Case Report

A Case of Corrected Transposition of the Great Vessels without Associated Intracardiac Anomalies

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CORRECTED transposition of the great vessels is a relatively rare congenital anomaly although being reported with increasing frequency. Corrected transposition of the great vessels is almost always associated with other intracardiac anomalies.\textsuperscript{1)-5)} Ventricular septal defect is the most common associated anomaly. Left-sided atrioventricular valvular anomalies including atrioventricular regurgitation and the Ebstein-type anomaly are said to be next in frequency. Pulmonary stenosis is also one of the associated anomalies frequently found.

Uncomplicated corrected transposition is an extremely rare condition.\textsuperscript{6),7)} In this case the circulation can be maintained in normal physiology and the transposition should be considered to be functionally totally corrected.\textsuperscript{1), 6), 7)} The term of transposition functionally totally corrected, associated with 'mitral' insufficiency is seen in the literature.\textsuperscript{7)} Cumming distinguished, however, corrected transposition of the great vessels without associated intracardiac anomalies from that associated with 'mitral' regurgitation.\textsuperscript{1)} He described 10 cases of this type of corrected transposition without associated anomalies from the literature prior to 1962 and added 2 cases of his own diagnosed clinically. In Cumming's 2 cases both showed the patency of the foramen ovale but there were no shunts detectable by oxymetry or dye dilution methods.

It is the purpose of this paper to report one clinically diagnosed case of corrected transposition of the great vessels without associated intracardiac anomalies.

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Case Report

A 13-year-old boy was referred to our clinic because of suspected congenital heart disease and was admitted for further examination on June 1, 1964. The patient was apneic following delivery and weighed 2,550 Gm. His mother had been well during the pregnancy and his 2 siblings are both healthy and without any symptoms of cardiac disease. A pediatrician noticed that the boy had a slightly asymmetric chest at the age of 3 years. There was no history of cyanosis or dyspnea. The patient did well in school and was able to swim and play baseball without unusual complaint. The school doctor found some auscultatory abnormalities in the patient who thereafter developed frequent episodes of slight chest pain and sometimes tightness of his chest mostly during exercise. Duration of the pain was only a few sec. and the location of the pain was very variable, occurring either the left or right side of the chest. Recently the patient consulted a doctor who found that he had a strongly accentuated second heart sound at the second left interspace without significant murmur and QRS abnormalities in the electrocardiogram.

Physical examination revealed a rather slender and alert boy, being 151 cm. tall and weighing 34 Kg. There were no signs of cyanosis, clubbing or dyspnea. Blood pressure was 110 systolic, 78 diastolic. There was no chest deformity. Pulsation was visible in the third and fourth left intercostal spaces with a slight heave.

The second sound was single and markedly accentuated at the second left interspace and a closure tap was palpable in the third left interspace. An early

![Fig. 1. Phonocardiograms taken at the left second interspace (above), right second interspace (middle) and apex (below). The upper record shows an accentuated second sound, systolic click and early diastolic murmur.](image-url)
A grade I, soft systolic murmur was heard over the heart and along the left sternal border. In the recumbent position a grade I, low-pitched, very short, early diastolic murmur was audible in the third left interspace. Phonocardiogram (Fig. 1) confirmed the auscultatory findings.

Postero-anterior chest X-ray (Fig. 2) showed a rounded and elongated contour of the left superior cardiac border. A short, slender mediastinal vascular shadow was observed above the cardiac shadow showing a slight convexity to the left. The right pulmonary artery was normal in size and position and the lung fields were normal. Left anterior oblique, right anterior oblique and lateral views of the chest were not remarkable.

The electrocardiogram (Fig. 3) showed normal sinus rhythm, marked left axis deviation around $-60^\circ$ and abnormal QS or q waves in leads II, III and aV$_F$. In chest leads QS was found in V$_{3R}$ and V$_{4L}$ with positive T waves. A tiny r was followed by an S in V$_{3R}$ through V$_{4L}$ and RS was found in V$_{3R}$ and V$_{6}$. Double two-step test was negative. Vectorcardiogram (Fig. 4) demonstrated QRS loop markedly
CORRECTED TRANSPOSITION OF GREAT VESSELS

Fig. 4. Vectorcardiogram by the Frank method. Frontal (above), horizontal (right, below) and right sagittal (left, below) planes. See text for discussion.

