Adolescent with Superior Mesenteric Artery Syndrome

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ABSTRACT

The underlying etiologies of paediatric bowel obstruction are wide ranging. It can be divided into proximal and distal bowel obstruction. Amongst the different etiologies of the proximal bowel obstructions at the level of the duodenum, there are a few etiologies including duodenal atresia, internal hernias, intestinal malrotation, annular pancreas etc. Superior mesenteric artery syndrome is amongst one of these differential diagnoses which is more prevalent in the adolescent age group. We describe the imaging features of this entity and its demographics, imaging characteristics, treatment and prognosis.

CASE REPORT

CASE REPORT

An 18-year-old male adolescent who had a known history of autism spectrum disorder with developmental delay, presented with 2 weeks history of bilious vomiting associated with increasing abdominal pain. The vomiting did not resolve with symptomatic medications given by the general practitioner. In between the episodes of vomiting, the patient however seemed well with good appetite. He also had no bowel output for the past 3 days. Otherwise, he did not have any infective symptoms, no fever or significant travel history.

On examination the abdomen was slightly distended on the epigastric and left upper quadrant area with hypoactive bowel sounds. The clinician noticed that the patient tossed and turned on palpation of the abdomen. The hernia sites and vital signs on admission were otherwise unremarkable.

Initial blood investigations revealed that the patient had normal total white count of 10.5 x 10^9/L (normal range: 4.5-11.0 x 10^9/L). The hemoglobin level was 9.3 g/DL (13.5 – 17.5 g/DL), low for age. The albumin was also slightly low at 33 g/L (normal range: 38 – 50 g/L). Otherwise, the rest of the liver enzyme panel and urine work up were unremarkable. The serum corrected calcium was 2.39 mmol/L (normal range: 2.10 – 2.55 mmol/L). The serum magnesium was low at 0.70 mmol/L (normal range: 0.86 – 1.17 mmol/L).

Subsequently abdominal radiographs performed showed dilated gastric bubble and duodenal loops. Normal gaseous distension of the large bowel was noted on the supine radiograph (Fig. 1).

This suggested that the bowel obstruction was in the proximal small bowel loops, probably the duodenum itself. The patient then went on to have a computerised tomography (CT) scan. The CT scan showed dilated proximal duodenum with a transition point at the 3rd part of duodenum, which was sandwiched between the superior mesenteric artery and abdominal aorta (Fig. 2 and Fig. 3).

An ultrasound study of the abdominal aorta and origin of the superior mesenteric artery was performed. An acute angle and narrowed distance were seen between the proximal segment of the superior mesenteric artery and the abdominal aorta (Fig 4). These radiographic findings were highly suspicious for superior mesenteric artery syndrome.

The patient subsequently underwent introduction of a peripherally inserted central catheter for total parenteral feeding with subsequent progression to nasojejunal tube feeding. He was treated conservatively and responded well. Ultrasound evaluation at one year follow up showed an increase in the angle between the superior mesenteric artery and aorta (Fig 5), with a concomitant weight gain of around 7 kilograms during the period.

DISCUSSION

Etiology & Demographics:

Superior mesenteric artery (SMA) syndrome, also known as Wilkie's syndrome, is a vascular compression syndrome, caused by compression of 3rd part of the duodenum by superior mesenteric artery. Other commonly described vascular compression syndromes of the abdomen are as follows [1]:

- Dunbar syndrome is compression of the coeliac trunk by the median arcuate ligament.
- Nutcracker syndrome is compression of the left renal vein by the aorta and the superior mesenteric artery.
- May-Thurner syndrome is compression of the left common iliac vein by the right common iliac artery.

The incidence of SMA syndrome is less than 0.3% of the population [2-3]. Majority of the patients are teenagers to young adults at the ages of 10 to 39, although outliers like geriatric patients have been reported. Slight higher prevalence in females might occur due to anorexia nervosa. The underlying cause for superior mesenteric artery syndrome is decreased thickness of the retroperitoneal visceral fat pad. Underlying etiologies can be divided into congenital or acquired causes. Congenital causes will include patients with anatomical variants including congenital low position of the superior mesenteric artery, naturally occurring acute SMA-aorta angle and short SMA-aorta distance and intestinal malrotation [1-3]. Acquired causes may include anorexia nervosa, severe burns, malabsorption, malignancy and recent surgeries including upper abdominal gastrointestinal surgery. Other causes also include patients who undergo scoliosis surgery of which increased tautness of the mesentery post-surgery may cause SMA syndrome. Patients who have severe neurological abnormalities, like cerebral palsy, may persistently hyperextend their spine, hence being prone to have narrowed distance between the SMA and aorta [2-3]. In this case, this patient had underlying autistic spectrum disorder with developmental delay, with a low body-mass index, probably predisposed to insufficient nutritional intake.

Clinical & Imaging findings:

Patients with SMA syndrome present with non-specific symptoms. These can be divided into the chronic type which is the majority and the acute presentation [4]. Both include post-prandial obstructive symptoms like nausea, vomiting, abdominal pain and lack of bowel output. The predominant symptom is abdominal pain in the epigastric location [4]. The patient usually experiences symptom relief when adopting a posture that relieves the obstruction, usually in the left lateral stance.

