Chapter 12
Redo Fallot: Surgery for Pulmonary Valve Implantation

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Abstract Since tetralogy of Fallot was first successfully repaired in 1954, an encouraging long-term survival has been reported. However, an increasing number of adult survivors will present late after repair for re-intervention, and pulmonary regurgitation has been the most common indication. The timing and indications for pulmonary valve replacement continue to evolve but the trend has now moved towards earlier intervention before irreversible changes in right ventricular function occur. Late survivors are also at an increased risk of developing arrhythmias due to right ventricular scarring related to corrective surgery and sudden death remains the commonest cause of late death. The need to restore pulmonary valve competency to preserve right ventricular function and reduce the arrhythmia burden have therefore become the important indications for late intervention in this disease. In addition to traditional surgical approaches, the role of percutaneous valve intervention is also emerging. It is anticipated that this strategy will improve the long-term mortality and morbidity and will be the focus of this chapter.

Keywords Tetralogy of Fallot • Redo-operation • Pulmonary regurgitation • Valve surgery • Arrhythmia surgery

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Introduction

Since tetralogy of Fallot was first successfully repaired by Lillihei and Varco in 1954 at the University of Minnesota [1], long-term survival of 85 % at 36 years has been reported [2]. The need to relieve the right ventricular outflow tract obstruction (RVOTO) is often accompanied by residual pulmonary regurgitation (PR). Surgical techniques to relieve RVOTO by means of infundibular resection and patch enlargement of the outflow tract and pulmonary annulus often render the pulmonary valve incompetent. Pulmonary regurgitation is the commonest residual lesion following complete repair in infancy.

Although pulmonary regurgitation is well tolerated early after surgery, it results in ventricular dilatation and, ultimately, right ventricular dysfunction in the long term. Late survivors also have an increased risk of developing ventricular arrhythmias as a result of scarring of the right ventricle related to corrective surgery. Arrhythmias requiring intervention has been reported in more than 40 % of survivors late after repair [3]. Sudden death remains the commonest cause of late death, and is most likely arrhythmogenic in etiology. Therefore, the need to restore pulmonary valve competence and reduce the arrhythmia burden have become the most important indications for late interventions in this disease.

Diagnosis and Imaging

Transthoracic echocardiography (TTE) is the first line imaging to assess the degree of pulmonary regurgitation, RV volume overloading, and any residual lesion such as RV outflow tract stenosis, pulmonary artery stenoses and ventricular septal defect. Echocardiography can also be used to assess tricuspid regurgitation, and thus estimate right ventricular and pulmonary artery pressure. Competence of the aortic valve, aortic root dilatation and ventricular function can also be assessed semi-quantitatively. We routinely perform a bubble contrast study to look for shunting at atrial or ventricular level using trans-esophageal echocardiography prior to cardiopulmonary bypass as this may alter our cannulation strategy.

Cardiac MRI (CMR) has emerged as the most important imaging for accurate quantification of pulmonary regurgitation, right ventricular volume and function. Branch PA anatomy can also be shown clearly and differential flow to each lung can be calculated to assess the functional effect of any branch stenosis visualized. Currently, CMR is performed routinely for each of our patients referred for pulmonary valve replacement. Gadolinium enhancement can demonstrate myocardial fibrosis, and provide useful information on the extent of myocardial scarring. There is some evidence to suggest that this information can be used as risk stratification for tachydysrhythmia and sudden cardiac death. Besides precise volumetric and hemodynamic evaluation, the proximity of cardiac structures/conduit to the sternum can also be assessed and is relevant to the planning of sternal reentry. MRI is not universally applicable of course in the context of patients with a pacemaker.

Cardiac catheterization may not be required unless percutaneous intervention is planned such as a percutaneous pulmonary valve implantation or stenting of the branch

pulmonary arteries. Angiography may also be considered to exclude coronary artery disease in older patients. In the latest guideline for ACHD (ESC 2010), pre-operative coronary angiography is recommended for men older than 40 years, post-menopausal women, and in patients with risk factors or sign of ischemic heart disease [4]. CT-angiography of the coronary anatomy is efficient in excluding significant CAD with high negative predictive value and can be used as a reliable alternative to coronary angiography in non-high risk patients without risk factors or symptoms of CAD [5, 6]. However, CT can overestimate the degree of any atherosclerotic obstruction, and before any decision making regarding revascularisation, coronary angiography is advised and fractional flow reserve quantification may be required in this setting [6].

Other useful investigations, which are sometimes performed in our practice include Holter monitoring, electrophysiology laboratory study and cardiopulmonary exercise testing. A formal cardiopulmonary exercise test provides objective exercise capacity (time, maximum oxygen uptake, peak oxygen consumption - peak VO_2) and can provide useful prognostic information, particularly regarding the timing of intervention [7]. Formal and serial exercise testing may also be useful to assess symptomatology particularly in the context of sedentary patients, which make up a large proportion of this group of patients.

Pre-operative Checklist

- Patency of peripheral vessels e.g., femoral vessels for peripheral cannulation if required
- The proximity of RV or any valved conduit behind the sternum to predict risk of cardiac injury on sternal re-entry.
- Electrophysiologic study may be needed if there is any history of arrhythmia.
- Intracardiac shunt: PFO/ASD, VSD to plan cannulation strategy and the need to arrest the heart.
- The anatomical relation between the aorta and pulmonary artery. A main pulmonary artery, which is rotated more posteriorly will require more extensive dissection to mobilize the heart.
- Aberrant coronary artery crossing the RV outflow tract
- Branch PA stenosis needing concomitant patch enlargement or stent insertion
- Tricuspid valve regurgitation – severity of regurgitation, degree of annular dilatation, any leaflet prolapse

Indications and Timing for Pulmonary Valve Replacement (Table 12.1)

The indications for replacing the pulmonary valve continue to evolve and the tendency has moved towards earlier intervention before irreversible myocardial damage has occurred. Pulmonary regurgitation (PR) remains the most common

Table 12.1 Indications for pulmonary valve implantation

PVR is indicated in the presence of criteria from I + II *OR* I + III
(I) Severe pulmonary regurgitation and/or stenosis
PR grading based on CMR: mild (regurgitation fraction, RF < 20 %), moderate (RF 20–40 %), and severe (RF > 40 %); Echo: broad regurgitant jet and diastolic retrograde flow seen at branch PA in the parasternal short axis RVOT view indicates severe PR
PS: severe stenosis as indicated by RV systolic pressure >60 mmHg, TR velocity >3.5 m/s
(II) Symptom(s): Dyspnea, reduced exercise capacity, heart failure, or arrhythmia
(III) In the absence of symptom:
Decrease in objective exercise capacity, VO_2 max <70 % of gender-age predicted or a decline >20 % on serial testing
Sustained atrial/ventricular arrhythmias
ECG CRITERIA: QRS duration 180 ms, QRS prolongation >3.5 ms/year
MRI CRITERIA: RVEF <40 %, RVESV > 80 mL/m^2, RVEDV >150 mL/m^2 (Z-score >4),
Progressive RV dysfunction or dilation on serial imaging
RVOT aneurysm
Tricuspid regurgitation: at least moderate or progressive TR
Residual RVOTO: RVSP > 2/3 systemic, RV systolic pressure >80 mmHg (TR velocity >4.3 m/s), branch PA stenosis
Coexisting cardiac lesions requiring surgery: Significant residual shunt (Qp:Qs > 1.5:1) or with LV volume overloading, severe aortic regurgitation (with symptoms or LV dysfunction) and/or aortic root dilatation

indication for pulmonary valve replacement (PVR) but it may also be indicated for residual RVOTO or mixed disease. Magnetic resonance imaging (MRI) has enabled precise quantification of RV volumes and reports have suggested that once the indexed right ventricular end-diastolic volume (RVEDV) exceeds 170 ml/m^2 or the end-systolic volume (RVESV) is greater than 85 ml/m^2 remodeling of the ventricle is unlikely even when the pulmonary valve competency is restored [8]. Therefore, PVR should be undertaken before these thresholds were reached to increase the likelihood that the RV volume will normalize after PV replacement and indexed RVEDV of 150 ml/m^2 has been recommended as a practical cut-off for intervention [8–10].

