OUTCOME OF SURGERIES IN SUPRAVALVULAR AORTIC STENOSIS.

PROJECT

BY

DR. ABHAY.S. JAIN

MCh. CARDIO VASCULAR THORACIC SURGERY RESIDENT

2014-2016

DEPARTMENT OF CARDIO VASCULAR THORACIC SURGERY
SREE CHITRA TIRUNAL INSTITUTE FOR MEDICAL SCIENCES AND TECHNOLOGY, TRIVANDRUM, KERALA, INDIA – 695011

DECLARATION

I hereby declare that this thesis entitled "**Outcome Of Surgeries In Supravalvular Aortic Stenosis**" has been prepared by me under the able guidance of Additional Professor **Dr. Vivek Pillai**, Additional Professor **Dr.Baiju S.Dharan,** Department Of Cardiovascular and Thoracic Surgery, at Sree Chitra Tirunal Institute For Medical Sciences & Technology, Thiruvananthapuram.

Date :
Place :

DR. ABHAY.S. JAIN
MCh. CVTS resident,
Department of Cardiovascular and thoracic surgery,
SCTIMST, Thiruvananthapuram

CERTIFICATE

This is to certify that this thesis entitled "**Outcome Of Surgeries In Supravalvular Aortic Stenosis**" has been prepared by **Dr ABHAY.S. JAIN**, MCh CVTS resident, Department of Cardiovascular and Thoracic Surgery at Sree Chitra Tirunal Institute for Medical Sciences & Technology, Thiruvananthapuram. He has shown keen interest in preparing this project.

(GUIDE)
Dr. Vivek Pillai
Additional Professor
Department of Cardiovascular and Thoracic Surgery,
SCTIMST, Thiruvananthapuram

(COGUIDE)
Dr.Baiju S.Dharan
Additional Professor
Department of Cardiovascular and thoracic surgery,
SCTIMST, Thiruvananthapuram

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Professor Jaykumar K.
Senior Professor and Head
Department of Cardiovascular and thoracic surgery,
SCTIMST, Thiruvananthapuram

Date :
Place :

ACKNOWLEDGEMENT

During the years working with this thesis, I have met many generous people who have shared with me their vast knowledge and enthusiasm for research. I would like to express my sincere gratitude to all my colleagues, friends and family who in different ways have contributed to this thesis.

It is a matter of great privilege for me to express my heartfelt gratitude and sincere regards for my Professor, Prof. Jayakumar K. (Professor and Head, Department of CVTS, Sree Chitra Tirunal Institute for medical Sciences and Technology), for his keen interest, constant inspiration and encouragement throughout the study. Mere words are insufficient to express my profound indebtedness and deep gratitude to my teacher and Guide Dr. Vivek Pillai, Additional Professor, Department of CVTS, Sree Chitra Tirunal Institute for medical Sciences and Technology. His depth of knowledge, readiness to help, understanding attitude, concern for excellence, limitless patience, invaluable guidance and able supervision has enabled me to undertake and complete the work on this project.

I am indebted to Dr. Baiju S.Dharan, Additional Professor, Department of CVTS, Sree Chitra Tirunal Institute For Medical Sciences and Technology my Co-Guide, for his scientific guidance, for sharing expertise and skills in scientific writing and also for making me push my limits and believing in me.

I also appreciate the quick and valuable comments on my manuscript and thesis frame. I am highly indebted to Dr.Varghese T Panikar, Additional Prof, of Department of CVTS, Sree Chitra Tirunal

Institute for medical Sciences and Technology, for his profound inspiration and ever helpfulness.

I am thankful to Dr.Bineesh K R.Assistant Prof, Department of CVTS, Sree Chitra Tirunal Institute for medical Sciences and Technology, for invaluable suggestions and being a source of inspiration. I am highly grateful for the immense support provided by my seniors and consultants Dr. Sudip Dutta baruah and Dr. Yadav Srinivasan, Assistant Prof in the department of CVTS, Sree Chitra Tirunal Institute for medical Sciences and Technology, who were always ready to help as elder brothers.

No words will be enough to acknowledge the great help provided by my colleagues particularly Dr. Neeraj Tapdiya, Dr. Saurabh Nanda, Dr. Chirag S. P. and Dr. Simon Philipose for their constant encouragement and support.

I would like to thank my colleague and friend Dr. Renjith S for his support throughout my study. I am thankful to my wife because without her support, encouragement and love, I could never have been what I am today. I am thankful to my family who provided constant support and encouragement, and to whom I owe everything in life.

ABBREVIATIONS

SVAS	-	Supra Valvular Aortic Stenosis
AR	-	Aortic Regurgitation
MR	-	Mitral Regurgitation
LVOT	-	Left Ventricular outflow tract
LVH	-	Left Ventricular hypertrophy
VSD	-	Ventricular septal defect
PS	-	Pulmonary stenosis
RPA	-	Right pulomonary artery
SAM	-	Systolic anterior motion
AVR	-	Aortic valve replacement
BAV	-	Bicuspid aortic valve
CPB	-	Cardio pulmonary bypass

CONTENTS

INTRODUCTION

INTRODUCTION

Supravalvular aortic stenosis (SVAS) is a rare anomaly in which there is an exaggerated narrowing at the sinotubular junction.It can either be a localised narrowing of STJ or a diffuse variety affecting the ascending aorta, aortic arch and its branches This anomaly is often associated with Williams syndrome. There may be generalized hypoplasia of the ascending aorta and more distal arterial tree as well as stenoses in the pulmonary artery tree.Williams syndrome is a congenital multisystem, developmental disorder resulting from the deletion of approximately 28 genes on chromosome 7q11.23, in 1 in 8000 live births. (1) It affects the vascular, connective tissue, and central nervous systems.(2) Williams syndrome been associated with congenital cardiac malformations in approximately 10% of patients and symptomatic narrowing of arteries in up to 80%.(3) Supravalvular aortic stenosis (SVAS) is reported to be the most common cardiovascular abnormality in Williams syndrome.

Despite considerable attention to the importance of maintaining the integrity of the aortic root during supravalvar reconstruction, there has been little focus on the management of other components of the aortic root and left ventricular outflow tract, including the aortic valve, subvalvular region, and coronary arteries.

Surgical techniques for repair of supravalvular aortic stenosis (SVAS) are numerous which include McGoon’s one patch, Doty’s two-patch, and Brom’s three-patch method. Data definitively supporting one technique over another have been elusive. No technique is considered gold standard for SVAS repair.Each technique have its own pros and cons.There is need to study the surgical outcomes of different procedures for SVAS to decide optimal procedure for it.

In this review we evaluated issues and clinical outcomes of these techniques at SCTIMST institute.

AIMS AND OBJECTIVES

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1. To evaluate and assess the morphology of ascending aorta and post operative gradient in patients after SVAS repair.

2. To evaluate the improvement in symptoms (NYHA functional class) following SVAS repair.

3. To evaluate the incidence of postoperative complications

MATERIALS AND METHODS

MATERIALS AND METHODS

Design

Retrospective, single-centre, Observational case series in a Tertiary referral centre (SCTIMST).

Participants:

14 patients with Supravalvar aortic stenosis operated in our institute between 2006 to 2015.

Inclusion criteria:

Patients with

1. Supravalvar aortic stenosis
2. Those who underwent SVAS repair between 2006 to 2015 will be enrolled in the study.

Exclusion criteria : Nil

Approval from Technical Advisory Committee: taken before commencing the study.

Approval from Institutional Ethics Committee: taken before commencing the study.

Proposed duration of study:

1 years

Funding:

Not required

Proposed study protocol:

- After obtaining permission from IEC, data was collected from the medical records of patients by Principle investigators or co-investigators. The data was kept by the principle investigator. Patient details kept confidential.
- Data was analysed.

Investigations:

2D echocardiography was done using multiple views, including parasternal, apical long-axis, and suprasternal views were taken to look for aortic gradients,aneurysm around the patch,stenosis around the patch,aortic regurgitation aortic valve annulus and aortic dimensions.

CT Aortogram was done for accurate measurement of aortic dimensions,aneurysm and stenosis around the patch.transverse diameter of Narrowest part of the ascending aorta to transverse diameter of arch of aorta at innominate artery origin ratio was taken to look for any stenosis or dialatation of ascending aorta with time.

ECG and Chest X-ray was taken at routine follow up of the patient in CSOPD.

Outcome parameters:

Evaluation of study Objectives.

Method of statistical analysis:

Quantitative variables were described by mean, sd, minimum and maximum values. Qualitative variables were described by percentage distribution. Between group comparison of qualitative variables were done

by chi- square test and Fisher's exact test and Comparison of quantitative data between two were analyzed by independent sample t test and that of more than two group was analysed by ANOVA. Pre test post comparison of quantitative variables were analysed by paired t test. a p value of 0.05 was taken as the level significance. Data analysis was performed using SPSS ver. 17.0.

CHI SQUARE TEST: Chi – Square test is a non-parametric test not based on any summary values of population. It is defined as $\chi^2 = \sum \frac{(0-E)^2}{E}$,

Where O is the observed frequency & E is the expected frequency of the same event (or for the same cell).

Expected frequency in a contingency table is calculated by the formula.

$$\text{Expected frequency} = \frac{\text{Row total} \times \text{column total}}{\text{Grand total}}$$

Comparison of two sample means. Independent sample t test

Let there be two different populations with means μ_1 and μ_2 and the standard deviations σ_1 and σ_2 respectively. Suppose the researcher draws two samples of sizes n_1 and n_2 from these populations and let $\bar{x}_1$ and $\bar{x}_2$ are the corresponding sample means, then

$H_0 : \mu_1 = \mu_2$

$H_A : \mu_1 \neq \mu_2$

The researcher wants to know whether the observed difference between $\bar{x}_1$ and $\bar{x}_2$ is statistically significant or not. If the difference is significant then the conclusion is that the difference is real and two samples

are from different populations having means μ_1 and μ_2. Z statistics is obtained as

$$Z = \frac{\left|\overline{x_1} - \overline{x_2}\right|}{SE(\overline{x_1} - \overline{x_2})} \text{ where}$$

$$SE(\overline{x_1} - \overline{x_2}) = \sqrt{\frac{\sigma_1^2}{n_1} + \frac{\sigma_2^2}{n_2}}$$

$\bar{x}_1$ = first sample mean

$\bar{x}_2$ = second sample mean

σ_1 = standard deviation of first sample

σ_2 = standard deviation of second sample

$SE(\overline{x_1} - \overline{x_2})$ = stand error of difference of means

If σ_1 and σ_2 *are not provided use S_1 and S_2 (first and second sample standard deviations)*

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REVIEW OF LITERATURE

REVIEW OF LITERATURE

Supravalvular aortic stenosis described for the first time in 1930 by an Italian pathologist,(4) has an estimated incidence of 1:20 000 live births.(5) The crude incidence of congenital heart defects is approximately 8 cases per 1000 live births. SVAS accounts for less than 0.05% of congenital heart defects. The sporadic form of SVAS is more common than the autosomal dominant form.

As previously mentioned, the sporadic form of SVAS is the most common (>50%) presentation(6) SVAS is a generalized disease of the arterial wall caused by the thickening of the media or intima layers, not related to atherosclerosis, which results in narrowing of the lumen of the ascending aorta or other arteries.

Supravalvular aortic stenosis (SVAS) is an uncommon but well-characterized inherited narrowing of the ascending aorta above the level of the coronary arteries.(3,7) The condition occurs as an isolated defect in individual patients with an autosomal dominant mode of inheritance or as part of Williams syndrome. Williams-Beuren syndrome is a genetic disorder characterized by mental retardation, ebullient personality, distinctive elf-like facial appearance, short stature, infantile hypercalcemia, abnormal vitamin D metabolism, and a wide spectrum of obstructive arteriopathies. It is caused by deletion of the elastin precursor gene on chromosome 7 (7q11.23) and several adjacent genes. More isolated elastin precursor gene deletions occur in familial and “sporadic” forms and are associated with similar cardiovascular manifestations without the additional manifestations of WilliamsBeuren syndrome. Cardiovascular manifestations are characterized by obstructive arterial lesions.

Supravalvar aortic stenosis (SVAS) lesion is variable and ranges from discrete ringlike thickening of the aortic media at the sinotubular junction to diffuse involvement with variable hypoplasia and thickening of the ascending, transverse arch, and descending aorta. Other left ventricular outflow tract abnormalities occur with SVAS, including aortic valve and coronary artery pathology.(8,9)

There is increased collagen, hypertrophied smooth muscle fibers, and a haphazard arrangement of thick, short elastic fibers, which are also reduced in number. Slit-like vascular spaces (lacunae) in the fibrotic intima and thick-walled vasa vasora are additional observations.(10)

Great systemic arteries that contain the largest number of ELN fibers in their media are the most affected. *ELN* mutations also result in peripheral pulmonary artery stenosis, such as supravalvular pulmonary stenosis or mesenteric and renal artery stenosis or coronary artery lesions.(9,11) Stenoses affecting different arteries are sometimes observed among different members within the same family, carrying the same *ELN* mutation.(5) Hypertension is often present in this patient group and is typically related to lack of systemic vessel distensibility, but occasionally may be secondary to renal artery stenosis.Intracranial focal and segmental stenotic artery disease can be responsible for stroke.(12,13)

Diagnosis of SVAS is established clinically by (1) systolic ejection murmur in the aortic area that radiates to the carotid arteries often accompanied by a thrill in the suprasternal notch, and (2) echocardiography that documents the SVAS distal to the valvular cusps.SVAS usually occurs as an hourglass stenosis above the aortic valve but it may also occur as a more diffuse thickening of the wall of the long segment of the aorta. SVAS is easily diagnosed by standard imaging methods, such as Doppler

echocardiography, which provides a more accurate definition of the lesions and their severity, and MRI, which gives information on associated vascular anomalies.

The molecular diagnosis of ELN arteriopathy relies on several methods that depend on the type of alteration: fluorescence in situ hybridization to detect *ELN* deletion in WBS, direct sequencing to identify point mutations or small insertion/deletion, Multiplex Ligation Probe Amplification and Real Time quantitative polymerase chain reaction to detect partial or complete *ELN* exon(s) deletions.(14)

Symptoms caused by SVAS usually develop in childhood. Rarely, symptoms may develop in infancy; in some cases, symptoms develop in the second or third decade of life. Most pediatric patients present because of a heart murmur or the features of Williams syndrome. Patients with Williams syndrome may also develop systemic hypertension and involvement of joints, peripheral pulmonary artery stenosis, coarctation of aorta, and mitral insufficiency.

Dyspnea on exertion, angina, and syncope develop in the course of the disease if SVAS is untreated. These symptoms indicate at least a moderate degree of LVOT obstruction. Because of the coronary artery involvement, angina may arise early and more often than in other obstructive LVOT lesions. Because of the risk of sudden death in SVAS, the development of angina and syncope should prompt immediate investigation.(15)

The physical examination focuses on upper extremity pulses, the precordium, heart sounds, and heart murmurs.

Asymmetrical upper extremities pulses

Discrepancies between carotid pulsations and upper extremity pulses and blood pressure are the characteristic clinical findings in SVAS. The discrepancies occur because the jet of blood flow from SVAS has a preferential trajectory into the brachiocephalic (innominate) artery (ie, Coanda effect).

Precordium

The precordium is usually hyperdynamic, and the apex of the heart is displaced laterally and inferiorly because of ventricular hypertrophy. A thrill in the suprasternal notch is usually felt because of the trajectory of the blood flow jet from SVAS.

Heart sounds

The first heart sound is generally normal. A narrowly split, single, or paradoxically split second heart sound and a fourth heart sound are present in severe SVAS.

Heart murmurs

The characteristic systolic murmur of SVAS is crescendo-decrescendo in shape, low pitched, and best heard at the base of the heart, sited higher than in valvular aortic stenosis. It mainly radiates to the right carotid artery and tends to peak during the last two thirds of ventricular systole if the obstruction is severe.

