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# A Rare Case Report of Scimitar Syndrome

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***Abstract:*** *Scimitar syndrome is characterized by partial or total anomalous pulmonary venous return from the right lung along with hypoplasia of the lung .Here we present a case of a 9month old female child with history of repeated lower respiratory tract infections with imaging findings consistent with Scimitar syndrome. We are reporting this case in view of the rarity.*
***Keywords:*** *scimitar syndrome,lower respiratory tract infections,female child*

## I. Introduction

Scimitar syndrome is a rare association of congenital cardiopulmonaryanomalies, and first described in 1836. The syndromeis a form of anomalous pulmonary venous drainagewhich implies partial or total failure of the pulmonary veinsto reach the left atrium. Instead, pulmonary venous drainageis anomalously connected to the systemic vein/s, typically tothe superior vena cava or inferior vena cava or directly tothe right atrium. The name Scimitar syndrome is derived from the curvilinear vascular pattern created on a chest radiogram by the anomalous pulmonary vein coursing along the right side of the heart towards the diaphragm, which resembles a "Scimitar" or Turkish Sword. Although radiographs play a role in the incidental detection and initial imaging evaluation in patients with clinicalsuspicionof congenital lung anomalies, computed tomography is frequentlyrequired for the confirmation of diagnosis, further characterization and preoperative evaluation.

## II. Case Report

A 9 monthold female child born to a 2nddegree consanguineous marriage came with complaints of repeated episodes of lower respiratory tract infections. Child had been visiting local doctor regularly for the same and was treated with oral antibiotics. Even after treating the symptoms were not subsided and child was referred to our hospital .On detailed clinical history,Antenatal and neonatal history was non-contributory.On examination, child had decreased breath sounds and crepitations, on right side on auscultation. Cardiac examination revealed shift of the apex beat to the right side in fourth intercostal space in right parasternal area without any murmur. Patient was subjected to various blood investigations and imaging modalities. The following findings were observed: Hyperinflated left lung with prominent left pulmonary veins and artery. Hypoplastic right lung affecting upper and mid lobe with decreased vascular markings with dextroposition of cardia. Anamolous pulmonary venous drainage (scimitar vein) noted draining into IVC below the diaphragm (infradiaphragmatic type).Hypoplastic hepatic segment of IVC noted .In our case systemic arterial blood supply to the right lung is noted arising from celiac trunk. Absent right pulmonary artery and right main bronchus. Based on these imaging findings a diagnosis of Scimitar syndrome was made.

