

ORIGINAL ARTICLES—Clinical science

The contribution of low birth weight to severe vision loss in a geographically defined population

Barbara J Crofts, Rosemary King, Ann Johnson

Abstract

Aims—To describe the birthweight specific rate of severe vision loss among babies born between 1 January 1984 and 31 December 1987 to mothers resident in a geographically defined area, to classify the causes of vision loss by time of origin, and to describe the associated sensory and motor impairments and disabilities.

Methods—Cases were identified from a population register of children with early childhood impairment, which uses multiple sources of ascertainment. Further clinical information was retrieved from hospital records and by asking ophthalmologists caring for the children.

Results—166 (1.25/1000 live births) children with severe vision loss diagnosed by the age of 5 years were identified. The rate among babies born weighing less than 1500 g at birth was 26 times higher than the rate for babies between 2500 g and 3499 g. These very low birthweight babies contribute 17.5% of all severely visually impaired children. Almost two thirds of children with severe vision loss have a lesion of prenatal origin. Other sensory or motor deficits are present in 69% of the children. Retinopathy of prematurity accounted for 5.4% of all visually impaired children and seven of the 166 children met the criteria for perinatal asphyxia.

Conclusions—Although the contribution made by babies with a low birth weight to overall severe vision loss in the community is small, many of these children have additional impairments and probably place considerable demands on health and educational services and families. Reduction in the frequency of vision problems in the preschool population as a whole is unlikely to occur until there are major advances in the understanding of the aetiology and prevention of eye conditions of genetic, prenatal, and developmental origin.

(*Br J Ophthalmol* 1998;82:9-13)

Babies with a birth weight under 1500 g are at a higher risk of later vision and ocular problems than larger babies. This is known from follow up studies of groups of babies

defined by birth weight.¹⁻⁴ In particular, a higher frequency of myopia, strabismus, and the long term effects of retinopathy of prematurity (ROP) have been described.⁵⁻⁷ As the number of babies born weighing under 1500 g who survive following neonatal intensive care has increased in the past few years⁸ there is a concern that the number of children with later vision and ocular problems has also increased and that some of these have severely reduced vision, possibly with additional sensory and movement disorders. It is likely that additional health and educational services will be required to meet the needs of these children.

The contribution made by children of low birth weight to the overall number of severely visually impaired children in England is difficult to estimate. This is because there are few reliable population data about children with severe or total vision loss. Figures from the BD8 notification system appear to underestimate the numbers of young children with severe vision loss and many previous surveys have been hospital or school based, thus having a selection bias.⁹⁻¹¹ It has been difficult, therefore, to estimate birthweight specific rates of severe vision loss in children, to monitor trends and variations in these rates over time, to answer questions about the contribution of ROP to childhood blindness in the UK, or to know the extent to which vision loss in children is associated with other impairments and disabilities.

In response to unanswered questions of this type the Oxford Register of Early Childhood Impairment (ORECI) was set up in 1984. The register collects information on children with severe vision loss, cerebral palsy, and/or sensorineural deafness who are born to mothers resident at the time of delivery in one of the four counties of the former Oxford Region—that is, Berkshire, Buckinghamshire, Northamptonshire, and Oxfordshire. Multiple sources of ascertainment are used and a minimal amount of information collected on each child.¹² Because diagnosis may be uncertain in young children, decisions are not made about whether or not a child is a "case" until the age of 3 years. The diagnosis and level of disability is rechecked when the child is 5 years old. The register has formed the basis of this more detailed study of all children in a geographi-

Oxford Eye Hospital,
Radcliffe Infirmary
NHS Trust
B J Crofts

Oxford Register of
Early Childhood
Impairments,
Women's Centre, John
Radcliffe Hospital,
Oxford
R King

Oxford Register of
Early Childhood
Impairments, National
Perinatal
Epidemiology Unit,
Oxford
A Johnson

Correspondence to:
Ms B J Crofts, Oxford Eye
Hospital, Radcliffe Infirmary
NHS Trust, Woodstock
Road, Oxford OX2 6HE.

