Clinical Policy: Pediatric Heart Transplant

Reference Number: LA.CP.MP.138 Coding Implications
Date of Last Revision: 11/23 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 Louisiana Healthcare Connections 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) Pulmonary hypertension and a potential risk of developing fixed, irreversible elevation of pulmonary vascular resistance that could preclude orthotopic heart transplantation in the future;
         b) Severe aortic or systemic AV valve insufficiency that is not considered amenable to surgical correction;
         c) Severe arterial oxygen desaturation (cyanosis) that is not considered amenable to surgical correction;

d) 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. Life expectancy in the absence of cardiopulmonary disease ≥ 2 years;

C. 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;

D. Does not have any of the following contraindications:
1. Glomerular filtration rate < 30 mL/min/1.73m2 unless being considered for multi-organ transplant;
2. HIV infection with detectable viral load, unless all of the following are documented:
   a. CD4 cell count >200 cells/mm^3,
   b. Absence of active AIDS-defining opportunistic infection (unless treated efficaciously or prevented, can be included on the heart transplant waiting list) or malignancy;
   c. Member/enrollee is currently on effective ART (antiretroviral therapy)[11];
3. Severe, irreversible, fixed elevation of pulmonary vascular resistance;
4. Severe hypoplasia of the central branch pulmonary arteries or pulmonary veins;
5. Any specific congenital heart lesion, except in circumstances noted in I.A.;
6. 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);
7. Retransplantation when performed during an episode of ongoing, acute allograft rejection, even in the presence of graft vasculopathy;
8. Retransplantation when performed during the first six months after primary transplantation;
9. Malignancy with high risk of recurrence or death related to cancer;
10. Acute liver failure, or cirrhosis with portal hypertension or synthetic dysfunction unless being considered for multi-organ transplant;
11. Acute renal failure with rising creatinine or on dialysis and low likelihood of recovery;
12. Other severe uncontrolled medical condition expected to limit survival after transplant;
13. Uncorrected atherosclerotic disease with suspected or confirmed end-organ ischemia or dysfunction;
14. Chronic infection with highly virulent and/or resistant microbes that are poorly controlled pre-transplant;
15. Active *tuberculosis* infection;
16. Progressive cognitive impairment;

17. Significant chest wall/spinal deformity expected to cause severe restriction after transplantation;
18. BMI ≥ 35 or BMI ≥ 120% of the 95th percentile (varies by sex and age), whichever is lower.; 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);
19. Inability to adhere to the regimen necessary to preserve the transplant, even with caregiver support;
20. Absence of an adequate or reliable social support system;
21. Active substance use or dependence including current tobacco use, vaping, marijuana use (unless prescribed by a licensed practitioner), or IV drug use without convincing evidence of risk reduction behaviors, such as meaningful and/or long-term participation in therapy for substance abuse and/or dependence (unless urgent transplant timelines are present, in which case a commitment to reducing behaviors is acceptable). Serial blood and urine testing may be used to verify abstinence from substances that are of concern.).

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 the chart corresponding to BMI for those with a male or female reproductive system to determine if the child's BMI falls at or above 120% of the 95th percentile BMI value.

BMI for those with a male reproductive system		
Age in years	**95th percentile BMI***	**BMI 120% of 95th percentile**
2 < 2.5	19.3 to 18.7	23.2 to 22.4
2.5 < 3	18.7 to 18.2	22.4 to 21.8
3 < 3.5	18.2 to 18.0	21.8 to 21.6
3.5 < 4	18.0 to 17.8	21.6 to 21.4
4 < 4.5	17.8	21.4
4.5 < 5	17.8 to 17.9	21.4 to 21.5
5 < 5.5	17.9 to 18.1	21.5 to 21.7
5.5 < 6	18.1 to 18.4	21.7 to 22.1
6 < 6.5	18.4 to 18.8	22.1 to 22.6
6.5 < 7	18.8 to 19.2	22.6 to 23.0
7 < 7.5	19.2 to 19.6	23.0 to 23.5
7.5 < 8	19.6 to 20.1	23.5 to 24.1
8 < 8.5	20.1 to 20.6	24.1 to 24.7
8.5 < 9	20.6 to 21.1	24.7 to 25.3
9 < 9.5	21.1 to 21.6	25.3 to 25.9
9.5 < 10	21.6 to 22.2	25.9 to 26.6
10 < 10.5	22.2 to 22.7	26.6 to 27.2

