The Surgical Management of Discrete and Diffuse Supravalvar Aortic Stenosis

JOHN F. KEANE, M.D., KENNETH E. FELLOWS, M.D., C. GRANT LAFARGE, M.D., ALEXANDER S. NADAS, M.D., AND WILLIAM F. BERNHARD, M.D.

SUMMARY Between 1956 and 1976, 18 patients underwent surgery for supravalvar aortic stenosis at The Children's Hospital Medical Center, Boston. Discrete obstruction, present in 11, was treated by insertion of a prosthetic gusset placed across the area of narrowing and extending into the noncoronary sinus of Valsalva. There was one operative death. Residual gradients (measured in five patients) ranged from 4-55 mm Hg, one of which was supravalvar in location. Significant aortic regurgitation was not common preoperatively. The diffuse form of supravalvar obstruction, a more difficult surgical problem, was present in seven patients. There were three operative deaths. Complete relief of the pressure gradient was achieved only in one instance by insertion of a left ventricular-aortic bypass shunt diverting the majority of the cardiac output into the descending thoracic aorta. This patient is now asymptomatic 20 months following operation. On the basis of this experience, it is suggested that patients with the diffuse form of supravalvar obstruction, and perhaps even those with a hypoplastic annulus alone, would benefit from a left ventricular-aortic bypass shunt.

CONGENITAL AORTIC STENOSIS is a relatively common form of heart disease with the anatomic site of obstruction usually located at the valvar level. Surgical management of this lesion is well established and can benefit a large number of patients with minimal risk. Occasionally, the level of obstruction may be supravalvar. The purpose of this report is to review the results of surgical management of both the discrete and diffuse forms of supravalvar aortic stenosis and to present a novel approach to the problem of diffuse supravalvar obstruction with a hypoplastic annulus.

Preoperative Evaluation

During the period 1956-1976, 18 patients with supravalvar aortic stenosis were operated upon at The Children’s Hospital Medical Center, Boston. The obstruction was discrete in 11 and involved the ascending aorta in a diffuse fashion in the remaining seven patients. Eleven of the 18 children were females. Thirteen complained of exercise intolerance, angina pectoris or syncope. The electrocardiogram (ECG) was abnormal in 17 (table 1). The remaining child, with an unremarkable ECG, was noted to have exercise intolerance and syncopal episodes. Seven patients were mentally retarded; five of these had discrete stenosis and “elfin-like” facies, an association first described by Williams et al.

At the preoperative cardiac catheterization, the left ventricular peak systolic ejection gradient (PSEG) ranged from 90-191 mm Hg (median 95 mm Hg). Angiography clearly delineated the sites of obstruction (figs. 1 and 2), and also quantitated aortic regurgitation (AR) in five patients (mild in three, and moderate and severe in one each [fig. 3]).

Five patients underwent a second preoperative cardiac catheterization during an interval of one to nine years (median 4 years). Among these, the PSEG remained unchanged in four but increased in one child with diffuse aortic obstruction (fig. 2) from 44 mm Hg at age three years to 122 mm Hg by age seven years with mitral regurgitation (MR) progressing simultaneously from mild to moderate.

Associated cardiac lesions were noted in 12 children (67%) and are outlined in table 2.

Surgical Results

At the time of operation, ages ranged from five to 27 years (median 14 years). Among 11 patients with the discrete form of supravalvar stenosis, there was one operative death (1960) due to a technical error in the placement of the aortic gusset. In the remaining 10 children, a Dacron or Teflon patch graft was employed to enlarge the narrowed aortic lumen. In six of these, mild valvar stenosis was also observed with fusion of commissures and thickening of the cusps. One patient with aneurysmal dilatation of the sinuses of Valsalva and fibrous retraction of all three cusps, required valve replacement (21 mm Bjork Shiley valve) to correct severe AR.

Three operative deaths (1957, 1960, 1968) occurred among the seven patients with the diffuse form of aortic obstruction. Marked hypoplasia of the ascending aorta was present in all; five also had hypoplasia of the aortic valve annulus. Additional subvalvar stenosis was noted in three and coarctation of the aortic isthmus in two. At the time of operation, surgical relief of the obstruction was not deemed possible in five of the patients, two of whom died at surgery, while insertion of a Dacron gusset in the hypoplastic aorta was carried out in the remaining two children, with one death.

