Early and mid-term outcomes after surgical repair of congenital supravalvular aortic stenosis with the Doty technique

Doğuştan supravalvüler aort stenozunun onarımında Doty tekniğinin erken-orta dönem sonuçları

Objective: The aim of this study was to evaluate the efficacy and safety of the Doty surgical approach in pediatric patients with congenital supravalvular aortic stenosis (SVAS) by examining early and mid-term outcomes.

Methods: Surgical repair using the Doty technique was performed in a total of 10 pediatric patients with SVAS between January 2005 and July 2015 at this clinic. These patients were evaluated retrospectively. Demographic characteristics, echocardiographic findings, and clinical outcomes were analyzed.

Results: The mean patient age was 4.8±3.9 years. Williams-Beuren syndrome was diagnosed in 4 of the patients. Early mortality was observed in 1 patient with diffuse SVAS. At the final follow-up (mean follow-up: 3.7±1.2 years; range: 6-61 months), echocardiograms revealed a mean pressure gradient of 14±4.2 mmHg. Two patients displayed minimal aortic insufficiency during the follow-up period. No reoperation or reintervention was required.

Conclusion: The Doty technique is an anatomically and technically effective surgical approach to treating SVAS.

Amaç: Doğuştan supravalvüler aort stenozu olan pediyatrik hastalarda Doty cerrahi yaklaşımın etkinliği güvenilirliği, kısa ve orta dönem sonuçlarını değerlendirmek.


Bulgular: Ortalama yaş 4.8±3.9 yıl idi. Williams-Beuren sendromu dört hastada tespit edildi. Erken mortalite diffuse supravalvüler aort stenozu olan bir hastada görüldü. En son yapılan kontrollerde (ortalama takip süresi 3.7±1.2 yıl; dağılım, 6-61 ay) ekokardiyografide ortalama basınç farkı 14±4.2 mm Hg olarak saptandı. Takiplerde iki hasta hafif aort yetersizliği saptandı. Hiçbir hastada yeniden cerrahi veya girişim gerekmedi.

Sonuç: Doty tekniği anatomik ve tekniğin etkin bir cerrahi yaklaşımıdır.

Congenital supravalvular aortic stenosis (SVAS) is the least common type of left ventricle outflow tract obstruction.[1,2] The disease is closely associated with chromosome 7 microdeletion (7q11.23), and is characterized by elastin arteriopathy. Williams–Beuren syndrome (WBS), a form of SVAS, causes reduced elasticity, increased shear stress, collagen deposition, and thickening of the aortic media.[2,3] Although arteriopathy is defined as stenosis of the aortic lumen at the level of the sinotubular junction, diffuse involvement of the ascending aorta, aortic arch and
branches, and the descending aorta, or involvement at different levels may also be observed.\cite{1,4,5}

SVAS requires a surgical repair because the natural history of SVAS is progressive.\cite{1,4,7} There are several surgical techniques available to repair SVAS. The first successful surgical repair was performed by McGoon in 1961 using a single-patch technique.\cite{8} Since then, various surgical modifications have been developed.\cite{9–13} The objective of this study was to present clinical experience and early and mid-term outcomes of patients who underwent surgery performed with the Doty technique.

### METHODS

The medical records of patients who underwent surgery between January 2005 and July 2015 with the diagnosis of congenital SVAS were retrospectively evaluated. The demographic data, perioperative findings, morbidity, mortality, and postoperative control echocardiographic findings of 10 patients were analyzed. Echocardiography and cardiac catheterization was conducted before surgical intervention in all cases. Computed tomography angiography was performed to rule out suspected coronary involvement and to evaluate the detailed morphology of SVAS and pulmonary arteries (PAs) in 4 patients before the surgical procedure because cardiovascular magnetic resonance imaging is not available at this center.

A median sternotomy incision was performed in the surgical procedure. Following systemic anticoagulation, arterial cannulation was performed from the aorta, and bicaval venous cannulation was performed selectively from the superior vena cava and the inferior vena cava. Right superior pulmonary venous cannulation was also performed. After initiation of cardiopulmonary bypass, diastolic arrest was achieved under moderate hypothermia using antegrade blood cardioplegia and topical application of cold. In all of the patients, the repair was performed using the technique defined by Doty et al.\cite{7} Discrete stenosis was detected in 9 patients, and diffuse stenosis was observed in 1 (Fig. 1a-d). The stenosis was repaired using an incision extending from the distal part of the stenosis to the non-coronary and right coronary sinus and with a reverse Y-shaped Dacron patch. The repair was performed using a non-absorbable, monofilament suture material. A supravalvular ridge was excised.

