



# A Case of Swyer–James–MacLeod Syndrome Mimicking Chronic Thromboembolic Pulmonary Hypertension in a 77-Year-Old Woman

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## ABSTRACT

Swyer–James–MacLeod syndrome (SJMS) is a rare condition, resulting from post-infectious obliterative bronchiolitis, usually diagnosed in childhood. We present a 77-year-old woman with a history of childhood respiratory infections, who developed progressive dyspnea and was referred to a specialized pulmonary hypertension (PH) clinic with suspected chronic thromboembolic pulmonary hypertension. (CTEPH). Echocardiography revealed elevated right ventricular systolic pressure, and a ventilation–perfusion (V/Q) scan showed multiple perfusion defects. However, CT pulmonary angiography excluded thromboembolic disease, instead demonstrating extensive unilateral bronchovascular depletion of the left lung, consistent with SJMS. The patient was diagnosed with SJMS complicated by PH, managed with long-term oxygen therapy and supportive treatment.

## CASE PRESENTATION

- 77-year-old woman with a six-month history of progressively worsening exertional dyspnea, who was on long-term oxygen therapy at 2.5 L/min for a few months
- No concomitant symptoms such as cough, chest pain, hemoptysis, or systemic symptoms
- Medical history: atrial fibrillation, arterial hypertension, dyslipidemia, type 2 diabetes mellitus, epilepsy.
- No smoker, previously worked in the textile industry
- History of respiratory tract infections in childhood.

### Work up:

- Echo: right atrial and right ventricular enlargement, TAPSE 1.7 cm, RVSP ~65 mmHg, moderate tricuspid regurgitation
- LFTS (Table 1): severe obstructive disorder with normal diffusion capacity
- CXR (Figure 1): left lung hyperlucency
- V/Q scan (Figure 2): multiple segmental and subsegmental perfusion defects, particularly in the left lung., without ventilation mismatch.
- CTPA (Figure 3): No intraluminal filling defects down to the subsegmental branches of the pulmonary arteries. Extensive bronchovascular depletion in the left lower lobe, lingula, and apicoposterior segment of the left upper lobe, with threadlike opacification of segmental and absence of subsegmental arterial branches. Bronchiectasis in the left lower lobe.

The overall clinical picture in association with the imaging findings was consistent with the diagnosis of Swyer–James–MacLeod syndrome.

### Management:

- long-term oxygen therapy
- optimization of comorbidities
- pulmonary rehabilitation

The patient remains under close follow-up with a stable functional and hemodynamic profile on supportive therapy so far

## DISCUSSION

- Swyer–James–MacLeod syndrome (SJMS) is also referred to as unilateral hyperlucent lung syndrome
- It is characterized by reduced pulmonary vascularity and regional hyperinflation, which may involve an entire lung or a single lobe
- May occur with associated bronchiectasis
- **Pathogenesis:**
  - It is considered a sequela of post-infectious bronchiolitis obliterans
  - Associated pathogens: adenovirus, measles, Bordetella pertussis, Mycoplasma pneumoniae, tuberculosis, respiratory syncytial virus, influenza A
- **Differential diagnosis:**
  - congenital lobar emphysema (CLE)
  - unilateral pulmonary artery agenesis
  - Pneumothorax
  - CTEPH

### Pulmonary function tests

FVC	1320 mL (56% of predicted)
FEV1	780 mL (43% of predicted)
FEV1/FVC	59.4%
FEF 25–75%	220mL
DLCO	87%
TLC	4390 mL (100%)
RV/TLC	149%

Table 1

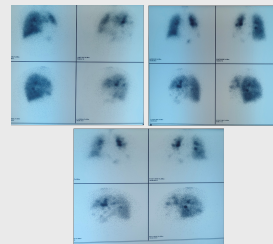


Figure 2

### Clinical picture:

- Many remain asymptomatic
- Dyspnea on exertion, cough, hemoptysis, frequent respiratory infections

### Diagnosis:

- Pulmonary function tests: usually airflow obstruction, may demonstrate mixed pattern
- CXR: different radiolucency between the two lungs (or lobes), more prominent on expiration
- CTPA or HRCT with contrast: small areas of normal tissue within generally hyperlucent regions, emphysema, hypoplastic pulmonary arteries, diminished peripheral vasculature, bronchiectasis
- Ventilation–Perfusion scan: matched defects in the affected lung



Figure 1

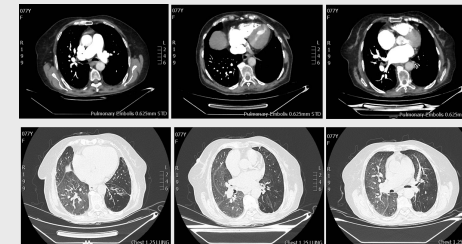


Figure 3

### Complications:

- Recurrent pulmonary infections (particularly in the presence of bronchiectasis)
- Lung abscess formation
- Spontaneous pneumothorax

### Management:

1. Conservative:
  - O2 therapy
  - Corticosteroids and inhaled bronchodilators
  - Chest physiotherapy, pulmonary rehabilitation
  - Pneumococcal and influenza vaccination
2. Surgical:
  - in cases of bronchiectasis and recurrent pulmonary infections or multiple episodes of pneumothorax
  - pneumonectomy, lobectomy or segmentectomy
  - occlusion of the main bronchus while the lung is left in place

## CONCLUSIONS

- The overlap in imaging features with more common conditions, particularly chronic thromboembolic pulmonary hypertension, can complicate the diagnostic process and potentially lead to misinterpretation.
- Careful integration of clinical history, advanced imaging modalities, and recognition of characteristic radiologic patterns is essential.

## REFERENCES

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