If the condition is severe, which can happen in the more acute type, it may even be complicated by bowel ischaemia or compartment syndrome, hence causing electrolyte derangements and even death [5].

A combination of imaging modalities has been used to workup for SMA syndrome, which includes radiographs, upper gastrointestinal barium imaging and computerised tomography mesenteric angiography. A double bubble sign may be seen in abdominal radiographs in which there is distension of the stomach and the proximal duodenum [2-3]. The classical fluoroscopic findings will be a dilated proximal duodenum with a transition point at the third part of the duodenum, presence of antiperistaltic waves just proximal to the obstruction, delayed gastric emptying and decrease in degree of obstruction with the patient in the left lateral or decubitus position [3].

Cross-sectional imaging will be useful to exclude other differentials of superior mesenteric artery syndrome [2-3]. The late angiographic phase is ideal to simultaneously evaluate the aorto-SMA relationship and the duodenum for tumours. The usual aortomesenteric angle in normal patient is between 25 to 60 degrees with an aortomesenteric distance of 10 to 28mm [1-3]. Most cases of superior mesenteric syndrome have an aortomesenteric angle of less than 22 degrees, with an aortomesenteric distance of less than 10 mm. Measurements of magnetic resonance imaging and ultrasound also show the same results [2,3,6].

Some of the patients undergo endoscopy as they might have been referred to a gastroenterologist in view of the abdominal symptoms. Case reports do describe features of duodenal dilatation with large amounts of gastric contents as suggestive of SMA syndrome on endoscopy [7]. This can be further augmented with endoscopic ultrasound to confirm the pulse-like nature of the obstruction with narrowed aortomesenteric distance. Endoscopic evaluation is also useful to exclude other pathologies, for example duodenal tumours or peptic ulcer disease that may also cause duodenal obstruction [3].

Treatment & Prognosis:

Treatment of SMA syndrome involves restoration of the retroperitoneal fat to decrease obstructive symptoms.

Conservative methods are first employed. The patient is supported with fluids to correct electrolyte imbalances [2-3]. The obstruction is bypassed with nasojejunal feeding and nursing in the left lateral or prone position. Parenteral nutrition may be employed to increase retroperitoneal fat. Sometimes medications were also prescribed including prokinetics and antacids [8]. The mean duration of medical treatment has been reported to be around 45 days. Results were worse with medical therapy beyond 6 weeks, hence surgical intervention can be considered [8].

Surgical methods including laparoscopic duodenojejunostomy may be considered if conservative methods are unsuccessful [9]. It has been reported to have the highest success rate. The other alternative surgeries will include the Strong procedure, which involves ligating the ligament of Treitz, and gastroduodenostomy. Strong procedure does not disrupt the natural bowel anatomy but has a failure rate quoted around 25%. Gastroduodenostomy may fail to release the duodenal obstruction, hence requiring a duodenojejunostomy. The recurrence rate overall was lower in patients with surgical treatment [9].

Differential Diagnosis:

Considering that the patient is a young adolescent, neonatal diagnoses for proximal bowel obstruction like duodenal atresia are unlikely. We would like to entertain a few differentials which are possible in this age group including internal hernia, intestinal malrotation, annular pancreas, bezoars and Crohn's disease.

Internal hernia

Internal hernia is an important differential with regards to proximal bowel obstruction. It is a rare entity, being reported in less than 1% of autopsies [10]. The paraduodenal hernias are the most common type of internal hernia, followed by pericaecal, foramen of Winslow, transmesenteric etc. Some reports do mention that transmesenteric hernias do occur more commonly in older children [11]. In our practice we encountered a rare case of hernia which is transomental hernia (Fig. 6), occurring in approximately 1 to 4% of internal hernias.

CT has been established as the best modality for workup of internal hernias due to the limitations of other modalities and the quick identification of the location and etiology of the hernia [10]. Traditional fluoroscopic studies will require a longer amount of time and administration of oral contrast media is not allowed for high grade obstruction.

In figure 6, we can see that the abdominal CT shows abnormal distribution of the small bowel loops within the lesser sac. The key point in cases of internal hernia is to look for evidence of closed loop obstruction. This can be seen by two distinct transition points in close spatial proximity to each other, resembling a "beak sign" [12]. The herniated bowel loops may be fluid-filled/distended. Next, the hernia orifice needs to be localised. This can be aided by looking for the confluence of vessels and bowel loops towards a point. The mesentery of the herniated loop also usually shows a "misty" appearance signifying venous congestion [12]. The functionality of radiographs and ultrasound are limited for characterisation of the internal hernia [11].

Intestinal Malrotation

Intestinal malrotation is a congenital abnormality where there is abnormal positioning of the bowel loops due to aberrant rotation during embryogenesis [13]. As the gut involutes back into the abdominal cavity, it should undergo a 270 degrees anticlockwise rotation where the duodenojejunal junction is on the left of the left L1 pedicle. Failure to do that will result in a short mesentery which may predispose the patients to complications like midgut volvulus.

