THE DIAGNOSIS OF CORRECTED TRANSPOSITION OF THE GREAT VESSELS

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Congenitally corrected transposition was, until recently, regarded as an extreme rarity, but is now being recognized more frequently during life. This anomaly in its uncomplicated form is associated with normal hæmodynamics and a normal expectation of life (Helmholz, Daugherty, and Edwards, 1956). It is, however, commonly complicated by abnormalities of the left atrio-ventricular (A-V) valve, and by other congenital heart lesions, notably ventricular septal defect (VSD), pulmonary stenosis, and single ventricle: the presence of corrected transposition introduces additional difficulties and dangers into the repair of these associated lesions. It is particularly important, therefore, that this anomaly should be recognized before operation; and it is the purpose of this article to review the clinical, radiological, cardiographic, and hæmodynamic features of corrected transposition, in the light of experience with our ten patients and of others that have been reported.

The pathological anatomy of three of these ten patients has been described elsewhere (Honey, Rushton, and Taylor, 1960; Honey, 1962) and the anatomical and physiological features of corrected transposition have been reviewed (Honey, 1962). Other detailed pathological studies have been reported by Lev and Rowlatt (1961) and by Schiebler *et al.* (1961).

The essential features are transposition with reversal of the antero-posterior relation of the aorta and pulmonary trunk, and inversion of the bulbo-ventricular portion of the heart so that the left-sided ventricle is morphologically a right ventricle, and vice versa. As a result, systemic venous blood returns to the right atrium and passes through a bicuspid right A-V valve into a right-sided ventricle which has the internal appearance of a left ventricle and lacks a crista supraventricularis and separate infundibulum: it is ejected into a right posterior pulmonary trunk. Pulmonary venous blood returns to the left atrium and passes through a tricuspid left A-V valve into a left-sided ventricle which has the internal appearance of a right ventricle with a crista supraventricularis separating its inflow portion from the anterior infundibulum: it is ejected into a left anterior aorta which passes upwards parallel to the pulmonary trunk without the normal spiral relation between the great vessels. The coronary arterial pattern is also inverted. The cusps, chordæ, and papillary muscles of the left A-V valve are frequently abnormal and the valve may therefore be incompetent (or less commonly, stenotic); and there may be a deformity resembling Ebstein's anomaly.

In a less common variant, there is situs inversus with dextrocardia and the appearance of the heart is the mirror-image of that just described: there is thus sinu-atrial inversion with normal bulbo-ventricular situs. Exceptionally, the common form may be associated with isolated dextrocardia or the less common form with isolated lævocardia. All the patients in the present series belong to the group with bulbo-ventricular inversion, and the discussion is therefore confined to this type of case. The developmental fault involved in all types of corrected transposition has been well discussed by de la Cruz *et al.* (1959).

There is no consistent practice in describing the laterality and morphology of the heart chambers and valves in corrected transposition, and the following rules are suggested. Ventricles should be referred to as left-sided or right-sided, according to their position in the body, and in addition should

be described, if possible, as morphological right or morphological left. The use of the terms "right" and "left" (with or without inverted commas) may be misleading. A "right" ventricle may be right-sided and morphologically left, or left-sided and morphologically right: both meanings have been used. The same rules should be applied to atria in the situs inversus heart. The A-V valves should properly also be referred to as right-sided or left-sided, but the use of the terms right and left A-V involves no loss of clarity: where possible they should, in addition, be described as bicuspid or tricuspid, as the case may be.

CASE REPORTS

The ten patients reported here included six with an associated VSD, in three of whom pulmonary stenosis was also present: two had left A-V valve abnormalities and two had a single ventricle. The electrocardiographic findings are illustrated in Fig. 1-5 and summarized in Table I. The chest radiographs of six patients are shown in Fig. 6-11. The observations made at cardiac catheterization are summarized in Table II. These three aspects are dealt with later in the discussion, so will be referred to only briefly in the Case Reports. The auscultatory signs were confirmed by phonocardiography.

Case 1. A boy, aged 3, was found to have congenital heart disease when he had pneumonia in infancy. He subsequently had repeated chest infections, but between these he was well and active.

He was pigeon-chested, with Harrison's sulci. A moderate parasternal heave suggested right ventricular hypertrophy, but there was no apical thrust. A chest radiograph confirmed moderate general enlargement of the heart, and showed pulmonary plethora and a narrow vascular pedicle (Fig. 6). A loud pansystolic murmur and thrill was maximal at the lower left border of the sternum and at the apex; and an apical crescendo-diminuendo mid-diastolic murmur followed the P wave, the unusual timing of this atrial systolic murmur being associated with partial heart block. Cardiac catheterization (Table II) confirmed the presence of a large VSD with moderate pulmonary hypertension.

At operation (Sir Russell Brock) for closure of the VSD, the great vessels were found to be transposed, the aorta (diameter 1.25 cm.) passing directly upwards from a large left-sided ventricle: the pulmonary artery (diameter 2 cm.) lay to the right of, slightly posterior to, and parallel with, the aorta, and also passed directly upwards. The presence of a VSD (diameter 1 cm.) in the pars membranacea was confirmed, and the defect was closed through a vertical incision in the right-sided ventricle, using complete cardiopulmonary bypass and potassium arrest.

His post-operative course was uneventful. Apical pansystolic and mid-diastolic murmurs were still present. Pulmonary vascularity was diminished, but the heart size was unchanged.

Comment. This boy had corrected transposition with a VSD. There was partial heart block (and transient complete heart block during catheterization), and in retrospect the appearance of the vascular pedicle might have suggested corrected transposition, but this was not diagnosed before operation. The A-V valve anatomy was not noted at cardiomy, but the persisting apical murmurs suggested the presence of mitral regurgitation, though the pulmonary artery wedge pressure record did not.

Case 2. A man, aged 19, was found to have a heart lesion at the age of 6 years. His disability was slight in childhood, but had increased particularly during the three months before admission for investigation. He was noted to have a slow pulse and, when he was 13, a cardiogram showed complete heart block.

He had a high-arched palate, dorsal scoliosis, and præcordial prominence of the chest. An apical thrust suggested left ventricular hypertrophy; and there was considerable cardiac enlargement on the radiograph which also showed increased vascular shadows in the upper zones and interlobular septal lines. There was a pulmonary systolic ejection click, a loud apical systolic murmur and thrill, and a short low-pitched apical diastolic murmur: the second sound was single or closely split and intense, and there was a high-pitched pulmonary early diastolic murmur. He had atrial fibrillation and complete heart block (Fig. 1). Cardiac catheterization (Table II) showed pulmonary hypertension with raised pulmonary resistance, and a doubtful small shunt at ventricular level.

Operation was not advised. His condition continued to deteriorate, he developed congestive cardiac failure and pulmonary infarcts, and died a year later at another hospital.

At necropsy, there was corrected transposition of the great vessels. The atrial and ventricular septa were intact. The left A-V valve was tricuspid; the arrangement of the chordæ and papillary muscles was abnormal and the base of the septal cusp displaced downwards; and the valve was clearly incompetent. Both ventricles were dilated and hypertrophied, and the pulmonary artery enlarged. The anterior descending branch arose from the right-sided coronary artery. The findings are described in detail elsewhere (Honey, 1962).

FIG. 1.—The electrocardiogram of Case 2. Below, lead III showing atrial fibrillation with complete heart block.

