



Pulmonary valve replacement after repaired Tetralogy of Fallot

Hideki Tatewaki¹ · Akira Shiose¹

Received: 17 February 2018 / Accepted: 28 April 2018 / Published online: 19 May 2018
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Abstract

In this review article, we describe pulmonary valve replacement (PVR) late after repaired Tetralogy of Fallot (TOF). Since the introduction of surgical intervention for patients with TOF in 1945, surgical management of TOF has dramatically improved early survival with mortality rates, less than 2–3%. However, the majority of these patients continue to experience residual right ventricular outflow tract pathology, most commonly pulmonary valve regurgitation (PR). The patients are generally asymptomatic during childhood and adolescence and, however, are at risk for severe PR later which can result in exercise intolerance, heart failure, arrhythmias, and sudden death. While it has been shown that PVR improves symptoms and functional status in these patients, the optimal timing and indications for PVR after repaired TOF are still debated. This article reviews the current state of management for the patient with PR after repaired TOF.

Keywords Tetralogy of Fallot · Pulmonary regurgitation · Pulmonary valve replacement · Adult congenital heart disease · Sudden death

Introduction

Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease. The first Blalock–Tausig shunt operation in 1945 improved life expectancy from a few months to years [1]. Lillehei et al. followed with the first successful intracardiac repair in 1954 [2]. The current early survival of TOF is excellent, at approximately 90–98% through the first two decades of life [3, 4]. While early mortality has improved, studies of adult survivors with repaired TOF showed that late complications included exercise intolerance, right heart failure, arrhythmias and sudden death [5, 6]. Many of these complications are related to the hemodynamic effects of pulmonary regurgitation (PR) and associated chronic right ventricular (RV) volume overload [5, 7, 8]. Patients with PR can remain asymptomatic for many years with a gradual deterioration of exercise performance inversely related to PR severity [9]. Because echocardiography is often inadequate for evaluating early RV dysfunction, cardiac magnetic resonance (CMR) imaging has become an

integral tool in evaluation of RV volumes, severity of PR, and biventricular function in following these patients later in life. It has been shown that Pulmonary valve replacement (PVR) alleviates symptoms, normalizes RV volumes, and improves RV function. It is unclear whether performing PVR decreases the risk of ventricular tachycardia (VT) or sudden death. There is strong evidence that elimination or significant reduction of PR after PVR is associated with symptomatic improvement, decrease in RV end-diastolic volume (RVEDV) and RV endo-systolic volume (RVESV), and no significant change in RV ejection fraction using CMR imaging [5, 10–12]. There is no consensus, however, regarding the indication and the timing for PVR. This article reviews the current state of management for the patient with PR after repaired TOF.

Pulmonary regurgitation

After intracardiac repair of TOF, the patient nearly always had residual right ventricular outflow tract (RVOT) pathology, most commonly PR. For many years, PR after repaired TOF was considered benign [13]. Young patients appeared to tolerate PR well and were subsequently lost to follow-up. It was later found that as these patients reached adulthood, the hemodynamic burden of chronic PR presented as late

✉ Hideki Tatewaki
tatewaki@heart.med.kyushu-u.ac.jp

¹ Department of Cardiovascular Surgery, Kyushu University Hospital, 3-1-1 Maidashi, Higashi-ku, Fukuoka 8128582, Japan

morbidity including exercise intolerance, right heart failure, and arrhythmias. The most common cause of mortality was sudden death. These adverse long-term outcomes in patients with TOF are demonstrated by a significant decrease in their survival curve in the second decade following repair [3].

RV systolic function in chronic PR is initially preserved. This stage may last for years and many patients remain relatively free of symptoms. As PR progresses, however, several studies have shown a close relationship between the severity of PR and RV end-diastolic volume [14, 15]. Severe PR is also known to impair biventricular function. Davlourous et al. demonstrated that left ventricular (LV) systolic dysfunction correlated with RV dysfunction in adults with repaired TOF, suggesting an unfavorable ventricular–ventricular interaction [16]. This study also showed three independent predictors of LVEF: length of time that patient remained palliated, aortic regurgitation fraction, and RVEF; the first two correlated inversely with LVEF. The mechanism that links RV dysfunction to LV dysfunction, however, is incompletely understood in repaired TOF.

