Long-Term Survival in Aged Patients with Corrected Transposition of the Great Arteries*

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Corrected transposition of the great arteries is a rare condition, and few patients with this abnormality survive past 50 years of age because of associated congenital defects or the subsequent development of atrioventricular valvular insufficiency or heart block or both. We describe four men with uncomplicated C-TGA. Our patients are of interest for the following reasons: (a) their condition is very rare; (b) the diagnosis of C-TGA traditionally has been verified through invasive cardiac catheterization procedures; however, in our latest two patients, recently developed noninvasive diagnostic techniques played the decisive role in the diagnosis of C-TGA; (c) in these modalities, they presented as a “natural experimental model” that the right ventricle submitted to a high systemic pressure load is capable of increasing muscle mass over long-term adaptation. Our four patients illustrate that patients with C-TGA, even with the associated cardiac anomalies, may live a normal life span with proper management. (Chest 1992; 101:1382-85)

Corrected transposition of the great arteries without associated intracardiac anomalies is a rare condition. The oldest patient in whom the diagnosis was made while living was described in the literature as being 80 years old; however, few patients with this anomaly survive past 50 years of age because of systemic (anatomic right) ventricular dysfunction or the subsequent development of AV valvular insufficiency or heart block or both. In this report we describe four elderly men more than 50 years old with uncomplicated C-TGA.

CASE REPORTS

Case 1

A 67-year-old man was referred to our hospital for evaluation of an abnormal ECG in June 1990. He had never experienced exertional dyspnea, chest pain, palpitations or ankle edema.

Physical examination revealed a healthy appearing, elderly man with a blood pressure of 154/84 mm Hg. The apical impulse was felt 1 cm lateral to the left middle-clavicular line. The second heart sound appeared to be single and its intensity was accentuated. A soft grade 1/6 holosystolic murmur was heard clearest at the left lower sternal border. The liver edge was felt at the right costal margin.

The cardiac silhouette was moderately enlarged with a tortuous descending aorta (Fig 1), and the ascending aortic shadow was not prominent in the right upper mediastinal border. A slightly increased pulmonary vasculature and an absent main pulmonary artery shadow were noted. The right pleura was thickened from an old pleurisy. On the ECG, a normal sinus rhythm was present, and the P-R interval was prolonged to 0.24 s. There were no septal Q waves in leads I, aVL, and V, and there were inverted T waves in leads I, aVL, and V,. The V lead showed an rsR pattern.

In two-dimensional echocardiography (Fig 2), the aorta arose from the anatomic RV which was identified by the presence of heavy trabeculation, and the three-leaflet tricuspid valve that inserted more apically than the bi-leaflet mitral valve. On the other hand, the pulmonary artery, identified by its right and left branches, arose from the anatomic LV. Color flow mapping revealed the presence of tricuspid regurgitation, but the severity was mild to moderate.

In ECG-gated MRI of a spine echo (1.250/20), the anatomic RV from which the aorta took origin had a much thicker wall than the

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FIGURE 1. Posteroanterior chest x-ray film of case 1.
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Figure 2. Two-dimensional echocardiograms of case 1. The parasternal long axis echocardiogram (top) and color flow mapping of tricuspid regurgitation (bottom) B, Ao, aorta; aRV, anatomic RV; aLV, anatomic LV; RA, right atrium; LA, left atrium.

Figure 3. Posteroanterior chest x-ray film of case 2.

Figure 4. ECG-gated MRI of case 2. Top: Upper horizontal MRI shows anterolaterally transposed ascending aorta, forming upper left cardiac border. AAo, ascending aorta; DAo, descending aorta; PA, pulmonary artery. Bottom: Lower horizontal MRI shows that the anatomic RV has a much thicker wall (2 cm) than the anatomic LV. aRV, anatomic RV; aLV, anatomic LV; RA, right atrium; LA, left atrium.

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2. Two-dimensional echocardiograms of case 1. The parasternal long axis echocardiogram (top) and color flow mapping of tricuspid regurgitation (bottom) B, Ao, aorta; aRV, anatomic RV; aLV, anatomic LV; RA, right atrium; LA, left atrium.

In a frontal projection of an IVDSA, the ascending aorta and main pulmonary artery were demonstrated to be reversed in origin from the respective ventricles, with the pulmonary valve located lower than the aortic valve.

On cardiac catheterization, the pressure study in the anatomic RV, LV and pulmonary artery was normal, and the cardiac index was 4.5 L/min/m².

Case 2

A 60-year-old man also was referred to our hospital for evaluation of an ECG abnormality in September 1990. He was asymptomatic and had a normal appearance with a blood pressure of 134/87 mm Hg. Apical pulse was felt at the left mid-clavicular line. The second heart sound was single and accentuated in the pulmonic area. There was a soft grade 2/6 holosystolic murmur heard clearest at the apex. The remainder of the physical examination was unremarkable.

On the chest x-ray film (Fig 3), the right superior mediastinal border was somewhat inconspicuous. A round left cardiac border and concave main pulmonary artery segment were noted. An old left clavicular fracture was noted. The ECG revealed left axis deviation with no septal Q waves in leads I, aVL, V₅ and V₆.

In two-dimensional echocardiography, atrioventricular and ventriculoarterial discordance without VSD or pulmonary stenosis were shown. Color flow mapping showed the presence of mild tricuspid regurgitation.

