Clinical Policy: Pediatric Heart Transplant
Reference Number: PA.CP.MP.138
Effective Date: 01/18
Last Review Date: 10/17

Description
Pediatric heart disease may be a progressive disease which affects cardiac structure and function in infants and children. Heart transplantation is treatment of choice for pediatric patients with end-stage heart disease. This policy establishes the medical necessity requirements for pediatric heart transplants and re-transplants.

Policy/Criteria
I. It is the policy of Pennsylvania Health and Wellness® that heart transplant for pediatric members (age < 21) with end-stage heart disease is medically necessary when all of the following conditions are met:
   A. End-stage heart disease due to any of the following indications:
      1. For heart transplantation
         a. Systemic ventricular dysfunction with cardiomyopathies or previously repaired/palliated congenital heart disease;
         b. Heart failure associated with severe limitation of exercise and activity.
         c. Heart failure associated with systemic ventricular dysfunction in patients with cardiomyopathies or previously repaired/palliated congenital heart disease (CHD) when heart failure is associated with significant growth failure attributable to the heart disease;
         d. Heart failure with associated near sudden death and/or life-threatening arrhythmias untreatable with medications or an implantable defibrillator;
         e. Restrictive cardiomyopathy disease associated with reactive pulmonary hypertension;
         f. Pulmonary hypertension and a potential risk of developing fixed, irreversible elevation of pulmonary vascular resistance that could preclude orthotopic heart transplantation in the future;
         g. Certain anatomic and physiological conditions likely to worsen the natural history of CHD in infant patients with a functional single ventricle, which can lead to use of heart transplantation as primary therapy, including any of the following:
            i. Severe stenosis (stenoses) or atresia in proximal coronary arteries;
            ii. Moderate to severe stenosis and/or insufficiency of the AV and/or systemic semilunar valve(s);
            iii. Severe ventricular dysfunction;
         h. Several anatomic and physiological conditions likely to worsen the natural history of previously repaired or palliated CHD that may lead to consideration for heart transplantation without severe systemic ventricular dysfunction, including any of the following:
            i. Severe aortic or systemic AV valve insufficiency that is not considered amenable to surgical correction;
            ii. Severe arterial oxygen desaturation (cyanosis) that is not considered amenable to surgical correction;
            iii. Persistent protein-losing enteropathy despite optimal medical/surgical therapy;
2. For heart re-transplantation
   a. Moderate to severe graft vasculopathy;
   B. American Heart Association Stage C or Stage D heart disease, as per Table 1;
   C. Adequate functional status with the ability for rehabilitation;
   D. Life expectancy in the absence of cardiopulmonary disease ≥ 1 year or is consistent with
      the certified PA Transplant Center criteria for life expectancy;
   E. Does not have any of the following contraindications:
      1. Severe, irreversible, fixed elevation of pulmonary vascular resistance;
      2. Severe hypoplasia of the central branch pulmonary arteries or pulmonary veins;
      3. Any specific congenital heart lesion;
      4. Malignancy in the past year, except for non-melanoma localized skin cancer that has
         been treated appropriately;
      5. Untreatable significant dysfunction of another major organ system unless combined
         organ transplantation can be performed;
      6. Uncorrected atherosclerotic disease with suspected or confirmed end-organ ischemia
         or dysfunction and/or coronary artery disease not amenable to revascularization;
      7. Acute medical instability, including, but not limited to, acute sepsis, myocardial
         infarction, and liver failure;
      8. Uncorrectable bleeding diathesis;
      9. Chronic or latent highly virulent infections that is poorly controlled pre-transplant,
         including any of the following:
         a. Hepatitis B
         b. Hepatitis C
         c. Uncontrolled HIV/AIDS, defined as:
            i. CD4 count ≤ 200 cells/µl;
            ii. HIV-1 RNA viral load is detectable or inconsistently suppressed;
            iii. Member is on stable antiviral therapy for < 3 months;
            iv. Active, untreated complications associated with or secondary to HIV (i.e.
                opportunistic infections such as aspergillus, tuberculosis, coccidiomycosis,
                or resistant fungal infections, or neoplasms such as Kaposi’s sarcoma or non-
                Hodgkin’s lymphoma);
      10. Evidence of active *Mycobacterium tuberculosis* infection;
      11. Significant chest wall/spinal deformity expected to cause severe restriction after
          transplantation;
      12. Current non-adherence to medical therapy or a history of repeated or prolonged
          episodes of non-adherence to medical therapy that are perceived to increase the risk
          of non-adherence after transplantation;
      13. Psychiatric or psychological condition associated with the inability to cooperate or
          comply with medical therapy;
      14. Absence of an adequate or reliable social support system;
      15. Severely limited functional status with poor rehabilitation potential;
      16. Substance abuse or dependence (including tobacco and alcohol) without convincing
          evidence of risk reduction behaviors, such as meaningful and/or long-term
          participation in therapy for substance abuse and/or dependence. Serial blood and
          urine testing may be used to verify abstinence from substances that are of concern.
Background
Pediatric heart disease incorporates a wide range of diseases and includes a variety of age ranges. Heart transplantation is recommended for end-stage pediatric heart disease. Cardiomyopathy is the most common indication for heart transplant in children and dilated cardiomyopathy is the most common form of cardiomyopathy in the pediatric population, followed by hypertrophic and restrictive diseases.1

