World blindness: a 21st century perspective

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Historical perspective

At this time, the real start of the new century let alone the new millennium, it is a good time to reflect on where we have been in terms of the prevention of blindness and where we are going. Most of us could watch the dawn break on the new millennium, but there were another 45 million around the world who could not see it because they were blind, and another 135 million who could see it only dimly because of their low vision.¹ Even more distressing is that without new and targeted programmes these numbers will double in the next 20 years. Millions more have vision threatening eye disease.

The causes of blindness have changed dramatically over the years in Western Europe. Two hundred years ago, smallpox was the major cause of blindness.² Smallpox typically affected young adults and often left scarred, blind eyes. However, great progress has been made. Jenner's application of that "traditional medicine", the use of cowpox to "vaccinate", led to a dramatic reduction of smallpox in Europe and the consequent decrease in related blindness. Although vaccination started in 1798, it was only 20 years ago that smallpox was finally eradicated globally.

With the reduction in smallpox and with the social disruption and crowding of the industrial revolution, venereal diseases—syphilis and especially gonorrhoea—became much more common, so that by the end of the 19th century the leading cause of blindness was blinding infections in newborn babies. One consequence of this was the founding of many blind babies' homes and schools for the blind.

In 1881, Credé discovered that bathing the eyes with a drop of silver nitrate could prevent infection and blindness. This proved to be a major breakthrough. Rapidly, institutions and societies were set up to promote Credé's prophylaxis and by the first world war the number of blind babies had been cut dramatically. In New York City, for example, the number was halved in just 15 years.³

At the start of the 20th century, the average life expectancy in most Western countries was about 40 years. We died young, and less than 10% lived their three score years and ten. By 1950, life expectancy had increased to about 60 years. With the increasing number of older people there was a dramatic change in patterns of eye disease and blindness.

In the 1950s most blindness was caused by cataract. The results of surgery were unpredictable and often poor. To see after surgery, people needed to wear thick aphakic lenses. Patients were told to wait for their cataracts to "mature". They had to wait until they were totally blind before the benefit of surgery outweighed the risks of surgery and the relatively poor quality of aphakic vision.

Also at this time alarming numbers of people were going blind from the ocular complications of diabetes. Before Banting and Best discovered insulin in 1921, people with diabetes, especially children, had a tortured but relatively short life as they effectively starved to death. Very few lived long enough to develop any of the other, now common, complications of diabetes. The discovery of insulin by the returned army surgeon and the medical student revolutionised the lives of those with diabetes, but as people with diabetes lived longer and longer, more and more became blind. In the 1950s there was no way to treat this blindness, and all sorts of desperate measures were tried, including pituitary gland ablation.

In the past 50 years there have been dramatic changes in our ability to treat both cataract and diabetic eye disease. The issues facing the health system and providers of eye care at the beginning of the 21st century are vastly different from those early in the 20th century.

As we start the new century, or the new millennium, the two leading causes of blindness that seemed insurmountable 50 years ago—cataract and diabetic eye disease—are essentially conquered in the developed world, although, of course, they will need a significant and sustained effort and commitment to continue to keep them at bay.

The world's population is generally getting older, albeit at somewhat different rates in different regions. Around the world, the number of older people will double in the next 20 years. In Australia, for example, at the turn of the last century, only 10% of people born would expect to reach the age of 80. Now, the average life expectancy is 80 years and two thirds of those who are 40 years old can expect to live to be 90! The day is not far off when the number of people reaching their hundredth year will be counted in tens of thousands.⁴ This will have a significant effect both on the number of people with impaired vision and the frequency of age

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related eye diseases such as macular degeneration and glaucoma.

Major ophthalmic epidemiological studies over the past decade or so have provided the important information needed for the planning of national eye health programmes and priorities. Studies such as the Baltimore and Beaver Dam eye studies in the USA,^{5,6} the Visual Impairment Project and Blue Mountains Eye Study from Australia,^{7,8} the Rotterdam Study (Netherlands), the Andra Pradesh Eye Study,⁹ (India) and the studies in Melton Mowbray¹⁰ and North London¹¹ in the UK have shown us the prevalence and causes of vision impairment. These studies have also elucidated a number of risk factors for eye disease and vision loss and also for the underutilisation of existing eye care services.

