Transposition of the Aorta and Pulmonary Artery

By Maurice Campbell, M.D., F.R.C.P., and S. Suzman, M.R.C.P.

Transposition of the aorta and pulmonary artery is now more frequently diagnosed in life. It must be distinguished from Fallot’s tetralogy because it is not helped by the Blalock-Taussig operation: indeed, the blood flow to the lungs is increased instead of reduced. This picture of pleomorphic lungs can be recognized by radioscopy and indicates a left to right shunt; when combined with central cyanosis, complete or partial transposition is the most common cause. The general picture seen in 25 cases, in 6 of which the diagnosis was confirmed by cardiac catheterization and angiography, is described. Two cases where the features of transposition were obscured by the presence of pulmonary stenosis are included.

When cyanotic children with congenital heart disease are examined on the x-ray screen, most show a concave pulmonary bay and light lung fields without pulsation; this suggests a diminished blood flow to the lungs. Some, however, show dense shadows around the hilum of the lung and for varying distances out into the lung fields, sometimes with the right and left pulmonary arteries much enlarged and with striking pulsation in these and even in the smaller branches out to the periphery; this means that there is an increased blood flow to the lungs, a condition we have called pulmonary pleonemia in contrast to pulmonary oligemia when the flow is diminished. The shape of the pulmonary arc is most often slightly concave (fig. 1) but may be convex, sometimes greatly so, generally owing to dilatation of the pulmonary artery (fig. 2), though the very varied position of the aorta (fig. 1, right) also changes the shape of the base of the heart.

The deep central cyanosis indicates a large right to left shunt, and the pleomorphic lungs indicate a large left to right shunt: we think that this combination generally indicates transposition of the aorta and pulmonary artery, though the transposition may often be partial rather than complete.

Present Views

Abbott1 collected 94 reported cases of transposition and found that the foramen ovale was open in 79, the ductus arteriosus in 63, and the ventricular septum in 31 of these. When there was no ventricular septal defect the average age at death was measured in weeks; and even when there was, it was under four years.

Until recently the picture was almost entirely based on cases seen post mortem. According to Brown1 the cyanosis generally dates from birth and may be only moderate; a systolic murmur is often present but may be absent if the defect is large; the pulmonary second sound is often accentuated; and there is right ventricular hypertrophy.

Taussig2 has made this diagnosis more possible in life, and describes the following picture. The heart increases in size with unusual rapidity. The pedicle is narrow in the anteroposterior view. In some cases the shadow of the right atrium may increase and decrease rhythmically with reversal of the shunt. Taussig does not think murmurs and thrills of much value in the diagnosis. Like Abbott, she gives a very bad prognosis, most patients dying within 14 months.

We think this description is mainly applicable to transposition in infancy. In older children we have not seen these rhythm changes in the atrium. In spite of the gloomy prognosis generally given, many of these patients are in reasonable health in their teens. The prognosis depends on the size of the shunts between the right and left sides of the heart; if these are large enough for adequate mixing of the two circulations, so that the infants survive, there is no reason for rapid deterioration, and these
are the only patients who will be seen in later childhood. We think dense lung fields and pulmonation of the pulmonary arteries in patients with central cyanosis are more important and
more constant than the other radiologic features, and even by themselves enough to allow the diagnosis of transposition.

At first, in 1947, we thought these signs indicated a septal defect with the cyanosis caused by some additional factor, and gradually became more sure that this was transposition,\(^5\) so that when the first patients had cardiac catheterization in 1948 we expected this to be confirmed.

\(\text{Taussig}^{12}\). p. 137 has a footnote saying that she is familiar with this type of case, but has only seen one necropsy, when there was found to be transposition. In 1948 when one of us was in Baltimore, Dr. Helen Taussig had come to look on this picture as characteristic of transposition, and this is also our view.

Cardiac catheterization and angiocardiography have not been done in all these patients because we are not at present advising operation for transposition. Blalock has, however, published a series of papers showing the difficulties and the measure of success he has achieved.\(^2\) \(^3\) It is clear, therefore, that many cases of transposition are diagnosed at the Johns Hopkins Hospital but little more has been published about the diagnosis.* Taussig

\* Since the writing of this paper, Taussig (1950) has kindly shown us her communication to the Inter-

and Bing\(^4\) have, however, described one case where the aorta was completely transposed and the pulmonary artery arose mainly from the right ventricle but overrode a ventricular septal defect. There was some fullness of the pulmonary conus with increased hilar shadows and faint expansile pulsation of the pulmonary arteries. The picture resembled Eisenmenger’s complex except that the cyanosis dated from birth.

**FIG. 2. Teleradiograms from cases of transposition; center and right with convexity of the pulmonary are, probably due to dilatation of the pulmonary trunk. (Left, Case 5). The lung fields show extreme mottling but the main pulmonary arteries are hidden by the heart. Pulsation became more obvious during two years observation. (Center, Case 4). Moderate increase of the root shadows and more prominence of the pulmonary are. Although pulsation was easily seen out to the periphery, the hila were less dense than in some cases. (Right, Case 3). The right pulmonary artery is dilated and expansile pulsation was seen here and in the smaller branches out to the periphery. Cases 3 and 4 contrast with the cases showing concavity of the are in figure 1 so that there is some similarity in shape to Eisenmenger’s complex.**

