


## Swyer–James–MacLeod syndrome—a rare diagnosis presented through two adult patients

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### Keywords

Air-trapping, emphysema, hyperlucent lung, obliterative bronchiolitis, pruned-tree appearance.

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### Abstract

Swyer–James–MacLeod syndrome (SJMS) is a rare syndrome of acute obliterative bronchiolitis following an early childhood infective insult to the lungs. This causes arrest of alveolarization, affecting lung development with hypoplasia of the ipsilateral pulmonary artery and results in a characteristic radiological pattern, such as a unilateral hyperlucent lung with expiratory air-trapping and pruned-tree appearance on pulmonary angiogram. The clinical presentation is either recurrent chest infections, exertional dyspnoea or it may be an incidental finding. Management involves early prevention of infection, airway clearance, and regular vaccinations.

We describe two adult patients with SJMS: A 51-year-old female of Indian ethnicity presenting with recurrent haemoptysis and a 40-year-old Indigenous male presenting acutely with sepsis and background history of recurrent chest infections.

These cases highlight the importance of being aware of and accurately recognizing this rare condition, to be able to manage patients appropriately and avoid incorrect and unnecessary treatment.

### Introduction

Swyer–James–MacLeod syndrome (SJMS) is an acquired rare clinical-pathologic entity resulting from an early childhood infective insult that results in post-infectious acute obliterative bronchiolitis with airflow limitation and concomitant vasculitis [1,2]. This causes arrest of alveolarization leading to hypoplasia of the affected lung (cut-off at fourth to fifth generation). The affected lung will be smaller than its counterpart along with evidence of air-trapping resulting in unilateral hyperlucency. There is associated reduced vascularity with hypoplasia of ipsilateral peripheral pulmonary vessels [1].

Swyer–James–MacLeod syndrome is usually found in childhood with clinical presentation of recurrent chest

infections but can be an incidental finding in adults. The prevalence is estimated to be 0.01% based on a survey of 17,450 chest radiographs [3]. It is often misdiagnosed as chronic obstructive pulmonary disease (COPD), asthma, pneumothorax, or pulmonary embolism due to similar presentations.

We describe herewith SJMS in two adult patients with diverse clinical presentations.

### Case Report

#### Case 1

A 50-year-old female of Indian ethnicity presented following recurrent haemoptysis. Her past history included an

episode of possible severe influenza at four years of age. She had been treated previously for suspected pulmonary tuberculosis without any significant improvement of her haemoptysis. She did not smoke or drink alcohol. Respiratory examination revealed reduced air entry on the left side. Her chest radiograph performed showed a small hyperlucent left hemithorax. The high-resolution computed tomography (HRCT) showed diffuse decrease in attenuation of the left hemithorax and expiratory air-trapping (Fig. 1A, B). The sputum examination was negative for acid-fast bacilli. A magnetic resonance angiogram was performed for the evaluation of pulmonary artery and its branches and revealed typical pruned-tree appearance on the left side (Fig. 1C). Due to the clinical presentation and radiological appearance, the diagnosis of SJMS was made.

## Case 2

An Australian Indigenous 40-year-old male presented acutely to the emergency department with cough, fever, and burning micturition. Respiratory examination showed reduced air entry on the left side and abdominal examination revealed suprapubic tenderness. He was concurrently diagnosed to have both left lower zone pneumonia and a prostatic abscess. The blood culture and abscess aspiration revealed *Staphylococcus aureus* and he was treated accordingly with vancomycin. However, he also described a history of recurrent chest infections. Given history of smoking, his symptoms were attributed to infective exacerbation of COPD, for which he had received treatment in the past. Further evaluation included a Pulmonary Function Test (PFT) which suggested a mixed obstructive–restrictive pattern with a reduced Forced Expiratory Volume in 1 second / Forced Vital Capacity (FEV1/FVC) ratio and reduced Total Lung Capacity (TLC). Chest radiograph showed a hyperlucent left hemithorax with ipsilateral small hila and

expiratory air-trapping (Fig. 2A). Hyperinflation of the right lung was also noted. A CT pulmonary angiogram (CTPA) (Fig. 2B) showed loss of pulmonary vasculature and a ventilation–perfusion (V/Q) scan (Fig. 2C) demonstrated matched V/Q defects. The above clinical presentation and investigations confirmed the diagnosis of SJMS.

