CORRECTED TRANSPOSITION OF THE GREAT VESSELS

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Received November 29, 1960

Corrected transposition of the great vessels is a congenital anomaly first described by von Rokitsansky in 1875. In its most common form it consists of transposition and inversion of the great vessels and inversion of the ventricles, while the atria and their venous connections are normally situated. The right atrium connects with the left ventricle via a bicuspid valve on the right side and this right-sided, but anatomically left, ventricle joins the pulmonary trunk arising posteriorly and to the right of the aorta. Similarly, the left atrium connects via a tricuspid valve to an anatomically right ventricle on the left side, from which arises the aorta anteriorly and to the left of the pulmonary trunk. The great vessels then ascend side by side without crossing over in the normal manner. In this way the aorta receives oxygenated blood and the pulmonary artery desaturated blood. In addition the coronary arteries show inversion and transposition. The plane of the interventricular septum is rotated through 180 degrees so that it runs from the back to the front and from left to right (De La Cruz *et al.*, 1959). The interior of the right ventricle has all the anatomical features of a normal left ventricle, while the left-sided ventricular chamber has the features of a right ventricle. Walmsley (1931) has demonstrated the reversal of the usual pattern of the conduction system in that the atrio-ventricular bundle and its branches proceed along the septal wall of the right-sided ventricle. Cardell (1956) and Anderson *et al.* (1957) have reviewed the various types of corrected transposition that may occur. These types depend on the presence of sinu-atrial, ventricular, and bulbar inversion in various combinations.

Various embryological explanations have been given in an attempt to understand the mechanism of development of this abnormality. Spitzer's (1929) conception of the role of torsion and detorsion has been accepted by some, while others like Walmsley (1931) believe that situs inversus of the ventricular loop is the basic defect. More recently, De La Cruz *et al.* (1959) have presented a theory based on the findings of Kramer (1942), Davis (1927), Streeter (1948), and Licata (1954) who have studied a large number of human embryos at the Department of Embryology of the Carnegie Institute, Washington and other universities. They conclude that corrected transposition, in any of its varieties, is a complex malformation in which two embryonic elements are involved, namely the bulbo-ventricular loop and the cono-truncal septum. They propose an embryological explanation of the defect in a normally situated heart, in mirror image dextrocardia, and in dextro-rotation.

Corrected transposition rarely exists as an isolated abnormality. Cardell (1956) mentions two patients without associated abnormalities, one of whom had heart block. Edwards *et al.* (1954) quote a patient, in whom the defect existed as an isolated asymptomatic lesion. More recently Schaefer and Rudolph (1957) reported a patient who died in congestive heart failure at the age of 36 with heart block, but there was no other abnormality. The vast majority of cases have some associated lesion, an atrial septal defect, ventricular septal defect, patent ductus, coarctation, or pulmonary stenosis. In Anderson's excellent paper (1957) the commonest anomalies were ventricular septal defect and pulmonary stenosis. Kernen (1958) reported two cases with ventricular septal defect, patent ductus, and preductal coarctation of the aorta. Gibbons (1956) reported three

Since this paper was submitted for publication we have encountered three more cases of corrected transposition, two with Fallot's tetralogy and one with ventricular septal defect.

patients with pulmonary stenosis. The association of corrected transposition with ventricular septal defect and atrio-ventricular heart block is found with sufficient frequency to constitute a clinical syndrome. It has been suggested (Anderson *et al.*, 1957) that severe pulmonary hypertension occurs more frequently in corrected transposition with ventricular septal defect than in uncomplicated ventricular septal defect. Another defect of importance, occurring with or without the other defects, has been stressed by the Mayo group (Helmholtz *et al.*, 1956; and Becu *et al.*, 1955), namely left atrio-ventricular valve insufficiency and sometimes this is an Ebstein type of malformation. Anderson *et al.* (1957) also reported one case of left atrio-ventricular valve stenosis.

Five patients with corrected transposition seen in this institution in the past year are presented in some detail. The clinical and catheter diagnosis as well as the possibility of surgical repair of associated defects are discussed.

CASE REPORTS

Case 1. M.B., a European girl of 5, was first seen at the age of 3 months with feeding difficulty, dyspnoea, and a chronic cough, congenital heart disease being diagnosed. The child had always been under-developed and was disabled.

