Lecturio Medical Knowledge Essentials – Aortic Dissection

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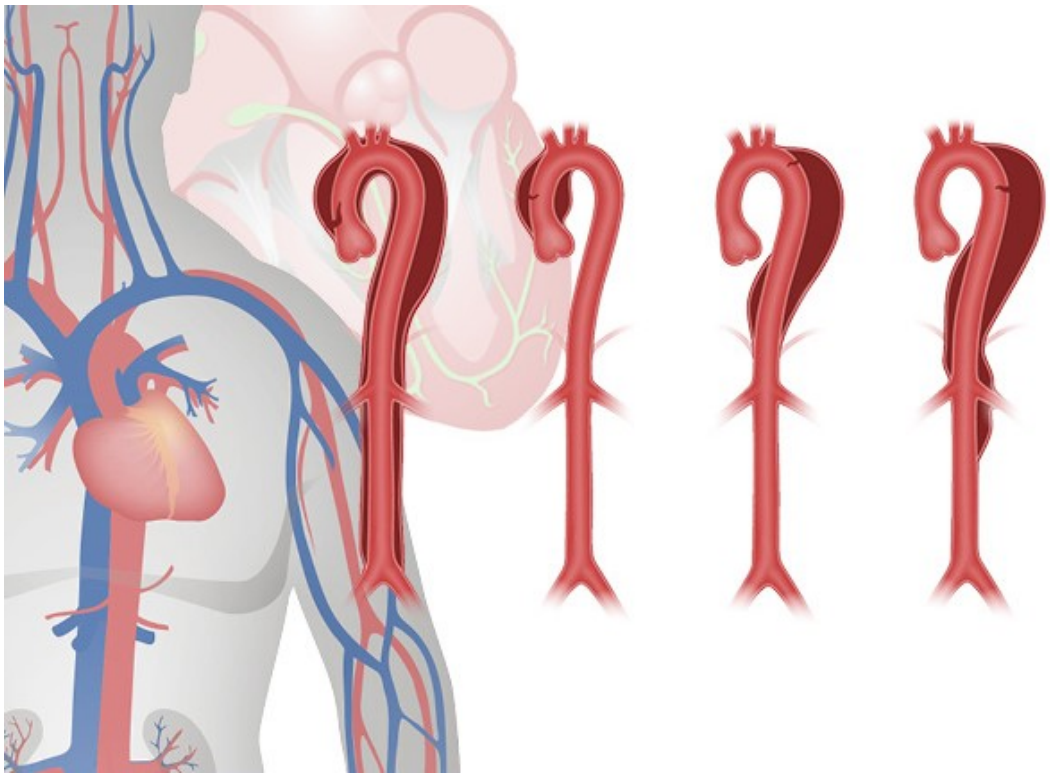

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Aortic dissection is a surgical cardiac emergency characterized by tears in the innermost layer (tunica intima) of the aorta leading to the separation of tunica intima and tunica media and the formation of a new false channel.

In aortic dissection, the tunica intima of the aortic wall develops a fissure that causes blood to enter the media layer of the wall to form a false channel, forcibly separating the layers of the wall and causing 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 resulting in insufficient supply of blood and nutrients to the vital organs. In severe cases, the aorta can rupture and cause rapid death.

For further review of this topic, including links to lectures by specialists in the field, follow this link: https://www.lecturio.com/concepts/aortic-dissection/

not be relied on as health or personal advice. **Always seek the guidance of your doctor** or other qualified health professional with any questions you may have regarding your health or a medical condition.

Epidemiology

Aortic dissection is a rare condition in which new cases are reported at a rate of 2–3.5 per 100,000 people every year. It is more common in men than women: 65% of all dissections are reported in males, mostly associated with hypertension.

People of age 40–70 years are more likely to develop this condition. It is more common in Afro—Caribbean people than Caucasians, similar to the risk of hypertension. Asians have the lowest incidence.

Dissections that occur individuals 30–40 years of age are usually associated with genetic and/or connective tissue diseases such as Marfan syndrome.

