ORIGINAL ARTICLE

Effects of cognitive, motor, and sensory disabilities on survival in cerebral palsy

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Background: Cerebral palsy presents with a range of severity of cognitive, motor, and sensory disabilities, which might affect survival.
Aims: To quantify the effects of motor, cognitive, and sensory disabilities, year of birth, birth weight, and gestational age on survival in cerebral palsy.
Methods: A cohort of children with cerebral palsy born between 1966 and 1989 to mothers resident in a defined geographical region was subdivided into early impairment (EICP: cerebral insult prenatally or within 28 days of birth) or late impairment (LICP: insult at least 28 days after birth). Deaths are notified by the National Health Service Central Register. Birth and disability details were obtained from clinical records. Survival analyses were carried out.
Results: Severe motor disability was associated with a 30 year survival of 42% and severe cognitive disability with a 30 year survival of 62%. Severe visual disability was associated with a 30 year survival of 38%, but the association of survival with hearing disability was weak. EICP had better survival than LICP but the difference was not significant after allowing for severity of functional disabilities. Normal birth weight infants (≥2500 g) showed no birth cohort effect, but the 10 year survival of low birth weight (<2500 g) infants declined from 97% for 1966 to 89% for 1989 births.
Conclusions: Survival in cerebral palsy varies according to the severity and number of functional disabilities and by birth weight. Among low birth weight children, survival declined steadily from 1966 to 1989 after allowing for disability. The disabilities reported do not capture all the factors affecting survival of preterm infants.

The effect of physical and mental disability on the survival of people with cerebral palsy has been described previously.[1–7] There is variation in the reported survival experience reported by these studies, and interest in the effect of hearing and visual disabilities.

It is now accepted that children with cerebral palsy may survive well into adulthood. Factors affecting their survival have impacted on the settlements in legal cases, and there is medicolegal interest in a more complete understanding of the factors associated with early death.

We present data on an updated and larger cohort from the Mersey Cerebral Palsy Register, with longer follow up than the previous report.[2] In addition, the effects of sensory disabilities are described.

SUBJECTS AND METHODS

Cerebral palsy register

The definition of cerebral palsy used is "a group of non-progressive, but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of its development".[8]

The cohort comprised all children with cerebral palsy born between 1966 and 1989, to mothers whose area of residence at the time of birth was in the counties of Merseyside and Cheshire. Multiple data sources were used for ascertainment of cases, as described previously.[9 10] The subjects on the register were flagged at the National Health Service Central Register (NHSCR) of the Office for National Statistics. Notifications of death are provided with copies of death registrations.

Children whose impairment was due to insult pre- or postnatally up to 28 days after birth were classified as early impairment cerebral palsy (EICP). Children who acquired the cerebral impairment as the result of an insult that occurred at least 28 days after birth and up to the age of 5 years were classified as late impairment cerebral palsy (LICP).

Classification of cognitive, motor, and sensory ability

The data used to classify the cognitive, motor, and sensory abilities were all obtained from the hospital and community child health clinical records. Motor ability was assessed in terms of ambulation and manual dexterity, and sensory ability in two modalities, hearing and vision.

Cognitive ability was based on the measurement of the intelligence quotient (IQ) and was classified into four categories, from normal to severe learning disability. A variety of test procedures were used to assess the IQ throughout the cohort, and the most recently determined value recorded in the clinical notes was used. A fifth category, "too young to test", occurred almost invariably with the most severe ambulatory and manual disability categories, and the child having died. This category was therefore combined with that for severe learning disability, as being the most likely level of cognitive disability.

Manual ability was classified into four categories, from normal to unable to feed and dress without assistance. Ambulatory ability was classified into five categories, from minimal disability to requiring a wheelchair with assistance for propulsion. Detailed definitions of these categories have been given previously.[2]

Hearing disability was classified into five categories: normal; moderate unilateral hearing loss; moderate bilateral hearing loss (40–69 dB); severe bilateral hearing loss (≤70 dB); and non-responsive, which might indicate severe hearing loss or severe cognitive disability with no apparent response.