Arrhythmias

Arrhythmias are also common sequelae of repaired TOF. Atrial arrhythmias may be present in approximately one-third of patients and are a major source of morbidity. Although the prevalence of sustained ventricular tachycardia (VT) is low, it is believed to be responsible for an increased incidence of sudden death in patients with TOF. Gatzoulis et al. in a multicenter study of 793 patients with repaired TOF at a mean of 8.2 ± 8 years, demonstrated that an older age at TOF repair and QRS duration > 180 ms were independent predictors of sudden death [5]. Khairy et al. in a multi-institutional study in 556 patients with repaired TOF reported a prevalence of atrial tachyarrhythmias (20.1%) and ventricular arrhythmias (14.6%) that markedly increased after 45 years of age. These arrhythmias were associated with the number of cardiac surgeries, QRS duration, and left ventricular diastolic dysfunction [17]. Targeted intraoperative cryoablation procedures during PVR appear to decrease the incidence of arrhythmias, at least in the short term [18]. In patients with risk factors for VT and sudden death, pre-operative electrophysiological study and implantation of a cardioverter–defibrillator should be considered [19].

Evaluation of pulmonary regurgitation and ventricular function

For many years, echocardiography has been used as the primary non-invasive imaging modality in the follow-up of patients with repaired TOF. The complex geometry of the

RV makes determining volume and function difficult using echocardiographic assessments. Doppler echocardiographic assessment of PR is also dependent on the observer's experience, and is limited by post-operative structural changes. CMR imaging has become the reference standard for assessment of biventricular size and function and offers a reliable method for the longitudinal assessment of PR severity and RV dilation in patients with repaired TOF [10, 20, 21]. Furthermore, CMR imaging is non-invasive and does not use ionizing radiation. Echocardiography and CMR imaging can be used in concert as a combined non-invasive imaging study in patients with repaired TOF. Yamasaki et al. demonstrated that cardiac computed tomography (CCT) can also assess RV function and pulmonary regurgitant fraction (PRF), compared with CMR imaging in patients with repaired TOF [22]. CCT is more useful than CMR for the patients with an implanted pacemaker/defibrillator, patients with claustrophobia or any clinical condition that prohibits a long CMR examination.

Indication and timing for PVR

PVR can be accomplished with a low early mortality in many centers, but should be performed before there is irreversible RV dysfunction and the increased propensity for VT and sudden death [23, 24]. The optimal management of patients who may have clinical or physiological indications for PVR has been widely researched and debated. Identification of risk factors for adverse clinical events is important for determining the optimal timing of PVR. There is agreement that PVR is indicated in symptomatic patients with significant PR, heart failure, or new arrhythmias. In the absence of these findings, physicians should consider a broad range of clinical data and longitudinal imaging modalities when considering PVR in the individual patient.

Several CMR imaging based studies have found that RV volumes return to normal after PVR if the pre-operative RVEDVI is < 150 – 170 ml/m² or the RVESVI is < 82 – 90 ml/m² [5, 10, 12]. Biventricular dysfunction and RV hypertrophy are associated with poor outcome in patients with repaired TOF [25].

Frigiola et al. advocated that a relatively aggressive PVR indicator (RVEDVI < 150 ml/m²) leads to normalization of RV volumes, improvement in biventricular function, and increased exercise capacity [26]. Frigiola also reported that patients with repaired TOF who were > 35 years of age without PVR and with normal exercise capacity showed mild residual RVOT obstruction and pulmonary annulus diameters $< 0.5Z$ [27].

Lee et al. suggest that pre-operative RVEDVI of 168 ml/m² and pre-operative RVESVI < 80 ml/m² are predictors for optimal outcome [10]. RV volume normalization is also

associated with a pre-operative QRS duration < 140 ms, whereas a QRS duration > 160 ms and an RV ejection fraction < 45% are associated with persistent RV dilation and dysfunction [11]. Therrien et al. showed that RV remodeling to normal size was unlikely once the RV end-diastolic volume (RVEDV) reached 170 ml/m² and the RV end-systolic volume (RVESV) reached 85 ml/m² [5].

Valve selection

Regarding the selection of prosthetic valve, the stented bioprosthetic valve is preferred for isolated PVR in adult patient, but structural valve deteriorations are unavoidable in the long term. A biological valve offers the advantages of not needing anticoagulation, whereas future replacement could be performed by transcatheter valve-in-valve procedures. A younger age at implantation with biological valve is associated with higher rates of valve deterioration [28].

