

Available online at [www.sciencedirect.com](http://www.sciencedirect.com)

journal homepage: [www.elsevier.com/locate/radcr](http://www.elsevier.com/locate/radcr)

## Case Report

# Median arcuate ligament syndrome (Dunbar syndrome): A diagnosis not to be underestimated ☆,☆☆

Asma Sghaier, PhD<sup>a,b,c,\*</sup>, Mohamed Amine Jelassi<sup>a,c</sup>, Khalil Fradi<sup>a,b,c</sup>,  
 Mohamed Hédi Mraidha<sup>a,b,c</sup>, Fehmi Hamila<sup>a,b,c</sup>, Sabri Youssef<sup>a,b,c</sup>

<sup>a</sup>Hospital Farhat Hached of Sousse, Rue Ibn El Jazzar, 4002 Sousse, Tunisia

<sup>b</sup>Department of Surgery at Hospital of Farhat Hached of Sousse, Rue Ibn El Jazzar, 4002 Sousse, Tunisia

<sup>c</sup>Faculty of Medicine of Sousse, Rue Mohamed Karoui, 4002 Sousse, Tunisia

### ARTICLE INFO

#### Article history:

Received 14 August 2023

Revised 23 October 2023

Accepted 26 October 2023

Available online 25 November 2023

#### Keywords:

Abdominal pain

MALS

Anatomy

Imaging

Diagnosis

### ABSTRACT

Medial arcuate ligament syndrome (MALS) is an anatomoclinical entity underestimated in most situations due to the lack of specific signs. However, the consequences can be disastrous if the diagnosis is not seriously considered and investigations are not fully undertaken to confirm this hypothesis. We report a case of Median Arcuate Ligament Syndrome in a young woman who presented to the Emergency Department with moderate postprandial pain. All investigations were carried out promptly, with no abnormalities, and the diagnosis was retained on the evidence of a well-performed and appropriately interpreted abdominal CT. This case illustrates a rare cause of epigastralgia simulating a surgical emergency. This condition should be considered when biological and often radiological investigations are inconclusive. The relative youth of the patient is often suggestive, but confirmation is based on a CT scan with injection of a well-technical contrast product, or angiography. Treatment is debatable between a conservative approach and surgery. Surgical approach is discussed depending on the degree of vascular stenosis and the impact on the digestive tract. However, the recurrence of symptoms may be a contributory factor in the therapeutic decision. The impact could be lethal depending on the degree of stenosis and consequently on the repercussions on the digestive tract. A multidisciplinary approach is required for its management.

© 2023 The Authors. Published by Elsevier Inc. on behalf of University of Washington.

This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

**Abbreviations:** MALS, median arcuate ligament syndrome; CT, computed tomography; US, ultrasonography; MRI, magnetic resonance imaging.

☆ Acknowledgments: All the authors read and approved the final manuscript.

☆☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

\* Corresponding author.

E-mail address: [asma.baya@gmail.com](mailto:asma.baya@gmail.com) (A. Sghaier).

<https://doi.org/10.1016/j.radcr.2023.10.058>

1930-0433/© 2023 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

---

## Introduction

Medial arch ligament syndrome median arcuate ligament syndrome (MALS) is still controversial as to its reality, its pathophysiological mechanisms, and even its therapeutic management. From a pathophysiological point of view, the ischemic theory of pain related to extrinsic compression of the celiac trunk is opposed to the neurogenic theory, which is irritation of the nerve fibers of the celiac plexus. Clinically, MALS is classically characterized by epigastric pain, vomiting, and weight loss [1]. The diagnosis of MALS is one of exclusion, which is made after ruling out all other causes of chronic abdominal pain, and is confirmed by invasive and/or non-invasive vascular radiological examinations and/or noninvasive vascular radiology [2]. In terms of treatment, the majority of authors accept the value of surgical treatment for symptomatic patients. The laparoscopic approach might be a potential alternative treatment. Few cases of MALS treated laparoscopically have been described in the medical literature. We report a case for which a conservative approach was retained after concertation with the medical staff. This case has been reported in line with SCARE criteria [3].

---

## Case presentation

A 44-year-old white female presented to the Emergency Department with moderate postprandial abdominal pain. The patient has a past medical history of open appendectomy for acute appendicitis with no postoperative complications. Apart from that, she has no known relevant personal, family, or psycho-social medical history. The patient reported a prior similar incident 5 years ago, which had spontaneously resolved without requiring any treatment. The semiological characteristics of these pains were such that pain, which was described as “burning,” was mainly located in the epigastrium and right hypochondrium. It appeared suddenly, approximately 1 hour after lunch, and was progressively worsening. Nausea and vomiting were also reported. Furthermore, a recent loss of appetite and early satiety were reported by the patient during the anamnesis.

