

DEFINITIONS

Aneurysm: An aneurysm is a focal permanent dilatation of an artery greater than 1.5 times the normal diameter for that particular artery. For e.g. if the normal aortic diameter in a patient is 20 mm, then any segment that is 30mm or more is aneurysmal.

Ectasia: Refers to a focal dilatation of an artery greater than the normal diameter for that artery but less than 1.5 times the normal diameter.

Arteriomegaly: Is similar to ectasia but the entire arterial segment is diffusely dilated.

Multiple aorto-iliac and infra-inguinal arterial aneurysms may occur in the setting of diffuse arteriomegaly in elderly patients. These are more difficult to treat and tend to have a poorer prognosis following surgical treatment. Multiple aneurysms may occur with intervening normal arterial segments; this is referred to as **aneurysmosis**.

An understanding of arterial aneurysms is essential considering that these aneurysms are prone to complications, some of which are life / limb threatening. These complications include:

- Rupture
- Acute or chronic thrombotic occlusion
- Acute or recurrent thrombo-embolism
- Pressure-related complications
- Spontaneous fistulisation

As a rule of thumb, aortic aneurysms are more likely to rupture, and non-aortic aneurysms are more likely to develop thrombotic or thrombo-embolic complications.

True aneurysms: The wall of the aneurysm (sac) incorporates all the conventional layers of an artery, i.e. the intima, media and adventitia.

False-aneurysms

(pseudoaneurysms): The wall of the aneurysm comprises adventitia and compressed surrounding tissue only. This pathology is generally seen in penetrating or blunt arterial injuries and at vascular anastomotic sites between vascular grafts and native arteries.

Arterial aneurysm may develop according to various pathophysiological mechanisms:

- **Degenerative aneurysms** generally evolve as a result of destructive of the media, the collagen and elastic fibres, with disordered fibrous healing resulting in weakening of the arterial wall. This classically occurs in non-specific degenerative aortic and peripheral aneurysms.
- **Infective aneurysms** may develop following various mechanisms. The organism may infiltrate the artery from an adjacent source e.g. tuberculous lymphadenitis or osteitis. Blood-borne organism (bacteraemia / viraemia) may lodge in the vasa vasorum of large arteries and generate a local inflammatory process. The infection may induce an immune response resulting in inflammatory aneurysms.
- **Connective tissue disorders** result in weakening of the arterial wall with dilatation and aneurysm formation. The defect in Marfan`s syndrome is related to the fibrillin 1 gene.

- **Trauma** results in a defect in the wall of the artery with resultant pseudo-aneurysm formation.

CLASSIFICATION OF ARTERIAL ANEURYSMS

There is no uniform classification for arterial aneurysms. They may be classified according to:

- **Anatomical location**
- **Aneurysm type: True or false (pseudoaneurysms)**
- **Morphology**
- **Size**
- **Aetiology**
- **Clinical presentation**

Classification based on anatomical location

1. Aortic aneurysms

- Abdominal aortic aneurysms (AAA)
- Thoraco-abdominal aortic aneurysms (TAAA)
- Thoracic aortic aneurysms (TAA)

2. Non-aortic aneurysms

- Peripheral aneurysms
- Renal and mesenteric aneurysms
- Carotid and subclavian aneurysms
- Other rare aneurysms e.g. ulna artery aneurysms related to the hypothenar-hammer syndrome, etc.

Morphological classification

Aneurysm may be classified according to shape:

1. Fusiform aneurysms. These aneurysms are spindle shaped.
2. Saccular aneurysms. These aneurysms are saccular outpunching's in an arterial segment. They are prone to complicate at a smaller diameter compared to fusiform aneurysms.

Classification based on size

1. Small aneurysms. The complication rate of asymptomatic small aneurysms, irrespective of location, is exceptionally low. Therefore these aneurysms, when asymptomatic, are generally observed over time rather than treated early.
2. Large aneurysms. Even when asymptomatic these aneurysms, irrespective of location, tend to complicate more frequently. These aneurysms are treated earlier as the risk of intervention is less than the risk of an aneurysm-related complication.