Vision 2020

The World Health Organization (WHO) and the International Agency for the Prevention of Blindness have developed a global initiative for the elimination of avoidable blindness by the year 2020; "Vision 2020: the right to sight".^{12,13} The name is suggestive both of the goal, the prevention of avoidable vision loss and blindness by the year 2020 and the notion of good vision, 20/20 (6/6) vision as the target.

Vision 2020 has identified five key areas for action—cataract, trachoma, onchocerciasis, childhood blindness, and refractive error and low vision. Three strategies are outlined for development—disease control, staffing development and training, and infrastructure and appropriate technology. Much information about this initiative is already available (www.v2020.org) and undoubtedly more will follow, both in the pages of this journal and elsewhere.

Some 90% of the blindness in the world occurs in developing countries.¹ In many ways, the problems faced here are similar to the problems faced by developed countries in the past. Cataract surgery is generally not available, or only available to the elite few who live in cities. Trachoma runs unchecked, particularly in more remote, rural areas. Children continue to go blind and die from vitamin A deficiency, often associated with measles. Minor corneal abrasions or conjunctivitis can lead to microbial keratitis and when treated with traditional medicines this may progress to corneal scarring or phthisis. Spectacles are available only in urban areas and for the rich, and low vision services basically do not exist. There is obviously a fertile area for a programme such as Vision 2020 and much work needs to be done.

However, Vision 2020 is a global initiative, and we must consider what Vision 2020 means for us, those who are privileged to live and work in developed areas. Is Vision 2020 something we can just dismiss as being "over there", or are there things we should also do at home? We would like to review what Vision 2020 means for the countries in the developed world. Much of this material is based on our experience in Australia, but this is broadly representative of the situation in other developed

countries. Population based data show that in Australia and other developed countries three quarters of blindness and vision loss are due to just five conditions.⁷ Is this not reminiscent of the rest of the world? It is just that the conditions are somewhat different.

Refractive error

The most surprising finding of recent studies is that refractive error is responsible for one quarter of blindness and half of low vision.⁷ This is especially surprising in a country like Australia which has universal healthcare coverage, good quality services that are well distributed and that include a mix of public and private services. Essentially "free" optometry consultations are available and subsidised spectacles are available for the indigent.

"Presenting" visual acuity indicates the functional vision of people—that is, their vision for driving, for work, and for other everyday activities. The comparison between presenting and best corrected visual acuity indicates the need for refractive services and corrective devices, be they spectacles, contact lens, or other means.

Until now the prevalence and importance of refractive error and its importance as a cause of vision impairment has been overlooked. Definitions of low vision and blindness were based on best corrected visual acuity. Surveys that use the WHO definition, for example, did not report the true prevalence of vision loss due to refractive error. The significance of refractive error has been recognised since studies such as those in Australia and India have reported "presenting visual acuity" in addition to "best corrected" acuity.^{7,14} In Australia we found that refractive error accounted for 53% of the vision loss ($>6/12$) and 24% of the "legal blindness" ($<6/60$).

The solution to eliminate refractive error is simple. It does not depend on finding a new gene or developing a new laser, it is just a pair of glasses. The three issues to be tackled to eliminate low vision and blindness due to refractive error are the human resources to perform refraction and prescription of correction, the availability of affordable spectacles, and adequate access to refraction services.

Despite having noticed a change in vision, many people do not seek eye care. Barriers reported by participants in the Visual Impairment Project to utilisation of services include insufficient time, that the change was not severe enough, it is normal for eyesight to deteriorate with age, fear of outcome, lack of knowledge and cost of services, and reluctance to wear glasses.

