Reviewer’s report

Title: Hepatic mitochondrial dysfunction in Friedreich Ataxia

Version: 1 Date: 28 September 2011

Reviewer: Francesc Palau

Reviewer’s report:

In this manuscript the authors investigated the mitochondrial function in the liver of 16 Friedreich ataxia patients using the 13C-methionine-breath-test. Overall the research is very well designed and the working hypothesis is well established. The clinical methodology is also well designed and results are expressed is easy understating way. Conclusions are based on experimental data and interpreted properly.

This is an interesting paper in the field of searching biomarkers in FRDA. In the last decade clinical trials in FRDA have increased but a few biomarkers are available to add to clinical scales when evaluating clinical response. In spite that the differences observed in patients do not correlated with the age of onset, severity or GAA expansion size, results are really important in the field. From my point of view this paper deserves to be published in BMC Neurology.

MINOR COMMENTS

1. At the end of the Discussion section the authors should state that measuring of hepatic mitochondrial activity by 13C-methionine-breath-test does not seem to be a good biomarker on the follow-up of natural history of the disease of the response to drugs in clinical trials.

2. References style should revised to fit that of BMC Neurology if not.

Level of interest: An article of outstanding merit and interest in its field

Quality of written English: Acceptable

Statistical review: No, the manuscript does not need to be seen by a statistician.

Declaration of competing interests:

I declare that I have no competing interests