Raynaud's phenomenon
Acute limb ischemia
Renal vascular diseases
Aortic diseases

Prof. Mazin Zamel Alshibani

Raynaud's phenomenon

Cold and emotional stimuli may trigger vasospasm, leading to the characteristic sequence of:

a) digital pallor due to vasospasm
b) cyanosis due to deoxygenated blood
c) redness due to reactive hypereamia.

Primary Raynaud's phenomenon (or disease)

- This affects 5–10% of young women aged 15–30 years in temperate climates and may be familial.
- It does not progress to ulceration or infarction, with no significant pain.
- The underlying cause is unclear.
- No investigation is necessary, the patient should be reassured and advised to avoid exposure to cold.
- Long-acting nifedipine may be helpful.

Secondary Raynaud's phenomenon (or syndrome)

* This occurs in older people in association with
1. CTD (most commonly CREST syndrome, also known as the limited cutaneous form of systemic sclerosis (lcSSc), is a multisystem CTD, refere to (calcinosis, R. phenomenon, esophageal dysmotility, sclerodactyly, telengectasia),
2. vibration-induced injury
3. thoracic outlet obstruction (cervical rib)

* Unlike primary disease, it is often associated with fixed obstruction of the digital arteries, fingertip ulceration, necrosis and pain.

* The fingers must be protected from cold exposur and trauma. Infection requires treatment with antibiotics, and surgery should be avoided if possible.

* Sympathectomy helps for a year or two.
* Prostacyclin infusions are sometimes beneficial.

Acute limb ischaemia

Symptoms and signs of acute limb ischemia:

- *Pain, pallor, pulselessness (may be absent in complete ischemia and can be present in chronic ischemia)*
- *Perishing cold: unreliable*
- *Parasthesia: and paralysis: important feature of impending irreversible ischemia*

✔ This is most frequently caused by acute thrombotic occlusion of a pre-existing stenotic arterial segment, thromboembolism, and trauma that may be iatrogenic.

✔ Pain on squeezing the calf indicates muscle infarction and impending irreversible ischaemia.

- ✔ Rx, if no contraindications (e.g acute aortic dissection or trauma, particularly head injury), an intravenous bolus dose of heparin (3000–5000 units) should be administered to limit propagation of thrombus and protect the collateral circulation.

- ✔ Acute limb ischaemia due to thrombosis in situ can usually be treated medically with intravenous heparin, antiplatelet agents, high-dose statins, and intravenous fluids to avoid dehydration, correction of anaemia, oxygen and sometimes prostaglandins.

- ✔ Embolism will normally result in extensive tissue necrosis within 6 hours unless the limb is revascularised.

- ✔ The indications for thrombolysis, remain controversial

Renal vascular diseases: (RVD)

Diseases which affect renal blood vessels may cause renal ischaemia, leading o acute or chronic kidney disease or secondary hypertension, it is an important cause of ESRD.

Renal artery stenosis (RAS)

* RAS is a relatively uncommon disorder, which presents clinically with hypertension.

* occur in about 2% of unselected patients with hypertension but may affect up to 4% of older patients with hypertension who have evidence of atherosclerotic disease.

* Most cases of RAS are caused by atherosclerosis (proximal part of RA) in elderly but fibromuscular dysplasia (distal) in younger patients.

* Rare causes include vasculitis, thromboembolism and aneurysms of the renal artery.

Pathophysiology

- RAS results in a reduction in renal perfusion pressure, which activates the renin–angiotensin aldosteron system (RAAS), leading to increased circulating levels of angiotensin II.

- This provokes vasoconstriction with increases aldosterone production by the adrenal and causing sodium retention.

- Significant reduction of renal blood flow occurs when there is greater than 70% narrowing of the artery.

- Atherosclerosis is the most common cause.

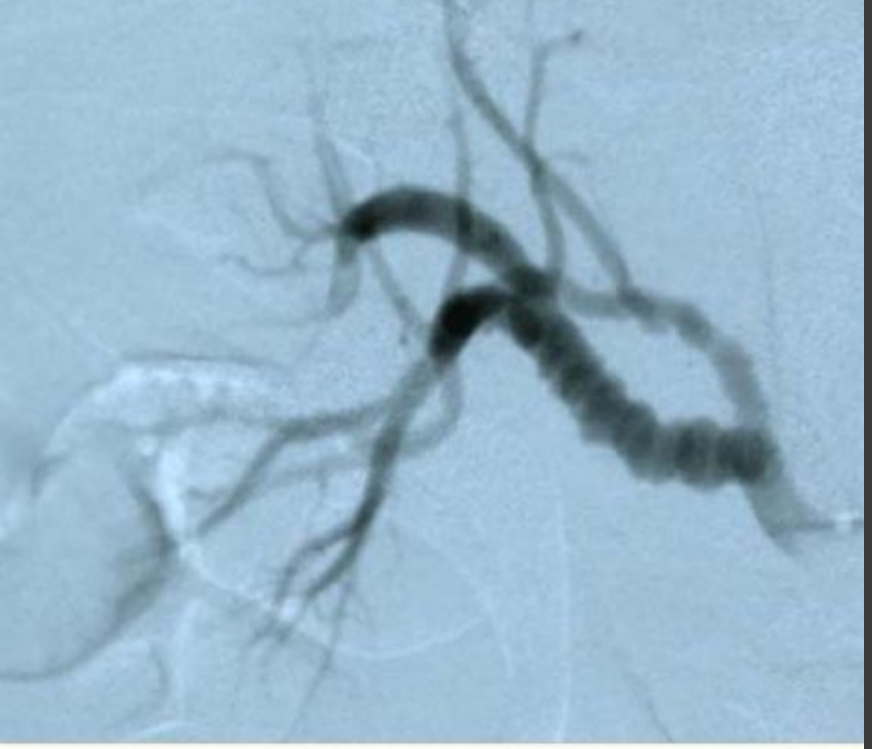
- The characteristic lesion is an ostial stenosis associated with atherosclerosis within the aorta and affects other major branches, particularly the iliac vessels.

