Surgical repair of congenital supravalvular aortic stenosis in children

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Abstract

Objective: Supravalvular aortic stenosis (SVAS) is an uncommon congenital cardiac anomaly characterized by varying degrees of left ventricular outflow tract obstruction beginning distal to the aortic valve. Methods: Between March 1962 and December 2000, 101 consecutive patients underwent surgical correction for congenital SVAS at Riley Children’s Hospital. There were 61 male (60%) and 40 female (40%) ranging in age from 3 month to 17 years (medium age, 6.1 years). Fourteen patients (14%) had Williams syndrome. Preoperatively, 11 patients were in New York Heart Association (NYHA) functional class I, 55 in class II, 28 in class III, and seven in class IV. Of the 101 patients, 73 (72%) had localized type SVAS and 28 (28%) diffuse type SVAS. Results: Those with localized SVAS were successfully treated with patch aortoplasty, whereas those with diffuse SVAS required either an apical aortic conduit or extensive endarterectomy with patch aortoplasty. The overall mean pressure gradient was reduced to 21 mmHg (P < 0.001) in the early postoperative period. There were one early death (30 days postoperatively) (1%), two (2%) late deaths, and 14 patients (14%) underwent one or two additional operation (n = 17) in a follow-up period ranging from 6 months to 30 years (medium 9.4 years). Postoperatively, there were 72 patients (73%) in NYHA functional class I and 26 (27%) in class II. Overall survival including operative mortality was 98% at 10 years, 97% at 20 and at 30 years. Conclusion: Good surgical outcome of congenital SVAS can be achieved with the appropriate method of treatment in patients with both localized and diffuse SVAS. © 2002 Elsevier Science B.V. All rights reserved.

Keywords: Congenital valve disease; Aortic stenosis; Children; Left ventricular outflow tract

1. Introduction

Supravalvular aortic stenosis (SVAS) is an uncommon congenital cardiac anomaly characterized by varying degrees of left ventricular outflow tract obstruction beginning distal to the aortic valve and is also associated with Williams-Beuren syndrome [1,2]. The obstruction in SVAS results from a thickening of the vessel wall secondary to excessive collagen and hypertrophied smooth muscle cells in the medial layer of the aorta [3].

Classically, three morphologic subtypes have been described: an hourglass type, a membranous type, and a diffuse type. Clinically, the hourglass and membranous types are managed similarly. The diffuse subtype, which involves hypoplasia of a variable length of ascending aorta, often necessitates different surgical approaches.

Since 1961 when McGoon and colleagues [4] reported successful surgical repair of this anomaly, the standard operation for SVAS has been a prosthetic or pericardial patch across the stenosis to widen the aorta at that point.

Doty and associates [5] described a more extensive repair that both relieves the obstruction and restores a more anatomical configuration to the aortic root. This operation involves placement of an inverted Y-shaped incision across the supravalvular-constricting ring and into the right and non-coronary sinuses of Valsalva, followed by insertion of a pantaloon-shaped Dacron fabric patch.

The aim of this report is to provide long-term follow-up of the large group of patients who underwent standard patch aortoplasty for SVAS.

2. Patients and methods

One hundred and one patients, between March 1962 and December 2000, underwent surgical repair for congenital SVAS at Riley Children’s Hospital at Indianapolis, IN, USA. Case histories of some of these patients have been presented previously [6–8]. The mean age at the time of surgery was 6.1 ± 2.8 years, with a range of 3 months to 17 years. Sixty-one (60%) patients were male and 40 were
female (40%). Seventy-three (72%) patients had localized type SVAS, and 28 (28%) had the diffuse type. Seventy-six of the 101 (75%) had one or more associated congenital cardiac anomalies (Table 1). The most common associated anomalies were valvular aortic stenosis, which was present in 35 patients (34%). Fourteen patients (14%) had Williams-Beuren syndrome, associated with mental retardation and the typical ‘elf-like’ faces. In this syndrome associated vascular or other anomalies were found in all patients (Table 2). Forty-one patients (40.6%) had previous cardiovascular operation (Table 3).

Eleven patients (11%) were free of symptoms; 55 (55%) were in New York Heart Association (NYHA) functional class II, 28 (27%) were in class III, and seven (7%) was in class IV. Preoperative symptoms included exercise intolerance in 32 patients, angina in 28 patients, and syncope in nine patients. Normal sinus rhythm was present in all patients.

