Unidirectional valved patch closure of ventricular septal defects with severe pulmonary arterial hypertension

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Abstract

Delayed presentation of ventricular septal defect (VSD) is common in developing countries. Such patients often have severe pulmonary arterial hypertension (PAH), which increases post-operative morbidity and mortality. To address these problems, we used our technique of unidirectional valved patch (UVP) for closure of VSD. Between January 2006 and December 2010, 17 patients (age 2–23 years, median 9 years) with a large VSD and severe PAH underwent VSD closure with UVP. Pre-operative mean indexed pulmonary vascular resistance (PVRI) was 10.9 ± 2.2 Wood units and mean pre-operative systemic saturation was 93.4 ± 2.6%. Shunt was bidirectional in 15 patients and predominantly right to left in two. After VSD closure, intra-operative transoesophageal echocardiography revealed a right to left shunt across the patch in three patients 2, 7 and 9 years of age who had pre-operative PVRI of 9.5, 9.8 and 11.1 Wood units, respectively. There were no in-hospital deaths and all patients had uneventful recovery. Mean follow-up was 30 ± 14.7 months and all patients are well without cyanosis. Echocardiography showed no shunt across the patch and all have systemic saturation >95%. We conclude that UVP is a promising technique in patients with large VSD and severe PAH.

Keywords: Ventricular septal defect • Pulmonary hypertension • Pulmonary vascular resistance

INTRODUCTION

In contrast to developed countries, delayed presentation and diagnosis of ventricular septal defect (VSD) is not uncommon in developing countries; therefore, early and late results are affected by development of pulmonary arterial hypertension (PAH) [1, 2]. These patients may require advanced pharmacological agents or even extracorporeal membrane oxygenation (ECMO) in early post-operative phase, but these are either unavailable or are too expensive [3, 4]. Surgical alternatives include leaving an inter-atrial communication or performing VSD closure with a fenestrated VSD patch or a unidirectional valved patch (UVP) [3–8]. In an earlier publication [5], we described our own technique of VSD closure, using a UVP. As our experience has grown with increasing follow-up, we are now in a position to present more detailed early- and mid-term results, using this technique.

PATIENTS AND METHODS

Between January 2006 and December 2010, 17 patients (Table 1) with VSD and basal indexed pulmonary vascular resistance (PVRI) of >8 Wood units underwent VSD closure with a fenestrated UVP. Hospital records of these patients were analysed in this retrospective study. Study protocol was approved by the institute Ethics committee with waiver of individual patient consent. Pre-operative data consisted of clinical history, physical examination, chest X-ray, electrocardiography, echocardiography and cardiac catheterization with complete haemodynamic data and oximetry at rest and following administration of high-flow oxygen.

Out of 17 patients, 12 were male. None had Down syndrome. Mean age was 9.2 ± 5.4 years (range, 2–23 years, median 9 years). Mean weight was 22 kg (median 15 kg, range, 8–45 kg). Recurrent respiratory tract infection and tachypnoea were chief complaints. Fifteen out of 17 were in NYHA class-III. None had cyanosis or haemoptysis. Chest X-ray revealed mild cardiomegaly in 5 patients and no cardiomegaly in 12 patients with signs of severe PAH. Echocardiography revealed a predominant right to left shunt in 2 patients and bidirectional shunt in remaining 15. A peri-membranous VSD was visualized in 15 patients. One patient had one peri-membranous and one additional muscular VSD. Another patient had two muscular VSDs. Four patients each had an associated ostium secundum atrial septal defect, and a patient ductus arteriosus, which were simultaneously repaired.

Pre-operative mean PVRI was 10.9 ± 2.2 Wood units (range, 8.7–16.1, median 10.6 Wood units). Following oxygen administration, PVRI decreased in all patients with an increase in left to right shunt. Mean PVRI following oxygen inhalation was 3.72 ± 1.98 Wood units (range, 1.3–9.8, median 4.2 Wood units). Mean
inotropes were weaned off. Post-operative transthoracic phenoxybenzamine and sildenafil were administered at the time of aortic cannulation. After cardiopulmonary bypass (CPB). Phenoxybenzamine (1 mg/kg) or milrinone prior to weaning from CPB. In the last 10 patients, sildenafil (5 mg/kg) was started after arrival in the intensive care unit (ICU) through the nasogastric tube. Following extubation, oral angiotensin converting enzyme inhibitors or phosphodiesterase inhibitors were administered and inotropes were weaned off. Post-operative transhysteric

d eventually proceeded with surgical therapy depending on the nature of the heart defect.

