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Institutional report - Congenital

# Pulmonary valve repair late after right ventricular outflow tract reconstruction in children and adolescents

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**Abstract**

Here, we report our experience with pulmonary valve (PV) repair using neoleaflet implantation in children and adolescents. Between April 2005 and May 2009, 20 patients underwent late PV repair with neoleaflet implantation for pulmonary regurgitation (PR) after reconstruction of the right ventricular outflow tract at <18 years of age. In patients with sufficient remaining native PV leaflets, the neoleaflet was implanted with a polytetrafluoroethylene membrane or fresh autologous pericardium to coapt with the remaining native PV leaflets. The median age at operation was 13 years (2–18 years). The median follow-up duration was 12 months (2–51 months). There was no early or late death. During follow-up, no significant PR was observed, but significant pulmonary stenosis (PS) developed in two patients who had a combined PS. The indexed right ventricular end diastolic dimension decreased during follow-up. At the latest follow-up, all patients were in New York Heart Association functional class I or II. PV repair with neoleaflet implantation can be performed safely and the outcome is satisfactory in the short-term, but attention must be paid in cases with combined PS. Longer follow-up should be carried out to investigate the changes in leaflet mobility and function, and pulmonary annular growth.


*Keywords:* Congenital heart disease; Acyanotic heart disease; Valve disease

## 1. Introduction

In many patients who undergo surgical reconstruction of the right ventricular outflow tract (RVOT), reoperation or reintervention is required for late pulmonary regurgitation (PR). Although PR is well tolerated in some cases, the prolonged exposure to PR can lead to progressive right ventricular distension, increasing tricuspid regurgitation, dysfunction of both ventricles, ventricular arrhythmia, and sudden death [1, 2]. Numerous studies have demonstrated that the restoration of pulmonary valve (PV) competence can preserve cardiac performance, control arrhythmias and improve symptoms [3–5]. However, debate continues over the ideal techniques and materials as well as the optimal timing for pulmonary valve replacement (PVR) [6–8]. In the pediatric age group, early degeneration and patient outgrowth of the bioprosthesis have been major concerns for PVR with a bioprosthesis. When opening the RVOT for PVR in the operating room, a well-preserved native PV leaflet is observed at times. Since 2005, we have used native PV leaflets for reconstruction of the RVOT with implantation of a neoleaflet. Here, we report the short-term outcome of this technique.

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## 2. Methods

### 2.1. Patients

Our Institutional Review Board and Ethical Committee approved this retrospective study and waived the informed consent requirement. Between April 2005 and May 2009, 20 patients underwent PV repair using neoleaflet implantation and native PVs for reconstruction of the RVOT at an age <18 years. The patient characteristics are described in Table 1.

PV repair was performed in patients who met the following criteria: a grade 3 or 4 PR with symptoms, arrhythmia, right ventricular distension, right ventricular dysfunction, left ventricular (LV) dysfunction and residual intracardiac lesions to be repaired and significant pulmonary stenosis (PS) at any level. Twelve patients had pure PR and the others had combined significant PS including three cases of subvalvar PS (pressure gradient >36 mmHg). Preoperative New York Heart Association functional class (NYHA fc) was I in 10, II in 9 and III in 1 patient. The median age and weight were 12.7 years (2.4–17.5 years) and 34.7 kg (13.4–61.7 kg).

### 2.2. Doppler echocardiography

It is widely accepted that the cardiac magnetic resonance imaging is the gold standard for evaluation of the severity