The reason why some patients appear more symptomatic with lesser degree of PR, and some with severe PR remain asymptomatic is not well understood. This paradox continues to confound decision-making but attempts to objectify exercise tolerance with cardiopulmonary exercise testing maybe helpful looking longitudinally in individual patients. The presence of severe PR alone does not necessarily indicate the need for pulmonary valve replacement, and, notwithstanding the potential subjectivity of symptoms mentioned above, the presence of symptoms or other clinical and imaging criteria need to be taken into account in decision making. Both the American (AHA 2008) and European (ESC 2010) guidelines for grown-up congenital heart patients advocated pulmonary valve implantation only in severe PR in symptomatic patients (Class 1 recommendation) [11, 12]. In the absence of symptoms, AHA and ESC recommend surgery when there is evidence of moderate to

severe progressive RV dilatation, RV dysfunction or sustained arrhythmia (Class IIa). The presence of less than severe PR is not addressed by current guidelines and the indications for valve replacement requires individualized decision-making.

Surgical Techniques for Pulmonary Valve Replacement

Pulmonary valve replacement is performed under cardiopulmonary bypass. In the absence of an intracardiac shunt, it can be accomplished on a beating heart using a single right atrial cannulation with a two-stage venous cannula. Otherwise, bicaval cannulation and cardioplegic arrest are required to close any intracardiac shunt. The residual VSD or ASD/PFO is closed first following cardioplegic arrest and the subsequent pulmonary valve replacement can be performed on a beating heart.

Following redo sternotomy, dissection is undertaken first to expose the cannulation sites, and to free the areas around the aorta, pulmonary artery, right atrium and the anterior right ventricle. If bicaval cannulation is planned, then the area around the IVC will also need to be exposed. The temperature of the patient on cardiopulmonary bypass should not be lower than 32 °C (Figs. 12.1, 12.2, 12.3, 12.4 and 12.5).

Sizing and Choice of Prostheses for PVR

The options for pulmonary valve replacement include mechanical, bioprosthetic or homograft (aortic or pulmonary). The ideal valve choice does not exist. In adolescents and adult patients, we prefer the use of a bioprosthetic valve over homografts due to their easy availability, preparation, and durability in the adult population. Additionally, the use of a bioprosthesis has the potential to limit further redo surgery by allowing transcatheter valve implantation (Medtronic Melody™ or Edwards SAPIEN™ Pulmonic Transcatheter Heart Valve) when this becomes necessary in the future. The use of a transannular patch to enlarge the pulmonary valve annulus allows a larger prosthesis to be implanted. Typically, in adult patients, the largest available aortic bioprosthesis is chosen, but consideration is also given to the valve size, which will allow the largest transcatheter valve to be implanted if required in the future (Fig. 12.6 and Table 12.2).

Studies suggest better durability of a bioprosthetic valve over a homograft when a larger size (>19 mm) can be implanted [13]. Although a bioprosthetic valve has a shorter durability compared to mechanical valve, it allows future transcatheter valve-in-valve implantation [14, 15]. Bioprostheses have an excellent 5-year durability with over 94 % freedom from valve replacement. This decreases to 36–51 % at 10-year follow-up [14, 16]. In a contemporary analysis, younger age appears to be the primary determinant of the durability of bioprosthetic valves in the pulmonary position [16]. In this study, all the structural valve

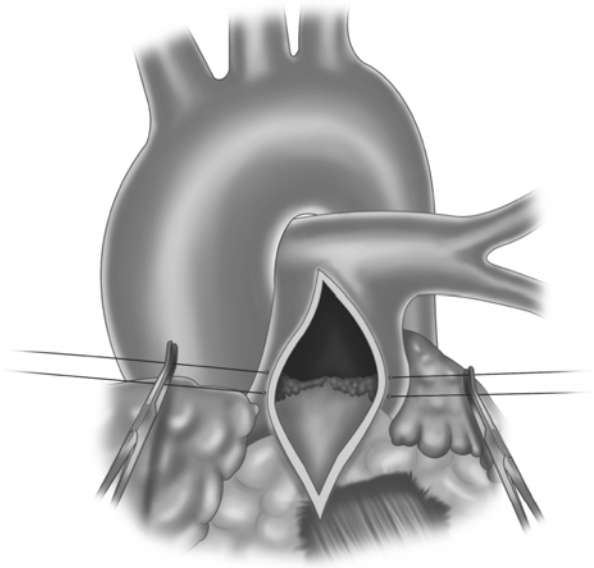


Fig. 12.1 An incision is made into the previous transannular patch, or in the absence of one, a transannular incision is created. The size of the incision should be adequate and starts at or just below the bifurcation of the main pulmonary artery. This technique augments the annulus and the right ventricular outflow tract to facilitate the implantation of a larger prosthesis

failure was observed in patients younger than 15 years, and re-intervention was only required in one patient older than 15 years due to endocarditis. There is no apparent advantage of using a pericardial valve over porcine valve, and both seem to be comparable [16]. Mechanical prostheses are more durable but large studies and long-term follow-up of their use in the pulmonary position is lacking. Besides anticoagulant related bleeding, thrombogenic complications have been the major concern associated with mechanical prostheses. In contemporary studies, valve thrombosis was not reported with adequate anticoagulation, but severe right ventricular dysfunction has been linked to a higher thrombogenic risk [15, 17]. Mechanical prostheses are also not immune from dysfunction, which can be due to thrombosis, fibrosis, or pannus formation and mandatory replacement with redo surgery is required. A percutaneous option is not possible in this context. Nevertheless, use of a mechanical valve may be justified in patients who are already on anticoagulation for other reasons such as a coexisting mechanical heart valve, compliant to anticoagulation medications, and those who have previously demonstrated accelerated degeneration of a bioprosthesis [15–18]. A generation of mechanical valves that are less thrombogenic and require lower levels of anti-coagulation, and the use of newer generation anticoagulants such as dabigatran may encourage the wider use of mechanical valves in the pulmonary position.