Decision to perform surgery was made by the surgeon and cardiologist following informed consent and acceptance of risk by the patient's family. Intra-operative transoesophageal echocardiography was performed after induction of anaesthesia and VSD was assessed. All patients were operated under standard anaesthesia and supported with dobutamine (5–10 µg/kg/min), nitroglycerine or nitroprusside (0.5–10 µg/kg/min), fentanyl (0.5 µg/kg) and vecuronium (0.1 mg/kg) for haemodynamic stabilization. The field was then prepared for sternotomy and CPB was instituted. A median sternotomy was performed and the aorta and innominate veins were cannulated. Patients 1, 2, 4 and 7 had associated atrial septal defect; Patients 9, 11, 13 and 17 had associated patent ductus arteriosus. The size Dacron patch is selected and a fenestrated (B) patch is folded upon itself in such a way that after it was folded, the dimensions were adequate for closure of septal defects in patients with severe pulmonary hypertension.

Table 1: Haemodynamic characteristic of patients undergoing valved patch VSD closure

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Basal SpO2 (%)</th>
<th>Basal PVRI (Wood units)</th>
<th>PVRI post O2 (%)</th>
<th>PVRI (Wood units)</th>
<th>Post O2 PVRI (Wood units)</th>
<th>Post O2 O2 (mmHg)</th>
<th>PASP post O2 (mmHg)</th>
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<th>Qp/Qs</th>
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Patients 1, 2, 4 and 7 had associated atrial septal defect; Patients 9, 11, 13 and 17 had associated patent ductus arteriosus.

SpO2: saturation; PVRI: indexed pulmonary vascular resistance; PAS: pulmonary artery systolic pressure; PADP: pulmonary artery diastolic pressure; Qp: pulmonary flow; Qs: systemic flow; Ao: aorta; LA: left atrium.

*Right to left shunt.

Figure 1: The technique of a unidirectional valved patch: (A) an appropriate size Dacron patch is selected and a fenestrated (B) patch is folded upon itself at the dotted line; (C) the patch is sutured (outer dots) to close the defect (D) mechanism of a right to left shunting through the patch in situ (arrow). The suture line has been removed for clarity. RV: right ventricle; LV: left ventricle. Reproduced with permission from: Talwar S, et al. Unidirectional valved patch for closure of septal defects in patients with severe pulmonary hypertension. Ann Pediatr Cardiol 2008;1:114–9. Copyright Medknow Publications.

echocardiography was used to assess the degree of shunting across the patch. All patients were followed up at 1, 3, 6 months and yearly thereafter. Follow-up consisted of regular clinical examination, pulse oximetry to measure oxygen saturation and echocardiography at regular intervals. Detailed cardiac catheterization is planned in a follow-up study.

**STATISTICAL ANALYSIS**

All pre-operative and post-operative data are reported as range and mean ± standard deviation. Median values are also reported.
RESULTS

All patients were uneventfully weaned off CPB. Mean aortic cross-clamp time was $32.2 \pm 5.7$ min (27–45 min, median 30 min) and mean CPB time was $52.2 \pm 11.4$ min (40–87 min, median 50 min).

Mean systemic saturation at the time of termination of CPB was $93.3 \pm 4.3\%$ (range, 84–100\%, median 93\%). Three out of 17 had a right to left shunt intra-operatively on transoesophageal echocardiography and they had episodes of systemic desaturation after termination of CPB and in the ICU. As described in Table 1, these patients were 2 years (Patient ID 13), 7 years (Patient ID 2) and 9 years (Patient ID 15) of age who had a pre-operative PVRI of 9.5, 9.8 and 11.1 Wood units, respectively. None of the patients required nitric oxide or ECMO support. There were no early deaths. Mean duration of mechanical ventilation was only $7.6 \pm 2.7$ h (range, 5–16 h, median 7 h). The mean systemic saturation in ICU was $96.3 \pm 4\%$ (87–100\%, median 98\%). The same three patients (18%) had episodes of systemic desaturation in the ICU, the lowest saturation being 87\%. These were documented as being due to a right to left shunt through the UVP on transthoracic echocardiography. These were managed by supplemental oxygen without the need for re-instituting mechanical ventilation. The inotropes could be weaned off on first post-operative day in all patients. Mean ICU stay was $1.9 \pm 0.3$ days (1–3 days, median 2 days). Mean hospital stay was $5.1 \pm 1.3$ days (median 5 days, range, 4–10 days).

