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Persistent Left Superior Vena Cava, Connected to the Left Atrium via the Left Superior Pulmonary Vein. An Unusual Cause of Air Lock

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Abstract

The intraoperative diagnosis of a persistent left superior vena cava (LSVC) that drains into the left atrium (LA) via the left superior pulmonary vein may be difficult because of its extrapericardial pathway. We report here the case of a 48-year-old man, operated for a mitro-aortic endocarditis complicating a Laubry-Pezzi syndrome. The opening of the LA was followed immediately by a sudden air lock. It was only after opening of the left pleura that we were able to find and then ligate the LSVC. The patient underwent mitral valve repair, closure of the infundibular septal defect and aortic valve replacement.

Keywords: Persistent left superior vena cava, Cardiopulmonary bypass, Air lock

1. Introduction

Persistent left superior vena cava (LSVC) that connects with the left atrium (LA) through a left superior pulmonary vein (LSPV) is an exceptional systemic venous drainage anomaly. In the present case report, we describe this extra pericardial communication, discovered intraoperatively following a sudden air lock of the cardiopulmonary bypass (CPB).

2. Case report

A 48-year-old man was referred for surgical treatment of mitro-aortic endocarditis, complicating a Laubry Pezzi syndrome. Transthoracic echocardiography objectified severe aortic valve regurgitation with mobile vegetations, left-to-right shunt through an infundibular ventricular septal defect (VSD) (Fig. 1), severe mitral valve regurgitation; a poor left ventricular ejection fraction (40%), and a pulmonary hypertension (72 mmHg).

The patient was operated via a median sternotomy, and CPB was established after an aortobicaval cannulation. Cardiac arrest was induced by cold blood cardioplegia, delivered through the coronary ostia. Vegetations and damaged aortic valve were excised and closure of the infundibular VSD was performed using an autologous pericardial patch. The opening of the LA was followed immediately by the entrance of high volume of air bubbles into the superior vena cava cannula which resulted in a sudden air lock of the venous outflow line. After a multiple slowings and cessations of pump flow, partial clamping of this cannula, resulted in flooding of the LA with venous blood. Since the luxation of the heart did not show a LSVC in its usual location, we widely opened the left pleura for further exploration. This allowed us to discover an exceptional variety of LSVC draining into the LSPV, which enters the pericardium to drain into the LA (Fig. 2). As this LSVC communicated with a left innominate vein of normal size (Fig. 3), we ligated it at both ends. This allowed full CPB, repair of the mitral

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valve using Alfieri's edge to edge technique, and implantation of a mechanical aortic valve prosthesis. The patient was discharged 10 days later. He was eupneic and had no neurological deficit. Transthoracic echocardiography objectified a residual mild-to-moderate mitral valve insufficiency; a completely closed VSD and no dysfunction of the aortic valve prosthesis. His left ventricular ejection fraction improved (51%) and his pulmonary pressures dropped considerably (36 mmHg).

3. Discussion

The LSVC is more common than is often assumed. It affects up to 10% of patients with congenital heart disease [1]. Typically it descends vertically, anterior, and to the left of the aortic arch and main pulmonary artery. It enters the pericardium then runs in the posterior atrio-ventricular groove [2].

In 90% of cases, the LSVC has no hemodynamic consequence, since it continues with the CS, or more rarely with the inferior vena cava or the hepatic veins, which drain into the right atrium [2].

In less than 10% of cases, the CS is unroofed and the LSVC drains directly into the LA [3]. Exceptionally, as described in this report, the LSVC do not enter the pericardium and drains directly into the LSPV which is normally connected to the LA, thus causing a right-to-left shunt [4].

Abbreviations

CPB	Cardiopulmonary bypass
CS	Coronary sinus
LA	Left atrium
LSPV	Left superior pulmonary vein
LSVC	Left superior vena cava
VSD	Ventricular septal defect

This anomaly should be carefully differentiated from a partial anomalous pulmonary venous drainage of the left upper lobe. In this case, the LSPV is not connected to the LA and drains directly into the persistent LSVC, thus causing a left-to-right shunt [5].

The use of transesophageal echocardiography with microbubble contrast agent application through the left arm intravenous line, can ascertain the presence of this right-to-left shunt. In our patient, microbubbles would have entered LA by the LSPV, mimicking intrapulmonary shunt [6]. Computed tomography or magnetic resonance imaging of the chest are recommended if persistent LSVC with atypical left atrial drainage is suspected.

