Requirement for repetitive surgical approaches at supravalvular aortic stenosis

Supravalvular aortic stenosis, which is a rare congenital cardiac anomaly, is associated with several lesions and has a progressive nature. Herein, we report a five-year-old girl with bicuspid aorta who underwent initial Doty operation at the age of nine months. A combined redo Doty operation and an aortic valve commissurotomy were performed two years later. Due to the rapidly progressing aortic regurgitation and both valvular and supravalvular gradient, a repeated surgery was required at the age of five years and an aortic homograft was successfully inserted with an annulus enlargement and the patient was discharged uneventfully. In conclusion, although Doty repair yields satisfactory results in most patients, certain cases with identified risk factors may require reoperations due to the progressive nature of the disease. Therefore, these patients should be kept under a close follow-up lifelong.

Keywords: Bicuspid aortic valve, doty repair, supravalvular aortic stenosis.

Supravalvular aortic stenosis (SVAS) is caused by a deletion of the elastin gene that ends up with narrowing of large elastic arteries, mostly the aorta. As it may present a feature of Williams-Beuren syndrome, it contains an autosomal dominant trait. In addition, several articles have been published reporting associated lesions such as coronary artery stenosis, bicuspid aortic valve, subvalvular aortic stenosis, and pulmonary stenosis in the literature.

To date, several surgical methods have been described and, currently, two sinus augmentation procedures with an inverted Y-patch (also known as Doty repair) and the three-patch technique (also known as Brom repair) have been widely utilized for the surgical correction of SVAS. Although large series have shown satisfactory and event-free late term results after surgical treatment due to the progressive course of SVAS, reoperations are not uncommon.

Herein, we present a case of SVAS who underwent Doty operation twice and, then, required a homograft replacement over a two-year interval.

CASE REPORT

A nine-month-old girl, who was under follow-up with the diagnosis of bicuspid aorta and SVAS since the neonatal period, was admitted to our institution with an increased gradient and need for a surgical treatment.
Initial transthoracic echocardiography (TTE) revealed 70 mmHg supravalvular gradient with bicuspid aorta; however, the aortic annulus was not narrowed and there was no valvular or subvalvular gradient. Computed tomography (CT) confirmed the diagnosis, and the heart and great vessels were assessed for associated lesions. Genetic tests excluded Williams-Beuren or other syndromes. Surgical correction was decided by the Pediatric Heart Council. During surgery, the narrowed segment relieved via a Dacron patch fashioned as inverted Y. There was no residual gradient at the time of discharge. At one year of follow-up, a significant gradient was measured via TTE and CT angiography was repeated. The CT images revealed supravalvular and valvular stenosis. In addition, TTE confirmed the findings and measured 85 mmHg supravalvular and 35 mmHg valvular gradient. Based on these findings, reoperation was concluded. During surgery, the supravalvular stenosis was enlarged via a Dacron graft as in the previous operation, and the aortic valve was evaluated. The aortic commissurotomy was applied, considering that only 10-mm Hegar bougie could pass through the aortic valve, although the appropriate Z score was 12.3. Following commissurotomy, the 12-mm-sized bougie was able to fit the annulus and, therefore, surgery was completed. The perioperative transesophageal echocardiography (TEE) revealed a remaining negligible supravalvular gradient without concurrent valvular gradient. On the other hand, trivial aortic regurgitation (AR) was detected. On Day 6 of hospitalization, the patient was discharged and scheduled for a closer follow-up. The onset of the valvular gradient and insufficiency began and developed in subsequent visits. The final TTE examination revealed 80 mmHg gradient with severe AR and the annular diameter was measured as 13 mm. Computed tomography angiography was repeated and the course of the coronary vessels were evaluated (Figure 1). Once more, these findings warranted the surgical repair. The adhesions complicated the release of the coronary buttons; therefore, a 15-mm-sized aortic homograft was inserted with an annular posterior enlargement procedure. Meanwhile, the aortic root replacement was avoided (Figure 2). Following excision of the previously inserted Dacron patch, SVAS was relieved.
by using a residual homograft tissue (Figure 3). The entire process was completed with a total of 188 min of cross-clamping and 201 min of cardiopulmonary bypass time. Postoperative TTE revealed normal valve function with no residual gradient (Figure 4). Then, the patient was discharged uneventfully on postoperative Day 6. Control TTE revealed similar findings at six months and the patient remained asymptomatic during follow-up. A written informed consent was obtained from each parent.

