Letters to the editor

'Comparison of survival in cerebral palsy between countries.'

SIR–The authors of the paper 'Life expectancy among people with cerebral palsy in Western Australia'[1] (see pages 508–15, this issue), kindly allowed us use of their database in order to compare with the large Californian database.[2,3]

We begin our comparison at age 5 years because measurements of intellectual level and severity of cerebral palsy (CP) in Western Australia are updated only until that age. Overall survival rate in the Western Australian population is 93% compared with 83% in California. This large disparity is not, however, due to differences in care in the two regions. It arises because the Californian data include nearly all the more severely affected individuals but comparatively fewer mild instances whereas the Western Australian database includes everyone with CP (most of whom are only mildly affected).

For a valid comparison, we stratified the population into four intellectual levels (IQ<20; 20–34; 25–49; 50–69) and three severities of motor dysfunction (mild, moderate, and severe), creating 12 groups.

When just one factor (severity or IQ) was controlled, the gap in survival rates narrowed considerably. When both were controlled, the survival curves became strikingly similar. Figure 1 illustrates two cases: severe impairment with IQ<20 and with IQ in the range 20–34.

Data from the UK study by Hutton and coworkers[4] of the 'three severe disabilities' group was compared to the Californian database in the same way and found to be remarkably similar.

To our knowledge, this is the first controlled comparison of survival in CP between countries.
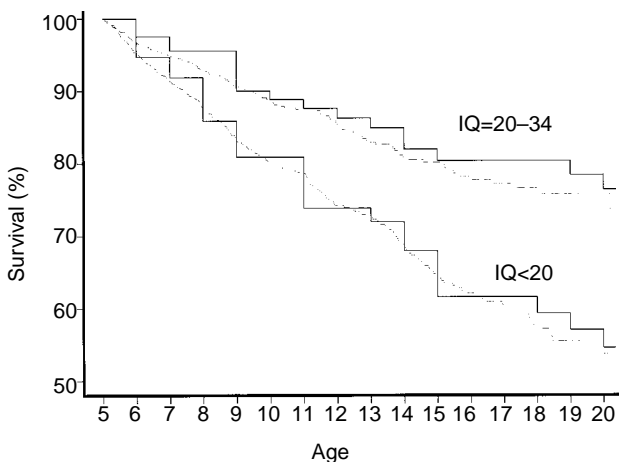

Figure 1: *Comparison of Western Australia (WA) and US survival curves for people with severe CP. All 12 pairs of curves either show similarity or have too few deaths to make comparisons meaningful. In no comparison was there a statistically significant difference; smallest p value=0.23.* ——, *data from WA (IQ 20–34, n=126; IQ<20, n=76);* -----, *data from US (IQ 20–34, n=828; IQ<20 n=974).*

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'Disability information improves reliability of cerebral palsy classification'

SIR – We were pleased to read the carefully detailed process by which the Surveillance of Cerebral Palsy in Europe (SCPE) collaborative network is standardizing the definition of CP across eight European countries.[1] Cerebral palsy (CP) suffers from a lack of precision in diagnosis and reporting due to the term's inconsistent use for a range of mild to severe motor abnormalities in children who are thought to have brain injuries. The authors identify four processes that result in differential detection of CP in the network's 14 geographically-based registries: definition and exclusion–inclusion criteria, case ascertainment, interobserver error, and method of classification and recording. Among these, the underlying impediment to accurate comparisons of CP prevalence over time and place is lack of a standard definition.

We have found that functional criteria are very important for the description of CP in a standardized form. But criteria for disability are not included in the common rules imposed by the network for a designation of CP. Of the five key elements specified, none require information as to age-appropriate activity limitations. By age 3 years, the lowest age of registration, major motor development milestones can be assessed and evidence for disability in walking, running, climbing stairs, jumping, dressing, feeding, and speech articulation should be perceptible and meaningful. The hierarchical classification scheme for subtypes of CP also does not use criteria for disability, although the authors acknowledge that disability information could help exclude ambiguous mild cases when estimating prevalence, and state encouragingly that work on functional loss is still ongoing.

