Surg Endosc (2017) 31:1180–1185
DOI 10.1007/s00464-016-5088-2

Laparoscopic duodenojejunostomy for superior mesenteric artery syndrome: intermediate follow-up results and a review of the literature

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Received: 15 March 2016 / Accepted: 5 July 2016 / Published online: 12 July 2016

Abstract
Background Superior mesenteric artery syndrome (SMAS) is a rare condition caused by partial obstruction of the third portion of the duodenum by the SMA anteriorly and aorta posteriorly. Laparoscopic duodenojejunostomy has been described as a safe and feasible surgical intervention with favorable short-term outcomes. However, descriptions of intermediate outcomes are lacking in the literature.
Methods A retrospective chart review was performed on patients who underwent minimally invasive duodenojejunostomy from March 2005 to August 2015 at our healthcare system with greater than 6-month follow-up.
Results Eighteen patients with mean age of 31.2 were identified. There were 4 men and 14 women. Patients' diagnosis was made by clinical presentation with radiographic confirmation. Mean weight loss preoperatively was 13.9 kg, representing 24.1 % total body weight loss. There were no intraoperative complications. Postoperatively, 2 patients developed prolonged ileus. One underwent exploratory laparotomy and washout for presumed leak, but none was identified. Three patients were readmitted within 30 days; 2 for intolerance to enteral intake with dehydration, and 1 for closed-loop obstruction requiring laparoscopic lysis of adhesions. The average and median length of follow-up were 27.7 and 26.0 months, respectively. Patients gained an average of 2.2 kg with an increase in body mass index of from 19.6 to 20.4 m/kg^2. Although 14 of 18 patients reported initial symptom improvement, at latest follow-up, only 6 patients reported symptomatic improvement or resolution. Three were diagnosed with global dysmotility, and 1 underwent intestinal transplant. Two were diagnosed with gastroparesis, and 1 underwent a laparoscopic gastric electric stimulator placement and pyloroplasty. There were no mortalities.
Conclusion Duodenojejunostomy is the most common surgical intervention in management of SMAS. Our intermediate follow-up reveals infrequent improvement and rare resolution of preoperative symptomatology. Patients had a modest average weight gain postoperatively. This may suggest that different preoperative workup and treatment is indicated.

Keywords Superior mesenteric artery syndrome · Duodenojejunostomy · Intestinal dysmotility · Gastroparesis · Chronic abdominal pain

Superior mesenteric artery syndrome (SMAS) is an uncommon cause of abdominal pain and failure to thrive [1–3]. SMAS refractory to medical therapy is referred for surgical management, most commonly laparoscopic duodenojejunostomy. Prior smaller case reports have described symptomatic relief in the immediate postoperative period [4]. Our group has previously reported early results with acceptable perioperative morbidity and modest symptom improvement [1]. Due to promising relief of symptomatology in the early postoperative period, we predicted similar results with intermediate follow-up. However, longer-term follow-up evaluating durability of these results

Presented at the SAGES 2016 Annual Meeting, March 16–19, 2016, Boston, Massachusetts.

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as well as safety of the procedure are lacking in the literature. We report the largest series of patients undergoing minimally invasive duodenojejunostomy for SMAS and review our experience at intermediate follow-up.

Methods

An institutional review board-approved retrospective chart review was performed on all patients with SMAS who underwent minimally invasive duodenojejunostomy from March 2005 to August 2015 in our healthcare system. All patients who received the diagnosis of SMAS and underwent a minimally invasive duodenojejunostomy including laparoscopic and robotic-assisted laparoscopic surgeries were included. Surgeries which required conversions to open procedures or those which were planned open procedures were not included in our review. In addition, only those with at least 6 months of follow-up were included in our cohort. Our patients underwent preoperative evaluation for their SMAS including imaging evaluation, most commonly upper gastrointestinal study (UGIS) with limited small bowel follow-through and/or CT imaging of the abdomen with angiogram to delineate the vascular anatomy and its relationship to the duodenum. Adjunctive evaluations were also considered including gastric emptying study, upper endoscopy, and CT scan of the abdomen and/or pelvis, as indicated by patient presentation. Data collected included patient demographics, clinical presentation, operative data, and outcomes such as perioperative and postoperative morbidity and mortality. Surgical technique of laparoscopic duodenojejunostomy was previously described by our group [1].

