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ESPS PEER-REVIEW REPORT

Name of journal: World Journal of Hepatology

ESPS manuscript NO: 31238

Title: Postpartum hyperammonemia crisis in a female patient with ornithine transcarbamylase deficiency

Reviewer's code: 02456449

Reviewer's country: China

Science editor: Fang-Fang Ji

Date sent for review: 2016-11-09 14:48

Date reviewed: 2016-11-27 16:00

CLASSIFICATION	LANGUAGE EVALUATION	SCIENTIFIC MISCONDUCT	CONCLUSION
<input type="checkbox"/> Grade A: Excellent	<input type="checkbox"/> Grade A: Priority publishing	Google Search:	<input type="checkbox"/> Accept
<input checked="" type="checkbox"/> Grade B: Very good	<input checked="" type="checkbox"/> Grade B: Minor language polishing	<input type="checkbox"/> The same title	<input checked="" type="checkbox"/> High priority for publication
<input type="checkbox"/> Grade C: Good	<input type="checkbox"/> Grade C: A great deal of language polishing	<input type="checkbox"/> Duplicate publication	<input type="checkbox"/> Rejection
<input type="checkbox"/> Grade D: Fair	<input type="checkbox"/> Grade D: Rejected	<input type="checkbox"/> Plagiarism	<input type="checkbox"/> Minor revision
<input type="checkbox"/> Grade E: Poor		<input type="checkbox"/> No	<input type="checkbox"/> Major revision
		BPG Search:	
		<input type="checkbox"/> The same title	
		<input type="checkbox"/> Duplicate publication	
		<input type="checkbox"/> Plagiarism	
		<input type="checkbox"/> No	

COMMENTS TO AUTHORS

Hyperammonemia after delivery in a female patient with ornithine transcarbamylase deficiency (OTCD) can be fatal. In this case report, authors have discussed the cause and treatment of hyperammonemia in a female patient with OTCD after delivery. This case indicates that it is important to perform early intervention before hyperammonemia occurs in patients with OTCD or carriers after delivery. I think that this paper is suitable for publication in your journal.

(Author's response)

Thank you for your comments and your consideration.



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Name of journal: World Journal of Hepatology

ESPS manuscript NO: 31238

Title: Postpartum hyperammonemia crisis in a female patient with ornithine transcarbamylase deficiency

Reviewer’s code: 00054001

Reviewer’s country: Japan

Science editor: Fang-Fang Ji

Date sent for review: 2016-11-09 14:48

Date reviewed: 2016-11-30 10:07

CLASSIFICATION	LANGUAGE EVALUATION	SCIENTIFIC MISCONDUCT	CONCLUSION
<input type="checkbox"/> Grade A: Excellent	<input type="checkbox"/> Grade A: Priority publishing	Google Search:	<input type="checkbox"/> Accept
<input type="checkbox"/> Grade B: Very good	<input type="checkbox"/> Grade B: Minor language polishing	<input type="checkbox"/> The same title	<input type="checkbox"/> High priority for publication
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		<input type="checkbox"/> Plagiarism	
		<input type="checkbox"/> No	

COMMENTS TO AUTHORS

The case the authors want to report may be interesting. However, I could not understand issues worth reporting in the current form. The authors should present their argument why this case is worth being discussed vigorously. For this purpose, the authors should address the followings. Major 1. What point(s) was rare in this case? 2. What point(s) was novel in this case? 3. Why the authors do consider this case to be worth reporting? Minor 1. The authors used terms “postpartum” or “after delivery”. These seemed misleading as newborn to me. I think other terms should be used for avoiding misleading: e.g. in a OTCD patient immediately after giving birth, etc. 2. The authors have reported their excellent outcomes of managing UCD patients without liver replacement. In “Introduction” section, I think they should show their know-how for their excellent outcomes. Almost all of descriptions in “Introduction” of the current form should be omitted and replaced by that. 3. More extensively detailed description must be required in Case presentation. Causes the authors considered at that time and interventions for eradicating those causes must be presented in Case presentation, not in Discussion. 4. In “Discussion” section, it should be more extensively

discussed whether this episode could be avoided with some preventive cares or not. Without that, lessons from this case will not benefit future cases. 5. Extensive English editing must be done.

(Author's response)

Thank you for your comments and your consideration.

Major 1. What point(s) was rare in this case?

(Author's response)

Thank you for your question. OTCD is an X-linked metabolic disorder, with an estimated prevalence of 1 per 80,000 live births. The clinical presentation in female patients with a heterozygous OTCD gene mutation is variable. Their metabolic crises can be easily provoked by non-predictable common disorders and sometimes may be fatal owing to genetic (private mutations and Lyonization) or external factors. Therefore, these patients may be unable to live a stable life in the long-term. The case presented in our report was of a female patient with OTCD with stable clinical course during pregnancy and immediately after delivery. Although other institutions have reported postpartum hyperammonemia crises in female patients with OTCD, we report a rare OTCD case of a patient who developed hyperammonemia with hepatic coma 6 days after delivery because we acquired her intact survival.

2. What point(s) was novel in this case?

(Author's response)

In female patients with OTCD, hyperammonemia crisis can develop 1 to 2 weeks after delivery, even if their clinical course was stable. Once the crisis occurs, it can persist for a relatively long time and can be fatal. Therefore, clinicians should aim to prevent it through hepatic coma, because it might be difficult to improve the patients' metabolic state. This point is novel in our study.

3. Why the authors do consider this case to be worth reporting?

(Author's response)

We reported a rare OTCD case and learned a lot from the clinical course of the disease in the patient. We want to report the knowledge acquired from this case and hope that the doctors caring for patients with OTCD can make use of this knowledge in their treatment strategies at delivery.

Minor 1. The authors used terms "postpartum" or "after delivery". These seemed misleading as newborn to me. I think other terms should be used for avoiding misleading: e.g. in a OTCD patient immediately after giving birth, etc.

(Author's response)

Thank you for your advice. Accordingly, we once have revised the title according as follows: "Hyperammonemia crisis in a female patient with ornithine transcarbamylase deficiency after giving birth." However, we are recommended to convert the title into "Hyperammonemia crisis following



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parturition in a female patient with ornithine transcarbamylase deficiency” by the native English speaker, and we changed “ after delivery” into “after parturition”.

2. The authors have reported their excellent outcomes of managing UCD patients without liver replacement. In “Introduction” section, I think they should show their know-how for their excellent outcomes. Almost all of descriptions in “Introduction” of the current form should be omitted and replaced by that.

(Author’s response)

Thank you for your advice. We have revised the Introduction according to your recommendations.

3. More extensively detailed description must be required in Case presentation. Causes the authors considered at that time and interventions for eradicating those causes must be presented in Case presentation, not in Discussion.

(Author’s response)

Thank you for your advice. We have added the cause and intervention about hyperammonemia crisis after giving birth in the Case presentation.

4. In “Discussion” section, it should be more extensively discussed whether this episode could be avoided with some preventive cares or not. Without that, lessons from this case will not benefit future cases.

(Author’s response)

Thank you for your advice. We have discussed, with descriptions, that this episode could be avoided with some preventive measures.

5. Extensive English editing must be done.

(Author’s response)

The initial submission had already been proofread, but we have resubmitted the revised manuscript to be checked for language, as you suggested.

In conclusion, We greatly thank the editor, two reviewers and Dr. Fang-Fang Ji for considering our manuscript.

Best regards

Jun Kido and Kimitoshi Nakamura