

# Complication of post-infectious bronchiolitis obliterans (Swyer-James syndrome)

## COMPLICAÇÃO DE BRONQUIOLITE OBLITERANTE PÓS-INFECCIOSA (SÍNDROME DE SWYER-JAMES-MACLEOD)

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### SUMMARY

Swyer-James syndrome is a complication of post-infectious bronchiolitis obliterans that causes inflammation and fibrosis of the bronchial walls. There are two types: asymptomatic, with most cases diagnosed in adults during routine radiological examinations; and symptomatic, most commonly found in children. Here, we report the case of a 6-year-old child with recurrent dyspnea since the age of 3, who showed signs and symptoms of bronchiolitis obliterans and radiological signs of bronchial wall thickening and air trapping. The clinical and radiological findings led to the diagnosis of Swyer-James syndrome. Treatment of this syndrome is intended to reduce the pulmonary lesions and improve the patient's quality of life.

**Keywords:** bronchiolitis, lung, hyperlucent, lung diseases.

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Article received: 9/9/2014

Accepted for publication: 9/16/2014

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<http://dx.doi.org/10.1590/1806-9282.61.05.404>

### INTRODUCTION

Bronchiolitis obliterans (BO) is a chronic obstructive pulmonary disease of the distal airway that is characterized by an inflammatory process caused by damage to the small airways. BO affects mainly male children during the first year of life.<sup>1-3</sup> Despite the lack of available epidemiology, there is a high prevalence in countries within the southern hemisphere, such as Argentina, Brazil, Chile and New Zealand.<sup>4,5</sup> There are several etiologies mentioned in the literature, including viral and bacterial infections, but the most frequent cause of BO in children is post-infectious, i.e. possibly related to *Paramyxovirus morbillivirus*, *Bordetella pertussis*, *Mycobacterium tuberculosis*, *Mycoplasma pneumoniae*, *Streptococcus* type B, *Legionella pneumophila*, influenza, parainfluenza, respiratory syncytial virus and adenovirus.<sup>5-7</sup>

One of the complications of post-infectious BO (PIBO) caused by adenovirus is Swyer-James syndrome (SJS), also known as unilateral hyperlucent lung syndrome.<sup>7,8</sup> SJS is defined as unilateral hyperlucency of a lobe or the entire lung due to pulmonary hypoperfusion, showing a decrease in the vascular network and volume of the affected lung

or lobe.<sup>9-11</sup> Functionally, SJS is characterized by a decrease in volume during inspiration and air trapping during expiration, which results from bronchiolar obstruction.<sup>12,13</sup>

Since this is a rare disease, it is important to understand the overall clinical picture of SJS to exclude the differential diagnosis of other diseases that are associated with bronchiolitis. For this purpose, we hereby report the case of a patient diagnosed with SJS.

### CASE REPORT

A 6-year-old male patient was referred to Hospital Infantil Cosme e Damião in Porto Velho, State of Rondônia, with the complaint of severe dyspnea. He had a history of bi-monthly dyspnea and wheezing since 6 months of age and was medicated with inhalation (fenoterol hydrobromide, ipratropium bromide, and saline solution) and amoxicillin. Starting at age 3, the patient was constantly dyspneic and required daily use of inhalation, showing a fortnightly intensification that led him to the city hospital, where he was treated for pneumonia. At the age of 6, he had an episode of severe dyspnea, intense headache, and coughing attacks, and sought emergency medical help, being then

medicated with inhalation, antibiotics, and oxygen therapy. His general condition worsened and he was referred to a pediatric hospital, where symptomatic treatment was performed. The initial physical examination revealed tachypnea (respiratory rate, 40/min) with intercostal and subcostal retractions, nasal flaring, and a lowered wishbone. A subsequent lung examination detected the presence of bilateral wheezing and rales, and the remainder of the physical examination was normal. The complete blood count showed normocytic and normochromic erythrocytes, and leukocyte atypia. Adequate laboratory tests ruled out the diagnoses of cystic fibrosis and tuberculosis.

