Coronary sinus septal defect (unroofed coronary sinus): a case report

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Introduction: Coronary sinus (CS) atrial septal defect (ASD) is a congenital abnormality of both the atrial septum and the CS that falls within a wide spectrum of unroofed coronary sinus syndrome (URCS)[1]. The rarest type of ASD called 'isolated CS ASD' can be found in less than 1% of all ASDs. This case report aims to highlight the symptoms and diagnostic approach in an elderly patient with CS ASD.

Case report: We present a 60-year-old man who complained of moderate effort dyspnea lasting more than 12 months. He was treated for arterial hypertension, atrial fibrillation and had a history of pulmonary hypertension of unknown etiology. Physical examination showed an accentuated second heart sound over the pulmonary ostium with a systolic murmur. Transthoracic echocardiography showed pulmonary hypertension, right ventricular hypertrophy and dilatation, enlargement of both atria, a dilated coronary sinus and no visible atrial septal defects. Right heart catheterization revealed post-capillary pulmonary hypertension, with mean pulmonary artery pressure (mPA = 48 mmHg), pulmonary capillary wedge pressure (PCWP) of 25 mmHg, a significant left-to-right shunt (Qp/Qs = 2.5:1) and pulmonary vascular resistance (PVR) of 2 Wood units. Cardiac CT (**Figure 1**) showed a large communication around 3.3 cm in diameter between both atria as well as a dilated CS of 1.1 cm diameter. Transesophageal echocardiography (**Figure 2**, **Figure 3**) with bubble test disclosed a communication between the left atrium and the CS consistent with diagnosis of CS ASD without a persistent left superior vena cava (PLSVC). After intensification of diuretic therapy, follow-up catheterization 2 months later showed a reduction of PCWP (10mmHg) and mPA (25 mmHg) leading to successful surgical repair.

Conclusion: Patients with left-to-right shunts due to CS ASD are usually asymptomatic throughout adulthood. However, once symptoms occur, this congenital heart malformation remains often misdiagnosed. Therefore, we emphasize the importance of multimodal imaging in these patients[2].

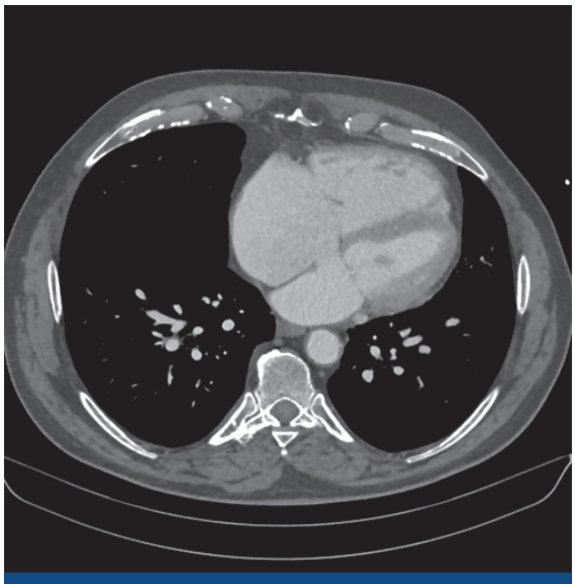

FIGURE 1. Heart computed tomography.

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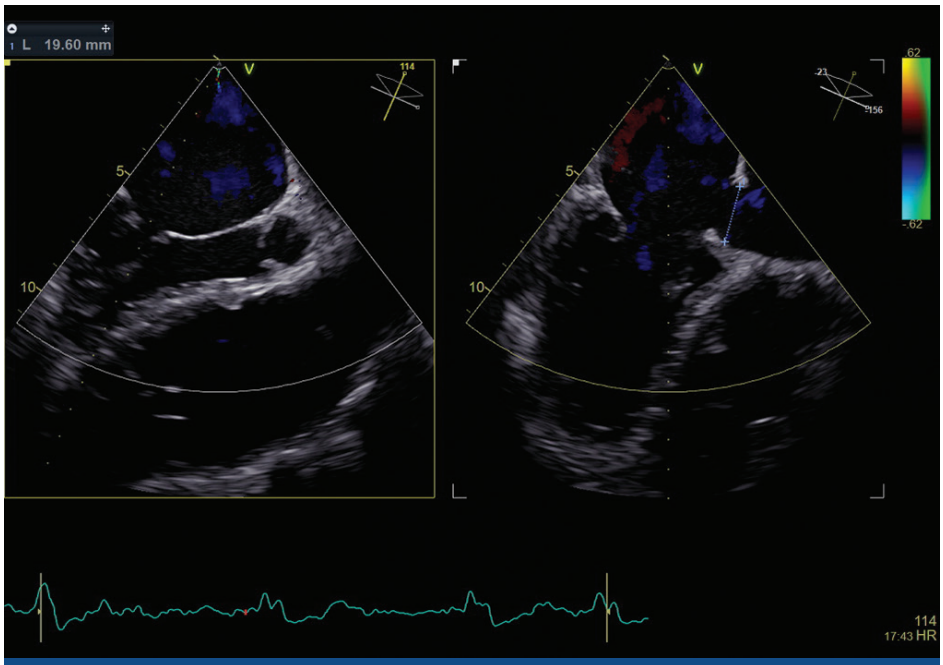

FIGURE 2. Transesophageal echocardiography.

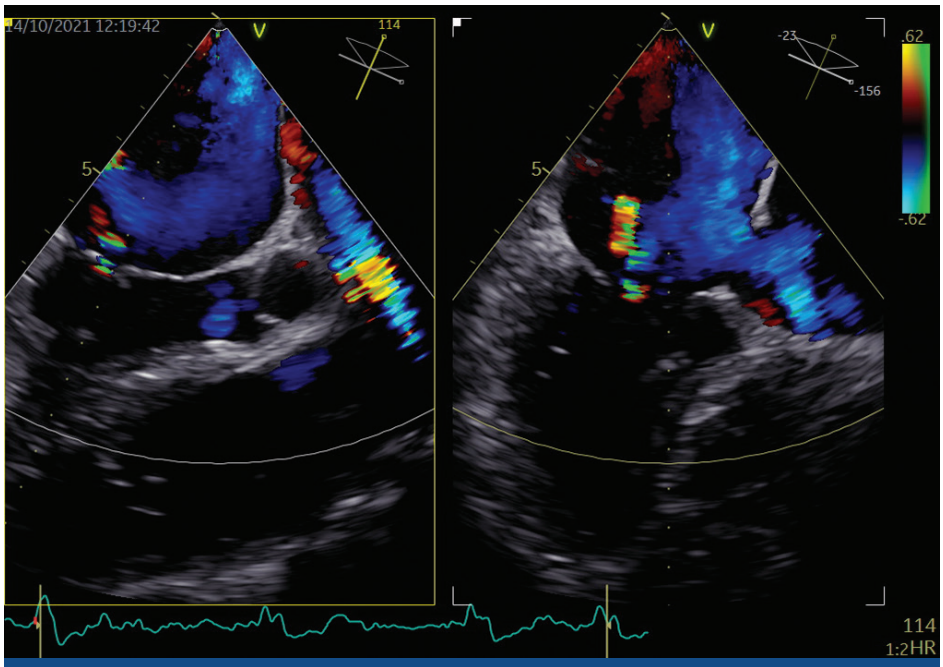

FIGURE 3. Transesophageal echocardiograph – color Doppler.

LITERATURE

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