POLICY AND PROCEDURE MANUAL

Policy Title: Small Intestine, Small intestine with Liver or Multivisceral Transplant	**Policy Number: F.16**
Primary Department: Medical Management **Affiliated Department(s): N/A**	**NCQA Standard: N/A** **URAC Standard: N/A**
Last Revision Date: 06/2019 **Revision Dates: 09/2011; 05/2012; 05/2014; 04/2015; 06/2016; 04/2017; 04/2018; 06/2019** **Effective Date: 06/24/2011**	**Next Review Date: 07/2020** **Review Dates: 06/24/2011; 09/23/2011; 07/13/2012; 06/28/2013; 06/27/2014; 06/26/2015; 06/24/2016; 06/28/2017; 06/27/2018; 6/26/2019**
Applicable Lines of Business: ☐MeridianCare ☒MeridianHealth ☐MeridianComplete ☒MeridianChoice	
Applicable States: ☐All ☒MI ☒IL ☐OH ☐_____ ☐_____	
Applicable Programs: ☒All ☐Other _____	
Policy is to be published: Internally Only ☐ Internally & Externally ☒	

Definitions:

Isolated Intestinal Transplantation	Indicated for members who have only isolated intestinal failure and no liver disease. 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 o small intestine.
Combined Intestinal and Liver Transplant	Indicated in those with intestinal failure and end stage liver disease. Combined small bowel/liver transplantation is transplantation of an intestinal allograft in combination with a liver allograft, either alone or in combination with 1 or more of the following organs: stomach, duodenum, jejunum, ileum, pancreas, or colon.
Multivisceral Transplant	Indicated in those with intestinal failure and the presence of neuropathy or extensive mesenteric thrombosis.

If this request is for a Medicare or MMP member please see NCD 260.5

Policy: Meridian Health Plan (MHP) covers intestinal transplantation for the purpose of restoring intestinal function in patients with irreversible intestinal failure. Intestinal failure is defined as the loss of absorptive capacity of the small bowel secondary to severe primary gastrointestinal disease or surgically induced short bowel syndrome. Short bowel syndrome (SBS) occurs after surgery or congenitally when the patient is left with < 200 cm of functional small intestine. It may be associated with both mortality and profound morbidity. Adaptation following disease or injury that leads to intestinal failure can occur over many months up to a year or more. The ability of the remaining gut to adapt to be able to support the patient with enteral nutrition alone is determined by a number of factors including the length of the remaining intestine, the segments

remaining, the presence of an ileocecal valve, the presence or absence of the colon and general motility patterns. A number of medical and surgical interventions are possible to help many of these patients avoid transplant.

Restoration of intestinal continuity, such as reanastomosis of small intestine with colon, should be performed whenever possible, because it can be performed with relatively low morbidity and mortality (often with discontinuation of TPN). Other forms of bowel lengthening surgery have significant associated morbidity and mortality, and therefore should be considered only in select patients. The evidence supports the fact that aged patients generally do not survive as well as younger patients receiving intestinal transplantation. Nonetheless, some older patients who are free from other contraindications have received the procedure and are progressing well, as evidenced by the United Network for Organ Sharing (UNOS) data.

It is essential to determine if associated liver pathology exists in patients being evaluated for potential intestinal transplantation; etiologies other than TPN or malabsorption should be considered. Hepatic aminotransferases, total bilirubin, albumin, international normalized ratio, and platelet count should be determined, and liver biopsy should be performed. Portal venous pressure should be measured to exclude portal hypertension, although normal results may be deceiving in patients who have had major intestinal resections, since most portal inflow will be missing. Patients and their families should meet with a social worker and psychiatrist who understand the complex medical, psychological, and social issues involved with organ transplantation. Living donation should be considered to eliminate waiting time, optimize HLA matching, and simplify coordination of donor-recipient procedures.

Procedure:
Criteria for Coverage:
This procedure is covered only when performed for patients who have irreversible intestinal failure, have failed total parenteral nutrition (TPN) and only when performed in centers that meet approval criteria.

