

Epidemiology of cerebral palsy in England and Scotland, 1984–9

P O D Pharoah, T Cooke, M A Johnson, R King, L Mutch

Abstract

Aims—To report on the epidemiology of cerebral palsy in England and Scotland, to provide information on the prevalence of cerebral palsy and the severity of the disability or any co-morbidity.

Methods—Cerebral palsy registers were compiled from multiple sources of ascertainment covering all of Scotland and the counties of Merseyside, Cheshire, Berkshire, Buckinghamshire, Northamptonshire and Oxfordshire in England. All cases of cerebral palsy born in 1984 to 1989, to mothers resident in the area, were included. Denominator number of live births and neonatal deaths for determining birthweight specific prevalence were obtained from birth and death registrations. Learning, manual, and ambulatory disabilities were graded for severity. Any co-existing sensory (hearing or visual) morbidity was also graded for severity.

Results—There were 789 411 live births in 1984–9, with 3651 neonatal deaths (neonatal mortality 4.6 per 1000 live births) and 1649 cases of cerebral palsy—a cerebral palsy prevalence of 2.1 per 1000 neonatal survivors. The birthweight specific cerebral palsy prevalence ranged from 1.1 per 1000 neonatal survivors in infants weighing ≥ 2500 g to 78.1 in infants weighing < 1000 g. There was no significant time trend in prevalence of cerebral palsy in any of the birthweight groups, in contrast to the fall in neonatal mortality observed in all birthweight groups. Of the 1649 cases of cerebral palsy, 550 (33.4%) had severe ambulatory disability (no independent walking), 390 (23.7%) had severe manual disability (incapable of feeding or dressing unaided), 381 (23.1%) had severe learning disability (IQ < 50), 146 (8.9%) had severe visual disability (vision $< 6/60$ in the better eye) and 12 had severe hearing disability (> 70 dB loss).

Conclusions—Registers fill an important gap left by the lack of routine data on the prevalence of disability in children. The ability to record trends in the prevalence and the severity of the disability should inform those who have responsibility for providing services for children with disabilities.

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Information on the prevalence and epidemiology of disability in children is not available from routine health service data systems. The Mersey Region Cerebral Palsy Register, the Oxford Region Register of Early Childhood Impairments, and the Scottish Register of Children with a Motor Deficit of Central Origin were developed with the intention of filling this gap in the routine data systems.¹ The impetus for this came from the technological advances in neonatal intensive care and the improving prognosis for survival of low birthweight infants^{2,3} among whom there was an increased prevalence of cerebral palsy. Even infants weighing < 1000 g at birth are now more likely to survive the neonatal period.⁴ Associated with improving survival among progressively lower birthweight infants there have been reports from different countries of increasing birth prevalence rates of cerebral palsy particularly among low birthweight infants.^{5–8}

The overlapping time period covered by the Mersey, Oxford, and Scottish registers has allowed the time trends and epidemiology of cerebral palsy to be examined, including the severity of the motor disability and the prevalence and severity of co-disabilities of cognition, vision, and hearing, that covers the total population of births in Scotland and over 10% of births in England from 1984 to 1989.

Methods

The Mersey Register covers the counties of Merseyside and Cheshire, and includes all children with cerebral palsy born to mothers resident in those counties at the time of the child's birth. The Oxford register similarly covers the counties of Berkshire, Buckinghamshire, Northamptonshire and Oxfordshire.⁹ The Scottish register is national, covering the whole of Scotland.¹⁰

The ascertainment of cases for the Mersey and Oxford registers was abstracted from several sources of data including: assessment centres, special schools, "handicap" and special needs registers; the York register of children claiming Family Disability Allowance; the Office for National Statistics for infants in whom cerebral palsy was recorded as a cause of death; and from regular reporting from physiotherapy departments, consultant and community paediatricians, and paediatric neurologists.

