

DIAGNOSTIC AND THERAPEUTIC CHALLENGES OF INFANTILE SCIMITAR SYNDROME IN A LOWER-MIDDLE INCOME COUNTRY: A CASE SERIES FROM PAKISTAN

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DOI: <https://doi.org/10.5281/zenodo.21213704>

Keywords

Article History

Received: 25 April 2026

Accepted: 04 June 2026

Published: 21 June 2026

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Abstract

Background: Infantile Scimitar syndrome (SS) is a rare, severe congenital anomaly involving anomalous right pulmonary venous drainage into the inferior vena cava (IVC). It poses significant diagnostic and management challenges in lower middle-income countries (LMICs) due to symptomatic overlap with endemic diseases.

Case Series: This series details three distinct presentations of infantile SS in Pakistan: **Case 1:** A 7-month-old female, initially misdiagnosed and treated empirically for pulmonary tuberculosis, was diagnosed with SS and an atrial septal defect (ASD). She underwent a successful intra-atrial pericardial patch baffle (Roger Mee procedure) at age two, demonstrating favorable long-term survival (>10 years). **Case 2:** A male neonate presented with severe persistent pulmonary hypertension (PPHN) and was diagnosed with SS associated with a rare horseshoe lung anomaly. Due to delayed treatment and parental refusal of timely admission, the patient clinically deteriorated and died at seven months. **Case 3:** A preterm male neonate presenting with dextroposition, severe right lung hypoplasia, and pulmonary sequestration underwent a successful life-saving right pneumonectomy on the 22nd day of life and was stabilized. **Conclusion:** Infantile SS exhibits high clinical variability and carries a poor prognosis if treatment is delayed. Advanced multimodality imaging is vital to avoid critical misdiagnoses, and early, tailored surgical interventions ranging from baffle repair to pneumonectomy are essential for survival.

Keywords: Scimitar syndrome; congenital heart disease; Baffle repair; Pneumonectomy; Pakistan.

1. Introduction

Scimitar syndrome (SS) is a rare, congenital cardiovascular anomaly. It is also referred to as Halasz syndrome, congenital venolobar syndrome, vena cava bronchovascular syndrome, mirror-image lung syndrome, and hypogenetic lung syndrome. (1) The characteristic feature is

drainage of the right pulmonary vein (RPV) into the inferior vena cava (IVC) instead of the left atrium, leading to a left-to-right shunt. (2) This is often accompanied by varying degrees of hypoplasia of the right lung and the right pulmonary artery, dextroposition of the heart, and abnormal systemic blood supply to the right lung

through a collateral branch of the descending aorta. (1,3) It is also associated with atrial septal defects (ASDs) and aortopulmonary collaterals. (1) Respiratory distress, tachypnea, recurrent pneumonia, cyanosis, and heart failure are common symptoms.

The name of the syndrome arises from the radiographic appearance of the anomalous, crescent-shaped RPV adjacent to the cardiac silhouette (Scimitar sign). (4) This resembles a curved Turkish sword called a "Scimitar". (1) Although it is reported in only 1-3 cases per 100,000 live births, the actual incidence is likely underestimated because many patients remain asymptomatic and are not diagnosed. (5)

SS can be divided into adult and infantile forms, with the latter being more severe and with higher rates of mortality. (6) Management of the syndrome requires a multidisciplinary approach, ranging from medical to surgical interventions. Surgical interventions also vary for each case and according to the preference of the surgeon. (1)

Since cases of SS have been rarely [documented in Pakistan](#), we present a case series of 3 patients who were diagnosed at our hospital.

2. Case Presentations

2.1 Case 1

2.1.1 Primary Presentation

A seven-month-old female infant, with a weight of 6.2 kg and a height of 68 cm, was brought to the emergency department with a one-week history of fever and cough, a two-day history of loose stools and vomiting, and a three- to four-month history of recurrent chest infections. Her parents had a consanguineous marriage, and she was born full-

term via cesarean section. She remained asymptomatic until four months of age. Due to the recurrent respiratory symptoms, she had been misdiagnosed as a case of pulmonary tuberculosis (TB) in her hometown and had been empirically treated as such.

On examination, the infant was in respiratory distress with reduced oral intake. Cardiovascular assessment revealed a hyperdynamic precordium with a pansystolic murmur audible at the lower left sternal border.

