INTRODUCTION

Ventricular septal defect (VSD) is the commonest congenital heart disease repaired surgically.\(^1\) VSD with left to right shunt gradually results in increased pulmonary vascular resistance (PVR) as the age progresses.\(^2\) PVR of more than 8 wood units had been considered inoperable and VSD closure at this stage may be very hazardous and is associated with high mortality and morbidity.

Closure of VSD should be performed at an early age before the progression of PVR for optimal results.\(^3\) In cases of adult VSDs with low PVR the results of surgical outcome are satisfactory.\(^4\) In Pakistan the patients present late with this problem of increased PVR which may be very hazardous and is associated with high mortality and morbidity.

Closure of VSD should be performed at an early age before the progression of PVR for optimal results.\(^3\) In cases of adult VSDs with low PVR the results of surgical outcome are satisfactory.\(^4\) In Pakistan the patients present late with this problem of increased PVR which may be very commonly encountered in clinical practice. Post operative prognosis depends upon the age and PVR at presentation and pulmonary hypertensive episodes is the major cause of morbidity and mortality.\(^5\) The increased morbidity and mortality range from 22.7 to 50% is due to the pulmonary hypertensive crisis, acute congestive heart failure and acute respiratory failure.\(^6\)\(^-\)\(^11\)

Nitric oxide (NO) and circulatory assist devices (extra corporeal membrane oxygenation - ECMO) are the methods to deal with the problem of pulmonary hypertensive episode. But no withdrawl can lead to rebound pulmonary hypertension while circulatory assist devices are costly for the developing countries like Pakistan.\(^12\)\(^,\)\(^13\) Gene transfer therapy and elastase inhibitors are still under trial.\(^14\)

Since December 2005, this institute is advancing its paediatric cardiac surgical programme in collaboration with International Children Heart Foundation (ICHF). The technique of double flap patch closure of VSD with elevated pulmonary vascular resistance was introduced by the chief surgeon of the organization. The technique has proved beneficial in reduction of morbidity and mortality.\(^15\) During periods of postoperative pulmonary hypertensive crisis, the unidirectional double flap valve act as a pop up for right ventricle resulting in right to left ventricular shunt thus maintaining cardiac output at the expense of oxygen saturation, thereby reducing the risk of early postoperative death. When pulmonary artery pressure gradually falls postoperatively and pressure

ABSTRACT

Objective: To determine the 30 days outcome measured in terms of morbidity and mortality in cases of ventricular septal defect (VSD) with increased pulmonary vascular resistance (PVR) managed with double flap patch closure.

Study Design: Case series.

Place and Duration of Study: Armed Forces Institute of Cardiology (AFIC/NIHD), Rawalpindi, from December 2005 to December 2008.

Methodology: Forty patients with VSD having PVR 9.58 ± 4.33 wood units underwent double flap patch closure. The patch was fenestrated as one half of the expected aortic annulus diameter. A separate flap patch 5 mm larger than fenestration was attached to superior upper one third margins of fenestration. The patch was placed with flap to open towards the left ventricular apex. Modified ultra filtration (MUF) was employed in every case and sildenafil was given postoperatively.

Results: The age of patients ranged from 1 to 28 years with a mean of 6.66 ± 5.70 years. There were 22 males and 18 females. All patients were weaned off from inotropic and ventilatory support as earlier as possible postoperatively with intensive care unit (ICU) stay of 77.15 ± 54.56 hours. Postoperative pulmonary artery pressures were reduced to 42.63 ± 10.86 mmHg as compared to pre-operative pulmonary artery pressures of 88.3 ± 15.2 mmHg. Postoperatively 11 patients with suprasystemic pulmonary artery pressures and desaturation went into pulmonary hypertensive crisis in which immediate 2D echo evidenced the functioning flap valve with right to left shunt. There was only one death (early) out of 40 patients with an overall mortality of 2.5% along with limited morbidity.

Conclusion: Double flap patch is an inexpensive, easy to construct technique with low morbidity and mortality in cases of VSD with raised PVR.

