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Unusual echocardiographic finding leading to diagnosis of pulmonary sequestration

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Introduction

Pulmonary sequestration is an embryonic mass of non- functioning lung tissue that does not communicate with the tracheobronchial tree and has a reported incidence of 0.15%-6.4% of all the pulmonary malformations.[1] This anomaly is classified as either intralobar or extralobar with the later variety lying outside the normal investment of visceral pleura. The arterial supply is predominantly by an anomalous artery usually arising from either abdominal or thoracic aorta, while the venous drainage occurs commonly via systemic rather than pulmonary veins.[2–3]

Identification of the anomalous arterial supply has therapeutic implication because the majority of infants clinically present large shunt lesions attributed to these channels in early infancy.[4] The diagnosis in such cases is usually established by computed tomography (CT), angiography, magnetic resonance angiography and conventional angiography[5]. We report a 28 day old neonate who presented with features of large shunt lesion, in which echocardiography was instrumental in the diagnosis of a large collateral supplying the sequestrated lung.

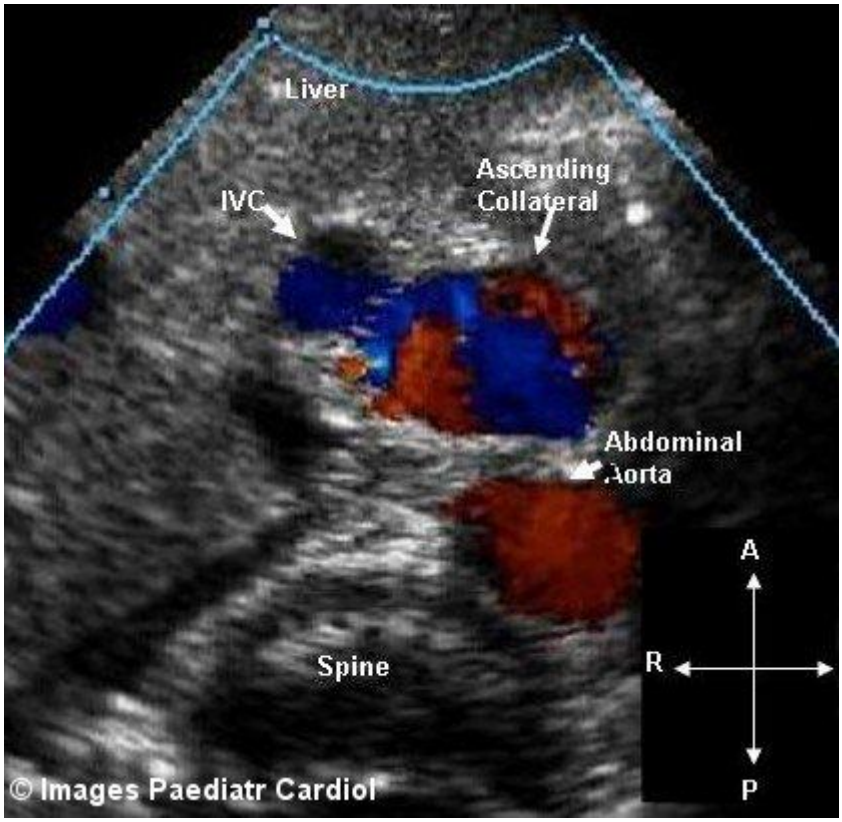
Patient

A 28 day old term baby boy weighing 2.1 kg (birth weight – 2.7 kg) presented to our hospital with a chief complaint of difficulty in feeding and failure to thrive since birth.

On physical examination the baby had a respiratory rate of 66/min, heart rate of 150/min and and was saturating at 94% in room air. Cardiovascular examination revealed wide and fixed splitting of the second heart sound and a pansystolic murmur at the left lower sternal border. He also had significant hepatomegaly. X-ray chest was suggestive of cardiomegaly with mild bronchovascular prominence. Echocardiography revealed situs solitus, levocardia, atrio-ventricular and ventriculo-arterial concordance with normal systemic venous drainage. The right lower pulmonary vein was draining into the IVC. The remaining pulmonary veins were

normally connected to the left atrium (LA). A sub-costal coronal view done to delineate situs showed an additional channel located anterior to the aorta (Fig 1).

Fig 1 Subcostal coronal view for situs delineation showing anomalous channel anterior to transverse section of abdominal aorta



Further echo examination showed fenestrated fossa ovalis atrial septal defects and small anterior ventricular septal defects with left to right shunt (Figs 2,3).

