Vascular Atypicals

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Aims

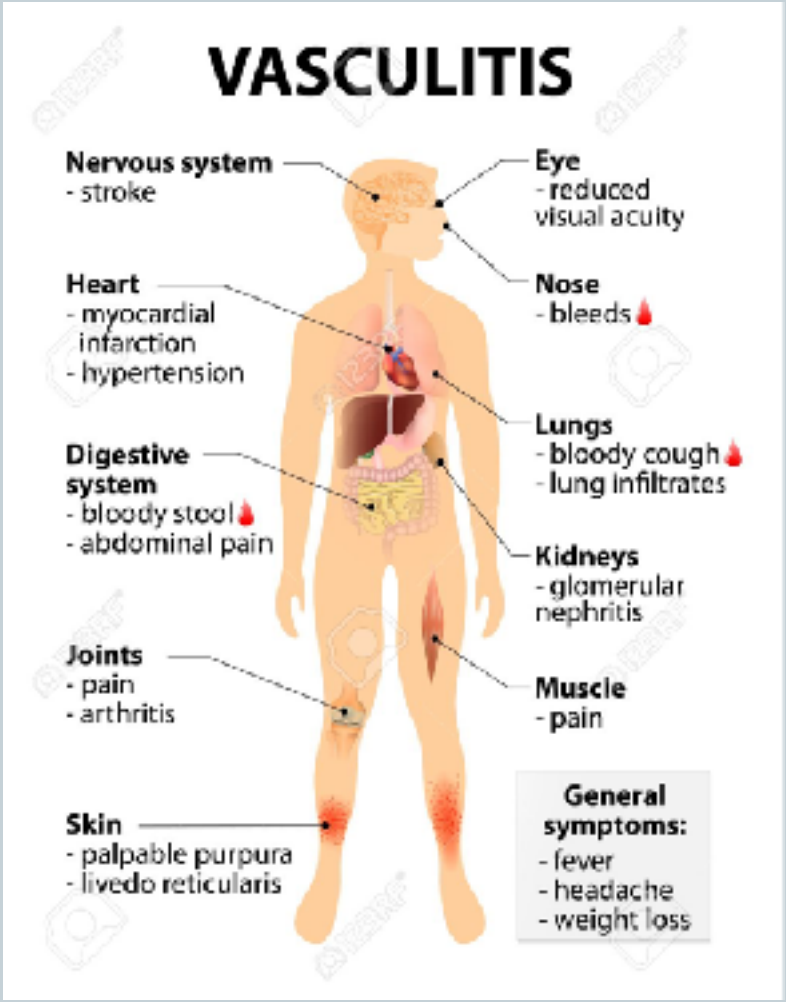
- To cover rare and obscure conditions that may present to the vascular practitioner
- Familiarise ourselves with their presentation, investigation and management.

Atypical Conditions

- Arteritis / Vasculitidies
- Fibromuscular Dysplasia
- Segmental Arterial Mediolysis
- Hypothenar Hammer Syndrome
- Median Arcuate Ligament Syndrome
- The Nutcracker Syndrome
- May-Thurner Sydrome
- Paget-Schroetter Syndrome
- Connective Tissue Disorders – Marfan's, Ehlers-Danlos, Loeys-Dietz Syndrome
- Ergotism

Vasculitis

- Vasculitis is a group of uncommon conditions, which results in inflammation of the blood vessels (predominantly arterial)
- Can be acute and/or chronic
- Vascular surgeons may treat the ischaemic component – fibrosis, stenosis and occlusion resulting in end-organ injury.
- Great number of disorders with unknown aetiology


VASCULITIS
Nervous system
- stroke
Eye
- reduced visual acuity
Nose
- bleeds
Heart
- myocardial infarction
- hypertension
Lungs
- bloody cough
- lung infiltrates
Digestive system
- bloody stool
- abdominal pain
Kidneys
- glomerular nephritis
Joints
- pain
- arthritis
Muscle
- pain
Skin
- palpable purpura
- livedo reticularis
General symptoms:
- fever
- headache
- weight loss

Categorisation

- Broadly Speaking Cutaneous Vasculitis vs Systemic Vasculitis

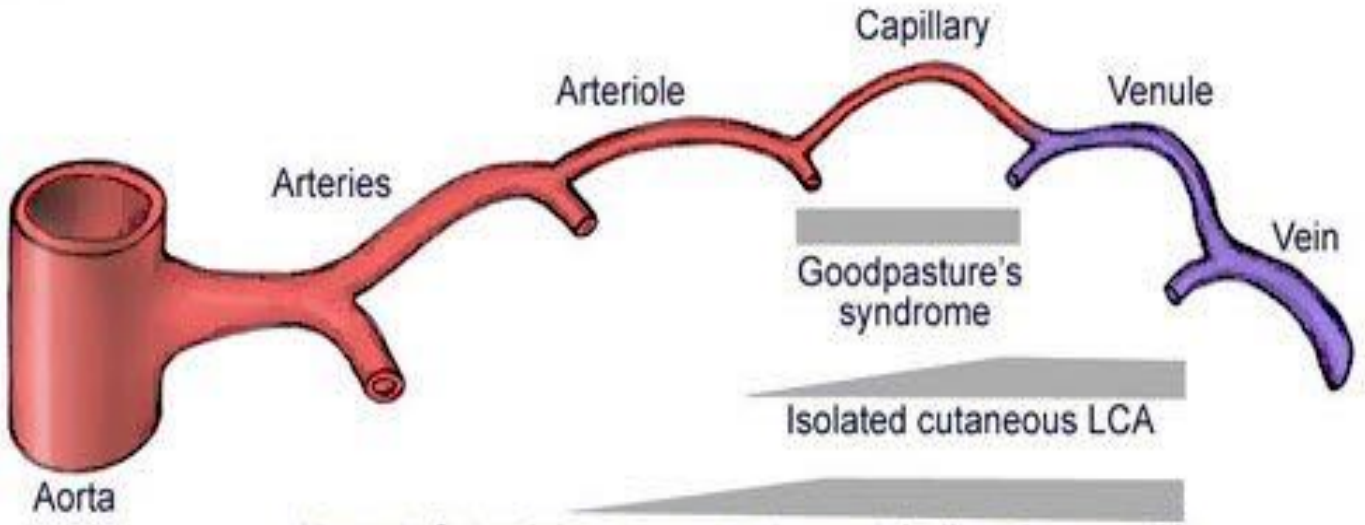
- Systemic is characterised by:
- Constitutional Symptons (malaise, fever, weight loss)
- End-organ symptoms
- Raised ESR/CRP

- Sub divided into vessel size

Small-Vessel Vasculitis
(e.g., microscopic polyangiitis, Wegener's granulomatosis)

Medium-Sized-Vessel Vasculitis
(e.g., polyarteritis nodosa, Kawasaki's disease)

Large-Vessel Vasculitis
(e.g., giant-cell arteritis, Takayasu's arteritis)

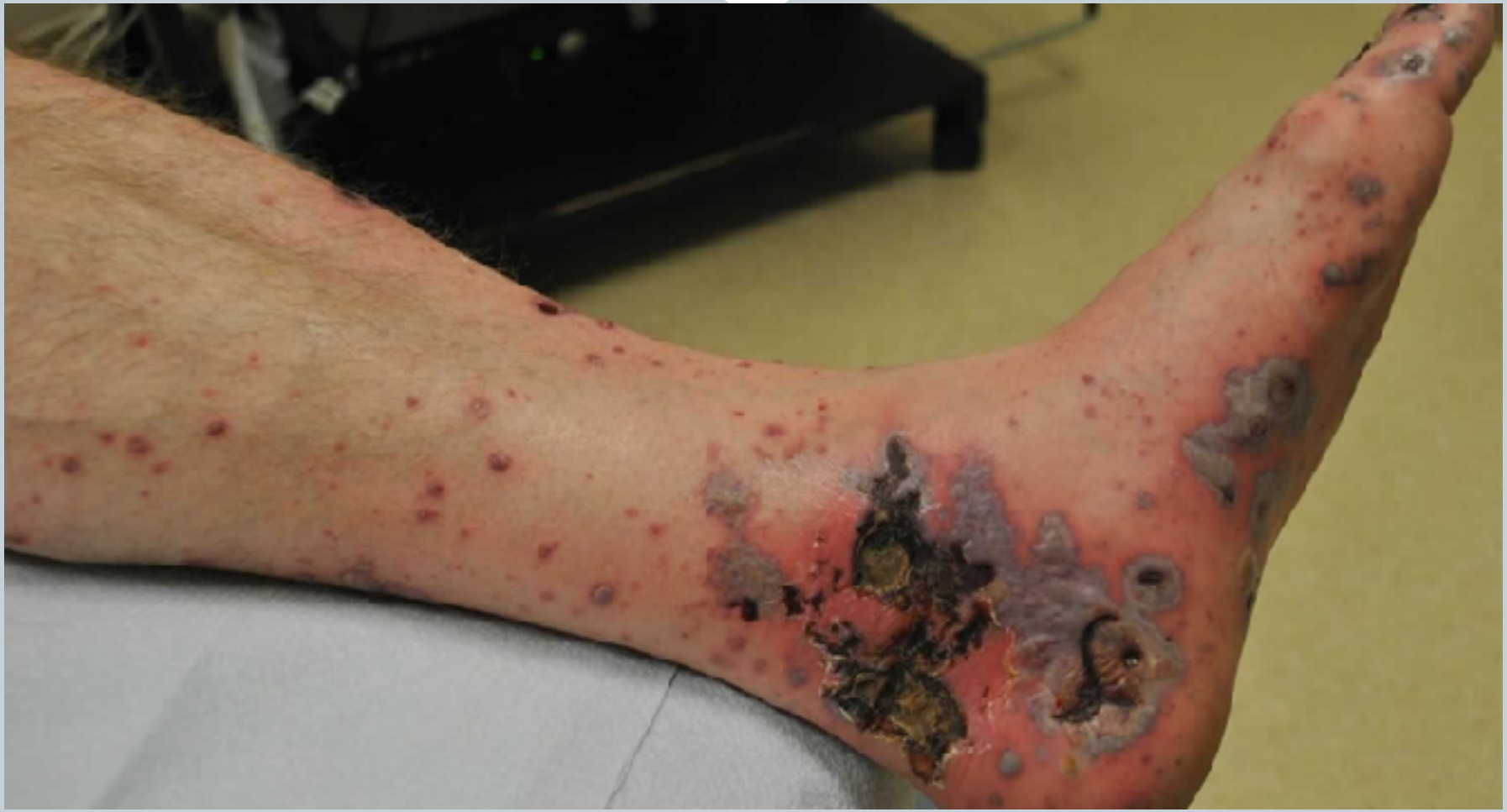


Microscopic polyangiitis, Wegener's granulomatosis, and Churg-Strauss syndrome

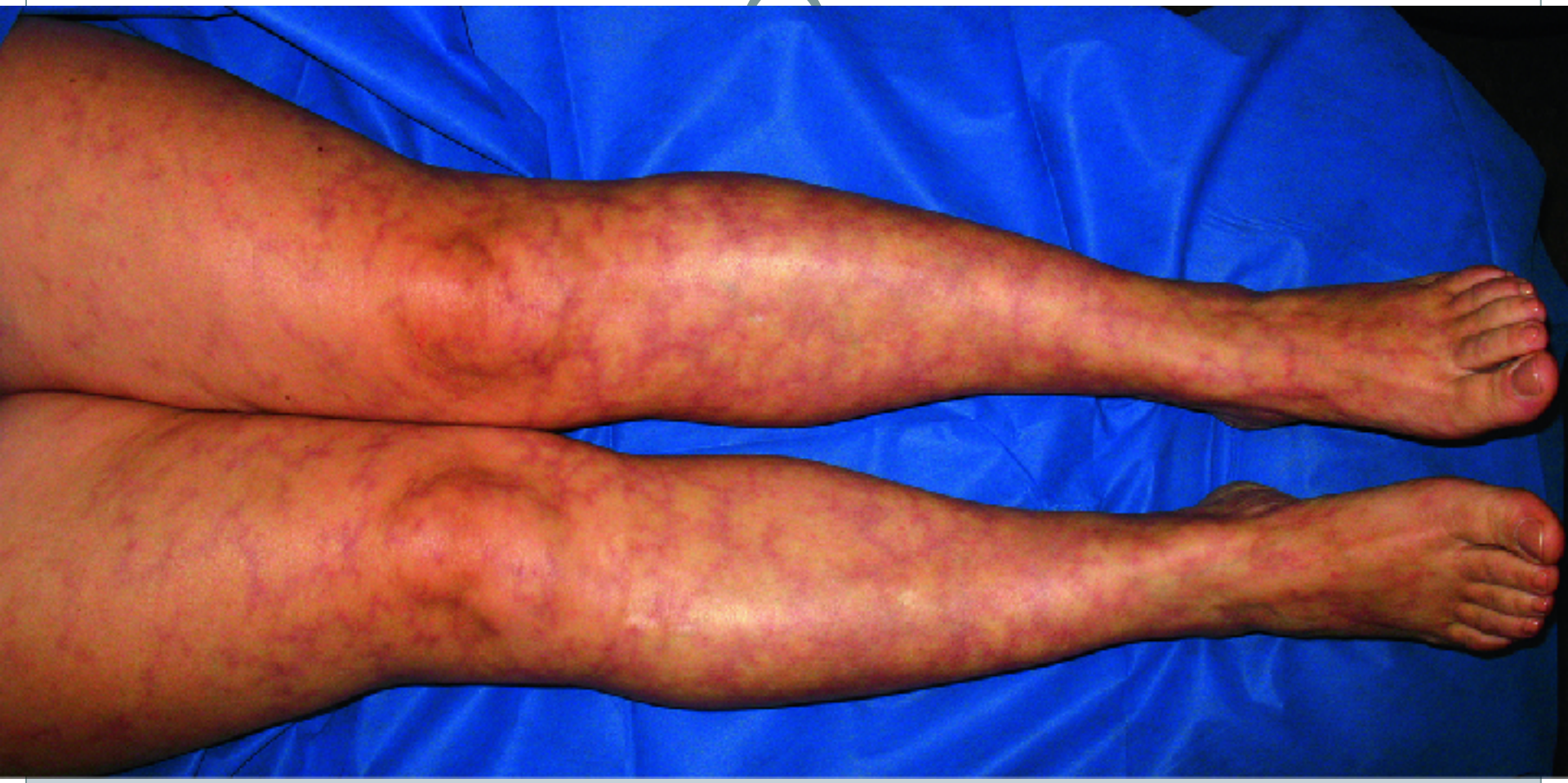
Clinical Picture and Presentation

- Cold hands and feet, cyanosis of digits, ulceration and gangrene.
- Nail fold infarcts and splinter haemorrhages
- Malaise, pyrexia
- Cutaneous Vasculitis
- Rarely aneurysmal dilatation

