Thallium Myocardial Scintigraphy in Congenitally-Corrected Transposition of the Great Arteries

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A case of congenitally-corrected transposition of the great arteries is presented with the correlation of thallium scintigraphic results with catheterization data. The essential features of the thallium scintigrams were marked counterclockwise rotation of the heart with perfusion abnormalities of the inferior wall and apex. Since patients with congenitally-corrected transposition of the great arteries may present with the symptom of chest pain, the diagnosis of transposition of the great arteries should be considered in patients with marked counterclockwise rotation of the heart and segmental perfusion abnormalities on thallium scintigraphy.

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Congenitally-corrected transposition of the great arteries (TGA) is a rare congenital heart condition. In this condition, there is discordance between both the atria and ventricles and the ventricles and great vessels: the anatomic left ventricle lies on the right side (pulmonic ventricle) and is connected to the pulmonic trunk, whereas the anatomic right ventricle lies on the left side and functions as the systemic ventricle. The atrioventricular valve between the right atrium and the pulmonic ventricle is morphologically the mitral valve, whereas the systemic atrioventricular valve is morphologically the tricuspid valve.

The presentation of congenitally-corrected TGA (without other associated congenital cardiac lesions) is often in adulthood with symptoms of chest pain or dyspnea. Despite the presentation with chest pain, reports of thallium scintigraphy in these patients are uncommon. We recently encountered a patient with congenitally-corrected TGA with an abnormal thallium scintigraphic study. The case illustrates abnormalities on thallium scintigraphy which differ from those previously attributed to congenitally-corrected TGA.

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CASE REPORT

The patient was a 34-yr-old female who presented to her personal physician with a 4-wk history of exertional chest pain. During an exercise tolerance test she developed chest pain and shortness of breath. She was referred to the University of Wisconsin Hospital and Clinics, Madison, for a thallium stress test. On physical exam, her weight was 88.6 kg, height 168 cm, blood pressure 124/80, pulse 90 bpm and regular. A faint Grade I/VI holosystolic murmur was heard at the apex. Examination was otherwise unremarkable. EKG showed sinus bradycardia, right shift in the QRS precordial forces and, left ventricular hypertrophy by voltage critera and repolarization abnormalities.

A thallium stress test was performed. The patient exercised to 6:25 min on the Bruce protocol achieving 91% of her maximum age-predicted heart rate. She developed chest pain at 4 min into exercise at a heart rate of 144/min; inferolateral ST depression of 2 mm was noted at peak exercise. One minute prior to the termination of exercise, the patient was injected with 2.4 Ci of ^{201}Tl-chloride, and gamma camera imaging was performed 4 min following exercise in three planar views of 45° left anterior oblique (LAO45), 80° left lateral in the right decubitus, and anterior using a small field of view gamma camera (300 Omega Starcam, General Electric, Inc., Milwaukee, WI). Images were obtained for 8 min using a general all-purpose low-energy collimator. Stress thallium imaging revealed abnormal cardiac anatomy. There was marked counter-clockwise rotation of the heart with the best views of the septum and the inferior wall in the anterior and LAO45 views, respectively. The inferior wall including the posterobasal segment appeared thinned. An apical perfusion defect was present in the LAO45 and left lateral views and was noted to involve the inferoapex in the anterior view (Fig. 1A). The uptake of the anterior wall, septum and the posterolateral wall appeared normal. The uptake of thallium by the pulmonic ventricle on the LAO45 and anterior views was similar to a normal right ventricle after exercise stress. Rest thallium imaging performed 4 hr and 24 hr later showed no change in the perfusion pattern (Fig. 1B, 24-hr delayed images not shown).

With an apparent abnormality in cardiac anatomy, an echocardiogram was then performed and a diagnosis of congenitally-corrected TGA was made. The alterations in cardiac anatomy made the study technically difficult; however, the systemic ventricle was slightly enlarged and mildly hypokinetic. There was mild regurgitation across the systemic atrioventricular valve.

