Mid-term outcome after surgical repair of congenital supravalvular aortic stenosis by extended aortoplasty

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Abstract

OBJECTIVES: Congenital supravalvular aortic stenosis (SVAS) is a rare arteriopathy associated with the Williams–Beuren syndrome (WBS) and other elastin gene deletions. Our objective was to review the mid-term outcomes of SVAS repair with extended aortoplasty.

METHODS: Congenital SVAS repairs from 2001 to 2010 were retrospectively reviewed. The follow-up records, reintervention and reoperation data and most recent echocardiograms were obtained.

RESULTS: From 2001 to 2010, 21 patients (15 males) underwent surgical repair of SVAS by extended aortoplasty with autologous pre-treated pericardium, which is a modification of the Doty technique. The mean age was 3.1 ± 4.2 years. WBS was diagnosed in 14 of the patients. There was no early mortality, but one late death was observed. At the latest follow-up (mean follow-up, 4.3 ± 2.9 years; range, 1–108 months), echocardiograms revealed a peak Doppler gradient across the aortic outflow tract of 15 ± 8 mmHg. The majority of the patients had minimal to mild aortic insufficiency. No reoperation or reintervention was required.

CONCLUSIONS: Extended aortoplasty provides excellent mid-term relief of SVAS and, in addition, reshapes the aortic root geometry to a much more favourable anatomical configuration. It can be performed without any increase in operative risks. The mid-term results are excellent.

Keywords: Supravalvular aortic stenosis • Outcome • Williams–Beuren syndrome

INTRODUCTION

Supravalvular aortic stenosis (SVAS) is an uncommon congenital cardiac anomaly characterized by varying degrees of left ventricular outflow tract obstruction that begins distal to the aortic valve and is also associated with Williams–Beuren syndrome (WBS) [1, 2]. Although it is not primarily involved, the more proximal part of the aortic root may undergo secondary pathological changes that may result in aortic valve insufficiency or coronary ischaemia [1–5]. 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 [2–6]. The surgical repair for SVAS has evolved considerably over the last 20 years. Initially, the one-patch aortoplasty technique described by McGoon et al. [2] addressed the stenotic aortic area with patch extension into the non-coronary sinus of Valsalva. The surgical technique then progressed to restore a more geometric configuration of the aortic root. The ‘Doty’ technique involves the placement of an inverted Y-shaped incision across the supravalvular-constricting aortic ring and into the right coronary and non-coronary sinuses of Valsalva, followed by the insertion of a pantaloon-shaped Dacron patch [1].

The aim of this study was to provide a mid-term follow-up of our experience with extended aortoplasty using an autologous glutaraldehyde-pretreated pericardial patch as a modification of the Doty technique.

PATIENTS AND METHODS

A retrospective review of the Leipzig Heart Center congenital cardiothoracic surgical database identified all of the patients who underwent surgery for SVAS from 2001 to 2010. Reviews of the medical charts and the database were used to determine demographics, operative details, perioperative events and complications. A mid-term follow-up was obtained from a combination of the cardiology and cardiac surgery database, the Leipzig Heart Center electronic medical records, the cardiology charts and the echocardiogram database.

Surgical technique

The surgical reconstruction consisted of a median sternotomy, a cardiopulmonary bypass with bicaval cannulation and a left ventricular venting through the right upper pulmonary vein. St. Thomas cardioplegia was used for myocardial protection. An extended aortoplasty...
with a glutaraldehyde-pretreated autologous pericardial patch in the shape of an inverted Y with extensive endarterectomy was performed (Fig. 1). A longitudinal aortotomy was extended from the non-stenotic segment of the ascending aorta distal to the supravalvular stenosis, deep into the right coronary and non-coronary sinuses of Valsalva (Fig. 2). The supravalvular ridge was excised carefully, and procedures on the aortic valve, root and subvalvular region were performed when necessary. The aorta was then augmented with a Y-shaped glutaraldehyde-pretreated autologous pericardial patch using a continuous non-absorbable monofilament suture (Fig. 3).

Follow-up and statistical analysis

The data were compiled and analysed using the BIAS software (BIAS for Windows, Version 7). The continuous variables are reported as the mean ± standard deviation for the normally distributed data or as the median and interquartile range for the non-normally distributed data. The continuous variables are expressed as the mean ± standard deviation, and the categorical variables are expressed as proportions throughout the manuscript. The statistical significance was defined as a P-value of <0.05.

RESULTS

A total of 21 patients underwent repair for SVAS at our institution from 2001 to 2010. The mean age of the patients was 3.1 ± 4.2 years (median: 1.56 years); 71% were male, and 29% were female. The mean weight was 15 ± 13.6 kg. All of the patients received the modified ‘Doty’ aortoplasty. Of the 21 patients, 14 had WBS.

Table 1 contains concomitant surgical procedures. The mean aortic cross-clamp time was 44 ± 24 min, and the mean bypass time was 112 ± 76 min (Table 2). No early death was observed. There were no major complications or mediastinitis during the postoperative period. The mean length of hospital stay was 8 ± 6 days. The peak aortic systolic gradient on echocardiogram at hospital discharge was 20 ± 10 mmHg. This value was significantly reduced from the preoperative peak gradient of 77 ± 34 mmHg. At hospital discharge, all of the patients reported no more than a mild aortic regurgitation. The follow-up data were obtained from

Figure 1: A longitudinal aortotomy with extension from the non-stenotic segment of the ascending aorta distal to the supravalvular stenosis.