The physical signs included normal peripheral pulses and a blood pressure of 100/70 mm. Hg. The jugular venous pulse showed a dominant "a" wave. The heart was enlarged, a marked apical thrust being felt in the anterior axillary line in the sixth intercostal space. There was a palpable parasternal heave and systolic thrill with a palpable second sound at the second left intercostal space. On auscultation, a harsh, moderately loud (grade 3 of 6 grades) systolic murmur, stopping before the aortic second sound, was heard maximally at the fourth left intercostal space. The second sound was loud and closely split and was followed by a short, early diastolic murmur. At the apex there was an opening snap and a moderately loud (grade 2/4), short mid-diastolic murmur (Fig. 1). The electrocardiogram showed sinus rhythm with a normal P wave and a P-R interval of 0.20 sec., atrio-ventricular dissociation occurred intermittently, and Q waves were present in leads III and in AVF. The præcordial leads showed an R wave of 6 mm. and an S wave of 26 mm. in lead V1, and an R wave of 20 mm. and an S wave of 16 mm. in V6. No Q waves were present in the præcordial leads (Fig. 2a). The postero-anterior radiogram showed a globular heart with considerable cardiomegaly and grade 2/4 pulmonary plethora. There was a narrow pedicle with an invisible pulmonary artery segment (Fig. 3A).

The clinical diagnosis was ventricular septal defect with pulmonary hypertension and a large left-to-right shunt. The opening snap suggested additional left atrio-ventricular valve disease. At cardiac catheterization, the right ventricular and pulmonary arterial systolic pressures were equal to systemic and there was evidence of arterialization at the ventricular level. No wedge pressure was obtained. In this instance there was no difficulty in entering the pulmonary artery. The pulmonary blood flow was calculated at 14.7 litres/min. and the systemic at 4 litres/min. The pulmonary vascular resistance was 3 units. No postero-anterior or lateral films were taken with the catheter in the pulmonary artery, since the case was catheterized before we were aware of this condition.

Surgical correction of the ventricular septal defect, utilizing cardio-pulmonary by-pass, was performed. At thoracotomy corrected transposition was noted (Fig. 4) the pulmonary artery being situated posteriorly and to the right of the aorta. A branch of the right coronary artery crossed in front of the outflow tract of the functional right ventricle near the origin of the pulmonary trunk. Ventriculotomy was performed avoiding the coronary vessel. Complete heart block occurred before the patient was put on by-pass. The interior of the right ventricle had the anatomy of a left ventricle. The ventricular septal defect was very easily seen and closed with an Ivalon patch. The right ventricular pressure dropped from systemic level to 35 mm. Hg.

Despite the persistent heart block the patient did well except for one Stokes-Adams attack. Although unable to take part in vigorous exercise she lived a reasonably normal and active life. On examination, in addition to the signs associated with complete heart block, pansystolic and mid-diastolic murmurs and an opening snap were present at the apex (Fig. 1). A loud single second sound at the second left intercostal space suggested persistence of pulmonary hypertension. Re-catheterization one year after operation showed corrected transposition, complete repair of the ventricular septal defect, and normal pulmonary arterial pressures. The loud component of the second heart sound at the second left intercostal space was therefore aortic and not pulmonary in origin. She died suddenly 15 months after operation. At necropsy there was corrected transposition and an abnormal tricuspid left atrio-ventricular valve.

FIG. 1.—The pre-operative (A) phonocardiogram (Case 1) at the second left intercostal space (PA), fourth left intercostal space (4LS) and medial to apex (MA-INT) shows an "ejection" type systolic murmur. The second sound is narrowly split and accentuated in keeping with pulmonary hypertension. The pansystolic murmur, opening snap, and mid-diastolic murmur at the apex (MA) indicate left atrio-ventricular valve disease, confirmed by their persistence after repair of the ventricular septal defect (B). After operation, which has produced complete heart block, the systolic murmur becomes pansystolic at all areas and the pulmonary regurgitant murmur persists. The pre- and post-operative tracings are not recorded at the same sensitivity and the bottom two PCG's are not synchronously recorded with the top two.

FIG. 2.—The electrocardiograms in Cases 1 to 4 (a–d respectively) are discussed in the text.

Case 2. A. E., a European boy aged 14 years, was referred by a school doctor because of a cardiac murmur. There was a history of cyanosis and frequent respiratory infections in infancy, which had been attributed to thymus enlargement and for which X-ray therapy was administered. In childhood he was subject to frequent coughs and colds, tired easily, and was moderately disabled by dyspnoea. He was mentally retarded.