Classification based on aetiology

The aetiology of arterial aneurysms may vary depending on location. These causes may be classified as follows:

1. Degenerative

- Non-specific generative (the terms "atherosclerotic" and "atherosclerosis-associated" are no longer used)
- Fibromuscular dysplasia (FMD)
- Intimo-medial mucoid degeneration (IMMD)

2. Infective

- Mycotic (related to infected embolism e.g. infective endocarditis, etc)
- Tuberculous
- Syphilitic
- Bacterial (Salmonella species, Brucellosis, etc)
- Viral (HIV-related, Hepatitis B, etc)
- Fungal (rare)

3. Inflammatory / Vasculitic

- Takayasu's disease
- Giant cell arteritis
- Kawasaki disease
- Behcet's disease
- Poly-arteritis nodosa (PAN)

- Systemic lupus erythematosus (SLE)
 - Pancreatitis-related
 - “Inflammatory” non-specific degenerative abdominal aortic aneurysm
- 4. Connective tissue disorders**
- Marfan`s syndrome
 - Ehlers-Danlos syndrome
 - Cystic media necrosis (Erdheim`s disease)
 - Berry aneurysms (cerebral artery aneurysms)
- 5. Post-dissection**
- Cystic media necrosis (Erdheim`s disease)
 - Trauma (e.g. blunt traumatic aortic injuries following motor vehicle accidents)
 - Idiopathic
- 6. Post-stenotic**
- Thoracic outlet syndrome (arterial type)
 - Coarctation of the aorta
- 7. Trauma**
- 8. Congenital**
- Tuberous sclerosis
 - Turner`s syndrome
 - Menke`s syndrome
- **Infra-renal:** ~ 90% of AAAs are infra-renal
 - **Juxta-renal:** the proximal neck (distance between the renal arteries and the aneurysm sac) is less than 8mm in length.
 - **Para-renal:** one of the renal arteries comes off the aneurysm itself
 - **Supra-renal:** both the renal arteries and or the mesenteric vessels come off the aneurysm
 - **Thoraco-abdominal aortic aneurysm (TAAA):** This may involve any part of the aorta between the left subclavian artery and the aortic bifurcation. Some of these TAAAS have a AAA component as well.

Approximately 25 – 40% of patients with a AAA will have an iliac artery aneurysm as well. Generally the common iliac arteries are involved and less commonly the internal iliac arteries. It is exceptionally rare to find external iliac artery aneurysms. Approximately 12 – 15% of patients with a AAA will have concomitant descending thoracic aortic aneurysms (DTAA) as well. Approximately 3 – 5% will also have associated peripheral aneurysms, usually lower extremity.

Pathophysiology

There are many theories why non-specific AAAs develop:

- Uncontrolled hypertension
- Hypercholesterolaemia
- Smoking
- Imbalances between proteases and anti-proteases resulting in destruction of collagen and elastin in the media. These enzymes include elastase and metallo-proteinase (MMP) 2 and 9
- Infection: chlamydia pneumonia

Abdominal aortic aneurysms are further classified according to size. This is important for treatment as the risk of rupture relates to the size (diameter) of the aneurysm.

ABDOMINAL AORTIC ANEURYSMS (AAA)

Abdominal aortic aneurysms (AAA) are the most common aneurysms seen in practice. The most common pathology is non-specific degenerative. The classic patient profile is an elderly Caucasian male. The community prevalence is ~ 7 – 8% of patients older than 65 years. The ratio of males to females is 6:1. The incidence is ~ 21 per 100 000 per annum.

Abdominal aortic aneurysms may be classified anatomically as follows:

Small aneurysms: By definition small AAAs are between 4 and 5.5 cm in diameter. The risk of rupture is small (~ 1-2% per annum)

Large aneurysms: These are > 5.5 cm in diameter. The risk of rupture is high.

Aneurysm size risk of rupture

< 4 cm	0% per year
4-5 cm	0.5 to 5% per year
5-6 cm	3 to 15% per year
7-8 cm	10 to 20% per year
7-8 cm	20 to 40% per year
>8 cm	30 to 50% per year

Clinical features

Abdominal aortic aneurysms may be:

1. Asymptomatic

- Most of the AAA currently is asymptomatic and detected by routine clinical examination or abdominal imaging.