**CLINICAL PICTURE IN OUR CASES OF TRANSPOSITION**

We have selected 25 cases with pleonemic lungs as examples of transposition from the national Paediatric Congress at Zürich in which she describes the main features of transposition as follows: It generally leads to progressive cardiac enlargement and death during infancy. Occasionally, a balance is established and it may then be compatible with life for a number of years. These children are greatly incapacitated and frequently squat when tired. They suffer from severe anoxia and develop early and severe polycythemia and clubbing. A systolic murmur and gallop rhythm are common. There is slight cardiac enlargement and absence of fullness of the pulmonary conus: the vascular markings extend nearly to the periphery of the lung fields; there are many discrete circular shadows in the hilar region due to the number of blood vessels viewed on end. As will be seen from our paper we are in close agreement with many of her observations.
first 400 patients with cyanotic congenital heart disease. Expansile pulsation of the pulmonary arteries has been the decisive criterion, though some have been included on the general appearance of the lung fields. Nine of the 25 were between 6 and 18 years of age and 16 were aged 5 years or less. Five of these children have died, all from among the younger 16; in the only one with necropsy, who died when 2 months old with congestive failure, there was complete transposition of the aorta and pulmonary artery. The others who died always had large hearts, the smallest car diothoracic ratio being 59 per cent. Seven other patients, aged 9 or more, have not been included as it seemed less certain whether they had Eisenmenger's complex or transposition. It is interesting that from this radiologic approach transposition and Eisenmenger's complex should prove somewhat difficult to distinguish, and that Holling and Zak from their experience of cardiac catheterization should conclude that the effect on the circulation of Eisenmenger's complex and of transposition is often the same.

Disability. Most of these patients can only walk 100 to 400 yards but some can walk a mile or more slowly. Many are not as incapacitated as might be expected, probably because only the most favorable cases survive. Our older patients have the smaller hearts which suggests that an absence of cardiac enlargement in early life is a good guide to the prognosis.

Cyanosis and Polycythemia. Cyanosis was as severe as in the average Fallot's tetralogy. It was noted even earlier; in one, with only partial transposition, from 7 months, in 5, from the second month, and in all the others, from birth. The hemoglobin percentages were in the same range as in Fallot's tetralogy: perhaps less often above 140, but in one of the children who died as high as 180 per cent.

Clubbing of the Fingers. This was generally severe, and was often well advanced even in children of 2 or 3 years. We have noticed that some very cyanosed children have great distension of the veins of the fingers, especially the middle and terminal phalanges, as well as the clubbing of the fingers. This is especially common in children with transposition and we think it is caused by the increased systemic blood flow in addition to the other factors that cause clubbing. Lovell\textsuperscript{12} has shown that the clubbed fingers of cyanotic congenital heart disease differ from the others by showing an increase of the volume of blood.

Squatting. Four of these children squatted habitually, and a few others frequently sat on the floor with legs crossed like a tailor. The incidence of squatting (1 in 5) was much lower than in Fallot's tetralogy (4 in 5).

Physical Signs. Most of the patients with transposition had a systolic murmur, usually maximal in the pulmonary area, but often heard at the apex and occasionally louder there. Eight, however, had no murmur. A systolic thrill was only noted in 4 cases and thus is much less common than in Fallot's tetralogy.

One-fourth of these children had a diastolic murmur—a rare occurrence with Fallot's tetralogy—generally maximal in the pulmonary area but often well heard toward the apex. In some it was constant and easy to hear. In others it was variable or a faint whiff was heard just after the second sound.

The most important physical sign is the quality of the pulmonary second sound. If the second sound is loud and booming, or obviously duplicated, or if pulsation is visible or still more if it is easily palpable, it suggests that large pulmonary arteries will be seen on screening and that there is a raised pressure in the pulmonary artery. This booming second sound may be enough by itself to exclude the diagnosis of Fallot's tetralogy, where it may be somewhat accentuated but never so much.

The patient must be examined sitting and lying and in different phases of respiration before deciding on the intensity of the second sound. In infants and very young children with transposition, the second sound may at first be passed as within normal limits of intensity.

Shape and Size of the Heart. The heart in transposition is nearly always enlarged and the range of cardiothoracic ratios is well above that found in Fallot's tetralogy (see fig. 3). In three it was 50, 51 or 53 but generally ranged from 55 to 67 and averaged 59 per cent. The x-ray appearance of the heart as a whole supported these figures.
The enlargement of the heart is mainly due to the right ventricle, though sometimes this is less obvious than usual in the left oblique view, because the anterior position of the aorta making the border one smooth continuous curve, almost in contact with the chest wall, may give a false impression that there is no enlargement of the right ventricle. Even allowing for this there sometimes seemed to be a little increase of the left ventricle as well. Two cases where the left ventricle was larger than the right, with left ventricular preponderance in the electrocardiogram, have been omitted.

None of these cases had the deep pulmonary bay of Fallot’s tetralogy. Two-thirds showed some concavity (figs. 1 and 6, left) and one-third moderate or great convexity in the pulmonary region (fig. 2). In some cases where the aorta is completely transposed, it may be responsible for the shadow that is ordinarily the pulmonary artery (fig. 1, top left) and pulsation of the aorta may be thought to be pulmonary. In the majority of cases there is some prominence on the left border of the heart below the pulmonary arc, indicating the infundibulum of the enlarged right ventricle (fig. 1, top left). The diameter of the heart is increased diagonally as well as transversely.