Abbreviations: EICP, early impairment cerebral palsy; IQ, intelligence quotient; LICP, late impairment cerebral palsy

Table 1 Birth characteristics of 1942 subjects with cerebral palsy

	Early impairment			Late impairment		
	1668 subjects			274 subjects		
Characteristic	No.	%*	No. dead	No.	%*	No. dead
Sex						
Female	714	43	90	118	43	21
Male	954	57	143	156	57	38
Birth weight (g)						
Missing	47		12	16		3
⩽1500	242	15	26	3	1	1
1501–2500	407	25	43	25	10	5
>2500	972	60	151	230	89	50
Gestational age (weeks):						
Missing	111		22	36		6
<32	253	16	23	2	2	1
32–36	340	22	33	22	9	3
⩾37	964	62	155	214	89	49
Year of birth:						
1966–69	248	15	39	75	27	16
1970–73	262	16	53	69	25	20
1974–77	204	12	26	35	13	7
1978–81	272	16	32	39	14	5
1982–85	333	20	38	34	12	5
1986–89	348	21	45	22	8	6

*Percentages for those with data available.

Visual disability was classified into five categories: normal; 6/9 or worse in the better eye; 6/60 or worse in the better eye; possible visual disability; and unable to test.

For all disabilities, the disability level was recorded as missing in children who had died before they could be assessed. The information on the severity of disability is based on assessments of the child after at least 5 years of age.

Statistical analysis

Subjects' ages in days, either to death or until 31 August 2000 were calculated. This date allows for a three month lag in notification of those who had died, as provided by the NHSCR. The χ^2 test was applied when comparing the difference between two proportions. Ten, 20, and 30 year survival estimates were determined by the Kaplan–Meier estimator, and comparisons between groups assessed using the Wilcoxon significance test.[11] Accelerated life models were used to model the combined effects of covariates on survival[11]; Akaike's information criterion[12] was used to select the final model. Likelihood ratio tests were used to combine categories. Missing data were investigated initially as separate categories, and then combined as appropriate.

RESULTS

There were 1942 children included in the analysis, of whom 1668 (86%) had EICP and 274 (14%) had LICP. The age at which the initial diagnosis of cerebral palsy was made was missing for 182 people. In the majority of children with both EICP (95%) and LICP (86%), the initial diagnosis was made before the age of 5 years. Of those with EICP, over two fifths of children were initially diagnosed before age 1 (43%), almost a third (31%) in their third year, and (13%) during their second year of life.These percentages are slightly lower for those with LICP: 40% before age 1, 23% aged 1, and 12% aged 2, as would be expected for later onset of cerebral palsy. The diagnosis of cerebral palsy may be prone to error if made during the first year of life.[13] Children were included in the register only if examination at later ages confirmed the initial diagnosis.

Summary statistics

Tables 1 and 2 give the characteristics of the 1942 children. By 31 August 2000, 233 (14.0%) with EICP and 59 (21.5%) with LICP had died ($p = 0.001$). The male to female ratio was 1.3:1 in both groups. In other respects the two groups differed. Most children with LICP were of normal birth weight (>2500 g; 89%) and gestational age (⩾37 weeks; 89%); whereas 60% of those with EICP were of normal birth weight and 38% were preterm (<37 weeks).

Compared with EICP, those with LICP had more severe cognitive, motor, and sensory disabilities. Over half of the LICP children had severe cognitive disability, in comparison to one third of those with EICP. Deaths were most frequent in those with the most severe disabilities.

Associations between birth weight, and motor, cognitive, and sensory disabilities

Among those with EICP, the proportion of children with severe disabilities was highest for those of normal birth weight compared with low birth weight cases (table 3). In contrast, fewer normal birth weight children had severe visual disability. Hearing disability was unrelated to birth weight group. For those with LICP, there was no association between severity of disability and birth weight group (table 3).

In children with EICP, one-third of those who were in the severe categories of cognitive and motor disability lacked normal hearing, in contrast to 6% of those with less severe cognitive or motor disability (table 4). A stronger relation was seen for LICP; one half of those with three severe disabilities were also hearing disabled (table 4). The proportion with visual disability increased as the number of severe cognitive or motor disabilities increased for both EICP and LICP.

Univariate survival estimates

Females with EICP or LICP had slightly better survival than males (table 5). For both birth weight and gestational age in conventional categories, survival was poorer for normal birth weight and full term infants, though not significantly so for those with LICP. For EICP, modelling the survival from birth with actual birth weight in grams, the survival was better for

