

Surgical Repair of Supravalvular Aortic Stenosis in Adults: Two Cases - Two Types

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Abstract

Isolated supravalvular aortic stenosis (SVAS) in adults is a rare form of left ventricular outflow tract obstruction. We report two surgical cases of isolated SVAS which illustrate the diversity and the complexity of this congenital aortic disease: one focal form in a young adult man treated with an extended aortoplasty, and one tubular and calcified in a middle aged woman treated with a 2-patch technique and a replacement of the ascending aorta.

Keywords: Supravalvular Aortic Stenosis; Aortic Root Repair; Congenital Heart Surgery

Introduction

Supravalvular aortic stenosis (SVAS) is an uncommon but well-characterized congenital narrowing of the ascending aorta above the level of the sinotubular junction and the attachments of the valve commissures [1,2]. Most of the reported surgical series have been small and surgery at the adult age is exceptional [3,4]. We report two recent surgical cases of isolated SVAS in adults, which illustrate the diversity and the complexity of this congenital aortic disease.

Case Report

Case 1 Presentation

A 18-year old male with regular follow-up for an asymptomatic and well documented SVAS was referred to surgery because of the occurrence of dyspnea on exertion and palpitations. Angio CT scan (Figure 1) and TEE (Figure 2) confirmed the diagnosis of a focal SVAS with a mean gradient of 40 mm Hg; the aortic valve was normal with a mild central regurgitation. There was no associated Williams syndrome or other heart abnormality. Surgical repair was done through median sternotomy using crystalloid cardioplegia arrest and moderate hypothermic cardiopulmonary bypass. After the opening of the ascending aorta with a longitudinal incision extended into the noncoronary sinus, the fibrous ridge was removed and a single diamond-shaped bovine pericardial patch (Edwards Lifesciences™) was inserted for aortic root reconstruction. The postoperative outcome was event-free. Cardiac echocardiography confirmed a good hemodynamic result with a residual gradient less than 10 mmHg. The patient has been asymptomatic till the last follow-up.

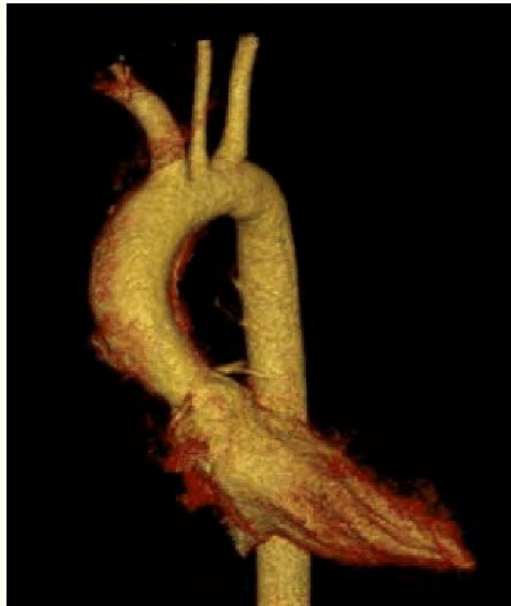


Figure 1: Case 1. Preoperative Angio CT scan. 3D reconstruction of the aorta.

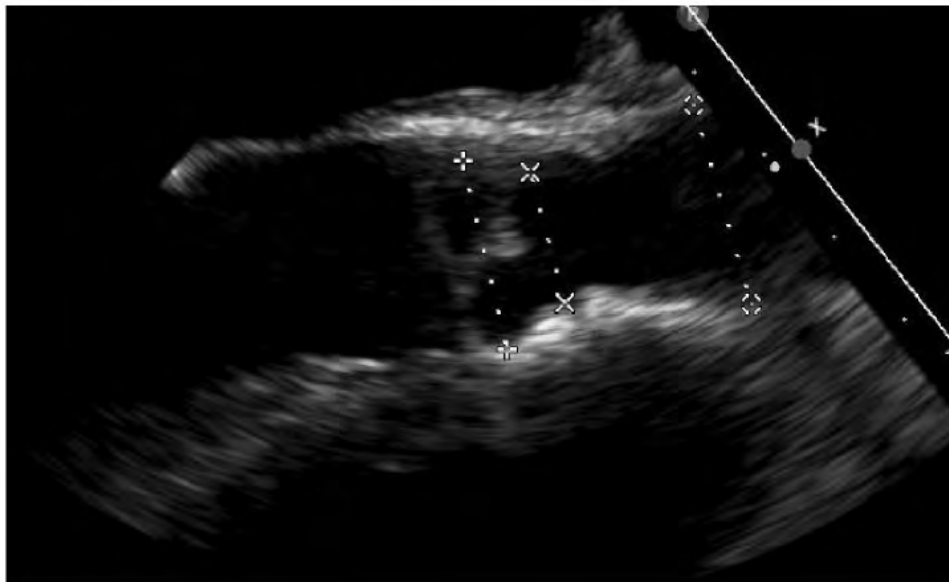


Figure 2: Case 1. Preoperative echocardiography. Longitudinal view of the aortic root with measurements: aortic annulus diameter 17 mm, aortic diameter at the aortic ridge 11 mm, diameter of the ascending aorta 19 mm.

