DOI: 10.1111/j.1540-8175.2009.00934.x

ECHO ROUNDS Section Editor: Edmund Kenneth Kerut, M.D.

A Heart Reversed Triply: Situs Inversus Totalis with Congenitally Corrected Transposition of the Great Arteries in a Middle-Aged Woman

Hung-Yu Chang, M.D.,* Wei-Hsian Yin, M.D., Ph.D.,* Ming-Chon Hsiung, M.D.,* and Mason-Shing Young, M.D., F.A.C.C.†

*Division of Cardiology, Cheng-Hsin Rehabilitation Medical Center, Taipei, Taiwan; and †Department of Internal Medicine, Cheng-Hsin Rehabilitation Medical Center, Taipei, Taiwan

(ECHOCARDIOGRAPHY, Volume 26, May 2009)

Address for correspondence and reprint requests: Wei-Hsian Yin, M.D., Ph. D., Division of Cardiology, Cheng-Hsin Rehabilitation Medical Center, No. 45, Cheng-Hsin Street, Pei-Tou, Taipei 112, Taiwan. Fax: +886-2-2826-1242; E-mail: amadeus0814@yahoo.com.tw

A 57-year-old woman was well in the past. She suffered mild dyspnea for 2 years. On physical examination, she was found to be normotensive without signs of heart failure. A grade 3/6 pansystolic murmur was heard over the right precordium. Chest radiography

Figure 1. *Chest radiography demonstrated a normal-sized heart with dextrocardia, right-sided gastric air bubble, aortic knob and descending aorta. The left hemidiaphragm was higher than the right one. This picture was compatible with situs inversus totalis.*

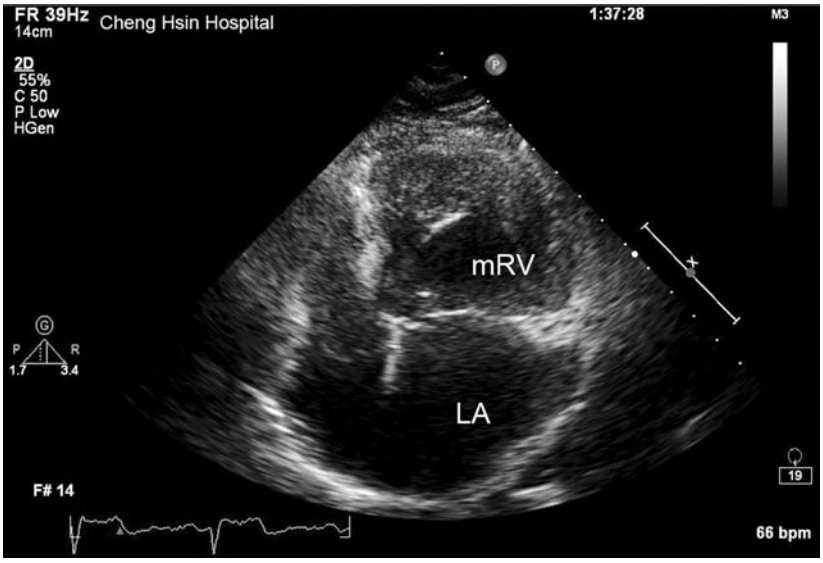

Figure 2. *Transthoracic echocardiographic four-chamber view showed a moderator band and thick trabeculations in the morphological right ventricle on the right side of the heart. mRV = morphologic right ventricle; LA = left atrium.*

