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Superior Mesenteric artery syndrome: A systematic review

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Abstract

Superior mesenteric artery syndrome (SAMS) or Wilkie syndrome is a rare clinical condition, the incidence of superior mesenteric syndrome reported in some previous studies can range from 0.013% to 0.78% but the true incidence is unknown. This syndrome is characterized by extrinsic constriction on the anterior surface of the duodenum by the superior mesenteric artery and the abdominal aorta, leading to the formation of an aortomesenteric angle < 25°, and a decrease in the normal distance between the aorta and the mesenteric artery to 2-8 mm. Objective: Therefore, the objective of the present study is to carry out a systematic review of the literature to describe the superior mesenteric artery syndrome. Methodology: This study is a systematic literature review, with a qualitative approach, with interpretation and analysis of the elements obtained. The research was carried out from the survey of scientific articles found in the main search platforms for scientific articles: ScienceDirect, PubMed, SciELO, Bireme, from 2017 to 2022. Results and discussion: For this review 15 articles were selected as the studies showed that the symptoms of Wilkie syndrome are confusing, epigastric pain, vomiting, heartburn may be signs of other gastrointestinal pathologies such as gastroesophageal reflux, as presented in the literature. In chronic patients, non-operative therapy is often prolonged due to the long period of hospitalization, with a low success rate ranging from 14 to 71%. Strong's operation, gastrojejunostomy, and duodenujejunostomy are treatment options for Wilkie syndrome. Final considerations: The diagnosis of Wilkie syndrome often depends on a high index of clinical exclusion of other diseases, resulting in ineffective symptomatic symptoms. For this reason, a detailed history should be taken and after careful endoscopic evaluation, CT images should be analyzed carefully and surgery should be considered in cases of longer duration with nutritional support, always emphasizing the importance of individual assessment of each patient.

Keywords: Syndrome. Superior mesenteric artery. SAMS Wilkie. Surgery.

1. Introduction

Superior mesenteric artery syndrome (SAMS) or Wilkie syndrome is a rare clinical condition, initially described by Rokitansky in an anatomy book from 1842. It is also known as Wilkie syndrome, the author who reported it in 1921 when he published his first comprehensive clinical and pathophysiological study of 75 patients. It is estimated that by the year 1989 about 400 cases had been described in the medical literature. However, a high number of doctors are still unable to diagnose correctly, as it is an infrequent pathology and sometimes of an insidious nature, in most cases patients often spend a long period of time with a history of

abdominal complaints (Hines; gore; Ballantyne, 1984; Fan *et al.,* 2021).

The superior mesenteric artery syndrome has as main characteristics the extrinsic constriction in the anterior aspect of the duodenum by the superior mesenteric artery, and by the abdominal aorta, leading to the formation of an aortomesenteric angle <25°. Obstruction occurs when the acute angulation of the SMA causes compression of the third part of the duodenum between the SMA and the aorta, leading to an obstruction of the small intestine. Symptoms range from postprandial nausea and bilious vomiting to pain and weight loss, and the severity of symptoms depends on the degree of compression (Chrysikos *et al.,* 2019; Ganss et al., 2019;).

The incidence of superior mesenteric syndrome reported in some previous studies can range from 0.013% to 0.78% but the true incidence is unknown. Several factors are correlated to the formation of the aortomesenteric angulation. One of the most common factors listed is significant weight loss (loss of fat in the space between the arteries) that leads to retroperitoneal fat loss, anorexia nervosa, paraduodenal hernias, chronic malnutrition, and abdominal aortic aneurysm (Table 1). The disease is also commonly associated with serious debilitating illnesses such as cancerous tumors, malabsorption syndromes, AIDS, trauma, and severe burns. Another factor also mentioned is the performance of surgeries that distort the anatomy, when they change the position of the mesenteric artery, leading to a decrease in the angulation, causing duodenal compression between the vessels. Among the surgeries, scoliosis correction can be mentioned (Mandarry *et al.,* 2010; Ganss *et al.,* 2019; Fan *et al.,* 2021).

Conservative treatment of SMA syndrome consists of fluid resuscitation, total parenteral nutrition, and gastric decompression, nutritional support is essential and usually provided by providing small, frequent meals to increase mesenteric fat, thereby relieving obstruction, surgical intervention is required when therapy conservative approach fails and the standard surgical procedure is laparoscopic duodenojejunostomy. This surgery bypasses the segment of the duodenum that is compressed creating a connection between the proximal duodenum and the jejunum (De Rodríguez *et al.,* 2017).

In this syndrome, as already mentioned, there is a decrease in the aorto-mesenteric angle to up to 6-25°, with the expected normal angulation being 38-46°, and consequently there is a decrease in the distance between the structures, which goes from 10- 20mm, and in patients with the syndrome it becomes 2-8mm. Complications of frequent vomiting can lead to progressive dehydration, severe hypovolemia, oliguria, severe electrolyte alterations such as: metabolic alkalosis, hypocalcemia, lesions in the esophageal epithelium, even vomiting with blood from spontaneous bleeding from the gastrointestinal tract (Welch *et al.,* 2021).

In addition, other complications can be mentioned such as cardiovascular collapse, stomach perforations with the appearance of peptic ulcer, aspiration pneumonia and the patient can evolve to death if the diagnosis is delayed. The diagnosis of SAMS syndrome can be aided by radiological examinations, angiographic, ultrasound and endoscopic studies, however, despite advances in medicine, the diagnosis of this pathology still represents a challenge for clinical practice (Merrett *et al*., 2009; Warncke *et al*., 2019). Therefore, the aim of the present study is to perform a systematic review on superior mesenteric artery syndrome.

2. Methodology

This study is a systematic literature review, with a qualitative approach, with interpretation and analysis of the elements obtained. The guiding question of the study was: “Superior mesenteric artery syndrome, challenges and advances”, after which the following steps were taken to identify the theme, select the hypothesis or research question, establish criteria for inclusion and exclusion, define the information and clinical studies to be included in the present review study, interpretation of results and presentation of the review with synthesis of knowledge.

First, the theme was defined to start the elaboration of the integrative review and proceed to the subsequent stages of the study, the guiding question was formulated (Mendes; Silveira; Galvão, 2008). Thus, in relation to the chosen theme, the guiding question was: Superior mesenteric artery syndrome, what are the challenges and main advances in the treatment of the disease?

