Medical Policy

BlueCross BlueShield of Kansas
An Independent Licensee of the Blue Cross and Blue Shield Association

Topic: Small Bowel/Liver and Multivisceral Transplant

Professional
Original Effective Date: January 11, 2005
Revision Date(s): February 25, 2010; February 10, 2011; July 19, 2011; November 19, 2012; February 28, 2014
Current Effective Date: February 10, 2011

Institutional
Original Effective Date: February 25, 2010
Revision Date(s): February 10, 2011; July 19, 2011; November 19, 2012; February 28, 2014
Current Effective Date: February 10, 2011

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DESCRIPTION

A multivisceral transplant is transplantation of an intestinal allograft in combination with a liver allograft, either alone or in combination with one or more of the following organs: stomach, duodenum, jejunum, ileum, pancreas, or colon.

Background

Small bowel transplants are typically performed in patients with short bowel syndrome, defined as an inadequate absorbing surface of the small intestine due to extensive disease or surgical removal of a large portion of small intestine. In some instances, short bowel syndrome is associated with liver failure, often due to the long-term complications of total parenteral nutrition (TPN). These patients may be candidates for a small bowel/liver transplant or a multivisceral transplant, which includes the small bowel and

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liver with 1 or more of the following organs: stomach, duodenum, jejunum, ileum, pancreas, and/or colon. A multivisceral transplant is indicated when anatomic or other medical problems preclude a small bowel/liver transplant.

POLICY

A small bowel/liver transplant or multivisceral transplant may be considered **medically necessary** for pediatric and adult patients with:

1. intestinal failure (characterized by loss of absorption and the inability to maintain protein-energy, fluid, electrolyte, or micronutrient balance), or
2. other end stage organ failure

Policy Guidelines

General

1. Potential contraindications to solid organ transplant (subject to the judgment of the transplant center):
    a. Known current malignancy, including metastatic cancer
    b. Recent malignancy with high risk of recurrence
    c. History of cancer with a moderate risk of recurrence
    d. Systemic disease that could be exacerbated by immunosuppression
    e. Untreated systemic infection making immunosuppression unsafe, including chronic infection
    f. Other irreversible end-stage disease not attributed to organ failure
    g. Psychosocial conditions or chemical dependency affecting ability to adhere to therapy

2. Intestinal failure results from surgical resection, congenital defect, or disease-associated loss of absorption and is characterized by the inability to maintain protein-energy, fluid, electrolyte, or micronutrient balance (adapted from reference 1). (1) Short bowel syndrome is one case of intestinal failure.

3. Candidates should meet the following criteria:
    a. Adequate cardiopulmonary status
    b. Documentation of patient compliance with medical management.

4. HIV [human immunodeficiency virus]-positive patients who meet the following criteria, as stated in the 2001 guidelines of the American Society of Transplantation, could be considered candidates for small bowel/liver or multivisceral transplantation:
    a. CD4 count greater than 200 cells per cubic millimeter for greater than 6 months
    b. HIV-1 RNA undetectable
    c. On stable anti-retroviral therapy >3 months

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d. No other complications from AIDS [acquired immune deficiency syndrome] (e.g., opportunistic infection, including aspergillus, tuberculosis, coccidiosis mycosis, resistant fungal infections, Kaposi's sarcoma, or other neoplasm), and meeting all other criteria for transplantation.

Small Bowel/Liver Specific

Evidence of intolerance of total parenteral nutrition (TPN) includes, but is not limited to, multiple and prolonged hospitalizations to treat TPN-related complications, or the development of progressive but reversible liver failure. In the setting of progressive liver failure, small bowel transplant may be considered a technique to avoid end-stage liver failure related to chronic TPN, thus avoiding the necessity of a multivisceral transplant.

Definition

Regarding multivisceral transplants, a visceral organ is defined as any organ within the chest or abdomen. Stedman's states that visceral organ is from the viscera, the plural of viscus. The viscus is, "an organ of the digestive, respiratory, urogenital, and endocrine system as well as the spleen, the heart, and great vessels; hollow and multilayered, walled organs studied in splanchnology." [Latin: the soft parts, internal organs] (Medical Director decision of 08/07/07.)

RATIONALE

The most recent literature search was performed for the period April 2011 through April 2012.

A 1999 TEC Assessment focused on multivisceral transplantation and offered the following conclusions:
Multivisceral transplantation in patients with small bowel syndrome, liver failure, and/or other gastrointestinal problems such as pancreatic failure, thromboses of the celiac axis and the superior mesenteric artery, or pseudo-obstruction affecting the entire gastrointestinal tract associated with poor patient and graft survival. Pediatric and adult patients have a similar 2- and 5-year survival of 33–50%. However, without this procedure, it is expected that these patients would face 100% mortality. (2)

The published literature consists of case series, mainly reported by single centers. Authors of these reports, as well as reviews, observe that while outcomes continue to improve, recurrent and chronic rejection and complications of immunosuppression continue to be obstacles to long-term survival.

