Transposition of the Great Vessels with Atrial Septal Defect

A Hemodynamic Study in Two Cases

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In complete transposition of the great vessels, survival is possible only when a route of access exists by means of which venous blood may enter the lungs. This report concerns 2 patients in relatively good health in whom a high degree of mixing of venous and arterial blood occurred through an atrial septal defect. The diagnosis was established in both patients by cardiac catheterization.

According to Keith and associates,1 complete transposition of the great vessels is found in approximately 12 per cent of patients who die in infancy with congenital heart disease. In the classic form of this condition, the aorta arises from the right ventricle and lies anteriorly and on a plane slightly to the right of the pulmonary artery. Depending on the degree of counterclockwise rotation from 90 to 180 degrees, the position of the aorta in relation to the pulmonary artery may be variable and thus many anatomic subvariations have been described. In the basic malformation there is no arrangement by which venous blood can pass through the lungs and then to the systemic circulation; therefore this condition is incompatible with life. Valve-competent foramen ovale or atrial septal defect, ventricular septal defect, or patent ductus arteriosus, alone or in combination are the commoner pathways that allow mixing of the blood between pulmonary and systemic circulations. In approximately 40 per cent of cases the interventricular septum is closed and in approximately 8 per cent an atrial septal defect is the only communication between the two sides of the heart. Anomalous pulmonary venous drainage is rarely an associated anomaly.

Transposition occurs more frequently in males than in females in a ratio of 3:1. Evidence of the clinical triad of central cyanosis, congestive heart failure and pulmonary plethora on a roentgenogram of the thorax should cause the clinician to suspect this diagnosis. Eighty-five per cent of the patients are cyanotic at birth, and congestive heart failure develops by the age of 6 months. A third of the patients do not have an audible murmur and the intensity of the systolic murmur, when present, is variable.2 The size of the heart is normal at birth and cardiac enlargement and the characteristic egg-shaped cardiac silhouette develop in the subsequent months. A narrow vascular pedicle in the anteroposterior position and a widened vascular pedicle in the left anterior oblique view with pulmonary plethora are often seen. The electrocardiogram is not characteristic but shows varying degrees of right-axis deviation and right ventricular hypertrophy.

The diagnosis of the transposition and any associated abnormality has been made by the use of selective angiocardiography.3,4 The aortic valve may lie at the level of the fourth to fifth thoracic vertebra instead of at the level of the seventh vertebra, but this relation is variable. The aorta is seen to arise anteriorly from the right ventricle.

It is the purpose of this paper to report 2 cases of complete transposition of the great vessels with atrial septal defect as the only communication between the 2 circulations. A
positive diagnosis was established during catheterization of the right side of the heart which included the use of indicator-dilution technics but did not include the use of angiocardiography.

CASE REPORTS

Case 1. The tenth child of a family was born at term after an uneventful pregnancy. He was intensely cyanotic at birth and spent the first week of his life in an incubator. A heart murmur was noted at the age of 3 months and the patient was referred to the Mayo Clinic for evaluation.

On examination he was found to be well developed for his age; he was moderately cyanotic without clubbing of the fingers or toes. A moderately loud systolic murmur was heard along the left sternal border, maximal in the third and fourth intercostal spaces. The second heart sound at the base was loud and was not duplicated. Both femoral arterial pulses were palpable; the blood pressure was normal, and the results of the remainder of the physical examination were within normal limits. The electrocardiogram was characterized by sinus rhythm, right-axis deviation, and evidence of right atrial enlargement and right ventricular hypertrophy. The hemogram and results of urinalysis were normal. A roentgenogram of the thorax revealed the size of the heart to be within normal limits; the pulmonary vascular markings were normal.

The patient was seen again at the age of 2½ years. He was well developed (weight, 33 pounds; length 37½ inches); he was moderately cyanotic with clubbing of the fingers; no squatting had been noted. The results of physical examination were essentially unchanged but the thoracic roentgenogram showed some evidence of cardiac enlargement (fig. 1a) and polycythemia had developed (hemoglobin, 16.8 Gm. per 100 ml of blood). The saturation of systemic arterial blood as determined by ear oximetry was 81 per cent at rest, 69 per cent during exercise while breathing air and 91 per cent while breathing oxygen at rest. The electrocardiogram is shown in figure 1b.

Catheterization² of the right side of the heart was carried out with the patient under balanced anaesthesia and breathing 100 per cent oxygen. The pressures in the great veins and right atrium were within the range of normal (table 1). The pressure in the right ventricle was severely elevated and identical to that in the femoral artery. At a later time the catheter passed across the midline at the atrial level to enter the left ventricle (fig. 2). There was no significant pressure gradient across the interatrial septum. The central great vessels were not entered.

