Median Arcuate Ligament Syndrome

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Disclosures
- none

“There is post(prandial) distress, usually cramping abdominal pain, perhaps with radiation to the back and occasionally with a sense of distention with constant abdominal ache. The distress usually develops fifteen to thirty minutes after eating and persists for one to three hours. Nausea, vomiting, or diarrhea may occur. Reluctance to eat along with malabsorption may produce weight loss.”

AJR 95: 731, 1965

Answer: none

Terminology
- Celiac Artery Compression is an anatomic finding
  - Autopsy, CTA, MRA, aortography
- Median Arcuate Ligament Syndrome describes a symptom complex that is the result of compression of the celiac artery and/or celiac ganglion
  - Classical findings post-prandial abdominal pain, weight loss and brut
  - Non-specific findings nausea, vomiting, diarrhea, unprovoked pain, exertional pain, bloating
11/15/2018

#1 Compression of the celiac artery is an irrelevant finding in most patients
- Up to one-third of autopsies
  - 33% of 75 autopsies Lindner Arch Surg 103: 600, 1971
  - 21% of 100 autopsies Derrick Ann Surg 149: 149, 1959
- MRI
  - 57% (55/99) of asymptomatic pts with at least "mild" compression during expiration Lindner Arch Surg 103: 600, 1971
- Diagnostic angiography
  - 7.3% (29/400) chemoembolization Park, Korean J Radiol 2: 8, 2001
- CT
  - 6.7% (18/284) Consecutive Abd CT Kazan, Vascular 21: 293, 2013
  - 3.4% (11/321) Kidney donors Petnys JVS 2018

#2 Symptoms associated with median arcuate ligament syndrome are non-specific
- All Abd CTs for 3 mos Celiac compression was present in 19 of 284 (6.7%)
- No symptom or group of symptoms was specific for MAL syndrome Kazan, Vascular 21: 293, 2013

Real-World Median Arcuate Ligament Syndrome
- Referred for evaluation since 2015 n=75
- 80% (60 of 75) incidental finding esp CTA
- F:M 2:1
  - >60 years – n=6 conventional revascularization
  - <60 Years - n=8 median arcuate ligament release +/- revasc

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<th>Unchanged</th>
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<td>Release + Revasc</td>
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Etiology of Median Arcuate Ligament Syndrome
- Neurogenic
- Vascular
- Psyche

Etiology of Median Arcuate Ligament Syndrome
- Neurogenic
  - Afferent pain fibers
  - Autonomic mediation of gut perfusion
- Vascular
  - Foregut ischemia due to inadequate collateral pathways SMA to Celiac
  - Pancreaticoduodenal to GDA to hepatic
  - Dorsal pancreatic to splenic
  - Note: increased risk of aneurysm formation in collateral pathways – 24% (9/37) (Kazan, Diagn Interv Radiol 24: 281, 2018.24%)
- Psyche
  - Depression, anxiety
Etiology of Median Arcuate Ligament Syndrome

- **Vascular**
  - Foregut ischemia due to inadequate collaterals (GDA, Dorsal Pancreatic)

- **Neurogenic**
  - Afferent somatic pain fibers
  - Autonomic control of gut perfusion

- **Psychogenic**
  - Depression, anxiety

#3 Symptom relief is **not** predicted by severity of post-operative celiac stenosis

- N=29 Lap release with 12 mo F/U* Patel JVS 2018
  - Clinical success (66%) did not correlate with severity of celiac stenosis

- N=39 patients Laparoscopic Release of Median Arcuate Ligament
  - Weber et al, Median arcuate ligament syndrome is not a vascular disease
  - Pre and post op inspiration/expiration duplex u/s
    - MAL Release was effective in most patients (85% - 33 of 39 improved)
    - Including 6 of 7 with persistent celiac stenosis/occlusion
    - Some patients (5 of 39 – 13%) had persistent symptoms despite improved celiac flow

![Celiac Artery Velocity Graph](image1)

![Celiac Artery Velocity Graph](image2)

#4 Stents are **not** effective in the presence of an intact median arcuate ligament

- Balloon expandable stents are prone to crushing
- Self-expanding stents are prone to fracture

