VENTRICULAR SEPTAL DEFECT IN INFANTS AND CHILDREN WITH INCREASED PULMONARY VASCULAR RESISTANCE AND PULMONARY HYPERTENSION - SURGICAL MANAGEMENT: LEAVING AN ATRIAL LEVEL COMMUNICATION

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Objective : To evaluate the surgical and medical efficacy of the patients operated for Ventricular Septal Defect (VSD) with Pulmonary Hypertension and Pulmonary Vascular Resistance (PVR). Infants and children with elevated PVR and Pulmonary Hypertension are associated with significant mortality and morbidity after surgical closure. Circulatory assist devices and sophisticated medicines may not be available to help in the management of infants and children with elevated Pulmonary artery pressure and resistance. We left Patent Foramen Ovale (PFO) or made atrial communication to decrease the morbidity and mortality associated with the closure of large VSD in this risky group. **Methods:** Sixteen infants and children were operated with median age of 12 months, operated by the same surgeon (IU), from January' 2004 to December' 2005 . They were with large VSD of elevated PVR (3.9+0.3) and underwent VSD closure leaving PFO or artificial ASD (5mm). Surgical approach was through right atrium. Post operatively, all the patients were electively ventilated for 36 hours. They were given intravenous dilators (Glyceral Trinitrate + Phentolamine) and oral Sildenafil up to 1mg /Kg, six hourly. Five cases went into acute pulmonary hypertensive crisis postoperatively, and were rescued by Prostacycline Nebulization. **Results**: Sixteen patients had VSD as the primary lesion that underwent operation. The overall early mortality was 6.25% (1/16). There have been no late deaths. **Conclusion**: Closure of large VSD with elevated PVR can be performed, leaving PFO or artificial ASD, with acceptable mortality and morbidity.

Key words:

INTRODUCTION

On the spectrum of congenital heart diseases Ventricular Septal Defect (VSD) is the most common congenital heart defect and surgical closure of VSD is one of the most common open heart procedure performed in paediatric cardiac surgery [1].

Large VSD with large left to right shunts commonly present in infants with respiratory symptoms and poor growth [2]. Large VSD usually results in PVR in later life. The closure of a large VSD is performed in developed countries at an early age before the onset of elevated PVR. It gives excellent long term results. The occurrence of pulmonary hypertensive crisis can contribute to morbidity and mortality. In most medically advanced countries, the pulmonary hypertensive crisis are managed with either sophisticated pharmacological agents such as Nitric acid or mechanical circulatory assist devices such as extracorporeal membrane oxygenation (ECMO) [3,4]. Children with large VSD and elevated PVR are rarely seen in surgically advanced countries. These children are at an increased risk of significant morbidity and mortality even when closure is performed in infancy [5]. To face this problem we planned to leave PFO as such or to make an artificial ASD to reduce the morbidity and mortality associated with surgery on infants and children with large VSD and increased PVR. It is one of the unique studies to leave PFO in a group of patients of VSD with pulmonary hypertension.

PATIENTS AND METHODS

Patients demographics

Sixteen Patients with a large VSD with pulmonary hypertension were operated between January'2004 and December'2005. Twelve patients (75%) were male, while four patients (25%) were female. Age of the patients ranged from 4 months to 10 years (median age: 12 months). 10 patients were infants (Fig 1). Weight of the patients ranged from 4.4 kg to 26 kg (median weight: 6.95Kg). Body surface area (BSA) ranged from 0.25/M2 to 1.00/M2 (Median BSA .365/M2). Patient's demographics are shown in Table 1. Abbreviations and Acronyms are labelled in Table 2.

Pre-operative evaluation

All children had pre-operative two dimensional echocardiography. Doppler evaluation in 5 patients was the sole pre-operative diagnostic test. 11 (68.75%) infants and children underwent cardiac

catheterization and manoeuvres to manipulate the PVR use of 100% oxygen. Preoperative diagnoses with associated lesion are listed in Table 3.

