

Median Arcuate Ligament Syndrome is Effectively Relieved with Minimally Invasive Surgery

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ABSTRACT

Objective: Median arcuate ligament (MAL) syndrome is a constellation of symptoms related to compression of the celiac artery trunk. Minimally invasive release of the ligament has been shown to improve these symptoms. This study describes one institution's experience with this procedure and reports on outcomes of minimally invasive release and patient quality of life.

Methods: We performed a retrospective chart review of all patients who underwent minimally invasive release of the MAL at our institution. Patients were mailed a survey consisting of the 36-Item Short Form and Visick questionnaires. If surveys were not returned after one month, patients were called and asked to complete them over the phone. Demographic and pre- and postoperative data were collected and analyzed.

Results: Eleven patients underwent a laparoscopic MAL release from January 1, 2015 to January 31, 2020. Most patients, 73%, reported epigastric pain as their primary symptom for a median of 18 months. All cases were successfully completed laparoscopically, with only one intraoperative complication. Mean hospital length of stay was 1.4 d. At the time of survey completion, the mean weight change was 2.3 kg. Additional interventions for resolution

of symptoms and celiac artery stenosis were required for two patients. Surveys were completed by eight patients. A mean Visick score of 1.8 showed resolution or improved symptoms for all patients. SF-36 scores were highest for physical functioning, emotional well-being, and social functioning health areas.

Conclusions: Minimally invasive release of the MAL is a safe and effective surgery for patients suffering from MALS. Symptoms improved after adequate release of the ligament, with minimal morbidity and additional postoperative procedures needed.

Key Words: Celiac artery decompression, Laparoscopic surgery, Median arcuate ligament release, Median arcuate ligament syndrome, Minimally invasive surgery.

INTRODUCTION

Median arcuate ligament syndrome (MALS), also known as celiac artery compression syndrome or celiac axis syndrome, is a rare condition that presents with chronic, recurrent abdominal pain. The pain is usually postprandial and may be accompanied by nausea, vomiting, weight loss, and occasionally an abdominal bruit.¹ These symptoms are due to the extrinsic compression of the celiac trunk and/or celiac ganglion by a fibrous band, the MAL, traversing the crura of the diaphragm.² It is thought that a higher take off of the origin of the celiac trunk or a lower diaphragmatic insertion puts some individuals more at risk of developing this condition, due to the increased likelihood of compression by the MAL in these situations.²

The pathophysiology of this syndrome is not well understood but is believed to be due to compression of the celiac artery leading to vascular insufficiency, compression of the celiac ganglion, adding a neuropathic pain component, or a combination of both.^{3,4} In an estimated 10% – 24% of individuals, the MAL crosses anterior to the celiac artery; however, not all patients experience stenosis, compression, or develop symptoms.⁵ Thus, this syndrome remains a diagnosis of exclusion.

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Patients often see multiple specialists, including gastroenterologists and vascular surgeons in addition to general surgeons. Patients often undergo a variety of tests and imaging studies before a diagnosis is made, and the order of the tests may depend on the order in which they encounter the various physicians. These can include an abdominal computed tomography (CT) scan, upper endoscopy, duplex abdominal ultrasonography (US), CT angiography (CTA), or traditional angiography.^{2,5,6} Treatment for symptomatic patients involves surgical division of the MAL and associated fibers, with possible celiac ganglionectomy, to decompress the celiac artery. In addition, celiac artery bypass or endovascular stenting may be performed. Traditionally, this operation was performed through an open laparotomy approach; however, in recent years, minimally invasive laparoscopic or robot-assisted techniques have become popular.⁷⁻¹⁰ In this study, we report our institution's experience with laparoscopic release for MALS in 11 patients, including intermediate-term outcomes and quality of life survey information.

