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PROJECTS

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POST DOCTORAL FELLOWSHIP IN CARDIO THORACIC SURGERY

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Projects:

1. Experience of Modified Shumacker and King Repair for Total Anomalous Pulmonary Venous Connection

2. Comparative study of single and double patch technique for sinus venosus atrial septal defect with partial anomalous pulmonary venous connection

Experience of Modified Shumacker and King Repair for Total Anomalous Pulmonary Venous Connection

Abstract

Objective: To assess the usefulness and reproducibility of modified Shumacker and King approach for all types of total anomalous pulmonary venous connection.

Methods: We retrospectively studied forty-two consecutive patients operated from July 2002 to June 2005 with 27 boys and 15 girls between 5 days and 9 years (median: 90 days). There were 20 supracardiac, 12 cardiac, 6 infracardiac and 4 mixed types of the lesion. The body weight ranged between 2 to 16 kg with a median of 4 kg. Eleven were operated as emergencies. Associated anomalies other than ductus were present in two patients. Modified Shumacker and King repair was done for all supracardiac, infracardiac, two cardiac and three mixed lesions.

Results: Mortality at one-month follow-up was 7/42(16.6%). Five presented as emergencies; four were infracardiac, one supracardiac, one mixed and one cardiac. Two patients were reoperated for pulmonary vein stenosis and pulmonary vein obstruction respectively (4.76%). Delayed vertical vein ligation was done in one patient. No patient had postoperative arrhythmia in cardiac type. Three patients of the modified Shumaker and King repair had rhythm problems at the end of one month 3/31(9.6%). Lung complications were present in 9(21.4%). Six patients had moderate to severe pulmonary artery hypertension. Low body weight and emergency repair were found as significant predictors of mortality.

Conclusion: Modified Shumacker and King repair is a safe and reproducible technique applicable to most anatomic forms of total anomalous pulmonary venous connection. Early diagnosis, referral and repair hold the key for better results in developing countries.

Introduction:

Total anomalous pulmonary venous connection (TAPVC) is a relatively rare congenital anomaly with an incidence of less than 3% of congenital heart disease[1, 2]. The condition is fatal if left untreated and has a mortality of 80% by one year[3, 4]. Shumacker and King described the classical technique for surgical repair of TAPVC. In this approach an incision is made parallel to

the atrioventricular groove in the right atrium and the septum excised, a transverse incision is further made extending across the atrial septal defect to the posterior wall of the left atrium, the underlying common pulmonary vein is opened, and anastomosis is performed from outside the heart[5]. A simple modification of this technique by excluding the first incision is safe and reproducible and is applicable to almost all anatomic forms of the disease. Although there are speculations of a higher incidence of postoperative arrythmias[6], there was no correlation between the presence of arrhythmia and the date of repair, anatomical subtype, operative approach or adequacy of repair[7]. This study summarizes our surgical experience with 42 consecutive patients who underwent TAPVC repair at our institute with a focus on immediate results of modified Shumacker and King repair.

Methods:

This is a retrospective study of forty-two consecutive patients undergoing repair of TAPVC from July 2002 to June 2005 in our institute. There were 27 boys and 15 girls the ratio being 1.8:1. Age ranged between 5 days to 9 years with a median of 90 days. There were 33 infants (78.7%). The median weight was 4kg (Table-1). The patients presented with tachypnoea, feeding difficulty, cyanosis and recurrent respiratory tract infections in infancy, with dyspnoea on exertion in older patients and with failure in emergency. Chest X-ray showed pulmonary venous congestion in all emergency cases off which one had bilateral lung consolidation. Preoperative echocardiography was the modality of diagnosis and the findings are shown in (Table-2). There were 12 cardiac (28.5%), 20 supracardiac (47.6%), 6 infracardiac (14.2%), 4 mixed (9.5%). Two supracardiac TAPVC had cortriatriatum dexter and supramitral ring respectively. Eleven patients (26.1%) presented as emergencies due to obstructed drainage, which included all infracardiac, 2 cardiac and 3 supracardiac cases. Associated anomalies other than PDA were present in two patients (4.7%), one case of supracardiac TAPVC with supramitral ring and one supracardiac TAPVC with cortriatriatum dexter. The modified Shumacker and King was done for all supracardiac, infracardiac, two cardiac and three mixed TAPVC the total number being thirty one (73.8%) (Table-3) and coronary sinus unroofing and pericardial patch rerouting was done for the

remaining cardiac TAPVC. In thirty patients (71.4%) the sternum was left open, with sterile adhesive film closure of the wound, which included all emergencies. The sternum was closed 18 to 76 hours after surgery with a mean of 32.8 ± 7.4 hours depending on the haemodyanamic status. Two patients were reoperated, one who was operated for cardiac TAPVC and presented with persistent symptoms of repeated respiratory infections and failure to thrive on follow up. Echocardiography in this patient showed stenosis between the common pulmonary vein and coronary sinus, which was missed during the first operation and was corrected by a subsequent modified Shumacker and King repair. The second patient had supracardiac TAPVC with vertical vein on the right side joining the azygous vein, a repair was done and azygous vein ligated. The patient developed right upper lobe congestion for which the ligation was removed and the azygous vein entry was closed with a pericardial patch from within the superior venacava, with widening of superior venacava with another pericardial patch. Both patients are on follow up and have a gradient free drainage of the pulmonary veins into the left atrium. A total of 42 patients underwent TAPVC correction clinical profile, patient characteristics, operative technique, postoperative outcomes and functional status are discussed.