Operative management

VSD closure were done by the author (IU). Routine Cardiopulmonary bypass (CPB), aorta, superior and inferior vein cannulation with moderate hypothermia (28 ^{o} C) were followed.

Cardioplegic arrest with blood based Cardioplegia was used in all the cases. The VSD closure patch was constructed of Sauvage Dacron. (C.R.Bard, Murray Hill, New Jersey USA).

The VSD Patch was tailored after inspection of the defect. The PDA interruption/ligation was done before going on CPB to save lungs from over flooding. The VSD was closed by interrupted suture technique. (Fig.2) 14 patients (78.5%) were intentionally left with an atria level communication. Cardiac Support medication and vasodilators were started before weaning from CPB. Conventional haemofiltration was done during CPB in all cases.

Transthoracic pulmonary artery catheter (9 cases) and transvenous pulmonary catheter (7 cases) was used to measure the PA Pressure.

Post operative management

Patients were ventilated electively up to 36 hours. Discontinuation of Inotropic drugs and intravenous vasodilators was accomplished as quickly as the clinical status allowed.

Follow up evaluation.

Follow up was conducted regularly through echocardiography and clinical evaluation. All 15 patients (93.75%) survived and attended for follow up.

RESULTS

Preoperative Data

A significant difference was found regardind sex of the patients i.e. 12 patients (75%) were male while 4 patients (25%) were female. The median age was 12 months for both groups. The median weight was 6.95kg while BSA was .365/M2.

For entire study population the preoperative catheterization data was obtained on 50% oxygen and 100% oxygen and it was labile. Preoperative catheterization data values are presented in Table 4.

Operative Procedures

All VSD were closed with Dacron patch . All patients were weaned from CPB . One patient (6.25%) had friable myocardium causing difficulty in suturing. One patient (6.25%) had acute cardiovascular collapse after CPB , so we went again on CPB and made the PFO patent. Haemofiltration was done in all patients, having a significant role postoperatively. The PA Transthoracic and Trans venous lines were passed and recorded Pulmonary artery pressure postoperatively. Two patients who had morbidity during weaning were less than 6 months of age, so we decided to close the PFO.

Postoperative data

One patient (6.25%) with age 4.5months with friable myocardium died on first postoperative day due to dysrythmias. 15 patients (93.75%) were electively ventilated for 36 hours in ICU. They were given intravenous dilators i.e Glyceral Trinitrate (GTN), Phentolamine and Phosphodiestrase type V inhibitor (oral Sildenafil) up to 1mg/kg/ 6 hourly through nasogastric tube and remained on Sildenafil , while weaned off from IV dilators. Five cases (31.25%) had acute pulmonary hypertensive crisis and were rescued by prostacycline nebulization .

Postoperative Echocardiography

Fifteen patients (93.75%) were evaluated in follow up and survived. Pulmonary hypertension estimated by Doppler (mean PA Pressure 27 mHg was present in all the survived cases. Four patients (25%) displayed right to left shunting through atrial communication . There was no correlation between the preoperative PVR and the postoperative doppler estimate of pulmonary artery pressure.

Early and late morbidity and mortality

Sixteen children were operated with Dacron patch with interrupted sutures technique closure of VSD. Two patient (12.50%) had closure of PFO. One patient (6.25%) had a acute cardiovascular collapse in operation theatre. Patient was rebypassed and PFO made patent and recovered while other patient with closed PFO died in intensive care unit(ICU). No thrombo-embolic event occurred todate. Mean survival time is 11 months (range: 5 months to 1 year and 11 months). No residual shunt was ducted at VSD patch level in all cases.

