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ORIGINAL ARTICLE

Superior Mesenteric Artery Syndrome: a Prospective Study in a Single Institution

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Abstract

Background Superior mesenteric artery syndrome (SMAS) is a rare cause of duodenal obstruction, resulting from the compression of the duodenum between superior mesenteric artery and aorta. This prospective registry aims to describe demographic, clinical, and outcome features of patients suffering from SMAS and to point out the indications for surgery.

Methods Between 2008 and 2016, patients with chronic gastrointestinal symptoms and diagnosis of SMAS were included. Demographics, clinical presentation, diagnosis, and surgical outcome were recorded. Symptoms were investigated with a standardized questionnaire. The diagnosis was achieved through barium swallow, CT/MR angiography (aortomesenteric angle ≤ 22°, distance ≤ 8 mm), endoscopy. All patients underwent duodenojejunostomy ± distal duodenum resection. At follow-up, symptom score and barium swallow were re-evaluated.

Results Thirty-nine patients (11 M/28 F, median age 38 years, median BMI 17.8 kg/m^2) were included. Barium swallow showed a gastroduodenal dilation in 57% of patients, and a delayed gastroduodenal emptying in 38%. Median aortomesenteric angle was 11° and distance was 5 mm. All patients underwent duodenojejunostomy, and in 32 patients, a distal duodenum resection was also performed. At a median follow-up of 47 months, the overall symptom score significantly dropped (10 vs. 32, $p < 0.0001$) and BMI increased (19.5 vs. 17.8, $p < 0.0001$). Barium swallow at 2 months postoperatively showed an improvement in terms of gastroduodenal dilation and emptying in 38% of patients with preoperative pathological findings.

Conclusions SMAS is a rare condition that should be suspected in cases of chronic, refractory upper digestive symptoms, particularly in females with low BMIs. Surgical treatment may improve symptoms and quality of life, although it is not curative in all cases.

ClinicalTrials.gov Identifier: NCT03416647

Keywords Superior mesenteric artery syndrome · Chronic duodenal obstruction · Duodenojejunostomy · Bile reflux

Introduction

Superior mesenteric artery syndrome (SMAS) is a rare cause of chronic duodenal obstruction, resulting from the compression of the third portion of the duodenum between superior mesenteric artery and the aorta. Over the years, it has been described by various names, i.e., Cast Syndrome, Wilkie's syndrome, arteriomesenteric duodenal compression, and duodenal vascular compression. These multiple terminologies have made it difficult to estimate the true frequency in the general population. The prevalence has been estimated at around 0.0024–0.3%, and the syndrome has been diagnosed more often in women in early adulthood.[1–3] Risk factors include significant weight loss (eating disorders, burn injury, bariatric surgery), abdominal or spinal surgery (proctocolectomy with ileoanal pouch for ulcerative colitis, spinal elongation for the treatment of scoliosis) and anatomic and congenital abnormalities (intestinal malrotation, low origin of the superior mesenteric artery, high insertion of the ligament of Treitz).[4–8] However, according to Akin et al., up to 40.4% of SMAS cases have no apparent cause.[9]

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Clinical presentation consists of chronic upper gastrointestinal symptoms, such as postprandial epigastric pain, nausea, vomiting, dyspepsia, abdominal bloating and weight loss secondary to decreased oral intake. A specific symptom score for patients with SMAS has not yet been developed. The differential diagnosis includes motility disorders, gastroesophageal reflux disease, pancreatitis, peptic ulcer disease, biliary colic, and mesenteric ischemia.[3,10] Given the poor specificity of the symptoms reported, the diagnosis remains challenging and is often rendered by excluding competing diagnoses.

Indeed, once the syndrome is suspected clinically, it must be confirmed radiologically by a barium swallow and a CT and/or MR angiography that represent the "gold standard". Esophagogastroduodenoscopy (EGD) can also provide additional information, such as the presence of esophagitis and/or gastritis caused by duodenogastric reflux. EGD may also help exclude differential diagnoses.

Therapeutic options for SMAS range from conservative management – including gastroduodenal decompression, correction of electrolyte imbalances and adequate nutritional support[11]—and surgical strategies to bypass or remove the obstruction. Surgery is recommended only when initial conservative treatment fails, or in severe cases.[12]

We prospectively investigated 39 patients with a history of chronic refractory upper gastrointestinal disorders, who underwent surgical correction for SMAS, in order to discuss clinical presentation, diagnostic workup and surgical outcome at a long-term follow-up. The following knowledge gaps were investigated and addressed: (a) a standardized diagnostic workup for superior mesenteric artery syndrome; (b) a specific symptom score, in order to standardize the evaluation of pre/postoperative symptoms; (c) the duration of conservative treatment before considering surgical correction of the syndrome; (d) thorough indications for surgery; (e) a theoretical comparison between different surgical techniques; (f) a precise knowledge of long-term outcomes after surgical correction of SMAS.

