The Prevalence of Co-Morbidities Associated with Median Arcuate Ligament Syndrome

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Abstract

Median Arcuate Ligament Syndrome (MALS) is a rare disorder that occurs when the celiac artery and celiac nerves are compressed by the median arcuate ligament. Symptoms of MALS can include significant postprandial abdominal pain, weight loss, nausea, and vomiting. It has been suggested that other co-morbid conditions may be associated with MALS (1, 2), however there has only been one study of 11 patients looking at multiple co-morbidities (3). In that study, 71.4% of patients had radiographic evidence of delayed gastric emptying, 18.2% had Ehlers-Danlos Syndrome (EDS), and 27.3% had Postural Orthostatic Tachycardia Syndrome (POTS), all of which are higher than the prevalence of these conditions in the general population. Another study of 51 patients diagnosed with MALS showed that 28% met the criteria for a psychiatric diagnosis (4).

We conducted a survey of individuals with MALS for self-reporting co-morbid conditions. 242 patients who self-identified as having MALS were surveyed. 52.7% of patients had a type of dysautonomia. Of those patients, 71.0% reported a diagnosis of POTS. EDS was reported in 31.7% of patients. Gastroparesis was reported in 37.8% of patients, and Mast Cell Activation Syndrome (MCAS) in 22.6% of patients. Diagnosable mental health conditions were reported in 66.1% of patients: 33.8% of these patients admitted to anxiety, and 29.6% admitted to depression. Autoimmune conditions were reported in 37.0% of patients, and 27.4% reported Small Intestinal Bacterial Overgrowth (SIBO).

The increased power in our study substantiates previous findings and reinforces that these conditions have a higher prevalence in MALS patients than in the general population. MCAS, SIBO and autoimmune conditions have not been previously studied in MALS patients, and thus we present the first association between MALS and these conditions. This data will help providers better care for their MALS patients by increasing awareness of co-morbid conditions.

Objective

The goal of this study was to determine the prevalence of co-morbid conditions associated with Median Arcuate Ligament Syndrome (MALS). Previous studies have looked at relatively small sample sizes with a limited number of studied co-morbid conditions. Our goal was to survey a larger population of MALS patients and broader categories for co-morbid conditions.

Introduction

The incidence of MALS in the population is predicted to be 2 cases per 100,000 patients, thus other possible diagnoses must be explored prior to a diagnosis of MALS is reached. It has been suggested that other co-morbid conditions may be associated with MALS (2). Despite this postulation, there has only been one small study of 11 patients looking at multiple co-morbidities of MALS (3). As the co-morbid conditions often associated with MALS are rare findings in the general population, we decided to explore the prevalence of these conditions in patients who already had been diagnosed with one relatively rare condition - MALS. In addition to the already loosely associated conditions of dysautonomia, EDS, gastroparesis and psychiatric conditions with MALS, we included an analysis of the prevalence of SIBO, autoimmune conditions and MCAS. This was the first time in the current literature that these conditions were studied in conjunction with a diagnosis of MALS.

Study Design

Participants:
Patients diagnosed with MALS were identified via three Facebook support groups and consent for posting the survey link was obtained via communication with the page administrators. The survey was posted on all three pages allowing for all page members to view the survey. The number of Facebook members in each group is 3,800, 4,400, and 2,700.

Inclusion criteria: Individuals must have been 18 years or older at the time of the survey and have a diagnosis of Median Arcuate Ligament Syndrome (Celiac Artery Compression Syndrome) diagnosed by a medical provider (MD/DO, PA, NP) at any time in the individual's life prior to completion of the survey. Exclusion criteria: Individuals who do not meet the inclusion criteria; those who had not been diagnosed with Median Arcuate Ligament Syndrome (Celiac Artery Compression Syndrome) by a medical provider (MD/DO, PA, NP) at any time in the individual's life prior to completion of the survey or were not 18 years of age or older.

