Cardiovascular Diseases in Athletes

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Disclosures: None

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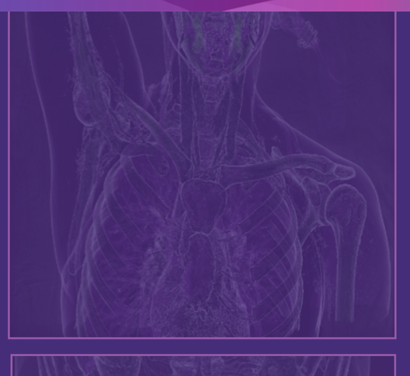
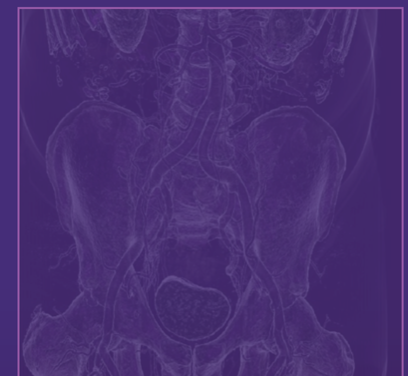
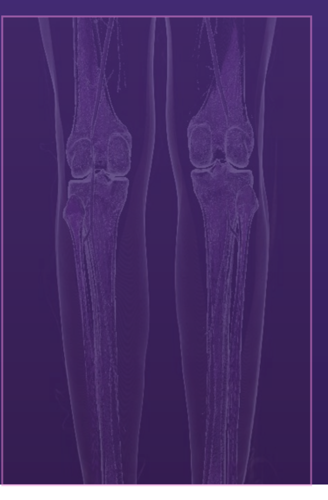
Questions – xraydoc97@yahoo.com

Background…….

- In athletes, MSK injuries are most common causes for pain
- Uncommon CV disorders may present with similar signs and symptoms
- Cardiac and Vascular diagnoses are easily overlooked
  - Young, healthy pts
  - May have plausible MSK diagnosis
  - Vascular history & physical exam needed
- Especially consider if chest pain, parasthesias, limb swelling, early fatigue, skin changes, evidence of emboli

Cardiovascular Diseases in Athletes

- Causes of Sudden Cardiac Death
  - Hypertrophic Cardiomyopathy
  - Malignant Coronary Anomalies
- Upper Extremity
  - Thoracic Outlet Syndrome (TOS)
- Pelvis
  - Iliac Endofibrosis (IE)
- Lower Extremity
  - Popliteal Entrapment Syndrome (PAES)

Cardiovascular Diseases in Athletes

- Causes of Sudden Cardiac Death in Athletes
  - **Hypertrophic Cardiomyopathy**
  - **Malignant Coronary Anomalies**

Sudden Cardiac Death in Athletes: Incidence: 1-2/100,000 athletes per year

Disease	Prevalence	Description	Imaging of choice
Hypertrophic Cardiomyopathy	1/500	A/D, myofibrillar disarray Septal, apical, diffuse variants	ECG, Echo, MRI
Anomalous Coronary artery	1/1000	Inter-arterial course = malignant	CTA, MRI
Commotio Cordis	?	Blunt chest trauma during T wave upstoke → VF	ECG, Echo, CCTA
Myocarditis	?	Myocardial inflammation #1 = viral	ECG, MRI Endomyocardial Bx
Long QT/short QT Brugada Syndrome	1/600-2500	Ion channelopathies Brugada = A/D	ECG
ARVD/C	1/5000	Lipofibromatous infiltration of RV > LV, arrhythmia	ECG, MRI
Marfan Syndrome	1/5000	Fibrillin gene mutation, A/D Aneurysms, rupture	ECG, Echo, CTA, MRI

Pugh, A. Postgrad Med J. 2012 Feb 23

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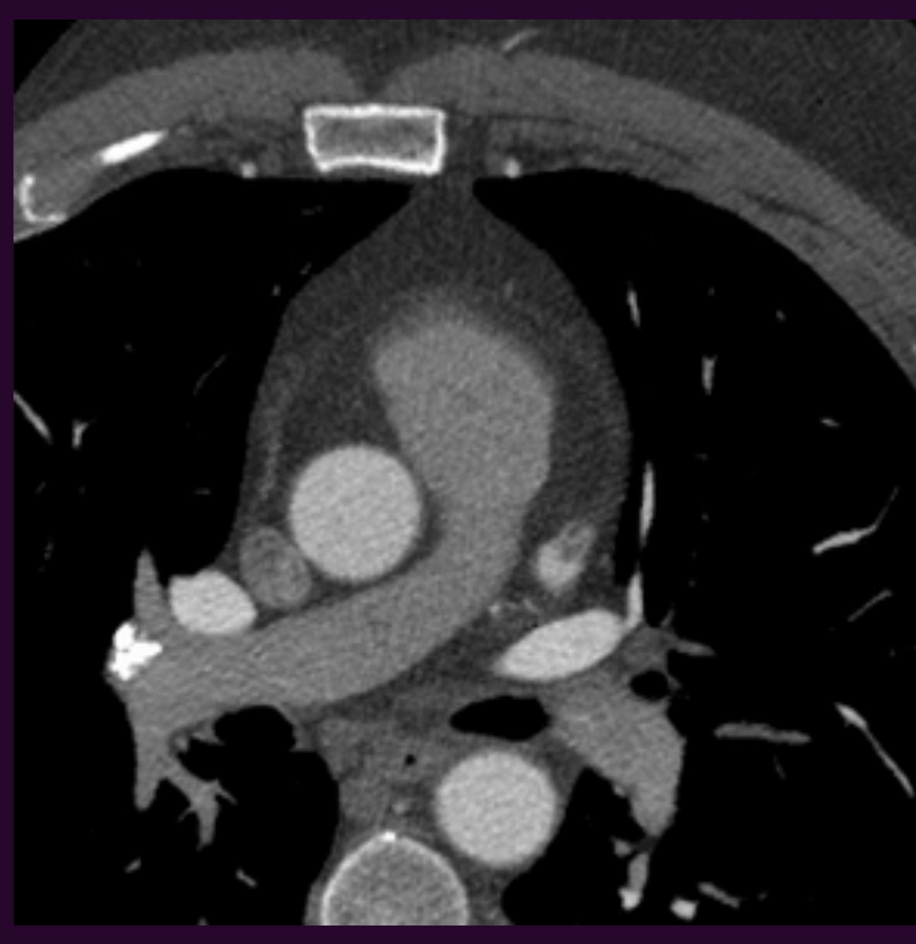
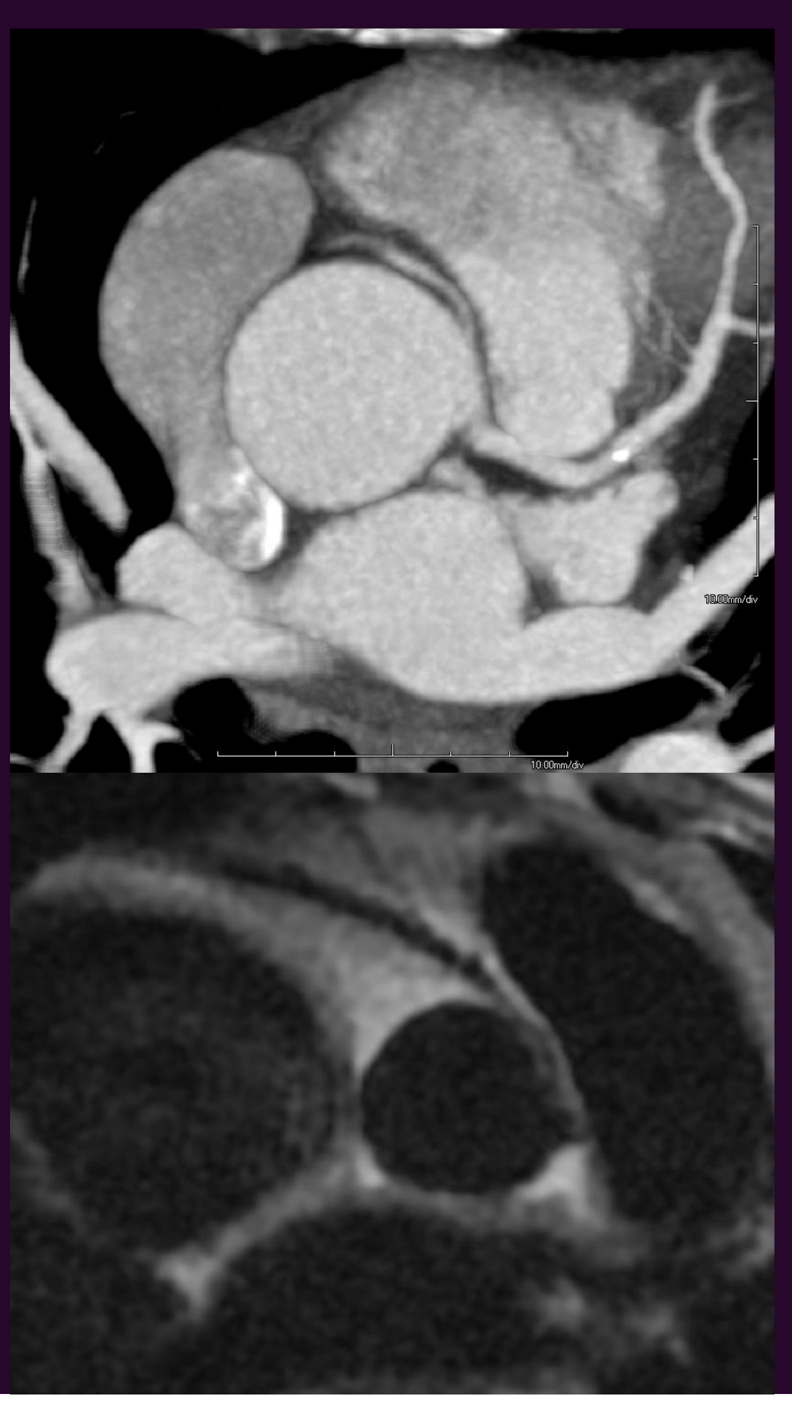
Malignant Coronary Anomalies

- Causes 5-35% of SCD in young athletes (#2)
- #1 malignant anatomy= Origin from Opposite Sinus with Inter-arterial Course
  - Transient compression causes torsion / pinching w/ exercise
  - Slit-like narrowing of ostium w/ acute angle origin
- Incidence: ~ 0.1% of population each
- In Athletes: Anomalous LEFT (57% mort.) > RIGHT (25%)

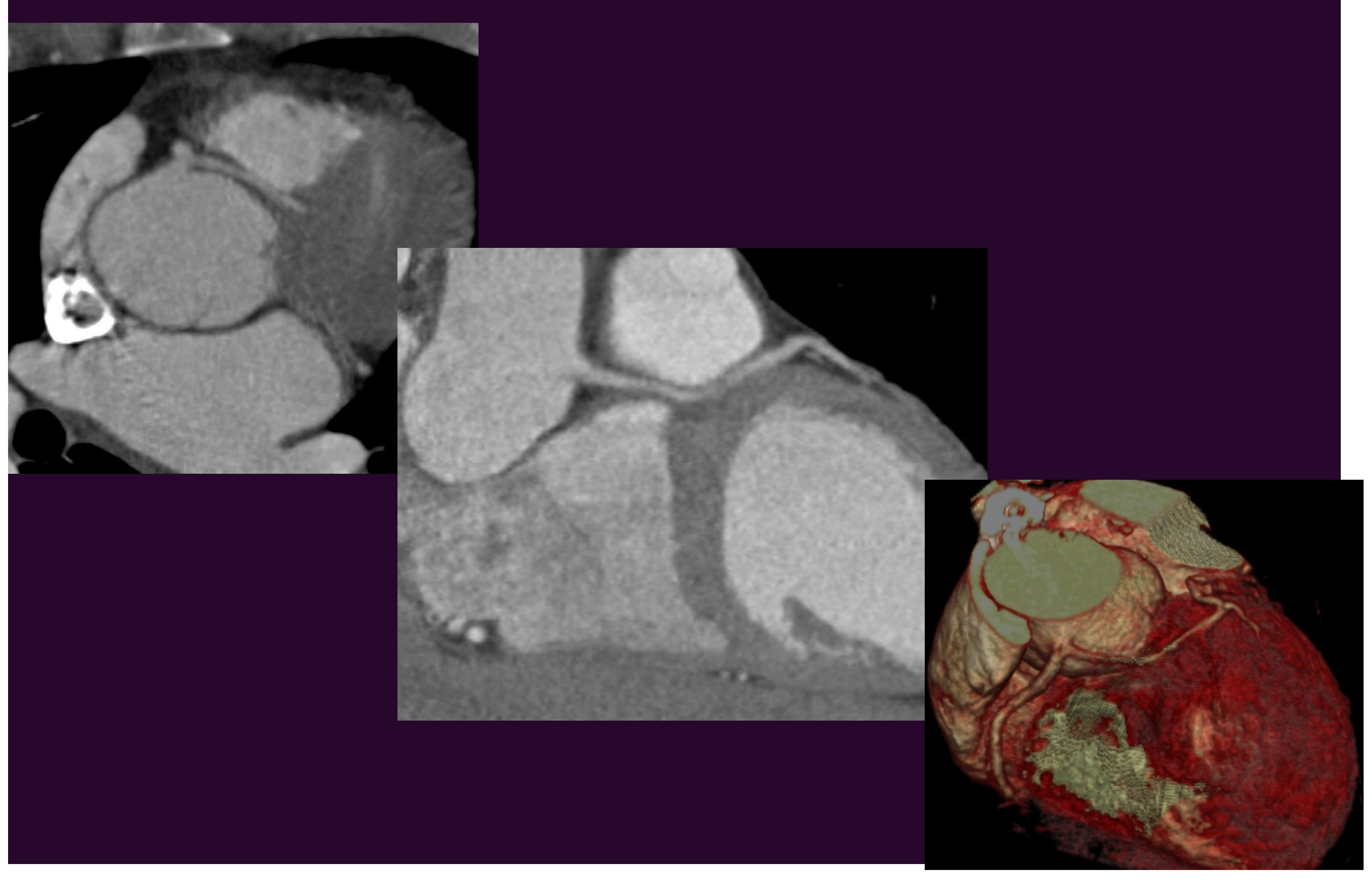
Virmani R, Burke AP, Farb A. The pathology of sudden cardiac death in athletes. chapter 15. In: Williams RA, editor. The athlete and heart disease: diagnosis, evaluation & management. Philadelphia: Lippincott Williams & Wilkins; 1999. p. 249–72

