

# Surgical Treatment of Median Arcuate Ligament Syndrome: Case Report and Review of Literature

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## SUMMARY

**Introduction** Median arcuate ligament (MAL) syndrome, also called celiac trunk compression syndrome (CACS) or Dunbar syndrome is a rare disorder caused by compression of the celiac artery by median arcuate ligament of the diaphragm, which leads to mesenteric ischemia and chronic abdominal angina. The typical clinical triad of symptoms includes postprandial epigastric pain, weight loss and vomiting. The gold standard for MAL syndrome diagnosis is selective angiography, while in symptomatic patients with angiographically verified stenosis the optimal therapy is surgical treatment.

**Case Outline** A 40-year-old male patient was presented with epigastric pain, followed by dyspepsia and weight loss. The upper endoscopy showed gastric and duodenal distention with prominent folds of gastric mucosa and slow peristalsis. Selective angiography showed stenosis (90%) of initial segment of the celiac trunk. Adhesiolysis with the transection of the median arcuate ligament was performed. Due to repeated symptoms, the patient was reoperated on the 10<sup>th</sup> postoperative day with performed adhesiolysis and gastrostomy for gastric nutrition. Two months later, the patient was rehospitalized for closure of gastrostomy. At five years follow-up, selective angiography showed no stenosis of the initial segment of the celiac artery.

**Conclusion** Despite the existing controversy concerning pathophysiological mechanism, the clinical presentation and treatment modalities of patients with MAL syndrome, it is evident that careful selection and adequate surgical treatment may significantly reduce symptoms in these patients.

**Keywords:** celiac artery; median arcuate ligament; diaphragm; arterial occlusive disease

## INTRODUCTION

Median arcuate ligament (MAL) syndrome, also known as celiac artery compression syndrome (CACS) is a rare disorder caused by compression of the celiac artery by the median arcuate ligament, a fibrous arch anterior to the aorta which is formed by a connection of the diaphragmatic crura [1]. Sporadically, the median arcuate ligament may insert at a lower level, thus compressing and narrowing the lumen of the celiac artery, especially during expiration. In that way, subsequent mesenteric ischemia may cause a typical triad of symptoms characteristic for MAL syndrome: postprandial abdominal pain, weight loss, nausea and vomiting [2]. The first celiac trunk compression by the median arcuate ligament was observed during autopsies by Lipshutz et al. [3] in 1917. However, the first clinical cases of celiac artery compression syndrome with angiographically visible stenosis of the celiac trunk were described by Harjola [4] and Dunbar et al. [5] in 1963 and 1965, respectively, who also defined celiac artery compression syndrome (often called Dunbar syndrome).

Treatment modalities for patients with a symptomatic compression of celiac artery and angiographically verified stenosis are percutaneous transluminal angioplasty and surgical

treatment. Here we report a case of a 40-year-old symptomatic patient with angiographically verified compression of celiac artery that was surgically treated and who had no symptoms during the 5-year follow-up period.

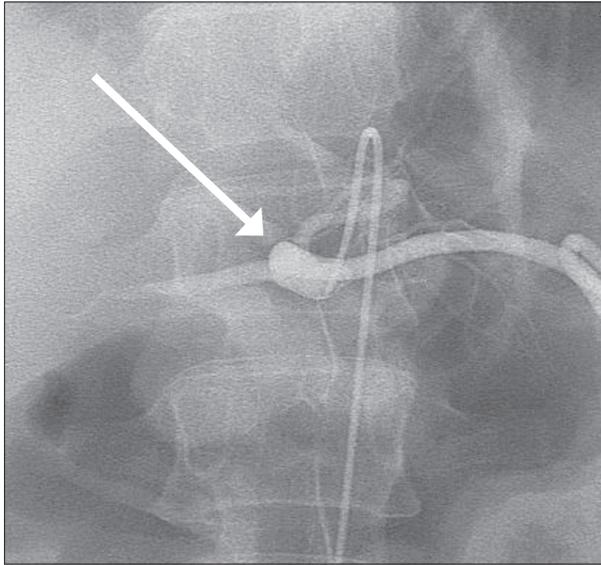
## CASE REPORT

A 40-year-old male patient presented with chronic epigastric pain which spread along both costal margins and not related to food intake, dyspepsia, weight loss and flatulence that have been ongoing on for over a year. The patient was occasionally using analgesics which only temporarily diminished abdominal pain. Physical examination and laboratory blood tests were unremarkable with normal findings on abdominal ultrasonography. Esophagogastroduodenoscopy showed distension of the stomach with prominent folds of gastric mucosa, as well as distended duodenum with slow peristalsis and rough appearance of its mucous layer. Abdominal X-ray verified air fluid levels in the small intestine with gastric distention.