The research was carried out from the survey of scientific articles found in the main search platforms for scientific articles: ScienceDirect, PubMed, Scientific Electronic Library Online (SciELO) and Latin American and Caribbean Center for Health Sciences Information (BIREME), the following databases were used: US National Library of Medicine (PUBMED).

Using the descriptors present in the Descriptors in Health Sciences (DeCS) which are: Superior mesenteric artery syndrome, treatment, diagnosis in Portuguese and English with the Boolean operator "e" in all bases, eventually associating the terms with the use of quotation marks (““). Considering the need to perform a broader search, these keywords and the Boolean operator were always used in the search field related to the abstract. These descriptors were combined in order to find as many publications as possible suitable for the review. The review was done from October 2021 to August2022.

For the collection of articles, the search limits were chosen (free full text, 6 years, humans) to specify the results and thus collect pertinent information to support the theme in question. Thus, in the first search, 1808 articles were found in Search I, after applying the filters in Search II, the search returned 516 articles and from these, 15 articles were selected for the systematic review. In the SCIELO database, from the Search II criteria, 15 articles were found, only 1 fit the inclusion criteria, however the same had already been found in another database. After the selection, the titles, keywords and abstracts of all publications resulting from the investigation carried out by the search strategy were carefully read in order to analyze the texts adequate to the previously established inclusion criteria and filters. All 15 selected articles were read in full to properly verify the purpose of the study, including clinical studies, cohort studies, reviews and meta-analyses.

In the last phase, the articles were analyzed and discussed in order to extract all the information that was relevant to the research for the formulation of the work, and all the questions were raised and propositions for the writing of the scientific work were made for the sedimentation of knowledge.

In this phase, the synthesis of the results was carried out through the analysis of the scientific material from the evaluation of the results and conclusions drawn from the selected articles. The document's formulation authorizes readers to assess the veracity of the procedures used in carrying out the review in question.

3. Results and discussion

The superior mesenteric artery (Figure 1) originates from the abdominal aorta, at the level of the intervertebral disc between the first and second lumbar vertebrae. The branches departing from the SMA are: jejunal and ileal arteries, ileocolic artery, right colic artery, middle colic artery and inferior pancreaticoduodenal artery. Briefly, this artery runs in its inferior course anteriorly to the uncinate process of the pancreas and to the third part of the duodenum, posteriorly located to the splenic vein, the body of the pancreas and the left renal vein, which separates it from the abdominal aorta. The superior mesenteric artery also passes anteriorly to the inferior vena cava and also to the right ureter and the right psoas major muscle, alongside the small intestine mesentery (Lippl *et al*., 2002; Ganss *et al*., 2019; Fan *et al*., 2021).

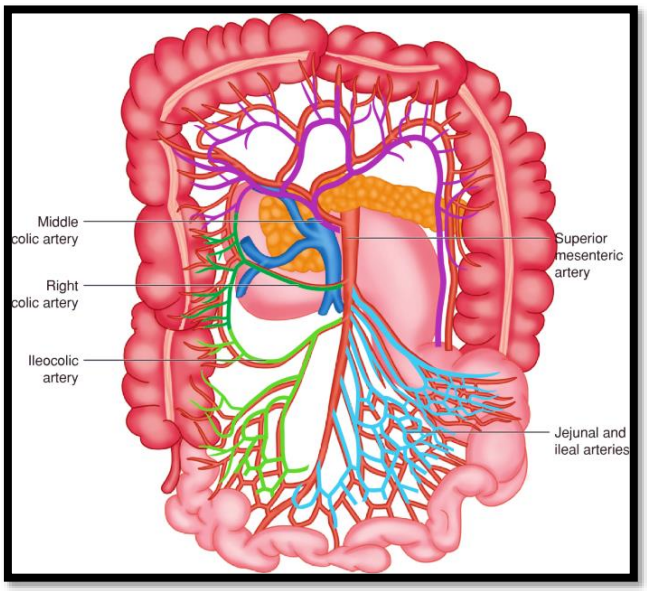

Figura 1- Superior mesenteric artery anatomical division (Khattab *et al*., 2020).

SMAS is characterized by obstruction of the gastrointestinal tract due to compression of the third part of the duodenum between the abdominal aorta and the superior mesenteric artery. The disease may also be known as mesenteric duodenal arterial compression, Cast syndrome or Willkie syndrome, aortomesenteric clamp, chronic duodenal ileus, plastered corset syndrome or duodenal vascular compression, the latter being very appropriate due to the role played by the superior mesenteric artery. on the duodenum in the etiopathogenesis of the disease (De Rodríguez *et al*., 2017; Welch *et al*., 2021).

Pathophysiologically, there is an intermediate mesenteric fat loss between the aorta and the SMA, which results in a narrower angle between the vessels leading to duodenal compression. The layer of fat keeps the SMA away from the spine, thus preventing duodenal compression. The normal aortomesenteric angle ranges from 38 to 65°. However, among SAMS patients, this angle reduces to less than 25º, which in turn reduces the distance to less than 10 mm (Biank; Werlin, 2006; Sinagra *et al*., 2018; Ganss *et al*., 2019).).

Important risk factors have already been identified such as: rapid growth in children, dietary conditions (anorexia nervosa and disease), hypermetabolism (trauma and burns), cachexia causing conditions such as AIDS, paraplegia and cancer. Other risk factors include abdominal aortic aneurysm, short or hypertrophic

Treitz ligament, duodenal malrotation, Ladd bands, hyperlordosis, visceroptosis, abdominal wall laxity, peritoneal adhesions, retroperimental tumors and catabolic states, mesenteric root neoplasia, surgical correction of scoliosis and adhesions (Welsch; Büchler; Kienle, 2007; Sinagra *et al*., 2018).

Treatment can be conservative or surgical, either way, the correction of hydroelectrolytic priority syndromes and/or nutritional deficiencies, in a special case, is da. The conservative therapeutic approach can be proposed as an initial therapeutic strategy in all patients, although it is usually reserved for patients with more latent symptoms and of significant importance to patients. It includes identification and correction of precipitating factors; Appropriately associated with prokinetics to achieve hypernutrition and weight gain, preferably orally, enterally (via nasojejunal tube passing through an obstruction) or parenterally; and correct positioning after meals (lateral decubitus (Recio-Barbero et al., 2019; Bronswijk et al., 2021).