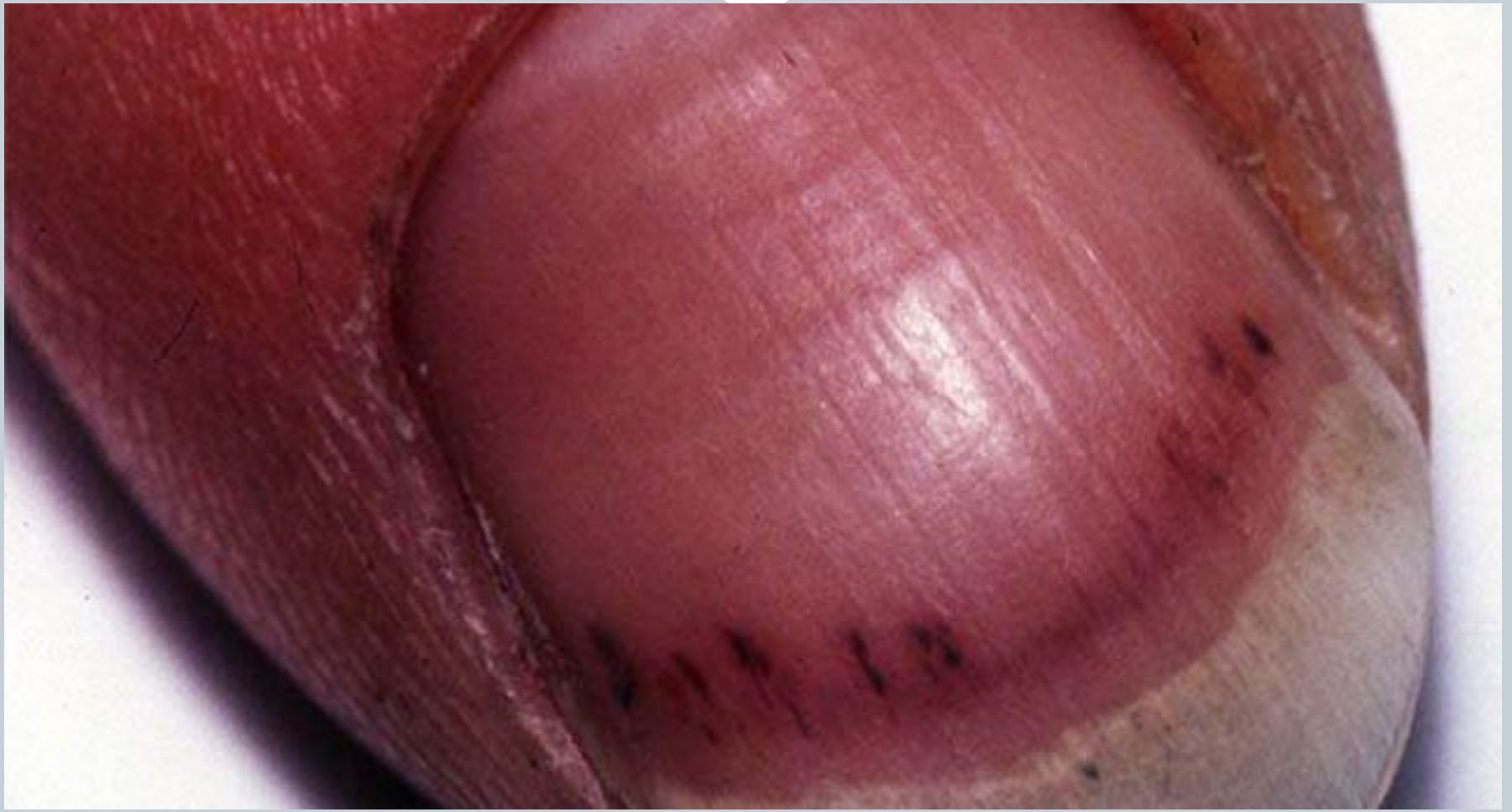
Cutaneous Vasculitis



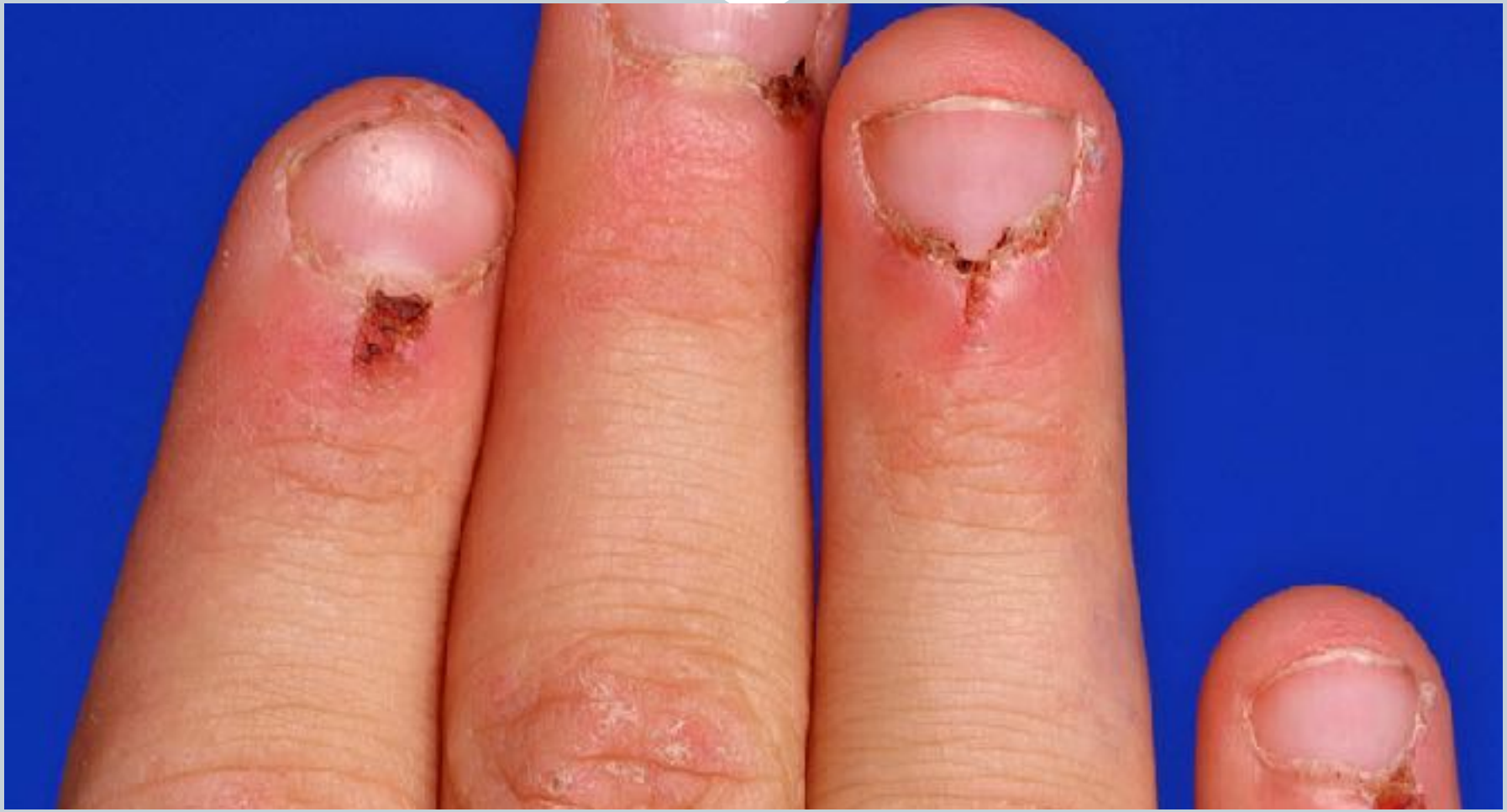
Livedo Reticularis



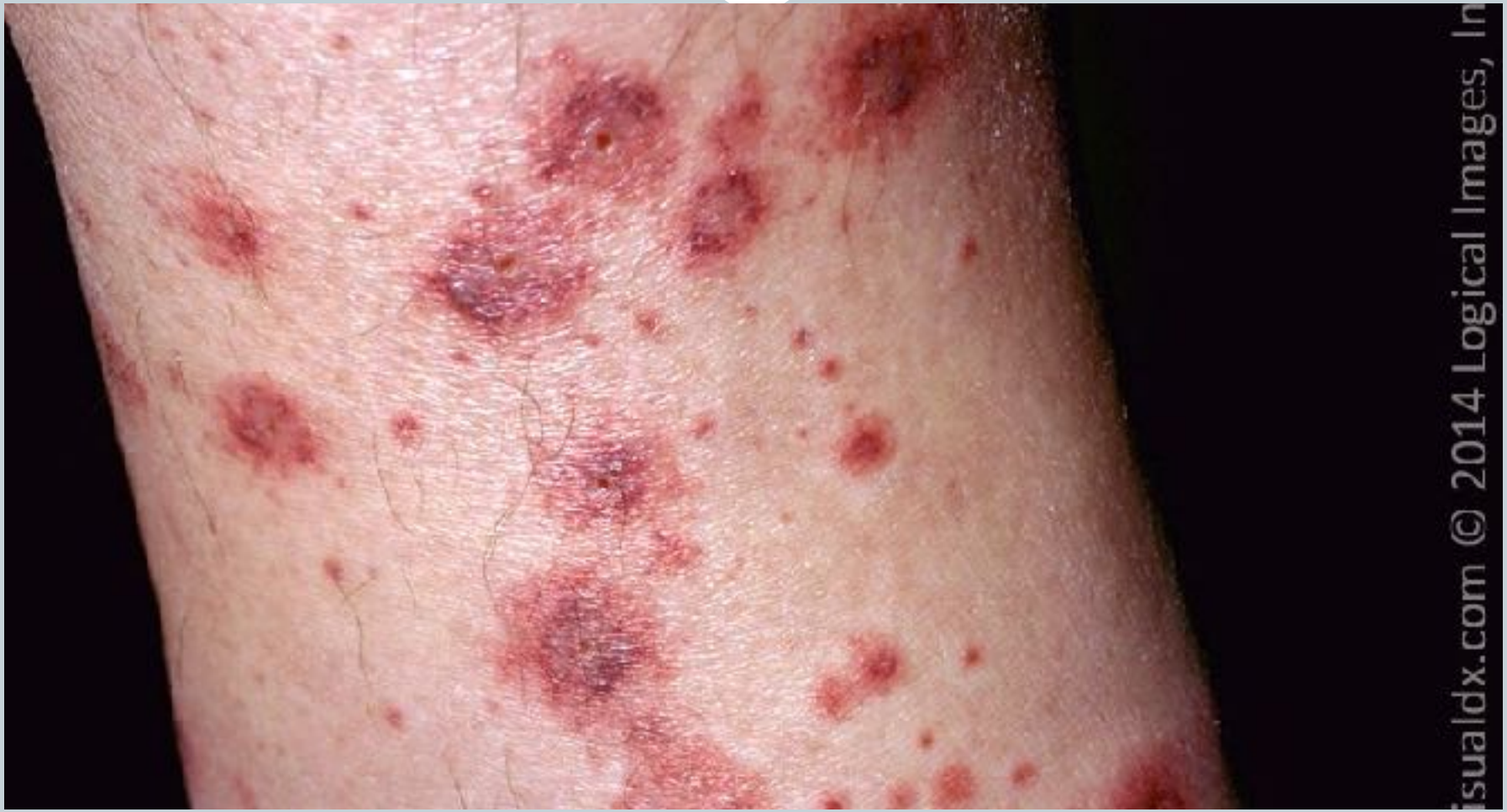
Splinter Haemorrhages



Nail-fold infarcts



Palpable Purpura



Investigations

- Clinical Findings
- Basic Serology – FBC, U&E, LFT's, TFT's, inflammatory markers (CRP, ESR, serum osmolality).
- Autoantibodies – Over 100 known. Interpretation takes experience.
- Diagnostic Imaging
- - CXR (cervical rib, Wegener's, rheumatoid disease)
- -Duplex: Halo Sign in Temporal Arteritis
- -Angiography
- Histopathology – microscopy or direct immunofluorescence.

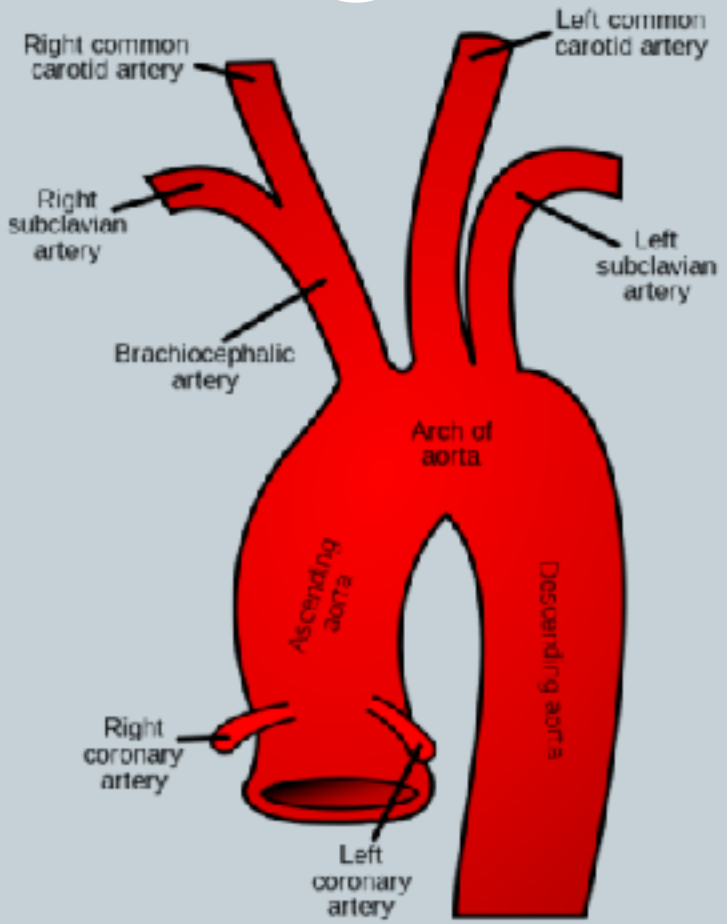
Management

- In mild cases – manage conservatively
- General Management – Smoking cessation especially Buerger's. Keep warm. Rest and elevation. Best medical therapy as per atherosclerosis.
- Treatment of Malperfusion - Revascularise
- Treatment of Systemic Inflammatory Component – Steroids, Cyclophosphamide, DMARD's etc.

Specific Conditions

- Takayasu's Arteritis
- Giant Cell Arteritis (temporal arteritis)
- Buerger's Disease
- Polyarteritis Nodosa (PAN)
- Cutaneous Vasculitis/Small Vessel Disease
- Behcet's Disease

Takayasu's Arteritis



Takayasu's Arteritis: Clincal Features

- Prevalent in South-East Asia. F:M 8:1, 10-30yrs
- An inflammatory and obliterative arteritis primarily affecting large and elastic arteries

- Two Phases
- 1. Acute Systemic Phase/Pre-pulseless phase with constitutional features (malaise, fever, weight loss).

- 2. Chronic Obliterative Phase – Vessel obliteration, stenosis and fibrosis. Hyptertension is common.

Takayasu's Arteritis: Diagnosis

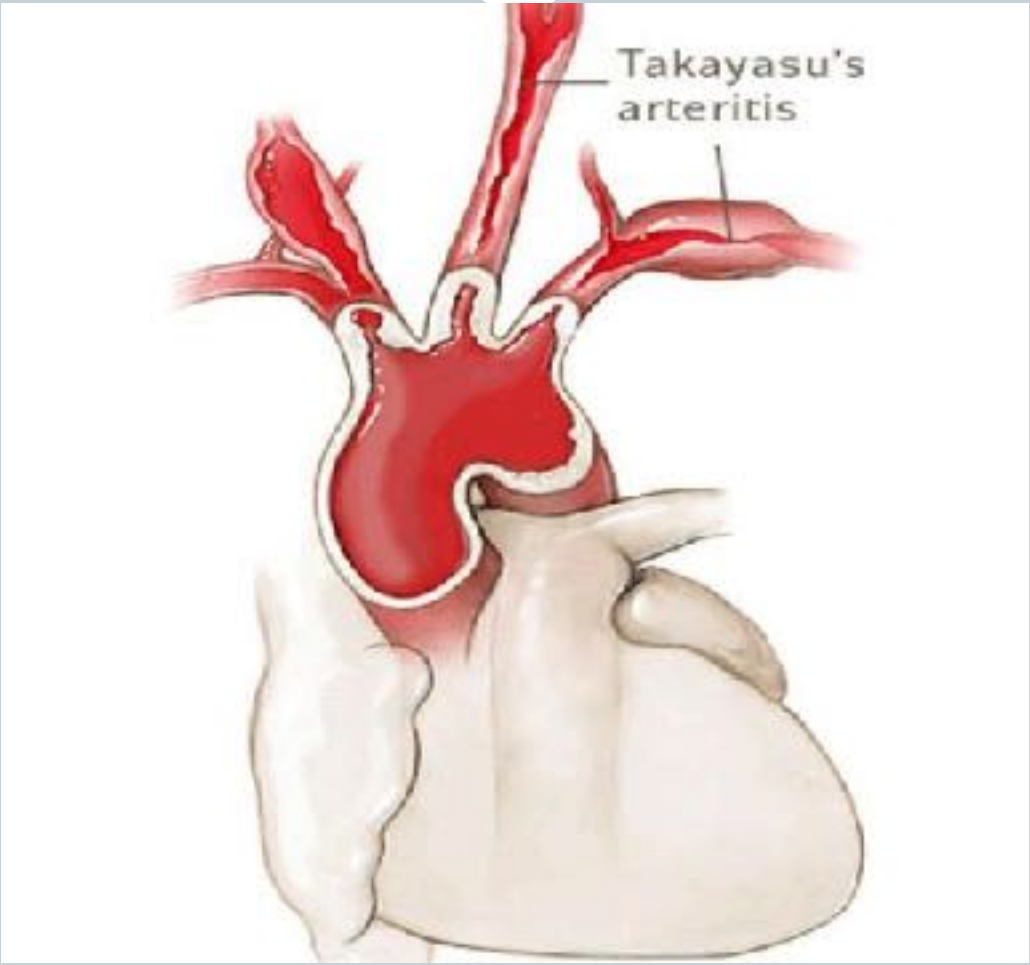
- Clinical Findings – May have reduced upper limb blood pressure.
- Basic Serology – ESR is elevated in most patients.
- Autoantibodies – None.
- Diagnostic Imaging – CE MRA. Image entire aorta.
- Histopathology – Inflammation of all three layers of the vessel wall, chronic granulomatous appearance.

Takayasu's Arteritis: Diagnostic Criteria

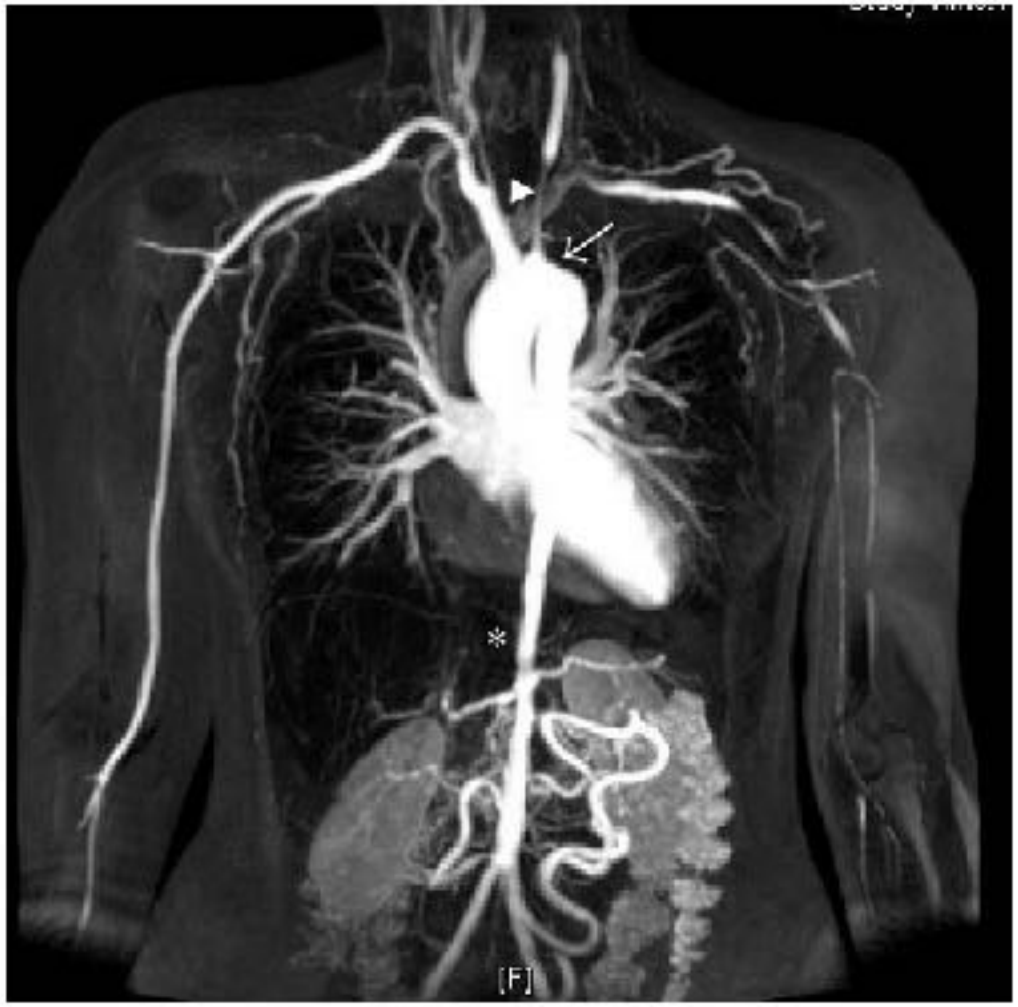
- American College of Rheumatology.
- -Onset <40 years
- -Claudication of an extremity
- -Decreased brachial artery pulse
- ->10mmHG difference between arms
- -Bruit over subclavian or aorta
- -Radiographic evidence of narrowing or occlusion of the entire aorta, it's primary branches, or large arteries in the proximal upper or lower extremities

- Sensitivity 90.5% Specificity 97.8%

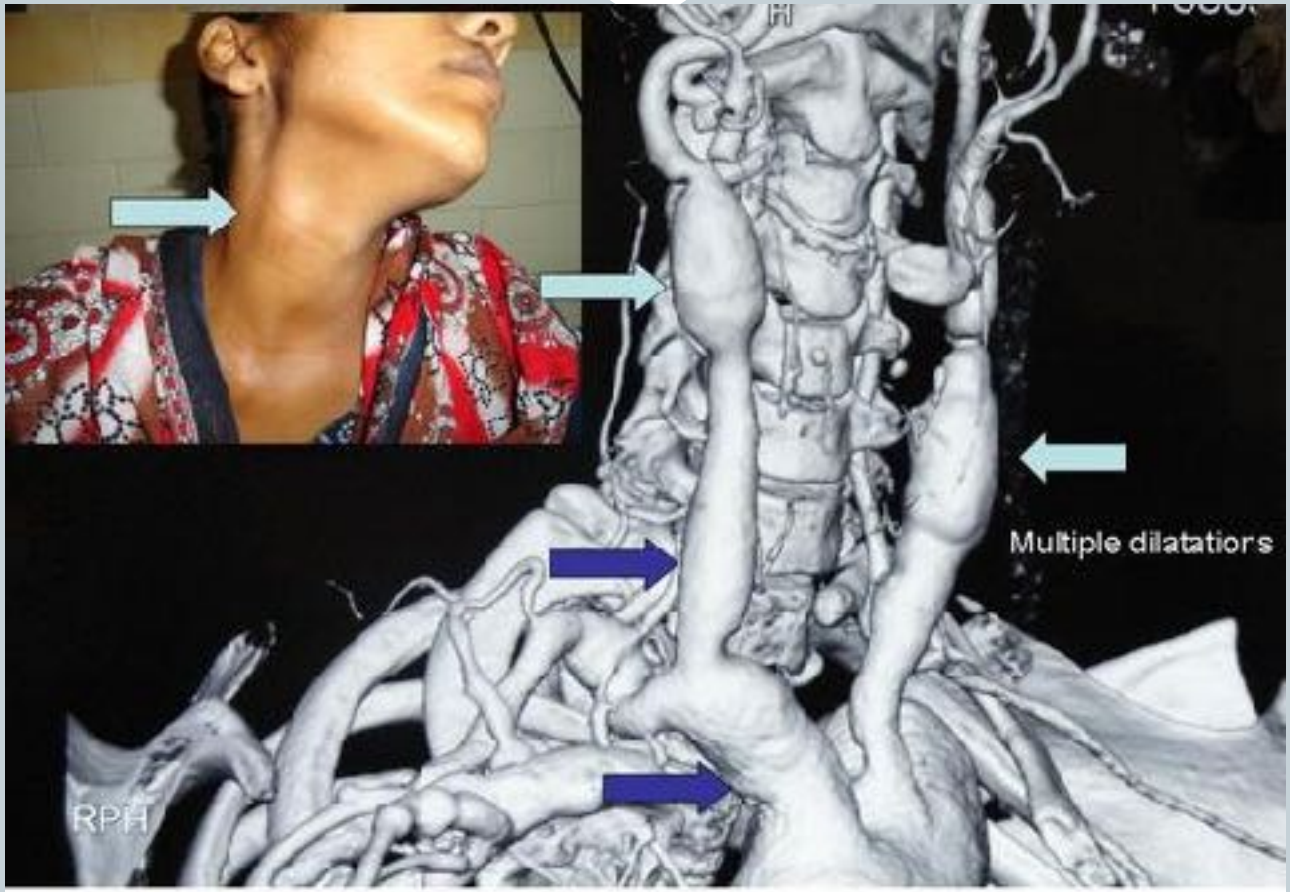
Takayasu's Arteritis



Takayasu's Arteritis



Takayasu's Arteritis



Large Vessel Vasculitis – Multisegmental involvement

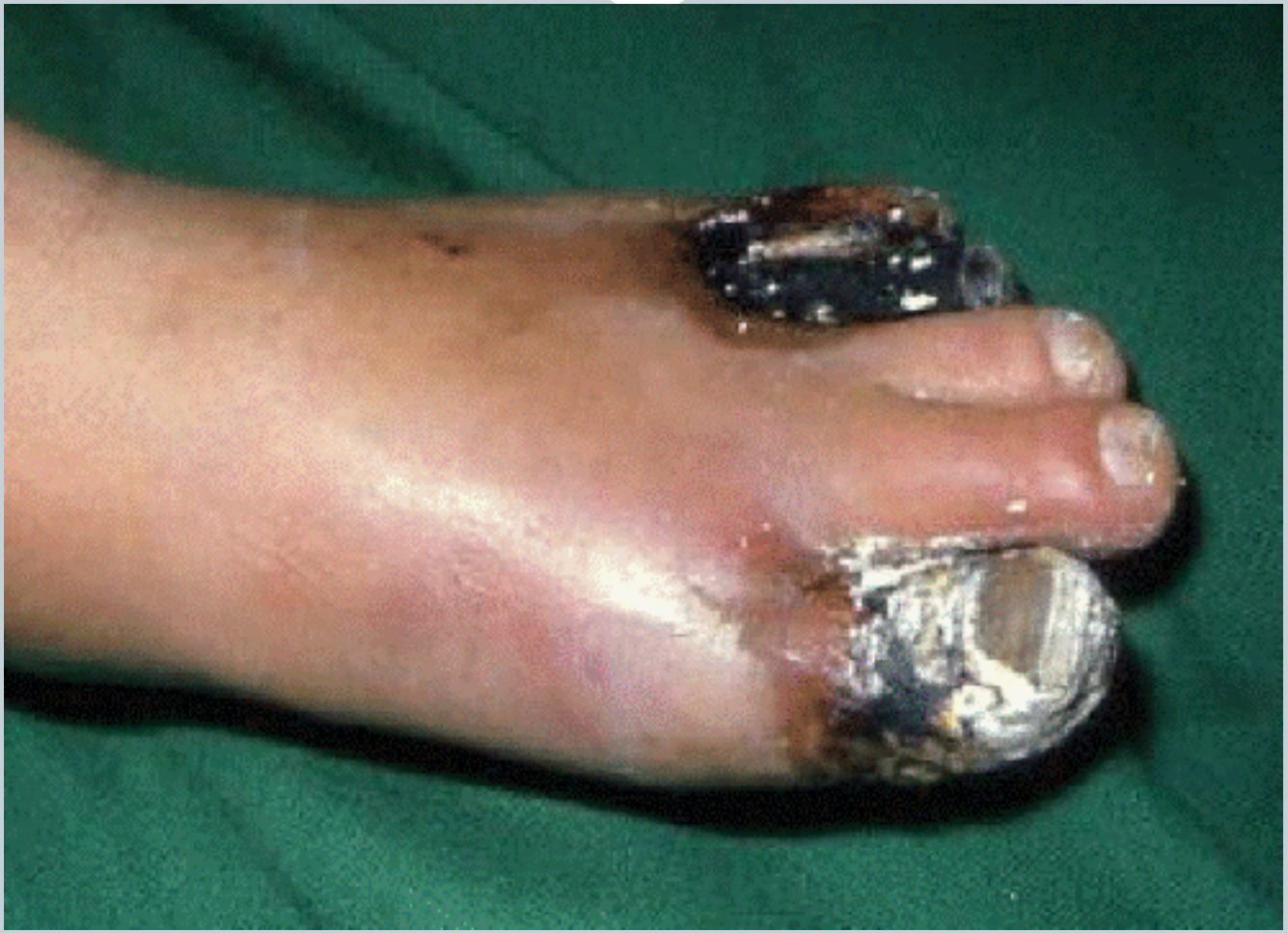
Takayasu's Arteritis: Treatment

- General Measures: Stop smoking. Keep hands warm. Aspirin/Clopidogrel.
- Treatment of malperfusion – can treat only, as symptomatically required, once acute phase is over.
- Treatment of systemic inflammation – Anti-inflammatories & steroids, escalated as necessary.

