Author's response to reviews

Title: Ventricular septal defect in a child with Alport syndrome: a case report

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Author's response to reviews:

Dear Editor,

I am pleased to enclose a revised version of the manuscript entitled “Ventricular septal defect in a child with Alport syndrome: a case report”. The manuscript has been revised taking into account recommendations received from Reviewers. We trust that all issues raised have been taken care of adequately. Looking forward to receiving your decision and thanking you for your kind attention.

Yours sincerely

Pier Paolo Bassareo

Dear Colleague,

thank you for the time you have spent in reviewing our paper. Please find below our detailed response to your suggestions:

In our opinion, the novelties of this paper are:

- A) the previously unreported association between Alport syndrome and a congenital heart disease

- B) the possible pathophysiological mechanism underlying the concomitant manifestation of Alport syndrome and ventricular septal defect. However, since the first release of the manuscript, we have specified in the Discussion section (first major compulsory revision) that the hypothesizes mechanism is merely putative and further studies should be undertaken to better clarify the relationship between the two disorders. This will likely not be an easy task due to the rarity of Alport disease

- C) in our clinical experience with paediatric and adult patients undergoing dialysis and suffering from secondary drug-resistant hypertension, clonidine is a very safe, effective option in normalizing their blood pressure. Even when all the other drugs are ineffectual, the addition of clonidine to the previously established pharmacological therapy is able to control high blood pressure. In this respect, a reference has been added [26]. According to your suggestion, in the Discussion section we have added that clonidine is not a first line drug in most cases of
hypertension as well (second major compulsory revision)
- D) as you so rightly point out, surgery for VSD closure is frequently complicated by the development of atroventricular blocks. According to your suggestion, in the Discussion section we have specified that AS patients may have an additional risk in developing this surgical complication (third major compulsory revision)
- E) lastly, the present case report is of particular interest in view of the high degree of severity of AS in such a young female, as the more severe forms of X-linked AS are generally manifested in males, with a later onset of end-stage renal disease

Minor essential revisions:
- The paper has been translated by another expert native English speaker
- The references has been corrected. All of them have been placed according to the Guidelines for authors of this Journal

Trusting that our revised manuscript may now be found suitable for publication.

Yours sincerely
Pier Paolo Bassareo

PS
The revised parts in the manuscript are in red

Dear Colleague,

thank you for the time you have spent in reviewing our paper. Please find below our detailed response to your suggestions:
- “appears to be responsible of” has been replaced with “appears to be responsible for”
- “basement” has been replaced with “basal”
- “niphedipine” has been replaced with “nifedipine”
- the sentence “the patient was discharged …” has been clarified in the text
- “generale” has been corrected
- the sentence “ even if type IV collagen…” has been corrected according to your suggestion
- the sentence “ these other types, maybe…” has been corrected according to your suggestion
- “theraphy” has been replaced with “therapy”
- the Figure 1 legend has been corrected

Trusting that our revised manuscript may now be found suitable for publication.

Yours sincerely
Pier Paolo Bassareo
PS
The revised parts in the manuscript are in red. Other changes have been performed, according to the suggestions of the other Reviewer.