



By: Baharak Moshiree MD MSc, Atrium Health, Carolinas HealthCare System, Digestive Health-Morehead Medical Plaza; Memi Rooker, IFFGD

International Foundation for Gastrointestinal Disorders (www.iffgd.org)

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What is Superior Mesenteric Artery Syndrome?

Superior mesenteric artery (SMA) syndrome or SMAS is a rare disease with an estimated incidence of about 0.1% to 0.3%. While anyone can get this disease, it is more often found in young adults and adolescents typically aged 10 to 39 years old. Females are more likely to be diagnosed over males; although, this could be due to the lower BMI also seen in these patients. SMAS is caused by the compression of the third part of the duodenum. The duodenum is the first section of the small intestine that connects the stomach to the middle of the small intestine or jejunum. The third part of the duodenum runs between the superior mesenteric artery (SMA) and the abdominal aorta. SMAS can go by many names, including chronic duodenal ileus, Wilkie syndrome, arteriomesenteric duodenal compression syndrome and cast syndrome.

Did You Know?

The first reported case of SMAS was in the 1800s by a man named Carl Freiherr Von Rokitsky. It remained undefined until 1927 when Sir David Percival Dalbreck Wilkie, a surgeon in Scotland, provided more details and findings about the disease in a case series of 75 patients.

What Causes Superior Mesenteric Artery Syndrome (SMAS)?

Duodenal compression occurs when the angle between the superior mesenteric artery (SMA) and the aorta (the aortomesenteric angle) is less than 25 degrees. A normal aortomesenteric angle is anywhere between 38-65 degrees. The most common cause of the decrease in this angle is significant weight loss due to any reason whether cancer, a motility disorder, an eating disorder or many other causes. Surrounding the SMA, there is mesenteric fat pad that acts as a cushion between the aorta and the SMA. This fat pad provides protection around the duodenum and keeps it from being compressed between the two arteries. Significant weight loss can lead to the loss of this fat pad. As a result, the aortomesenteric angle will decrease and cause duodenal compression. This type of

weight loss is usually associated with conditions causing significant weight loss such as:

- Dietary Conditions (anorexia nervosa, malabsorptive diseases)
- Motility disorders causing weight loss such as gastroparesis or functional dyspepsia

Learn more about gastroparesis or functional dyspepsia by visiting IFFGD's websites:
<https://aboutgastroparesis.org>
<https://aboutgimotility.org>

- Surgery or trauma
- Cachexia causing conditions such as cancer, congestive heart failure, or other chronic conditions where lack of muscle mass is seen
- Situations of hypermetabolism like hyperthyroidism or infection

Cachexia – an illness brought on by chronic conditions that results in extreme weight and muscle loss which can include the loss of fat.

Hypermetabolism – an increased state of metabolism typically as the result of injury or illness

Other causes include:

- Surgical correction of scoliosis (youth)
- Congenitally short or enlarged suspensory ligament of the duodenum (ligament of Treitz). This connects and supports the duodenum and jejunum.
- Peritoneal adhesions or bands of scar-like tissue that form inside your abdomen following surgery.
- Duodenal malrotation which is when your duodenum is not where it normally should be.
- Abdominal aortic aneurysm or the weakening and bulging of the abdominal part of the aorta.
- Lumbar lordosis (the exaggerated inward curve of the lower back)

Symptoms

The decreased aortomesenteric angle as mentioned before causes an obstruction or blockage of the duodenum due to compression. This in turn causes pain especially experienced when the patient eats leading to a fear of eating. Depending on the severity of the obstruction the pain can be occasional or last for long periods of time. Lying flat on your stomach (prone), on your left side, or knee to chest may help alleviate this pain. This, however, is not the only symptom associated with SMAS. Other symptoms include:

- Nausea and vomiting
- Weight loss
- Pain or discomfort just below the ribcage in the upper abdominal region also known as epigastric pain
- Abdominal distention- a visible swelling of the abdomen
- Early satiety or feeling full after only consuming a few bites of food

Diagnoses

Due to the rareness of this disorder and the symptoms resembling that of other illnesses diagnosing SMAS can be difficult. Initially, given the symptoms, an upper endoscopy may be done to rule out other causes of weight loss. However, tests can be done to differentiate SMAS from other disorders. The most common method of diagnosis is a computed tomography (CT) scan of arteries called CT angiogram. This scan allows for the measurement of the aortomesenteric angle, which helps to confirm SMA syndrome. Other imaging tests such as a barium x-ray, endoscopy, , and magnetic resonance angiography (MRA) can be done. The most cost effective and preferred test is an Ultrasound with dopplers, also known as a doppler ultrasound.

Barium x-ray – using barium as a contrast agent, this type of x-ray will highlight the gastrointestinal tract making it easier to see any abnormalities. This is also known as a small bowel follow through.

Doppler Ultrasound – a noninvasive test that measures blood flow through arteries and veins. This is the best way to diagnose this condition

Magnetic Resonance Angiography (MRA) – an imaging test that shows blood vessels and blood flow

Computed Tomography (CT) Angiography – A test done to highlight blood vessels and tissues using a dye.

Differential Diagnosis

With superior mesenteric artery syndrome being so rare, not many people know about it. This makes it easy to misdiagnose or confuse with another disorder. Some of these disorders include:

- Anorexia nervosa or other eating disorder
- Gastroparesis
- Cancer
- Duodenal ileus
- Peptic ulcer disease

Treatment

Treatment of superior mesenteric artery syndrome will vary from person to person as there are multiple different causes. With weight loss being the most common cause, a typical treatment plan would be to restore the weight lost. This can be done through various dietary and feeding strategies. Including a dietician to help with these strategies may prove to be beneficial. Surgery is also an option to treat SMAS but is typically only considered when other treatment methods have failed.

IFFGD's **Dietitian Listing** is a resource that allows you to search for a dietitian that is in your area or treats a specific condition.

<https://www.iffgd.org/resources/dietitian-listing.html>

About IFFGD

The International Foundation for Gastrointestinal Disorders (IFFGD) is a 501(c)(3) nonprofit education and research organization. We work to promote awareness, scientific advancement, and improved care for people affected by chronic digestive conditions. Our mission is to inform, assist, and support people affected by gastrointestinal disorders. Founded in 1991, we rely on donors to carry out our mission. Visit our website at: www.iffgd.org.

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537 Long Point Road, Unit 101
Mt Pleasant, SC 29464

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