2. Symptomatic

- Vague abdominal pain
- Recent onset backache
- Vomiting (duodenal compression)
- Constipation (colonic compression)
- Flank pain (ureteric compression)
- Chronic venous disease (venous compression)

3. Complicated

- Acute lower extremity limb ischaemia (macro-embolism)
- Blue toe syndrome (micro-embolism): These patients have bluish digits and dermal staining but bounding foot pulses
- Rupture
 - Free intra-peritoneal rupture
 - Contained retro-peritoneal rupture

- Chronic contained leak
- Aorto-enteric fistula – seen with “inflammatory” non-specific AAAs
- Aorto-caval fistula – seen with “inflammatory” non-specific AAAs

Diagnosis

The diagnosis is confirmed using the following imaging modalities.

- **Abdominal duplex ultrasound (DUS).** This is a first line investigation for AAA detection. It is cheap and readily available. The results are operator-dependent and imaging may be affected by obesity and bowel gas.
- **Computed Tomography Angiography (CTA).** Multi-detector CTA is requested when ultrasound findings are equivocal or ultrasound imaging is inadequate. Multi-detector CTA is the imaging tool of choice in treatment planning for AAA when indicated.
- **Magnetic Resonance Angiography (MRA).** This is an alternative to CTA but it's availability and cost remains an issue.
- **Conventional Digital Subtraction Angiography (DSA).** This imaging modality is rarely used for diagnostic considerations currently. It's use is limited to interventional treatment of AAAs

Screening for AAA

The rationale for screening includes the following.

- The overall mortality for a ruptured AAA is ~ 90%
- Approximately 70% of patients are asymptomatic prior to rupture
- Up to 75% die before reaching
- Operative mortality ruptured AAA is ~ 50%

- Operative mortality for elective AAA repair is < 5%
- Ultrasound is cheap and available
- Studies have documented a > 50% AAA-related mortality reduction using screening programmes

However screening the entire population for AAA is not cost-effective. Therefore selective screening is recommended.

- Elderly Caucasian males > 65 years
- Elderly patients with documented peripheral aneurysms
- Patients with documented thoracic aortic aneurysms
- Family history of AAAs

Treatment

All patients need life-style modifications and medical treatment.

- Smoking cessation strategies
- Weight loss
- Anti-platelet therapy
- Lipid-lowering strategies
- B-Blockers
- Optimum blood pressure control

Surveillance programme for small AAAs

Small AAAs do not benefit from early intervention. This applies to both open repair and to endovascular repair. Consequently a surveillance programme is recommended for patients with small AAAs. A surveillance programme has been shown to be safe. Approximately 70% of patients on a surveillance programme will reach a threshold size for intervention within 5 years.

- Annual ultrasound or CTA is recommended for small AAAs
- Six monthly ultrasound or CTA is recommended for aneurysms > 4.5 cm

Indications for intervention (only in patients fit for treatment)

- All symptomatic AAAs
- All complicated AAAs
- Asymptomatic AAA > 5.5 cm in males
- Asymptomatic AAAs > 5 cm in females
- Small AAAs on surveillance with rapid enlargement (> 1 cm after 1 year on repeat scan)
- Asymptomatic AAA with a large iliac aneurysm > 3cm
- Asymptomatic saccular AAA > 3 cm (these tend to rupture at smaller diameters)

Open repair for AAA

Open repair is the standard of care for patients who are young, fit and have a predicted life expectancy > 5 years. Open repair involves a laparotomy with repair of the aneurysm using a tube or bifurcated prosthetic vascular graft. The peri-operative mortality is 3 – 5% but the morbidity may be up to 30%.

