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REVIEW ARTICLE

Laparoscopic options in superior mesenteric artery syndrome in children: systematic review

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Abstract

Purpose Superior mesenteric artery syndrome (SMAS) refers to the compression of the third portion of the duodenum between the aorta and the superior mesenteric artery, and usually results from weight loss or alteration in spine anatomy. This study reviewed laparoscopic options regarding SMA S in children.

Methods MEDLINE/PubMed was reviewed. Studies in patients under 16 years of age and published in English/Spanish were included, and selected by two independent reviewers. Data were collected for age, gender, weight/body mass index, comorbidities, symptoms, surgical technique, complications, conversions, recurrence and mortality. Descriptive statistics were used to analyze the quantitative portion of the study, with results presented as percentages, means and medians.

Results Twelve papers with $n = 16$ patients were included. Mean/median age were 10.7 and 13 years, respectively, with female predominance ($n = 11$, 68.75%). Emesis ($n = 15$, 93.75%) and abdominal pain ($n = 11$, 68.75%) were the most common symptoms. Anorexia nervosa was present in $n = 1$, and $n = 1$ patient had recent history of spinal fusion for idiopathic scoliosis. Regarding surgical technique, eleven cases consisted of duodenojejunostomy with side-to-side anastomosis (one associated with feeding jejunostomy); two Strong's procedures; and three Ladd's procedures. There were no conversions, and $n = 2$ (12.5%) minor complications (self-limited upper gastrointestinal bleed and persistence of vomiting for 48 h postoperatory). There was no mortality. Mean and median follow-up were 44.5 and 48 weeks, respectively, with no recurrences.

Conclusions SMAS is uncommon in children. The preferred laparoscopic approach is duodenojejunostomy, which can provide definitive relief of the obstruction with minor complications and low recurrence.

Keywords Cast syndrome · Duodenal obstruction · Laparoscopy · Mesenteric duodenal compression syndrome · Superior mesenteric artery syndrome · Wilkie's syndrome

Background

Superior mesenteric artery syndrome (SMAS) is an uncommon cause of upper gastrointestinal obstruction resulting from compression of the third part of the duodenum between the aorta and the superior mesenteric artery [1, 2]. It is also known as Wilkie's syndrome, Cast syndrome and mesenteric duodenal compression syndrome, and may result from weight loss or alteration in spine anatomy [1, 3, 4].

The classical presentation is recurrent postprandial pain, nausea, vomiting and early satiety [1, 5, 6], and first-line treatment is usually clinical management, which includes stomach decompression and nutritional support. Surgery is recommended if conservative therapy fails [5, 7].

This is the first systematic review on SMAS in pediatric patients, and the aim is to analyze laparoscopic options in this age group, with focus on surgical technique, complications, conversions, recurrence and mortality.

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Methods

Systematic search of MEDLINE/PubMed database was performed with the following terms: superior mesenteric artery syndrome/Cast syndrome/Wilkie syndrome/mesenteric duodenal compression syndrome and laparoscopy/laparoscopic. All published papers found were considered for this review, regardless the publication year. The inclusion criteria were reports with description of cases of SMAS in patients under 16 years of age, and papers published in English or Spanish. Exclusion criteria were: (a) unclear case documentation; and (b) full article unavailable. Before exclusion, the authors were contacted to clarify information or provide full text, but there was no reply in any case.

Eligibility assessment and data extraction were independently performed by two reviewers. Data were collected for age, gender, weight/body mass index (BMI), comorbidities, symptoms, surgical technique, complications, conversions, recurrence and mortality. Descriptive statistics were used to analyze the quantitative portion of the study, with results presented as percentages, means and medians.

