

Dear Dr. Ma:

We would like to express our sincere thanks to you and the reviewers for the constructive suggestions and positive comments, which we have made some changes accordingly.

Replies to Reviewer 1:

Specific comments

The study is merit, but I have minor concerns 1- Minor English polishing is required.

[Answer: Thank you for your kind reminder and we will revise the grammar.](#)

2- More information about CRB2 should be included in the introduction section.

[Answer: Thank you for your kind suggestion, we have modified accordingly.](#)

3- More references are required.

[Answer: We have modified accordingly.](#)

Replies to Reviewer 2:

Lu and colleagues report two siblings with steroid-resistant nephrotic syndrome where they identified two novel compound heterozygous mutations in CRB2. The sentence "In the research of primary NS, more attention is paid to detecting the genes" is misleading. The aim is to detect underlying mutations; genes will be present anyway.

[Answer: Thank you for pointing out this error, we have corrected the expression.](#)

"So far, more than 20 different single gene mutations such as NPHS1, NPHS2 and PLCE1 have been reported to cause SRNS". WT1 mutations should be mentioned (PMID: 18516627 and Refs. therein).

[Answer: Thank you for the recommended literature. Wilms' tumor suppressor gene WT1 is an important regulator in kidney development, and mutations in this gene have been associated with Wilms' tumor, Frasier syndrome, and Denys-Drash syndrome, as well as isolated glomerular disease. We have added this gene in our manuscript accordingly.](#)

In the discussion, some sentences regarding the potential function of CRB2 in kidney podocytes should be included.

[Answer: Thank you for your good suggestion, and we have modified accordingly.](#)

Best wishes,

Yours sincerely,

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