Median Arcuate ligament syndrome

Imaging, Diagnosis and Management

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J BARD MRI

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- Radiology Residency UCLA
- Neuroradiology Fellowship Stanford
- Diagnostic Radiology Associates of Dallas – Partner
- Solo Practice 2003- Present with Envision Radiology
- Married, 2 boys, 15 and 11
- Hobbies include piano lessons, Texas Hold'em, Bass, Music (Blues)
- Disclosure Statement – No financial interest to disclose

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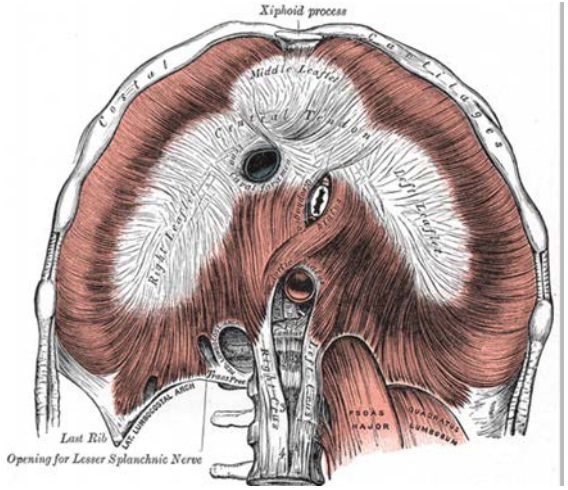
Median Arcuate Ligament syndrome

MALS

- Celiac artery compression syndrome
- Dunbar syndrome, Horjola-Marable syndrome
- Compression of the celiac trunk by the median arcuate ligament
- Worse with expiration

Median arcuate ligament



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MALS symptoms

Common symptoms

- Post prandial pain
- Emesis, bloating, weight loss
- Diagnosis of exclusion and with Doppler ultrasound, CTA

Preoperative workup

- Colonoscopy
- Upper endoscopy
- Testing for Celiac Disease
- Testing for H Pylori
- Upper GI barium swallow
- Ultrasound and HIDA scan
- Psychological and Pain evaluation

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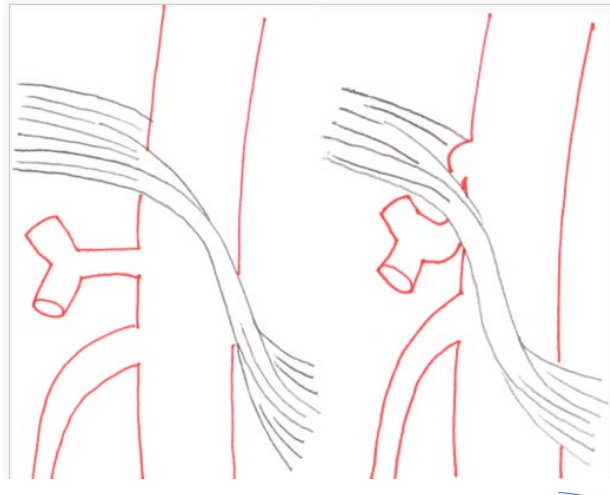
MALS anatomy

Theories

- Abnormally inferior and anterior location of MALS
- Compresses the celiac axis, causing ischemia
- Possible compression of the celiac ganglia
- However, this occurs in up to 25% of normal individuals

Normal on left, abnormal on right



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Celiac axis Anatomy

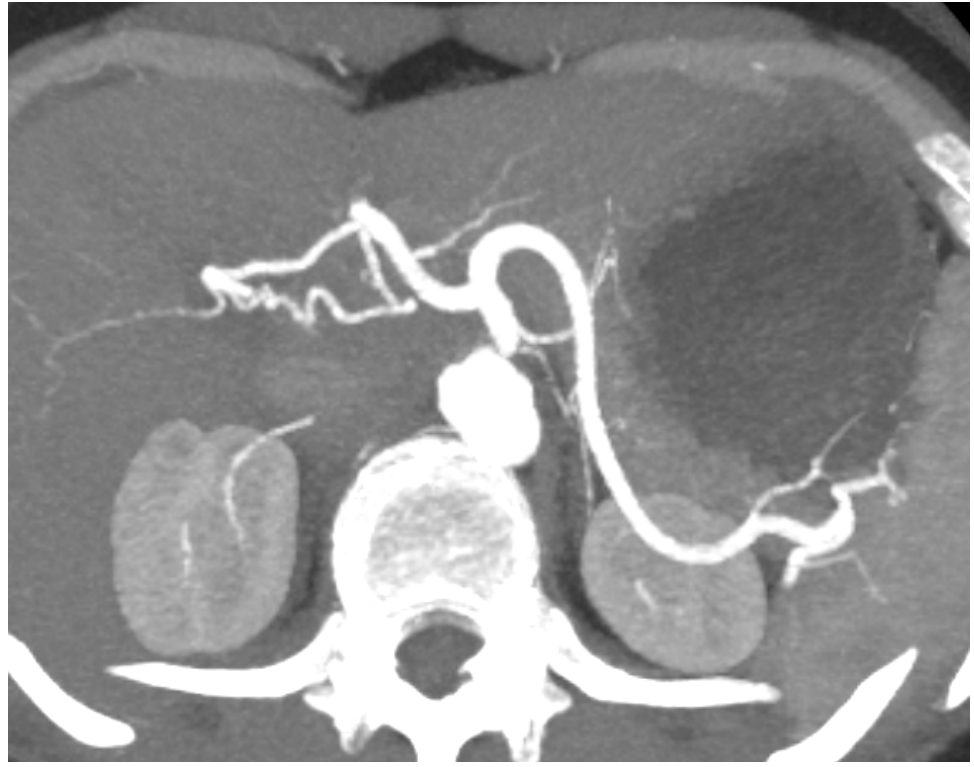
Celiac trunk

Left Gastric

Splenic artery

Common Hepatic artery

Gastroduodenal artery



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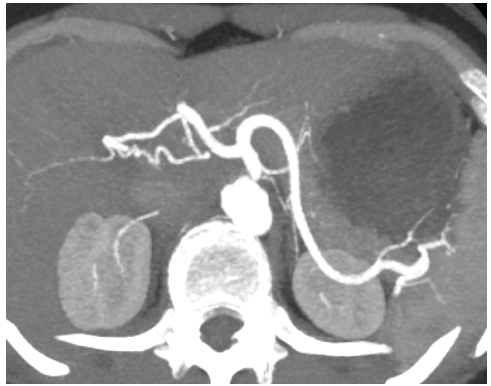