Accepted for publication
28 May 1997

Table 1 Children with severe vision loss, born in years 1984–7, diagnosed by the age of 5 years, by birthweight group

Birth weight (g)	No of live births	Children with severe vision loss			
		No	%	Rate/1000 live births	95% CI
<1500	1 118	29	17.5	25.9	17.4–37.0
1500–2499	7 548	27	16.3	3.6	2.2–4.9
2500–3499	74 358	78	47.0	1.0	0.8–1.2
3500 and more	49 392	32	19.3	0.7	0.4–0.9
All weights	132 416	166	100.0	1.3	1.0–1.4

cally defined population born between 1984 and 1987 with severe vision loss.

In this study we describe birthweight specific rate of severe vision loss in a geographically defined population of children in England, classify the underlying causes of vision loss by the likely time of origin, and describe the associated motor, other sensory and developmental impairments and disabilities. Based on this information, we have estimated the likely effect of increasing survival rates of low birthweight babies on the numbers of children with severe vision loss in the UK. In addition, we discuss the extent to which severe vision loss is preventable in young children in England.

Methods

A list was obtained of all children born between 1 January 1984 and 31 December 1987 who were included as a “case” of severe vision loss on the Oxford Register of Early Childhood Impairment (ORECI) and who had been diagnosed by the age of 5 years. The definition of a case is a child with a visual acuity in the better eye of 6/18 or less. If visual acuity could not be measured, as in the presence of an associated neuromotor deficit or developmental delay, an assessment of the degree of visual impairment was made on the behavioural responses of the child. Birth weight, gestational age at birth, and place of birth were available from the register. In addition, the register has information on whether or not the child has cerebral palsy, sensorineural hearing loss, seizures, or developmental delay. After contacting the ophthalmologist looking after the child, information was abstracted from the hospital notes of mothers and babies. Data extracted included family history, condition at birth, and neonatal course. Birth asphyxia was defined as an Apgar score of 3 or less at 1 minute after birth, with evidence of neonatal encephalopathy. In addition, events in the first 3 years of life which appeared to have contributed to vision loss were recorded. Abnormalities of the eyes and visual pathways as described in the hospital notes were recorded. If the diagnosis was

Table 2 Estimated time of origin of vision impairment by birthweight group

Birth weight (g)	Time of origin of impairment									
	Prenatal		Perinatal		Postnatal		Not known		Total	
	No	%	No	%	No	%	No	%	No	%
<1500	8	27.6	18	62.1	2	6.9	1	3.4	29	100.0
1500–2499	20	74.1	5	18.5	2	7.4	0	0	27	100.0
2500–3499	49	62.8	13	16.7	10	12.8	6	7.7	78	100.0
>3500	23	71.9	4	12.5	3	9.4	2	6.3	32	100.0
All weights	100	60.3	40	24.1	17	10.2	9	5.4	166	100.0

unclear, the examining ophthalmologist was contacted for further details.

Based on this information, a judgment was made of the likely time of origin of the vision/ocular disorder and the children allocated to one of four groups:

- (1) Prenatal—conditions present at birth including conditions of known genetic origin, a developmental abnormality of the eye or visual pathways, or the result of intrauterine infections.
- (2) Perinatal—conditions assumed to be related to events in late pregnancy, during labour and delivery, in the neonatal period, or in the time up to hospital discharge if hospitalisation extended beyond 28 days.
- (3) Postnatal—conditions originating after the neonatal period or after hospital discharge if hospitalisation extended beyond 28 days after birth up to the age of 5 years.
- (4) Unclassifiable.

Results

In the years 1984 to 1987 inclusive, there were 132 416 live births to mothers resident in the four counties of the former Oxford Region. Of these, 166 (1.25/1000 live births) were diagnosed as having a vision loss by the age of 5 years and are included in this cohort. Twenty six of these children have died.

SEVERE VISION LOSS BY BIRTHWEIGHT GROUP

The rate of severe vision loss among babies weighing under 1500 g at birth was 26 times higher than the rate for babies weighing between 2500 g and 3499 g at birth (Table 1). Although the risk of severe vision loss was highest in the smallest babies, two thirds (110/166) of the visually impaired children in the population had a birth weight of 2500 g or more. Babies weighing 1500–2499 g at birth had a fourfold increase in rate of severe vision loss compared with babies weighing more than 2500 g at birth.

