Prevalence and disabilities in 4 to 8 year olds with cerebral palsy

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SUMMARY A register of children born between 1970 and 1979 in the South East Thames Regional Health Authority, and diagnosed as having pre-, peri-, and postnatal cerebral palsy, was set up between 1978 and 1981. We report the 527 children born between 1970 and 1974 for whom ascertainment is virtually complete. The estimated prevalence was 2.2 per 1000, with 7.4% multiple births, and 58% boys. Birthweight distribution is as expected, with 35% weighing less than 2500 g at birth. Parental permission for release of detailed medical information was sought, and the clinicians responsible gave us data on the type of cerebral palsy; details of impairments, disabilities, and anticipated future prospects; and an opinion on the probable timing of the cause. There was a high incidence of orthopaedic defects which must represent a considerable use of resources, although the prevalence of hearing and vision defects suggested that some of these may be undetected.

The setting up of a geographically based register of children diagnosed as having cerebral palsy has been strongly recommended.^{1,2} The aims include the provision of epidemiological data for aetiological studies; the identification of trends in prevalence which may act as an outcome measure for changes in obstetrical or neonatal management; and the provision of an information base from which to plan services needed by the affected children.

Such a register was assembled between 1978 and 1981 in the South East Thames Regional Health Authority (SETRHA) with regard to resident children diagnosed as having any form of cerebral palsy and born between 1970 and 1979. We report here on children born between 1970 and 1974.

Setting up the register presented many problems, in particular the potential conflict between obtaining parental agreement to the registration and the need to obtain complete information, and problems arising from using data obtained as part of the routine care of the children, rather than carrying out a structured and validated examination of each child. The present report shows what could and could not be obtained from routinely available data.

Methods

Selection of cases. The register comprised children

with cerebral palsy who had been born in or whose families were resident in SETRHA at the time of ascertainment. The boundaries of SETRHA were as defined in 1974. It was hoped that the number of affected children born in the region, who had moved out before ascertainment, would be approximately balanced by the number of children born outside but resident at the time of ascertainment.

Ascertainment. Ascertainment was from multiple sources: in the first instance from the Spastics Society (18% of all those on our register); secondly from paediatricians in the region (51%); thirdly from area health authority handicap registers or special files (an additional 26%); and lastly from other sources including special schools and orthopaedic clinics (5%). A total of 816 cases born between 1970 and 1979 was assembled, mostly ascertained from more than one source.

This paper is based on the findings concerning the 527 children born in the period 1970 to 1974.

An independent check on the completeness of ascertainment is available for children born in 1970, 1971, or 1973 with a birthweight of 2000 g or less. These were included in a prospectively studied cohort,³ and using the sources listed above, retrospective ascertainment was complete except for one

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very mild case and two in whom the diagnosis was doubtful or had changed over the years.

Data obtained. The parents or guardians of the children were asked by the professionals who first notified us, whether we might approach them. Where an agreement was obtained permission was sought for the release of medical information, and for the place of birth and birthweight of the child. Of the 527 children in the subsample, parental forms were received for 316 (60%), of which 99% gave permission. No reply was received from 159 parents and a further 52 (10%) were not approached for a variety of social and other reasons. In a few cases the clinicians considered it appropriate to release information in the absence of explicit parental permission.

For 331 cases (63%), we then received a clinical form, signed by one of 50 or so collaborating clinicians, and this described each child's impairments, disabilities, anticipated future prospects and needs, and the probable timing of any insult thought to have caused the cerebral palsy. Obstetric details were obtained for 264 (50%) of the pregnancies. Where no clinical form was received and permission (parental or medical) was given, any available clinical data were abstracted from the local authority records.

