COMPLETE TRANSPOSITION OF THE AORTA AND A LEVOPOSITION OF THE PULMONARY ARTERY

CLINICAL, PHYSIOLOGICAL, AND PATHOLOGICAL FINDINGS

HELEN B. TAUSIG, M.D., AND RICHARD J. BING, M.D.

BALTIMORE, MD.

THE clinical findings presented in the following case report represent a syndrome which we have seen not infrequently in recent years in the cardiac clinic of the Harriet Lane Home. This is, however, the first instance in which it has been possible to correlate the clinical findings with the autopsy findings. The case clarified the nature of the malformation and unfortunately demonstrated one of the most serious dangers of angiocardiography. Hence, the case is reported in detail.

CASE REPORT

P. A. W. (H. L. H. A-60186), a 5½-year-old white girl, was referred to the clinic for diagnosis of her cardiac abnormality.

The family history was noncontributory. The mother had not had German measles nor any rash or unexplained fever during her pregnancy. There was no familial history of congenital abnormalities.

The past history indicated that cyanosis was noted at birth and persisted throughout her life. At 3 weeks of age, a murmur was heard over the precordium. During infancy she gained weight slowly. At the age of 1 year she weighed 7.6 kilograms. Her development was also slow: she sat alone at 9 months and walked at 2 years. When about 3 years of age she frequently squatted down to rest, but soon outgrew the habit.

Physical Examination.—The temperature was 37° C., pulse 120, respiration 30, height 110 cm., weight 15.6 kilograms, and blood pressure 100/80. The child was an intelligent, moderately cyanotic, poorly developed girl who suffered from dyspnea at rest. There was suffusion of the conjunctivae. The lips and buccal mucous membranes were deeply cyanotic. The tonsils were small. The teeth were in good condition. The heart was slightly enlarged. The rhythm was regular. A systolic murmur was audible over the precordium which was definite but not loud; no thrill could be felt. The lungs were clear to percussion and auscultation. The liver and spleen were not palpable. The pulse in the femoral artery was of good quality. There was cyanosis and clubbing of the fingers and toes.

Laboratory Data.—The red blood cell count was 9.3 million. Hemoglobin concentration was 23.5 grams. The hematocrit was 77. Arterial blood analysis showed an oxygen content of 17.4 volumes per cent, oxygen capacity of 30.8 volumes per cent, oxygen saturation of 57 per cent, and carbon dioxide content of 25.8 volumes per cent.

*From the Departments of Pediatrics and Surgery of the Johns Hopkins Hospital and from the Cardiac Clinic of the Harriet Lane Home, Johns Hopkins Hospital, Baltimore, Md.

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Teleroentgenogram.—The heart was slightly enlarged. There was fullness of the pulmonary conus and the hilar markings were increased (See Fig. 1).

Fluoroscopy.—The findings in the x-ray film were confirmed and, in addition, fluoroscopy revealed faint expansile pulsations of the hilar shadows. Delineation of the esophagus with a barium-opaque mixture showed a left aortic arch and no evidence of left auricular enlargement.

Electrocardiogram.—There was a normal sinus mechanism, sinus tachycardia, normal P-R interval, high P waves in the second lead, right axis deviation, and right ventricular hypertrophy.

Clinical Impression.—The clinical findings were characteristic of an Eisenmenger complex in that there was cyanosis, clubbing, and polycythemia; the heart was slightly enlarged with x-ray evidence of fullness of the pulmonary conus and increased hilar shadows, which upon fluoroscopy showed faint expansile pulsations. However, the fact that cyanosis dated from birth made us suspect some totally different malformation.

Fig. 1.—Teleroentgenogram of the chest, anterior-posterior position.

In Norway in the summer of 1947, one of us (H. B. T.) had been told of an infant with a similar clinical history. In this instance, examination of the heart showed an unusual anomaly of the great vessels: the aorta, which was abnormally small, arose from the right ventricle and the pulmonary artery was greatly enlarged and overrode the ventricular septum. For this reason, in the case under discussion, an overriding pulmonary artery was postulated, but, because of the
age of the patient and child's comparative well-being, the transposition of the aorta was not suspected.

Inasmuch as the diagnosis in this instance was obscure, the patient was referred to the physiological laboratory for special studies by one of us (R. J. B.) and to the x-ray department for angiocardiography.

Results of Physiological Studies.—Results obtained from the standard exercise test showed that the oxygen consumed per liter of ventilation fell from 17 to 12 cubic centimeters. From this, it was inferred that the effective pulmonary blood flow through the lungs did not increase normally with exercise.

