Vision impairment in Liverpool: prevalence and morbidity

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Abstract

A database related to the activities of the Liverpool vision assessment team was used to identify all children with vision impairment aged 0–16 years, resident in Liverpool, UK, on 1 April 1995. Prevalence rates were calculated for all children with vision impairment, and separately for two groups: those with uncomplicated vision impairment, and those with additional pathology. Visual tract pathologies were tabulated and compared. Associated handicapping conditions were defined and the extent of multiple disability was investigated for all vision impaired children, for very low birthweight children, and for those with cortical visual impairment. Of 199 children with vision impairment, 69 (35%) had uncomplicated impairment and 130 (65%) had additional and usually multiple pathology. There were 111 boys (56%); the excess of males was not statistically significant. Prevalence rates per 10 000 population were 18.1 for all vision impairment, 6.3 for uncomplicated vision impairment, and 11.8 for vision impairment complicated by additional pathology. Genetically determined disease accounted for over half the cases of uncomplicated vision impairment. Among the 130 children with additional pathology, cortical visual impairment was the commonest visual tract finding, affecting 64 (49%); 86% had learning difficulties; 53% had cerebral palsy. Multidisability (two or more disabling conditions in addition to vision impairment) affected half the entire childhood vision impairment population. These data should assist health and education authorities to determine the size of the vision impairment problem and how it relates to other disabilities in childhood, and can facilitate resource allocation and service planning. (*Arch Dis Child* 1996; 74: 299–303)

Keywords: childhood visual impairment, prevalence, morbidity, multihandicap.

education (Liverpool City Council) and health (Liverpool Health Authority) are coterminous, and the community and hospital paediatric services are now integrated within the Royal Liverpool Children's NHS Trust. The team aims to see, or at least to have information about, every vision impaired child in Liverpool, and its concern with functional development and educational aspects of vision is complementary to hospital based ophthalmology and paediatrics.

The recent setting up of a database (on a Psion 3a 'palm-top' computer) relating to the team's activities has made it possible to study both the prevalence and morbidity associated with childhood vision impairment. The dataset for each child includes administrative details and basic clinical information derived from the team's assessments, supplemented by access to clinical records at Alder Hey Children's Hospital (particularly those of paediatric ophthalmology, paediatric neurology, the Child Development Centre and community child health) and to education records.

Methods

CASE DEFINITION

A child with vision impairment has corrected vision sufficiently abnormal to interfere with development or to have ongoing educational implications. In cases where distance acuity can be measured and is relevant (see discussion below), this equates with acuity of 6/18 (Snellen) or worse in the better eye.

CASE ASCERTAINMENT

All visually impaired children known to the vision assessment team, resident in Liverpool on 1 April 1995, and aged 0–16 years were identified from the database by age, sex, and the absence or presence of pathology in addition to visual tract dysfunction. On this date there were a few cases of vision impairment not known to the team: these might have included very young children not yet referred, vision impairment of insidious onset in school age children, and those who had very recently moved into the city. Ascertainment was therefore a slight underestimate.

PREVALENCE

The total 0–16 year population in Liverpool was calculated from the Office of Population and Census Surveys (OPCS) mid-1993 population estimates, which were the most recent available. Prevalence rates in this study, and those

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The Liverpool vision assessment team^{1–3} was set up in 1976 to provide multidisciplinary assessment and support of children with vision impairment. The core members of the team are a senior educational psychologist, the senior advisory teacher of visually impaired children, and a consultant community paediatrician: this structure facilitates close links between health and education services. The geographical boundaries of

quoted from elsewhere, are all per 10 000 population.

VISUAL TRACT MORBIDITY

The clinical features of dysfunctional vision were tabulated separately for children with uncomplicated vision impairment and for those with additional pathology. Sex distribution was determined overall and among children with the most frequently occurring visual tract disorders.