Table 2 Disabilities of 1942 subjects with cerebral palsy

	EICP			LICP		
	1668 subjects			274 subjects		
Characteristic	No.	%*	No. dead	No.	%*	No. dead
Ambulation						
Missing	27		10	4		0
Minimal disability	809	49	12	121	45	2
Limited mobility	135	8	4	22	8	5
Walking aids	229	14	8	24	9	2
Self propelled wheelchair	86	5	7	10	4	1
Other wheelchair	382	24	192	93	34	49
Manual dexterity						
Missing	35		11	5		0
Normal	142	9	3	5	2	0
Mild disability	860	53	21	123	46	4
Moderate disability	259	16	12	54	20	5
Severe disability	372	23	186	87	32	50
Mental ability						
Missing	62		39	13		6
Normal	709	44	14	56	21	0
Mild disability	148	9	1	29	11	0
Moderate disability	223	14	8	42	16	3
Severe disability	526	33	171	134	51	50
Hearing						
Missing	41		27	7		1
Normal	1517	93	179	237	89	44
Unilateral	21	1	2	9	3	2
Bilateral 40–69 dB loss	40	2	9	4	2	1
Bilateral ⩾70 dB loss	35	2	4	7	3	2
Unresponsive	14	1	12	10	4	9
Vision						
Missing	76		19	9		1
Better than 6/60 in better eye	1279	80	60	165	62	11
6/60 or worse in better eye	121	8	62	44	17	22
Possible disability or unable to test	192	12	91	56	21	25

*Percentages for those with data available.

Table 3 Association between severe disabilities and birth weight in people with cerebral palsy

	With severe disability			Without normal	
Birth weight (g)	Ambulation %	Manual %	Mental %	hearing %	vision %
EICP					
<1500	21	18	26	8	53
1500–2499	21	20	30	13	51
⩾2500	25	25	36	11	44
Total number	371	361	514	173	728
LICP					
<2500	33	33	60	22	63
⩾2500	34	31	50	22	52
Total number	87	80	112	56	133

2000 g infants (30 year survival rate 86%), than for 1000 g (84%), 3000 g (81%), or 4000 g (80%) infants. The best survival was for 5000 g infants (93%); but this pattern was mainly due to the absence of death before 2 years in heavy infants. For those children who survived at least two years, survival to age 30 was 89% at 1000 g, and declined to 84% at 3000 g and 78% at 5000 g. The pattern of survival from birth with gestational age in weeks was similar, with those born at 32 weeks having the best survival: 10 year survival rate 94%, in contrast to those born at 25 weeks and 40 weeks, who had 10 year survival rates of 88% and 90% respectively.

There was no obvious improvement in survival of the four year cohorts (table 5), but when year of birth was entered as a continuous variable, with an interaction with low birth weight (⩽2500 g), for EICP, there was an association. Children of normal birth weight had the same 20 year survival rate (85%) throughout the period. For low birth weight children, the 20 year survival rate was better than that of normal birth weight children in the 1960s (95% in 1966), but has declined steadily to become worse by the late 1980s (83% in 1989). The survival rate to age 20 years for those surviving to age 2 was 91% for normal birth weight, and declined from 97% to 91% for low birth weight infants (fig 1).

Survival rates decrease as the severity of cognitive, motor, and sensory disabilities increased; the pattern held for both

Table 4 Association between number of severe motor and cognitive disabilities, and sensory disabilities in people with cerebral palsy

No. of severe cognitive or motor disabilities	EICP without normal		LICP without normal	
	hearing %	vision %	hearing %	vision %
None	6	33	7	26
One	6	50	15	65
Two	6	58	12	75
Three	34	86	48	88
Total number	166	702	54	140

Table 5 Survival by birth characteristics of 1942 people with cerebral palsy

	EICP				LICP			
	% surviving to				% surviving to			
Characteristic	10 y	20 y	30 y	p*	10 y	20 y	30 y	p*
Sex				0.29				0.39
Female	91.9	87.8	84.8		91.4	82.7	80.2	
Male	91.2	85.9	82.1		91.0	81.4	69.6	
Birth weight (g)				0.08				0.69
⩽1500	90.5	89.6	89.6					
1501–2500	93.9	90.1	85.9		85.7	77.5	77.5	
>2500	91.4	85.5	81.7		92.1	82.5	72.9	
Gestational age				0.02				0.60
<32 (weeks)	91.3	90.9	90.9					
32–36 (weeks)	95.0	90.0	88.0		91.7	82.2	82.2	
⩾37 (weeks)	90.6	84.9	80.9		91.1	81.7	70.9	
Year of birth				0.15				0.14
1966–69	93.6	87.9	86.3		97.3	85.0	78.1	
1970–73	89.7	85.1	79.2		92.8	81.2		
1974–77	92.7	88.2			85.7	80.0		
1978–81	93.4	88.6			89.7	87.2		
1982–85	91.0				94.1			
1986–89	89.7				72.7			

*Wilcoxon test.

