

Surgical Management of Discrete Supravalvular Aortic Stenosis by Extended Patch Aortoplasty

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Objective: Supravalvular aortic stenosis is the rarest form of left ventricular outflow obstruction. Several techniques for symmetric reconstruction of the aortic root in congenital supravalvular aortic stenosis had been described. In this study, we demonstrate our experience with the extended patch aortoplasty technique.

Materials & Methods: Between October 2001 and March 2005, 11 patients with discrete supravalvular aortic stenosis had pantaloon patch aortoplasty in Abu-El Reech students' Hospital. They were 4 males, and 7 females ranging in age from 1½-9 years (mean age 5.2 years). Three patients (33%) had manifestations of Williams syndrome. All of the patients had the typical harsh ejection systolic murmur. One patient (9%) had associated hypoplastic aortic arch. No associated other cardiac anomalies or significant valvular aortic stenosis were present. The peak gradient across the obstruction ranged from 70-120 mmHg. All the patients underwent pantaloon shaped pericardial patch aortoplasty through an inverted Y-shaped incision in the aorta under deep hypothermia (28o) to enlarge the right and non coronary sinuses.

Results: No operative mortality or morbidity occurred in our study. The peak gradient across the obstruction dropped markedly [0-30 mmHg (mean 10)]. During surgery four patients (40%) had thickened aortic cusps without significant valvular aortic stenosis. After surgery, the typical harsh ejection systolic murmur disappeared completely.

Conclusions: Extended patch aortoplasty produces symmetrical enlargement of the right, and non coronary sinuses. It provides more physiologic pattern, preserves the valve function better than augmentation of the non-coronary sinus only. It is technically easy, yet it should be done accurately.

Supravalvular aortic stenosis (SVAS) represents an important feature of Williams syndrome (Elfin facies, and mental retardation), but it is also found in a familial form or as sporadic cases. The underlying cause is now known to be a spontaneous or an inherited mutation of the elastin gene on chromosome 7 [1]. The defining feature of the malformation is aortic narrowing at the level of the sinotubular junction but in some cases, there is narrowing of the entire ascending aorta, and arch branches. SVAS may also occur as part of a generalized arteriopathy involving both the systemic, and pulmonary arteries[2]. There are three morphologic variations: (a) Seventy five percent of supravalvular anomalies consists of discrete hour glass deformity, which consists of an extreme apparent wasting of the aorta at distal extent of sinuses of Valsalva, i.e. above the aortic valve commissures in association with some dilatation of sinuses of Valsalva, and absence of post-stenotic dilatation Fig. (1).

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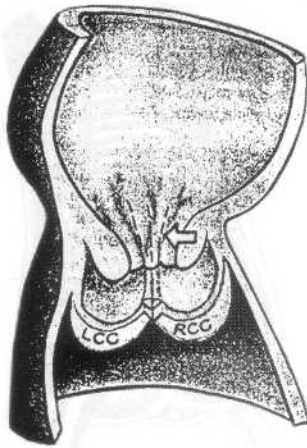


Fig. (1): Typical hourglass deformity of the aorta (After Flaker et al., 1983)

(b) More diffuse narrowing is detected in 25% of children. It consists of thickening of the aortic wall with a narrow lumen that may extend over a variable distance and may reach the aortic arch, and its branches. (c) In addition there may be a localized membrane immediately above the valve[3]. Histologic examination of the hourglass and membranous types revealed intimal thickening, medial disorganization, and thickening, with areas of necrosis, and calcification. The intimal thickening may involve the edges of the aortic valve leaflets occasionally which may become adherent to the intimal shelf producing stenosis of the inlet into the sinus of Valsalva thereby causing coronary insufficiency. In extreme cases, the proliferative process may extend into, and narrow or even obstruct the ostium of the coronary artery [4,5].

It was also found that significant coronary artery abnormalities were found in 49% of cases with thickening of the intima and media. In addition to the effects of left ventricular pressure after load, patients are at risk for myocardial ischemia due to coronary ostial stenosis, decreased blood flow to the coronary sinus, and hypertension related coronary arteriosclerosis[2]. Associated anomalies include valvular aortic stenosis, multiple peripheral pulmonary artery stenosis, narrow aortic arch which may involve its branches and VSD [4, 5].