A high-pitched, short, early diastolic aortic regurgitation murmur is uncommon in SVAS unless the aortic valve has become damaged due to the

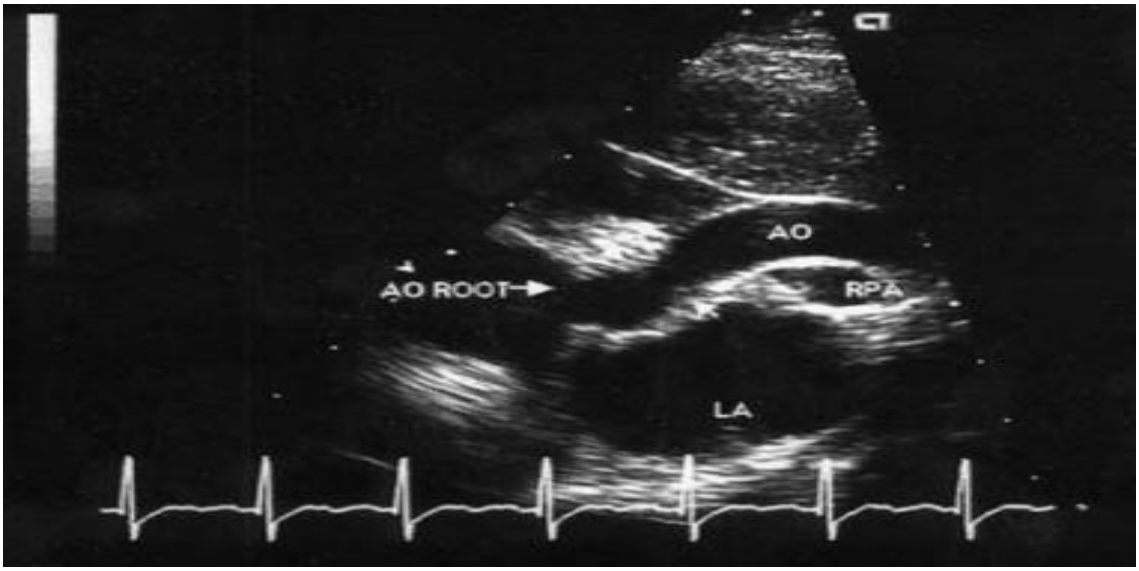
supravalve obstruction and has become regurgitant. An ejection click is absent.(15)

Clinical and echocardiographic findings in patients with *ELN* mutations vary widely, even within the same family, and range from calcifications of the ascending aorta in older individuals with minimally increased flow velocity to significant narrowing with impressively increased flow velocity. The phenotype may also include cases with isolated mild pulmonary stenosis.(16) The aortic valve may also be affected in SVAS, causing an additional source of left ventricle outflow tract obstruction. Varying degrees of aortic valve leaflet thickening or leaflet adhesion is seen in 45% of cases. In contrast to pulmonary circulation, arterial stenosis of the systemic circulation may worsen with time; thus, lifelong monitoring of the cardiovascular system is important.(14)

SVAS produces abnormalities that are evident on electrocardiography (ECG) and chest radiography. These include increased left ventricular voltages from left ventricular hypertrophy. ST and T wave changes may be present if there is coronary involvement. Additionally, if right ventricular outflow tract obstruction is present, there may be voltage criteria for right ventricular hypertrophy.(17)

The principal diagnostic test, however, is two-dimensional echocardiography. Cardiac catheterization along with angiography may be performed at an increased risk as indicated, but it may be necessary to evaluate the severity of the lesion and to confirm the coexisting anomalies prior to surgery if they cannot be accurately assessed with other modalities. Magnetic resonance imaging (MRI) may be utilized to evaluate for stenosis of the arch vessels or for better delineation of the anatomy if cardiac catheterization is not performed.(17)

The anatomic diagnosis of SVAS can reliably be made from two-dimensional echocardiography that uses multiple views, including parasternal, apical long-axis, and suprasternal (seen in the image below).



Two-dimensional suprasternal echocardiographic image of supravalvar aortic stenosis.(17)

In SVAS with hourglass deformity and diffuse hypoplasia, the diameter of the ascending aorta is smaller than that of the aortic root. In SVAS with fibrous diaphragm, the external ascending aortic diameter is normal, although an echogenic membrane is commonly observed above the sinuses of Valsalva.

Turbulent color flow mapping indicates the site of hemodynamically significant obstruction in relation to the origin of the coronary ostia. The incidence of coronary artery involvement is high in SVAS.(18)

Doppler peak gradient overestimates and, therefore, does not predict catheter-measured gradient well in patients with SVAS and may not be reliable in assessing its severity and guiding the need for intervention.(19)

Children and adolescents with catheter peak-to-peak (or Doppler mean) gradient of 50 mm Hg or more should have surgical intervention. The choice of procedures in these patients is similar to that indicated for valvar aortic stenosis.(20) Children and adolescents with catheter peak-to-peak (or Doppler mean) gradient of 30-50 mm Hg may be considered for surgical intervention if they are symptomatic, with angina, syncope, or dyspnea on exertion (class I). Asymptomatic patients who have developed ST/T-wave changes over the left precordium on ECG at rest or with exercise should also be considered for surgical intervention (class I). Aortic valve involvement and lower supravalve gradients may also warrant surgical intervention.

Surgical resection of the supravalvar obstruction and patch aortoplasty and multiple-sinus reconstructions (inverted bifurcated patch plasty and 3-sinus reconstruction) are the procedures of choice for the fibrous diaphragm and hourglass deformities.(21)

Associated coronary artery involvement is addressed with the following measures, which are performed at the same time as aortoplasty:

- Patch aortoplasty encompassing the left main ostium for circumferential narrowing of the left main ostium
- Excision of the fused leaflet from the aortic wall for ostial obstruction caused by a fusion of the aortic cusp to the supravalvar ridge.(22)
- Bypass grafting for diffuse narrowing of the left main coronary artery

In patients who have SVAS with diffuse narrowing, the ascending aorta and the arch of the aorta can be reconstructed using an aortic allograft or a pulmonary autograft.Surgical treatment of associated abnormalities of aortic valve and aortic arch vessels should be undertaken at the same time to

optimize the overall surgical outcome.(11) By limiting the lowest point of the aortci patch to the middle of the aortic sinus,the incidence of post operative aortic incompetence can be reduced.

Standard postoperative care and precautions for pediatric cardiac patients are also required for patients with SVAS. Postoperative complications include aortic insufficiency (in 25% of patients).

Exercise recommendations for children with SVAS and no coronary artery involvement are as follows:

- Mild stenosis (< 20 mm Hg), normal ECG findings, no symptoms - Full sports participation
- Moderate stenosis (21-49 mm Hg), mild left ventricular hypertrophy (LVH), no symptoms - Low static or moderately dynamic sports participation
- Severe stenosis (>50 mm Hg) or moderate degree of stenosis with symptoms - No competitive sports participation (at most recreational)
- Coronary artery stenosis or abnormal anatomy - No competitive sports participation (at most recreational and dependent on the level of obstruction)(23)

Williams syndrome, which is found in many children with SVAS, may be associated with infantile hypercalcemia with some risk of nephrocalcinosis, osteosclerosis with progressive joint limitation and abnormal gait, and neurodevelopmental delay. These children require multidisciplinary support. Use a coordinated management approach. They are also at risk of higher mortality than the normal population is, because of cardiac and noncardiac causes.(23)

Dwight McGoon and John Kirklin (24) from the Mayo Clinic first reported a one-patch teardrop-type repair of supravalvar aortic stenosis in 1961. Donald Doty (25) reported the use of an inverted Y-shaped patch which extended into two of the aortic valve sinuses in 1977. The technique described by A. Gerard Brom,(24) with three patches placed into the three aortic valve sinuses. This frequently also requires a patch in the distal ascending aorta. Steinberg's modification to the Doty's repair in which an extra patch is inserted in the left coronary sinus in addition to the inverted Y-shaped patch in the right and non-coronary sinuses.(26) More recently, John Myers (27) described an approach with three incisions into the three coronary sinuses and corresponding counterincisions into the distal ascending aorta which then insert into the openings created in the proximal ascending aorta. This technique has the advantage of not requiring autologous patch material, but is technically more demanding.Most recently Seo and collegues described sliding aortoplasty for SVAS repair.(28)

Surgical History of Operations for Supravalvar Aortic Stenosis(24)

Surgeon	Technique	Number of Sinuses Opened	Year Reported
McGoon	Single-patch	1	1961
Doty	Inverted "Y" Patch	2	1977
Brom	Three-patch	3	1988
Myers	Three-sinus incision	3	1993
DongMan Seo	Modified simple sliding aortoplasty	1	2007

When the stenosis is far from the valves and the coronary ostia the repair technique used by us is that described by McGoon et al,(29). Nevertheless, when the stenosis is very near the valves and the coronary ostia, the use of the Doty et al(25) technique may be preferred, which involves an inverted Y patch.

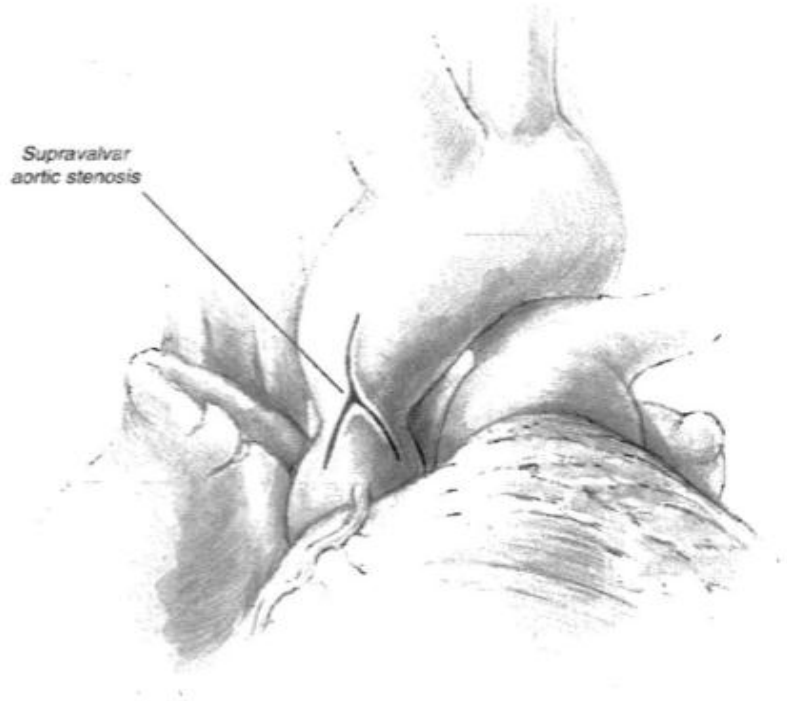
At present, there are several variations in the surgical technique for correcting this type of defect, although Hazekamp et al(30) did not find any significant differences in change in valve function, and found the efficacy of reducing the pressure gradient was similar and acceptable with various techniques. Their findings are in contrast with those of Stamm et al(31) in Boston, who analyzed cases occurring between 1957 and 1998 that included 75 patients, of whom 7 died perioperatively and the remainder of whom had a survival rate of 100% at 5 years and 77% at 20 years; at the end of the study the authors showed that diffuse stenosis had an influence on the outcome of this type of patient and that plasty of the 3 valves reduced the gradient more efficaciously than simple plasty of the noncoronary chest.

In the cases of recurrent serious stenosis, an alternative has even been to use the anastomosed valve graft of the of the free wall from the left ventricle to the descending aorta.(32) Similarly, another option that must be mentioned not only for cases of recurring stenosis but also for complex cases of diffuse stenosis, is repair with an autologous arterial graft from the pulmonary artery, as described by Al-Halees et al(33) in a 6-year-old patient.

Various surgical procedures for SVAS repair

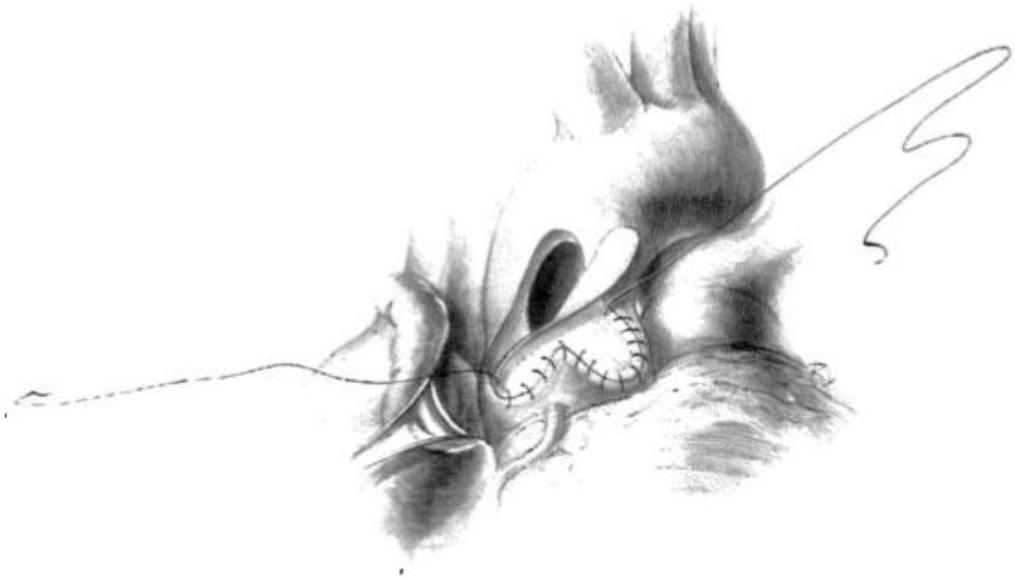
A. Inverted Bifurcatcd Patch Technique (34)

This technique was originally described by Doty and is appropriate for moderate or moderate to severe supravalvular aortic stenosis that does not involve important narrowing of the left coronary sinus of Valsalva.



A.1 With ascending aortic cannulation and a single venous cannula in the right atrium and following the application of the aortic cross clamp and infusion of cardioplegia solution, a longitudinal incision is made on the anterior surface of the proximal ascending aorta. The incision is bifurcated into the middle of the coronary sinus as well as into the right coronary sinus to the left of the right coronary ostium passing through the thickened sinotubular ridge. It is important that the right coronary ostium be carefully visualized and that the incision has adequate clearance from the right coronary ostium to allow subsequent suturing. Following completion of the

bifurcated incision the right coronary ostium sits on a small triangle of tissue directly anteriorly.(34)



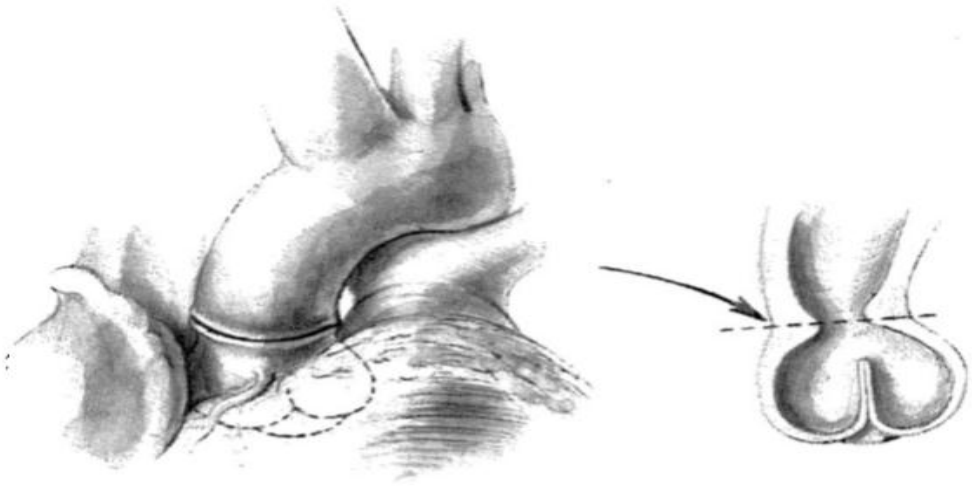
A.2 A generous pantaloon shaped patch is now sutured into the two sinuses of Valsalva. It is important to understand that the goal is to create bulging sinuses of Valsalva similar to those seen normally so that the patch should appear quite a bit larger than one would initially anticipate. Interestingly despite placing very generous patches in the two anterior sinuses it is rare that sufficient distortion of the aortic valve is created that aortic regurgitation ensues. (34)

The choice of patch material is dependent on the age of the patient. In the smaller, younger patient it is preferable to use autologous pericardium treated with 0.6% glutaraldehyde for 20-30 minutes. Pericardium is very much more hemostatic than synthetic alternatives. In the larger older patient it is probably wise to use collagen impregnated crimped Dacron (eg, II emashield). It is not wise to use PTFE (eg, Goretex) because of excessive needlehole bleeding in this location even when PTFE suture is employed.

