Laparoscopic therapy of the coeliac artery compression syndrome: a critical analysis of the current standard procedure

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ABSTRACT

INTRODUCTION Median arcuate ligament syndrome has been known anatomically for approximately 100 years and results from a compression of the coeliac axis by fibrous attachment of the diaphragmatic crura. Owing to the rarity of the disease and limited available data, many aspects of treatment are controversial. Currently, laparoscopic decompression is considered by several authors as standard surgical procedure. We present an analysis of the clinical routine of MALS therapy.

METHODS We conducted a prospective observational trial in patients with MALS between March 2016 and August 2018, in which clinical symptoms, diagnostic evaluation, procedures with complication analysis and follow-up data were recorded.

RESULTS A total of 18 patients (12 female, 6 male) with MALS, aged between 15 and 65 years, were included in this study. All patients presented with long-standing abdominal pain. Preoperative Doppler ultrasonography showed a flow velocity of the coeliac artery averaging 289.9cm/second in mid-position of the diaphragm, 285.9cm/second in expiration and 199.0cm/second in inspiration. All operated patients underwent laparoscopic decompression; two patients received an angiographic intervention. Postoperatively, a significant decrease of the flow velocity in mid-position of the diaphragm was detected ($P = 0.018$). At follow-up after 5.2 months, 50.0% of the patients were pain-free, 37.5% reported symptomatic relief and 12.5% showed evidence for a recurrence.

CONCLUSION MALS is challenging both diagnostically and therapeutically. Laparoscopy with release of the median arcuate ligament is an essential part of the therapy and can be confirmed by Doppler ultrasonography. Disease outcome is also influenced by several predictive factors.

KEYWORDS

Dunbar syndrome – Median arcuate ligament syndrome – Laparoscopic release – Chronic abdominal pain

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Introduction

Median arcuate ligament syndrome (MALS) is a rare medical condition which still poses great challenges for accurate diagnosis and therapy. Anatomically, MALS results from the external compression of the coeliac artery and the coeliac ganglion by the median arcuate ligament. The ensuing abdominal neurovascular chronic pain syndrome causes a significant reduction in quality of life for patients. It is known that in 10–25% of normal individuals, the median arcuate ligament passes the aorta at a lower level, thus compressing the coeliac artery,¹ but only in a small subset of patients is it pathological.

Anatomically, this abnormality was described more than 100 years ago by Benjamin Lipshutz.² The first clinical reports of a surgical treatment for MALS by decompression were published by Harjola in 1963 and two years later by Dunbar *et al.*^{3,4} In clinical use, MALS is thus often referred to as Dunbar syndrome. Typical symptoms of MALS are severe postprandial abdominal pain, nausea, vomiting, diarrhoea, fainting, tachycardia, blocked inspiration and weight loss. Moreover, epigastric bruits have been noted during clinical examination. The pain is typically localised in the epigastrium. Many of these symptoms, taken individually, are also seen in other, more frequently occurring

disorders, which complicates the diagnosis, even if taking MALS into account in the clinical routine. Nevertheless, the combination of these symptoms is highly suggestive of a MALS. Most patients with MALS consult numerous physicians and clinicians before the diagnosis of MALS is eventually made.

In our experience, it is critical to clearly differentiate an intrinsic vascular stenosis from an external compression of the coeliac trunk. Important in this respect is proving a correlation between the compression of the coeliac artery with an unfixed stenosis and the existing symptoms,⁵ since a significant coeliac stenosis without MALS is similarly demonstrated in other diseases.^{6,7}

Typical diagnostic options in MALS are colour Doppler ultrasonography, computed tomography, magnetic resonance imaging, angiography of the coeliac artery and endoscopy. Colour Doppler ultrasonography of the coeliac artery in inspiration and expiration is a particularly important diagnostic tool. Evidence of an increased blood flow velocity in the coeliac artery in comparison with aortic flow velocity in the mid- and expiratory positions of the diaphragm, as well as a flow velocity decline in inspiration are typical findings in MALS and exclude a fixed internal (sclerotic) stenosis.⁸ Therapeutic options, mostly published in single case studies or with small patient cohorts, involve open decompression, laparoscopic release of the median arcuate ligament, robotic-assisted techniques or several vascular procedures.⁹ Recent studies also address predictive factors and their influence on therapeutic outcome.^{10–12}