Table I. Cardiac Catheterization Data

<table>
<thead>
<tr>
<th>Location</th>
<th>Pressure (mm. Hg)</th>
<th>Oxygen (vol. %) (Van Slyke)</th>
</tr>
</thead>
<tbody>
<tr>
<td>SVC</td>
<td></td>
<td>12.3</td>
</tr>
<tr>
<td>Right Atrium</td>
<td>7-2</td>
<td>12.1</td>
</tr>
<tr>
<td>Right-sided Ventricle</td>
<td>22/3</td>
<td>12.4</td>
</tr>
<tr>
<td>Main Pulmonary Artery</td>
<td>29/15</td>
<td>12.4</td>
</tr>
<tr>
<td>Brachial Artery (O₂ Saturation)</td>
<td>98/62</td>
<td>16.6 (96.2%)</td>
</tr>
</tbody>
</table>

Fig. 5. Postero-anterior view of catheter entering the right pulmonary artery taken from TV screen. See text for description.
deviated to the left, posteriorly and superiorly.

The diagnosis of corrected transposition of the great vessels was suspected because of the strongly accentuated second heart sound at the left interspace, left axis deviation in the electrocardiogram and chest X-ray findings mentioned above.

Right cardiac catheterization was carried out on June 3, 1964. Catheter was introduced into the right pulmonary artery passing an unusually deviated course to the right at the base of the heart (Fig. 5). No intracardiac shunts were demonstrated and all pressure determinations were normal, as shown in Table I. Intracardiac electrocardiogram (Fig. 6) demonstrated the initial positivity of QRS complex showing rS pattern in the right-sided ventricular cavity.

Biplane angiocardiogram was performed with a catheter placed in the right
atrium. Anterior and lateral views (Fig. 7A and 7B) demonstrated the pulmonary trunk given off medially and posteriorly at an unusual position from the heart. The right-sided ventricle showed no infundibulum but a 'tail' formed toward the apex. The ventricle showed a very smooth intracavitary surface without trabeculation. Later films (Figs. 8A and 8B) demonstrated the aorta given off from the ventricle anteriorly and to the left of the pulmonary trunk. A distinct infundibulum and a crista supraventricularis could be seen in the lateral view of the left-sided ventricle.

**DISCUSSION**

Corrected transposition of the great vessels without associated intracardiac anomalies is an extremely rare congenital condition.1), 6), 7) Only 12 cases were reviewed by Cumming including 2 cases of his own.7) To our knowledge there have been no subsequent reports. A 13-year-old boy reported in this paper was suspected of having this type of corrected transposition of the great vessels because of characteristic auscultatory, radiologic and electrocardiographic features and was confirmed by cardiac catheterization and angiocardiography. Only 5 cases were diagnosed clinically out of 12 cases.7)

In corrected transposition of the great vessels the aorta arises to the left and anteriorly from the left-sided ventricle, and the pulmonary artery arises to the right and posteriorly from the right-sided ventricle. In addition, the left-sided ventricle has all the anatomical features of the usual 'right ventricle' with the infundibulum formed by protrusion of the crista supraventricularis
while the right-sided ventricle shows the anatomical features of the usual 'left ventricle', triangular in shape and with a less-trabeculated, smooth intracavitary surface. The right-sided ventricle (anatomical 'left') is connected invertedly to the right atrium and the left-sided ventricle (anatomical 'right') to the left atrium. As a consequence, the right-sided ventricle pumps venous blood from the right atrium to the pulmonary artery, and the left-sided ventricle pumps arterial blood from the left atrium to the aorta. This is the reason that this type of transposition of the great vessels is referred to as 'corrected'.

