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# Multimodality imaging of scimitar syndrome in adults: A report of four cases

Calvin Ngai MD[1] | Robin S. Freedberg MD[1] | Larry Latson MD[2] | Michael Argilla MD[3] | Ricardo J. Benenstein MD[1] | Alan F. Vainrib MD[1] | Robert Donnino MD[1,4] | Muhamed Saric MD, PhD[1]

[1]The Leon H Charney Division of Cardiology, New York University Langone Health, New York City, New York

[2]Department of Radiology, New York University Langone Health, New York City, New York

[3]Department of Pediatrics, New York University Langone Health, New York City, New York

[4]Department of Medicine (Cardiology), VA New York Harbor Health Care System (Manhattan Campus), New York City, New York

**Correspondence**: Muhamed Saric, The Leon H Charney Division of Cardiology, New York University Langone Health, 550 First Avenue, New York, NY 10016 (Muhamed.Saric@nyumc.org).

Partial anomalous pulmonary venous return (PAPVR) comprises a group of congenital cardiovascular anomalies associated with pulmonary venous flow directly or indirectly into the right atrium. Scimitar syndrome is a variant of PAPVR in which the right lung is drained by right pulmonary veins connected anomalously to the inferior vena cava. Surgery is the definitive treatment for scimitar syndrome. However, it is not always necessary as many patients are asymptomatic, have small left-to-right shunts, and enjoy a normal life expectancy without surgery. We report multimodality imaging in four adults with scimitar syndrome and the implications for management of this rare syndrome.

**KEYWORDS**

cardiac magnetic resonance imaging, congenital cardiac anomalies, partial anomalous pulmonary venous return, scimitar syndrome, transesophageal echocardiogram, transthoracic echocardiogram

## 1 | INTRODUCTION

Scimitar syndrome or pulmonary venolobar syndrome was first described as an autopsy finding by the French physician Raoul Chassinat (1807–1891) in 1836 (Figure 1).[1] It is a rare variant of a group of congenital cardiovascular disorders, known as partial anomalous pulmonary venous return (PAPVR),[2] in which pulmonary venous flow terminates in the right atrium (RA) rather than the left atrium (LA). In scimitar syndrome, a part or all of the right lung is drained by right pulmonary veins connected anomalously to the inferior vena cava (IVC).

The term "scimitar" (a Western rendering of the Persian word شمشیر : shamshīr) was suggested by Nicolas Halász in 1956 to characterize the radiographic finding of a curvilinear, scimitar- or Middle Eastern sword-shaped density along the right cardiac border.[3] This finding is now recognized to be an anomalous vein that connects the right pulmonary vein to the IVC, rather than to the LA, creating a left-to-right shunt. But the "scimitar sign" (Figure 2) is present on the chest X-rays of fewer than half of patients with this syndrome.[3]

Scimitar syndrome is a very rare disorder that occurs in about 2 of every 100 000 births, affects more women than men,[4] and is usually associated with other congenital pulmonary and cardiac defects. Typically, right lung hypoplasia with associated right pulmonary artery hypoplasia and cardiac dextroposition accompany extracardiac left-to-right shunting through a scimitar vein. There are often associated forms of pulmonary sequestration, including abnormal collateral arterial supply to segments of the right lung from the descending aorta, as well as abnormalities of the bronchial tree. While scimitar syndrome can be discovered incidentally on imaging, patients often present with symptoms of right heart failure, as well as pulmonary complications like recurrent pneumonia and hemoptysis.[5]

The diagnosis of scimitar syndrome can be made by echocardiography, but the work-up often includes magnetic resonance imaging (MRI), computed tomography (CT), or cardiac catheterization, to evaluate for other associated abnormalities with important implications

  