Surgery is indicated in symptomatic patients when conservative treatment fails. There is no clear time limit on the duration of medical treatment, as the treatment time for symptoms has been observed to be 12 days, but it can last up to 2 days. Duodenal atony after massive dilation may persist even after duodenal decompression and delay normal gastrointestinal function (Warncke *et al.,* 2019).

Surgical treatment is indicated after failure of conservative treatment and in long-term adult patients in whom inpatient medical treatment has a low probability of success. There are several techniques, one of them is Strong's Technique (Figure 2) which was described for the first time the division of the Treitz ligament with mobilization of the transverse and ascending duodenum for caudal displacement of the duodenum (Pillay, 2016; De Rodríguez *et al.,* 2017).

As advantages of this one are that it does not violate the intestine and therefore it is the least invasive, quickest and safest procedure. It has been correlated with earlier postoperative recovery. How inferior correction can occur or due to corrections and how can the displacement of the duodenum caudal occur, due to the possibility of aggravation with the correction of the pancreaticoduo vessel to the duodenum, due to a failure rate of 25%. (Pillay, 2016; De Rodríguez *et al.,* 2017; Recio-barbero *et al.,* 2019; Zhang, 2018; Hamidi *et al.,* 2019).

Figure 2- Strong's procedure. Strong's procedure mobilizes the duodenum by dividing the ligament of Treitz.

Once the duodeno-jejunal junction is mobilized, the duodenum is positioned to the right of the superior mesenteric artery (Pillay, 2016).

Another type of surgery also performed in patients with SAMS is gastrojejunostomy (Figure 3). The surgery is performed with a side-to-side anastomosis that is performed between the stomach and a jejunal loop. It allows gastric decompression in patients with significant abdominal distention, but does not relieve the obstruction to which the third portion of the duodenum is subject, which is why recurrences are frequent. Persistent obstruction can lead to blind loop syndrome, gastric bile reflux and ulceration (Bronswijk *et al.,* 2021).

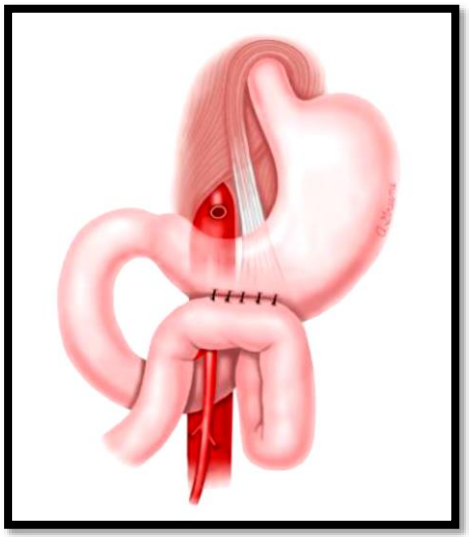

Figure 3- Gastrojejunostomy with side-to-side anastomosis (Sah *et al*., 2020).

Side-to-side or Roux-en-Y duodenojejunostomy can also be mentioned as a surgical procedure for SAMS, which was described for the treatment of this pathology in 1908 by Stavely. It is admitted that it presents better results than the previous ones, but it also favors the development of bacterial overgrowth. To avoid this, the mobilization of the fourth duodenal portion is associated, performing the side-to-side duodenojejunostomy with the proximal portion (Figure 4) (Barkhatov et al., 2018).

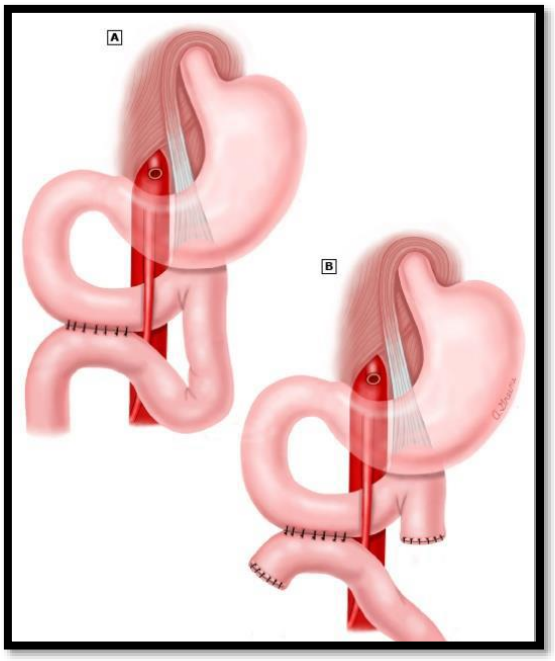

Figure 4: Latero-lateral duodenojejunostomy without separation of the 4th duodenal portion (A) and with separation (B) (Pillay, 2016).

Surgical therapy is indicated when: a) conservative treatment has not resulted, b) prolonged disease with progressive weight loss and duodenal dilatation with stasis, c) in complicated peptic ulcer and pancreatitis secondary to stasis and reflux bile, d) other local pathologies requiring laparotomy, e) the patient's preference for surgical correction, rather than prolonged conservative procedures (Wang, Takada, 1984; Pillay et al., 2016). Despite the disease having been described in 1842, the literature is still relatively lacking in complete studies on the disease.

In the present study, 15 articles were selected, Table 1 shows the distribution of articles, according to authors, titles and type of study, all of these articles were published in the ScienceDirect, PubMed, Bireme and Lilacs databases. One of this studies the Alzerwi's research (2021), was a retrospective hospital-based case-control study conducted over a period of 2018-2020 at the Department of Gastroenterology at Qimat Rai Gupta Central Hospital, Haryana. Case records of 2,100 patients who visited the health facility with chronic or acute pain in the abdomen were selected. Of these, there were 1,524 cases of acute abdominal pain and 576 cases of chronic abdominal pain. Superior mesenteric syndrome was suspected among 10 cases, who were advised contrast-enhanced computed tomography (CBT). Of these, only seven patients underwent investigations and were confirmed to have SAMS syndrome.