The physical signs at 5, 10, and 14 years were essentially the same. The pulse was always small and the blood pressure at times unrecordable. The jugular venous pressure was normal. The apex beat was thrusting and displaced slightly outwards. There was a marked parasternal lift and a palpable second sound in the second left intercostal space. A harsh, grade 3/6 short "ejection" systolic murmur maximal at the fourth left space, but radiating medially to the second space to the left and the right of the sternum was heard, and its intensity was increased after amyl nitrite inhalation. The second sound was greatly accentuated and closely split at both aortic and pulmonary areas where a systolic ejection sound was recorded. A soft, early diastolic murmur was audible at the fourth left intercostal space and there was a third sound and a doubtful mid-diastolic murmur at the apex.

The electrocardiogram (Fig. 2b) showed sinus rhythm with a P–R interval of 0.20 sec. In V1 there was a 26-mm. R wave and a 16-mm. S wave, in V6 a 12-mm. R wave and no S wave. Small Q waves were present in leads I, V5, and V6. The postero-anterior skiagram showed normal heart size and a convex shadow on the upper left-hand border (Fig. 3B), which pulsed freely on screening.

The clinical diagnosis was aortic stenosis with a left-to-right shunt at ventricular level. At cardiac catheterization the pulmonary trunk was entered, though with difficulty, and a systolic pressure of 80 mm. Hg was recorded there and in the right ventricle. The left ventricle was entered via a foramen ovale and the systolic pressure in the left ventricle and in a systemic artery were both 100 mm. Hg, thus excluding aortic stenosis. Unfortunately no samples were taken from the right ventricle or pulmonary artery. A shunt was not demonstrated at the atrial level. An angiogram from the left ventricle (Fig. 5a) showed the aorta arising from the left ventricle and running upwards along the left border of the heart. The pulmonary artery was also opacified suggesting a left-to-right shunt at the ventricular or pulmonary artery level. A week later he was re-catheterized from the left arm but on this occasion the right ventricle could not be entered. The right atrio-ventricular valve appeared to be abnormally placed. An angiocardigram from the right atrium (Fig. 5b) showed filling of the pulmonary arteries, the pulmonary trunk being medially placed.

FIG. 3.—(A) Anterior view of Case 1, showing the globular heart with considerable cardiomegaly and pulmonary plethora. The pedicle is narrow with an invisible pulmonary artery segment. (B) Case 2 has a normal-sized heart but there is abnormal shadow with a lateral convexity on the left upper border of the heart. (C) Anterior view in Case 3 shows a normal-sized heart with a pulmonary bay, a large right pulmonary artery and pulmonary oligæmia. (D) Anterior view of Case 4. Dextrocardia with situs inversus is shown and the heart is slightly enlarged. A prominent bulge above the left pulmonary artery is related to the superior vena cava and there is considerable pulmonary plethora.

A dye-dilution curve recorded at the brachial artery following injection into the right atrium showed considerable distortion of the disappearance slope consistent with a moderate left-to-right shunt. A catheter diagnosis was made of corrected transposition with pulmonary hypertension and a moderate left-to-right shunt presumably at ventricular level. Surgical correction of the ventricular septal defect is being considered.

FIG. 4.—Case 1. The heart, exposed at operation, shows the transposition of the great vessels with the aorta and pulmonary arteries lying side by side. The “right” coronary artery can be seen crossing the outflow tract of the right ventricle.

Case 3. L. D., a white boy aged 7 years, was first discovered to have congenital heart disease at the age of 9 months, when he became cyanosed during a respiratory infection. Since that time, his mother had observed cyanosis in cold weather and effort dyspnoea but no squatting. At the age of 2 he was admitted for excision of an accessory left cervical auricle. Thereafter his disability had increased, so that by the time of admission he was spending more time out of school than in. On examination, moderate cyanosis and clubbing was noted, the jugular venous pressure and pulses were normal and the blood pressure was 110/80 mm. Hg. The apex was in the normal situation, there was a lift in the second left intercostal space with a palpable second sound and a systolic thrill. A long, loud (grade 4/6) systolic murmur was present partially obscuring the aortic sound (Fig. 6A) radiating widely to the neck, the right of the sternum and the apex. A soft delayed pulmonary sound was heard at the second left intercostal space. Phonocardiography confirmed these findings (Fig. 6A), the A2–P2 interval being 0.08 sec. At the second right intercostal space the systolic murmur was quite intense and P2 was recorded. The murmur softened with amyl nitrite inhalation.