Results

Types of cerebral palsy reported. The collaborating clinicians stated the type of cerebral palsy on the form according to their interpretation of the current nomenclature. More than 40 different diagnostic labels, and numerous combinations thereof, were used. This led to considerable difficulties when we attempted to allocate each case to a diagnostic subgroup. Only broad groupings could be used with any confidence, and these were distributed as in Table 1. Within the group described as spastic it was

Table 1 *Distribution of types of cerebral palsy*

Diagnosis	No	% of total number	% of those of known type
Dyskinetic	52	9.8	12.5
Spastic	270	51.2	64.7
Ataxic	39	7.4	9.4
Dyskinetic spastic	40	7.6	9.6
Dyskinetic ataxic	3	0.5	0.7
Ataxic spastic	13	2.5	3.1
Other types and type not known*	110	21.0	
Total	527	100	100

*The dyskinetic group includes choreoathetosis and dystonia; six cases were described only as 'hypotonic', but these are included under 'other types' as it was not clear how many had true hypotonic cerebral palsy.

possible to identify several subgroups, as shown in Table 2, although it was clear that different clinicians used these terms in different ways.

Age at diagnosis. The age at firm diagnosis was reported for 352 cases. Only 49% had been diagnosed by the first birthday, 79% by the second, and 95% by the fourth, but for many the presence of cerebral palsy had been suspected much earlier.

Prevalence, multiple births, and sex distribution. There were about 236 000 livebirths within the SETRHA in the years 1970 to 1974, so that the estimated prevalence was 2.2 per 1000.

For 354 cases, the clinicians gave an opinion on the probable timing of any insult thought to have caused the cerebral palsy. Thirty previously normal children (8.5%) were thought to have acquired the condition, most commonly after an infection affecting the central nervous system. These 'postnatal' cases are included in the analysis.

Of those where plurality was known (393), 7.4% were multiple births. More boys were affected than girls, and there is an excess of boys in all the major groups and subgroups, except for the diplegic group as shown in Table 3.

Birthweight and gestational age. Birthweight was known for 438 (83%) cases, the range being 790 to 4791 g, and estimated gestational age for 384, the range being 27 to 44 weeks. Eleven per cent weighed less than 1500 g, 6% between 1500 and 1749 g, 7% between 1750 and 1999 g, and 11% between 2000 and 2499 g; overall, 35% weighed less than 2500 g at birth.

Table 2 *Distribution of subgroups described as spastic*

	No	%
Hemiplegia	97	36
Diplegia	74	28
Quadriplegia	71	26
Monoplegia	6	2
Paraplegia	5	2
Other	17	6
Total	270	100

Table 3 *Percentage of boys by clinical type (524 cases)*

	%		%
Dyskinetic	62	Dyskinetic spastic	67
Ataxic	54	Dyskinetic ataxic	67
Spastic	56	Ataxic spastic	77
Hemiplegia	63		
Quadriplegia	63		
Diplegia	45		

Impairments. The forms completed by the clinicians or by our researchers from district handicap records gave details of impairments, which are summarised in Table 4. Two different percentages are given for each category. The first relates only to those cases for whom a clinical form was available, and may be an overestimate of the prevalence of impairment. This is because clinicians may have selectively chosen to complete clinical forms for severe cases, and some milder cases may no longer be under regular surveillance. The second percentage is estimated out of all cases assigned to that diagnostic group, whether or not clinical data were available. This figure may be an underestimate of the prevalence of impairment, because of the number of cases for whom information was incomplete. In the text, figures quoted are for those cases with clinical forms, unless otherwise stated.

Disabilities. Table 5 presents the findings related to mental ability, manual dexterity, and ambulation. As one would expect, 90% of all children in the study were reported as having a problem with ambulation, and more surprisingly 76% were described as having problems with manual dexterity.

Seventy seven per cent were said to have some mental disability. The children with spastic dyskinesia and with spastic quadriplegia were the most severely disabled, often with multiple, severe problems. (Data concerning the spastic subgroups is available on request).

Handicap: anticipated future prospects. The clinicians were asked to give their assessment of each child's likely future prospects using the categories listed in Table 6, and this was the best measure of handicap available from this study.

Clearly the children in this study were very young at the time the forms were completed, but the clinicians' views of their anticipated prospects was gloomy. A need for permanent care was foreseen for at least 23%.