The patient subsequently underwent cardiac catheterization including biplane ventriculography and coronary angiography. Hemodynamic parameters of both the pulmonic and systemic

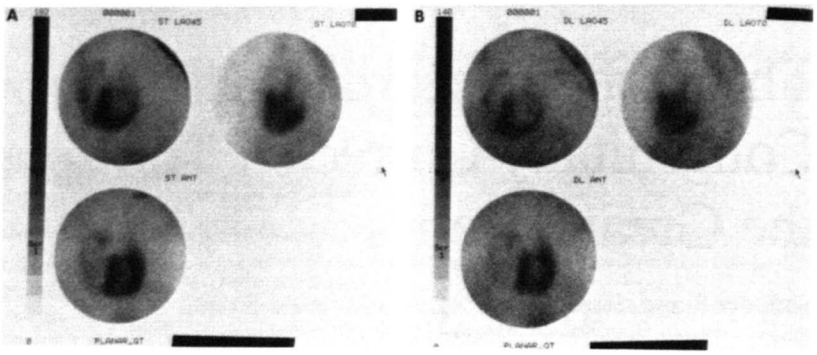

FIGURE 1. Planar thallium images are displayed in the 45° left anterior oblique (LAO45), left lateral (LAO70), and anterior (ANT) views. (A) Stress (ST) thallium images and (B) 4-hr delayed (DL) thallium images. The same grey scale for the delayed images was used as for the stress images with a correspondingly lower maximal pixel count.

ventricles were within normal limits (pulmonic ventricle 28/8 mmHg, systemic ventricle 104/5 mmHg, cardiac index 4.5 liters/min/m^2) except for elevation in the pulmonic ventricular end-diastolic pressure to 17 mmHg. The typical anatomic features of congenitally corrected TGA with L-transposition were identified. The pulmonic ventricle (morphologically the left ventricle with smooth walls) was placed to the right and connected to the pulmonary trunk which was located posteriorly, low and to the right (Fig. 2A). The systemic ventricle (morphologically the right ventricle with trabeculated walls) was on the left side and connected to the aorta which was placed anteriorly, high and to the left (Fig. 2B). The systemic ventricle appeared to have mild, diffuse hypokinesis. A probe-patent foramen ovale was also present. The coronary arteries were also transposed with the morphologic right coronary artery supplying the left-sided, systemic ventricle and the morphologic left coronary artery supplying the pulmonic ventricle (Fig. 3).

DISCUSSION

About half of the patients with congenitally-corrected TGA, especially those without associated defects, do not present until adult life. These patients may present between the third and sixth decades of life with symptoms of congestive heart failure or varying degrees of AV block (*1,2*). In rare instances, these patients may also present with symptoms of angina (*3,4*).

The current case is an example of late presentation with angina. Cardiac catheterization confirmed the findings of congenitally corrected TGA with L-transposition. An abnormality in cardiac anatomy was first suspected on thallium scintigraphy. The principal abnormalities on thallium scintigraphy were: (1) marked counter-clockwise rotation of the heart; (2) apical and inferoapical perfusion defect without redistribution; (3) thinning of the inferior wall; and (4) normal pulmonic ventricular uptake of thallium. There are few reports of thallium scintigrams in patients with congenitally-corrected TGA. Abdel-Dayem et al. (*4*) have reported a resting thallium study on a patient with congenitally-corrected TGA who presented with dyspnea on exertion and incomplete right bundle branch block. In addition to the marked counter-clockwise rotation of the heart, the thallium study in their case was significant for: (1) equivalent uptake of thallium by both ventricles, and (2) decreased septal uptake of thallium (*4*). Nakajima et al. also reported a case of congenitally-corrected TGA in which a tomographic thallium study demonstrated reverse curvature of the septum and inhomogeneous uptake of thallium by the myocardium (*5*).

The current report confirms the presence of marked counter-clockwise rotation of the heart on thallium scintigraphy seen on the previous planar study of Abdel-Dayem et al., but differs in the amount of pulmonic ventricular uptake and location of perfusion abnormalities. The septum was best visualized on both studies in the anterior position and both ventricles were visualized. However, instead of septal perfusion defects, as reported in the previous study, our patient had fixed perfusion defects in the apical and inferoapical segments and thinning of the inferior wall and posterobasal segment. These portions of the systemic ventricle were supplied by the morphological right coronary artery. The perfusion abnormalities are

