Unroofed coronary sinus: A case vignette emphasizing the role of three-dimensional transesophageal echocardiogram

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DOI: 10.33963/v.kp.98404

Received:
October 10, 2023

Accepted:
December 4, 2023

Early publication date:
December 7, 2023

A 42-year-old man was admitted to our outpatient cardiac center for chronic exertional dyspnea. His medical history was unremarkable. Cardiovascular examination revealed a 2/6 systolic murmur at the lower left sternal border. An electrocardiogram showed normal sinus rhythm. A transthoracic echocardiogram showed normal biventricular systolic functions, mild tricuspid regurgitation, mildly enlarged right atrium (area: 21.4 cm^2) and right ventricle (basal diameter: 4.5 cm). Contrast echocardiography from the right antecubital vein showed a right-to-left transition of bubbles in the first three cycles (Figure 1A–B; Supplementary material, *Video S1*). Transesophageal echocardiography demonstrated marked turbulent flow in the right atrium from the coronary sinus (CS). There was a defect on the roof of the CS in the left atrium (1.5 × 0.7 cm) (Figure 1C–D; Supplementary material, *Videos S2–S6*). The patient was referred for surgical repair.

This case illustrates the challenging aspects of defining the dilatation of the right

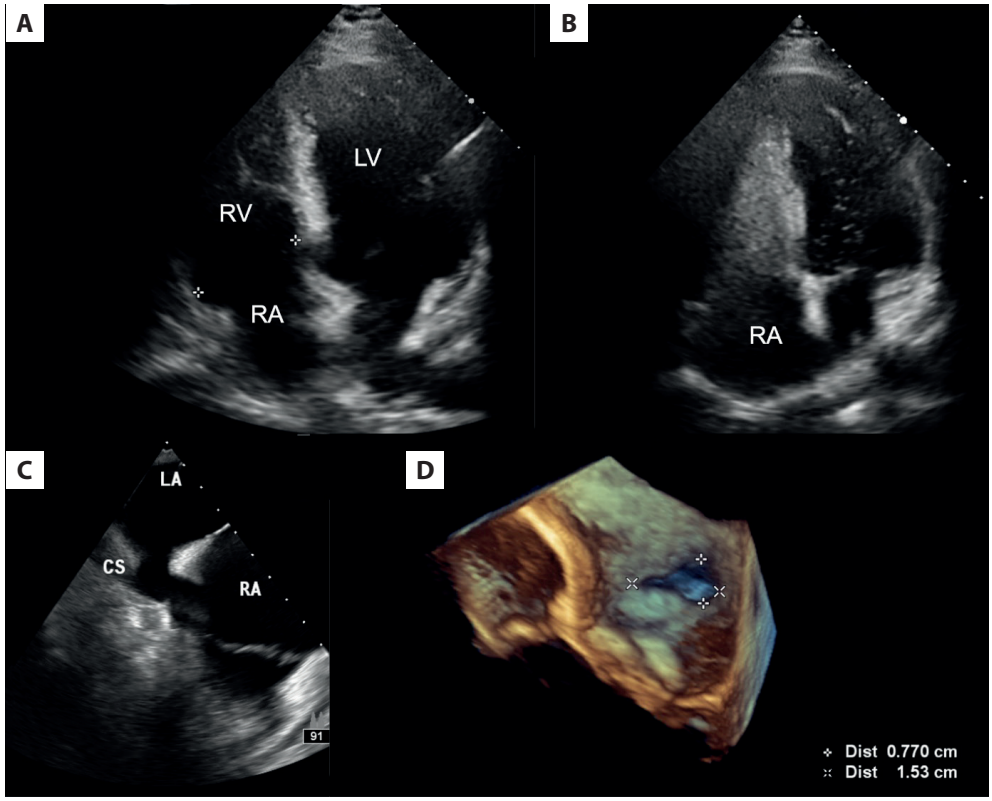

Figure 1. A. Apical four-chamber view demonstrates the mildly dilated right heart chambers. **B.** Contrast echocardiography from the right antecubital vein showed a right-to-left transition of bubbles in the first three cycles. **C.** Transesophageal echocardiography revealed a defect between the left atrium and the coronary sinus. **D.** Three-dimensional echocardiography provided visualization of the pear-shaped defect

Abbreviations: CS, coronary sinus; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle

heart chambers. Meticulous examination of the interatrial septum is crucial in finding a left-to-right shunt when the right ventricle is dilated. Unroofed coronary sinus is a rare congenital cardiac anomaly, characterized by a partially deficient or completely absent wall of the CS within the left atrium leading to a left-to-right shunt [1]. Because it can accompany a secundum atrial septal defect, it is crucial to fully delineate the anatomy before percutaneous intervention [2]. Computed tomography scans may not visualize the exact dimensions of the opening of the unroofed coronary sinus but additional abnormalities (i.e., persistent left superior vena cava or partial pulmonary venous return abnormality) can be delineated in detail [3]. Classically, patients with large unroofed coronary sinus are surgically corrected although a transcatheter therapeutic approach to the coronary sinus could be feasible [3–5]. Our Heart Team decided on the surgical correction due to the local expertise on congenital heart disease. The thorough assessment of the interatrial septum *via* a three-dimensional echocardiogram had a pivotal role in the diagnosis and management of the patient.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/kardiologia_polska.

Article information

Conflict of interest: None declared.

Funding: None.

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