Materials and methods

This prospective observational trial included data analysis from 18 patients with MALS who were treated in a single centre from March 2016 to August 2018. Among them were two patients with a recurrent compression after previous external surgery. Patient demographics, preoperative symptoms, diagnostic, intra- and postoperative data, including complications, and follow-up data were collected. Published study protocols were analysed to allow a comparative analysis of different aspects of this rare disease. Data were evaluated using a Microsoft Excel spreadsheet.

All operations were performed by a single surgeon. Patients were positioned in a reversed Trendelenburg position. A pneumoperitoneum was induced above the navel using an insufflation needle and placement of the 10-mm optical trocar. Four 5mm working trocars were introduced parabolically in the epigastric angle. The left lobe of the liver was held in the cranial direction by a retractor. In the first operations, the small omentum was opened to reach the operative target region as typically described in the literature. In our own experience, however, we saw an optimisation of the procedure in dissecting the gastrocolic ligament and a cranial positioning of the stomach. After exposure of the right crus of the diaphragm, the abdominal aorta with coeliac trunk and MAL was exposed at its caudal end (Fig 1). The MAL,

including nerve fibres and lymphatics, was divided using an Ultracision® harmonic scalpel (Ethicon Endo-Surgery, Guaynabo, Puerto Rico) and the tissue was examined histologically (Fig 2). Finally, the coeliac trunk presented with strong pulsation (Fig 3). The retractor and trocars were removed under visual control.

The analysis was performed using Microsoft Excel. Statistical analysis software SPSS 24.0 was used. A *P*-value of less than 0.05 was considered statistically significant. We used the U-test and the Wilcoxon sign test for paired samples.

Results

The patients came from 11 different federal states of Germany, as well as from Switzerland and Finland. Two patients were treated for recurrence after previous surgery. The mean age of the patients was 39.2 years (standard deviation, SD, ± 15.1 ; median 42.5 years; range 15–65 years); the ratio of male to female was 1 : 2 (sex vs age, *P* = 0.102). Mean body mass index (BMI) was 21.6kg/m² (SD ± 3.6 kg/m²; sex vs BMI, *P* = 0.0180) and American Society of Anesthesiologists score ASA I was for 6 (33.3%) and ASA II for 12 (66.6%). Table 1 shows further demographic data for the 18 patients in the study period. Table 2 represents the preoperative symptom characteristic of MALS. All patients reported chronic abdominal pain; 66.7% of the patients reported postprandial pain, 16.7% nausea and vomiting and 55.6% weight loss. The mean weight loss was 12.2kg (SD ± 4.5 kg).

Imaging prior to treatment included colour Doppler ultrasonography for all patients, magnetic resonance imaging (MRI) for nine patients (50%), computed tomography (CT) in two patients (11.1%); two patients (11.1%) underwent angiography. In the mid-position of the diaphragm, the flow velocity in the coeliac trunk reached 180–446cm/second (mean 289.9cm/second ± 80.1 cm/second) and dropped to 65–436cm/second (mean 199.0cm/second ± 110.6 cm/second) in inspiration. In expiration, the flow velocity reached 148–508cm/second (mean 285.9cm/second ± 123.6 cm/s). Aortic flow velocity was 59–171cm/second (mean 106.8cm/second ± 30.6 cm/second). In 13 patients (72.2%), a nutcracker syndrome of the left renal vein was additionally diagnosed. Sixteen patients underwent laparoscopic MAL release; two patients did not consent to surgery and thus received endovascular intervention as part of the treatment algorithm for MAL. Mean operating time for laparoscopic surgery was 82.7 minutes (SD ± 33.8 minutes; range 46–175 minutes; median 66.5 minutes). Intraoperative bleeding occurred in two patients (12.5%) and four patients required conversion to open surgery.

