Modified simple sliding aortoplasty for preserving the sinotubular junction without using foreign material for congenital supravalvar aortic stenosis

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Abstract

Objective: The surgical approach for treating supravalvar aortic stenosis (SVAS) has evolved from a plain patch technique to a three-dimensional patch repair, which has some drawbacks. Here, we report on the midterm outcomes after using our modified simple sliding aortoplasty preserving sinotubular junction without foreign material for surgical correction of SVAS.

Methods: Between June 2001 and February 2010, 18 children (median age, 6.2 years; range 0.5—2.2 years) with discrete SVAS underwent surgical repair. After a standard median sternotomy, cardiopulmonary bypass and cardioplegic arrest, the aorta was transected obliquely just distal to the point of stenosis. An incision was then made into the non-coronary sinus of the proximal aorta, and a counterincision was made into the lesser curvature of the ascending aorta, after which, the proximal and distal aorta were anastomosed directly with a running suture. Peak pressure gradients were estimated using echocardiography and the data were reviewed retrospectively.

Results: The median follow-up period was 39.6 months (range, 1—104.5 months). There was no early or late death. No patients required re-operation. Ten patients had Williams—Beuren syndrome. There were eight cases of concomitant pulmonary artery angioplasty. The mean pressure gradient decreased from 65.9 ± 18.4 mmHg preoperatively to 15.2 ± 8.9 mmHg at the final follow-up (P = 0.01). There was no significant, more than mild aortic regurgitation.

Conclusions: Our modified simple sliding aortoplasty showed excellent surgical results, and may be a good option for discrete SVAS.

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Keywords: Congenital supravalvar aortic stenosis; Williams—Beuren syndrome

1. Introduction

Congenital supravalvar aortic stenosis (SVAS) is a relatively uncommon obstructive lesion of the left-ventricular outflow tract. The typical feature of this malformation is an aortic narrowing at the level of the sinotubular junction (STJ), although, in some cases, there is narrowing of the entire ascending aorta and arch branches [1]. The first successful surgical relief for SVAS was reported in 1961 [2]. Since then, the surgical approach has evolved from the plain patch technique to a three-dimensional patch repair [3—6]. However, no technique has demonstrated clear superiority.

We have modified a previously described sliding aortoplasty approach for preserving the STJ without using foreign material for surgical correction of SVAS. Our modified approach is quicker and easier, as it does not require the creation of three identical sinuses. The present study examined the midterm outcomes following the use of this method.

2. Material and methods

2.1. Study patients

This retrospective study involved all patients younger than 18 years of age, who underwent surgical repair for discrete congenital SVAS at our institution between June 2001 and February 2010. Early and late outcomes were reviewed. The study was approved by our institution ethics committee/institutional review board.

The study involved 18 patients, 13 male and five female. Ten patients had Williams—Beuren syndrome (56%). The median age at SVAS surgery was 6.2 years (range: 6 months to 12.2 years). The median body weight was 22.5 kg (range: 7.8—47.9 kg).

No patient had undergone a prior sternotomy or SVAS repair. One or more concomitant procedures were performed...
in 10 patients (Table 1). One patient (6%) had a bicuspid aortic valve, and underwent balloon valvuloplasty before concomitant aortic valvotomy at SVAS repair. There was no co-arctation of the aorta or subaortic stenosis.

2.2. Surgical techniques

A standard median sternotomy was used for all patients. Cardiopulmonary bypass was instituted with a cannula for arterial return in the ascending aorta and a venous single cannula in the right atrium. On bypass, meticulous dissection around the arch vessels was performed to reduce tension on the future suture line. Cardiac arrest was achieved using a cold antegrade cardioplegic solution. The aorta was transected obliquely just distal to the point of stenosis, and the narrowest segment several millimeters above the STJ was cut and resected down to the level just above the commissure. An incision was then made into the non-coronary sinus of the proximal aorta anterior, and a counterincision was made into the lesser curvature of the ascending aorta posteriorly to create the appropriate diameter for the new STJ (Fig. 1(A)) [7]. The tethered fibrous tissue was then excised, and the thickened commissural tissue was mobilized. The proximal and distal aorta were then anastomosed directly with a 5/0 or 6/0 Prolene running suture (Ethicon, Somerville, NJ, USA) as is routine in arterial switch operations (Fig. 1(B)).

