



# Medical Coverage Policy

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## Intestinal and Multivisceral Transplantation

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#### INSTRUCTIONS FOR USE

The following Coverage Policy applies to health benefit plans administered by Cigna Companies. Certain Cigna Companies and/or lines of business only provide utilization review services to clients and do not make coverage determinations. References to standard benefit plan language and coverage determinations do not apply to those clients. Coverage Policies are intended to provide guidance in interpreting certain standard benefit plans administered by Cigna Companies. Please note, the terms of a customer's particular benefit plan document [Group Service Agreement, Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer's benefit plan document always supersedes the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Each coverage request should be reviewed on its own merits. Medical directors are expected to exercise clinical judgment and have discretion in making individual coverage determinations. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations.

### Overview

This Coverage Policy addresses primary and repeat deceased donor intestinal and multivisceral organ transplantation. Contraindications for transplantation are also addressed.

### Coverage Policy

**Primary and repeat deceased donor intestinal transplantation and primary and repeat deceased donor multivisceral organ transplantation are considered medically necessary in an individual with total irreversible intestinal failure for ANY of the following indications:**

- failure, contraindication, or intolerance to parenteral nutrition with ANY of the following:
  - impending or overt liver failure
  - impending loss of central vein access (e.g., thrombosis)
  - recurrent, life-threatening sepsis
  - frequent episodes of dehydration
- high risk of death
- severe short bowel syndrome

- frequent hospitalizations for complications directly related to intestinal failure
- pseudo-obstruction

**Deceased donor intestinal or multivisceral transplantation are considered not medically necessary when ANY of the following absolute contraindications to transplantation exist (this list may not be all-inclusive):**

- malignancy that is expected to significantly limit future survival
- unsuccessfully treated major or systemic infections
- systemic illness or comorbidities that would be expected to substantially negatively impact the successful completion and/or outcome of transplant surgery
- a pattern of demonstrated patient noncompliance which would place a transplanted organ at serious risk of failure
- human immunodeficiency virus (HIV) disease unless **ALL** of the following are noted:
  - CD4 count greater than 200 cells/mm<sup>3</sup>
  - HIV-1 ribonucleic acid (RNA) undetectable
  - stable anti-retroviral therapy for more than three months
  - absence of serious complications associated with or secondary to HIV disease (e.g., opportunistic infection, including aspergillus, tuberculosis, coccidioidomycosis; resistant fungal infections; or Kaposi's sarcoma or other neoplasm)

## General Background

Intestinal (i.e., small bowel only), liver-intestinal, or multivisceral transplantation are accepted therapeutic options for highly selected adults with irreversible intestinal and/or multivisceral organ failure who have failure, contraindication, or intolerance to total parenteral nutrition (TPN). Irreversible gastrointestinal system failure is defined as the inability to maintain nutrition or adequate fluid and electrolyte balance without special support, when currently available medical and surgical treatments fail to improve intestinal adaptation and gut function. Causes may differ among children and adults (Reyes, 2020; Matarese, 2007). Although TPN is the standard of care for patients with temporary or permanent intestinal failure, it severely affects quality of life and may be associated with a number of highly morbid and sometimes fatal complications and has a treatment-related mortality of 7–28% (Markman, 2012). Transplantation should be considered once it has been clearly shown that the bowel cannot adapt to allow full enteral autonomy from parenteral nutrition (Braun, 2007). Additional life-threatening indications include impending or overt liver failure, loss or impending loss of central venous access (e.g., thrombosis), recurrent, systemic sepsis, frequent episodes of dehydration, high risk of death, severe short bowel syndrome, frequent hospitalization, or pseudo-obstruction (Reyes, 2020; Avitzur, 2010; Centers for Medicare and Medicaid Services [CMS], 2006; American Gastroenterological Association [AGA], 2003, Kaufman, 2001).

