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## Superior mesenteric artery syndrome

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### INTRODUCTION

Superior mesenteric artery syndrome is an unusual cause of proximal intestinal obstruction. It has been referred to by a variety of other names, including Cast syndrome, Wilkie syndrome, arteriomesenteric duodenal obstruction, and chronic duodenal ileus [1,2]. The syndrome is characterized by compression of the third portion of the duodenum due to narrowing of the space between the superior mesenteric artery and aorta and is primarily attributed to loss of the intervening mesenteric fat pad.

There remains some controversy surrounding a diagnosis of superior mesenteric artery syndrome since symptoms do not always correlate well with abnormal anatomic findings on radiologic studies, and symptoms may not resolve completely following treatment [3,4]. Furthermore, the diagnosis may be confused with other anatomic or motility-related causes of duodenal obstruction [5].

The diagnosis and treatment of superior mesenteric artery syndrome will be reviewed here. The management of bowel obstruction is discussed elsewhere. (See "Etiologies, clinical manifestations, and diagnosis of mechanical small bowel obstruction in adults".)

### **ANATOMY**

The third portion of the duodenum passes between the aorta and the superior mesenteric artery ( figure 1 and image 1). The duodenum typically crosses anterior to the aorta at the level of the third lumbar vertebral body suspended by its attachment to the ligament of Treitz.

The superior mesenteric artery arises from the anterior aspect of the aorta at the level of the L1 vertebral body. It is enveloped in fatty and lymphatic tissue and extends in a caudal direction at an acute angle into the mesentery. In the majority of patients, the normal angle between the superior mesenteric artery and the aorta is between 38 and 65° due, in part, to the mesenteric fat pad [6]. This angle correlates with body mass index [7]. The aortomesenteric distance is normally 10 to 28 mm [8].

In superior mesenteric artery syndrome, the angle can be narrowed to as low as 6° with the aortomesenteric distances as low as 2 mm, which minimizes the space between the superior mesenteric artery and aorta, potentially leading to duodenal compression ( figure 1). The left renal vein may also be compressed, which can result in nutcracker syndrome [9]. (See "Etiology and evaluation of hematuria in adults", section on 'Rare conditions'.)

### **PATIENTS AT RISK**

Several factors can decrease the angle between the aorta and superior mesenteric artery. The most common is significant weight loss leading to loss of the mesenteric fat pad as a consequence of medical disorders, psychological disorders, or surgery. Anatomic abnormalities (congenital or acquired) can also contribute.

Superior mesenteric artery syndrome in adults is most commonly associated with severe, debilitating illnesses, such as malignancy, malabsorption syndromes, AIDS [10-12], trauma [13], and burns [14]. It has also been described in a variety of other disorders associated with extreme weight loss, including bariatric surgery [15-18], spinal cord injury, paraplegia [19], drug abuse [20], prolonged bed rest, and anorexia nervosa [21-24].

Weight loss is not responsible for all cases. An interesting case report suggests that weight loss is not always necessary for the diagnosis of superior mesenteric artery syndrome in the pediatric population and that it can occur in otherwise healthy adolescents after insufficient weight gain relative to height growth [25]. This idea that weight loss is not necessary for the development of SMA syndrome in pediatric population has also been noted in other reviews [26-28]. In a series of 22 pediatric cases, an absence of weight loss was noted in 50 percent of children [26].

In younger patients, superior mesenteric artery syndrome is most commonly described following corrective spinal surgery for scoliosis [8,29-32]. This procedure lengthens the spine cranially, displacing the superior mesenteric artery origin, decreasing the mesenteric artery's lateral mobility, and reducing the aortomesenteric angle [33]. Under this circumstance, it may be referred to as "cast syndrome." Other surgeries may also be complicated by the development of superior mesenteric artery syndrome due to distortion of normal anatomy, such as esophagectomy [34].

Rarely, a patient may have a congenitally short ligament of Treitz suspending the duodenum in an abnormally cephalad position. A case report of identical twins with this disorder [35] and another case diagnosed in utero [36] suggest there may be a genetic predisposition in some patients. The origin of the superior mesenteric artery may also be abnormally low.

There have been several reported cases of superior mesenteric artery syndrome coexisting with celiac axis compression syndrome. One report documented a patient with celiac axis compression, which was believed to be the cause of superior mesenteric artery syndrome [37]. (See "Celiac artery compression syndrome".)