2.1.2 Diagnosis

On chest x-ray, a linear, serpiginous opacity was observed over the mid and lower right thorax, most likely corresponding to an anomalous venous channel draining the right lung.

Echocardiography demonstrated partial anomalous pulmonary venous return (PAPVR), with the RPV draining into the IVC, as well as a 7-8 mm ASD. Significant dilatation of the right heart chamber and the IVC was also seen.

Subsequent CT angiography confirmed these findings and also identified an abnormal systemic arterial supply from the abdominal part of the descending aorta to the right lower lobe of the lungs. A second small arterial branch arising from the aorta at the level of the diaphragm (at T9) also supplied the right lower lobe. Multiple venous channels from the right upper and lower lobes coalesced into a large vein that coursed through the right diaphragm and drained into the intrahepatic IVC. (refer to Fig 1)

Based on these findings, a diagnosis of SS with associated ASD was established.

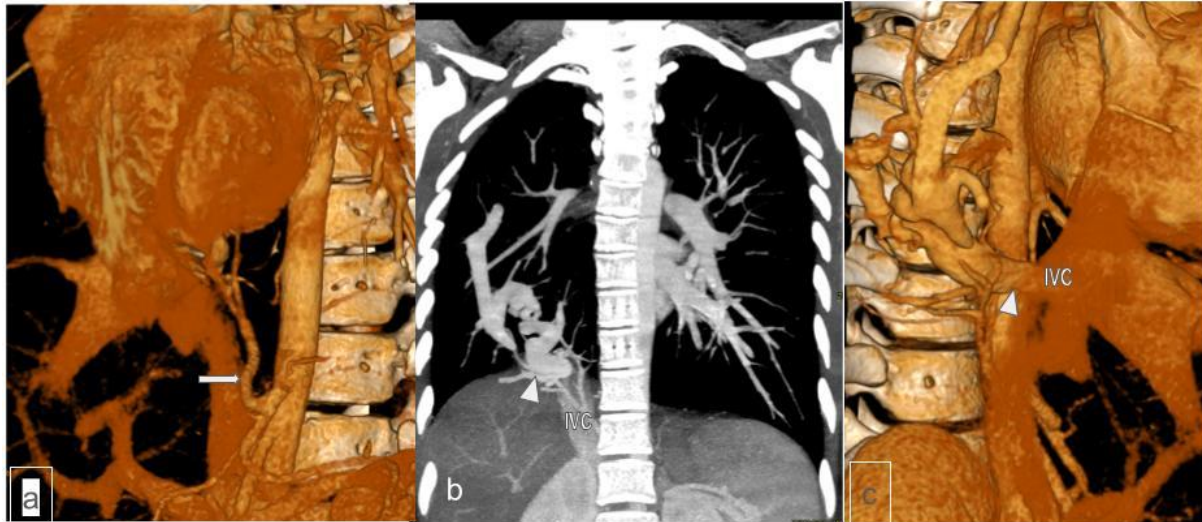


Fig1, a, showing arterial supply of the right lower from from celiac trunk (arrow) and venous drainage (arrow head) into the IVC (b,c)

2.1.3 Management

The infant was medically stabilized and discharged on a regimen of spironolactone, captopril, furosemide, and antibiotics.

At the age of two years, the patient presented to the outpatient clinic with recurrent chest infections. A corrective surgical procedure was planned and performed two months later. The patient underwent a total repair of the PAPVR and closure of the ASD via median sternotomy.

The anomalous venous drainage (Scimitar vein) was rerouted to the left atrium through an intra-atrial pericardial patch baffle (Roger Mee procedure). The IVC was transected, the posterior wall of the left atrium was incised, and a baffle was constructed to partition the IVC and re-establish the posterior wall between the IVC and left atrium. The right atrium was then anastomosed to the IVC, and the ASD was closed using the same baffle. A small foramen ovale was intentionally left patent. Postoperatively, the patient's recovery was uneventful.

2.1.4 Future Course

Approximately nine years after the surgery (age: 11.5 years), the patient presented to the outpatient

clinic with complaints of palpitations, night wakening, and shortness of breath. Another two years later, she again presented with complaints of palpitations for the past six months. She also experienced episodes of tachycardia and sweating, 1-2 times a week. These episodes were accompanied by her skin turning yellow. This was the most recent follow-up, and the patient is alive and undergoing regular clinical visits.