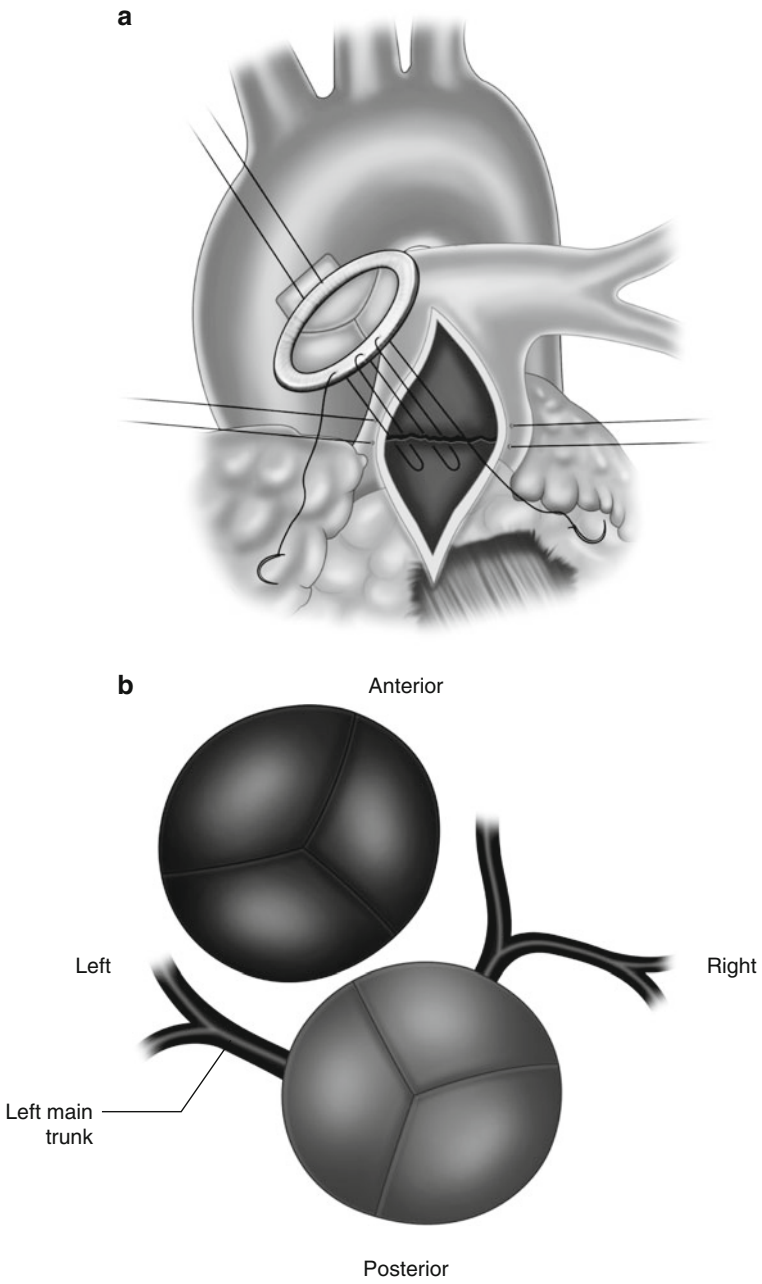


Fig. 12.2 PV leaflets are not routinely excised and can be used as tissues to anchor valve sutures during implantation. The prosthesis is first sutured on the posterior annulus starting with 4/0 Prolene™ using a continuous suture. We prefer to start closer to Y and suture towards X before parachuting the prosthesis down after several stitches. Deep bites on the posterior annulus are avoided to prevent any inadvertent injury to the left common coronary trunk which courses behind the main pulmonary artery. (**a**) Suturing valve in place (**b**) Relationship of pulmonary valve to left main coronary artery

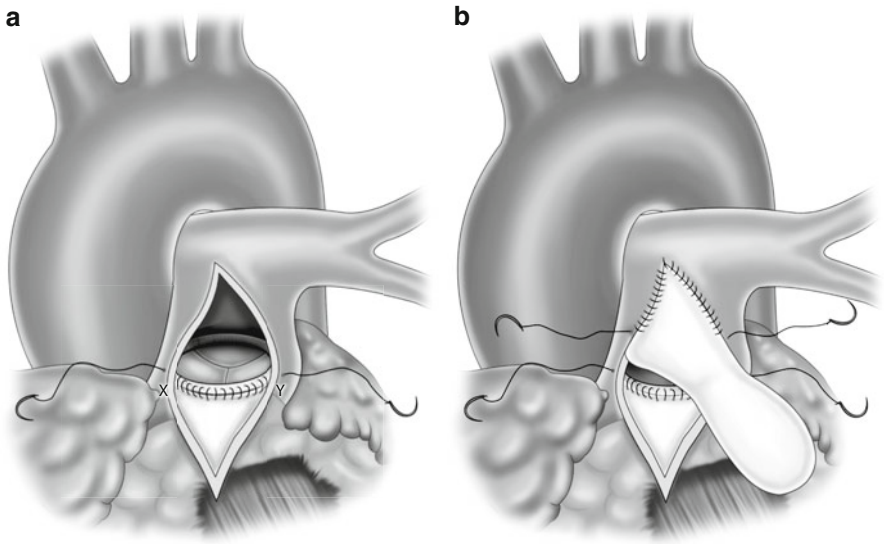


Fig. 12.3 A bovine pericardial patch (Peri-Guard®, Synovis, Deerfield, IL, USA) is measured to fit the size of the augmented RVOT. The patch is first sutured at the apex adjacent to the pulmonary artery bifurcation using double-armed 4/0 Prolene™ and brought on each side towards the prosthesis. At points X and Y, the needle is brought out from the edge of ventriculotomy and tied to the previous stitch. (**a**) Running 4/0 ProleneTM sutures anchoring the valve in place brought to outside the opened pulmonary artery (**b**) Pericardial patch being sewn in place

Surgical Ablation for Ventricular Tachycardia during Redo Surgery for Tetralogy of Fallot

In a multi-institutional study of 556 patients, 43 % of those older than 18 years of age had a documented sustained arrhythmia during adulthood and/or an arrhythmia needing intervention late after repair [3]. This may occur as a supraventricular and/or ventricular tachyarrhythmia with a prevalence of 20.1 % and 14.6 % respectively [3]. In several other studies, the incidence of late sustained ventricular tachycardia (VT) is 3–14 % and the risk of sudden death is 3–6 % amongst survivors at 25–30 years after corrective surgery [2, 19–21]. Due to a significant incidence of tachyarrhythmia among late survivors, cryoablation of the RVOT pathway is sometimes required concomitantly with the pulmonary valve replacement. The presence of ventricular fibrosis was shown to be the major predisposing factor for inducible monomorphic ventricular tachycardia in non-ischemic cardiomyopathy patients [22]. Based on intra-operative mapping studies, the mechanism for inducible VT in post repair patients is most commonly secondary to monomorphic and macro-reentrant circuits around the ventriculotomy scars or surgical patches [23].

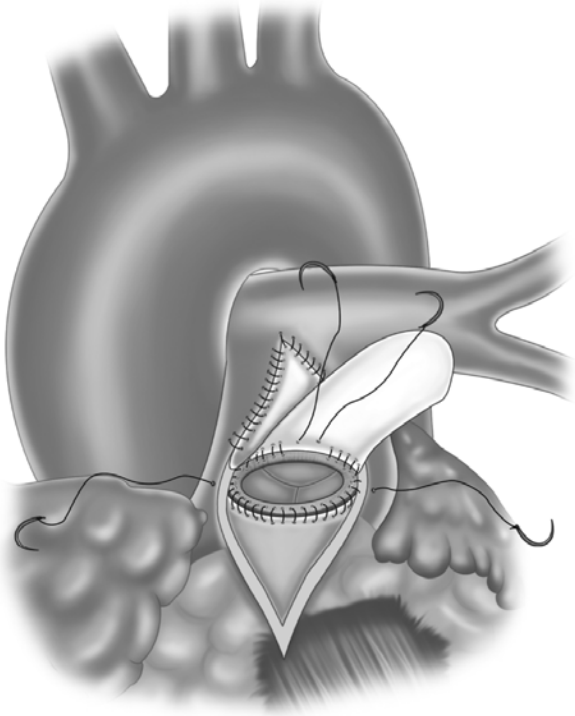


Fig. 12.4 The prosthesis is adjusted so that it is seated in a desirable position before it is fixed anteriorly. One of the needles at junction X/Y will be used to suture the prosthesis anteriorly and the other needle to suture the pericardial patch to the infundibulum. The needle is passed from outside to inside through the pericardial patch and the prosthesis. Once the prosthesis is secured anteriorly, the bottom half of the pericardial patch is sutured to the edge of the ventriculotomy. Before the patch is fixed on its lower half, the prosthetic valve leaflets are checked to ensure that they are not caught by any suture

