

# Imaging in pediatric small bowel transplantation

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## Abstract

Small bowel transplantation, alone or with other organs as multivisceral transplantation, is performed for patients with chronic intestinal failure. With advancing surgical techniques and improved post-surgical management, survival of these patients has increased tremendously in the last two decades. The radiologist has an important role in the preoperative and postoperative management of these patients. Knowledge of surgical techniques and post-surgical complications seen in the transplant recipient is necessary for adequate management of these patients.

**Key words:** Bowel transplantation; pediatric small bowel transplantation; pediatric transplantation; small bowel transplantation; transplantation

## Introduction

Intestinal failure is the end result of various conditions that cause permanent failure of the small intestine to maintain adequate function, and can be life threatening in children.<sup>[1]</sup> Patients with intestinal failure are usually treated with bowel rest and parenteral nutrition. The long-term mortality ranges from 65 to 80%, depending on the cause of intestinal failure.<sup>[1]</sup> The definitive treatment for intestinal failure is small bowel transplantation (SBT), which is an innovative surgical technique that has dramatically changed the prognosis of children and adults with this condition.<sup>[1-7]</sup> Children of age 0-5 years constituted 41.2% of the total number of patients waiting for SBT in 2011.<sup>[3]</sup>

Over the last two decades, surgical advances in SBT, along with improved preoperative evaluation and postoperative care have significantly reduced the mortality and morbidity in patients undergoing transplant.<sup>[1,2]</sup> Following the first case of SBT in 1964, the infant survived only 12 h.<sup>[4]</sup> The number of living pediatric recipients with a functioning SBT has progressively increased, with 600 recipients in 2011 compared to 400 in 2007.<sup>[3]</sup> The longest surviving living donor intestinal transplant is 11 years and cadaveric transplant is 18 years.<sup>[5]</sup>

SBT may be performed alone, in combination with a liver transplant, or as part of a multivisceral transplant (MVT) to include any combination of liver, stomach, pancreas, colon, spleen, and kidney depending on the underlying cause of intestinal failure.<sup>[1-4]</sup> The radiologist has an important role in a multidisciplinary team for SBT to evaluate the anatomy and early and delayed post-transplant complications. This article will provide a complete review of SBT, including indications, imaging protocols, anatomy, and pre- and post-transplant imaging evaluation.

## Indications

Intestinal failure occurs when the intestine can no longer absorb enough water and nutrients to provide adequate

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growth and development.<sup>[1,2]</sup> In such cases Total Parenteral Nutrition (TPN) can be administered, but it is often associated with long-term complications. In addition to complications related to venous access catheter for TPN such as blockage, sepsis, and thrombosis of central veins, children are also susceptible to hepatobiliary complications.<sup>[6]</sup> These include cholestasis, cholelithiasis and hepatic dysfunction, and failure.

Failure of TPN can also be considered as an indication for SBT when there is development of hepatic dysfunction and failure, loss of two or more central veins for venous access, and recurrent catheter line infections.<sup>[1,2]</sup>

In children, intestinal failure occurs for many reasons. It can be divided broadly into two types. The first occurs when the small intestine is abnormally shortened, and is referred to as "short gut syndrome." The second occurs when the small intestine has lost its normal function.<sup>[1]</sup> The small intestine may become short due to complications and/or surgery related to midgut volvulus, gastroschisis, intestinal atresia, necrotizing enterocolitis, or intestinal polyposis.<sup>[1-7]</sup> Intestinal failure can occur in children secondary to aganglionosis, pseudo-obstruction, and microvillus inclusion disease.<sup>[6,7]</sup>

#### Pre-transplant evaluation

Important components of pre-transplant evaluation include a thorough history and physical examination with details of all previous operative procedures, laboratory evaluation of organ function, nutritional evaluation, and imaging to evaluate for anatomy, radiological evidence of disease, and contraindications for surgery.<sup>[1]</sup> Depending upon institutional guidelines and protocols, routine laboratory testing such as complete blood count (CBC), coagulation profile, liver function tests (LFTs), and urinalysis are performed. A complete nutritional evaluation of the patient should be performed, as many patients have not learned or have forgotten to eat because they have been on long-term TPN.<sup>[1]</sup> Liver function assessment is important to determine any long-term effects of TPN therapy. Hepatocellular reserve can be evaluated with coagulation profile, albumin level, and ammonia level. Liver biopsy may also be performed, if necessary.<sup>[1]</sup>

Imaging protocols vary with the clinical scenario, but may include multiple modalities such as fluoroscopic upper and lower gastrointestinal barium studies, abdominal ultrasound (US), radiographic examinations, computed tomography (CT), and magnetic resonance imaging (MRI) [Table 1]. The intestinal tract should be evaluated for anatomy and the extent of small bowel disease. The extension of disease into the stomach and colon should also be evaluated. If there is history of previous surgery, portion (s) of the bowel may be missing or anatomically altered in position. Motility studies and absorption studies can be performed when appropriate.<sup>[1,7]</sup>

In addition to imaging evaluation of the bowel itself, all patients being evaluated for SBT should have imaging to evaluate the anatomy of associated vasculature and the abdominal wall, to exclude any evidence of malignancy in the abdominal viscera, or to identify any other contraindications for surgery.<sup>[2]</sup> US evaluation of liver in gray-scale and Doppler can assess the liver disease and portal vein thrombosis. If there is difficulty in evaluation of the portal vein due to bowel gas or anatomical difficulty, CT or MRI in portal venous phase should be performed. Portal vein thrombosis or occlusion is not an absolute contraindication for SBT, but would indicate the necessity of MVT.

MRI is frequently utilized for the evaluation of liver disease and inflammatory bowel disease. However, its role in patients being evaluated for SBT is still not clear. MRI can provide multiphase imaging, without the radiation of CT; this is advantageous over CT, since these patients are young and will require multiple examinations during their lifetime.

CT imaging typically should include the abdomen and pelvis, although institutional policy to include the chest may vary. The abdomen and pelvis should be evaluated in both arterial and portal venous phases. The arterial phase helps to assess the number and location of native arteries including the iliac arteries and evaluate incidental stenosis, aneurysms, and presence of accessory arteries.<sup>[2]</sup> The portal venous phase helps to evaluate portal venous

**Table 1: Pre-transplant imaging evaluation**

Area of interest	Imaging study	Technique	Indication
Native bowel	Small bowel follow-through	Water-soluble contrast or barium	Assess residual small bowel
Abdominal vessels	Doppler US or CT angiography	US: Gray-scale and color Doppler with spectral waveform analysis of the aorta, bilateral common iliac arteries and veins, portal venous system, and IVC for IIT. Liver inflow and outflow assessed for LIT. CT: Arterial and portal venous phases	Identify variant anatomy and pre-existing conditions, such as IVC thrombosis, that may affect suitability of recipient
Liver	US or MRI	US: Gray-scale imaging	Assess liver for TPN-induced liver disease that could be addressed with liver-intestinal transplantation
Peritoneal cavity	CT or MRI	Manual or computer-generated volume	Assess sufficiency of intra-abdominal volume to accommodate graft

