The management of complete transposition of the great arteries remains one of the most challenging problems in the operative treatment of congenital heart disease. Although a completely corrective operation is feasible,\(^1,2\) in most centers the mortality associated with this procedure is prohibitive, and it is likely to remain high among the seriously ill infants who form the vast majority of patients with this malformation. While total correction is technically less difficult in older children, the presence of serious and possibly irreversible changes in the pulmonary vascular bed may limit the effectiveness of the procedure in this age group as well. Instead of a corrective operation, therefore, surgeons have most commonly utilized a palliative procedure for complete transposition of the great arteries; these operations include creation or enlargement of an atrial septal defect in order to improve mixing between the two circulations,\(^3\) establishment of a systemic-pulmonary artery anastomosis or superior vena cava-pulmonary artery anastomosis to increase pulmonary blood flow,\(^4,5\) banding of the pulmonary artery or closure of a patent ductus arteriosus or both to reduce pulmonary blood flow,\(^6,7\) inferior vena caval-left atrial anastomosis with transplantation of the right pulmonary veins,\(^8\) and various combinations of these procedures.\(^9\)

The purpose of this report is to describe the rationale of a new palliative operative procedure for complete transposition of the great arteries, creation of a systemic-pulmonary arterial shunt combined with ligation of the main pulmonary artery, and to present preliminary results of the application of this operation. This new procedure is based on the consideration that the circulatory derangement in patients with persistent truncus arteriosus with normal, or nearly normal, pulmonary blood flow is significantly less serious than that which exists in patients with transposition of the great arteries. In the latter anomaly, the oxygen content of systemic arterial blood is usually less than that of pulmonary arterial blood, while in persistent truncus arteriosus the oxygen saturation of the systemic and of the pulmonary arterial blood is essentially identical. The operative procedure described in this report converts transposition of the great arteries to a functional equivalent of truncus arteriosus. Ligation of the main pulmonary artery directs highly oxygen saturated left ventricular blood across the ventricular septal defect into the aorta, and the oxygen saturation of systemic arterial blood is thus increased. The creation of a systemic-pulmonary arterial anastomosis of appropriate size provides a controlled and normal, or nearly normal, inflow of blood into the pulmonary arteries. Since pulmonary arterial blood contains a larger fraction of right ventricular blood after ligation of the pulmonary artery, its oxygen saturation falls. To prevent interruption of pulmonary blood flow during operation, the anastomosis is established before the main pulmonary artery is ligated (figs. 1 to 4).
At catheterization, and roentgenograms revealed 54%. The electrocardiogram revealed a ventricular septal defect, pulmonary atresia, and origin of the aorta from the right ventricle; the pulmonary arteries filled through a small patent ductus arteriosus. The patient’s clinical condition continued to deteriorate and accordingly, at the age of 12 months, a right subclavian-pulmonary artery anastomosis was constructed. A continuous murmur was heard postoperatively, and the child’s color and general condition have shown distinct improvement. Three months after operation the concentration of hemoglobin had fallen to 14.5 g./100 ml.

S. M. (no. 03-95-02), a 4½-year-old girl, had been cyanotic at birth. She was admitted to the National Heart Institute at the age of 3 years because of tachypnea, increasing cyanosis, and poor growth. Marked clubbing, cyanosis, cardiomegaly, and a continuous murmur were noted. The hemoglobin was 19.6 g/100 ml and the hematocrit was 72%. Right axis deviation, right atrial enlargement, and right ventricular hypertrophy were evident from the electrocardiogram, while the chest roentgenograms showed generalized cardiomegaly, a right aortic arch and descending aorta, and diminished pulmonary vascularity. The systemic arterial oxygen saturation was 51% and the pressures in the two ventricles. A right ventricular angiocardiogram revealed a ventricular septal defect, pulmonary atresia, and origin of the aorta from the right ventricle; the pulmonary arteries filled through a small patent ductus arteriosus. The patient’s clinical condition continued to deteriorate and accordingly, at the age of 12 months, a right subclavian-pulmonary artery anastomosis was constructed. A continuous murmur was heard postoperatively, and the child’s color and general condition have shown distinct improvement. Three months after operation the concentration of hemoglobin had fallen to 14.5 g./100 ml.

Prior to clinical application of this concept, it was deemed desirable to determine the circulatory state which might be achieved by this operative procedure. Accordingly, systemic-pulmonary arterial anastomoses were established in two patients with transposition of the great arteries and pulmonary atresia.