Electrocardiography. Often the P waves were large and pointed but sometimes they were not remarkable. On the whole, the electrocardiograms show no distinguishing features from those seen in Fallot’s tetralogy.

The Lung Fields. There are two main features: general mottling of the lung fields and dilatation of the pulmonary arteries. In most patients both features are evident (fig. 2, center; in some, one or another predominates. Sometimes it looks as if the pulmonary artery is represented by many branches that produce a special mottled appearance of the hilum extending for varying distances into the fields (fig. 1, bottom left). Sometimes enlargement of the right and left pulmonary arteries may be hidden by the large heart (fig. 2, left). An unusual degree of dilatation of the main pulmonary arteries is shown in figure 1, top left.

The mottled density throughout the lung fields is generally obvious on the film but sometimes is better judged on radioscopy and should always be confirmed in this way. Pulsation in the pulmonary arteries is one of the main criteria for the selection of patients, and in at least half of them it can be seen easily in small branches out to the periphery. There is often a hilar dance. In very young children these mottled shadows may be the first sign of the increased pulmonary circulation and it is only in the course of a year or two that the full picture emerges more clearly. Sometimes the picture is very characteristic as early as 24 or even 12 months.

When a diastolic murmur is easily heard and maximal in the pulmonary area or when it fol-

![Fig. 3. Chart showing the cardiothoracic ratio in these 25 cases of transposition of the aorta and pulmonary artery. (Each case being shown by a solid circle) compared with 50 cases of Fallot’s tetralogy that have been divided into four groups, enlarged, large normal, normal, and small normal. In general, the heart is larger (average ratio 59.4 per cent) than in Fallot’s tetralogy.

ows a loud booming second sound, it probably indicates pulmonary regurgitation. We thought there might be a close correlation between this and the hilar dance, but it was not so. Of 13 cases of transposition in which pulsation was seen well out into the lung fields, 5 had diastolic murmurs in the pulmonary area and 8 had no diastolic murmur. A diastolic murmur in the pulmonary area is not therefore a requisite for increased pulmonary artery pulsation, nor is the degree of pulsation dependent on the presence or absence of a pulmonary diastolic murmur. On the other hand, when such a murmur is present it is unusual not to have some increased pulsation in the lung fields. In cyanotic cases with pulsation in the lung fields, diastolic
murmurs have been heard in a much larger proportion of our cases.

Differential Diagnosis. Difficulty in diagnosis may arise when there is increased collateral circulation to the lungs because of pulmonary atresia or severe stenosis in Fallot's tetralogy. As a general rule collateral arteries are shown on x-ray films by an increase of streakiness or linear markings in the lung fields, or by small round spots when the vessels are seen in cross section. This contrasts with the denser and large patchy shadows, generally with clearly defined edges which show expansile pulsation or changing density on radioscopy; these are rarely pathognomonic.

The dilatation of the artery beyond a pulmonary valvular stenosis may cause some superficial resemblance to these cases, but then pulsation will be slight or absent and the branches of the pulmonary arteries will be small and the lung fields light and clear.

Some cases of Eisenmenger's complex may be confused with transposition and the differentiation may be difficult, especially when there is convexity of the pulmonary arc. The late onset of cyanosis is one point of distinction, and radiologically there is less dilatation of the smaller branches and pulsation and the mottled appearance of the lung fields are much less.

TABLE 1.—Oxygen Percentage Saturation Obtained by Catheterization in Cases of Transposition

<table>
<thead>
<tr>
<th>Case no.</th>
<th>O2 capacity (vols. %)</th>
<th>S. V. C.</th>
<th>I. V. C.</th>
<th>Right atrium</th>
<th>Right ventr.</th>
<th>Pulm art.</th>
<th>Systemic artery</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Aorta</td>
</tr>
<tr>
<td>1</td>
<td>29.5</td>
<td>39</td>
<td>41</td>
<td>41</td>
<td>50</td>
<td>—</td>
<td>53</td>
</tr>
<tr>
<td>2</td>
<td>26.7</td>
<td>57</td>
<td>—</td>
<td>61</td>
<td>62</td>
<td>—</td>
<td>71</td>
</tr>
<tr>
<td>3</td>
<td>24.8</td>
<td>53</td>
<td>58</td>
<td>50</td>
<td>61</td>
<td>84</td>
<td>—</td>
</tr>
<tr>
<td>4</td>
<td>25.3</td>
<td>60</td>
<td>—</td>
<td>54</td>
<td>75</td>
<td>82</td>
<td>—</td>
</tr>
<tr>
<td>5</td>
<td>24.2</td>
<td>60</td>
<td>65</td>
<td>64</td>
<td>84</td>
<td>85</td>
<td>—</td>
</tr>
<tr>
<td>6</td>
<td>20.9</td>
<td>52</td>
<td>63</td>
<td>65</td>
<td>79</td>
<td>—</td>
<td>—</td>
</tr>
</tbody>
</table>

The italic figures indicate the place where the left to right shunts become manifest, either through a ventricular septal defect or because the pulmonary artery is arising from the left ventricle. None of these patients showed any evidence of an atrial septal defect.