1. Failed TPN: TPN failure includes the following:
    a. Impending or overt liver failure due to TPN induced liver injury. The clinical manifestations include elevated serum bilirubin and/or liver enzymes, splenomegaly, thrombocytopenia, gastroesophageal varices, coagulopathy, stomal bleeding or hepatic fibrosis/cirrhosis.
    b. Thrombosis of the major central venous channels; jugular, subclavian, and femoral veins. Thrombosis of two or more of these vessels is considered a life threatening complication and failure of TPN therapy. The sequelae of central venous thrombosis are lack of access for TPN infusion, fatal sepsis due to infected thrombi, pulmonary embolism, Superior Vena Cava syndrome, or chronic venous insufficiency.
    c. Frequent line infection and sepsis. The development of two or more episodes of systemic sepsis secondary to line infection per year that requires hospitalization indicates failure of TPN therapy. A single episode of line related fungemia, septic shock and/or Acute Respiratory Distress Syndrome are considered indicators of TPN failure.
    d. Frequent episodes of severe dehydration despite intravenous fluid supplement in addition to TPN. Under certain medical conditions such as secretory diarrhea and non-constructable gastrointestinal tract, the loss of the gastrointestinal and pancreatobiliary secretions exceeds the maximum intravenous infusion rates that can be tolerated by the cardiopulmonary system. Frequent episodes of dehydration are deleterious to all body organs particularly kidneys and the central nervous system with the development of multiple kidney stones, renal failure, and permanent brain damage.

Required Documentation:

1. Transplant team evaluation recommending listing for transplant, documenting indications and contraindications, if any.
2. Psychological evaluation to assess behavioral or psychiatric disorders likely to compromise adherence to strict medical regimens and post-transplant follow-up.
3. Social work evaluation to confirm adequate family/social support post-transplant.
4. Blood group and HLA typing.
5. CBC, LFT's, Renal profile, serum creatinine <2.5 mg/dl (<1.5 mg/dl in children) or GFR.35 ml/min, coagulation profile.

6. Serologic testing: CMV, EBV, Hepatitis (A,B, and C,) HIV
7. Radiologic evaluation of the bowel to determine bowel length and function.
8. Duplex Doppler sonography of intraabdominal vascular system (abdominal aorta, superior mesenteric artery, portal vein, superior mesenteric vein).
9. Nutritional consult to confirm TPN failure.
10. If liver transplant requested, then liver biopsy should be performed.
11. Documentation of blood or urine screening for alcohol, tobacco, and illicit drug use. (See Compliance Section)
12. Cardiac evaluation with assessment of cardiac risk factors in all patients over 40 years of age and for those younger than 40 with multiple risk factors for coronary artery disease. This must include EKG and echocardiogram as an initial screening test and stress test and/or cardiac catheterization where clinically indicated.
13. Pulmonary function tests if indicated for pulmonary disease.
14. Documentation of age-appropriate screening for extrahepatic malignances (e.g., colonoscopy, mammogram, pap smear, skin cancer screening by dermatologist) and abdominal CT or MRI to screen for hepatocellular carcinoma.
15. Liver function tests with transaminases <3X upper limit of normal and total bilirubin <2.5 mg/dl.
16. Carotid Doppler ultrasound (if patient has known coronary artery disease OR >age 50)-Abnormal findings evaluated further. Intervention and/or clearance required for abnormal findings.
17. Dental exam, x-rays, and treatment completed to eliminate sources of infection in the oral cavity.
    a) Examples include, but are not limited to gum disease, tooth decay, tooth abscesses, and poor oral hygiene.

*For additional selection criteria see the special instructions sections below.

