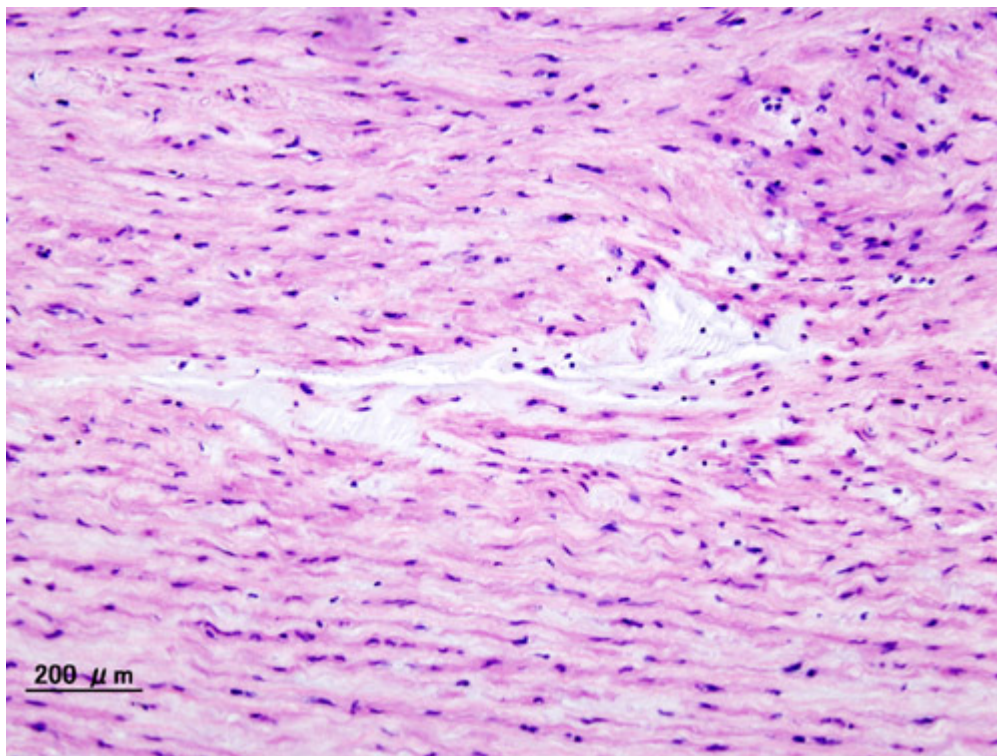


# Aortic Dissection — Classification and Diagnosis

[See online here](#)

**Aortic dissection is a rare but serious medical condition. It can be suddenly fatal and needs to be detected early. 40 % of people do not survive once the aorta dissects and therefore despite its rare nature, patients with related symptoms should be thoroughly investigated.**