TABLE 2.—Intracardiac Pressures Found at Cardiac Catheterization in cases of Transposition

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Pressure mm. Hg*</th>
<th>Pulmonary blood flow L/min./sq.m.</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Right atrium</td>
<td>Right ventr.</td>
</tr>
<tr>
<td>1</td>
<td>7</td>
<td>55</td>
</tr>
<tr>
<td>2</td>
<td>12/0</td>
<td>89/16</td>
</tr>
<tr>
<td>3</td>
<td>5/0</td>
<td>93/0†</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>35</td>
</tr>
<tr>
<td>4</td>
<td>10</td>
<td>47</td>
</tr>
<tr>
<td>5</td>
<td>8</td>
<td>65/10</td>
</tr>
<tr>
<td>6</td>
<td>12/6</td>
<td>85/8</td>
</tr>
</tbody>
</table>

* Single figures are mean pressures and double figures systolic and diastolic pressures.
† The reading in the left ventricle was 100/0 with a mean pressure of 45.
‡ Direct reading in aorta: brachial artery pressures have been taken with sphygmomanometer.

really pathognomonic of increased pulmonary blood flow. Sometimes they may need looking for carefully, especially in infants.

The Findings on Cardiac Catheterization and Angiocardiography

Six patients had cardiac catheterization and 5 of these and 2 others had angiocardiography. In 3 the catheter entered the pulmonary artery, and in one of these the aorta also. All these 3 showed a much higher oxygen saturation in the pulmonary artery (84 per cent) than in the aorta and its branches (70 per cent), proving that, functionally at least, there was transposition. In the other 3 the pulmonary artery was not entered but the oxygen saturation of the aorta was even lower (65 per cent); in 2 of these the catheter entered the aorta only, but in the third no main vessel could be entered.

In all the 6 cases there was evidence of a large left to right shunt through a ventricular septal defect, but no evidence of an atrial septal defect. In all, the systemic pressure was rather low but there was a high pressure in the right ventricle and in the pulmonary artery in the 3
Angiocardiograms were of value in showing the aorta arising entirely or partly from the right ventricle in every case. In case 1 the aorta came entirely from the right ventricle in the position that is normally occupied by the pulmonary artery, and the pulmonary artery came mainly or entirely from the left ventricle. In some of the others, the transposition was less complete though the aorta arose mainly or partly from the right ventricle. They showed late fillings of the pulmonary arteries in spite of their large size and the large pulmonary flow. They provided some direct evidence that the pulmonary arteries filled mainly or entirely from the left ventricle, except perhaps in case 2 where the hilar shadows were suggestive of lungs filling from bronchial arteries (fig. 1, bottom left; ref. 7). Angiocardiograms did not always give as clear a picture as was hoped, but if there are shunts in both directions, complete clarity can only be expected in the very early stages after the injection of the Diodone. The arm to lung and arm to tongue circulation times were very close, giving further evidence of a right to left shunt.

### Table 3.—Functional Relationships of Aorta and Pulmonary Artery to the Two Ventricles in Transposition

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age when first seen &amp; sex</th>
<th>Clubbing</th>
<th>Squatting</th>
<th>Degree of cyanosis</th>
<th>Onset of cyanosis</th>
<th>Aortic arch</th>
<th>Pulmonary artery</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 (0052)</td>
<td>0 M</td>
<td>4</td>
<td>(S)*</td>
<td>4</td>
<td>4</td>
<td>Birth</td>
<td>Entirely from L. V.</td>
</tr>
<tr>
<td>2 (0145)</td>
<td>4 M</td>
<td>4</td>
<td>No</td>
<td>3</td>
<td>3</td>
<td>3 birth</td>
<td>Mainly from L. V.†</td>
</tr>
<tr>
<td>3 (0109)</td>
<td>11 M</td>
<td>4</td>
<td>No</td>
<td>3</td>
<td>2</td>
<td>Birth</td>
<td>Mainly (½ths) from L. V.</td>
</tr>
<tr>
<td>4 (0076)</td>
<td>13 M</td>
<td>4</td>
<td>S</td>
<td>2-3</td>
<td>3-4</td>
<td>7 mo.</td>
<td>Some from R. V.*</td>
</tr>
<tr>
<td>5 (0A06)</td>
<td>6 M</td>
<td>4</td>
<td>S</td>
<td>3-4</td>
<td>3-4</td>
<td>Birth</td>
<td>About half from R. V.*</td>
</tr>
<tr>
<td>6 (0175)</td>
<td>2 F</td>
<td>4</td>
<td>No</td>
<td>3</td>
<td>4</td>
<td>Birth</td>
<td>About half from R. V.*</td>
</tr>
</tbody>
</table>

* Indicates that the catheter entered this vessel from the right ventricle.
† Films suggest lungs are partly supplied by bronchial arteries.
‡ Catheterization suggests mainly from right ventricle but angiocardiography suggests more from left ventricle.

