FMD 101

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FMDSA Annual Meeting

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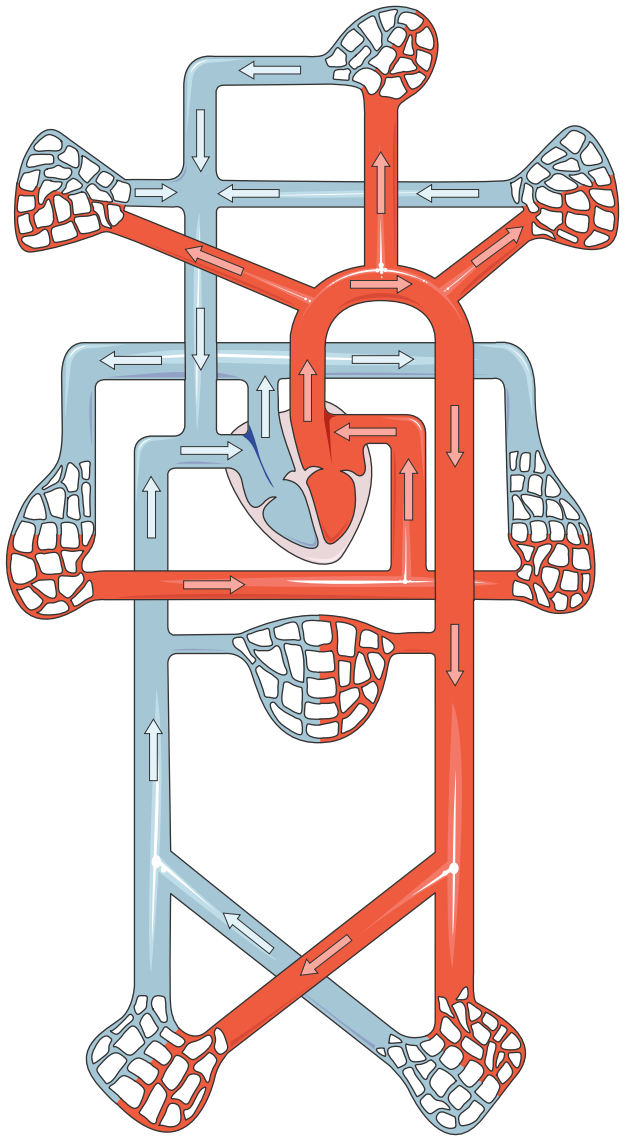
Back to basics…

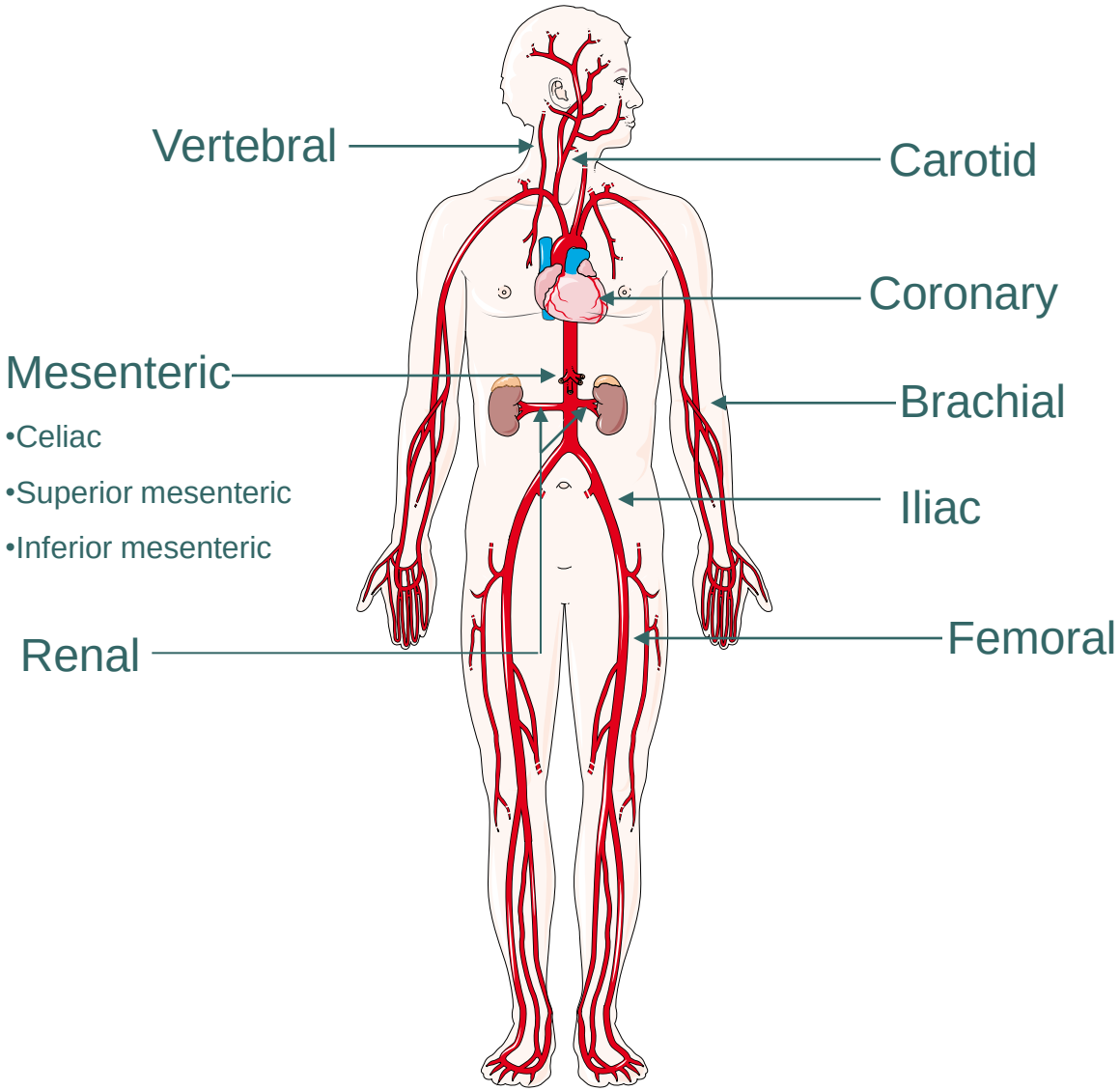
- Blood
  - Bodily fluid that transports necessary substances (oxygen, nutrients, antibodies, hormones, etc) and waste to and from cells in the body
- Blood vessels
  - Circulates blood to organs

The Circulatory System

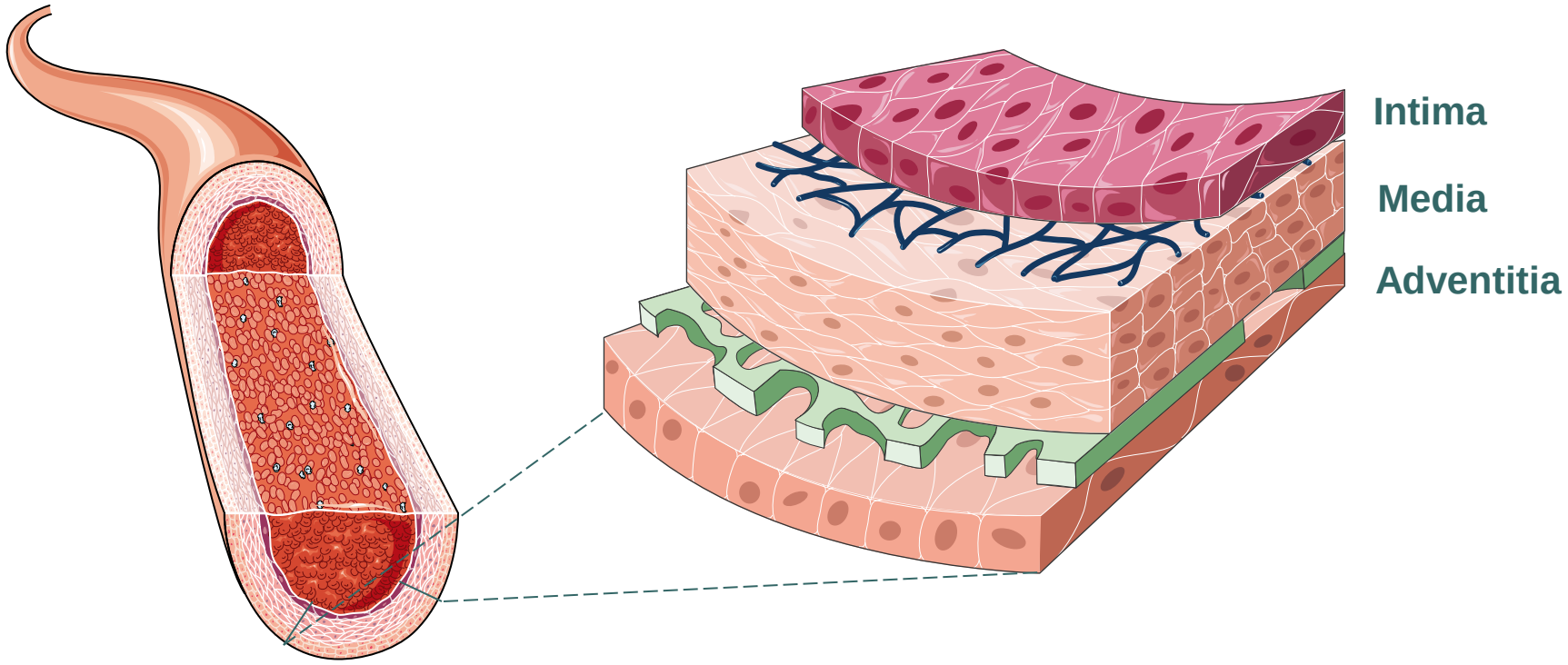
- Heart
- Arteries
  - Away from the heart
- Veins
  - Towards the heart
- Lymphatics
  - Part of the immune system



