Reviewer’s report

Title: HDR Syndrome with a Novel Mutation in GATA3 Mimicking a Congenital X-linked Stapes Gusher: a case report

Version: 2 Date: 16 Aug 2017

Reviewer: M. Andrew Nesbit

Reviewer’s report:

All my previous queries have been dealt with.

1 more point

"140 The molecular analysis of our patient revealed a heterozygous deletion of AT nucleotides 141 in exon 6 (c.1201_1202delAT) of the GATA3 gene causing a frameshift at the 401st codon 142 with a premature stop codon at the 106th codon of the reading frame (p.Met401Valfs*106)."

This cannot be described as a premature stop codon as the normal stop codon is at 444. Here there is a C-terminal extension until 106 codons after the frameshift at codon 401. Thus, the total protein is 506 amino acids long which includes 106 missense amino acids at the C-terminus. Your nomenclature for the mutation is correct, your interpretation for what this means is not.

Please revise text accordingly.

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?
If not, please specify which controls are required in your comments to the authors.

Yes

Are the conclusions drawn adequately supported by the data shown?
If not, please explain in your comments to the authors.

Yes

Are you able to assess any statistics in the manuscript or would you recommend an additional statistical review?
If an additional statistical review is recommended, please specify what aspects require further assessment in your comments to the editors.

Not relevant to this manuscript

**Quality of written English**
Please indicate the quality of language in the manuscript:

Acceptable

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