Reviewer's report

Title: A Novel Pathogenic Variant in OSBPL2 Linked to Hereditary Late-onset Deafness in a Mongolian Family

Version: 1 Date: 03 Jan 2019

Reviewer: Barbara Vona

Reviewer's report:

The revised manuscript entitled, "A novel pathogenic variant in OSBPL2 linked to hereditary late-onset deafness in a Mongolian family" has improved significantly compared to the first submitted version. I have reviewed the revision and have identified a few minor suggestions for changes to the text to improve clarify. I have also outlined several areas that need English improvement and made suggestions when possible.

Minor revisions:

-Abstract: include the p. position after the c. position

-Include the OMIM number for OSBPL2

-Figure 2: enlarge the figures. The x- and y-axes are unclear and the x-axis does not look to be completely labelled

-State the LOD scores in the results section

-Line 222: check that OSRB should be OSBP

English suggestions:

-Line 43: seriously affects

-Line 48: Based on whether…

-Line 53: …60-75% is autosomal recessive,… (note, this number seems a bit low for describing the proportion of hearing loss that is autosomal recessive. I would recommend finding an updated/newer reference that provides updated statistics about this.)

-Line 54: …X-linked or mitochondrial [4].

-Line 56: Replace the word delayed with late-onset
-Lines 148-149: instead of saying "at the end of chromosome 20," list the cytogenetic band.

-Line 168: … c.158_159delAA, co-segregated in the family…

-Line 176: OSBPL2 encodes a receptor

-Line 177: To find out whether the frameshift…

-Lines 177-178: It sounds too strong of a statement to say lipid metabolism leads to hearing loss, especially when lipid levels are normal in individuals with hearing loss?

-Lines 178-179: in members of this family with the confirmed genotype…

-Line 180: use the word indistinguishable instead of not distinguishable

-Line 196: …starting from the 53rd amino acid…

-Line 204: The pathogenic variant in the OSBPL2 gene…

-Line 208: …in the OSBPL2 gene was…

-Line 231: …as well as by studying animal models with…

-Figure 1 legend: Modify to something like, "Members with genotypes noted were recruited for this study"

-Figure 3 legend: line 154: …around 1.5 (shorten this sentence).

-Figure 3 legend: line 155: B. OSBPL2 is located on chromosome 20q13.33 (red bar).

-Supplementary Table 3: check the spelling of "panel", as well as "autosomal recessive" and "X-linked"
Are the methods appropriate and well described?
If not, please specify what is required in your comments to the authors.

Yes

Does the work include the necessary controls?
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