An unusual form of double outlet right ventricle

Çift çıkışlı sağ ventrikülün nadir bir formu

Mehmet Yokuşoğlu, Ali Cevat Tanalp, Mehmet Uzun, Nail Bulakbaşı, Ersoy Işık*

From Departments of Cardiology and Radiology*, Gülhane Military Medical Academy, Ankara, Turkey

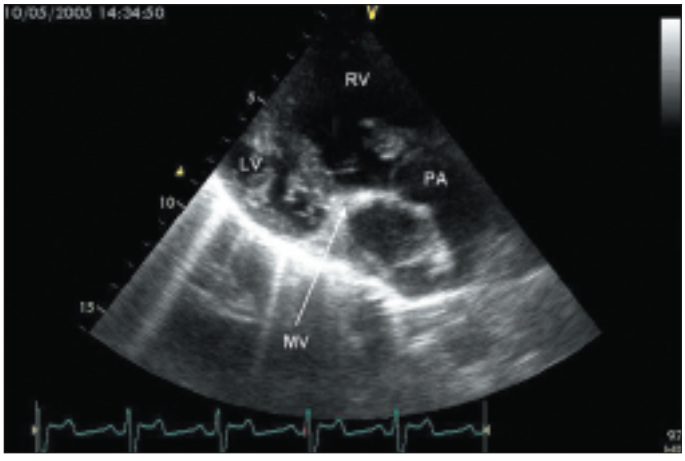

Figure 1. Parasternal long-axis view showing the atretic mitral valve, hypoplastic left ventricle and the posteriorly located pulmonary artery

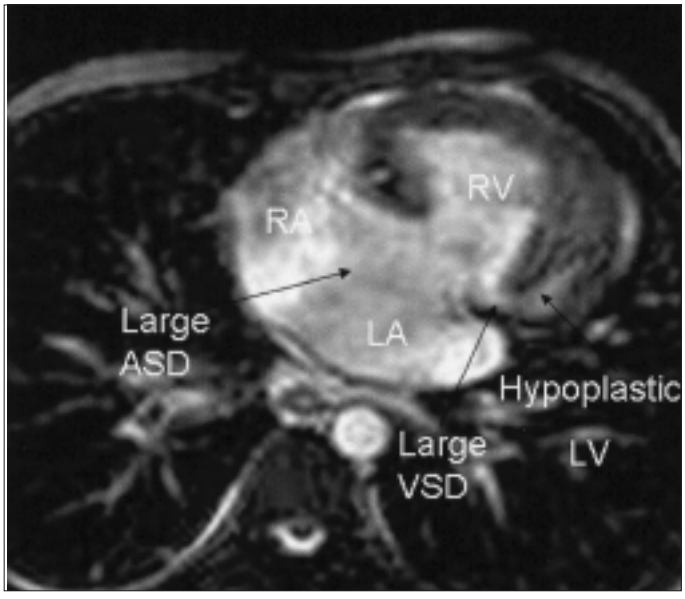

Figure 2. Magnetic resonance imaging demonstrating the atrial septal defect, ventricular septal defect and the hypoplastic left ventricle

A 20-year-old male was admitted to the hospital with cyanosis, exercise intolerance and failure to thrive. Physical examination revealed cyanosis, clubbing and 3/6th grade systolic murmur in the second left and right intercostal spaces. Transthoracic echocardiographic examination demonstrated a hypoplastic left ventricle with atretic mitral valve (Fig. 1). Both great vessels were arising from the right ventricle, which was communicated with hypoplastic left ventricle via a subaortic ventricular septal defect (VSD). The aorta was anterior to the main pulmonary artery. There was also a wide secundum atrial septal defect (ASD). A 16 mmHg gradient was measured by the pulmonary valve and a persistent left superior vena cava

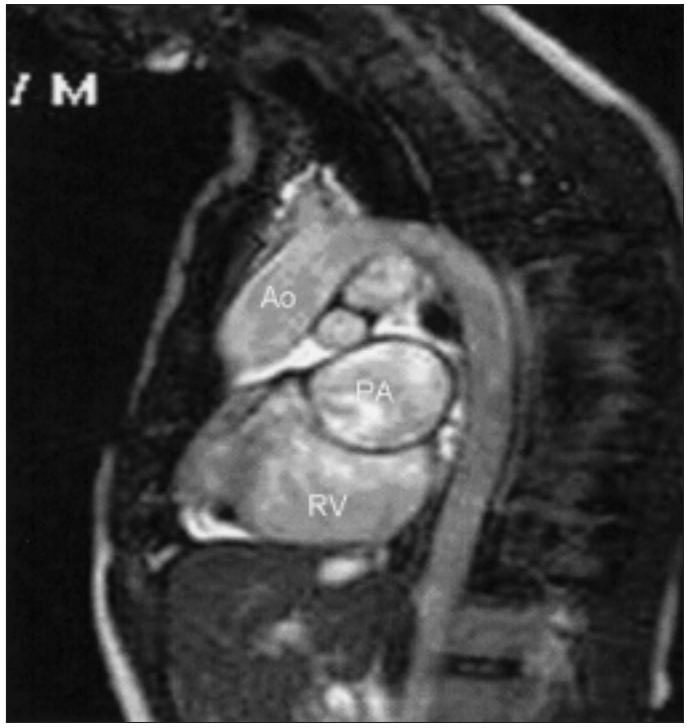

Figure 3. Magnetic resonance imaging demonstrating the localization of aorta anterior to the main pulmonary artery

Ao- aorta, ASD- atrial septal defect, LA- left atrium, LV- left ventricle, MV- mitral valve, PA- pulmonary artery, RA- right atrium, RV- right ventricle, VSD- ventricular septal defect

Address for Correspondence: Mehmet Yokuşoğlu, MD, Gülhane Askeri Tıp Akademisi Kardiyoloji Anabilim Dalı, 06018 Etlik, Ankara, Türkiye
Tel.: +90 312 304 42 67 Faks: +90 312 304 42 50 E-mail: myokusoglu@gata.edu.tr

was identified. Cardiac magnetic resonance imaging confirmed the hypoplastic left ventricle, ASD, VSD and the spatial relationship of the great vessels (Fig. 2-3). The patient underwent cardiac catheterization, which showed Eisenmenger physiology, and the patient was referred to heart-lung transplantation list. Double outlet right ventricle (DORV) represents a continuum of congenital heart diseases that ranges from VSD with significant override of the aorta to a common arterial trunk arising completely from the right ventricle. Patients rarely present with varying degrees of left ventricular hypoplasia and mitral valve anomalies such as stenosis or atresia. To our knowledge, a case of DORV with mitral atresia and left ventricular hypoplasia who survived beyond 20 years without surgery has not been reported.