Controversy still remains with respect to the best bioprosthesis to implant. Fiore et al. reviewed the performance of the mosaic porcine, bovine pericardial, and homograft prostheses in a cohort of patient who underwent PVR at a mean of 15.3 years after initial outflow tract reconstruction and reported that all three prostheses performed similarly for 3 years and PR developed more frequently in homografts [29].

Use of a mechanical valve could be an alternative especially for patients with high surgical risk including multiple prior surgery and patients already using anticoagulation agents. Pragt et al. demonstrated that freedom from valvular thrombosis was 86% at 10 year post-PVR and freedom from reoperation 91% at 10 years at a median follow-up duration of 4.26 years (range, 0–27 years) in 364 patients (69.8% with cardiac diagnosis of TOF and mean age at time of implantation was 27.2 ± 12.2 years) [30]. Propensity for thrombosis was 1.7% per patient year and incidence for reoperation was 1.0% per patient year. They concluded that mechanical PVR is associated with a limited risk of valvular thrombosis and thrombolysis was an effective treatment with a success rate up to 88%. Fleling et al. also demonstrated a 10% incidence of thrombosis or pannus valve-related complications at a mean follow-up duration of 5.5 years in 66 patients following mechanical PVR (77% with cardiac diagnosis of TOF and mean age at time of implantation was 35 ± 13 years) [31].

Mechanical valve in the pulmonary position seems not to do worse. Mechanical valve is suitable for the selected patients. These selected patients, we think, are who had multiple prior operations and ventricular dysfunction or who need left-sided valve surgery with mechanical valve simultaneously. Further data of both biological valve and mechanical valve in the long term will be necessary.

Other complications

Coexisting problems such as tricuspid regurgitation, RVOT obstruction, residual shunts, and aortic regurgitation or dilation requiring surgical intervention should also be considered. Management of functional tricuspid valve regurgitation at the time of PVR remains controversial. However, most surgeons tend to take an aggressive approach to concomitant tricuspid valve annuloplasty at the time of PVR when tricuspid valve regurgitation was more than moderate. Previous studies have indicated that as many as 13–41% of patients with repaired TOF undergoing PVR will undergo a concomitant tricuspid valve repair for tricuspid regurgitation [32–36]. Kogon et al. reported that in 35 patients with at least moderate tricuspid regurgitation, significant improvement in tricuspid valve function and right ventricular size occurs in the first post-operative month after pulmonary valve replacement, irrespective of concomitant tricuspid valve annuloplasty [37]. Cramer et al. demonstrated that after pulmonary valve replacement with or without tricuspid valve repair, there is significant improvement in the degree of tricuspid regurgitation and right ventricular size. They also concluded that 6 months after pulmonary valve replacement there were no statistical differences between those patients undergoing concomitant tricuspid valve repair and those undergoing pulmonary valve replacement alone [38]. We totally agree with the concept of Kogon and Cramer's papers when tricuspid regurgitation is only central due to RV dilatation. However, some patients have a structural deformity of the tricuspid valve, especially between anterior and septal leaflets where the patch had been sewn. Decision making to perform tricuspid valve repair or not should be made not only by the degree of tricuspid regurgitation but by the combined assessment with the tricuspid valve structure.

RV remodeling and late outcome of PVR

PVR is increasingly used to treat the chronic RV volume overload from pulmonary valve regurgitation. The procedure can be performed by a transcatheter technique or surgically, using one of the many available bioprosthetic valve. The early results of PVR have been well described by several groups, demonstrating consistent results characterized by resolution or marked reduction of PR, 30–40% reduction in RVEDVI and RVESVI, unchanged RV ejection fraction (EF), slightly increased left ventricular (LV) size with unchanged LV EF, decrease in RV systolic pressure in those with pre-operative RVOT obstruction, and consistent improvement in NYHA functional class without

a clear change in objective exercise parameters or arrhythmia burden [9, 35, 39–41]. In a study of 71 patients, Frigola et al could not demonstrate an improvement in peak oxygen consumption or anaerobic threshold in patients undergoing PVR for RVOT [26].

