

Clinical Policy: Pediatric Heart Transplant

Reference Number: LA.CP.MP.138

Date of last Revision: 1/23

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 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 indications¹:
 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. 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 < 40 mL/min/1.73m² unless being considered for multi-organ transplant;
 2. HIV infection with detectable viral load;
 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 6 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 $\geq 120\%$ 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 smoking, 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. 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	95 th percentile BMI*	BMI 120% of 95 th 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
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
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

BMI for those with a female reproductive system		
Age in years	95 th percentile BMI*	BMI 120% of 95 th 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
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	30.2 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
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	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

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 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

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 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.

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
33940	Donor cardiectomy (including cold preservation)

HCPCS Codes	Description
N/A	

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	5/22	8/13/22

Reviews, Revisions, and Approvals	Revision Date	Approval Date
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		
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

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 November 29, 2022.
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 November 29, 2022.
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

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 this clinical policy. This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved.

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 LHCC administrative policies and procedures.

This clinical policy is effective as of the date determined by LHCC. 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. LHCC 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/enrollees. This clinical policy is not intended to recommend treatment for members/enrollees. Members/enrollees should consult with their treating physician in connection with diagnosis and treatment decisions.

Providers referred to in this clinical policy are independent contractors who exercise independent judgment and over whom LHCC has no control or right of control. Providers are not agents or employees of LHCC.

This clinical policy is the property of LHCC. Unauthorized copying, use, and distribution of this clinical policy or any information contained herein are strictly prohibited. Providers, members/enrollees and their representatives are bound to the terms and conditions expressed herein through the terms of their contracts. Where no such contract exists, providers, members/enrollees and their representatives agree to be bound by such terms and conditions by providing services to members/enrollees and/or submitting claims for payment for such services.

©2020 Louisiana Healthcare Connections. All rights reserved. All materials are exclusively owned by Louisiana Healthcare Connections and are protected by United States copyright law and international copyright law. No part of this publication may be reproduced, copied, modified, distributed, displayed, stored in a retrieval system, transmitted in any form or by any means, or otherwise published without the prior written permission of Louisiana Healthcare Connections. You may not alter or remove any trademark, copyright or other notice contained herein. Louisiana Healthcare Connections is a registered trademark exclusively owned by Louisiana Healthcare Connections.