Patients and Methods

Between October 2008 and March 2016, we prospectively screened 254 consecutive patients with aspecific upper digestive symptoms, lasting from at least 5 months, who were referred to the Department of Surgery, Oncology and Gastroenterology at the University-Hospital of Padua (Italy), a referral center for complex upper gastrointestinal surgery. We excluded 215 (85%) patients based on the following criteria: inability to provide the informed consent (2.8%); malignancies (7.4%); bowel motility disorders (57.2%); severe psychiatric illness (9.3%); pregnancy (2.3%); impossibility to perform the required diagnostic workup (20.9%).

Thirty-nine enrolled patients underwent duodenal surgery due to clinical and radiological criteria. They presented with one or more of the following clinical features: (a) severe and frequent upper digestive symptoms (occurring at least once a week), associated with poor quality of life and refractory response to medical treatment; (b) a condition of underweight (BMI < 18.5 kg/m^2) associated with difficulty eating; (c) severe complications of SMAS (e.g., gastric perforation, acute pancreatitis, aspiration pneumonia). In all patients, the following features were recognized at imaging studies: (a) suggestive findings of SMAS at barium swallow; (b) diagnostic aortomesenteric angle and distance at CT/MR angiography.

This study was performed according to the Declaration of Helsinki Principles. It was reviewed and approved by the Internal Review Board in Padua, Italy. Written consent was obtained from all patients prior to study enrolment.

Preoperative Assessment

The following demographic and clinical data were prospectively collected: age, weight, height, body mass index (BMI), symptom duration, past and current medical therapy, and previous surgical and endoscopic procedures.

A symptom score was collected before and after surgery. Symptoms were assessed at baseline and during follow-up by using a detailed Lickert-scale based questionnaire for epigastric pain, nausea, vomiting, abdominal bloating and regurgitation. For each symptom, a score was calculated by combining severity with frequency, as shown in Table 1. The total symptom score was obtained from the sum of the scores of all the symptoms, a maximum value of 55, and a minimum value of 25 was judged necessary to suspect a diagnosis of SMAS.

The clinical diagnosis required confirmation by radiologic investigations, such as barium swallow, CT and/or MR angiography with multiplanar 3-dimensional reconstructions, and EGD.[13] Typical findings at barium swallow are gastrectasy, proximal duodenal dilation, an abrupt vertical or oblique compression of the third portion of the duodenum, duodenogastric reflux, and a delayed transit (Fig. 1). Radiological criteria at CT/MR angiography are a reduction of the aortomesenteric angle to 22° or less (normal 25°–60°) and a decrease of the aortomesenteric distance to 8 mm or less (normal 10–28 mm).[14] The aortomesenteric distance was defined as the maximum distance at the level where the duodenum passes between the vessels (Fig. 2a); the aortomesenteric angle was measured at the same level on sagittal images (Fig. 2b). Both values were blindly reviewed and calculated by two expert radiologists in order to standardize the method. EGD cannot rule out the diagnosis, but it can show the presence of bile reflux, suggestive for duodenal obstruction, and exclude malignant diseases.

Table 1 Symptom score. The following questionnaire was administered before and after surgery

1. Report the **FREQUENCY** of symptoms you are experiencing using the numbering system below:
0 = Never, 1 = Occasionally, 2 = Once a month, 3 = Every week, 4 = Twice a week, 5 = Daily

SYMPTOMS	0	1	2	3	4	5
Epigastric pain						
Nausea						
Abdominal bloating						
Vomiting						
Regurgitation						

2. Report the **SEVERITY** of your symptoms using the ratings list below:
0 = None, 2 = Mild, 4 = Moderate, 6 = Severe

SYMPTOMS	0	2	4	6
Epigastric pain				
Nausea				
Abdominal bloating				
Vomiting				
Regurgitation				

Surgical Intervention and Postoperative Assessment

Duodenojejunostomy via a short upper midline incision was the treatment of choice, with or without a distal resection of the duodenum. The surgical technique of duodenojejunal bypass consisted in dissection of the visceral peritoneum covering the duodenum at the base of the mesocolon, and fashioning of a side-to-side transmesocolic loop-duodenojejunostomy. In patients who underwent a distal resection of the duodenum, a division of the ligament of Treitz, duodenojejunal resection and an anterior transposition of the jejunum through a rent in the transverse mesocolon were performed.