### Table 4.—Some Clinical Data with Heart Size and Blood Counts

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age when first seen &amp; sex</th>
<th>Clubbing</th>
<th>Squatting</th>
<th>Degree of cyanosis</th>
<th>Onset of cyanosis</th>
<th>Aortic arch</th>
<th>Blood count</th>
<th>Heart size</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>0 M</td>
<td>4</td>
<td>(S)*</td>
<td>4</td>
<td>4</td>
<td>Birth</td>
<td>148</td>
<td>50</td>
</tr>
<tr>
<td>2</td>
<td>4 M</td>
<td>4</td>
<td>No</td>
<td>3</td>
<td>3</td>
<td>Birth</td>
<td>130</td>
<td>59</td>
</tr>
<tr>
<td>3</td>
<td>11 M</td>
<td>4</td>
<td>No</td>
<td>3</td>
<td>2</td>
<td>Birth</td>
<td>122</td>
<td>56</td>
</tr>
<tr>
<td>4</td>
<td>13 M</td>
<td>4</td>
<td>S</td>
<td>2-3</td>
<td>3-4</td>
<td>7 mo.</td>
<td>130</td>
<td>51</td>
</tr>
<tr>
<td>5</td>
<td>6 M</td>
<td>4</td>
<td>S</td>
<td>3-4</td>
<td>3-4</td>
<td>Birth</td>
<td>116</td>
<td>62</td>
</tr>
<tr>
<td>6</td>
<td>11 M</td>
<td>4</td>
<td>(S)*</td>
<td>3-4</td>
<td>3-4</td>
<td>Birth</td>
<td>115</td>
<td>66</td>
</tr>
<tr>
<td>7</td>
<td>3 F</td>
<td>4</td>
<td>—</td>
<td>3</td>
<td>4</td>
<td>Birth</td>
<td>107</td>
<td>56</td>
</tr>
<tr>
<td>8</td>
<td>2 F</td>
<td>4</td>
<td>No</td>
<td>3</td>
<td>4</td>
<td>5 wks.</td>
<td>146</td>
<td>55</td>
</tr>
</tbody>
</table>

* No true squatting but sitting with legs crossed like a tailor.

Cases where it was measured (see tables 1, 2, 3, 4). The blood flow to the lungs was increased in these 3, and the systemic flow was thought to be increased in all, but not always greatly so. Holling and Zak[1] have referred shortly to 3 of these cases.

### Case 1 (0052). The heart showed some enlargement of the right ventricle only. There was a concave pulmonary bay and great dilatation of both pulmonary arteries (fig. 1, top left). These showed expansile pulsation right out to the periphery with a hilar dance.

On cardiac catheterization the right ventricle
was entered easily and the mean pressure (55 mm. Hg) was much increased. The aorta was entered easily from the right ventricle (fig. 1, top right) but the pulmonary artery could not be entered. There was a left to right shunt through a ventricular septal defect, the oxygen saturation rising steadily from 44 per cent low in the right ventricle to 53 per cent in the aorta.

The aorta therefore comes entirely from the right ventricle and arises exactly in the position normally occupied by the pulmonary artery (our fig. 4, and reference 8, fig. 18). A very similar case has been reported by Goodwin and co-workers. 10

Case 2 (0145). The heart was enlarged, especially the right ventricle, and the pedicle was narrow. The density of the lung fields was greatly increased, especially above the right hilum (fig. 1, bottom left), and expansile pulsation was easily seen, sometimes out to the periphery.

On cardiac catheterization the right ventricle and aorta were entered easily (fig. 1, bottom right). A moderate left to right shunt through a ventricular septal defect must have been high, as it had more effect on the oxygen saturation in the aorta than in the right ventricle. The pulmonary artery was not entered.

The aorta therefore rises mainly from the right ventricle but acquires some blood from the left ventricle direct or through a high ventricular septal defect, which is the site of the left to right shunt.

Angiocardiography showed the aortic arch well filled at two seconds and the subclavian already filling, which is unusually early. The pulmonary artery was not well seen and the lungs hardly started to fill until three seconds, probably from the left ventricle, though some filling by the bronchial arteries could not be excluded.

Case 3 (0109). The heart was moderately large with a convex pulmonary arc (fig. 2, right). Density of the lung fields was increased and there was expansile pulsation, sometimes out to the periphery.

The oxygen saturation of 70 per cent in the aorta and of 84 per cent in the pulmonary artery proved a functional transposition. The right ventricle, and from it the aorta, was entered easily. Later, the pulmonary artery was entered apparently after passage through a fairly large ventricular septal de-
prominence of the pulmonary arc (fig. 2, center). Pulmonary arteries and their branches were moderately dilated, with pulsation out to the periphery and a hilar dance.

Cardiac catheterization (September 15, 1948). The rise in oxygen saturation from right atrium to right ventricle and from right ventricle to pulmonary artery indicated a left to right shunt through a high ventricular septal defect, and a pulmonary artery overriding both ventricles. Between one-third and one-half of the blood in the pulmonary artery comes from the left ventricle.

![Image of angiocardiograms](http://circ.ahajournals.org/)

**Fig. 5.** Angiocardiograms, both at four seconds, from cases with transposition showing filling of the aorta from the right ventricle with very little filling of the lungs. (Left, Case 5). The right atrium and ventricle and the aorta and its branches are filled, but very little has passed to the left ventricle or to the pulmonary arteries. (Right, Case 4). At four seconds. The heart has filled fairly generally and the aorta and its branches are still seen as they had been from two seconds. The pulmonary arteries are only just starting to fill, probably from both ventricles, but the lungs show little increase in density.

The aorta was not entered, but the low systemic oxygen saturation showed that it comes mainly or entirely from the right ventricle and further away from the ventricular septal defect than the pulmonary artery, as it was less affected than the latter by the left to right shunt through the ventricular septal defect.

