

# An Unusual Neonatal Presentation of Scimitar Syndrome

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## Abstract

### Keywords

- neonatology
- perinatology
- pediatric cardiology
- pulmonology

Scimitar syndrome is characterized by partial or total anomalous pulmonary venous return from the right lung along with pulmonary hypoplasia. We present a case of a 37 weeks' gestation male infant with antenatal ultrasound findings of suspected partial anomalous pulmonary venous return (PAPVR) and coarctation of the aorta. The newborn presented with respiratory distress, a chest X-ray and chest computed tomography (CT) angiogram confirmed the diagnosis of scimitar syndrome. The combination of scimitar syndrome with aortic coarctation is extremely rare with only a few cases previously reported.

## Case History

A 37 weeks' gestation male infant was born to a 31 years old gravida 2, para 1 mother with a dichorionic diamniotic twin pregnancy. Parents were nonconsanguineous and family history was noncontributory. She had a spontaneous healthy pregnancy with protective antenatal serology. Her Group B streptococcus (GBS) status was unknown and gestational diabetes screen was normal. First trimester screening for trisomy 18, 13, and 21 was negative. At 20 weeks' gestation, ultrasound findings were suggestive of partial anomalous pulmonary venous return (PAPVR) and suspected coarctation of aorta for twin A. There was, however, no growth discordance between the twins.

A fetal echocardiogram at 26 weeks and 6 days' gestation age confirmed findings of PAPVR with right inferior pulmonary vein draining into the right atrium, narrowing/hypoplasia of the right branch of pulmonary artery, and a suspicion of coarctation of the aorta. Subsequent ultrasounds confirmed cardiac findings in twin A with growth restriction at 35 weeks' gestation. Both twins had ultrasounds findings of normal amniotic fluid indices and biophysical profiles.

The twins were born at 37 weeks' gestation by cesarean section because of evolving growth discordance. Twin B weighed 3.5 kg at birth with no respiratory distress and

preductal oxygen saturations of 90% in room air. Twin B remained well to discharge home.

Twin A had a decreased breath sounds in the right hemithorax with an ejection systolic murmur of grade 3/6 intensity heard in the pulmonary area. Heart rate was 160/min and blood pressure in the right arm was 65/40 mm Hg. Lower limbs were cool to touch with weak femoral and posterior tibial arterial pulsation. Preductal blood pressure measured with right arm: 64/36, mean 48 mm Hg, while postductal blood pressure measured with right leg: 54/28, mean 37 mm Hg.

Postnatal echocardiography confirmed coarctation of the isthmus of the aorta, following which a prostaglandin infusion was started. He required a maximum of 1 L/min of nasal cannula flow with 25% fraction of inspired oxygen (FiO<sub>2</sub>) to maintain preductal oxygen saturations greater than 90%. The first chest X-ray (►Fig. 1) showed atelectasis of the right hemithorax with a mediastinal shift to the right which raised concern of the diagnosis of scimitar syndrome. A chest computed tomography (CT) angiogram performed on day 3 of life showed hypoplastic right lung (►Fig. 2) with only two bronchi, a hypoplastic right pulmonary artery and PAPVR of the right pulmonary vein to the inferior vena cava (IVC) essentially at the level of the atrial–caval junction (►Figs. 3–5). There were no identified systemic collaterals, or evidence of scimitar vein stenosis. Imaging of the lung bases demonstrated no evidence

