Abdominal Aortic Aneurysm (AAA) — Risk Factors and Prognosis

Abdominal Aortic aneurysm results from loss of strength and thinning of the aortic wall. It often implies the threat of rupture of aneurysm demanding of emergency interventions. Being asymptomatic, delayed discovery of AAA is a common phenomenon. The exact etiology of AAA is not known. Males develop AAA more commonly than females. The risk factors of AAA involves hyperlipidemia, hypertension, heart disease, smoking and family history. With the proper evolution of the case, elective surgical repair can be opted as definite cure.

Definition and Background of Aortic Aneurysm

An aortic aneurysm is a permanent and irreversible widening of the aorta to more than 1.5 times its normal diameter. It is generally asymptomatic and undiagnosed until it ruptures. Aneurysms are frequently located in the abdomen but can occur anywhere along the aorta, including the thoracic aorta. Ruptured AAAs are associated with high mortality. Patients aged above 65 years are at the greatest risk. Other risk factors include multiple cardiovascular and genetic factors or pre-existing peripheral vascular diseases.
Epidemiology of Aortic Aneurysm

The incidence of asymptomatic aortic aneurysms ranges between 5% and 10% and are more common in males than in females. Most cases occur in men who are aged above 50 years and in women aged above 60. A sharp increase in the number of cases has been reported within this age bracket.

AAAs are more common in white Caucasians than in those of African American, Asian, and Hispanic ethnicities. Ruptured abdominal aortic aneurysm causes 15,000 deaths a year in the United States alone, making it the 13th most common cause of death. In 2013, 15,200 deaths were attributed to aortic aneurysms, an increase from 10,000 in 1990.

Etiology of Aortic Aneurysm

Aortic aneurysms are triggered by a degradation of the aortic walls, which weakens them. The etiological factors for aortic aneurysm include:

- Atherosclerosis
- Genetic disease affecting the structure and function of connective tissues/proteins (e.g., collagen and elastin) in the walls of the aorta — Marfan’s
syndrome, Ehler-Danlos, Collagen vascular-diseases (more likely to cause thoracic aortic aneurysms)
- Infection/mycotic aneurysms due to the local invasion of the intima and media by infective pathogens (most commonly gram-positive organisms: Escherichia coli, Salmonella) leading to abscess formation and dilatation.
- Trauma
- Aortitis (usually syphilitic — TAA > AAA)
- Hypertension
- Alcohol abuse

Risk Factors

Modifiable Risk Factors

- Cardiovascular and peripheral vascular disease
- Smoking
- Hypercholesterolaemia
- Hypertension

Unmodifiable Risk Factors

- Age: > 55 years
- Sex: male
- Genetic Conditions (e.g., Marfan’s, Ehlers-Danlos syndromes)
- Family history
- Atherosclerotic disease can be genetically more susceptible

Classification of Aortic Aneurysm

Aneurysms are classified as follows:

<table>
<thead>
<tr>
<th>Classification</th>
<th>Description</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>True aneurysm</td>
<td>Involves all 3 layers of the vessel wall — intima, media, and adventitia.</td>
<td>Atherosclerosis, infection (syphilitic), ventricular and congenital</td>
</tr>
<tr>
<td>False aneurysm (pseudoaneurysm)</td>
<td>Blood that has leaked out of artery/vein but is held around the vessel by the surrounding tissue — this cavity can clot or rupture eventually</td>
<td>Trauma, surgery (percutaneous), artery injection</td>
</tr>
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</table>

Aortic aneurysm classification based on the aneurysm location:

<table>
<thead>
<tr>
<th>Aortic root aneurysm</th>
<th>Rare</th>
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<tbody>
<tr>
<td>Thoracic aortic aneurysms (TAA)</td>
<td>Ascending, aortic arch and descending</td>
</tr>
<tr>
<td>Abdominal aortic aneurysms (AAA/Triple-A)</td>
<td>Most common</td>
</tr>
</tbody>
</table>

Morphology

Fusiform

- Spindle-shaped, with variable length and diameter, and affects the entire circumference of the vessel
- Most commonly affecting aorta and iliac arteries rarely

Saccular

- Spherical and affecting the vessel partially
Pathophysiology of Aortic Aneurysm

Aortic aneurysms are **caused by the degradation of the aortic wall**, which results in a permanent and irreversible dilatation. The degradation consists of **loss of elastic lamellae and smooth muscle cells, increased proteolysis, and leucocytic infiltration**. The dilatation is considered as an aneurysm once the diameter reaches 1.5 times the diameter of the normal aorta (abdominal → 3 cm/thoracic → 4.5 cm).