There have also been cases of superior mesenteric artery syndrome reported in patients with nutcracker syndrome [38-41]. (See "Evaluation of microscopic hematuria in children", section on 'Nutcracker syndrome' and "Etiology and evaluation of hematuria in adults".)

#### **CLINICAL EVALUATION**

Patients may present acutely (such as following surgery) or more insidiously with progressive symptoms [3]. In both cases, symptoms are consistent with proximal small bowel obstruction. Patients with mild obstruction may have only postprandial epigastric pain and early satiety, while those with more advanced obstruction may have severe nausea, bilious emesis, and weight loss. Patients may also have symptoms of reflux. (See "Approach to the adult with nausea and vomiting".)

Symptoms may be relieved when patient is lying prone, in the left lateral decubitus, or in a kneechest position [2]. These positions remove tension from the mesentery and superior mesenteric artery opening the space between the superior mesenteric artery and aorta.

Findings on physical examination are nonspecific but can include abdominal distension, a succussion splash, and high-pitched bowel sounds. Laboratory examination can be normal, or, in patients with severe vomiting, significant electrolyte abnormalities may be present. (See "Causes of metabolic alkalosis", section on 'Gastrointestinal hydrogen loss'.)

Diagnosis is often delayed and may result in significant complications, including [42,43]:

- Fatalities due to electrolyte abnormalities
- Fatalities due to gastric perforation
- Gastric pneumatosis and portal venous gas
- Formation of an obstructing duodenal bezoar

**Differential diagnosis** — The differential diagnosis of superior mesenteric artery syndrome includes other causes of bowel obstruction as well as diseases associated with duodenal dysmotility (and "megaduodenum"), including diabetes mellitus, collagen vascular diseases, scleroderma, and chronic idiopathic intestinal pseudo-obstruction [44]. (See "Etiologies, clinical manifestations, and diagnosis of mechanical small bowel obstruction in adults".)

Additional causes of dyspepsia and reflux should be considered in patients with nonspecific symptoms and equivocal radiographic findings. (See "Approach to the adult with dyspepsia" and "Clinical manifestations and diagnosis of gastroesophageal reflux in adults".)

Patients with a significant smoking history or other risk factors for atherosclerosis who have symptoms of chronic food intolerance associated with weight loss are more likely to have chronic mesenteric ischemia, a condition that is unlikely to be confused with superior mesenteric artery syndrome. Patients with chronic mesenteric ischemia usually have reproducible pain with eating and will not have obstructive symptoms. (See "Chronic mesenteric ischemia".)

## **DIAGNOSTIC STUDIES**

A high index of suspicion is required since symptoms can be nonspecific. A diagnosis of superior mesenteric artery syndrome is often a diagnosis of exclusion. If not already performed, patients should undergo judicious testing for other disorders that can cause similar symptoms. (See "Etiologies, clinical manifestations, and diagnosis of mechanical small bowel obstruction in adults".)

The diagnostic evaluation begins with plain abdominal films and radiography with oral contrast. These studies may be diagnostic in patients for whom there is heightened suspicion based upon the clinical setting. Additional testing may be needed for those patients in whom the diagnosis remains unclear.

**Plain abdominal films** — Diagnostic evaluation should begin with abdominal radiographs. Although plain films are frequently nonspecific, they may reveal findings suggestive of proximal

small bowel obstruction, such as gastric distension, dilation of the proximal duodenum, and, occasionally, an abrupt vertical cutoff of air in the third portion of the duodenum.

**Oral contrast studies** — Upper gastrointestinal series usually demonstrate marked delay in passage of the contrast from the duodenum into the more distal small bowel ( image 2A-B) [3]. Passage of contrast typically halts abruptly at the third portion of the duodenum. The proximal duodenum and stomach are dilated and show prolonged retention of barium. Similar findings can be seen with computed tomography (CT).

- Ultrasound A noninvasive method to evaluate the mesenteric artery anatomy is using transabdominal ultrasound, which can identify and measure the aortomesenteric angle ( image 3). Positional maneuvers not typically performed with routine examination may identify alterations in the aortomesenteric angle when the patient is in the lateral decubitus position or even standing [45,46]. The advantage of ultrasound is its low cost. Endoscopic ultrasound has also been used to demonstrate the anatomic abnormalities associated with superior mesenteric artery syndrome [47].
- Arteriography Conventional arteriography was traditionally performed simultaneously
  with barium contrast radiography to demonstrate the superior mesenteric artery
  superimposed upon the barium-filled duodenum. Lateral mesenteric arteriography
  demonstrates the reduction of the aortomesenteric angle ( image 1).