In addition to the recommended indications for PVR, the risks and benefits should be weighed in each patient. All bioprosthetic valves inserted in the pulmonary position have a limited life expectancy, with wide variations in rates of freedom from valve failure and reoperation. Freedom from RVOT reintervention after PVR has been reported to be 58–96% at 10 years [9, 42, 43]. Younger patients have more consistent improvements in RV function with PVR and, however, are associated with higher rate of valve failure and early reoperation [44–46]. Hallbergson et al. reported that early reduction in RV size showed a gradual return toward pre-operative values by 7–10 years after PVR. Late adverse RV remodeling was associated with increased RV volume and pressure loads. These findings highlight the palliative nature of PVR and the importance of continued longitudinal surveillance [47].

Operative considerations

Careful pre-operative planning is crucial to minimize the risk of surgery. Previous operative records should be reviewed to evaluate the individual anatomical features. Pre-operative electrocardiographic (ECG) gated multidetector CT is useful modality to evaluate the risk of resternotomy. Ultrasound studies should be routinely performed to confirm the patency of the femoral vessels. Especially in patients with adult congenital heart disease, femoral vessels may have been stenosed or occluded during previous surgical intervention, cardiac catheterization, or thrombosed as a consequence of vascular access.

Alternate cannulation sites including femoral vessels and axillary artery should be identified and prepped into the operative field before resternotomy. In the case of high risk resternotomy, both femoral vessels should be exposed to start cardiopulmonary bypass (CPB). Resternotomy is undertaken with an oscillating saw. Once resternotomy has been completed, the pericardial contents are dissected out.

Kyushu University hospital experience

The decision to proceed to PVR was made by the primary cardiologist in consultation with our cardiologist and cardiac surgeon based on the evidence of valve dysfunction. In the case of PR, documentation of severe PR, defined by regurgitant fraction > 40% by CMR imaging, exercise intolerance, arrhythmia, and excessive RV dilatation, defined

by RVEDVI > 150 ml/m² by CMR imaging, were used as criteria.

Surgical procedures at our institution for PVR after repaired TOF are described below. CPB is established using aortic and bicaval cannulation and mild hypothermia. We routinely employ aortic cross-clamping and cardioplegia for PVR to eliminate the risk of air embolism. An antegrade cardioplegia cannula is placed in the ascending aorta to give cold intermittent blood cardioplegia. LV is vented through the aortic root or right superior pulmonary vein. Our current practice is to use a stented bovine pericardial valve and commonly implant a 21–25 mm valve. Selection of the valve size is performed after intraoperative assessment of the opened RVOT. A longitudinal incision is made into the proximal outflow tract and all calcifications were debrided as appropriate. Residual pulmonary valve tissue and any hypertrophied muscular trabeculations in the subjunctional region are removed to create a wide-open pathway. A stented bioprosthetic valve is sewn into the orthotopic position in the RVOT. The posterior two-thirds of the bioprosthetic valve are sutured to the pulmonary annulus or just above the annulus with interrupted mattress sutures. Special caution should be taken to avoid coronary compression or injury when the surgeon put the stitches on the RVOT. Precise pre-operative CT evaluation may help to avoid these complications. Another technical consideration is to put the prosthetic pulmonary valve properly to keep laminar blood flow from RVOT to both pulmonary arteries. The RVOT is augmented with a diamond-shaped patch as necessary to accommodate the valve. Once the procedure is complete, the patient is placed in Trendelenburg position, deairing confirmed, and the cross-clamp is released. Intraoperative transesophageal echocardiography is uniformly performed to rule out the presence of air in the left side of the heart, and to make sure the procedures are satisfactory. Consideration should be given to placement of a substernal Gore-Tex to facilitate predictable resternotomy. All patients were treated with Coumadin for 3 months.

