Complete transcatheter correction of variant scimitar syndrome—a case report

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Received 18 March 2022; first decision 22 June 2022; accepted 2 November 2022; online publish-ahead-of-print 4 November 2022

Background
Scimitar syndrome (SS) comprises of an anomalously draining right pulmonary vein (APV), to the inferior vena cava (IVC), maldevelopment of the right pulmonary artery (RPA), and the right lung, with variable number of aorto-pulmonary collaterals (APC) to the right lung. It can cause pulmonary hypertension if left untreated. Surgical correction is the method of choice. We report a case of variant SS with dual drainage of the APV to the IVC and left atrium (LA) that was addressed with a transcatheter approach.

Case summary
A 13-year-old child was evaluated for dyspnoea. Chest x-ray and transthoracic echo (TTE) were suggestive of SS with an additional central atrial septal defect (ASD). Cardiac computed tomography (CT) revealed dual drainage of the APV to the IVC and via a meandering vein to the LA and three APCs. The ASD was closed, and the APCs were coiled. The connection of the APV to the IVC was closed with a device, rerouting the pulmonary vein blood to the LA. The child is doing well on follow-up after 2 years.

Discussion
Variant forms of SS are rare. Our case had ASD, multiple APCs, well-developed RPA and right lung and a dual drainage of the APV. This allowed for transcatheter management. Otherwise, surgery is the default choice. Multimodality imaging with TTE, CT, magnetic resonance imaging, and cardiac catheterization will help in diagnosis and anatomical delineation.

Keywords
Case report • Pulmonary veins • Heart defects • congenital

ESC Curriculum
2.2 Echocardiography • 7.4 Percutaneous cardiovascular post-procedure

Learning points
• Variant presentations of scimitar syndrome with dual drainage of the pulmonary veins to IVC and LA need multimodality imaging for diagnosis and can be addressed with complete transcatheter management.
• These cases may be asymptomatic and often detected incidentally, but rerouting of the pulmonary vein to LA is essential to avoid development of pulmonary hypertension.

Introduction
Scimitar syndrome is a rare association of cardiovascular defects—encompassing either total or partial anomalous right pulmonary vein (APV) drainage to the inferior vena cava (IVC), hypoplastic right pulmonary artery (RPA), an underdeveloped right lung, dextroposed heart, and multiple aortopulmonary collaterals (APCs). It can present variably: in infants, with intractable pulmonary hypertension, and cardiac failure, and in adults, with dyspnoea, recurrent lung infections, or it is detected incidentally.¹ Scimitar syndrome is managed surgically, with baffling of the APV(s) to the left atrium (LA). An additional atrial septal defect (ASD) can be seen in 25% of these patients.² Sometimes, there are scimitar variants—with dual drainage of the right lower pulmonary vein to both IVC and the LA.³ We present a case of a
variant scimitar syndrome where the constellation of defects had a favorable anatomy and allowed for complete transcatheter management.

**Timeline**

| Date        | Event Description                                                                 |
|-------------|-----------------------------------------------------------------------------------|
| July 2018   | Presentation to hospital. Chest x-ray, ECG, and transthoracic echo were done, and the diagnosis was made. |
| August 2018 | Cardiac CT performed for anatomic delineation and planned for transcatheter intervention. |
| September 2018 | Transcatheter closure of ASD, coiling of collaterals, and device closure of the right lower pulmonary vein (RLPV) to inferior vena cava junction performed, rerouting the RLPV to left atrium. |
| September 2021 | Asymptomatic, 3 years follow-up. The child has normal pulmonary artery pressures and is thriving well. |

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**Case presentation**

A 13-year-old child with no past medical history was evaluated for dyspnoea, noticeable over 1 year. He was acyanotic with stable vital signs. There were palpable pulsations in the left second space. Second heart sound was widely split and fixed with a normal pulmonary component. A 2/6 ejection systolic murmur and a mid-diastolic flow murmur were heard over the pulmonary and the tricuspid areas, respectively. Chest X-ray (Figure 1A) showed a dextroposed heart and a hyperdensity parallel to the right heart border, consistent with a scimitar syndrome. Electrocardiogram showed a right axis deviation and partial right bundle branch block (Figure 1B). Transthoracic echocardiogram (TTE) revealed a 14 mm ostium secundum ASD with a left to right shunt and an anomalously draining right lower pulmonary vein (RLPV) to the junction of IVC and the right atrium (RA), with mild turbulence (Figure 1C). There was mild tricuspid regurgitation with mildly elevated pulmonary artery systolic pressure (PASP). There was no significant gradient at the RLPV → IVC junction.