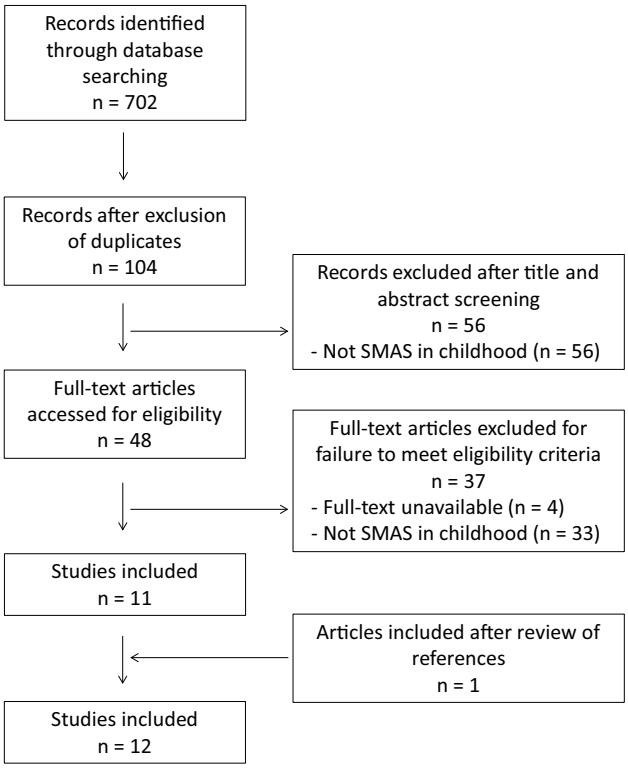

Fig. 1 Flowchart of the study literature search. SMAS: superior mesenteric artery syndrome

Results

The literature search revealed 702 papers, with 12 studies meeting the inclusion criteria [8–19], as detailed in the flow diagram (Fig. 1). All studies consisted of case reports or case series, as shown in Table 1. A total of 16 patients were included. The mean and median ages were 10.7 and 13 years, respectively (SD 5.5, range 6 months–16 years). There were five (31.25%) males and 11 (68.75%) females.

The mean and median periods from the onset of symptoms to surgical management were 620 and 270 days, respectively (SD 1045.6, range 4 days–10 years). In one case, the duration of symptoms was not mentioned and another report described an acute presentation. Emesis ($n = 15$, 93.75%) and abdominal pain ($n = 11$, 68.75%) were the most common symptoms (Fig. 2).

Associated conditions were present in three children (18.8%): one case of anorexia nervosa, another case of intrauterine growth restriction, right esotropia, familial short stature, congenital hip dysplasia and idiopathic scoliosis, and the third patient had precocious puberty, polycystic ovarian syndrome and recurrent right patellar dislocation.

Previous surgery was described in three patients (18.75%): one child with idiopathic scoliosis and recent history of spinal fusion, second patient was post-open extraction of stomach bezoar, and third was a case of cholecystectomy (performed three years prior to development of SMAS) with recent history of tooth extraction complicated by local abscess.

Regarding nutritional status prior to onset of symptoms, the three infants were above 50th percentile (considering weight for gender and age) [20]. In five other patients, weight loss was reported, but not specified: $n = 1$ after pneumonia treatment, $n = 1$ anorexia nervosa, $n = 1$ lost 13.6 kg after oral infection, and the loss/reason was not specified in $n = 2$. For eight patients, weight loss was not mentioned. Therefore, five patients (31.25%) were reported to have a history of weight loss prior to SMAS symptoms.

Management consisted of duodenojejunostomy with side-to-side anastomosis in 11 cases (one associated with feeding jejunostomy; eight using stapler, and two did not specify instruments); two Strong's procedures; and three Ladd's procedures (Table 1).

There were no conversions, and $n = 2$ (12.5%) complications ($n = 1$ self-limited upper gastrointestinal bleed and $n = 1$ persistence of vomiting for 48 h post-operatory). There was no mortality.