Table 1
Patient characteristics

| No. | Age (years) | Wt. (kg) | Dx. | Interval from initial procedure (years) | Preoperative PV function | | Neoleaflet material | Preoperative NYHA fc | Additional intracardiac procedures | FU interval (months) | Postoperative PV function | | Postoperative NYHA fc |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | | | PR (grade) | PS (mmHg) | | | | | PR (grade) | PS (mmHg) | |
| 1 | 16 | 50.3 | ASD, PS | 13.2 | 3 | | PTFE | 1 | | 42 | 2 | | 1 |
| 2 | 10 | 26.9 | TOF | 9.9 | 4 | | PTFE | 2 | | 20 | 1 | | 1 |
| 3* | 13 | 36.9 | TOF | 11.1 | 3 | 70 | PTFE | 2 | RVOT patch widening | 13 | 2 | 54 | 1 |
| 4 | 11 | 30.4 | TOF | 10.9 | 3 | 44 | PTFE | 1 | RVOT m. resection | 16 | 2 | | 1 |
| 5 | 16 | 61.7 | TOF | 14.4 | 3 | 50 | PTFE | 2 | PV commissurotomy | 18 | 1 | | 1 |
| 6 | 15 | 58 | TOF | 13.1 | 4 | | PTFE | 2 | | 12 | 2 | | 1 |
| 7 | 5 | 17.3 | TOF | 4.1 | 3 | 49 | PTFE | 2 | TVP | 51 | 2 | | 1 |
| 8 | 16 | 53 | TOF | 14.1 | 3 | | PTFE | 1 | | 38 | 1 | | 1 |
| 9 | 6 | 20 | TOF | 5.2 | 3 | 81 | PTFE | 2 | TVP, RVOT m. resection | 12 | 2 | | 1 |
| 10 | 8 | 21.6 | TOF | 7.0 | 3 | | PTFE | 1 | | 16 | 1 | | 1 |
| 11 | 13 | 43 | TOF | 12.2 | 4 | | PTFE | 2 | | 10 | 2 | | 1 |
| 12 | 7 | 22 | TOF | 7.2 | 3 | | PTFE | 1 | Residual ASD closure | 11 | 2 | | 1 |
| 13 | 10 | 32.6 | TOF | 8.8 | 3 | 36 | FP | 2 | | 7 | 2 | | 1 |
| 14 | 14 | 24.4 | TOF | 12.4 | 4 | | FP | 1 | | 6 | 1 | | 1 |
| 15* | 17 | 56.9 | TOF | 16.8 | 3 | 36 | FP | 3 | | 9 | 1 | 46 | 2 |
| 16 | 11 | 23.5 | TOF | 10.3 | 4 | | FP | 2 | TVP | 6 | 2 | | 1 |
| 17 | 13 | 48.4 | TOF | 12.1 | 3 | | FP | 1 | | 8 | 2 | | 1 |
| 18 | 2 | 13.4 | TOF | 2.0 | 3 | 74 | FP | 1 | RVOT m. resection | 7 | 2 | | 1 |
| 19 | 14 | 42.4 | TOF | 13.6 | 4 | | FP | 1 | | 6 | 1 | | 1 |
| 20 | 14 | 44.6 | TOF | 13.6 | 4 | | FP | 1 | | 2 | 1 | | 1 |

*Patients who had late pulmonary valve dysfunction; No., patient's serial number; Wt., weight; Dx., diagnosis; PV, pulmonary valve; FU, follow-up; NYHA fc, New York Heart Association functional class; PR, pulmonary regurgitation; PS, pulmonary stenosis; ASD, atrial septal defect; PTFE, polytetrafluoroethylene; TOF, tetralogy of Fallot; RVOT, right ventricular outflow tract; m., muscle; TVP, tricuspid valvuloplasty; FP, fresh autologous pericardium.

of PR and right ventricular volume. However, this modality is not easily accessible and is costly, therefore, it may be impractical for the serial follow-up in our center. For this reason, we used Doppler echocardiography as a tool for evaluation of the PV function and right ventricular volume during follow-up. The PR was measured with degree of regurgitant Doppler color jet in the parasternal short-axis imaging plane and graded from 0 to 4. To evaluate the change of right ventricular volume, right ventricular end-diastolic dimension (RVEDD) was measured and indexed to the patient's body surface area (RVEDDi). The PS gradient (peak) was calculated by the modified Bernoulli equation and graded by the recommendation of the 2006 American College of Cardiology/American Heart Association guidelines [9].

### 2.3. Technique

PV repair was performed using moderate hypothermic cardiopulmonary bypass. Cardioplegic arrest was required in five patients for additional intracardiac procedures. The mean bypass time was 117.6±29.0 min and the mean ischemic time ($n$=5) was 13.1±24.2 min. We considered a PV repair possible when the preoperative echocardiogram showed a remnant mobile native PV leaflet. After the RVOT was opened, we determined whether to proceed with the valve repair procedure, the details of which are as follows (Fig. 1).

