Molina Clinical Policy
Transcatheter Pulmonary Valve Replacement
Policy No. 148

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OVERVIEW

Congenital heart disease (CHD) are conditions that are present at birth that affect the heart and proximal vasculature. Some forms of CHD may affect the structure and function of the heart. CHD ranges from mild to severe and requiring treatment. CHD which includes tetralogy of Fallot, pulmonary atresia, and transposition of the great arteries, are generally treated by surgical repair at an early age. Right ventricular outflow tract (RVOT) dysfunction, a problem with how the blood flows as it leaves the heart and goes to the lungs, is one type of CHD. The repair of RVOT requires reconstruction of the RVOT and pulmonary valve by means of placing a tube or surgical conduit to allow the blood to flow correctly. However, over a period of time the conduit can become narrowed, or a specific valve can become leaky, and a second valve replacement surgery may be required. This second surgery is usually done as an open surgery. Interventions for correction of pulmonary stenosis include open heart surgery with valve replacement, balloon dilatation, or percutaneous stenting. Interventions for pulmonary regurgitation are primarily surgical: either reconstruction of the RVOT conduit or replacement of the pulmonary valve. **Transcatheter pulmonary valve implantation (TPVI)** is a non-surgical option to restore pulmonary valve function in children and adults with such forms of CHD. The procedure offers a less invasive alternative to, or an improvement on, surgical pulmonary valve implantation for patients with CHD, who would otherwise require open surgical pulmonary valve replacement or reconstruction for RVOT obstruction. This procedure may be also important for non-congenital, destructive lesions of the pulmonary artery valve.

There are ongoing post approval studies to assess long-term clinical performance of the Melody TPV and the SAPIEN XT Transcatheter Heart Valve – Pulmonic after transcatheter implantation in participants with dysfunctional RVOT conduits; however, there are no randomized controlled trials to compare the transcatheter approach to open-heart surgical technique.

Regulatory Status

Devices for TPVI were initially cleared from marketing by the FDA through the humanitarian device exemption (HDE) process or used off-label until FDA-approved through the premarket approval (PMA) process. FDA product code: NPV.

Melody. The Melody valve system consists of two components: the Melody TPV (bovine jugular valve with stent) and the Ensemble Transcatheter Valve Delivery (TVD) System (Medtronic) were originally approved under HDE (H080002) on January 25, 2010. The approval of the Melody device was amended to a PMA due to the determination by the FDA that the device represents a breakthrough technology (FDA 2015). The PMA was based, in part, on two prospective clinical studies: the Melody TPV Long-term Follow-up Post Approval Study (PAS) and the Melody TPV New Enrollment PAS. The Melody TPV and Ensemble TVD System received PMA (P140017) on January 27, 2015 for use as an adjunct to surgery in the management of pediatric and adult patients with the following clinical conditions:

- Existence of a full (circumferential) RVOT conduit that was equal to or greater than 16 mm in diameter when originally implanted, and
- Dysfunctional RVOT conduits with a clinical indication for intervention, and either:
  - Regurgitation: ≥ moderate regurgitation, or

- Stenosis: mean RVOT gradient ≥ 35 mmHg.

In February 2017, approval of the Melody system was expanded to include patients with a dysfunctional surgical bioprosthetic valve (valve-in-valve).

SAPIEN XT. The Edwards SAPIEN XT Transcatheter Heart Valve and accessories (Edwards Lifesciences) received FDA approval for pulmonary valve use through the PMA process on February 29, 2016. The Edwards Sapien XT Transcatheter Heart Valve (Pulmonic) is composed of a stainless-steel frame with bovine pericardial tissue leaflets and available in 23- and 26-mm sizes. It includes a delivery accessories system. The Edwards SAPIEN was originally approved for aortic valve use in 2011. Approval was based on clinical evidence from the Congenital Multicenter trial of Pulmonic valve regurgitation studying the SAPIEN Interventional (COMPASSION) THV trial (NCT00676689). According to the PMA approval order, this device is indicated for use in pediatric and adult patients with the following clinical conditions:

- A dysfunctional, non-compliant RVOT conduit with a clinical indication for intervention and either:
    1. Regurgitation: ≥ moderate regurgitation, and/or
    2. Stenosis: mean RVOT gradient ≥ 35 mmHg.

Harmony. The Harmony Transcatheter Pulmonary Valve (Medtronic) received breakthrough technology status in 2019 and PMA in March 2021. It is first non-surgical heart valve to treat pediatric and adult patients with a native or surgically-repaired RVOT to stop severe pulmonary valve regurgitation caused by CHD. Harmony TPV is composed of self-expanding nitinol wire struts, a knitted polyester fabric graft, and a porcine pericardial tissue valve. It includes a delivery accessories system and is indicated for use in the management of pediatric and adult patients with severe pulmonary regurgitation (i.e., severe pulmonary regurgitation as determined by echocardiography and/or pulmonary regurgitant fraction > 30% as determined by cardiac magnetic resonance imaging) who have a native or surgically-repaired right ventricular outflow tract and are clinically indicated for surgical pulmonary valve replacement.

Refer to 'Supplemental Information' section of policy for the 'Regulatory Status of TPVI Devices' (Table 1).

COVERAGE POLICY

Transcatheter pulmonary valve implantation (TPVI) using an FDA approved valve is considered medically necessary for members with congenital heart disease and current right ventricular outflow tract obstruction (RVOT) or regurgitation when **ALL** of the following criteria are met:

1. Existence of a full (circumferential) RVOT conduit that was equal to or greater than 16 mm in diameter when originally implanted; **AND**

2. Definitive diagnosis of ONE of the following:

    a. Dysfunctional RVOT conduit with a clinical indication for intervention, and **EITHER** of the following:
        - Moderate or greater pulmonic regurgitation; **OR**
        - Pulmonic stenosis with a mean RVOT gradient greater or equal to 35 mmHg.

    OR

    b. Dysfunctional non-conduit, patch-repaired RVOT

LIMITATIONS AND EXCLUSIONS

PPVI are **contraindicated and may not be authorized** if **ANY** of the following circumstances are present:

1. History of endocarditis or other active infection within 6 months of PPVI
2. RVOT size is not appropriate for stent valve delivery (size range depends on the valve system)
3. Venous occlusions that do not permit percutaneous femoral or jugular vein access
4. Vessel size and characteristics in which the placement of a 22- to 24-Fr introducer sheath would not be safe
5. Morphology of the RVOT does not permit a percutaneous approach
6. Presence of coronary artery compression
7. Weight < 30 kg

8. Pregnancy

The following are considered **experimental, investigational and unproven** based on insufficient evidence:

1. Any indications other than those listed above

Exceptions (case-by-case review): There are rare patients without CHD who may have pulmonary valve dysfunction requiring replacement. Native outflow tract pulmonary valve implantation with balloon-expandable valves while considered off-label may be considered appropriate as an alternative to surgery in some cases and may be an exception. There may also be cases where the exclusion criteria are not met but a multidisciplinary cardiology team may determine that TPVI is favorable in exceptional scenarios.