Surgical technique- Modification of the Shumacker and King repair

Supracardiac (n=18), Infracardiac (n=6), cardiac (n=2), mixed (n=3)

Surgery is performed through a midline sternotomy, the thymus excised, aortic and bicaval cannulation was performed in all cases. Patent Ductus Arteriosus is ligated in all cases as a routine. The vertical vein is dissected extrapericardially in all cases. Aorta is clamped, heart arrest is instituted with cold sanguinous cardioplegia and patients are cooled to 18 degrees. In smaller infants less 2.5 kg double venous cannulation with innominate vein cannulation is performed, as the area of exposure is less. Complete circulatory arrest was instituted in 9 patients all of whom were infants below 2.5kg for the whole repair and the remaining patients were operated in low flows with or without a short period of circulatory arrest. A short period of circulatory arrest was used in few patients to suture the left end of the anastomosis if required. The horizontal pulmonary venous confluence is dissected during cooling from the right side. A transverse incision is made a short distance from the AV groove. This incision is further extended

across the fossa ovalis or the atrial septal defect into the left atrium towards the base of left atrial appendage. An incision is made in the common venous chamber parallel to the left atrial incision and a direct anastomosis fashioned between the two structures using 6-0 or 7-0 prolene sutures. The ASD is closed using a pericardial patch. The anomalous vertical vein is snared and weaning of bypass attempted and if tolerated the vertical vein is ligated. Only one patient had a delayed ligated of the vertical vein at the time of delayed sternal closure after 48 hours

Unroofing of coronary sinus and pericardial patch rerouting

Cardiac (n=10) and mixed TAPVC (n=1)

All cases of cardiac TAPVC except two underwent the repair wherein the coronary sinus is unroofed towards the LA and the tissue between the coronary sinus and the septal defect excised. The resulting septal defect is closed with a pericardial patch. One cardiac TAPVC had obstruction proximal to the coronary sinus and modified Shumacker and King repair was done in that case. The second case after discharge presented with recurrent symptoms and a modified Shumacker and King repair was done to relieve the persistent pulmonary venous gradient, which was present between the common pulmonary vein and coronary sinus which was missed at the time of first operation.

Sternum left open (n=30)

At the end of surgery in all neonates, patients weighing less than 3.5 Kg, all emergency cases and if there was significant myocardial edema, diffuse mediastinal bleeding or haemodynamic instability, the sternum was left open, with sterile adhesive film closure of the wound.

Delayed sternal closure (n=28)

The sternum was closed 18 to 76 hours with a mean of 32.8 ± 7.4 hours after surgery depending on the haemodynamic status. Two patients died before sternum could be closed.

Postoperative management

All patients were electively ventilated for at least 48 hours. All infants were ventilated with the pressure control mode of ventilation and for all above one year of age volume control ventilation was used. The partial pressure of carbon dioxide was attempted to maintain at 25-30mm Hg. We

have monitored the pulmonary artery pressure to assess pulmonary hypertension by daily echocardiography, which helped us in deciding weaning patients off ventilator.

A variety of inotropes and vasodilators have been used to maintain the cardiac output and the pulmonary artery pressure like isoprenaline, adrenaline, milrinone, dobutamine, dopamine, nitroglycerine, sodium nitoprusside. Nitric oxide, ECMO and assist devices are not available in our institute.

If there was a fall in urine output despite the use of diuretics, peritoneal dialysis was started. If patients required ventilation for more than 48 hours enteral feeding was started. Total parenteral feeds were started only if enteral feeding was not tolerated.

