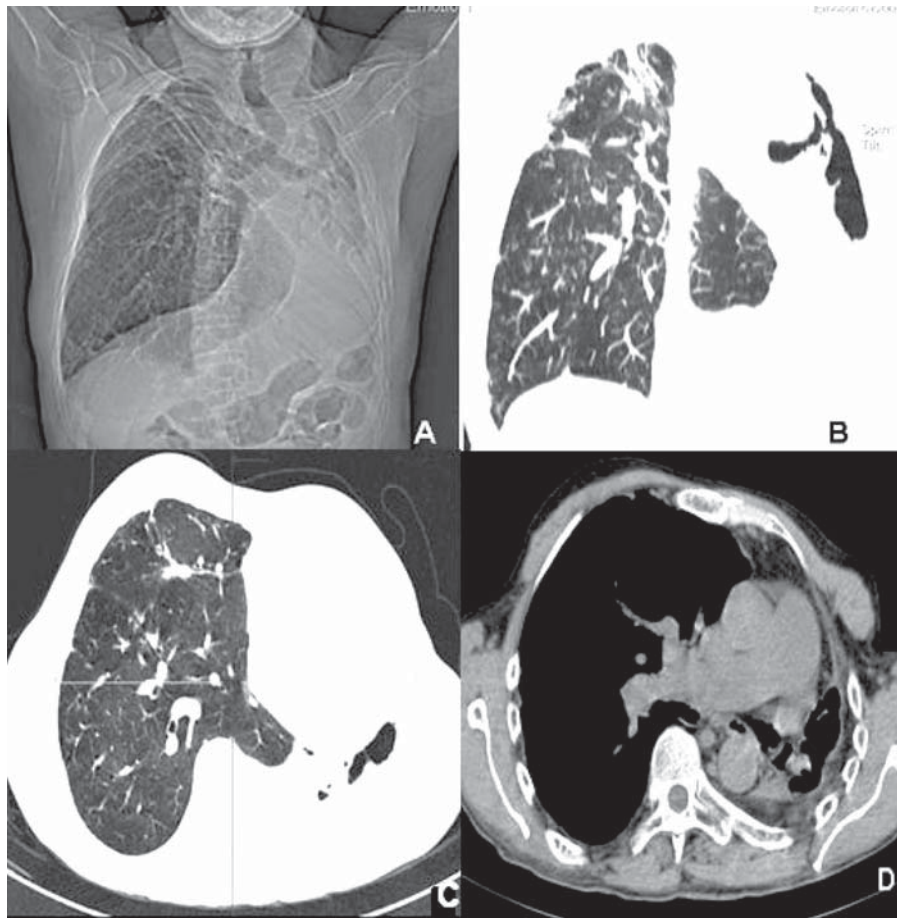


## Photoclinic



**Figure 1.** A) Chest topogram showing almost total opacity of the left hemithorax, with homolateral cardiac and mediastinum deviation. B, C, and D) Computed chest tomography revealing reduced left hemithorax, wall thoracic hypotrophy and trachea deviation to the left; dilated trachea (greater than 3 cm), main bronchi, and some bronchiectasis were observed. The right lung appears hyperinflated with herniations to the left hemithorax. Mediastinal venous tributaries appeared dilated.

A 66-year-old male presented with productive cough and breathlessness after an upper airway infection. There was no fever or other symptoms. Main antecedents were birth at home, long-term use of formoterol and budesonide to control episodes of breathlessness and wheezing since infancy. There was a family history of asthma. Diagnosis was elsewhere established by diurnal variation in his peak expiratory flow of more than 20% over a two-week period and an increase of more than 200 mL and 12% in FEV1 after bronchodilator use. He denied a history of tuberculosis, foreign body aspiration or recurrent chest infections. The patient was an ex-smoker (15 pack years) who quit smoking 14 years earlier. Physical examination showed the following: BMI of 17.9 kg/m<sup>2</sup>; heart rate, 144 bpm, blood pressure, 120/70 mmHg, and respiratory rate of 32 ipm. His thorax was deformed with scoliosis, scapulae asymmetry, and left costal retraction. Hyper-reso-

