Clinical Policy: Pediatric Heart Transplant

Reference Number: CP.MP.138
Review Date: 12/20

Coding Implications
Revision Log

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/enrollees (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, one of the following:*
   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 any of the following:
      i. Severe limitation of exercise and activity, evidenced by peak maximum oxygen consumption < 50% predicted for age and sex;
      ii. 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;
      iii. Near sudden death, and/or life-threatening arrhythmias untreatable with medications or an implantable defibrillator;
      iv. Restrictive cardiomyopathy disease associated with reactive pulmonary hypertension;
      v. 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;
      vi. Certain 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:
         a) Severe aortic or systemic AV valve insufficiency that is not considered amenable to surgical correction;
         b) Severe arterial oxygen desaturation (cyanosis) that is not considered amenable to surgical correction;
         c) Persistent protein-losing enteropathy despite optimal medical/surgical therapy;

c. 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;

2. *For heart re-transplantation*, 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. Amyloid light-chain (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, except for non-melanoma localized skin cancer that has been treated appropriately, a malignancy that has been completely resected, or a treated malignancy determined to have a small likelihood of recurrence and acceptable future risks;
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;
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. BMI ≥120% of the 95th percentile (varies by sex and age; see https://www.cdc.gov/growthcharts/clinical_charts.htm for BMI percentile by age, and refer to Appendix A for 120% of the 95th percentile values);
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.

E. All reversible causes of heart failure have been ruled out such as, but not limited to, anemia, hypertension, renal failure, acidosis, obesity, malnutrition, respiratory disorders and thyroid disorders.

Appendix A:
Steps to determine 120% of 95th BMI percentile by age:

1. Calculate BMI by age, with percentile, here: https://www.cdc.gov/healthyweight/bmi/calculator.html
2. In the row corresponding to the child's age, look at the last column of either the male or female chart to determine if the child's BMI falls at or above 120% of the 95th percentile BMI value.

Male BMI Chart		
Age in years	**95th percentile BMI***	**BMI 120% of 95th percentile**
2- < 2.5	19.3-18.7	23.2-22.4
2.5- < 3	18.7-18.2	22.4-21.8
3- < 3.5	18.2-18.0	21.8-21.6
3.5- < 4	18.0-17.8	21.6-21.4
4- < 4.5	17.8	21.4
4.5- < 5	17.8-17.9	21.4-21.5
5- < 5.5	17.9-18.1	21.5-21.7
5.5- < 6	18.1-18.4	21.7-22.1
6- < 6.5	18.4-18.8	22.1-22.6
6.5- < 7	18.8-19.2	22.6-23.0
7- < 7.5	19.2-19.6	23.0-23.5
7.5- < 8	19.6-20.1	23.5-24.1
8- < 8.5	20.1-20.6	24.1-24.7
8.5- < 9	20.6-21.1	24.7-25.3
9- < 9.5	21.1-21.6	25.3-25.9
9.5- < 10	21.6-22.2	25.9-26.6
10- < 10.5	22.2-22.7	26.6-27.2
10.5- < 11	22.7-23.2	27.2-27.8
11- < 11.5	23.2-23.7	27.8-28.4
11.5- < 12	23.7-24.2	28.4-29.0
12- < 12.5	24.2-24.7	29.0-29.6
12.5- < 13	24.7-25.2	29.6-30.2
13- < 13.5	25.2-25.6	30.2-30.7
13.5- < 14	25.6-26.0	30.7-31.2