Complications include:

- Iatrogenic injuries (bowel, veins, ureter, etc)
- Haemorrhagic complications
- Wound healing complications (sepsis, dehiscence, hernias, etc)
- Acute renal dysfunction / failure
- Pulmonary complications (atelectasis/ pneumonias)
- Cardiac complications (myocardial infarction, cardiac failure, cardiac arrhythmias)
- Ischaemic colitis
- Erectile dysfunction
- Buttock / perineal ischaemia / necrosis
- Acute limb ischaemia (macro-embolism)
- Thrash foot (micro-embolism)
- Graft occlusion
- Graft sepsis

However open repair is extremely durable and generally does not need post-operative surveillance imaging. Studies have confirmed that open

repair is associated with better long-term survival and decreased re-intervention rates compared to the endovascular procedure.

Endovascular aneurysm repair (EVAR)

Endovascular aneurysm repair (EVAR) has evolved as a suitable alternative or complimentary treatment strategy to open repair. In essence the AAA is excluded internally using a large aortic covered stent (aortic stentgraft / endografts). The device is deployed using endovascular techniques. Most of these devices have multiple components which overlap during deployment. The procedure may be performed using local, regional or general anaesthesia. It avoids a laparotomy as the device is deployed via the groin vessels using a small groin incision and/or percutaneous access.

The peri-operative mortality is low (< 1%). Patient recovery is quicker. The length of hospital stay is shorter. However these patients need long-term surveillance imaging with CTA or ultrasound because of complications unique to EVAR and the associated risk of rupture (~ 1-2%/annum). These patients consequently have a higher reintervention rate than patients with open repair.

Complications of EVAR:

- **Endoleaks.** While the aneurysm sac may be excluded from the general circulation, the sac may still be perfused from various sources. This is referred to as an endoleak.
 - **Type 1 endoleak.** There is an endoleak around the proximal or distal ends of the stentgraft (inadequate seal)
 - **Type 2 endoleak.** The sac is perfused via side branches e.g. lumbar arteries.
 - **Type 3 endoleak.** The components if the stentgraft have separated resulting in sac perfusion
 - **Type 4 endoleak.** There is increased stentgraft porosity.
 - **Type 5 endoleak.** Endotension (the sac continues to enlarge but no endoleak is seen)
- Stentgraft limb kinking / occlusion
- Stentgraft migration
- Late rupture
- Stentgraft sepsis
- Access vessel related complications (pseudo-aneurysms, etc)

Laparoscopic-assisted AAA repair & Robotic-assisted AAA repairs

These procedures are performed in a few centres, are time-consuming, costly and have shown very little additional benefit.

Ruptured AAA

The overall mortality associated with ruptured AAA has remained the same over the past few decades (~ 85 – 90%). Only 40-50% of patients admitted to hospital will survive following surgery.

Patients with a ruptured AAA may not always present with the classic triad:

- Sudden onset severe backache
- Shock
- Pulsatile abdominal mass

Some may present with vague abdominal pains and a pulsatile abdominal mass only. Some patients present *in extremis* and need to be operated on immediately. Some patients present with bluish discoloration involving the scrotum, around the umbilicus (**Cullen's sign**) or involving the flanks (**Grey-Turner sign**).

Pre-treatment imaging and patient work-up

Current practice favours pre-operative CTA imaging in stable patients to guide the choice of intervention, and to guide therapeutic strategies. An emergency room ultrasound may assist with the diagnosis but will not provide adequate information for the treating vascular surgeon.

All patients should have the following evaluated on admission:

- A full blood count
- Creatinine levels
- Electrocardiogram (ECG)
- Serial blood pressure recordings
- The patients age
- The patients co-morbidities
- History of previous abdominal / vascular surgery

Treatment

A universal strategy to treat all patients with a ruptured AAA is not cost-effective and will consume desperate resources unnecessarily. Therefore predictive scoring systems have been developed to aid decision-making to favour those most likely to survive an intervention. The most widely used predictive scoring system is the **Hardman risk index** which incorporates 5 variables:

- **Age > 79 (octogenarians or older)**
- **Blood pressure persistently < 90 mmHg systolic**
- **Creatinine > 179 uMol/L**
- **Haemoglobin level < 9 g/dL**
- **Ischaemic ECG**

A patient with three or more variables will have an operative mortality of 100%.

A patient with two variables will have an operative mortality of 70% (These patients may be offered surgery)

Patients with one or less variables will benefit enormously from surgery. Note that even if a patient has no predictive variables the operative mortality is still 18%.