These exceptional cases require the following as deemed appropriate by the Molina clinical reviewer: a peer-to-peer with a Molina Medical Director, consult with multidisciplinary cardiology team, and relevant clinical documentation(s) or supporting evidence to assess whether a TPVI is favorable in an individual member's case.

DOCUMENTATION REQUIREMENTS. Molina Healthcare reserves the right to require that additional documentation be made available as part of its coverage determination; quality improvement; and fraud; waste and abuse prevention processes. Documentation required may include, but is not limited to, patient records, test results and credentials of the provider ordering or performing a drug or service. Molina Healthcare may deny reimbursement or take additional appropriate action if the documentation provided does not support the initial determination that the drugs or services were medically necessary, not investigational or experimental, and otherwise within the scope of benefits afforded to the member, and/or the documentation demonstrates a pattern of billing or other practice that is inappropriate or excessive.

SUMMARY OF MEDICAL EVIDENCE

The published literature on TPVI consists of the registration trials for FDA approval, small case series, and cohort studies. The Melody valve is the longest studied TPV with the largest body of clinical evidence.

Melody TPV and the Ensemble® Transcatheter Valve Delivery System are used together for percutaneous replacement of a dysfunctional pulmonary valve. The Melody valve consists of a section of bovine jugular vein with an intact native venous valve. The valve and surrounding tissue are sutured within a platinum-iridium stent scaffolding. The transcatheter delivery system consists of a balloon-in-balloon catheter with a retractable sheath and distal cup into which the valve is placed. The procedure is performed on a beating heart without the use of cardiopulmonary bypass. The Melody valve is first crimped to fit into the delivery system. It is introduced through the femoral vein and advanced into the right side of the heart and put into place at the site of the pulmonary valve. The inner balloon is inflated to open the artificial valve, and then the outer balloon is inflated to position the valve into place.

The multicenter U.S. Melody TPV trial was a prospective uncontrolled trial designed to assess the safety, procedural success, and short-term effectiveness of the Melody TPV (McElhinney et al. 2010; Zahn et al. 2009). The Summary of Safety and Probable Benefit (SSPB) to support the approval of a humanitarian device exemption to market the Melody TPV was based on clinical data from 99 subjects who were catheterized for potential implantation with the TPV from January 2007, through December 2008, with expected follow-up and adverse event data on these subjects current through March 2009 (FDA Summary of Safety and Effectiveness Data: Melody TPV, 2015). Approved indications included RVOT dysfunction, defined as pulmonic regurgitation (moderate or greater) or pulmonic stenosis (mean gradient, ≥35 mm Hg). Also, a circumferential RVOT conduit should exist that is 16 mm or greater in diameter when originally implanted.

US Investigational Device Exemption (IDE) Study

The multicenter US Melody TPV trial is a prospective uncontrolled trial from 5 clinical sites that was designed to study the safety, procedural success, and short-term effectiveness of the Melody transcatheter pulmonary valve (McElhinney et al. 2010; Zahn et al. 2009). Beginning in January 2007, the Melody TPV was implanted in 150 patients at 5 US centers under an IDE protocol for treatment of RVOT dysfunction. In January 2010, enrollment in the US Melody Valve IDE trial was completed, and the Melody valve was approved for placement in dysfunctional RVOT conduits as a palliative measure aimed at delaying surgical intervention (McElhinney, et al., 2011).

Zahn et al. reported data on 30 patients with a success rate of the procedure in 29 Melody valve implantations. Early results of the Melody U.S. Clinical Trial were published by Zahn et al. (2009). This trial was designed to evaluate the safety, procedural success, and short-term effectiveness of the Melody transcatheter pulmonary valve in patients with dysfunctional right ventricular outflow tract conduits. 34 patients underwent catheterization for intended Melody valve implantation at three centers between January and September 2007. Implantation was successful in 29 of 30 attempts, and not attempted in four patients. The mean age was 19.4 ± 7.7 years. Doppler mean gradient was 28.8 ± 10.1 mm Hg, and 94% of patients had moderate or severe pulmonary regurgitation. Complications included one conduit rupture requiring urgent surgery and device removal, one distal pulmonary artery guidewire perforation, and one instance of wide complex tachycardia. Peak systolic conduit gradient fell from 37.2 ± 16.3 mm Hg to 17.3 ± 7.3 mm Hg. None of the patients had more than mild PR. At 6-months, conduit Doppler mean gradient was 22.4± 8.1 mm Hg, and pulmonary regurgitation fraction as measured by magnetic resonance imaging was significantly improved (3.3 ± 3.6% vs. 27.6 ± 13.3%, p<0.0001). Stent fracture occurred in 8 of 29 implants. Three of these patients were subsequently treated with a second Melody valve for recurrent stenosis during follow-up. The authors concluded that implantation of the Melody valve for RVOT conduit dysfunction can be performed by experiences operators and appears safe and has encouraging acute and short-term outcomes. During the study follow-up, 100% of the patients were free of new procedures, and 79% of the 24 patients with NYHA functional class ≥ II showed functional class improvement. This study was continued and culminated in another study that resulted in the approval of the Melody prosthesis by the FDA under the aforementioned HDE provision. The study investigators concluded that a longer duration for follow-up and a larger patient experience are required to determine the definitive role of this therapy in the treatment of conduit dysfunction (Zahn et al. 2009).

McElhinney et al. (2010) conducted a multicenter trial of 136 patients (median age, 19 years) who underwent catheterization for intended Melody valve implantation. Implantation was attempted in 124 patients. In the other 12, TPV placement was not attempted because of the risk of coronary artery compression (n=6) or other clinical or protocol contraindications. There was 1 death and 1 explanted valve after conduit rupture. The median peak RVOT gradient was 37 mmHg before implantation and 12 mmHg immediately after implantation. Before implantation, pulmonary regurgitation was moderate or severe in 92 patients. No patient had more than mild pulmonary regurgitation early after implantation or during follow-up. Freedom from stent fracture was 77.8+/-4.3% at 14 months. Freedom from valve dysfunction or reintervention was 93.5+/-2.4% at 1 year. A higher RVOT gradient at discharge and younger age were associated with shorter freedom from dysfunction. The results demonstrated an ongoing high rate of procedural success and encouraging short-term valve function. All re-interventions in this series were for RVOT obstruction, highlighting the importance of patient selection, adequate relief of obstruction, and measures to prevent and manage stent fracture (NCT00740870).

McElhinney et al. (2011) reported patient-related and procedural risk factors in the US Melody Valve Trial. From January 2007 to January 2010, 150 patients (median age, 19 years) underwent TPV implantation in the Melody valve Investigational Device Exemption trial. Existing conduit stents from a prior catheterization were present in 37 patients (25%, fractured in 12); 1 or more new pre-stents were placed at the TPV implant catheterization in 51 patients. During follow-up (median, 30 months), Melody stent fracture (MSF) was diagnosed in 39 patients. Freedom from a diagnosis of MSF was 77+/-4% at 14 months (after the 1-year evaluation window) and 60+/-9% at 39 months (3-year window). On multivariable analysis, implant within an existing stent, new pre-stent, or bioprosthetic valve (combined variable) was associated with longer freedom from MSF (P<0.001), whereas TPV compression (P=0.01) and apposition to the anterior chest wall (P=0.02) were associated with shorter freedom from MSF. Freedom from RVOT reintervention was 86+/-4% at 27 months. Among patients with a MSF, freedom from RVOT reintervention after MSF diagnosis was 49+/-10% at 2 years. Factors associated with reintervention were similar to those for MSF. The authors concluded that MSF was common after TPV implant in this multicenter experience and was more likely in patients with severely obstructed RVOT conduits and when the TPV was directly behind the anterior chest wall and/or clearly compressed. A TPV implant site protected by a pre-stent or bioprosthetic valve was associated with lower risk of MSF and reintervention.