2.3. Data analysis

Peak pressure gradients were estimated using echocardiography (by the Bernoulli equation). Medical records were reviewed retrospectively. Data are presented as medians with ranges or means with standard deviations. Continuous variables were compared using Wilcoxon’s signed rank test. A $P$ value less than 0.05 was set as the level of statistical significance.

Analysis was performed using Statistical Package for Social Sciences (SPSS) for Windows version 17.0 (SPSS Inc., Chicago, IL, USA).

3. Results

All patients completed follow-up, and the median follow-up period was 39.6 months (range, 1—104.5 months). The mean cardiopulmonary bypass time was $93.66$ min and the mean cross-clamp time was $32.14$ min. There were no early or late deaths. All patients were discharged from the hospital with no complications except for one patient, who experienced postoperative accidental hypoxic brain damage — that patient remains in hospital for rehabilitation. All patients underwent postoperative echocardiography or computed tomographic (CT) scans, all of which showed good surgical correction results (Fig. 2). No patients required re-operation. The mean preoperative pressure gradient was $65.9 \pm 18.4$ mmHg, the mean immediate postoperative pressure gradient was $17.5 \pm 5.5$ mmHg, and the mean final follow-up pressure gradient was $15.2 \pm 8.9$ mmHg, which was lower than the mean preoperative values ($P = 0.01$) (Fig. 3). There were three (17%) cases of mild aortic regurgitation (AR) (Table 2). The median hospital stay was 7 days (range 5—41 days, excluding the one patient who

<table>
<thead>
<tr>
<th>Procedure</th>
<th>$n$</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Branch pulmonary angioplasty</td>
<td>8</td>
<td>44.4</td>
</tr>
<tr>
<td>LV myotomy</td>
<td>1</td>
<td>5.6</td>
</tr>
<tr>
<td>Aortic valvotomy</td>
<td>2</td>
<td>11.1</td>
</tr>
<tr>
<td>Pulmonary valvotomy</td>
<td>1</td>
<td>5.6</td>
</tr>
<tr>
<td>MPA angioplasty</td>
<td>2</td>
<td>11.1</td>
</tr>
<tr>
<td>Tricuspid annuloplasty</td>
<td>1</td>
<td>5.6</td>
</tr>
</tbody>
</table>

*Abbreviations: LV, left ventricle; MPA, main pulmonary artery.*
remains in hospital). The mean ratio between the aortic annulus diameter and STJ diameter of echocardiographic data from the last follow-up was $91.4 \pm 5.1\%$ (median ratio $91.5\%$, range $80.9—97.8\%$).

4. Discussion

The defining feature of congenital SVAS is an aortic narrowing at the level of the sinotubular junction (STJ), which results in the typical hourglass appearance. This distorted geometry of the aortic root has important implications for surgical therapy. The aortic root is a dynamic rather than static structure. Brewer et al. [8] reported an aortic root dimensional change of 16% during each cardiac cycle, which would result in a marked fluctuation of the fatigue stress on the tissue. The normal root and flexible STJ expand in systole to aid in flattening and straightening of the aortic leaflet. Lansac et al. [9] also reported asymmetric aortic root expansion in an animal (sheep) study. They observed asymmetrical expansion of the aortic root. During systole, each sinus of Valsalva differed in terms of perimeter area expansion: the right expanded more than the left, and more than the non-coronary sinus. This asymmetry resulted in changes in the root’s twist and tilt angles. During systole, the curved cylindrical root became straighter. This geometric change might act as a shock-absorber mechanism to reduce shear stress on the leaflets while left ventricular (LV) pressure is dramatically falling. In SVAS, the STJ is narrowed and inflexible, and the redundant free edges of the aortic leaflets are apparently prone to degeneration [10]. Although the main goal of surgical intervention is to relieve obstruction to the outflow tract, consideration should be given to restoring the physiological anatomy of the aortic root [11]. The aim of SVAS surgery is to restore STJ flexibility and size, and enlarge the narrowing lesion.

