Author’s response to reviews

Title: Patients affected with Fabry disease have an increase incidence of progressive hearing loss and sudden deafness: an investigation of twenty-two hemizygous male patients

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Dear Editor,

We have now carefully revised our manuscript and we would like to thank the reviewers for their constructive comments. We have followed all recommendations of the reviewers:

Reviewer 1
1) A table with demographics and clinical characteristics of patients, including a column on hearing status has been added. The presence or absence of renal, cardiac, and cerebrovascular disease has been noted and the values for GFR have been given in parenthesis (Table 1).
2) A recommendation to systematically perform an audiogramm prior to initiate enzyme replacement therapy has been clearly mentionned (page 16)

Reviewer 2
1) The comment of the reviewer is quite relevant. However it was not possible for us to perform exhaustive ENT investigations of unaffected FD members due to the amount of work this would represent. Of note, no family history of inherited deafness was found for any FD uninvolved family member during medical history records. This point has been added to the manuscript. In addition, it should be noted that standardised values from the World Health Organisation have been used for statistical analysis, further strengthening our results.
2) The historical exposure to aminoglycosides is indeed an important point which had been searched during medical history. For clarity, this point has now been added in the Material and Method section (page 4).
3. The original table 1 has been deleted and replaced by a revised table showing demographics and clinical data.

We also carefully checked all items of the manuscript formatting checklist.
We hope that the manuscript is now satisfactory for publication in BMC Medical Genetics.

Yours sincerely,

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