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Editorial

## Life expectancy in hydranencephaly

This issue of *Clinical Neurology and Neurosurgery* contains a case report on the so far longest survival of an individual with hydranencephaly documented in the clinical literature. The report is a significant addition to the few prior publications on the topic, which has never been subjected to systematic study. The lack of formal survivorship data for hydranencephaly leaves scant grounds for prognosis when the condition is diagnosed, and adds to the many question marks that surround our knowledge of the consequences of drastic loss of hemispheric tissue early in development.

As a neuroscientist concerned with subcortical competences, and those of the brainstem after telencephalic loss in particular, I have taken an interest in hydranencephaly as one source of information in this regard. My orientation about the condition has included participation in an Internet-based network of families with members diagnosed with hydranencephaly. It was founded in 1998 by Barb Aleman, and to date somewhere near 350 families world-wide have been active in the internet exchanges. This is a sizeable population of individuals with hydranencephaly, but it does not constitute a cohort from which survivorship statistics can be calculated directly. Membership in the network is in constant flux, and the fate of some of its individuals with hydranencephaly is currently unknown. As a proxy for a full survivorship analysis I have therefore plotted a survivorship curve based solely on the 116 individuals of this population for whom known dates of birth and death are available, presented in Fig. 1.

A striking and previously unknown fact emerges from Fig. 1. To a first approximation, the survivorship pattern exhibited here conforms to a so-called Type II survivorship curve, defined by the main descending diagonal of the graph. This means that beyond the steeper mortality during the initial 2 years of life, life expectancy in hydranencephaly is largely independent of age. Put differently, a child with hydranencephaly that has survived its first 2 years survives “indefinitely”, subject only to the average attrition rate. For the deceased individuals in the plot, after the first 2 years this rate averages a yearly attrition of 16%, but as noted in the fig-

ure legend, the exclusion of currently surviving individuals means that this figure is an over-estimate. The oldest of these excluded individuals is currently 28 years old, and even older cases are known, such as the woman with hydranencephaly described informally by Dr. S. Allen Counter, first seen at age 32, but surviving for a number of years more [1].

Given these circumstances, it stands to reason that quality of care must have a significant impact on life expectancy in hydranencephaly. These children are medically fragile. Their first year of life is typically marked by medical emergencies occasioned by uncontrolled epileptic seizures, pulmonary sequels to reflux and aspiration, unregulated intracranial pressure (hydrocephalus), problems with temperature regulation, and more. Presumably the steeper mortality in the first year of life is related to these problems, and then settles towards the diagonal as the problems are brought under control by medication, shunting, and other interventions.

The type and quality of care is also a factor to consider in relation to the neurological status and competences of individuals with hydranencephaly. The case report in this issue conforms well to the diagnosis of developmental or persistent vegetative state commonly applied to hydranencephaly [2], but there are also cases of hydranencephaly exhibiting a responsiveness to their surroundings incompatible with the classification vegetative state [3]. The four individuals reported by Shewmon et al. were cared for in their homes, in family settings providing rich stimulation and individual attention, and were also assessed in their familiar environments, while the patient reported here by Bae et al. comes from and was assessed in institutional settings. In a recent publication I have provided a detailed examination of functional reasons for expecting individuals with early and drastic (even complete) loss of cortical tissue to display forms of coherent responsiveness to their surroundings incompatible with a diagnosis of vegetative state [4]. Children with hydranencephaly are an important source of information in this regard, and careful assessment of their behavioral competences and responsiveness under optimal conditions promises to add significantly to our understanding of the spectrum of subcortical competences.

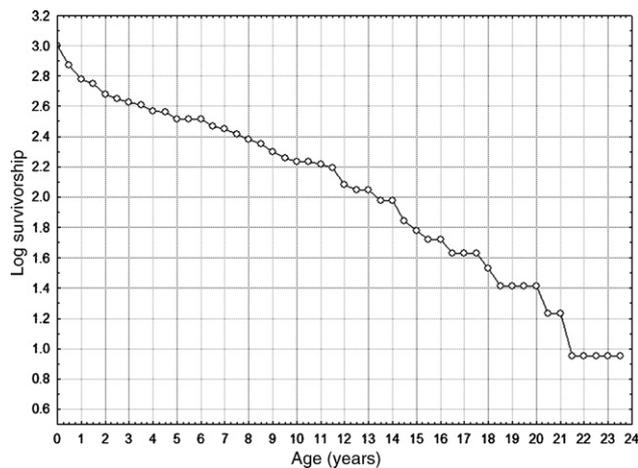


Fig. 1. Pseudo-survivorship curve for 116 deceased individuals diagnosed with hydranencephaly. Data were converted to survivorship per thousand, logarithmically transformed, and plotted as a function of age at half-year intervals. Note that others in the population from which these individuals were drawn are surviving beyond the oldest individual plotted in the graph, and that the exclusion of surviving individuals has the effect of inflating attrition figures.

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