Comment. Corrected transposition was not recognized during life or at necropsy. On subsequent review the diagnosis was suggested by the association of complete heart block with clinical features of mitral regurgitation and this was confirmed by re-examination of the heart. The heart showed also the features of a partial left-sided Ebstein anomaly.

Case 3. A man, aged 19, was found to have a heart murmur at the age of 4 years, but was free from symptoms until four weeks before his admission to hospital with atrial fibrillation and cardiac failure.

The heart was large, with much right and left ventricular enlargement; and a chest radiograph showed a narrow vascular pedicle and pulmonary congestion. At the apex there was a harsh systolic murmur, a loud third sound, and a long soft rumbling diastolic murmur cardiogram; see Fig. 3 and Table I. He died within twenty-four hours of admission.

At *necropsy*, there was corrected transposition of the great vessels. The atrial and ventricular septa were intact: the right A-V valve was composed of a single continuous leaf, and the left A-V valve was tricuspid and incompetent.

Comment. This patient was known to have congenital heart disease and had the clinical features of mitral regurgitation, though a VSD had also been suspected. There was no heart block, but the vascular pedicle was surprisingly narrow. These features led Dr. E. W. Hancock to suspect the presence of corrected transposition.

Case 4. A man, aged 21, was found to have congenital heart disease in infancy. His disability was slight.

There was complete heart block (Fig. 2 and Table I), with a regular pulse (60 a minute) and cannon waves in the neck. There was biventricular hypertrophy on clinical examination, and the radiograph (Fig. 7)

FIG. 2.—The electrocardiogram of Case 4.

confirmed slight cardiac enlargement, with slight pulmonary plethora, and left atrial enlargement fluoroscopically. A long loud harsh systolic murmur was maximal down the left border of the sternum: at the pulmonary area, the murmur was an ejection murmur and there was a soft diminuendo early diastolic murmur. At the apex, a loud third sound and a short mid-diastolic murmur varied in intensity from cycle to cycle. The second sound was widely split and the pulmonary component soft.

Cardiac catheterization (Table II) seven years previously had shown a VSD with pulmonary stenosis, probably at valve level: the catheter passed into the ascending aorta which was shown to be to the left of the pulmonary trunk. On *angiocardiography*, injections were made into polythene catheters, passed into the right-sided ventricle by the saphenous route, and retrogradely into the left-sided ventricle. Both series (antero-posterior view) showed the ascending aorta passing upwards and to the right, lying to the left of the medially placed pulmonary trunk, before arching over the left pulmonary artery and descending on the left side. The ventricular septum and septal defect were clearly outlined, especially after injection into the left-sided ventricle: early films also showed regurgitation into an atrium, probably the left. A diagnosis of VSD and pulmonary stenosis with corrected transposition was made. Operation was advised, but has not yet been undertaken.

Comment. The association of complete heart block with congenital heart disease and the unusual position of the cardiac catheter in the aorta led to a confident diagnosis of corrected transposition; and the angiocardiograms confirm this. There was no clinical evidence of "mitral" regurgitation but the probable early appearance of opaque medium in the left atrium suggests that regurgitation through the left A-V valve may have occurred.

Case 5. A boy, aged 9, was found to have congenital heart disease when he had bronchitis in infancy. His disability was moderate but increasing, and he became cyanosed and occasionally squatted on exertion.

On examination, there was cyanosis and clubbing, and the jugular venous pulse showed prominent *a* waves. The heart was slightly enlarged, with a parasternal heave, and the chest radiograph confirmed slight enlargement but was not otherwise abnormal. A moderately loud systolic ejection murmur extended up to the loud palpable single second sound: a separate apical pansystolic murmur was noted.

Cardiac catheterization (Table II) showed a VSD with right-to-left shunt, due to presumed pulmonary stenosis. The pulmonary trunk was not entered, but the pulmonary vein wedge pressure was normal. Right and left ventricular pressures were identical, and the catheter was passed into a right pulmonary vein, being withdrawn through left atrium, left-sided ventricle, and probably right-sided ventricle, to the right atrium. Dye dilution curves from the ventricles were dissimilar, and single ventricle seems improbable. The catheter entered the aorta and right subclavian artery (Fig. 12): its course in the ascending aorta appeared unusually far to the left.

Angiocardiography. Injections were made into the superior vena cava and by percutaneous puncture into the left-sided ventricle: the needle-tip lay in the region of the VSD and most of the opaque medium passed directly into the right-sided ventricle. Both series showed a medially placed right-sided outflow tract and pulmonary trunk, and the site of the evident pulmonary stenosis was below the normal position of the pulmonary valve. The VSD was clearly seen, with dye streaming through it into the outflow tract of the left-sided ventricle and ascending aorta.

A diagnosis of VSD, pulmonary stenosis, and corrected transposition was made, and operation has been advised.

Comment. In this patient with a complex congenital abnormality, corrected transposition could only be diagnosed on re-examination of angiocardiograms made three years previously, the position of the catheter in the aorta being suggestive but not diagnostic in a cyanotic patient. There were no other features to point to this condition.

Case 6 (Case 27 of Campbell, 1961). A boy, aged 13, had been blue from birth, was very breathless on exertion, and squatted. When 5 years old, he was intensely cyanosed and clubbed; there was a pulmonary systolic murmur and a loud single second sound; and the cardiogram showed left ventricular dominance. A chest radiograph (Fig. 8) showed slight cardiac enlargement, and the straight upper left border of the cardiac silhouette was unbroken by the left pulmonary artery which was not visible. An angiocardiogram was thought to be suggestive of pulmonary atresia, but in view of the cardiographic pattern tricuspid atresia was diagnosed. A Blalock operation (end-to-side left subclavian-pulmonary anastomosis) was performed by Mr. B. B. Milstein, who observed that there was a large left ventricle from which arose a large aorta, but no obvious pulmonary trunk.

Following operation, his exercise tolerance improved greatly, he squatted less and the cyanosis and clubbing diminished. Later, his symptoms increased again and he was reinvestigated. At this time, he was

undersized, with moderate cyanosis and clubbing. There was a prominent *a* wave in the jugular venous pulse. In addition to the previous signs there was a widely-heard anastomotic murmur, and a third sound and short mid-diastolic murmur at the apex. A chest radiograph showed some increase in cardiothoracic ratio, but little other change.

Angiocardiography. A polythene catheter was passed from the saphenous vein to the right-sided ventricle. In the antero-posterior view, both ventricles filled and the VSD was clearly outlined: the outflow tract of the left-sided ventricle and the ascending aorta formed the upper left border of the heart. The pulmonary trunk was situated to the right of the aorta, the valve being unusually low and its orifice small. The lateral films showed the anterior aorta and the posterior direction of the outflow tract of the right-sided ventricle and pulmonary trunk.

A diagnosis of VSD, pulmonary stenosis, and corrected transposition was made. Operation has been advised.

Comment. This patient has most of the features of Fallot's tetralogy. The associated corrected transposition was recognized only on angiocardiology, though the surgeon had observed the anterior situation of the ascending aorta. The left axis deviation and apparent left ventricular hypertrophy had led to the suspicion of tricuspid atresia.

Case 7. This boy, aged 13, has been described elsewhere (Honey *et al.*, 1960). On clinical examination and at catheterization he was thought to have a VSD with pulmonary hypertension. He died following operation and was found to have a large common ventricle, with a rudimentary subaortic outflow chamber on the left side; the aorta was anterior, to the left of, and parallel to the pulmonary trunk; and both A-V valves opened into the common ventricle, the left being tricuspid and the right bicuspid.

