Scimitar syndrome is a rare congenital disorder with an incidence of approximately 2 per 100 000 live births. The hallmark of this syndrome is partial or complete anomalous venous return from the right lung, directly into the inferior vena cava (IVC), thereby creating a left-to-right shunt. The syndrome is almost invariably associated with other cardiac and extracardiac abnormalities including atrial septal defects (ASD), dextrocardia, abnormal right lung lobation, and right lung hypoplasia. The infantile syndrome is highly associated with ASDs, ventricular septal defects, patent ductus arteriosus, coarctation, and tetralogy of Fallot.

The abnormal venous drainage gives the characteristic radiographic appearance akin to that of a “scimitar” (Turkish sword) with a shadow on the right side of the heart progressively widening as it approaches the cardiophrenic angle. The expected natural course of uncorrected scimitar syndrome is similar to uncorrected large ASD with eventual right heart failure, and therefore even asymptomatic adults with Qp:Qs ratio of more than 1.5:1 should undergo corrective surgery. Catheterization is often recommended and advised to assess pulmonary vascular resistance and suitability for operative repair. Other imaging modalities include preoperative echocardiography, cardiac MRI, or computed tomography (CT). Which of these latter techniques is chosen depends on institutional preference and experience, with factors such as cost, the potential need for patient sedation, exposure to ionizing radiation, and exposure to intravenous contrast material also to be considered in the choice of imaging modality. Perioperative transesophageal echocardiography (TEE) has previously been shown to be useful for demonstrating the abnormal scimitar vein anatomy and also for confirming unobstructed blood flow after anastomosis of the scimitar vein to the left atrium after repair. In this article, we...

Figure 1. A, Preoperative chest radiograph demonstrates dextroversion of the heart and hypoplasia of the middle and lower lobes of the right lung. The typical “scimitar” appearance from the aberrant venous drainage is not seen on plain film. B, Right heart catheterization with selective pulmonary artery contrast angiography. The image illustrates the late (venous) phase with contrast draining from the right lower and middle lobes forming the typical “scimitar” shape, with a narrow base at the hilum progressively becoming wider before entering the inferior vena cava below the diaphragm. C, Live imaging from the surgical head camera shows the aberrant vein as it exits the right lung and travels caudally, parallel to the right side of the pericardium, and disappears through the diaphragm.
describe the utility of 3D TEE as a complementary intraoperative imaging modality to visualize the abnormal scimitar vein. To illustrate our case, we have composed an imaging vignette that includes images and movies from the preoperative right heart catheterization, the intraoperative TEE, and video recorded from a surgical head camera demonstrating the surgical repair (see online-only Data Supplement Movies).

The patient is a 25-year-old woman who was initially diagnosed with dextrocardia as a child. Although asymptomatic for most of her life, lately, over the course of several months, she developed symptoms of heart failure with peripheral edema and dyspnea. Her diagnostic workup included a chest radiograph (Figure 1A), right heart catheterization with selective pulmonary artery contrast angiography (Figure 1B), and transthoracic echocardiography. These studies demonstrated partial anomalous venous return (PAVR) of the right, middle, and lower lobes (Figure 1B), hypoplasia of the right lung, and dextroversion of the heart with a Qp:Qs ratio of 1.3, confirming the diagnosis. Left ventricular function was normal, but the right ventricle was dilated with mildly elevated pulmonary artery pressures.

The patient underwent corrective surgery, during which time a comprehensive 2D and 3D TEE was performed (iE33; Phillips Medical Systems; Andover, MA). Both 2D and 3D TEE allowed visualization of the scimitar vein and its entrance into the IVC (Figure 2A). Images from 3D TEE seemed particularly helpful in demonstrating the scimitar vein orientation in reference to the hepatic veins and the IVC. This en face view acquired with the 3D full-volume mode demonstrates the entry of the scimitar vein into the IVC. The image is rotated so that the echocardiographic point of view is caudally from the IVC looking cranially. Dimensions of the scimitar vein measured on the captured 3D dataset illustrate dimensions of the scimitar vein that correlated well with the surgical findings (1.93 ± 0.95 cm).

Figure 2. A, Full-volume 3D transesophageal echocardiography illustrates the entry of the anomalous pulmonary vein into the inferior vena cava (IVC) from a right superior direction. The anatomic relationship to the left and right hepatic veins is clearly seen. B, Full-volume 3D TEE with color flow Doppler clearly distinguishes a high-velocity jet coming from the anomalous pulmonary vein from laminar flow in the hepatic vein and in the IVC. This en face view acquired with the 3D full-volume mode demonstrates the entry of the scimitar vein into the IVC. The image is rotated so that the echocardiographic point of view is caudally from the IVC looking cranially. D, Multiplanar reconstruction of the 3D volume dataset illustrates dimensions of the scimitar vein that correlated well with the surgical findings (1.93 ± 0.95 cm).
recovery and was discharged home 5 days after the operation. The outcomes of surgical repair of scimitar syndrome are excellent, with a very high rate of reoperation-free survival, although risk of operative mortality or reoperation is higher in patients presenting with symptoms in infancy.

In conclusion, we demonstrate that 3D TEE is a powerful complementary perioperative imaging modality in complex cases of PAVR repair. Three-dimensional TEE was especially helpful in understanding the unique orientation of the scimitar anatomy, demonstrating the course of the scimitar vein into the IVC and its relation to the hepatic veins.

Disclosures

None.

References


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Figure 3. After the repair, 2D color flow Doppler (A) and 2D pulsed-wave Doppler (B) ensure that there is no obstruction or kinking at the anastomosis site of the aberrant pulmonic vein to the left atrium. Color Doppler demonstrates laminar flow in the pulmonary vein while pulse wave Doppler demonstrates a deep A-wave in the spectral tracing, presumably due to acute volume loading of the left ventricle.
Three-Dimensional Transesophageal Echocardiography Enhances Multimodal Imaging of a Successful Repair of a Case With Scimitar Syndrome

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