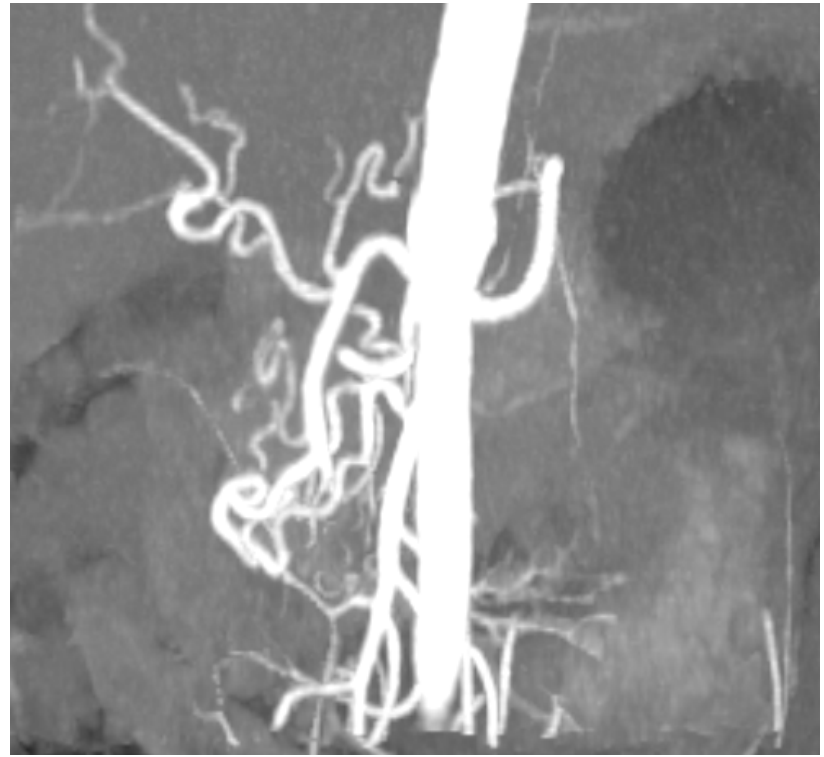

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Celiac axis Anatomy

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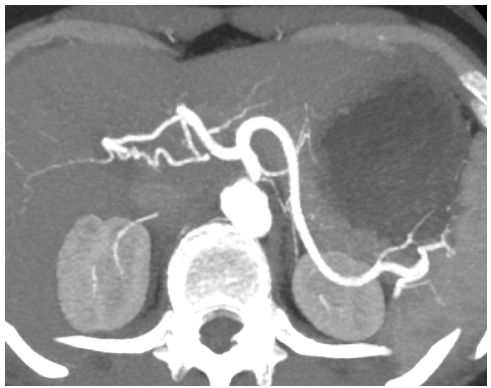

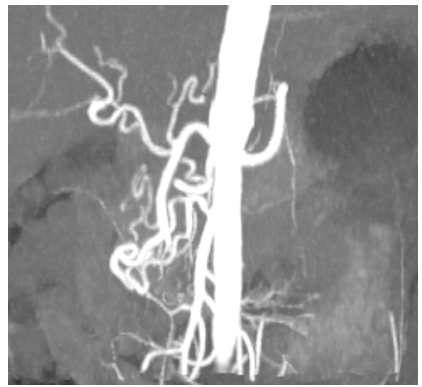

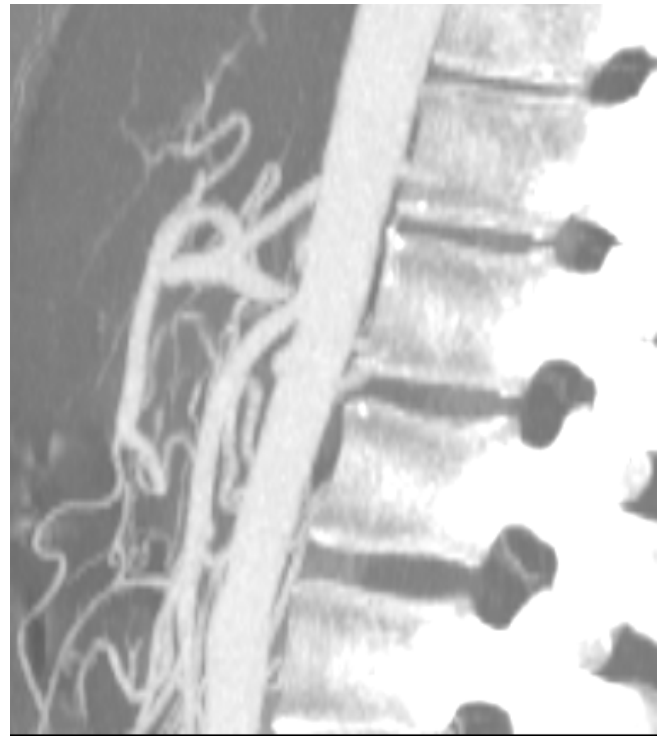

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MALS Management Theory

- Compression of the Celiac axis leads to symptoms
- Vascular - Bowel ischemia
- Neurogenic - Celiac Ganglia compression cause pain
- Laparoscopic treatment of MALS can help resolve symptoms by decompression of the artery, but also releasing or destroying the adjacent ganglia
- Chronic Functional abdominal pain Syndrome overlaps with MALS
  - CFAP is similar to IBS, but no bowel changes (diarrhea, constipation)

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Web and other Resources

- Horton KM, Talamini MA, Fishman EK (2005). "Median arcuate ligament syndrome: evaluation with CT angiography". *Radiographics* **25** (5): 1177–82.doi: 10.1148/rg.255055001. PMID 16160104.
- YouTube “SMA Syndrome and Median Arcuate Ligament Syndrome: True Syndromes or Fantasy” ctisus
- https://www.youtube.com/watch?v=L9ftfucwLWI
- University of Chicago MALS Program
  - http://www.ucmals.com/mals.html

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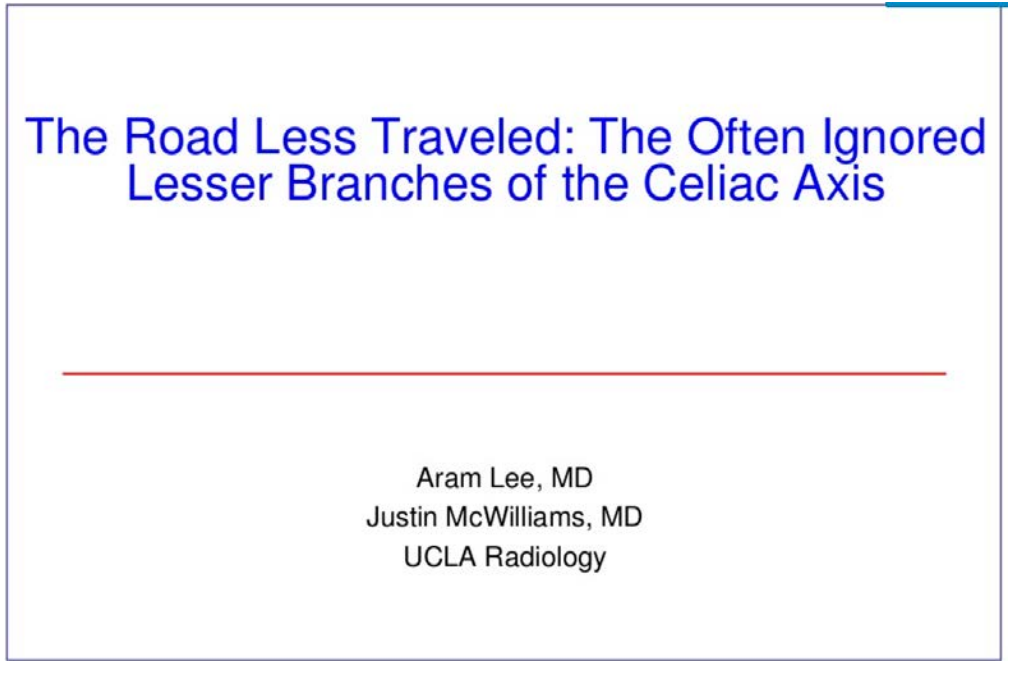