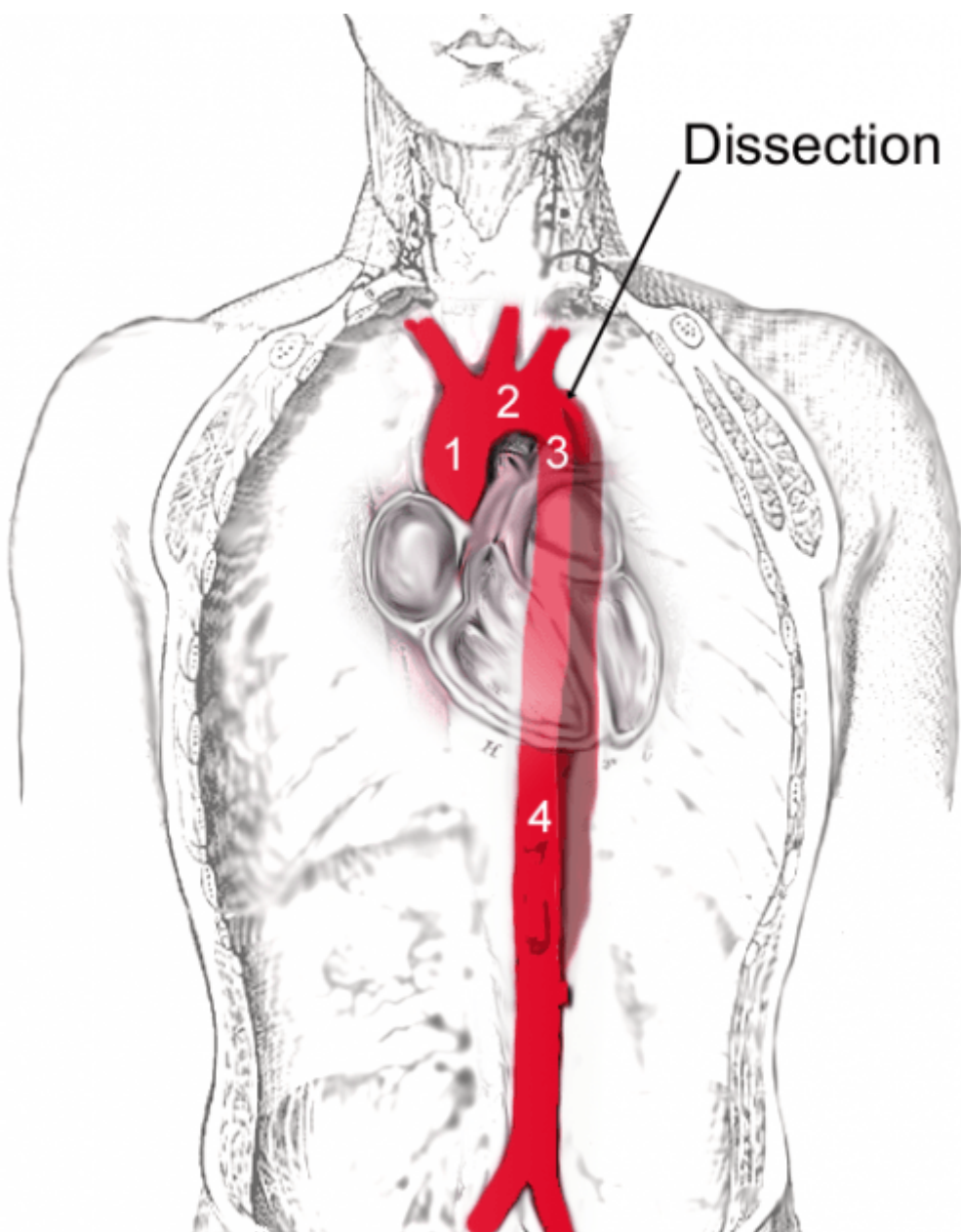


## Definition and Background of Aortic Dissection

### What is an aortic dissection?

Aortic dissection occurs when the inner coat (**tunica intima**) of the aortic wall develops a fissure that causes blood to enter the media layer of the wall to form a channel of [blood](#). The pressure causes the layers to be forced apart and causes severe pain, characteristically known as a tearing pain. This is a serious medical emergency as it can partially occlude branches of the aorta and reduce blood flow to the rest of the body causing vital organs to be starved of blood and nutrients. If severe, the aorta can rupture and cause rapid death.

There are risk factors associated with aortic dissection, such as **hypertension**, and there are also [genetic diseases](#) that can markedly increase the chances of occurrence.



**Image:** "Dissection of the descending part of the aorta (3), which starts from the left subclavian artery and extends to the abdominal aorta (4). The ascending aorta (1) and aortic arch (2) are not involved." by JHeuser.  
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## Epidemiology of Aortic Dissection

### Who is at risk of aortic dissection?

Aortic dissection is a rare condition which occurs at a rate of 2—3.5 per 100,000 people every year. It is more common in men than women as 65 % of all dissections occur in males. There is a link with pregnant females, as this accounts for half of all dissections in women under 40 (rare). Peak incidence is around 50 - 65 years of age and nearly all occur between 40 - 70 years.

Dissections that occur around 30-40 years of age are usually associated with genetic and/or connective tissue diseases.

