3

Blindness: Epidemiological Methods

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3.1 Epidemiological Studies on Blindness

The necessity of epidemiological studies into the causes and incidence of blindness, both nationally and internationally, has long been recognised as a pre-requisite for both the future planning of the care of the blind, and the furtherance of scientific understanding of this distressing handicap. Furthermore, given our current level of knowledge, without new and targeted prevention and treatment programmes, the numbers of blind (45 million worldwide), will double in the next 20 years. (1)

3.2 Data Collection on Blindness

Since the last century, epidemiological data on blindness and visual handicap have usually been collected by a mixture of governmental, non-governmental, medical and charitable agencies. Prior to that time, periodic census returns were the only source of statistics on the blind and this method, together with the lack of an adequate definition of blindness, added to the difficulties inherent in such works. (2)

3.3 Definitions of Blindness

Visual Acuity as a Measure

The use of visual acuity and fields of vision as a means of creating a coherent definition on which to base policy, originated during the 1930s in the USA where unemployment was high and there was concern for the welfare requirements of the visually impaired. As a consequence, a committee of the Section of Ophthalmology of the American Medical Association was appointed to develop a scientific definition of blindness suitable for the development of statutes and several definitions of blindness were created in 1934. (2)

WHO Definition of Blindness

The WHO also addressed the variations in the definition of blindness and visual impairment and set up a standard definition of blindness in 1993. Under this, blindness was defined as: *'A visual acuity of less than 3/60 (20/400, 0.05), or a visual field loss in each eye of less than 10º from fixation.'* Whilst low vision was defined as: *'visual acuity of less than 6/18 (20/60, 0.3) but equal to or better than 3/60 in the better eye with best possible correction (corresponding to visual impairment categories 1 and 2 in ICD-10)* (equivalent to current categories 2 and 3 respectively). *Category 1* (equivalent to current category 2) *is visual impairment, less than 6/18 to 6/60, and category 2* (equivalent to current category 3) *severe visual impairment, less than 6/60 to 3/60'*. (3)

3.4 Blind Registration

In 1935 a definition of 'economic blindness' was modified and accepted as an administrative aid in blind programmes and subsequently spread to most industrialised nations. The definition for blindness used at this time was 'Blindness in the US is defined as 'visual acuity, in the better eye with correction, of not more than 20/200 or a defect in the visual field so that the widest diameter of vision subtends an angle no greater than 20 degrees'. (2)

In the UK, prior to the introduction of statutory benefits payable to the blind under the Blind Persons Act of 1920, the recording of blindness had been through a simple declaration of blindness on the census returns. Registration, which required a certificate from any medical practitioner, had a poor take-up initially but whilst early registration was poor, the statutory benefit payable involved examination by an ophthalmologist after 1950, with a diagnosis and a rigidly enforced rule of 3/60 visual acuity irrespective of age or other factors. (4)

It was Sorsby who wrote in 1950 that 'the lack of an adequate definition of blindness is paralleled by the lack of uniform methods for the enumeration of the blind.' He believed that the explanation for conflicting tendencies in census returns was due to different criteria being used in different censuses and believed that it demonstrated the unsatisfactory nature of census returns as a measure of the frequency of blindness. These problems were solved in part by blind registrations although this was confined to a few countries only and the majority continued to rely on the census returns. (5)

This inadequacy remained until very recently and, as late as 1989, Johnson and Minassian reported that there were still at least 65 different definitions of blindness and visual impairment in use throughout the world. This was despite the fact that a WHO study group in 1972 recommended a standardised method of testing to give a uniform definition of blindness and visual impairment so that global comparisons could be made. (6)

In the UK, the introduction of statutory benefits payable to the blind in 1920 also led to registration but this had a poor take-up initially. After 1950, registration involved examination by an ophthalmologist and included a diagnosis. (5)

A DHSS bulletin of 1988 however pointed out that because of the varying degrees of response, the number of blind registration forms (BD8s) available for analysis of causes in any one year did not necessarily equal the total number of new registrations in that year as notified through SSDA 902 form. The fact also remained that not all persons who were blind or partially sighted chose to be registered for reasons such as the stigma of being registered as blind, or the fear of losing employment, so that the true prevalence was therefore higher. (7)