Specific Diagnoses:
Isolated Small Bowel Transplant

A. A small bowel transplant using a cadaveric intestine may be considered MEDICALLY NECESSARY in adult and pediatric patients with life threatening complications attributable to irreversible intestinal failure, associated with all of the following criteria:
    a. Reduced gastrointestinal absorption resulting in the need for parenteral nutrition, AND
    b. Long-term dependency (minimum of two years) on total parenteral nutrition (TPN), AND
    c. Severe complications due to failed TPN (e.g., thrombosis of two or more major central venous channels, repeated central line-related sepsis, frequent episodes of severe dehydration).
B. A small bowel transplant using a living donor may be considered MEDICALLY NECESSARY only when a cadaveric intestine is not available for transplantation in a patient who meets the criteria noted above for a cadaveric intestinal transplant.
    a. A small bowel transplant using living donors is considered NOT MEDICALLY NECESSARY in all other situations. The long-term outcomes of living intestine donors have not yet been reported.
C. A small bowel transplant is considered INVESTIGATIONAL AND/OR EXPERIMENTAL for adults with intestinal failure who are able to tolerate TPN.

Small Bowel/Liver Transplant

A combined small bowel and liver transplant may be considered medically necessary for pediatric and adult patients with intestinal failure (characterized by loss of absorption and the inability to maintain protein-energy, fluid, electrolyte, or micronutrient balance) who have been managed with long-term total parenteral nutrition (TPN) and who have developed evidence of impending end-stage liver failure.

A. Small bowel / liver transplants using a cadaver donor may be considered MEDICALLY NECESSARY in pediatric and adult patients with short bowel syndrome (SBS) who meet all of the following:
    a. Patient has irreversible small bowel failure; and
    b. Patient has been managed with long-term TPN for a minimum of two years; and
    c. There is evidence of impending end-stage liver failure such as:
        i. Prolonged prothrombin time (PT) > 2 times the laboratory "control" value with normal range ordinarily 11 to 13.5 seconds; or

ii. Decreasing albumin to < 3.0 with normal range ordinarily 3.4 to 5.4 g/dL; and
iii. There is evidence of severe complications from TPN (e.g., liver dysfunction, repeated infections, thrombosis, difficult venous access for TPN or fluids)

Multivisceral Transplant
A multivisceral transplant may be considered medically necessary for pediatric and adult patients who meet criteria above for the combined small bowel and liver transplant and require 1 or more abdominal visceral organs to be transplanted due to concomitant organ failure or anatomical abnormalities that preclude a small bowel and liver transplant.
Multi-visceral transplants are considered experimental and investigational for individuals with neuroendocrine pancreatic tumors.

A. Simultaneous multi-visceral transplants (including small bowel and liver but can also include the stomach, duodenum, jejunum, ileum, pancreas, or colon), also known as cluster transplant, using a cadaver donor may be considered MEDICALLY NECESSARY in pediatric and adult patients with SBS who meet all of the following:
    1. Patient meets the criteria above for isolated small bowel transplant, **AND**
    2. Patient must have evidence of severe liver dysfunction
    3. Prolonged prothrombin time (PT) > 2 times the laboratory "control" value with normal range ordinarily 11 to 13.5 seconds, **OR**
    4. Decreasing albumin to < 3.0 with normal range ordinarily 3.4 to 5.4 g/dL, **AND**
    5. Patient must have evidence other than small bowel problems such as pancreatic failure, thromboses of the celiac axis, and the superior mesenteric artery or pseudo-obstruction affecting the entire gastric and small bowel tract, **AND**
    6. Patient must have tried or considered all other medically necessary medical and surgical therapies that might have been expected to yield both short and long-term survival comparable to that of transplantation, **AND**
    7. Pediatric patient should have a parent or legal guardian who has received pre and post-transplant education and informed consent of the range of clinical outcomes that may be encountered, **AND**
    8. Adult patients should also have received pre and post-transplant education and informed consent of the range of clinical outcomes that may be encountered, **AND**
    9. Patient agrees to follow the prescribed regimen of pre and post-transplant follow up.

A combined small bowel and liver transplant or multivisceral transplant for pediatric and adult patients performed for any other conditions not listed above will be considered not medically necessary.

Living donor intestinal or multivisceral transplantation for any indication is considered INVESTIGATIONAL AND/OR EXPERIMENTAL and NOT MEDICALLY NECESSARY.

Small intestine/liver or multivisceral transplant:
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 intestine/liver transplant or a multivisceral transplant, which includes the small intestine and liver with one 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 intestine/liver transplant.