Thoraco-abdominal aortic aneurysms

Thoraco-abdominal aortic aneurysms (TAAA) comprise 2 – 5% of all aneurysms. These originate distal to the left subclavian artery and involves both the descending thoracic and abdominal aorta. There are five types based on the extent of disease.

Pathology

- Non-specific degenerative
- Takayasu's disease
- Intimo-medial mucoid degeneration
- HIV-related aneurysms
- Mycotic aneurysms
- Tuberculous aortitis with aneurysms

Clinical features

- High interscapular backache
- Chest and/or abdominal pains
- Dysphagia
- Dysnoea
- Stridor, hoarseness, superior vena cava syndrome (these are more common in aortic arch aneurysms)

Treatment

1. **Open surgical repair.** This generally involves a thoracotomy and a laparotomy. Patients may require limited cardiac bypass. There is a higher complication rate compared open AAA repair. Complications include:
 - Significant blood loss (a cell-saver is usually employed)
 - High operative mortality rate (~ 10% on average)
 - High paraplegia rate (~ 7% on average)
 - Acute renal dysfunction / failure
2. **Hybrid procedures.** These employ open surgical and endovascular techniques.

Benefits include avoiding a thoracotomy, less blood loss, avoids cardiac bypass and less paraplegia rates.

3. **Endovascular repairs.** Select centres are using branched stentgraft technology to treat these aneurysms in patients not suitable for open repair.

Dissecting aortic aneurysm

This develops as a complication of a chronic aortic dissection. The arterial wall dissects via an intimal tear resulting in a true lumen and a false lumen. The false gradually enlarges to become aneurysmal. Considering that most Stanford A (arising from the ascending aorta) acute aortic dissections are operated upon, most dissecting aortic aneurysms are Stanford B (arising distal to the left subclavian artery – most of these are managed medically)

Clinical features

- Features of compression (chest pain, dysnoea, dysphagia, etc)
- Rupture (overall mortality > 95%)
- Malperfusion. The dissection may extend into aortic branch vessels resulting in ischaemic complications viz. stroke, mesenteric ischaemia, renal dysfunction, lower extremity ischaemia, paraplegia.

Treatment

1. **Open surgical repair**
2. **Endovascular thoracic aneurysm repair (TEVAR)**
3. **Hybrid procedures**

Peripheral aneurysms

These are aneurysms involving the lower extremity and, less commonly, the upper extremity.

Clinical features

1. **Thrombo-embolic complications**

- Acute limb ischaemia
- Claudication
- Critical limb ischaemia (masquerading as peripheral arterial disease)
- Blue toe syndrome / Blue digit syndrome

2. **Features of compression**

- Regional / discomfort
- Limb swelling / distended veins (venous compression)
- Nerve compression

3. **Rupture.** This is exceptionally rare in peripheral aneurysms (< 5%)

POPLITEAL ANEURYSMS

These are the commonest peripheral aneurysms, excluding iliac aneurysms.

- **Approximately 70% of peripheral aneurysms are popliteal aneurysms.**
- **Approximately 50% are bilateral**
- **Approximately 50% have an associated AAA**
- **Approximately 40% – 50% will have an associated femoral aneurysm**
- **Approximately 50% will present clinically with acute limb ischaemia**

Pathology

- **Non-specific degenerative.** More than 90% are elderly caucasian males. Male to female ratio is 30:1. Age range: 60 – 80 years
- **Trauma**
- **HIV-related aneurysm**
- **Tuberculous aneurysm**

Indications for Treatment

- All symptomatic or complicated aneurysms
- All asymptomatic non-specific degenerative aneurysms > 2cm

Treatment

1. Open surgical repair.
2. Endovascular popliteal aneurysm repair (EVPAR) using peripheral covered stents.

FEMORAL ANEURYSMS

These are the second commonest peripheral aneurysms.

