Transposition of the Great Vessels

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The vast majority of cases of transposition of the great vessels can be diagnosed in the first few weeks of life with electrocardiogram, x-ray and fluoroscope. The clinical pattern and the diagnostic features are described in detail. The degree of cyanosis, the characteristic heart shape and progressive increase in size coupled with generous hilar shadows all suggest this diagnosis. A new suggestion is made regarding possible surgical therapy using an extracorporeal circulation.

After the tetralogy of Fallot, transposition of the great vessels appears to be the leading cause of death in congenital heart disease. This has not been generally appreciated since most authors refer to complete transposition of the vessels as a somewhat rare malformation. Kato reviewed the literature up to 1930 and reported five cases of his own. Hanlon and Blalock brought the total collected in the literature up to 123 by their paper in 1948. More recently Astley and Parsons (1952) reported 30 cases from the Childrens' Hospital in Birmingham.

Reviewing the postmortem records of the past 10 years at the Hospital for Sick Children in Toronto, we have been able to collect 62 cases. Gardiner and Keith recorded it as the second leading cause of death from congenital malformation of the heart in the Cardiac Registry reported in 1950. In this Registry it will be noted that approximately one-seventh of the cases dying of congenital heart disease had this defect. Thus throughout Canada one might estimate there are approximately 100 deaths from this cause each year. In the United States of America there would be 10 times as many.

Both because of its relative prevalence and its high mortality it thus assumes a role of importance in the field of congenital heart disease, and it is of general interest to study the pathology, diagnosis and possible surgical cure. Kerley indicated that the diagnosis was difficult during life without angiocardiography. In other congenital defects familiarity with the clinical details has lead to accurate diagnosis with relative ease in most instances, and we believe the same may soon be said of transposition of the great vessels.

Pathology

Among the 62 cases which have come to postmortem examination at the Hospital for Sick Children and which could be classified as complete transposition of the great vessels there are 18 that belong in a somewhat separate category. The hearts in five of these are now referred to as Taussig heart. In eight cases there was a single ventricle and they thus presented a different clinical picture. In one the tricuspid valve was over-riding both ventricles and in two there was pulmonary stenosis or hypoplasia with the transposition. There were two cases with tricuspid atresia as an associated defect. This leaves 47 that have come to the postmortem table with transposition of the great vessels. The pulmonary artery arose from the left ventricle and the aorta from the right ventricle. Three of these cases appeared at first glance to be identical with the others but on detailed examination slight over-riding of the aorta was considered to be present. Thus 44 cases of complete transposition of the great vessels were available for detailed study and the following report is based on them.

A Summary of the Postmortem Findings Is As Follows:

The foramen ovale was patent in all cases examined at postmortem examination. An auricular septal defect was present in two

Aided by a grant from the Ontario Heart Foundation.

Drs. Neill and Vlad were holders of Cardiac Fellowships, President’s Research Fund, University of Toronto.
instances. A ventricular septal defect was found in 18 cases and a patent ductus arteriosus in 25. One had a minor mitral valve anomaly which did not interfere with normal function. These defects were all relatively minor in their size and type except for one very large ventricular septal defect. There were four cases that showed major defects. These included one with situs inversus, two with infantile coarctation of the aorta. Thus the majority of cases had relatively unimportant associated defects and if they had not had their great vessels transposed would have been able to lead a normal life.

A comparison of the heart weights and x-ray measurements indicated a greater degree of hypertrophy than could be appreciated by x-ray study alone. The heart enlarged rapidly after birth and usually attained a weight that was about twice normal within four to six weeks. The transverse diameter as shown by x-ray films increased during the first two months of life but did not portray the great increase in heart weight that was in progress concurrently. The transverse diameters are compared with a normal average in figure 1.

The position of the coronary vessels on the aorta is important in considering possible surgical procedures. In 37 cases the coronaries were dissected. The right coronary was found to arise behind the posterior cusp in all but three instances. In these three it arose from behind the left anterior cusp. The left coronary arose from behind the left anterior cusp in all but three instances; in them it appeared behind the posterior cusp. These findings are depicted in figure 2. The practical significance of these observations is that in transposition of the great vessels both the coronaries arise in close proximity to the pulmonary artery in such a way that it is possible to transpose them to the other great vessel.

**SITUATION of the AORTIC CUSPS**

<table>
<thead>
<tr>
<th>Normal Heart</th>
<th>Transposition of Great Vessels</th>
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</thead>
<tbody>
<tr>
<td>Normal x-ray</td>
<td>37 cases</td>
</tr>
<tr>
<td>Diameter</td>
<td>85%</td>
</tr>
<tr>
<td>Heart</td>
<td>34 cases</td>
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<tr>
<td>Ao</td>
<td>R.C.</td>
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<tr>
<td>L.C.</td>
<td>PA.</td>
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**Fig. 2.** The situation of the origin of the coronary vessels in transposition of the great vessels is compared with the arrangement in normal hearts.