http://www.slideshare.net/pryce27/rsna-final-2?qid=1560f210-f9be-489a-b48f-1231cf91008d&v=qf1&b=&from_search=6

http://www.theparisreview.org/blog/2015/09/11/the-most-misread-poem-in-america

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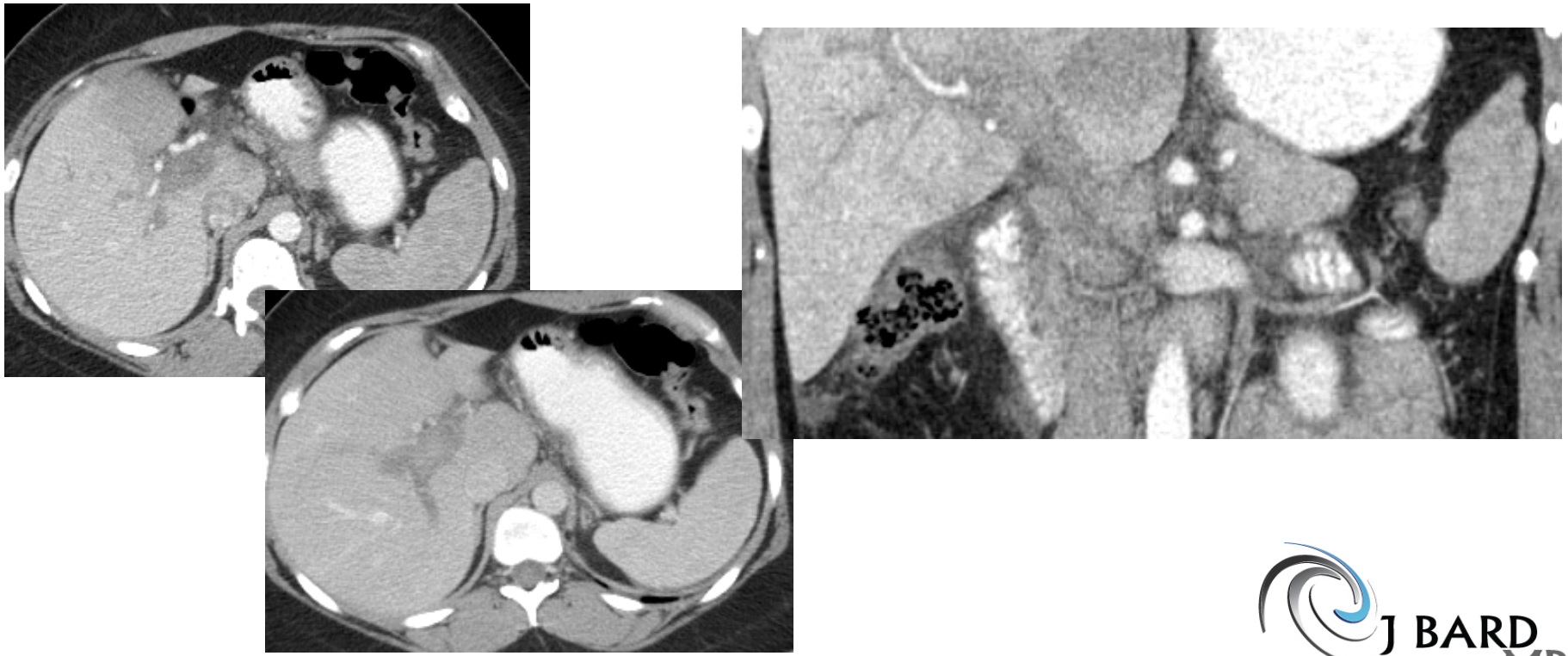
The Road Not Taken- Case 1



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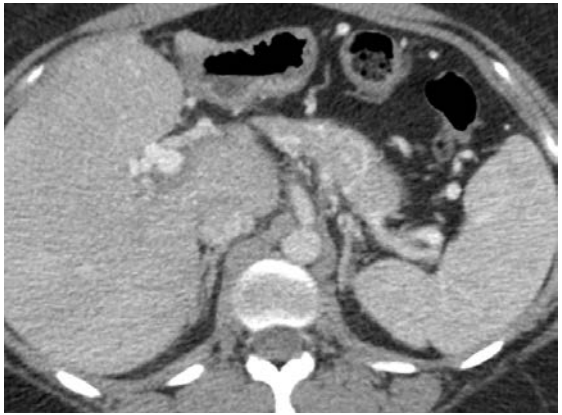
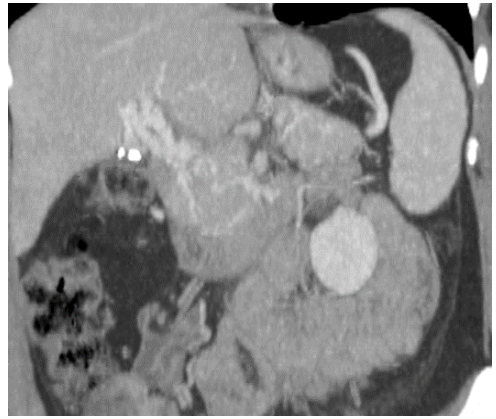
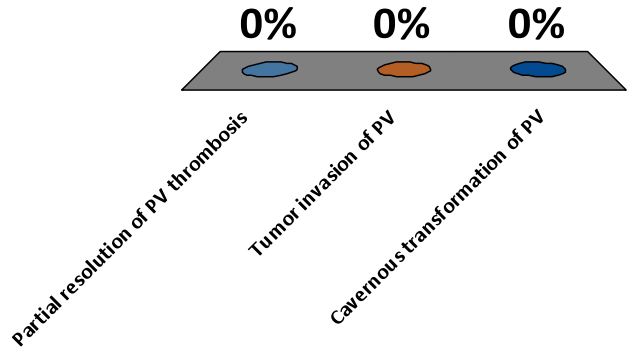
What has happened?