On angiocardiography the aortic arch fills at two seconds and the subclavian at three seconds. The aorta, therefore, comes off the right ventricle but not in the same position as in case 1, and may override the left ventricle somewhat. A high dilated pulmonary artery also shows up at two seconds and more easily at three seconds, and also the border of the right ventricle, showing that the pulmonary artery comes, partly at least, from the right ventricle. The poor filling of the pulmonary branches, although they are large and pulsatile, shows that their main supply is from the left, not from the right ventricle (fig. 5, right). The left ventricle is filled at four seconds so there is a septal defect. At 15 seconds the left heart is still opaque instead of being clear by 10 seconds, further evidence that the aorta does not rise entirely from the right ventricle. This case corresponds most closely to the one reported by Taussig and Bing.

Case 5 (OA06) The heart was large, especially the right ventricle; the pulmonary arc was concave and the pedicle broadened in the oblique view (fig. 2, left). The pulsation and the density of the lung fields increased during two years observation.

On cardiac catheterization the right ventricle and the left pulmonary artery were entered easily but the route seemed somewhat unusual. There was a large left to right shunt through a ventricular septal defect, the oxygen saturation rising from 64 to 84 per cent. The oxygen saturation in the pulmonary artery was close to that in the right ventricle; from this the pulmonary artery seemed to arise normally, but angiocardiography suggested that it overrides the left ventricle to a great extent (fig. 5A).

The blood in the systemic arteries was only 70 per cent saturated. As the blood in the right ventricle was 84 per cent saturated and that in the left ventricle must have been more than this (to ac-
count for the increasing oxygenation caused by the left to right shunt through the ventricular septal defect the aorta must have come entirely from a part of the right ventricle that was well away from the effects of the ventricular septal defect, while the catheter must have been much nearer it when the right ventricular figures were obtained. The only other possibility, that the catheter was in the left ventricle, seems unlikely.

Case 6 (0175). The heart was much enlarged; there was extreme dilatation of a main trunk in the normal position of the pulmonary artery. The lung fields seemed typical of transposition and pulsation was easily seen out to the periphery, with a hilar abundance. This case is not really helpful, as neither the aorta nor the pulmonary artery was entered on catheterization and there was thought to be a single ventricle. The angiocardiograms are also difficult to interpret.

Another child has recently had angiocardiograms done by Dr. C. G. Parsons; we would like to thank him for letting us see these and agree that they confirm our clinical diagnosis of transposition.

Two Cases Where the Diagnosis Was Made at Operation

The findings at operation in 2 other children who were wrongly diagnosed as Fallot's tetralogy supported the diagnosis of transposition. We are indebted to Mr. R. C. Brock for these observations.

Case 7 (H113). Doubt was felt about the density of the lungs, but they were accepted as consistent with a reduced blood flow and heavy lung markings from a collateral circulation. At operation, the pulmonary artery was large and pulsed vigorously. The pressure inside it was well above 85 cm. of saline (the manometer did not measure above this), and was probably the same as the systemic pressure, which was only 90 mm. Hg. The patient was then thought to have Eisenmenger's complex, but transposition seems more likely. On screening two years later, pulsation was easily seen in the pulmonary arteries and in some of the rounded patches in the dense lung fields.

Case 8 (P029). On x-ray examination there was a narrow pedicle; in retrospect, it seems obvious that the blood flow to the lungs was increased, but pulsation, which could be recognized later, was missed on screening the increased shadows were thought to be due to collateral circulation (fig. 6, left). On angiocardiography the right side of the heart and the aorta and its branches were well filled at 2 seconds without any change in the density of the lungs, which only started filling slowly at 3 seconds.

At operation the left pulmonary artery, about 1.5 cm. in diameter and with a high pressure, was larger than the aorta. On opening the pericardium the diagnosis of transposition was confirmed. The small aorta arose from the right ventricle and the large pulmonary trunk, fully 3 cm. in diameter, from the left ventricle. She made an uneventful recovery.
These two cases reinforce the points we have given in the differential diagnosis of increased density due to transposition and due to a collateral circulation. Unless the site of the stenosis can be demonstrated, angiocardiography does not always distinguish between lungs that are not filling because there is pulmonary stenosis, or because the pulmonary artery arising from the left ventricle cannot fill until later when the blood has reached this chamber.

Transposition with Pulmonary Infundibular Stenosis

We are adding 2 cases with necropsies where complete transposition of the aorta and partial or complete transposition of the pulmonary artery were complicated by infundibular stenosis. This last prevents the development of the changes generally found with transposition, so that there may be rather prominent lung shadows without increased pulsation. Both these

![Fig. 7. (Left). External view of the heart of case 9. The aorta arises from the right ventricle. The pulmonary trunk and pulmonary arteries with walls spread out are seen alongside, with a probe entering the narrow pulmonary valve orifice. A dilated coronary vessel, cut open down its length, and the interventricular groove can be seen. (Right). The cavity of the hypertrophied right ventricle (R.V.) and the large defect high up in the interventricular septum (B). The aorta is seen on the right with a large aortic valve sinus (A). To the left the cavity of the right atrium (R.A.) and the slit communicating with the left atrium are seen.](http://circ.ahajournals.org/).
cases were thought to have Fallot’s tetralogy, perhaps with some other complications. The first, who died after operation, had already reached the age of 24 years and is reported here. In the second (case 10), there was complete transposition, high grade infundibular stenosis affecting the outflow tract to the pulmonary artery, and almost a single ventricle. His case will be reported later with some other examples of single ventricles.

Case 9 (1088), aged 24, was cyanosed from birth, and could walk a quarter of a mile but no more. When 16, and again when 20, she had hemoptysis. She could only do easy housework but married and had one miscarriage.

