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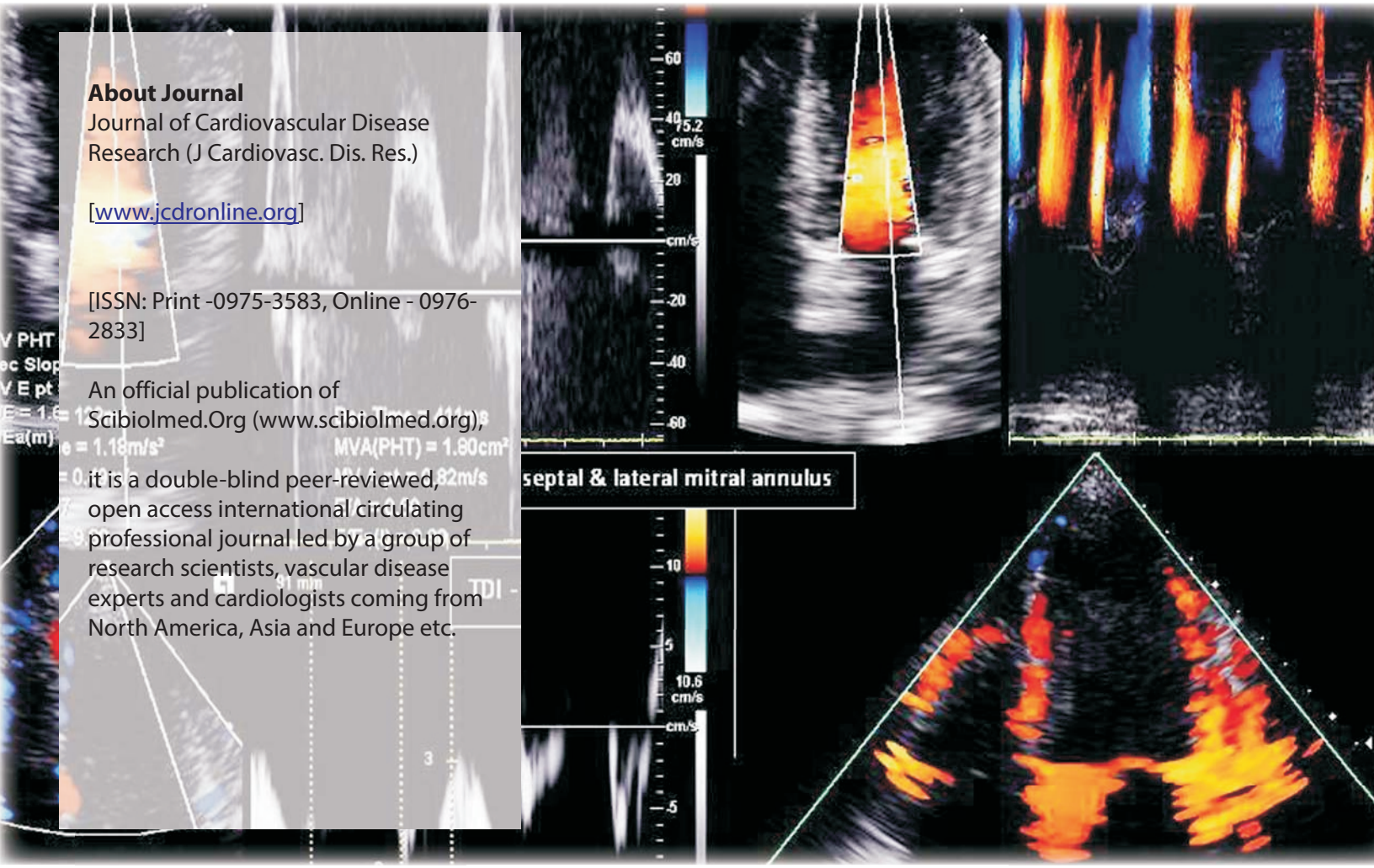

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Case Report

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 for repair of supravalvular aortic stenosis (SVAS). Supravalvular aortic stenosis is a rare malformation as a result of an abnormal thickening of the aortic wall. SVAS may present in two forms: a localized form (affecting only the aortic sinotubular junction) and a diffuse form, where the aortic arch and its side branches are also affected. We present the novel four-patch technique as modification of Brom three patch technique. After transection of the aorta at the sinotubular junction, three longitudinal incisions are made into the three sinuses. The aortic root geometry is then restored by placement of three 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 the aortic anatomy with insignificant gradient and good short-term and potentially good long-term results.

