Congenital Corrected Transposition of the Great Vessels in a 58-Year-Old Man*

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This is the case report of a 58-year-old living man with uncomplicated congenital corrected transposition of the great vessels proved by cardiac catheterization and selective cineangiocardiology. Only six patients (3 percent) have lived beyond the age of 40 years despite the theoretic speculation of a normal life span. Selective coronary arteriograms revealed that the left coronary artery supplied the anatomic right ventricle, and the right coronary artery the anatomic left ventricle.

The term, congenital corrected transposition of the great vessels, was introduced by Schiebler and associates in 1961 to differentiate from surgically corrected transposition of the great vessels which can be accomplished after a Mustard procedure.

The true incidence of congenital corrected transposition of the great vessels is not known. Fontana and Edwards reported this condition to occur in 1.4 percent of 357 specimens of congenital cardiac disease. More than 200 cases have been reported since the first clear anatomic description by Rokitansky in 1875. Uncomplicated congenital corrected transposition is rarely recognized in life because of normal cardiac hemodynamics, while the majority of the cases with associated intracardiac defects are recognized in early childhood. There were only 18 patients who were older than 20 years in several series totaling more than 100 reported cases. There are six patients who have reached the age of 40 years, the oldest patient being a 73-year-old man.

This paper reports the case of a 58-year-old man with congenital corrected transposition of the great vessels without any associated intracardiac anomalies.

**Case Report**

A 58-year-old man, father of five living children, was admitted to Good Samaritan Hospital on March 15, 1970, with a history of dizzy spells for the past 14 years. These spells were increasing in number and severity two months prior to admission. A heart murmur was noted to be present for a number of years associated with the "electrocardiographic abnormalities." The patient was informed that he had narrowing of one of the heart valves. His dizzy spells have been associated with diaphoresis and mild dyspnea without any definite chest pain. There was no history of syncopal episodes, paroxysmal nocturnal dyspnea, orthopnea, or angina pectoris. The dizziness was precipitated by exercise and relieved by rest of 10 to 20 minutes. There was no history of rheumatic fever, and the family history was negative for congenital heart disease.

Physical examination revealed a healthy-appearing white man who appeared slightly younger than his stated age of 58 years. There was no cyanosis, clubbing of fingers or peripheral edema. The blood pressure was 130/84 on both arms. Pulse rate was 82 per minute with frequent irregularities due to extrasystoles. The neck veins showed normal "a" and "v" waves, and they were not distended. The lungs were clear to auscultation. The precordium was quiet and there were no thrills. The first heart sound was normal at the mitral area. The second heart sound was single and accentuated at the pulmonic area. There was a soft grade II/VI systolic ejection murmur best heard at the second left intercostal space, at the sternal border (Fig 1). A fourth heart sound was heard over the mitral area. The abdomen was soft, and the liver and spleen were not enlarged. All peripheral pulses were normal.

The electrocardiogram and vectorcardiogram taken at the time of admission are shown in Figure 2. The phonocardiogram, shown in Figure 1, confirmed the auscultatory findings. The chest x-ray picture is shown in Figure 3.

The patient was subjected to right and left heart catheterization and cineangiocardograms. The hemodynamic data are summarized in Table 1. The pressure on the venous ventricle was 20/4 mm Hg, and the pulmonary artery pressures were 20/6/12 mm Hg. With some difficulty, the catheter was passed across the pulmonary valve, and the "venous" ventri-
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CORRECTED TRANSP.—GREAT VESSELS

50-500 cps.

MA

TA

PA

AA

VT

L II

Figure 1. Phonocardiogram at the mitral (MA), tricuspid (TA), pulmonic (PA) and aortic areas (AA) with the carotid tracing (CT) and lead II of the electrocardiogram (see text).

Figure 2. Electrocardiogram and Frank vectorcardiogram. Note the presence of marked degree of left axis deviation resembling the pattern of left anterior hemiblock and abnormal T waves. The QRS-T angle is also increased.

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Figure 3. Chest x-ray films in PA, lateral, right and left anterior oblique projections. Note the round contour of the apex.

