Flap valved closure of ventricular septal defects with increased pulmonary vascular resistance

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Abstract

Closure of ventricular septal defect (VSD) in children with elevated pulmonary vascular resistance (PVR) is associated with significant morbidity and mortality with pulmonary hypertensive (PH) episodes being a major postoperative problem. Flap valved closure of VSD is reported to decrease morbidity and mortality. We report our experience of closure of a VSDs in patients with severe PH, using a valved patch in an effort to reduce the risk of operation. Eighteen consecutive patients with a large VSD with severe PH (mean PVR > 8 Wood units) underwent flap valved closure (as described by Novick et al.) of VSD during a one-year study period. The mean age at surgery was 8.3 ± 3.9 years (range: 3–13 years). Mean PVRI was 13.02 ± 4.05 Wood units. In-hospital 30-day mortality was 5.6% (1/18). Mechanical ventilation time averaged 11.6 ± 8.1 hours. Postoperative pulmonary artery pressures were significantly reduced. Four patients had PH crisis postoperatively. Obvious opening and closing of the flap valve was detected by echocardiography in eight patients. There were no late deaths due to cardiac causes. Closure of a large VSD in patients with severe pulmonary hypertension could be performed with low morbidity and mortality when a flap valve patch was used.

Keywords: Ventricular septal defect; Pulmonary vascular resistance; Flap valve patch; Cardiopulmonary bypass; Low cardiac output

1. Introduction

Congenital heart disease with septal defects and a large left to right shunt often causes pulmonary artery (PA) hypertension. The course of the disease is variable and depends on the size of the defect, magnitude of the left to right shunt, and the pulmonary vascular response to increased pulmonary blood flow and pressure. PA pressure is hyperkinetic in the early phase but may eventually become a fixed elevation due to a fixed increase of pulmonary vascular resistance (PVR) [1]. They clinically manifest with decreased exercise tolerance, cyanosis, congestive cardiac failure, often hemoptysis, and finally death (Eisenmenger’s syndrome).

Closure of a ventricular septal defect (VSD) with increased PVR is associated with significant morbidity and mortality [2, 3]. Acute congestive heart failure, pulmonary hypertensive (PH) crisis, and acute respiratory failure are the principal causes of postoperative death. In some patients the postoperative increase of PVR may be more marked, may regress slowly, may show intermittent spontaneous or precipitated crisis-like exacerbation, and may require prolonged ventilation and specific strategies to maintain a lower PVR [4].

Many patients in developing nations come at a stage of near eisenmengerization and their management perioperatively is especially difficult with a high mortality and morbidity. We report our experience of closure of a VSDs in patients with severe PH, using valved patch in an effort to reduce risk of operation.

2. Patients and methods

2.1. Patients

Between August 2008 and July 2009, 18 patients presenting to our unit with a VSD and increased PVRI (mean PVRI > 8 Wood units) were included in the study. The patients were evaluated preoperatively by history and physical examination, chest radiography, electrocardiography, transthoracic echocardiography (TTE), and cardiac catheterization.

Complete hemodynamic data were determined by cardiac catheterization. Pulmonary and systemic flow and resistance were measured at rest and 100% oxygen. Color M-mode echocardiography was done to assess the direction of shunting across VSD. The decision for surgery was based on the presence of any augmentation of left to right shunt or increase in pulmonary vascular blood flow following sublingual nitrate administration on preoperative echocardiography even if the patients showed an unfavorable PVR on formal hemodynamic testing in the cardiac catheterization laboratory. The study was approved by the Hospital...
Research Ethics Committee, and written informed consent was obtained from the parents of each child.

2.2. Surgical procedure and cardiopulmonary bypass

Surgery was performed with standard cardiopulmonary bypass (CPB) with moderate hypothermia and cold blood cardioplegia was used in all patients. Inj phenoxybenzamine (0.5 mg/kg i.v.) was given prior to CPB. Zero balance ultrafiltration was done throughout CPB. The operative approach was transatrial.

