CASE REPORTS

Corrected Transposition of the Great Arteries

Associated Ventricular Rotation

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SUMMARY
Two patients with corrected transposition of the great arteries and ventricular rotation are described. The ventricular rotations encountered were about the longitudinal and the anteroposterior axes, respectively. The ventricular rotation affected the plane of the ventricular septum, the inflow and outflow tracts of the ventricles, and the interrelationship of the great arteries. Surgical closure of an associated ventricular septal defect was not without its problems.

Additional Indexing Words:
Ventricular septal defect
Cardiac embryology
Open-heart operation
Pulmonary stenosis

CORRECTED TRANSPOSITION of the great arteries (CTGA) is defined as transposition of the great arteries associated with ventricular inversion. Usually, when the heart is in situs solitus, transposition is of the levo form (aorta anterior and to the left), the mitral valve and morphologic left ventricle (pulmonary ventricle) are right-sided, and the tricuspid valve and morphologic right ventricle (systemic ventricle) are left-sided. The ventricular septum typically occupies a sagittal plane.

Occasionally, CTGA is complicated by bizarre positioning of the ventricles and ventricular septum. The heart may rotate about an anteroposterior or longitudinal axis, considerably modifying the interrelationships of the ventricles. In the two patients to be described, the ventricles occupied superior-inferior and anteroposterior positions, respectively. Both cases illustrate the difficulties imposed on surgical repair of an associated ventricular septal defect. Throughout the descriptions, the terms "right ventricle" and "left ventricle" refer to the respective morphologic characteristics of the ventricle, rather than to its laterality.

Report of Cases

Case 1
A 3½-year-old white boy was admitted to the Mayo Clinic on March 21, 1973. Heart disease was first noted when he was hospitalized for pneumonia at the age of 4 months. He had had three further attacks of pneumonia during the first year of his life. After cardiac catheterization at another institution when he was 7 months old, a diagnosis of CTGA, ventricular septal defect, and atrial septal defect was made and he was treated with digitalis. He was moderately active but tired easily, and had excessive sweating. Cyanosis of the lips with crying had been noted since he was 4 months of age. Syncope occurred on one occasion after a coughing spell. The patient had no siblings, and there was no family history of congenital heart disease.

At examination, the boy weighed 14.2 kg (25th percentile) and was 100 cm tall. There was no cyanosis or clubbing of the fingers. The heart was overactive, a grade 3/6 ejection systolic murmur was heard along the left sternal border, and an apical diastolic rumble was present. The second sound was loud and single.

A roentgenogram of the chest showed an enlarged heart (cardiothoracic ratio 0.66) and markedly increased pulmonary vascularity with attenuation of the peripheral pulmonary arteries. The electrocardiogram showed right bundle branch block, right ventricular...

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hypertrophy, and a small Q wave in V\textsubscript{6}. The hemoglobin level was 12.9 g/100 ml.

Cardiac catheterization was carried out before operation (table 1). The systemic flow was 3.7 L/min/m\textsuperscript{2}, and the pulmonary flow 6.5 L/min/m\textsuperscript{2}. The pulmonary resistance was 6.5 units m\textsuperscript{2}, the pulmonary-to-systemic flow ratio (Qp/Qs) was 1.8, and the pulmonary-to-systemic resistance ratio (Rp/Rs) was 0.4. Selective angiocardiograms were performed from the left (fig. 1) and right (fig. 2) ventricles. The atria were in situ solitus and both atroventricular valves appeared competent. The ventricles were rotated and tipped so that the left ventricle was inferior, the right ventricle superior, and the ventricular septum oriented transversely (fig. 2). This had the effect of elongating the left ventricular outflow tract and displacing the dilated pulmonary artery to the left so that it straddled the ventricular septum and faced the right (systemic) ventricle (fig. 3).

At operation, the external anatomy was typical for CTGA with ventricular septal defect. The aorta was small and located anteriorly and to the left of a dilated pulmonary artery. A 3 mm patent foramen ovale was palpated in the atrial septum. A large (1.5 by 1.3 cm) ventricular septal defect was located adjacent to the mitral annulus. The morphologic right (systemic) ventricle was located superiorly and the morphologic left (pulmonary) ventricle inferiorly. The pulmonary artery connection to the heart was displaced markedly to the left, producing a functionally double-outlet right ventricle, although there was still fibrous continuity between the right atroventricular valve and pulmonary artery posteriorly (fig. 3).