Her cyanosis, clubbing, and polycythemia were extreme. There was a systolic murmur loudest below the pulmonary area, sometimes with a soft systolic thrill, and the pulmonary second sound was increased. There was no diastolic murmur. The blood pressure was 115/85. The electrocardiogram showed gross right ventricular preponderance with a large pointed P wave in lead II.

The heart was not generally enlarged (cardio-thoracic ratio 49), but only the right ventricle. On screening, the base shadows seemed increased with some mottling of the lungs (fig. 6, right); in the absence of pulsation this was thought to be due to collateral circulation. On catheterization the pulmonary artery was not entered; a pressure of 80 mm. Hg in the right ventricle and an arterial oxygen saturation of 65 per cent were thought to support the diagnosis of pulmonary stenosis and a large right to left shunt.

Operation was decided on after some hesitation because of the lung fields, since she was becoming more disabled. Various difficulties prevented completion of an anastomosis. She remained drowsy afterwards and was not seen to move her right arm; she deteriorated and died 24 hours after operation.


The heart was enlarged (400 Gm.) and rather quadrilateral in shape. The right ventricular muscle was hypertrophied, its thickness at the apex being 14 mm. compared with 11 mm. for the left ventricle. The right atrium was dilated, with a large coronary sinus and a slit-like defect in the anterior part of the septum measuring 5 by 2 mm. There were three small vegetations, partly calcified, on the tricuspid valve. The right ventricle communicated with the left through a large defect of the upper part of the septum, 30 mm. in diameter (fig. 7, right).

The pulmonary artery was normal but thin-walled, and its diameter was 10 mm. against 24 mm. for the aorta; it overrode the septal defect, slightly to the left of the aortic orifice. The infundibulum, 20 mm. below the pulmonary valve ring, measured only 5 mm. across and a projecting mass of endocardium increased the infundibular stenosis. There was partial fusion of two pulmonary valve cusps, but no stenosis. The dextroposed aorta (fig. 7, left) arose from the right ventricle.

The right coronary artery was normal. The left arose from the left anterior aortic cusp and was as large as a normal carotid artery, but soon divided into a normal left and an aberrant branch running downwards over the right ventricle to the right anterior branch of the left coronary artery. There were many arteries passing to the lung hilum, some apparently bronchial arteries.

The spleen was enlarged (400 Gm.). The diagnosis of thrombocytopenic purpura had been made in life. The other findings were not remarkable and no gross abnormality of the brain was found.

Sections showed hypertrophy of the muscle cells and much replacement fibrosis, diffusely spread through the whole muscle of the right ventricle. There was no hypertrophy but a few similar patches of fibrosis in the left ventricle.

Summary and Conclusions

Most cyanotic children with congenital heart disease have a diminished blood flow to the lungs (pulmonary oligemia). Some, however, show increased shadows around the hilum and for varying distances out into the lung fields, often with large right and left pulmonary arteries and with striking pulsation there and even in the small branches out to the periphery. These changes are somewhat like those seen in atrial septal defect in acyanotic cases and indicate an increased blood flow (pulmonary pleonemia).

In addition to the large left to right shunt there is an extra factor giving rise to a right to left shunt and to the cyanosis of these children; this extra factor is generally complete or partial transposition of the aorta and pulmonary artery, with a septal defect that is usually ventricular.

The prognosis is generally bad, the patient dying in the first few months. If, however, the septal defects are large enough for adequate mixing of the circulations of both sides of the heart, so that the child survives infancy, there is no reason why it should not survive much longer.

We thought 25 children with pleonemic lungs among the first 400 with cyanotic congenital
heart disease had complete or partial transposi-
tion of the aorta and pulmonary artery. The
selection was made on the appearance of the
lung fields on radiography and teleradiograms;
and expansile pulsation of the pulmonary
branches was the main criterion.

Six of our children had cardiac catheterization,
and 5 of the 6 and 2 others have had
angiocardiography. These results have been
described fully. In 3, the oxygen saturation of the
pulmonary artery was much higher than that
of the aorta, proving that, functionally at least,
there was transposition; and in the other 3 this
diagnosis was supported indirectly. Angiocar-
diography proved that the aorta arose from the
right ventricle in 2, and in one of these in the
position normally occupied by the pulmonary
artery. In the others the aorta probably over-
 rode both ventricles. In all these there was evi-
dence that the pulmonary artery arose—entirely
or mainly or at least half—from the left
ventricle. In one of these there may have been
a single ventricle. In two others the diagnosis
was made after the findings at operation.

In these cases of transposition, cyanosis is as
severe as in Fallot’s tetralogy, and much more
constantly has been noticed from birth. The
general appearance and the clubbing of the
fingers are similar but the hemoglobin is not so
often above 140 per cent. In addition, there is
often an unusual degree of dilatation of the
veins over the dorsal aspect of the fingers that
may be suggestive of the diagnosis of transposi-
tion. Only a few of these children squat habitually.

A systolic murmur, generally maximal on the
left side in the pulmonary area, is present in
two-thirds of these cases. A thrill may accom-
pany this, but much less often than with Fal-
lot’s tetralogy. A diastolic murmur, generally
maximal in the pulmonary area, is present in
one-fourth of these cases.

An important physical sign is the quality of
the pulmonary second sound, which is generally
loud and booming and may be palpable; this
alone may be enough to exclude Fallot’s tetral-
yogy. It suggests that large pulmonary arteries
will be seen on screening, probably with high
pulmonary pressure.