Men aged 40 to 60 with hypertension make up 90 % of cases. It is more common in

Afro—Caribbean people than Caucasians, as is hypertension. Asians have the lowest occurrence.

## Etiology of Aortic Dissection

### Causes of aortic dissection

- Hypertension
- Genetic disease/connective tissue abnormalities that affect the aorta; affects the structure and function of connective tissue/proteins (e.g. collagen and elastin) in the walls of the aorta — **Marfan syndrome** (more likely to be proximal dissections), **Ehler—Danlos syndrome**
- **Turner syndrome** (causes aortic root dilatation)
- Pregnancy
- Blunt chest trauma (e.g car accidents, though they are deceleration injuries that more commonly cause an aortic transection, that is a laceration of all three layers of the aorta) or iatrogenic trauma (during catheterization or intra-aortic balloon pump counterpulsation)
- **Atherosclerosis**
- Bicuspid aortic valve (congenital) — increases the chance of ascending aortic dissection
- **Syphilis** (tertiary stage)
- Cardiac surgery — especially aortic valve replacement as aortic regurgitation can cause dilatation and aortic wall weakening.

### Risk Factors

Modifiable Risk Factors	Unmodifiable Risk Factors
Hypertension (can be unmodifiable if genetic or secondary)	Age
Atherosclerosis — lipid levels	Sex — Male
Iatrogenesis	Genetic Conditions (e.g. Marfan, Ehlers—Danlos)
	<b>Family History</b>
	Pregnancy
	Bicuspid aortic valve
	Coarctation of the aorta
	Arteritis and syphilis-induced swelling of vessels

## Classification of Aortic Dissection

### Stanford and DeBakey systems




There are several systems of classification for aortic dissection based on anatomy or duration of onset of symptoms.

The **Stanford system** is the most commonly used one:

<b>Type A</b> 70 - 75 %	<b>Ascending aorta</b> +/- aortic arch, possibly descending aorta. <b>Can involve the aortic valve</b>	Tear can originate in any 3 (rarely originates in the descending aorta) <b>Includes DeBakey 1 + 2.</b> <b>Requires primary surgical treatment</b>
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<p><b>Type B</b> 25 - 30 %</p>	<p>Descending aorta or the aorta (distal to the left subclavian artery) <b>without the involvement of the ascending aorta</b></p>	<p><b>Includes DeBakey 3.</b> Mortality is not improved by surgery unless it is complicated — therefore <b>the primary treatment is medical (control blood pressure)</b></p>
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In contrast, the **DeBakey system** is based on anatomy:

<p><b>Type 1</b> 60 %</p>	<p>Origin — ascending aorta, extends to the aortic arch and often beyond. <b>Most lethal</b> and often seen in patients &lt;65.</p>	 <p><small>Image: "DeBakey Type 1" by JHeuser. License: CC BY-SA 3.0</small></p>
<p><b>Type 2</b> 10 - 15 %</p>	<p>Origin — ascending aorta and is confined here</p>	 <p><small>Image: "DeBakey Type 2" by JHeuser. License: CC BY-SA 3.0</small></p>
<p><b>Type 3</b> 25 - 30 %</p>	<p>Origin — descending aorta — rarely goes proximally but commonly goes distally. Elderly with hypertension and atherosclerosis.</p>	 <p><small>Image: "DeBakey Type 3" by JHeuser. License: CC BY-SA 3.0</small></p>