**Abdominal aortic aneurysm (AAA)**

AAA is the most common type of aortic aneurysm, with 90% of the cases occurring below the **L1-L2 vertebra**. The location correlates with the level of renal arteries (infrarenal). AAAs are more common than TAAs because the abdominal aortic walls contain less **elastin**.

Elastin is a protein, which enables vascular elasticity, and repeated expansion and contraction of a vessel. The thoracic aorta is usually under higher pressure as it is closer to the heart, and therefore carries higher levels of this load-bearing protein.

A lower level of elastin increases the strain on the abdominal aorta. Diseases of the abdominal aorta decrease the time taken to degrade all the elastin present in the AAA compared with the elastin levels in the TAA.

**Proteolytic degradation of tunica media** is the main mechanism underlying AAA development. Increased **metalloproteinase** activity leads to increased destruction of the media.

The media contains the muscular, structural and elastic tissues, which contribute to the strength of the vessel walls. The abdominal aorta also contains fewer vasa vasorum compared with the TAA. Therefore, as the cells within the abdominal aortic wall rely on diffusion, it **increases the risk of damage, especially in atherosclerotic vessels**.
Thoracic aortic aneurysm (TAA)

TAAs are aneurysms that are located in the thoracic aorta, above the diaphragm. Undetected TAAs can be very fatal, as rupture can trigger death due to their close proximity to the heart. They are rarer than AAAs.

TAAs are more likely to be syphilitic, genetic, congenital or traumatic in origin. Descending aortic aneurysms are primarily atherosclerotic, whereas those of the aortic arch is generally attributed to atherosclerosis or dissection.

Hypertension

If the patient is hypertensive, this can lead to a rapid progression of the disease as the weakened walls of the aorta are under more strain.

Atherosclerosis

Atherosclerosis occurs when the atherosclerotic plaque builds up within the aorta and reduces blood diffusion to the media of the aortic wall by increasing the diffusion distance. Atherosclerosis induces atrophy of the media due to reduced blood supply. The decrease in size and the strengthening of this layer leads to larger and irreversible dilatation because the media includes smooth muscle cells and elastic tissue, which usually absorb the blood pressure generated by the heart.

Genetic

Marfan’s syndrome is a connective tissue disorder, which is characterized by the misfolding of fibrillin-1. Fibrillin-1 is a protein that forms elastic tissue and plays a role in signaling. One such role includes binding to TGF-beta. In the case of Marfan’s syndrome, mutant fibrillin-1 fails to bind TGF-beta and induces accumulation of TGF-beta in various tissues including the aorta. This results in the formation of weakened tissue with abnormal structure and function.

Ehlers-Danlos syndrome is a genetic condition causing incorrect production and processing of collagen, an essential protein involved in tissue structure, which can lead to weakened vessel walls that quickly become aneurysmal.

Pathology of Aortic Aneurysm

The changes within the aorta wall include:

- Foam cells, cholesterol, ulceration, thrombosis, and ruptures in the tunica media and intima, consistent with atherosclerosis
- Inflammatory infiltration of the adventitia
- Loss of smooth muscle cells and elastic lamellae

Further evidence of atherosclerosis includes abnormal connective tissue structure in genetic conditions, infection and/or inflammation.

Symptoms of Aortic Aneurysm

Aortic aneurysm is usually asymptomatic if not ruptured.
Most aneurysms are asymptomatic until rupture occurs and are usually detected in routine health checks or incidentally during radiological investigations.

**Eventual enlargement of an aneurysm can lead to:**
- flank/back pain
- abdominal pain
- pulsatile mass, usually in the abdomen, which increases in size over months and years.

An aneurysm can compress the surrounding structures such as organs and nerves depending on its size and location.