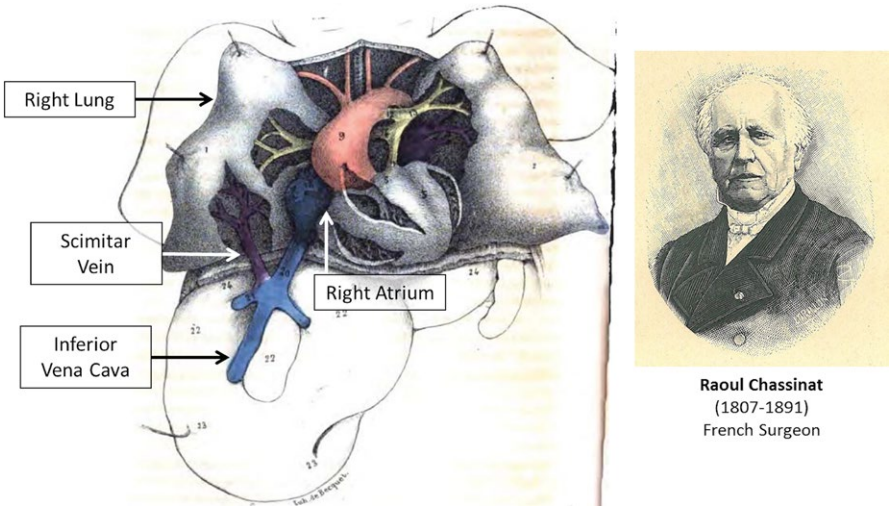


**FIGURE 1** Original Scimitar Description by Raoul Chassinat. This is the drawing from the 1836 paper by Raoul Chassinat with the original description of what is now known as the scimitar syndrome with added labels. In this original report, Chassinat refers to the scimitar vein as "système pulmonaire droit réduit à un seul vaisseau qui traverse le diaphragm et se rend dans la vein cave ascendante" or "right-sided pulmonary veins reduced to a single vessel which traverses the diaphragm and empties into the inferior vena cava"

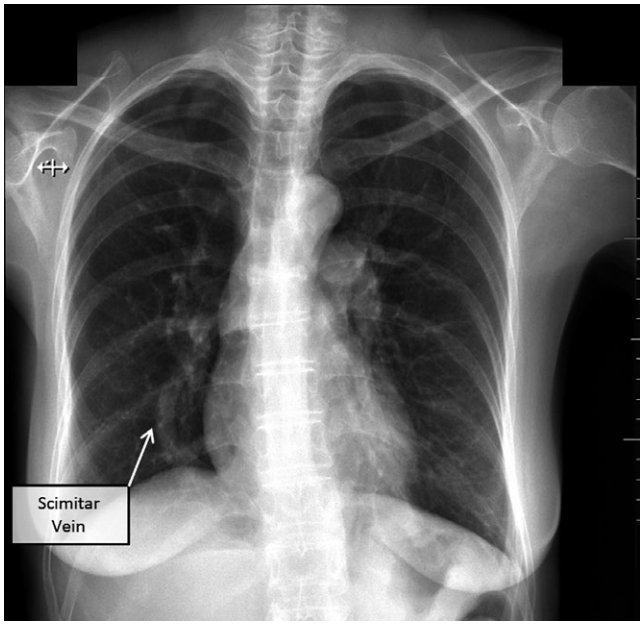


**FIGURE 2** Scimitar Vein on Chest Radiograph. The arrow points to a curved density created by the scimitar vein

for ongoing management. We describe multimodality imaging in four adults with scimitar syndrome, highlighting the typical findings, clinical presentations, and implications for management of this rare disorder.

## 2 | CASE 1

A 73-year-old woman with a history of chronic obstructive pulmonary disease (COPD), long QT syndrome with family history of sudden death, s/p implantable cardioverter/defibrillator (ICD), hypertension, and hyperlipidemia presented with worsening dyspnea and bilateral leg edema. Physical examination revealed jugular venous distension, lower extremity edema, and a pulsatile liver.

A transthoracic echocardiogram (TTE) demonstrated right heart dilatation with a hypokinetic right ventricle, severe pulmonary hypertension, and unusual high-velocity venous flow into the inferior vena cava (Figure 3). No intra-cardiac shunt was demonstrated on TTE.

A chest X-ray demonstrated evidence of a small right lung with rightward deviation of the cardiac silhouette (Figure 4A). Subsequent cardiac MRI with contrast showed a dilated pulmonary artery and left-to-right shunt (Figure 4B,C) with a ratio of pulmonary to systemic blood flow (Qp:Qs) of 2:1. It also demonstrated a single pulmonary vein emerging from the hypoplastic right lung and emptying into the supradiaphragmatic IVC (Figure 5), consistent with the scimitar syndrome variant of PAPVR.

Cardiac catheterization revealed normal coronary arteries and severe pulmonary arterial hypertension with normal pulmonary artery wedge pressures. The anomalous scimitar vein was also well visualized on a retrograde venous angiogram and a 3D transesophageal echocardiogram (TEE) (Figure 6).

The patient's right heart failure was medically managed, and she was discharged with cardiology follow-up. Her scimitar vein remains unrepaired.

## 3 | CASE 2

An 81-year-old woman with known scimitar syndrome presented to a pulmonologist for management of her chronic shortness of breath. He diagnosed asthma and treated her with inhaled mometasone and formoterol. Subsequent pulmonary function testing, however, demonstrated components of both obstructive and restrictive lung disease.