demonstrated a normal-sized heart with dextrocardia, right-sided gastric air bubble, aortic knob, and descending aorta (Fig. 1). A transthoracic echocardiographic four-chamber view showed a moderator band and thick trabeculations in the morphological right ventricle on the right side of the heart (Fig. 2 and Video clip 1). Color flow imaging revealed a severe central regurgitation jet into the morphologic left atrium, which received pulmonary venous drainage (Fig. 3 and Video clip 2). Although the tricuspid valve may be identified by its relatively more apical position compared with the mitral valve anterior leaflet, the atrioventricular valve between morphologic right ventricle and left atrium was examined by real time three-dimensional echocardiography, and a trileaflet tricuspid valve was clearly demonstrated (Fig. 4 and Video clip 3). A modified right parasternal long-axis view showed the anteriorly placed aorta arising from morphologic right ventricle and the posteriorly placed pulmonary artery arising from morphologic left ventricle. Two great arteries were parallel rather than having the normal spiraling pattern (Fig. 5 and Video clip 4). There was no ventricular septal defect or pulmonic stenosis. The right-sided heart with both atrioventricular and ventriculoarterial discordance indicated situs inversus with

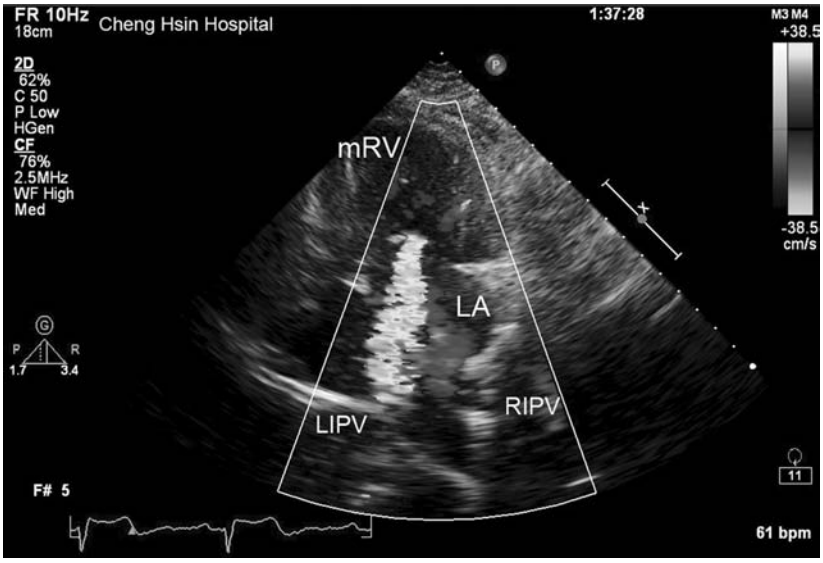

Figure 3. *Color flow imaging revealed a severe central regurgitation jet into the morphologic left atrium, which received pulmonary venous drainage. mRV = morphologic right ventricle; LA = left atrium; RIPV = right inferior pulmonary vein; LIPV = left inferior pulmonary vein.*

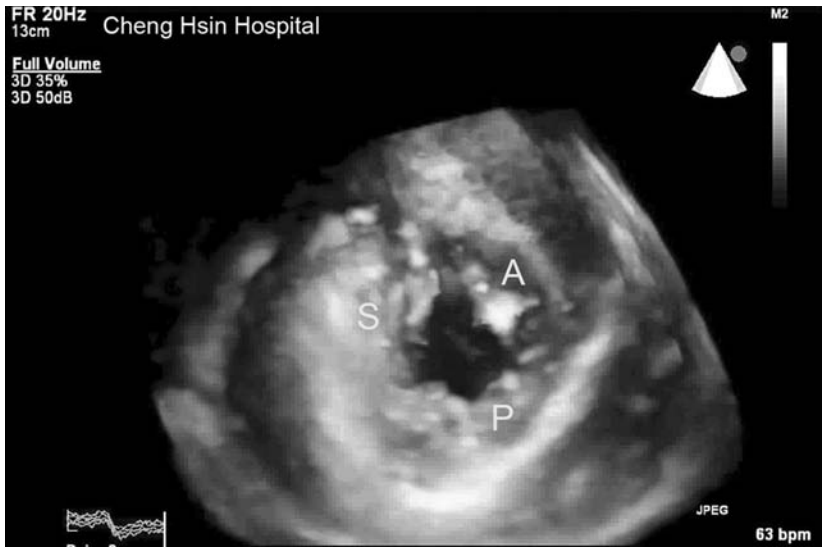

Figure 4. *Real time three-dimensional echocardiography obtained by parasternal long-axis imaging showed three leaflets of tricuspid valve. A = anterior leaflet; P = posterior leaflet; S = septal leaflet.*