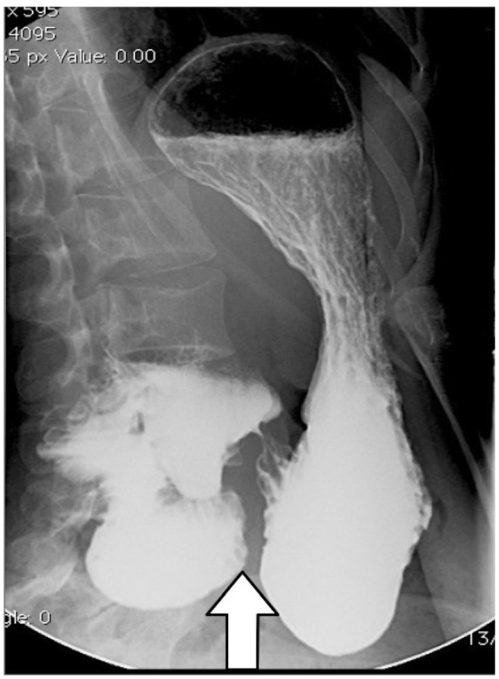

Fig. 1 Typical findings at barium swallow are gastrectasy, proximal duodenal dilation, and a remarkable duodenal narrowing at the site of passage between the superior mesenteric artery and the abdominal aorta

All patients underwent water-soluble contrast swallows (with Gastrografin®, Bracco, Milan, Italy) on postoperative day 6 to exclude anastomotic leakage. On the same day, they progressed to a liquid diet, then to soft foods on postoperative day 7.

Short- and Long-term Follow-up

At follow-up, a clinical and radiological evaluation was performed. This included a barium swallow at 2 months post procedure, while clinical outcome was assessed by means of the same standardized questionnaires used at baseline and at ≥ 12 months after postoperatively. We considered a positive response to surgery to be a reduction in symptom score of greater than one third of the initial value. Moreover, BMI and weight were also evaluated and considered in determining a positive/negative outcome, a positive outcome being weight gain. Postoperative quality of life was evaluated through a short questionnaire in order to stratify patient satisfaction regarding their surgery in the following categories: satisfied/neutral/unsatisfied.

Statistical Analysis

Continuous data were expressed as medians and interquartile range (IQR). Categorical data were compared between the preoperative and postoperative period using

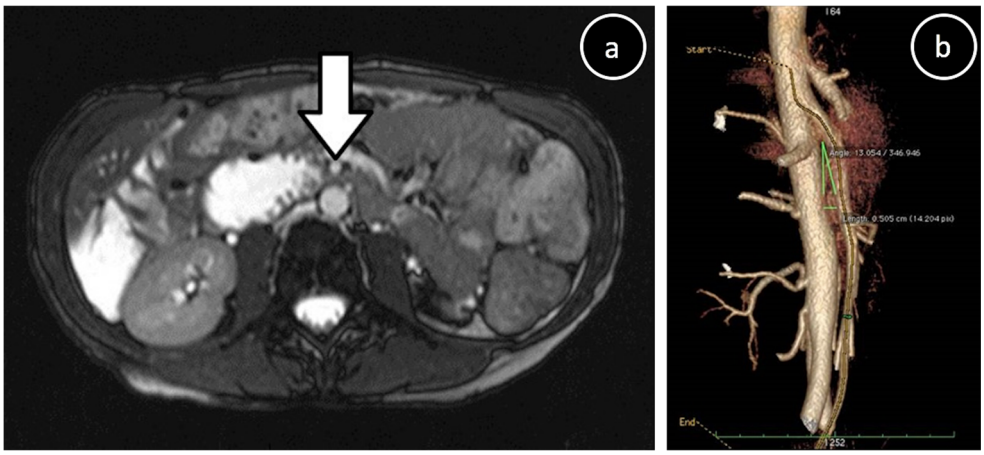

Fig. 2 Aortomesenteric distance and angle: **a** MR angiography showing dilation of the second duodenal portion, caused by a decrease in the aortomesenteric distance to 4 mm. **b** CT angiography with vascular reconstructions, showing duodenal compression by the superior mesenteric artery and narrowing of the aortomesenteric angle to 13°

Fisher's test, and continuous data using the Mann-Whitney non-parametric test. Pre versus postoperative variations in continuous data were assessed within each group using Wilcoxon's non-parametric test for paired data. A p value of less than 0.05 was considered significant. Statistical analyses were performed using SAS 9.1 software.