Physical examination revealed a solid McBurney incision scar, moderate abdominal distension, and epigastric tenderness. The electrocardiogram (ECG) was normal.

As routine, blood tests were performed, and no biological stigma of inflammatory systemic response was found. Acute hepato-biliary conditions were ruled out. The pancreatic blood test was normal, with lipasemia within the usual range.

An upper gastrointestinal endoscopy was performed to exclude gastritis, peptic ulcer disease was normal.

The abdominal injected CT-scan (Fig. 1) that was performed showed no evidence of occlusive syndrome.

The liver, the gall bladder, the bile ducts, the stomach, and the intestines were unscathed. But an abnormal striction of the celiac trunk, at its origin, followed by a poststenotic dilation of the same artery, was unveiled. This striction seemed to be caused by an extrinsic anatomic structure: the median arcuate ligament. We estimate 15% of the degree of stenosis. In

addition, a nonmodal presentation of the right hepatic artery had been described, with the artery arising directly from the aorta (Figs. 2 and 3).

Patient has been symptomatic treatment based on Level I analgesics and proton pump inhibitors. Evolution was good. After 72 hours, she has been then referred to the vascular Surgery Department, in order to benefit from adequate symptomatic treatment. The latter was normal. It was only at this stage of investigation that the assumption of a causality link between the abdominal pain and the abnormal anatomic position of the median arcuate ligament seemed plausible.

The decision after the medical staff concertation was in favor of conservative management because the degree of stenosis was not important and with no effect on the vascular system and neither consequence on digestif tract subject. Number of recurrences was not significant. Nevertheless, regular clinical and radiological follow-up was indicated.

---

## Discussion

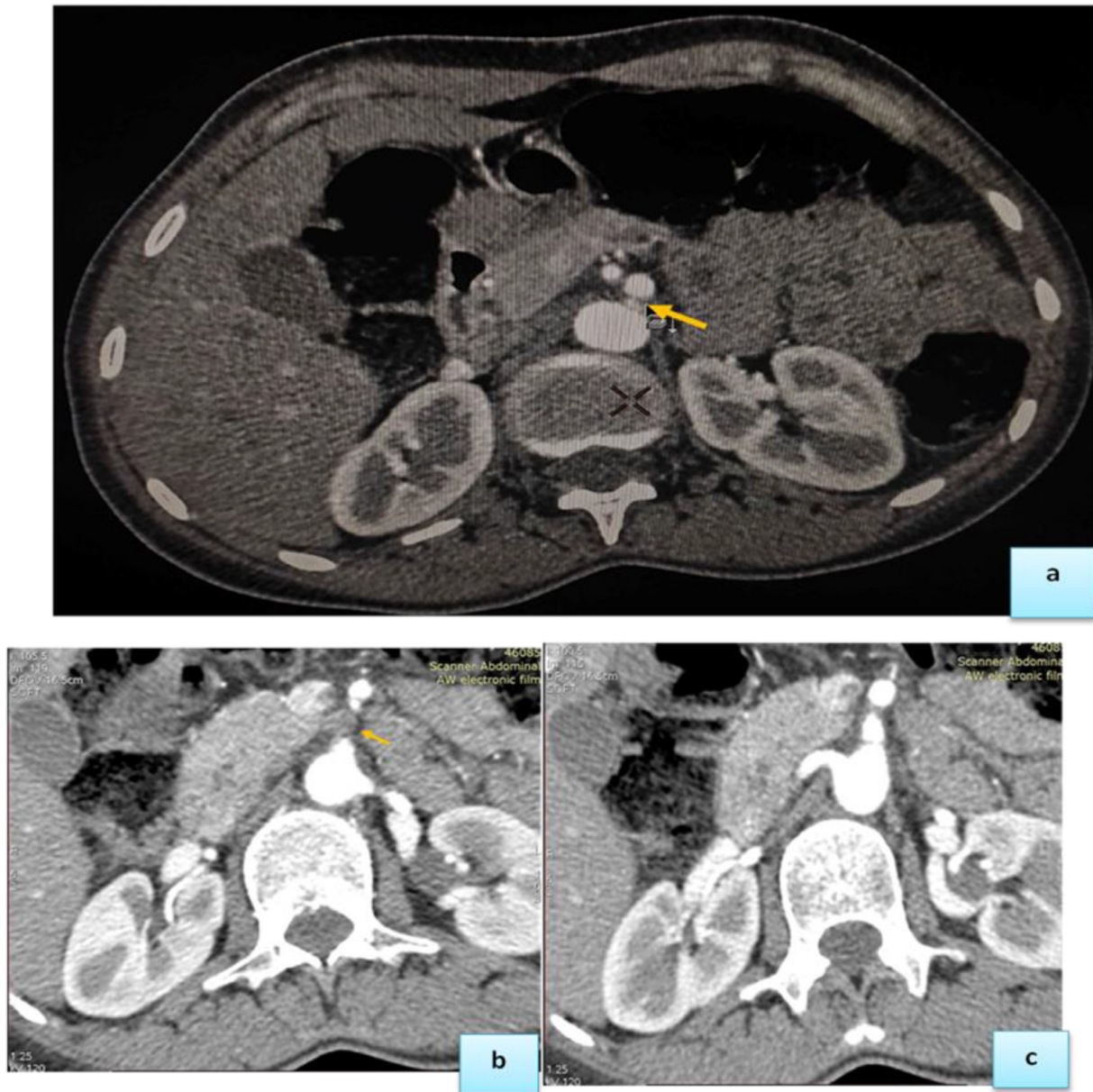
Also known as Dunbar syndrome, celiac artery compression syndrome, or celiac axis syndrome, Median Arcuate Ligament Syndrome (MALS) is a rare vascular compression syndrome, found only in 2 out of 100,000 patients [4]. Reduced blood flow through this artery is thought to be accountable for the variety of symptoms observed in this syndrome. Celiac ganglion compression in MALS is a contributory factor [5,6].