Key words: Supravalvular aortic stenosis, Brom three patch technique, Pericardial patch.

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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] It accounts for less than 7% of all fixed forms of congenital LVOT obstructive lesions. Histology reveals an abnormal thickening of the aortic media, as well as anintimal hyperplasia of the aortic wall. It is presented as a characteristic lesion in patients affected by Williams-Beuren syndrome, but SVAS can also be found in non-syndromic familiar forms or sporadic cases. 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 and Khonsari technique;[5]

4. Aortoplasty with enlargement of the three sinuses of the distal aorta without use of prosthetic material, Sousa *et al*. Technique.[6]

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 REPORT

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 80º, and overloaded left ventricle in the form of deep q waves and increased qrs amplitude in V5 and V6. 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.

The surgery was performed with cardiopulmonary bypass, cannulation of aorta and vena cava using anterograde cold blood cardioplegia (4ºC) for myocardial protection, with the patient at 31ºC. Intraopt TEE shown supravalvar aortic stenosis (Figure 2). Pericardial patch harvested (Figure 3). Figure 4 shows the anatomical aspect of supravalvular aortic stenosis before surgical correction. Transverse incision was performed in the aortajust above the stenosis, which showed: significant stenosis of the sinotubular junction, extensive fibrosis of the aortic wall. The aortic valve was normal with adequate cooaptation.

In the case reported in this article, we used the new four patch technique modification of Brom's technique, which consists of large dissection of

