Prevalence of blindness and eye disease: discussion paper

G J Johnson MD FRCS(C) **D C Minassian** FRCS MSc *Institute of Ophthalmology, 27/29 Cayton Street, London EC1V 9EJ*

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Introduction

Prevalence is a measure of disease frequency. Prevalence (or point prevalence), is the number of people with a certain disease existing at a given time. This number is related to the number of persons at risk and is usually expressed as a proportion (period prevalence, an infrequently used measure, is the number of people with a certain disease at a given time plus all the new cases of the disease that occur during a certain period of observation relative to the persons at risk). For example, in the ophthalmic survey conducted in Framingham, Massachusetts, among individuals 52-85 years of age, there were 310 of the 2477 persons examined who had cataract at the time of the survey. The prevalence of cataract in that age group was therefore 310 per 2477, or 12.5%.

Incidence is the number of illnesses or cases of a condition that occur within a specified period of time; for example the incidence of open angle glaucoma in a defined population in 1985 would be the number of people in that population who were newly diagnosed as having glaucoma during 1985. This could be expressed as a percentage or per 1000 of the population at risk.

The difference between these terms prevalence and incidence perhaps needs further emphasis because they are frequently confused. Prevalence refers to all cases in the population, new and old, whereas incidence refers only to new cases.

Prevalence is a proportion. The term 'prevalence rate' is frequently loosely used, as is 'interest rate' in financial matters. Strictly speaking 'rate' is a measure of speed of change[1]. Incidence is also usually a proportion but it can be measured as a rate.

Prevalence is usually regarded as being an inferior measure of disease frequency. It depends on both incidence of the disease and survival from the disease, creating the paradox that diseases with a high incidence and high mortality have lower prevalence.

However, when a disease is chronic, does not greatly affect life expectancy and has a stable incidence, prevalence gives a useful indication of both incidence and the burden of the disease in the population. Therefore, prevalence is a particularly useful measure of blindness. The exceptions are in special cases where blindness affects life expectancy as in children in rural Africa or conversely when the disease causing blindness also affects life expectancy as in proliferative diabetic retinopathy.

Why is prevalence important?

In order to attempt any preventive measure against a disease, some knowledge of the distribution of that disease in the population is essential, if only to be able to assess the impact of intervention. It is inadequate to sit in a hospital and expect all the cases of the disease concerned to attend the hospital. Estimates of prevalence thus determined may be seriously biased either way. Experts in specialized areas will attract cases from far and wide creating a mistaken impression of increased prevalence. Alternatively, prevalence of a disease which is associated with being unable to leave the house will be seriously underestimated.

Once prevalence has been established, further investigative epidemiological research can be undertaken to discover disease associations and causes. Recalling and re-examining the same sample after a period of time converts prevalence to incidence.

How can we measure prevalence?

The only fully accurate way of determining the prevalence of blindness or eye conditions in a population would be to examine every individual. For a small village or community this is often possible, but clearly it is not feasible for a whole region or country.

The alternative is to take a *sample*, which must be as representative as possible of the whole population. In order to be able to extend statistical conclusions from the sample to the whole population, the sample must be taken according to strict principles. Each individual or cluster of people in the whole population must have an equal or known chance of being included in the sample.

Example: the Malawi survey

In the autumn of 1983 a random population-based survey was conducted in the Lower Shire Valley of Malawi[2]. This was a joint project of the Government of Malawi, and the International Centre for Epidemiologic and Preventive Ophthalmology in Baltimore, with the support of several voluntary agencies.

The design of the survey was rather simple. The information required was the prevalence and causes of blindness in the rural population, so two urban areas were excluded. The remaining 694 villages, with a population of 226 000, comprised the sampling frame. These were arranged geographically and numbered, and a computer selected the sample of 60 villages at random.

All the children under six years of age were examined in these 60 villages, and the entire population in a sub-sample of 10 of the villages, using three teams of health workers, each led by one ophthalmologist. Socio-economic and nutritional data were collected about each village, each household, and each individual.

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No-one informed the computer whether all the villages chosen were accessible by ordinary means of transport. One was on an island in the middle of the river. Others could not be reached by roads. But to have exchanged these difficult places for villages near the main road would have introduced bias into the sample.

In the children the teams were looking particularly for the grades of trachoma infection and signs of xerophthalmia. It was necessary to standardize the observers at the outset, and then re-standardize at intervals to avoid diagnostic drift. The same children were examined by all three observers and then re-examined in a different order.

From these results the intra-observer and inter-observer agreements could be calculated, expressed as a Weighted Kappa Statistic[3]. Kappa is a measure of agreement achieved beyond chance, relative to the maximum possible agreement beyond chance. Thus a Kappa of 1 indicates perfect agreement. When there are more than two possible categories or grades the Kappa can be weighted according to seriousness of the disagreement[4].

Results

The coverage among those aged 6 years or over was 87% (1664 persons examined), and below six years more than 99% (5436 children examined).