CT and magnetic resonance (MR) arteriography have largely replaced conventional arteriography since they are noninvasive and provide additional anatomic detail, such as the amount of intra-abdominal and retroperitoneal fat ( image 4A-B) [48,49]. However, arteriography may be needed if a diagnosis remains unclear.

**Diagnostic imaging criteria** — As a general rule, the following criteria should be present on imaging [3,45,46]:

- Duodenal obstruction with an abrupt cutoff in the third portion and active peristalsis.
- An aortomesenteric artery angle of ≤25° is the most sensitive measure of diagnosis, particularly if the aortomesenteric distance is ≤8 mm [45,46].
- High fixation of the duodenum by the ligament of Treitz, abnormally low origin of the superior mesenteric artery, or anomalies of the superior mesenteric artery.

#### CONSERVATIVE THERAPY

The goals of conservative treatment of superior mesenteric artery syndrome are alleviation of obstructive symptoms and reversal of any precipitating factors. As noted above, weight loss is commonly associated. However, if surgery has altered the anatomy, the likelihood that conservative therapy will be successful is low.

**Gastrointestinal decompression** — All patients should have a nasogastric tube placed and correction of electrolyte abnormalities. A nasogastric tube decompresses the dilated stomach and proximal duodenum, improving patient comfort and aiding in monitoring fluid losses. (See "Inpatient placement and management of nasogastric and nasoenteric tubes in adults".)

**Correction of electrolyte abnormalities** — Patients who are acutely ill may have electrolyte abnormalities, which should be monitored and corrected aggressively. Emesis can lead to hypovolemia, hypokalemia, and metabolic alkalosis. Fluid and electrolyte repletion should be initiated. (See "Treatment of metabolic alkalosis".)

Following relief of duodenal obstruction, refeeding syndrome can contribute to electrolyte abnormalities. (See "Anorexia nervosa in adults and adolescents: The refeeding syndrome".)

**Nutrition support** — The major component of therapy is nutrition support. Psychiatric evaluation may be needed to help manage an eating disorder. (See "Eating disorders: Overview of prevention and treatment".)

Nutrition support is usually required, at least in the initial stages until patients are able (and willing) to increase oral intake. Enteral nutrition is preferred and often administered through a nasojejunal feeding tube placed distal to the obstruction. Parenteral nutrition may be necessary if enteral feeding is not an option [50]. Once significant weight gain is noted, the diet may be advanced slowly.

In adults who have a brief history of symptoms and in children who tend to present acutely, conservative management with nutrition support has good success [14,26]. The adult patient with more chronic symptoms is less likely to benefit from nutrition support alone. For these patients, correction of electrolytes and decompression with a short course of nutrition should be followed soon by surgical management, if symptoms are not relieved [9].

## SURGICAL MANAGEMENT

There are several surgical options for the treatment of superior mesenteric artery syndrome if conservative therapy fails to resolve the condition. These include Strong's procedure

( figure 2), gastrojejunostomy, and duodenojejunostomy with or without division or resection of the fourth part of the duodenum.

Prior to surgery, the patient's nutritional status should be reevaluated to ensure adequacy of wound healing. A multidisciplinary approach including dieticians and psychiatry consultation should be used to ensure the patient's well-being before and after surgery.

## **OPEN SURGERY**

- Strong's procedure Strong's procedure mobilizes the duodenum by dividing the ligament of Treitz [51]. Once the duodenal-jejunal junction has been fully mobilized, the duodenum is positioned to the right of the superior mesenteric artery so it does not lie within the space between the aorta and the superior mesenteric artery [52]. The integrity of the bowel is maintained; however, the short branches of the inferior pancreaticoduodenal artery may limit the ability of the duodenum to fall away from the aorta.
- Gastrojejunostomy Gastrojejunostomy is performed by bringing a loop of jejunum up to the stomach and performing a side-to-side anastomosis ( figure 3).
- Duodenojejunostomy With duodenojejunostomy, the duodenum can be left intact or divided and the proximal jejunum brought through the right mesocolon to perform a side-to-side duodenojejunostomy ( figure 4).

**Laparoscopic approach** — Successful laparoscopic duodenojejunal bypass has also been described. Although experience is limited, it offers a less invasive surgical option [53-55]. Advanced laparoscopic techniques have been described for the Strong procedures and duodenojejunostomy [54,56].