CLASSIFICATION OF SEVERE VISION LOSS BY TIME OF ORIGIN

Three fifths of the visually impaired children were considered to have a problem of prenatal origin (Table 2). Prenatal factors accounted for a higher proportion of the visual impairment in the group of children who had weighed more than 1500 g at birth than in those who had weighed less than 1500 g. Almost a quarter of the 166 children were considered to have a perinatal origin to their vision loss. Of the 40 children in this group, almost a half weighed less than 1500 g at birth. Ten per cent of severely visually impaired children had an identifiable event in infancy or early childhood associated with the onset of severe vision loss.

ASSOCIATED IMPAIRMENTS

Over two thirds of the visually impaired children had an additional motor or sensory deficit or developmental delay (Table 3). Children in the lowest birthweight group were more likely to have associated deficits than the children with higher birth weights. Over 70% of severely visually impaired children who

Table 3 Severe vision loss and associated impairments by birthweight group

Birth weight (g)	Vision only		Associated impairments		Not known		Total	
	No	%	No	%	No	%	No	%
<1500	5	18.5	21	72.4	3	10.3	29	100.0
1500–2499	6	22.2	19	70.4	2	3.7	27	100.0
2500–3499	23	30.7	45	57.7	10	9.3	78	100.0
>3500	17	53.1	14	43.8	1	6.2	32	100.0
All weights	51	30.7	99	59.6	16	9.6	166	100.0

weighed less than 1500 g at birth had an additional motor, sensory, or intellectual deficit compared with only 43.8% of those weighing 3500 g or over.

The commonest associated problem was cerebral palsy (CP). Two fifths (68/166) of the children with visual impairment on the register had some degree of motor deficit. This was most frequent among the children with vision loss of perinatal origin (31/40; 78%). Of the 100 children with vision loss of prenatal origin 26 had associated CP, compared with 11 of 17 children with vision loss of postnatal origin.

TYPE OF OCULAR/VISION LESION

The ophthalmological diagnoses are given in Table 4. Among the recognised causes of prenatal vision impairment were metabolic and neurodegenerative disorders (13), cerebral developmental abnormalities (11), and intrauterine infection (2). In addition, there were a number of children with a history of consanguinity or siblings affected, in whom a genetic origin was likely. Nine children with a lesion of perinatal origin had retinopathy of prematurity. These accounted for 5.4% of all severely visually impaired preschool children in the population. All but one of these weighed less than 1000 g at birth. Most of the children in this perinatal group with cortical visual impairment or optic atrophy also had cerebral palsy. In seven of these 23 children there was a history of birth asphyxia; this accounts for

Table 4 Causes of severe vision loss in a 4 year birth cohort (1984–7) in a geographically defined population of preschool children

Time of origin of impairment	Diagnosis	No of children	
Prenatal:	Cortical visual impairment	27	
	Cataract	16	
	Congenital nystagmus	10	
	Albinism	9	
	Optic atrophy	8	
	Retinal dystrophy	11	
	Anophthalmos/microphthalmos	4	
	Colobomata	4	
	Optic nerve hypoplasia	4	
	Severe myopia	2	
	Buphthalmos	1	
	<i>Other: (one each)</i>		
	Peters' anomaly		
	Rieger's		
Moebius syndrome			
Other syndrome			
Total		100	
Perinatal:	Cortical visual impairment	23	
	Retinopathy of prematurity	9	
	Severe myopia (assoc with ROP)	1	
	Optic atrophy	7	
Total		40	
Postnatal:	Cortical visual impairment	10	
	Optic atrophy	6	
	Retinoblastoma	1	
Total		17	
	Unclassifiable	9	

4.2% of all visually impaired preschool children in the population.

All but one of the children in the postnatal group had either a diagnosis of cortical visual impairment or optic atrophy. Their underlying causes included four "near miss" sudden infant deaths, six children with meningitis/encephalitis, two with trauma, two with hydrocephalus, one with Reyes' syndrome, and one following a prolonged period of status epilepticus.

