Scimitar Syndrome

Added Value by Isotropic Flow-Sensitive Four-Dimensional Magnetic Resonance Imaging With PC-VIPR (Phase-Contrast Vastly Undersampled Isotropic Projection Reconstruction)

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Scimitar, or pulmonary venolobar, syndrome is a rare but well-known congenital cardiovascular defect that includes a hypoplastic right pulmonary artery and right lung, which leads to displacement of cardiac structures into the right hemithorax, anomalous systemic arterial supply to the right lung, and a characteristically curved anomalous right pulmonary vein that drains into the inferior vena cava and resembles the curved Middle Eastern sword “scimitar.”

A variety of congenital thoracic abnormalities are associated with this specific type of partial anomalous pulmonary venous return.

Imaging, and specifically findings from magnetic resonance imaging, in an 18-month-old male (11 kg body weight) with known congenital right pulmonary venolobar syndrome with increasingly frequent cyanotic episodes are presented. Findings on chest radiography and contrast-enhanced computed tomography of the chest performed when the patient was 4 days old included right lung hypoplasia and partial anomalous pulmonary venous return with scimitar vein to the supradiaphragmatic inferior vena cava (Figure 1). Echocardiography identified the scimitar vein and an atrial septal defect. Cardiac magnetic resonance imaging, including 4-dimensional flow-sensitive magnetic resonance imaging, confirmed these findings but also identified additional cardiovascular abnormalities, including an additional partial anomalous pulmonary venous return from the upper right lung to the superior vena cava and an anomalous systemic artery from the upper abdominal aorta to the lower right lung (Figure 2). Further comprehensive analysis of flow, blood flow quantification, and detection of blood flow direction was feasible in all analyzed vessels.

With advanced magnetic resonance imaging approaches, simultaneous anatomic and functional hemodynamic imaging can be obtained by use of 4-dimensional flow-sensitive sequences such as PC-VIPR (phase-contrast vastly undersampled isotropic projection reconstruction). PC-VIPR mag-

Figure 1. Chest radiograph (left) and chest computed tomography (right, A–D) in a 4-day-old boy with scimitar syndrome. Although right lung hypoplasia and shift of the mediastinal structures to the right are well delineated on the chest radiograph, the anomalous pulmonary venous return (“scimitar vein”; black arrowheads) cannot be readily appreciated. The scimitar vein is better appreciated with computed tomography (performed at age 4 days; white arrowheads), which also confirmed right lung hypoplasia and mild compression of the right lower lobe. Additional partial anomalous pulmonary venous return vessels were not identified, possibly owing to the small anatomic scale at that age and a lack of information on blood flow direction.
Nuclear resonance imaging was performed on a clinical 1.5T Signa HDx MR system (GE Medical Systems, Milwaukee, Wis) equipped with an 8-element phased-array cardiac coil and TwinSpeed gradient performance in “whole” mode (gradient strength = 40 mT/m, maximum rise time = 288 μs). Data were acquired during free breathing with respiratory gating. Parameters for the 4-dimensional flow sequence (PC-VIPR) were adapted to the specific anatomic demands: Echo time/ repetition time = 3.08/9 ms; flip angle = 10°, bandwidth = 62.5 kHz; velocity-encoding sensitivity = 100 cm/s; field of view = 256 × 256 mm; slab thickness = 14 cm; 3-dimensional radial acquisitions with 256 data points in the readout direction; image volume = 256 × 256 × 140 voxels; spatial resolution = 1 × 1 × 1 mm³; and 12 time frames per cardiac cycle at a heart rate of 137 bpm. Offline image visualization was performed with Vitrea Advanced software (Vital Images Inc, Minnetonka, Minn) and MIMICS (Mimics Innovation Suite, Materialise, Ann Arbor, Mich) for image segmentation and morphology (Figure 2; online-only Data Supplement Movie I), which subsequently was used to specify regions for flow visualization and analysis with EnSight 9.0 (CEI, Apex, NC; Figure 3; online-only Data Supplement Movies I and II).

Four-dimensional flow imaging not only enables the analysis of cardiovascular morphology but also provides quantitative flow parameters and blood flow patterns all from a single acquisition, thereby aiding in the diagnosis, identifica-

![Figure 2](image-url)

**Figure 2.** A, Maximum-intensity projection of the phase-contrast angiogram in sagittal oblique direction as seen from 30° left anterior oblique view. In addition to the pronounced scimitar vein (ScimV), the hypoplastic right pulmonary artery (*), an additional partial anomalous pulmonary venous return vein in the right upper lobe (open white arrow), and the anomalous systemic artery from the celiac trunk to the right lower lung (white arrowheads) can be appreciated. B, Posterior view of segmented PC-VIPR angiography data with color-shaded surface display. For detailed understanding and ready apprehension, the oxygenized arterial (red), oxygenized partial anomalous pulmonary venous return (pink), deoxygenized venous and right ventricular (blue) structures, and portal venous system (yellow) were color-coded. SVC indicates superior vena cava; AAO, ascending aorta; LPA, left pulmonary artery; LA, left atrium; RA, right atrium; IVC, inferior vena cava; and DAO, descending aorta.

![Figure 3](image-url)

**Figure 3.** Color-coded particle trace representation of blood flow contribution and hemodynamics in the right atrium from a posterior view. As opposed to previous descriptions, the blood flow in the right atrium showed overt changes in flow patterns, with a backward rotation of the superior vena cava inflow. Animated blood flow behavior can be appreciated in online-only Data Supplement Movie I and II. SVC indicates superior vena cava; IVC, inferior vena cava; ASD, atrial septal defect; ScimV, scimitar vein; and RA, right atrium.
tion, and characterization of vessels. The derived high-resolution angiogram with isotropic spatial resolution depicts the altered cardiovascular anatomy with multiple partial anomalous pulmonary venous return veins and the hypoplastic right pulmonary artery (8 mm in diameter compared with 14 mm in the left pulmonary artery) in great detail without the need for intravenous contrast material (Figure 2; online-only Data Supplement Movie I). Furthermore, with these techniques, the velocity fields could be visualized and flow rates, directions, and volumes in any region of interest could be analyzed subsequent to the scan without the need for multiple 2-dimensional acquisitions. In this patient, flow quantification revealed a pulmonary-to-systemic flow ratio \((Q_p/Q_s)\) of 1.33, with contribution of the scimitar vein to the inferior vena cava equal to 0.42 L/min and a left-to-right shunt through the atrial septal defect equal to 1.34 L/min. Four-dimensional flow furthermore depicted the various contributions to right atrial filling and mixture, which were less organized than described previously.5

Without the need for an interventional procedure or multiple magnetic resonance acquisitions, in-depth visualization of hemodynamics by pathlines from the scimitar vein through the supradiaphragmatic inferior vena cava into the right atrium could be clearly separated from flow through the superior vena cava and atrial septal defect (Figure 3; online-only Data Supplement Movie II). Despite its thus far limited availability thus far and different clinical standards, including echocardiography, standard cardiovascular magnetic resonance imaging, computed tomography, and catheter angiography, the availability of anatomic and quantitative information from a single 5- to 10-minute acquisition could be especially suited for children with congenital cardiovascular abnormalities.

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None.

References
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