The overall prevalence of blindness, (defined according to the World Health Organization criterion of best corrected central vision less than 3/60 in the better eye), amongst those aged six years or over was 1.27% with a 95% confidence interval (CI) of 0.76 to 1.96% (Table 1). This means that the prevalence of blindness in the sample was 1.27% and there was a 95% probability that the true prevalence in the total population was within those limits; however, there still remained a one in twenty chance that the true figure could lie outside those limits. The three main causes of blindness were cataract (prevalence 0.57%), corneal scarring (0.38%) and glaucoma (0.19%). They account for 40%, 30% and 15% respectively of all blindness. These have been the main causes of blindness in several smaller surveys in Africa.

The overall prevalence in children under six years was 0.11% (95% CI, 0.04-0.24%) and the causes of blindness were quite different compared to the older group. In all of these children the cause of blindness was corneal scarring, probably the result of vitamin A deficiency.

Table 1. Southern Malawi: causes and prevalence of bilateral blindness among persons aged at least six years (1983). Data available on 1574 persons

Cause	*No.*	*Prevalence (%)*	*95% confidence interval*
Cornea related	6	0.38	(0.14-0.83)
Cataract	8	0.51	(0.22-1.09)
Glaucoma	3	0.19	(0.04-0.56)
Retinal disease	1	0.06	(0.01-0.36)
Unknown	2	0.13	(0.01-0.46)
Total	20	1.27	(0.76-1.96)

Surveys of whole countries

The only nations for which population surveys of the whole country for ocular conditions have so far been completed are The Gambia[5], Saudi-Arabia[6] and Nepal[7].

The Gambia survey

In order to make sure that the sample was representative of the whole country, the design of this survey was more elaborate. It was a stratified and multistage sample. If you suspect that there may be variation in disease prevalence between different regions of a country, stratification can create groups of areas within which there is less variation than in the whole. This increases the efficiency of sampling, so that less numbers will be needed for the same precision. It also allows you to make reliable estimates separately for each region, provided the samples are of adequate size; for example, the number of blinding cataracts awaiting surgery in the Eastern Region.

The urban area was considered separately, and each of the three main regions sampled individually. Sampling units were also taken from north and south of the river. Within each of these sub-regions, one or two of the smaller administrative sub-units were selected at random. All the villages within each of these sub-units then provided the sampling frame for the next stage, and the appropriate number of villages were selected.

In the smaller villages, of up to 200 inhabitants, every person was examined. Within the larger villages all the houses were listed, and households then randomly selected. The survey data estimated that 0.7% of the population of The Gambia were blind in both eyes by the WHO standards, so that there were approximately 5500 blind people in the total population of 800 000. When standardized by the aged structure of England and Wales, this corresponded to a blindness prevalence of 1.7%, more than eight times higher than most industrialized countries. The estimated number with low-vision (best vision less than 6/18 but equal to or better than 3/60) was 11 000, making a total with blindness and low-vision of 16 500 (95% CI, 14 000-19 000).

One of the values of this survey in terms of planning was that it gave a figure, within fairly tight confidence limits, of the numbers of people requiring cataract surgery (5500), lid surgery for trichiasis (4600), and medical treatments and broke them down into the numbers for each region. It also showed the excess risk of blindness in females.

In order to achieve even more accurate figures for blindness, enquiry should be made in each village examined, for any children who might be away at blind school.

Nepal survey

The Nepal survey in 1980/81 was a stratified two-stage probability sample of people in 105 sites, the more than 3000 panchayats, or political units, being grouped into 12 strata on the basis of terrain and other characteristics. Ninety-seven rural panchayats were selected, and a single site then selected at random from each. A selection of eight sites was made from urban areas. A total of 39 887 patients were examined.

It was estimated that there were 117 623 blind people in Nepal (0.84% of the population). The causes are shown in Table 2. The distribution of conditions

Table 2. Nepal: causes of blindness (1981)

	Per cent
Cataract	66.8
Iatrogenic sequelae of cataract	5.3
Retinal disease	5.3
Glaucoma	3.2
Infectious other than smallpox or trachoma	2.8
Trachoma	2.4
Trauma	2.4
Smallpox	2.2
Amblyopia	1.3
Nutritional aetiology	0.9
Combinations or undetermined	9.3

was very uneven across the country. There was wide variation in the prevalence of trachoma and of blindness from this cause in different sub-groups.

Such surveys, have been criticized because of the cost involved. However, in a developing country they provide the substantial benefits of recruiting and training a large number of local health staff in eye examination, and also in galvanizing interest throughout the country in the prevention of blindness.

The Western industrialized countries

In general we depend for our working figures of the prevalence and causes of blindness in the Western world on estimates derived from registration. The problems with this have been outlined[8]. Blindness registers are usually maintained in order that blind people will receive appropriate financial benefits and services. Their primary purposes are not medical. The chief weakness is variation in the indications for registration. The extent of under-registration is usually not known, and itself varies according to socioeconomic and racial sub-groups within the population. The only western European country where blindness registration is at present compulsory is Norway.

The figures usually referred to in the UK for causes of blindness are those of Sorsby published in 1966, analysing registrations up to 1962[9]. Already they are more than 25 years out of date.