Anomalous RCA from Left Sinus

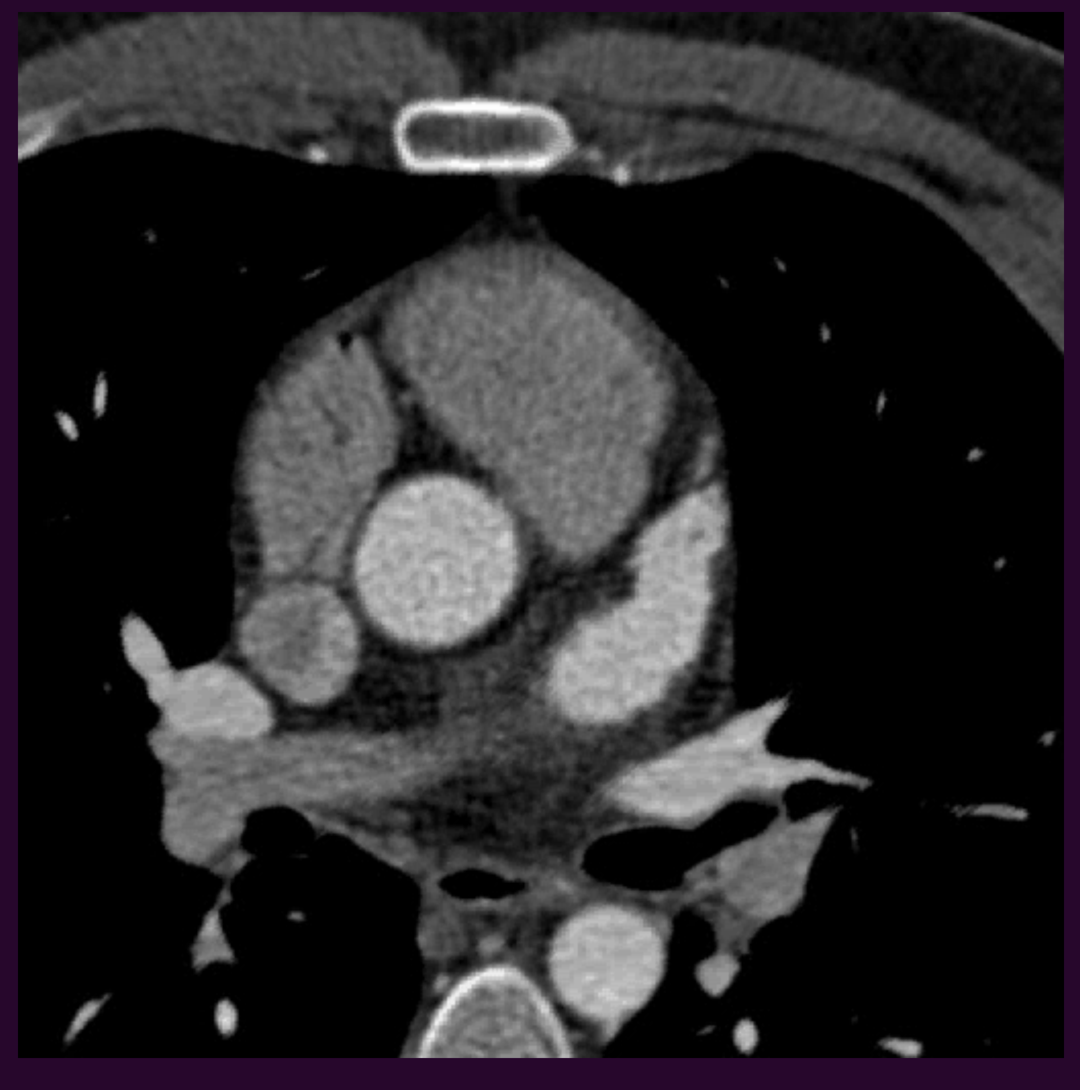


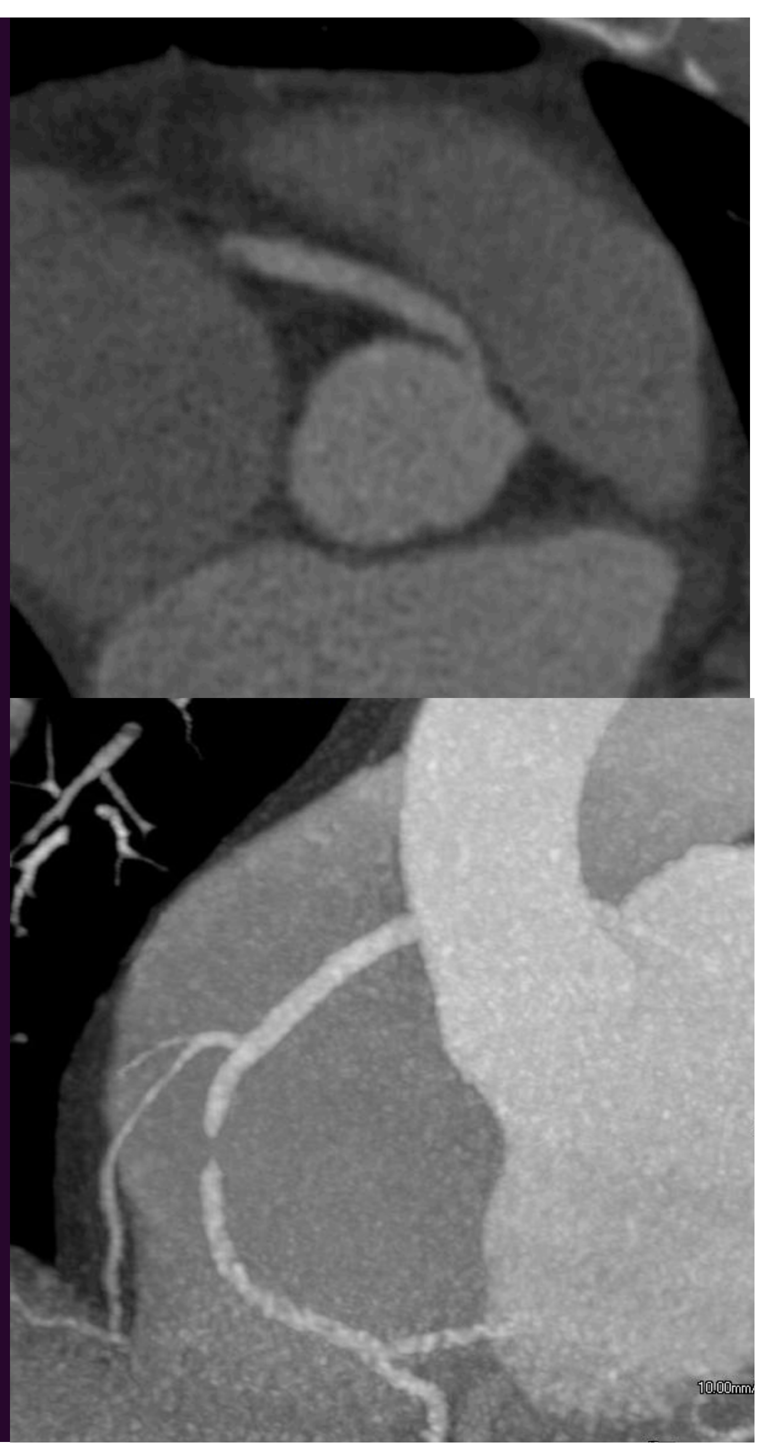


Anomalous LCA from RCA



27 yo Bullfighter, Intermittant CP





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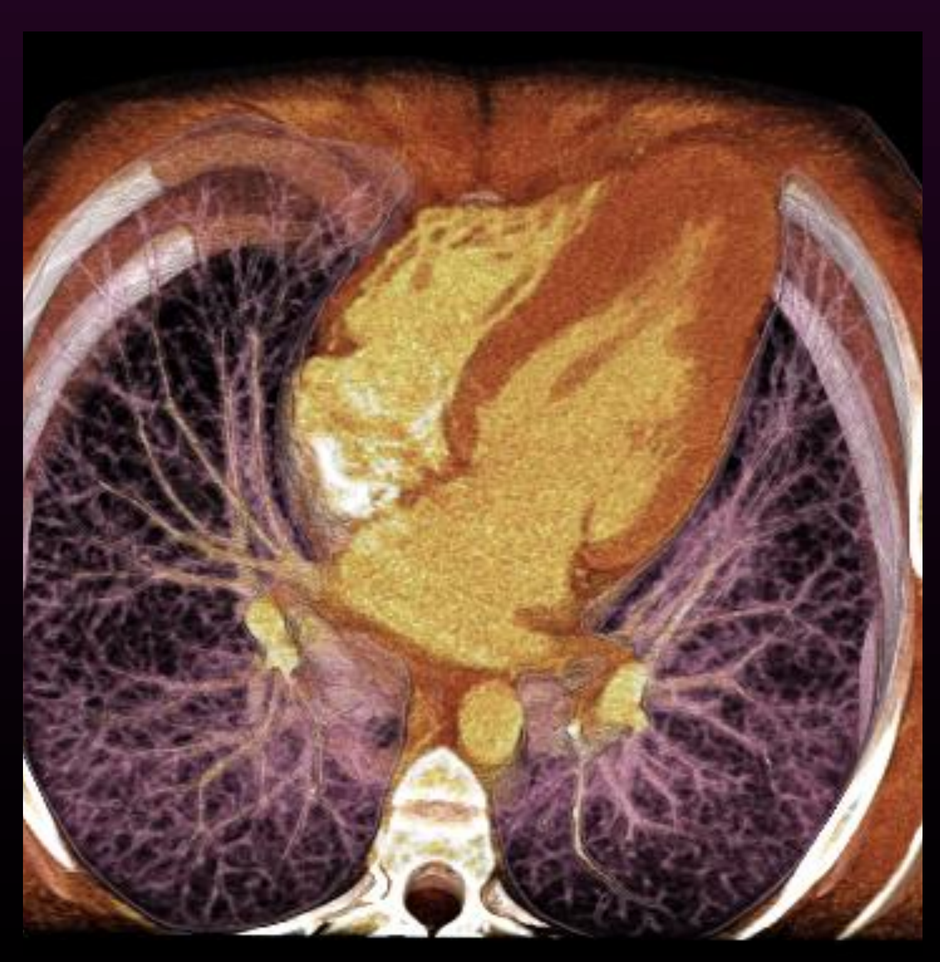
Hypertrophic Cardiomyopathy

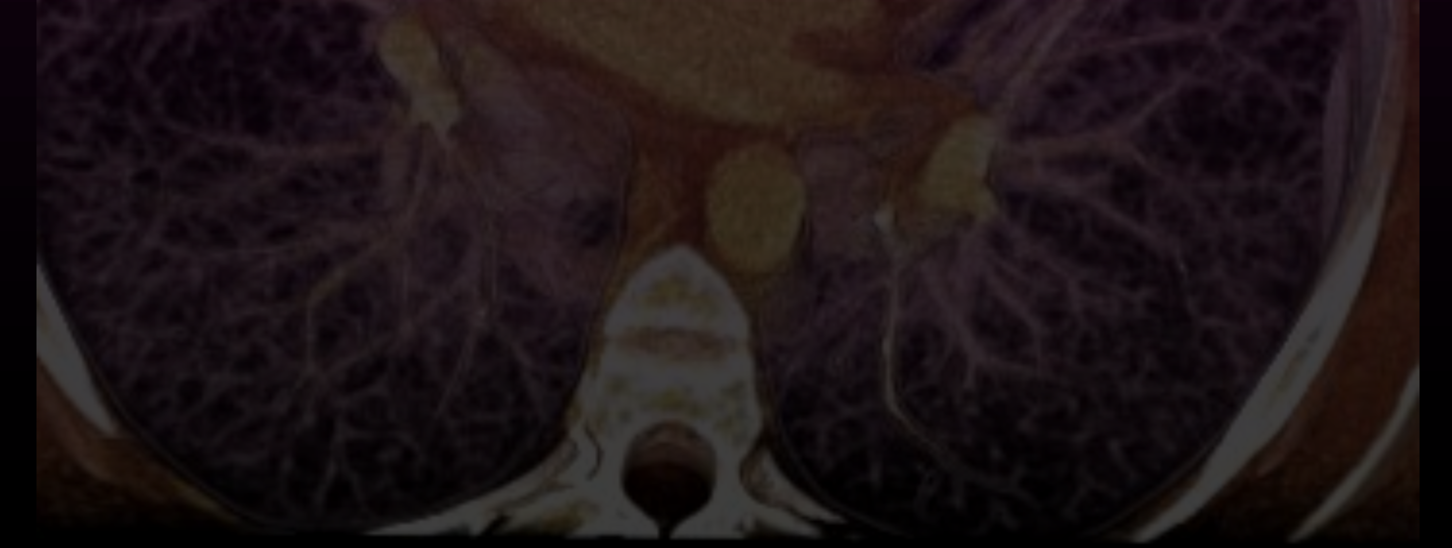


Hypertrophic Cardiomyopathy

- aka HCM, HOCM, IHSS
- Heterogeneous, usually familial (50-60%)
  - A/D inheritance
- Most common cause of sudden cardiac death in athletes and in pts < 35 y.o.

Athlete’s Heart vs. HCM

	Athlete’s Heart	HCM
ED Septum Thickness	<15 mm	>15 mm
LV symmetry	YES	NO (ASH, apical)
Family HX	NO	Possibly
Deconsitioned Response	Reduction within 3 mos.	None

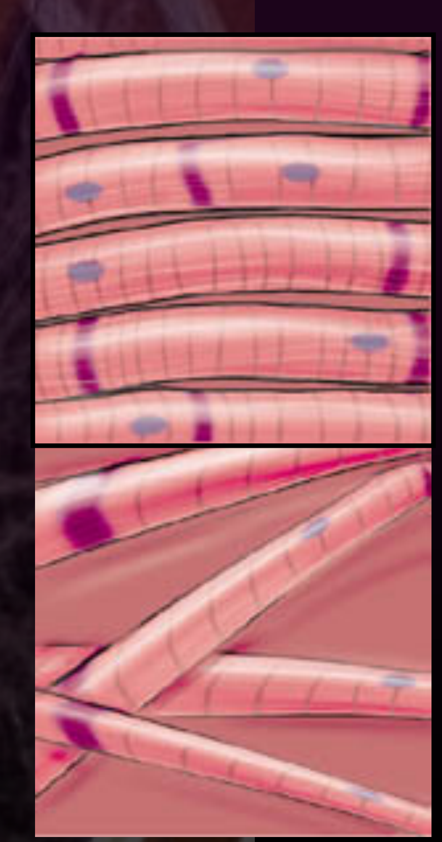


Histopathology of HCM

- Extensive disarray of hypertrophied myocardial cells
- Scarring
- Small intra-mural coronary arteries affected
- At least 10 genes involved in sarcomere protein synthesis implicated

Histology of HOCM

- Myofibrillar disarray
- Intertwined hypertrophied myocytes with bizarre-shaped nuclei
- Focal or widespread interstitial fibrosis
  - Both matrix collagen and replacement fibrosis found in increased amounts



HCM

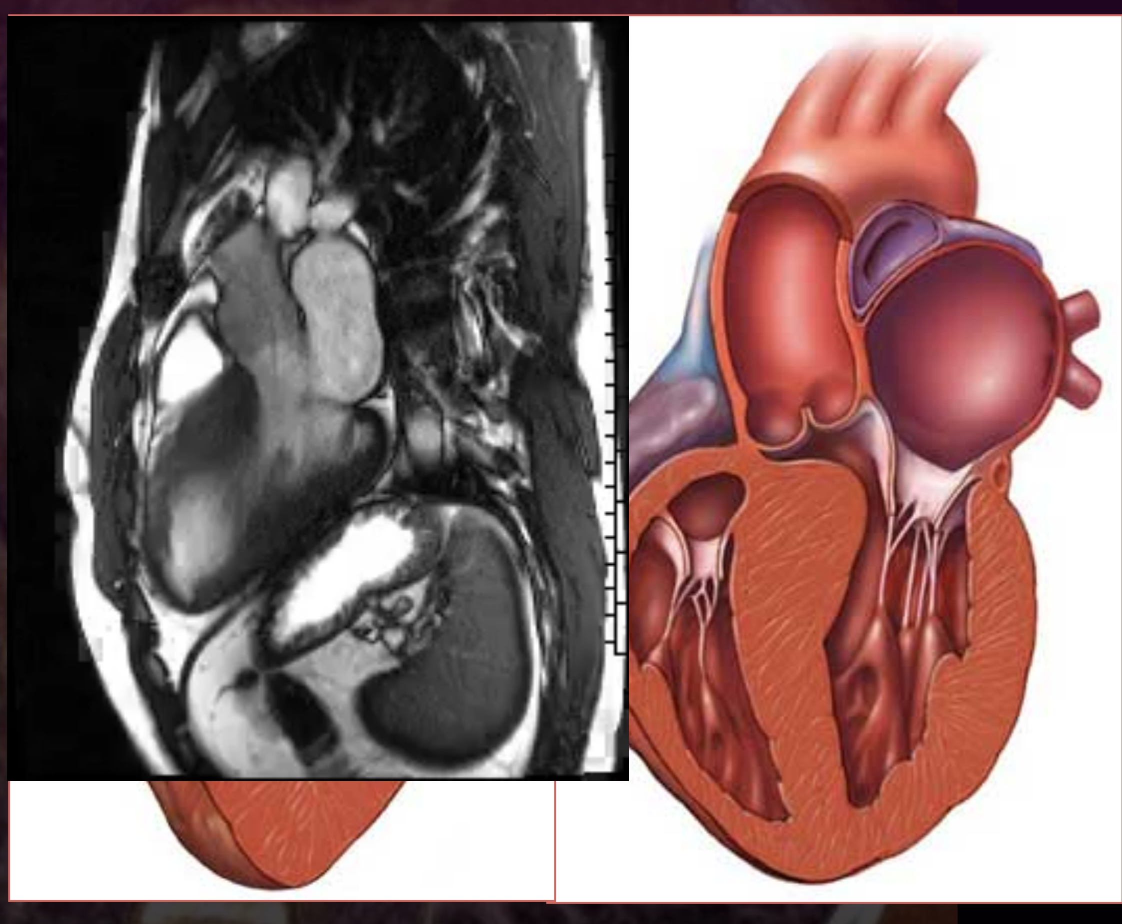
- LV hypertrophy
  - Asymmetric septal hypertrophy (ASH): 65%
  - May be concentric (25%)
  - May involve only a focal segment of septum
  - May be apical only (10%)
    - Higher incidence in Japanese
  - Enhanced contractility
- **S**ystolic **A**nterior **M**otion of mitral valve (SAM)

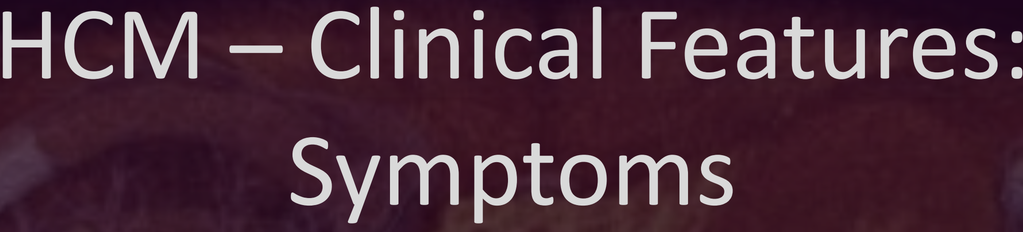
SAM

Anterior leaflet → into LVOT → **LVOT obstruction**

Venturi Effect (pulling) vs. abnormal MV leaflets and anteriorly positioned pap muscles (pushing) into LVOT

RESULT→ mitral regurg





- Dyspnea (stiff LV - diastolic dysfunction)
- Syncope (fibrosis , Fiber disarray - arrhythmias)
- Angina (intramural coronary dz - ischemia)
- LVOT obstruction (SAM)

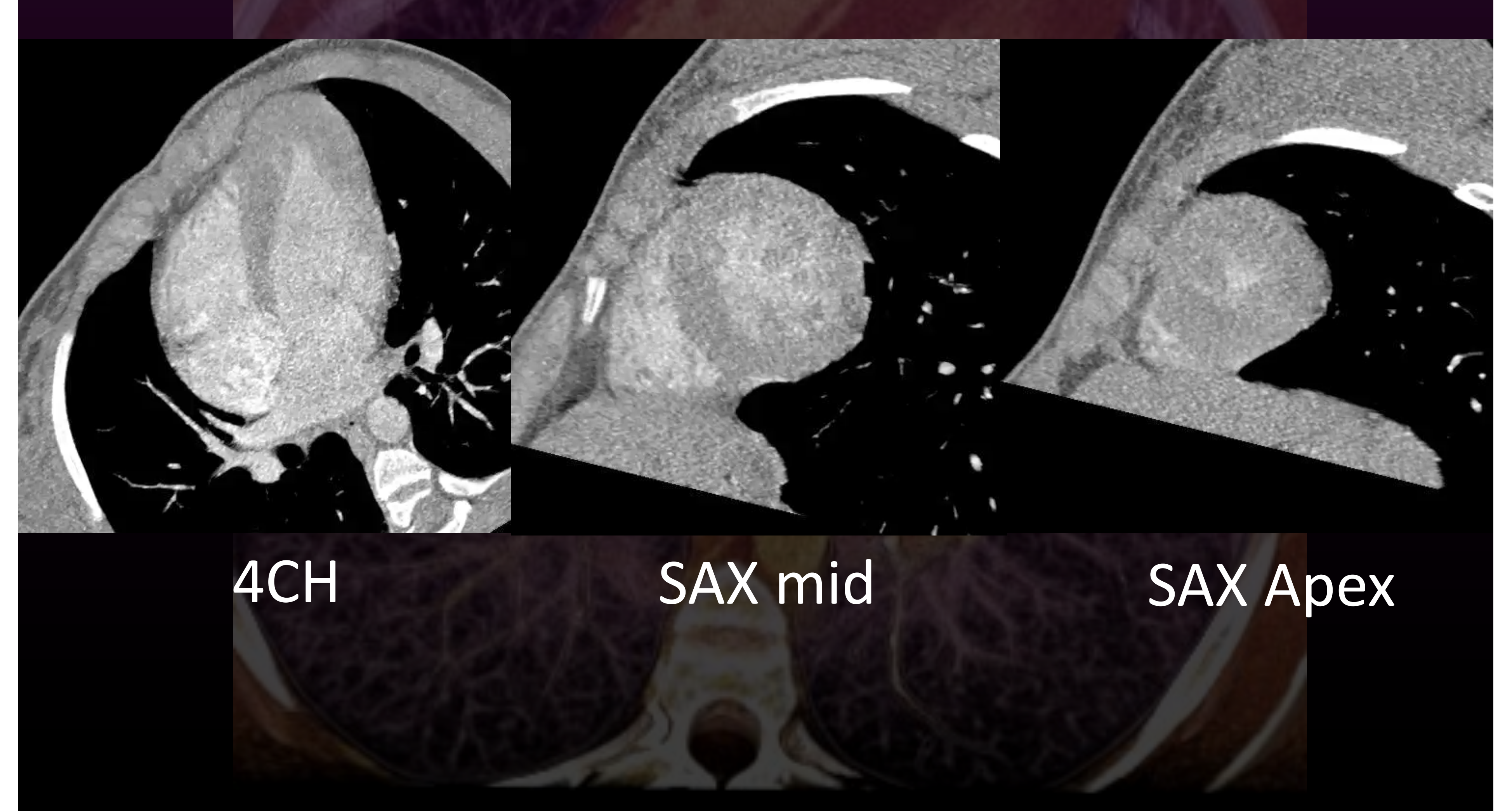
Screening for HCM

- Genetics: Complex and not cost effective for screening
  - Used for family risk assesement in clear HCM patients
- ECG: abnl. In 70 % Asx pts. but not specific
- Echo: (+) in ≥80%
  - LVH can be absent in <13 yo
- **MRI: Gold standard**

CT

- Calculate myocardial mass, define anatomy
- Contraindication to MR
- Epicardial coronaries usually large and non-stenotic
- Cine images → SAM, volumes, EF, cavity obstruction
- ? DE scar ?

