

Scimitar Syndrome

Ulf Gudjonsson and John W. Brown

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 Cooper⁴ 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, Chassinat⁵ documented similar findings on a necropsy.

The first diagnosis in a live (and asymptomatic) patient was made in 1949 by Dotter et al⁶ on cardiac catheterization.

Section of Cardiothoracic Surgery, James W. Riley Hospital for Children and Indiana University School of Medicine, Indianapolis, IN.

Address reprint requests to John W. Brown, MD, Section of Cardiothoracic Surgery, Indiana University School of Medicine, 545 Barnhill Dr, EH 215, Indianapolis, IN, 46202-5123; e-mail: jobrown@iupui.edu

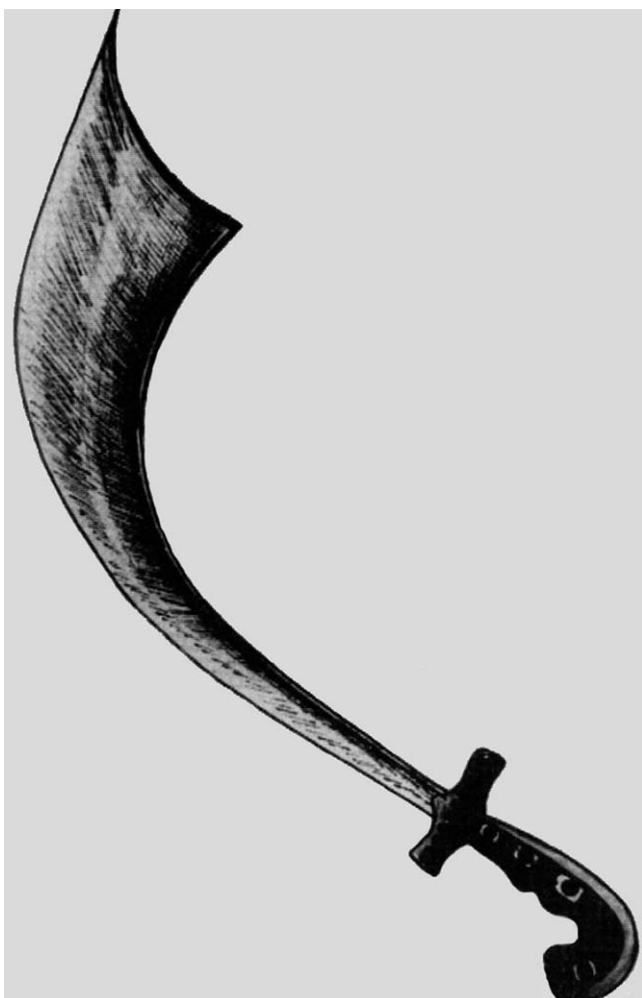


Figure 1 Sketch of scimitar.

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 al⁸ 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 from Chile that focused on the radiologic diagnosis of the syndrome, Koch and Silva⁹ 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 lung¹² 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 hyperarterial, 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.

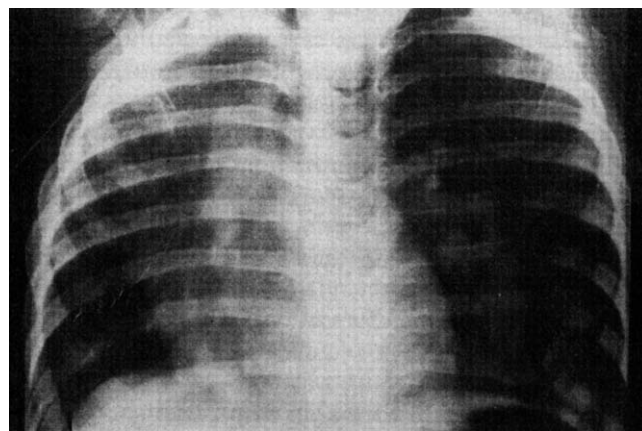


Figure 2 Frontal chest x-ray of a patient with Scimitar vein.

