Reviewer's report

Title: The multiple systemic artery to pulmonary artery fistulas resulting in severe irreversible pulmonary arterial hypertension in patient with previous history of pneumothorax.

Version: 1 Date: 30 Nov 2018

Reviewer: Andrea Lombi

Reviewer's report:

I read very carefully this paper and I found it very interesting, because it will help to improve knowledges about a rare and difficult-to-approach pathology like pulmonary arterial hypertension.

I think that some minor reviews could enhance the quality of the manuscript.

First of all, I found the title too long and full of details that could be omitted in this context (making it more "catchy"), for example "irreversible" (related to pulmonary hypertension" and "and right-sided pneumothorax thirty five years ago".

Then, a complete review of written english shoud be performed: there are several minor imperfections that make the text less fluent (for example: "we describe A first case of...", "arterial pulmonary hypertension" (instead of pulmonary arterial hypertension), "was admitted to cardiology clinic due to exertional dyspnea and THE signs of.." and so on.

I suggest you to pay attention to acronyms and initials, they should be uniformed! For example, "Systemic artery to pulmonary artery fistulas" is abbreviated as "SA-PAFs", "SA-PVF", "SAPVF" and "SAPVS". Other example "angio-CT" and "angio CT" etc...

Third minor point is about RHC: PVR are always expressed as PVRI in dyna·cm·s^5/m^2, it could be more useful to express them in Wood Units (WU - accepted and described in the last PH guidelines); cardiac index could be an interesting value that may help to describe severity of PAH in this case. Last notice about RHC: why has vasoreactivity test been performed in this complex manner?

Last suggestion about conclusion: the fact that, after closing AV fistulas, patient condition went dramatically worse could be related to the loss of beneficial effect of right-left shunting with "decompress" of right ventricle, as it happens after closure of interatrial defects with R-L shunts.

In conclusion I approve this paper for pubblication, after this minor review.
Are the methods appropriate and well described?
If not, please specify what is required in your comments to the authors.

Yes

Does the work include the necessary controls?
If not, please specify which controls are required in your comments to the authors.

Unable to assess

Are the conclusions drawn adequately supported by the data shown?
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Yes

Are you able to assess any statistics in the manuscript or would you recommend an additional statistical review?
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