Key words: Ventricular septal defect (VSD). Pulmonary vascular resistance (PVR). Double flap patch.
gradients between the right and left sides of the circulation normalize, the valve closes and prevents a significant left to right shunt.\textsuperscript{16} It acts as a safety mechanism at the time of pulmonary hypertensive crisis.

The study was carried out to determine the early (thirty days) morbidity and mortality in cases of VSD with raised PVR managed with double flap closure.

**METHODOLOGY**

From December 2005 to December 2008, 40 patients having VSD with increased PVR underwent VSD closure with double flap patch. Patients having VSD with PVR more than 6 wood units were included in this study with reversible PVR and the criteria for reversibility was post-oxygen reduction in PVR. Cases included solitary VSDs and VSDs along with other associated anomalies which were managed in the same sitting as well. Cases of VSD operated by simple patch closure were excluded from this study. After clinical assessment all patients were subjected to laboratory biochemical investigations along with chest radiography, electrocardiography, two dimensional doppler echocardiography and cardiac catheterization before surgery.

Routine cardiopulmonary bypass (CPB) with moderate hypothermia was employed for all cases. Cold cardioplegic arrest with blood based cardioplegia was used in every case. The double flap patch VSD closure was constructed of Sauvage Dacron (C.R.Bard, Murray Hill, New Jersey) or Gore-Tex (W.L.Gore and associates, Newark, Delaware) depending upon the surgeon preference.

The VSD patch was tailored according to the size of the defect and fenestrated half the size of expected aortic root diameter. A separate flap patch 5 mm, larger than the fenestration was sewn onto the superior margin of the fenestration along one third of the circumference. A separate tethering stitch was placed at the inferior apex of the flap valve and tied loosely over a Hegar dilator that was of the same size as the fenestration. Thus, the tethering stitch length approximated the diameter of the fenestration. The VSD patch was then sewn into place orienting the patch so that the flap valve was placed on the left ventricular (LV) side and directing the flap so that it would open toward the LV apex. Patch was sewn with continuous or interrupted polypropylene suture (Figure 1). No patient was left with an atrial level communication. Cardiac support medications and vasodilators were started according to the requirements before weaning from CPB. All patients were weaned from CPB successfully and modified ultrafiltration (MUF) was employed in every case. Transvenous pulmonary catheter was passed through internal jugular vein in all cases at the time of induction to monitor postoperative pulmonary artery pressures. All the patients were shifted to ICU.

In the ICU, patients were allowed to be extubated earlier. Inotropic agents and intravenous vasodilators were discontinued according to clinical status. Inotropic support duration was recorded. Sildenafil first dose was given in OT at the time of induction through NG tube (0.5 mg/kg with max upto 3 mg/kg). Its dose was increased in cases of PAP > 50% of systemic pressures. Serial PA pressures were measured in ICU through transvenous PA line and findings were recorded till the patient was shifted from the ICU. Postoperative lowest saturation was noticed in correlation to pulmonary artery pressure for determining shunting.

Statistical analysis was performed using SPSS version 12.0. All data were expressed as mean ± standard deviation, frequency and percentages.

**RESULTS**

Total number of patients were 40 in which there were 22 (55%) males and 18 (45%) were females. The age ranged from 1 to 28 years (mean = 6.66 ± 5.70 years) and weight ranged from 7 to 55 kg (mean = 19.85 ± 9.27 kg). The diagnostic spectrum of anomalies and pre-operative variables are described in Table I and II respectively. All patients were weaned off from ventilatory support in the early postoperative period with mean of 8.61 ± 11.18 hours and mean ICU stay of 77.15 ± 54.56 hours. As compared to pre-operative pulmonary artery pressures 88.3 ± 15.2 mmHg a marked reduction was noticed in pulmonary artery pressures at post-bypass, at 6 hours and 24 hours in ICU (Table III).

All patients were weaned off from inotropic supports within 33.14 ± 30.75 hours. Only one patient died out of 40 patients in early postoperative period with a mortality of 2.5%. He did not have a marked fall in PA pressures inspite of good functioning of double flap patch on echocardiographic evaluation.