The trial was initially designed to follow patients for 5 years after implantation or until ex-plantation but was modified in 2011 to allow follow-up out to 10 years in patients who provided supplemental written informed consent (Cheatham, et al., 2015).

Cheatham et al (2015) reported on outcomes up to 7 years following TPVI for the 148 patients who received and were discharged with a TPV in the U.S. Melody TPV trial (of 171 patients enrolled). Of the 171 patients enrolled, 167 underwent catheterization, 150 had a Melody valve implanted, and 148 of those survived to discharge with the Melody valve in place. On echocardiogram at discharge, pulmonary regurgitation was absent/trivial or mild in 140 patients and 5 patients, respectively, which represented a significant improvement from baseline. Over a median follow-up of 4.5 years (range, 0.4-7.0 years), 4 deaths occurred. During the follow-up period, 32 patients required a reintervention on RVOT, 25 of which were TPV reinterventions. A total of 11 patients required Melody valve explantation. Among the 113 patients who were alive and free from reintervention at a median of 4.5 years post-implantation, the most recent RVOT gradient was unchanged from early after valve implantation. Functional outcomes generally improved during the study: before TPVI, 14% of patients were in NYHA class I and 17% were in class III or IV. At every post-implantation annual evaluation, at least 74% of patients were in class I and no more than 1% to 2% were in class III or IV.

Melody TPV Post Approval Study (PAS)
Armstrong et al. (2014) published 1-year follow-up results of the Melody TPVI PAS, a prospective, non-randomized study designed to evaluate the short-term hemodynamic changes following device implantation. The study used historical controls from the Melody IDE trial to investigate whether the short-term effectiveness of the device was noninferior to results shown in the IDE trial. The study enrolled 120 subjects, 101 of whom underwent attempted TPVI. Patient selection was based on the criteria used in the IDE trial but did not include the age (≥5 years of age) and weight (≥30 kg) limitations. Procedure-related significant AEs occurred in 16 patients (13.3% of total cohort of 120; 15.8% of those who had an attempted TPVI), the most common of which was a confined conduit tear. Procedural success occurred in 99 subjects (98% of those with an attempted TPVI). At 1-year follow-up, the proportion of patients in NYHA class I heart failure increased from 35% at baseline to 89%. Of the 99 patients implanted for at least 24 hours, 87 had acceptable TPV hemodynamic function confirmed at 6 months (96.7% of those with evaluable echocardiographic data, 87.9% of entire cohort) and 82 had acceptable TPV hemodynamic function at 1 year (94.3% of those with evaluable echocardiographic data, 82.8% of the entire cohort). Following the procedural period, serious device-related AEs occurred in 8%, most commonly endocarditis (n=3 patients).

Gillespie et al. (2015) evaluated results of TPVI after a Ross procedure in a retrospective review of pooled findings from the Melody TPV trial and post-approval study and an additional European registry, the manufacturer-sponsored Melody TPV Post-Market Surveillance Study which was conducted in Canada and Europe (NCT00688571). In the pooled sample (N=358), 67 (19%) had a prior Ross procedure. A Melody valve was successfully implanted in 56 of 67 (84%) of the Ross patients who underwent catheterization with intent for TPVI. Six patients (9%) had symptomatic coronary artery compression after TPVI or did not undergo implantation due to the risk of compression. RV hemodynamics generally improved after TPVI, but RVOT reinterventions were required in 12 of 55 patients who were discharged from the implant hospitalization with the Melody valve in place.

Additional Studies
Eiken et al. (2011) published results of 102 consecutive PPVI performed at two centers in Germany between 2006 and 2010. The median patient age was 21.5 years. Sixty-one patients had undergone surgical correction of a Tetralogy of Fallot/pulmonary atresia with ventricular septal defect, and14 had a common arterial trunk; the remaining patients had been treated surgically for transposition of the great arteries (n=9) or aortic stenosis (n=8) or had a variety of other cardiac lesions (n=10). The majority of conduits (79) used during previous surgery were homografts. The median peak systolic RVOT gradient between the right ventricle and the pulmonary artery decreased immediately following the procedure from 37 mmHg (29–46 mmHg) to 14 mmHg (9–17 mmHg, p< 0.001). Pulmonary regurgitation assessed by MRI was reduced from a median of 16% (5–26%) to 1% (0–2%, p<0.001). The median end-diastolic RV-volume index also decreased significantly (p=0.001). One patient died due to compression of the left coronary artery. At a median follow-up of 357 days (99–388 days), the mean doppler gradient in the RVOT decreased from a pre-procedure median of 36 mmHg (26–44) to a median of 15 mmHg (12–20) at the latest follow-up (p<0.0001). The authors concluded that PPVI can be performed by an experienced structural heart disease interventionalist in patients with RVOT dysfunction. However, medium and long-term follow-up needs to be assessed to document sustained benefit. It remains to be proven whether the improvements in hemodynamics persist, and the goal to reduce the number of cardiothoracic operations during the lifetime of the patient can be achieved.

There are case series with smaller numbers of patients with patient populations ranging from 7 to 64 that reported generally similar results as the larger series, with high procedural success and relatively low rates of serious complications (Khambadkone et al. 2005; Momenah et al. 2009; Nordmeyer et al. 2006; Nordmeyer et al. 2008; Vezmar M et al. 2010; Muller et al. 2014; Butera et al. 2013; Fraisse et al. 2014). Borik et al., who evaluated 51 patients who underwent TPVI with the Melody valve at a single institution, reported the longest follow-up duration of 4.5 years (mean; range: 0.9-6.9 years), freedom from any reintervention was 87% and 68% at 3 and 5 years, respectively, and freedom from surgery was 90% at 5 years. Overall, RV functional parameters did not change with longer follow-up.
Hasan et al. (2011) described the immediate and short-term results of Melody valves implanted in a high-pressure environment. Definitions of a high-pressure system were established for Melody valves implanted in the systemic (i.e., aortic or mitral position) and pulmonary (i.e., right ventricular outflow tract conduit or tricuspid valve annulus) circulations. Implants in these environments were ascertained from databases of the 5 centers that participated in the US Investigational Device Exemption trial. Thirty implants met the inclusion criteria: 23 pulmonary circulation implants (all in the pulmonary position) systemic circulation implants (5 in the native aortic position, 1 in a left ventricle-to-descending aorta conduit, and 1 in the mitral annulus). All pulmonary circulation implants were performed percutaneously in the catheterization laboratory. A hybrid approach (surgical exposure for transcatheter implant) was used for 4 of the aortic implants. There were no procedure-related deaths. Three patients died of nonprocedural- and nonvalve-related causes. At 1 year, freedom from moderate to severe regurgitation was 100%, and freedom from mild regurgitation was 90%. Freedom from moderate to severe stenosis was 86% at 1 year. The authors concluded that short-term performance of the Melody valve in high-pressure environments is encouraging, with good valve function in all patients. The Melody valve may provide a reasonable option for transcatheter therapy in pediatric patients who are poor candidates for surgical valve replacement in high-pressure systems.

