

## Case report - Thoracic non-oncologic

# Swyer–James–MacLeod syndrome; repeated chest drainages in a patient misdiagnosed with pneumothorax

Abdulrazzaq Sulaiman\*, Alaric Cavaille, Brigitte Vaunois, Olivier Tiffet

*Unité de Chirurgie Général et Thoracique, Hôpital Nord, Av. Albert Raimond, CHU de St. Etienne, 42055 St. Etienne, France*

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### Abstract

Swyer–James–MacLeod Syndrome (SJMS) occurs as a result of childhood bronchiolitis obliterans. Typically, this disorder is diagnosed in childhood after evaluations for recurrent respiratory infections. One of the reasons to explain the difficulty in diagnosis is that when patients develop little bronchiectasis, and therefore, few symptoms, then this syndrome may not be recognized until adulthood. Here, we are presenting a 22-year-old female patient who was diagnosed with SJMS who was initially misdiagnosed with a pneumothorax and treated by multiple chest tube drainages. This case highlights the significance of taking a careful history, the application of computed tomography and scintigraphy in confirming the diagnosis of SJMS and in eliminating other diseases.

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**Keywords:** MacLeod syndrome; Bronchiolitis obliterans; Computed tomography

### 1. Case report

A 22-year-old female who was a non-smoker presented to the Accident and Emergency Department with acute left-sided chest pain. The first chest X-ray showed left-sided hyperlucency (Fig. 1a), and therefore, a left-sided pneumothorax with compression of the left lung were suspected. The doctors at the A&E Department inserted the first chest tube, however, the control chest X-ray (Fig. 1b) showed no pulmonary expansion, and there was no air leakage from the drain. Twenty-four hours later, they decided to remove the chest tube and replace it with another one, but the condition remained unchanged (Fig. 1c). After 48 h of the second drain, they attempted with a third drain (Fig. 1d), but neither lung expansion, nor air leakage from the chest tube could be observed. Finally, they decided to consult a thoracic surgeon. The thoracic surgeon took a careful history from the patient and her parents, and it revealed that she had a history of acute neonatal bronchopulmonary infections which required hospitalization. During the past two years, she also noticed repeated chest infections.

On physical examination, she weighed 44 kg and her height was 154 cm. Respiratory examination revealed reduced chest expansion and crackles were detected in the left lung. Chest radiographs showed evidence of asymmetry and of radiolucency between two lung fields. Both clinical and radiologic findings suggested the diagnosis of Swyer–James Syndrome, which was further confirmed by the

following investigations: the result of the pulmonary function test was consistent with a diffusely obstructive airway disease with a certain degree of restrictive patterns [in our patient VC = 1.76 l (54% of estimated value), FEV<sub>1</sub> = 1.43 l (of estimated value)]. Computed tomography (CT) of the thorax (Fig. 2a,b) documented hyperlucency and diminished vascularity in the left upper lobe with hyperinflation of the pulmonary parenchyma. No endobronchial lesion or bronchiectasis was identified. Pulmonary ventilation perfusion scans (Fig. 2c,d) revealed markedly diminished blood flow to all hyperlucent areas that were defined by chest radiographs and CT-scan.

In order to improve the respiratory function and to relieve the symptoms, surgical treatment was considered after obtaining an informed consent. Under general anesthesia using one-lung ventilation, a left upper lobectomy was done by lateral thoracotomy.

### 2. Comment

Swyer–James–MacLeod Syndrome (SJMS) is a rare condition characterized by unilateral hyperlucency of a part of or the entire lung which was first described in 1953 by Swyer and James [1, 2], and further detailed by MacLeod. It is considered to be an acquired disease secondary to viral bronchiolitis and pneumonitis in childhood.

Initially, SJMS was thought to be congenital in origin and attributed to hypoplasia of the pulmonary artery. This theory, however, was later disproved because the number of bronchial generations and vascular branches in these patients are normal [3]. SJMS is now considered a postinfectious manifestation of childhood bronchiolitis obliterans

\*Corresponding author. Tel.: + (33) 4 77 82 84 74; fax: + (33) 4 77 82 84 62.

E-mail address: sulaiman@creatis.insa-lyon.fr, abdulrazzaq.sulaiman@chu-st-etienne.fr (A. Sulaiman).