In spite of the breathing of 100 per cent oxygen, systemic arterial blood was desaturated to 89 per cent and the saturation of mixed venous blood was reduced to 70 per cent. There was arterialization of the blood in the right atrium equivalent to a left-to-right shunt of approximately 40 per cent. The saturations of femoral arterial and right ventricular blood were not significantly different. The saturation of pulmonary venous blood was 100 per cent, and it must be presumed that a further complement was present in dissolved form, since the patient continued to breathe 100 per cent oxygen. The oxygen saturation of left ventricular blood was significantly less than that of the pulmonary venous blood. Assuming the oxygen content of pulmonary venous blood to exceed capacity by 1.8 ml per
100 ml. of blood, the pulmonary blood flow was calculated to be 10.7 L per minute per M.² of surface, whereas the systemic flow was 3.9 L.

An understanding of the indicator-dilution curves seen in transposition of the great vessels with intact ventricular septum may be facilitated by figure 3, which illustrates the path taken by the indicator following its injection into the right and left ventricles.

The dilution curves obtained in case 1 are shown in figure 4. The differences in appearance times and peak concentration values after injection of the dye into the 2 ventricles are apparent. All the dye injected into the right ventricle appeared to have passed directly to the systemic arterial system. In contrast, a longer appearance time, reduced peak deflection, and prolongation of the disappearance slope characterize the curve obtained after left ventricular injection. The latter are indicative of pulmonary recirculation.

The dye curves recorded after injection of indicator into the superior and inferior venae cavae are essentially similar to one another in appearance time and contour. They differ from the curve obtained after injection into the right ventricle in that the peak concentrations are smaller, and the disappearance phase fails to show the nearly complete return to zero concentration shown on the right ventricular curve. On comparison of the caval dilution curves with the latter, it appears that approximately 45 per cent of the blood draining from the superior vena cava is shunted across an interatrial communication into the pulmonary circulation and an even greater amount of blood from the inferior vena cava reaches the left atrium. This is analogous to the demonstration of a larger right-to-left shunt from the inferior than from the superior vena cava in patients without transposition who have an interatrial communication located in the fossa ovalis.

The striking difference in the drainage pattern of blood from the pulmonary vein and the venae cavae indicates that the pulmonary veins are connected to the left atrium, while in comparison with the curve obtained after injection into the left ventricle, the shorter appearance time and greater initial deflection for the pulmonary vein curve are evidence for arteriovenous shunting at the atrial level.

**Case 2.** A 3-year-old boy, the first child of his family, was born at term after an uneventful pregnancy. He had been cyanotic since birth, when a heart murmur was noted. During the first 2½ years of his life this child was hospitalized many times with otitis media and repeated infections of the upper respiratory tract. At the age of 9 months he had had staphylooccal septicaemia with cerebral embolism. Left residual hemiplegia and expressive aphasia developed subsequent to this illness.

On examination he was fairly well developed (weight, 31½ pounds; length, 35½ inches) with moderate cyanosis and clubbing. A moderately loud systolic murmur was heard along the left sternal border and the second sound in the pulmonary area was not duplicated. The results of neurologic examination were consistent with those associated with a right cerebral lesion.

The hemoglobin was 18.4 Gm. per 100 ml. of blood. The electrocardiogram showed the mean

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<th>Table 1.—Pressure Measurements and Oxygen Saturations of Blood Samples in Great Vessels and Chambers of Heart in Two Cases of Transposition with Atrial Septal Defect</th>
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<td>Site</td>
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*These samples were not taken simultaneously.
Fig. 3. Schematic representation of the heart in transposition of the great vessels with atrial septal defect showing the paths taken by dye introduced into the right ventricle (left) and into the left ventricle (right). Note the short direct circulatory path to the arterial system taken by material introduced into the right ventricle and the much longer and more complex path followed by material introduced into the left ventricle. The actual arterial dilution curves recorded in the two patients considered in this paper are shown in figures 4 and 7.

Fig. 4. Indicator-dilution curves recorded at right femoral artery in case 1. The instant of injection of dye is indicated by the vertical arrows. Note: 1. The large initial deflection followed by an almost complete clearance of dye after injection into the outflow portion of the right ventricle (fig. 2, left). This alone indicates that no significant flow of blood to the lungs occurs at or distal to the right ventricle. 2. The differences in magnitude of peak deflection and in contour between the curves recorded after injection into superior and inferior venae cavae and right ventricle. 3. Evidence for pulmonary recirculation in the curves recorded after injection into pulmonary vein and left ventricle. 4. Longer appearance time for curves recorded after injection into left ventricle in contrast to that for pulmonary vein, localizing the left-to-right shunt to atrial level.
entered from the right ventricular outflow tract (fig. 6a and b). During the procedure the catheter entered the left atrium through an atrial septal defect and was advanced into the left ventricle (fig. 6c) where the pressure was low. There was an apparently significant pressure gradient between the 2 atria. The pulmonary artery was not entered.