Conventional radiographs may show proximal bowel distension with absence of distal gas [13]. Upper gastrointestinal series remain the main modality of diagnosis. Kimberly et al. recommends documentation of the duodenojejunal junction on the left of the L1 vertebral body on the antero-posterior projections. On the lateral projection the DJ junction should be posterior in location, in keeping with a retroperitoneal structure. Failure of visualisation of the above will result in radiological diagnosis of intestinal malrotation. Delayed images can be performed to locate the caecum which will not be in the right iliac fossa in 80% of patients with malrotation. A corkscrew duodenum will be seen in a patient with malrotation complicated by midgut volvulus, which requires emergency surgical intervention [13]. A false positive rate of up to 15% has been reported due to variations in the duodenojejunal junction anatomy including wandering duodenum or displacement of the junction due to enlarged stomach. Cross-sectional imaging and ultrasound might be useful in showing a reversal of the normal SMA-superior mesenteric vein (SMV) relationship with the SMA being on the right of the SMV in cases of malrotation [13] which can be seen in 60% of patients.

Annular pancreas

Annular pancreas is a congenital extrinsic type of duodenal obstruction [14]. It arises when the dorsal bud fails to completely rotate fully, causing it to encircle the duodenum. It may be an infiltrative process occurring within the duodenal fibres which predisposes to duodenal ulceration; alternatively, it may remain external to the duodenum, but making the patient prone to duodenal obstruction.

Radiographs will reveal the double bubble sign [15]. Fluoroscopic study will show the transition point at the 2nd part of the duodenum [15,16]. Ultrasound will show an echogenic ring of pancreatic tissue around the duodenum with a duct extending into the circumferential component of the pancreas [16]. Cross-sectional imaging shows focal narrowing of the 2nd part of the duodenum with the enlarged pancreas wrapped around it [14,17]. With magnetic resonance imaging (MRI), the duct encircling the duodenum can be visualised, further confirming pancreatic annulus. Duodenal obstruction is seen in 33 to 40% of annular pancreas [17]

Other extrinsic lesions also include duodenal haematomas which may occur in the setting of trauma or anticoagulation.

Duodenal masses (intraluminal) – bezoar

Bezoars are masses containing indigestible fibre [18]. The type of bezoar is classified according to their composition, ranging from plant fibre to ingested hair. Rapunzel syndrome is used to describe the presence of a trichobezoar extending into the small or even large bowel causing gastric outlet obstruction [19].

Radiographs may show a dilated gastric antrum and proximal duodenum. An erect chest radiograph will be useful to look for visceral perforation [18]. Fluoroscopic studies may show an intraluminal filling defect without a connection to the bowel wall [19]. Ultrasound might help to locate an intraluminal mass within the duodenum. It will show a hyperechoic arc border with hypoechoic shadowing, due to the multiple foci of gas [18]. Computerised tomography will reveal a mass with mottled appearance. To differentiate from small bowel faeces, bezoars will show a floating fat density sign, better appreciated by shifting the window level to around -50HU [20].

Crohn's disease

Crohn's disease is a bowel inflammatory disease. It shows skip lesions with transmural involvement of the bowel. Duodenal involvement in Crohn's disease ranges from 0.5 to 4% [21].

Clinical presentation will depend on the extent of disease involvement which may have complications ranging from bowel obstruction to fistula formation [22].

On radiographs it might not be possible to find any significant findings. Fluoroscopic studies will reveal the classical findings of aphthous ulcerations, fissures, cobble-stoning, pseudopolyps and skip lesion involvement [22]. Sonography will show increased bowel thickness in the affected bowel loop, named as the most important sonographic sign of Crohn's disease. Ultrasound will also be useful in detecting complications such as abscesses [22]. An increase in vascularity with doppler interrogation suggests active disease while decreased vascularity as compared to the surrounding bowel suggests fibrosis. Casciani E. et al. describes ultrasound with oral contrast, which was useful in dynamic evaluation of the bowel loop, with differentiation of the active and fibrotic phases [22]. In computerised tomography, there will be duodenal thickening with hyperenhancement and adjacent fat stranding. In bowel with submucosal oedema, enhancement will be targetoid in nature. Fibrotic bowel will show hypoenhancement. The mesentery also displays the comb sign, which is inflammation of the mesenteric vessels [22]. Magnetic resonance imaging is the preferred modality due to the lack of ionising radiation and superior soft tissue contrast, ideal especially for paediatric patients requiring multiple follow ups. The different sequences also provide additional information of disease activity. Signs of active disease include thickened wall, increased T2W mural signal, and presence of mural differentiation on post contrast images. The comb sign can also be appreciated, similar to the findings on CT. Complications can occur with the formation of fistula or peri-colic abscesses which also can be picked up on CT and MRI cross-sectional imaging [22].

TEACHING POINT

The underlying etiologies of paediatric duodenal obstruction are wide ranging and many of the entities are rare. Of these, superior mesenteric artery syndrome is one which can be demonstrated primarily with an obstruction at the third part of the duodenum and narrowed aortomesenteric angle and distance. Clinicians and radiologists should be mindful of this diagnosis as part of the differential diagnosis of paediatric duodenal obstruction in the appropriate clinical context.