A retrospective analysis of our experience with PVR among repaired TOF patient was approved by Kyushu University Hospital Human Research Review Board. Between 2003 and 2017, 52 patients greater than 15 years of age underwent a first-time PVR following repair of TOF. The mean age of intracardiac TOF repair was 5.5 ± 6.3 years. All patients underwent PVR with a bioprosthetic bovine pericardium valve at a mean age of 29.6 ± 10.5 years. Concomitant procedures included peripheral pulmonary artery plasty in 13, tricuspid valve plasty in 9, cryoablation for arrhythmias in 7, other procedures in 15. Mean follow-up time was 4.3 ± 3.8 years. There were no early and late deaths. No pulmonary valve reintervention have been required with a longest follow-up time of 14.6 years. Two other reoperations were performed including residual atrial septal defects closure in

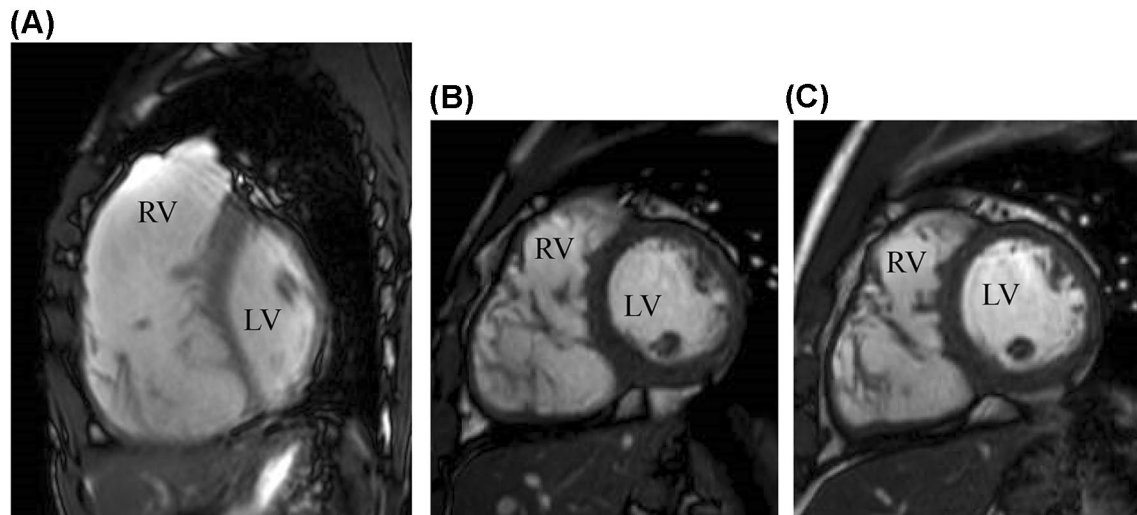


Fig. 1 Cardiac MR imaging in a 43-years-old patient with repaired TOF before and after PVR. **a** Pre-operative MRI shows severe RV dilatation (RVEDVI, 518.3 ml/m²) and flattened intraventricular septum with severe PR (pulmonary regurgitant fraction 50%), severe tricuspid regurgitation, moderate mitral regurgitation and RV dysfunction (ejection fraction, 18%) This patient underwent PVR with

tricuspid valve repair and mitral valve repair simultaneously. **b** MRI 1 year after PVR demonstrated improved RV dilatation (RVEDVI, 222.1 ml/m²), and improved interventricular septum shift. **c** MRI 2 years after surgery showed further improvement in RV dilatation (RVEDVI, 183.1 ml/m²)

one and Bentall operation in another. Compared with pre-PVR measurement, RVEDVI significantly decreased from 193.4 ± 87.0 to 114.0 ± 38.4 ml/m² ($p=0.002$) (Fig. 1) and pulmonary regurgitant fraction also significantly decreased from 44.4 ± 15.3 to $4.5 \pm 5.0\%$ ($p=0.003$). Both LVEF and RVEF remained unchanged from 47.6 ± 10.9 to $47.2 \pm 9.1\%$ and plasma brain natriuretic peptide levels improved from 77.0 ± 85.0 pg/dl to 38.5 ± 31.8 pg/dl. All but one patients had NYHA functional class I following PVR.

Conclusions

Repaired TOF can be characterized as a latent chronic right heart failure disease whose cause is a gradual increasing PR grade into the second decade of life. Decreasing RV size and PR fraction can be achieved early with PVR and with a low mortality. However, improved long-term clinical outcome is determined also using careful pre-operative evaluations, concomitant cardiac procedures to improve RV function, longitudinal cardiac imaging modalities to assess RV geometry and function over time and the use of optimal criteria for and timing of PVR as earlier PVR has been shown to improve biventricular function, while young adults requiring PVR may require several reoperations in their life time.

Compliance with ethical standards

Conflict of interest The authors declare no conflicts of interest.

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