The results of cardiac catheterization are given in Fig. 2. It may be seen that the oxygen content of the right ventricular blood was significantly higher than that of the right auricle, indicating the presence of a ventricular septal defect. Of special interest was the finding that the oxygen content of the pulmonary arterial blood exceeded that of the right ventricular blood by 4.4 volumes per cent. The finding suggested admixture of oxygenated blood with right ventricular blood. A gradient of this magnitude between the right ventricular blood and the pulmonary arterial blood could have been the result of a ductus arteriosus, or of a communication between the pulmonary artery and the left ventricle through a high ventricular septal defect with the pulmonary orifice overriding the lower portion of the ventricular septum. The clinical findings, however, rendered unlikely the diagnosis of a patent ductus arteriosus as there was no continuous machinery-like murmur. Furthermore, the peripheral arterial oxygen saturation was only 57 per cent. It seemed almost certain that a large patent ductus arteriosus would increase the effective pulmonary blood flow sufficiently to raise the oxygen saturation of peripheral arterial blood to a higher level. It was, therefore, assumed that the pulmonary artery received oxygenated blood directly from the left ventricle. Fig. 2 shows that the oxygen content of peripheral arterial blood was 7.6 volumes per cent less than that of the pulmonary arterial blood and 3.2 volumes per cent less than that of the right

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**Fig. 2.—Diagram illustrating intracardiac hemodynamics.** $O_2$ indicates oxygen content of the blood in volumes per cent. Oxygen content of the blood in pulmonary vein was calculated on the assumption that the blood was 96 per cent saturated.2 Figures in boxes give volume of blood flow in cubic centimeters per minute per square meter of body surface. It may be seen that the large volume of the right auricular blood flows directly from the right ventricle into the aorta and only a small volume of blood passes into the pulmonary artery. This latter represents the effective pulmonary blood flow.
ventricular blood. This finding indicated that the aorta must receive a large quantity of un-oxygenated blood. This could be the result of a high septal defect with the aorta overriding the ventricular septum or a complete transposition of that vessel. Although the latter possibility seemed likely, the physiological data alone were insufficient to establish the diagnosis. The pressures recorded in the pulmonary artery were 57/45 mm. of mercury; those in the right ventricle were 42/19 mm. of mercury (Fig. 3).

![Graph](image_url)

**Fig. 3.**—Continuous tracing of direct strain-gauge recording obtained in the pulmonary artery and in the right ventricle. The arrow indicates the point at which the tip of the catheter passes through the pulmonic valve into the right ventricle.

**Angiocardiography.**—After an initial test dose, the patient was given 19 c.c. of 70 per cent Diodrast intravenously through a canula and a series of eight films were taken in eight seconds. Thirty minutes later she was given another dose of 19 c.c. and a second series of eight films were taken. Because of a mechanical defect, no exposures were obtained. Therefore, fifteen minutes later, a third dose of 19 c.c. of Diodrast was injected and a series of eight films were photographed in eight seconds. There was no immediate reaction, but three minutes thereafter the child sat bolt upright and the heart stopped. All effort at resuscitation failed.

The angiocardiograms showed that the dye entered the right auricle and then the right ventricle; immediately thereafter the aorta was promptly visualized. Very little dye was seen in the pulmonary artery or the lungs. The circulation of the dye could not be traced further. The second series of films taken in the lateral position showed that the aorta appeared to arise from the anterior portion of the right ventricle. Again, the circulation of the dye could not be traced into the lungs, nor to the left side of the heart.
Final Clinical Diagnosis.—The physiological studies and angiograms indicated a transposition of the great vessels. The x-ray and fluoroscopic findings indicated that such was not the case in that the pulmonary artery appeared to arise from the right ventricle.

