Cardiovascular System
ISSN 2052-4358 | Volume 4 | Article 1

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Case report Open Access

New four patch repair [Modified Brom's] technique for supravalvular aortic stenosis

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Abstract

The objective of this paper is to describe the novel four-patch technique [modified Brom's] for repair of supravalvular aortic stenosis (SVAS). After transection of the aortaat the sinotubular junction, three longitudinal incisions are made into thethree sinuses. The aortic root geometry was restored by placement ofthree separate patches of autologous pericardium in the opened sinuses. One additional diamond shaped pericardial patch placed anteriorly between proximal and distal aorta. This modified four patch technique provides a complete and symmetric restoration of theaortic anatomy with insignificant gradient and good short-term and potentially good long-term results.

Keywords: Supravalvular aortic stenosis, brom three patch technique, pericardial patch

Introduction

Supravalvar aortic stenosis (SVAS) is a fixed form of congenital left ventricular outflow tract (LVOT) obstruction that occurs as a localized or diffuse narrowing of the ascending aorta beyond the superior margin of the sinuses of Valsalva [1]. In cases with Williams syndrome, SVAS can be associated with peripheral pulmonary artery stenosis [2]. SVAS is the result of a spontaneous or inherited microdeletion in theelastin gene, localized at chromosome 7. The sporadic form of SVAS is the most common (>50%) presentation. The anatomic diagnosis of SVAS can reliably be made from 2-dimensional (2D) echocardiography that uses multiple views, including parasternal, apical long-axis, and suprasternal. Cardiac catheterization or MRI may be indicated to evaluate the coronary artery or aortic arch anatomy. Surgery is the primary treatment for SVAS.

For surgical correction of the SAS it is reported the use of four techniques:

1. Enlargement using simple patch in the non-coronary sinus of Valsalva, McGoon et al., technique [3].
2. Enlargement using Y bifurcated patch in right noncoronary and coronary sinuses of Valsalva, Doty technique [4].
3. Enlargement of the three sinuses of Valsalva with patch, Brom&Khonsari technique.
4. Aortoplasty with enlargement of the three sinuses of the distal aorta without use of prosthetic material, Sousa et al., technique.

In this report we present the case of a young female patient of age 14 years diagnosed with supravalvular aortic stenosis, who had undergone reconstruction of the sinotubular junction, associated with the enlargement of three sinuses of Valsalva using new four patch repair technique using autologous pericardium with postopt uneventful course and insignificant gradient.

Case presentation

14-year-old female, referred to the cardiologist for progressive limitation to physical effort and tachycardia. The cardiovascular examination revealed a hyperdynamic pulse at apex in the fifth intercostal space at anterior axillary line and low volume radial pulses. On auscultation, the heart sounds were rhythmic, with paradoxical second sound, and pansystolic murmur predominantly aortic area radiating to left sternal border, right and left supraclavicular region and back. Electrocardiogram showed sinus rhythm, axis at 80°, and overloaded left ventricle. Echocardiographic examination revealed supravalvar aortic stenosis. On angiography, the images were compatible with severe supravalvular aortic stenosis (**Figure 1**) with manometric measurements in the aorta of 100 mmHg and in the left ventricle of 150 mmHg.

Pericardial patch harvested. **Figure 2** shows the anatomical aspect of supravalvular aortic stenosis before surgical correction. The aortic valve was normal with adequate cooaptation. In

doi: 10.7243/2052-4358-4-1

the case reported in this article, we used the new four patch technique modification of Brom's technique, which consists of large dissection of the rootup to the aortic arch, transverse section of the ascending aorta just above the stenotic area, performing three sections: the first toward the non-coronary sinus and the second toward the bottom of the right coronary sinus, atleft of the ostium of the right coronary artery and the third section toward the left coronary sinus, at right of the left coronary artery ostium. In all these three sections three triangular pericardial patch sutured.