Comment. The patient had cor triloculare biatriatum (single ventricle), with transposition of the great vessels and bulbo-ventricular inversion, the aorta arising from a left-sided bulbar chamber (type II A 1 of Harley, 1958). The association of transposition with bulbo-ventricular inversion is characteristic of corrected transposition and this was recognized at operation, though unsuspected beforehand: the presence of a single ventricle dominated the physiological situation but did not impair the functional correction, since a low pulmonary vascular resistance permitted a high pulmonary flow and normal arterial oxygen saturation.

Case 8. A boy, aged 6, was found to have congenital heart disease in infancy. He subsequently had repeated attacks of bronchitis, and was breathless on exertion, when he became cyanosed.

FIG. 3.—The electrocardiogram of Case 3. Präcordial leads half-standard.

FIG. 4.—The electrocardiogram of Case 8. Leads III and aVF and præcordial leads half-standard.

He was undersized, and there was slight central cyanosis and clubbing: signs of early heart failure disappeared with treatment. The heart was moderately enlarged, with apical and parasternal thrust. A loud ejection systolic murmur with a thrill was maximal at the pulmonary area, and there was a pansystolic and a short atrial systolic murmur at the apex. The second sound was single on auscultation: a phonocardiogram (Fig. 17) showed a late soft pulmonary component at the lower left border of the sternum, but not at the

aortic or pulmonary areas. (Cardiogram; see Fig. 4 and Table I.) The chest radiograph (Fig. 9) showed a prominent bulge on the left border of the heart below the usual position of the pulmonary artery, and a smooth unbroken convex contour above this.

He was catheterized twice (Table II). The findings supported a diagnosis of single ventricle with pulmonary valve stenosis. The catheter was passed with difficulty into the abnormally medial pulmonary trunk (Fig. 13) but neither main branch was entered. The left atrium was entered both through an interatrial communication, and from the single ventricle. The oxygen saturation of samples from the ventricle varied from 70 to 89 per cent, and brachial and pulmonary arterial samples were both 83 per cent saturated.

Angiocardiography (right atrial injection). Dye was seen to pass through an interatrial communication into the left atrium and there was a large common ventricle. The pulmonary trunk was long and narrow and abnormally medial and the pulmonary valve abnormally low: the aorta arose from the upper left part of the ventricle, probably from a separate small outflow chamber. In the lateral view, the aortic root was anterior.

A diagnosis was made of single ventricle, with transposition of the great vessels and bulbo-ventricular inversion, pulmonary valve stenosis, and atrial septal defect. Surgical treatment was not advised.

Comment. The patient with single ventricle resembled Case 7 in being type II A 1 (Harley, 1958), but in addition had pulmonary stenosis. The radiological appearances led to a confident diagnosis of corrected transposition and the electrocardiogram was consistent with this: the abnormal position of the great vessels was confirmed by catheterization and angiocardiography. Though the transposition was anatomically corrected, functional correction was prevented by the presence of a single ventricle. The pulmonary flow was relatively low as a result of pulmonary stenosis and the arterial oxygen saturation was therefore low.

Case 9. This girl, aged 15, had been found to have a cardiac murmur at 5 years. Though advised to limit her activities, she had no disability.

There was no cardiac enlargement, but the right ventricle was hyperdynamic. There was a loud early systolic ejection click, an accentuated palpable single second sound, a soft systolic murmur, and a long moderately loud diminuendo blowing diastolic murmur, best heard at the pulmonary area. (Cardiogram; see Fig. 5, Table I.) A chest radiograph (Fig. 10) showed a prominence of the upper left border of the heart, but narrow upper mediastinal shadow.

FIG. 5.—The electrocardiogram of Case 9.

At cardiac catheterization (Table II) considerable difficulty was experienced in entering the pulmonary trunk. An antero-posterior radiograph showed the catheter to be abnormally medial in the right-sided ventricle and pulmonary trunk (Fig. 14), and on withdrawal the pulmonary valve was noted to be abnormally low. No other abnormality was found.

Angiocardiography (injection into the right-sided ventricle). One early lateral film showed a puff of dye passing into the posterior (left-sided) ventricle. In the antero-posterior view (Fig. 15A) the right-sided ventricle was triangular with a smooth outline and a tail directed towards the cardiac apex, and the medial position of the pulmonary trunk was confirmed. Later films (Fig. 15B) showed the aorta arising from the left-sided ventricle on the upper left border of the heart. The aortic valve was higher than the pulmonary valve and slightly anterior to it (Fig. 16).

A diagnosis of a very small VSD, aortic or pulmonary regurgitation, and corrected transposition was made. No treatment was advised.

Comment. The radiographic and cardiographic appearances and the signs of pulmonary hypertension in the absence of cardiographic right ventricular hypertrophy led to the suspicion of corrected transposition. This was confirmed by cardiac catheterization and angiocardiography. There was no evidence of left A-V valve abnormality.

Case 10. A man, aged 27, had been known to have a murmur since infancy. He had been subject to winter bronchitis and he was limited by dyspnoea and fatigue.

There was moderate cardiac enlargement and an apical thrust and parasternal heave suggested hypertrophy of both ventricles. There was a moderately loud systolic murmur maximal at the lower left border of the sternum; the second sound was closely split and accentuated and there was a short soft pulmonary early diastolic murmur; and at the apex there was a third sound and moderately loud mid-diastolic murmur, following the P wave. The cardiogram (see Table I) showed partial heart block with periods of complete A-V dissociation, ending with a ventricular capture beat. The chest radiograph (Fig. 11) showed pulmonary plethora; the left upper mediastinal shadow was formed by a smooth straight line sloping upwards and medially, unbroken by the pulmonary trunk or by its left main branch; and there was also a slight prominence on the left upper border of the heart. *Cardiac catheterization* showed a large left-to-right shunt at ventricular level; the pulmonary arterial pressure was at systemic level (Table II).

At operation (Mr. I. M. Hill), the pulmonary trunk was twice the diameter of the aorta, and though its surface lay anterior to that of the aorta, the centre of origin of the pulmonary trunk lay posterior and to the right of the centre of origin of the aorta: the great vessels ran upwards and to the right, remaining parallel to one another. The left-sided ventricle had an infundibulum resembling that of the normal right ventricle in the pattern of the external muscle fibres and superficial coronary arterial branches. The right-sided coronary artery divided immediately into two, giving off a descending interventricular branch, and continued in the right A-V groove sending a large branch to the anterior aspect of the right-sided ventricle. The left-sided coronary artery gave rise to a left marginal branch and continued in the posterior A-V groove. The left atrium was normal and there was no systolic thrill over it. The interior of the heart was explored through the right atrial appendage. The right A-V valve was bicuspid, and the right-sided ventricle lacked a crista supraventricularis. A crista could be felt through the VSD separating the inflow and outflow portions of the left-sided ventricle, but the anatomy of the left A-V valve could not be determined. It was decided not to attempt to close the VSD. He survived the operation.

Comment. This man had corrected transposition with a VSD. There was no evidence of any left A-V valve incompetence. There was partial heart block and the appearance of the left upper mediastinum was very suggestive of corrected transposition. Though the presence of the anomaly had been suspected, the course of the catheter was not recognized as abnormal.