Table 1 Patient demographics

Mean age (years)	31.1
Male (%)	4 (22.2 %)
Female (%)	14 (77.8 %)
Comorbid conditions (%)	16 (88.9 %)
Psychiatric disease	11 (61.1 %)
Prior abdominal surgery	10 (55.5 %)
GERD	4 (22.2 %)
Chronic constipation	2 (11.1 %)
Chronic opiate therapy	2 (11.1 %)
Inflammatory bowel disease	2 (11.1 %)

GERD gastroesophageal reflux disease

Table 2 Patient presentation

Symptom duration (months)	
Mean	15.2
Median	8.0
Symptomatology (%)	
Abdominal pain	18 (100 %)
Nausea/vomiting	16 (88.9 %)
Weight loss	10 (55.5 %)
Bloating	4 (22.2 %)
Preoperative BMI (kg/m^2)	
Mean	19.9
Median	20.5
Mean preoperative weight loss (kg)	14.0
Abnormal imaging	
Upper GI	10 (55.5 %)
CTA	14 (77.8 %)

Results

Of 26 patients who underwent minimally invasive duodenojejunostomy for SMAS at our institution, 18 were followed for 6 months or more from the initial procedure. Preoperative patient demographics are featured in Table 1. The mean age at time of surgery was 31.1 years, and there were 4 men and 14 women. Sixteen of 18 patients (88.9 %) had other comorbid conditions. The most common was psychiatric disease, present in 11 of 18 patients (61.1 %). Ten of 18 patients had prior abdominal procedures. Other common comorbid conditions included: 4 patients had gastroesophageal reflux disease (GERD); 3 had chronic constipation; 2 carried a preoperative diagnosis of chronic abdominal pain; and 2 were on chronic opiate therapy.

Data regarding patient presentation is found in Table 2. The most common presenting symptom was postprandial abdominal pain which all 18 patients reported. The second most common symptom was nausea and vomiting which was present in 16 of 18 patients (88.9 %). Ten of 18 patients (50 %) reported preoperative weight loss secondary to their symptoms with a mean weight loss of 13.9 kg/m^2, representing 24.1 % total body weight loss. Mean preoperative body mass index (BMI) was 19.7 kg/m^2. Three of 18 patients were maintained on total parenteral nutrition (TPN) preoperatively for nutritional supplementation. Patients' diagnoses were made by clinical presentation, elimination of other diagnoses, and radiographic confirmation, either a CT of the abdomen with angiography demonstrating a narrowed aortomesenteric angle (8 of 18 patients) or an upper gastrointestinal series demonstrating extrinsic obstruction at the third portion of the duodenum with dilation of the first and second portions of the duodenum (4 of 18 patients) or both (6 of 18) (Fig. 1). The duration of symptoms prior to diagnosis ranged from 1 to 51 months (average 15.5 months).

Operative data is presented in Table 3. Sixteen patients underwent a laparoscopic duodenojejunostomy, while 3

Fig. 1 Upper gastrointestinal series demonstrating obstruction at the level of the third portion of the duodenum due to compression by the superior mesenteric artery (*red* arrow) (Color figure online)

underwent a robotic-assisted laparoscopic duodenojejunostomy. There were no intraoperative complications in our series. One patient had concomitant replacement of a prior venting gastrostomy tube; another had a feeding jejunostomy placed at time of her operation for nutritional supplementation; and 2 had concomitant extensive lysis of adhesions for significant intraabdominal adhesions. The average operative time was 143.9 min, and the average estimated blood loss was 21.4 mL. Postoperatively, 2 patients developed prolonged ileus, and 1 required postoperative TPN supplementation. One developed symptoms concerning for an anastomotic leak including tachycardia with abdominal pain and leukocytosis requiring an exploratory laparotomy and washout; however, no anastomotic perforation was identified during the exploration. Median length of stay was 5 days.