NON-VISUAL MORBIDITY

Additional pathology was investigated in terms of six broad diagnostic categories – (1) cerebral palsy; (2) hydrocephalus; (3) hearing loss; (4) epilepsy; (5) learning difficulties; (6) other relevant conditions – using the database supplemented by medical and educational records. Cerebral palsy, hydrocephalus, and epilepsy were clinical diagnoses made by appropriate paediatric specialists. Hearing loss was defined as loss of 60 dB or more in the better ear, which is the local criterion for receiving specialist advisory support in school. Learning difficulties comprised (a) developmental delay in preschool children deemed likely to indicate later severe or moderate learning difficulties, (b) severe or moderate learning difficulties in older children as indicated in statements of special educational need. 'Other relevant conditions' covered causes of appreciable dysfunction (judged subjectively) not covered in the first five categories (see table 5). Emotional and behavioural problems were excluded from this classification because of difficulties with categorisation and assessment.

The number of categories of additional pathology was determined for each child, and tabulated for all children and for those of very low birthweight (VLBW, <1500 g) and with cortical visual impairment. The educational placements of children with and without additional non-visual pathology were also compared.

Results

One hundred and ninety nine visually impaired children were identified, of whom 111 (56%) were boys: the male/female ratio was not statistically significant ($p=0.14$, sign test). Sixty

nine children (35%) had uncomplicated vision impairment; the remaining 130 (65%) had additional pathology.

PREVALENCE

The 0–16 years population of Liverpool was 110 200. Prevalence rates for visual impairment were 18.1 for all vision impaired children, 6.3 for those with uncomplicated vision impairment, and 11.8 for children with vision impairment and additional pathology.

VISUAL TRACT MORBIDITY

The causes of visual dysfunction in 69 children with uncomplicated vision impairment are shown in table 1. All had a clear cut ophthalmological diagnosis accounting for visual impairment. Secondary manifestations (for example, non-idiopathic nystagmus, coincidental refractive error, and so on) have not been listed. Seven of the nine children with uncorrectable refractive error had high myopia. At least 39 (56%) of this group had genetically determined disease (genetic data were not available for all relevant cases).

Findings related to visual dysfunction in the 130 children with vision impairment and additional pathology are shown in table 2. A few children had a straightforward diagnosis accounting for impaired vision, but this group was characterised by multiple vision related findings, the relative contributions of each of which to the resulting visual handicap were often unclear. Cortical visual impairment was the commonest single diagnosis, affecting 64 children (49%). Nine of the 13 children with visual field defects had hemianopia associated with hemiplegic cerebral palsy.

SEX DISTRIBUTION

Table 3 shows distribution by sex for the most frequently identified causes of visual problems in the two groups. Nine out of 13 children with hereditary retinopathy, and 15 out of 27 with optic atrophy, were girls. Otherwise boys predominated, most notably in congenital idiopathic nystagmus (10 of 11).

Table 1 Causes of visual dysfunction in Liverpool children aged 0–16 years with uncomplicated vision impairment

	Number	Per cent
Albinism	15	22
Hereditary retinopathy	13	19
Congenital idiopathic nystagmus	11	16
Refractive error	9	13
Congenital cataract	6	9
Retinopathy of prematurity	5	7
Tumour	2	3
Microphthalmia + cataract	2	3
Optic atrophy	2	3
Various*	4	5
Total	69	100

*One case each of optic nerve hypoplasia, optic neuritis, coloboma, keratitis.

Table 2 Findings related to visual dysfunction in Liverpool children aged 0–16 years with vision impairment and additional pathology: n=130

	Number*	Per cent*
Cortical visual impairment	64	49
Nystagmus	29	22
Refractive error	28	22
Optic atrophy	27	21
Squint	23	18
Visual field defect	13	10
Retinopathy	6	5
Cataract	6	5
Amblyopia	5	4
Retinopathy of prematurity	5	4
Optic nerve hypoplasia	4	3
Tumour	4	3
Anophthalmia/microphthalmia	3	2
Ptosis	3	2
Coloboma	2	1.5
Various†	4	3

*More than one finding applied to each child in many cases
†One case each of corneal opacity, albinism, glaucoma, gaze paresis.