RESULTS

Hospital mortality has been 7/42 (16.6%). All deaths were in the postoperative period of which five presented as emergency; four were infracardiac, one supracardiac one mixed and one cardiac. Four patients died as a result of myocardial failure following uncontrolled pulmonary hypertensive crisis, one died of septicemia following prolonged ventilation, one patient expired due to ARDS and one due to multiple organ dysfunction syndrome. Sternum was left open in 30 patients for delayed closure of which 28 could be closed and two died before sternal closure. Two patients were reoperated one for pulmonary vein obstruction and one for stenosis between the common chamber and coronary sinus (4.76%), which was missed during the first surgery. Delayed vertical vein ligation was done in one patient as the child did not tolerate coming off bypass and was ligated during sternal closure. The incidence of postoperative arrhythmia was none in the cardiac type. Seven patients of the Shumaker King repair had immediate post op rhythm abnormalities in the form of supraventricular tachycardia, junctional rhythm and atrial fibrillation, of which three patients had persistent rhythm problems at the end of one month 3/31(9.6%). Two had junctional rhythm and one had atrial fibrillation. At 9 months follow up the patient having atrial fibrillation was found to have fast ventricular rate and was cardioverted. This patient has been on follow up since then and is still in atrial fibrillation after 3 years but is asymptomatic. The duration of ventilation was 20 to 116 hours with a mean of 65.5 ± 23.9 hours. Major morbidity included moderate to low ventricular function at the time of discharge in three

patients. Two of these patients had pulmonary arterial hypertension (PAH) of 56mm and 52 mm respectively and the other had no PAH. Lung complications were present in 9 patients of which one had ARDS and died, 1 had bilateral lung consolidation developed septicemia and died, 5 had lung collapse, 1 had bronchopneumonia and one had right pleural effusion with underlying collapse. One patient had recurrent seizures in the postoperative period. CT scan was done which was normal; the patient was subsequently put on antiepileptics, which resulted in resolution of the seizures. Six patients had moderate to severe pulmonary artery hypertension at one month. One patient of supracardiac TAPVC had a persistent pulmonary venous gradient of 30mm Hg post repair and a pulmonary pressure of 35mm Hg and is awaiting re-surgery. Twelve patients were subjected to post operative peritoneal dialysis due to postoperative acute renal failure (Table-4). All patients required inotropic support postoperatively for 36 hours to 12 days. The ICU stay ranged from 3 to 26 days with a mean of 9.8 ± 5.05. The mean hospital stay was 18.3 ± 6.4 days (Table-5). The significant risk factors, which were identified to determine death, were emergencies due to obstructed drainage and low body weight. A total of 29 patients (69.05%) were below the 5th percentile and 39 patients (92.8%) were below the 25th percentile in body weight. Both these factors were found to be statistically significant predictors of mortality. Using Fishers exact tests P=. 034 for body weight and mortality and P=. 001 for emergency repair and mortality.

Discussion

TAPVC is a rare congenital anomaly with an unfavorable natural history. Only 20% survive after their first year of life if left untreated[3, 4]. The hospital mortality has been decreasing over time due to standardized operative technique and good perioperative and postoperative care[8, 9]. The mortality has been 19% before 1995 and in the current era it is around 5% [10]. In emergency cases, which present with obstructed drainage we use warm induction of cardioplegia before reducing the temperature of the perfusate to prevent the heart from becoming hard and fibrillating. This is believed to be due to preoperative acidosis or due increased wall tension, which limits the perfusion of the inner layers of the heart[11]. In cardiac TAPVC there are two approaches first is to excise the roof of coronary sinus to enable it to communicate with the LA and fossa ovalis, in the

second approach the foramen ovale is enlarged, roof of the coronary sinus is excised and the foramen ovale and coronary sinus orifice are sutured separately[11]. We enlarge the foramen ovale and remove the tissue between the fossa ovalis and coronary sinus and excise the roof of coronary sinus, the resulting interatrial defect is closed with a single pericardial patch, this prevents occurrence of stenosis at the repair site. For the other types various approaches have been described but we prefer the modified Shumacker and King repair, which can be performed in almost all anatomic types of TAPVC. The Shumacker and King repair was, devised in 1961. In the classical Shumacker and King repair a longitudinal incision is made on the right atrium parallel to the AV groove, another transverse incision is made beneath the first almost perpendicular to it extending across the atrial septum at the level of the atrial septal defect into the left atrium. The modification is, the first incision is omitted and the transverse incision starts a short distance from the AV groove making it longer than the transverse incision of the classical technique Shumacker and King repair[12]. This approach offers the best possible exposure and is reproducible besides the fact that a wide stenosis free anastomosis can be performed. The usefulness of this approach is increased in cases where there is stenosis at the pulmonary vein common chamber junction, which was encountered by us in one of our cases wherein a wider incision and anastomosis can be easily performed. It is also a better approach in cases of TAPVC associated with other congenital anomalies like cortriatriatum dexter or the supramitral ring where a good exposure is obtained. Lastly it involves minimal or no handling of the ventricle and is applicable to all anatomic forms of TAPVC. Although a group of surgeons feel that it is associated with a higher incidence of postoperative arrythmias due to the long atrial incision[6], some authors have a long term follow up with a similar modification with no correlation between the technique and arrythmias[7, 8]. Surgeons adopting the other approaches[13] have also been experiencing important ventricular as well as supraventricular ectopy. Bogers et al. who have adopted this technique have no late atrial arrhythmias in their series where the mean duration of follow up has been 12 years[8]. Jonas et al have shown no correlation between the presence of arrhythmia and the date of repair, anatomical subtype, operative approach or adequacy of repair[7]. Our early results too did not show any increase in arrhythmias with this approach. The atrial

septal defect was always closed with a patch, which avoids any narrowing of the anastomosis, and it also helps to supplement the size of the left atrium.[7]