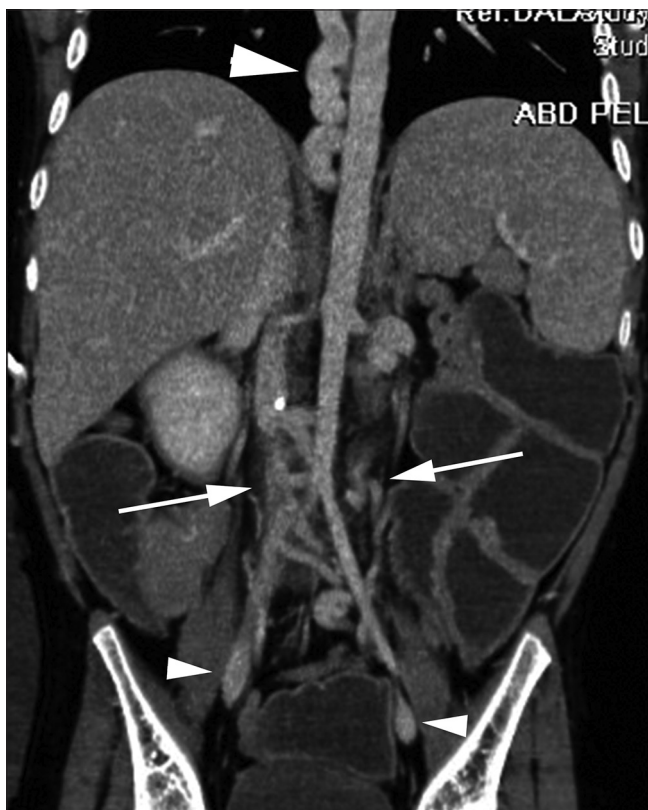
MRI=Magnetic resonance imaging, CT=Computed tomography, IVC=Inferior vena cava, IIT=Isolated intestinal transplantation, isolated intestinal transplantation, TPN=Total parenteral nutrition, LIT=Liver-intestinal transplantation

thrombosis, its extent, if present, and also assess any incidental anomalous venous drainage [Figure 1]. If kidneys are also being transplanted, a delayed CT phase to assess the collecting system and ureters may also be needed. The chest and pelvis can be covered during the portal venous phase. MRI of abdomen and pelvis would include similar phases; however, the chest should be evaluated with CT using a low-dose protocol and contrast should be used when indicated.

CT or MRI can also be used to assess the abdominal volume. Patients with history of intestinal failure and long-term TPN can have a scaphoid abdomen due to loss of intra-abdominal fat. SBT or MVT would need sufficient intra-abdominal volume to accommodate the graft within the abdomen.<sup>[1]</sup> Tissue expanders and plastic surgery may be used for expanding the intra-abdominal volume.<sup>[1]</sup> The anterior abdominal wall should also be evaluated as there may be fistulas, adherence of small bowel to the abdominal wall, or scarring from previous surgeries.<sup>[9]</sup>

#### Types of small bowel transplants

SBT can be divided into isolated intestinal transplantation



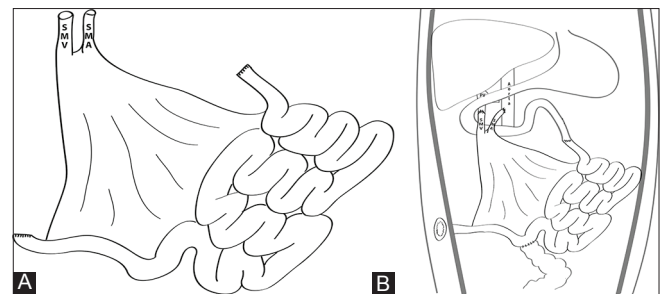
**Figure 1:** An 8-year-old boy with intestinal failure related to pseudo-obstruction. Coronal MIP from a pre-transplant CT angiogram shows infrarenal thrombosis of the IVC, with tapering of the distal IVC with multiple adjacent, enlarged collateral veins (arrows). Portions of the bilateral iliac veins (small white arrowheads) are still patent. A dilated and tortuous azygous vein is noted (large arrowhead). Thrombosis of the IVC may hinder feasibility of transplantation. Furthermore, patients with IVC thrombosis have poorer outcomes after transplantation

(IIT), liver-intestinal transplantation (LIT), and MVT, depending on whether the small bowel is transplanted alone, with liver, or with multiple organs which may include the stomach, liver, pancreas, colon, spleen, and kidney.

IIT [Figure 2] is performed when only the intestine is diseased and the other abdominal organs are functioning normally. Surgical techniques may have some minor differences based on institutional preferences, but the overall technique is similar.<sup>[2,7,8]</sup> In all types of procedures, a distal loop ileostomy is formed using the donor small bowel to allow visual and ileoscopic examination for rejection and can be closed 3-6 months after SBT.<sup>[7,8]</sup> The donor intestines may be placed either orthotopically or heterotopically in the recipient.<sup>[7,8]</sup> The proximal end of small bowel is divided close to the ligament of Treitz and the distal end is transected proximal to the ileocecal valve. If the ileocecal valve is preserved, an ileoileal anastomosis is performed; otherwise, the donor ileum can be anastomosed to the residual recipient colon. For patients with previous proctocolectomy, a terminal ileostomy is performed.<sup>[8]</sup>

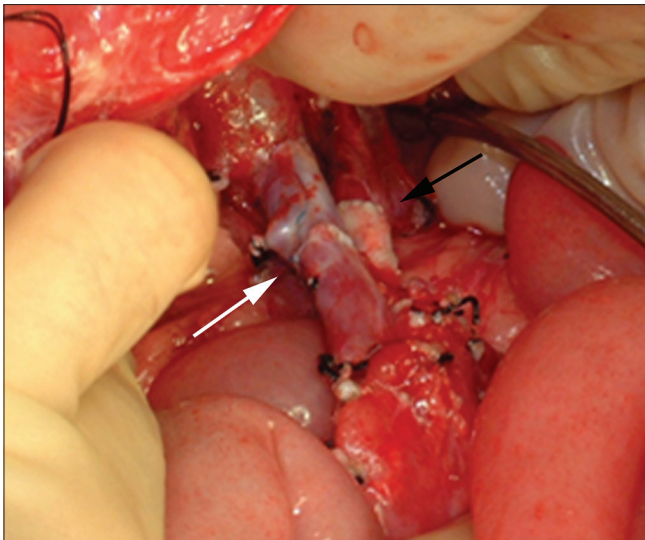
In orthotopic transplantation, the donor superior mesenteric artery (SMA) is anastomosed in an end-to-side fashion to the recipient infrarenal abdominal aorta above the inferior mesenteric artery (IMA) and the donor superior mesenteric vein (SMV) is anastomosed to the recipient SMV or portal vein [Figure 3].<sup>[8]</sup> In heterotopic transplantation, an end-to-side anastomosis between the donor SMA and the recipient infrarenal abdominal aorta below the origin of the IMA or common iliac artery is performed and the venous anastomosis is between the recipient inferior vena cava (IVC) or common iliac vein and the donor SMV.<sup>[8]</sup>

