Clinical Experience With the Use of a Valve-Bearing Conduit to Construct a Second Left Ventricular Outflow Tract In Cases of Unresectable Intra-Ventricular Obstruction

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Two patients, ages 7 and 17, with unresectable obstructions within the left ventricular cavity, have been managed by interposing a conduit bearing a porcine aortic valve between the apex of the left ventricle and the infra-renal abdominal aorta. The younger child had idiopathic hypertrophic subaortic stenosis (IHSS) recognized in infancy. At age three, a right ventricular myomectomy and a trans-aortic left ventricular myotomy were performed. Symptoms were progressive with congestive failure, diaphoresis, syncope, and angina pectoris. Following construction of a second left ventricular outflow tract with relief of intraventricular obstruction, the patient has become asymptomatic. The second patient has fibrous tunnel obstruction of the left ventricular outflow tracting providing a 100 mm Hg gradient. Fibrous tissue was resected in part through the trans-aortic route, and a second outflow tract was constructed. A postoperative cardiac catheterization revealed an obliteration of the previous intraventricular gradients and an equal distribution of left ventricular output through the two available outflow tracts. She remains asymptomatic.

TWO CHILDREN, age 7 and 17, with severe forms of intra-cavitary left ventricular obstruction, have been managed by the surgical creation of a second left ventricular outflow tract. A polyester woven prosthesis bearing a glutaraldehyde-treated porcine aortic valve was inserted between the apex of the left ventricle and the infra-renal abdominal aorta. The alternative left ventricular outflow tract located proximal to the site of obstruction successfully removed systolic pressure gradient from the cavity of the left ventricle. Similar procedures have previously been performed for relief of obstruction at the aortic valve.

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Case Reports

Case 1. A 7-year-old boy was first seen at the St. Louis Children's Hospital at the age of 16 months with congestive heart failure. Cardiac catheterization revealed hypertrophic muscular cardiomyopathy providing obstruction in the mid portion of both the right and left ventricles. A gradient of 119 mm Hg was recorded within the right ventricle and a gradient of 57 mm Hg was recorded in the left ventricle. Over the next two years, frequent episodes of pneumonia and congestive heart failure was treated with antibiotics, digitalis, diuretics, and propranolol. At the age of three, because of a continuing deterioration in his clinical condition, a second cardiac catheterization was performed. At this time, the systolic gradient in the left ventricle was noted to have increased and measured 100 mm Hg. In January of 1971, an operation was performed under cardiopulmonary bypass and at moderate hypothermia. Obstructing muscle was removed from the right ventricle and a trans-aortic left ventricular septal myotomy was performed. The postoperative course was not complicated, but the patient continued to have evidence of severe cardiac decompensation and was re-admitted to the hospital one and one-half months after surgery to be re-evaluated. A third cardiac catheterization revealed that the right ventricular gradient had been reduced to a level of 14 mm Hg, but the systolic gradient in the left ventricle remained at 100 mm Hg. His electrocardiogram demonstrated premature ventricular contractions and evidence of right bundle branch block. There was a vascular re-distribution with excessive blood flow to the upper lobes and persistent cardiomegaly was noted on his chest film. There was a progressive decrease in his exercise tolerance. A fourth cardiac catheterization was performed in 1975 (Fig. 1) which revealed that the peak systolic right ventricular gradient was 15 mm Hg and that the left ventricular systolic gradient measured 80 mm Hg. Profound cardiac hypertrophy was demonstrated

