*Editorial*

# The many challenges of childhood blindness

There are an estimated 45 million blind people in the world of whom only 3% are children.[1] This dramatic difference in numbers of blind adults compared with children accounts in part for the relatively minor importance that has been attributed to the problem surrounding childhood blindness. Certainly, the well organised advocacy groups for the elderly in many developed countries are not matched by comparable ones for children. The result of this can be seen in the difference in resources made available for health services and research for adult blindness versus childhood blindness. One hopes that, now that the World Health Organization (WHO) and International Agency for Prevention of Blindness have developed a global initiative to eliminate avoidable blindness and have included childhood blindness as one of its five key areas, this will change.[2] In this issue of the *BJO* (p 1149) Kocur and co-workers report on the causes of severe visual impairment (visual acuity in the better eye less than 6/60) and blindness (visual acuity in the better eye less than 3/60) in the Czech Republic. This is an excellent study and the authors raise issues that go well beyond the borders of the Czech Republic. We wish to highlight only two of these issues.

First, and foremost, is the issue that the authors emphasise themselves—the continuing havoc resulting from retinopathy of prematurity (ROP). In this study ROP was the leading cause of blindness—41.9% had ROP. Although this figure appears to be staggering at first, similar studies in Bulgaria and Cuba have reported incident rates of 25.9% and 38/6% respectively.[3] The ever changing epidemiology of ROP is difficult to summarise precisely. The simplistic view that ROP is becoming less of a problem in the more developed nations while being a major problem in emerging nations, who are just now beginning to establish intensive care neonatal units, is misleading. Isolated reports have suggested that the incidence of ROP is decreasing in developed nations.[4 5] A study in Denmark found a decrease in the incidence in ROP for infants with birth weights between 1251 and 1750 g but not for infants weighing less than 1251 g.[6] These smaller premature infants are more likely to suffer from ROP and the neurological sequelae of intraventricular haemorrhage and hypoxia.[7] The Light-ROP study showed no reduction in incidence of ROP.[8] At the present time in San Francisco, approximately 20% of children referred for preschool services to the Variety Club for Blind Babies Foundation have ROP. Increasingly, they are multiply handicapped with severe neurological and developmental problems. While it is true that improved neonatal care has resulted in an improved survival and quality of life for premature infants, ROP remains an important cause of childhood visual impairment in the developed world. Finally, although it is too little spoken of, the incidence of ROP in developed nations is significantly affected by the guidelines established by neonatal units for resuscitating and supporting very ill tiny premature babies. This is an ethical dilemma in which open discussion and debate would be welcomed.

In the Third World where no neonatal units are available, ROP is, of course, not a problem. However, as emerging nations develop neonatal care nurseries ROP becomes a larger part of the picture of childhood blindness. Establishing guidelines for screening and treating ROP are essential in these countries. The study of Kocur and co-workers identified 96 children with ROP. Yet, only four of these children had been treated with cryotherapy. Surely, this suggests that many of these children were cared for in neonatal units that did not have adequate screening or treatment programmes for ROP. The emphasis of the authors on establishing such programmes is well placed. Certainly, in some parts of the world this is a staffing issue with too few available ophthalmologists trained to screen for ROP. Of course, even with a well established programme of screening for ROP our treatment options remain limited and not entirely effective.[9] ROP remains a problem for all but the poorest nations without neonatal intensive care units.

