Persistent Left Superior Vena Cava (PLSVC) connected to the left atrium

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Abstract: A persistent left superior vena cava draining into the left atrium was diagnosed in an adult patient, scheduled for surgical correction of a large inferiorly located sinus venosus atrial septal defect. In the majority of cases a persistent left superior vena cava is found incidentally and causes little or no symptoms. Nevertheless, anaesthesiologist should be aware of its occurrence, because of different technical difficulties and clinical problems that can be encountered. Echocardiography plays an key role in the detection of a persistent left superior vena cava.

Key words: Persistent left superior vena cava; atrial septal defect.

INTRODUCTION

Atrial septal defect is a common congenital heart defect. It can be classified into primum, secundum, and sinus venosus defects. Sinus venosus atrial septal defect (SVASD) encompasses 4%-11% of all atrial septal defects, mostly of the superiorty located type (1).

Persistent left superior vena cava (PLSVC) is the most commonly seen systemic venous anomaly with an overall incidence of 0.5% to 2%, and in association with congenital cardiac disease, an incidence of up to 10% (3). In a minority of cases the PLSVC is connected to the left atrium.

In this report we describe the rare association of a large inferiorly located SVASD and a PLSVC draining into the left atrium. We focus on the importance of a PLSVC in anaesthesia practice.

CASE REPORT

A 28-year old woman was hospitalized and treated for a community-acquired pneumonia. On physical examination a heart murmur was noted. Transthoracic echocardiography revealed an inferior SVASD, an important dilatation of the right atrium and ventricle, mild regurgitation of the tricuspid valve and a preserved left ventricular function. Additional MRI imaging showed a PLSVC connected to the superior part of the left atrium, the absence of the innominate vein and coronary sinus, and a normal pulmonary venous connection.

The patient reported minor exertional dyspnea already present since childhood. Her further medical history was limited: at 10 months she underwent surgical correction of a neck abscess.

The arterial saturation of the patient was 91% on room air. The exercise intolerance and cyanosis suggested a large shunt and as a consequence the patient was planned for surgical repair.

After induction of anaesthesia a transoesophageal echocardiography (TEE) was performed, which confirmed the transthoracic echocardiographic findings. Additionally, agitated saline contrast, injected from the left arm, first appeared in the left atrium, demonstrating the presence of a PLSVC draining into the left atrium. To avoid an aberrant left-sided course, the central venous line was placed in the right internal jugular vein.

A median sternotomy was performed, resulting in a successful correction of both anomalies.

The intra- and post-operative clinical course was uneventful. The patient was rapidly extubated during an overnight stay on the intensive care unit. The patient left the hospital in good clinical condition, after a seven-day stay. Echocardiographic follow-up showed a normal left ventricular function and an already slightly better right ventricular function. Six weeks later, she had recovered well, reporting no more dyspnea.


**DISCUSSION**

Atrial septal defect is the second most common congenital heart defect in children and adults, with an incidence of 2.1 per 1000 live births, more frequent in females than males. SVASD encompasses 4%-11% of all atrial septal defects (1). It may be asymptomatic in childhood, but may become symptomatic with age. Unrepaired SVASD eventually leads to right heart volume overload and can ultimately lead to hypertensive pulmonary vascular disease (H PV D ) (2). Arrhythmia and paradoxical embolism are potential complications of SVASD. The considerable long-term risks for HPVD development and the presence of symptoms were the key factors in the decision for surgery.

During preoperative workout, another congenital anomaly was found: a PLSVC draining into the left atrium. PLSVC is the most common systemic venous anomaly with an estimated incidence of 0.5% to 2% in general population and up to 10% in patients with other congenital heart defects (3). Almost 40% of the patients with PLSVC have associated cardiac anomalies, such as atrial septal defect, bicuspid aortic valve, coarctation of aorta, absence of coronary sinus, and cor triatriatum. During embryonic development the venous blood of the upper part of the body drains into the right atrium via bilateral and symmetrically running veins: the right and left anterior cardinal veins. During the 8th week of gestation the left brachio-cephalic (innominate) vein develops as a bridge between the left and right anterior cardinal veins. The cephalic portion of superior cardinal veins forms the internal jugular veins, while the caudal portion of right superior vein forms the normal right-sided superior vena cava. The portion of the left superior cardinal vein caudal to the innominate vein normally obliterates to form the ligamentum of Marshall (5). When this process of regression doesn’t occur a left superior vena cava persists.

In approximately 90% of PLSVC a right superior vena cava is present. A bridge between these two superior cava veins, the innominate vein, is observed in 35% of the cases (4). There are variations in the insertion of the PLSVC: in 8% the PLSVC is connected to the left atrium, resulting in a right-to-left shunt (6). In the remaining cases it is connected to the right atrium, either directly or through a dilated coronary sinus (7). The diagnosis of a PLSVC is nearly almost incidental, mainly during anaesthesiological or cardiovascular procedures, or during diagnostic exams like echocardiography, CTscan or MRI (6). On TEE (Fig. 1) the coronary sinus is reliably visualized in a modified Four Chamber view, focussing on the right atrium and coronary sinus. The maximum normal diameter of the coronary sinus is less than 10 mm. In figure 2 a dilated coronary sinus is shown. When injecting contrast, it travels from the left arm to a persistent left vena cava draining into the coronary sinus. On chest X-ray widening of the aortic shadow, para-median bulging and a para-median strip or crescent along the left heart border can be encountered (8).

Most of the patients are usually asymptomatic or minimally symptomatic. PLSVC has clinical importance in many ways. When a left superior approach is used for insertion of a central venous line, Swan-Ganz catheter or for implantation of a pacemaker or a cardioverter-defibrillator, serious complications can occur. Although with a relative low incidence, arrhythmia, cardiogenic shock, cardiac tamponade, and coronary sinus thrombosis have been reported (4). PLSVC can be associated with structural abnormalities of sinus node, AV node or His Bundle. Cases of Wolf-Parkinson-White syndrome, sick sinus syndrome and even sudden death have been described (7). The radiographic appearances produced by insertion of a central venous line into a PLSVC may be mistaken for arterial placement, location outside the central venous circulation or an entirely extravascular site. Blood-gas analysis and echocardiographic examination of a central venous line whose precise location is not obvious is recommended (8).

If the PLSVC is draining into the left atrium, a right-to-left shunt is present, causing unexplained cyanosis or clubbing (8). A right-to-left shunt also forms a theoretical substrate for paradoxical
embolism, which may result in stroke or the possible development of intracranial abscesses (9).

Because of the previously described association, echocardiography is advisable to detect a PLSVC in all patients undergoing cardiac surgery for congenital anomaly. In the majority of cases of a PLSVC, a dilated coronary sinus is visualized. Diagnosis can be confirmed by contrast echocardiography (Fig. 2): saline contrast injected into a left arm vein passes through the PLSVC into the coronary sinus and the right atrium (10) or the left atrium, when the PLSVC is draining into the left atrium.

In summary, different specialists such as anaesthesiologists, intensivists, cardiologists and cardiac surgeons, should be aware of the potential clinical problems and technical difficulties caused by a PLSVC, especially when it is draining into the left atrium. In case of an aberrantly positioned central venous catheter, the existence of a PLSVC should be considered. Perioperative echocardiography is an important non-invasive diagnostic step in the detection of a PLSVC.

References


Fig. 2. — 2D-TEE: Modified Four Chamber view showing a dilated coronary sinus. The injected contrast travels from left arm into the coronary sinus through a persistent left SVC.