Takayasu's Arteritis: Malperfusion

- 2018 Meta-analysis Open vs Endovascular approaches to revascularisation
- 770 patients with 1363 lesions. 389 Endo vs 420 Open
- Restenosis more common in Endo OR 5.18 ($P<0.001$)
- Stroke less common in Endo OR 0.33 ($p<0.003$).

Buerger's Disease (thromboangitis obliterans)



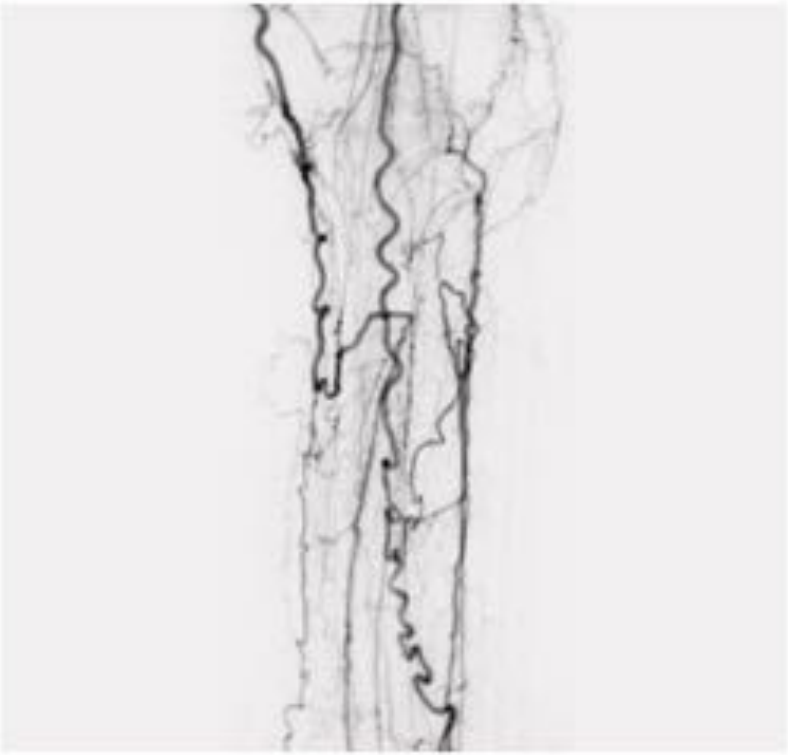
Buerger's Disease: Clincal Features

- Largely a disease of young male smokers (<40)
- Non-atherosclerotic, inflammatory and thrombotic occlusive disorder
- Affects small and medium vessels (both arteries and veins)
- Lower limbs (>95%) and upper limbs (25%)

Buerger's Disease: Diagnosis

- Clinical Findings – Foot claudication, digital ischaemia, cyanosis, shiny skin, hair loss, rest pain
- Basic Serology – Non specific
- Autoantibodies - None
- Diagnostic Imaging – Angiography shows abrupt vessel cut off and typical corkscrew collaterals.
- Histopathology – Hypercellular thrombus infiltrated with lymphocytes, fibroblasts and later giant cells.

Buerger's Corkscrew Appearances



Angiography showing corkscrew appearance of vessels

Buerger's Disease

- Olin 2000 proposed this diagnostic criteria (NEJM):
- 1. Age 20-40, predominantly male
- 2. Current, or recent, tobacco use.
- 3. Distal extremity ischaemia (claudication, rest pain, gangrene), documented by imaging.
- 4.Exclusion of other autoimmune diseases, hypercoagulable states and DM
- 5. Exclusion of a proximal source of emboli by ECHO and arteriography
- 6.Consistent arteriographic findings in the clinically involved and noninvolved limbs

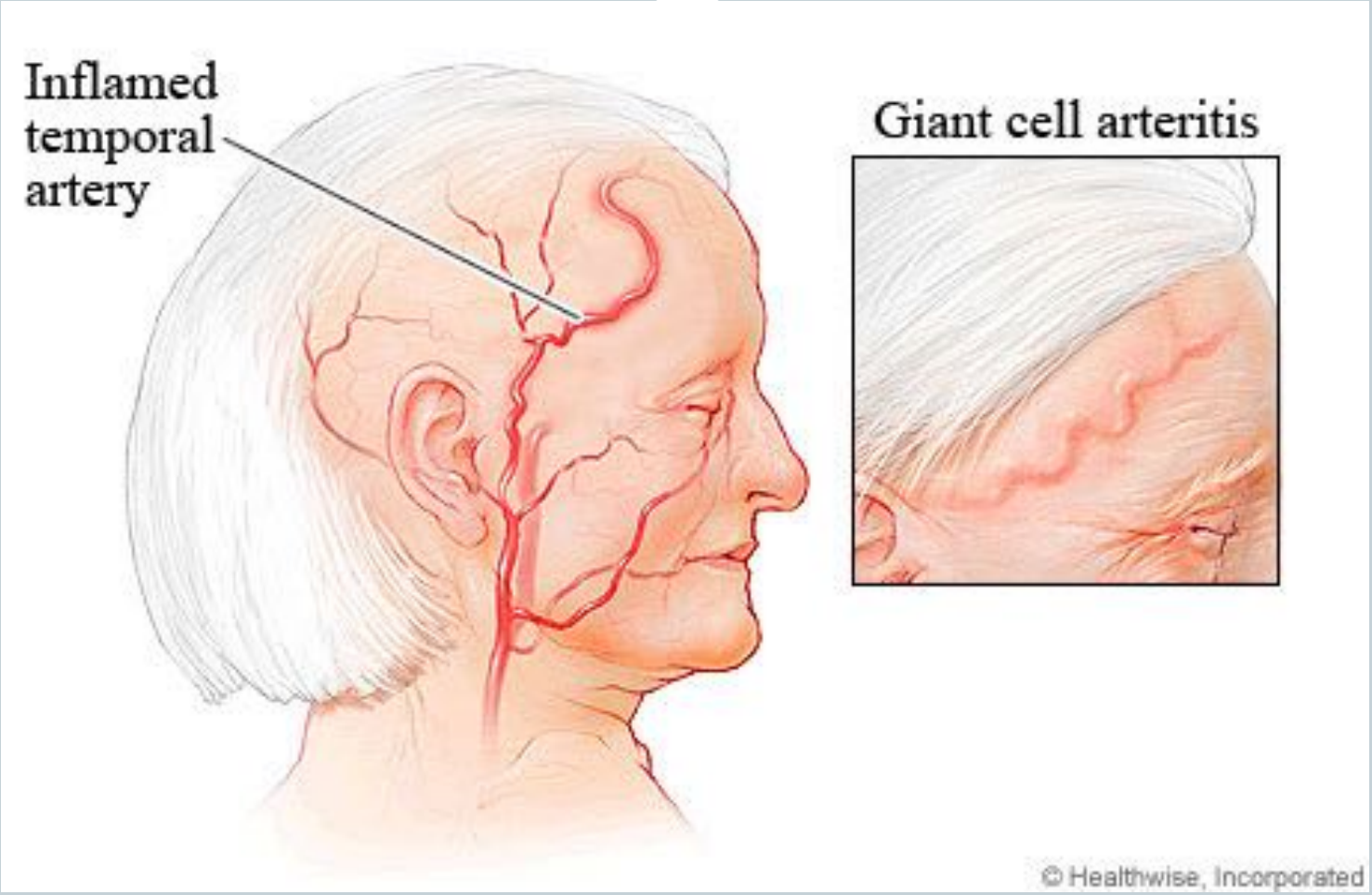
Buerger's Disease: Treatment

- General – STOP SMOKING. Keep digits warm. Exercise. Vasodilators to improve blood flow – iloprost.
- Treatment of malperfusion - Typically no target vessel. A sympathectomy may be beneficial later on.
- Treatment of systemic inflammation – Unlikely to be of significant benefit.

Buerger's Disease

- Retrospective clinical audit 2018, Journal of Diving Metabolic Medicine
- 97 patients with non healing painful ulcers secondary to Buerger's treated with hyperbaric oxygen therapy
- Significant improvement 10 months after starting treatment – both for wound healing and rest pain control

Giant Cell Arteritis



Giant Cell Arteritis: Clinical Features

- Disease of the middle aged and elderly
- 1:15,000 people >50 per year
- F:M 5:1
- Affected small and medium sized arteries
- Common in smokers and pre-existing atherosclerosis

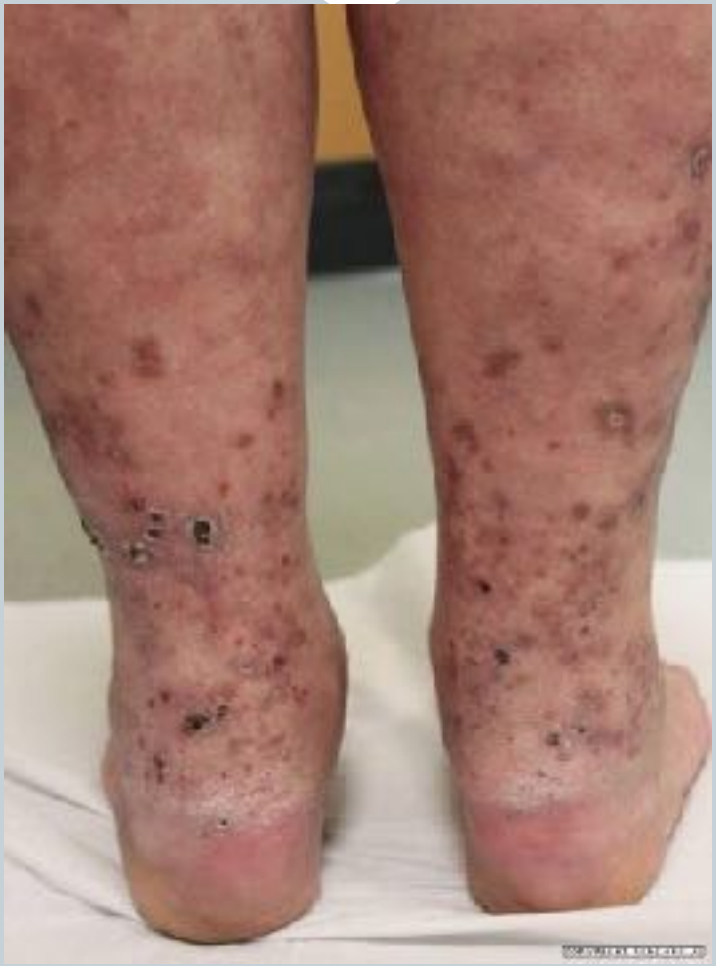
Giant Cell Arteritis: Diagnosis

- Clinical Findings – Non-specific systemic involvement. Localised end-organ symptoms.
- Basic Serology – ESR is typically but not always elevated.
- Autoantibodies - None
- Diagnostic Imaging – Duplex may show a halo sign.
- Histopathology – Gold Standard showing a granulomatous arteritis. Remember skip lesions = 50% negative sampling.

Giant Cell Arteritis: Treatment

- General – STOP SMOKING. No evidence for Aspirin.
- Treatment of malperfusion - None
- Treatment of systemic inflammation – Steroid are the mainstay of treatment, escalated if required. Treatment is initiated before pathology results due to the risk of blindness. Maintain for around two years.

Polyarteritis Nodosa



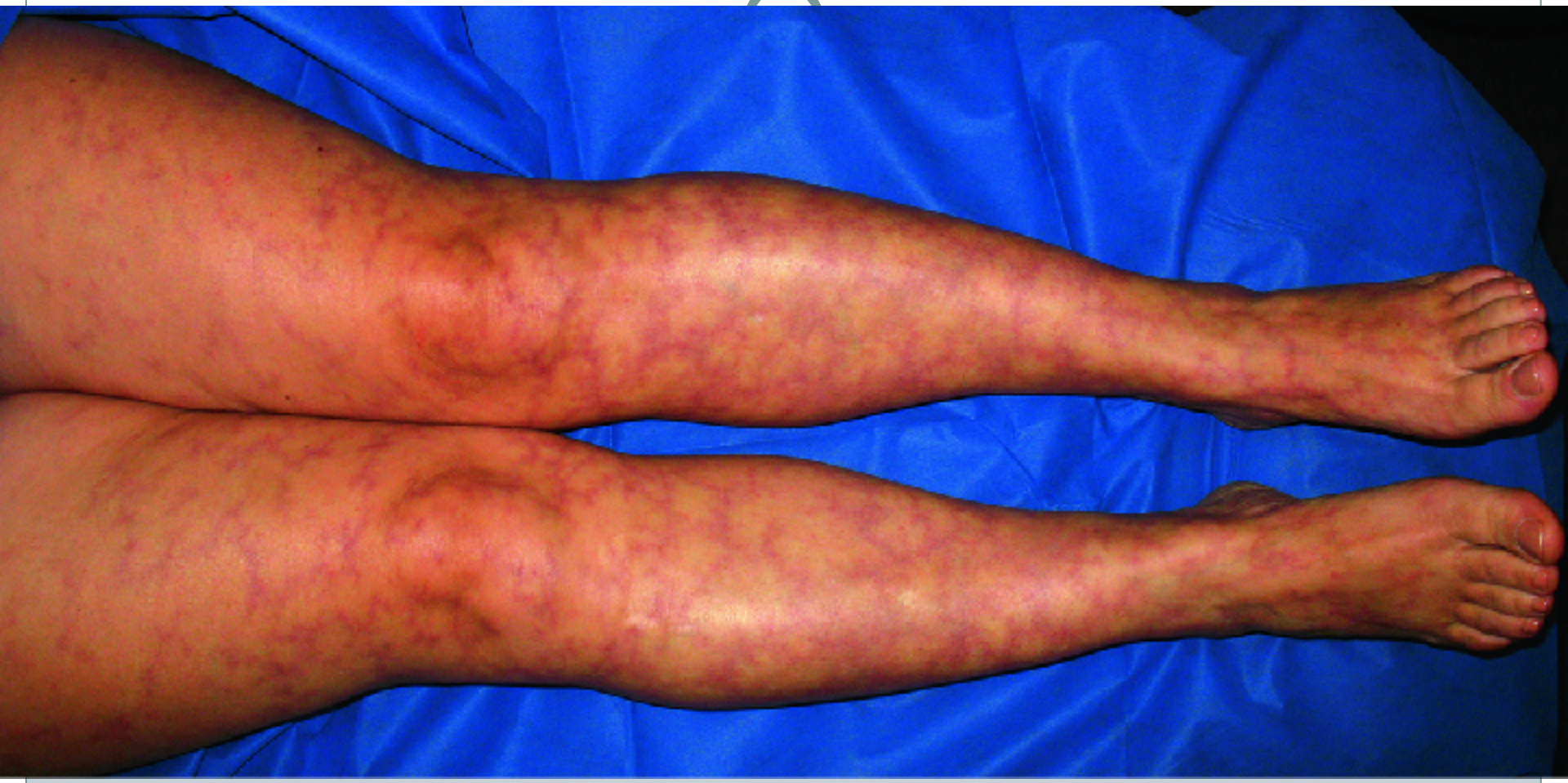
Polyarteritis Nodosa: Features

- A disease of the middle-aged (40-60)
- Male preponderance 2:1
- It is a systemic necrotising vasculitis affecting small and medium-sized arteries
- Strong link with Hepatitis B Infection

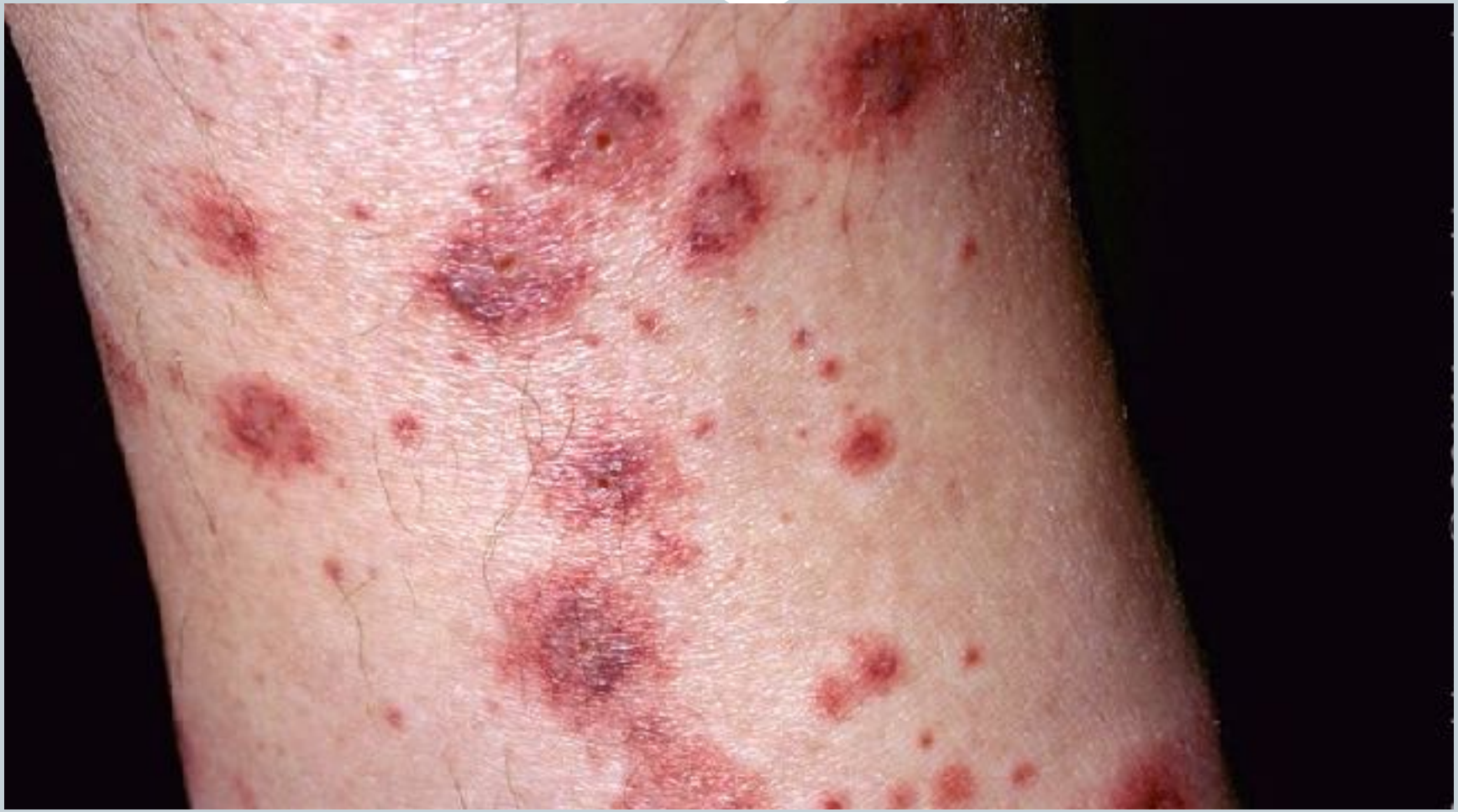
Polyarteritis Nodosa: Clinical Findings

- 1. Cutaneous Polyarteritis Nodosa (25-50%) patients
- Palpable Purpura
- Livedo reticularis
- Ulceration
- Digital Infarcts

Livedo Reticularis



Palpable Purpura



Polyarteritis Nodosa: Clinical Findings

- 2. Microscopic Polyangitis
- 40% also have c-PAN

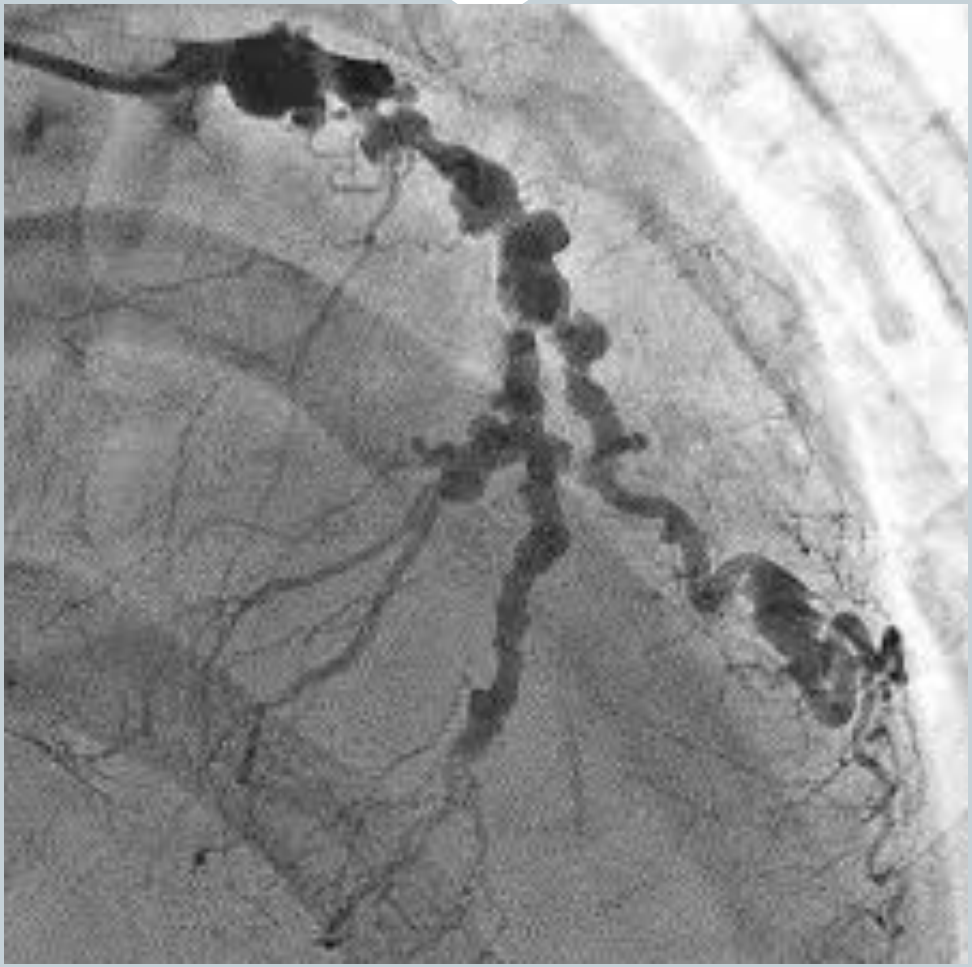
- Systemic features

- End-organ dysfunction
- Renal 70%
- Pulmonary
- Gastrointestinal – Pain and vomiting.
- Acute Events – Bowel ischaemia, perforation, haemorrhage.
- Aneurysmal Formation
- Testes and Retina

Polyarteritis Nodosa: Diagnosis

- Basic Serology – Non specific, ESR may be elevated
- Autoantibodies – pANCA, HepBsAG
- Diagnostic Imaging – Saccular or fusiform aneurysm.
- Histopathology – Gold Standard showing fibrinoid necrosis of the vessel wal, microaneurysms, thrombosis and tissue infarction.