Table 1: Patient Demographics

No	Age yrs/mth	Sex	Weight (kg)	Body surface area (BSA)/M 2	Diagnosis	Procedures	Survived
1	06 yrs	Male	20	.75	VSD+PFO+PDA	Patch Closure of VSD+PDA Ligation	+
2.	02 yrs	Male	10	.47	VSD	Patch Closure of VSD	+
3.	1.5 yrs	Male	5.6	.37	VSD+PFO+PDA	Patch Closure of VSD+PDA Ligation	+
4.	05 mth	Male	4.4	.25	VSD+PFO+PDA	Patch Closure of VSD+PDA Ligation	+
5.	10 mth	Female	5.4	.30	VSD+PFO+PDA	Patch Closure of VSD+PDA Ligation	+
6.	1.6 yrs	Male	7.8	.40	VSD+PFO	Patch Closure of VSD	+
7.	1.0 yrs	Male	7.4	.35	VSD+PFO+PDA	Patch Closure of VSD+PDA Ligation	+
8.	1.4 yrs	Male	7.6	.36	VSD+PDA	Patch Closure of VSD+PDA Interruption	+
9.	08 mth	Female	5.8	.37	VSD+PFO+PDA	Patch Closure of VSD	+
10.	1.0 yrs	Female	6.5	.36	VSD+PFO	Patch Closure of VSD	+
11.	07 mth	Male	5.9	.32	VSD	Patch Closure of VSD	+
12.	1.0 yrs	Female	5.5	.33	VSD+PDA	Patch Closure of VSD+PDA Interruption	+
13.	2.0 yrs	Male	8.0	.40	VSD+PFO+PDA	Patch Closure of VSD+PDA Interruption	+
14.	10 yrs	Male	26.0	1.00	VSD+PFO+PDA	Patch Closure of VSD+PDA Interruption	+
15.	10 yrs	Male	7.5	.36	VSD+PFO+PDA	Patch Closure of VSD+PDA Interruption	+
16.	4.5 mth	Male	5.0	.30	VSD+PFO	Patch VSD Closure	-

+ = Survival, - = Mortality

Table-2: Abbreviations and Acronyms

ASD	=Atrial Septal Defect
BSA	=Body surface Area
CPB	=Cardiopulmonary bypass
ECMO	=Extracorporeal membrane oxygenation
ICU	=Intensive Care Unit.
PA	=Pulmonary Artery
PFO	=Patent Foramen Ovale
PDA	=Patent Ductus Arteriosus
PAs/AOs	=ratio of PA systolic pressure to arotic systolic Pressure.
PVR	=Pulmonary Vascular Resistance
Qp/Qs	=The ratio of pulmonary blood flow to systemic blood flow.
SpO2	=Arterial hemoglobin oxygen saturation.
VSD	=Ventricular septal defect.

Table-3: Preoperative diagnoses with associated lesions.

S/No.	Defects	Number of Patients	Percentage
1.	Solitary VSD	02	(12.5%)
2.	VSD+ASD/PFO	03	(18.75%)
3.	VSD+PDA	02	(12.5%)
4.	VSD+ASD+PDA	09	(56.50%)

Table-4: Preoperative catheterization Data VSDs.

Variables	50% Oxygen	100% Oxygen
PAP/AOP	0.60±0.10	0.55±0.10
PVR (Woodunits)	3.9±0.3	3.0±0.5
QP/QS	1.9±0.5	2.3±0.3
SPO2 (%)	93±4.0	96±3.0

Values are expressed as mean ± SD.
PAP/AOP Peak systolic pulmonary artery pressure/peak. Systolic aortic Pressure PVR=Pulmonary Vascular Resistance, QP/QS=Pulmonary blood flow/systemic blood flow, SPO2 (%) = Systemic arterial Saturation.