Supraventricular arrhythmias can occur in the form of intra-atrial reentrant tachycardia or atrial fibrillation [3]. Typical atrial flutter is the most common form of intra-atrial reentrant tachycardia and usually originates from the isthmus between the tricuspid valve and inferior vena cava [24]. The occurrence of sustained ventricular tachyarrhythmia is the most concerning and carries the highest risk of sudden cardiac death, a leading cause of mortality among late survivors after repair of tetralogy of Fallot. In the most recent multi-centers study (N = 873), RV hypertrophy, ventricular dysfunction and atrial tacharrhythmia are predictive of sustained ventricular tachycardia and sudden death in adults with repaired Fallot [25]. These high-risk patients should not be managed with anti-dysrhythmia agents alone. More definitive therapy in the form of catheter or surgical ablation and an implantable cardiac defibrillator is warranted [4, 12].

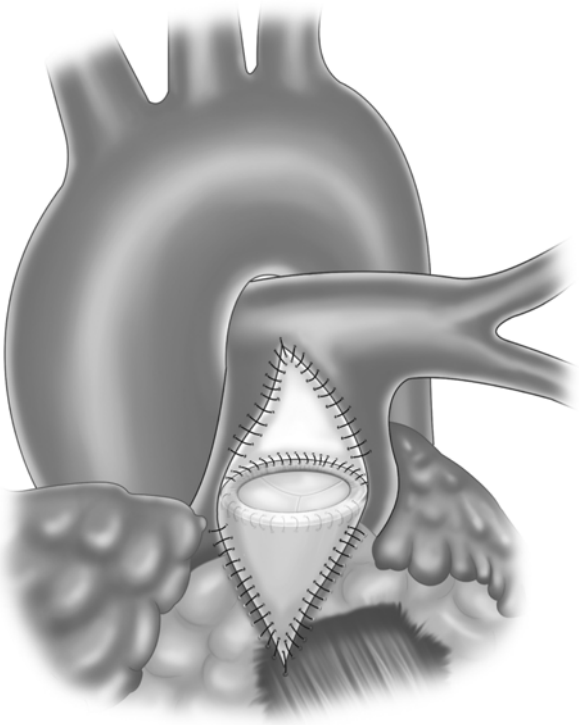


Fig. 12.5 When the pericardial patch is sutured down, a closer bite is taken on the pulmonary artery and ventriculotomy edges as opposed to a larger bite on the patch itself. This technique allows the pericardial patch to billow when the heart is full and ejecting

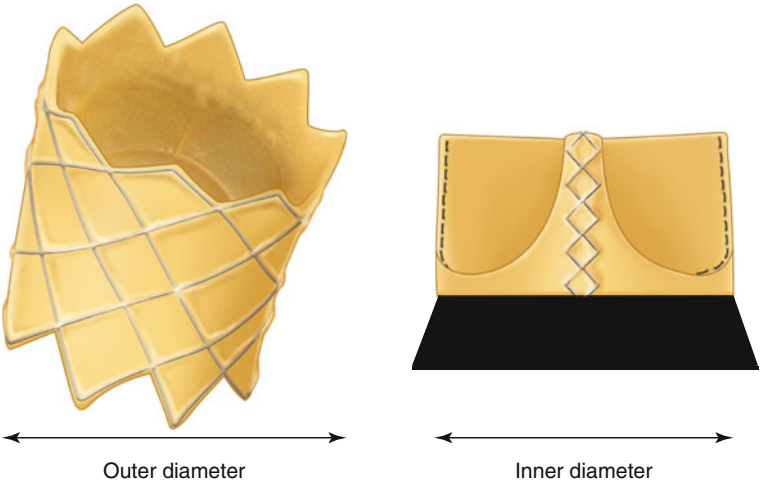
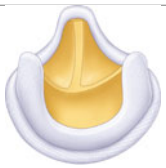


Fig. 12.6 The inner diameter of an aortic bioprosthesis should accommodate the outer diameter of a Melody™ valve. A size 27 mm Sorin Aortic Mitroflow™ (*right*) (outer diameter of 27.3 mm, inner diameter 22.9 mm) will facilitate the largest available Melody™ valve (*left*) (a Melody™ valve deployed with a 22 mm balloon diameter has an outer diameter of approximately 22–24 mm

Table 12.2 The dimensions of the Melody™ valve system and available aortic bioprostheses (the sizes which accommodate the largest Melody™ valve 22 mm are highlighted)

Melody® transcatheter pulmonary valve, Medtronic Inc		Valve size	Outer ballon pressure-outer diameter
		18 mm (smallest)	1 atm – 17.93 mm
			4 atm – 20.06 mm
		20 mm	1 atm – 19.65 mm
			4 atm – 22.42 mm
		22 mm (largest)	1 atm – 21.80 mm
			2 atm – 22.79 mm
			3 atm – 24.06 mm
Carpentier-Edwards PERIMOUNT aortic heart value		Valve size	Inner diameter
		19 mm (smallest)	18 mm
		21 mm	20 mm
		23 mm	22 mm
		25 mm	24 mm
		27 mm	26 mm
		29 mm (largest)	28 mm
Sorin aortic mitroflow		Valve size	Inner diameter
		19 mm (smallest)	15.4 mm
		21 mm	17.3 mm
		23 mm	19.0 mm
		25 mm	21.0 mm
		27 mm (largest)	22.9 mm
Medtronic Hancock II aortic bioprosthesis		Valve size	Inner diameter
		21 mm (smallest)	18.5 mm
		23 mm	20.5 mm
		25 mm	22.5 mm
		27 mm	24.0 mm
		29 mm (largest)	26.0 mm

Indications for Electrophysiologic (EP) Study and Ablation

The American and European Guidelines (AHA 2008, ESC 2010) recommend electrophysiologic study and/or ablation for symptomatic patients with suspected or documented clinical arrhythmia (atrial or ventricular) [4]. Unexplained syncope is alarming and symptoms such as palpitations and dizziness should raise suspicion for serious arrhythmias and prompt assessment including cardiac catheterization and electrophysiology studies. A catheter ablation of the ventricular tachycardia circuit may be performed if stable monomorphic ventricular tachycardia can be induced and sustained sufficiently to permit mapping in the electrophysiology laboratory [12]. If unsuccessful, surgical ablation with or without intraoperative mapping can be performed as part of the pulmonary valve replacement surgery [12]. A concomitant Cox-Maze procedure during surgery should also be considered in the

presence of atrial flutter or fibrillation. A more aggressive approach to perform electrophysiologic studies in all patients undergoing surgical pulmonary valve replacement is adopted in some centers, but whether this aggressive protocol will influence late outcome remains to be seen.