Female BMI Chart		
Age in years	**95th percentile BMI***	**BMI 120% of 95th percentile**
2- < 2.5	19.1-18.6	22.9-22.3
2.5- < 3	18.6-18.3	22.3-22.0
3- < 3.5	18.3-18.1	22.0-21.7
3.5- < 4	18.1-18.0	21.7-21.6
4- < 4.5	18.0-18.1	21.6-21.7
4.5- < 5	18.1-18.2	21.7-21.8
5- < 5.5	18.2-18.5	21.8-22.2
5.5- < 6	18.5-18.8	22.2-22.6
6- < 6.5	18.8-19.2	22.6-23.0
6.5- < 7	19.2-19.6	23.0-23.5
7- < 7.5	19.6-20.1	23.5-24.1
7.5- < 8	20.1-20.7	24.1-24.8
8- < 8.5	20.7-21.2	24.8-25.4
8.5- < 9	21.2-21.8	25.4-26.2
9- < 9.5	21.8-22.4	26.2-26.9
9.5-< 10	22.4-23.0	26.9-27.6
10- < 10.5	23.0-23.6	27.6-28.3
10.5- < 11	23.6-24.1	28.3-28.9
11- < 11.5	24.1-24.7	28.9-29.6
11.5- < 12	24.7-25.2	29.6-30.2
12- < 12.5	25.2-25.8	20.2-31.0
12.5- < 13	25.8-26.3	31.0-31.6
13- < 13.5	26.3-26.8	31.6-32.2
13.5- < 14	26.8-27.3	32.2-32.8

14- < 14.5	26.0-26.4	31.2-31.7
14.5- < 15	26.4-26.8	31.7-32.2
15- < 15.5	26.8-27.2	32.2-32.6
15.5- < 16	27.2-27.6	32.6-33.1
16- < 16.5	27.6-27.9	33.1-33.5
16.5- < 17	27.9-28.2	33.5-33.8
17- < 17.5	28.2-28.6	33.8-34.3
17.5-< 18	28.6-29.0	34.3-34.8
18-<18.5	29.0-29.4	34.8-35.3
18.5-<19	29.4-29.7	35.3-35.6
19-19.5	29.7-30.1	35.6-36.1
19.5- <20	30.1-30.6	36.1-36.7

14- < 14.5	27.3-27.8	32.8-33.4
14.5- < 15	27.8-28.2	33.4-33.8
15- < 15.5	28.2-28.5	33.8-34.2
15.5- < 16	28.5-28.9	34.2-34.7
16- < 16.5	28.9-29.3	34.7-35.2
16.5- < 17	29.3-29.6	35.2-35.5
17- < 17.5	29.6-29.9	35.5-35.9
17.5-< 18	29.9-30.3	35.9-36.4
18-<18.5	30.3-30.6	36.4-36.7
18.5-<19	30.6-31.0	36.7-37.2
19-19.5	31.0-31.4	37.2-37.7
19.5- <20	31.4-31.8	37.7-38.2

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 rates 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 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

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 patency of a ductus arteriosus; mechanical ventilatory and/or mechanical circulatory support

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 2020, 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.

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

ICD-10-CM Diagnosis Codes that Support Coverage Criteria

ICD-10-CM Code	Description
I25.10– 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
Code I25.1 changed to I25.10. In D.15, replaced "Class II or III obesity (body mass index ≥35.0 kg/m$^{2)}$ with BMI ≥120% of the 95th percentile and added a link to the CDC clinical growth Charts. Added Appendix A with the 95th percentile values. Specialist reviewed.	01/20	01/20

Reviews, Revisions, and Approvals	Date	Approval Date
Edited malignancy contraindication to not specify within 2 years, and added exceptions of cancer that has been completely resected, or that has been treated and poses acceptable future risk. Removed coronary artery disease not amenable to revascularization from list of contraindications.	05/20	05/20
Reformatted criteria to group all class C heart failure scenarios together and added additional exclusion of ruling out reversible causes of heart failure. References reviewed and updated. Replaced "member" with "member/enrollee" in all instances.	12/20	12/20

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 website. www.uptodate.com. Published June 5, 2019. Accessed December 2, 2020.
3. Thrush PT, Hoffman TM. Pediatric heart transplantation—indications and outcomes in the current era. *J Thor Dis* 6.8 (2014): 1080.
4. Elfriede P, Dipchand AI, Burch M. Heart transplantation for heart failure in children." *Heart Fail Clin* 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 theme: retransplantation. *Jour Heart Lung transplant* 33.10 (2014): 985.
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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 website. www.uptodate.com. Published January 13, 2020. Accessed December 13, 2020.
9. Bolling, CF, Armstrong, SC, Reichard, KW, et al. Metabolic and bariatric surgery for pediatric patients with severe obesity. *Pediatrics* Dec 2019, 144 (6) e20193224.

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

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Note: For Medicaid members/enrollees, 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/enrollees, 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.