

## Case Series

# A study in patients of median arcuate ligament syndrome undergoing laparoscopic management

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## ABSTRACT

Median arcuate ligament syndrome (MALS), a rare disease characterized as a result of extrinsic compression by diaphragmatic fibers arching on the celiac artery. This syndrome presents a challenging diagnostic workup due to its various clinical presentations and incomprehensible pathophysiology, requiring various investigations to exclude other causes. Laparoscopic surgery is gaining popularity. The aim was to study presenting age, sex, clinical symptoms, postoperative symptom relief and re-establishment of celiac artery blood flow after laparoscopic surgery. The prospective study was carried out in a tertiary care hospital for 10 cases of MALS over 24 months. All patients underwent extensive evaluation with no definitive diagnosis and persistent symptoms. Radiologically proven cases were operated laparoscopically and post-operative celiac artery Doppler was done on 6 months follow-up. All demographic data, clinical characteristics, diagnostic studies, management and outcomes were reviewed. 70% cases of MALS present between 3rd to 5th decades with 85% female preponderance. Postprandial epigastric pain, most common presenting symptom in 60% cases. Laparoscopic surgery is extremely effective and considered to be safe, having substantial reduction in symptoms in 90% patients and re-establishment of celiac artery blood flow (PSV) in 100% patients on 6 months follow-up. Surgeons evaluating patients with chronic abdominal pain should consider MALS as a differential diagnosis. Prompt diagnosis and treatment are critical for resolving symptoms and improving patients' quality of life. Radiological investigations are pivotal in diagnosis and post-operative evaluation. Laparoscopic management is recommended due to its shorter postoperative stay, less post-operative pain and cosmetic suture line.

**Keywords:** MALS, Postprandial epigastric pain, Celiac artery blood flow, Peak systolic velocity, Laparoscopic surgery

## INTRODUCTION

Median arcuate ligament syndrome (MALS), also known as Dunbar syndrome is a rare vascular disorder that characterized by the compression of the celiac artery due to the downward positioning of fibrous connections originating from the median arcuate ligament and diaphragmatic crura, described by Harjola and the first surgical treatment was described after two years by Dunbar.<sup>1-3</sup> Median arcuate ligament forms the fibrous arch, forming a junction between right and left crura of

the diaphragm. It forms the anterosuperior boundary of aortic hiatus at the level of 12th thoracic vertebra just above the origin of the celiac artery.<sup>4</sup> It may cause foregut ischemia due to low number of collaterals or mid gut ischemia due to diversion of blood flow to foregut. Apart from the compression of the extrinsic vessels, the multifactorial etiology of medial arcuate ligament syndrome has led to the suggestion that it may be a neurogenic disease affecting the somatic nerves in the splanchnic plexus, causing altered sensation and pain.<sup>5</sup>

Symptoms are due to an extrinsic compression which result in a narrowing of the celiac artery due to a low insertion of the median arcuate ligament or high origin of the celiac artery. A number of gastrointestinal symptoms, including chronic postprandial epigastric pain, nausea, vomiting, abdominal fullness, weight loss, anorexia, and occasionally abdominal pain during physical activity due to intermittent visceral ischemia, are variable presenting clinical symptoms. An estimated 10-24% of people may have MALS, yet its true prevalence remains uncertain due to its variable clinical expression.<sup>2,6</sup> This syndrome presents a challenging diagnostic workup due to its various clinical presentations and incomprehensible pathophysiological mechanism, requiring various investigations to exclude other causes of abdominal pain. Though a number of imaging and diagnostic modalities, such as Duplex ultrasonography, contrast enhanced computed tomography, CT angiography, MR angiography, gastric tonometry and angiography, can suggest findings consistent with median arcuate ligament syndrome, the condition is mostly diagnosed after other more common conditions have been ruled out. CT with 3-D reconstruction is the gold standard to diagnose MALS. It includes information regarding the anatomy and exclude other causes like atherosclerosis.<sup>7</sup> Using the percentage of stenosis seen on an angiography, the degree of external compression can be calculated.

The general principle is to decompress the celiac artery. Options for treatment include open, laparoscopic or robot-assisted release of the median arcuate ligament, retroperitoneal endoscopic release and percutaneous transluminal angioplasty with stenting. Historically, median arcuate ligament release was accomplished by an open surgical technique with higher morbidity. Percutaneous transluminal angioplasty (PTA) and stenting have also been used; however, they were associated with early restenosis due to external compression from the ligament.<sup>8</sup> Laparoscopic approach for the correction of median arcuate ligament syndrome has gained significant popularity over time and considered to be feasible and safe.

## CASE SERIES

We performed an observational prospective study of 10 cases of MALS in the surgery department of tertiary care hospital over a period of 24 months.

### *Inclusion criteria*

Patients with symptoms of median arcuate ligament syndrome and radiologically confirmed external compression of celiac artery by median arcuate ligament; patients undergoing laparoscopic median arcuate ligament release surgery; patients who gave consent for the study were included.

### *Exclusion criteria*

All cases with incidental radiological finding of celiac artery compression; patients undergoing conservative management and surgery through an open or endoscopic approach; patients who lost follow up were excluded.