MVT is described first due to overlap of the surgical technique with LIT. In MVT, besides the liver and small bowel, the pancreas, part of the stomach, and the duodenum are transplanted [Figure 4]. The various organs from the



**Figure 2:** Schematic of the IIT graft before (A) and after (B) transplantation. The graft may be placed orthotopically, as shown. Alternatively, the graft may be placed heterotopically, typically with a venous anastomosis between the SMV and IVC, and an arterial anastomosis between the SMA and right common iliac artery. End ileostomy of the graft facilitates inspection, endoscopy, and biopsy (Reproduced from Phillips, Bhargava, Stanescu, *et al.* Pediatric intestinal transplantation: Normal radiographic appearance and complications. *Pediatric Radiology* 2011;41:1028-39; with permission)

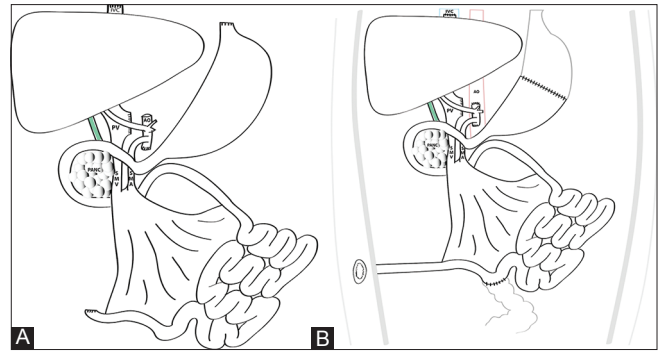




**Figure 3:** Intraoperative view of the SMV (white arrow) and SMA (black arrow) anastomoses of an isolated intestinal transplant. Photo courtesy of Dr. Jorge Reyes (Reproduced from Phillips, Bhargava, Stanescu, et al. Pediatric intestinal transplantation: Normal radiographic appearance and complications. Pediatric Radiology 2011;41:1028-39; with permission.)

donor are mobilized *en bloc* without manipulation of the portal venous system.<sup>[8]</sup> In the recipient, the liver, pancreas, spleen, part of the stomach, and the small bowel are also removed. The proximal bowel anastomosis is performed by connecting the proximal portion of the recipient stomach to the donor stomach. The distal bowel anastomosis is performed as described in the discussion of IIT. The donor aorta is excised above the celiac trunk and directly below the SMA to preserve the origins of the renal arteries. Below the SMA, the aorta is closed, forming a conduit for the vascular flow to reach the donor SMA and celiac trunk once an end-to-side aorto-aortic anastomosis is established with the infrarenal recipient aorta.<sup>[8]</sup> Venous anastomosis is performed in an end-to-end fashion between the suprahepatic and infrahepatic IVC or by using a piggyback technique with a graft caval stump end-to-side or side-to-side with the recipient IVC.<sup>[7]</sup> In addition, cholecystectomy and splenectomy are performed, as well as a pyloroplasty to prevent gastric outlet obstruction since the stomach is denervated.<sup>[8]</sup>

LIT is performed in patients who have small bowel failure and hepatic disease either due to TPN or other diseases. The liver and small bowel, along with the biliary system, portion of duodenum, and adjacent pancreatic tissue are transplanted *en bloc* to avoid an additional anastomosis, reducing the complications seen in older techniques [Figures 5 and 6].<sup>[6]</sup> The stomach, duodenum, and pancreas are preserved in the recipient. The residual recipient portal vein is anastomosed to the native suprarenal IVC in an end-to-side fashion. The rest of the vascular supply is anastomosed analogous to MVT. Proximal bowel



**Figure 4:** Schematic showing the MVT graft consisting of foregut and midgut before (A) and after (B) transplantation. Arterial and venous anastomoses are similar to those in the liver-intestinal transplant. The donor IVC may be joined end-to-end with the recipient IVC, or alternatively, either end-to-side or side-to-side using a piggyback technique (Reproduced from Phillips, Bhargava, Stanescu, et al. Pediatric intestinal transplantation: Normal radiographic appearance and complications. Pediatric Radiology 2011;41:1028-39; with permission.)

anastomosis is made between the recipient duodenum or jejunum and the donor proximal jejunum.<sup>[8]</sup> The distal anastomosis is similar to IIT and MVT.

#### The living donor

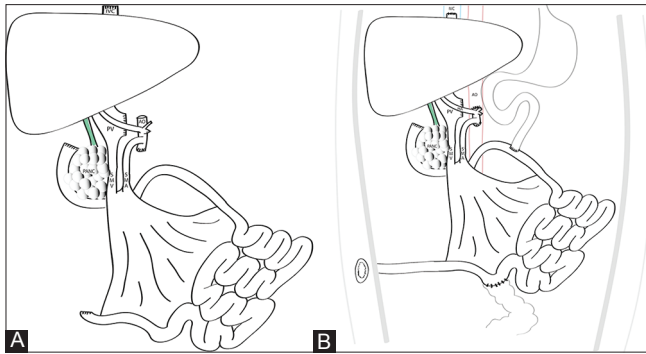
Preoperative imaging of the living donor is performed to assess the vascular anatomy and to exclude any contraindications for surgery.<sup>[10]</sup> CT angiography (CTA) is preferred over conventional angiography due to its superiority in assessing the venous system.<sup>[11]</sup> CT offers the additional advantage of assessment of the bowel and other organs for any unsuspected mass.

Approximately 120-160 cm of the small bowel is obtained from the donor for pediatric SBT, starting from 20 cm away from the ileocecal valve.<sup>[10]</sup> At least 60% of the bowel is left in the donor.<sup>[10]</sup> After resection, donor intestinal continuity is reestablished with an end-to-end anastomosis. The donor is closely monitored for any early or late complications. Postoperative imaging is only performed if a complication is suspected.<sup>[10]</sup>

For donors with only SBT, complications are the same as any small bowel surgery including post-surgical ileus, small bowel obstruction (SBO), leakage, collections/abscesses, infections, and vasculature complications. CTA is the preferred modality for all the above indications. Post-transplant diarrhea is commonly encountered and usually not evaluated with imaging.<sup>[10]</sup>

#### Post-transplant evaluation

The imaging modality selected for post-transplant evaluation is dictated by the type of post-surgical complication in question [Table 2]. Upper gastrointestinal (GIT) and small bowel series are only occasionally performed for obstruction and motility.<sup>[12]</sup> Motility can also be evaluated with US, but



**Figure 5 (A and B):** Schematic of the *en bloc* LIT graft with a portion of the pancreatic head, before (A) and after (B) transplantation. The graft is typically harvested with a contiguous segment of donor aorta that preserves the origins of the celiac axis and SMA, from which an aortic conduit may be fashioned. Recipient portal vein is typically diverted with a portocaval shunt. Donor biliary system remains intact (Reproduced from Phillips, Bhargava, Stanescu, *et al.* Pediatric intestinal transplantation: Normal radiographic appearance and complications. Pediatric Radiology 2011;41:1028-39; with permission.)

