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Partial atrioventricular septal defect: An unexpected anomaly in a 68-year-old patient

Short title: Partial atrioventricular septal defect

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A 68-year-old man consulted a cardiologist because of rapid fatigue, shortness of breath, a feeling of heaviness in the chest, and sweating. Occasionally, there are fainting spells and palpitations. He suffered a cerebrovascular insult 5 years ago, but he is currently without neurological consequences. On clinical examination, the first and second heart sounds were normal and a systolic murmur 3/6 was heard at the apex with radiation to the left axilla. Auscultation revealed a normal breathing sound, without pathological murmurs. The electrocardiogram showed sinus rhythm, frequency about 70/min, levogram, anterior left hemiblock, without signs of acute ischemia and myocardial lesions, without rhythm disturbances (**Figure 1A**). A chest X-ray was unremarkable. After transthoracic echocardiography (**Figure 1B–C**; Supplementary material, *Video S1*), transesophageal echocardiography was performed. An atrial septal defect of the ostium primum type with a cranio-caudal diameter of 15 mm and a cross-sectional area of 1.9 cm² was registered, through which a bidirectional but still dominantly left-right shunt was registered

(Supplementary material, *Videos S2–3*). An anterior mitral cusp cleft and moderately severe mitral regurgitation with an eccentric jet were seen (**Figure 1C–E**; Supplementary material, *Videos S3–S4*). The diameter of the mitral annulus was 3.9 cm. Moderate tricuspid regurgitation and pulmonary hypertension (right ventricular systolic pressure 59 mm Hg) were also registered. The medical documentation was shown to the heart team, who indicated the surgery. As part of the preoperative preparation, coronary angiography was performed. There were no significant lesions on the coronary arteries.

The intraoperative findings corresponded to the transesophageal echocardiography description. Through the sizeable defect of the atrial septum, a large cleft of the anterior mitral cusp was approached (**Figure 1F**) and repaired with a 4-0 Ticron suture, and the widened mitral orifice was repaired with an annuloplasty with a rigid band No. 32 (Supplementary material, *Video S5*). The atrial defect was closed with a 4 × 3 cm bovine pericardium (Supplementary material, *Video S6*). Tricuspid annuloplasty was performed, also. The postoperative course was uneventful, and the patient was discharged from the hospital on the seventh postoperative day.

A partial atrioventricular septal defect is a congenital heart malformation consisting of an ostium primum atrial septal defect and a cleft dividing the anterior mitral valve leaflet [1, 2]. There is a single atrioventricular valve annulus with two atrioventricular valve orifices (mitral and tricuspid), which share a common hinge point at the crux of the heart. There is no ventricular septal defect component. A partial atrioventricular septal defect is a form of endocardial cushion defect that is infrequent in adults, with only sporadic case reports published [3]. It is commonly repaired in the first years of life. The curiosity in this case is that the patient lived to his old age with this anomaly. So far, only a few patients who survived the sixth and seventh decades have been described [3]. To the best of our knowledge from the English-language literature, this is one of the oldest patients operated for this anomaly.

