷⁵.

9.10 Childhood Blindness in Germany

9.10.1 Prevalence

The proportion of blind children in Germany, which applies to the rest of the industrialised countries, from the total blind population has dropped significantly in the past 100 years from 12% in the 19th Century to about 5% at the present time. Until 1901 it ranged between 9.6% - 26% in the various studies in Germany, came down to 8.8% in 1926 and 0.1-12.6% in 1952, reaching a constant rate from 1973 onwards. Currently it averages 3.9%.

9.10.2 Causes

The causes also varied as expected. These are shown in Table 9-5 below⁶⁶⁹. Schmidt et al¹⁹⁸ highlighted the changes in the course of CB including the significant reduction in the number of blind children since the 1960s and especially since the 1970s. Corneal causes secondary to infections, which formed 25% of childhood blindness until 1940, had dropped to 2%. There was also an obvious reduction in blindness due to uveitis from infections and chorioretinitis and the total eradication of blindness from trauma, with no case of sympathetic ophthalmia since 1950. The incidence of congenital cataract and congenital glaucoma remained unchanged, although they are rarely associated with blindness as a result of improvement in the

management of these conditions.

The incidence of blindness caused by retinal detachment and retinoblastoma remained unchanged, though rare. The commonest causes of morbidity were optic atrophy (20%) and retinal degenerations (16-19%) whose incidence has remained constant and high. On the other hand, a major increase was noted in ocular malformations and developmental anomalies including ROP. The latter was responsible for 38% of childhood blindness.

In a more recent study in south Germany (1996), Haassler et al²⁰⁹ reported the aetiologies of visual impairment and mental retardation in 239 multi-handicapped blind and partially sighted

children born between 1981 and 1987. Marked aetiological heterogeneity was found. The commonest causes were due to perinatal factors and late gestation (41%). Amongst these lesions in the visual pathways in preterm children were very frequent and were explained by periventricular keratomalacia in the majority of instances. The distribution of the major aetiological groups was different from populations with isolated mental retardation and populations of isolated visual impairment, but similar to the Swedish population with spastic quadriplegia. The prevalence of the above visual impairment was found to be low at 0.67/1000 live births²¹⁰.

9.11 Childhood Blindness in Switzerland

Figures from Switzerland date back to 1987 when 262 children with blindness and visual impairment were examined; children with severe brain damage and multiple handicaps were not included in the study. The Congenital malformations accounted for 53% of the cases, ROP for 25%, infect-

<u>Years</u>	<u>Causes</u>
1885 – 1939	O/N and cornea
1984	Injuries
1950	Malformations
1967	Genetic
1972	ROP
1973 – 1978	OA, congenital
1984	ROP
1985	Congenital causes
O/N:	Ophthalmia Neonatorum
OA:	Optic atrophy

Table 9-5 Causes of childhood blindness since 1885 in Germany⁶⁶⁹

ions for 10% and tumours for 10%. In the visually impaired group, metabolic disorders, and accidents were additional causes ²¹⁴.

9.12 Childhood Blindness in the PSE Countries

Figures from countries that belong to the PSE Group are available from 4 countries namely; the Czech Republic, Slovakia, Poland and Hungary ^{97, 100, 103, 112}. In the Czech Republic ⁹⁷ (42%) of CB is caused by ROP, and in Poland ROP contributes to 19% of cases ⁹⁸. In the latter, a 70% increase in the number of visually disabled children took place in the 20 years between 1979 and 1999 with the major cause being optic nerve atrophy (21.66%). Hereditary factors accounted for the majority of cases in Hungary and Slovakia at 50% in the former and 90% and 48% in the latter in one blind school and hospital admissions respectively ²¹⁵. In Hungary there was relatively more AD than AR and sex-linked conditions, while acquired causes accounted for about 40% of the cases ^{100, 103, 112}. The prevalence of severe visual handicap was 0.43/1000 ¹¹¹.

9.13 Childhood Blindness in France

In France, data in English is scarce. In the mid 1990s, cataract and congenital nystagmus were the main causes of blindness as found in one study of 340 children and adolescents in two specialised schools in northern France ¹⁹¹.