Statistical analysis was performed using SPSS for Windows, Version 16.0 (SPSS Inc., Chicago, IL, USA). Continuous variables were expressed as median and minimum-maximum, while categorical variables were expressed as a percentage (%). Statistical significance was defined as a p value of <0.05.

### RESULTS

Demographic and clinical characteristics of the patients are provided in Table 1. Operative and postoperative details, as well as echocardiographic measurements are listed in Table 2.

Of a total of 10 patients who were operated on with the diagnosis of SVAS, 6 (60%) were male and 4 (40%) were female. The mean age was 4.8±3.9 years (range: 0.5 to 8 years). The mean weight of the patients was 17±10.1 kg (range: 6 to 25 kg).

WBS was diagnosed in 4 (40%) patients. None of the patients had stenosis in the main PAs. The gradient of 2 patients with peripheral pulmonary stenosis was found to be 40 mm Hg and 55 mm Hg, re-

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![Figure 1. Preoperative angiography images revealing severe supravalvular aortic stenosis. (A) Discrete stenosis, (B) diffuse stenosis. (C, D) A preoperative computed tomography image demonstrating hourglass-type supravalvular aortic stenosis.](image-url)
Congenital supravalvular aortic stenosis

Pulmonary arterioplasty was performed in these cases, in addition to SVAS repair. One of these patients underwent xenograft pericardial patch repair of short segment stenosis of the right PA. Membrane excision as an additional surgical intervention was performed for 1 patient with a subaortic discrete membrane. Two patients had a bicuspid aortic valve. Aortic regurgitation was present in 4 patients: mild in 3 cases and moderate in 1. The patients with bicuspid aortic valve had a mild degree of aortic valve stenosis. Surgical intervention was not required for aortic valve stenosis. The mean duration of aortic cross-clamp was 57.6±18.2 minutes (range: 42–80 minutes) and the mean duration of cardiopulmonary bypass was 79.7±20.5 minutes (range: 61–105 minutes).

Mortality was observed in 1 patient (10%). This patient was a 6-month-old infant with a low body weight (6 kg) and diffuse SVAS. The patient was separated from cardiopulmonary bypass with high doses of inotropic agents and died as a result of low cardiac output in the intensive care unit on the first postoperative day. In addition to the diffuse SVAS surgical repair with the Doty technique, augmentation of the ascending aorta and hemiarch with Dacron grafts was performed. The right PA was augmented from the bifurcation to the hilum with a xenograft pericardial patch. Preoperative cardiac catheterization with coronary and aortic angiography had not detected this coronary abnormality in this patient, but during the operation, a slight circumferential thickening of the left main ostium and right coronary artery was found. In this patient, there was no need for coronary surgical osteoplasty.

A postoperative complication was observed in 1 patient. There was a prolonged intensive care unit stay (3 days) and total hospital stay (14 days) due to pneumonia.

The mean peak systolic gradient seen in preoperative echocardiography was 90±25.5 mm Hg (range: 65–118 mm Hg). The mean peak systolic gradient on echocardiography performed before discharge and at the last follow-up was 20±8.4 mm Hg (range: 12–28 mm Hg; p<0.01) and 14±4.2 mm Hg (range: 10–18 mm Hg; p<0.05) respectively (Fig. 2, 3). During follow-up, it was found that aortic valvular functions were well preserved, and there was no progression observed in patients with aortic insufficiency (AI). There was regression of moderate AI to mild AI in 1 patient. During follow-up of 3 patients originally classified as having mild AI, no further AI was detected in

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**Table 1. Demographic and clinical data of the patients**

<table>
<thead>
<tr>
<th></th>
<th>n</th>
<th>Mean±SD</th>
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</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>Gender</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>Age (years)</td>
<td>4.8±3.9</td>
<td></td>
</tr>
<tr>
<td>Weight (kg)</td>
<td>17±10.1</td>
<td></td>
</tr>
<tr>
<td>Williams-Beuren syndrome</td>
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<td></td>
</tr>
<tr>
<td>Supravalvular stenosis type</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Discrete</td>
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<td></td>
</tr>
<tr>
<td>Diffuse</td>
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<td></td>
</tr>
<tr>
<td>Additional cardiovascular abnormality</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bicuspid aortic valve</td>
<td>2</td>
<td></td>
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<tr>
<td>Peripheral pulmonary stenosis</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Subaortic discrete membrane</td>
<td>1</td>
<td></td>
</tr>
</tbody>
</table>

SD: Standard deviation.