Small intestine/Liver transplant or multivisceral transplant may be considered medically necessary in pediatric and adult patients with intestinal failure who have the loss of absorption and the inability to maintain protein-energy fluid, electrolyte, or micronutrient balance, who have been managed with TPN, and have developed evidence of impending end-stage liver failure. In addition, the patient usually has a history of surgery, abdominal trauma, motility disorders, tumor or other etiologies that warrant the multivisceral transplant. The multivisceral transplant can include the liver, stomach, small intestine (duodenum, jejunum and ileum), pancreas and/or colon. Kidney transplant may also be included if the recipient has end stage renal failure.

Indications:
The following are pediatric conditions causing intestinal failure:

- Short bowel syndrome following extensive bowel surgeries (midgut volvulus)
- Congenital malformations (e.g. intestinal atresia, gastroschisis, aganglionosis such as in Hirschsprung's disease)
- Absorptive impairment (e.g. microvillus involution disease, chronic intestinal pseudo-obstruction)
- Infections of gastrointestinal tract (e.g. necrotizing enterocolitis)
- Trauma
- Massive resection secondary to tumor
- Radiation enteritis

The following are the leading causes of intestinal failure in adults:

- Crohn's disease
- Tumors of the mesenteric root and retroperitoneum (e.g. desmoid tumor)
- Short bowel syndrome following extensive surgeries secondary to mesenteric ischemia (following thrombosis, embolism, volvulus, trauma, or gastroschisis)
- Chronic intestinal pseudo-obstruction
- Small bowel tumors such as Gardner's Syndrome (familial colorectal polyposis)
- Trauma
- Volvulus

The chronic use of TPN is often associated with life-threatening complication including:

- Catheter related sepsis
- Catheter related thrombosis
- Severe dehydration
- Parenteral nutrition associated liver disease (PNALD)

Absolute Contraindications:

1. Persistent noncompliance, see criteria in Member Assessment of Compliance
2. Inadequate support system as documented by a formal social worker evaluation
3. BMI > 40 – relative contraindication
4. Malignancy present with exception of non-melanotic skin cancer
5. Systemic illness or comorbidities that would be expected to substantially and negatively impact the successful completion and/or outcome of transplant surgery, such as, but not limited to:
    a. Severe cardiac disease (ejection fraction less than 40%)
    b. Severe pulmonary disease (diffusion capacity (DLCO) < 65% of predicted or dyspnea at rest requiring O2 or FEV1<1 L or FVC <50%,)
    c. Severe end stage organ damage including: Severe diabetes mellitus with end organ damage, irreversible severe pulmonary disease, with FEV1 <1 L or FVC <50%, irreversible severe hepatic disease, irreversible severe renal disease
    d. Poor kidney function (creatinine clearance less than 50 ml/min/kg)
    e. Advanced neurological disorders (e.g., neuroaxonal dystrophy, Tay-Sachs disease, Niemann-Pick disease and its variants, neuronal ceroid lipofuscinosis, and Huntington disease)
6. Significant infection that could be exacerbated by immunosuppressive therapy (bacterial, fungal or viral, such as chronic active viral hepatitis, hepatitis B, hepatitis C and AIDS-SEE APPENDIX A). **NOTE:** HIV infection must be controlled as evidenced by the following:
    a. CD4 counts > 200 for more than 6 months
    b. HIV-1 RNA undetectable
    c. The member is stable on anti-retroviral therapy more than 3 months