Lurz et al. (2011) reported the results of early versus late functional outcome after successful PPVI. Sixty-five patients with sustained hemodynamic effects of PPVI at 1 year were included. Patients were divided into 2 subgroups based on pre-procedural predominant pulmonary stenosis (PS) (n = 35) or predominant pulmonary regurgitation (PR) (n = 30). Data from magnetic resonance imaging and cardiopulmonary exercise testing were compared at 3 time points: before PPVI, within 1 month (early) and at 12 months (late) after PPVI. There was a significant decrease in right ventricle end-diastolic volume early after PPVI in both subgroups of patients. Right ventricle ejection fraction improved early only in the PS group (51 ± 11% vs. 58 ± 11% and 51 ± 12% vs. 50 ± 11%, $p < 0.001$ for PS, $p = 0.13$ for PR). Late after intervention, there were no further changes in magnetic resonance parameters in either group (right ventricle ejection fraction, 58 ± 11% in the PS group and 52 ± 11% in the PR group, $p = 1.00$ and $p = 0.13$, respectively). In the PS group at cardiopulmonary exercise testing, there was a significant improvement in peak oxygen uptake early (24 ± 8 ml/kg/min vs. 27 ± 9 ml/kg/min, $p = 0.008$), with no further significant change late (27 ± 9 ml/kg/min, $p = 1.00$). In the PR group, no significant changes in peak oxygen uptake from early to late could be demonstrated (25 ± 8 ml/kg/min vs. 25 ± 8 ml/kg/min vs. 26 ± 9 ml/kg/min, $p = 0.48$). The authors concluded that in patients with a sustained hemodynamic result 1 year after PPVI, a prolonged phase of maintained cardiac function is observed. However, there is no evidence for further positive functional remodeling beyond the acute effects of PPVI.

Boudjemline et al (2012) evaluated the use of the Melody valve for hemodynamically significant isolated pulmonary regurgitation. Procedural and short-term outcomes data from13 patients who underwent Melody valve implantation for a large RVOT with significant pulmonary regurgitation as the primary lesion were analyzed. All procedures were successful. The mean follow-up period was 30 +/- 4 months after the procedure. There was no incidence of stent fracture, migration or embolization. Only 1 patient who underwent the jailing technique developed a significant paraprosthetic leak and is scheduled for redilatation of the Melody valve. The authors concluded that careful patient selection, balloon sizing and RVOT preparation with pre-stenting using the Russian dolls technique and/or the PA jailing technique are required to modify the RVOT for transcatheter valve implantation. Short-term follow-up showed competent valves with no stent fracture or migration and appears promising. Wider experience with long-term outcomes may be required to standardize the procedure in such a subset of patients.

Adverse Events (AEs)
Butera et al. (2013) conducted a prospective, observational, multicenter web-based registry study of PPVI with the Melody valve through the Italian Society of Pediatric Cardiology (SICP) registry. The registry study included 63 patients between October 2007 and October 2010 (median age: 24 years; range 11-65 years). Results suggest that PPVI has good procedural and mid-term success and might delay surgical intervention in more than 80% of patients. Early results of the SICP registry on transcatheter Melody pulmonary valve implantation show that the procedure is safe and

successful. However, serious complications can occur, and valve failure occurred in almost 20% of patients during follow-up. The authors concluded that longer follow-up and larger series are needed.

Amat-Santos et al. (2015) described the incidence, features, predisposing factors, and outcomes of prosthetic valve endocarditis (PVE) after transcatheter valve replacement, both aortic and PPVI, published from 2000 to 2013. Among 28 publications (n=60; 32 aortic valve, 28 pulmonary valve), aortic valve patients had a high risk, with mean logistic EuroSCORE of 30.4 ± 14.0. Patients in the aortic valve group received either the Sapien/Sapien XT or CoreValve devices, while all patients in the pulmonary valve group received the Melody device. Data on antibiotic prophylaxis were not detailed and in-hospital complications were only reported for aortic valve patients. Pulmonary valve patients were much younger than aortic valve patients (mean age 19 ± 6 versus 80 ± 7 years). In patients undergoing aortic valve replacement, PVE was located in the transcatheter valve in 71.9% of cases compared with 100% of cases in the PPVI group. The median time between valve replacement and infective endocarditis was 5 months (IQR, 2 to 9 months). A higher incidence of enterococci was reported in the aortic group (34.4%), while Staphylococcus aureus was most reported among the pulmonary group (29.4%). Approximately 60% of aortic valve patients with PVE were managed medically, but with valve ex-plantation rates from 23% to 57% and in-hospital mortality of 34.4%. Seventy-five percent of pulmonary valve patients were managed surgically, with in-hospital mortality of 7.1%.

Virk et al. (2015) reported clinical results from 12 observational studies (n=677) for periprocedural mortality (death within 30 days of PPVI), complications, and freedom from RVOT reintervention. A DerSimonian-Laird random-effects model was used for analysis. There were 9 studies of the Melody device, 2 of the Sapien device, and 1 of both devices. Pooled periprocedural mortality was 1.4% (95% CI, 0.7% to 2.8%), while complication rates were relatively low and included: coronary artery compression (1.2%; 95% CI, 0.6 to 2.5), pulmonary artery obstruction (1.2%; 95% CI, 0.5% to 2.6%), valve embolization (2.4%; 95% CI, 1.3% to 4.3%), and conduit rupture (2.6%; 95% CI, 1.5% to 4.3%). Conversion to open surgery occurred in 2.8% of patients (95% CI, 1.7% to 4.6%). Incidence at latest follow-up was reported for stent fracture at 12.4% (95% CI, 7.6% to 19.6%) and infective endocarditis at 4.9% (95% CI, 3.2% to 7.6%). Freedom from RVOT reintervention ranged from100% at 4 months follow-up to 70% at 70 months follow-up.

Edwards Sapien XT Transcatheter Heart Valve (Pulmonic)
COngenital Multicenter trial of Pulmonic vAlve regurgitation Studying the SAPIEN InterventIONal THV (COMPASSION). The study is a prospective, non-randomized, multicenter center study assessed the safety and effectiveness of pulmonic implantation of the SAPIEN THV in patients with dysfunctional RVOT conduit requiring treatment for moderate or severe pulmonary regurgitation by TTE and/or RVOT conduit obstruction with a mean gradient of 35 mmHg or higher by TTE. Patients were treated between April 2008 and November 2014. The database supplement reflects data collected through March 2015 and includes 81 patients.

