Introduction

Congenital heart disease is a term that means a person was born with heart problems. These difficulties affect the heart’s function and structure. Congenital heart disease can range from mild, which may not need treatment, to severe, which often does. One congenital heart defect is known as right ventricular outflow tract (RVOT) dysfunction. Essentially, it’s a problem with how the blood flows as it leaves the heart and goes to the lungs. Repairing it requires reconstructing certain areas of the heart and placing a tube (conduit) to allow the blood to flow correctly. Over a long period of time the conduit can become narrowed or a specific valve can become leaky. A second valve replacement surgery may be needed in this situation. This second surgery is usually done as an open surgery. However, surgery using a long, thin tube (a heart catheter) instead of open heart surgery can be done in certain situations. This policy describes when an additional RVOT surgery using a catheter may be considered medically necessary.

Note: The Introduction section is for your general knowledge and is not to be taken as policy coverage criteria. The rest of the policy uses specific words and concepts familiar to medical professionals. It is intended for providers. A provider can be a person, such as a doctor, nurse, psychologist, or dentist. A provider also can be a place where medical care is given, like a hospital, clinic, or lab. This policy informs them about when a service may be covered.
Transcatheter pulmonary valve implantation (TPVI)

Transcatheter pulmonary valve implantation is considered medically necessary for patients with congenital heart disease and current right ventricular outflow tract obstruction (RVOT) or regurgitation including the following indications:

- Individuals with right ventricle-to-pulmonary artery conduit with or without bioprosthetic valve with at least moderate pulmonic regurgitation
- OR
- Individuals with native or patched RVOT with at least moderate pulmonic regurgitation
- OR
- Individuals with right ventricle-to-pulmonary artery conduit with or without bioprosthetic valve with pulmonic stenosis (mean RVOT gradient at least 35 mm Hg)
- OR
- Individuals with native or patched RVOT with pulmonic stenosis (mean RVOT gradient at least 35 mm Hg).

Transcatheter pulmonary valve implantation is considered investigational for all other indications.

### Coding

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>CPT</td>
<td>Transcatheter pulmonary valve implantation, percutaneous approach, including pre-stenting of the valve delivery site, when performed</td>
</tr>
</tbody>
</table>

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Evidence Review

Description

Transcatheter pulmonary valve implantation (TPVI) is a less invasive alternative to open surgical pulmonary valve replacement or reconstruction for right ventricular outflow tract (RVOT) obstruction. Percutaneous pulmonary valve replacement may be indicated for congenital pulmonary stenosis. Pulmonary stenosis or regurgitation in a patient with congenital heart disease (CHD) who has previously undergone RVOT surgery are additional indications. Patients with prior CHD repair are at risk of needing repeated reconstruction procedures.

Background

Congenital Heart Disease

Congenital heart disease, including tetralogy of Fallot, pulmonary atresia, and transposition of the great arteries, is generally treated by surgical repair at an early age. This involves reconstruction of the right ventricular outflow tract (RVOT) and pulmonary valve using a surgical homograft or a bovine-derived valved conduit. These repairs are prone to development of pulmonary stenosis or regurgitation over long periods of follow-up.

Because individuals with surgically corrected congenital heart disease are living into adulthood, RVOT dysfunction following initial repair has become more common. Calcification of the RVOT conduit can lead to pulmonary stenosis, while aneurysmal dilatation can result in pulmonary regurgitation. RVOT dysfunction can lead to decreased exercise tolerance, potentially fatal arrhythmias, and/or irreversible right ventricular dysfunction.¹

Treatment

Interventions for RVOT dysfunction often require numerous repeat open heart procedures for patients who live into adulthood. Treatment options for pulmonary stenosis are open surgery
Interventions for pulmonary regurgitation are primarily surgical, either reconstruction of the RVOT conduit or replacement of the pulmonary valve. The optimal timing of these interventions is not well understood.

Transcatheter pulmonary valve replacement offers a less invasive treatment option for patients with prior surgery for congenital heart disease and RVOT dysfunction. It is possible that a less invasive valve replacement technique could spare patients from multiple repeat open heart procedures over long periods of follow-up.

