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: 2 Date: 07 Feb 2019

Reviewer: Rocco Rinaldo

Reviewer's report:

I read with interest the very complete authors' answer and the new version of the manuscript. The complexity of the story of the patient and the interpretation from the authors is now more clear, also mentioning in a more clear way the possible differential diagnosis.

Nevertheless, I found that two points have not been addressed:

- please, include the complete data from the second RHC, as you did for the first one (if there is not enough room for that you could remove the data from the combined sildenafil+NO, as the first test is already good for demonstrating the non-reversibility)

- given the series of rare events you hypothesize and the peculiar timing of the story, this single case report cannot be considered as a solid base to state your conclusion (which is pretty inclusive as many common conditions could possibly give a fistula). I, thus, suggest reconsidering the final conclusion, changing it, for example, in something like: "non congenital SA-PAFs are extremely rare, but still the diagnosis should be considered when a shunt is suspected during the diagnostic work up of pulmonary arterial hypertension, especially in patients with risk factors".

- In addition, related to the new version of the manuscript, you can remove the fact that a normal PWCP excludes PVOD, as the condition is usually pre-capillary, in terms of hemodynamics.
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.

Yes

Are the conclusions drawn adequately supported by the data shown?
If not, please explain in your comments to the authors.

No

Are you able to assess any statistics in the manuscript or would you recommend an additional statistical review?
If an additional statistical review is recommended, please specify what aspects require further assessment in your comments to the editors.

Not relevant to this manuscript

Quality of written English
Please indicate the quality of language in the manuscript:

Acceptable

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