The diagnosis of supravalvular aortic stenosis depends mainly on echo cardiography, as the patient is usually asymptomatic. Right ventricular hypertrophy may suggest concomitant pulmonary obstruction, coronary angiography may be needed to show any coronary insufficiency. Supravalvular aortic stenosis usually occurs with idiopathic hypercalcemia of infancy, since both conditions have a similar set of features. During

clinical examination there may be difference in the measurement of blood pressure between the two limbs due to peripheral arterial stenosis [3].

Surgical repair was first described by placement of a tear-drop or diamond shaped patch across the supra-valvular ring. The short term results were acceptable, and it remained the technique of choice for many years. However, a report by Keane, and his colleagues noted that a significant portion of patients (75%) of patients has a gradient greater than 30 mmHg. Flaker and his colleagues also reported residual gradient and aortic insufficiency[6,7], Fig. (2, 3).

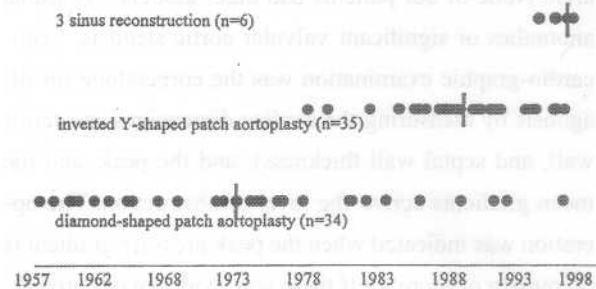


Fig. (2): Date of operation grouped according to surgical technique for augmentation of aortic root in SVAS (After Stamm et al., 1999).

The extended patch technique, described by Flaker and his colleagues in 1975, provides more symmetric augmentation of the aortic root. This is done by inserting an inverted bifurcated patch into the non coronary, and the right coronary sinuses[6]. The aim of this report is to demonstrate our experience with this technique.

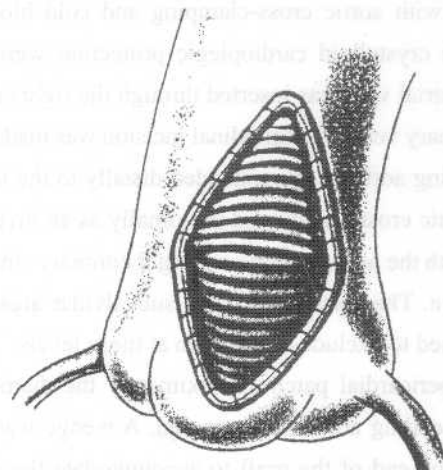


Fig. (3): Surgical technique used for relief of supravalvular aortic stenosis. A diamond shaped patch in the non coronary sinus (After Stamm et al., 1999).

Patients & Methods

Between October 2001 and March 2005, 11 patients underwent surgical correction for congenital supra-valvular aortic stenosis in Abu El Reech students' Hospital. They were 4 males and 7 females ranging in age from 1½-9 years. Three patients had Williams syndrome. All of them were discovered during routine medical examination by the presence of a murmur.

All of the patients had typical harsh ejection systolic murmur. One patient had associated hypoplastic aortic arch. None of our patients had other associated cardiac anomalies or significant valvular aortic stenosis. Echocardiographic examination was the cornerstone for diagnosis by measuring the cardiac dimensions (posterior wall, and septal wall thickness), and the peak, and the mean gradients across the level of obstruction. The operation was indicated when the peak pressure gradient is 50 mmHg or more, or if there was evidence of coronary insufficiency either by angiography (none of our patients reported an ischemic chest pain). In our patients, the peak gradient ranged from 70-120 mmHg.

Surgical Technique

Cardiopulmonary bypass was established after cannulation of the distal ascending aorta and the right atrial appendage in the standard fashion. Deep hypothermia (28%) with aortic cross-clamping and cold blood antegrade crystalloid cardioplegic protection were used. A left atrial vent was inserted through the right superior pulmonary vein. A longitudinal incision was made in the ascending aorta. It was extended distally to the level of the aortic cross-clamp, and proximally as an inverted Y into both the non coronary, and right coronary sinuses of Valsalva. The aortic valve, and subvalvular areas were examined to exclude obstruction at those levels.

