

# Celiac Artery Compression by the Median Ligament: An Uncommon Cause of Abdominal Pain in the Emergency Department. Report of a Case

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**Abstract:** The Authors report a case of a patient presenting in the Emergency Department (ED) with severe epigastric pain refractory to therapeutic treatment, with onset after forced ice water ingestion caused by joking compression of a soft PET<sup>®</sup> bottle during drinking.

The Angio MR and angiography performed, after exclusion of suspected esophagus rupture and other esophago-gastric diseases, to rule out an ischemic origin of the pain demonstrate a stenosis of the celiac artery resulting from median arcuate ligament narrowing, the so-called Dunbar's Syndrome.

The Dunbar's Syndrome is uncommon and some aspects such as the vascular etiology of symptoms are still controversial.

In this report clinical presentation, differential diagnosis and pathophysiology of this disease are discussed.

**Keywords:** Celiac artery stenosis, Dunbar's syndrome, Median arcuate ligament syndrome, Abdominal pain, Emergency Department.

## INTRODUCTION

Celiac artery compression also known as median arcuate ligament or Dunbar's syndrome is an uncommon disorder characterized, in symptomatic patients, by transient post-prandial or exercise-related or chronic epigastric pain, hyperemesis, vomiting, diarrhoea and weight loss [1-6].

It may be considered also in asymptomatic patients presenting abdominal bruit or with palpated thrill in the upper epigastrium at physical examination [2,3,5,7].

The incidence of median arcuate ligament is not clearly established, the majority of literature consisting of isolated reports [1-4, 8].

Either ischemic or neurogenic etiology have been proposed to explain the abdominal pain. However, based on observation of celiac artery compression in asymptomatic patients, questions have emerged as to whether celiac artery compression syndrome really exists [1,7,9].

We report a case of Dunbar's syndrome in a young boy visited in an Emergency Department (ED) after forced ice water ingestion. Our case report seems to confirm the vascular ischemic etiology of the Dunbar's syndrome which has to be considered for the differential diagnosis of epigastric pain in the ED.

## CASE REPORT

A 16 years old boy was referred to our ED for evaluation of recent onset abdominal pain beginning after about 30 minutes after forced ice water ingestion caused by joking compression of a soft PET<sup>®</sup> bottle during drinking. Additional symptoms included nausea, and vomiting. He did not drink alcohol or smoke and no risk factors for Hiv were present. There was not previous anamnestic relevant medical history or surgery.

Physical general exam was normal in contrast with the severe epigastric pain. Rectal examination was unremarkable. Patient's tympanic temperature, heart rate and blood pressure were normal. Other workup including electrocardiographic examination, laboratory investigations including haemogram, c reactive protein, amylase, and lipase, abdominal and chest x ray and abdominal ultrasound were normal. Pain was unresponsive to protonic pump inhibitors or analgesic non steroidal inflammatory drugs medication (NSAIDs).

Correlation with pain onset and forced ice water ingestion suggested to the emergency physician a suspect of traumatic esophagus rupture, like Boerhaave syndrome, and to detect a pneumomediastinum and parietal esophageal abnormality a thoraco-abdominal CT scan, an esophagogram and esophago-endoscopy were performed. But they did not show any abnormality.

Patient showed a spontaneous pain relief and was admitted to hospital stay in the Observation Unit where after 24 hours experienced post prandial epigastric pain and diarrhoea. An angio-MR of the abdomen was requested to rule out ischemic origin of pain correlated to an aortic dissection.

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Angio MR (Fig. 1) pointed out a celiac trunk stenosis (Dunbar's syndrome) confirmed by angiography. Patient was referred to vascular surgeon for surgical treatment.

## DISCUSSION

The median arcuate ligament is formed by muscular fibers that bridge the right and left crura of the diaphragm just anterior to the aorta at T 12–L1 level and it defines the anterior margin of the aortic hiatus [10]. The ligament is highly variable, with appearances ranging from a well defined ligamentous mass to an amorphous area of connective tissue. Supplementary fibers to the celiac plexus normally form a thick, resistant shield immediately ventral to the median arcuate ligament and may contribute to compression of the celiac artery [11,12].

