Predictors of Surgical Outcome in Patients with Median Arcuate Ligament Syndrome

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Abstract

Background: The median arcuate ligament is a normal anatomical feature of the aortic hiatus, attached to the posteromedia aspect of the diaphragm. Median arcuate ligament syndrome (MALS), also known as celiac artery compression syndrome (CACS), is a presumed rare congenital defect thought to cause compression of the celiac trunk, in addition to possible impingement of the celiac plexus. Symptomatic patients, depending on age and stage of disease complain of post-prandial abdominal pain, post-exertional abdominal pain, nausea, vomiting, and weight loss. Due to rarity of the disease in conjunction with vague physical complaints and unclear etiology, a universal method for surgical screening has not been developed and predictors of surgical outcome are not widely understood. This systematic review addresses how providers can determine surgical candidacy based upon outcome predictors and symptomatology.

Methods: A search was conducted using Google Scholar, MEDLINE via PubMed, ScienceDirect, and CINAHL via Ebsco. Keywords included were median arcuate ligament syndrome, celiac artery compression syndrome, predict*, and outcomes. Studies were assessed for quality using GRADE criteria.

Results: This systematic review includes 3 studies that discuss predictive models for screening MALS surgical candidates. One study evaluated the surgical outcomes of MAL decompression in 42 patients using SF-36 to evaluate symptom improvement and resolution, and it was deduced that age and baseline celiac artery expiratory velocity served as predictors of positive surgical outcome. Another study evaluated 67 patients instead using SF-12 and the Visick score as symptom questionnaires. This study found that pre-operative post-exertional abdominal pain suggested a positive surgical outcome, and vomiting and unprovoked pain were associated with negative surgical outcomes. A prospective study was lastly analyzed. This study, which included 135 patients, determined that the development and grade at which collateral circulation was formed within the abdomen correlated with increasingly negative surgical outcomes.

Conclusion: Surgical decompression of the celiac artery is often successful, despite pre-operational predictors that might suggest incomplete symptom resolution. Predictors of positive surgical outcomes include post-exertional pain, low age of diagnosis, and reduced development of collateral circulation. Predictors of negative surgical outcome include vomiting, unprovoked pain, and increased formation of collateral circulation.

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Keywords
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Faculty Advisors: Annjanette Sommers, PA-C, MS and  
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Biography

[redacted]
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**Conclusion:** Surgical decompression of the celiac artery is often successful, despite pre-operational predictors that might suggest incomplete symptom resolution. Predictors of positive surgical outcomes include post-exertional pain, low age of diagnosis, and reduced development of collateral circulation. Predictors of negative surgical outcome include vomiting, unprovoked pain, and increased formation of collateral circulation.

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**Acknowledgements**
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List of Abbreviations

CA Celiac Artery
CACS Celiac Artery Compression Syndrome
MAL Median Arcuate Ligament
MALS Median Arcuate Ligament Syndrome

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Visick Score
Gastrointestinal Symptom Rating Scale
12-Item Short Form Health Survey (SF-12)
SF-36 Questionnaire
Predictors of Surgical Outcome in Patients with Median Arcuate Ligament Syndrome

BACKGROUND

Median arcuate ligament syndrome (MALS), also known as celiac artery compression syndrome (CACS) is a set of symptoms stemming from a rare congenital malformation of the median arcuate ligament of the diaphragm or high emergence of the celiac artery from the aorta. Abnormal insertion of the median arcuate ligament in relation to the celiac artery causes vascular compression and in some cases compression of the celiac plexus. The celiac artery supplies blood to the upper abdominal organs, and compression often results in ischemia which may be dependent on respiration, exertion, and/or eating.¹

Patients present with numerous symptoms, often varying and dependent on age, stage of disease, and whether the celiac plexus is also occluded. Universally, patients present with chronic, recurrent, severe abdominal pain. Varying symptoms include postprandial abdominal pain, post-exertional abdominal pain, mid-epigastric bruit, nausea, vomiting, weight loss, and anorexia. These vague symptoms are often complicated by pre-existing conditions.¹ Studies have shown correlation with pre-existing psychiatric disorders,² cardiovascular disease,³ and presence of collateral mesenteric arterial circulation.⁴

While controversial, the favored treatment in recent years has been laparoscopic decompression of the celiac artery by release of the arcuate ligament. Arterial bypasses, endovascular angioplasty, and stenting have been used in conjunction with release of the ligament in cases where revascularization is necessary and/or for relief of persistent symptoms.¹ Sympathectomy of the celiac plexus is also
occasionally done for patients who show improvement in their symptoms with a celiac ganglion nerve block.\textsuperscript{5}

Often, most or all symptoms are relieved after surgical release of the median arcuate ligament and decompression of the celiac artery. In many cases though, patients’ symptoms continue or recur months to years later. Additionally, the revascularization and surgical procedure occasionally lead to complications such as gastroparesis, pancreatitis,\textsuperscript{6} and abdominal aneurysms resulting in hospital readmission. This review aims to correlate patients’ presenting symptoms with their expected anatomical variant to determine surgical candidacy and better predict surgical outcome.

