Reviewer’s report

Title: Long-term continuous N-carbamylglutamate treatment in frequently decompensated propionic acidemia: a case report

Version: 0 Date: 19 Sep 2017

Reviewer: Anupam Chakrapani

Reviewer's report:

1. Do you believe the case report is authentic?

Yes

2. Do you have any ethical concerns? Please consider if local Institutional Review Board approval or ethical approval was obtained (if appropriate) and if the patient (or their parent or guardian in the case of children under 18) gave written, informed consent to publish this case and any accompanying images. A statement to this effect should appear in the manuscript.

Comments:

No

3. Does the Introduction explain the relevance of the case to the medical literature?

Yes

4. Does the article report the following information? Where information is missing, please specify.

a. The relevant patient information, including:
   - De-identified demographic information (age, gender, ethnicity) – Yes
   - Main symptoms of the patient – Yes
   - Medical, family and psychosocial history – Yes
   - Relevant past interventions and their outcomes - Yes
b. The relevant physical examination findings - Yes

c. Important dates and times in this case (if appropriate, organized as a timeline via a figure or table); if specific dates could lead to patient identification, consider including time relevant to initial presentation, i.e. initial presentation at T = 0, follow up at T = 1 month. Yes, but some further detail should be provided - see comments below

d. Diagnostic assessments, including:
   - Diagnostic methods –Yes
   - Challenges (e.g., financial, language/cultural) - Not applicable
   - Reasoning and prognostic characteristics (e.g., staging), where applicable - not applicable

e. Types and mechanism of intervention - Yes

f. A summary of the clinical course of all follow-up visits - Yes

Comments:

In relation to the time course of events, the frequency of admission is mentioned as 78 admissions in the first 9 years and twice in the next 6 years. It would be more relevant to compare the annual admission rates in the 2 or 3 years immediately before and after using long-term Carbaglu. This could be presented as a graph or a table.

5. Is the interpretation (discussion and conclusion) well balanced and supported by the case presented?

Comments: Yes

6. Is the anonymity of the patient protected? Please consider any identifying information in images such as facial features or nametags, whether the patient is named etc. If not, please detail below.

   Yes

7. Is the Abstract representative of the case presented?
Comments: Yes

8. Does the case represent a useful contribution to the medical literature?

Comments: Yes

9. Additional comments for the author(s)?

**Level of interest**
Please indicate how interesting you found the manuscript:

An article of importance in its field

**Quality of written English**
Please indicate the quality of language in the manuscript:

Acceptable

**Declaration of competing interests**
Please complete a declaration of competing interests, considering the following questions:

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2. Do you hold any stocks or shares in an organisation that may in any way gain or lose financially from the publication of this manuscript, either now or in the future?

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If you can answer no to all of the above, write 'I declare that I have no competing interests' below. If your reply is yes to any, please give details below.

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