    d. The member has no other complications from acquired human immunodeficiency (AIDS) (e.g. opportunistic infection including aspergillus, tuberculosis, coccidioses mycosis, resistant fungal infections, Kaposi's sarcoma, or other neoplasm).
7. Systemic diseases affecting multiple organ systems that could be exacerbated by immunosuppression including but not limited to:
    a. Scleroderma
    b. Amyloidosis
    c. Diffuse atheromatous disease
8. Multisystem organ failure
9. Cerebral edema
10. Irreversible severe brain damage
11. Social and Psychiatric Issues-refer for psychosocial evaluation and/or psychiatry consultation for guidance.
    a. Emotional instability, significant depression or other psychiatric illness that cannot be controlled that would impact ability to comply with a complex evaluation process, surgical procedure and post-transplant plan of care and/or ability to give informed consent (and does not have a representative/guardian/conservator.
    b. Limited cognitive ability (memory loss, dementia, etc.) that would impact ability to comply with a complex evaluation process, surgical procedure and post-transplant plan of care and/or ability to give informed consent (and does not have a representative/guardian/conservator).
    c. Lack of psychosocial support as indicated by either no identified caregiver or an uncommitted caregiver. This would include the lack of ability to adhere to the requirements of transplant related treatment plan. A care contract may be needed.
    d. History of non-adherence that has not been successfully remediated.
    e. Inability to give informed consent. If the patient has an authorized representative/guardian/conservator or patient in the case of a minor, that individual must understand and support the ongoing health care needs of the patient.
12. Post-transplant lymphoproliferative disease (PTLD) unless no active disease demonstrated by negative positive emission tomography (PET) scan and resolved adenopathy on computed tomography (CT) and/or magnetic resonance emission tomography (CT) and/or magnetic resonance imaging.
13. Limited irreversible rehabilitative potential
14. Abdominal wall defects that would complicate closure.
15. Concurrent GI disorders (diverticulitis, bleeding peptic ulcer, chronic hepatitis.

Repeat Transplant

1. Retransplantation in individuals with graft failure of a combined small bowel and liver transplant or multivisceral retransplant due to technical reasons, hyperacute or chronic rejection, or return of disease may be considered medically necessary if the transplant criteria above have been met.

Member Compliance with Plan of Care (applicable for ages 10 and above):

- Alcohol screen- abstinence for the past 6 months prior to actual transplant approval, if member history includes use of alcohol. If no history exists then 1 negative alcohol screen must be submitted for members with no history of past alcohol use
- Drug screen-abstinence for the past 6 months prior to actual transplant approval if history exists of drug use. If no history exists then 1 negative drug screen must be submitted for members with no history of positive drug screen.
- Nicotine screening- abstinence for the past 6 months prior to actual transplant approval if history of smoking. If no history exists then 1 negative cotinine level must be submitted

Refusal or failure to undergo monthly testing for those members with a history of alcohol, tobacco, and/or drug use will be interpreted as a positive test result.

Six month abstinence period may be shortened in cases where patient's condition is sufficiently advanced that mortality is reasonably expected before the full abstinence period can be completed. Patients granted a waiver of the six month abstinence period require documentation of participation in a formal outpatient treatment program, when practical, as well as serial blood or urine testing no less frequently than monthly. A positive test result at any time prior to the procurement phase will result in denial.
Developmental or Acquired Cognitive Impairment and Dementia: Psychosocial and guardianship support as well as reversibility of impairment must be assessed and documented prior to non-compliance determinations.

Line of Business Applicability:
This policy applies to Michigan Medicaid, Illinois Medicaid, and Individual plans.

For **Medicaid/Medicaid Expansion Plan** members, this policy will apply. Coverage is based on medical necessity criteria being met and the codes being submitted and considered for review being included on either the Michigan Medicaid Fee Schedule (located at: http://www.michigan.gov/mdch/0,1607,7-132-2945_42542_42543_42546_42551-159815--,00.html), or the Illinois Medicaid Fee Schedule (located at: http://www.illinois.gov/hfs/MedicalProviders/MedicaidReimbursement/Pages/default.aspx). If there is a discrepancy between this policy and either the Michigan Medicaid Provider Manual (located at: http://www.michigan.gov/mdch/0,1607,7-132-2945_5100-87572--,00.html), or the Illinois Medicaid Provider Manual (located at: http://www.illinois.gov/hfs/MedicalProviders/Handbooks/Pages/default.aspx)the applicable Medicaid Provider Manual will govern.

For **Individual** members, consult the individual insurance policy. If there is a discrepancy between this policy and the individual insurance policy document, the guidelines in the individual insurance policy will govern.