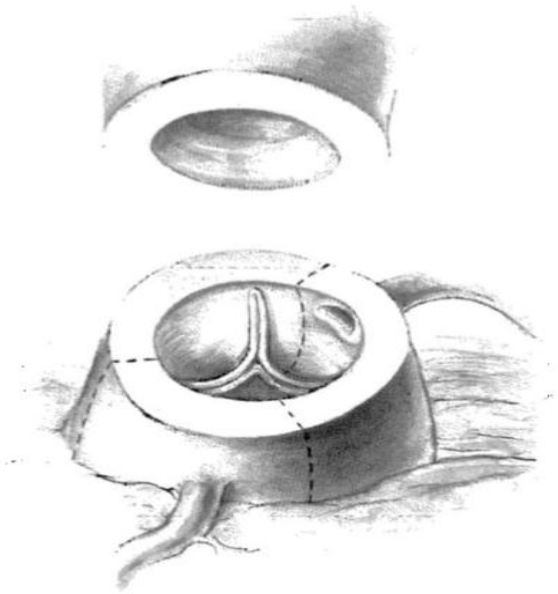
If this procedure is undertaken in conjunction with patching of the ascending aorta and the arch, one patch started in the arch and distal ascending aorta is used. The usual maneuvers are undertaken for de-airing the left heart including allowing an aortic vent site to bleed freely at the time of release of the aortic cross clamp.

B. Symmetric Three Patch Approach(34)

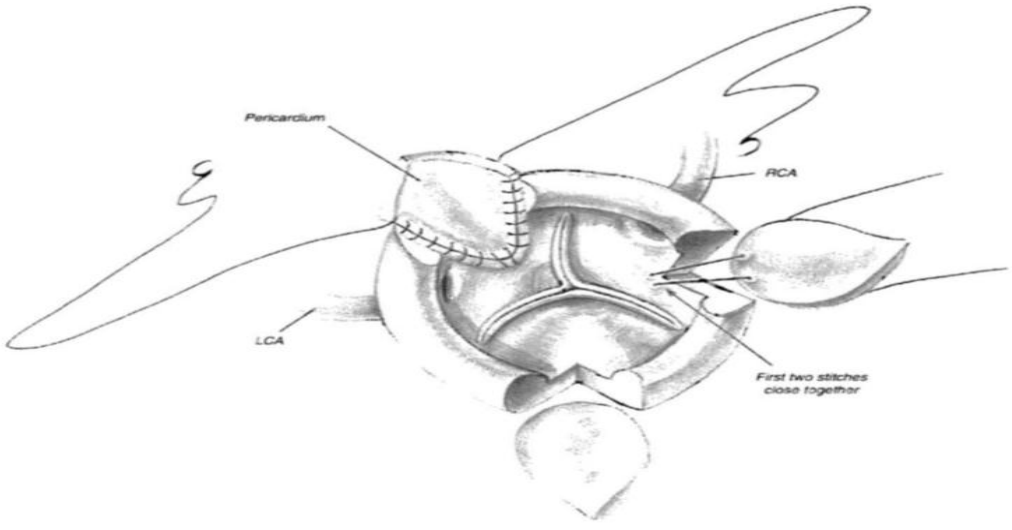
If there is important narrowing of the left coronary sinus as is often seen with severe forms of supravalvular stenosis, one option is to place three independent patches, one in each sinus of Valsalva. This can be achieved by advancement of the ascending aorta (see below). An alternative is to use autologous pericardium.



B.1 The ascending aorta is divided transversely at the level of the sinotubular junction or slightly above.(34)



B.2 Incisions are carried down into each of the sinuses of Valsalva. In the case of the left coronary sinus the incision is just to the right of the left coronary ostium. In the case of the right coronary sinus the incision is just to the left of the right coronary ostium. The incision is carried well into the sinus of Valsalva with care to avoid injury to the valve leaflets.(34)



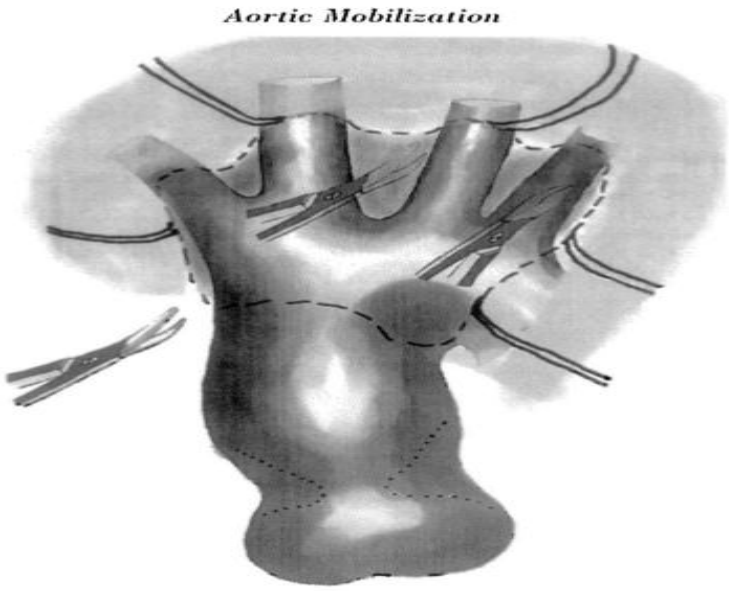
B.3 Teardrop shaped patches of autologous pericardium treated with 0.6% glutaraldehyde for 30 minutes are sutured into each of the sinuses of Valsalva. It is important to use a differential suturing technique, particularly

at the apex of the incision. Bites are widely spaced on the pericardium and closely spaced in the sinus of Valsalva. This differential spacing allows optimal supplementation of the sinus of Valsalva. As with the pantaloon shaped patch the goal should be to achieve a bulging normal shaped sinus of Valsalva so the pericardial patch is relatively large and redundant.(34)

It is very common to extend the two anterior patches into the ascending aorta. A longitudinal incision is made on the anterior ascending aorta. The two anterior patches are sutured together along their contiguous margins and extend into the distal aortotomy. Posteriorly the aorta is reconstituted directly to the supplemented sinotubular junction.

C. Direct Anastomosis Technique(34)

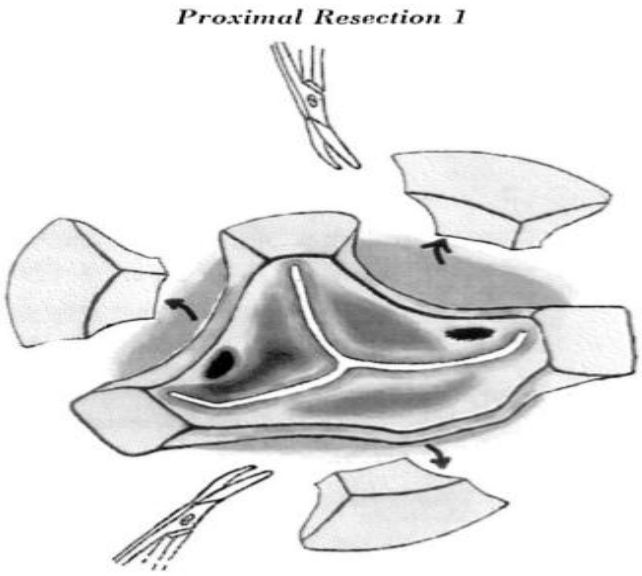
The technique of direct anastomosis can be applied when supravalvular aortic stenosis is well circumscribed and limited to the area in and around the sinotubular junction.



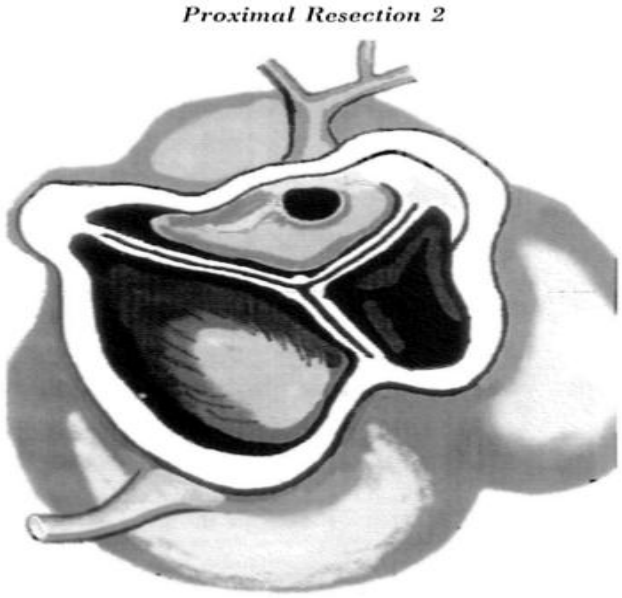
C.1 Mobilization of the distal ascending aorta and arch branches allows sufficient mobility for resection of discrete supravalvular aortic stenosis

with direct anastomosis. For those cases with more extensive involvement of the ascending aorta and aortic arch these techniques can still be combined with patching techniques to achieve lasting relief of obstruction while preserving aortic valve function and growth.(34)

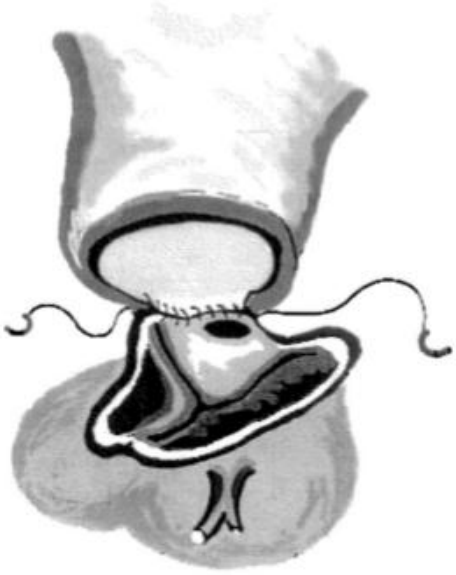
C.2 Complete resection of the thickened aortic wall is essential for adequate relief and involves resection of this tissue from proximal and distal stumps after division of the aorta just distal to the aortic valve commissural peaks.(34)



C.3 Resection lines must extend into the aortic sinuses and closely skirt the coronary orifices. This is most safely achieved by resecting the tissue in the noncoronary sinus first to allow the proximal stump to open sufficiently to view the other sinuses accurately.(34)



C.4 The proximal aortic stump opens very well after adequate resection. Note the proximity of the coronary orifices to the intended reconstruction suture line.(34)

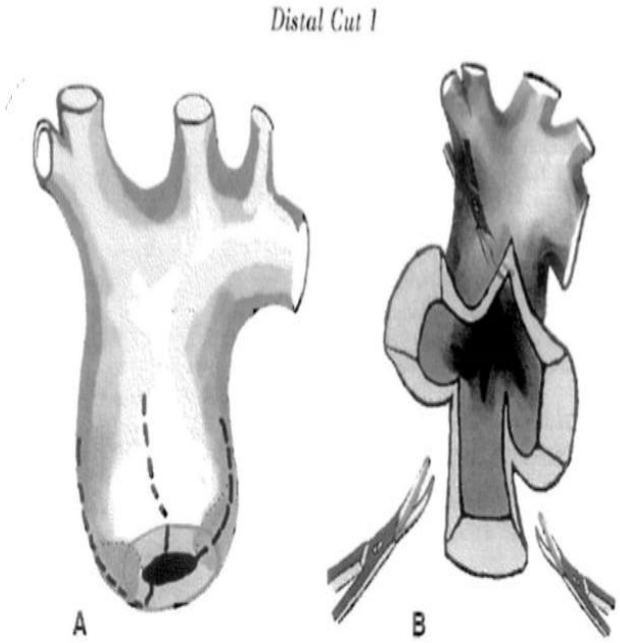


C.5 Reconstruction should start near the left coronary orifice so that this can be well seen as the anastomosis proceeds. Sutures will of necessity be on the verge of the left coronary orifice. The suture line should finish away from the right coronary orifice for the same reason.

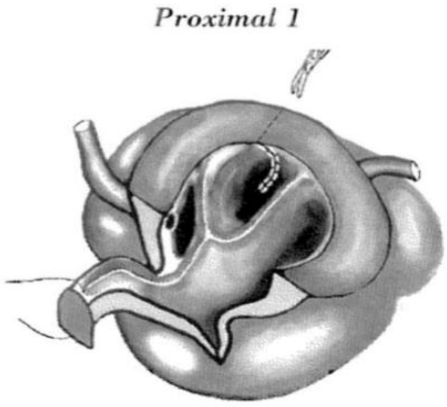
C.6 The mobilization of the arch branches allows approximation without tension and tissue resection allows a normal sized reconstructed sinotubular junction with sustained relief of supravalvular aortic stenosis.

D. Modified Direct Anastomosis Technique(34)

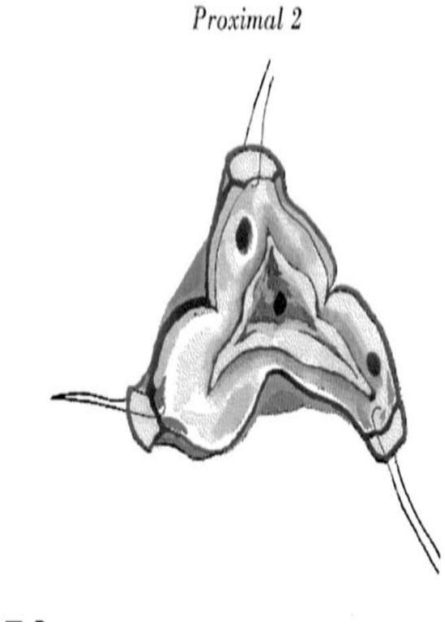
This technique also employs autologous aortic reconstruction of the sinuses of Valsalva. Rather than direct anastomosis, however, the ascending aorta is spatulated in such a fashion as to interdigitate with the incisions in the three sinuses of Valsalva as originally described by Myers and coworkers.



D.1 The three lines of incision should extend well into the distal ascending aorta and correspond to the length of the incisions into the aortic sinuses and be positioned so that there are 120 degrees to these sinus incisions. This allows the three tongues of tissue cut to advance into the corresponding sinuses.

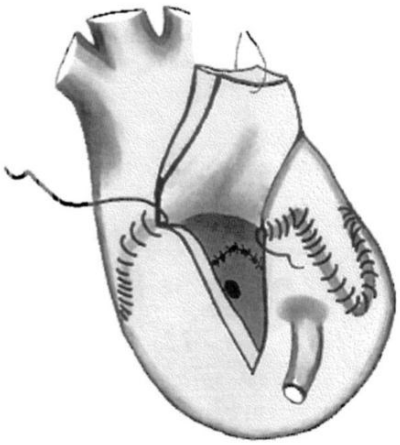


D.2 The first incision into the proximal stump should be into the noncoronary sinus so that subsequent cuts can more fully appreciate the positioning of the left and right coronary arteries within their respective sinuses.



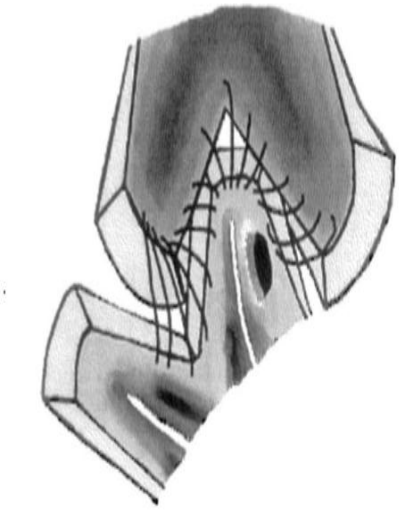
D.3 The proximal stump opens completely after these incisions and the normal nature of the aortic valve leaflets is easily appreciated. The closeness of the coronary orifices to the lines of incision is often exquisite.

Anastomosis 2

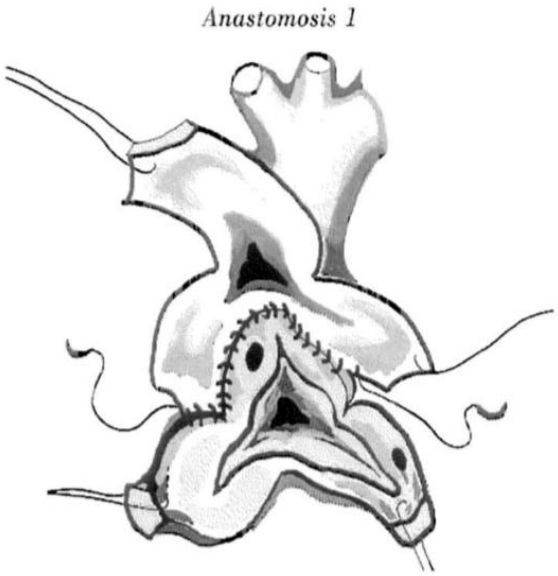


D.4 The three tongues of tissue that advance into the proximal incisions require careful near and far suturing to maintain the accuracy of the three tongues positioning around the circumference to the proximal stump.

