

Cognitive effects of cerebellar disorder

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Scientific abstract

The project addresses the effect of cerebellar disorder on cognition. This aspect of cerebellar disorder has received less attention than motor deficits, but represents an additional burden for patients and their carers that is inadequately defined. The work will test the hypothesis that cerebellar disorders produce cognitive deficits, and examine the specific cognitive profile. The work will be carried out on a cohort of greater than twenty patients with spinocerebellar ataxia type 6 who have been identified and characterised fully in terms of the genotype and neurological phenotype by Professor Chinnery. They will undergo detailed structural imaging with MRI to define the extent of the cerebellar damage. The cognitive profile will be assessed using sensitive, robust and internationally accepted measures of cognitive function including WAIS III, WMS III and the Delis-Kaplan Executive Function system, under the supervision of Dr Welch. The group will additionally undergo testing of auditory perceptual analysis in the laboratory of Professor Griffiths to test the specific hypothesis that they have a deficit in predictive coding of sensory stimuli. Proof of concept for the study is provided by cognitive data on an initial cohort of ten subjects that has been reported to the World Congress of Neurology in 2006.

Lay summary

This work tests the hypothesis that disorders of the cerebellum affect not only movement but also the ability of patients to carry out certain types of computation. Specifically, there is evidence based on our pilot work that these disorders affect the perception of space and time and our ability to predict the way in which visual patterns and sound change over time. If the concept can be proven it will have important implications for patients who suffer from any cerebellar disorder. Such deficits in sensory prediction may be practically important in contributing to falls, and have been comparatively little studied compared to the motor deficit. We propose to carry out the work on a group of patients with an inherited ataxia where the disease only affects the cerebellum (SCA6) to allow us to obtain clear information about the effect of cerebellar damage in the absence of other confusing factors.

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