Table 2 Demographic and clinical data, expressed as median and interquartile range

Patients, *n*	39
Male, *n* (%)	11 (28%)
Female, *n* (%)	28 (72%)
Age, years	38 (26–46)
≤ 19, *n* (%)	4 (10%)
20–29, *n* (%)	10 (26%)
30–39, *n* (%)	6 (15%)
40–49, *n* (%)	12 (31%)
50, *n* (%)	7 (18%)
Weight loss, kg	10 (4–15)
BMI, kg/m^2	17.8 (15.6–19.8)
Duration of symptoms, months	66 (12–120)
Symptom score, *n*	32 (26–39)
Weight loss, *n* (%)	34 (87%)
Nausea, *n* (%)	31 (80%)
Abdominal pain, *n* (%)	29 (74%)
Bloating, *n* (%)	27 (69%)
Vomiting, *n* (%)	24 (62%)
Regurgitation, *n* (%)	22 (56%)
Complications	
Poor nutrition, BMI scored as "underweight," *n* (%)	22 (56%)
Dehydration, electrolyte imbalance requiring parenteral hydration, *n* (%)	6 (15%)
Acute pancreatitis, *n* (%)	8 (21%)

Results

Demographic and Clinical Data

Demographic and clinical data are shown in Table 2.

Thirty-four (87%) of our SMAS cases were idiopathic, while 5 female patients (13%) reported a possible risk factor in their clinical history. Two of the 5 patients had a history of an eating disorder (anorexia and bulimia, respectively), 2 patients suffered from ligamentous laxity (Marfan and Ehlers-Danlos syndrome), while one patient had a prior proctocolectomy with ileoanal J-pouch performed at another center. A single case of familial correlation (mother and son) was also recorded.

Thirty-four patients (87%) reported mild to severe weight loss, leading to a condition of underweight in 22 of them. Eight patients (21%) had suffered an episode of acute pancreatitis, requiring surgical or endoscopic treatment (cholecystectomy, endoscopic sphincterotomy) in 2 of these. Previous surgical history is recorded in Table 3, while Table 4 shows duration and effectiveness of the conservative initial treatment.

Radiological and Endoscopic Data

Barium swallow showed a gastroduodenal dilation in 22 cases (57%), and a delayed gastroduodenal emptying in 15 (38%). At CT/MR angiography, median aortomesenteric angle was 11° (8–15), and aortomesenteric distance was 5 mm (4–7). Moreover, EGD was further performed to detect the presence of gastritis (59%, with Helicobacter Pylori infection present in 11% of these), bile reflux (38%), esophagitis (13%), or duodenitis (6%). In 16% of cases, no pathological findings were discovered.

Surgical Intervention and Outcome Data

Due to patients' refractory response to medical therapy and poor quality of life, all patients underwent open

Table 3 Previous surgical history

Procedure or surgical intervention	Indication	No. patients	Total no. of procedures
Fundoplication	Gastroesophageal reflux disease	8	13 (*)
Cholecystectomy	Dyspepsia and gallstones	3	3
	Acute pancreatitis	2	2
Endoscopic papillotomy of the ampulla of Vater	Acute pancreatitis	2	8 (*)
Adhesiolysis	Chronic small bowel obstruction	6	7 (*)
Pyloroplasty	Severe gastric dilation after fundoplication	1	1
Proctocolectomy with ileoanal J-pouch	IBD (**)	1	1
Colectomy with definitive ileostomy	IBD	1	1
Gastrojejunostomy	SMAS	2 (***)	2
Strong's procedure	SMAS	1 (***)	2 (*)
Gastrostomy + jejunostomy	SMAS	1 (***)	1
Nasojejunal tube	SMAS	1 (***)	1

(*) In some patients, the same procedure/intervention was performed more than once