The electrocardiogram (Fig. 2C) showed sinus rhythm with a P–R interval of 0.18 sec. In V1 there was a 6 mm. R wave and a 2.5 mm. S wave, and in V6 a 9 mm. R wave and a 2 mm. S wave. Small Q waves were present in leads 1, AVL and V6. Deep, wide S waves were present in leads V2–V5. The anterior X-ray (Fig. 3C) showed a pulmonary bay, a prominent right pulmonary artery, and pulmonary oligæmia. The clinical diagnosis of Fallot's tetralogy was made.

At cardiac catheterization the left atrium and left ventricle were entered from the right atrium, but the pulmonary trunk could not be entered from the right ventricle. The systemic and right ventricular pressures were identical at rest and after circulatory manipulation with amyl nitrate and phenylephrine, proving the presence of a large ventricular septal defect (Vogelpoel *et al.*, 1959, 1960). Saturation data and dye dilution curves established a bidirectional shunt at atrial and ventricular levels. The major shunt was at the atrial level, mainly right to left. An angiogram from the right ventricle (Fig. 6B) showed infundibular stenosis

(a)

(b)

FIG. 5.—Case 2. A selective angiogram from the left ventricle (a) outlines the aorta arising from the left ventricle and running up along the left upper heart border. The selective angiogram from the right auricle (b) shows filling of the pulmonary arteries, the pulmonary trunk being medially placed.

FIG. 6.—Case 3 (A). A long systolic murmur partially obscuring A2 is shown at the 2nd left and right intercostal spaces radiating to the 4th left intercostal space and apex. A soft pulmonary second sound at the 2nd left and right intercostal spaces the A2–P2 interval being 0.08 sec. (B) The selective angiogram from the right ventricle outlines the narrowed outflow tract of the right ventricle and the pulmonary arteries displaced to the right. The right pulmonary artery is far larger than the left. The aorta ascended to the left of the pulmonary trunk.

and displacement of the pulmonary arteries markedly to the right. The right pulmonary artery was disproportionately enlarged compared with the left. An angiogram from the left ventricle showed the aorta to the left of the pulmonary arteries, which were slightly filled from the left ventricle. An angiogram from the left atrium filled all chambers, aorta, and pulmonary trunk. The catheter diagnosis was “pentalogy” of Fallot, with bidirectional shunts at both atrial and ventricular levels and corrected transposition. Although the pulmonary trunk could not be entered, there was sufficient evidence to support the presence of severe pulmonary stenosis. Operation was not advised.

Case 4. C. G., a European man aged 35 years, was discovered to have heart disease at the age of 6 months. He had always been cyanosed and was moderately disabled by effort dyspnoea. The physical signs included slight cyanosis and clubbing of fingers and toes, a full peripheral pulse at 45 a minute, and a blood pressure of 130/75 mm. Hg. Dominant “a” waves were seen in the jugular venous pulse and at times these became “cannon” waves. There was complete dextrocardia with situs inversus. The cardiac impulse was normal and there was a systolic thrill maximal over the third right intercostal space to the right of the sternum. The first heart sound was constant and wide splitting of the second heart sound could be heard in the second to fourth right intercostal spaces. The pulmonary component was of normal intensity but markedly delayed (0.08 sec. in held expiration). A grade 4/6 pansystolic murmur maximal in the second right intercostal space was present and after amyl nitrite inhalation there was no change in heart rate nor in intensity of the murmur. There was no diastolic murmur.

The cardiogram (Fig. 2d) showed a bradycardia of 45 a minute and a P–R interval of 0.12 sec. The rhythm varied between high and low nodal. The P waves were inverted in leads 2, 3, AVF and across the right præcordium from V4R to V7R, and were upright in AVR, AVL, V3R, V2, and V1. Reversing the right and left limb leads did not alter the polarity of the P waves, thus favouring an abnormal origin of the impulse. The QRS complex showed an rS pattern in V2, a 5-mm. R wave and a 56-mm. S wave in V3R and a 12-mm. R wave and no S wave in V7R. Small q waves were present in V6R and V7R.

The X-ray (Fig. 3D) showed dextrocardia with situs inversus and a slight increase in heart size with a prominent bulge above the left pulmonary artery and moderate pulmonary plethora.

The clinical diagnosis was dextrocardia with situs inversus and mild pulmonary stenosis with a bi-directional shunt at atrial level. Corrected transposition was suspected because of the arrhythmia.