A cardiac computed tomography (CT) scan corroborated the findings and additionally demonstrated a normal right bronchus without pulmonary sequestration and a well-developed RPA. The RLPV was noted to have dual drainage to the LA (via a meandering vein) and IVC-RA junction with a constriction (Figure 1D). Other pulmonary veins

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**Figure 1** Diagnosis. (A) Chest X-ray showing situs solitus with a dextroposed heart and a right para cardiac shadow—the ‘scimitar sign’. (B) Electrocardiogram demonstrating incomplete right bundle branch block. (C) Two-dimensional transthoracic echocardiogram showing the right lower pulmonary vein draining into the inferior vena cava. (D) Coronal computed tomography scan showing the right lower pulmonary vein with its dual drainage to inferior vena cava and via a meandering vein into left atrium.
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had normal drainage. Three indirect APCs to the right lung were noted arising from the celiac, right hepatic, and right renal arteries.

Under sedation, right femoral artery and vein were accessed and right heart catheterization was performed. Oximetry run showed a step-up of 11% between mixed venous and the pulmonary artery. Qp:Qs was 2.3:1. Mean PA pressure was 22 mmHg. The RLPV was cannulated from the IVC (see Supplementary material online, Video S1), and as the CT showed a constriction, a pullback was performed, with no gradient. RPA angiogram showed a good-sized vessel with normal distal arborization. Levophase demonstrated that the RLPV had dual drainage, into IVC-RA junction and LA through a meandering vein (Figure 2; see Supplementary material online, Video S2). These findings—dual drainage of the RLPV, normal sized RPA, good lung anatomy and an ASD was consistent with a variant scimitar syndrome. This was amenable for a complete transcatheter correction. We proceeded after obtaining informed consent from the parents.

The three APCs (see Supplementary material online, Video S3) were selectively entered and occluded with appropriately sized 0.035” MREye coils (Cook medical, USA). The ASD was closed with an 18 mm Amplatzer septal occluder (Abbott medical, USA). The communication between the RLPV and the IVC was closed with a 6 x 4 amplatz ductal occluder-1 (ADO1, Abbott medical, USA) after confirming the unobstructed drainage of IVC→RA (see Supplementary material online, Video S4) and RLPV→LA through the meandering vein (Figure 3). The procedure was uneventful (see Supplementary material online, Video S5). This rerouted the RLPV→LA via the meandering venous channel (Figure 4). The child was started on aspirin for 6 months. At 36 months post procedure, the child underwent follow-up with CT and echocardiography that demonstrated both the devices in position, with normal PA pressures (Figure 5, A and B).

Discussion

Scimitar syndrome has an incidence of 1–3 per 100000 live births.1 It is usually suspected on CXR showing a dextroposed heart with a “scimitar sign”—a right paracardiac shadow cast by the anomalous RLPV, resembling a Turkish sword. TTE confirms the diagnosis, but cardiac CT gives better anatomical delineation, clarifies the pulmonary venous drainage, and highlights the APCs. Magnetic resonance imaging additionally provides hemodynamic information by quantifying pulmonary

![Figure 2. Cardiac catheterization. (A) Pigtail angiogram in the descending aorta demonstrating three collaterals to the right lung. (B) Hand contrast injection into the anomalously draining right lower pulmonary vein through a multipurpose diagnostic catheter shows the dual drainage pattern, into the inferior vena cava and through a meandering venous channel, to the left atrium. (C) and (D) Pigtail angiogram of the right pulmonary artery in the levophase shows the right lower pulmonary vein and its dual drainage pattern. The inferior vena cava is seen in the lower half of the image and the meandering venous channel to the left atrium in upper half.](image-url)
blood flow. Cardiac catheterization is not required for diagnosis and is performed when a transcatheter intervention is planned.