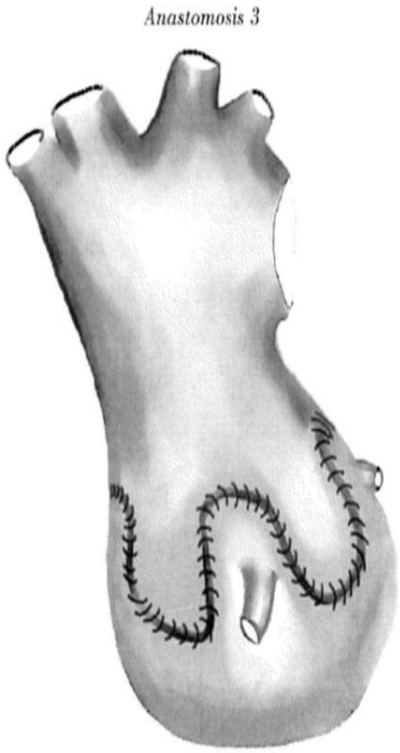
Near and Far Suturing



D.5 Joining of the aorta should begin posteriorly and proceed to completion anteriorly away from the right coronary orifice. The thickened nature of the tissue is no impediment to an accurate suture line.



D.6 Tissue advancement into the sinuses is very adequate.



D.7 The completed long suture line successfully enlarges the sino tulular junction without tissue resection and provides sustained relief of supravalvular aortic stenosis with preservation of aortic growth and normal valve function.

E.Modified simple sliding aortoplasty(28)

The aorta was transected obliquely several millimeters distal to the point of stenosis.

This was to allow resection of the narrowed segment under direct visualization from the luminal side as the proximal resection must be just above the commissure between the left and right coronary cusps. As a result, the resected segment may be around 1 cm in length. An incision was then made into the noncoronary sinus of the proximal aorta, and a counter incision into the lesser curvature of the ascending aorta as shown in Fig E1 was made so as to create the appropriate diameter of the new sinotubular junction. The tethered fibrous tissue was then excised, and the thickened commissural tissue was mobilized. The proximal and distal aorta was then anastomosed directly with 5-0 or 6-0 Prolene running suture (Ethicon, Somerville, NJ) as is routine in arterial switch operation (Fig E2). Absorbable sutures or an interrupted suture technique may be used in anastomosis.

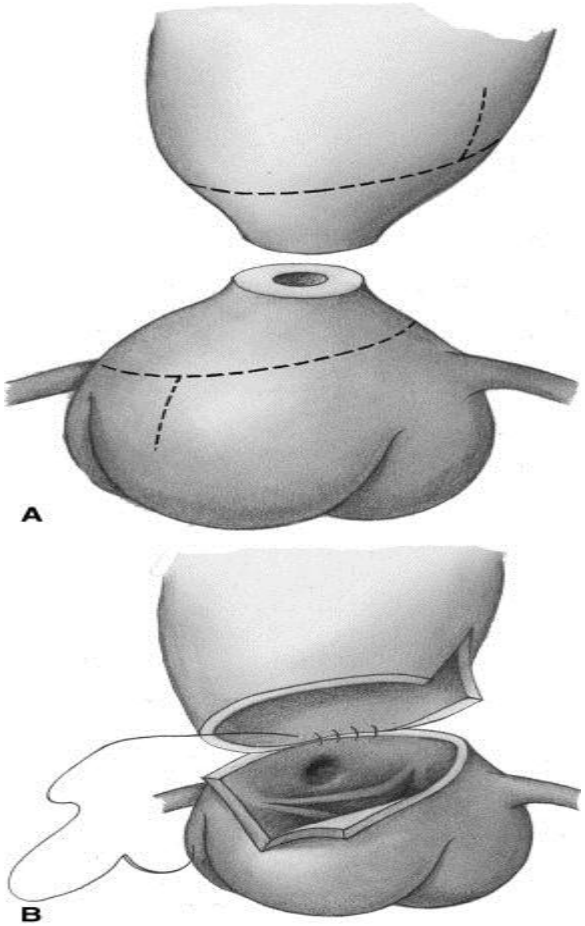


Fig E

(1) Dashed line depicts the incision into the noncoronary sinus of the proximal aorta and another one into the lesser curvature of the ascending aorta. (2) The proximal and distal aorta is then anastomosed directly with 5-0 or 6-0 Prolene running suture (Ethicon, Somerville, NJ).

This technique is easier to perform, more timesaving, and has the advantage of allowing for aortic growth in children.

OBSERVATIONS AND RESULTS

OBSERVATIONS AND RESULTS

AGE:

In our study, 14 patients who underwent surgery for SVAS were analyzed.

The age of presentation ranged from 1 year to 34 years respectively with the mean age of 15 years.

SEX:

There were 8 males and 6 female patients in our study.

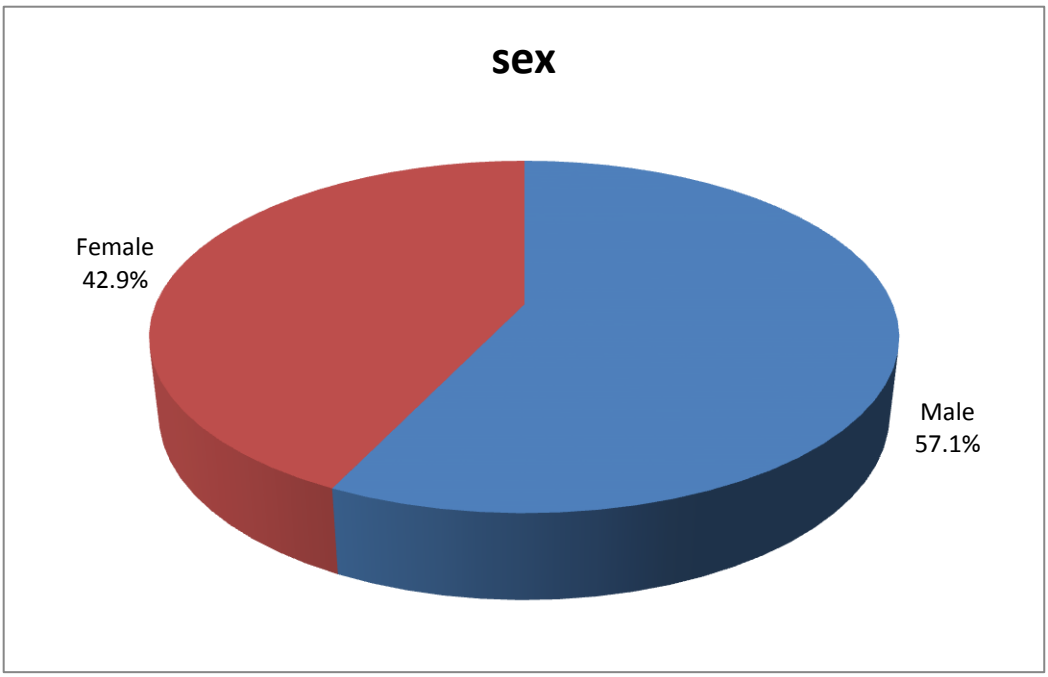


Fig. 1 Sex distribution

Coexisting cardiovascular anomalies:

9 out of 14 patients (64%) did not have any coexisting cardiovascular anomaly. However 1 patient had severe AR with anterior leaflet motion, 1 patient had biscuspid aortic valve, 1 patient had moderate AR moderate MR and RPA stenosis and 1 patient had subpulmonic VSD, PS and RPA origin stenosis.

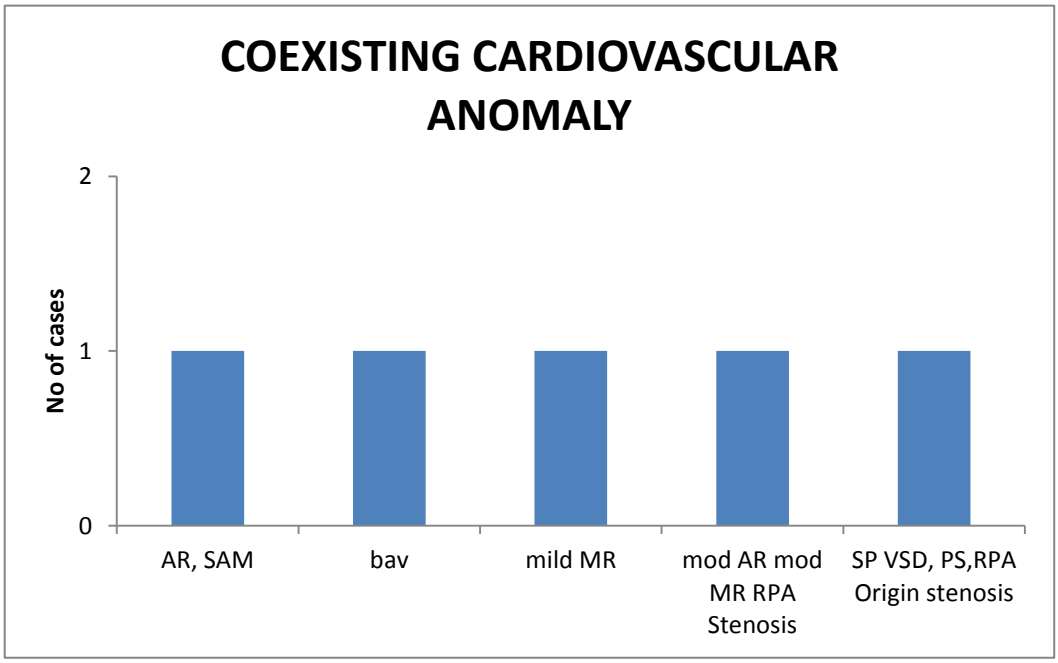


Fig.2 Coexisting cardiovascular anomaly

Concomitant Procedure performed along with SVAS repair:

Out of 14 patients, 11 patients underwent exclusive svas repair procedure (78.6%) however 3 patients underwent concomitant procedures along with svas repair. The concomitant procedures involved AVR, RPA plasty, Pulmonary valve commisurotomy, Infandibular resection, VSD closure and SAM excision.

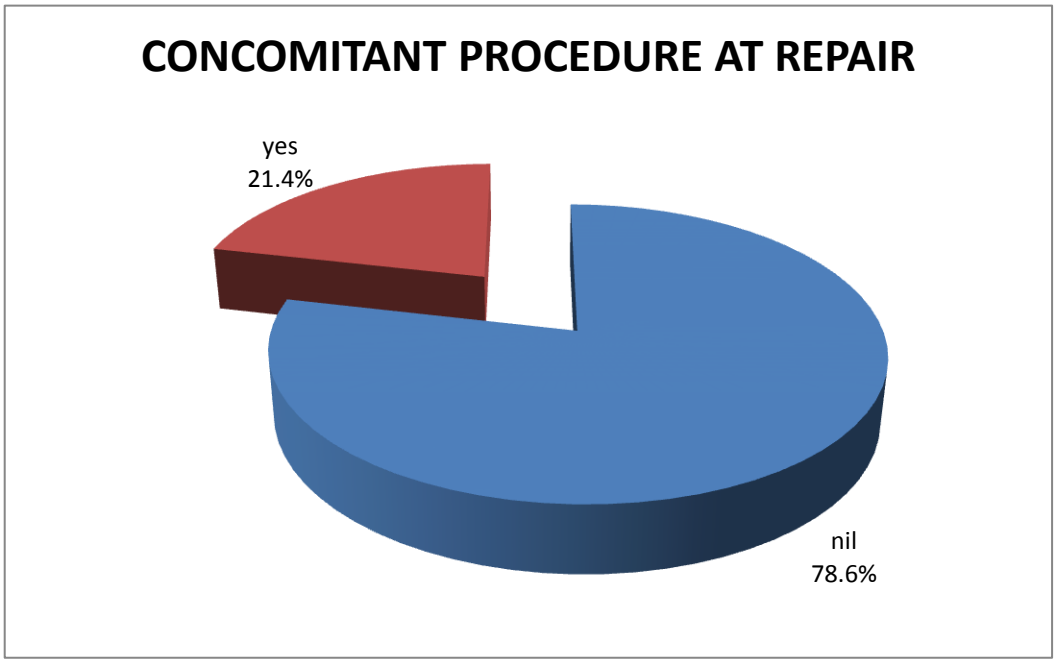
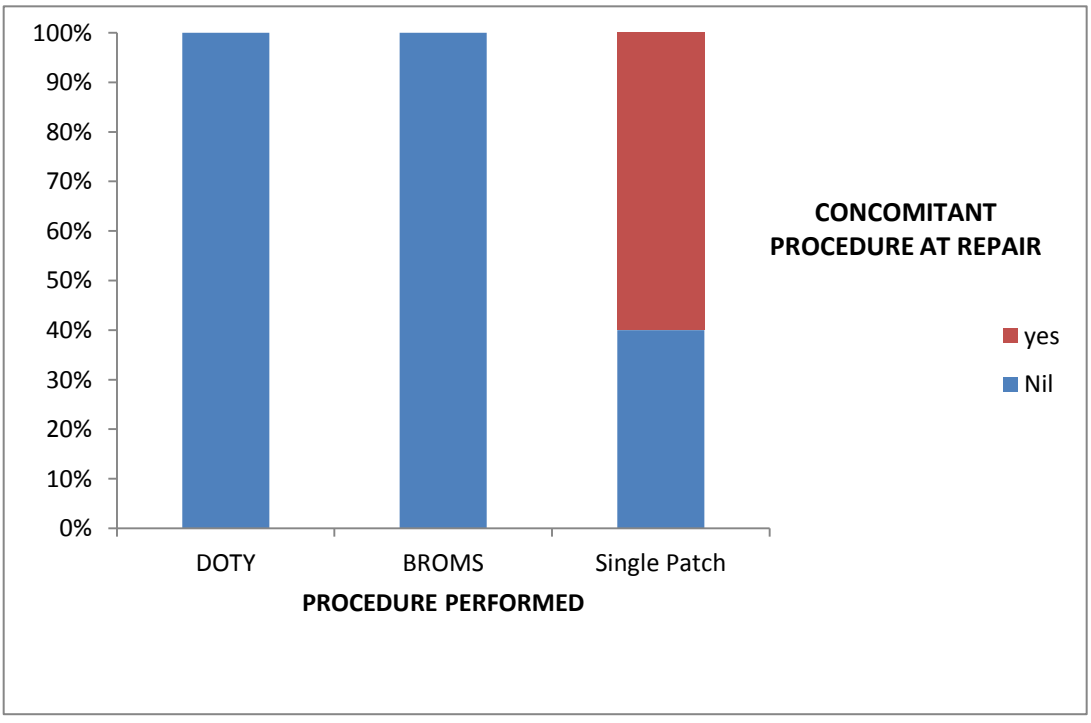


Fig.3 Concomitant procedure at repair

However, all 3 patients who underwent concomitant procedures underwent single patch aortoplasty for SVAS.

CONCOMITANT PROCEDURE AT REPAIR	PROCEDURE PERFORMED						**Total**	
	DOTY		BROMS		Single Patch			
	N	**%**	**N**	**%**	**N**	**%**	**N**	**%**
Nil	6	100	3	100	2	40	11	79
Yes	0	0	0	0	3	60	3	21
Total	6	100	3	100	5	100	14	100



CPB time and Cross clamp time:

Mean CPB time for patients who underwent concomitant procedures at SVAS repair was 155.3 mins and for whom it was not done was 106.5 mins.

CONCOMITANT PROCEDURE AT REPAIR	N	CPB TIME In minutes		t	p
		Mean	sd		
Done	3	155.3	52.3	1.955	.074
Not done	11	106.5	34.8		

Mean clamp time for patients who underwent concomitant procedures at SVAS repair was 104.3 mins and for whom it was not done was 68.5 mins.

CONCOMITANT PROCEDURE AT REPAIR	N	CLAMP TIME in minutes		t	p
		Mean	sd		
Done	3	104.3	27.0	1.939	.076
Not done	11	68.5	28.6		

Williams syndrome:

William syndrome was present in 50 % of the patients with SVAS.