Although symptoms and signs of corrected transposition depend on the presence of associated abnormalities, several diagnostic features have been described and are valuable even in complicated cases. Strongly accentuated second heart sound at the left second interspace is the only and very characteristic auscultatory feature of corrected transposition of the great vessels. The aortic valve is placed not only to the left, anteriorly and closer to the chest wall but also at an abnormally higher position of the left-sided ventricle and this produces the unusual loudness of the second sound at the left second interspace. Genesis of an early systolic click heard in the sitting position and a soft, low-pitched, very short, early diastolic murmur in the recumbent position is not clear in our case. Both findings were not considered to indicate any incompetence of semilunar valves.

In the chest roentgenogram the straightened or rounded, elongated border of the upper left cardiac shadow, narrow cardiac waist, inconspicuous aortic knob and globular heart are suggestive features. In our patient a globular heart with rounded and elongated left upper border, narrow cardiac waist and small mediastinal vascular shadow and normal pulmonary markings are highly suggestive of corrected transposition in the presence of an accentuated second sound at the left second interspace.

In the electrocardiogram several characteristic features have been given attention although they are not diagnostic. Left axis deviation, QS or Qr pattern over the right precordial leads and absence of q waves in the left precordial leads are suggestive. A qR, qRS or QS pattern in III and aVF also have been reported by Fink et al. and Shiebler et al. A-V block of all degrees is frequently found. In our patient P-R interval was normal but marked left axis deviation around -60° and QRS abnormalities were observed. Abnormal QS or qrS patterns were observed in leads V3R, V1, II, III and aVF. Although the precise mechanism of the electrocardiographic abnormalities described in corrected transposition is not clear, the inversion of the conduction system is the probable explanation. Intracardiac electrocardiogram revealed rS pattern in the right-sided ventricle.
in our patient which might be incompatible with the above explanation. But conclusive evidence regarding this point would require left-sided catheterization and determination of left-sided intracavitary potentials. This was not done.

Routine right cardiac catheterization can be of great value in suggesting or confirming the diagnosis of corrected transposition. The course of the catheter into the pulmonary artery is unusually displaced medially to the right and does not pass the normal pulmonary conus. Difficulty in passing the catheter into the pulmonary trunk also has been reported. The catheter was inserted into the right pulmonary artery without difficulty in our patient and showed the markedly deviated pulmonary trunk to the right. Determinations of the pressure and oxygen content revealed no shunts and no stenosis or regurgitation.

Angiocardiography is the most conclusive method in establishing the diagnosis. Transposed great vessels are clearly demonstrated by using biplane angiocardiography. A smooth-walled, right-sided ventricle, triangular in shape forming a 'tail' toward the apex and without an infundibulum is definitive evidence that the ventricle is anatomically the 'left' one. The left-sided ventricle shows the infundibulum and crista supraventricularis forming a long outflow tract particularly in the lateral view and a well-trabeculated wall, both indicating the anatomical 'right' ventricle. As shown in Figs. 7 and 8, angiocardiography demonstrated characteristic features of corrected transposition of the great vessels in our patient. The angiogram revealed that the rounded and elongated left upper cardiac border was formed by the root of the aorta originating to the left, anteriorly and superiorly from the outflow tract of the left-sided ventricle.

The diagnosis of corrected transposition of the great vessels without associated intracardiac anomalies was suspected in our patient before catheterization and angiocardiography which confirmed it. The combination of the auscultatory, electrocardiographic and roentgenologic findings was valuable in establishing the diagnosis. The patient has been well except for slight chest pain. No anomalous coronary artery was demonstrated in the angiocardiography and the patient had no sign of cardiac dilatation nor left-sided heart failure. The chest pain was of very short duration on some occasions and seemed to develop after he became conscious of his cardiac abnormalities.

Coronary arteries are also usually inverted in the corrected transposition and the left-sided ventricle with the structure of a 'right' ventricle is given a high work load. This may be a contributory factor in the chest pain of our patient but remains speculative.
SUMMARY

Corrected transposition of the great vessels is almost always associated with other intracardiac anomalies. Patients without associated anomalies are extremely rare.

A 13-year-old boy with corrected transposition of the great vessels without associated intracardiac anomalies is presented and discussed. The characteristic combination of auscultatory, electrocardiographic and roentgenologic features was present.

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REFERENCES