Reviewer’s report

Title: Divergent phenotypes in siblings with identical novel mutations in the HNF-1α gene leading to maturity onset diabetes of the young type 3

Version: 0 Date: 29 Mar 2016

Reviewer: Helen Lunt

Reviewer's report:

I enjoyed reading this revision, as it was easy to follow.

I have a few discretionary comments.

1. Whilst patient therapy (and response to it) is not the focus of the article, it is nevertheless of interest to clinical readers and helps place this Case Report within a clinical context. I felt this information became ’buried’ by including it only in Figure 1. It should have been included in the main text.

2. Background. Regarding the statement 'MODY ...(has a)..clinical appearance similar to type 2 diabetes', maybe a better statement would be that there is overlap in the clinical characteristics of both conditions.

3. Determination of diabetes related antibodies. The Table indicates that GAD, ICA and IA2 were measured but the text (page 4 first paragraph) also mentions tyrosine phosphatase and insulin antibodies. Consistency of terminology and reporting would reduce the potential for confusion.

4. Results section - first line on Page 5. There is discussion about diabetic complications during pregnancy. Am assuming these are obstetric diabetic complications rather than traditional non pregnancy related complications? This sentence was confusing and could be reworded.

5. Page 6, first paragraph; 'Temporal decrease in triglycerides to 4.5mmol/L'. I did not follow what was meant by this as I could not find a value that equated to 4.5mmol/L in Table 2.
6. It was not clear to me whether Patient 1 and Patient 2 omitted their anti-diabetic therapy prior to their OGTT and the lipid tolerance tests. (Apologies in advance if I missed this within the text). Clarification of this, perhaps as a footnote after the Tables, would help with interpretation of the Tables.

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.

I am able to assess the statistics

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

Acceptable

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