CT – Apical HCM
15 yo F soccer player



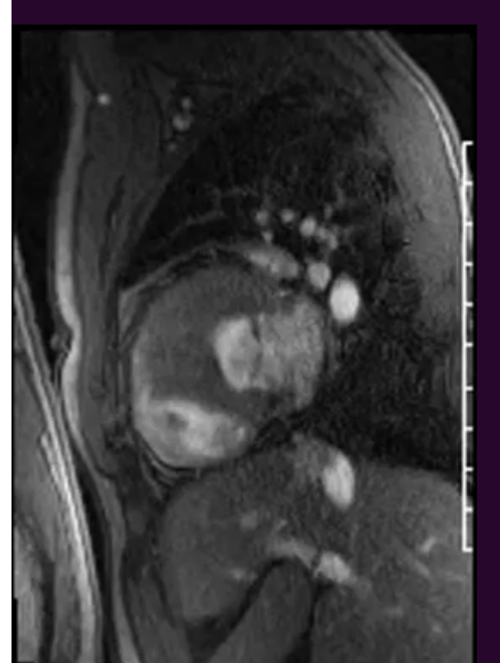
MR in HCM

- Best for quantification of myocardial mass
- Visualize SAM, calculate gradients
- Visualize scar / fibrosis (delayed enhancement)
  - May be 2^0 to fibrosis (more) or AMI (less)
    - Repetitive ischemic events?
  - Presence of DE implies poorer prognosis than lack of DE (^{201}Tl data)

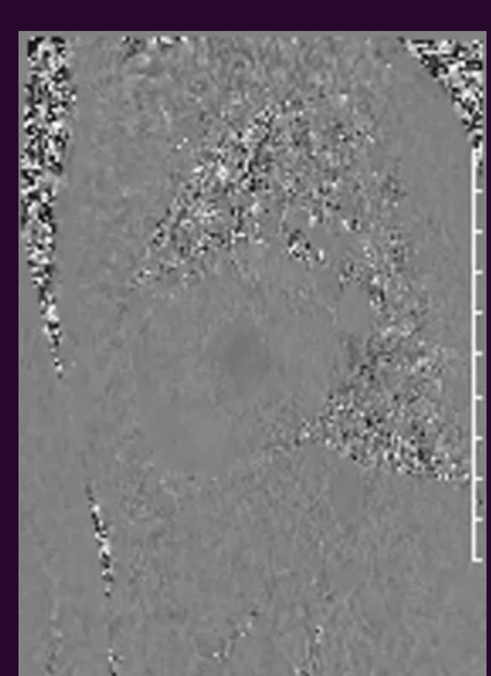
ASH – 4CH

ASH – SAX

SAM – LVOT Obstr.
3CH



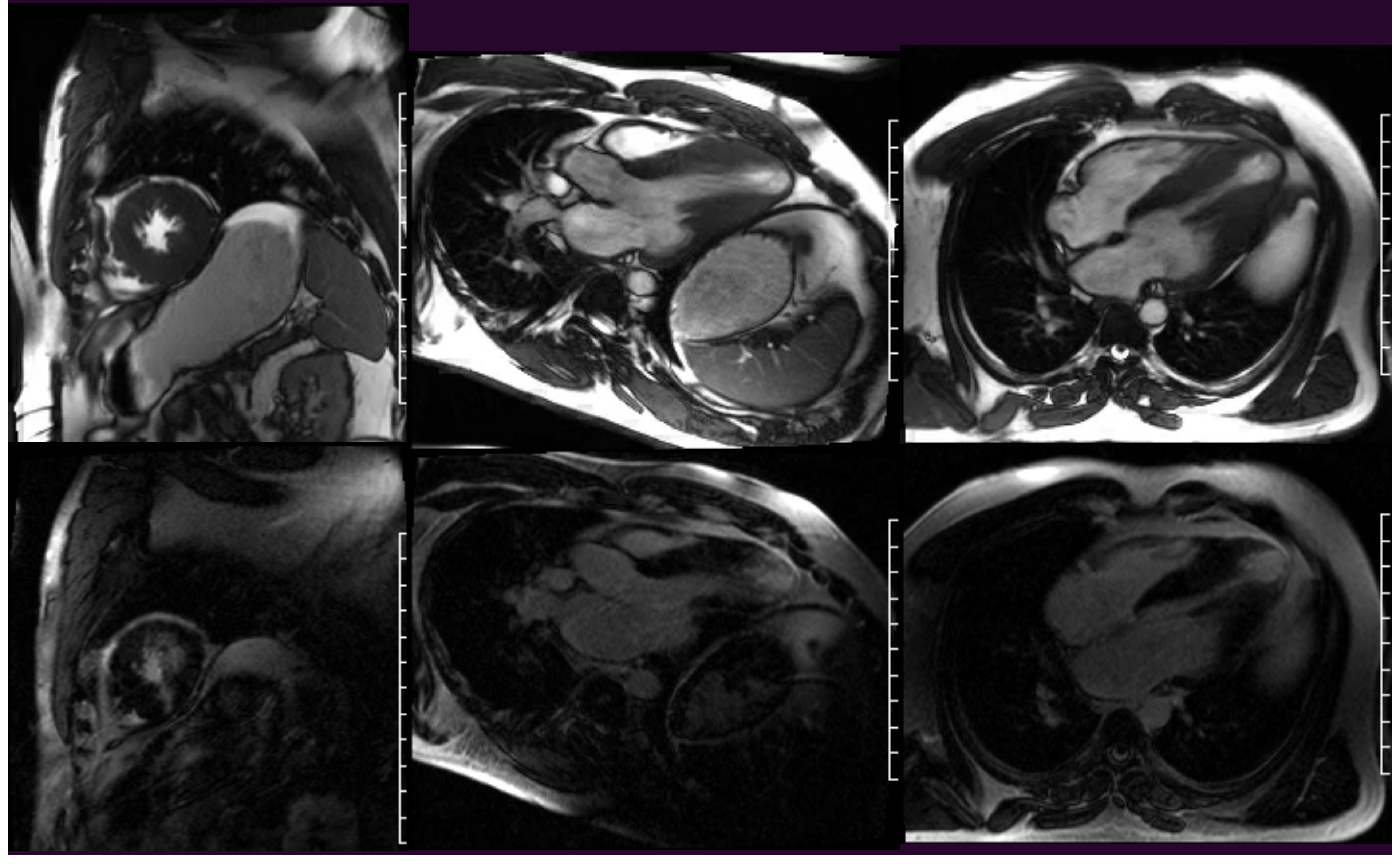
PHASE CONTRAST



27 YO male, SOB/syncope

Mid-cavity Obliteration, apical dyskinesis, DHE

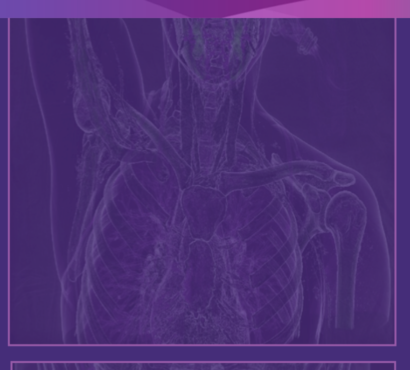
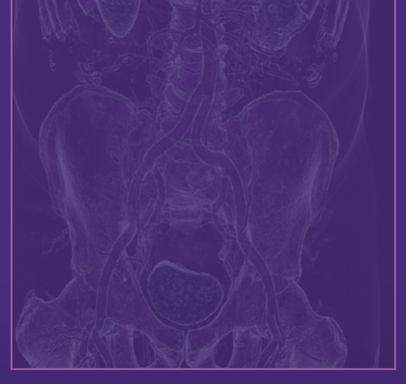
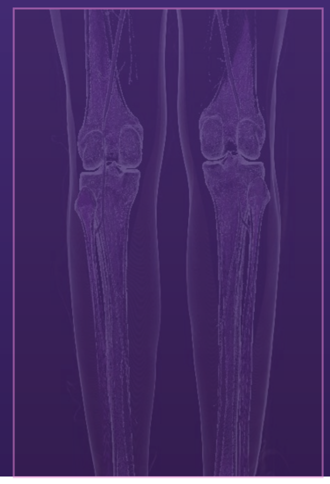
38 yo Male w/ CP



Screening for HCM in Athletes

- U.S.: No national testing
  - 15 M athletes, <100 deaths /yr (1 death / 220,000 athletes
  - AHA does not endorse
  - Many colleges / pros screen w/ Echo
- Italian model: by law all "elite" athletes screened:
  - H&P, UA, rest/stress ECG, PFT
  - Abnormals → ECHO
  - 89% decrease in SCD after screening instituted

Vascular Diseases in Athletes

- Upper Extremity
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- Pelvis
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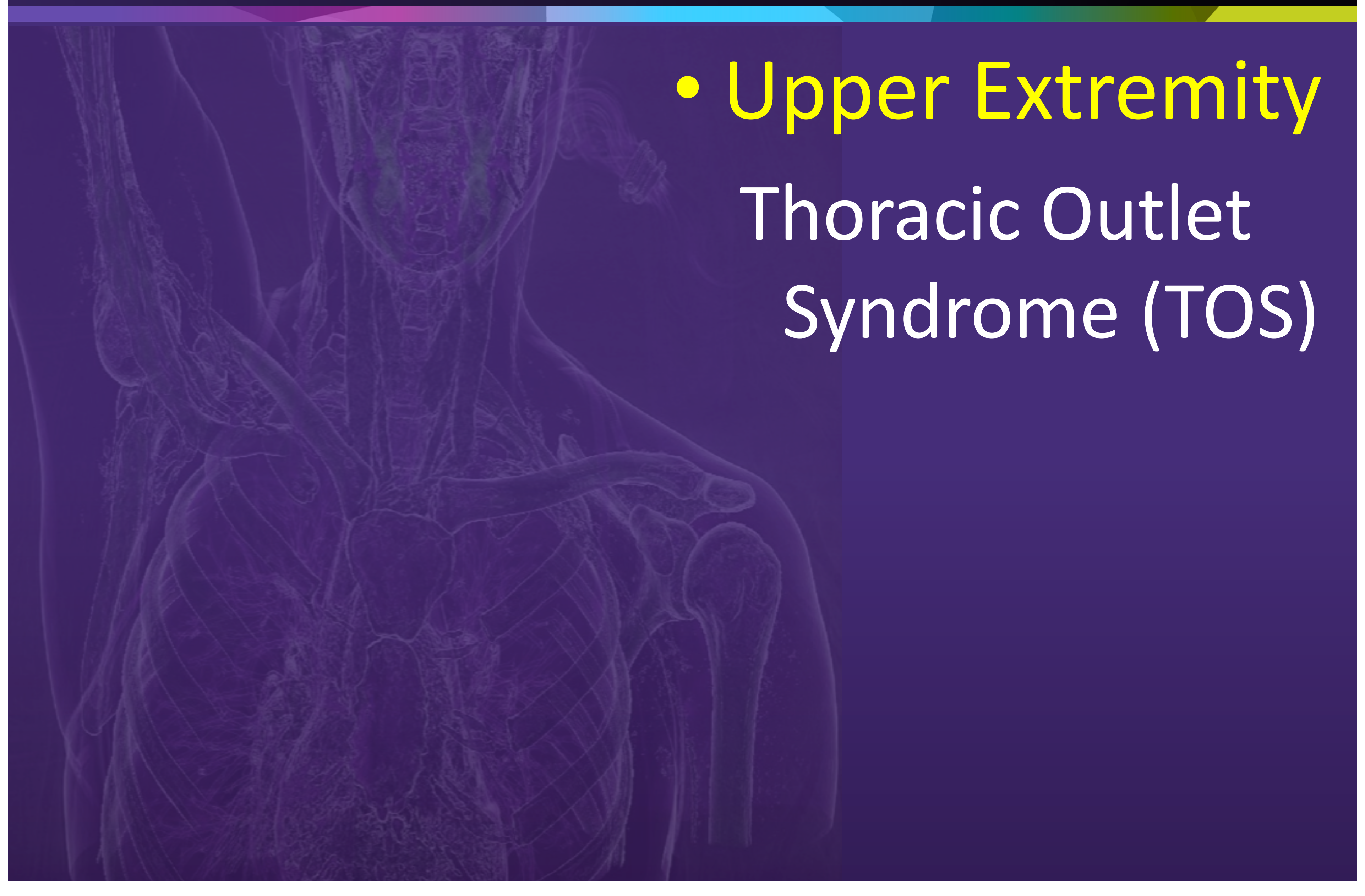
- Lower Extremity
  - Popliteal Entrapment Syndrome (PAES)

Vascular Diseases in Athletes



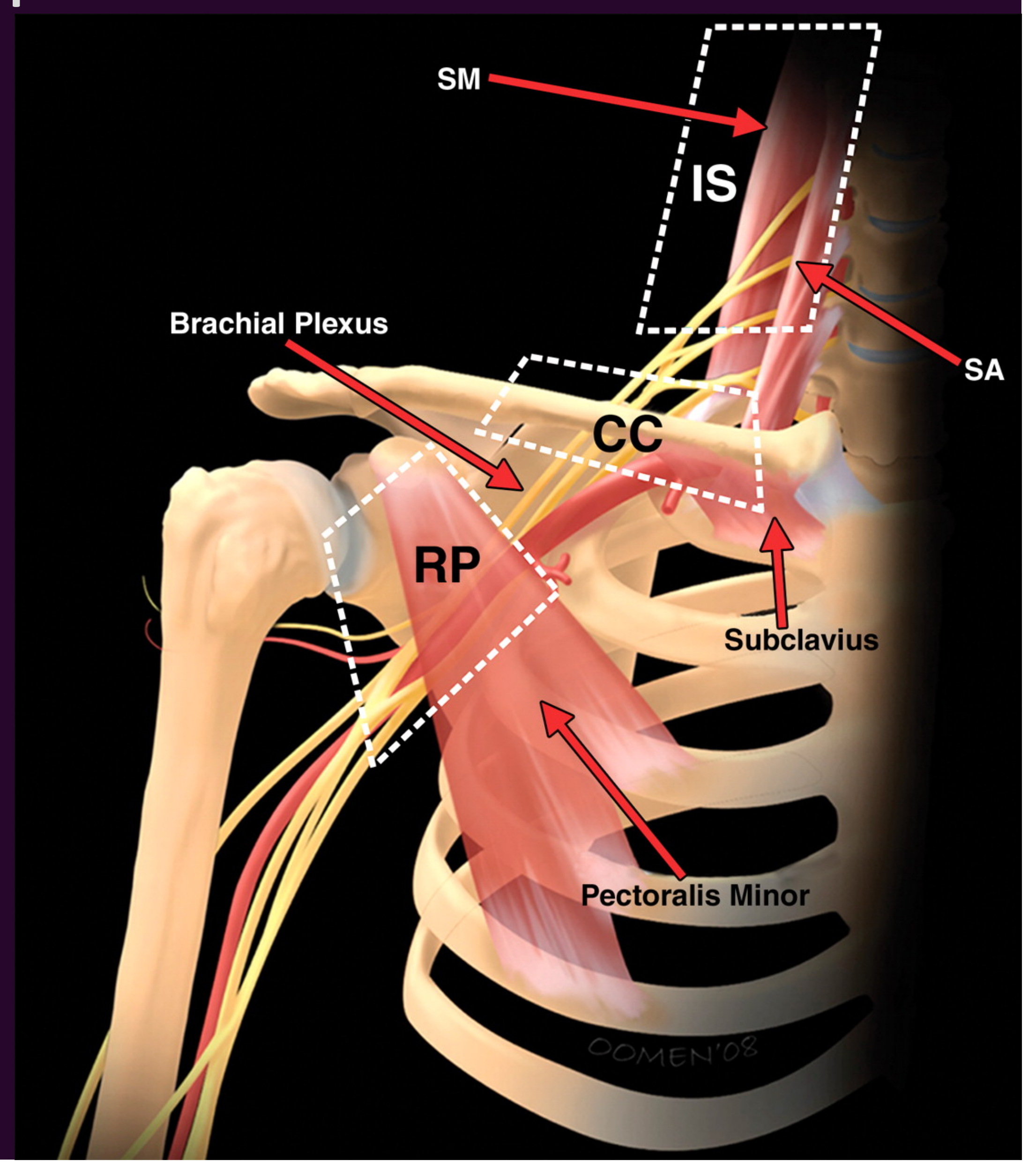
Thoracic Outlet Syndrome (TOS)

- Symptomatic extrinsic compression/ entrapment of the neurovascular **structures** of the upper extremity by bone and/or soft tissue as they pass through the cervicoaxillary canal

aka "Cervico-axillary Compression Syndrome"

Components of Cervico-Axillary Canal

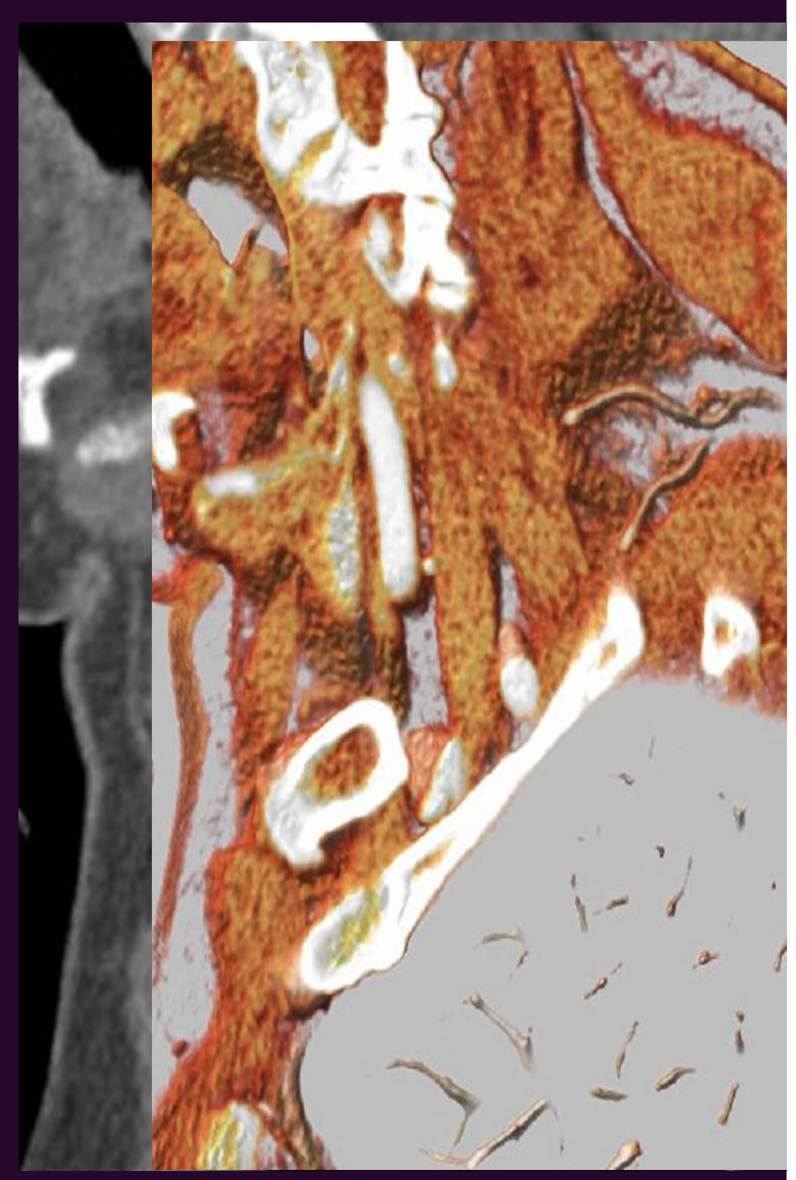
- Interscalene Triangle: #1 site of compression
- Costoclavicular space: #1 site vascular TOS
- Retro-pectoralis space: #1 site for masses



Linda DD et al. Radiographics 2010;30:1373-1400

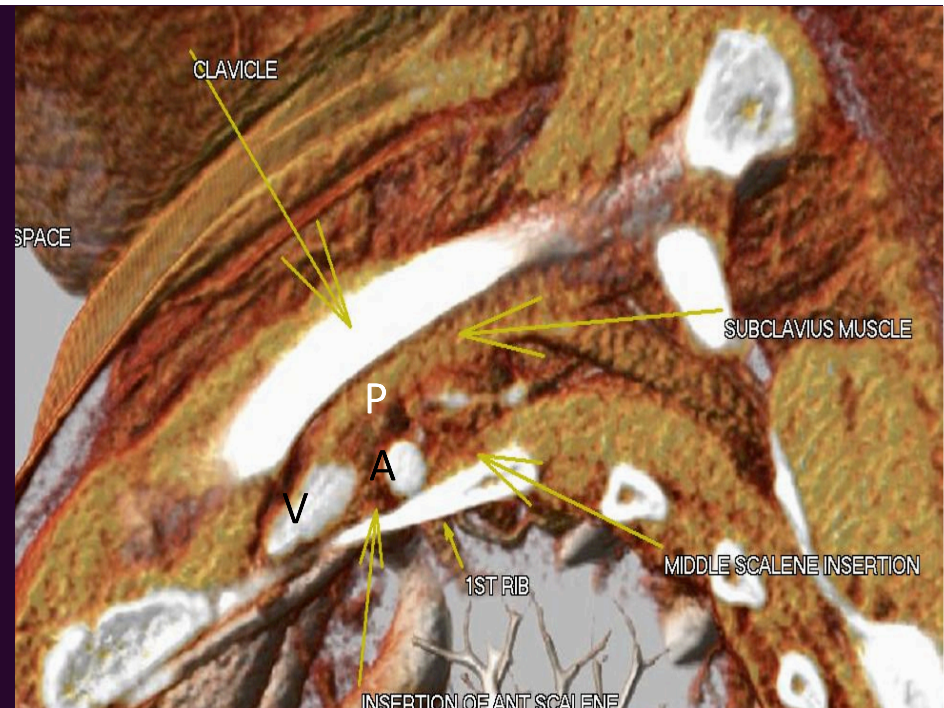
Interscalene Triangle

- # 1 site of compression
- Borders:
  - Anterior: anterior scalene
  - Posterior: middle/post. scalenes
  - Inferior: 1st rib
- Contents:
  - Subclavian Artery
  - Brachial Plexus (3 trunks)
  - NOT subclavian vein !!