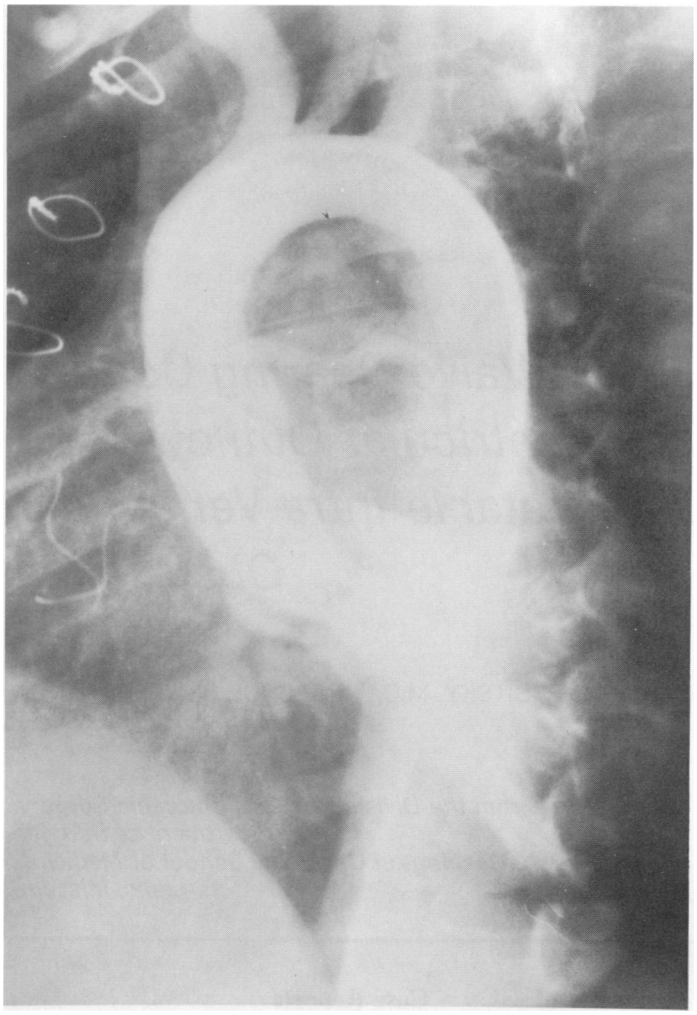

FIG. 1. Case 1. Left ventriculogram obtained between second and third operations. The configuration of the ventricle is typical for severe familial asymmetrical septal hypertrophy. Thickness of the myocardium is demonstrated by the large distance between coronary arteries and the ventricular cavity. An intracavitary gradient of 100 mm Hg in systole was recorded.

at this study with marked diminution in the size of the left ventricular cavity. No change in the left ventricular gradient could be demonstrated at the time of catheterization with a test dose of intravenously injected propranolol. Because of his continuing clinical deterioration, he was transferred to another institution in the hope of having a second left ventricular myotomy performed. At that institution, echocardiographic and electrocardiographic examinations of parental hearts demonstrated that the child's father had evidence of asymmetrical septal hypertrophy. An examination of the muscle removed from the right ventricle revealed histological changes compatible with the diagnosis of asymmetrical septal hypertrophy. A cardiac exploration was performed, but it was felt that the size of the aorta was small and that the left ventricle which had been severely rotated posteriorly by the enlarged right ventricle made a myomectomy by either the trans-aortic or trans-ventricular approach impossible. The patient was returned to the St. Louis Children's Hospital. In October of 1975, because of increasing loss of exercise tolerance, frequent spells of near syncope and diaphoresis, and continuing severe congestive heart failure, a third operation was performed. Because of the massive cardiomegaly, exposure of the heart required an extension of the median sternotomy through the left seventh intercostal interspace. With the child supported on a cardiopulmonary bypass, a modified commercially available 20 mm Hancock prosthesis was inserted into the left ventricular cavity through the left ventricular apex and sutured into position. The other end of this valve-bearing arterial prosthesis was led through a surgically created defect in the diaphragm, passed posterior to the stomach through the transverse mesocolon, and was sutured to the infra-renal aorta. The postoperative course was uncomplicated. A fifth cardiac catheterization was performed on the fourteenth postoperative day just prior to hospital discharge. At that study, a peak systolic gradient pressure of 35 mm Hg was measured between the aorta and the left ventricle. This pressure difference was interpreted as the consequence of catheter impingement, as there was no demonstrated obstruction to flow through the second and newly created left ventricular outflow tract. Left ventriculography demonstrated that the majority of left ventricular output exited through the apical-aortic shunt (Fig. 2). Flow through this route filled the aorta distally and in a retrograde fashion to the level of the left subclavian artery, a flow distribution which accounts for approximately two-thirds of left ventricular output. The child was discharged from the hospital with no medication. Since the time of his operation, he has demonstrated a progressive increase in exercise tolerance.

