Peer Review File
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Review Comments:
Reviewer A

General comments:
The authors showed two adult patients with unilateral absence of pulmonary artery (UAPA). Case 1, with severe pulmonary arterial hypertension, experienced massive hemoptysis related to UAPA. Despite treatment with medication, he died of cardiac arrest. Case 2 presented with asthma exacerbation, for whom medical therapy was effective. She successfully underwent a complex structural intervention. These cases are rare and include educational points. Also, the manuscript is well-described; however, several points should be addressed to improve the manuscript.

Specific comments:

1. Please describe how the authors treated massive hemoptysis in Case 1.
   1-1. Did it improve spontaneously?
   1-2. Did the contrast-enhanced CT show any findings related to massive hemoptysis?
   1-3. Was selective embolization considered a treatment option for massive hemoptysis in the case?

   Reply 1: Thank you for your thorough review and insightful comments that have helped enhance the quality and clarity of our manuscript. Regarding the management of massive hemoptysis in case 1. Initially, the patient was stabilized with fluid resuscitation, which was followed by a spontaneous cessation of the hemoptysis. No further episodes were observed while the patient was in the emergency room. Subsequent imaging, specifically contrast-enhanced CT, did not reveal any active bleeding sites. Despite the absence of identified bleeding on the CT scan, we proactively consulted with the interventional radiology (IR) team to assess the possibility and preparedness for embolization, should it become necessary. Fortunately, the patient's symptoms resolved spontaneously without the need for further invasive interventions, and there was no recurrence of hemoptysis.
   Changes in the Text: We have included this detailed account of the patient's management in response to your queries 1-1, 1-2, and 1-3 in the revised manuscript. (page 5, line 90-97)

2. Did the authors perform an echocardiogram to investigate the complication of other congenital cardiac defects in Case 1?

   Reply 2: Thank you for your query regarding the investigation of potential congenital cardiac defects in Case 1. A limited echocardiogram was indeed performed on the patient. The echocardiographic assessment revealed a dilated right ventricle with

reduced function. Importantly, this examination did not uncover any evidence of congenital cardiac defects. This finding helped to guide our clinical management and focus on addressing the primary issues identified without the added complexity of congenital cardiac anomalies.
Changes in text: We have updated the manuscript to include this information, providing clarity on the cardiac evaluation and its outcomes. (page 6, line 107-109)

3. What was the cause of cardiac arrest that led to death in Case 1?

Reply 3: Thank you for your question regarding the cause of the cardiac arrest that led to the unfortunate demise of the patient in Case 1. The patient experienced a progressive worsening of symptoms associated with right ventricular failure. This deterioration was accompanied by persistent signs of dysrhythmia, which ultimately culminated in a cardiac arrest. It is within this context of advancing right ventricular failure and dysrhythmia that the patient's condition led to a fatal cardiac arrest.
Changes in text: We have included this detailed explanation in the revised manuscript to provide a clear understanding of the sequence of events leading to the patient's death. (page 7, line 115-116)

4. In Case 2, Figure 3 included the finding of chest X-ray, which is duplicated in Figure 4. Please clarify that.

Reply 4: Thank you for pointing out the duplication of the chest X-ray image in Figures 3 and 4 for Case 2.
Changes in text: Figure 3 has been corrected.

5. Please add the findings of aortogram to Figure in Case 2.

Reply: Thank you for your suggestion to include the findings of the aortogram in the figures for Case 2. Unfortunately, we are unable to provide formatted images of the aortogram due to a change in the radiology reporting application, which has affected our access to suitable images for inclusion in the manuscript. We have, however, ensured that the textual description within the manuscript accurately reflects the aortogram findings to provide readers with a comprehensive understanding of the case despite the absence of these specific images. We apologize for any inconvenience this may cause and appreciate your understanding of the situation.

Reviewer B

The authors present an excellent case series with UAPA. The case presentations and outcomes are done well with with some minor modifications needed.

Major comments: None
Minor comments:
P2 L 56- Embolization should be the term used.

Reply to P2 L 56: Thank you for your recommendation to use the term "embolization" in our manuscript.
Changes in text: We have reviewed the document and made the necessary changes to reflect this terminology accurately. (page 4, line 59)

P3 L 93- Please explain poorly controlled COPD exacerbations

Reply to P3 L 93: Thank you for seeking clarification on the description of poorly controlled COPD exacerbations mentioned in our manuscript. The patient's medical history was significant for these exacerbations, primarily attributed to noncompliance with prescribed medications. This non-adherence to treatment regimens had led to multiple visits to the emergency room, indicating a lack of effective management of his COPD condition.
Changes in text: We have elaborated on this aspect in the revised manuscript to provide a clearer understanding of the patient's health background and the challenges in managing his COPD exacerbations. (page 6, lines 90-92)

P3 L 95- respiratory wheezing - inspiratory or expiratory?

Reply to P3 L 95: Thank you for your query regarding the nature of the respiratory wheezing described in our manuscript. We have clarified this detail within the text: the wheezing observed in the patient was expiratory.
Changes in text: This specification has been added to the manuscript to ensure a precise and comprehensive clinical description of the patient's condition. (page 6, line 94)

P4 L111- Why was the patient on Spironolactone- please explain.
Any other meds considered fro PAH?

Reply to P4 L111: Thank you for your question. The decision to initiate spironolactone was made to address the ongoing right ventricular (RV) failure and to optimize the patient's volume status. In addition to spironolactone, the patient was also started on ambrisentan, an endothelin receptor antagonist, specifically indicated for the treatment of PAH. This medication was chosen as part of a comprehensive management approach to address the underlying PAH pathology and optimize the patient's cardiovascular function.
Changes in text: We have included this explanation in the manuscript to provide clarity on the rationale behind the patient's medication regimen. (page 7, Line 113-

115)

L 129- "Inflated" bronchial A-sounds very layman and should be changed

Reply to L 129: Thank you for your feedback regarding the terminology used to describe the bronchial artery in our manuscript.
Changes in text: We have duly noted your suggestion and revised the text accordingly. The term "inflated" has been replaced with "hypertrophied" to better reflect the medical terminology and ensure clarity in the description of the bronchial artery. (page 8, Line 139)