The actual completion date is December 31, 2019. Data from this clinical study were the basis for the PMA decision for the pulmonary valve implantation indication. Last Update Posted: February 26, 2020: (Clinicaltrials.gov number NCT00676689)

Kenny et al. (2018) reported 3-year outcomes of COMPASSION (Congenital Multicenter Trial of Pulmonic Valve Regurgitation Studying the SAPIEN Transcatheter Heart Valve) study. Patients with moderate to severe pulmonary regurgitation and/or RVOT conduit obstruction were implanted with the SAPIEN transcatheter heart valve. Fifty-seven of the 63 eligible patients were accounted for at the 3-year follow-up visit from a total of 69 implantations in 81 enrolled patients. Indications for implantation were pulmonary stenosis (7.6%), regurgitation (12.7%) or both (79.7%). Functional improvement in NYHA functional class was observed in 93.5% of patients. Mean peak conduit gradient decreased from 37.5 ± 25.4 to 17.8 ± 12.4 mmHg, and mean right ventricular systolic pressure decreased from 59.6 ± 17.7 to 42.9 ± 13.4 mmHg. Pulmonary regurgitation was mild or less in 91.1% of patients. When implanted in patients with moderate to severe pulmonary regurgitation and/or RVOT conduit obstruction, the SAPIEN valve was associated with favorable outcomes at 3 years, with low rates of all-cause mortality, reintervention and endocarditis and no stent fractures. Freedom from all-cause mortality at 3 years was 98.4%. Freedom from reintervention was 93.7% and from endocarditis was 97.1% at 3 years. There were no observed stent fractures. The authors concluded that TPV replacement using the Edwards SAPIEN THV demonstrates excellent valve function and clinical outcomes at 3-year follow-up.

Harmony Transcatheter Heart Valve
The FDA assessed the safety and effectiveness of the Harmony TPV device through a prospective, non-randomized, multi-center clinical study. During the study, physicians implanted the Harmony TPV in a total of 70 patients (FDA Summary of Safety and Effectiveness, 2021). The trial included 70 patients with severe pulmonary regurgitation and a clinical indication for surgical placement of a pulmonary artery conduit or prosthetic valve. Of the 70 patients, 20 were in the feasibility phase, 31 in the pivotal phase with the current TPV 22 and TPV 25 devices, and 19 were in the pivotal cohort with an earlier version (cTPV 25). Technical success was achieved in 95.7% of implantations, and the clinical endpoint of acceptable hemodynamic function without reintervention at 6 months was met in 89.2% of patients. The proportion of patients with severe pulmonary regurgitation decreased from 84.4% at baseline to 1.7% at 6 mo. 4 out of 70 patients (5.7%) required explant of the TPV; 2 were in the feasibility phase and 2 were with a prior version of the device. There were no explants with the current devices in the pivotal study and no mortalities up to the 6-month follow-up. Quality of life, measured by the 36-item short form survey, was improved most in the areas of physical functioning and role limitations due to physical health. All patients were scheduled for follow-up examinations at the start of the study, at implant procedure, discharge, and post implant at 1-month, 6-months, and annually through five years. The follow-up has been extended to 10 years as part of the post-approval study. The primary safety endpoint was no procedure- or device-related death within 30 days following the implant, which 100% of patients attained. The primary effectiveness endpoint was percentage of patients with no additional surgical or interventional procedures related to the device and acceptable heart blood flow function at 6 months. Among patients with evaluable echocardiography data, 89.2% of them achieved the primary effectiveness endpoint. Adverse events observed during the clinical study included irregular or abnormal heart rhythms (23.9%, including 14.1% ventricular tachycardia), leakage around the valve (8.5%, including 1.4% major leakage), minor bleeding (7.0%), narrowing of the pulmonary valve (4.2%), and movement of the implant (4.2%).

Systematic Review of Transcatheter Versus Surgical Pulmonary Vein Replacement
Ribeiro et al. (2020) performed a systematic review and meta-analysis of 18 nonrandomized comparative studies of SPVI and TPVI. No RCTs were identified. There were no significant differences in age or gender between the groups, but there were significant differences in anatomic and functional characteristics. Patients undergoing TPVR were more likely to have pulmonary stenosis (29% vs 12%), while those undergoing SPVR were more likely to have pulmonary regurgitation (57% vs 22%). There were large numerical differences in the presence of a native ventricle outflow tract/transannular patch (TPVR: 16%, SPVR: 60%), but this difference did not achieve statistical significance. Meta-analysis suggested a reduction in peri-procedural complications 16.5% vs 41.3%, p=.01) and length of hospital stay (-4.32 days) with the percutaneous approach, with an increased risk of infective endocarditis (5.8% vs 2.7%). There were no significant differences in early mortality, late mortality and need for reintervention. Interpretation is limited by the differences in baseline characteristics between the 2 groups and the possibility of selection bias. The authors noted that a number of patients underwent SPVR because they were not candidates for TPVR due to RVOT anatomy and/or other cardiac defects.

Professional Society Guidelines

American College of Cardiology (ACC)/American Heart Association (AHA)/et al.
The ACC and AHA and 6 other societies published comprehensive guidelines on the management of patients with CHD in 2018 (Stout et al. 2018). Recommendations for treatment included pulmonary stenosis, pulmonary regurgitation and tetralogy of Fallot.

ACC/AHA Guidelines on the Management of Patients with Tetralogy of Fallot		
Recommendation	**Strength of Recommendation**	**Level of Evidence**
Pulmonary valve replacement (surgical or percutaneous) for relief of symptoms is recommended for patients with repaired tetralogy of Fallot and moderate or greater pulmonary regurgitation with cardiovascular symptoms not otherwise explained.	Strong	Non-randomized (moderate quality evidence)
Pulmonary valve replacement (surgical or percutaneous) is reasonable for preservation of ventricular size and function in asymptomatic patients with repaired tetralogy of Fallot and ventricular enlargement or dysfunction and moderate or greater pulmonary regurgitation.	Moderate	Non-randomized (moderate quality evidence)

Surgical pulmonary valve replacement may be reasonable for adults with repaired tetralogy of Fallot and moderate or greater pulmonary regurgitation with other lesions requiring surgical interventions.	Weak	Consensus of expert opinion
Pulmonary valve replacement, in addition to arrhythmia management, may be considered for adults with repaired tetralogy of Fallot and moderate or greater pulmonary regurgitation and ventricular tachyarrhythmia.	Weak	Consensus of expert opinion

National Institute for Health and Care Excellence (NICE) published a guidance, *Percutaneous pulmonary valve implantation for right ventricular outflow tract dysfunction (2013)*, which indicates that the evidence on PPVI for RVOT dysfunction shows good short-term efficacy. There is little evidence on long-term efficacy, but it is well documented that these valves may need to be replaced in the longer term. With regard to safety there are well-recognized complications, particularly stent fractures in the longer term, which may or may not have clinical effects. Patients having this procedure are often in poor health and might otherwise need open heart surgery (typically re-operative) with its associated risks. The procedure should be performed only in specialist units and with arrangements in place for cardiac surgical support in the event of complications. Patient selection should be carried out by a multidisciplinary team including a cardiologist with a special interest in congenital heart disease, an interventional cardiologist and a cardiothoracic surgeon with a special interest in congenital heart disease. Additionally, this is a technically challenging procedure that should only be performed by clinicians with training and experience in interventional cardiology and congenital heart disease.