Table 1 Reports on superior mesenteric artery syndrome in children with regards to the publication year, number of patients, management and postoperative complications

Author	Publication year	Number of patients	Management	Postoperative complications
Castro et al. [16]	2018	2	Duodenojejunostomy	None
Bing et al. [10]	2017	3	Ladd's procedure	None
Bohanon et al. [17]	2016	1	Duodenojejunostomy	None
Kumar et al. [8]	2016	1	Duodenojejunostomy	None
Cullis et al. [9]	2015	1	Duodenojejunostomy	None
Rabie et al. [18]	2015	1	Strong's procedure	None
Record et al. [19]	2015	2	Duodenojejunostomy	Upper gastrointestinal bleed
Sato et al. [12]	2014	1	Duodenojejunostomy	None
Kurbegov et al. [13]	2010	1	Duodenojejunostomy	None
Makam et al. [14]	2008	1	Duodenojejunostomy	None
Palanivelu et al. [15]	2006	1	Duodenojejunostomy	None
Berchi et al. [11]	2001	1	Strong's procedure	Post-operatory vomiting

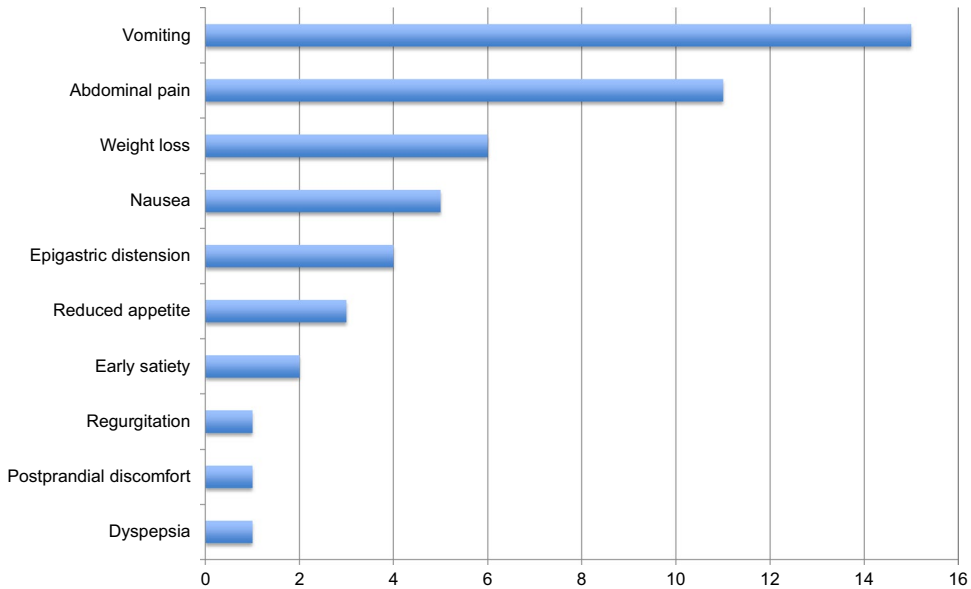

Fig. 2 Symptoms presented by the children with superior mesenteric artery syndrome included in this review

The follow-up period was mentioned in 13 cases, with mean and median period of 44.5 and 48 weeks, respectively (SD 32.5, range 2–136 weeks).

Discussion

SMAS results from the compression of the third part of the duodenum between the aorta and the superior mesenteric artery, that leads to marked narrowing of aortomesenteric angle and reduced aortomesenteric distance [1, 5, 8, 9].

The mesenteric fat and lymphatic tissue that surrounds the duodenum serves as a cushion, preventing this entrapment [1, 8, 21]. It has been postulated that any condition that predisposes the patient to weight loss, or if a patient gains height rapidly without a concomitant increase in weight, the fatty cushion around the artery diminishes, and may result in SMAS [1, 15]. Less than 32% of the patients included in the present study were reported to have had weight loss before the onset of SMAS symptoms.

Other factors that may narrow the vascular angle and contribute to the development of SMAS include surgical treatment of scoliosis, high insertion of the duodenum/short ligament of Treitz, peritoneal adhesions, duodenal malrotation, and low origin of the superior mesenteric artery [1, 2, 16, 22–24].

Genetic factors have also been proposed. Iwaoka et al. [25] reported SMAS in identical 28-year-old twins and Caspi et al. [24] reported a case with intrauterine manifestation of SMAS. Ortiz et al. [26] reported a family of eight, in which five members presumably suffered from SMAS. Our study also included a report of monozygotic 13-year-old

twins and three infants with SMA, supporting the genetic predisposition theory [10, 16].