In a horizontal section of ECG-gated MRI (93/20) (Fig 4), four chambers were shown. The lateral ventricular chamber showed an approximately 2-cm thick wall in early diastole, with the ascending
aorta transposed anterolaterally and the main pulmonary artery related posteromedially.
In a frontal projection of IVDSA (Fig 5), the ascending aorta, which lay left of the pulmonary artery, arose from the trabeculated, anatomic RV. The anatomic LV chamber was smooth in contour and interposed between the pulmonary artery and the right atrium to which the superior vena cava was committed. The main pulmonary artery located posteroinferiorly to the ascending aorta was well demonstrated in the lateral view.

CASE 3
A 54-year-old man was admitted to our hospital with the chief complaint of dizziness in April 1988. He appeared healthy with a blood pressure of 120/80 mm Hg and a regular arterial pulse rate of 36 beats per minute. A grade 1/6 holosystolic murmur was heard at the apex. There was no peripheral edema.
A chest x-ray film (Fig 6) showed a rounded left cardiac border without enlargement. The ascending aortic shadow and the main pulmonary artery segment were absent. An ECG revealed complete AV block with a ventricular rate of 40 beats per minute.

Two-dimensional echocardiography showed the presence of heavy trabeculation and a three-leaflet tricuspid valve in the left-sided ventricle. Cardiac angiography disclosed that the left-sided ventricle was structurally identical with the anatomic RV. The right coronary artery was found to be bifurcate, while the left coronary artery was nonbifurcate. The pressure study was normal and the cardiac index was 3.2 L/min/m². Attempts to record the His bundle ECG were unsuccessful. Permanent epicardial pacemaker insertion was performed in May 1988.

CASE 4
A 55-year-old man began to have palpitations and dyspnea during exercise in 1982. Afterward, his condition progressively worsened to cause orthopnea usually at night. At the time of hospitalization in June 1982, he complained of difficulty in breathing, with a blood pressure of 100/72 mm Hg and an irregular arterial pulse rate of 87 beats per minute. The second heart sound was single and accentuated; a grade 3/6 holosystolic murmur was heard at the apex. The liver was palpable two finger-breadths below the costal margin. There was mild peripheral edema.
At admission, a chest x-ray film revealed marked cardiomegaly with pulmonary congestion. An ECG demonstrated atrial fibrillation with a rapid ventricular response and Q-S pattern in right precordial leads.

Two-dimensional echocardiography revealed that the aorta arose from the anatomic RV, the contractility of which was moderately and diffusely reduced.

Management of the patient with furosemide and digoxin quickly improved the state of congestion. On cardiac catheterization performed three months after the hospitalization, the pressure study in the RV, LV and pulmonary artery was normal and the cardiac index was 2.4 L/min/sq m. Left ventriculography disclosed moderate tricuspid regurgitation and an enlarged left atrium. The left-sided ventricle was structurally identical with the anatomic RV.
At discharge in October 1982, a chest x-ray film (Fig 7) revealed the straight left cardiac border because of the position of the ascending aorta. An ECG revealed a regular sinus rhythm with left axis deviation. Q-S pattern in leads V₁ and V₂, and inverted T waves in leads 1, aVL, V₁, and V₅.

**DISCUSSION**

We have described four unusual cases of uncomplicated C-TGA diagnosed in patients more than 50 years old. Our patients are of interest for the following reasons: (a) Their condition is very rare; only 60 comparable cases have been reported and there have been only a few cases reported in patients more than 50 years of age. (b) The diagnosis of C-TGA is sometimes presumed, based on clinical, electrocar-
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diographic or radiologic grounds, but traditionally has been verified through invasive cardiac catheterization procedures. In our latest two cases (cases 1 and 2), recently developed noninvasive diagnostic techniques such as MRI and IVDSA played the decisive role in the diagnosis of C-TGA. In these modalities, they presented as a "normal experimental model" that the RV, submitted to a high systemic pressure load, is capable of increasing muscle mass over long-term adaptation.

The clinical course of C-TGA is determined primarily by the severity of associated defects. It is estimated that only about 1 percent of individuals with C-TGA have an otherwise normal heart. Congestive heart failure associated with a large VSD has been the most common cause of death, with most fatalities occurring within the first year of life. Also common are varying degrees of heart block, which often progress to complete heart block due to an abnormally prolonged His bundle, as seen in case 3. Similarly, the left AV valve, which anatomically is the tricuspid valve, sometimes is congenitally incompetent, and regurgitation may occur at any age, as seen in case 4.

Even without associated malformations, or following surgical correction of these abnormalities, the question remains as to whether the anatomic RV is capable of sustaining adequate cardiac output over a normal life span. Benchimol and co-workers are dubious of a normal life span in patients with C-TGA. Graham et al also suggest that there may be gradual deterioration of the anatomic RV in adulthood in affected patients. On the other hand, the reports by Cumming and Rotem and Hultgren reviewing C-TGA without associated anomalies suggest that affected patients may live a normal life span with only minor impairment of cardiac function. Dimas et al also suggest that the anatomic RV can function appropriately over a long period in adult patients with C-TGA even in the presence of associated cardiac lesions. Our four cases also suggest that patients with C-TGA may live a normal life span; cases 1 and 2 have been healthy without medical management, case 3 has been asymptomatic with permanent pacemaker implantation, case 4 also had been free of symptoms with medication for eight years, but died suddenly of cerebral infarction in September 1990.

As noninvasive diagnostic techniques become more popular and established, as shown in case 1 and 2, it is anticipated that more cases of asymptomatic C-TGA will be diagnosed with greater ease and certainty. Our four elderly cases illustrate that patients with C-TGA, even with the associated cardiac anomalies, may live a normal life span by proper management. It is hoped that additional data on survival and ventricular performance in this group of patients will be forthcoming.

REFERENCES