One of the four survivors with diffuse obstruction underwent a second operation in 1974 (10 years following the initial exploration) because of severe exercise intolerance and angina and a residual PSEG of 128 mm Hg. At surgery, a newly designed left ventricular-aortic shunt incorporating a single xenograft valve was implanted. The rigid U-shaped prosthesis (Thermo Electron Corporation, Waltham, Mass.), 9.5 cm in length and 2.0 cm in width, shunted blood from the body of the left ventricle through an attached segment of 25 mm Dacron graft containing a xenograft valve (Hancock Laboratories Inc., Anaheim, Calif.) to the descending thoracic aorta. The rigid end of the prosthesis, 20 mm in diameter, was inserted through the apex of the left ventricle to maintain a constant-sized outflow tract. The
configuration of the apical section of the composite device directed blood through a 145-degree angle from the ventricular apex toward the descending aorta. Anastomosis of the woven Dacron graft portion of the device containing the tissue valve to the aorta was accomplished above the diaphragm at the level of the 8th rib posteriorly to permit easy coupling to the apical conduit. After implantation, the composite prosthesis was fixed to the diaphragm with several interrupted sutures to stabilize the rigid section, and to avoid kinking of the attached Dacron graft. The device fitted easily beneath the left lower lobe of the lung without compression or trauma. The blood-contacting surface within the metal shunt consisted of a fine layer of Dacron fibrils (25 microns in diameter and 250 microns in length) which encouraged the development of a well-attached pseudoendothelial lining. This bypass shunt has functioned satisfactorily for 20 months, during which time two hemodynamic evaluations have been performed.

**Table 1. Electrocardiographic Features**

<table>
<thead>
<tr>
<th>Left ventricular hypertrophy</th>
<th>Preoperative</th>
<th>Postoperative</th>
</tr>
</thead>
<tbody>
<tr>
<td>With strain</td>
<td>9</td>
<td>6</td>
</tr>
<tr>
<td>Without strain</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Strain pattern alone</td>
<td>6</td>
<td>3</td>
</tr>
<tr>
<td>Biventricular hypertrophy</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Right ventricular hypertrophy only</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Absent anterior forces only</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Normal</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>18</td>
<td>14</td>
</tr>
</tbody>
</table>

**Table 2. Supravalvar Aortic Stenosis: Associated Cardiac Lesions 12/18 (67%)**

<table>
<thead>
<tr>
<th></th>
<th>Discrete (6/11)</th>
<th>Diffuse (6/7)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypoplastic aortic valve annulus</td>
<td>0</td>
<td>5</td>
</tr>
<tr>
<td>Valvar aortic stenosis</td>
<td>6</td>
<td>2</td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Valvar</td>
<td>2</td>
<td>4</td>
</tr>
<tr>
<td>Peripheral</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Subvalvar aortic stenosis</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Stenosis of brachiocephalic arteries</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Coarctation</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Mitral regurgitation</td>
<td>0</td>
<td>1</td>
</tr>
</tbody>
</table>

**Postoperative Evaluation**

No deaths have occurred during the follow-up period, ranging from 10 months to 15 years (median 11 years).

**Discrete Stenosis**

Among the 10 survivors, seven are asymptomatic while three have exercise intolerance, one of whom (a 41-year-old patient with Williams syndrome, systemic hypertension and bilateral bypass grafts for carotid artery stenoses) has angina in addition. Aortic regurgitation is evident clinically in three patients, being mild in two and moderate in the other. The ECG (table 1) has improved in five, but is normal in only two. Postoperative cardiac catheterization has been performed in six patients. The left ventricle was entered in five and the residual PSEG ranged from 4–55 mm Hg (median 30 mm Hg), in contrast to 50–120 mm Hg (median 80 mm Hg) which was present preoperatively (fig. 4). The area of ascending aorta containing the prosthetic gusset was visualized at angiography (fig. 5). In the patient whose left ventricle was not entered, there was no supravalvar residual gradient.

**Figure 1. Supravalvar aortic stenosis, discrete type.** The stenotic segment is well located immediately above the aortic sinuses of Valsalva. The distal ascending aorta (Ao) is normal in size. LV = left ventricle.

