

# Resolution of Refractory Superior Mesenteric Artery Syndrome with Laparoscopic Duodenojejunostomy: Pediatric Case Series with Spectrum of Clinical Imaging

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## ABSTRACT

**Background:** Superior mesenteric artery (SMA) syndrome is an uncommon condition resulting in partial small bowel obstruction because of external compression of the third portion of the duodenum between the SMA anteriorly and the aorta posteriorly. SMA syndrome often presents with postprandial nausea, bilious vomiting, and abdominal pain with associated weight loss. Onset of symptoms can be acute (occurring in the setting of rapid weight loss because of trauma/surgery) or can be vague and chronic over many years.

**Case Reports:** We present two cases of female adolescents who presented with symptoms of duodenal obstruction attributed to SMA syndrome. Both failed conservative treatment with weight gain and underwent successful laparoscopic duodenojejunostomy procedures with resolution of duodenal obstruction.

**Conclusion:** In the differential diagnosis of persistent nausea and bilious vomiting, even in the setting of an eating disorder, SMA syndrome should be considered. Upper gastrointestinal examination is the primary modality for diagnosing SMA syndrome, but ultrasound is an inexpensive, rapid screening tool for patients with unexplained abdominal pain. Abdominal computed tomography may also be helpful in selected patients.

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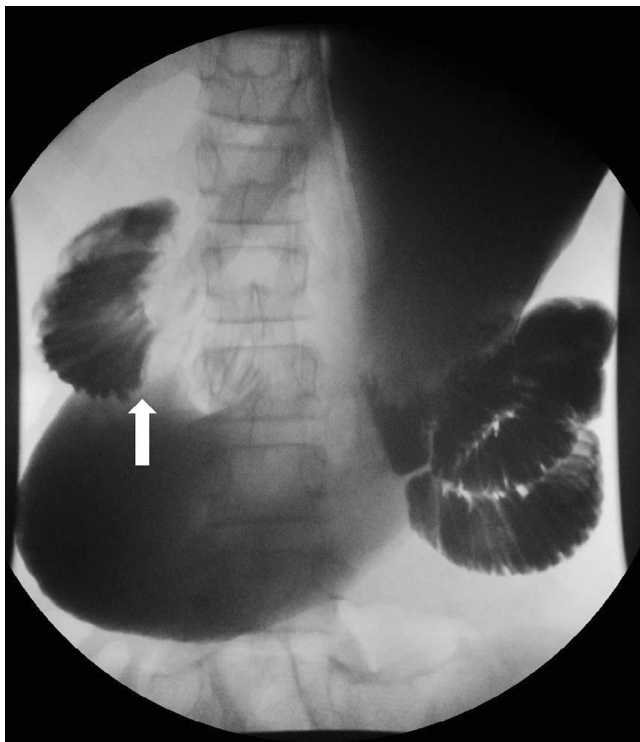
Conservative therapy consisting of nutritional support to enhance weight gain is usually sufficient and is accomplished with placement of a nasojejunal feeding tube past the point of duodenal compression. When conservative therapy fails, laparoscopic duodenojejunostomy can provide definitive relief of the obstruction.

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## INTRODUCTION

Superior mesenteric artery (SMA) syndrome is an uncommon condition resulting from partial small bowel obstruction because of external compression of the third portion of the duodenum between the SMA anteriorly and the aorta posteriorly.<sup>1,2</sup> The aortomesenteric angle is narrowed, typically because of loss of the mesenteric fat pad in the setting of rapid weight loss, scoliosis surgery, or a high insertion of the ligament of Treitz.<sup>3,4</sup> SMA syndrome often presents with postprandial nausea, bilious vomiting, and abdominal pain with associated weight loss. Onset of symptoms can be acute (occurring in the setting of rapid weight loss because of trauma/surgery) or can be vague and chronic over many years.