- FMD is an uncommon disorder of unknown cause. It is characterized by hypertrophy of the media (medial fibroplasia). It most commonly presents with HT in patients aged 15–30 years, and women are affected more frequently than men. Irregular narrowing (beading) may occur in the distal renal artery

- Rarely, RAS may occur as a complication of large-vessel vasculitis, such as Takayasu's arteritis and PAN.

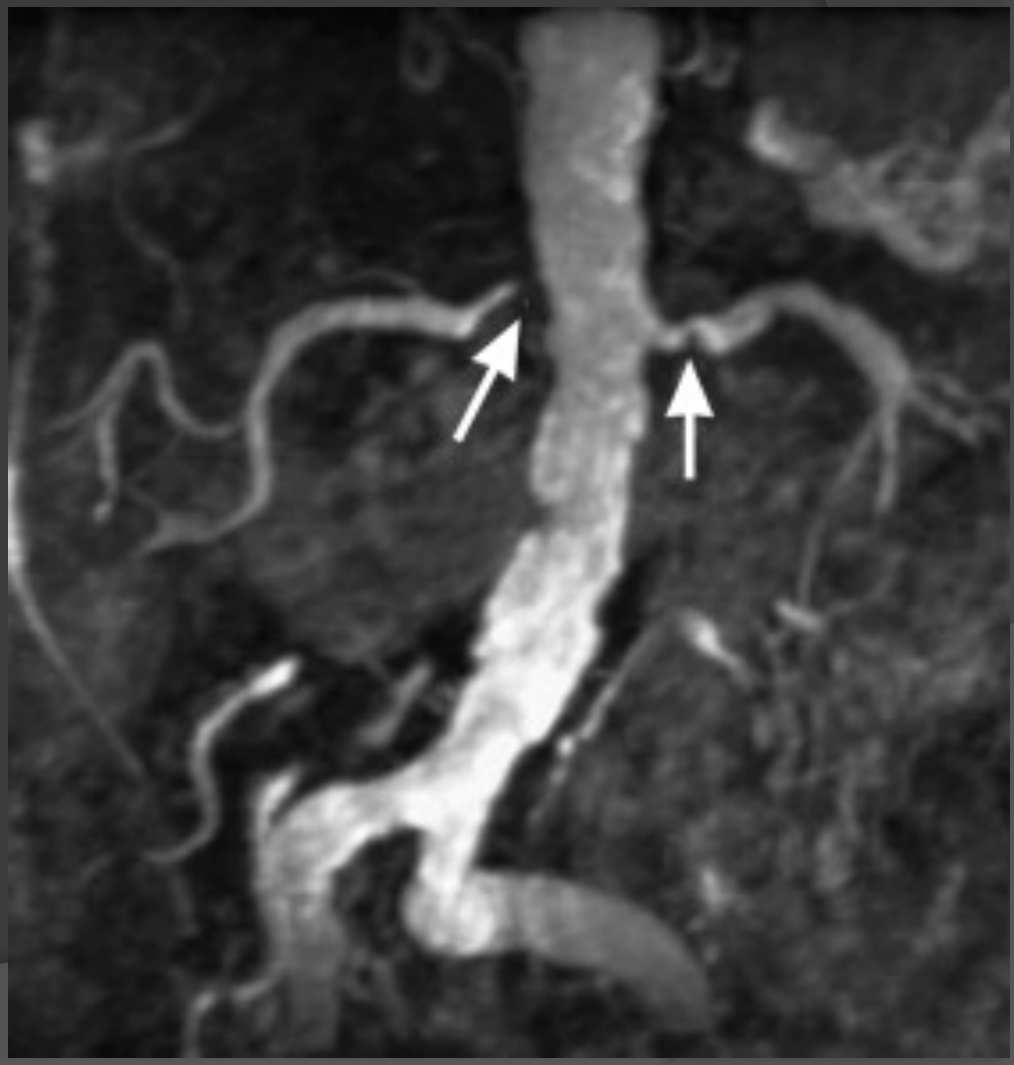
- If the progression is gradual, collateral vessels may develop and some kidney function may be preserved, preventing infarction and loss of kidney structure.