The postoperative outcomes are given in Table 4. Three patients were readmitted within 30 days. Two were admitted for dehydration and intolerance to oral ($n = 1$) or tube feedings ($n = 1$) and were successfully managed medically. One patient was readmitted with a closed-loop obstruction and underwent a laparoscopic lysis of adhesions, after which she was discharged in stable condition with resolution of her symptoms.

Eighteen patients were available for intermediate follow-up defined as at least 6-month follow-up with an average and median length of follow-up was 27.7 and 26.0 months, respectively. Patients gained an average of 2.2 kg with an increase in body mass index of from 19.6 to 20.4 m/kg^2. Fourteen of 18 patients reported initial improvement of symptoms such as postprandial epigastric pain, nausea and vomiting, frequency of emergency room visits and hospital admissions, and weight stability. Symptomatic improvement was determined in postoperative follow-up clinic

Table 3 Operative data

Patient	Operative time (min)	EBL (mL)	LOS (days)	Intraoperative details
1	91	10	25	
2	86	10	7	Replacement venting gastrostomy
3	163	25	5	
4	123	0	3	
5	242	50	4	Robotic
6	454	20	13	Robotic
7	123	20	3	Robotic
8	231	25	3	
9	75	0	6	
10	80	25	3	
11	64	0	4	
12	146	25	8	Concurrent feeding jejunostomy
13	77	25	3	
14	79	25	10	Extensive lysis of adhesions
15	85	25	8	
16	216	25	4	
17	129	25	7	
18	127	50	6	Extensive lysis of adhesions

EBL estimated blood loss, *LOS* length of hospital stay

Table 4 Postoperative outcomes

Patient	BMI at latest follow-up (kg/m^2)	Duration of follow-up (mo.)	Postoperative course
1	15.6	8	Persistent poor PO requiring TPN, eventually underwent intestinal transplantation after diagnosis of global dysmotility
2	17.9	10	Persistent nausea and abdominal pain requiring venting PEG and TPN supplementation. Smart pill study showed global dysmotility
3	23	7	Multiple ED admissions for persistent abdominal pain; CT imaging with patent anastomosis. Persistent abdominal pain, no improvement
4	22.1	12	*Symptoms resolved*
5	28.2	12	Chronic abdominal pain on gabapentin, no improvement
6	20.5	30	*Symptoms improved*
7	23.6	35	*Symptoms improved*
8	22.7	20	Chronic abdominal pain with hospitalizations and subsequent C difficile colitis, no improvement
9	17.2	27	Persistent nausea and abdominal pain requiring venting PEG and TPN supplementation. Diagnosed with global dysmotility, undergoing intestinal transplant evaluation
10	23.6	24	*Symptoms resolved*
11	19.5	22	Later diagnosis of gastroparesis s/p GES followed by pyloroplasty
12	14.7	28	Cyclic nausea/vomiting requiring PEJ and tube feed supplementation
13	19.1	29	*Symptoms improved*
14	17.3	33	Chronic abdominal pain on tricyclic antidepressants and gabapentin
15	20.5	43	Initially with weight gain but with recurrence of symptoms including weight loss, nausea, and abdominal pain at recent follow-up visits
16	21.6	59	Persistent abdominal pain, diagnosed with gastroparesis, managed medically
17	19.7	64	Chronic nausea and vomiting s/p laparoscopic jejunostomy and tube feed supplementation
18	19.5	11	*Symptoms resolved*

Italics were to make more visually obvious which patients had symptomatic resolution/improvement

BMI body mass index, *mo.* months, *PO* per oral nutrition, *TPN* total parenteral nutrition, *PEG* percutaneous endoscopic gastrostomy, *s/p* status post, *GES* gastric electric stimulator, *PEJ* percutaneous endoscopic jejunostomy

