Eye (2010) 24, 112–117

www.nature.com/eye

CLINICAL STUDY

Childhood sight impairment: a 10-year picture

JM Durnian, R Cheeseman, A Kumar, V Raja, W Newman and A Chandna

Abstract

Introduction **Registering a child as visually impaired is a potentially traumatic, but necessary, milestone in paediatric ophthalmology. Registration enables the provision of services essential to maximise the child's potential.**

Purpose **This study was carried out to investigate the changes over a 10-year period in the rates of registration of childhood blindness at a tertiary paediatric ophthalmology department. Particular attention was given to diagnosis, whether the disease was preventable, time to registration, age at registration, and the socioeconomic status of the patient's family.**

Methods **A retrospective analysis of all children registered blind or partially sighted over a 10-year period until December 2006.**

Results **A total of 256 children were registered blind or partially sighted over the 10 years. All cases were analysed. Of these, 58.2% were male and the average age at registration was 76 months; 52.0% were registered as severely sight-impaired. The most common primary diagnosis was cerebral visual impairment (CVI) in 27% cases, followed by optic atrophy in 16%, and the commonest anatomical site involved was the retina in 30.9%. An average of 25.6 (SD 8.0) registrations were carried out each year. The number of registrations per year is increasing. Seven cases (2.7%) were deemed avoidable and 61 cases (23.8%) were deemed potentially treatable. The mean index of multiple deprivation (IMD) score for the English cases (45.1) was significantly higher than of the surrounding area ($P<0.0001$)**

Conclusions **The yearly rates of registration increased slowly over the 10 years. The most common underlying cause for registration remains CVI, with the yearly proportion of registrations because of CVI not altering. The average age of registration was 6.3 years. A significant proportion of the cases of visual impairment, are because of potentially modifiable causes.**

Eye (2010) **24,** 112–117; doi:10.1038/eye.2009.32; published online 27 February 2009

Keywords: childhood blindness; paediatrics; cerebral visual impairment; sight impairment; registration

Alder Hey Children's NHS Foundation Trust, Merseyside, UK

Correspondence: A Chandna, Department of Paediatric Ophthalmology, Alder Hey Children's NHS Foundation Trust, Eaton Road, Liverpool, Merseyside L12 2AP, UK
Tel: +44 0151 228 4811; Fax: +44 0151 228 4812.
E-mail: arvind.chandna@alderhey.nhs.uk

Received: 7 March 2008
Accepted in revised form: 24 January 2009
Published online: 27 February 2009

Introduction

In 1992, the World Health Organization (WHO) estimated that nearly 1.5 million children in the world were blind or severely sight-impaired, with less than 5% of them being outside Africa, Asia, and Latin America.[1] In the UK, nearly 6 children per 10 000 live births will be diagnosed as being severely sight-impaired by their sixteenth birthday.[2] In the developed world, cerebral visual impairment (CVI) is the leading cause of vision loss in children;[2–5] however, the situation in the developing world is very different. The leading causes there, which are mainly avoidable by western standards, are: corneal scarring, cataract formation, and vitamin A deficiency.[6–9] The VISION2020 target for severe visual-loss in children is to reduce the global incidence from 0.75 to 0.4 per 1000 children by 2020, mainly by addressing these avoidable causes.[10,11] Is avoidable vision loss a problem in the United Kingdom?

As residents of the UK, we are more fortunate than those in the developing world in that if vision loss occurs, help is provided. From the appointment of 'The Royal Commission on the Blind, the Deaf, and Dumb' in 1886 to the introduction of the Certificate of Vision Impairment in 2005, formal recognition of sight impairment has been encouraged in the UK and has been used as an entry for any social service provisions available. A pilot paediatric form was introduced in 2007 (paediatric CVI extended version) by the Royal College of Ophthalmologists, allowing easier and more

precise classification of paediatric eye disease, hopefully allowing international comparison of data.

Registering a child as being sight impaired is potentially a traumatic event in ophthalmic clinical practice, often causing further anxiety in families in which there may be serious concurrent medical problems. However, it is a fundamental part of the treatment of these children, allowing them access to services that will help them fulfil their ultimate potential both in and out of school. When counselled correctly, this step can be the most helpful aspect provided by ophthalmologists in the care of these children.