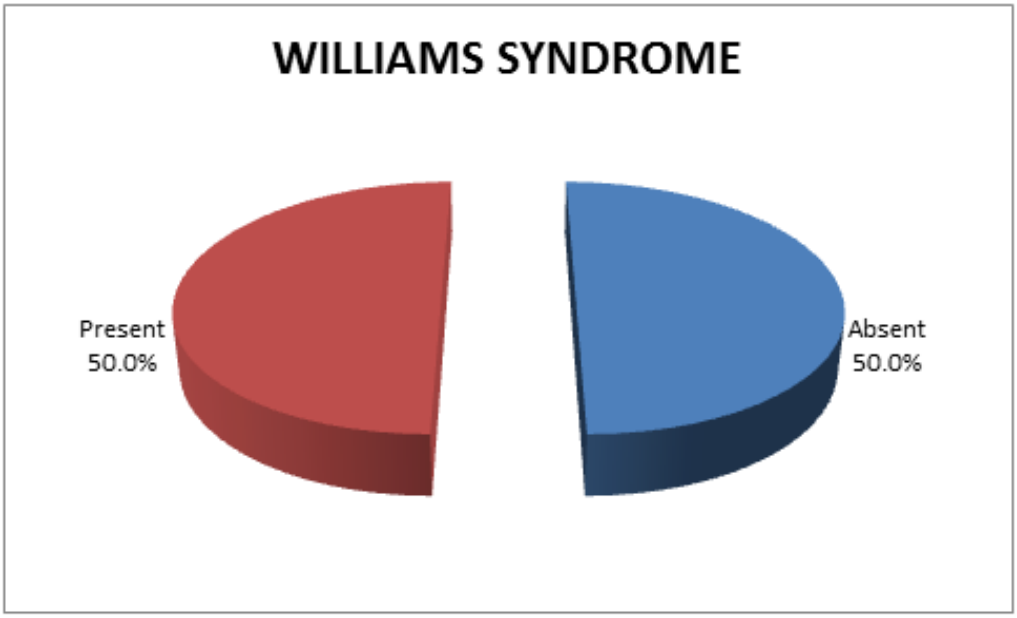
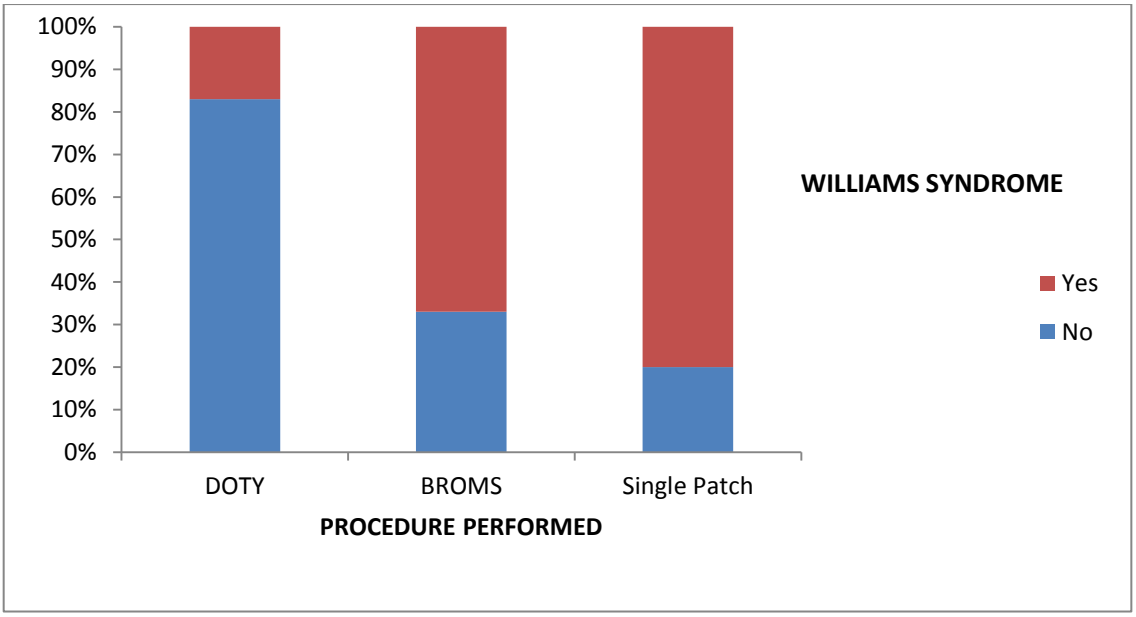


Fig. 4 William syndrome

Out of 7 patients who had Williams syndrome only 1 patient had diffuse disease whereas 6 patients(85.7%) had localized stenosis. So our study shows these two variables are not statistically related.(p>0.05)

DIFFUSE DISEASE	WILLIAMS SYNDROME				**Total**	
	No		Yes			
	N	**%**	**N**	**%**	**N**	**%**
No	6	85.7	6	85.7	12	85.7
Yes	1	14.3	1	14.3	2	14.3
Total	7	100.0	7	100.0	14	100.0

p=1.000



WILLIAMS SYNDROME	PROCEDURE PERFORMED						**Total**	
	DOTY		BROMS		Single Patch			
	N	**%**	**N**	**%**	**N**	**%**	**N**	**%**
No	5	83	1	33	1	20	7	50
Yes	1	17	2	67	4	80	7	50
Total	6	100	3	100	5	100	14	100

$\chi^2 = 4.800$ *df* $= 2$ $p = 0.091$

Out of 7 patients who had Williams disease,4 patients underwent single patch aortoplasty,2 patients underwent Broms repair and 1 patient underwent Doty repair which is not statistically significant as p value is

> 0.05.Choice of the procedure is not dependent on presence of Williams disease.

Diffuse Disease:

Diffuse disease was present in only 2 (14.3%) out of 14 patients.

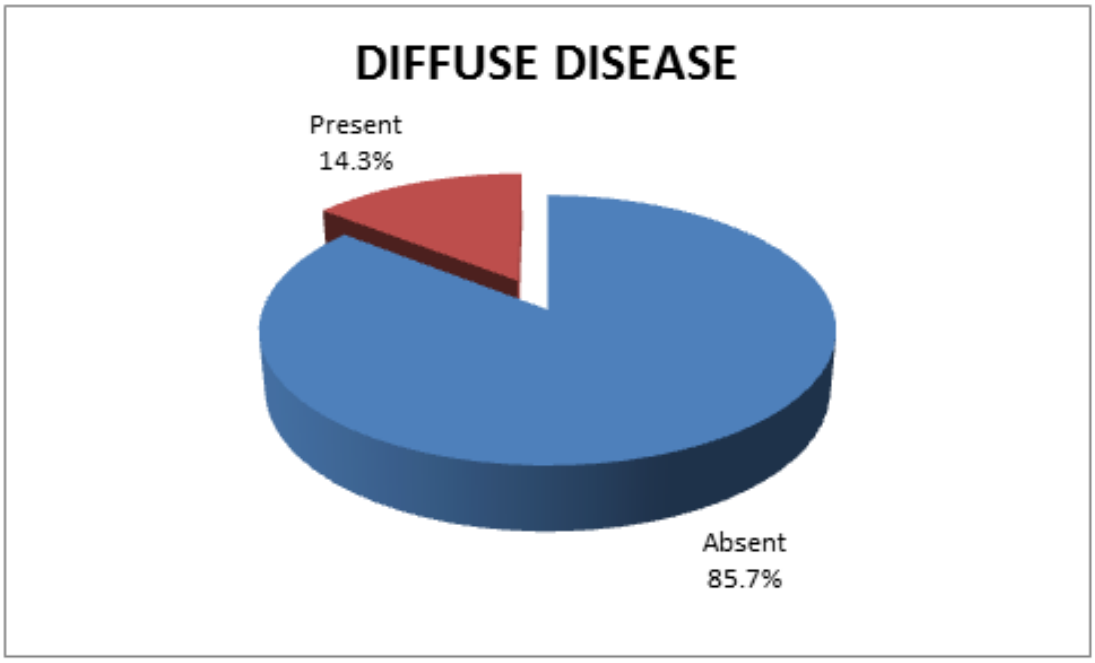


Fig. 5 Diffuse aortic disease

Ascending aorta Dimensions:

With the help of CT Aortogram,Ratio of Diameter of Narrowest part of Ascending aorta to Diameter of distal part ascending aorta at innominate artery origin was calculated with CT aortogram. All 14 Patients had ratio between 0.8 to 1.30 with mean ratio of 1.055 which denotes no dialatation or stenosis has developed around the patch.

Bicuspid Aortic valve:

Bicuspid aortic valve was present in only 1 out of the 14 patients in this study.

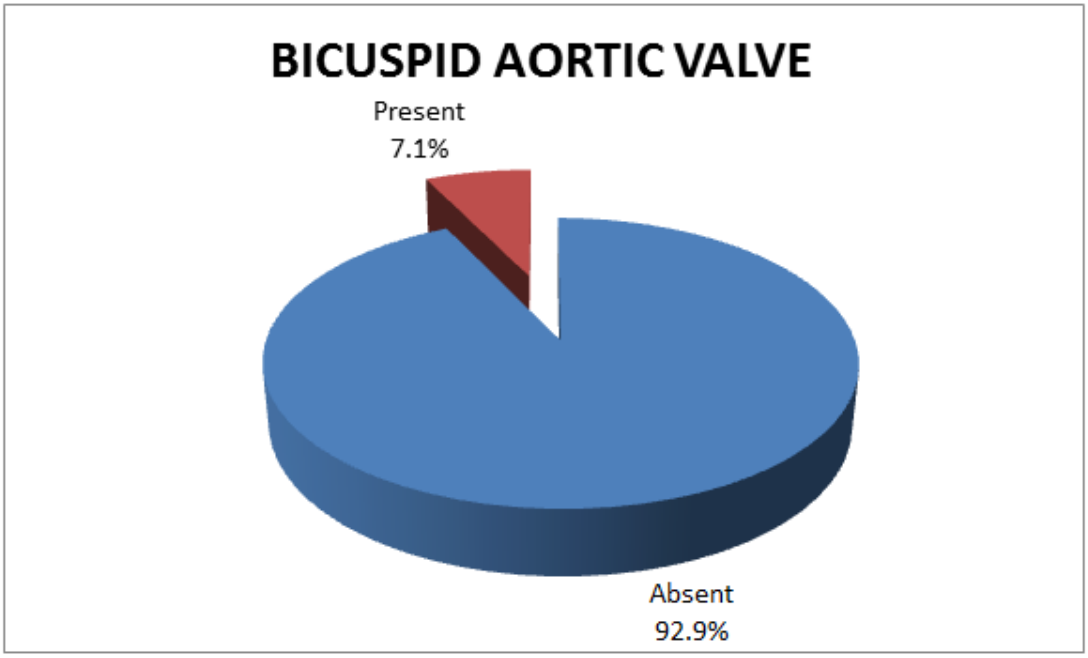
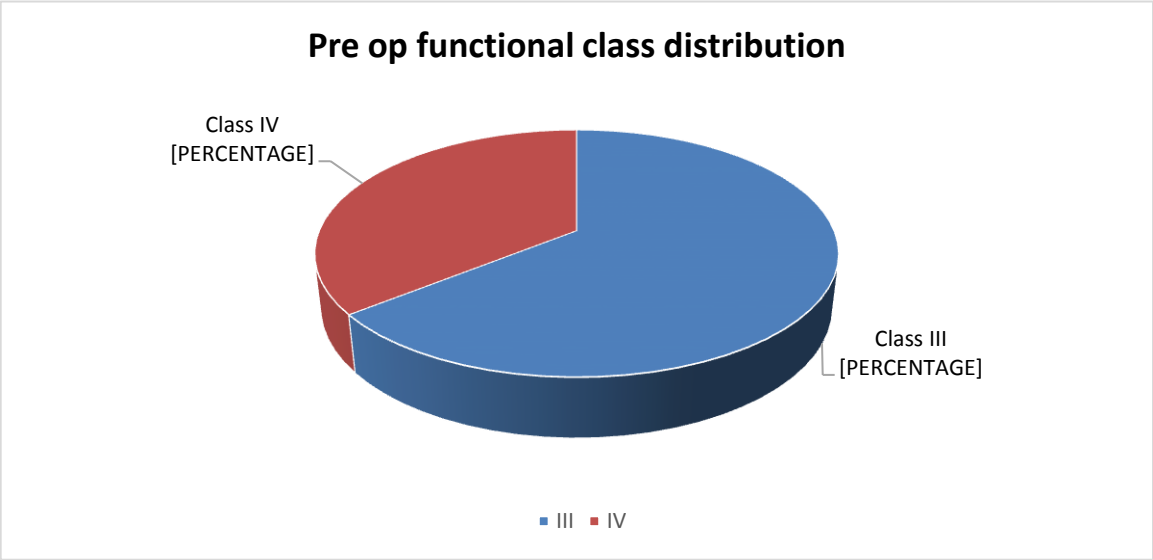


Fig.6 Bicuspid aortic valve

NYHA Functional class:



Pre-operatively all the patients were in NYHA functional class III and IV.9 patients were in class III and 5 were in class IV.After SVAS surgery all patients showed improvement in functional class in early and late follow up.Post operatively ,10 patients had NYHA Functional class I and 4 had class II

	Functional class		Post OP		Total
			I	II	
DOTY	Pre Op	III	4 (80%)	1(20%)	5
		IV	1(100%)	0(0%)	1
		Total	5(83.3%)	1(16.7%)	6
BROMS	Pre Op	III	1 (50%)	1(50%)	2
		IV	1(100%)	0(0%)	1
		Total	2(66.7%)	1(33.3%)	3
Single Patch	Pre Op	III	2 (100%)	0(0%)	2
		IV	1(33.3%)	2(66.7%)	3
		Total	3(60%)	2(40%)	5

Out of 5 patients who underwent Doty procedure 4 patients were in functional class III who improved to class I and 1 patient improved to class II.

Out of 2 patients who underwent Broms repair 1 patient improved to class I and the other to class II.Out of 3 patients who underwent single patch repair 2 patients improved to class I and one to class II.However there is no statistical relation in improvement in functional class and type of procedure.

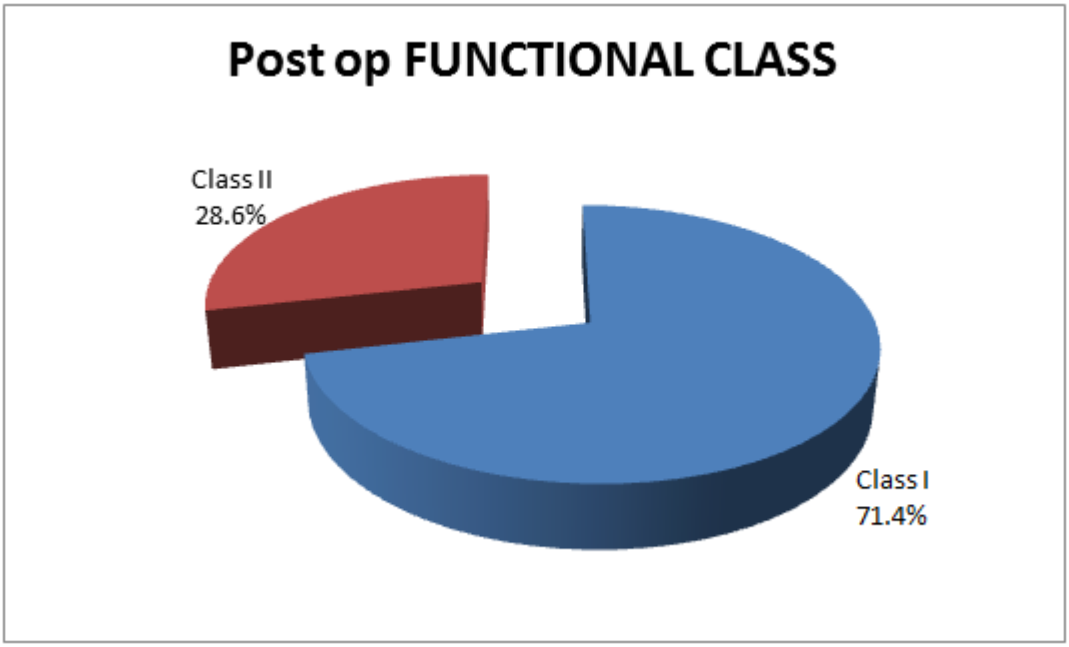


Fig 7. NYHA Functional class

Aortic Regurgitation:

Aortic regurgitation was present in 6(42.9 %) patients in late follow up who underwent SVAS surgery. Out of these 6, only 2 patients had 2 + AR whereas 4 patients had 1 + AR.

