

On Table Detection of Persistent Left Superior Vena Cava Draining into Superior Aspect of Left Atrium in a Case of Ostium Secundum Atrial Septal Defect - A Case Report

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Abstract: *Persistent Left Superior Vena Cava (LSVC) draining into coronary sinus is not a rare entity but LSVC draining directly into Left Atrium (LA) is very rare. It is quite often overlooked on transthoracic echocardiography and many a time escaped detection during right and left heart catheterisation. We present a case of ostium secundum (OS) atrial septal defect (ASD) where LSVC was present and it was draining directly into superior aspect of LA. This was diagnosed on table. Once it is identified, operative correction is mandatory to prevent right to left shunt and its known CNS complication.*

Keywords: Left Superior venacava, Atrial septal defect, Left Atrium

1. Introduction

The embryological development of systemic and pulmonary veins is very complex. The symmetrical cardinal veins gives rise to superior systemic venous channels while the splanchnic plexus of the foregut give rise to the pulmonary venous channels. Most of the left sided cardinal system disappears except coronary sinus which drains the cardiac veins. Many congenital variation have been described^{1,2}. One of the common variations is persistent LSVC draining into coronary sinus³. Very rarely LSVC can directly drain into LA resulting in systemic arterial desaturation⁴. This variation can most of the time associated with other congenital heart disease like ASD^{6,9}. When associated with ASD the clinical finding resembles those of ASD with only mild arterial desaturation. It can be overlooked on Echo as well as on cardiac catheterisation, as in our patient. But when it is diagnosed even on table it has to be treated to prevent its known potential risk of CNS complication.

2. Case Report

A 6 year old boy presented to us with chief complain of shortness of breath since 2 years of age. He also had frequent upper respiratory tract infection for the same duration. Shortness of breath occurs mostly on exertion and subsides on rest. It was not associated with any cyanosis or syncopal attack. He was born at term by normal vaginal delivery with no specific antenatal maternal illness or exposure to any teratogenic drugs. None of his brother or sisters or any family members have any cardiac disease. He was told by his physician of suspicion of some cardiac disease and was referred to us. We investigated him. His pulse was 90/min, B.P -110/76. No pallor, icterus or clubbing. The 1st Heart sound was slightly increased in intensity with wide fixed split 2nd heart sound. A grade 3/6 systolic murmur was present over left upper sternal border and a low pitched mid diastolic rumbling sound present at

the left lower sternal border. The ECG suggestive of right axis deviation and mild right ventricular hypertrophy. The chest X-ray revealed cardiomegaly with enlarged right atrium (RA) and right ventricle. 2D Echo study suggestive of enlarged right atrium and ventricle (volume Overload picture), a 28 mm ostium secundum type ASD with left to right shunt with ejection fraction of 70% and dilated main pulmonary artery.

He was posted for ASD closure. Median sternotomy done. Thymus dissected and brachiocephalic vein was found to be hypoplastic. SVC, IVC, LSVC and Aorta was cannulated and put on total CPB. RA opened parallel to RA groove. We inspected LA properly and was found an extra opening in LA just above LA appendage (Fig.1). The coronary sinus was normal. We decided to construct a pericardial baffle to route LSVC to RA since the innominate was hypoplastic. Pericardial patch was harvested which we started suturing to the superior aspect of the LA in such a manner (Fig.2) so that, opening of the LA appendage and all the pulmonary veins remain in the LA cavity proper and a tunnel at the superior aspect made by the pericardial baffle for draining of the LSVC into the RA. Another pericardial patch was taken and suturing done starting from the inferior margin of the ASD up to the mid portion of the defect and attached to the right margin of the baffle (Fig.3). He was weaned from CPB and tolerated the operative procedure well. His post-operative Pao₂ was 196 and saturation was 100%. He was discharged after 7 days and no complication occurred during his recovery and ward stay.