At cardiac catheterization, the catheter was advanced from the right basilic vein into the superior vena cava and right atrium situated on the left. The right ventricle was entered but not the pulmonary trunk. The catheter was repeatedly and readily passed from right atrium into the left atrium and left ventricle. The pressure in the right ventricle was 60/5 and in the left 140/10, a simultaneous pressure in the femoral artery

FIG. 7.—Case 5. The electrocardiogram shows right ventricular hypertrophy. The phonocardiogram shows a short "ejection" type systolic murmur associated with pulmonary flow and/or a ventricular septal defect with balanced pressures and left-to-right shunt. The second sound at the second left intercostal space (PA) is normally split and accentuated, being followed by an early diastolic murmur. At the second right intercostal space (AA) a typical continuous murmur is recorded.

being 140/75 mm. Hg. The pressure in the left atrium was 8/4 with a mean of 4, and the right atrium 4/2 with a mean of 3 mm. Hg. The peculiar bulge along the left upper cardiac border was related to the superior vena cava. There was clear cut evidence of arterialization in the superior vena cava and the right atrium. Samples from the left atrium, left ventricle, and femoral artery had an identical saturation of 82 per cent while that from the right ventricle, was 79 per cent. A selective angiogram from the left ventricle showed the aorta arising from the left ventricle and occupying the upper right cardiac border, whereas an angiogram from the right ventricle showed a centrally placed pulmonary trunk, establishing the presence of corrected

transposition. Dye dilution curves recorded at the femoral artery following injection into the right ventricle showed disproportionate prolongation of the disappearance slope suggesting a large left-to-right shunt, whereas the curve following injection into the superior vena cava showed a large right-to-left shunt in addition to the prolonged disappearance slope, thus proving a right-to-left shunt at atrial level. The catheter findings therefore revealed the presence of a bidirectional shunt at atrial level. Since the left and right atrial pressure pulses were so dissimilar, a common atrium seemed unlikely. Atrial septal defect with partial anomalous venous drainage into the superior vena cava appeared to be the most likely diagnosis. Dextrocardia and corrected transposition have been shown and there may be mild pulmonary stenosis. The patient has refused operation.

Case 5. S. N., a Bantu girl of 4, was first seen at the age of 9 months because of failure to thrive. A diagnosis of ventricular septal defect was made on the basis of a loud parasternal murmur and thrill. By the age of 2, effort intolerance had developed and evidence of left ventricular enlargement was present with an apical mid-diastolic murmur as well as the widespread systolic murmur. Frequent colds and poor development remained prominent features and since she failed to improve she was admitted for investigation.

Slight clubbing of the fingers was noted and cyanosis suspected (always difficult to assess in the darkly pigmented races). The peripheral pulses were normal and the blood pressure was 100/70 mm. Hg. The jugular venous pulsations and pressures were normal. The heart was enlarged and an apical thrust was felt in the sixth space in the mid-axillary line. There was a palpable parasternal heave and the second sound could be felt in the second left intercostal space. A widespread systolic thrill maximal at the fourth parasternal space was present. At the apex there was a soft blowing systolic murmur followed by a third sound and short soft diastolic murmur, while at the fourth parasternal space the systolic murmur was louder (4/6) but also short, stopping clearly before the second sound (Fig. 7). At the second left intercostal space the second sound was normally split (0.03 sec.) with a loud pulmonary component and an early diastolic murmur. At the second right intercostal space radiating to the midline there was a loud continuous murmur with all the features of a patent ductus.

The electrocardiogram (Fig. 7) showed sinus tachycardia of 110 a minute with a P-R interval of 0.16 sec. Right axis deviation was present with an R in lead I of 7.5 mm. and an S of 15 mm. In V1 the R wave was 28 mm. with an intrinsicoid deflection of 0.04 sec. and the S wave was 4 mm. A minute Q wave was present in V6 and V7. In V6 the R wave was 13 mm. and the S 8 mm. The postero-anterior radiogram showed a globular heart with a prominent left upper border, a narrow pedicle and marked pulmonary plethora (Fig. 8a).

The clinical diagnosis was ventricular septal defect with pulmonary hypertension, a right-sided patent ductus, and pulmonary regurgitation. At the first cardiac catheterization (from the right arm) the aorta was readily entered from the right ventricle, but the pulmonary trunk could not be entered. The right ventricular and aortic systolic pressures were identical and there was evidence of a bi-directional shunt at ventricular level with an arterial saturation of 88 per cent. Selective angiocardiograms showed good filling of a small aorta which was higher and more lateral than usual, both aorta and pulmonary arteries filling from the right ventricle.