When the left innominate vein is hypoplastic or atretic, a persistent LSVC connected to the LA, is usually responsible for flooding the operating field with venous blood occurring at the opening of the LA. However, in the case of duplicate superior vena

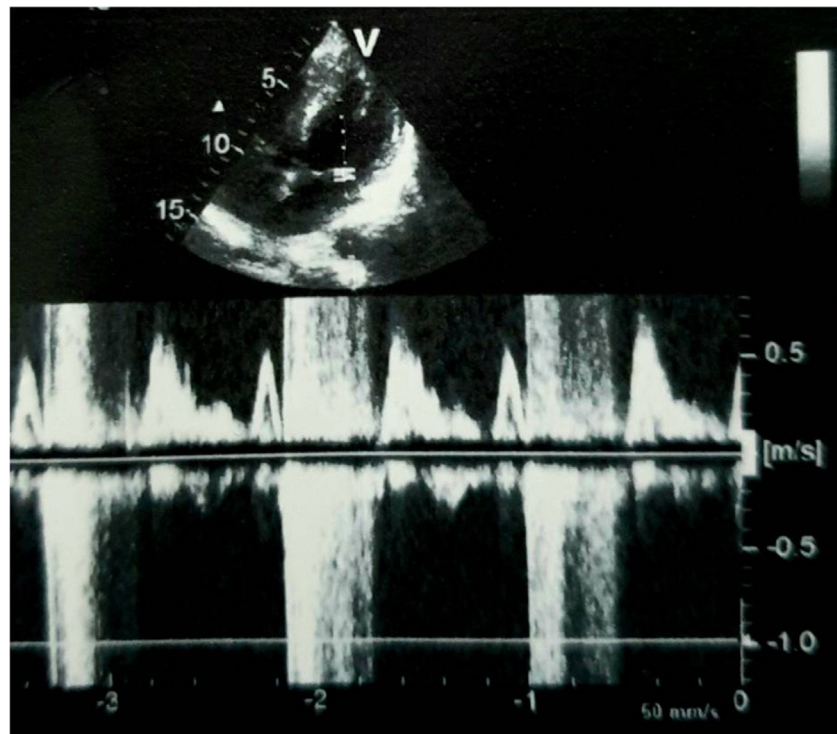


Fig. 1. Transthoracic echocardiography showing a ventricular septal defect (VSD), and a severe mitral regurgitation.

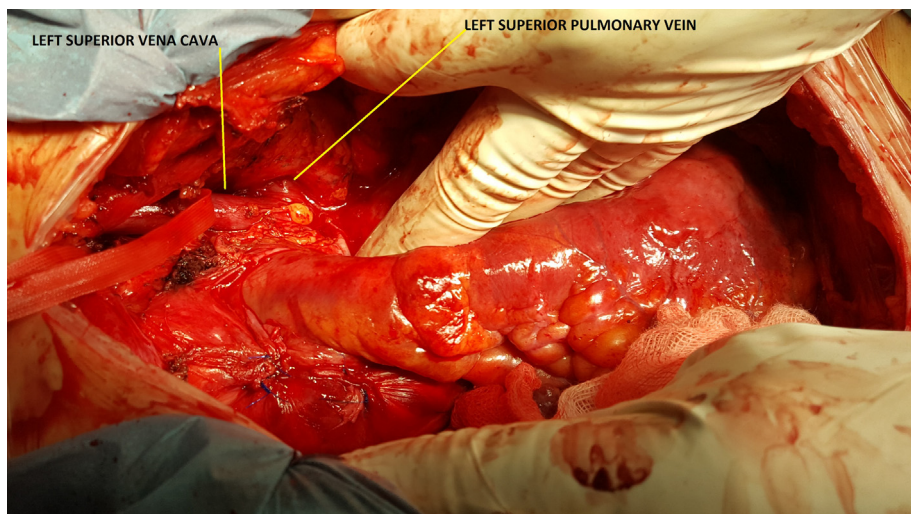


Fig. 2. Connection between the left superior vena cava (LSVC) and the left superior pulmonary vein, which drains into the left atrium. Exposure of the totally extrapericardial path of the LSVC after wide opening of the left pleura.

cava, with an innominate bridging vein of normal size, the negative pressure generated by the siphon effect of the CPB venous drainage, can lead from the opening of the LA to a massive air entry into the venous line, thus causing a sudden air lock.

