

# Unusual Cause and Treatment of Uncontrolled Hypertension



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## ABSTRACT

Median arcuate ligament syndrome (MALS) is a rare benign condition typically affecting young females. It usually presents with chronic abdominal pain, nausea, vomiting and weight loss. They are usually diagnosed incidentally on MDCT angiography. We report a rare adult male of MALS which presented with a recurrent hypertensive urgency without any abdominal pain leading to an extensive workup and incidental diagnosis of MALS which was successfully treated by angioplasty with stenting of celiac trunk without any complication.

## Introduction

**D**unbar syndrome (DS), also called median arcuate ligament (MAL) syndrome/celiac artery compression syndrome/Harjola–Marable syndrome, is characterized by external compression of celiac artery root by the MAL [Figure 1 A-B]. Harjola first described Dunbar Syndrome in 1963 [1].

The reported incidence of 2 per 100,000 is between 1.76% and 4%, [2] more commonly seen in young (20–40 years of age) and thin women who present with postprandial epigastric pain (80%), nausea (9.7%), weight loss (48%), and diarrhea (7.5%). CA

compression has been found in 34% of autopsies without prior reporting of symptoms [3].

Traditionally, the treatment of this syndrome is surgery such as a classic opening or a novel laparoscopic division of the median arcuate ligament to relieve the extrinsic compression [4]. Percutaneous endovascular treatment is an alternative technique and may be considered in selected cases, for which the traditional surgery failed or was a treatment option. [5]

## Case Presentation

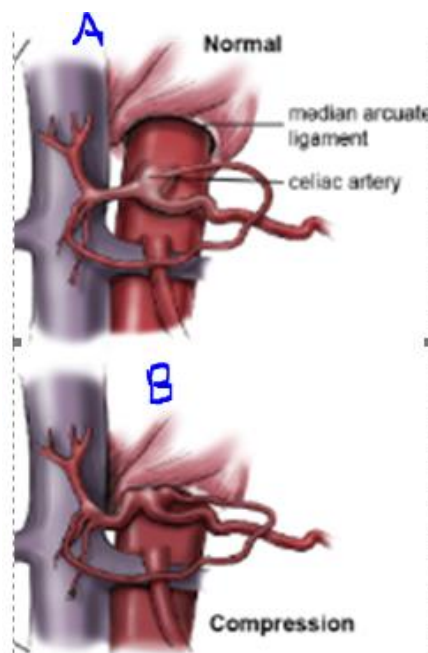
A 17-year-old male patient without history of any

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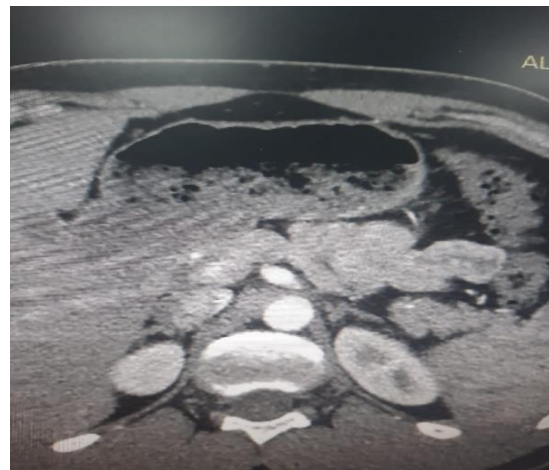
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**Fig. 1.** (A) Normal anatomy of the median arcuate ligament passes anterior to the aorta and superior to the celiac artery. (B) In case of median arcuate ligament crosses proximal portion of the celiac artery resulting in compression. Illustration by Scott Holmes, CMI, Michael E. DeBaakey Department of Surgery



**Fig. 2.** (three Dimensional-sagittal) CT Angiography suggested severe stenosis in ostial celiac trunk ( blue arrow)



**Fig. 3.** Axial CT angiogram of the abdomen. The image at the level of origin of celiac trunk (blue arrow), in which a linear hypodense structure (orange arrow) separates it from the abdominal aorta and indicates the arcuate ligament compression.