BMI for those with a female reproductive system		
Age in years	**95th percentile BMI***	**BMI 120% of 95th percentile**
2- < 2.5	19.1 to 18.6	22.9 to 22.3
2.5- < 3	18.6 to 18.3	22.3 to 22.0
3- < 3.5	18.3 to 18.1	22.0 to 21.7
3.5- < 4	18.1 to 18.0	21.7 to 21.6
4- < 4.5	18.0 to 18.1	21.6 to 21.7
4.5- < 5	18.1 to 18.2	21.7 to 21.8
5- < 5.5	18.2 to 18.5	21.8 to 22.2
5.5- < 6	18.5 to 18.8	22.2 to 22.6
6- < 6.5	18.8 to 19.2	22.6 to 23.0
6.5- < 7	19.2 to 19.6	23.0 to 23.5
7- < 7.5	19.6 to 20.1	23.5 to 24.1
7.5 < 8	20.1 to 20.7	24.1 to 24.8
8 < 8.5	20.7 to 21.2	24.8 to 25.4
8.5 < 9	21.2 to 21.8	25.4 to 26.2
9 < 9.5	21.8 to 22.4	26.2 to 26.9
9.5 < 10	22.4 to 23.0	26.9 to 27.6
10 < 10.5	23.0 to 23.6	27.6 to 28.3

BMI for those with a male reproductive system		
Age in years	**95th percentile BMI***	**BMI 120% of 95th percentile**
10.5 < 11	22.7 to 23.2	27.2 to 27.8
11 < 11.5	23.2 to 23.7	27.8 to 28.4
11.5 < 12	23.7 to 24.2	28.4 to 29.0
12 < 12.5	24.2 to 24.7	29.0 to 29.6
12.5 < 13	24.7 to 25.2	29.6 to 30.2
13 < 13.5	25.2 to 25.6	30.2 to 30.7
Age in years	**95th percentile BMI***	**BMI 120% of 95th percentile**
13.5 < 14	25.6 to 26.0	30.7 to 31.2
14 < 14.5	26.0 to 26.4	31.2 to 31.7
14.5 < 15	26.4 to 26.8	31.7 to 32.2
15 < 15.5	26.8 to 27.2	32.2 to 32.6
15.5 < 16	27.2 to 27.6	32.6 to 33.1
16 < 16.5	27.6 to 27.9	33.1 to 33.5
16.5 < 17	27.9 to 28.2	33.5 to 33.8
17 < 17.5	28.2 to 28.6	33.8 to 34.3
17.5 < 18	28.6 to 29.0	34.3 to 34.8
18 < 18.5	29.0 to 29.4	34.8 to 35.3
18.5 < 19	29.4 to 29.7	35.3 to 35.6
19 < 19.5	29.7 to 30.1	35.6 to 36.1
19.5 < 20	30.1 to 30.6	36.1 to 36.7

BMI for those with a female reproductive system		
Age in years	**95th percentile BMI***	**BMI 120% of 95th percentile**
10.5 < 11	23.6 to 24.1	28.3 to 28.9
11 < 11.5	24.1 to 24.7	28.9 to 29.6
11.5 < 12	24.7 to 25.2	29.6 to 30.2
12 < 12.5	25.2 to 25.8	20.2 to 31.0
12.5 < 13	25.8 to 26.3	31.0 to 31.6
13 < 13.5	26.3 to 26.8	31.6 to 32.2
Age in years	**95th percentile BMI***	**BMI 120% of 95th percentile**
13.5 < 14	26.8 to 27.3	32.2 to 32.8
14 < 14.5	27.3 to 27.8	32.8 to 33.4
14.5 < 15	27.8 to 28.2	33.4 to 33.8
15 < 15.5	28.2 to 28.5	33.8 to 34.2
15.5 < 16	28.5 to 28.9	34.2 to 34.7
16 < 16.5	28.9 to 29.3	34.7 to 35.2
16.5 < 17	29.3 to 29.6	35.2 to 35.5
17 < 17.5	29.6 to 29.9	35.5 to 35.9
17.5 < 18	29.9 to 30.3	35.9 to 36.4
18 < 18.5	30.3 to 30.6	36.4 to 36.7
18.5 < 19	30.6 to 31.0	36.7 to 37.2
19 < 19.5	31.0 to 31.4	37.2 to 37.7
19.5 < 20	31.4 to 31.8	37.7 to 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 one, five, and ten years after transplantation is approximately 90, 80, and 60%, respectively.[2] The median survival is 22.3 years for infants, 18.4 years for children ages one through five, 14.4 years for children ages six to ten, and 13.1 years for children ages 11 years or older at the time of trasnplanation.[10] 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 one to five, 16% aged between six to ten 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 2022, 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 and may not support medical necessity. 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.

NOTE: Coverage is subject to each requested code's inclusion on the corresponding LDH fee schedule. Non-covered codes are denoted (*) and are reviewed for Medical Necessity for members under 21 years of age on a per case basis.

