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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CORRECTED TRANSPOSITION IN A 73-YEAR-OLD MAN

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 diff-

Table 1
Pulmonary Function Studies

<table>
<thead>
<tr>
<th></th>
<th>Value</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Expiratory capacity (ml)</td>
<td>1450</td>
<td>62*</td>
</tr>
<tr>
<td>Expiratory reserve volume (ml)</td>
<td>350</td>
<td>44*</td>
</tr>
<tr>
<td>Vital capacity (ml)</td>
<td>1800</td>
<td>57*</td>
</tr>
<tr>
<td>Maximal voluntary ventilation (L/min)</td>
<td>31.3</td>
<td>40*</td>
</tr>
<tr>
<td>Forced expiratory volume (L/min)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 sec</td>
<td>1100</td>
<td>88†</td>
</tr>
<tr>
<td>2 sec</td>
<td>1250</td>
<td>100†</td>
</tr>
<tr>
<td>3 sec</td>
<td>1250</td>
<td>100†</td>
</tr>
<tr>
<td>Peak expiratory flow (L/min)</td>
<td>200</td>
<td>43*</td>
</tr>
</tbody>
</table>

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

Electrocardiogram.

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, 23</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>
<tr>
<td></td>
<td>(2.6 L/min/m²)</td>
</tr>
</tbody>
</table>

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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 P₉₂ of 55 mm Hg. There was no edema or congestion. On the twenty-fifth hospital day he suddenly expired.

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**Figure 3**

(A) Left ventricular cineangiogram in right anterior oblique position. Note moderate A-V valvular insufficiency and heavy trabeculation of ventricular wall. (B) Supravalvular cineangiogram in left anterior oblique position. Mild aortic insufficiency is present. Note root of aorta just posterior to rib cage. Both coronary arteries are visualized, the dark spot in the right coronary artery represents its division into anterior descending and circumflex branches.

**Figure 4**

Heart in situ at postmortem examination. String is around the base of the aorta, as it arises anterior to the pulmonary artery.

**Figure 5**

Arterial ventricular cavity demonstrating a tricuspid valve with many chordae tendineae arising from the ventricular wall. The transected coronary artery shows negligible disease.
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, the arterial 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
lesion often is clinically significant cannot be
denied, as five deaths (all in adult age
range) have probably been attributable to
this complication. 3, 7, 8, 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 compli-
cations at the age of 35, 36, and 45 years,
respectively. 11, 12

The relationship of the mild aortic insuf-
iciency 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 val-
ular diseases at postmortem examination.

A 15-year-old boy with associated unex-
plained severe aortic insufficiency has been
reported 14 and there have been other patients
with murmurs compatible with aortic insuffi-
cency, 4, 7, 8 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 "cor-
rected" transposition. It seems likely that
some of these patients may go on to develop
the same complications as those with the
naturally acquired disease.

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