A. Partial resolution of PV thrombosis
B. Tumor invasion of PV
C. Cavernous transformation of PV







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Ultrasound of MALS

- **Mesenteric/celiac duplex ultrasound interpretation criteria revisited.**
- J Vasc Surg. 2012 Feb;55(2):428-436.e6; discussion 435-6. doi: 10.1016/j.jvs.2011.08.052. Epub 2011 Dec 21.
- he PSV threshold that provided the highest OA for ≥50% stenosis was **≥240 cm/s and** for ≥70% stenosis was **≥320 cm/s**

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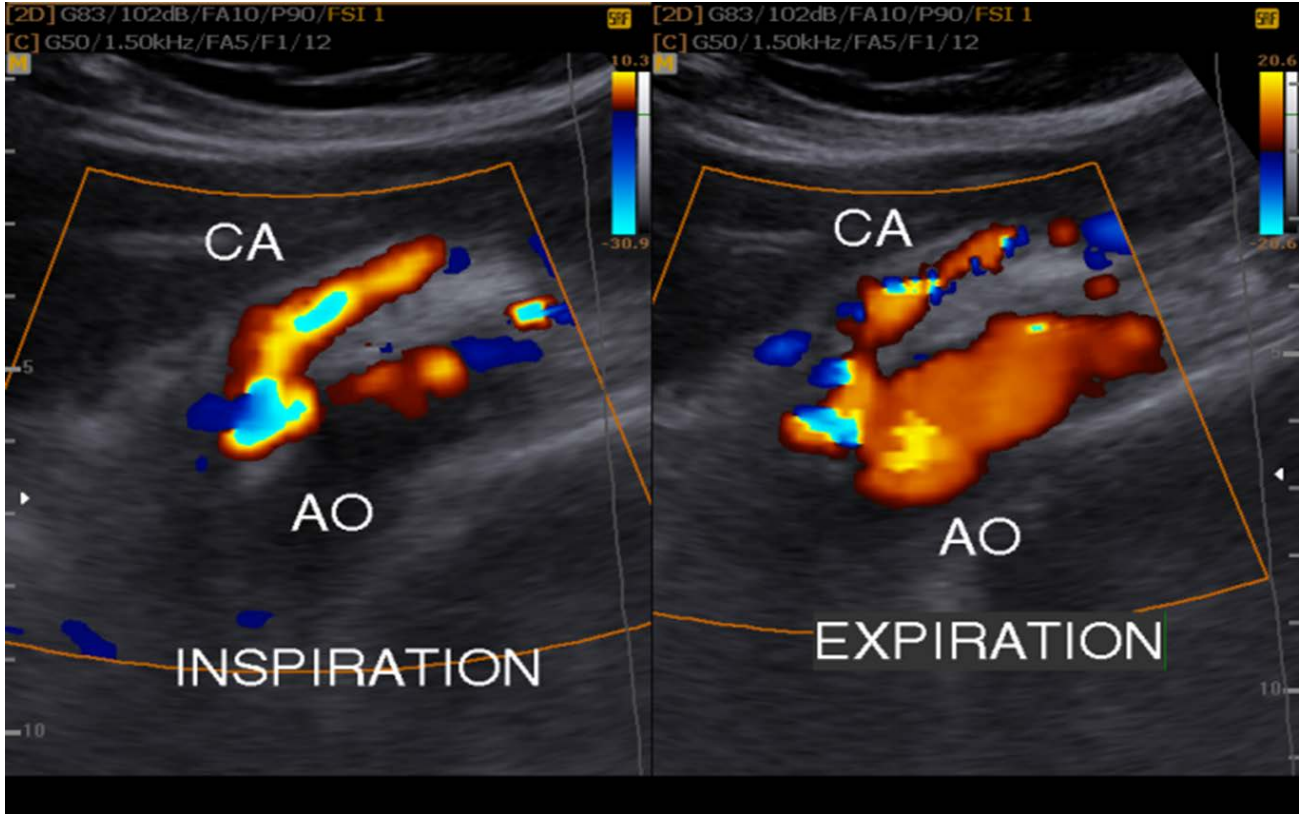
[2D] G83/102dB/FA10/P90/FSI 1
[C] G50/1.50kHz/FA5/F1/12
CA
AO
INSPIRATION
[2D] G83/102dB/FA10/P90/FSI 1
[C] G50/1.50kHz/FA5/F1/12
CA
AO
EXPIRATION

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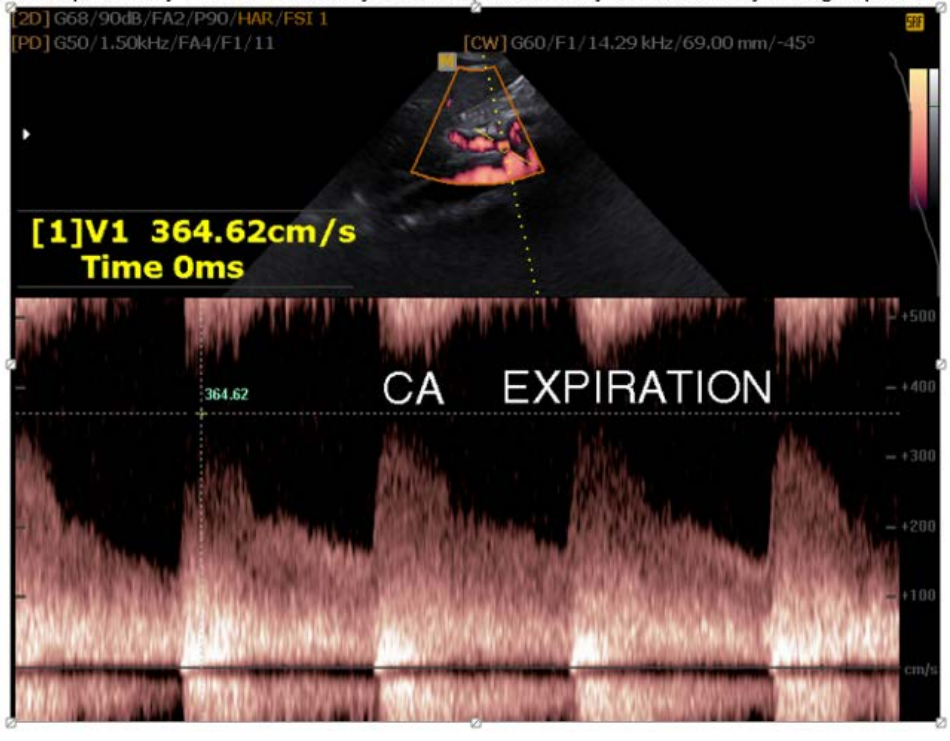
Figure 3B. Color duplex study revealed markedly elevated peak velocity of celiac artery during expiration



Ashraf Talaat Youssef et al. Evaluation of Asymptomatic Patients with Median Arcuate Ligament Syndrome (Mals) Using Color Duplex Ultrasound and Computed Tomographic (Ct) Angiography. American Journal of Cardiovascular Disease Research, 2013, Vol. 1, No. 1, 7-11. doi:10.12691/ajcdr-1-1-2

CTA of MALS

- Narrowed Celiac Axis
- Post stenotic dilation
- Hooked proximal Celiac artery after stenosis
- Vessel collaterals
- Protocol varies, need 1mm recons, but arterial phase done in expiration, venous or delayed in normal respiration
- Need thin MIPS for diagnosis and stenosis, thick MIPS for collaterals