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

Reviews, Revisions, and Approvals	Revision Date	Approval Date
Converted corporate to local policy.	12/31/2020	
In I.C., replaced "adequate functional status with ability for rehabilitation" and contraindications regarding past or current nonadherence to medical therapy, and psychological condition associated with the inability to comply with medical therapy with "Inability to adhere to the regimen necessary to preserve the transplant, even with caregiver support." Changed "review date" in header to "Date of Last Revision" and "Date" in the revision log header to "Revision Date." Added "and may not support medical necessity" to coding implications. Annual review. Revised I.C.13, from "BMI ≥ 120% of the 95th percentile (varies by sex and age)" to "BMI ≥ 35 or BMI ≥ 120% of the 95th percentile (varies by sex and age), whichever is lower." References reviewed and updated. Reviewed by specialist.	2/22	
Moved criterion "all reversible causes of heart failure have been ruled out…" to I.C, and moved contraindications to I.D. Edited contraindications: added GFR rate; added "Acute liver failure or cirrhosis…", added acute renal failure; added HIV infection with detectable viral load; added septic shock; added progressive cognitive impairment; replaced "untreatable significant dysfunction of another major organ system.." with "Other severe uncontrolled medical condition expected to limit survival after transplant"; slightly reworded substance use contraindication; removed "acute medical instability…" and "uncorrectable bleeding diathesis;" replaced "malignancy, except for non-melanoma…" with "Malignancy with high risk of recurrence or death related to cancer." Added "and may not support medical necessity" to Coding Implications section	5/22	8/13/22
Annual review. Appendix A tables updated to remove dashes. Removed ICD-10 codes. References reviewed and reformatted. Added CPT code 33940.	1/23	4/10/23
Annual review. Added additional criteria I.A.1.b.vi.a., pulmonary hypertension and a potential risk of developing fixed, irreversible elevation of pulmonary vascular resistance that could preclude orthotopic heart transplantation in the future. Updated I.D.1. from GFR < 40 mL/min/1.73m2 to GFR < 30 mL/min/1.73m2. Expanded I.D.2. with qualifying criteria for members who are HIV positive. Updated I.D.21. to exclude marijuana use when prescribed by a licensed practitioner and include required commitment to reducing substance use behaviors if urgent transplant timelines are present. Background reviewed and updated. References reviewed and updated. Reviewed by external specialist.	10/23	1/23/24

References

1. Canter CE, Shaddy RE, Bernstein D, 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 [published correction appears in Circulation. 2007 Apr 3;115(13):e385. Friedman, Allen H [corrected to Friedman, Alan H]]. *Circulation*. 2007;115(5):658 to 676. doi:10.1161/CIRCULATIONAHA.106.180449
2. Singh RK, Singh TP. Heart failure in children: management. UpToDate. www.uptodate.com Published June 5, 2019. Accessed August 30, 2023.
3. Thrush PT, Hoffman TM. Pediatric heart transplantation-indications and outcomes in the current era. *J Thorac Dis*. 2014;6(8):1080 to 1096. doi:10.3978/j.issn.2072-1439.2014.06.16
4. Pahl E, Dipchand AI, Burch M. Heart transplantation for heart failure in children. *Heart Fail Clin*. 2010;6(4):575-x. doi:10.1016/j.hfc.2010.05.010
5. Dipchand AI, Edwards LB, Kucheryavaya AY, et al. The registry of the International Society for Heart and Lung Transplantation: seventeenth official pediatric heart transplantation report--2014; focus theme: retransplantation. *J Heart Lung Transplant*. 2014;33(10):985 to 995. doi:10.1016/j.healun.2014.08.002
6. Hsu DT, Lamour JM. Changing indications for pediatric heart transplantation: complex congenital heart disease. *Circulation*. 2015;131(1):91 to 99. 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;58(4):303 to 312. doi:10.1016/j.pedneo.2017.01.001
8. McKenna WJ. Treatment of amyloid cardiomyopathy. UpToDate. www.uptodate.com. Published March 23, 2021. Accessed August 31, 2023.
9. Bolling CF, Armstrong SC, Reichard KW, Michalsky MP; SECTION ON OBESITY, SECTION ON SURGERY. Metabolic and Bariatric Surgery for Pediatric Patients With Severe Obesity. *Pediatrics*. 2019;144(6):e20193224. doi:10.1542/peds.2019-3224
10. Dipchand AI. Current state of pediatric cardiac transplantation. *Ann Cardiothorac Surg*. 2018;7(1):31-55. doi:10.21037/acs.2018.01.07
11. Agüero F, Castel MA, Cocchi S, et al. An Update on Heart Transplantation in Human Immunodeficiency Virus-Infected Patients. *Am J Transplant*. 2016;16(1):21-28. doi:10.1111/ajt.13496
12. Roest S, Hesselink DA, Klimczak-Tomaniak D, et al. Incidence of end-stage renal disease after heart transplantation and effect of its treatment on survival. *ESC Heart Fail*. 2020;7(2):533-541. doi:10.1002/ehf2.12585

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. LHCC makes no representations and accepts no liability with respect to the content of any external information used or relied upon in developing

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