A pericardial patch approximately the diameter of the ascending aorta was prepared. A wedge was taken from one end of the graft to accommodate the area of aorta around the right coronary ostium giving a pantaloons shape to the patch Fig. (4).

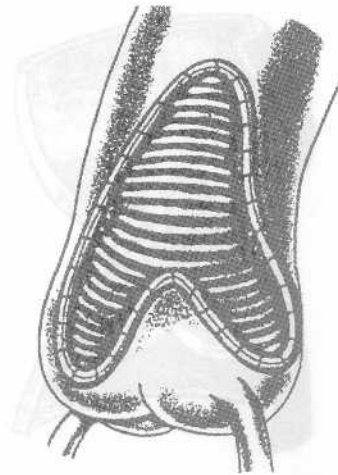


Fig. (4): Surgical technique used for relief of supra-valvular aortic stenosis. A Pantaloons shaped patch in the right and non coronary sinuses (After Stamm et al., 1999).

The patch was sewn to the aortic incision using running 6/0 polypropylene suture.

After completion of the anastomosis, the suture line was tested, and the aorta was declamped. Before closure of the sternum, control of systemic blood pressure was mandatory by vasodilators (Nipride or Tridil). In the ICU, the patient was weaned from the ventilator when the hemodynamics became stable and the chest tube drainage was average. A postoperative echo was done immediately after surgery to measure the gradient across the level of obstruction. The patients didn't receive anticoagulation.

Results

No operative mortality or morbidity in our study. The peak gradient across the obstruction dropped to 0-30 mmHg (mean 10) immediately after surgery. During surgery four patients (40%) had thickened aortic cusps without significant gradient across the valve. After surgery, the typical harsh ejection systolic murmur disappeared.

Discussion

The morphologic spectrum of SVAS had been well defined since 1970. The concept of an isolated supra-valvular membrane had been abandoned, and it had become clear that in most but not all cases the supra-valvular narrowing is part of a general disease of the arterial wall with a genetic origin. However, the reason that the stenosis is most prominent at the sinotubular junction remains subject to speculation. Because the sinotubular

junction is at the level of the tops of the valve commissures, the geometry of the entire aortic valve apparatus is disturbed[7]. Abnormalities of the valvular tissues are said to be found in 30-54% at operation or necropsy. Stamm and his colleagues, examined the angiograms, and the echocardiograms of 37 patients, and studied eight pathologic specimens. They found that partial adhesion of the leaflets to the stenosing ridge was observed in 54% of cases, and the leaflets were thickened, and less mobile in 30% of cases. Forty-five percent of angiograms showed evidence of coronary ostial stenosis. The sinuses of Valsalva were significantly enlarged in 75% of cases[8].

The goals of surgical repair should be (1) relief of stenosis (2) restoration of normal root geometry and (3) allowance of growth of site of repair[9].

In the present study, only two cases (2/11) were found to have bicuspid aortic valves during surgery. This was contrary to the usual description of the morphology of the disease by Delius, and his colleagues who reported that 47% of these patients had bicuspid aortic valves[10].

During surgery 40% of patients had thickened aortic valve cusps but without significant obstruction at the valvular level as detected by preoperative echocardiographic examination. This explains why aortic valvotomy was not done during the surgical procedure. On the other hand Stamm, and his colleagues performed digital or sharp valvotomy of the aortic cusps in 5 cases (5/56), mobilization of the tethered aortic cusps in 5 cases (5/56), and aortic valve replacement in 2 cases despite the absence of echocardiographic evidence of obstruction in their report[2].

The study of Stamm was conducted to compare between three techniques to repair discrete supra-avalvular aortic stenosis; the diamond shaped patch, the extended patch aortoplasty by pantaloon patch (1975), and 3-sinus reconstruction technique described by Brom in (1988). They concluded that the last two techniques are more physiologic. They added that they provide symmetrical sinus enlargement and are associated with lower mortality and reoperation rates than the first one[2]. This may be attributed to the fact that this technique only enlarges the noncoronary sinus.

