

A 28-year-old Female with Multiloculated Pneumothorax Secondary to Lymphangioliomyomatosis

Trevor Gauthier, M.D., James Onigkeit, M.D.
Department of Anesthesiology and Perioperative Medicine, Mayo Clinic, Rochester, MN

BACKGROUND

- Lymphangioliomyomatosis (LAM) is a rare, slowly progressive neoplasm of unknown origin that causes cystic lung disease, chylous pleural effusions, and abdominal angiomyolipomas
- The most common presenting symptoms are progressive dyspnea on exertion secondary to cystic lung disease or acute dyspnea secondary to spontaneous pneumothorax
- mTOR inhibitors are the mainstay of treatment for LAM, however if symptoms progress lung transplantation can be considered
- Chest Xray is limited in detection of pneumothorax compared to CT scan, with a sensitivity of 40% and a specificity of 96%

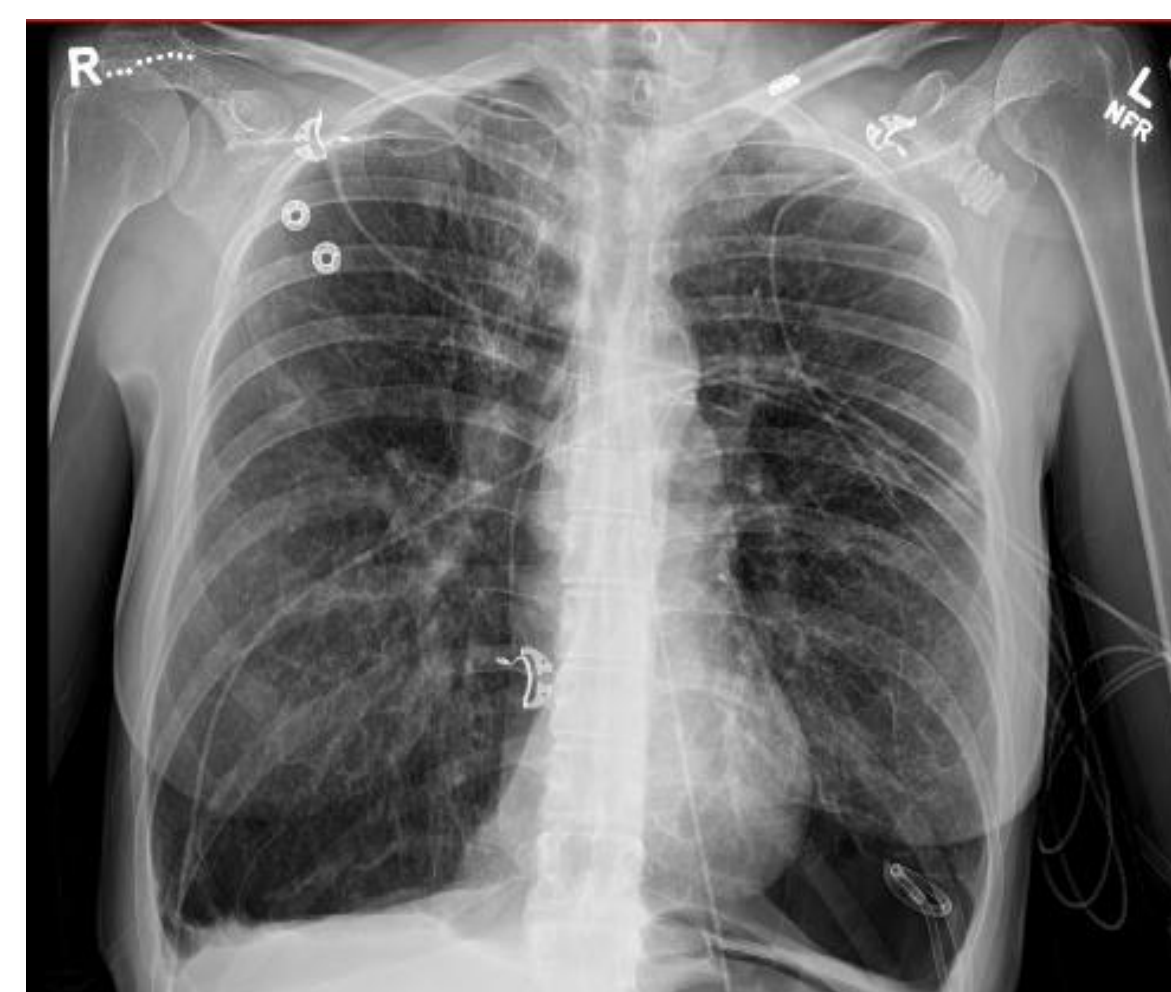
CASE PRESENTATION

Chief Complaint

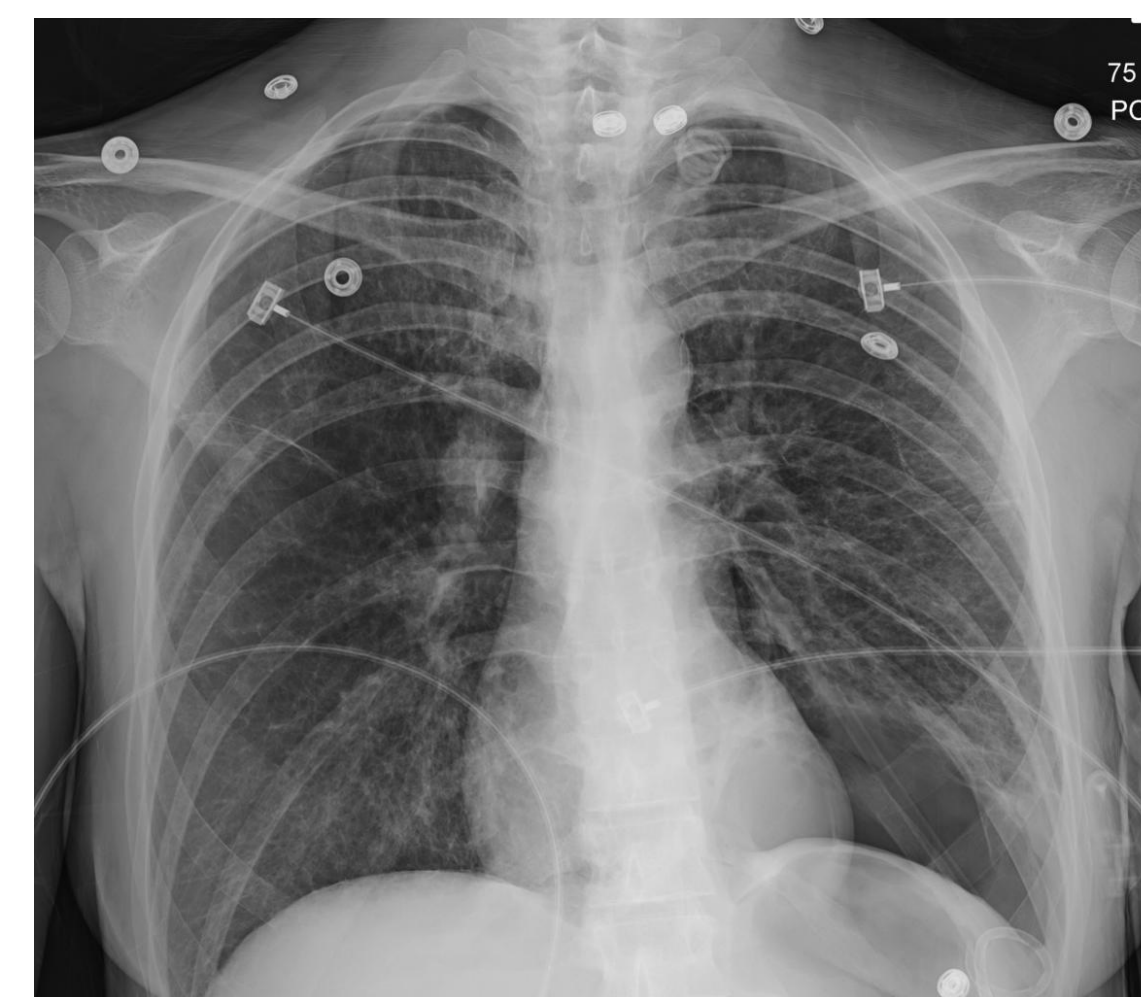
- 28-year-old female transferred to ICU due to dyspnea and hypoxemia

HPI

- Admitted for a left pneumothorax s/p endobronchial valve placement and subsequent removal