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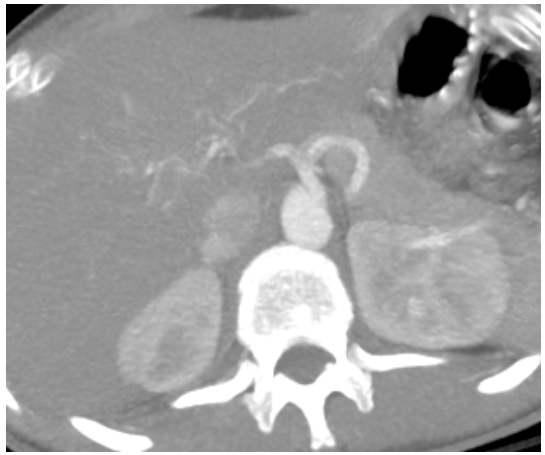
Motion artifact full expiration



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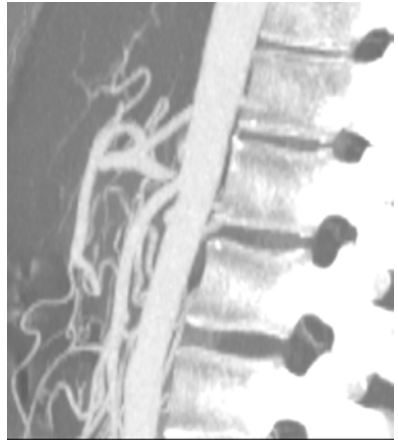
Same Patient in earlier slides – thin MIPS

High Grade stenosis of the Celiac trunk



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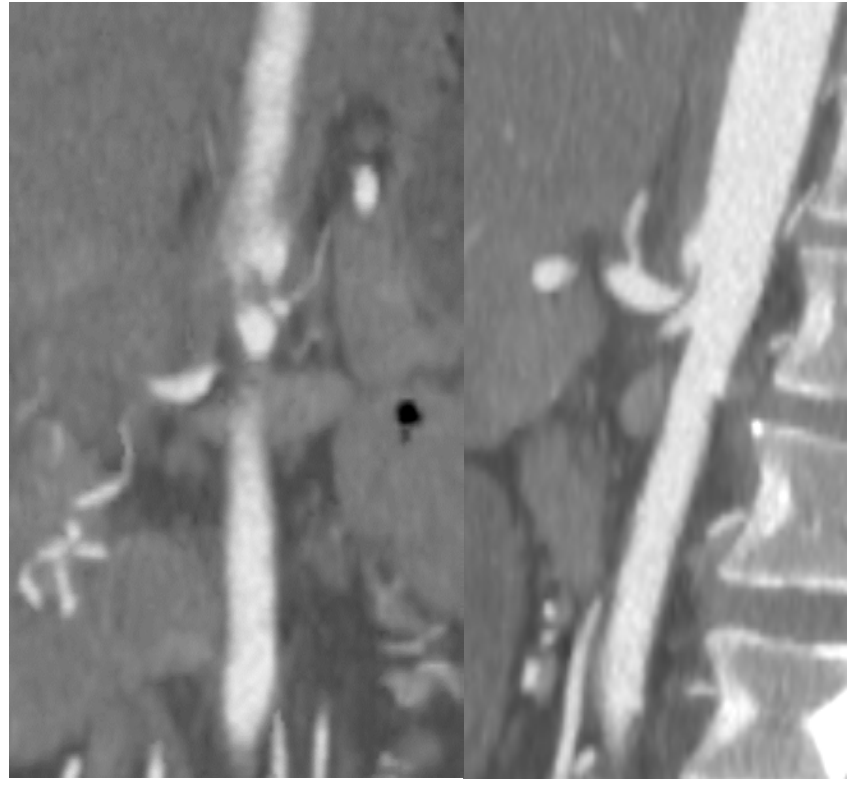
Same Patient – thin MIPS

High Grade stenosis of the Celiac trunk

J BARD MRI

Same Patient – thin MIPS

High Grade stenosis of the Celiac trunk



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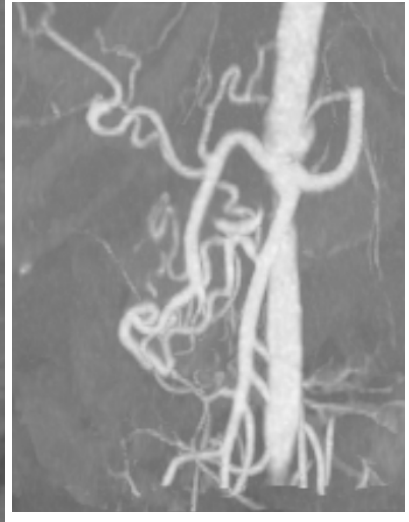
MALS - Collaterals

- SMA to Gastroduodenal Collaterals
- Same patient

J BARD MRI

MALS - Collaterals

- SMA to Gastroduodenal Collaterals
- Same patient



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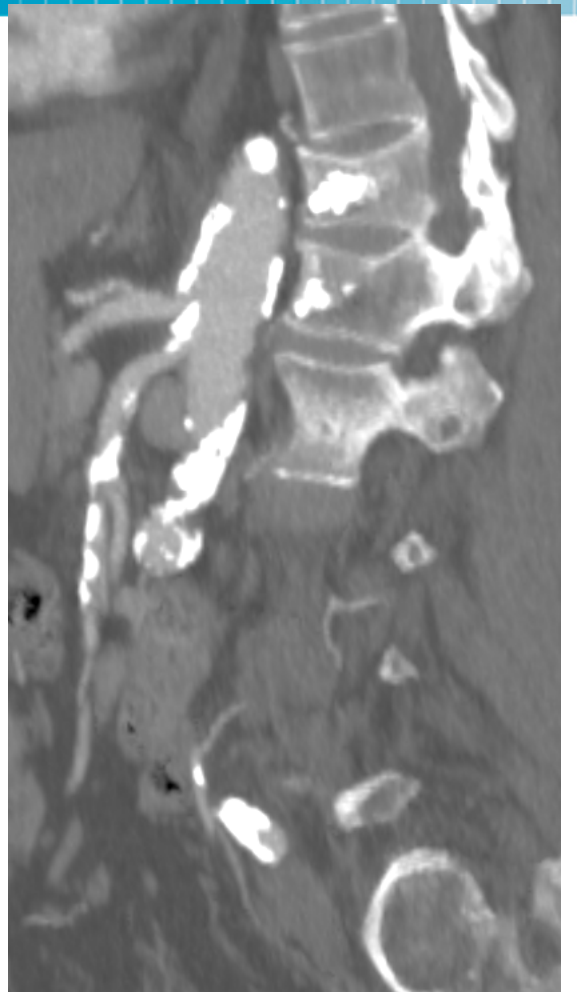
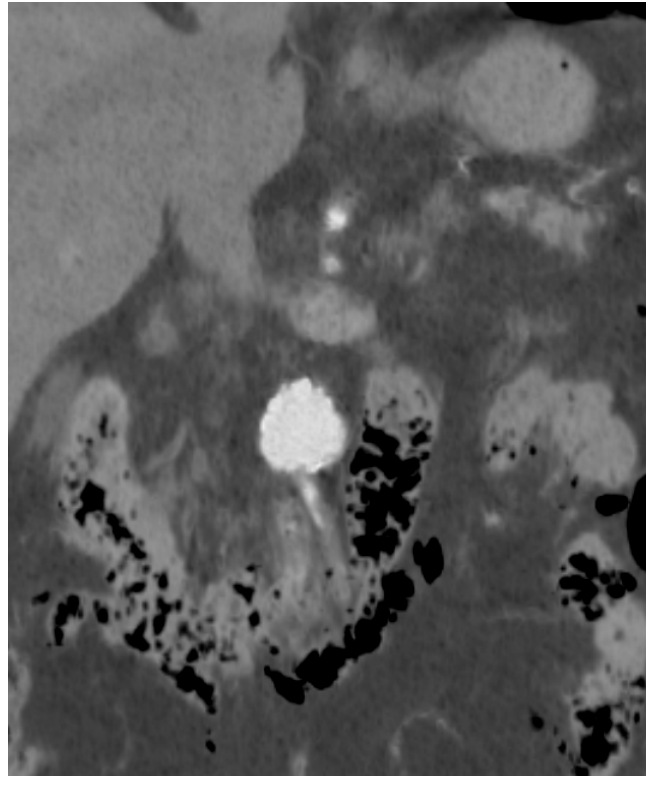
Atherosclerotic Disease





