

### Clinical Policy: Intestinal and Multivisceral Transplant

Reference Number: PA.CP.MP.58

Effective Date: 01/18

Date of Last Revision: 09/2023

Coding Implications
Revision Log

#### **Description**

This policy describes the medical necessity criteria for the review of intestinal and multivisceral transplant requests.

#### Policy/Criteria

It is the policy of Pennsylvania Health & Wellness<sup>®</sup> (PHW) that any of the intestinal and/or multivisceral transplantation procedures listed in **I** are **medically necessary** for pediatric and adult members/enrollees to restore function in those with irreversible intestinal failure when meeting the criteria in section **II**:

#### I. Transplantation Procedures

- **A.** Isolated *intestinal transplantation is indicated* for members/enrollees who have only isolated intestinal failure and no liver disease.
- **B.** Combined *intestinal and liver transplant is indicated* in those with intestinal failure and end stage liver disease.
- C. Multivisceral transplant is indicated in those with intestinal failure and gastrointestinal motility disorders (e.g., chronic idiopathic intestinal pseudo-obstruction, visceral myopathy, visceral neuropathy, total intestinal aganglionosis, and some forms of mitochondrial respiratory chain disorders that affect gastrointestinal motor function), or extensive mesenteric thrombosis.
- **II. Procedure Criteria:** Members/enrollees must have one of the indications in **A** and none of the contraindications in **B**:
  - **A.** Indications, any one of the following:
    - 1. Failure of total parenteral nutrition (TPN) as indicated by one of the following:
      - a. Impending or overt liver failure due to TPN, indicated by elevated serum bilirubin and/or liver enzymes, splenomegaly, thrombocytopenia, gastro-esophageal varices, coagulopathy, peristomal bleeding, or hepatic fibrosis/cirrhosis;
      - b. Thrombosis of  $\geq$  two central veins, including jugular, subclavian, and femoral veins:
      - c. Two or more episodes of systemic sepsis due to line infection, per year, or one episode of septic shock, acute respiratory distress syndrome, and/or line related fungemia:
      - d. Frequent episodes of dehydration despite IV fluid supplementation;
      - e. Other complications leading to loss of vascular access;
    - 2. High risk of death if transplant is not performed;
    - 3. Severe short bowel syndrome (gastrostomy, duodenostomy, and/or residual small bowel <10 cm in infants and <20 cm in adults);
    - 4. Frequent hospitalizations for complications directly related to intestinal failure;
    - 5. Significant hepatic cirrhosis associated with diffuse post-mesenteric thrombosis;
  - **B.** Does not have ANY of the following contraindications:
    - 1. Malignancy with high risk of recurrence or death related to cancer;

#### **CLINICAL POLICY**

#### **Intestinal and Multivisceral Transplant**



- 2. Other severe uncontrolled medical condition expected to limit survival after transplant;
- 3. Glomerular filtration rate < 40 mL/min/1.73m<sup>2</sup> unless being considered for multi-organ transplant;
- 4. HIV infection with detectable viral load;
- 5. Presence of other GI diseases;
- 6. Acute liver failure, or cirrhosis with portal hypertension or synthetic dysfunction unless being considered for multi-organ transplant;
- 7. Septic shock;
- 8. Progressive cognitive impairment;
- 9. Stroke, acute coronary syndrome, or myocardial infarction (excluding demand ischemia) within 30 days;
- 10. Chronic infection with highly virulent and/or resistant microbes that are poorly controlled pre-transplant;
- 11. Inability to adhere to the regimen necessary to preserve the transplant, even with caregiver support;
- 12. Absence of an adequate or reliable social support system;
- 13. 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 (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.

#### **Background**

Intestinal transplantation is a therapeutic option for patients with intestinal failure. Intestinal failure is the loss of absorptive capacity of the small bowel secondary to severe primary gastrointestinal disease or surgically induced short bowel syndrome (SBS). The normal small intestine length varies widely, ranging from three to eight meters. SBS occurs when there is approximately < 200 cm of small bowel remaining.