In Scotland case finding relied on the routine health services reporting of cases. This was usually by community paediatricians, particularly those involved in recording children as having special needs, those dealing with special schools, by some assessment centres and by one paediatric neurology clinic. Unlike the Mersey and Oxford regions, a dedicated

FSID Unit of
Paediatric and
Perinatal
Epidemiology,
Department of Public
Health,
University of Liverpool
Liverpool L69 3GB
P O D Pharoah
T Cooke

National Perinatal
Epidemiology Unit,
Radcliffe Infirmary,
Oxford
M A Johnson
R King

Public Health
Research Unit,
University of Glasgow
Glasgow
L Mutch

Correspondence to:
Professor Pharoah.

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Table 1 Birthweight specific trends in cerebral palsy prevalence*

Year	Number of live births	Number of neonatal deaths	Neonatal mortality rate per 1000 livebirths (95% CI)	Number of survivors	Number with cerebral palsy	Prevalence of cerebral palsy per 1000 neonatal survivors (95% CI)
<i>Birthweight <1000 g</i>						
1984	367	193	527.3 (474.8–579.4)	174	9	51.7 (23.9–95.9)
1985	361	205	567.9 (515.0–619.6)	156	10	64.1 (31.2–114.7)
1986	397	208	523.8 (473.5–574.0)	189	18	95.2 (57.4–146.3)
1987	403	197	488.8 (439.0–538.8)	206	21	101.9 (64.2–151.6)
1988	391	175	447.6 (397.6–498.4)	216	14	64.8 (35.9–106.4)
1989	385	174	451.9 (401.5–503.2)	211	18	85.3 (51.3–131.5)
1984–89	2304	1152	500.0 (479.4–520.6)	1152	90	78.1 (63.3–95.2)
<i>Birthweight 1000–1499 g</i>						
1984	760	115	151.3 (126.6–178.8)	645	41	63.6 (46.0–85.2)
1985	752	109	144.9 (120.6–172.2)	643	38	59.1 (42.2–80.2)
1986	756	97	128.3 (105.3–154.3)	659	44	66.8 (48.9–88.6)
1987	814	108	132.7 (110.1–157.9)	706	47	66.6 (49.3–87.5)
1988	839	117	139.5 (116.7–168.8)	722	44	60.9 (44.6–81.0)
1989	781	93	119.1 (97.2–143.9)	688	53	77.0 (58.2–99.6)
1984–89	4702	639	135.9 (126.2–146.0)	4063	267	65.7 (58.3–73.8)
<i>Birthweight 1500–2499 g</i>						
1984	7133	153	21.5 (18.2–25.1)	6980	67	9.6 (7.4–12.2)
1985	7524	129	17.1 (14.3–20.3)	7395	74	10.0 (7.9–12.5)
1986	7490	112	15.0 (12.3–18.0)	7378	80	10.8 (8.6–13.5)
1987	7514	102	13.6 (11.1–16.5)	7412	75	10.1 (8.0–12.7)
1988	7360	106	14.4 (11.8–17.4)	7254	84	11.6 (9.2–14.3)
1989	7156	124	17.3 (14.4–20.6)	7032	63	9.0 (6.9–11.4)
1984–89	44177	726	16.4 (15.3–17.7)	43451	443	10.2 (9.2–11.2)
<i>Birthweight ≥2500 g</i>						
1984	119767	209	1.7 (1.5–2.0)	119558	140	1.2 (1.0–1.4)
1985	122199	199	1.6 (1.4–1.9)	122000	154	1.3 (1.1–1.5)
1986	122167	209	1.7 (1.5–2.0)	121958	151	1.2 (1.0–1.5)
1987	123887	182	1.5 (1.3–1.7)	123705	149	1.2 (1.0–1.4)
1988	126284	182	1.4 (1.2–1.7)	126102	128	1.0 (0.8–1.2)
1989	120761	150	1.2 (1.1–1.4)	120611	118	1.0 (0.8–1.2)
1984–89	735065	1131	1.5 (1.4–1.6)	733934	840	1.1 (1.1–1.2)

*There were nine children for whom birthweight was not recorded.

research assistant did not maintain the register and any difference in the completeness of ascertainment could be attributed to this. Deaths with a mention of cerebral palsy were reported from the Office of the Registrar General for Scotland.