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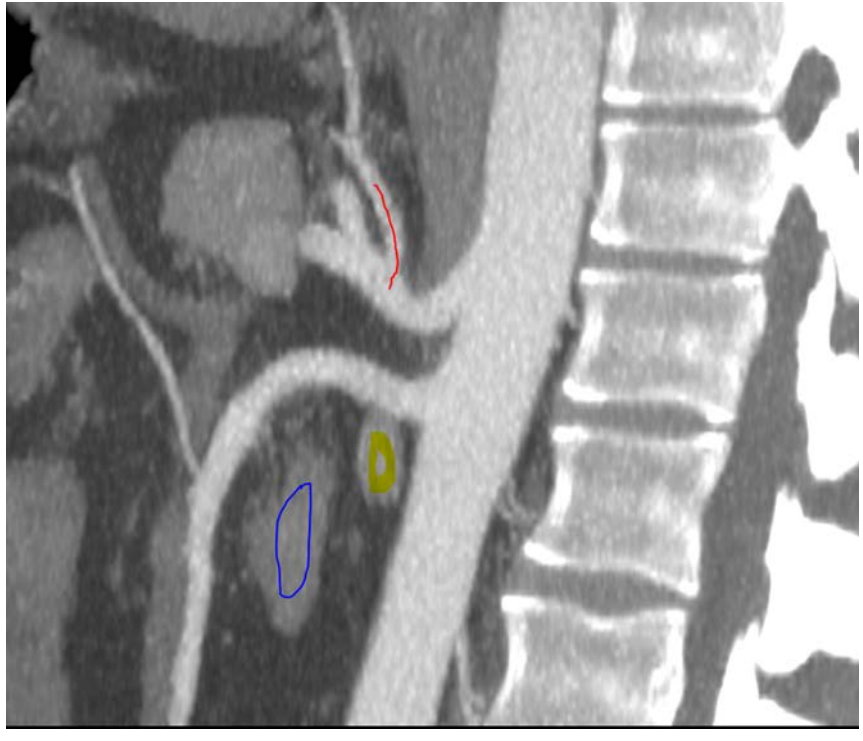
Second Year Case



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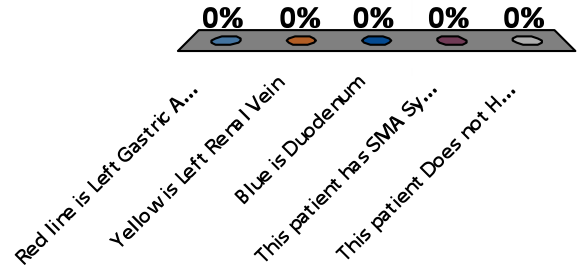
Which of the following is false?

A. Red line is Left Gastric Artery
B. Yellow is Left Renal Vein
C. Blue is Duodenum
D. This patient has SMA Syndrome
E. This patient does not have MALS



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SMA Angle

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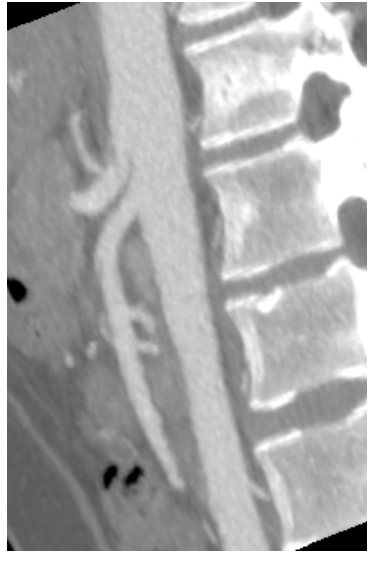
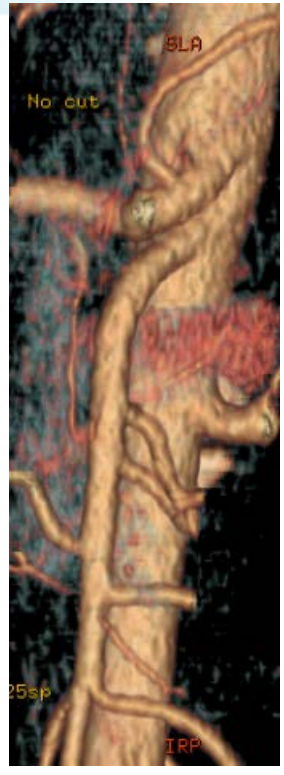
Nutcracker phenomenon

- Congestion of left renal vein from SMA and Aorta
- Some have urinary symptoms
- Pelvic congestion syndrome cause?
- Renin angiotensin system activation
- Treatment

Is this MALS?