Case 2. A 17-year-old girl was first seen at the St. Louis Children's Hospital at the age of 7 months because of a loud systolic heart murmur. Right ventricular and left atrial enlargement were noted on the chest x-ray and a provisional diagnosis of interventricular septal defect or acyanotic type tetralogy of Fallot was made. The child remained acyanotic, but had a noticeable decrease in exercise tolerance and accentuation of the systolic murmur. At the age of four years,

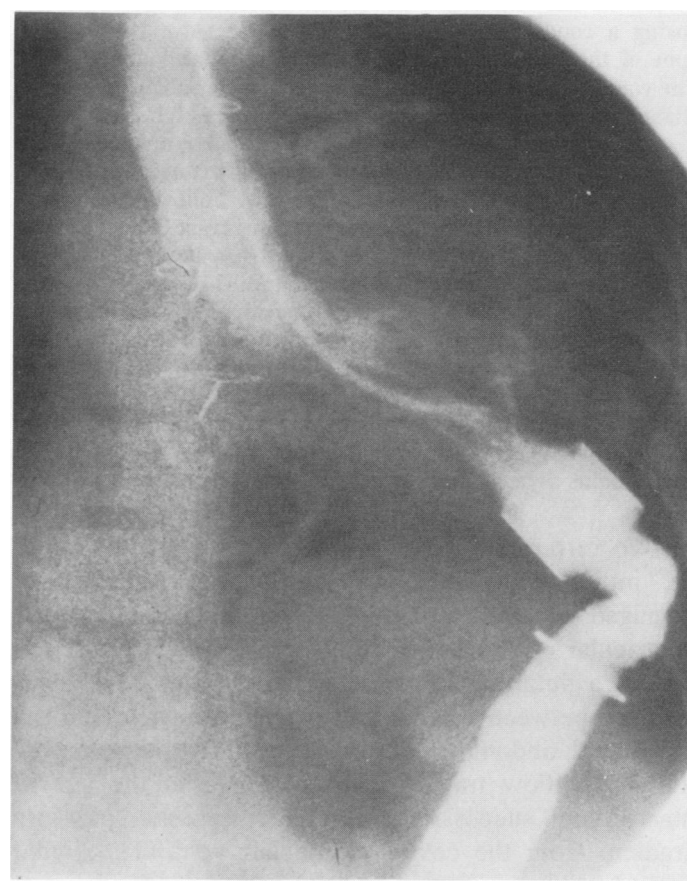

FIG. 2. Case 1. Left ventriculogram following the placement of an apical-aortic shunt. There is no longer any obstruction to left ventricular ejection. The metal cylinder projects into the left ventricular cavity to avoid obstruction at the apex.

