Corrected Transposition of the Great Vessels without Associated Anomalies: Report of a Case with Congestive Failure at Age 45*

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The clinical course and laboratory findings in the case of a 45-year-old woman with corrected transposition of the great vessels and no associated defects are reported. Spontaneous systemic ventricular failure occurred at the age of 45 without obvious cause. The possibility is raised that this represents failure of the anatomic right ventricle to sustain systemic pressures for a normal lifetime. This patient is discussed in relation to present surgical techniques for correcting transposition of the great vessels which leave the right ventricle as the systemic pump.

Isolated congenital corrected transposition of the great vessels is an uncommon anomaly that results in normal circulating pathways. Although several types of corrected transposition are theoretically possible, the form of transposition with bulboventricular inversion represents the "classical" form of corrected transposition in the individual with situs solitus and is the only type of corrected transposition with a significant clinical incidence.1

The majority of reported cases of congenital corrected transposition of the great vessels have had one or more associated defects.2-11 In addition, interference with atrioventricular conduction resulting in all degrees of AV block has been a very common accompaniment with or without other defects.4,12

Much less frequent is the finding of congenitally corrected transposition of the great vessels without significant associated defects.12 The actual incidence of this anomaly is unknown as it does not frequently cause symptomatology, and, therefore, an indeterminate number of unrecognized cases undoubtedly exist.

The following case documents the onset of systemic ventricular failure (anatomic right ventricle) without an obvious inciting cause in a 45-year-old woman with uncomplicated corrected transposition of the great vessels.

CASE REPORT

A 45-year-old white woman enjoyed good health until April 1968 at which time she developed shortness of breath on exertion, orthopnea, increasing fatigueability with cough in the supine position, and paroxysmal nocturnal dyspnea. There were no symptoms of chest pain. She was in overt left-sided (anatomic right ventricle) congestive failure with a sinus tachycardia, presystolic and protodiastolic gallops, a holosystolic regurgitant apical murmur of left-sided atrioventricular valve insufficiency. Chest roentgenograms confirmed cardiomegaly and pulmonary congestion (Fig 1). This patient made a full recovery after treatment with bed rest, diuretics and digitalis. The gallop sounds and apical regurgitant murmur disappeared with cardiac compensation. She was then referred to Letterman General Hospital for further evaluation.

At the time of evaluation at Letterman General Hospital in August 1968, she was asymptomatic on digitalis. She was normal appearing and acyanotic, with a blood pressure of 130/80 mm Hg, and a regular pulse of 76 beats per minute. Jugular venous pulsations were normal and lung fields were clear to auscultation. Cardiac examination revealed no abnormal heaves or thrills. The point of maximum impulse was in the midclavicular line in the fifth intercostal space. The first sound was of normal intensity. The second sound was single and loudest in the left second interspace. There was a short grade II/VI systolic ejection murmur at the lower left sternal border which did not transmit well in any direction. There were no diastolic murmurs or gallop sounds. The remainder of the examination was unremarkable.

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Chest roentgenogram revealed a normal cardiac silhouette and clear lung fields (Fig 1). Cardiac series with barium swallow revealed no cardiac chamber enlargement and the barium outlined a normally positioned left-sided stomach. Electrocardiogram showed extreme left axis deviation with the initial forces abnormally leftward and posterior (Fig 2).

Diagnostic cardiac catheterization was performed. Catheter passage into the pulmonary artery was medial to the normal position of the pulmonary outflow tract and actually overlaid the spine. Right heart pressures and pulmonary wedge pressures were normal at rest and with exercise in the supine legs-up position. Retrograde left heart catheterization was performed, and there were normal left-sided pressures with no gradient across the aortic valve. The left heart catheter demonstrated that the aorta was positioned to the left of the pulmonary artery anterior to its normal position (Fig 3). Left ventricular cineangiogram demonstrated no evidence of atrioventricular valve incompetence or left-to-right shunting from a ventricular septal defect. The systemic left-sided ventricle had the configuration of a trabeculated chamber with an infundibulum which is usually characteristic of an anatomic right ventricle. The aorta arose to the left of the pulmonary artery anterior to its normal position and approximately side by side with the pulmonary artery. Selective coronary arteriography, performed by the Judkins' technique, showed widely patent and unobstructed vessels ruling out angiographically demonstrable occlusive disease. The major coronary artery arose posteriorly and to the left, and branched into three main vessels. One vessel was directed posteriorly along the atrioventricular groove and subsequently gave rise to the posterior descending vessels, supplying the diaphragmatic surface of the heart. The other major branch of the left coronary artery branched into three vessels which supplied various aspects of the systemic ventricle. The second major artery, or right coronary artery, was not entered selectively, but filled adequately on systemic ventricular and aortic root angiograms. This vessel arose to the right and anteriorly and supplied the anterior surface of the heart. It was a much smaller vessel as compared to the left and was unobstructed.

In summary, the catheterization revealed corrected transposition with bulboventricular inversion in situ solitus and no associated defects. At the time of catheterization, the patient was well-compensated with no evidence of abnormal pressures.