Autopsy (No. 21039, Performed by Dr. Edmund Novak).—The chief interest centered about the heart. It weighed 180 grams. The right auricle was not greatly enlarged. The superior vena cava and the inferior vena cava opened into it in the normal fashion. The foramen ovale was completely covered by a valve, but there was no patency of the valve for a distance of 1.0 cm. along its margin. The tricuspid valve, which was slightly thickened, opened into the right ventricle. That chamber was tremendously hypertrophied; its wall measured 1.5 cm. in thickness. The pulmonary artery arose approximately in its normal position. The aorta was transposed; it arose entirely from the right ventricle. The aortic orifice lay adjacent to the pulmonary orifice and to the ventricular septum as shown in Figs. 4 and 5. The aortic valve had three cusps and the coronary arteries were given off from the aorta in the normal manner. The aortic ring measured 3.5 cm. in circumference. The aorta and its branches appeared to be normal. The maximum circumference of the ascending aorta was 4.0 centimeters. At the base of the ventricular septum, the septal wall was defective for a distance of 1.2 cm. and the defect extended downward toward the apex for approximately 0.6 centimeter. The superior portion of the ventricular septum deviated to the right to such an extent that the pulmonary orifice overrode the septal defect by a few millimeters. From the upper margin of the ventricular septum close to the defect, a muscular ridge extended forward to the outer wall of the right ventricle. This ridge separated the aorta from the pulmonary artery. Consequently, the aorta arose entirely from the right ventricle and only the pulmonary orifice overlay the ventricular septum. Thus, the pulmonary artery not only received blood from the right ventricle, but also received blood directly from the left ventricle. The pulmonary artery and its branches were greatly dilated. The pulmonary orifice measured 5.8 cm. in circumference and the main pulmonary artery above the ring had a circumference of 6.3 centimeters. The left main branch measured 4.0 cm. in circumference; the right

Fig. 4.—Drawing of the heart showing size and position of the aorta and pulmonary artery and their relation to the septal defect.
This malformation produces a syndrome which is clinically similar to that associated with an Eisenmenger complex. In both conditions the pulmonary artery arises from the right ventricle; in both, the contour of the heart in the x-ray film shows fullness of the pulmonary conus. Both have large pulmonary arteries, which upon fluoroscopy usually show expansile pulsations. In both malformations the heart is but slightly, if at all, enlarged; both have a systolic murmur; both show evidence of right axis deviation and right ventricular hypertrophy. Both conditions are compatible with life for a number of years. In both, the habit of squatting is either entirely absent or of short duration. The outstanding clinical difference between this malformation and the Eisenmenger complex is that in the former, cyanosis dates from birth, whereas the late development of cyanosis, at or about the time of puberty, is characteristic of the Eisenmenger complex. Both conditions lead to polycythemia and clubbing of the extremities which, however, occur at a later date in patients with an Eisenmenger complex than with this malformation.

Anatomically, this malformation differs from an Eisenmenger complex in that the aorta is not dextroposed; that is, it does not arise from the left ventricle and partially override the ventricular septum, but it is transposed and arises entirely from the right ventricle. Furthermore, it is the pulmonary artery, not the aorta, which overrides the ventricular septum.

The origin of the aorta from the right ventricle means that the blood from the right ventricle is pumped directly into the aorta; this readily explains the early appearance of cyanosis. Indeed, the only oxygenated blood to reach the aorta is that which is shunted from the left ventricle through the septal defect into the right ventricle. Inasmuch as the pulmonary artery overrides the ventricular septum, blood from the left ventricle is readily directed into the pulmonary artery.

Functionally, this malformation closely resembles the malformation in which both the aorta and the pulmonary artery arise entirely from the right ventricle and the septal defect lies beneath the pulmonary artery. This last mentioned malformation is also mentioned by Pernkopf and has been classified by some as an Eisenmenger complex, but is totally different from the malformation originally described by Eisenmenger and, to use Dr. Maude Abbott’s words, “is not to be confused with an Eisenmenger complex.” Therefore, the authors feel that the term “Eisenmenger complex” should be limited to the type of malformation originally described by Eisenmenger and that the combination of a transposed aorta with a pulmonary artery which arises from the right ventricle and partially overrides the ventricular septum represents a separate clinical and pathological entity. Furthermore, the malformation, in which both great vessels arise from the right ventricle and in which the septal defect is adjacent to the posterior margin of the pulmonary orifice, is functionally more closely related to the malformation under discussion than to the Eisenmenger complex.

The origin of the aorta from the right ventricle means that venous blood is directed into the aorta and, consequently, the oxygen saturation of the arterial blood is abnormally low. Exercise causes a further fall in the oxygen saturation
of the arterial blood and a fall in the oxygen consumption per liter of ventilation. The latter finding is similar to that which occurs in a patient with a tetralogy of Fallot and, not infrequently, with a complete transposition of the great vessels, but is contrary to that which occurs in a patient with an Eisenmenger complex.

Cardiac catheterization reveals a high pressure in the right ventricle and a markedly higher oxygen content in the right ventricle than in the right auricle (Fig. 2). Therefore, if the pulmonary artery is not catheterized, the findings are similar to those in a tetralogy of Fallot. If the pulmonary artery is catheterized, the pulmonary pressure will be found to be high and the oxygen content in the pulmonary artery will be higher than that in the femoral artery.