- About 85% of patients with RAS will not develop progressive renal impairment, and the stenosis may be insignificant and not responsible for coexisting essential HT



Angiogram showing fibromuscular RAS

Atherosclerotic RAS



Clinical features

- RAS can present with sever HT, renal failure (with bilateral disease), a deterioration in renal function when ACE inhibitors or ARBs are used, or acute pulmonary oedema.
- Although many patients experience a slight drop in GFR when commencing these drugs, an increase in serum creatinine of 25% or more raises the possibility RAS.
- Acute pulmonary oedema is particularly characteristic of bilateral renovascular disease.
- Clinical evidence of PAD may be observed, particularly in the legs, in older patients with atherosclerotic RAS

Investigations

- Imaging of the renal vasculature with either CT angiography or MR angiography.
- Biochemical testing may reveal impaired renal function and an elevated plasma renin level, sometimes with hypokalaemia due to hyperaldosteronism
- Ultrasound may also reveal a discrepancy in size between the two kidneys.

Patient with high likelihood of RAS:

1. Onset of HT ≤30 years of age or ≥55 years of age
2. Malignant, accelerated or resistant HT
3. Unexplained renal dysfunction
4. Development of azotemia when using ACE inhibitors or ARBs
5. Unexplained kidney size discrepancy of >1.5 cm
6. Multivessels CAD
7. PAD
8. Unexplained flash pulmonary edema
9. Unexplained CHF or refractory angina pectoris

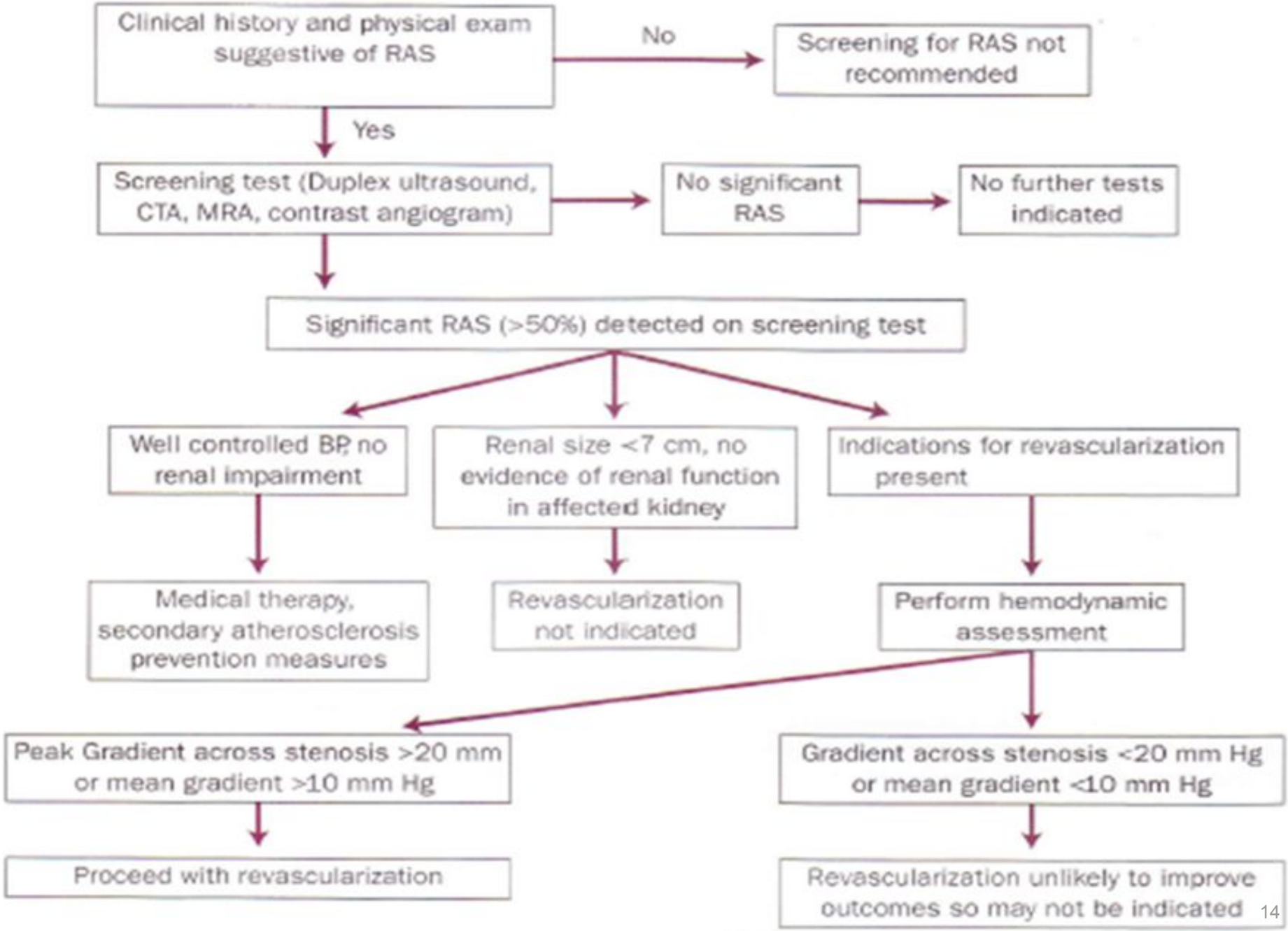
Indications for revascularization:

1. Accelerated HT, resistant HT, malignant HT, HT with unilateral small kidney & HT with intolerance to medication

2. Renal insufficiency

3. Recurrent CHF or flush pulmonary edema, refractory HF or refractory angina pectoris

Approach to Patients With Renal Artery Stenosis



Management

- In elderly with RAS:

1. Medical therapy with antihypertensive drugs, with statins and low-dose aspirin in those with atherosclerotic disease.

2. Angioplasty and stenting can be successful in atherosclerotic disease. The risks of angioplasty and stenting include renal artery occlusion, renal infarction, and atheroemboli

- In young with RAS: interventions to correct the vessel narrowing should be considered in the presence of indication for revascularization. Angioplasty is the most commonly used technique in FMD, where correction of the stenosis has success in improving blood pressure and protecting renal function. .