visits as well as via phone questionnaires. However, at latest follow-up, only 3 patients reported symptomatic improvement, while 3 patients reported symptomatic resolution at latest follow-up. Of the remaining 12 patients, 3 patients were subsequently diagnosed with global intestinal dysmotility and 1 patient has since undergone intestinal transplantation, while 1 patient remains on the intestinal transplant list. Two patients were diagnosed with gastroparesis postoperatively; 1 underwent a gastric electric stimulator placement followed by a pyloroplasty, while the other is being managed medically for his symptoms. The remaining 7 patients remain severely symptomatic at latest follow-up and comparable to their preoperative symptomatology. Five patients still require nutritional supplementation with TPN or tube feeds due to continued poor per oral (PO) intake secondary to persistent symptoms of nausea and abdominal pain. Symptomatic patients have undergone extensive workup including endoscopy and imaging studies which have revealed patent anastomosis as well as normal transit time through the duodenojejunal anastomosis. Three patients are being managed by chronic pain management for their abdominal pain and nausea with multimodal pain therapy including gabapentin and tricyclic antidepressants. None of the patients in our series have suffered from symptoms of blind loop syndrome like new-onset bloating, diarrhea, or greasy stools. There were no mortalities in our series.

Discussion

Superior mesenteric artery syndrome (SMAS) is defined as compression of the third portion of the duodenum between the superior mesenteric artery and the aorta and was first described by Carl Freiherr von Rokitansky in 1861 as an autopsy finding [1]. Not long afterward it was described by Willet in a patient in a full-body cast in 1878 [5], leading to the eponym "cast syndrome" and subsequently was clinically expanded on by Wilkie in a series of 64 patients in 1927 [6]. It is a rare cause of abdominal pain most commonly seen after a pathologic amount of weight loss and is usually a diagnosis of exclusion.

The normal angle between the aorta and the SMA in humans ranges from 25° to 60°, while the normal aortomesenteric distance is 10–28 mm [4]. Loss of the mesenteric fat pat with an angle of less than 20° between the SMA and the aorta due to severe weight loss, anatomic abnormalities secondary to congenital or acquired spinal deformities, or high insertion of the ligament of Treitz may result in compression of the third portion of the duodenum against the aorta and the spine at the level of L2 or L3 [1, 3, 4, 6]. The diagnosis of SMAS in our cohort was confirmed with either dynamic upper gastrointestinal studies which demonstrated obstruction at the second and third portion of the duodenum or CT angiography which was used to measure the aortomesenteric angle and confirm that the takeoff of the SMA was less than 20°.

Symptoms of SMAS include postprandial abdominal pain, nausea and vomiting, and weight loss [1, 4–6]. The most common presenting symptom in our patient population was postprandial abdominal pain followed by nausea and weight loss. The incidence of SMA syndrome is quoted to be 0.0024–0.34 % [3], but this incidence may be higher in chronically ill institutionalized patients as well as in children with delayed development [2, 4]. SMAS is more commonly described in women, perhaps due to the higher incidence of concomitant eating disorders that may contribute to the disease process [4], and in our series the overwhelming majority of patients were women (14:4).

In the largest series of medically managed SMA patients [2], chronic illnesses associated with weight loss such as tuberculosis as well as diseases of the nervous system such as cerebral palsy, muscular dystrophy, and Parkinson's disease were common comorbid conditions seen in patients with SMAS [2]. However, the most common comorbid condition seen in that cohort was psychiatric disease, similar to our series in which this was the most common comorbidity seen in 11 of 18 patients (61.11 %).

Nutritional optimization including hyperalimentation to increase mesenteric distance is the first step in management of SMAS in an effort to increase the aortomesenteric angle [4, 6]. Our institution provides nutritional supplementation in the form of oral supplementation, escalating to feeding tubes and lastly to total parenteral nutrition. Three of 18 patients were on TPN prior to their surgical procedures. Failure of medical management has traditionally been an indication for surgical intervention. The strong procedure maintains intestinal integrity and aims to relieve intestinal obstruction by dividing the ligament of Treitz, thus allowing the third portion of the duodenum to fall away from the SMA. However, tethering of the inferior pancreaticoduodenal artery may explain the up to 25 % symptomatic failure rate seen with the procedure compared to duodenojejunostomy [4, 6, 7]. Gastrojejunostomy has largely been abandoned as the proximal duodenum is not decompressed and may result in blind loop syndrome [6, 8]. The most common and successful surgical procedure for SMAS is minimally invasive duodenojejunostomy. In brief, this operation mobilizes a loop of jejunum approximately 20 cm distal to the ligament of Treitz is anastomosed to the dilated second portion of the duodenum to decompress and bypass the obstructed area. This operation has a reported 90 % success rate in the literature [6, 8]; however, case series in the literature report only short-term outcomes for small series.