congenitally corrected transposition of the great arteries (ccTGA). Contrast-enhanced computed tomography confirmed the diagnosis. The morphologic left atrium received pulmonary venous drainage and emptied into morphologic right ventricle (Fig. 6). The aorta arose from the right ventricular outflow tract. The aortic valve was located anteriorly and to the right of the pulmonic valve. The left-hand sinus gave rise to the left main coronary artery, and the right-hand sinus gave rise to the right coronary artery (Fig. 7). Compared to the normal anatomy in situs inversus totalis, the cusps rotated 120° clockwise. Reconstructed cardiac angiogram demonstrated the coronary artery anatomy (Fig. 8). The dominant right coronary artery ran toward the right-sided cardiac apex. There was no significant coronary artery stenosis.

This is a rare but typical case of situs inversus totalis with congenitally corrected TGA. In congenitally corrected TGA patients, the most common associated intracardiac defects are ventricular septal defect (90%) and pulmonary stenosis (80%).[1] A complete heart block is acquired at a rate of 2% per year and is thought to result from an abnormally long His bundle.[2] Even without any other cardiac anomalies,

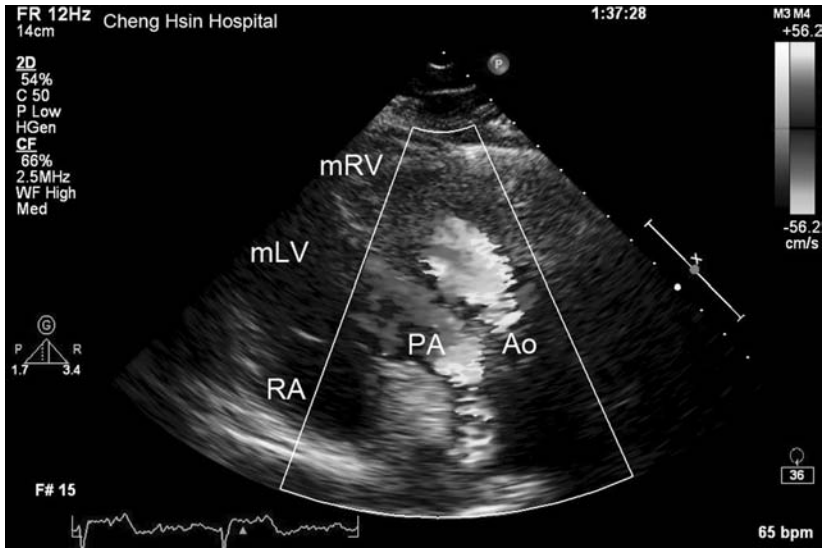

Figure 5. *Transthoracic echocardiography demonstrated the anteriorly placed aorta arising from morphologic right ventricle and the posteriorly placed pulmonary artery arising from morphologic left ventricle. Two great arteries were parallel rather than having the normal spiraling pattern. mRV and mLV = morphologic right and left ventricle; Ao = aorta; PA = pulmonary artery; RA = right atrium.*

