Chapter 23 In Patients with Superior Mesenteric Artery Syndrome, Is Enteric Bypass Superior to Duodenal Mobilization?

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Abstract Superior mesenteric artery syndrome is an infrequent cause of duodenal obstruction within the narrowed aortomesenteric angle. The condition is characterized by vague and elusive symptomatology thus often making it difficult to diagnose. In addition, the etiology remains poorly defined and standard diagnostic criteria are lacking. Following a trial of supportive medical management, the surgical treatment options include traditional open or minimally invasive duodenojejunostomy, division of the ligament of Treitz (Strong's procedure), or gastrojejunostomy. Duodenojejunostomy has been the favored surgical technique historically and most described in the literature. Due to the rarity of the syndrome and overall inconsistencies in diagnosis and treatment, there is a paucity of evidence in the literature to strongly recommend one technique over the other. Available case series and case reports lack appropriate follow-up. Based on the existing data and our personal experience, our preference is to perform a laparoscopic duodenojejunostomy for the treatment of medically refractory SMA syndrome. However, larger and more rigorous studies will be needed to make more evidence-based recommendations.

Keywords Superior Mesenteric Artery Syndrome • Wilkie's Syndrome • Cast Syndrome • Aortomesenteric Obstruction • Chronic Duodenal Obstruction • Duodenojejunostomy • Strong's Procedure

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Introduction

Superior mesenteric artery (SMA) syndrome is an uncommon cause of duodenal outlet obstruction and is often difficult to diagnose due to its vague and elusive symptomatology. Clinically it is characterized by postprandial epigastric abdominal pain, nausea, bilious vomiting and weight loss. Pain is classically relieved by assuming a prone, knee-to-chest or lateral decubitus position [1, 2]. SMA syndrome affects predominantly young women between 10 and 39 years of age and thin, asthenic build [1, 3, 4]. While it is a very rare disease with an exact prevalence that is unknown, it is estimated to have an incidence of 0.013-0.3% based on upper gastrointestinal barium studies [4, 5]. Yet because there is no gold standard imaging test to confirm the diagnosis, its incidence is likely overrepresented [2, 6]. SMA syndrome was originally described in 1861 by Carl von Rokitansky based on his post-mortem observations of young asthenic females. He described acute gastric and duodenal dilation as a result of compression of the duodenum by the root of mesentery [7, 8]. The first large series consisting of 75 patients was published in 1921 by D. P. D. Wilkie, in which he described treatment options that are used today and still remain effective [1]. Along this historical context, the entity is still often referred to as Wilkie's Syndrome. The condition has been given many names over the years; chronic duodenal ileus, Cast syndrome (pernicious vomiting that resulted from the application of body cast [9, 10]), arteriomesenteric duodenal compression [11], aorto-mesenteric artery compression syndrome [12]. Wilkie proposed that congenital alteration in the relationship of the vessels to the duodenum, aggravated by an acute insult, leads to symptom onset and worsening [1]. Guthrie proposed that the disease is the result of man's upright posture acquired late in evolution [8].

The superior mesenteric artery originates at an acute angle off the aorta behind the neck of the pancreas at the level of first lumbar vertebrae. The aortomesenteric angle contains retroperitoneal fat, lymphatics, the uncinate process of the pancreas, and the left renal vein as it crosses over the aorta [13] (Fig. 23.1). The interposed adipose tissue within the aortomesenteric window is thought to displace the SMA anteriorly to a degree sufficient to allow for the duodenum to cross through the window without extrinsic compression. When this is no longer the situation, the etiology of SMA syndrome is believed to be due to vascular compression of the third portion of the duodenum as a result of a narrowed aortomesenteric angle. Classically explained, significant weight loss leading to critical loss of the fat pad within this angle is the proposed etiopathogenesis of SMA syndrome [6, 14, 15]. Congenital or acquired anatomic variations, such as short and high insertion of the ligament of Treitz, low origin of superior mesenteric artery, lumbar lordosis, or malrotation may also predispose to the syndrome [4, 6, 14, 16]. Symptom onset is reported to be precipitated by acute insults that lead to rapid weight loss and depletion of the abdominal adipose tissue (malabsorption, cancer, trauma, burns, neurological disorders, eating disorders, bariatric surgery), external compression (cast), intra-abdominal compression (dissecting aortic aneurysm), or mesenteric tension due to surgical alterations (proctocolectomy with ileoanal pouch anastomosis,

