

# Median Arcuate Ligament Syndrome: A Case Report

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## ABSTRACT

**Background:** The median arcuate ligament passes superior to the origin of the celiac artery and is a continuation of the posterior diaphragm that wraps over the aorta. If it lies too low on the aorta, the ligament may cause symptoms of abdominal pain related to compression of the celiac artery.

**Case Report:** An abdominal ultrasound in a 22-year-old woman with longstanding abdominal pain after eating showed elevated celiac artery velocities of  $>300$  cm/s upon inspiration. Computed tomography angiography of the abdomen showed stenosis of the origin of the celiac artery and confirmed the diagnosis of median arcuate ligament syndrome. Laparoscopic release of the median arcuate ligament resulted in relief of the patient's symptoms.

**Conclusion:** The diagnosis of median arcuate ligament syndrome should be considered in patients with postprandial abdominal pain that does not have a clearly established etiology.

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## INTRODUCTION

The median arcuate ligament is a fibrous arch that unites the diaphragmatic crura on either side of the aortic hiatus. The ligament usually passes superior to the origin of the celiac artery near the first lumbar vertebra. In the general population, 10-24% of people may have indentation caused by an abnormally low ligament.<sup>1</sup> Few of these patients have hemodynamically

significant stenosis that would cause symptoms. We present the case of a patient with median arcuate ligament syndrome that caused abdominal pain associated with nausea, emesis, and bloating.

## CASE REPORT

A 22-year-old woman with no significant past medical history presented to the clinic with a 3-year history of intermittent epigastric abdominal pain. The pain was associated with nausea, nonbilious emesis, and bloating. The pain became worse when she ate fatty foods; the nausea worsened with any oral intake and relieved with bowel rest. She had rare diarrhea and denied any radiation of the pain to other locations in her body. The patient was adopted and did not know of any medical problems in her biological family. She denied excessive alcohol, any illicit drug, or any tobacco product use. She had lost 20 pounds over 3 years. Her physical examination revealed epigastric tenderness to palpation but no other abnormalities.

Electrolyte, *H. pylori* titers, liver function tests, amylase, lipase, and complete blood count were all within normal limits. Right upper quadrant ultrasound showed an insignificant liver hemangioma and no evidence of cholelithiasis. Gastric emptying study showed minimal delay in gastric emptying with a half-life of the tracer expelled of 100 minutes (normal 60-90). A hepatobiliary scan showed minimal evidence of biliary dyskinesia with an ejection fraction of 25% (normal  $>35\%$ ) and no reproduction of symptoms upon administration of cholecystikinin. Esophago-gastroduodenoscopy (EGD) showed a small hiatal hernia and erosive gastropathy. Colonoscopy showed granularity of the terminal ileum. Biopsies of both endoscopies did not show any significant abnormalities. A mesenteric ultrasound showed elevated celiac artery velocities of 155 cm/s that augmented with inspiration to 308 cm/s. Higher velocities suggest increased stenosis, and 200 cm/s suggests 70% stenosis (Figures 1 and 2). Superior mesenteric artery velocities matched those of the aorta and indicated no abnormalities.

Based on the elevated celiac artery velocities, we diagnosed the patient with median arcuate ligament syndrome (celiac artery compression syndrome). Computed tomography angiography (CTA) of the

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**Keywords:** Celiac artery, constriction—pathologic, stenosis from compression by median arcuate ligament of diaphragm

The authors have no financial or proprietary interest in the subject matter of this article.



Figure 1. Mesenteric ultrasound of the celiac artery.

abdomen showed a high-grade stenosis involving the origin of the celiac axis without significant atherosclerotic plaque or calcification (Figures 3 and 4). These findings confirmed the diagnosis.

The patient underwent laparoscopic surgery to release the median arcuate ligament impingement on her celiac artery. The patient was placed in the supine position, and 5 laparoscopic ports were placed. The aorta was identified under the diaphragmatic crura and dissection was carried inferiorly on the aorta until the origin of the celiac artery was identified. The artery was completely skeletonized, releasing any external compression on the artery due to the median arcuate ligament. The patient tolerated this procedure well and remained in the hospital for 23-hour observation. She had an uneventful hospital stay and was tolerating a liquid diet at discharge.

At follow-up in our general surgery clinic 2 weeks after surgery, the patient's postprandial abdominal pain had significantly improved. She tolerated a diet without difficulty and had no complaints of nausea, emesis, or bloating.

## DISCUSSION

Median arcuate ligament syndrome (also known as Dunbar syndrome or celiac artery compression syndrome) was first described by Harjola in 1963.<sup>1</sup> A patient who presented with postprandial abdominal pain and an epigastric bruit was found to have his

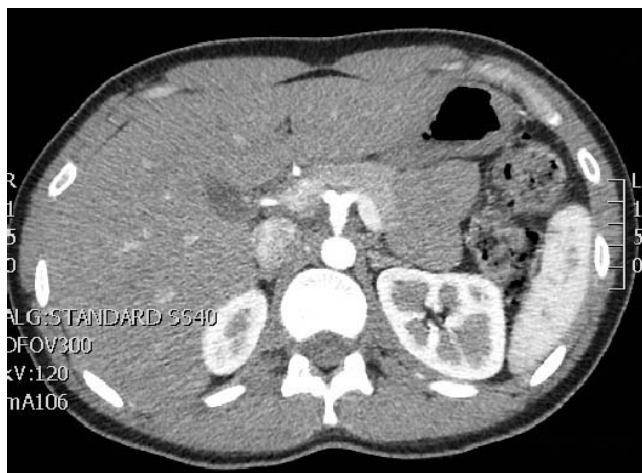


Figure 2. Ultrasound velocities were augmented to  $>300$  cm/s upon inspiration.

celiac artery encased with thick ganglionic tissue at the time of surgery. The patient experienced full relief of symptoms following removal of this thick fibrotic tissue from the celiac artery.

