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Case Report

A New Method for Supravalvular Aortic Stenosis Repair

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Abstract

Conventional supravalvular aortic stenosis repair methods are associated with risks of aortic regurgitation, restenosis, and bleeding at the anastomosis site. Our method involves peeling the inner thickened aortic wall from below the innominate artery to above the coronary ostium, followed by end-to-end anastomosis. Our simple method involves less bleeding and fewer geometrical changes in the sinuses of Valsalva.

Keywords: Supravalvular Aortic Stenosis; Congenital Heart Surgery; New Method; Restenosis; Aortic Regurgitation

Case Report

A 10-year-old female patient weighing 47.9 kg underwent a routine clinical workup. A grade 2 systolic murmur was noted at the right sternal border during physical examination. The heart did not appear enlarged on chest radiography. Electrocardiography revealed a normal sinus rhythm with left ventricular hypertrophy. Echocardiography revealed supravalvular stenosis and a normal ejection fraction.

To clearly visualize the vessel structure and the surrounding anatomy, we performed three-dimensional computed tomography, which revealed supravalvular aortic stenosis. Cardiac catheter examination revealed a pressure gradient of 60 mmHg.

Surgical repair was performed through a median sternotomy. Cardiopulmonary bypass was established after heparinization. The incision line was marked to the ascending aorta to avoid injury to the coronary ostium, and the patient was operated under hypothermic chemical cardiac arrest. The ascending aorta had a thickened wall and a narrow lumen. We peeled out the inner portion of the thickened aortic wall just below the innominate artery at the distal side and just above the coronary ostium on the proximal side (Figure 1). After the procedure, the aortic wall was well dilated (Figure 2), and end-to-end anastomosis was performed. After rewarming, the patient was easily weaned from the cardiopulmonary bypass without arrhythmias, and the postoperative hemodynamic status of the patient was stable. On postoperative day 3, echocardiography revealed non-obstructed blood flow through the anastomosis site and good cardiac function. The patient was discharged from our hospital on postoperative day 10 in good condition. Two years have passed since the operation, and restenosis of the ascending aorta has not yet been observed. We plan to continue annual medical follow-up using echocardiography or computed tomography.

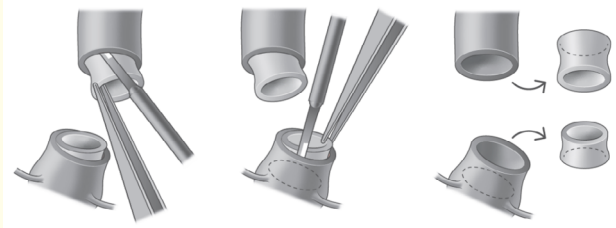

Figure 1: We peeled out the inner portion of the thickened aortic wall just below the innominate artery at the distal side and just above the coronary ostium at the proximal side.

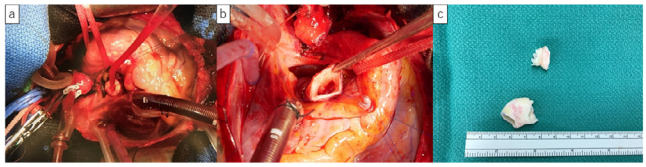

Figure 2: (a) Before the resection, ascending aorta wall was well thickened. (b) After the procedure, the aortic wall was well dilated. (c) Resected thickened aortic wall tissue.

Discussion

McGoon., *et al*. first reported supravalvular aortic stenosis repair using the single-patch augmentation technique in 1961 [1]. Since then, multiple surgical techniques have been developed and have evolved. These techniques for surgical repair of supravalvular aortic stenosis include the McGoon single-patch repair, Doty pantaloon-patch repair, Brom 3-patch technique, and Myers' autologous sliding aortoplasty technique [1-8]. Although the operating time is not significantly different among the various surgical approaches, the Doty and Brom aortoplasties restore normal hemodynamics and reduce the need for reoperation compared with the classic 1-patch technique. However, the overall freedom from reoperation for left ventricular outlet tract obstruction at 10 years was 70.3% [3,4]. These conventional surgical treatments consist of making an incision between the commissures of the aortic valve and patch enlargement for one to three commissures. Larger patches cover a greater diameter but carry the risk of reducing the coaptation area of the aortic valve by lateral displacement of the commissural attachments of the valve, possibly resulting in aortic regurgitation. There also exists a risk of bleeding because the surgical procedure is complicated and involves many suture lines. Late mortality may occur due to left or right coronary artery ostium stenosis as the incision line is close to the coronary ostium in patients who receive a larger patch. Our new surgical repair approach for supravalvular aortic stenosis is straightforward. The suture line is very simple and does not affect the coronary ostium or valve geometry.

In patients with supra-aortic stenosis, the wall of the ascending aortic route is very thick and edematous. Consequently, this lesion is the very problem of this disease. Recurrent aortic stenosis by this lesion persists in other surgical treatments. Our method involves resecting this tissue so that the risk of a recurrent event is reduced.

However, dilation of the ascending aorta remains a key concern. Thinning of the aortic wall raises another concern about the risk of developing an aortic aneurysm in the future. However, prior studies suggest that only abnormal tunica media is resected and that the main middle and adventitia membrane remains [9]. The middle elastic membrane is the core structure of the vascular wall. Hence, we do not have to be concerned about risks such as future dilation of the aortic wall. Nevertheless, long-term follow would be required to confirm this assumption.

Conclusion

We propose our novel surgical method as the new option to treat supravalvular aortic stenosis to reduce the risks of bleeding from the anastomosis site, aortic regurgitation, and recurrent aortic stenosis.

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Conflicts of Interest

None.

Ethical Standards

We obtained informed consent verbally and documented it in the medical record. We also obtained approval from the ethics committee of Juntendo University.

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