**METHODS**

An exhaustive literature search was conducted using Google Scholar, MEDLINE via PubMed, ScienceDirect, and CINAHL via Ebsco. Keywords included were median arcuate ligament syndrome, celiac artery compression syndrome, predict and outcome. The bibliographies of relevant studies were used to find additional sources. Our search was limited to studies on humans, written in English and published in the last 20 years. Included were studies with patients who had a confirmed diagnosis of MALS/CACS, and considered surgical intervention in the treatment of the condition. Case studies were used for background information. Exclusion criteria also included involvement of the superior mesenteric artery, or if the focus of the study was on diagnostic procedures. We assessed the quality of each article using the Grading of Recommendations, Assessment, Development, and Evaluation (GRADE).\textsuperscript{7}
RESULTS

The search initially yielded 708 articles: 628 on Google Scholar, 12 on CINAHL via Ebsco, 24 on MEDLINE via PubMed, and 44 on ScienceDirect. The articles were reviewed for relevance, duplicates, inclusion and exclusion criteria, resulting in 3 studies for this systematic review.

Ho et al

This study is a retrospective records review of patients identified as having MALS between 1998 and 2013 from Princess Alexandra Hospital, the Royal Brisbane and Women’s Hospital, and Queensland Vascular Diagnostics. Patients were contacted via mail and telephone to assess their outcomes, which were measured subjectively by Visick score, Gastrointestinal Symptom Rating Scale (GSRS), and 12-Item Short Form Health Survey (SF-12).

The study included 67 patients diagnosed with MALS: 43 were treated surgically and 24 were managed nonoperatively. Pre-treatment characteristics, including clinical features, past medical history, investigations performed, and imaging results were defined and analyzed for each group. There were 38 (88%) patients who underwent laparoscopic decompression, 1 (3%) patient who was converted to an open operation, 5 (12%) patients who underwent open decompression, and 2 patients who had additional measures taken for revascularization following the initial surgery due to persistent symptoms. Pre- and post-treatment Visick scores, GSRS and SF-12 scores were compared. Median follow-up was 25 months, at which time 93% of patients who were treated surgically had improvement in their symptoms, including 37% who were asymptomatic. An additional 5 (26%) patients reported being asymptomatic at later follow-up. One patient required an open
aortoceliac bypass due to recurrence of symptoms 9 months after decompression. Residual celiac artery stenosis was present in 5 (26%) patients. “No clinical features were associated with significant differences in GSRS scores or adjusted SF-12 physical or mental scores;” however, “several clinical features were associated with Visick scores.” Postexertional pain was associated with lower Visick scores (improved symptoms) and cure. Unprovoked abdominal pain and vomiting were associated with higher Visick scores and persistence of symptoms. “Age at presentation, gender, postprandial pain, weight loss >9kg, nausea, bloating, and presence of an epigastric bruit were not significantly associated with the level of the Visick score or being cured.”

Of the 24 patients who were treated nonsurgically, at a mean follow-up of 24 months, 8 patients (33%) had less severe symptoms including 1 patient who reported being asymptomatic. Twelve patients (50%) had no change in symptoms, and 4 (17%) had worsening symptoms. “Age at presentation, gender, postprandial abdominal pain, post-exertional abdominal pain, unprovoked abdominal pain, weight loss of >9kg, nausea, vomiting, diarrhea, and epigastric bruits were not associated with any outcome measures.” Non-operative treatment type was not associated with any outcome measures.

Brody et al

In this study, 42 MALS patients underwent surgical decompression of the median arcuate ligament. Of these patients, the mean age was 35.5, mean weight was 65.9 kg, and mean BMI was 23.4. 97.6% were female and 45.2% were taking narcotics pre-operatively. Improvement was measured in eight scales, using the global SF-36 scale: function, physical, bodily pain, general health,
vitality, social function, emotional, mental health, and global score. Though all 8 scales had significant improvements, the most affected score was bodily pain, with a 64% improvement. The SF-36 data was collected 28.5±18.8 months following surgical decompression. Poor postoperative outcomes included pancreatitis (1 patient), tachycardia and/or epigastric abdominal pain (4 patients), for which 3 patients were treated inpatient with supportive measures, while 1 patient required treatment for a pulmonary embolism. Gastroparesis with nausea and vomiting was noted in 4 patients. According to the study, “based on SF-36 scores, 31 (73.8%) patients had a good outcome, 6(14.3%) had a neutral outcome, and 5(11.9%) had a poor outcome.”