The results of TAPVC repair have drastically improved in recent years due to improvement in the surgical technique with better myocardial preservation and better intraoperative and postoperative care[14, 15]. In the past various factors like the anatomic type, younger age, preoperative metabolic acidosis, presence of obstruction and pulmonary artery pressure were considered to influence the operative mortality[15, 16]. In the current era these factors are less important [17,18]. Our mortality of 16.6%, which is high as compared to the western literature where the mortality has dropped to 5% in recent years, this can be attributed to low body weight and delayed referrals. Out of the 7 deaths in our series, 5 had presented as emergencies and had multiple problems like respiratory tract infection in 3, severe PAH in all and with deranged liver function in 2 patients on arrival, all due to delayed referrals. Secondly low body weight, which can be attributed to malnutrition partly due to cardiac cachexia, has also been responsible for increased mortality as 69.05% of the patients were below the 5th percentile and 92.8% were below the 25th percentile. All patients who expired had a body weight below the 3rd percentile. These patients are prone to variety of infections and react poorly to cardiopulmonary bypass. Both these factors were statistically significant predictors of mortality. Due to financial constraints we do not have facilities like nitric oxide and ECMO. With the availability of these facilities in the future our results would probably improve in this subset of patients.

To conclude modified Shumacker and King repair is a very safe and reproducible technique for correcting most of the anatomical forms of TAPVC and has been effectively used in our institution. Long incision and atrial suture line has not been associated with increase in the post-operative rhythm abnormalities in the immediate postoperative period. The results of surgery for TAPVC have drastically improved in recent years; early detection, referral and surgery hold the key to better survival in the developing countries.

References:

1.Cope JT, Kron IL. Anomalies of pulmonary venous return and cortriatriatum. In: Kaiser LR, Kron IL, Spray TL, eds. Mastery of cardiothoracic surgery. Philadelphia: Lippincott-Raven Publishers, 1998:876-79

2. N Sinzobahamvya, Arenz C, Brecher AM, Blaschczok HC, and Urban AE. Early and long term results for correction of total anomalous pulmonary venous drainage (TAPVD) in neonates and infants. Eur J Cardiothor Surg 1996;10:433-8

3.Rowe RD. Anomalies of venous return .IN: Keith JD, Rowe RD, Vlad P, eds. Heart disease in infancy and childhood. 3rd ed. New York: Macmillan, 1978:554.

4. Burroughs JT, Edwards JE. Total anomalous pulmonary venous connection. Am Heart J 1960;59:913-31

5.Shumacker HB Jr, King H. A modified procedure for complete repair of total anomalous pulmonary venous drainage. Surg Gynaecol Obstet 1961 ;112:763

6.Alain Serraf, Emre B,Daniel R, Miguel S, Fancois L. Claude P. Modified superior approach for repair of supracardiac and mixed total anomalous pulmonary venous drainage. Ann Thorac Surg 1998;65 1391-3

7. Jonas RA . In: Anomalies of the pulmonary vein: Comprehensive Surgical Management of congenital heart disease, first edition, Arnold publications 2004:392- 410

8. Bogers AJJC, Baak R, Lee PC, Boersma, Meijboom F.J, Witsenburg M. Early results and long term follow up after corrective surgery for total anomalous pulmonary venous return. Eur J Cardiothor Surg 1999;16:296-9

9.Caldarone CA, Najm HK, Kadletz M, Smallhorn JF, Freedom RM, Williams WG, and Coles JG. Surgical management of total anomalous pulmonary venous drainage: impact of coexisting cardiac anomalies. Ann Thorac Surg 1998;66 1521-6.

10. Kirshbom PM, Myung RJ, Gaynor JW, Ittenbach RF, Paridon SM, DeCampli WM. Preoperative pulmonary venous aostruction affects long-term outcome for survivors of total anomalous pulmonary venous connection repair Ann Thorac Surg 2002;74:1616-20.

11. Kouchoukos NT, Blackstone EH, Donald DB, Hanley F, Robert BK. Total Anomalous Pulmonary Venous Connection. Kirklin/Barratt-Boyes Cardiac surgery, third edition, 2003: 753-780

12. Stark J. Total anomalous pulmonary venous drainage: surgical repair. Rob and Smiths operative surgery, Cardiac surgery 4th edition 129-137

13.Bhan A, Umre MA, Choudhary SK, Saxena A, Sharma R, Airan B. Cardiac arrythmias in surgically repaired total anomalous pulmonary venous connection: a follow up study. Indian Heart J 2000 ;52: 427-30.