STJ size plays a crucial role in the optimization of aortic valve competence. Furukawa et al. [12] created a model of STJ dilatation using canine hearts and reported that dilatation of the STJ with outward deviation of the commissure caused AR, regardless of sinus dilatation. Indeed, a larger-than-necessary STJ was invariably associated with tethering and bending of the free margin of the aortic leaflets, which reduced the co-aptation reserve, causing valve regurgitation. Kunzelman et al. [13] analyzed the mathematical relationship between normal non-pressurized human aortic root components and found that STJ and aortic valve annulus diameters were $81\%—2\%$ and $97\%±2\%$ of the diameter at the sinus level, respectively.

The ‘adjustable STJ’ principle can represent the solution for both overestimation and underestimation of the neo STJ, which can result, respectively, in a residual AR or improper co-aptation level of the aortic valve leaflets [14]. Considering the dynamic aortic root and important size of STJ, what is the adequate size of the STJ? Maselli et al. [15] suggested that the optimal ratio between the aortic annulus diameter and STJ diameter in a reconstructed aortic root is 1:1. However, Kunzelman reported the STJ diameter is 10—15% smaller than the diameter of the annulus [13]. In our study, the mean ratio between the aortic annulus diameter and STJ diameter was $91.4 \pm 5.1\%$, which was comparable to Kunzelman’s study. Therefore, we propose that a neo-STJ diameter range from 90% to 100% of the annulus diameter to preserve proper coaptation of aortic valve.
Surgical repair for SVAS was first accomplished by insertion of a diamond-shaped prosthetic patch across an incision into the non-coronary sinus of Valsalva [2]. Most operative techniques use patch material to enlarge the STJ of one or more sinuses [2—5]. However, a prosthetic patch cannot maintain a flexible STJ, and cannot handle the increase in aorta size as the patient grows. The most widely used technique is Brom’s three-patch method in which the three Valsalva sinuses are made to the same size, which is not the natural situation [7]. As long ago as 1975, McAlpine reported that, under normal physiological conditions, the differences in annular circumferences compassing the various sinuses can be as great as 20—25% [16]. Kunzelman et al. [13] also found that the non-coronary was the largest of the three leaflets, and the right tends to be the smallest. As such, using our present technique, we did not aim to make the Valsalva sinuses same size. We just need to restore flexibility and adequate STJ size, and do not need to make sinus symmetry. Indeed, the sinuses are not symmetrical. In 1993, Myers et al. [6] reported on an all-autologous three-sinus repair in which the distal ascending aorta flaps were used to augment the STJ. Scott et al. [17] described that the advantages of this technique were improved growth potential and preservation of more normal symmetry over time when compared with prosthetic patch techniques. However, that technique is time-consuming and complex as it requires making three symmetrical sinuses. In addition, it is difficult to create an adequate neo-STJ size using that approach, which is important for maintaining a constant strain to minimize fatigue stress [10]. To address those issues, we developed the present technique, which is a previously described modified simple sliding aortoplasty-preserving STJ [7]. Our technique is easier, faster, does not require the creation of three individually sized sinuses, and allows for growth as the patients mature. There were no mortalities or re-operations required. The mean cross-clamp time was 32 ± 14 min, which is shorter than reported for other methods (almost 60 min for patch repair) [17—19], and is almost less than half the time reported by others for Brom’s repair cross-clamping (65 ± 17 min and 104 ± 21 min) [18,19]. There were three cases of mild late AR in the present study, with one due to aortic valve commissurotomy. By comparison, a study of Brom’s repair reported one moderate AR and five mild ARs in eight patients [19] (Table 3). We believe our focus on restoring STJ flexibility and adequate size, as well as enlarging the narrowing lesion, explains the present low incidence of AR and no re-operations. In addition, we believe the good surgical outcomes and nil mortality are because this