Acute renal infarction

- This is an uncommon condition that occurs as the result of sudden occlusion of the renal arteries.
- The presentation is typically with loin pain of acute onset, usually in association with haematuria, but pain may be absent.
- Severe HT is common but not universal.
- Blood levels of LDH and CRP are commonly raised.
- The condition may be caused by local atherosclerosis (atheroembolic) or by thromboemboli from a distant source, where occlusion may occur in branch arteries distal to the main renal artery.

* If occlusion of the main renal arteries is bilateral or if there is occlusion of a single functioning kidney, the presentation is with AKI and the patient is typically anuric

* Patients with bilateral occlusion usually have evidence of widespread vascular disease and may show evidence of aortic occlusion, with absent femoral pulses and reduced lower limb perfusion.

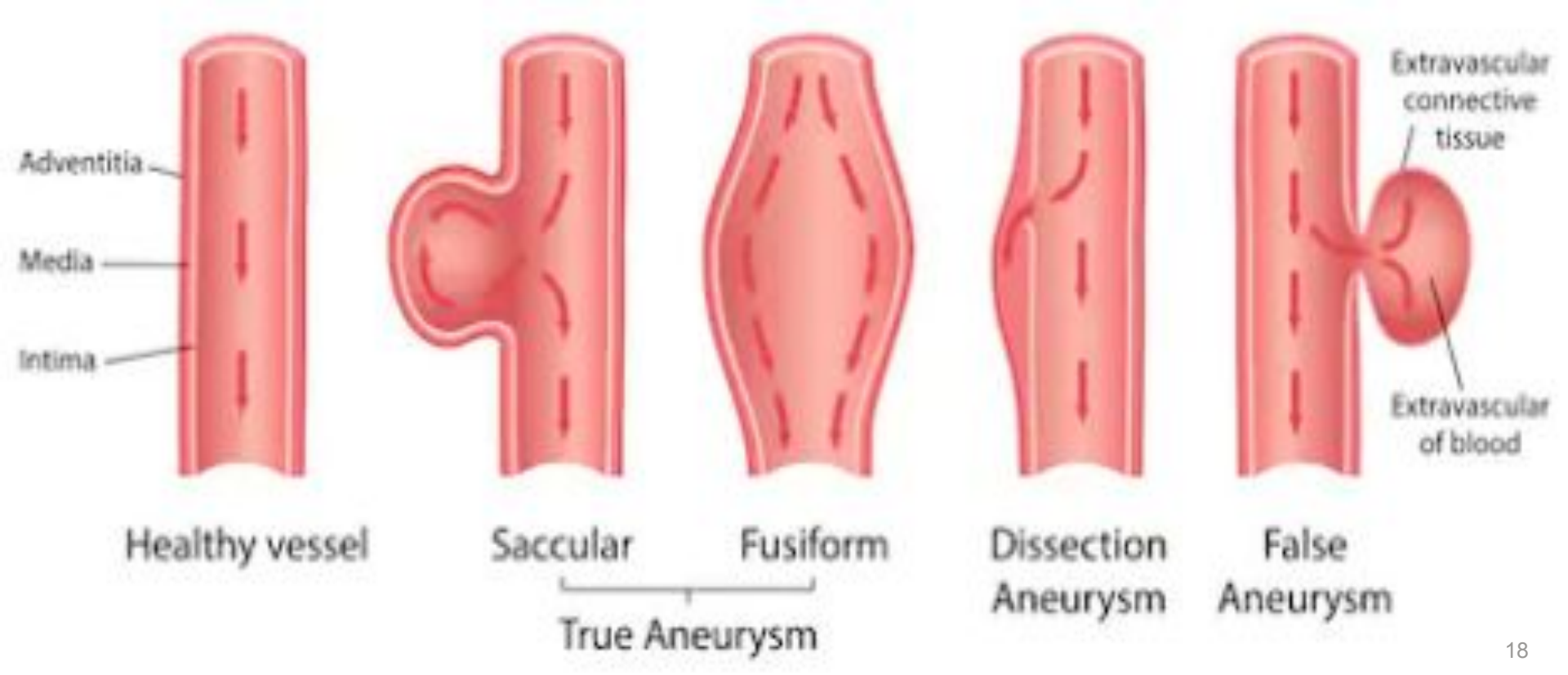
* Management is largely supportive, and includes anticoagulation if a source of thromboembolism is identified.