9.14 Childhood Blindness in North America

9.14.1 Changing Pattern in Canada

In North America and Australia the picture, including the changing trends, are in line with the rest of the EME countries. Earlier studies in 1965 in Canada, by MacDonald revealed that approximately 30% of a total number of 24,605 on the blind register (of whom 1,865 were aged 0-8 years) had genetically determined eye diseases, with the predominance in cataract (41% of all cases of cataract), together with retinal lesions ¹⁰¹. The incidence of acquired causes of visual impairment had dropped by two thirds from 6 to 2 /100,000 between 1960 and 1989 as reported from analysing data on children younger than 19 years of age with visual impairment equal to and less

than 6/60²³⁷. The quality of information began to improve in the early 1970s. Cataract (13%), optic atrophy (12%) and nystagmus (10%) surfaced as the three most common causations of CB in 1046 children under the age of 20 years²³⁸. ROP formed a small group at that time (6%)²³⁷. Earlier data were not sufficient to establish the aetiologies of these conditions, whether hereditary or environmental. The predominance of genetic causations were later established by Robinson and Jan²³⁷ in a retrospective analysis of data in 1989. These were followed by tumours, injuries, infections and autoimmune disorders. Acquired causes formed 25% of the aetiologies, whilst optic nerve atrophy together with retinal disorders rose to 90% of all ocular pathology. They also demonstrated that the percentage of children with neurological disorders had increased in the past 30 years because more children with profound brain damage had survived²³⁷. In the highly inbred region of Newfoundland and Labrador, single-gene disorders accounted for 30% of total blindness in children (and also adults) and congenital defects for another 10-11% in 1986²².

9.14.2 Childhood blindness in the USA

There is a general lack of comprehensive information available for children in the USA in the recent past¹⁶⁷. There is no national Registry of the blind in the USA, and most of the schools for the blind do not keep data regarding the cause of blindness in their students¹⁰⁴. Of the few early studies available on the causes of childhood blindness were those of Delaney in 1962¹¹⁷, Gillespie et al in 1963²³¹, Hilgartner in 1967¹¹⁶, Fraser¹⁰² and DeCarlo²²⁴ in 1999.

Delaney's¹¹⁷ study was based on the records of 834 pupils in Pennsylvania in 1962. He found retinopathy of prematurity to be the largest single cause of childhood blindness affecting 307 (36.9%) of pupils, an expected finding for that period of time, and he anticipated a 30% reduction in the following decade. Congenital cataract was found to affect 99 (11.9%) of pupils followed by optic atrophy in 79 (9%) of pupils.

Hilgartner¹¹⁶ studied the records of pupils attending the Texas State School for the Blind during several periods; 1929-30, 1939-40, 1948-49 and 1966-67. Over that period of time the three main conditions to have declined

in importance were noted as sympathetic ophthalmia, O/N, and syphilis. As with the Delaney study, the greatest problem found in the later period of the study was ROP, with the numbers entering the school with this diagnosis rising from 1 in 1955 to 10 in 1966. He also noted that congenital conditions had remained at more or less the same percentage during the preceding 37 years, the causes of which included congenital cataract, albinism, microphthalmos, congenital glaucoma, dislocated lens and congenital absence of the anterior segment of the globe.

Along with the rest of the EME countries, a further change in the trend of blindness has taken place in the USA, that is a shift to optic nerve pathologies as demonstrated by DeCarlo²²⁴ in his retrospective study of the medical records of 123 students at the Alabama school for the blind between 1996-1997. Optic nerve pathologies constituted 30.9% (optic atrophy 13%; Leber's optic atrophy 4.1%; optic nerve hypoplasia 5.7%; and glaucoma 8.1%). Other common causes included congenital malformations (other than aniridia) 12.2%; cataracts/aphakia 13.8%; albinism 13.0%; and retinitis pigmentosa 8.1%. Nystagmus, chorioretinitis, and other conditions accounted for the remaining 22% of diagnoses. Visual acuities ranged from 6/6 to NLP, with 44.3% having acuity better than 6/60; 26.2% having 6/60 to <3/60; 13.1% having measurable acuity of 3/60 or worse; 9% having LP; and 7.4% having NLP.

Leonard¹¹⁸ revealed that in 1990, data on legal blindness in the USA indicated that approximately 2,600 children under 5 years of age, and approximately 51,000 between the ages of 5-19 were legally blind. Among the children under 5 years, prenatal cataract was the leading cause of legal blindness, accounting for 16% of all cases. This was followed by OA (12% of all cases) and retinopathy of prematurity (9% of all cases).

Steinkuller¹⁰⁴ noted that of those blind schools in the USA that did keep information on the causes of blindness, the top 3 causes were cortical visual impairment, ROP and optic nerve hypoplasia. There has been a significant increase in both cortical vision loss and ROP in the preceding 10 years. It was noted that there were marked regional differences both in the prevalence and causes of paediatric blindness, but as with elsewhere studied these were based on socio-economic factors. As noted earlier, there is no

national registry of the blind in the USA and the authors concluded that there was a need for more complete and uniform data based on the WHO reporting format.