**Figure 2. Supravalvar aortic stenosis, diffuse type.** Narrowing in the ascending aorta begins above the aortic valve (lower arrow) and extends throughout the ascending aortic segment to the origin of the brachiocephalic vessels (upper arrow). In this patient, the aortic arch and descending aorta appear hypoplastic also.
Diffuse Stenosis

Among the four survivors, three including the patient with the bypass shunt are asymptomatic, while exercise intolerance is present in the remaining patient. Mild AR is present in one patient and moderate MR persists in another. The ECG (table 1) remains unchanged in three and has improved in the patient with the bypass shunt. Postoperative cardiac catheterization has been performed in three patients, the PSEG remaining unchanged compared to preoperative levels in two, while being completely eliminated in the 22-year-old male with the left ventricular-aortic shunt (fig. 4).

In this latter patient, the left ventricular ejection fraction was normal (0.64), although the left ventricular end-diastolic pressure was elevated to 22 mm Hg, being only 6-8 mm Hg preoperatively, but 22 mm Hg five years earlier. This, perhaps, represents a loss of ventricular compliance resulting from the long-standing pressure overload. Left ventricular angiography disclosed contrast material leaving the heart via both the aortic valve and the bypass shunt (fig. 6).

Discussion

Congenital aortic stenosis is more common in males with the anatomic site of obstruction usually being located at the valvar level in approximately 70% of surgical patients.\(^1\) Supravalvar aortic stenosis involving the ascending aorta, first described by Mencarelli in 1930\(^0\) is relatively rare (approximately 7% of our surgical patients),\(^1\) but affects both sexes equally.\(^7\) In a review of 68 autopsied patients, including eight of their own, Peterson identified a localized hourglass deformity in 45, hypoplasia of the ascending aorta in 14 and a membranous type of obstruction in the remaining nine patients.\(^7\) He further noted that the lesions, in some, had features of more than one type of obstruction. Among our 18 patients, 11 had the hourglass type of deformity, which we termed discrete, while the remaining seven had diffuse hypoplasia of the ascending aorta.

The high incidence of associated cardiac anomalies, including coarctation of the aorta, abnormalities of the aortic valve, coronary arteries, brachiocephalic vessels, pulmonary valve and pulmonary arterial tree, has been well documented in literature reviews.\(^7\)\(^-\)\(^9\) Mitral valve abnormalities, although less frequent, have also been described.\(^10\)\(^-\)\(^17\) The majority of these anomalies were present in our series, although none of our patients had coronary arterial obstructive lesions. In addition, the aortic valve annulus was noted to be hypoplastic in five of our seven patients with diffuse hypoplasia of the ascending aorta. Of interest was the progressive nature of both the diffuse obstruction and mitral regurgitation documented in one of our patients by repeat cardiac catheterization over a four-year preoperative interval. The rather frequent association

### Table 3. Changes in PSEG* at Cardiac Catheterization: Discrete Supravalvar Aortic Stenosis

<table>
<thead>
<tr>
<th>Author</th>
<th>Preop (mm Hg)</th>
<th>Interval</th>
<th>Postop (mm Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Farrowi(^1)</td>
<td>75</td>
<td>?</td>
<td>40</td>
</tr>
<tr>
<td>Shumacker(^2)</td>
<td>79</td>
<td>5 months</td>
<td>30</td>
</tr>
<tr>
<td>Nordstrom(^3)</td>
<td>122</td>
<td>3 years</td>
<td>27</td>
</tr>
<tr>
<td></td>
<td>134</td>
<td>2 years</td>
<td>24</td>
</tr>
<tr>
<td>Rastelli(^4)</td>
<td>63</td>
<td>3 years</td>
<td>78 (valvar)</td>
</tr>
<tr>
<td></td>
<td>83</td>
<td>2 years</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>116</td>
<td>4 years</td>
<td>0</td>
</tr>
<tr>
<td>Weiss(^5)</td>
<td>90</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>75</td>
<td>7-44</td>
<td>8</td>
<td></td>
</tr>
<tr>
<td>65 months</td>
<td>30 (valvar, subvalvar)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>161 postop</td>
<td>90 (subvalvar)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>40 for group</td>
<td>4</td>
<td></td>
<td></td>
</tr>
<tr>
<td>80</td>
<td>40 (valvar, subvalvar)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Present Series</td>
<td>50</td>
<td>2 years</td>
<td>37 (valvar)</td>
</tr>
<tr>
<td>70</td>
<td>8 years</td>
<td>30 (subvalvar)</td>
<td></td>
</tr>
<tr>
<td>80</td>
<td>10 years</td>
<td>55 (valvar, supravalvar)</td>
<td></td>
</tr>
<tr>
<td>90</td>
<td>6 years</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>120</td>
<td>11 years</td>
<td>10</td>
<td></td>
</tr>
</tbody>
</table>