(**) IBD: Inflammatory bowel disease

(***) In one patient, after the failure of a twice-attempted Strong's procedure (first laparoscopic, then open) and of a Roux-en-Y gastrojejunostomy, the gastrojejunal anastomosis was disassembled because of a bleeding anastomotic ulcer. First, a jejunostomy for enteral feeding was performed, then an intravenous catheter for parenteral nutrition was obtained, and finally, a gastrostomy was performed to prevent vomiting

duodenojejunostomy. In 32 of these (82%), a distal duodenum resection was also performed and all specimens were sent for histopathologic examination, which was later reported as showing normal ganglion cells in both the submucosal and myenteric plexus. Ten patients (26%) also underwent a fundoplication for GERD. Gastrojejunal bypass was disassembled in one patient and gastrostomy was removed in another one so as to restore physiologic digestive continuity. Median operating time was 120 min (90–175), without a significant difference between patients who had previously undergone surgery ($n = 22$; median time 120 min, IQR 94–176) and those who had not ($n = 17$; median time 120 min, IQR 90–130). The need for fundoplication in 10 patients with proven reflux disease slightly prolonged surgical time (median time 133 min, IQR 126–180).

Postoperative mortality was nil. Complications occurred in 6 patients (15%). These included melena (treated conservatively with blood transfusions ($n = 2$)), acute pancreatitis (medically treated ($n = 2$)), intestinal obstruction due to adhesions (required surgical exploration for adhesiolysis ($n = 1$)), hemoperitoneum (surgical revision for hemostasis ($n = 1$)).

In all patients, a water-soluble contrast swallow was performed on postoperative day 6, and since no anastomotic leak was observed, realimentation proceeded on day 7; patients were discharged on postoperative day 9 (8–11). Postoperative pancreatitis and reoperations prolonged the hospital stay (median 24 days; IQR 20–27). Barium swallow at 2 months postoperatively demonstrated an improvement in terms of gastroduodenal dilation and emptying in 12 patients (38% of the patients with preoperative pathological findings). A wide and patent duodenojejunal anastomosis was demonstrated in all series.

At a median follow-up of 47 months (34–72), a significant increase in BMI was recorded and in 28 cases (72%) symptom score decreased markedly. Also, the need for antireflux and prokinetic therapy significantly reduced after surgery. Patient-

Table 4 Medical treatment

Medication	Patients, *n*	Patients reporting partial symptom relief, *n (%)*	Median duration of treatment (IQR), months
Antacids/PPIs (*)	39	7/39 (18%)	78 (17–123)
Prokinetic therapy	30	6/30 (20%)	60 (12–108)
Total parenteral nutrition	4	0 (0%)	3 (2–12)
Enteral feeding (**)	1	0 (0%)	38
Neuroleptics (***)	15	Not evaluated	102 (60–150)

(*) PPIs: Proton pump inhibitors

(**) via nasojejunal tube

(***) Prescribed elsewhere, for presumed depressive or anxiety disorders

Table 5 Comparison between preoperative and postoperative clinical data and medical therapies. Data are expressed as medians and interquartile range

	Before surgery	After surgery	*P* value
Symptom score	32 (26–39)	10 (4–21)	< 0.0001
BMI, kg/m^2	17.8 (15.6–19.8)	19.5 (17.6–21.8)	< 0.0001
Weight variation, kg	− 10 (4–15)	+ 5 (0–9)	< 0.0001
PPI therapy, *n* (%)	39 (100%)	7 (18%)	< 0.0001
Prokinetic therapy, *n* (%)	30 (77%)	13 (33%)	0.02

reported satisfaction with surgery was the following: 69% satisfied ($n = 27$), 13% neutral ($n = 5$), 18% unsatisfied ($n = 7$). Among the 7 unsatisfied patients (postoperative vs. preoperative symptom score: 27 vs. 29), further postoperative diagnosis was made: associated severe gastrointestinal dysmotility ($n = 5$), unspecified psychiatric disease ($n = 2$). Four patients underwent further surgical interventions: gastric resection with Roux-en-Y anastomosis ($n = 2$), Mikulicz pyloroplasty ($n = 1$), mesenteric artery stenting for Dunbar syndrome ($n = 1$).

A comparison between preoperative and postoperative clinical data and need for medical therapy is shown in Table 5.

Discussion

Superior mesenteric artery syndrome is a rare but disabling condition caused by a mechanical obstruction of the duodenum leading to gradual gastroduodenal dilation and impaired motility of the upper digestive tract. SMAS often results in severe weight loss and can be potentially life-threatening due to malnutrition, dehydration, electrolyte imbalances or other severe complications, such as gastric perforation, acute pancreatitis[15] or aspiration pneumonia.[16,17] Unfortunately, most clinicians are not aware of this disease, and as a result, patients are often misdiagnosed and the syndrome is often recognized after a lengthy period of ongoing abdominal complaints and a myriad of treatment failures.