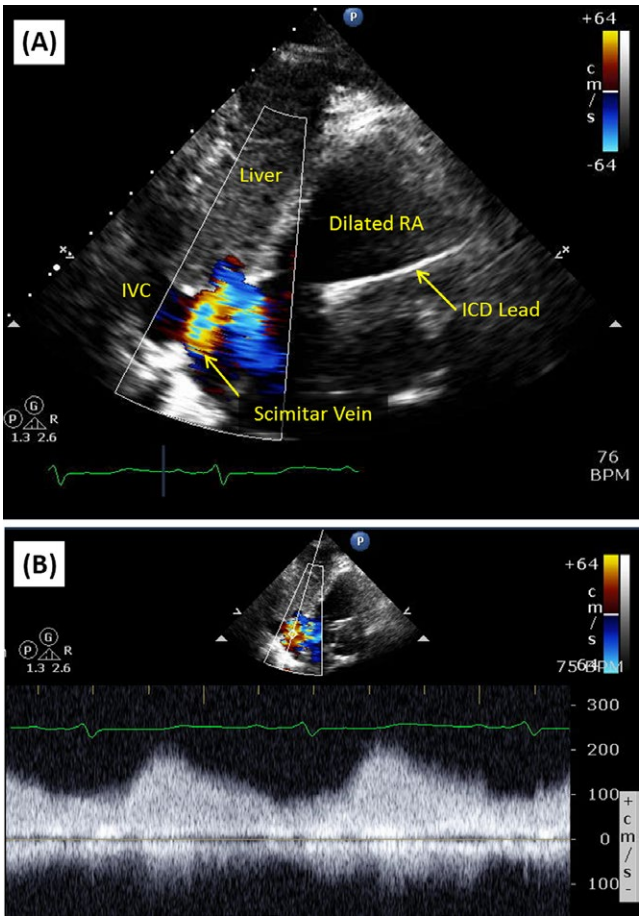


**FIGURE 3** Case 1 TTE. Subcostal views of TTE showing color Doppler flow (Panel A) and spectral Doppler (Panel B) of the scimitar vein draining into the IVC. Movie S1 corresponds to Panel A. ICD, implantable cardioverter-defibrillator; IVC, inferior vena cava; RA, right atrium

TTE revealed a normal left heart, mild pulmonary hypertension, rightward displacement of the heart, and an anomalous vein draining into the terminal portion of the IVC just proximal to the RA (Figure 7), consistent with the scimitar syndrome. Contrast-enhanced CT confirmed cardiac dextroposition and demonstrated hypoplasia of the right lung as well as the entire course of the scimitar vein (Figure 8).

The patient declined further cardiac evaluation and was lost to follow-up.

## 4 | CASE 3

A 62-year-old woman undergoing preoperative cardiac evaluation before adrenal schwannoma surgery was found to have a scimitar vein on routine TTE. She subsequently underwent uncomplicated left nephrectomy and adrenalectomy.

Several months later, the patient presented with progressive shortness of breath. Cardiac catheterization confirmed PAPVR via the scimitar vein with a Qp:Qs of 1.9:1 and revealed severe stenosis of her proximal left anterior descending artery (LAD). Biplane pulmonary angiography demonstrated the entire course of the scimitar vein with its drainage into the IVC in the venous phase (Figure 9). She was referred for surgical repair of scimitar syndrome in conjunction with coronary artery bypass grafting. Redirection of the scimitar vein flow was accomplished by intra-cardiac baffling of the right pulmonary veins into the LA via atrial septostomy (Figure 10).

She had an uncomplicated postoperative course and remained asymptomatic for 6 months. Thereafter, however, she redeveloped shortness of breath. Work-up revealed narrowing of the baffle and inability to demonstrate flow from the right pulmonary veins into the left atrium. Repeat cardiac catheterization showed persistence of a left-to-right shunt with a Qp:Qs of 1.5:1.

