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Congenitally corrected transposition of the great vessels: A case of very late presentation at old age and survival till 9th decade

Büyük damarların konjenital düzelmiş transpozisyonu: Doksanlı yıllarda hala hayattaki, yaşlı bir olgunun çok gecikmiş tanısı

Dear Editor,

Congenitally corrected transposition of the great vessels (CCTGV) characterized by atrioventricular (AV) and ventriculoarterial discordance is a rare congenital anomaly which accounts for about 1% of all congenital heart disease cases (1). Only 1% of these patients are without other congenital anomalies (1) Commonly associated anatomic lesions include large atrial or ventricular septal defects, pulmonary stenosis, Ebstein's anomaly and single ventricle (2). The natural history is quite variable and it is dependent on the presence and severity of associated malformations (3). In uncomplicated cases, survival patterns are good but not normal (3). Most patients present in their 50's with the onset of systemic ventricular failure due to left AV regurgitation or suffer from arrhythmias and heart blocks (4). In these patients, the diminutive papillary muscles, the morphology of the tricuspid valve, and the single-vessel perfusion of the morphological right ventricle by the right coronary artery, make right ventricle inferior to left ventricle in the long-term maintenance of systemic circulation (5).

Here, we report a case of a 79- year- old female with no significant past medical history who was admitted for new onset congestive heart failure (CHF). Chest radiogram was significant for moderate pulmonary congestion and abnormal cardiac silhouette with the apex directed rightward and inferiorly. Transthoracic echocardiogram showed the right sided systemic ventricle to be rounded with prominent trabeculae and diminutive papillary muscles. The right sided ventricle had smooth wall and well-formed papillary muscles. The right AV valve was basal to the left sided AV valve. There was severe left sided tricuspid regurgitation. Transesophageal echocardiogram confirmed these findings and demonstrated the aorta and pulmonary trunk to be positioned in a parallel manner (Fig. 1). AV discordance with apical displacement of left sided tricuspid valve consistent with CCTGV was also noted (Fig. 2). Electrocardiogram revealed a normal sinus rhythm, normal P wave axis, normal P-R interval and left ventricular conduction delay. She was treated for new onset CHF, stabilized in the hospital and subsequently discharged home for outpatient follow up. She declined further diagnostic testing and lived till 86 years before she had an unwitnessed sudden death.

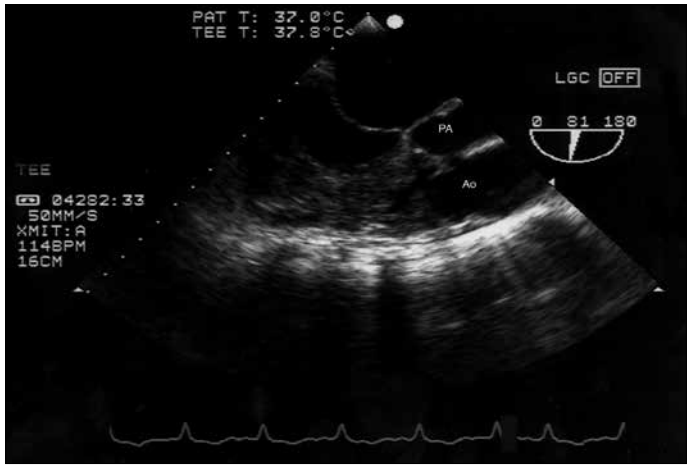

Figure 1. Transesophageal echocardiogram, shows ascending aorta (Ao) and pulmonary artery (PA) in parallel

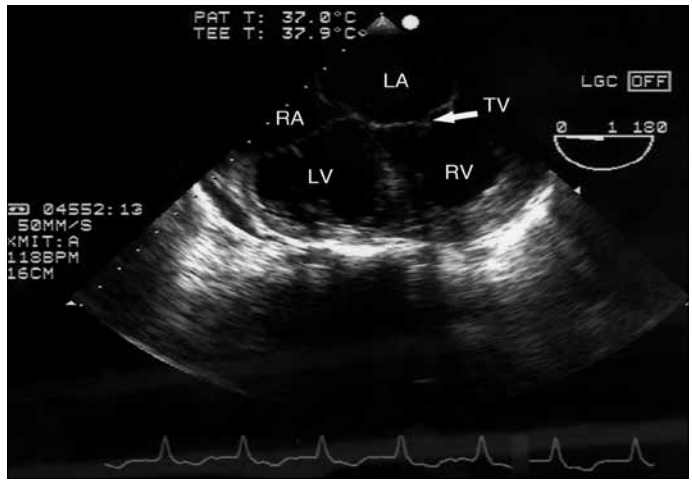

Figure 2. Transesophageal echocardiogram, 4-chamber view, shows atrioventricular discordance with apically displaced left atrioventricular (tricuspid) valve consistent with congenitally corrected transposition of great vessels

LA - left atrium, LV - morphological left ventricle, RA - right atrium, RV - morphological right ventricle, TV - tricuspid valve

The limit of the RV adaptation to the systemic afterload in isolated CCTGV may not be reached until the 9th decade in some patients. CTGV may have a benign clinical course and should be recognized as a rare cause of CHF in very old patients.

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