**Summary of Evidence**

For individuals who have a history of congenital heart disease (CHD) and current right ventricular outflow tract (RVOT) obstruction who receive transcatheter pulmonary valve implantation (TPVI) with a Food and Drug Administration (FDA)-approved device and indication, the evidence includes prospective, interventional, noncomparative studies and multiple prospective and retrospective case or cohort series. Relevant outcomes are overall survival, symptoms, functional outcomes, quality of life, hospitalizations, and treatment-related morbidity and mortality. Results of the case series have indicated that there is a high rate of procedural success and low procedural mortality, although the rates of serious procedural adverse events reported ranges from 3.0% to 7.4%. Most valves have demonstrated competent functioning by Doppler echocardiography at 6- to 12-month follow-ups, but complications (eg, stent fractures, need for re-interventions) were reported in an FDA analysis to occur at rates of 18% and 7%, respectively. Other publications with longer follow-up have reported stent fractures in up to 26% of patients; however, most stent fractures did not required reintervention. Studies with follow-up extending to a maximum of 7 years postprocedure have suggested that the functional and hemodynamic improvements are durable, but a relatively high proportion of patients (20%-30%) have required reintervention on the pulmonary valve. No comparative studies were identified, and there is no direct evidence that TPVI reduces future open heart procedures. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have a history of CHD and current RVOT obstruction who receive TPVI with a non-FDA-approved device or indication, the evidence includes case series. Relevant outcomes are overall survival, symptoms, functional outcomes, quality of life, hospitalizations, and treatment-related morbidity and mortality. There is limited evidence on the off-label use of TPVI, including the use of a non-FDA-approved valve, or use of an approved valve for a non-FDA-approved indication. The published case series enrolled relatively few patients and are
heterogeneous regarding devices used and indications for TPVI. The evidence is insufficient to determine the effects of the technology on health outcomes.

**Ongoing and Unpublished Clinical Trials**

Some currently unpublished trials that might influence this review are listed in Table 1.

**Table 1. Summary of Key Trials**

<table>
<thead>
<tr>
<th>NCT No.</th>
<th>Trial Name</th>
<th>Planned Enrollment</th>
<th>Completion Date</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Ongoing</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>NCT01186692a</td>
<td>Melody® Implantation of the Medtronic Melody Transcatheter Pulmonary Valve (TPV) in Patients With Dysfunctional Right Ventricular Outflow Tract (RVOT) Conduits: A Post-Market Approval Study</td>
<td>131</td>
<td>Jul 2017 (ongoing)</td>
</tr>
<tr>
<td>NCT00676689a</td>
<td>Implantation of the SAPIEN Transcatheter Heart Valve (THV) in the Pulmonic Position</td>
<td>70</td>
<td>Nov 2019</td>
</tr>
<tr>
<td>NCT00740870a</td>
<td>Implantation of the Medtronic Melody Transcatheter Pulmonary Valve in Patients With Dysfunctional RVOT Conduits: A Feasibility Study</td>
<td>150</td>
<td>Jan 2020</td>
</tr>
<tr>
<td>NCT02744677a</td>
<td>COgenital Multicenter Trial of Pulmonic vAlve Dysfunction Studying the SAPIEN 3 interventIONal THV</td>
<td>58</td>
<td>Dec 2022</td>
</tr>
<tr>
<td>NCT02979587</td>
<td>The Medtronic Harmony™ Transcatheter Pulmonary Valve Clinical Study</td>
<td>40</td>
<td>Dec 2023</td>
</tr>
<tr>
<td>NCT02987387a</td>
<td>New Enrollment SAPIEN XT Post-Approval Study</td>
<td>191</td>
<td>Jan 2024</td>
</tr>
</tbody>
</table>

NCT: national clinical trial  
*a* Denotes industry-sponsored or cosponsored trial

**Clinical Input Received from Physician Specialty Societies and Academic Medical Centers**

While the various physician specialty societies and academic medical centers may collaborate with and make recommendations during this process, through the provision of appropriate
reviewers, input received does not represent an endorsement or position statement by the physician specialty societies or academic medical centers, unless otherwise noted.

2018 Input

In response to requests, clinical input on the use of transcatheter pulmonary valve implantation (TPVI) was received from 2 specialty society-level respondents while this policy was under review in 2018. The combined clinical input response incorporated input from a panel including physicians affiliated with academic medical centers.