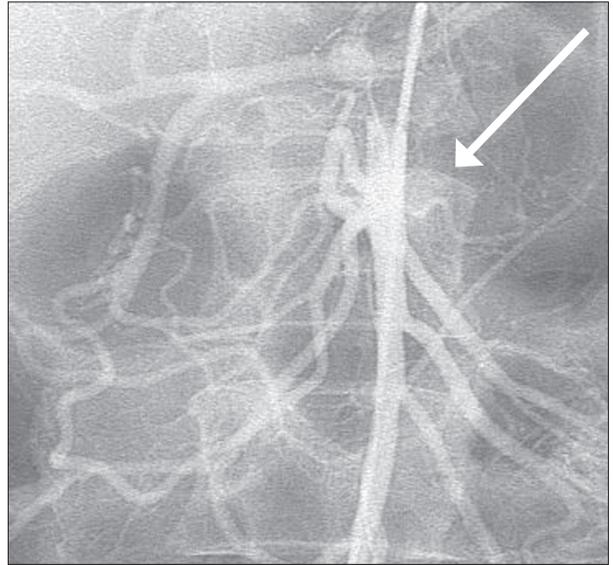
Selective angiography of the celiac trunk and superior mesenteric artery showed stenosis (90%) of initial segment of the celiac artery caused by median arcuate ligament compression. The stenosis was respiratory-dependent:

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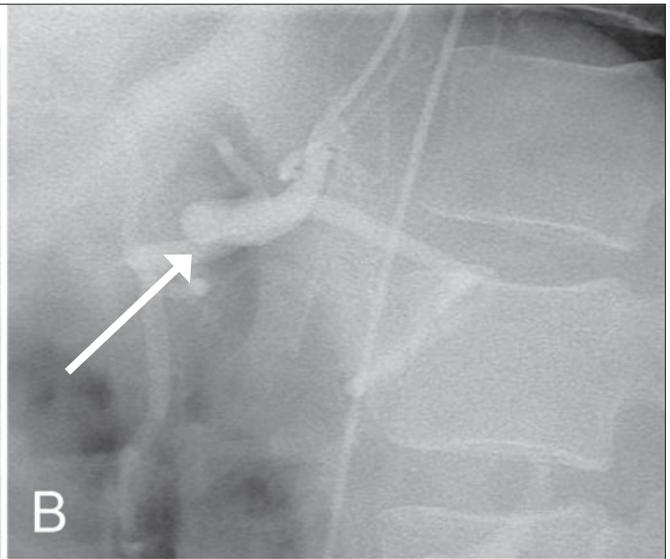
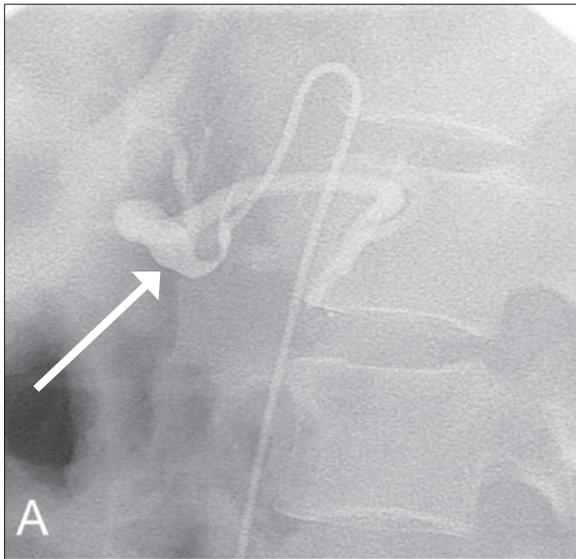
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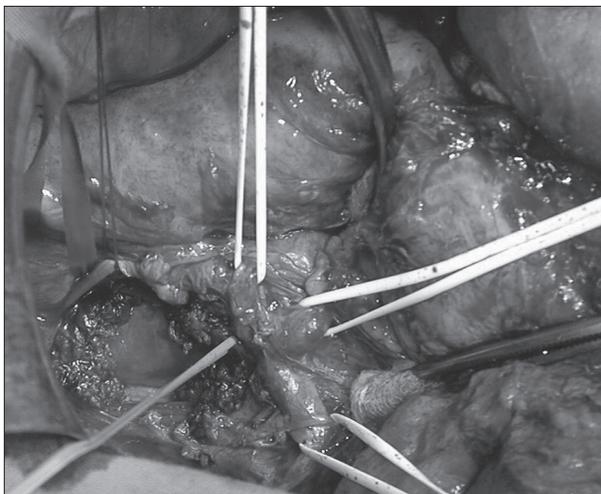
**Figure 1.** Frontal projection of visceral arteries