## Pathophysiology of Aortic Dissection

Conditions observed with aortic dissection

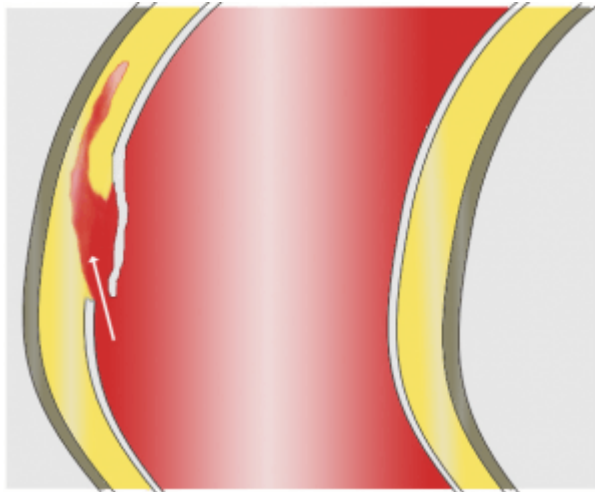


Image: "scheme of aortic dissection" by JHeuser. License: [CC BY—SA 3.0](#)

In aortic dissection, blood enters the intima and the media layers. The high pressure of blood tears the media apart in a laminated plane. The plane is usually between the inner 2/3rd and the outer 1/3rd. The dissection can extend proximally and/or distally for variable distances.

Most dissections originate in the ascending aorta, usually within 10 cm of the aortic valve. These tears are commonly 1 - 5 cm long and are transverse or oblique in orientation, with rough edges.

- **Antegrade dissection** — spreads towards the iliac bifurcation and sometimes all the way down to the iliac and femoral arteries.
- **Retrograde dissection** — spreads towards the aortic root and heart.

Sometimes, the dissection can spread through the intima, media, and adventitia causing an external rupture. This, of course, causes huge internal bleeding or **cardiac tamponade** if the dissection extends through the adventitia but into the pericardial sac, forming a hemopericardium. Both scenarios are life-threatening and can lead rapidly to death.

It is possible for the dissection to enter the aortic walls and then recanalize the lumen of the aorta through another second distal intimal tear. This condition is less serious for patients as there is a pathway for the blood to leave preventing a serious build-up of pressure. Eventually, this can lead to a chronic dissection and the channel can become covered in endothelium leading to a **double-barrel aorta** (there are now two vascular channels within the aorta). Retroperitoneal and pericardial ruptures are possible.

When the blood enters the intima and tears through the media, it creates a **false lumen**. The **true lumen** is the natural physiological lumen of the vessel. In between both of these lumens is a layer of intima which is known as the **intimal flap**. As stated above, the false lumen may recanalize into the true lumen.

There are different types of aortic dissection as discussed above, but generally, 65 % originate in the **ascending aorta**, 10 % in the **aortic arch** and 20 % occur in the **descending thoracic aorta** (distal to the **ligamentum arteriosum**).

The reason the intimal tear occurs is unknown. It can occur as a result of intimal ischemia from increased shear forces due to hypertension, or due to genetic connective tissue diseases where there is a paucity of healthy elastic tissue such as **Marfan syndrome**. In Marfan, the key issue is that the collagen and elastin within the media are degenerative,

unstructured and dysfunctional — causing **cystic medial necrosis** (as discussed below).

In approximately 10 % of cases, there is no evidence of an intimal tear. These dissections may be caused by bleeding within the medial layer of the vessel causing a **secondary aortic dissection** — this is still treated in the same way.

## Genetic disease implications

Marfan syndrome is a connective tissue disorder which involves the misfolding of **fibrillin—1**. This is a protein that forms elastic tissue and has roles in signaling. One such role includes binding to **TGF—beta**; the mutated fibrillin—1 fails to do this and this causes an accumulation of TGF—beta in various tissues including the aorta, causing weakened tissue with an abnormal structure and function.

**Ehlers—Danlos syndrome** is where a genetic problem causing incorrect production and processing of **collagen**, an essential protein involved in the structure of tissues. This can lead to weakened [vessel walls](#) which can quickly become [aneurysmal](#).

## Pathology of Aortic Dissection

### Lesions associated with aortic dissection



**Image:** "Histopathological image of dissecting aneurysm of the thoracic aorta in a patient without evidence of Marfan trait. The damaged aorta was surgically removed and replaced by an artificial vessel. Victoria blue & HE stain." by KGH. License: [CC BY—SA 3.0](#)

The most commonly identified lesion within the aortic wall is **cystic medial degeneration** — decrease in [smooth muscle](#), necrosis, fragmentation of elastic tissue and deposition of the proteoglycan-rich extracellular matrix. Cystic medial degeneration is usually related to a genetic disease such as **Marfan syndrome**. There may be further evidence of atherosclerosis and abnormal connective tissue structure in genetic conditions. Inflammation is absent. Dissection can also occur when no identifiable [histological](#) lesions are present and can be spontaneous.

