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Ultrasound of the median arcuate ligament syndrome: a new approach to diagnosis.

Hannes Gruber[1], Alexander Loizides[1], Siegfried Peer[1], Ingrid Gruber[2]

[1] Innsbruck Medical University, Department of Radiology, Innsbruck, Austria
[2] Innsbruck Medical University, Department of Vascular Surgery, Innsbruck, Austria

Abstract

Introduction: The currently accepted pathophysiological concept behind median arcuate ligament syndrome (MALS) is vascular compromise by atypically positioned median arcuate ligament. Despite many articles on MALS, only sparse data on the outcome in general and the rather poor outcome after intervention have been reported and this makes at least questionable the current concepts of the underlying disease.

Material and methods: A total of 364 patients with suspected celiac trunk pathology underwent a standardized ultrasound assessment procedure: suspected diagnostic features for MALS such as typically elevated peak flow velocities (PV) in the celiac trunk or atypical celiac trunk deflection-angles (DA) were defined in patients and in 20 matched volunteers.

Results: All the 6 retrospectively clearly diagnosed MALS-patients as well as 40% (8/20) of volunteers presented a DA of over 50°. MALS-patients presented a mean inspiratory PV of 172cm/s (± 40.9 cm/s), a mean expiratory PV of 425cm/s (±130.1 cm/s) with a PV-amplitude of 249.1% (±68.9%). Volunteers presented a mean inspiratory PV of 126.9cm/s (±42 cm/s), a mean expiratory PV of 209.9cm/s (±80.1 cm/s) with a PV-amplitude of 169.4% (±54.3%).

Conclusions: The combination of a maximum expiratory PV of over350 cm/s and a DA higher than 50° seems to be a most reliable indicator for MALS in this small series of patients. Based on these data we propose that functional ultrasound should be the first line in screening for MALS. However, a clear pathophysiological definition of MALS remains still obscure.

Keywords: median arcuate ligament syndrome, Doppler sonography, celiac trunk.

Introduction

Atypically positioned median arcuate ligament – extension of the aortic hiatus – are blamed for mechanically causing the so called "Dunbar-" or "median arcuate ligament syndrome" (MALS). This concept was raised by Dunbar et al in 1965 [1] on 15 young symptomatic patients examined by conventional angiography. Of these 15 subjects 13 underwent surgery with subsequent relief of symptoms. In 1972 Levin et al assessing asymptomatic subjects, found that the angiographic changes, defined according to Dunbar et al, are also seen in 24% of unselected healthy people [2]. When in 1985 Reilly et al [3] evaluated patients with such vascular changes and only 53% and 76%, became asymptomatic after surgery ± revascularization, the concern, that "reliable diagnosis awaits definition … as are the pathophysiologic mechanisms", was raised for the first time.

In the subsequent years a flood of investigations followed (to date about 100 PubMed entries), which transferred the initially defined angiographic features to, at that time, "new" modalities such as ultrasonography (US), computed tomography (CT) and magnetic resonance imaging (MRI). When assessing MALS with CT-Angiography (CTA) in 2005 Horton et al [4] stated very vaguely: "median arcuate ligament syndrome is a controversial entity", "abnormal insertion of the arcuate ligament can be found in asymptomatic people", "in a

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Corresponding author: Hannes Gruber, MD
Innsbruck Medical University,
Department of Radiology
Anichstrasse 35, 6020 Innsbruck, Austria
Tel: +43/512/504/22761
Fax: +43/512/504/22758
E-mail: hannes.gruber@i-med.ac.at

small subset of patients surgery helps" and "CTA can play a role in diagnosis". In a later publication Gloviczki et al [5] asked the question "does it [i.e. MALS; author´s remark] really exist based on De Cecchis et al [6] paper and on currently questionable data". However, there was still little concern regarding the rather sparse data and poor or unexplainable outcome of intervention in patients with suspected MALS. Loukas et al [7] in their anatomical review ended at the same point where Levin et al [2] or Reilly et al [3] had already stated decades ago "celiac artery compression syndrome is a broadly interpreted phenomenon" with "unknown pathophysiology", "… which result in inaccurate diagnosis and consequently in inaccurate therapy". The most recent publication even implemented ECG-gated CT-Angiography [8] with the result that "the decision to treat the complications of MALS is clear, but to intervene when a median arcuate ligament variant anatomy is diagnosed incidentally is less so", "not the presence of the ligament sling alone … that should prompt consideration of surgical management". To date reliable diagnostic features are still vague as is our understanding of the "true" pathophysiologic mechanisms. Thus indications for treatment remain in question.