SMAS has been reported more commonly among females, children, and adolescents.[18,19] Similarly, a higher prevalence in women and young adults has been confirmed in our patient population (Table 2). Low BMI and weight loss are clinical features of SMAS,[4–7] as well as consequences of reduced oral intake due to SMAS symptoms. This is evident in our cohort in which 87% of patients reported a rapid weight loss secondary to their symptoms, resulting in being underweight in 59% of them.

The diagnosis of SMAS is made using a combination of clinical and radiological features and requires a high index of suspicion. As underlined by Goin and Wilk in 1956, the syndrome is "endemic" in areas where radiologists are more familiar with the diagnosis.[20] The very existence of SMAS as a disease entity has been controversial because of confusion surrounding other causes of megaduodenum or duodenal ileus, including collagen vascular disorders and chronic idiopathic intestinal pseudoobstruction.[21]

All our patients presented with protracted symptomatology (≥ 5 months) with the median time interval between symptom onset and diagnosis being 66 months (IQR 12–120). During this period of time, they were misdiagnosed, treated medically and experienced either partial relief, persistence, or even worsening of symptoms. Furthermore, most of our patients underwent several endoscopies, but when no anatomical explanation for their symptoms was detected, many of them were referred to psychiatric treatment (see "Neuroleptics", Table 4).

In order to evaluate and standardize the clinical presentation of SMAS, we opted to use a symptom score that has proved to be very useful for many diseases, such as GERD, gastroparesis, achalasia, and irritable bowel syndrome.[22–25] To date, no standardized symptom score for SMAS and no universal "gold standard" for evaluation of outcome after SMAS surgery has been validated. This symptom score overlaps with the score proposed by Zaninotto et al. for GERD,[26] with only minor changes. Due to the rarity and uniqueness of this disease, proper validation had not been performed.

As to the instrumental diagnostic workup, an algorithm of preoperative evaluation has been proposed by Sun et al..[27] Based on the clinical picture and available literature, we suggest our diagnostic flow-chart (Fig. 3): (a) EGD, to exclude other organic causes of gastroduodenal obstruction; (b) barium swallow, to provide both anatomic and functional information on gastroduodenal area; (c) CT or MR angiography, which provide equivalent information on the aortomesenteric distance and angle,[27] although MR should be preferred in young female patients in order to avoid radiation.

The best treatment for SMAS is unclear. In our case series, most patients were misdiagnosed and all of them received empirical treatment, either medication, such as PPIs, antacids, H2-receptor blockers, prokinetics, neuroleptics, or with enteral or parenteral hyperalimentation. Despite protracted symptomatology, only 2 patients had been correctly diagnosed with SMAS in other centers and unsuccessfully treated. Some reports support conservative management as first-line treatment;[11,13] however, thus far, there has been no data on the appropriate duration of medical therapy before consideration of surgical intervention.[2,13,28] Earlier studies on SMAS in children reported that patients should have a trial of at least 4 to 6 weeks of conservative treatment with optimal nutritional support before surgery[29] and the outcomes seemed to be worse ($p = 0.018$) when the time limit for the duration of medical treatment exceeded 6 weeks.[30] Of note, Merrett et al.[2] surmised that medical treatment in adult patients with chronic symptoms is often a prolonged hospital admission with poor success rates. In our opinion, a medical trial of no more than