Supplementary material

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

Article information

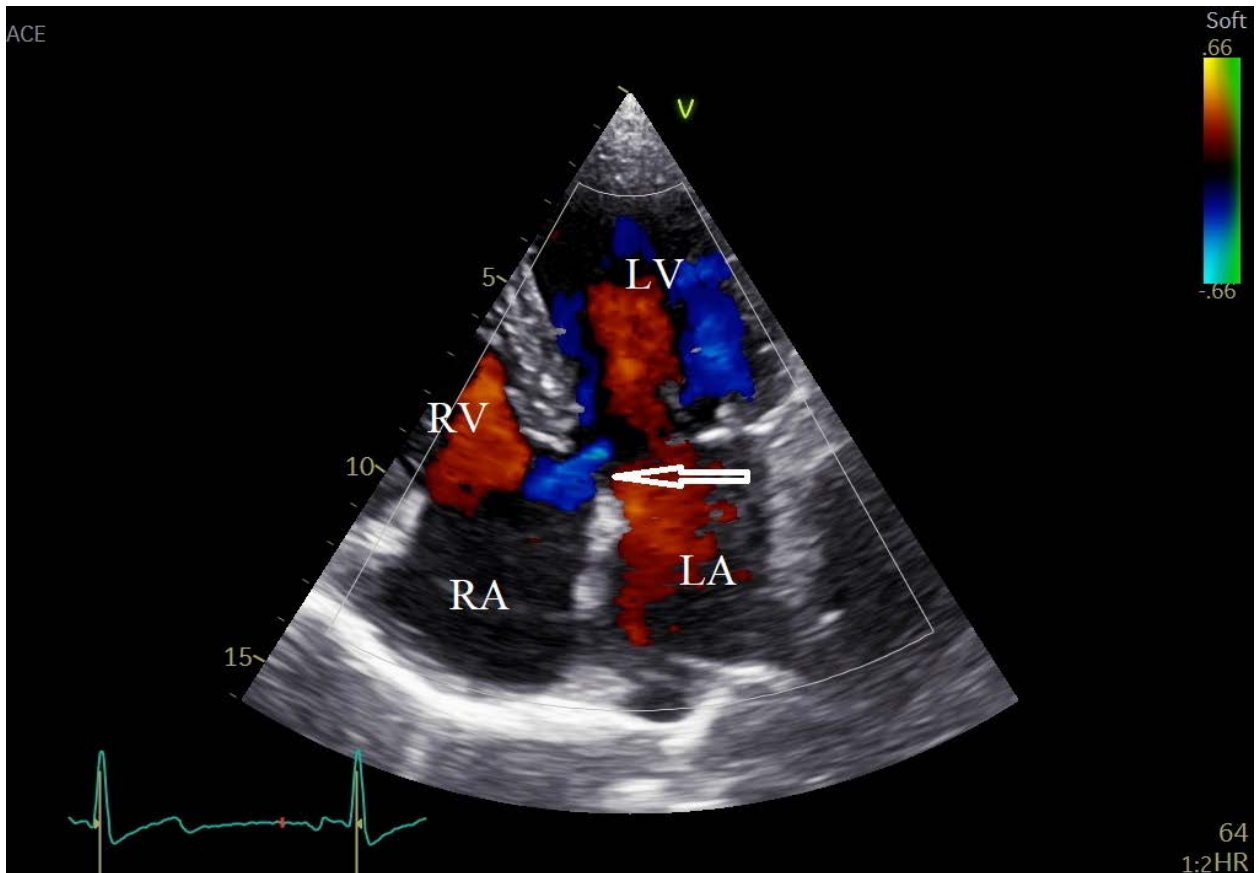
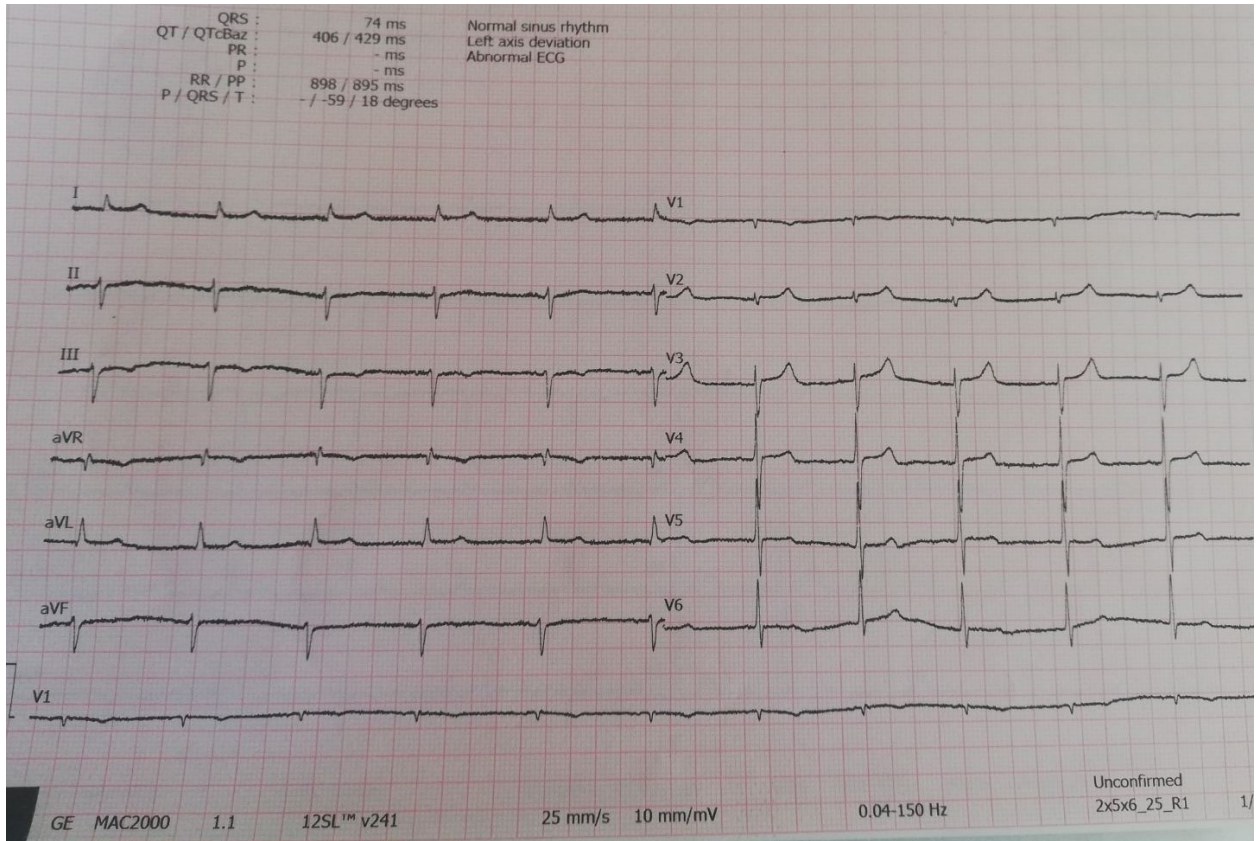
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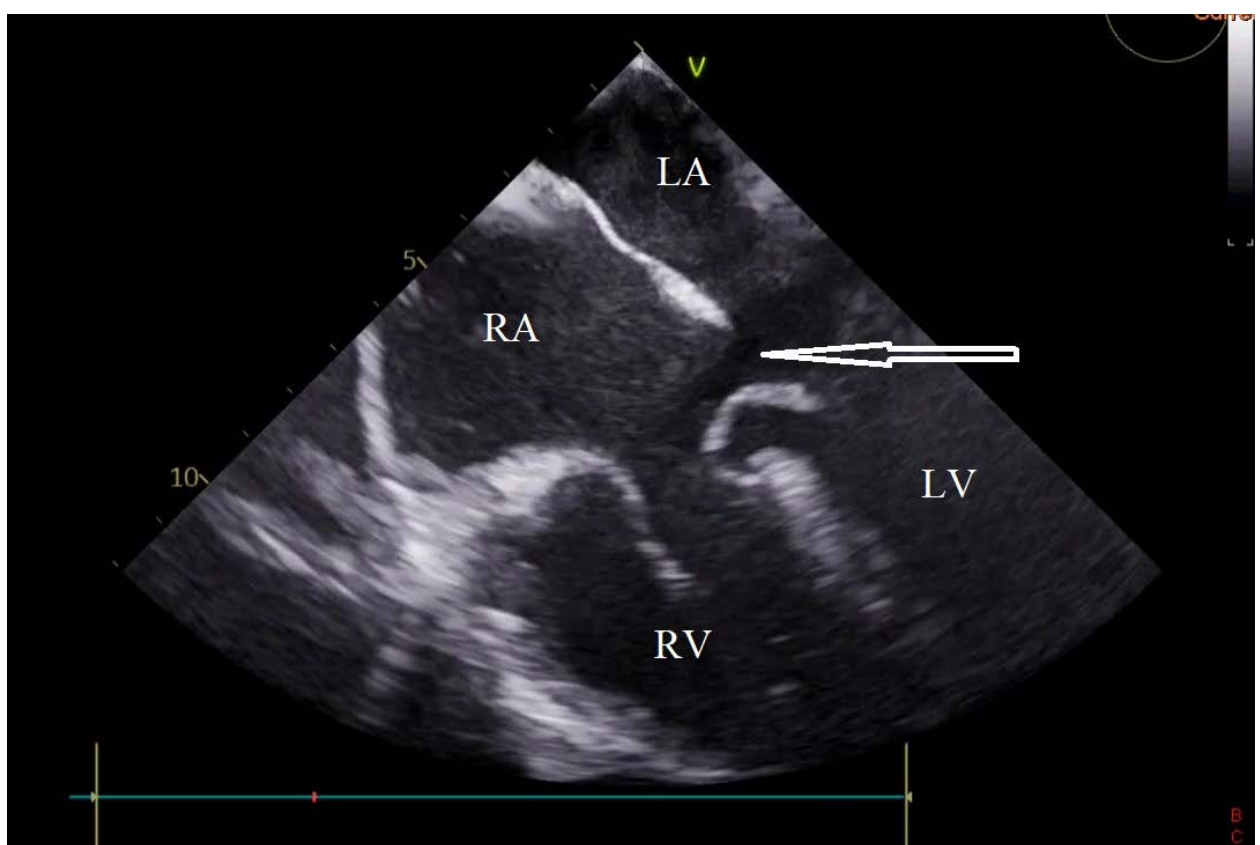