The diagnosis and pressure gradients were established by cardiac catheterization preoperatively in all patients. Postoperative peak gradients were measured in the operating room after the patient was weaned from cardiopulmonary bypass by direct needle puncture. Peak gradients were determined at long-term follow-up by two-dimensional echocardiography with Doppler flow analysis. In addition, 13 patients also underwent late cardiac catheterization. Preoperatively, the peak pressure gradient ranged from 45 to 200 mmHg (mean 90 ± 33 mmHg, P = NS). Aortic insufficiency was observed in 24 patients: mild in four, moderate in 14, severe in six.

2.1. Operative procedures

Standard techniques for cardiopulmonary bypass were used. Profound hypothermia and circulatory arrest were added for some patients who underwent placement of ascending-descending aorta grafts or aortic arch reconstruction. Cold potassium crystalloid or blood cardioplegia was used for myocardial protection since 1976. Subsequently, a generous standard patch was sutured into the incision to increase the circumference of the aorta. Although Dacron (CR Bard, Covington, GA), autologous pericardium were used as patch material in the past, Haemashield fabric (Boston Scientific Corp., Natick, MA), which has given excellent early and late results, has been our prosthetic material of choice for many years.

2.1.1. Procedures to repair supravalvular aortic stenosis

SVAS was repaired using one of several techniques. In 12 patients with diffuse type, extended aortoplasty with a patch in the shape of an inverted Y with extensive endarterectomy was performed. A longitudinal aortotomy was extended from the non-stenotic segment of ascending aorta distal to the supravalvular stenosis, deep into the right and non-coronary sinuses of Valsalva. The supravalvular ridge was excised carefully, and procedures on the aortic valve, root and subvalvular region were performed if necessary. The aorta was then augmented with a Y-shaped patch using continuous non-absorbable monofilament suture.

For some patients (n = 5) with diffuse SVAS and valvular or supravalvular aortic stenosis operated on early in the series with apical aortic conduit (AAC) inserted were the

Table 1

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>Patients</th>
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<tr>
<td></td>
<td>No.</td>
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<td>35</td>
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<tr>
<td>Aortic insufficiency</td>
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<tr>
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<tr>
<td>Coarctation of aorta</td>
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<td>Ventricular septal defect</td>
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<tr>
<td>Patent ductus arteriosus</td>
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<td>Complex LVOTO</td>
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<tr>
<td>Branch and/or peripheral pulmonary stenosis</td>
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<tr>
<td>Interrupted aortic arch</td>
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Table 2

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Table 3

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<td>Repair of IAA</td>
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<td>Open aortic valvotomy</td>
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<tr>
<td>Repair TAPVR</td>
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<tr>
<td>Bidirectional Glenn</td>
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</tr>
<tr>
<td>Total</td>
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*a PS, pulmonary stenosis.
commercially available woven Dacron graft containing a glutaraldehyde-preserved porcine valve (Hancock-Extra-corpooreal) or aortic homograft. The conduits ranged in size from 8 to 22 mm: 8–12 mm conduits were used in children less than 3 years old, and 22 mm conduits for adults. AAC was used in accordance with the technique described previously [8].

A diamond-shaped patch was inserted in the ascending aorta after longitudinal incision through the stenotic region extending in to the non-coronary sinus in 80 cases. Extensive resection of the stenotic ridge was frequently attempted but proved impossible in most cases because the ridge usually represented a construction of the thickened aortic wall at the level of the leaflet hinge point, rather than a circumscribed fibrous structure.

In four patients, a Ross (n = 2) or Ross-Konno (n = 2) procedure was performed. In all of these, the segment of ascending aorta distal to the pulmonary autograft was augmentation with single diamond-shaped patch of aortic homograft tissue.

Of the 73 patients with localized SVAS (Fig. 1), 71 had a single, diamond-shaped patch placed, two had a Ross procedure. The 28 patients with diffuse narrowing of the ascending aorta (Fig. 2) were initially treated with a diamond-shaped (n = 9) or inverted bifurcated patch with extensive endarterectomy (n = 12) extending to the origin of innominate artery, Ross-Konno procedure (n = 2), and five patients had AAC plus a diamond-shaped patch. Among the 28 patients with diffuse SVAS, the aortic root and proximal ascending aorta were enlarged in ten with standard patch.