Specific indications for intestinal and multivisceral transplantation may include the following (Reyes, 2020; Matsumoto, 2018; Bharadwaj, 2017; Lauro, 2007; Matarese, 2007; Sudan, 2007; Reyes, 2006; Carter, 2006; Dove and Brown, 2004; AGA, 2003; Abu-Elmagd, 2001):

### Common Indications for Intestinal and Multivisceral Transplantation

Children	Adults
Aganglioneosis (Hirschsprung's disease)	Autoimmune enteritis
Autoimmune enteropathy	Crohn's disease
Congenital epithelial mucosal disease (microvillus inclusion disease, tufting enteropathy)	Desmoid tumors
Crohn's disease	Gardner's syndrome/familial polyposis
Familial polyposis	Hollow visceral myopathy
Gastroschisis	Inflammatory bowel disease
Inflammatory pseudotumor	Ischemia
	Radiation enteritis

Intestinal atresia Intestinal failure-associated liver disease Intestinal pseudo-obstruction Microvillus inclusion disease Necrotizing enterocolitis Pseudo-obstruction Radiation enteritis Short gut syndrome Tufting enteropathy Trauma Volvulus	Secretory diarrhea Short gut syndrome Surgical adhesions Trauma Vascular occlusion Volvulus
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Intestinal and multivisceral transplantations are more challenging than other types of solid organ transplantation due to the intestine's large number of immune competent cells and colonization of the gut with microorganisms. Intestinal allografts may be transplanted alone, as in an isolated intestine graft, or as a composite graft which may include the liver, duodenum, and pancreas. If the recipient operation includes replacement of the entire gastrointestinal graft and liver, it is generally referred to as a multivisceral transplantation (Reyes, 2020). The type and number of transplanted organs is dictated by the extent of the abdominal pathology and the functional status of the organs at the time of transplantation (Matsumoto, 2018; Bharadwaj, 2017; Matarese, 2007; Abu-Elmagd, 2006).

Isolated intestine transplantation may be indicated when there is a permanent need for total parenteral nutrition (TPN) after failure of intestinal adaptation and failed attempts of medical and surgical rehabilitation (Matarese, 2007). Combined liver-intestinal transplantation may be appropriate for individuals with combined intestinal and TPN-associated liver failure, liver failure associated with portal and mesenteric venous thrombosis, intestinal failure due to a hypercoagulable state associated with enzyme deficiencies that can be corrected by a liver graft (e.g., mesenteric vascular thrombosis secondary to protein C or S deficiency), or documented end-stage hepatic disease. In adults, such disease is associated with refractory ascites, spontaneous bacterial peritonitis, refractory variceal bleeding, chronic encephalopathy, hepatorenal syndrome, failure to thrive, or a severe compromise in the quality of life (Abu-Elmagd, 2001). In children, end-stage hepatic disease is suggested by hyperbilirubinemia persisting beyond three to four months of age, combined with features of portal hypertension, such as splenomegaly, thrombocytopenia, or prominent superficial abdominal veins.

A full multivisceral transplantation involves the en bloc transplantation of the stomach, liver, duodenum and pancreas with the intestine. In a modified procedure only one or two organs may be transplanted. It is indicated for patients with irreversible failure of their abdominal visceral organs, including the small bowel. The aims of multivisceral transplantation are to replace as many functional digestive units as possible, restore gastric emptying, ileocecal valve function, rectal continence, and improvement of surgical and oncological margins of resection (Braun, 2007). Conditions include symptomatic extensive thrombosis of the splanchnic vascular system, massive gastrointestinal polyposis or neoplasm, and generalized hollow visceral myopathy or neuropathy (Abu-Elmagd, 2001). Multivisceral transplantation may also be indicated for diffuse gastrointestinal disorders such as dysmotility syndromes, hereditary neoplasms, and extensive vascular thrombosis (Matarese, 2007).

