Cite as: Smereczyński A, Kołaczyk K, Kiedrowicz R: New perspective on median arcuate ligament syndrome. Case reports. J Ultrason 2021; 21: e234–e236. doi: 10.15557/JoU.2021.0037.

Submitted: 02.11.2020
Accepted: 30.12.2020
Published: 16.08.2021

New perspective on median arcuate ligament syndrome. Case reports

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DOI: 10.15557/JoU.2021.0037

Keywords
median arcuate ligament syndrome, coeliac trunk, ultrasound

Abstract

A large group of patients with significant asymptomatic or low-symptomatic coeliac trunk stenosis require deeper consideration. On angiography, CT and MRI, 10–24% of examined patients are found to have their coeliac trunk compressed by the median arcuate ligament of the diaphragm. The associated median arcuate ligament syndrome, which is also called coeliac trunk compression syndrome or Dunbar syndrome, is rarely fully symptomatic. It is estimated that there are up to 7% of patients with such a clinical presentation. An asymptomatic or low-symptomatic course of the disease in patients with the syndrome is mainly explained by a developed collateral circulation, particularly involving the arterial arcades of the head of pancreas. In such cases, CT angiography detects collateral circulation in 22–69.6% of examined patients. The present authors often observed coeliac trunk blood flow to normalise in a standing position. According to them, the main causative factor for this phenomenon is the deflection of the coeliac trunk and its compression against the aorta by a lowered left lobe of the liver. The researchers observed it in many individuals; in this study, 5 cases are presented.

Introduction

Median arcuate ligament syndrome (MALS), which is also called coeliac trunk compression syndrome or Dunbar syndrome, is rarely fully symptomatic. It is estimated that there are up to 7% of all cases with such a clinical presentation; therefore, the syndrome is diagnosed primarily by excluding other diseases with a similar set of symptoms[1–4].A possible explanation for the disproportion between fully symptomatic cases and low-symptomatic or asymptomatic ones is the development of competent collateral circulation originating mainly from the superior mesenteric artery, and a low degree of stenosis[5]. A known phenomenon in MALS is a haemodynamically significant change of blood flow velocity in the coeliac trunk at extreme respiratory displacements, which is observed in patients in a supine position. The flow improves and stabilises radically in a standing position, which was demonstrated in a number of case studies[6,7] and in one large study including 180 cases[8]. The authors of those publications explain this change by the pressure exerted by the arcuate ligament of the diaphragm on the coeliac trunk being released. The present authors' observations indicate that another important factor which straightens the coeliac trunk is a caudal descent of the left lobe of the liver.

Case reports

Out of a group of 50 patients with MALS, the authors of the present study selected five individuals who met the criterion proposed by Moneta *et al.*[9], i.e. PSV >200 cm/s. Comparative sonograms were obtained at breathhold in a supine and standing position by applying a 2–5 MHz convex transducer to the epigastric fossa.

Case 1 is that of an 18-year-old woman with sonographic signs of MALS in a supine position. Once the patient returned to an upright position, the left lobe of the liver descended caudally and straightened the coeliac trunk. The difference in the distance from the dorsal surface of the

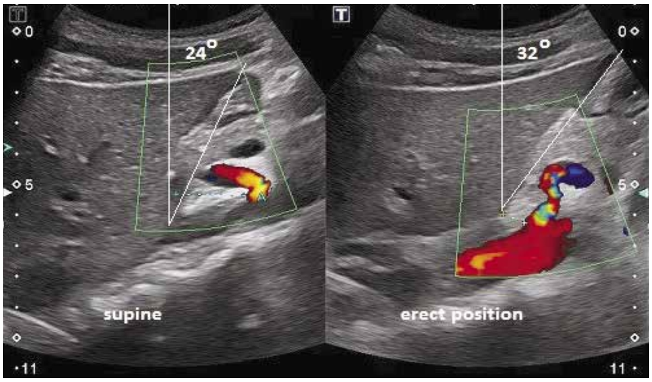

Fig. 1. *An 18-year-old woman with MALS. The left side of the sonogram shows an image of the anomaly in a supine position and the right side of the sonogram shows an image of the anomaly in a standing position*

