Clinical Policy: Pediatric Heart Transplant
Reference Number: CP.MP.138
Review Date: 01/19

See Important Reminder at the end of this policy for important regulatory and legal information.

Description
Pediatric heart disease may be a progressive disease, affecting cardiac structure and function in infants and children. Heart transplantation is the 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 health plans affiliated with Centene Corporation® that heart transplant for pediatric members (age < 18) 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. Stage D heart failure (see table 1) associated with systemic ventricular dysfunction with cardiomyopathies or previously repaired/palliated congenital heart disease (CHD);
         b. Stage C heart failure associated with severe limitation of exercise and activity, evidenced by peak maximum oxygen consumption < 50% predicted for age and sex;
         c. Stage C heart failure associated with systemic ventricular dysfunction in patients with cardiomyopathies or previously repaired/palliated CHD when heart failure is associated with significant growth failure attributable to the heart disease;
         d. Stage C heart failure with associated near sudden death, and/or life-threatening arrhythmias untreatable with medications or an implantable defibrillator;
         e. Stage C heart failure in restrictive cardiomyopathy disease associated with reactive pulmonary hypertension;
         f. Stage C heart failure associated with reactive 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 atroventricular (AV) and/or systemic semilunar valve(s);
            iii. Severe ventricular dysfunction;
         h. Certain anatomic and physiological conditions likely to worsen the natural history of previously repaired or palliated CHD in patients with stage C heart failure that may lead to consideration for heart transplantation without severe systemic ventricular dysfunction, including any of the following:
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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 cardiac graft vasculopathy;
   B. Adequate functional status with the ability for rehabilitation;
   C. Life expectancy in the absence of cardiopulmonary disease ≥ 2 years;
   D. 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, except in circumstances noted in I.A.;
      4. AL amyloidosis (exceptions may be made where curative therapy of amyloidosis has been performed or is planned, such as with stem cell transplantation in primary amyloidosis, or with liver transplantation in familial amyloidosis);
      5. Retransplantation when performed during an episode of ongoing, acute allograft rejection, even in the presence of graft vasculopathy;
      6. Retransplantation when performed during the first 6 months after primary transplantation;
      7. Malignancy in the past two years, except for non-melanoma localized skin cancer that has been treated appropriately;
      8. Untreatable significant dysfunction of another major organ system unless combined organ transplantation can be performed;
      9. Uncorrected atherosclerotic disease with suspected or confirmed end-organ ischemia or dysfunction and/or coronary artery disease not amenable to revascularization;
      10. Acute medical instability, including, but not limited to, acute sepsis, myocardial infarction, and liver failure;
      11. Uncorrectable bleeding diathesis;
      12. Chronic infection with highly virulent and/or resistant microbes that are poorly controlled pre-transplant;
      13. Evidence of active *Mycobacterium tuberculosis* infection;
      14. Significant chest wall/spinal deformity expected to cause severe restriction after transplantation;
      15. Class II or III obesity (body mass index ≥ 35.0 kg/m²);
      16. 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;
      17. Psychiatric or psychological condition associated with the inability to cooperate or comply with medical therapy;
      18. Absence of an adequate or reliable social support system;
      19. 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.
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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 and 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 congenital 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 the proportion of transplant recipients by age accordingly: 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

<table>
<thead>
<tr>
<th>Classification</th>
<th>Characteristics</th>
</tr>
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<tbody>
<tr>
<td>A</td>
<td>At high risk for developing heart failure</td>
</tr>
<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 patency of a ductus arteriosus; mechanical ventilatory and/or mechanical circulatory support</td>
</tr>
</tbody>
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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 2019, 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.
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<tr>
<th>CPT® Codes</th>
<th>Description</th>
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<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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<tr>
<th>HCPCS Codes</th>
<th>Description</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>
</tr>
<tr>
<td>I42.0 – I42.9</td>
<td>Cardiomyopathy</td>
</tr>
<tr>
<td>I50.1 – I50.9</td>
<td>Heart failure</td>
</tr>
<tr>
<td>Q20.0 – Q28.9</td>
<td>Congenital malformations of circulatory system</td>
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**Reviews, Revisions, and Approvals**

<table>
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<th>Date</th>
<th>Approval Date</th>
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<td>12/16</td>
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<td>01/19</td>
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</table>

**References**

1. Canter, CE. 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.
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Important Reminder
This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical policy; and other available clinical information. The Health Plan makes no representations and accepts no liability with respect to the content of any external information used or relied upon in developing this clinical policy. This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved. “Health Plan” means a health plan that has adopted this clinical policy and that is operated or administered, in whole or in part, by Centene Management Company, LLC, or any of such health plan’s affiliates, as applicable.

The purpose of this clinical policy is to provide a guide to medical necessity, which is a component of the guidelines used to assist in making coverage decisions and administering benefits. It does not constitute a contract or guarantee regarding payment or results. Coverage decisions and the administration of benefits are subject to all terms, conditions, exclusions and limitations of the coverage documents (e.g., evidence of coverage, certificate of coverage, policy, contract of insurance, etc.), as well as to state and federal requirements and applicable Health Plan-level administrative policies and procedures.

This clinical policy is effective as of the date determined by the Health Plan. The date of posting may not be the effective date of this clinical policy. This clinical policy may be subject to applicable legal and regulatory requirements relating to provider notification. If there is a discrepancy between the effective date of this clinical policy and any applicable legal or regulatory requirement, the requirements of law and regulation shall govern. The Health Plan retains the right to change, amend or withdraw this clinical policy, and additional clinical policies may be developed and adopted as needed, at any time.

This clinical policy does not constitute medical advice, medical treatment or medical care. It is not intended to dictate to providers how to practice medicine. Providers are expected to exercise professional medical judgment in providing the most appropriate care, and are solely responsible for the medical advice and treatment of members. This clinical policy is not intended to recommend treatment for members. Members should consult with their treating physician in connection with diagnosis and treatment decisions.
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**Note: For Medicaid members**, when state Medicaid coverage provisions conflict with the coverage provisions in this clinical policy, state Medicaid coverage provisions take precedence. Please refer to the state Medicaid manual for any coverage provisions pertaining to this clinical policy.

**Note: For Medicare members**, to ensure consistency with the Medicare National Coverage Determinations (NCD) and Local Coverage Determinations (LCD), all applicable NCDs, LCDs, and Medicare Coverage Articles should be reviewed prior to applying the criteria set forth in this clinical policy. Refer to the CMS website at [http://www.cms.gov](http://www.cms.gov) for additional information.

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