40 Pulmonary Valve Replacement

Varun Aggarwal, Henri Justino, Charles D. Fraser Jr.

Longstanding RVOT stenosis or regurgitation are common in patients with congenital anomalies involving the RVOT. There are 2 relatively distinct groups of patients requiring RVOT intervention. The first group consists of patients who underwent transcatheter or surgical intervention on the pulmonary valve (PV) or RVOT and now suffer from a *dysfunctional native RVOT* (i.e., without a preexisting conduit or artificial valve). The other group includes patients who received an RV-PA conduit or a bioprosthetic pulmonary valve and now suffer from a *dysfunctional conduit or bioprosthetic pulmonary valve.*

Chronic PR in the setting of repaired tetralogy of Fallot leads to RV dilation and dysfunction, which is linked to the development of exercise intolerance, arrhythmias and death. Current techniques for RVOT intervention include both transcatheter and surgical options.

Indications for Pulmonary Valve Replacement (PVR)

Criteria for PVR continue to be refined as information accrues concerning the late effects of compromised RV function. In the current era, surface echocardiography remains the primary screening tool and in most patients, it can provide semiquantitative information about RV size and function. Other important information includes level of RVOT obstruction, presence/degree of TR (and assessment of causation), and presence of intracardiac shunts. Anatomic and functional MRI has become the primary diagnostic modality for patient assessment for PVR and is possible in most patients. Pertinent information includes RV end-diastolic volume indexed for body surface area (RVEDVi), RV and LV ejection fraction, pulmonary regurgitant fraction, branch PA distortion, and assessment for intracardiac shunts. Less frequently, cardiac catheterization can be a useful adjunct to the decision-making process. Finally, in patients with pacing systems that are not MRI compatible, the decision to proceed with PVR may be facilitated by cardiac CTA.

The specific indications for PVR in patients with a dysfunctional conduit or bioprosthetic pulmonary valve are still debated. In general, PVR is indicated in the following circumstances (Tretter et al. 2016):

- Presence of symptoms from a dysfunctional RVOT
- For patients with predominant PI: an RVEDVi >150 ml/m2, ± pulmonary regurgitation fraction > 40%, or an indexed RV end-systolic volume (RVESVi) >80 ml/m2
- For patients with predominant PS: RVOT stenosis with mean Doppler gradient >35 mmHg

Additional criteria may include a dysfunctional RVOT with moderate-severe accompanying TR or the risk of long-term arrhythmia (QRS ≥180 msec) and progressive LV dysfunction.

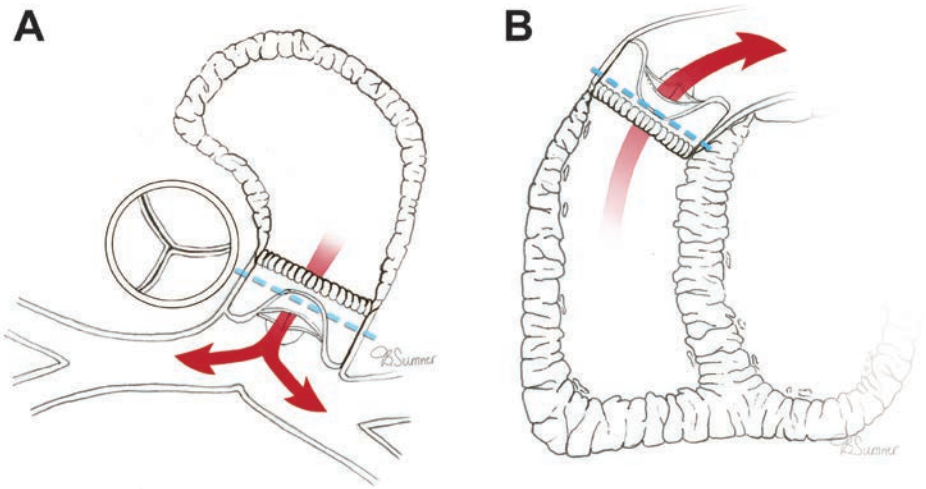

Figure 40-1. Axial and sagittal views illustrating the correct orientation of a surgically implanted PV in order to avoid branch PA obstruction,

Transcatheter Pulmonary Valve Replacement (TPVR)

Since the initial description of TPVR in 2000, TPVR has gained widespread acceptance as a nonsurgical alternative among patients who have dysfunctional RV-PA conduits or dysfunctional bioprosthetic valves. With the advent of larger transcatheter valves, the usage has also been extended to select patients with dysfunctional native RVOT without a preexisting conduit or bioprosthetic valve. The 2 FDA approved valves for pulmonic implantation are the Melody™ valve (Medtronic, Minneapolis, Minnesota, USA) and the Sapien™ XT valve (Edwards Lifesciences Inc., Irvine, CA).