nant percussion notes were elicited throughout the right hemithorax, in addition to ronchi, scattered wheezing, and bronchovesicular sounds. Upon examination of the left hemithorax, percussion notes were dull with a distinct reduction in respiratory sounds and absence of the vocal fremitus in the thoracic basal area. Other clinical findings were unremarkable, without any stigma of a co-existent congenital disorder. Blood determinations revealed hemoglobin, 121 g/L; hematocrit, 37.2%; leukocytes, 13.5×10<sup>9</sup>/L (segmented: 82%, lymphocytes: 10%, monocytes: 8%) and platelets, 283×10<sup>9</sup>/L. Tuberculin test and culture for microorganisms both in sputum and the bronchial aspirate were negative, including acid-fast mycobacteria. Evaluation of cardiac function was unremarkable.

A chest imaging study showed the presence of an opaque left hemithorax with mediastinum deviation, hyperinflated right lung, no lymph node enlargement and free pleural spaces.

Treated with corticosteroids, theophylline, and a short-acting beta-2 agonist, he became asymptomatic and was discharged to outpatient follow-up seven days after admission. One year later, he remains in good clinical condition and is able to perform normal activities.

**What is your diagnosis?  
See the next page for diagnosis.**

Vitorino Modesto dos Santos MD PhD<sup>1,2</sup>, Vitor Laerte Pinto Junior MD PhD<sup>1</sup>, Guilherme Teixeira Guimarães Paixão MD<sup>2</sup>, Késsia Danielle Sampaio de Faria MD<sup>1</sup>

**Authors' affiliations:** <sup>1</sup>Internal Medicine Department, Catholic University Medical Course, Brasília-DF, Brazil, <sup>2</sup>Internal Medicine Department, Armed Forces Hospital, Brasília-DF, Brazil.

**Corresponding author and reprints:** Vitorino Modesto dos Santos MD PhD, SMPW Quadra 14, Conjunto 2, Lote 7, Casa A. 71.741-402, Brasília-DF, Brazil. Tel.: +55-61-3380-2666, fax: +55-61-3233-1599, E-mail: vitorinomodesto@gmail.com.

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Opaque hemithorax can represent gross displacement of mediastinal structures, trachea, and heart to the affected side, with compensatory hyperinflation of the contralateral lung and herniation across midline in the upper thorax. This uncommon condition is usually due to pleural effusion, consolidation, collapse, massive tumor, fibrothorax, pneumectomy, or a maldeveloped lung (agenesis, aplasia, or hypoplasia). Hypoplasia is characterized by a decrease in the numbers or size of the airways, vessels, and alveoli, resulting in a small fibrotic and non-functioning lung, sometimes associated with other congenital anomalies and bronchiectasis.<sup>1,2</sup> Consensual clinical diagnostic criteria are lacking to facilitate the identification of lung hypoplasia but computed tomography (CT), angiography and fiberoptic bronchoscopy often contribute to the diagnosis. CT scans are useful to confirm lung under-development and differentiate hypoplasia from conditions that may mimic it on chest radiography images.<sup>1</sup> Lung hypoplasia has been described in asthmatic patients, but this association is very rare.<sup>3-6</sup>