3. Operative Photographs

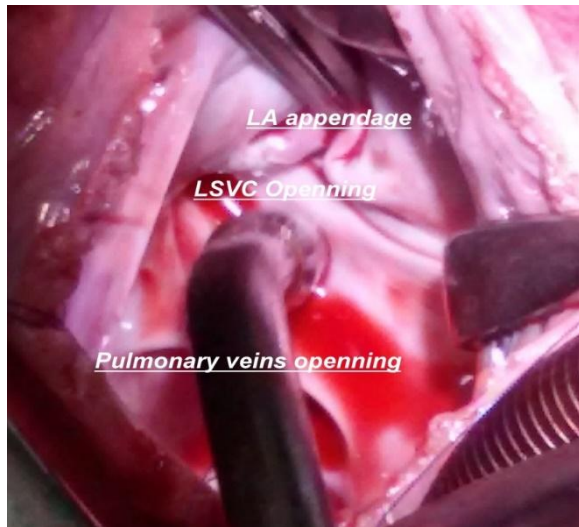


Figure 1: Showing the opening of LSVC, Pulmonary veins and LA appendage

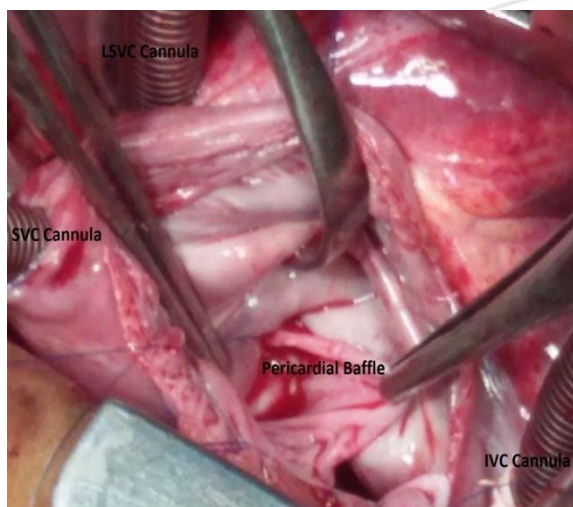


Figure 2: A tunnel made by the pericardial baffle is being created at the superior aspect of the LA for draining of the LSVC into the RA.

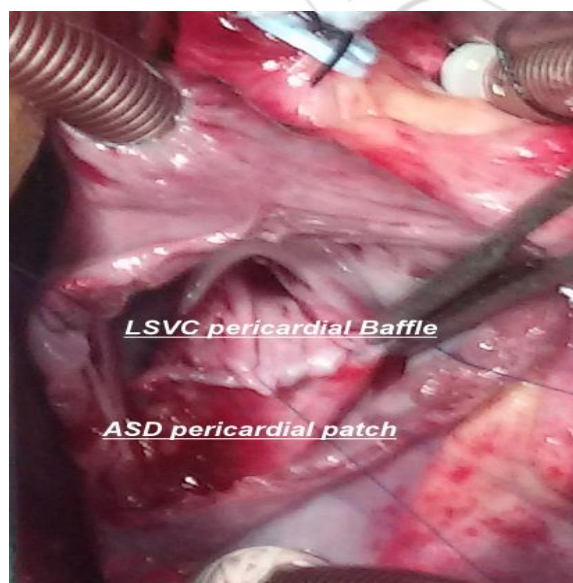


Figure 3: The pericardial patch used for closing the ASD is being sutured with the LSVC pericardial baffle thus completing the inter atrial septum.

4. Discussion

The incidence of persistent LSVC in general population is 0.35% and 3% to 10% in patients with congenital heart disease^{3,5}. Rarely may it present as isolated lesion. The embryology of the venae cava has been described by Campbell and Deuchar⁵. Embryologically the superior vena cava is formed by the right common cardinal vein and the proximal portion of the right anterior cardinal vein¹. LSVC is caused by the persistence of left anterior cardinal vein². When LSVC is present, it most commonly drains into coronary sinus⁷, but in around 7.5% of cases it drains directly into LA⁴. Persistence of LSVC is of little surgical significance unless it enters the left atrium giving rise to left to right shunt. In most cases this does not produce any obvious clinical symptoms except some variable degree of cyanosis but some serious potential complications can occur attributed right to left shunt, e.g., risk of embolism to CNS and brain abscess make operative correction very necessary. Most commonly persistent LSVC is associated with ASD as in our patient^{3,6,9}.

Mostly patients do not give any history related to persistent LSVC opening in LA, only clue clinically is mild cyanosis. Our patient does not have clinical cyanosis or any significant arterial oxygen desaturation at rest. It can be easily missed on echo as happened in our case. It can be recognized only by cardiac catheterisation if done through the left arm. Mild oxygen desaturation in a peripheral artery or left heart chamber would also suggest the presence of right to left shunt.

When opening of persistent LSVC in LA is identified then operative correction is mandatory to prevent a right to left shunt and its known CNS complication. There are many ways of interrupting the LSVC into the LA. Simple method is to ligate the LSVC but there must be an adequate innominate vein connecting both left and right SVC and also after demonstrating no rise in left jugular pressure than normal on temporary occlusion of the LSVC. Several other procedures have been explained to tackle this abnormal shunt such as intra atrial roofing, intraatrial baffle rerouting, reimplantation into RA or pulmonary artery and graft interposition to the right atrium. In our patient we constructed a pericardial baffle to route venous blood to the RA.

5. Conclusion

A persistent LSVC draining directly into LA is very rare entity but clinically significant condition. It can be easily missed on echo and even during cardiac catheterisation. When identified, even after initiation of bypass, its surgical correction is mandatory to prevent its potential complication such as brain abscess, risk of embolization to CNS and rarely systemic cyanosis.

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