*Peak systolic ejection gradient.
of mental retardation and “elfin-like” facies with supravalvar stenosis, first reported by Williams, was also noted among our patients, in both the discrete and diffuse groups.

Discrete Stenosis

The surgical relief of the discrete form of supravalvar stenosis, by insertion of a gusset of prosthetic material across the narrowed area was first described in 1961. In a subsequent report in 1966 by Rastelli, 14 patients had undergone this procedure without mortality. In earlier years, mortality rates among smaller series were variable, while in a recent report, there were no deaths among eight patients operated upon between 1969 and 1973. We have had one death (1960) among our 11 surgically treated patients with this type of obstruction.

Table 3 summarizes the results of surgery, as measured at postoperative catheterization, from the literature and the
present study. It may be seen that the majority of the survivors studied, particularly in the excellent recent series of Weisz, had only mild residual gradients, if any, and some of these were at the subvalvar or valvar level.

Diffuse Stenosis

It has been felt for some time that surgery for the diffuse form of supravalvar obstruction was an unrewarding undertaking. Among our seven patients there were three operative deaths, and postoperatively, cardiac catheterization in three survivors revealed no reduction in PSEG.

From a historical standpoint, a number of investigators have attempted experimentally to bypass the aortic valve using a conduit. In 1955, Sarnoff and co-workers devised a rigid polished Lucite tube incorporating a Hufnagel ball valve which was inserted between the left ventricle and descending aorta in a series of dogs, five of the 17 animals surviving for more than six months.

In 1967, following an extended series of experiments performed by Bernhard, development of a left ventricular-aortic prosthesis which functioned satisfactorily in calves for periods up to one year was reported. Successful laboratory studies conducted during the ensuing seven years were the basis for the first clinical bypass shunt procedure performed in 1974. The composite prosthesis used in this patient consisted of a rigid U-shaped apical tube coupled to a 25 mm Dacron graft containing a xenograft valve.

In a recent report by Cooley, three additional patients, 7, 10 and 15 years of age, with supravalvar stenosis, have undergone a somewhat different left ventricular-aortic bypass procedure employing only the use of a small-sized (14 mm) Hancock-valved conduit. The proximal end of the Dacron graft was sutured directly to an opening made in the left ventricular apex and brought through the left hemidiaphragm to the abdominal aorta above the celiac axis. The initial procedure was undertaken in August 1975, and cineangiograms performed 10 days postoperatively indicated filling of the graft in early and mid-systole with pinching off of the ventricular end of the conduit in late systole, without, however, any significant pressure gradient. The longest follow-up period for these patients is three months, and no postoperative hemodynamic data are available for the two most recent patients.

The main objections to anastomosing a Dacron graft directly to the hypertrophied ventricular myocardium are related to encroachment by muscle upon the lumen of the graft during ventricular systole and narrowing of the ventricular anastomosis by scar formation. In addition, kinking or twisting of the conduit in its circutious path from the left hemidiaphragm to the abdominal aorta is also possible, and in young patients, the fate of the grafts as growth continues remains uncertain.

Postoperative cardiac catheterization of the 22-year-old Children’s Hospital patient with the bypass graft was performed seven months postoperatively, and revealed complete elimination of the left ventricular-aortic pressure gradient. During angiography, contrast material was seen exiting mainly through the conduit, although some dye was also noted traversing the aortic valve, and then entering the ascending aorta (fig. 6). This patient remains asymptomatic 20 months following operation.

Use of a composite left ventricular-aortic shunt represents a new approach to the problem of diffuse hypoplasia of the ascending aorta and aortic valve annulus. In addition, the procedure may be of value in certain patients with a hypoplastic valve annulus alone, and in others with severe left ventricular outflow obstruction due to the long, tunnel-like variety of subvalvar aortic stenosis.

