Coincidence of cor triatriatum sinistrum and bicuspid aortic valve in an adult patient

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A 58-year-old female with uncontrolled arterial hypertension and obesity (body mass index [BMI], 35 kg/m^2) was admitted to the emergency department with elevated blood pressure (180/100 mm Hg) with concomitant dyspnea at rest. The patient had a 6-month history of worsening exertional dyspnea along with non-adherence to antihypertensive therapy which included ramipril 5 mg/day, nebivolol 5 mg/day, and doxazosin 4 mg/day. The patient discontinued amlodipine 5 mg/day due to intolerance. Upon admission to the hospital, physical examination showed mild lower extremity edema and a diastolic murmur on aortic valve auscultation. Lung auscultation was normal. The B-type natriuretic peptide and serum troponin I levels were within the normal range. Electrocardiogram presented sinus rhythm 75/min without ST-T changes. Blood pressure lowering therapy was administrated, and the patient was transferred to the Department of Cardiology for further assessment. Transthoracic and transesophageal echocardiography demonstrated preserved ejection fraction (60%) without left ventricle wall motion abnormalities, bicuspid aortic valve (BAV) with a fusion of non-coronary cusp and right-coronary cusp (Figure 1B and D; Supplementary material, *Video S2*, *Video S4*) with moderate central regurgitation jet (vena contracta width <0.4 cm, pressure half-time 380 ms), without any visible valve calcifications. The ascending aorta was dilated to 45 mm, while the size of the sino-tubular junction was 34 mm and the aortic root diameter at the Valsalva level was 38 mm. Left ventricular diastolic and systolic diameters were 51 mm and 44 mm, respectively. The left atrium (LA) was slightly enlarged (43 mm) and divided by a membrane into two parts with pulmonary veins draining into the "accessory" superior chamber of LA. The LA appendage without thrombus was connected with the "true" inferior chamber of LA (Figure 1A and D; Supplementary material, *Video S1*, *Video S3*). A wide single large opening in the intra-atrial membrane (Group 3 of Loeffler cor triatriatum classification [1]) was present in the inferomedial part of LA. Continuous wave Doppler across the orifice in the intra-atrium membrane did not reveal flow acceleration. Multislice computed tomography confirmed an intra-atrial membrane and a moderate dilatation of the aorta (Figure 1C and F). As coronary arteries were not sufficiently visible, coronary angiography was performed showing no coronary lesions. During the hospital stay, anti-hypertensive pharmacotherapy was optimized with good clinical response. It was assumed that the main pathology that was not sufficiently treated was hypertension, and other abnormalities were not responsible for the symptoms. At discharge, the patient was prescribed ramipril 10 mg/day, nebivolol 5 mg/day, doxazosin 4 mg/day, torsemide 5 mg/day, and spironolactone 25 mg/day for anti-hypertensive treatment. A periodic check of the aortic diameter and aortic regurgitation was scheduled. The 1-year follow-up was uneventful.

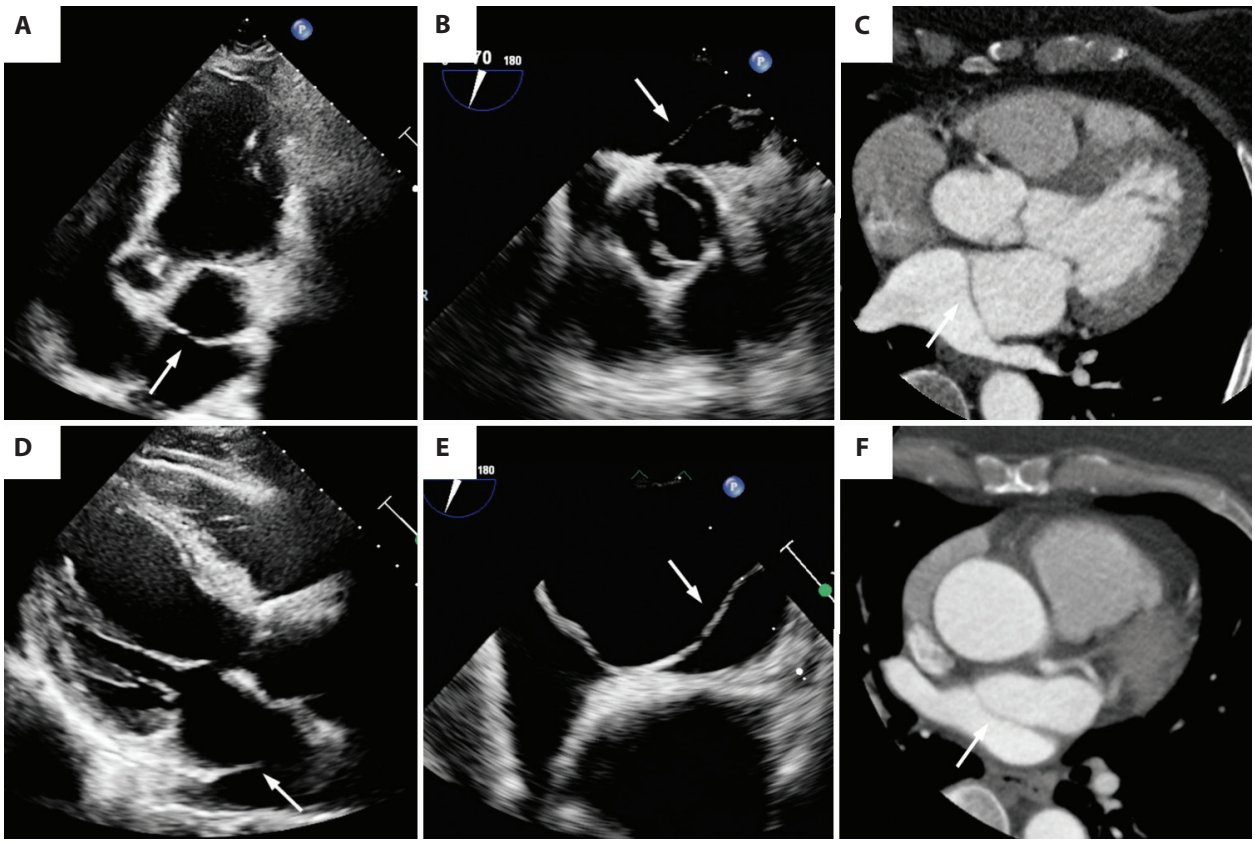

Figure 1. A. Transthoracic echocardiography — apical view. **B.** Transesophageal echocardiography — the bicuspid valve and left atrium membrane. **C.** Computed tomography angiography. **D.** Transthoracic echocardiography — long axis view. **E.** Transesophageal echocardiography — the left atrium membrane. **F.** Computed tomography angiography. The white arrow in all panels indicates the left atrium membrane

Cor triatriatum sinistrum (CTS) is a unique congenital abnormality with prevalence of 0.1%–0.4% in congenital heart disease [1]. It is usually corrected in childhood either surgically or percutaneously in highly symptomatic CTS group 1 (no connection between chambers) and 2 (small connection between chambers) according to the Loeffler classification [2]. It is incidentally recognized in adults in CTS group 3 (wide connection) [3]. The clinical presentation of CTS is similar to mitral stenosis. The coincidence of CTS with BAV is a unique finding — with only a few reports in adult patients available so far [4, 5].

Supplementary material

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

Article information

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