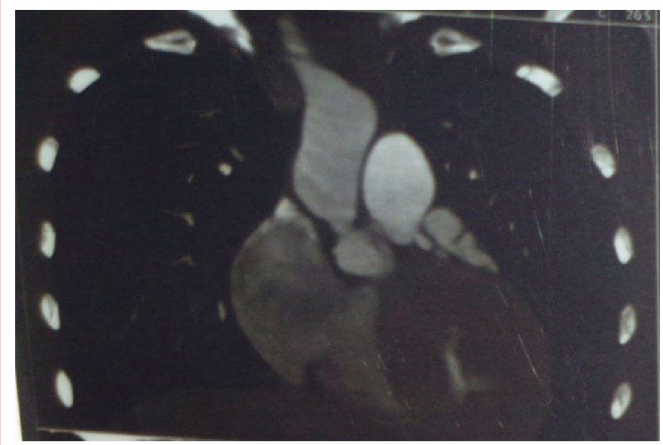

Figure 1: CT Aortogram Showing Supravalvar Aortic Stenosis

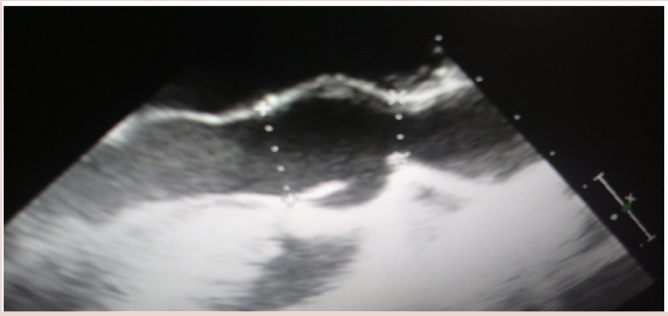

Figure 2: Transesophageal Echocardiography Showing Supravalvar Aoric Stenosis

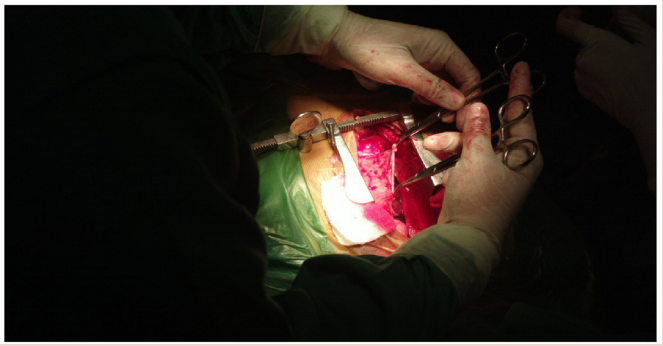

Figure 3: Pericardial Patch Harvesting

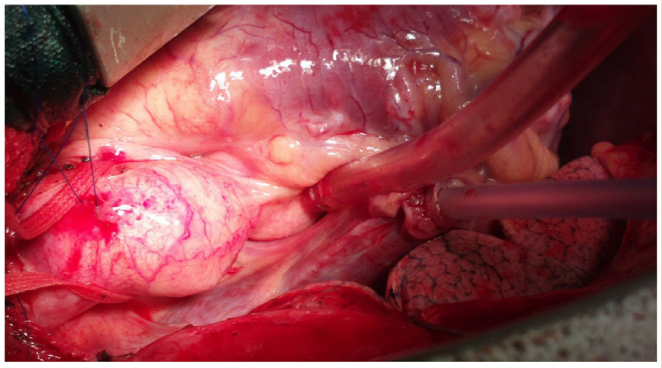

Figure 4: Supravalvar Aortic Stenosis

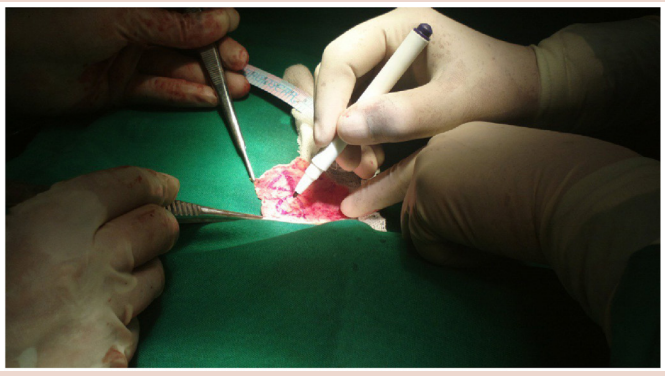

Figure 5: Marking of Pericardial Patch and Cutting

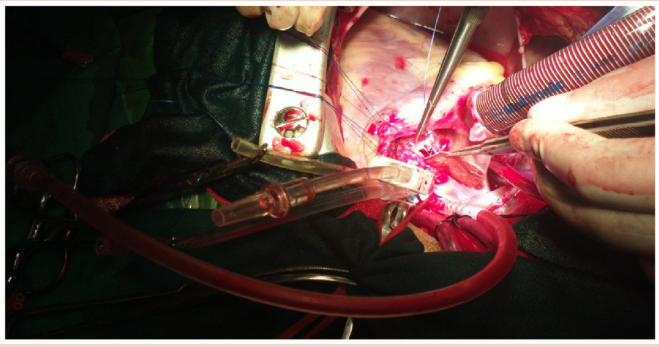

Figure 6: Suturing the Traingular Pericardial Patch in the Sinus Inscision

the root up 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, at left 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 (Figure 5 and 6).

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 7). Line diagram shows how the four pericardial patches are sutured to widen supravalvar aortic area (Figure 8).