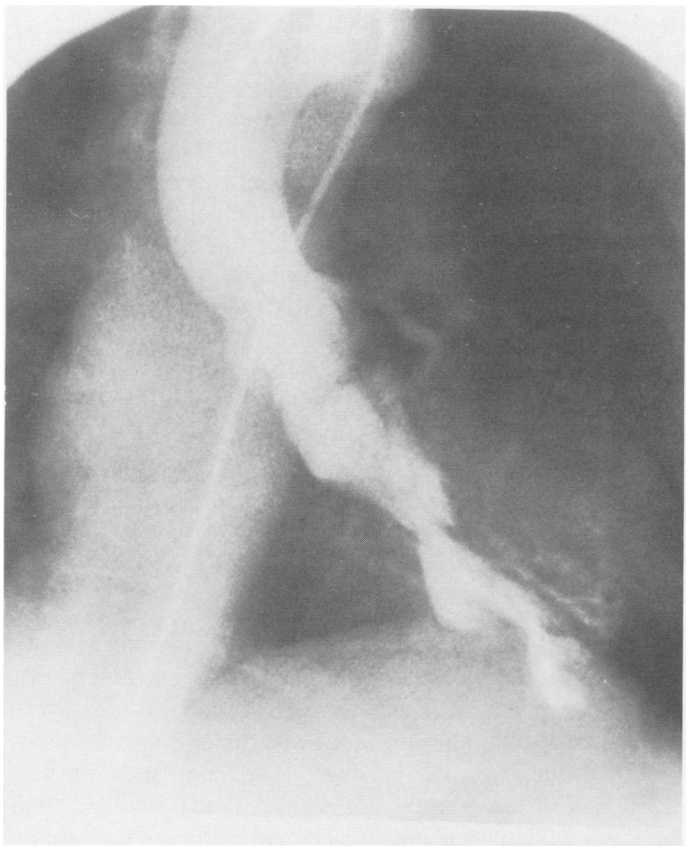

FIG. 3. Case 2. Left ventriculogram obtained prior to surgery. Both subaortic and mid left ventricular obstructions are apparent.

she was re-admitted to the hospital and a cardiac catheterization was performed which was erroneously interpreted as showing valvular pulmonic stenosis. Re-evaluation at the age of 13 included an auscultatory examination of the heart which suggested aortic stenosis and pulmonary stenosis. By the age of 17, she had severe complaints of exercise intolerance with dyspnea on exercise. Examination revealed left ventricular enlargement, prominent pulmonary vascular markings and electrocardiographic evidence of left ventricular hypertrophy. A cardiac catheterization was performed (Fig. 3), and this demonstrated a series of gradients within the left ventricle. There was a gradient of 50 mm Hg across a demonstrable subaortic diaphragmatic obstruction and an additional gradient of 80 mm Hg across a mid left ventricular obstruction, producing a total systolic gradient of 130 mm Hg between the body of the left ventricle and the aorta. In addition, a right ventricular gradient at the level of the infundibulum measured 50 mm during systole. Electrocardiograms in the postcatheterization period suggested a myocardial infarction and this was confirmed by a transient rise in the myocardial muscle creatinine phosphokinase and a positive myocardial scan performed with technitium pyrophosphate. An operation was performed approximately one month following the catheterization. With the patient supported on cardiopulmonary bypass, the left ventricular cavity was exposed through an aortotomy. The aortic valve was found to be bicuspid and showed commissural fusions. A commissurotomy of this bicuspid valve was performed. Beneath the valve, there was a well developed typical, subaortic, fibrous membrane most prominent along the ventricular septum, but extending onto the anterior leaflet of the mitral valve. This membrane was excised. Digital exploration of the left ventricular cavity then revealed a very severe stenosis at the level of the mid left ventricle which could not be well visualized, but which felt to be more fibrous than muscular. The intra-operative interpretation of these findings was that this represented a fibrous tunnel obstruction of the left ventricular outflow tract. The aortotomy was closed. The apex of the left ventricle was exposed. A circle of tissue was removed from the apex of the left ventricle and a specially modified 22 mm prosthesis bearing a glutaraldehyde-treated porcine aortic valve was interposed between the apex of the left ventricle and the infra-renal aorta. After inserting the rigid end of the prosthesis into the left ventricular cavity, the distal end was led through the diaphragm, passed posterior to the stomach, through the transverse mesocolon, and anastomosed to the infra-renal abdominal aorta. The interposition of the prosthesis in this case was similar to the first case except that the porcine valve was located above the diaphragm in the first case, and placed below the diaphragm in the second case. This change was instituted specifically to facilitate future replacement of the xenogeneic valve in case of future valve failure. Muscle resected from the apex of the left ventricle suggested the pattern seen with asymmetrical septal hypertrophy and suggests that this child also had that condition as the basis for her intra-ventricular obstruction. An electrocardiogram in the postoperative period demonstrated Q waves in leads 2, 3, and AVF, but a myocardial scan performed with technetium pyrophosphate failed to confirm the presence