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FIGURES

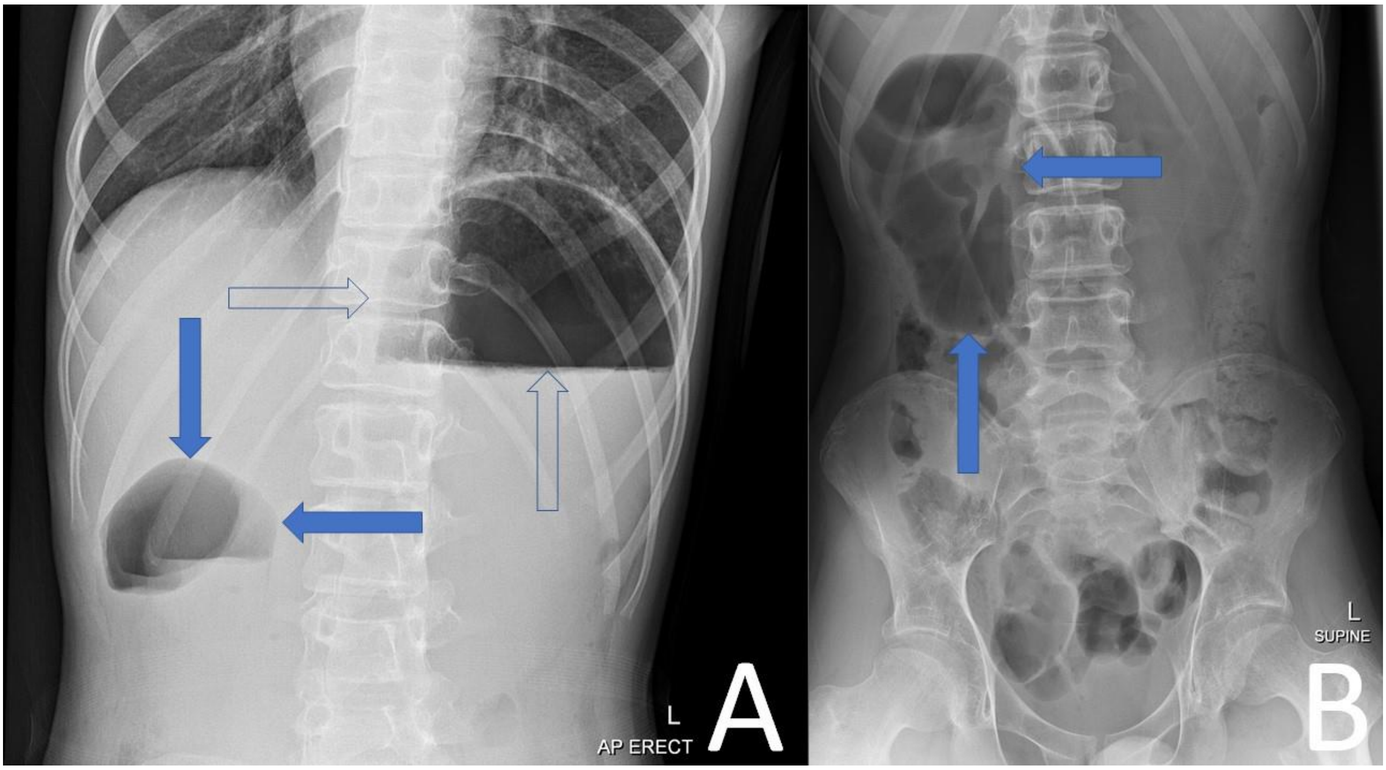


Figure 1: 18-year-old male adolescent with superior mesenteric artery syndrome.

Findings: Erect (a) and supine (b) abdominal radiographs. Note the dilated gastric bubble (blank arrows) and the proximal duodenal loops (solid arrows). Normal gaseous distension of the large bowel is noted on the supine radiograph.

Technique: 90 kVp x 20 mAs.

