

## Partial Anomalous Pulmonary Venous Return: Scimitar Vein

### Abstract

Scimitar syndrome is a rare association of congenital cardiopulmonary anomalies characterized by partial anomalous pulmonary venous return, in which an abnormal right pulmonary vein drains into the inferior vena cava. This case exemplifies the role of transesophageal echocardiography in perioperative management and surgical decision-making.

**Keywords:** *Cardiothoracic anesthesia, transesophageal echocardiogram, transthoracic echocardiography*

### Introduction

Scimitar syndrome is a rare association of congenital cardiopulmonary anomalies characterized by partial anomalous pulmonary venous return, in which an abnormal right pulmonary vein drains into the inferior vena cava. Although frequently diagnosed in infancy, patient may remain asymptomatic until adulthood. Surgical correction is indicated in adults with signs of pulmonary volume overload or right heart dilation.<sup>[1,2]</sup> During surgical correction, transesophageal echocardiography (TEE) can be utilized to define anatomy of the lesions and confirm the adequacy of the repair.

### Case Report

A 41-year-old female presented with recurrent pulmonary infections and dyspnea on exertion. A right lower lobe with vertical, linear opacification extending to her right diaphragmatic margin was seen on chest X-ray. Evaluation with computed tomography, transthoracic echocardiography, and right heart catheterization demonstrated a scimitar vein – a large, right lower pulmonary vein with anomalous drainage to the inferior vena cava (IVC) [Figure 1]. Additional findings included an atretic right upper pulmonary vein, a patent foramen ovale (PFO), normal biventricular and valvular function, and normal right-sided pressures. The patient was referred for surgery.

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Intraoperative transesophageal echocardiography (TEE) was performed. Employing midesophageal views, an aneurysmal atrial septum and a PFO with right-to-left flow confirmed by color flow Doppler (CFD) were identified. Left but not right pulmonary veins were identified. Although commonly atretic, the diameter of the visualized right pulmonary artery was normal. Turning to the patient's right from a transgastric, midpapillary view and at an omniplane angle of 45°, the scimitar vein was imaged at the level of the hepatic vein (HV) and IVC confluence [Figure 2].

CFD interrogation of these vessels revealed flow from scimitar vein into the IVC [Figure 3]. Pulse wave Doppler (PWD) interrogation of the HV displayed late systolic flow reversal [Figure 3b]. In the absence of severe tricuspid regurgitation, this may be explained by the large volume of flow from the scimitar vein. PWD of the scimitar vein revealed a higher velocity, monophasic waveform compared to the characteristic lower velocity, and biphasic waveform of the HV [Figure 4].

Surgical correction was achieved by anastomosing a tube graft to the atrial septal defect and then positioned to traverse and exit the right atrium and pericardium lateral and posterior to the phrenic nerve. The opposite end of the tube graft was anastomosed to the scimitar vein

**How to cite this article:** Heinke T, Stewart SR, Steinberg T, Hand WR, Abernathy JH. Partial anomalous pulmonary venous return: Scimitar vein. *Ann Card Anaesth* 2017;20:259-61.

**Received:** May, 2016. **Accepted:** January, 2017.

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#### Access this article online

**Website:** [www.annals.in](http://www.annals.in)

**DOI:** 10.4103/aca.ACA\_82\_16

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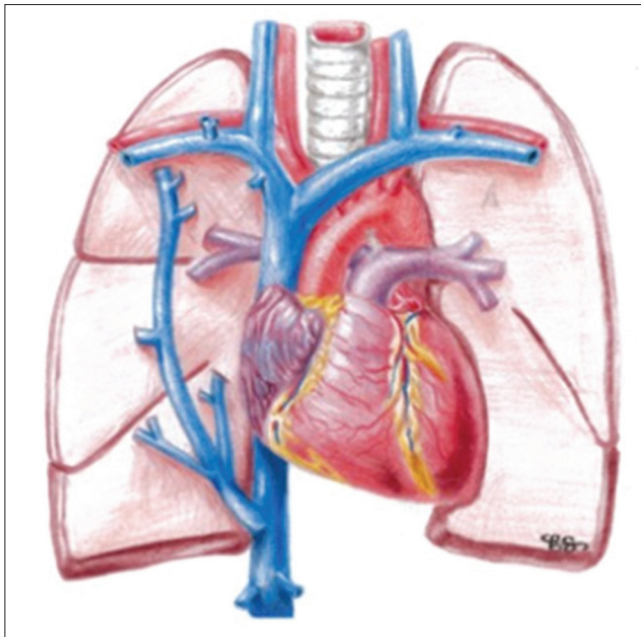


Figure 1: Scimitar vein anatomy

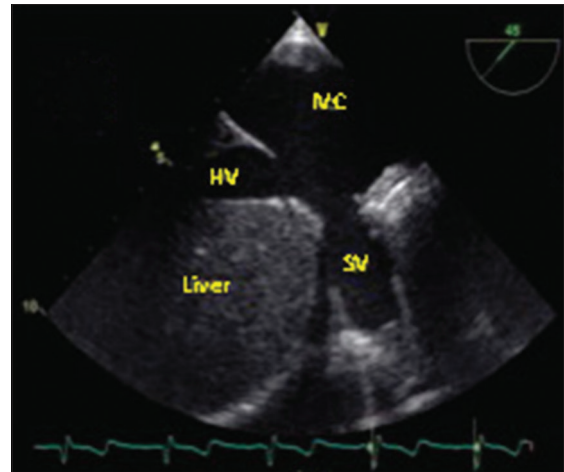


Figure 2: Two-dimensional transesophageal echocardiography of scimitar vein anatomy