Fig 2 Subcostal 4 chamber view shows small fenestrations in the atrial septum

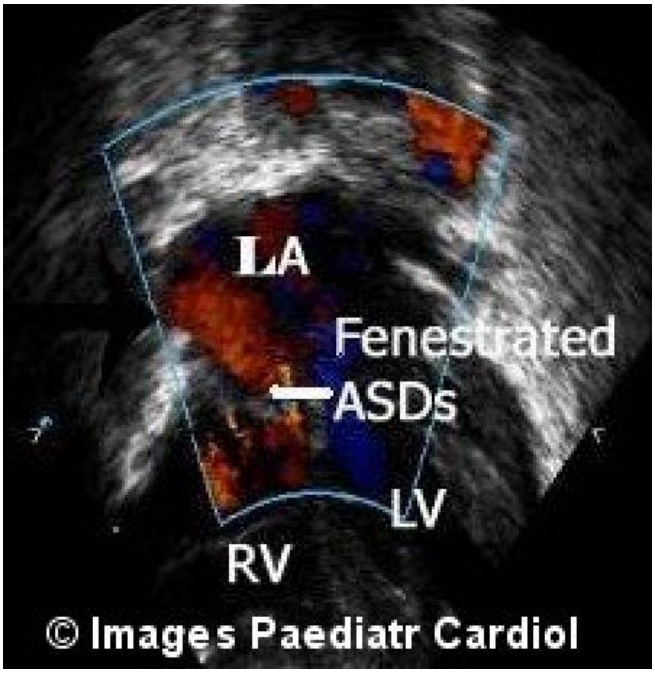
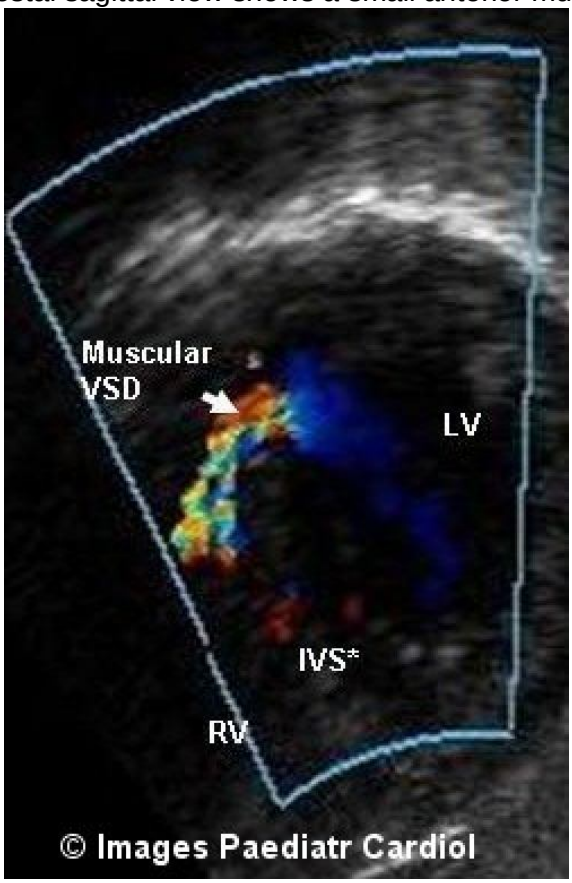


Fig 3 Subcostal sagittal view shows a small anterior muscular VSD



Interrogation of the descending aorta in sagittal view revealed an anomalous ascending channel similar in caliber to that of aorta and arising at the level of L2-L3 vertebrae. It was seen communicating with the sequestrated pulmonary tissue at a level just above the left dome of the diaphragm (Figs 4,5).

Fig 4 Subcostal sagittal view showing anomalous ascending channel from descending aorta.

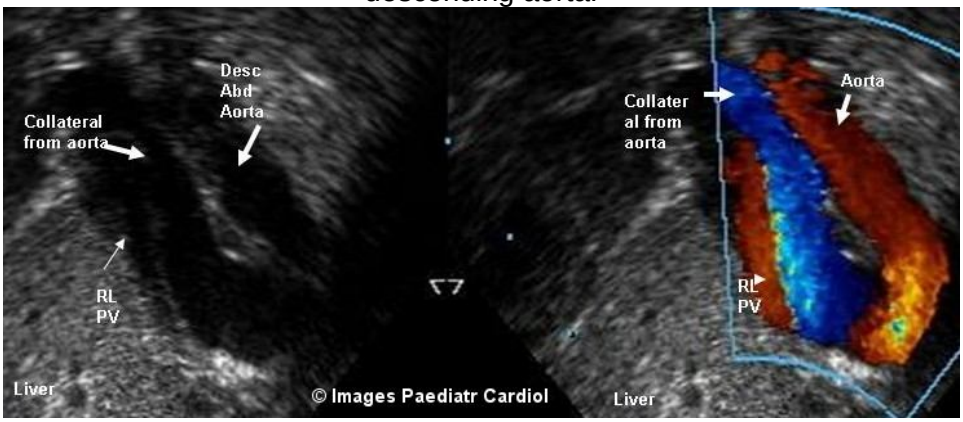
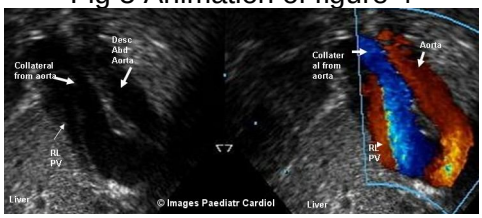


Fig 5 Animation of figure 4



Doppler examination of the descending aorta, though narrow beyond this point, was essentially normal otherwise. The venous drainage of the sequestrated segment was through the portal venous system (Figs 6,7).