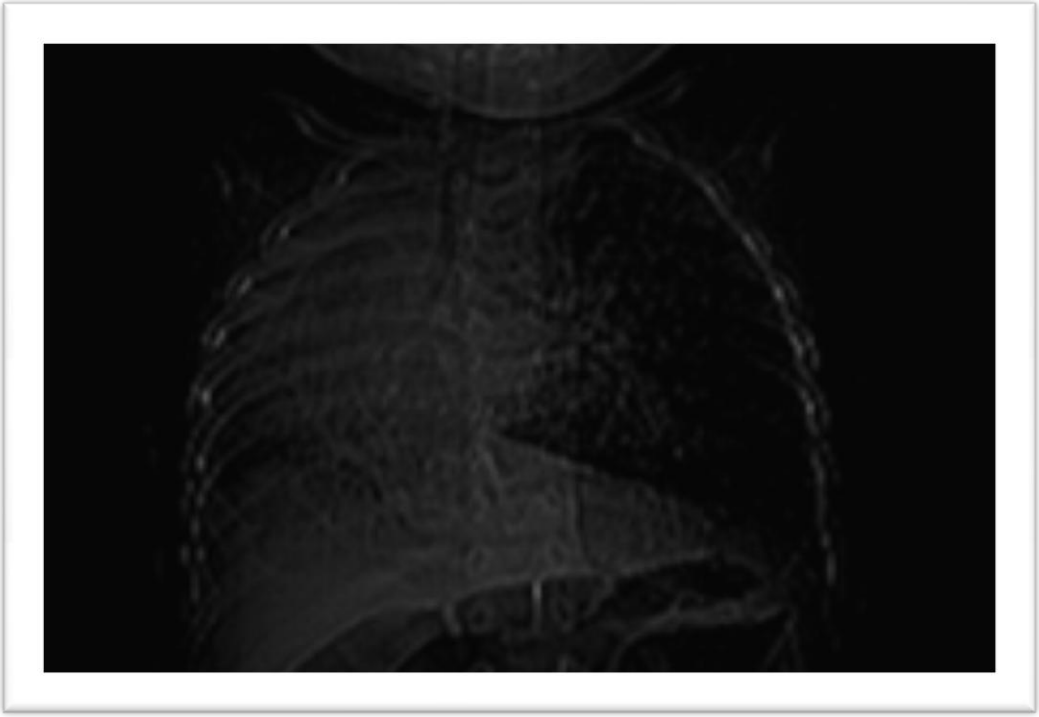

**Fig1:** Scanogram showing decreased right lung volume, trachea and cardia pushed to the right side

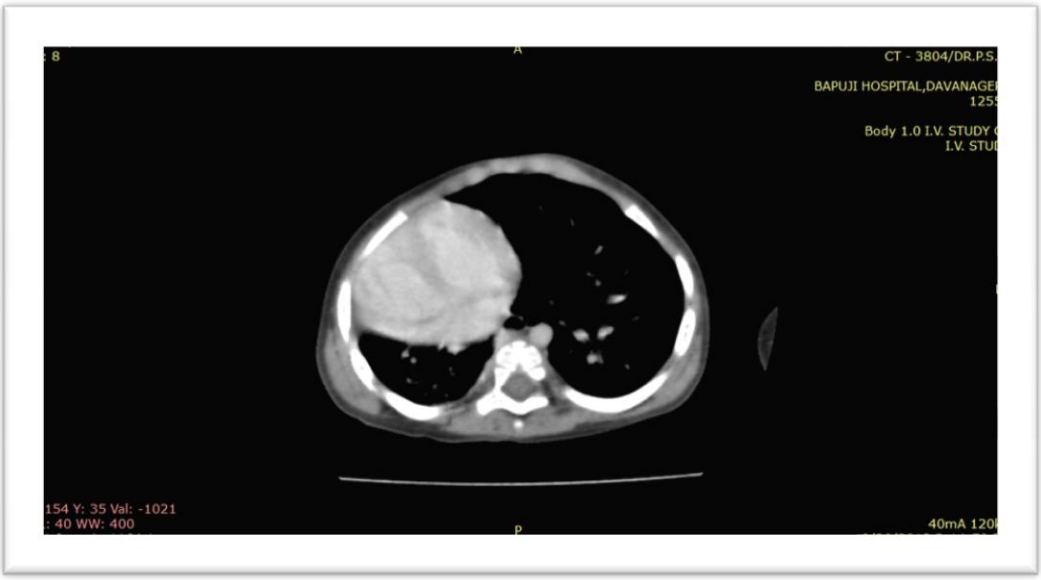


**Fig 2**: Axial CECT Chest demonstrates dextroposition of Cardia with hypoplasia of right lung and compensatory hyperinflated left lung