Registration is a voluntary step, and lack of registration does not bar the child from accessing services, just makes it more difficult. The voluntary nature of registration means that data derived from any studies are open to bias and can be incomplete. Specifically, when measuring rates from across different departments, the differing attitudes to timing and suitability of registration can lead to selection bias. Apart from the methods used by the British Childhood Visual Impairment Study Group (BCVISG),[2] all studies relating to the incidence of visual loss are open to selection bias, whether because of a selected group of patients in a 'school for the blind', consultant interpretation of specific conditions, or patient unwillingness to be registered.

The aim of this study was to investigate the 10-year picture of visual impairment registration of children at our institution—a tertiary referral centre for paediatric ophthalmology in Merseyside. We report the epidemiology of these patients in addition to their clinical picture and investigate whether our patients differ from studies published earlier. We also report whether visual loss was potentially modifiable.

Materials and methods

We report the rates of sight impairment registration over a 10-year period, from 1997 to 2006, from the tertiary referral centre for paediatric ophthalmology at Alder Hey Children's NHS Foundation Trust. UNICEF defines a child as an individual aged less than 16 years; hence, although we deal with adolescents above this age, this definition was adopted. WHO defines blindness/severe sight impairment as best-corrected visual acuity in the better eye of less than 6/60; however, we included the presence of visual field defect (in older children) in the registration process.

We performed a retrospective review of all children who had been registered as severely sight impaired/ sight impaired from January 1997 to December 2006.

All patients were identified from internal databases and correspondence. Once a child had been registered, their details were entered into a departmental computer and copies of the registration documents were kept separately, meaning two systems were in place to ensure that all patients who had been registered were identified. All case notes were analysed, and in the minority of cases in which the notes could not be found, typed clinic letters were retrieved to investigate the patient's history.

We believe our study minimises bias as much as possible as; (1) all patients/families are intensively counselled over registration when required, and, in our experience, very rarely refuse registration; (2) consultant staff has been relatively stable over the period and all concerned are keen on the step being taken; (3) consultant input into every child's care means that all eligible patients are identified.

Once the patients had been identified, data were collected with respect to age at registration, time to registration, ethnic group (according to the Office of National Statistics' taxonomy), sex, socioeconomic group according to registered address at time of registration, diagnosis, and visual acuity/behaviour. Statistics are generally descriptive with Welch's *t*-test being used to compare the index of multiple deprivation (IMD) scores.

We recorded all diagnoses, but only report the primary diagnosis causing visual failure and classify it according the new guidelines issued by the Royal College of Ophthalmologists for paediatric patients (October 2007). In those cases of CVI or optic atrophy being listed as the cause of visual impairment, the underlying diagnosis is also reported.

Socioeconomic status was assigned using the patient's postcode at time of registration and the IMD 2004 for the English postcodes and the Welsh IMD 2005 for the Welsh postcodes. The two indices (IMD and WIMD) cannot be directly correlated with each other as there are no indices combining English and Welsh scores. The distribution of deprivation scores for the English children was compared with the deprivation score for the children of the North West of England as a whole and that of the Welsh children were compared with that of Wales as a whole.

The diagnosis was assigned depending on whether the underlying condition was modifiable, a phrase that can include both preventable and treatable causes. We adopted the methods used in the British Ophthalmological Surveillance Unit (BOSU) study allowing meaningful comparison, so that each case was documented as being (1) entirely unavoidable/ untreatable, (2) entirely preventable, or (3) the condition being potentially treatable.

In those cases of perinatal ischaemia and autodominant disease, arguments could be made as to whether these are modifiable or not. Perfect antenatal care may prevent perinatal ischaemia and brain cooling,[12] or allopurinol[13] may treat it, whereas genetic

counselling may prevent genetic disease. We made the decision to classify these conditions as unavoidable and untreatable to avoid deep ethical discussion. In some cases, the treatment of the primary condition—for example, optic chiasm glioma—may have led to visual loss; these were still classified as treatable.

Ethics Committee approval for the study was sought but deemed not to be necessary.

