

# Clinical Policy: Pediatric Heart Transplant

Reference Number: CP.MP.138

Review Date: 01/19

[Coding Implications](#)

[Revision Log](#)

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## 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<sup>®</sup> 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<sup>1</sup>:
    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 atrioventricular (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:

- 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  $\geq 2$  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  $\geq 35.0$  kg/m<sup>2</sup>);
  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.<sup>1</sup>

The American Heart Association has published a scientific statement specifically to address the requirements for heart transplantation and re-transplantations in pediatric heart disease.<sup>1</sup> 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.<sup>2</sup> 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.<sup>3</sup> 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.<sup>3</sup>

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.<sup>5</sup>

**Table 1: Heart Failure Stages in Pediatric Heart Disease**

<b>Classification</b>	<b>Characteristics</b>
<b>A</b>	At high risk for developing heart failure
<b>B</b>	Abnormal cardiac structure and/or function; no symptoms of heart failure
<b>C</b>	Abnormal cardiac structure and/or function; past or present symptoms of heart failure
<b>D</b>	Abnormal structure and/or function; continuous infusion of intravenous inotropes or prostaglandin E <sub>1</sub> to maintain patency 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	01/17
Added AL amyloidosis as a contraindication.	01/18	01/18
Specified for each indication the stage of heart failure required, and removed general criteria stating that patient is in stage C or D heart failure. Removed “severely limited functional status with poor rehab potential,” as it is included in criteria requiring “adequate functional status with the ability for rehabilitation.”	05/18	
References reviewed and updated	01/19	01/19

**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.
2. Singh RK, Singh TP. Heart failure in children: management. UpToDate, Arnsby C. (Ed), UpToDate, Waltham, MA. Accessed January 9, 2019.
3. Thrush PT, Hoffman TM. "Pediatric heart transplantation—indications and outcomes in the current era." *Journal of thoracic disease* 6.8 (2014): 1080.
4. Elfriede P, Dipchand AI, Burch M. "Heart transplantation for heart failure in children." *Heart failure clinics* 6.4 (2010): 575-589.
5. Dipchand AI, et al. "The registry of the International Society for Heart and Lung Transplantation: seventeenth official pediatric heart transplantation report--2014; focus

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- theme: retransplantation." *The Journal of heart and lung transplantation: the official publication of the International Society for Heart Transplantation* 33.10 (2014): 985.
6. Hsu DT, Lamour JM. Changing indications for pediatric heart transplantation: complex congenital heart disease. *Circulation*. 2015 Jan 6;131(1):91-9. doi: 10.1161/CIRCULATIONAHA.114.001377.
  7. Masarone D, Valente F, Rubino M, et al. Pediatric heart failure: a practical guide to diagnosis and management. *Pediatr Neonatol*. 2017 Aug;58(4):303-312. doi: 10.1016/j.pedneo.2017.01.001.
  8. McKenna WJ. Treatment of amyloid cardiomyopathy. UpToDate. Kyle RA (Ed), UpToDate, Waltham, MA. Accessed January 10, 2019.

### **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.

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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> for additional information.

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