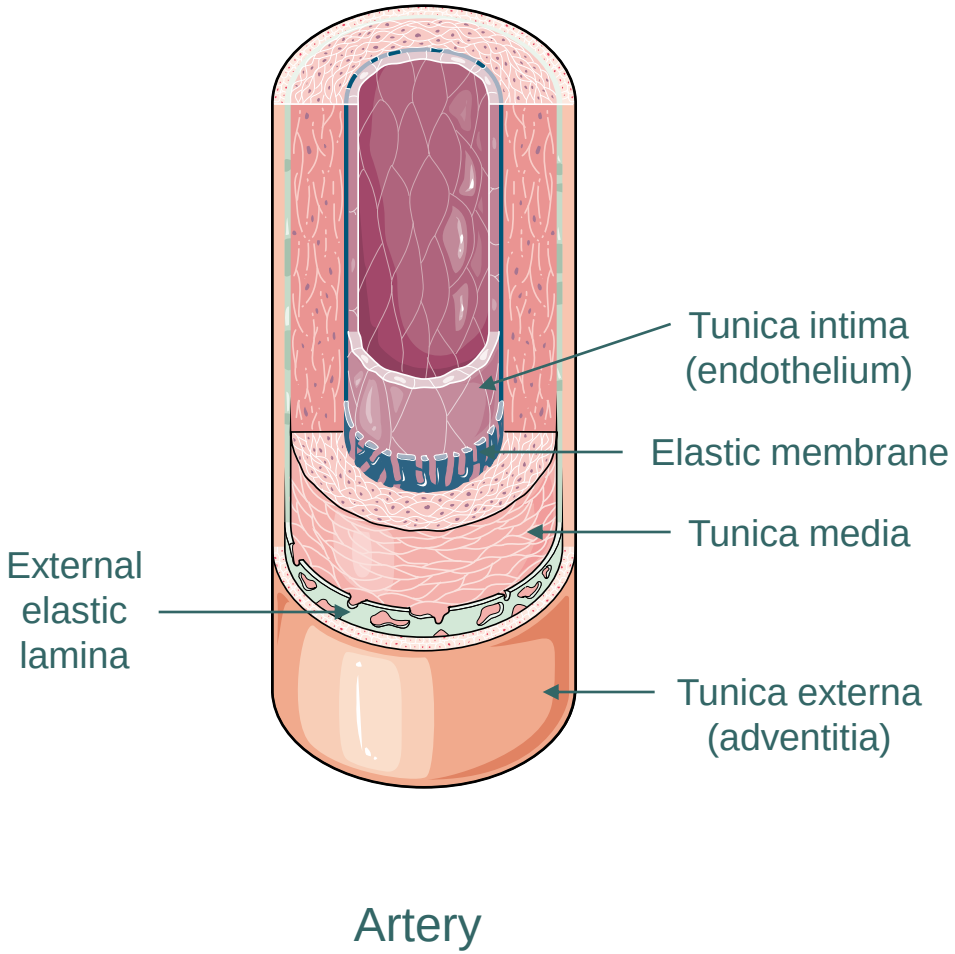
Arteries


Vertebral
Carotid
Coronary
Mesenteric
•Celiac
•Superior mesenteric
•Inferior mesenteric
Brachial
Iliac
Renal
Femoral

Arterial layers



Arterial layers

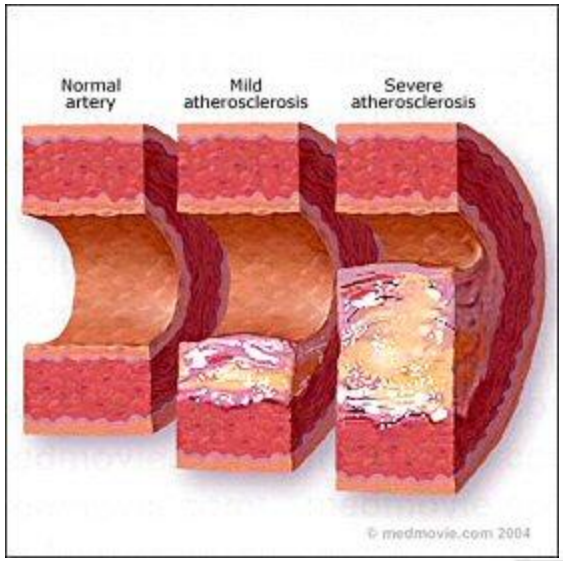


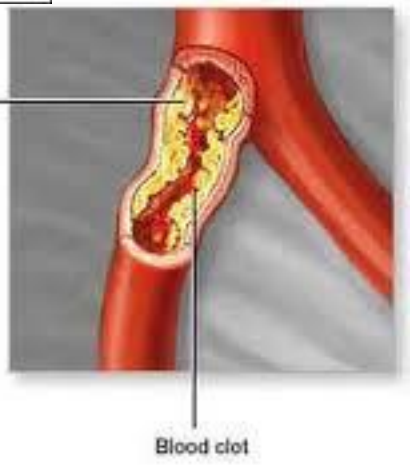
Artery

- Intima
  - One layer of endothelial cells
  - Many functions:
    - Selective permeability barrier
    - Inflammation
    - Blood clotting
    - Vascular growth and remodeling
    - Control of vascular tone
- Media
  - Smooth muscles and elastic tissue
  - Vascular tone
- Adventitia
  - Connective tissue
  - Anchors and stabilizes vessel

Arterial Pathologies

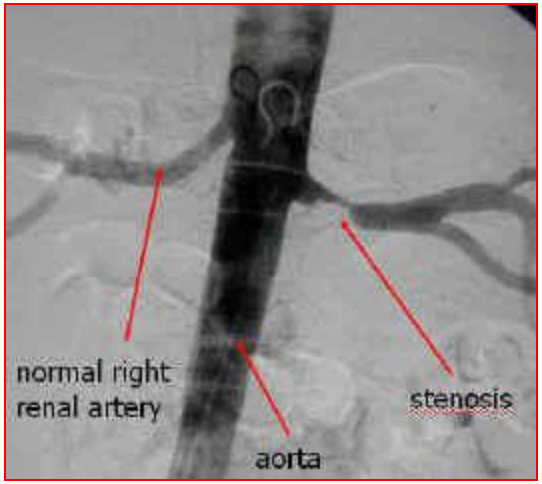
Atherosclerosis

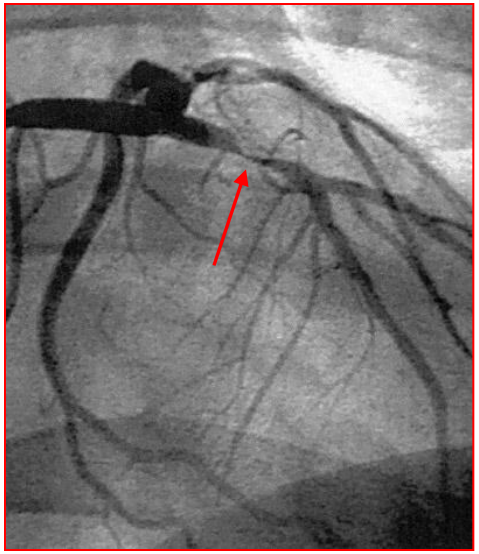


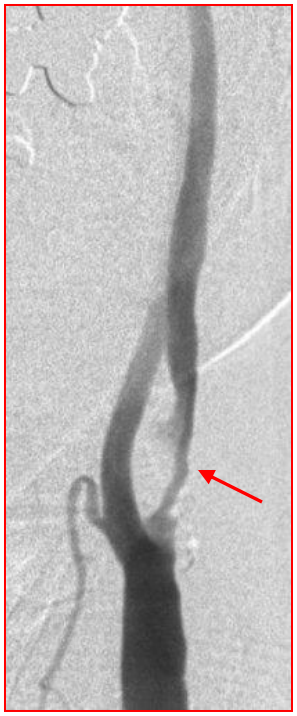


- Hardening, thickening, and narrowing of arteries due to buildup of fatty materials (cholesterol)
- Most common cause of cardiovascular disease in the USA
- Most common cause of heart attack and stroke
- Traditional risk factors include age, smoking, high blood pressure, high cholesterol, family history, and gender
- May cause significant narrowings in many vascular beds but may also result in aneurysm and dissection