Pitfalls of BD8 Certification

The pitfalls of BD8 certification in the UK have been assessed in several studies from 1986 to 1998. (4), (3), (8), (9) It was estimated that whilst 90% of eligible blind people in the UK were registered, only 50% of the partially sighted eligible were registered. (3) The degree of under registration in the UK is unknown but the RNIB estimated that out of every 100 blind people, 64 were not registered and 87 out of 100 partially sighted people were not registered. (4) The problem of the cause of visual disability recorded by the ophthalmologist is not always adequately transmitted into the statistical analyses. (8) Data collection during registration can be biased towards permanent, non-treatable causes of visual loss and those which affect central rather than peripheral vision leading to under certification of certain subgroups of the visually impaired. (9) It has been suggested that a numerical system replace the currently used terminologies (containing the word blind) or even using the term 'visual impairment' may be a workable alternative. (10)

Trends in Blind Registration in the UK

Despite this, the numbers of new registrations for blindness have risen considerably over the years between 1965, 1975, and 1985. (11) This was attributed to both changes in the age structure of the population and under-registration in 1965.

The increase in new registrations for partial sight was significantly more than would be expected, even after changes in the population structure are allowed. Registration rates for macular degeneration and glaucoma are increasing in both males and females, and rates for cataract are at a significantly higher level for women than for men. (11) Of all registrations in Bradford, 64% were found to be blind and 36% were partially sighted. (12)

Blind Registration in Other Countries

In addition to the anomalies of registration in some countries, or lack of it in other parts of the world, different countries continued to use different definitions for blindness for their own legal and social purposes.

In Canada, Hameed found that health administrative databases, despite containing a wide cross-section of diagnoses, were usually limited in systematic information and that more comprehensive validation of large registers was needed to provide the foundation for a longitudinal ocular surveillance system. (13)

In Norway and Finland, registration has become compulsory, making registration data more accurate in these countries. Accurate data has also been found in some European countries with small populations such as Greenland and Iceland. (14) In addition, in Sweden, an elaborate special register for children in the Nordic countries was set up in 1990. The individual record contains information on sex, year of birth, year of registration, classification of visual impairment, ocular diagnosis, systemic diagnosis, aetiology and additional disorders. There is also a coding system for additional impairments. (15)

3.5 Problems in Data Collection Observer Bias

Data collection remains problematic and despite the advantages of blind registration data in industrialised countries, this source of data is subject to both observer bias and selection bias. Whilst using the same International Classification of Diseases for coding, different ophthalmologists will not necessarily code the condition in the same way. (16), (17)

Variations in Definitions

Variations in definitions also made it extremely difficult to compare epidemiological studies of the visually impaired. Added to this are the ongoing ways in which data are collected; European data are derived mainly from blind registrations whilst those from developing countries come mainly from surveys. Given that most population-based surveys try to assess the prevalence for a whole population, and as blindness rates increase with age, such surveys are usually too small to give accurate estimates for children. (17)

Under Reporting

Worldwide, Foster and Gilbert have pointed to the dearth of data available from population-based studies. They believed that the prevalence and incidence data from blind registers usually underestimated the true figures because of under-reporting. (18)

Similarly, in developing countries where blind registrations are not available and prevalence data is derived from population-based surveys, (usually of blind institutes), not all blind children will be in schools for the blind and so such surveys do not represent the total blind child population. This is in addition to the lack of reliable methods of assessing visual acuities, the exclusion of children in residential schools, and selection bias. (17)

However, epidemiological data are becoming more standardised but comprehensive worldwide studies for the purposes of comparison are still sparse.

3.6 Data Collection in Children

Different Diagnostic Criteria

Changes in our knowledge of the aetiologies of disease have also produced dissimilarity, and relatively more difficulties in comparing changes in the pattern of disease. At an international level, the WHO directed attention to data collection on blindness following the recommendations of the 25th World Health Assembly in 1972 and this has provided the earliest epidemiological information on the magnitude and pattern of blindness globally. The methods used to collect these data in population surveys in many countries were different which led the WHO to produce guidelines and protocols for this purpose in 1978. (19) Standardized protocols for collecting data for use in a simplified grading system for community-based trachoma surveys and childhood blindness were later produced. (17), (20), (21), (16), (17), (6), (19), (20), (21), (22), (23), (24), (25), (26), (27)