Published case reports or case series on the use of US, with its possibility to measure flow and function under real-time conditions even upright subjects [9-11] pointed out possible functional or collateral mechanisms that might be demonstrated by sonography but are not supported by data from other (non-functional) modalities.

Our **aim** was to provide clear functional imaging data by comparing definite MALS patients with healthy volunteers and as such to take the first step to "true" diagnosis based on better therapeutic concepts.

Material and methods

Between August 1999 and March 2011 364 patients were assigned to standardized US assessment for evaluation of celiac trunk pathology ("possible MALS") during daily routine. Using gray-scale and Doppler US, features possibly indicative for MALS such as celiac trunk ex- and inspiratory peak velocity (PV; including percent of amplitude change) and the end-expiratory upturn-angle (deflection-angle; DA) of the trunk versus its maximum inspiratory position were assessed and documented. Conscious patients were examined in supine position with a C5-2 broadband curved-array transducer on a HDI 5000 or iU22™ (ATL-Philips, Wash., USA). Arterial wall calcifications (relevant atherosclerotic stenosis was excluded by preliminary expiratory PV assessment) in the celiac trunk and its excursion angle in maximum inspiration vs. maximum expiration were assessed on gray-scale images and measured using the standard caliper available on the institutional Agfa®-PACS (Agfa AG, Mortsel, BEL): a DA of more than 50° was assigned a "positive deflection angle" (DA+) (fig 1, fig 2). The threshold of 50° was defined empirically in retrospect but arbitrarily based on the deflection angles of the MALS-patients. Subsequently, PV in the celiac artery was measured three times each in maximum expiration and inspiration. Measurement values were averaged to correct for deviations in individual measurements.

Out of these 364 patients only 6 (mean age 35.8 years ± 17.2) retrospectively clearly fulfilled the criteria for inclusion in this evaluation, i.e. were "true" MALS patients based on clinical signs and symptoms: clinical data had to include chronic, postprandial pain ± vomiting and weight loss with exclusion of any other possible causes. If available, other imaging modalities (CT, MRI, conventional angiography) had to present consistent features of MALS

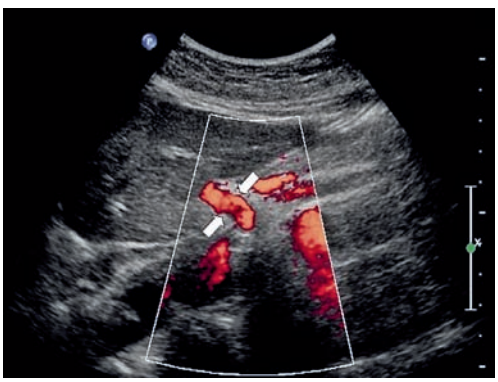

Fig 1. Doppler ultrasound scan of a 21 year old female patient with a positive deflection angle (DA+) of the celiac trunk (arrows) of about 80° during maximum expiration.

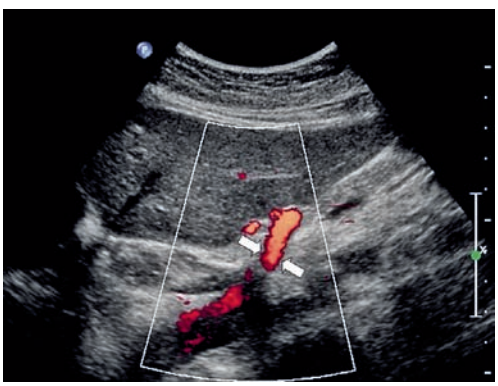

Fig 2. The same case as in fig 1 during maximum inspiration.

such as "deformed" celiac trunk angiograms ± depiction of a externally compromising soft tissue string (median arcuate ligament). For all patients but one a corresponding CT (including CT-Angiography) was available; one patient had undergone even magnetic resonance imaging (MRI) (including contrast enhanced MR-Angiography) and two had undergone conventional Angiography. Only one patient had undergone MR-Angiography (including contrast enhanced MR-Angiography). Of these patients three had undergone surgical repair (decompression) ± percutaneous transarterial angioplasty (PTA) and three are still under clinical observation while scheduled for intervention.