A second investigation (via the saphenous vein) confirmed the left ventricular desaturation (right-to-left shunt). Contrast material injected into the left ventricle outlined the transposed aorta (Fig. 8c) and showed the left ventricle occupying the upper border of the heart normally formed by the right ventricular outflow tract. A left-to-right shunt at ventricular level was confirmed and contrast medium injected into the right ventricle showed simultaneous opacification of the aorta and pulmonary arteries with medial displacement of the latter (Fig. 8b). The pulmonary trunk and main branches were considerably larger than the small aorta. A lateral angiogram from the right ventricle showed the aorta and pulmonary trunk superimposed, both arteries lying side by side in the same horizontal plane.

The diagnosis following cardiac catheterization was corrected transposition with ventricular septal defect and bi-directional shunt. Although the pulmonary trunk was not entered there was no phonocardiographic or clinical evidence of pulmonary stenosis and we thus assumed high grade pulmonary hypertension. The continuous murmur audible on the right side was attributed to a right pulmonary artery stenosis rather than an aortico-pulmonary communication.

DISCUSSION

Although once considered a rare anomaly, the increasing reports in recent years suggest that corrected transposition is not uncommon and will be more frequently recognized as clinicians and

FIG. 8.—Case 5. The anterior X-ray (a) shows a globular heart with a prominent left upper border, a narrow pedicle, and much pulmonary plethora. A selective angiogram from the right ventricle (b) shows simultaneous filling of the aorta and pulmonary arteries with medial displacement of the latter. A selective angiogram from the left ventricle (c) shows the abnormally placed left ventricle forming the left upper border of the heart, usually formed by the right ventricular outflow tract, and the transposed aorta is clearly delineated.

pathologists become aware of its existence. This is illustrated by our experience of finding five cases in one year after our attention had been drawn to the condition.

In Cardell's report of published cases (Cardell, 1956) there was an equal sex ratio, and most other reports show no sex bias (Kernen, 1958). Anderson *et al.* (1957), noted a preponderance of males as in ordinary transposition. In our series three were male and two female.

As uncomplicated corrected transposition produces no physiological disturbance, the diagnosis can usually only be made at necropsy as described by Edwards *et al.* (1954), Cardell (1956), and Schaeffer and Rudolph (1957). Diagnostic clues during life may be provided by the presence of complete heart block or signs relating to the anomalous position of the aortic and pulmonary valves. Thus splitting of the second sound may be better heard along the right sternal border than along the left (Gasul *et al.*, 1959). In some cases a loud second heart sound may be heard in the second left interspace due to aortic valve closure, but this is generally misinterpreted as pulmonary valve closure indicating pulmonary hypertension.

The presence of associated abnormalities, however, is almost invariable and these direct attention to the heart. In our series, three had ventricular septal defects, one had Fallot's pentalogy, and one was a complex abnormality with a bi-directional shunt at atrial level, dextrocardia, and possibly pulmonary stenosis. Malformation of the left atrio-ventricular valve was suspected in one patient who had an opening snap and mid-diastolic murmur which persisted after surgical closure of a ventricular septal defect (Fig. 1): at necropsy an abnormal deformed left tricuspid valve was found.

The clinical diagnosis of the associated abnormalities is usually not difficult and certain features may suggest the presence of corrected transposition. When pulmonary stenosis is present the systolic murmur may be better heard along the lower left sternal border than the second and first intercostal spaces and the loud aortic component of the second sound may be best heard in the second left interspace (Gibbons *et al.*, 1956; Anderson *et al.*, 1957). These findings were not present in Case 4 where pulmonary stenosis was suspected but not proven owing to difficulty encountered in entering the pulmonary trunk. We were unable to confirm better splitting of the second heart sound along the right sternal border than along the left; in our Case 3 with tetralogy of Fallot and an atrial communication the second heart sound was widely split in both areas. In ventricular septal defect with pulmonary hypertension (Cases 1, 2, and 5) the second sound was unusually loud at all areas, but particularly so in the second intercostal space on the left, and the two components were superimposed.

Anderson *et al.* (1957) drew attention to certain important electrocardiographic features. Atrio-ventricular block is frequently encountered and P-R intervals are often greater than 0.20 sec. dissociation or heart block occurring in 6 of their 17 cases. In Cardell's (1956) report 4 of 25 cases had atrio-ventricular block.