Postoperatively, retrogastric abscess formation was observed in one patient (6.2%). Postoperatively, with respect to the typical vegetative MALS symptoms, 12.5% of the patients still complained of abdominal pain and 6.3% patients suffered from vomiting during their inpatient stay.

Despite the considerable residential distances of our patients, follow-up examinations were done for 50.0% of

the patients post-surgery. Mean follow-up duration was 5.25 months $\pm$ 3.6 months. Postoperatively measured values showed a reduction of the flow velocity of the coeliac

Figure 1 Exposition of the abdominal aorta with coeliac trunk and median arcuate ligament.

Figure 2 Division of the median arcuate ligament and resection of nerve fibres and lymphatics.

Figure 3 Final presentation of the coeliac trunk with strong pulsation.

artery to 126–278cm/second (mean 168.5cm/second $\pm$ 47.6cm/second) in a resting expiratory position, 126–291cm/second (mean 178.3cm/second $\pm$ 53.7cm/second) in expiration and 76–227cm/second (mean 150.6cm/second $\pm$ 41.5cm/second) in inspiration. A comparison of preoperative with follow-up values revealed a significant difference of the values in mid-position of the diaphragm ($P = 0.018$). The P -value in inspiration was 0.0833; the P -value in expiration was 0.285. Table 3 shows coeliac artery mean velocities.

Some 50.0% of the patients followed up were completely symptom free; in 50% of the operated patients, abdominal pain was still detectable at follow-up; 75.0% of the patients with postoperative pain reported a reduction on the pain scale compared with preoperative findings. Nausea and vomiting was present in 25.0% of the patients and abdominal bruits in 37.5%. In one patient, a recurrence of MALS was assumed after Doppler ultrasonography.

A subgroup analysis revealed that patients who still indicated abdominal pain in the follow-up showed a flow velocity of 142cm/second $\pm$ 17.9cm/second in the coeliac trunk in the mid-position of the diaphragm during Doppler ultrasonography. This value is lower than the flow velocity of 195cm/second $\pm$ 55.6cm/second in patients without abdominal pain. The difference was significant ($P = 0.029$). The preoperatively determined average flow velocity of 291.7cm/second in patients reporting abdominal pain at follow-up was higher compared with the preoperative average flow velocity of 259.7cm/second measured in the patient group without abdominal pain at follow-up.

Discussion

MALS poses a clinical challenge in diagnostics and treatment. A review in 2016 analysing the literature from 1995 to 2015 summarises that many aspects of diagnosis and therapy are still unclear.⁹ Most findings are based on case reports or publications with small numbers of patients. Frequently, very large time frames are needed to collect patient groups with MALS. Cienfuegos *et al*, for example, required a time period from 2001 to 2013 to reach 15 patients;¹¹ Khrucharoen *et al* took from 2010 to 2017 for the same number of patients. Our analysis included patients from March 2016 to August 2018.²⁰ Most patients were diagnosed in highly specialised ultrasound practices and thus came from 11 federal states of Germany, from Finland and from Switzerland. The rarity of MALS reports complicates any compilation of a randomised study with appropriate statistical power. In the literature, a dominance of women, with a ratio four to one, is documented; the ratio in our study was two to one. The age of our patients ranged from 15 to 65 years (mean 39.2 years $\pm$ 15.1 years). Furthermore, a slender physique was found.¹³ The mean BMI of our patient cohort was 21.6kg/m².

Typical symptoms of MALS are chronic abdominal pain, postprandial pain, nausea/vomiting and unintended weight loss. Our findings confirmed abdominal pain in 100.0% of the patients, followed by 66.7% with postprandial pain, 16.7% with nausea/vomiting and 55.6% of patients with

weight loss (weight loss averaged $12.2\text{kg} \pm 4.5\text{kg}$). The variety of rather unspecific symptoms complicates the diagnosis. It is therefore crucial to note the coincidence of pain with a variety of vegetative symptoms, mostly nausea, fainting and blocked inspiration, to raise the suspicion of median arcuate ligament syndrome. In the hand of an experienced sonographer, the quantitative and functional colour Doppler examination is the most valuable tool for establishing the diagnosis and for re-evaluating patients postoperatively.