2.1.2. Procedures on the left ventricular outflow tract and coronary arteries

In addition to the four patients who underwent a Ross or Ross-Konno operation, 31 procedures were performed on the aortic valve. These included, in addition to the Ross procedure, mobilization of restricted leaflets (commissurotomy and detachment of leaflets partially fused to the supra-valvular ridge) in 26 patients, thinning of the leaflets in three, resuspension of one or more leaflets in one, and augmentation of a leaflet in one.

Resection of subvalvular left ventricular outflow tract

![Fig. 1. Surgical results of discrete type of supravalvular aortic stenosis (SVAS). CoA, coarctation of aorta; AVR, aortic valve replacement; and AH, aortic homograft.](image1)

![Fig. 2. Surgical results of diffuse type of supravalvular aortic stenosis (SVAS). AAC, apicoaortic conduit; RVOT, right ventricular outflow tract; and GT, Gore-Tex.](image2)
obstruction was performed in 25 patients, including two of four who underwent a Ross procedure. Subvalvular membranectomy and myectomy were employed as indicated in each of these patients.

Four patients underwent procedures on the coronary arteries. Two patients were treated with patch aortoplasty encompassing the left main ostium and SVAS. Another two patients were treated with excision of the fused leaflet from the aortic wall and patch aortoplasty.

2.1.3. Other procedures

Nine of the 14 patients with Williams’ syndrome underwent augmentation of the pulmonary arteries with a patch. In all of these patients, augmentation consisted of an inverted-Y patch extending into the sinuses of the pulmonary root, along with an extensive augmentation of the branch pulmonary arteries and partial pulmonary valvectomy.

2.2. Statistical analysis

Variables that were analyzed with the \( \chi^2 \) or Student’s test were as follows: age; sex; presence of associated cardiovascular anomalies; presence of associated aortic valve disease; form of SVAS (discrete or diffuse); preoperative NYHA functional class; previous cardiovascular operations; preoperative, intraoperative post repair, and late postoperative left ventricular-aortic gradients; presence of Williams-Beuren syndrome; standard versus pantaloon-shaped patch; year of operation; need for previous or concomitant aortic valve replacement. Survival estimates were made with Kaplan-Meier method and comparisons between survival distributions were made with the log-rank and Gehan Wilcoxon tests. Mean medium, range and standard deviations were also calculated.

3. Results

3.1. Early and late results

There was one hospital death (1%). This resulted from intraoperative cardiac failure in the early part of the series (1962). In this patient Dacron patch enlargement of the aorta had been confined to the root. There were no other deaths, significant complications, or reoperations in the perioperative period. The mean cardiopulmonary bypass time was 128 ± 64 min (range, 42–202 min), and the mean duration of aortic cross-clamp time was 42 ± 26 min (range, 35–112 min). Patients remained in the hospital for a mean of 6 ± 4 days after surgery (range, 4–28 days).

Follow-up ranged from 6 months to 30 years (median 9.4 years). There were two (2%) late deaths. The first patient had with localized SVAS underwent total aortic arch replacement with aortic homograft elsewhere 3 years after the primary operation due to restenosis of the supravalvular area (gradient 80 mmHg). The single late death occurred in a second patient with residual supravalvular and tunnel subaortic stenosis. The operative procedure for insertion of the conduit and the hospital course were unremarkable. This patient was discharged on the 14th postoperative day but returned 3 weeks later with fever and left pleural effusion. Although the pleural fluid was sterile initially, it became secondarily infected and led to a staphylococcal empyema. The secondarily infected conduit was removed, and aortoventriculoplasty (Konno procedure) was performed. The patient died of renal failure and continued sepsis 2 weeks following the operation.

Overall survival estimated by the Kaplan-Meier method including early mortality, was 98% (70% CI, 78–99%) at 5 years and 97% (70% CI, 76–99%) at 20 and 30 years (Fig. 3A). Univariate and multivariate analysis identified none of the other tested variables as risk factors of deaths.

