Reviewer's report

**Title:** Cognition is only minimally impaired in Spinocerebellar Ataxia type 14 (SCA14): a neuropsychological assessment of ten Norwegian subjects

**Version:** 1  **Date:** 24 September 2013

**Reviewer:** Ludger Schöls

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Wedding and colleagues systematically investigated cognitive function in 10 individuals from 2 Norwegian families with spinocerebellar ataxia type 14 (SCA14). They found focal deficits in verbal executive function, psychomotor speed, working memory, attention and visual learning. This is a well-structured approach that considered motor handicaps in selection of the test battery. Only 10 subjects were investigated but this is a reasonable cohort given the rarity of SCA14. All patients carried the same PKCG mutation, p.H139Q.

One weakness of this study is the lack of a matched control group especially with the same linguistic background (mother tongue). The authors used published control cohorts for comparison but social background and mother tongue may influence test performance and especially rapid generation of words starting with specific letters as one of the tasks with major deficits in the SCA14 cohort. This may also explain some of the differences in neuropsychological testing observed in comparison to other SCA14 cohorts described before.

Within the SCA14 group the authors included one individual who claimed to carry the mutations but was clinically asymptomatic. If included in this study at all, genetic test results must be confirmed since this individual is almost 10 years older than the latest age of onset in the other patients. Anyway, her results should be presented separately and should not be merged with patients with manifest disease to prevent bias.

In this cross-sectional neuropsychological study the authors did not observe accentuation of cognitive deficits with duration of disease. On the contrary, they speculate that the tendency to better test performance with longer disease duration reflects compensatory brain plasticity that helps to improve cognitive function with time. This is highly speculative and neither a significant result nor supported by longitudinal data. Such speculation should be omitted at least from the abstract and conclusions.

**Level of interest:** An article whose findings are important to those with closely related research interests

**Quality of written English:** Acceptable
**Statistical review:** No, the manuscript does not need to be seen by a statistician.

**Declaration of competing interests:**

I declare that I have no competing interests