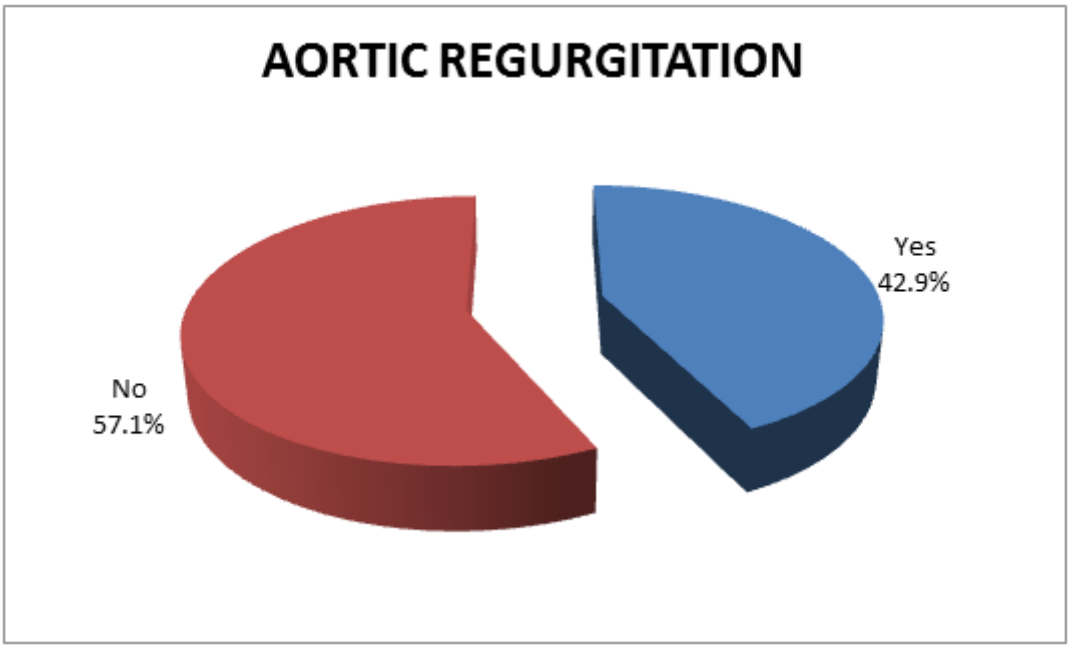
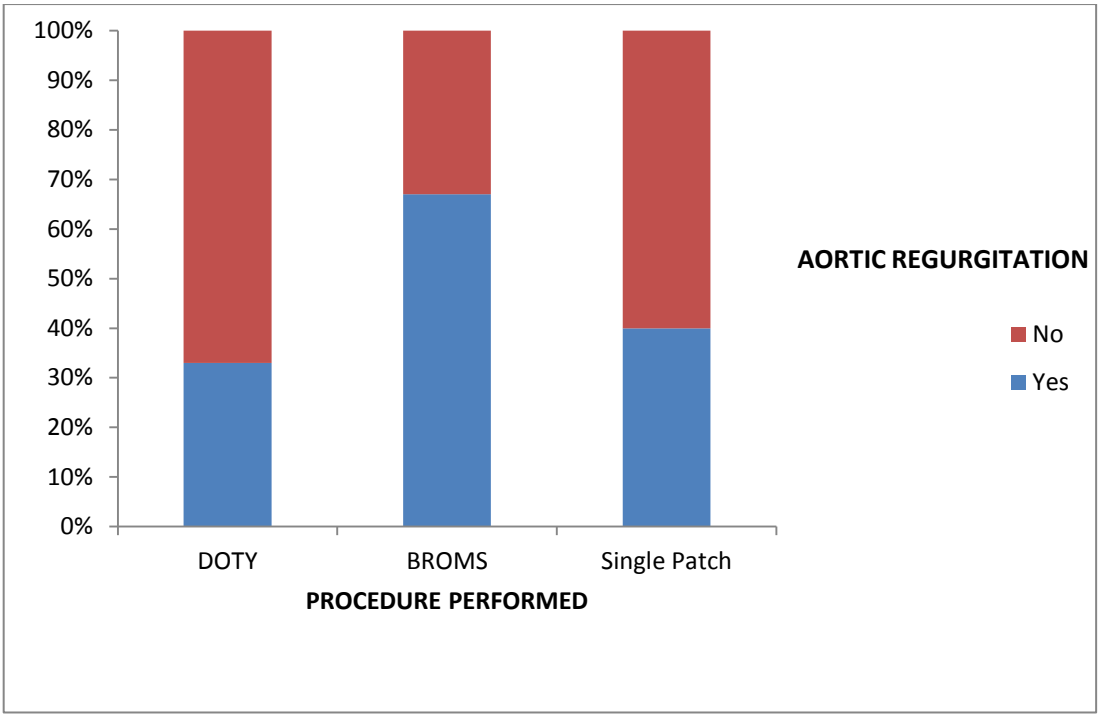


Fig. 8 Aortic regurgitation in patients

Out of these 6 patients, Doty, Broms and Single patch repair was done in 2 patients each. Hence development of AR postoperatively is not related to the type of procedures performed for SVAS (p value > 0.05). However 33% of patients who underwent Doty procedure developed AR. For Broms and Single patch repair it was 67% and 40% respectively.



Procedure Performed for SVAS repair:

Out of 14 patients, 6 patients(42.9%) underwent Dotys aortoplasty, 5 patients(35.7%) underwent Single patch aortoplasty while 3 patients(21.4%) underwent Broms aortoplasty for SVAS repair.

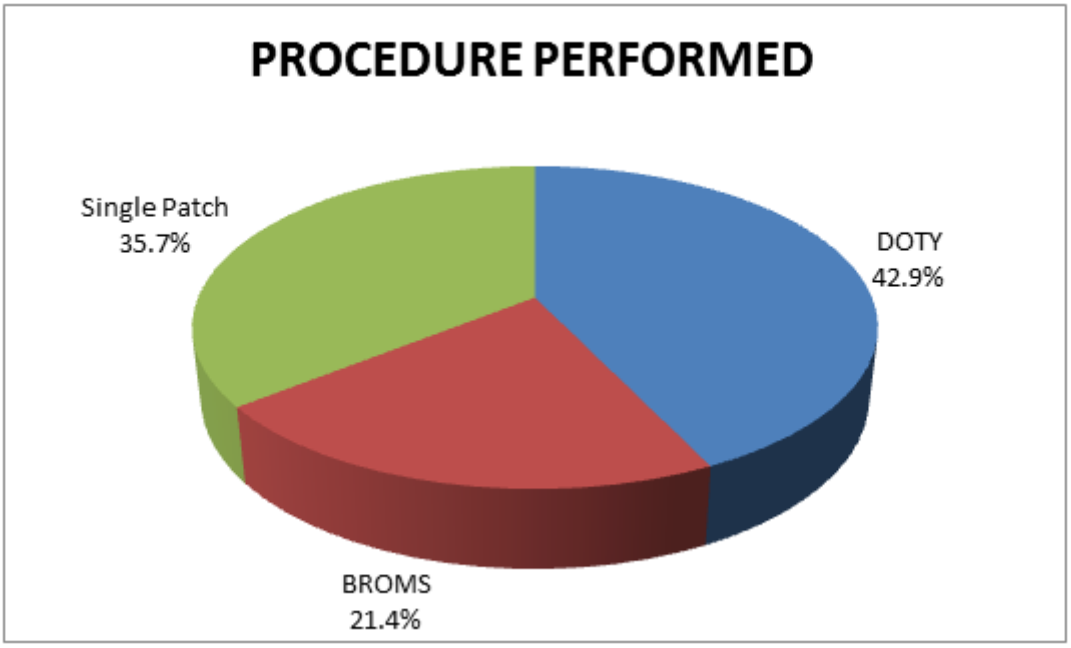


Fig. 9 Surgical procedures performed

In this study, CPB time of these surgical procedures – Dotys, Broms and Single patch aortoplasty was analysed.

The mean CPB time was 90.3 minutes for Dotys procedure, 128.4 minutes for single patch aortoplasy while it was 151.3 minutes for Broms procedure.

PROCEDURE PERFORMED	N	CPB TIME In minutes		p
		Mean	sd	
DOTY	6	90.3	25.2	.083
BROMS	3	151.3	17.6	
Single Patch	5	128.4	52.4	
Total	14	117.0	42.3	

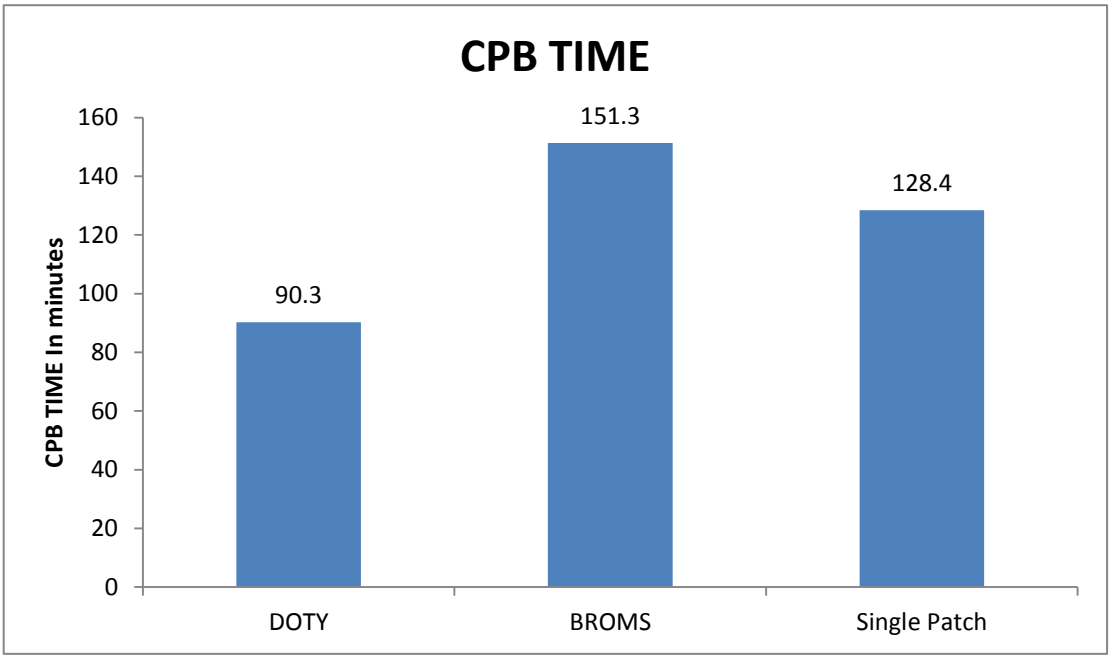


Fig.10 CPB time in different surgical procedures

The mean clamp time was 58.5 minutes for Dotys procedure, 80.4 minutes for single patch aortoplasty and 104.7 minutes for Broms procedure.

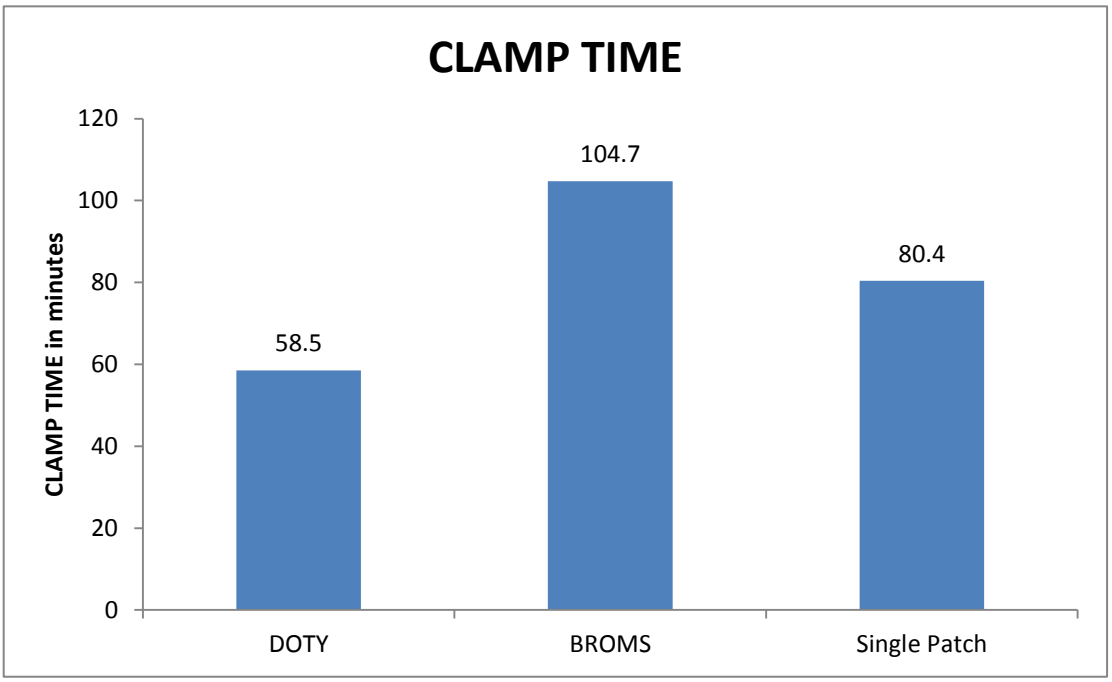


Fig.11 Clamp time in surgical procedures

Though Doty procedure showed decreased CPB time and clamp time in our study it is not statistically significant($p>0.05$).

PROCEDURE PERFORMED	N	CLAMP TIME in minutes		p
		Mean	sd	
DOTY	6	58.5	16.9	.095
BROMS	3	104.7	19.6	
Single Patch	5	80.4	38.9	
Total	14	76.2	31.2	

Hospital Stay:

Hospital stay was analysed with respect to procedure performed.For Doty procedure mean hospital stay was 10 days, For Broms repair it was 9.7 days whereas for single patch repair it was 11.4 days.However our study showed no statistical significance between hospital stay and procedure. .performed.(P>0.05).Hospital stay for single patch repair was more can be explained as 3patients who underwent single patch repair had undergone concomitant procedures for other heart lesion thereby increasing hospital stay.

For SVAS surgery mean hospital stay was 10.4 days which included all the three procedures.

PROCEDURE PERFORMED	N	HOSPITAL STAY (DAYS)		p
		Mean	sd	
DOTY	6	10.0	2.8	.693
BROMS	3	9.7	0.6	
Single Patch	5	11.4	4.2	
Total	14	10.4	3.0	

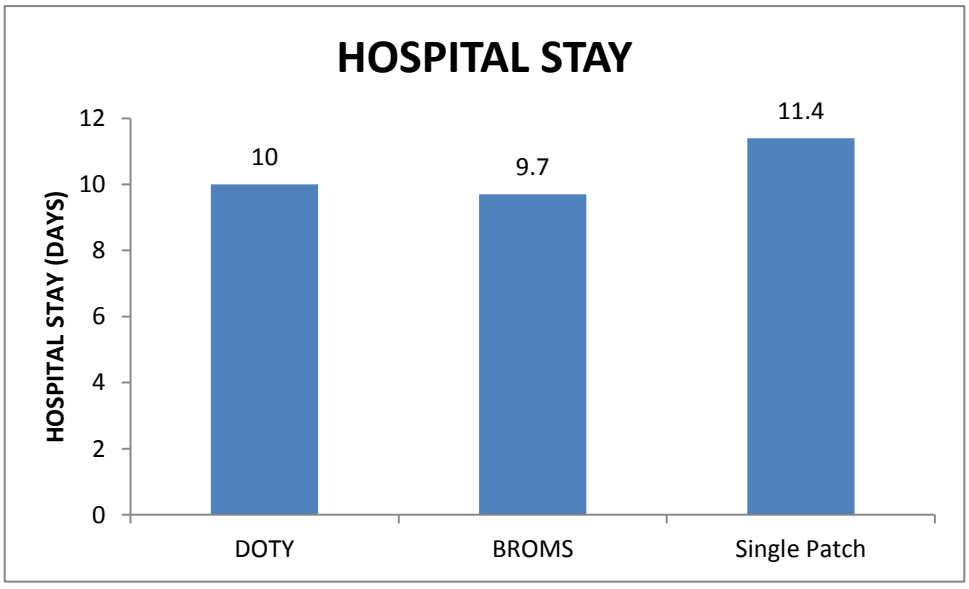
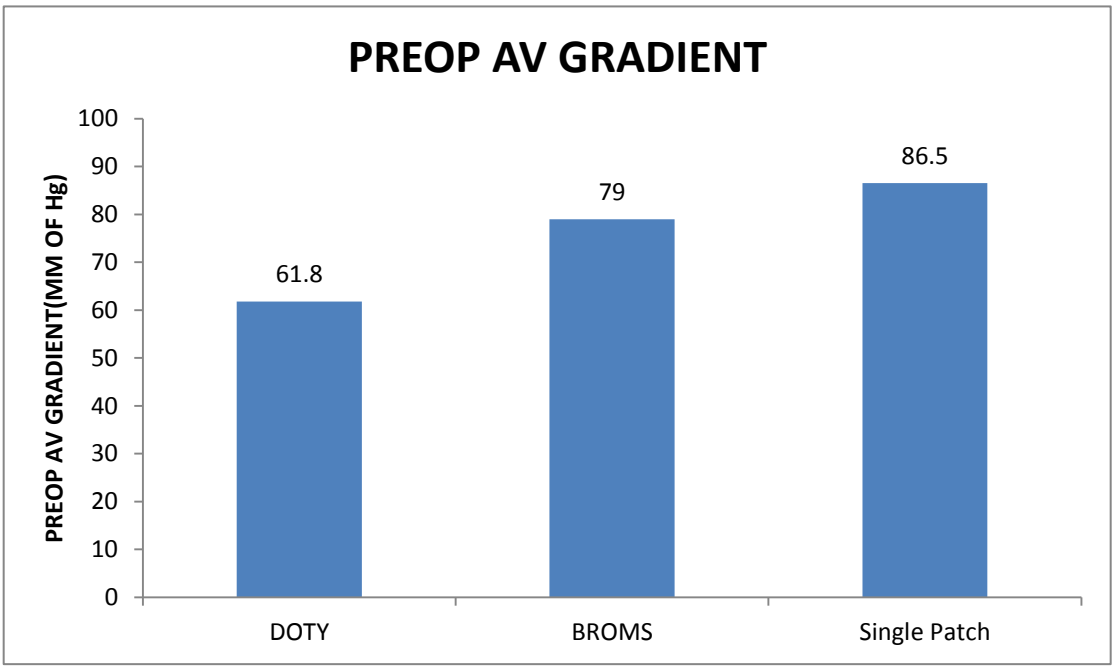