In a period of two years, the authors found 2,100 records of cases that underwent esophagogastroduodenoscopy, of which 07 cases were confirmed as having mesenteric artery syndrome. Thus, the prevalence of SMA syndrome was found to be 0.3% among patients who arrive with acute or chronic abdominal pain, data that are in accordance with the frequency described in the literature (Alzerwi, 2021). Demographic and clinical characteristics of SMAS patients in this study were as follows: 4 had dysmotility with dyspepsia, unexplained weight loss (n=2), and reflux with dyspepsia (n=1). Five patients had

comorbidities such as spina bifida (n=2), anorexia nervosa (n=2) and Crohn's disease (n=1). Regarding the duration of symptoms, patients with acute abdominal pain, chronic abdominal pain and those diagnosed with SAMS was the mean duration of symptoms onset was 8 (6-12) months , The median age of patients in Group I was 26 (23-38) years, The proportion of women with SAMS corresponded to 71.4% (n=5), the median body mass index (BMI) was 20 (18-22) kg/m2 (Alzerwi, 2021)

Tabela 1- Select Articles

Authors	Title	Year of publication	Type of study
FARINA, *et al.*	Super ior mesenter ic artery syndrome (Wilkie Syndrome) with unusual clinical onset: Description of a rare case ☆	2021	Case report
CHRYSIKOS, D. *et al.*	Superior mesenteric artery syndrome: a rare case of upper gastrointestinal obstruction.	2019	Case report
SINAGRA, E. *et al.*	Superior mesenteric artery syndrome: clinical, endoscopic, and radiological findings.	2018	Case report
YILMAZ, T. U, *et al.*	Laparoscopy versus laparotomy for the management of penetrating abdominal trauma: a systematic review and meta-analysis.	2016	Case report
GANSS, A. *et al.*	Superior mesenteric artery syndrome: a prospective study in a single institution. Journal of Gastrointestinal Surgery	2019	Retrospective observation study
ARAUJO, A. O. *et al,*	Superior mesenteric artery syndrome–An uncommon complication following surgical corrections of spinal deformities.	2021	Case report
ALZERWI, N.	Predictors of Superior Mesenteric Artery Syndrome: Evidence from a Case-Control Study	2020	Retrospective observation study
ZARAKET, V.; DEEB, L.	Wilkie's syndrome or superior mesenteric artery syndrome: fact or fantasy. Case reports in Gastroenterology,	2017	Case report
ZHANG, R *et al.*	Diagnosis and treatment of superior	2021	Retrospective

	mesenteric artery compression syndrome complicated with gastroesophageal reflux disease.		observation study
FARINA, R *et al.*	A man with the rare simultaneous combination of three abdominal vascular compression syndromes: median arcuate ligament syndrome, superior mesenteric artery syndrome, and nutcracker syndrome.	2021	Case report
SILVA, A. L *et al.*	Epigastric pain and weight loss–A case of Wilkie's syndrome.	2020	Case report
YILMAZ, T. U *et al.*	Superior Mesenteric Artery Syndrome-Process of Diagnosis and Treatment of Problematic Cases.	2017	Retrospective observation study
WELCH B *et al.*	Superior Mesenteric Artery Syndrome: A Community Hospital Case Series.	2021	Retrospective observation study
ZEE, K.; STEPHENS, M.; FABISZAK, M.	Superior Mesenteric Artery Syndrome: Could it Involve a Potential Familial Pattern?.	2021	Case report
SALEHZADEH, F.; SAMADI, A.; MIRZARAHIMI, M.	Superior Mesenteric Artery Syndrome in a 6-Year-Old Girl with Final Diagnosis of Celiac Disease.	2019	Case report

Computed tomography findings of constraint showed that the median aortomesenteric angle in Groups I (SAMS patients), II (Acute abdominal pain) and III (chronic abdominal pain) was 22 (20-24)º, 52.5 (43.5 - 58.5) º, 52 (43.5-58.5) º respectively. Likewise, the distance from the aortomesenteric artery found was 6 (5-6) mm, 11 (11-12) mm and 11 (10.5-11.5) mm, that is, the median aortomesenteric angle and the distance were significantly lower in Group I compared to Group II (p value <0.001) and Group III (p value <0.001). A statistically significant positive correlation was found between BMI and aortomesenteric angle and distance (Table 4). This is an important finding as well as a new aspect of this study (Alzerwi, 2021).

Likewise, the aortomesenteric angle and distance in Group I was smaller than in both groups. This indicates that as a patient's BMI decreases, there is a corresponding reduction in SAMS angle and distance as reflected by correlation coefficients. Thus, this finding presents a window of opportunity for hypotheses that the gastroenterologist or assistant physician may suspect to perform more specific tests to confirm the syndrome. Rather than just relying on endoscopic findings to guide whether or not to suspect the presence of SAMS syndrome, a reduced BMI along with a detailed history of numerous risk factors for increased weight loss, metabolic diseases, or a history of scoliosis (Table 2) should ring the bell for the possibility of the

syndrome (Alzerwi, 2021).

Table 2- Predisposing conditions for the development of superior mesenteric artery syndrome (Welsch Büchler; Kienle, 2007; Fan *et al*., 2021).

Risk Factors
Chronic debilitating illnesses: **Cancer** **cerebral palsy** **Paraplegia** **juvenile rheumatoid arthritis** **cardiac cachexia** **Drug abuse**
Trauma: **burn injuries** **multiple injuries**
Eating disorders: **anorexia nervosa** **malabsorption** **food intolerances**
Postoperative states **Bariatric surgery** **Proctocolectomy and ileonal pouch anastomosis** **Nissen fundoplication** **Aortic aneurysm repair** **Spinal instrumentation, scoliosis surgery**
Anatomical and congenital anomalies: **High insertion of the ligament of treitz** **Intestinal malrotation, peritoneal adhesions** **Low origin of superior mesenteric artery** **Increase in lumbar lordosis** **intestinal malrotation**
Local pathology: **Neoplastic growth in the mesenteric root** **Dissecting aortic aneurysm**

The research by Ganss et al (2019) was carried out with the aim of prospectively investigating 39 patients with a history of chronic refractory gastrointestinal disorders, who underwent surgical correction for SAMS, in order to discuss, the clinical presentation of these patients, diagnosis and surgical outcome in a

long-term follow-up.

The researchers screened between October 2008 and March 2016, 254 consecutive patients with symptoms of digestive diseases, lasting at least 5 months, these patients were referred to the Department of Surgery, Oncology and Gastroenterology of the University Hospital of Padua (Italy), a referral center for complex upper digestive surgery. We excluded 215 (85%) patients based on the following criteria: inability to provide informed consent (2.8%); malignancies (7.4%); intestinal motility disorders (57.2%); psychiatric illness (9.3%); pregnancy (2.3%); impossibility of carrying out the necessary diagnostic investigation (20.9%) Thirty-nine enrolled patients underwent duodenal surgery by clinical and radiological criteria. They presented with one or more of the following clinical features: (a) severe and frequent upper digestive symptoms (occurringat least once a week), associated with poor quality of life and refractory response to medical treatment; (b) a condition of low weight (BMI < 18.5 kg/m2) associated with difficulty eating; (c) serious complications of SMAS (eg, gastric perforation, acute pancreatitis, aspiration pneumonia) (Ganss *et al.,* 2019).