Case 2 Presentation

A 35-year old female with a severe familial hypercholesterolemia was referred to surgery because of the worsening of her symptoms related to a calcified and extended SVAS. Angio CT scan (Figure 3) and TEE (Figure 4) confirmed the aortic obstruction with a huge protrusive calcification and a partial porcelain aorta; the mean gradient was 50 mmHg and there was a mild aortic regurgitation due to thick leaflets. Preoperative coronary angiography detected a pre-occlusive ostial lesion of the right coronary artery (RCA) which needed to be bypassed. There was no associated Williams syndrome. Surgical repair was done through median sternotomy using crystalloid cardioplegia arrest and moderate hypothermic cardiopulmonary bypass. The proximal RCA was first bypass using a short segment of saphenous vein graft. Then, the distal ascending aorta was transected showing the impressive protrusive intra-luminal calcifications (Figure 5) and all the calcified aorta was resected till the sinotubular junction. The noncoronary and right coronary sinuses were free of calcifications and they were incised till the aortic annulus for the implantation of two triangular pericardium patches (Edwards Lifesciences™) to augment both sinuses (Figure 6, 7); the left coronary sinus and the left main ostium were mild calcified and they were not involved in the aortic root reconstruction to avoid any technical issue. Then, the ascending aorta was replaced with a 24 mm prosthetic graft (Figure 8) and finally, the vein graft was implanted into the prosthetic tube. Postoperative outcome was event-free. Patient recovered very well and CT scan at 1-month follow-up showed a good anatomic result without echocardiographic residual gradient but with a persistent mild aortic valve regurgitation.

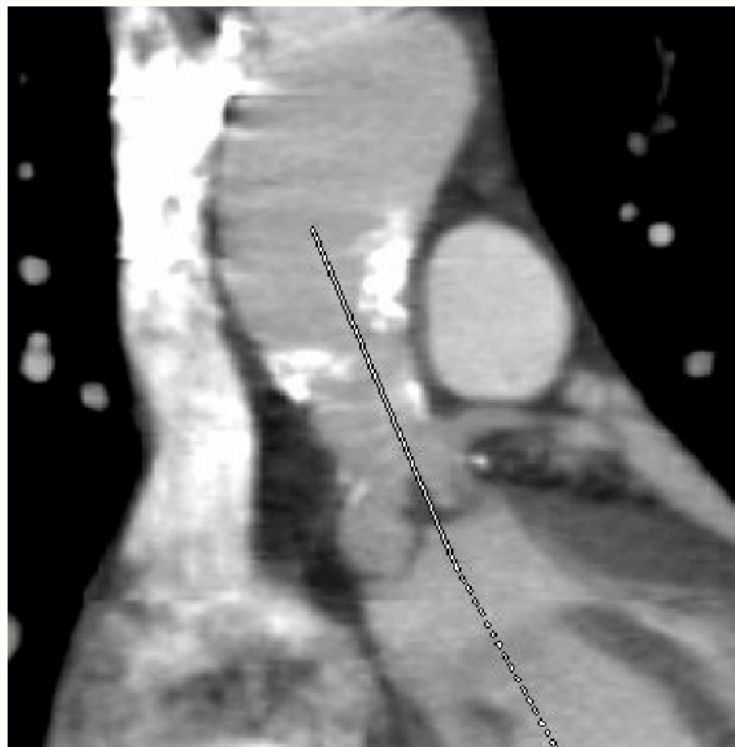


Figure 3: Case 2. Preoperative Angio CT scan of the ascending aorta showing the protrusive and calcified ridge; the left main is free of lesion with mild calcifications.



Figure 4: Case 2. Preoperative echocardiography. Longitudinal view of the aortic root showing the aortic valve with thick leaflets and the protrusive calcified ridge. Measurements: aortic annulus diameter 20 mm, aortic diameter at the ridge 11 mm, diameter of the ascending aorta 28 mm.