2.2 Case 2

2.2.1 Birth History and Presentation

A male neonate was born at term to a primigravida mother via an emergency cesarean section due to meconium grade III meconium-stained liquor. The mother also tested positive for hepatitis B. The neonate had a good APGAR score (7 at 1 minute; 9 at 5 minutes) at birth but subsequently developed respiratory distress and was managed for meconium aspiration syndrome for three days before being discharged.

At 17 days of age, the neonate was admitted to the NICU with severe respiratory distress and refusal to feed. Echocardiogram showed severe pulmonary hypertension, and he was managed for persistent pulmonary hypertension of the

newborn (PPHN) with mechanical ventilation, dopamine, and sildenafil. After improvement, sildenafil was stopped because a left-to-right shunt was seen on a subsequent echocardiogram. The neonate was transitioned to furosemide and extubated onto high-flow support. However, his respiratory status deteriorated, leading to re-intubation.

2.2.2 Diagnosis

On chest x-ray, cardiomegaly with bilateral haziness of the lung fields was observed, which may be secondary to infection.

Echocardiography demonstrated small apical muscular ventricular septal defects, a small patent ductus arteriosus, anomalous right-sided pulmonary venous drainage into the IVC, a patent foramen ovale, and severe pulmonary arterial hypertension.

Subsequent CT angiography confirmed cardiomegaly and identified anomalous venous drainage of the right lung into the infracardiac portion of the IVC via a large tubular vascular channel. A tortuous systemic vessel originating from the descending aorta was seen supplying the right lung. Additionally, a thin, thread-like pulmonary vein from the right upper lobe drained into the left atrium, measuring less than 1 mm. The basal portion of the left lung also exhibited anomalous venous drainage to the IVC with near-complete collapse of the left lung and partial collapse of the posterior basal segments of the right upper and lower lobes. A parenchymal band connecting the right and left lungs was identified, consistent with a horseshoe lung. (refer to Fig 2) Based on these findings, a diagnosis of SS with associated horseshoe lungs was established.

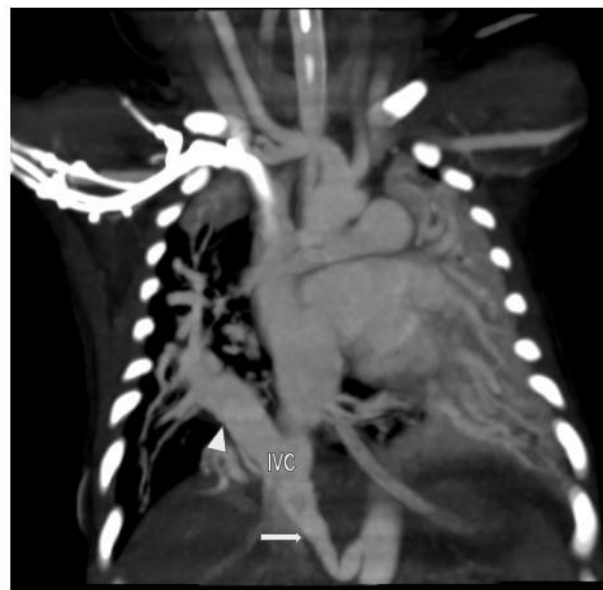


Fig2. showing arterial supply of the right lower from from celiac trunk (arrow) and venous drainage (arrow head) into the IVC

2.2.3 Management and Future Course

The neonate was treated medically and was discharged once he stabilized. However, two days later, he returned with acute respiratory distress and was re-admitted to the NICU. He was intubated and treated medically. Right heart catheterization confirmed the earlier diagnosis of

PPHN. His condition stabilized, and he was discharged after six days.

Three months later, the infant presented again with acute respiratory distress and an oxygen saturation of 70%. Given the diagnosis of SS with PPHN, inpatient admission for further management was advised. However, the family

declined admission and chose to leave against medical advice.

Another two months later, the infant presented with a four-day history of fever and cough and was admitted in a gasping state. He was intubated and placed on mechanical ventilation. After initial improvement and extubation, he developed recurrent respiratory distress, tachycardia, and oliguria, requiring re-intubation and inotropic and diuretic support. Blood cultures grew gram-negative organisms. Despite maximal ventilatory and inotropic management, he developed pneumothorax requiring chest drainage. His condition worsened; he suffered cardiac arrest and was declared dead at the age of approximately seven months.