**DISCUSSION**

In this article, we report an interesting case who required multiple surgical approaches due to SVAS. We preferred the Doty technique as usual in our institution and only the present case required reintervention, since the last decade, among 15 patients operated by the same surgical team in our clinic. Although the Doty technique provides a non-asymmetrical correction compared to the Brom technique, it is not yet clear that the latter yields superior outcomes.\(^{[3-5,7]}\) Therefore, we routinely utilize the Doty technique to increase overall suture line to avoid bleeding. In addition, Mongé et al.\(^{[3]}\) reported that cross-clamp time was longer for the three-patch technique. Besides, Roemers et al.\(^{[4]}\) published their 52-year results for SVAS. They observed that freedom from reoperation rates were 100% and 88% for 10 and 20 years, respectively. Nonetheless, these results did not differ from the Brom group. In particular, the authors highlighted that, in patients with a bicuspid aortic valve, they did not prefer a symmetrical reconstruction and enlarged the sinuses with one or two patches, depending on the most ideal appearance.

More interestingly, in another study, despite satisfactory surgical results, the survival in SVAS cohort was found to be significantly lower, compared to the general population.\(^{[5]}\) Considering the progressive nature and underlying genetic disorder, reinterventions are not infrequent. In the largest, multicenter, retrospective study including 301 patients using data from the European Congenital Heart Surgeons Association was conducted by Padalino et al.\(^{[5]}\) They found 12.6% (n=30) reoperation rate. Eight of them were redo SVAS and seven were aortic valve replacement. Two of the redo SVAS cases were previously operated with the Doty technique and SVAS was relieved by redo patch plasty similarly to our case. However, repair in infancy was determined as a risk factor for reintervention and long-term survival in this study, as previously reported.\(^{[3-6]}\)

Liu et al.\(^{[6]}\) reported a case with bicuspid aorta which was initially treated with the Doty technique, followed by the McGoon repair and aortic valvuloplasty 2.5 years later. They observed residual aortic stenosis in 14 of 90 patients and found that male gender, bicuspid aorta, infancy, and preoperative peak gradient of greater than 90 mmHg to be risk factors. Similarly, our case had bicuspid aorta and was initially operated when she was nine months old with a high gradient, suggesting prior findings.

Aortic regurgitation may develop following surgical treatment over time. Furthermore, the most common reason for reoperation was identified as aortic valve replacement.\(^{[7]}\) Some authors emphasized that non-symmetrical techniques remained a theoretically improved risk as they could lead to aortic regurgitation and constructed an inadequate adaption to new aortic root geometry.\(^{[6]}\) On the contrary, Kari et al.\(^{[8]}\) reported a case of AR progression from mild-to-severe within three months following commissurotomy and that valvular complications might manifest in the late term. Therefore, the patient was ultimately treated with a homograft replacement. In the aforementioned study, increased AR rates in the late postoperative period of SVAS were also evaluated, compared to perioperative values. In another study, Fricke et al.\(^{[7]}\) presented a case who underwent aortic valve replacement with a 19-mm mechanical valve seven years after the initial Doty procedure at the age of four. Then, the Konno procedure was performed as the third operation at the age of 20 years. In the study of Padalino et al.,\(^{[5]}\) the median interval from the first operation for aortic valve replacement was found to be 5.7 (range, 0.8 to 22.3) years, and one of seven cases was treated with aortic homograft in this serial. In a meta-analysis, Etnel et al.\(^{[9]}\)
compared the Ross procedure, mechanical valve and homograft replacement in the aortic valve position. They found that the Ross procedure provided better outcomes in terms of low early and late mortality rates. On the other hand, homograft replacement was found to be associated with a significantly higher rate of reoperations. However, we performed homograft replacement due to the adhesions causing unease to relief the coronary buttons. Due to the absence of the appropriate mechanical valve size, the parents of the patient were unwilling to the insertion of mechanical valve and long-term warfarin treatment, and this option was abandoned.

In addition, an emerging aortic valve replacement method using the glutaraldehyde-treated autologous pericardium (Ozaki procedure) has been introduced in the last decade and has become worldwide. Although some satisfactory results have been obtained in children with this technique, further studies are still needed.

In conclusion, although the Doty repair can be achieved with satisfactory results and event-free survival in most patients to other well-described techniques, occasional cases with identified risk factors may require reoperations due to the progressive nature of the disease. Considering these findings and evidences regarding significantly low survival rates, compared to the general population, patients should be kept under a close follow-up lifelong.

**Declaration of conflicting interests**

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

**Funding**

The authors received no financial support for the research and/or authorship of this article.

**REFERENCES**