There were no late deaths. Mean follow-up was $30 \pm 14.7$ months (range, 4–51 months). All patients were in NYHA class-I up except one 23-year old male patient (Patient ID 5 in Table 1) who was NYHA class-II. No patient reported cyanosis or required re-intervention.

DISCUSSION

Previous reports have shown that results after closure of a large VSD with PAH are dependent upon the Heath-Edwards classification at the time of closure [1]. VSD closure in children with elevation in PVR preoperatively that decreases after closure of the defect has been described [2, 9]. Kanan et al. [9] recommended VSD closure in such patients. Castaneda et al. [2] reviewed 55 such patients and found that at 1 year after surgery, the PVR fell but remained above normal in most patients. Hallidie-Smith et al. [10] reported 36 patients with VSD and PVR of at least 8 Wood units. Twenty-eight of the 36 patients survived after VSD closure. At cardiac catheterization, 18 of 25 patients had normal to moderately elevated pulmonary artery pressure, and 9 had PVR of 3 Wood units or less. Recent reports in patients with primary PAH who have improved haemodynamics with decreased pulmonary pressures and improved exercise capacity following administration of high-dose calcium channel blocker verapamil [11] and phosphodiesterase inhibitors and prostacyclin [12] suggest that pulmonary vascular remodelling may take place in children. This seems to make VSD closure an attractive option even with elevated PVRI.

Mortality and morbidity for patients with VSD and PAH have been high in the past and even in the present era [2, 6]. Following CPB and later following administration of protamine, vasoactive substances such as thromboxane A2 and catecholamines are released, which aggravate PAH and respiratory failure, which may be the cause of death in these patients [13].

Multiple surgical strategies have been described to manage such patients. One is prior pulmonary arterial banding [14] but it carries high morbidity and is always associated with a need for a later definitive surgery and its potential complications. Most surgeons leave an inter-atrial communication to decompress the right atrium and right ventricle during pulmonary hypertensive crisis. However, shunting across this communication is often unreliable with the potential for a left to right shunt when the pulmonary artery pressures decrease. Another approach is a fenestrated VSD patch that converts a large VSD into a small VSD. The fenestration allows for a right to left shunt in the case of pulmonary hypertensive crisis. With sustained PAH, the fenestration would allow for a right to left shunt as in case of UVP but once the pulmonary artery pressures decrease it has the potential for left to right shunt with associated complications such as infective endocarditis and systemic embolism and need for subsequent surgical or interventional closure [4]. A UVP has been described for closure of large VSDs with borderline operability [4–8]. The patch is designed to function like a flap mechanism with unidirectional opening that allows a right to left shunt during an episode of pulmonary hypertensive crisis but closes and does not allow a left to right shunt once the pulmonary artery pressures come down. Thus, it prevents acute right ventricular failure and death from refractory pulmonary hypertension.

The UVP was first described by Zhou et al. [8]. They used a Dacron patch with a fenestration 0.5–1 cm slightly off centre with pericardium attached to three sides of the Dacron patch. Novick et al. [3, 4] reported their technique in 1998 and followed it up with their experience with the same technique in 2005. Another modification was proposed by Zhang et al. [8] who used an aortic homograft with attached anterior mitral leaflet to construct the UVP. All techniques described above suffer from the limitation that they are often time-consuming. The sizing may be inaccurate, and fenestration may be misplaced if the patch is constructed before inspection of the defect. When the patch is constructed after cardioplegic arrest, it adds to the cardioplegia time. To minimize this, we devised an innovative way of constructing the UVP [5] that takes <30 s. The median ischaemic time was 30 min and median CPB time was 50 min in our patients, which is fairly close to the routine ischaemic and CPB times for an isolated VSD closure.

Results with UVP have been gratifying. It is difficult to compare the results of various techniques as patient populations are heterogeneous and the number of patients is small. In our study, none of the patient had any complication with UVP and all patients could be extubated uneventfully with a median time to extubation 7 h. Median ICU stay was 2 days and median hospital stay was 5 days, which is comparable to our other routine patients. No patient developed any significant complication after surgery. At follow-up, most of the patients were in NYHA class-I and were off medications. None required any re-intervention. There was no death. This technique of UVP is safe, effective and easily reproducible in our experience.