The heart is larger than in Fallot’s tetralogy,
the average cardiothoracic ratio being 59 per
cent, and is also of a different shape. The pul-
monary arc is typically concave but may be
convex, sometimes greatly so from dilatation
of the pulmonary trunk or from misplacement of
the aorta. The right ventricle is nearly always
enlarged on radiography and there may be some
increase of the left ventricle also.

There are three main features in the lung
fields; (1) the general mottling and increased
density, (2) the dilatation of the pulmonary
arteries and of their main branches, and (3) the
expansile pulsation. The first two features are
well seen on the film, but may be better assessed
on radiography because then the degree of pulsa-
tion can also be taken into account.

Sometimes the pulsation seen on screening
amounts to a well-marked hilar dance. There is
no close correlation between the presence of a
hilar dance and of a diastolic murmur to sug-
gest that the former is due to pulmonary re-
gurgitation. Nevertheless, a pulmonary dias-
tolic murmur in a cyanotic case should suggest
the probability of visible pulsation in the lung
fields.

Right ventricular preponderance is the gen-
eral rule and there is often a large pointed P
wave in lead II, so that the electrocardiogram
is very similar to that of Fallot’s tetralogy.

Difficulty in diagnosis may arise when there
is a heavy collateral circulation to the lungs.
Generally this shows as an increase of linear
markings in the lung fields or small round spots
when the vessels are seen in cross section. In
contrast, cases with transposition show patchy
shadows, often with clearly defined edges, with
expansile pulsation or changing density on radi-
ography. These changes are pathognomonic of
increased pulmonary blood flow and in a case
with central cyanosis generally indicate partial
or complete transposition of the aorta and pul-
monary artery.

We have also described 2 cases of transposi-
tion of the aorta and pulmonary artery with
infundibular stenosis where the diagnosis was
proved by necropsy. The picture of transposi-
tion is obscured by the effects of the stenosis
and may be confused with Fallot’s tetralogy.

**Appendix: Case Notes**

*Case 1 (0052)*, seen in 1947, aged 9 years. Able to
walk only a few yards at a time. Cyanosis dating
from birth fairly severe, with gross clubbing of the fingers. No squatting but sitting with legs crossed like a tailor.

Slight systolic murmur at apex and in second left intercostal space, with triple rhythm at apex due to a split first sound. No thrill and no diastolic murmur. Pulmonary second sound variable from slightly to greatly accentuated and drum-like, especially during expiration. Blood pressure 100/60. Extreme right ventricular preponderance with large and pointed P waves in leads II and III in electrocardiogram.

Case 2 (0145), seen in 1948, aged 4 years. Cyanosis of moderate severity since birth, not progressive. Moderately severe clubbing. Could walk only 150 yards slowly. No squatting.

Harsh systolic murmur left side, maximal at apex, with some splitting of first sound. Second sound in pulmonary area (and towards apex) moderately accentuated with short diastolic whiff following.

Electrocardiogram: some right axis deviation but not characteristic of right ventricular preponderance in chest leads; QRS 0.08 second.

Case 3 (0109), seen in 1948, aged 11 years. Able to walk only 200 yards but got about the house fairly well. Moderately cyanosed from birth with fairly severe clubbing and moderate polycythemia. Had not squatted.

Systolic murmur in pulmonary area and at apex. Pulmonary second sound very loud and booming, and sometimes with a diastolic murmur after it and also at apex. Blood pressure, 90/70. Extreme right ventricular preponderance; some increase of P waves.

Case 4 (0076), seen in 1947, aged 13 years. Fairly severe and progressive cyanosis from 7 months old. Severe clubbing of fingers. “Pneumonia” at 2, 9, and 11 years of age. Recurrent bronchitis and asthma, often with hemoptysis. Sometimes able to walk slowly up to half a mile or more, improving during last two years. Began to squat at the age of nine.

Harsh systolic murmur maximal in pulmonary area with faint systolic thrill. Second sound in pulmonary area always accentuated and palpable, sometimes strikingly so. Short diastolic murmur sometimes heard after pulmonary second sound, especially when this is greatly accentuated, and also heard at apex. Blood pressure, 110/80. Electrocardiogram, right ventricular preponderance with large and pointed P waves in lead II.

Case 5 (0A06), seen in 1947, aged 6 years. Cyanosis of moderate severity from birth with moderate clubbing of fingers. Can now walk only 100 to 200 yards slowly without panting for breath; has become worse. Squats, but less than many patients.

Widespread systolic murmur, maximal in the second left intercostal space, and faint systolic thrill. Diastolic murmur well heard at apex. Second sound in pulmonary area much accentuated. Electrocardiogram, extreme right preponderance with large and pointed P wave in lead II.

Case 6 (0175), seen in 1948, aged 11 years. Moderate cyanosis from 6 weeks old, and moderate clubbing. Can generally walk only 200 yards but sometimes nearly a mile slowly. No true squatting but sit with legs crossed like a tailor.

Rough systolic and diastolic murmurs in pulmonary area and at apex. Systolic and diastolic thrill at apex. Pulmonary second sound accentuated and drumlike.

Electrocardiogram, some left axis deviation in standard leads, but right ventricular preponderance in chest leads, large R and absent S with T wave inversion in leads V1 through V5. QRS complexes widened and notched with P-R interval of 0.28 second. P waves large and rather broad in leads I and II.

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