In the distal ascending aorta, longitudinal incision was made anteriorly and a diamond shaped pericardial patch sutured in that gap created. After suturing the four patches in place, an end-to-end aortic anastomosis was made connecting the reconstructed aortic root to the ascending aorta (**Figure 3**).

Postoperative transesophageal echocardiographic assessment was performed. A complete evaluation of the aortic valve, left ventricle outflow tract gradients and ventricle function was useful to evaluate the final result of the aortic restoration. The

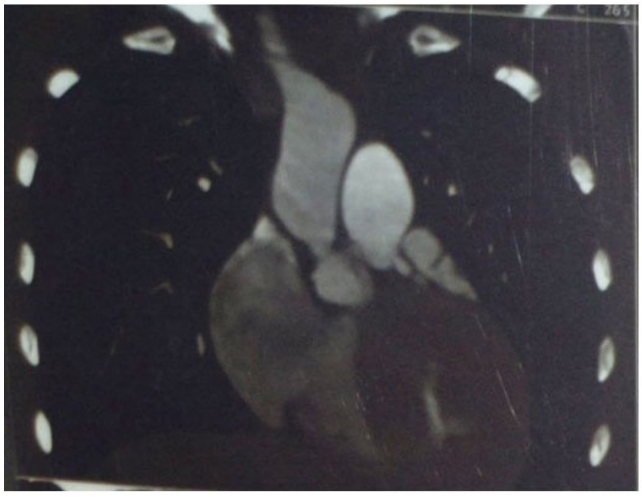

Figure 1. CT-aortogram showing supravalvar aortic stenosis.

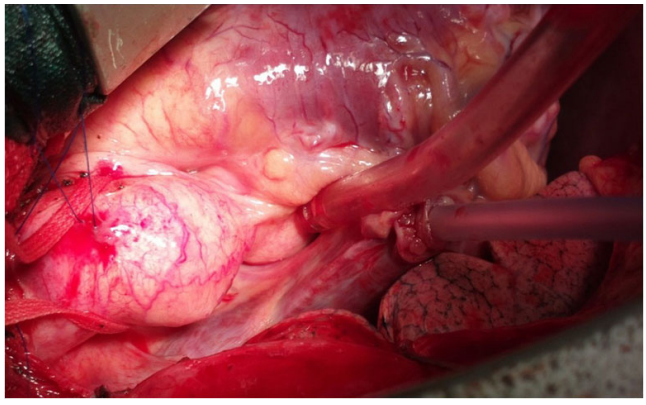

Figure 2. Supravalvar aortic stenosis.

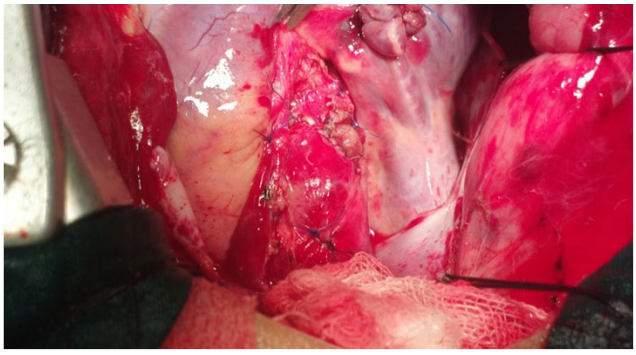

Figure 3. Four patch repair [Modified brom's] of supravalvar aortic stenosis.

postoperative follow-up was uneventful, with a maximum systolic gradient of 18 mmHg and mean of 8 mmHg, good mobility of valves without reflux (transthoracic echocardiogram). The patient was discharged from hospital on the 11th day.

After six month of follow-up, a patient was in functional class I, without cardiovascular symptoms. During this period the echocardiogram showed tricuspid aortic valve, slightly thickened with minimum reflux under color-Doppler, generating peak systolic valve gradient of 10 mmHg and supravalvular of 15 mmHg. The diameter of the aorticannulus measured 20 mm and 19mm for the supravalvular region (**Table 1**).