Therefore, after excluding the usual sources of air entry in the venous outflow line (loose atrial purse string, atrial tear, and open intravenous access) we must first luxate the heart to look for a persistent LSVC which usually ends in the LA, between the left appendage and the left pulmonary veins. If this maneuver does not find it in its usual location, as was the

case for our patient, one should not omit to open the left pleura widely in order to look for the extrapericardial variety of the LSVC draining directly into the LSPV.

This exceptional anatomical variety must be known because the recurrence of air lock it causes, and the multiple slowings and cessations of pump flow that may result, can lead to irreversible brain and kidney damages, particularly in normothermic CPB.

Surgery of the LSVC depends on whether or not it communicates with the right superior vena cava but also on how it enters the heart chambers:

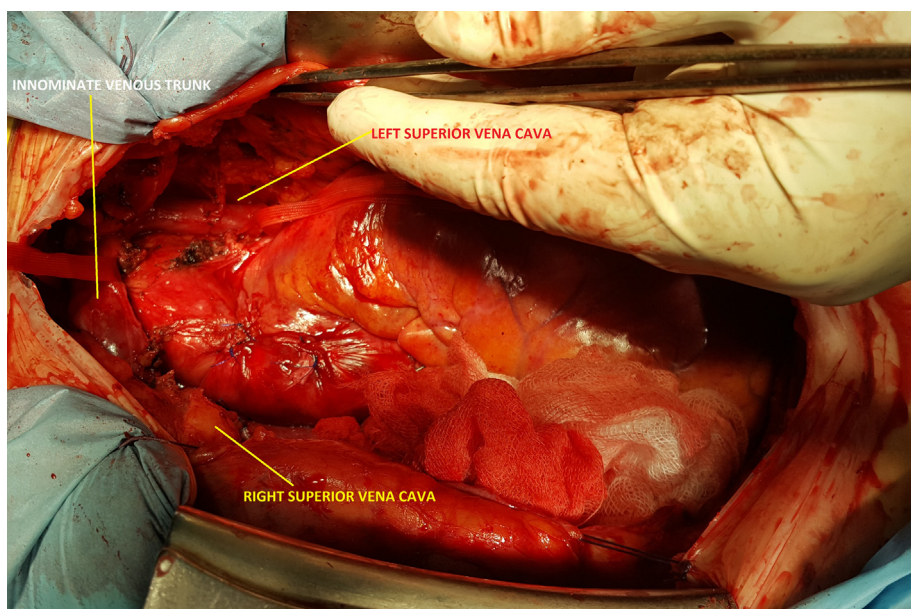


Fig. 3. Connection between the left superior vena cava and the innominate vein, which is of normal size. Note the presence of a right superior vena cava draining into the right atrium.

- When the two superior venae cavae communicate via a left innominate vein and the LSVC drains into the right atrium through a CS, it must be respected if a right atriotomy is not required. Otherwise, it is often necessary to clamp it temporarily, or to cannulate it through the CS.

On the other hand, if the LSVC drains into the LA, its ligation before initiating the CPB is necessary.

- If the two venae cavae are not connected, a LSVC draining into the right atrium must be respected if there is no need to open this chamber. Otherwise, its clamping is prohibited and therefore the LSVC must be cannulated, either through the CS or directly on its extracardiac path. In the case of a LSVC draining into the LA, correction can be achieved by rerouting LSVC flow into the right atrium using intra-atrial baffle techniques [7] or extracardiac anastomosis [8] between the LSVC and (a) the right atrial appendage, (b) the base of the right superior vena cava, or (c) the left pulmonary artery.

4. Conclusion

An undiagnosed LSVC connected to the left atrium is usually responsible for flooding the left atrium and operating field with venous blood, but can also cause a sudden air lock in case of duplicate superior venae cavae, with an innominate bridging vein of normal size.

The LSVC must be sought not only inside but also outside the pericardium, after a wide opening of the left pleura, as is the case of its extrapericardial variety draining directly into the LSPV.

Author contribution

Conception and design of Study: AB, MEAN. Literature review: AB. Acquisition of data: AB.

Analysis and interpretation of data: AB, MEAN. Data collection: AB. Drafting of manuscript: AB. Revising and editing the manuscript critically for important intellectual contents: AB. Data preparation and presentation: AB.

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Conflict of interest

Nothing to declare.

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