Anomalous Pulmonary Venous Return: Scimitar Vein

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Introduction: A scimitar vein (SV) is a rare congenital anomalous pulmonary venous return. Our case focuses on the pathophysiology of this disease and the transesophageal echocardiographic (TEE) findings.

Case Presentation: A 41 year old female with recurrent pulmonary infections presented to the OR for repair of a right sided anomalous pulmonary vein that emptied into the hepatic vein (HV) near the inferior vena cava (IVC). The SV was identified via intraoperative TEE by locating the SV at the level of the HV and IVC confluence. Color flow Doppler (CFD) demonstrated different flow velocities in the IVC, HV and SV. Pulse wave Dopler (PWD) revealed the characteristic HV waveform with S, D and a waves, while the SV, as a result of aortopulmonary malformations, had a more arterial-like waveform.

Post repair, 2D echocardiography, CFD, and PWD revealed a patent conduit connecting the SV to the left atrium and SV flow exclusion.

Discussion: Scimitar veins are rare congenital anomalies characterized by anomalous pulmonary venous drainage, partial to complete in nature, into the IVC(1). Angiography delineates the curvilinear scimitar sword appearance of the anomalous venous flow to the IVC with arteriogram and TEE exams confirming the diagnosis.

There is a paucity of published information that exists on TEE imaging of this pathology and due to the aberrant nature of these veins; imaging will vary from case to case. Some commonalities of scimitar syndrome are increased caval flow into the right atrium with dilation of right sided chambers, interatrial septal defects, a blunted pulmonary vein-left atrial confluence, and aortopulmonary collateral arteries(2,3,4).

The TEE examination may consist of imaging the right atrium using the bicaval view to evaluate and compare flows in the IVC versus the superior vena cava using PWD and CFD. A systematic scan of the IVC using 2D echo, PWD and CFD can be used to locate and compare flows of the IVC and SV. Reports have shown that the location at which the SV empties into the IVC may differ, ranging anywhere from the right atrium to the subdiaphragmatic IVC. A SV is best located by using CFD while scanning the IVC, looking for turbulent, nonlinear flow patterns(3). Depending on the type of surgical correction, which includes reimplantation of the scimitar vein to the left atrium or, as in our case, the use of an interatrial conduit, post-bypass TEE can confirm the re-establishment of normal pulmonary venous flow.

Scimitar vein pathology may be rare but knowledge of this physiology is important in the care of these patients and allows for a more accurate TEE examination which can aid in the management and surgical decision making process as well as confirm the patency and effectiveness of the surgical correction(4).

References:
1) Circ 2010;122(12):1159-66
2) Arch Inst Cardiol Mex 2005;75(2):165-9
3) JASE 1996;9(2):174-81
4) JASE 1998;11(4):387-92