Wilkie published the first case series analyzing the condition in 1927.<sup>5</sup> Since then, there has been controversy in the medical literature regarding whether SMA syndrome is an actual entity or the result of other gastrointestinal pathology.<sup>6-8</sup> Symptoms do not always correlate with imaging and do not necessarily resolve with treatment.<sup>9,10</sup> Additionally, because of the rarity of this condition and its overlap with numerous other gastrointestinal pathologies, patients with SMA syndrome can easily be misdiagnosed, especially those with chronic cases. Advances in medical imaging, however, now allow physicians to more clearly define this process through better visualization of the anatomy and the ability to measure the aortomesenteric angle and distance between the SMA and the aorta. Typical aortomesenteric angles



**Figure 1.** Upper gastrointestinal examination with barium obtained on initial presentation in case 1 revealed a massive enlargement of the stomach and proximal duodenum with abrupt cutoff point as the duodenum crosses the midline (arrow). Contrast did not pass through the horizontal portion of the duodenum while the patient was supine; however, contrast easily passed through with the patient in the left lateral decubitus or prone positions.

are between  $38^{\circ}$  and  $65^{\circ}$ , whereas in patients with SMA syndrome this angle is significantly reduced, ranging from  $6^{\circ}$  to  $25^{\circ}$ .<sup>11-13</sup>

Conservative treatment with weight gain is the preferred management of SMA syndrome.<sup>10,14</sup> If patients are unable to tolerate oral intake because of the severity of their symptoms, weight gain can be achieved with nasojejun tube feeding or, if needed, total parenteral nutrition (TPN). The goal is to increase the mesenteric fat pad and thus enlarge the aortomesenteric angle, relieving duodenal compression. For patients in whom weight gain is not sufficient to resolve their symptoms, surgery is an option. The most conservative surgical treatment described in the literature is the Strong procedure that involves mobilization of the fourth portion of the duodenum by mobilization of the ligament of Treitz.<sup>3</sup> Unfortunately, the failure rate for this procedure is high.<sup>15</sup> Duodenojejunostomy has proven to be a more successful approach for these patients. In 1998, Gersin and Heniford reported the first case of laparoscopic duodenojejunostomy for the treatment

of SMA syndrome.<sup>16</sup> Because laparoscopic duodenojejunostomy is minimally invasive and can provide definitive treatment, this approach is becoming the preferred treatment when conservative therapy fails.

We present the cases of two female adolescents who presented with symptoms of duodenal obstruction attributed to SMA syndrome and failed conservative treatment with weight gain. Because of the refractory nature of their symptoms, the patients were evaluated for surgical intervention. Both patients underwent successful laparoscopic duodenojejunostomy procedures with resolution of duodenal obstruction. We present these cases to highlight the patients' clinical imaging and the successful management of their uncommon condition with laparoscopic surgery.

### CASE 1

A 13-year-old female of European descent with a 6-day history of midabdominal pain and epigastric distention that progressed to postprandial nonbilious vomiting and inability to tolerate oral intake presented at our facility. Her symptoms improved with postural change to the knee-to-chest position. She was underweight at approximately 37 kg at the time of initial evaluation, but her weight loss was presumed to be secondary to her symptoms. Fluoroscopic upper gastrointestinal examination revealed massive dilation of the stomach and proximal duodenum. Contrast passed easily into the proximal duodenum but abruptly stopped in the third portion of the duodenum prior to crossing the midline (Figure 1). Contrast eventually passed through this area when the patient was placed in the knee-to-chest position. SMA syndrome was suspected, and because of the severity of the patient's illness at presentation, she was admitted to the pediatric surgery service.

A nasogastric tube was placed to decompress her stomach. Computed tomography (CT) of the abdomen and pelvis revealed a massively distended stomach spanning from the diaphragm into the pelvis. The proximal duodenum was distended and came to an abrupt taper in the third portion that appeared to be compressed by the SMA (Figure 2). She had significant mass effect from her dilated stomach and duodenum on her other abdominal organs, including hydronephrosis of the left kidney. Because of obscuration by massive gastroduodenomegaly, the exact aortomesenteric angle was difficult to measure but was approximately  $15^{\circ}$  (Figure 3). She required intravenous hydration, TPN, and electrolyte replacement. Her nausea, vomiting, and abdominal pain improved after nasogastric tube placement, and she gained weight with TPN.