Arterial stenosis

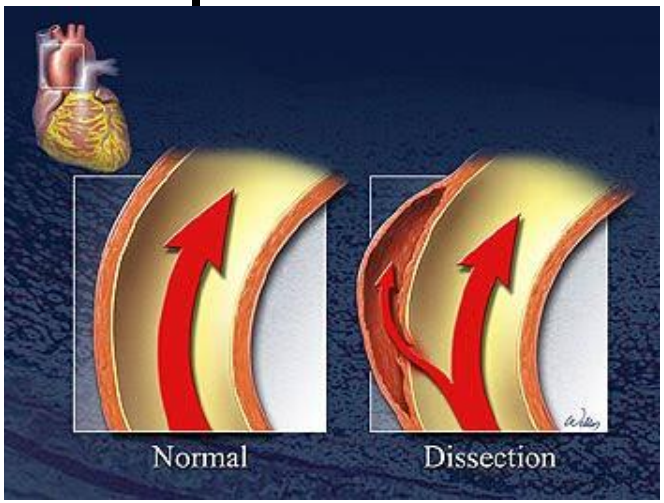


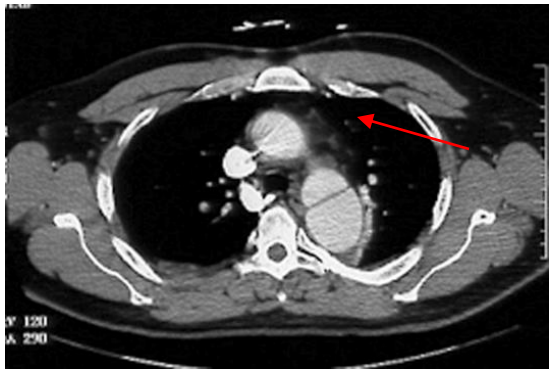




- Abnormal narrowing in a blood vessel
- May be any blood vessel
- Most common cause is atherosclerosis
- Can result in a “bruit”

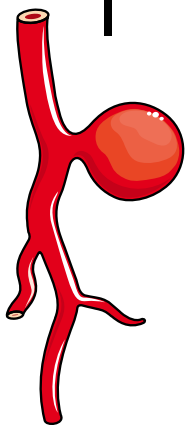
Arterial Dissection



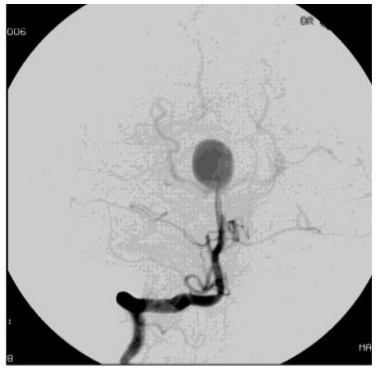


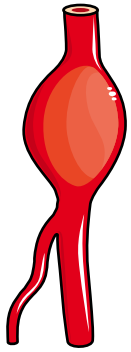
- Tear within the artery wall allowing blood to separate the wall layers
- Most common within the aorta but can happen in smaller vessels
- Weakening in wall from dissection → “pseudo”aneurysm
- FMD
  - Cervical artery dissection
  - Coronary dissection
  - Renal artery dissection
  - Mesenteric artery dissection

Arterial aneurysm



Saccular



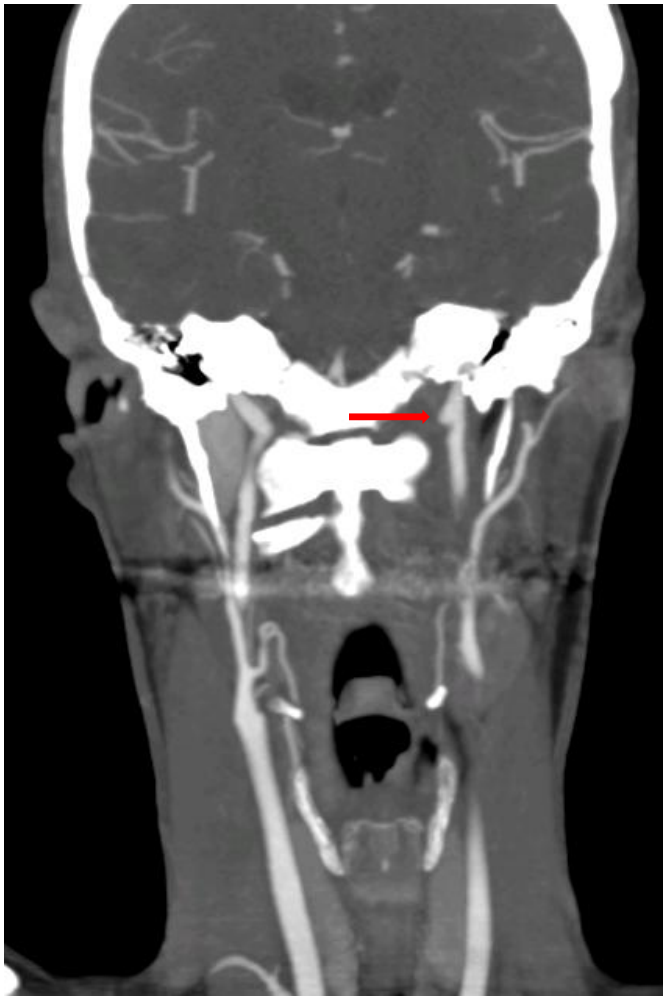


Fusiform

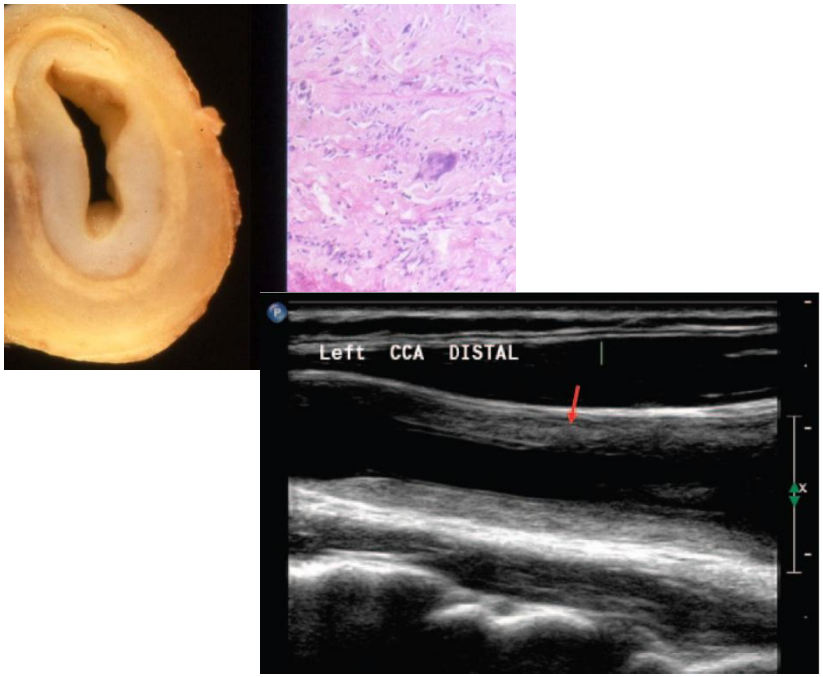
- Aneurysm
  - An abnormal bulge or ballooning of an artery caused by weakening of the arterial wall
  - Usually 2x normal caliber
- Types
  - Saccular – sac or pouch on one side of vessel wall
  - Fusiform – outward bulging in all directions
- “Ectatic”
  - Dilated artery wall but not quite large enough to be considered an aneurysm

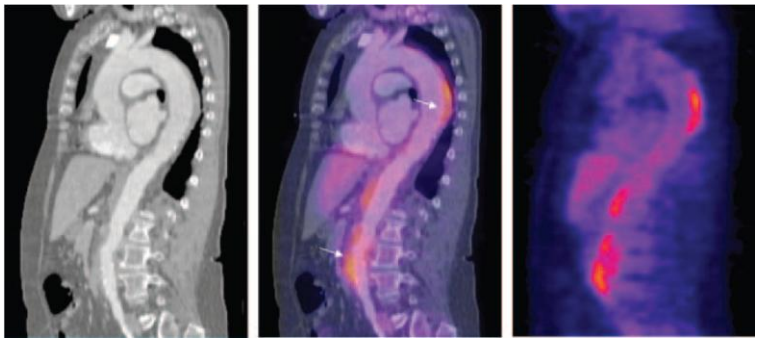
Pseudoaneurysm

- Not a true aneurysm
- Outpouching of the vessel in an area of prior dissection



Vasculitis





- Inflammation of the blood vessels, small, medium, or large
  - Polyarteritis nodosum, Takayasu's arteritis, giant cell arteritis
- Infectious or autoimmune
- May cause stenosis, aneurysm, dissection
- Treatment is with immunosuppression