Report of Cases

P. K. (no. 05-96-20), a 1-year-old girl, had had a heart murmur and faint cyanosis at birth. She was referred to the National Heart Institute at the age of 5 months because of increasing cyanosis, poor weight gain, and tachypnea. On examination she exhibited marked cyanosis and clubbing, a short, grade-II/VI systolic ejection murmur along the left sternal border and moderate hepatomegaly. The value for hemoglobin was 16.2 g/100 ml and the hematocrit reading was 54%. The electrocardiogram showed right axis deviation and right ventricular hypertrophy, and roentgenograms revealed hypovascular lungs. At catheterization, an atrial septal defect was

Diagrammatic representation of the central circulation in transposition of the great arteries and ventricular septal defect. Symbols: SVC, superior vena cava; IVC, inferior vena cava; RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle; Ao, aorta; PT, pulmonary trunk; RPA, right pulmonary artery; LPA, left pulmonary artery; INN.A., innominate artery; LCCA, left common carotid artery; LSA, left subclavian artery.

Figure 1

Diagrammatic representation of the central circulation in transposition of the great arteries and ventricular septal defect after creation of a subclavian-left pulmonary artery anastomosis.

Figure 2
OPERATION FOR COMPLETE TRANSPOSITION

The favorable clinical results achieved in these two patients are not surprising, since others have reported on the value of systemic-pulmonary artery shunts in patients with transposition of the great arteries and pulmonary atresia. However, the preliminary experience with these patients was of considerable interest since the functional arrangement of their central circulations post-operatively was essentially identical to that which would result from ligation of the main pulmonary artery and creation of an aortopulmonary shunt in a patient with transposition of the great arteries without pulmonary atresia. Accordingly, this operation was carried out on the next such patient whom we encountered.

J. McN. (no. 03-98-17) had a heart murmur and cyanosis at birth. He was admitted to the National Heart Institute at the age of 1½ years because of poor growth, tachypnea, and progressive cyanosis. The concentration of hemoglobin was 19 g/100 ml; the hematocrit value was 70%, and the arterial oxygen saturation, 62%. At catheterization, an atrial septal defect was crossed and cineangiograms demonstrated an atrial septal defect, a large ventricular septal defect, and complete transposition of the great arteries. There was a moderate increase in pulmonary vascularity. During the next 2½ years motor development lagged and polycythemia progressed.

The onset of seizures and focal neurological signs prompted the child's second admission at the age of 4 years. A left parietal brain abscess, containing hemolytic microaerophilic alpha streptococci, was successfully aspirated. After an extended course of antibiotic therapy his neurological signs regressed and his condition stabilized.

Comment

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At the age of 4½ years he was readmitted and was found to be intensely cyanotic and underdeveloped. Cardiomegaly and a grade III/VI systolic ejection murmur along the upper left sternal border were noted. The concentration of hemoglobin was 21.6 g/100 ml, and the hematocrit reading was 79.5%. The electrocardiogram showed right axis deviation, right atrial enlargement, and right ventricular hypertrophy. Generalized cardiac enlargement, a right aortic arch and descending aorta, and increased pulmonary vascularity were noted on the chest roentgenograms (fig. 5A).

The arterial oxygen saturation was 57.5%, and at catheterization the pressures in the two ventricles were equal. The catheter crossed an interatrial communication. The left atrial oxygen saturation was 98.2%. A right ventricular angiocardiogram again confirmed the presence of ventricular septal defect and transposed great arteries. In addition, discrete subpulmonic stenosis, that is, subvalvular obstruction to left ventricular outflow was present (fig. 6).

In view of the patient’s extreme cyanosis and functional disability, he was operated upon on March 18, 1965. The main pulmonary artery, which was 9 mm in diameter, lay posterior and to the left of the aorta, and a systolic thrill was palpable in it. The pressure in the right ventricle was 88/0 mm Hg, and the mean pulmonary arterial pressure was somewhat elevated, 24 mm Hg.

An end-to-side left subclavian-pulmonary artery anastomosis was constructed. The main pulmonary artery was then completely occluded with a snare. During the occlusion the heart action remained good and the color of the heart and other tissues improved distinctly. The mean arterial pressure was 80 mm Hg and the mean pulmonary arterial pressure 24 mm Hg.

Figure 5

Figure 6
Lateral view of angiocardiogram with right ventricular injections obtained preoperatively. RV, right ventricle; LV, left ventricle; Ao, aorta; PA, pulmonary artery; PV, pulmonary valve; AV, aortic valve; S.P.O., subpulmonic obstruction.
pulmonary artery pressure distal to the occlusion was normal, 11 mm Hg. The pulmonary artery was then ligated at its base with umbilical tape. After occlusion there was no change in left ventricular pressure.

