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

E.D. Blume, MD; T. Chung, MD; F.A. Hoffer, MD; T. Geva, MD

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 viscerocaudal 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 al1 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


From the Departments of Cardiology (E.D.B., T.G.) and Radiology (T.C., F.A.H.), Children’s Hospital, and the Departments of Pediatrics (E.D.B., T.G.) and Radiology (T.C., F.A.H.), Harvard Medical School, Boston, Mass.

Correspondence to Tal Geva, MD, Department of Cardiology, Children’s Hospital, 300 Longwood Ave, Boston, MA 02115.

E-mail geva_t@al.tch.harvard.edu

The editor of Images in Cardiovascular Medicine is Hugh A. McAllister, Jr, MD, Chief, Department of Pathology, St Luke’s Episcopal Hospital and Baylor College of Medicine.

Circulation encourages readers to submit cardiovascular images to Dr Hugh A. McAllister, Jr, St Luke’s Episcopal Hospital and Texas Heart Institute, 6720 Bertner Ave, MC1-267, Houston, TX 77030.

(Circulation. 1998;97:1207.)

© 1998 American Heart Association, Inc.
Anatomically Corrected Malposition of the Great Arteries {S,D,L}
E.D. Blume, T. Chung, F.A. Hoffer and T. Geva

Circulation. 1998;97:1207
doi: 10.1161/01.CIR.97.12.1207

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1998 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/97/12/1207

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/