Double-inlet left ventricle with transposition of great arteries in an asymptomatic adult

A 34-year-old woman was admitted with palpitation. Physical examination revealed IV/VI systolic murmur with thrill over the entire precordium. An increased cardiothoracic ratio was seen on the telecardiogram. ECG revealed a sinus rhythm. Transthoracic echocardiography showed that both atria were connected at the same level to a chamber forming the morphologic left ventricle (MLV), which was separated from the hypoplastic right ventricle via a rudimentary septum with large ventricular septal defect. This MLV was connected to the transposed anteriorly located aorta and to the posteriorly located pulmonary artery. The atrioventricular valves were connected to the MLV (Fig. A, B). There was a subvalvular hypertrophic band leading to a maximum systolic gradient of 68 mmHg in the pulmonary outflow tract.

The diagnosis was made as double-inlet left ventricle with rudimentary right ventricle, transposition of the great arteries, and subvalvular pulmonary stenosis. Subvalvular pulmonary stenosis probably prevented pulmonary hypertension providing a favorable survival. Holter monitoring revealed monomorphic ventricular extrasystoles with two couplets and 26 triplets. Beta-blocker medication was the final decision for her treatment.

Figures. On transthoracic echocardiography, (A) an apical moderated five-chamber view showing pulmonary artery in continuity with the mitral valve and large outlet ventricular septal defect. The pulmonary artery and aorta are transposed. The aorta is in concordance with the hypoplastic right ventricle. (B) An apical four-chamber view showing the left and right atrioventricular valves at the same level and in concordance with the morphologic left ventricle.