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Scimitar Syndrome

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Scimitar syndrome is a rare congenital anomaly consisting in part of right pulmonary venous return to the inferior vena cava. There is a clear bimodal presentation of this syndrome with either an infantile manifestation or a pediatric/adult form. The infantile variant is marked by a higher incidence and severity of associated defects, heart failure, pulmonary hypertension, and significant mortality. The patient with the pediatric/adult form is less severely affected and may be asymptomatic on diagnosis. In this article, we review the historical aspects, presentation, and pathophysiology of Scimitar syndrome and discuss available treatment strategies. We emphasize the safe and effective approach developed at Indiana University that obviates both the need for an intra-atrial baffle or use of cardiopulmonary bypass. The results with our alternative approach to Scimitar syndrome are summarized and they compare favorably with other published reports.
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Definition and Brief History

The diagnosis of Scimitar syndrome rests on the demonstration of partial anomalous pulmonary venous return to the inferior vena cava (IVC), the inferior cavo-atrial junction, or low on the right atrium. In two thirds of cases the Scimitar vein (SV) provides drainage for the entire right lung, but in one third the SV drains only the lower portion of the right lung with a normally connected upper pulmonary vein. Few reports have documented cases with SVs from the left lung.[1,2]

The SV is typically single, courses anterior to the hilum of the lung, and pierces the diaphragm en route to the IVC where it enters just superior, posterior, and lateral to the right hepatic vein orifice. Considerable variation from the above schema has been noted; SV running a course posterior to the hilum, instances of duplicated SV, and the anomalous entry point has been noted as far afield as the right hepatic vein and the azygous vein. In addition, the SV may be stenotic at or just distal to its junction with the IVC or right atrium in 10% to 20% of cases.[3]

The SV is the sine qua non of this syndrome, but is rarely the only abnormal finding. In descending order of frequency, the following anomalies are associated with the SV: abnormal right lung lobation and right lung hypoplasia (virtually 100%, with widely varying degrees of hypoplasia); dextroposition of the heart; hypoplasia of the right pulmonary artery (60%); systemic arterial blood supply to the right lower lung from the infradiaphragmatic aorta (60%); atrial septal defect (ASD) of the secundum type (40% overall, 80% to 90 % in the infantile variant); right-sided diaphragmatic hernia (15%); and horseshoe lung (parenchymal continuity between the right and left lung behind the heart and anterior to the esophagus). The infantile Scimitar syndrome, in addition to its high incidence of ASD, has an association with a litany of cardiovascular anomalies including ventricular septal defect, patent ductus, hypoplastic aortic arch, coarctation, tetralogy of Fallot, anomalous origin of the left coronary artery, and truncus arteriosus.

SV syndrome is a rare constellation, estimated to occur in two out of 100,000 births, with a 2:1 female preponderance. Hence, the bulk of the literature on this topic is in the form of individual case reports or small case series from large centers with patient numbers in single digits or low teens, with data collected over decades.

This anomaly was first described by Cooper[4] in London in 1836 during an autopsy of an infant. Note was made of dextroposition of the heart and hypoplasia of the right lung in this specimen. In Paris in same year, Chassinat[5] documented similar findings on a necropsy.

The first diagnosis in a live (and asymptomatic) patient was made in 1949 by Dotter et al[6] on cardiac catheterization.

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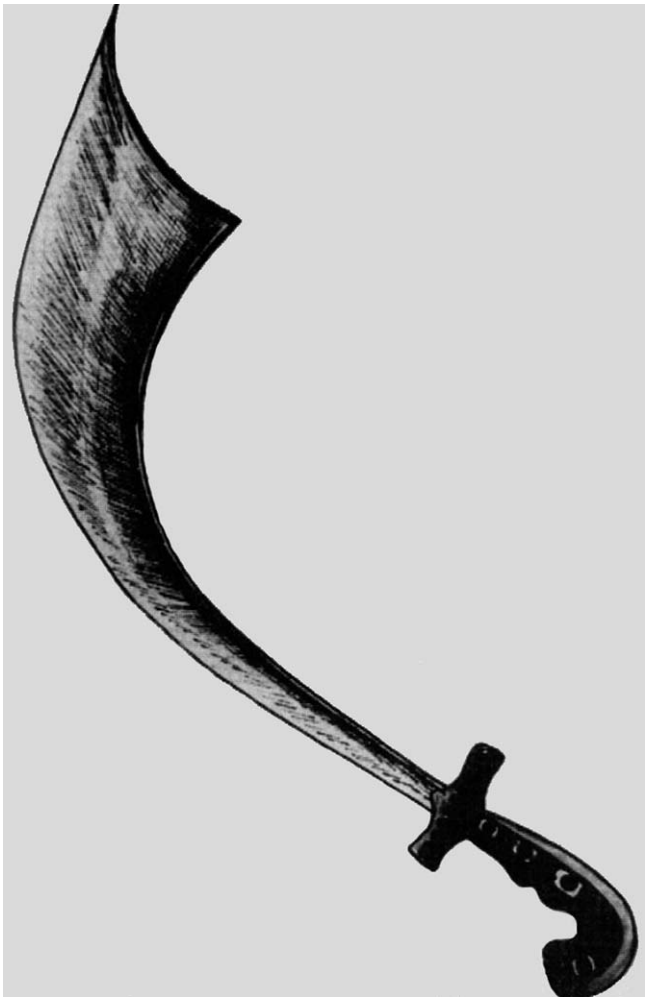