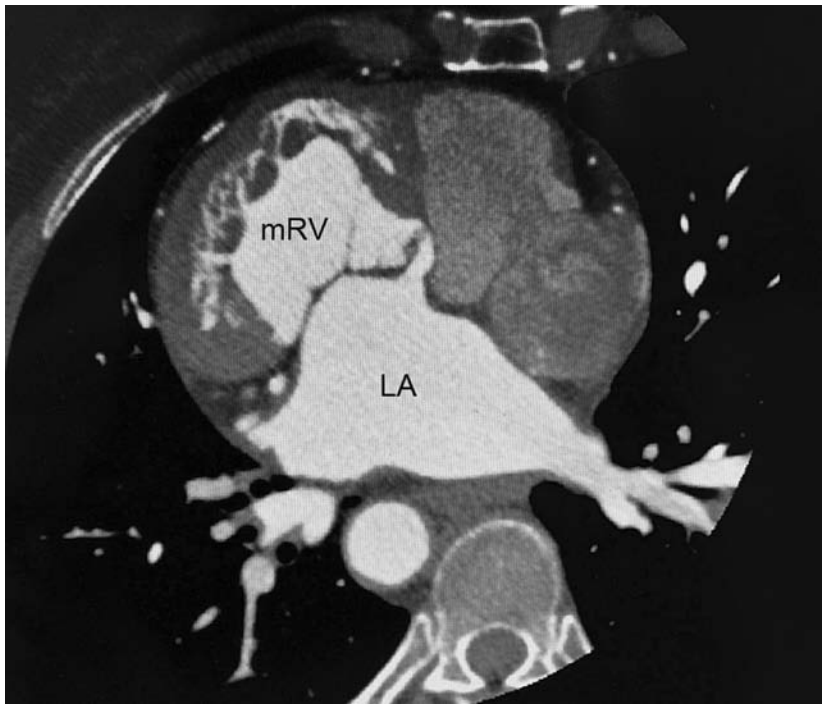

Figure 6. *Contrast-enhanced computed tomography. The morphologic left atrium received pulmonary venous drainage and emptied into morphologic right ventricle. mRV = morphologic right ventricle; LA = left atrium.*

chronic pressure overload of the systemic right ventricle and atrioventricular valve regurgitation may still lead to contractile dysfunction and heart failure.[3] Simultaneous visualization of all leaflets of the atrioventricular valve is challenging from standard two-dimensional echo imaging. In our case, we examined the right atrioventricular valve by real time three-dimensional echocardiography and clearly demonstrated the three leaflets of

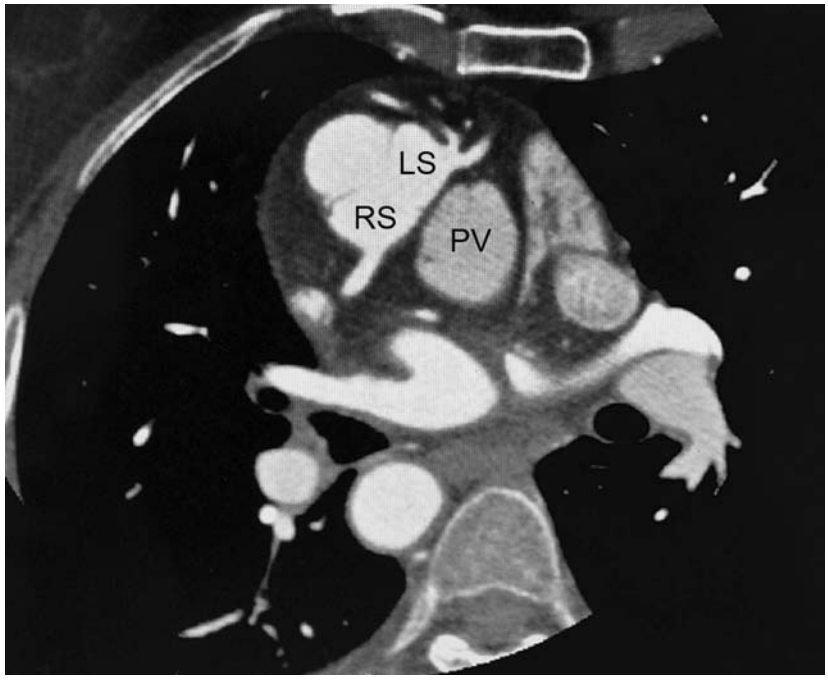

Figure 7. *Contrast-enhanced computed tomography. The aortic valve located anteriorly and to the right of the pulmonic valve. The left-hand sinus gave rise to the left main coronary artery, and the right-hand sinus gave rise to the right coronary artery. PV: pulmonic valve; LS: left-hand sinus; RS: right-hand sinus.*