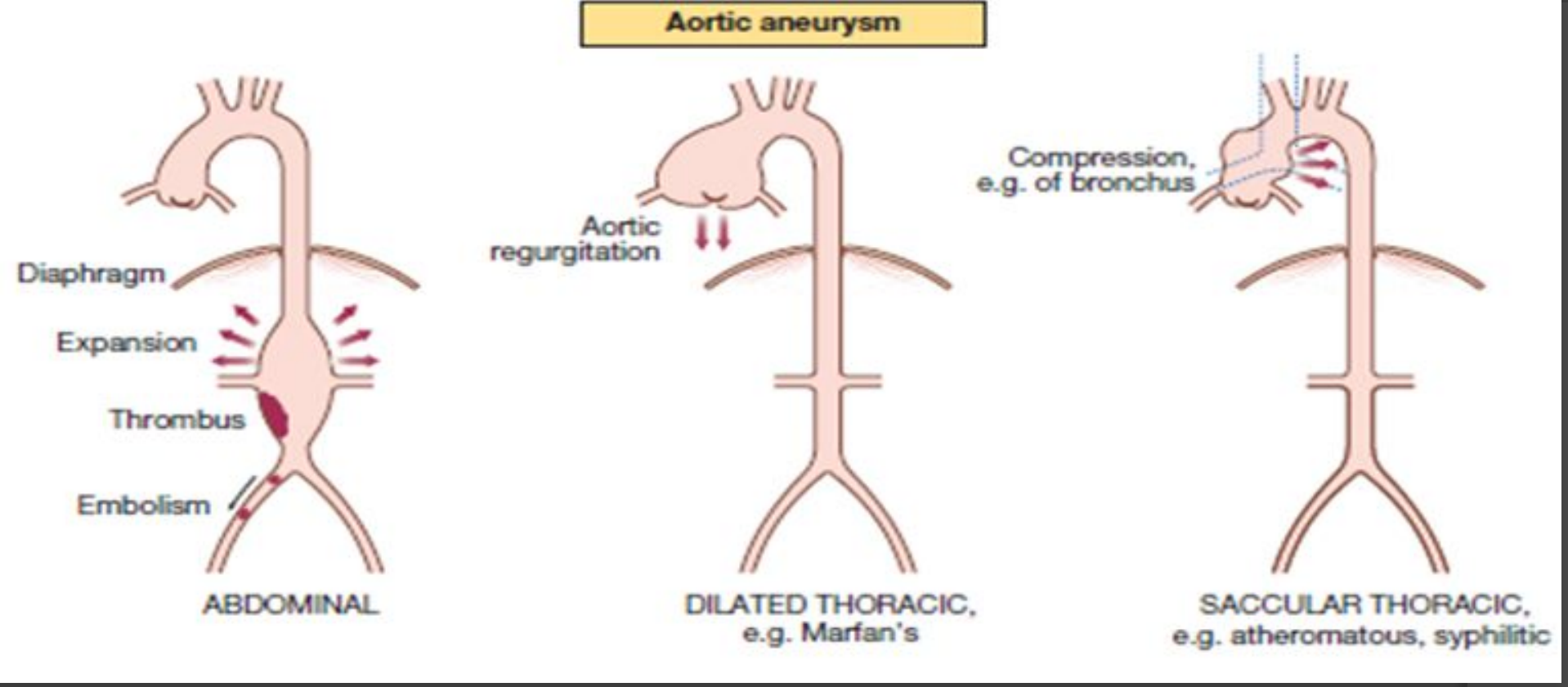
* Sometimes possible to stenting of an acutely blocked main renal artery to try to restore renal blood flow and kidney function

Diseases of the aorta

Aortic aneurysm

Abnormal dilatation of the aortic lumen; a true aneurysm involves all layers of the wall, against false aneurysm or pseudoaneurysm (collection of blood leaking completely out of an artery, but confined next to the vessel by the surrounding tissue)





Aetiology and types of aneurysm:

1. *Non-specific aneurysms:* may occur in association with occlusive disease.

- Aneurysmal disease tends to run in families.
- The most common site for 'nonspecific' aneurysm formation is the infrarenal abdominal aorta.
- The suprarenal abdominal aorta and a variable length of the descending thoracic aorta may be affected in 10–20% of patients
- Ascending aorta is usually spared

2. Marfan's syndrome

* CTD inherited as an AD trait and is caused by mutations in the *fibrillin* gene on chromosome 15.

* Affected systems include the skeleton (arachnodactyly, joint hypermobility, scoliosis, chest deformity and high arched palate), the eyes (dislocation of the lens) and the CVS (aortic disease and MR).

* Weakening of the aortic media leads to aortic root dilatation, AR and dissection

* Pregnancy is particularly hazardous.

* Chest X-ray, echo, MRI or CT may detect aortic dilatation at an early stage and can be used to monitor the disease.

* Treatment with BB reduces the rate of aortic dilatation and the risk of rupture.

3. Aortitis

* Syphilis is a rare cause of aortitis that produces saccular aneurysms of the ascending aorta containing calcification.

* Other rare conditions associated with aortitis include Takayasu's disease, Reiter's syndrome, giant cell arteritis and ankylosing spondylitis.