Fig.12 Hospital stay in patients following surgery

Aortic Gradients(Pre op,Early Post-op & Late Post-op):

The mean Pre-op AV gradient in patients with svas who underwent surgery was 73.4 with minimum mean gradient of 40 mm Hg and maximum of 100 mm Hg. However relation between the choice of procedure performed and preop gradient is not statistically significant in our study.(p>0.05)

PROCEDURE PERFORMED	N	PREOP AV GRADIENT(MM OF Hg)		p
		Mean	sd	
DOTY	6	61.8	20.2	.095
BROMS	3	79.0	8.7	
Single Patch	5	86.5	12.1	
Total	14	73.4	18.7	

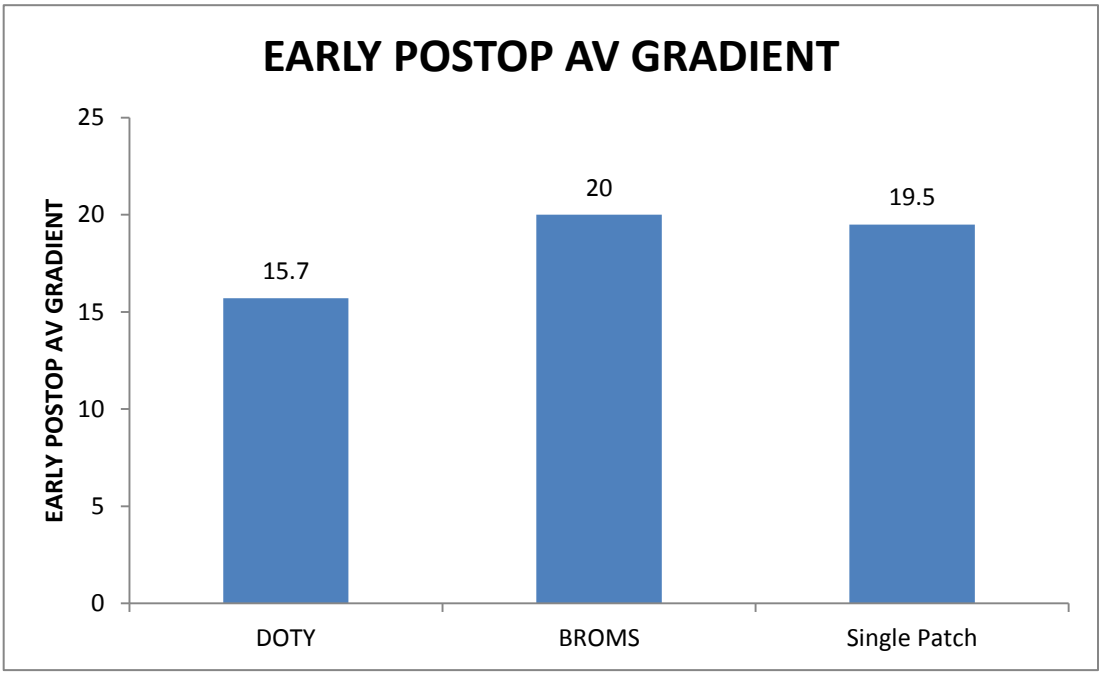


Mean Post op AV gradient in early and late followup was 17.8 mm of Hg and 15.7 mm of Hg respectively.

Mean early postop AV gradients (in mm of Hg) for Doty was 15.67, for Broms was 20 and for single patch repair was 19.5.

Early Post op AV gradient which was observed within 3 months after surgery showed considerable improvement as compared to Pre op gradients, however our study did not show any significant difference in improvement in early post op gradients depending on the procedures performed.

PROCEDURE PERFORMED	N	EARLY POSTOP AV GRADIENT		p
		Mean	sd	
DOTY	6	15.7	5.5	.586
BROMS	3	20.0	9.5	
Single Patch	5	19.5	6.8	
Total	14	17.8	6.6	

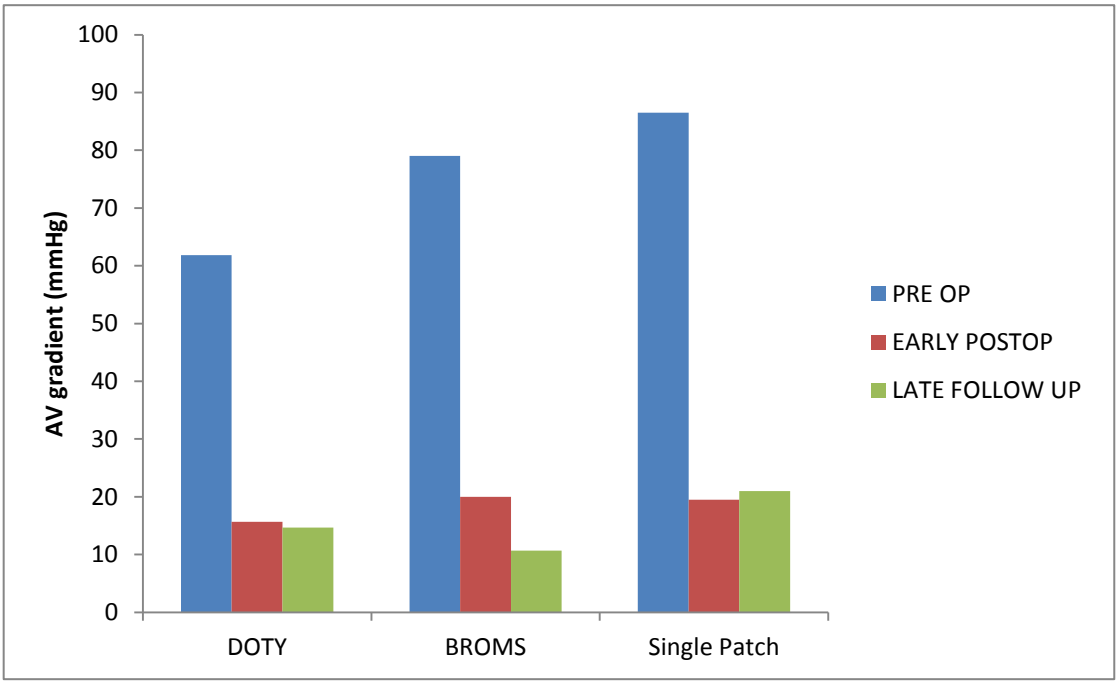


Mean late follow up postop AV gradients (in mm of Hg) for Doty was 14.7, for Broms was 10.7 and for single patch repair was 21.On Late follow up (considered atleast 3 years following surgery), the Post op AV gradient showed no statistical difference as compared to early post op gradients. However our study did not show any significant difference in late post op gradients depending on the procedures performed.

PROCEDURE PERFORMED	N	LATE FOLLOW UP GRADIENT		p
		Mean	sd	
DOTY	6	14.7	6.0	.126
BROMS	3	10.7	2.9	
Single Patch	5	21.0	7.8	
Total	14	15.7	6.9	

AV Gradient (mm of Hg)	DOTY		BROMS		Single Patch	
	mean	sd	mean	sd	mean	sd
PRE OP	61.83	20.22	79.00	8.72	86.50	12.07
EARLY POSTOP	15.67	5.50	20.00	9.54	19.50	6.81
LATE FOLLOW UP	14.67	6.02	10.67	2.89	21.00	7.79
Comparison between PRE OP and early POST OP - p	.001		.016		.001	
Comparison between PRE OP and Late POST OP - p	.001		.005		.004	
Comparison between Early POST OP and Late POST OP - p	.041		.148		.817	

When the gradients in preop and early postop period were compared, our study showed significant difference(p value <0.05). Similarly when the gradients in the preop and late postop period were compared, our study showed significant difference. However when early and late postop gradients were compared, there was no statistically significant difference.

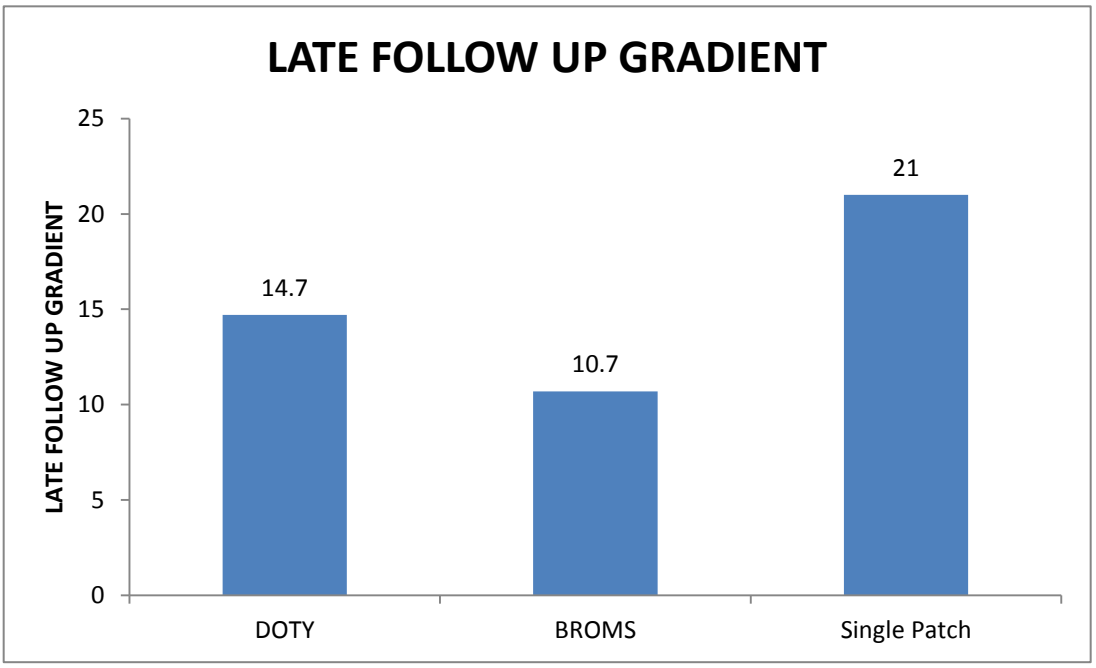


PROCEDURE PERFORMED	N	% reduction in AV gradient From Pre OP to Early Post OP		p
		Mean	sd	
DOTY	6	74.6	3.9	.753
BROMS	3	74.5	11.7	
Single Patch	5	77.7	6.3	
Total	14	75.5	6.4	

In our study, there was no significant difference in % reduction in AV gradient from preop to early postop period for these 3 procedures for svas surgery. Therefore the percentage reduction in AV gradients from preop to early postop is independent of the type of surgery performed.

PROCEDURE PERFORMED	N	% reduction in AV gradient From Pre OP to Late Post OP		p
		Mean	sd	
DOTY	6	76.6	4.7	.148
BROMS	3	86.5	3.2	
Single Patch	5	74.9	12.1	
Total	14	78.4	8.3	

In our study, there was no significant difference in % reduction in AV gradient from preop to late postop period for these 3 procedures for svas surgery. Therefore the percentage reduction in AV gradients from preop to late postop is independent of the type of surgery performed.



DISCUSSION

DISCUSSION

Our study consisted of 14 patients with SVAS who underwent surgical repair with either Doty,Broms or single patch repair from year 2006 to year 2015.

There were 8 males (57.17%) and 6 female patients in our study who had SVAS.Sampayo F et al studied sex distribution of congenital cardiopathies and reported 71 % of male preponderance in SVAS.(35)

The age of presentation ranged from 1 year to 34 years with the mean age of 15 years. Brenda Fabiola Cruz-Castañeda et al. studied 9 patients of SVAS and found out that all patients were between 5yrs to 14 yrs.(36) Between August 1956 and May 2009,Salil V. Deo et al. analysed 78 patients of SVAS. Median age was 10.4 years with minimum age of 16 days and maximum of 55.2 years.(37) The age of presentation ranged from 1 year to 34 years with the mean age of 15 years. Brenda Fabiola Cruz-Castañeda et al. studied 9 patients of SVAS and found out that all patients were between 5yrs to 14 yrs.(36) Between August 1956 and May 2009,Salil V. Deo et al. analysed 78 patients of SVAS. Median age was 10.4 years with minimum age of 16 days and maximum of 55.2 years.(37)

9 out of 14 patients (64%) did not have any coexisting cardiovascular anomaly. However 1 patient had severe AR with systolic anterior leaflet motion, 1 patient had biscuspid aortic valve, 1 patient had moderate AR, moderate MR and RPA stenosis and 1 patient had subpulmonic VSD, PS and RPA origin stenosis.

Salil V .Deo et al in his study also analysed coexisting cardiovascular anomalies with SVAS. They found out 29% patients had Aortic valve

stenosis.Aortic regurgitation was present in 87% patients.3 patients had sub aortic stenosis.14 patients (18%) had coronary artery involvement.

In our study we did not get any patient with coronary artery involvement.

Ralph E Delius et al. in his study with 47 patients analysed correlation of bicuspid aortic valve and SVAS.Sixteen patients (34%) had a bicuspid aortic valve that was competent and nonobstructive, and 31 (66%) had a tricuspid aortic valve.we got only 1 patient with bicuspid aortic valve along with supravalvular aortic stenosis.(38)

Sharma BK et al. studied 73 patients with SVAS from 1960 to 1989 and found out association of Williams syndrome, SVAS and peripheral pulmonary artery stenosis.(39)

William syndrome was present in 50 % of the patients with SVAS.

SVAS was the cardiovascular lesion first reported by Williams et al (40) and has been found to be the most common cardiovascular abnormality associated with williams syndrome(41) The incidence of SVAS has been reported to be 45% to 75% in patients with WS.(42)

Two types of SVAS are typically seen in patients with WS: a discrete, hourglass narrowing at the sinotubular junction or a diffuse, long-segment stenosis of the ascending aorta(43)

In our study, Out of 7 patients who had Williams syndrome only 1 patient had diffuse disease whereas 6 patients(85.7%) had localized stenosis. So our study shows these two variables are not statistically related.($p>0.05$)however Discrete type is more common in williams disease.

This finding is similar to study done by Zalzstein E et al., Stamm C et al and Hickey EJ et al. who opined that the hourglass type of SVAS is the more common of the two,(31,44) occurring in 75% of children.(45)

In our study,Pre-operatively all the patients were in NYHA functional class III and IV.9 patients(64%) were in class III and 5(36%) were in class IV.After SVAS surgery all patients showed improvement in functional class in early and late follow up.

Post operatively ,10 patients had NYHA Functional class I and 4 had class II.However in our study 100 % survival was observed.

Good surgical outcome of congenital SVAS can be achieved irrespective of the type of procedure performed in patients with both localized and diffuse SVAS.

Brown JW et al. studied 101 patients of SVAS from 1962 to 2000 and found out 11 % were in NYHA class I,55 % in class II,28% in class III and 7% in class IV. Postoperatively, there were 72 patients (73%) in NYHA functional class I and 26 (27%) in class II. Overall survival including operative mortality was 98% at 10 years, 97% at 20 and at 30 years.(46)

Findings of Brown JW et al. is similar to our findings.