14. Raisher BD, Grant JW, Martin TC, Strauss AW, Spray TL. Complete repair of total anomalous pulmonary venous connection in infancy. J Thorac Cardiovasc Surg 1992;104:443-8

15. Bowe EL, de Leval MR, Taylor JFN, Macartney FJ, Szarnicki RJ, Stark J. Infradiapraghmatic total anomalous pulmonary venous drainage: surgical treatment and long term results. Ann Thorac Surg 1981; 31:544-50.

16. Hammon JW Jr, Bender HW Jr, Graham TP Jr, Boucek RJ Jr, Smith CW, Erath HG, Jr. Total anomalous pulmonary venous connection in infancy. Ten years experience including studies of post-operative ventricular function. J Thorac Cardiovasc Surg 1980;80:544-51

17.Lupinetti FM, Kulik TJ, Beekman RH, Crowley DC , Bowe EL. Correction of total anomalous pulmonary venous connection in infancy. J Thorac Cardiovasc Surg 1993;106:880-5

18.Cobanoglu A, Menashe VD. Total anomalous pulmonary venous connection in neonates and young infants: repair in current era. Ann Thorac Surg 1993;55:43-9

TABLES

TABLE 1 -Patient demography

Statistical parameters	Age in days	Weight in kilograms
1) Mean	318.36	5.34
2) Median	90	4
3) Standard deviation	587.141	3.804
4) Minimum	5	2
5) Maximum	3285	16

Table 2
Anatomical forms of TAPVC and their association with obstructed drainage and mortality

Type of TAPVC	Number (Percentage)	Obstructed	Mortality
Supracardiac	20 (47.6%)	3	1
Cardiac	12 (28.5%)	2	1
Infracardiac	06 (14.2%)	6	4
Mixed	04 (9.5%)	0	1

Table 3 :Operative technique

Modified Shumacker and King repair	Number (%)
Supracardiac	20/20 (100%)
Infracardiac	6/6 (100%)
Cardiac	2/12 (16.6%)
Mixed	3/4 (75%)

Table 4
Complications

Complications	S/K repair	Rerouting with patch
Arrhythmias	3	0
Lung Complications	7	2
CNS complications	1	0
Renal failure	9	3
Moderate to low ejection frac.	3	0
Persistent PAH	5	1
Pulmonary venous gradient	1	0

Table 5
Duration of ventilation, ICU stay and hospital stay

Statistical parameters	Duration of ventilation in hours	ICU stay in days	Hospital stay in days
Mean	65.55	9.83	18.31
Median	62	10	17
Standard deviation	23.919	5.055	6.471
Minimum	20	3	6
Maximum	116	26	33

Statistical data:

Frequencies

SEX

		Frequency	Percent	Valid Percent	Cumulative Percent
Valid	F	15	35.7	35.7	35.7
	M	27	64.3	64.3	100.0
	Total	42	100.0	100.0	

Frequencies

Statistics

		AGEDAYS	WEIGHT	HSTAY	VENT	ICU_STAY
N	Valid	42	42	35	42	35
	Missing	0	0	7	0	7
Mean		318.36	5.34	18.31	65.55	9.83
Median		90.00	4.00	17.00	62.00	10.00
Std. Deviation		587.141	3.804	6.471	23.919	5.055
Minimum		5	2	6	20	3
Maximum		3285	16	33	116	26

Mortality between weight groups and surgery type (emergency/others); both are statistically significant P=. 034 and P=. 001 respectively, based on Fishers exact tests.

WTGRP * DEATH

Crosstab

			DEATH		
			0 No	1 Yes	Total
WTGRP	0 Others	Count	22	1	23
		% within WTGRP	95.7%	4.3%	100.0%
	1 <3	Count	13	6	19
		% within WTGRP	68.4%	31.6%	100.0%
Total		Count	35	7	42
		% within WTGRP	83.3%	16.7%	100.0%

Chi-Square Tests

	Value	df	Asymp. Sig. (2-sided)	Exact Sig. (2-sided)	Exact Sig. (1-sided)
Pearson Chi-Square	5.555[b]	1	.018		
Continuity Correction[a]	3.768	1	.052		
Likelihood Ratio	5.921	1	.015		
Fisher's Exact Test				.034	.025
Linear-by-Linear Association	5.423	1	.020		
N of Valid Cases	42				

a. Computed only for a 2x2 table

b. 2 cells (50.0%) have expected count less than 5. The minimum expected count is 3.17.