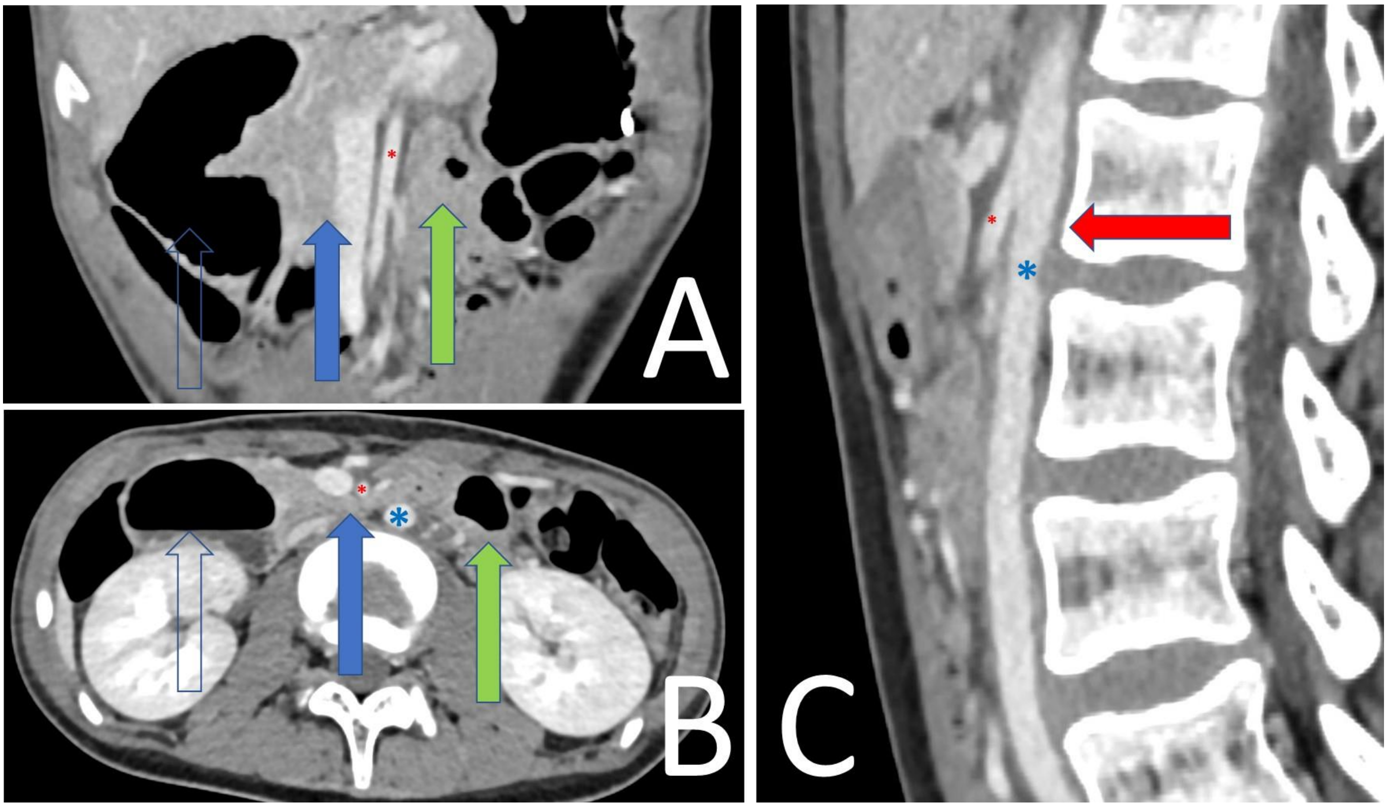


Figure 2: 18-year-old male adolescent with superior mesenteric artery syndrome.

Findings: Coronal (a), axial (b) and sagittal (c) contrast enhanced CT scan of the abdomen and pelvis. The proximal duodenum was dilated (blank blue arrow). The third part of the duodenum was sandwiched between the superior mesenteric artery and the aorta (blue solid arrow). The post-stenotic segment of the duodenum shows normal calibre (green arrow). The blue asterisk is the aorta and the red asterisk is the superior mesenteric artery. Note the acute angle between the proximal segment of the superior mesenteric artery and the abdominal aorta, in keeping with superior mesenteric artery syndrome (red solid arrow).

Technique: CT scan of the abdomen and pelvis with multiplanar reconstruction. 80 mls of Omnipaque 300 was injected intraveously. 90 kv x 144 mAs. 5mm slick thickness.

