Reviewer's report

Title: Carnitine deficiency presenting with encephalopathy and hyperammonia in patients with chronic enteral tube feeding

Version: 2 Date: 21 March 2012

Reviewer: Philippe Lheureux

Has the case been reported coherently?: Yes

Is the case report authentic?: Yes

Is the case report ethical?: Yes

Is there any missing information that you think must be added before publication?: Yes

Is this case worth reporting?: Yes

Is the case report persuasive?: Yes

Does the case report have explanatory value?: Yes

Does the case report have diagnostic value?: No

Will the case report make a difference to clinical practice?: Yes

Is the anonymity of the patient protected?: Yes

Comments to authors:

Journal of Medical Case Reports

Manuscript: Carnitine deficiency presenting with encephalopathy and hyperammonia in patient with chronic enteral tube feeding.

Authors: P. Ling et al.

General comments

Carnitine deficiency has been rarely reported in patients with long term tube feeding and this clinical observation draws the attention on a likely underestimate problem. Structure and length of the manuscript are adequate.

Specific comments

Title:
Hyperammonemia, rather than hyperammonia

Introduction:

Carnitine is an essential agent to allow mitochondrial beta-oxidation of fatty acids. In carnitine deficiency, cytosolic omega-oxidation of fatty acids will be used as an alternative pathway. Unoxidized fatty acids and byproducts of omega-oxidation may be responsible for adverse consequences of carnitine deficiency.

Case presentation: some data could be added if available and would increase the value and soundness of this report

- Was there any sign suggestive of carnitine deficiency in the medical history before the accident (muscle weakness, cardiac or liver dysfunction,…) ? Presence of such sign could suggest an underlying carnitine palmitoyl transferase II deficiency (adult form) for example. If the answer is negative, it could be briefly mentioned.

- Psychiatric medications: please provide details about patient’s medications.

- What is the composition of Isosource HN: lipids ?, carbohydrates ?, proteins ?. Carbohydrate and lipid content of the feeding could play a role in the development of carnitine deficiency. Proteins may influence ammonia production.

- Did the patient present any sign (clinical, echocardiography, ECG, 24 h-recording, biomarkers…) of cardiac dysfunction ?

- Was a test for ketones performed on urines ? renal function ?

- Presence of myopathy was difficult to assess clinically. Was there any biological sign of muscular dysfunction (creatine kinase, myoglobinuria) ?

- Was the excretion of acylcarnitine measured in urine, before and after supplementation?

Consent:

Accompanying image: no image is joined to the paper

References:

Year is lacking for ref. 8

Which of the following following best describes what type of case report this is?

- An unexpected event in the course of observing or treating a patient
- Findings that shed new light on the possible pathogenesis of a disease or an adverse effect

**Quality of written English:** Acceptable

**Declaration of competing interests:**

'I declare that I have no competing interests'