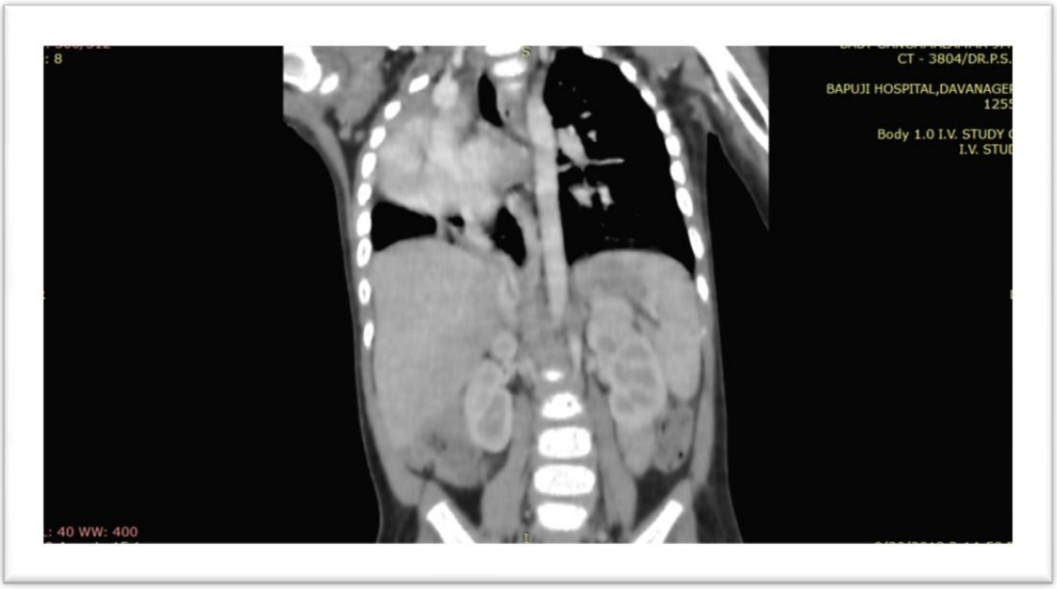


**Fig 3**: Coronal Reformatted CECT showing anamolous pulmonary venous drainage (scimitar vein)draining into IVC below diaphragm