**Table 2: Imaging of suspected intestinal transplant complications**

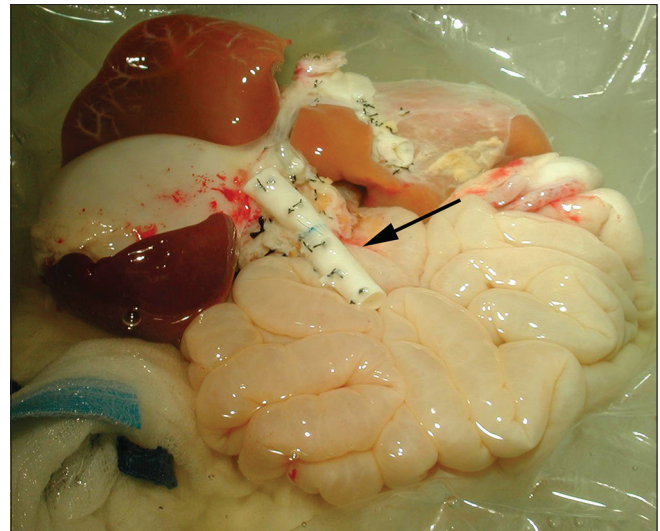
Complication	Imaging study	Technique
Rejection	None	Endoscopy/Biopsy
Free air	Abdominal radiographs	Supine and left lateral decubitus (or upright)
Obstruction or dysmotility	Fluoroscopy	Water-soluble contrast
Infection	US	Focused US to address the clinical question (such as "rule out abscess")
Vascular compromise	Doppler US	Assess inflow and outflow of the transplant; vascularity of individual loops may also be subjectively assessed
PTLD	CT	With both oral and IV contrast

PTLD=Post-transplant lymphoproliferative disorder

it is operator dependent and patients with SBT frequently have gaseous distension and ileus hampering optimal evaluation of the bowel.<sup>[12]</sup> However, US has the advantage of being available bedside for sick or critical patients and unlike CT, does not expose the patients to unnecessary radiation, which is especially important for the pediatric population.<sup>[7]</sup>

US is routinely performed for vascular assessment, and often performed to guide aspiration of large peritransplant fluid collections and to assess interval change for postoperative ascites and small collections. Doppler US is routinely performed on postoperative day 1 after SBT to establish a baseline and to detect unsuspected immediate postoperative complications.<sup>[5]</sup> The normal donor SMA waveform shows variable resistive index as seen in the native SMA, depending upon the fasting or postprandial state of patient.<sup>[13]</sup>

CT scan is the primary imaging modality used in both the immediate and late postoperative periods to evaluate SBT patients for complications [Figure 7].<sup>[12]</sup> It has the



**Figure 6:** Composite LIT allograft shown with a sizeable portion of donor aorta attached (arrowhead). Photo courtesy of Dr. Jorge Reyes (Reproduced from Phillips, Bhargava, Stanescu, *et al.* Pediatric intestinal transplantation: Normal radiographic appearance and complications. Pediatric Radiology 2011;41:1028-39; with permission)

capability to assess the bowel, vasculature, and abdominal cavity in SBT and MVT patients.<sup>[12]</sup> In patients with renal failure, contrast can be avoided with an unenhanced CT or a noncontrast MRI can be considered. The disadvantage of MRI in pediatric population is the need for anesthesia, especially for young children. The transplanted bowel could be evaluated with MRI, but the enteric anastomosis is often difficult to assess.<sup>[12]</sup> Rejection of SBT is best evaluated with repeated biopsies by endoscopy in the first few weeks following transplant.<sup>[1]</sup> CT and US-guided biopsy of the transplanted liver, pancreas, and kidney in MVT patients can be performed safely.<sup>[15]</sup>

### Imaging complications

#### Rejection

Acute cellular rejection is the leading cause of graft loss in the first 2 months after SBT.<sup>[14]</sup> SBT patients are closely monitored for rejection as it is the most common complication. The small bowel is periodically evaluated by manual inspection of the ileostomy site and by serial endoscopic examinations.<sup>[1,5]</sup> Biopsies are obtained from several locations to detect early evidence of rejection and initiate immediate treatment to save the transplanted bowel.<sup>[14,16]</sup> Symptoms of rejection include fever, nausea, vomiting, diarrhea, abdominal pain, and increased and/or bloody stool.<sup>[14,16]</sup>

The imaging appearances of rejection include focal or diffuse bowel wall thickening and hyperenhancement [Figure 8].<sup>[2,7,8]</sup> Similar appearance can be seen with other complications, most commonly infection and blood flow compromise.<sup>[7,8]</sup> Therefore, endoscopy with biopsy is routinely performed on all SBT patients even if they are clinically stable, as CT



findings are nonspecific. CT can be useful in evaluating the extent of small bowel involvement and to detect other complications.<sup>[2]</sup> With advances in technology, MRI has become an excellent modality for evaluation of inflammatory bowel disease, but its role in graft evaluation is still not well established. The role of MRI should be further investigated, especially in children, as it does not require radiation and SBT patients often need multiple examinations.

Chronic rejection is the main cause of late intestinal graft dysfunction.<sup>[14]</sup> Clinically, patients with chronic rejection have a prolonged, insidious course and lack early specific clinical symptoms or mucosal findings. Full-thickness biopsies are needed to demonstrate the nonspecific changes of fibrosis which are often seen with chronic rejection.<sup>[14]</sup> Mucosal biopsies are often noncontributory. On imaging, the bowel may appear thickened with loss of mucosal folds.<sup>[14]</sup>

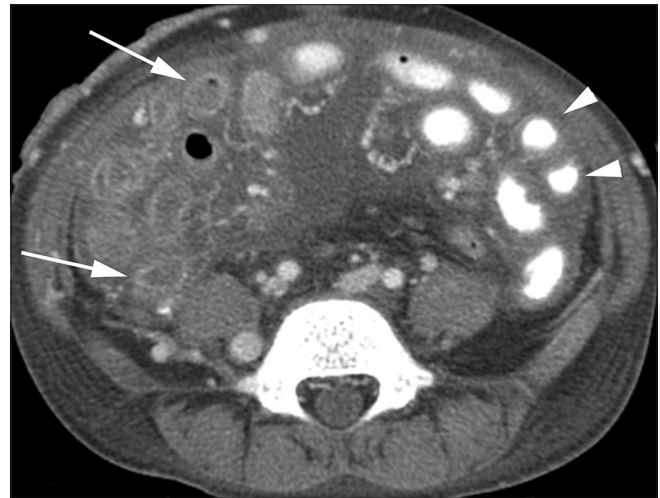
### Vascular complications

The most serious vascular complication is arterial or venous graft thrombosis that accounts for 10% of intestinal transplant patient mortality.<sup>[7]</sup> Other vascular complications include stenosis [Figure 9], dehiscence, hemorrhage, and pseudoaneurysm.<sup>[2,12]</sup> US is often used as first-line imaging to evaluate vascular compromise because of its portability and availability. Gray-scale and color Doppler US with spectral waveforms can directly visualize the thrombus as a hypo- to hyperechoic mass with absence of color flow and loss of venous Doppler signal. In addition, color Doppler is also useful to assess bowel perfusion. However, US can be confounded by technical difficulties from bowel gas and edematous bowel. With the advances in CT technology, CT/CTA has become an essential diagnostic tool in patients with complex vascularity and post-surgical changes. If vascular patency is still in question following US and CTA, catheter angiography can be performed to detect venous thrombosis and arterial complications.<sup>[2]</sup> Complicated anastomosis and collateral vasculature can make interventional radiology procedures challenging.<sup>[17]</sup>