Her symptoms were initially controlled with prolonged nutritional support, and during the next year she had minimal symptoms but required hospitaliza-



**Figure 2. Coronal computed tomography image demonstrates the massively distended esophagus, stomach, and proximal duodenum in case 1. There is an abrupt cutoff point of the duodenum just prior to it crossing the midline (white arrow) at the level of the superior mesenteric artery (black arrow). The distal bowel is collapsed and compressed.**

tion twice for exacerbation of symptoms. Repeat imaging was obtained prior to the second admission, and both abdominal CT and upper gastrointestinal examination revealed a massively distended stomach and proximal duodenum. The patient was admitted, and a nasojejun tube was placed. Because of the worsening severity of her symptoms and the failure of conservative treatment, surgical options were discussed with the patient and her parents. They decided to proceed with laparoscopic duodenojejunostomy. The patient's postoperative course was complicated by a self-limited upper gastrointestinal bleed; however, she did not require transfusion. She was discharged on postoperative day 9 and at the time of discharge was tolerating a regular diet without symptoms. At her 1-month follow-up in the surgery



**Figure 3. Sagittal computed tomography image shows the narrowed aortomesenteric angle (arrow) in case 1.**

clinic, she was still tolerating a regular diet without any nausea or vomiting. At a well-child visit approximately 1 year after surgery, she reported doing well with no new issues or complaints.

## CASE 2

A 16-year-old female of European descent with a 2-year history of postprandial nausea, abdominal pain, and bilious vomiting presented at our facility. She was on continuous nasojejun tube feeds. She had initially been diagnosed with SMA syndrome 2 years prior and had undergone conservative therapy with an attempt at weight gain to resolve her symptoms. Her weight varied during this time from 33-47 kg, and she had no resolution of her symptoms. She was diagnosed with anorexia nervosa and spent time in an eating disorder facility with no improvement of her symptoms. She presented seeking surgical options because she had failed 2 years of conservative therapy.

The patient's nasojejun tube was removed, and an upper endoscopy was performed to evaluate for an intraluminal cause of obstruction. The observed esophagus and stomach were within normal limits; however, the third portion of the duodenum exhibited a luminal deformity and narrowing consistent with extrinsic compression (Figure 4). The patient then underwent an upper gastrointestinal examination with barium. With the patient supine, contrast initially passed without difficulty from the esophagus into the stomach and first and second portions of the duodenum; however, antiperistaltic flow of contrast was observed refluxing from the first portion of the



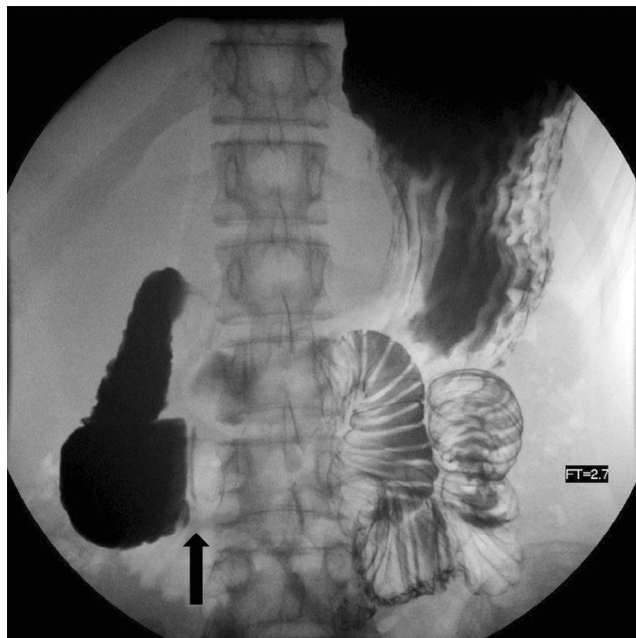
**Figure 4.** Endoscopic view shows deformation and narrowing of the third portion of the duodenum in case 2. No etiology of obstruction intrinsic to the bowel was identified on this examination.

duodenum into the stomach. Fluoroscopic images revealed an abrupt vertical line across the third portion of the duodenum just proximal to the ligament of Treitz consistent with an external compression (Figure 5). A small amount of contrast was able to travel distally to this obstruction. Given her constellation of symptoms and the results of the upper gastrointestinal examination, the etiology of these findings and the patient's symptoms was considered to be most likely SMA syndrome.