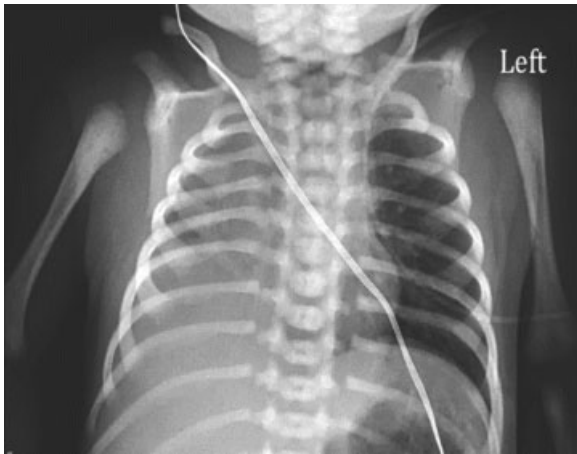
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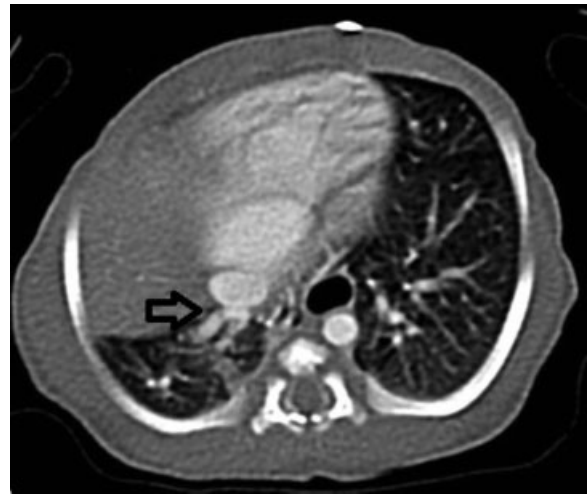




**Fig. 1** Anteroposterior (AP) supine chest radiograph: There is abnormal added dense opacity and volume loss in the right hemithorax. The left heart border and cardiac size are normal. There is no focal abnormality of the left lung or bony thorax. Decreased right thoracic loss of volume may suggest hypogenetic lung in a child with known partial anomalous pulmonary venous return (PAPVR).

of a sequestration, but definite leftward herniation of the medial basal right lower lung parenchyma across the midline was present in the central inferior chest anterior to the distal esophagus and descending thoracic aorta, consistent with the presence of a pseudohorseshoe lung malformation (► **Figs. 6 and 7**).

A nonstenotic bicuspid aortic valve and coarctation of aorta with hypoplasia of the distal aortic arch were identified. Subsequent echocardiography studies demonstrated increased pulmonary pressure, a large patent ductus arteriosus (PDA) and a small anterior muscular ventricular septal defect (VSD). In our case, the PAPVR anomaly included one of the pulmonary veins draining into the right atrium that was detected antenatally, and another pulmonary vein draining



**Fig. 3** Axial image from contrast-enhanced computed tomography (CT) chest (lung windows) showing small right lower lung and anomalous pulmonary venous drainage (scimitar vein) draining into the posterior inferior vena cava (IVC) near the level of the junction with the right atrium.

to the IVC at the level of the atrial–caval junction, as per the chest CT angiogram findings.

He had surgical repair of the aortic coarctation by subclavian arterial flap repair on day 16 of age. Pulmonary hypertension improved on sildenafil following successful postoperative extubation.

## Discussion

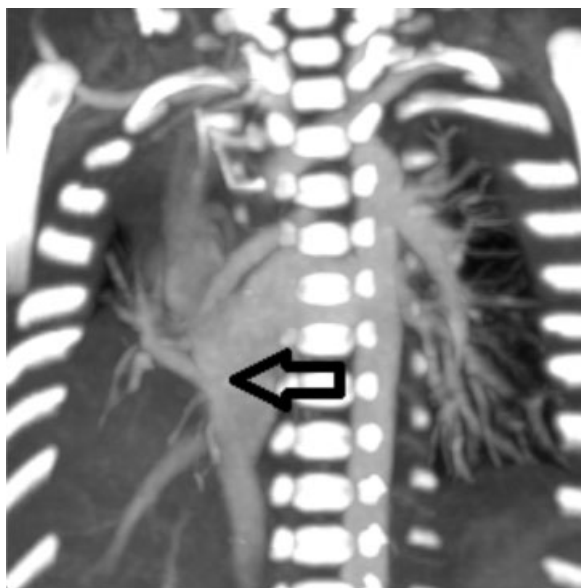
Scimitar syndrome was first described in 1836 by George Cooper during the autopsy of a 10-month-old infant. The