THE DIAGNOSIS OF CORRECTED TRANSPOSITION

Clinical Examination. Symptoms depend exclusively, and signs largely, on the presence of associated abnormalities. There are, however, certain relatively unobtrusive features that may point to the presence of the anomaly either in its pure form or as part of a more complex syndrome.

The pulmonary valve is situated further to the right, more caudally, and further from the anterior chest wall than normally: as a result the pulmonary component of the second sound may be inaudible, or audible only at the conventional aortic area (Gasul, Graettinger, and Bucheleres, 1959) or at the lower left sternal border as in Case 8 (Fig. 17). This was not a helpful sign in this series: when the sound of pulmonary valve closure is accentuated, splitting of the second sound may be widely heard; and when it is normal or diminished, it may be inaudible in all areas. Similarly, the systolic murmur of pulmonary stenosis or ASD may be lower down, as in pure infundibular stenosis, or to the right of the sternum when it may simulate aortic stenosis, and may be less intense than usual (Anderson, Lillehei, and Lester, 1957; Kjellberg *et al.*, 1959; and Schiebler *et al.*, 1961).

The aortic valve on the other hand is more superficially placed than in the normal heart and is situated further to the left. As a result, the single second sound may be louder in the pulmonary area than in the aortic area. This has led to the erroneous suspicion of pulmonary hypertension (Schiebler *et al.*, 1961; Goodman and Kuzman, 1961; and Badawi *et al.*, 1961); in Case 9, an early

systolic ejection sound, loud single second sound, and blowing diminuendo diastolic murmur combined to give a very convincing impression of pulmonary hypertension.

Congenital mitral regurgitation is rare except in corrected transposition. Other cases have been described by Edwards and Burchell (1958), Kjellberg *et al.* (1959), Malers *et al.* (1960), Talner, Stern, and Sloan (1961), and Proudfit and McCormack (1961) and others, and when mitral regurgitation is first recognized in an adult or older child, a rheumatic aetiology is usually presumed. However, when the physical signs of mitral regurgitation are known to have been present from an early age or when, in addition to the features of some other congenital heart lesion, there is also an apical pansystolic murmur, corrected transposition may reasonably be suspected. This was so in Cases 2, 3, 5, and 8 in this series, and in Case 1, with a VSD, the pansystolic murmur was louder even at the apex than at the usual Roger area.

Heart block may be recognized clinically and then be a valuable clue to the diagnosis of corrected transposition. The association of heart block with signs of mitral regurgitation may lead to the suspicion of an endocardial cushion defect (as in Case 2), particularly if the absence of wide fixed splitting of the second sound can be attributed to pulmonary hypertension. In Cases 1 and 10, as a result of a prolonged P-R interval, atrial systole coincided with the onset of ventricular filling and was responsible for an unusually intense mid-diastolic apical flow murmur.

Sex Incidence. Cardell (1956) and Malers *et al.* (1960) found an equal sex incidence in their reviews of reported patients, and Lester *et al.* (1960) reported 18 female and 22 male patients in their series. However, Schiebler *et al.* (1961) reported 21 male and 12 female patients and in the present series 9 out of the 10 patients were male.

THE ELECTROCARDIOGRAM

Though no cardiographic pattern is diagnostic of corrected transposition and no feature occurs in all patients with this anomaly, attention has been drawn to several unusual characteristics of the cardiogram that may be seen: all of these have been present in some of the patients in this series (Table I).

Heart block of all degrees is common. Walmsley (1931) and Abbott (1936) reported complete heart block and subsequently this has been observed by Anderson *et al.* (1957), Schaefer and Rudolph (1957), Fink *et al.* (1958), Walker *et al.* (1958), Kjellberg *et al.* (1959), Malers *et al.* (1960), Badawi *et al.* (1961), and Lev and Rowlett (1961), and in 6 out of 29 patients in the Mayo Clinic series (Schiebler *et al.*, 1961). 2:1 A-V block has been reported by Cardell (1956) and by Schiebler *et al.* (1961) in 4 patients. Lengthening of the P-R interval beyond the normal for age and rate is even more common: this occurred in 11 of Schiebler's patients (1961), in 10 of the 17 reported by Anderson *et al.* (1957), and in patients reported by Gasul *et al.* (1959), Kjellberg *et al.* (1959), Badawi *et al.* (1961), and Beck *et al.* (1961). Walker *et al.* (1958) had a patient with partial heart block and intermittent dropped beats. The term complete A-V dissociation is sometimes used loosely to include complete heart block but this arrhythmia in the more limited sense of the term was present in two patients (Anderson *et al.*, 1957). Different degrees of heart block may be present at different times in the same patient and the development of complete heart block during cardiac catheterization was reported by Schiebler *et al.* (1961), and occurred in our Case 1.

Atrial fibrillation was present in adult patients with "mitral" regurgitation reported by Schaefer and Rudolph (1957) and Malers *et al.* (1960), both of whom had complete heart block, and in two adult patients without "mitral" regurgitation by Schiebler *et al.* (1961). The Wolff-Parkinson-White syndrome with paroxysmal supraventricular tachycardia has been reported once by the last authors in a patient with a complete Ebstein anomaly of the left A-V valve: this is interesting in that the W-P-W pattern is often seen in the usual right-sided type of Ebstein anomaly. In the present series, complete heart block was present in two patients (one of whom developed atrial fibrillation) and a prolonged P-R interval in three others: intermittent A-V dissociation occurred in one of the latter. Atrial fibrillation was seen in two patients, one of whom had been previously observed in sinus rhythm: both were adults with "mitral" regurgitation. Though all degrees of heart block are

TABLE I
THE ELECTROCARDIOGRAM IN CORRECTED TRANSPOSITION

Case no., age, and sex	Associated lesions	Rhythm	P-R interval (sec.)	P wave Height (mm.)	Duration (sec.)	QRS duration (sec.)	Mean QRS axis	Lead III	Lead aVF	Lead V1	Lead V6	Other features
1 3 M	VSD, ? MR	Sinus but C.H.B. during cathet. (A 100, V62) Atrial fibr.	0.22	5	0.10	0.06	LAD	r1, S16, r'0.5	r5, S15	r3, S10	R19, S16	Large biphasic RS in V3-V5
2 19 M	MR, Partial L-sided Ebstein	C.H.B. (V43) (before, sinus, C.H.B. A90, V42)	—	—	—	0.11	LAD	Q(r)S11	Q(r)S9	QS16	R13, s3	Deep S V2-4
3 19 M	MR	Atrial fibrillation	—	—	—	0.14	LAD	q2, r2, S15 (notched) R8	q1, r2, S10 (notched) R10	q3(r), S30 (V2: QS48) QS11	R32, S18 (notched) r3.5, S12	Splintered QRS in V4 (rsr'sr's'') Deep S V3-5
4 21 M	VSD, PS, ? MR	Sinus, C.H.B. (A92, V60)	—	6.5	0.10	0.10	Normal	q1, R24	R21	r3, S8 (V3R: r3, s1)	r3, S8	Deep S V2-4
5 9 M	VSD, PS	Sinus	0.12	3	0.07	0.08	RAD	q1, R24	R21	r3, S8 (V3R: r3, s1)	r3, S8	Deep S V2-4
6 13 M	VSD, PS	Sinus	0.15	2.5	0.11	0.07	Normal	r2, S4, R5 q8, R40	R5 (notched) q3, R38	r4, S26	R22, S9	Large biphasic RS V2-5
7 13 M	Single ventricle	Sinus	0.22	4.5	0.12	0.10	RAD	q4, R46	q3, R38	R10, s3	R15, s3	Large biphasic RS V2-5
8 6 M	Single ventricle	Sinus	0.15	2	0.09	0.10	RAD	Q(r)S9	Q3, r1, S5	R16, S10 (V3R: q2, R16)	R26, s3	Deep S V2-4
9 15 F	PS, ASD Very small VSD, AR or PR	Sinus	0.14	1.5	0.08	0.08	LAD	q9, R33	q2, R27	q(r)S33 (V2: r0.5, S5, r3 (or Qr) (V3R: q5, R15)	R10, s3	Deep S V2-4
10 27 M	VSD	Sinus*	0.26	3	0.12	0.11	Slight RAD	q9, R33	q2, R27	q(r)S33 (V2: r0.5, S5, r3 (or Qr) (V3R: q5, R15)	R38, S48 (V7: q6, R54)	Deep S V2-6

