Complete Transposition of the Great Vessels

Unusual Longevity in a Case with Subpulmonary Stenosis

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Complete transposition of the great vessels is a congenital abnormality usually incompatible with prolonged survival. About 85 per cent of patients succumb within the first 6 months of life. The following is the report of a case showing unusual survival to the age of 3 years, with clinical improvement and a further 8 years of life following surgical intervention.

Case Report

A male infant was noted to be cyanotic at birth. The first year of life was characterized by choking spells while feeding, delay in growth and development, and recurrent, unexplained fever. At 1 year of age a diagnosis of complete transposition of the great vessels was made. At this time, the oxygen saturation of the systemic arterial blood was only 40 per cent. The hemoglobin concentration of the blood was 14.5 Gm. per 100 ml. and the hematocrit value was 51 per cent. Some improvement in the rate of body development was noted during the second year of life and, when the patient was 3 years of age, operation was recommended.

At the time of operation (performed by Drs. Alfred Blalock and F. C. Spencer) the pulmonary arterial pressure was noted to be significantly lower than the systemic arterial pressure. The aortic arch was on the left side and the right subclavian artery was anomalous, arising as the fourth branch of the arch.

Two procedures were performed. The first was the creation of an end-to-side anastomosis between the right subclavian artery and the right pulmonary artery. The second step of the operation was to create an artificial atrial septal defect.

In the 6-month period after operation mild congestive cardiac failure was evident but this was controlled with digitalis. Following this period, the rate of body growth accelerated and despite some cyanosis with severe exercise, the patient did not limit his activities. When the patient was 11 years old and while playing ball at school, he suddenly became cyanotic and unconscious. He was pronounced dead when brought to the hospital.

Necropsy showed the heart to exhibit the characteristic features of complete transposition of the great vessels associated with a ventricular septal defect (fig. 1), subpulmonary stenosis, and patent ductus arteriosus.

Figure 1

Right ventricle showing large ventricular septal defect (point of arrow) lying postero-inferiorly to the parietal and septal limbs of the crista supraventricularis. Characteristic of complete transposition, the right ventricle (R.V.) gives rise solely to the aorta.
The venous connections with the atria were normal and the aorta arose from the right ventricle, while the pulmonary trunk arose from the left ventricle.

The ventricular septal defect was large, measuring 2.0 cm. in diameter. From the right ventricular view (fig. 2a) the defect lay postero-inferiorly to the parietal and septal limbs of the crista supraventricularis. The posterior wall of the defect was formed by the conjoined tissue of the septaltricuspid and anterior-mitral leaflets.

From the left ventricular aspect the ventricular

**Figure 2**

a. Basal aspect of left ventricle (L.V.). The ventricular septal defect (D.) lies anterior to the mitral valve (A.L. and P.M., anterolateral and posteromedial papillary muscles, respectively, of the mitral valve). The unopened inlet to the outflow tract of the left ventricle (S.P.S.) is markedly stenotic. b. The left ventricle (L.V.), the subpulmonary stenotic tract (S.P.S.), and the pulmonary valve (P.V.) have been opened in conventional manner. Remaining abbreviations as in a.

**Figure 3**

Interior of aortic arch and descending thoracic aorta. The right subclavian artery (R.S.) arises anomalously as the fourth branch of the arch. Beyond the aortic ostium of the patent ductus arteriosus (P.D.) the branches of the descending aorta are wide. It is assumed that the latter were channels for collateral blood supply to the lungs.
septal defect occupied a posterior position, lying anteriorly to the posteromedial papillary muscle of the mitral valve (fig. 2b). Anterior to the ventricular septal defect the inlet to the outflow tract of the left ventricle was found to be narrow, having a diameter of but 9 mm. The pulmonary trunk was thin-walled but of wide diameter (2.8 cm.; ascending aorta 3.6 cm.). The pulmonary valve was bicuspid but seemingly not stenotic.

In the descending thoracic aorta (fig. 3) the ostia of intercostal and other (presumably bronchial) arteries were unusually wide. The largest diameter of these vessels was 3 mm. The patent ductus arteriosus measured 7 mm. in diameter at its aortic end and 4 mm. in diameter at is pulmonary arterial end. Each ventricular wall was hypertrophied, the right measuring 1.4 cm. in thickness, the left, 1.2 cm.

In addition to the findings named, the posterior aspect of the atrial septum showed a large defect measuring 3 by 4 cm. (fig. 4). It was presumed that this opening was the surgically created atrial septal defect. The region of the fossa ovalis lay below the zone of the defect.

The right subclavian artery-right pulmonary arterial anastomosis, which had been performed 8 years before death, was widely patent. The lumen at the anastomosis measured 1 cm. in diameter. Opposite the opening of the anastomosis the lining of the right pulmonary artery was rough, a phenomenon considered to represent multiple “jet lesions.” The observation made at operation that the right subclavian artery arose anomalously as the fourth branch of the aortic arch was confirmed.

Discussion

It will be recalled that the subject of this report was a patient with complete transposition who survived to the relatively uncommon age of 3 years without operation and lived an additional period of 8 years following creation of an atrial septal defect and a subclavian-pulmonary arterial anastomosis.

It is of interest to attempt an explanation for the favorable hemodynamics that allowed this patient to live considerably longer than is average for patients with complete transposition. In attempting this explanation a consideration first will be given to the hemodynamic features assumed to have been present both before and after operation.

In this patient, in the natural state, the subpulmonary stenosis in combination with a
ventricular septal defect seems to have been a favorable factor in causing the pulmonary arterial pressure to be lower than the systemic pressure. This pressure pattern would favor delivery of systemic venous blood from the aorta into the pulmonary arterial system through the bronchial arteries and the demonstrated patent ductus arteriosus (fig. 5a).

The subpulmonary stenosis of the left ventricle lay distally to the ventricular septal defect. This arrangement would have favored a situation in which highly oxygenated blood reaching the left ventricle would preferentially have been shunted into the right ventricle for delivery to the aorta.

Following the operation the favorable direction of blood flow present preoperatively was augmented (fig. 5b). The subclavian-pulmonary arterial anastomosis provided an additional channel for delivery of systemic venous blood into the pulmonary arterial system. The created atrial septal defect represented yet another route, in addition to the ventricular septal defect, for delivery of highly oxygenated blood from the left side to the right side of the heart and ultimately to the aorta.

Summary
A case of survival to the age of 11 years is reported in a patient with complete transposition of the great vessels. Additional malformations in the form of subpulmonary stenosis, ventricular septal defect, and patent ductus arteriosus may have contributed to the unusually long period of survival. Surgical creation of an atrial septal defect and of a subclavian-pulmonary arterial anastomosis when the patient was 3 years old may have augmented the beneficial effects of the existing associated malformations.
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