MALS was described first anatomically in 1917 by Lipshutz [7]. The clinical entity was described by Harjola in 1963 [8]. This surgeon was also the first to perform the first median arcuate ligament release in the world. Later, a larger case series of patients have been published.

The median arcuate ligament (MAL) is a strong fibrous structure linking the left and right diaphragm crura. These 3 structures form, together, the aortic hiatus (next to the vertebrae T<sub>12</sub>-L<sub>1</sub>) [9,10]. The position of the MAL and the origin of the celiac trunk may vary between individuals [11].

MALS is a rare, but not-to-forget, compression syndrome due to abnormally lower insertion of the diaphragm crura and MAL, or abnormally higher origin of the celiac artery, leading to extrinsic compression of the proximal celiac artery. The stress on the compressed artery is likely to increase at expiration, due to the upward movement of the celiac axis [12]. Hyperplastic intimal changes in the celiac artery may be associated with the compression [5]. Neuropathic pain is thought to be an aggravating factor in MALS. This is explained by the fact that the celiac ganglion is located close to the celiac artery [5,6]. MALS is more common in women (sex ratio = 4:1) aged between 30 and 50 years old [6–10]. No other risk factors have been found in literature. A variety of symptoms can be observed in a patient with MALS. A study describing a 20-year experience of surgical treatment of MALS revealed that the most frequent symptoms are abdominal pain (94%), postprandial abdominal pain (80%), weight loss (50%), bloating (39%), nausea and vomiting (55.6%), and abdominal pain triggered by exercise (8%) [14].

Clinical examination findings can include abdominal bruits, epigastric tenderness, and weight loss [8]. Most of



**Fig. 1 – Abdominal scanner performed as an emergency. (A) Contrast-enhanced CT axial image of the abdomen shows proximal stenosis of the celiac artery accompanied by poststenotic dilatation, although it is better appreciated on a Sagittal CT image. (B, C) Section with magnification.**

the time, MALS is an asymptomatic condition. The lack of symptoms could be explained by the development of collateral circulation, usually from the superior mesenteric artery (SMA) [15].