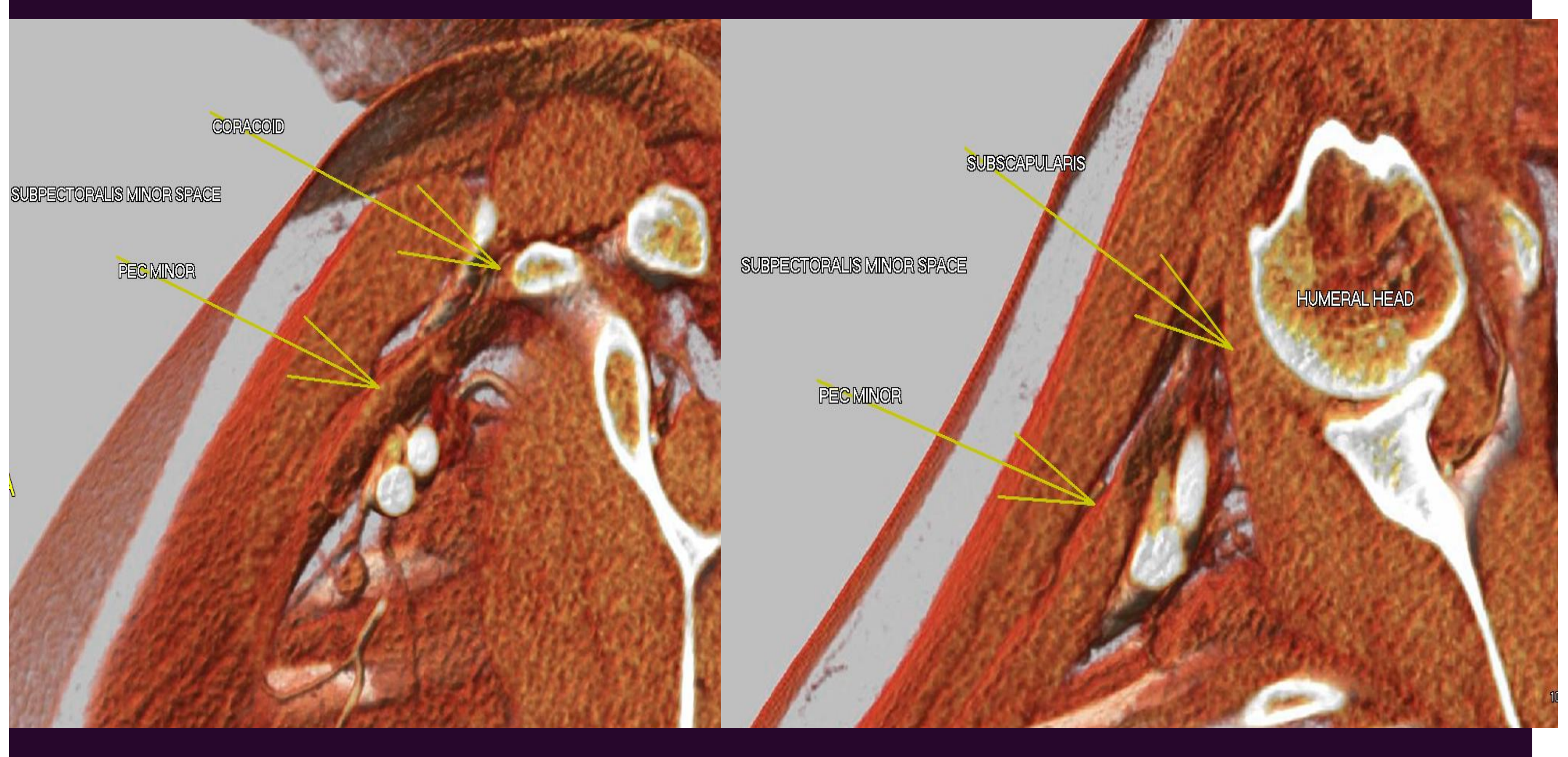
Costoclavicular Space

Borders:



- Inferior: 1st rib
- Superior: clavicle + subclavius ms.
- Anteromedial: Clavicopectoral fascia, rhomboid ligament & sternum
- Posterolateral: Middle scalene muscle
- Anterior scalene muscle divides space into anterior and posterior divisions

Retro-pectoralis Space



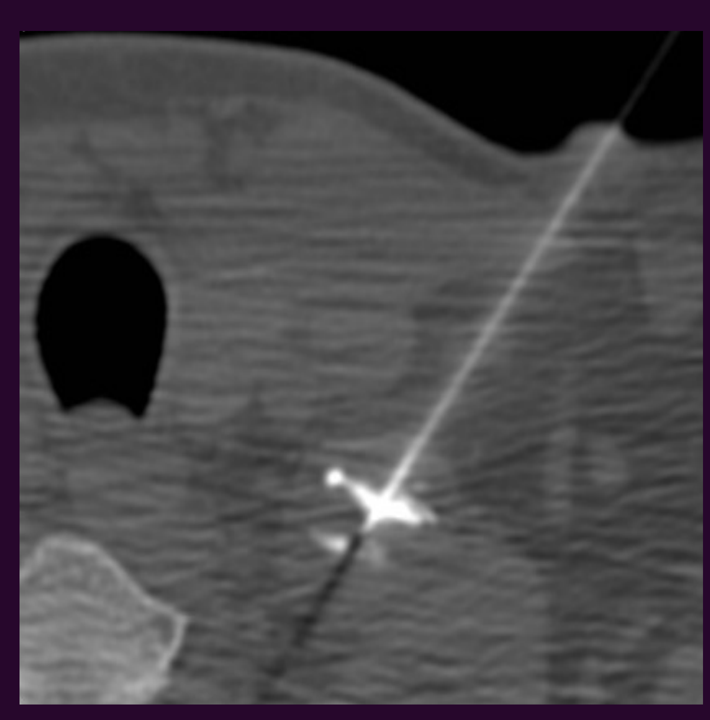
3 Types of TOS

- Neurogenic: 90%, F>M
  - May be post-traumatic, compression, post-XRT
  - Parasthesias, pain → worse with ABduction
  - motor weakness in 10%
  - Usually responds to PT, NSAIDs, massage, postural change, etc
  - MRI → brachial plexus distortion, muscle atrophy, etc.
- Vascular: 10%, M>F
  - Arterial
  - Venous

Demondion X, et al. Radiographics 2006 vol. 26 (6) pp. 1735-50

Neurogenic TOS

- SX
  - Arm pain & paresthesias -> 95%,
    - Pain in neck, shoulder, arm, hand
    - Ulnar nerve paresthesias
  - Motor weakness -> 10%
  - May follow trauma / MVA
- Workup:
  - Nerve conduction velocities (NCV)
  - Electromyography
  - CXR (cervical rib or anomaly)
  - CT/MRI
  - Interscalene block



Arterial TOS

- Least common TOS
- Overhead athletes: Baseball, swimming, volleyball, weight-lifting
- Cause: Repetitive compression injury
  - Anatomic predisposition (tight CCS)
  - Post-traumatic, bony callus
  - Scalene hypertrophy
- Sx:
  - cool, weak, easy fatigability arm & hand, diffuse arm pain (ischemic neuritis of brachial plexus)
  - Thrombosis / Distal emboli / pseudoaneurysm
  - Raynaud's phenomenon

Venous TOS: “Effort Thrombosis”

- Paget-Schroetter syndrome (PSS), “axillo-subclavian venous thrombosis”
  - Overhead athletes
- SX:
  - Edema, venous congestion, arm heaviness, cyanosis of hand and arm
  - PE in up to 1/3!! *
  - Post-thrombotic syndrome (later)

* Perlowski AA. Vasc Med (2010) vol. 15 (6) pp. 469-79

Vascular TOS: Workup

- Clinical Tests (Adson, Roos, etc)
- Plain films
- Ultrasound
- CTA / MRA
- Catheter angiography

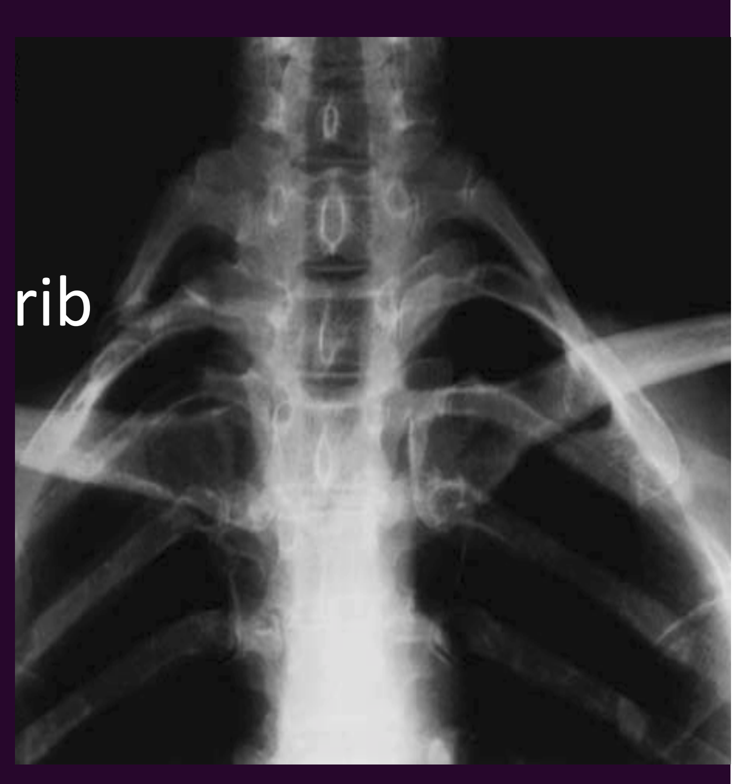
DYNAMIC EVALUATION IS IMPORTANT !!

Clinical Tests

- Adson: supination + 15^0 abduction, head turned to Sx side, lose radial pulse w/ deep inspiration
  - Most sensitive and specific
- Roos
- Costoclavicular compression
- Wright

Effort Thrombosis: Imaging

- Radiography: Valuable 1st step
  - Cervical rib
  - Elongated C7 trans. process
  - Callus/Deformity of clavicle / 1st rib
  - Exostosis
- Ultrasound



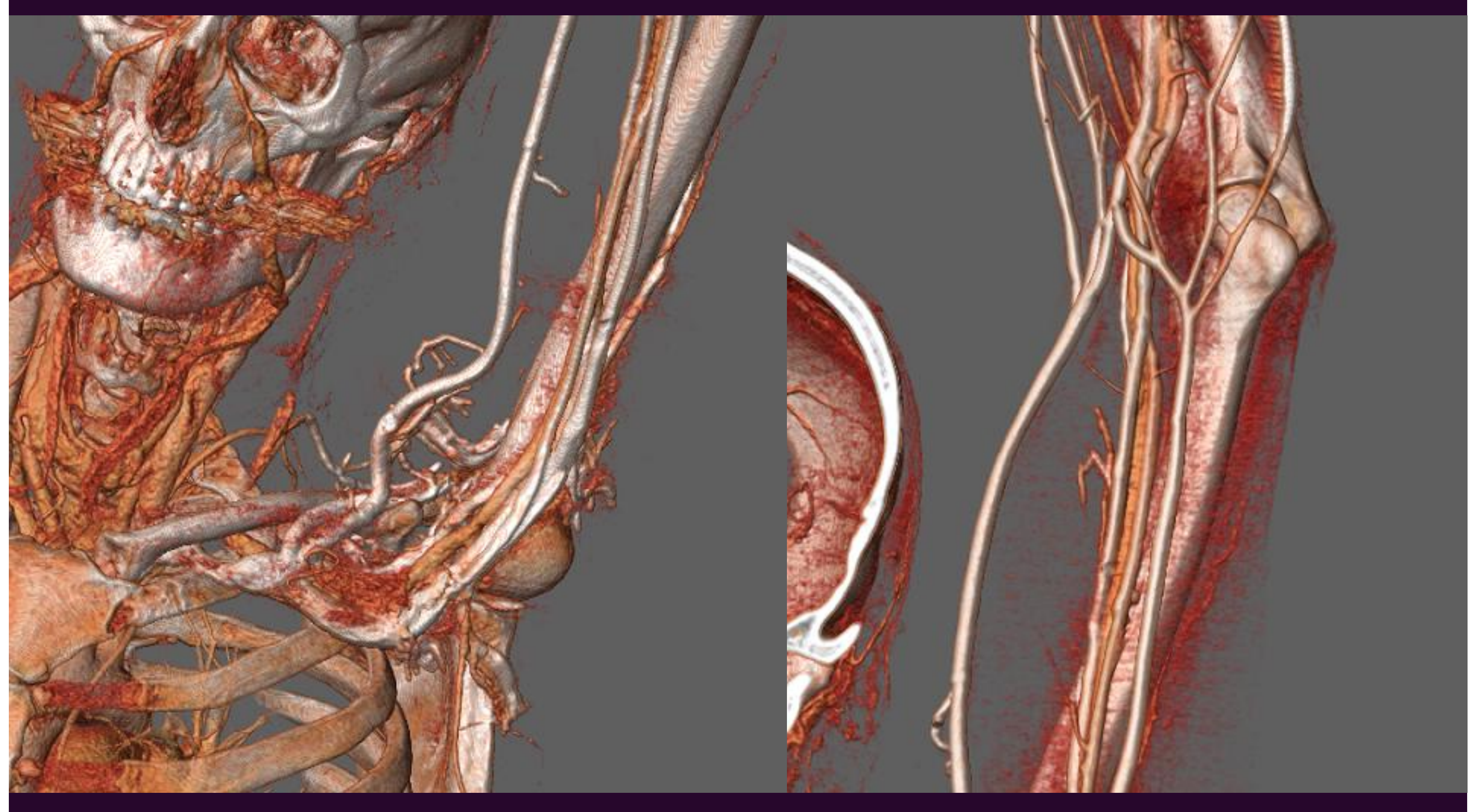
CTA Imaging in TOS - Considerations

- Dynamic evaluation
- Image with arm at side and over head
  - Positioning!!
- Which arm to inject?
- One injection or two ?
- Timing?

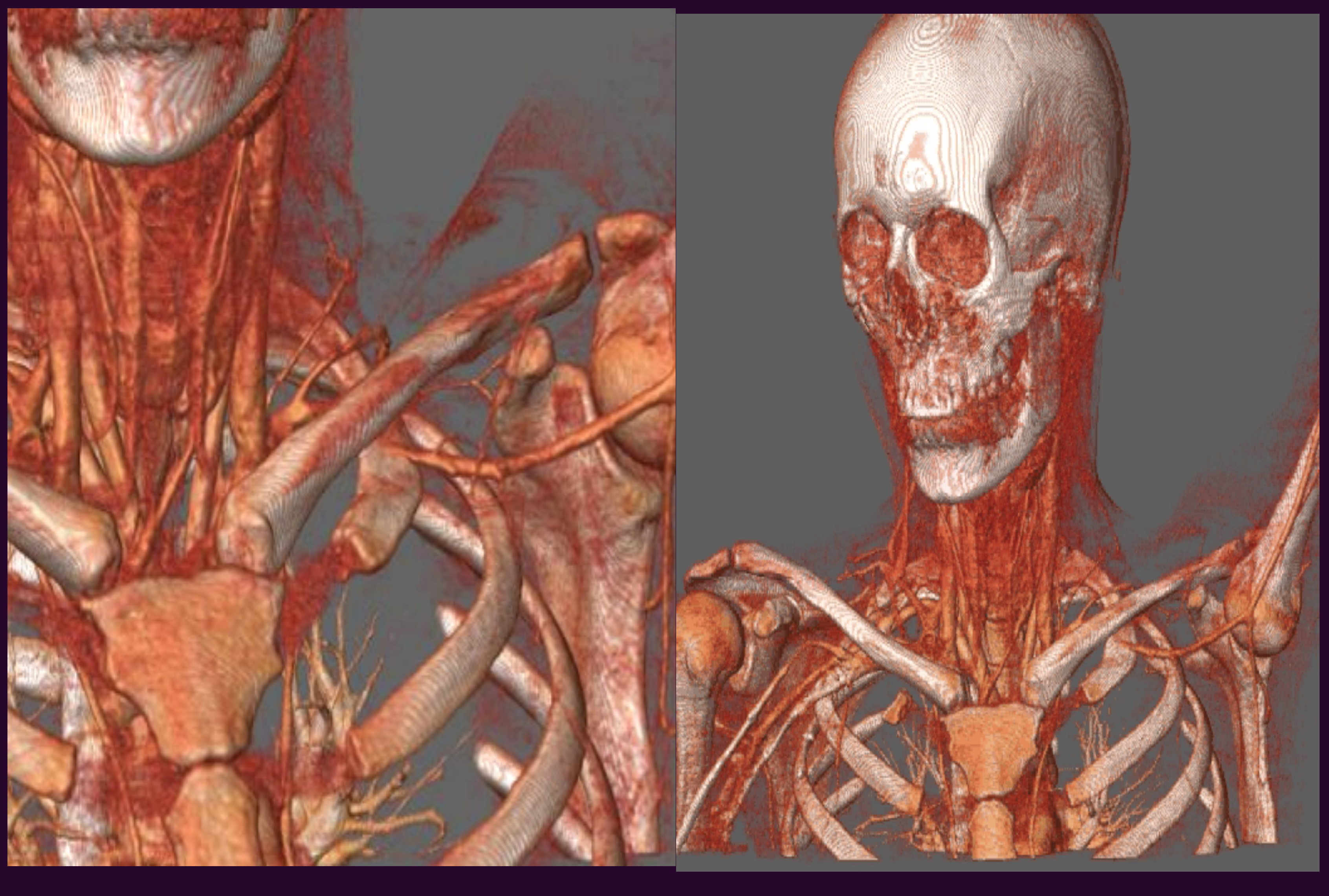
CTA for TOS: Combo Direct / Indirect CTA

- Ipsilateral IV, arm over head w/ palm taped up
- Bolus: 120 mL full-strength @ 4ml/s
- Chase: 100 mL dilute (10%) contrast @2.5 ml/s
- 65 sec empiric delay, scan caudo-cranial
- Arm down, immediate re-scan cranio-caudal
- **Volumetric Review**

