Diseases of Aorta

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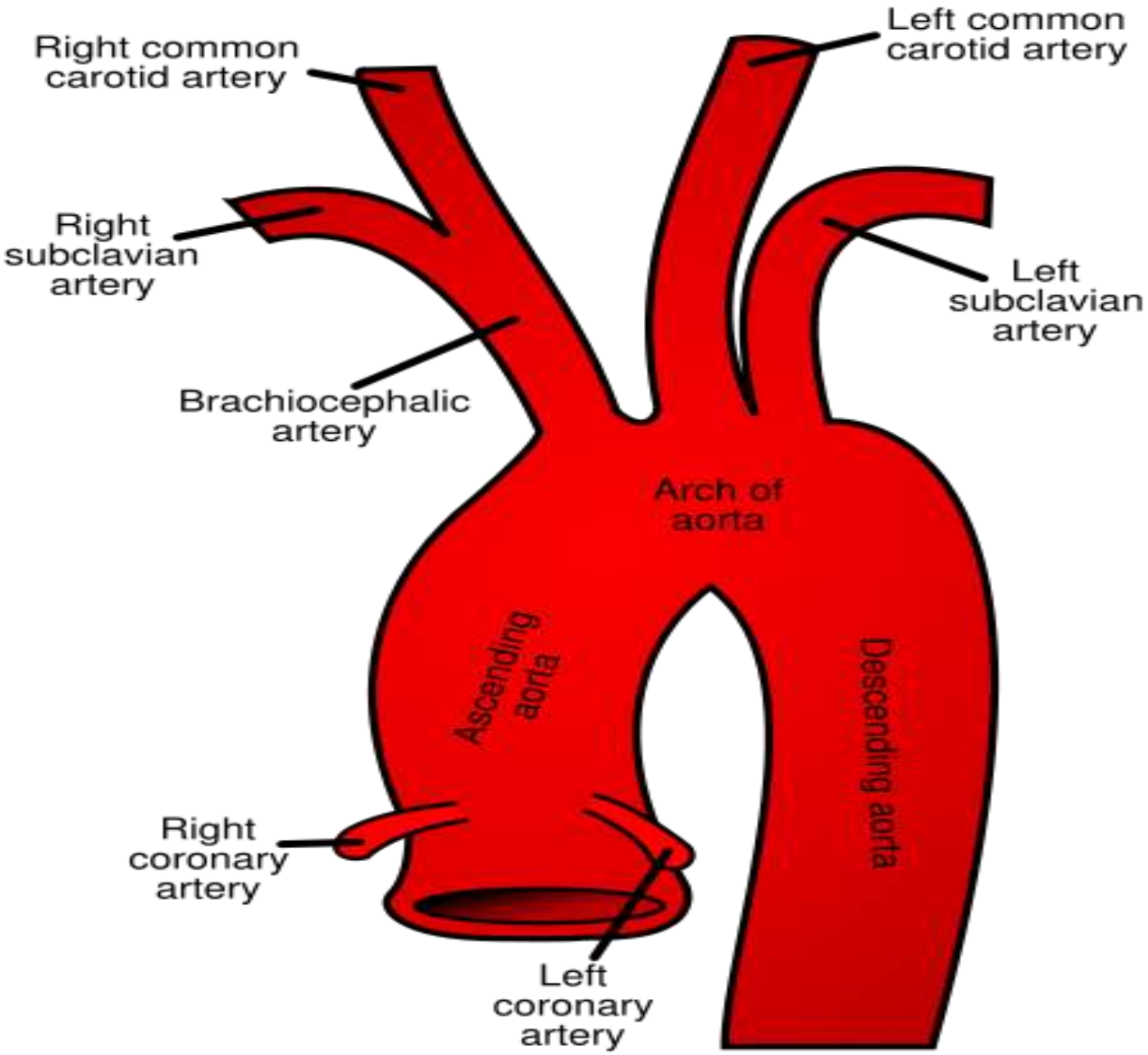
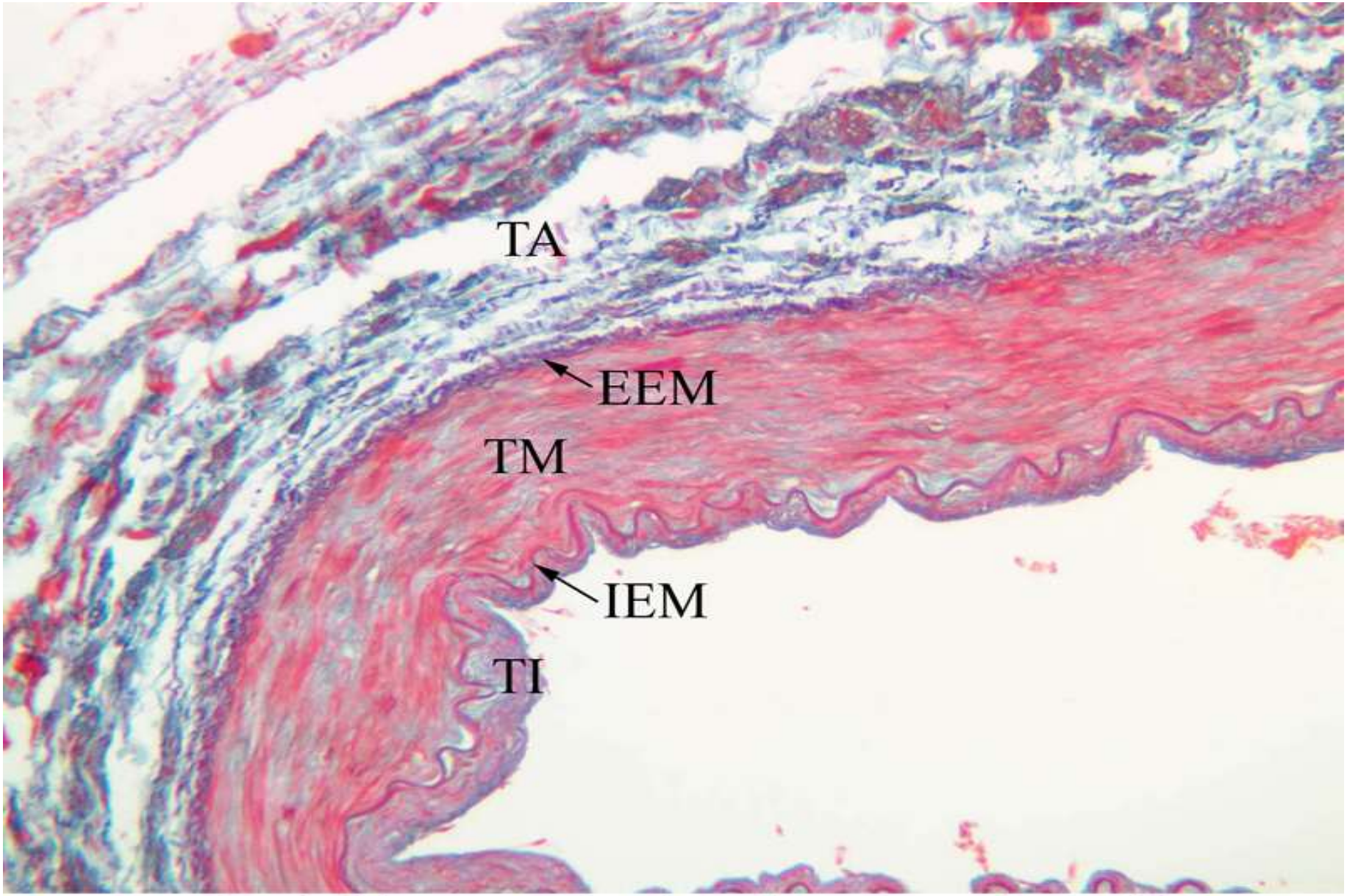
- Aorta is the conduit through which blood ejected from the left ventricle is delivered to the systemic arterial bed.