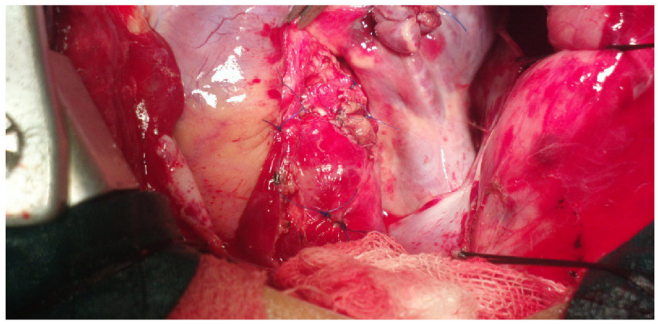

Figure 7: Four Patch Repair [Modified Brom's] of Supravalvar Aortic Stenosis

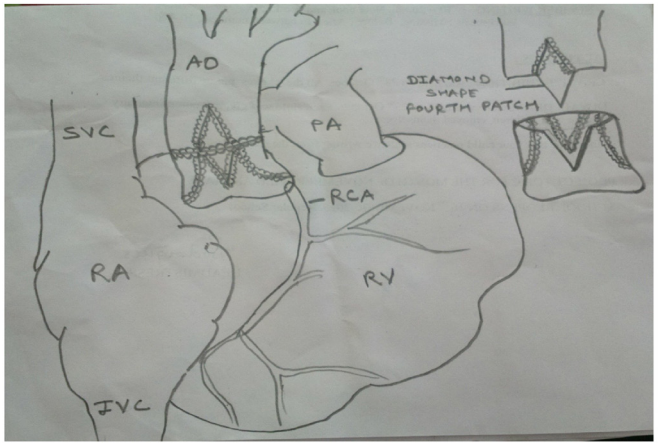

Figure 8: Four Patch Repair [Modified Brom's] of Supravalvar Aortic Stenosis-Line Diagram

We peformed the rewarming of the patientat 37°C, removal of air from the left cavities and aorta, declamping of the aorta, heart rate return to normal, disconnection of cardiopulmonary bypass (CPB), decannulation, protamine administration, review of hemostasis. Postoperative transesophageal echocardiographic assessment was performed. A complete evaluation of the aortic valve, left ventricle outflow tractgradients and ventricle function was useful to evaluate the final result of the aortic restoration. The 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 aortic annulus measured 20 mm and 19 mm for the supravalvular region.

DISCUSSION

Supravalvular aortic stenosis was first described in 1842 by Chevers as an isolated malformation affecting only the narrowing of the aortic lumen. It was in 1958, when Denie and Verheugt described the supravalvular aortic stenosis with a more morphologic and detailed description, focusing the narrow lesion at the aortic sinotubular junction. One year later, Morrow and colleagues reported the presence of aortic valve lesions associated to the supravalvular aortic stenosis. Complications of SVAS also include progressive coronary osteal stenosis, infective endocarditis, and sudden death.[7,8]

Several surgical techniques have been described since the first supravalvular aortic stenosis description appeared. 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 symmetricen largement 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 providesa more symmetric reconstruction and less distortion of the aortic root and the ascending aorta.[7] Autologous pericardium has never resulted in aneurysm formation and appears to behave as normal thickness vascular tissue. Only the three-patch technique or the sliding plasty as described by Myers, allow for effective restoration of the left coronary sinus.[7] The Myers technique has the advantage of not using any patch material. 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 patchestoo much as this may lead to loss of aortic valve coaptation and valve leakage. Different materials have been used in SVAS repair. We feel more comfortable with the use of pericardium alone, in terms of achieving better postoperative gradients.

The presented case demonstrates several aspects. Due to the wide margin of fibrosis and the thickness of the aortic wall, it was performed its resection, respecting the mobility of the leaflets. Therefore, for the treatment of significant stenosis it was 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 moredistal ascending aorta. It is important to emphasize not to use artificial prosthetic material, to ensure development and growth of the sinotubular junction of aorta.

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Grant Medical College, Mumbai, Maharashtra.

CONFLICT OF INTEREST

No potential conflict of interest to this article.

ABBREVIATION USED

SVAS: Supravalvar aortic stenosis

LVOT: Left ventricular outflow tract

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