Figure 1 Sketch of scimitar.

Surgery for SV syndrome was first performed in 1950 by Drake and Lynch.[7] They resected the right lower lung in a patient with recurrent right-sided pneumonias who was found to have a SV draining the bronchiectatic lower right lung, with good results.

The first corrective surgery was performed in 1956 by Kirklin et al[8] using cardiopulmonary bypass on a patient with SV and an ASD (the SV was anastomosed to the right atrium in proximity to the ASD) and this portion of the right atrial wall was then sutured to the margin of the ASD, closing the ASD and routing SV flow to the left atrium.

In a 1960 report fro Chile that focused on the radiologic diagnosis of the syndrome, Koch and Silva[9] reported on direct anastomosis of the SV to the left atrium, but no details were provided on the technical aspects of the operation.

The term "Scimitar syndrome" has become firmly affixed to this constellation of anomalies since the 1960 report by Neill et al in the *Bulletin of John Hopkins Hospital* entitled "The familial occurrence of hypoplastic right lung with systemic arterial supply and venous drainage-'Scimitar syndrome'." There is some irony to how pervasive the subsequent use of the term Scimitar syndrome has become for two reasons: first, to our knowledge there are only two other reports of a familial incidence of the syndrome,[10,23] and second, the frontal radiographic finding for which the syndrome is named is only present in approximately half of all patients and less than 10% of infants with the syndrome.

The scimitar (or Turkish sword; Fig 1) sign refers to the half crescent described by the descent of the anomalous pulmonary vein (the tip of the crescent points inferiorly and medially to the diaphragm/right heart border junction). The concavity of the crescent is adjacent to the right heart border. Tributary veins may be seen converging on the SV as the "hilt" of the dagger (Fig 2).

However, this sign is often absent. Reasons cited for the absence include the hypoplasia of the right lung and resultant abnormal rightward positioning of the heart, obfuscating the view of the vein in frontal projection. In infants, the prominent thymic shadow has been implicated in the absence of the scimitar sign.

Embryology and Pathologic Findings

The developmental errors accounting for the observed anatomy in Scimitar syndrome are not understood at present.

It is known that in the course of normal lung development, pulmonary venous drainage to the left atrium is in place by week 11 of gestation. It is also known that as the advancing lung bud develops, its primary blood supply changes from a plexus derived from the post-branchial descending aorta to the portion of the sixth aortic arch that becomes the pulmonary artery,[11] a transition that is complete after the seventh week. Some insult presumably culminates in the failure of this "hand-over," resulting in the observed persistence of systemic arterial supply to the right lung from the abdominal aorta, and the underdevelopment of the right pulmonary artery and right lung. Gross examination of the right lung[12] reveals a small lung with equal incidence of uni-, bi-, and tri-lobar lung. In some cases of bilobar lung, the bronchial pattern is found to be hyparterial, with a mirror image configuration with respect to the larger left lung. Diverticulae or cystic changes of the bronchi have been noted in 20% of specimens.

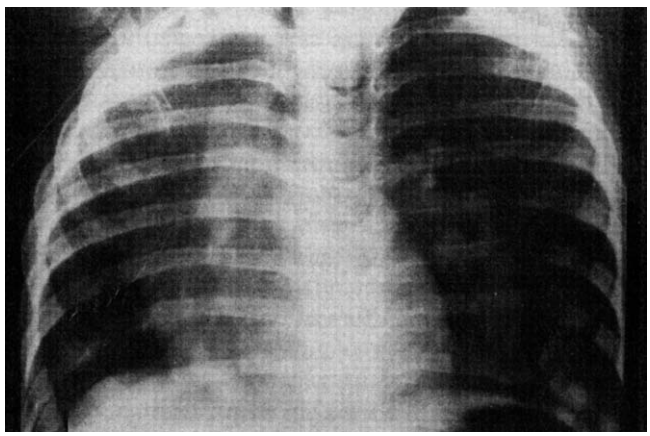

Figure 2 Frontal chest x-ray of a patient with Scimitar vein.