Thoracic Aortic Aneurysms

See also separate Abdominal Aortic Aneurysms, Ruptured Aortic Aneurysm and Aortic Dissection articles.

Definitions[1]

An aneurysm is a permanent and irreversible dilatation of a blood vessel by ≥50% of its normal expected diameter. Aortic aneurysms are classified as abdominal (the majority) or thoracic. The normal size of the mid-descending thoracic aorta is 26-28 mm. A 'true aneurysm' involves dilatation of all three layers of the arterial wall.

Thoracic aortic aneurysms (TAAs) may affect various parts of the thoracic aorta:

- Ascending aorta - between aortic annulus and innominate artery (these may cause aortic valve regurgitation if they are proximal enough).
- Aortic arch.
- Descending aorta - from the left subclavian artery distally.
- Thoraco-abdominal aorta - distal thoracic aorta and abdominal aorta.

Epidemiology[2]

- The true incidence and mortality rate of TAAs is not known.
- A population-based study reported an incidence of 5.9 new aneurysms per 100,000 person-years in a Midwestern community over a 30-year period with median ages of 65 years for men and 77 years for women.
- 51% of aneurysms were in the ascending aorta, 11% in the arch and 38% in the descending thoracic aorta. Prevalence may be at least 3-4 % of those aged >65 years and is probably increasing.[3,4]

Aetiology

Pathogenesis[3,4]
The pathogenesis of TAA is complex and probably involves inflammation, proteolysis and reduced survival of smooth muscle cells in the aortic wall. There is probably an imbalance of proteolytic enzymes versus their inhibitors. Once the aorta reaches a critical diameter (about 6 cm in the ascending aorta and 7 cm in the descending aorta), it loses all distensibility so that a rise in blood pressure to around 200 mm Hg (as can occur physiologically during stress or exertion) can exceed the arterial wall strength and may trigger dissection or rupture.

Causes[3]

- Genetic - there is a strong genetic component.[4] In some families, there appears to be an autosomal dominant trait causing TAA.[5,6]
- Certain connective tissue disorders:
  - Marfan's syndrome.
  - Ehlers-Danlos syndrome type IV.
  - Loeys-Dietz syndrome (which partly resembles Marfan's syndrome).[7]
- Infections:
  - Tertiary syphilis
  - Mycotic aneurysm[8]
  - HIV[9]
Aortitis - eg, from giant cell arteritis, rheumatoid arthritis, Behçet's disease, Takayasu's arteritis or retroperitoneal fibrosis.

Aortic dissection may be a cause (or a result) of TAA.

Trauma.

Weightlifting, cocaine and amfetamine use may be involved in causing TAA and/or aortic dissection - perhaps because of the large rises in blood pressure which occur during these activities.

Risk factors[^3, 10, 11]

These factors probably result in an increased rate of growth of aneurysms:[^3]

- Hypertension.
- Increasing age.
- Smoking.
- Bicuspid or unicuspid aortic valves.
- Atherosclerosis.
- Chronic obstructive pulmonary disease (COPD).
- Chronic kidney disease.
- Previous aortic aneurysm repair.
- Turner syndrome - may be a risk factor.

Presentation[^2, 4]

Most TAAs are asymptomatic (about 95%). They may only be diagnosed incidentally, or if complicated by dissection, rupture, or other complications. Possible presentations are:

- Pain - located in the chest, neck, upper back, mid-back or epigastrium - the site may relate to which section of aorta is involved:
  - Acute or increasing pain suggests expansion, with the risk of impending rupture or dissection.
  - Acute aortic dissection presents with sudden onset, very severe, tearing or sharp pain which is maximal at the time of onset (in contrast to the more dull, crescendo pain of myocardial infarction).[^4]

- Aortic regurgitation - if a proximal TAA involves the aortic valve.
- Systemic symptoms (eg, fever) - if there is an infective cause.
- Thrombo-embolic presentations:
  - Arterial emboli.
  - Disseminated intravascular coagulation - eg, bruising, petechiae.

- Symptoms due to compression of local structures:
  - Hoarseness (recurrent laryngeal nerve).
  - Cough, stridor, or dyspnoea.
  - Superior vena cava obstruction.
  - Dysphagia.
  - Back pain (vertebral body erosion).


This may present with:

- Severe, sharp pain in the chest, neck or back. The pain may migrate as the dissection progresses.
- Ischaemia in the territory of any affected artery - eg, myocardial infarction, neurological features, visceral or limb ischaemia.
- Unequal upper limb pulses.
- The dissection may progress to rupture.
Ruptured TAA
This may present with:

- Acute pain - in the chest, neck or back.
- Collapse, shock or sudden death.
- If the aneurysm erodes into a nearby structure, there may be:
  - Haematemesis (aorto-oesophageal fistula).
  - Haemoptysis (aorto-bronchial fistula).
  - Haemothorax.
  - Cardiac tamponade (ruptures into the pericardium).
  - An acute left to right shunt (aortopulmonary artery fistula).[12]

Differential diagnosis
This is wide, and TAAs are easily missed or mis-diagnosed. Differential diagnoses include other causes of chest pain, thoracic back pain, arterial ischaemia or collapse.

With acute chest pain, myocardial infarction[4] and pulmonary embolus[13] are important to differentiate, as these require thrombolysis or anticoagulation, which is contra-indicated for TAA.