Polyarteritis Nodosa



Polyarteritis Nodosa: Treatment

- General Management – Smoking cessation. Keep warm. Rest and elevation
- Treatment of Malperfusion – No evidence for BMT
- Treatment of Systemic Inflammatory Component – Steroids escalated as required.

Cutaneous Vasculitis

- Primarily small vessels and post-capillary venules.

- Idiopathic Cutaneous Vasculitis:
- -Most common form of vasculitis in the skin
- -Palpable purpura below knee most frequently
- -Tends to be symmetrical, worse on sitting
- -Lesions appear as crops, macular erythema then to purpura

- Biopsies

Cutaneous Vasculitis

- Necrotising Vasculitis Secondary to :
- -Drugs e.g. Abx, Diurectics, NSAIDs, Anticonvulsants
- -Infection, e.g. URTI, Streptococcus, Hep B, HIV
- -Immunological disorders e.g. connective tissue disorders

Cutaneous Vasculitis

- Cutaneous manifestations of systemic disease
- -Connective Tissue Disorder
- -Mixed Cryogolbulinaemia
- -Allergic granulomatosis (Churg-Strauss)
- -Behcet's disease
- -HIV

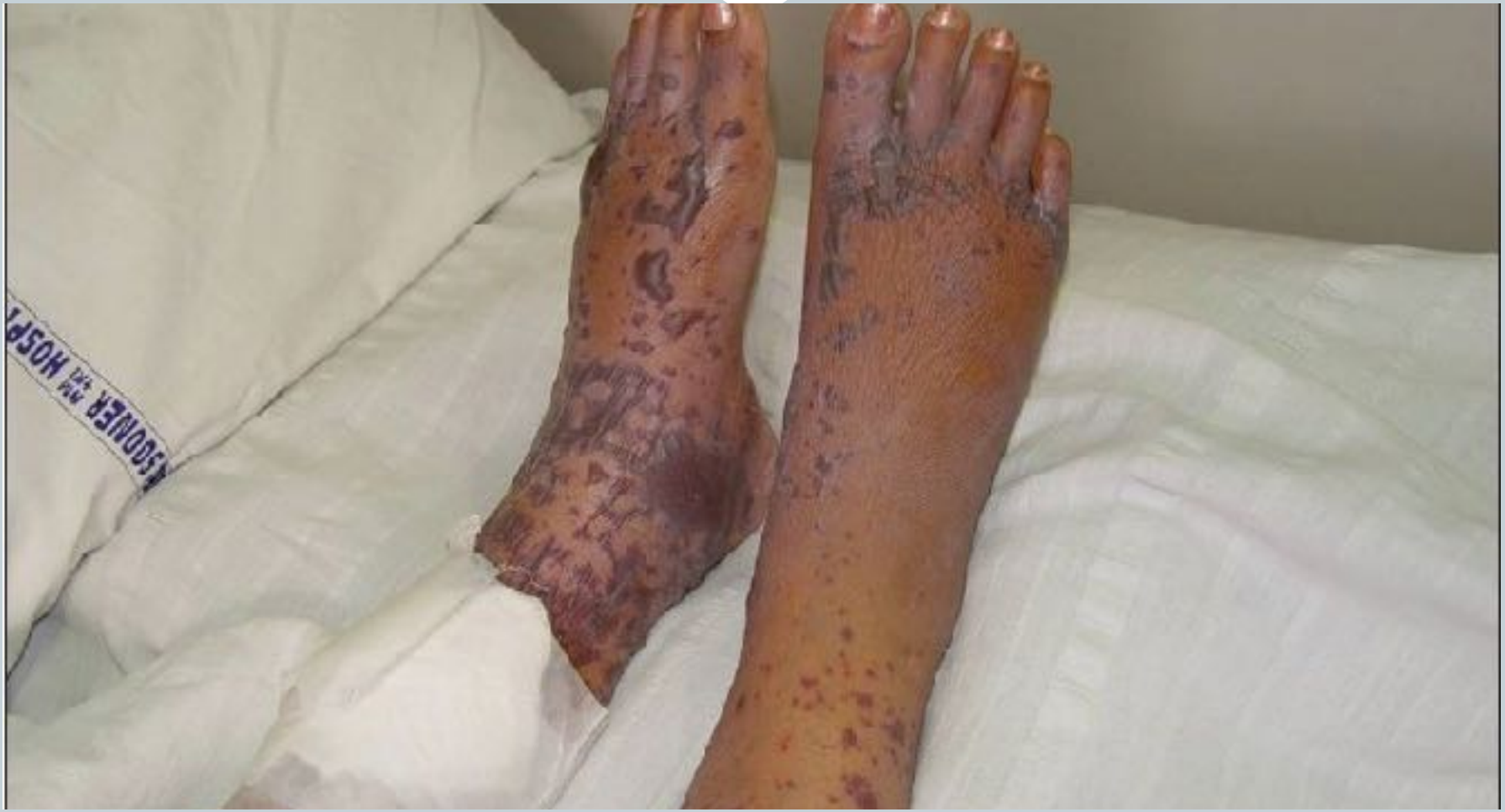
Cutaneous Vasculitis: Treatment

- Most are short lived
- Limited to the skin
- Biopsy confirms the diagnosis
- NSAIDs or Anti-histamines, steroids rarely.

Churg-Strauss

- A macropopular vasculitis in the extremities, often with vesicles and bullae
- An eosinophilic granulomatosis with polyangitits, affects small and medium sized vessels, in patients with a history of airway sensitivity.
- Treated with steroids

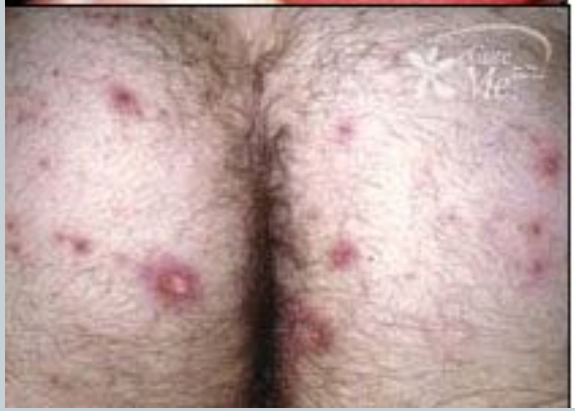
Churg-Strauss

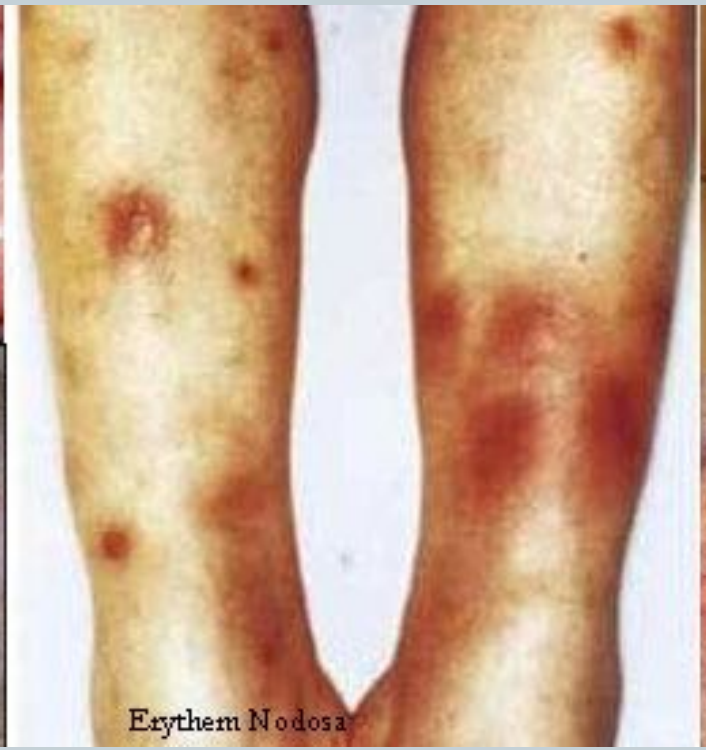


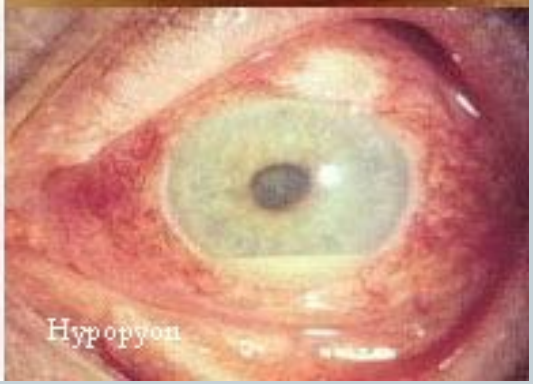
Behcet's Disease / Silk-Road Disease

- A multisystem recurrent vasculitis
- Affects small/medium sized vessels – 90% venous system. Causes DVT, thrombophlebitis & Budd-Chiari
- Asian and middle-eastern populations – HLA B51
- Classical Triad of oral/genital ulceration, chronic uveitis and vasculitis
- Standard treatments

Behcet's



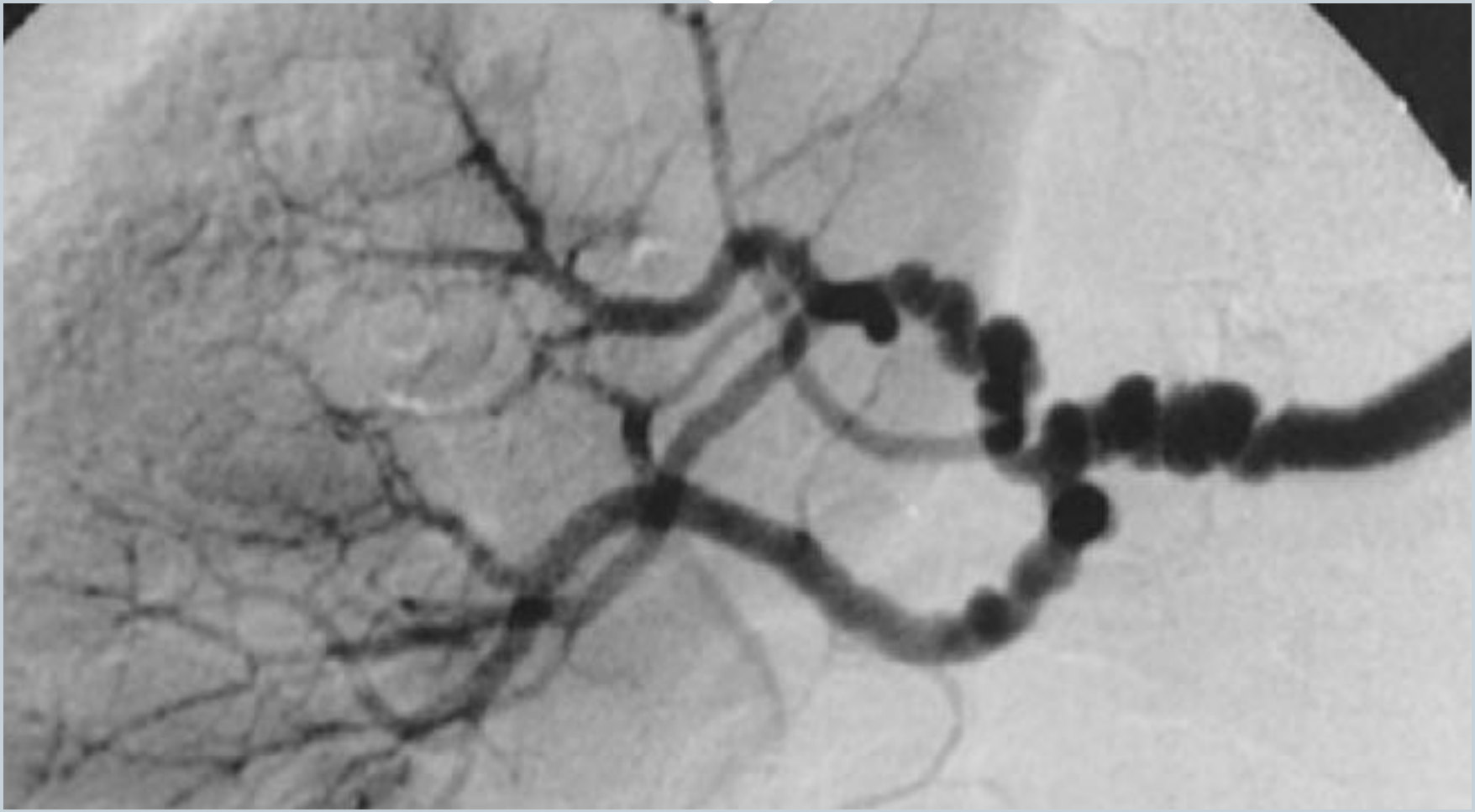




Vasculitis Summary

- May present to the vascular surgeon as skin ischaemia
- Constitutional symptoms of fever, malaise, weight loss and pyrexia are often clues to the diagnosis
- Key investigations are ESR, CRP, plasma viscosity, autoantibodies and biopsy.
- Immunnosuppression is the cornerstone treatment of vasculitis

Fibromuscular Dysplasia



Fibromuscular Dysplasia

- First described 1938, Leadbetter & Burkland
- Non atherosclerotic, noninflammatory disorder
- Affects Renals (60-75%), Carotids (25-30%)
- Unknown aetiology and prevalence
- 5 Histological Subtypes - Intimal Medial (80%) and Adventitial fibroplasia
- 3 Radiological Subtypes – i.) Alternating stenoses & aneurysms (80%), ii.) unifocal or multifocal tubular stenoses, iii.) thinning of the vessel with saccular aneurysmal dilatation

Fibromuscular Dysplasis

- FMD registry 2012, 447 patients.
- Affects females to males 9:1, mainly middle aged
- Mean age at first symptom 47 (5-83)
- Mean age at diagnosis 51

Fibromuscular Dysplasia: Common Presenting Symptoms

- Hypertension 63%
- Headache 52%
- Pulsatile Tinnitus 27.5%
- Dizziness 26%
- Cervical Bruit 22%
- Cervical Artery Dissection 12%
- Hemispheric TIA 8.7%
- Stroke 6%
- Renal Artery Dissection 3%
- Asymptomatic 5.6%

Fibromuscular Dysplasia

- Co-Morbidities
- -Hypertension 72% starts age 43.
- -Significant Headaches 60%, Migraine 32%, Weekly 13%, Daily 12%
- -Post menopausal 60%
- -History of OCP 69%
- -Hyperlipidaemia 42%
- -Smoking 37%

- FH 7-10%

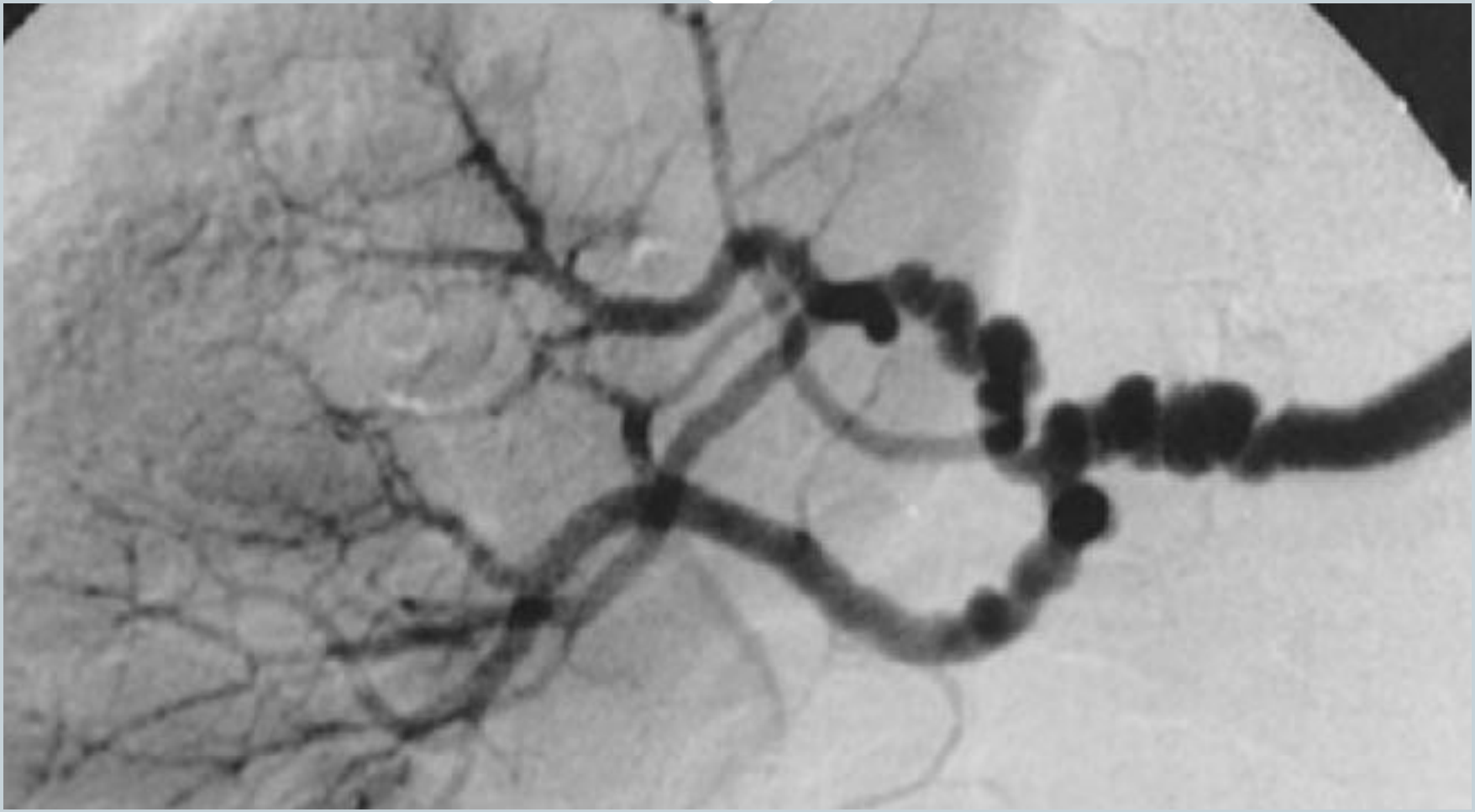
Fibromuscular Dysplasia

- Physical Examination:
- BMI of around 25
- B.P within normal limits
- Horner Syndrome 12%
- Cranial Nerve Abnormalities 9%
- Carotid Bruits 30%

Fibromuscular Dysplasia: Arterial Distribution

- Renal 79%
- Extracranial Carotid 74%
- Vertebral 36%
- Mesenteric 26%
- 60% of cases are bilateral.