**Table 2. Operative and postoperative details and echocardiographic measurements of the patients**

<table>
<thead>
<tr>
<th></th>
<th>n</th>
<th>Mean±SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Additional surgery</td>
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<td></td>
</tr>
<tr>
<td>Pulmonary arterioplasty</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Subaortic discrete membrane resection</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Aortic X-Clamp time (min)</td>
<td>57.6±18.2</td>
<td></td>
</tr>
<tr>
<td>Total bypass time (min)</td>
<td>79.7±20.5</td>
<td></td>
</tr>
<tr>
<td>Intensive care stay (days)</td>
<td>1.5±0.4</td>
<td></td>
</tr>
<tr>
<td>Hospital stay (days)</td>
<td>6.2±1.2</td>
<td></td>
</tr>
<tr>
<td>Aortic insufficiency (preoperative/postoperative)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>3/3</td>
<td></td>
</tr>
<tr>
<td>Moderate</td>
<td>1/0</td>
<td></td>
</tr>
<tr>
<td>Balloon pulmonary angioplasty (postoperative)</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>SVAS peak gradient (mm Hg)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Preoperative</td>
<td>90±25.5</td>
<td></td>
</tr>
<tr>
<td>Postoperative before discharge</td>
<td>20±8.4</td>
<td></td>
</tr>
<tr>
<td>Last follow-up</td>
<td>14±4.2</td>
<td></td>
</tr>
<tr>
<td>Duration of follow-up (years)</td>
<td>3.7±1.2</td>
<td></td>
</tr>
</tbody>
</table>

SVAS: Supravalvular aortic stenosis; SD: Standard deviation. 

respectively. Pulmonary arterioplasty was performed in these cases, in addition to SVAS repair. One of these
commonly seen. In more serious cases, hypoplasia advancing up to aortic arch and iliac bifurcation can be observed. In our case series, diffuse hypoplasia was present in 1 patient. Luminal hypoplasia was observed throughout the aorta on preoperative catheter angiography images. Peripheral pulmonary stenosis was also recorded in this patient, with a diagnosis of WBS.

The aim in surgical repair of SVAS is to remove the left ventricular outflow tract obstruction. In the last 40 years, several surgical techniques have been defined for this purpose; however, the optimal surgical approach for SVAS still remains to be identified. The first of these was the single-patch aortoplasty technique defined by McGoon et al., but it does not provide symmetrical enlargement. Doty et al. later designed the reverse Y-shaped patch extending toward the non-coronary and right coronary sinus, which provides more geometrical enlargement. Brom et al. modified the technique by patching and enlarging all 3 sinuses during the repair. Furthermore, Myers et al. performed the repair by creating a flap in the distal and proximal part of the obstruction in the ascending aorta without using foreign material in order to preserve both the growth potential and geometry. Recently, a modified arterioplasty technique has been published in the literature as a different surgical modification. Although the qualities of the different techniques have been discussed, none has yet been uniformly judged superior.

1 patient (mild AI remained in 2 patients). At the final visit, it was determined that all of the patients were asymptomatic (New York Heart Association Class 1). Balloon pulmonary angioplasty was required in 1 case during a follow-up period of 3.7±1.2 years (range: 1.2–6.1 years) due to segmental pulmonary stenosis. The desired gradient reduction response was achieved using angioplasty: the gradient was reduced from 45 mm Hg to 18 mm Hg.

**DISCUSSION**

The disease pattern and the degree of arterial lesion vary widely, depending on the degree of elastin defect that develops according to the microdeletion damage. Elastin arteriopathy is evaluated as either discrete or diffuse. The discrete type of stenosis, which is similar to a localized ring, is seen in most cases, while diffuse tubular hypoplasia, which affects the ascending aorta, aortic arch, and its branches, is less commonly seen. In more serious cases, hypoplasia advancing up to aortic arch and iliac bifurcation can be observed. In our case series, diffuse hypoplasia was present in 1 patient. Luminal hypoplasia was observed throughout the aorta on preoperative catheter angiography images. Peripheral pulmonary stenosis was also recorded in this patient, with a diagnosis of WBS.