Etiology

Acquired Causes

- Hypertension
- Atherosclerosis
- Blunt chest trauma (e.g., car accidents, though they are usually deceleration injuries that more commonly cause a complete aortic transection or iatrogenic trauma (during catheterization or intra-aortic balloon pump counterpulsation)
- Pregnancy, especially in the third trimester and in the postpartum period
- Syphilis (tertiary stage) as it causes vasculitis with aortic involvement.
- Amphetamines and cocaine use.
- Cardiac surgery—especially aortic valve replacement, since aortic regurgitation can cause dilatation and aortic wall weakening.

Congenital Causes

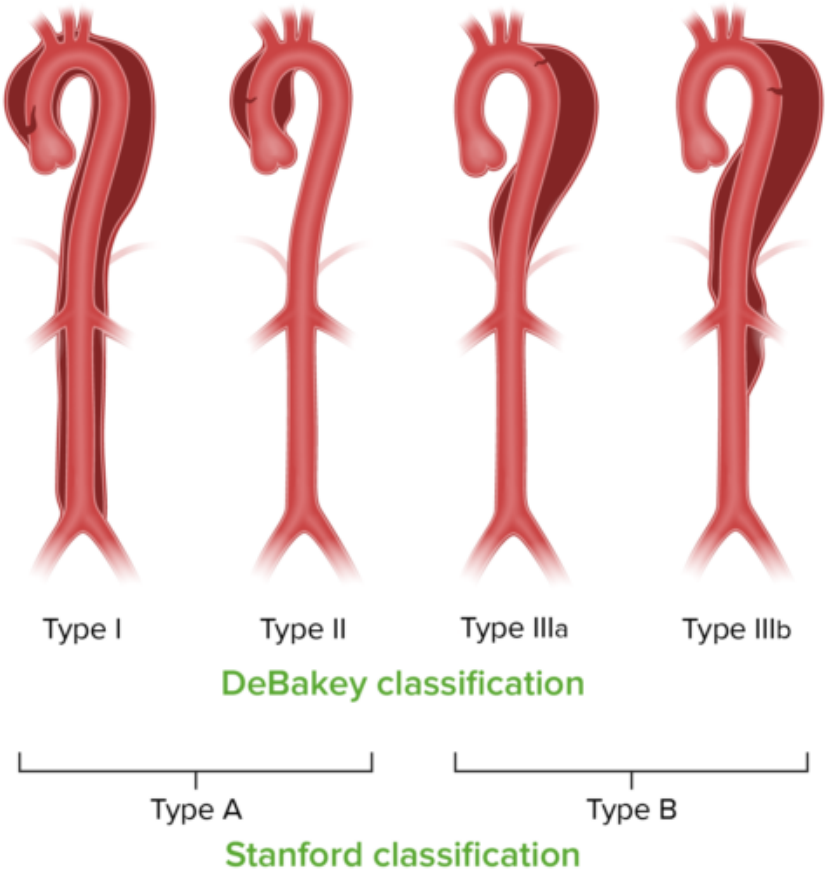
- Genetic disease/connective tissue abnormalities that affect the aorta; affects the structure and function of connective tissue/proteins (e.g., collagen and elastin) in the walls of the aorta — Marfan syndrome (more likely to be proximal dissections), Ehlers-Danlos syndrome
- Turner syndrome (causes aortic root dilatation)
- Bicuspid aortic valve increases the chance of ascending aortic dissection.
- Coarctation of the aorta

High-yield facts:

- Hypertension is the most common cause of ascending aortic aneurysm.
- 70 % of patients with aortic dissection have uncontrolled high blood pressure.

