An 82-year-old woman presented with recurrent syncope. Clinically, she had a slow rising pulse, right parasternal heave, and an ejection systolic murmur loudest at the right sternal edge with a quiet pansystolic murmur at the apex. ECG demonstrated prolonged PR interval and extreme leftward axis. Transthoracic and transesophageal echocardiography revealed atrioventricular and ventriculoarterial discordance, suggesting congenitally corrected transposition of the great arteries (ccTGA). The systemic right ventricle (RV) was hypertrophied and mildly dilated with normal systolic function, but there was severe subvalvular stenosis with a peak velocity of 4.5 m/s (Figures 1 and 2). The aortic valve was trileaflet and anterior to the pulmonary valve, and there was mild left atrioventricular valve regurgitation.

Cardiac magnetic resonance imaging confirmed a systemic RV with marked hypertrophy (RV mass, 112 g; mass index, 72 g/m2) but normal volumes and function (RV end-diastolic volume, 95 mL; RV end-systolic volume, 27 mL; ejection fraction, 0.72). There was minimal systemic atrioventricular valve regurgitation. There was a muscle band across the systemic RV outflow tract, causing dynamic obstruction (Figures 3 and 4). The subpulmonary left ventricle was thin with normal volumes and function without obstruction of the outflow tract or main or branch pulmonary arteries. Coronary angiography confirmed a dominant left coronary artery with a posterior origin and a small right coronary artery. Right heart catheterization confirmed normal pulmonary artery pressures.

The patient underwent surgery to relieve the RV outflow tract obstruction. At operation, there was a tunnel-like outflow tract obstruction. The myocardium was resected to allow passage of a Heger dilator into the morphological RV, but the aortic valve was not modified. Histology of the resected myocardium confirmed myocyte hypertrophy with fibrosis and marked endocardial thickening. Postoperative recovery was rapid and uneventful, with discharge from hospital on day 6.

ccTGA is uncommon, accounting for 1% of patients born with congenital heart disease, and only 1% of these patients have uncomplicated ccTGA (without associated abnormalities such as pulmonary stenosis, ventricular septal defect, or an Ebstein-like tricuspid valve).1 ccTGA may frequently present in adulthood. In 1 study, 66% of patients with ccTGA presented in adulthood, with 17% of those patients being >60 years of age.2 Morbidity and mortality in this group are due predominantly to systemic atrioventricular valve regurgitation and systemic RV failure. In 1 multicenter study, 25% of patients with uncomplicated ccTGA had developed significant RV dysfunction by 45 years of age.3 However, very late presentation of ccTGA, in the eighth decade of life, is unique and is notable for the presence of severe systemic ventricle outflow tract obstruction but with preserved systemic ventricular function without major atrioventricular valve regurgitation.

Sources of Funding

Dr Myerson received support from the Oxford NIHR Biomedical Research Centre programme.

Disclosures

None.

References


From the Department of Cardiology, John Radcliffe Hospital, Oxford, UK.
Correspondence to Elizabeth A. Orchard, MA, MBBS, MRCP, Department of Cardiology, John Radcliffe Hospital, Headley Way, Oxford OX3 9DU, UK. E-mail Elizabethorchard@hotmail.com

(Circulation. 2010;122:e441-e444.)

© 2010 American Heart Association, Inc.

Circulation is available at http://circ.ahajournals.org

DOI: 10.1161/CIRCULATIONAHA.109.919068 e441
Figure 1. Transthoracic echocardiographic images. Shown are representative images from transthoracic echocardiography at mid diastole (A) and mid systole (B). The systemic RV is severely hypertrophied, is filled from the left atrium (LA), and ejects into the aorta (Ao). The RV outflow tract (asterisk) is severely narrowed by a muscle bridge (arrowhead), most markedly during systole.
Figure 2. Transesophageal echocardiographic images. Transesophageal images at early systole (A), late systole (B), and diastole (C). The RV outflow tract (asterisk) is narrowed most severely during mid and late systole. LA indicates left atrium; Ao, aorta.
Figure 3. Magnetic resonance images. A, A 4-chamber view demonstrating the subsystemic RV (*) and subpulmonary left ventricle with the mitral valve displaced more basally (arrowed). B, Basal short-axis view showing the bileaflet mitral valve (MV) in a more anterior position and the trileaflet tricuspid valve (TV) in the more posterior position.

Figure 4. Magnetic resonance images. View of the RV outflow tract into the aorta (Ao). A, Diastole, showing position of the aortic valve (short arrow). B, Systole, showing turbulence in the narrowed outflow tract (long arrow).
Congenitally Corrected Transposition of the Great Arteries Presenting in a Nonagenarian
Elizabeth A. Orchard, Oliver Ormerod, Saul Myerson and Stephen Westaby

Circulation. 2010;122:e441-e444
doi: 10.1161/CIRCULATIONAHA.109.919068

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2010 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/122/9/e441

An erratum has been published regarding this article. Please see the attached page for:
http://circ.ahajournals.org/content/suppl/2011/03/22/122.9.e441.DC1

Data Supplement (unedited) at:
http://circ.ahajournals.org/content/suppl/2011/03/22/122.9.e441.DC1

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/
In the article by Orchard et al, “Congenitally Corrected Transposition of the Great Arteries Presenting in a Nonagenarian,” which was published in the August 31, 2010 issue of the journal *(Circulation. 2010;122:e441–e444)*, the authors neglected to name a source of funding. Dr. Myerson received support from the Oxford NIHR Biomedical Research Centre programme.

The text has been corrected in the online version of the article. The authors regret the error.

DOI: 10.1161/CIR.0b013e31823238e7