- In adults, its diameter is approximately 3 cm at the origin and in the ascending portion, 2.5 cm in the descending portion in the thorax, and 1.8–2 cm in the abdomen.

- The aortic wall consists of a thin intima composed of endothelium, subendothelial connective tissue, and an internal elastic lamina; a thick tunica media composed of smooth muscle cells and extracellular matrix; and an adventitia composed primarily of connective tissue enclosing the vasa vasorum and nervi vascularis.


Right common carotid artery
Left common carotid artery
Right subclavian artery
Left subclavian artery
Brachiocephalic artery
Arch of aorta
Ascending aorta
Descending aorta
Right coronary artery
Left coronary artery



TI - tunica intima TM - tunica media TA - tunica adventitia
IEM - internal elastic membrane EEM - external elastic membrane

- In addition to the conduit function, it distends during systole to allow a portion of the stroke volume and elastic energy to be stored, and recoils during diastole so that blood continues to flow to the periphery.

- Because of its continuous exposure to high pulsatile pressure and shear stress, the aorta is particularly prone to injury and disease resulting from mechanical trauma.

Congenital Anomalies

- Symptoms such as dysphagia, stridor, and cough may occur if an anomaly causes a ring around or otherwise compresses the esophagus or trachea.

- Anomalies associated with symptoms include double aortic arch, origin of the right subclavian artery distal to the left subclavian artery, and right-sided aortic arch with an aberrant left subclavian artery.

- A Kommerell's diverticulum is an anatomic remnant of a right aortic arch.