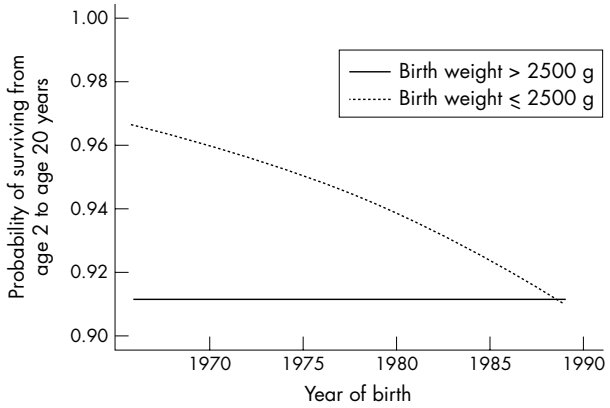

Figure 1 Survival rates to age 20 from age 2 by year of birth.

EICP and LICP (table 6). This effect was strongest for motor and sensory disabilities.

Severe disability in one area is usually associated with disability in other areas. It is therefore essential to consider all these factors in combination.

Combined effects of covariates on survival

Accelerated life log logistic models, in which the risk of dying, having survived to a given age (that is, the instantaneous death rate or hazard rate, increases after birth and then decreases again), provide a good model or summary of these data. For these models, the median survival times are adjusted by factors for each covariate.

Early impairment cerebral palsy

Severe ambulatory, manual, and cognitive disability were all strongly jointly associated with decreased survival. When vision was added to the cognitive and motor disabilities, those with normal sight had the best survival; the next level had poorer survival. The survival of those with 6/60 or worse vision was not distinguishable from that of those with possible disability, or who were impossible to test or had missing data. As the difference between those with normal vision and those with minor visual impairment was marginal, the contrast between these two groups and the remainder was used in the model. For hearing, those with normal hearing or moderate impairment could not be distinguished from those with severe impairment. Those who were unresponsive or for whom hearing data were missing had substantially poorer survival. As these hearing categories for the 55 people concerned essentially indicate that the child was severely brain damaged and died early, the factor contrasting this group with other levels of hearing did not imply a significant association of survival and severity of hearing. Rather, it indicated that the lack of information on hearing was probably due to the death of the child before it could be fully assessed. The subset of data excluding these 55 children was also analysed, and the conclusions were essentially the same.

Table 6 Survival by functional disabilities of 1942 people with cerebral palsy

Characteristic	EICP* % surviving to			LICP* % surviving to		
	10 y	20 y	30 y	10 y	20 y	30 y
Ambulation						
Minimal disability or limited mobility	99.5	98.6	97.5	98.6	96.2	93.6
Walking aids	98.3	96.6	96.6	97.1	90.5	90.5
Self propelled wheelchair	96.5	94.2	88.7			
Other wheelchair	68.6	51.3	42.0	77.3	55.9	37.9
Manual dexterity						
Normal, or mild disability	98.9	97.7	96.9	99.2	96.6	96.6
Moderate disability	98.1	95.9	93.8	98.2	94.3	88.4
Severe disability	68.8	51.5	41.1	74.4	52.0	32.5
Mental ability						
Normal, or mild disability	99.1	98.5	97.7	100.0	100.0	100.0
Moderate disability	99.1	96.7	94.8	100.0	92.1	92.1
Severe disability	82.3	69.4	61.9	87.2	70.5	54.0
Hearing						
Normal or unilateral loss	93.6	88.6	85.0	92.8	84.2	77.2
Bilateral loss	88.0	81.0	81.0	100.0	68.4	—
Unresponsive or missing	38.2	34.1	30.7	63.6	56.6	32.3
Vision						
Better eye: <6/60	98.2	95.8	93.5	98.2	94.0	91.7
Better eye: worse than 6/60	64.5	46.4	37.7	80.0	50.4	39.8
Possible, unable to test	69.3	55.6	47.3	80.0	69.1	49.0

*Wilcoxon test: $p \leq 0.001$ for all disabilities.