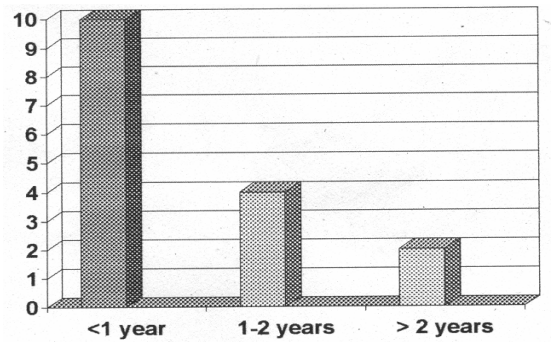

Fig. 1. Number of patients vs Age group

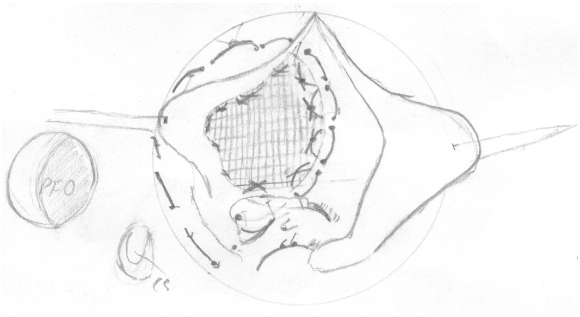

Fig.2 Illustration of VSD Closure, Dacron patch with interrupted suture technique through right atrium leaving PFO patent.

DISCUSSION

Closure of large VSD with pulmonary hypertension carries significant morbidity and mortality even in developed countries.[6] Two dimensional Doppler Echocardiography (DD echo) has got a significant role in diagnosis and primary repair of VSD[7] , but catheterization and angiography is more fruitful regarding PVR. Nitric Oxide use and ECMO rescue has reduced the mortality but on account of morbidity and significant cost, many developing countries can not afford these sophisticated modalities. We recommend to leave the natural inter-atrial communication i.e. PFO or make artificial ASD (5mm) each. This is, by the grace of Allah, Almighty, the natural option for the management of children with pulmonary hypertension and an elevated PVR. It provides a simple, natural physiologic mechanism for unloading the right ventricle during periods of severe pulmonary hypertension whether acute and transient or sustained. According to Knott C et al, even in the surgical management of the Tetrology of Fallot, closure of PFO is a significant variable in mortality [8]. Novick WM and colleagues[9] designed a simple fenestrated flap valve VSD closure patch, known as (Flap valve Double Patch closure of VSD) to reduce the morbidly and mortality associated with surgery on children with a large VSD and increase PVR. They described the use of the flap valve double patch VSD closure technique as a method which could provide surgeon with an inexpensive option for the management of children with pulmonary hypertension and elevated PVR. They have used it in simple and complex defects. They have been able to extubate the children rapidly and thereby diminish the need for prolonged mechanical ventilation. Kannan and colleagues [10] published their results on the closure of a large VSD in Patients with high PVR. The early mortality was 13.1% .Children in this study were older (7.5Year) than those in Novick's study i.e. (4.0year) while PVR in Kannan's study was 7.6±1.8 wood units as compared to 10.5±4.9 wood units in Novick's study. In our study the age of the infants and children was (12 months), while the PVR was 3.9±0.3 wood units in 50%. We have not noticed a higher PVR and so our result with leaving PFO inter-atrial communication as a flap valve with mortality of 1 case (6.25%). We believe that this difference supports our idea, leaving inter-atrial communication provides for a lower mortality with pulmonary hypertension and elevated PVR. Wessel et al [11] outlined strategy of heartlung and heart transplantation in cases of VSD with very high PVR. In our setup, it is almost impossible. We can step wise do this technique for older children with high PVR (10 wood units). In addition Trachte AL et al [12] has concluded that oral Sildenafil is an effective agent for treatment of postoperative pulmonary hypertension and can be used to facilitate weaning of inhaled and intravenous pulmonary vasodilators. In our patients we had used Sildenafil postoperatively with satisfactory results. Surgical closure of VSD is still having superior role than interventional Transcatheter closure.[13].

CONCLUSION

We conclude that with leaving inter-atrial communication (PFO or ASD) in infants and children with pulmonary hypertension and elevated PVR, can undergo operation with acceptable morbidity and mortality. Intermediate survival is expected and long term survival need continuous observation. It is also useful in high risk high flow pulmonary hypertensive with normal PVR infants. We recommend the future treatment strategy i.e. leaving PFO, early extubation, Ionodilators (Phosphodiestrase type III inhibitors i.e. Milrinon) and Phosphodiestrase type V inhibitors i.e. Sildenfil.

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