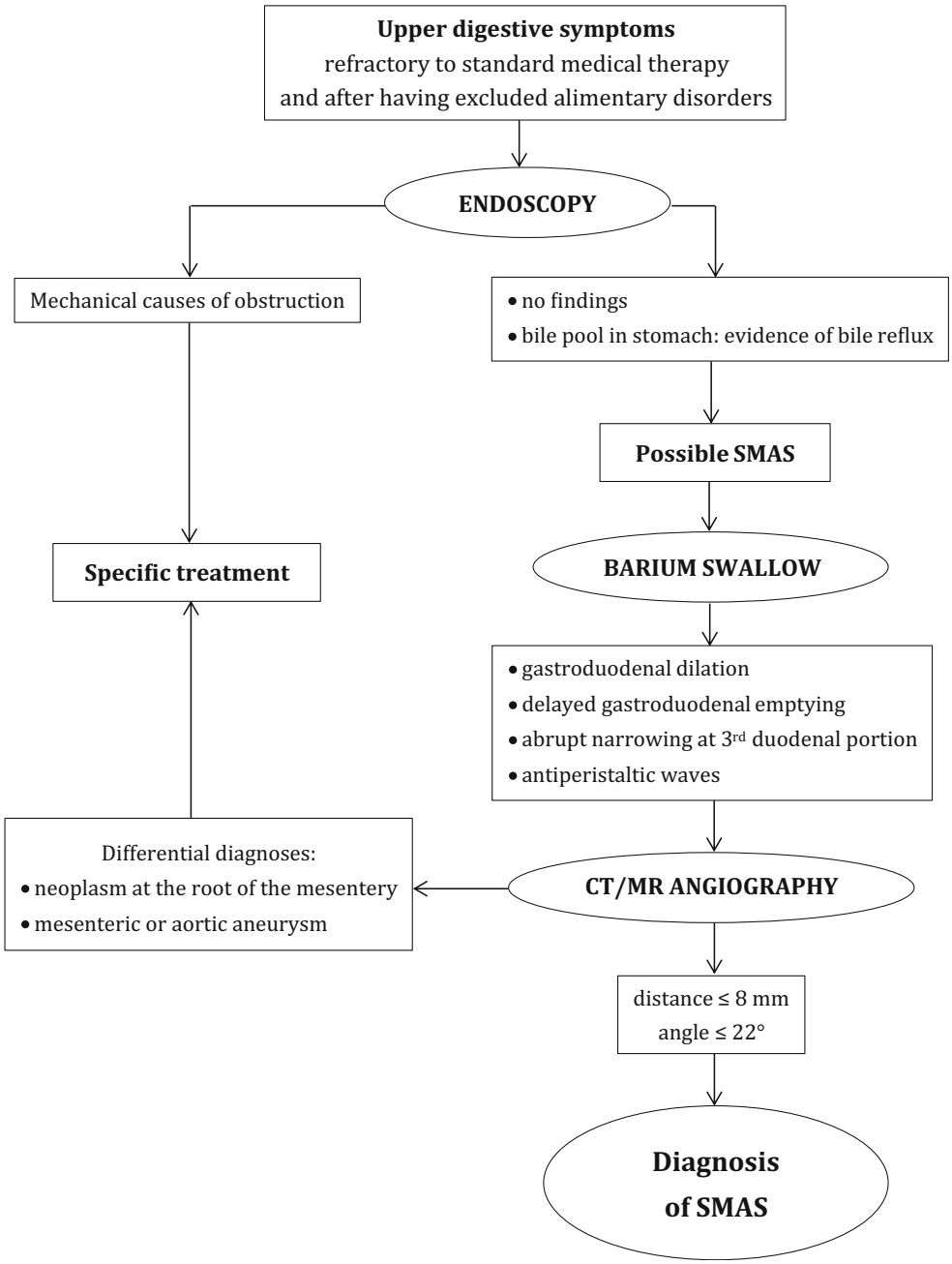

Fig. 3 Diagnostic flow-chart

3 months should be considered before surgery, in order to avoid chronicity of the disease.

Welsch et al.[13] recommend surgery in symptomatic patients when medical approaches fail. In a recent retrospective study of 80 patients with SMAS,[28] 57 were managed medically, showing an overall success rate of 71.3% and a recurrence rate of 15.8% at a median follow-up of 5 months (IQR 1–84). The overall success rate of surgical management in 14 patients was higher (92.9%) and the recurrence rate during the median follow-up period of 12 months was nil (0%).

Other indications for surgery are as follows: (a) severe presentation (i.e., aspiration pneumonia), (b) complications of TPN or EF (hepatic steatosis, cholestasis, catheter-related bloodstream infection or sepsis, tube displacement, patient's refusal), (c) complications of SMAS that require surgical exploration themselves (i.e., gastric perforation). As stated by Sun et al.,[27] when the diagnosis of SMAS is clinically and radiologically clear, a more aggressive approach in terms of surgical treatment should be adopted. In particular, the placement of a nasojejunal tube or jejunostomy for EF in young patients should be avoided since vomiting, nausea and epigastric pain persist despite tube placement, resulting in mediocre quality of life. In contrast, surgery can lead to complete symptom relief and restore oral feeding, although it is not curative in all cases.