**Follow-up** — Contrast studies are performed at one to two weeks postoperatively to demonstrate patency of the repair and normal emptying of the duodenum. Patients are followed for resolution of their preoperative symptoms, and weight gain is monitored.

**Outcomes and complications of surgery** — There are few reports of long-term outcomes in patients with superior mesenteric artery syndrome who have undergone surgery. One of the largest series included 16 patients who were followed seven years after surgery [4]. Weight loss had been corrected in all patients. However, symptoms were essentially unchanged with the exception of vomiting, which was significantly decreased. In another series of eight patients, symptoms were improved; however, significant weight gain was not seen [9].

Each of the surgical approaches has advantages and disadvantages. Strong's procedure maintains the integrity of the bowel; however, failure occurs in up to one fourth of patients. Gastrojejunostomy decompresses the stomach, but the failure to relieve the duodenal obstruction may result in recurrent symptoms requiring a second procedure, and the unrelieved obstruction may result in blind loop syndromes or peptic ulceration. Duodenojejunostomy is generally accepted as having superior results to both Strong's procedure and gastroenterostomy. Duodenojejunostomy with division of the fourth part of the duodenum establishes bowel continuity and minimizes the issues associated with a blind loop [4].

Some authors resect the third portion of the duodenum altogether. The rationale for this practice is the feeling that superior mesenteric artery syndrome is a variant of a motility disorder rather than a true mechanical obstruction. However, there is little evidence, either pathological or physiologic, to support this notion [9]. Following relief of obstruction, normal duodenal motility can be demonstrated with gastrointestinal contrast studies. (See 'Follow-up' above.)

## SUMMARY AND RECOMMENDATIONS

- Anatomy and clinical features Superior mesenteric artery (SMA) syndrome is characterized by symptoms of proximal small bowel obstruction due to compression of the third portion of the duodenum related to a decreased angle between the aorta and superior mesenteric artery. There remains some controversy surrounding SMA syndrome, since symptoms do not always correlate well with radiologic findings and do not always improve following treatment. Symptoms can be confused with other anatomic or motilityrelated causes of duodenal obstruction. (See 'Anatomy' above and 'Clinical evaluation' above.)
- **Risk factors** The most common risk factor for SMA syndrome is significant weight loss because of medical disorders, psychological disorders, or surgery. Anatomic abnormalities (congenital or acquired) can also contribute. (See 'Patients at risk' above.)
- **Diagnosis** A diagnosis can be challenging because SMA syndrome is uncommon, and symptoms can be nonspecific. Thus, a high index of suspicion is required in the appropriate clinical setting. The diagnosis is established in patients with clinical features that suggest duodenal obstruction and noninvasive imaging that demonstrates an abnormal angle between the aorta and the superior mesenteric artery. Patients should undergo judicious testing for other disorders that can cause similar clinical features.

Conventional arteriography may be needed for those patients in whom the diagnosis remains unclear. (See 'Diagnostic studies' above.)

- **Conservative management** Patients may initially require gastric decompression, fluid resuscitation, and correction of electrolyte abnormalities. We suggest reversal of weight loss as initial therapy for superior mesenteric artery syndrome (**Grade 2C**). This generally requires enteral nutrition support, but some patients may require parenteral nutrition. (See 'Conservative therapy' above.)
- **Surgical management** For patients diagnosed with superior mesenteric artery syndrome whose condition is not improved with conservative management, several surgical options are available, including mobilization of the ligament of Treitz, gastrojejunostomy, and duodenojejunostomy. Of these, duodenojejunostomy may provide the best results. (See 'Surgical management' above.)

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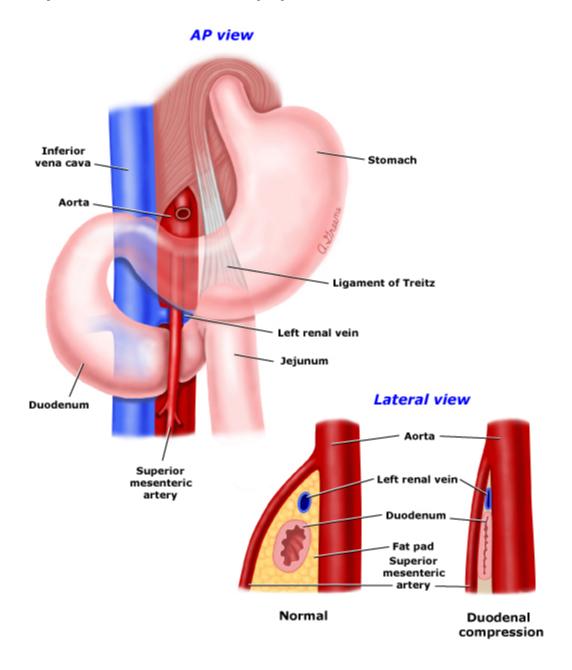
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#### **GRAPHICS**