Classification

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



Two different classifications of aortic dissections are used. Of the two, the Stanford classification predominates. Both the DeBakey classification and the Stanford classification are used to separate aortic dissections into those that need surgical repair, and those that usually require only medical management. DeBakey Type I involves the ascending aorta, arch, and descending thoracic aorta and may progress to involve the abdominal aorta); DeBakey Type II is limited to the ascending; DeBakey Type IIIa involves the descending thoracic aorta distal to the left subclavian artery and proximal to the celiac artery; DeBakey Type IIIb involves the thoracic and abdominal aorta distal to the left subclavian artery. In the Stanford classification, Type A involves the ascending aorta and may progress to involve the arch and thoracoabdominal aorta, and Type B involves the descending thoracic or thoracoabdominal aorta distal to the left subclavian artery without involvement of the ascending aorta. The treatment of type A dissections surgical, unless the patients would not survive surgery. Type B dissections can usually be managed medically, but surgery or endovascular intervention may be used if there are complications or progressive symptoms Other classifications of aortic dissections are also used, to improve specificity and reporting standards (J Vasc Surg. 2020;71(3):723). Image by Lecturio.

Stanford Classification

Mnemonic:

Stanford A = Affects ascending aorta

Stanford B = Begins beyond brachiocephalic vessels

The Stanford classification is the most commonly used in aortic dissection.

Type A 70–75%	• Ascending aorta +/- aortic arch, possibly descending aorta. • Can involve the aortic valve	• It requires primary surgical treatment
Type B 25–30%	• Descending aorta or the aorta (distal to the left subclavian artery) without the involvement of the ascending aorta	• It is generally treated conservatively by controlling blood pressure and heart rate. • Surgery is indicated in complicated cases only.

DeBakey system

In contrast, the DeBakey system is based on anatomy:

Type 1 **60%**	Origin — ascending aorta, extends to the aortic arch and often beyond. Most lethal and often seen in patients < 65.	 Image: "DeBakey Type 1" by JHeuser. License: CC BY-SA 3.0
Type 2 **30–35%**	Origin — ascending aorta and is confined here	 Image: "DeBakey Type 2" by JHeuser. License: CC BY-SA 3.0
Type 3 **10–15%**	Origin — descending aorta — rarely goes proximally but commonly goes distally. Elderly with hypertension and atherosclerosis.	 Image: "DeBakey Type 3" by JHeuser. License: CC BY-SA 3.0