9.14.3 Childhood Blindness in Australia

In Australia, Fraser¹⁰² undertook a study into the causes of blindness in two schools in South Australia with a total of 50 pupils between the ages of 5-18 years. Several were the offspring of immigrants (27 of the 100 parents had been born outside Australia and three of the children). Of the 50 children examined, 25 had acquired, prenatal causes of blindness of which the major cause (20%; 10 cases) was due to prenatal rubella, followed by retrolental fibroplasias (ROP) (10%; 5 cases). The number of children affected by genetic causes of blindness was 17, of which retinal aplasia (LCA) produced the highest number (n = 6). Blindness from uncertain causes was recorded in 8 children. Of the 10 children diagnosed as rubella embryopathy, 5 were severely deaf and 4 had milder hearing loss whilst 6 of the 10 had been born in 1958 suggesting a particularly severe epidemic in the second half of 1957. One of the other interesting features noted was the presence of 2 children in this small series with sex-linked myopia. Another large unconnected group was also found in South Australia leading Fraser to conclude that the prevalence of this condition was unusually high in males in this region and possibly attributable to 'founder effect'.

9.15 Childhood Blindness in Latin America and the Caribbean

9.15.1 Introduction

Whilst Central, and South America (including the Caribbean) cover an area of extreme diversity geographically and socio-economically, very little is known about the magnitude of childhood blindness within the region¹⁶⁷. In 1988, Foster¹²⁰ indicated that 25% of childhood blindness in Latin America resulted from corneal scarring as a consequence of malnutrition and ocular infections, occurring mainly in the rural areas.

Gilbert et al^{122, 79} highlighted that ROP was becoming a major cause of potentially preventable blindness in middle income countries, in essence a second epidemic, in middle income countries that have introduced neonatal

intensive care services. In Cuba for example, ROP accounts for 38.6% of childhood blindness. The true size of the problem is not known, as many of these countries do not have blind registers.

9.15.2 Childhood Blindness in Jamaica

A study which does not fall easily into the above two categories is one by Moriarty in 1988¹¹⁹ in Jamaica of 108 children aged 5-15 years. This study showed similarities to that of Fraser's⁹⁰ in the UK in that the rate of hereditary disease was 48% (UK 50%). Moriarty highlighted that, in Jamaica, rubella often affects those of childbearing age, being the leading preventable cause of CB accounting for 22% of children examined. This is in contrast to the studies in Cyprus, Lebanon, Saudi Arabia and Nigeria, where rubella is usually contracted at a pre-childbearing age. The major difference between this study and Fraser's UK study is the total absence of blindness from perinatal causes in Jamaica resulting from the lack of premature babies care. Whilst there was little consanguinity in the Jamaican population, the village population is endogamous and the social practice whereby a man may father children by several women, makes it difficult to establish precise lines of heredity. It is interesting to note also that the recent economic climate in Jamaica, which had been characterised by increasing poverty, unemployment and rising food prices, led to an increase in hospitalisation for malnutrition and VAD in addition to the reappearance of xerophthalmia after an absence of ten years.

9.15.3 Childhood Blindness in Chile

In a study by Gilbert et al in 1994¹²¹ of 421 children in Chile in ten schools for the blind, 318 (76%) were found with severe visual loss. Of these, 29.6% were attributable to hereditary factors; 22.5% to perinatal factors; 11.2% to childhood factors and 8.2% to intra-uterine factors. Aetiology could not be determined in 28.5%. ROP accounted for 17.6% of all children with severe visual loss suggesting that it was becoming an increasingly important cause of blindness. It was also estimated that 50% of children had avoidable blindness.

9.15.4 Childhood Blindness in Peru

In Peru ¹⁷⁰, Rojas et al studied 202 children at the school for the blind in Lima, the referral centre for all severely visually impaired Peruvian children. Some 53% of blindness was caused by congenital and hereditary disorders. The leading causes are leucoma (15.3%), congenital glaucoma, congenital cataract and retinal degenerations (11.9% each), and optic atrophy and retinoblastoma (8.4% each). The latter reflected the advanced stage of the disease in these children. Measles was the cause of the leucoma in 51% of that category; a finding similar to that reported by Olurin ¹²⁴ in Nigeria. Measles accounted for 10% of blindness.

9.15.5 Onchocerciasis in Children in LAC

Onchocerciasis remains a problem in 6 countries in Central and South America (Brazil, Colombia, Ecuador, Guatemala, Mexico and Venezuela) in localised areas affecting the extremely poor ^{167, 168}. However, it is not perceived as a serious health problem in these countries ¹⁶⁹. OEPA has been active in these countries since 1990¹⁶⁷.

9.16 Childhood Blindness in Sub-Saharan Africa (SSA)

9.16.1 The Size of the Problem in SSA

In Africa (and also Asia), the picture of childhood blindness is very different from the countries visited so far. An IAPB Report ¹²³ in 1984 stated, "ignorance, poverty, superstition, adverse cultural practices and some fatalistic indifference, contributed to the gravity of the problem in Africa". Seventy percent of blindness in SSA was due to corneal scarring, the main causes of which were xerophthalmia and measles, a pattern that echoes in many African countries including Malawi, Kenya, Tanzania, Ethiopia and Nigeria ^{126, 127, 133, 134}. Reduction in corneal scarring from improved measles immunisation coverage rates has been achieved in some African countries such as ^{77, 131}.