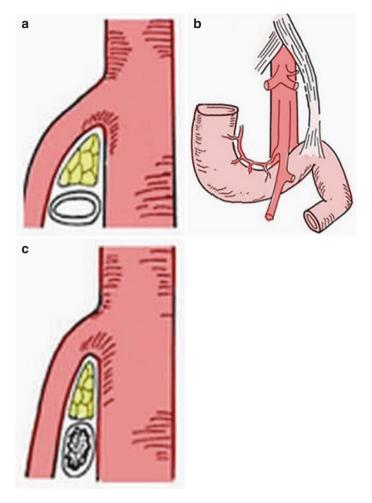


Fig. 23.1 Superior mesenteric artery and the aorta form an acute aorto-mesenteric angle (a). In superior mesenteric artery syndrome, the angle is markedly narrowed resulting in compression of the third portion of the duodenum (b, c)

corrective spinal surgery) [6, 8, 17–21]. The extent to which these disorders are also associated with an acquired gastric and duodenal motility disorder contemporaneous with a diagnosis of SMA, and the extent to which each contributes to the symptoms, is unstudied and therefore unknown.

Although patient demographics and presenting symptoms are similar between superior mesenteric artery syndrome and megaduodenum, the former is postulated to be a mechanical obstruction without underlying myopathy ruled out by duodenal biopsies whereas the latter is a hereditary motility disorder [4, 6, 14]. SMA syndrome often remains an ambiguous diagnosis as it can be overshadowed by

co-exisitingmedical conditions involving severe malnutrition, psychosocial eating-related disorders and substance abuse [14, 22]. Surgeons tend to be consulted for SMA syndrome when medical therapy and conservative management fail and generally focus on the mechanical plausibility of the diagnosis based on imaging. The indications for surgery remain a challenge since the diagnosis of SMA syndrome is typically made clinically since there is much variation in the interpretation and significance of imaging studies. It should be noted that imaging studies do not rule in the diagnosis of SMA syndrome, most often they rule it out. For these reasons, the diagnosis of SMA syndrome often remains ambiguous at best with the diagnosis confirmed when patients symptoms improve following surgery. Results of surgical outcomes are by and large incomplete and thus should be viewed with caution.

In general the diagnosis of SMA syndrome is suspected when patients can no longer maintain their weight without exogenous nutritional support, display symptoms suggestive of duodenal obstruction, and have had all other potential causes ruled out. It is good practice to first have patients screened in an eating disorders clinic by a specialist including a dietician. Once the possibility of an eating disorder is ruled out, the diagnosis is considered when an upper gastrointestinal contrast study and CT angiogram are together suggestive of SMA syndrome. Median arcuate ligament syndrome, which can cause symptoms similar to SMA syndrome, should also be ruled out, as well as any endoluminal or extrinsic obstructive cause of duodenal obstruction.

The radiologic criteria for the diagnosis of SMA syndrome can be highly subjective and a comprehensive review of the literature is beyond the scope of this review. In general, upper barium study should be performed by an experienced radiologist who is familiar with the diagnosis. Additionally a CT angiogram should confirm that there is narrowing of the aortomesenteric window. Surgeons considering intervention should realize that there is much variability in the measurement of the aortomesenteric window from one radiologist to another and much subjectivity in the interpretation of the upper barium study. While the degree of angulation at the aortomesenteric site is used as criteria with specific numerical cutoffs, there is no consensus among radiologists as to how the angle is measured. Consideration of surgery should involve clear communication between the radiologist and surgeon as to the findings on imaging.

Once conservative measures have failed and the patient can no longer maintain their weight within a healthy range, surgery should be considered. Surgical options include enteric bypass (side-to-side duodenojejunostomy or gastrojejunostomy) or mobilization of the duodenum at the ligament of Treitz (Strong's procedure).