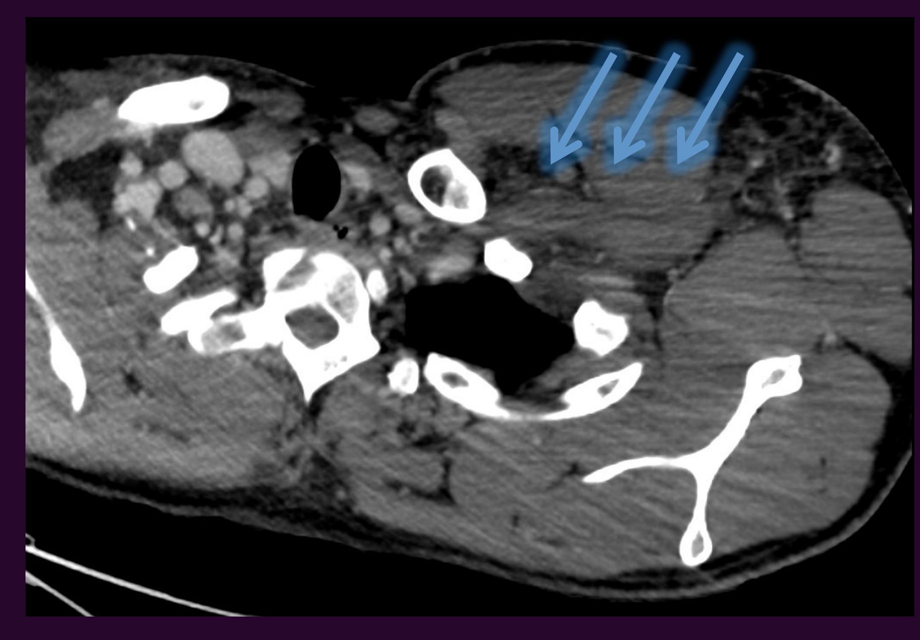
Combo Direct / Indirect CTA

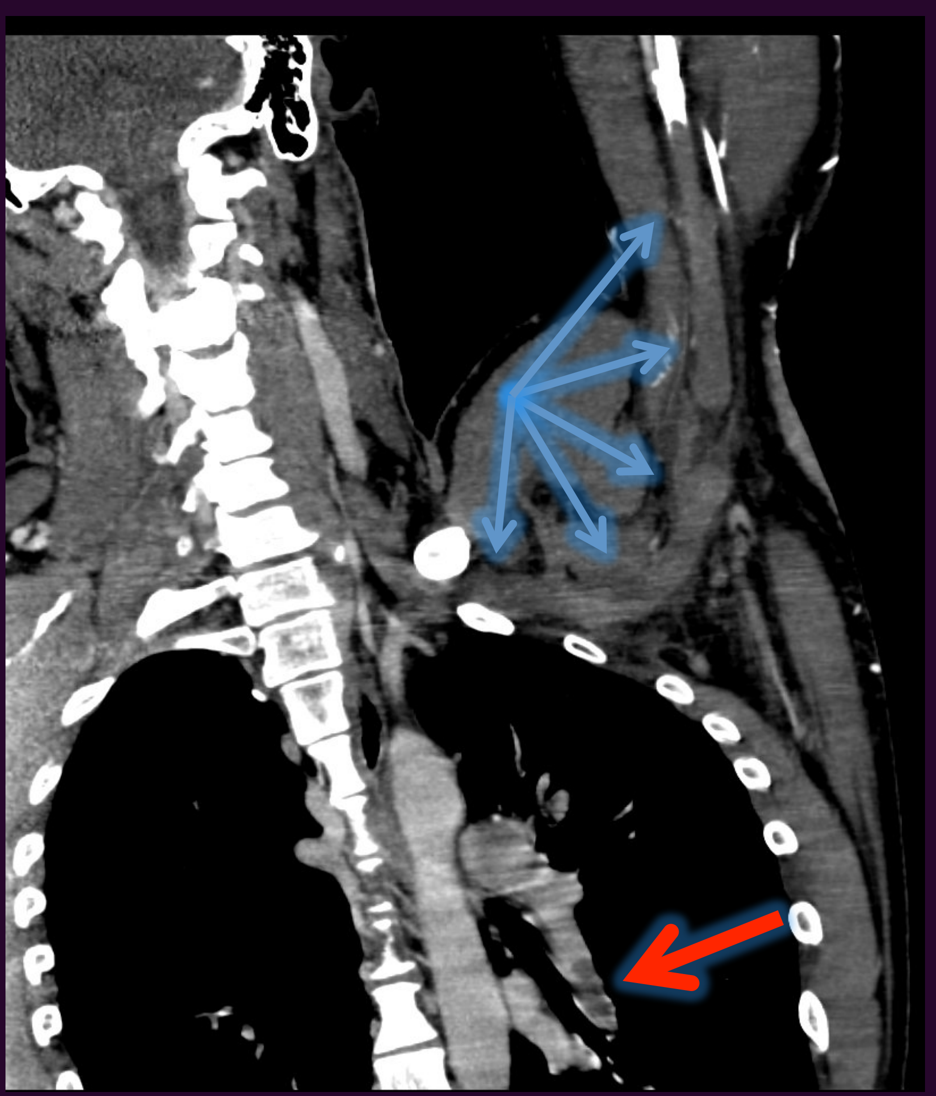


Arterial Compression



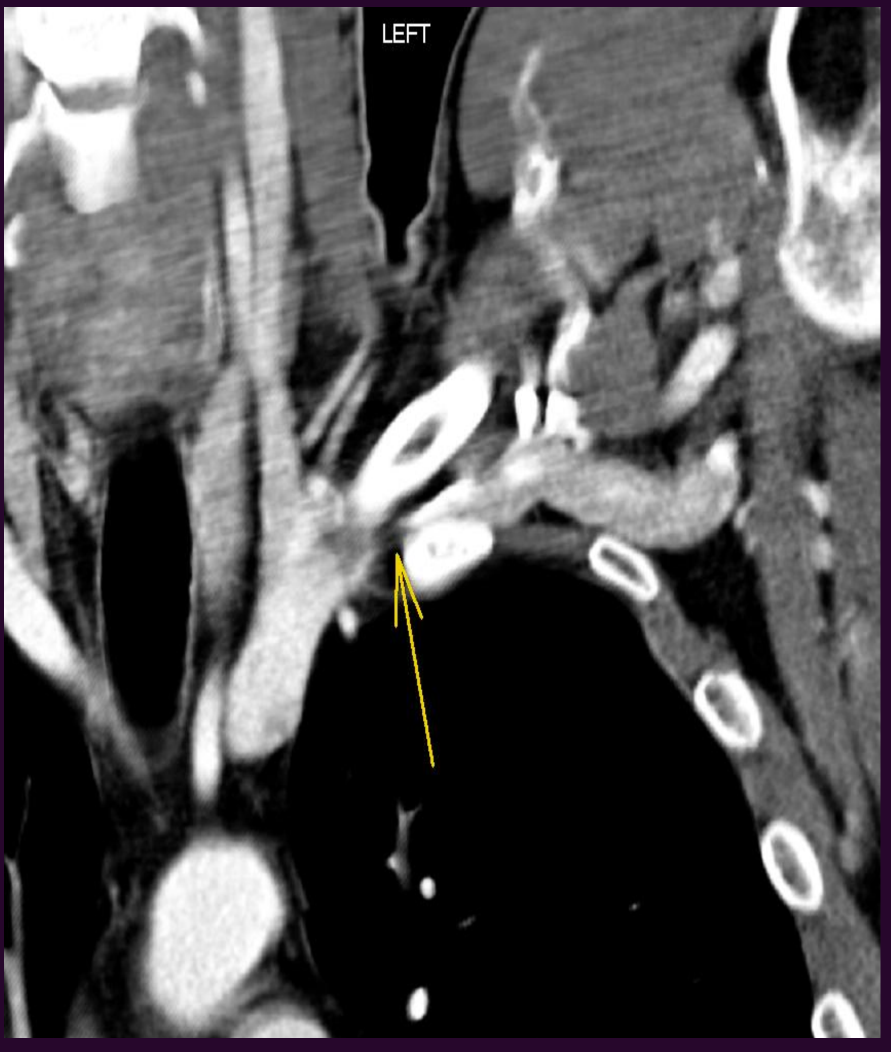
Effort Thrombosis – 26 year old pitcher



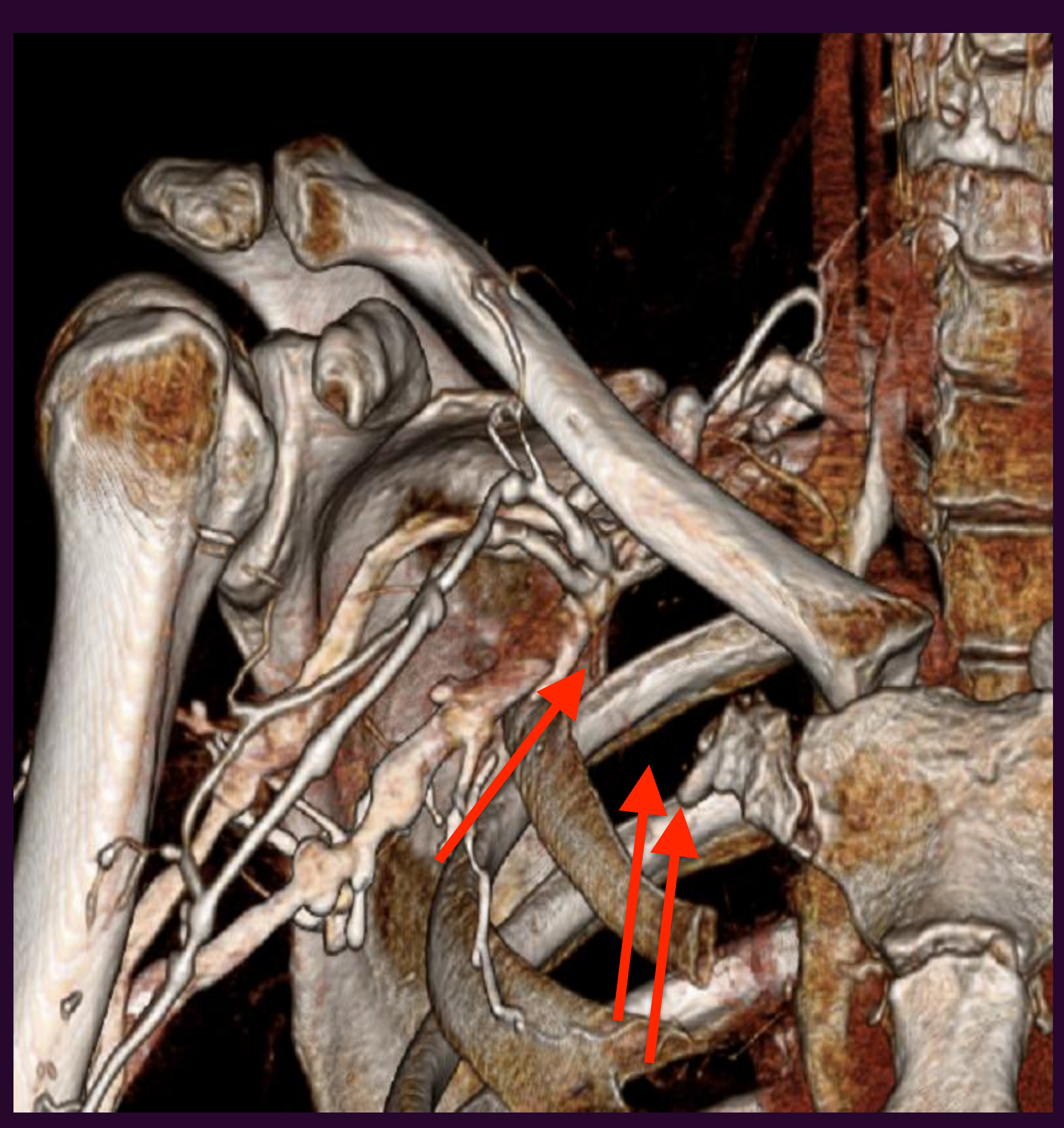


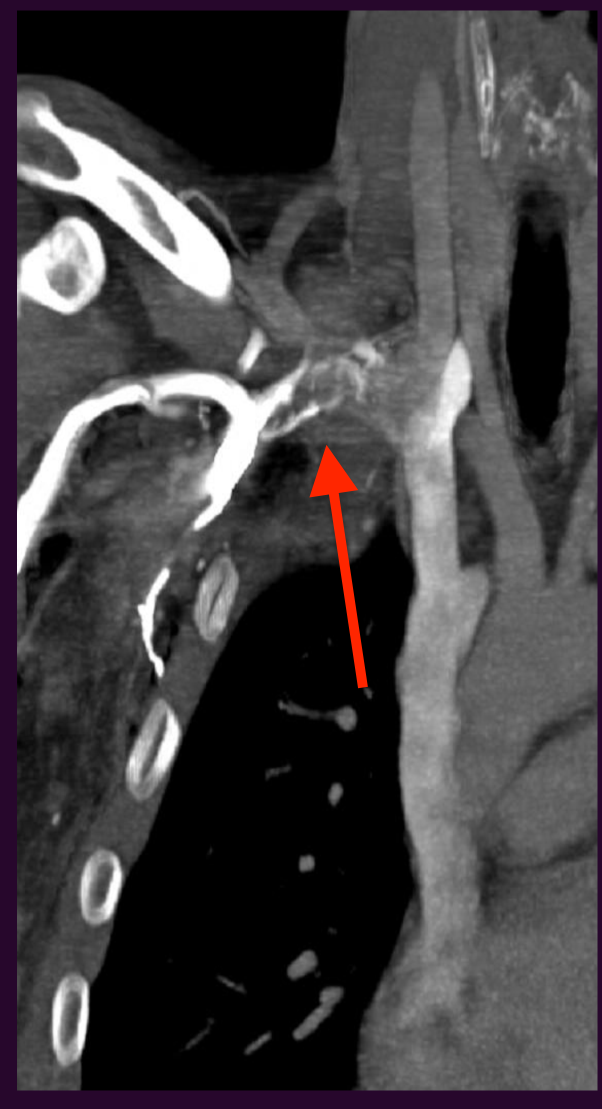
Courtesy Lt.Cmdr Scott Alexander, MD

Effort Thrombosis:
36 YO weightlifter

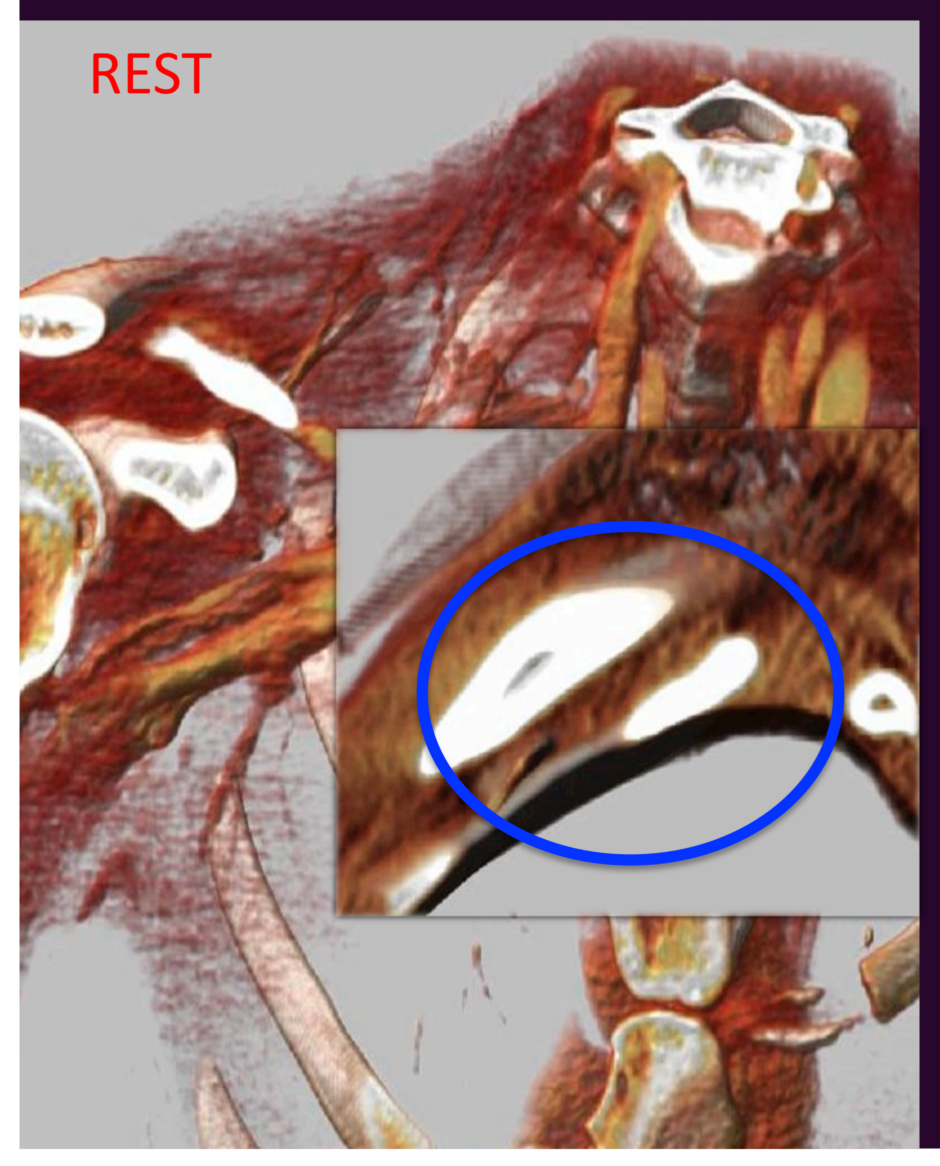


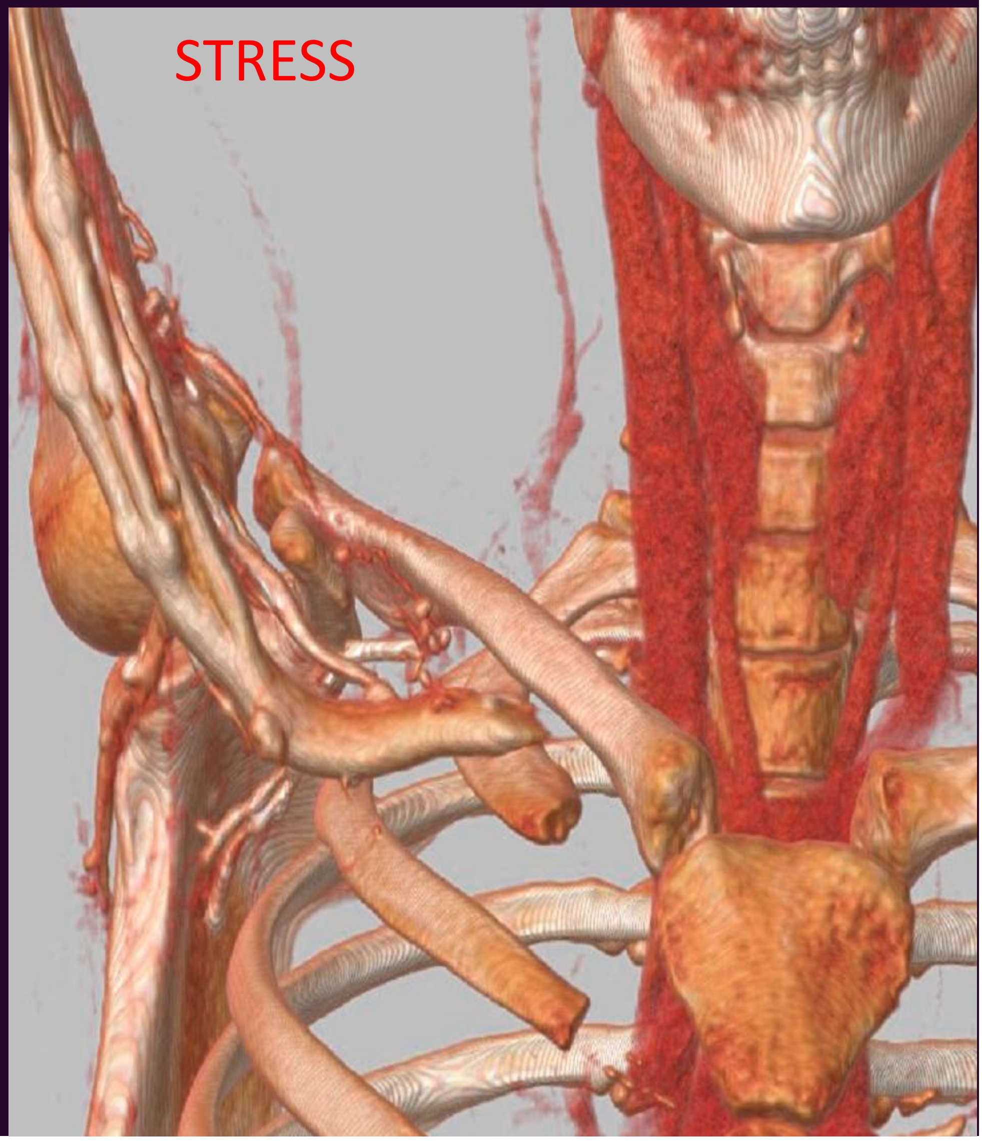
Post-Op 1st rib resection





Arterial and Venous TOS: 16 YO Volleyball Athlete





Flow limitations in the Athletic Pelvis

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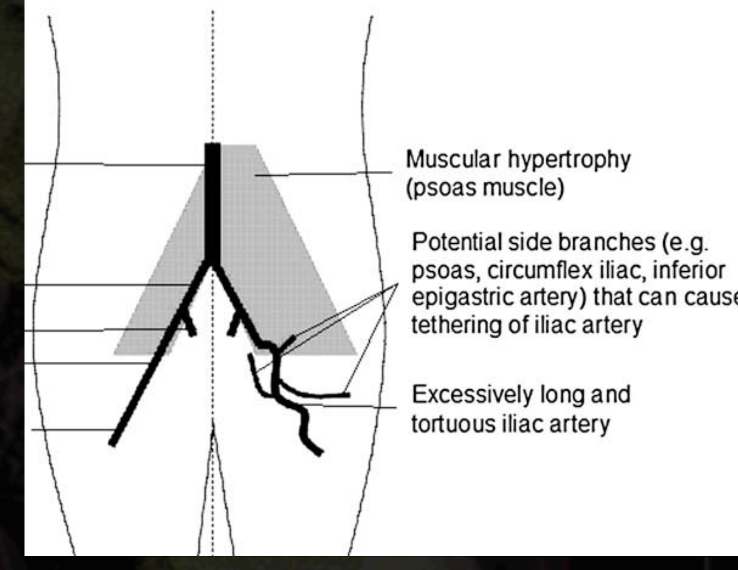
Flow limitations in the pelvis in athletes



- Static: Iliac endofibrosis
- Dynamic:
  - Elongated / tortuous vessels
  - Kinking with or w/o stenosis (elongation/ tethering)
  - compression (psoas, ligaments)

Lim CS, et al., Eur J Vasc Endovasc Surg 2009:38:180-6.