**CLINICAL FINDINGS**

If operative therapy is to have a place in transposition of the great vessels an early and accurate diagnosis will be needed. Taussig pioneered in this field and showed that the diagnosis could be made during life in many cases. Increasing familiarity with the clinical and postmortem picture has added to our diagnostic criteria and we have found the following facts to be useful: age, degree of cyanosis, presence of dyspnea, heart size and shape, hilar shadowing and the angiogram. The following diagnostic criteria are based on clinical findings of 44 cases that have had the diagnosis verified at postmortem examination.

It will be noted in the accompanying chart that the average age at death in our series was 3 months. (See fig. 3.) There were 22 under 1 month, 16 between 1 and 6 months, and 6 cases over 6 months of age. Thus one is dealing with a severe congenital defect of the heart in the
early months of life. It is of considerable interest that of 44 cases there were 31 males and only 13 females.

The second diagnostic point is cyanosis. Out of 33 of 44 who had cyanosis noted at birth, in 11 it was not obvious enough to attract the attention of the attending doctor, and the saturation varied between 20 and 65 per cent; crying brought it to an extremely low level. The percentage saturation will vary considerably over a period of one hour as is shown in the accompanying chart (fig. 4).

When over-riding of the aorta occurs or when a Taussig heart is present the baby may be much less cyanotic. Then the arterial oxygen saturation will usually vary between 50 and 80 per cent.

The question of heart murmurs did not prove as sterile as was anticipated. There were 28 out of 44 that had a systolic murmur over the precordium. Fourteen had no murmur and in one case the presence or absence of a murmur was not known. There were 18 cases that had a ventricular septal defect and 13 of these had a systolic murmur over the precordium. There were 14 cases without a systolic murmur and only two of these had a ventricular septal defect and one of these died at 5 days of age. Twenty-six had no ventricular septal defect and of these eight had systolic murmurs. From these facts there are two points of value in considering murmurs. The first is that one-third of these babies with transposition of the great vessels had no murmurs and thus the absence of murmurs is of some significance in making a diagnosis. The second point is that
if a murmur is not present it is unlikely that one is dealing with an associated ventricular septal defect. The x-ray is a most useful tool in making a diagnosis. At birth the heart in transposition of the great vessels is close to normal in size and shape as is shown in figure 6. After three to four weeks of life a characteristic shape and appearance has usually emerged. (See fig. 5.) The heart becomes enlarged to a considerable degree to both right and left. The left cardiac surface assumes a convex border (the opposite of tetralogy of Fallot) and the apex points down and out. The left border can be readily shown at postmortem examination as composed of the left ventricle. The shoulder of the left ventricle comes up to the area where one expects to see the pulmonary artery. Taussig has shown that since in many cases the pulmonary artery is posteriorly placed the great vessel area is thus narrowed in the anteroposterior view and widened in the left anterior oblique view. This approach to diagnosis is most useful at birth since later when the left ventricle has hypertrophied it often enlarges upwards to the point of obscuring the pulmonary artery in either the anteroposterior or the lateral views. The narrow neck is then due to the tip of the arch of the aorta with the vertebral column above it. The pulmonary artery is buried in the cardiac shadow below the aorta and below the point where the left cardiac border meets the great vessel or supracardiac shadow.

Another diagnostic point is the relatively rapid enlargement of the heart after 3 to 4 weeks. This is shown in figure 6; case 4 is a good example. At the age of 3 days the heart
appears about normal size. At the age of 2 months it has enlarged considerably and at 10 weeks it is larger again. The postmortem heart weights confirm this increase in size and demonstrate that the gain in weight is actually greater than is indicated by the changes in x-ray films.

An enlarged heart was noted in 33 of the 36 cases whose x-ray films were available for study. Diagrammatic outlines of these hearts after the manner of Astley and Parsons are shown in the accompanying chart (fig. 7). There is a similar pattern in most which, taken with the age of the baby and the marked cyanosis, is highly suggestive of transposition of the great vessels. There are a few other conditions such as abnormal venous return, Taussig heart or Eisenmenger's complex with marked dextroposition of the aorta that may simulate complete transposition of the great vessels. However such cases usually live longer, are less cyanotic and reveal a different angiocardio-graphic pattern.

