Summary of neurological deficits and methods of assessment

Over the last decade, neuropsychological impairments have been increasingly described in patients with Langerhans cell histiocytosis. Neurological deficits can range from minimal signs, such as mild tremor, to severe symptoms, such as seizures, ataxia, and dysarthria which may affect "quality of life."1-4

Cognitive testing has shown lower-than-average scores in intelligence tests, memory, learning and language problems, and academic attainments5. Significant neurological symptoms are seen in a few patients. Single-center long-term follow-up studies have shown that LCH patients with multisystem disease, and in particular those with CNS changes, show decreased cognitive function in comparison to LCH patients without such lesions. Another intriguing finding was the detection of lower scores in tests for short-term auditory memory which are subtests of the Verbal IQ evaluation. Interestingly, these deficits can be seen not only in patients with obvious intracranial lesions, but also in those with minimal changes on MRI or psychological problems. Brainstem acoustic-evoked potentials are a promising tool to assess impairment in the central auditory pathway, which might partly be responsible for problems in short-term auditory memory.

It is very important that neuropsychological deficits be recognized early, as providing specific support to these patients may improve learning and outcome. Therefore, it seems warranted for LCH patients who suffer from multisystem disease and/or intracranial disease to be tested at regular intervals. For neurological assessment, the widely accepted Ataxia Rating Scale6 and the Expanded Disability Status Scale7 are used, while the age-appropriate Wechsler Scales for IQ and memory are used to evaluate neuropsychological function. Behavioural development and life quality can be evaluated by the Child Behaviour Check List and appropriate questionnaires for health-related quality-of-life assessment, such as the Health Utility Index (HUI)8 (Torrance G, Feeny D, et al. 1996).

References: