Reviewer’s report

Title: Mutations at codon 178, 200-129, and 232 contributed to the inherited prion diseases in Korean patients

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Reviewer: Inga Zerr

Reviewer’s report:

This is an interesting and well-written report on genetic prion disease patients from Korea. Of interest is the case description of the patient with 178 mutation. This mutation has been linked to fatal familial insomnia with a distinct clinical syndrome before but increasing evidence is available that even in these patients clinical heterogeneity exists. Of interest is also the observation on MRI and CSF abnormalities in these patients, which has not been reported before. Most data on the CSF in FFI from the literature report negative 14-3-3 test in patients with this mutation and normal MRI scans. This should be stressed and the literature should be amended.

Level of interest: An article of importance in its field

Quality of written English: Needs some language corrections before being published

Statistical review: No, the manuscript does not need to be seen by a statistician.

Declaration of competing interests:

I have no competing interests