Pathophysiology

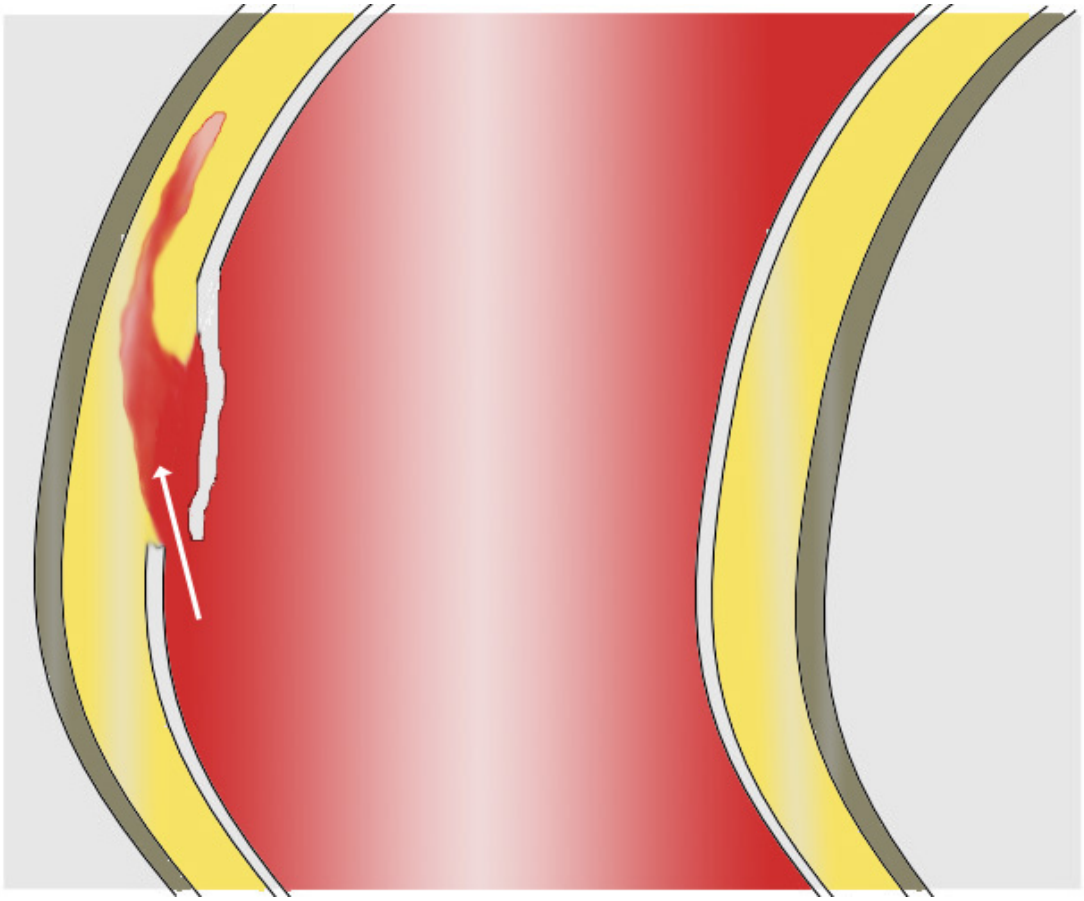


Image: “Scheme of aortic dissection” by JHeuser. License: CC BY-SA 3.0

In aortic dissection, blood enters the intima from the media layers. The high pressure exerted by blood tears the media apart in a laminated plane. The plane is usually between the inner 2/3rds and the outer 1/3rd. The dissection can extend proximally and/or distally for variable distances. This establishes a connection between the media and intima through a false lumen.

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 external rupture. This results in 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 rapidly lead to death.

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. 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 behind why 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 such as Marfan syndrome. In Marfan, 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 resulting in secondary aortic dissection.

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; inappropriate functioning of the mutated fibrillin-1 causes an accumulation of TGF-beta in various tissues including the aorta, resulting in weakened tissue with an abnormal structure and function.
- **Ehlers-Danlos syndrome** is a genetic condition characterized by insufficient production and processing of collagen (an essential protein involved in the structure of tissues). This can lead to weakened vessel walls that can develop an aneurysm