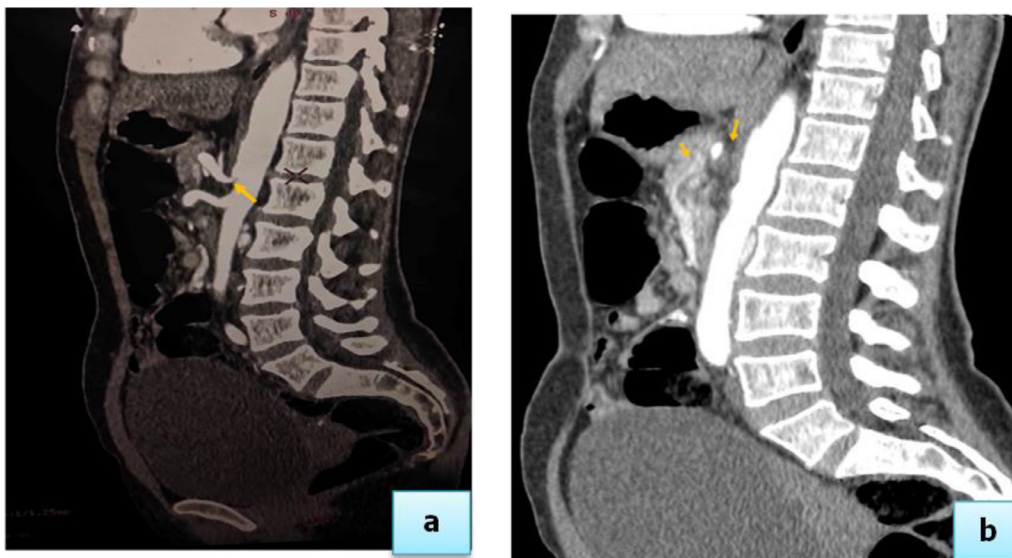
Celiac artery compression by the MAL was found in only 2.8% of cases in a retrospective database analysis where CT abdominal angiography was performed for various indications. Among them, only 3 presented with symptoms of MALS [16].

As MALS is an exclusion diagnosis [3,13–17], alternative causes of abdominal pain and other digestive symptoms should always be ruled out. A formal gastroenterology review is advisable. When clinical suspicion of MALS arises, it is

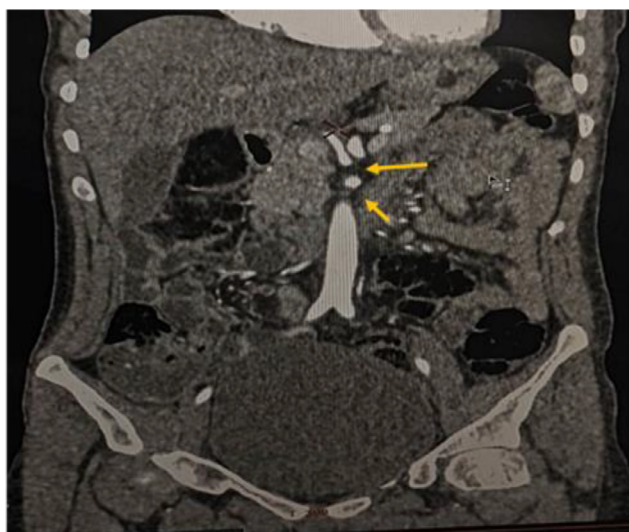
recommended to perform a duplex ultrasound (DUS) study of the celiac artery [17–19].

DUS can be a good initial screening tool as it has no risk of ionizing radiation and is cheaper. However, it is operator-dependent and needs an experienced operator to assess and show the modifications. DUS signs include: (1) poststenotic dilatation of the celiac trunk, and (2) elevated blood velocities exaggerated during expiration.

Computed tomography angiography (CTA) allows a special visualization of the pressed celiac trunk. However, compared to DUS, CTA involves ionizing radiation and needs contrast, which can be crippling in patients with renal dysfunction.



**Fig. 2 – Axial reconstruction. (A) Contrast-enhanced CT of the abdomen, parasagittal reconstruction, shows kinking of the proximal celiac artery, resulting in a characteristic hooked appearance with poststenotic dilatation. Note the absence of atherosclerosis. (B) Contrast-enhanced CT of the abdomen, parasagittal reconstruction, shows poststenotic dilatation of the celiac artery.**



**Fig. 3 – Abdominal scanner performed as an emergency. Frontal section showing stenosis of the celiac trunk due to the arcuate ligament.**

Magnetic resonance angiography (MRA) is a radiation-free alternative modality.

Conventional angiography remains “the gold standard to show dynamic compression of the celiac artery” [17]. Breathing maneuvers can be very helpful for diagnosis. Angiography is also indicated to evaluate patients as they develop symptoms in the postoperative period [20].

The aims of the treatment of MALS are (1) decompression of celiac trunk so that normal blood flow is restored, and (2) pain management by celiac ganglionectomy [17]. Many treat-

ment options have been tried and described. Nowadays, the laparoscopic release of coeliac artery and celiac neurolysis (with intraoperative duplex ultrasound) has established itself as a usual surgical option [17–20]. Advantages of laparoscopic method include small incisions, low risk of complications, and improved view during surgery [17]. But during a laparoscopic procedure, it is more likely to incompletely release the celiac artery and, there is a higher risk of abdominal aorta injury [21]. Robotic-assisted release of compression and celiac neurolysis is the new trend [22,23].