The intracardiac hemodynamics of this patient are illustrated in Fig. 2. Her oxygen consumption was 105 c.c. per minute per square meter of body surface. As shown in Fig. 2, the systemic flow was 3,920 c.c. per minute per square meter of body surface and the pulmonary artery flow was 2,300 c.c. per minute per square meter of body surface. Thus, the systemic flow exceeded the pulmonary artery flow by 1,620 cubic centimeters. Both were calculated according to formulas published in a previous communication. The effective pulmonary blood flow is the quantity of blood which, after having been returned to the right auricle from the body, is eventually aerated in the lung. In this patient, it will be represented by the volume of mixed venous blood which enters the pulmonary artery from the right ventricle. Consequently, it can be calculated from the oxygen content of the blood in the right auricle and the oxygen content of the blood returned to the left auricle. In this instance, the effective pulmonary blood flow was found to be 710 cubic centimeters. This means that, although the volume of the pulmonary blood flow is 2,300 c.c., only 710 c.c. are mixed venous blood; the remainder is arterial blood which is recirculated through the lungs. Furthermore, in order to keep the pulmonary flow at its calculated constant value, 710 c.c. must be shunted from the left ventricle into the aorta. Since this represents the oxygenated component of the blood supplied to the systemic circulation, it represents the effective systemic flow. The remaining 3,210 c.c. of the systemic flow is mixed venous blood from the right auricle which passes into the right ventricle and is pumped out into the aorta and recirculated through the body.

The relatively small volume of blood entering the aorta from the left ventricle furnishes the only means by which oxygenated blood reaches the body. This explains the low oxygen saturation in the peripheral arterial blood and the severe cyanosis. Consequently, any diminution of this volume may have dangerous consequences. This may explain the fatal outcome of angiocardiography.

*It is our belief that any condition in which the injection of Diodrast decreases the supply of oxygen to the individual is extremely dangerous. The danger of angiocardiography in pulmonary arteriovenous aneurysms is well known. Under such circumstances, the dye is laced in the aneurysms and interferes with the exchange of oxygen in the lungs. Angiocardiography also proved fatal in a man with a cor pulmonary in whom the pulmonary arteriolar disease caused difficulty in the circulation of the blood through the lungs and in the oxygenation of the blood in the lungs. Recently, a child with an extreme pulmonary stenosis and no ventricular septal defect died after the injection of a single dose (9 c.c.) of Diodrast. In this instance, the orifice into the pulmonary artery was only 1.0 mm. in diameter and the expulsion of dye through this tiny orifice cut off the entire blood supply to the lungs and thereby deprived the child of its sole supply of oxygenated blood.

In this malformation, the increased pressure in the right side of the heart may have blocked the supply of oxygen to the systemic circulation.
In this instance, the rapid injection of the Diodrast into the superior vena cava raised the pressure in the right side of the heart and, consequently, decreased the left-to-right shunt. For this reason, angiocardiography was exceptionally dangerous for this patient. Furthermore, angiocardiography did not clarify the nature of the malformation and therefore is not necessary to establish the diagnosis.

SUMMARY

A new clinical syndrome is described. The malformation consists of a transposed aorta, a large pulmonary artery which arises primarily from the right ventricle and partially overrides the ventricular septum, a high ventricular septal defect, and right ventricular hypertrophy.

Clinically, in this instance the heart was but slightly, if at all, enlarged; there was a systolic murmur and thrill. Cyanosis dated from birth. Clubbing of the extremities developed at an early age. The red blood cell count, the level of the available hemoglobin, and the hematocrit reading were increased. The electrocardiogram showed evidence of right ventricular hypertrophy. The x-ray films of the heart showed fullness of the pulmonary conus and increased hilar shadows. Upon fluoroscopy, the pulmonary vessels showed faint expansile pulsations. The oxygen saturation of the arterial blood was abnormally low and fell still further with exercise.

In brief, the clinical syndrome associated with this malformation resembled that of an Eisenmenger complex, except that cyanosis dated from birth.

The two conditions showed a further difference in that, in the Eisenmenger complex, exercise causes an increase in the consumption of oxygen per liter of ventilation, whereas in this malformation, exercise causes a decline in the oxygen consumption per liter of ventilation.

Intracardiac catheterization studies revealed a higher oxygen content in the pulmonary artery than in the femoral artery. It is probable that the volume of blood which entered the pulmonary artery from the right ventricle was equal to that directed from the left ventricle into the aorta. The extensive intimal changes in pulmonary arterioles appeared to be sufficient to account for the increased resistance in the pulmonary vascular bed.

REFERENCES