Results

During the 10 years from 1997 to 2006, 276 patients were registered as blind or sight impaired. Twenty of these cases were in adolescents aged over 16 and they have been excluded, leaving 256 children eligible (Table 1). One hundred and forty-nine cases were male (58.2%), 241 were white (94.1%), and the average age at registration was 76 months. Of the total, 133 (52.0%) were registered as being blind/severely sight impaired. There were 14 cases of siblings being registered in the 10 years.

A minority of case notes were not available for analysis and in these cases typed and stored clinic letters gave the required information. The primary diagnoses are listed in Table 2. Cerebral visual impairment (CVI) was the most common primary diagnosis with 69 (27.0%) cases affected by it. Optic atrophy (OA) was the second most common diagnosis with 41 cases (16.0%). Hence, combined CVI/OA accounted for 110 cases (43.0%) and Table 3 shows the underlying diagnoses for all cases of CVI and OA. The most common anatomical site for pathology causing visual impairment is the retina, 79 cases (30.9%), closely followed by visual pathways and cortex, 78 cases (30.5%). The rates of registration according to the anatomical site are shown in Figure 1.

Table 1 Patient characteristics

		Number	*%*
General 100.0	Children registered	256	
	Severely sight impaired	133	52.0
	Sight impaired	123	48.0
Sex	Male	149	58.2
	Female	107	41.8
Age (months)	Minimum age	1	
	Maximum age	192	
	Mean age (SD)	76 (55)	
Ethnicity	White	241	94.1
	Other	15	5.9
Socioeconomic Status			
	Mean IMD score-English	45.1 (SD 22.9)	
	Mean WIMD score-Welsh	20.7 (SD 15.2)	

IMD: index of multiple deprivation, WIMD: Welsh index of multiple deprivation.

We report an average of 25.6 registrations per year (SD 8.0); however, over the 10 years there is a gradual increase in the numbers registered per year (Figure 2) with the proportion of cases because of CVI/OA remaining relatively stable at 44.6% of registrations per year (SD 11%).

Visual acuity was measured in various ways over the period and, hence, is difficult to quantify together. Of the total, 121 cases had visual acuity equivalent to 6/60 or less, with 10 cases being documented as no light perception (NPL).

The mean IMD score for the English children ($n = 231$) was 45.1 (SD 22.9)—the higher the IMD score, the more deprived the area. The score for the entire North West region was 28.5 (SD 19.2, $n = 4460$), meaning that our English population has a significantly higher deprivation

Table 2 Causes of visual impairment

	Number	*%*
Total number children registered	256	100
Visual pathways & cortex		
Cerebral visual impairment	69	27.0
Other	3	1.2
Nystagmus	6	2.3
Whole globe and anterior segment		
Anophthalmos/microphthalmos	6	2.3
Anterior segment anomaly	6	2.3
Primary glaucoma	3	1.2
Amblyopia		
Strabismic	1	0.4
Refractive	1	0.4
Lens		
Cataract	9	3.5
Uvea		
Aniridia	7	2.7
Coloboma	9	3.5
Uveitis	1	0.4
Retina		
Retinopathy of prematurity	2	0.8
Retinal dystrophy	35	13.7
Retinoblastoma	1	0.4
Albinism	27	10.5
Retinal detachment	3	1.2
Other	11	4.3
Optic Nerve		
Hypoplasia	13	5.1
Optic atrophy	41	16.0
Other		
Uncertain	2	0.8

Table 3 Underlying causes for CVI/OA

Cerebral Visual Impairment (n = *69*)	
Perinatal ischaemia	16
Seizure disorder	14
Infection	7
Perinatal interventricular haemorrhage	7
Congenital brain malformation	7
Unknown	7
Premature birth	2
Rett syndrome	2
Familial metabolic encephalopathy	1
Hydrocephalus	1
Infantile respiratory arrest	1
Non-accidental injury	1
Post pertussis vaccine	1
Succinic semialdehyde dehydrogenase deficiency	1
Tuberous sclerosis	1
Optic Atrophy (n = *41*)	
Cerebral tumour	16
Hereditary	6
Hydrocephalus	3
Porencephalic cyst	2
Benign intracranial hypertension	1
Optic nerve glioma	1
Infantile Krabbe's disease	1
Cerebral infarct	1
Intraventricular haemorrhage	1
Leigh syndrome	1
Meningitis	1
Neurofibromatosis-1	1
ODI syndrome	1
Optic chiasm glioma	1
Perinatal ischaemia	1
Trauma	1
Tuberous sclerosis	1
Williams syndrome	1