Fibromuscular Dysplasia



Fibromuscular Dysplasia: Vascular Events

- -Coronary events are uncommon
- -A cerebrovascular event, including stroke/TIA (19%) or amaurosis (6%), is common
- -Renal and mesenteric ischaemia is rare
- -Dissection is found in 20% of patients (carotids 75%)
- -Aneurysms 17% - Renal 32%, Carotid 21%

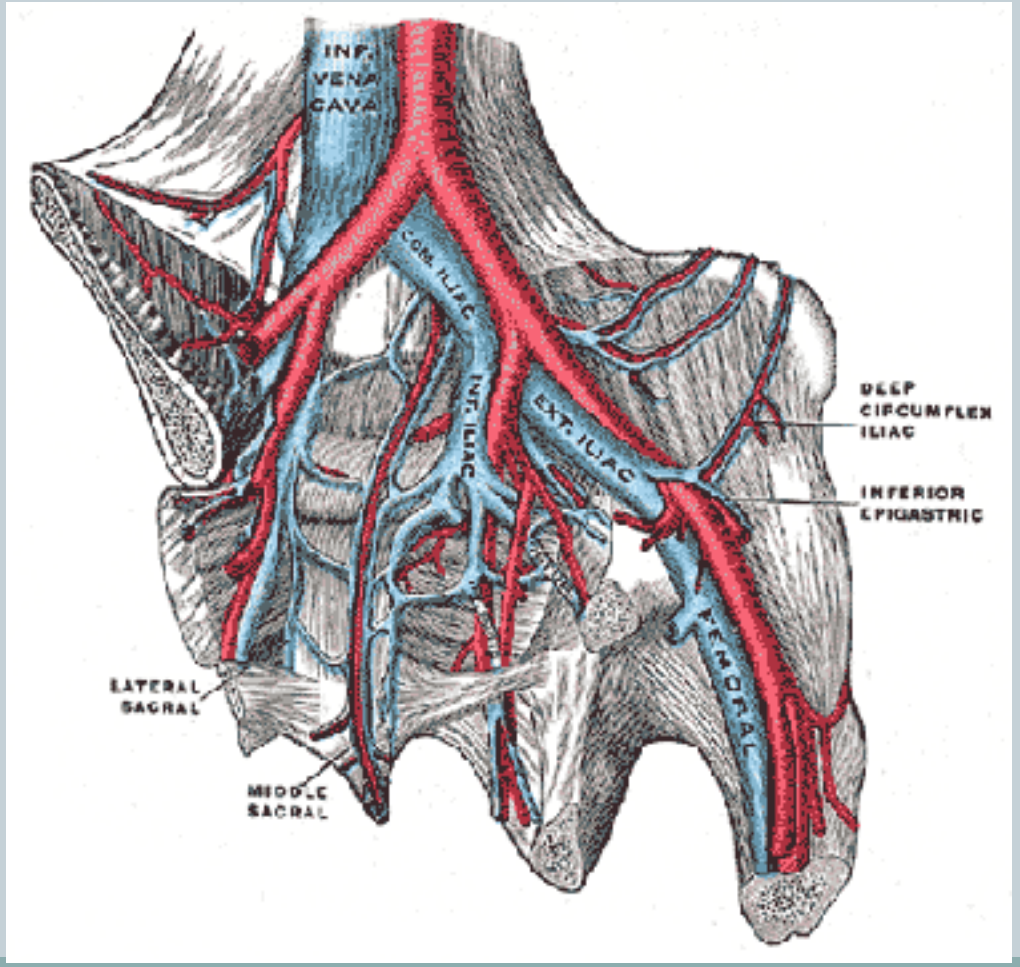
Fibromuscular Dysplasia: When to consider?

- HTN <35years
- Resistant hypertension
- Epigastric bruit & HTN
- Cervical bruit in patients <60
- Pulsatile tinnitus
- Severe & recurrent headaches
- TIA/Stroke <60
- Dissection of a peripheral artery
- Aneurysm in visceral or intracranial vessel
- Aortic Aneurysm in patient <60
- SAH
- Renal Infarction

Fibromuscular Dysplasia: Treatment

- Predominantly medical
- Renal → B.P. Control, PTA to relieve hypertension
- Cerebrovascular → Medically managed to reduce risk of stroke. Typically anti-platelets.
- Surveillance – no prognosis/regimes described
- Surgery – interposition grafts if required.
- Summary: Non atherosclerotic stenotic disease typically affecting middle aged women.

May-Thurner Syndrome



May-Thurner Syndrome

- MTS, Cockett Syndrome or Iliac-Vein Compression Syndrome – Compression of the common venous outflow of the leg leading to discomfort, swelling, pain & DVT's.
- Virchow's Triad – Stasis, Endothelial injury, hypercoagulability.
- Non-thrombotic Iliac Vein Lesions (NIVL) – Broader classification involving left and right iliac veins.

May-Thurner Syndrome: Clinical Features

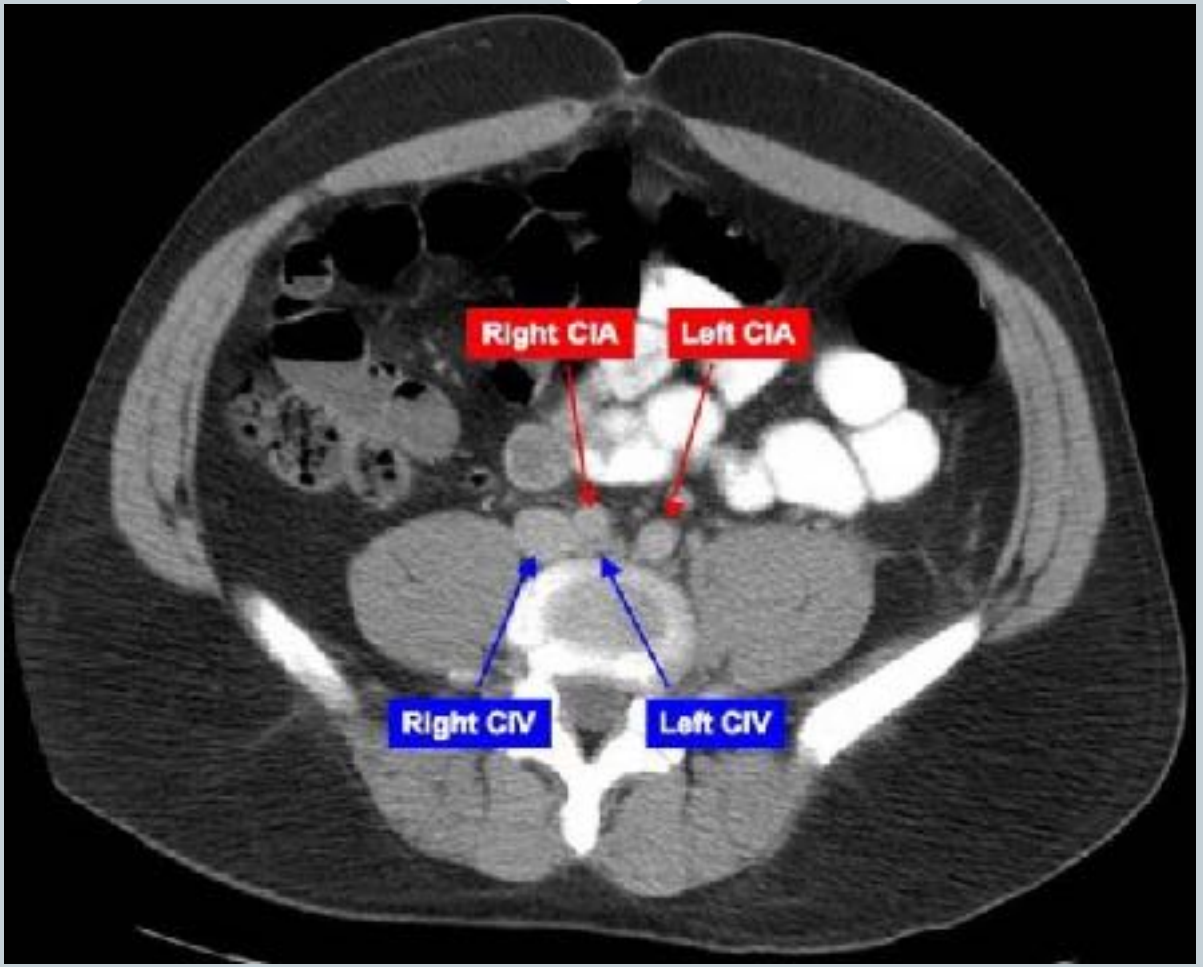
- Female to Male 3:1 – believed to be the sharper pelvic angle
- Younger – typically 20-40
- Permissive lesion – an intravenous web which facilitates thrombus at times of physiological stress e.g. surgery, pregnancy
- 2-5% of DVT's of lower limb venous disorders from MTS
- Must rule out other disorders such as antithrombin, Protien C&S deficiencies, Factor V Leiden & prothrombin G20210A

May-Thurner Syndrome: Diagnosis

- Typical referral – Young woman from peripheral hospital with left leg swelling & pain. US demonstrates proximal DVT.

- Imaging – Although most commonly CT/MRA are used, intravenous ultrasound is the gold standard for diagnosing venous webs/fibrosis.

May-Thurner Syndrome



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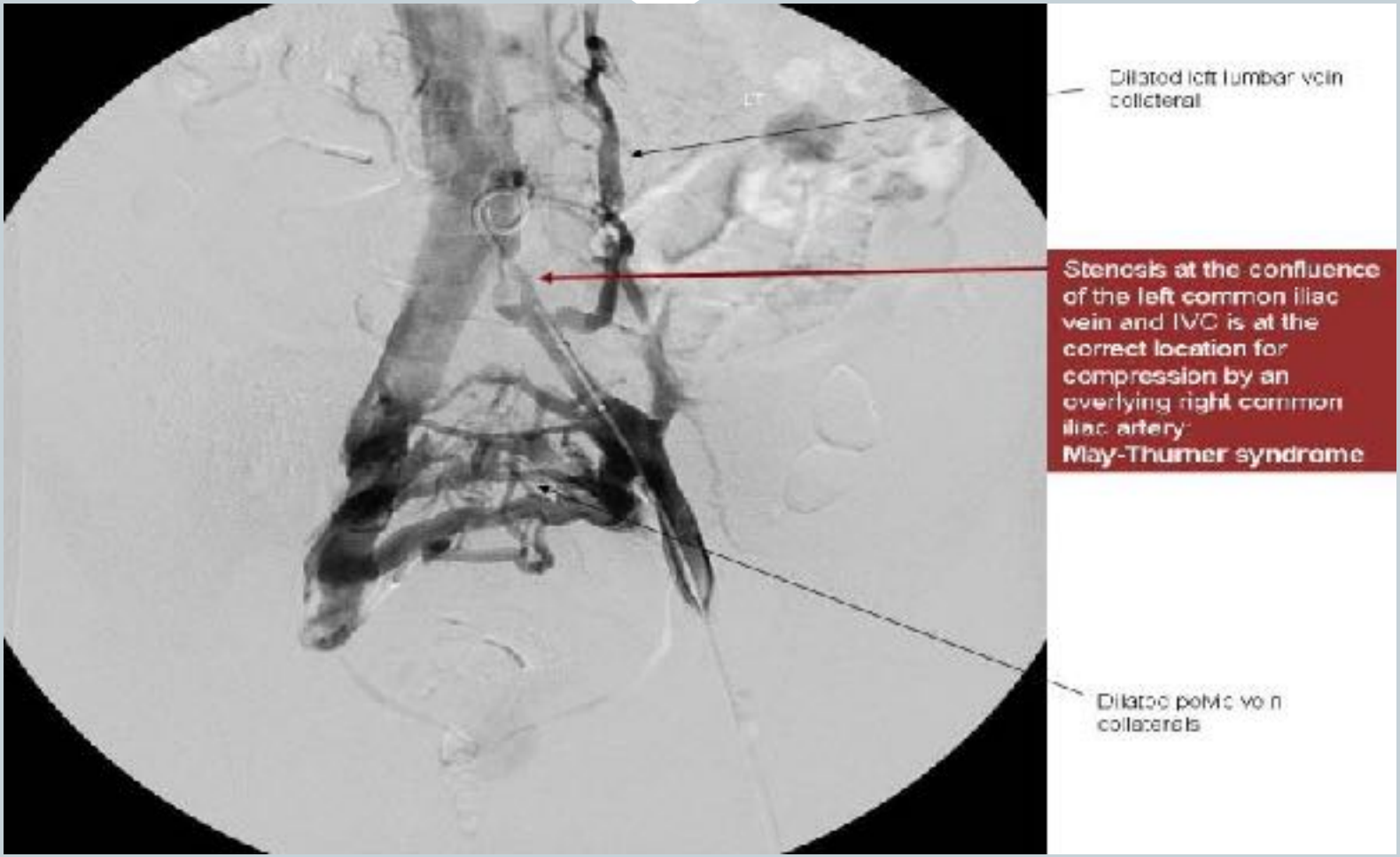


May-Thurner Syndrome

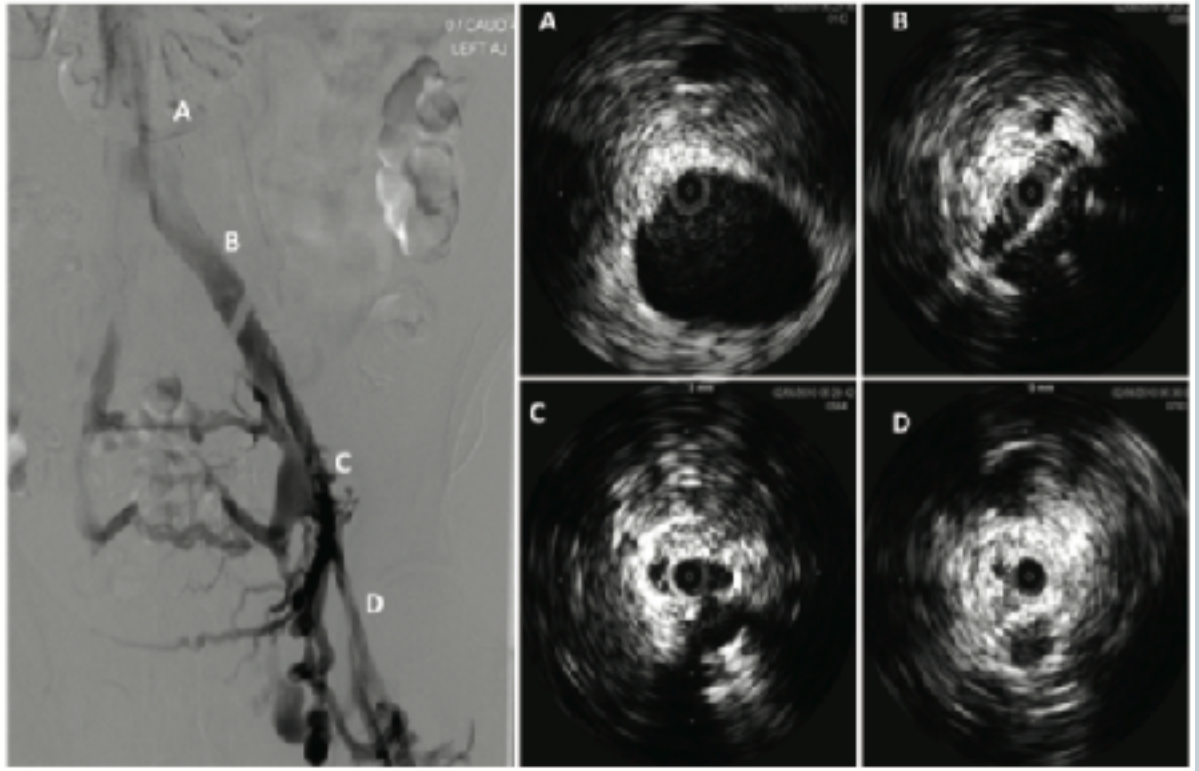


Figure 2. Left lower extremity venogram with view of the iliac veins and IVC (left) and corresponding IVUS images (right, A, B, C, D). Venographic findings of iliofemoral obstruction include "pancaking" of contrast (B), filling of multiple collateral veins, and venous stenosis (D). However, venography is much less sensitive than IVUS, which clearly shows multiple areas of severe stenosis (B, C, D).

May-Thurner Syndrome: Treatment

- Post thrombotic syndrome – Valve destruction from DVT leads to symptomatic venous reflux.
- NICE Clinical Guideline CG 144, November 2015 – Offer treatment for those patients with symptoms of <14 days, good functional status, life expectancy > 1 year and low risk of bleeding.
- Anti-coagulation as per local guidelines.
- Graduated compression stocking to manage symptoms.
- Investigation for cancer: physical examination, chest x-ray, blood tests, urinalysis & mammogram for women >40

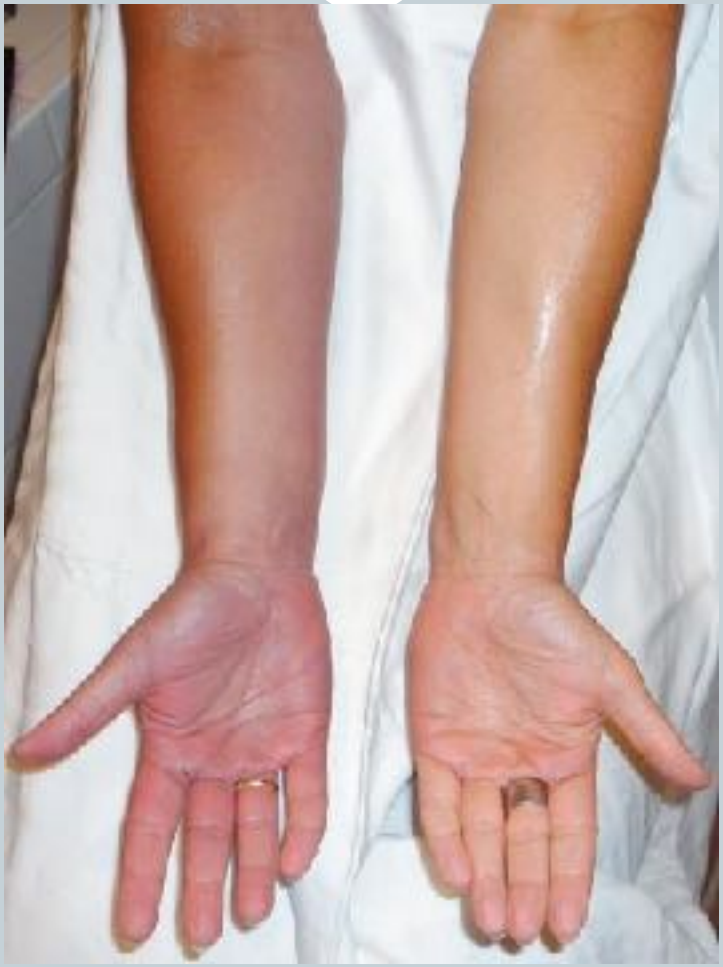
May-Thurner Syndrome

- Watson et al 2014, Cochrane Review
- Compared thrombolytic therapy & anti-coagulation vs anti-coagulation
- Complete clot lysys in Rx group RR 4.91 (p0.0004). Similar findings for venous patency.
- PTS Rates were significantly lower in the treatment group. RR 0.64. p0.00001
- Bleeds were more likely in treatment group. RR 2.23. 3 strokes occurred in patient all pre 1990.
- Stents may be required if the angioplastied vein rebounds immediately.