Denominator numbers of live births and neonatal deaths for determining the birthweight specific prevalence of cerebral palsy were obtained from the legally required birth and death registrations.

Four clinical types of cerebral palsy were recognised: hemiplegia; diplegia; quadriplegia; and "other," which included the ataxic, dyskinetic, hypotonic and mixed varieties of the syndrome. Assignment to a clinical category in Oxford and Scotland was made from the reporting form which was based on the standard format for recording motor deficit of central origin.¹¹ In Mersey the clinical type of cerebral palsy was that recorded by the paediatrician in the case notes and no attempt was made to standardise the recording of signs.

Levels of functional disability were categorised for ambulatory, manual, learning, hearing and visual disabilities. For the Mersey register, after a child was ascertained as having cerebral palsy, paediatric records were subsequently abstracted to obtain details of any functional disability.

Ambulatory disability was classified as "none" if fluent and stairs could be climbed without difficulty; "mild" if ambulation was limited and non-fluent, if there was difficulty in climbing stairs; "moderate" if there was obvious reduced mobility and/or the child required walking aids; and "severe" if there was no independent walking even with aids.

Manual disability was classified as "none" if there was no apparent problem; "mild" if the child had some difficulty using both hands together but could feed or dress unaided; "moderate" if feeding and dressing unaided could be done only with considerable difficulty and "severe" if the child was incapable of feeding or dressing unaided.

Various methods of assessment were used for ascertaining the IQ of the child. The IQ was used to categorise the level of learning disability and, if an assessment of IQ had been made on more than one occasion, the most recent assessment was used for analysis. For the Oxford cases, formal IQ assessments were not recorded but paediatricians were asked whether there was minor, moderate, or severe learning disability. In the Scottish register intellectual impairment was sometimes assessed using Griffiths developmental assessment¹² and occasionally was determined as an IQ by a psychologist, but was often a general view based on observation of the child by the examining doctor or teacher, grading the children as mildly, moderately, or severely delayed. In Merseyside most children had had a formal assessment by a psychologist. The assumption was made that minor or no learning disability was equivalent to an IQ >70, moderate learning disability was equivalent to an IQ in the range 50 to 70, and severe learning disability corresponded to an IQ <50.

Visual disability was classified as "mild" if vision was >6/18 in the better eye, "moderate" if 6/18 to 6/60 in the better eye, "severe" if <6/60 in the better eye or there was no useful vision.

Table 2 Clinical type of cerebral palsy by birthweight group

Type of cerebral palsy	<1000 g	1000–1499 g	1500–2499 g	≥2500 g	*All birthweights
	n (%)	n (%)	n (%)	n (%)	n (%)
Hemiplegia	24 (26.7%)	53 (19.9%)	88 (19.9%)	305 (36.3%)	470 (28.5%)
Diplegia	14 (15.6%)	78 (29.2%)	117 (26.4%)	79 (9.4%)	289 (17.5%)
Quadriplegia	31 (34.4%)	101 (37.8%)	165 (37.2%)	274 (32.6%)	575 (34.9%)
Other [†]	21 (23.3%)	35 (13.1%)	73 (16.5%)	182 (21.7%)	315 (19.1%)
All CP	90 (100%)	267 (100%)	443 (100%)	840 (100%)	1649 (100%)

χ^2 , 9DF = 121.9; $p < 0.0001$.

[†]Includes nine cases in whom the birthweight was not recorded.

[‡]Includes 20 cases with type of cerebral palsy not recorded.

