

Childhood dementia

Background

Cognitive decline in children has received little attention to date. The literature is replete with rare syndromes, metabolic pathways and uncertainty as to management and prognosis. What the conditions have in common has frequently been sacrificed to their distinguishing characteristics with the result that the research, clinical and service efforts associated with these conditions have been fragmented. What has not been known is how often childhood dementia occurs within the community and the range and frequency of its various aetiologies.

Objectives

- To identify the incidence of childhood dementia in Australia
- To determine the perceived adequacy of resources

Case Definition

The definition was changed during the study.

Initial definition

Any child who suffers from an illness fulfilling the following criteria:

*Multiple losses of already attained development skills **with***

*Duration of illness greater than 3 months **and***

*Skill loss most likely due to CNS dysfunction **and***

*Evidence of generalised (not focal) brain dysfunction **and***

The condition must not be explicable in terms of acute drug toxicity, prolonged ictal confusion or other causes of delirium.

Final definition

*As above except that the multiple losses needed to be **progressive**.*

Results and discussion

For consistency, only cases with progressive deterioration are discussed. Overall there were 179 reported cases in 18 months. 148 cases were confirmed or probable and 31 cases were uncertain. This yielded an annual incidence of definite or probable childhood dementia in Australia, excluding cases with Rett syndrome, of 2.6/100,000 children under age 15. However, under-reporting is known to have occurred and as such the above figure, although still the best available for this disorder in Australia, is an underestimate of the true incidence of this disorder.

Of the 148 confirmed cases 49 (33%) were idiopathic. The fact that the cause of such a large proportion of cases remains undetermined is of interest and suggests the need for further research in this field.

Following completion of the childhood dementia study, the British Paediatric Surveillance Unit commenced a study of progressive intellectual and neurological deterioration (PIND), including Creutzfeldt-Jakob disease (CJD) from May 1997. This condition is of interest in the UK with recent concern about possible human disease following ingestion or contact with Bovine Spongiform Encephalopathy (BSE) affected cattle. The study definition is very similar to that used for the childhood dementia study and attests the need for further work in this field.

Investigators

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