

Dear Reviewer:

Thank you very much for your comments and professional advice. These Opinions help to improve academic rigor of our article. Based on your suggestions, I have made corrected modifications on the revised manuscript. Meanwhile, the manuscript had be reviewed and edited by language services of **LetPub**. I hope that our work can be improved again.

Furthermore, we have responded to the review comments one by one, as follows:

Answering Reviewer #1:

Suggestion 1: keywords: some are too generally (variant, hereditary, pathogenicity)

Answer 1: This is an excellent suggestion and I have modified these three keywords. According to the research topic, I have selected **"Rare disease"; "Genetic counseling"; "Assisted reproductive technique"** from the MeSH to replace them.

Suggestion 2: Background: Is a rare hereditary tomor. A rare hereditary neoplastic disorder is more correct.

Answer 2: Thanks for your suggestion, I have changed **"tomor"** to **"neoplastic disorder"** in the Background.

Suggestion 3: Line 62: 2436, please specify variants

Answer 3: Thanks for your suggestion, I checked the ClinVar database again and made updated statistics on the number of found variants. The variants here refers to the number of all the variations recorded in the database with uncertain significance, which has been explained in detail. The changes are as follows: **"A search for *STK11* gene variants in the ClinVar database revealed a total of 2481 entries with clinical significance, of which 974 (39.26%) were variants of uncertain significance (VUS), predominantly (79.98%, 779/974) missense variants until December 18, 2023".**



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Suggestion 4: Results: a tale with clinical features could be useful.

Answer 4: I have taken your suggestion and added a sentence in the results section, “The identification of the pathogenic mutations in these two clinically characterized PJS patients had helped these patients select the most appropriate mode of pregnancy assistance”.

Suggestion 5: Discussion: I suggest a separate paragraph for the limitations.

Answer 5: Thanks for your suggestion, I have adjusted the limitations of the article to a separate paragraph at the end of the discussion.

“Limitations

Several limitations should be noted in our study. First, because of the rarity of PJS, the number of patients included is relatively small, which may reduce the statistical analysis power. Second, the follow-up period was short; the longest follow-up was 8 years. Because the patients with PJS were generally young when they visited the doctor, no malignant tumors were found till the date of the final follow-up; thus, the correlation between PJS and cancer development could not be discussed. Third, because our hospital mainly caters to patients who need assisted reproduction and genetic counseling, there may be some bias in the selection of patients included; that is, patients visit the doctor for assisted reproduction, leading to a high proportion of patients choosing PGT”.

Thank you very much for your attention and time. Look forward to hearing from you.

Yours sincerely,

Jin Liu

29 Dec, 2023

Hunan Normal University