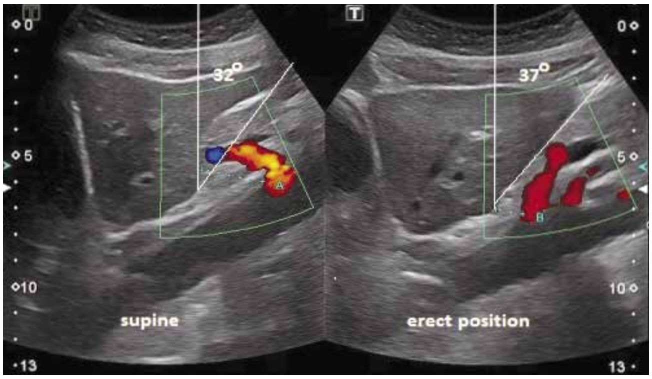

Fig. 2. *A 48-year-old woman with MALS. The left side of the sonogram shows an image of the anomaly in a supine position and the right side of the sonogram shows an image of the anomaly in a standing position*

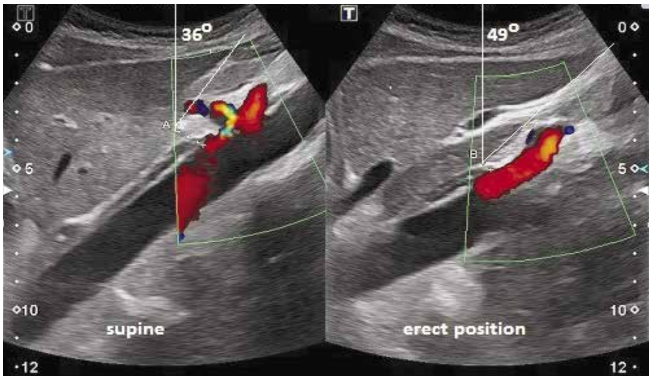

Fig. 3. *A 35-year-old woman with MALS. The left side of the sonogram shows an image of the anomaly in a supine position and the right side of the sonogram shows an image of the anomaly in a standing position*

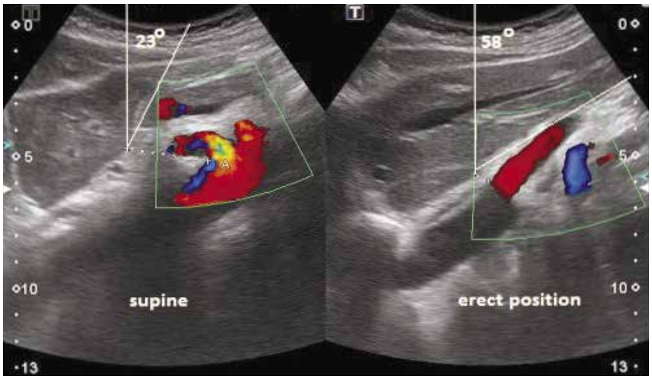

Fig. 4. *A 27-year-old man with MALS. The left side of the sonogram shows an image of the anomaly in a supine position and the right side of the sonogram shows an image of the anomaly in a standing position*

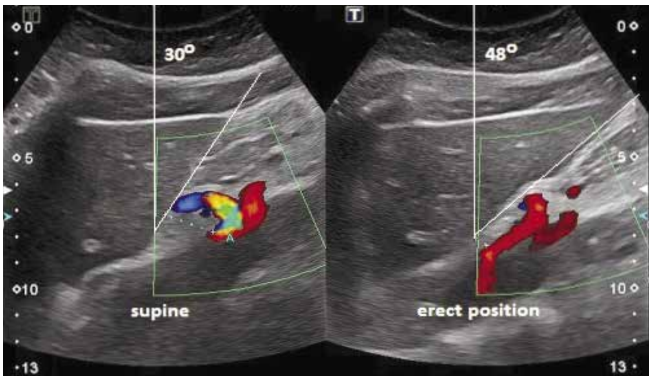

Fig. 5. *A 42-year-old woman with MALS. The left side of the sonogram shows an image of the anomaly in a supine position and the right side of the sonogram shows an image of the anomaly in a standing position*

left lobe of the liver to the origin of the coeliac trunk was reduced by 13.9 mm (Fig. 1). The angle of rotation of the liver was 8°.

Case 2 is that of a 48-year-old woman with sonographic signs of MALS in a supine position. Upon assuming an upright position, the left lobe of the liver straightened the coeliac trunk and approached it by 12.7 mm (Fig. 2). The angle of rotation of the liver was 5° (Fig. 2).