CXR obtained after endobronchial valve removal



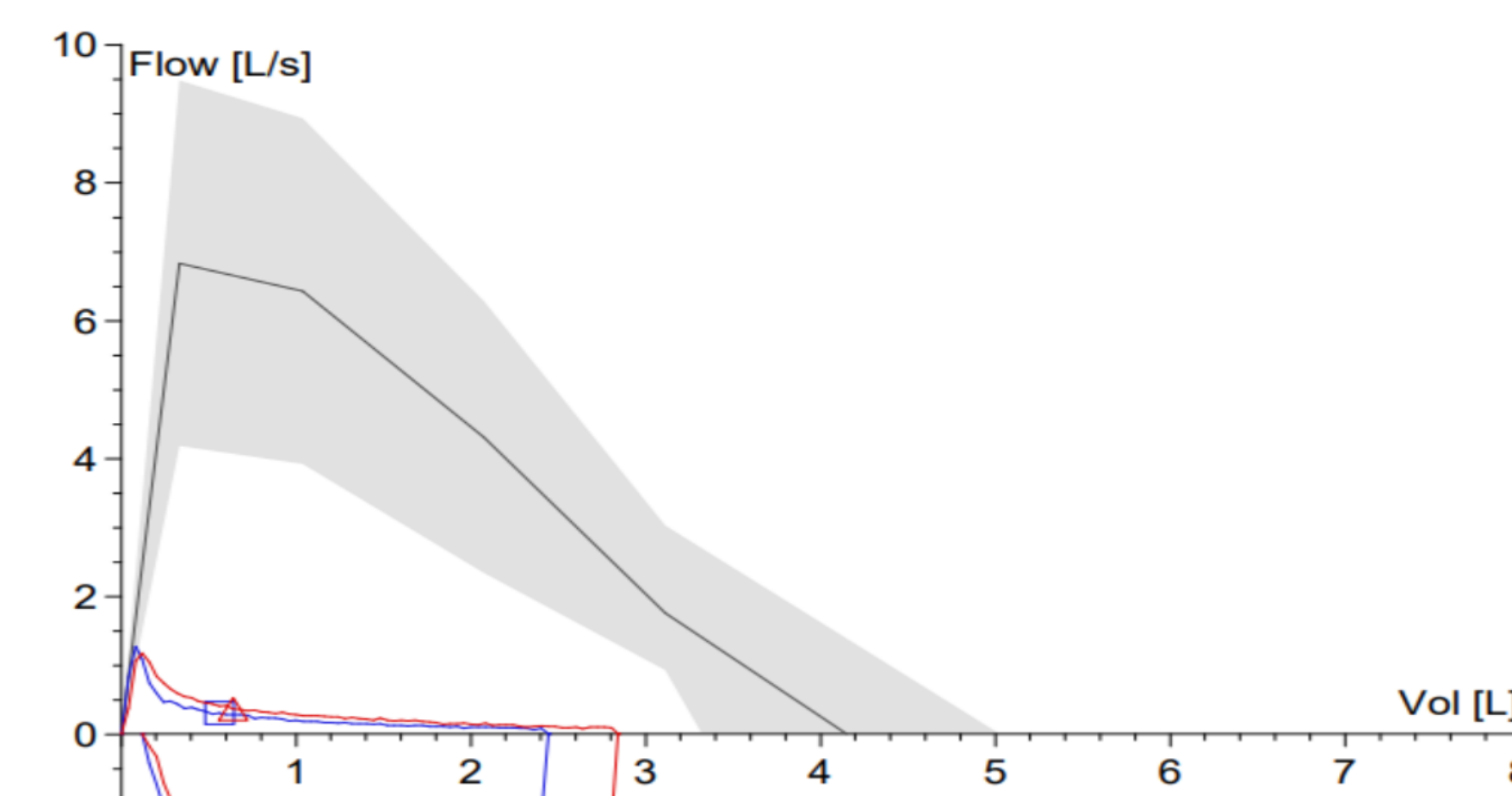
CXR obtained upon ICU transfer

Pertinent PMH

- Initial spontaneous pneumothorax in 2023 at age 27
- Recurrent pneumothorax in early 2024, s/p bilateral VATS with talc pleurodesis
- Diagnosed with LAM and initiated on sirolimus 1 g daily



CT chest of patient prior to admission, demonstrating diffuse cystic lung disease