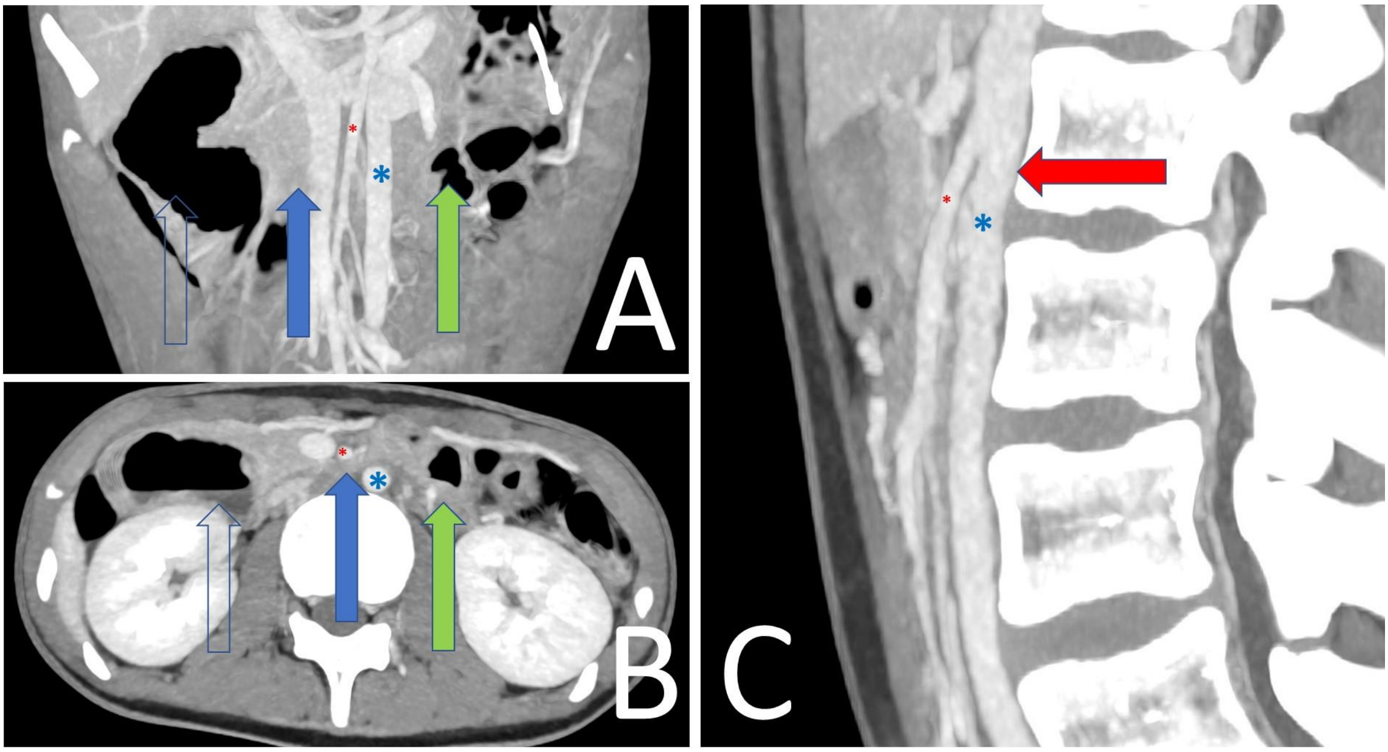


Figure 3: 18-year-old male adolescent with superior mesenteric artery syndrome.

Findings: Coronal (a), axial (b) and sagittal (c) contrast enhanced CT scan of the abdomen and pelvis. The proximal duodenum was dilated (blank blue arrow). The third part of the duodenum was sandwiched between the superior mesenteric artery and the aorta (blue solid arrow). The post-stenotic segment of the duodenum shows normal calibre (green arrow). The blue asterisk is the aorta and the red asterisk is the superior mesenteric artery. Note the acute angle between the proximal segment of the superior mesenteric artery and the abdominal aorta, in keeping with superior mesenteric artery syndrome (red solid arrow).

Technique: CT scan of the abdomen and pelvic, maximal intensity projection. 80 mls of Omnipaque 300 was injected intraveously. 90 kv x 144 mAs. 10mm slick thickness.

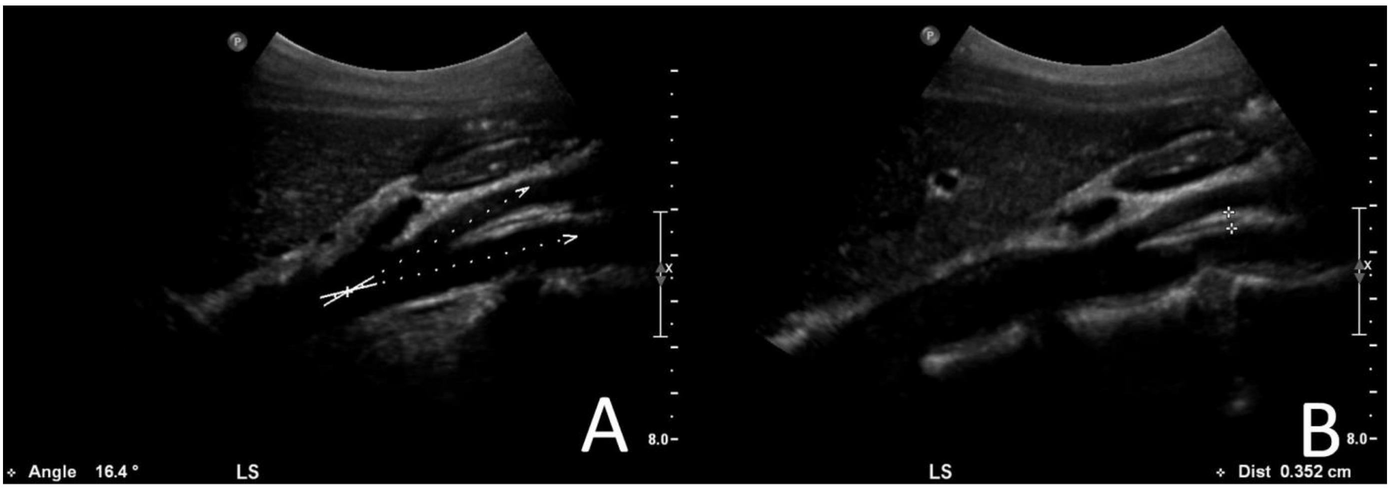


Figure 4: 18-year-old male adolescent with superior mesenteric artery syndrome.