The morbidity in early postoperative period was dysrhythmias in 5 (12.5 %) cases that were treated with epicardial pacing and antiarrhythmic pharmacological agents, 2 (5 %) cases had pleural effusion managed with chest intubation and 2 (5 %) had bleeding from the periosteum of sternum who underwent reopening to secure haemostasis.
Double flap patch closure of VSD with elevated pulmonary vascular resistance: an experience at AFIC/NIHD

Table I: The spectrum of cardiovascular anomalies.

<table>
<thead>
<tr>
<th>Diseases</th>
<th>Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Solitary ventricular septal defect</td>
<td>30 (75%)</td>
</tr>
<tr>
<td>VSD + ASD</td>
<td>03 (7.5%)</td>
</tr>
<tr>
<td>VSD + Patent ductus arteriosus</td>
<td>02 (5%)</td>
</tr>
<tr>
<td>VSD + Supra mitral membrane</td>
<td>01 (2.5%)</td>
</tr>
<tr>
<td>Multiple VSD</td>
<td>01 (2.5%)</td>
</tr>
<tr>
<td>VSD + Sub aortic membrane</td>
<td>01 (2.5%)</td>
</tr>
<tr>
<td>VSD + DORV</td>
<td>01 (2.5%)</td>
</tr>
<tr>
<td>VSD + Hypoplastic arch</td>
<td>01 (2.5%)</td>
</tr>
</tbody>
</table>

Total 40

ASD = Atrial septal defect; DORV = Double outlet right ventricle.

Table II: Pre-operative haemodynamic data.

<table>
<thead>
<tr>
<th>Variables</th>
<th>Mean</th>
<th>Std. deviation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic systolic pressure (mmHg)</td>
<td>65.43</td>
<td>± 11.74</td>
</tr>
<tr>
<td>PA systolic pressure (mmHg)</td>
<td>57.31</td>
<td>± 12.47</td>
</tr>
<tr>
<td>PAP / AOP (mmHg)</td>
<td>88.3</td>
<td>± 15.2</td>
</tr>
<tr>
<td>Oxygen saturation (%)</td>
<td>91.53</td>
<td>± 5.84</td>
</tr>
<tr>
<td>PVR (air) wood units</td>
<td>9.79</td>
<td>± 5.50</td>
</tr>
<tr>
<td>PVR (oxygen) wood units</td>
<td>6.81</td>
<td>± 2.48</td>
</tr>
</tbody>
</table>

PAP = Pulmonary artery pressure. AOP = Aortic pressure. PVR = Pulmonary vascular resistance.

Table III: Post-operative change in variables.

<table>
<thead>
<tr>
<th>Variable</th>
<th>Mean</th>
<th>Std. deviation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic cross clamp time (min)</td>
<td>52.03</td>
<td>± 25.59</td>
</tr>
<tr>
<td>Cardiopulmonary bypass (min)</td>
<td>85.82</td>
<td>± 32.07</td>
</tr>
<tr>
<td>PAP/AOP post bypass (mmHg)</td>
<td>66.75</td>
<td>± 13.47</td>
</tr>
<tr>
<td>PAP/AOP at 6hrs in ICU (mmHg)</td>
<td>54.75</td>
<td>± 9.93</td>
</tr>
<tr>
<td>PAP/AOP at 24 hours in ICU (mmHg)</td>
<td>42.63</td>
<td>± 10.86</td>
</tr>
<tr>
<td>Time to extubation (hours)</td>
<td>8.61</td>
<td>± 11.18</td>
</tr>
<tr>
<td>ICU stay (hours)</td>
<td>77.15</td>
<td>± 54.56</td>
</tr>
<tr>
<td>Inotropic duration (hours)</td>
<td>33.14</td>
<td>± 30.75</td>
</tr>
</tbody>
</table>

Postoperatively 11 (27.5%) patients went into pulmonary hypertensive crisis having suprasystemic pulmonary artery pressures with low oxygen saturation in correlation to these raised pulmonary artery pressures depicting right to left ventricular shunt (pop off phenomenon) confirmed by 2D echocardiography. Those patients were kept on ventilatory support along with sedation and muscle relaxation in addition to 100% oxygen inhalation, keeping PCO₂ < 35 mmHg and hemocrit > 0.30. Injection milrinon (0.7-1 µgm/kg) and tablet sildenafil (upto 2 mg/kg/6 hours) were adjusted accordingly. Oxygen inhalation continued for 10 days even after shifting from ICU.