**Figure 2.** Hepatic "steal"; retrograde visualization of the gastroduodenal artery via the superior mesenteric artery



**Figure 3.** Profile angiography of the celiac artery showing (A) dynamic stenosis of the celiac artery during normal respiratory phase and (B) decompression of the celiac artery during forced inspiration



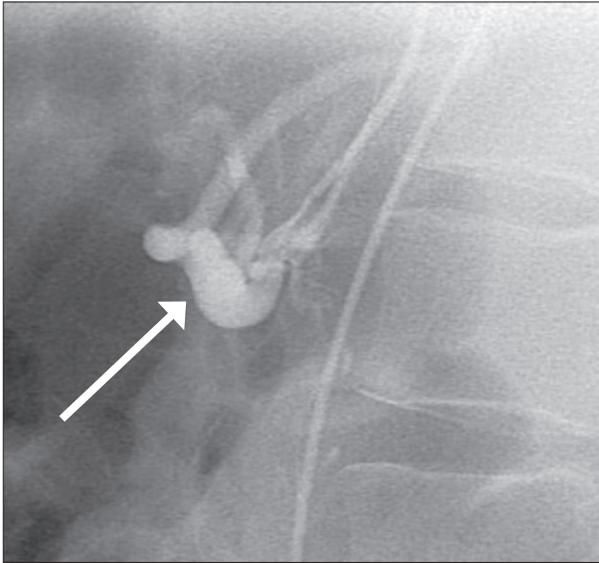
**Figure 4.** Intraoperative transection of the median arcuate ligament

it was present in expiration while it disappeared during the deep inspiration.

Angiography of the visceral arteries in the antero-posterior projection showed the entire celiac trunk, but gastroduodenal artery could not be viewed (Figure 1). The gastroduodenal artery was visualized retrogradely via the superior mesenteric artery (hepatic "steal") due to lower pressure in the celiac trunk system (Figure 2).

Selective angiography showed dynamic stenosis of the celiac artery due to diaphragmatic ligament compression; during expiration extrinsic compression reduced the celiac artery lumen for 80% (Figure 3A). During the forced inspiration, the lumen of the celiac artery was decompressed with reestablished adequate blood flow (Figure 3B).

After adequate preoperative assessment, adhesiolysis with transection of the median arcuate ligament was performed (Figure 4).



**Figure 5.** Selective angiography showing no reduction in the lumen of the initial segment of the celiac artery during expiration

However, early remission was followed by repeated symptoms; abdominal pain, nausea followed by vomiting, and the patient refused any oral nutrition. Therefore, the patient was reoperated on the 10<sup>th</sup> postoperative day, with performed adhesiolysis and gastrostomy for gastric nutrition. Postoperative course was without complications, intrahospital stay was prolonged in order to stabilize the patient's condition and nourishment, and the patient was discharged on the 45<sup>th</sup> postoperative day. Two months after discharge, the patient was rehospitalized for the closure of gastrostomy, and was back on oral nutrition with normal digestive functions.