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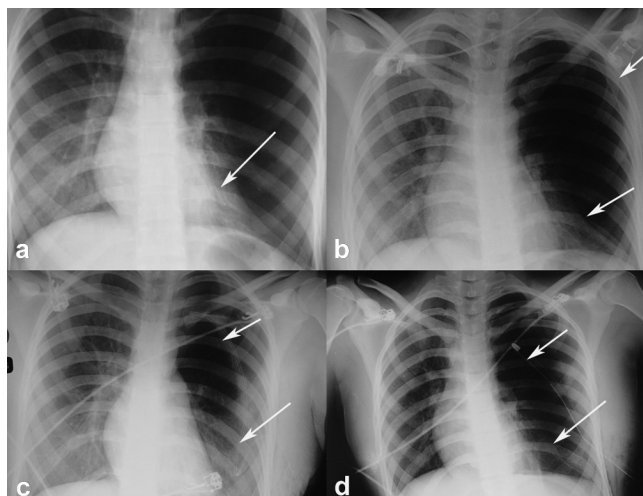


Fig. 1. (a) Chest X-ray shows a hyperlucent and over-expanded left lung with a small right lung with left lower lobe limitation, 'long arrows' in (a). (b, c and d) Three chest tube drains indicated by the 'short arrows' with the same limits of the lower lobe.

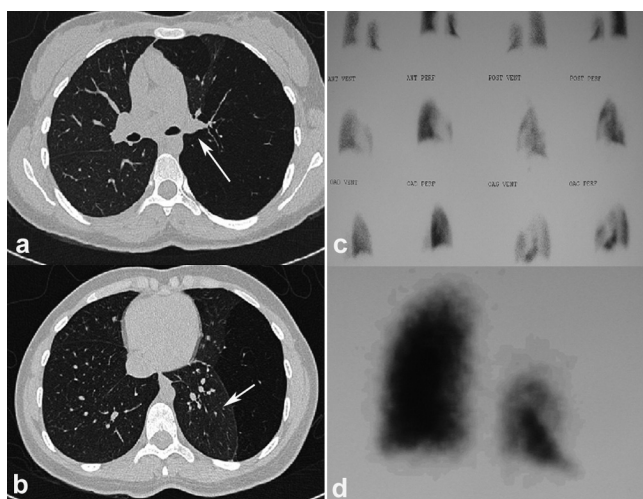


Fig. 2. Computed tomography (a) hyperlucency of nearly all the left upper lobe due to diminished vasculature with a hypoplastic left pulmonary artery (arrow). (b) Normal left lower lobe. Ventilation perfusion scintigraphy; (c and d) showed both ventilation and perfusion defects on single breath images in the left upper lobe.

[4]. These respiratory infections include measles, whooping cough by *Bordetella pertussis*, tuberculosis, *Mycoplasma pneumoniae*, and influenza A. Adenovirus types 3, 7, and 21 appear to be the agents which are most commonly associated with bronchiolitis obliterans in this population [5]. Our patient had an episode of acute neonatal bronchopulmonary infection.

Bronchiolitis obliterans results in inflammation and fibrosis in the walls and contiguous tissues of the membranous and respiratory bronchioles with narrowing of their lumens [6]. Fibrosis of the interalveolar septae causes obliteration of the pulmonary capillary bed and secondarily diminishes blood flow to the major pulmonary artery segments, resulting in the hypoplastic arterial development as observed in SJMS. In addition, the reduction in ventilation causes a

compensatory decrease in perfusion. Hyperexpansion of the terminal air sacs secondary to bronchiolar obstruction of the peripheral airways offers additional mechanical resistance to flow through the alveolar capillaries and contributes to atrophy of the vascular beds [3].

Hypoplasia of the pulmonary arteries is a reflection of this decrease in blood flow. Because of the decreased parenchymal perfusion, this syndrome is often referred to as translucent or hyperlucent unilateral lung [7]. Although classically considered to affect one lung or one lobe, CT has made it apparent that the disease may be more heterogeneous in distribution than previously thought and that contralateral parenchymal lesion may be present [8]. The percentage of patients that develop SJMS after bronchiolitis obliterans is not known, although, the condition is considered unusual. The onset of symptoms typically occurs during infancy or early childhood in association with frequent respiratory infections. Patients usually present with productive cough, shortness of breath, and dyspnoea on exertion. Occasionally, haemoptysis may be seen. These symptoms correlate with the degree of bronchiectasis present in individual patients in association with this syndrome. Patients who have little or no bronchiectasis have minor symptoms or are asymptomatic and may, therefore, miss their diagnosis until adulthood. These adult patients with SJMS are often diagnosed after a chest radiograph obtained for another reason. The classic chest radiographic finding is a pronounced one-sided hyperlucency due to oligoemia of the involved segments of the lung [9]. A mediastinal shift toward the affected side may occur on inspiration. Expiratory radiograph may demonstrate air trapping or a shift of the mediastinum towards the unaffected side. Bronchiectatic changes may also be observed on chest radiographs.

