Reviewer's report

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

Version: 1 Date: 21 October 2003

Reviewer: Andrew Cant

Reviewer's report:

This is an extremely well written paper that clearly and concisely describes the clinical, immunopathological and molecular features of this kindred with CD40 Ligand deficiency. None of the findings are novel but this first complete description in a sub-Saharan African family is important, as it highlights the importance of considering genetically defined primary immunodeficiency in this patient population. The methods are appropriate and well described or referenced. The data appear sound. Their is a paucity of clinical detail with no mention of whether neutropenia was present nor isolation of organisms causing pneumonia. The manuscript adheres very well to standards for reporting. The discussion and conclusions are balanced and supported by the data; the title and abstract very accurately convey what has been found and the writing is excellent.

a) Discretionary Revisions (which the author can choose to ignore)

i) More clinical detail - if available.
ii) A sentence in the introduction on interaction between CD40 Ligand on T cells & CD40 on Kupffer cells and pulmonary macrophages, and how this might allow susceptibility to PCP and Cryptosporidial infection.

b) Minor Compulsory Revisions (such as missing labels on figures, or the wrong use of a term, which the author can be trusted to correct)

None

c) Major Compulsory Revisions (that the author must respond to before a decision on publication can be reached)

None

What next?: Accept after discretionary revisions

Level of interest: An article whose findings are important to those with closely related research interests

Quality of written English: Acceptable

Statistical review: No

Declaration of competing interests:

None