Iliac Endofibrosis

- Rare, M>F, average age : 25 years
- 90% of pts are cyclists (15 % professional, 48% top amateur) requires 8000-35000 km/yr or 150,000 km lifetime
- Also: speed skaters, runners, wt lifters, XC skiers, and rugby players
- 90% external iliac artery (EIE)
- Pathology: intimal fibroplasia, medial hypertrophy, and adventitial hyperplasia. Involved segments universally free from atherosclerosis.

Lim CS, et al. Eur J Vasc Endovasc Surg 2009:38:180-6.

Clinical evaluation of EIE

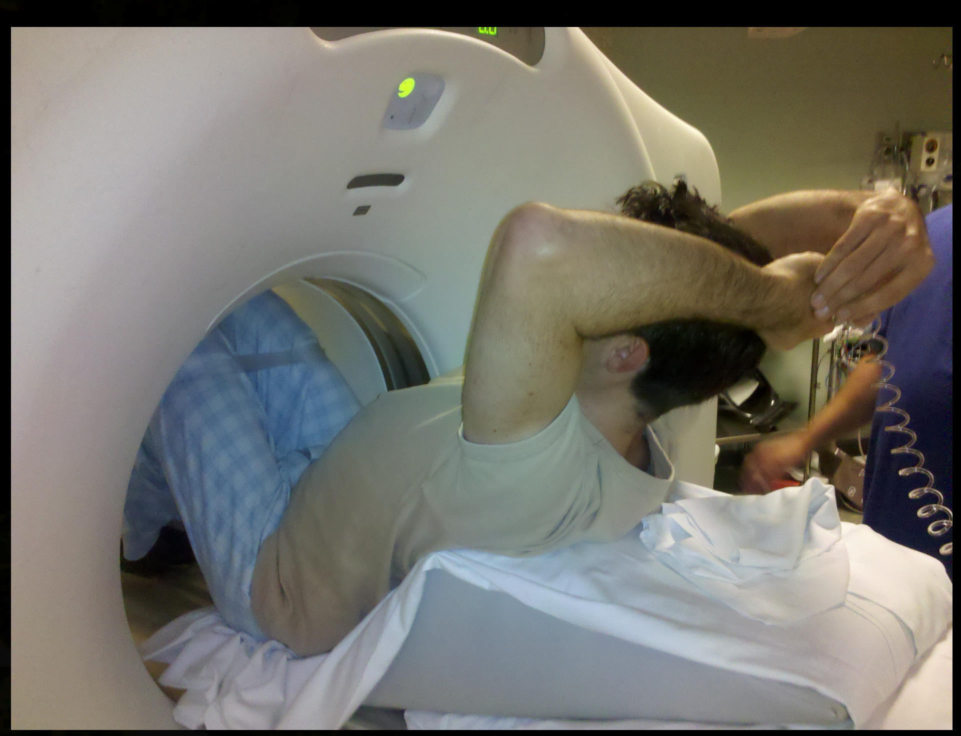
- SX: performance decline, weakness, lack of power
  - bilateral in up to 15%
- Exertional ABI: flat or drop (false negatives)

Vascular Lesion of EIE

- Smooth, eccentric, non-calcified
- Imaging Options:
  - CTA
  - MRA
  - US
  - DSA (with IVUS)
- Provocative maneuvers useful

CTA: Imaging technique

- Two phases in relaxation and hip flexion
  - Try simulate cycling position as close as possible considering space within CT gantry (almost 90 deg)



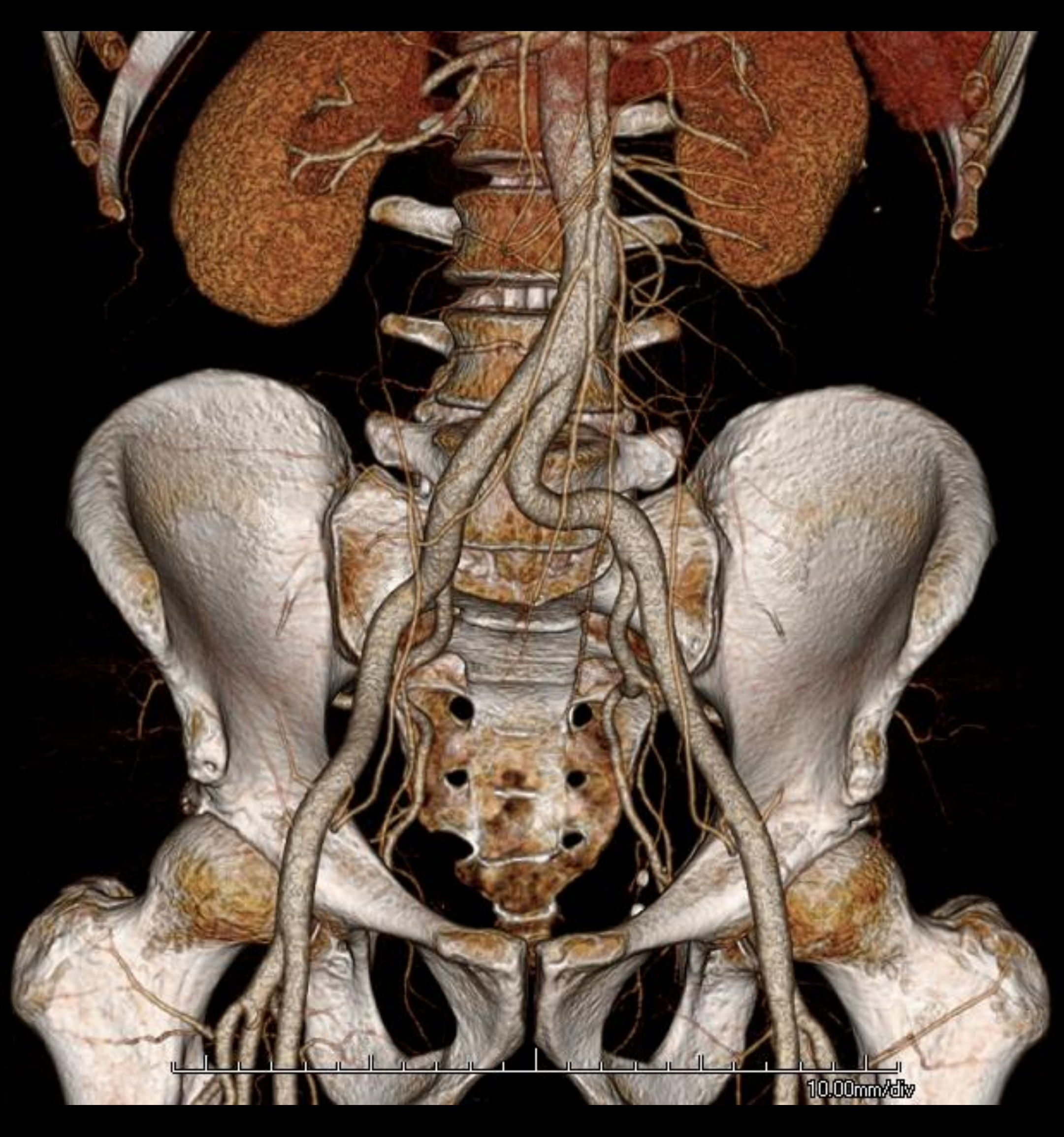
CTA: Imaging technique

- Two phases in relaxation and hip flexion
- Coverage ~ 40 cm
- Recon: overlapped 0.625 - 1.25 mm (30 % ASIR)
- Relaxation – 100 kVp, flexion – 120 kVp
- 80 - 100 cc of IV contrast for each phase, 20 sec injection.
- Saline flush at same rate
- Approx 10 sec scan

Case I

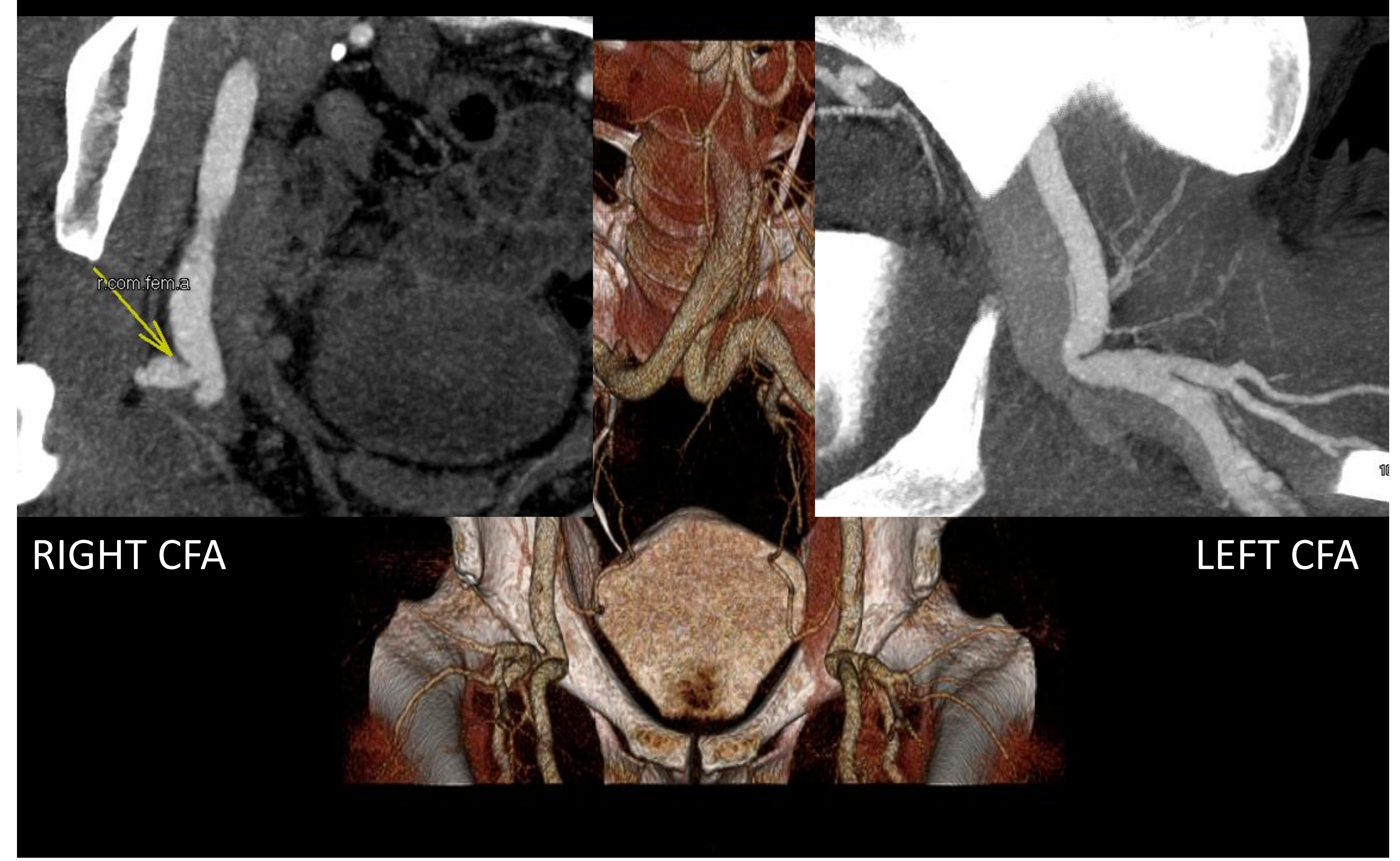
- 45 yo avid cyclist
- Proximal thigh pain, cramping with exertion
- ABI drops with exertion

Supine, legs extended



Dynamic Flow Restriction

Hip flexion

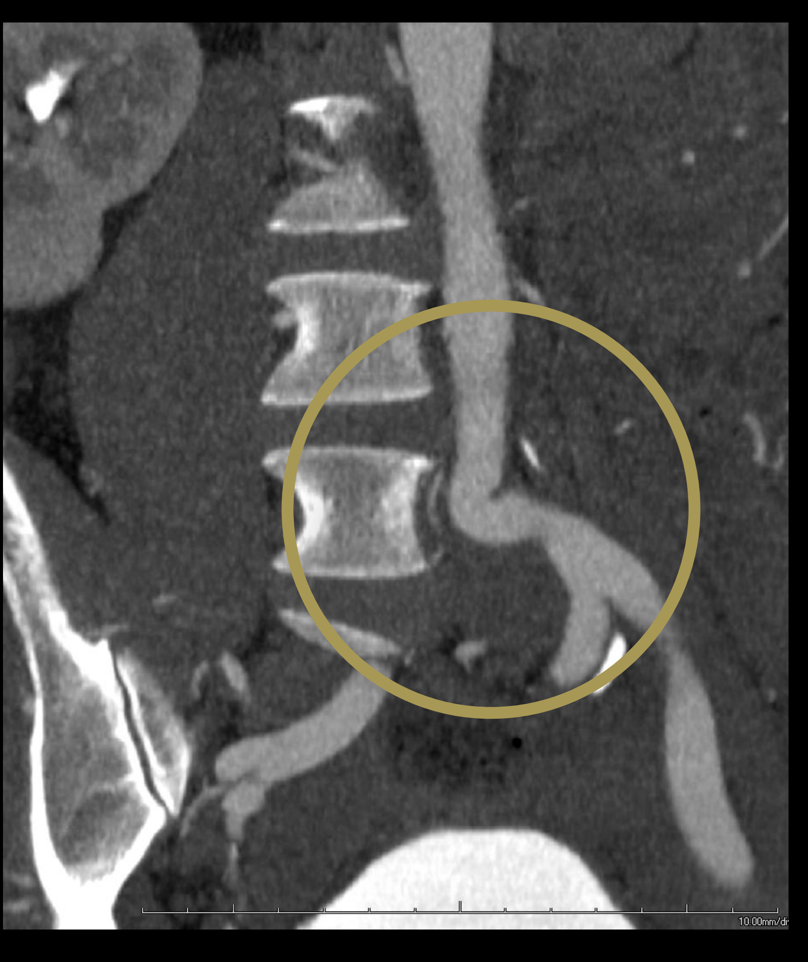


Case 2

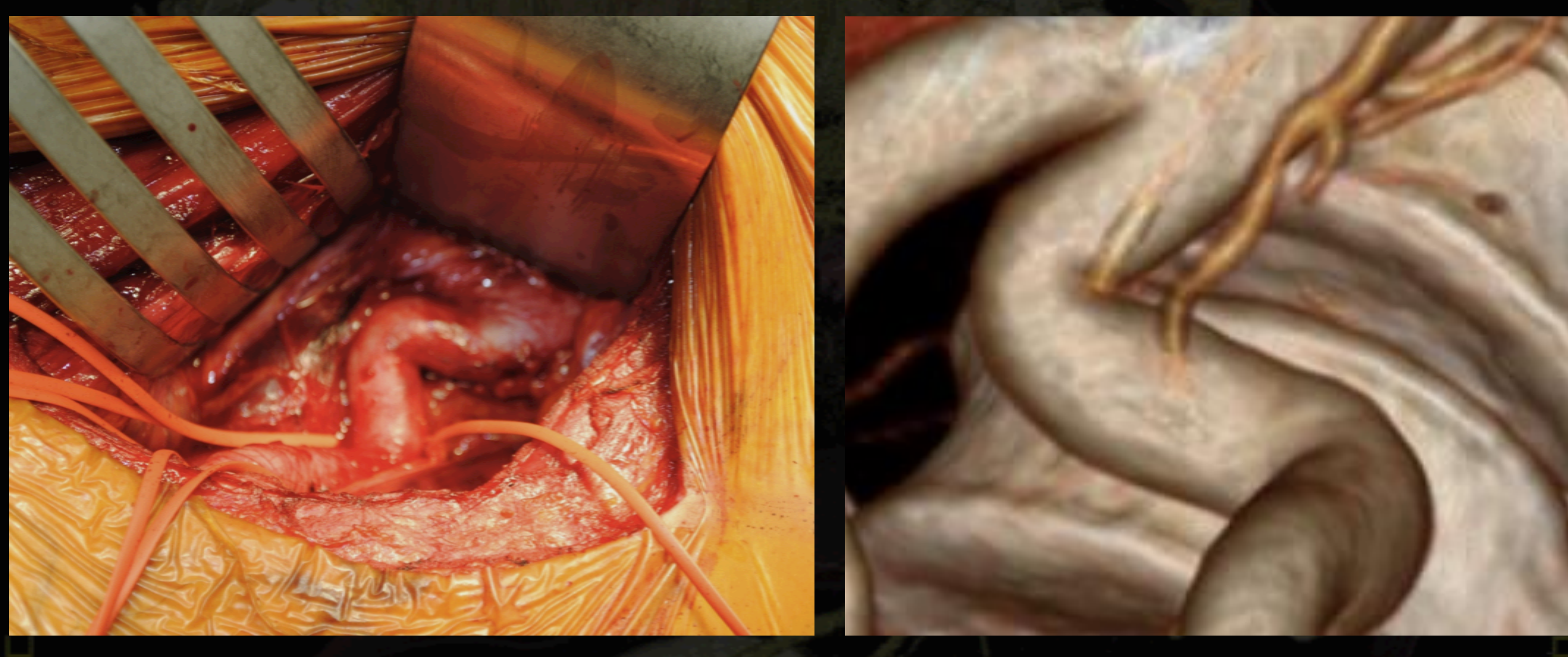
- 49 yo avid cyclist since 1980's
- left thigh and buttock pain at high performance levels.
- Pain described as a "deep burn" and would occur more frequently on flat surfaces as opposed to pedaling on an incline.

