

Valve patch technique for repair of ventricular septal defect: long-term results

Asian Cardiovascular & Thoracic Annals

0(0) 1–4

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DOI: 10.1177/0218492320962923

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Abstract

Objective: This study aimed to show the long-term results in patients who underwent unidirectional valve patch repair of ventricular septal defect with pulmonary artery hypertension.

Methods: Thirty-five acyanotic patients aged 2 to 26 years (mean 9.3 years) with a large ventricular septal defect and elevated pulmonary vascular resistance (mean 9.5 Wood units) underwent surgery in Madani Heart Hospital. The medical records and clinical outcomes were reviewed from March 1998 to March 2017.

Results: Five patients died in the first postoperative week. In the long-term follow-up (mean 11 years), two patients were lost to follow-up. Pulmonary artery hypertension gradually decreased in 17 patients within 6–12 months with significant improvement in right ventricular end-diastolic diameter, New York Heart Association functional class, and tricuspid regurgitation. Eleven patients with persistent pulmonary artery hypertension were divided into tolerable and non-tolerable groups. Six patients in the tolerable group had satisfactory conditions compared to before the operation, and gained weight with improved functional class despite echocardiographic findings of persistent elevated pulmonary artery pressure. One had a full-term delivery by caesarean section in the fifth postoperative year. Five patients in the non-tolerable group gradually developed right heart failure and complications such as extremity edema, ascites, pleural effusions, and died after 10–30 months.

Conclusion: Although relatively high mortality occurred during long-term follow-up, surviving patients were in a better condition and functional class despite persistent pulmonary artery hypertension. Therefore, fear of persistent pulmonary artery hypertension should not prohibit surgery in this group of patients.

Keywords

Cardiac surgical procedures, ventricular septal defect, pulmonary artery hypertension

Introduction

Ventricular septal defect (VSD) is one of the most frequent forms of congenital heart disease, and closure is a common procedure in cardiac surgery. A large hole in the ventricular septum causes enormous shunting of blood into the right ventricle and increases pulmonary arterial pressure, which sometimes induces irreversible damage in the pulmonary arterial bed. The size and location of the VSD as well as the pulmonary vascular resistance (PVR) are possible risk factors for the development of complications. In general, surgical closure is promising in this critical group of patients but sometimes, decision-making for surgery is challenging. Furthermore, different techniques with a wide variety of unidirectional valve patches (UVP) have been

designed for VSD closure to reduce the rates of mortality and morbidity.^{1–3} We have already shown our early and short-term results of VSD closure with our own designed UVP,⁴ and now report the long-term clinical course and survival in these high-risk cases.

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Patients and methods

From March 1998 to March 2017, 35 acyanotic patients with a large VSD and severe pulmonary artery hypertension (PAH) underwent surgery at Madani Heart Hospital, Tabriz University of Medical Sciences, Tabriz, Iran. The age range was 2 to 26 years and 48% were male. History, physical examination, electrocardiography, chest radiography, two-dimensional and Doppler echocardiography were the basic clinical evaluations in these patients. Functional class determined by two-step walking, pulmonary artery arborization on chest radiography, large left ventricular size on two-dimensional echocardiography, and a bidirectional shunt on color Doppler echocardiography were the criteria for inclusion in the study. Clinically cyanotic patients were excluded. The baseline characteristics of the 35 patients are summarized in Table 1.

Cardiac catheterization, hemodynamic data, oximetry, and 100% oxygen for at least 10 minutes were specified for the evaluation of PVR in all cases. In the included patients, PaO₂ was significantly increased under O₂ inhalation. Obtaining informed consent from the patient's family was the basis for making a decision for surgery.