Prevention strategies should include routine vision testing of groups at risk of vision loss, mainly older people. The threshold for referral for refractive services (or medical care) should be vision of less than 6/12. Visual acuity of less than 6/12 has a profound impact on the quality, and length, of life. Even this mild loss of vision is associated with a doubling of mortality, morbidity, and social isolation.¹⁵⁻¹⁷

Glaucoma

Glaucoma was described by Fred Hollows as a disease we cannot define, we cannot diagnose, and we do not know how to treat. Although this is still true, well established glaucoma is easily recognisable and at last studies have confirmed the notion that the reduction of intraocular pressure will reduce the rate of loss of vision associated with glaucoma.^{18, 19} So that even though we would prefer to have better treatments available, and much work is being done in this field, the treatment we currently have is much better than nothing. The problem with glaucoma is that at least half the people with glaucoma are not diagnosed, and as a consequence they are receiving no treatment at all. The challenge for us is to identify these people.

Population based studies have shown that one person in 10 will eventually develop glaucoma and this makes it a very common disease.²⁰ The conceptual breakthrough in the past 5 years for glaucoma was the identification of a genetic basis for at least some cases of glaucoma.

People with a family history of glaucoma have an approximately four times increased risk of developing glaucoma.²¹ The identification of the first genes associated with glaucoma was a major breakthrough.²² Although these genes only account for a small percentage of cases, they clearly indicate that a family history is important. This has great public health significance, as it immediately identifies a group of people at a significantly increased risk of developing glaucoma, those with a positive family history. However, studies done in Victoria show that many people with a true family history of glaucoma are unaware of this.²³ It has not been customary for patients with glaucoma to be told that they have a hereditary condition, nor have they been encouraged to pass this information on to family members so that their relatives can have the appropriate checks. The simple messages from this are obvious.

The challenge with glaucoma screening is to try to detect glaucoma early in its course, even though the early changes are the most difficult to define. Although tonometry was originally used as the gold standard for glaucoma, many studies have shown that it has a very low sensitivity and a very poor specificity, missing half of the people with glaucoma and referring nine or 10 times more people for further examination than needed.²⁴

Exciting new developments in psychophysical testing, such as frequency doubling perimetry and developments in optic nerve imaging, such as the scanning laser ophthalmoscope, offer the potential to develop new screening modalities that are both more sensitive and more specific for use in community based screening for glaucoma. They, together with the targeting of those with a positive family history of glaucoma, are likely to significantly decrease the proportion of people with glaucoma who are undiagnosed and not under treatment.

Cataract

Although cataract is no longer a major cause of blindness in most developed countries, this is only because of the very high volume of cataract surgery that is undertaken.

The amount of cataract surgery, or cataract surgery rate (the number of operations per million people per year), is affected by a number of factors. One of the most important factors is age, and the increasing age of the community will lead to a corresponding increase in the amount of cataract surgery required. Basically, the amount of cataract doubles with each decade of life over the age of 40. By the age of 90, one person in two will have had cataract surgery.²⁵

Modern cataract surgery with intraocular lenses has dramatically changed the quality of vision after cataract surgery, and this has led to a dramatic alteration in the visual threshold for which cataract surgery is indicated. There is an exponential increase in the amount of cataract surgery that is required as the visual threshold for surgery is reduced.²⁶ In addition, the amount of cataract surgery required will vary with the functional demands of the individuals, as well as access to and availability of services. Overall, the cataract surgery rate in developed countries varies from 3000 to 6000 per million people per year.