Melody™ Valve

The first US implantation of the Melody™ valve was in 2007. The FDA approved the use of the Melody™ valve under humanitarian device exemption in January 2010 and subsequent premarket approval was given in February 2015. Currently available Melody™ transcatheter pulmonary valves in the US are composed of 16 mm or 18 mm bovine jugular veins sewn within a Cheatham-Platinum stent. These valves can be deployed using 18 mm, 20 mm, or 22 mm delivery systems. It is important to note that the diameter of the Melody™ valve therefore represents an *internal diameter* of 18, 20, or 22 mm.

In a recent meta-analysis (Virk et al. 2015), the overall periprocedural mortality was 1.4% and procedural complications included conduit rupture (2.6%), valve embolization (2.4%), coronary artery compression (1.2%), and PA obstruction (1.2%). Conversion to surgery was reported in 2.8% of patients. The incidence of stent fracture and infective endocarditis were 12.4% and 4.9%, respectively. The long-term outcome of the US Melody™ investigational device exemption trial (Cheatham et al. 2015) demonstrated 5-year freedom from reintervention and explantation of 76±4% and 92±3%, respectively. The main cause of valve dysfunction was stent fracture. It is noteworthy that the rate of

Melody™ stent fracture decreased by 65% when conduits were treated with placement of bare metal stents prior to implantation of a Melody™ valve, a practice known as "prestenting", which has become standard of care when Melody™ valves are placed within conduits.

Sapien™ Valve

Initially introduced for aortic valve replacement, the first use of the Sapien™ valve in the pulmonary position was reported in 2006. The COMPASSION trial (Kenny et al. 2011) demonstrated the successful deployment of the Sapien™ valve in 34 attempts in 33 patients. Valve migration was noted in 3 patients. Freedom from reintervention was 97%, with 1 patient undergoing elective placement of a second valve due to distortion of the initial implant.

Initially, the Sapien™ valve was available in the US in 23 mm and 26 mm sizes for use in RVOT ranges of 18-25 mm. In March 2016, the FDA approved the use of the Sapien™ XT valve (available in 23, 26, and 29 mm diameters) for the pulmonary position in patients with dysfunctional RV-PA conduits. The Sapien-3™ valve is available in 20, 23, 26, and 29 mm diameters, and represents a later iteration of the Sapien™ family. Although not FDA approved for pulmonic implantation, it is commonly being used off label in the pulmonary position. Of note, the Sapien™ valves are labeled by their *outer diameters* rather than by their internal diameters. Because they are available in larger diameters than the Melody™ valve, they are also commonly used off label in the native RVOT in patients who have received patch augmentation of the RVOT as part of tetralogy of Fallot repair.

Preprocedural Evaluation

TTE (2D, color, and spectral Doppler) is the first line imaging modality for all patients. Assessment of biventricular function, degree of RVOT/conduit stenosis and/or insufficiency, and evaluation of branch PAs is important. While Doppler-derived mean RVOT pressure gradients correlate reasonably well with catheter-derived gradients, echocardiography is often less accurate in estimating the severity of PR and RV size and function relative to cardiac MRI. Thus, in patients with predominant PR, cardiac MRI is generally an important component of the preprocedural evaluation.

Intraprocedural Evaluation

A complete right-heart catheterization is performed, carefully documenting the degree and levels of RVOT obstruction. A retrograde left-heart catheterization is generally also performed. Angiographic measurements of the conduit or bioprosthetic valve should be performed in several planes (ideally in a biplane catheterization laboratory). In general, degenerated conduits that are significantly shrunken down compared to their original (nominal) implanted diameters require successive balloon angioplasties to restore the conduits to an adequate diameter for TPVR. Balloon angioplasties are generally performed with noncompliant balloons, in 2 mm increments, with angiography after each angioplasty to look for signs of conduit disruption. Small pseudoaneurysms commonly develop as a consequence of conduit angioplasty. Ideally, conduits should be restored to a diameter suitable for the patient's body size and for TPVR; this implies generally a minimal diameter of 18 mm to implant a Melody™ valve. Of note, even relatively

small conduits with nominal diameters <16 mm can frequently be overexpanded to enable TPVR.

Coronary angiography should be performed at baseline, and again after successive conduit angioplasty prior to TPVR. With the balloon inflated across the conduit using dilute contrast, coronary angiography is then performed to determine if there is coronary compression during balloon expansion of the conduit. Coronary compression is an absolute contraindication to conduit stenting and TPVR. Distally, aortic root angiography should be performed during test dilation of the conduit, to determine if there is significant distortion of the aortic root, as this would also preclude conduit stenting or TPVR. In the absence of coronary or aortic root compression, prestenting of the conduit is then performed with 1-3 heavy stainless steel stents to prevent conduit recoil, followed by TPVR. Prestenting is not necessary for the Sapien™ valves because their stent is much less prone to fractures. Placement of a polytetrafluoroethylene (PTFE)-covered stent may be necessary to seal any substantial pseudoaneurysms or frank extravasations of contrast prior to prestenting and TPVR.

TPVR within bioprosthetic valves is similar to TPVR within conduits, with the exception that the frame of the bioprosthetic valve provides sufficient resistance from recoil, such that prestenting is often unnecessary, and pseudoaneurysms are much less common.