* Intermittent A-V dissociation with ventricular capture beats (A75; V77).

common and Walker *et al.* (1958) suggested that the combination of VSD, heart block, and corrected transposition is a recognizable triad, many patients have had sinus rhythm with normal P-R interval: Schiebler *et al.* (1961) had 8 such patients out of 28, Anderson *et al.* (1957) 4 out of 17, and there were 4 in the present series.

Abnormal P waves indicating right- or left- or bi-atrial hypertrophy have been frequently noted. P waves, 0.3 mV or more in amplitude in lead II, were recorded in 6 patients in the present series, including one who later fibrillated: in Cases 1, 7, and 10, the appearance suggested left- or bi-atrial enlargement and in the others (Cases 2, 4, and 5) right atrial enlargement. However, as abnormal P waves are commonplace in congenital heart disease, their appearance, however striking, in patients with corrected transposition is of little help in diagnosis.

Anderson *et al.* (1957) were the first to suggest that reversal of the typical right and left ventricular QRS patterns in the præcordial leads was a characteristic feature of corrected transposition: when this occurs, right præcordial leads show a qR or qRs pattern and left præcordial leads an rS or RS pattern. In Anderson *et al.*'s series, 9 out of 17 patients had qR or qRs in V1 and two a monophasic Q or Qr. In the Mayo Clinic series (Schiebler *et al.*, 1961) 8 had qR (or qRs, qRS, qrS), 2 a monophasic Q (QS), and 6 others a deep Q (QR, QRs, Qrsr' or Qr). Of Kjellberg *et al.*'s (1959) 4 patients, one had a qRS and another a QS in V1. Three of 4 patients of Fink *et al.* (1958) had a qR and one a monophasic Q in V1. Morgan *et al.* (1962) found a q in V1 to be a valuable sign of corrected transposition in patients with a VSD. On the other hand 18 patients in the series of Anderson and Schiebler did not show a q wave in V1; none of the 5 patients reported by Beck *et al.* (1961) showed this pattern; nor did patients of Kernen (1958) or Goodman and Kuzman (1961). A qR pattern may, however, be seen in V4R but not in V1 (Anderson *et al.*, 1957). Two patients in this series (Cases 8 and 10) had a qR pattern in a right præcordial lead (V3R or V4R) but not in V1; four others (Cases 2, 3, 4, and 9) had QS waves in one or more right præcordial leads, with a tiny embryonic r. A qR pattern could be attributed to hypertrophy of the right-sided ventricle, but in the absence of corrected transposition this is a relatively uncommon finding even with considerable right ventricular hypertrophy, and it has been very reasonably suggested that the qR and similar patterns are associated with inversion of the right and left branches of the bundle of His (as has been demonstrated by Walmsley, 1931), so that the septum is activated from the right side instead of the left. The QS and Qr patterns on the other hand may occur in the absence of a raised pressure in the right-sided ventricle as in the series of Schiebler *et al.* (1961) and Anderson *et al.* (1957) and in Case 9 of the present series. Kjellberg *et al.* (1959) suggested that because of an abnormal plane of the right A-V valve, cavity potentials may be recorded in V1; but it is noteworthy that neither in their patients with this pattern, in Case 9 of the present series, nor in cardiograms illustrated by other authors was P inverted in V1.

The absence of q from the left præcordial leads is a much less valuable sign. Though a q was present in V6 in only one of Anderson *et al.*'s (1957) patients and two of Schiebler *et al.*'s (1961) and in V7 in one of the present series, the absence of q in left ventricular leads is by no means infrequent in the absence of corrected transposition, and in one series (Beck *et al.*, 1961) a small q was present in a left præcordial lead in 3 out of 5 patients. This sign obviously gains in significance when associated with a q in V1. Azevedo *et al.* (1958) reported a patient with a VSD and pulmonary hypertension in whom there was a curious splintering of the initial portion of QRS (rsr'S') in V5-6, and they suggested that this pattern might be helpful in diagnosis. Case 3 in the present series, with "mitral" regurgitation, also showed this feature, particularly in V4: both patients had also considerable left axis deviation.

Fink *et al.* (1958) drew attention to prominent Q waves in leads III and aVF in their 4 patients. This was also observed by Badawi *et al.* (1961, Case 1). Schiebler *et al.* (1961) have reported qR, qRS, or QS in lead III in 22 of their 28 patients. A qR pattern commonly occurs in these leads in a vertical heart as a reflection of left ventricular potentials. It seems possible that in corrected transposition, lead aVF may depict events in the right-sided ventricle when this is relatively posterior. In the present series, one patient (Case 10) had deep q waves in III and aVF, and two (Cases 2 and 9)

showed a deep notched Q or QrS in leads III and aVF: in the former, however, at necropsy the body of the right-sided ventricle was relatively anterior, and the explanation and significance of the presence of deep Q waves in the "posterior leads" remains obscure.

Gibbons *et al.* (1956) found a deep S in the right and mid-præcordial leads in three patients with corrected transposition and pulmonary stenosis, and suggested that this was due to hypertrophy of the posterior right-sided ventricle. Kjellberg *et al.* (1959) attempted to correlate the præcordial QRS pattern with the degree of rotation of the interventricular septum, pointing out that this was very variable. In the present series, such attempts were unrewarding: in Case 2, deep S waves over the right præcordium were probably related to dominance of the left-sided ventricle; and in Case 4 with pulmonary stenosis and VSD, rS complexes extended from V1-6, but though the right-sided ventricle was probably hypertrophied, angiocardiology showed the septum to be in the sagittal plane. Furthermore, dominant S waves extending over most or all the recorded præcordial leads are not uncommon in other situations and it is hard to accept Gibbons' claim that this pattern is really suggestive of corrected transposition.

RADIOGRAPHIC FEATURES

The cardiac contour, like the symptoms and signs, is largely dependent on the presence of associated lesions. The anomalous position of the great vessels does, however, result in unusual appearances of the upper mediastinum, which may be more or less suggestive of corrected transposition. The exact configuration of that part of the cardiac silhouette normally occupied by the aortic knuckle and by the pulmonary trunk and left pulmonary artery depends on the size of the aorta and of the pulmonary trunk, on how far to the left the aorta arises, dependent itself on the degree of general cardiac enlargement and on the extent of cardiac rotation, and on the direction of the parallel great vessels as they course upwards.

The aortic arch lies more nearly in the antero-posterior plane than normal and as a result the aortic knuckle is never conspicuous and is usually not seen in the postero-anterior chest film. This

FIG. 6.—Chest radiograph: Case 1.