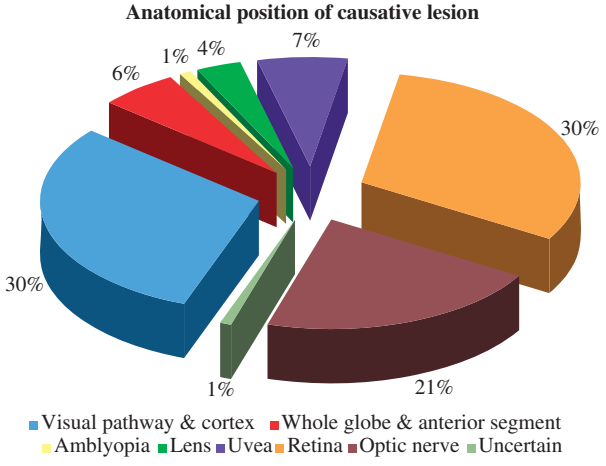

Figure 1 Registration rates according to the anatomical position of the causative lesion.

Table 4 Classification of modifiable causes

Entirely unavoidable/untreatable:	178 (69.5%)
Unknown underlying diagnosis:	9 (3.5%)
Entirely preventable:	
Total	7 (2.7%)
Amblyopia	2
ROP	2
Trauma	2
NAI	1
Potentially treatable:	
Total	61 (24.2%)
Brain tumour	21
Cataract	9
Infection	9
Hydrocephalus	5
Myopia	5
Sticklers	3
Glaucoma	3
Porencephalic cyst	2
Retinal haemorrhage	1
Uveitis	1
Retinoblastoma	1
Respiratory arrest	1

index than that would be expected for the area ($t = -12.7$, d.f. = 4689, $P < 0.0001$). For the Welsh children ($n = 24$), the WIMD mean was 20.7, (SD 15.2) with the score for Wales as a whole being 21.7 (SD 14.3, $n = 1896$), which is not a significant difference ($t = 0.34$, d.f = 1918, $P = 0.7338$). There was one patient from the Isle of Man for whom there was no deprivation score.

We deemed 68 cases (26.6%) of blind registrations to be either avoidable or treatable (Table 4). Seven cases (2.7%) were deemed entirely avoidable; two cases each of amblyopia, retinopathy of prematurity (ROP), and trauma, with a single case of non-accidental injury. Sixty-one cases were deemed treatable in some manner. In nine cases a unifying, underlying diagnosis was never found so these have been separated out.

Conclusions

This is the largest longitudinal study detailing vision impairment registration rates in children in the UK. The report by the BCVISG[2] details, in great depth, a snapshot of rates of visual impairment in 2000, but we aim to add to this body of knowledge by reporting the rates of registration over a period of time relatively consistently. There are always limitations when reporting visual impairment rates gained from registration forms of any type, but we feel that we have minimised these as far as possible for the reasons stated above. All cases registered for children aged between 0 and 16 have been included

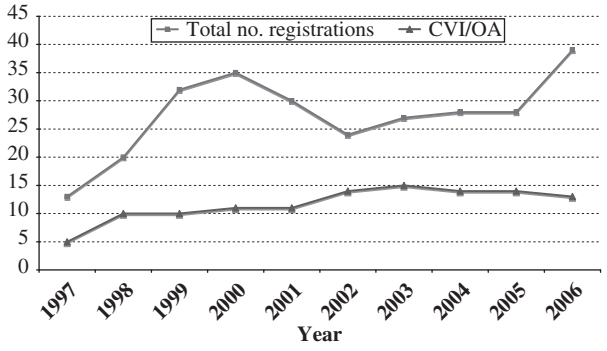

Figure 2 Yearly rates of registration.

and all notes have been analysed, giving a good insight into the trends of reporting over 10 years at our institution.

