

Survival in children with severe cerebral palsy: a further international comparison

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SIR—International comparisons of survival are of particular interest because significant differences may be indicative of the effects of the quality and the type of care provided in different countries. Research evidence from around the world demonstrates that the severity of disabilities is the primary determinant of survival for children with cerebral palsy.^{1–4} Valid inferences from different countries may thus only be drawn from controlled comparisons of persons who have similar patterns of disabilities. This ensures that observed differences are not simply due to imbalances in the ‘case mix’.

Several international comparisons of survival in children with cerebral palsy have been reported in the literature. The 2001 study of Shavelle et al.² found remarkable similarity in the 20-year survival experience of severely disabled 5-year-old children from California and Australia with IQ<20 and 20–34. In total, these authors considered twelve comparison groups based on combinations of intellectual functioning (IQ<20; 20–34; 35–49; and 50–69) and motor dysfunction (mild, moderate, and severe) and found that survival was not significantly different between the two countries for any group (lowest $p=0.23$).

More recently, the 2008 study of Strauss et al.³ demonstrated very similar 20-year survival for 4-year-old children from California and the UK with four severe disabilities in ambulation, cognition, hand use, and vision. Conducting controlled comparisons of this type for other countries has proven

difficult in practice because common definitions of severity are not always available. Further, even when common definitions are found, stratified study samples often become too small to yield precise survival probability estimates.

The recently published Westbom et al.¹ study of survival for Swedish children with cerebral palsy provides the most recent opportunity to re-examine this important question. These authors used the Gross Motor Function Classification System (GMFCS),⁴ a five-level scheme that describes the severity of motor disabilities, to stratify their population. Their original article presents a Kaplan–Meier survival curve based on data from 102 children identified as GMFCS level V (highest severity), of whom 25 died during the 1994 to 2010 study period. There were not enough observed deaths to provide reliable estimates for less severely affected children.

We plotted the Westbom et al.¹ probabilities of survival for children in GMFCS V, conditioned on survival to age 3. Each age-specific conditioned survival probability was calculated as the ratio of the probability of surviving to that age to the probability of surviving to age 3. The actual survival probabilities for the Westbom et al.¹ cohort were digitally measured from the published curves. Using data from California, we identified 1283 children aged 3 years in GMFCS level V who were formally assessed between 1988 and 2002. We followed each child’s vital status until 31 December 2006 (the most recent available data) and found that 322 had died. We constructed the Kaplan–Meier survival curve for this group. The survival estimates for the two countries were quite similar, with 63% and 67% survival from age 3 to 19 years for Swedish and Californian children respectively (see Fig. 1).

As discussed in several articles from California, the UK, and Sweden, gastrostomy-dependence is consistently identified as a significant marker for increased mortality.^{1,3,5} The large California sample allowed further within-sample comparisons of survival for children in GMFCS V according to the presence of a feeding tube (see Fig. 1). In the vast majority of cases in California, well over 90%, feeding tube refers to a gastrostomy; by contrast, nasogastric feeding is rarely used for extended periods. There were 1007 orally-fed children of whom 221 died, and 276 tube-fed children of whom 101 died. The difference in survival was significant (log-rank $p<0.001$), with 72% and 55% survival to age 19 years for the two subgroups respectively.

The California and Sweden comparison here is consistent with previous research demonstrating similarities in survival between California, UK, and Australian children with cerebral palsy who have similar patterns of disabilities. It represents a significant additional piece of evidence that survival probabilities are remarkably similar for very severely disabled children in developed countries. This may suggest that there are not

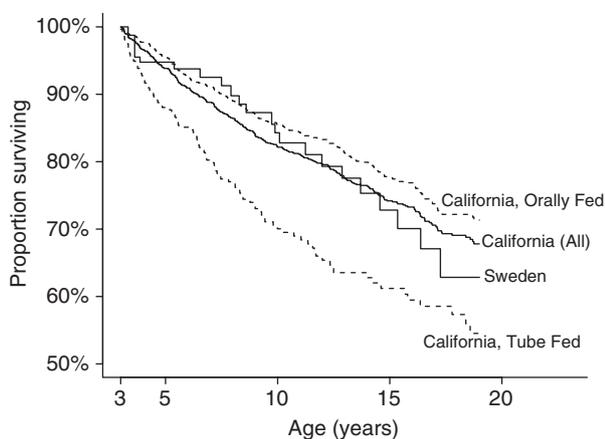


Figure 1: Survival in Sweden and California, Gross Motor Function Classification System level V.

major differences in the type and quality of care delivered in these countries.

We encourage other authors who report on health outcomes to continue to use properly validated reporting measures, like the GMFCS, that facilitate such controlled

comparisons. Finally, we note that there are several other factors (e.g. dependence on a feeding tube) that should be considered in the estimation of survival probabilities or the life expectancy for an individual child.

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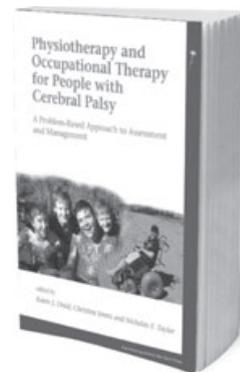
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