METHODS

After approval was obtained from the Institutional Review Board, all patients diagnosed with MALS between January 1, 2015 and January 31, 2020 and seeking treatment were identified. A retrospective review of medical records was conducted for all patients undergoing treatment for MALS with a minimally invasive release of the ligament performed by a senior minimally invasive surgeon. Information, including demographics, pre-operative symptoms and workup, operative details, and postoperative information, was collected, recorded, and maintained using a Microsoft Excel (Microsoft, Inc., Seattle, WA.) database. All identified patients were mailed letters notifying them of and explaining the study, along with a survey to complete. Completion and return of the survey constituted informed consent from the patient. Data analysis was performed using Excel.

Demographic information collected included age, gender, body mass index (BMI; calculated as kg/m²), past medical and surgical history, and current medications. Pre-operative symptoms of interest were nausea, vomiting, pain, anorexia or early satiety, weight loss, and length of time any symptoms were present. Imaging findings were recorded for abdominal CT/CTA, magnetic resonance angiography (MRA), duplex US, and arterial angiography. Diagnosis was made in consultation with a gastroenterologist, vascular surgeon, and advanced minimally invasive

surgeon based on history, symptoms, and imaging findings consistent with MALS. Operative details included length of operation, estimated blood loss (EBL), and length of stay postprocedure. Continued symptoms, weight, and any additional studies obtained, or surgery performed after the index operation was also collected.

A survey was mailed to each patient in 2021, consisting of the 36-Item Short Form Survey (SF-36)¹¹ and the Visick score. The follow-up period was \geq one year for all patients and six years for the first patient. If patients did not return a completed survey within one month, they were called by a team member and asked to complete the survey over the phone. Two phone calls with messages left were attempted to reach each patient.

Measures

The self-reported survey consisted of the Visick score and SF-36.

The Visick score is a subjective score that looks at symptom severity after surgery, helping to evaluate the effectiveness of the procedure.¹² It consists of four options given a numerical value of 1 to 4: no symptoms/resolved, improved, unchanged, or worsened. A lower Visick score corresponds to better symptom outcomes after surgery. It has been previously used to evaluate outcomes of MALS surgery.^{12,13} If a patient failed to return a completed survey and was unable to be contacted by phone, a Visick score was calculated from the clinical notes taken during the last follow-up appointment. If inadequate data was available to calculate a score, the patient was excluded from the study.

The SF-36 is a 36-item questionnaire, developed at RAND as part of the Medical Outcomes Study, that measures quality-of-life outcomes in healthcare¹¹ and has previously been used to evaluate patient reported outcomes following surgery.^{14,15} Questions relate to eight health areas including physical functioning, bodily pain, role limitation due to physical health problems, role limitation due to personal or emotional problems, emotional well-being, social functioning, energy/fatigue, and general health perceptions. Results of the health survey were scored using the scoring key available online, with higher scores representing a more favorable health state.¹⁶

Surgical Procedure

All surgical procedures for MAL release were performed by one senior minimally invasive surgeon at our institution.

The same surgical technique was used for each participant. Briefly, the patient is placed in the low lithotomy position. Five-millimeter trocars are placed in the supraumbilical, left lateral subcostal, and right and left epigastric positions for the camera and working ports, as well as the subxyphoid position to be used for a liver retractor (**Figure 1**). Dissection is begun at the lesser curvature along the left gastric artery with a Harmonic scalpel. Using blunt dissection and the Harmonic scalpel, the common hepatic artery is exposed and dissected free towards the celiac trunk and down to the level of the aorta until fully visible. The ligament compressing the celiac trunk and any celiac nerves present are divided allowing the celiac trunk to be freed circumferentially. This is performed using a combination of the Harmonic scalpel and electrocautery. Full release is verified visually; no intraoperative US is performed. The dissection is considered complete when the gastric, hepatic, and splenic arteries are skeletonized for a distance of 2 cm, and the junction of the celiac trunk and the aorta has been exposed. There is a typically a visible change in caliber of the vessels as they are released, obviating a need for intraoperative US. The dissection also necessarily destroys the