The oxygen saturation of femoral arterial blood of 68 per cent indicated severe systemic desaturation and was identical with that of the right ventricle and aorta. The saturation of mixed venous blood was likewise reduced. There was good evidence of arterIALIZATION of venous blood at the atrial level, equivalent to a left-to-right shunt of 28 per cent. Assuming a pulmonary venous oxygen saturation of 98 per cent, the drop in oxygen saturation of blood in the left atrium must have been due to a right-to-left shunt across the atrial defect. The pulmonary blood flow, as calculated by Fick's principle, measured 8.3 L per minute per M.2 and the systemic flow measured 5.2 L per minute per M.2

The dye-dilution curves are shown in figure 7. There is an obvious difference in the appearance time, peak deflection and disappearance slope of the 2 curves after injection of dye into the right and left ventricles respectively.

Dye injected into the right ventricle appeared at the femoral artery almost immediately. The resultant dilution curve had a rapid upstroke and sharp downstroke with almost complete return to the base line, followed in 10 seconds by a second deflection. This was almost certainly due to the passage into the arterial system of indicator which had returned to the heart from the systemic circulation. Dye injected into the left ventricle passed through the pulmonary circulation before reaching the sampling site. This curve is characterized by a longer appearance time, reduced peak concentration, and a marked prolongation of the disappearance slope.

The curves obtained after dye was injected into the aorta and right ventricle, respectively, were identical and excluded any hemodynamically significant communication to the pulmonary vascular bed at or distal to the ventricular level.

Injection of dye into the superior vena cava resulted in a curve that differed from those obtained after injection into the right ventricle and aorta in that the area subtending this curve was reduced by approximately 25 per cent and the clearance of dye on first circulation was less complete. The reduction in the area of the initial portion of the curve is due to the loss of dye that had been shunted across the atrial septal defect into the left atrium while the incomplete clearance is explained

Fig. 5. Case 2. a. The electrocardiogram shows extreme right axis deviation with evidence of atrial enlargement and right ventricular hypertrophy. b. Anteroposterior view of thorax. Note slight cardiac enlargement and a moderate increase in pulmonary vascular shadows. The area usually occupied by the main pulmonary artery is concave. The aortic arch is on the left.

QRS axis to lie between 170° and 180°; there was evidence of right atrial enlargement and right ventricular hypertrophy; the left ventricular potential was not well represented by the standard electrocardiographic leads (fig. 5a). The roentgenogram of the thorax showed evidence of generalized cardiac enlargement with increased pulmonary vascular markings (fig. 5b).

The pressure in the right atrium was normal on catheterization of the right side of the heart (table 1). The right ventricular pressure was elevated and identical with that in the aorta, which was
Fig. 6. Roentgenograms of the thorax showing the position of catheters in the outflow portion of right ventricle (left), the root of the aorta (middle) (proved by indicator-dilution curves) and the left ventricle (right). The aortic valve was not displaced cephalad and to the right as usually seen in cases of transposition. Unfortunately a lateral roentgenogram was not obtained. In each instance systolic and diastolic pressures and oxygen saturation values are given.

by the circulation of the shunted dye through the lungs. A part then re-enters the right side of the heart (shunted left to right) across the atrial septal defect. The curve recorded after injection of dye into the left atrium showed a shorter appearance time compared to that obtained after injection into the left ventricle, which served to localize the shunt to the atrial level.

**DISCUSSION**

In both cases the following facts were established during catheterization of the heart:

1. There are 2 anatomically and functionally separate ventricles. The systemic ventricle, which is a high-pressure chamber and supplies the high-resistance systemic arterial bed, is the anatomic right ventricle. The anatomic left ventricle is a low-pressure ventricle that supplies only the low-resistance pulmonary vascular bed. The pressure measurements alone prove the presence of separate pathways of egress from each ventricle.

2. The oxygen saturation of the femoral arterial blood is identical with that from the right ventricle and aorta and is much less than that of blood from the left ventricle. Arterialization of the caval blood takes place at atrial level, and this mixture supplies the systemic arterial system.