Anticipated future prospects seemed, at least in the minds of the clinicians, to be very closely linked with mental ability. Of those who were of normal or near normal mental ability, only 4% were expected to require permanent care; of those expected to require permanent care, 96% were educationally subnormal. The anticipated future prospects of the spastic children are shown in Table 7.

Table 4 *Impairments by clinical type of cerebral palsy*

	<i>Ataxic</i>		<i>Spastic</i>		<i>Dyskinetic</i>		<i>Dyskinetic spastic</i>		<i>*All cases in study</i>	
	<i>Clin form</i>	<i>(All) (cases)</i>	<i>Clin form</i>	<i>(All) (cases)</i>	<i>Clin form</i>	<i>(All) (cases)</i>	<i>Clin form</i>	<i>(All) (cases)</i>	<i>Clin form</i>	<i>(All) (cases)</i>
Number of cases	32	(39)	191	(270)	34	(52)	28	(40)	332	(527)
<i>Hearing (%)</i>										
Severe defect	0	(0)	2	(2)	6	(4)	4	(2)	3	(2)
Moderate defect	3	(3)	6	(4)	6	(4)	4	(3)	6	(4)
Defect of unknown severity	0	(3)	1	(3)	0	(1)	0	(0)	1	(3)
No defect	97	(82)	88	(64)	82	(58)	92	(65)	87	(57)
Missing data	0	(12)	3	(27)	6	(33)	0	(30)	3	(34)
<i>Vision (%)</i>										
Severe defect	3	(3)	9	(8)	9	(6)	4	(2)	7	(6)
Moderate defect	6	(5)	17	(13)	15	(10)	18	(13)	16	(10)
Squint	19	(15)	17	(16)	17	(15)	32	(27)	19	(16)
Other defect, eg nystagmus	22	(21)	2	(3)	0	(0)	4	(3)	4	(4)
No defect	50	(46)	51	(36)	56	(38)	43	(30)	51	(33)
Missing data	0	(10)	4	(24)	3	(31)	0	(25)	3	(31)
<i>Orthopaedic (%)</i>										
Right hip defect	0	(0)	0	(0)	3	(4)	4	(2)	1	(1)
Left hip defect	0	(0)	2	(2)	6	(4)	4	(2)	2	(1)
Both hips	0	(0)	7	(5)	0	(2)	11	(8)	5	(3)
'Other' defect(s)	16	(13)	40	(32)	32	(23)	46	(33)	36	(26)
Hip(s) and 'other' defects	0	(0)	4	(3)	3	(2)	7	(5)	4	(3)
Defect(s) of unknown type	3	(5)	2	(4)	0	(2)	0	(10)	2	(4)
No defect	72	(59)	38	(27)	50	(34)	25	(18)	44	(28)
Missing data	9	(23)	7	(27)	6	(29)	3	(22)	6	(34)

*Includes cases for whom diagnostic type is not known, as well as cases in smaller diagnostic groups.