Conspicuous by its insignificance in the study of Kocur and co-workers is cortical visual impairment. They found only four children in their study with this diagnosis. This is striking and deserves comment. In the developed world ocular causes of visual impairment and blindness in children have decreased in frequency during the 20th century. In contrast, various central nervous system disorders have become the most common causes of visual impairment in children in many countries.[10 11] The term cortical visual impairment has been used to describe these central nervous system disorders. The original definition of cortical visual impairment was loss of vision due to bilateral dysfunction of the optic radiations and/or visual cortex. Recently, however, it has regrettably been applied to a myriad of disorders including autism, learning disabilities, and attention deficit disorders. For the purpose of this discussion we shall use cortical visual impairment to describe children with visual loss due to optic radiation, striate cortex, and peristriate cortex damage. Causes of cortical visual impairment in children include, but are not limited to, perinatal hypoxia, near drowning episodes, hydrocephalus, trauma (including non-accidental), meningitis, and periventricular leucomalacia. Many of these children have damage to non-visual portions of the central nervous system and are therefore significantly handicapped in functions other than vision. Many have severe neurodevelopmental problems and for this reason often are excluded from residential schools for the blind. Since Kocur and co-workers performed their study in 10 primary schools for visually handicapped children this may account at least in part for why they found only four children with the diagnosis of cortical visual impairment.

The child with cortical visual impairment is challenging to his/her parents, physicians, and teachers. Standard techniques to evaluate visual function are often inadequate to describe precisely the extent and nature of visual impairment. Educational approaches for intervention designed for the child with ocular causes of visual impairment are often unsuccessful. Recently, educators have developed specific instructional intervention strategies for the child with cortical visual impairment; nevertheless, the potential for the child with cortical visual impairment to live an independent and productive life is often not good.

In San Francisco, the two leading causes of childhood blindness and visual impairment are ROP and cortical visual impairment. In some ways they both result from

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improved medical care and technology which has allowed very premature and/or severely brain damaged children to survive. It is not altogether precise to refer to them as iatrogenic disorders but there are complex difficult ethical issues raised by these disorders which should remind us that advances in medical technologies are frequently accompanied by significant adverse effects. If our goal is to eliminate preventable causes of blindness by 2020, ROP and cortical visual impairment must be involved in a major portion of the effort on behalf of visually impaired children in the developed world.

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1 World Health Organization. *Preventing blindness in children.* Report of a WHO/IAPB scientific meeting. WHO/PBL/00.77. Geneva: WHO, 2000.
2 Thylefors B. A global initiative for the elimination of avoidable blindness. *Am J Ophthalmol* 1998;**125**:90–3.
3 Gilbert CE, Rahi J, Eckstein M, *et al*. Retinopathy of prematurity in middle-income countries. *Lancet* 1997;350:12–14.
4 Bullard SR, Donahue SP, Feman SS, *et al*. The decreasing incidence and severity of retinopathy of prematurity. *J AAPOS* 1999;**3**:46–52.
5 Rowlands E, Ionides ACW, Chinn S, *et al.* Reduced incidence of retinopathy of prematurity. *Br J Ophthalmol* 2001;**85**:933–5.
6 Fledelius HC, Dahl H. Retinopathy of prematurity, a decrease in frequency and severity. Trends over 16 years in a Danish county. *Acta Ophthalmol Scand* 2000;**78**:359–66.
7 Volpe JJ. Intraventricular hemorrhage and brain injury in the premature infant. Neuropathology and pathogenesis. (Review) *Clin Perinatol* 1989;**16**: 361–386.
8 Reynolds JD, Hardy RJ, Kennedy K, *et al*. Light reduction in retinopathy of prematurity (Light-ROP) Cooperative group. Lack of efficacy in preventing retinopathy of prematurity. *N Engl J Med* 1998;**328**:1572–6.
9 Good WV, Gendron RL. Gene therapy for retinopathy of prematurity: the eye is a window to the future. *Br J Ophthalmol* 2001;**85**:908–11.
10 Rosenberg T, Flage T, Hansen E, *et al.* Incidence of registered visual impairment in the Nordic child population. *Br J Ophthalmol* 1996;**80**:49–53.
11 Huo R, Burden SK, Hoyt CS, *et al*. Chronic cortical visual impairment in children: aetiology, prognosis, and associated neurological deficits. *Br J Ophthalmol* 1999;**83**:670–5.