All surgical procedures were performed by the same surgeon with conventional cardiopulmonary bypass, moderate hypothermia, and cold blood cardioplegia. All patients were treated with oral sildenafil and low-dose diuretics from the time of diagnosis until the day before surgery. Milrinone infusion was initiated for 30 min before the operation and continued for at least 2–3 days after VSD repair. Before establishing cardiopulmonary bypass, transesophageal echocardiography was used to determine exact size and location of the VSD to construct a precise UVP using a Gore-Tex patch (WL Gore, Newark, DE, USA) and a piece of the patient's pericardium with a central perpendicular slit of 5–8 mm in length, depending on the VSD size and PVR.⁴ The operative approach was frequently through the right atrium. To repair a subpulmonic or subarterial VSD,

both the main pulmonary artery and right atrial approaches were commonly used. Transesophageal echocardiography was performed in all patients before leaving the operating room to evaluate ventricular function, pulmonary arterial pressure, unidirectional shunt function, and the severity of tricuspid regurgitation (TR). In the intensive care unit, patients were sedated with fentanyl and supported by milrinone infusion and a ventilator in mild to moderate hypocarbia for 18–24 hours after the operation. Then, according to hemodynamic parameters, arterial blood gases, echocardiography findings, and in the absence of a significant right-to-left shunt via the UVP, the patients were allowed to wake up and were extubated. Unstable patients with marked signs of a low PaO₂ or any associated organ dysfunction were kept on the ventilator during the second and third postoperative days.

Data were compared with the Student *t* test, Mann-Whitney *U* test, or Wilcoxon matched-pairs test, as appropriate, and are reported as mean ± standard deviation.

Results

The VSD types are listed in Table 1. Baseline cardiac catheterization data before the operation are shown in Table 2. Five patients died in the first postoperative week due to low PaO₂, a right-to-left shunt via the valve patch, persistent severe PAH, and acute right heart failure. The remaining patients were attended to by pediatric cardiologists after the operation. Patients with echocardiographic findings of declining PVR and improvement in functional class were followed up every two months, and those with high and sustained PAH were monitored monthly and managed closely. The mean follow-up duration was 11 years (range 5 to 17 years). In the long-term follow-up, we lost contact with two patients. PAH gradually decreased in 17 patients 6–12 months postoperatively, with significant improvement in right ventricular end-diastolic diameter, functional class, and TR. These patients received oral sildenafil, low-dose diuretics, aspirin, and recently,

Table 1. Baseline characteristics of 35 patients with ventricular septal defects.

Variable	No. of patients
Age (years)	9.3 ± 8.8
Follow-up (years)	11 ± 4.5
Male	17 (48.57%)
Female	18 (51.43%)
Ventricular septal defect type	
Perimembranous	23 (65.71%)
Doubly committed	10 (28.57%)
Subpulmonic	1 (2.86%)
Muscular	1 (2.86%)

Table 2. Catheterization parameters before ventricular septal defect closure in 35 patients.

Parameter	Mean value
PA systolic pressure (mm Hg)	96.30 ± 19.082
PA diastolic pressure (mm Hg)	51.61 ± 10.08
AO systolic pressure (mm Hg)	106.70 ± 12.64
AO diastolic pressure (mm Hg)	67.90 ± 8.42
Systemic O ₂ saturation (room air)	88.6% ± 5.2%
PA/AO pressure	0.91 ± 0.14
PVR (Wood units)	9.5 ± 2.26

AO: aorta; PA: pulmonary artery; PVR: pulmonary vascular resistance.

Bosentan, during the first year. The other patients with persistent PAH were continued on these drugs. Eleven patients who had persistent PAH were divided into tolerable and non-tolerable groups. Six patients with tolerable PAH are in a satisfactory condition and have gained weight with improved functional class; one of them had a full-term delivery by caesarean section in the fifth postoperative year. These cases are under close observation by a pediatric cardiologist every 3 months. In five patients with non-tolerable PAH and right heart failure, complications such as cyanosis due to a right-to-left shunt via the UVP on color Doppler echocardiography, extremity edema, ascites, and pleural effusion developed, and they died at 10 to 30 months postoperatively (Table 3).

Discussion

Treatment is important in patients with elevated PAH because the size of the VSD affects the pressure gradient between the ventricles and the shunt volume, which play major roles in the physiopathology of VSD.⁵ Early surgery is indicated based on the severity of the symptoms; surgical treatment of cases of severe PAH after infancy is controversial. Unoperated patients are likely to develop Eisenmenger syndrome so decision-making for surgery of these borderline patients is challenging. Large left ventricular size on two-dimensional echocardiography, pulmonary vascular marking on chest radiography, and functional class are important factors to consider before surgery in these cases. Patients who walked two steps with little shortness of breath and no cyanosis had a smooth postoperative period, but this is no guarantee against failure and decreasing PVR. Catheterization findings and increasing PaO₂ on inhaled O₂ are important indications for surgery.

