

## **Wilkie syndrome: Case Report**

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### **Abstract**

Wilkie syndrome or superior mesenteric artery syndrome (SMAS) or cast syndrome is a rare condition caused by intermittent obstruction of the third portion of the duodenum compressed between the Superior Mesenteric Artery (SAM) anteriorly and the aorta and lumbar spine posteriorly.

It is a pathology that is often seen in children and young adults, and in cases of severe undernutrition. there are no specific clinical or biological criteria. CT scan, coupled with the ingestion of a radiopaque product or a gastroduodenal transit generally allows the diagnosis.

The management of this condition varies between observation, with correction of hydroelectrolytic disorders and the early initiation of nutrition, and surgery, depending on each particular case.

We aim to analyze the means of diagnosis and management of Wilkie syndrome in a pediatric population.

**Keywords:** Wilkie syndrome; Duodenum; Superior mesenteric artery; Computed Tomography

### **Introduction**

Wilkie syndrome is an uncommon condition resulting from an extrinsic compression of the third portion of the duodenum compressed between the SAM anteriorly and the aorta and lumbar spine posteriorly. First described by von Rokitansky in 1842, SMAS was associated with orthopedic casting by Willet in 1878. Wilkie in 1927 formally characterized SMAS in a series of 75 patients [1,2].

SMAS usually presents more acutely than chronically with symptoms of small bowel obstruction and contrast-enhanced spiral computed tomography scan, coupled with the ingestion of a radiopaque product or a gastroduodenal transit generally allows the diagnosis. the evolution is fatal in absence of adequate management.

### **Case Report**

A 12-year-old girl, with history of chronic intermittent abdominal pain, vomiting and gastrointestinal problems apart from a weight-loss of approximately 6 kg over the last 4 months. It was only on the evening was admitted to emergency department for acute abdomen and vomiting. An abdominal CT scan showed a large gastric distension with the stenosis at the level of part III of the duodenum, and a reduced aorto mesenteric angle.

First, she was managed by using a nasogastric tube and with correction of hydroelectrolytic disorders, then a laparoscopic

approach was performed to restore digestive continuity.

### **Discussion**

SMAS is a rare form of upper gastrointestinal obstruction with a reported incidence of 0.013% to 0.3% [3].

Clinical findings include chronic intermittent abdominal pain, vomiting, nausea, early satiety and anorexia. The diagnosis of SMAS is commonly confirmed by upper gastrointestinal radiography but the diagnosis may be made by computerized tomography or laparotomy [4].

Endoscopic ultrasound at the site of duodenal compression has proven to be of substantial value, but angiography or angiography MRI are the best technique for determination of the aortomesenteric angle.

CT scan calculate the angle between the SMA and the aorta which is reduced from 7 ° to 22 °, whereas it is normally between 25 ° and 60 °. The aorto-mesenteric distance is also reduced and measures between 2 -8 mm, while the normal distance is 10 to 28mm [5]. In our patient the angle between the AMS and the aorta calculated on the CT images was 13°.

Surgical treatment is indicated in case of failure of medical treatment. It consists of a bypass by gastrojejunostomy or duodeno-jejunostomy, which can be performed laparoscopically [6] or modifying the anatomical conditions by mobilizing and

