Persistent left superior vena cava: What an anesthesiologist needs to know?

Madam,

Persistent left superior venacava (PLSVC) is a venous anomaly of the thorax that may affect up to 2% of the general population. Absence of right-sided superior venacava with PLSVC is rare. PLSVC may be an isolated finding without any structural heart disease or may be associated with congenital heart diseases in up to 10% of cases, such as atrial septal defect, tetralogy of Fallots, cor triatrium, and mitral stenosis. It is the most common thoracic anomaly reported.

We discuss here the case of a 16-year-old boy admitted for anterograde dilatation of esophagus following trachea-esophageal fistula (TEF) repair presenting with severe dysphagia. Central venous access was required for poor peripheral venous access. After induction of general anesthesia, left internal jugular vein was canulated with a 7.5-Fr triple lumen catheter under ultrasound guidance for parenteral nutrition as the right internal jugular vein (IJV) was found to be thrombosed in Doppler imaging. The procedure was uneventful. A postoperative chest X-ray showed the catheter following a left para mediastinal course. A paired blood sample was obtained from the most distal port of the triple lumen catheter, and peripheral vein and blood gas analysis of these samples found that both sample had identical PO₂. Previous hospital records showed that the child had undergone a total correction of tetralogy of Fallot and operative notes mentioned the presence of a PLSVC opening into the coronary sinus along with a normally draining RSVC.

The embryological drainage of the heart is different from that of the adult heart. The cardinal system that drains the embryo heart undergoes partial resorption on the left side to form the Ligament of Marshall. If this remains patent, it forms the PLSVC and drains into the right atrium via the coronary sinus. If the right cardinal vein develops normally, the patient has a double SVC, which is the more common case. If the vein undergoes resorption (meaning, an absent right SVC), the right side venous blood drains via a brachiocephalic vein into the left SVC that ends in the left atrium either directly or through an unroofed coronary sinus in around 10% of the cases. This forms a right to left shunt. A schematic diagram showing development of LSVC has been depicted in Figure 2.

Figure 1: Chest X-ray showing central venous catheter in persistent left superior venacava

Persistent left-sided SVC almost always an incidental finding as 80% occur along with a right SVC. Almost 92% of PLSVCs drain into the coronary sinus and therefore into the right atrium. The first suspicion of a PLSVC usually arises with the incidental discovery of a dilated coronary sinus in a trans-esophageal echocardiography (TEE) done for some cardiac condition, or a pacemaker implantation, a left sided IJV cannulation, etc. The dilated sinus is seen in the posterior atrioventricular groove in the parasternal long axis view and the PLSVC is seen anterior to the left pulmonary artery in the parasternal short axis view. Agitated saline through the left peripheral vein appearing first in the coronary sinus followed by right atrium and ventricle confirms presence of PLSVC draining into the coronary sinus. This is contrary to that of normal patients where dye or bubble reaches the right atrium, followed by ventricle and at last the coronary sinus after getting filtered in the pulmonary bed. This can be done on table intraoperatively if a dilated coronary sinus is seen on TEE. Further, the blood gas analysis of samples taken will yield a VBG if the PLSVC ends on the right atrium.

Figure 2: A schematic diagram showing development of persistent LSVC
side circulation, and arterial or mixed if PLSVC ends in the left atrium.

PLSVC with a right SVC in situ is asymptomatic when not accompanied by other heart defects. Arrhythmias may occur with a PLSVC and absent RSVC though. The left atrial draining PLSVC can present as cyanosis. An undiagnosed PLSVC can complicate the placement of vascular access into the right side of the heart via a left subclavian approach, more often used in Swan-Ganz catheter placement and pacemaker implantation. Stimulation of the coronary sinus and hence the SA and AV node nearby can cause serious complications such as shock, cardiac arrest, angina, and perforation or rupture of the coronary sinus or brachiocephalic vein.

One should keep in mind the additional risk of a direct embolus into the systemic circulation in case of PLSVC draining into the left atrium.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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