3.2. Reoperation

Fourteen patients underwent one or two reoperations, the majority for aortic valvular disease (Table 4). Reasons for aortic valve re-replacement (with aortic homograft or pulmonary autograft) included multilevel obstruction and severe aortic regurgitation. Three of the four patients with AAC had undergone reoperations: one – replace AAC (Hancock-Extracorporeal porcine valve) and later re-reoperation – Ross procedure, and two – removed of AAC. Risk factors for reoperation by univariate analysis were associated aortic valve disease (\( P = 0.03 \), presence

![Fig. 3. Kaplan-Meier estimated 30-year overall survival (A); and freedom from reoperation (B). Error bars indicate the lower 70% CI. Number of patients at risk are shown in parentheses.](image-url)
of late left ventricular – aortic gradient greater than 35 mmHg \((P = 0.05)\), associated left ventricular outflow tract obstruction (LVOTO) \((P = 0.003)\), and the need for previous or concomitant aortic valvotomy or AAC \((P = 0.05)\). By multivariate analysis, the only risk factor for reoperation was associated LVOTO \((P = 0.04)\). Overall freedom from reoperation or intervention estimated by the Kaplan-Meier method was 86.9% \((70\% \text{ CI, } 66–90\%)\) at 10 years, 83.8% \((70\% \text{ CI, } 48–76\%)\) at 20 years, and 82.1% \((70\% \text{ CI, } 35–64\%)\) at 30 years (Fig. 3B).

The late mean left ventricular-aortic gradient was lower in the patients with localized SVAS \((12 \pm 10 \text{ mmHg}, \text{ range } 0–35 \text{ mmHg})\) than in those with diffuse SVAS \((22 \pm 17 \text{ mmHg}, \text{ range } 0–65 \text{ mmHg}; P = 0.01)\). Late aortic insufficiency, present in 18 patients, was mild in 16, and moderate in two. All patients with aortic insufficiency had associated aortic stenosis: no patient with a normal aortic valve had late aortic insufficiency.

Postoperatively, there were 72 patients \((73\%)\) in New York Heart Association class I and 26 patients \((27\%)\) – in class II (Fig. 4). Two of the seven patients in class IV died, as did one of the 23 in class III. In four patients in whom a severe pulmonary branch stenosis was relieved by patch enlargement, there was echocardiographic evidence of persistence of the pulmonary artery branches. The other two patients with unoperated mild pulmonary branch stenosis remained free of symptoms.

4. Discussion

SVAS is a complex form of LVOTO with distinctive morphologic features, which influence the results of corrective surgery [9,10]. Our study demonstrates that standard patch aortoplasty provides good postoperative results with asymptomatic patients for up to 30 years. However, residual abnormalities may remain, including gradients across the aortic valve and aortic valve insufficiency. These abnormalities appear related to the amount of attachment between the aortic valve cusps and residual anterior supravalvular tissue [11].

McGoon and associates [4] first described successful surgical repair for SVAS with standard patch in 1961. This technique remained the standard operative procedure for localized SVAS. In 1964, Shumacker and Mandelbaum at the Indiana University [6] treated patients with the diffuse form of SVAS with an extended patch into the entire ascending aorta. In 1978, Brom [12] introduced symmetric aortoplasty with enlargement of all three sinuses by the 3-patch technique. The aorta is transected just distal to the level of maximal obstruction, and three pericardial patches are inserted into the aortic sinuses, leading to a normalized geometry of the aortic root with complete relief of the aortic stenosis.
obstruction. The technique that has been reported by Myers and associates obviates the use of prosthetic patch material [10]. The aorta is transected just above the level of stenosis and all thickened tissue is removed. Three vertical incisions are made in the Valsalva sinuses. In the ascending aortic end, three longitudinal incisions are made that correspond to the commissures of the aortic valve. With an up-and-down running suture line, the continuity of the aorta is restored, obtaining three flaps of aortic tissue that are sewn into the sinuses. Other procedures to correct SVAS include circumferential resection of the stenosing ring followed by primary anastomosis of the proximal and distend of the ascending aorta [13], and the use of an apical aortic conduit to bypass the lesion [8,14–16].

The aortoplasty technique was modified by Doty and associates [5] by incising the supravalvar ring at two points in the non-coronary and the right coronary sinus of Valsalva. The repair was achieved with an inverted V-shaped Dacron patch to provide a wide aortic cross-sectional area. This extended aortoplasty resulted in complete relief for patients with localized SVAS [17,18].

Patch aortoplasty has been shown to be quite effective in reducing the supravalvar gradient, but there have been reports of a persistent gradient across the region of the aortic root [18]. Data from several reports [17,19] demonstrate the postoperative gradient to average 12–30 mmHg. The attachment of the right and left coronary cusps to the fibrous intimal ridge after patch enlargement of the non-coronary sinus may account for some of these postoperative aortic gradients [18,20]. In our series, the postoperative gradient was extremely low in both groups (12 ± 10 mmHg in localized SVAS; 22 ± 17 mmHg in diffuse SVAS). The absence of important gradients in our patients may have been due to the use of a large patch, extending from the valve hinge in the sinus of Valsalva to the ascending aorta well above the obstructive waist and wide enough to produce a normal aortic diameter at the level of obstruction.