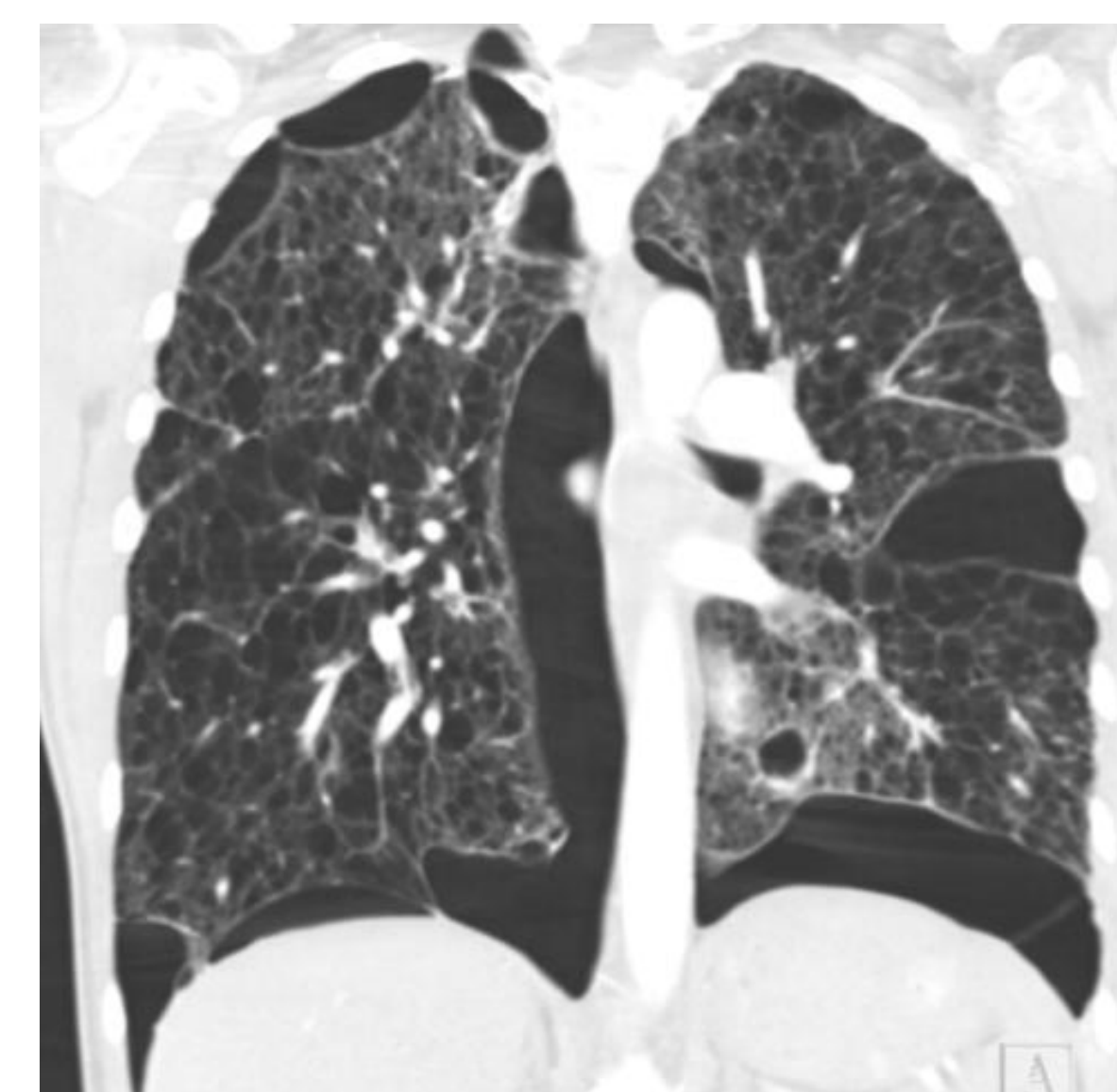


Patient flow/volume curve demonstrating severe obstructive lung disease

ICU Course

