Reviewer's report

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

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Reviewer: Wendy H Raskind

Reviewer's report:

The question posed by the authors is well defined and appropriate. Although cognitive impairment, particularly affecting executive function, in SCA14 has been attributed to mutations in PRKCG, the data are scanty and corroboration is a worthy goal. It is also not clear whether the cognitive phenotype is restricted to certain genotypes. The authors used an appropriate and well-described battery of tests in this physically impaired group of subjects and present all the data in an understandable way.

Major Compulsory Revisions

1. Unfortunately, a major limitation of the work was not even mentioned: the absence of comparison data from unaffected biologic relatives. Both genetic and environmental factors contribute to performance on the tasks used in this study and the affected and unaffected members of the families share these contributors. You cannot just assume that these families would have exactly population-average performance on all the tests. In fact, the authors acknowledge that the families are from industrial areas and may lack in academic experience. Therefore, the conclusions are difficult to assess.

2. Similarly, the title may be wrong. What if the unaffected people in the family have superior performance on the tests? Then the impairment related to SCA14 may be more than minimal. Given the importance of the topic, it is worth administering the battery of tests to the first-degree relatives of the affected people.

Discretionary Revisions

1. When cognitive test results for the unaffected family members are available, a related, but less important, comparison of interest would be to analyze the families independently. This might provide insight on whether there is a similar cognitive phenotype in the two families that share the same PRKCG mutation on different genetic and environmental backgrounds.

2. With respect to the relationship of IQ to duration of disease, the number of subjects is so small that, without a better comparison group, “trends” seem meaningless to me.

Level of interest: An article of outstanding merit and interest in its field
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 I that I have no competing interests.