The microscopic appearance of the hypoplastic lung has been reported as normal,¹² while in others a pattern of disorganized bronchioles with absence of alveolar formation has been cited. Infants succumbing with SV show microscopic changes of pulmonary hypertensive arteriopathy.¹

Pathophysiology and Presentation

There are unequivocally two forms of Scimitar syndrome in terms of clinical presentation: (1) an infantile syndrome associated with significant mortality and (2) a child/adult presentation that is a milder form of the syndrome and in fact is frequently asymptomatic, with diagnosis being made incidentally because of radiographic abnormalities. Perusal of the literature reveals similar numbers of patients in each category. Infantile cases are perhaps more likely to be counted because of referral to major centers and their higher associated morbidity and mortality.

As a rule, patients with the infantile syndrome are diagnosed in the first few months of life, the median age at presentation being 2 months. Failure to thrive, tachypnea, and heart failure are the dominant features at presentation in these severely ill patients, although cyanosis may be observed if pulmonary hypertension and the anatomy at hand predispose to right-to-left shunting of blood. Mortality is quoted at 45%.¹¹

Qp/Qs ratio was found to be 3.1 in the series by Huddlestone et al¹³ which focused on 12 infants in St Louis, MO. The mean pulmonary systolic pressure was 74 mm Hg in this series, which is consistent with the other sizeable series of infants, that of Gao et al¹ from Toronto, Canada, which systematically evaluated at the catheterization data, associated anomalies, and outcomes in 13 patients.

The uniformly markedly elevated Qp/Qs and pulmonary hypertension in the infantile syndrome is in sharp contrast to the milder adult variant. The SV, by participating in the inefficient circulation of pulmonary venous blood back to the right atrium, would be expected to increase Qp/Qs, but not to the extent seen in these infants. The flow from the infradiaphragmatic aorta to the lower right lung is a definite factor in increasing Qp/Qs, especially as the aberrant systemic pulmonary artery had a diameter $\geq 40\%$ of the diameter of the infradiaphragmatic aorta, from which it arose in nine of 13 patients in the series by Gao et al.¹ Reports by Gikonyo et al,¹⁴ Huddlestone et al,¹³ and Gao et al¹ document a 75%, 91%, and 93% association, respectively, of the SV with a cardiovascular lesion(s) that would further contribute to increased ineffective pulmonary flow and hypertension. ASD was by far the most common anomaly (80%), followed by patent ductus (75%), ventricular septal defect (30%), and pulmonary vein stenosis (20%). Left-sided obstructive lesions such as subaortic stenosis, aortic coarct, arch hypoplasia, and an infant with Shone's complex were also documented; these findings would be expected to aggravate left-to-right shunting and failure in the presence of septal defects.

Huddlestone and Mendeloff¹¹ point out that although in

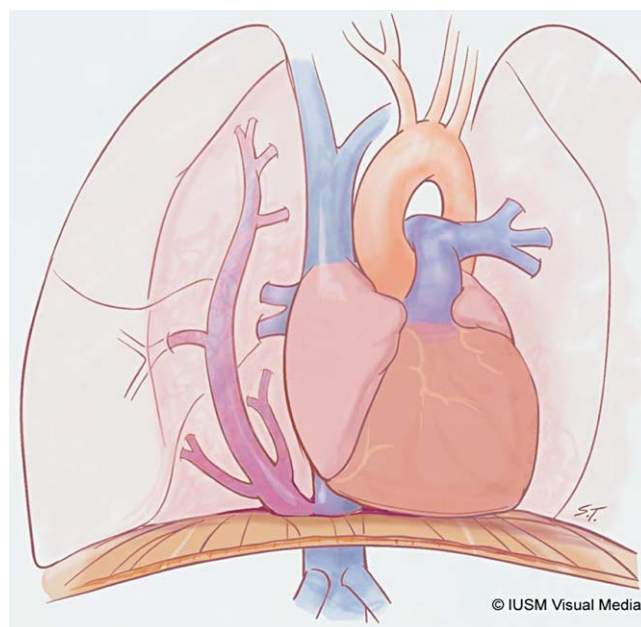


Figure 3 Illustration of a Scimitar vein.