Hearing loss was averaged across frequencies 0.5–4 kHz and the disability classified as “mild” if hearing loss was 21–45 dB, “moderate” for a 45–70 dB loss, “severe” with a hearing loss >70 dB. Assessment of accurate hearing levels was often impossible in children with concomitant intellectual impairment.

The cases for whom disability levels were recorded as “not known” were predominantly those who had died before the level of disability could be assessed. Inevitably, this will be a biased group because it would contain a disproportionate number who were more severely disabled.

In spite of the differing methods of ascertainment of cases in the three regions and the minor differences in categorising the severity of the disabilities, the results were similar for each region and it was felt to be justified to combine the three datasets.

The statistical tests used were either the normal approximation to the binomial for the difference between two proportions, the χ^2 test or the χ^2 test for trend.

Results

During 1984–9, there were 196 439 live births in the Mersey Region, 204 250 in the Oxford Region, and 388 722 in Scotland. The total live birth population covered by the three registers was therefore 789 411 among which there were 3651 neonatal deaths and 1649 cases of cerebral palsy among the survivors, giving a neonatal mortality rate of 4.6 per 1000 live births and a cerebral palsy prevalence of 2.1 per 1000 neonatal survivors.

Of the total 1649 cases of cerebral palsy, 840 (50.9%), 443 (26.9%), 267 (16.2%) and 90 (5.5%) were of birthweights (2500 g, 1500–2499 g, 1000–1499 g and <1000 g, respectively. In nine cases the birthweight was not known.

Table 1 shows the birthweight specific trends in the prevalence of cerebral palsy. There was no discernible trend over this short period of time in any of the birthweight groups. In the lowest birthweight group (<1000 g), an early rise was succeeded by a fall in prevalence but the small number of cases each year means that

Table 3 Severity of functional disability by birthweight group

	<1000 g	1000–1499 g	1500–2499 g	≥2500 g	*All birthweights
<i>Ambulatory disability:</i>					
None	5 (5.6%)	10 (3.7%)	21 (4.7%)	64 (7.6%)	102 (6.2%)
Mild	30 (33.3%)	97 (36.3%)	162 (36.6%)	333 (39.6%)	624 (37.8%)
Moderate	16 (17.8%)	64 (24.0%)	102 (23.0%)	144 (17.1%)	327 (19.8%)
Severe	35 (38.8%)	90 (33.7%)	147 (33.2%)	275 (32.7%)	550 (33.4%)
Not known	4 (4.4%)	6 (2.2%)	11 (2.5%)	24 (2.9%)	46 (2.8%)
Total	90 (100%)	267 (100%)	443 (100%)	840 (100%)	1649 (100%)
<i>Manual disability:</i>					
None	15 (16.7%)	49 (18.4%)	78 (17.6%)	83 (9.9%)	226 (13.7%)
Mild	30 (33.3%)	94 (35.2%)	150 (33.9%)	317 (37.7%)	593 (36.0%)
Moderate	22 (24.4%)	70 (26.2%)	102 (23.0%)	187 (22.3%)	385 (23.3%)
Severe	19 (21.1%)	46 (17.2%)	98 (22.1%)	226 (26.9%)	390 (23.7%)
Not known	4 (4.4%)	8 (3.0%)	15 (3.4%)	27 (3.2%)	55 (3.3%)
Total	90 (100%)	267 (100%)	443 (100%)	840 (100%)	1649 (100%)
<i>Learning disability:</i>					
None/mild	45 (50.0%)	157 (58.8%)	223 (50.3%)	413 (49.2%)	839 (50.9%)
Moderate	16 (17.8%)	43 (16.1%)	72 (16.3%)	148 (17.6%)	283 (17.2%)
Severe	18 (20.0%)	38 (14.2%)	81 (18.3%)	241 (28.7%)	381 (23.1%)
Not known	11 (12.2%)	29 (10.9%)	67 (15.1%)	38 (4.5%)	146 (8.9%)
Total	90 (100%)	267 (100%)	443 (100%)	840 (100%)	1649 (100%)
<i>Hearing disability:</i>					
None/mild	71 (78.9%)	245 (91.8%)	408 (92.1%)	789 (93.9%)	1520 (92.2%)
Moderate	5 (5.6%)	3 (1.1%)	3 (0.7%)	5 (0.6%)	16 (1.0%)
Severe	9 (10.0%)	5 (1.9%)	5 (1.1%)	12 (1.4%)	31 (1.9%)
Not known	5 (5.6%)	14 (5.2%)	27 (6.1%)	34 (4.0%)	82 (5.0%)
Total	90 (100%)	267 (100%)	443 (100%)	840 (100%)	1649 (100%)
<i>Visual disability:</i>					
None/mild	56 (62.2%)	193 (72.3%)	293 (66.1%)	598 (71.2%)	1145 (69.4%)
Moderate	14 (15.6%)	41 (15.4%)	80 (18.1%)	106 (12.6%)	241 (14.6%)
Severe	14 (15.6%)	16 (6.0%)	38 (8.6%)	77 (9.2%)	146 (8.9%)
Not known	6 (6.7%)	17 (6.4%)	32 (7.2%)	59 (7.0%)	117 (7.1%)
Total	90 (100%)	267 (100%)	443 (100%)	840 (100%)	1649 (100%)