Search Strategy

A literature search of English language publications from 1921 to 2014 was used to identity published data on surgical treatment of superior mesenteric artery syndrome. Databases searched were PubMed, Ovid, and GoogleScholar. Terms used in

the search were "Superior Mesenteric Artery Syndrome", "Wilkie's Syndrome", "Cast Syndrome", "Duodenal Ileus", "Aortomesenteric Compression Syndrome", "Duodenojejunostomy" AND ("Open" OR "Laparoscopic"), "Gastrojejunostomy" AND ("Open" OR "Laparoscopic"), "Strong's Procedure" AND ("Open" OR "Laparoscopic"), and "Duodenal Mobilization" AND("Open" OR "Laparoscopic"). Reference lists of the retrieved publications were manually reviewed for additional publications. We noted that majority of large, comprehensive series dated back to 1960–1980s, while most recently only small case series and case reports are available. The data was classified using the GRADE system (Table 23.1).

Results

Clinical Results of Duodenojejunostomy

Duodenojejunostomy was first described by Bloodgood in 1907 and performed by Stavely in 1908 (Fig. 23.2). It was shown to be a successful treatment option for SMA syndrome by Wilkie in 1921 [1]. Since, it has been the most frequently utilized operative procedure for treatment of this condition, having a published success rate of around 80% [23-25]. In 1978 Lee and Mangla published a review of 146 patients surgically treated for SMA syndrome, concluding that duodenojejunostomy had superior outcomes to both Strong's procedure and gastrojejunostomy [26]. Their quoted success rate was 90% in terms of symptomatic relief. In 1984, Gustafsson et al. published a 100 % success rate in ten patients treated with duodenojejunostomy [3]. In 1989, a case series of 16 operative patients showed the opposite results; only one patient achieved complete symptom resolution, while the only significant improvement was decreased frequency of vomiting in the others [4]. The first successful laparoscopic duodenojejunostomy was described in 1998 by Gersin and Heniford [27]. More recently in 2009, Merrett et al. described eight patients treated with duodenojejunostomy with duodenal division, reporting 100% success rate demonstrating no evidence of obstruction on imaging and weight gain in all eight patients post-operatively; however, the details of post-operative assessments and symptom resolution were omitted [14]. In 2010, Munene et al. published a literature review of nine case reports of patients with SMA syndrome treated with laparoscopic duodenojejunostomy reporting a 100% success rate for the operation in ten patients. However, follow-up data was lacking and the criteria used for determination of operative success was missing [28]. In 2012, Lee at al. published a

Table 23.1 PICO table

P (patients)	I (intervention)	C (comparator)	O (outcomes)
Patients with superior mesenteric artery syndrome	Duodenojejunostomy	Duodenal mobilization (Strong's procedure)	Symptom resolution

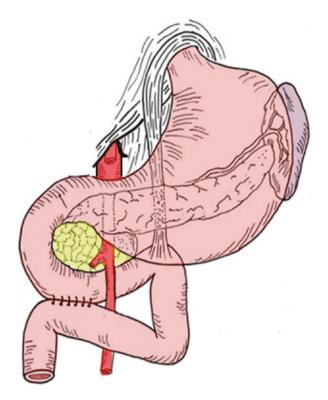


Fig. 23.2 Duodenojejunostomy

100% success rate for eight patients who underwent laparoscopic duodenojejunostomy and a 100% success rate for two patients who underwent open duodenojejunostomy [22]. Retrospective review by Pottorf et al. of 12 cases of SMA treated with laparoscopic duodenojejunostomy report 92% success in symptom improvement [29]. Most published studies suffer from a very small sample size, short follow-up, and lack of information regarding the criteria used to determine long term success (Table 23.2). Other small case reports consisting of one or two patients, revealed similar conclusions and suffer from the same lack of objective preoperative assessment tools compared to blinded postoperative assessment in the long term [30, 31, 35].

