Author's response to reviews

Title: Subacute Sclerosing Panencephalitis: Results of the Canadian Paediatric Surveillance Program and review of the literature.

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Author's response to reviews: see over
August 14, 2005

Dear BioMed Central Editorial Board:

RE: MS: 8847892156591942

Thank you for providing the reviewers’ comments to us and I apologize again for the delay in our response, given the summer season it was difficult to get all the authors feedback in a timely manner.

Below are the responses to reviewers’ comments in the same format they provided them and attached is a new copy of the manuscript.

Reviewer: Brenda Banwell

General comments:
1. We have reduced the manuscript by shortening the case reports and removing the section on pathophysiology.
2. We have attempted to remove comments on our personal views as outlined in many of the reviewers specific points later in the review
3. Table 4. and the comments in the text although similar do present the data in two different forms. The text details the disease treatment according to drug whereas the table approaches therapy by levels of evidence. We have tried to eliminate the redundancies in other tables/text but feel that specifically Table 4. should remain.

Abstract
1. The wording has been changed from ‘will’ to ‘may’.

Introduction
1. The comment has been changed.
2. The sentence has been changed.

Methods
1. The suggested change has been made

Case reports
1. The phrase brief sleeping episodes was a symptom describe by the parents and as such we feel this should be reported as we have done. ‘Obvious ataxia’ has been removed.
2. No autopsy was done on any of the cases. Certainly not on case 1 and 2 and we have indicated this in the manuscript. For case 4 the report came through the CPSP and it was not indicated that this child had an autopsy.
3. The immunization record for case 2 has been clarified in the manuscript.
4. Only abnormal results will be numerically indicated. For longer lists of investigations that were normal we feel it appropriate to indicate ‘within normal’. In the cases 1, 3 and 4 the EEG findings suggested the diagnosis quickly and little
investigation beyond measles immunoglobulin from CSF were needed. Case 2 provided more diagnostic challenge hence the long list of investigation. We have absolutely tried to be consistent – in providing all the pertinent investigations done.

5. We report the fact that case 1 and 4 received immunization on entry to Canada to show all the possible exposures to measles virus. We are not aware that it makes any difference to the establishment or course of SSPE.

6. We do not have the specific results of the brain biopsy in case 4 as it was provided to us via the CPSP and we were limited in our access to information.

7. The patient had an Ommaya reservoir.

Discussion
1. Deleted ‘endemic areas’
2. We have not changed the presentation of the incidence figure because in the case of rare disease it is not uncommon to report cases per million population and in most of the epidemiology studies quoted in this paper the figures are quoted per million.
3. We have deleted the section on pathophysiology as suggested by both reviewers.
4. We have changed the description of the seizure semiology.
5. A comment about the importance of translation services has been made.
6. This degree of disability as measured by the NDI measure has been used to define fulminate course in past studies. This has been clarified in the manuscript.
7. We have deleted surprisingly
8. We have tried to improve the wording of this paragraph.
9. We have removed the statement ‘in the same manner’.
10. We have altered the numerous phrases the reviewer objected to and added a comment on the importance of palliative care programs.

Reviewer: Benedikt Weissbrich

Major compulsory revisions
1. We have tried to shorten the cases as suggested. We have added a statement about the difficulty of diagnosis in case 2.
2. We have added considerably to the epidemiologic component of the manuscript but due to concerns about the length of the manuscript we have placed the studies in Table 2 and reduced the text section.
3. We have deleted the section on pathophysiology and Figure 1. as suggested but wish the reviewer had specifically pointed out the inaccuracies for our educational benefit.
4. We have deleted the reference to this being a possible vaccine related case.
5. We have added a statement on the incidence of measles infection in Canada, but did not have available figures back to 1985 but to 1990 published in the Journal of Infectious Disease.

Minor Essential Revisions
6. We have changed the wording to ‘only known exposure’.
7. We have added dates in the case reports where available.
8. These changes have been added to case 2.
9. The CSF findings have been changed.
10. The CSF measles IgG was elevated.
11. We are not aware why a brain biopsy was performed given the diagnostic information. This case was not one that the authors were directly involved in and so our information was limited to that provided by the CPSP reporting physician.
12. No the CPSP for SSPE has not been continued and we know of only one other case since that time but this is known to us only through discussion with colleagues and not from any form of epidemiologic investigation.

Discretionary revisions
13. We hope that the formatting of the tables for publication will help to improve clarity.

Thank you again for considering our submission, we look forward to your response.

Sincerely,

Craig Campbell MD FRCPC