Long-term survival in patients with severely elevated pulmonary vascular resistance and pulmonary artery pressures is controversial as discussed above and it is not possible to conclusively prove that a UVP actually decreases the mortality and morbidity, unless there are prospective randomized trials with pre-operative and post-operative resting and exercise oximetry with demonstration of direction of shunt by echocardiography and quantification of shunt in immediate and late post-operative period. It is however difficult to justify these trials for ethical reasons. One of
the limitations of our study is the lack of availability of long-term echocardiographic and cardiac catheterization data. We expect to shortly provide more data in this direction. Our findings are in sharp contrast to a recent study from China [15] in which 876 patients with VSD and PAH underwent VSD closure. In 195 of these, a UVP was used to close the VSD. The authors performed a multiple logistic regression analysis with propensity score matching, and found that in the 138 propensity-matched pairs, there were no significant differences in early and late survival. Whether these findings hold true for our patient population will be known only after more follow-up of our patients.

CONCLUSION

UVP for closure of VSD helps in tiding over immediate post-operative period in patients with borderline operability. Early- and mid-term results of this technique are promising but long-term results are awaited. The use of UVP in patients with established Eisenmenger's syndrome needs further evaluation.

Conflict of interest: none declared.

REFERENCES


eComment. An oval-shaped unidirectional check-valved patch for treating ventricular septal defects

Authors: Murat Ugurluclan, Ahmet H. Arslan, Yahya Yildiz and Sertac Cicik
Anadolu Medical Center Hospital, Cardiovascular Surgery Clinic, Istanbul, Turkey
doi:10.1093/icvts/ivs145
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We read with great interest the manuscript of Talwar et al. [1], in which the authors presented their experience of ventricular septal defect (VSD) closure with a unidirectional valved patch in patients with severe pulmonary hypertension. We would like to comment on a few points of the authors’ paper.

As a referral centre for the region, we receive similar patients, i.e. who are still operable but are high risk candidates for VSD closure due to increased pulmonary vascular resistance, and we use a similar technique. All patients receive detailed echocardiographic examinations and cardiac catheterization studies preoperatively. As soon as the patients are prepared for operation, a transesophageal echocardiography probe is inserted. The dimensions, shape, and boundaries of the VSD are once again verified. A surgeon prepares the valve patch while the other starts the operation so that the delay during cardiac arrest is minimized. A hole is created on a square Dacron patch with an aortic punch. Another ‘U’ shaped Dacron patch is sewn on this patch 4–5 mm away from the margins of the fenestration. The open end of the ‘U’ is left unseen. The exact size of the patch is adjusted when the VSD is explored. Most often, although three-dimensional, the VSDs are round or oval. An oval-shaped patch has smooth edges and is also easy for VSD closure. Thus we usually trim the reconstructed patch into a round or oval-shape. The authors used a folded, fenestrated patch [1]. Although it seems easier and faster to construct such a patch, size adjustment, if needed, does not seem easy, since trimming may encounter the suture lines. Moreover, since the fenestration is directed at the left ventricular apex, the sharp margin of the patch may be faced towards the tricuspid or semilunar valves, which may perhaps have valvaral consequences. Additionally, when open, the valve of the patch may lead to outflow tract obstruction and leaflet damage in the long term.

Another point is regarding the postoperative management of the patients. Authors stated that none of their patients had required nitric oxide during the intensive care unit stay [1]. In our practice we routinely administer inhaled nitric oxide to facilitate postoperative recovery in pulmonary hypertensive patients. In the manuscript, mean systemic saturation was 96.3±4% (87% being the lowest) and 18% of the patients had episodes of systemic desaturation indicating a right-to-left shunt due to increased pulmonary pressure [1]. We believe that nitric oxide would have been very beneficial, especially in those patients.

We would like to congratulate the authors for their successful management strategy of such a high risk patient group and for stressing once again the importance of unidirectional valved patches in treating patients with increased pulmonary vascular resistance.

Reference


eComment. Ventricular septal defect correction in patients with severe pulmonary hypertension

Authors: Leo A. Bockeria and Sergey Gorbachevsky
Bakauel Scientific Center for Cardiovascular Surgery of RAMS, Moscow, Russian Federation
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