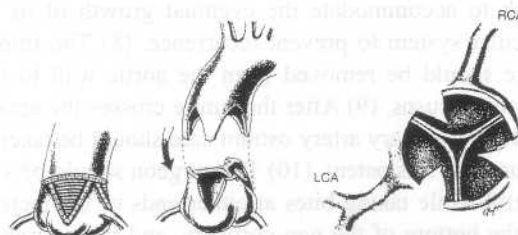


Fig. (5): Surgical technique used for relief of supra-avalvular aortic stenosis. The 3-sinus reconstruction technique (After Stamm et al., 1999)

Resection of the fibromuscular ridge above the left coronary cusp which was carried out in one case in the present study was frequently attempted by many surgeons. However it was proved impossible in most cases because the ridge usually represent a constriction of the thickened aortic wall rather than a circumscribed fibrous stricture[2]. Such resection allows the left cusp to assume a normal position. However, in some cases an incision into the left sinus and patch repair might be required before normal positioning of the left cusp[10].

In the present work pericardial patch was used which was concomitant with what Delius and his colleagues reported. They stated that pericardium or polytetrafluoroethylene were preferable as patch materials. Such assumption was based upon the failure of neointimal lining of Dacron patch leaving a thrombotic surface resulting in transient ischaemic attacks in one of their cases[10].

After surgery the gradient dropped markedly, this coincides with results of Brown and his colleagues who showed that the mean pressure gradient was reduced to 21 mmHg in the early postoperative period[11].

The extended patch aortoplasty technique first described by Doty and his colleagues should be done accurately. The surgeon should take care of the following points: (1) The longitudinal limb of the incision should extend up as far as the cross clamp, while the transverse limb should extend down to the bottom of the coronary sinus. (2) While making the aortotomy incision, the bifurcation point should start well above the area of discrete narrowing. (3) The incisions across the fibrosing ring should be done approximately 180 degrees apart. (4) The limb of the incision into the right coronary sinus is made to the left of the right coronary ostium this means that the patch is put straddling the right coronary ostium. (5) In cases with previous aortotomy, the incision is made at the site of previous aortic incision. (6) The suture line starts at the point of bifurcation of the inverted Y incision, and then extends to either side. (7) The surgeon should intend to oversize the width of the

patch to accommodate the eventual growth of the vascular system to prevent recurrence. (8) The fibrous ridge should be removed from the aortic wall to free the aortic cusps. (9) After the suture crosses the area of the right coronary artery ostium care should be taken to ensure that it is patent. (10) The surgeon should be very careful while taking bites at either ends of the incision (in the bottom of the non-coronary, and right coronary sinus), because taking additional sutures at this area, after release of the aortic clamp is extremely difficult. (11) Closure of the pericardium or part of its length is done, as some patients may need aortic valve replacement in the future.

Delius, and his colleagues, studied the long term results of 15 cases who had extended aortoplasty between 1975 and 1983, they concluded that the main risk factors for residual gradient after surgery or reoperation is the pathologic condition of the aortic valve and bicuspid aortic valve[10]. Pathologic changes of coronary arteries, and myocardial ischemia in SVAS have been described by many authors. Although they are rare, they are strong indications of early surgery. The pathologic changes in the arterial wall may be attributed to elevated pressure inside the vessels due to obstruction at the sinotubular junction or due to generalized arteriopathy[12, 13]. In addition to severe ventricular hypertrophy, coronary circulation of patients with severe arteriopathy may be more sensitive to changes in blood pressure during the perioperative period[14]. In the present study, none of our cases had coronary insufficiency.

Limitations of the Study

Due to rarity of the disease, the number of patients in the present study was few, that is 11 cases were operated upon during a period of 4 years. The largest study which was conducted by Brown and colleagues collected the data of 73 patients over a period of 28 years. They were operated upon between 1962-2000. This would represent the same average number of patients per year in our study.

Conclusions

Supravalvular aortic stenosis is a rare form of left ventricular outflow tract obstruction. It occurs mostly with Williams syndrome. There are three types, hour-glass, membranous, and diffuse. The first two types have the same pathological features, and the same surgical management. Many years ago surgical procedures entailed putting a teardrop or diamond shaped patch.

This was modified later on by Doty and his colleagues, who described the technique of extended aortoplasty. It produces symmetrical enlargement of right and non coronary sinuses and more physiologic flow pattern, and preserves the aortic valve function. It is also associated with the least re-operation rate.

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