Despite characteristic findings by chest radiography, CT is the imaging technique of choice in establishing the diagnosis of SJMS [10]. On CT, SJMS appears as hyperlucent areas due to decreased pulmonary perfusion of the lung without an anteroposterior gradient attenuation. As stated earlier, the hyperlucency may not be confined to one lung. In addition, the distribution of hyperlucency within the involved lung may be inhomogeneous with patches of normal lung attenuation demonstrable. Ventilation/perfusion scans are an important modality in the diagnosis of SJMS. Ventilation/perfusion scans document matched ventilation and perfusion defect [4]. In our patient, her chest pain with unilateral hyperlucency chest X-ray in the emergency unit carried the diagnosis of a left-sided pneumothorax; she underwent three chest tube drainages and numerous chest radiographs without SJMS being suspected. Chest radiography may, therefore, underestimate the prevalence of this syndrome. Despite numerous chest radiographic examinations, SJMS was not diagnosed until after another imaging study was obtained on the advice of a thoracic surgeon. This finding illustrates the serendipity that often occurs in the diagnosis of this syndrome.

Finally, this case reaffirms the utility of CT-scan and scintigraphy in establishing the presence of SJMS.

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### eComment: Swyer–James–MacLeod syndrome: addendum to the clinical appearance

**Authors:** *Thorsten Walles, Schillerhöhe Hospital, Solitudestrasse 18, 70839 Gerlingen, Germany; Thomas Kyriass, A. Sibylle Maier, Godehard Friedel*

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With great interest we read the recent case study of Sulaiman and colleagues on the Swyer–James–MacLeod syndrome [1]. The authors presented a comprehensive review of this rare clinical picture. Our institute's surgical experience with this disease has been similar to that of the authors; however, we have one addendum to make to complete the report.

The authors shed light on the radiological and clinical findings going along with secondary obliteration of the pulmonary capillary bed and the disappearance of blood flow in the major pulmonary artery segments, resulting in the hypoplastic arterial development observed. Besides the described pathological perfusion changes in the diseased lung sections, their occasionally bizarre hyperinflation results in a compression of healthy lung areas,

ultimately causing their atelectases. In one case, a middle lobe Swyer–James–MacLeod syndrome caused complete chronic atelectasis of the healthy right upper lobe. The patient's afflictions and medical condition are deteriorated accordingly. Radiologists and thoracic surgeons alike may be misguided by computed tomography scans of these patients, feigning two non-coherent pathologies.

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### eComment: Diagnostic and surgical considerations in Swyer–James–MacLeod syndrome

**Authors:** *Nikolaos Barbetakis, Thoracic Surgery Department, Theagenio Hospital, A. Simeonidi 2, 54007 Thessaloniki, Greece; Dimitrios Paliouras, Christos Asteriou, Georgios Samanidis*  
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We read with great interest the article by Sulaiman et al. [1] regarding a case of a patient with Swyer–James–MacLeod syndrome and we would like to congratulate them and highlight a few points.

Swyer–James–MacLeod syndrome is morphologically characterized by the presence of constrictive bronchiolitis with dilatation and destruction of alveolar structures, resulting in significant air trapping and lung hypoperfusion. The main pathogenetic factor seems to be acute bronchiolitis with obliteration of small airway in childhood [2].

Ventilation perfusion (VQ) scan is very helpful in determining the extent of the disease and correlates well with high resolution computed tomography (HCRT) which seems to be the most valuable technique. The volume and antero-posterior attenuation gradient of the affected lung, size and distribution of central and peripheral pulmonary arteries, air trapping, the patency of main airways and presence of bronchiectasis are investigated. The appearance of the lungs on forced expiration is important in the assessment of Swyer–James–MacLeod syndrome with HCRT and therefore the patient's cooperation is essential. The patient should be placed in the prone position to help identify the typical mosaic pattern of the syndrome.

Surgical treatment is predicated upon the principle that a reduction in lung volume can favorably affect the mechanics of the diaphragm, chest wall and airways. Video-assisted thoracic surgery techniques could be very useful and effective [2].

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