A total of 20 age matched asymptomatic volunteers (mean age 37.7 years ± 8.8; mean body mass index [BMI] 22.4 kg/m² ± 2.8) underwent the same standardized ultrasound algorithm for comparison and validation of patient features.

Informed consent on the utilization of anonymized data for study purposes was obtained from all subjects according to the World Medical Association Declaration of Helsinki (59th WMA Assembly, Seoul, 2008) [12]. Institutional review board approval was granted by means of a general waiver for studies with retrospective data analysis (Ethikkommission, Med. Univ. Innsbruck; 2009-02-20). Each image, measurement and assessment was documented for statistical evaluation using SPSS® (PASW Statistics, Version 18.0.0, Chicago, IL, USA) in an Microsoft Excel®-file (Microsoft Corp., Redmont, Wash., USA) or in our institutional Agfa®-PACS (Agfa AG, Mortsel, BEL).

Specificity, sensitivity, positive and negative predictive value (PPV; NPV) and prevalence were calculated for inspiratory PV, expiratory PV, percent of amplitude change, the DA+ and the combination of a DA+ and the according expiratory PV. Correlation tests (Kendall-τ; Spearman-ρ) were performed to assess possible coherences between volunteer data and BMI.

Statistical differences of the Doppler-US data between patients and volunteers were calculated by non-parametric Mann-Whitney-U(MWU)-testing (significance level $\alpha \leq 0.05$ for 2-tailed and $\alpha \leq 0.025$ for 1-tailed testing) and by contingency testing concerning statistical independence of cross-tabulated categorical data with a χ^2- test based Φ-coefficient calculation (significance on stochastic independence at $\alpha=0{,}01$; χ^2 1-tailed with 1 degree of freedom) based on the following empirically defined thresholds: inspiratory PV =150 cm/s, expiratory PV = 350 cm/s and DA+ according to the distribution of features in the available data.

Box-plots were built for illustration of inspiratory PV-, expiratory PV-, and PV-amplitude-distribution.

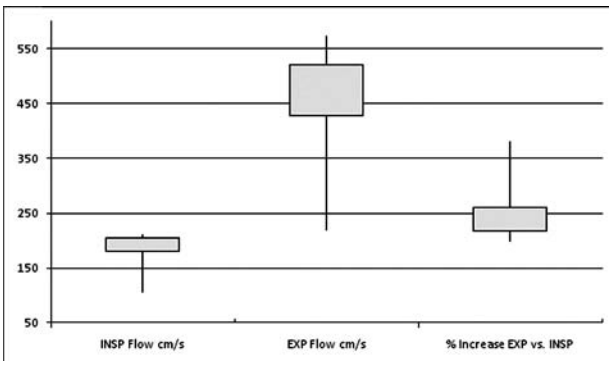

Fig 3. Distribution of (Doppler-) US values in patients.

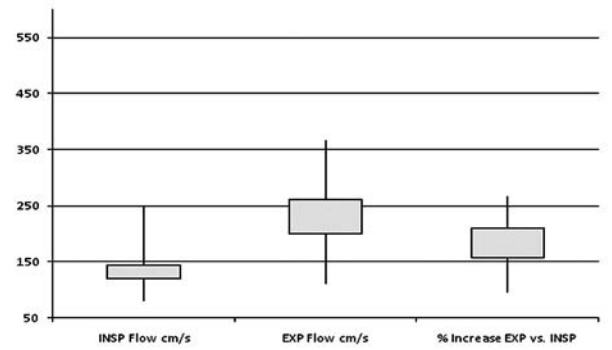

Fig 4. Distribution of (Doppler-) US values in volunteers.