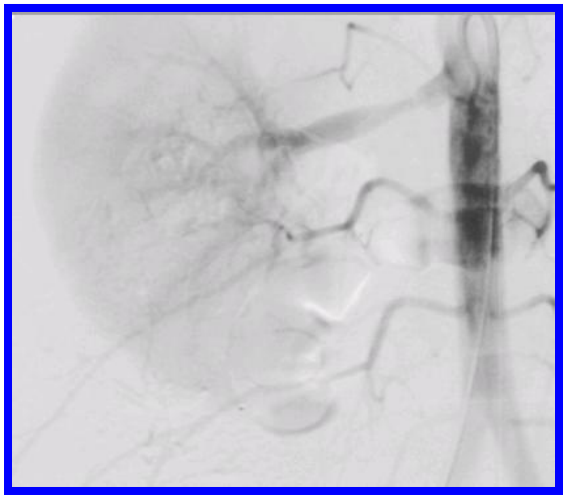
Fibromuscular Dysplasia

- Non-inflammatory, non-atherosclerotic disorder of the arteries
- Thickening of one of the layers of the artery resulting in
  - Arterial stenosis
  - Aneurysm
  - Dissection
- Type is determined according to angiographic appearance

Medial Fibroplasia

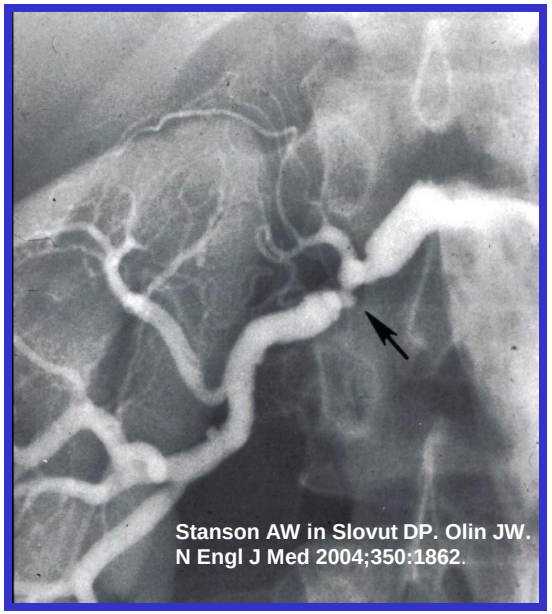
- Most common angiographic variant
- > 85% of cases
- Multiple areas of stenosis and aneurysmal dilatation
  - “String of beads”
  - “String of pearls”
  - “Stack of coins”
  - “Sausage links”

Intimal Fibroplasia



•< 10% of cases
•Variable angiographic appearance
  •Focal, severe concentric stenosis
  •Longer, tubular lesions:

Perimedial Fibroplasia

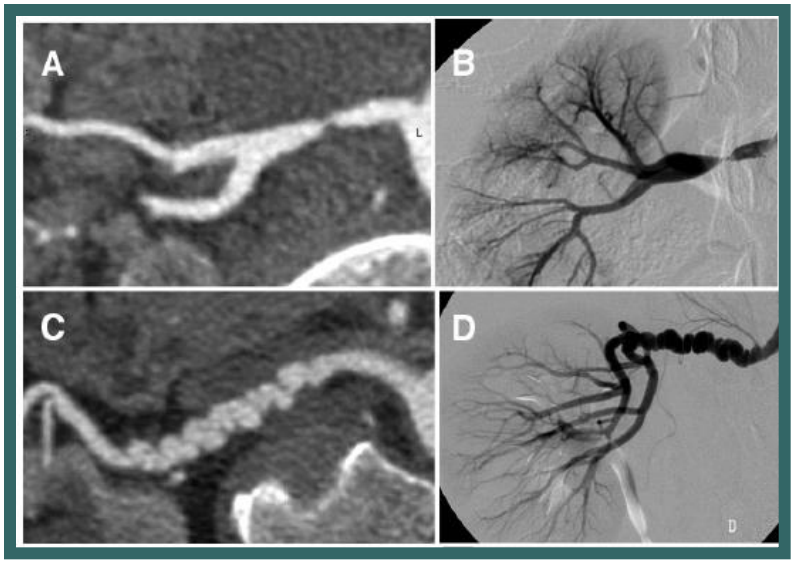


- Very rare
- Few beads of small caliber
- Often associated with severe stenosis

Classification can get complex

- Medial hyperplasia?
- Intimal fibroplasia?

- FMD no longer a primarily pathologic diagnosis
- Radiographic findings
- Nomenclature in evolution?
  - Unifocal
  - Multifocal

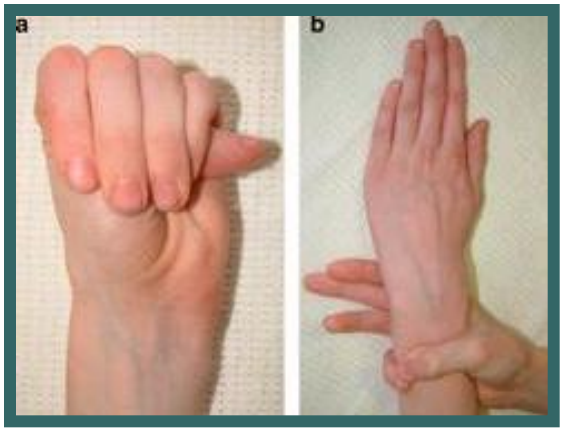


Savard et al. Circulation 2012;126:3062-3069

Physical Exam

Horner’s sign

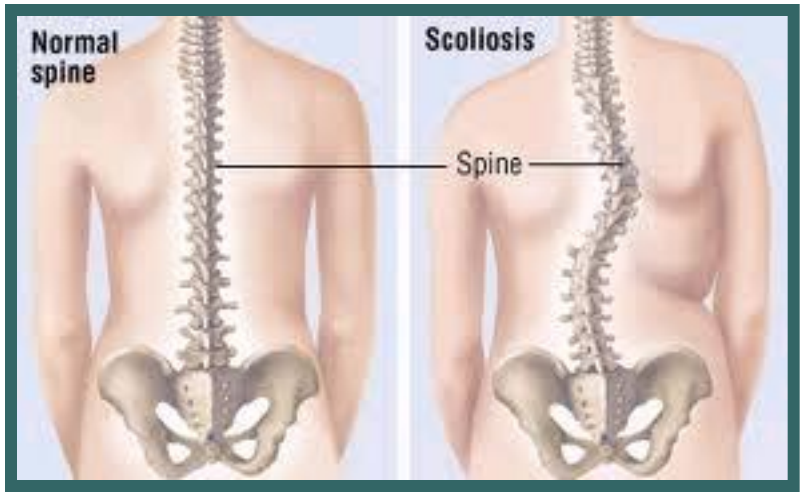
Torus Palantinus



Thumb and Wrist Sign

Elbow Hyperextension

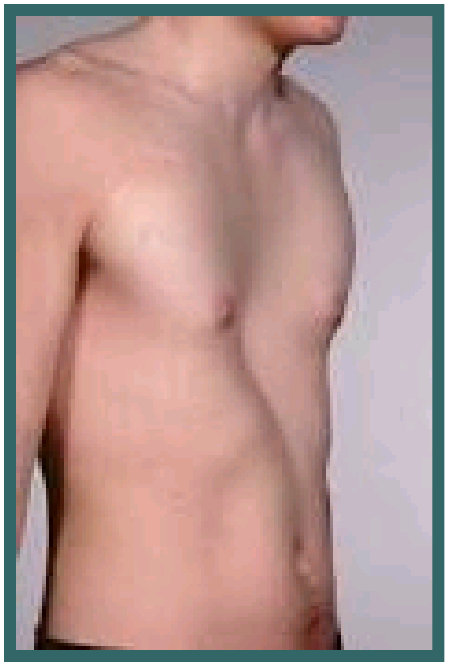
Knee Hyperextension



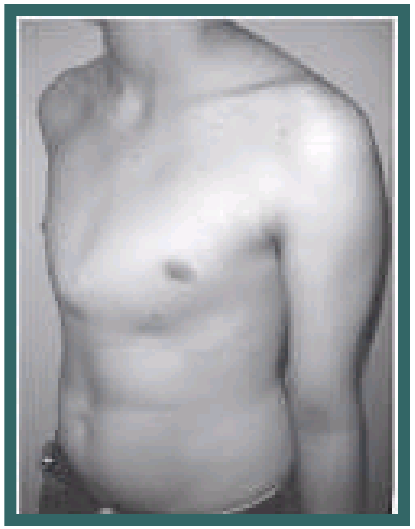
Scoliosis

Hyperextension

Skin Laxity

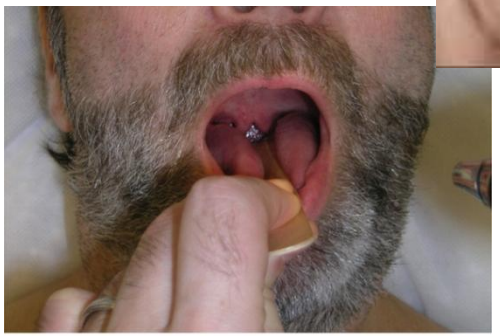


Pectus excavatum



Pectus carinatum

Connective Tissue Disease



- Any disease that targets the connective tissues of the body, including the blood vessels
- Heritable
  - Marfan's
  - Ehlers-Danlos
  - Neurofibromatosis
  - Loeys-Dietz
- Varied manifestations
  - Skin, blood vessels, joints, facial features