The aim in surgical repair of SVAS is to remove the left ventricular outflow tract obstruction. In the last 40 years, several surgical techniques have been defined for this purpose; however, the optimal surgical approach for SVAS still remains to be identified. The first of these was the single-patch aortoplasty technique defined by McGoon et al., but it does not provide symmetrical enlargement. Doty et al. later designed the reverse Y-shaped patch extending toward the non-coronary and right coronary sinus, which provides more geometrical enlargement. Brom et al. modified the technique by patching and enlarging all 3 sinuses during the repair. Furthermore, Myers et al. performed the repair by creating a flap in the distal and proximal part of the obstruction in the ascending aorta without using foreign material in order to preserve both the growth potential and geometry. Recently, a modified arterioplasty technique has been published in the literature as a different surgical modification. Although the qualities of the different techniques have been discussed, none has yet been uniformly judged superior.
In previous studies, surgical interventions during infancy, diffuse SVAS etiology, bicuspid valvular structure, coronary obstruction, and the presence of WBS syndrome have been found to be associated with early mortality. Early mortality has been reported between 2% and 11% in the literature. In our series, early mortality was observed in 1 patient (10%) who had 3 risk factors: surgery at an early age, pulmonary stenosis, and diffuse SVAS. The patient died following sudden cardiac arrest at the postoperative eighth hour.

Although the early results seem good in some of the larger series, the rates of late mortality and reoperation are significant. Preoperative patient characteristics had a greater role in outcomes than techniques. Reintervention after SVAS surgery was associated with the presence of left ventricular outflow tract obstruction at follow-up. These cases were also associated with the diffuse form of SVAS with aortic arch stenosis (hypoplasia or coarctation), age <12 months, and an aortic valve pathology that often required a late reintervention (either surgical or interventional). Reinterventions typically occurred over a decade after the primary surgery. These mid-term follow-up findings confirm that the Doty technique has a low reoperation rate and a low incidence of aortic regurgitation. Similar observations have also been made in other studies. Elastin arteriopathy may also involve other large arteries, such as the PAs, either at the central, branching, segmental, or subsegmental level.

Although PA stenosis is frequently diagnosed preoperatively in cases of SVAS, an operation on the PA tree is not always required since the peripheral vessels are most commonly affected. Currently, no consensus exists about the best approach for these patients.

Some advocate for catheter intervention alone and some prefer surgery, while others combine these 2 methods. Whatever the technique used, reintervention is the most common cause of long-term complications, and long-term follow-up is required. The associated presence of preoperative PA stenosis is a risk factor for late death and catheter intervention. Thus, reinforcing the importance of close follow-up in patients with PA stenosis is essential. In our series, 2 patients with PA stenosis required an associated PA procedure during SVAS repair. PA patch augmentation was performed in a concomitant procedure on the right PA. Balloon pulmonary angioplasty was required in 1 case within a 3-year follow-up period due to segmental pulmonary stenosis. The desired response in gradient was achieved in this WBS patient following balloon pulmonary angioplasty.

Coronary obstruction is often a concern in cases with this lesion, as well as the surgical management. The incidence of a preoperative diagnosis of coronary anomalies among patients with SVAS ranges from 36% to 70%. Deo et al. reported a preoperative rate of 18% coronary ostial obstruction that was not treated in surgery. Coronary stenosis and ischemia with various etiologies (isolation of the coronary ostia from fused cusps, arteriosclerosis resulting from intracoronary hypertension, and discrete or diffuse ostial stenosis) may be associated with SVAS. Stamm et al. reported an incidence of 23% marked dilation or tortuosity of coronary arteries and 45% coronary arterial orifice stenosis. In addition, severe left ventricular hypertrophy further compounds coronary subendocardial malperfusion, causing myocardial ischemia. The presence of coronary anomalies must be assessed to provide effective surgical repair of SVAS, both preoperative evaluation using imaging modalities and peroperative inspection of the coronary ostium and anatomy. In 1 of our cases, despite a good clinical assessment and further imaging, slight circumferential thickening of the left main ostium and right coronary artery was found peroperatively. Yet, there was no need for coronary surgical ostioplasty due to the absence of signs or symptoms associated with coronary artery obstruction.

As the disease is rare, the number of patients in our series was limited and the data were not sufficient for an analysis of risk factors. Mortality was observed in 1 patient. Furthermore, the retrospective nature of the study is another limitation.

In conclusion, although satisfactory results have been achieved with the Doty aortoplasty technique,
the number of patients with the accepted risk factors and the length of follow-up was limited in our series. Consequently, we may not yet have seen all eventual late mortality and reoperation with the 3-patch repair. Longer follow-up is needed. The Doty technique has been preferred in our clinic for yielding good midterm results without increasing morbidity and mortality, no progression in the gradient, no impairment in coaptation, and no chest pain, as well as being a technique that can be performed easily.

**Peer-review:** Externally peer-reviewed.

**Conflict-of-interest:** None.


**REFERENCES**


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Keywords: Doty technique; supravalvular aortic stenosis; Williams-Beuren syndrome.

Anahtar sözcükler: Doty tekniği; supravalvüler aortik stenozu; Williams-Beuren sendromu.