Childhood Blindness was preventable in 50.7% of the cases and an additional 17.3% were blind from causes that were potentially treatable. These figures for avoidable/preventable blindness were reported across the board in SSA including Malawi (67.2%) and Uganda (56.7%), Kenya (28.6%) and South Africa (38.8%) ^{128, 130, 131}.

9.16.2 Causes of Childhood Blindness in SSA

The above report ¹²³ split the causes of blinding conditions geographically; showing onchocerciasis present in most countries between the latitudes 15° north and 15° south, the worst areas being the savannah zones. Trachoma was endemic in the Sahelian belt and the Sudan as well as many countries in central and southern Africa. Congenital cataract was common throughout Africa and this, combined with congenital glaucoma, was found to account for blindness prevalence ranging from 1% in East Africa, to 4-16% in West Africa, thus demonstrating wide regional differences, although the reasons for this are unclear ¹²⁰.

Measles remains a major problem in Africa and many other developing countries. It affects 30 million children a year, causes up to 1 million deaths annually and is the single leading cause of blindness among children in low-income countries ²⁴¹.

9.16.3 Childhood Blindness in Nigeria

Olurin in Nigeria in 1970 ¹²⁴, in a study of 140 children (age range 2 months to 14 years), found the major pathology to be keratitis (21%) of which the largest cause was measles. This was followed by cataracts (19%), (only 5 of the 26 cases being genetic in origin), and optic atrophy (14%; all due to acquired causes). Thirty-three years later Ezegwui et al ²⁴² studied 142 resident of 3 schools for the blind in southeast Nigeria. Childhood factors from corneal scarring, predominantly as a result of measles, have remained a major cause of blindness in the country. It is interesting to note that hereditary factors have increased from 12.7% in the 15 years plus cohort, to 19.6% in the <15 years cohort. However, in another study in 2002, by Akinsola et al ²⁸³, of 26 children below the age of 16 years with visual impairment in a blind school in Lagos, Nigeria, there were very few cases of corneal scar (measles-/VAD). Hereditary conditions formed 38.5% of the cases, intrauterine factors 23.1%, other causes 15.4%, and in 23% the cause was unknown. Retinal dystrophy was the most common cause of low vision and blindness, while congenital cataract and glaucoma were the major causes of avoidable blindness. Anatomical sites of diseases leading to low vision and blindness in

these children were retina (30.8%), lens (23.1%), glaucoma (19.2%), cornea (11.5%) and optic nerve (7.7%).

9.16.4 Childhood Blindness in Malawi

Chirambo's study¹²⁵ in 1976 of 270 students attending 17 blind schools in Malawi, found that 73% of the total number were blind before the age of 3 years. The most common cause for the blindness was ocular infections (75.2%). Measles as a single cause was responsible for 43.7% of the cases, and smallpox for 5.2%. Bacterial infections were incriminated in 26.3% of cases. Hereditary factors as causes of blindness were found in only 7.8% of the cases. These included congenital cataract (2.6%), optic atrophy of unknown origin (3%), and microphthalmos (1.5%). Similar findings were reported by Benezra and Chirambo in 1977 who estimated the prevalence of blindness in children < 5 years of age as 0.34/1,000 children. Direct ocular infections were responsible for blindness in 32% of the cases (bacterial infections 20% and measles 12%)⁴⁴⁷. Traditional eye medicines (TEM) added to the severity of the ocular morbidity, leading to total melting of the cornea and protrusion of the uvea in all cases. Congenital conditions (excluding retinoblastoma) formed 30.8% of the total, 11 of these were cases of CC. A high incidence of cortical blindness (9.3%) was noted. Trauma was implicated in 5.3%, while retinoblastoma was found in 8%.

All cases of bilateral blindness in this age group in Malawi in 1983 were considered to be due to VAD. The disease was not only a leading cause of blindness in this area, but may have an important impact on child survival as well⁴³⁹.

9.16.5 Childhood Blindness in Central African Republic (CAR)

The causes of blindness in the CAR in 30 children were; corneal scarring (17%), congenital cataract and optic atrophy (13.5% each), iritis and microphthalmos (10% each), glaucoma (7%), and trachoma and phthisis bulbae (3.5% each)¹⁷⁶.