Pre- and post-operative celiac artery mean velocities were measured at rest and during inspiration and expiration. Surgical decompression significantly reduced the mean velocity in all patients in all three phases. Resting mean velocities were measured at 318.5±114.9 cm/s pre-op, to 256.5±116.5 cm/s post-op. Inspiratory values were 219.3±82.4 cm/s to 183.9±76.7 cm/s, and expiratory values were 333.4±118.4 cm/s to 265.4±137.8 cm/s pre- and post-operative, respectively. P-values were less than 0.05 for all data. Along with baseline CA expiratory velocity, age was determined as another predictor of good surgical outcome. Using these 2 variables together, a predictive model was produced to determine poor, neutral, and good outcomes, based on the SF-36. For example, a MALS patient of the age of <15 years could expect a poor outcome at a baseline CA expiratory value of ≥ 365 cm/s and a good outcome at ≤ 210 cm/s. For a MALS patient over the age of 55, a poor outcome is predicted with a baseline CA expiratory value of ≥ 733 cm/s and a good outcome with a value of ≤ 461 cm/s.
In this retrospective study,\textsuperscript{4} which evaluated 129 patients from 2002 to 2013 who underwent surgical decompression of the median arcuate ligament, patients were assessed by presence of developed collateral circulation. Collateral circulation was typed by grade 0, 1, or 2, with grade 0 as, “no visible collaterals or partial filling of the recipient artery on selective angiography,” grade 1 as, “at least one collateral clearly visible on selective but not on non-selective angiography,” and grade 2 as, “at least one collateral clearly visible on non-selective angiography.”\textsuperscript{4}

Of the 129 patients evaluated, 29 had grade 0 collaterals, 52 patients had grade 1 collaterals, and 48 patients had grade 2 collaterals. Anatomic and clinical outcomes were measured. There were 99/129 (77\%) of patients who had anatomic success postoperatively. Twenty-five patients underwent additional percutaneous endovascular angioplasty and 3 patients underwent open reintervention. After the additional treatment for those 28 patients, 120/129 (93\%) had anatomic success. There were 92 patients (71\%) who had improvement in their pain and were considered to have a successful clinical outcome. Sixteen patients (12\%) had a temporary improvement in their pain. Twenty-one patients (16\%) had no change in their pain, and 0 patients had worsening pain. The presence of extensive collaterals (grade 2), on preoperative mesenteric angiography showed a lessened positive correlation to postoperative clinical success in comparison with lower grades. “Clinical success rate was 81\% in patients with grade 0 collaterals (39 of 48 patients) and 89\% in patients with grade 1 collaterals (25 of 28 patients). In patients with grade 2 collaterals, the clinical success rate was 52\% (23 of 44 patients; P<.001).”\textsuperscript{4}
DISCUSSION

Median arcuate ligament syndrome, also known as celiac artery compression syndrome, has 2 main presentations with a spectrum of symptomatology. The earliest detected symptom is often post-exertional abdominal pain, which typically presents at a younger age. As a patient with MALS/CACS ages, other symptoms may become prominent. Patients with a longer history of this particular condition, whether undiagnosed or not, may experience physical symptoms such as postprandial abdominal pain and later unprovoked abdominal pain, weight loss, nausea, and vomiting. Occult pathology may be present, specifically extensive developed collateral circulation, arterial stenosis, and abdominal vessel aneurysms.

Upon diagnosis, the mainstay of treatment is surgical decompression of the celiac artery, which is completed by release of the median arcuate ligament. Currently, this procedure is most done laparoscopically, but for patients with special cases, surgeons may opt for an open approach. Surgeons may also consider stenting, bypass, sympathectomy or celiac ganglionectomy, or celiac plexus blocks as adjunct to surgical decompression.

Post-operative outcomes of surgical decompression of the celiac artery include complete resolution of symptoms, continued/recurrent symptoms, or complications of surgery such as pancreatitis, gastroparesis, aneurysms and death.

MALS/CACS presents differently based on solely vascular versus neurovascular involvement as well as age. According to Brody et al,⁶ “the longer duration of symptoms may correlate with more impingement of the CA and plexus and this may correlate with higher CA velocities. Ultimately, patients with longer duration of symptoms may not respond as readily or as well to a MAL release.” Weight loss,
gender, postprandial abdominal pain, while useful symptoms to establish the diagnosis of MALS, were not useful surgical outcome prediction. However, post-exertional abdominal pain was associated with a positive surgical outcome and vomiting, unprovoked abdominal pain, and presence of Grade 2 collateral circulation were associated with less surgical success.