3. There is no further drop in the oxygen saturation of blood from the left ventricle as compared to that of the left atrium. This alone, in the presence of the significant pressure gradient across the ventricular septum, excludes any significant shunt at ventricular level.

4. The dye-dilution curves confirm the presence of 2 separate drainage pathways from the ventricles. The longer appearance time and prolonged disappearance phase found after injection into the right ventricle show that blood from this chamber first passes through the pulmonary vascular bed before a part reaches the systemic circulation. Similarly, the contour and appearance time of the dye curves after injection into the right ventricle and the aorta prove that they alone
supply the systemic circulation. At the same time the practically complete clearance of dye shown by these curves excludes any communication of significant size between the pulmonary and systemic circulations at, or distal to, the right ventricle.

The preferential shunting of blood from the inferior vena cava and the decreased peak concentration of the curves after injection of dye into the venae cavae strongly suggest the presence of an atrial septal defect located in the fossa ovalis. The short appearance time that follows injection of dye into the left atrium as compared with the longer appearance time after injection into the left ventricle localizes, in addition, a left-to-right shunt at atrial level.

5. The low pressure in the left ventricle excludes severe pulmonary stenosis or significant pulmonary hypertension.

6. The pulmonary blood flow significantly exceeds the systemic flow in both cases. The absolute magnitude of the volume shunted in both directions across the atrial septum is considerable and is related to the degree of intermixing between the right and left atrial chambers.

These findings establish the diagnosis of transposition of the great vessels with intact ventricular septum and atrial septal defect. The pressure measurements, oxygen saturation data, and dye-dilution curves cannot be explained on the basis of any other anatomic or hemodynamic abnormality.

Two additional points are worthy of comment. From the clinical point of view it is remarkable that at no time did either patient have any symptom or sign of congestive heart failure considered characteristic of this condition. This is almost certainly related to the substantial degree of mixing that was apparently occurring freely at atrial level.

In figure 6b the roentgenogram made with the tip of the cardiac catheter lying just above the aortic valve suggests that the location of this valve may be normal and not at the level of the fourth thoracic vertebra as has been described in complete transposition of the great vessels.1 This may represent transposition type I of Castellanos and associates.

**Summary**

The cardiac catheterization findings in 2 patients, aged 2½ and 3 years, with moderate physical disability due to cyanotic congenital heart disease are reported. In each patient, 2 ventricles were entered, the left with a low level of pressure and a high blood-oxygen saturation, the right with pressure equal to systemic arterial pressure and a reduced blood-oxygen saturation equal to that of systemic arterial blood. It was demonstrated by indicator-dilution curves that all blood from the right ventricle passed into the systemic circulation, while all blood from the left ventricle passed into the pulmonary circulation. Mixing of systemic venous and pulmonary venous bloods at atrial level was also demonstrated. On these hemodynamic data the diagnosis of complete transposition of the great vessels with atrial septal defect was made.

**Summario in Interlingua**

Es reportate le resultatos de catheterismo cardiac in 2 patientes, de 2½ e 3 annos de etate, con moderate grades de invaliditate physic como effecto de congenite morbo cyanotic del corde. In ambe casos, penetration de ambe ventriculos eseva effectuate, le ventriculo sinistre con un bассe nivello de pression e un alte saturation de oxygeno, le ventriculo dexter eun pression equal al pression arterial in le circulation major e un saturation oxygenic del sanguine reducete al nivello trovate in le sanguine arterial del circulation major. Esseva demonstrate per curvas de dilution de un indicator que omne le sanguine ab le ventriculo dexter passava a in le circulation major durante que omne le sanguine ab le ventriculo sinistre passava a in le circulation pulmonar. Esseva etiam demonstrate le mixture de sanguines venose del duo circulationes al nivello atrial. Super le base de iste datos hemodynamic le diagnose de un transposition complete del grande vasos in association con defecto atrio-septal eseva formulate.
REFERENCES


Truly America has a great future before her; great in toil, in care, and in responsibility; great in true glory if she be guided in wisdom and righteousness; great in shame if she fail. I cannot understand why other nations should envy you, or be blind to the fact that it is for the highest interest of mankind that you should succeed; but the one condition of success, your sole safeguard, is the moral worth and intellectual clearness of the individual citizen. Education cannot give these, but it may cherish them and bring them to the front in whatever station of society they are to be found; and the universities ought to be, and may be, the fortresses of the higher life of the nation.—Thomas H. Huxley. American Addresses with a Lecture on the Study of Biology. London, MacMillan and Co., 1877, p. 126.
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Circulation. 1959;20:842-849
doi: 10.1161/01.CIR.20.5.842
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

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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