Pathology

Lesions associated with aortic dissection

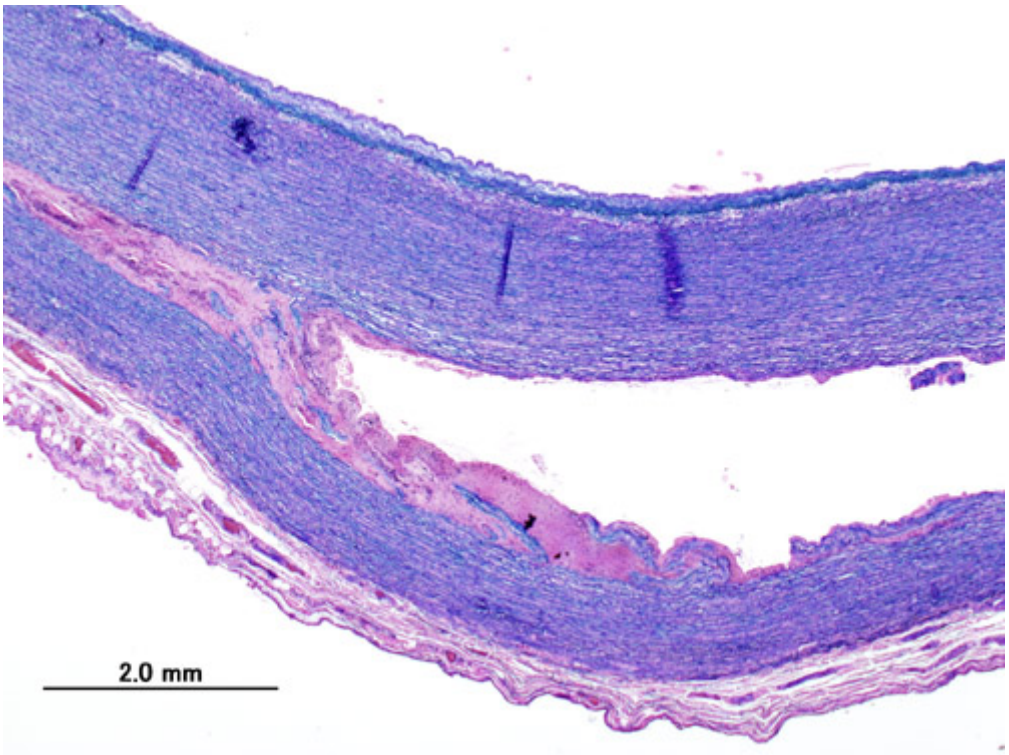


Image: “Histopathological image of dissecting aneurysm of thoracic aorta in a patient without evidence of Marfan’s trait. The damaged aorta was surgically removed and replaced by artificial vessel. Victoria blue & HE

stain." by KGH. License: CC BY-SA 3.0

Clinical Features

Symptoms

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

- Site: Chest pain depends on the location of dissection (can mimic myocardial infarction pain). This pain occurs as a result of 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 severe at onset, when compared to 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 in 20% of cases:

- 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
- Hoarseness due to compression of the laryngeal nerve

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 orthopnea and dyspnea.
- 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 it extends 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 Ehlers-Danlos syndrome.

Signs

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

- Blood pressure that is unequal in both arms, usually with a difference of > 20 mm Hg between left and right arms (may be normal in 20%) due to dissection obstructing the branches of the aorta
- Aortic regurgitation characterized by bounding (collapsing/water hammer) pulse, wide pulse pressure, diastolic murmur
- Signs of congestive heart failure secondary to acute severe aortic valve dysfunction leading to orthopnea, dyspnea, elevated JVP, and bibasilar crackles
- Possibly unconsciousness
- Cardiac tamponade, characterized by distension of jugular veins, hypotension, pulsus paradoxus, Kussmaul's sign
- SVC obstruction can cause to SVC syndrome in rare cases
- 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.
- Numbness and tingling in the upper and lower limbs due to peripheral ischemia
- Signs of hemothorax may be present if the dissection ruptures into the pleura: see rapid shallow breathing, sharp pleuritic pain.
- Acute arterial insufficiency in the lower or upper limbs, as indicated by weak pulses, pallor, low body temperature, loss of sensation—paresthesias, paralysis
- Signs of connective tissue disorders such as Marfan and Ehlers-Danlos syndromes.

Complications

Aortic dissection might cause the following complications:

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

Diagnosis

Methods of Detecting Aortic Dissection

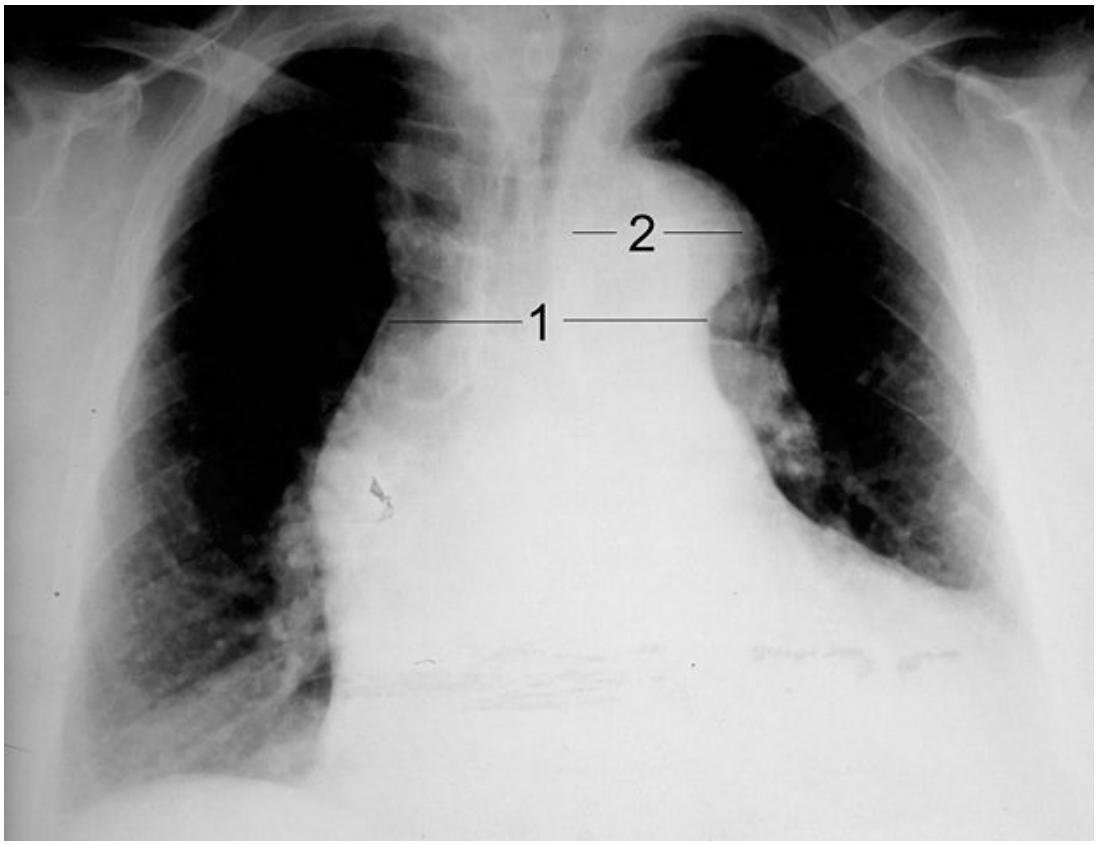
Diagnosis of aortic dissection needs to be rapid and accurate. Diagnosis should be suspected from the history and physical examination as previously explained.

Investigations

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

Chest Radiograph (CXR)

- Initial imaging that shows mediastinal widening; pleural effusions may be visible
- Calcium sign—the calcified intima is separated from the outer aortic soft tissue border by one cm (rare)
- Obliteration of the aortic knob



Chest X-ray shows mediastinal widening (line labeled as 1) and a prominent aortic knob (line labeled as 2) in Stanford type A aortic dissection. Image: “Chest x-ray of aortic dissection type Stanford A” by JHeuser. License: CC BY-SA 3.0

Transesophageal echocardiogram (TEE)

- High sensitivity and specifically for hemodynamically unstable patients
- Fast, minimally invasive, and can be used in unstable patients or in those with renal insufficiency or contrast allergy
- Can determine if valves or ostia of the coronary arteries are involved
- Does not provide a full view, hence MRI is recommended