Bruit
(french for “noise”)

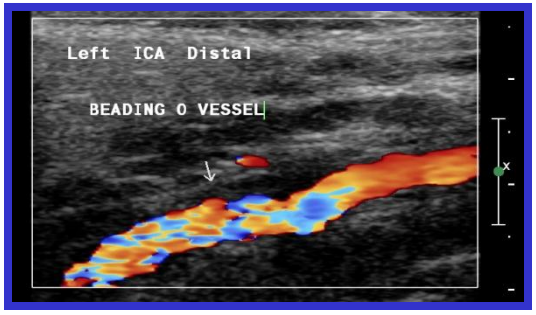
“the unusual sound that blood makes when it rushes past an obstruction in an artery when the sound is auscultated with a stethoscope” – Dr Wikipedia

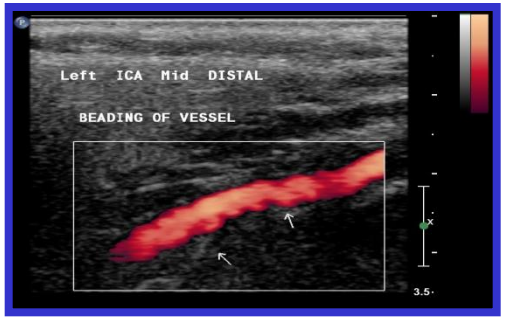
Imaging

Ultrasound

Ultrasound

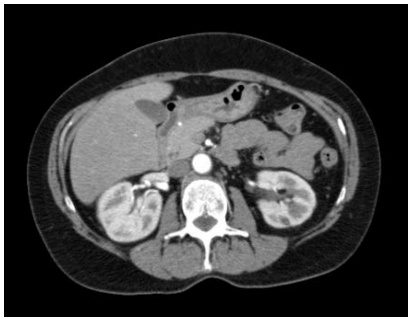
- Uses sound waves to create an image
- Non-invasive
- Can be used to image most arteries involved in FMD
- Test of choice for initial diagnosis and follow-up in most cases
- Pros: non-invasive, velocity information, no need for contrast
- Cons: technician dependent

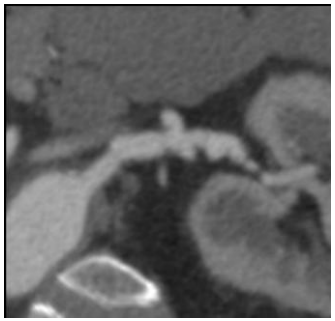


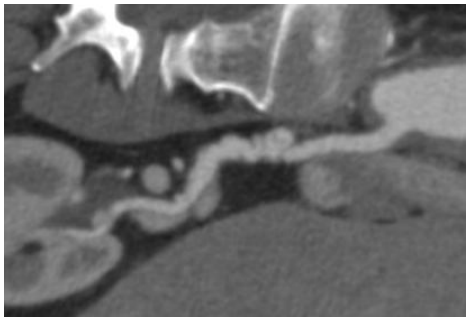


CT scan

- CT stands for computed tomography
- Uses x-rays to produce cross-sectional images of the body
- Much more resolution than traditional x-ray
- Pros: great detail
- Cons: radiation, contrast, no physiologic information



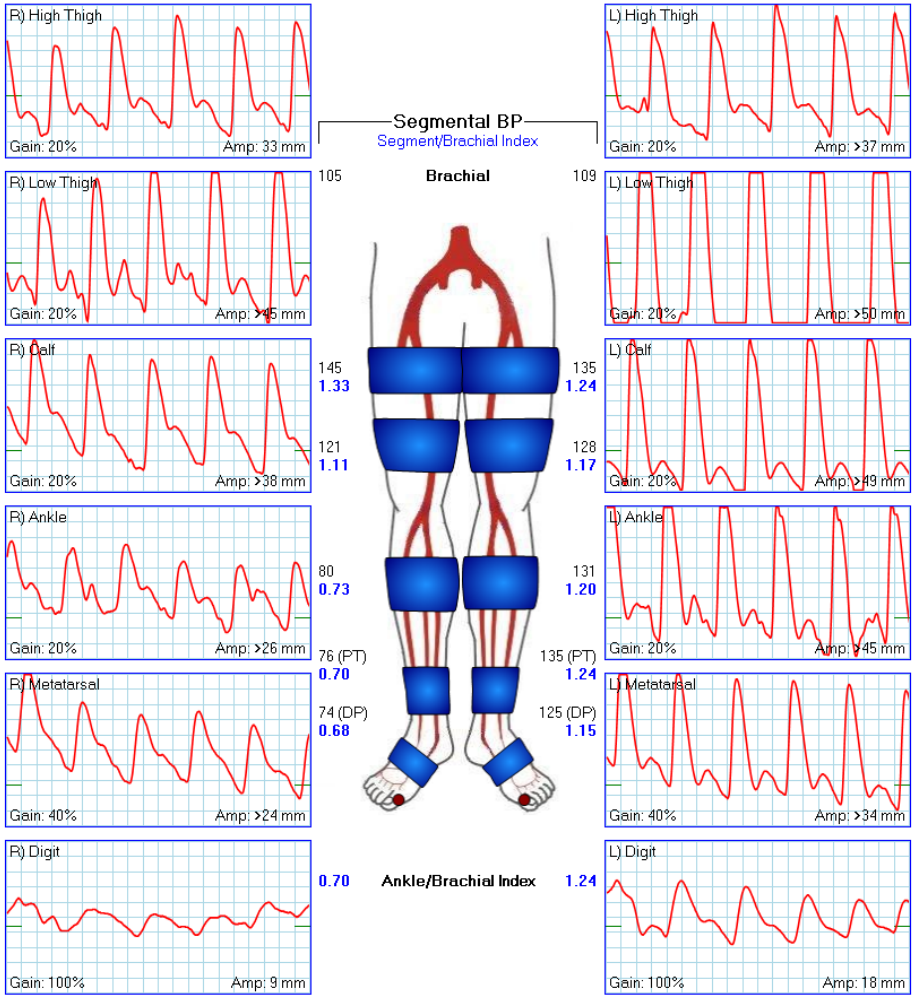




MRI

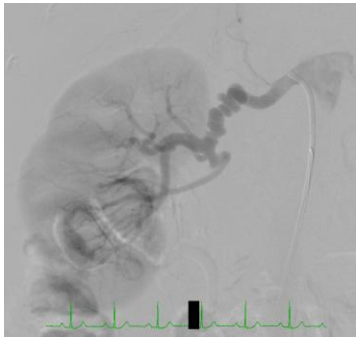
- Stands for magnetic resonance imaging
- Uses a magnet to detect magnetic fields emitted by atoms in the body to produce an image
- Can image the arteries well
- Pros: no ionizing radiation, can sometimes be done without contrast
- Cons: cannot be done in patients with metal implants or pacemakers, noisy during scan, more expensive, longer scanning time, claustrophobic
- Can be institution dependent – many protocols

ABI/PVR



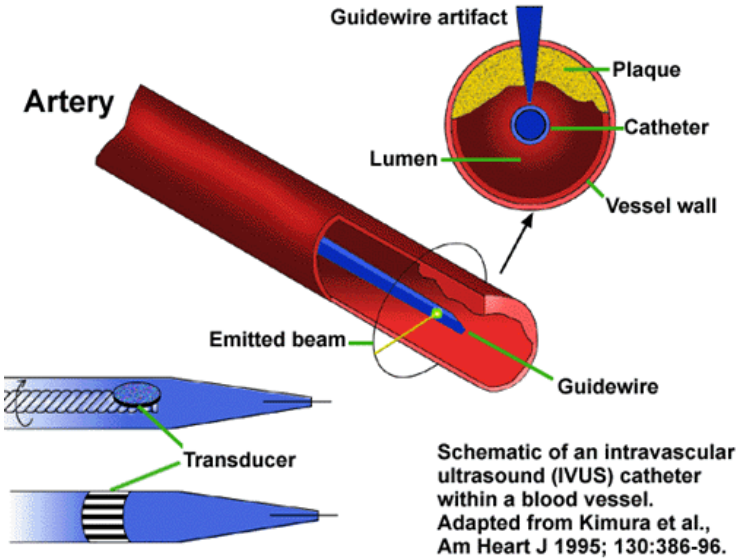
- Stands for ankle brachial index and pulse volume recording
- Series of blood pressure cuffs on the legs
- Can use exercise as well
- Pros: non-invasive, office-based, physiologic/functional test
- Cons: not a direct visualization of the arteries