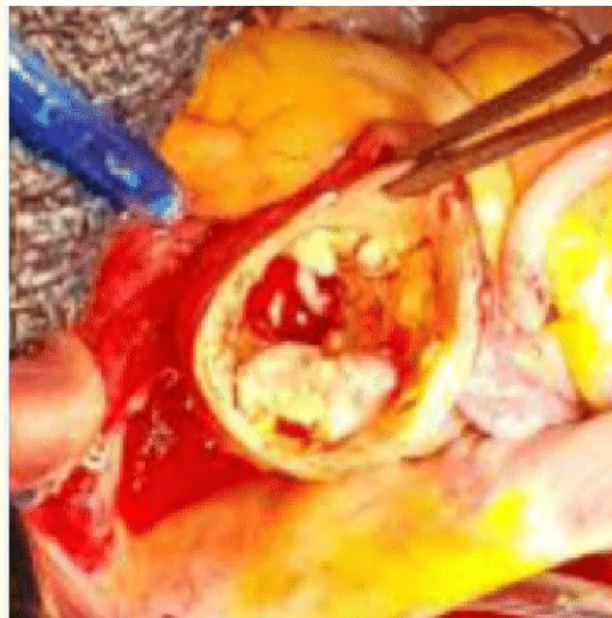


Figure 5: Case 2. Operative view of the ascending aorta with the protrusive calcifications.

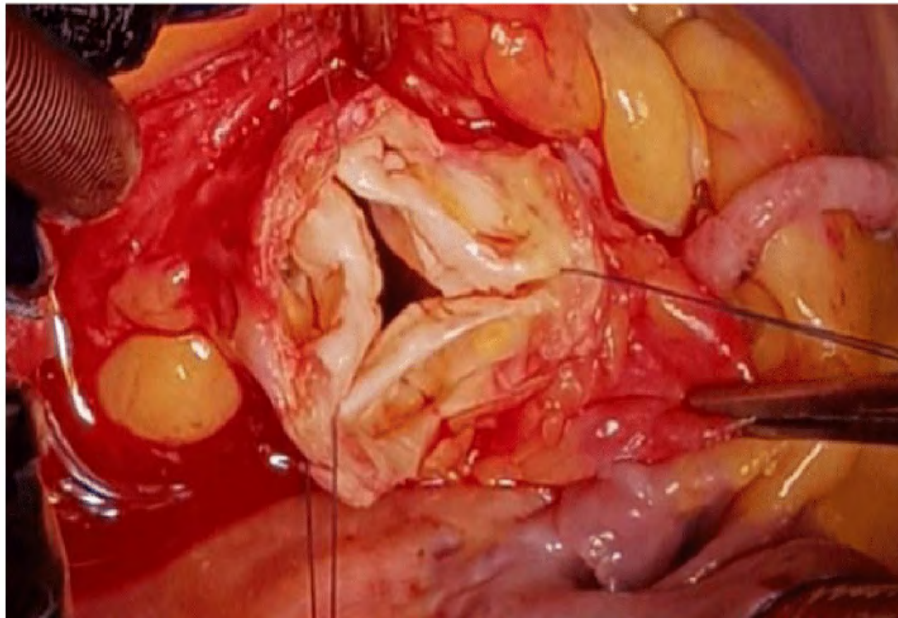


Figure 6: Case 2. Aortic root after resection of the ascending aorta and V shape incision of the non-and right-coronary sinus.



Figure 7: Case 2. Aortic root after implantation of triangular bovine pericardium patch in both sinuses.