Addendum

Since this manuscript was completed, a 26-year-old symptomatic male with a very small aortic valve annulus successfully underwent placement of a left ventricular-aortic bypass shunt. He had undergone an aortic valvulotomy 17 years ago, and had a preoperative residual PSEG of 95 mm Hg. Cardiac catheterization was performed 22 days following operation which revealed normal left ventricular pressure and elimination of the systolic gradient. The patient was asymptomatic and discharged an anticoagulant therapy. Seven days later, left-chest pain commenced and a prothrombin time of 30 seconds was reported at another hospital. Two days later chest pain increased in severity, hemoptysis appeared, thoracic bleeding became evident on X-ray and he was transferred to Children’s Hospital where he died later that day at surgery. At autopsy extensive bleeding had occurred from a small leak at the junction of the dacron graft with the descending aorta.

References


Transatrial Resection of the Obstructed Right Ventricular Infundibulum

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SUMMARY Obstructions of the right ventricular infundibulum were resected through the orifice of the tricuspid valve in 21 patients, 15 of whom had tetralogy of Fallot. At operation the systolic pressure difference between the right ventricle and pulmonary artery after repair averaged 18 mm Hg (range 0–40 mm Hg). In patients with tetralogy, cardiac index four hours after operation averaged 2.8 L/ M²/min. One patient with tetralogy and severe pulmonary hypertension died. Twelve patients with tetralogy were rehydrated 10 to 186 days after operation. The mean systolic pressure difference between right ventricle and pulmonary artery was 23 mm Hg. Residual obstructions were in the pulmonary valvular annulus. Cineangiograms did not show paradoxical motion of the right ventricular wall. Transatrial resection of right ventricular infundibular obstructions carries with it none of the consequences that often follow right ventriculotomy and this surgical approach satisfactorily relieves infundibular obstructions.

A TRANSVERSE OR VERTICAL INCISION in the right ventricle is generally used to resect muscular or fibrous obstruction of the infundibulum. An incision in the right ventricle reduces myocardial contractility, may be associated with complete right bundle branch block, and causes necrosis of adjacent myocardium. In a few patients a major coronary arterial branch may be transected or the right ventricular free wall may develop akinesis or become aneurysmal. An especially long ventriculotomy, the addition of an outflow patch, or creation of pulmonary insufficiency further impairs right ventricular performance after surgical correction of infundibular fibromuscular obstructions.

This report describes our experience with a technique for resecting infundibular obstructions through a right atriotomy and the orifice of the tricuspid valve. The technique is an extension of methods developed for transatrial closure of high ventricular septal defects. Our initial experience with 21 patients, 15 of whom had tetralogy of Fallot, is reported.

Operative Technique

The operation is performed through a median sternotomy

using conventional cardiopulmonary bypass, hemodilution, and hypothermia to 28–30° C. The right atrium is incised and the left ventricle is vented through the fossa ovalis or right superior pulmonary vein if a ventricular septal defect is present. Often the pulmonary valve is exposed through a longitudinal arteriotomy and the valvular obstruction is relieved.

The anterior leaflet of the tricuspid valve is retracted cephalad and anteriorly. Chordae attached to the papillary muscle of the conus are retracted caudally. An incision is made in the crista supraventricularis parallel to the plane of the ventricular septum. This incision is carried laterally toward the aortic annulus and anteriorly beneath the retracted anterior leaflet of the tricuspid valve into the endocardium of the free right ventricular wall. With scissors, the surgeon establishes a plane of dissection between the fibromuscular obstructing tissue and the aorta and free right ventricular wall. Fingers of the left hand partially invaginate the anterior-right lateral portion of the right ventricular infundibulum as the dissection proceeds cephalad to the pulmonary annulus. The free right ventricular wall is left approximately 4–7 mm thick.

Hypertrophied septal bands are resected similarly. The left lateral portion of the crista supraventricularis is incised in the plane of the ventricular septum. This incision is extended caudally and laterally to the base of the anterior papillary muscle of the tricuspid valve. The incision then curves anteriorly into the endocardium of the free right ventricular wall. A plane of dissection is established with

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