May-Thurner Syndrome: Conclusion

- Web/spur accommodates DVT
- Typically young women
- Venous thrombolysis significantly reduces PTS

Paget-Schroetter Syndrome



Paget-Schroetter Syndrome

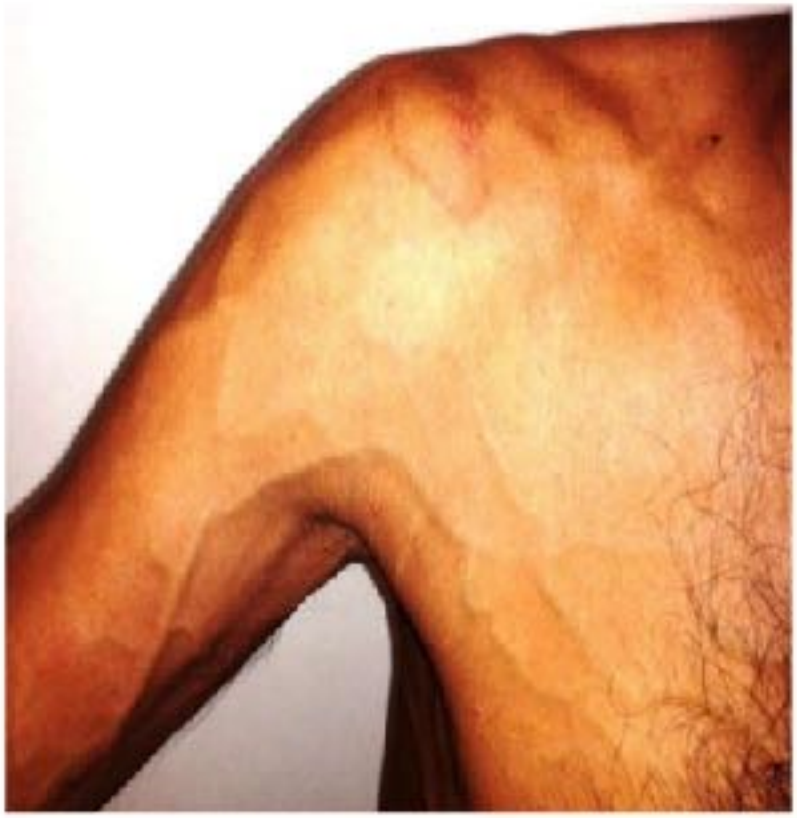
- Upper Limb Venous Occlusion,
- Young fit males aged 30, typically exercise induced
- Right limb 3x more common
- Symptoms of pain and swelling, discolouration, worsens with exercise

Paget-Schroetter: Diagnosis

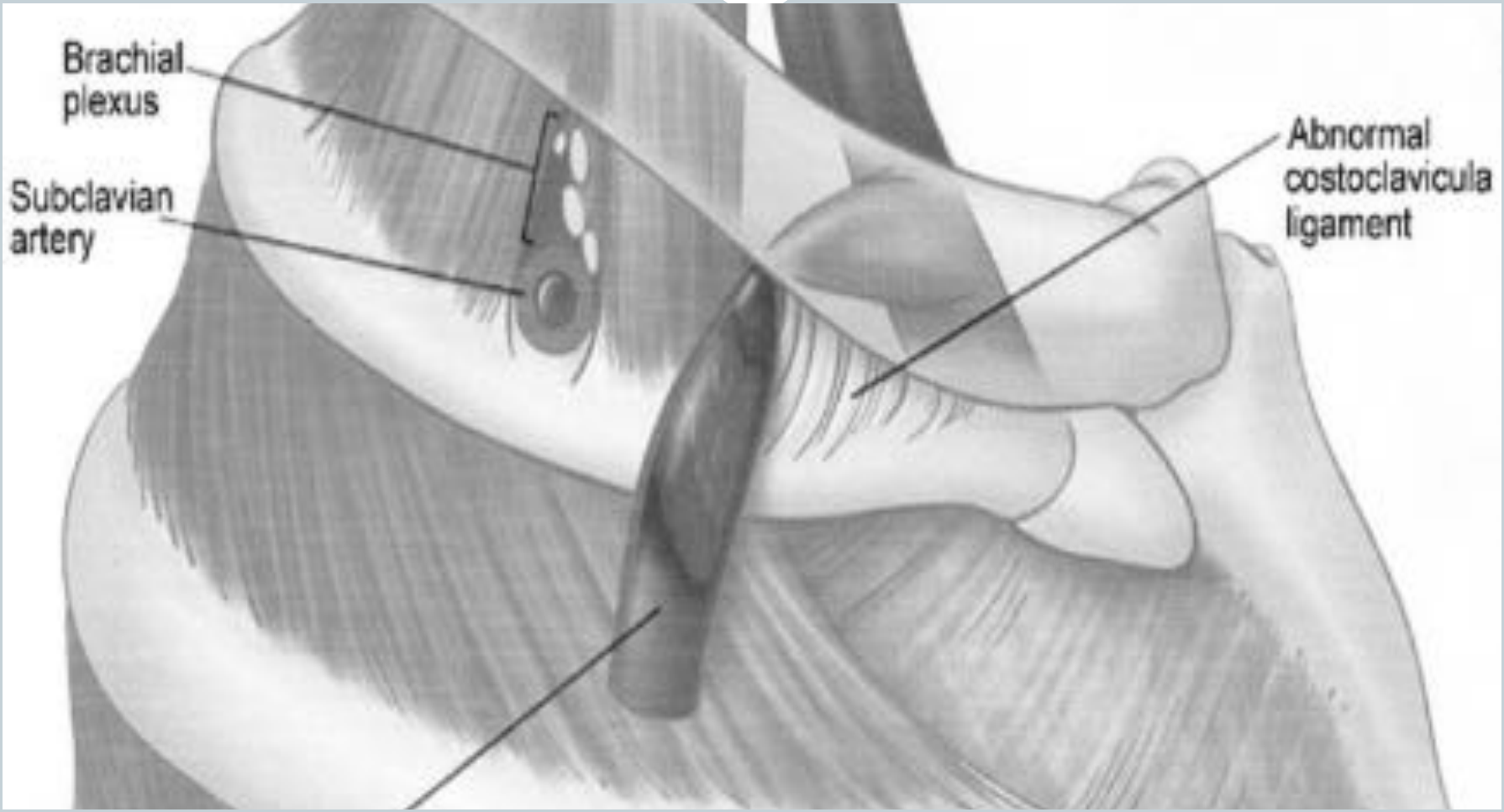
- Inspection – Swollen, cyanosed, shoulder girdle collaterals

- Investigation
- -Duplex. Sensitivity of 94% & Specificity of 96%
- -MRV – poor.
- -CTV role is undetermined.
- -Venography is preferred, via the basilic vein.

Paget-Schroetter

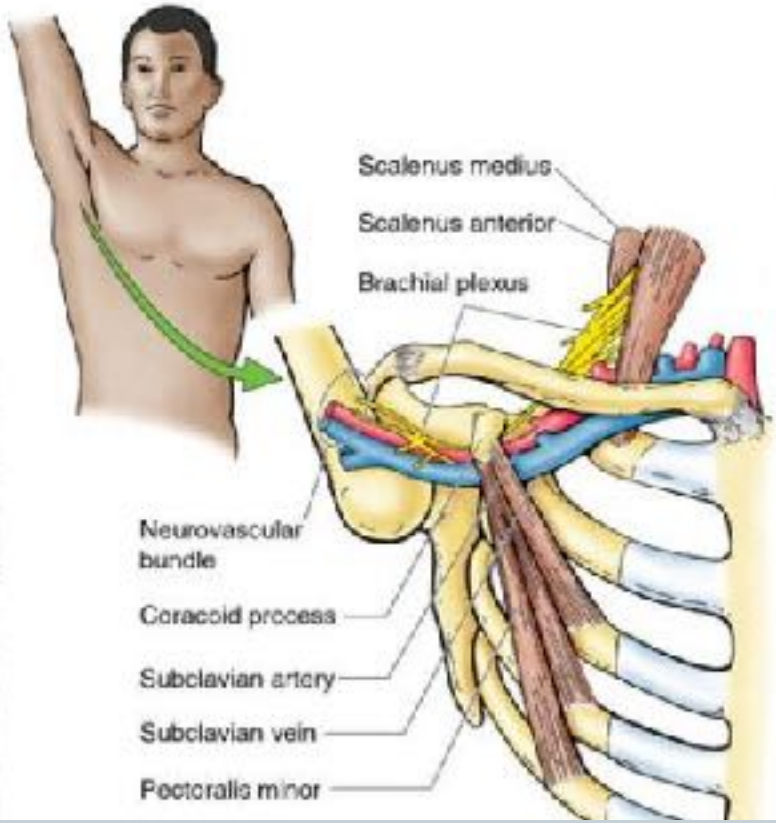


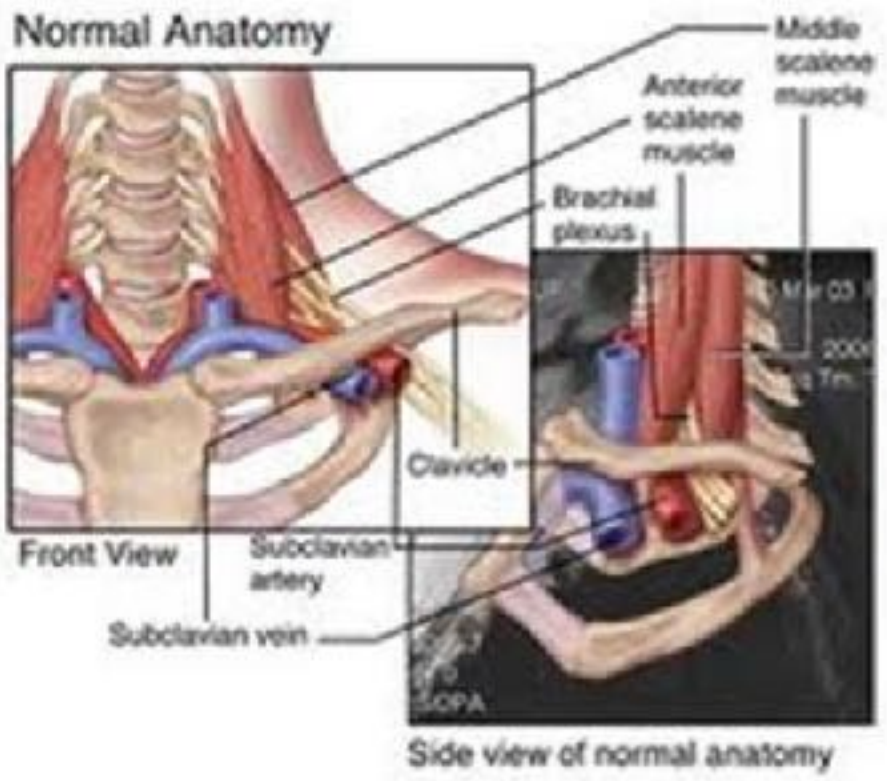
Paget-Schroetter



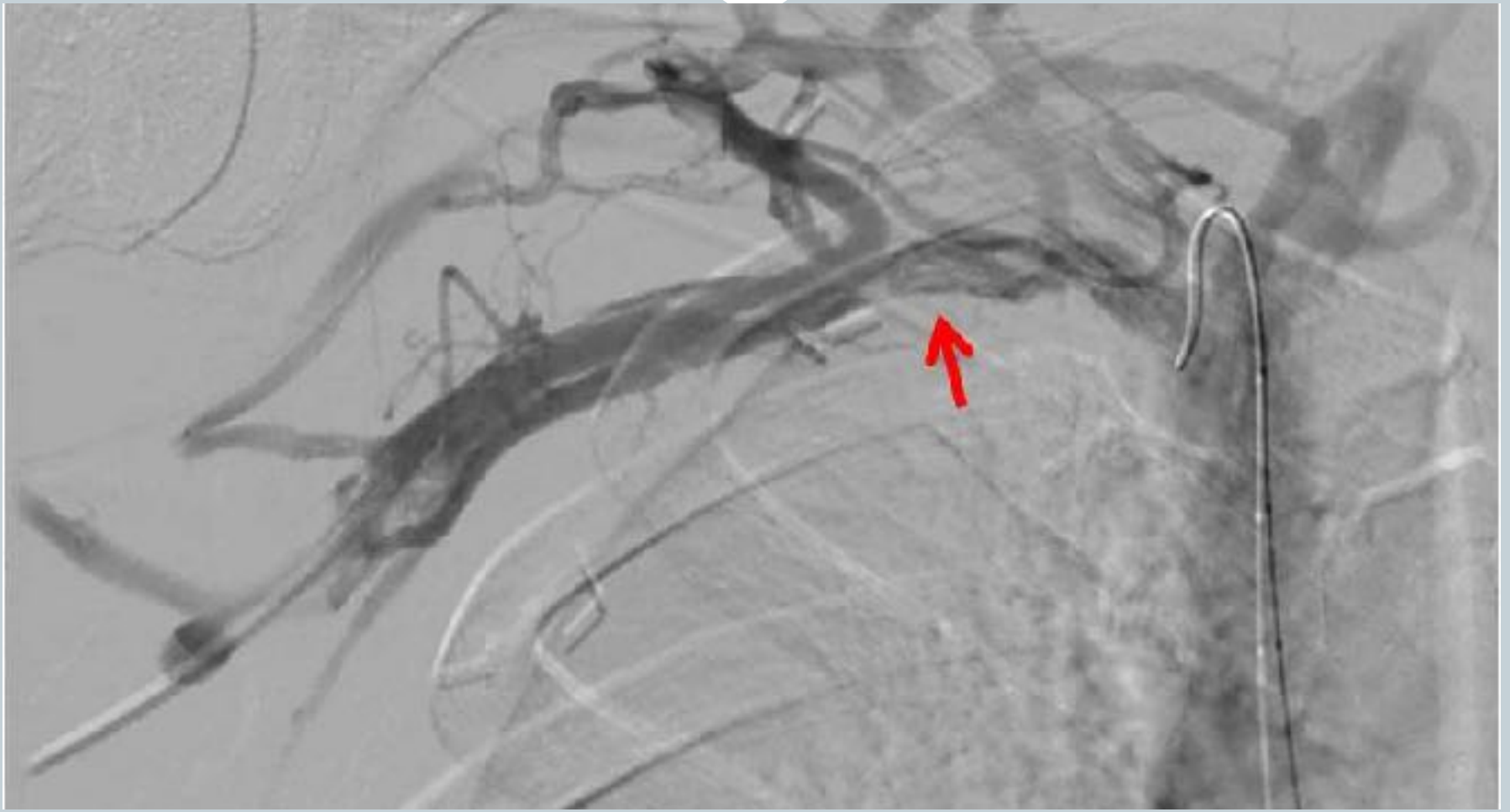
Paget-Schroetter

Thoracic Outlet





Paget-Schroetter



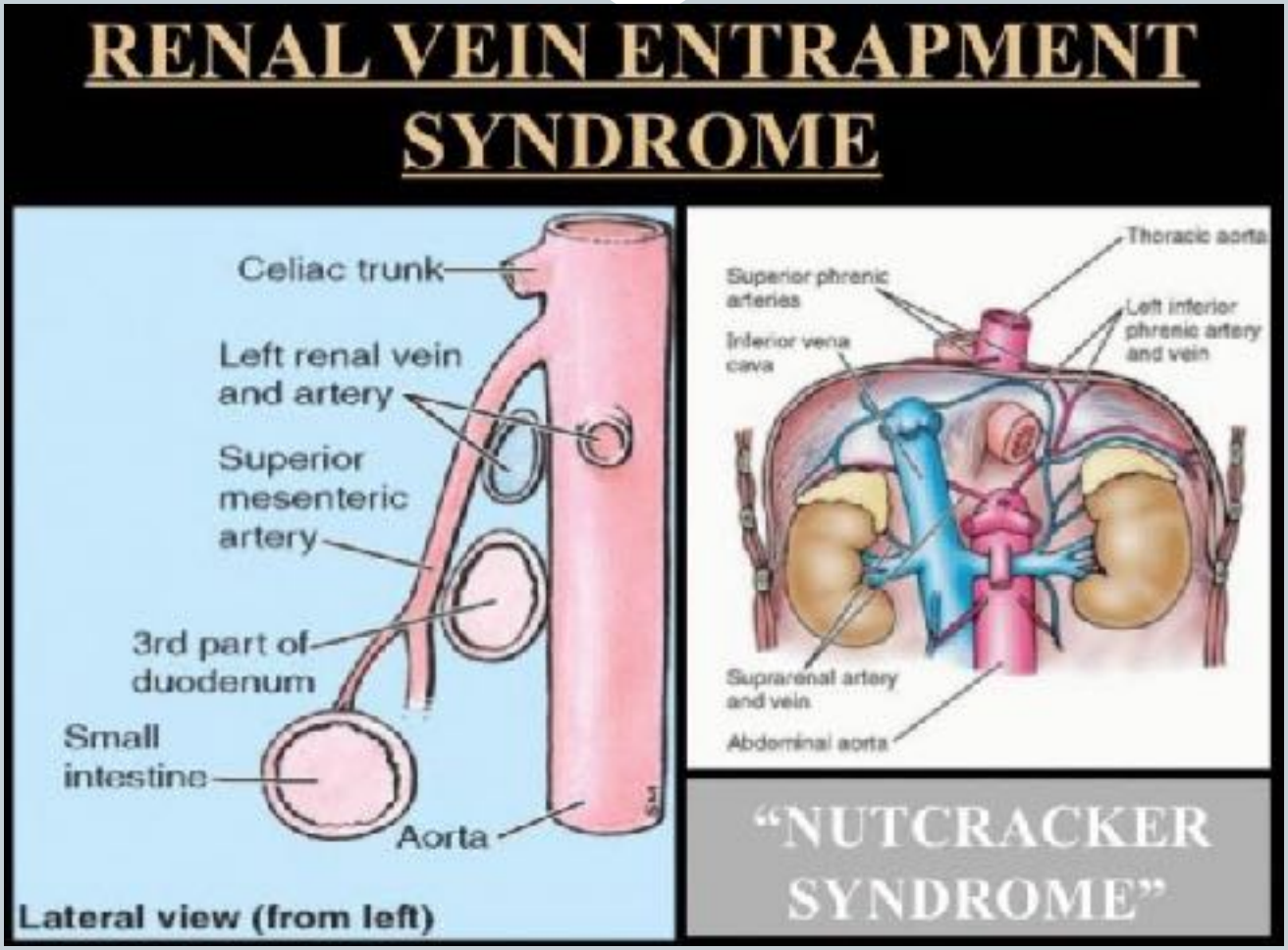
Paget-Schroetter: Treatment

- Conservative – rest, elevation & anti-coagulation is associated with a high morbidity.
- Thrombolysis/Thrombectomy both effective
- Anti-coagulation is temporary (3-6 months)
- First rib resection – if patients have evidence of venous compression at the time of lysis, rethrombosis is likely without rib resection

Paget-Schroetter: JVS 2010

- Comprehensive review 2010
- Good results
- 1. 50-60% Conservative & anti-coagulation
- 2. 80-90% with thrombolysis
- 3. 90-95% triple intervention
- Anti-coagulate for 3-6 months

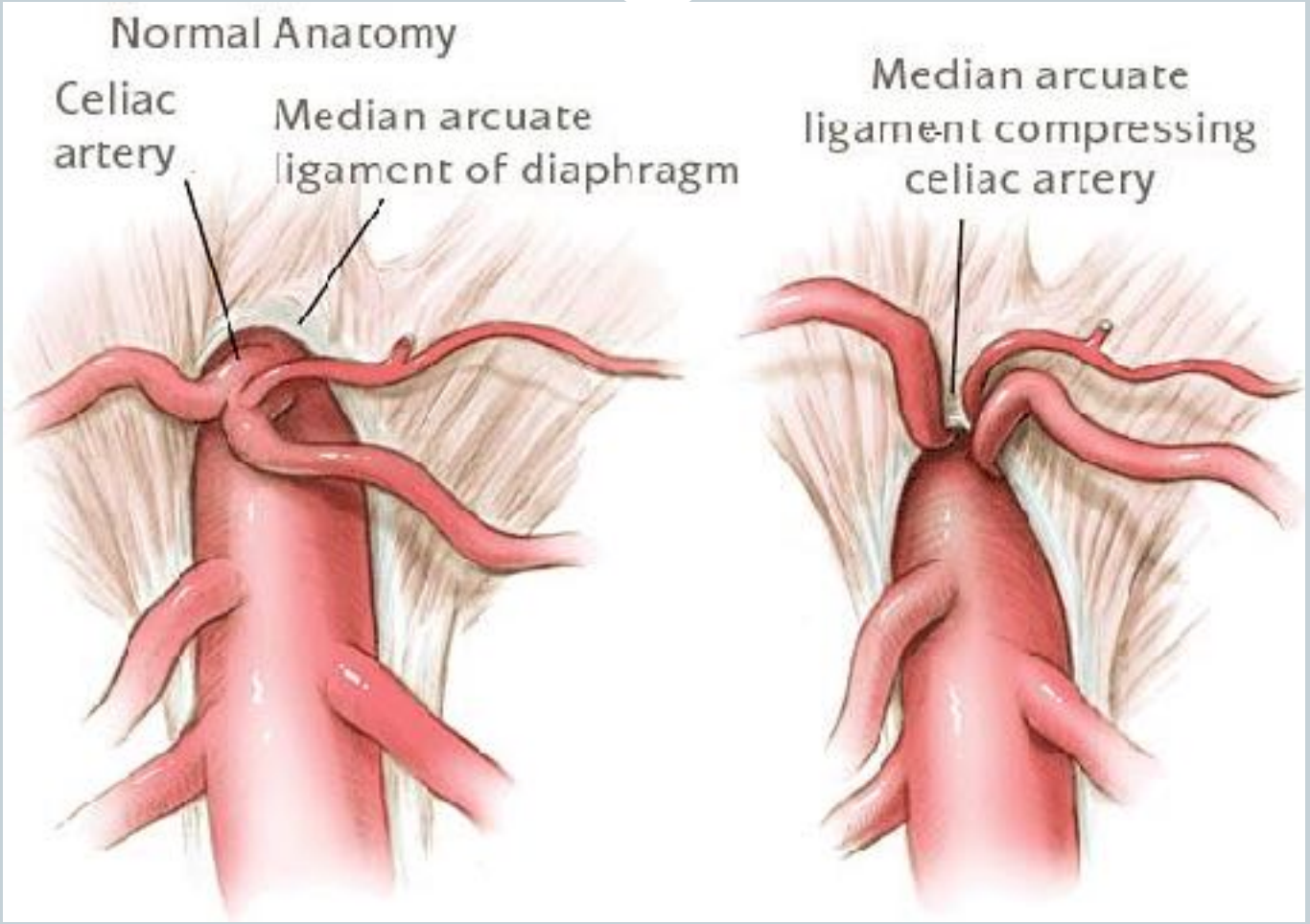
Nutcracker Syndrome



Nutcracker Syndrome

- Left renal vein entrapment betweek aorta and SMA
- S&S → Haematuria, left flank or pelvic pain
- Diagnosis → Left renal venography, CT, US
- DD → Pelvic congestion syndrome, renal stones, gentourinary malignancy
- Treatment → Stenting, renal vein reimplantaion, gonadal vein embolisation

