

Double-outlet right atrium in an asymptomatic 8-year-old girl

Sekiz yaşında asemptomatik kız çocuğunda çift çıkışlı sağ atriyum

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Betül Çınar

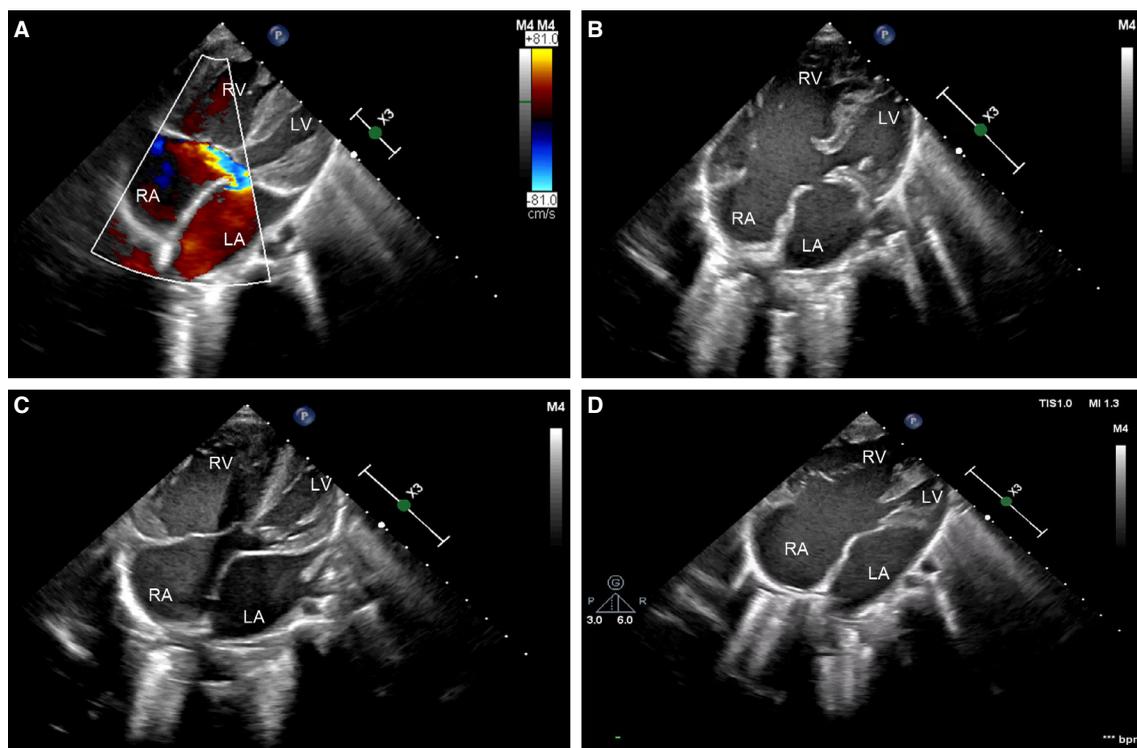
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An asymptomatic 8-year-old girl was admitted to the outpatient clinic for further evaluation of a previously detected heart murmur. A physical examination was unremarkable with the exception of a 2/6 systolic murmur heard at the left second

intercostal space. The oxygen saturation level recorded was 95% and the heart rate was 95 beats per minute. An electrocardiogram revealed a normal sinus rhythm with a superior QRS axis and a left anterior hemiblock pattern. A transthoracic echocardiographic examination demonstrated a common atrioventricular (AV) junction with a leftward rotation of the atrial septum resulting the right atrium draining into both ventricles (double-outlet right atrium [DORA]). Pulmonary venous drainage from the left atrium was transiting to the sys-

temic ventricle through the right atrium via a 10-mm primum-type interatrial defect (Fig. A-D and videos). The cardiac catheterization results were saturation from the superior vena cava: 65%, pulmonary artery (PA): 85%, aorta: 96%; PA pressure: 45/22 mmHg (mean: 33 mmHg); and pulmonary vascular resistance index: 2.4 WU.m². A diagnosis of DORA was confirmed during surgical correction. DORA is a rare congenital anomaly defined by a defect in atrioventricular septal development in which the right atrium opens into both the right and left ventricles. Three anatomical requisites should be present to classify a case as DORA associated with atrioventricular canal defect. First, there is a peculiar leftward deviation of the “true” atrial septum, a distinctive feature of a cor triatriatum. Second, the presence of a cephalic and posterior left atrial chamber receiving all 4 pulmonary veins and with a normally located left atrial appendage can be observed. Finally, specific anatomical features of the AV valves, consisting of either 2 separate patent valves or a common AV valve, are present.



Figures– (A) Transthoracic apical 4-chamber color Doppler images showing (A) passage from the left atrium to the right atrium through the septum primum defect, (B) the right atrium draining into both ventricles, and (C, D) the common atrioventricular valve. LA: Left Atrium, LV: Left ventricle. *Supplementary video files associated with this presentation can be found in the online version of the journal.