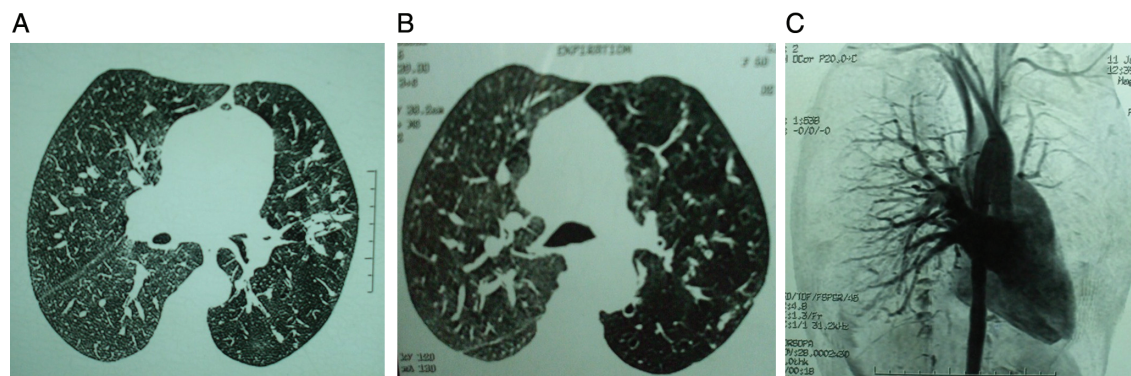
## Discussion

Swyer–James–MacLeod syndrome, also called unilateral hyperlucent lung syndrome is a rare complex lung disorder initially described by English physician William Macleod [4], and simultaneously by Canadian physician Paul Swyer and radiologist George James in the 1950s [5].