In cases with a previous transannular repair ($n$=14), the previously placed patch was incised longitudinally without extension toward the intact right ventricle (Fig. 1a). In cases with a preserved annulus ($n$=6), the incision was extended toward the right ventricle no more than 10 mm. Preserving the native PV leaflet, the neoleaflet was fashioned and implanted with a 0.1-mm-polytetrafluoroethylene (PTFE) membrane (GoreTex; W.L. Gore & Associates, Inc, Newark, USA) or fresh autologous pericardium (FP) on the leaflet-defected pulmonary annulus. The length of the

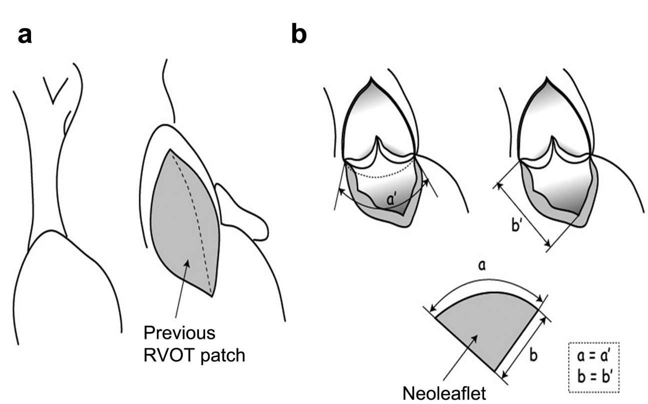


Fig. 1. Diagram for pulmonary neoleaflet implantation. (a) Dotted line indicates the length of longitudinal incision along with the previous RVOT patch; (b) opened RVOT and neoleaflet. RVOT, right ventricular outflow tract.

free-margin of the neoleaflet was determined to conform to the length of the leaflet-defected pulmonary annulus, and the length of both lateral margins of the neoleaflet was the length between the most proximal point of the longitudinal incision (endocardial surface) and just above the pulmonary annulus along with the longitudinal incision (Fig. 1b). In cases using FP, thick paper was used for easy handling and designing the neoleaflet. If the commissure between the native valve leaflets was loose, a commissural reinforcement suture was added. The Z score of the pulmonary annulus was more than 1 after the valve repair.

### *2.4. Follow-up and data analysis*

Echocardiography was performed just after the patient was weaned off of bypass, just before discharge and during follow-up in all patients. We evaluated the leaflet motion, development and progression of PR or PS and changes in the right ventricular dimension. The median duration of follow-up was 12 months (2–51 months).

Data are presented as the mean $\pm$ standard deviation (S.D.) or median with range. To compare variables between groups, the $t$-test for continuous variables and $\chi^2$-test for categorical variables were performed. A $P<0.05$ was considered statistically significant.

## 3. Results

There were no early deaths or operative morbidity. The median length of intensive care unit and hospital stay was 2 days (1–7 days) and 7.5 days (5–18 days) respectively. During follow-up, there was no late death or reintervention. Transesophageal echocardiography, just after discontinuation of bypass, and transthoracic echocardiography, before discharge, showed a PR less than grade 2 and no significant PS in all patients. During follow-up, no significant PR developed, but in two patients, significant PS developed (Table 1). Leaflet mobility was intact in all patients at the latest follow-up echocardiography (up to 51 months). Among 12 patients who had pure PR preoperatively, no PV dysfunction developed; however, it developed in two out of eight patients with combined preoperative PS ($P=0.147$) (Table 1).

The mean RVEDDi decreased from $36.1 \pm 10.0$ mm/m$^2$ preoperatively to $29.7 \pm 8.0$ mm/m$^2$ postoperatively ($P=0.003$) (Fig. 2). At the latest follow-up, all patients were graded as NYHA fc I or II and were not taking any cardiac medication except for short-term (three months) antiplatelet agents.

## 4. Discussion

For many years, xenoprostheses or homografts have been the major valve substitutes for the PV in the pediatric age group. Although the outcome regarding survival and reoperation is acceptable, the late valve function has been disappointing. The possible factors associated with late valve dysfunction are the patient outgrowing the prosthesis, degeneration with subsequent calcification attributed to the immune response and suboptimal mechanical properties. In addition, young children have few options for valve substitutes, because of the limited availability of