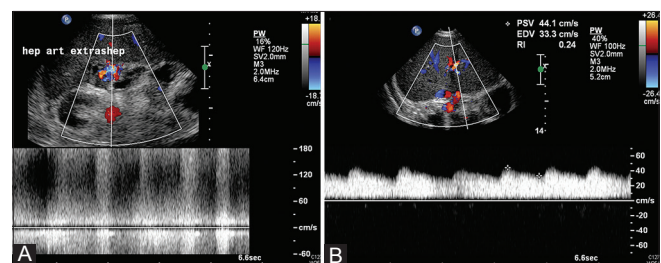
On CTA, a thrombus will appear as an intraluminal partial or complete filling defect with or without vein dilatation.<sup>[12]</sup> CT also has the advantage of evaluating the intra-abdominal organs that may have been affected by vascular compromise. The size of hematomas can be well outlined, and the associated mass effect on surrounding structures can be well seen. Portal venous thrombosis will result in ascites and splenomegaly.<sup>[12]</sup> The bowel may become edematous and dilated. In arterial narrowing, the percentage stenosis can be measured. Bowel necrosis and infarction due to arterial compromise are associated with bowel wall thickening, non-enhancement of bowel wall, pneumatosis of bowel wall, and porto-mesenteric air.<sup>[18]</sup>



**Figure 7:** A 1-year-old boy who underwent LIT for intestinal failure related to gastroschisis and small bowel atresia, now presenting with gram-negative sepsis. Contrast-enhanced CT (CTDIvol 4.39 mGy; DLP 120.05 mGy cm) shows normal appearance of the native (arrowheads) and transplant (arrow) pancreas, as well as nondistended transplant bowel (asterisk)



**Figure 8:** An 11-year-old boy with fever, increased ostomy output, and suspected infection who underwent IIT for pseudo-obstruction 3 years before. Axial image from contrast-enhanced CT (CTDIvol 4.3 mGy, DLP 179.28 mGy cm) shows thickened bowel loops (arrowheads) and mucosal hyperemia (arrows) suggesting bowel ischemia. Severe cellular rejection with mucosal necrosis was observed at biopsy

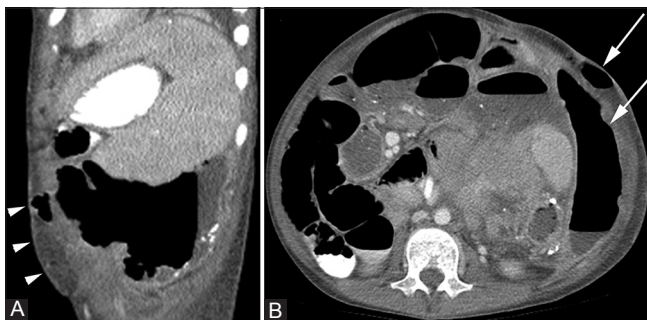


**Figure 9 (A and B):** A 2-year-old girl who underwent LIT for gastroschisis, short gut syndrome, and TPN-related liver disease. (A) Aliasing is seen of the celiac arm of the aortic conduit (B) Interrogation of the hepatic artery shows tardus parvus waveform distal to the stenosis

### Infection

Immunosuppressive therapy presents a risk for developing opportunistic infections in transplant patients.<sup>[14]</sup> Small bowel infections, particularly bacterial and fungal, are more common in patients with SBT due to immunosuppression.<sup>[7]</sup> Guaraldi *et al.* ascertained that the causative organisms of infections in decreasing order were bacteria (94%), viruses (67%), and fungi (28%).<sup>[16]</sup> Bacterial infections, which are the most common, have been reported to affect more than 80% of SBT patients within 2 months after transplant.<sup>[14]</sup> Viral infections are the second most frequently occurring infection, with rotavirus being the most common in the pediatric population.<sup>[14]</sup> Other viral infections affecting the gastrointestinal tract are cytomegalovirus (CMV), Epstein-Barr virus (EBV), and adenovirus.<sup>[14]</sup> CMV is the most serious viral infection, as it can result in graft loss and sometimes death.<sup>[7]</sup> Fungal infections occur later compared to bacterial and viral infections, with *Candida albicans* being the most common organism.<sup>[14]</sup>

The clinical and radiological appearance of SBT rejection or intestinal infection is similar, but management is dramatically different; therefore, histopathology plays a vital role in establishing the correct diagnosis.<sup>[14]</sup> The CT appearance of infection can include bowel wall thickening; the bowel wall may also have diffuse or focal areas of hyperenhancement similar to changes found with rejection and vascular compromise. Bowel wall edema can appear in the first 2 months after SBT in normal patients, likely related to a harvesting injury.<sup>[7]</sup> Infection in the abdomen can also manifest as abscess, peritonitis, and fistula formation [Figure 10].<sup>[12]</sup> The bowel loops appear dilated on radiographs and are unchanged in appearance (fixed loops of bowel) on serial abdominal radiographs in patients with peritonitis. Oral contrast may help delineate a fistula and communication with an abscess. CT may also help the interventional radiologist in image guidance for diagnostic aspiration to evaluate the type of pathogen for effective antimicrobial therapy or therapeutic drain placement.<sup>[17,19]</sup>



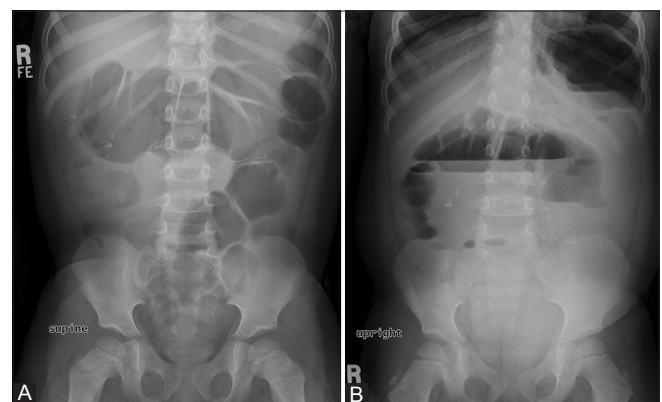
**Figure 10 (A and B):** A 7-year-old boy who underwent LIT for intestinal failure secondary to gastroschisis, small bowel resections, and TPN-related liver disease. Sagittal (A) and axial (B) Images from a contrast-enhanced CT show a mixed collection of gas and fluid within the left flank (arrows) and anterior abdominal wall (arrowhead) soft tissues, consistent with fistula formation

Infection can also occur in other regions of the body due to immunosuppression in patients with SBT. Short- and long-term venous access lines and catheters can get infected.<sup>[2]</sup> Pulmonary infections are also common. If chest radiographs are normal, CT may help in detecting the underlying pathology. Even if CT findings are nonspecific, they may help identify the region of lung involved for sampling.<sup>[2]</sup> Those patients with MVTs can have a source of infection in other transplanted organs besides the small bowel, such as kidneys or liver. Therefore, CT with contrast is usually the most appropriate investigation when patients with SBT get symptoms and signs of infection.

