Experience with Anastomosis of Superior Vena Cava to Pulmonary Artery (Glenn Procedure)

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Certain children with cardiac malformations and increasing cyanosis require immediate palliation. They cannot wait until they reach the minimum body size required for a safe corrective operation with perfusion or until a corrective procedure is devised for their lesions. Systemic to pulmonary artery anastomosis has provided palliation for many of these patients. Anatomical findings, however, frequently exclude a subclavian to pulmonary artery anastomosis or lead to its occlusion or inadequacy as the child grows. Glenn et al.\textsuperscript{1, 2} suggested the physiological superiority of the anastomosis of the systemic venous system to the pulmonary artery in the absence of pulmonary hypertension. The reported collective experience following superior vena cava to pulmonary artery anastomosis is not only small, but has a relatively short follow-up. We have summarized our own experience with 15 cases, describing our operative technique and reviewing the indications for perfusion during the anastomosis, the operative mortality, and the late results of treatment. Suggestions for improved anesthetic management of these difficult cases have also been included.

Patients

Eight boys and seven girls with congenital heart disease and severe cyanosis have had anastomosis of the superior vena cava to the pulmonary artery. A cardiac abnormality was recognized in 10 at birth, and in all before the first birthday. Their ages, at the time of operation, ranged from 2 months to 14 years. Eleven children were between the ages of two and eight years. Nine of these children had ten previous operations, nine being Blalock anastomoses and one a Potts anastomosis. All operations except one were done when the patient was under the age of 30 months. Eight of these procedures were followed by obvious clinical improvement. In two patients, the postoperative course and the absence of a continuous murmur suggested early closure of the anastomosis. At the time of the superior vena cava to pulmonary artery anastomosis, a continuous murmur suggested that seven of the previous anastomoses were patent.

There were six cases of tetralogy with hypoplasia or atresia of the main pulmonary artery, three cases of transposition with pulmonic stenosis, three cases of tricuspid atresia or stenosis, two Ebstein-like anomalies, and one atrioventricular canal with pulmonic stenosis. In every case, the correct diagnosis was made preoperatively. Three of these patients had associated congenital anomalies: one tetralogy case had hypoplasia of the left pulmonary artery and lung; situs inversus accompanied one transposition of the great vessels with pulmonic stenosis and the case of atrioventricular canal defect.

The symptoms and physical findings elicited were as follows: cyanosis, 15; fatigue and limited activity, 14; clubbing, 14; cardiomegaly, 11; dyspnea, 10; underdevelopment, 10; hypercyanotic spells, 9; squatting, 5; cardiac failure, 2; and hemiparesis from cerebral thrombosis, 1.

Laboratory Data

The preoperative hemoglobin, hematocrit, and red blood cell values were elevated, as expected. The hemoglobin values ranged from 17.4 to 25.0 Gm. per cent, with an average of 20.5. The hematocrit values in 13 cases ranged from 60 to 80 per cent, with an average value of 67 per cent. In 10 cases, the red blood cell count ranged from 5.54 to 10.20 million, with an average of 8.05 million.

Routine posteroanterior roentgenograms frequently suggested the diagnosis. Cardiomegaly was seen in 11 cases. Thirteen cases revealed a diminished vascularization of the lungs—the two exceptions being transposition of the great vessels with pulmonic stenosis. Cineangiograms were completed in eight cases. They were diagnostic in seven—the exception was a case of Ebstein-like deformity. It was the primary diagnostic feature in the atrioventricular canal defect, tetralogy, transposition, and tricuspid atresia cases.
Electrocardiograms

The electrocardiograms were most useful in diagnosing tricuspid atresia, showing in every case P. pulmonale and left ventricular hypertrophy. In other conditions it was mainly confirmatory.

Cardiac Catheterization

Cardiac catheterization was not attempted or was incomplete in eight cases. Five cases of tetralogy were studied. Four of the five demonstrated an increase in the oxygen saturation in the right ventricle. All five had right ventricular systolic pressures over 100 mm. Hg. In only two cases did the catheter enter the pulmonary artery, demonstrating a gradient of 90 and 100 mm. Hg, respectively.

Anesthesia

Premedication

Ten patients received morphine, 1 mg. per 5 Kg. body weight, and five patients received meperidine, 1 mg. per pound of body weight, as premedication. There was no respiratory depression, but three of the five patients who received meperidine sustained a 50 mm. Hg fall in blood pressure upon induction. Thirteen patients received atropine, 0.1 mg. per 10 lbs. body weight, and 11 suffered tachycardia of 120 to 160 before induction, obviously compromising cardiac reserve. Recent evidence suggests that this dose of atropine precipitates arrhythmia rather than preventing it. Five patients received pentobarbital, 1 to 2 mg. per pound, and their induction was smooth.