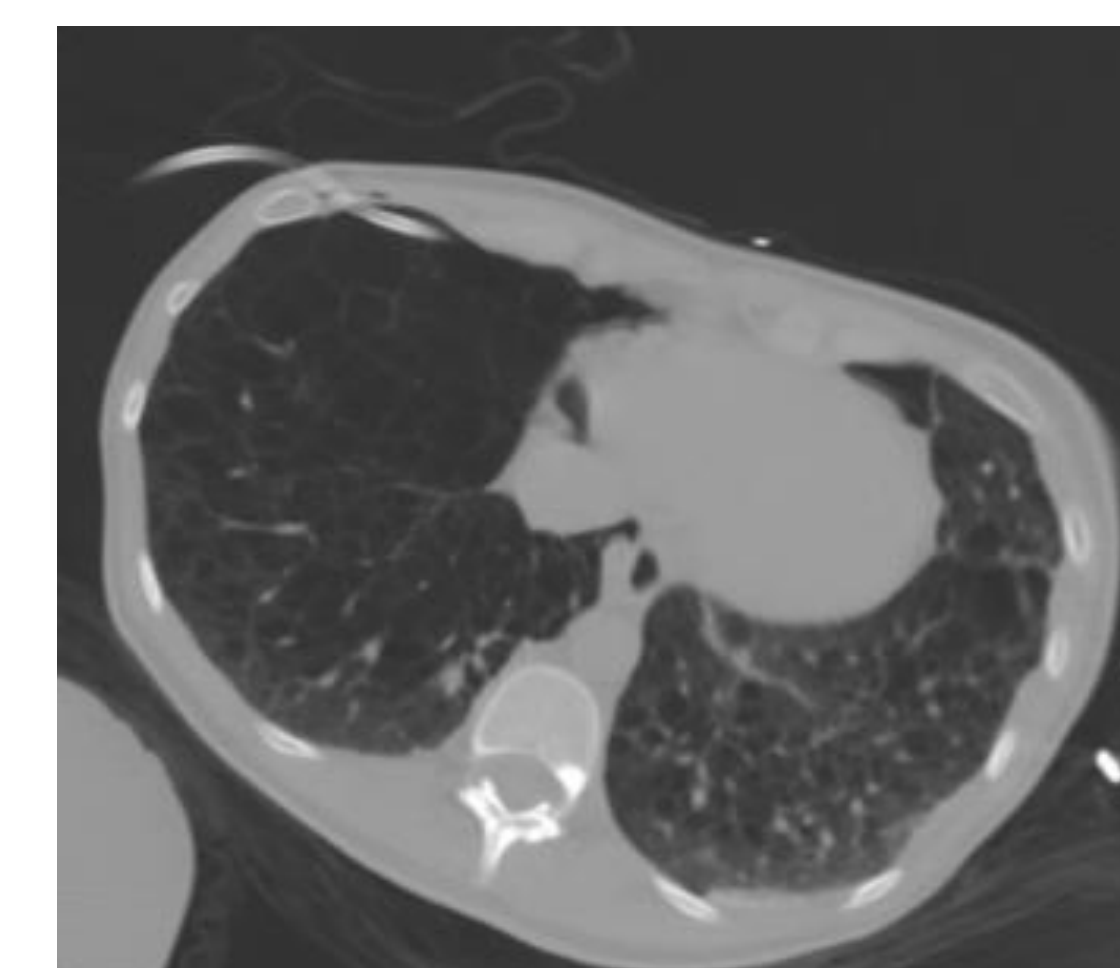
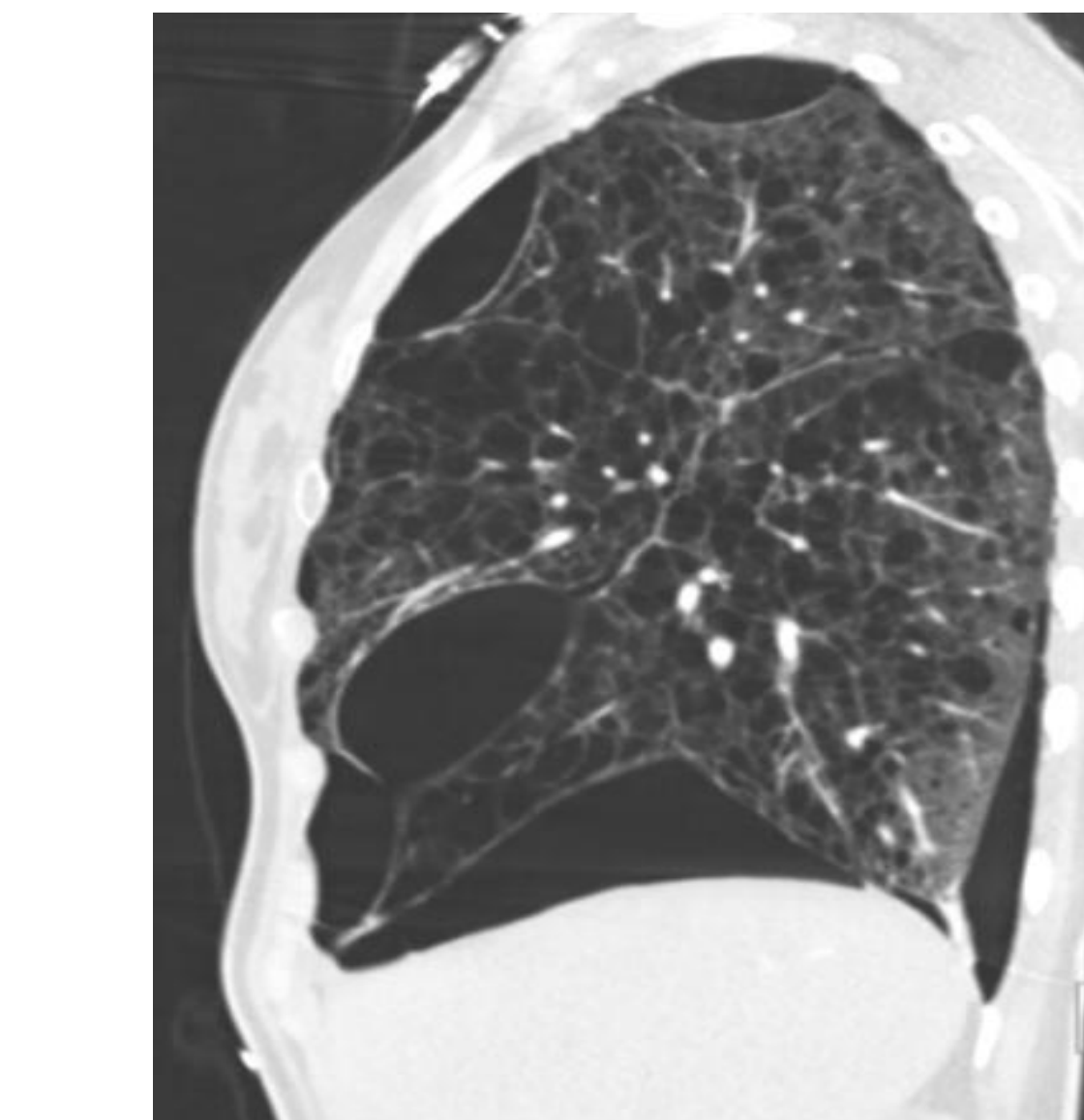
ABG: pH 7.24, PaCO₂ 70 mmHg, HCO₃ 29 mEq/L