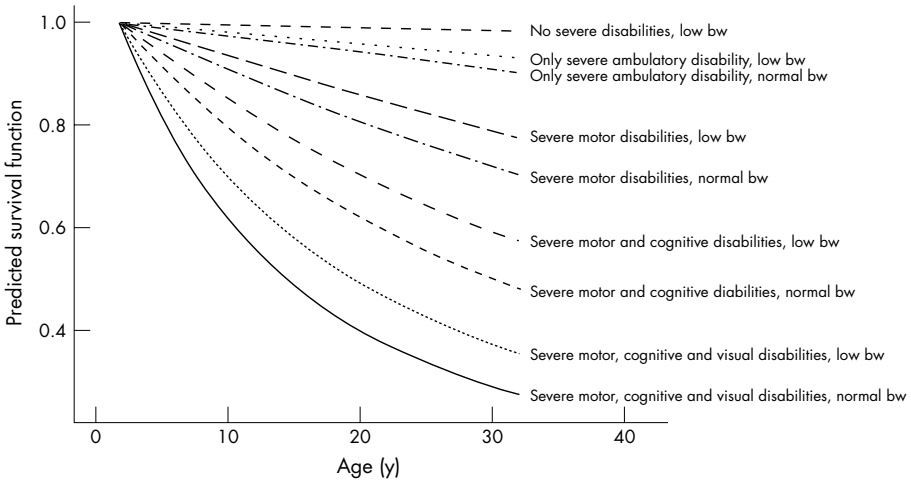

Figure 2 Predicted survival from age 2 years. Normal bw, birth weight >2500 g; low bw, birth weight ≤2500 g.

Visual disability had explanatory value in addition to the motor and cognitive disabilities. Those with normal or reasonable sight had better survival than those without. After allowing for motor, cognitive, and visual disabilities, and whether a child had birth weight ≤2500 g, the year of birth was additionally associated with survival. Low birth weight children had slightly better survival, as did children born earlier in this cohort. For example, the age to which 95% of children will survive was reduced by the factor 0.84 for 1980 births compared to 1970 births. Gestational age and birth weight were not significantly associated with survival after allowing for motor, cognitive, and visual disabilities.

As there might be some missing cases among children who die early, and were therefore not included in the register, further analyses of the survival from age 2 years of the 1890 children who lived at least two years were carried out. The conclusions were very similar. The estimated rate of decline with year of birth was essentially unchanged, though no longer significant.

Figure 2 presents survival curves for various combinations of severe disabilities and birth weight group. The substantial deterioration in survival with increasing numbers of severe disabilities is clearly shown.

Late impairment cerebral palsy

As there were only 274 people with LICP, there was limited power to investigate multivariate models. Considered individually, severe manual disability was most strongly associated with survival, followed by severe ambulatory, cognitive, visual, and hearing disability. Once severe manual disability was taken into account, ambulatory and visual disability had no additional association with survival, but severe mental and hearing disability were associated with poorer survival.

All cerebral palsy

When models for the entire data set were investigated, a variable indicating whether the person has EICP or LICP had no explanatory power once their disabilities were taken into account. Analysis from age 2 years gave essentially similar results.

DISCUSSION

The effect of sensory disabilities on survival has not been reported previously. Severe visual disability, and lack of information on hearing, were associated with reduced survival. The effect of severe visual disability on survival quantiles was the same in those with no severe motor or cognitive disabilities as in those with severe motor and cognitive disabilities.

The univariate patterns of survival for cognitive and motor disabilities were similar to results reported by us previously, and for that reported in other studies.[1–7] Nevertheless, when interpreting the effects on survival of all the variables examined in this study, ascertainment bias must be taken into consideration. Compilation of the Mersey Cerebral Palsy Register began in 1980 but retrospectively collected cases born from 1966 onwards. It is possible that some cases of cerebral palsy, born in the earlier years of this study cohort, may have died and been removed from the sources of ascertainment before 1980. This would particularly bias the results in that it is likely the most severely disabled children would have been excluded.

A further source of ascertainment bias arises in association with the increase in prevalence of cerebral palsy observed among low birth weight infants.[9 10] This has been attributed to improvements in neonatal care that have led to a significant increase in the survival of preterm infants. These preterm infants now survive to an age that allows the diagnosis of cerebral palsy of prenatal origin to be made. A corollary to this increased ascertainment among preterm infants is that there was a trend towards an increased severity of disability among them.[14]

Motor, cognitive, and visual disability modalities contribute in a multivariate model, and low birth weight and year of birth were additionally significantly associated with survival. Survival was better for low birth weight than normal birth weight infants, but declined steadily for both groups from 1966 to 1989, after allowing for the effect of four disability modalities. This suggests that the disabilities reported here do not capture all the factors affecting mortality and morbidity of premature infants, far more of whom now survive long enough for cerebral palsy to be diagnosed.