State specific special instructions:

None: ☐

ALL: All

- The Chief Medical Officer and/or Senior Medical Director must receive notification for all possible approved requests by the UM team.

MI: Medicaid

- Member must be in Compliance with MHP's Member Compliance Medical Policy I.07.

IL: Medicaid

- Member must be in Compliance with MHP's Member Compliance Medical Policy I.07.
- Small Bowel transplants are not covered for members over the age of 21 as per the Illinois Administrative Code

OH:

References:

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13. Hayes. Health Technology Brief. Living Related Donor Small Bowel Transplantation for Intestinal Failure. June 26, 2014. Updated April 8, 2019.
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15. Overview of intestinal and multivisceral transplantation Farrukh A. Khan, M.D., FACS, Gennaro Selvaggi, M.D. UpToDate.com. Updated April 8, 2019

State Letters/Bulletins					
CMS National/Local Coverage Determination (NCD/LCD)	NCD 260.5 Intestinal and Multi Visceral Transplant, (v.2 May, 2006)				
CMS Local Coverage Determination (LCD)					
Medicare Managed Care Manual:					
Medicaid CFR:					
State Administrative Codes:			[441] 78.1(20)(d), page 8 (April 2012)		
Contract Requirements:					
Related Policies:	I.7 Member Compliance Medical Policy				

Appendix A
AIDS-Defining Conditions

Certain serious and life-threatening diseases that occur in HIV-positive people are called "AIDS-defining" conditions. When a person gets one of these illnesses, he or she is diagnosed with the advanced stage of HIV infection known as AIDS.

The Centers for Disease Control and Prevention (CDC) has developed a list of these conditions (see below). No single patient is likely to have all of these problems. Some of the conditions are rare.

1. Bacterial infections, multiple or recurrent (a)
2. Candidiasis of bronchi, trachea, or lungs
3. Candidiasis of esophagus (b)
4. Cervical cancer, invasive (c)
5. Coccidioidomycosis, disseminated or extrapulmonary
6. Cryptococcosis, extrapulmonary
7. Cryptosporidiosis, chronic intestinal (>1 month's duration)
8. Cytomegalovirus disease (other than liver, spleen, or nodes), onset at age>1 month
9. Cytomegalovirus retinitis (with loss of vision) (b)
10. Encephalopathy, HIV related
11. Herpes simplex: chronic ulcers (>1 month's duration) or bronchitis, pneumonitis, or esophagitis (onset at age >1 month)
12. Histoplasmosis, disseminated or extrapulmonary
13. Isosporiasis, chronic intestinal (>1 month's duration)
14. Kaposi sarcoma (b)
15. Lymphoid interstitial pneumonia or pulmonary lymphoid hyperplasia complex (ab)
16. Lymphoma, Burkitt (or equivalent term)
17. Lymphoma , immunoblastic (or equivalent term)
18. Lymphoma, primary, of brain
19. *Mycobacterium avium* complex or *Mycobacterium kansaii,* disseminated, or extrapulmonary (b)
20. *Mycobacterium tuberculosis* of any site, pulmonary (bc) , disseminated (b), or extrapulmonary (b)
21. *Mycobacterium,* other species or unidentified species, disseminated (b) , or extrapulmonary (b)
22. *Pneumocystis jiroveci* pneumonia (b)
23. Pneumonia, recurrent (bc)
24. Progressive multifocal leukoencephalopathy
25. *Salmonella* septicemia, recurrent
26. Toxoplasmosis of brain, onset at age >1 month (b)
27. Wasting syndrome attributed to HIV

Legend:

(a) Only among children aged <13 years. (CDC. Revised classification system for human immunodeficiency virus infection in children less than 13 years of age. MMWR 1994; 43 [No. RR-12].)

(b) Condition that might be diagnosed presumptively.

(c) Only among adults and adolescents aged ≥ 13 years. (CDC. 1193 Revised classification system for HIV infection and expanded surveillance case definition for AIDS among adolescents and adults. MMWR 1992; 41 [No. RR-17])