The patient underwent a laparoscopic duodeno-jejunosotomy to bypass the compression, and she did well in the postoperative period. On postoperative day 5, her nasogastric tube was clamped and removed, and her diet was slowly advanced. On postoperative day 6, the patient tolerated a regular consistency diet without difficulty, and she was considered safe to discharge home. At the time of her postoperative follow-up appointment 2 weeks after discharge, she continued to do well without any nausea or vomiting, and her weight was stable from discharge.

## DISCUSSION

In patients with signs and symptoms of mechanical small bowel obstructions, plain abdominal radiographs are the initial study of choice. In patients with middistal small bowel obstruction, plain radiographs are often diagnostic. In duodenal and other proximal small bowel obstructions, findings are generally nonspecific and may include a dilated stomach and proximal duodenum with air/fluid levels. If the index of suspicion for SMA syndrome or other proximal small bowel obstruction is high, further evaluation with



**Figure 5.** Upper gastrointestinal examination with barium demonstrates distention of the stomach and proximal duodenum and a sharp vertical line across the third portion of the duodenum (arrow) consistent with extrinsic compression in case 2. During this examination, there was antiperistaltic reflux of contrast into the stomach.

fluoroscopic upper gastrointestinal series examination can be obtained. In patients with SMA syndrome, imaging will show an abrupt cutoff in the third portion of the duodenum just prior to crossing the spine. Antiperistaltic reflux of contrast from the proximal duodenum into the stomach can be associated with this obstruction. During this dynamic study series, the patient can be placed prone, knee-to-chest, or in the left lateral decubitus position to partially relieve obstruction and further confirm diagnosis.

If the diagnosis is unclear from the upper gastrointestinal series examination or if other underlying etiology is suspected, contrast-enhanced CT with multiplanar reformats can be obtained, allowing measurement of the aortomesenteric angle and distance in the sagittal plane. The aortomesenteric angle in patients with SMA syndrome is reduced to  $6^{\circ}$  to  $25^{\circ}$  with the aortomesenteric distance  $<8$  mm.<sup>11-13</sup> Ultrasonography with Doppler can also be used to evaluate the aortomesenteric angle and distance; such screening results correlate well with CT findings. Because of its low cost and wide availability, ultrasound may prove to be a beneficial screening tool in patients with abdominal pain of unclear etiology.<sup>17</sup> Direct visualization of the duodenum with endoscopy can also be used to evaluate for a mechanical intraluminal cause of obstruction in

conjunction with other studies prior to proceeding with surgical therapy in unclear cases.<sup>10,18,19</sup>

Patients with SMA syndrome in whom medical management fails can have the obstruction relieved by surgical bypass of the compressed duodenum. Several cases in the literature report laparoscopic duodenojejunostomy being used for SMA syndrome. The first, by Richardson and Surowiec in 2001, reported two patients who were treated effectively, had short hospital stays, and had no significant complications.<sup>20</sup> The technique used in these two patients involved initial placement of a 12 mm trocar in the umbilicus using open technique and a Hasson trocar, followed by the placement of 5 mm trocars in both lower quadrants and in the suprapubic region. A 5 mm telescope was introduced through the suprapubic port for the remainder of the procedure, allowing introduction of the endoscopic stapler through the umbilical port for the side-to-side duodenojejunal anastomosis.

## CONCLUSION

Our two cases highlight the importance of recognizing and treating SMA syndrome. Although often a diagnosis of exclusion, SMA syndrome should be considered in the differential diagnosis of persistent nausea and bilious vomiting, even in the setting of an eating disorder. The presence of bilious emesis should always raise the concern of a mechanical component. Upper gastrointestinal examination is the primary modality for diagnosing SMA syndrome, but ultrasound is an inexpensive, rapid screening tool for patients with unexplained abdominal pain. Abdominal CT may be helpful in selected patients.

Conservative therapy consisting of nutritional support to enhance weight gain is usually sufficient and accomplished with placement of a nasojejunal feeding tube past the point of duodenal compression. When conservative therapy fails, laparoscopic duodenojejunostomy can provide definitive relief of the obstruction.

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