International Guidelines

The European Society of Cardiology (ESC) 2010 guidelines for the management of adult CHD endorsed by the Association for European Pediatric Cardiology (AEPC) include the following exclusion criteria for percutaneous pulmonic valve implantation (PPVI):

- Evidence of risk of coronary compression by the expanded implant as determined by balloon testing
- Central vein occlusion or significant obstruction
- Active infection (such as endocarditis) or high risk of infection (such as intravenous drug abuse)
- Surgery is preferred when additional interventions are considered such as tricuspid annuloplasty, coronary artery bypass, or arrhythmia surgery

The European Society of Cardiology (ESC)/Association for European Pediatric Cardiology (AEPC) 2010 guidelines for the management of adult CHD include the following indications for surgical intervention or PPVI in patients with right ventricular to pulmonary artery conduits:

- Intervention is recommended in symptomatic patients with RV systolic pressure >60 mmHg (TR velocity >3.5 m/s; may be lower in cases with reduced flow) and/or moderate to severe pulmonic regurgitation.
- Intervention is suggested in asymptomatic patients with severe right ventricular outflow tract obstruction and/or severe pulmonic regurgitation when at least one of the following criteria is present:
  - Decrease in exercise capacity on cardiopulmonary exercise testing
  - Progressive RV dilation
  - Progressive RV systolic dysfunction
  - Progressive TR (at least moderate)
  - RV systolic pressure >80 mmHg (TR velocity >4.3 m/s)
  - Sustained atrial/ventricular arrhythmias

SUPPLEMENTAL INFORMATION

Table 1. Regulatory Status of TPVI Devices

Device	Manufacturer	Date Approved	PMA No.	Indications
Melody® Transcatheter Pulmonary Valve (TPV)	Medtronic	Jan 2010	H080002 (HDE)	Pulmonary valve replacement for pediatric and adult patients with a dysfunctional, noncompliant RVOT conduit
Melody® TPV	Medtronic	Jan 2015	P140017	Pulmonary valve replacement for pediatric and adult patients with a dysfunctional, noncompliant RVOT conduit

Melody® TPV	Medtronic	Feb 2017	P140017/S005	Valve-in-valve for patients with a dysfunctional surgical bioprosthetic pulmonary valve
SAPIEN XT™ Transcatheter Heart Valve (pulmonic)	Edwards Lifesciences	Feb 2016	P130009/S037	Pulmonary valve replacement for pediatric and adult patients with a dysfunctional, noncompliant RVOT conduit
Harmony™ TPV	Medtronic	Mar 2021	P200046	Pulmonary valve for pediatric and adult patients with severe pulmonary regurgitation

HDE: humanitarian device exemption; PMA: premarket approval; RVOT: right ventricular outflow tract.

CODING & BILLING INFORMATION

Covered CPT Codes

CPT	Description
33477	Transcatheter pulmonary valve implantation, percutaneous approach, including pre-stenting of the valve delivery site, when performed

Covered HCPCS Codes

HCPCS	Description
	N/A

CODING DISCLAIMER. Codes listed in this policy are for reference purposes only and may not be all-inclusive. Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement. Listing of a service or device code in this policy does guarantee coverage. Coverage is determined by the benefit document. Molina adheres to Current Procedural Terminology (CPT®), a registered trademark of the American Medical Association (AMA). All CPT codes and descriptions are copyrighted by the AMA; this information is included for informational purposes only. Providers and facilities are expected to utilize industry standard coding practices for all submissions. When improper billing and coding is not followed, Molina has the right to reject/deny the claim and recover claim payment(s). Due to changing industry practices, Molina reserves the right to revise this policy as needed.

APPROVAL HISTORY

12/08/2021	Policy reviewed and revised. IRO Peer Review: IRO reviews, 11/30/2021; 12/1/2021. Practicing physician board-certified in Pediatric Cardiology; Interventional Cardiology. Updated references, clinical studies and content of policy. Notable updates include: • Added diagnosis of 'Dysfunctional non-conduit, patch-repaired RVOT' indication • Added Harmony™ TPV (Medtronic) approved in March 2021 for the indication of pulmonary valve for pediatric and adult patients with severe pulmonary regurgitation • Previous criteria were specific to Melody TPV (MedTronic) and Edwards SAPIEN™ XT Transcatheter Heart Valve; revised criteria to address TPVI devices • Revised and updated 'Summary of Medical Evidence' section • Added 'Table 1: Regulatory Status of TPVI Devices' in Supplemental Information section • Added pregnancy to exclusion criteria • Added exceptions statement for case-by-case review • Assessed Sapien S3 valve for inclusion in medical necessity as percutaneous transcatheter pulmonary valve replacement, however its long-term effectiveness has not been established. References addressing Sapien S3 valve, including two studies reported off-label PPVI with the SAPIEN S3 valve with short follow-up, added to references section.
12/9/2020	Policy reviewed. No changes to coverage criteria. Updated references
12/10/2019	Policy reviewed. Updated with inclusion of the Edwards Sapien XT Transcatheter Heart Valve FDA approval. Updated description of PPVI procedure, revised coverage criteria to include device specific criteria, updated contraindications section, added new references and clinical trial information and updated guideline information. IRO Peer Review: 10/13/2019. Practicing physician board certified in Pediatrics, Pediatric Cardiology.
7/10/2018	Policy reviewed. No changes to coverage criteria. Updated references.
9/19/2017	Policy reviewed. No changes to coverage criteria. Updated references.
11/2016	Policy reviewed. No changes to coverage criteria. Medical evidence summary and references sections updated.
12/16/2015	Policy reviewed. No changes.
10/30/2013	New policy. IRO Peer Review: 9/20/2013. Practicing physician board certified in Pediatrics, Pediatric Cardiology.

REFERENCES

Government Agency

Centers for Medicare and Medicaid Services (CMS). Medicare Coverage Database. National Coverage Determination (NCD).Available at: CMS [Search: Transcatheter pulmonary valve; pulmonary valve; Melody valve]

Center for Devices and Radiological Health (CDRH). Premarket Approval (PMA) Database [search: NPV]. Transcatheter Heart/Pulmonary Valve. Food and Drug Administration [website]. Available at: FDA. Accessed December 2021

ClinicalTrials.org.

