

Acquired cerebral palsy

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The heterogeneity associated with the diagnostic label of cerebral palsy has led to the formulation of several definitions of the syndrome. Early reports by Little, Osler, and Freud described cerebral palsy not as a disease entity but as a collection of motor disorders dating from the time of birth or early childhood and due to lesions of the brain.¹ The current burgeoning of interest in the epidemiology of the cerebral palsies has been largely instigated by its possible use as an outcome measure of the effectiveness of neonatal intensive care. This interest has drawn attention to some of the differences in, for example, the definitions given by the Little Club of London,² Bax,³ and Nelson and Ellenberg.⁴ It is common to these definitions that the lesion is of the brain, is non-progressive, and leads to a motor disorder resulting in aberrant posture and control. Some of the variation in definition is associated with the timing of the presumed insult, which may be in the early years of life,^{2, 4} or of the immature brain.³

In examining the epidemiology of cerebral palsy, the present tendency is to separate those in whom the presumed aetiology is postnatal—that is, 'acquired'—from those where the aetiology is pre-natal or perinatal in origin. This report is an account of the epidemiology of acquired cerebral palsy in the Mersey region.

Methods

A register was compiled of all cases of cerebral palsy born in the years 1966–77, whose mother's area of residence at the time of birth was within the boundary of the Mersey regional health authority, which has a total population of approximately 2.5 million. Several sources of data were used to compile the register, these included the handicap registers maintained in each district health authority, the national Rowntree Trust register of all families claiming a family disability allowance, records of all children attending schools for the educationally subnormal (moderate and severe) and the physically handicapped, and a search by the Office of Population Censuses and Surveys for all death certificates in which the underlying cause of

death on Part I, or as other significant condition contributing to the death on Part II, of the certificate was cerebral palsy.

The definition of cerebral palsy used was that proposed by Bax, namely a non-progressive group of disorders of movement and posture due to a defect or lesion of the immature brain. Because the definition only includes non-progressive disorders and lesions of the immature brain, all cases in which the underlying pathology was a tumour or a lesion of the spinal cord were excluded. The cerebral palsy was classified as acquired if the damage to the brain was considered to have taken place after the 28th postnatal day and up to the end of the 5th year.

The categorisation of the type of cerebral palsy was made on the basis of information in the clinical records of each child. If there was uncertainty about the timing of the insult, the aetiology was presumed congenital and not acquired.

The cases were grouped into spastic quadriplegia, spastic hemiplegia, and 'others'. Spastic quadriplegia included quadriplegia, double hemiplegia, and mixed quadriplegia and double hemiplegia. Spastic hemiplegia included the mixed hemiplegias. Others included the dyskinesic, ataxic, hypotonic, mixed spastic diplegic, and unclassified types of cerebral palsy.

The severity of functional disability as it affected ambulation was classified as minimal, moderate, or severe. Minimal disability meant that the child required no aid for walking though aid for climbing stairs may have been necessary. Moderate disability involved the use of an aid to enable the child to be mobile, either crutches or a self propelled wheelchair. Severe disability necessitated the use of a wheelchair that was not self propelled.

Almost all children were assessed by a clinical or educational psychologist at least once, usually when school placement was being considered. A variety of assessment procedures was used. Where possible mental disability was graded according to the intelligence quotient (IQ). Severe retardation was determined by an IQ <50, moderate retardation by an IQ 50–69, and normal or minimal retardation by an IQ ≥70.

Results

There were 833 cases of cerebral palsy recorded in the register of which 147 (18%) were classed as acquired—that is, the brain damage was considered to have taken place after the 28th day of life and up to the end of the 5th year. These 147 cases are the index cases for this report, 82 were boys and 65 were girls, a male:female ratio of 1.3:1. The cases relate to a denominator population of 443 531 survivors of the neonatal period giving an incidence of acquired cerebral palsy of 3.31/10 000. There was no discernible trend in incidence over time, the annual incidence rate by year of birth is shown in table 1.

TYPE OF CEREBRAL PALSY

Spastic quadriplegia with 76 cases (52%) was the commonest type of acquired cerebral palsy. There were 61 cases (41%) of spastic hemiplegia. 'Others' included 10 cases which were comprised of ataxia (n=3), dyskinesia (n=2), hypotonia, spastic diplegia, and mixed type (one case each), and there were two cases in whom the type of cerebral palsy was not recorded.

AETIOLOGY

The presumed cause of the cerebral palsy was grouped using a similar classification to that of Blair and Stanley and is shown in table 2.⁵ The commonest causal group was infection, which consisted predominantly of infections of the brain or meninges. A significant number of cases, however, were attributed to circulatory collapse consequent upon other infections such as septicaemia, respiratory tract infections, and shigella dysentery.