A comparison of the survival of this cohort with others has been reported.[7] It may not be valid to compare of the results we present with those using the California service register.[4 5] The Californian Register included people with cerebral palsy or other motor dysfunction and provides no information on the censoring mechanisms, nor on missing data. It is difficult, therefore, to determine how different the Californian Register population is from the Mersey cohort.

This report has medicolegal implications. Medically qualified experts may not be conversant with the statistical complexities of determining life expectancy,[15] and it is not valid to use the results given here to determine more than an approximate estimate of the life expectancy for an individual child with cerebral palsy. The life expectancy will be affected by the age to which the child has already survived.

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COMMENTARY

As Hutton and Pharoah have pointed out, there are a variety of reasons why there is increasing interest in studying the survival of individuals who have cerebral palsy.

The appropriate context for this must surely be the increasing recognition that the implications of making a diagnosis of cerebral palsy in a child are life long, both for the individual and the family. It follows that as much as possible needs to be learned about aetiology, natural history, intervention modalities and their relevance, and survival.

In this paper, Hutton and Pharoah have extended the work presented in their earlier Liverpool study by now looking at the survival of children born up to 30 years previously. They have shown both for children with early impairment (damage occurring no more than 28 days after birth) and later impairment cerebral palsy, that the large majority survived the study period.

In elucidating those who are rather more or less likely to survive, a number of characteristics have been examined, some of which are novel when contrasted with both their previous studies and those of other workers. Fundamentally, these relate to sensory—that is, visual and hearing impairments. These have been studied by analysis of the relevant medical records, and Hutton and Pharoah suggest in consequence, firstly that a significant minority of children with visual problems but only a few with hearing problems are demonstrable in their population, and secondly, that severe visual impairment is a marker for increased likelihood of early demise.

While these data are of interest, it is unfortunate that more clinical information has not been provided. For example, the authors infer that severe visual impairment is a marker for more severe disability generally, and yet we are not told even whether the clinical findings are those primarily of ophthalmic pathology, for example retinopathy of prematurity, or of cerebral visual impairment. Indeed, no information is given as to how the visual acuity measurements were obtained.

While it is possible—at least in theory—to understand that there might be a correlation between severe visual impairment of brain origin and severity of disability, it is much more difficult to understand, either on an aetiological or pathological

basis, the correlation between severe hearing impairment (defined as a bilateral loss of more than 70 dB) and brain abnormalities leading to cerebral palsy. The authors have nevertheless found that no fewer than 2% of their early impairment group (35 subjects) and 3% of their late impairment group (seven subjects) have a severe hearing loss.

It may be that this hearing loss has occurred coincidentally to the cerebral palsy, and the fact that there is no correlation between severe hearing loss and increased mortality in their study might support that. What a pity it is that the authors have not emphasised the need for further evaluation of the individuals with sensory impairment; this point is returned to further below.

Hutton and Pharoah also make clear that their study of the clinical records confirms that, so far as cognitive impairments are concerned, the greatest mortality has occurred in those who are severely disabled—that is, have a recorded IQ below 50. Here again, an opportunity may well have been missed. Clinical experience strongly suggests that among this grouping, it is those individuals who have profound cognitive impairment (which can be taken as equivalent to an IQ of 20 or less) have the shortest survival, especially when this is combined with severe motor and other impairments. This would also be consistent with the work of Blair and colleagues.[1] It is unfortunate therefore that no comment has been made on whether, from the data available, the authors could distinguish between subjects with profound cognitive impairment and those who were less severely affected but still had an IQ below 50.

What lessons can be learned from this study? The first is the reaffirmation that individuals with cerebral palsy who have severe and multiple disabilities are nevertheless likely, on a statistically derived basis, to survive into adult life and that we must plan services accordingly.

Secondly, it is clear that there is a reasonable consistency between Australian, North American, and British studies on survival in cerebral palsy when methodological variations are accepted.

Thirdly, for clinicians, epidemiologists, and statisticians, the newly provided data are helpful when being asked to estimate the probability of survival for individual children within a medicolegal context.

However, perhaps the most important conclusion is that unless opportunities are taken by statisticians, epidemiologists, and clinicians to work more coherently and productively together, opportunities for maximising the use of data will be lost. This is a message not so much for individual workers as for funding bodies. Databases such as that from Liverpool are very important repositories of information. They need to be used to provide and test hypotheses at a time when subjects are available for study. Merely to look at the data from this or other databases retrospectively will not promote the optimal provision of services for disabled children.

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