The x-ray reveals one other diagnostic point recorded in the right ventricle was as 110.0; in the left ventricle it was 67.0. Thus the pressure in the left ventricle, while raised, is not up to the systemic and therefore does not produce as large hilar shadows as are present with a Taussig heart or in an Eisenmenger's complex nor is pulsation as readily noticeable. In the first year of life one can easily discern hilar pulsations in complete transposition of the great vessels.

Angiocardiography was carried out in 18 cases. It added to the diagnostic clarity in 17

Fig. 7. Figures 6 and 7 show the x-ray silhouettes of 29 cases of complete transposition of the great vessels and illustrate the relatively common typical pattern described above.
cases in which it was performed in the left oblique or lateral position. The aorta could be seen clearly coming off the right ventricle in a position farther to the right and more anteriorly placed than in the normal heart or in the tetralogy of Fallot. This is demonstrated in figure 8. The postmortem specimen is shown beside the angiocardiogram.

When the baby is quite cyanosed and the x-ray and fluoroscopic appearance of the heart transposed. The left auricle may fill quickly but it is difficult to tell whether this is through an auricular septal defect or the foramen ovale which is invariably patent. Most ordinary ventricular septal defects are small (usually less than 0.5 cm.) in infants with transposition of the great vessels and do not show in the angiocardiogram. There are, however, a few infants, usually less cyanotic than the rest, who have the aorta over-riding both ventricles.

are characteristic, the diagnosis is usually not in doubt. The angiocardiogram is probably of most use in studying the cases with only moderate cyanosis or those whose cardiac silhouettes are not typical. In the series reported here such problem cases were the exception rather than the rule but when they did occur the angiocardiogram was useful.

With angiocardiography it is usually difficult to demonstrate clearly the associated defects that may occur when the great vessels are to a sufficient degree to show simultaneous filling of the aorta and pulmonary artery. Here the aorta fills prominently early and the pulmonary artery less prominently and a little later. In the Taussig heart the aorta fills in the way usual in transposition but the pulmonary artery may be seen to over-ride both ventricles.

The angiocardiogram is useful in the problem cases but even there it does not always clarify all the anatomic details one is searching for. The electrocardiograms were available in
30 of 50 cases and all showed right axis deviation. In six cases precordial leads had been taken and these showed right ventricular hypertrophy with some balanced hypertrophy of both ventricles. Such changes were within normal limits in the first month or two of life but as the age increased the right ventricular hypertrophy became more noticeably abnormal.

In the differential diagnosis of cyanosis at birth there are many infants with cyanosis due to atelectasis. At times it may be difficult to tell them apart from those with transposition of the great vessels, especially in the first few days of life. One useful point is the deepening of cyanosis with crying in infants with transposition. Infants who show cyanosis with atelectasis may show a decrease of cyanosis with crying. The administration of oxygen relieves the cyanosis of those with transposition to a moderate degree only, while cyanosis due to atelectasis is markedly improved by oxygen.

**Possible Correction of Transposition of the Great Vessels**

Blalock has done a good deal of pioneer surgery in the field of transposition of the great vessels and in a recent publication lists 12 methods of attempting to improve the circulation in these babies. The procedure that he found most effective was a combination of two operations carried out at the same time. One transposed the subclavian artery to a branch of the right pulmonary and the second produced an auricular septal defect. With these procedures the patients were more likely to survive the operation and be somewhat improved following it. The arterial oxygen saturation did not show any significant rise in most cases and in others increased only 10 to 20 per cent so that the results were not considered highly satisfactory, not nearly as satisfactory as those produced by the Blalock-Taussig operation in the tetralogy of Fallot.

Most of the attempts at corrective surgery have thus been aimed primarily at transposing the venous return. An auricular septal defect increases the cross circulation but to attempt to transpose the great veins entering the heart with the pulmonary veins would lead the surgeon into several major difficulties.

In some respects it would appear simpler to transpose the arterial trunks since there are only two of these and since they are placed side by side. However two major problems arise. First that of maintaining the circulation to the rest of the body during the procedure and second that of providing a sufficient blood flow to the coronary arteries.

Mustard and Chute have devised an artificial heart-lung preparation that aerates the blood with the lungs of monkeys and can effectively maintain the circulation without the heart functioning as the pump. It was suggested to them that transposition of the great vessels would be a suitable problem to tackle with this apparatus since practically all of these infants die in the first few months of life, thus justifying somewhat heroic surgery.

Four infants with transposition of the great vessels have had their circulation maintained on the heart-lung machine for periods of 10 minutes to 5 hours. Adequate oxygen content and blood pressures were maintained.

To transfer the great vessels requires cutting them near their origin and then suturing them on to their proper ventricles. As soon as the aorta is cut it becomes necessary to maintain the coronary circulation. (If the heart is allowed to stop beating and no coronary circulation is maintained it is likely to fibrillate and when the circulation is re-established the heart cannot adequately maintain the circulation.) With this in mind two small cannulas were prepared which can be inserted into the mouths of the coronary vessels and a sufficient blood flow kept up to supply the needs of the heart muscle at rest and out of the circulation.