A patch was constructed using the technique described by Novick et al., using a Gore-Tex patch approximately as large as the defect to be closed. A fenestration that was half of the expected aortic annulus diameter was made in the patch. Following this, a separate flap patch at least 2 mm larger than the fenestration was constructed and 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 the same size as the fenestration. This tethering stitch length approximated the diameter of the fenestration. The VSD patch was sewn into place orienting the patch in such a way that the flap valve was placed on the left ventricular (LV) side with the flap opening towards the LV apex to avoid subaortic obstruction. Patent foramen ovale (PFO) was left open in all patients. Tricuspid valve competence was checked and repaired if necessary.

2.3. Weaning from CPB

Prior to discontinuation of CPB, dobutamine 3 μg/kg/min, nitroprusside 0.5–2 μg/kg/min, and the phosphodiesterase inhibitor milrinone 0.35 μg/kg/min were started. Inj phenoxybenzamine 0.5 mg/kg/day were given as infusion for 48 h followed by oral formulation. Left atrial (LA) line was placed if required and systemic vasoconstrictors were started through it. After weaning from CPB, modified ultrafiltration was used in all the patients. Epicardial echocardiography was used to assess the degree of shunting across the fenestration.

2.4. Postoperative management

Pulmonary and systemic artery pressures were monitored continuously. Mild to moderate hypocarbia was maintained during mechanical ventilation. Serial postoperative TTE was used to assess the degree of shunting across the fenestration. Once the patients are fully awake and hemodynamically stable, they were extubated. Inhaled nitroprusside and milrinone nebulisation were given intermittently (every 2 hours) or when PA pressure surges occur. Inotropes were weaned-off once patients were hemodynamically stable. Milrinone infusion was stopped within 36–48 hour provided PA pressure surges stopped or no right to left shunting across PFO. Oral phenoxybenzamine was administered at 48 hours and sildenafil (1 mg/kg 4 hourly) was administered on the first postoperative day (POD). Patients were monitored for seven days in ICU/intermediate care for PH crisis.

2.5. Follow-up

All patients were followed-up at regular intervals (every three months) with clinical examination, and TTE to estimate the PA pressure, assess the right ventricular function and to demonstrate any right-to-left shunting.

2.6. Statistical analysis

All data are expressed as mean ± standard deviation. Continuous variables at baseline and following surgery were compared with unpaired t-test. A P-value of 0.05 or lower on any test was considered to indicate statistical significance. Data were analyzed using SPSS software (SPSS Inc, Chicago, IL, USA).

3. Results

3.1. Patient characteristics

The mean age at surgery was 8.3 ± 3.9 years (range: 3–13 years) and female to male ratio was 1.25:1. One patient with PVR 13.3 Wood units had Down’s syndrome.

3.2. Preoperative data

A perimembranous VSD was diagnosed in 80% of patients. Muscular VSD was seen in 20% of patients. Cardiac catheterization showed the ratio of systolic pulmonary artery pressure to systolic systemic arterial blood pressure ranged from 0.8 to 0.9 (mean: 0.85). Mean PVR was 13.02 ± 4.05 Wood units (range: 8.5–23 Wood units) and after oxygen administration was 8.34 ± 2.5 Wood units. PVR was more than 10 Wood units in 11 patients and was >15 Wood units in four patients. The ratio of pulmonary artery flow to systemic arterial blood flow (Qp/Qs) ranged from 0.5 to 2.0 (mean: 1.44 ± 0.36) and increased to a mean of 1.91 ± 0.34 after oxygen administration, and was <1.5 in eight patients. Mean pulmonary artery pressure (MPAP) ranged from 57 to 80 mmHg (mean: 68.8 ± 9.7 mmHg) and was >70 mmHg in nine patients. Arterial oxygen saturation ranged from 88 to 99% (mean: 95.2 ± 3.04%). Preoperative catheterization data are summarized in Table 1.

3.3. Operative data

Eighteen patients underwent flap valved closure of VSD out of which four patients could not be weaned-off CPB initially and required placement of LA line, through which systemic vasoconstrictors (noradrenaline and vasopressin) were started and then patients could be weaned from CPB.