4. Thoracic aortic aneurysms

* These may produce chest pain, AR, compressive symptoms such as stridor (trachea, bronchus) and hoarseness (recurrent laryngeal nerve), and SVC syndrome. If they erode into adjacent structures, e.g. aorto-oesophageal fistula, massive bleeding occurs.

5. Abdominal aortic aneurysms

- Abdominal aortic aneurysms (AAAs) are present in 5% of men aged over 60 years and 80% are confined to the infrarenal segment.
- Men are affected three times more commonly than women.
- The usual age at presentation is 65–75 years for elective presentations and 75–85 years for emergency presentations.
- Ultrasound is the best way for diagnosis and following up in asymptomatic aneurysms.
- CT provides more accurate information.

Management:

* All symptomatic AAAs should be considered for repair.

* Distal embolization is a strong indication for repair, regardless of size.
* Most patients with a ruptured AAA do not survive to reach hospital, but if they do , surgery is thought to be appropriate.

* Open AAA repair has been the treatment of choice in both the elective and the emergency settings, and replacing the aneurysmal segment with a prosthetic (usually Dacron graft).

* Endovascular aneurysm repair (EVAR), using a stent-graft introduced via the femoral arteries. It is cost effective and likely to become the treatment of choice for infrarenal AAA.

Common presentation of aortic aneurysm:

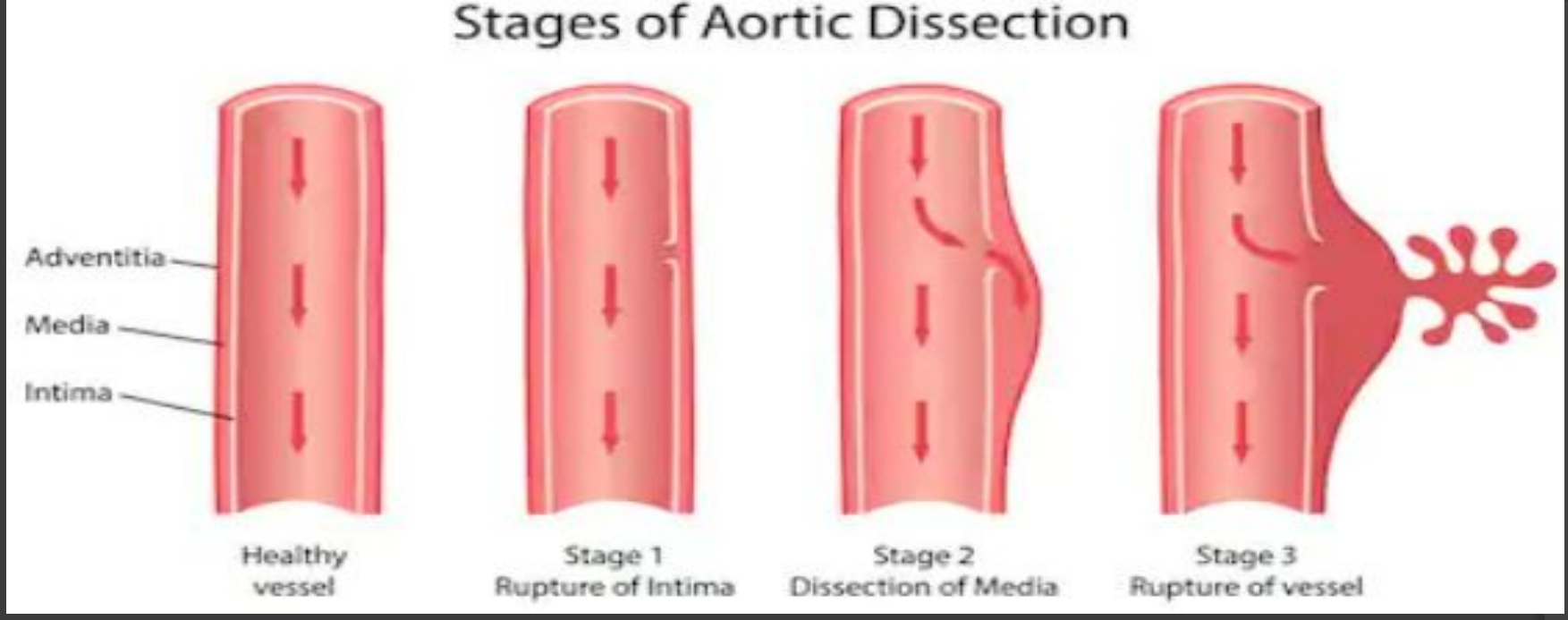
- Incidental: on physical examination, plain X-ray, most commonly abdominal US, sometime large AAA can be difficult to feel, so many remain undetected until rupture
- Pain: in the central part of abdomen, back, loin, iliac fossa, and groin
- Thromboembolic complication: thrombus in aneurysmal sac may act as a source for emboli to the lower limb, less commonly, the aorta may undergo thrombotic occlusion
- Compression: obstruction to the surrounding structure as duodenum causing obstruction with vomiting, pressure to IVC, may cause odema and DVT
- Rupture: into the retroperitoneum, peritoneal cavity.