In 2010, Nayyar and colleagues reported that there had been improvements in 5-year actuarial patient and graft survival after liver/small bowel transplant since the use of rabbit antithymocyte globulin (rATG) induction began to be used in their pediatric center in 2002 (81% vs. 58% and 76% vs. 52%, respectively). (3) In addition to innovations in immunosuppressive therapy, the authors cited new approaches to management of short gut syndrome including hypoallergenic formulas and modification of enteral nutrition to prevent total parenteral nutrition (TPN)-induced cholestasis. The authors noted that better understanding of the protective role of the liver in

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preventing chronic rejection of the small bowel allograft could improve long-term survival after isolated small bowel transplantation.

Abu-Elmgagd et al., reporting on experience with 500 intestinal and multivisceral transplantations, also found that the best outcomes in their series were in the intestine-liver allografts. (4) The study found 1- and 5-year patient survival of 92% and 70%, respectively.

Several recent series have focused on complications after small bowel and multivisceral transplantation. For example, in 2011 Wu and colleagues reported on 241 patients who underwent intestinal transplantation. (5) Of these, 147 (61%) had multivisceral transplants, 65 (27%) had small bowel transplants, and 12% had small bowel/liver transplants. There were 151 children (63%) and 90 adults. A total of 22 patients (9%) developed graft-versus-host disease (GVHD). Children younger than 5 years-old were more likely to develop GVHD; the incidence in this age group was 16 of 121 (13.2%) compared to 2 of 30 (6.7%) in children between 5 and 18 years and 9 of 90 (4.4%) in adults older than 18 years. In addition, a 2012 article retrospectively reported on bloodstream infections among 98 children younger than age 18 years with small bowel/combined organ transplants. (6) Seventy-seven (79%) patients underwent small bowel transplant in combination with a liver, kidney or kidney-pancreas, and 21 had an isolated small bowel transplant. After a median follow-up of 52 months, 58 (59%) patients remained alive. The 1-year survival rate was similar in patients with combined small bowel transplant (75%) and those with isolated small bowel transplant (81%). In the first year after transplantation, 68 patients (69.4%) experienced at least one episode of bloodstream infection. The 1-year survival rate for patients with bloodstream infections was 72% compared to 87% in patients without bloodstream infections (p-value= 0.056 for difference in survival in patients with and without bloodstream infections).

HIV-Positive Transplant Recipients

This subgroup of recipients has long been controversial, due to the long-term prognosis for HIV positivity and the impact of immunosuppression on HIV disease. Although HIV-positive transplant recipients may be a research interest of some transplant centers, the minimal data regarding long-term outcome in these patients primarily consist of case reports and abstract presentations of liver and kidney recipients. Nevertheless, some transplant surgeons would argue that HIV positivity is no longer an absolute contraindication to transplant due to the advent of highly active antiretroviral therapy (HAART), which has markedly changed the natural history of the disease. In 2001, the Clinical Practice Committee of the American Society of Transplantation proposed that the presence of AIDS could be considered a contraindication to kidney transplant unless the following criteria were present. (7) These criteria may be extrapolated to other organs:

- CD4 count greater than 200 cells/mm-3 for more than 6 months
- HIV-1 RNA undetectable
- On stable antiretroviral therapy for more than 3 months
- No other complications from AIDS (e.g., opportunistic infection, including aspergillus, tuberculosis, coccidioses mycosis, resistant fungal infections, Kaposi's sarcoma, or other neoplasm).
- Meeting all other criteria for transplantation.

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In 2006, the British HIV Association and the British Transplantation Society Standards Committee published guidelines for kidney transplantation in patients with HIV disease. (8) As described above, these criteria may be extrapolated to other organs.

The guidelines, which are similar to those cited here, recommend that any patient with end-stage organ disease with a life expectancy of at least 5 years is considered appropriate for transplantation under the following conditions:

- CD4 greater than 200 cells/mL for at least 6 months
- Undetectable HIV viremia (<50 HIV-1 RNA copies/mL) for at least 6 months
- Demonstrable adherence and a stable HAART regimen for at least 6 months
- Absence of AIDS-defining illness following successful immune reconstitution after HAART.