### **Contraindications to Intestinal and Multivisceral Transplantation**

Contraindications for intestinal and multivisceral transplantation are similar to those for other types of solid organ transplantation. Absolute contraindications include severe uncontrolled infection, multiorgan failure, nonresectable or disseminated malignancy, significant cardiopulmonary insufficiency, acquired immunodeficiency syndrome, the existence of life-threatening uncontrollable intra-abdominal or systemic infections, and noncompliance (Braun, 2007; Matarese, 2007; Abu-Elmagd, 2001; Kaufman, 2001). In addition to the absolute contraindications noted, relative contraindications which may also negatively affect survival, may include, but not be limited to (Kaufman, 2007; Abu-Elmagd, 2001):

- current, ongoing substance abuse, including tobacco, alcohol and narcotic/other addictive pain medications

- profound neurologic disabilities
- severe congenital or acquired immunological deficiencies
- multisystem autoimmune diseases
- progressive neuropathy or myopathy that is not amenable to rehabilitation
- any active medical process that is currently not optimally treated and/or stable and that is likely to result in end-organ damage or medical emergency without appropriate management, such as active peptic ulcer disease, diverticular disease, active hepatitis, cholecystitis, pancreatitis, diabetes mellitus, hypertension, autoimmune disease, or cytopenia
- advanced age
- positive cross-match

### **Deceased (Cadaver) Donor Intestinal and Multivisceral Transplantation**

Intestinal and multivisceral transplantations most frequently involve the use of cadaveric, or deceased, donors. Although randomized clinical trial data are not available, there are several case reports and retrospective studies demonstrating improved outcomes. Demographic characteristics of candidates on the intestine only transplant waiting list show that 82.4% are under 17 years old, 52.9% are female, with a race/ethnicity breakdown of 58.8% white, 21.3% Black, 14% Hispanic and 2.9% Asian. For candidates waiting for intestine and liver transplant, the age and race/ethnicity demographic characteristics are similar, however 62.2% are males. The demographic characteristics of intestine transplant recipients reported 26.8% are <17 years old, 61% are female, with a race/ethnicity breakdown of 63.4% white, 24.4% Black, 9.8% Hispanic and 2.4% Asian (United States Organ Procurement and Transplantation Network and the Scientific Registry of Transplant Recipients [OPTN/SRTR], 2021). According to national data from the OPTN/SRTR, one-, three-, and five-year patient survival outcomes for individuals undergoing initial or primary deceased donor intestinal transplantation from 2008-2015 (as of Oct 22, 2021) are 82.8%, 68.9%, and 58.9%, respectively. Neither graft nor patient survival data are available for multivisceral transplantation from the OPTN.

### **Literature Review**

Abu-Elmagd et al. (2009) reported results of a retrospective review of 453 patients who received 500 visceral transplants at a single transplant facility. Of 453 recipients, 198 (44%) received intestine, 142 (31%) received combined liver-intestine, and 113 (25%) received multivisceral grafts: 84 with liver (full) and 29 without liver (modified). Actuarial patient survival was 85% at 1-year, 61% at 5-years, 42% at 10-years, and 35% at 15-years with respective graft survival of 80%, 50%, 33%, and 29%. With a 10% retransplantation rate, second/third graft survival was 69% at 1-year and 47% at 5-years. Although limited by uncontrolled study design, long-term survival data suggest acceptable overall survival rates for primary and repeat intestinal and multivisceral transplantation.

### **Retransplantation**

The overall effect of intestinal and multivisceral retransplantation in the United States has not been evaluated in a comprehensive manner and there are limited data in the published, peer-reviewed medical literature. Causes of graft loss may include acute cellular rejection, chronic rejection, post transplant lymphoproliferative disorder, graft dysmotility or dysfunction, severe infection, arterial graft aneurysm, or allograft liver failure (Mazariegos, 2008). Careful patient selection, post transplant immunosuppression, and patient management are essential for successful long-term outcomes (Mazariegos, 2008). Based on Organ Procurement Transplant Network and the Scientific Registry of Transplant Recipients (OPTN/SRTR) data for repeat intestine transplants performed between 2008-2015 Kaplan-Meier one-, three-, and five-year patient survival rates (as of Oct 22, 2021) were 70.2%, 53.9% and 49.8%, respectively. Individuals undergoing repeat intestinal or multivisceral transplantation should meet all of the eligibility criteria for primary transplantation and should not have absolute contraindications to transplantation.