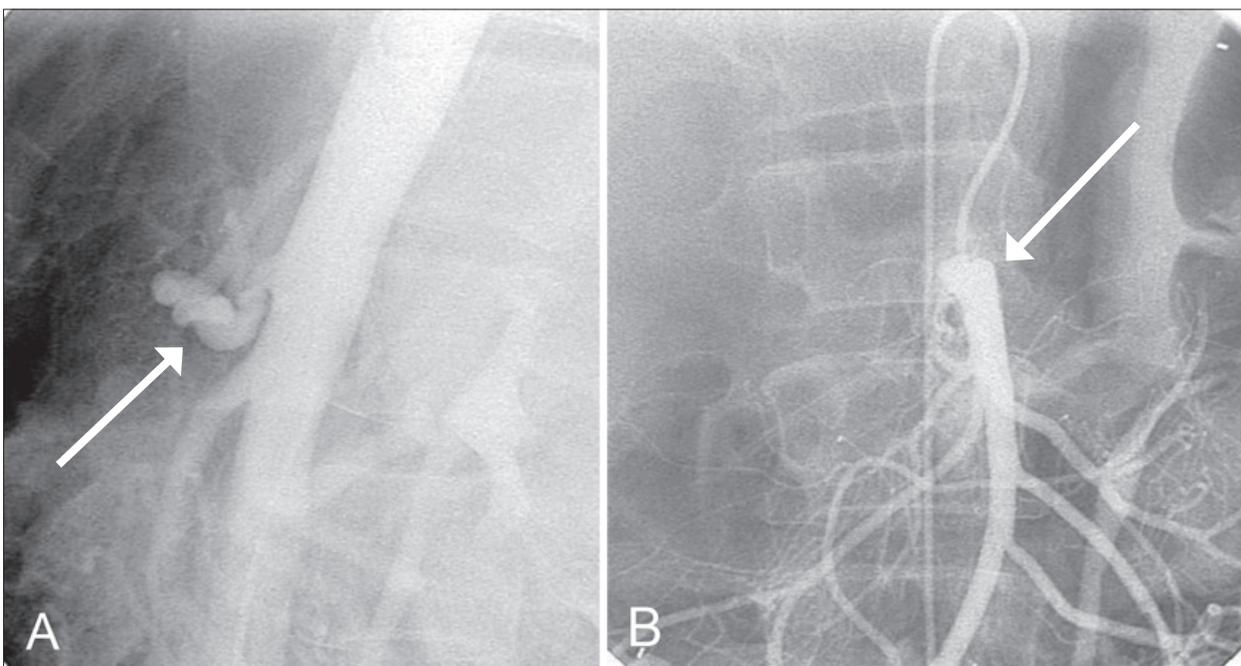
Selective angiography of the celiac trunk was performed five years after the operation, which showed no reduction

in the lumen of the initial segment of the celiac artery during expiration (Figure 5). Also, selective angiography of the upper mesenteric artery failed to show reverse blood flow via the gastroduodenal artery (Figure 6) which corroborated well with the finding of the patent' ostium of the celiac artery and its successful decompression.

## DISCUSSION

MAL syndrome is a rare disorder caused by extrinsic compression and narrowing of the celiac artery by a relatively low insertion of the median arcuate ligament of the diaphragm. The exact pathophysiological mechanism underlying this disorder is still unclear [6, 7, 8]. Namely, although celiac artery shows reduced blood flow, the superior and inferior mesenteric artery are widely patent, which should provide a normal intestinal perfusion. Therefore, there are two proposed theories which tend to elucidate the existing symptoms of MAL syndrome. According to the first theory, compression of the mesenteric artery leads to mesenteric ischemia, either directly or indirectly, via "steal phenomenon" by the collaterals which connect the superior mesenteric and celiac artery [9]. Still, there are rare cases of co-existing stenosis of the celiac artery and superior mesenteric artery was reported by Sianesi et al. [10].

The second neurogenic theory implies the involvement of the celiac ganglion and plexus and its subsequent stimulation, which may lead to splanchnic vasoconstriction [11]. In the majority of patients extrinsic compression of the celiac artery is caused by the median arcuate ligament, but several cases of celiac ganglion fibrosis have been described as a potential cause of extrinsic celiac artery compression [12, 13], which confirms the latter



**Figure 6.** Profile angiography showing (A) the ostia of the celiac artery and superior mesenteric artery and (B) selective angiography of the superior mesenteric artery

pathogenesis theory of this syndrome. Albeit, controversies still exist, since it has been reported that 13-50% of healthy subjects may exhibit a certain degree of angiographically visible compression of the celiac artery during expiration [6].

MAL syndrome is typically presented in young to middle-age adults, although it has been described in infants and children [14]. It is characterized by a clinical triad of postprandial epigastric pain, weight loss and vomiting.

The typical manifestation of abdominal angina is seen only in about 40% of patients, while more than 80% of cases have symptoms which vary from postprandial cramp-like abdominal pain to non-specific epigastric pain [15].