Results

The 6 MALS-patients and 40% (8/20) of volunteers presented a DA+. The patients presented a mean inspiratory PV of 172 cm/s (± 40.9 cm/s), a mean expiratory PV of 425 cm/s (± 130.1 cm/s) with an amplitude of 249.1% ± 68.9 (fig 3). The volunteers presented a mean inspiratory PV of 126.9 cm/s (± 42 cm/s), a mean expiratory PV of 209.9 cm/s (± 80.1cm/s) with amplitude of 169.4% ± 54.3 (fig 4).

The statistical assessment defined a significant inverse correlations of the probands BMI and the PV measured (Kendall-τ; Spearman-ρ) between -0.33 (Kendall-τ) and -0.58 (Spearman-ρ) and no relevant correlation in PV-change (inspiratory vs. expiratory PV change) was found. The MALS-patients group was too small therefore the calculations could not be taken as reliable evidence.

The non-parametric Wilcoxon-Mann-Whitney test (MWU) showed significant differences between volunteers and MALS-patients for: the inspiratory PV ($p^{[2\text{-tailed}]}=0.023$; $p^{[1\text{-tailed}]}=0.012$; U=97 at $U^{critical}=27$) and the

Table I: Differentiability by (Doppler-)US-features defined by contingency testing concerning statistical independence of cross-tabulated categorical data including sensitivity, specificity, PPV and NPV at a prevalence of 23% [95%-Confidence Interval (CI): 9% .. 44%].

Discriminator **Calculation**	**Expiratory PV of the celiac trunk [> 350 cm/s]**	**DA [(+)]**	**Combination of DA and the according expiratory PV of the celiac trunk [> 350 cm/s and (+)]**
χ^2- test based Φ-coefficient calculation (1-sided;$\chi^{2critical}$=5.41)	χ^2= 15.95 at a Φ- coefficient of 0.78	χ^2= 6.69 at a Φ- coefficient of 0.51	χ^2= 20.63 at a Φ- coefficient of 0.89
Sensitivity	**83%**(95%-CI: 36% .. 100%)	**100%**	**83%**(95%-CI: 36% .. 100%)
Specificity	**95%**(95%-CI: 75% .. 100%)	**60%**(95%-CI: 36% .. 81%)	**100%**
PPV	**83%**(95%-CI: 36% .. 100%)	**43%**(95%-CI: 18% .. 71%)	**100%**
NPV	**95%**(95%-CI: 75% .. 100%)	**100%**	**95%**(95%-CI: 76% .. 100%)

PV- peak velocity, DA- deflection-angle, PPV- positive predictive value, NPV- negative predictive value

expiratory PV ($p^{[2\text{-tailed}]}$=0.001; $p^{[1\text{-tailed}]}$<0.001; U=110 at $U^{critical}$=27).

Contingency testing revealed no probability for the discrimination of pathologic (MALS) and healthy subjects via the above mentioned discriminators for inspiratory PV and amplitude change, but for expiratory PV of the celiac trunk (χ^2= 15,95 at a Φ- coefficient of 0,78; $\chi^{2\ critical}$ = 5,41), for a DA+ (χ^2= 6,69 at a Φ- coefficient of 0,51; $\chi^{2\ critical}$ = 5,41) and for the combination of DA+ and the according expiratory PV (χ^2= 20,63 at a Φ- coefficient of 0,89; $\chi^{2\ critical}$ = 5,41) (table I).

Discussions

To date, according to current literature more or less reliable, several features for the diagnosis of MALS are under debate. As mentioned in the introduction, however, some of these are vague and none of these can definitely diagnose MALS with reasonable probability and thereby select patient-cohorts with an immanent necessity for therapy.

While the true nature of MALS is not understood – and we were also not able to elicit it in our evaluation – we tried to define reliable features for diagnosis of MALS. Through the application of very restricted inclusion criteria we methodically excluded patients with the least evidence for diseases other than MALS.

Our data were able to confirm that ultrasound detects features highly associated with MALS, namely the flow changes of the celiac trunk and functional-geometric changes such as celiac trunk deflection during respiration and thus diaphragmatic excursion. In addition, the body mass dependence of inspiratory and expiratory PV in the celiac trunk could be verified: slim persons tend to have a significantly higher PV while the amplitude of PV change remains without significant association with the body mass.