2.3 Case 3

2.3.1 Birth History and Presentation

A male neonate was born late preterm at 35 weeks of gestation to a primigravida mother via emergency cesarean section due to a suspicious cardiotocograph, with a weight of 2.7 kg and a height of 50 cm. His APGAR score at one minute

was 8. On the first day of life, he developed tachypnea, and clinical examination was notable for right-sided heart sounds, raising suspicion of dextrocardia or right lung collapse. He was shifted to the NICU and intubated.

2.3.2 Diagnosis

On chest x-ray, a complete collapse of the right lung was seen.

Echocardiography demonstrated a dextroposed heart with a hypoplastic right pulmonary artery, a small patent ductus arteriosus, and trace pericardial effusion.

Subsequent CT angiography revealed the RPVs draining into the IVC, with a hypoplastic right lung. The right lower lobe of the lung received an anomalous systemic arterial supply from the subdiaphragmatic aorta, consistent with pulmonary sequestration. Additionally, diffuse ground-glass opacities were noted in multiple segments of the right lower lobe. (refer to Fig 3)

Based on these findings, a diagnosis of SS was established.

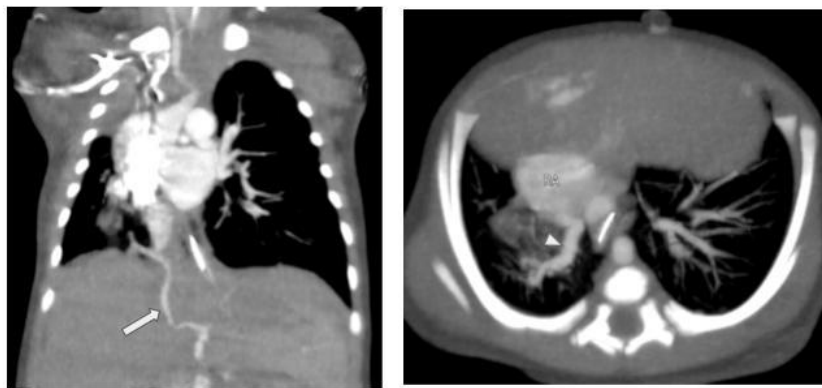


Fig3. showing arterial supply of the right lower from from celiac trunk (arrow) and venous drainage (arrow head) into the RA.

2.3.3 Management and Future Course

It was decided that if the neonate could be successfully extubated, conservative management would be pursued. However, on the tenth day of

life, he failed extubation and his condition deteriorated, marked by worsening respiratory distress and cyanosis. This necessitated re-intubation. Due to the severe lung hypoplasia and

sequestration leading to refractory respiratory failure, a decision was made to proceed with surgical intervention.

On the twenty-second day of life, the patient underwent a right pneumonectomy and placement of a peritoneal dialysis (PD) catheter via a right posterolateral thoracotomy. During the procedure, the right inferior pulmonary vein (scimitar vein), superior pulmonary vein, and the systemic arterial feeder were divided and oversewn. The hypoplastic right main pulmonary artery and right main bronchus were similarly divided, and the stumps were oversewn and buried. The post-operative course was complicated by transient bradycardia, but the patient’s vitals eventually stabilized.

Regular chest physiotherapy was conducted. Antibiotic therapy was also administered. During the removal of the PD catheter, evisceration of the bowel occurred. An emergency laparotomy and bowel reduction with wound closure were performed. After stabilization, the infant was extubated and was eventually discharged following step-down care and caregiver training.

One month following discharge, the patient was brought to the clinic with a new-onset umbilical hernia.

3. Discussion

This case series highlights the unique presentations and treatment options for SS, a rare congenital cardiovascular anomaly. The main feature of this syndrome is PAPVR of the right lung to the IVC, which is accompanied by hypoplasia of the right lung and the right pulmonary artery, dextroposition of the heart, and abnormal systemic blood supply to the right lung from the descending aorta. (1) Patients with SS also commonly exhibit congenital abnormalities such as ASDs, ventricular septal defects (VSD), patent ductus arteriosus (PDA), patent foramen ovale (PFO), and horseshoe lung. SS was first described by George Cooper (1) in 1836, but the name was given by Catherine Neill (7) in 1960 because the radiographic appearance resembled a curved Turkish sword called a “Scimitar”.

Our series describes three patients, all of whom presented with the core signs of infantile SS. However, we observed variation in the associated defects, management strategies, and outcomes. The defects found on radiological modalities (X-ray, echocardiography, and CT angiography) for each patient are shown in Table 1.