Table 5 *Disabilities by clinical type of cerebral palsy*

	<i>Ataxic</i>		<i>Spastic</i>		<i>Dyskinetic</i>		<i>Dyskinetic spastic</i>		<i>*All cases in study</i>	
	<i>Clin form</i>	<i>(All) (cases)</i>	<i>Clin form</i>	<i>(All) (cases)</i>	<i>Clin form</i>	<i>(All) (cases)</i>	<i>Clin form</i>	<i>(All) (cases)</i>	<i>Clin form</i>	<i>(All) (cases)</i>
Number of cases	32	(39)	191	(270)	34	(52)	28	(40)	332	(527)
<i>Mental ability (%)</i>										
ESN (S)	19	(20)	30	(24)	44	(40)	53	(45)	34	(26)
ESN (M)	41	(33)	20	(15)	26	(19)	25	(17)	24	(16)
Slight disability	31	(26)	20	(14)	12	(8)	18	(13)	19	(12)
Some disability										
severity not known	0	(0)	0	(4)	0	(0)	0	(0)	0	(4)
No disability	6	(8)	27	(20)	18	(14)	4	(5)	21	(15)
Missing data	3	(13)	3	(23)	0	(19)	0	(20)	2	(27)
<i>Manual dexterity (%)</i>										
Disability (S)	16	(13)	25	(18)	53	(36)	50	(35)	28	(18)
Disability (M)	69	(59)	45	(32)	41	(27)	50	(35)	47	(30)
Some disability,										
severity not known	0	(3)	1	(1)	0	(0)	0	(0)	1	(2)
No disability	12	(10)	26	(19)	0	(0)	0	(0)	21	(13)
Missing data	3	(15)	3	(30)	6	(37)	0	(30)	3	(37)
<i>Ambulation (%)</i>										
Bedridden or chair,										
not self propelled	0	(0)	20	(14)	50	(35)	39	(27)	22	(14)
Wheelchair,										
self-propelled	0	(0)	1	(1)	9	(6)	18	(13)	4	(2)
Sticks, crutches,										
rollator etc	13	(10)	11	(8)	9	(6)	18	(13)	12	(8)
Limited ambulation										
but no aids used	25	(20)	17	(13)	9	(6)	21	(15)	18	(12)
Minimal disability	50	(41)	41	(29)	14	(9)	4	(2)	33	(21)
Some disability,										
severity not known	0	(3)	1	(1)	0	(0)	0	(0)	1	(1)
No disability	9	(8)	5	(3)	3	(2)	0	(0)	7	(5)
Missing data	3	(18)	4	(31)	6	(36)	0	(30)	3	(37)

*Includes cases for whom diagnostic type is not known, as well as cases in smaller diagnostic groups.
ESN=educationally subnormal; S=severe; M=moderate.

Table 6 *Anticipated prospects in children with cerebral palsy*

	<i>No</i>	<i>% of total no</i>	<i>% with replies</i>
Normal	31	6	10
Normal school but some handicap	65	12	20
Special school but 'employable'	96	18	30
Permanent care	120	23	37
Impaired prospects (details not given)	10	2	3
Not known	205	39	—
Total	527	100	100

Table 7 *Anticipated prospects in spastic children by major subgroups*

	<i>Hemiplegics (%)</i>	<i>Diplegics (%)</i>	<i>Quadriplegics (%)</i>
Normal	20	16	2
Normal school but some handicap	45	24	4
Special school but 'employable'	22	46	13
Permanent care	10	10	80
Impaired prospects, (details not given)	3	4	1

Discussion

All features of the epidemiology of this group are closely in accord with that expected from the published reports. The prevalence of about 2 per 1000 livebirths, the pattern of birthweight and gestation, the excess of boys, the 7.4% multiple births, and the 8.5% of postnatal cases are within the range of other studies.⁴ It seems therefore that the present data do give some useful information for planning purposes; and it seems likely that these children are a fairly complete sample of cerebral palsy in the region in births between 1970 and 1974.

Orthopaedic impairments. The reported prevalence of hip defects, which was directly asked for, is likely to represent accurately the situation at the time of the child's most recent assessment. The form did not distinguish between congenital and acquired defects. The data concerning 'other' orthopaedic impairments must be regarded as incomplete, as details were often omitted from the clinical form. It has been very difficult to find any previous data with which to compare the prevalence of hip defects

reported in this study. Muckart⁵ reported on the orthopaedic aspects of a geographically based cerebral palsy study undertaken in the 1950s. In a group of 37 cases of spastic quadriplegia he surveyed, at least 16 (43%) had a hip problem, 12 flexed adducted hips, one a dislocated hip after arthritis, and three a flexion deformity due to muscle imbalance. Unfortunately it is difficult to make a direct comparison with the present study, but it seems that the overall prevalence of hip problems in spastic quadriplegia of a maximum of 28% (data available on request) may be a little lower in the 1970s compared with the 1950s.