Percutaneous transluminal angioplasty (PTA) is indicated when persistent celiac flow abnormalities are noted on preoperative duplex ultrasound postoperative imaging, or in cases of postchirurgical recurrence [17]. Vascular reconstruction is a last resort weapon [17,20].

At least Finally, we need to be cautious about the main differential diagnoses, which include

- Hyperalgesic ulcer attack Epigastric pain is the most common clinical sign. Pain is improved by taking proton pump inhibitors. Diagnosis is confirmed by upper gastrointestinal endoscopy. Imaging is of no interest in confirming the diagnosis.
- Acute pancreatitis There are many similarities with MALS in the clinical presentation and pain semiology. Imaging in this case is of considerable prognostic value. Furthermore, in specific situations, this examination can be of genuine therapeutic benefit
- A perforated ulcer the symptoms of ulcer perforation are characteristic. Imaging is of primary diagnostic importance. Standard radiography may show a gaseous crescent in 40% of cases. in appropriate cases, the abdominal CT scan may show direct signs of perforation: an extra digestive gas bubble indirect signs: intraperitoneal fluid effu-

sion, or densification of the prebulbar fat. The abdominal CT scan with upper digestive opacification with hydrosolubles will show an extra digestive fluid leak at the bulbous region.

- Aorto-mesenteric clamp syndrome Superior mesenteric artery (SMA) syndrome, also known as Willkie syndrome or aortomesenteric duodenal compression syndrome, is a rare acquired vascular compression disorder in which acute angulation of the SMA results in compression of the third part of the duodenum, leading to obstruction. Patients with SMA syndrome may present acutely, with chronic symptomatology, or with an acute exacerbation of chronic symptoms: acute presentation is usually characterized by signs and symptoms of duodenal obstruction chronic cases may present with long-standing vague abdominal symptoms, early satiety and anorexia, or recurrent episodes of abdominal pain, associated with vomiting. CT-tomography and magnetic resonance angiography (CTA/MRA) enable visualization of vascular compression of the duodenum and measurement of aortomesenteric distance. Normally, the aortomesenteric angle and aortomesenteric distance are 28°–65 and 10–34 mm, respectively. In SMA syndrome, both parameters are reduced, with values of 6°–22° and 2–8 mm.

Through this case, we have chosen to focus on the diagnostic aspect of this syndrome and the possible difficulties we may encounter in making the diagnosis. The therapeutic approach remains a subject of debate and no consensus has yet been established. undoubtedly, the degree of stenosis is the cornerstone for the surgical option.

The classical, laparoscopic, and even robotic approaches and the performance of each one will be the subject of better prospective studies and randomized controlled trials to assess the validity of one compared with the other.

These considerations also apply to the plethora of additional examinations to be carried out as part of the diagnostic confirmation strategy.

## Conclusion

MALS is a rare condition. Its clinical suspicion arises when usual medical conditions causing abdominal pain or other digestive symptoms are ruled out. Duplex Ultrasonography (DUS) and CT abdominal angiography are nowadays' most used imaging techniques when MALS is highly suspected. MALS treatment is based on celiac trunk decompression with celiac lymphadenectomy, followed, in specific cases, by a percutaneous transluminal angioplasty. This can be done through open surgery, laparoscopic or robot-assisted routes. Vascular reconstruction of celiac trunk remains a solution in sectioned cases.

## Ethical approval

N/A Ethical approval is exempt/waived at our institution.

## Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2023.10.058](https://doi.org/10.1016/j.radcr.2023.10.058).