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Challenges to MALS diagnosis and management

- Defining Significant stenosis on CTA
- Excluding other diagnoses (Nutcracker, SMA syndrome)
- Recurrent symptoms after laparoscopic surgery
- Recurrent stenosis
- Repeat CTAs are done only on patients with recurrent symptoms
- Here are some failures

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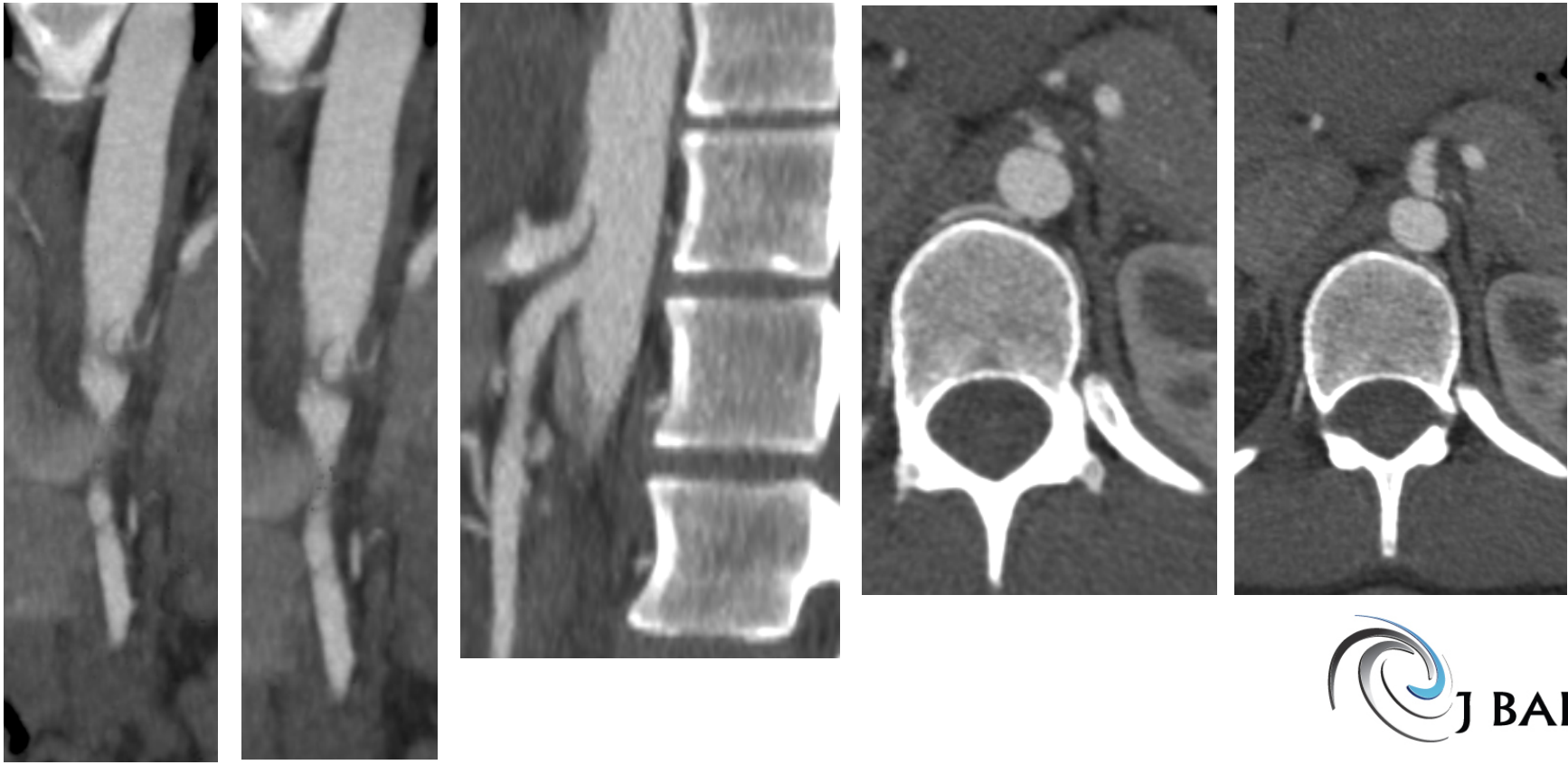
Initial study March 2015 pre-surgery



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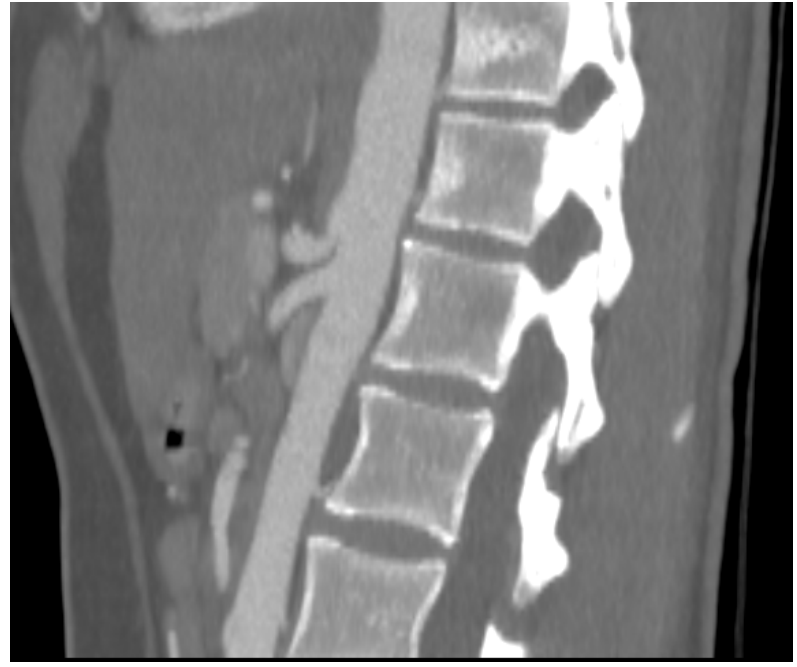
Post Surgery



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Preoperative MALS

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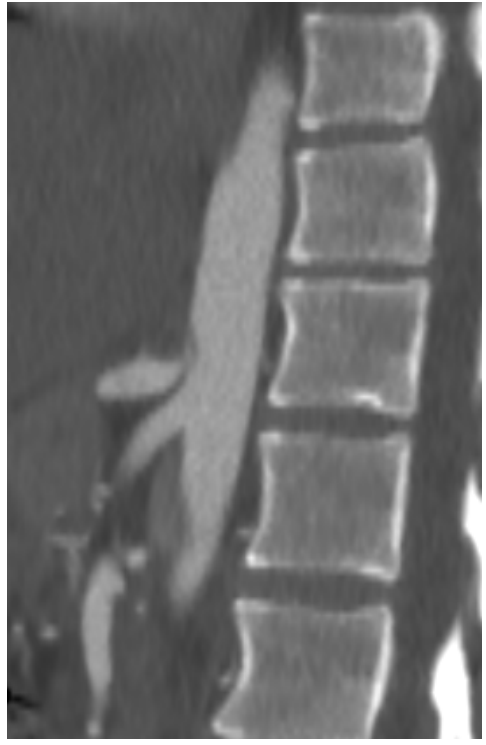
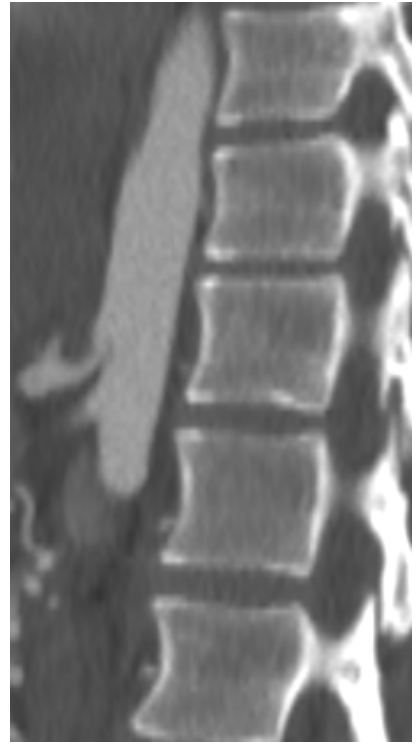
Recurrent post op stenosis