Discussion

This survey was based on a population register of children with severe vision loss. Details of the vision and ocular problems were derived retrospectively from hospital notes. This process lacks the precision of the much more expensive prospective follow up studies of groups of babies at particular risk of vision loss, such as those of low birth weight. On the other hand, the register provides a unique opportunity to collect information on all children in the population with severe vision problems, to assess the contribution made by VLBW babies to the numbers of visually impaired children in the population, and to monitor trends in vision and ocular problems which may be associated with changes in obstetric and neonatal management. This information, which is useful to both purchasers and providers of health care, is difficult to find in most areas of the UK. Information systems maintained at regional level have largely been abolished and it has proved difficult to retrieve these data from computerised child health information systems.

Because we use a system of multiple ascertainment of cases,¹² we believe that most children with a severe vision problem in the area are known to us. The population rate of 1.25/1000 live births is higher than on other registers,¹³ but this may be partly accounted for by differences in the definition of a case. We also included children with a severe neurological deficit or developmental delay who, in the opinion of an ophthalmologist, had behavioural responses suggesting low vision. Level of vision loss can be difficult to assess in these children and it is possible that ascertainment among these children may be less precise.

Our study confirmed the higher risk of severe vision loss among babies born with a low birth weight compared with larger babies. The numbers of such babies in a population will be influenced by the mortality rate among this birthweight group. There are some variations by geographic area in the UK in these mortality rates. For example, in 1992 the overall infant mortality rate for babies weighing less than 1500 g at birth in England and Wales was 240/1000 live births. The rate per 1000 live births by area varied from 192 to 304; the region covered by this register had a rate of 211 per 1000 live births. The impact of such variations on the number of visually impaired children will not be great however, as birth under 1500 g accounts for only 1% of all live births.

The raised rate of severe visual impairment in this low birthweight group was accounted for mainly by vision loss associated with cerebral palsy rather than the end result of retinopathy of prematurity. Hence, it seems that the vulnerability of the preterm brain to ischaemia leading to periventricular leucomalacia involving both motor and visual pathways is a greater problem for immature babies than the risk of irreversible damage to the immature retina resulting directly or indirectly from hyperoxia. Some degree of ROP is undoubtedly common in preterm babies but with the level of neonatal intensive care available to these babies and the level of screening and treatment now available, blinding ROP is rare in our area.

Twenty five years ago ROP may have been a more frequent cause of blindness in early childhood.¹⁴ Similarly, higher prevalences may be seen in other populations especially in developing countries where neonatal intensive care units are now being developed.¹⁵ It also remains a possibility that the use of very aggressive therapy in the UK for babies who are at the borderline of viability, and are probably very vulnerable to ROP, may lead to an increase in the number of surviving babies with severe grades of ROP. Routine screening and early intervention when appropriate will continue to play a key role in reducing the impact of this condition in preschool children.^{16 17}

Babies who weigh between 1500 and 2499 g at birth are of particular interest. Many of the babies with low vision in this weight group have a lesion of prenatal origin. This suggests that an adverse prenatal environment may have contributed to both the eye lesion and to the poor fetal growth which has resulted in a low birth weight. Babies in this weight group also account for almost as many children in the population of severely visually impaired children as do babies in the very low birthweight group.

Despite this increased risk of vision and ocular problems among the immature babies, most of the children in the community with severe vision loss did not have a low birth weight.¹⁸ In order to understand the extent to which severe vision loss was preventable, we attempted to classify the children by the likely time of origin of the vision/ocular lesion. Although there is a well described genetic origin to many conditions, and others are clearly developmental or anatomical anomalies, albeit of unknown aetiology, the time of origin can be difficult to ascertain in some children. For example, it is likely that some of the children with "cortical visual impairment" have cerebral ischaemic lesions of prenatal origin. This may be so even in the presence of clinical signs of "birth asphyxia". Recent work has shown that such signs may reflect preceding cerebral injury rather than an acute intrapartum event.¹⁹ Alternatively, the ischaemic brain damage may be the end result of a continuum of adverse events in a vulnerable baby. Better understanding of the risk factors for cerebral ischaemic injury will help to identify interventions which

might reduce the frequency of vision/ocular lesions of this type.

