Reviewer's report

Title: Incidence rates of progressive childhood encephalopathy in Oslo, Norway: a population based study

Version: 2 Date: 11 March 2007

Reviewer: Paul Uvebrant

Reviewer's report:

General

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Major Compulsory Revisions (that the author must respond to before a decision on publication can be reached)

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Minor Essential Revisions (such as missing labels on figures, or the wrong use of a term, which the author can be trusted to correct)

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Discretionary Revisions (which the author can choose to ignore)

The paper Incidence rates of progressive childhood encephalopathy in Oslo Norway: a population based study, by Stromme et al. deals with the complicated and important issue progressive encephalopathies in children. It is almost impossible to delineate the group and the contributing disorders vary between geographic areas and ethnic groups. With this in mind the authors have done the best possible attempt to classify and define the group and a little more vaguely the population at risk.

The paper contributes important information that will help others to structure their thinking of the concept and to get an opinion about the size of the problem. The single disorders are rare, sometimes extremely rare, but taken together they constitute a number of children that equals other large neuroimpairment groups such as neural tube defects, subtypes of cerebral palsy and autism spectrum disorders.

The method used is adequate but is complicated by the fact that the diagnostic possibilities have developed during the years studied. It is difficult to interpret the incidence of for example mitochondrial disorders as they were hardly known and the diagnostic methods were not developed in the early years of the study. Was there a shift from unknown to defined diseases over the years?

There is also a lack of information about the population studied. The only criterion is that the child should live or have lived in Oslo at any time of the study period. What was the proportion of immigrants? Did that proportion change over time? The number of live births is used but the population at risk is not necessarily born in Oslo but in other parts of the world.

The use of incidence rates per person years is probably adequate but difficult to interpret, despite the extensive description of the method. It would be helpful if some references were given to other studies using this method, for example in childhood epilepsy or other disorders.

Is Figure 1 really necessary?

Some details: Was age at diagnosis or age at onset of symptoms used? These two may differ by years in these disorders. In Methods is stated that both were recorded but the result is not presented. To use the time when “a downhill course” was recognized as the age at diagnosis could be questioned as small children develop fast and the first symptom of a degenerative disorder may be a slowing or arrest of development, rather than the loss of acquired abilities or a “downhill” course.

The Results are adequately presented but the last two sentences may be omitted as they only describe what was not found in the study. There are certainly more than the four diseases mentioned that did not occur during the study period.

The Discussion mainly repeats rather than discusses and interprets the results but is otherwise appropriate.
The Conclusion is a short summary of the findings but do not add anything about the importance, implications or meaning of them. The last sentence that there was a delay from onset to diagnosis in children with neurodegenerative disorders is not supported by any results presented.

The Title and Abstract are adequate and supported by the results.

**What next?:** Accept after discretionary revisions

**Level of interest:** An article of importance in its field

**Quality of written English:** Acceptable

**Statistical review:** Yes, but I do not feel adequately qualified to assess the statistics.

**Declaration of competing interests:**

I declare that I have no competing interests