Furthermore, as of November 2010, the United Network for Organ Sharing (UNOS) policy on identification of transmissible diseases in organ recipients states, "A potential candidate for organ transplantation whose test for HIV is positive should not be excluded from candidacy for organ transplantation unless there is a documented contraindication to transplantation based on local policy." (9)

No studies that reported on outcomes in HIV-positive patients who received small bowel/liver or multivisceral transplants have been identified in literature searches.

Summary

Evidence for small bowel/liver and multivisceral transplant consists of case series. Though infrequently performed, the procedures are demonstrated to provide a survival benefit, and the procedure is considered medically necessary for patients who have been managed with long-term total parenteral nutrition and who have developed evidence of impending end-stage liver failure.

CODING

The following codes for treatment and procedures applicable to this policy are included below for informational purposes. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage of these services as it applies to an individual member.

CPT/HCPCS

44120	Enterectomy, resection of small intestine; single resection and anastomosis
44121	Enterectomy, resection of small intestine; each additional resection and anastomosis (List separately in addition to code for primary procedure)
44132	Donor enterectomy (including cold preservation), open; from cadaver donor
44133	Donor enterectomy (including cold preservation), open; partial, 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

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44721 Backbench reconstruction of cadaver or living donor intestine allograft prior to transplantation; arterial anastomosis, each

44799 Unlisted procedure, intestine

47133 Donor hepatectomy (including cold preservation), from cadaver donor

47135 Liver allotransplantation, orthotopic, partial or whole, from cadaver or living donor, any age

47136 Liver allotransplantation, heterotopic, partial or whole, from cadaver or living donor; any age

47140 Donor hepatectomy (including cold preservation), from living donor; left lateral segment only (segments II and III)

47141 Donor hepatectomy (including cold preservation), from living donor; total left lobectomy (segments II, III, or IV)

47142 Donor hepatectomy (including cold preservation), from living donor; total right lobectomy (segments V, VI, VII, and VIII)

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 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

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

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ICD-9 Diagnoses

570	Acute and subacute necrosis of liver
572.8	Other sequelae of chronic liver disease
579.3	Other and unspecified postsurgical nonabsorption

ICD-10 Diagnoses (Effective October 1, 2014

K72.00	Acute and subacute hepatic failure without coma
K72.01	Acute and subacute hepatic failure with coma
K76.2	Central hemorrhagic necrosis of liver
K72.10	Chronic hepatic failure without coma
K72.11	Chronic hepatic failure with coma
K72.90	Hepatic failure, unspecified without coma
K72.91	Hepatic failure, unspecified with coma
K91.2	Postsurgical malabsorption, not elsewhere classified

REVISIONS

01-26-2010	Policy added to the bcbsks.com web site.
02-25-2010	In Definition Section: Updated definition of multivisceral transplant to: "...multivisceral transplant, which includes the small bowel and liver with 1 or more of the following organs: stomach, duodenum, jejunum, ileum, pancreas, and/or colon."
02-10-2011	Description section updated
	In Policy section: ▪ Revised wording of “evidence of impending end stage liver failure” to “other end stage organ failure” ▪ Removed criteria requiring “long term management with total parenteral nutrition (TPN)” ▪ Removed Policy Guidelines
	Rationale section updated
	References section updated
07-19-2011	Rationale section updated
	In Coding section: Updated wording for CPT Codes: 44121, 44721, 47135, 47136, 47141, 47142, 47144, 47145, 47147
	References section updated
11-19-2012	Description section updated
	In Policy section: ▪ Created a Policy Guideline section ▪ Moved multivisceral definition from Description section to Policy Guideline section
	Rationale section updated
	In Coding section: ▪ Added CPT codes 44132, 44133 ▪ Updated Diagnosis nomenclature
	References updated
02-28-2014	In Coding section: ▪ ICD-10 Diagnoses codes added

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3. Nayyar N, Mazariegos G, Ranganathan S et al. Pediatric small bowel transplantation. Semin Pediatr Surg 2010; 19(1):68-77.
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6. Florescu DF, Qiu F, Langnas AN et al. Bloodstream Infections during the First Year after Pediatric Small Bowel Transplantation. Pediatr Infect Dis J 2012; In Press.
7. Steinman TI, Becker BN, Frost AE et al. Guidelines for the referral and management of patients eligible for solid organ transplantation. Transplantation 2001; 71(9):1189-204.
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9. United Network for Organ Sharing. Identification of transmissible diseases in organ recipients. Available online at: optn.transplant.hrsa.gov/PoliciesandBylaws2/policies/pdfs/policy_16.pdf. Last accessed April, 2012.
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Other References

1. Medical Director e-mail of August 7, 2007 on Small Bowel / Liver and Multivisceral Transplant definition.

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