## Superior mesenteric artery syndrome



The superior mesenteric artery arises from the anterior aspect of the aorta at the level of the L1 vertebral body. It is enveloped in fatty and lymphatic tissue and extends in a caudal direction at an acute angle into the mesentery. In the majority of patients, the normal angle between the superior mesenteric artery and the aorta is between 38 and 65 degrees. Superior mesenteric artery syndrome is characterized by compression of the third portion of the duodenum due to narrowing of the space between the superior mesenteric artery and aorta and is primarily attributed to loss of the intervening mesenteric fat

pad. With superior mesenteric artery syndrome, the angle between the superior mesenteric artery and the aorta can be narrowed to as little as 6 degrees.

Graphic 74899 Version 4.0

# A lateral arteriogram in a patient with SMA Syndrome



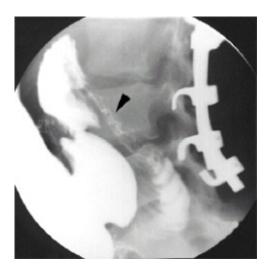
The angle between the aorta (open arrowhead) and the SMA (closed arrowhead) is reduced causing compression of the third portion of the duodenum.

SMA: superior mesenteric artery.

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Graphic 61740 Version 3.0

## **Upper gastrointestinal contrast study**

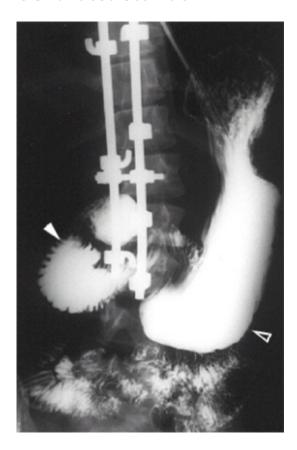


This upper gastrointestinal contrast study (UGI) in an oblique projection demonstrates filling of the stomach and proximal duodenum with contrast (left side of photograph). Distal to this (dark arrowhead), there is compression of the third portion of the duodenum with only a scant amount of contrast in this region. This patient had repair of his scoliosis as evidenced by the hardware (right side of photograph).

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## **UGI** dilated stomach

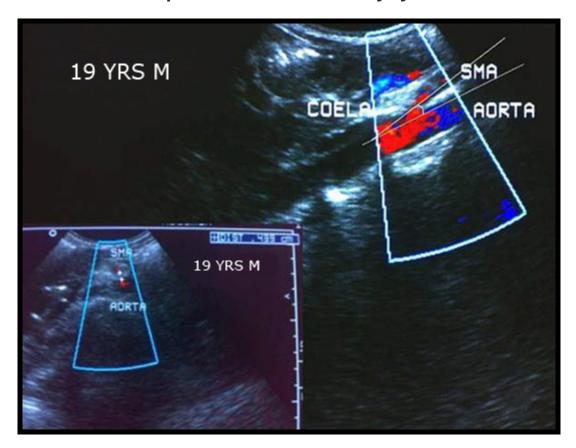


This is the UGI study on the same patient demonstrating the dilated stomach (open arrowhead) and a dilated proximal duodenum (closed arrowhead).

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## Ultrasound in superior mesenteric artery syndrome



Sagittal ultrasonography image shows SMA-aortic angle of 12° (white constructed angle). A normal angle should be between 38 and 65°.

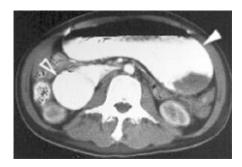
Inset: Transverse ultrasonography image showing SMA-aorta distance of 4.4 mm.

SMA: superior mesenteric artery; COELA: celiac artery.