Compression of the celiac axis by extraluminal structures was first described in 1917 by the anatomist Lipshutz [13] but the association with a clinical syndrome was initially described by Harjola [14] in 1963, and by Dunbar [15] *et al.* in 1965 and it is a well documented anatomic variant, reportedly seen in 12.5%-49.7% of patients [16,17].

The median arcuate ligament or Dunbar's syndrome has been documented in monozygotic twins, which suggests that the anatomical relationships, responsible for the disease, are congenital [18].

The stenosis of the celiac trunk occurs either in a too cranial emergence of the celiac artery from the aorta, or in a too caudal insertion of the left crux of the diaphragm on the lumbar vertebral column [7]. Results of conventional angiographic studies dating to the early 1970s showed that the position of the median arcuate ligament, celiac artery, and

aorta varies considerably during respiration and that median arcuate ligament compression is often accentuated during expiration [19,20].

In patients affected by median arcuate ligament syndrome, celiac artery compression occurs during expiration and is more evident with the patient in the erect position [8].

At expiration, in fact, the aorta and its major branches, including the celiac artery move cephalad and this causes worsening of compression, while with inspiration the celiac artery descends lower in the abdominal cavity resulting in a more vertical orientation, which often relieves compression. Additional findings include poststenotic dilatation of the distal celiac trunk, dilated peripancreatic collateral vessels, pancreaticoduodenal artery aneurysms, atheromatous plaques in the aorta [21-23] and mitral valve prolapse [24].

Most of patients are young [1-4,8], thin individuals with acute post prandial (beginning from 15 to 30 minutes after meals), exercise related [1] or chronic epigastric pain, hypopermesis, vomiting, diarrhoea and weight loss [1-6].

Clinical manifestations are vague and diagnosis may rely on findings at duplex and color flow Doppler sonography [2,8], angiographic [2,3,19,20,21,25], CT [3,4,9,21] or MR [2,6,22] imaging but differentiation between clinically relevant celiac artery compression and incidental narrowing may be difficult [16,22].

Physical examination may reveal, classically, a bruit that varies with respiration and position [7] in the midepigastic region or a palpated thrill in the upper epigastrium [2,3,5,7].

The pathophysiologic origin of these symptoms is not clearly understood but the most accepted hypothesis is that



Fig. (1). Stenosis of the celiac artery resulting from median arcuate ligament narrowing (arrow).

compression of the celiac artery by the median arcuate ligament of the diaphragm causes intestinal or diaphragmatic ischaemia, which produces symptoms [1,7,9].

The vascular etiology of symptoms however is still uncertain and controversial.

Others investigators have proposed that these signs and symptoms have a neurogenic origin [26] depending from a compression of the celiac ganglion or a fibrous celiac plexus overlying the celiac trunk which determine an altered gastric electrical rhythm [25].

Questions have emerged also as to whether celiac artery compression syndrome exists from the frequent observation of celiac compression in asymptomatic patients [2,16,22] but it might be due to the presence of an extensive well developed collateral pathways between the celiac artery and the mesenteric circulation [27].

Our case seems to confirm the vascular ischemic etiology of the symptomatology related to Dunbar's syndrome. Abdominal pain emerging 30 minutes after forced ice water ingestion may be interpreted due to decreased gastrointestinal blood flow and increasing metabolic requests correlated to mucosal cold water contact and successive discrepancy ischemia related to gastric parietal vessels vasodilatation.

Despite these controversies, there is a general agreement that patients with celiac axis compression which are symptomatic may benefit from proper open or minimally invasive laparoscopic surgical treatment [3,5,7,28].

It remains to individuate and recognize these patients for a correct differential diagnosis of epigastric pain even in the ED in order to choose the optimal treatment.

#### CONFLICT OF INTEREST

None of the authors have any financial or personal relationship with other people, or organizations, that could inappropriately influence this work.

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