Cardiopulmonary bypass was instituted through caval catheters placed via the right atrial appendage and a metal cannula placed in the ascending aorta. Because the apex of the systemic ventricle could not be reached easily, the left side of the heart was vented through the left atrium anterior to the right pulmonary veins. The pulmonary artery was opened longitudinally, and the anatomy was inspected. Both ventricular cavities, particularly the right ventricle, were visible through the pulmonary valve. The margins of the ventricular septal defect could not be delineated clearly, so the right atrium was incised. The patent foramen ovale was closed by direct suture, and the cavity of the inferiorly placed left ventricle was...
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Figure 3

Case 1. (Upper Left) Intracardiac anatomy of usual case of corrected transposition of great arteries, as viewed through a cutaway of anterior walls of left and right ventricles. Intracardiac arrows show flow of blood, and external arrow illustrates direction of the additional rotation of ventricles which occurred in case 1. (Middle) Cutaway of anterior wall of right ventricle. Note marked displacement of pulmonary valve to left, resulting in overriding of ventricular septum. Mitral valve (MV) is visible through ventricular septal defect. (Upper Right) Diagram of surgical repair, as described in text.

inspected. As visualized through the mitral valve, the orifice of the elongated left ventricular outflow tract looked deceptively like a ventricular septal defect of the type seen in complete transposition of the great arteries, and this was inadvertently closed with a patch. Only when the anatomy was inspected again through the pulmonary artery was the error appreciated. It was not possible to reach the superior margin of the ventricular septal defect through either the posteriorly located pulmonary artery or the right atrium, so it was necessary to open the right (systemic) ventricle between two coronary arteries. The original patch was removed, and a larger patch was inserted in such a manner as to direct the systemic venous blood behind the patch and out the pulmonary artery (fig. 3). Bypass was discontinued without difficulty, and pressures (mm Hg) were measured: left (pulmonary) ventricle 55/5, right (systemic) ventricle 110/5, and pulmonary artery 45/15. The postoperative course was uneventful, and the patient was discharged on the 10th postoperative day in normal sinus rhythm.

Case 2

An 8-year-old white boy was admitted to the Mayo Clinic on May 7, 1973. His mother had contracted rubella 10 days after conception. The pregnancy was otherwise uneventful, but the patient was cyanotic and had a heart murmur at birth. Cardiac catheterization when the patient was 3 weeks old confirmed congenital heart disease of indeterminate type. In the 12 months prior to admission, the patient had noted an increase in cyanosis and a marked reduction in exercise tolerance. A second cardiac catheterization, also performed at another institution, demonstrated corrected transposition of the great arteries, ventricular septal defect, and pulmonary valve stenosis.

At examination, the boy weighed 21.6 kg and was 121 cm tall (less than the 10th percentile). There were moderate cyanosis and clubbing of the nail beds. The heart was quiet, a systolic thrill was palpable, and a grade 4/6 systolic murmur was present at the left sternal border. The aortic and pulmonary closure sounds were narrowly split.

A roentgenogram of the chest showed mild cardiomegaly (cardiothoracic ratio 0.53) and normal pulmonary vasculosity. The electrocardiogram showed sinus rhythm with a PR interval of 0.17 sec and intermittent nodal rhythm. There was no Q wave in V5 or V6, and the T wave was upright in V1 and V3R. Vectorcardiography showed an abnormally posteriorly directed QRS vector. The hemoglobin level was 14.9 g/100 ml.

Cardiac catheterization was repeated before operation (table 1). The systemic flow was 5.5 L/min/m2, and the pulmonary flow was 4.5 L/min/m2. The pulmonary resistance was 3.2 units m2, the pulmonary-to-systemic flow ratio (Qp/Qs) was 0.8, and the pulmonary-to-systemic resistance ratio (Rp/Rs) was 0.3. Selective angiograms were performed from the right (fig. 4) and left (fig. 5) ventricles. The atra were in situ solitus and both atriopulmonary valves appeared competent. The ventricles were rotated so that the left ventricle was posterior, the right ventricle was anterior, and the ventricular septum was oriented in a coronal plane (fig. 6). This resulted in elongation of the left atrium (fig. 5) and a rightward displacement of the aorta (fig. 6). A severe obstruction of the pulmonary outflow tract was noted (fig. 4).

At operation, the external anatomy of the heart was remarkably similar to that of complete transposition of the great arteries. The aorta was large and located anteriorly and to the right of a small pulmonary artery. The atrial septum was intact. A large (1.2 by 0.8 cm) ventricular septal defect was located to the right of the tricuspid orifice, adjacent to the septal leaflet of the tricuspid valve. The morphologic right (systemic) ventricle was located anteriorly and the morphologic left (pulmonary) ventricle was located posteriorly. The pulmonary valve was bicuspid with moderate fusion of the anterior commissure and some thickening of the cusps. Severe subvalvular stenosis, located 8 mm beneath the valve, was caused by a diaphragm of fibromuscular tissue.