Despite these difficulties in classification, it is clear that most severe vision loss in early childhood is associated with conditions of genetic origin or due to other prenatal factors influencing eye development, most of which cannot be identified. We estimated that 60% of all the visually impaired children in our population were in this group. This is similar to the estimates derived from both the large Nordic register²⁰ in which 66% of visually impaired children were considered to have prenatal aetiological factors and the large survey of Irish children²¹ in which 55% of the children were classified in this way. Ways of reducing the numbers of children in this group are limited. Progress has been made; for example, new cases of blindness due to congenital rubella syndrome are rarely seen since the introduction of widespread immunisation programmes. Nevertheless, reduction will not be achieved until there is a clearer understanding of the inter-relation of genetic and environmental factors which affect the development of the eye.²²

For one in 10 children with severe vision loss, it arises from an event occurring well after birth. Many of these children also have evidence of motor and cognitive deficit. In this group the challenge is prevention as most are attributable to accidents and infections.²³

Most of the children with severe vision loss classified as perinatal were born preterm. The extent to which the vision problems in these babies is preventable is not clear. It is possible that interventions which reduce the risk of brain haemorrhage and ischaemia may result in a reduction of the risk of severe vision loss and other neurological deficits.

Almost two thirds of children under 5 with a severe vision problem also have other impairments and disabilities. The association of vision disorders and cerebral palsy has been well documented in a study in a school for motor impaired children.²⁴ On the Nordic register, a register of visually impaired children in Denmark, Finland, Iceland, and Norway, 57% of the children have additional intellectual or motor disability.¹³ These associated disabilities need to be taken into account when planning the type of educational provision they will need.

Conclusions

In summary, although the chance of survival among babies born very early and very small continues to increase and there may be larger numbers of visually impaired survivors, their contribution to the total number of visually impaired children in the population will be small.

Hence, although it is appropriate to continue to find ways of reducing the risk of retinopathy of prematurity and haemorrhagic lesions of the brain in very preterm babies, further reduction in the prevalence of blindness in preschool children in the UK will result from preventive strategies directed at the genetic, developmental, and prenatal causes.

This study illustrates the value of a population register in monitoring rates of early childhood impairment.

We are grateful to the ophthalmologists who allowed access to medical notes and to the Steering Committee of the Oxford Register of Early Childhood Impairments (ORECI) who gave permission for access to register data. ORECI is funded by Anglia/Oxford Research and Development programme. Ann Johnson is funded by the Department of Health. The study was supported by the British Council for Prevention of Blindness.