Patients with the diffuse form of SVAS present a special problem. Because relief of the pressure gradient by patch repair is often incomplete [19,20], an apical aortic conduit has been recommended [14,17]. Our 28 patients with this form of the disease had good results with patch aortoplasty and apical aortic conduit. A diamond-shaped patch aortoplasty was successful in nine patients and three patients had four reoperations (Fig. 2). Inverted bifurcated patch aortoplasty with extensive endarterectomy was successful in twelve patients and only one patient had two reoperations. The next five patients were treated with an apical aortic conduit and patch aortoplasty. Three of these patients (60%), in follow-up period, had four reoperations. In addition, associated complications, including infective endocarditis, conduit stenosis, and porcine valve dysfunction, have made this approach less favorable. We agree with others [5,14,16,21] that all operations should begin in the aortic root so that any necessary repairs of the aortic valve, subaortic area, or coronary ostial obstructions can be performed.

Aortic insufficiency after repair has been attributed to the persistent attachment of the right and left coronary leaflets to the residual fibrous intimal ridge [16,22].

Although most reoperations had patients with complex aortic stenosis and are the result of recurrent subaortic stenosis [20], a substantial numbers are performed because of aortic valve pathology. In most cases reoperation is caused by the aortic insufficiency that can result from subvalvular aortic stenosis.

The possibility of obstruction to coronary artery blood flow in patients with SVAS has been recognized for lately. Edwards [23] documented adhesion of the right aortic cusp to the site of the aortic obstruction thus resulting in a blind sac from which the right coronary artery arose. The surgical importance of this mechanism of coronary ischemia has led to modifications in corrective techniques. We believe resection of as much supravalvar membrane as possible is also important not only to improve access to blood flow but also to free up the cusps to allow greater mobility. Some patients have anatomic coronary ostial stenosis that may necessitate separate patch enlargement or extension of the aortic patch into the stenotic coronary ostium [20,21,24]. In addition to isolation of a coronary arterial orifice, the coronary arteries may be of concern in patients with SVAS for several reasons. As was the case in two of our patients and several previously reported cases, stenosis [16] or atresia [25] of a coronary ostium and proximal coronary artery may occur, necessitating either ostial endarterectomy, coronary bypass grafting, or other reconstructive techniques [25]. As several authors have noted [10,21], dilatation of the arterial system may occur in patients with this lesion, and may be associated with medial thickening and intimal fibrosis. Although the long-term significance of this process has not been confirmed clinically, it is intuitive to suppose that such patients may be at increased risk of developing atherosclerotic coronary artery disease.

Our experience indicates that enlargement of the aortic root with a standard patch leads to excellent long-term results, although this technique may not completely restore the geometry of the aortic sinuses. Important technical points include using standard patch rather than a pantalon-shaped patch and patch large enough to restore the aorta to a normal diameter.

In previous surgical series, procedures on the aortic valve have been performed in up to 42% of patients undergoing repair of SVAS. In most cases, simple commissurotomy was performed for partial fusion of a bicuspid valve. One-third to half of the procedures performed on the aortic valve in these series, however, has been valvar replacement with a mechanical or bioprosthesis. In the present era, the use of prostheses for replacement of the aortic valve in children is controversial, but the Ross procedure has provided another, arguably superior, option in many patients requiring replacement [21]. In addition, congenital heart surgeons today have become more aggressive about salvaging the native valve with valvuloplasty, whether it is augmentation,
The role of the Ross procedure in the management of SVAS with associated valvular, with or without subvalvular, pathology has not been elucidated. Over the past several years, this procedure has become an important strategy in the management of systemic outflow tract disease in children. In addition to older patients with combined aortic regurgitation and stenosis, replacement of the aortic valve with the pulmonary autograft has revolutionized the surgical management of complex, multilevel obstruction of the left ventricular outflow tract in infants and young children. We have performed a Ross procedure in four patients with SVAS, an additional six patients underwent a Ross procedure after repair of SVAS, and all have done well.

We believe that the standard patch aortoplasty provides a more anatomical repair for SVAS than other techniques. The procedure can be done at no greater operative risks and will provide excellent relief of the pressure gradient between the left ventricle and the aorta.

References