## REFERENCES

- [1] Jarra J, Berard X, Ducasse E, Biscayb D, Paillet A, Sassoust G, et al. Traitement du syndrome du ligament arcué médian par voie coelioscopique. *J Mal Vasc* 2008;33:30–4.
- [2] Narwani P, Khanna N, Rajendran I, Kaawan Hesham, Al-Sam Rafid. Median arcuate ligament syndrome diagnosis on computed tomography: what a radiologist needs to know. *Radiol Case Rep* 2021;16:3614–17.
- [3] Agha RA, Franchi T, Sohrabi C, Mathew G for the SCARE Group. The SCARE 2020 Guideline: Updating consensus Surgical CAse REport (SCARE) Guidelines. *Int J Surg* 2020;84:226–30.
- [4] Iobst TP, Lamb KM, Spitzer SL, Patel RN, Alrefai SS. Median arcuate ligament syndrome. *Cureus*. 14(2):e22106.
- [5] Bech FR. Celiac artery compression syndromes. *Surg Clin North Am* 1997;77(2):40924.
- [6] Sultan S, Hynes N. PS160. Management of median arcuate ligament syndrome (MALS) with decompression and coeliac ganglion sympathectomy (CGS) for chronic mesenteric ischaemia (CMI). Procedural, clinical and enduring results with quality-adjusted time spent without symptoms of disease and toxicity of treatment (Q-TWiST). *J Vasc Surg* 2010;51:62S 6, Supplement.
- [7] Lipshutz B. A composite study of the coeliac axis artery. *Ann Surg* 1917;65:159–69.
- [8] Harjola PT. A rare obstruction of the coeliac artery. Report of a case. *Ann Chir Gynaecol Fenn* 1963;52:547–50.
- [9] Gaillard F, Moore C, Glick Y, et al. Celiac artery compression syndrome. Reference article, [Radiopaedia.org](https://radiopaedia.org) (accessed on 06.06.23)
- [10] Loukas M, Pinyard J, Vaid S, Kinsella C, Tariq A, Tubbs RS. Clinical anatomy of celiac artery compression syndrome: a review. *Clin Anat* 2007;20:612–17.
- [11] Goodall R, Langridge B, Onida S, Ellis M, Lane T, Davies AH. Median arcuate ligament syndrome. *J Vasc Surg* 2020;71(6):2170–6.
- [12] Reuter SR, Bernstein EF. The anatomic basis for respiratory variation in median arcuate ligament compression of the celiac artery. *Surgery* 1973;73(3):381–5.
- [13] Kim EN, Lamb K, Relles D, Moudgill N, DiMuzio PJ, Eisenberg JA. Median arcuate ligament syndrome-review of this rare disease. *JAMA Surg* 2016;151(5):471–7.
- [14] Iqbal S, Chaudhary M. Median arcuate ligament syndrome (Dunbar syndrome). *Cardiovasc Diagn Ther* 2021;11(5):1172–6.
- [15] Heo S, Kim HJ, Kim B, Lee JH, Kim J, Kim JK. Clinical impact of collateral circulation in patients with median arcuate ligament syndrome. *Diagn Interv Radiol* 2018;24(4):181.

- 
- [16] Gümüş H, Gümüş M, Tekbaş G, Önder H, Ekici F, Çetinçakmak MG, et al. Clinical and multidetector computed tomography findings of patients with median arcuate ligament syndrome. *Clin Imaging*. 2012;36(5):5225.
- [17] Iqbal S, Chaudhary M. Median arcuate ligament syndrome (Dunbar syndrome). *Cardiovasc Diagn Ther* 2021;11(5):1172–6.
- [18] Scholbach T. Celiac artery compression syndrome in children, adolescents, and young adults: clinical and color duplex sonographic features in a series of 59 cases. *J Ultrasound Med* 2006;25:299–305.
- [19] Björck M, Koelemay M, Acosta S, Bastos Goncalves F, Kölbl T, et al. Editor's Choice - Management of the Diseases of Mesenteric Arteries and Veins: Clinical Practice Guidelines of the European Society of Vascular Surgery (ESVS). *Eur J Vasc Endovasc Surg* 2017;53(4):460–510.
- [20] Duffy AJ, Panait L, Eisenberg D, Bell RL, Roberts KE, Sumpio B. Management of median arcuate ligament syndrome: a new paradigm. *Ann Vasc Surg* 2009;23(6):778–84.
- [21] Takach TJ, Livesay JJ, Reul GJ Jr, Cooley DA. Celiac compression syndrome: tailored therapy based on intraoperative findings. *J Am Coll Surg* 1996;183:606–10  
Erratum in: *J Am Coll Surg* 1997;184:439.
- [22] Do MV, Smith TA, Bazan HA, Sternbergh WC 3rd, Abbas AE, Richardson WS. Laparoscopic versus robot-assisted surgery for median arcuate ligament syndrome. *Surg Endosc* 2013;27:4060–6.
- [23] Jaik NP, Stawicki SP, Weger NS, Lukaszczuk JJ. Celiac artery compression syndrome: successful utilization of robotic-assisted laparoscopic approach. *J Gastrointest Liver Dis* 2007;16(1):93–6.