

# Complex subvalvular aortic stenosis, a bicuspid aortic valve with ventricular septal defect, patent ductus arteriosus, and aortic coarctation: A challenging combination

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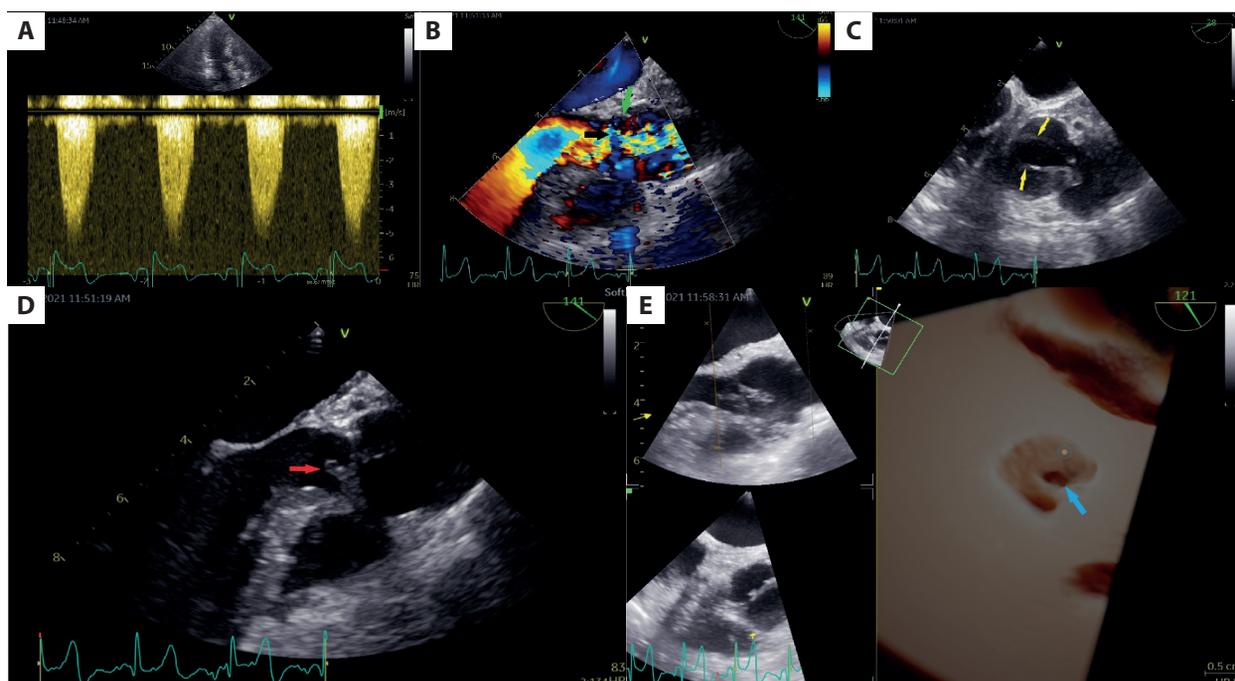
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Subaortic stenosis (SubAS) is frequently associated with aortic valve dysfunction and has to be differentiated from obstructive hypertrophic cardiomyopathy and Shone's complex [1–4]. Both bicuspid aortic valve (BAV) and coarctation of the aorta (CoA) often coexist (in 85% of cases) and may be related with subvalvular (SubAS), valvular, or supra- valvular aortic stenosis [1, 5].

We present a case of an 18-year-old man with a rare combination of SubAS, bicuspid aortic valve after surgical correction of ventricular septal defect (VSD), patent ductus arteriosus (PDA), and coarctation of the aorta in childhood. The patient was referred because of a symptomatic aortic stenosis: New York Heart Association class III and two episodes of syncope. Moreover, he had a history of mild atypical autism, Gilbert's syndrome, bone marrow hypoplasia, and thrombocytopenia (the mean platelets count was about  $91 \times 10^3/\text{UL}$ ). On physical examination, there was a loud, grade 5/6 systolic murmur best heard over the aortic area, which radiated to the carotid arteries. Clinical examination showed normal body development with short stature (150 cm) and low body weight (48 kg). The ECG revealed sinus rhythm, negative T-waves in leads I and aVL as well as features of left ventricular hypertrophy.

On transthoracic echocardiography, the heart chambers were of normal sizes with unclear morphology of the aortic valve; however, the transaortic systolic gradient was very high (Figure 1A; Supplementary material, Videos S1, S2). Calculation of the aortic valve area was not reliable because of the acceleration in the left

ventricular outflow tract (Figure 1B; Supplementary material, Video S3). Transesophageal echocardiography revealed a bicuspid, bicommissural aortic valve with slightly thickened valve leaflets (Figure 1C, Supplementary material, Video S4). The aortic valve area assessed by planimetry was 1.8 cm<sup>2</sup>. The membrane was partially fused with the base of the right coronary leaflet of the aortic valve in the left ventricular outflow tract (Figure 1D, Supplementary material, Video S5). In the posterior part of the membrane, an orifice with a surface area of about 0.4 cm<sup>2</sup> was visualized (Figure 1E). The patient was consulted by the Heart Team and qualified for surgical removal of the subvalvular membrane. The surgery risk estimated by EuroSCORE II (0.5%) was low. However, the STS Prom calculator, taking into account thrombocytopenia, immunodeficiency, and liver disease, already indicated an intermediate procedural risk (5.5%). The operation was successfully completed, and the native aortic valve was maintained. Unfortunately, in the postoperative period, the patient developed severe pneumonia with multiorgan failure leading to a fatal outcome. This is a unique case of a patient with congenital heart disease previously surgically corrected, with membranous subvalvular AS located very high in the left ventricular outflow tract, partially fused with the right-coronary leaflet of the aortic valve. This raised concerns about the risk of intraoperative damage of this leaflet during its membrane removal. It is important to be aware that there is a high probability of coexistence of multiple defects (in this case detected late) in patients with congenital heart disease [3].



**Figure 1.** **A.** Continuous wave Doppler – transaortic gradient:  $V_{max} = 5.24\text{m/s}$ ,  $PG_{max} = 110\text{ mm Hg}$ ,  $PG_{mean} = 68\text{ mm Hg}$ . Transesophageal echocardiography: **B.** Color Doppler — left ventricular outflow tract flow acceleration at the level of the hole of the subvalvular membrane (black arrow). Aortic valve — purple arrow. **C.** Bicuspid, bicommissural aortic valve with slightly thickened valve leaflets (yellow arrows). **D.** The membrane partially fused with the base of the right-coronary leaflet of the aortic valve (red arrow). **E.** 3D reconstruction — the hole in the posterior part of the subvalvular membrane (blue arrow)

### Supplementary material

Supplementary material is available at [https://journals.viamedica.pl/kardiologia\\_polska](https://journals.viamedica.pl/kardiologia_polska).

### Article information

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