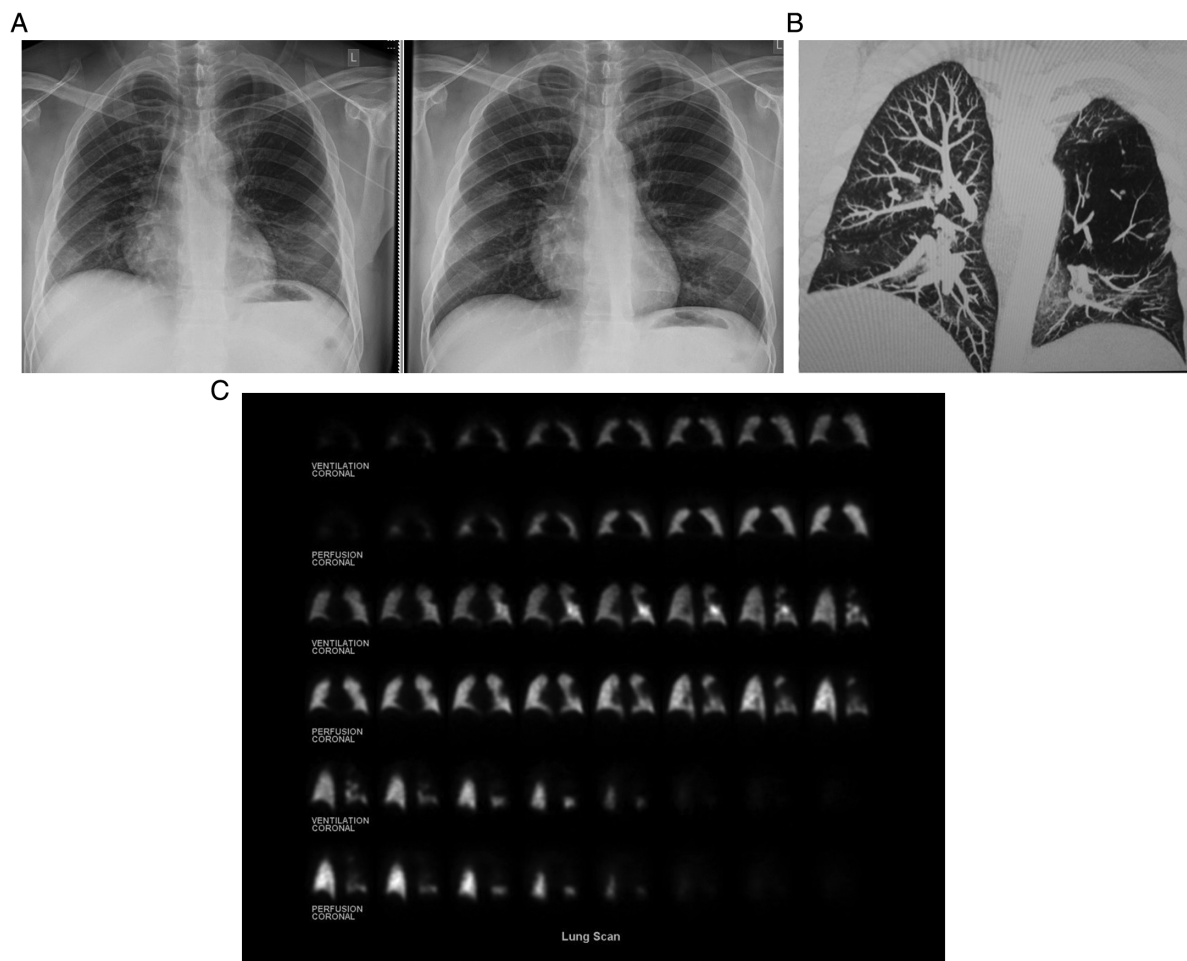
Swyer–James–MacLeod syndrome is currently considered to be an acquired condition due to infectious insults occurring during infancy or childhood. The agents, which are implicated, include adenovirus types 3, 7, and 21 (most commonly implicated), paramyxovirus, *Bordetella pertussis*, mycobacterium, mycoplasma, influenza, *Streptococcus pneumoniae*, and *S. aureus* [1]. The infective insult is thought to cause acute obliterative bronchiolitis with resultant arrested growth of lung and reduced vascularity.

Bronchoalveolar lavage (BAL) in SJMS reveals striking neutrophilia and slight lymphocytosis with increased CD8 to CD4 ratio suggesting that the damage seen is an active inflammatory process [2]. There is hypoperfusion due to pulmonary artery hypoplasia, which may be the result of concomitant vasculitis and reflex vasoconstriction of pulmonary vessels. These may be protective mechanisms to reduce V/Q mismatch [3,6].

All these pathologic changes result in altered lung dynamics with abnormal time attenuation curves and expiratory air-trapping with resultant bronchial and bronchiolar abnormalities. The affected lung does not develop properly and is smaller than its counterpart with evidence of air-trapping resulting in unilateral hyperlucency.



**Figure 1.** (A, B) High-resolution CT thorax of case 1 demonstrating air trapping on expiration. (C) Magnetic resonance angiogram (MRA) of case 1 showing pruned-tree appearance of pulmonary artery on the left side.



**Figure 2.** (A) Chest radiograph images showing a hyperlucent left hemithorax with ipsilateral small hila and expiratory air-trapping. (B) CT pulmonary angiogram (CTPA) of case 2 showing loss of the pulmonary vasculature on left side. (C) Ventilation–perfusion (V/Q) scan of case 2 demonstrating matched V/Q defects on the left side.

Swyer–James–MacLeod syndrome is quite a heterogeneous entity clinically. Majority of the cases are diagnosed in childhood, with usual presentation being recurrent bouts of chest infection [7]. The less common clinical presentations include productive cough, wheezing, exertional dyspnoea, and reduced exercise tolerance. Haemoptysis as seen in one of our patients is an extremely rare presentation.