most cases the increased pulmonary flow and resultant hypertension can be blamed on associated lesions, in some infants with relatively mild-appearing anatomic substrate (the kind seen typically with the pediatric/adult syndrome; eg, one symptomatic infant who was found to have left-sided SVC as the only other abnormality other than SV), the patient follows the same morbid course as infants with more severely aberrant anatomy. They postulate that these patients have a pulmonary circulation that fails to adapt normally after birth in the presence of even a mild right-to-left shunt that is associated with the SV and thus present with symptoms while others are able to complete the transition to low pulmonary resistance of the normal infant.

In the pediatric/adult form of the syndrome, symptoms tend to be mild or absent (Fig 3). Frequently, a history of recurrent pulmonary infections involving the right lung can be elicited. Pulmonary function studies show mild deficits in vital capacity and FEV-1 (approximately 80% predicted).¹⁵ The scimitar sign is present in 70% of the pediatric/adult group, and right lung hypoplasia is both less severe and prevalent when compared with the infantile group. Some patients are diagnosed because of atrial fibrillation associated with chronic right-sided overload. Electrocardiography in 50% of cases shows evidence of right ventricular hypertrophy, often in the form of right bundle branch block. Pulmonary artery pressures are normal or only mildly elevated, Qp/Qs averages 2 when several European and North American adult case series are combined.^{3,15,22} The infradiaphragmatic aortic-to-pulmonary branch is absent in half of these cases and often miniscule. ASD occurs in 20% of the pediatric/adult group. Most pediatric/adult series show an equal distribution across the first three decades of life. In our nine-patient series treated via right thoracotomy and without bypass, eight patients were in their first decade and two in their fifth.

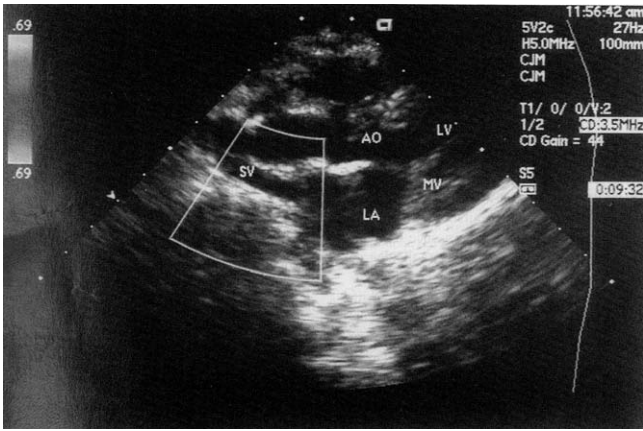


Figure 4 Transthoracic echocardiogram showing patent pulmonary vein/left atrial anastomosis in a patient 2 years after off-pump repair via right thoracotomy.

Evaluation

Patients suspected of having Scimitar syndrome should always have cardiac catheterization to confirm the diagnosis, identify the specific course of the anomalous pulmonary venous drainage, assess stenosis in the SV or pulmonary veins, assess pulmonary arterial anatomy and pressure, degree of left-to-right shunting, and associated cardiac anomalies. Measurement of oxygenation step-up at the site of anomalous drainage may shed light on the value of proceeding with a corrective procedure. Catheterization also allows delineation of the aberrant systemic arterial supply to the right lung, which may be occluded in the catheterization suite using embolization techniques if the clinical scenario demands it.

Ventilation/perfusion scans are recommended as a routine by some investigators¹⁵ both for initial evaluation and follow-up. This group also recommends routine bronchoscopy. Magnetic resonance imaging is a complex and costly diagnostic tool probably not justified as a routine but has been used in Scimitar syndrome¹⁶ to provide excellent images with comprehensive data on hemodynamics, intracardiac anatomy, as well as excellent images of bronchial anatomy and parenchymal abnormalities.