EMER * DEATH

Crosstab

			DEATH		Total
			0 No	1 Yes	
EMER	0 No	Count	30	1	31
		% within EMER	96.8%	3.2%	100.0%
	1 Yes	Count	5	6	11
		% within EMER	45.5%	54.5%	100.0%
Total		Count	35	7	42
		% within EMER	83.3%	16.7%	100.0%

Chi-Square Tests

	Value	df	Asymp. Sig. (2-sided)	Exact Sig. (2-sided)	Exact Sig. (1-sided)
Pearson Chi-Square	15.396[b]	1	.000		
Continuity Correction[a]	11.923	1	.001		
Likelihood Ratio	13.854	1	.000		
Fisher's Exact Test				.001	.001
Linear-by-Linear Association	15.029	1	.000		
N of Valid Cases	42				

a. Computed only for a 2x2 table

b. 1 cells (25.0%) have expected count less than 5. The minimum expected count is 1.83.

Comparative study of single and double patch technique for sinus venosus atrial septal defect with partial anomalous pulmonary venous connection

Abstract:

Objective: Surgical correction of sinus venosus atrial septal defect with partial anomalous pulmonary venous connection to the superior venacava has been associated with obstruction to the venous return and sinus node dysfunction. We present our follow up of two approaches of tackling the lesion thereby comparing their postoperative results.

Methods: Forty-four patients were operated from March 1999 to January 2005. Out of these 37 patients are on follow-up (84.09%). They were divided into patients who underwent the single patch repair (group A) and patients who underwent the double patch repair (group B). Group A had 18 patients and group B had 19 patients. Their age ranged from 3 to 50 years. Echocardiography was done for assessing the intactness of the pericardial patch, gradient across superior venacava, gradient across the right pulmonary vein at the level of the patch and pulmonary artery pressure. Electrocardiography was done for reassessing the rhythm. Both these investigations were done 7 days after surgery and during subsequent follow up.

Results: The mean duration of follow up was 22.56 months. There was no early or late postoperative death. There were no residual defects in any of these patients. Six patients of group A and 2 patients of group B had turbulence and a significant superior venacava - right atrium pressure gradient of more than 6mm of Hg. Nine patients of group A had a significant gradient causing turbulence across the right superior pulmonary vein at the level of the patch while no patient of group B had turbulence across the pulmonary vein. Four patients of group A and none of group B had postoperative rhythm abnormalities in the form of junctional rhythm on late follow up. There was no other major complication.

Conclusion: Sinus venosus atrial septal defect with partial anomalous pulmonary venous connection can safely tackled by multiple techniques with very low morbidity. The two-patch technique offers better results in terms of superior venacava narrowing and gradient across the

pulmonary vein at the level of the patch without any increase in complications. It is technically reproducible and can be used for all forms of the lesion.

Introduction:

Sinus venosus syndrome accounts for 10% of the patients presenting for surgery with an atrial septal defect (ASD)[1]. Sinus venosus syndrome comprises of a sinus venosus ASD with partial anomalous pulmonary venous connection (PAPVC) to the superior venacava. Inappropriate surgery in this condition may lead to sinus node dysfunction, residual shunt and obstruction of the pulmonary veins or superior venacava (SVC)[2-4]. The goal of sinus venosus repair is to eliminate the intracardiac shunt without causing stenosis of the pulmonary veins or the superior venacava and without injuring the sinus node. Various surgical techniques involving incisions across cavoatrial junction[2,5], right atrial free wall muscle flaps[6], and transection and relocation of the SVC to the right atrial appendage[7] are adopted. Two commonly adopted techniques are the single patch and the double patch repair. The purpose of this study was to compare the postoperative results of both these approaches with respect to superior venacava narrowing, right superior pulmonary vein narrowing, residual ASD and rhythm.

Materials and methods

Between March 1999 and January 2005 forty-four patients underwent repair for sinus venosus ASD with PAPVC to the SVC of which 37 patients are on follow up (84.09%). The mean follow was 22.5 months. Their age ranged from 3 years to 50 years (mean: 15.1 years). Twenty-eight of the 37 patients (75.6%) patients were asymptomatic before surgery, whereas the remaining nine patients complained of easy fatigability, chest pain or reduced exercise tolerance. All the symptomatic patients were above 40 years. Preoperative echocardiography revealed pulmonary hypertension of more than 30mm of Hg in seven patients. Documented arrythmias were absent before surgery in all patients. Right bundle branch block pattern was present in 10 (27%) of them. None of the patients were subjected to angiography. Follow up included physical examination, electrocardiography and transthoracic echocardiography as part of postoperative protocol. Colour doppler was used to asses the venous structures.

Operative technique: In all patients, the SVC was cannulated above the anomalous venous drainage and a curved cannula was used for the inferior venacava. The surgery was performed under mild hypothermia and cardioplegic arrest. Eighteen patients (group A) underwent a single patch technique wherein a patch was used to close the ASD and reroute the anomalous pulmonary vein or veins no patch was used to enlarge the SVC and 19 patients underwent a double patch technique where a patch was used to enlarge the SVC as well.