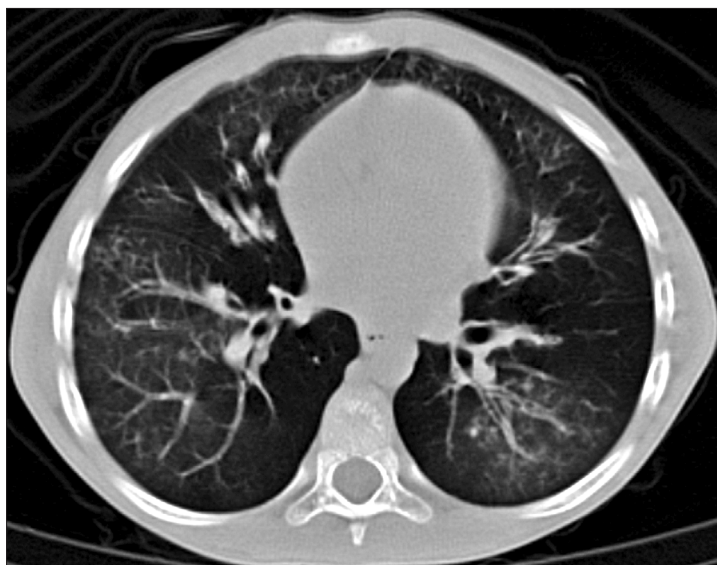
A chest radiograph revealed hypertransparency and hypoperfusion of the left lung (Figure 1). This finding

led to computed tomography (CT) of the chest, which showed marked diffuse thickening of the bronchial walls associated with air trapping areas in addition to areas of diffuse oligemia randomly distributed throughout both lungs, showing topographical signs compatible with relevant inflammatory bronchopathy (Figure 2). Thus, the diagnosis of SJS was made through the patient's clinical evaluation and radiological findings.

Based on this diagnosis, the patient was suggested to do the following: respiratory physiotherapy, vaccination against pneumococcal diseases and influenza, and decrease their exposure to episode triggers, as well as corticosteroid and bronchodilator use. The use of antibiotics was restricted to exacerbations only.



**FIGURE 1** Severe bulbo-duodenitis with mild diffuse bleeding.



**FIGURE 2** At close inspection with narrow-band imaging (NBI), enlarged and rigid duodenal villousities are seen.

## DISCUSSION

The physiopathology of SJS is explained by the consequences of BO, which include inflammation and fibrosis of the bronchial walls that results in lumen narrowing, reduced ventilation, and vasoconstriction that leads to decreased perfusion. Fibrosis of the interalveolar *septa* causes obliteration of the pulmonary capillary, reducing blood flow to the pulmonary artery segments and triggering arterial hypoplasia.<sup>8</sup>

There are two types of SJS: asymptomatic, with most cases being diagnosed in adults during routine radiological examinations; and symptomatic, which is most commonly found in children.<sup>13</sup> The clinical manifestations of SJS may vary, but typically include productive cough, dyspnea on exertion, and sometimes hemoptysis. On physical examination, the patient may show hypomobility on the side affected with hypertympanism to percussion, decreased breath sounds, and occasionally crepitant rales.<sup>14-15</sup> The diagnosis can be made through anamnesis and the report of recurrent lower respiratory infections; on physical examination, changes in the respiratory system on chest radiograph reveal specific radiological aspects.<sup>11</sup> The pulmonary function test can also be performed to demonstrate a ventilator change, reinforcing the existence of an obstructive airway disease.<sup>15</sup>

CT can address other data associated with the syndrome, such as the presence of bronchiectasis and agenesis of the pulmonary artery, which is useful in excluding other differential diagnoses.<sup>11</sup> The differential diagnosis of SJS should include congenital disorders, namely agenesis or occlusion of the pulmonary artery, congenital absence of the pectoralis major muscle, and congenital lobar emphysema, as well as central airway obstruction caused by foreign body aspiration or lesions inside the bronchi, lung cysts, and pneumatoceles.<sup>15,16</sup> SJS is currently considered one of the forms of presentation of PIBO.<sup>16</sup>

There are currently few data on the treatment of patients diagnosed with the Swyer-James syndrome, and most of the measures to be taken are merely supportive, to help minimize the formation of new pulmonary lesions and improve the life quality of those patients.

## RESUMO

Complicação de bronquiolite obliterante pós-infecciosa (Síndrome de Swyer-James-Macleod)

A síndrome de Swyer-James-Macleod é uma complicação da bronquiolite pós-infecciosa, ocasionando inflamação

e fibrose das paredes dos bronquíolos. Pode se manifestar de duas formas: assintomática, sendo a maioria diagnosticada na fase adulta, quando o paciente se submete a exames radiológicos de rotina, e a forma sintomática, que é mais encontrada em crianças. Relatamos um caso de uma criança de 6 anos de idade com crises de dispneia de repetição desde os 3 anos, apresentando sinais e sintomas de bronquiolite obliterante e sinais radiológicos de espessamento brônquico e aprisionamento aéreo. Por meio da clínica e achados radiológicos, foi feito o diagnóstico de síndrome de Swyer-James-Macleod. O tratamento dessa síndrome visa a reduzir as lesões pulmonares e a melhorar a qualidade de vida do paciente.

**Palavras-chave:** pulmão hipertransparente, bronquiolite, pulmão, criança.

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