Corrected Transposition of the Great Vessels in a 73-Year-Old Man

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SUMMARY
The case of a 73-year-old white man with corrected transposition of the great vessels and associated mitral and mild aortic regurgitation is reported. The patient's survival was the longest of any patient with this defect reported to date. Although the condition is theoretically compatible with a normal life span, few patients with this lesion survive past 40 years of age because of associated congenital defects or the subsequent development of A-V valvular insufficiency or heart block, or both.

Additional Indexing Words:
Mitral insufficiency       Aortic insufficiency

Corrected transposition of the great vessels is a relatively uncommon congenital defect. Approximately 200 cases have been reported to date. In contrast, transposition of the great vessels is a common congenital defect, always life threatening, and only recently becoming amenable to corrective surgery. Correction of this anomaly, however, using the Mustard procedure does not result in a normal anatomic situation, but one that simulates that of corrected transposition; namely, a normal right ventricular chamber, with its tricuspid valve, acting as a systemic ventricle.1

Since this surgically produced abnormality will occur with increasing frequency, the natural history of corrected transposition assumes greater importance in an attempt to prognosticate the late results of this type of surgery. With this in mind, the case of a patient who survived to the age of 73 years with corrected transposition of the great vessel is reported.

Report of Case
E. A., a 73-year-old white man, was admitted to the Indianapolis Veterans Administration Hospital for the eighth time on February 9, 1968, with a history of increasing shortness of breath. He had had an extensive cardiac evaluation in November 1966, at which time the diagnosis of corrected transposition of the great vessels was established. His previous admission had also been prompted by increasing shortness of breath, thought to be due to chronic obstructive pulmonary disease.

Physical examination revealed a regular pulse of 80, and blood pressure of 130/72 mm Hg. Examination of the chest revealed changes consistent with chronic obstructive lung disease. Cardiac examination revealed a normally placed point of maximal impulse in the fifth interspace, an extremely loud single second heart sound in the pulmonic area, a grade II/VI holosystolic murmur at the apex, and a grade II/VI decrescendo diastolic murmur along the left sternal border. Chest x-rays (fig. 1) revealed absence of the normal pulmonary artery segment of the cardiac silhouette, in addition to pulmonary changes of chronic parenchymal and pleural disease. Results of the pulmonary function test are given in...
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Figure 1
Postero-anterior roentgenogram of the thorax on November 7, 1966.

Table 1. Repeated ECGs (fig. 2) was interpreted as showing an incomplete left bundle-branch block without any evidence of A-V block.

Cardiac catheterization findings are reported in table 2. The salient findings included a large pulse pressure, an elevated pulmonary artery pressure, an elevated left ventricular end-diastolic pressure and a subnormal cardiac index. Cineangiography revealed the aortic arch to arise anteriorly to the pulmonary artery, moderate arterial A-V valvular insufficiency (fig. 3A), mild aortic valvular insufficiency (fig. 3B), and mirror-image coronary arterial architecture. No difficulty was encountered in repeatedly entering the pulmonary artery from the right basilic vein.

On previous admissions the patient had responded well to therapy which consisted of antibiotics, intermittent positive pressure breathing, bronchodilators, and digitalis. During his final admission he was initially semicomatose, but once again responded to therapy, and recovered.

Table 2
Results of Cardiac Catheterization of Patient (November 1966)

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressure (mm Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left brachial artery</td>
<td>126/55, 82</td>
</tr>
<tr>
<td>Right atrium</td>
<td>A-6, V-4, 3</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>40/5</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>40/10, 25</td>
</tr>
<tr>
<td>Wedge</td>
<td>A-18, V-18, 13</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>118/16</td>
</tr>
<tr>
<td>Cardiac output</td>
<td>4.2 L/min</td>
</tr>
</tbody>
</table>

(2.6 L/min/m²)

*Per cent of normal.
†Normal is 83% at 1 sec; 92% at 2 sec; and 97% at 3 sec in our laboratory.

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sufficiently to be ambulant on the ward. ECG on admission was unchanged except for varying first and second degree A-V block, of the Wenckebach variety which subsequently cleared with administration of O₂ and discontinuance of digitalis. Blood gases, drawn in 1968 when the patient was no longer semicomatose but still had respiratory distress, showed a P₀₂ of 60 mm Hg and P₇₅ of 55 mm Hg. There was no edema or congestion. On the twenty-fifth hospital day he suddenly expired.
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thick. No shunt or obstructive defects were found, and the A-V valves were normally attached to their respective valve rings. No gross aortic or mitral valvular lesion was found to explain the insufficiency previously demonstrated.