Our findings agree with the accepted fact of CVI/OA being the most common cause for visual loss in children in the developed world. The most recent survey of blind registration in the UK as a whole found that CVI/Optic nerve disorders were by far the most common underlying pathologies in the 0–15 age group, with 41.2% of registrations being attributed to this.[14] This is remarkably similar to our figure of 43.0%. The BCVISG report found CVI alone was the main cause of visual impairment in 48% of children.[2] However, if we take the causes of vision impairment according to anatomical location, we find that retinal pathology accounts for the highest proportion (30.9%) which, again, is in direct agreement with all literature published earlier. Retinal pathology encompasses two relatively large areas of childhood visual problems (retinal dystrophies and albinism) accounting for the large numbers.

The numbers of children registered each year do seem to be increasing as can be seen in Figure 2. Analysis of this is difficult because of the numbers of registrations each year being relatively small with the trend probably being artifactual because of the much lower number of registrations in 1997. It is well reported that childhood blindness is directly correlated with survival and if this trend does prove to be true, it may be because of improvements in paediatric care as a whole—although one would probably expect the numbers of CVI/OA cases to also increase.

We found that the vast majority of cases (94.1%) were of a White, British extraction. This is in disagreement with the BCVISG report, which found a higher proportion of registered children being from an ethnic background. This may be explained as, according to the 2001 Census, Merseyside having a higher proportion of White, British population (95.3%) than do England and Wales (91.3%).[15] Although we are a tertiary referral centre and take patients from outside the Mersey area, the majority of our patients are from Merseyside—184 of the 256 cases in our study. The difference in ethnicity can be attributed to the selection bias of a restricted population.

It has been widely reported that visual impairment is more common in those areas of higher deprivation and we confirm that for our study, with the added finding relating to our area of practice. We felt that comparisons of our study population to that of England as a whole would be inappropriate and a more accurate picture would be gained by comparison with the hospital catchment area. All of our English children came from the North West area so we believe that the approach used is valid. Those patients registered as sight impaired in England have a statistically significant higher chance of coming from a more deprived area. For our Welsh patients, when compared with those in Wales as a whole, there was no significant difference in deprivation index; this may be because of the numbers included (24) being too few for meaningful statistical analysis.

We found the average age of registration to be just over 6 years of age (76 months), which may seem to be surprisingly high. However, only a minority of children were born with a condition that can be instantly recognised as giving the child a definite poor prospect of vision. What is important is that the visual impairment was formally recognised at a time that would enable the proper services to be implemented to help the child fulfil his/her educational potential. When taking the step of registering a child as sight impaired, one must balance difficult subjects of; labelling the child, being realistic about the child's visual potential, allowing visual development to happen, allowing the child's parents to come to terms with the diagnosis and future ramifications, and enabling the correct services to be implemented at an opportune time. This combination is one that should be treated with caution in order to maintain a good working relationship with the parents and, as such, should not be rushed, even when clinical judgement may suggest that a baby will have extremely poor visual development. However, in the UK, formal recognition of poor visual function can aid the family in setting in place social service provision for family care, financial support, education, and so-called 'statementing', and this is often prompts the start of the registration process. If clinicians pre-empt this in children in whom the prospect of visual recovery is nil, then the entire process may be made slightly easier on these vulnerable families.

A significant proportion of our cases did fall into our classification of modifiable—26.6%. However, the vast majority of these cases are classified as treatable and non-preventable. We deemed only seven cases (2.7%) to be preventable. This is in close agreement with the BCVISG

study, which, if autosomal dominant diseases are discounted—as in our study—found 6% of childhood blindness being because of preventable causes. We report that 23.8% of our cases were potentially treatable, which is higher than reported elsewhere earlier. This may be because we have included brain tumours causing CVI/OA as potentially treatable. Our rationale for this was that early diagnosis of such lesions may have led to less morbidity after any treatment. In nine cases, no underlying diagnosis was found after extensive investigation by ourselves and the paediatricians. Seven of these cases had significant other neurological abnormalities and were registered as having CVI—these have been separated out from the analysis. However, it is safe to assume that these cases were unavoidable if no diagnosis was found after confirmation of profound visual loss.