Fig 6 Subcostal coronal view showing collateral to sequestrated lung and draining venous channel.

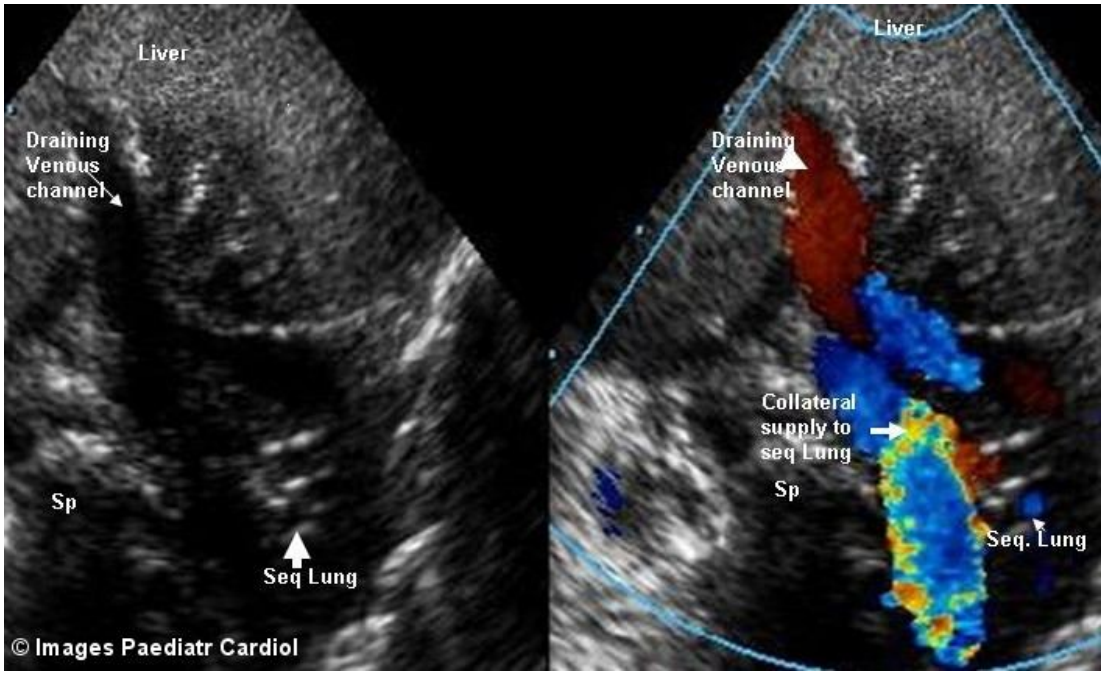
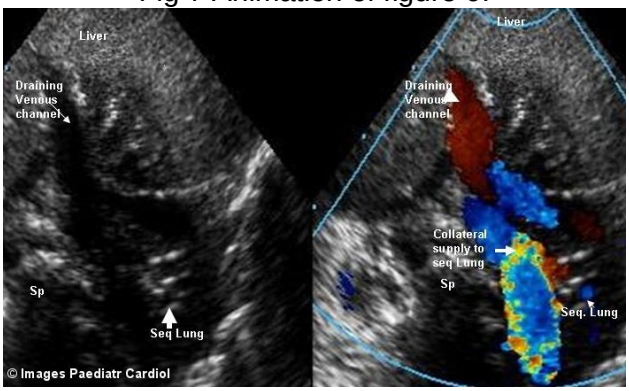


Fig 7 Animation of figure 6.



Ultrasonography (USG) and subsequent plain CT chest and upper abdomen confirmed the presence of a sequestrated pulmonary segment in the left supra-diaphragmatic area.
This confirmed pulmonary sequestration with arterial supply from the abdominal aorta and draining into the portal venous system. CT angiography was planned, but deferred in view of the deranged renal function.

Discussion

Pulmonary sequestration with or without scimitar syndrome is a well recognized entity. The subset presenting early in life have typical manifestations of a shunt lesion.

Collaterals supplying the sequestrated tissue form a major extra-cardiac site for left to right shunting. These children are usually treated with either surgical or catheter guided occlusion of the collaterals. Those with late presentation generally have respiratory distress, chronic cough, difficulty in feeding, recurrent chest infections and failure to thrive as the presenting complaints. The diagnosis is most often made by radiological findings on X-ray, bronchography, CT, USG and MRA.[5]

Our patient presented with the classical triad of congestive heart failure, failure to thrive and a murmur suggestive of an acyanotic shunt lesion. In view of the deranged renal function, echocardiography was the only modality available to establish the diagnosis. In the absence of radiological scimitar sign and other associations, scimitar syndrome and scimitar variant were ruled out.

Echo images of the ascending channel were the only pointer towards the diagnosis in our reference case and this unique finding, to the best of our knowledge, has never been reported earlier in the literature.

References

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