Angiography



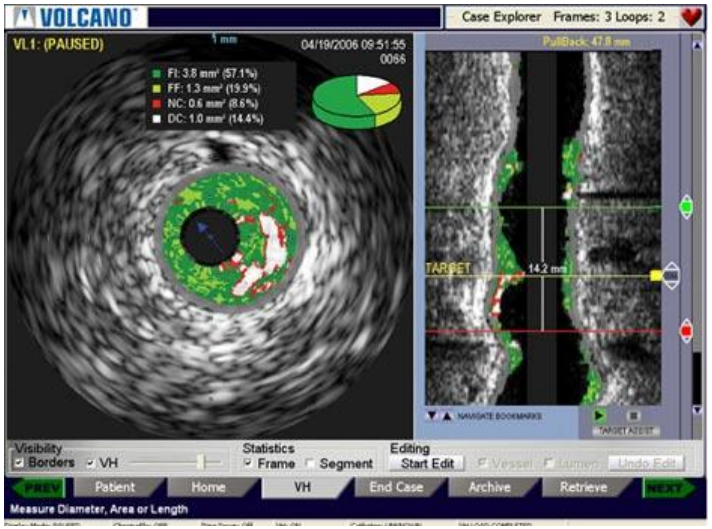
- Gold standard for the diagnosis of FMD
- Uses contrast dye to fill the lumen of the artery while a series of xrays are taken
- Pros: gold standard, able to take physiologic measurements, can proceed directly to intervention
- Cons: invasive, procedural risk, contrast

Intravascular Ultrasound

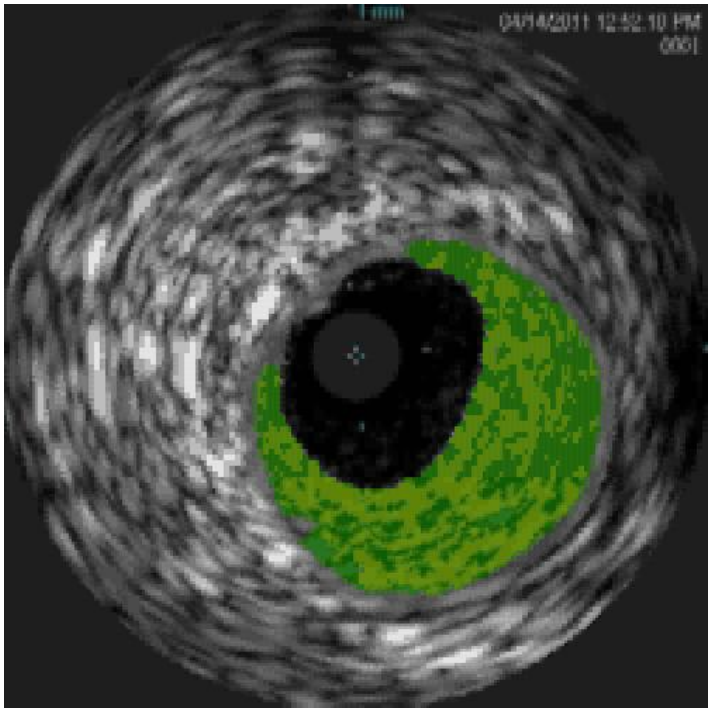


Schematic of an intravascular ultrasound (IVUS) catheter within a blood vessel. Adapted from Kimura et al., Am Heart J 1995; 130:386-96.

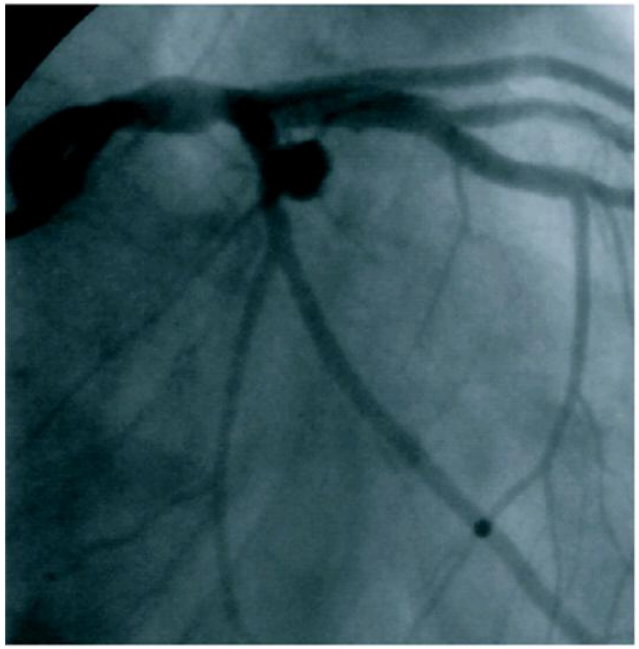
- Ultrasound from inside an artery
- Catheter based during an angiography
- Pros: ability to visualize inside the vessel for stenosis, webs, plaque
- Cons: invasive, not yet widely available