In all patients, the following features were recognized on imaging studies: (a) findings suggestive of SMAS in Barium Swallow; (b) diagnostic aortomesenteric angle and distance on CT/MR angiography. Typical findings in barium swallow are gastrectasia, proximal duodenal dilatation, an abrupt vertical or oblique compression of the third portion of the duodenum, duodenogastric reflux, and delayed transit (GANSS *et al*., 2019).Thirty-four (87%) of the patients with SAMS were idiopathic, while 5 female patients (13%) reported a possible risk factor in their medical history. Two of the 5 patients had a history of an eating disorder (anorexia and bulimia, respectively), 2 patients suffered from ligament laxity (Marfan and Ehlers-Danlos syndrome), while one patient had a J-shaped ileoanal pouch proctocolectomy performed at another center. A single case of familial correlation (mother and child) was also reported (Ganss *et al.,* 2019).

Thirty-four patients (87%) reported severe weight loss, leading to an underweight condition or severe malnutrition in 22 of them. Eight patients (21%) suffered an episode of acute pancreatitis and required surgical or endoscopic treatment (cholecystectomy, endoscopic sphincterotomy) in 2 of them. Radiological and endoscopic data show that 22 (57%) patients had gastroduodenal dilatation and delayed gastroduodenal emptying in 15 (38%). On CT/MR angiography, the median aortomesenteric angle was 11° (8–15), and the aortomesenteric distance was 5 mm (4–7). In addition, UDE was also 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 (Ganss *et al.,* 2019).

The procedure chosen for the surgical approach was the Duodenojejunostomy. All patients underwent special x-ray tests used to examine their throat, esophagus, stomach and the first part of the intestine. After surgical treatment, patients continued to be followed up for an average of 47 months (34-72), a significant increase in BMI was recorded and in 28 cases (72%), in addition, the need for therapy for reflux and prokinetic therapy with that of D2 receptor antagonists, significantly reduced after surgery. A positive response to surgical treatment was considered to be a reduction in the symptom score of more than one third of the initial value, an increase in weight and in the value of BMI, as well as an improvement in postoperative quality of life. Regarding patient satisfaction, 69% (27) reported satisfaction with the surgery, 13% neutral (n=5), 18% dissatisfied (n=7). Among the 7 patients who were dissatisfied with the postoperative diagnosis and of these

still had severe gastrointestinal dysmotility (n = 5), and 2 had unspecified psychiatric illness (Ganss *et al.,* 2019).

In another study by Sinagra *et al* (2018) where they prospectively evaluated 10 cases of SAMS (2 men, 8 women), with a prevalence of 0.005%. Median age was 40 years (range 14-40), and BMI was 21.5 kg/m2, patients had symptoms between 6 and 24 months (mean 18 months). In the group of patients with the disease, the symptoms reported were: postprandial discomfort syndrome (epigastric pain and discomfort, nausea and vomiting), dyspepsia" (p = 0.02), according to the Rome IV criteria, and loss of weight (median weight loss before diagnosis was 6 kg), while in the control group, the most common presentation was "epigastric pain and dyspepsia syndrome" (p = 0 01), according to Rome IV criteria, with a loss less marked weight loss (median weight loss before diagnosis was 0.5 kg).

Predisposing conditions were present in 5 patients (anorexia nervosa in 2 patients and G6PDH deficiency, spina bifida and Crohn's disease in 3 patients), the findings revealed a median aortomesenteric angle of 22° (p = 0.001), and the median aorta distance -SMA was 6 mm (p < 0,001). All patients in this study had an improvement in conservative treatment, despite the surgical consultation proposed for each patient. Treatment strategies involved strategies such as nasogastric decompression and hyperalimentation followed by frequent feeding and small meals, through clinical follow-up under the supervision of a gastroenterologist and a nutritionist (Sinagra *et al.,* 2018).

In conclusion, the authors showed that, as in other reports in the literature, a marked weight loss and a symptomatology of severe gastric symptoms for more than 8 months are significantly related to a diagnosis of SAMS syndrome confirmed on contrast-enhanced CT after endoscopic suspicion. Majority of patients were female, ranging from children, adolescents and even adult individuals with a history of sudden weight loss, all patients in this study opted for the conservative treatment that can be successful in acute cases of compression, a hypercaloric diet that takes weight gain can alleviate or even remove symptoms. The objective of the conservative approach with jejunal or parenteral nutrition is the restoration of the aorthomesenteric adipose tissue that in the normal individual displaces the SMA previously away from the aorta, thus avoiding duodenal compression.

A retrospective survey by Zhang *et al.* (2021) found 30 cases (eight males and 22 females) with a male-to-female ratio of 4:11, and the mean age was 41.3 ± 16.4 years. The body mass index (BMI) was 18.7 kg/m2 (15.6, 23.3 kg/m2), and the disease course was 4.0 years (2.1, 30.5 years) . The main symptoms were anorexia in 18 cases, heartburn in 17 cases and acid regurgitation in 14 cases. CT showed that the angle between the abdominal aorta and the superior mesenteric artery was between 10° and 22°, twelve patients who received only drug treatment (drugs for gastrointestinal motility, gastric mucosal protectors) and nutritional support demonstrated relief of their symptoms (Zhang *et al.,* 2021).

Laparoscopic Toupet Fundoplication combined with Treitz releasing ligament (LOTR) was performed in 18 remaining patients, the postoperative and hospital stay was 7.0 days (5.0, 19.0 days). After 2 years of follow-up, the patients' BMIs were significantly higher to an average of 20.8 kg/m2 (18 before surgery to 24.7 kg/m2).According to the patients' assessment of the curative effect of the surgery, the total effective rate was 100%, with 17 cases (57%) being cured and 13 cases (43%) considering the treatment effective. Therefore,

these data show that SAMS combined with GERD seriously affects an individual's quality of life. Thus, appropriate treatments must be selected based on the clinical picture and characteristics of each patient. Favorable results obtained after laparoscopic Toupet Fundoplication combined with ligament of Treitz release confirming that this is an appropriate and effective surgical procedure for SAMS combined with GERD (Zhang *et al.,* 2021).