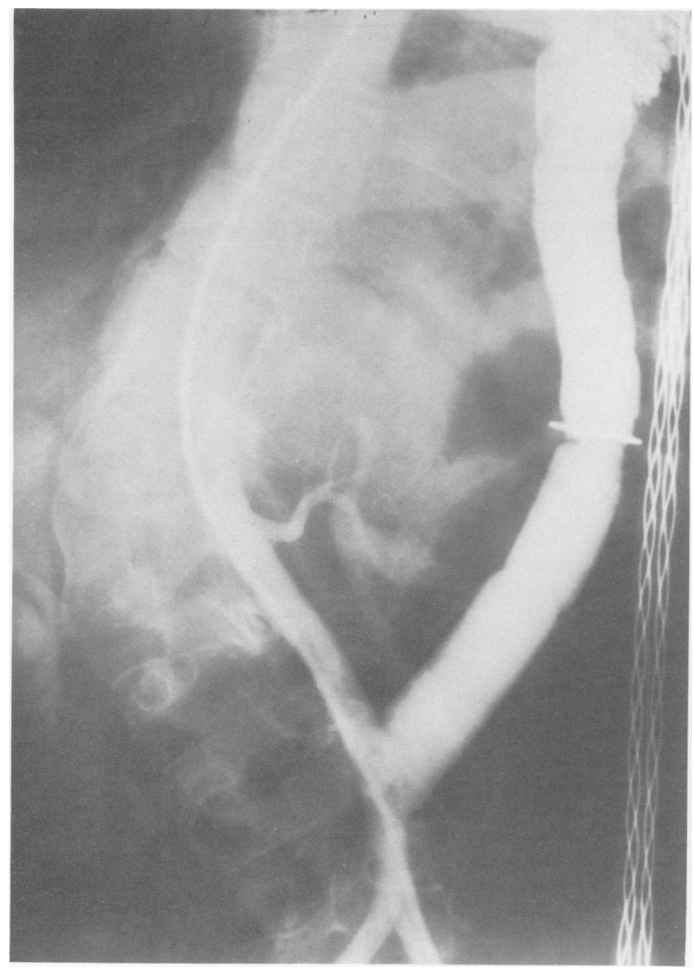

FIG. 4. Case 2. Appearance of the second left ventricular outflow tract (apical-aortic conduit). The porcine valve is in an intra-abdominal position. The left ventricle empties into the infra-renal aorta and provides retrograde flow to the level of the diaphragm.

of an infarct. It was concluded that the abnormal electrocardiographic pattern was a consequence of the left ventricular apical resection. The postoperative course was uncomplicated and a repeat cardiac catheterization was performed on the fourteenth postoperative day just prior to discharge (Fig. 4). A pull-back tracing from the infrarenal abdominal aorta through the prosthesis into the left ventricular cavity and out the aorta demonstrated an absence of any systolic pressure gradient. More remarkably, the gradient had also disappeared from the right side of the heart where there was no longer any septal impingement and normal pressures were recorded in the right ventricle. A left ventricular injection demonstrated that the newly created left ventricular outflow tract and dye filled the abdominal aorta to the level of the diaphragm. We estimated that the left ventricular-aortic conduit accounted for approximately 50% of left ventricular output. The patients has remained asymptomatic since discharge from the hospital.