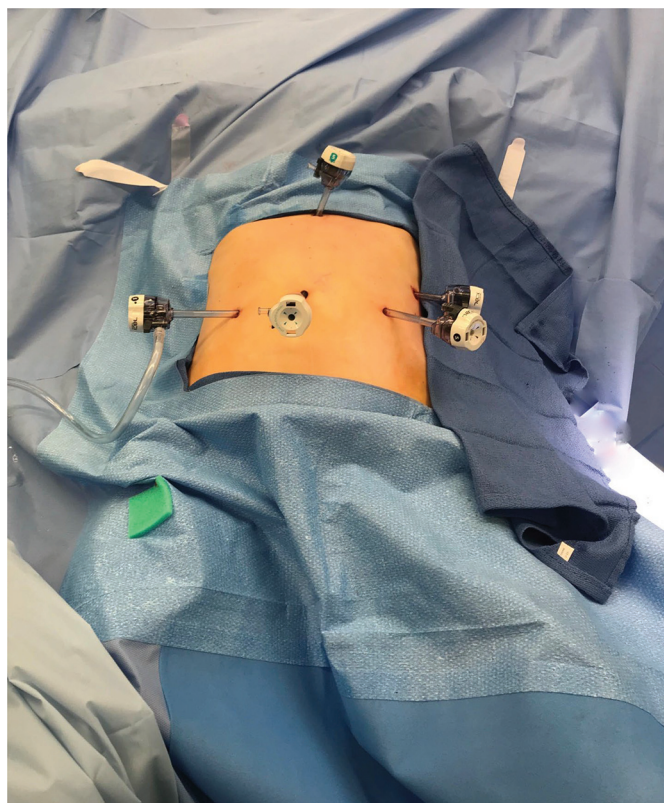


Figure 1. Placement of the camera, retractor, and working ports for a laparoscopic median arcuate ligament release.

connective tissue elements and neural fibers immediately adjacent to the celiac artery. While this is not a formal ganglionectomy, it does disrupt neural pathways in immediate proximity to the celiac axis. Patients are discharged from the hospital when their pain is well controlled and able to tolerate an oral diet.

RESULTS

A total of 11 patients underwent a laparoscopic release of the median arcuate ligament from January 1, 2015 to January 31, 2020. **Table 1** summarizes the baseline patient demographics and characteristics. There were eight female and three male patients, with a mean age of 42 years and median of 47 years (range 15 to 62 years). Mean BMI was 20.4 (range 15.1 to 26.5) pre-operatively, and the median was 20.1. Epigastric pain was the most reported symptom (73%), followed by postprandial pain, nausea, and weight loss, each reported by 64% of patients. Antiemetics were taken by five patients and six were on prescribed pain medications. Median length of time symptoms were present was 18 months (range 3 to 180 months). Pre-operative imaging included esophagogastroduodenoscopy (EGD), CT/CTA, MRA, mesenteric duplex US, and arterial angiography. Six patients underwent a CT or CTA, with all but one showing stenosis or narrowing of the celiac artery. Similar findings of stenosis were seen in all three MRAs obtained. An arterial angiogram was performed in three patients, all of which had findings consistent with MALS. Mean (\pm standard deviation [SD]) pre-operative celiac artery peak velocity was 376 (\pm 138) cm/s for the nine patients for which it was reported although one patient did not have a reported peak systolic velocity (PSV). One patient underwent a celiac nerve plexus block about a week before surgery, reporting a moderate reduction in their pain symptoms.

All 11 patients underwent a laparoscopic procedure without conversion to open. Nine cases reported total operative time and EBL; for the remaining two, both done in 2015, this information was unavailable. One case had an intraoperative complication. An arteriotomy was made on the side of the splenic artery and a large branch avulsed during dissection around the aorta. Both injuries were controlled with 5-millimeter clips, resulting in cessation of bleeding. Total EBL for this case was 750 ml. Mean EBL, excluding the 750 ml, was 150.6 ml.