FIG. 7.—Chest radiograph: Case 4.

FIG. 8.—Chest radiograph: Case 6.

FIG. 9.—Chest radiograph: Case 8.

feature, with the medial position of the pulmonary trunk, sometimes even when this is enlarged as a result of high pulmonary flow or pressure, may make the vascular pedicle appear abnormally narrow as in Cases 1, 3, and 9 (Fig. 6 and 10), and in patients reported by Becu *et al.* (1955), Anderson *et al.* (1957, Fig. 9), Lester *et al.* (1960, Fig. 5A), and Beck *et al.* (1961, Fig. 3A). When the aorta is large and arises well to the left, its ascending portion characteristically produces a smooth slightly convex bulge in the situation of the normal pulmonary artery; this is well seen in Case 8 (Fig. 9) and in illustrations in articles by Anderson *et al.* (1957, Fig. 7), Thilenius *et al.* (1959, Fig. 5), Lester *et al.* (1960, Fig. 3A and 4A), Beck *et al.* (1961, Fig. 3B), and Schiebler *et al.* (1961, Fig. 6A). In other patients, the lateral border of the ascending aorta forms a smooth straight line sloping upwards and medially, producing a gently tapering outline of the upper left portion of the cardiac shadow: this

FIG. 10.—Chest radiograph: Case 9.

FIG. 11.—Chest radiograph: Case 10.

contour is unbroken by the left pulmonary artery which lies medial to it. This appearance is well illustrated by Cases 6 and 10 (Fig. 8 and 11) and by patients reported by Anderson *et al.* (1957, Fig. 8), Walker *et al.* (1958, Fig. 5), Gasul *et al.* (1959), Lester *et al.* (1960, Fig. 6A), and Talner *et al.* (1961, Fig. 3). In other patients (e.g. Case 2), the appearance is not obviously unusual; and this is particularly the case when the contour of the ascending aorta is interrupted by the left pulmonary artery and its main branches. The position of the catheter in the aorta or pulmonary trunk (Fig. 12, 13, and 14) may also be useful.

FIG. 12.—Case 5. Cardiac catheter in the aorta and right subclavian artery, illustrating abnormal position of ascending aorta, on left of upper mediastinum (P.A. view).

FIG. 13.—Case 8. Cardiac catheter in pulmonary trunk, showing its abnormally low and medial situation (P.A. view).

Left atrial enlargement occurred in almost half of the patients reported by Schiebler *et al.* (1961), as a result of left A-V valve regurgitation or Ebstein anomaly; and it may be considerable (Malers *et al.*, 1960). In only two patients in this series (Cases 2 and 3) was there more than slight left atrial enlargement. This may result in a prominence of the left atrium on the left border of the heart as well as a double contour on the right. A prominence in the same situation continuous with the ascending aorta may also be seen in patients with a left-sided infundibulum; this was well seen in the patients with a single ventricle (Cases 7 and 8) and in Case 10, and in others a similar appearance may have been due to this cause (Fig. 7, 9, 10, 11).

Pulmonary plethora is to be expected when there is an associated left-to-right shunt, but pulmonary venous congestion and Kerley lines (as in Case 2) may point to the presence of pulmonary venous hypertension and so lead to the suspicion of a left A-V valve lesion. Anderson *et al.* (1957), Lester *et al.* (1960), Schiebler *et al.* (1961), and Goodman and Kuzman (1961) have observed indentation of the barium-filled œsophagus by the medially-placed pulmonary trunk.

CARDIAC CATHETERIZATION

Routine right heart catheterization (Table II), including dye dilution techniques, gives information about the associated lesions but can also be of great value in suggesting or confirming the diagnosis of corrected transposition.

TABLE II
OBSERVATIONS AT CARDIAC CATHETERIZATION

Case No.	Oxygen saturation percentage						Pressure (mm. Hg)†				Systemic artery	PA	Shunt	PVR units	Catheter course
	SVC	PV	RA	LA	R-sided V	L-sided V	PA	LA	PA wedge	R-sided V	L-sided V				
1	81	—	82	—	89	—	94	—	12/7	51/8	—	51/23	Large L to R	Low	Difficulty in entering P.T. course not noted to be abnormal
2	62	—	59	—	66	—	67	—	—	78/6	—	82/42	Doubtful L to R	8	Course not noted to be abnormal
4	—	—	72	—	89	—	85	—	—	110/0	—	23/8	L to R (2:1)	1	Catheter in aorta to left of P.T.
5 (1957)	64	94	62	94	62, 64	76	—	(5)	—	93/3	95/-1	(8)	R. to L.	Low	RA→R-sided V→L-sided V→LA→PV
5 (1960)*†(1)	67	—	65	90	65, 71	81, 84 (near LA), 82, 83 (near aorta)	—	(10)	—	94/-1	98/5	—	R to L.	—	As before, but PV not entered; catheter in ascending aorta on left of upper mediastinum
7	68	—	63	—	Single ventricle 95	—	94	—	—	Single ventricle 70/2	—	72/43	Large L. to R	2-3	Course not noted to be abnormal
8 (1957)	58	—	63	93	Single ventricle 79, 86, 94, 95 (right)	—	83	(3)	—	Single ventricle 85/0	—	30/10	Bidirectional	—	Catheter in P.T. abnormally medial. RA→single ventricle→LA
8 (1961)*†(2)	65	—	64	—	70 (near R.A.-V valve), 78, 83 (intermediate)	—	83	(2)	—	120/6	—	22/10	Bidirectional (pulmonary/systemic flow ratio 1.4:1)	2	Catheter in pulmonary trunk abnormally medial (Fig. 13)
9	63	—	68	—	66	—	65	(3)	(7)	21/4	—	20/7	—	1	Catheter in pulmonary trunk abnormally medial (Fig. 14)
10 (1958)*†(3)	65	—	68	—	89	—	85	(0)	—	74/0	—	70/39	L. to R. (3.4:1)	3	Course not noted to be abnormal
10 (1961)	71	—	70	—	87	—	83	(0)	(12)	110/8	—	118/43	L. to R. (2.3:1)	4	Course not noted to be abnormal

*Dye dilution curves: (1) Injection into R-sided ventricle; R.-to-L. shunt. Injection into L.-sided ventricle: normal curve. (2) Injections into RA and single ventricle: complete mixing. Appearance time: 2 sec. (3) Injection into pulmonary trunk: L.-to-R. shunt.

† Reference zero for pressure measurements: Cases 1 to 7, mid-thorax; Cases 8 to 10, sternal angle.

Owing to the proximity of the pulmonary and right A-V valves, the cardiac catheter must make a particularly sharp bend upwards and medially to enter the pulmonary trunk. The difficulty experienced in passing the catheter into the pulmonary trunk has been reported by several authors: it was not possible to enter it in 8 of the 24 patients catheterized by Schiebler *et al.* (1961), in 4 of the 5 reported by Beck *et al.* (1961) and in 9 of the 17 reported by Anderson *et al.* (1957). In the present series, there was only 1 failure in 8 patients catheterized, but in 3 others (Cases 1, 8, and 9) considerable patience was required. Helmholz *et al.* (1956) and Anderson *et al.* (1957) remarked that it was necessary to advance the catheter in the right-sided ventricle with a sharp curve on the catheter and the tip directed upwards and medially: in two of the three patients mentioned (all personally catheterized) this was confirmed.