Sachin Talwar et al. studied 13 patients with SVAS and their surgical outcome and found out that post SVAS surgery all patients except one converted back to NYHA class I group.(47)

The mean Pre-op AV gradient in patients with svas who underwent surgery was 73.4 with minimum mean gradient of 40 mm Hg and maximum of 100 mm Hg. However relation between the choice of procedure

performed and preop gradient is not statistically significant in our study.($p>0.05$)

Deo S V et al. study in 78 patients who underwent surgical correction of SVAS had shown mean preoperative gradient of 57.2±21.9 mm Hg .(37)

Delius RE et al. in their study of long-term follow-up of extended aortoplasty for supravalvular aortic stenosis also found mean preoperative gradient of 90mm of Hg range being 50 to 150 mm of Hg.(9)

Mean Post op AV gradient in early and late followup was 17.8 mm of Hg and 15.7 mm of Hg respectively.

Brenda et al. studied 9 patients with SVAS and compared transaortic pre op gradient and post op gradient. In all patients who survived, postoperative gradients were improved (range, 0–16 mmHg.(36)

Elena Arnáiz et al in their study of Surgery for supravalvular aortic stenosis in 15 patients found out mean post operative gradient of 10 mm Hg.(48)

The above mentioned studies have similar opinion like ours that after SVAS surgery in late followup,aortic gradients tend to remain low.

This finding is contradicted by Sachin Talwar et al. study which was done on 13 patients.They found out after follow-up of 57 months, gradients across the LVOT progressed to 10–130 mmHg (37.3 ±31.6, median 30 mmHg). In seven (58%), the gradients were 30 mmHg or more at 5 years of follow up.(49) More than 50% of these patients developed significant gradients and they opined Recurrent LVOT obstruction is an ongoing issue even after satisfactory initial repair of SVAS which is different from our results.(49)

In our study none of the patients developed significant restenosis or increased AV gradient. However 6 patients developed AR postoperatively which was mild and did not require any intervention.

Other contradictory study is by Delius RE et al. in which long-term follow-up of extended aortoplasty for supravalvular aortic stenosis was observed and they found progressive increase in aortic gradient after extended aortoplasty with time.(9)

Mean early postop AV gradients (in mm of Hg) for Doty was 15.67, for Broms was 20 and for single patch repair was 19.5.

Early Post op AV gradient which was observed within 3 months after surgery showed considerable improvement as compared to Pre op gradients,however our study did not show any significant difference in improvement in early post op gradients depending on the procedures performed.

Mean late follow up postop AV gradients (in mm of Hg) for Doty was 14.7, for Broms was 10.7 and for single patch repair was 21mm of Hg.On Late follow up (considered atleast 3 years following surgery), the Post op AV gradient showed no statistical difference as compared to early post op gradients. However our study did not show any significant difference in late post op gradients depending on the procedures performed.

Metton O et al. in their study Surgical management of supravalvular aortic stenosis: does Brom three-patch technique provide superior results?,analysed 34 patients who underwent SVAS repair with different techniques and opined that there is significant difference in post-op aortic gradients between different SVAS repair techniques.(50)

At last follow-up, left ventricle to aorta peak gradient was 45 +/- 28 mm Hg after one-patch repair, 30 +/- 9 mm Hg after Doty operation, and 11 +/- 18 mm Hg after symmetric Brom procedure. Brom repair was associated with a low incidence of residual obstruction (peak gradient > or = 40 mm Hg) (2 of 22; 9.1%) and moderate aortic insufficiency (1 of 22; 4.5%)

In Our study we got no significant post-op aortic gradient difference between single patch ,Doty and Broms technique.In contrast to our study,

Kaushal S et al. in their study Midterm outcomes in supravalvular aortic stenosis demonstrate the superiority of multisinus aortoplasty opined that Doty and Broms repair are superior to single patch repair in terms of post op aortic gradient.(51)

Hazekamp et al(30) did not find any significant differences in change in valve function, and found the efficacy of reducing the pressure gradient was similar and acceptable with various techniques which is the finding similar to our study.

Sachin Talwar et al. study which was done on 13 patients of SVAS were not able to conclusively define a co-relation between the type of the operation and the severity of the pre-operative gradients to the progression of the gradients on follow-up.This observation is similar to our study.

Hospital stay was analysed with respect to procedure performed.For Doty procedure mean hospital stay was 10 days, For Broms repair it was 9.7 days whereas for single patch repair it was 11.4 days.However our study showed no statistical significance between hospital stay and procedure performed.($P>0.05$).Hospital stay for single patch repair was more can be explained as 3patients who underwent single patch repair had undergone concomitant procedures for other heart lesion thereby increasing hospital

stay. For SVAS surgery mean hospital stay was 10.4 days which included all the three procedures.

Kaushal S et al. in their study ‘Midterm outcomes in supravalvular aortic stenosis demonstrate the superiority of multisinus aortoplasty’ found out that mean postoperative length of stay was 10 +/- 10.6 days which is similar to our study.

CONCLUSION

CONCLUSION

1. Post-operatively aortic gradient reduces significantly irrespective of the type of surgical procedure performed.Single patch technique,Doty repair and Broms repair have similar results in terms of percentage reduction in aortic gradient .

2. Symptom improvement after SVAS surgery is significant irrespective of type of surgical procedure performed.All patients operated for SVAS returned in NYHA functional class I or II.

3. Though Post operative complications are not uncommon,Aortic valve insufficiency is the most common complication.However AR is of mild grade and can be managed conservatively rarely requiring intervention.

4. Prospective randomized multi-institutional trial with larger sample size is required to define the optimal management strategy and surgical outcomes for these patients.

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ANNEXURES

Technical Advisory Committee (Clinical Studies)

SREE CHITRA TIRUNAL INSTITUTE FOR MEDICAL SCIENCES & TECHNOLOGY
THIRUVANANTHAPURAM – 695011, INDIA

TAC Registration No: SCT-/S/2015/417 **Date:** 28.01.2016

Project title: OUTCOME OF SURGERIES IN SUPRAVALVULAR AORTIC STENOSIS

Principal Investigator:
Name: Dr. Abhay Suresh Jain Degree: DNB (general surgery), M.B.B.S Senior Resident, Department of Cardiovascular and Thoracic Surgery, SCTIMST, Trivandrum
Co-Principal Investigator(s)
Name: Dr.Vivek Pillai Degree: M.Ch. (Cardiothoracic surgery), M.S. (General Surgery), M.B.B.S. Associate Professor, Department of Cardiovascular and Thoracic Surgery, SCTIMST
Name: Dr.Jayakumar K, Degree: M.Ch. (Cardiothoracic surgery), M.S.(General Surgery), M.B.B.S. Professor, Department of Cardiovascular and Thoracic Surgery, SCTIMST, Trivandrum.
Name: Dr. Baiju S Dharan Degree: M.Ch. (Cardiothoracic surgery), M.S. (General Surgery), M.B.B.S. Additional Professor, Department of Cardiovascular and Thoracic Surgery , SCTIMST

Members who participated in the TAC meeting on 08/01/2016

Dr. Rupa Sreedhar (Chairperson)
Dr. Thomas Koshy
Dr. Lissy K Krishnan
Dr. Sylaja P.N
Dr. Krishnamoorthy KM
Dr. Biju Soman
Dr. Bejoy Thomas
Dr. Syam. K
Dr. K. Shivakumar (Member Secretary)

Dr. Thomas Koshy, Dr. Syam K, Dr. Bejoy Thomas and Dr. Sylaja.P.N stayed away from the proceedings when the projects in which they are involved (# 406, 415, 410, 413, 424, 426) as investigators were discussed.

Risk Classification of the project (Minimum/ Moderate/ High): Minimum
Requirement of DSMB: No
Recommended members of DSMB: Not applicable

Recommendations of TAC:
Recommended for consideration of IEC.

The PI may note that there can be no additions / alterations in the documents approved by TAC when they are submitted to the IEC.

Signature of the Member Secretary, TAC (Clinical Studies)

Note for IEC
Copy of the investigator's responses to questions/suggestions from TAC is attached (Appendix-1).

श्री चित्रा तिरुनाल आयुर्विज्ञान और प्रौद्योगिकी संस्थान, त्रिवेन्द्रम
तिरुवनन्तपुरम - ६९५०११, केरल, इंडिया
SREE CHITRA TIRUNAL INSTITUTE FOR MEDICAL SCIENCES AND TECHNOLOGY, TRIVANDRUM
Thiruvananthapuram - 695 011, Kerala, India
(An Institute of National Importance under Govt. of India)
Grams : Chitramet, Phone : +91-471-2443152, Fax : +91-471-2550728 / 2446433, E-mail : sct@sctimst.ac.in, Website : www.sctimst.ac.in

Institutional Ethics Committee
(IEC Regn No. ECR/189/Inst/KL/2013)

SCT/IEC/867/FEBRUARY-2016 15.09.2016

Dr. Abhay Suresh Jain
Senior Resident
Department of CVTS
SCTIMST, Thiruvananthapuram

Dear Dr. Abhay Suresh Jain,

The Institutional Ethics Committee reviewed and discussed your application to conduct the study entitled "OUTCOME OF SURGIRES IN SUPRAVASCULAR AORTIC STENOSIS" (IEC/867) on 20th February, 2016.

The following documents were reviewed:

Original submission

1. *Covering letter with HOD forwarding dated, 1/2/2016*
2. *Full proposal*
3. *IEC Application Form*
4. *Forwarding Letter from Guide/HOD*
5. *Protocols*
6. *TAC Approval*
7. *CV of PI and Co-PI*

Revised submission

1. *Covering letter addressed to the Secretary, IEC, SCTIMST with check list*
2. *IEC Application Form*
3. *IEC Recommendation Letter dated 08.03.2016*
4. *Project Proposal*
5. *Modified Proforma*
6. *CV of Principal Investigator and Co. Investigators*

The following members of the Ethics Committee were present at the meeting held on 20th February, 2016 at G. Parthasarathi Board Room, AMCHSS, SCTIMST

SL. No.	Member Name	Highest Degree	Gender	Scientific /Non Scientific	Affiliation with Institution(s)
1.	Justice Gopinathan. P.S	BSc. LLB	Male	Legal Expert (Chairperson)	No
2.	Dr. Asha Kishore	MD, DM	Female	Clinician (Neurologist)	Yes
3.	Shri. O.S. Neelakandan Nair	BE	Male	Engineer	Yes
4.	Dr. Meenu Hariharan	DM	Female	Clinician (Gastro-Enterologist)	No
5.	Dr. Rema M. N	MD	Female	Pharmacologist	No
6.	Dr. V. Raman Kutty	MPH(Harvard) MPhil, MD	Male	Public Health	Yes
7.	Dr. K R S Krishnan	ME, PhD	Male	Biomedical Scientist/Engineer	No
8.	Dr. Kala Kesavan. P	MD	Female	Pharmacologist	No
9.	Dr. Christina George	MD	Female	Psychiatrist	No
10.	Dr. Mala Ramanathan	MSc, PhD, MA	Female	Ethicist/Social Scientist (Member Secretary)	Yes

IEC Decision

The IEC approved the conduct of the study in the present form.

Remarks:
The Institutional Ethics Committee expects to be informed about the progress of the study, any SAE occurring in the course of the study, any changes in the protocol and patient information/informed consent and asks to be provided a copy of the final report.

There was no member of the study team who participated in voting / decision making process. The ethics committee is organized and operated according to the requirements of Good Clinical Practice and the requirements of the Indian Council of Medical Research (ICMR).

Sincerely,

Mala Ramanathan
Member Secretary, IEC

Abhay Jain_Thesis.odt

ORIGINALITY REPORT

12%

SIMILARITY INDEX

PRIMARY SOURCES

1	Sachin Talwar. "Outcomes following surgery for supravalvular aortic stenosis", Indian Journal of Thoracic and Cardiovascular Surgery, 12/2008 Crossref	93 words — 2%
2	circ.ahajournals.org Internet	82 words — 2%
3	www.ncbi.nlm.nih.gov Internet	66 words — 2%
4	intl-ejcts.ctsnetjournals.org Internet	53 words — 1%
5	ats.ctsnetjournals.org Internet	45 words — 1%
6	Jonas, . "Left ventricular outflow tract obstruction : aortic valve stenosis, subaortic stenosis, supravalvar aortic stenosis", Comprehensive Surgical Management of Congenital Heart Disease, 2004. Crossref	44 words — 1%
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9	Delius, R.E.. "Should a bicuspid aortic valve be replaced in the presence of subvalvar or supravalvar	21 words — 1%

aortic stenosis?", The Annals of Thoracic Surgery, 199810
Crossref

10 Collins, R.T.. "Abnormalities of Cardiac Repolarization in Williams Syndrome", The American Journal of Cardiology, 20101001 — 21 words — 1%
Crossref

EXCLUDE QUOTES ON
EXCLUDE BIBLIOGRAPHY ON
EXCLUDE MATCHES OFF

Slno	**1**	**2**	**3**	**4**	**5**	**6**	**7**
sex	m	v	m	m	f	f	m
DATE	14/10/14	13/07/06	02/06/15	22/08/09	07/06/07	11/09/12	03/05/10
AGE	28	15	14	18	2	9	34
WEIGHT	61kg	39kg	32 kg	24kg	9kg	25.4kg	55kg
COEXISTING CARDIOVASCULAR ANOMALY	nil	nil	nil	nil	nil	bav	nil
CONCOMITANT PROCEDURE AT REPAIR	nil	nil	nil	nil	nil	nil	nil
WILLIAMS SYNDROME	no	yes	yes	yes	yes	no	yes
DIFFUSE DISEASE	no	no	no	no	yes	no	no
BICUSPID AORTIC VALVE	no	no	no	no	no	yes	no
CPB TIME	49mins	161min	131min	121min	82min	104min	94min
CLAMP TIME	34mins	107min	84min	83min	32min	69min	57min
PROCEDURE PERFORMED	doty	broms	broms	doty	single patch	doty	single patch
HOSPITAL STAY (DAYS)	6days	10days	9 days	8days	8days	12days	10days
PREOP AV GRADIENT(MM OF Hg)	60	83	69	70	68	64	85
EARLY POSTOP AV GRADIENT	17	30	19	14	18	17	14
LATE FOLLOW UP GRADIENT	17	14	9	13	14	17	14
SURVIVAL	yes	yes	yes	yes	yes	yes	yes
ANEURYSM	no	no	no	no	no	no	no
FUNCTIONAL CLASS{pre op}	grade3	grade3	grade3	grade4	grade3	grade3	grade3
FUNCTIONAL CLASS{post op}	grade1	grade2	grade1	grade 1	1	1	1
ARRYTHMIA	no	no	no	no	no	no	no
ANNULAR SIZE	20mm	22mm	22mm	23mm	11mm	19mm	20.6mm
AORTIC REGURGITATION	no	no	2+	no	no	2+	no
Narrowest part of Ascending Ao/Prox Arch	0.95	0.9	0.8	1.3	1.18	1.13	0.92

8	9	10	11	12	13	14
m	f	f	f	m	f	m
27/09/10	07/10/09	27/09/12	05/08/11	27/05/14	19/02/13	18/06/14
29	1	18	28	5	13	5
40kg	7.5kg	71kg	50kg	15	25kg	15kg
mod AR mod MR RPA Stenosis	SP VSD, PS,RPA Origin stenosis	mild MR	AR, SAM	nil	nil	nil
AVR RPA plasty	RPA Plasty Pulmonary valve commisurotomy,infandibular resection,vsd closure	nil	SAM excision, AVR	nil	nil	nil
yes	no	no	yes	no	no	no
no	no	no	no	no	thickened ascending aorta	no
no	no	no	no	no	no	no
171min	198min	84min	97min	104min	162min	80min
110min	128min	57min	75min	60min	123min	48min
single patch	single patch	doty	single patch	doty	broms	doty
13days	18days	9days	8days	13days	10days	12days
100	90	95	71	42	85	40
22	28	25	14	9	11	12
25	15	24	30	7	9	10
yes	yes	yes	yes	yes	yes	yes
no	no	no	no	no	no	no
grade4	grade4	grade 3	grade4	grade3	grade4	grade3
2	1	2	2	1	1	1
no	no	no	no	no	no	no
20mm	15mm	21mm	19mm	15mm	18mm	15mm
1+ intravalvular	no	no	1+	1+	2+	no
1.16	1.3	0.88	0.93	1.2	1.16	0.96