Results:

The mean duration of follow up was 22.56 months. There was no early or late postoperative death. There were no residual defects in any of these patients. Six patients of group A and 2 patients of group B had a significant superior venacava - right atrium pressure gradient causing turbulence and a gradient of more than 6mm of Hg and was found to be statistically significant ($p=0.005$). One patient from group A having a gradient of 13 had symptoms of SVC narrowing in the form of recent onset headache, frequent episodes of giddiness and had few dilated veins in the neck. He is being investigated for corrective surgery.

Nine patients of group A had a significant gradient causing turbulence across the right superior pulmonary vein at the level of the patch while no patient of group B had turbulence across the pulmonary vein ($p=0.0001$). Off the nine patients, two patients having a gradient of 9 and 8.4 had right upper and middle lobe haziness in repeated X rays. Both these patients are asymptomatic and are on follow up.

Four patients of group A had postoperative junctional rhythm with intermittent sinus escapes. All these patients are asymptomatic and are on follow up. None of the patients from group B had any rhythm disturbance. Four off the seven patients who had elevated pulmonary artery pressure had reduction in pulmonary artery pressure post surgery.

Discussion:

The surgical management of sinus venosus ASD with partial anomalous pulmonary venous connection has evolved over the years. It has been ranging from closure of the ASD with either leaving or ligating the anomalous veins or total correction with rerouting of the anomalous veins to the left atrium[8,9].

The commonly done procedures include the single or the two patch technique as shown by various reports[10, 11,12]. There have been speculations that compartmentalization of SVC with patch rerouting were followed by SVC obstruction[13, 14]. We have been able to show that the double patch method is superior to the single patch method with regards to the SVC narrowing. The patients operated without SVC augmentation have a statistically significant number (p=0.005) of cases compared to the cases which have underwent a patch augmentation. Significant SVC gradient had been taken as 6mm of Hg by Trusler et al[2]. We have also had turbulence in the SVC in the region of the patch as visualized in doppler when the gradient is above 6mm.

None of the patients in group B had any gradient across the pulmonary vein due to the patch but 9 patients of group A have turbulence at that region in postoperative follow up echocardiography (p=0.0001). This may be related to the fact that smaller patches are used in single patch method in order to prevent any redundancy of the patch, which may eventually cause SVC narrowing. The size of the patch is of utmost importance in this method as a smaller patch may cause pulmonary vein narrowing and a larger patch may contribute towards SVC narrowing. Some surgeons perform the single patch method for PAPVC to the SVC right atrium junction and a double patch method for any other anomalous connection above the junction[12]. All our patients of the single patch who had turbulence across the pulmonary veins as well as significant SVC gradient including the patient who had clinical and Echocardiographic features of SVC stenosis had PAPVC only to the SVC right atrium junction. Four patients from group A developed turbulence across the pulmonary vein at the level of the patch off these two patients have X ray findings of venous congestion. They are presently asymptomatic and are on follow up.

Rhythm disturbances are known complications of the surgery. We have encountered four patients with postoperative junctional rhythm all of whom underwent the single patch repair. This was due to intraoperative damage to either the SA node or the artery to the SA node. There were no rhythm abnormalities in the remaining patients of either of the groups in long term follow up. There are reports that incision in the SVC or the across the cavoatrial junction may cause sinus node dysfunction in follow up even if there is no injury to the node or the artery due to fibrosis in that area [15]. We have not encountered such events in long-term follow-up. DeLeon et al. have had

rhythm abnormalities only in two of the 18 patients wherein the cavoatrial incision has been made through the sinus node[16]. Therefore, if the incision is properly planned and sutured carefully the chances of rhythm abnormalities are very less. The only risk is the varied course of the artery to the SA node, which may lie in the lateral aspect of the SVC right atrium junction but this risk persists for all the techniques except the Gustafson, Warden and Murray procedure[7]. This modification involves transection of the SVC with anastomosis to the right atrial appendage has the disadvantage of a venous anastomosis with the possibility of acute thrombosis or subsequent stenosis[17]. The transcaval repair involves an incision only in the superior venacava and is applicable for PAPVC to the SVC-RA junction besides the patch should not be redundant whereby it can cause SVC obstruction [18]. To conclude the two-patch technique of performing repair for Sinus venosus ASD with PAPVC is a safe, simple and reproducible technique. It does not cause SVC or pulmonary venous obstruction and maintains normal sinus node function in the long term follow up

References:

1. Davia JE, Cheitlin MD, Bedynek JL. Sinus venosus atrial septal defects: analysis of 50 cases. Am Heart J 1973;85:177-85
2. Trusler GA, Kazenelson G, Freedom RM, Williams WG, Rowe RD. Late results following repair of partial anomalous pulmonary venous connection with sinus venosus atrial septal defect. J Thorac Cardiovasc Surg 1980;79:776-81
3. Friedli B, Gherin R, Davignon A, Fouron JC, Stanley P. surgical treatment of partial anomalous pulmonary venous drainage . A long term follow up study. Circulation 1972;45:159-70
4. Anderson PD, Glaser SP, Czarnecki S, Hopeman AR. Three unusual complications resulting from attempted repair of partial anomalous pulmonary venous drainage. Chest 1976;69:384-7
5. Kyger ER 3, Frazier OH, Cooley DA, Gillette PC, Reul GJ Jr, Sandiford FM. Sinus venosus atrial septal defect: early and late results following closure in 109 patients. Ann Thorac Surg 1978;25:44-50

6. Robicsek F, Daugherty HK, Cook JW, Serle JG. Sinus venosus type of atrial septal defect with partial anomalous pulmonary venous return. J Thorac Cardiovasc Surg 1979; 78:559-62
7. GustafsonRA, Warden HE, Murray GF. Partial anomalous pulmonary venous connection to the superior venacava. Ann Thorac Surg 1995;60:S614-7
8. Kirklin JW, Ellis FH, Wood ED. Treatment of anomalous pulmonary venous connections in association with interatrial communications. Surgery 1956;39:389-98
9. Gustafson RA, Warden HE, Murray GF, Hill RC, Rozar GE. Partial anomalous pulmonary venous connection to the right side of the heart. J Thorac Cardiovasc Surg 1989;98:861-8
10. Ohmi M, Mohri H. A single pericardial patch technique for repair of partial anomalous pulmonary venous drainage associated with sinus venosus atrial septal defect. Ann Thorac Surg 1988;46:360-1
11. Kirklin/Barratt-Boyes . Cardiac surgery. Philadephia: Churchill Livingstone 3rd edition. 715-752
12. Backer CL, Mavroudis C. Paediatric Cardiac Surgery. Philadelphia: Mosby 3rd edition 283-297
13. Kirklin JW, Ellis FH, Wood ED. Treatment of anomalous pulmonary venous connections in association with interatrial communications. Surgery 1956;39:389-98
14. Robicsek F, Daugherty HK, Cook JW, Selle JG. Sinus venosus type of atrial septal defect with partial anomalous pulmonary venous return. J Thorac Cardiovasc Surg 1979;78:559-62.
15. Tung KSK, James TN, Effler DB, McCormack J. Injury of the sinus node in open-heart operations. J Thorac Cardiovasc Surg 1967;53:814-29
16. DeLeon SY, Jenny EF, Ilbawi MN, Husayni TS, Quinones JA, Ow EP, Bell TJ, Pifarre R. Surgical techniques in partial anomalous pulmonary veins to the superior venacava. Ann Thorac Surg 1993;55:1222-6

17. Pathi V, Guererro R, MacArthur KJ, Jamieson MPG, Pollock JCS. Sinus venosus defect:single patch repair with caval enlargement. Ann Thorac Surg 1995;59:1588-9

18. Nicholson IA, Chard RB, Nunn GR, Cartmill TB. Transcaval repair of sinus venosus syndrome. J Thorac Cardiovasc Surg 2000;119:741-44

Statistical data:

T-Test

Group Statistics

	VN	N	Mean	Std. Deviation	Std. Error Mean
SVCVEL	1 single	18	1.151	.2659	.0627
	2 double	19	.923	.1861	.0427
scv gradient	1 single	18	5.7117	2.70321	.63715
	2 double	19	3.6074	1.74350	.39999
RUPV_VEL	1 single	18	1.192	.2088	.0492
	2 double	19	.835	.1843	.0423
rupv gradient	1 single	18	6.0044	2.07170	.48830
	2 double	19	2.8884	1.29348	.29674

Independent Samples Test

		t-test for Equality of Means						
		t	df	Sig. (2-tailed)	Mean Difference	Std. Error Difference	95% Confidence Interval of the Difference	
							Lower	Upper
SVCVEL	Equal variances assumed	3.027	35	.005	.227	.0751	.0749	.3799
scv gradient	Equal variances assumed	2.829	35	.008	2.1043	.74372	.59447	3.61413
RUPV_VEL	Equal variances assumed	5.521	35	.000	.357	.0647	.2257	.4882
rupv gradient	Equal variances assumed	5.520	35	.000	3.1160	.56447	1.97010	4.26195

Frequencies

Statistics

SEX

N	Valid	37
	Missing	0

SEX

		Frequency	Percent	Valid Percent	Cumulative Percent
Valid	f	19	51.4	51.4	51.4
	m	18	48.6	48.6	100.0
	Total	37	100.0	100.0	

Descriptives

Descriptive Statistics

	N	Minimum	Maximum	Mean	Std. Deviation
AGE	37	3	50	15.11	12.507
Valid N (listwise)	37				