Maintenance Agents

Halothane was used in all cases, usually at a concentration of 0.1 per cent, and never more than 0.5 per cent. Nitrous oxide, while never the primary agent, was used as a fluorothane vehicle in all cases. Oxygen concentration was never less than 50 per cent, and it was increased whenever cyanosis deepened. Ether was used in only three cases and with fluorothane. It was never an azo trope, and was apparently well tolerated.

Relaxants

Succinylcholine chloride aided 50 per cent of the intubations. In four cases, a 0.2 per cent intravenous drip of succinylcholine chloride permitted less general anesthesia and was associated with an elevation in the blood pressure, but not with apnea.

Operation

Thirteen Glenn operations were done by the closed technique, and two using total body perfusion with the Clark heart-lung machine. Perfusion was first used in this series in an attempted correction of an Ebstein-like deformity. After the "open" operation failed, a superior vena cava to right pulmonary artery anastomosis was completed. Certainly the long perfusion and attempted correction of the deformed tricuspid valve was responsible for this death, and not the superior vena cava to right pulmonary artery anastomosis. The other case in which perfusion was used was in a child with tetralogy of Fallot with hypoplasia of the left lung and left pulmonary artery.

Upon induction of anesthesia, a plastic cannula was inserted into the left saphenous vein; plasma was administered through the cannula to replace blood loss. We have not withdrawn blood preparatively to reduce the hematocrit levels. The patients were placed on the left side in a posterior oblique position. Obviously, the right side was down in cases of situs inversus. The incision, beginning at the sternal border in the inframammary fold, was carried around the right anterolateral hemithorax into the intrascapular space. In cases of situs inversus, the same incision was used to open the left thorax. The chest was entered through the fourth intercostal space. The phrenic nerve was mobilized and retracted anteriorly. Meticulous hemostasis was required about the hilum of the lung in dividing the profuse bronchial collateral circulation. The pericardium was opened by a longitudinal incision that allows adequate exploration of the heart and divides the pericardial reflection on the superior vena cava (fig. 1A).

The azygos vein was divided and the superior vena cava mobilized extensively. Next, the right (or left) pulmonary artery was mobilized from its bifurcation to its division within the hilum. The pulmonary artery was ligated and transfixed with silk just distal to the bifurcation (fig. 1B and C). The superior vena cava was grasped with fine forceps at each end of the proposed anastomosis (usually at the azygos vein insertion). It was rotated in a clockwise fashion, posterior to anterior. This rotation was important for positioning the anastomosis without torsion and/or kinking. The section between the forceps was excluded with a vascular clamp that does not completely obstruct flow in the vena cava (fig. 1D). An ellipse of the vena cava was excised. The anastomosis to the distal most end of the pulmonary artery was completed with alternating interrupted end-on and mattress sutures of 5-0 black silk or 6-0 merselene. The clamp and occluding distal ligatures were removed, allowing blood to flow through the anastomosis.

The superior vena cava was divided between vascular clamps placed between the anastomosis and the heart. Each end was closed with silk sutures (fig. 1E). The pericardium was closed loosely. The chest was drained with underwater drainage and closed.

Results

There were three hospital deaths, represent-
ing a 20 per cent operative mortality. One death followed a long perfusion in which the superior vena cava–pulmonary artery anastomosis was done as a desperate measure when a correction of a tricuspid valve lesion failed. This child had an Ebstein-like deformity with transposition of the atrioventricular valves, stenosis of a low, malpositioned right bicuspid valve, and an ostium seconum defect. The right atrioventricular valve was elevated to the annulus, and the atrial defect closed. Her pressure could not be maintained without perfusion and the atrial septal defect was opened. Glenn anastomosis was performed in desperation. The total perfusion time was five and a half hours, two and a half merely being supportive. She never regained consciousness, and died 12 hours later. Cerebral edema, neuron necrosis, and alveolar hemorrhage were the causes of death.

The second child also had an Ebstein-like deformity. She developed bradycardia and cardiac arrest during the anastomosis. Her heart could not be resuscitated. Her heart weighed 1,210 Gm., with right ventricular dilatation, left ventricular hypertrophy (9-
mm. thick), tricuspid valve deformity and insufficiency, and an atrial secundum defect. It is believed that her huge heart, compressing the left lung during the anastomosis (when the oxygen saturation of the blood is low), created lethal hypoxia.