Discussion

Corrected transposition of the great vessels has been stated on theoretical grounds to be compatible with a normal life span.2-4

The literature, however, does not support the theoretical benignity of this lesion, since only six patients whose cases have been reported previously have survived to reach the age of 40 years.3,5,6 In seven series reviewed which included 101 cases,2-4,6-9 only 18 patients were 20 years of age or older.

While approximately 80% of the patients whose cases were reviewed have had associated lesions which have limited the survival, 20% have been considered to have “uncomplicated” lesions. In reality, though, this is not true, as almost all of the additional 20% had complications either from birth or by the subsequent development of A-V valvular insufficiency, heart block or both. In fact, of 24 “uncomplicated” cases collected by Rotem and Hultgren, only three were free of these complications.4

The present patient seems to have been unusually fortunate in many respects. The mitral insufficiency was probably late in onset and not of a severe degree. A-V block was never a problem. The aortic insufficiency was insignificant. There was little atherosclerosis of the coronary vessels for his age.

While it is difficult to document the time of onset of the patient’s mitral insufficiency, no murmurs were noted while he was in the military service. When examined for employment by the Veterans Administration, no murmurs were documented nor was a murmur documented during the first three admissions to the Veterans Administration Hospital. That mitral insufficiency may be a late development is suggested by the fact that, of 25 adults (16 years of age or older) with this lesion, 40% had mitral insufficiency, as opposed to 18.5% of 76 patients in the pediatric age group. That early development of this

Figure 6
Left ventricular cavity demonstrating a normal aortic valve. Heavy trabeculation, a widely patent coronary artery, and the crista supraventricularis can also be seen.

At postmortem examination no specific precipitating cause of death was found. The aorta arose directly anteriorly to the pulmonary artery (fig. 4). The anterior coronary cusp was the non-coronary cusp, and the atrial appendages were juxtaposed to the left of the great vessels. The official pathological report stated that “there were three normal aortic cusps” and we could find no abnormality of the aortic valve when we examined the heart. The right coronary artery gave rise to the anterior descending and circumflex arteries. The left coronary artery passing to the left simulated the pattern of the right coronary artery but in mirror-image position. Only minimal atherosclerotic changes of the coronary vessels were present. The heart weighed 500 g. The arterial ventricle showed the anatomic configuration of a normal right ventricle (figs. 5 and 6) with a tricuspid type of A-V valve, a crista supraventricularis, and a heavily trabeculated chamber. Many chordae tendineae, including all those to the small cusp, arose directly from the ventricular musculature and not from the papillary muscles. The pulmonary artery arose directly posterior to the aorta from a normal-appearing left ventricle, which displayed a bicuspid A-V valve and a smooth interventricular septum. The systemic ventricle was 20 mm thick, while the ventricle leading into the pulmonary artery was 10 mm thick.
lesion often is clinically significant cannot be denied, as five deaths (all in adult age range) have probably been attributable to this complication.10

Heart block in the present case occurred only during overdigitalization. Its absence at other times is noteworthy as 35% of the 18 patients over 20 years old in the collected series of 101 cases had first degree heart block and 41% third degree heart block. This compares to 25% and 18%, respectively of those under the age of 9 years. This complication appears to be of major significance in the death of three patients without other complications at the age of 35, 36, and 45 years, respectively.11-13

The relationship of the mild aortic insufficiency to the patient’s other congenital lesion is difficult to determine. There was no evidence of syphilis of the aorta, and no evidence of aortic, aortic ring, or aortic valvular diseases at postmortem examination.

A 15-year-old boy with associated unexplained severe aortic insufficiency has been reported14 and there have been other patients with murmurs compatible with aortic insufficiency,4-7 but it is difficult at this point to rule out possible fortuitous association of these two lesions.

The case herein reported is significant in that it testifies to the fact that corrected transposition of the great vessels can be a benign lesion. That ideal circumstances are rarely met is obvious from the infrequency with which this lesion is reported or found at routine postmortem examination in the adult. While modern cardiovascular surgery is likely to have a beneficial effect on the course of corrected transposition by correction of the associated defects, it is likely to produce many more patients with surgically produced “corrected” transposition. It seems likely that some of these patients may go on to develop the same complications as those with the naturally acquired disease.

References
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