Treatment of Complete Transposition of the Great Vessels with the Blalock-Hanlon Operation

By John L. Ochsner, M.D., Denton A. Cooley, M.D., Leonard C. Harris, M.D., and Dan G. McNamara, M.D.

TRANSPPOSITION of the great vessels is not a rare anomaly. It accounts for 8 per cent and is the fourth most common of all congenital heart diseases, being surpassed in frequency only by patent ductus arteriosus, ventricular septal defect, and tetralogy of Fallot. Furthermore, transposition is a notorious first cause of death from congenital heart disease in childhood. At the Texas Children's Hospital transposition of the great vessels is the most frequent congenital heart malformation found at autopsy, accounting for 64 (21 per cent) of the 300 deaths from congenital heart disease.

The high mortality from transposition of the great vessels is not usually due to a delay in diagnosis, since the most prominent symptoms, cyanosis, dyspnea, and growth failure usually cause the parents to seek medical help early. Nor is it due to difficulty in diagnosis, which according to Taussig can usually be made clinically with the help of an electrocardiogram and a roentgenogram, although confirmation by angiocardiogram is often desirable. The prognosis of this disease is so grave that the great majority of cases do not survive beyond the first year of life and only the occasional case survives to adolescence or adult life. Blalock and Hanlon found an average life expectancy of 19 months in 123 cases, and if one excludes six of their patients who lived 10 years or longer, the average duration of life was only 5½ months. Keith and co-workers reported that 52 per cent of infants with transposition of the great vessels were dead at 1 month and 86 per cent dead at 6 months, the average length of survival being only 3 months.

The frequency and poor prognosis of this condition have stimulated the surgical development of many palliative and some corrective procedures, but in general the unsatisfactory results have discouraged most clinicians. The purpose of this paper is to present our experience in the management of patients with complete transposition of the great vessels and to report the results of palliative surgery for this disease by the creation of an atrial septal defect by the use of the Blalock-Hanlon technique.

Since success of the various palliative and attempted corrective procedures depends on some alteration of the hemodynamics, it is pertinent to review briefly the physiology of this condition. In utero the origin of the aorta from the right ventricle causes little physiologic disturbance, since the right ventricle serves as a systemic ventricle during fetal life. After birth, in the absence of a shunt between the right and left sides of the heart, two separate circulations exist (fig. 1). Unsaturated systemic venous blood returns to the right atrium and enters the right ventricle and transposed aorta. Saturated blood returns from the lung to the left atrium and recirculates through the pulmonary system via the transposed pulmonary artery. Obviously life cannot be sustained if there is no interchange of blood between these two independent circuits. A shunt, resulting in partial saturation of the systemic blood, must be present for survival; to prevent overloading of the pulmonary or systemic circulations the shunt must be in two directions. The shunt may be bidirectional at one site or in opposite directions at two or more sites. Shunts may

From the Cora and Webb Mading Department of Surgery and Department of Pediatrics, Baylor University College of Medicine, Houston, Texas.

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be present as a ventricular septal defect, patent foramen ovale, and patent ductus arteriosus, and various combinations may be demonstrated at cardiac catheterization.\textsuperscript{17}

Among 64 cases of transposition of the great vessels autopsied at the Texas Children’s Hospital from 1956 to 1960, the most common site of a shunt was through a ventricular septal defect (table 1). Although a patent foramen ovale was present in 25 cases, a true atrial septal defect was encountered only six times. This is significant, since the valvular nature of a patent foramen ovale tends to prevent free bidirectional shunting of blood, and left-to-right shunting of blood is usually retarded. A patent ductus arteriosus, though often present in infants with transposition, does not increase the chance of survival.\textsuperscript{1} The inadequacy of a patent ductus arteriosus or foramen ovale to provide adequate shunting of blood was evident in 10 of our autopsied newborn infants who had only these associated defects. Rare sites of shunting in our series included anomalous venous return from the lungs and aorticopulmonary septal defect (table 1). Dilated bronchial arteries may carry unsaturated blood to the left side of the heart for oxygenation. Communications have been described between the right internal jugular vein and left atrium and between the azygos vein and left atrium, permitting systemic venous blood to be oxygenated by passing through the pulmonary circulation.

The severity of symptoms in a given case is determined by the degree of anoxemia and congestive heart failure. The degree of anox-
emia is firstly dependent on the adequacy of the intracardiac shunt, usually through either a ventricular septal defect, atrial communication, or both. Secondly, it is dependent on the degree of pulmonary vascular resistance. Although pulmonary stenosis was described in more than one third of the cases reported by Becker and Brill, Edwards found it to be uncommon. In our autopsy series pulmonary stenosis occurred in six instances, or 9 per cent of the 64 cases of transposition of the great vessels. A mild-to-moderate pulmonary stenosis or increased pulmonary arteriolar resistance in the presence of an adequate left-to-right shunt tends to improve mixing and thereby increase longevity.

During the past 6 years surgical treatment of the critically ill infant admitted to our hospital with complete transposition of the great vessels consisted in most instances of creation of an interatrial septal defect with use of a modified Blalock-Hanlon technic.

