Reviewer’s report

Title: Patients affected with Fabry disease have an increase incidence of progressive hearing loss and sudden deafness

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Reviewer: Gregory Grabowski

Level of interest: A paper of considerable general medical or scientific interest

Advice on publication: Accept after discretionary revisions

This is an extensive description and characterization of hearing loss in an unselected population of Fabry hemizygotes. The conclusion is that hearing loss and ear functional abnormalities are a significant and underreported clinical problem in Fabry disease. The studies are very well executed and important for the future developments in therapies for this disease.

Comments:

1. Because deafness is common in the general population, it would be useful to provide information about the Fabry disease uninvolved family members to support the contention that hearing loss is related to Fabry disease.

2. The authors address the issue of a variety of ototoxic agents, but historical exposure to aminoglycosides might be an important addition to the manuscript.

3. The tables, especially table 1, are somewhat tedious, but complete. Some attention to shortening table 1 or condensing might be appropriate.

Competing interests:

Have been a consultant to TKT and Genzyme.
Do not hold stock in any biotech company including the above.
No other competing financial interest.
No non-financial competing interests.