9.16.6 Childhood Blindness in Ethiopia

This is a country with a low socio-economic status in which children make up almost 50% of its 65 million inhabitants. An U5MR of 176/1000 live births is an unacceptably high figure, even by sub-Saharan standards⁷⁷. A study covering 3 blind schools visited in 2001 found that the causes of CB were attributable mainly to VAD and measles (49.8%); whilst hereditary disease, such as retinal dystrophies accounted for only 3.4%⁷⁷. It was estimated that 70% of blindness was due to either corneal opacity or phthisis bulbi; and in 40% of patients with non-congenital bilateral corneal opacities or phthisis bulbi, which was preceded by measles²⁴⁵.

9.16.7 Childhood Blindness in South Africa

O'Sullivan in 1997¹²⁸ documented hereditary causation in 33% of the 1,311 children examined whilst acquired conditions accounted for only 11.5%. In 41.5% it was not possible to determine the underlying cause, although it was estimated that in 38.8% of those studied the cause was avoidable. There were major variations in causes between the different ethnic groups, the major difference being the higher proportion of ROP in white and Indian children.

9.16.8 Corneal Scarring in Children in SSA

There has been controversy over the major causes of corneal scarring in Africa, with debate centring around VAD; keratitis caused by human alpha herpes virus 1 or 2; the use of TEM; measles; and exposure keratopathy. There is a close correlation between measles and VAD that can result in xerophthalmia, with corneal ulceration, keratomalacia and subsequent corneal scarring or phthisis bulbae. Herpes simplex keratitis predisposed to 20% of measles related corneal ulcer in Tanzania¹³⁴. The importance of these conditions varies geographically and temporally depending on famine, measles epidemics and the availability of eye services¹²⁰.

The pathogenesis of corneal scarring and the ensuing blindness in Africa has been well explained in the literature^{120, 134}. Whatever the cause, corneal ulceration leading to corneal scarring was found to be responsible for 70% of childhood blindness and 1-4% of African children with acute measles

will develop corneal ulceration. In Tanzania 84% of bilateral corneal ulcers were found to be due to VAD ¹²⁹ and in northern Kenya, corneal scarring from xerophthalmia (and trachoma) was the main cause of blindness not only in children but also up to the age of 35 years ¹³².

9.17 Diversity Of Childhood Blindness In Asia

Asia is a vast and diverse continent whose countries fall in 5 different categories of the World's Bank ranks: the EME (Japan), India, China, OAI and the MEC which is being echoed in the causation of CB ^{115, 139}.

The lack of blind registration create difficulties for assessing the prevalence of CB in Asia together with the significant scarcity of surveys of blind schools. The socio-economic diversity in this continent produces a vivid mix of causations ranging from those seen in Europe to those seen only in the poorest countries such a nutritional blindness, as is the case in many parts of South East Asia. In the middle, there are many countries that have overcome the odds and achieved good progress in combating this problem, such as Sri Lanka, Myanmar, and Korea ³²³. The magnitude of VAD is so great with 83 million school-aged children (23.4%), of whom 9 millions (10.9%) were found to have mild xerophthalmia (night blindness or Bitot's spots). Fortunately, potentially blinding corneal xerophthalmia is negligible in these children ²⁴⁰.

9.18 Childhood Blindness In India

9.18.1 Prevalence

In India, an economic entity group in its own right ^{115,139}, estimates of the number of children with SVI/BL in 1995 was at least 200,000 from a total blind population of the world of 5 millions. Of these, approximately 15,000 were in schools for the blind ¹⁴² confirming that only 5-10% of blind children in India are in special educational establishments ¹⁴⁰.

Significant interstate variations were recorded in the prevalence of SVI/BL ranging from 7.5% in Kerala to 26.7% in Madhya Pradesh. In addition, major differences were found between urban and rural locations where the percentage was 7.5% in the capital city blind school compared with 30.4% in a blind school in a rural area of the Tamil Nadu state ³²⁸. In

two population based studies of 6935 children at or below the age of 15 years in the southern Indian state of Andhara Pradesh, the prevalence of SVI/BL was found to be 0.17%³²⁶.

9.18.2 Causes of Childhood Blindness in India

The causes of CB in India appear to have changed little in the span of 28 years since Jain's study in 1968¹⁴³. In a more recent study by Sil and Gilbert in 2001¹⁴⁰ of 2,283 children in 11 Indian states, 50% of cases were still either preventable or treatable. The single most important disease causing SVI/BL in India is VAD, affecting 18.6% of children³²⁸.

Anterior segment pathologies remain the major cause of blindness. A study in 1993 found that corneal scarring/phthisis bulbi formed 38.4% of the cases of SVI/BL followed by retinal causes at 22.6%. The latter was mainly made up of retinal dystrophies (21.3%).

Hereditary factors account for 26%¹⁴⁰ to 29.8%⁴¹. Retinal blindness formed 22.6% of the causes of blindness and was made up mainly of retinal dystrophies accounting for between 60-75% of hereditary causes (21.3%). Congenital eye anomalies were another important cause³²⁶.