Subsequent to this study the patient remained compensated on digitals and diuretics for over a year but has recently redeveloped evidence of mild left-sided failure. In addition, several episodes of paroxysmal atrial tachycardia have been documented and are presently controlled on quinidine.
CORRECTED TRANSPOSITION OF GREAT VESSELS WITHOUT ASSOCIATED ANOMALIES

FIGURE 2. Electrocardiogram of a 45-year-old woman with uncomplicated corrected transposition of the great vessels demonstrating left axis deviation and altered initial forces which are directed leftward and posteriorly.

FIGURE 3. Postero-anterior projection of heart with arterial catheter retrograde from aorta into systemic ventricle (large arrow) and venous catheter from superior vena cava through right atrium, venous ventricle and into left pulmonary artery (small arrow). This demonstrates the aorta to the left of the pulmonary artery and the medial position of the venous ventricular outflow tract.

DISCUSSION

The preceding case documents the spontaneous onset of systemic ventricular failure (anatomic right ventricle) during the natural course of uncomplicated congenital corrected transposition. No inciting cause such as myocarditis, coronary atherosclerosis, valvular defects, or arrhythmias could be found to explain this break in cardiac compensation.

Complete transposition of the great vessels as opposed to corrected-transposition, accounts for about 8 percent of all congenital cardiac defects recognized at birth and 90 percent of these children die by the age of one if no surgical intervention is undertaken. A multitude of surgical procedures have been attempted at both palliating and correcting this lesion. Of the “corrective” procedures, the Mustard procedure, an intra-atrial pericardial baffle redirects arterialized pulmonary venous return to the tricuspid valve, right ventricle and hence the aorta. The systemic venous return is directed to the mitral valve, left ventricle and to the pulmonary artery. This results in physiologic correction of blood flow but does not correct the basic anatomy and the right ventricle remains the systemic pump. This procedure has gained wide application with an acceptable surgical mortality rate.

The question to be answered is whether or not the right ventricle can maintain its job as a systemic pump indefinitely. The naturally occurring counterpart of the surgically corrected transposition of the great vessels is the congenital defect demonstrated by the present case, namely, that of congenital corrected transposition of the great vessels due to bulboventricular inversion. In this defect, the anatomic right ventricle is the systemic pump. Greater than 90 percent of the reported cases of corrected transposition have associated defects and are not analogous to this situation. However, there are a small number of cases with isolated corrected transposition reported where the natural history might allow some insight to the durability of the right ventricle as the systemic pump.

It has been claimed that when corrected transposition is not associated with other anomalies, a fairly normal longevity is to be expected. In a few reported cases the patients have lived into the fifth and sixth decade. One patient, age 60, in a series reported by Schiebler and associates, had an isolated corrected transposition which was an incidental finding at autopsy. A 73-year-old man reported by Lieberson and co-workers had associated mild left atrioventricular and aortic regurgitation. Moss and colleagues have recently reported a case of uncomplicated corrected transposition in a 54-year-old man who had exertional breathlessness and nodal tachyarrhythmia and, although exercise pressures are not mentioned, he apparently had normal hemodynamics when he was studied. These cases point out that in certain instances, the anatomic right ventricle can act as a
systemic pump for long periods, even in the presence of valvular incompetence. However, these cases may indeed be the exception. Lieberson and associates\(^\text{16}\) in reviewing previously reported cases of corrected transposition, could find only six persons who had survived to age 40.

Of the reported cases of corrected transposition that are alleged to be uncomplicated, the majority have had either varying degrees of left AV valvular insufficiency, complete heart block, or both.\(^\text{12}\) With the onset of complete heart block, congestive failure is often superimposed and may be the cause of death. Berben and Adicoff\(^\text{18}\) have reported two cases of corrected transposition without associated defects who developed complete heart block in their mid 30's. In one of these cases, despite an artificial pacemaker, an inexorable course of congestive heart failure ensued over the next 1\(\frac{1}{2}\) years and finally death. This suggests that the systemic ventricle was unable to handle the pressure load for a sustained period of time.

The present case was not associated with either mitral insufficiency or complete heart block and yet the patient developed systemic or "left" ventricular failure without an inciting cause. It is conceivable that the patient's congestive failure was precipitated by unrecognized myocarditis or arrhythmia, but the possibility exists that the congestive failure was due to the inability of the anatomic right ventricle to sustain the pressure load. If so, this certainly is important knowledge for the cardiac surgeon who, in performing corrective or palliative procedures for complete transposition of the great vessels, has assumed that the right ventricle can sustain systemic pressures for a long period of time. While it is tempting to talk of the Mustard procedure as being a "total correction," it should be remembered that the correction is physiologic only. Whether or not this is a truly corrective procedure or just another, albeit highly successful, palliative procedure depends largely upon whether the right ventricle can maintain a systemic workload for a normal life span. It has been said that subtle differences between the ventricles fit them for their respective high and low pressure loads. In lesions where the RV functions at high pressure and high resistance, life expectancy has been greatly shortened with but few exceptions.

\[\text{References}\]


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