**Fig. 2** Axial chest computed tomography (CT) angiogram (lung windows): The right lung is small compared with the normal sized left lung and the mediastinal structures are shifted rightwards due to volume loss in the right thorax. The right pulmonary artery is small/hypoplastic (open arrow) compared with left pulmonary artery (black arrow).



**Fig. 4** Maximum intensity projection (MIP) sagittal reformatted contrast-enhanced computed tomography (CT) image demonstrates abnormal obliquely and vertically oriented draining vein from the right lower lobe entering the posterior aspect of inferior vena cava (IVC) at its junction with the right atrium.

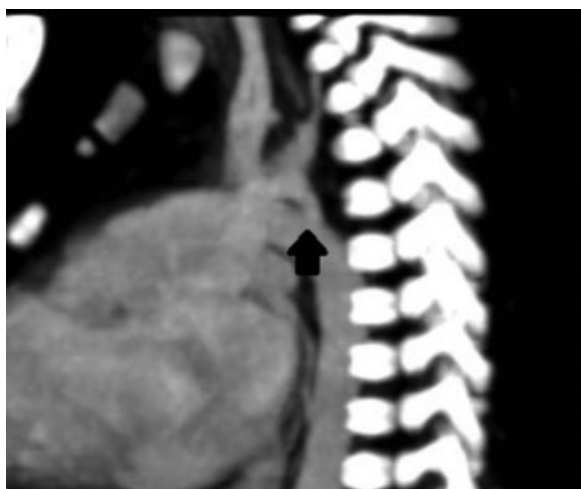


**Fig. 5** Maximum intensity projection (MIP) coronal reformatted contrast-enhanced computed tomography (CT) image demonstrates abnormal pulmonary venous drainage. The right inferior pulmonary vein is joining the posterolateral aspect of the right atrium/superior vena cava (RA/SVC) junction.

incidence of scimitar syndrome is estimated at 1 to 3/100,000 live births.<sup>1,2</sup> However, its true incidence may be much higher as some of those affected may be asymptomatic.

The diagnosis is made by echocardiography, characteristic chest X-ray findings, and confirmatory CT chest findings. CT chest angiography provides excellent visualization of the vascular anatomy of this complex congenital defect noninvasively.<sup>3</sup>

Scimitar syndrome is a rare association of congenital cardiopulmonary anomalies characterized by anomalous pulmonary veins (scimitar vein) that drains into the IVC, a hypoplastic right lung, and dextroposition of the heart. It has been reported in 3 to 6% of patients with partial anomalous pulmonary venous connection. Patients are either diagnosed



**Fig. 6** Contrast-enhanced computed tomography (CT) chest maximum intensity projection (MIP) sagittal reformatted image demonstrating coarctation of the distal transverse aorta at the isthmus (black arrow).



**Fig. 7** Axial image from contrast-enhanced computed tomography (CT) chest (lung windows) showing bulging pulmonary parenchyma (isthmus) from the medial right lung base crossing the midline with adjacent enveloping pleural reflection or pseudohorseshoe lung (open arrow). No abnormal contralateral bronchi or pulmonary arterial branches were detected.

early with severe symptoms (infantile type) or late with minimal symptoms (childhood/adult type).<sup>4,5</sup>

Schramel et al recognized two different types of scimitar veins: type 1 is simple classic vein that runs from the middle of the right lung to the cardiophrenic angle and type 2 is double arched vein in the upper and lower lung zones, with drainage into the left atrium and inferior caval vein.<sup>4,5</sup>

