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Raghib's syndrome with the absence of the right superior vena cava

Vena kava süperiyor yokluğunun olduğu Raghib sendromu

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A 40-year-old female diagnosed with a 42 mm atrial septal defect (ASD) (Figure 1a), drainage of the persistent left superior vena cava (PLSVC) into the left atrium (Figure 1b), and an absence of both the coronary sinus (Figure 2a) and right SVC (Figure 2b) was referred to our department. Cardiac catheterization showed the persistent drainage of the LSVC into the left atrium. During the catheterization, the mean pulmonary artery pressure (PAP) was 22 mmHg, and the volume of pulmonary flow (Qp) / the volume of systemic flow (Qs) was 1.5. The patient then underwent cardiac surgery. Arterial cannulation was performed from the ascending aorta and venous cannulation was carried out from the PLSVC and inferior vena cava (IVA). Under cardiopulmonary bypass (CPB), the ASD was closed with a pericardial patch, and the PLSVC was separated from the left atrium and anastomosed to the left pulmonary artery (LPA). The patient's postoperative course was uneventful. Five years have passed since the surgery, and she has no upper extremity or facial edema. Furthermore, the most recent control computed tomography (CT) angiography showed patent anastomosis (Figure 2b). In addition, control echocardiography detected a systolic PAP of 22 mmHg and clearly indicated an intact interatrial septum with drainage from the PLSVC into the LPA.

Raghib's syndrome is a developmental condition characterized by the combination of abnormal drainage from the LSVC into the left atrium, the presence of ASD, and the absence of a coronary sinus.[1] Surgical correction for Raghib's syndrome involves patching the ASD and draining the PLSVC into the right system either by dividing and reimplanting the PLSVC to the right atrium with a graft, using an intra-atrial baffle to divert flow from the PLSVC to the right atrium followed by anastomosing it to the LPA, or applying a simple ligation of the PLSVC.[2]

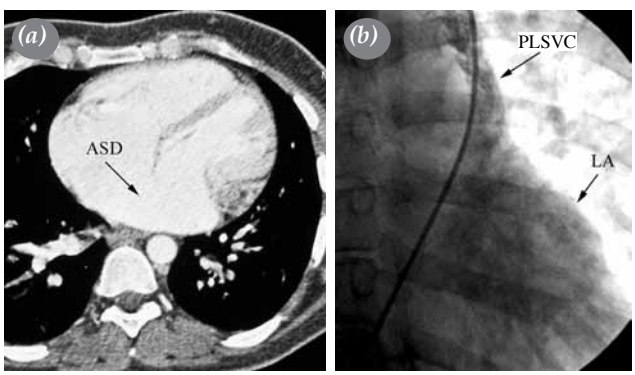

Figure 1. (a) Computed tomography showing the atrioventricular septal defect. **(b)** Cardiac catheterization indicating the PLSVC in the left atrium. ASD: Atrial septal defect; PLSVC: Persistent left superior vena cava; LA: Left atrium.

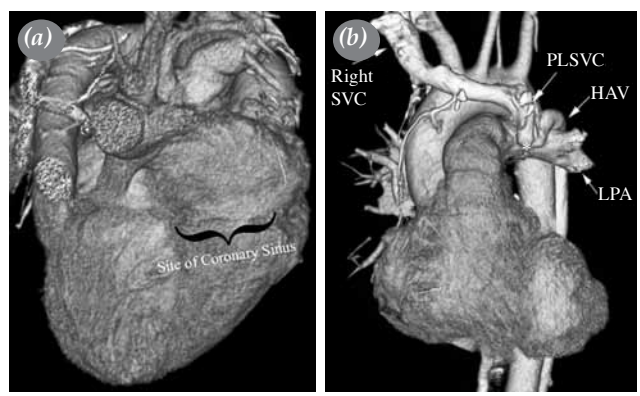

Figure 2. (a) Postoperative computed tomography revealing the absence of the coronary sinus. **(b)** Postoperative computed tomography showing the absence of the right SVC. The asterix indicates the anastomosis of the PLSVC and the LPA. SVC: Superior vena cava; PLSVC: Persistent left superior vena cava; HAV: Hemiazygos vein; LPA: Left pulmonary artery.

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We avoided anastomosing the PLSVC to the right atrium because of the risk of thrombosis. The patient had a large ASD, so it was not possible to divert flow from the PLSVC to the right atrium via the baffle. Hence, we chose to anastomose the PLSVC to the LPA. However, it is well known that pressures above 20 mmHg constitute a high risk for cava-pulmonary anastomosis, and the mean PAP was 22 mmHg for our patient. The increase in PAP depends on the shunt due to large atrial septal defect relatively was thought because Qp/Qs was 1.5. After banding the shunt, the PAP decreased.

According to our knowledge, our case was the first in the literature in which Raghib's syndrome was accompanied by the absence of the RSVC. The patient is currently doing well, and our results seems to confirm that anastomosis of the PLSVC to the right atrium was the most appropriate option for this case.

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