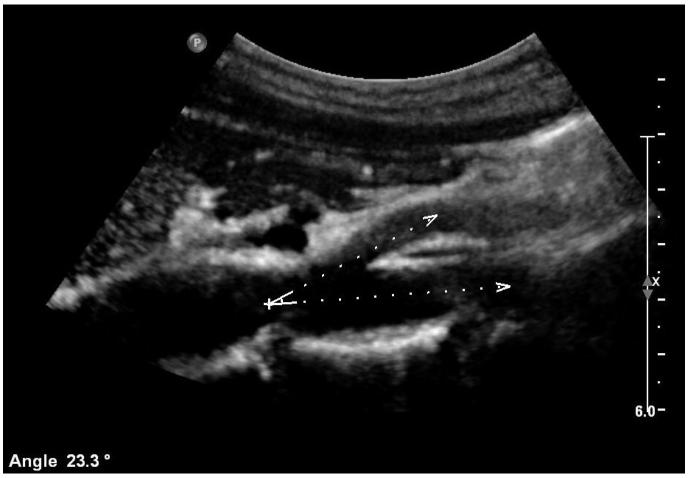
Findings: Longitudinal ultrasound images of the abdominal aorta and the origin of the superior mesenteric artery. Note the acute angle (a) and narrow distance (b) between the proximal segment of the superior mesenteric artery and the abdominal aorta.

Technique: Real time ultrasound of the aorta using curved array transducer at 40Hz.

Figure 5 (right): One year interval follow up ultrasound of a 19-year-old male adolescent with superior mesenteric artery syndrome.

Findings: Longitudinal ultrasound images of the abdominal aorta and the origin of the superior mesenteric artery. Note that the angle between the proximal segment of the superior mesenteric artery and the abdominal aorta has increased as compared to the prior study.

Technique: Real time ultrasound of the aorta using curved array transducer at 40Hz.



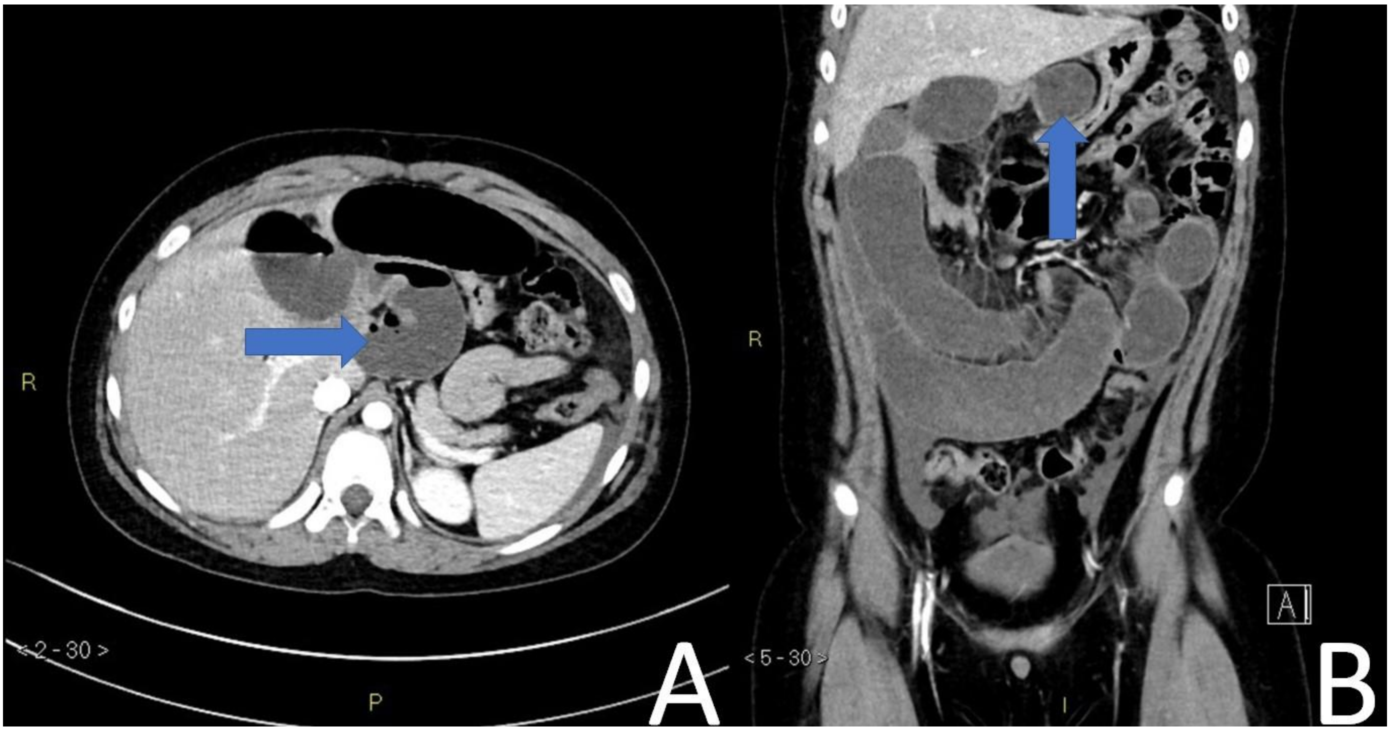


Figure 6: 9 years old male child with transomental internal hernia into the lesser sac.