- COMPASSION: COngenital Multicenter Trial of Pulmonic VAlve Regurgitation Studying the SAPIEN InterventIONal THV. NCT00676689. First received: May 28, 2008. Last updated Feb, 2020. Accessed at: Link

Food and Drug Administration (FDA)

- Summary of Safety and Effectiveness: Harmony Transcatheter Pulmonary Valve. 2021. https://www.accessdata.fda.gov/cdrh_docs/pdf20/P200046B.pdf.
- Summary of Safety and Effectiveness Data: Melody™ Transcatheter Pulmonary Valve, models PB1016 and PB1018; Ensemble™ Transcatheter Valve Delivery System 2017; https://www.accessdata.fda.gov/cdrh_docs/pdf14/p140017s005b.pdf.
- Medtronic Melody Transcatheter Pulmonary Valve. P140017. New Humanitarian Device Approval. 2015. Accessed at: https://www.accessdata.fda.gov/cdrh_docs/pdf14/P140017a.pdf
- Summary of Safety and Effectiveness Data: Melody™ Transcatheter Pulmonary Valve. 2015; https://www.accessdata.fda.gov/cdrh_docs/pdf14/p140017b.pdf
- Summary of Safety and Probable Benefit: Melody Transcatheter Pulmonary Valve and Ensemble Transcatheter Valve Delivery System. 2010; https://www.accessdata.fda.gov/cdrh_docs/pdf8/H080002b.pdf. A
- Premarket Approval (PMA). Edwards SAPIEN XT™ Transcatheter Heart Valve. P130009. Accessed at: https://www.accessdata.fda.gov/cdrh_docs/pdf11/p110021b.pdf

Peer Reviewed Publications

Amat-Santos IJ, Ribeiro HB, Urena M, et al. Prosthetic valve endocarditis after transcatheter valve replacement: a systematic review. JACC Cardiovasc Interv. 2015;8(2):334-346.

Armstrong AK, Balzer DT, Cabalka AK, et al. One-year follow-up of the Melody transcatheter pulmonary valve multicenter post-approval study. JACC Cardiovasc Interv. 2014;7(11):1254-1262.

Boudjemline Y, Brugada G, Van-Aerschot I et al. Outcomes and safety of transcatheter pulmonary valve replacement in patients with large patched right ventricular outflow tracts. Archives of Cardiovascular Diseases. 105 (8-9) (pp 404-413), 2012.

Cheatham JP, Hellenbrand WE, Zahn EM, et al. Clinical and hemodynamic outcomes up to 7 years after transcatheter pulmonary valve replacement in the US Melody valve investigational device exemption trial. Circulation. 2015;131(22):1960-1970.

Eiken A, Ewert P, Hager A, Peters B, Fratz S, Kuehne T, et al. Percutaneous pulmonary valve implantation: two-centre experience with more than 100 patients. Eur Heart J. 2011 May;32(10):1260-5. Epub 2011 Jan 27.

Esmaeili A, Khalil M, Behnke-Hall K, et al. Percutaneous pulmonary valve implantation (PPVI) in non-obstructive right ventricular outflow tract: limitations and mid-term outcomes. Transl Pediatr. 2019;8(2):107-113. doi:10.21037/tp.2019.04.02.

Gillespie MJ, McElhinney DB, Kreutzer J, et al. Transcatheter Pulmonary Valve Replacement for Right Ventricular Outflow Tract Conduit Dysfunction After the Ross Procedure. Ann Thorac Surg. Sep 2015; 100(3):996-1003. PMID 26190388

Hasan BS, McElhinney DB, Brown DW et al. Short-term performance of the transcatheter Melody valve in high-pressure hemodynamic environments in the pulmonary and systemic circulations. Circ Cardiovasc Interv. 2011 Dec 1;4(6):615-20. doi: 10.1161/CIRCINTERVENTIONS. 111.963389. Epub 2011 Nov 9.

Hascoet S, Dalla Pozza R, et al. Early outcomes of percutaneous pulmonary valve implantation using the Edwards SAPIEN 3 transcatheter heart valve system. EuroIntervention. 2019 Jan 20;14(13):1378-1385. doi: 10.4244/EIJ-D-18-01035. PMID: 30418156.

Kenny D, Rhodes JF et al. 3-Year Outcomes of the Edwards SAPIEN Transcatheter Heart Valve for Conduit Failure in the Pulmonary Position from the COMPASSION Multicenter Clinical Trial. JACC Cardiovasc Interv. 2018 Oct 8;11(19):1920-1929

Kreutzer J, Armstrong AK, Rome JJ, et al. Comparison of the Investigational Device Exemption and Post-Approval Trials of the Melody Transcatheter Pulmonary Valve. Catheter Cardiovasc Inter. 2021; 1-13.

Malekzadeh-Milani S, Ladouceur M, Patel M, et al. Incidence and predictors of Melody(R) valve endocarditis: a prospective study. Arch Cardiovasc Dis. Feb 2015; 108(2):97-106. PMID 25445752

McElhinney DB, Hellenbrand WE, Zahn EM, et al. Short- and medium-term outcomes after transcatheter pulmonary valve placement in the expanded multicenter US melody valve trial. Circulation. 2010 Aug 3;122(5):507-16. Epub 2010 Jul 19. PMID 20644013

McElhinney DB, Cheatham JP, Jones TK, et al. Stent fracture, valve dysfunction, and right ventricular outflow tract reintervention after transcatheter pulmonary valve implantation: patient-related and procedural risk factors in the US Melody Valve Trial. Circ Cardiovasc Interv. Dec 1 2011; 4(6):602-614. PMID 22075927

Ribeiro JM, Teixeira R, Lopes J, et al.(2020) Transcatheter Versus Surgical Pulmonary Valve Replacement: A Systemic Review and Meta-Analysis. Ann Thorac Surg. Nov 2020; 110(5): 1751-1761. PMID 32268142

Van Dijck I, Budts W, Cools B, et al. Infective endocarditis of a transcatheter pulmonary valve in comparison with surgical implants. Heart. May 15 2015; 101(10):788-793. PMID 25539944

Vezmar M, Chaturvedi R, Lee KJ, Almeida C, Manlhiot C, McCrindle BW, Horlick EM, Benson LN. Percutaneous pulmonary valve implantation in the young 2-year follow-up. JACC Cardiovasc Interv. 2010 Apr;3(4):439-48.

Virk SA, Liou K, Chandrakumar D, Gupta S, Cao C. Percutaneous pulmonary valve implantation: a systematic review of clinical outcomes. Int J Cardiol. 2015;201:487-489.

Zahn EM, Hellenbrand WE, Lock JE, McElhinney DB. Implantation of the melody transcatheter pulmonary valve in patients with a dysfunctional right ventricular outflow tract conduit early results from the U.S. Clinical trial. J Am Coll Cardiol. Oct 27, 2009;54(18):1722-1729. PMID 19850214

Case Series

- Khambadkone S, Coats L, Taylor A, et al. Percutaneous pulmonary valve implantation in humans: results in 59 consecutive patients. Circulation. 2005; 112(8):1189-1197.
- Momenah TS, El Oakley R, Al Najashi K, et al. Extended application of percutaneous pulmonary valve implantation. J Am Coll Cardiol. 2009; 53(20):1859-1863.
- Nordmeyer J, Coats L, Bonhoeffer P. Current experience with percutaneous pulmonary valve implantation. Semin Thorac Cardiovasc Surg. 2006; 18(2):122-125.
- Nordmeyer J, Coats L, Lurz P, et al. Percutaneous pulmonary valve-in-valve implantation: a successful treatment concept for early device failure. Eur Heart J. 2008; 29(6):810-815.
- Vezmar M, Chaturvedi R, Lee KJ, et al. Percutaneous pulmonary valve implantation in the young 2-year follow- up. JACC Cardiovasc Interv. 2010; 3(4):439-448.
- Muller J, Engelhardt A, Fratz S, et al. Improved exercise performance and quality of life after percutaneous pulmonary valve implantation. Int J Cardiol. May 15 2014; 173(3):388-392. PMID 24713459
- Butera G, Milanesi O, Spadoni I et al. Melody transcatheter pulmonary valve implantation. Results from the registry of the Italian Society of Pediatric Cardiology. Catheter Cardiovasc Interv. 2013 Feb;81(2):310-6. doi: 10.1002/ccd.24518. Epub 2012 Nov 14.
- Fraisse A, Aldebert P, Malekzadeh-Milani S, et al. Melody (R) transcatheter pulmonary valve implantation: results from a French registry. Arch Cardiovasc Dis. Nov 2014; 107(11):607-614. PMID 25453718
- Borik S, Crean A, Horlick E, et al. Percutaneous pulmonary valve implantation: 5 years of follow-up: does age influence outcomes? Circ Cardiovasc Interv. Feb 2015; 8(2):e001745. PMID 25652317