Table 1. Comparision of patient symptoms and Doppler data before and after the operation.

	Before operation	After operation
Breathlessness on exertion	Class 3	Class 1
Supravalvar Aortic gradient	Peak 100 mm mean 70 mm of hg	Peak 15 mm mean 8 mm of hg

Discussion

Supravalvular aortic stenosis was first described in 1842 by Chevers as anisolated malformation affecting only the narrowing of the aortic lumen. It was in 1958, when Denie and Verheugt described the supravalvular aorticstenosis with a more morphologic and detailed description, focusing the narrow lesion at the aortic sinotubular junction. Complications of SVAS also include progressive coronary osteal stenosis, infective endocarditis, and sudden death [**5**,**6**].

The original simple patch enlargement of sinotubular junction at the non-coronary sinus of Valsalva, proposed in 1961 by McGoon and associates, was the first surgical technique described to widen the supravalvular stenosis. This procedure became the standard surgical procedure until the description of more extended aortoplasties. Multiple surgical options of treatment of supravalvular aortic stenosis have been suggested. Doty, Polansky and associates, in 1977, described an extended aortoplasty that involved two sinuses instead of one, using

a bifurcated patch(Y-shaped Dacron patch). In this case, an inverted Y-shape incision is made into the non-coronary sinus and the right coronary sinus and the bifurcated patch is placed. This technique appeared as a more symmetric enlargement of the aortic root, but the narrowing above the left coronary ostium remains unsolved by this extended aortoplasty. Some years later, Brom (1988) described a new surgical approach that enlarged all three sinuses of Valsalva . In this procedure, the aorta is transected just above the stenosis, and three longitudinal incisions are made in each sinus of Valsalva and three pericardial patches are inserted.

The three-patch technique for repair of supravalvular aortic stenosis designed by Brom, provided in the past a new concept of restoration of the aortic tract that nowadays continues to be the ideal model to repair the supravalvular aortic stenosis. The advantages of this technique in contrast to other techniques reported in literature are multiple. It provides a more symmetric reconstruction and less distortion of the aortic root and the ascending aorta [**5**]. Autologous pericardium has never resulted in aneurysm formation and appears to behave as normal thickness vascular tissue. However, we believe that Brom's three-patch technique may be technically easier while the individual patches can be designed in such a way that aortic root restoration will be optimal. Care should be taken not to widen the patches too much as this may lead to loss of aortic valve coaptation and valve leakage.

The presented case demonstrates several aspects. Due to the wide margin of fibrosis and the thickness of the aortic wall, we has chosen the modified Brom's technique ie four patch technique, thus avoiding distortion of the sinuses with full valve competence.

Conclusion

We consider this new four patch technique for SVAS repair to be safe and it produced effective anatomic restoration with no gradient along with good short-term and potentially good long-term results. Brom's three patch technique was modified into four patch technique for enlargement of the more distal ascending aorta with good aortic valve coaptation. It is important to emphasize not to use artificial prosthetic material, to ensure development and growth of the sinotubular junction of aorta.

Competing interests

The authors declare that they have no competing interests.

Author contributions

Dr. Suraj Wasudeo Nagre-analysing and interpreting the patient data also literature search, and image editing. Dr Suhas Bendre and Dr. Suraj Wasudeo Nagre-done surgery.

Acknowledgment

Grant Medical College and J J Group of Hospitals, Mumbai, Maharashtra.

Publication history

Editor: Xupei Huang, Florida Atlantic University, USA.
Received: 15 December 2015 Revised: 23 January 2016
Accepted: 10 February 2016 Published: 18 February 2016

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Citation:
Nagre S.W and Bendre S. **New four patch repair [Modified Brom's] technique for supravalvular aortic stenosis**. *Cardio Vasc Syst*. 2016; **4**:1.
http://dx.doi.org/10.7243/2052-4358-4-1