Author’s response to reviews

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

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Version: 3 Date: 6 May 2009

Author’s response to reviews: see over
Dear Editor (Sir)

We are very glad to submit our revised manuscript (MS number is 1600596070258814) to the BMC Infectious Diseases. The point-by-point responses to the concerns follow as next page. We also addressed the comments in memo boxes in revised manuscript and attached the ethics document for this study to editorial@biomedcentral.com. We are looking forward to hearing from you soon.

Sincerely,

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Itemized list of our specific responses to each of the reviewer’s comments and request.
(with red phrases)

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1. **The name and details of the ethics committee which approval for this study.**

   • The name is『KOREA CENTERS FOR DISEASE CONTROL AND PREVENTION INSTITUTIONAL REVIEW BOARD (IRB)』
   
   -> Attached the approved document to BioMed Central Editorial <editorial@biomedcentral.com>

2. **Referee 1 : 1065093662623749 comment**

   Major Compulsory Revisions

   1. Figure 3. Brain MRI. I see the lesions in DWI but not in T2-FLAIR images. Moreover, in the DWI image of the M232R patient there is high signal in the occipital area. Is that a lesion or an unspecific signal?
   
      • FLAIR figure is exchanged to new one with a lesion in the parietal lobe.

   2. Figure 3. EEG. The EEG on the left does not seem typical for CJD. I would suggest to choose a better EEG for this patient.
   
      • The EEG was that of the early phase, so we exchanged to the MRI figure of the early phase. Instead of, T2-FLAIR image was that of the follow up stage.

   3. Discussion. In the discussion the Author should avoid to repeat what has been already described in the result session.
   
      • Corrected or removed -> In manuscript

   Minor Essential Revisions

   1. PRNP should be in italics
   
      • Changed -> In manuscript

   2. Background, line 8. In this context, it would be better to use the generic term “genetic TSE” rather than familial CJD.
   
      • Changed -> In manuscript

   3. The first sentence of the discussion should be removed.
   
      • Removed -> In manuscript

   Quality of written English: Needs some language corrections before being Published
   
      • Edited by native speaker in professional editing organization.