*Includes nine cases in whom the birthweight was not recorded

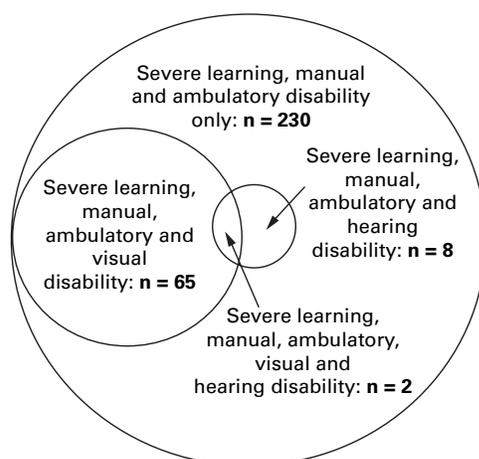


Figure 1 All birthweights: severe learning, manual, and ambulatory disability ($n=305$).

the 95% confidence interval of each annual estimate was wide and the annual prevalence rates did not differ significantly.

In contrast to the stability of the birthweight specific cerebral palsy prevalence over the 6 year study period, the birthweight specific neonatal mortalities all showed a significant decline, ranging from 14% in infants of birthweight <1000 g to 29% in those weighing ≥ 2500 g.

In 20 cases there was no record of the clinical type of cerebral palsy. These were because the infant died with a diagnosis of cerebral palsy recorded but with no information on the type. Quadriplegia comprised almost two fifths and hemiplegia almost one third of the remaining 1629 cases (table 2). The type of cerebral palsy differed significantly among the different birthweight groups (χ^2 , 9 df = 121.9; $p < 0.0001$). This was mainly attributable to relatively fewer cases of diplegia in the smallest and the largest birthweight groups.

Table 3 shows the severity of the motor, learning, and sensory disabilities. After excluding those of unknown severity, about a third of the children, irrespective of birthweight, are unable to walk independently. Of the heaviest infants, a significantly greater proportion have severe manual disability compared with those <2500 g (226/813 (27.8%) vs 163/779 (20.9); $p < 0.001$); they also have the highest proportion with severe learning difficulty. In the birthweight group ≥ 2500 g, 31.3% have a severe learning difficulty compared with 19.2% in those infants weighing <2500 g. This difference is highly significant ($p < 0.001$).

In contrast to the motor and learning disabilities, the sensory disabilities are more severe in those of lower birthweight. After excluding those children whose severity of hearing disability is not known, 10.2% of infants weighing <1000 g have severe hearing disability compared with 1.4% of the remainder ($p < 0.001$). For severe visual disability, the respective proportions are 16.1% and 9.5% ($p < 0.05$).