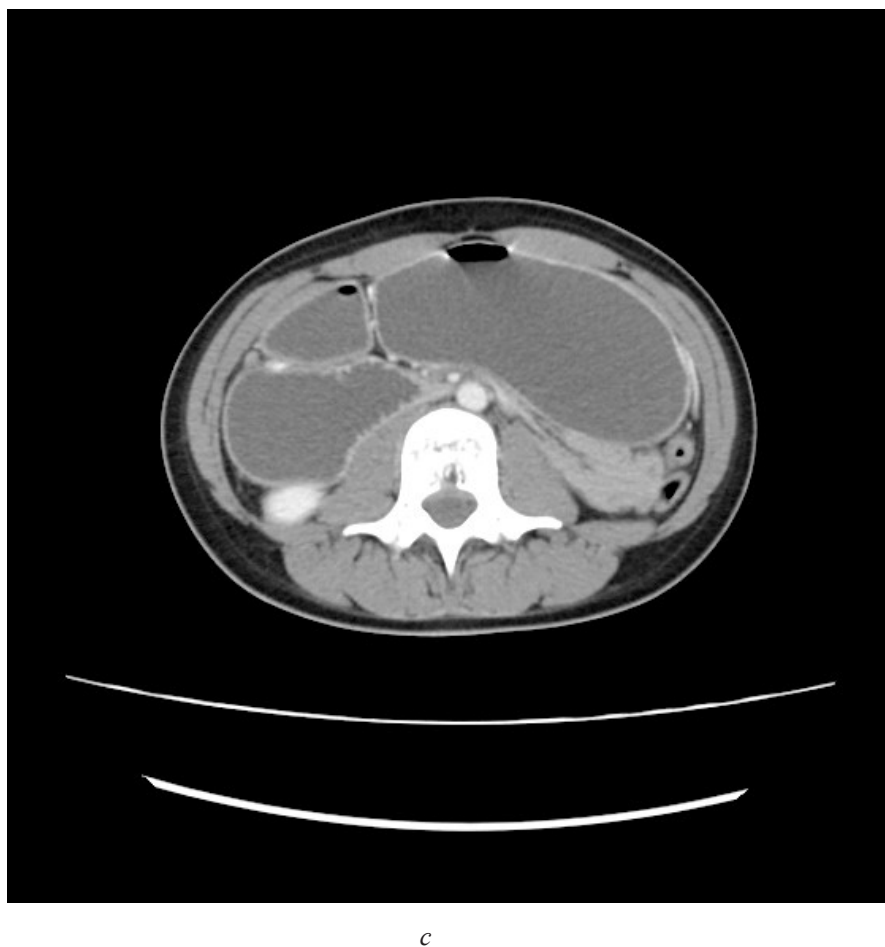
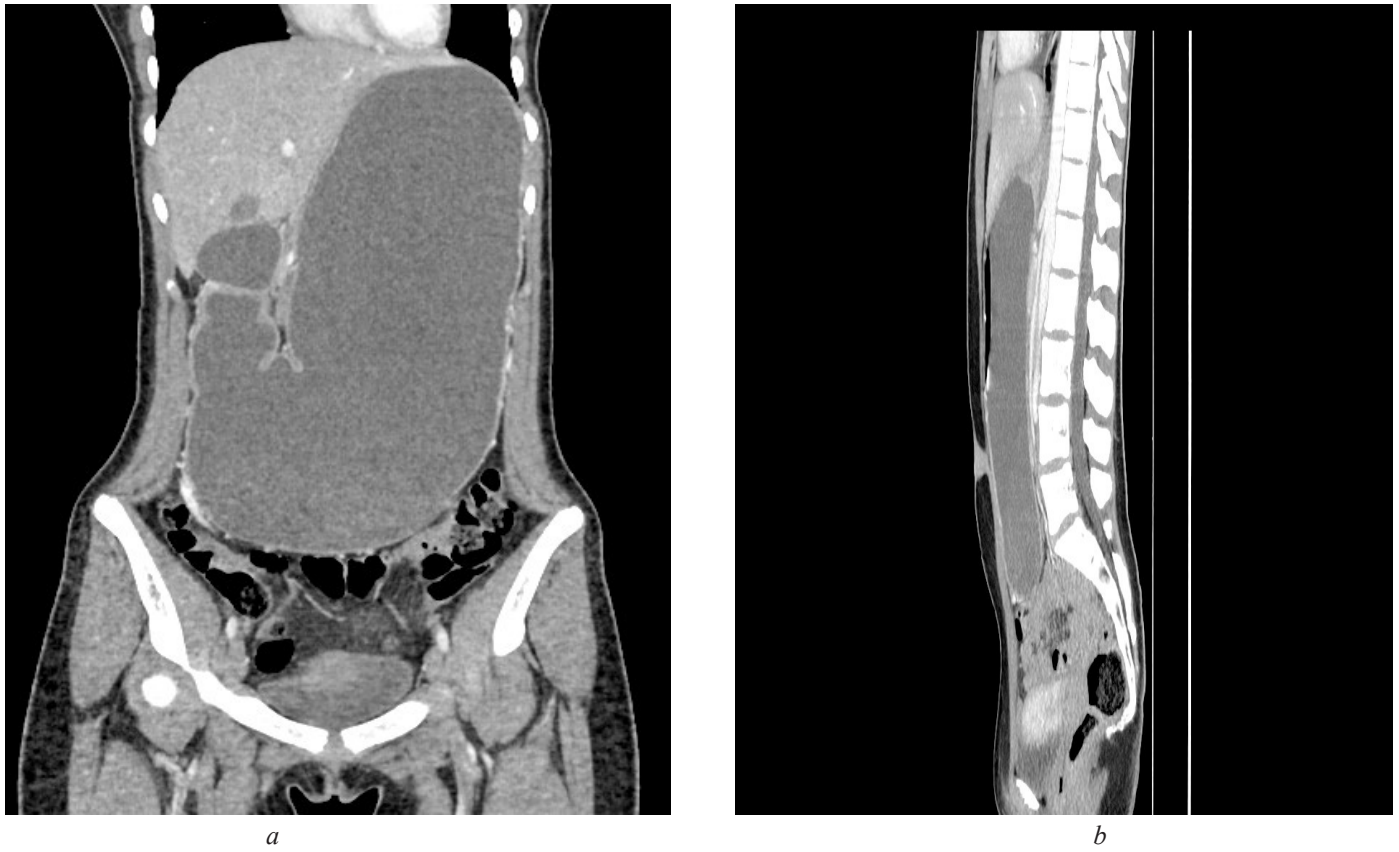


Figure: CT images in coronal (a) section showing a massive gastric distension, sagittal (b) and axial (c) sections, showing SMA (red arrow) exiting at an angle less than  $13^\circ$  from the aorta.

uncrossing the duodeno-jejunal angle by positioning the jejunum to the right of the SMA after section of the Treitz muscle according to Strong's method [7].

### Conclusion

Wilkie syndrome is an uncommon condition which should be suspected in case of small bowel obstruction symptoms and scan. Conditioning of the patient must begin before confirmation of the diagnosis by contrast-enhanced spiral computed tomography CT scan, angiography or angio-MRI.

### Conflicts of Interest

The authors report no conflicts of interest.

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