Much work has been done on the identification of risk factors for cataract but this has not yet resulted in effective forms of prevention.²⁷ Exciting recent data have identified the importance of genetic factors in the development of age related cataract.^{28, 29} The role of antioxidants and the efficacy of vitamin supplements are still being ascertained in clinical trials. However, to date the only two established, but modifiable, risk factors for cataract are cigarette smoking and exposure to UVB radiation. Targeted public health interventions should address these two risk factors. These two factors have a combined attributable risk of about 15%. Extrapolation from our data shows that a modest delay of only 14% in the rate of development would delay the onset of cataract by a decade and this would halve the amount of surgery that is required.³⁰

At present, the treatment for cataract is cataract surgery, which is one of the most cost effective public health interventions.¹ In developing countries cataract surgery may cost \$15–32 (£10–21) per DALY (disability adjusted life year),¹ but even in the US cataract surgery only costs \$1796 (£1197) per DALY.³¹

Before the advent of intraocular lens surgery, surgery was delayed until the vision was 6/60 or less. This level of acuity resulted in economic blindness. Now the level of visual acuity that could be described as economic "blindness" is less than 6/12. This is the level of visual acuity usually needed to hold a driver's licence. Vision less than this not only has an economic impact, but it also affects independence and mobility in areas where driving a motor car is almost essential for daily living. As mentioned above the changing threshold for surgery has a dramatic impact on the number of operations required. With the demographic changes that

will occur over the next 20 years, the need for cataract surgery will double and we will have to do twice as many cataract operations to maintain the status quo.

In developing countries, the barriers of access to cataract surgery include cost, distance, lack of awareness of the benefits of surgery, poor quality outcomes, and lack of adequate human resources for both case finding and surgery. These barriers do not apply equally to all people and there are usually groups of people at greater disadvantage. Older people and women are most likely to remain blind from cataract.³² In developed countries the lack of trained personnel is usually not an issue, but the other barriers may exist to a greater or lesser degree as was demonstrated in north London where cataract was still a major cause of vision loss.¹¹

Diabetic retinopathy

Diabetes is a major health problem in industrialised countries and a rapidly emerging problem in urban areas in developing countries. We are facing a global epidemic of diabetes.³³ In developed countries the prevalence of diabetes in adults is around 5%. However, if one includes those with adult onset diabetes who have not yet been diagnosed these numbers will usually double. The implications for the provision of eye care are evident as everyone with diabetes will eventually develop diabetic retinopathy if they live long enough.

Timely treatment can prevent up to 98% of vision loss from diabetic retinopathy.³⁴ Despite this, vision loss from diabetic retinopathy still occurs. In many areas less than half of those with diabetes have had their eyes examined for retinopathy at the recommended frequency.³⁵

Screening needs to include the assessment of both visual acuity and retinal examination. The retina can be examined by either dilated funduscopy or with non-mydriatic photography.³⁶ The minimum sensitivity for any method to be effective if it is repeated at the recommended interval is 60%.³⁷ This level of sensitivity can be achieved with ophthalmoscopy through dilated pupils by suitably trained observers (principally ophthalmologists, optometrists, general practitioners, or physicians) or with non-mydriatic photography.

Screening for diabetic retinopathy needs to be community based in addition to clinic based services and can include a range of examination modalities. The use of the non-mydriatic camera empowers an additional cadre of health professionals who can participate in screening programmes.

Screening of people with diabetes is cost effective with the maximum savings gained if compliance reaches 80% to 90%.^{37, 38} The funds invested to increase compliance are a small but vital component of the costs of a screening programme.³⁹

Knowledge of the need for screening, especially in the absence of symptoms, is a major barrier to regular screening for many people with diabetes.⁴⁰ Learning about the need for screening prompts many people to

commence regular screening, but other barriers exist such as time, cost, and the ability to attend appointments play a part. Even in developed countries, attitudes and beliefs about prevention, treatment, and the inevitability of disease and vision loss are important. Some barriers may be country specific owing to the nature of the healthcare system, but surprisingly many appear to be universal.

Thus the development of awareness of the need for regular eye examinations is of prime importance in the community in general and in people with diabetes and their families specifically. Some successful community based programmes have been developed to do this. For example, through the Lions Clubs International, the SightFirst programme has funded awareness campaigns in the USA, UK, Japan, Canada, and Australia. These Lions Eye Health programmes (LEHP) have developed videos, print, and broadcast materials incorporating the evidence based information so that communities can use well designed and accurate materials to encourage eye examinations. These LEHP activities are carried out through national and local partnerships with eye and healthcare groups and other community organisations to plan and implement awareness campaigns tailored to the needs of their local communities.