Multi-visceral transplantation includes the stomach, duodenum, pancreas, liver, and small intestine. A modified version excludes the liver if the recipient's liver is normal. A kidney transplant is occasionally included if the recipient has end-stage renal disease.<sup>4</sup>

Common indications for intestinal transplantation in children include:

- Small bowel atresia
- Gastroschisis
- Aganglionosis (Hirschsprung's disease)
- Infections such as necrotizing enterocolitis and mesenteric ischemia
- Intestinal pseudo-obstruction

- Microvillus inclusion disease
- Short gut syndrome
- Trauma
- Crohn's disease
- Midgut volvulus
- Massive resection secondary to tumor

Common indications for intestinal transplantation in adults include:

- Short gut syndrome
- Mesenteric ischemia following thrombosis, embolism, volvulus, or trauma
- Crohn's disease

- Small bowel tumors
- Small bowel secretory disorders
- Tumors of mesenteric root and retroperitoneum

# **CLINICAL POLICY Intestinal and Multivisceral Transplant**



- Trauma
- Volvulus

- Pseudo-obstruction
- Radiation enteritis

#### Guideline Recommendations

The British Society of Gastroenterology recommends patients with SBS, including irreversible intestinal failure, expected to die prematurely on TPN, should be referred for consideration of short bowel transplant where appropriate.<sup>13</sup>

The American Society of Transplantation issued a position paper on indications for pediatric intestinal transplantation. The AST recommends intestinal transplantation only for TPN-dependent children with intestinal failure who have or are likely to develop life-threatening TPN-related complications such as liver disease, recurrent sepsis, and threatened loss of central venous access. The AST stated that intestinal transplantation should not be performed solely because of continued dependence on TPN.<sup>8</sup>

#### **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. 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
44135	Intestinal allotransplantation; from cadaver donor
44136	Intestinal allotransplantation; from living donor
44715	Backbench standard preparation of cadaver or living donor intestine allograft prior to transplantation, including mobilization and fashioning of the superior mesenteric artery and vein
44720	Backbench reconstruction of cadaver or living donor intestine allograft prior to transplantation; venous anastomosis, each
44721	Backbench reconstruction of cadaver or living donor intestine allograft prior to transplantation; arterial anastomosis, each
47135	Liver allotransplantation, orthotopic, partial or whole, from cadaver or living donor, any age
47143	Backbench standard preparation of cadaver donor whole liver graft prior to allotransplantation, including cholecystectomy, if necessary, and dissection and removal of surrounding soft tissues to prepare the vena cava, portal vein, hepatic artery, and common bile duct for implantation; without trisegment or lobe split
47144	Backbench standard preparation of cadaver donor whole liver graft prior to allotransplantation, including cholecystectomy, if necessary, and dissection and removal of surrounding soft tissues to prepare the vena cava, portal vein, hepatic artery, and common bile duct for implantation; with trisegment split of whole liver



# **CLINICAL POLICY Intestinal and Multivisceral Transplant**

<b>CPT</b> <sup>®</sup>	Description
Codes	
	graft into two partial liver grafts (i.e., left lateral segment (segments II and III) and right trisegment (segments I and IV through VIII)
47145	Backbench standard preparation of cadaver donor whole liver graft prior to allotransplantation, including cholecystectomy, if necessary, and dissection and removal of surrounding soft tissues to prepare the vena cava, portal vein, hepatic artery, and common bile duct for implantation; with lobe split of whole liver graft into two partial liver grafts (i.e., left lobe (segments II, III, and IV) and right lobe (segments I and V through VIII)
47146	Backbench reconstruction of cadaver or living donor liver graft prior to allotransplantation; venous anastomosis, each
47147	Backbench reconstruction of cadaver or living donor liver graft prior to allotransplantation; arterial anastomosis, each