At our institution (Indiana University School of Medicine, Indianapolis, IN), we have relied heavily on echocardiography in our follow-up (Fig 4), specifically to assess the patency of our re-implanted SV as SV or baffle thrombosis in most series is the *Achilles' heel* of the reparative approach.

Treatment

Medical treatment is indicated in the infantile presentation to offset heart failure and allow growth before undertaking surgical repair. However, the presence of pulmonary hypertension or failure of response to medical therapy demands prompt intervention.

At the time of angiography in these infants, the abnormal systemic artery from the abdominal aorta to the right lung should be coil embolized because this intervention may help

and is unlikely to harm the patient. There are several reports in the literature of infants whose deteriorating course was reversed after embolization.^{1,3,12,17,18} In most of these cases, the catheter-based intervention bought valuable time and allowed for a repair under elective conditions. In the series by Gao et al,¹ two patients under 2 months of age had the need for further intervention delayed until they reached 1 year of age. Huddleston et al¹³ had two patients in their series who had embolization without appreciable effect on pulmonary artery pressures or left-to-right shunt, which indicates that only cautious optimism is justified with regards to the results of embolization, with more benefit likely the larger the aberrant artery being coiled. The pulmonary tissue subtended by the abnormal artery seems to be protected from infarction, perhaps via bronchial or pulmonary artery collaterals, as only one case of pulmonary infarction has been documented after coiling.¹

In adults, operation is advised for symptoms of recurrent lung infection or Qp/Qs greater than 1.5 in an asymptomatic patient, with similar rationale (except paradoxical embolus and reversal of left-to-right shunt should not be a concern without intracardiac shunt) in effect when advocating ASD repair in an adult, to avoid the sequelae of chronic right heart overload.

Dupuis et al²³ cast doubt on the utility of corrective surgery in adults with Scimitar syndrome. Their review of 37 patients treated surgically versus 85 patients who were observed showed that only 12 surgical patients were reported to have a “good” result; whereas four patients died and 21 had thrombosis of the SV-to-left atrium connection, presenting with lung infarction, pulmonary hypertension, or hemoptysis, all requiring lung resection. Patients who were simply observed had a “good” course in 79 of 85 cases and were felt to lead normal lives. The counterpoint to this argument is that less

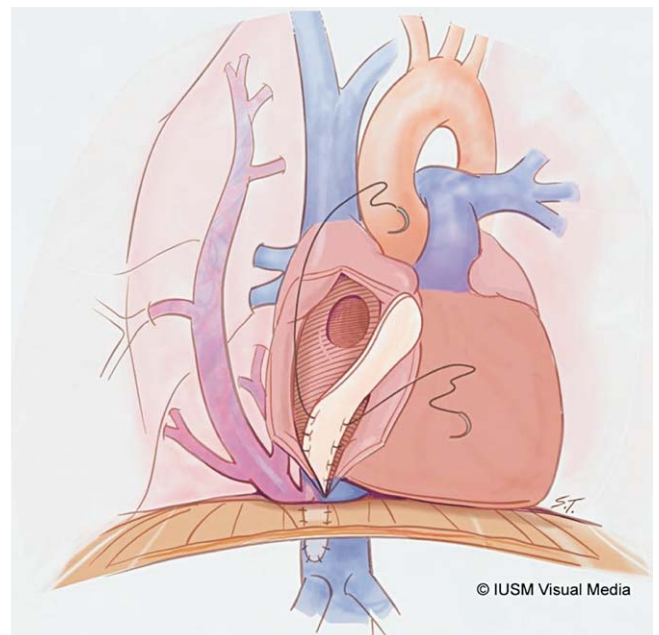


Figure 5 Correction of Scimitar syndrome with a long baffle from IVC to ASD.