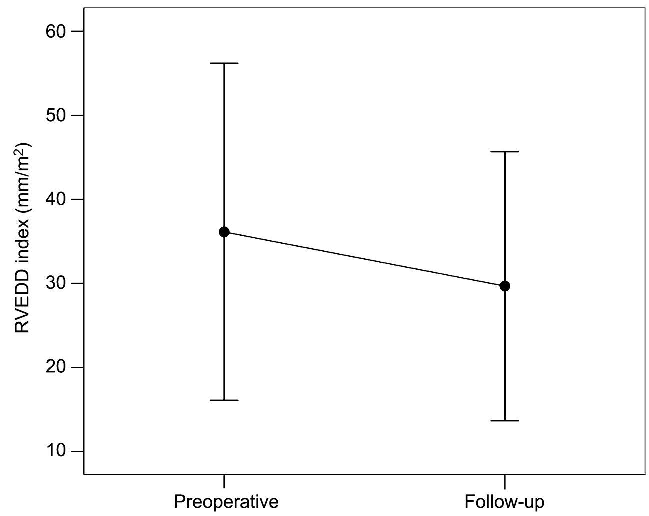


Fig. 2. Change in RVEDDi. RVEDDi, right ventricular end diastolic dimension index.

small-sized commercial xenoprostheses and homografts. Some efforts to avoid these biological valves for PVR have been made infrequently with excellent short-term outcomes regarding valve function [10, 11].

With our simple repair technique, we expected that the annulus anchoring native leaflets might grow. PTFE and FP as a material for a valve substitute have some advantages as a substitute for the PV. PTFE is chemically inert and has low friction and very low tissue affinity, which might lead to reduced cellular or fibrinous deposition and resistance to tissue degeneration or destruction. Turrentine and colleagues [12] found no calcification in the PTFE membrane and a rather well-vascularized layer of non-obstructive fibrocollagenous tissue. Kalangos and coworkers [13] observed that in 41 children who underwent aortic cusp extension with FP, cusp mobility and coaptation were preserved. In this report, aortic regurgitation had been aggravated from grade 1 to 2 in only one of the early survivors at three years of median follow-up. The authors noted that the autologous pericardium elicited a reduced inflammatory reaction and had expansion potential, compared to cases with treated autologous pericardium or xenopericardium. In reoperated cases, similar to the cases in this report, mediastinal adhesions and a thickened pericardium may preclude obtaining truly FP. However, we could harvest a FP from the right side above the right phrenic nerve or the diaphragmatic surface without much difficulty.

In our series, PV dysfunction (all were stenosis) developed in two patients out of the eight patients who had PR and combined stenosis preoperatively. Despite the absence of statistical significance, PV dysfunction was only observed in patients who had preoperative combined PS ($P=0.147$). This finding might be attributed to the rather small annulus to be enlarged, an increased need for a native valve procedure and poor subvalvar morphology that may influence the mechanism of the neoleaflet. Although reoperation has yet to be required in our patients, this procedure of PV repair with neoleaflet formation must be considered with caution in patients with combined PS, and longer follow-up is essential.

When a considerable amount of the native PV leaflet showed good mobility and adequate thickness in the operating room, we used the native PV leaflet for PV repair. In most patients, the PV leaflet was dysplastic and thickened at the initial procedure. Papadopoulos and associates [11] observed similar findings even in patients with pulmonary atresia and noted that the regurgitant jet might stimulate the development or growth of the lunular structures. Butcher et al. [14] noted that valvular endothelial and interstitial cells respond to a variety of mechanical stimuli, often with unique responses that are putatively relevant to valve remodeling. Ikhumetse and coworkers [15] performed experiments on porcine PV leaflets to investigate the biological properties of PV leaflets subjected to normal aortic pressures. They observed that the PV leaflets adapted to changes in mechanical conditions by increasing the collagen and sulfated glycosaminoglycan (sGAG) content. Therefore, it can be inferred that the PV leaflet can be remodeled in changed circumstances around the RVOT through a process that has yet to be clearly identified. In addition, it is recommended that at the initial procedure, valvectomy should be avoided to allow for future valve repair if possible.

This study was limited by inclusion of a small number of patients, short duration of follow-up, retrospective methodology and no definite criteria regarding the choice of the neoleaflet material. In addition, the parameters used to evaluate functional outcomes were subjective, and cardiac performance and dimension were measured with echocardiography, which is not the most reliable method. Since the follow-up period was short, additional objective measures to evaluate the functional capacity, cardiac dimension, performance status and pulmonary annulus size, which is one of our matter of concerns, could not be performed in all patients.

In conclusion, PV repair with neoleaflet implantation can be performed safely. Over the short-term, the outcome was satisfactory in patients with pure PR. However, in cases with combined PS, this procedure should be performed with caution. Longer follow-up is essential for the evaluation of the changes in leaflet mobility and function, and pulmonary annular growth.