Clinical Results of Gastrojejunostomy

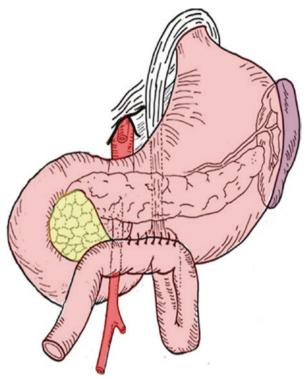
Gastrojejunostomy allows for gastric decompression; however, inadequate relief of duodenal obstruction may lead to failure of symptom resolution and complications such as blind loop syndrome, bile reflux and ulcers [14, 26] (Fig. 23.3). Even back

Table 23.2 Quality of follow up data in more recently published studies

	Number of		6 month outcome	Details of follow	Ouality of follow
Reference	patients	Intervention	reported	dn	up (0-3 points)
Gustafsson et al. [3]	10	Open duodeno-jejunostomy	Yes – successful in 10/10	Minimal	1
Ylinen et al. [4]	16	Open duodeno-jejunostomy	Yes – successful in 3/16 only	Detailed	3
Gersin et al. [27]	1	Laparoscopic duodeno-jejunostomy	No	Lacking	0
Richardson et al. [29]	2	Laparoscopic duodeno-jejunostomy	No	Lacking	0
Kim et al. [30]	2	Laparoscopic duodeno-jejunostomy	Yes – successful in 2/2	Minimal	1
Merett et al. [14]	8	Open duodeno-jejunostomy	Yes - successful in 8/8	Minimal	1
Singaporewalla et al. [31]	1	Laparoscopic duodeno-jejunostomy	No	Lacking	0
Munene et al. [28]	1	Laparoscopic duodeno-jejunostomy	No	Lacking	0
Lee et al. [22]	8	Laparoscopic duodeno-jejunostomy	Yes – successful in 8/8	Minimal	1
	4	Open duodeno-jejunostomy	Yes – successful in 4/4	Minimal	1
	2	Open gastro-jejunostomy	Yes – successful in 1/2	Minimal	1
Pottorf et al. [29]	12	Laparoscopic duodeno-jejunostomy	No	Lacking	0
Massoud [32]	4	Laparoscopic duodenal mobilization	Yes – successful in 3/4	Detailed	2
Villalba et al. [33]	1	Open duodenal mobilization	Yes – not successful	Detailed	2
Ha et al. [34]	19	Open duodenal derotatation	Yes – successful in 18/19	Detailed	2
Welsch et al. [6]	1	Open duodenal mobilization	Yes – successful in 1/1 Minimal	Minimal	0
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Quality of follow up is determined by following grading system: strategy for follow up and patient outcomes described (1 point), follow-up performed at or after 6 months from surgery (1 point), questionnaire used for post-operative symptom assessment (1 point). Scoring: 0 - Follow up absent, 1 - Poor, 2 - Fair, 3 - Good



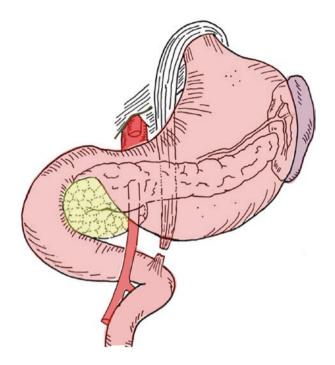


in 1921, Wilkie described treatment of SMA syndrome by gastrojejunostomy as a "mistake" due to "post-operative troubles" [1]. It has been largely abandoned as a treatment of SMA syndrome, but remains an option in cases where the other two procedures are deemed unsafe due to duodenal scarring or ulceration.

Clinical Results of Mobilization of the Duodenum

In 1958, Strong introduced lysis of the ligament of Treitz and lowering the duodenojejunal flexure away from the narrow aortomesenteric axis as a surgical option claiming the added benefits of a shorter duration of the procedure and avoidance of a bowel anastomosis [36] (Fig. 23.4). Over the next decade, the procedure was repeated infrequently [37, 38]. The disadvantage of this operation is its potential inadequate caudal displacement of the duodenum due to short inferior pancreaticoduodenal artery or adhesions leading to failure of symptom resolution and scar formation leading to symptom recurrence and potential increased difficulty at reoperation [24]. There is limited published data available for this procedure. In