- 1 Gibson NA, Fielder AR, Trounce JQ, Levene MI. Ophthalmic findings in infants of very low birthweight. *Dev Med Child Neurol* 1990;**32**:7–13.
- 2 Gallo JE, Lennerstrand G. A population-based study of ocular abnormalities in premature children aged 5 to 10 years. *Am J Ophthalmol* 1991;**111**:539–47.
- 3 Burgess P, Johnson A. Ocular defects in infants of extremely low birth weight and low gestational age. *Br J Ophthalmol* 1991;**75**:84–7.
- 4 Dowdeswell HJ, Slater AM, Broomhall J, Tripp J. Visual deficits in children born at less than 32 weeks' gestation with and without major ocular pathology and cerebral damage. *Br J Ophthalmol* 1995;**79**:447–52.
- 5 Robinson R, O'Keefe M. Follow up study on premature infants with and without retinopathy of prematurity. *Br J Ophthalmol* 1993;**77**:91–4.
- 6 Page JM, Schneeweiss S, Whyte HEA, Harvey P. Ocular sequelae in premature infants. *Pediatrics* 1993;**92**:787–90.
- 7 Powls A, Botting B, Cooke TWI, Stephenson G, Marlow N. Visual impairment in very low birthweight children. *Arch Dis Child* 1997;**76**:F82–7.
- 8 Alberman E, Botting B. Trends in prevalence and survival of very low birthweight infants, England and Wales: 1983–1987. *Arch Dis Child* 1991;**66**:1304–8.
- 9 Baird G, Moore AT. Epidemiology. In: Fielder AR, Best AB, Bax MCO, eds. *The management of visual impairment in childhood. Clinics in Developmental Medicine No 128*. London: Mackeith Press, 1993.
- 10 Evans J, Rooney C, Ashwood F, Dalton N, Wormald R. Blindness and partial sight in England and Wales: April 1990–March. 1991. *Health Trends* 1996;**28**:5–12.
- 11 Walker E, Tobin M, McKennell A. *Blind and partially sighted children in Britain: the RNIB survey 1992*. London: HMSO.
- 12 Johnson A, King R. A regional register of early childhood impairments: a discussion paper. *Community Med* 1989;**11**:353–63.
- 13 Riise R, Flage T, Hansen E, Rosenberg T, Rudanko S-L, Viggosson G, Warburg M. Visual impairment in Nordic children 1 Nordic registers and prevalence data. *Acta Ophthalmol* 1992;**70**:145–54.
- 14 Fraser GR, Friedman AI. *The causes of blindness in childhood*. Baltimore: Johns Hopkins Press, 1967.
- 15 Gilbert CE, Canovas R, Kocksch de Canovas R, Foster A. Causes of blindness and severe visual impairment in children in Chile. *Dev Med Child Neurol* 1994;**36**:326–33.
- 16 Report of a working party. *Retinopathy of Prematurity: Guidelines for screening and treatment*. London: The Royal College of Ophthalmologists and British Association of Perinatal Medicine, 1995.
- 17 Cryotherapy for Retinopathy of Prematurity Cooperative Group. Multicentre trial of cryotherapy for retinopathy of prematurity. *Arch Ophthalmol* 1996;**114**:417–24.
- 18 Stayte M, Johnson A, Wortham C. Ocular and visual defects in a geographically defined population of 2-year-old children. *Br J Ophthalmol* 1990;**74**:465–8.
- 19 Gaffney G, Squier MV, Johnson A, Flavell V, Sellers S. Clinical associations of prenatal ischaemic white matter injury. *Arch Dis Child* 1994;**70**:F101–6.
- 20 Rosenberg T, Flage T, Hansen E, Rudanko S-L, Viggosson G, Riise R. Visual impairment in Nordic children. Actiological factors. *Acta Ophthalmol* 1992;**70**:155–64.
- 21 Goggin M, O'Keefe M. Childhood blindness in the Republic of Ireland: a national survey. *Br J Ophthalmol* 1991;**75**:425–9.
- 22 Gilbert C. Prevention of childhood blindness. In: Fielder AR, Best AB, Bax MCO, eds. *The management of visual impairment in childhood. Clinics in Developmental Medicine No 128*. London: Mackeith Press, 1993.
- 23 Robinson GC, Jan JE. Acquired ocular visual impairment in children. 1960–1989. *Am J Dis Child* 1993;**147**:325–8.
- 24 Black P. Visual disorders associated with cerebral palsy. *Br J Ophthalmol* 1982;**66**:46–52.



The contribution of low birth weight to severe vision loss in a geographically defined population

Barbara J Crofts, Rosemary King and Ann Johnson

Br J Ophthalmol 1998 82: 9-13

doi: 10.1136/bjo.82.1.9

Updated information and services can be found at:

<http://bjo.bmj.com/content/82/1/9.full.html>

References

These include:

This article cites 19 articles, 10 of which can be accessed free at:

<http://bjo.bmj.com/content/82/1/9.full.html#ref-list-1>

Article cited in:

<http://bjo.bmj.com/content/82/1/9.full.html#related-urls>

Email alerting service

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Topic Collections

Articles on similar topics can be found in the following collections

[Retina](#) (1207 articles)

Notes

To request permissions go to:

<http://group.bmj.com/group/rights-licensing/permissions>

To order reprints go to:

<http://journals.bmj.com/cgi/reprintform>

To subscribe to BMJ go to:

<http://group.bmj.com/subscribe/>