## References


[1] Ebert PA. Second operations for pulmonary stenosis or insufficiency after repair of tetralogy of Fallot. Am J Cardiol 1982;50:637–640.

[2] Geva T, Sandweiss BM, Gauvreau K, Lock JE, Powell AJ. Factors associated with impaired clinical status in long-term survivors of tetralogy of Fallot repair evaluated by magnetic resonance imaging. J Am Coll Cardiol 2004;43:1068–1074.

[3] Bove EL, Kavey RE, Byrum CJ, Sondheimer HM, Blackman MS, Thomas FD. Improved right ventricular function following late pulmonary valve replacement for residual pulmonary insufficiency or stenosis. J Thorac Cardiovasc Surg 1985;90:50–55.

[4] Warner KG, O'Brien PKH, Rhodes J, Kaur A, Robinson DA, Payne DD. Expanding the indications for pulmonary valve replacement after repair of tetralogy of Fallot. Ann Thorac Surg 2003;76:1066–1072.

[5] Discigil B, Dearani JA, Puga FJ, Schaff HV, Hagler DJ, Warnes CA, Danielson GK. Late pulmonary valve replacement after repair of tetralogy of Fallot. J Thorac Cardiovasc Surg 2001;121:344–351.

[6] Davlourosa PA, Karatza AA, Gatzoulisa MA, Shore DF. Timing and type of surgery for severe pulmonary regurgitation after repair of tetralogy of Fallot. Int J Cardiol 2004;97:91–101.

[7] Geva T. Indications and timing of pulmonary valve replacement after tetralogy of Fallot repair. Semin Thorac Cardiovasc Surg Pediatr Card Ann 2006;9:11–22.

[8] Lim C, Lee JY, Kim W-H, Kim S-C, Song J-Y, Kim S-J, Choh J-H, Kim CH. Early replacement of pulmonary valve after repair of tetralogy: is it really beneficial? Eur J Cardiothorac Surg 2004;25:728–734.

[9] Bonow RO, Carabello BA, Chatterjee K, de Leon AC Jr, Faxon DP, Freed MD, Gaasch WH, Lytle BW, Nishimura RA, O'Gara PT, O'Rourke RA, Otto CM, Shah PM, Shanewise JS, Smith Jr SC, Jacobs AK, Adams CD, Anderson JL, Antman EM, Faxon DP, Fuster V, Halperin JL, Hiratzka LF, Hunt SA, Lytle BW, Nishimura R, Page RL, Riegel B. ACC/AHA 2006 Practice Guidelines for the Management of Patients with Valvular Heart Disease: Executive Summary: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Revise the 1998 Guidelines for the Management of Patients with Valvular Heart Disease). J Am Coll Cardiol 2006;48:598–675.

[10] Quintessenza JA, Jacobs J, Morell VO, Giroud JM, Boucek RJ. Initial experience with a bicuspid polytetrafluoroethylene pulmonary valve in 41 children and adults: a new option for right ventricular outflow tract reconstruction. Ann Thorac Surg 2005;79:924–931.

[11] Papadopoulos N, Esmaeili A, Zierer A, Bakhtiary F, Ozaslan F, Moritz A. Secondary repair of incompetent pulmonary valves. Ann Thorac Surg 2009;87:1879–1884.

[12] Turrentine MW, McCarthy RP, Vijay P, McConnell KW, Brown JW. PTFE monocusp valve reconstruction of the right ventricular outflow tract. Ann Thorac Surg 2002;73:871–880.

[13] Kalangos A, Beghetti M, Baldovinos A, Vala D, Bichel T, Mermillod B, Murith N, Oberhansli I, Friedli B, Faidutti B. Aortic valve repair by cusp extension with the use of fresh autologous pericardium in children with rheumatic aortic insufficiency. J Thorac Cardiovasc Surg 1999;118:225–236.

[14] Butcher JT, Simmons CA, Warnock JN. Review: mechanobiology of the aortic heart valve. J Heart Valve Dis 2007;17:62–73.

[15] Ikhumetse JD, Konduri S, Warnock JN, Xing Y, Yoganathan AP. Cyclic aortic pressure affects the biological properties of porcine pulmonary valve leaflets. J Heart Valve Dis 2006;15:295–302.


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