Abstract:
SMA syndrome is an unusual cause of proximal intestinal obstruction. It is a clinical entity characterized by compression of the third, or transverse, portion of the duodenum between the aorta and the superior mesenteric artery. This results in chronic, intermittent, or acute complete or partial duodenal obstruction. We present a case of a 40 years old lady who presented with postprandial abdominal pain at the epigastric region, colic type, without radiation accompanied by nausea, postprandial vomiting and weight loss. She was evaluated and diagnosed as SMA syndrome with duodenojejunal diverticulosis. She was taken up for surgery due to chronic malnutrition not improving with conservative management. Duodenojejunostomy with transgastric feeding jejunostomy was done. To alleviate postoperative refractive gastroparesis, kinking and angulation, transgastric feeding jejunostomy was done in contrast to modified witzels technique.

The patient was free of symptoms and recurrence during follow up.

Keyword: Cast syndrome, chronic duodenal ileus, Arterio-mesentric duodenal compression syndrome, Wilkie syndrome, Duoduenojejunal diverticulosis.

BACKGROUND: Superior mesenteric artery syndrome was first described in 1861 by Von Rokitansky,[1] who proposed that its cause was obstruction of the third part of the duodenum as a result of arterio-mesentric compression. It was later popularised by Wilkie [2]. The syndrome is characterized by compression of the third portion of the duodenum due to narrowing of the space between the superior mesenteric artery and aorta and is primarily attributed to loss of the intervening mesenteric fat pad. It has been referred to by a variety of other names including Cast syndrome, Wilkie syndrome, arterio-mesentric duodenal obstruction, and chronic duodenal ileus [3].
Despite the fact that about 400 cases are described in the English language literature, existence of duodenojejunal diverticulosis makes it as second case to be reported[4]. Furthermore, the diagnosis may be confused with other anatomic or motility-related causes of duodenal obstruction [5].

CASE REPORT:
We present 40 year old thin built housewife weighing 29kg presented to our department with complaints of postprandial vomiting for 1 year. Vomiting is of bilious in nature, 30 min to 60 minute after food intake, with increased frequency for past 45 days. She also had colicky post prandial abdominal pain, recurrent abdominal distension, ball rolling movements, weight loss despite good appetite. She had no history of altered bowel habits, hematemesis, malena, jaundice. She had no co-morbidities. Her laboratory parameters of complete hemogram, liver function, renal function, viral markers were unremarkable. UGI endoscopy revealed D1 &D2 distended, narrowing at D3, lumen appears normal with duodenal diverticulosis. Contrast enhanced computed tomography (CECT) revealed reduced aorto-mesentric distance of 7 mm (Fig 1). Aorto-mesentric angle also reduced to 10 degrees with compression of D3 stomach. Left renal vein is also compressed between aorta and SMA features suggestive of SMA Syndrome. Thus based on the history, the clinical and imaging findings, the diagnosis of SMA syndrome was concluded. Due to chronic malnutrition with increased frequency of symptoms she underwent duodenojejunosotomy bypass procedure. The first and second part of duodenum were grossly dilated and hypertrophied. The dilated duodenum abruptly ended at the third part of duodenum where the superior mesenteric artery crossed the duodenum. Multiple duodenojejunal diverticula were found with largest measuring 3 cm (Fig 3 ). SMA was thick and compressing D3.

These findings confirmed the diagnosis of SMA syndrome and side to side duodenojejunostomy in two layers was performed. (Fig 4) To alleviate postoperative refractive gastroparesis transgastric feeding jejunostomy was done. (Fig 5) The postoperative course was uneventful. She had complete relief of symptoms without any recurrence in the follow up period. She gained weight of 12 kg on follow up at 6th month and remained asymptomatic.