Two other reported cases were of a 54-year-old female who was vomiting after ingesting solid foods and liquids and had lost 12 kg in 4 months and a 68-year-old white female patient who presented to the hospital with abdominal pain and concomitant loss of 15 kg. in the last 6 months that was unintentional, patient 1 underwent longitudinal ultrasound to study the abdominal aorta and superior mesenteric artery.

The examination showed a reduction in the aortomesenteric area angle (15°) measured approximately 1 cm from the bifurcation, and a consequent decrease in the perivascular fat that surrounds the abdominal aorta and the upper portion of the mesenteric artery (diameter 2 mm). The patient followed a hypercaloric diet and 2 months later, after the disappearance of emetic symptoms, another ultrasound showed an aorto-mesenteric angle of 32° and an increase in the thickness of the adipose tissue (diameter 8 mm) (Farina *et al.,* 2017; Chrysikos *et al.,* 2019).

Patient 2 after confirming the diagnosis of SAMS by CT of the upper and lower abdomen showed an acute angle of 15°, however she was unwilling to attempt nasojejunal feeding or total parenteral nutrition feeding. The patient underwent laparotomy, the duodenum was obstructed at the point where the superior mesenteric artery crossed the third part of the duodenum at the time of surgery the patient presented hemodynamic instability and the Strong procedure was performed with rupture of the ligament of Treitz and mobilization of the duodenum, however one week after the first operation (Sinagra *et al.,* 2018; Chrysikos *et al.,* 2019).

Subsequently, she underwent a new laparotomy where a Gastrojejunostomy was performed with latero-lateral anastomosis, and after one month the postoperative examination showed the unobstructed passage of the stomach contents to the jejunum (Fig 7). Total parenteral nutrition, as previously discussed, has also been widely used, but eventually 50-70% of all cases will relapse and may require surgery, physicians at the time of patient 2's surgery performed after mobilization of the duodenojejunal flexure dividing it. if the ligament of Treitz (Strong procedure) as it happened in this case, this surgical option fails in 25% of patients (Sinagra *et al.,* 2018; Chrysikos *et al.,* 2019).

As already highlighted, the diagnosis of the syndrome is not always performed correctly, which was what happened with the reported clinical case Silva et al. (2021). The patient was a 46-year-old female who presented to the outpatient clinic of our internal medicine service with a 2-year history of epigastric pain, nausea, early satiety, and weight loss of 15 kg. Postprandial pain was also observed. On examination, the patient was emaciated, had epigastric pain and weighed 45 kg (Sinagra *et al.,* 2018; Chrysikos *et al.,* 2019).

The patient had been previously evaluated by gastroenterology, by general surgeons and psychiatrists, who had performed an exhaustive study, but with inconclusive results. Upper endoscopy and colonoscopy showed esophagitis, which persisted after treatment. An abdominal computed tomography (CT) scan was inconclusive. Laboratory tests were normal. The symptoms were then attributed to an anxiety disorder and the

patient was prescribed antidepressants, with no clinical improvement. Soon after, the patient underwent CT enterography, which revealed distention of the proximal duodenum and compression of the third portion of the duodenum between the aorta and the superior mesenteric artery (aortomesenteric clamp), with an aortomesenteric distance of 5 mm and an angle of 18°. .6°. These findings were consistent to confirm the diagnosis of SAMS, the work did not show the treatment chosen or the approach that was performed on the patient (Silva *et al.,* 2021).

An equally difficult prognosis was reported by Yılmaz et al (2019) who describe a 25-year-old woman referred to our outpatient clinic with epigastric pain, postprandial discomfort, swelling, bilious vomiting, and inability to gain weight. Her symptoms improved with postural change to knee position. In her medical history she has suffered from abdominal pain and retrosternal burning and swelling. She had undergone Nissen Fundoplication 2 years ago. However, 1 month later after the procedure the symptoms of postprandial bloating and discomfort worsened. She underwent laparotomy with suspected paralytic ileus.

No pathology was found, but segmental small bowel resection and anastomosis were performed due to iatrogenic injury. Two months later, the patient was operated on and a bridectomy was performed. Her symptoms did not improve, she was hospitalized for 10 days with a diagnosis of acute pancreatitis and received therapy with proton pump inhibitors. The patient was also consulted in psychiatry with suspected anorexia neurosis and was followed up by the psychiatrist for 6 months. When she was admitted, she had been on a liquid diet for 3 months and her BMI was 15.4. In the endoscopic evaluation, a megoduodenum and non-digestion of food residues in the duodenum were observed (Figure 5). With the suspicion of SMAS, measurement of the aortic angle was performed and SAMS was confirmed. The patient was treated with a side-to-side or Roux-en-Y duodenojejunostomy and gained 8 kg of weight at follow-up at six months (Yılmaz *et al.,* 2019).

Welch *et al.* 2021 evaluated Thirteen patients diagnosed with SAMS The median age at diagnosis for this cohort was 29 (24-43) years. Four patients were male and nine were female. Median duration of symptoms was 180 days. The mean mesenteric aortic angle was 23.75 (±10.0) degrees, the mean (±SD) of the aortomesenteric angle was 5.4 (±1.9) mm, and the mean BMI (±SD) was 21.7 (±3.1) kg/m2. Nine of the patients were managed conservatively (69.9%) and four patients required surgical intervention (30.1%, three duodenal-jejunostomies 75% and a Strong procedure 25%) showing once again that not all patients may receive the same choice of treatment, as limitations the authors pointed out that they were not able to obtain patient data after treatment.

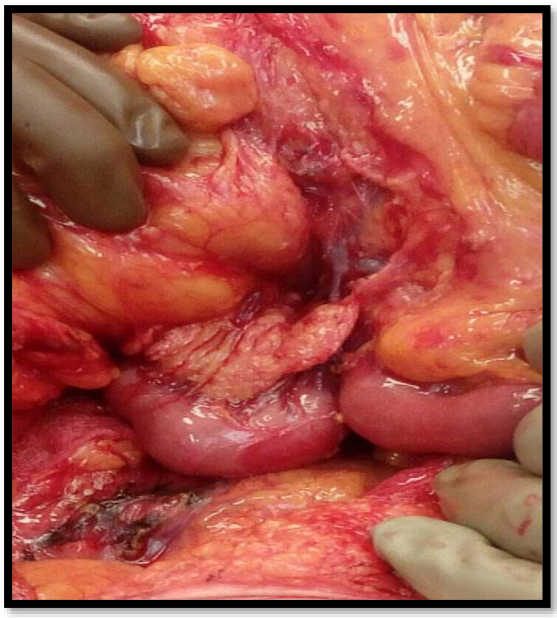

Figure 5 - SMA syndrome compression of the third part of duodenum (Chrysikos *et al.,* 2019).