In our experience, an exaggerated lordosis of the lumbar spine is the triggering mechanism for the development of a median arcuate ligament syndrome. This also explains why many patients with MALS suffer from other vascular compression syndromes, predominantly nutcracker syndrome. Individual observations clearly show

that the degree of the compression can be influenced by the actual body posture of the patient. A kyphotic posture of the lumbar spine tends to reduce the traction of the median arcuate ligament towards the coeliac trunk and thus reduces its compression. This is in accordance with repeated reports from patients, who constantly stress the effectiveness of a crouching embryonic position to relieve their pain in postprandial pain attacks. Traditionally, as summarised by Kim *et al*,⁹ a variety of pathophysiological mechanisms are considered and a multifactorial genesis for this intriguing disease is favoured.¹⁴ One of the most frequently published theories is a compression of the coeliac artery and coeliac ganglion by abnormally thickened ligament fibres, a higher aortic origin of the coeliac artery or a descensus of the diaphragm due to severe lumbar lordosis or accelerated growth.^{15,16}

Vascular steal of blood flow by larger collateral vessels is also considered to cause symptoms of MALS.¹⁵ The vegetative symptoms result in this case from the compression and overstimulation of the coeliac ganglion.¹⁵ Often, arteriosclerotic stenoses of the coeliac trunk remain undetected, since many patients show no clinical symptoms. This debate lacks a sound basis since it does not take into account fundamental pathophysiological and anatomical discrepancies between an internal obstruction of the coeliac artery, mostly by arteriosclerotic plaque, and an external compression of the vessel, which also pinches the nervous fibres surrounding the origin of the coeliac artery.^{6,7} From our point of view, the origin of all symptoms is not the reduction of blood supply to the liver, stomach, spleen and pancreas, but the mechanical irritation of the coeliac plexus. It is therefore mandatory for successful interventions not only to restore the lumen of the coeliac trunk but also to prepare the nervous tissue carefully to reduce scar tissue after long-standing compressions and to ensure that the aortic hiatus is enlarged sufficiently to avoid postoperative strangulation of the remaining coeliac plexus.

In addition to abdominal sonography, CT, MRI, angiography and endoscopy to exclude other diagnoses, colour-coded duplex sonography seems to be of particular importance in diagnosis. Evidence of high blood flow velocity in the coeliac artery in a resting respiratory position and in expiration, as well as a less increased velocity in inspiration are typical of MALS and exclude a fixed stenosis.⁸ Dunbar syndrome is characterised by flow velocities in the coeliac trunk that substantially exceed the flow velocity in the aorta. Other authors set a coeliac flow velocity above 200cm/second as a diagnostic criterion.¹⁴ In our experience, is it more reliable to evaluate the flow velocity in the coeliac trunk in relation to the velocity in the aorta. It is often observed that very low aortal flow velocities exist, in the range of 50–70cm/second. In these cases, a coeliac arterial flow of 180cm/second is clearly a substantial flow acceleration, which raises the suspicion of MALS.

If no change in flow velocity is observed, a classic fixed stenosis exists.⁸ In our patients, Doppler ultrasonography showed, on average, a flow velocity in the coeliac artery in

Table 1 Demographic data for 18 patients.

	Patients	
	(n)	(%)
Hypertension	2	11.1
Gastro-oesophageal reflux disease	1	5.6
Tobacco use	3	16.7
Narcotic use	3	16.7
Psychiatric history	4	22.2
Nutcracker syndrome	13	72.2

Table 2 Clinical presentation for 18 patients.

	Patients	
	(n)	(%)
Abdominal pain	18	100
Postprandial abdominal pain	12	66.7
Vomiting	3	16.7
Weight loss	10	55.6
Abdominal bruit	1	5.6

Table 3 Coeliac artery mean velocities.