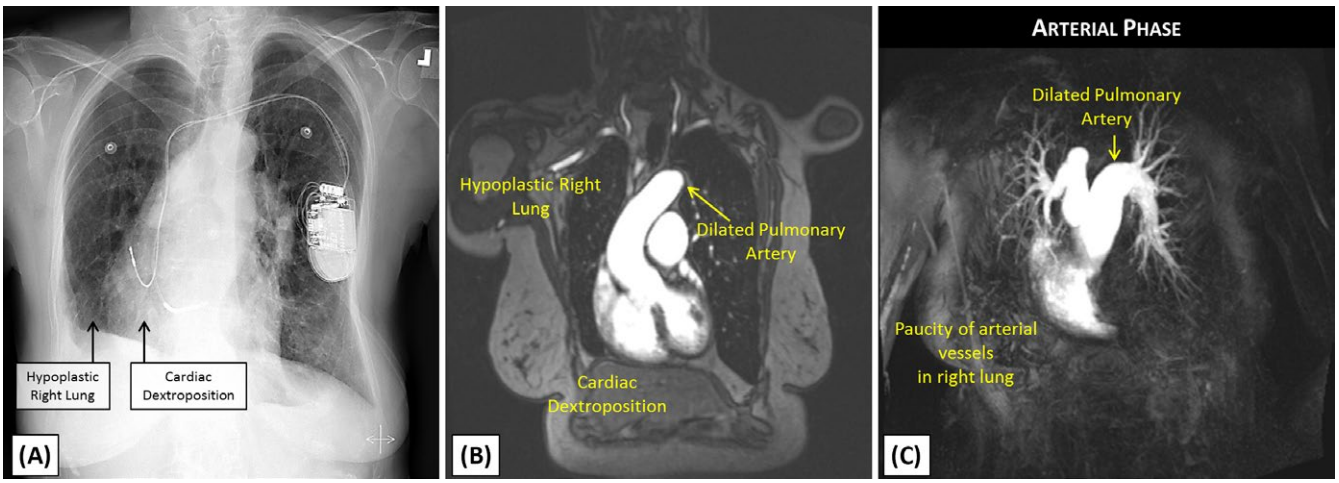


**FIGURE 4** Case 1 CXR and MRI, part 1. Panel A, Chest X-ray depicting two of the three anatomic hallmarks of scimitar syndrome, namely right lung hypoplasia and cardiac dextroposition. Panel B, MRI showing a dilated main pulmonary artery in addition to right lung hypoplasia and cardiac dextroposition. Panel C, Arterial phase of the contrast-enhanced MRI (i.e, MR angiogram) demonstrating paucity of arterial vessels in right lower lung during pulmonary artery phase. Review of systemic arterial phase did not show collaterals from the aorta

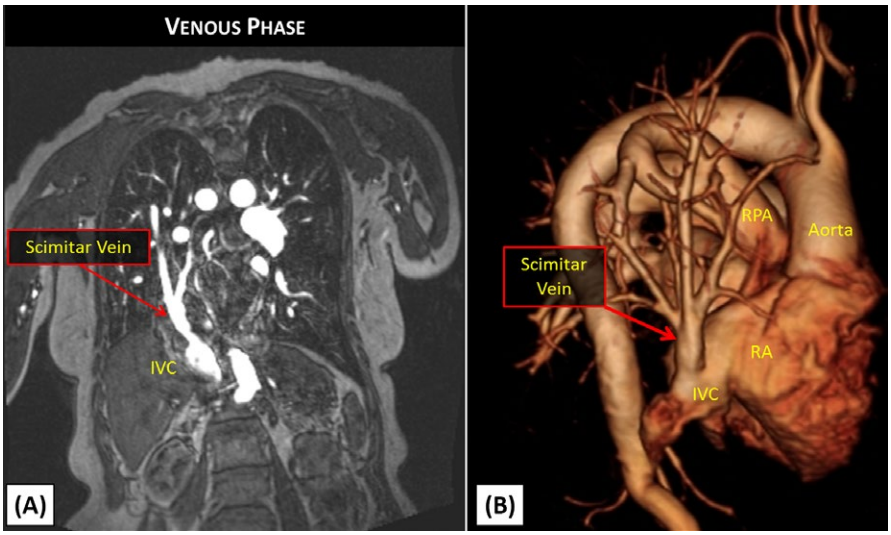


**FIGURE 5** Case 1 MRI, part 2. Panel A, Venous phase of the contrast-enhanced MRI (MR venogram) demonstrating the scimitar vein draining into the IVC. Panel B, 3D MRI rendering of the scimitar vein draining into the IVC. IVC, inferior vena cava; RA, right atrium; RPA, right pulmonary artery

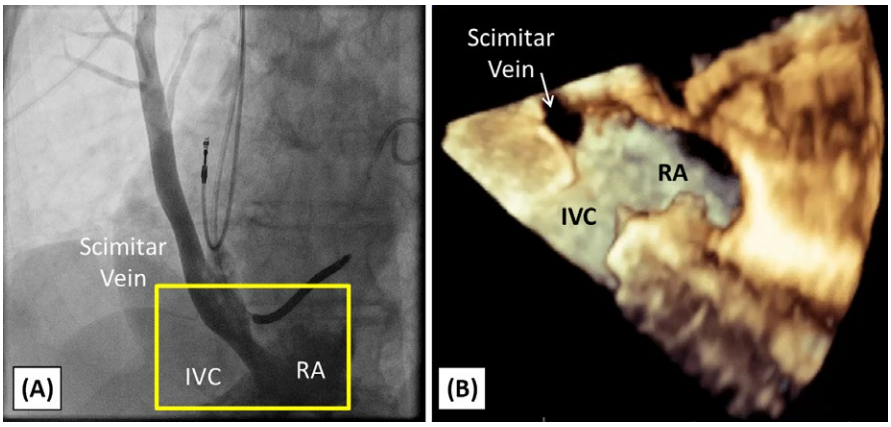


**FIGURE 6** Case 1 Cardiac Catheterization and 3D TEE. Panel A, Angiogram of the scimitar vein draining into the junction of the IVC and RA. Movie S2 corresponds to this panel. Panel B, 3D TEE image of the scimitar vein draining into the IVC/RA junction. Movie S3 corresponds to this panel. IVC, inferior vena cava; RA, right atrium