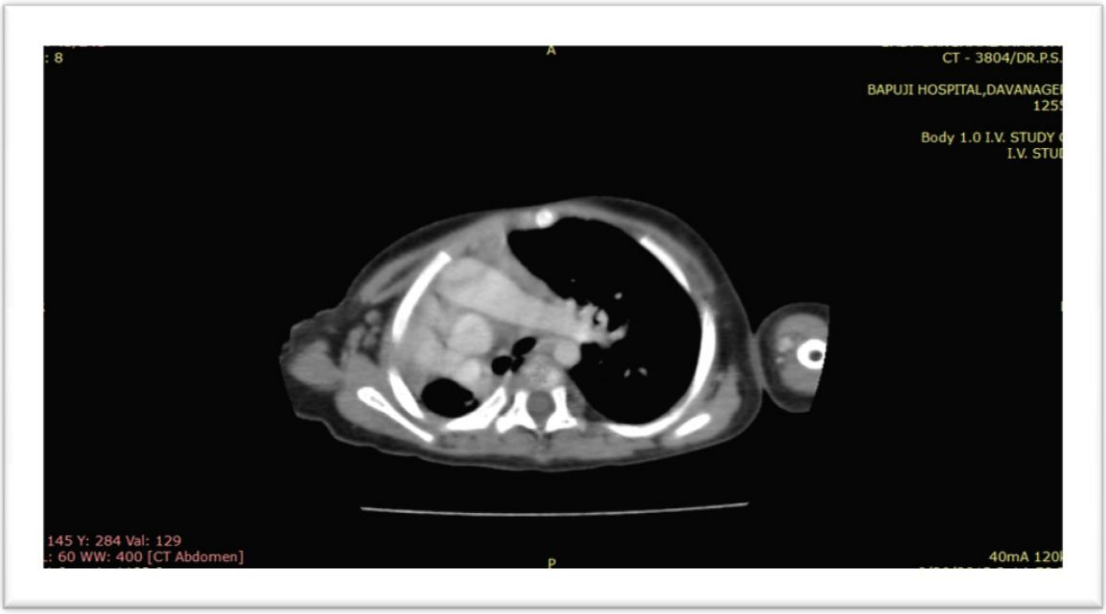


**Fig 4**: Axial CECT chest demonstrates absent right pulmonary artery with prominent left pulmonary artery

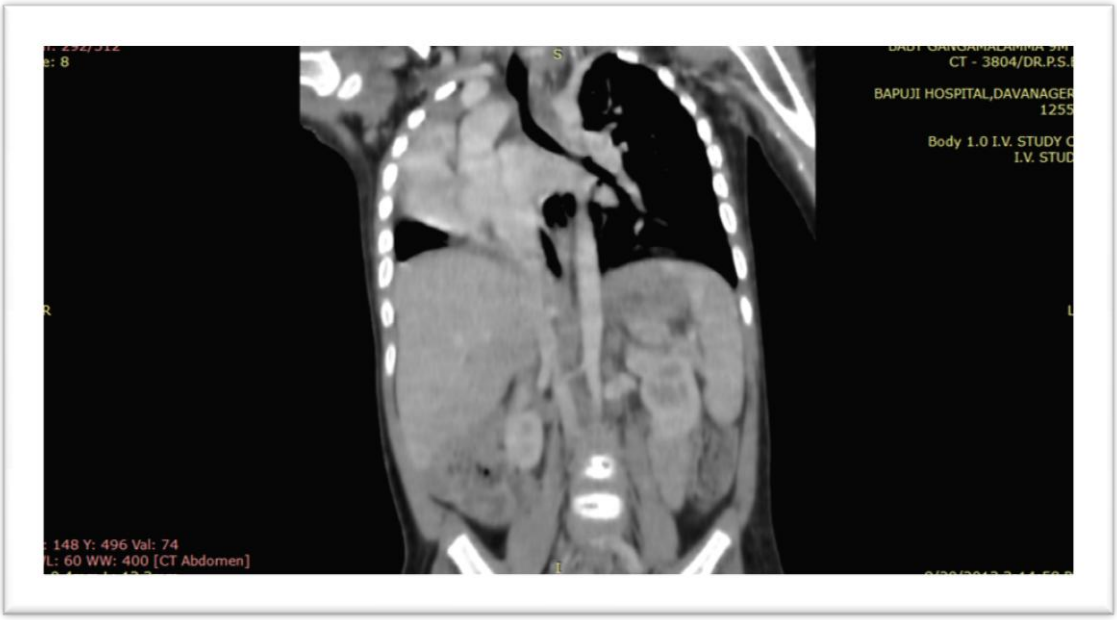


**Fig 5**: Coronal Reformatted CECT showing absent right main bronchus

## III. Discussion

Scimitar syndrome was first described in 1836 by George Cooper during an autopsy of a 10 month old infant. The incidence of scimitar syndrome is 1-3/100,000 live births [1,6]. However, its true incidence may be much higher in view of some asymptomatic cases. The disease often presents in early infancy but may present later in childhood or rarely in adults. In infants it usually presents with tachypnea, congestive heart failure, and pulmonary hypertension. It may lead to recurrent lower respiratory tract infections as in our case. Recurrent episodes of wheeze have also been described in the literature. Hemoptysis may be the presenting symptom rarely [7]. Scimitar syndrome is associated with congenital heart disease, mostly ostium secundum type atrial septal defect. In some cases, ventricular septal defect, patent ductus arteriosus, aortic stenosis, aortic arch anomalies, Shone complex, and Fallot's tetralogy can be seen. Other anomalies associated with this syndrome are sequestration of the lower lobe of the right lung, horseshoe lung and anomalous perfusion from the abdominal aorta [5,8-10].

Diagnosis is made with the help of echocardiography, characteristic chest X-ray, CT chest findings. CT angiography provides excellent visualization of vascular anatomy of this complex congenital defect noninvasively [11].

Cardiac catheterization and angiography are extremely useful tools to confirm the diagnosis and clarify the exact anatomy and degree of pulmonary hypertension. Radionuclide perfusion scan can reveal abnormal arterial supply while ventilation scan can identify bronchial connection in a patient with Scimitar syndrome. In sequestration, aeration can occur through pores of Kohn or through anomalous fistulous connection. Scintigraphic screening is helpful to rule out significant vascular shunting [12].

Treatment primarily involves surgical correction of the anatomical anomaly. However, the most effective surgical method has not been clearly identified. Surgical options include direct anastomosis of the Scimitar vein to the left atrium or division with reimplantation of the anomalous pulmonary vein into the right atrium with baffle insertion to redirect the flow into the left atrium. An intra-atrial patch may be used to create a tunnel, redirecting flow from the anomalous pulmonary vein to the left atrium through an ASD [13]. Uthaman et al. described coil embolization of Scimitar vein and artery, followed by definitive surgery later in life as a method of treatment [14]. Pneumonectomy is required in certain cases. Baskar et al. performed pneumonectomies for three of the six patients in their case series, citing low weight to be the reason for opting out of reimplantation surgery [10].

Pulmonary hypertension is a problem seen in infants and older children with scimitar syndrome [15]. Pulmonary artery pressure was significantly reduced by pulmonary venous stenting in combination with coiling of aberrant vascular supply to the sequestered lung segment in a report by Awasthy et al. [16]. Further experience with such procedures could be useful as it suggests that interventional procedures such as pulmonary venous stenting could be an emergent palliative treatment to relieve pulmonary hypertension in children with Scimitar syndrome.

The age of presentation 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.

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