chronic disease presents with recurrent attacks of headache, palpitation and uncontrolled blood pressure in the last 8 months. The patient complains from exertional shortness of breath, dizziness upon standing, frequent headaches, and palpitations. He also described fluctuation in her blood pressure with high systolic blood pressures ranging between 170 to 190 mmHg. A documented maximum systolic pressure of 210 mmHg was also reported, other than normal physical examination. Laboratory work-up for hypertension was normal renal and liver function test, normal complete blood count, and no cardiac abnormalities. Electrocardiogram demonstrated

normal sinus rhythm. Renal duplex ultrasonography suggested bilateral renal artery as normal. A CT angiogram of the aorta showed normal bilateral renal arteries, no Coartication of Aorta, no suprarenal mass, but ostial focal narrowing 90% at the origin of celiac trunk which is most compatible with the appearance of compression by the median arcuate ligament was observed. [Figure 2, 3]

Despite extensive investigations and evaluation for the cause of his hypertension, no underlying etiology



**Fig. 4.** (A) Selective celiac trunk angiography suggested severe ostial stenosis, (B) After deployment of a 8 x 25 mm Bare-metal stent in focal stenosis, (C) successful angioplasty with stenting in ostial celiac trunk.

could be identified. Then selective celiac angiography was done by an interventional cardiologist which showed severe ostial celiac artery stenosis with post stenotic dilatation and the patient was planned for celiac artery angioplasty and stenting. Thereafter endovascular treatment involved deployment of an 8 × 25 mm bare-metal stent at the proximal celiac artery [Figure 4 A-C], with successful final angiographic result and without residual stenosis. The patient was discharged on the second day with improved symptoms and no complications.

## Discussion

The Median Arcuate Ligament Syndrome or Celiac Artery Compression Syndrome is a rare cause of postprandial pain and weight loss. The etiology is celiac artery compression by the medial arcuate ligament (MAL) resulting in compromised blood flow and symptom causation [1,4,6,7]. However, some patients are asymptomatic due to sufficient collateral supply from superior mesenteric circulation. This syndrome is more frequent among young females with a thin body habitus and comprises the classical triad: postprandial abdominal pain, epigastric bruit, and presence of extrinsic celiac compression revealed by vascular imaging [8]. Other clinical symptoms are associated with vegetative symptoms including nausea, vomiting, dizziness, tachycardia, diarrhea, weight loss and sweating [3,9].

In this case, the patient didn't experience any abdominal pain, but he presented with unusual symptoms like exertional shortness of breath, dizziness upon standing, frequent headaches, palpitations and he had uncontrolled hypertension with unknown cause despite many investigations conducted and MDCT. Abdominal angiography incidentally discovered Median Arcuate Ligament Syndrome. Three-dimensional volume rendering plays a significant role in establishing the diagnosis.

MAL thickness >4 mm is considered abnormal. [10] Dunbar Syndrome is diagnosed with MDCT abdominal angiography, which demonstrates a characteristic focal narrowing of the proximal celiac axis with a typical "hooked" configuration caused by the inferior displacement of the celiac artery by the MAL on sagittal views [6]. Sugae *et al.* [11] proposed a grading system of celiac artery stenosis caused by MAL compression based on stenosis grade, length of a stenosis, distance from the abdominal aorta, and presence of small collateral vessels found on CT angiography.

The goal of MALS treatment is restoring normal blood flow in the celiac axis [2]. Classically, a simple surgical division of the fibrous ligament was performed. Other complex surgical procedures like vascular reconstruction of the celiac artery with patch angioplasty, aortoceliac bypass, and reimplantation of the celiac artery may be needed in some patients. However, open surgery is more invasive and increases the morbidity rate [2,12]. The recent application of laparoscopy in the division of MAL has proven to be a novel technique because it is less invasive and involved a lower morbidity rate than open surgery [13]; yet, the results from both procedures are equal. However, the outcomes from several of these studies are based on only short follow-up periods [13].

Percutaneous angioplasty with stenting is an alternative technique for the treatment of MALS. It is a minimally invasive technique, characterized by short hospitalization and low morbidity rate. However, there are a few studies that have reported successful endovascular treatment. One of them is the study by Silva *et al.* [14], in which stenting was employed in four patients with extrinsic compression of the celiac artery with immediate excellent results. Only one of the four patients had a 3-year symptom-free follow-up period. However, long term prognosis of the angioplasty and stenting is still unknown because a few treated cases were reported. In this case, endovascular treatment

was done with beneficial effect and symptoms were relieved.

## Conclusion

MALS is a rare condition and it may present with atypical presentations like palpitation, exertional shortness of breath, dizziness upon standing, frequent headaches and uncontrolled hypertension. Percutaneous angioplasty with stenting is an alternative technique for the treatment of selected cases of MALS with successful results.

## Ethical Considerations

### Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

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### Conflict of Interests

The authors have no conflict of interest to declare.

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