Infantile presentations of scimitar syndrome can have severe PAH and poor prognosis despite early surgical intervention. Severe PAH in this group is multifactorial, due to (i) hypoplasia of the right lung and RPA, (ii) atelectasis of the right lung causing recurrent infections, (iii) obstruction to the RLPV outflow at the IVC, and (iv) high pressure APC supply to the right lung, causing segmental PAH. Transcatheter options in infantile scimitar syndrome are limited to either (i) coil closure of APCs or (ii) balloon dilatation and/or stenting of the constricted RLPV-IVC junction.

In older children and adults, the decision to treat hinges the symptoms—often dyspnoea or a demonstrated increase in Qp:Qs with right-sided chamber dilatation. Asymptomatic patients with a Qp:Qs < 1.5 can be followed up medically. Our patient was 13 years old and had symptoms of dyspnea as well as a high Qp:Qs ratio.

Multiple scimitar variants are described (i) with anomalously draining left sided pulmonary veins instead of right sided veins, (ii) dual drainage of the RLPV—both to the IVC and through a venous channel to the LA, (iii) multiple APCs, or (iv) with a normal RPA and right lung. Our patient had a variant with multiple APCs, normal RPA, and dual drainage of RLPV to the IVC and the LA via a meandering vein. The closure of RLPV to IVC with an occluder device baffled the pulmonary venous flow into the LA and corrected the anatomic defect in this patient. PA pressures remained stable during the procedure. Post procedure, RPA injection in the levophase clearly demonstrated the RLPV draining into the LA. The APCs transmit the high aortic pressure to the pulmonary capillary bed and potentiate PAH if left unoccluded. The ASD was occluded with a device. We followed up the child for 3 years post procedure and the PA pressures were normal at each follow-up. The child is asymptomatic and is thriving well. Earlier reports demonstrated

![Figure 3](image-url) Device closure. (A) The indirect aorto-pulmonary collaterals were closed with coils. (B) The secundum atrial septal defect was occluded with an 18 mm Amplatzer septal occluder using the standard left upper pulmonary vein approach. (C) A 6 x 4 mm Amplatzer ductal occluder-1 was positioned at the right lower pulmonary vein–inferior vena cava junction. A sheath injection in the inferior vena cava showed smooth inferior vena cava flows into the right atriumand no flow into the right lower pulmonary vein, suggesting complete occlusion. Pulmonary artery pressure was normal right lower pulmonary veinand after deployment of the device. (D) The devices and coils in-situ after the procedure.

![Figure 4](image-url) Schematic description. (A) A schematic describing the anatomy prior to intervention, demonstrating the dual drainage of the right lower pulmonary vein to the inferior vena cava and the left atrium. (B) Post procedure, after diversion of right lower pulmonary vein flow into the left atrium.
transcatheter management of variant scimitar syndromes variously with a device or coils to occlude the RLPV-IVC junction. Our case has a longer follow-up, and we achieved complete transcatheter correction with occlusion of the ASD and multiple APCs.

**Conclusion**

Variant forms of scimitar syndromes are rare. Multimodality imaging with CT, MRI and TTE will help delineate the anatomy. Significant shunts will require closure even in asymptomatic patients to avoid development of PAH. Dual drainage of the APV to IVC and LA can be managed with transcatheter interventions with good long-term results.

**Lead author biography**

Dr Sowmya Kasturi is an Assistant Professor of Pediatric Cardiology at the state run Sri Jayadeva Institute of Cardiovascular Sciences and research, Bangalore, India.

**Supplementary material**

Supplementary material is available at European Heart Journal – Case Reports

**Acknowledgements**

We acknowledge the support staff at the cardiac catheterization laboratory, pediatric wards, and intensive care unit in caring for this patient.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

**Consent:** The authors confirm that written consent for submission and publication of this case report including media and associated text has been obtained from the parents of the patient in line with COPE guidance

**Conflict of interest:** None declared.

**Funding:** None declared.

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