Another point that was discussed in one of the works was the genetic part of the syndrome. The patient had a family history of SMA syndrome in her mother, with a surgical history of cholecystectomy and hysterectomy. Computed tomography of her abdomen was performed, which revealed a severely distended small intestine and dilated duodenal C-loop to the level where the duodenum crosses between the aorta and the SMA (Figure 1). Three days after admission to the hospital, the patient underwent a side-to-side or Roux-en-Y duodenojejunostomy. SMA syndrome is currently not considered an inherited condition, however the condition has been observed in twin studies indicating the possibility of a family inheritance pattern of the syndrome. More studies should be performed to correlate the condition with a congenitally shortened ligament of Treitz or a point of inferior origin of the SMA (Zee; Stephens; Fabiszak, 2021).

One of the reported cases was of a patient who did this methamphetamine to lose weight quickly, she was 19 years old suffered from bipolar disorder and had a weight loss of 27.7 kg last year with about 13.6 kg lost in the last year. last month. Physical examination showed a BMI of 18.03 kg/m2, decreased sounds and diffuse abdominal tenderness. CT revealed severely distended stomach and proximal duodenum with air-fluid levels and midline dilated to uncompressed duodenum transition point as the third portion of the duodenum passed inferiorly with an aortomesenteric angle measured 8 degrees (limit normal: 38-65 degrees) with a aortomesenteric distance of 6.0 mm (normal limit: 10-28 mm), which was compatible with SAMS syndrome (Johnson; Paladugu, 2019).

The patient admitted to restrictive eating behavior in the months prior to the use of weight loss drugs, her symptoms began to improve over the course of her hospital stay. On day 4 of her admission, the nasogastric tube was discontinued and her diet progressed as tolerated with no return of symptoms. She was discharged with psychiatric primary care and follow-up, these data once again confirm the main risk factor is sudden weight loss and anorexia (Johnson; Paladugu, 2019).

And, in conclusion, the symptoms of Wilkie syndrome are confusing, epigastric pain, vomiting,

heartburn can be signs of other gastrointestinal pathologies such as gastroesophageal reflux as presented in the literature. In chronic patients, non-operative therapy is often prolonged due to the long period of hospitalization, with a low success rate ranging from 14 to 71%. Strong's operation, gastrojejunostomy, and duodenujejunostomy are treatment options for Wilkie syndrome. The diagnosis of Wilkie syndrome often relies on a high index of suspicion and is often made by a process of exclusion, resulting in ineffective symptomatic symptoms. inadequate therapies and investigations, this delay in diagnosis can lead to a delay in treatment resulting in unnecessary operations. For this reason, a detailed history should be taken and after careful endoscopic evaluation, CT images should be analyzed carefully and surgery should be considered in cases of longer duration with nutritional support, always emphasizing the importance of individual assessment of each patient.

5. Conclusion

Superior mesenteric artery syndrome (SAMS) or Wilkie syndrome is a rare clinical condition with an incidence of 0.013% to 0.78% but the true incidence is unknown. However, a high number of health professionals still cannot diagnose correctly, as it is an infrequent pathology and sometimes of an insidious nature, in most cases patients go through a long period with a history of abdominal complaints, epigastric pain, vomiting, heartburn can be signs of other gastrointestinal conditions.

The diagnosis of Wilkie syndrome often relies on a high index of suspicion and is often made by a process of exclusion, resulting in ineffective symptomatic symptoms. inadequate therapies and investigations, this delay in diagnosis can lead to a delay in treatment resulting in unnecessary operations. Treatment can be conservative or surgical, however in adult patients non-operative therapy is often a prolonged period of hospitalization with a low success rate ranging between 14 and 71e% (De Rodriguez *et al.,* 2017). Strong's operation, gastrojejunostomy, and duodenujejunostomy are treatment options for Wilkie syndrome. For this reason, a detailed history should be taken and after careful endoscopic evaluation, CT images should be analyzed carefully and surgery should be considered in cases of longer duration with nutritional support, always emphasizing the importance of individual assessment of each patient.

6. References

Alzerwi, N. A. (2020). Predictors of Superior Mesenteric Artery Syndrome: Evidence from a Case-Control Study. *Cureus*, *12*(8).

Araujo, A.O.D., Oliveira, R.G.D., Arraes, A.J.D.C., Mamare, E.M., Rocha, I.D., & Gomes, C.R. (2021). Superior Mesenteric Artery Syndrome–An Uncommon Complication After Surgical Corrections of Spinal Deformities. Brazilian Journal of Orthopedics, 56, 523-527.

Biank, V., & Werlin, S. (2006). Superior mesenteric artery syndrome in children: a 20-year experience. *Journal of pediatric gastroenterology and nutrition*, *42*(5), 522-525.

Biank, V., & Werlin, S. (2006). Superior mesenteric artery syndrome in children: a 20-year experience. *Journal of pediatric gastroenterology and nutrition*, *42*(5), 522-525.

Bronswijk, M., Fransen, L., Vanella, G., Hiele, M., & van der Merwe, S. (2021). Successful treatment of

superior mesenteric artery syndrome by endoscopic ultrasound-guided gastrojejunostomy. *Endoscopy*, *53*(02), 204-205.

Chrysikos, D., Troupis, T., Tsiaoussis, J., Sgantzos, M., Bonatsos, V., Karampelias, V., ... & Mariolis-Sapsakos, T. (2019). Superior mesenteric artery syndrome: a rare case of upper gastrointestinal obstruction. *Journal of Surgical Case Reports*, *2019*(3), rjz054.