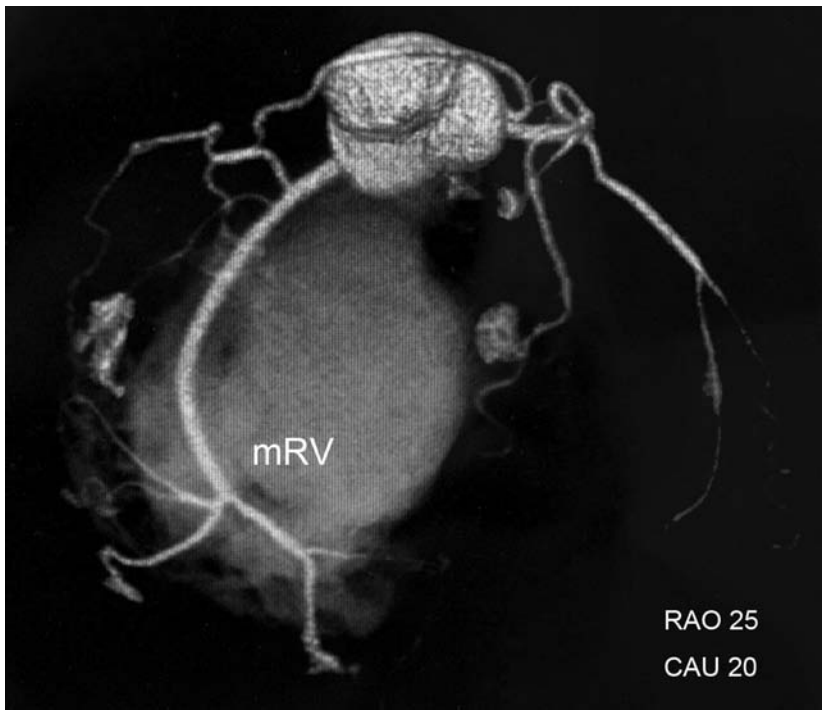

Figure 8. *Reconstructed computed tomography cardiac angiogram demonstrated the coronary artery anatomy. The dominant right coronary artery ran toward the right-sided cardiac apex. There was no significant coronary artery stenosis. mRV: morphologic right ventricle; RAO: right anterior oblique projection; CAU: caudal projection.*

tricuspid valve. This case demonstrates the usefulness of echocardiography and contrast-enhanced computed tomography in the assessment of this rare congenital heart disease.

References

1. Connelly MS, Liu PP, Williams WG, et al: Congenitally corrected transposition of the great arteries in the adult: Functional status and complications. *J Am Coll Cardiol* 1996;27:1238–1243.
2. Huhta J, Maloney J, Ritter D: Complete atrioventricular block in patients with atrioventricular discordance. *Circulation* 1983;67:1374–1377.
3. Hornung TS, Bernard EJ, Celermajer DS, et al: Right ventricular dysfunction in congenitally corrected transposition of the great arteries. *Am J Cardiol* 1999;84:1116–1119, A10.

Supporting Information

Additional Supporting Information may be found in the online version of this article:

Video clip 1. Video clip for figure 2.
Video clip 2. Video clip for figure 3.
Video clip 3. Video clip for figure 4.
Video clip 4. Video clip for figure 5.

Please note: Wiley-Blackwell is not responsible for the content or functionality of any supporting information supplied by the authors. Any queries (other than missing material) should be directed to the corresponding author for the article.