National and Specialty Organizations

American College of Cardiology/American Heart Association

- Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *Circulation*. 2019;139:e698–e800. doi: 10.1161/CIR.0000000000000603

National Institute for Health and Care Excellence (NICE): Percutaneous pulmonary valve implantation for right ventricular outflow tract dysfunction. IPG436. January 2013. Accessed at: http://guidance.nice.org.uk/IPG436

American College of Cardiology (ACC) Website. Philipp Lurz, MD; Johannes Nordmeyer, MD; Alessandro Giardini, MD et al. Early Versus Late Functional Outcome After Successful Percutaneous Pulmonary Valve Implantation: Are the Acute Effects of Altered Right Ventricular Loading All We Can Expect? J Am Coll Cardiol. 2011;57(6):724-731. doi:10.1016/j.jacc.2010.07.056

Nishimura RA, Otto CM, Bonow RO, et al.; ACC/AHA Task Force Members.

- 2017 AHA/ACC Focused Update of the 2014 AHA/ACC Guideline for the Management of Patients With Valvular Heart Disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines [correction appears in Circulation. 2014;130(13):e120]. Circulation. 129(23):e521-e643. Available at: http://circ.ahajournals.org/content/early/2017/03/14/CIR.0000000000000503
- Stout KK, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. Journal of the American College of Cardiology 2018; Online. DOI: 10.1016/j.jacc.2018.08.1029. (Reaffirmed 2018 Aug)

Other Peer Reviewed Publications

UpToDate: [website]. Waltham, MA: Walters Kluwer Health; 2021 [via subscription only]

- Doyle T. Kavanaugh-McHugh A. Overview of the management of tetralogy of Fallot.
- Fratz S. Percutaneous pulmonic valve implantation.

Dynamed: In DynaMed [database online]. EBSCO Information Services 2021. [via subscription only]

- Tetralogy of Fallot in infants and children.
- Tetralogy of Fallot in adults.

References (not cited in policy)

- Baumgartner H, Bonhoeffer P, De Groot NM, et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2010; 31:2915.
- Biernacka EK, Piotrowicz E, Fronczak A, et al. Influence of percutaneous pulmonary valve implantation on exercise capacity: Which group of patients benefits most from the intervention? Cardiol J. 2015a;22(3):343-350.
- Biernacka EK, Rużyłło W, Demkow M, et al. Transcatheter pulmonary valve implantation in patients with right ventricular outflow tract dysfunction: early and mid-term results. J Invasive Cardiol. 2015b;27(6):E82-E89.
- Bonhoeffer P, Boudjemline Y, Saliba Z, et al. Percutaneous replacement of pulmonary valve in a right-ventricle to pulmonary-artery prosthetic conduit with valve dysfunction. Lancet 2000;356:1403-1405.
- Cools B, Budts W, Heying R, et al. Medium-term follow-up after percutaneous pulmonary valve replacement with the Melody valve. IJC Heart & Vasculature. 2015;7:92-97.
- Fathallah M, Krasuski RA. Pulmonic valve disease: review of pathology and current treatment options. Current Cardiology Reports 2017;19(11):108. DOI: 10.1007/s11886-017-0922-2.
- Hascoet S, Dalla Pozza R et al. Early outcomes of percutaneous pulmonary valve implantation using the Edwards SAPIEN 3 transcatheter heart valve system. Euro Intervention. 2019 Jan 20;14(13):1378-1385.
- Plessis J et al. Edwards SAPIEN Transcatheter Pulmonary Valve Implantation: Results from a French Registry. JACC Cardiovasc Interv. 2018 Oct 8;11(19):1909-1916.
- Steinberg ZL, Jones TK et al. Early outcomes in patients undergoing transcatheter versus surgical pulmonary valve replacement. Heart 2017;103(18):1455-1460. DOI: 10.1136/heartjnl-2016-310776.

Treatment of Non-Conduit, Patch-Repaired RVOT

- Boshoff DE, Cools BL, Heying R, et al. Off-label use of percutaneous pulmonary valved stents in the right ventricular outflow tract: Time to rewrite the label? Catheter Cardiovasc Interv. 2013;81(6):987-995.
- Cools B, Brown SC, Heying R, et al. Percutaneous pulmonary valve implantation for free pulmonary regurgitation following conduit-free surgery of the right ventricular outflow tract. Int J Cardiol. 2015;186:129-135.
- Demkow M, Rużyłło W, Biernacka EK, et al. Percutaneous Edwards SAPIEN(™) valve implantation for significant pulmonary regurgitation after previous surgical repair with a right ventricular outflow patch. Catheter Cardiovasc Interv. 2014;83(3):474-481.
- Grundmann S, Hoehn R, Kroll J, Grohmann J. Percutaneous pulmonary valve implantation in the patched right ventricular outflow tract 51 years after surgical repair for tetralogy of Fallot. J Heart Valve Dis. 2016;25(5):648-650.
- Esmaeili A, Bollmann S, Khalil M, et al. Percutaneous pulmonary valve implantation for reconstruction of a patch-repaired right ventricular outflow tract. J Interv Cardiol. 2018;31(1):106-111.

Sapien 3

- Edwards SAPIEN 3 Transcatheter Heart Valve System with Edwards Commander Delivery System – P200015.Accessed at: FDA
- COMPASSION S3 - Evaluation of the SAPIEN 3 Transcatheter Heart Valve in Patients With Pulmonary Valve Dysfunction. ClinicalTrials.gov Identifier: NCT02744677. Accessed at: https://clinicaltrials.gov/ct2/show/NCT02744677
- Hascoet S, Dalla Pozza R, Bentham J, et al. Early outcomes of percutaneous pulmonary valve implantation using the Edwards SAPIEN 3 transcatheter heart valve system. EuroIntervention 2019; 14:1378.
- Sinha S, Aboulhosn J, Asnes J, et al. Initial results from the off-label use of the SAPIEN S3 valve for percutaneous transcatheter pulmonary valve replacement: A multi-institutional experience. Catheter Cardiovasc Interv 2019; 93:455.

APPENDIX

Reserved for State specific information *(to be provided by the individual States, not Corporate). Information includes, but is not limited to, State contract language, Medicaid criteria and other mandated criteria.*

Medicare National Coverage There is no national coverage determination (NCD) identified. In the absence of a national coverage determination, coverage decisions are left to the discretion of local Medicare carriers.