Lee et al. [1] reported that almost 1/4 of their 80 SMAS patients with median age of 28 years had normal BMI. Similar to our findings, Biank et al. [6] reported no weight loss in 50% of their 22 cases of SMAS in children and young adults, suggesting that low weight/BMI and weight loss are not requirements for the syndrome.

Our results agree with previous observations that SMAS affects girls more often than boys, since almost 70% of our cases were female [1, 2, 6].

SMAS confirmation can be given by upper gastrointestinal series (UGI), Doppler ultrasound, computed tomography and magnetic resonance. [8, 12–14] In general, these exams show dilation of the stomach and proximal duodenum with signs of extrinsic compression of its third part and a decreased aortomesenteric angle [18]. Abdominal angiography can also be performed, but, as a much more invasive diagnostic tool, is rarely used [12].

There is no consensus on optimal therapy for SMAS in children, but the initial management is usually conservative [1, 2, 6]. The goal is to increase the mesenteric fat pad, enlarging the aortomesenteric angle. It may include gastric decompression, fluid resuscitation, anti-reflux therapy and nutritional support (post-pyloric high caloric feeding, parenteral nutrition, and/or oral diet if tolerated), with psychiatric consultation in cases of anorexia nervosa [8, 10, 17, 19]. Postural maneuvers and prokinetic agents have also been used [9]. The maneuvers can be "tested" on UGI, observing if prone, left lateral decubitus or knee–chest position relieves the obstruction [13]. Up to date, there are no irrefutable data on the appropriate duration of medical treatment before the consideration of surgical correction for children with SMAS [12, 16].

For patients unresponsive to conservative management, and for those with complications such as peptic ulcer, surgical options are available, and consist of bypass procedures or duodenal mobilization. They include open or laparoscopic duodenojejunostomy, gastrojejunostomy, Ladd's procedure and Strong's operation [4, 7, 12, 27].

Strong's procedure, the dissection of the ligament of Treitz with duodenal mobilization for lowering the duodenojejunal flexure, has the advantage to avoid anastomosis and allow for future bypass procedures if the patient continues to have symptoms. However, this approach has a significant failure rate, presumably due to the short branches of the inferior pancreaticoduodenal artery that do not allow the duodenum to fall inferiorly [4, 28].

Ladd's procedure has been proposed for SMAS as another anastomosis-free approach, and consists of right colon mobilization facilitating access, lysis of the retroperitoneal attachments of the third portion of the duodenum and widening the mesentery of the midgut. As a result, the third portion of the duodenum is rotated out of the superior mesenteric artery [10, 27].

Gastrojejunostomy has been abandoned, since it does not address the duodenal obstruction and is associated with significant complications including blind loop syndrome, dumping syndrome, and ulceration [27].

Laparoscopic duodenojejunostomy is usually the operation of choice in the treatment of SMAS, with a success rate up to 90% [1, 4, 29]. It was, as expected, the approach of choice for the patients included in this review. Complications were minor, and no recurrence or mortality was reported.

The present study has limitations: it included only papers published in English and Spanish, consisted of case reports and small series, and some reports contained incomplete data (weight/BMI/nutritional status, symptoms duration, surgical instruments used and follow-up period). The reports all mention good post-operatory evolution, but none detail the nutritional status during follow-up.

However, the strength of this study is the provision of much-needed recent information about laparoscopic options in pediatric SMAS.

Conclusions

This is the first systematic review on laparoscopic options for SMAS in children. Given its rarity and inspecific symptoms, the diagnosis requires a high suspicion index. It occurred more frequently in female patients, at a median age of 13 months, and 31% of surgical cases had a history of weight loss/low BMI before the onset of symptoms. Conservative management is the first option for most authors, and surgery is indicated if it fails or in the presence of complications. We found that laparoscopic approach is a safe and effective treatment for SMAS, duodenojejunostomy being the preferred technique, but more data are needed to establish if surgery should be considered earlier in the disease course.

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