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# CT scan of the abdomen in a patient with SMA Syndrome



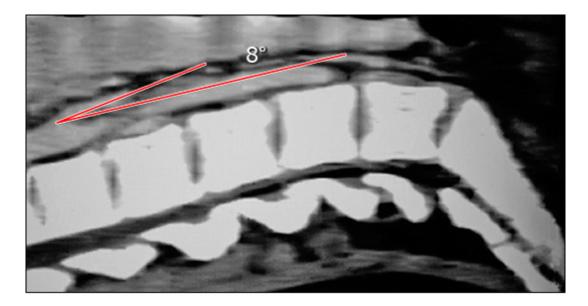
There is a distended, contrast-filled stomach (closed arrowhead) and a dilated, contrast-filled proximal duodenum (open arrowhead). The duodenum can be seen narrowing as it approaches the aorta where it is compressed between the aorta and the SMA.

CT: computed tomography; SMA: superior mesenteric artery.

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# Abdominal CT scan showing superior mesenteric artery syndrome



Computed tomography of the abdomen with intravenous contrast showing a narrowed angle between the superior mesenteric artery and the aorta measuring eight degrees.

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Graphic 76655 Version 2.0

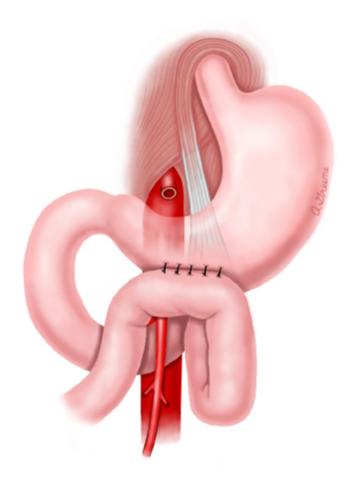
## **Strong procedure**



Strong's procedure mobilizes the duodenum by dividing the ligament of Treitz. Once the duodenal-jejunal junction is mobilized, the duodenum is positioned to the right of the superior mesenteric artery.

Graphic 74196 Version 2.0

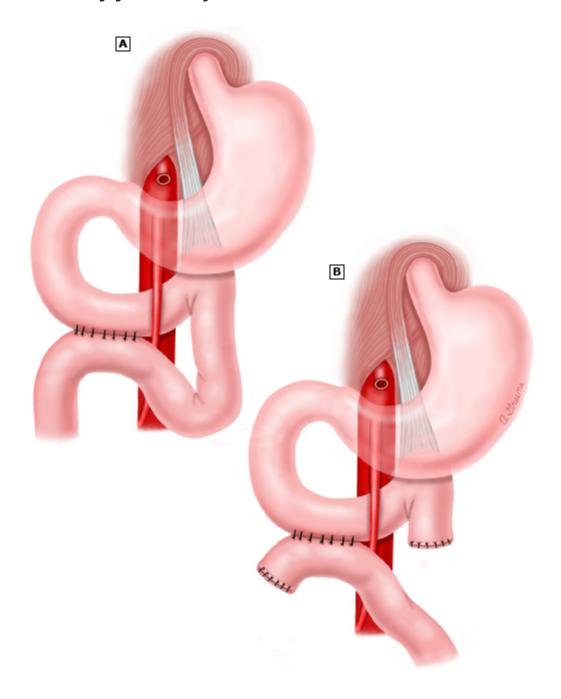
## Gastrojejunostomy



A gastrojejunostomy can be fashioned to bypass an obstructed segment of duodenum using sutures or staples.

Graphic 53190 Version 3.0

## Duodenojejunostomy



Duodenojejunostomy is accomplished without (A) or with (B) division of the 4th portion of the duodenum.

Graphic 66614 Version 2.0

#### **Contributor Disclosures**

Sherry Scovell, MD, FACS No relevant financial relationship(s) with ineligible companies to disclose. Allen Hamdan, MD No relevant financial relationship(s) with ineligible companies to disclose. J Thomas Lamont, MD Equity Ownership/Stock Options: Allurion [Weight loss]. Consultant/Advisory Boards: Teledoc [Gastrointestinal diseases]. All of the relevant financial relationships listed have been mitigated. John F Eidt, MD Grant/Research/Clinical Trial Support: Syntactx [Clinical events and data/safety monitoring for medical device trials]. All of the relevant financial relationships listed have been mitigated. Joseph L Mills, Sr, MD No relevant financial relationship(s) with ineligible companies to disclose. Kathryn A Collins, MD, PhD, FACS No relevant financial relationship(s) with ineligible companies to disclose.

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