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Standard variant of the systemic veins with significance for operations with cardiopulmonary bypass

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Case description

We report on a 5-year-old boy who was referred to our department as a newborn for evaluation of a heart murmur. Echocardiography revealed an atrial septal defect (ASD) of the secundum type (ASD II) (*Figure 1A*); a secondary finding was a significantly enlarged coronary sinus (CS) (*Figure 1B*).

An enlargement of the CS can be found in pathologies such as total anomalous pulmonary venous return, which is a severe congenital heart defect. It can also be found in standard variants of the systemic veins like a persistent left superior vena cava (pLSVC) with drainage into the right atrium via the CS.[1] During intrauterine development, there is a right and left anterior cardinal vein. Normally, the right SVC develops from the right anterior cardinal vein while the left anterior cardinal vein regresses. A pLSVC results from a failed regression of the left anterior cardinal vein. In most of these cases, the right SVC coexists, resulting in bilateral SVC with or without bridging vein. In a minority (0.09–0.13%), the right SVC does not develop and the venous blood of the upper body is solely drained through the pLSVC. In ∼90%, the pLSVCs drain into the CS, which leads to a dilatation of that same. This is however without haemodynamic implications.[1,2]

In the illustrated case transoesophageal echocardiography (TEE)-guided interventional closure of the ASD was targeted. However, the anatomy of the ASD did not prove suitable (*Figure 1A*); there was only an inadequate aortic rim, and the required device size was inappropriate for the patient's septal diameter and adjacent cardiac structures. For this reason, the therapeutic strategy was changed towards surgical closure of the defect. Computed tomography was performed before elective surgical closure of the ASD II to confirm the echocardiographic diagnosis of a pLSVC with agenesis of the right SVC (*Figure 1C* and *D*). This anatomical variant is of utmost interest especially if a minimally invasive approach is chosen, since the pLSVC cannot be reached for cardiopulmonary bypass (CPB) cannulation. Moreover, cardioplegia infused retrogradely through the CS would not be effective. In case of bilateral SVC, cannulation of only the right SVC due to undiagnosed pLSVC would result in excessive blood return via pLSVC and CS into the right atrium during CPB.[3]

Finally, after recommendation for surgical closure of the ASD, the patient was lost to follow-up until the age of 4.5 years. A reassessment of the echocardiographic findings at this time gave us reason to hope that interventional closure of the defect could be achieved. In the end, TEE-guided interventional closure of the ASD with an

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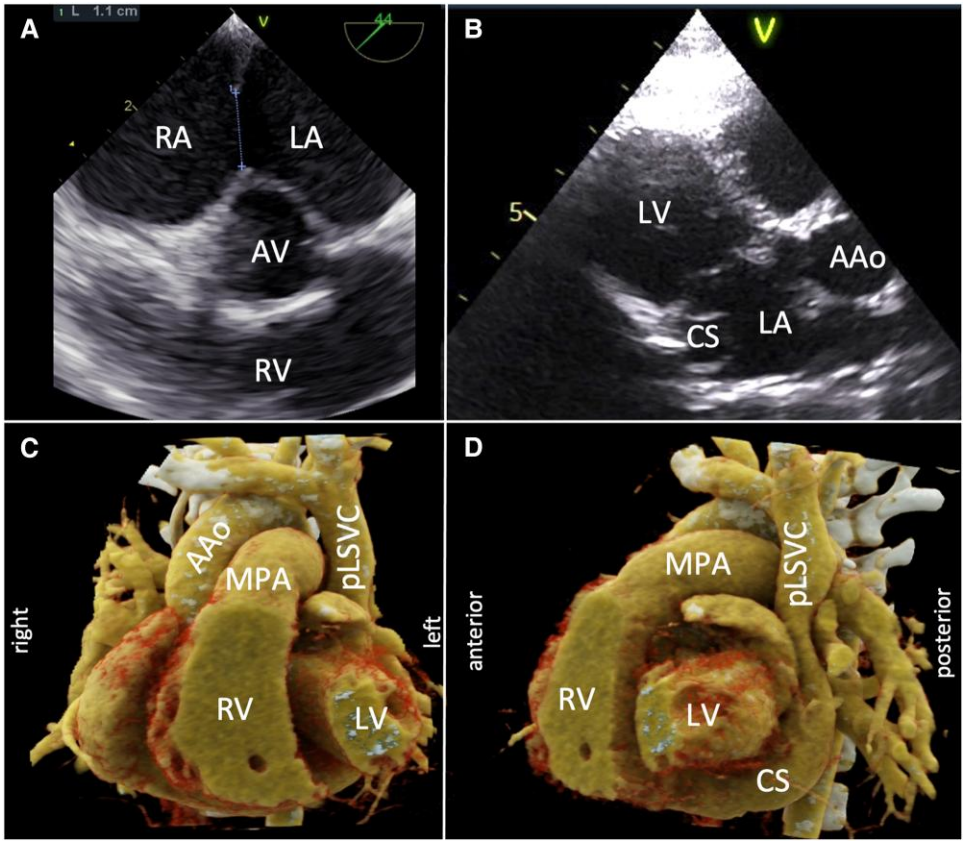

Figure 1 Transoesophageal echocardiography (modified mid-oesophageal aortic valve short-axis view, *A*)—atrial septal defect of 1.1 cm with almost no rim towards the aorta. Transthoracic echocardiography (modified long-axis view, *B*)—dilated coronary sinus can be seen in the left atrium. Computed tomography (coronary plane, *C*; sagittal plane, *D*)—persistent left superior vena cava with opening into a dilated coronary sinus and agenesis of the right superior vena cava. AAo, ascending aorta; AV, aortic valve; LA, left atrium; LV, left ventricle; MPA, main pulmonary artery; RA, right atrium; RV, right ventricle.

Amplatzer Septal Occluder (12 mm) was successfully performed. Follow-up did not show any residual shunt nor did any complication related to the Amplatzer Septal Occluder occur.

In conclusion, the anatomy of the systemic veins is of relevance for planning cannulation for CPB during paediatric cardiac surgery procedures.[2–4] Therefore, it is important to assess the anatomy of the systemic veins preoperatively.

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Data availability

All relevant data are presented within the manuscript.

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