The patient was under longstanding follow-up because of asthma-related symptoms. Previously, his thoracic changes had been evaluated by pulmonary perfusion scan with Tc 99m, which showed normal distribution of the tracer on the right lung and no uptake on the contralateral lung parenchyma. This finding was then attributed either to a left lung underdevelopment or vascular compressive phenomenon that occurred in early childhood. Furthermore, fiberoptic bronchoscopy disclosed indicative features of hypoplasia of the left bronchial branches (dilated main bronchus, and narrow openings of the upper and lower lobe bronchi),<sup>1-3</sup> while the right tracheobronchial tree was normal. Recent high resolution followed by helicoidal CT tomographies revealed images of left lung hypoplasia, with reduction of left hemithorax volume, wall muscle hypotrophy and trachea deviation to the left. The right lung was hyperinflated and developed extensive herniations to the left hemithorax. The left lung was hypoplastic, with a dilated main bronchus, and some saccular and fusiform bronchiectasis. There was no residual functioning left lung parenchyma or air trapping, and the mediastinum venous tributaries appeared dilated (Figure 1D). Of note, the left pulmonary artery branching was not clearly observed on CT contrast administration.<sup>1,3,6,7</sup> As a whole, the aforementioned data were strongly indicative of left lung hypoplasia, but the timing and exact mechanism of hypoplasia could not be unequivocally established. Imaging data contributed to rule out atelectasis from other causes, such as bronchiectasis with collapse, advanced fibrothorax and Swyer-James-MacLeod syndrome (SJMS).<sup>1</sup> Despite the absence of other anomalies or association with known etiologic factors, the lung hypoplasia reported here could be idiopathic (1 – 2 per 12,000 births) or secondary to decreased hemithorax volume, lung vascular perfusion, fetal respiratory movement, or lung fluid.<sup>1</sup> Trachea and main bronchi dilation were considered secondary to chronic retraction forces. Another major concern was the possibility of asthma and right apical tuberculosis in early childhood, which evolved with left lung sequels,<sup>4,6,7</sup> but this hypothesis was not confirmed.

Similar to the present report, Kant described right lung hypoplasia (pertaining to the fourth group of Monaldi classification), in a 20-year-old male who presented with bronchiectasis and no other developmental abnormalities<sup>1</sup>; nevertheless, his patient had no asthma. Unilateral agenesis, aplasia or hypoplasia of the lung

have been scarcely described, mainly in patients with asthma, and the degree of pulmonary development arrest has been variable.<sup>1-7</sup> Booth and Berry reviewed 200 cases of unilateral pulmonary agenesis, including 17 of their own patients, and found diverse associated congenital abnormalities (cardiovascular, facial, gastrointestinal, spinal, and urogenital). Of note, when the right lung was affected, the outcome was poorer.<sup>2</sup> Sharma et al. described unilateral pulmonary agenesis in two patients without coexistent congenital disorders (isolated). One showed agenesis of the right lung and was affected by *S. pneumoniae*; while the other presented with aplasia of the left upper lobe and tuberculosis.<sup>7</sup> Arrest of lung development has been very rarely described in association with bronchial asthma.<sup>3-6</sup> In fact, studying radiographic features of 1016 asthmatic adults, Pickup et al. found a unique patient (0.09%) with pulmonary hypoplasia.<sup>4</sup> Kushwaha and colleagues reported isolated left upper lobe aplasia and lower lobe hypoplasia in a 36-year-old male with asthma. Their imaging studies showed complete opacification of the left hemithorax, with hyperinflation and herniation of the right lung in addition to the mediastinum shifting to the left.<sup>3</sup> Rathkopf et al. reported isolated right-sided pulmonary agenesis in a 23-year-old female with asthma since early childhood.<sup>5</sup> Left-sided opaque hemithorax with homolateral shifting of the mediastinum and right lung herniation was also described by Sahay and colleagues in an 8-year-old asthmatic boy who presented with isolated aplasia of the left lung.<sup>6</sup> Although one major concern in the case reported here could be potential misdiagnosis involving asthma and SJMS as recently reported by Walia et al.,<sup>8</sup> the respiratory symptoms of this patient were always relieved by classic treatment for asthmatic episodes. Moreover, the diagnosis of asthma was confirmed by the respective lung function tests.<sup>3,6</sup> Therefore, these findings were consistent with concomitant asthma and lung hypoplasia.

Despite the weaknesses inherent to single case studies, this report could increase physicians' awareness about the possibility of coexistent asthma and underdeveloped lungs.

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