Horseshoe lung in a young child at Tygerberg Hospital, South Africa

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Horseshoe lung is a very rare congenital malformation in which the bases of the right and left lungs are fused to each other by a narrow isthmus. Although rare, a hyperlucent area in the lower left lung, close to the vertebral column, may represent a horseshoe lung. Horseshoe lung is often associated with scimitar syndrome. Here, we present a case of a 2-year-old girl who presented with recurrent chest infections and wheezing associated with horseshoe lung. The right lung appeared to be hypoplastic.

Keywords. horseshoe lung; scimitar syndrome; congenital venolobar syndrome; bronchoscopy; hypoplastic lung.

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Horseshoe lung is a very rare congenital malformation in which the bases of the right and left lungs are fused to each other by a narrow isthmus posterior to the pericardial reflection but anterior to the aorta and oesophagus. The isthmus originates from the hypoplastic lung.[1] The embryological mechanism for horseshoe lung is not known but non-separation of splanchnic mesodermal mass has been suggested.[2]

Hyperlucent area in the lower left lung, close to the vertebral column, may represent a horseshoe lung. Parenchymal, airway and vascular abnormalities are associated with horseshoe lung. Horseshoe lung is most seen in association with scimitar syndrome or other hypoplastic conditions of the right lung. The scimitar syndrome was first described in 1836 by Chassinat.[3] It is also referred to as congenital venolobar syndrome, as a constellation of cardio-pulmonary anomalies. The characteristic abnormality is anomalous pulmonary venous return from a part of or the entire right lung to the inferior vena cava.[4] The anomalous venous connection is seen as curvilinear shadow close to the right heart border and resembles a curved Turkish sword. Less than 50 cases have been reported in the literature.[5]

Case

A 2-year-old girl presented with a history of recurrent chest infections and persistent wheezing that were not responding to treatment. She was HIV-negative, there was no history of contact with tuberculosis and all tuberculosis testing including Gene Xpert MTB/RIF was negative. Plain frontal chest radiograph demonstrated a well-defined lucency in the left lower lobe medially, with a curvilinear outline resembling an inferior accessory lobe and fissure. The right lung appeared smaller than the left, but the heart was normally positioned (Fig. 1).

Bronchoscopy was performed and demonstrated anterior mid-tracheal compression with pulsation. The bronchial configuration was abnormal on the right side with an absent bronchus intermedius.

A contrasted chest computed tomography (CT) scan showed an ‘isthmus’ of the lung in continuity with the right lung, extending from the right hemithorax across the midline (between the heart and descending aorta) into the left hemithorax, consistent with a horseshoe lung (Fig. 2). The arterial supply was from the right lung branches (Fig. 3A) and there was a clearly defined pleural separation from the left lung (the presumed ‘isthmic fissure’). Arterial supply to the isthmus was via the right lower lobe vessels and venous drainage appeared to be directly into the right atrium. A small bronchial...
branch of the left lower lobe bronchial tree was noted to supply the isthmus, but this appeared attenuated across the vertebral body and descending aorta, resulting in selective air-trapping of the isthmic segment (Fig. B).

Even though the plain radiograph demonstrated a possible smaller right lung, there were no CT features of right (or left) pulmonary hypoplasia. The pulmonary arteries were of adequate size, if not slightly plump. The heart was normally positioned on the left. There were, however, some significant associated venous abnormalities in the form of a scimitar-type vessel, which ascended from the right lung base peripherally to join another scimitar-type vessel descending from union of two superior segmental intervertebral veins, to enter the superior vena cava as it feeds into the right atrium (Fig. 3C). The right atrium was noted to be prominent.

Other CT scan findings included an absent minor fissure and a bovine variant of the aortic arch with the left common carotid artery arising together with the brachiocephalic trunk from the aortic arch, a common variant. This appeared to cause some anterior impression on the trachea superiorly.

Discussion
Horseshoe lung is a very rare congenital malformation in which the bases of the right and left lungs are fused to each other by a narrow isthmus posterior to the pericardial reflection. Fusion occurs via the parietal pleural defect that allows communication of pleural cavities either directly without a fissure, or indirectly through intervening visceral pleura (isthmic fissure). Very few cases have been described in the literature, with the first case report appearing in the textbook by Spencer[6] in 1962. Most cases involve lung hypoplasia and/or vascular anomalies.

The most common associated abnormality is scimitar syndrome, which may be seen in up to 85% of cases.[7] A few reported cases have been associated with other cardiovascular anomalies including atrial septal defect, ventricular septal defect, tetralogy of Fallot, and hypoplastic left ventricle.[8] Right lung hypoplasia is much more common than left lung hypoplasia, which is seldom reported.[9]

When there is a pneumothorax in a child with horseshoe lung, it may affect both lungs due to the band of parenchymal tissue, which causes communication between the two lungs.

Plain chest X-rays may be suggestive due to the air trapping in the parenchyma of the horseshoe lung, which has a lower density on X-ray.

Conclusion
A hyperlucent area in the lower left lung is a clue to the diagnosis of horseshoe lung.

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Author contributions. PG, JM, LM provided clinical care to the patient. SA and PG interpreted the imaging and other data. All authors approved the manuscript for publication.

Fig. 2. Axial multidetector computed tomography (MDCT) on lung window demonstrates an isthmus of basal lung in continuity with the right lung, extending across the midline between the heart anteriorly and the aorta and vertebral body posteriorly. There is a distinct pleural interface between the isthmus and the left lung.

Fig. 3. Coronal oblique reconstructions demonstrating vascular supply, bronchial supply, and the scimitar vessel. (A) Coronal oblique reconstruction using maximum intensity projection (MIP) with a slab thickness of 7 mm, demonstrating the arterial origin of the vessels arising from the right inferior pulmonary artery branches extending across the midline to supply the isthmus of lung. (B) Coronal oblique reconstruction using minimum intensity projection (MinIP) with a slab of 5 mm, demonstrating a small attenuated bronchial branch from the right lung extending across the midline to supply the isthmus which has air-trapping. (C) A contrast-enhanced coronal oblique maximum intensity projection (MIP) reconstruction on mediastinal window demonstrates the enhancing curvilinear structure ascending from the right lung base (scimitar vein) joining a second peripheral venous structure before entering the superior vena cava at its entry into the right atrium.
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