Method

Modifying the Hancock Prosthesis (Fig. 5)

Commercially available Hancock prostheses were used. This prosthesis consists of a polyester knitted arterial prosthesis into which a glutaraldehyde treated porcine aortic valve has been inserted. The prosthesis was designed for the creation of a new right ventricular outflow tract usable in those situations where there is a lack of continuity between the right ventricle and the pulmonary circulation. Left ventriculograms were studied preoperatively to measure the thickness of the left ventricular wall at the ventricular apex. Using these measurements, a thin cylinder of stainless steel was machined so that its diameter equalled the internal diameter of the prosthesis and its height was 5 mm greater than the measured thickness of the left ventricular wall. A rectangle of flexible dacron cloth was then sutured together to form a cylinder. This cloth cylinder was made so that its length was slightly in excess of twice the height of the stainless steel cylinder. The stainless steel cylinder was placed over the cloth cylinder and the end of the cloth drawn down over the cylinder and sutured to itself. In this way, the stainless steel cylinder was covered on either side with polyester porous cloth. The cloth covered cylinder was sewn to the appropriate end of the Hancock prosthesis and as a final step, a strip of felt was used to cover this suture line forming a felt washer at the distal end of the cloth covered cylinder. The purpose of this washer was to facilitate fixation to the heart at the epicardial surface.

Inserting the Prosthesis

With the heart exposed through a median sternotomy and supported by a cardiopulmonary bypass, the apex of the left ventricle was elevated. A circular defect was created in the diaphragm just opposite the normal position of the left ventricular apex. A circular incision was made in the left ventricular apex slightly smaller than the rigid end of the prosthesis. An incision was made through to the left ventricular cavity so that a piece of left ventricular apex shaped as a truncated cone was