The presentation of the syndrome could be as early as immediately after birth, and the diagnosis should be suspected when the signs of respiratory distress and/or heart failure are present in association with radiological findings of cardiac dextroposition and haziness or suspicion of atelectasis of the right lung.<sup>6</sup>

Scimitar syndrome often presents as an isolated finding with a benign outcome; however, when associated with other cardiac defects and pulmonary arterial hypertension, there is an increased risk of congestive heart failure and mortality. The etiology of pulmonary hypertension in infants with scimitar syndrome is variable and may be related to a decreased right-sided pulmonary vascular bed, scimitar vein stenosis, and unappreciated systemic collateral blood flow along with intracardiac shunts, or additional comorbidities, such as developmental lung disease. Horseshoe lung is a rare pulmonary malformation associated with scimitar syndrome. Eighty percent of reported cases occur in patients with congenital pulmonary venolobar (scimitar) syndrome.<sup>7</sup> Typical features of horseshoe lung include an isthmus of the pulmonary parenchymal tissue arising from the right lung base, bridging the right and left lungs, which resides posterior to the inferior aspect of the middle mediastinum. Pulmonary arterial and bronchial supply extend from the right lower lobe pulmonary artery and bronchus. It has been suggested that when the isthmus is completely encased in a pleural membrane, as it was in our patient, this be termed pseudohorseshoe lung.<sup>8</sup>

Pulmonary hypertension is a problem seen in infants and older children with scimitar syndrome.<sup>9</sup> The age of presentation

**Table 1** Review of literature of published cases with scimitar syndrome that associated with aortic arch abnormalities

Author	Year	Number of cases	Cardiac anomaly	Outcome
Gikonyo et al <sup>14</sup>	1986	2	Case #1: Bicuspid aortic valve, tubular hypoplasia of aortic arch, ASD, VSD, PDA. Persistent left superior vena cava to coronary sinus	Died at the age of 7 d in a state of congestive failure following right ventricular angiography
			Case # 2: Tubular hypoplasia of aortic arch, VSD, PDA, anomalous venous drainage	Experienced intraoperative cardiac arrest, and resuscitation was unsuccessful
Oshima et al <sup>12</sup>	2003	1	Coarctation of aorta, extremely hypoplastic right pulmonary artery and right lung, pulmonary sequestration, and ASD	At 35 d of life, patient underwent successful repair of coarctation and PAPVR
Marcondes et al <sup>15</sup>	2014	1	Mild aortic arch hypoplasia, no coarctation, PAPVR; hypoplastic right pulmonary artery. Aortopulmonary collaterals arising from the descending aorta	At 7 wk of life patient underwent successful surgical repair
Rezaei et al <sup>16</sup>	2016	1	Coarctation of aorta, right pulmonary artery stenosis, and right to left ductus arteriosus flow	Discharged on day 22nd of life postcoarctation correction

Abbreviations: ASD, atrial septal defect; PAPVR, partial anomalous pulmonary venous return; PDA, patent ductus arteriosus; VSD, ventricular septal defect.

and the presence of associated anomalies are important in predicting the outcome. In general, presentation in infancy and presence of heart failure are poor prognostic factors.

The combination of scimitar syndrome with aortic coarctation is rare, with only few cases reported.<sup>10–12</sup> The literature describes the correction of associated cardiac defects with the therapeutic occlusion of anomalous arterial supply to the affected lung can led to benign outcomes comparable to that in primarily isolated forms.<sup>13</sup>

We did a review of literature of published cases with scimitar syndrome that associated with aortic arch abnormalities (→Table 1).

## Conclusion

Although scimitar syndrome has been described in the literature, it is rare and variable in its presentation, especially in the neonatal period. This diagnosis should be considered when right-sided “atelectasis” does not improve or when dextroposition of the heart is associated with right lung hypoplasia. Scimitar syndrome can be associated with cardiac anomalies but only few cases are reported with aortic coarctation. Our case report highlights this rare combination.

### Conflict of Interest

None.

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