### Graft dysfunction/dysmotility

Small bowel dysmotility and ileus is not unusual in the first 3 days after surgery.<sup>[20]</sup> It is difficult to differentiate between ileus and mechanical SBO in the immediate postoperative period as imaging findings can be very confusing [Figure 11].<sup>[20]</sup> Serial abdominal radiographs may show persistent bowel dilatation that should gradually reduce over time in ileus with worsening dilatation; however, there is overlap between both entities that can make diagnosis with radiographs unreliable.<sup>[20]</sup>

Fluoroscopic water-soluble small bowel studies and real-time US can evaluate intestinal motility, which can be delayed up to 5 h in the immediate postoperative period.<sup>[21]</sup> Often US is suboptimal due to increased bowel gas in patients with bowel distension. Fluoroscopic examination has the added benefit of assessing the site and cause of obstruction, to include volvulus, stricture [Figure 12], or adhesions. Fluoroscopic studies may also be able to assess dehiscence with free leakage into the abdominal cavity or a contained collection.<sup>[2,7,12]</sup> The mucosal pattern can be edematous in normal patients and in early rejection. With more severe



**Figure 11 (A and B):** A 4-year-old girl with past history of LIT at 2 years of age for intestinal failure related to microvillus inclusion disease. Supine (A) and upright (B) Conventional radiographs show dilated, gas-filled bowel loops with air-fluid levels. No obstruction was found on subsequent small bowel follow-through. This radiographic pattern may be seen with intestinal dysmotility (Reproduced from Phillips, Bhargava, Stanescu, *et al.* Pediatric intestinal transplantation: Normal radiographic appearance and complications. Pediatric Radiology 2011;41:1028-39; with permission)



rejection, dysmotility will be present with ablation of the normal mucosal pattern on fluoroscopic exam.<sup>[21]</sup> In patients with MVT, there can be delayed gastric emptying due to the denervation of gastric supply; therefore, pyloroplasty is routinely performed to avoid gastric outlet obstruction.<sup>[2,8]</sup>

In patients with clinical suspicion of bowel obstruction, CT is more effective in differentiating between postoperative dysmotility and obstruction.<sup>[20]</sup> CT sensitivity and specificity were 100% compared to combined clinical and plain film findings with sensitivity of 19% in distinguishing between postoperative ileus and mechanical small bowel obstruction in an older study.<sup>[20]</sup> Extraluminal causes of obstruction such as abdominal hematoma or fluid collection, intramural hematoma in the early postoperative period, and sclerosing peritonitis, which may be seen in chronic graft failure,<sup>[20,22]</sup> may also be better seen with CT [Figures 13 and 14].

#### Post-transplant lymphoproliferative disorder

Post-transplant lymphoproliferative disorder (PTLD) is a serious but rare complication in patients with transplantation, which can occur in the transplanted organ, in the recipient's remaining bowel, or even other organs.<sup>[12]</sup>

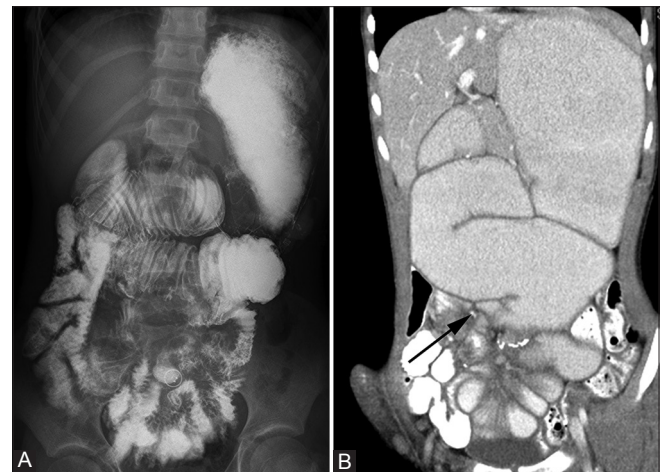


**Figure 12:** A 2-year-old girl who underwent LIT for intestinal failure related to microvillus inclusion disease. Spot fluoroscopic image from a water-soluble contrast enema performed per rectum shows luminal narrowing (arrow) of a segment of distal small bowel in the left upper quadrant

PTLD is a lymphoid tumor characterized by proliferation of recipient B-lymphocytes, which is caused by the EBV.<sup>[1,2]</sup> In a study of 127 patients, a total of 27 patients developed PTLD after SBT, with an overall frequency of 21%.<sup>[23]</sup> It may appear either as a primary infection or as reactivation of remotely acquired disease.<sup>[2]</sup>

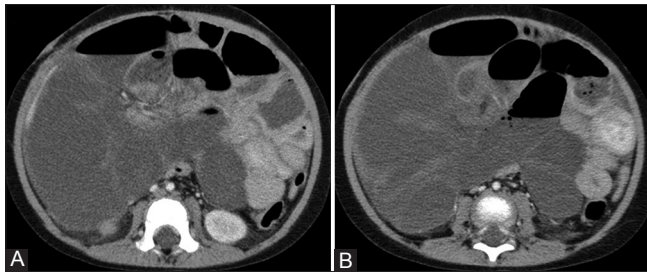
PTLD is seen more commonly in SBT recipients than in solid organ transplant patients.<sup>[11]</sup> PTLD is seen more commonly in children than in adults after transplantation, and in those children with more severe history of organ rejection who were given tacrolimus.<sup>[2]</sup> Patients with MVT also have a higher incidence of PTLD than isolated intestinal transplant patients.<sup>[7]</sup> Routine surveillance for EBV is done for early detection of infection.<sup>[11]</sup>

On imaging, the presentation of PTLD is variable. The most common presentation of PTLD is lymphadenopathy, which may be seen on US or CT.<sup>[1,2]</sup> A sudden and new onset of enlarged intra-abdominal lymph nodes in a patient with history of transplant warrants early biopsy to evaluate for PTLD. The transplanted organ itself or the patient's remaining bowel can be involved; however, usually lymphadenopathy is also present. The bowel may be thickened, usually more than that seen with infection or rejection, and may even appear mass-like [Figure 15]. Central nervous system (CNS) presentation is associated with a poorer prognosis.<sup>[11]</sup> CT is useful for planning and guiding lymph node biopsy, or to assist in identification of sites of bowel involvement for endoscopic biopsy.<sup>[11]</sup> PTLD can occur anytime, but usually occurs after a few months to several years.<sup>[11]</sup> PTLD frequency can be reduced