Excluding the case with an intraoperative complication (329 minutes), the mean (\pm SD) operative time was 165 (\pm 27.9) minutes (range 127 to 212 minutes). All patients

Table 1.
Baseline Patient Demographics and Characteristics

Patient Number	Age (years)/sex	Body Mass Index	Pre-operative symptoms	Symptom duration (months)	Pre-operative Studies	Pre-operative celiac artery PSV, cm/s
1	62/F	15.8	Epigastric pain, anorexia, nausea, diarrhea, weight loss	36	EGD, MRA, Angiogram, Duplex US	303
2	21/M	20.1	Epigastric pain, postprandial pain, anorexia	36	Duplex US	488
3	52/F	20.2	Postprandial pain, weight loss	3	Duplex US	461
4	31/F	25.7	Epigastric pain, diarrhea	180	EGD, CT, Duplex US	421
5	57/M	26.5	Epigastric pain, early satiety, nausea, weight loss	120	CTA, Duplex US	568
6	15/F	15.1	Epigastric pain, early satiety, nausea	60	EGD, CT, Duplex US	342
7	61/F	18.8	Epigastric pain, postprandial pain, nausea, weight loss	4	EGD, MRA, Angiogram, Duplex US	143
8	60/F	24.0	Epigastric pain, postprandial pain, early satiety, weight loss	6	EGD, CTA, Angiogram, Duplex US	177
9	29/F	15.8	Postprandial pain, early satiety, nausea, emesis, weight loss	9	EGD, CTA, Duplex US	484
10	47/M	19.9	Epigastric pain, postprandial pain, anorexia, nausea, emesis, weight loss	18	EGD, MRA, Duplex US	Not provided in report
11	23/F	22.3	Postprandial pain, nausea	18	CTA, MRI	Not done

Abbreviations: PSV, peak systolic velocity; EGD, esophagogastroduodenoscopy; MRA, magnetic resonance angiography; US, ultrasound; CT, computed tomography; CTA, computed tomography angiography; MRI, magnetic resonance imaging.

were discharged between one and three days (mean of 1.4 days). There was one readmission at one month for abdominal pain and nausea. Imaging obtained was normal and symptoms were found to have a somatization component. Another patient returned on postoperative day five due to constipation and mild abdominal pain, however, did not require admission.

Postoperative angiograms were performed in two patients, both for suspected recurrence with planned intervention. The first required an angioplasty and stent placement two months after surgery due to recurrent abdominal pain and stenosis on imaging (**Figures 2A and 2B**). Five months later another balloon angioplasty was performed due to recurrent symptoms; they subsequently underwent a renal vein transposition due to Nutcracker syndrome and are currently pain free. In another

patient, balloon angioplasty was performed two years post-procedure for stenosis and progression of plaque in the celiac artery. Duplex US was obtained for five patients postoperatively. Mean (\pm SD) PSV decreased to 222.8 (\pm 64.6) cm/s, however, was not statistically different from the pre-operative value.

Survey responses were provided by eight patients. Current postoperative weight was reported on the survey or retrieved from the last clinic visit if no survey was completed. The mean weight change was 2.3 kg (range -7.2 to + 12.4 kgs), with a mean postoperative BMI of 21.3 (range 15.4 to 28.1). No statistically significant change in BMI was noted ($P = .12$). The mean Visick score was 1.8 (range 1 – 3), with no patients excluded from the score calculation (**Figure 3**). Eighty-two percent of patients reported either no symptoms or improved symptoms