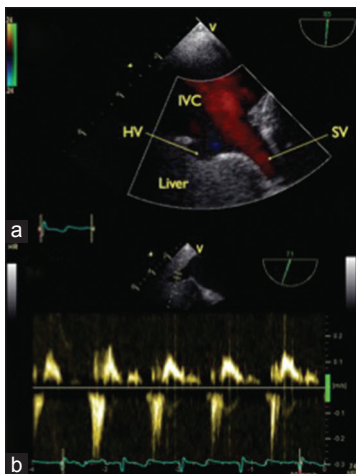


Figure 3: (a) Scimitar vein flow. Color flow Doppler showing flow from the scimitar vein into the inferior vena cava. (b) Pulse wave Doppler interrogation of the hepatic vein flow

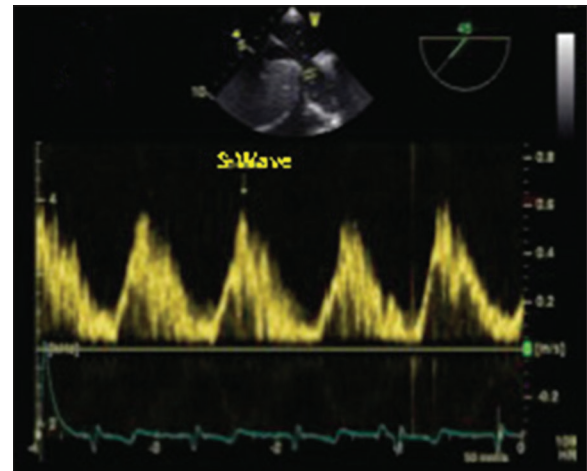


Figure 4: Pulse wave Doppler interrogation of the scimitar vein flow

proximal to its insertion on the IVC. The scimitar vein was ligated distal to the conduit. Following correction, TEE using CFD and PWD confirmed the restoration of right-sided pulmonary venous drainage into the left atrium. Doppler interrogation of the scimitar and HVs demonstrated abolition of flow from the scimitar vein into the IVC and restoration of a normal hepatic venous Doppler tracing [Figure 5].

### Discussion

Scimitar syndrome is an association of congenital cardiopulmonary anomalies characterized by partial anomalous pulmonary venous return, in which an abnormal right pulmonary vein drains into the IVC

either above or below the diaphragm. In addition, frequently present is an abnormal right pulmonary arterial supply consisting of aortopulmonary collaterals. Adults often present with dyspnea and recurrent pulmonary infections. Additional findings include right ventricular hypertrophy, right bundle branch block, and an atrial septal defect. Pulmonary artery hypertension if present is mild.<sup>[3]</sup> Surgical correction is indicated in adults with signs of pulmonary volume overload or right heart dilation.<sup>[1,2]</sup> The scimitar vein is frequently initially discovered on chest X-ray by the curvilinear “scimitar sword” appearance of the anomalous venous flow to the IVC. Subdiaphragmatic scimitar veins can be imaged by obtaining transgastric midpapillary views and turning the TEE probe clockwise while adjusting the omniplane angle to visualize the IVC. The sensitivity of detecting the scimitar vein can be increased using CFD while scanning the IVC, looking for turbulent, nonlinear flow patterns, in contrast to typical laminar flow in the IVC.<sup>[4]</sup> Identifying the HV anastomoses to the IVC can further

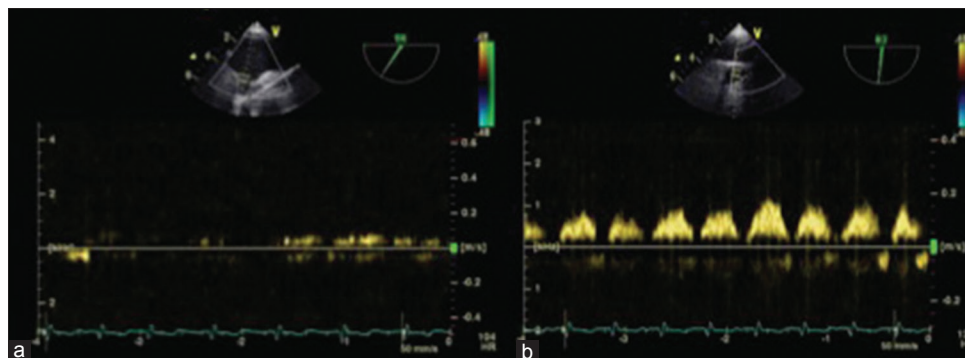


Figure 5: Postrepair pulse wave Doppler interrogation of (a) the ligated scimitar vein and (b) the hepatic vein. HV: Hepatic vein, IVC: Inferior vena cava, SV: scimitar vein

define the location of the scimitar vein–IVC confluence. Methods for the two-dimensional (2D) identification and the PWD interrogation of the HVs and the characteristic waveform comprising antegrade systolic (S) and diastolic (D) waves and a retrograde (A) wave have been well described.<sup>[5]</sup> PWD interrogation of the scimitar vein demonstrates a monophasic, high-velocity waveform [Figure 2].<sup>[6]</sup> This may be explained by the large volume of blood flowing into a compliant IVC. Following correction, 2D and Doppler examination can confirm restoration of anatomically correct blood flow.

In summary, although scimitar syndrome is rare, TEE plays a significant role in the perioperative management and surgical decision-making as well as confirming the effectiveness of surgical correction.

#### Financial support and sponsorship

Nil.

#### Conflicts of interest

There are no conflicts of interest.

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