Decreasing the VSD size by making a fenestration in the patch was the technique used in the past. This procedure may cause a decrease in systemic PaO₂ due to a bidirectional shunt. Although in cases of persistent PAH, shunting via a fenestration will be permanently

right-to-left with cyanosis, in others with reduced PVR, a small VSD will have to be closed by a device. Today, two main surgical strategies, a unidirectional valve patch and a simple patch are used in these patients. Gan and colleagues⁶ emphasized that a unidirectional valve patch provides no benefits in terms of early or long-term survival, whereas some authors have reported better early results in children and adults with increased PVR.^{1,3}

Major concerns in the early days after the operation are pulmonary hypertensive crisis, acute right heart failure, and acute respiratory failure. Release of vasoactive substances such as thromboxane A₂ and catecholamines during and after the operation may predispose to pulmonary vasoconstriction and pulmonary hypertension crisis.^{7,8} Therefore, persistent PAH and shunting via a unidirectional valve to the left heart may prevent acute cardiopulmonary disturbance. Unlike Novick and colleagues,¹ we usually support patients on a ventilator during the night of the operation to keep PaO₂ high and PaCO₂ between 30 and 35 torr for late extubation. Unfortunately, we lost one extubated patient in the night of the operation due to pulmonary failure; reintubation and cardiopulmonary resuscitation were not effective. Early mortality in this study occurred in 5 patients: 3 due to cardiopulmonary failure in the intensive care unit and 2 in the ward with sudden death before discharge.

Survivors to hospital discharge were categorized into two groups: group 1 comprised patients whose pulmonary artery pressure and TR started to diminish on echocardiography, and group 2 was those who had echocardiographic signs of persistent PAH. All patients and their families were educated to do less activity during the postoperative period at home. Serial echocardiograms were performed every two months in patients in group 1, and close follow-up with medical treatment such as sildenafil and a calcium channel blocker with monthly echocardiograms was routine in group 2. Two patients were lost to follow-up and excluded from the study. During follow-up of 28 patients, 17 had echocardiographic findings of mild to moderate PAH with decreased right ventricular size and mild to moderate TR. Approximately 50% of our patients were back to near normal life at one year after the operation. The remaining 11 patients with the persistent elevated PVR were divided into two subgroups according to functional class and severity of right heart failure: there were 6 patients in subgroup 2a (tolerated PAH) and 5 in subgroup 2b (non-tolerated PAH). In subgroup 2a, no clinically overt right heart failure was found. The condition of these patients slowly improved with weight gain and increasing functional capacity. Despite warnings, one 26-year-old woman who was in the 5th postoperative

Table 3. Follow-up data of 35 patients after ventricular septal defect closure.

Variable	No. of patients
Mean follow-up (years)	11
Death	
Intensive care unit	3
Ward (sudden death)	2
Lost to follow-up	2
Decreased pulmonary hypertension	17
Persistent pulmonary hypertension	
Tolerable	6
Non-tolerable	5

year had a full-term delivery by caesarean section. Subgroup 2b had a worse postoperative period with clinical right heart failure requiring intermittent readmission to the hospital with dyspnea, extremity edema, ascites, and pleural effusion, and in some, cyanosis from two months after the operation. Cyanosis occurred in cases of a right-to-left shunt via the patch, the youngest patient was a two-year-old boy with severe ascites and cachexia refractory to medical treatment. The condition of all patients in subgroup 2b gradually worsened and they lost their lives during the first two years. These cases were challenging because if they did not undergo the operation, they would have survived with an Eisenmenger pattern for months or years, and living with this syndrome is not favorable.

We concluded that operative and postoperative mortality and morbidity in these cases are high. Although the valve patch technique is a promising way to save some of these patients in the early postoperative period, persistent PAH may lead to early cyanosis. However, in some surviving patients with PAH who feel in good condition, the valve patch may protect them from right heart failure.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

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