Figure 4. Ventricular angiogram. The contrast agent was injected in the left-sided arterial ventricle which connects with the aorta. This ventricle has the morphology of the anatomic right ventricle. The patient is in a right anterior oblique projection.
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Table 1—Hemodynamic Data

<table>
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<tr>
<th>Pressures mm Hg</th>
<th>SVC a-V-mean</th>
<th>RA a-V-mean</th>
<th>RV S/D</th>
<th>PA S/D/M</th>
<th>P Wedge S/D</th>
<th>LV a-V-mean</th>
<th>Aorta S/D/M</th>
<th>BSA M²</th>
<th>CI L/min/M²</th>
<th>SI ml/beat/M²</th>
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<tr>
<td>6/3/4</td>
<td>6/3/4</td>
<td>20/4</td>
<td>20/6/12</td>
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<td>116/67/88</td>
<td>1.95</td>
<td>2.5</td>
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<tr>
<td>HR beat/min</td>
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<td>300</td>
<td>1914</td>
<td>226</td>
<td>150</td>
<td>1607</td>
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Abbreviations: SVC: Superior vena cava RA: Right atrium P Wedge: Pulmonary wedge LV: Left ventricle HR: Heart rate VP: Ventricular power SP: Stroke power SER: Systolic ejection rate RV: Right ventricle PA: Pulmonary artery BSA: Basal surface area CI: Cardiac index ET: Ejection time TTI: Tension time index

Discussion

Congenital corrected transposition of the great vessels is characterized by transposition of the great vessels with inversion of the ventricles and of the atrioventricular valves. The right-sided ventricle receives venous blood from the right atrium through a bicuspid valve ("mitral valve"). This ventricle has the structure of the left ventricle which includes a bicuspid atrioventricular valve resembling the "normal mitral valve," smooth endocardial surface without trabeculation, and the absence of infundibulum or crista supraventricularis. The left-sided ventricle received arterial blood through a tricuspid valve, but has the structure of the normal right ventricle, which includes a tricuspid valve, infundibulum, crista supraventricularis, trabeculations, and a papillary muscle on its septal surface. Circulation in this type of congenital heart disease is usually normal. Systemic venous inflow reaches the normal right atrium and across the "bicuspid" valve (mitral valve) into the "venous" ventricle. This ventricle ejects blood into the normal pulmonary artery that arises posteriorly in relation to the aorta instead of its normal anterior position. The arterial oxygenated blood returning from the lungs reaches a normal left atrium, crosses a "tricuspid" left-sided valve, and reaches the "arterial" ventricle. Blood is then ejected into the aorta, which is abnormally placed anteriorly and to the left of the pulmonary artery. Therefore, despite the transposition of the great vessels, the resulting hemodynamic events are normal in that the inverted ventricles "correct" the transposition so that the systemic venous (unox ygenated) blood flows into the pulmonary artery, whereas the pulmonary venous (oxygenated) blood flows into the aorta.

The important feature in this case is the recognition of this congenital lesion in the older age group. This emphasizes the fact that this form of corrected transposition of the great vessels is compatible with long-term survival provided that there are no associated intracardiac defects or significant valvular insufficiency. Cardiac catheterization with selective cineangiography is mandatory to establish the correct diagnosis. Of additional interest in this case is that the coronary arteries were not transposed except for the minor abnormality of the early take off of the left anterior descending artery and the circumflex arteries (Fig 5). The left coronary artery supplied the anatomic right ventricle and the right coronary artery the anatomic left ventricle. Despite this unusual distribution of blood supply, the patient had no evidence of coronary insufficiency. The usual coronary artery distribution in this condition has been previously described.

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to the anterior descending branch and then continues in the right atrioventricular groove. The left coronary artery usually arises from the left aortic sinus and proceeds into the left atrioventricular groove with a left circumflex distribution, giving off a marginal branch and a posterior descending branch. However, there are many variations of the above pattern.4 9 10 The anterior aortic cusp is usually the noncoronary cusp.

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