Operative Technic

The anesthetized patient is placed in the left lateral position and a right lateral incision is made through the fifth intercostal space (fig. 2). The lung is retracted and the pericardium is entered anterior and parallel to the course of the right phrenic nerve. Confirmation of the diagnosis may be made by aspirating blood from the aorta and right pulmonary artery and demonstrating blood of higher oxygen content in the pulmonary artery than the aorta. The two major pulmonary veins to the right lung are dissected inside the pericardium and encircled with heavy silk ligatures for traction and temporary occlusion. The right main pulmonary artery is also encircled with a heavy ligature. During the period of temporary occlusion of the pulmonary veins the pulmonary artery must be occluded to prevent engorgement of the lung and development of hemorrhagic edema. A curved vascular clamp is placed across the base of the pulmonary veins at the junction with the left atrium (fig. 2). The pulmonary artery and veins are first occluded with the ligatures. The vascular clamp is closed incorporating a portion of the right atrium. Two incisions are made in the heart, one entering the right atrium anteriorly and the other entering the left atrium posteriorly. The tissue between these incisions, which is attached to the atrial septum, is then grasped with a hemostatic forceps. As the arterial clamp is partially released, the septum is withdrawn and trimmed progressively, a fragment being excised approximately 2 cm. in diameter (fig. 2). Usually the fossa ovalis is pulled into view medially during this maneuver. After the septal fragment is excised, the septum retracts back inside the heart. The two margins of incision are approximated with a continuous 5-0 black silk suture and the occluding ligatures on the pulmonary artery and veins are removed. Usually the period of venous occlusion is brief, being between 6 and 8 minutes, and therefore pulmonary congestion does not occur. The patient’s color and general condition improve immediately. The pericardium is closed loosely. The thoracotomy incision is repaired and intercostal underwater seal drainage is used for 24 hours.

Clinical Material

Over the past 6 years 45 patients with transposition of the great vessels underwent operation for the creation of an atrial septal

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Table 1

<p>| Associated Congenital Defects in 64 Autopsied Cases of Transposition of Great Vessels |
|---------------------------------|----------------|</p>
<table>
<thead>
<tr>
<th>Defect</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventricular septal defect</td>
<td>43</td>
</tr>
<tr>
<td>Isolated ventricular septal defect</td>
<td>20</td>
</tr>
<tr>
<td>Single ventricle</td>
<td>18</td>
</tr>
<tr>
<td>Taussig-Bing anomaly</td>
<td>5</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>31</td>
</tr>
<tr>
<td>Patent foramen ovale</td>
<td>25</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>6</td>
</tr>
<tr>
<td>Coarctation of aorta</td>
<td>7</td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
<td>6</td>
</tr>
<tr>
<td>Aortic arch atresia and hypoplasia</td>
<td>5</td>
</tr>
<tr>
<td>Pulmonary atresia</td>
<td>4</td>
</tr>
<tr>
<td>Mitral atresia</td>
<td>3</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
<td>3</td>
</tr>
<tr>
<td>Anomalous pulmonary venous return</td>
<td>2</td>
</tr>
<tr>
<td>Aorticopulmonary septal defect</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>136</td>
</tr>
</tbody>
</table>
defect by the Blalock-Hanlon technic. There were 33 males and 12 females, a male predominance of 2.75 to 1. A wide variation in age distribution was noted, the youngest child being 26 hours old and the oldest 9 years. The majority of the children, however, were operated upon during the first year of life (table 2). Twenty-two (49 per cent) were less than 3 months of age at the time of surgery.

The prime criterion for selection of a patient for surgical treatment was severe anoxemia. The additional complication of congestive heart failure was not considered a contraindication for operation on the assumption that cardiac failure was accompanied by anoxemia. One patient operated upon at 2 months of age with severe anoxemia and congestive heart failure was still improved 1½ years later. Cases of transposition of the great vessels associated with single ventricle, pulmonary atresia, mitral atresia, or tricuspid atresia were not included in this study because these cases have different hemodynamics, functioning as a single ventricle.

Results

The over-all survival rate of 45 patients in whom the creation of an atrial septal defect by the Blalock-Hanlon technic was performed was 67 per cent (table 3). Fifteen (33 per cent) of the 45 cases in this study have died since operation. Thirteen (29 per cent) succumbed in the immediate postoperative period, whereas two died after discharge from the hospital, having survived for periods of 2 months and 9 months, respectively. During the first 3 years of this study only 9 of 17 patients (53 per cent) survived the early postoperative period and two more subsequently died, the mortality rate for this group being 59 per cent. In contrast there were only five deaths in the 28 patients operated upon during the last 3 years (a mortality rate of 18 per cent). One of these five patients died 7 days after surgery without an autopsy but was known to have a birth injury with a fractured skull.

Blood and ear oxygen saturations were determined before and after surgery in 13 cases (table 4). All except two showed a significant improvement in saturation and the average increase in the 13 cases was 17.5 per cent. Four of the cases who had preoperative determinations of oxygen saturation died after surgery. Only two of the four showed a rise in oxygen saturation of more than 10 percentage points, and one of these, who was improved by surgery died 8 months afterwards from a cerebral hemorrhage.