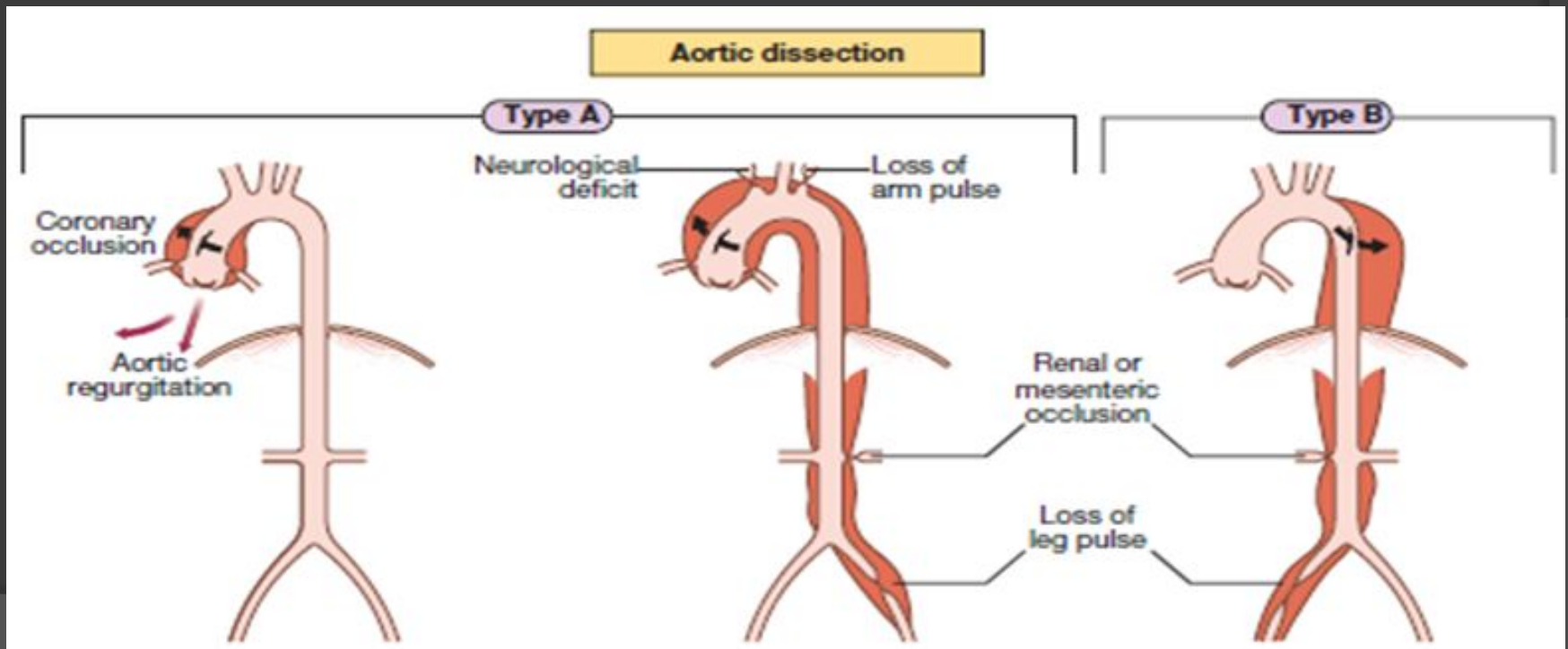
Aortic dissection

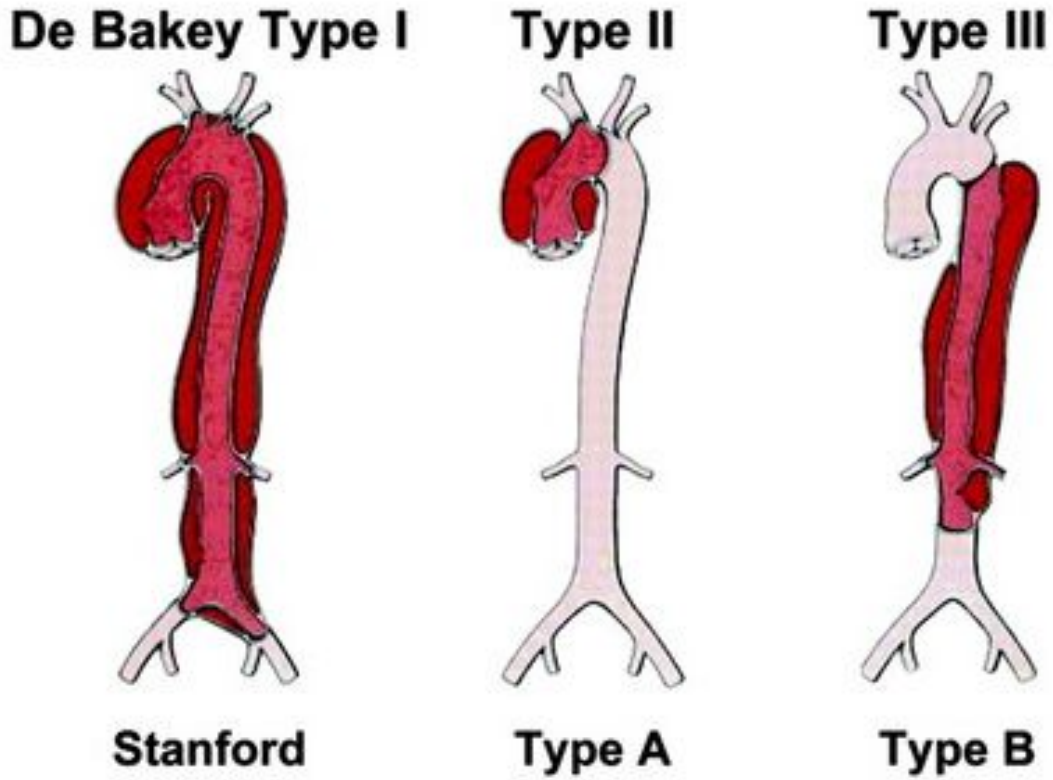
* A breach in the integrity of the aortic wall allows arterial blood to enter the media, which is then split into two layers, creating a ‘false lumen’ alongside the existing or ‘true lumen’.