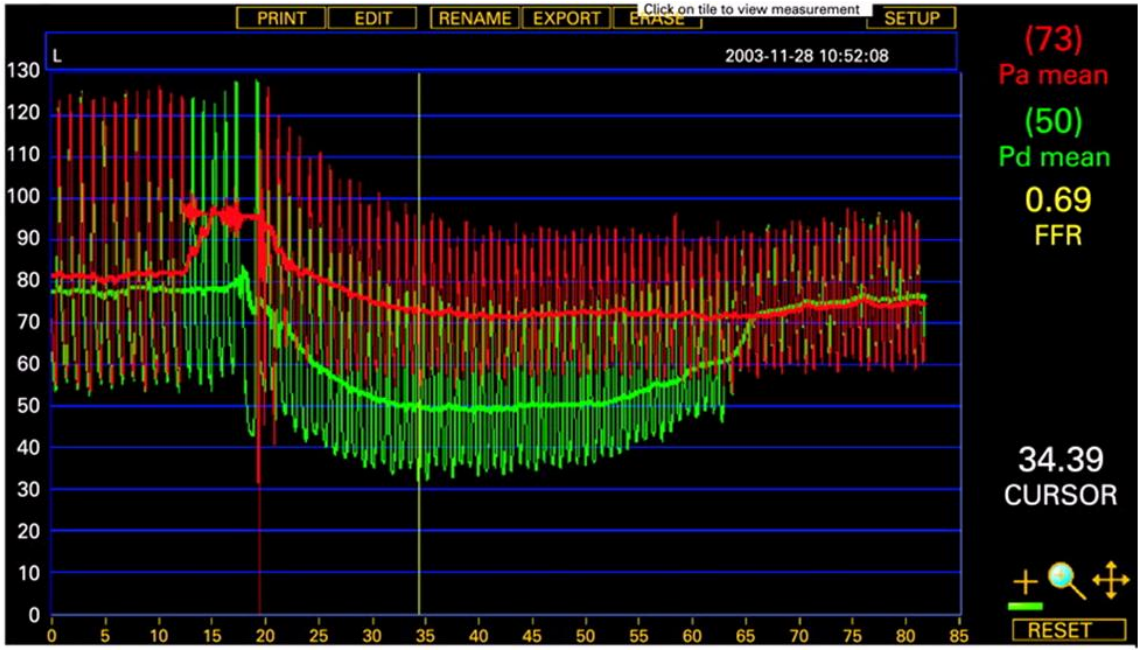


Intravascular Ultrasound



Fractional Flow Reserve

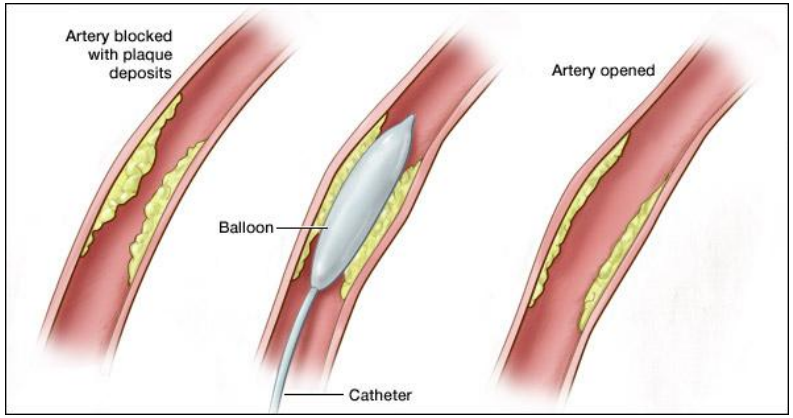


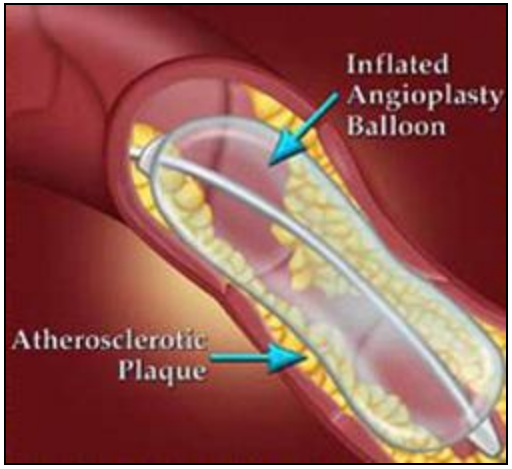


Treatment

Invasive Treatment

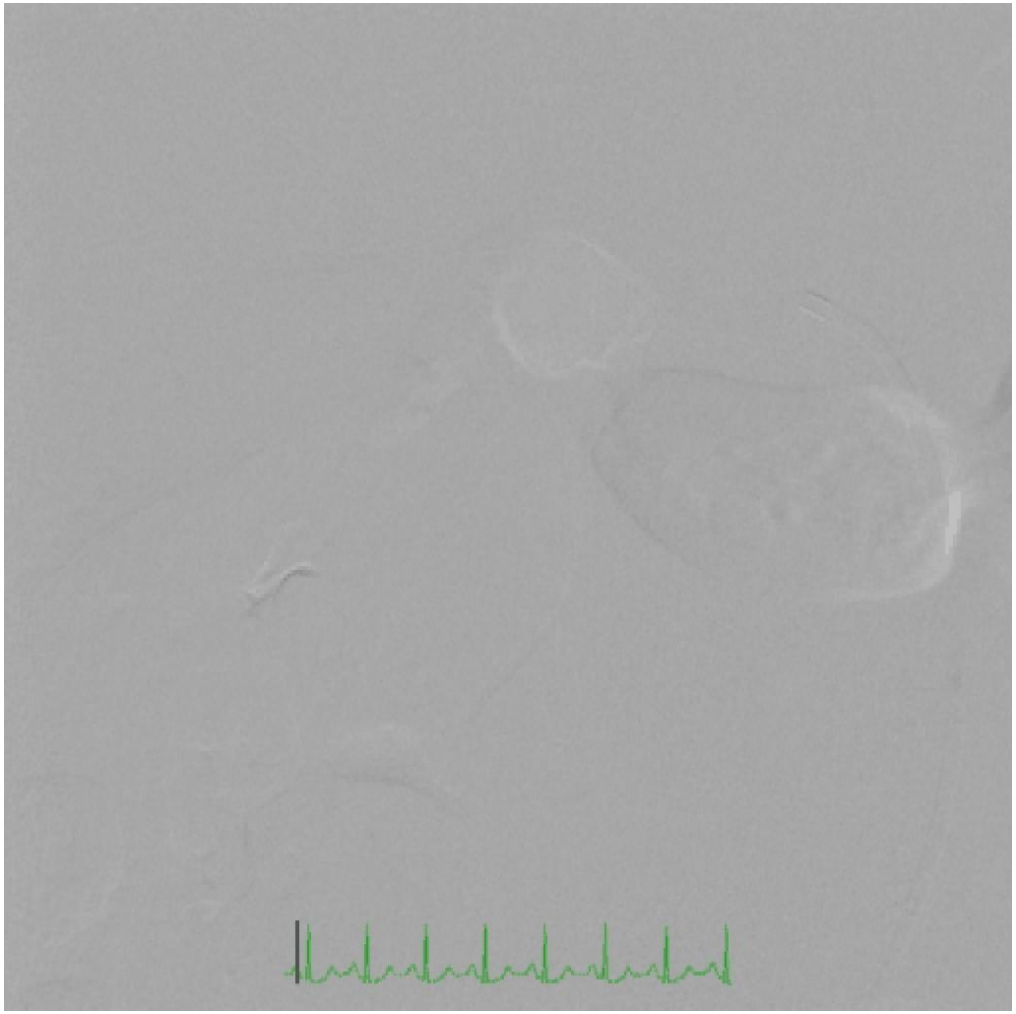
Angioplasty



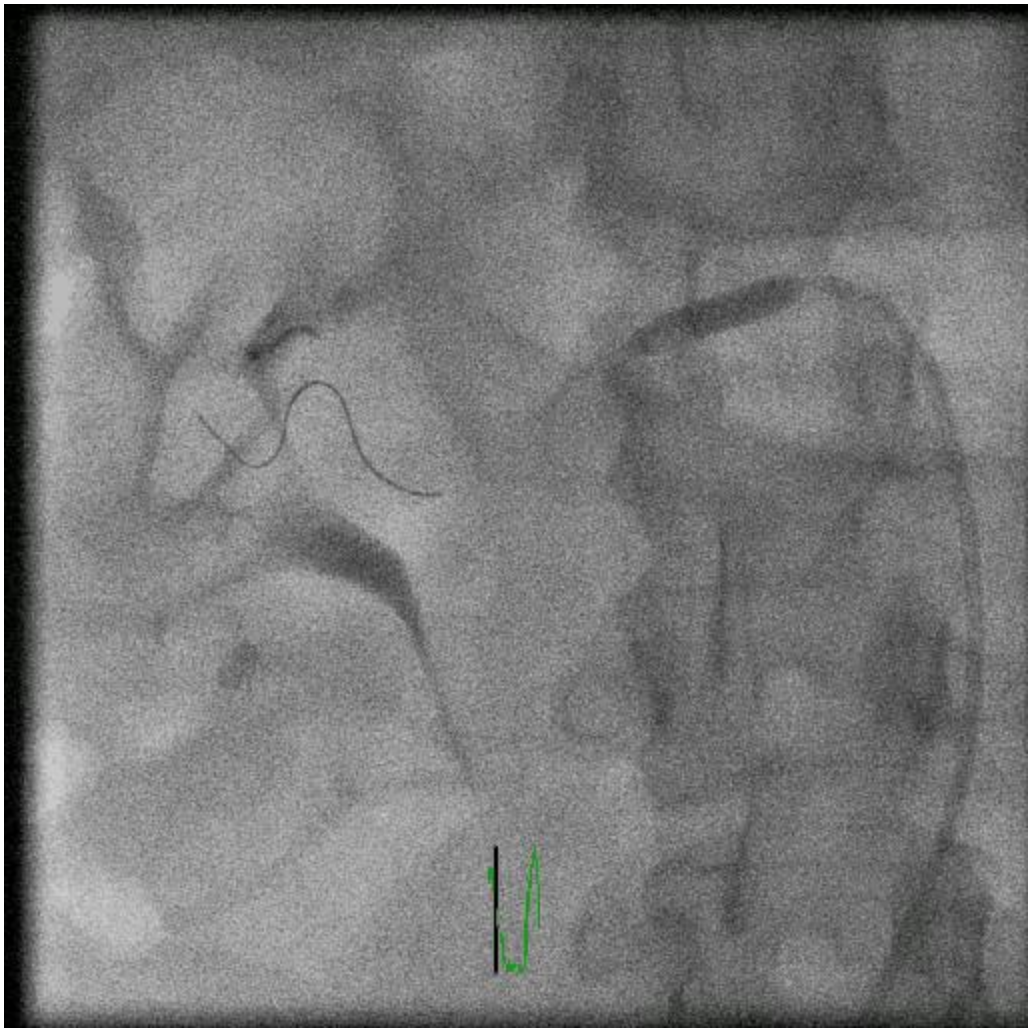


- Mechanically opening a narrowed vessel with a balloon mounted on a catheter
- Usually the treatment of choice for FMD, if necessary