- **Approximately 70% are bilateral**
- **Approximately 25% are associated with a AAA**

Clinical features (similar to popliteal artery aneurysms)

Pathology

- **Pseudoaneurysms.** These are anastomotic aneurysms and are, by far, the most common femoral aneurysms.
- **Non-specific degenerative aneurysms**
- **HIV-related aneurysms**
- **Tuberculous aneurysms**
- **Mycotic aneurysms**
- **Trauma**

Indications for treatment:

- All symptomatic or complicated femoral aneurysms
- All asymptomatic non-specific degenerative aneurysms > 2.5 cm

Treatment

The only treatment is open surgical repair.

SUBCLAVIAN ARTERY ANEURYSMS

These are uncommon aneurysms.

Pathology

- **Post-stenotic.** This is related to the **thoracic outlet syndrome –arterial type (a-TOS)**. A bony anomaly, e.g. a cervical rib or ligamentous band may compress the subclavian artery producing a stenotic lesion. The turbulent flow may produce a post-stenotic dilatation of the artery. Eventually this part of the artery becomes aneurysmal with a thick wall and areas of ulceration and adherent clot.
- **Non-specific degenerative type**
- **Hiv-related**
- **Intimo-medial mucoid degeneration**
- **Tuberculous aneurysms**
- **Trauma**
- **Fibromuscular dysplasia (FMD)**

Clinical features

- Upper limb ischaemia
- Brachial plexus compression
- Limb swelling (venous compression)
- Stroke (rare complication)
- Hoarseness
- Stridor
- Pulsatile mass

Treatment

The vast majority are symptomatic and need treatment. Small asymptomatic non-specific degenerative aneurysms < 2.5 cm may be observed.

- **Open surgical repair.** This is the standard of care for treatment.
- **Endovascular repair with covered stent.** Generally reserved for trauma patients

EXTRACRANIAL CAROTID ANEURYSMS

These are uncommon aneurysms

Pathology

- **Non-specific degenerative**
- **HIV-related**
- **Tuberculous**
- **Trauma**
- **Anastomotic** - following carotid surgery

Clinical features

- Pulsatile mass
- Stroke
- Transient ischaemic attacks (TIAs)
- Hoarseness
- Stridor
- Dysphagia
- Odynophagia
- Nerve compression (e.g. hypoglossal n, facial n)
- Horner's syndrome

Treatment

- **Open surgical repair**
- **Endovascular repair.**
Generally reserved for trauma patients or to avoid a sternotomy. Not advisable for infected aneurysms

NB. All infected aneurysms, irrespective of location, should ideally have a resection of the aneurysm with extra-anatomical bypass grafting to restore circulation.

MESENTERIC ARTERY ANEURYSMS

Splanchnic artery aneurysms are uncommon aneurysms. Splenic artery aneurysms are the most common (60%), followed by hepatic artery aneurysms (20%) and superior mesenteric artery aneurysms (6%). Involvement of other splanchnic arteries are rare.

Pathology

- Non-specific degenerative aneurysms
- HIV-related
- Intimo-medial mucoid degeneration
- Connective tissue disorders, e.g. Ehlers-Danlos syndrome
- Polyarteritis nodosa
- Pregnancy-related

Clinical features

These aneurysms may be asymptomatic, symptomatic or complicate with rupture. The significance of these aneurysms relate to the high mortality rates following rupture, approaching 100% for ruptured splenic aneurysms during pregnancy.

Treatment

- **Open surgical repair.**
Generally involves a laparotomy.
- **Endovascular treatment.**
Covered stents are deployed using percutaneous endovascular techniques.
- **Transcatheter coil embolization**

RENAL ARTERY ANEURYSMS

These aneurysms are exceptionally rare. Most true renal artery aneurysms are extra-renal (90%). Approximately 75% of these are saccular.

Pathology

- Non-specific degenerative
- Fibromuscular dysplasia (FMD)
- Ehlers-Danlos syndrome
- Polyarteritis nodosa
- Trauma
- Dissection

Clinical features

- Most are asymptomatic and detected on routine angiography
- Pain
- Haematuria
- Renovascular hypertension
- Rupture. This is the most feared complication.

Treatment

- ***Open surgical repair.*** May require bench-top aneurysm resection and vascular reconstruction.
- ***Transcatheter coil embolization***
- ***Endovascular treatment using covered stents***



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