Our research group has found that without evidence of disabling conditions, CP had poor reliability of classification across international cohorts. We analyzed neurological findings from three population-based cohorts of very-preterm babies born in Canada, the US, and The Netherlands in the late 1970s through to the mid 1980s. Up to five pediatricians with expertise in diagnosing CP reviewed 33–51 case records (omitting the CP diagnosis) from weighted random samples of children in each cohort

with disabling CP (DCP), non-disabling CP (NDCP), and no neurological abnormality. DCP was much more reliably classified than NDCP, yielding kappa measures of interobserver agreement in a range from fair to excellent. The one cohort producing only fair agreement for classifying DCP versus no DCP or any CP increased its kappa into the range of excellent when functional motor assessments were made available to reviewers along with neurological information. [2,3]

To promote reliable assessment of this important public health burden, we recommend that disabling characteristics be added to CP definitions, and that disabling CP always be included as a distinct outcome of interest among those investigating CP trends in differing time periods and locations.

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'Surveillance of Cerebral Palsy in Europe (SCPE) reply'

SIR–Firstly we are very encouraged by the interest of this research group from Michigan in the European collaborative work on cerebral palsy (CP).

We accept the general principle in the letter by Lenski and colleagues – that level of functional loss associated with CP is a very important outcome, indeed the most important outcome as far as parents are concerned. In addition we recognize that it is likely that disagreement over inclusion and exclusion of children with 'non-disabling' CP could contribute to some of the variation in prevalence rates between centres. With this in mind, we included in the 5 key elements of our definition: 'a disorder of movement and/or posture and motor function' (p 819). A child presenting with neurological signs without any disorder of motor function, is not included as 'a CP case' on the European database. Although not fully described in our paper, we also included in the standard minimum data set used to describe children on the European database, information on level of walking at the age of 4 or 5 years. This allows us the possibility of defining three groups of children, those who are walking unaided, those walking with aids, and those who are unable to walk even with aids. Prevalence rates excluding the mildest group can then be reported.

We recognize that this is a rather simple approach to including 'disability' when reporting prevalence rates. We do not have information on children at several ages. Most of the children are registered only after the age of 4 to 5 years and the question is, whether the degree of motor disability changes very much after this age. To our experience, it seems it does not: if you have not learnt to walk independently at the age of 5, there is a low probability that you will learn it afterwards in a way which is functionally relevant. We plan, however, to continue to discuss ways of recording a reliable, age-related composite measure of disability. Using the Gross Motor Function Classification is one way of doing this, although this would be difficult to achieve at international level. We are not clear how 'disability' at 2 years of age was defined by the Michigan group as their work is reported only in abstract form, and we would be very pleased to know more about it.

We share the correspondents' concern that in order to compare prevalence rates of CP over time and between areas, we need a consensus on definition and inclusion and exclusion criteria and we need to use a scale measuring the level of functional loss. We look forward to collaborating with them and other groups around the world who share this interest.

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'School career of children is at risk before diagnosis of epilepsy only'

SIR–In childhood, 'epilepsy only'[1] is less benign than the clinical impression holds. Although heterogeneous in aetiology and seizure characteristics, it is clinically a clearly discernible category, comprising approximately two-thirds of the total population of childhood epilepsy. Before the onset of epilepsy, the development of these children is generally normal. There are, by definition, no associated diseases and the seizures react favourably to antiepileptic drug (AED) treatment in 70% of patients.[2] However, Austin and colleagues in Minnesota, USA, found 44% of children with 'epilepsy only' had repeated at least one year at school, which is twice as many as a control group of children with asthma.[3] All children had been on AEDs for at least one year before inclusion. Bailet and Turk, in Florida, USA, found 34% of 74 children with recent onset and established idiopathic epilepsy to have repeated a year of school, compared with 13% of 23 older siblings.[4] Special educational services have been more frequently offered to children with idiopathic epilepsy (19%) than to control siblings (4%).[4] European school data are scarce.