After discharge from the hospital, follow-up consisted of clinical examination and monitoring of PA pressure by echocardiography. Thirty six (90 %) patients reported for follow-up at 4 weeks with marked improvement clinically and on echocardiography. Long-term survival requires continued observation of these patients to determine any progressive pulmonary hypertension.

**DISCUSSION**

Many techniques exists for the management of VSD with elevated pulmonary vascular resistance. Each has its own advantages and disadvantages. The VSD closure necessitates adequate sizing and shape of the patch.17

Charles P. Bailey applied flap-valves made of a compressed ring of polyvinyl sponge in 8 patients with atrial and ventricular septal defects in whom the conventional closure appeared too risky because of severe pulmonary hypertension. Seven of the 8 patients survived.18

Interatrial communication has been left in the form of patent foramen ovale (PFO) or made in cases of VSD with raised PVR. In the study by Inamullah et al. 16 patients had VSD as the primary lesion that underwent closure of large VSD with elevated PVR leaving PFO or artificial ASD, with acceptable mortality and morbidity. The overall early mortality was 6.25% (1/16) and there were no late deaths.19

A one-way, valved, atrial septal patch to serve as an artificial one-way foramen ovale has been tested. By permitting right-to-left shunt, this device decompresses the failing right ventricle and maintains systemic cardiac output. This patch provides the same hemodynamic benefits as an ASD, together with the added advantages of a controlled opening pressure and the ability to close, without the need for a subsequent invasive procedure.20

Zhou et al. described the closure of VSD with severe pulmonary hypertension by using unidirectional valved patches (UVP). Total 4 out of 24 patients (16.66%) died in early postoperative period.21

The first modification was by Novick et al., who used UVP from sovage dacron. The weight of the child was used to decide the size of fenestration. Out of the 18 patients there were no early deaths and only one late death occurred.15 Later on Novick et al. used a modified technique with a fenestration size of half of the expected aortic annulus diameter for each patient. There were 7 early deaths and 7 late deaths. Postoperative echocardiography studies revealed persistent pulmonary arterial hypertension (PAH) in 25 patients.22 The same technique was practiced in this study in which only one early postoperative death was observed.

Zhang et al., used the aortic homograft along with the attached mitral leaflet for the UVP construction. They have 2 early deaths due to raised PAH out of 27 patients (7.40%).23

Talwar et al., experienced modified UVP technique for VSD closure by using a patch of knitted polyester fabric. Out of 21 patients no early or late deaths were noticed.24

Afrasiabi used a Gore-Tex patch with a 5-8 mm longitudinal slit in the center in cases of VSD with raised PVR. A piece of pericardium was sewn around the slit on one side of the patch, except for the upper quarter and defect closure was done in such a manner that the pericardial aspect was placed on the left ventricular side.
to allow right-to-left shunting. Two patients (12.5%) died in the early postoperative period due to frequent episodes of pulmonary hypertensive crisis. It seems to be a promising technique to decrease morbidity and mortality in severe pulmonary arterial hypertension. Double flap patch is an inexpensive option for the management. Utilizing this technique, we were able to extubate the patients rapidly and thereby diminish the need for prolonged mechanical ventilation and ICU stay. This reduces the use of extra resources.

Improvements in anaesthesia techniques and myocardial protection, modified ultrafiltration and medicines like Sildenafil are effective in reducing the pulmonary hypertension. Oral Sildenafil can be used to facilitate weaning off inhaled and intravenous pulmonary vasodilators. The role of Sildenafil has been established among a group of postoperative children with large septal defects with moderate to severe pulmonary hypertension. It is effective oral therapy as it is safe and easily applied but determination of its efficacy, safety and optimal dosage further studies are necessary. In this study Sildenafil was used postoperatively with dose adjustment according to pulmonary artery pressures postoperatively.

CONCLUSION

Ventricular septal defect with elevated PVR can be managed with double flap patch. The latter is easy to construct, is inexpensive and has a role as a pop off phenomenon during episodes of pulmonary hypertensive crisis postoperatively resulting in low morbidity and mortality.

REFERENCES