The course of the catheter in the right-sided ventricle and pulmonary trunk as seen on the x-ray screen is often obviously abnormal, owing to the medial position of the latter. The catheter in the right pulmonary artery may form a smaller loop than normal as a result of its abnormally medial position: though the catheter in the ventricle is gently convex to the left as in the normal heart, the increased distance from the upper left border of the cardiac silhouette is distinctive. This is well illustrated by Case 9 (Fig. 14A), and by figures published by Walker *et al.* (1958), Azevedo *et al.*

FIG. 14.—Case 9. Cardiac catheter in right pulmonary artery. (A) P.A. view, showing medial position of catheter, with unusually great separation between it and the upper left heart border. (B) Lateral view.

(1958, Fig. 3), Goodman and Kuzman (1961), and Badawi *et al.* (1961, Fig. 3). A similar catheter course may be seen when the right pulmonary artery is entered from the saphenous vein (Azevedo *et al.*, 1958, Fig. 6; Lester *et al.*, 1960, Fig. 10A; and Badawi *et al.*, 1961, Fig. 7). This appearance can, however, be misleading, and in a patient with severe aortic stenosis and considerable left ventricular hypertrophy, the catheter course was thought to be suggestive of corrected transposition; but a lateral film showed the catheter in pulmonary artery and right ventricular outflow to be normally anterior. A lateral radiograph is, therefore, advisable under these circumstances, though the degree of posterior displacement may be unimpressive, as in Case 9 (Fig. 14B). Unless a true lateral film is taken, slight obliquity may give a spurious impression of a posteriorly placed pulmonary trunk, as happened in a patient with severe pulmonary valve stenosis and in a patient reported by Beck *et al.* (1961).

The catheter appears to enter the left pulmonary artery as readily as the right in many cases, and

the course of the catheter is then even more distinctive, its curve in the right-sided ventricle often being *concave* to the left, sometimes strikingly so (Helmholz *et al.*, 1956; Anderson *et al.*, 1957, Fig. 16; and Schiebler *et al.*, 1961, Fig. 8C). A similar appearance may be seen when the catheter tip is in the pulmonary trunk (as in Case 8, Fig. 13), when it may be impossible to advance the catheter into either lung field. A wedge pressure may be unobtainable for this reason or on account of pulmonary hypertension with obstructive pulmonary vascular disease. Careful observation of the catheter tip and pressure record during withdrawal from the pulmonary trunk may show the pulmonary valve to be abnormally low, as was noted in Cases 8 and 9.

The medial position of the pulmonary trunk gains added significance when the catheter also enters the ascending aorta which is then seen to lie to the left of the pulmonary trunk. This may occur during right heart catheterization when a VSD is traversed (Cases 4 and 5): the catheter passes up on the left side of the upper mediastinum and either continues up into subclavian or carotid artery as in Case 5 (Fig. 12), or turns down into the descending aorta (Lester *et al.*, 1960, Fig. 10C). A similar course may be taken by a catheter traversing a persistent ductus or in persistent truncus arteriosus, but these conditions can usually be distinguished at catheterization, and in persistent truncus the catheter can only very rarely be passed into either pulmonary artery. Schiebler *et al.* (1961) demonstrated the relative positions of the great vessels by simultaneous right heart and retrograde aortic catheterization with antero-posterior and lateral radiographs: this was not done in any of the present series and though it is undoubtedly a useful procedure, the same information can be obtained by angiocardiology. The same relative position of the aorta and pulmonary trunk may be seen in a few patients with uncorrected transposition (Noonan *et al.*, 1960).

In two patients, the catheter appeared to pass from the right atrium in turn to right-sided ventricle, left-sided ventricle, and, retrogradely through the left A-V valve, to the left atrium (and in one case into a pulmonary vein). One of these patients (Case 8) was shown to have a single ventricle with both A-V valves opening into a common chamber, but in the other (Case 5) the evidence did not favour this interpretation. In corrected transposition, the proximity of the ventricular septal defect to the left A-V valve and its separation from the aortic valve by the crista supraventricularis makes it more likely that a catheter entering the left-sided ventricle will pass into the left atrium than into the aorta: it is tentatively suggested that when the catheter takes this course, this is likely to be due to single ventricle or to corrected transposition.

The presence of "mitral" regurgitation may be suspected from the contour of the wedge pressure record if this shows tall *v* waves with steep *y* descent, and usually a raised mean pressure (Helmholz *et al.*, 1956; Kjellberg *et al.*, 1959; Badawi *et al.*, 1961; Talner *et al.*, 1961; and Schiebler *et al.*, 1961). A diagnostic left atrial pressure record (*v*₆₁, *y*₂₂, mean 34) obtained by direct puncture in a patient with "mitral" regurgitation was reported by Malers *et al.* (1960). When no wedge pressure is obtainable, a high pulmonary arterial pressure in the absence of a demonstrable intracardiac shunt may be due to "mitral" regurgitation (as in Case 2). However, though features indicating mitral regurgitation in a patient with congenital heart disease are very suggestive of corrected transposition, they do not prove its presence; but when other clinical, cardiographic, radiological, or catheter findings are suggestive, the presence of mitral regurgitation is valuable supporting evidence.

Other points that may be helpful but have not been observed in the present series include an abnormally high location of the right A-V valve orifice (Schiebler *et al.*, 1961). At left heart and aortic catheterization, the aortic valve may be seen to be abnormally high and lateral and further from the left A-V valve than normal.

When a patient with cyanotic congenital heart disease is found to have transposition of the great vessels, it is necessary to distinguish between uncorrected and corrected transposition with an associated lesion impairing the functional correction. In the former, the aorta is usually anterior and to the right but not invariably so, whereas in the latter the aorta is anterior and to the left. Furthermore, in uncorrected transposition, the systemic arterial saturation is inevitably less than the pulmonary arterial saturation. When the pulmonary artery cannot be entered, the distinction can only be made by angiocardiology.

ANGIOCARDIOGRAPHY

This is the most certain method of establishing the diagnosis of corrected transposition and is of particular value when the pulmonary trunk is not entered at cardiac catheterization, and in cyanosed patients.

Selective angiocardiology is preferable to peripheral venous injection, both to minimize the effect of dilution by large intracardiac shunts and to avoid superimposition of the right atrial opacity on the right-sided ventricle and pulmonary trunk, particularly in the lateral view. Injection into the right-sided ventricle demonstrates the medial position of the pulmonary trunk and the low situation of the pulmonary valve. When there is a right-to-left shunt through a VSD (as in Cases 5 and 6), the aorta and left-sided ventricle are also opacified: the origin of the aorta is seen to be high on the left side of the heart, the aortic valve being higher than the pulmonary valve; and the septal defect may be outlined and the ventricular septum seen in profile when it lies in the sagittal plane. In the absence of a significant right-to-left shunt, the left-sided ventricle and aorta may be outlined in the later films.

When the left heart and aorta are not well seen following injection into the right-sided ventricle, the anatomy can be demonstrated by injection into the left-sided ventricle, either through a catheter passed through a VSD or through a patent foramen ovale and left A-V valve (Beck *et al.*, 1961, Cases 5 and 2), or through a catheter passed retrogradely from a peripheral artery (as in Case 4), or through a needle introduced percutaneously, as in Case 5 and in the patient of Malers *et al.* (1960). When the left-sided ventricle is relatively small, percutaneous puncture may be difficult and the retrograde method is therefore preferable. Regurgitation of dye into the left atrium and enlargement of the atrium may be seen when there is left A-V valve incompetence. The left-sided anatomy may also be shown by left atrial injection, when left atrial puncture has been done to obtain pressure records (Malers *et al.*, 1960).

