

## Absent right and persistent left superior vena Cava, interrupted Inferior vena cava: a case report

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#### ESC curriculum 2.1 Imaging modalities • 2.2 Echocardiography • 2.4 Cardiac computed tomography • 9.7 Adult congenital heart disease



**Figure 1** (A) Chest X-ray showing the absence of right superior vena cava shadow creating a notch (arrow) giving a clue pre-procedure; (B) transoesophageal echocardiogram showing large mal-aligned ostium secundum atrial septal defect with left to right shunt (arrow); (C) left superior vena cava (arrow) going inferiorly to coronary sinus; (D) common iliac vein (arrow) continuing as hemiazygos vein crossing to left of the spine; (E) hemiazygos vein (arrow) draining into left superior vena cava in turn into coronary sinus; (F) right brachiocephalic vein injection draining into left superior vena cava via connecting vein (arrow), right superior vena cava absent; (G) left superior vena cava angiogram with catheter course from right femoral vein to hemi-azygos vein to left superior vena cava, showing drainage of left superior vena cava to coronary sinus; (H) cardiac computed tomography showing right brachiocephalic (arrows) vein draining into left superior vena cava (arrow head) which in turn draining into coronary sinus.

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A 29-year-old asymptomatic gentleman was found to have atrial septal defect on routine check-up. His clinical examination corroborated large atrial septal defect with significant shunt. Electrocardiogram showed incomplete right bundle branch block, normal PR interval. Chest X-ray showed mildly increased pulmonary blood flow, mild cardiomegaly and conspicuous notching at right heart border raising suspicion of absent right superior vena cava (SVC; Figure 1A). Echocardiogram revealed normal left ventricular systolic function, mildly dilated right ventricle, right ventricular systolic pressure of 55 mmHg, 20 mm high ostium secundum atrial septal defect (OS-ASD) with left to right shunt and persistent left SVC (Figure 1B and C). During right femoral venous access, an interrupted inferior vena cava was documented as the catheter was progressing towards the left side of the spine (Figure 1D and Supplementary material online, Video S1), continuing up as left-sided hemiazygos vein (Figure 1E and Supplementary material online, Video S2) and draining into the left SVC in turn into coronary sinus (CS; see Supplementary material online, Videos S3–S6). Left and right innominate vein injection confirmed the absence of right SVC and the presence of left SVC, draining via CS (Figure 1F and G, and Supplementary material online, Videos S3 and S4). There was no un-roofing of the CS (see Supplementary material online, Video S7), and normal pulmonary venous drainage was evident (see Supplementary material online, Video S8).

Catheterization study showed  $Q_p/Q_s$  (pulmonary/systemic blood flow) 1.67:1, pulmonary vascular resistance 3.19 Woods unit, and pulmonary/systemic vascular resistance 0.1. Cardiac computed tomography showed normal veno-atrial concordance with hepatic veins

draining into right atrium. Inferior vena cava was interrupted with hemiazygos continuation draining into left SVC, no un-roofing of CS, no right-sided SVC, and no additional shunt lesions (*Figure 1H* and Supplementary material online, *Figure S1*). This case highlights the importance of tracing systemic venous drainage besides pulmonary venous drainage in a case of ASD to avoid cath-lab misadventures.

### **Supplementary material**

Supplementary material is available at European Heart Journal – Case Reports online.

**Consent:** The authors confirm that consent for submission and publication of this case report has been obtained from the patient in line with COPE guidance.

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#### Data availability

The data underlying this article are available in the article and in its online supplementary material.