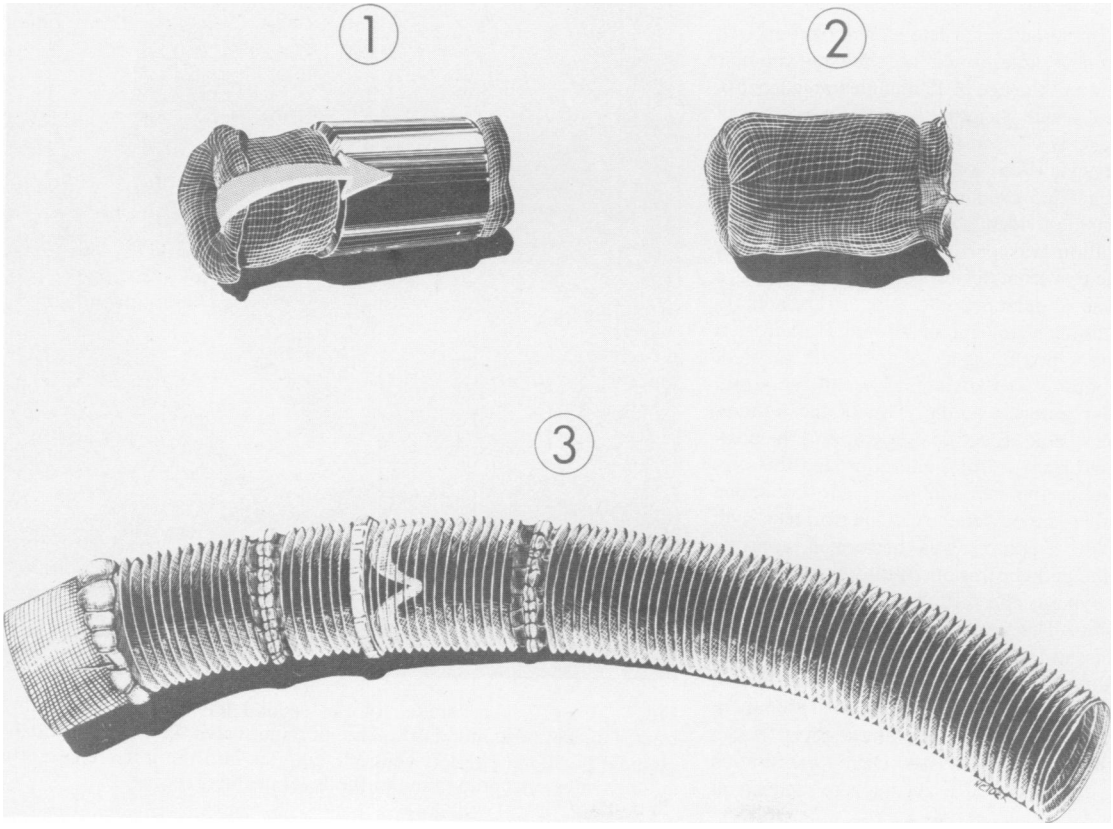

FIG. 5. Artist's illustration of the technique for modifying a commercially available valve bearing arterial prosthesis for use as an apical-aortic shunt. (1) A metal cylinder is drawn over a cloth cylinder. (2) The cloth is doubled over the metal cylinder and sewn to itself, thus enclosing the metal cylinder in porous polyester cloth. (3) The cloth covered cylinder is sewn to the prosthesis and a ring of felt is used to cover the junction.

removed. The incision on the endocardial surface was kept purposely smaller to avoid any damage to the papillary muscle attachments. The rigid end of the prosthesis was then firmly forced into the incision so that the felt cuff butted against the epicardium, and it was then sutured into place with a series of mattress sutures reinforced with teflon felt, drawn through the epicardium of the left ventricle, and through the felt washer on the prosthesis. The other end of the prosthesis was delivered through the diaphragmatic defect, passed posterior to the stomach, and through the transverse mesocolon, and brought into approximation with the infra-renal aorta. An anastomosis between the beveled end of the prosthesis and the side of the infra-renal aorta was made. In the first case, the prosthesis had been cut so that the valve was situated above the diaphragm. In the second case, it was purposely placed within the abdominal cavity to facilitate later replacement of the prosthetic valve. Consideration has been given to placing the prosthesis entirely in a retroperitoneal position, but this was not done in either case. Prior to completing the anastomosis between the aorta and the prosthesis, the prosthesis was allowed to fill with left ventricular blood and clamped within the abdomen. In both cases, the infra-renal abdominal aorta was exposed through a midline abdominal incision and was completed while the patient was still being supported with a cardiopulmonary bypass.

Discussion

Recorded among the many experimental achievements of Alexis Carrel is an attempt to divert blood from the ascending aorta to the descending aorta, employing a bypass between the left ventricular cavity and the distal aorta. Carrel was searching for a technique which would permit a safe resection of the aortic arch without damage to the spinal cord. In 1910,[7] he described his efforts employing paraffin rubber tubes and jugular vein to create bypasses from the left ventricular cavity to the descending aorta. In 1923, Jeger[13] improved upon these experiments by inserting a valve bearing conduit and managed to keep alive, over a four-day period, an experimental animal fitted with such a device and challenged by a ligation of the ascending aorta. Bailey and his associates[1] in 1950, searching for a technique to treat aortic valve stenosis, described some unsuccessful experiments in which they created ventriculo-aortic shunts using canine aortic homografts. Hufnagel[12] in 1951 described similar experiments. In 1954, Donovan and Sarnoff[11] described apical-aortic anastomoses used in acute animal experiments, and by 1955, Sarnoff[20] and his co-workers were able to report on 7 long-term dog survivors with valve bearing conduits inserted between the apex of the left ventricle and the aorta. The technique was applied clinically in the early 1960's by Templeton,[23] who inserted a rigid Hufnagel type prosthesis into five patients with aortic stenosis, interposing this prosthesis between the apex of the left ventricle and the thoracic aorta. One of these patients is said to have survived for a 13-year period. No further effort with the technique has been reported until 1975 when Bernhard[4] inserted a rigid apical-aortic conduit into a 22-year-old man for the treatment of a hypoplastic aortic annulus. This prosthesis was manufactured from a stainless steel tube covered with flocked dacron fibrils. A porcine xenograft glutaraldehyde-treated tissue valve was mounted into a polyester sleeve and interposed between the rigid conduit which had been inserted into the left ventricle and the thoracic aorta. This effort was a direct outgrowth of Bernhard's experience with the use of left ventricular assist devices which are designed to remove blood from the left ventricle and pump it into the descending thoracic or abdominal aorta.[3] The device which he used was indeed a modification of the device used for left ventricular assistance. In September of 1975, following a symposium on idiopathic hypertrophic subaortic stenosis at the International Surgical Congress in Edinburgh, Scotland, Cooley[8] discussed a similar patient with a small aortic annulus in whom he had inserted by direct suture a commercially available Hancock prosthesis between the left ventricle and the supra-renal abdominal aorta. It was these remarks which led directly to the authors' present reported effort. Cooley and Norman have recently reported this experience and similar success in three additional cases.[10] The reported postoperative left ventriculogram of their initial case demonstrates a marked narrowing at the anastomosis between the graft and the left ventricular apex.