Of the 1649 cases of cerebral palsy, complete data on the level of disability in all five functional categories—learning, manual, am-

bulatory, hearing and visual disability—was available in 1338 (81%). Of these, 230 (17%) had severe learning, manual, and ambulatory disability, but their vision and hearing were not severely affected. An additional eight (0.6%) also had a severe hearing disability and 65 (5%) had co-existent severe visual disability. Two children (0.1%) were severely affected in all functional disability categories. These data are shown as a Venn diagram (fig 1).

Discussion

The stability of the birthweight specific cerebral palsy prevalence contrasts with that reported from other studies which have drawn attention to the increasing prevalence among those of the lower birthweight groups.⁵⁻⁸ This could be because the prevalence rates have reached a plateau or that the period covered by the study was too short to reveal a trend that is distinct over and above random variation. Alternatively, if there was incomplete ascertainment, it would affect the more recent rather than the earlier years of the study, particularly those children least severely affected in whom the diagnosis is most likely to be delayed. As several years have elapsed since the end of the reported study period, it is unlikely that failure of ascertainment would have had more than a marginal effect.

The birthweight specific prevalence of cerebral palsy shows a sharp rise with decreasing birthweight, with the highest prevalence in the lowest birthweight group (<1000 g). This is in contrast to previously reported series in which the prevalence of cerebral palsy in infants <1000 g was less than that in the 1000–1499 g group. In the past this would have been attributed to an artefact of the poor survival of the lowest birthweight group of infants who died before the diagnosis of cerebral palsy was made. But as survival improves, there is sufficient time for cerebral palsy to be diagnosed and counted so that the prevalence in those <1000 g now exceeds that in the 1000–1499 g group.

It has long been recognised, however, that heavier infants tend to have more severe cognitive and motor disabilities than those of low birthweight. Indeed, the higher prevalence of severe learning disability among infants of normal birthweight (≥ 2500 g) compared with those of <2500 g was considered to indicate a difference in the aetiology of cerebral palsy.¹³ An alternative explanation is the bias introduced as a result of differential survival. Among normal birthweight infants, even those with extensive cerebral damage may now survive; the neonatal mortality in this birthweight group was 1.2 per 1000 live births in 1992.¹⁴ In the lowest birthweight groups the neonatal mortality is much greater and infants with cerebral palsy would be more likely to die than unimpaired infants. The corollary to this is that, as survival in those of very low birthweight further improves, there will be a higher prevalence of cerebral palsy among the survivors.

In contrast to the higher proportion with severe cognitive and motor disability among

the heavier infants, the severest sensory disabilities of hearing and vision are more common in the lowest birthweight group. This cannot be due to bias attributable to differential survival and is, therefore, more likely to be directly associated with preterm delivery. Retinopathy of prematurity accounts for some, but not all, of the excess of severe visual disability.

About one in five children has severe multiple disability; the clinical and social impact is considerable because most require continuous care. On a more positive note, about half of these children are not intellectually impaired. Nevertheless, these children do need appropriate educational provision to help maximise their potential despite their physical disabilities.

Controlled trials have shown the effectiveness of surfactant¹⁵ and antenatal steroid use¹⁶ in reducing mortality and morbidity associated with hyaline membrane disease. The major impact of these interventions should be seen in cohorts born in the late 1980s and early 1990s, and their effects on cerebral palsy rates should continue to be monitored. Birthweight specific neonatal and postneonatal mortality is measured and published routinely in the United Kingdom but there is, as yet, no routine recording of disability, such that national trends in the prevalence of disability can be monitored. As long as this continues, cerebral palsy registers provide one source of information on long term disability in childhood. Not only should rates of disability be followed up but the prevalence of the various grades of severity of disability needs to be recorded if

monitoring is to be comprehensive and if there is to be rational planning for the needs of the disabled.

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