Anatomically Corrected Malposition of the Great Arteries {S,D,L}

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B.M. is a previously healthy 18-year-old man referred for a second opinion on an abnormal echocardiogram. He had been followed in the first year of life for a nonspecific murmur. He presented again to medical attention secondary to Navy prequalification requirements and was referred to a local cardiologist. His physical examination, chest radiograph, and ECG were normal. On transthoracic and transesophageal echocardiograms performed in the referring institution, “the aorta did not connect to the left ventricle” and the superior aspect of the ventricular septum was “prominent.” Cardiac MRI revealed levocardia with viscerocardiac situs solitus and D-ventricular loop. The great arteries originated above the appropriate ventricles (ventriculoarterial concordance) (Fig 1), but their spatial position and orientation were abnormal: the aortic valve was anterior, superior, and leftward relative to the pulmonary valve and the great vessels were side-by-side (Figs 1 and 2). There was bilateral conus with aortic-mitral and pulmonary-tricuspid discontinuity and a well-developed subaortic chamber without obstruction. The patient had normal coronary anatomy and normal ventricular function.

Anatomically corrected malposition of the great arteries is a rare form of congenital heart disease in which the great arteries are abnormally related to the ventricles and to each other but nonetheless arise above the anatomically correct ventricles. This abnormal relationship was first reported in 1895 by Theremin and was characterized by Van Praagh et al in 1975. In the absence of associated malformations, anatomically corrected malposition is associated with normal physiology and may be detected incidentally. MRI proved useful in establishing the diagnosis noninvasively in this patient with limited acoustic windows.

Reference


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