Multiple Handicaps

Those with multiple disabilities, including blindness, are less likely to be registered and the causes will be weighted in favour of conditions not associated with additional handicaps. In addition, blind registration does not take into account those who have died after being registered. There is an obvious absence of clearly defined criteria for registering children. (16), (17)

Visual Improvement and Delayed Visual Maturation (DVM)

A relevant point in evaluating CB is the observed possible improvement of visual functions in visually disabled children with increasing age. This was first 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, 13% were deceased, while in 23% the vision had

improved to 0.3 and above (6/18). 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. (22), (23)

WHO Recommended Methodology in Children

On childhood blindness, the WHO recommended a standard methodology of reporting causes of childhood blindness based on the two criteria of anatomical site of abnormality and underlying aetiology. (20) Only more recent studies have used this approach thus making it difficult to compare previously available data. The pitfalls of the blind register in England and Wales are well recognised for several reasons including; their lack of defined visual criterion for registration of children, data do not take into consideration children who had died, the difficulty in determining the level of visual acuity in younger children, and the delayed onset of visual impairment in some conditions until a later age. (17) Most cases of visual impairment in children do not appear on the statutory blind or partially sighted register which is another deficiency that makes these registers of limited value for service development. (24), (25)

3.7 Global Perspective

There are an estimated 1.5 million blind children worldwide (28) with an additional 5 million visually disabled. (29) Among those, 90% live in the developing countries, (30) with some 1.3 million living in Asia and Africa (31) and some 72,000 of them in Europe, the USA and Japan. (27)

Approximately one child goes blind every minute in the world, (28) making a total of 500,000 children who go blind each year worldwide, 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. (30), (32) In the UK, this figure is 10%. (33) In Africa, the prevalence of infant and childhood mortality (and also childhood blindness) is the highest in the world although this has shown a decline in recent years. (32), (34)

3.8 Impact of Childhood Blindness

The impact of blindness in childhood is enormous and is summed up in Table 3.1.

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 the childhood population at 6,849 person-years, compared to 270 for diabetes and 430 person-year for age related macular degeneration. (35)

Table 3.1 The impact of blindness on childhood (Adapted from Gilbert: Eliminating Childhood blindness) (25)

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

3.9 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. (36), (37), (38), (39), (40), (41), (42) 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. (33), (43), (44), (45), (46), (47), (48)

Male predominance has been attributed to several factors, (Table 3.2), although in many instances it remains inexplicable or has not been addressed by the authors of these studies. (36), (37), (38), (39), (40), (41), (42) 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 1968, (49) when the M:F ratio was 14-fold higher in boys (12.8:1), to the current ratio of 1.7:1. (41) Male favouring in blind registration was put forward by Schappert-Kimmijserin in the Netherland, (50) but this has been questioned in recent studies from the same country (51) and discarded in other countries. (40)

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 (39)' (52) in both the genetic and non-genetic cases. (52) 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, choroideraemia, X-linked juvenile retinoschisis, Lowe syndrome and X-linked cataract. Genetic factors (X-linked) are found especially in the Danish and Finnish groups. (52) (Table 3.3) The significant prepond-erance of males found in the prenatal asphyxia and prematurity categories 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. (52) The same applies to optic atrophy and perinatal lesions, which may be due to X-linked disordersinducing pre-term birth and perinatal asphyxia. (53)

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.

Table 3.2 Causes of Male Predominance in Childhood Blindness

- Social factors favouring boys education
- Inclusion on the register [223 (Rosenberg T)]
- X-linked conditions [90, 96, 135]
- Perinatal affections [90, 96, 135, 669]
- Cataract with perinatal difficulties [90]
- Optic atrophy [96, 135]
- Buphthalmos [90]
- Unexplainable [97, 669]

Table 3.3 Male predominance in relation to aetiological categories

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

3.10 Blindness and Other Disabilities

Mental Retardation in the UK Cohorts

Mental retardation was the most commonly defined additional impairment, being present in 29 (27%) of those examined and many of the children had multiple impairments due to brain injury e.g. cerebral palsy, mental retardation 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. (33)

Associations 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, hear-ing 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. (15)' (39)

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