Case 3 is that of a 35-year-old woman with classic clinical and sonographic signs of MALS. In a standing position, the left lobe of the liver approached the origin of the coeliac trunk by 6.5 mm and the angle of rotation of the liver reached 13° (Fig. 3).

Case 4 is that of a 27-year-old man. In that patient, in an upright position the distance between the liver and the origin of the coeliac trunk was shortened by 23.7 mm. In addition, the liver rotated 35° (Fig. 4).

Case 5 is that of a 42-year-old woman with sonographic signs of MALS. In a standing position, her liver approached the origin of the coeliac trunk by 13.8 mm and rotated 18° (Fig. 5).

Discussion

In MALS, variable displacement of the coeliac trunk is assigned to a changing position of the median arcuate ligament of the diaphragm during the respiratory cycle. During expiration, the ligament is considered to press against the coeliac trunk as a result of diaphragm movement, which moves it caudally, while during inspiration, the ligament is moved in a cephalad direction, thus releasing the pressure. This mechanism is hard to question; however, research by the authors of the current study demonstrated that the left lobe of the liver also participates in caudal deflection of the coeliac trunk, provided that the lobe is well-developed. According to the observations by these researchers, such an anatomical variant of the left lobe of the liver (in the sagittal plane) is found in slim individuals with coeliac trunk compression, who account for a decisive majority of individuals with MALS[10]. It is worth pointing out that the liver is the largest organ in the human body, which weighs between 1500 and 1700 g in deceased men and is 500–800 g heavier when filled with blood; the respective measurements in women are 200 g lower. In an upright position, as a result of gravity, it is primarily the liver that lowers the diaphragm in a caudal direction, with the posterior surface of the left lobe inverting the coeliac trunk in the same direction. This also happens as a result of a slight anterior rotation of the liver in the transverse axis, since it is firmly attached to the posterior abdominal wall by the bare area and hepatic veins, which drain into the inferior vena cava. This movement is controlled from the top by the coronary ligament and falciform ligament and from the front by the round ligament of the liver. In addition, the mechanism of the descent and rotation of the liver is supported by the gravitation of the inferior parts of the lungs and the heart, which is located in the anterior part of the thoracic cavity. Thus, in the majority of patients with a fluctuating movement of the coeliac trunk during inspiration when lying on their back and in an upright position, the caudal deflection of the coeliac trunk will be caused by two concomitant factors: release of pressure exerted by the arcuate ligament of the diaphragm, and deflection of the distal segment of the trunk by a descending left lobe of the liver. Another factor that presses the liver dorsally, and, consequently, the coeliac trunk as well, is the activation of abdominal muscles in a standing position which stabilise the trunk. As a result, it is not surprising that the majority of patients with functional stenosis of the coeliac trunk have no symptoms of upper gastrointestinal tract ischaemia upon eating a meal in a standing position. However, ischaemia is expected if such a patient lies down after a meal or eats in a similar body position (a bedridden patient). The symptoms of MALS will become permanent if, as a result of many years of an alternating mechanism, coeliac trunk stenosis becomes persistent. In such cases, endothelial hyperplasia, proliferation of elastic fibres in the tunica media and changes in the adventitia were observed[5]. In addition, a blood clot can fill the lumen of the trunk. In patients with MALS, it is not uncommon to see an aneurysm-like dilation of the coeliac trunk past the site of stenosis. In this syndrome, one can also expect to see the development of aneurysms, particularly on the pancreaticoduodenal arteries and gastroduodenal artery, which are sometimes diagnosed upon rupturing and are fatal in approximately 30% of cases[5]. Therefore, MALS can result in a number of serious vascular sequelae. When considering clinical symptoms and surgical treatment of the condition, one should also take into account changes in the coeliac plexus, which are also the result of the pressure exerted by the arcuate ligament of the diaphragm. Upon analysis of comparative sonograms of the five MALS cases presented in this study, in the first case a slight descending motion of the pancreas was also revealed. However, upon the patient returning to an upright position, it is primarily the liver that straightens the coeliac trunk. Together with the pancreas, the liver plays an important role in the pathological mechanism leading to renal nutcracker syndrome by compressing the superior mesenteric artery and subsequently constricting the left renal vein, as demonstrated by Yun *et al*.[11].

Conflict of interest

The authors do not report any financial or personal affiliations to persons or organisations that could adversely affect the content of or claim to have rights to this publication..

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