- ABI R/L: 1.3/1.2
- Exercise ABI R/L: 1.5/1.2

CTA at Rest



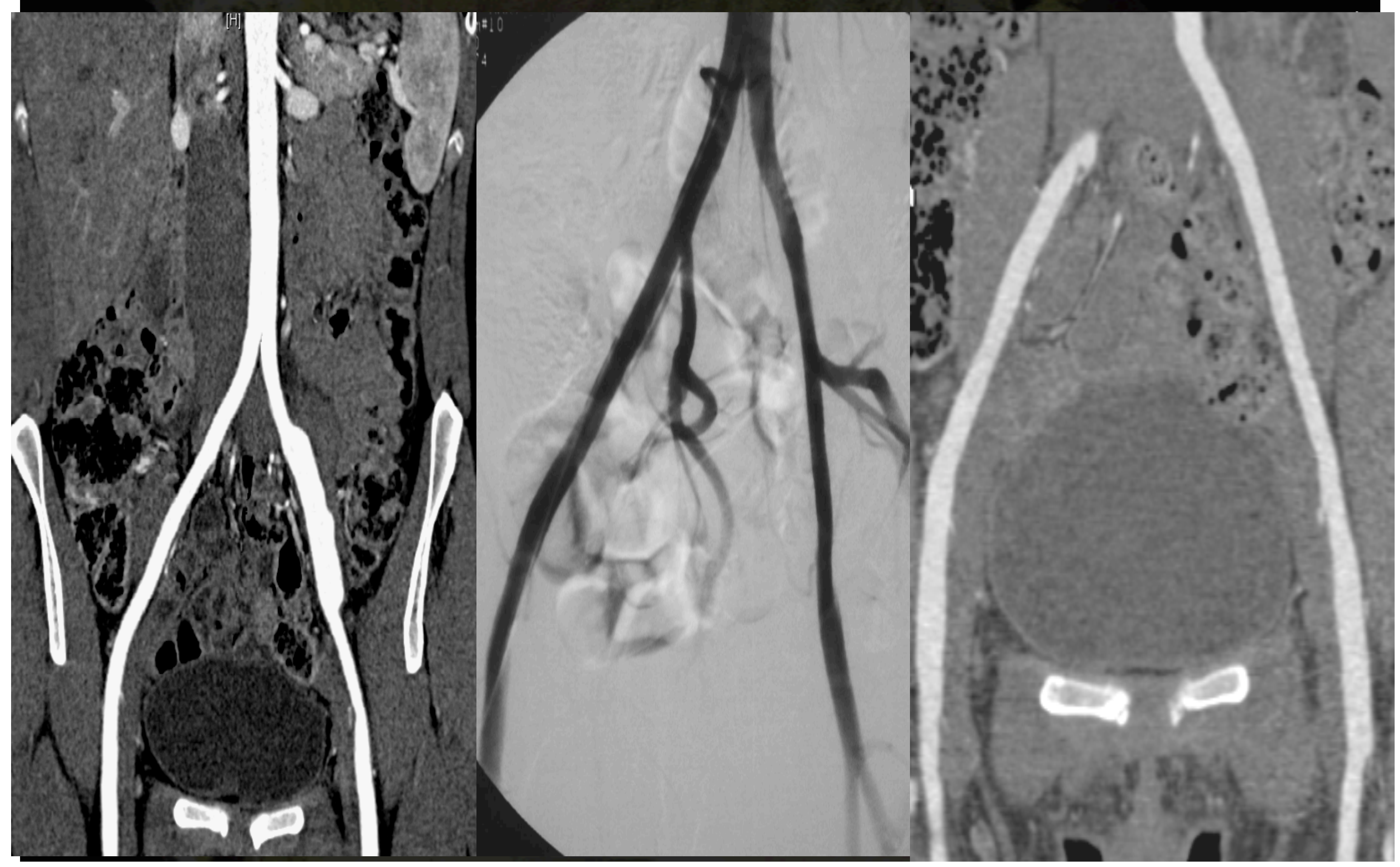
Operative Findings

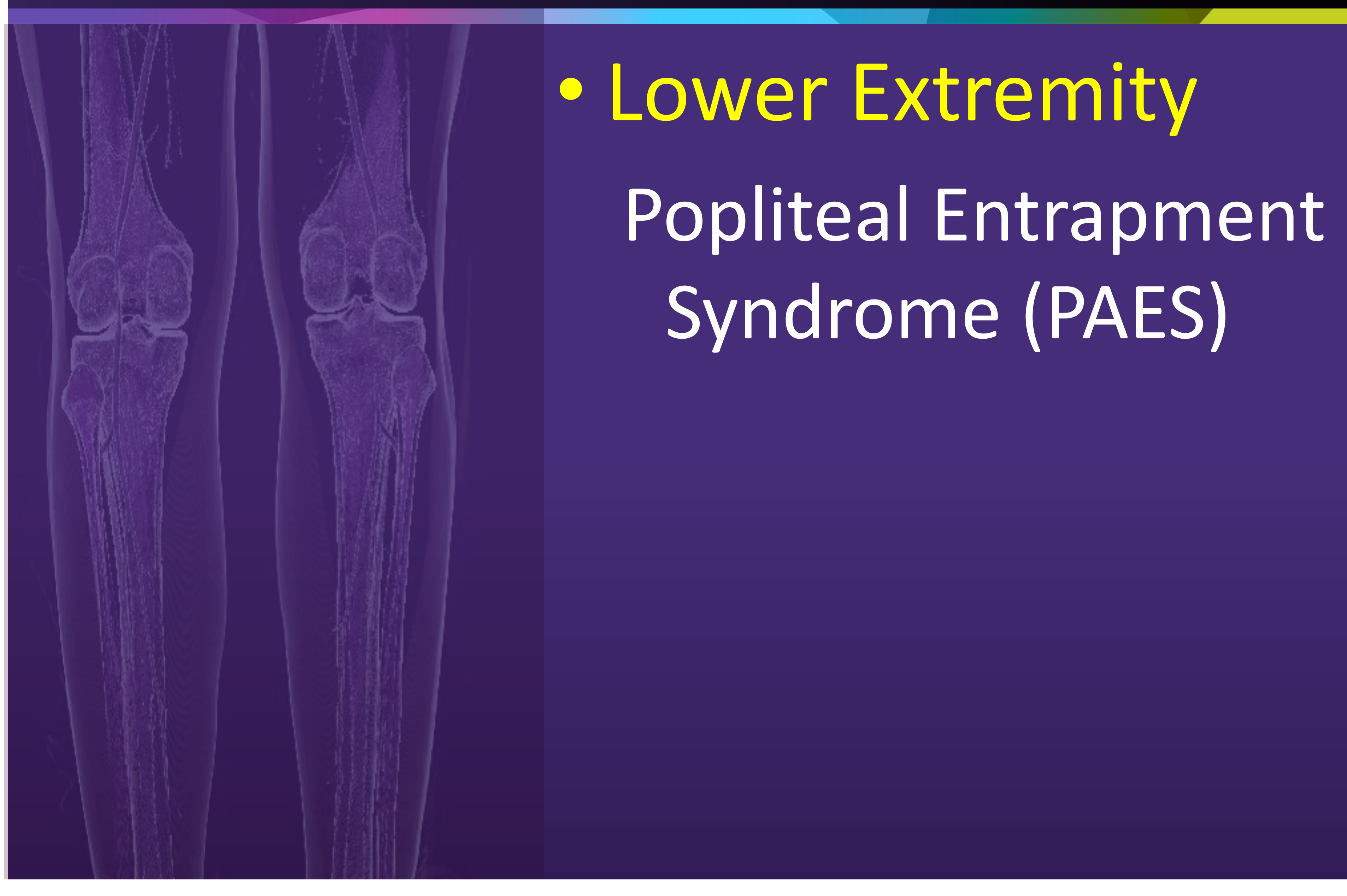


- Pathology: intimal thickening and fibrosis
- No inflammatory change

- 30 yo elite female cyclist
- left thigh and buttock pain at high performance levels.
- Exercise ABI drop on left from 1.1 → 0.5

30 FEMALE ELITE CYCLIST





Vascular Diseases in Athletes
• Lower Extremity
Popliteal Entrapment
Syndrome (PAES)

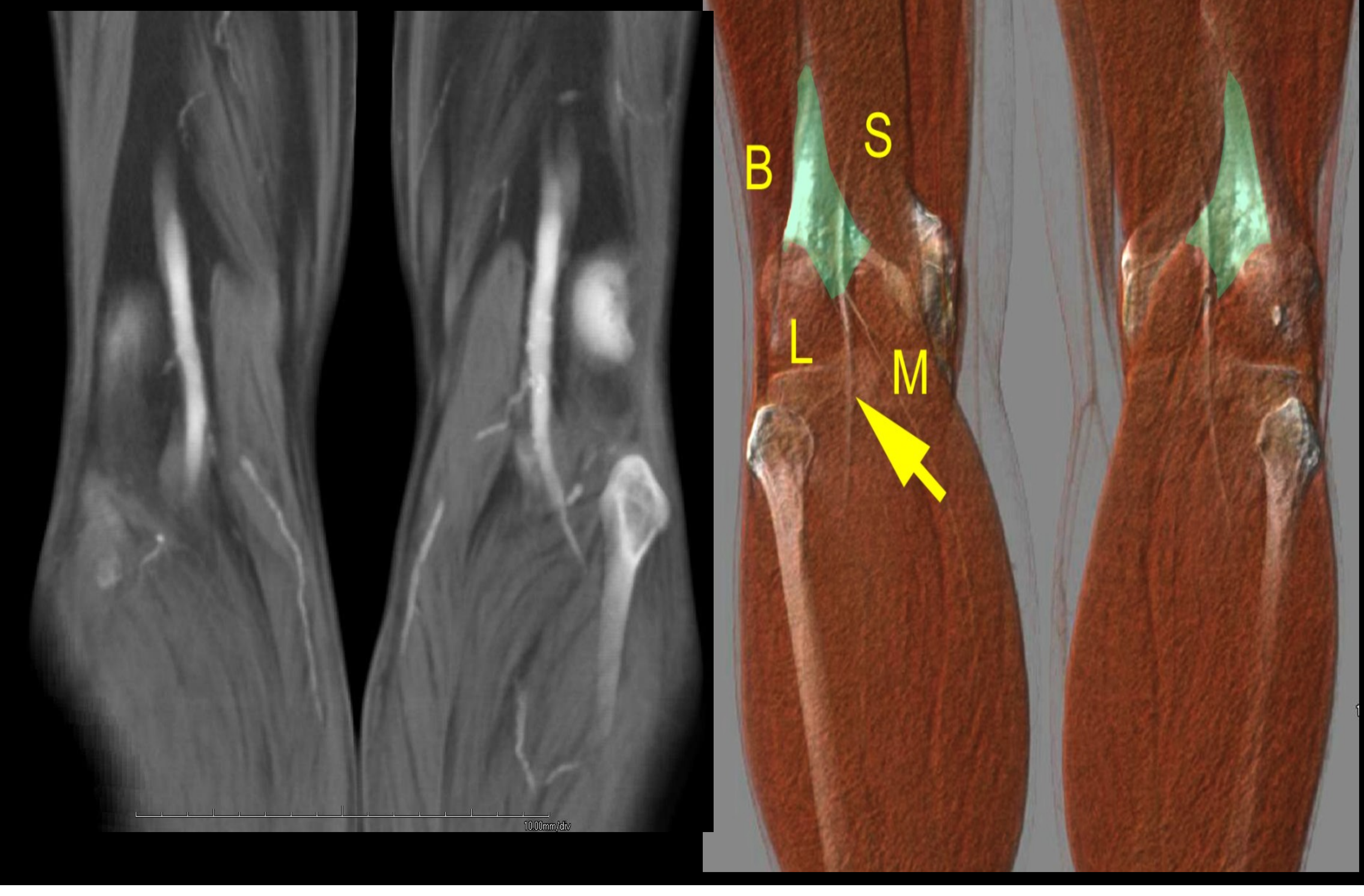
PAES – Background

- Anatomic incidence up to 3.5%
- Clinical Sx in ~ 0.17% young, active adults
  - Claudication in 90%
  - Walking rather than running
- Acute / Chronic Limb Ischemia in 10%
  - Parasthesias, rest pain, tissue loss

PAES - Embryology

- In utero, medial head of gastrocnemius (MHG) migrates from lateral to medial across popliteal fossa
- The 3 segments of popliteal artery are fusing at same time
- If delayed or abnormal migration → **MHG too far lateral**
  - Pop fossa space is limited, compression can result

Popliteal Fossa Anatomy



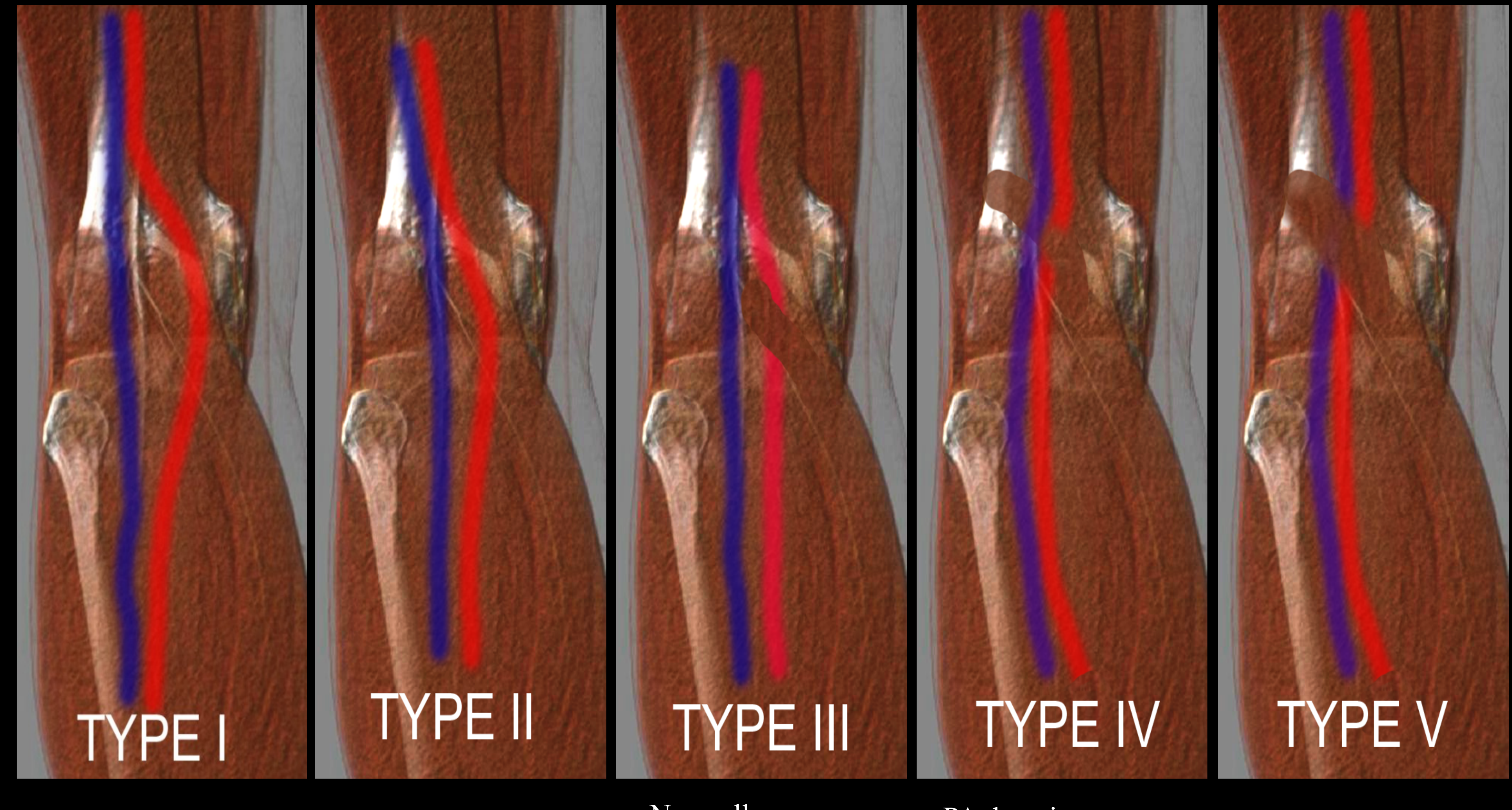
Etiologies of PAES

- Anatomic Compression
  - Anomalous course of PA
  - Anomalous origin of MHG
  - Accessory muscle slip
  - Fibrous band
- "Functional" compression: Typical Sx in absence of anomalous musculotendinous structures

Functional Popliteal Entrapment (Type VI)

- Tends to occur in younger population
  - almost exclusively in highly conditioned athletes
- Compression by hypertrophic gastrocnemius +/- soleal sling
- Longer segment involvement (vs. anatomic PAES)
- Treatment:
  - Try conservative management first
  - Surgical release, debulking of muscle if needed

* Whelan TJ. In: Haimovici H, Ed. Vascular surgery: principles and techniques. New York: McGraw-Hill, 1984: 557-67



TYPE I	TYPE II	TYPE III	TYPE IV	TYPE V
PA travels aberrantly, medial to normally positioned MHG	Anomalous lateral and inferior origin of MHG, PA displaced medially	Normally positioned PA compressed by muscular slip or aberrant band from MHG	PA deep in popliteal fossa, entrapment from aberrant band or popliteus muscle	Any type of entrapment **involving popliteal vein**

Alternate Classification of PAES

- Heidelberg Classification*:
  - I. Abnormal course PA
  - II. Abnormal muscle anatomy
  - III. Both

* Hoelting T. Br J Surg 1996; 85: 338-341.

Testing for PAES

- Positional Stress Test (PST)
- Doppler US
- Cath Angio
- MRI
  - time-resolved imaging[1]
  - blood pool contrast agent[2]
- CTA

FUNCTIONAL EVALUATION NECESSARY!!

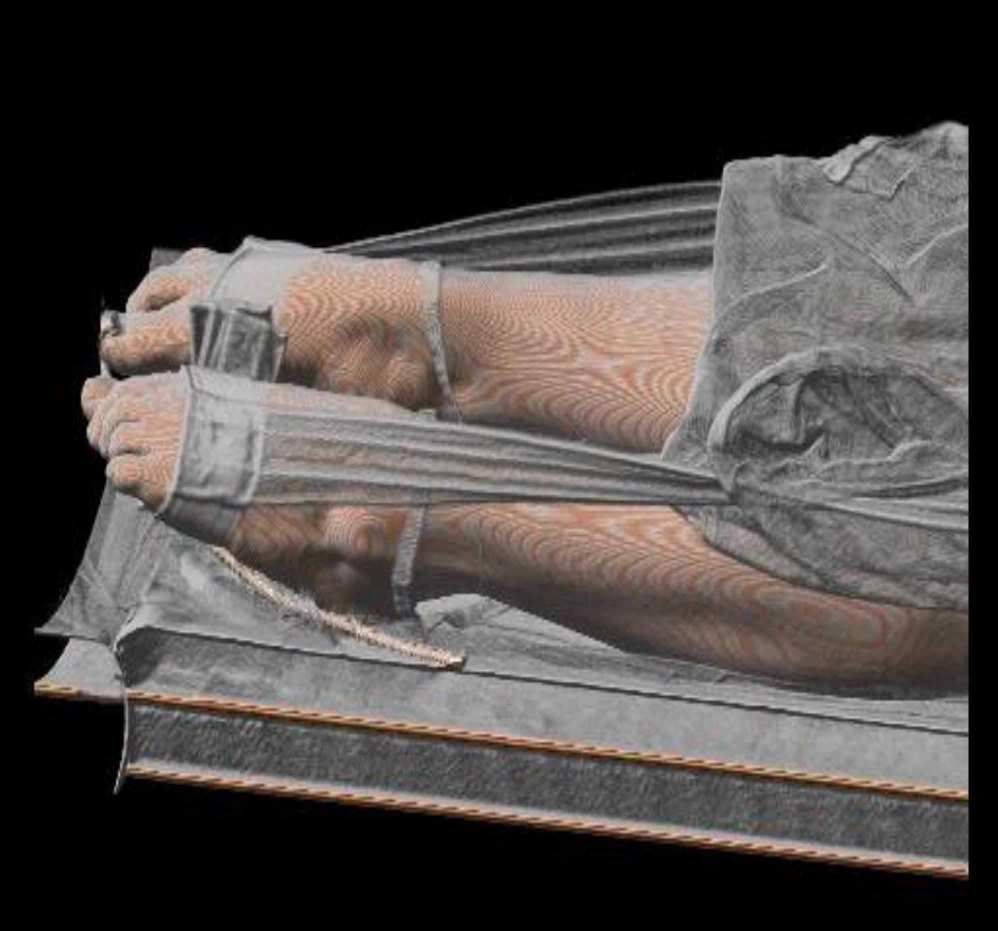
[1] Blackham, KA et al. Amer J Roentgen 2011. 196 (5) W613-20
[2] Beitzke, D et al. Cardiovasc Intervent Radiol 2011. 34 (Suppl 2) S12-6

