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Median arcuate ligament syndrome in the pediatric population☆,☆☆

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Abstract

Objectives: Median arcuate ligament syndrome (MALS) is a vascular compression syndrome with symptoms that overlap chronic functional abdominal pain (CFAP). We report our experience treating MALS in a pediatric cohort previously diagnosed with CFAP.

Patients and Methods: We prospectively evaluated 46 pediatric (<21 years of age) patients diagnosed with MALS at a tertiary care referral center from 2008 to 2012. All patients had previously been diagnosed with CFAP. Patients were evaluated for celiac artery compression by duplex ultrasound and diagnosis was confirmed by computed tomography. Quality of life (QOL) was determined by pre- and postsurgical administration of PedsQL™ questionnaire. The patients underwent laparoscopic release of the median arcuate ligament overlying the celiac artery which included surgical neurolysis. We examined the hemodynamic changes in parameters of the celiac artery and perioperative QOL outcomes to determine correlation.

Results: All patients had studies suggestive of MALS on duplex and computed tomography; 91% (n = 42) positive for MALS were females. All patients underwent a technically satisfactory laparoscopic surgical release resulting in a significant improvement in blood flow through the celiac artery. There were no deaths and a total of 9 complications, 8 requiring a secondary procedure; 33 patients were administered QOL surveys. 18 patients completed the survey with 15 (83%) patients reporting overall improvement in the QOL. Overall, 31/46 patients (67%) reported improvement of symptoms since the time of surgery.

Conclusions: MALS was found to be more common in pediatric females than males. Laparoscopic release of the celiac artery can be performed safely in the pediatric population. Surgical release of the artery and resultant neurolysis resulted in significant improvement in the blood flow, symptoms, and overall QOL in this cohort. The overall improvement in QOL outcome measures after surgery leads us to conclude that

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MALS might be earlier diagnosed and possibly treated in patients with CFAP. We recommend a multidisciplinary team approach to care for these complex patients.

The majority of chronic abdominal pain in children is thought to be functional (CFAP), that is, without demonstrable evidence of an underlying anatomic, metabolic, infectious, inflammatory, or neoplastic disorder [1,2]. Many of these difficult patients carry a host of symptom-based diagnoses, including functional dyspepsia, abdominal migraine, and especially, irritable bowel syndrome (IBS), all within the domain of what is now better known as functional gastrointestinal disorders (FGID). This classification was most recently updated as the Rome III criteria in 2006 [1–6]. The pathophysiology for FGID is poorly understood, but is thought to involve abnormalities in the enteric nervous system leading to dysregulation of brain-gut communication to explain altered bowel motility, visceral hypersensitivity, and stress-mediated effects in the pathogenesis of functional abdominal pain [1–4]. IBS is perhaps the best example where annual direct and indirect management costs are estimated to be $8 and $25 billion respectively with evidence pointing to its origin in childhood of those with CFAP [1,2,7–11]. As opposed to chronic abdominal pain where there is demonstrable pathology, i.e. celiac disease, inflammatory bowel disease, etc., for which there are established treatment strategies with mostly predictable outcomes, treatment for FGID remains unproven and published results mostly difficult to interpret [12–17].

Median arcuate ligament syndrome (MALS), also known as celiac artery compression syndrome, was first described by Harjola in 1963 [18]. The hallmark symptoms of postprandial abdominal epigastric pain, nausea, occasional diarrhea and weight loss, overlap with those of CFAP. Although MALS has been advocated as an unusual cause of abdominal pain, the evidence has been based principally on anecdotal or small single-center retrospective analysis rather than level 1 or level 2 evidence [18–20]. In anatomical terms, MALS is felt to be caused by a compressive anatomic relationship of the diaphragmatic crura to the celiac vessels leading to decreased flow, a steal phenomenon and resultant postprandial abdominal pain [18,21,22,19]. Similarly it has been suggested that neurogenic compression may lead to the clinical symptoms [23]. Only recently have advances in noninvasive, high definition duplex ultrasound scan and CT or MR angiography allowed vascular occlusive diseases such as MALS to be more readily diagnosed based on objective measurement of vessel flow velocity and alterations in vascular architecture. Leveraging these advances in diagnostic imaging in the context of an apparent noncoincidental overlap of GI symptoms between MALS and FGID, we prospectively evaluated MALS in 46 pediatric patients diagnosed with CFAP within the domain of FGID, and we report on the surgical outcomes and quality of life of these patients.

1. Patients and methods

1.1. Patients

We evaluated 46 patients (42 females, 4 males; ages 8.6–20.5 years; median 16.6; mean 16.2 ± 0.5 years) with diagnosis of CFAP with duplex US at a single tertiary care institution between August 2008 and January 2012. This was performed under an IRB approved protocol. A complete GI work up was performed by the patient's gastroenterologist. Typical workup included a complete battery of studies shown in table 1. Study requirements included 1) diagnosis of FGID with chronic abdominal pain by a gastroenterologist and 2) thorough GI evaluation for abdominal pain with a minimum of upper endoscopy and abdominal/pelvic CT scan. Patients with chronic abdominal pain with demonstrable pathology were excluded. Informed consent was obtained from the patient or guardian (<18 y) as required by the University of Chicago Institutional Review Board (#16997A).

1.2. Duplex ultrasound protocol

Prior to any intervention, all patients had a duplex ultrasound in our accredited Intersocietal Accreditation Commission (IAC) laboratory for vascular testing to fully evaluate the visceral vessels. All patients fasted overnight to minimize the amount of abdominal gas present at the time of

Table 1 Guidelines for standard GI workup in patients with possible MALS.

1. Complete blood count with differential, platelet count
2. ESR, C-reactive protein
3. Amylase, lipase
4. Comprehensive metabolic panel (including liver function tests)
5. Prealbumin
6. Thyroid fuction tests (T4, TSH)
7. Serum IgA, tissue transglutaminase IgA, IgG, deamidated antigliadin IgG, and IgA
8. UGI alone or UGI with small bowel follow through
9. Upper endoscopy with biopsy
10. Abdominal ultrasound
11. Urinalysis, ± toxicology screen if indicated, pregnancy test in adolescent females if indicated
12. Growth charts (over previous 2–3 years), BMI

ESR: erythrocyte sedimentation rate. UGI: upper gastrointestinal. BMI: body mass index.