**Figure 13 (A and B):** A 9-year-old boy with past history of IIT for intestinal failure secondary to midgut volvulus. (A) UGI series shows a transition zone between dilated proximal and normal-caliber distal small bowel loops, suggesting partial small bowel obstruction (b) Correlative coronal reformatted image from a contrast-enhanced CT (CTDIvol 2.81 mGy, DLP 119.68 mGy cm) shows markedly dilated proximal small bowel loops with a transition zone (arrow) in the right lower abdomen. Pathology of the enterectomy specimen was consistent with sclerosing peritonitis, a rare late complication of small bowel transplantation





**Figure 14 (A and B):** A 2-year-old girl who underwent LIT for microvillous inclusion disease. (A and B) Axial images from contrast-enhanced CT demonstrate dilated bowel loops in the right abdomen with decreased enhancement. Volvulus of a portion of the allograft was confirmed at surgery, necessitating partial allograft excision

by aggressive EBV surveillance in peripheral blood and supplemental donor bone marrow infusion.<sup>[23]</sup> PTLD may also present as small bowel obstruction and intussusception, especially in children, which requires further workup.<sup>[24]</sup>

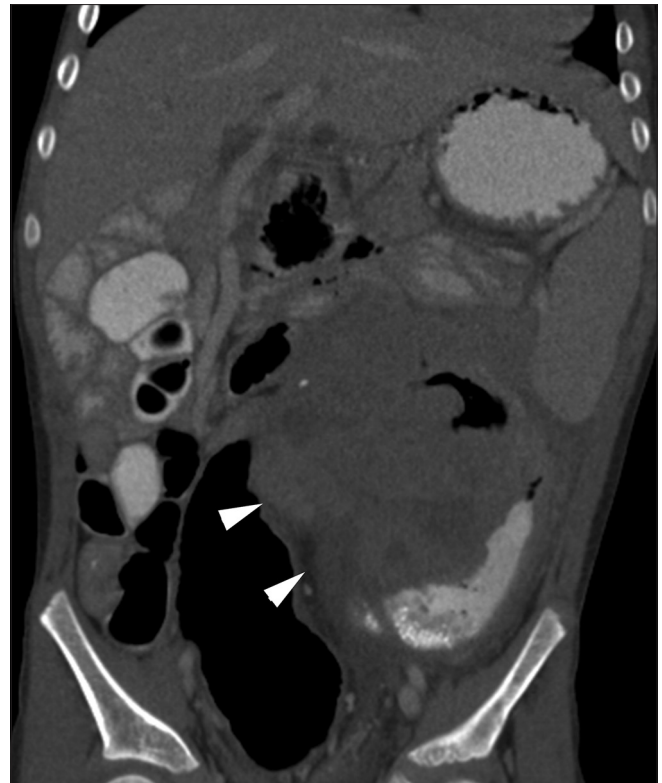
#### Post-transplant fluid collections

It is not unusual to have a small amount of ascites and small pleural effusions in immediate post-transplant patients, especially in those with MVT. Percutaneous drainage can be performed if ascites is moderate in size and causing patient discomfort. If it is large and reoccurs after drainage, further investigation for portal vein thrombosis should be performed.<sup>[12]</sup>

Other early postoperative collections include seromas or hematomas.<sup>[12]</sup> If these are small, they can be followed by serial US until resolution [Figure 16]. If they are large and cause small bowel compression or patient discomfort, then the drain can be placed with either US or CT guidance depending on their location and size. Fluid collections containing gas can develop if there is communication with bowel.<sup>[2]</sup> Fluoroscopic examination may help delineate a communication; however, CT with oral contrast better evaluates the anatomy as well as provides guidance for drainage. Air can also develop in a collection due to infection by gas-forming organisms. Percutaneous drainage is a preferred technique over surgery for managing abscesses of the abdomen and pelvis.<sup>[25]</sup> Diagnostic samples can also be obtained to identify the microorganism and its sensitivity to different antimicrobials.

#### Other transplant-associated complications and MVT organ dysfunction

Graft versus host disease (GVHD) occurs when immunocompetent donor lymphoid cells damage recipient tissues after allogeneic transplantation.<sup>[26]</sup> GVHD is a clinical diagnosis, wherein histopathology and immunocytology are used to confirm the disease.<sup>[26]</sup> The most common lesions of GVHD are on the skin and mucosal surfaces; therefore, clinical examination in transplant patients is vital.<sup>[2]</sup> GVHD is treated with steroids and tacrolimus immunosuppression.<sup>[26]</sup> The incidence of histologically



**Figure 15:** A 7-year-old boy who underwent LIT for intestinal failure secondary to gastroschisis, small bowel resections, and TPN-related liver disease. Coronal reformatted image from contrast-enhanced CT (CTDIvol 2.81 mGy, DLP 175.30 mGy cm) shows a hypodense mass (arrowheads) centered in the small bowel wall. Biopsy was consistent with PTLD

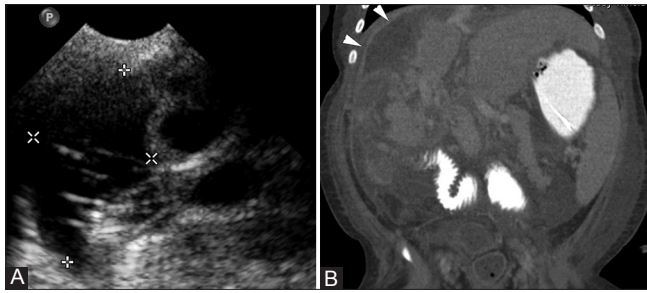
proven GVHD after clinical intestinal transplantation is 6.5% in children and 4.7% in adults.<sup>[26]</sup>

MVTs can result in a wide array of complications involving the transplant organs. Rejection can occur in almost any organ. The pancreas can develop inflammation, often including fluid collections and pseudocyst formation.<sup>[12]</sup> The liver can undergo fatty degeneration.<sup>[12]</sup> Traumatic denervation of the phrenic nerve during surgery can result in elevation of one or both hemidiaphragms.<sup>[21]</sup>

Patients on immunosuppression therapy after transplant with tacrolimus can develop posterior reversible encephalopathy syndrome (PRES), which is a small-vessel microangiopathy of the cerebral vasculature.<sup>[27,28]</sup> Many other neurological problems have been described in the pediatric population after combined liver and small bowel transplant, including seizures, encephalopathy, CNS infection, cerebrovascular accident, peripheral neuropathy, transient blurring of vision, auditory hallucinations, and choreoathetosis.<sup>[28]</sup>

#### Conclusions

Small bowel and multivisceral transplantations have become accepted treatments for patients with chronic intestinal failure.