The majority of patients had significant and often striking symptomatic improvement following creation of the atrial septal defect. They have shown a decrease in cyanosis and an increase in exercise tolerance. A few were able to live an almost normal life, engaging in most activities enjoyed by other children of equal age.

Discussion

Complete correction of transposition of the great vessels is obviously the ideal treatment and continues to provide a challenging technical and physiologic problem. Surgical correction could be achieved either by transposing the aorta and pulmonary artery or by converting the systemic and pulmonary veins to conform with the arterial transposition.

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Figure 2

Drawing showing technic of creation of interatrial septal defect in treatment of complete transposition of great vessels. In (a) an incision is made in the right fifth intercostal space. In (b) a curved clamp is placed across base of the right pulmonary veins and right atrium. Dotted lines show position of incisions into right and left atria. In (c) and (d) portion of atrial septum is withdrawn from the heart as the clamp is partially released. Fossa ovalis is visible at apex of septal specimen. In (e) the atrial incision is closed with a continuous fine silk suture.

The latter method has a distinct advantage in eliminating the necessity of transposing the coronary blood supply, which is derived from the aortic annulus at its origin from the right ventricle. Experimental and clinical efforts toward complete correction of this anomaly in our hospital were directed toward atrial repositioning, excising the interatrial septum and replacing the septum with a spiraled cloth prosthesis. Results with this technic were not satisfactory. Recently Senning reported a unique and ingenious method of transposing the two atria during cardiopulmonary bypass. He reported one clinical success with this method, and Kirklin and associates have recently had four patients survive this operation among 11 attempted total corrections. Unfortunately, the complicated nature of a totally corrective operation may limit the usefulness of this procedure in the gravely ill newborn infant.

Since most of the patients with this malformation die before 6 months of age, and because of the technical difficulties and detrimental physiologic alterations associated with open-heart surgery in infants of this age, a palliative surgical procedure of less magnitude seems desirable. On the basis of our recent experience with 82 per cent survival in patients in whom an adequate atrial septal defect was created, this palliative treatment of complete transposition of the great vessels appears to
be satisfactory. At the same time, the presence of an atrial septal defect will not hinder an intraatrial complete repair at a later date when the patient reaches a more optimum size and general physical state for radical repair.

Certain recommendations for management of these cases seem justified at this time. Creation of an interatrial septal defect by the Blalock-Hanlon technic should be employed in all severely anoxicem infants with complete transposition of the great vessels. Angiocardiography done in the lateral position is a useful method of demonstrating the absence of an adequate interatrial communication. Lack of early opacification of the left atrium provides the indication for creation of an adequate interatrial communication. The larger the atrial defect produced the more efficient will be the mixing of the unoxygenated and oxygenated blood. The size of the defect created will naturally be limited by the size of the heart; however, even in an infant less than 3 months of age the defect which is created should be 2.0 cm. in diameter.

Summary

Transposition of the great vessels is the most frequent cause of death from congenital heart disease in childhood. The high incidence and poor prognosis of this condition have stimulated the development of many palliative and corrective surgical procedures, but in general their results have been discouraging.

During the past 6 years at the Texas Children’s Hospital 45 patients with transposition of the great vessels have undergone palliative operation by the creation of an atrial septal defect, using the Blalock-Hanlon technic. The prime criterion for the selection of a patient for surgical treatment was severe anoxemia with or without associated congestive heart failure. The majority of cases were under 1 year of age and 49 per cent less than 3 months of age at the time of surgery.

Among 28 patients operated upon during the past 2 years 23, or 82 per cent, survived operation with general improvement. Use of this palliative operation is recommended for the small infant or critically ill patient, and totally corrective procedures should be reserved until conditions are more favorable for survival.

References


TENTH OBSERVATION
Of Scirrhus of the Lungs, and its Symptoms

XXXVIII. By scirrhus of the lungs I mean the degeneration of the natural spongy substance of the organ into an indolent, fleshy mass.

A portion of sound lung swims in water, but this carniform degeneration sinks. There is often observed a vast difference in the character of these scirrhi, in respect of hardness, color, and component parts. Thus in inflammatory diseases of the chest proving fatal on the fifth, seventh, or ninth day, the lung is very often found so completely gorged with blood as to resemble liver in every respect, both as to colour and consistency. . . .

XXXIX. The presence of scirrhus of the lungs, in its primary, unsoftened condition, may be suspected from the following signs:

Together with diminution or entire loss of the natural sound over the affected part, there is an infrequent cough without any expectoration, or with only a scanty excretion of viscid and crude sputa. During a state of quiescence there is nothing to be observed much amiss, either in the condition of the pulse or respiration; but upon any considerable bodily exertion, or after speaking for some time, these persons become speedily exhausted, anxious, and breathless, and complain of a sense of dryness and roughness in the throat. All the above symptoms are more severe in proportion as the scirrhus is more extensive.—From On Percussion of the Chest. Published in 1761. Translated by John Forbes, M.D. In: Classics of Medicine and Surgery. New York, Dover Publications, Inc., 1959, p. 136.
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