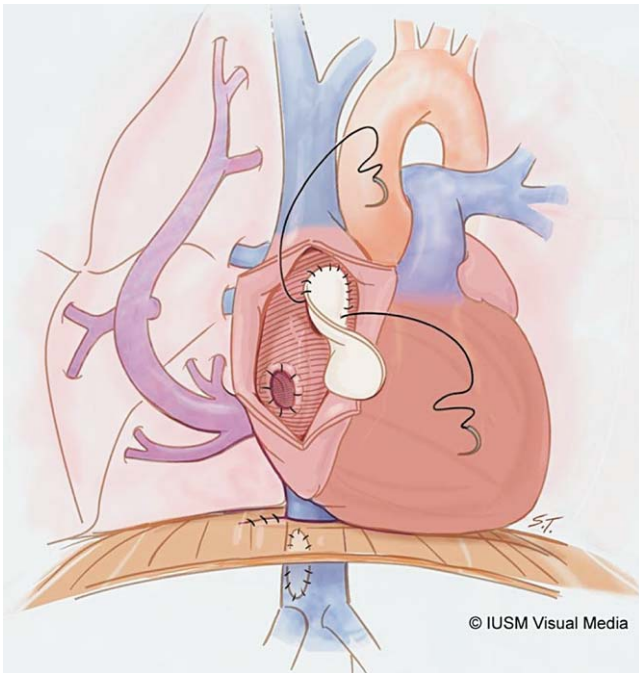


Figure 6 Repair of Scimitar syndrome with reimplantation of SV in the right atrium and use of a short baffle.

than half of the observed patients were followed for more than 10 years, and that the observed rate of SV thrombosis in this series is quite high.

In broad terms, the physiologic aberration of Scimitar syndrome may be surgically corrected in two ways. Either resection of the right lung drained by the SV, or by a corrective approach rerouting SV flow to the left atrium. In either approach, any abnormal systemic arteries to the right lung are ligated and intracardiac lesions will require standard repair.

The abnormal lobation of the hypoplastic lung often makes any resection short of pneumonectomy difficult. Sur-

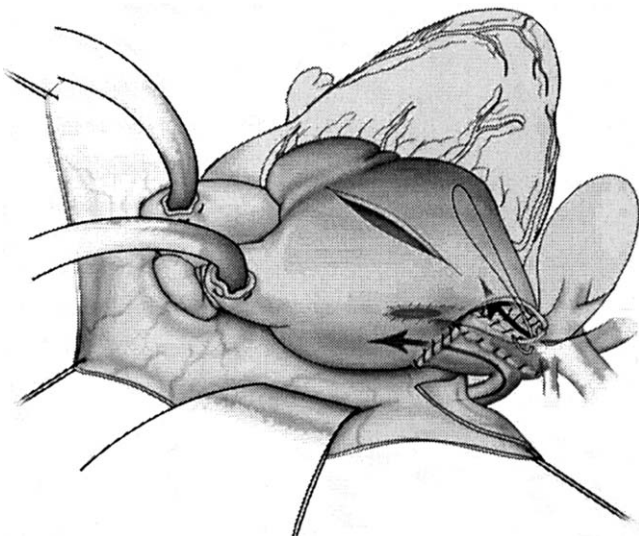


Figure 7 Completed repair of the type advocated by Calhoun and Mee.²²

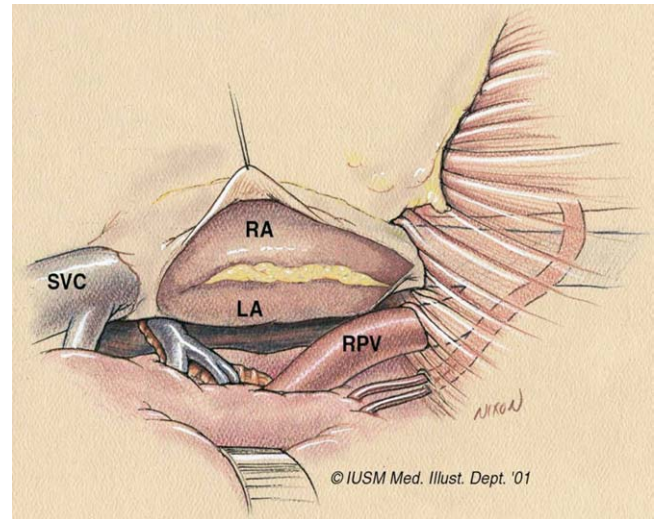


Figure 8 Indiana University modification of SV repair via right thoracotomy and without the use of cardiopulmonary bypass.