Table 3 Sex distribution of commonest causes of visual dysfunction, in Liverpool children with vision impairment aged 0–16 years. Values are numbers of cases

	Male	Female
<i>Uncomplicated vision impairment</i>		
Albinism	8	7
Hereditary retinopathy	4	9
Congenital idiopathic nystagmus	10	1
Refractive error	5	4
<i>Vision impairment with additional pathology</i>		
Cortical visual impairment	35	29
Nystagmus	16	13
Refractive error	18	10
Optic atrophy	12	15

NON-VISUAL MORBIDITY

Table 4 shows the numbers of children with conditions falling into each of the six diagnostic categories of additional non-visual pathology. Multiple pathology was common (see table 6). Of the 112 children with learning difficulties (56% of all children with vision impairment), 71 (63%) had severe and 18 (16%) moderate learning difficulties, and 23 children of pre-school age (21%) had clinically ascertained developmental delay. Thirteen (54%) of the 24 children with hydrocephalus had associated optic atrophy. Eleven children had educationally significant hearing loss. There were no cases of congenital rubella syndrome.

MULTIPLE PATHOLOGY

Table 6 shows how many of the six categories of additional non-visual pathology applied to each child. Ninety eight children (49% of all with vision impairment) suffered from at least two other disabling conditions. In 54 children vision impairment was combined with cerebral palsy and severe learning difficulties; 30 of these also had epilepsy.

Morbidity associated with very low birthweight

There were 22 VLBW children (11% of all children with vision impairment). Mean birthweight was 926 g (range 622–1460 g); mean gestation was 26.5 weeks (range 24–30 weeks). Relevant visual tract findings are shown in table 7.

Ten children (45% of the VLBW group) developed vision impairment as a result of retinopathy of prematurity (ROP), despite cryotherapy or laser photocoagulation in seven. Five had uncomplicated vision impairment which was severe in all (for example, requiring teaching by non-sighted methods).

Of the 12 children (55%) whose vision impairment was not ROP related, seven

Table 4 Non-visual pathology associated with vision impairment in Liverpool children aged 0–16 years: numbers of children with conditions in the six diagnostic categories shown: *n* = 130

Diagnostic category	Number*	Per cent*
Cerebral palsy	69	53
Hydrocephalus	24	18
Hearing loss	11	8
Epilepsy	47	36
Learning difficulties	112	86
Other†	32	25

*Multiple pathology was common: see table 6. †See table 5.

Table 5 Non-visual pathology associated with vision impairment in Liverpool children aged 0–16 years: 'other' pathology not covered in its dysfunctional consequences by the remaining five diagnostic categories (see table 4) *n* = 32. Values are numbers of cases

Severe respiratory disease	6
Asthma (frequent inpatient treatment)	3
Bronchopulmonary dysplasia (severe)	2
Repeated severe infections	1
Cleft lip and palate	5
Endocrine disorder	4
Post-tumour	3
Post-meningitis	1
Severe feeding difficulties	3
Gastrostomy	2
Nasogastric feeding	1
Scoliosis	3
Meningomyelocoele	2
Facial abnormalities	2
Limb abnormalities	2
Paraplegia	2
Various*	8
Total	37†

*One case each of: agenesis of corpus callosum, CHARGE association, congenital cardiac defect, Fanconi syndrome, Prader-Willi syndrome, neurofibromatosis, autism, hydrotic ectodermal dysplasia.

†Five children had multiple 'other' pathology.

developed ROP which regressed (following treatment in two). The remaining five children, all born before 1988, did not have eye examinations by an ophthalmologist until visual dysfunction became manifest; transitory ROP could therefore have been missed. All these 12 children had additional pathology.

Fourteen (64%) of the VLBW group had ultrasound evidence of intraventricular haemorrhage (IVH). Eight had hydrocephalus (shunted in seven), of whom four developed optic atrophy. One child developed a large porencephalic cyst following IVH. The overall pattern of multiple disability in VLBW children is shown in table 6. Five (23%) had uncomplicated vision impairment, and 12 (55%) had two or more additional disabilities.