Based on the evidence and independent clinical input, the clinical input supports that the following indications provide a clinically meaningful improvement in the net health outcome and are consistent with generally accepted medical practice:

- Use of TPVI for individuals with right ventricle-to-pulmonary artery conduit with or without bioprosthetic valve with at least moderate pulmonic regurgitation;
- Use of TPVI for individuals with native or patched right ventricular outflow tract (RVOT) with at least moderate pulmonic regurgitation;
- Use of TPVI for individuals with right ventricle-to-pulmonary artery conduit with or without bioprosthetic valve with pulmonic stenosis (mean RVOT gradient at least 35 mm Hg); or
- Use of TPVI for individuals with native or patched RVOT with pulmonic stenosis (mean RVOT gradient at least 35 mm Hg)

2011 Input

In response to requests, input was received from 6 academic medical centers while this policy was under review in 2011. Overall response to whether TPVI was investigational was mixed, with 2 of 5 reviewers agreeing with the investigational status, and 3 reviewers indicating partial support. Most reviewers (4/5) indicated that there is a subpopulation of patients who are at high risk for surgery or who are not candidates for surgery, and for whom there are no other available options. These reviewers felt TPVI was a viable alternative that offered potential benefit for these patients.
Practice Guidelines and Position Statements

*Society for Cardiovascular Angiography and Interventions et al*

The Society for Cardiovascular Angiography and Interventions, American Association for Thoracic Surgery, American College of Cardiology (ACC) and the Society of Thoracic Surgeons (2015) published a consensus-based report on operator and institutional requirements for TPVI. Recommendations to qualify for a TPVI program included 150 catheterizations per year, association with a surgical program, submission of all cases to a national registry, and, for patients, 80% freedom from re-intervention at 1 year.

*American Heart Association and American College of Cardiology*

The 2014 American Heart Association (AHA) and the ACC guidelines on the management of patients with valvular disease and the 2017 AHA and ACC focused update do not make specific recommendations on the treatment of primary pulmonary valve disease (stenosis or regurgitation), but instead referred to the 2008 guidelines for the management of adults with congenital heart disease.

In 2008, the AHA and ACC (in collaboration with other medical societies) issued guidelines for the management of adults with congenital heart disease. For patients with isolated valvular pulmonary stenosis, the guidelines make recommendations on balloon valvulotomy or surgical intervention; however, TPVI was not addressed. In 2015, an AHA scientific statement on congenital heart disease in older adults was published and meant to complement the 2008 ACC and AHA guidelines for adults with congestive heart disease. The intent was to outline the natural history, ramifications of childhood repair, and late initial diagnosis of congestive heart disease in older adults. The statement commented on the emerging use of the currently available transcatheter valve repair devices for both pulmonary stenosis and pulmonary valve regurgitation primarily after repair of tetralogy of Fallot. There was a specific comment that contemporary morbidity, mortality, and durability of surgical pulmonary valve regurgitation are unknown and, therefore, no contemporaneous benchmark against which to compare transcatheter valve implantation.
Medicare National Coverage

State or federal mandates (eg, Federal Employee Program) may dictate that certain U.S. Food and Drug Administration–approved devices, drugs, or biologics may not be considered investigational, and thus these devices may be assessed only by their medical necessity.

Regulatory Status

Devices for transcatheter pulmonary valve implantation were initially cleared from marketing by the U.S. Food and Drug Administration (FDA) through the humanitarian device exemption (HDE) process or used off-label until approved by FDA through the premarket approval (PMA) process between 2015 and 2016 (see Table 2).

<table>
<thead>
<tr>
<th>Device</th>
<th>Manufacturer</th>
<th>Date Approved</th>
<th>PMA No.</th>
<th>Indications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Melody® Transcatheter Pulmonary Valve (TPV)</td>
<td>Medtronic</td>
<td>Jan 2010</td>
<td>H080002 (HDE)</td>
<td>Pulmonary valve replacement for pediatric and adult patients with a dysfunctional, noncompliant RVOT conduit</td>
</tr>
<tr>
<td>Melody TPV</td>
<td>Medtronic</td>
<td>Jan 2015</td>
<td>P140017</td>
<td>Pulmonary valve replacement for pediatric and adult patients with a dysfunctional, noncompliant RVOT conduit</td>
</tr>
<tr>
<td>Melody TPV</td>
<td>Medtronic</td>
<td>Feb 2017</td>
<td>P140017/S005</td>
<td>Valve-in-valve for patients with a dysfunctional surgical bioprosthetic pulmonary valve</td>
</tr>
<tr>
<td>SAPIEN XT™ Transcatheter Heart Valve (pulmonic)</td>
<td>Edwards Lifesciences</td>
<td>Feb 2016</td>
<td>P130009/S037</td>
<td>Pulmonary valve replacement for pediatric and adult patients with a dysfunctional, noncompliant RVOT conduit</td>
</tr>
</tbody>
</table>

HDE: humanitarian device exemption; PMA: premarket approval; RVOT: right ventricular outflow tract.
The Melody® Transcatheter Pulmonary Valve (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.