HCPCS	Description
Codes	
S2053	Transplantation of small intestine and liver allografts
S2054	Transplantation of multivisceral organs
S2055	Harvesting of donor multivisceral organs, with preparation and maintenance of
	allografts; from cadaver donor
S2152	Solid organs(s), complete or segmental, single organ or combination of organs;
	deceased or living donor(s), procurement, transplantation, and related complications;
	including: drugs; supplies; hospitalization with outpatient follow-up;
	medical/surgical, diagnostic, emergency, and rehabilitative services; and the number
	of days of pre- and post-transplant care in the global definition

Reviews, Revisions, and Approvals	Date	Approval
		Date
Policy developed	11/17	1/1/18
Specialist review (Surgical Transplant)		
References reviewed and updated	09/18	10/18
References reviewed and updated. Added CPT-47135	12/19	1/30/2020
Edited malignancy contraindication to not specify within 2 years, and	2/26/2021	
added exceptions early stage prostate cancer, cancer that has been		
completely resected, or that has been treated and poses acceptable		
future risk. Clarified in I.C that multivisceral transplants are		
indicated in gastrointestinal motility disorders, along with examples		
of such. Added ICD 10 Q43.1 References reviewed and updated.		
Specialist reviewed. Annual Review performed.		
Annual Review performed. Replaced contraindications of "severely		
limited functional status with poor rehabilitation potential" and those		
regarding past or current nonadherence to medical therapy, and		
psychological condition associated with the inability to comply with		

# **CLINICAL POLICY Intestinal and Multivisceral Transplant**



Reviews, Revisions, and Approvals	Date	Approval Date
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." Edited contraindications:		
Replaced "non-hepatic malignancy" with malignancy with high		
risk of recurrence or death"; added GFR restriction, added HIV		
infection with detectable viral load, added stroke, acute coronary		
syndrome, or MI; added acute renal failure; 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"; removed		
"uncorrectable bleeding diathesis." Specialist reviewed.		
Annual review. Updated verbiage in II.B.13. to "Active substance	09/2023	
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 (unless		
urgent transplant timelines are present, in which case a commitment		
to reducing behaviors is acceptable)." References reviewed, updated,		
and reformatted. Specialist reviewed.		

#### References

- 1. Buchman AL, Iyer K, Fryer J. Parenteral nutrition-associated liver disease and the role for isolated intestine and intestine/liver transplantation. *Hepatology*. 2006;43(1):9-19. doi:10.1002/hep.20997
- 2. National coverage determination: intestinal and multi-visceral transplantation (260.5). Centers for Medicare and Medicaid Services Web site. <a href="https://www.cms.gov/medicare-coverage-database/search.aspx">https://www.cms.gov/medicare-coverage-database/search.aspx</a>. Published May 11, 2006. Accessed April 12, 2022.
- 3. Khan FA, Selvaggi G. Overview of intestinal and multivisceral transplantation. UpToDate. <a href="https://www.uptodate.com">www.uptodate.com</a>. Published August 17, 2022. Accessed April 5, 2023.
- 4. Gilroy RK. Intestinal and multivisceral transplantation. Medscape. <a href="https://reference.medscape.com/">https://reference.medscape.com/</a>. Published May 24, 2018 (updated February 22, 2021). Accessed April 5, 2023.
- 5. Andacoglu, OM. Intestinal transplantation. Medscape. <a href="https://reference.medscape.com/">https://reference.medscape.com/</a>. Published January 18, 2017 (updated March 16, 2021). Accessed April 5, 2023.
- 6. Kato T, Tzakis AG, Selvaggi G, et al. Intestinal and multivisceral transplantation in children. *Ann Surg.* 2006;243(6):756-766. doi:10.1097/01.sla.0000219696.11261.13
- 7. Kaufman SS, Atkinson JB, Bianchi A, et al. Indications for pediatric intestinal transplantation: a position paper of the American Society of Transplantation. *Pediatr Transplant*. 2001;5(2):80-87. doi:10.1034/j.1399-3046.2001.005002080.x
- 8. Kubal CA, Mangus RS, Tector AJ. Intestine and multivisceral transplantation: current status and future directions. *Curr Gastroenterol Rep.* 2015;17(1):427. doi:10.1007/s11894-014-0427-8