One of the most quoted population-based prevalence studies was the survey of glaucoma in three Welsh villages by Hollows and Graham[10]. The aim was to examine everybody aged 40-74 years; 91.9% responded. This survey provided invaluable prevalence data for glaucomas in a Caucasian population, but was nevertheless concerned with only one group of diseases.

Cullinan assessed the degree of visual disability in the elderly, as part of a 15 000 household survey[11]. The vision was measured by 150 untrained interviewers. This was correlated with a history of each person's functioning in different circumstances. It was not possible in this survey to determine the causes of visual impairment. The disturbing feature of this survey was that a high proportion of the population, around 40%, had never had a specialist assessment of their visual problem.

The Framingham Eye Study was designed to provide a description of characteristics, prevalence and severity of the four diseases thought to be the major causes of visual impairment in a general population of American adults - cataract, glaucoma, diabetic retinopathy and age related macular degeneration[12]. Ideally the population studied would be representative of the nation or of some large geographic region, but this was not considered feasible economically or logistically. Therefore a study of the adult population of one local area was undertaken. A cohort of adults, aged 52-85 years in 1973, in a town in Massachusetts was chosen. These people had been studied regularly since 1948 in the Framingham Heart Study. Of 3977 subjects still alive and available, 2675 were examined by the ophthalmic team - a response of 67%.

The only recent population-based random sample survey carried out in the UK was that by Gibson *et al.*[13] of 484 subjects of 76 years and older living in Melton Mowbray. There was a 71.5% response. The non-responders did not specifically differ from the responders in age, sex distribution, or whether they lived in town or village.

The prevalence of eye disease was calculated by two methods: (a) assuming the non-attenders had no eye disease, to give a minimum prevalence, and (b) assuming the non-attenders had the same amount of eye disease as those who attended. It is to be hoped that other age groups in this population can be sampled in the future.

Problems in expressing prevalence of blindness

Definitions of blindness

There are at least 65 definitions in use around the world, as discussed by Cullinan[14]. In order that international comparisons can be made, visual acuity should also be recorded at registration or in surveys for each country according to WHO definitions.

How to classify and code the causes of blindness?

There are many possible choices for the criteria for a classification of diseases. The anatomist may desire a classification based on the part of the body or organ affected, the pathologist in the nature of the disease process, the public health practitioner in aetiology, and the clinician in a particular manifestation. A statistical classification of disease and injury will depend upon the use to be made of the statistics to be compiled.

It has been customary in many national blindness registries to record causes under two separate headings: 'Site and type of affection', and 'aetiology'. The categories have not always been helpful.

The present world-wide trend is to adopt the International Statistical Classification of Diseases (ICD 9)[15]. The origins of ICD lay in a list of causes of death, first agreed in 1893. At the Sixth Revision, the classification was extended to cover non-fatal conditions. Later the classification has been shown to be useful for the purposes of hospital indexing.

ICD 9 contains some codes for aetiological agent, notably infections and parasitic diseases, neoplasms, and endocrine and nutritional disorders, and trauma. In these situations an eye disease or cause of blindness can be classified under two entries in ICD 9, so that an aetiology is stipulated. With other disorders, however, the opportunity in the present coding does not exist to stipulate the likely aetiology: for example, there is not a separate entry for genetic disease per se.

There is a need for a revised classification of aetiologies of visual impairment for the purposes of prevalence studies, taking into account contemporary views of causation and aimed at the possibilities for prevention.

One eye or two eyes?

There is at present no agreed convention as to how different causes of blindness in each eye should be recorded.

The simplest solution is to list exactly the numbers for each situation and cause; either:

(A1) binocular blindness, same cause in each eye, and

(A2) bilateral blindness, with each eye having a different cause; or:

(B) bilateral blindness, but listed by eyes: expressed as total eyes. This approach is well illustrated in a paper by Thompson and Chumbley[16].

Another solution is to record for each person the causation of the blinding event, that is the event in the second eye. For example, first eye blind due to amblyopia, second eye due to trauma: the causation here would be trauma. This is convenient, but not a very accurate description of the total causative factors affecting the vision of that population.

A further problem is when there is more than one cause in the same eye; for example, corneal opacity due to trachoma and cataract - both appearing to contribute to loss of vision. If either were treated, vision would become 3/60 or better.

In both The Gambia and the Nepal surveys, the decision was made to assign whichever of the two causes of blindness was the most avoidable, i.e. preventable or curable. For example, a person with cataract in one eye and phthisis due to trauma in the other was diagnosed as being blind from cataract, because a successful operation could restore sight. The latest instructions for the coding of the WHO/PBL Eye Examination Record (PBL/88.1, unpublished) recommend that when there are co-existing primary disorders in the same or different eyes, the principal disorder to be marked is that which is most readily curable or, if not curable, that which is most easily preventable.

Conclusion

We now have more accurate measures of the prevalence of blindness and its causes in several countries of Africa, and in other developing countries, than we do in Europe. In the developing countries these have been invaluable in planning the priorities and manpower training for prevention of blindness programmes. If representative population based sample surveys could be applied in the United Kingdom they would focus effort to prevent avoidable disability. Such surveys could give particularly valuable information on the relative prevalence of different eye diseases in socio-economic and racial subgroups of the population.

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