Surgical Techniques for Cryoablation of RVOT pathway

Cryoablation of RVOT pathways to ablate macro re-entrant circuits is usually undertaken with pulmonary valve replacement under cardiopulmonary bypass. The cryo-ablation is performed after the RVOT is opened and prior to implanting a prosthetic valve. Several ventricular isthmuses have been identified as critical in the pathogenesis of re-entrant monomorphic VT and ablation lines are performed in these critical regions (Fig. 12.7) [22, 26, 27]. A cryoablation probe is applied at each ablation line for 60 s, with caution to avoid collateral damage to adjacent structures. The cryothermy (Cardioblate® CryoFlex™ Argon-powered Surgical Ablation System, Medtronic, Inc) consists of a metal probe, which can be rapidly cooled using Argos gas to reach −150 °C. The metal probe is flexible with an adjustable insulation sheath, which is malleable and conforms to the endocardial

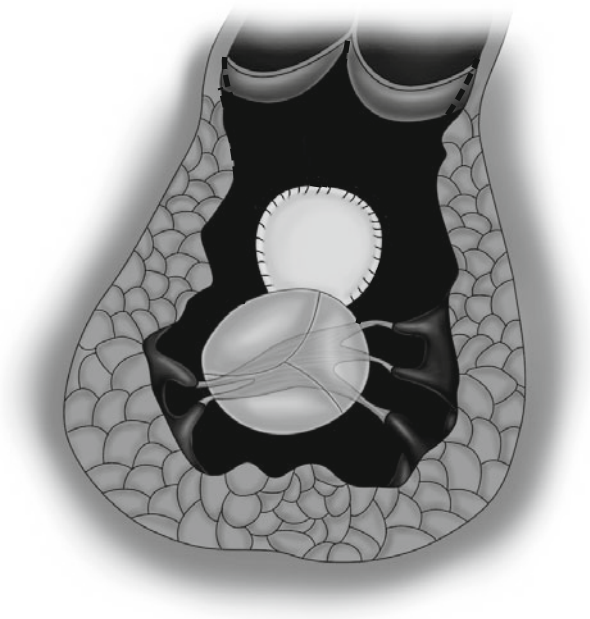


Fig. 12.7 Ablation lines in ventricular isthmuses which serve as critical substrate for VT: (i) from the posterior pulmonary annulus to the VSD patch (ii) from both edges of the ventriculotomy to the VSD patch (iii) from the edges of ventriculotomy on each side towards tricuspid annulus

surface to achieve good contact. The ideal application time is unknown, but in our experience we have observed complete heart block with a longer contact time, hence each ablation is reserved to 60 s. After completion of each ablation line, warm saline is used to rinse the probe, which allows easy removal from the endocardial surface.

Tetralogy of Fallot with Pulmonary Atresia

At the severe end of the spectrum of tetralogy of Fallot the pulmonary valve is completely atretic in approximately 20 % of patients [28]. This group represents a complex spectrum of malformation in itself and the use of a conduit to restore right ventricle to pulmonary artery continuity when confluent pulmonary arteries are present or to grow the pulmonary arteries is an important part of the surgical management.

An ideal conduit does not exist and patients require repeated intervention throughout life. The homograft conduits are used in countries where they are readily available [29, 30]. The early experience of using aortic homograft in pulmonary position was off set by rapid calcification especially in the very young patients and also lack of reliable preservation technique [30]. However, the durability of homograft has improved with cryopreservation technology and introduction of pulmonary homograft. A pulmonary homograft is thinner with less transvalvular gradient, is easier to work with, begets less calcification, and clinical studies suggest that it is more durable than an aortic homograft [30, 31]. More recently, decellularized homografts have been introduced to reduce immunogenicity and potentially attenuate the host–graft response and may improve homograft longevity [32].

Homograft conduits are not universally available and in some countries homograft availability is scarce thus alternatives are required. Xenograft and synthetic material have been used with satisfactory results in clinical series, either implanted as an extracardiac conduit or in an orthotopic position such as in the Ross procedure [33]. These various alternatives include stented (e.g., Hancock, Carpentier Edwards), or stentless xenografts (e.g., Medtronic Freestyle, RVOT élan, Contegra valved bovine jugular vein) [33]. Non-valved conduits have also been used but are associated with the deleterious long-term effects of free pulmonary regurgitation.

Each conduit has a finite life span. Whilst there is no single conduit that has been shown to be consistently superior compared to the others, younger age (under 1-year old), smaller conduit size, lower weight, and extra-anatomical conduit implantation had emerged as important risk factors across studies [34–36]. A truly ideal conduit that grows and does not degenerate will not exist in the foreseeable future. Currently, an acceptable conduit to restore RV to PA continuity should be one that will provide durable competent valve function without obstruction, be resistant to calcification, easy to handle, and is hemostatic when implanted.

Redo Surgery in Patients with Right Ventricle to Pulmonary Artery Conduit

RV to PA conduit may need replacement, either because it became stenotic, incompetent, or both. The criteria for PV replacement as outlined above can be used to decide timing of intervention. Redo surgery is not always necessary, as the availability of percutaneous transcatheter pulmonary valve has reduced the need of resternotomy to replace an old conduit in the current era. When the percutaneous option is deemed not suitable (common reason for this is the conduit size is too big for the largest available Melody™ valve), a redo surgery will then become necessary.

In the presence of an old, calcified conduit, sternal re-entry needs to be approached with an extreme caution due to high risk of the conduit being adherent to the back of sternum. We routinely placed a Goretex patch overlying the conduit in the first surgery to reduce risk of complication during resternotomy. Pre-operative imaging provides information of the risk of conduit injury and pre-emptive measures may be required, including preparation of femoral or iliac vessel to go on bypass immediately in any event of conduit injury or alternatively, peripheral cardiopulmonary bypass can be instituted prior to sternal re-entry.

The old conduit does not necessarily need to be explanted and replaced with a new one. Instead, the surgeon can simply lay open the previous conduit and implant a stented bioprosthetic valve in-situ with or without an overlying patch to fit in an appropriate size valve (Fig. 12.8). In our experience, this is achievable even in young children and this

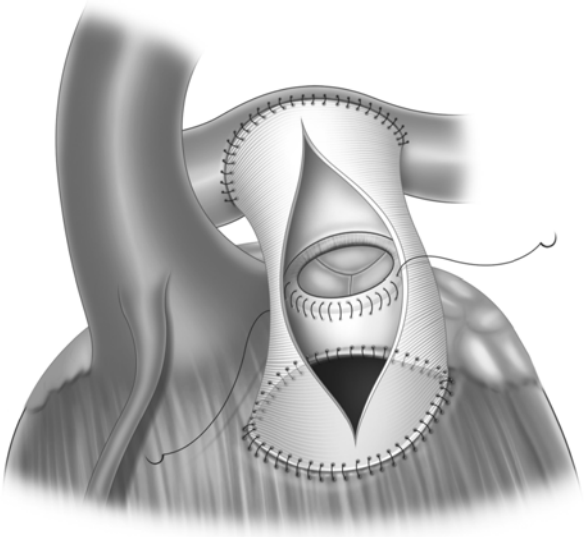


Fig. 12.8 The old conduit is opened anteriorly and the prosthesis is sutured in-situ. This can be achieved providing that the old conduit was not too calcified for implantation of the prosthesis. A patch can be used to accommodate larger prosthesis is needed

strategy will also allow replacement with a percutaneous valve in the future. Occasionally, the whole conduit may need to be explanted and replaced in the setting of endocarditis, when it is excessively aneurysmal or when it is too calcified to implant a prosthesis or achieve a hemostatic suture closure. When conduit replacement is required, it's important to ensure that the stent of the valve does not impinge on the left coronary artery posteriorly ("S" in Fig. 12.9). The conduit is trimmed as such that the valve within the conduit sits close to the central PA bifurcation (Fig. 12.9). The surgery can be usually completed on a beating heart when there is no intra-cardiac shunt present.