Fig. 23.4 Strong's procedure (mobilization of the ligament of Treitz)



1995 Massoud reported a case series of four patients treated with laparoscopic release of ligament of Treitz with 75% success rate [32]. The largest recent retrospective review published by Ha et al. describes a modification to the procedure with mobilization of the right colon, terminal ileum and their respective mesenteries (duodenal derotation) in order to facilitate access to the third portion of the duodenum and to reduce the angular torque on the duodenum. They report nineteen adolescent patients who underwent the above procedure and quote a success rate of 95% in terms of symptom relief [34]. This is one of the largest and more complete retrospective reviews of the topic; however the utility of the additional steps of the operation remains questionable.

Recommendations Based on the Data

Due to the rarity of the superior mesenteric artery syndrome, randomized controlled studies are unavailable. By the late 1980s, most of the comprehensive literature on the topic had been published. Since then, the more current available literature includes mostly small case series and individual case reports.

The available limited results and their incomplete interpretation and analysis does not provide sufficient statistical power to allow for evidence based recommendations for one particular operation versus the other. Historically, duodenojejunostomy has been the preferred operation due to reports published in the early history of the disease. However, it is difficult, if not outright impossible, to determine the actual results of these studies, as most lack the appropriate criteria for follow up and tracking of symptom resolution in patients afflicted by SMA syndrome. An optimal study would require the following study elements: (1) objective pre-operative symptoms assessment via a comprehensive questionnaire-based assessment tool performed by a non-treating clinician, (2) a standardized consensus based diagnosis of SMA syndrome involving surgeon, radiologist, and gastroenterologist, standardization of the surgical procedure and (3) long term objective follow-up assessment using a multi-element assessment tool performed by a nontreating clinician. This long term assessment would include post-operative symptom resolution, discontinuation of prior medical treatments, significant weight gain, and re-imaging showing complete resolution of the obstruction and lack of any pre-SMA angle duodenal dilatation previously observed on imaging. Unfortunately, in general, most studies we reviewed fail to outline the criteria used to determine surgical success including the degree of symptom resolution.

Regarding SMA syndrome, laparoscopic duodenojejunostomy has been shown to be an effective and safe operation when performed by an experienced surgeon. It is the preferred method by many, including our group. It should be considered in patients with chronic symptomatology who have failed other approaches. Minimally invasive application of the Strong's procedure is a viable alternative in younger patients with acute onset of disease.

A Personal View of the Data

We recommend laparoscopic duodenojejunostomy for the surgical treatment of the SMA syndrome. In experienced hands, it is safe, simple and potentially curative. We believe it is the most direct and logical way to alleviate obstruction at the SMA angle and therefore it should theoretically have the lowest rate of recurrence since the actual obstructing lesion is completely bypassed. We recommend caution in diagnosing SMA syndrome and vigilance to avoid misinterpretation of the results of previously reported case studies.

Given the incomplete assessments of the long term results of one operation versus the other and the lack of accounting for the placebo effect of general anesthesia and surgery and the confounding variables of postoperative pain management and continuous medical management, we recommend a team approach to surgical treatment of patients with SMA syndrome.

Recommendations

- Recommendation for a thorough psychiatric assessment, especially eating disorders, prior to surgical intervention. Grade of recommendation 1C (evidence quality low; strong recommendation)
- Recommendation against performing a gastrojejunostomy for the treatment of SMA Syndrome, unless other approaches are not safe due to duodenal scarring or ulceration. Grade of recommendation 1C (evidence quality low; strong recommendation)
- Cannot recommend for or against laparoscopic duodenojejunostomy over duodenal mobilization for the treatment of SMA Syndrome. Grade of recommendation 2C (evidence quality low; weak recommendation)
- Recommend consideration of laparoscopic duodenojejunostomy in patients with chronic symptomatology who have failed other approaches. Grade of recommendation 2C (evidence quality low; weak recommendation)

Recommend consideration of laparoscopic duodenal mobilization procedure in younger patients with acute onset of disease. Grade of recommendation 2C (evidence quality low; weak recommendation)

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