In January 2010, the Melody® TPV and the Ensemble® Transcatheter Valve Delivery System (Medtronic) were approved by FDA under the HDE program 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 is 16 mm or greater in diameter when originally implanted, and
- Dysfunctional RVOT conduits with clinical indication for intervention, and either:
  - regurgitation: moderate-to-severe regurgitation, or
  - stenosis: mean RVOT gradient ≥35 mm Hg

On January 27, 2015, approval of the Melody® system was amended to a PMA because FDA determined that the device represented a breakthrough technology. The PMA was based, in part, on 2 prospective clinical studies, the Melody TPV Long-term Follow-up Post Approval Study and the Melody TPV New Enrollment Post Approval Study.

On February 24, 2017, approval of the Melody® system was expanded to include patients with a dysfunctional surgical bioprosthetic valve (valve-in-valve).

The Edwards SAPIEN XT™ Transcatheter Heart Valve (Pulmonic) (Edwards Lifesciences) 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. On February 29, 2016, it was approved by FDA as a supplement “for use in pediatric and adult patients with a dysfunctional, noncompliant Right Ventricular Outflow Tract (RVOT) conduit with a clinical indication for intervention and:

- pulmonary regurgitation ≥ moderate and/or
The approval for the pulmonic valve indication is a supplement to the 2014 PMA for use of the Edwards SAPIEN XT™ Transcatheter Heart Valve System for relief of aortic stenosis in patients with symptomatic heart disease due to severe native calcific aortic stenosis and who are judged by a heart team, including a cardiac surgeon, to be at high or greater risk for open surgical therapy (ie, Society of Thoracic Surgeons operative risk score ≥8% or at a ≥15% risk of mortality at 30 days). FDA product code: NPV

References


### History

<table>
<thead>
<tr>
<th>Date</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>01/10/12</td>
<td>New Policy – Policy created with literature search through June 15, 2011; considered medically necessary for patients who are high risk for open surgery and are poor surgical candidates due to multiple prior thoracotomies for open heart surgery. Considered investigational for all other indications. Clinical vetting information added.</td>
</tr>
<tr>
<td>09/27/12</td>
<td>Update Coding Section – ICD-10 codes are now effective 10/01/2014.</td>
</tr>
<tr>
<td>01/29/13</td>
<td>Replace policy. Policy updated with literature review, references 4, 5, 13-15, 17 added. Medically necessary statement amended to include “when performed according to FDA-approved indications”.</td>
</tr>
<tr>
<td>01/21/14</td>
<td>Replace policy. Policy updated with literature review, references 4, 5, 13-15, 17 added. Medically necessary statement amended to include “when performed according to FDA-approved indications”. Policy updated with literature review through September 30, 2013. References 13, 14, 16, 18, 21 added. No change to policy statement. Remove ICD-9 procedure and diagnosis codes; remove all ICD-10 codes except 02RH4JZ (which specifically applies) – these will not be used for adjudication.</td>
</tr>
<tr>
<td>09/23/14</td>
<td>Update Related Policies. Add 2.02.30.</td>
</tr>
<tr>
<td>01/19/16</td>
<td>Coding update. New CPT code 33477, effective 1/1/16, added to policy.</td>
</tr>
<tr>
<td>02/01/16</td>
<td>Coding update. Added 93799.</td>
</tr>
<tr>
<td>09/01/16</td>
<td>Annual Review, approved August 9, 2016. Policy updated with literature review through April 28, 2016; references 3, 5, 7-8, 16, 19, and 36-37 added. Policy statement unchanged. CPT coding updated.</td>
</tr>
<tr>
<td>08/01/18</td>
<td>Annual Review, approved July 10, 2018. Policy updated with literature review through May 2017; references 9, 22, and 43 added. Clinical input was obtained and policy statement changed to: Transcatheter pulmonary valve implantation is considered medically necessary for patients with congenital heart disease and current right ventricular outflow tract obstruction or regurgitation when specified indications are</td>
</tr>
</tbody>
</table>
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fechas para mantener su cobertura médica o ayuda con los costos. Usted
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