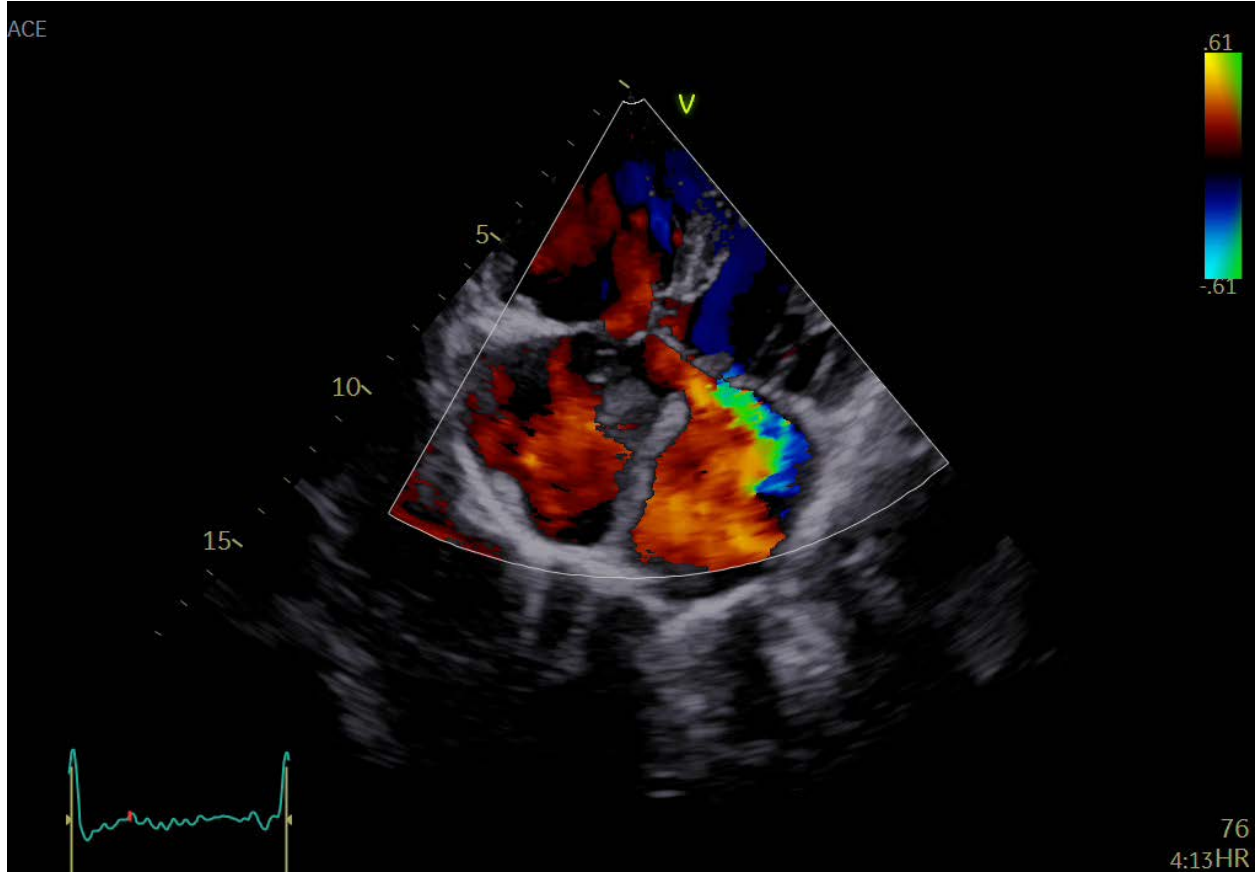
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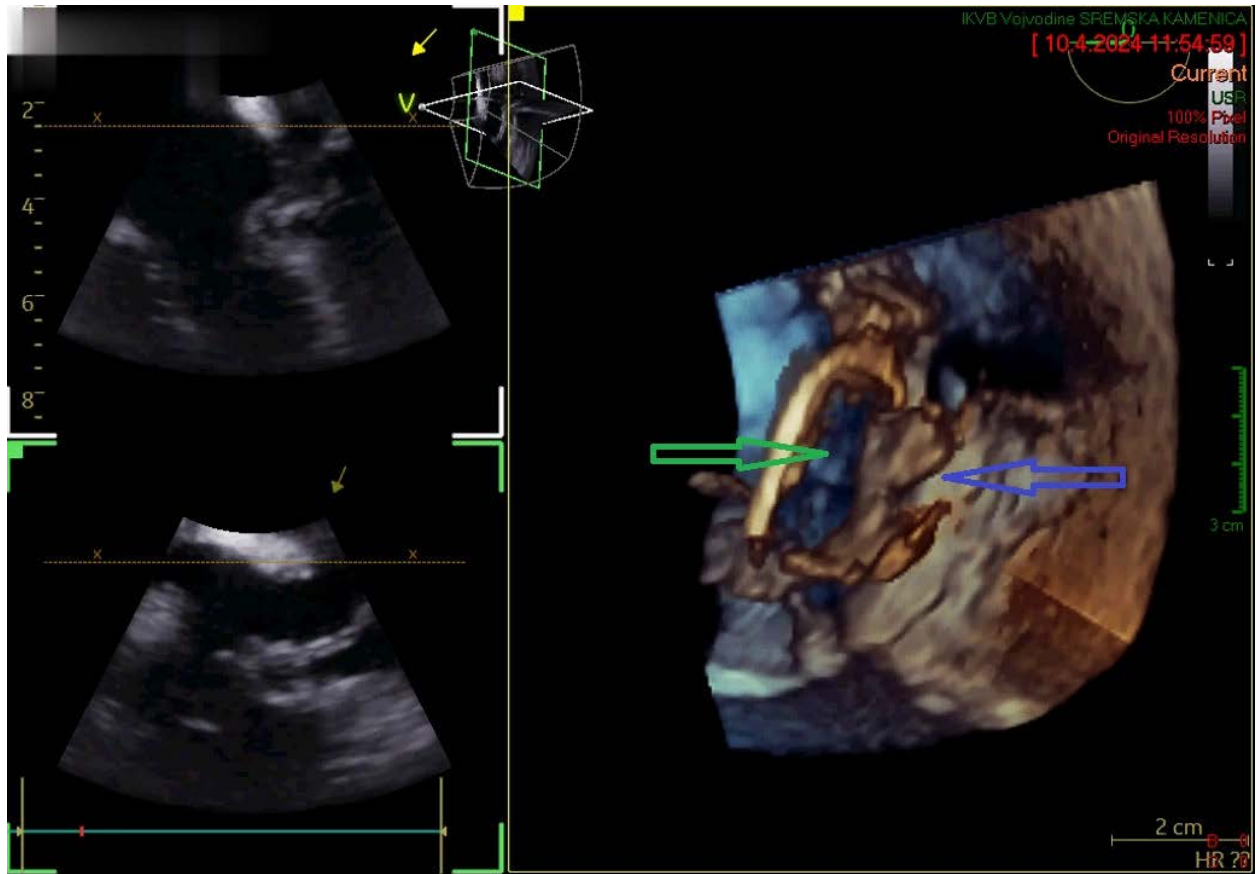
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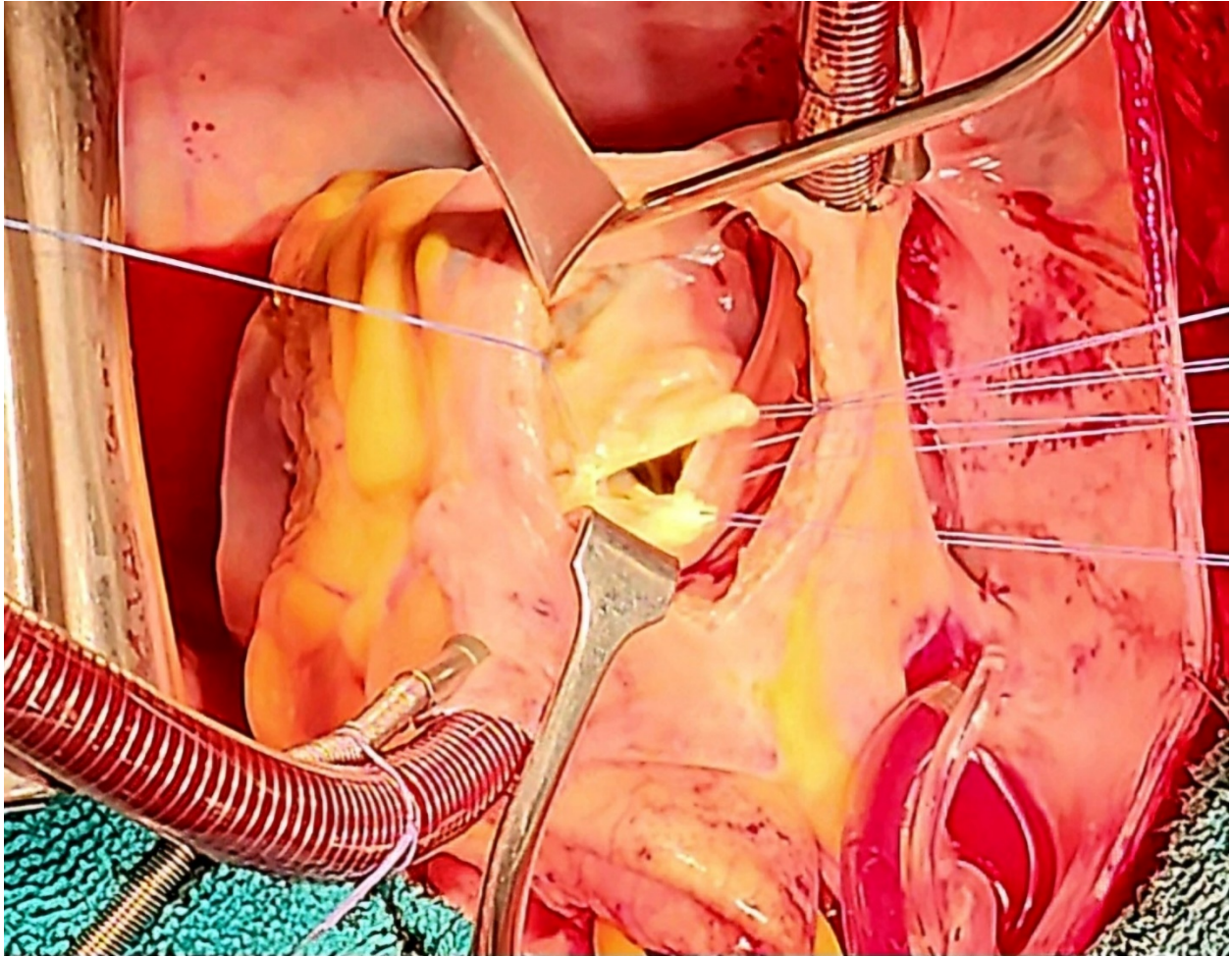


Figure 1. A. The electrocardiogram showed sinus rhythm, frequency about 70/min, levogram, anterior left hemiblock, without signs of acute ischemia and myocardial lesions, without rhythm disturbances. **B.** Two-dimensional transthoracic echocardiography (TTE) (apical 4-chamber view) showing an atrial septal defect of the ostium primum type (arrow). **C.** Two-dimensional TTE showing mitral regurgitation with an eccentric jet. **D.** Transesophageal echocardiography mid-esophageal 4-chamber view of the ostium primum (arrow). **E.** Three-dimensional reconstruction of the ostium primum (green arrow) and cleft of the anterior cusp of the mitral valve (blue arrow). **F.** Surgical view of a mitral valve cleft pulled through an atrial septal defect into the right atrium
Abbreviations: LA – left atrium; LV – left ventricle; RA – right atrium; RV – right ventricle