The pathophysiology of the disease is external compression of the celiac artery by an abnormally low lying ligament. The compression worsens with expiration as the diaphragm moves caudally during expiration, causing compression of the celiac trunk. This compression leads to visceral ischemia and postprandial abdominal pain. Some also claim that this causes a steal phenomenon from blood flow being diverted away from the superior mesenteric artery via collaterals to the celiac axis, causing midgut ischemia.<sup>2</sup> Overstimulation of the celiac ganglion is also believed to cause chronic pain in these patients. Sustained compression of the celiac axis may lead to changes in vascular layers such as intimal hyperplasia, proliferation of elastic fibers in the media, and disorganization of the adventitia.

Patients are usually young thin women between the ages of 30 and 50 and typically have had extensive workups for other sources of abdominal pain.<sup>2</sup> Pain is located in the epigastric area and worsens after meals, with exercise, or with leaning forward. The pain is also associated with nausea, emesis, bloating, and diarrhea. Patients may also experience sitophobia, or food fear, because of these symptoms. Patients may get transient relief of these



**Figure 3. Computed tomography angiography showing stenosis of the celiac artery without any evidence of atherosclerosis.**

symptoms by bringing their knees to their chest. This position decreases impingement of the arcuate ligament on the celiac artery by pushing it cephalad relative to the artery as expiration does. Epigastric pain may be present, and physical examination may reveal epigastric bruit in as many as 83% of patients.<sup>2</sup> This bruit may increase on expiration.

In a workup of these patients, other causes of visceral pain should be excluded, including biliary sources and ulcer disease because celiac artery compression syndrome is a diagnosis of exclusion. Therefore, abdominal ultrasound, EGD, and gastric emptying studies are usually performed to rule out other sources of pain. A mesenteric ultrasound is a good screening tool for patients with suspected median arcuate ligament syndrome. The ultrasound should show elevated peak systemic velocities on expiration that may normalize on inspiration or with standing erect. Other observed abnormalities at the origin of the celiac artery would rule out this disease. Reversal of flow in the hepatic artery may also be seen.

Angiography has been the gold standard in the diagnosis of this disease in the past. On lateral views, one may see focal narrowing of the celiac axis with poststenotic dilatation and increased collaterals from the superior mesenteric artery. Angiography has largely been supplanted by multidetector CT scanners with 3-dimensional software, allowing reconstructions at various anatomical planes. A CT scan will be able to detect focal narrowing of the celiac axis, particularly in sagittal views. This narrowing has a characteristic hooked appearance similar to that seen in our patient's CT. Collateral vessels may also be noted. Gastric tonometry has also been used to aid in the diagnosis. Faries et al saw a normalization on



**Figure 4. Sagittal view of the computed tomography angiography showing compression of the celiac artery.**

gastric pH (measured via tonometry catheter) upon release of the median arcuate ligament. In that study, a gastric pH of less than 7.32 indicated significant ischemia.<sup>3</sup> Mensink et al also used tonometry measurements that correlated well with relief of symptoms.<sup>4</sup> In that study, 83% of patients with abnormal gastric pH measurements obtained relief after operative release of the median arcuate ligament. Our patient did not need gastric pH measurements because of the characteristic CT findings.

Surgical median arcuate ligament release has been the mainstay of treatment. The largest follow-up series of open surgical patients was done in 1984 by Reilly et al.<sup>5</sup> A total of 51 patients underwent surgery for median arcuate ligament syndrome: 16 patients underwent decompression only, 17 patients underwent decompression and dilatation, and 18 patients underwent decompression and reconstruction. At 10-year follow-up, 53% of patients who had decompression only had resolution of symptoms compared to 76% of patients with decompression and revascularization.<sup>5</sup> Patient characteristics that predicted relief of symptoms after surgery were postprandial pain, age

40-60 years, female gender, and weight loss greater than 20 pounds. Current results using laparoscopic techniques show relief in nearly 80% of patients undergoing this surgery. Typically, pain relief is immediate, but because postoperative pain can mimic preoperative symptoms and may take up to 6 weeks to resolve, it may take that long to determine if the procedure was successful. Persistent symptoms have been successfully treated with angioplasty.<sup>2</sup>

## CONCLUSION

Median arcuate ligament syndrome is a difficult diagnosis to obtain in a majority of patients. Most patients have had extensive workups or various surgical procedures for postprandial abdominal pain. A patient with suspected compression of the celiac artery should undergo a mesenteric ultrasound with evaluation of artery velocities. Confirmation of this diagnosis can be performed with conventional angiography or CT angiography. Patients who have evidence of median arcuate ligament syndrome

should undergo surgical decompression, which can be accomplished laparoscopically.

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