The Role of Percutaneous Pulmonary Valve Replacement

Since the first report of percutaneous pulmonary valve replacement in 2000, Melody™ valve, Medtronic Inc (previously by VenPro Corp) has been used widely as an alternative to surgical replacement of RV-PA conduit [37]. The available

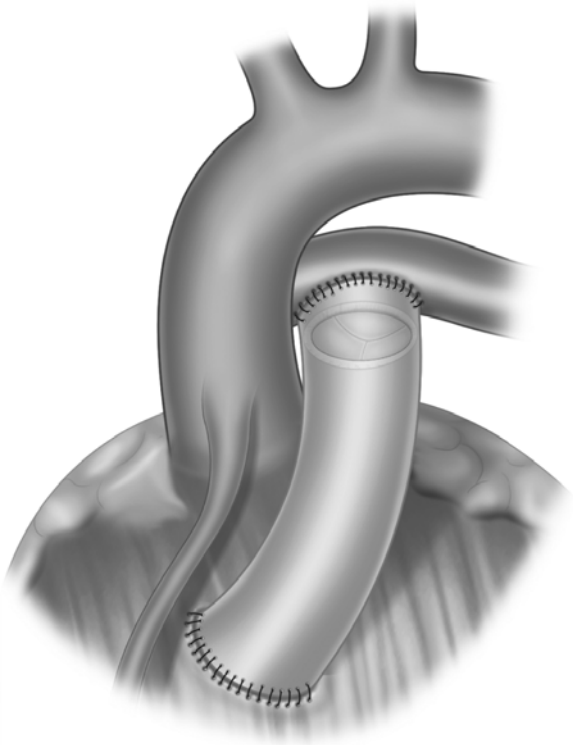


Fig. 12.9 RV to PA conduit implantation: the conduit is trimmed so that the valve sits close to the bifurcation. The LCA can be potentially compressed by stent (S) of the valve and sometimes, removal of the stent may be required to allow a stentless implantation

technology has been augmented with comparable results by the more recent use of the Edwards SAPIEN™ valve [38]. Transcatheter valve implantation has been shown to improve hemodynamics by reducing obstruction and abolishing regurgitation resulting in mechanical and electrical 'remodeling' of the right ventricle [39–42]. The topic of transcatheter pulmonary valve replacement is comprehensively covered in another chapter of this book. We would like to briefly state its role in the context of surgical pulmonary valve replacement in our practice.

It is acknowledged that any type of valve replacement in any position will have finite longevity before failure with stenosis or regurgitation or both intervene. The longevity and hemodynamic efficiency of a valve in the pulmonary position may be constrained by the younger age of the patient, the effects of somatic growth, the existing ventricular substrate, the relationship of the great arteries, the branch pulmonary artery anatomy, proximity of the implant to the posterior aspect of the sternum and the more imponderable chronic effects of inflammation and its accompanying results of neo-intima and calcification. The inevitability of repeated surgery in this group of patients and the prospect of procrastinating reoperation using a technique, which in practical terms did not make subsequent surgery more hazardous supported the inception of catheter based technology. This has been incorporated into the clinical problem of conduit dysfunction of predominant stenosis, predominant regurgitation or, more commonly, combined stenosis and regurgitation.

At our institution, we support an active transcatheter pulmonary valve replacement service and see our roles as complementary. We have therefore tailored our approach such that when our surgically implanted valve fails as a consequence of the factors above, we have effectively left a legacy of a 'landing site' for the implantation of a transcatheter valve. A Sorin Aortic Mitroflow™ in the pulmonary position has an outer diameter of 27 mm and inner diameter of 22.9 facilitates the interventional deployment of the Melody™ using the largest (22 mm inner diameter) delivery system (Ensemble Medtronic) ensuring maximal gradient reduction when present and greatest available competent valve area (Fig. 12.6, Table 12.2). Arguably, with the recently available Edwards SAPIEN™ valve for transcatheter deployment in the pulmonary position one could justify, where substrate permits, to use even larger bioprostheses, as the SAPIEN™ valve is available in 23 and 26 mm diameters.

Surgical Outcomes after Pulmonary Valve Replacement with or without Ablation Surgery

Despite the perceived benefits of PVR, current evidence has not shown a survival benefit. Nonetheless, a meta-analysis of 3118 patients demonstrated: (1) Remodeling of the RV with improvement in volume and function; (2) Improvement in left ventricular function; (3) Decreased QRS duration and (4) Improvement in functional status. Surgery can be performed with low mortality and the pooled 30-day mortality from this meta-analysis was 0.87 % (47 studies; 27 of 3100 patients). At midterm follow-up, the pooled 5-year mortality was 2.2 % (24 studies; 49 of 2231

patients) and the pooled 5-year re-intervention rate was 4.9 % (15 studies; 88 of 1798 patients) [43].

Unlike studies in atrial fibrillation following the Cox-Maze procedure, available outcome data following ablation for VT are from small series, largely non-surgical, with mixed cohorts of structural and non-structural heart disease [44, 45]. Early results following ablation are satisfactory but long-term success may be seen in only 60 % of patients [45]. Ablation therapy should not be considered as sole therapy to prevent sudden cardiac death in high-risk patients and an implantable defibrillator (ICD) should be considered [46]. Those patients with documented sustained VT or cardiac arrest are at high risk and should receive an ICD regardless of the early success of ablation therapy as the late outcome of ablation is too uncertain [12]. In others, a repeat EP assessment and implantation of an ICD only in the presence of inducible VT seems to be a reasonable approach.

Tricuspid Regurgitation in Late Survivors

Tricuspid regurgitation can be seen in late survivors who present for pulmonary valve replacement. Moderate to severe regurgitation has been reported in one-fifth to one-third of survivors of Fallot surgery [47, 48]. Despite these observations, in our experience, the need of concomitant tricuspid valve surgery has been very rare. Tricuspid regurgitation is usually secondary to annular dilatation consequent on right ventricular enlargement from severe pulmonary regurgitation, and pulmonary valve replacement alone is usually all that is required to reverse this process. Studies have suggested that RV remodeling occurs with PV replacement, and improvement in TR is usually observed as a consequence of this [49]. Much less commonly, TR can occur as a result of injury to the valve and supporting apparatus during the initial reparative surgery. Tricuspid leaflet injury can occur during a trans-atrial approach to close the VSD and to relieve RV outflow tract obstruction. Injury of the chordae tendinae can also occur during the resection of obstructive muscle bundles, leading to TR.

We reserve surgery for TR only when it is severe and we will strive to avoid prosthetic valve replacement in this position. Our philosophy is to reduce the severity of TR and to allow any lesser degree of residual TR to improve with RV remodeling, which we believe is always better than achieving perfect tricuspid competency with a prosthetic valve replacement. A repair is tailored according to the underlying mechanism of the TR. An annuloplasty ring (e.g., Edwards MC3) sparing the AV node region is usually adequate to address annular dilatation. When compared with the De Vega technique without a ring, the placement of an annuloplasty ring has been associated with better long-term survival and freedom from recurrent TR [50]. Less frequently, leaflet repair, or neo-chordae may be needed when tricuspid valve leaflets or subvalvar apparatus have been injured.