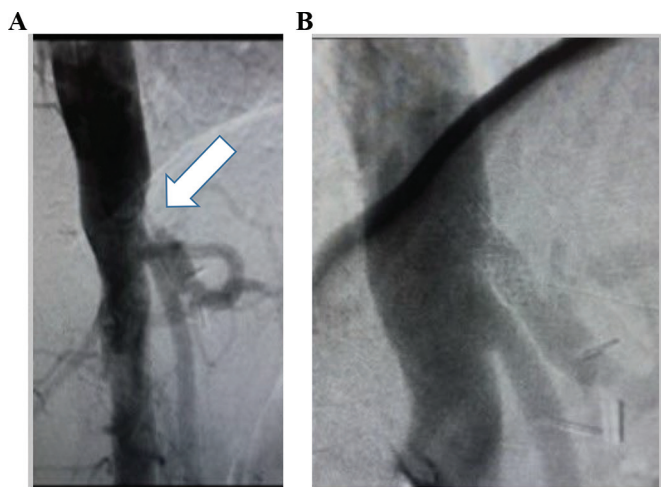


Figure 2. (A) Arterial angiogram, post-laparoscopic median arcuate ligament release, showing celiac artery stenosis (arrow). (B) Arterial angiogram with resolution of stenosis after celiac artery angioplasty and stent placement.

following the procedure. The SF-36 health area scaled scores for each patient, as well as overall means are provided in **Table 2**. The scores for physical functioning, emotional well-being, and social functioning were the highest reported by the patients, suggesting better function and lower disability in these areas. Energy/fatigue and general health had the lowest reported scaled scores. Only one area, general health, had a mean score under 50. Six of the areas had mean scores over 65. When comparing their health to a year ago, three patients reported much better health, four patients reported somewhat

better health, and one patient reported their health was about the same.

DISCUSSION

This single-institution experience with 11 patients who underwent laparoscopic MAL release demonstrates this is a safe and effective treatment option for patients with symptomatic MALS. No cases needed conversion to an open procedure and only one intraoperative complication occurred. One patient required readmission, giving a low readmission rate (< 10%). At the time of the survey, most patients reported no or improved symptoms indicating a successful outcome of the procedure.

In 2000, one of the first reports of a minimally invasive technique for treatment of MALS was described.¹⁷ Since then, this technique has gained in popularity with additional small case series and studies reported.^{7-9,14,18,19,24} More recently, robot-assisted MAL release has been characterized in the literature.^{10,20,21} Both of these operative techniques provide the benefits seen with other minimally invasive procedures to MALS patients, including less post-operative pain, shorter hospital stay, and quicker recovery time. These benefits were also reported in this study's results. For example, the average hospital length of stay of 1.4 days was consistent with, and often shorter, than what has previously been reported.^{7,9}

The demographics of this study were consistent with prior studies and reports^{2,7,9,20,23} on MALS with 73% of patients being female and a mean age of 41.6 years. Three patients

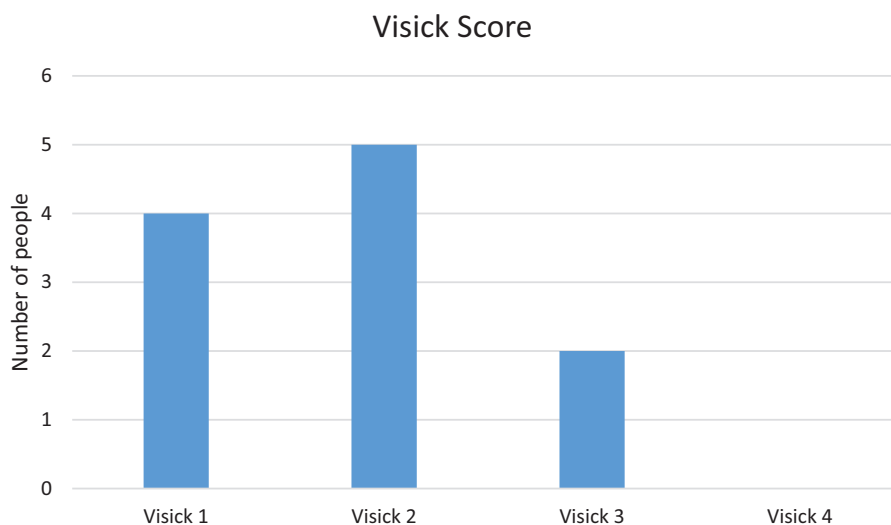


Figure 3. Visick score for eleven study patients.