### Table 2. Patients characteristics.

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Age (year)</th>
<th>Gender</th>
<th>Body weight (kg)</th>
<th>Preoperative Pr. gradient (mmHg)</th>
<th>Last postoperative Pr. gradient (follow-up interval, months)</th>
<th>Pr. gradient reduction (mm Hg)</th>
<th>AR</th>
<th>Length of follow-up (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3.3</td>
<td>Male</td>
<td>13.3</td>
<td>85</td>
<td>22 (81.2)</td>
<td>63</td>
<td>Mild</td>
<td>104.5</td>
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<tr>
<td>2</td>
<td>8.2</td>
<td>Male</td>
<td>29.6</td>
<td>80</td>
<td>19 (29.0)</td>
<td>61</td>
<td>Trivial</td>
<td>103.4</td>
</tr>
<tr>
<td>3</td>
<td>5.6</td>
<td>Male</td>
<td>11.1</td>
<td>45</td>
<td>11 (62.6)</td>
<td>34</td>
<td>None</td>
<td>96.7</td>
</tr>
<tr>
<td>4</td>
<td>12.2</td>
<td>Male</td>
<td>42.4</td>
<td>82</td>
<td>30 (68.8)</td>
<td>52</td>
<td>Trivial</td>
<td>70.9</td>
</tr>
<tr>
<td>5</td>
<td>9.4</td>
<td>Female</td>
<td>23.7</td>
<td>46</td>
<td>0 (0.2)</td>
<td>46</td>
<td>None</td>
<td>67.2</td>
</tr>
<tr>
<td>6</td>
<td>7.0</td>
<td>Male</td>
<td>16.9</td>
<td>58</td>
<td>16 (60.2)</td>
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<td>Trivial</td>
<td>61.6</td>
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<tr>
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<td>4.2</td>
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<td>18.8</td>
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<td>20 (6.9)</td>
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<tr>
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<td>Female</td>
<td>47.9</td>
<td>63</td>
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<td>50.0</td>
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<tr>
<td>9</td>
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<td>Female</td>
<td>7.8</td>
<td>92</td>
<td>10 (38.9)</td>
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<td>Trivial</td>
<td>43.3</td>
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<td>10</td>
<td>10.1</td>
<td>Male</td>
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<td>90</td>
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<tr>
<td>11</td>
<td>5.8</td>
<td>Female</td>
<td>23.2</td>
<td>45</td>
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<td>12</td>
<td>12.7</td>
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<td>38.7</td>
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<td>8 (18.7)</td>
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<tr>
<td>13</td>
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<td>21.8</td>
<td>65</td>
<td>7 (13.7)</td>
<td>58</td>
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<td>23.0</td>
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<tr>
<td>14</td>
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<td>Male</td>
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<td>97</td>
<td>8 (2.7)</td>
<td>90</td>
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<tr>
<td>15</td>
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<td>Male</td>
<td>11.5</td>
<td>38</td>
<td>8 (6.6)</td>
<td>30</td>
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<td>11.0</td>
</tr>
<tr>
<td>16</td>
<td>1.0</td>
<td>Male</td>
<td>13</td>
<td>50</td>
<td>8 (0.4)</td>
<td>42</td>
<td>None</td>
<td>2.7</td>
</tr>
<tr>
<td>17</td>
<td>11.7</td>
<td>Female</td>
<td>45.6</td>
<td>60</td>
<td>27 (0.2)</td>
<td>33</td>
<td>Mild</td>
<td>1.0</td>
</tr>
<tr>
<td>18</td>
<td>1.1</td>
<td>Male</td>
<td>9.3</td>
<td>74</td>
<td>31 (0.1)</td>
<td>43</td>
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<td>1.0</td>
</tr>
<tr>
<td>Median</td>
<td></td>
<td></td>
<td>6.2</td>
<td>22.5</td>
<td>13.5</td>
<td>46.5</td>
<td>None</td>
<td>39.6</td>
</tr>
</tbody>
</table>

**Abbreviations:** AR, aortic regurgitation.

### Table 3. Results reported in studies of supravalvar aortic stenosis surgery.

<table>
<thead>
<tr>
<th>Study</th>
<th>Year</th>
<th>Aortic cross-clamp time, min</th>
<th>Aortic regurgitation</th>
<th>Early death</th>
</tr>
</thead>
<tbody>
<tr>
<td>Scott et al.</td>
<td>2009</td>
<td>63 ± 21</td>
<td>Mild (10/23, 43.3%)</td>
<td>1/25 (4%) 1 Myers repair</td>
</tr>
<tr>
<td>Metton et al.</td>
<td>2009</td>
<td>54 ± 15</td>
<td>Moderate (1/23, 4.4%)</td>
<td></td>
</tr>
<tr>
<td>Kaushal et al.</td>
<td>2010</td>
<td>60 ± 14</td>
<td>One-patch repair (mild 3, moderate 4)</td>
<td>1/20 (5%) 1 One-patch repair</td>
</tr>
<tr>
<td>Current study</td>
<td>2018</td>
<td>32 ± 14</td>
<td>Trivial (4/18, 22.2%)</td>
<td>0 (0%)</td>
</tr>
</tbody>
</table>
technique does not require the creation of three symmetrical sinuses, and is hence quicker, and the result is more ‘physiological’.