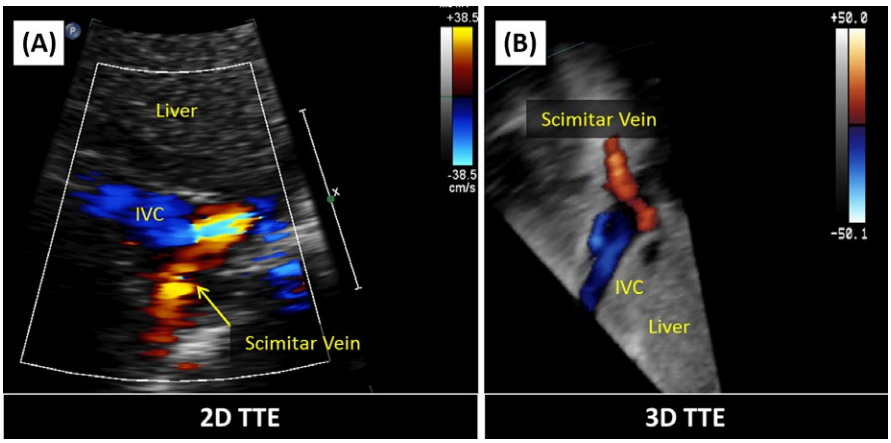


**FIGURE 7** Case 2 TTE. TTE showing the scimitar vein draining into the IVC on 2D image (panel A; subcostal view) and 3D image (Panel B). IVC, inferior vena cava. Movie S4 corresponds to panel A, and Movie S5 corresponds to panel B

Following discussion with her cardiologist, the patient declined further surgical intervention.

## 5 | CASE 4

A 48-year-old man with no significant past medical history presented for a routine pre-employment physical examination. His chest X-ray was interpreted as showing a right lung tumor. Subsequently, the patient developed chest tightness and shortness of breath. A CT angiogram of the chest excluded pulmonary embolism but revealed two right pulmonary veins draining into the inferior vena cava, and he was referred for surgical repair of his newly diagnosed scimitar syndrome.

The scimitar vein drainage into the IVC was visualized on the intra-operative TEE (Figure 11). The surgical repair of his PAPVR

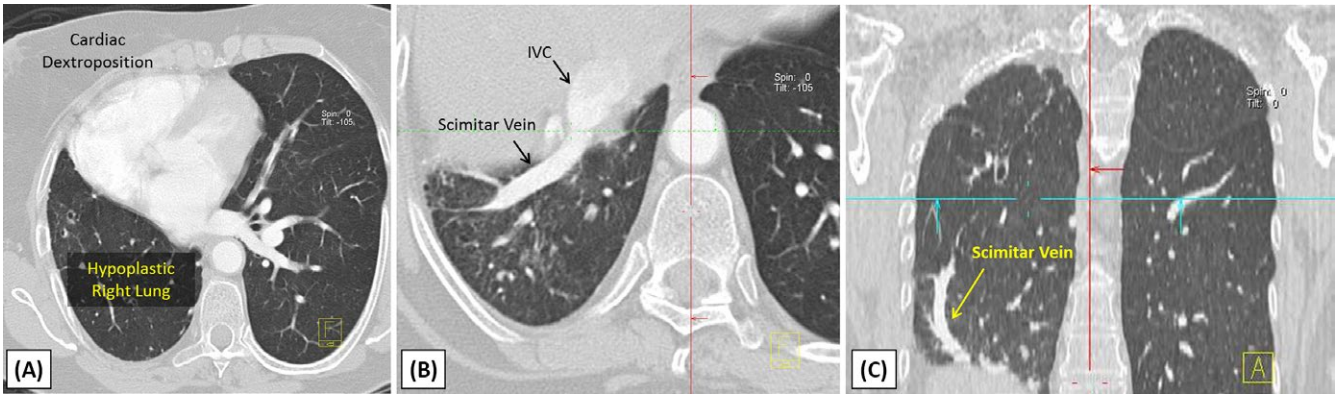


**FIGURE 8** Case 2 CT. Panel A, Axial view of chest CT showing a hypoplastic right lung with cardiac dextroposition. Panel B, Close-up axial view of a chest CT showing the course of the scimitar vein as it drains into the IVC. Panel C, Coronal view of a chest CT demonstrating the course of the scimitar vein. IVC, inferior vena cava

