

A 61-year-old woman presented with insidious onset of worsening dyspnoea on exertion for 2 years. Chest X-ray showed cardiomegaly (CTR=0.62) and hilar mass. She had been worked-up for a 'lung lesion' in China. Computed tomography (CT) imaging of the chest showed a dilated pulmonary artery, but no lung masses.

Subsequent echocardiogram showed a dilated right ventricle with moderate tricuspid regurgitation. There was pulmonary hypertension (estimated pulmonary arterial systolic pressure, 55 mm Hg). Transthoracic Doppler studies showed a left-to-right shunt with a shunt ratio of 2:1. However, the ductus arteriosus was not patent and there were no septal defects. The coronary sinus was dilated and there was suspicious communication between the left atrium and coronary sinus on a modified apical four-chamber view. 64-Slice CT was performed, which revealed an unroofed coronary sinus defect, a rare type of atrial septal defect with a left-to-right shunt (Figs 1 and 2). The pulmonary artery was grossly dilated. There was associated persistent left superior vena cava (LSVC). There was no anomalous pulmonary venous drainage. Cardiac catheterization confirmed the diagnosis of a completely unroofed coronary sinus defect with a LSVC. There was an associated patent foramen ovale. The patient underwent baffle repair surgery and correction of the LSVC. The postoperative period was uneventful.

Discussion

An unroofed coronary sinus defect is a very rare type of atrial septal defect, in which there is no partition between the left atrium and coronary sinus. The defect is frequently associated with a persistent LSVC that drains directly into the coronary sinus. Persistent LSVC occurs in 0.1 to 0.5% of the general population, with 8% draining into the left atrium. Unroofed coronary sinus defect is seen in over 70% of patients with a LSVC that drains into the left atrium.¹ Unroofed coronary sinus defect is difficult to diagnose by clinical signs and symptoms alone. It should be suspected in patients with persistent LSVC and a history of paradoxical embolism or brain abscess. Coronary sinus defect has been reported to occur with other congenital heart diseases, such as cor triatriatum, pulmonary atresia, tetralogy of Fallot and anomalous pulmonary venous drainage.²

The morphological type can be classified as: type I, completely unroofed with LSVC, as in our case; type II, completely unroofed without LSVC; type III, partial unroofed mid portion; and type IV, partial

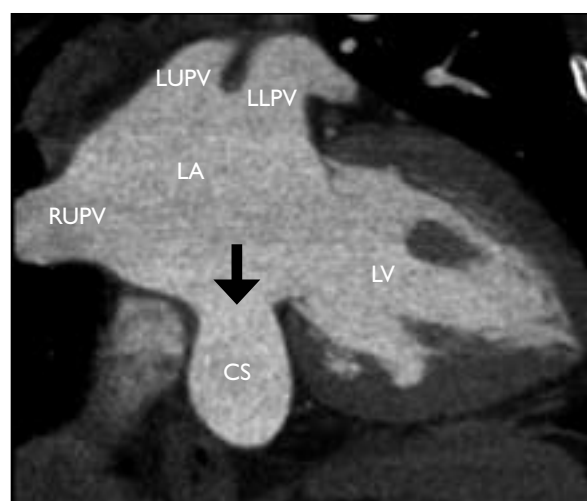


FIG 1. 64-Slice computed tomographic imaging of the heart (longitudinal two-chamber view) showing the unroofed coronary sinus defect (arrow)
CS denotes coronary sinus, LA left atrium, LLPV left lower pulmonary vein, LUPV left upper pulmonary vein, LV left ventricle, and RUPV right upper pulmonary vein

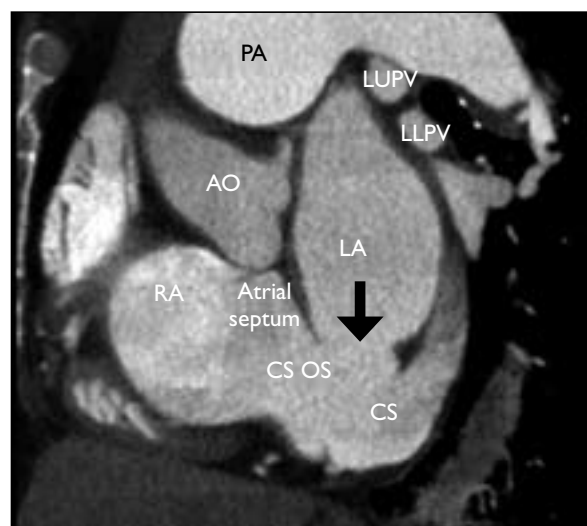


FIG 2. 64-Slice computed tomographic imaging of the heart (sagittal view) showing the left-to-right communication through the unroofed coronary sinus defect (arrow)
AO denotes aorta, CS OS opening of coronary sinus, LA left atrium, LLPV left lower pulmonary vein, LUPV left upper pulmonary vein, PA pulmonary artery, and RA right atrium

unroofed terminal portion.¹

Non-invasive imaging with 64-slice CT and cardiac magnetic resonance imaging can provide

excellent high-resolution imaging of the anatomy in cases of complex congenital heart disease.

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