

Persistent Left Superior Vena Cava with Absent Right Superior Vena Cava: Out of Mind is Out of Sight?

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Introduction

Persistent Left Superior Vena cava (LSVC) is the most common venous cardiac anomaly found in 0.3 - 0.5% [1-4] and in 4.3% of those with congenital cardiac anomaly. However, its association with absent Right Superior Vena Cava (RSVC) is rare (0.09 - 0.13%) [5]. It can be associated with situs inversus. It gives rise to increased risk of paradoxical embolism since it is associated with interatrial septal defect, unroofed coronary sinus or direct communication of the vein to left atrium [2]. A genetic culprit may be genes for left- right signaling [6]. Since it is a rare anomaly, the key is to keep it 'in mind' so that when we see it, we recognize it. We report a case of Persistent LSVC with absent RSVC discovered during pacemaker implantation.

Case

52 year old male with Marfan's syndrome, coronary heart disease and metallic aortic valve replacement presented with progressive shortness of breath and palpitations. He denied any chest pain, syncopal attacks or limb swelling. Surgical history included aortic valve replacement and cardiac catheterization. Medications were metoprolol and warfarin. Examination revealed a tall lean gentleman with bradycardia, hypertension, thoracotomy scar, displaced cardiac apex and systolic murmur with metallic click. His lungs were clear on auscultation and he had no pedal edema with intact peripheral pulses. Electrocardiogram revealed complete heart block. Hence he was deemed to be a candidate for pacemaker placement.

Intra-operatively, a persistent LSVC was found draining both right and left systems with an absent RSVC. (Figure 1) Leads were advanced through left brachiocephalic vein which drained into LSVC. They were further advanced into the coronary sinus and then fixed in the right atrium and ventricle. Normal sensing and capture were obtained confirming successful placement of a dual chamber pacemaker.

Discussion

Persistent LSVC is asymptomatic and discovered incidentally during cardiovascular imaging or surgery. During the formation of the

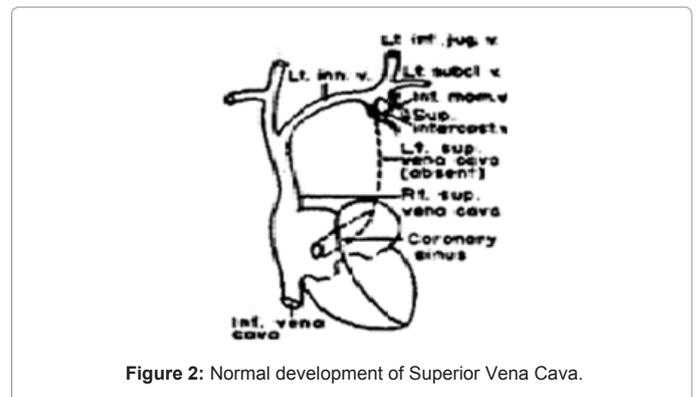


Figure 2: Normal development of Superior Vena Cava.

embryo, the superior vena cava is usually formed by the right common cardinal vein and the proximal portion of the right anterior cardinal vein. (Figure 2)

Persistent LSVC is a venous anomaly resulting from the persistence of the left anterior cardinal vein and obliteration of the common cardinal and proximal part of the anterior cardinal veins on the right [7] (Figure 3).

In our case, in addition to the persistent LSVC, the RSVC was absent. A prospective study of 2676 patients spanning over 15 years looked at the prevalence of persistent LSVC [8]. All of these patients were undergoing either pacemaker or cardioverter- defibrillator implantation. Persistent LSVC was found in 4 of 2676 (0.41%) patients and absent RSVC was found in only 4 of these 11 (0.36%) patients. LSVC persistence in these patients was thus similar to the general population. It is associated with conduction abnormalities and 40% of cases are associated with other structural congenital cardiac anomalies.