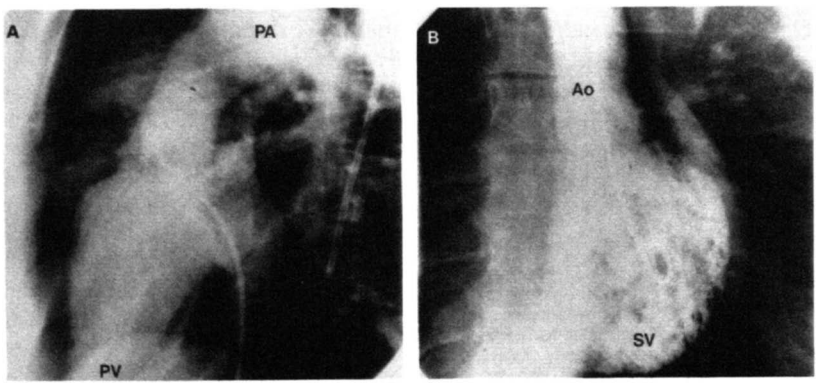

FIGURE 2. Ventriculograms of both ventricles. (A) pulmonic ventricle (PV) in the left anterior oblique projection (PA, pulmonary artery) and (B) systemic ventricle (SV) in the right anterior oblique projections (Ao, aorta).

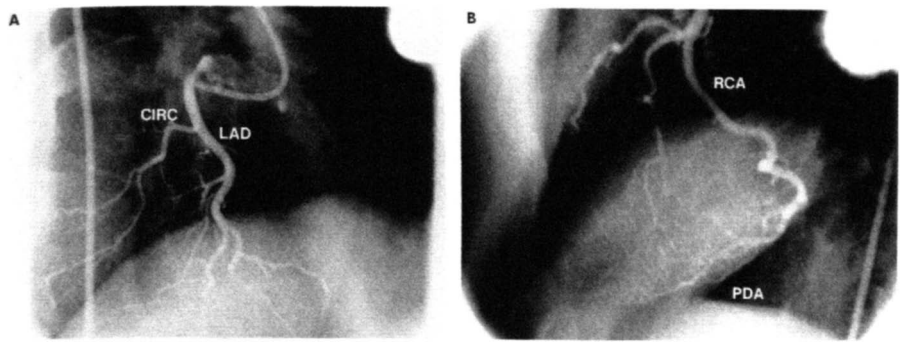

FIGURE 3. Arteriograms of the left coronary artery supplying the pulmonic ventricle in the right anterior oblique projection (panel A; LAD, left anterior descending artery; CIRC, circumflex artery) and the right coronary artery supplying the systemic ventricle in the left anterior oblique projection (panel B; RCA, right coronary artery; PDA, posterior descending artery).

probably due to inadequate perfusion by the right coronary artery of the systemic ventricle. The difference in perfusion abnormalities between the case of Abdel-Dayem et al. and the current study is probably due to difference in coronary anatomy. Hypertrophy of the morphologic right ventricle occurs to accommodate the increased ventricular afterload of the systemic circulation. With patients developing ventricular dysfunction or angina with congenitally-corrected TGA, it is apparent that coronary perfusion of the right ventricle is insufficient for the degree of hypertrophy. It is unknown whether the inadequacy of myocardial perfusion is secondary to an epicardial defect with the morphologic right coronary supplying the systemic ventricle or a microvascular phenomenon similar to acquired left ventricular hypertrophy as in systemic hypertension, for instance. The alterations in myocardial perfusion in congenitally-corrected TGA was also suggested by the case of Nakajima et al. (*5*).

The second major difference between the study of Abdel-Dayem et al. and this study is the amount of pulmonic ventricular uptake of thallium. In the former study, uptake was equal in both ventricles as opposed to greater uptake by the systemic ventricle in this study. This differential in ventricular activity was at least in part related to the performance of exercise stress in the current study.

In summary, patients with corrected TGA often escape detection until adult life when they may present with various symptoms including angina. Thallium scintigraphy in such patients may reveal abnormal cardiac anatomy with marked counter-clockwise rotation of the heart. The septum in these patients is best visualized in the anterior view, with both ventricles usually seen as well. Perfusion abnormalities may occur in either the septum or inferior wall and apex. Such an appearance on thallium scintigraphy should, in the appropriate situation, suggest a diagnosis of congenitally-corrected TGA and lead to further evaluation to confirm the diagnosis.

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