Hearing impairment. The fact that only 10% of children with a clinical form were recorded as having any impairment of hearing may be seen as a cause for concern. The true incidence of hearing loss in normal children is difficult to determine. The National Child Development Study⁶ follow up of 7 year olds found that 5.7% had a hearing loss of 35 dB or more on at least two frequencies. More recently, however, a small study on a class of normal 8 year olds in a relatively deprived area of London⁷ found the incidence of a similar degree of hearing loss to be 26.3%. The incidence of hearing loss in children with cerebral palsy was discussed by Mowat⁸ who concluded that while studies using routine clinical records gave figures of 3 to 7%, the true incidence, shown by systematic investigation of hearing, was probably between 20 and 25% in the cerebral palsied population as a whole, and much higher among those attending special schools.

While it is possible that medical advances in the last two decades may have resulted in a genuine fall, it is likely that the prevalence of 10% in the present study is an underestimate, and this may mean that some children are not getting the help they need.

Visual impairment. In the present study half of those with clinical forms were thought to have no visual defect, but it is not known how many of these had been examined by an ophthalmologist. It is to be hoped that the figure of 7% for those with severe visual defects is not a gross underestimate. As the clinicians were not specifically asked about squints, the reported incidence of 19% is known to be an underestimate, and no doubt some of those with squints are grouped under 'moderate defect'. Douglas⁹ found that nearly half of the patients with cerebral palsy were affected by a squint. Black¹⁰ described the visual problems of children with cerebral palsy attending a Spastics Society school, and commented that a high proportion of the children had never been examined by an ophthalmologist. He also found that just over half of the

children at the spastics school had a visual defect, and was concerned at the large number of children with amblyopia who had not received any treatment.

As the most common visual defects in children with cerebral palsy are amenable to treatment, it is vital that this opportunity to enhance their communication with the world is not lost.

Disabilities of locomotion. Of the children for whom a clinical form had been received, 22% were bedridden or confined to a wheelchair which they did not propel themselves. A further 16% relied on a self propelled wheelchair or other aid for their mobility. Future follow up may, however, show that with the passage of time additional children have become unable to maintain independent mobility, or have resorted to aids in order to increase the distances they are able to cover as they seek greater independence.

Mental ability and future prospects. Anticipated future prospects are closely linked with mental ability, which is the overriding factor in all but the most severely physically handicapped cases, and 34% were reported to be severely educationally subnormal. This is slightly higher than the 27% of children born in the 1940s and 1950s, with an IQ of under 50,¹¹ but as explained above the figure for cases with clinical forms may be an overestimate.

Value of the study. These were results obtained from data available in the routinely kept medical notes, and no attempt was made to examine the children in a standard fashion. Any child with a clinical diagnosis of cerebral palsy was included, and in some the diagnosis might subsequently be revised. Moreover, we accepted the data limitations imposed by seeking parental permission for medical disclosure of records.

The quality of the data obtained needs to be looked at with regard to each of these constraints, for they had different effects. Firstly, it was obvious that the unstructured and idiosyncratic nature of the clinical records, kept by more than 50 different clinicians, prevented any rigorous systematic analysis of the different types of cerebral palsy and the impairments, disabilities, or handicaps. It would be a very considerable advance if some agreement could be reached, regarding an informative standardised and structured description of each, independent of individual classification preferences. Secondly, it is time for open discussion on the ethics of releasing data to bona fide researchers without explicit parental permission in such important conditions as cerebral palsy, a subgroup of which is still

regarded as an indicator of the quality of obstetric and paediatric care. Prospective studies with full parental agreement are the ideal, but are rarely feasible.

Finally, among recent arguments about the causes of, or trends in prevalence of cerebral palsy, the needs of the affected children seem to have been somewhat neglected. The present study, incomplete though it is, shows the number and complexity of the impairments and disabilities in these children, of whom some 100 new cases a year will occur in a health region with some 50 000 births, and in whom mortality has now been much reduced. It seems that the clinical picture of the group has not changed substantially since the 1940s and 1950s. Their orthopaedic problems alone must be a major drain on the paediatric surgical services, and there is still little agreement as to their best management. Sensory and communication problems still loom large, and again it is unclear to what extent these are being either fully diagnosed or compensated for through the use of new technology.

We owe it to these children to at least keep records that are standard and informative, and to follow them at regular intervals to see to what extent their needs are being met.

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