This trend was confirmed by another study 2 years later where the major causes of SVI/BL were corneal staphyloma, scar and phthisis bulbi (mainly attributable to VAD) 26.4%; microphthalmos, anophthalmos and coloboma, 20.7%; retinal dystrophies and albinism 19.3%; and cataract, uncorrected aphakia and amblyopia 12.3%¹⁴².

9.18.3 Avoidable Blindness

Of the 50% avoidable blindness in India, preventable causes formed 30% of cases and treatable 20%¹⁴⁰. Refractive errors are an important cause and formed 33.3% of cases in the state of Andhara Pradesh, 16.6% of which were due to preventable causes (8.3% each due to VAD and amblyopia after cataract surgery)³²⁶. Priorities for action to reduce CB in the context of Vision 2020 were considered to be refractive error, cataract related amblyopia, and corneal diseases³²⁶. It has been suggested that this mixed pattern puts the population in India in an intermediate position between the developing countries and industrialised countries¹⁴⁵.

9.19 Childhood Blindness in Bangladesh

Bangladesh, known as earth of mud and water ¹⁵³, is one of the poorest countries in the world where in most parts diarrhoeal diseases are prevalent. In a survey conducted in 1976 of a few union councils of the districts of Chittagong, Comilla and Noakhali, in a population of 114,624, 1,085 cases of blindness were identified (243 children and 842 adults).

The most common cause of blindness in children was corneal infection (cataract in adults), followed by trauma in 26.7% of children versus 10.2% in adults ¹⁴⁷.

There has also been an emphasis in the research on evaluating the prevalence and magnitude of nutritional blindness from VAD in Bangladesh ^{79, 147, 148}. A prevalence of 1.09/1000 and 0.64/1000 in the urban and rural population respectively was found in a nationwide survey conducted in 1985 on 22,000 children aged 3 months to 6 years in rural and slum urban areas ⁷⁹. Night blindness combined with conjunctival xerosis and Bitot's spots were present in 1.69/1000 person and all combined stages of active xerophthalmia were seen in 0.06/1000 ¹⁵¹. Night blindness alone was found in only 0.03/1000 and males had a significantly higher prevalence than females at 2.9/1000 and 1.2/1000 respectively. The rate of keratomalacia scar was 0.31/1000, whilst corneal opacities from other causes were seen in 2.25/1000 populations.

An intervention programme was conducted between 1986 and 1989 in the northern Ranjpure district of Bangladesh to reduce the morbidity of nutritional blindness. The efficacy of the programme was evaluated in two cross-sectional studies ¹⁴⁸ in 1986 and 1989 and concentrated on children <9 years of age. A reduction in the prevalence of night blindness from 50.7% to 26.7% was found over the period. However, the post intervention prevalence varied between areas. There was no statistically significant difference in the sex ratio. Higher father income, higher mother literacy, and smaller family size were associated with low prevalence of night blindness in the 1989 cross-sectional study.

9.20 Childhood Blindness in Sri Lanka

The pattern of childhood blindness seen in Sri Lanka was typical of a growing number of South East Asian countries undergoing rapid development, where VAD was not a significant cause of visual morbidity^{175,144}. In a study of 226 children with blindness or severe visual impairment, cataract was found to be responsible for 17% and was the commonest avoidable cause of SVI/BL. Bilateral microphthalmos accounted for one-quarter of those examined¹⁷⁵.

9.21 Childhood Blindness in Nepal

Nepal is another poor country in Asia where VAD is common and a major cause of blindness and loss of vision among children in the eastern plains of country¹⁵⁴. Active or past xerophthalmia was found in 15.4% of 4601 children aged 0-10 years by Hennig et al¹⁵⁷. The peak age for active corneal xerophthalmia was 3 years and that of non-corneal xerophthalmia was 5 years. Prevalence of blindness in children under 15 was found to be 0.63/1000 children in a nationwide survey reported in 1988⁷⁹. Trachoma is another ocular disease that is prevalent in Nepal, especially the follicular type. Intense inflammatory trachoma was relatively rare and scarring was absent in the preschool population. It was concluded that the population studied were not at high risk of repeat infections leading to blindness in adulthood¹⁵⁸.

A survey in the only available school for the blind in Eastern Nepal was conducted by Panda et al in 1999^{243, 244} and reported that 41.25% of causations were retinal in origin. Avoidable conditions formed 46.25% of cases, with 38.75% preventable and the rest treatable. Lenticular causes formed only 4% of the eye conditions.

9.22 Childhood Blindness in Indonesia

The only study available from Indonesia was that of 16 students in a school for the blind in Bandung in 2003; 96.4% of the students were blind and 3% severely visually impaired¹⁵⁵. The main causes of SVI/BL were corneal pathologies (staphyloma, scarring and phthisis bulbi) followed by retinal dystrophies at 20.6% (mainly LCA and early onset RP). Congenital cataract formed 13.3% of the series and microphthalmos/anophthalmos

10.9%. The whole globe was the main anatomical site of primary pathology (32.7%) followed by the retina (20.6%), cornea 17.6%, lens (13.3%), optic nerve (6.1%) and uvea (4.3%). Hereditary and infective causes constituted 42.4% and 29.7% respectively.