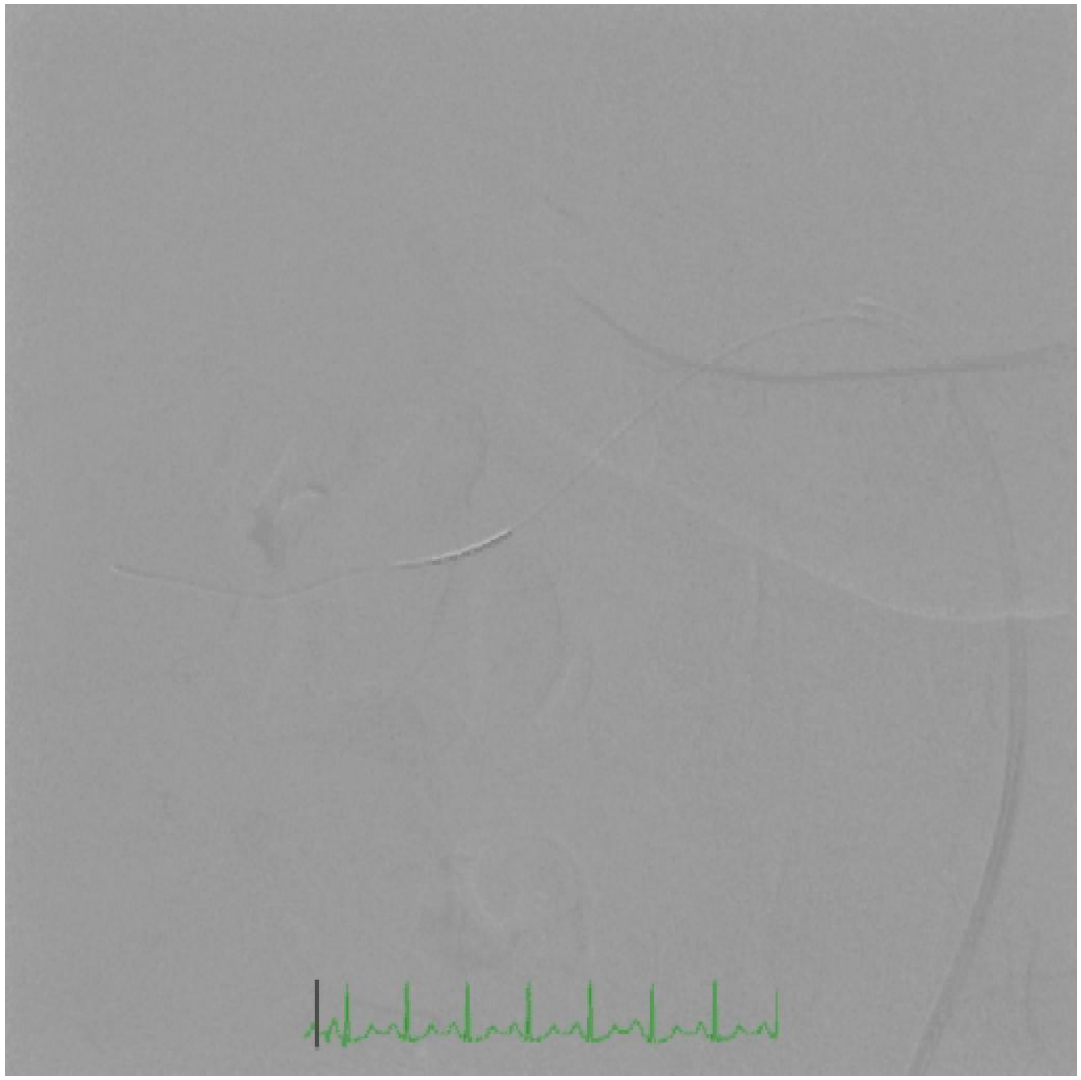
Pre-angioplasty



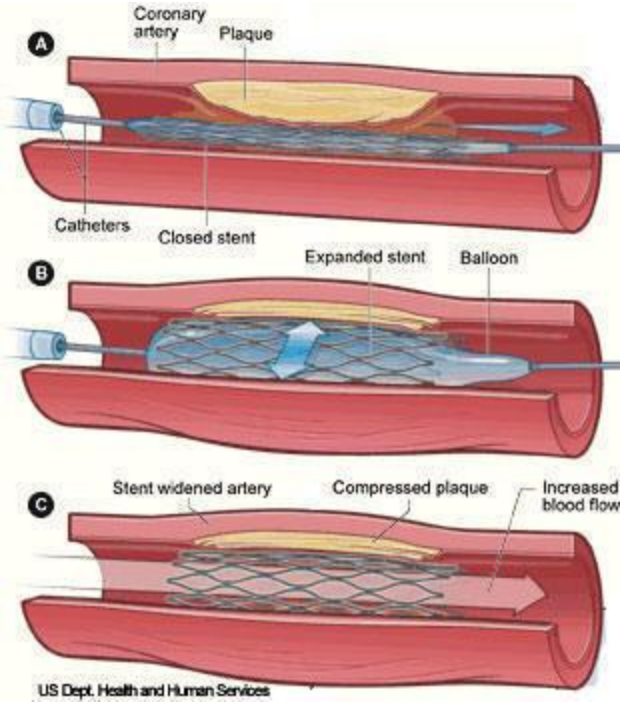
Angioplasty



Post-angioplasty

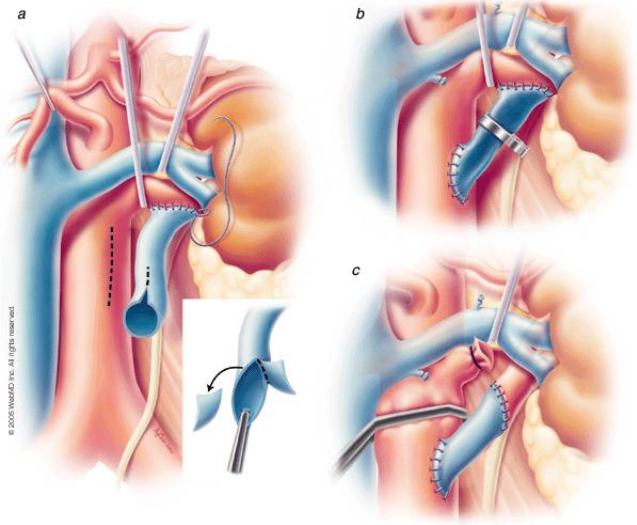


Stent



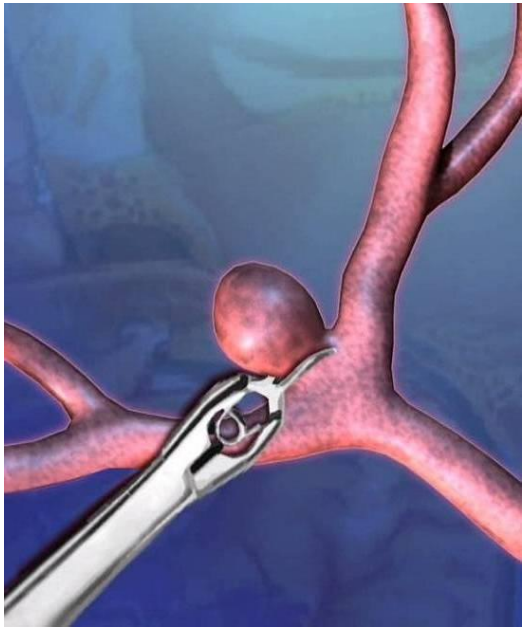
- Artifical tube inserted into a natural passage in the body
- Vascular stents are metallic, can be bare metal, drug eluting, or covered
- In FMD, use is generally reserved for cases of dissection

Bypass Surgery



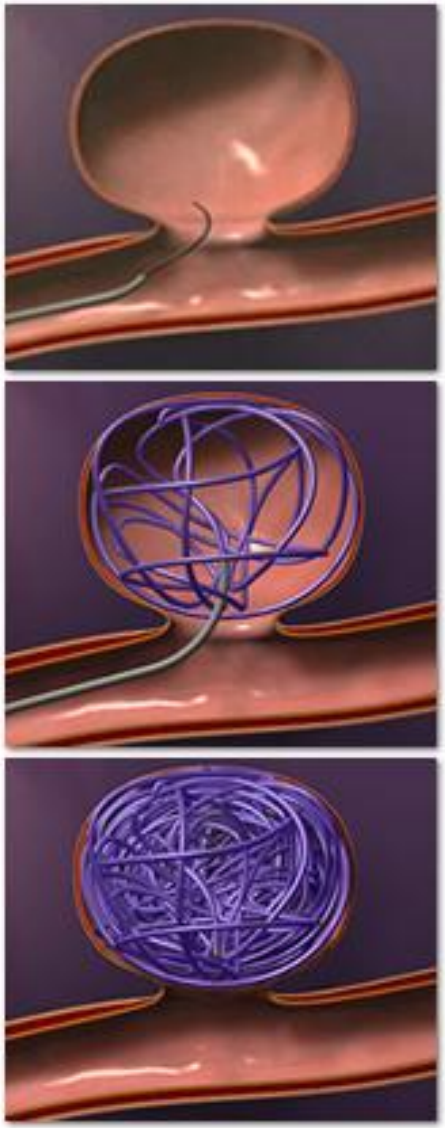
- Surgical procedure where an artery or vein from elsewhere in the body or an artificial graft is used the bypass a diseased artery and supply blood flow to the organ
- Generally reserved for cases not amenable to PCI

Aneurysm Clipping



- Surgical procedure for treatment of cerebral aneurysms
- Requires craniotomy (removal of part of skull)
- Aneurysm is clipped with a titanium clip

Aneurysm Coiling



- Endovascular procedure
- Tiny platinum coils deployed into aneurysm
- Coils block blood flow into the aneurysm and prevent rupture

Thank you