Morbidity associated with cortical visual impairment

The distribution of additional pathology among the 64 children with cortical visual impairment is shown in table 6. This was a severely disabled group: none had uncomplicated vision impairment, and 34 (53%) had three or more other pathologies. Sixty two (97%) had established learning difficulties. Twenty six (41%) had evidence of anterior

Table 6 Morbidity in Liverpool children aged 0–16 years with vision impairment (VI): showing numbers and percentages with no additional pathology (0) and with 1–5 additional disabling pathologies (see table 4): for all children with VI, very low birthweight (VLBW) children, and those with cortical visual impairment (CVI)

No of additional pathologies	All children with VI		VLBW children with VI		Children with CVI	
	No	%	No	%	No	%
0	69	35	5	23	0	0
1	32	16	5	23	7	11
2	42	21	4	18	23	36
3	42	21	7	32	25	39
4	10	5	1	4	7	11
5	4	2	0	0	2	3
Total	199	100	22	100	64	100

Table 7 Visual tract findings causing or contributing to vision impairment in very low birthweight children aged 0–16 years in Liverpool: *n*=22. Values are numbers of cases*

Retinopathy of prematurity	10
Myopia	6
Optic atrophy	5
Visual field defect (hemianopia)	4
Nystagmus	3
Cortical visual impairment	2

*More than one finding applied to each child in some cases.

visual pathway disease in addition to cortical visual impairment.

EDUCATIONAL PLACEMENT

The education environment in Liverpool emphasises mainstream integration of children with vision impairment wherever possible. As table 8 shows, whereas 70% of children with uncomplicated vision impairment received mainstream education, only 13% of the children with additional pathology did so: in fact only two children of secondary school age with multiple disabilities were attending mainstream schools.

Discussion

It is usual^{4–6} and, on the face of it, straightforward to define vision impairment in terms of distance acuity, but there are drawbacks. In the first place, despite improved techniques such as preferential looking, Keeler cards, and Cardiff acuity cards, many children with vision impairment are too young or too disabled to cooperate sufficiently to obtain measurements. Thirty six per cent of children in this study were under five, 56% had learning difficulties, and 49% had two or more additional disabilities (these groups, of course, overlap considerably). In a West Sussex study of children with vision impairment 46% 'were unable to perform formal tests of visual acuity'.⁵ A case definition which cannot be applied to almost half a study population is at best of limited value.

Secondly, an emphasis on distance acuity diverts attention from other aspects of visual function such as near vision and accommodation,⁷ eye movements, visual fields, and contrast sensitivity. All of these, alone or in combination, may cause or contribute to a child's visual difficulties, especially at school.

Unless and until succinct quantifiable criteria emerge, which can be applied at all ages and across the spectrum of childhood disability, an overarching criterion of developmental and

educational dysfunction, consistently applied with regular case review, remains the most reliable approach to case ascertainment.

Prevalence of vision impairment has probably been underestimated in the past. This is partly explained by the use of social security registration data^{5 8 9}: the criteria for registration do not cover all children with vision impairment, and some children who would qualify are not registered. Riise *et al*⁴ reported prevalence among Danish 0–15 year olds of 4.1, but although ascertainment was closely linked to registration (Hyvarinen L, personal communication), many children with educationally significant vision impairment would have been excluded by the case definition of 3/60 or worse visual acuity in the better eye. In the West Sussex study,⁵ an overall prevalence rate of 6.5 suggests under-reporting, unless impaired vision is less common in West Sussex than in Liverpool. The Royal National Institute for the Blind (RNIB) analysed returns from 33 local education authorities; the mean prevalence was 10.3, with a range from 1.8 to 26.2.¹⁰ OPCS, using census data and household sampling, suggested an overall rate of 20,¹¹ which is in line with this study.

