Author's response to reviews

Title: X-linked Hyper IgM (HIGM1) in an African kindred: The first report from South Africa

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Dr E Parkin
Editorial Administrator
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Dear Dr Parkin

MS: 6386098622198204
X-linked Hyper IgM (HIGM1) in an African kindred: The first report from South Africa

In response to the reviewer's comments the following modifications were made to the manuscript:

Reviewer: John Ziegler

1. 'Phorbol' the spelling error was corrected.
2. Family tree: The pedigree of the affected family was added (Figure 1) and the remaining figures re-labeled.
3. The 'affected boy' in the results section under "Nucleic acid analysis" was changed to 'the second eldest, affected male'.
4. Last sentence of the first paragraph of the discussion: The word "Unfortunately" was removed and the sentence re-phrased.
5. Last statement of the paper was changed from: 'In order to gain a greater understanding of the spectrum of immunological and genetic disorders in sub-Saharan Africa, existing laboratory infrastructure should be strengthened to enable more detailed analysis.' TO 'Recent progress should lead to a greater understanding of the spectrum of immunological and genetic disorders in sub-Saharan Africa.'

Reviewer: Andrew Cant

6. More clinical detail: Under Clinical and Laboratory Data (Results Section) the following comment was added about neutropaenia - 'Neutropaenia was intermittently documented during the clinical course of the two younger affected males. Neutropaenia was not documented in the eldest male.'

We are unable to provide information about the organisms causing pneumonia as detailed studies of the respiratory secretions were not done.
7. The following comment on the susceptibility to PCP and cryptosporidial infection was added to the second last paragraph of the introduction: 'Failure to isotype switch, as well as failure to activate Kupffer cells and pulmonary macrophages increases the risk for Pneumocystis carinii and Cryptosporidial infection [7].'

Furthermore, an additional paragraph was added to the discussion section (2nd last paragraph) which explains why carrier detection was only extended to the mother.

Yours sincerely

Dr Brian Eley