Reviewer's report

Title: Metabolic phenotype of methylmalonic acidemia in mice and humans: the role of skeletal muscle

Version: 1 Date: 1 June 2007

Reviewer: Moacir Wajner

Reviewer's report:

General
This is an original and nicely well planned and written article which demonstrates that methylmalonic acid (MMA) is highly produced in extra-hepatic tissues, like particularly skeletal muscle, kidneys and brain, of a murine model with a null allele at the methylmalonyl-CoA mutase mimicking methylmalonic acidemia. This probably explains why amino acid restricted formula does not substantially decrease tissue MMA levels and speak against the current treatment with liver transplantatron. Therefore I believe that this is To my mind, these data may clarify the pathomechanisms involved in the neuropathology of methylmalonic acidemia patients and besides creates new approaches that may be useful to treat patients affected by this disorder. Therefore, I believe that this is an important paper that deserves publication. I have no suggestion to improve this manuscript.

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

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

Discretionary Revisions (which the author can choose to ignore)

What next?: Accept without revision

Level of interest: An article of importance in its field

Quality of written English: Acceptable

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