<table>
<thead>
<tr>
<th>Variables</th>
<th>Mean value</th>
</tr>
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<tbody>
<tr>
<td>PVR1</td>
<td>13.02 ± 4.05</td>
</tr>
<tr>
<td>Qp/Qs</td>
<td>1.44 ± 0.37</td>
</tr>
<tr>
<td>SpO2 (%)</td>
<td>95.2 ± 3.04</td>
</tr>
<tr>
<td>PAs/Aos</td>
<td>0.88 ± 0.5</td>
</tr>
</tbody>
</table>

PAs/Aos, ratio of pulmonary artery systolic pressure to aortic systolic pressure; PVR1, pulmonary vascular resistance index; Qp/Qs, ratio of pulmonary blood flow to systemic blood flow; SpO2, arterial hemoglobin oxygen saturation.
One patient with PVR of 9.3 Wood units had a persistent rise in PA pressure >70% of systemic pressure, inspite of maximizing pulmonary vasodilators and had significant LV dysfunction. She had her chest kept open and was shifted to the ICU.

3.4. Postoperative period

Mechanical ventilation time ranged from 8 to 36 hours (mean: 11.6±8.1 hours). Three patients with a PVR of 18.43, 8.8 and 15 Wood units had an episode of PH crisis on the day of surgery and were managed with administration of inhaled pulmonary vasodilators, oxygen administration, and support with inotropic and vasodilator drugs.

One patient with PVR of 9.3 Wood units required splinting open of the sternum. She had persistent elevation of the PA pressure in the postoperative period and developed repeated episodes of PH crisis on first POD. She developed intractable supraventricular tachycardia followed by low cardiac output (LCO) and expired on the second POD.

Three patients with a PVR of 18.43, 9.3 and 18.6 Wood units, respectively went into supraventricular tachycardia on the day of surgery and were managed with administration of amiodarone as a bolus followed by infusion. Six patients developed right lower lobe collapse of which three required pleural drain insertion for concomitant effusion and two required bronchoscopy. One patient with PVR of 18.43 Wood units had an episode of PH crisis on the third POD, which was triggered by right lower lobe collapse due to bronchomalacia requiring reintubation. The patient underwent tracheostomy on the eighth POD and was in the ICU for one month and was later discharged at their request. She expired after 48 days due to bronchomalacia which required stenting but her parents refused treatment for it. The patient with Down’s syndrome could not be weaned-off the ventilator due to upper airway obstruction caused by a large tongue. He required a tracheostomy on the second POD. Take down of tracheostomy was done on 10th POD and he was discharged on the 14th POD. The other patients had an uneventful postoperative course.

3.5. Echocardiographic assessment

Eight patients had a right-to-left shunt through the valved patch as determined by TTE (eight patients up to the second POD, one patient on the third POD and no patients up to the seventh POD). Three patients had right to left shunting across the PFO. Postoperative TTE results are summarized in Table 2. All patients had preserved right ventricular function (by visual estimation on echocardiography) and none of the patients had significant tricuspid regurgitation.

3.6. Clinical outcome

Thirty-day mortality was 5.6%. One patient with PVR of 18.43 Wood units expired after 48 days due to bronchomalacia. There have been no subsequent deaths on follow-up. At a mean follow-up of nine months (range: five to 12 months), one patient with PVR of 13.56 Wood units had TTE evidence of moderate PH at three months, postoperatively. Sildenafil and phenoxybenzamine were continued for the next two months and were stopped when follow-up PA pressure was persistently low. The others were asymptomatic with normal or near normal PA pressure. Postoperative hemodynamics are given in Table 3.

We compared our data to a historical control of 18 patients with PVRI >8 Wood units, where the mortality was 12.6% and the mean ventilation time was 86±12 hours.

4. Discussion and conclusions

Historically, surgical closure of a large VSD with severe PH was associated with a high mortality rate [4, 5]. Even in the present era, postoperative PH remains a significant risk factor for morbidity and mortality.