The age at which the damage to the brain occurred differed according to the causal group. As

Table 1 *Incidence of acquired cerebral palsy by year of birth*

<i>Year of birth</i>	<i>No of cases</i>	<i>No of neonatal survivors</i>	<i>Incidence/10 000 neonatal survivors</i>
1966	15	44 485	3.37
1967	22	42 982	5.12
1968	8	42 529	1.88
1969	11	41 242	2.67
1970	8	40 982	1.95
1971	17	40 170	4.23
1972	21	36 973	5.68
1973	12	33 552	3.58
1974	14	32 113	4.36
1975	5	30 885	1.62
1976	8	29 472	2.71
1977	6	28 146	2.13
Total	147	443 531	3.31

Table 2 *Acquired cerebral palsy grouped by cause*

<i>Apparent cause</i>	<i>No</i>	<i>(%)</i>
Head injury:		
Road traffic accident	13	(8.8)
Non-accidental	12	(8.2)
Other head injury	10	(6.8)
Anoxia:		
Suffocation	3*	(2.0)
Electrocution	1	(0.7)
Postoperative cardiac arrest	3	(2.0)
Postepileptic	13†	(8.8)
Cerebrovascular accident	5	(3.4)
Gastroenteritis and dehydration	22	(15.0)
Infections	63	(42.9)
Miscellaneous	2	(1.4)
Total	147	100%

*One case non-accidental; †one postoperative.

might be expected, head injury after a road traffic accident was more common in the older child, 12 out of 13 cases were incurred after the 2nd birthday. In contrast, non-accidental injury took place early in life with 11 out of 12 cases occurring within 18 months of birth. Acquired cerebral palsy attributable to infections was distributed throughout the first five years but was predominantly in the first year (34/63, 54%) and 27% in the second year. Gastroenteritis and dehydration as the causal factor showed an even greater preponderance in the first year when 20 out of 22 cases (91%) occurred. In five cases the onset of the cerebral palsy was noted to have followed the administration of pertussis vaccine. The time relationship was not stated, however, and cause and effect cannot be assumed.

There were 13 cases in whom the cerebral palsy was classified as postepileptic. Although in all but one it is unlikely that convulsions caused the brain damage. Thus in eight of these cases the presenting feature was convulsions in infancy, and the cerebral palsy was diagnosed subsequently. It is difficult to be certain of cause and effect in these cases as an underlying pathology may have been responsible for both the epilepsy and the cerebral palsy. In a further two cases convulsions were associated with hypernatraemia and severe dehydration. In one child hypoxic brain damage was diagnosed after admission to hospital aged 7 weeks with recurrent fits and severe hypothermia, in another the cerebral palsy was attributed to convulsions and cardiac arrest which occurred postoperatively and there was only one case in which cerebral palsy was attributed to prolonged generalised convulsions when aged 3.

MOTOR AND MENTAL DISABILITY

The association between motor and mental disability

Table 3 Correlation of mental and motor disability

Motor disability (ambulation)	No (%) with mental disability			Total
	Normal or minimal	Moderate	Severe	
Minimal	30 (86)	20 (77)	16 (20)	66 (46)
Moderate	5 (14)	5 (19)	7 (9)	17 (12)
Severe	0 (0)	1 (4)	58 (72)	59 (42)
Total	35 (100)	26 (100)	81 (100)	142 (100)*

*Data on mental or physical disability not available for five cases.

ity is shown in table 3. A higher proportion of children show severe mental retardation as classified by the criteria in this study. A child with acquired cerebral palsy whose mobility is severely affected is almost invariably severely mentally retarded also but the converse does not hold true. These differences are reflected in the type of school attended. Sixteen (11%) of the 147 children were attending a normal school, 33 (22%) a school for the physically handicapped and 67 (46%) a school for the educationally subnormal (ESN). Of the 67 most (56) were at an ESN (severe) school. A further 24 children were not at school because they had died before attaining school age or because they were too young to be attending school.