**Vocal hoarseness**
- The arch of the aorta aneurysms can irritate the left recurrent laryngeal nerve (branch of the vagus, which goes underneath the aortic arch before turning back upwards to supply blood to the laryngeal muscles involved in voice generation).

**Urinary symptoms**
- Ureteral compression

**Thromboses/emboli**
- As a result of pressure on surrounding vessels

**Emboli symptoms**
- The plaque that usually lines atherosclerotic aortic walls can break off and as a result block a downstream vessel.

**Symptoms of systemic disease(s)**
- Patients may have established disease(s) related to an aortic aneurysm and could, therefore, manifest symptoms of peripheral vascular disease, infection, trauma, Marfan’s syndrome or Ehler-Danlos syndrome.

**Signs**

**Inspection**, which reveals comfort at rest and visible abdominal pulsation

**Auscultation** indicating bruits in the central abdominal region

**Manual palpation on either side of aorta** indicating pulsatile and expansive abdominal masses; and expansion of the mass when the palpation is outwards along the borders of the aorta

**Signs of peripheral vascular disease**
- Arterial insufficiency in the lower limbs, e.g., arterial ulcers, weak pulses, pallor, cold, loss of hair, gangrene, loss of sensation (paraesthesia), and paralysis
- Patient in shock; cold, clammy, pale, tachycardia, tachypnoea
- Abnormal carotid, brachial and femoral pulses
- Anxiety and sense of impending doom
- Marfan’s syndrome-related signs (for e.g., high arched palate)
- **Ehlers-Danlos syndrome**-related signs
- Infection-related signs (for e.g., fever)
- Trauma signs (for e.g., knife wound)

**Complications**

**Ruptured aortic aneurysm** is a major complication, and is especially dangerous when the diameter of an aneurysm is greater than 5 cm. It is a severe life-threatening emergency and the patient usually presents with shock if they survive the journey to the emergency department:

**Patient presents with a classic triad:**

- Flank pain – severe tearing pain that usually radiates towards the back
- Hypotension (from blood loss)
- Pulsatile abdominal mass

**Patient can manifest:**

- Tachycardia
- Altered mental status
- Flank bruising if there is retroperitoneal bleeding.
- Skin with a blue complexion
- Loss of consciousness
- Hypotension and shock (hypovolemic) – eventually death by **exsanguination** (blood loss)
- Permanent disability from a stroke (**CVA**)
- Global **ischemia** – e.g., mesenteric, bowel, renal, spinal cord, visceral ischemia/infarction
- Compression – esophagus, left recurrent laryngeal nerve (hoarseness and vocal cord paralysis).

**Diagnosis of Aortic Aneurysm**

Rapid and accurate diagnosis of AAA is needed using the following procedures:

- **Physical examination** via auscultation revealing pulsatile and expansile abdominal mass with bruits; failure to detect this is **not** enough to rule out AAA.
- **Anamnesis**: In stable patients, a detailed history should be taken to assess risk factors, risk of rupture, symptom duration, and onset and any family history or genetic conditions.
- **Imaging** is used to diagnose and reveal the extent of the disease

**Imaging methods**

The following imaging methods are used to detect aortic aneurysms:

**Ultrasound**

- Gold standard diagnostic test for an aortic aneurysm
- Used to monitor aortic aneurysms over time if the patients do not require immediate surgery.
- Quickest and easiest option
- It can be done at the bedside
- Reveals size and extent of disease

**CT > (MRI) Scan**

- Used after ultrasound diagnosis to obtain more insight into size and anatomy for monitoring purposes and to assist surgeons
- CT scan has 99% sensitivity
- CT scan uses contrast dye to measure blood flow through the aorta

**X-Ray Scan**

- Abdominal/kidney ureter and bladder (may be discovered after KUBXR for urinary symptoms caused by ureteral compression)
- Reveals calcification of the aorta
Aortic angiography

- Catheterization of the aorta (via femoral artery) and injection of contrast whilst X-rays of the aorta are taken are useful in determining the luminal size and examining any aortic dissections that may be present.