geons loathe resecting viable lung tissue, and pneumonectomy is not a popular option, but it has its place; an acceptable example would be in a patient with a diffusely scarred bronchiectatic lung. Huddleston et al¹³ are of the opinion that a posteriorly located SV in an infant may be impossible to reroute to the left atrium without kinking and thrombosis. Four patients in their series report good long-term results at a median 9-year follow-up after pneumonectomy. Postpneumonectomy concerns in infants include scoliosis, respiratory insufficiency, and postpneumonectomy syndrome where the mediastinal contents and left lung rotate posteriorly into the post-pneumonectomy space and cause tracheo-bronchial obstruction and malacia. However, postpneumonectomy syndrome is rare and treatable and the scoliosis and reduction in lung capacity do not seem to be significant issues

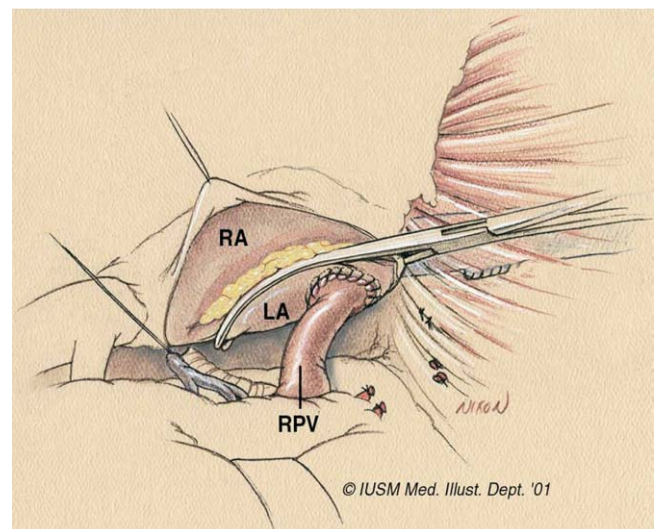


Figure 9 Completion of repair of SV-Indiana University modification. SV has been implanted into the left atrium and the clamp is about to be removed from the left atrium

Table 1 Comparison of Results of Treatment of Scimitar Syndrome in Five of the Larger Series in the Literature

Study	No of Patients	Overall Mortality	Medical Therapy	Occlusion of Anomalous Syst. Artery	PDA Ligation/ CoA Repair	Complete Repair	Pulmonary Resection
Huddleston (1999)	12	4	2 (2)	1 (1)	0 (0)	7 (1)	5 (0)
Najm (1996)	32	5	7 (1)	5 (3)	2 (2)	17 (0)	1 (0)
Dupuis et al ²³ (1993)	25	16	10 (7)	6 (1)	5 (5)	0 (0)	3 (2)
Torres (1993)	14	1	0 (0)	3 (1)	0 (0)	10 (0)	1 (0)
Indiana University (2005)	10	0 (0)	0 (0)	3 (0)	0 (0)	10 (0)	0 (0)

Abbreviation: PDA, patent ductus.

in the 30-year follow-up study performed by Laros and Westermann.¹⁹

There are several surgical corrective options for Scimitar syndrome. One method for correction was described by Zubiate in 1962²⁰ that involved creating a synthetic patch or pericardial tunnel baffling flow from the orifice of the SV in the IVC through the right atrium and then suturing the baffle to the sides of the ASD to deliver SV blood to the left atrium. Deep hypothermic circulatory arrest is used to allow exact SV orifice visualization and avoidance of hepatic veins without interference from an IVC cannula. Drawbacks to this approach (other than the need for deep hypothermic circulatory arrest) include inability to fix stenosis of the SV and the fact that blood is baffled through a long tunnel 180° into the left atrium, which predisposes this repair to thrombosis, especially in infants (Fig 5). The IVC may also need a patch enlargement.