Table 2.
Postoperative SF-36 Health Area Scaled Scores Listed by Participant and Mean Score

Patient Number	Physical functioning	Role limitations due to physical health	Role limitations due to personal or emotional problems	Energy/fatigue	Emotional well-being	Social functioning	Pain	General health
1	100	75	100	60	88	75	67.5	70
2	75	100	100	80	92	87.5	55	25
3	100	75	100	60	100	100	90	20
4	100	100	100	80	88	100	90	95
5	95	0	33.3	15	72	62.5	67.5	40
8	10	0	0	0	44	37.5	22.5	5
10	75	100	100	80	92	100	77.5	75
11	95	100	0	40	44	37.5	77.5	50
Mean	81.2	68.7	66.7	51.9	77.5	75.0	68.4	47.5

were considered underweight (< 18.5) per their calculated BMI and one patient was just inside the normal range with a BMI of 18.8. Two of those patients reported weight loss as a symptom and all three reported either early satiety or anorexia, likely contributing to their low pre-operative BMIs. MALS is commonly seen in people with a thinner body habitus and those more resistant to weight gain.^{2,19} In these patients, the closer proximity of the celiac trunk and ganglion to the MAL can increase the likelihood that compression of these structures will lead to symptoms.

As MALS is a diagnosis of exclusion, multiple imaging modalities are usually obtained. In our experience, over 80% of patients received at least two different pre-operative studies. Celiac artery peak systolic velocity is obtained from a duplex US and is considered elevated with values > 200 cm/s.¹ By that criteria, 7 out of 9 of the patients in this study were positive. Several patients had angiography rather than CTA or MRA. It is our preference to rely on cross-sectional imaging; however, given the variation in practice patterns of referring physicians, the authors did not have control of the workup for most patients.

Patient selection is critical in this condition. A thoughtful gastroenterologist and a high index of suspicion is important in the early workup. US criteria are a useful guide and unintentional weight loss may be the most important single determinant. A celiac plexus block is a method sometimes used to select patients that could benefit from a MAL release.^{2,19} Temporary resolution of symptoms suggests a high chance of success following the procedure. One of our patients underwent this procedure and did receive temporary pre-operative symptom relief. Postprocedure, they had

an increased BMI and reported improved symptoms overall. Including a celiac plexus block could be a good option, especially for patients who are unsure about undergoing a MAL release.

Over 70% of patients responded to the survey for this current study. The mean Visick score was low indicating that most patients reported no or improved symptoms following this procedure. There was a range of responses for the SF-36 questionnaire, with some participants reporting better health in some of the areas than others. This could be due to a variety of factors. Some patients reported having difficulty with other medical conditions at the time of the survey which could have biased the responses. Additionally, five of our patients had psychiatric conditions noted in their medical history or were taking prescribed medications for these conditions, also noted in other MALS studies.^{6,22} This could influence responses to the survey, especially for questions relating to energy/fatigue, emotional well-being, and general health areas. However, even with the varied responses on the survey, overall means for seven out of the eight health areas were still over 50, indicating better outcomes than before surgery. This, combined with the fact that only two patients needed additional vascular interventions postoperatively, suggests overall positive outcomes for patients and successful treatment for MALS.

There were a few limitations to this study. First, it was limited by the small sample size. MALS is a relatively rare condition making it difficult to have large numbers of patients. It is possible that some of the pre- and

postoperative datapoints examined, for example BMI and PSV, may have reached statistical significance if the number of patients had been larger. Additionally, Visick and SF-36 questions were only asked in the post-operative setting. Having pre-operative scores to compare to would have been ideal and could have provided additional information. Although these questionnaires have been used often to provide information about surgery outcomes, and in MAL release specifically, recall bias by patients is possible. Future studies should include a multicenter approach to help increase the sample size and pre- and postoperative questionnaires with longer-term follow-up to see if the positive effects of this procedure continue.

CONCLUSION

Laparoscopic release of the median arcuate ligament is a safe and effective technique for treating MALS. Patients benefit by an improvement in or resolution of their symptoms with relatively low morbidity associated with this procedure.

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