DISCUSSION
Superior mesenteric artery (SMA) syndrome is a rare but well recognized clinical entity characterized by compression of the third portion of the duodenum between the aorta and the superior mesenteric artery. First described by Von Rokitansky in 1861 [1]. The reported prevalence in the general population varies between 0.013% and 0.78% [6]. The superior mesenteric artery usually forms an angle of approximately 45° (range, 38-56°) with the abdominal aorta.[7]. The third part of the duodenum crosses caudal to the origin of the superior mesenteric artery, coursing between the superior mesenteric artery and aorta. Any factor that sharply narrows the aortomesenteric angle to approximately 6-25° can cause entrapment and compression of the third part of the duodenum resulting in SMA syndrome.[8] In the majority of patients, the normal angle between the superior mesenteric artery and the aorta is between 38 and 65° due, in part, to the mesenteric fat pad [9]. This angle correlates with body mass index [10]. The aortomesenteric distance is normally 10 to 28 mm [11].
Wilkie’s syndrome tends to affect young female adults aged between 17-39 years old. Probable predisposing factors include immobilization in the supine position or application of body casts, considerable and rapid weight loss such as in intensive diets or in anorexia nervosa, malrotation or paraduodenal hernias and abdominal aorta aneurysm [12-14]. Alternatively other causes implicated include high insertion of duodenum at the Ligament of Treitz, a low origin of SMA and compression of duodenum due to peritoneal adhesions [15]. The main symptoms of Wilkie’s syndrome are epigastric discomfort and pain followed by vomiting which as the disease progresses they become more severe, frequent and causing fluid and electrolyte disturbances with weight loss [16]. An acute presentation is uncommon [17]. The diagnosis of the disease is confirmed by radiological imaging [18]. The most helpful roentgenologic sign is a line of obstruction in the third part of the duodenum passing obliquely towards the right lower quadrant corresponding to the course of the superior mesenteric vessels. Thus, the findings of SMA syndrome in upper GI study are very important. It reveals the characteristic dilatation of the first and second parts of the duodenum, with an abrupt vertical or linear cutoff in the third part with normal mucosal folds. Very little barium is seen to pass into jejunum during the early part of the examination. The barium meal will show obstruction of the third part of the duodenum with proximal dilation of the organ and a clear cut line which demarks the obliteration of the duodenal lumen by external compression of the SMA. These radiographic features usually disappear when placing the patient in the knee – chest position as well as after application of pressure at the lower abdomen (Hayes maneuver) [19]. Computed tomographic (CT) and magnetic resonance (MR) arteriography have largely replaced conventional arteriography since they are noninvasive and provide additional anatomic detail such as the amount of intra-abdominal and retroperitoneal fat excluding other causes of duodenal obstruction (tumors, annular pancreas etc..) [20, 21]. Conservative management is the rule for acute cases [22]. It consists of frequent meals of fluid or blenderized food, diluted to the consistency of soup and then the patient lying on a the left side prone or in the knee - chest position. Total parenteric nutrition has also been used but eventually 50-70% of all cases will come to surgery. Surgery is indicated for chronic cases and failure of conservative management. The operative options include duodenojjejunostomy, section of the ligament of Treitz and relocation of the duodenojejunal junction (strong’s procedure), gastrojjejunostomy, duodenal anterior replacement [23]. Duodenojjejunostomy is the procedure of choice and is effective in 90% of patients. Strong’s procedure mobilizes the duodenum by dividing the ligament of Treitz. Once the duodenal-jejunal junction has been fully mobilized, the duodenum is positioned to the right of the superior mesenteric artery so it does not lie within the space between the aorta and the superior mesenteric artery [24]. The integrity of the bowel is maintained; however, the short branches of the inferior pancreaticoduodenal artery may limit the ability of the duodenum to fall away from the aorta. Successful laparoscopic duodenojjejunal bypass has also been described. Although experience is limited, it offers a less invasive surgical option [25-27].
Gastroparesis after correction surgery is a frequently encountered problem related to gastric and duodenal atony. Although the presence of such persistent symptoms has been described in the literature, there is little information on their management. In our patient transgastric feeding gastrostomy was done in contrast to modified witzel's technique. This particular attention has avoided kinking and angulation in addition to the post operative gastroparesis. Further powered studies are needed to address this has technique in alleviating the post operative refractive gastroparesis.[28].

REFERENCES:


![Fig.2- Reduced aortomesentric angle](image-url)
Fig.1 - Reduced aortomesentric distance

Fig.3 Distended and dilated stomach and duodenum with duodenojejunal diverticulosis
Fig.4 Duodenojejunostomy bypass

Fig.5 Transgastric feeding jejunostomy