Cardiopulmonary bypass was instituted through caval catheters placed via the right atrial appendage and a metal cannula placed in the ascending aorta. The right atrium was first incised inferiorly from the appendage, and the ventricles were explored through the mitral valve. The ventricular septal defect could not be visualized, so a longitudinal incision was made in the right ventricle; the defect then could be easily seen and closed with a patch. The pulmonary artery was incised longitudinally, the fused commissure of the pulmonary

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ventricular rotation in CTGA

Figure 4
Case 2. Right ventriculogram, lateral view. Note coronal plane of ventricular septum, elongated right ventricle (RV), and obstructed pulmonary outflow tract (POT). LV = left ventricle; PA = pulmonary artery; Ao = aorta.

valve was opened, and a large specimen of fibromuscular tissue was excised from beneath the valve.

Hemodynamics were generally good after the discontinuation of bypass although the pressure in the left (pulmonary) ventricle remained at systemic levels. Final pressures (mm Hg) were right ventricle 100/5, left ventricle 100/5, main pulmonary artery 50/20, aorta 105/70, left atrium 30/20, and right atrium 15/8. The residual pulmonary gradient was considered to be preferable to the resumption of bypass and the insertion of a conduit from the posteriorly located, relatively inaccessible left ventricle to the pulmonary artery. Cardiac pacing was instituted because of the presence of complete heart block.

The postoperative course was complicated by intractable heart failure and respiratory insufficiency. The patient died 3 weeks after operation. At postmortem examination, the anatomic findings were confirmed (fig. 7) and the repair of the ventricular septal defect was found to be accurate. Heart failure and death were probably related to a combination of the necessity for making a ventriculotomy in the systemic (right) ventricle, residual pulmonary stenosis, and heart block.

Discussion

During cardiac development only the intrapericardial, freely movable part of the embryonic heart (the bulboventricular loop) participates in rotation of the cardiac tube; the fixed, extrapericardial portions (the atria, sinus venosus, and trunco-aortic sac) cannot. Normally, a 90° rotation of the bulboventricular loop occurs about a longitudinal axis to the right (d-loop), but in CTGA it rotates an equal degree to the left (l-loop).2 However, occasionally in CTGA, for reasons that are obscure, the bulboventricular loop continues its rotation about the longitudinal or a different axis,3 with concomitant alteration in the plane of the septum, the inflow and outflow tracts of the ventricles, and the relationship of the proximal portions of the major arteries to each other.

Figure 5
Case 2. Venous phase of left ventriculogram, lateral view. Catheter has recoiled into right ventricle (RV). Note elongated “tunnel” of left atrium (LA) leading to tricuspid valve (TV). LV = left ventricle; Ao = aorta.

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Figure 6
Case 2. (Upper Left) Diagram of usual location of ventricles in corrected transposition of great arteries. External cardiac arrow indicates direction of additional rotation of ventricles in case 2. (Middle) Cutaway view of interior of right ventricle. Arrow indicates direction of flow of blood from venae cavae to posteriorly located left ventricle and thence out pulmonary artery. Note coronal plane of septum. (Upper Right) Diagram of surgical repair, as described in text.
In case 1, further rotation occurred about an anteroposterior axis, thus tipping the apex of the ventricles cephalad and to the left. Hence, the ventricular septum came to occupy almost a transverse plane, thus pulling the base of the left ventricle caudally, with the necessity for the left ventricular outflow tract to elongate markedly. A continued 90° counterclockwise (from below) rotation about a longitudinal axis occurred in case 2, thus rotating the right ventricle anteriorly. Consequently, the ventricular septum occupied a coronal plane; the left atrial-right ventricular inflow tract became markedly elongated and directed forward (the position of the left atrium remains relatively fixed), and the aorta and pulmonary artery rotated about each other proximally to assume the position of dextro-transposition (instead of the usual levo-transposition with ventricular inversion).

The cases described illustrate the surgical difficulties encountered as a result of ventricular rotation as well as inversion. There is a natural reluctance in CTGA to open the systemic ventricle, already handicapped in its requirement to develop systemic pressure by virtue of being a morphologic right ventricle. In most instances, the ventricular septal defect can be closed through the right atrium and mitral valve or pulmonary artery. However, in both patients in this report, a right ventriculotomy was unavoidable. In case 1, the superior margin of the ventricular septal defect could be visualized only through the right ventricle; indeed, incomplete exposure through the right atrium resulted in preliminary inadvertent closure of the left ventricular outflow tract. The right ventricle was directly anterior in case 2 and consequently was the only chamber through which the ventricular septal defect could be adequately visualized.

Acknowledgment

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References


Figure 7

Case 2. Autopsy. (A) Heart and lungs. Note anterior position of right ventricle, coronal plane of septum, and rightward location of aorta in relation to pulmonary arteries. (B) Close-up view of great arteries and cutaway of right ventricle.
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