Acute congestive heart failure, PH crisis, and acute respiratory failure are the principal causes of postoperative death. CPB, infusion of protamine, and other factors that could cause the release of vasoactive substances, such as thromboxane A2 and catecholamines could result in pulmonary vasoconstriction and acute PH crisis [6, 7].

Creation of an intracardiac shunt to prevent right ventricular failure has been used with success. Novick and colleagues used a flap valved double-patch for closure of VSDs in children with increased PVR [3, 8]. The double patch technique provides a simple physiological mechanism for unloading the right ventricle during periods of severe PH, whether acute and transient or sustained. The modification allowed more patients to be extubated within 48 hours and with lower operative mortality. The flap valve allows early extubation and prevents supra-systemic pulmonary pressures that can be rapidly fatal in the postoperative period and allows patients to ride through PH crisis without the need for prolonged intubation and multiple maneuvers albeit at the cost of transient desaturation, while maintaining adequate systemic cardiac output which is tolerated well until the PH crisis is resolved.

In our study, all patients were children with high PVR. Thirty-day mortality was 5.6%. One patient with a PVR 9.3 Wood unit died on the second POD with frequent episodes of PH crisis and intractable supraventricular tachycardia followed by LCO. Early extubation was possible in most of our patients thereby diminishing the need for prolonged mechanical ventilation. In addition, prolonged stay in the

<table>
<thead>
<tr>
<th>Time</th>
<th>No right to left shunt (cases)</th>
<th>Right to left shunt (cases)</th>
</tr>
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<tbody>
<tr>
<td>Day 2</td>
<td>10</td>
<td>8</td>
</tr>
<tr>
<td>Day 7</td>
<td>17</td>
<td>0</td>
</tr>
<tr>
<td>Three months</td>
<td>16</td>
<td>0</td>
</tr>
<tr>
<td>Six months</td>
<td>16</td>
<td>0</td>
</tr>
<tr>
<td>Nine months</td>
<td>14</td>
<td>0</td>
</tr>
</tbody>
</table>

Table 3. Preoperative and postoperative hemodynamic status

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Before repair</th>
<th>After repair</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>MPAP (mmHg)</td>
<td>68.8±9.67</td>
<td>34.4±11.42</td>
<td>0.0001</td>
</tr>
<tr>
<td>PAs/Aos</td>
<td>0.88±0.5</td>
<td>0.52±0.2</td>
<td>0.013</td>
</tr>
<tr>
<td>SpO2</td>
<td>95.2±3.04</td>
<td>97.9±2.4</td>
<td>0.005</td>
</tr>
</tbody>
</table>

MPAP, mean pulmonary artery pressure; PAs/Aos, ratio of pulmonary artery systolic pressure to aortic systolic pressure; SpO2, arterial hemoglobin oxygen saturation.
ICU is associated with an increase in septic complications [9] and other adverse events. Early extubation reduces the use of resources that are already limited in a number of developing countries.

Severe reactive postoperative PH may occur despite a technically successful operation and active conventional management, including the administration of high inspired oxygen, hyperventilation, sedation, neuromuscular blockade, and support with inotropic and vasodilator drugs [6].

When these therapies fail, patients can be considered for inhaled nitric oxide and extracorporeal life support (ECLS) which ensures adequate oxygenation and systemic perfusion in the face of a markedly elevated PVR and may allow time for the PH crisis to resolve [10].

PH after closure of a large VSD continues to cause significant morbidity and mortality even in industrialized countries. The use of nitric oxide and extra corporeal membrane oxygenation rescue has reduced the mortality, but with significant cost and patient morbidity. These expensive and sophisticated modalities are not available in many countries throughout the world [10].

Patients with VSD with severe PH who would otherwise be considered for lung transplantation can undergo conventional cardiac surgery using flap valve method of VSD closure in a significant subset. Reports show that patients with PH improved their hemodynamics and exercise capability after closure of the septal defect. It implies that pulmonary vascular remodeling might take place but requires long-term follow-up [11].

We conclude that a valved patch in cases of severe PH can undergo operation with reasonable morbidity and mortality in the absence of sophisticated pharmacological or mechanical intervention.

References