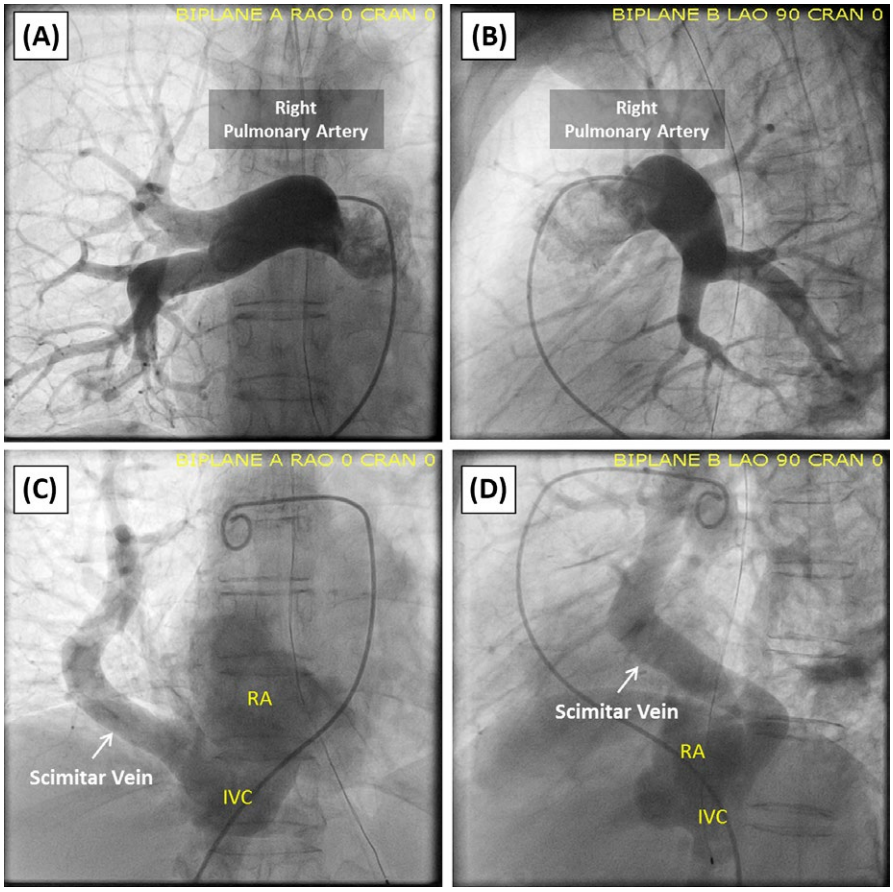


**FIGURE 9** Case 3 Angiogram. Panels A and B are biplane views of the angiogram showing dilated right pulmonary artery. Movie S6 corresponds to these two panels. Panels C and D are biplane views of the angiogram showing scimitar vein draining in the IVC/RA junction. Movie S7 corresponds to these two panels. IVC, inferior vena cava; RA, right atrium

consisted of right pulmonary vein connection to the LA via a RA baffle and atrial septostomy. TTE with agitated saline intravenous contrast injection during a vigorous Valsalva maneuver was used to demonstrate the absence of a baffle leak (Figure 12).

He has been asymptomatic and remained well during the several years since surgery.

## 6 | DISCUSSION

These 4 distinct cases are examples of the clinical presentation, diagnosis, and management of scimitar syndrome in adults. Collectively, they illustrate the use of multiple modalities—TTE, TEE, MRI, CT, and cardiac catheterization with pulmonary arterial and retrograde

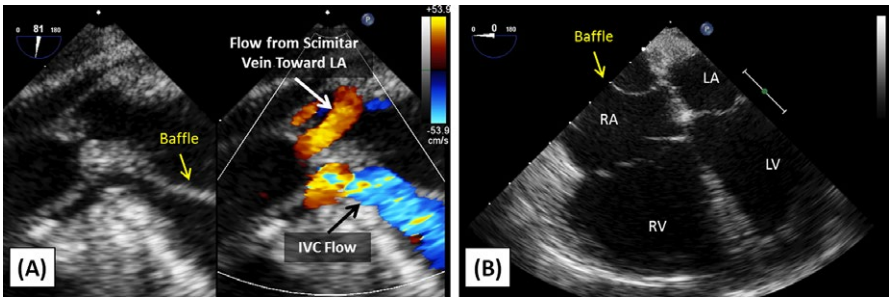


**FIGURE 10** Case 3 Post-op TEE. Panel A, Post-operative TEE of the baffle diverting the scimitar vein flow into the LA during imaging of the IVC (Panel A) and in the 4-chamber view (Panel B). Movie S8 corresponds to panel A, and Movie S9 corresponds to panel B. IVC, inferior vena cava; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle

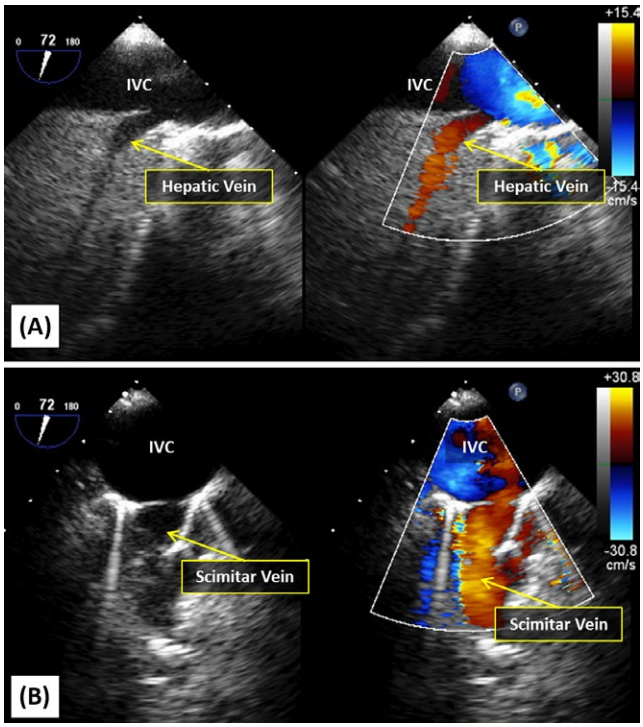


**FIGURE 11** Case 4 Intra-operative TEE. Panel A shows the normal hepatic vein drainage into the IVC; this flow should be distinguished from the scimitar vein flow into the IVC seen on Panel B. Movie S10 corresponds to panel A, and Movie S11 corresponds to panel B. IVC, inferior vena cava

venous angiography—in the anatomic and physiologic evaluation of this rare congenital anomaly. The last two cases also include the evaluation of the integrity of its repair by TEE and TTE with saline contrast.

The anatomic hallmarks of scimitar syndrome are PAPVR to the IVC, right lung hypoplasia with associated right pulmonary artery hypoplasia, and cardiac dextroposition. The physiologic consequence of the shunting of oxygenated pulmonary venous blood to the right heart is measured by the ratio of pulmonary to systemic blood flow (Qp:Qs).

Patients with scimitar syndrome who present as adults or older children typically have less severe disease and a better prognosis than those who present as infants.[6] Symptoms of infantile scimitar syndrome include poor feeding and failure to thrive, cyanosis, and lethargy. Adults, on the other hand, usually present with fatigue, dyspnea, and signs of right heart failure as seen in some of the patients in our case series. The severity of these symptoms depends on the degree of left-to-right shunting generated by the scimitar vein. In addition, the coexistence of other congenital cardiac and pulmonary defects, including atrial septal defects and lung hypoplasia, can complicate the clinical picture.[3]

Patients with only a single anomalous pulmonary vein and without any associated defect are typically asymptomatic and have normal lifespans. A French study examining the clinical and hemodynamic findings in 122 adult patients with scimitar syndrome concluded that although hypoplasia and vascular abnormalities in the lung were common, significant left-to-right shunts were present in less than 25 percent of patients and pulmonary artery pressures were normal in 94 of the patients. Of the 85 of 122 patients who were treated medically in that study, 79 had favorable outcomes and led normal lives.[7] Another multicenter study exploring the natural history of patients with a scimitar vein in 44 individuals found that the course is often benign when no other congenital cardiac abnormalities are present. However, when it was associated with other cardiac defects or pulmonary hypertension, there was an increased risk of heart failure and mortality.[8]

The chest X-ray may provide the first clue to scimitar syndrome, but the initial diagnosis is usually made by echocardiography[9] and confirmed by other imaging modalities like MRI or CT. The diagnosis can also be made at cardiac catheterization, by pulmonary arterial angiography or retrograde contrast injection of the anomalous vein. In addition, data derived during catheterization regarding hemodynamics and shunting, such as pulmonary artery pressure and Qp:Qs, may guide management of the scimitar syndrome. Therapeutically, interventional cardiologists can also occlude small anomalous connections to reduce left-to-right shunting.

Asymptomatic individuals with scimitar syndrome resulting in small shunts can lead normal lives without surgical intervention. But surgery is the definitive treatment for symptomatic scimitar syndrome. The approach to surgical management depends on the anatomy of the PAPVR. One technique separates the scimitar vein from the IVC and implants it directly into the LA using a polytetrafluoroethylene interposition tube graft.[10] Other surgical options