## Symptoms of Aortic Dissection

## Chest pain and other indications for aortic dissection

Thoracic aortic dissection should be considered in all patients with **chest pain**. This pain usually has the following characteristics:

- **Site:** Chest pain; depends on where the dissection arises (can mimic myocardial infarction pain). This pain is due to the interruption of blood flow to the coronary arteries causing ischemia (usually when arch or root are affected). Pain is usually more sudden and is maximal at the onset, unlike MIs. The dissection is painless in 10 % of patients.
- **Onset:** sudden
- **Character:** excruciating tearing/ripping pain (**tearing pain between the shoulder blades is usually associated with descending aortic dissection**).
- **Radiation: to the back and/or in-between the shoulders.** Can radiate to the neck or jaw (usually occurs with arch dissection which spreads into the branches of the aorta).
- **Severity:** usually excruciating (can be mild in some cases)

The following **neurological symptoms** can be the presenting complaint (20 %):

- **Syncope** (hypovolemia, arrhythmia, increased vagal tone)
- Altered [mental status](#)
- **Stroke** (CVA) — hemiparesis or hemiplegia with hemianesthesia
- Change in sensation (tingling, paresthesia, pain) and motor function (weakness) can occur if peripheral nerves are affected by the lack of blood supply.

Additionally, other types of symptoms may occur alongside aortic dissection:

- **Cardiovascular symptoms:** There may be acute severe aortic valve compromise leading to secondary congestive left [heart failure](#). This leads to orthopnoea and dyspnoea.
- **Hypertension:** underlying hypertension or an increase in circulating catecholamines.
- **Hypotension:** poor prognostic sign as may be the result of cardiac tamponade, hypovolemia or increase vagal tone.
- Symptoms of **esophageal compression:** dysphagia.
- **Abdominal pain** if spread to the abdominal aorta.
- **Flank pain** if the renal arteries are involved.
- Symptoms of **systemic disease(s):** Patients may have established disease(s) associated with the aortic dissection and could, therefore, have symptoms of the peripheral vascular disease, [infection](#), Marfan syndrome or Ehler—Danlos syndrome.

## Signs of aortic dissection

The following list comprises the most common symptoms of aortic dissection:

- Blood pressure: a difference of >20 mm Hg between left and right arms (20 % don't have this).
- **Aortic regurgitation:** bounding (collapsing/water hammer) pulse, wide pulse pressure, murmurs (diastolic).
- Signs of congestive heart failure secondary to acute severe aortic valve

dysfunction leading to orthopnoea, dyspnoea, elevated JVP and bibasal crackles.

- Possibly **unconsciousness**.
- **Cardiac tamponade**: distension of jugular veins, hypotension, pulsus paradoxus, Kussmaul's sign
- Rarely SVC obstruction that can lead to **SVC syndrome**.
- Signs of **stroke** — e.g., body leaning to one side due to hemiparesis.
- Patient in **shock**; cold, clammy, pale, tachycardia, tachypnea.
- **Horner's syndrome** may be present due to compression of the cervical sympathetic chain.
- New diastolic murmur and/or asymmetrical pulses.
- **Pulse**: carotid, brachial and femoral may be abnormal.
- **Bruits**: may be present initially or may develop later.
- Signs of hemothorax may be present if the dissection ruptures into the pleura — rapid shallow breaths, sharp pleuritic pain.
- Anxiety, a sense of impending doom.
- **Acute arterial insufficiency** in the lower or upper limbs, as indicated by weak pulses, pallor, low body temperature, loss of sensation — paraesthesia, paralysis
- Marfan syndrome signs — e.g. high arched palate
- **Ehlers—Danlos syndrome** signs