Once people have their first eye examination, reminder systems are required to encourage continuing eye examinations. Reminder systems can range from a simple reminder card to tracking software for eye or healthcare providers.

Working towards a higher level of patient compliance is one part of the solution to the prevention of vision loss from diabetic retinopathy. The other significant component is practitioner compliance with screening and treatment guidelines. Professional organisations such as the Royal College of Ophthalmologists, the American Academy of Ophthalmology, national governments (Australian National Health and Medical Research Council), and international organisations (St Vincent Declaration) have produced guidelines for practitioners. However, the issuing of guidelines on their own does not necessarily lead to major changes in professional practice.⁴¹ Regular audits of clinical management can be very useful to indicate areas of compliance and areas where more effort is needed.⁴²

Age related macular degeneration

Age related macular degeneration (AMD) is now the leading cause of blindness in developed countries. The prevalence of AMD increases dramatically with age. Although cases of AMD were described during the last half of the 19th century, AMD was regarded as an uncommon disease until after the second world war. Although AMD is uncommon before the age of 50 or 60 years, by their 90s, nearly two out of three people will have developed early AMD and one in four will have lost vision from AMD.⁴³ Thus, with the ageing population, the frequency of AMD has increased dramatically.

Recent data show that the only identifiable risk factor for macular degeneration which can be altered is cigarette smoking.⁴⁴ Those who smoke cigarettes have some three to five times higher risk of developing macular degeneration than non-smokers. AMD must be added to the list of diseases associated with cigarette smoking and this information used in campaigns to encourage people to quit smoking.

It is apparent that AMD runs in families, although the genetic study of this is exquisitely difficult.⁴⁵⁻⁴⁶ As the disease is not manifested until late in life, people who may become affected are phenotypically normal for most of their life, and this makes the normal genetic association studies almost impossible to perform. Studies with sibs or particularly twins with AMD may be useful, as will be the study of the earlier onset hereditary macular degenerations that may share some similarities.

The treatment of macular degeneration also leaves much to be desired. Focal photocoagulation of classic subretinal neovascularisation is appropriate only in a small proportion of cases and slows rather than prevents further vision loss.⁴⁷ The use of photosensitising dyes may offer a new perspective to treatment, but at present this expensive treatment needs to be repeated on a regular basis in the majority of cases.⁴⁷ Surgical techniques to excise subretinal tissue, or translocate or transplant various layers of the retina are being studied. They are still highly experimental and await further development before they can be generally applied.

There is no question that macular degeneration remains the biggest challenge in terms of prevention of blindness in developed areas.

Although at present we are unable to do much to prevent or treat macular degeneration, a great deal can be done to enhance the function of those who are affected. Early rehabilitation and the use of low vision services can make a tremendous difference in the visual function of people with low vision. The development in low vision devices, from simple magnifiers to closed circuit TV reading devices, can revolutionise the functional ability of people with low vision. Despite this, as many as two out of three of those with low vision are not referred or do not utilise low vision services,⁴⁸ and frequently those who are referred are not referred until they are essentially blind. There is a real need to increase the availability and utilisation of low vision services, particularly for the elderly with earlier degrees of visual impairment.

Conclusion

In developed countries, blindness and vision loss increase threefold for each decade over 40. This means that the number of people who are vision impaired or blind will double in the next 20 years, just as an effect of the ageing of our population. Similarly, the need for cataract surgery will more than double in the next 20 years, as will the number of people needing screening or treatment for diabetic eye disease. This will need both an increased allocation of resources and the more efficient use of those

we currently have. The challenge in the years ahead will be to find ways to prevent, delay, or treat these diseases such as glaucoma and macular degeneration, diseases that seem to result from our living longer than our eyes.

Vision 2020, the global elimination of avoidable blindness around the world, clearly has significant implications for us in developed countries where much still needs to be done, in addition to the more obvious issues in developing areas.

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