Based on our empiric test-discriminators (thresholds) for inspiratory PV at 150 cm/sec, for expiratory PV at 350 cm/sec and for the amplitude of PV change at 210% and a positive DA of 50°, we were able to clearly differentiate volunteers from patients: the combination of the feature “expiratory PV higher than 350 cm/sec” (as a stand-alone feature with a PPV of 83% and a NPV of 95%) and the feature “positive DA” (as a stand-alone feature with a PPV of 43% and a NPV of 100%) ended in an exceptionally high sensitivity of 83% and a specificity of 100% (PPV 100%; NPV 95%) for our cohorts.

However, the pathophysiological entity MALS still remains a rather questionable syndrome also concerning the most appropriate choice of therapy. While this is beyond the scope of this study, we believe the mere temporary mechanical restriction of blood-flow in subjects with otherwise healthy vasculature does not explain oligemia in the well collateralized arterial vas-

cular system of the upper gastrointestinal tract. Many of our volunteers presented rather high arterial flow velocities and a pronounced DA of the celiac trunk. Thus, the actual mechanism of this disease might be some mechanically triggered, vegetative dysregulation of blood flow in the mesenteric system, possibly due to mechanical impairment at the region of the median arcuate ligament (celiac plexus). The verification or exclusion of this suspicion will at least be rather difficult or never achieved. For this reason we find functional US imaging maybe the best diagnostic option to define subjects with an acceptable high probability of suffering from MALS: the combination of a maximum expiratory PV of > 350 cm/s and a deflection angle higher than 50°.

A limitation of our study was the small number of patients who fulfilled the criteria for inclusion in this evaluation. These data and all thresholds might be debated as they were chosen empirically and arbitrarily and therefore must be subsequently validated in a randomized, prospective trial to define the overall effectiveness of such features for daily routine.

Conclusion

Based on these data we propose that functional ultrasound should be the first line in screening for MALS. However, attempts must be made to clearly define the true pathophysiologic findings behind MALS as – although our sonographic features and data proposed are noticeable – the underlying mechanisms are still unexplained by our data.

Conflict of interest: none

References

1 Dunbar JD, Molnar W, Beman FF, Marable SA. Compression of the celiac trunk and abdominal angina. Am J Roentgenol Radium Ther Nucl Med 1965; 95: 731-744.
2 Levin DC, Baltaxe HA. High incidence of celiac axis narrowing in asymptomatic individuals. Am J Roentgenol Radium Ther Nucl Med 1972; 116: 426-429.
3 Reilly LM, Ammar AD, Stoney RJ, Ehrenfeld WK. Late results following operative repair for celiac artery compression syndrome. J Vasc Surg 1985; 2: 79-91.
4 Horton KM, Talamini MA, Fishman EK. Median arcuate ligament syndrome: evaluation with CT angiography. Radiographics 2005; 25: 1177-1182.
5 Gloviczki P, Duncan AA. Treatment of celiac artery compression syndrome: does it really exist? Perspect Vasc Surg Endovasc Ther 2007; 19: 259-263.
6 De Cecchis L, Risaliti A, Anania G, et al. Dunbar's syndrome: clinical reality or physiopathologic hypothesis? Ann Ital Chir 1996; 67: 501-505.
7 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: 612-617.
8 Manghat NE, Mitchell G, Hay CS, Wells IP. The median arcuate ligament syndrome revisited by CT angiography and the use of ECG gating--a single centre case series and literature review. Br J Radiol 2008; 81: 735-742.
9 Sproat IA, Pozniak MA, Kennell TW. US case of the day. Median arcuate ligament syndrome (celiac artery compression syndrome). Radiographics 1993; 13: 1400-1402.
10 Erden A, Yurdakul M, Cumhur T. Marked increase in flow velocities during deep expiration: A duplex Doppler sign of celiac artery compression syndrome. Cardiovasc Intervent Radiol 1999; 22: 331-332.
11 Wolfman D, Bluth EI, Sossaman J. Median arcuate ligament syndrome. J Ultrasound Med 2003; 22: 1377-1380.
12 Williams JR. The Declaration of Helsinki and public health. Bull World Health Organ 2008; 86: 650-652.