## Complications

### Risks inherent in aortic dissection

Aortic dissection might cause the following complications:

- **Hypotension and shock** (hypovolemic): eventually death by exsanguination (blood loss)
- Permanent **disability** from stroke (CVA)
- **Acute aortic regurgitation**: proximal dissection spreading to the sinus of Valsalva and aortic root.
- **Pulmonary edema**: acute aortic valve regurgitation
- **Pericardial tamponade**: due to blood in the pericardial sac (hemopericardium)
- **Myocardial ischemia** — reduction in blood flow to the coronary arteries
- **Aortic insufficiency**
- **Myocardial infarction**
- **Global ischemia** e.g. mesenteric, bowel, renal, spinal cord, visceral ischemia/infarction
- **Compression**: **esophagus**, SVC, ganglia (sympathetic chain causing Horner's syndrome), airway, left recurrent **laryngeal nerve** (hoarseness and vocal cord paralysis).
- **Aortic aneurysm**

## Diagnosis of Aortic Dissection



## Methods of detecting aortic dissection

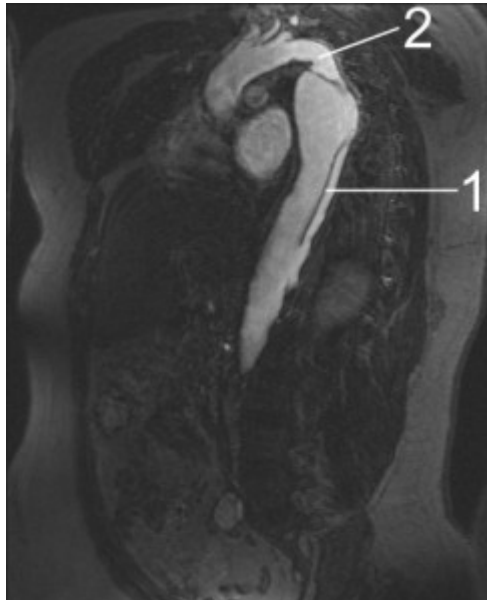
Diagnosis of aortic dissection **needs to be rapid and accurate**. The following procedures are recommended:

- **Physical examination:** signs as stated above.
- **History:** In stable patients, a detailed history should be taken to assess risk factors, hypertension status, the risk of rupture, symptom duration, and onset and any family history or genetic conditions.

## Investigations

Investigations are used to diagnose and reveal the site of the intimal tear and extent of dissection:

### MRI



- [Image:](#) "MRT scan of aortic dissection" by Dr. Lars Grenacher. License: [CC BY—SA 3.0](#)

**The gold standard** for detection and assessment. Sensitivity and specificity of 98 %

- Can create a 3D reconstruction and can determine the intimal tear location (unlike CT scans) and extent of the dissection.
- Non—invasive
- No iodinated contrast needed
- Quantifies the level of aortic insufficiency
- May only be available at larger hospitals. Takes longer than CT scans and therefore may be less useful.

### CT Scan



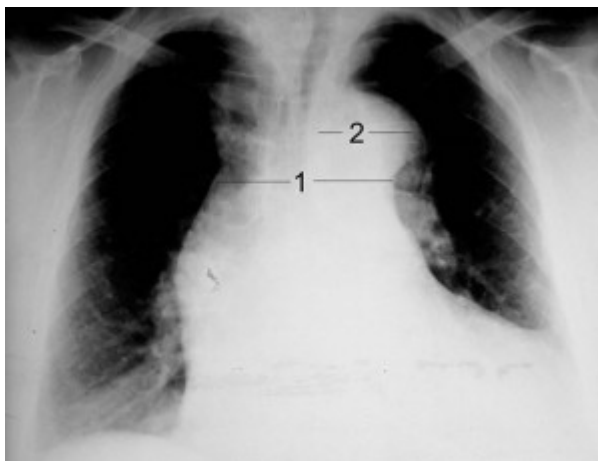
- [Image:](#) " CT scan of aortic dissection" by James Heilman, MD.  
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Non—invasive, rapid and accurate test that can give a 3D view of the aorta — especially useful for surgical interventions