**Figure 16 (A and B):** A 1-year-old boy who underwent size-reduced LIT for intestinal failure related to gastroschisis and small bowel atresia. (A) Postoperative transverse sonogram on day 1 after transplantation shows a complex right upper quadrant fluid collection (asterisk marks boundaries) (B) Contrast-enhanced CT 13 days after transplantation shows persistence of the collection and enhancement and thickening of the peritoneum (arrowheads)

Many patients are being transplanted with better outcomes due to advances in surgical techniques and improved postoperative management. Therefore, it is important for the radiologist to have a strong understanding of the indications, surgical anatomy, and potential postoperative complications associated with these procedures.

## References

- Kocoshis SA, Reyes J, Todo S, Starzl TE. Small intestinal transplantation for irreversible intestinal failure in children. *Dig Dis Sci* 1997;42:1997-2008.
- Godfrey EM, Upponi SS, See TC, Cheow HK, Sivaprakasam R, Butler AJ, *et al.* A radiologist's guide to small bowel and multivisceral transplantation. *Clin Radiol* 2013;68:983-91.
- Healthcare Systems Bureau, Division of Transplantation. Department of Health and Human Services. 2011 Annual Report of the U.S. Organ Procurement and Transplantation Network and the Scientific Registry of Transplant Recipients: Transplant Data 1997-2009. Rockville, MD: Health Resources and Services Administration; 2011.
- Watson AJ, Lear PA. Current status of intestinal transplantation. *Gut* 1989;30:1771-82.
- Cai J. Intestine and multivisceral transplantation in the United States: A report of 20-year national registry data (1990-2009). *Clin Transpl* 2009;83-101.
- Kelly DA. Liver complications of pediatric parenteral nutrition-epidemiology. *Nutrition* 1998;14:153-7.
- Phillips GS, Bhargava P, Stanescu L, Dick AA, Parnell SE. Pediatric intestinal transplantation: Normal radiographic appearance and complications. *Pediatr Radiol* 2011;41:1028-39.
- Unsinn KM, Koenigsrainer A, Rieger M, Czermak BV, Ellemunter H, Margreiter R, *et al.* Spectrum of imaging findings after intestinal, liver-intestinal, or multivisceral transplantation: Part 1, posttransplantation anatomy. *AJR Am J Roentgenol* 2004;183:1275-83.
- Cipriani R, Contedini F, Santoli M, Gelati C, Sgarzani R, Cucchetti A, *et al.* Abdominal wall transplantation with microsurgical technique. *Am J Transplant* 2007;7:1304-7.
- Benedetti E, Holterman M, Asolati M, Di Domenico S, Oberholzer J, Sankary H, *et al.* Living related segmental bowel transplantation: From experimental to standardized procedure. *Ann Surg* 2006;244:694-9.
- Panaro F, Testa G, Balakrishnan N, Rao B, Bogetti D, Jarzembowski T, *et al.* Living related small bowel transplantation in children: 3-dimensional computed tomography donor evaluation. *Pediatr Transplant* 2004;8:65-70.
- Unsinn KM, Koenigsrainer A, Rieger M, Czermak BV, Ellemunter H, Margreiter R, *et al.* Spectrum of imaging findings after intestinal, liver-intestinal, or multivisceral transplantation: Part 2, posttransplantation complications. *AJR Am J Roentgenol* 2004;183:1285-91.
- Coley BD. Pediatric applications of abdominal vascular Doppler: Part II. *Pediatr Radiol* 2004;34:772-86.
- Remotti H, Subramanian S, Martinez M, Kato T, Magid MS. Small-bowel allograft biopsies in the management of small-intestinal and multivisceral transplant recipients: Histopathologic review and clinical correlations. *Arch Pathol Lab Med* 2012;136:761-71.
- Lee BC, McGahan JP, Perez RV, Boone JM. The role of percutaneous biopsy in detection of pancreatic transplant rejection. *Clin Transplant* 2000;14:493-8.
- Kato T, Tzakis AG, Selvaggi G, Gaynor JJ, David AI, Bussotti A, *et al.* Intestinal and multivisceral transplantation in children. *Ann Surg* 2006;243:756-66.
- Culp WC, Vonk BN, McCowan TC, Sudan DL. Utilization and tailoring of interventional radiology procedures in small bowel transplants. *Transplant Proc* 2002;34:929-30.
- Furukawa A, Kanasaki S, Kono N, Wakamiya M, Tanaka T, Takahashi M, *et al.* CT diagnosis of acute mesenteric ischemia from various causes. *AJR Am J Roentgenol* 2009;192:408-16.
- Guaraldi G, Cocchi S, Codeluppi M, Di Benedetto F, De Ruvo N, Masetti M, *et al.* Outcome, incidence, and timing of infectious complications in small bowel and multivisceral organ transplantation patients. *Transplantation* 2005;80:1742-8.
- Frager DH, Baer JW, Rothpearl A, Bossart PA. Distinction between postoperative ileus and mechanical small-bowel obstruction: Value of CT compared with clinical and other radiographic findings. *AJR Am J Roentgenol* 1995;164:891-4.
- Reyes J, Tzakis AG, Todo S, Nour B, Starzl TE. Small bowel and liver/small bowel transplantation in children. *Semin Pediatr Surg* 1993;2:289-300.
- Macedo C, Sindhi R, Mazariegos GV, Abu-Elmagd K, Bond GJ, Reyes J. Sclerosing peritonitis after intestinal transplantation in children. *Pediatr Transplant* 2005;9:187-91.
- Nalesnik M, Jaffe R, Reyes J, Mazariegos G, Fung JJ, Starzl TE, Abu-Elmagd K. Posttransplant lymphoproliferative disorders in small bowel allograft recipients. *Transplant Proc* 2000;32:1213.
- Joo SH, Acun Z, Stefanovic A, Blieden CR, Ikpat OF, Moon J. Post-transplant lymphoproliferative disorder presented as small bowel intussusception in adult liver transplant patient. *J Korean Surg Soc* 2012;82:50-3.
- Cronin CG, Gervais DA, Castillo CF, Mueller PR, Arellano RS. Interventional radiology in the management of abdominal collections after distal pancreatectomy: A retrospective review. *AJR Am J Roentgenol* 2011;197:241-6.
- Mazariegos GV, Abu-Elmagd K, Jaffe R, Bond G, Sindhi R, Martin L, *et al.* Graft versus host disease in intestinal transplantation. *Am J Transplant* 2004;4:1459-65.
- Barbas AS, Rege AS, Castleberry AW, Gommer J, Ellis MJ, Brennan TV, *et al.* Posterior reversible encephalopathy syndrome independently associated with tacrolimus and sirolimus after multivisceral transplantation. *Am J Transplant* 2013;13:808-10.
- Fernandez D, El-Azzabi TI, Jain V, Lloyd C, Wassmer E, Peake D, *et al.* Neurologic problems after pediatric liver transplantation and combined liver and bowel transplantations: A single tertiary center experience. *Transplantation* 2010;90:319-24.

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