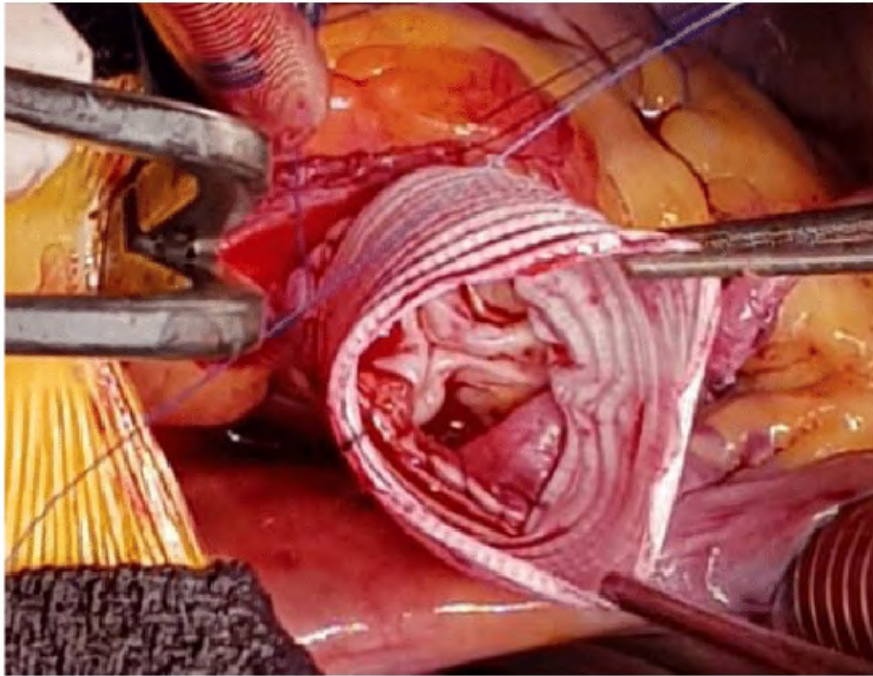


Figure 8: Case 2. Operative view of the prosthetic aortic graft and the reconstructed aortic root showing a good morphology of the aortic valve with thick leaflets.

Discussion

Supravalvular aortic stenosis is characterized by narrowing of the aorta, starting at the sinotubular junction and sometimes extending up to the ascending aorta. There are 3 types of SVAS: focal segmental (hourglass), which is the most typical; tubular (diffuse); and membranous, the least prevalent type [2,5]. SVAS represents an important feature of Williams syndrome but may also occur as familial forms or sporadic cases. Regarding the natural history of the disease, it is advocated to observe a wait-and-see approach in patients with moderate lesions and to indicate surgical repair only in children with severe and symptomatic lesions [6,7]. However, occurrence of symptoms can be delayed and surgery may be indicated at adult age as in our reported cases.

Our two patients had isolated sporadic SVAS which is rare; they did not have Williams syndrome or associated heart abnormality. In the younger patient, it was a simple and focal type; in the middle age lady, it was a tubular type with diffuse calcifications related to the ancientness of the disease and probably to the severe associated familial hypercholesterolemia which could explain also the associated coronary lesion observed even if SVAS are prone to coronary premature atherosclerosis [5].

In surgery to treat SVAS, the aim is to reduce the transaortic gradient as much as possible. The surgical repair is focused on restoring the geometry of the aortic root through various iterations of patch aortoplasty. In 1961, McGoon [8] described the single-patch repair of the noncoronary sinus. Later, Doty [9] introduced a pantaloons patch repair of the noncoronary and right coronary sinuses. However, up to 40% of patients who have undergone single or double-patch repairs have required later operations for left ventricular outflow tract obstruction or aortic valve regurgitation [10,11]. Subsequently, Brom [12] described a three-patch repair of all aortic sinuses, aiming at restoration of the aortic root configuration with a symmetric reconstruction of all 3 sinuses. This technique has improved midterm results in terms of relief of the obstruction and incidence of aortic regurgitation [5,6,10,11].

However, in the two reported cases the three-patch repair technique was not used and the surgical technique was adapted to the specificity of each patient. In the younger patient, the aortic wall was flexible and the intraluminal ridge could be easily removed; consequently a simple single-patch repair of the aorta extended to the noncoronary sinus was done without impact on the aortic regurgitation. In the second and more complex case, all the calcified ascending aorta had to be removed till the sinotubular junction to reach flexible tissues and a modified Brom technique was done with a only two-patch repair to avoid getting involved the calcified left main and left coronary sinus in the repair of the aortic root and having to do a total root replacement; consequently surgery was completed by a simple sus-valvular replacement of the aorta.

In both case, the associated mild aortic regurgitation was a concern. An asymmetric reconstruction of the root as well as a patch over-sizing may compromise the geometry and the stability of the aortic valve coaptation leading to a significant aortic regurgitation [6,10]. In both case, per-operative TEE and postoperative echo assessment confirmed the stability of the aortic regurgitation; but a strict follow-up is mandatory. The associated lesion of the RCA was also a concern in the second case; it was an ostial lesion without any connection with the aortic ridge or the leaflet, as it could be observed in child [2]; it is described as the consequence of the hemodynamic disturbances in the sinus [5,11], probably heightened by the severe lipid disorder in this case. The ostium of the RCA was virtual; a patch angioplasty was not appropriate and a vein grafting of the vessel was the best option; the postoperative CT scan showed an evolution to a complete occlusion of the proximal RCA. In the two cases, the postoperative outcome was event-free with a quick recovery and postoperative imaging tests confirmed good hemodynamic and functional results. However, both patient have a short follow-up, less than 1 year, and they need a regular assessment regarding the potential worsening of the aortic valve status despite a good anatomic reconstruction of the aortic root.

Conclusion

Sporadic SVAS case in adult is exceptional. For surgical repair, extensive calcifications and/or aortic valve regurgitation are challenging and lead to adapt the technique in each case, with the 3-patch aortoplasty as the reference technique to improve gradients and avoid reoperation.

Disclosure

The authors declare no conflict of interest.

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