The patient had a benign postoperative course. Arterial oxygen saturation had risen to 75.3% prior to discharge and was 76.9% 2 months later. Striking improvement in the patient's color and exercise tolerance was noted immediately after operation and this improvement has been sustained. The chest roentgenogram no longer shows pulmonary hypervascularity (fig. 5B). At present, January 1966, the child has gained 5½ pounds, has learned to walk for the first time, and can now run and play. Clubbing and cyanosis have improved markedly.

Discussion

The clinical improvement exhibited by all three patients supports the proposed concept that patients with functional truncus arteriosus and normal, or nearly normal, pulmonary blood flow have significantly less functional derangement of the circulation than do patients with complete transposition of the great arteries. The courses of these patients suggest that creation of a systemic-pulmonary arterial anastomosis and occlusion of the main pulmonary artery may prove to be a useful palliative procedure in some patients with transposition of the great arteries and increased pulmonary blood flow.

Two considerations are pertinent to the selection of patients for this operation. First of all, it is essential that a large ventricular septal defect be present, so that left ventricular outflow will not be impeded when the pulmonary artery is ligated. Secondly, the operation is not contraindicated by either the presence or absence of pulmonic or subpulmonary stenosis. A more impressive increase in arterial oxygen saturation might be anticipated in patients without than in those with obstruction to left ventricular outflow, since ligation of the main pulmonary artery would divert a larger fraction of oxygenated blood from the left ventricle across the ventricular septal defect and into the aorta. However, ligation of the pulmonary artery should result in some elevation of the arterial oxygen saturation and clinical improvement even in patients with moderately severe obstruction to left ventricular outflow. Ligation may be expected to elevate arterial oxygen saturation to a higher level than banding the pulmonary artery, since it will divert all of the oxygenated blood from the left ventricle across the ventricular septal defect and into the aorta.

The specific method utilized for establishing the systemic-pulmonary artery shunt will primarily depend upon the size of the patient. In older children, as in patient J. McN., a subclavian artery-pulmonary artery shunt would appear to be the procedure of choice, while in infants below the age of 1 year an anastomosis between the descending aorta and the pulmonary artery or between the ascending aorta and the right pulmonary artery might be more appropriate. In any event, the presence of a patent anastomosis must be clearly apparent before the pulmonary artery is ligated. Measurement of pressures in the left ventricle and pulmonary artery during temporary occlusion of the pulmonary artery ensure that the ventricular septal defect is large enough to decompress the left ventricle, and that the shunt is of appropriate size to produce normal or near normal pulmonary artery pressure. It is generally recognized that an anastomosis between the end of the main subclavian artery and the side of the pulmonary artery delivers a flow of shunted blood approximately equal to systemic flow when pulmonary vascular resistance is normal. Similarly, an aortic-pulmonary anastomosis 5 mm in diameter will usually have the same effect in an infant.

Theoretically, one of the most important potential benefits which may accrue from the palliative procedure described is the reduction of pulmonary artery pressure, which in turn may arrest the progression, or hopefully even allow the regression of the pulmonary vascular changes which ultimately develop in patients with complete transposition of the great arteries and increased pulmonary blood flow. If such favorable changes occur and are accompanied by growth of the child, a completely corrective procedure as described by Senning or Mustard might well be considered. Such an operation would, of course,
require reestablishment of continuity of the main pulmonary artery, a procedure which could be accomplished by techniques similar to those utilized at the time of complete correction in children with ventricular septal defect who have been initially treated by pulmonary artery banding.

Although both the theoretical considerations and the preliminary clinical experience presented herein suggest that ligation of the main pulmonary artery and creation of a systemic-pulmonary shunt may be a useful palliative procedure in selected patients with transposition of the great arteries, ventricular septal defect, and increased pulmonary blood flow; only further clinical trials will determine the ultimate usefulness of this procedure.

**Summary**

A new palliative operation is suggested for certain patients with transposition of the great arteries and ventricular septal defect. A systemic-pulmonary artery shunt is created, and the main pulmonary artery ligated. The operation converts transposition of the great arteries to a functional equivalent of truncus arteriosus with normal or nearly normal pulmonary blood flow, a malformation in which the functional derangement of the circulation is not as severe as it is in transposition. The hemodynamic consequences of the procedure were initially evaluated in two patients with pulmonary atresia and transposition, and both improved after shunts were created. The procedure was then carried out in a patient with transposition of the great arteries, ventricular septal defect, mild subpulmonic stenosis, and increased pulmonary blood flow. Postoperatively, the patient has exhibited considerable clinical improvement and significant elevation of arterial oxygen saturation.

**References**

Ligation of the Main Pulmonary Artery and Systemic-Pulmonary Arterial Anastomosis: A New Palliative Operation for Complete Transposition of the Great Arteries
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