Ocular toxoplasmosis was found to be a common sight-threatening disease in Indonesia among children in a laboratory study evaluating 41 children under the age of 12 years, 56 of whom were blind ¹⁵⁶.

9.23 Childhood Blindness in Malaysia

A study by Reddy in 2001 in Malaysia found that hereditary causes were responsible for 29.5%, whilst intrauterine factors and perinatal factors were responsible for only 4.5% and 9% respectively. The aetiology was unknown in 49.1% of cases which included congenital anomalies of the globe. The author concluded that perinatal screening for intrauterine factors and hereditary diseases would help in reducing the prevalence in this region ¹⁶⁰.

9.24 Childhood Blindness in Thailand and The Philippines

Gilbert and Foster in 1993 examined 256 children in schools for the blind in Thailand (1 school) and the Philippines (3 schools) ¹⁵⁹. The causes of visual loss were similar in both regions with perinatal factors accounting for 20% and 23%, and hereditary disease 13.8% and 17.7% in Thailand and the Philippines respectively. In the total study, 15% of children were blind as a result of ROP and 9% from corneal scarring attributed to VAD. A total of 58% of children were suffering from avoidable causes of visual loss.

An earlier study of 129 students between the ages of 3 to 19 years (the predominant age group being 6-15 years) from the School for the Blind in Khon Kaen, Thailand found that pathologies affecting the whole eye were the leading cause of blindness (36.2%) and that nearly half of the cases of blindness were preventable or treatable. Only 26 students (20.5%) had positive family histories. The M/F ratio was 2.63:1. Of all the students seen, 26 students (20.5%) were suitable for treatment that succeeded in improving vision in 42% of them ³³⁰.

Vitamin A deficiency has been evaluated in the above two countries in separate studies ^{165, 329}. Vitamin A deficiency was considered an important

public health problem by Bloem et al in rural Thailand ¹⁶⁵, as demonstrated in the prevalence of night blindness, Bitot's spots, and deficient serum retinal levels. The prevalence of night blindness was 1.3% and Bitot's spots 0.4% in children aged 1-5 years in rural areas, while this pathology was absent in urban regions. Of the children between 1-8 years, 9.6% had deficient retinal levels. In the Philippines ³²⁹ a survey of VAD was conducted in 1991 on the island of Mindanao on 248 preschool children in five randomly selected rural communities on the outskirts of Davao City. Subclinical levels were found in 29% and clinical levels in 6%. A recent history of diarrhoeal disease, reported night blindness, maternal education less of than 9 years, and infrequent consumption of eggs, mangoes, and liver were associated with an increased risk of VAD.

9.25 Childhood Blindness in Vietnam

In Vietnam, trachoma and xerophthalmia are very common and ROP is emerging as a cause of visual morbidity. The risk and severity of ROP appears to be higher in larger and older infants in Vietnam than the rest of the world ³²⁵.

9.26 Childhood Blindness in China

9.26.1 Epidemiological Studies

China is grouped separately in the economical categorisation of nations ^{115, 139}, and studies from it are scarce. One by Hu ¹⁶¹ in 1987 revealed the results of a mass screening of 700,000 people with more than 5,000 pedigrees of genetic eye disease. Unfortunately this did contain a breakdown of the age groups studied. It is interesting to note however that as early as 1978 a clinic for genetic eye disease had been established with a genetic counselling service.

9.26.2 Causes of CB in China

Another study conducted in 1999 of 1245 blind school student between the age of 5 to 15 years from 36 blind schools in 18 provinces by Hornby et al ¹⁶³, found 91% of the students in the SVI/BL WHO category. The commonest anatomical sites for visual impairment were the whole globe

(essentially microphthalmos) in 25.5%, retina (mainly retinal dystrophies) in 24.9%, lens in 8%, optic nerve in 13.6%, and glaucoma in 9%. Corneal conditions were not a significant cause of visual morbidity. Hereditary factors contributed to 30.7% of cases and childhood causes to 14%. An earlier study in 1992 reported that hereditary conditions formed 84.46% of the causes of blindness and low vision in China ²⁶⁴.

A similar pattern was demonstrated in a more recent study undertaken in 2002 by Shi-Yu and Zuomin-Xu who assessed 385 children attending seven blind schools in Eastern China between April 1998 and May 1999 using the WHO examination record. Of the 385 children examined 356 (92.5%) were blind or severely visually impaired with the commonest anatomical sites being the lens (27.5%), followed by the retina (22.5%), and the whole globe (15.2%). Hereditary factors were found in 35.1% followed by prenatal factors in 6.2% (ie drugs or alcohol taken during pregnancy) and meningitis in 5.3% of cases ¹⁶².