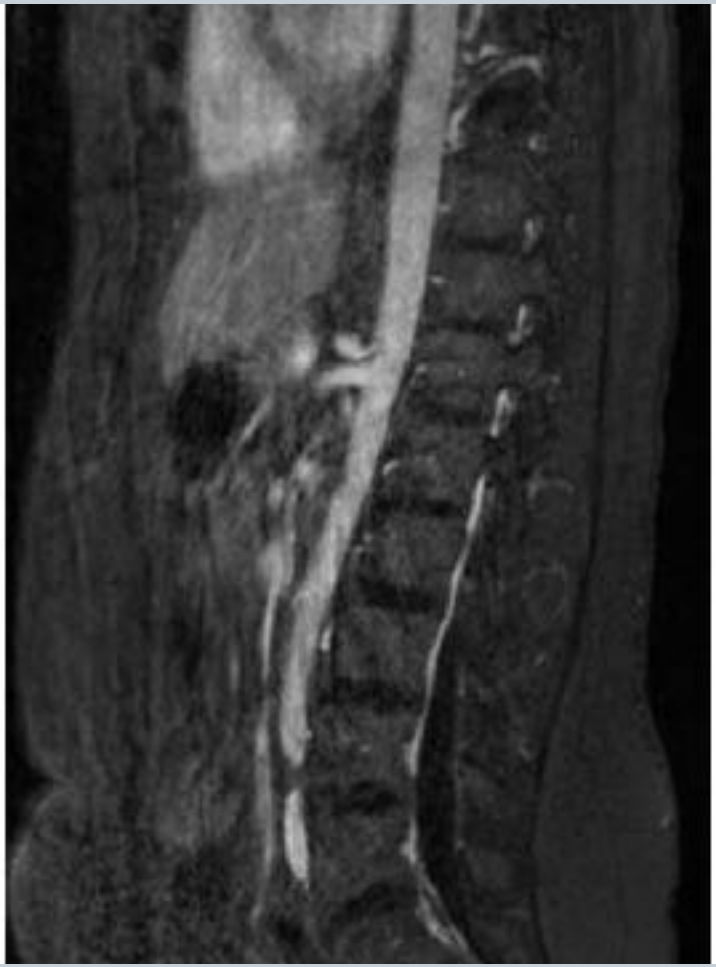
Median Arcuate Ligament Synrome

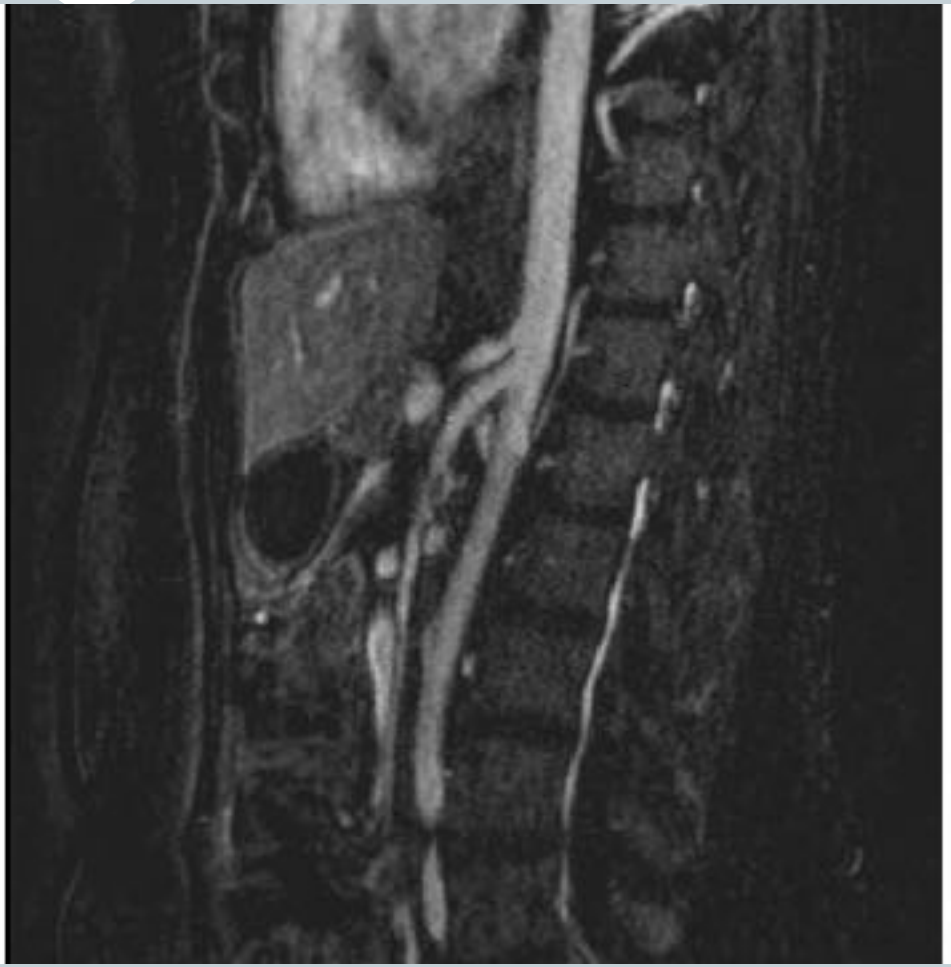


Median Arcuate Ligament Syndrome

- MALS or Coeliac Artery Compression Syndrome
- Anatomy MAL formed by joining the L&R hemidiaphragm near the twelfth thoracic vertebra. Forms the anterior aspect of the aortic hiatus through which the aorta, thoracic duct and azygous vein pass.
- In a quarter of individuals the ligament passes right in front of the coeliac compressing it. Pain may be due to ischaemia or ganglia compression
- Low indidence Affects 1% of those with low ligament, particulary women 20-40. Realistically it's a diagnosis of exclusion.
- Symptoms Epigastric pain +/- eating. Leads to anorexia and weight loss. Other signs are nausea, lassitude & exercise intolerance. Diarrhoea is common. Rarely a bruit may be heard.
- Aneurysmal dilatation of the superior and inferior pancreaticoduodenal arteries may occur.
- Diagnosis one of exclusion. Typical triad is one of post prandial pain, weight loss and an abdominal bruit
- US – PSV >2m/s usually diagnostic.
- CT – Narrowing with post stenotic dilatation. A hook-shaped artery is exaggerated on expiration.
- Treatment - surgical division and decompression of the ligament. Coeliac ganglia resection. PTA is reserved for those who have had failed surgical management

Median Arcuate Ligament Syndrome





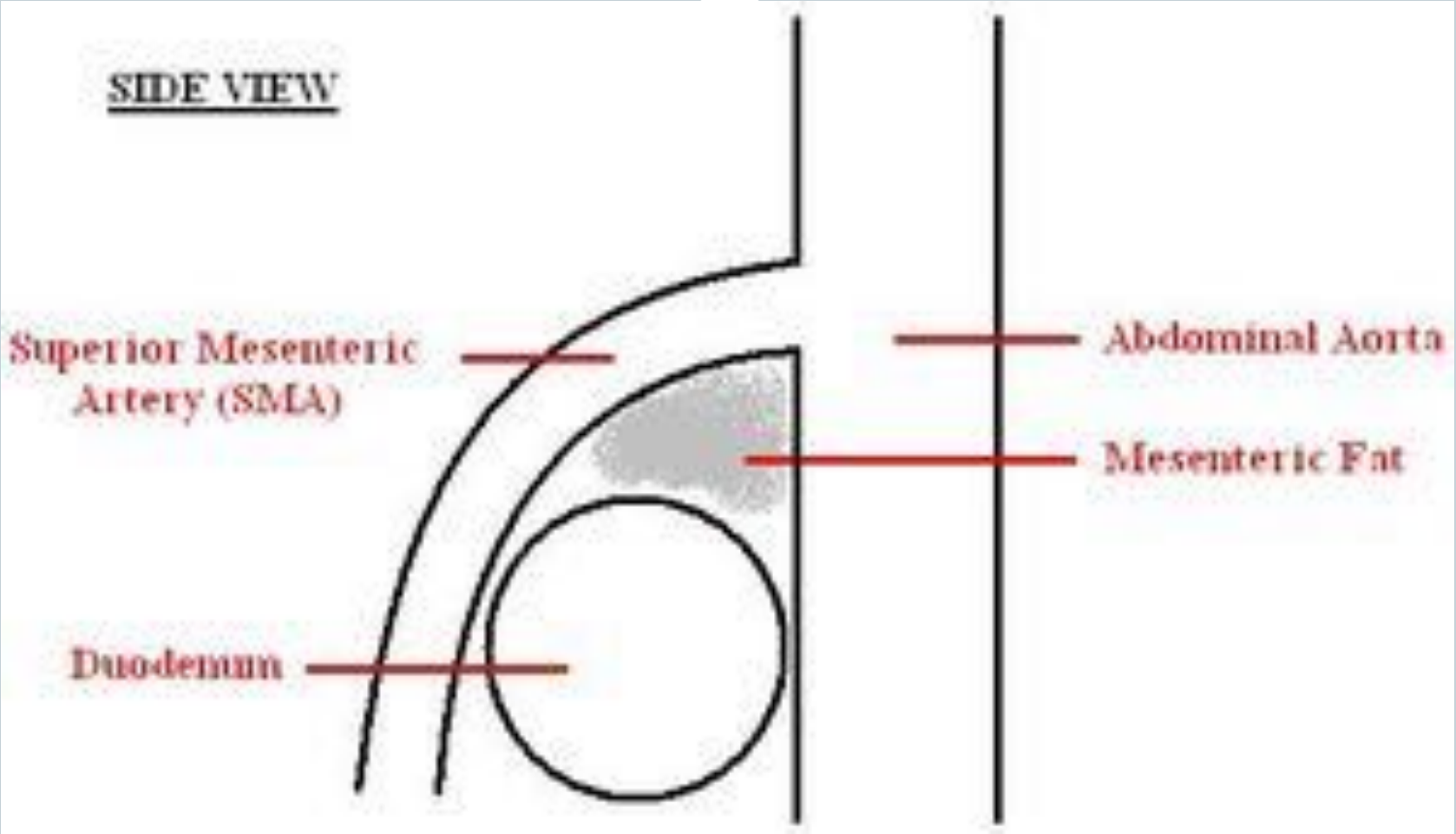
Median Arcuate Ligament Syndrome

- Duncan 2008 – Current Rx Options Cardiovascular Medicine

- Predictors of good surgical outcome:
- -Age 40-60
- -Lack of psychiatric illness/alcohol use
- -Post prandial pain
- -Weight loss >20lb

- 51 Patients underwent surgical intervention. 75% asymptomatic at follow up.

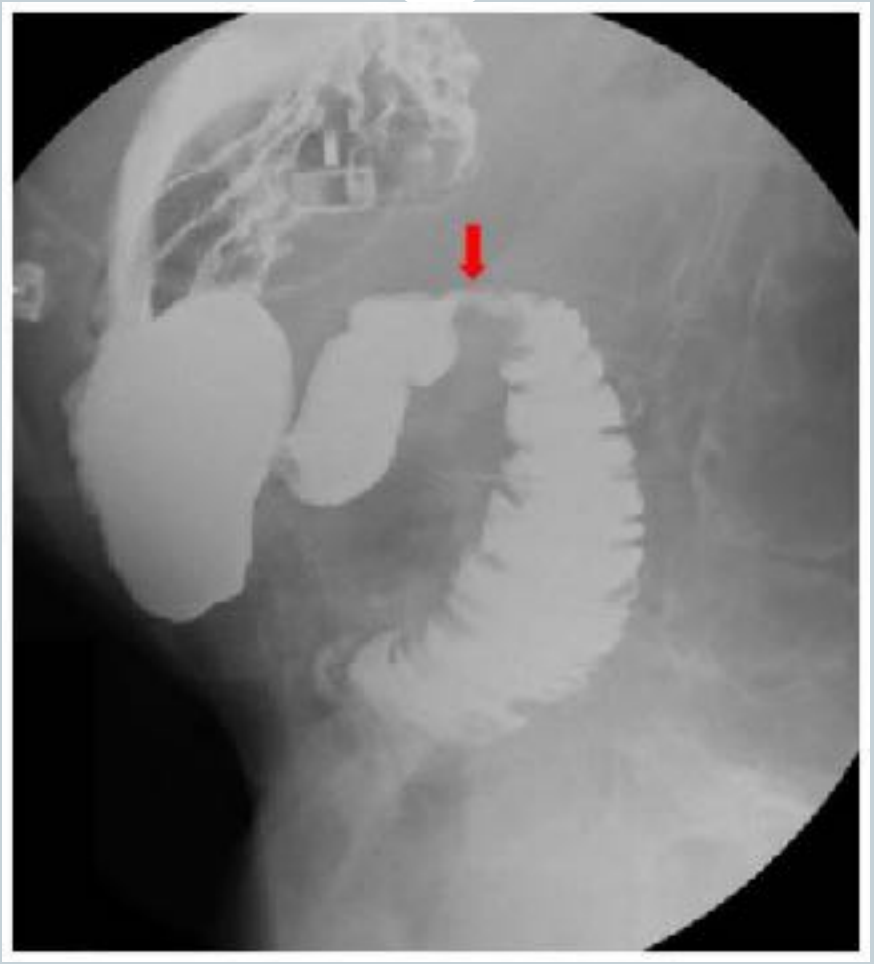
Superior Mesenteric Artery Syndrome



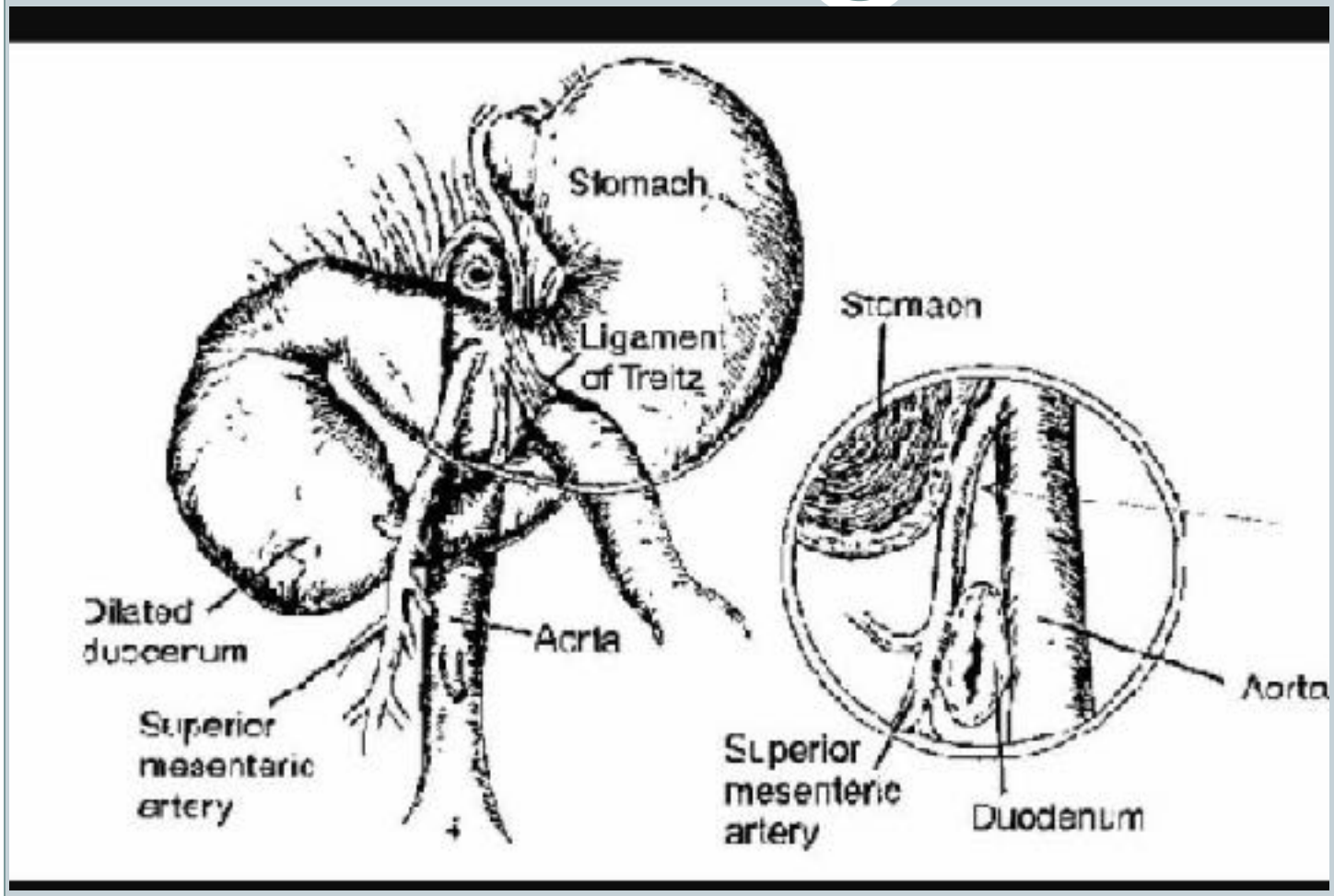
Superior Mesenteric Artery Symptoms

- SMAS or Wilke's syndrome. Compression of the third part of the duodenum between the aorta and the SMA.
- Radiographic diagnosis. Short angle of <30 degrees between aorta and sma, normally because of a lack of retroperitoneal fat.
- S&S → early satiety, N&V, stabbing abdominal pain, bloating and eructations(burping). In severe cases malnutrition can occur worsening the symptoms.
- Hayes manouevre – pressure below umbilicus cephalad and dorsal, takes pressure off SMA root relieving pain.
- Diagnosis: One of exclusion. CT, UGI series, US demonstrating increased velocity. This condition is controversial. Affects females 10-30years old.
- Treatment: Medically in 70% with weight gain being the primary concern. Surgically duodenojejunostomy or Roux-En-Y. Strong's operation is to divide the ligament of Trietz. Some patients do not improve after surgery presumably because of reverse peristalsis.

SMA Syndrome

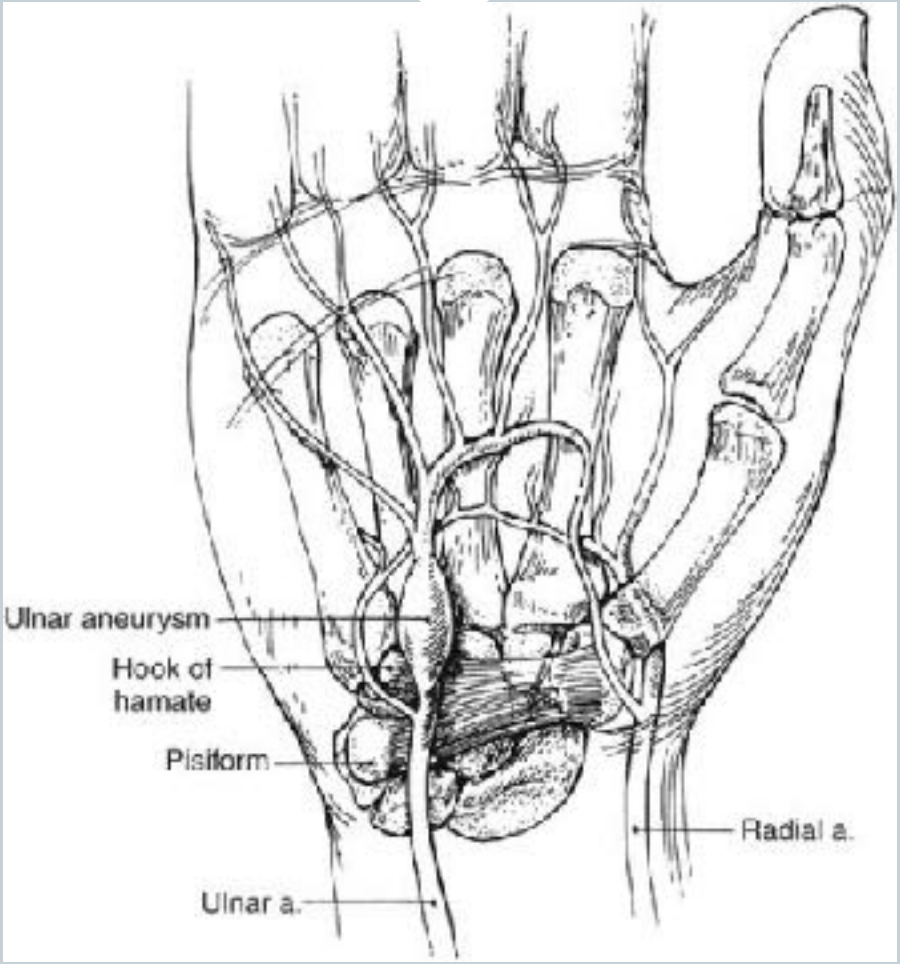


SMA Syndrome



The suspensory muscle of the duodenum, or the ligament of treitz is a thin muscle connecting the juntcion of the duodenum, jejunum and DJ flexure to connective tissue surrounding the SMA.

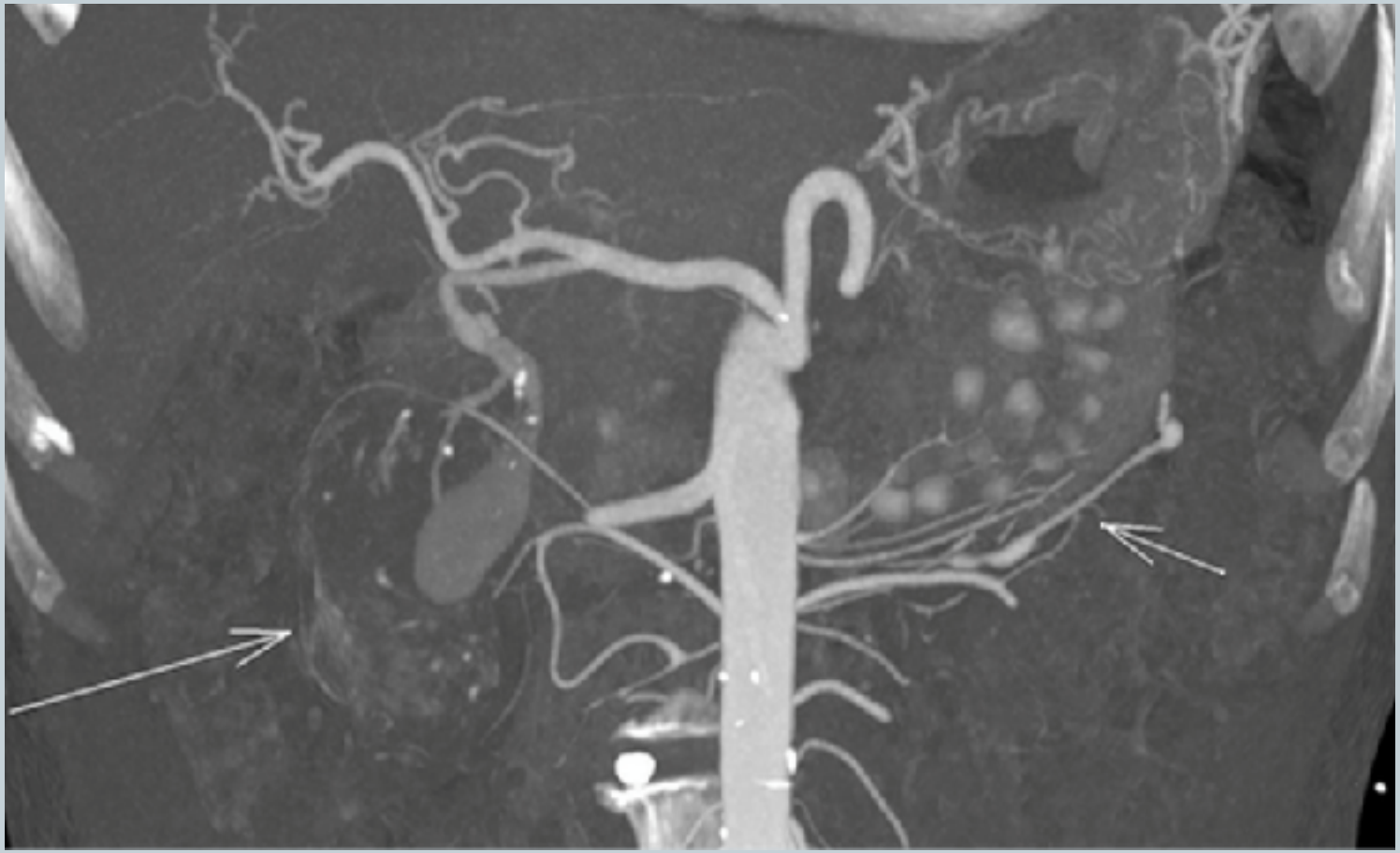
Hypothenar Hammer Syndrome



Hypothenar Hammer Syndrome

- Repetitive trauma to hypothenar eminence
- Occlusion, thrombosis, aneurysm or embolisation of the ulnar artery as it passes over the hamate.
- M:F 9:1 typically in 40's or 50's
- Examination: Blanching and mottling +/- cyanosis/gangrene. Unusual tenderness over hypothenar eminence, callous formation.
- Fingertip ulcerations and splinter haemorrhages.
- Allen's test may be positive.
- Treatment is often supportive although thrombolysis and vasodilatory agents may be used in acute ischaemia.
- Aneurysms should be removed to treat the source of emboli.

