An interesting case of an absent right-sided AV connection with an atrioventricular septal defect and double-outlet left atrium

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A 41-year-old female presented with exertional presyncope without palpitation and easy fatigability since 7 years. On examination she had cyanosis, clubbing, and room air oxygen saturation of 88%. The electrocardiogram showed 2:1 atrioventricular (AV) block. Transthoracic (Panels A–C and see Supplementary data online, Video S1 and S2) and transoesophageal (Panel D, arrowhead and see Supplementary data online, Video S3) echocardiography revealed situs solitus, levocardia, usual atrial arrangement, absent right AV connection, and double outlet left atrium (DOLA) connected through a common AV valve to both the ventricles. There was no primum atrial septal defect (ASD) and no obvious ventricular septal defect (VSD). The outflow from the RA was through a secundum ASD. There was ventriculoarterial concordance with no outflow tract obstruction. The common AV valve on the short-axis view seems to have an inferior and superior bridging leaflet, a large mural leaflet, and probably small right AV valve anterior and inferior leaflets. The common AV valve was mildly regurgitant. A plain and contrast-enhanced cardiac magnetic resonance study confirmed the findings of echocardiography. DOLA is a ventriculo-atrial malalignment defect. We report an interesting case of an absent right-sided AV connection with an atrioventricular septal defect and DOLA with 2:1 AV block and longest reported survival of 41 years. Moreover, patient underwent a successful dual-chamber epicardial permanent pacemaker implantation for symptomatic 2:1 AV block.

(Panel A) Two-dimensional echocardiography (2D echo) with colour Doppler in the apical four-chamber view showing right atrial outlet atresia and DOLA. (Panel B) Parasternal long-axis view with colour Doppler showing DOLA. (Panel C) Parasternal short-axis view showing a common AV valve. (Panel D) TEE showing an ostium secundum ASD with a right-to-left shunt.

Supplementary data are available at European Heart Journal – Cardiovascular Imaging online.

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