Many surgical strategies have been proposed to bypass or remove the obstruction. These include (1) gastrojejunostomy; (2) Strong's procedure, consisting in the division of the ligament of Treitz and caudal mobilization of the ascending duodenum[31]; (3) duodenojejunostomy, with or without resection of the third duodenal portion. Conventional open surgical techniques or laparoscopic techniques can be used.[32] The data available in the literature does not provide sufficient evidence to identify superiority of one surgical procedure, and randomized controlled trials are not available.[13] Gastrojejunostomy has, nonetheless, been associated with insufficient release of the duodenal obstruction, blind loop syndrome or severe complications such as peptic ulceration due to massive duodenogastric reflux, with a higher risk of anastomotic bleeding or gastric cancer.[13,33,34] One of our patients had previously undergone a Roux-en-Y gastrojejunostomy, but the gastrojejunal anastomosis had to be disassembled because of a bleeding anastomotic ulcer. Strong's procedure is less invasive, quicker and safer, since it maintains bowel integrity,[33] but its unacceptably high failure rate is well known.[35] As further proof of its ineffectiveness, one of our patients underwent Strong's procedure twice, without any benefit. Most surgeons consider duodenojejunostomy superior to other techniques, since it has yielded the best results in severe cases and was significantly better when compared to gastrojejunostomy and Strong's procedure.[1,33] It also has a low risk of postoperative adhesions. Very good results after duodenojejunostomy have been reported in the literature: 79% (161 patients) in Barner's series,[29] 90% in 50 cases reviewed by Lee and Mangla[33] and 100% in Lee's series of 7 patients.[33] Most of the data regarding large studies of SMAS was published over 30 years ago[28] and descriptions of long-term outcomes are lacking in the literature. The longest follow-up, reported by Chang et al., on a recent retrospective case series of a small cohort of 18 patients had a mean length of 27.7 months.[36] Cases of blind loop syndrome after duodenojejunal bypass have been reported.[1] Since the first 7 patients of our series experienced blind loop syndrome after duodenojejunostomy without resection, we adopted the technique of duodenojeunal resection followed by antevascular side-to-side duodenojejunal anastomosis, in order to definitively remove the stenosis and prevent reversal peristalsis. Laparoscopic duodenojejunostomy is also a well-established approach with known advantages[28,36] but we prefer an open approach, as it allows a careful dissection of the connective tissue between the duodenum and pancreas and a wide anastomosis on the inferior surface of the duodenum.

As for the surgical outcomes, Chang et al. reported an immediate postoperative symptomatic improvement in all 18 patients; only one third of patients ($n = 6$) experienced ongoing relief at the latest follow-up (≥ 6 months),[36] while our patients experienced an incomplete regression of digestive symptoms at short-term follow-up and a progressive improvement in the long term. This was more obvious in patients with preoperative gastrectasy and delayed gastric emptying as evidenced by barium swallow several months post-surgery, thus requiring a prolonged course of prokinetic therapy. As stated by Welsch et al., duodenal atony after massive dilatation may persist even after duodenal decompression,[13] therefore lengthy symptom duration may be responsible for impaired gastric and duodenal peristalsis, precipitating a secondary motility disorder. Care must be taken to properly inform patients about the possibility of a delayed definitive improvement of symptoms.

In summary, we suggest the following indications for surgery: (a) a lengthy period of severe and frequent upper digestive symptoms (occurring at least once a week for > 6 months) with diagnostic characteristics at imaging studies; (b) refractory response to medical treatment, attempted for at least 3 months; (c) severe complications of SMAS.

Conclusions

To the best of our knowledge, this is the only prospective single-institution study on SMAS available in the medical literature, with a long-term follow-up. The following conclusions can be drawn from our experience:

1. SMAS should be considered as a differential diagnosis in patients with nonspecific chronic digestive symptoms of unknown origin.
2. A standardized diagnostic workup for SMAS must include EGD, barium swallow, and CT/MR angiography.
3. Symptom score is a useful tool for the evaluation of subjective patients' response to surgery.
4. Early diagnosis and surgical treatment may prevent gastroduodenal dilatation and the onset of a secondary motility disorder, and may improve symptoms and quality of life.
5. Duodenojejunal resection should be considered as a better surgical option compared to bypass, since it prevents blind loop syndrome and avoids reversal peristalsis.

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Authors' Contributions Angelica Ganss: study conception and design, acquisition of data, analysis and interpretation of data, drafting of the manuscript, critical revision of the manuscript

Sabrina Rampado: drafting of the manuscript, critical revision of the manuscript

Edoardo Savarino: critical revision of the manuscript

Romeo Bardini: study conception and design, critical revision of the manuscript

Compliance with Ethical Standards

This study was performed according to the Declaration of Helsinki Principles. It was reviewed and approved by the Internal Review Board in Padua, Italy. Written consent was obtained from all patients prior to study enrolment.

Conflict of Interest The authors declare that they have no conflict of interest.

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