Chrysikos, D., Troupis, T., Tsiaoussis, J., Sgantzos, M., Bonatsos, V., Karampelias, V., ... & Mariolis-Sapsakos, T. (2019). Superior mesenteric artery syndrome: a rare case of upper gastrointestinal obstruction. *Journal of Surgical Case Reports*, *2019*(3), rjz054.

de Rodríguez, E. J. O., Hernández-Villegas, A. C., Serralde-Zúñiga, A. E., & del Carmen Reyes-Ramírez, A. L. (2017). The two sides of superior mesenteric artery syndrome treatment: conservative or surgical management?. *Nutrición Hospitalaria*, *34*(4), 997-1000.

Fan, Y., Cai, M., Wang, J., & Xia, L. (2021). Superior Mesenteric Artery Syndrome Following Scoliosis Surgery: A Systematic Review of Case Reports. *Annals of Vascular Surgery*, *76*, 514-535.

Farina, R., Foti, P. V., Coronella, M., Pennisi, I., Libra, F., Di Mari, A., ... & Basile, A. (2021). Superior mesenteric artery syndrome (Wilkie Syndrome) with unusual clinical onset: Description of a rare case. *Radiology Case Reports*, *16*(10), 2998-3002.

Hamidi, H., Rahimi, M. T., Maroof, S., & Soroush, F. A. (2019). Computed tomography features and surgical treatment of superior mesenteric artery syndrome: A case report. *Radiology Case Reports*, *14*(12), 1529-1532.

Hines, J. R., Gore, R. M., & Ballantyne, G. H. (1984). Superior mesenteric artery syndrome: diagnostic criteria and therapeutic approaches. *The American journal of surgery*, *148*(5), 630-632.
in Gastroenterology, *9*(2), 194-199.

Johnson, B. M., & Paladugu, G. (2019). Superior mesenteric artery syndrome secondary to anorexia nervosa and methamphetamine use. Cureus, 11(11).

Khattab, A., Hashmi, S., Ehrenpreis, E.D., Coffey, J.C. (2021). Vascular Anatomy of the Mesentery. In: Ehrenpreis, E.D., Alverdy, J.C., Wexner, S.D. (eds) The Mesenteric Organ in Health and Disease. Springer, Cham. https://doi.org/10.1007/978-3-030-71963-0_4.

Lippl, F., Hannig, C., Weiß, W., Allescher, H. D., Classen, M., & Kurjak, M. (2002). Superior mesenteric artery syndrome: diagnosis and treatment from the gastroenterologist's view. *Journal of gastroenterology*, *37*(8), 640-643.

Mandarry, M. T., Zhao, L., Zhang, C., & Wei, Z. Q. (2010). A comprehensive review of superior mesenteric artery syndrome. *European Surgery*, *42*(5), 229-236.

Matsevych, O., Koto, M., Balabyeki, M., & Aldous, C. (2018). Trauma laparoscopy: when to start and when to convert?. *Surgical Endoscopy*, *32*(3), 1344-1352.

Mendes, K. D. S., Silveira, R. C. D. C. P., & Galvão, C. M. (2008). Revisão integrativa: método de pesquisa para a incorporação de evidências na saúde e na enfermagem. *Texto & contexto-enfermagem*, *17*, 758-764.

Merrett, N. D., Wilson, R. B., Cosman, P., & Biankin, A. V. (2009). Superior mesenteric artery

syndrome: diagnosis and treatment strategies. *Journal of Gastrointestinal Surgery*, *13*(2), 287-292.

Pillay, Y. (2016). Superior mesenteric artery syndrome: a case report of two surgical options, duodenal derotation and duodenojejunostomy. *Case Reports in Vascular Medicine*, *2016*.

Recio-Barbero, M., Fuertes-Soriano, S., Cabezas-Garduño, J., López-Atanes, M., Peña-Rotella, A., & Sáenz-Herrero, M. (2019). Delayed diagnosis of an eating disorder in a male patient with superior mesenteric artery syndrome: results from a case study. *Frontiers in Psychiatry*, *10*, 731.

Silva, A. L., Antunes, D., e Cunha, J. C., Nogueira, R., Fernandes, D., Salazar, T., & Pinto, C. M. (2020). Epigastric Pain and Weight Loss–A Case of Wilkie's Syndrome. *European Journal of Case Reports in Internal Medicine*, *7*(5).

Sinagra, E., Raimondo, D., Albano, D., Guarnotta, V., Blasco, M., Testai, S., ... & Midiri, M. (2018). Superior mesenteric artery syndrome: clinical, endoscopic, and radiological findings. *Gastroenterology research and practice*, *2018*.

Sinagra, E., Raimondo, D., Albano, D., Guarnotta, V., Blasco, M., Testai, S., ... & Midiri, M. (2018). Superior mesenteric artery syndrome: clinical, endoscopic, and radiological findings. *Gastroenterology research and practice*,

Wang, Y., & Takada, T. (1984). Superior mesenteric artery syndrome: report of four cases. *Gastroenterologia Japonica*, *19*(5), 479-485.

Warncke, E. S., Gursahaney, D. L., Mascolo, M., & Dee, E. (2019). Superior mesenteric artery syndrome: a radiographic review. *Abdominal Radiology*, *44*(9), 3188-3194.

Welch, B., Schaal, A., O'Shea, T. F., & Cantu, R. (2021). Superior Mesenteric Artery Syndrome: A Community Hospital Case Series. *Case Reports in Vascular Medicine*, *2021*.

Welsch, T., Büchler, M. W., & Kienle, P. (2007). Recalling superior mesenteric artery syndrome. *Digestive surgery*, *24*(3), 149-156.

Welsch, T., Büchler, M. W., & Kienle, P. (2007). Recalling superior mesenteric artery syndrome. *Digestive surgery*, *24*(3), 149-156.

Yılmaz, T. U., Güler, S. A., Pösteki, G., & Alponat, 2017. Superior Mesenteric Artery Syndrome-Process of Diagnosis and Treatment of Problematic Cases. Okmeydanı Tıp Dergisi, n. 33, p. 57-59.

Zaraket, V., & Deeb, L. (2015). Wilkie's syndrome or superior mesenteric artery syndrome: fact or fantasy. *Case reports*

Zee, K., Stephens, M., & Fabiszak, M. (2021). Superior Mesenteric Artery Syndrome: Could it Involve a Potential Familial Pattern?. *Cureus*, *13*(5).

Zhang, R., Li, Z. T., Han, X. W., Liang, L. D., Wang, Z. G., & Ji, F. (2021). Diagnosis and treatment of superior mesenteric artery compression syndrome complicated with gastroesophageal reflux disease. *Chinese medical journal*, *134*(11), 1382-1384.