In the study method, all admitted patients underwent extensive evaluation of their symptoms with no definitive diagnosis and persistence of symptoms. The refractory nature of these symptoms, coupled with suspicion of MALS. Patients were evaluated about the chronology of symptoms and previous conservative management. All patient underwent basic hematological tests including complete blood count, liver function test, renal function test, coagulation studies, serum electrolytes and lipid profiles. ECG and echocardiography were done to evaluate the patient's overall health. Upper and lower endoscopy were also performed to rule out any abnormality of gastrointestinal tract. They were comprehensively evaluated to exclude other causes, and the diagnosis of MALS was confirmed by CECT abdomen and CT aortogram. All the patients underwent laparoscopic surgical release of the median arcuate ligament. All surgeries were performed by the same team of surgeons with average length of postoperative stay was only 1-2 days. Improvement in Doppler ultrasound peak systolic velocity was noted on 6 month follow up in all patient. All demographic data, baseline clinical characteristics, diagnostic studies, management, and outcomes were reviewed and recorded.

### *Surgical technique*

Patient was placed in the french position with the surgical table in reverse Trendelenburg position. After insertion of first trocar for camera in midline, carbon dioxide was insufflated. Then other 4 trocar inserted, a 10 mm trocar on the right axillary line below the costal arch for the liver retractor; two 5 mm trocars, one on the right and one on the left mid-clavicular line; and a 5 mm trocar on the left anterior axillary line under the left costal arch for the assistant. After inspecting the peritoneal cavity, left lobe of the liver was retracted and lesser omentum was divided along the lesser curvature. Esophagus identified, isolated and retracted with umbilical tape. Dissection of the tissue was completed and supraceliac aorta posterior to the esophagus identified and dissected toward the origin of celiac artery. Fibers of the median arcuate ligament identified and resected until no external compression present.

In our study, 7 patients that suffered from median arcuate ligament syndrome were of 31-50 years of age (70%). Median age for MALS was 40.6 years in our study.

Median arcuate ligament syndrome was observed to have female preponderance with 8 patients (80%) being female. 6 patients were female in 31-50 year age group.

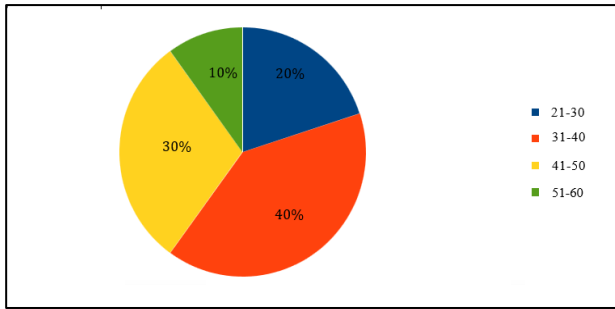


Figure 1: Age distribution.

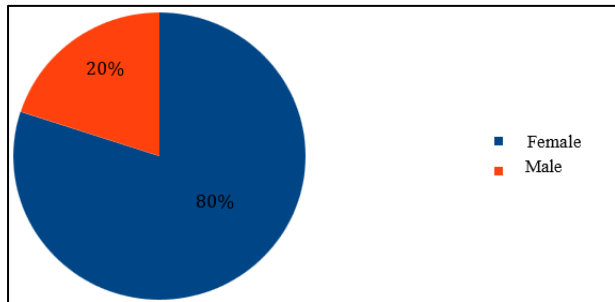


Figure 2: Gender distribution.

Table 1: Chief complaints.

Chief complaints	No. of patients	Percentage
Epigastric pain	6	60
Postprandial fullness	2	20
Weight loss	1	10
Anorexia	1	10

8 out of 10 patients presented with epigastric pain (60%) or postprandial fullness (20%) as presenting complaints. Weight loss and anorexia were found least commonly (10%) as chief complaint although these were frequently found as associated complaints with either epigastric pain or postprandial fullness.

Table 2: Radiological (angiogram) investigations.

Percentage of compression (%)	No. of patients	Percentage
20 to 30	1	10
30 to 40	2	10
40 to 50	4	40
50 to 60	2	20
60 to 70	1	10

Angiogram is considered as an important tool in assessing the severity of the disease and subsequently predicting the chances of complications and prognosis post operatively. In our study it was observed that 60% of the patients had 30-50% compression and 70% patients had less than 50% compression.

Table 3: Post-operative relief of symptoms.

Relief of symptoms	No. of patients	Percentage
Complete	7	70
Partial	2	20
No relief	1	10

Complete relief from symptoms could be achieved in 7 patients (70%) and no relief in just 1 patient (10%). Overall, 90% of the patients showed improvement from their pre-operative state after laparoscopic release of median arcuate ligament.

Table 4: Post-operative celiac artery re-establishment of blood flow.

Re-establishment	No. of patients	Percentage
Complete	10	100
Partial	0	0
None	0	0

Celiac artery Doppler was done in follow up after 6 months to evaluate the adequacy of the laparoscopic median arcuate ligament release. In our study all patients achieved complete re-establishment of blood flow (PSV) on postoperative celiac artery Doppler study.

