

Peer Review File

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**Round 1**

**Reviewer A**

Comment 1 - Can the authors specify at what age the diagnosis of lung agenesis was made in the present patient?

Reply/Changes in Text: The patient reported that he was given this diagnosis in his around age 6.

Changes in Text: "He reported first receiving a diagnosis of right lung underdevelopment when he was six years old." Page 3/line 17-18

Comment 2 - Can the authors argue for the use of theophylline in this patient?

Reply: The patient had been on theophylline for at least 18 years prior to his pneumonectomy. Based on our chart review, due to patient preference and gradual (as opposed to rapid) progression of his respiratory status, the medication was started and continued by his pulmonology team. He is no longer on this medication. We presume that theophylline was started given its historically higher use, but since this cannot be confirmed, we have elected to not include this supposition in the manuscript.

Comment 3 - How long in total has the patient been treated with voriconazole for the past 10 years?

Reply/Changes in Text: The patient received voriconazole for a total of 46 months (all not consecutively as noted earlier in the transcript) during this 10 year period. We have included this information in Page 4 line 7-8

Comment 4 - How many times has the patient been treated for bacterial pneumonia or acute bronchitis (number of episodes/year) in the last 10 years?

Reply: The patient received treatment for pneumonia or acute bronchitis at least 16 times in the last 15 years. The antibiotic courses tended to be longer durations (10-14 days as opposed to 5-7 days). There did not appear to be a pattern regarding these episodes (ie. No significant increase in frequency in the more recent years).

Changes in text: "Additionally, he was empirically treated for bacterial pneumonia or acute bronchitis at least 16 times over a 15-year period in the setting of worsening cough

and intermittent pleurisy.” Page 4 line 10-13

Comment 5 - Has the patient experienced hemoptysis? If so, how many episodes have been recorded?

Reply: The patient experienced mild hemoptysis once, just prior to starting his first course of voriconazole. He did not have recurrence of hemoptysis.

Changes in text: “He experienced mild hemoptysis prior to starting his first course of voriconazole, though did not have any recurrence of this symptom later in life.” Page 4 Line 4-6

Comment 6 - What is the follow-up from the pneumonectomy?

Reply: The patient has done well postoperatively. We have included additional details below to expand on his follow up.

Changes in text: “He had an uneventful postoperative course and is currently doing well, approaching one year from his surgery. He is using albuterol as needed for symptom control and is gradually increasing his exercise tolerance. Serial follow up imaging has shown a well aerated left lung.” Page 5, line 1-4

Comment 7 - Discussion: In the last paragraph, the authors should specify the risks associated with pneumonectomy, particularly on the right side and in the elderly patient, which should be taken into account in the decision.

Reply: We agree that discussion of the particular risks is important to include. We have done so as indicated below with additional references.

Changes in text: “While older age is generally thought to be a risk factor for post-operative morbidity and mortality, studies have suggested that age itself should not be a contraindication for pneumonectomy and instead should be one component of a larger, comprehensive risk-assessment approach (11-12). Additionally, there may be higher rates of perioperative complications for right-sided versus left-sided pneumonectomies, including but not limited to bronchopleural fistulas and pulmonary edema, though further investigation is needed to clarify true differences in outcomes between the two procedures (13).” – Page 7, line 4-11

Comment 8 - Figure 1: The authors should provide multiple CT views.

Reply: We appreciate this feedback and have added another view to the figure.

Changes in text: Figure 1

**Reviewer B**

This paper describes a 72-year-old male with recurrent aspergillus right lung infections who underwent right pneumonectomy as a therapeutic treatment for the fungal infection. In different sections of the paper the condition is described as unilateral lung agenesis, pulmonary agenesis and bronchial atresia and in another area congenital atresia of the right lung and pulmonary artery with diffuse bronchial atresia and peripherally distributed cystic lung. The condition described does not fit the definition of pulmonary agenesis.

I do not think this case report would add to the medical literature.

Reply: We appreciate this feedback and realize there was need to clarify both the patient's lung disorder and the educational value of this case report. Through our further review of congenital lung disorders, we have decided that this patient's lung disease is more accurately described as hypoplasia as opposed to agenesis. This change has been made throughout the manuscript. We believe the rarity of this condition presenting well into adult remains notable. Moreover, we have expanded upon the discussion section to include considerations for pneumonectomies in this patient population. We intend for this report to guide expectations for both surgical and non-surgical specialties alike in the management of similar patients.

**Round 1**

**Review comments:**

We thank the authors for their revised manuscript. We have no additional comments.