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References

1. Lillehei CW, Cohen M, Warden HE, Read RC, Aust JB, Dewall RA, Varco RL. Direct vision intracardiac surgical correction of the tetralogy of Fallot, pentalogy of Fallot, and pulmonary atresia defects; report of first ten cases. Ann Surg. 1955;142:418–42.
2. Nollert G, Fischlein T, Bouterwek S, Böhmer C, Klinner W, Reichart B. Long-term survival in patients with repair of tetralogy of fallot: 36-year follow-up of 490 survivors of the first year after surgical repair. J Am Coll Cardiol. 1997;30:1374–83.
3. Khairy P, Aboulhosn J, Gurvitz MZ, Opotowsky AR, Mongeon FP, Kay J, Valente AM, Earing MG, Lui G, Gersony DR, Cook S, Ting JG, Nickolaus MJ, Webb G, Landzberg MJ, Broberg CS, for the Alliance for Adult Research in Congenital Cardiology (AARCC). Arrhythmia burden in adults with surgically repaired tetralogy of Fallot: a multi-institutional study. Circulation. 2010;122:868–75.
4. Baumgartner H, Bonhoeffer P, De Groot NMS, de Haan F, Deanfield JE, Galie N, Gatzoulis MA, Gohlke-Baerwolf C, Kaemmerer H, Kilner P, Meijboom F, Mulder BJM, Oechslin E, Oliver JM, Serraf A, Szatmari A, Thaulow E, Vouhe PR, Walma E, Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC), Association for European Paediatric Cardiology (AEPC), ESC Committee for Practice Guidelines (CPG). ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J. 2010;31:2915–57.
5. Task Force Members, Montalescot G, Sechtem U, Achenbach S, Andreotti F, Arden C, Budaj A, Bugiardini R, Crea F, Cuisset T, Di Mario C, Ferreira JR, Gersh BJ, Gitt AK, Hulot J-S, Marx N, Opie LH, Pfisterer M, Prescott E, Ruschitzka F, Sabate M, Senior R, Taggart DP, van der Wall EE, Vrints CJM, ESC Committee for Practice Guidelines, Zamorano JL, Baumgartner H, Bax JJ, Bueno H, Dean V, Deaton C, Erol C, Fagard R, Ferrari R, Hasdai D, Hoes AW, Kirchhof P, Knuuti J, Kolh P, Lancellotti P, Linhart A, Nihoyannopoulos P, Piepoli MF, Ponikowski P, Sirnes PA, Tamargo JL, Tendera M, Torbicki A, Wijns W, Windecker S, Document Reviewers, Valgimigli M, Claeys MJ, Donner-Banzhoff N, Frank H, Funck-Brentano C, Gaemperli O, Gonzalez-Juanatey JR, Hamilos M, Husted S, James SK, Kervinen K, Kristensen SD, Maggioni AP, Pries AR, Romeo F, Rydén L, Simoons ML, Steg PG, Timmis A, Yildirir A. 2013 ESC guidelines on the management of stable coronary artery disease: the Task Force on the management of stable coronary artery disease of the European Society of Cardiology. Eur Heart J. 2013;34:2949–3003.
6. Task Force on Myocardial Revascularization of the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS), European Association for Percutaneous Cardiovascular Interventions (EAPCI), Kolh P, Wijns W, Danchin N, Di Mario C, Falk V, Folliguet T, Garg S, Huber K, James S, Knuuti J, Lopez-Sendon J, Marco J, Menicanti L, Ostojic M, Piepoli MF, Pirlet C, Pomar JL, Reifart N, Ribichini FL, Schalij MJ, Sergeant P, Serruys PW, Silber S, Sousa Uva M, Taggart D. Guidelines on myocardial revascularization. Eur J Cardiothorac Surg. 2010;38 Suppl:S1–52.
7. Diller G-P, Dimopoulos K, Okonko D, Li W, Babu-Narayan SV, Broberg CS, Johansson B, Bouzas B, Mullen MJ, Poole-Wilson PA, Francis DP, Gatzoulis MA. Exercise intolerance in adult congenital heart disease: comparative severity, correlates, and prognostic implication. Circulation. 2005;112:828–35.
8. Therrien J, Provost Y, Merchant N, Williams W, Colman J, Webb G. Optimal timing for pulmonary valve replacement in adults after tetralogy of Fallot repair. Am J Cardiol. 2005;95:779–82.
9. Dave HH, Buechel ERV, Dodge-Khatami A, Kadner A, Rousson V, Bauersfeld U, Prêtre R. Early insertion of a pulmonary valve for chronic regurgitation helps restoration of ventricular dimensions. Ann Thorac Surg. 2005;80:1615–20; discussion 1620–1.
10. Buechel ERV. Remodelling of the right ventricle after early pulmonary valve replacement in children with repaired tetralogy of Fallot: assessment by cardiovascular magnetic resonance. Eur Heart J. 2005;26:2721–7.

11. McDonagh T. ESC guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J. 2010;31:2915–57.
12. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, Del Nido P, Fasules JW, Graham Jr TP, Hijazi ZM, Hunt SA, King ME, Landzberg MJ, Miner PD, Radford MJ, Walsh EP, Webb GD. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: executive summary: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to develop guidelines for the management of adults with congenital heart disease). Circulation. 2008;118:2395–451.
13. Dearani JA, Danielson GK, Puga FJ, Schaff HV, Warnes CW, Driscoll DJ, Schleck CD, Ilstrup DM. Late follow-up of 1095 patients undergoing operation for complex congenital heart disease utilizing pulmonary ventricle to pulmonary artery conduits. Ann Thorac Surg. 2003;75:399–411.
14. Lee C, Park CS, Lee CH, Kwak JG, Kim SJ. Durability of bioprosthetic valves in the pulmonary position: long-term follow-up of 181 implants in patients with congenital heart disease. J Thorac Cardiovasc Surg. 2011;142:351–8.
15. Stulak JM, Dearani JA, Burkhart HM, Connolly HM, Warnes CA, Suri RM, Schaff HV. The increasing use of mechanical pulmonary valve replacement over a 40-year period. Ann Thorac Surg. 2010;90:2009–15.
16. Chen X-J, Smith PB, Jaggers J, Lodge AJ. Bioprosthetic pulmonary valve replacement: contemporary analysis of a large, single-center series of 170 cases. J Thorac Cardiovasc Surg. 2013;146:1461–6.
17. Dos L, Muñoz-Guijosa C, Mendez AB, Ginel A, Montiel J, Padro JM, Subirana MT. Long term outcome of mechanical valve prosthesis in the pulmonary position. Int J Cardiol. 2011;150:173–6.
18. Stulak JM, Dearani JA. Technique of mechanical pulmonary valve replacement. Oper Tech Thorac Cardiovasc Surg. 2006;11:200–6.
19. Gatzoulis MA, Balaji S, Webber SA, Siu SC. Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study. Lancet. 2000;356:975–81.
20. Murphy JG, Gersh BJ, Mair DD, Fuster V. Long-term outcome in patients undergoing surgical repair of tetralogy of Fallot. N Engl J Med. 1993;329:593–9.
21. Aliot EM, Stevenson WG, Almendral-Garrote JM, Bogun F, Delacretaz E, Bella Della P, Hindricks G, Jaïs PJ. EHRA/HRS Expert Consensus on Catheter Ablation of Ventricular Arrhythmias: developed in a partnership with the European Heart Rhythm Association (EHRA), a Registered Branch of the European Society of Cardiology (ESC), and the Heart Rhythm Society (HRS); in collaboration with the American College of Cardiology (ACC) and the American Heart Association (AHA). Heart Rhythm, Oxford University Press; 2009. p. 771–817.
22. Misaki T, Tsubota M, Watanabe G, Watanabe Y, Matumoto Y, Ishida K, Iwa T, Okada R. Surgical treatment of ventricular tachycardia after surgical repair of tetralogy of Fallot. Relation between intraoperative mapping and histological findings. Circulation. 1994;90:264–71.
23. Kurzidim K, Schneider H-J, Kuniss M, Sperzel J, Greiss H, Berkowitsch A, Pitschner HF. Cryocatheter ablation of right ventricular outflow tract tachycardia. J Cardiovasc Electrophysiol. 2005;16:366–9.
24. Khairy P, Stevenson WG. Catheter ablation in tetralogy of Fallot. Heart Rhythm. 2009;6:1069–74.
25. Valente AM, Gauvreau K, Assenza GE, Babu-Narayan SV, Schreier J, Gatzoulis MA, Groenink M, Inuzuka R, Kilner PJ, Koyak Z, Landzberg MJ, Mulder B, Powell AJ, Wald R, Geva T. Contemporary predictors of death and sustained ventricular tachycardia in patients with repaired tetralogy of Fallot enrolled in the INDICATOR cohort. Heart. 2014;100:247–53.
26. Zeppenfeld K, Schalij MJ, Bartelings MM, Tedrow UB, Koplan BA, Soejima K, Stevenson WG. Catheter ablation of ventricular tachycardia after repair of congenital heart disease: electroanatomic identification of the critical right ventricular isthmus. Circulation. 2007;116:2241–52.