In the developing world, the main pathology leading to blindness is corneal scarring as a result of either vitamin A deficiency, measles infection, ophthalmia neonatorum, or harmful traditional remedies.[11] There were no such cases in our cohort, so comparisons with the situation in the developing world would be impossible and worthless. Even so, we have shown that avoidable visual impairment does occur in the developed world, perhaps on a more subtle scale than elsewhere. We only found two cases of ROP in our cohort, which is much less than expected. This is likely to be because of the unusual situation in our area where the ROP screening, subsequent care, and any registration is done by a medical retina specialist outside of the paediatric service. However, the rates of blindness owing to ROP are falling in the developed world because of the success of the screening protocols. ROP is now becoming an important, avoidable cause of blindness in middle income countries owing to improvements in perinatal medicine.[16]

This is the largest longitudinal study detailing vision impairment registration rates in children in the UK. We report that there may be a subtle upward trend in registration numbers over the 10-year period. We have shown that the most common primary diagnosis is cortical visual impairment/optic atrophy, but pathology affecting the retina is the most common anatomical site leading to vision loss. Patients are being registered at an opportune time, balancing the child's needs, the scope for visual improvement, and diagnosis confirmation. Patients who are registered tend to be from more deprived areas and, in our region, are more likely to be from a White, English background. Over a quarter of all registrations are because of causes that are modifiable and as a medical community we must strive to reduce this number, as the remaining children have visual loss not amenable to intervention with current medical practices.

References

1 *Preventing blindness in children: report of a WHO/IAPB scientific meeting* World Health Organisation: Geneva 2000 (unpublished document WHO/PBL/00.77).
2 Rahi JS, Cable N. on behalf of the British Childhood Visual Impairment Study Group (BCVISG). Severe visual impairment and blindness in children in the UK. *Lancet* 2003; **362**: 1359–1365.
3 Rogers M. Vision impairment in Liverpool: prevalence and morbidity. *Arch Dis Child* 1996; **74**: 299–303.
4 Foster A. Childhood blindness. *Eye* 1988; **2**(suppl): 27–36.
5 Rosenberg T, Flage T, Hansen E. Incidence of registered visual impairment in the Nordic child population. *Br J Ophthalmol* 1996; **80**: 49–53.
6 Sitorus RS, Abidin MS, Prohartono J. Causes and temporal trends of childhood blindness in Indonesia: study at schools for the blind in Java. *Br J Ophthalmol* 2007; **91**: 1109–1113.
7 Muhit MA, Shah SP, Gilbert CE, Foster A. Causs of severe visual impairment and blindness in Bangladesh: a study of 1935 children. *Br J Ophthalmol* 2007; **91**: 989.
8 Titiyal JS, Pal N, Murthy GV, Gupta SK, Tanson R, Vajpayee RB *et al*. Causes and temporal trends of blindness and severe visual impairment in children in schools for the blind in North India. *Br J Ophthalmol* 2003; **87**: 941–945.
9 Kello AB, Gilbert C. Causes of severe visual impairment and blindness in children in schools for the blind in Ethiopia. *Br J Ophthalmol* 2003; **87**: 526–530.
10 *Preventing blindness in children* World Health Organization: Geneva 1999 WHO/PBL/00.77.
11 Gilbert C, Foster A. Childhood blindness in the context of VISION 2020–The right to Sight. *Bull World Health Organ* 2001; **79**: 227–232.
12 Jacobs S, Hunt R, Tarnow-Mordi W, Inder T, Davis P. Cooling for newborns with hypoxic ischaemic encephalopathy. *Cochrane Database Syst Rev* 2007; CD003311.
13 Chaudhari T, McGuire W. Allopurinol for preventing mortality and morbidity in newborn infants with suspected hypoxic ischaemic encephalopathy. *Cochrane Database Syst Rev* 2008; DC006817.
14 C Bunce, R Wormald. Causes of blind certications in England and Wales: April 1999–March 2000. *Eye* 2007 ; doi:10.1038/sj.eye.6702767 (advance online publication).
15 http://www.statistics.gov.uk/census2001/s104.asp
16 Gilbert C, Rahi J, Eckstein M, O'Sullivan J, Foster A. Retinopathy of prematurity in middle income countries. *Lancet* 1997; **350**: 12–14.