Between January 1997 and October 1998, we assembled a group of 69 children with recently diagnosed 'epilepsy only' from 10 hospitals.* Inclusion was consecutive. All children had full investigations with EEGs and MRI if appropriate. 'If appropriate' refers to the fact that, in agreement with good practice, children with primary generalized and those with Rolandic epilepsy had no MRIs. Children with temporal lobe epilepsy who on MRI had mesial temporal sclerosis

were excluded. Healthy classmates, matched to the children with epilepsy for age and gender, participated as controls. All attended normal schools. Both groups were followed for a period of one year. Sample attrition was negligible: one control child dropped out from the second investigation and two more from the third one. AEDs were prescribed to fifty children with epilepsy. At the end of the year, 38 children had a remission of at least six months. We report here on school careers: repeating years, remedial teaching, and academic progress. The Teacher's Report Form[5] and a questionnaire addressing teachers' assessment of any change, yielded data on school performance at intake (within 48 hours after diagnosis and before AED-use), and at three and 12 months later.

To our surprise, before the diagnosis, 15 children (22%) with epilepsy had repeated a year at school, compared with an average, in The Netherlands, of 11% in mainstream primary schools.[6] Because of age requirements, repeaters with epilepsy tended to be matched with classmates who had also repeated a year (McNemar p=0.078). Over the one-year follow-up, six more children with epilepsy, including one boy for the second time, and only two controls had to repeat a year. In The Netherlands, almost 9% of children are allowed to continue at kindergarten, usually for reasons of maturation.[6] Among the children with epilepsy, the number of repeaters (10/21) in higher years is particularly striking, as this is counter to Dutch educational policies.[6]

We also found that, already before the diagnosis, remedial teaching had been requested for more children with the later diagnosis of 'epilepsy only' (54%) than for their healthy classmates (23%; χ^2, p=0.003). Over the year, the necessity for remedial teaching did not change significantly. None of the children, however, had to be sent to schools for special education.

At the time of diagnosis, significantly more children with epilepsy than their non-epileptic classmates had lost ground (Table I). Twelve months after diagnosis, only a few children with epilepsy had overcome academic stagnation.

The cause of poor school performance before diagnosis requires further study. Epilepsy and control groups did not differ significantly in general intelligence (epilepsy mean IQ 98, SD 17.3; classmates mean IQ 100, SD 16.1) or educational level of the parents. The majority of the children in both groups were of Dutch origin and children with epilepsy who belonged to ethnic minorities were matched with children from their ethnic group. Repeating a year was not statistically significantly associated with particular epilepsy subgroups. More children who were treated with AEDs over the year, showed deteriorating performances compared with untreated children (Fisher's exact test, p=0.043) but no relationship with remission was found.

In conclusion, 'epilepsy only' is less benign than the clinical impression holds. Not only did 45% of the children in our sample still have seizures after one year, but already at diagnosis progress at school was worse than in controls, in spite of a significantly greater amount of educational assistance. Management of children with this 'benign' category of epilepsy should, therefore, include careful monitoring of school progress.

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Table I: Progress of children with 'epilepsy only' and their non-epileptic classmates with respect to overall rate of progress; teachers' reports at diagnosis, and three and twelve months later.

Time	Obtained reports, n (%)		Worsened, n		Remained stable, n		Improved, n		Overall change
	Epilepsy	Control	Epilepsy	Control	Epilepsy	Control	Epilepsy	Control	*p*
Before diagnosis	59 (86)	47 (68)	14[c]	0	40	45	5	2	0.000
After 3 months	60 (87)	53 (78[a])	10[b]	0	45	51	5	2	0.001
After 12 months	49 (71)	41 (62[a])	9[b]	1	30	36	10[d]	4	0.008

[a]One classmate dropped out after three months and 2 more after twelve months. Fisher's exact test – fallen off versus stable, [b] $p \leq 0.01$; [c] $p \leq 0.001$, gained versus stable; [d]Tendency (p=0.06).