A variety of successful operative techniques have been developed and successfully employed for the relief of left ventricular obstruction. These include plastic reconstruction of the aortic root for supra-valvular stenosis,[17] commissurotomy for congenital valvular stenosis,[22] resection of subvalvular membranes for the relief of subvalvular aortic stenosis,[16] and finally, aortic valve replacement for the management of aortic stenosis not suitable for commissurotomy.[21] Patients with an hypoplastic valve annulus have presented special problems since the size prosthesis which can readily be fitted into the subcoronary position provides for unacceptable obstruction to flow. In recent months, at least three different techniques[15,19,5] have been proposed and used for enlarging such a small aortic annulus and inserting an adult size prosthesis. Similarly, a number of operations have been proposed and successfully applied for the relief of intra-cavitary left ventricular obstruction, especially as it occurs in asymmetrical septal hypertrophy. These proposals include septal myotomy, trans-aortic resection of hypertrophied septum,[18] transventricular resection of hypertrophied septum,[2] resection of the septum through the right

ventricle to permit a displacement of the hypertrophied septum,[14] and replacement of the mitral valve with a low profile prosthesis.[9] Two of these operations, namely, resection of the septum from the right side and myotomy through the aorta, failed to relieve the left ventricular obstruction in our first patient. In both of our patients, the obstruction within the left ventricle was regarded as unresectable. The technique of creating a second left ventricular outflow tract by the construction of a conduit between the left ventricular apex and the aorta is an old one, but has been facilitated greatly by the recent availability of a flexible prosthesis bearing a dependable tissue valve. Our modification of this prosthesis for use in constructing a left ventricular outflow tract is based on the work of Brown[6] who demonstrated that muscle bundles within the left ventricle will eventually occlude the orifice of a conduit which does not extend into the left ventricular cavity for a minimum of 5 mm beyond the endocardial surface. The choice of the infra-renal abdominal aorta as a site of entry into the arterial system was dictated in our first patient by abnormal anatomy. In that case, the left ventricular apex lay directly against the thoracic aorta making the interposition of any conduit impossible. This site has the additional advantage, however, of improving the ease with which the prosthesis can be inserted. The concept of placing the valve within the abdominal rather than the thoracic cavity was added in our second case because of a strong conviction that no available prosthetic or tissue valve has any durability which approximates normal life expectancy. With the view that such a valve inserted into a child will require replacement in the future, it was felt that such a valve will be more readily accessible when situated in the abdominal cavity. This same reasoning suggests the possibility that apical-aortic shunting may have advantage over valve replacement into the subcoronary position when prosthetic replacement of the aortic valve is required in young children.

The creation of two left ventricular outflow tracts does, of course, create a permanent division of aortic blood flow. At the present time, there is insufficient experimental study of such a flow arrangement to determine whether or not it will have long term detrimental effects. It is recognized, however, that the fetus both develops and survives with a division of aortic blood flow.

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