* The aortic valve may be damaged and the branches of the aorta may be compromised. Typically, the false lumen eventually re-enters the true lumen, creating a double-barrelled aorta, but it may also rupture into the left pleural space or pericardium with fatal consequences.

* The primary event is often a spontaneous or iatrogenic tear in the intima of the aorta; multiple tears or entry points are common.


Stages of Aortic Dissection
Adventitia
Media
Intima
Healthy vessel
Stage 1
Rupture of Intima
Stage 2
Dissection of Media
Stage 3
Rupture of vessel


Aortic dissection
Type A
Type B
Neurological deficit
Loss of arm pulse
Coronary occlusion
Aortic regurgitation
Renal or mesenteric occlusion
Loss of leg pulse



De Bakey

Type I	Originates in the ascending aorta, propagates at least to the aortic arch and often beyond it distally.
Type II	Originates in and as confined to the ascending aorta.
Type III	Originates in the descending aorta and extends distally down the aorta or, rarely retrograde into the aortic arch and ascending aorta.

Stanford

Type A	All dissections involving the ascending aorta, regardless of the site of origin.
Type B	All dissections not involving the ascending aorta.

- Other dissections are triggered by primary haemorrhage in the media of the aorta, which then ruptures through the intima into the true lumen. This form of spontaneous bleeding from the vasa vasorum is sometimes confined to the aortic wall, when it may present as a painful intramural haematoma.

- Aortic disease and HT are the most important causes.

- Chronic dissections may lead to aneurysmal dilatation of the aorta, and thoracic aneurysms may be complicated by dissection.

- The peak incidence is in the sixth and seventh decades but dissection can occur in younger patients, usually in association with Marfan's syndrome, pregnancy or trauma; men are twice as frequently affected as women. Aortic dissection is classified anatomically and for management purposes into type A and type B, involving or sparing the ascending aorta, respectively. Type A dissections account for two thirds of cases

Factors that may predispose to aortic dissection:

1. HT (in 80%)
2. Aortic atherosclerosis
3. Non-spesific aneurysm
4. Coarctation of aorta
5. Collagen disorders (Marfan`s syndrome, Ehller-Danlos syndrome)
6. FMD
7. Previous aortic surgery (CABG, AVR)
8. Pregnancy (usually third trimester)
9. Trauma
10. Iatrogenic (Cardiac catheterization, IABP)

Clinical features:

* Involvement of the ascending aorta typically gives rise to anterior chest pain, and involvement of the descending aorta to intrascapular pain.

* The pain is typically described as ‘tearing’ and very abrupt in onset; collapse is common.

* Unless there is major haemorrhage, the patient is mostly hypertensive. There may be asymmetry of the brachial, carotid or femoral pulses and signs of AR.

* Occlusion of aortic branches may cause MI (coronary), stroke (carotid), paraplegia (spinal), mesenteric infarction with an acute abdomen (coeliac and superior mesenteric), renal failure (renal) and acute limb (usually leg) ischaemia.

Investigations

* The chest X-ray characteristically shows broadening of the upper mediastinum and distortion of the aortic ‘knuckle’, but these findings are variable and are absent in 10% of cases. A left-sided pleural effusion is common.

* The ECG may show LVH in patients with hypertension, or rarely changes same as that of inferior MI.

* Doppler echo may show AR, a dilated aortic root and, occasionally, the flap of the dissection.

* TEE is particularly helpful because transthoracic echocardiography can only provide images of the first 3–4 cm of the ascending aorta.

* CT and MRI angiography are both highly specific and sensitive.

Management

- Early mortality of acute dissection is approximately 1–5% per hour.
- Initial management comprises pain control and antihypertensive treatment.
- Type A dissections require emergency surgery to replace the ascending aorta.
- Type B dissection are treated medically unless there is actual or impending external rupture, or vital organ (gut, kidneys) or limb ischaemia.
- The aim of medical management is to maintain a mean arterial pressure 60–75 mmHg.

- First-line therapy is with BB; the additional α-blocking properties of labetalol make it with more benefit.
- Rate-limiting calcium channel blockers, such as verapamil or diltiazem, are used if BB are contraindicated.
- Sodium nitroprusside may be considered if these fail to control BP.
- Percutaneous or minimal access endoluminal repair is sometimes possible and involves either ‘fenestrating’ (perforating) the intimal flap so that blood can return from the false to the true lumen, or implanting a b graft stent.


THANK YOU