The American Heart Association has published a scientific statement specifically to address the requirements for heart transplantation and re-transplantations in pediatric heart disease.1 Canter, et al, addresses the indications for heart transplants, as well as defines the staging of heart failure as illustrated in Table 1.

The current survival in pediatric recipients 1, 5, and 10 years after transplantation is approximately 90, 80, and 60%, respectively.2 The median survival is 19.7 years for infants, 16.8 years for children ages 1-5, 14.5 years for children ages 6-10, and 12.4 years for children ages 11-17 at the time of transplantation.3 Several risk factors contribute to the decreasing survival in older ages groups, including immature immune system in infants, the absence of preformed antibodies in infants, sensitization in the older children due to surgical repair for congestive heart disease, and medication non-compliance in older children.3

Dipchand, et al, analyzed the Registry of the International Society for Heart and Lung Transplantation and reported that the proportion of transplant recipients by age remains similar with 24% infants, 25% aged between 1 and 5 years, 16% aged between 6 and 10 years, and 35% aged between 11 and 17 years.5

![Table 1: Heart Failure Stages in Pediatric Heart Disease](image)

<table>
<thead>
<tr>
<th>Classification</th>
<th>Characteristics</th>
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<tbody>
<tr>
<td>A</td>
<td>At high risk for developing heart failure</td>
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<tr>
<td>B</td>
<td>Abnormal cardiac structure and/or function; no symptoms of heart failure</td>
</tr>
<tr>
<td>C</td>
<td>Abnormal cardiac structure and/or function; past or present symptoms of heart failure</td>
</tr>
<tr>
<td>D</td>
<td>Abnormal structure and/or function; continuous infusion of intravenous inotropes or prostaglandin E1 to maintain of a ductus arteriosus; mechanical ventilatory and/or mechanical circulatory support</td>
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Coding Implications
This clinical policy references Current Procedural Terminology (CPT®). CPT® is a registered trademark of the American Medical Association. All CPT codes and descriptions are copyrighted 2015, American Medical Association. All rights reserved. CPT codes and CPT descriptions are from the current manuals and those included herein are not intended to be all-inclusive and are included for informational purposes only. Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.
**Clinical Policy**

**Pediatric Heart Transplant**

<table>
<thead>
<tr>
<th>CPT® Codes</th>
<th>Description</th>
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<tbody>
<tr>
<td>33944</td>
<td>Backbench standard preparation of cadaver donor heart allograft prior to transplantation, including dissection of allograft from surrounding soft tissues to prepare aorta, superior vena cava, inferior vena cava, pulmonary artery, and left atrium for implantation</td>
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<tr>
<td>33945</td>
<td>Heart transplant, with or without recipient cardiectomy</td>
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<th>HCPCS Codes</th>
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**ICD-10-CM Diagnosis Codes that Support Coverage Criteria**

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<thead>
<tr>
<th>ICD-10-CM Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>I25.1 – I25.9</td>
<td>Chronic ischemic heart disease</td>
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<tr>
<td>I42.0 – I42.9</td>
<td>Cardiomyopathy</td>
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<tr>
<td>I50.1 – I50.9</td>
<td>Heart failure</td>
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<tr>
<td>Q20.0 – Q28.9</td>
<td>Congenital malformations of circulatory system</td>
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**Reviews, Revisions, and Approvals**

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<th>New policy developed, specialist reviewed</th>
<th>Date</th>
<th>Approval Date</th>
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<td></td>
<td>12/16</td>
<td>1/17</td>
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**References**


2. Singh RK. “Management of heart failure in infants and children” In: UpToDate, Armsby C. (Ed), UpToDate, Waltham, MA. (Accessed on September 26, 2016.)

