Clinical Policy: Pediatric Heart Transplant

Reference Number: PA.CP.MP.138
Effective Date: 01/18
Last Review Date: 10/17

Coding Implications
Revision Log

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]:

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, coccidiodomycosis, 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 trasnplanation.[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

Classification	Characteristics
A	At high risk for developing heart failure
B	Abnormal cardiac structure and/or function; no symptoms of heart failure
C	Abnormal cardiac structure and/or function; past or present symptoms of heart failure
D	Abnormal structure and/or function; continuous infusion of intravenous inotropes or prostaglandin E_1 to maintain of a ductus arteriosus; mechanical ventilatory and/or mechanical circulatory support

Coding Implications

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CPT® Codes	Description
33944	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
33945	Heart transplant, with or without recipient cardiectomy

HCPCS Codes	Description
N/A	

ICD-10-CM Diagnosis Codes that Support Coverage Criteria

ICD-10-CM Code	Description
I25.1 – I25.9	Chronic ischemic heart disease
I42.0 – I42.9	Cardiomyopathy
I50.1 – I50.9	Heart failure
Q20.0 – Q28.9	Congenital malformations of circulatory system

Reviews, Revisions, and Approvals	Date	Approval Date
New policy developed, specialist reviewed	12/16	1/17

References

1. Canter, Charles E., et al. "Indications for Heart Transplantation in Pediatric Heart Disease A Scientific Statement from the American Heart Association Council on Cardiovascular Disease in the Young; the Councils on Clinical Cardiology, Cardiovascular Nursing, and Cardiovascular Surgery and Anesthesia; and the Quality of Care and Outcomes Research Interdisciplinary Working Group." *Circulation* 115.5 (2007): 658-676.
2. Singh RK. "Management of heart failure in infants and children" In: UpToDate, Armsby C. (Ed), UpToDate, Waltham, MA. (Accessed on September 26, 2016.)
3. Thrush, Philip T., and Timothy M. Hoffman. "Pediatric heart transplantation—indications and outcomes in the current era." *Journal of thoracic disease* 6.8 (2014): 1080.
4. Pahl, Elfriede, Anne I. Dipchand, and Michael Burch. "Heart transplantation for heart failure in children." *Heart failure clinics* 6.4 (2010): 575-589.
5. Dipchand, Anne I., et al. "The registry of the International Society for Heart and Lung Transplantation: seventeenth official pediatric heart transplantation report--2014; focus theme: retransplantation." *The Journal of heart and lung transplantation: the official publication of the International Society for Heart Transplantation* 33.10 (2014): 985.