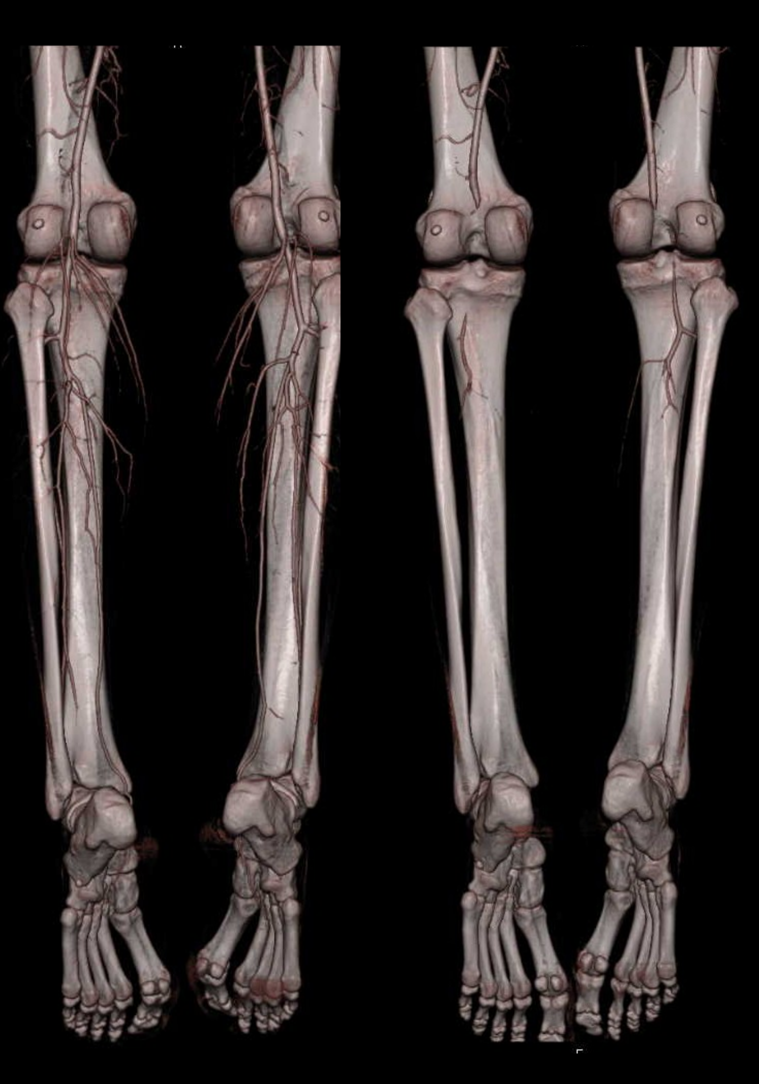
PAES: CTA Imaging Technique

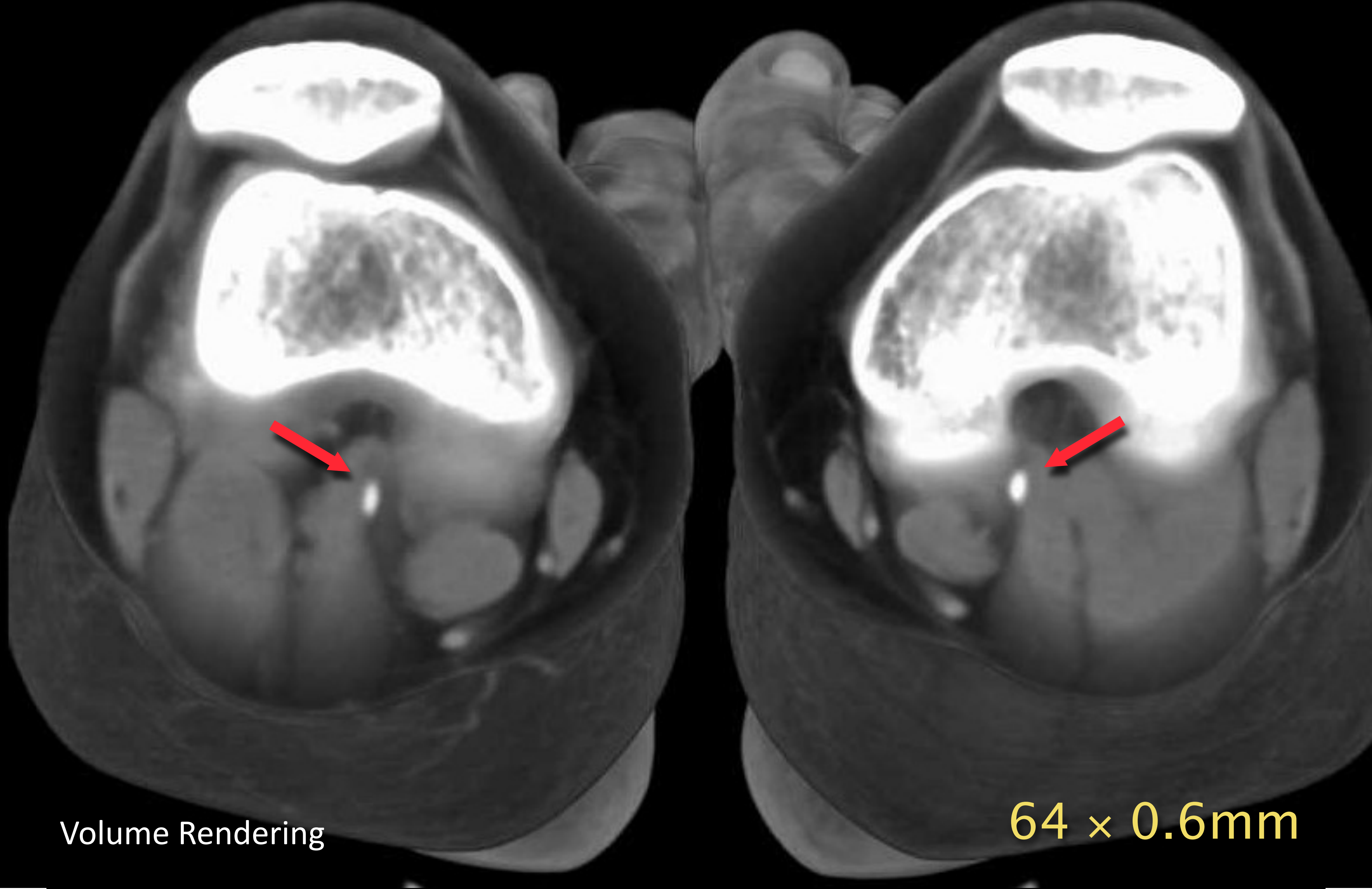
- 2 phases – relaxed and active plantar flexion
- ~ 80 mL of contrast (4mL/s) for each phase regardless of weight, followed by saline flush at same rate
- Bolus track distal SFA
- Scan time: 12-15 sec
- Pulse oximeter on symptomatic large toe
- Actively plantar flex without bearing down
  (straps)

PAES: CTA Imaging Technique

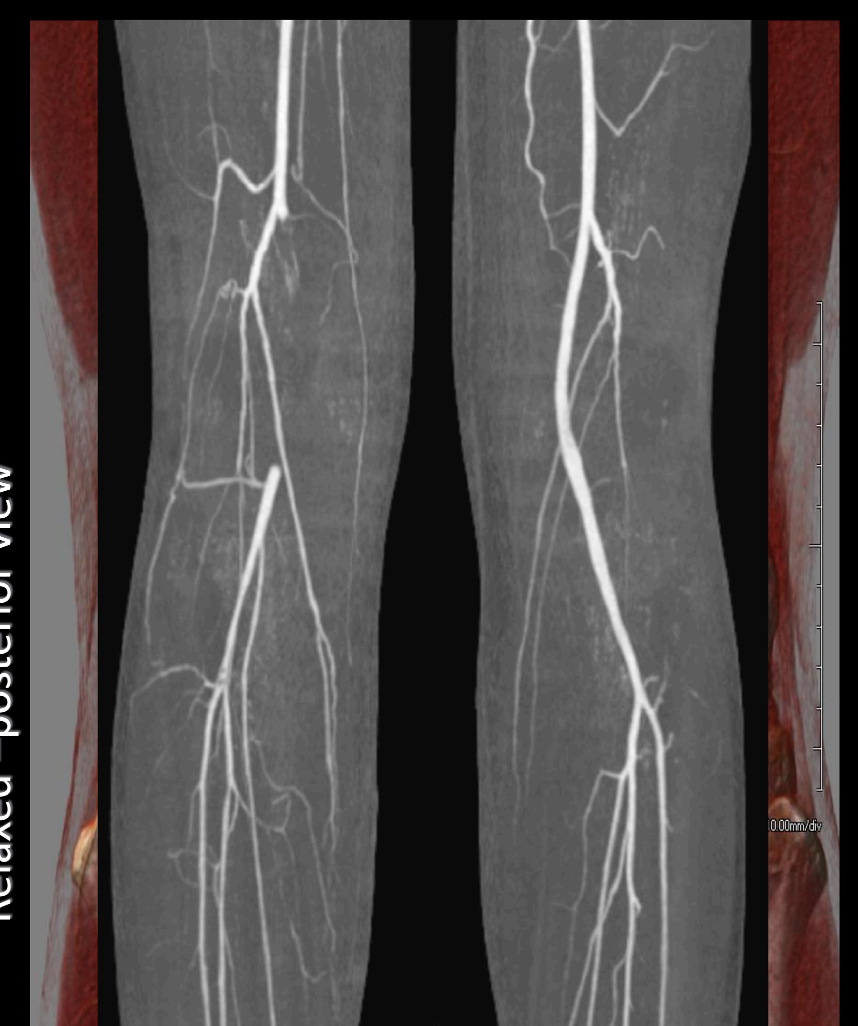
PAES: CTA Imaging Technique

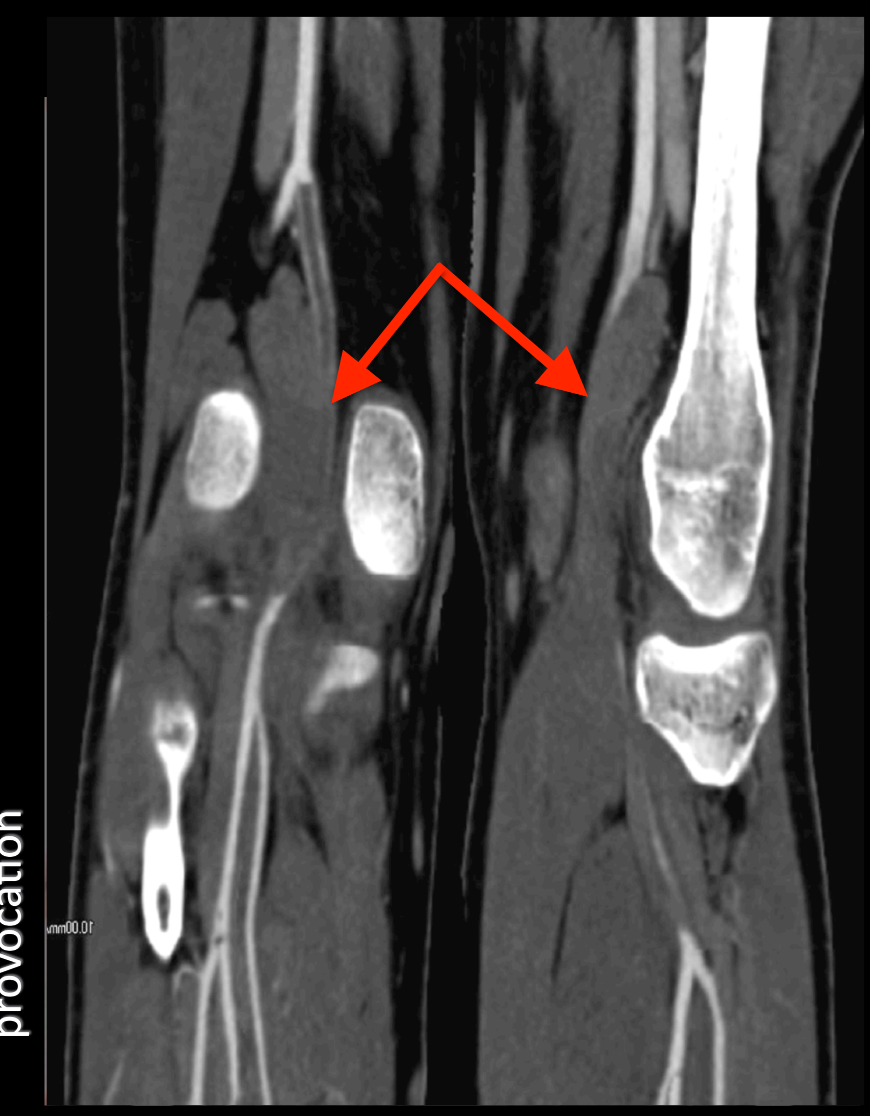





Soccer Player - Type I PAES
Volume Rendering
64 × 0.6mm

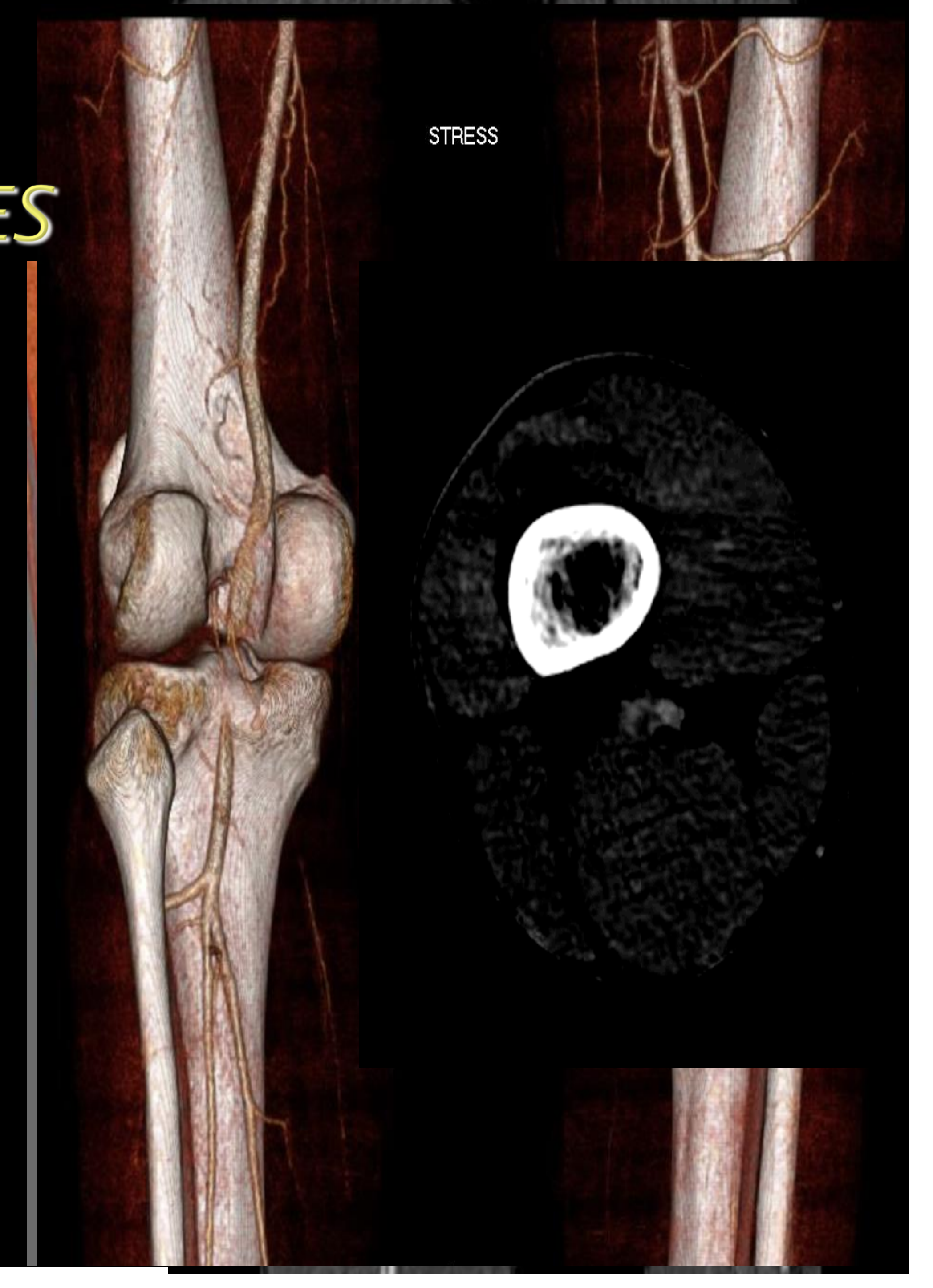
Type III PAES
Thrombosis of Left popliteal a.



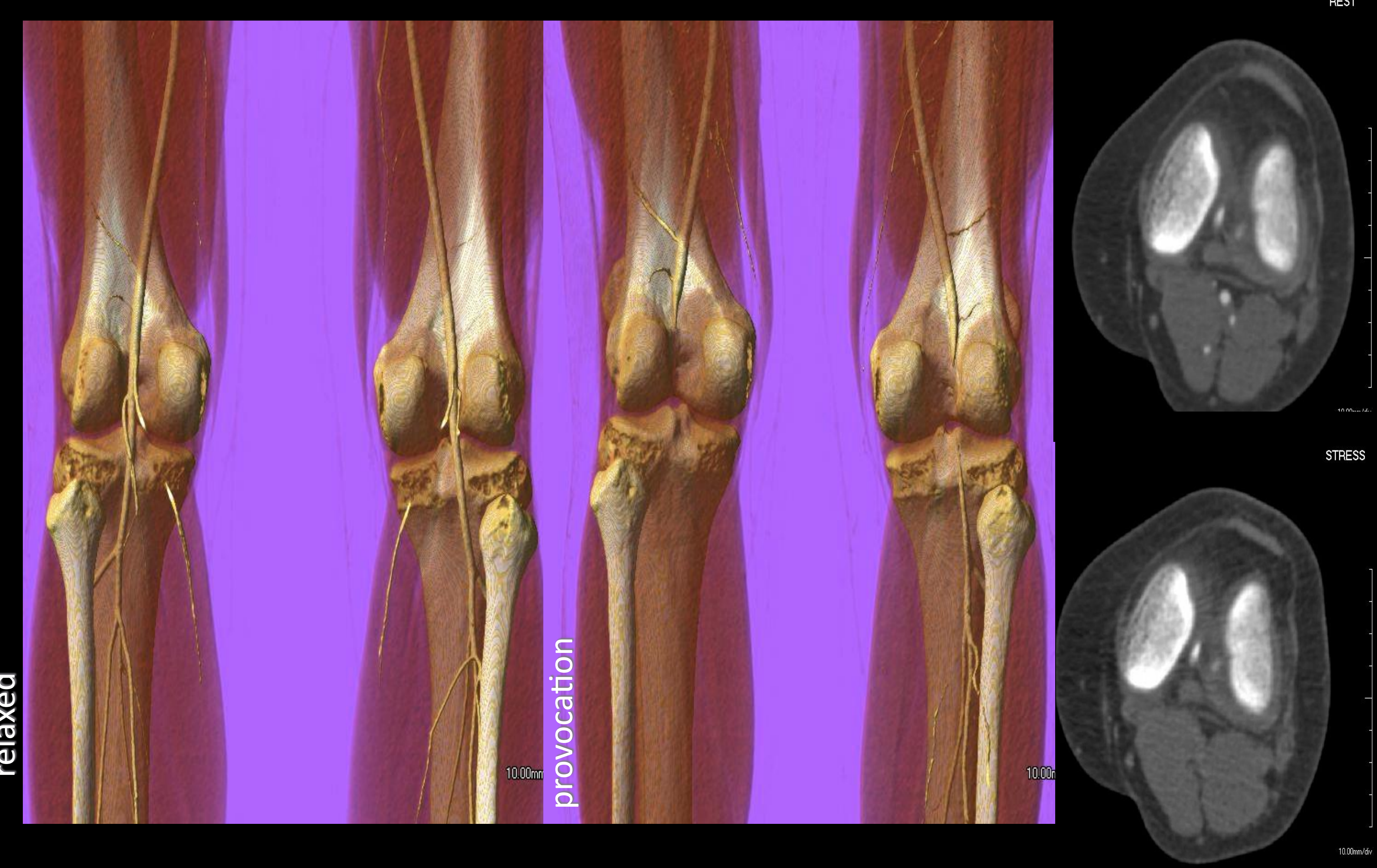


23 M Water Polo Athlete: Type V PAES

- Bilateral accessory slip MHG
- L popliteal a. occlusion @ rest
- Bilateral Popliteal v. thrombosis (Type V)



Functional (Type VI) PAES



Conclusions

- **Hypertrophic Cardiomyopathy** is the #1 cause of sudden death in athletes and young adults. MRI is the gold standard for diagnosis.
- Vascular diseases in athletes can be a significant source of disability (need HIGH index of suspicion)
- Functional imaging is paramount for accurate detection and characterization of vascular entrapment / stenotic syndromes of the shoulder, pelvis, and lower extremities
- CTA allows rapid, functional evaluation of TOS, entrapment syndromes and endofibrosis

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Deirdre Sheahan, MD
Lt. Cmdr. Scott Alexander, MD

Handouts Available:

www.stanford.edu/~hallett

Choose folder "Mayo"