We previously reported the largest single-institution series of patients to undergo minimally invasive duodenojejunostomy for SMA syndrome [1]. In this series, all patients had symptomatic improvement immediately following surgery. In this current series, we present the largest case series of patients who have undergone minimally invasive duodenojejunostomy for SMA syndrome with the longest follow-up of at least 6 months. We have found that with intermediate follow-up of at least 6 months and an average of 27.7 months, the vast majority of patients experienced immediate symptomatic improvement in the postoperative period. However, we found that only 33.3 % of patients (6 of 18) reported symptomatic improvement or resolution at intermediate follow-up. Of the patients with symptom recurrence, endoscopic and imaging workup revealed patent duodenojejunal anastomosis without obstruction, and it is unclear if symptoms may be functional. Chronic pain syndromes and gastrointestinal tract dysmotility syndromes have common symptom overlap with SMAS.

Of patients with symptomatic recurrence, two patients were diagnosed with gastroparesis following their surgery. Neither carried a preoperative diagnosis of gastroparesis and neither had a gastric emptying study prior to their procedure. Both patients had abnormal imaging studies that were diagnostic for SMAS (both underwent an UGIS and a CT); one patient went on to undergo a gastric electric stimulator followed by a pyloroplasty with some relief of symptoms, while the other is being managed medically with suboptimal relief of symptoms.

Three patients were diagnosed with global intestinal dysmotility following their surgeries. Their diagnoses were confirmed with gastrointestinal transit studies by SmartPill™, and none carried a preoperative diagnosis of dysmotility syndromes. Given that intestinal dysmotility is a functional disorder, whereas SMAS is classically described as a structural disease, after the diagnosed of SMAS is established, patients do not routinely undergo SmartPill™ evaluations or further workup to rule out intestinal dysmotility at our institution. However, given the overlapping vague symptomatology associated with the diseases including postprandial abdominal pain and weight loss, it may be a consideration in the future in the workup of the

SMAS, perhaps prior to surgical intervention. More importantly, the radiographic findings of SMAS should not necessarily exclude the concurrent diagnosis of intestinal dysmotility syndromes, especially given similar presenting symptoms.

Given the small number of patients with relief of symptoms after surgery in our series, it is difficult to conclude whether we can predict which patients may respond to minimally invasive duodenojejunostomy. In our series, there were no appreciable differences in preoperative comorbid conditions between those who experienced relief after DJ versus those patients with persistent symptoms. In addition, patient presentation in terms of symptomatology was similar between those who responded to surgery and those who did not. In our series, there are minimal perioperative and intermediate complications associated with the procedure. However, given the unknown long-term symptomatic relief following surgery, better prospective studies are needed to elucidate the best, most durable treatment option in SMA syndrome.

SMAS is a diagnosis of exclusion with a sometimes vague constellation of symptoms. Diagnosis is typically confirmed with radiographic findings, which were well documented in our cohort. However, we were disappointed to find that initial symptomatic relief did not appear durable at intermediate follow-up, leading us to speculate whether the finding of obstruction at the third portion of the duodenum or the narrowed aortomesenteric are secondary to other various pathologies rather than a single disease process in and of itself. Limitations in our study include that this is a retrospective analysis from a single institution, possibly introducing bias inherent in retrospective analyses and potentially missing patients that sought care or underwent interventions at other institutions.

Conclusion

Duodenojejunostomy is the most common surgical intervention in management of SMAS. In this largest series of minimally invasive duodenojejunostomy for SMA syndrome at intermediate follow-up, we find infrequent improvement and resolution of preoperative symptomatology. In contrast, symptomatic relief in the immediate postoperative period was much higher in this patient population. This may suggest that different preoperative workup and treatment is indicated to find a more accurate diagnosis prior to surgical intervention for SMA syndrome.

Compliance with ethical standards

Disclosures Julietta Chang, Mena Boules, John Rodriguez, Matthew Walsh, Raul Rosenthal, and Matthew Kroh have no conflicts of interest or financial ties to disclose.

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