The position of the coronary arteries as they arise from the aorta is in close proximity to the pulmonary artery and this permits transposing either one or both of the coronary arteries at the time the great vessels are surgically transposed. This would not always be so in the normal heart where the right coronary may arise a considerable distance from the pulmonary artery.

A retransposition operation has been attempted on four cases. Details are being presented by Mustard and co-workers in a separate publication. These patients have all died,
indicating that there are several major difficulties to surmount if this approach is to be successful but since it offers some promise the result of one case will be referred to here.

J. B., who had signs indicating complete transposition of the great vessels, was operated on in March 1952. The left femoral vein and artery and left jugular vein were cannulated and joined to the artificial heart-lung preparation. A continuous reading was recorded on the oximeter and is shown in the accompanying figure 9.

In figure 9 the rise in oxygen is shown when the anesthetic was started and the baby asleep and completely relaxed. Another point of note in the oximetry curve is when the artificial heart-lung preparation takes over the circulation. Here the oxygen rises to approximately 100 per cent. At this time the aorta and pulmonary artery were clamped at their bases and then transposed to their proper ventricles. During this time the oxygen saturation continued at 100 per cent. When this procedure was completed and the left coronary only had been dissected out and transferred under the aorta to the left ventricle, the clamps were then removed and the blood allowed to circulate through the retransposed vessels.

The heart beat fairly well under the new conditions with aorta coming off the left ventricle and pulmonary artery off the right ventricle, but the heart beat was not strong enough to maintain the circulation without the help of the heart-lung preparation. At point X on figure 9 the circulation was thrust upon the babies heart alone and fibrillation occurred. The heart finally stopped altogether.

The attempts at surgical correction of transposition of the great vessels are being reported elsewhere by Mustard and co-workers. Many problems need further study and elaboration.

These include the technic of transposing the coronary vessels; the type of suturing used to implant these in the wall of the transposed vessel; the maintenance of adequate coronary circulation during the procedure; the control of bleeding at the site of the suture line after vessel transfer is completed. The present report is presented as a discussion of the pathology underlying the conception of transposing the arteries to their proper ventricles and special stress is placed on the early and accurate diagnosis of this severe and commonly fatal congenital anomaly.

**SUMMARY**

Familiarity with clinical and postmortem findings in complete transposition of the great
vessels now makes diagnosis possible in the vast majority of cases during life. One usually finds a baby in the first few weeks of life who has been markedly cyanotic since birth. The chances are two to one that it is a boy. He is underdeveloped and the gain in weight has been slow. A third of these babies have no murmur when examined. In all the electrocardiogram shows right axis deviation. An x-ray film of the heart shows a relatively normal contour at birth, but three or four weeks later there is a fairly characteristic shape. The heart by that time is enlarged to both right and left and there is a convex left border. The great-vessel area appears narrow in the anteroposterior view. The hilar shadows are increased. Angiocardiography shows the aorta coming clearly off the right ventricle. Sometimes this procedure may outline the pulmonary artery as well and aid in determining the presence or absence of associated defects. These diagnostic criteria have proved very satisfactory and the high natural mortality permits one to check the diagnosis frequently.

Operative technics for this condition have been mainly directed to the venous return. It is now suggested that transposing the arterial trunks to their proper ventricles might prove to be a more satisfactory procedure. To carry this out an artificial heart and lung preparation is necessary. Furthermore the coronary circulation must be maintained during the procedure and carried over with the aorta to the proper ventricle. Recent work by Mustard and Chute suggests that this procedure may be feasible.

ACKNOWLEDGMENT

This work was begun in the hope that some adequate surgical procedure will eventually be evolved. The authors would like to express their appreciation of the continuous cooperation, interest and encouragement from Dr. Mustard and the Surgical Service of the Hospital for Sick Children, Toronto.

SUMARIO ESPAÑOL

La gran mayoría de los casos de transposición de los grandes vasos pueden ser diagnosticados en las primeras semanas de vida con el electrocardiograma, radiografía y el fluoroscopio. El cuadro clínico y los puntos diagnósticos se describen en detalle. El grado de cianosis, la silueta cardíaca característica y el progresivo incremento en tamaño acompañado de sombras de hilo aumentadas sugieren este diagnóstico. Una nueva sugerencia se hace en cuanto al posible tratamiento quirúrgico usando circulación extracorpórea.

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Circulation. 1953;7:830-838
doi: 10.1161/01.CIR.7.6.830

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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