Of our 5 patients 3 showed some conduction disturbance: the first had a P-R interval of 0.20 sec. and intermittent atrio-ventricular dissociation and the second a P-R interval of 0.20 sec. The third had varying nodal rhythm with an ectopic pacemaker similar to a case reported by Anderson *et al.* (1957). Atrio-ventricular node disturbance, although highly suspicious, is not diagnostic of corrected transposition since we have a patient with ventricular septal defect and complete heart block in whom the great vessels were normally orientated. In former years, the association of ventricular septal defect with congenital heart block was stressed but this has not been confirmed in recent reports on large series (Kjellberg (1955), and Downing (1956)). Peaked P waves in lead II was a common finding in the series of Anderson *et al.* (1957) and was present in our Case 4. The QRS pattern is often described as reversed in that Q waves are present over the right præcordial leads and not over the left and the T waves are positive from V4R or V1 across the left of the chest; this may be due to depolarization of the septum occurring from the right to the left. However, similar changes may be found in ventricular septal defect with marked right ventricular hypertrophy in the absence of corrected transposition. In three patients reported by Kernen (1958), there was neither heart block nor a reversal of QRS pattern. In our 5 patients Q waves were present over the left præcordial leads in Cases 2, 3, and 4 but not in the other two. In the presence of pulmonary stenosis

Gibbons *et al.* (1956) described the mean QRS vector as directed quite posteriorly resulting in deep S waves in mid and right præcordial leads. Only one of Anderson's patients showed this feature but it was a striking finding in the tracing of our fourth patient with dextrocardia and suspected pulmonary stenosis. The presence of this type of electrocardiographic pattern may therefore be quite helpful.

The appearance of the anterior chest film may occasionally be of assistance as pointed out by Anderson *et al.* (1957). The ascending aorta coursing along the left upper border of the heart may produce a shadow with a straight or concave lateral border or at times a distinct convexity in the region normally occupied by the main pulmonary artery. The medially placed main pulmonary artery may produce an indentation of the barium-filled œsophagus in the anterior view, and occasionally the left pulmonary artery is seen to arise more medially than normally as in our Case 5, though Anderson *et al.* (1957), claim that this latter finding is occasionally encountered in uncomplicated ventricular septal defect. Just as frequently, however, there is nothing remarkable about the cardiac outline, except for the changes due to the associated defects. In Case 2 (Fig. 3), the typical radiological appearance was present; in Cases 3 and 5 there were certain suggestive features whereas in Cases 1 and 4 there were none.

The findings at routine cardiac catheterization may not be at all helpful in making the diagnosis unless steps are taken to identify the position of either the pulmonary trunk or the aorta, or preferably both great vessels, in the antero-posterior and lateral projections. The right ventricle may be entered by the catheter in an unusual way (as in Case 2), since the plane of the right atrio-ventricular valve is transverse rather than vertical. Many reports have emphasized the difficulty in entering the pulmonary trunk which is due to the acute bend the catheter must take, since the right atrio-ventricular valve and the pulmonary valve are close together as in the case of the aortic and mitral valves in the normal heart. The pulmonary trunk could not be entered in four of our patients. An antero-posterior X-ray of the catheter in the pulmonary trunk may not appear abnormal or it may show the trunk to be more medial than usual. The lateral film, however, will clearly show that the pulmonary trunk is far posterior to the anterior cardiac border and this will establish the diagnosis. It is essential that a true lateral picture be taken (Fig. 9).

FIG. 9.—It is essential that a true lateral picture is taken with the catheter in the pulmonary trunk. An incorrect diagnosis of corrected transposition was made in this case on these plates, the lateral view being too oblique.

If the pulmonary trunk cannot be entered, a catheter inserted into the ascending aorta from the femoral or brachial arteries will show the ascending aorta running along the left upper cardiac

border and in the lateral view it is abnormally anterior. If the left ventricle is entered, the aortic valve appears to be placed at a higher level than normal.

The simplest and most accurate way of demonstrating corrected transposition is by angiocardiology. Injection of opaque medium into the right ventricle will show the centrally and posteriorly placed pulmonary trunk (Fig. 5, 6, 8). If there is a ventricular septal defect the aorta may be outlined in its abnormal position (Fig. 8); if not, the aorta can be filled by the injection of dye directly into the left ventricle (Fig. 5, 8). The angiogram may show that the right ventricle lacks its usual conus and that the cavity of the right ventricle is smooth and devoid of significant trabeculae. There is a tail-like extension of the right ventricle towards the apex behind the left ventricle in many cases.