**Blood tests** - FBC, U+E, ESR/CRP, Clotting screen, lipid profile

**Blood pressure tests**

**Differential Diagnoses**

<table>
<thead>
<tr>
<th>Kidney</th>
<th>Pyelonephritis, nephrolithiasis</th>
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<tbody>
<tr>
<td><strong>GI system</strong></td>
<td>Acute gastritis, perforated peptic ulcer, bleeding, ischemic bowel, large bowel obstruction, pancreatitis, gallstones, diverticulitis, IBS, IBD, appendicitis</td>
</tr>
<tr>
<td><strong>Heart</strong></td>
<td>Myocardial infarction, aortic dissection</td>
</tr>
<tr>
<td><strong>Gynecological</strong></td>
<td>Cystitis, ruptured ovarian cyst, ovarian torsion</td>
</tr>
</tbody>
</table>

**Treatment of Aortic Aneurysm**

**AAA**

<table>
<thead>
<tr>
<th>Conservative</th>
<th>Surgical Repair</th>
<th>Medication</th>
</tr>
</thead>
</table>
| • Surveillance usually by ultrasound to monitor the progression  
• Reduction of modifiable risk factors | • Open aneurysm repair (OR)  
• Endovascular aneurysm repair (EVAR)  
• Immediate surgery for rupture | • To monitor and treat risk factors  
• Lipids (statins)  
• Strict blood pressure measurement (antihypertensives) |

**TAA**

TAA’s are defined as aneurysmal after reaching a diameter > 4.5 cm. However, TAA with a diameter > 6 cm are indicated for treatment and repair via endovascular or open surgery.
Prognosis

The larger an aneurysm, the more likely it is to rupture. However, it is important to note that smaller diameter aneurysms also rupture. The following table presents the results of a 2003 study by the Society for Vascular Surgery, which shows the relationship between the diameters of AAA’s and the annual risk of rupture.

<table>
<thead>
<tr>
<th>AAA Size (cm)</th>
<th>Annual Rupture Risk (%)</th>
</tr>
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<tbody>
<tr>
<td>3—3.9</td>
<td>0</td>
</tr>
<tr>
<td>4—4.9</td>
<td>0.5—5</td>
</tr>
<tr>
<td>5—5.9</td>
<td>3—15</td>
</tr>
<tr>
<td>6—6.9</td>
<td>10—20</td>
</tr>
<tr>
<td>7 &gt;</td>
<td>20—50</td>
</tr>
</tbody>
</table>

Ruptured AAAs have a significantly worse prognosis compared with unruptured AAAs. The postoperative mortality of a ruptured AAA is still > 40% (Brown et al, 2002), compared with 1–6% if the AAA is repaired before rupture (Greenhalgh et al, 2004).

Screening

Screening programs for the detection of aortic aneurysms exist worldwide. The main criteria usually involve men over the age of 65 years who may be at higher risk, due to risk factors such as a history of smoking. It is not certain whether screening for women is useful.

Increased screening is recommended for those with family history and/or genetic diseases with known links to aortic aneurysms. Early detection is key to reducing the high mortality rate of aortic aneurysms.

Prevention

The risk of aortic aneurysms can be reduced via close monitoring of cholesterol and blood pressure. Reduction of modifiable risk factors also decreases the chances of worsening or developing aortic aneurysms – e.g., stop smoking, reduce alcohol intake and eat a healthy diet.

Regular health checks can help detect any aortic aneurysm early and prevent progression and rupture.

<table>
<thead>
<tr>
<th>Eligible population</th>
<th>Test</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Men aged between 65 and 75 years who have a smoking history &gt; 100 cigarettes per year, with or without a family history of AAA</td>
<td>Ultrasound of abdominal aorta</td>
<td>One time</td>
</tr>
<tr>
<td>Men aged between 55 and 75 years who have a family history of AAA</td>
<td>Ultrasound of abdominal aorta</td>
<td>One time</td>
</tr>
<tr>
<td>Women aged between 55 and 75 years who both have a smoking history &gt; 100 cigarettes and a family history of AAA</td>
<td>Ultrasound of abdominal aorta</td>
<td>Consider one time</td>
</tr>
<tr>
<td>Women of any age who have neither a smoking history nor a family history of AAA</td>
<td>Screening not recommended</td>
<td>N/A</td>
</tr>
</tbody>
</table>
References


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