Instrument:
We developed an online survey. The questionnaire first obtained informed consent from the participants and ensured that they met the inclusion criteria. Demographic information included age and gender. Health history information regarding a diagnosis of dysautonomia, EDS, SIBO, mental health conditions, gastroparesis, autoimmune conditions, MCAS, and SIBO were obtained.

Results

What are the demographics of the 242 MALS patients surveyed?

Gender	Number of MALS Patients
Female	228
Male	11
Nonbinary	2
Gender fluid	1

Table 1. Gender of 242 respondents of self-identified MALS patients.

-The overwhelming majority of those surveyed were female (94.2%). This fits with the cited demographic of "young adult females" being most likely to be affected (1).
-Young adults made up the majority of those surveyed, with the largest group being between the ages of 18 and 22.

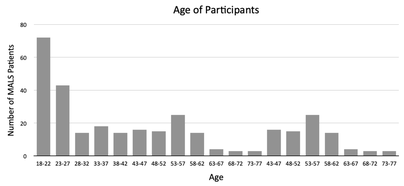


Figure 1. Age of 242 respondents of self-identified MALS patients.

What is the prevalence of the co-morbidities evaluated?

-All co-morbidities were higher in the MALS population surveyed as compared to the general population (3).
-Dysautonomia, EDS, and mental health conditions were higher in our surveyed population than the small studies that had previously reported these conditions linked to MALS.
-Gastroparesis was reported to be lower than the previous study with 37.8% reporting gastroparesis in our study compared to the radiographic evidence present in 71.4% of patients in the previous small study (3).
-MCAS, Autoimmune conditions, and SIBO had not been previously studied and we showed that these conditions had a higher prevalence in the MALS patients studied than in the general population

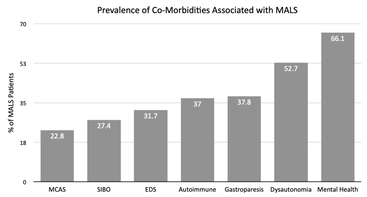


Figure 2. Percentage of co-morbidities present in 242 respondents of self-identified MALS patients.

What type of dysautonomia do MALS patients report?

-POTS was found to be the most prevalent type of dysautonomia (72%) among those who had MALS and a diagnosed dysautonomia (52.7%)

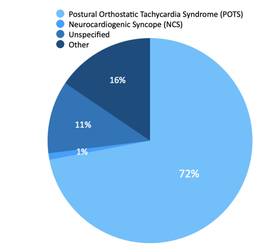


Figure 3. Type of dysautonomia of 126 respondents of self-identified MALS patients with self-identified dysautonomia.

What type of Ehlers-Danlos Syndrome (EDS) do MALS patients report?

-Hypermobile EDS (hEDS) was found to be the most prevalent type of dysautonomia (77%) among those who had MALS and a diagnosed type of Ehlers-Danlos Syndrome (31.7%)

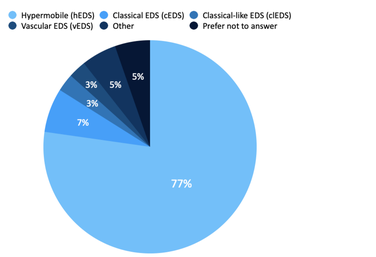


Figure 4. Type of Ehlers-Danlos Syndrome (EDS) of 75 respondents of self-identified MALS patients with self-identified EDS.

What type of autoimmune condition(s) do MALS patients report?

-Various types of autoimmune conditions were present in 37% of MALS patients surveyed. Other types of autoimmune conditions accounted for 32%, Inflammatory Bowel Disease (IBD) comprised 19%, Celiac disease was present in 12% of those with autoimmune conditions and Hashimoto's Thyroiditis also showed 12%.