Initial Management: HFNC at 45 L/min, FiO₂ 60% achieving SpO₂ 97%; STAT Chest CTA



Case Resolution

- CT-guided placement of chest tubes
- Discharged with Heimlich valves on chest tubes
- Bilateral lung transplantation two months later



KEY POINTS

- Multiloculated pneumothorax is rare
- CT is superior to chest film for determining size and cause of pneumothorax
- If chest Xray is negative for pneumothorax but suspicion is still high, chest CT should be obtained

REFERENCES

- Elia, D. et al. (2023). Presse Med. 2023;52(3):104173.
- Expert Panel on Thoracic Imaging, Laroia AT, Donnelly EF, et al. ACR Appropriateness Criteria® Intensive Care Unit Patients. *J Am Coll Radiol.* 2021;18(5S):S62-S72. doi:10.1016/j.jacr.2021.01.017
- McCarthy, C. et al. (2021). Lancet Respir Med. 9(11):1313-1327.
- Winkler MH, Touw HR, van de Ven PM, Twisk J, Tuinman PR. Diagnostic Accuracy of Chest Radiograph, and When Concomitantly Studied Lung Ultrasound, in Critically Ill Patients With Respiratory Symptoms: A Systematic Review and Meta-Analysis. *Crit Care Med.* 2018;46(7):e707-e714. doi:10.1097/CCM.0000000000003129