Segmental Arterial Mediolysis



Segmental arterial mediolysis with gastroduodenal artery aneurysm.

Segmental Arterial Mediolysis

- Disease of the elderly - Sudden spontaneous haemorrhage within the splanchnic arterial branches.They can also develop fusiform aneurysms
- Non-inflammatory, non-atherosclerotic arteriopathy
- Histologically there is lysis of the smooth muscle of the outer media resulting in intramural haemorrhage, saccular or dissecting aneurysms, thrombosis and haemorrhage.
- Typically affects the SMA in a skip pattern. Some histopathological similarities to FMD but the clinical features are usually diagnostic.
- Patients typically present shocked and in pain

Segmental Arterial Mediolysis

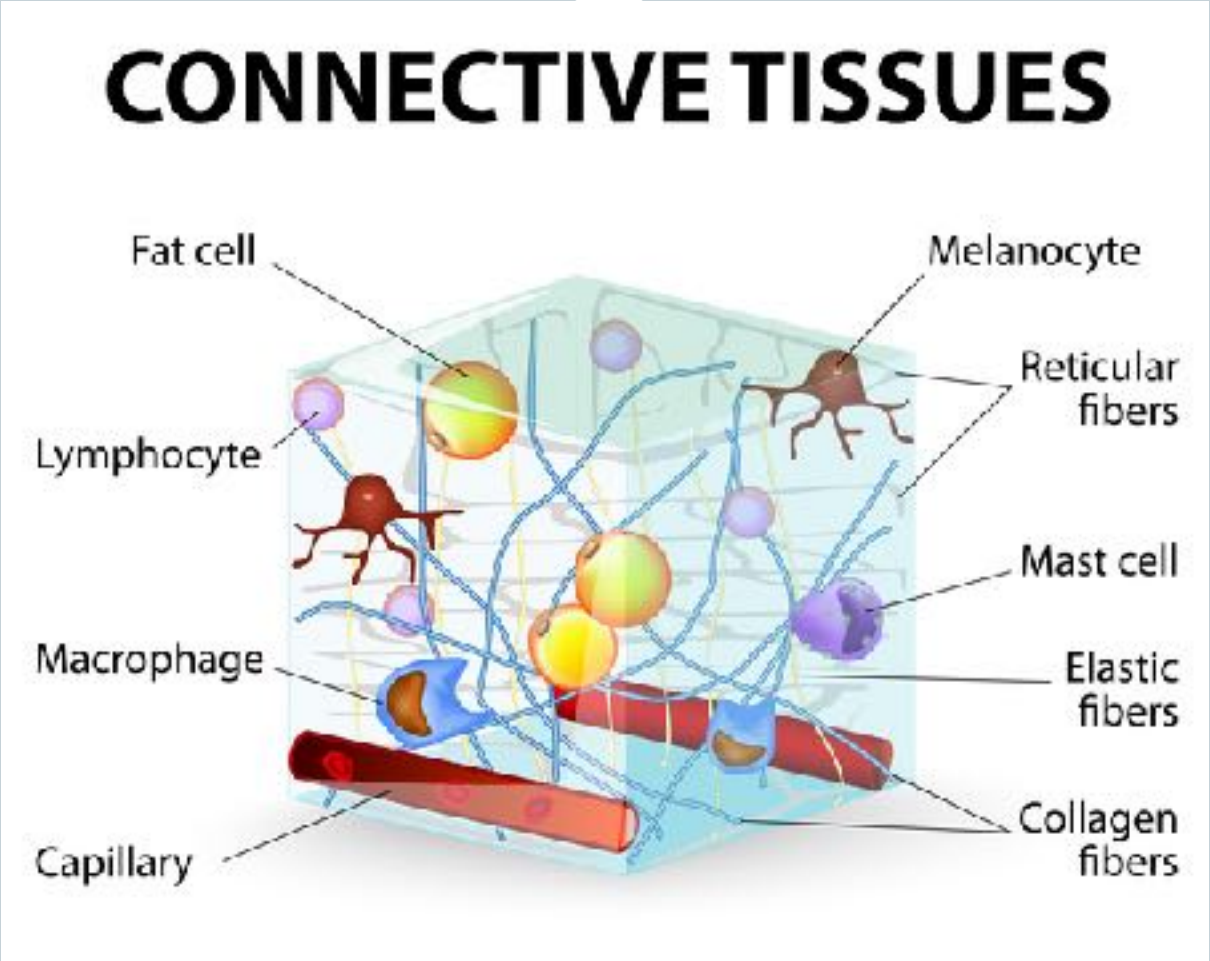
- CT:
- -Mesenteric or intraperitoneal haemorrhage
- -Fusiform aneurysms
- -Stenoses
- -Occlusions
- -Dissections

- Typical string of beads appearance

Segmental Arterial Mediolysis: Treatment

- Depends on severity at presentation
- Immunnosuppression is couterproductive
- Rx aneurysms >10mm
- Interventional / Open surgical correction

Connective Tissue Disorders



Ehlers-Danlos Syndrome

- EDS is a group of connective tissue disorders, with a defect in type 3 collagen, most of which are inherited and autosomal dominant. Symptoms may include loose joints, stretchy skin and abnormal scar formation. These can be noticed at birth or in early childhood. Complications may include aortic dissection, joint dislocations, scoliosis, chronic pain or early OA.

- Type III Collagen is a fibrillar collagen consisting of only one alpha chain, with three alpha chains wrapped round each other. It is the main component of the interstitial matrix

- Genetic Defects: More than a dozen genetic defects identified such, subgrouped into fibrous proteins and enzymes

- Incidence is 1:150,000

- Types of ED. There are 13 EDS with a significant overlap in features. Examples of this include Vascular, Classical, Hypermobile, Kyphoscoliosis, Cardiac-Valvular etc.

Ehlers-Danlos Syndrome

- Symptoms clearly depend on the type of EDS the patient has. We are interested in incresed elasticity of the skin, joint hyper-mobility, thin skin and easy bruiability. We can see varicose veins and aneurysmal dilatation, particularly thoracic and abdominal aorta. Spontaneous arterial rupture is possible along with gravid uterine +/- intestinal rupture.

- Diagnosis Evaluation of medical history and clinical observation. Beighton criteria is used to asses joint mobility. DNA testing and collagen typing via skin biopsy.

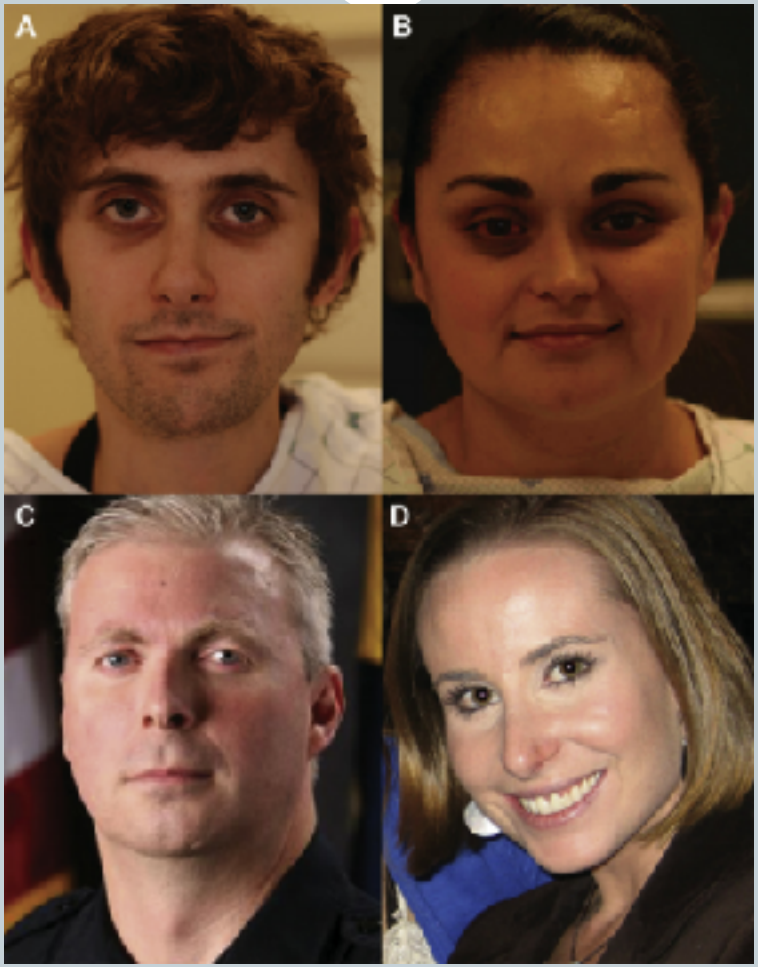
- Management- N o cure so management is supportive with surveillance.

- Surgical Intervention - It is best not to intervene in EDS patients where possible. Of note they have considerably longer recovery time and require particularly patient tissue handling. Resistance to xylocaine and bupivacaine are not uncommon.

- Life expectancy is dependent on the type of EDS, those with vessel problems are at risk of sudden deat. The average life expectancy of a patient with vascular EDS is 48 years.

Ehlers-Danlos Syndrome

Ehlers-Danlos Facial Features



Marfan's Syndrome

Once again, Marfan's is an autosomal dominant disorder, this time of Fibrillin-1. People are tall and thin with long fingers toes and limbs.

Marfan's Syndrome

- Vascular problems – Prone to mitral valve prolapse & aortic aneurysms.

- Diagnosis – Ghent criteria
- Management – NO cure but life expectancy has improved to normalise now.
- Physical activity should be limited and has classified certain physical activities as low, medium or high risk. Bodybuilding is high risk whilst running is medium
- Patients are often prescribed beta blockers such as propranolol or ACEi.
- Surgery is as per the size of aneurysms. Preferably elective.
- During pregnancy women are at very high risk of dissection. They should be screened every few weeks with ECHO.

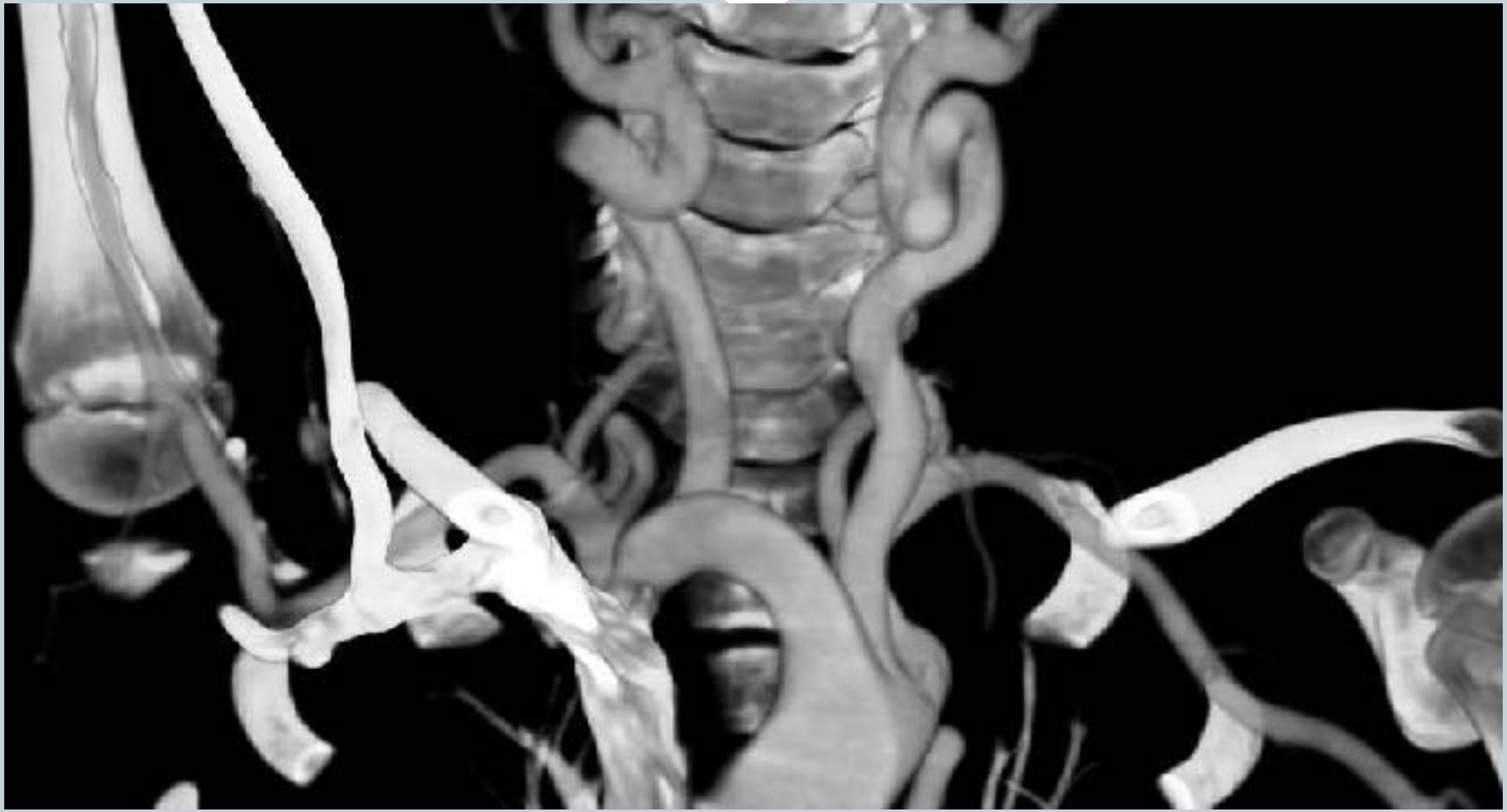
Marfan's Syndrome – Ghent Criteris

CMJ

Table 1. Ghent diagnostic nosology

System	Major criterion	Involvement
Cardiovascular	Dilatation of the aortic root	Mitral valve prolapse
	Dissection of the ascending aorta	Dilatation of the pulmonary artery, below age 40
		Calcified mitral annulus, below age 40
		Other dilatation or dissection of the aorta
Skeletal	At least 4 of the following features:	2 of the major features, or 1 major feature and 2 of the following
	Pectus carinatum	Pectus excavatum
	Pectus excavatum requiring surgery	Joint hypermobility
	Upper: lower segment ratio <0.86 or span: height >1.05	High palate with dental
	Wrist and thumb signs	Crowding
	Scoliosis >20° or spondylolisthesis	Characteristic face
	Reduced elbow extension (<170°)	
	Pes planus	
	Protrusio acetabulae	
Ocular	Lens dislocation (ectopia lentis)	Flat cornea
		Increased axial length of globe (causing myopia)
		Hypoplastic iris or ciliary muscle (causing decreased miosis)
Pulmonary	None	Spontaneous pneumothorax
		Apical blebs
Skin/integument	None	Striae atrophicae
		Recurrent or incisional hernia
Dura	Lumbosacral dural ectasia	None
Genetic findings	Parent, child or sibling meets these criteria independently	None
	Fibrillin 1 mutation known to cause Marfan syndrome	
	Inheritance of DNA marker haplotype linked to Marfan syndrome in the family	

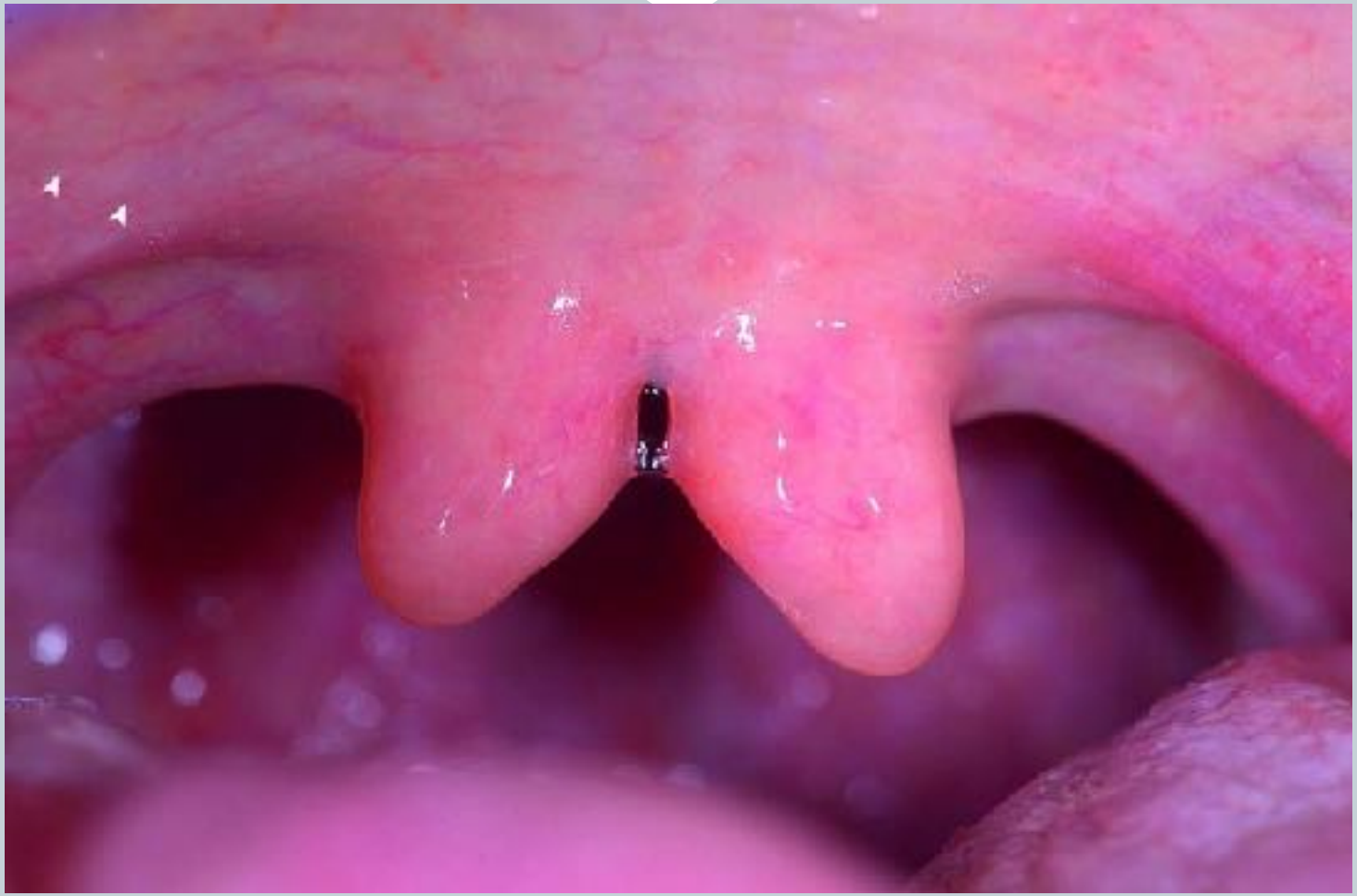
Loeys Dietz Syndrome



Loeys Dietz

- Genetic – Autosomal dominant
- Five subtypes TGFBR1 & 2, SMAD3, TGFB2&3 –Thse genes code for non functional proteins failing to produce collagen
- Symptoms – Weak skin and joints, stenrnal & cardiac abnormalities
- Aneurysms +/- Dissections in any vessel
- Treatment: Losartan may block TGF Alpha, slowing or halting the progress of aneurysms. B-Blockers may also reduce shearing stress.
- People should be surveilled and aneurysms treated.

Loeys Dietz – Bifid Uvula



Ergotism – The Psychedelic Nightmare

Ergotism

- Ergotism is the effect of long-term ergot poisoning, typically by the ingestion of alkaloids produced by the Claviceps Purpurea fungus.

- It infects rye and other cereals. Alkaloids are mainly nitrogen atoms.
- It is also caused by the use of ergoline based drugs which vasoconstrict (5-HT receptor antagonists.) used in the treatment of parkinson's and migraines.

- Symptoms are convulsive and gangrenous. Dry gangrene is due to vasoconstriction. Affects distal structures with desquamation. Weak pulses, neuropathy, oedema and tissue ischaemia

- Treatment includes vasodilators such as nitroprusside or nitroglycerine.