Dear Dr. Wang,

Thank you for the editors We would like to thank the editors and reviewers for their careful assessment of our article; their suggestions have strengthened the manuscript. And also we have revised the paper according to the review's comments.

Sincerely yours,

Xiaodong Sun

Response to editor's comments:

(1) Science editor:

This manuscript is well designed. The author give enough information of the cases and disscused in depth. Language Quality: Grade B (Minor language polishing)

Scientific Quality: Grade B (Very good)

Response: Thank you very much for your review with valuable suggestions. We greatly appreciate the time you have contributed to the review process.

(2) Company editor-in-chief:

I have reviewed the Peer-Review Report, the full text of the manuscript, and the relevant ethics documents, all of which have met the basic publishing requirements of the World Journal of Clinical Cases, and the manuscript is conditionally accepted. I have sent the manuscript to the author(s) for its revision according to the Peer-Review Report, Editorial Office's comments and the Criteria for Manuscript Revision by Authors. Before its final acceptance, the author(s) must provide the Signed Informed Consent Form(s) or Document(s) of treatment in Chinese. Please provide the original figure documents. Please prepare and arrange the figures using PowerPoint to ensure that all graphs or arrows or text portions can be reprocessed by the editor. Please upload the approved grant application form(s) or funding agency copy of any approval document(s).

Response: Thank you very much for your review with valuable suggestions. We greatly appreciate the time you have contributed to the review process. We have provided the Signed Informed Consent Form, the original figure documents and the approved grant application form.

Response to Reviewer's comments:

Reviewer #1: *In this manuscript, the authors presented an interesting case about HNF1A gene mutation in a 22-year-old MODY3 patient. The mutation resluted in a good repsonse to sulfonylurea therapy. Generally I think the topic is very interesting and the approach is attractive, but I simply could not follow how the authors framed and then conducted their analysis. In order to make the draft more illustrated, the authors should add some contents in the disscusion for how sulfonylurea benifit from the mutation in glycemic control.*

Response: Thank you very much for your review with valuable suggestions. We greatly appreciate the time you have contributed to the review process. We have added the frame and the contents in the discussion section for how sulfonylurea benefit from the mutation in glycemic control (line 118 and line 139-144).

Reviewer #2:

Q1. *Major comments: The medical history of the patient is not fully described, some information is missing. There are no data about the presence of glycosuria, as well about the length of the time the patient has been treated with metformin, saxagliptin and glargine.*

Response: Thank you very much for your review with valuable suggestions. We greatly appreciate the time you have contributed to the review process. We have added the

information in the TREATMENT section.

Urine tests were positive for glucose. After admission, the patient was treated with insulin glargine, and blood glucose was controlled within a week. Subsequently, we changed treatment with metformin (0.5g/d) and saxagliptin (5mg/d). Two months later, Hb1Ac was showed 5.5%. Meanwhile, we have achieved the genetic testing results.

Q2. *Some data is redundant unnecessarily, e.g. twice the HBA1c level at the beginning of the therapy, while the final effect after the inclusion of the SU in the therapy.*

Response: We have deleted redundant data. Additionally, we have added the final Hb1Ac results (Hb1Ac 6%).

Q3. *The dose of glibenclamide appears only in the discussion.*

Response: Thank you for the suggestion. We have added it in the TREATMENT section.

Q4. *The authors do not mention about the imaging test results.*

Response: We have added the Imaging examinations results. The ankle-brachial index and somatosensory potential findings were normal. Abdominal ultrasound showed fatty liver (light-medium) (line 114).

Q5. *The study lacks a description of the research methods. Only in Figure 1 there is a signature that this is Sanger sequencing.*

Response: We have added it.

Q6. *Minor comments: line 50 – has to be "replaced" instead of "rheplaced" Finally the paper is worth publishing after revision.*

Response: We have revised it.

Reviewer #3: *I studied the manuscript entitled "A novel HNF1A gene mutation in maturity-onset diabetes of the young" by Xu et al. It deals with a well-documented case of MODY3 attributed to a novel mutation. The information provided is of great interest to all specialists focusing on diabetes and, as such, deserves prioritized publication.*

Response: Thank you very much for your positive comments. We greatly appreciate the time you have contributed to the review process.

Reviewer #4: *It is a well-design study adding new information to the literature. Authors in a clear and simply way managed to give their results as well as the relative literature. I have no comments to make and in my opinion the article can be published unaltered.*

Response: Thank you very much for your positive comments. We greatly appreciate the time you have contributed to the review process.