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4th Year case - Pain

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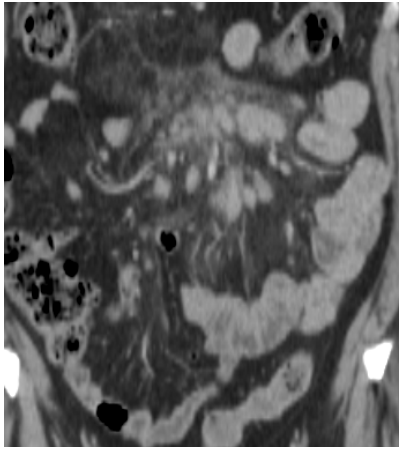
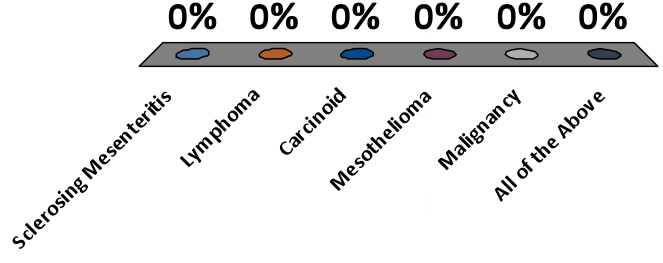
Differential?

A. Sclerosing Mesenteritis
B. Lymphoma
C. Carcinoid
D. Mesothelioma
E. Malignancy
F. All of the above





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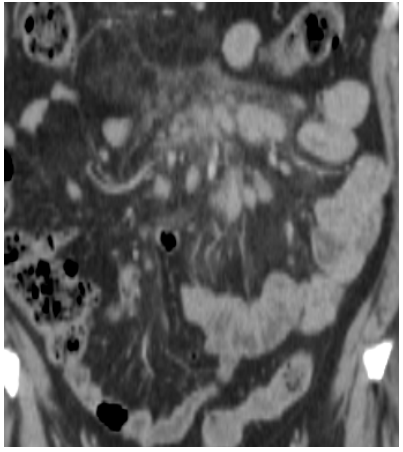
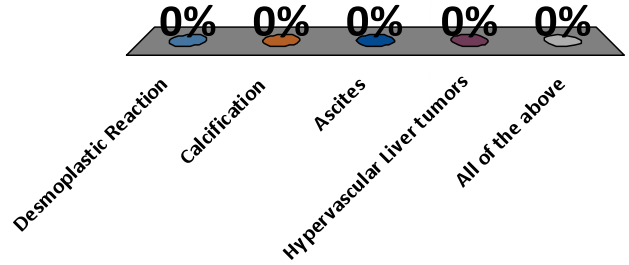
What would aid in narrowing the differential?

A. Desmoplastic Reaction
B. Calcification
C. Ascites
D. Hypervascular Liver tumors
E. All of the above





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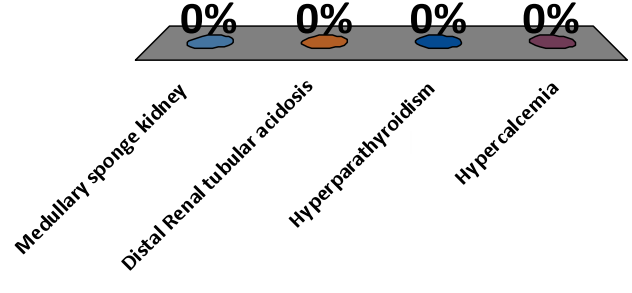
First Year –
What Is the most common cause of this finding?

A. Medullary sponge kidney
B. Distal Renal tubular acidosis
C. Hyperparathyroidism
D. Hypercalcemia



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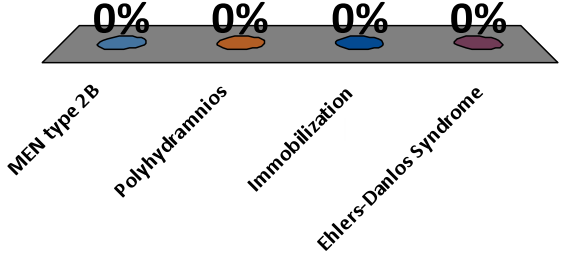
With What is this finding NOT associated?

A. MEN type 2B
B. Polyhydramnios
C. Immobilization
D. Ehlers-Danlos Syndrome



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Ehlers Danlos syndrome Q79.6

- Inherited Connective tissue disorder
- 6 major types (consolidated from 10)
  - Multiple genes, can be x-linked or Dominant or Recessive
- 1 in 2,500-5,000 people.
- Type 3 Hypermobility type most common

  Joint instability, MSK Symptoms, early arthritis

  Also Mitral valve prolapse, Ascending Aortic Rupture

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EDS Type 3

- 1 in 10-15,000, most common subtype
- Scoliosis
- Fragile skin
- Nerve compression disorders
- Hiatal Hernia
- Ascending Aorta Rupture
- Raynaud's Phenomenon
- Postural orthostatic Tachycardia Syndrome

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POTS
(Postural Orthostatic Tachycardia Syndrome)

- Increase of at least 30 bpm when going from supine to upright.
- No orthostatic hypotension
- Blood pressure does not drop
- Chronic fatigue syndrome – up to 50 % may have POTS
- EDS type 3 patients have POTS up to %40 comorbidity
- Theory – Underlying Dysautonomia

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POTS
(Postural Orthostatic Tachycardia Syndrome)

- Theory– Underlying Dysautonomia
- Hyper adrenergic POTS – hypovolemia or autonomic neuropathy
- Neuropathic POTS – Denervation of sympathetic nerves, impaired constriction of vessels
- Causes
  - Genetics
  - Post viral illness or parasitic infection, Lyme, MS, Lupus
  - Many others
  - Syrinx – Chiari type 1

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Symptomatic but no stenosis

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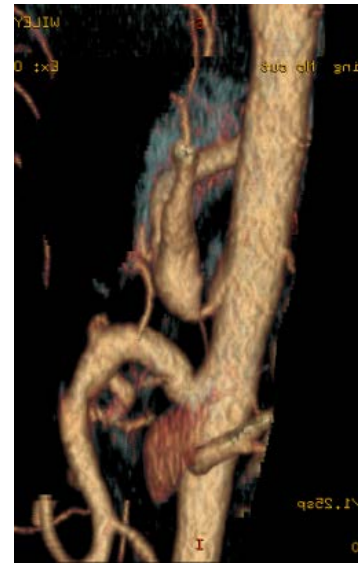
Recurrent Stenosis Post operative



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Summary

- MALS Diagnosis is not as straightforward as we thought
- Treatment failures cause reevaluation of imaging studies
- Is there something on the images that can guide management and alter the outcome? I suspect so
- Data are still being collected – 60 patients so far, 65 studies total

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THANK YOU