The gold standard of the diagnostic method for MAL syndrome is selective angiography performed in inspiration as well as in expiration. Treatment modalities include endovascular and surgical procedures (open or laparoscopic surgery). Although percutaneous transluminal angioplasty with stent implantation is a less invasive procedure, it often does not solve the problem of the underlying extrinsic compression of the celiac trunk and frequently requires surgical interventions.

Classic treatment for MAL syndrome is represented by open surgery with celiac artery decompression. The largest series of surgically treated patients with MAL syndrome included 51 patients [16]. Surgical modalities included celiac trunk decompression in 16 patients, decompression and dilatation in 17 patients, and decompression and reconstruction either by primary reanastomosis or interposition of the graft in 18 patients. The best results of symptoms relief were achieved in cases of decompression and certain forms of celiac revascularization (76%). Also, Grottemeyer et al. [17] in their series of 18 surgically treated patients observed that open surgical therapy in its

various forms is a safe and reliable procedure with either no mortality or low morbidity rate.

Since the beginning of laparoscopic era, there have been numerous reports of different laparoscopic techniques for treating patients with MAL syndrome [18, 19, 20]. Baccari et al. [21] (2009) have reported a successful laparoscopic decompression of the celiac artery in a group of consecutive patients with complete resolution and symptom-free period of 28.3 months. Despite the postoperative effectiveness of laparoscopic procedures, no long-term follow-up is still available.

Tsujimoto et al. [22] have used intraoperative Doppler ultrasound for the confirmation of the decompressed celiac artery after successful laparoscopic intervention in patients with MAL syndrome. Recently, several authors have reported robotic-assisted treatment of MAL syndrome as a new, safe and efficacious modality treatment [23, 24, 25].

Although MAL syndrome has been described in the 1960s, controversies concerning pathophysiological mechanism, clinical presentation and treatment modalities still exist. Regardless of the controversial viewpoints, the majority of authors agree that symptomatic patients with angiographically confirmed celiac artery compression will benefit more from surgical treatment.

In this paper we presented a case of a 40-year-old symptomatic patient with angiographically verified stenosis of the celiac artery that was surgically treated. The patient had no symptoms during a 5-year follow-up period, which can be considered as a long-term follow-up.

Many questions regarding MAL syndrome have not been clarified yet, but it is evident that careful selection and adequate treatment can significantly reduce symptoms in these patients while the choice of treatment must depend on the specific clinical situation for each patient.

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## Хирушко лечење синдрома медијалног аркуатног лигамента дијафрагме – приказ болесника и преглед литературе

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### КРАТАК САДРЖАЈ

**Увод** Синдром медијалног аркуатног лигамента дијафрагме, такође познат као синдром компресије целијачног трункуса или Данбаров (Dunbar) синдром, редак је поремећај узрокован компресијом целијачне артерије медијалним аркуатним лигаментом дијафрагме који доводи до мезентеричне исхемије и хроничне ангине абдомена. Типично клиничко тројство симптома укључује постпрандијални епигастрични бол, губитак тежине и повраћање. Златни стандард у дијагностици овог синдрома је селективна ангиографија, док је за болеснике који имају симптоме и ангиографски потврђену стенозу оптимални третман хируршко лечење.

**Приказ болесника** Мушкарац стар 40 година пожалио се на бол у епигастријуму праћен диспепсијом и губитком телесне тежине. Ендоскопски преглед горњих органа указала је на надутост желуца и дуоденума с истакнутим наборима гастричне мукозе и спором перисталтиком. Селективна ангиографија указала је на стенозу (90%) иницијалног сегмен-

та целијачног трункуса. Обављена је адхезиолиза уз пресецање медијалног аркуатног лигамента. Због поновљених симптома, болесник је опет оперисан десетог дана након прве операције, када је урађена адхезиолиза и креирана нутритивна гастростомија. Два месеца касније болесник је поново примљен на болничко лечење, како би се извршило затварање нутритивне гастростомије. Пет година након хируршког лечења контролном селективном ангиографијом није утврђена стеноза иницијалног сегмента целијачне артерије.

**Закључак** Упркос опречним ставовима у вези с патолошким механизмима настанка и клиничком сликом Данбаровог синдрома, те облицима лечења болесника с овим обољењем, јасно је да пажљив одабир одговарајућег хируршког приступа може значајно умањити симптоме код ових болесника.

**Кључне речи:** целијачна артерија; медијални аркуатни лигамент; дијафрагма; артеријска оклузивна болест

Примљен • Received: 10/04/2014

Прихваћен • Accepted: 03/06/2014