Sequelae of SJMS include bronchiectasis, which can be present in up to 78% cases [8], lung abscesses, and spontaneous pneumothorax [9]. The clinical manifestation and prognosis is affected by the presence and type of bronchiectasis, with worst prognosis associated with saccular form [6]. Those with little or no associated bronchiectasis, have minor symptoms with diagnosis often being incidentally made on chest radiography.

Important differential diagnosis of SJMS include congenital lobar emphysema, pulmonary hypoplasia, bullous lung disease, cystic lung disease, post lobectomy compensatory emphysema, pulmonary embolic disease, pneumothorax, and foreign body in the airway [3,6,10].

There have been situations where SJMS has been misdiagnosed resulting in inappropriate management. The patients are often misdiagnosed as having COPD or asthma as demonstrated by our second case. He was treated with several courses of steroids for recurrent exacerbations, which could potentially increase the risk of infection. There are other cases where patients were misdiagnosed as having a pneumothorax and received multiple chest drains [10]. Moreover, one of our patients had received unnecessary anti-tuberculosis treatment previously.

The diagnosis of SJMS is made on radiological findings. Radiological modalities usually performed include chest radiograph, HRCT, CT angiography, magnetic resonance angiography, angiography, and V/Q scanning.

A classical triad has been described which confirms the diagnosis [11]:

1. Chest X-ray and HRCT thorax demonstrating unilateral hyperlucent lung with small ipsilateral hila and pulmonary artery with air-trapping on expiration. Expiratory air trapping is sine-qua-non for making the diagnosis, and is a reflection of airway obstruction.
2. Diffuse decrease in ventilation of the affected lung.
3. Matching decreased perfusion of the affected lung identified as ipsilateral pruned-tree appearance on angiography or matched defects on V/Q scan [12].

high-resolution computed tomography thorax with thin collimation in both phases of respiration is required to demonstrate air trapping. Prone position may be required for demonstration of mosaic pattern. Paucity of bronchial subdivisions and proximal bronchiectasis may be present [13]. Marked air-trapping may be noted in the washout phase of the V/Q scan [12]. Angiography helps confirm the diagnosis by demonstrating smaller pulmonary artery and its branches on the affected side. The narrowed attenuated arteries coursing through the radiolucent lung will produce a “pruned-tree” appearance. Invasive investigations such as thoracoscopy is generally not required for diagnosis but if done will show hypovascular lung parenchyma and multiple sub-pleural air-trapping [14].

PFT, if performed, will show a mixed obstructive–restrictive ventilatory defect [1,7], as was seen in one of our patients.

The treatment of SJMS is often conservative, and usually involves managing current and preventing future pulmonary infections. Airway clearance should be emphasized especially if there is concomitant bronchiectasis. Influenza and pneumococcal vaccines as per protocol should be administered. Surgical procedures, such as pneumonectomy or lobectomy, are only indicated in patients who suffer from recurrent infections due to affected lung segments [11].

Swyer–James–MacLeod syndrome is considered to be a small airways disease with fixed airflow limitation. Therefore, inhaled bronchodilators and corticosteroids are not usually recommended as therapy options. However, recently it has been suggested that combination inhaler therapy with inhaled corticosteroids/long acting antimuscarinic agents plus long acting beta-agonists may have a role in selected situations [15], but further studies are needed.

In conclusion, SJMS is a post-infective obliterative bronchiolitis with a variable clinical course and prognosis influenced by the presence of underlying bronchiectasis. The syndrome should be suspected in cases of presumed pulmonary emphysema with an atypical distribution, pneumothorax, or stable patients with unilateral hyperlucency. A chest X-ray may underestimate the presence of SJMS syndrome and further imaging should be considered. Swyer–James–MacLeod syndrome should be considered in the differential and diagnosed early, especially in patients not responding to conventional treatment to achieve better prognosis and also to prevent inappropriate treatment.

### Disclosure Statements

No conflict of interest declared.

Appropriate written informed consent was obtained for publication of this case series and accompanying images.

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