In conclusion, our modified simple sliding aortoplasty technique showed excellent surgical results with no mortality, no re-operation, and no significant AR. This technique may be a good option for discrete SVAS.

References


Appendix A. Conference discussion

Dr V. Rybin (Moscow, Russia): Preservation of aortic valve competence during correction of supravalvar aortic stenosis represents a real surgical challenge. I believe in the valuable method described by Myers and his colleagues in 1993 where they warned against the use of prosthetic materials for aortic root enlargement. Your results clearly demonstrate the importance of asymmetrical enlargement of the aortic root. I have a couple of questions I would like to ask.

Presenting your surgical technique you said, ‘... the aorta was transected obliquely just distal to the point of stenosis.’ However, on your picture it seems that you have removed the narrowed part of the ascending aorta prior to making an incision into the non-coronary sinus. Can you comment on this important point?

And the second one: What would you do if you found a bicuspid aortic valve or a diffuse narrowing of the ascending aorta during surgery?

Dr Shin: Let me show you my operative technique. We usually cut the incision into the most stenotic part and excise the most stenotic portions. And after that, we incise the non-coronary sinus and bevel it to the lesser curvature, so we create a new sinotubular junction.

And regarding your second question, we encountered only one case of bicuspid aortic valve.

Dr M. Haw (Southampton, UK): Looking at the technique, I had not been aware of this before, but it looks like a very viable option. But do you treat every case of supravalvar stenosis this way? Because it strikes me that in some of the more severe forms of anatomy, the left coronary artery can be really involved in the area of narrowing, in which case the incision would be rather dangerous.

And also the post commissures of the valve are involved in the narrowing. You might chop the top of the valve off; have you got any comments about that?

Dr Shin: You mean the left coronary artery?

Dr Haw: The left coronary, in my experience anyway, seems, in some of the more severe forms of this, to be very high up in the sinus right involved in the shelf. So if you were to excise the whole of the shelf, you would be almost on your left coronary.

And the second point was that in the more severe forms, the commissures and even some of the tissue of the valve itself are also involved, in which case you might well be removing part of the valve structure.

Dr Shin: Our incision is uppermost, a safe distance from coronary os or aortic valve commissure. So there is no concern about damaging the coronary artery or valve commissure. Don’t worry.

Dr W. Brawn (Birmingham, UK): It has been alluded to in the past, but often the disease is much more extensive than illustrated in your diagram. And I just wonder whether all the patients that you have with supravalvar aortic stenosis have been able to be dealt with in this way. Because in our experience, it is not unusual for the disease to extend into the aortic arch or even into the head vessels. The last speakers mentioned proximal aortic valve involvement as well. So it is often more diffuse than just a discrete lesion.

And then, it would seem to me you cannot apply this to all lesions, and I wonder whether or not you have had cases where you have not been able to use this particular technique.

Dr Shin: I did not catch that.

Dr Brawn: How many have you not been able to use this technique on? Have you applied this to all patients that you see with supravalvar aortic stenosis?

Dr Shin: For all discrete supravalvar aortic stenosis.

Dr Brawn: And the other ones that do not have this sort of localized lesion?

Dr Shin: No. All patients.

Dr Brawn: All patients?

Dr Shin: Yes.

Dr Brawn: So you do not see more diffuse lesions. The other comment, then, would be that there is a difference in ethnicity and the type of lesion you see in different countries because this certainly happens with other lesions. But certainly in our population, it is usually much more diffuse than this.

Dr Seo: I am an operator of these operations, and I will just comment to Dr Brawn and the previous speaker.

Yes, this is just discrete cases. In many cases, supravalvar stenosis is mainly diffuse with progressive disease. In such cases, we have to tailor the technique to the individual, but we can adapt this principle to the supravalvar area, not indicating for left main or ostial coronary lesion and diffuse arch type stenosis.

Even in Asia in the diffuse type arch-involving cases, we can use this concept to augment the ascending aorta and the transverse arch. Our objective is not to use prosthetic material and to maintain the sinotubular junction in those group of patients.