FIG. 15.—Angiocardiograms in Case 9. Antero-posterior view. (A) Early film showing right-sided ventricle and pulmonary artery. (B) Later film showing left-sided ventricle and aorta.

The position of the aorta on the left and the pulmonary trunk on the right, in the antero-posterior view, is diagnostic of corrected transposition in the acyanotic patient (Fig. 15). The lateral view merely gives confirmation of the anterior situation of the aorta, though in some patients (e.g. Case 9, Fig. 16) the two great vessels lie almost side by side, the pulmonary valve being only slightly posterior.

The relative positions of the ventricles are somewhat variable (Kjellberg *et al.*, 1959), depending on the degree of rotation of the heart. According to Schiebler *et al.* (1961), the commonest arrangement is for the right-sided ventricle to be posterior, with the septum running from right anterior to left posterior. In Case 2, the right-sided ventricle was in normal (right anterior) relation to the left; when this is the situation, the right-sided ventricle often shows a tail-like projection directed towards

FIG. 16.—Angiocardiogram in Case 9. Lateral view. (A) Early film showing right-sided ventricle and pulmonary artery. (B) Later film showing left-sided ventricle and aorta.

the apex of the heart: the cavity of the ventricle is then roughly triangular, with the pulmonary and right A-V valves occupying the base of the triangle, there being no infundibular portion of the ventricle. This was well illustrated by Anderson *et al.* (1957), Gasul *et al.* (1959), Lester *et al.* (1960), Malers *et al.* (1960), and Badawi *et al.* (1961) and by Case 9 in the present series (Fig. 15). When the ventricles lie side by side with the septum in the sagittal plane, this characteristic contour is not seen: the septum may be seen in profile when both ventricles are opacified simultaneously (as in Cases 4, 5, and 6). In Case 8, with a single ventricle and pulmonary stenosis, a separate subaortic outflow chamber could be distinguished. Sometimes the left-sided ventricle may appear small and it may be difficult by angiocardiography alone to determine whether this is a true ventricle, into which left atrial blood drains through the left A-V valve, or merely an outflow chamber communicating only with a large common ventricle through a bulbo-ventricular orifice.

In addition to its unusual shape the right-sided ventricle is commonly said to show a smooth internal surface on angiocardiograms, "with minimal trabeculation" (Lester *et al.*, 1960) like a normal left ventricle; the left-sided ventricle may show increased trabeculation (Malers *et al.*, 1960) and an infundibulum. Though the trabeculae in the normal left ventricle and in the right-sided ventricle of corrected transposition are in fact more numerous than in the contralateral ventricle, they are finer and consequently present a less striking appearance on contrast radiography than the less numerous but thicker trabeculae of the normal right ventricle and left-sided ventricle of corrected transposition.

In complete uncorrected transposition the aorta is sometimes anterior and to the left of the pulmonary artery: in the cyanotic patient, therefore, the relative position of the great vessels does not distinguish this situation from corrected transposition complicated by a defect producing right-to-left shunting. However, the pulmonary artery fills mainly or entirely from the right-sided ventricle in

corrected transposition, and from the left-sided ventricle in uncorrected transposition. Thus in the former, the ventricles, their outflow tracts, and the great vessels lie side by side in the antero-posterior view, whereas in the latter the outflow tract of the right ventricle crosses in front of the left ventricle. This enables the differentiation of these two conditions to be made on the angiocardigram in those patients in whom both pulmonary and systemic arterial samples have not been obtained (e.g. Cases 5 and 6).

Retrograde aortography was advocated as a diagnostic procedure by Thilenius *et al.* (1959) as the inverted coronary arterial pattern may be recognizable and the position of the aortic root can be clearly identified without being obscured by shadows due to filling of other chambers. However, the intracardiac anatomy is not revealed and, certainly in cyanotic patients, angiocardiology is to be preferred.

SUMMARY AND CONCLUSIONS

Ten patients with corrected transposition have been reported. This was associated with a ventricular septal defect in six, and in three of them there was also pulmonary stenosis, in one pulmonary hypertension, and in one probable pulmonary regurgitation. Two had a single ventricle, one with and the other without pulmonary stenosis: the remaining two had "mitral" regurgitation, with a left-sided Ebstein type of anomaly in one. The clinical features and the results of special investigations in the present series and in previously reported cases have been reviewed in order to establish diagnostic criteria.

The association of any degree of heart block, particularly when this is complete, with features suggesting mitral regurgitation, or of either or both of these with a congenital heart lesion, particularly ventricular septal defect (with or without pulmonary stenosis), is very suggestive of corrected transposition. Clinical examination is not often helpful, but suspicion may be aroused when pulmonary systolic murmurs are maximal low down or to the right of the sternum, when the pulmonary component of the second sound is inaudible in the pulmonary area, but can be heard or recorded elsewhere (Fig. 17); or when there are auscultatory signs of pulmonary hypertension without evidence of right ventricular hypertrophy.

FIG. 17.—Case 8. Phonocardiogram. Upper tracing, low frequency record from apex; lower tracing, high frequency records: (on left) from second left intercostal space (PA); (in middle) from second right intercostal space (AA), and (on right) from fourth left intercostal space. The amplification was unchanged during the recording from these three positions.

The chest radiograph may provide valuable information if the ascending aorta gives a straight or convex smooth contour to the left upper portion of the heart shadow normally occupied by the

pulmonary artery, or if the arrangement of the great vessels is such that the vascular pedicle appears unusually narrow. In the electrocardiogram there may be, in addition to heart block, a Q wave in right chest leads (Qr, QR, or qR, with or without S, qrS, or QS) and an absence of q from the left chest leads; an abnormally prominent Q in leads III and aVF; or deep S waves in all the præcordial leads in patients with hypertrophy of the right-sided ventricle. All of these clinical, radiological, and cardiographic features may, however, occur in other circumstances; and may all be absent in some patients with corrected transposition. Their presence, however, should lead to a search for further evidence from cardiac catheterization and particularly from angiocardiology.

At cardiac catheterization, the pulmonary trunk is seen to be abnormally medial and it may be difficult to enter. When the tip is in the left pulmonary artery, the course of the catheter in the right-sided ventricle may even be concave to the left. The pulmonary valve is lower than normal. A lateral radiograph shows the catheter to be abnormally posterior. A catheter passed from the right-sided ventricle or retrogradely into the aorta shows that this is situated to the left of and anterior to the pulmonary trunk. Pulmonary arterial wedge or direct left atrial puncture records may provide evidence of "mitral" regurgitation.

Selective angiocardiology shows the parallel great vessels, the aorta being anterior and to the left: the aorta fills from the left-sided ventricle and the pulmonary trunk from the right-sided ventricle. It may be necessary to perform left ventricular angiocardiology, preferably through a catheter passed retrogradely through the aortic valve; and this may show the coronary artery inversion in addition to the abnormal position of the aorta.

The differentiation of corrected transposition with an associated cyanotic form of congenital heart disease from complete uncorrected transposition may be difficult. The oxygen saturation in a systemic artery is above that in the pulmonary artery, unless there is complete mixing of the pulmonary and systemic venous returns. If no pulmonary arterial sample is obtained, the angiogram should be diagnostic.

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