- Injected iodinated contrast medium used
- Very sensitive and specific investigation
- Unable to diagnose the site of the intimal tear

### [Chest X-ray](#)

- Mediastinal widening
- Pleural effusions may be visible
- Calcium sign — the calcified intima is separated from the outer aortic soft tissue border by 1 cm (rare)
- Obliteration of the aortic knob



- [Image:](#) " chest x-ray of aortic dissection type Stanford A" by JHeuser.  
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- Catheterization of the aorta (via the femoral artery) and injection of contrast whilst x—rays are taken of the aorta. Useful for determining the luminal size and to determine and aortic dissections that may be present

## Blood test

- D—Dimer <500 ng/ml may rule out aortic dissection in low-risk patients. It is not specific as D—Dimer can be high for other reasons, therefore, if suspicious, confirm with imaging.
- Cannot be used as a single diagnostic tool

## Transoesophageal echocardiogram

- High sensitivity and specificity
- Fast, minimally invasive and fairly inexpensive
- Can determine if valves or Ostia of the coronary arteries are involved
- Does not provide a full view, hence MRI is recommended

## Aortogram

- The aorta is catheterized and contrast is injected.
- X—rays are taken to determine flow within the aorta.

**Standard blood tests** — to confirm the diagnosis and rule out differential diagnoses

## Differential Diagnoses

Consider aortic dissection with all chest pain presentations:

<b>Heart</b>	Myocarditis, aortic regurgitation, aortic stenosis, cardiogenic shock, myocardial infarction, aortic aneurysm
<b>MSK</b>	Mechanical back pain, muscular pain
<b>Respiratory</b>	Pleural effusion, pulmonary embolism

## Treatment of Aortic Dissection

### Possible therapy for aortic dissection

Acute Stanford Type A (DeBakey 1+2)	Surgery > Medical
Acute Stanford Type B (DeBakey 3)	Medical > Surgical

If the patient presents with chronic aortic dissections (present for > 2 weeks) that they have survived, the treatment varies as the mortality is much higher for **acute aortic dissections** (66 % cases).

## Surgery

Surgical treatment may be indicated for acute dissections type A. The surgery is high risk due to its proximity to the heart and the importance of the aorta. The surgery aims to remove the damaged aorta section and repair the false lumen to prevent the entry of blood. The intimal tear can be removed. The surgery can be carried out via open surgery or endovascularly. Surgery can replace the damaged aorta with a tube graft and/or repair the damaged aortic valve if it is implicated in the aortic dissection.

## Medications

Any patient who presents with aortic dissection needs immediate **blood pressure control**. The target is a mean arterial blood pressure of 60 - 75 mmHg. This is to be managed long-term which may involve medication and therapy to control emotional and

psychological factors.

- **Beta-blockers** — first-line treatment.
- **Vasodilators** — if beta-blockers do not sufficiently reduce blood pressure.
- **Calcium channel blockers** — if beta-blockers are contraindicated (e.g. [asthma](#))

## Prognosis of Aortic Dissection

40 % of all patients die suddenly with acute aortic dissection. Of the remaining 60 %, 1 % of these die every hour; this highlights the importance of rapid diagnosis and referral for surgical repair (or medical treatment if indicated). The surgery itself is an extremely high risk with a mortality of 5—20%.

Ascending aortic dissections have a much worse prognosis in comparison to thoracic aortic dissections.

There are risk factors which affect the prognosis postoperatively: increased preoperative evaluation time, older age, leakage of an aneurysm, cardiac tamponade, preexisting heart pathology (MI, [Coronary artery disease](#)), previous stroke, shock, kidney failure ([acute/chronic](#)).

## Screening

The increased screening takes place for those with family history / those individuals suffering from a genetic condition known to increase the chance of developing aortic dissection (e.g. Marfan and [Ehler—Danlos syndromes](#)).

## References

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