Investigations

Urgent scenario
For an acutely ill patient with suspected thoracic aortic rupture or dissection, the most relevant investigations are:

- Blood tests - FBC, clotting screen, renal and liver function and cross-match.
- ECG.
- CT scan with contrast.
- Magnetic resonance angiography will also provide good images but probably only for stable patients.[3]

Non-acute scenario
Investigations aim to evaluate the detailed anatomy of the aneurysm, any treatable causes and the patient’s fitness for surgery:

- Blood tests:
  - FBC, clotting screen, renal function.
  - ESR and/or CRP if an inflammatory cause is suspected.
  - LFTs and amylase, if embolisation or dissection is suspected
  - Investigate for a suspected cause, if relevant - eg, syphilis or HIV serology, connective tissue disease.

- ECG.
- Lung function tests.
- Ultrasound:
  - Transthoracic echocardiogram shows aortic valve and aortic root.
  - Transoesophageal echocardiogram shows from the aortic valve to the proximal descending aorta.
  - Abdominal ultrasound - to look for associated abdominal aneurysms.

- Scans:
  - CT scan with contrast medium is the most widely used diagnostic tool. This defines the precise anatomy and can show the aneurysm, dissection, thrombus or haematoma. Detailed views are needed for endovascular repair planning.
  - CT angiography is possible and non-invasive, although the contrast material is nephrotoxic.
  - Magnetic resonance imaging (MRI) also shows the anatomy well; the advantage is that magnetic resonance angiography is not nephrotoxic.
Management

See also separate Ruptured Aortic Aneurysm and Aortic Dissection articles.

Who needs surgery?[4]

- Immediate/urgent surgery is needed for:
  - Ruptured TAA.
  - Some types of aortic dissection.
  - Acute symptoms (because these suggest expansion and impending rupture/dissection).

- Symptomatic TAAs merit surgery regardless of their size (because there is a higher risk that they will rupture or dissect).
- Asymptomatic TAAs are assessed to evaluate the relative risks/benefits of surgery. The risk of rupture depends on:
  - Aneurysm diameter. This is the most important factor predicting rupture. Generally, aneurysms of diameter >5.5 cm (ascending aorta) or >6 cm (descending aorta) merit repair.
  - Using body surface area as well as aneurysm diameter gives a more accurate risk profile.
  - Patients with Marfan’s syndrome or a strong family history of TAA merit surgery earlier (at a smaller aneurysm diameter).
  - The rate of expansion is also important.

Medical management

- Regular monitoring of the aneurysm - eg, by CT or MRI scans every six months.
- Rigorous blood pressure control with use of beta-blockers to reduce shear stress across the aortic wall.
- Smoking cessation.
- Treating the underlying cause where feasible - eg, infection.
- Treating other cardiovascular risk factors - eg, hyperlipidaemia.

Driving

There are Driver and Vehicle Licensing Agency (DVLA) restrictions for aneurysms above a certain size, and requirements for notification.[14]

Surgical repair

- This involves thoracotomy to replace the diseased aorta with a Dacron® graft.
- Replacement of the aortic root ± the aortic valve may also be necessary.
- There are significant perioperative mortality and complication rates (see ‘Prognosis’, below).

Endovascular repair[1]

Endovascular repair is also known as ‘endovascular stenting’, EVAR (= endovascular aneurysm/aortic repair) or TEVAR (= thoracic endovascular aneurysm/aortic repair). It is a minimally invasive technique which is relatively new. It delivers a stent via a remote vessel.

- Reports from non-randomised studies suggest that endovascular repair may improve early outcomes, with reduced mortality and paraplegia rates.
- However, TEVAR can have additional complications, ie endoleaks, stent fractures, stent graft migration, iliac artery rupture, retrograde dissection and aorto-oesophageal fistula. These may require re-intervention.
- Patients receiving stents require frequent follow-up with CT scans.
Prognosis

Natural history:

- The size of the TAA is the major factor predicting rupture.
- The natural course is for the aneurysm to increase in size slowly.
- The five-year mortality rate of TAAs >6 cm is 38-64%.
- Rupture is the most common cause of death in patients not receiving surgery.
- When rupture occurs, patients mostly die within six hours. Although about half arrive alive at the hospital, the overall mortality from ruptured TAAs is up to 97%.

With elective surgical repair:

- Perioperative 30-day mortality is 4.8% with open surgery.
- Possible complications particular to aortic repair are:
  - Paraplegia due to spinal cord ischaemia - with descending aorta repairs.
  - Stroke.
  - Acute kidney injury.
- A more detailed review of outcomes after surgery is available.

Prevention and screening

- For athletes - one expert advises routine screening of all athletes involved in weightlifting or heavy athletics. [4]
- For patients with Marfan's syndrome or Ehlers-Danlos syndrome - lifelong beta-blockers, moderate restriction of physical activity and regular imaging of the aorta.
- For patients with a bicuspid aortic valve, assessment of the ascending aorta may be advisable. [15]

Further reading & references

- Aortic Aneurysm, Familial Thoracic 1, AAT1; Online Mendelian Inheritance in Man (OMIM)
- Aortic Aneurysm, Familial Thoracic 2, AAT2; Online Mendelian Inheritance in Man (OMIM)
- Loeye-Dietz Syndrome, Type 1A, LDS1A; Online Mendelian Inheritance in Man (OMIM)
- Assessing fitness to drive: guide for medical professionals; Driver and Vehicle Licensing Agency