Surgical management of MALS/CACS differs depending on presentation and symptoms. According to the Brody et al study, “the celiac plexus may play a significant role in visceral pain. Therefore, a meticulous division of the celiac plexus is a critical step along with the division of the MAL...” Currently, celiac ganglionectomy and sympathectomy are excluded as first line adjuncts to surgical decompression of the celiac artery in patients treated for MALS. Another option surgeons may choose to incorporate into surgical treatment may include stenting, in the case of atherosclerotic stenosis of the celiac artery and mesenteric arteries, in conjunction with MALS. These treatments are adjunct at the discretion of individual surgeons, given patient symptomatology.

Study limitations included risk of bias due to small community of surgeons and doctors who treat MALS/CACS patients and variability across studies. Worth considering is the observation of reported data not corresponding with supplied figures and numerical data in the Van Petersen et al study. When communication was attempted in regards to clarification of the data, correspondence via email was unfortunately left incomplete. The corresponding author was queried but has yet to respond.

Based on the studies evaluated during this analysis, some recommendations for further study include celiac plexus involvement versus vascular involvement and presentation differences between
either pathology, age of presentation comparison, and surgical decompression alone versus decompression with concurrent sympathectomy. Poor surgical outcomes associated with diaphragmatic injuries following surgical decompression may be a useful evaluation to include in further studies. Given more positive outcomes with lower age of diagnosis and treatment, further studies may have benefit to reducing morbidity in this population and may better unify guidelines for detecting a likely often underdiagnosed syndrome.

**CONCLUSION**

Surgical outcome can be predicted based off initial presentation and neurovascular involvement using a series of prognostic assessments and imaging, allowing clinicians to better inform patients of surgical expectations for this uncommonly diagnosed condition. Age of presentation and age at which surgical decompression is achieved act as the most influential predictive factors as each accounts for comorbidities and overall physical baseline of a MALS patient. As post-exertional pain often presents at an earlier stage than symptoms of vomiting, unprovoked abdominal pain, and Grade 2 collateral development, the benefit of further studies would be provision of more efficient guidelines for detecting and treating MALS at an earlier age. While surgical outcomes may differ with involvement of the celiac plexus versus solely vascular involvement, symptomatology, or development of collateral circulation, surgical decompression remains the most optimal treatment for MALS.
References


# GRADE

**Table 1: Quality Assessment of Reviewed Articles**

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<td>Limitations</td>
<td>Indirectness</td>
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The Ho et al study\(^2\) has an RR of 0.1045 and RRR of 0.8955
**Figure 1.** A three-dimensional reconstruction of the aorta and its main branches in patient 8 shows the extrinsic compression to the celiac trunk during expiration (*arrow*). ¹

**Figure 2.** A three-dimensional aortic reconstruction of patient 8, after laparoscopic release of the median arcuate ligament, shows good patency of the celiac trunk (*arrow*) at the 1-month follow-up. ¹
Appendix: Scale and Score Definitions

**Visick Score:** “A subjective comparison of pretreatment and post-treatment symptom severity whereby patients describe their symptomatic outcome with four options (asymptomatic, improved, unchanged, and worsened).” 3

**Gastrointestinal Symptom Rating Scale (GSRS):** “A 15-item survey assessing the degree of “bother” from 15 gastrointestinal symptoms on a scale of 1(none) to 7 (very severe).” 3

**12-Item Short Form Health Survey (SF-12):** “A 12-item survey that assessed the health-related quality of life. Results from this survey were converted into two scores, the physical composite score and the mental composite score, using the standardized SF-12 formula available online. Adjusted composite scores were then calculated from the difference between each patient’s composite scales and the published Australian norms of the corresponding gender and age groups. A positive adjusted score can be interpreted as a higher quality of life of the patient compared with the Australian norm. A negative adjusted score can be interpreted as a lower quality of life of the patient compared with the Australian norm.” 3

**SF-36:** “Patient-reported outcomes were measured using the SF-36 v.2. This scale measures eight dimensions of health to assess patient quality of life and the potential burden of disease. Scores were
computed for physical functioning, physical role functioning, bodily pain, general health, vitality, social functioning, emotional, and mental health by summing the item scores within each scale. A global mean score was measured pre- and post-surgery. Since Antonescu et al. [30] observed that a 10 percent increase in the SF-36 score correlated with significant surgical success, a 10 percent increase was used similarly to indicate a good outcome. A neutral outcome was defined as an SF-36 score increase between 1 and 10 percent. Poor outcome was defined as an SF-36 global score that decreased or was unchanged.”