27. Moore JP, Seki A, Shannon KM, Mandapati R, Tung R, Fishbein MC. Characterization of anatomic ventricular tachycardia isthmus pathology after surgical repair of tetralogy of fallot. Circ Arrhythm Electrophysiol. 2013;6:905–11.
28. Ferencz C, Rubin JD, Mccarter RJ, Brenner JI, Neill CA, Perry LW, Hepner SI, Downing JW. Congenital heart disease: prevalence at livebirth. The Baltimore-Washington Infant Study. Am J Epidemiol. 1985;121:31–6.
29. Ross DN. Homograft replacement of the aortic valve. Lancet. 1962;2:487.
30. Campbell DN, Clark DR. Use of the allograft aortic valved conduit. Ann Thorac Surg. 1990;50:320–2.
31. Livi U, Kay P, Ross DN. The pulmonary homograft: an improved conduit for RVOT reconstruction. Circulation. 1986;74(Supplement 2):250.
32. Ruzmetov M, Shah JJ, Geiss DM, Fortuna RS. Decellularized versus standard cryopreserved valve allografts for right ventricular outflow tract reconstruction: a single-institution comparison. J Thorac Cardiovasc Surg. 2012;143:543–9.
33. Caldarone CA, McCrindle BW, Van Arsdell GS, Coles JG, Webb G, Freedom RM, Williams WG. Independent factors associated with longevity of prosthetic pulmonary valves and valved conduits. J Thorac Cardiovasc Surg. 2000;120:1022–30; discussion 1031.
34. Vitanova K, Cleuziou J, Hörer J, Kasnar-Samprec J, Vogt M, Schreiber C, Lange R. Which type of conduit to choose for right ventricular outflow tract reconstruction in patients below 1 year of age? Eur J Cardiothorac Surg. 2014;46:961–6.
35. Forbess JM, Shah AS, St Louis JD, Jaggers JJ, Ungerleider RM. Cryopreserved homografts in the pulmonary position: determinants of durability. Ann Thorac Surg. 2001;71:54–9.
36. Sierra J, Christenson JT, Lahlaidi NH. Right ventricular outflow tract reconstruction: what conduit to use? Homograft or Contegra? Ann Thorac Surg. 2007;84:606–10.
37. Bonhoeffer P, Boudjemline Y, Saliba Z, Merckx J, Aggoun Y, Bonnet D, Acar P, Le Bidois J, Sidi D, Kachaner J. Percutaneous replacement of pulmonary valve in a right-ventricle to pulmonary-artery prosthetic conduit with valve dysfunction. Lancet. 2000;356:1403–5.
38. Faza N, Kenny D, Kavinsky C, Amin Z, Heitschmidt M, Hijazi ZM. Single-center comparative outcomes of the Edwards SAPIEN and Medtronic Melody transcatheter heart valves in the pulmonary position. Catheter Cardiovasc Interv. 2013;82:E535–41.
39. Chowdhury SM, Hijazi ZM, Rhodes J, Kar S, Makkar R, Mullen M, Cao Q-L, King L, Akin J, Shirali G. Early echocardiographic changes after percutaneous implantation of the Edwards SAPIEN transcatheter heart valve in the pulmonary position. Echocardiography. 2013;30:786–93.
40. Hasan BS, McElhinney DB, Brown DW. Short-term performance of the transcatheter Melody valve in high-pressure hemodynamic environments in the pulmonary and systemic circulations. Circ Cardiovasc Interv. 2011;4:615–20.
41. Harrild DM, Marcus E, Hasan B. Impact of transcatheter pulmonary valve replacement on biventricular strain and synchrony assessed by cardiac magnetic resonance feature tracking. Circ Cardiovasc Interv. 2013;6:680–7.
42. Plymen CM, Bolger AP, Lurz P, Nordmeyer J. Electrical remodeling following percutaneous pulmonary valve implantation. Am J Cardiol. 2011;107:309–14.
43. Ferraz Cavalcanti PE, Sá MP, Santos CA, Esmeraldo IM, de Escobar RR, de Menezes AM, de Azevedo Jr OM, de Vasconcelos Silva FP, Lins RF, Lima Rde C. Pulmonary valve replacement after operative repair of tetralogy of fallot: meta-analysis and meta-regression of 3,118 patients from 48 studies. J Am Coll Cardiol. 2013;62:2227–43.
44. Furushima H, Chinushi M, Sugiura H, Komura S, Tanabe Y, Watanabe H, Washizuka T, Aizawa Y. Ventricular tachycardia late after repair of congenital heart disease: efficacy of combination therapy with radiofrequency catheter ablation and class III antiarrhythmic agents and long-term outcome. J Electrocardiol. 2006;39:219–24.
45. Morwood JG, Triedman JK, Berul CI, Khairy P, Alexander ME, Cecchin F, Walsh EP. Radiofrequency catheter ablation of ventricular tachycardia in children and young adults with congenital heart disease. Heart Rhythm. 2004;1:301–8.

46. Le Gloan L, Khairy P. Management of arrhythmias in patients with tetralogy of Fallot. Curr Opin Cardiol. 2011;26:60–5.
47. Mahle WT, Parks WJ, Fyfe DA, Sallee D. Tricuspid regurgitation in patients with repaired tetralogy of fallot and its relation to right ventricular dilatation. Am J Cardiol. 2003;92:643–5.
48. Kobayashi J, Kawashima Y, Matsuda H, Nakano S, Miura T, Tokuan Y, Arisawa J. Prevalence and risk factors of tricuspid regurgitation after correction of tetralogy of Fallot. J Thorac Cardiovasc Surg. 1991;102:611–6.
49. Kogon B, Patel M, Leong T, McConnell M, Book W. Management of moderate functional tricuspid valve regurgitation at the time of pulmonary valve replacement: is concomitant tricuspid valve repair necessary? Pediatr Cardiol. 2010;31:843–8.
50. Tang GHL, David TE, Singh SK, Maganti MD, Armstrong S, Borger MA. Tricuspid valve repair with an annuloplasty ring results in improved long-term outcomes. Circulation. 2006;114:I577–81.