Findings: Annotated axial (a) and coronal (b) contrast enhanced CT scan of the abdomen and pelvis showing a closed loop small bowel obstruction within the lesser sac (solid blue arrow), suggestive of an internal hernia. The proximal small bowel loops were dilated. This was subsequently surgically proven to be a transomental hernia.

Technique: CT scan of the abdomen and pelvis with multiplanar reconstruction. 70 mls of Omnipaque 300 was administered intravenously. 90 kv x 178 mAs. 3mm slice thickness.

	Plain radiograph	Fluoroscopy	Ultrasound	CT/MRI
Superior mesenteric artery syndrome	Dilated bowel loops until the third part of the duodenum. "Double bubble" sign.	Dilated proximal duodenum with a transition point at the third part of the duodenum, presence of antiperistaltic waves just proximal to the obstruction, delayed gastroduodenal emptying and decrease in degree of obstruction with the patient in the left lateral or decubitus position	Aortomesenteric angle < 22º, aortomesenteric distance < 10 mm.	Aortomesenteric angle < 22º, aortomesenteric distance < 10 mm.
Internal hernia	Dilated bowel loops proximal to obstruction.	Abnormal cluster of bowel loops. Transition point at the afferent limb of the hernia.	Dilated bowel loops.	Clustering of bowel loops at the hernia. "Beak" sign suggesting close loop obstruction.
Malrotation	Dilated bowel loops proximal till duodenum. "Double bubble" sign.	Failure of the duodenojejunal junction to be on the left and inferior to the L1 vertebral body. "Corkscrew sign" suggests midgut volvulus.	Reversal of the normal SMA-SMA relationship.	Failure of the duodenojejunal junction to be on the left and inferior to the L1 vertebral body. Reversal of the normal SMA-SMA relationship.
Annular pancreas	Dilated bowel loops till duodenum. "Double bubble" sign.	Transition point at the 2nd part of the duodenum.	Echogenic ring of pancreatic tissue around the duodenum	Focal narrowing of the 2nd part of the duodenum with the enlarged pancreas wrapped around it. The duct may be visualised on MRI.
Duodenal bezoar	Dilated stomach and proximal duodenum.	Intraluminal filling defect without a connection to the bowel wall.	Hyperechoic arc border with hypoechoic shadowing.	Mass with mottled appearance. Floating fat density sign.
Crohn's disease	Dilated duodenum.	Aphthous ulcerations, fissures, cobble-stoning, pseudopolyps and skip lesion involvement.	Increased bowel thickness. Increase in vascularity suggests active disease while decreased vascularity as compared to the surrounding bowel suggests fibrotic stage.	Bowel thickening with hyperenhancement and adjacent fat stranding. In bowel with submucosal oedema, enhancement will be targetoid in nature. Fibrotic bowel will show hypoenhancement. Signs of active disease on MRI include thickened wall, increased T2W mural signal, and presence of mural differentiation on post contrast images.

Table 1: Differential diagnosis table for superior mesenteric artery syndrome

Etiology	Congenital or acquired (see risk factors)	
Incidence	Less than 0.3% of the population	
Gender ratio	Slight prevalence in females	
Age predilection	Usually presents in the young child to young adult age group at the ages of 10 to 39. Some geriatric patients have also been reported.	
Risk factors	Congenital	• Congenital variations of the SMA such as low insertion of the SMA • Congenital neurological abnormalities
	Acquired	• Anorexia nervosa • Malabsorption • Malignancy • Upper abdominal gastrointestinal and scoliosis surgery
Treatment	Conservative	• Obstruction is bypassed with nasojejunal feeding and nursing in the left lateral or prone position. • Parenteral nutrition may be employed to increase retroperitoneal fat
	Surgical	• Laparoscopic duodenojejunostomy • Strong procedure (ligating the ligament of Treitz) • Gastroduodenostomy
Prognosis	Good if responsive to medical treatment or have little surgical complications. Bad prognosis if the patient has an acute presentation with severe complications.	
Imaging findings	Plain radiograph	Dilated bowel loops until the third part of the duodenum.
	Fluoroscopy	Dilated proximal duodenum with a transition point at the third part of the duodenum, presence of antiperistaltic waves just proximal to the obstruction, delayed gastric emptying and decrease in degree of obstruction with the patient in the left lateral or decubitus position
	Ultrasound	Aortomesenteric angle < 22º, aortomesenteric distance < 10 mm
	Computerised Tomography	Aortomesenteric angle < 22º, aortomesenteric distance < 10 mm
	Magnetic resonance imaging	Aortomesenteric angle < 22º, aortomesenteric distance < 10 mm

Table 2: Summary table for superior mesenteric artery syndrome

ABBREVIATIONS

DJ junction = Duodenojejunal junction
MRI = Magnetic resonance imaging
SMA = Superior mesenteric artery
SMA syndrome = Superior mesenteric artery syndrome
SMV = Superior mesenteric vein

KEYWORDS

Superior mesenteric artery syndrome; Wilkie's syndrome; abdomen vascular compression syndrome; computerised tomography; ultrasound; fluoroscopy