Shumacker and Judd²¹ advocated excising the SV orifice with a cuff of IVC or right atrium and reimplanting it on the atrium adjacent to a pre-existing ASD or by creating an ASD and baffling flow across to the left atrium. This approach returns blood more directly to the left atrium. SV stenosis can be addressed with spatulation. The use of IVC or right atrium margin provides more robust tissue for anastomosis than the fragile SV (Fig 6). The IVC is typically patched to avoid stenosis.

Calhoun and Mee²² devised an approach for infants where the IVC is transected under deep hypothermic circulatory arrest and divided into posterior (SV blood) and anterior (IVC, hepatic veins) compartments with a flap of pericardium (Fig 7). An incision is then made into the inferior aspect of the left atrium and carried to the atrial septum, and the posterior half of the IVC is sutured to the left atriotomy and the pericardial patch is used to close the septal defect. The anterior lip of the IVC is then sewn to the right atrium. The purported advantage of this approach is again that SV blood is baffled through a relatively gentle angle to the left atrium.

An approach we have used with good results involves a fifth interspace right lateral thoracotomy with no need for baffles or bypass (unless an intracardiac lesion also needs to be fixed) (Fig 8). The right lung is freed from adhesions, the SV is dissected out in its thoracic course to allow for mobilization, the inferior pulmonary ligament is taken down and any abnormal systemic arteries coursing through it are ligated and divided. The diaphragm is opened and the SV freed

all the way to the IVC junction. Opening the pericardium posterior to the phrenic nerve and taking down pericardial attachments around the IVC improves exposure of the SV-IVC confluence.

The right pulmonary artery is circumferentially dissected and encircled with a vessel loop. The patient is heparinized and, with an ACT in excess of 400 seconds, the pulmonary artery and SV are occluded. We have cooled the lung with ice slush and a topical pad. A curved vascular clamp is placed on the IVC at the SV entrance and the SV divided from the IVC with a caval cuff. The opening in the IVC is then closed with a running absorbable monofilament suture.

A window of pericardium is then excised posterior to the phrenic nerve and through this a generous portion of left atrium is exposed through dissection in the interatrial groove; a partial occluding clamp is then placed across a swathe of the posterior, right-sided aspect of the left atrium, which is then opened along the axis of the clamp and then sutured to the spatulated end of the SV in an end-to-side fashion with absorbable 6-0 monofilament (Fig 9). Spatulation may need to be more aggressive if a SV stenosis is present, but we have not encountered problems with undue tension on our repairs in these instances.

We have been very satisfied with how the operation “sets up” in the left lateral decubitus position and the contour of the SV reimplantation through this approach. To date, none of our patients in a series of 12 has had stenosis of the reimplanted vein at a mean echocardiographic follow-up of 5 years (Table 1). We have not had to reoperate on any of these patients and have had no mortality in our series to date. On the basis of this information, we would strongly contest the opinion of Dupuis et al²³ and advise intervention on all asymptomatic patients with Qp/Qs in excess of 1.5, barring some compelling comorbidity that would be expected to elevate the patient’s risk for surgery or reduce long-term survival. We make this recommendation because we would expect the natural history of these patients to parallel that of patients with unrepaired ASD with significant shunt in respect to the late (fourth and fifth decade and beyond) development of right heart failure, atrial arrhythmias, and pulmonary hypertension.

We believe this approach also lends itself well to the repair of posteriorly located SV, a situation for which some have suggested pneumonectomy. We would tackle patients with SV and ASD in an identical manner, except for a run of bypass

for the ASD closure. In our series, we have one patient with infantile presentation that was treated successfully with a staged repair, that is to say ligation of abnormal systemic to pulmonary arteries which was successful in alleviating pulmonary hypertension as a neonate followed by repair of SV through right thoracotomy (as just described) at 2 years of age. In our view, the contraindication to a right thoracotomy approach would be in a patient who has an associated lesion requiring a sternotomy (eg, anomalous origin of the left coronary).

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