9.26.3 Avoidable Childhood Blindness in China

Avoidable blindness in China ranged between 36.5% ²⁶⁴ to 46.5% ¹⁶² of cases of blindness. Potentially preventable cases formed 15% ²⁶⁴ to 23% ¹⁶² and potentially treatable 22.5% ²⁶⁴ to 24.4% ¹⁶². These results reveal that this country has gained from improvements in health and socio-economic status so that nutritional and infective causes of blindness are uncommon and hereditary factors are very much to the fore.

It was concluded that the results reflected the improved health and socioeconomic status of China, although it was felt that it might also reflect a bias, with higher socioeconomic students over represented as a result of a bias in admission to, and location of blind schools. The study demonstrated a shift from nutritional infective causes to hereditary and unknown factors in childhood blindness ^{162, 264}.

9.26.4 VAD

The prevalence and spatial distribution of VAD in China among children under six years of age were addressed by Lin et al ³²⁷ who studied 8,600 children under 6 years of age in 14 cities and 28 counties of 14 provinces

using a stratified cluster sampling for survey, including interview with questionnaire for their family information and nutritional status. Vitamin A deficiency was found to exist in children, especially in the remote and poverty-stricken rural areas of China and it was recommended that vitamin A supplementation was urgently needed for the children in these regions. In this survey, 8 cases of night blindness and seven cases of xerophthalmia were found among the children at ages of 2-5 and 61 mothers of the children in this group were also found to be suffering from night blindness. All the cases of night blindness and xerophthalmia both in children and mothers were living in rural areas.

The prevalence of VAD was 15.0% in the rural areas and 5.8% in urban areas, in contrast to 23.3% in the poverty-stricken counties. In the coastal, inland and remote areas, the prevalence was 5.8%, 11.5% and 16.8% respectively. There was a significant difference in serum levels of vitamin A between ages, but no significant difference between genders. Babies under six months of age accounted for 33.4%, and those at ages of 4 - 5 years for 8%.

9.27 Childhood Blindness in Mongolia

In Mongolia a study by Bulgan and Gilbert found a prevalence estimate that was lower than anticipated together with a pattern of SVI/BL similar to that found in blind school children in China and very different from other Asian countries¹⁶⁴. A total of 64 students were examined who had been referred from a variety of sources in 10 of the 18 provinces of Mongolia. The source of referral was; family doctors (52); home visits (3); hospital records (4) and schools for the blind (5). Lesions of the lens were the most common cause (34%), central nervous system disorders (19%) and retinal conditions (12.5%). Whilst 27% of causes were hereditary, 17% of children were blind following acquired conditions, meningococcal meningitis being the most common preventable cause. Other preventable causes were rare and the authors concluded that better management of conditions requiring surgery and low visual aids were necessary.

9.28 Childhood Blindness in Japan

9.28.1 Prevalence

Japan is the only Asian country which belongs to the EME rank of countries)^{115, 139}. The two studies by Nakajima¹⁶⁶ and Tsukahara⁹² in Japan have shed light on CB in that country. Nakajima's study¹⁶⁶ in 1982 revealed that the prevalence of blindness among school children dropped from 0.65/1000 in 1900 to 0.08/1000 by 1980.

9.28.2 Changing Pattern Role of Consanguinity

Blindness from infection and malnutrition were replaced by congenital causes.

One of the major differences in Japan was the role of consanguinity. According to Nakajima, the rate of first cousin marriage in 1950 was 5%; this had dropped to 0.1% by 1980.

Tsukara's study⁹² in 1985 of 67 pupils at Yamanashi Blind School, found 5 of the pupils were the product of first cousin marriages and there was a history of consanguinity in 12.2% of those blind from hereditary causes. This study found that hereditary causes were very probable in 41.8% of cases; probable in 10.4%; and possible in 20.9%. This broke down into an incidence of Autosomal recessive inheritance being very probable in 42.9%; probable in 14.3%; and autosomal dominant in 10.2%. When compared to Phillips⁹¹ and Fraser's study, the pattern of hereditary diseases was fairly similar with cataract being the main followed by RP, choroidoretinal degeneration and ROP. There were no cases of blindness associated with multiple handicaps in Yamanashi School.

9.29 Conclusions on Interpreting Data from Blind Schools Surveys

Whilst examining the above figures, it is necessary to point out that in some developing countries, only about 10% of blind children are in blind schools and therefore the surveys of blind school pupils are potentially biased. This must of course be balanced against the advantages of one examiner using a standard method and examining a large number of children in a relatively short period of time¹⁴¹.