The third death occurred in a small child with a tetralogy of Fallot. She had had two previous Blalock anastomoses, only the left being patent. Her hemoglobin was 25 Gm. per cent. She, too, developed bradycardia and cardiac arrest during the anastomosis. Although a normal beat was restored, she never regained consciousness and died 24 hours later. No autopsy was permitted; however, the clinical picture suggested cerebral thrombosis.

During the procedure, hypotension was experienced in 11 cases. Five patients developed bradycardia and cardiac arrest; three of them expired. Bradycardia and cardiac arrest occurred in the very cyanotic children.

In the postoperative hospital course, all of the survivors demonstrated an improvement in color. Their hemoglobin values dropped from 3 to 6.5 Gm. per cent, the average fall being 5.2 Gm. per cent. The postoperative complications encountered were edema of the face in three patients, atelectasis in two; staphyloccocal pneumonia in one; cardiac arrest in one, and atrioventricular dissociation with a bundle-branch block in one. All except the last responded to therapy, and the complication in this case was not disabling. The edema of the face was minimal, transient, and apparently less than is frequently reported.

Follow-up

This follow-up is brief, ranging between one and three years. There have been two deaths after discharge from the hospital. One child died two weeks after discharge. This three-month-old patient had tricuspid atresia with an atrial septal defect, endocardial fibrosis, and multiple neurological defects. The anastomosis was small, and we considered this a technical failure.

The second child, with the atrioventricular canal defect, was readmitted four months later with congestive failure. While the initial response to medical therapy was gratifying, he became more refractory to treatment and died one year postoperatively. His atrioventricular defect was really a cor biloculare associated with transposition of the great vessels and pulmonic stenosis. The heart was enormous, the right ventricle being 16-mm. thick and the left 6-mm. There was one large atrioventricular valve with a mural thrombus. Pulmonary infarcts were seen in the right lower lobe.

Of the 10 remaining survivors, all have maintained their initial improvement in color. In all there has been a progressive increase in exercise tolerance and a reduction in dyspnea. There has been no recurrence of squatting or hypercyanotic spells. We have repeated the laboratory studies on nine children. The hemoglobin values are below the preoperative levels by from 0.6 to 7 Gm. per cent, with an average reduction of 3.2 Gm. per cent. The hematocrits have fallen 7 to 26 per cent since operation, with an average reduction of 18 per cent on comparison with preoperative figures. The change in all except one was dramatic. This exception was a small child, two years and ten months old, with tetralogy of Fallot and hypoplasia of the left pulmonary artery and lung.

Discussion

Subtracting the death following revision of the tricuspid valve, which cannot be attributed to the Glenn anastomosis, leaves an operative mortality of 14 per cent. This mortality reflects poor myocardial function and hypoxic metabolic abnormalities rather than difficult operative techniques.

To our knowledge, this is the first report of using total body perfusion to accomplish this anastomosis. The anatomical necessity (hypoplasia of the left lung and pulmonary artery) was obvious. The high incidence of bradycardia and cardiac arrest during the anastomosis suggests the further use of partial or total body perfusion to perform the anastomosis. The already low oxygen saturation of the arterial blood is dangerously reduced during the anastomosis. Further preoperative

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study must delineate when this reduction is lethal and perfusion is necessary.

The margin of safety is extremely narrow in patients of this type and, therefore, anesthetic agents which may further reduce myocardial function or cause acidosis are contraindicated. Meperidine is a particularly dangerous agent, in our experience, as shown by the associated fall in blood pressure. Morphine and pentobarbital produced no untoward reactions and are the premedications of choice. Atropine, when given, should be used in small doses, 0.04 to 0.05 mg. per 10 lbs. body weight, to avoid the tachycardia seen in 11 of our cases. This dose will adequately suppress salivation.

All agents commonly used for maintenance of general anesthesia depress cardiac output and alter tissue metabolism, producing acidosis. Obviously, it is desirable to give less of these agents. All maintenance agents were administered by controlled respiration, with a Bennett Respirator, through a closed system with endotracheal intubation. Succinylcholine was used to control movement, reducing the need for general anesthesia. There were no harmful effects, and blood pressure was maintained in the normal range. Local or regional anesthesia has been used in other intrathoracic procedures with the elimination of general anesthesia. Its use in cyanotic patients is being explored, and will be the subject of a future presentation.

Summary

Fifteen children with uncorrectable cyanotic heart disease were treated with the Glenn procedure. The hospital mortality was 20 per cent. Total and partial body perfusions were used in two cases to accomplish the anastomosis. The indications for perfusion have been discussed. Modifications in anesthesia were presented to reduce the high incidence of hypotension, bradycardia, and cardiac arrest.

References

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