Surgical intervention is required only for the correction of the associated cardiovascular defects. Fink *et al.* (1958) state that prompt repair should be undertaken for surgically correctable defects, because the circulatory pathway is then restored to normality. Walker *et al.* (1958) reported on three cases of corrected transposition with complete heart block in which associated ventricular septal defects were successfully repaired. It would appear that established heart block is well tolerated during open-heart surgery for repair of ventricular septal defects. According to Kernen (1958), surgical experience indicates that the additional anomaly of corrected transposition does not alter the prognosis following repair of simple ventricular septal defects. When, however, cases have either no conduction defect or only partial heart block, the occurrence of complete heart block will be just as serious as in cases of uncomplicated ventricular septal defect undergoing heart surgery. Furthermore in cases without pre-existing heart block the predisposition to surgically induced complete heart block is apparently much greater than in uncomplicated ventricular septal defect. This may be related to the way in which the conduction system runs down the right ventricular aspect of the septum, making it difficult or impossible to avoid the bundle when placing the sutures. In our Case 1, heart block developed during operation before the ventricle was opened (or the defect exposed), emphasizing the ease with which heart block can develop even without surgical interference with the bundle.

The surgical approach to the septal defect is modified in the presence of corrected transposition for several anatomical reasons. The anterior descending branch of the "right" coronary artery which runs down over the outflow tract of the right ventricle is not a serious problem (Fig. 4), and both the pulmonary valve and the ventricular septal defect can usually be adequately exposed by tunnelling underneath this vessel. The absence of a crista-supraventricularis in the right ventricle permits the ventricular septal defect to be more easily exposed, as in our first case. However, difficulty may be encountered in repairing an abnormality of the left atrio-ventricular valve which is usually tricuspid. If this valve is grossly incompetent, and the repair at the time of open-heart surgery inadequate, the outcome may be disastrous. Pre-operative assessment of the state of the left atrio-ventricular valve is therefore important. Because of these considerations it seems highly desirable that the surgeon should know of the presence of corrected transposition before he is confronted with it at thoracotomy.

SUMMARY AND CONCLUSIONS

Corrected transposition without associated defects can usually be diagnosed only at necropsy.

When attention is drawn to the heart in life because of associated defects, certain features raise the possibility of coexisting corrected transposition. Thus, splitting of the second heart sound may be better heard at the right sternal border than on the left and an unusually loud aortic component can sometimes be heard at the second left intercostal space. In the presence of large shunts or pulmonary hypertension however, these signs become obscured. Signs of concomitant "mitral" valve disease may also arouse suspicion.

An important clue in some patients is the unusual appearance of the left upper cardiac border in the anterior X-ray due to inversion of the aorta and pulmonary trunk as they arise from the ventricles.

Perhaps the most suggestive feature of all is the presence of some degree of atrio-ventricular block on the electrocardiogram.

The diagnosis can be established at cardiac catheterization. If the catheter can be advanced into the main pulmonary artery (often difficult or even impossible) the anomalous position of the pulmonary trunk can frequently be identified in the anterior X-ray film and with more certainty in the lateral projection. The most certain method of establishing the diagnosis is by angiocardio-graphy which clearly demonstrates the anomalous position of the great vessels. The peculiar anatomy of the ventricles may also be identified.

The surgical treatment of the associated defects, particularly ventricular septal defect, is complicated by the difficulty of opening the "right" ventricle due to the abnormal course of the coronary arteries. The presence of uncorrectable "mitral" valve disease also adversely affects the surgical results. The frequent occurrence of heart block following surgical intervention is probably the major reason for advocating a conservative policy in the management of these cases.

We wish to thank Dr. G. Sutin for his co-operation, the members of the staff of Groote Schuur Hospital for referring cases for investigation and the superintendent, Dr. J. A. Burger, for permission to publish. We would also like to thank our surgical colleagues Mr. C. Barnard and Mr. W. Phillips for their continued co-operation.

We should like to acknowledge the great assistance received from Mr. L. W. Piller our chief technician and the help of Miss S. Joseph.

Part of the expenses for this work have been defrayed by grants from the Council for Scientific and Industrial Research. Our thanks are due to the City Council of Cape Town for their financial help to the Cardiac Clinic during the past years.

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