

February 4, 2014

Dear Editor,

Please find enclosed the edited manuscript in Word format (file name: 7047-review.doc).

**Title: ARC syndrome with high GGT cholestasis caused by VPS33B mutations**

**Author:** Jian-She Wang, Jing Zhao, Li-Ting Li

**Name of Journal:** *World Journal of Gastroenterology*

**ESPS Manuscript NO: 7047**

The manuscript has been improved according to the suggestions of reviewers:

1 Format has been updated

2 Revision has been made according to the suggestions of the reviewer

**(1) Response to Reviewer 00006675**

COMMENT-1 An interesting data is the parallel increase in serum levels of GGT and total bile acids. This should be described in Results section and commented in the Discussion.

Thanks for your valuable advice. Revision is made as suggested. Please see the blue sentences in the Results and Discussion sections.

COMMENT-2 Can the authors provide any possible speculation regarding the mechanistic link among the mutation, the dysfunction in bile formation and the elevation of serum GGT?

Revision is made as suggested. Please see the blue sentences in the Discussion section.

COMMENT-3 Improve consistence regarding the use of GGT or gGt

Thanks for your comment. Revision is made as you suggested.

**(2) Response to Reviewer 2860826**

COMMENT-1 In the abstract the authors illustrate only one case, but in the Cases reports they describe two cases. Please modified the “cases report” combining the two cases. I suppose that is more understandable if the second case is inserted into the medical history of the proband.

Thanks for your advice. Revision is made as suggested.

COMMENT-2 The authors use sometimes the term “patient” and sometimes “proband” . Please use only one of the two terms.

Thanks for your comment. We chose to use “proband” in our manuscript.

COMMENT-3 In the manuscript I find GGT, gGT and  $\gamma$  GT. Please use the first one (GGT), cited in the abstract.

Thanks for your comment. Revision is made as you suggested.

COMMENT-4 Page 4 paragraph 2: “ Epstein-Barr virus; blood cytomegalovirus DNA, and chromosome G bands.” Please correct in : Epstein-Barr virus, blood cytomegalovirus DNA and chromosome G bands.

Thanks for your correction. Revision is made accordingly.

COMMENT-5 Page 3, paragraph 1: detailed premiers. I suppose that was detailed primers.

Thanks for your correction. Revision is made accordingly.

COMMENT-6 The authors use “, and” : please remove “,” before “and” .

Thanks for your correction. Revision is made accordingly.

**(3) Response to Reviewer 02860576**

COMMENT-1 blood ceruloplasmin is not related to this age ,should be deleted.

Thanks for your comment. Revision is made as you suggested.

COMMENT-2 Ultrasound abdomen should be re written without repetitions.

Thanks for your comment. Revision is made as you suggested.

COMMENT-3 family history should follow history not physical examination and onvestigations.

Thanks for your comment. Revision is made as you suggested.

**(4) Response to Reviewer 2861186**

COMMENT: none

Thank you.

3 References and typesetting were corrected

Thank you again for publishing our manuscript in the *World Journal of Gastroenterology*.

Sincerely yours,



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