![Clinical improvement despite worse blood flow](image3)

![Clinical improvement despite worse blood flow](image4)
#5 Co-morbid psychiatric diagnoses are common in patients with suspected MALS

- N=51 Preoperative psychological assessment*
  - Surgery improved QOL in most patients (66%)
  - 31% (16/51) Required adjunctive procedures
    - Celiac plexus block, PTA, ERCP
  - 28% (14/51) Carried psyche diagnosis
    - Depression, anxiety, PTSD, adjustment disorder, ADHD
    - Psyche Dis associated with worse patient-reported QOL outcomes

*Skelly et al, Impact of psychiatric comorbidities on patient-reported surgical outcomes in adults treated for median arcuate ligament syndrome, JVS 2018

So how select patients?
- Classic history – post-prandial pain, >20# weight loss, age 40-60
- Provocative testing
  - Gastric ischemia
  - Neurogenic pain
  - Psychiatric diagnosis

Predictors of outcome
- Success
  - Age 40-60, post-prandial pain and weight loss >20# Reilly, 1985
  - Age combined with peak expiratory velocity – Brody 2018
  - Post-exercise abdominal pain – Ho 2017
- Failure
  - Emesis – Ho 2017
  - Unprovoked abd pain – Ho 2017
  - Psychiatric diagnosis – Skelly 2018

Tests to identify gastric ischemia
- SMA Intra-arterial vasodilator (Kalapatapu, Vasc and Endovasc Surg 43: 46, 2009)
  - Symptom reproduction with injection of vasodilator; Reduced filling of foregut collaterals due to SMA STEAL
  - 8 of 9 with positive test improved with MAL release
  - 10 of 10 with negative test have been successfully managed non-operatively

- Gastric exercise tonometry (Mensink, JVS 44: 277, 2006)
  - Detects gastric mucosal ischemia
  - Requires a-line, NS tube tonometer, and bicycle ergometer for 30 minutes
  - 29 of 43 symptomatic patients had POSITIVE test for gastric mucosal ischemia
  - 24 of 29 (83%) asymptomatic after OPEN MAL release (22/29) + revasc (7/29) at 3 years
  - Postop tonometry correlated with symptom persistence

Tests to identify neurogenic etiology
- Celiac plexus block
  - Positive response to celiac plexus block may predict successful median arcuate ligament release

Sun et al, Celiac Plexus block as a predictor of surgical outcome for sympathetically-mediated abdominal pain in a case of suspected median arcuate ligament syndrome, A&A Practice 12: 78, 2018
Tests to identify psychogenic component of MALS

- Preoperative psychological testing

Skelly et al, Impact of psychiatric comorbidities on patient-reported surgical outcomes in adults treated for median arcuate ligament syndrome, JVS 2018

Predictive model for success of neurolysis and median arcuate ligament release*

- Severity of expiratory celiac compression combined with patient age predict QOL outcomes
- N=42 Median Arcuate Ligament Release
  - 74% (31/42) - Good (>10% increase in SF-36 score)
  - 14% (6/42) - Neutral
  - 12% (5/42) - Poor
- Expiratory celiac peak velocity was NEGATIVELY correlated with outcome. The tighter the stenosis, the less likely surgery would be beneficial
- Post-op celiac velocity did not predict QOL outcomes suggesting that the degree of celiac stenosis was a marker for severity of compression of the celiac neural plexus. Supports a NON-Vascular etiology

Brody, Surgical Endoscopy published online May, 2018, Washington DC VAMC

What I do

- H&P – esp post prandial pain and >20 lbs weight loss
- CTA
- Insp/Exp Duplex - >200 cm/sec
- Consider angiography with vasodilator injection
- Laparoscopic release – (not all laparoscopists are equivalent!)
- Post op Duplex –
  - PTA for residual stenosis in symptomatic patients
  - Open reconstruction for recurrent/persistent symptoms

Summary

- There remains uncertainty regarding the etiology and optimal management of MALS
- Success achieved in 65-85%
- Significant risk of early recurrence
- There appear to be three interrelated components:
  - Foregut ischemia
  - Neurogenic pain
  - Psychogenic factors
- Provocative testing may reduce the risk of failed surgery