Off-label use of the current transcatheter valves has also been reported successful in native dysfunctional RVOT. In this case, a very compliant sizing balloon is used to determine if a suitable landing zone is present in the native RVOT. Generally, a suitable landing zone should be several millimeters smaller than the intended valve to be deployed. Similar precautions to look for signs of aortic root compression and coronary compression are necessary prior to TPVR in the native RVOT.

Postprocedural Care

Patients are observed overnight in the hospital for any arrhythmia and for hemodynamic monitoring. They undergo a 2-view CXR and echocardiogram on the following day to evaluate for valve position, any residual stenosis or insufficiency, perivalvar leaks, or pericardial effusion. The vascular access site is monitored for any bleeding or hematoma, given the large bore delivery sheaths needed for these procedures. Patients are started on aspirin 81 mg daily to be taken indefinitely, and maintenance of good oral hygiene and bacterial endocarditis prophylaxis cautions are required.

Surgical PVR

As per above, all patients with abnormal RV to PA connections face the ultimate need for surgical PVR. TPVR has become a very attractive and relatively safe option to *delay* the ultimate need for surgical PVR. Unfortunately, neither methodology confers lifelong security from need for reintervention, whether surgical or transcatheter. For a given patient, the goal should be to coordinate current technological alternatives in a careful strategic roadmap to optimize RV function and minimize morbity.

Preoperative Evaluation

Many patients undergoing surgical PVR have undergone previous cardiac surgery. They may have extra-anatomic RV-to-PA connections with calcified conduits, indwelling stent material, and other complex associations. The cardiac structures including aorta, PA, conduits, high-pressure cardiac chambers, and patches may be densely adherent to the sternum and/or other aspects of the chest wall. In the present era, even in the setting of multiple previous cardiac surgeries, safe sternal reentry should be expected and careful planning is paramount. CTA or MRI are very useful in determining the relationship of cardiac structures to the sternum. Options for peripheral cannulation for CPB should be carefully evaluated and the patient prepped accordingly. The OR team, including the perfusion team members, must be prepared to various contingency solutions for safe conduct of the operation. Some surgeons advocate for the routine use of femoral cannulation before sternal reentry in complex situations. We have found that with careful planning, this is not necessary in most cases.

Surgical Procedure

Sternal reentry is facilitated with the use of an oscillating saw that is used to divide the outer table and cortex, but *largely leave intact the inner table.* Using gentle upward traction and under direct vision, the adjacent cardiac structure are carefully dissected away from the undersurface of the sternum and the inner table progressively divided from a caudal to cranial direction. Once sternal reentry is gained, meticulous dissection facilitates the operation and optimizes hemostasis.

There are presently numerous options for surgical placement of a competent PV with or without a conduit. There are no data available to definitively demonstrate superiority of one over another. In our experience, in *small children and early adolescents* where significant somatic growth is still anticipated, human cadaveric cryopreserved valved conduits (pulmonary homograft valved conduits) or bovine jugular vein valved conduits (Contegra® conduits) offer attractive options. However, in our experience, bovine jugular vein conduits have a higher, and possibly prohibitive, risk of late endocarditis 7 years after implantation (Mery et al. 2016). Other options include the Hancock® composite porcine valved Dacron® conduits, which have the additional attractive feature of a supporting ring at the valve annulus. Some centers have utilized surgically constructed thin-walled Gore-Tex® valved conduits, although we do not have experience with that option. In extra-anatomic situations, where the distance from the RV to the PAs may be quite long, we have occasionally used aortic homograft valved conduits or have constructed composite extensions of homografts with Gore-Tex® tube grafts. Finally, in rare circumstances, surgeons do consider placing mechanical valves in the pulmonary position. We have uniformly avoided this alternative because of the need for significant anticoagulation.

For isolated PVR in *patients large enough to receive an adult-sized valve*, we have commonly used supported heterograft valves placed in an orthotopic position. We have favored porcine heterograft valves (such as the Epic™ and Epic™ Supra stented tissue valves) as their thin leaflets close better than bovine pericardial leaflets in the setting of the lower pressures that exist in the PA compared to the aorta. For implantation of these valves, the previous patch is longitudinally incised or removed, the

valve is implanted by suturing the posterior two-thirds circumference to the RVOT at the level of the annulus, and a Dacron® patch is used to cover the anterior third of the RVOT. Attention is made to align the valve properly in order to avoid obstruction of the branch PAs (Figure 40-1). While these valves will eventually degenerate, they have the additional attractive feature of acting as a suitable "landing zone" for future transcatheter valves.

We frequently perform PVR on CPB support with a beating heart. It is therefore very important that no residual intracardiac shunts exist. A bubble-contrast TEE study is performed on all patients prior to initiating the operation. Any residual intracardiac shunts preclude beating-heart PVR. Where possible, all intracardiac shunts should be closed. The subject of tricuspid valve repair is frequently considered in the setting of RV dilation associated with PS or PI. A tricuspid repair is performed for any degree of TR greater than mild.

Suggested Readings

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