### **Literature Review**

Desai et al. (2012) performed an analysis of United Network of Organ Sharing (UNOS) registry data relative to outcomes for intestine retransplantation performed in children and adults from 1987-2009. Of 1822 isolated intestine transplants (ITx) in 1664 patients during the study period, 149 patients (adults, n=72; children, n=77) received repeat transplantation. Nine of these were third transplants, all in children. Of 41 adult isolated ITx, patient survival was 80.1%, 47.4%, and 28.5% at 1, 3, and 5 years, respectively, which is lower than outcomes

seen with primary isolated ITx (p=0.005). For combined liver/ITx retransplantation in adults (L-ITx, n=31), patient survival at one-, three-, and five-years was 63.1%, 56.1%, and 46.8%, respectively, compared with primary L-ITx retransplantation (p=0.07). Isolated ITx retransplantation in children (n=28) resulted in patient survival of 80.7%, 74%, and 57.5% at one-, three-, and five-years, respectively. One-, three-, and five-year patient survival in children receiving L-ITX was 42%, 42%, and 42%, respectively. Although data suggests lower survival rates for retransplantation compared to primary transplantation, outcomes are acceptable in this population of individuals for which alternative treatment options are limited.

**Professional Societies/Organizations**

**American Society of Transplantation (AST):** On behalf of the AST, Kaufman (2001) published guidelines regarding the indications for pediatric intestinal transplantation. These include progressive parenteral nutrition-associated liver disease, recurring sepsis, impending loss of central venous access, extreme short-bowel syndrome, and congenital intractable epithelial (mucosal) disorders. The Society notes that intestinal transplantation is a lifesaving therapy for the child with intestinal failure. Transplantation should be considered when intestinal failure has been, or will probably become, refractory to conventional management, the mainstay of which remains parenteral nutrition therapy.

**American Gastroenterological Association (AGA):** The AGA’s medical position statement: Short Bowel Syndrome and Intestinal Transplantation (2003) notes the following indications for intestinal transplantation:

- impending or overt liver failure
- thrombosis of major central venous channels
- frequent central line-related sepsis
- frequent severe dehydration

**Use Outside of the US**

No relevant information.

**Medicare Coverage Determinations**

	Contractor	Policy Name/Number	Revision Effective Date
NCD	National	Intestinal and Multi-Visceral Transplantation (260.5)	6/26/2006
LCD		No Local Coverage Determination found	

Note: Please review the current Medicare Policy for the most up-to-date information.

**Coding/Billing Information**

**Note:** 1) This list of codes may not be all-inclusive.  
 2) Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

**Considered medically necessary when used to report primary and repeat deceased donor intestinal transplantation and primary and repeat deceased donor multivisceral organ transplantation:**

CPT®* Codes	Description
44132	Donor enterectomy (including cold preservation), open; from cadaver donor
44135	Intestinal allotransplantation; from cadaver donor
44137	Removal of transplanted intestinal allograft, complete
44140	Colectomy, partial; with anastomosis
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

<b>CPT®*</b> <b>Codes</b>	<b>Description</b>
44721	Backbench reconstruction of cadaver or living donor intestine allograft prior to transplantation; arterial anastomosis, each
47133	Donor hepatectomy (including cold preservation), from cadaver donor
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 graft into 2 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 2 partial liver grafts (i.e., left lobe (segments II, III, and IV) and right lobe (segments I, 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
47399†	Unlisted procedure, liver
48550	Donor pancreatectomy (including cold preservation), with or without duodenal segment for transplantation
48551	Backbench standard preparation of cadaver donor pancreas allograft prior to transplantation, including dissection of allograft from surrounding soft tissues, splenectomy, duodenotomy, ligation of bile duct, ligation of mesenteric vessels, and Y-graft arterial anastomoses from iliac artery to superior mesenteric artery and to splenic artery
48552	Backbench reconstruction of cadaver donor pancreas allograft prior to transplantation, venous anastomosis, each
48554	Transplantation of pancreatic allograft
48556	Removal of transplanted pancreatic allograft

†**Note:** When used to represent liver allotransplantation; heterotopic, partial or whole, from cadaver donor, any age.

<b>HCPCS</b> <b>Codes</b>	<b>Description</b>
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 organ(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

\***Current Procedural Terminology (CPT®) ©2020 American Medical Association: Chicago, IL.**

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