CT Scan

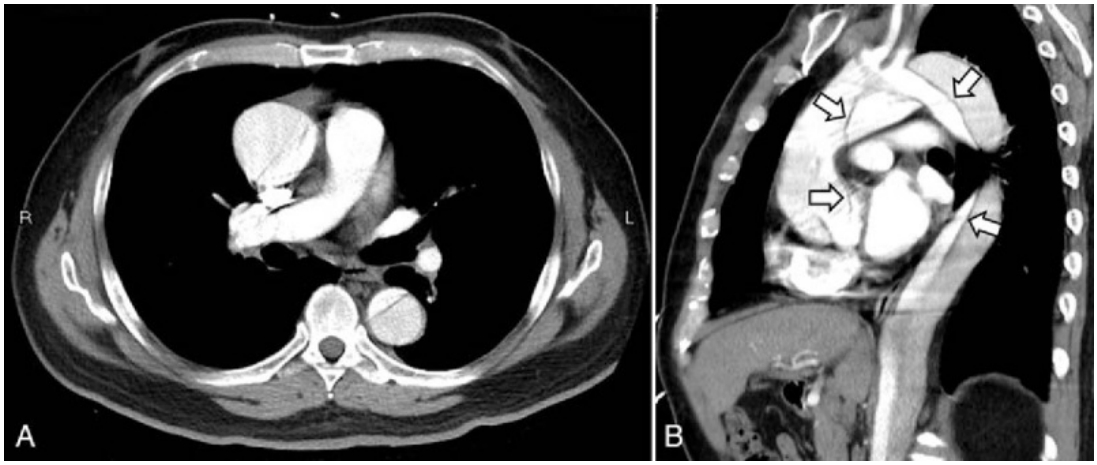


Image: Contrast-enhanced computed tomography of chest indicated type A aortic dissection from aortic root. (A) Axial view; (B) sagittal view
Arrows indicate intimal flap. By: Medicine. License: CC BY 4.0

- Noninvasive, rapid and accurate test that can give a 3D view of the aorta—especially useful for surgical interventions
- It is used in stable patients
- Injected iodinated contrast medium used
- Very sensitive and specific investigation
- Unable to diagnose site of intimal tear

MRI

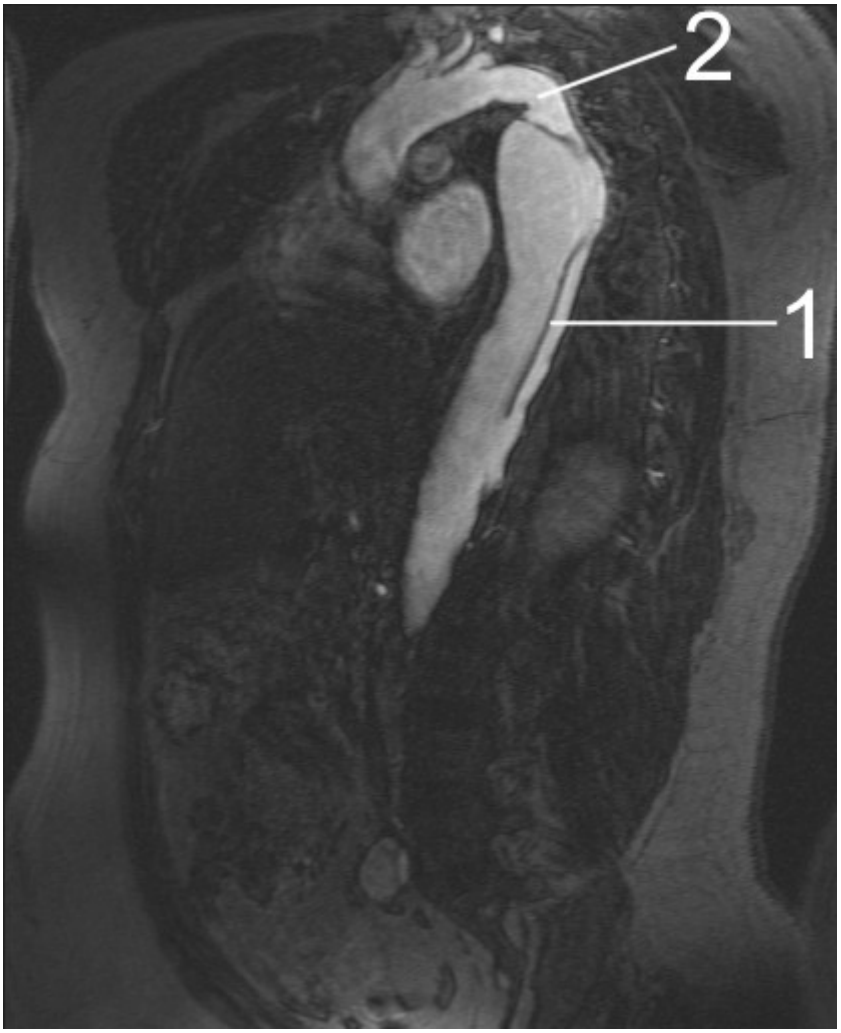


Image: “MRT scan of aortic dissection” by Dr. Lars Grenacher. 1: descending aorta with dissection; 2: aortic arch. License: CC BY-SA 3.0

- Gold standard for detection and assessment.
- Sensitivity and specificity of 98%
- It is used if contrast-enhanced CT is contraindicated in stable patients.
- Can create a 3D reconstruction and can determine the intimal tear location (unlike CT scans) and extent of the dissection
- Noninvasive
- 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.

