Dear Editor, Dec-05, 2019

Thank you very much for sending us the decision letter of 04-Dec-2019 for our manuscript number 52524, entitled "***OFD1* mutation induced renal failure and polycystic kidney disease in a childhood male twins in China**" by Hongwen Zhang, Baige Su, and Yong Yao.

We appreciate very much for the decision and comments. We have done our best to modify the manuscript according to the reviewer`s recommendations. We now resubmit our revised manuscript to your journal "World Journal of Clinical Cases" after we made a great effort to revise the manuscript. Thank you again.

In the revised manuscript all additions were marked in red. The point-by-point response to the referees is listed on pages 2.

Note: the page number may be mismatched because of differences in stationary page size (e.g. European A4 versus American "letter" size papers) and file transformation (e.g. Microsoft Word file is transformed into PDF file).

We would be grateful to your assistance with the English grammar if possible, though we have done our best to correct our typographical and grammatical mistakes.

We appreciate again for your response and effort, and look forward to hearing from you soon.

With the best wishes,

Sincerely Yours,

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Reply to reviewer's Comments:

Reviewer: 1

00503254

Conclusion: Accept (High priority)

Scientific Quality: Grade A (Excellent)

Language Quality: Grade B (Minor language polishing)

In this manuscript, the authors report rare cases of renal failure and polycystic kidney disease induced by OFD1 mutation in a childhood male twins. This case report is clinically interesting and useful. However, there is one point that needs to be addressed. Minor comment: There is a typing error. "Compliment" should be corrected to "Complement".

Answer: Thank you very much. We agree on you advices. We have corrected it. See line 18, page 6.

Reviewer: 2

00503228

Conclusion: Minor revision

Scientific Quality: Grade C (Good)

Language Quality: Grade B (Minor language polishing)

- The title isn't proper, since OFD1 mutation is supposed to make PKD and renal failure, but you better specify something specific in your cases. For example this particular missense mutation in exon 19 is not reported to my knowledge. So you may say instead "An exon 19 c.2524G>A (p. G842R) hemizygous mutation in OFD1 gene in 14 y/o Chinese male twins with PKD as the sole

disease presentation' or something –

Answer: Thank you very much. We agree on you advices. However, the c.2524G>A (p. G842R) mutation has been reported, and the pathogenicity also needed further studies, so we think the title is proper. See line 11-17, page 7.

Instead of using 'vs.' in giving data of the two patients better to say “&” also better to assign a number to each patient (p.1 & p.2) so a reader can distinguish differential results between the two cases.

Answer: Thank you very much. We agree on you advices. We have used “&” instead of “vs”, and we also assigned a number to each patient (p1 & p.2). See table 1, pages 11-12.

-Instead of listing all the laboratory tests in the text, you better give them in a table - Instead of stating 'no abnormal dysmorphic features' you should at least specifically name the most prevalent dismorphies like buccal, nasal, digital & intellectual problems (you may find a list in a series [doi: 10.1136/jmg.2004.027672]) –

Answer: Thank you very much. We agree on you advices. We have given the laboratory tests and dysmorphic features in table 1, thanks again. See table 1, pages 11-12.

Had they any family history of renal failure/PKD in their maternal side? - You specified that "The twins are monozygotic. ". Have you confirmed it or just due to their feature similarities; - In table 1, you specify your patients as elder and younger, but they were supposed to be monozygotic twins! - "they denied ... hematuria" You should confirm with laboratory tests.

Answer: Thank you very much. There was no family history of renal failure/PKD in their maternal side, we have added it, see line 2, page 6. The twins are monozygotic which was confirmed by the hospital where they birthed. We used patient 1 and 2 instead of elder and younger. They

denied ……, or gross hematuria, that is just presentations. We confirm with laboratory tests. See table 1, pages 11-12.

- Try to find disparities in the disease presentations between the two, it is very interesting to see if the same mutation in a monozygotic twin(?) might represent different features and at what extent? - Have you not related the ankle pain to their present illness? How both of them presented the ankle pain in such a short time interval? and what was its relation to their renal signs and symptoms (have you found their renal disease by routine check or there were any complaints. If so, be very precise in giving details in a time trail.

Answer: Thank you very much. We agree on you advices, it is very interesting to see if the same mutation in a monozygotic twin, but there were no different features between our twins. We added this. See lines 14-15, page 9. As for their ankle pain it might be related to osteoporosis due to renal failure. One of the twin test showed renal failure, and then the other. It was a pity that there were no any other complaints.