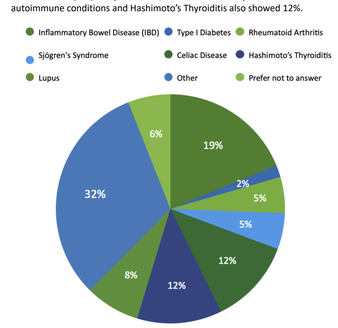


Figure 5. Type of autoimmune condition(s) of 85 respondents of self-identified MALS patients with self-identified autoimmune condition(s).

What type of psychiatric condition(s) do MALS patients report?

-Anxiety was found to be the most prevalent type of psychiatric diagnosis (34%), with depression as a second (30%) among those who had MALS and a diagnosed mental healthy condition (66.1%)

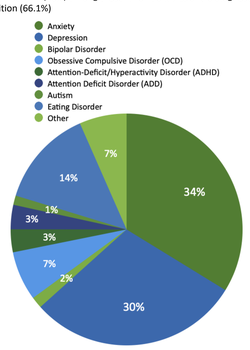
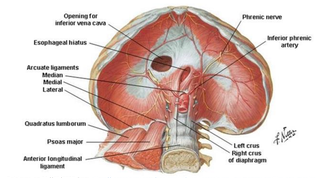


Figure 6. Type of mental health condition(s) of 152 respondents of self-identified MALS patients with self-identified mental health condition(s).

Discussion

- Our sample size of 242 is much larger than previous studies that included 11 and 51 MALS patients while looking at co-morbid conditions (3, 4).
  - Our study employed self-identification of patients with a MALS patient instead of using retrospective chart review.
  - Our sample demographics contained an overwhelming majority of young females which fits with the data in the literature citing this as the main population affected (1). This could also be due to bias where young females could be more likely to participate in Facebook support groups.
- The prevalence of all co-morbid conditions including Dysautonomia, Ehlers-Danlos Syndrome (EDS), Gastroparesis, Mast Cell Activation Syndrome (MCAS), mental health conditions, autoimmune diseases and Small Intestinal Bacterial Overgrowth (SIBO) were higher in our surveyed MALS population than in the general population.
- Dysautonomia: 52.7% of those surveyed reported a diagnosed type of dysautonomia, and 72% reported a diagnosis of POTS.
- Ehlers-Danlos Syndrome (EDS): Our study found that the prevalence of EDS in patients with MALS could be much higher, reported at 31.7% (Figure 2), with hypermobile EDS (hEDS) as the most prevalent type of EDS at 77% (Figure 4).
- Gastroparesis was reported in 37.8% of patients surveyed, which was lower than the 71.4% in a previous studying when radiographical evidence was used. This discrepancy is most likely due to the lack of radiographic evidence, patient reporting, or the small sample size of the previous study.
- Psychiatric condition(s) were reported in 66.1% of MALS patients surveyed which is higher than the previous study's findings of 28% of patients with MALS (4). This may be due to patient reporting, and duration of time to diagnosis. Often a delay in MALS diagnosis results in physiologic distress and subsequent mental illnesses or misdiagnosis of eating disorders as reported by patients studied.
- Autoimmune conditions, SIBO, and MCAS had not been previously studied in relation to MALS patients. We found that all three of these diagnoses may have a higher prevalence in MALS patients than in the general population with 37%, 27.4% and 22.8% of MALS patients reporting these diagnoses respectively (Figure 2).



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Conclusion

Median Arcuate Ligament Syndrome (MALS) is a condition characterized by significant post-prandial abdominal pain, weight loss, nausea and vomiting. MALS has been proposed to be associated with multiple other co-morbid conditions in a few small studies (3, 4). We found an increased prevalence of Dysautonomia, Ehlers-Danlos Syndrome, Mast Cell Activation Syndrome, Small Intestinal Bacterial Overgrowth, psychiatric conditions and autoimmune conditions among those with MALS. This data will help providers and patients better understand the prevalence of other conditions. This study may also decrease suffering and the time to diagnose patients with MALS if other co-morbidities are recognized in patients. Further research into the prevalence of MALS symptoms in those who carry diagnoses of co-morbid conditions is needed to further explore this possible implication.

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