## DISCUSSION

MALS, also known as Dunbar syndrome is a rare vascular disorder that characterized by the compression of the celiac artery due to the downward positioning of fibrous connections originating from the median arcuate ligament and diaphragmatic crura. MALS is a rare disorder that is often misdiagnosed and not considered in the differential diagnosis, resulting in the exhaustive negative workup, after which suspicion may arise for this entity. The condition tends to favor a female phenotype (4:1) and typically emerges within the age range of 30 to 50 years.<sup>9</sup> Lipshutz et al first described MALS in 1917 with only a few later cases documented, such as in 1963 and 1965.<sup>10</sup> Interestingly, epidemiologic review of cases revealed higher incidence in females (4:1), notably between the ages of 40 and 60 years old.<sup>11</sup> The majority of patients in our study presented in 3rd to 5th decade of life with a mean age of 40.6 years with 80% being female patients, whereas in Naveen et al study, mean age of patients was 52 years.<sup>12</sup> In a study of Diab et al. the reported median age was 46 years with 83.3% female preponderance.<sup>13</sup>

A triad of symptoms including weight loss, nausea, vomiting, and postprandial epigastric pain, is frequently observed.<sup>14</sup> Despite its infrequency, MALS remains a subject of medical interest, yet its true prevalence remains uncertain due to its variable clinical expression. Presenting clinical symptoms are various gastrointestinal symptoms. The doctors evaluating patients with nonspecific symptoms such as postprandial pain and

bloating should consider MALS as a diagnosis. In this study, epigastric pain (60%) and postprandial fullness (20%) were the most common chief complaints. A Mayo clinic study of 36 patients, Cusati et al reported 94% patient with nonspecific abdominal pain and 80% with postprandial abdominal pain.<sup>15</sup> In Diab et al the presenting complaint of all the patient was postprandial abdominal pain.<sup>13</sup>

Detailed clinical history and physical examination should precede medical evaluation with radiological investigation. The clinical examination is often unremarkable and non-specific with vague mild epigastric tenderness, with an occasional finding of a mid-abdominal systolic bruit on auscultation.<sup>16</sup> Julius' findings described that epigastric bruits were present in 16% of asymptomatic individuals and 30% of younger patients with MALS, however, we did not identify these features in our experience.<sup>17</sup>

Diagnosis relies on the utilization of imaging modalities like CECT abdomen, CT angiogram and color Doppler ultrasound with the respiratory variation of celiac blood flow. Doppler vascular ultrasound, to evaluate respiratory variation can help determine if compression is clinically significant and may be contributing to abdominal pain.<sup>18</sup> CTA helps in the diagnosis allowing three-dimensional visualization of compressed celiac artery. CT Angiogram has high resolution and can show changes like proximal smooth narrowing of artery, post stenotic dilatation and hooked appearance. In study done by Ilica et al all patients showed radiological changes in CT Angiogram and also they classified the arterial stenosis into four grades: grade I-0-49% diameter reduction as mild stenosis; grade II-50-74% diameter reduction as moderate stenosis; grade III-75-99% diameter reduction as severe stenosis; grade IV-total occlusion.<sup>19</sup>

The stenosis of the celiac artery was grade III in 7 patients and grade II in 3 patients in our study, whereas Naveen et al. reported 3 patients with grade III and 2 patients with grade II stenosis.<sup>12</sup> In Diab et al the degree of occlusion confirmed by CTA was <50% in one case, 70% in one case, and the remaining four were  $\geq 90\%$ .<sup>13</sup>

The surgical approach of minimally invasive laparoscopic technique focuses on releasing the ligament and overcoming the compression of the celiac artery from which the symptoms are emanating. Symptomatic relief is seen in a majority of patients after undergoing minimally invasive treatment, as was seen in our patient. Complete reversal of symptoms could be achieved in 70% of patients and partial relief in 20% of patients in our study. Overall, 90% of the patients showed improvement from their pre-operative state on 6 months follow up. In a series of 400 patients, Jimenez et al reported 85% experienced immediate post-operative relief of symptoms.<sup>20</sup> Jesse et al reported that Five of the six patients had complete resolution of their symptoms post-operatively, with one patient reporting improvement

without complete resolution of his abdominal pain.<sup>21</sup> Finally, it is important for the long-term follow-up of these patients to make sure that they are symptom-free, thus confirming the sustainability of the surgery and the accuracy of the diagnosis.

## CONCLUSION

This case series highlight the significance of considering MALS as a cause of abdominal pain and emphasize the need for further research to improve our understanding and management of this uncommon vascular condition. Prompt diagnosis and treatment following suspicion of the syndrome are critical for resolving symptoms and improving patients' quality of life. Laparoscopic management is gaining popularity and recommended owing to the basic advantages of minimal invasive approach clubbed with acceptable learning curve and ease of performing, along with excellent visualization of the celiac trunk origin and the median arcuate ligament. Laparoscopic surgery has the additional benefit of having shorter postoperative stay, less post-operative pain and more cosmetic suture line.

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