Period	Preoperative (cm/second)		Postoperative (cm/second)		P-value
	(mean)	(SD)	(mean)	(SD)	
Mid-position of diaphragm	289.9	± 80.1	168.5	± 47.6	0.018
Peak expiration	285.9	± 123.6	178.3	± 53.7	0.285
Peak inspiration	199.0	± 110.6	150.6	± 41.5	0.833

the mid-position of the diaphragm of 289.9cm/second, in expiration of 285.9cm/second and in inspiration of 199.0 cm/second. This is consistent with the findings of Klimas *et al*, who reported flow velocities of 190–450cm/second.¹⁶ Duplex sonography performed by a specialist was found to be a critical diagnostic tool for diagnosing MALS in our patient cohort.

The data available in the literature provide no indication for a superior therapeutic method. In recent publications, laparoscopic median arcuate ligament release is increasingly regarded as current standard surgical management.⁹ Numerous surgical and vascular surgical procedures have been published, including decompression of the median arcuate ligament in an open procedure, as conventional laparoscopy or as a robotic-assisted laparoscopic technique.¹⁷ The open procedure can be conducted as sole decompression of the median arcuate ligament after laparotomy. In addition to procedures with decompression and reconstruction, various vascular reconstructive techniques have also been described. Grottemeyer *et al* published aortocoeliac vein interpositions, aortohepatic vein interpositions, resections of the coeliac artery and end-to-end anastomosis, patch plastic of the coeliac artery and transaortic removal of a stent of coeliac artery.¹⁸ The large number of conceivable surgical options points to the lack of knowledge with respect to an optimal therapy of this disease. Thus, it still remains important today to document and evaluate the therapeutic data of patients with MALS.

Considering the large number of therapeutic procedures, follow-up data are of particular importance. Treatment results vary considerably. Klimas *et al* published, similar to several other authors, a symptom-free status in 93.1% of patients after laparoscopy.¹⁶ In a smaller group of patients, Do *et al* reported 67% of symptom-free patients in the laparoscopic group and 50% in the robotic group.¹⁹ In another publication concerning the robotic-assisted technique, the results showed a freedom of symptoms in 46.1%, symptom relief in 15.4% and symptom recurrence in 38% of patients.²⁰ Grottemeyer *et al* reported a complete absence of symptoms in 73.3% of patients after vascular surgical therapy.¹⁸

Another critical issue for analysis is the difference between immediate postoperative outcome and later follow-up data. For example, results were published in which only one-third of patients showed symptomatic recurrence in the follow-up after 19 months.²⁰ The mean follow-up duration in our analysis was 5.25 months $\pm$ 3.6 months, after which 50.0% of the patients indicated no further abdominal pain.

Several studies have focused on factors influencing the outcome of MALS in addition to surgery. Brody *et al* published a predictive model that demonstrates a correlation of postoperative quality of life with age and baseline coeliac artery expiratory velocity.¹⁰ A correlation also exists between high age and high flow velocity of the coeliac artery, with a poor outcome in the Short Form Health-36 questionnaire. Cienfuegos *et al* saw a correlation between long-term outcome and severity of occlusion in their

analysis.¹¹ The higher the CT occlusion grade, the better the postoperative result. Skelly *et al* reported a connection between a poorer-quality life outcome and psychiatric comorbidity in patients with MALS.¹² All these data confirm the hypothesis that a large variety of factors have a significant impact on the outcome and suggest a re-evaluation of the importance of the surgical procedure.

According to these studies, a median arcuate ligament release is nevertheless essential, but still not a guarantee for an excellent outcome. After median arcuate ligament release, a decompression of the coeliac artery is measurable via the flow velocity, but the extent to which the compression of the coeliac ganglion has been reduced cannot be determined. In the follow-up analysis of our subgroups, the average flow velocity in resting expiratory position of 142cm/second after median arcuate ligament release in the patient group with abdominal pain was lower than the flow velocity of 195cm/second in the group without pain ($P = 0.029$). The correlation of a worse outcome with higher preoperative flow velocity, as previously published by Brody *et al*, was also confirmed by our data.¹⁰

Conclusions

Laparoscopy, either classical or robotic-assisted, offers all the advantages of minimally invasive surgery for patients with MALS. The final surgical aim is, however, always a median arcuate ligament release and decompression of the coeliac nervous plexus. According to our data, the outcome after surgical therapy is considerably improved, but the many further factors contributing to this disease are not well understood and are extremely difficult to assess.

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