



## Swyer-James-MacLeod syndrome: an uncommon conundrum

### Síndrome de Swyer-James-MacLeod: un enigma poco común

Vitorino Modesto-dos Santos,\*  
Lister Arruda Modesto-dos Santos†

\*Medicine Department from  
Armed Forces Hospital and Catholic  
University of Brasília-DF, Brazil.  
ORCID: 0000-0002-7033-6074;

†General Surgery of State Worker's  
Hospital, Sao Paulo-SP, Brazil.  
ORCID: 0000-0003-4647-4044.

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Dear editor:

The Swyer-James-MacLeod syndrome (SJML) is an uncommon cause of obstructive pulmonary disease with unilateral hyperlucent lung, often in children, with estimated prevalence of 0.01% in over than 17,000 chest X-rays, related to restrictive respiratory tests and lung infections, and the data on the outcomes during pregnancy are scarce.<sup>1-5</sup>

We read the illustrative report in this Journal by Torres-Rodríguez ST *et al.*<sup>5</sup> of a 31-year-old woman who was incidentally diagnosed with SJML when searched medical attention due to a mild insidious retrosternal pain and the imaging studies revealed the hyperlucent left lung that was displacing the mediastinum to the right, and abnormal pulmonary vasculature with hypoperfusion and hypoplasia of the left

pulmonary artery. The authors emphasized her childhood and adolescence history of respiratory diseases like common colds, bronchitis, pneumonia; and an uncomplicated COVID-19 infection. They also stressed the diagnostic cornerstones (unilateral lung hyperlucency, reduction of vascularity and perfusion loss), besides the conservative management of this patient.<sup>5</sup> Considering the important role of case studies about uncommon and scarcely reported diseases, it seems opportune add short comments of novel literature data about SJML.<sup>1-4</sup>

Al-Bakri O *et al.*<sup>1</sup> reported a 29-year-old primipara in the prenatal evaluation, who had the diagnosis of SJML seven years before by typical images in the left lung, besides asthma and pneumonia in the early infancy, followed by recurrent respiratory infections. The lung function tests at 29 weeks showed FEV1 1.79 L (59% predicted), and FEV1/FVC ratio 64%; and at 34 weeks the FEV1 1.99 L (65% predicted), and FEV1/FVC ratio 66%.<sup>1</sup> She underwent budesonide/formoterol and salbutamol inhalers for the month prior to delivery, and had an uncomplicated vaginal delivery under early epidural control of pain.<sup>1</sup> The authors emphasized the uneventful pregnancy and vaginal labor of a patient with SJMS and moderate obstructive pulmonary disease maintained under a close vigilance.<sup>1</sup> They also stressed the epidural control of pain to reduce oxygen consumption and minute ventilation during the first and second labor stages of women with respiratory diseases.<sup>1</sup> Chlapoutakis S *et al.*<sup>2</sup> described a 63-year-old heavy smoker man with coronaropathy and chronic obstructive lung disease, using beclomethasone, formoterol, and glycopyrronium; and more than three hospitalizations due to infectious exacerbations the last six months. The pulmonary function tests revealed that the FEV1 and the FVC values were of 65%/67% and 99%/102% of predicted values, pre- and post-bronchodilator, respectively.<sup>2</sup> Chest images showed emphysema, bronchiectasis, and an hyperlucent left lower lobe; with diagnosis of SJML he underwent salbutamol/ipratropium, corticosteroid, and antibiotics, besides recommendation for smoking cessation and regular vaccinations.<sup>2</sup> The authors emphasized the uncommon diagnosis of SJML syndrome in adulthood, mainly in

absence of antecedent significant pulmonary infection during the childhood.<sup>2</sup> Cheng YH, *et al.*<sup>3</sup> reported a 17-year-old male, who had repeated pneumonia since the childhood, presenting with acute left thoracic pain and dyspnea; and the chest imaging studies showed almost complete atelectasis and bronchiectasis in the right lung, hyperlucency of left lung, mediastinal deviation to the right, and bilateral pneumothorax. The chest X-ray images were very similar to bilateral pneumothorax and due to air leak for over a week a thoracoscopic procedure with chemical pleurodesis was performed.<sup>3</sup> The overexpansion of the left lung by SJMS caused atelectasis of the contralateral lung; the authors stressed the pneumothorax as an ominous complication because of the dysfunction of the other lung, but a closed thoracostomy drainage may be lifesaving.<sup>3</sup> Fontes CP, *et al.*<sup>4</sup> described a 34-year-old male with antecedent of pulmonary tuberculosis in adolescence, who had a prolonged fever, right-sided pleuritic pain and purulent sputum. The chest imaging studies showed a hyperlucent right upper lobe (as seen on ancient lung images), homolateral opacities, besides a reduced pulmonary vasculature and perfusion.<sup>4</sup> Cultures of sputum and bronchoalveolar lavage were negative, and ventilation/perfusion scintigraphy showed a matched ventilation and perfusion defect of the affected areas; these evaluations and the clinical manifestations confirmed the SJMS as final diagnosis.<sup>4</sup> The authors highlighted the role of recognition of this syndrome in adulthood, because of higher possibility of misdiagnosis and adverse effects of the inappropriate management.<sup>4</sup>

Worthy of note, an occasional diagnosis of SJMS during adulthood may constitute a more challenging task, inclusive because some of the differential hypotheses like pneumothorax and pulmonary hypertension are also complications of the syndrome;<sup>2-4</sup> and if accurately followed, affected women who desire can have uneventful pregnancy.<sup>1</sup> The authors believe that the case studies can lessen the underdiagnosis and misdiagnosis.

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*Correspondence:*

**Vitorino Modesto dos Santos MD, PhD;**

**Lister Arruda Modesto dos Santos MD.**

**E-mail:** vitorinomodesto@gmail.com

## Reply

### Respuesta

Servio Tulio Torres-Rodríguez,\*

Karla Marleny Reyes-Zúñiga,\*

Iván Estuardo Chang-Castillo,\*

Danilo Herrera-Cruz\*

\*Hospital San Vicente,  
Guatemala, Guatemala.

I believe that as more cases are published, the recognition of this entity will be much better understood, recognized and treated. The history of lung disease must be kept in mind, since many architectural changes of the bronchial tree are a consequence of them. Although bronchiolitis is more common in children, it can affect adults. The disease can progress to severe obstructive respiratory failure, especially in the case of constrictive bronchiolitis.<sup>1</sup> The radiological image of a hyperlucent lung should guide us to the suspicion of this syndrome and the treatment is in principle conservative and aimed at preventing recurrent infections.<sup>2</sup> Surgical treatment

should be reserved for advanced cases and the magnitude of the type of surgery will be justified by the structural damage to the lung, or by the implications of the hyperinflated lung that can act as a tension pneumothorax by displacing the mediastinum so severely that it can limit healthy lung function; Therefore, surgical intervention is reserved for: a) patients with recurrent lung infections, b) patients who do not respond, or c) patients whose symptoms are not adequately controlled with optimal medical treatment.<sup>3</sup>

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*Correspondence:*

**Servio Tulio Torres-Rodríguez, MD.**

Hospital San Vicente, Guatemala, Guatemala.

**E-mail:** stuliotr@gmail.com