Venography, computerised tomography and magnetic resonance imaging can be used for diagnosis. Also the role of transthoracic saline echocardiography should be mentioned as a diagnostic tool of this extremely rare congenital anomaly [9]. It should be suspected when echocardiography shows 1) dilated CS confirmed by bubble study, 2) enhancement of CS prior to right atrium when contrast injected

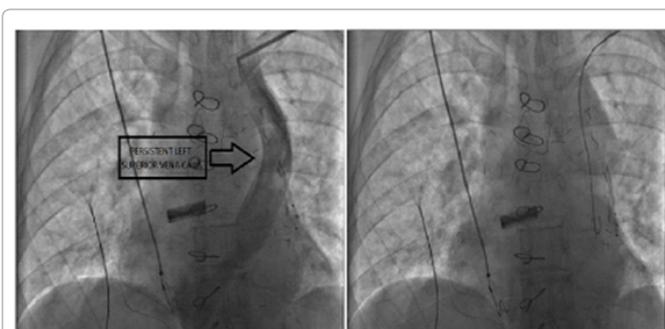


Figure 1: Pacemaker lead assuming an atypical path due to Persistent Left Superior Vena Cava.

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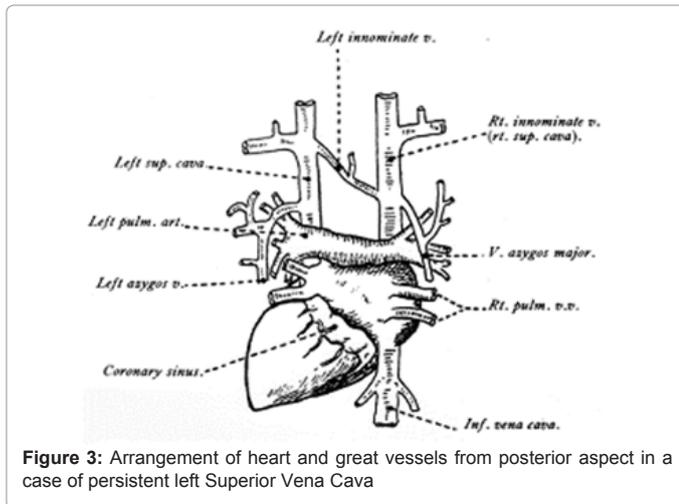


Figure 3: Arrangement of heart and great vessels from posterior aspect in a case of persistent left Superior Vena Cava

through left arm vein, while normal enhancement transit if injected through right arm. In another case report published in the Texas Heart Institute's Journal the coronary sinus was nearly 5cm in diameter and presented as a para-cardiac mass [10].

This anomaly is very elusive and can be easily missed during cardiac catheterizations and open heart surgeries [5]. Persistent LSVC should be suspected if a catheter or guide wire inserted via left subclavian vein takes an unusual left sided downward course. The sole access to the right side of heart becomes via LSVC which drains directly in the coronary sinus. Upon entering through the coronary sinus direction of movement of leads are away from the tricuspid valve and the septum rendering lead positioning and fixation in the right ventricle challenging and in some cases nearly impossible during pacemaker implantation. In such cases passive implantation has been described with intermittent loss of capture. Other complications include thrombogenesis and restriction in canula size. This can be overcome by use of hand shaped stylets and active fixation.

In the prospective study [8] the leads were placed from the left subclavian approach in 5 out of 7 patients receiving a pacemaker, 2 received an elective right sided approach due to physician preference. All patients had the device placed in the left pectoral region with a single-coil lead: defibrillator placement was successful in the long term in all but one patient, who required the addition of a subcutaneous array.

Conclusion

Awareness of embryological anatomical variants among physicians and sonographers is essential for recognition of persistent LSVC and absent RSVC. The left-sided implant required for placement of pacemakers and implantable cardioverter defibrillators in patients with persistent LSVC and absent RSVC may be arduous during lead placement; however, this task can be accomplished in the majority of cases, with reliable short-term and long-term outcomes.

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