Differential Diagnoses

Myocarditis, myocardial infarction, aortic aneurysmal rupture, and mechanical chest pain (pain referred from the cervical or thoracic spine or from a costo-chondral or chondro-sternal joint), are the main differential diagnoses.

Treatment

Acute Stanford Type A (DeBakey 1+2) Acute ascending aortic dissection	A surgical emergency; medical therapy alone only if major comorbidities
Acute Stanford Type B (DeBakey 3) Descending aortic dissection	Medical initially and surgical if complications (occlusion of major branch, severe hypertension, chest pain, propagation of the dissection aneurysmal expansion, expanding hematoma, or rupture)

Note:

- Type A aortic dissection is usually treated by emergent open surgery, less often by endovascular stent-grafting if there are major comorbidities, and a hybrid approach has been used (surgical repair of the ascending aorta, and endovascular stent-graft for the descending aorta).
- Type B aortic distension is initially treated by beta-blockers, vasodilators, or calcium channel blockers; open surgical repair is if there are complications (occlusion of major branch, severe hypertension, chest pain, propagation of the dissection aneurysmal expansion, expanding hematoma, or rupture)

Early Treatment

Beta-blockers given via intravenous route are the first-line early treatment of AD, with the goal of decreasing the heart rate to 60 bpm. The next step is to control hypertension which can be achieved with beta-blockers alone or with an add-on therapy with nitroprusside if systolic BP > 120 mm Hg; calcium channel blockers can be used if beta blockers are not tolerated IV labetalol, esmolol, and propranolol are the most commonly used beta-blockers in this setting.

Note:

Initial treatment should be beta-blockers before vasodilators to avoid reflex tachycardia.

Note:

Beta-blockers lower the heart rate, reducing shearing forces with the aorta.

Surgery

Aortic dissection involving the ascending aorta is a surgical emergency. The surgery involves excision of the intimal tear, obliteration of the proximal entry point into the false lumen, reconstitution of the aorta with a synthetic graft, and repair or replacement of the aortic valve.

Prognosis

Acute aortic dissection has a high mortality rate, 40% at time of presentation. Of the remaining 60%, 1–2% die every hour without therapy, which highlights the importance of rapid diagnosis and referral for surgical repair (or medical treatment if indicated). The surgery itself has a 7–36% risk of mortality for type A dissections.

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

There are risk factors which affect the prognosis postoperatively:

- Increased preoperative evaluation time

- Older age
- Leakage of an aneurysm
- Cardiac tamponade
- Pre-existing cardiac pathology (MI, coronary artery disease)
- Previous stroke
- Shock
- Kidney failure (acute/chronic)

Screening

High-risk individuals such as those with family history of collagen-disease or AD should be screened, especially if they develop hypertension.

Review Questions

1. **The most common type of aortic dissection is…?**
    A. …Stanford Type A.
    B. …Stanford Type B.
2. **Which of the radiological investigations, (assuming all are available at the hospital) is the most useful for diagnosing and managing aortic dissection?**
    A. CT Scan
    B. X-ray
    C. Aortogram
    D. MRI Scan
    E. Echocardiogram
3. **In aortic dissection, what is the strand of tissue known as the tissue that splits off but remains in the lumen?**
    A. Intimal tear
    B. Intimal flap
    C. Medial slice
    D. Medial tear
    E. Adventitial flap

Answers: 1A, 2D, 3B

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Notes