Table 1

Patient	Right pulmonary vein draining into the inferior vena cava (also called PAPVR)	Right lung hypoplasia	Right pulmonary artery hypoplasia	Abnormal systemic blood supply to the right lung	Dextroposition of the heart	Cardiomegaly	Persistent pulmonary hypertension of the newborn (PPHN)	Atrial septal defect (ASD)	Ventricular septal defect (VSD)	Patent ductus arteriosus (PDA)	Patent foramen ovale (PFO)
Case 1	+	-	-	+	-	-	-	+	-	-	-

Case 2	+	+	-	+	-	+	+	-	+	+	+
Case 3	+	+	+	+	+	-	-	-	-	+	-
Key: + present; - absent											

SS presents a significant diagnostic challenge as its clinical manifestations can overlap with other respiratory illnesses, leading to misdiagnosis. This is exemplified by the patient in case 1, whose history of recurrent chest infections resulted in the diagnosis and empirical treatment of pulmonary TB. A similar misdiagnosis has also been reported in Ethiopia, another region with a high prevalence of TB, where a 20-year-old female was misdiagnosed as a case of TB. (8) These errors highlight the importance of utilizing imaging studies to reach a diagnosis, to prevent the use of unnecessary medications, and to avoid delays in the appropriate management of SS.

The initial suspicion of SS is raised by chest radiography, which often reveals variable opacification of the right hemithorax along with dextroposition of the heart, findings that were also observed in our patients. (9) The characteristic “scimitar sign” is diagnostic (10), but is usually obscured due to the opacity and hence, more advanced imaging is required for a definitive diagnosis.

Echocardiography is a useful technique used to identify the anomalous connection of the right pulmonary vein (the scimitar vein) with the IVC, and abnormal systemic blood supply to the right lung. (11) It also helps assess pulmonary hypertension (as seen in case 2) and associated cardiac defects, such as ASD, VSD, PDA, and PFO. Additionally, CT angiography accurately shows the presence and anatomical course of anomalous veins, associated cardiovascular defects, and any other lung or vascular anomaly. (12,13) Particularly in the patient in case 2, CT angiography revealed the presence of horseshoe lung, another congenital anomaly that has been associated with SS. (14) These advanced modalities especially assist in visualizing the complete vascular network, which identifies the

abnormal systemic arterial supply, which was observed in all three patients in our series.

Management of SS is unique to the patient’s clinical presentation and the specific anatomical defects, but it often requires a multidisciplinary approach, usually involving pediatrics, cardiology, radiology, and cardiothoracic surgery. Patients presenting with heart failure or pulmonary hypertension require immediate treatment and are initially managed medically with diuretics and inotropic support. Nonetheless, surgical correction is the definitive treatment in most cases. This was evident in case 2, where parent refusal of hospital admission for further management resulted in rapid deterioration of the infant and ultimately death upon delayed presentation.

The optimal surgical treatment for SS remains a debate. (15) In case 1, an intra-atrial pericardial patch baffle was used to connect the scimitar vein with the left atrium, while also closing the ASD. This approach is referred to as the Roger Mee procedure and was first described by Calhoun et al. (16) in 2003 to correct SS in infants due to the short course of the scimitar vein. Our surgical outcome supports the use of this technique, as the patient has spent more than 10 years since the surgery. In case 3, the right lung was hypoplastic and sequestered, making it nonfunctional. Therefore, a right pneumonectomy was performed to remove the right lung and eliminate the left-to-right shunting. This has also been described as a treatment option in multiple studies with good outcomes. (17,18)

The prognosis of infantile SS is known to be poor, even after surgical correction. A retrospective review reported a 1-year survival rate of 62.5% in surgically treated infants with symptomatic SS, compared to 54.5% among those managed conservatively. (5) In our series, case 1 demonstrated favorable long-term survival

following timely and appropriate surgical intervention. Similarly, case 3 also underwent successful surgical treatment, although long-term outcomes remain to be determined given the recent presentation and treatment. In contrast, the fatal outcome observed in case 2 highlights the potential consequences of delaying treatment. This case also highlights the need for effective parental counselling in such situations.

4. Conclusion

This case series illustrates the variability in clinical presentation and management of infantile SS, while also highlighting the necessary imaging modalities for a timely diagnosis. The poor prognosis, especially in the case of delay in treatment, is also depicted, and two unique surgical treatment options are highlighted based on the difference in presentation of the infants.

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