Figure 2: Surgeons view on the aortic root with attention of aortotomy extended deep into the right and non-coronary sinuses.

Figure 3: The augmentation of the aortic root with a Y-shaped pericardial patch using continuous non-absorbable monofilament suture.

Table 1: Associated cardiovascular anomalies (n)

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>n</th>
</tr>
</thead>
<tbody>
<tr>
<td>Branch and/or peripheral pulmonary artery stenosis</td>
<td>5</td>
</tr>
<tr>
<td>Aortic arch vessel stenosis</td>
<td>2</td>
</tr>
<tr>
<td>Infundibular and/or valvular pulmonary stenosis</td>
<td>3</td>
</tr>
<tr>
<td>Valvular aortic stenosis</td>
<td>7</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>2</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>2</td>
</tr>
<tr>
<td>Subvalvular aortic stenosis</td>
<td>4</td>
</tr>
</tbody>
</table>
Table 2: Patient characteristics and perioperative data

<table>
<thead>
<tr>
<th>Variable</th>
<th>n (men/women)</th>
<th>Age (years)</th>
<th>Weight (kg)</th>
<th>Williams-Beuren syndrome (n)</th>
<th>Presence of bicuspid aortic valve (n)</th>
<th>Aortic cross-clamp time (min)</th>
<th>Total bypass time (min)</th>
<th>Rectal temperature (°C)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>21 (15/6)</td>
<td>3.1 ± 4.2</td>
<td>15 ± 13.6</td>
<td>14</td>
<td>3</td>
<td>44 ± 24</td>
<td>112 ± 76</td>
<td>28 ± 3.5</td>
</tr>
</tbody>
</table>

Table 3: Echocardiographic data

<table>
<thead>
<tr>
<th>Variable</th>
<th>Preoperative</th>
<th>At discharge</th>
<th>At the latest follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild AR</td>
<td>1</td>
<td>19</td>
<td>19</td>
</tr>
<tr>
<td>Moderate AR</td>
<td>0</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>$P_{mean}$ (mmHg)</td>
<td>77</td>
<td>34</td>
<td>20 10 15 ± 8</td>
</tr>
</tbody>
</table>

the cardiovascular surgery database, cardiology charts, hospital electronic medical records and the echocardiogram database. These data were obtained for all of the patients who survived to discharge. The mean duration of the follow-up was 4.3 ± 2.9 years. There was 1 late sudden cardiac death. No reoperations or reinterventions were required. At the last follow-up, the mean peak gradient was 15 ± 8 mmHg. The aortic regurgitation was graded as mild in 19 patients and moderate in 1. At the last follow-up, this patient presented an asymptomatic clinical course. There was no sign of coronary ischaemia on a routine electrocardiogram (Table 3).

Comments

At first glance, SVAS appears to be a relatively simple lesion. Attention is usually focused on the constricting supravalvular ring. However, SVAS is actually a complex lesion that often involves the entire left ventricular outflow tract [4–5]. The primary abnormality involves intimal hyperplasia and a thickening of the aortic media immediately above the sinuses of Valsalva, which results in a narrowing of the aorta [4]. This fibrosing ring can have many secondary effects. The sinuses themselves may be hypoplastic. The stenosing ring often affects the relationships of the commissures, which leads to a distortion of the aortic cusps, particularly the free edges [3]. Occasionally, the free edges of the cusp may even fuse to the aortic wall, effectively isolating the sinuses of Valsalva and the coronary ostia from the lumen of the aorta [4]. A significant number of patients also present dysplasia and irregular fibrous thickening of the aortic cusps [5].

Multiple surgical approaches have been described to relieve SVAS, including Doty’s patch aortoplasty and Brom’s three-patch technique [1–4]. We have adopted this technique since 2001 for all of our patients regardless of the associated lesions in the left ventricular outflow tract. In our opinion, the advantages of this technique include the complete augmentation of the supravalvular narrowing, a reshaping of the physical and symmetrical geometry of the aortic root, and the prevention of growth limitations by patch augmentation of the right coronary and non-coronary sinuses [1].

In our series and in other reports, the majority of SVAS patients had WBS, which is an underlying elastin arteriopathy involving the aorta and the pulmonary arteries [5]. This elastin disorder results from a microdeletion of the elastin gene on chromosome 7q11.23. Because of the reduction in elastin production, the shear stress in the aorta may result in smooth muscle hypertrophy and increased collagen deposition, causing medial thickening. Furthermore, intimal thickening may occur as a result of fibrous growth [5]. The elastin arteriopathy may range from the discrete type of narrowing of the supravalvular area to the diffuse type that extends the narrowing throughout the aortic arch and branch vessels [5]. In addition, the elastin arteriopathy may also involve other large arteries, such as the pulmonary arteries, either at the central or branching level. The natural history of the pulmonary arterial stenosis may decrease spontaneously over time, which limits the surgical indications in these patients [5–8].

Our study demonstrates the use of a modified Doty technique with no operative mortality and a low complication rate for the repair of SVAS. Based on our experience, the associated lesions in the left ventricular outflow tract, subvalvar stenosis and the supra-aortic vessels should be aggressively addressed during the initial surgical repair. Whether an operation is indicated should be decided at the time of diagnosis. The mid-term follow-up confirms that this modified Doty technique is associated with a low reoperation rate and a low incidence of aortic regurgitation; similar observations have also been made by several other groups [4–8]. We continue to use this technique routinely in patients with SVAS.

Conflict of interest: none declared.

REFERENCES