Surveys which distinguish children with additional pathology have reported proportions varying between 33% and 50% in different Nordic countries,⁴ 61% in West Sussex,⁵ and 56% in the RNIB survey.¹⁰ OPCS estimated that 83% of children with a sight problem had an additional disability.¹¹

The preponderance of boys probably reflects the increased vulnerability of boys to pathological insult. Otherwise, in terms of individual clinical conditions, numbers were too small for more detailed conclusions to be drawn, except that X linked inheritance of congenital idiopathic nystagmus presumably explained the 10:1 excess of boys with this condition. The sex ratio reached statistical significance in the national registers of Denmark and Finland,¹² ascribed to a combination of X linked inheritance and undetermined prenatal and perinatal factors.

Comparison of the causes of vision impairment in different studies is complicated by differing methodologies and diverse patterns of aetiology in different parts of the world.¹³ Among Nordic children¹⁴ optic atrophy, ROP, cerebral amblyopia, and congenital cataract were identified as the most important causes. In a recent study of blind children in Chile,¹⁵ 30% had hereditary disease and 18% had vision impairment caused by ROP.

ROP continues to be a major cause of concern,^{13–17} but it accounted for less than half the cases of vision impairment associated with VLBW in Liverpool, and it is important to be clear that the issue is vision impairment caused by ROP, not the incidence or prevalence of ROP itself. The risk of ROP occurring is inversely related to birthweight and gestation,^{13 16} and the survival of more very small babies is bound to mean that cases of ROP continue to occur. Spontaneous regression is common, however, and in selected cases

Table 8 Educational placement of children with vision impairment (VI) aged 0–16 years in Liverpool: showing numbers and percentages under 5 (preschool), and in mainstream and special education.

	Uncomplicated VI		VI with additional pathology	
	No	%	No	%
Preschool	9	13	15	12
Mainstream	48	70	17	13
Special	12	17	98	75
Total	69	100	130	100

treatment seems to be effective.^{16 17} Vision impairment caused by ROP is not inevitable, and can be kept to a minimum by ophthalmological monitoring as an integral part of neonatal intensive care.¹⁶ It should never be assumed that a history of ROP accounts for any subsequent vision problems, which are at least as likely to be caused by sequelae of VLBW such as intraventricular haemorrhage.¹⁸ Myopia is also a well recognised complication of prematurity.¹⁹

The increasing prevalence of cortical visual impairment is probably both apparent, in the sense of increased professional awareness, and real because of the survival of more children with brain damage. Jan has commented that the study of cortical visual impairment 'is still in its infancy',²⁰ although the functional implications for affected children are beginning to be addressed.^{21 22}

Neither vision impairment itself nor most of the disabling conditions used to analyse coexistent non-visual pathology are precise clinical concepts, and each is associated with a spectrum of dysfunction ranging from borderline normality to severe disability. The survey of morbidity in this study is broad brush and descriptive, delineating and to some extent quantifying the concept of 'multidisabled visual impairment' (MDVI) which is already widely used in education. Comparison with further United Kingdom and European studies might show significant geographical variations in, for example, genetically determined eye disease. Well validated special needs registers, once these are achieved, could be used for this purpose.²³ They could also address issues not covered by this study, for example the numbers and proportions of children with cerebral palsy, learning difficulties, hydrocephalus, and so on, who are also visually impaired. This would help to place vision impairment realistically in the broader context of childhood disability.

Assuming that Liverpool's experience is typical of the United Kingdom, what are the practical implications? Reliable prevalence rates and an overview of associated morbidity should be used to inform service planning, resource allocation, training, and working practices. As examples, commissioners and providers of health care can plan on the basis that about two per 1000 children have impaired vision, and the majority of these have complex disabilities: education authorities should provide specialist teacher support and equipment accordingly; community paediatricians need to acquire experience of vision impairment so that they can contribute; and cooperative and complementary working

relations between ophthalmologists and paediatricians in this field are a necessity, not a desirable optional extra.

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