Tables 4 and 5 show the severity of mental and motor disability respectively associated with the type of cerebral palsy. As would be expected, the

Table 4 Mental disability according to type of cerebral palsy. Results are No (%)

Mental disability	Type of cerebral palsy		
	Spastic hemiplegia	Spastic quadriplegia	Others
Normal/minimal	27 (46)	6 (8)	2 (25)
Moderate	16 (27)	8 (11)	2 (25)
Severe	16 (27)	61 (81)	4 (50)
Total	59 (100)	75 (100)	8 (100)

Table 5 Motor disability according to type of cerebral palsy. Results are No (%)

Motor disability	Type of cerebral palsy		
	Spastic hemiplegia	Spastic quadriplegia	Others
Minimal	52 (88)	10 (13)	5 (63)
Moderate	3 (5)	13 (17)	1 (13)
Severe	4 (7)	52 (69)	2 (25)
Total	59 (100)	75 (100)	8 (100)

children with quadriplegia suffer a significantly greater degree of mental retardation and functional motor disability than the hemiplegias. Even among those with hemiplegia, moderate or severe mental retardation predominates, occurring in 32 of 59 cases, whereas functional motor disability is much less evident almost all being mobile without requiring aid.

BIRTHWEIGHT SPECIFIC INCIDENCE OF ACQUIRED CEREBRAL PALSY

Among the cases of acquired cerebral palsy, 11% were of low birth weight (<2500 g) compared with 6.1% of all neonatal survivors—that is, the risk of a child acquiring cerebral palsy is almost double in the low birthweight infant. In contrast, there is almost a seven fold increase in risk for congenital cerebral palsy in low birth weight compared with normal birthweight infants.⁶ Birthweight specific rates for acquired and congenital cerebral palsy are shown in table 6.

Discussion

Most definitions of cerebral palsy refer to the condition being the result of damage to the immature brain. Crucial to this definition is the age at which the brain is no longer considered immature. Blair and Stanley included those cases where the potentially brain damaging event had been recorded before the age of 5 years.⁵ On the other hand Cussen *et al* include cases up to the age of 2 years,⁷ while Swinyard *et al*, in a study from New York, take the upper limit as 15 years.⁸ These differences in definition will profoundly affect the relative preponderance of the casual factor, the older the age limit at which the brain damage occurs the greater the relative importance of road traffic accidents in particular.

There is similar variation in the age at which the insult occurs for differentiating between 'congenital' and 'acquired' cerebral palsy.⁵ Blair and Stanley included individuals born free of brain damage who

Table 6 *Birthweight specific rates of acquired and congenital cerebral palsy*

Birth weight (g)	Acquired cerebral palsy		Congenital cerebral palsy	
	No of cases	Prevalence/10 000 neonatal survivors	No of cases	Prevalence/10 000 neonatal survivors
≤1500	1	7.1	35	270.7
1501-2500	13	5.6	145	62.4
>2500	117	3.1	440	11.7

These rates exclude infants born in 1966 because of lack of birthweight specific denominator data for that year. The data for the congenital cerebral palsy have been previously published.⁶

suffered some subsequent potentially brain damaging event which was unrelated to a perinatal cause. Others draw the dividing line at 7 days,⁷ 14 days,⁸ or 28 days after birth.⁴⁻⁹ As the clinical diagnosis of cerebral palsy is not made in most cases until some months or even years after birth, it is often not possible to be certain when the brain damaging event took place. This uncertainty is greatest when the presumed insult is in the perinatal period. For example, in a premature and small for dates infant that has a stormy intrapartum and early neonatal history and is subsequently found to have cerebral palsy, it may be difficult to distinguish cause and effect. Modern investigative procedures such as ultrasound scanning may help to narrow the uncertainty. Even in the older child, when the developmental delay is only recognised after a serious infective episode, there may occasionally be uncertainty in determining cause and effect.

In the study reported here, the observation that 18% of cases of cerebral palsy are acquired is significantly higher than that from several other studies where the proportion ranges from 3-13%.¹⁰ Several factors may be responsible, apart from the differences in the definition of timing of an insult, there are variations in the methods by which cases are ascertained and in the population from which they arise. These differences in definition and ascertainment bedevil all epidemiological studies of cerebral palsy and render invalid almost all comparisons between studies. Standardisation of data recording is an essential step in solving these issues and a step in the right direction has been made.¹¹

It is pertinent to comment on the recorded association of pertussis vaccination and cerebral palsy. These cases were diagnosed at a time when there was intense interest and speculation on the neurological sequelae attributable to pertussis vaccination. The fact that five cases of cerebral palsy in the survey reported here were noted to follow vaccination must be interpreted with caution. Clear documentation of the time relationship between the two events was not available and a causal association is not a valid assumption.

The major tragedy of acquired cerebral palsy is that a considerable proportion of cases is potentially preventable. Furthermore, comparing the severity of mental retardation in congenital cerebral palsy reported in an earlier paper¹² with these data on acquired cerebral palsy, the latter show a significantly higher proportion of cases with severe or moderate mental retardation. This has important implications for health and social service provision and for the burden of care experienced by the family of an affected child.

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