#### **CLINICAL POLICY**

#### **Intestinal and Multivisceral Transplant**



- 9. Levitsky J, Doucette K; AST Infectious Diseases Community of Practice. Viral hepatitis in solid organ transplantation. *Am J Transplant*. 2013;13 Suppl 4:147-168. doi:10.1111/ajt.12108
- 10. Lladó L, Fabregat J, Castellote J, et al. Management of portal vein thrombosis in liver transplantation: influence on morbidity and mortality. *Clin Transplant*. 2007;21(6):716-721. doi:10.1111/j.1399-0012.2007.00728.x
- 11. Mangus RS, Tector AJ, Kubal CA, Fridell JA, Vianna RM. Multivisceral transplantation: expanding indications and improving outcomes. *J Gastrointest Surg*. 2013;17(1):179-p.187. doi:10.1007/s11605-012-2047-7
- 12. Nightingale J, Woodward JM; Small Bowel and Nutrition Committee of the British Society of Gastroenterology. Guidelines for management of patients with a short bowel. *Gut*. 2006;55 Suppl 4(Suppl 4):iv1-iv12. doi:10.1136/gut.2006.091108
- 13. Holzheimer RG, Mannick JA, eds. *Surgical Treatment: Evidence-Based and Problem-Oriented*. Munich: Zuckschwerdt; 2001.
- 14. Tzakis AG, Kato T, Levi DM, et al. 100 multivisceral transplants at a single center. Ann Surg. 2005;242(4):480-493. doi:10.1097/01.sla.0000183347.61361.7a
- 15. U.S. Department of Veteran Affairs. HIV. Laboratory Tests and HIV: Entire Lesson. <a href="https://www.hiv.va.gov/patient/diagnosis/labtests-single-page.asp">https://www.hiv.va.gov/patient/diagnosis/labtests-single-page.asp</a>. Accessed April 7, 2023.
- 16. Vianna RM, Mangus RS, Kubal C, Fridell JA, Beduschi T, Tector AJ. Multivisceral transplantation for diffuse portomesenteric thrombosis. Ann Surg. 2012;255(6):1144-1150. doi:10.1097/SLA.0b013e31825429c0
- 17. Wu G, Cruz RJ. Liver inclusion improves outcomes of intestinal retransplantation in adults. [Corrected] [published correction appears in Transplantation. 2015 Aug;99(8):e118]. *Transplantation*. 2015;99(6):1265-1272. doi:10.1097/TP.0000000000000488
- 18. Bharadwaj S, Tandon P, Gohel TD, et al. Current status of intestinal and multivisceral transplantation. *Gastroenterol Rep (Oxf)*. 2017;5(1):20-28. doi:10.1093/gastro/gow045
- 19. Pironi L, Arends J, Bozzetti F, et al. ESPEN guidelines on chronic intestinal failure in adults [published correction appears in Clin Nutr. 2017 Apr;36(2):619]. *Clin Nutr.* 2016;35(2):247-307. doi:10.1016/j.clnu.2016.01.020
- 20. Hawksworth JS, Desai CS, Khan KM, et al. Visceral transplantation in patients with intestinal-failure associated liver disease: Evolving indications, graft selection, and outcomes. *Am J Transplant*. 2018;18(6):1312-1320. doi:10.1111/ajt.14715
- 21. Camilleri M. Chronic intestinal pseudo-obstruction: Etiology, clinical manifestations, and diagnosis. UpToDate. <a href="www.uptodate.com">www.uptodate.com</a>. Published February 17, 2022. Accessed April 7, 2023.