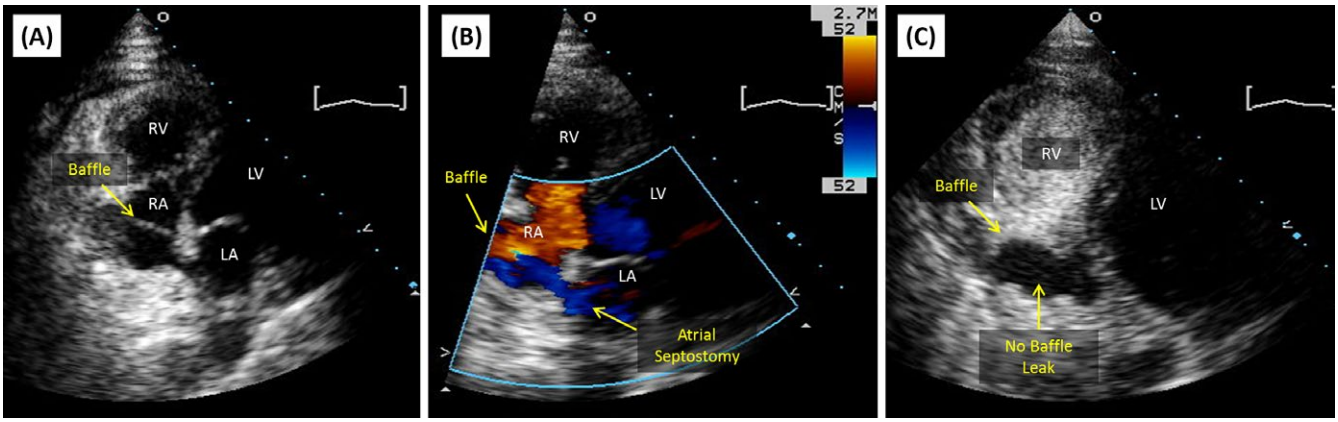


**FIGURE 12** Postoperative TTE with saline. This demonstrates the use of agitated saline intravenous contrast to confirm the absence of a baffle leak postsurgical correction of scimitar syndrome. Panel A shows the baffle directed toward the inter-atrial septum. Movie S12 corresponds to this panel. Panel B shows the color Doppler flow within the baffle entering the LA through the hole created by atrial septostomy. Movie S13 corresponds to this panel. Panel C shows no evidence of baffle leak with agitated saline contrast injection into systemic venous circulation with simultaneous Valsalva maneuver. Movie S14 corresponds to this panel. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle

include baffling of the anomalous venous return into the LA. This is accomplished using the anterior wall of the RA to form a tunnel through which the scimitar vein flow is diverted toward the LA, and septostomy through which the baffled blood passes into the LA.[11]

Surgical correction should be considered in patients who have hemodynamically significant left-to-right shunting (Qp:Qs greater than 2:1), evidence of right heart failure, or recurrent pulmonary infections, and in those undergoing surgical repair of other major cardiac lesions.

In conclusion, our case series demonstrates how challenging it can be to diagnose scimitar syndrome in adult patients and shows the incremental value of various imaging techniques in elucidating the anatomic and physiologic aspects of the syndrome. We do not imply that every patient with scimitar syndrome should undergo every single imaging modality presented; we hope that the variety of presented imaging techniques will provide guidance to clinicians for individualized approach in each patient.

## ORCID

*Calvin Ngai* http://orcid.org/0000-0002-9799-0869
*Alan F. Vainrib* http://orcid.org/0000-0002-0832-8452

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## SUPPORTING INFORMATION

Additional supporting information may be found online in the Supporting Information section at the end of the article.

**Movie S1.** Transthoracic echocardiogram (TTE) showing color Doppler flow of the scimitar vein draining into the inferiorvena cava (IVC).

**Movie S2.** Angiogram of the scimitar vein draining into the junction of the IVC and right atrium (RA).

**Movie S3.** 3D transesophageal echocardiogram (TEE) of the scimitar vein draining into the IVC/RA junction.

**Movie S4.** 2D TTE showing the scimitar vein draining into the IVC in subcostal view.

**Movie S5.** 3D TTE showing the scimitar vein draining into the IVC in subcostal view.

**Movie S6.** Biplane angiogram showing dilated right pulmonary artery in a patient with scimitar syndrome.

**Movie S7.** Biplane angiogram showing scimitar vein draining in the IVC/RA junction.

**Movie S8.** Postoperative TEE of the baffle diverting the scimitar vein flow into the LA during imaging of the IVC.

**Movie S9.** Postoperative TEE of the baffle diverting the scimitar vein flow into the LA in a 4-chamber view.

**Movie S10.** TEE shows normal hepatic vein drainage into the IVC.

**Movie S11.** TEE shows the scimitar vein flow into the IVC.

**Movie S12.** Postoperative TTE demonstrates the baffle directed toward the interatrial septum.

**Movie S13.** Postoperative TTE demonstrates the color Doppler flow within the baffle entering the LA through the hole created by atrial septostomy.

**Movie S14.** Postoperative TTE shows no evidence of baffle leak with agitated saline contrast injection into systemic venous circulation with simultaneous Valsalva maneuver.

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