Diseases of Aorta

- Aortic aneurysm
- Acute aortic syndromes (aortic dissection, acute intramural hematoma, penetrating atherosclerotic ulcer)
- Aortic occlusion
- Aortitis

Aortic Aneurysm

- An aneurysm is defined as a pathologic dilation of a segment of a blood vessel ie. 1.5 times to that of expected normal diameter

- A fusiform aneurysm affects the entire circumference of a segment of the vessel, resulting in a diffusely dilated artery

- Saccular aneurysm involves only a portion of the circumference, resulting in an outpouching of the vessel wall

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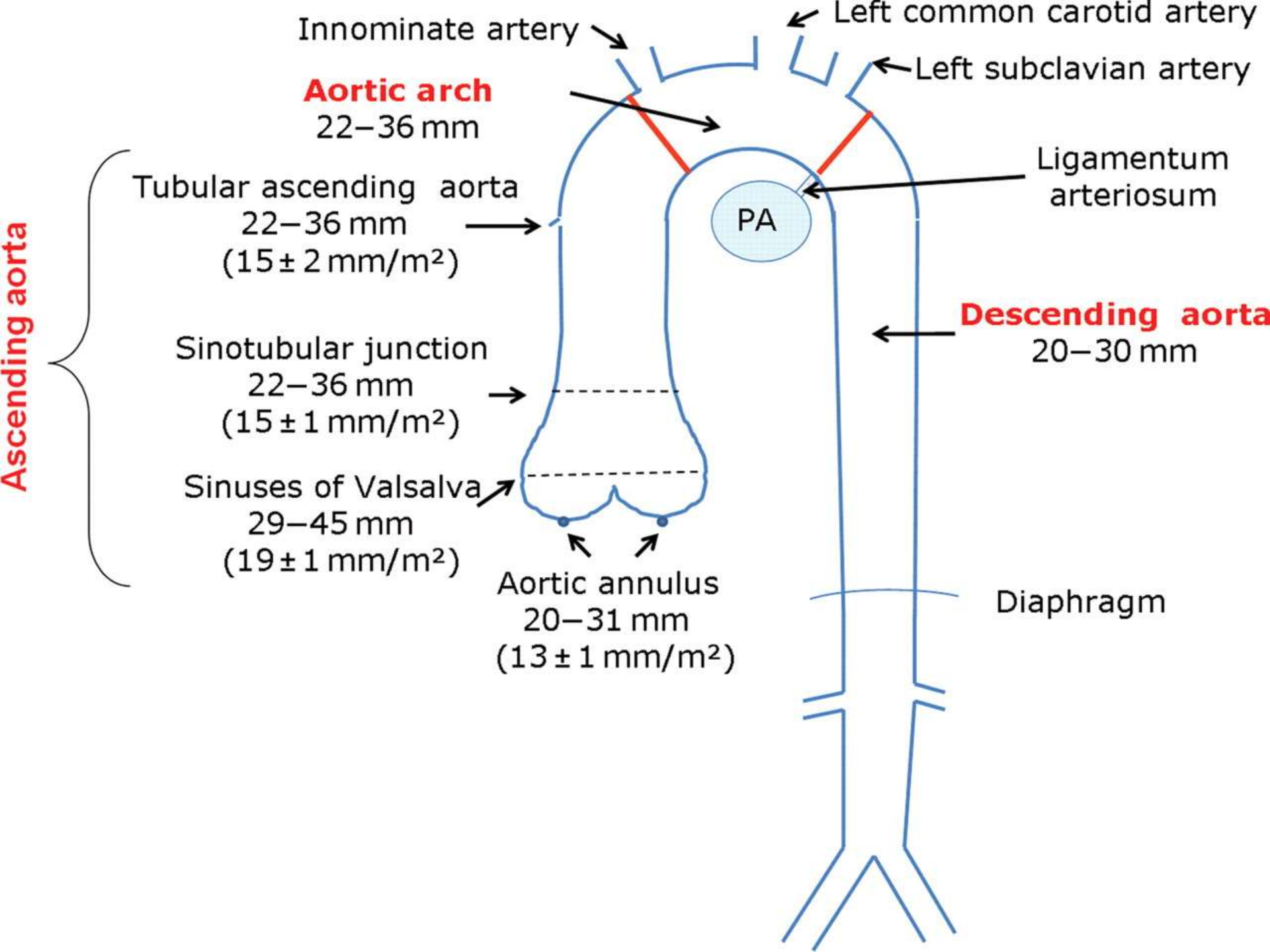

Innominate artery
Left common carotid artery
Left subclavian artery
Aortic arch
22–36 mm
Ligamentum arteriosum
PA
Ascending aorta
Tubular ascending aorta
22–36 mm
(15 ± 2 mm/m²)
Descending aorta
20–30 mm
Sinotubular junction
22–36 mm
(15 ± 1 mm/m²)
Sinuses of Valsalva
29–45 mm
(19 ± 1 mm/m²)
Aortic annulus
20–31 mm
(13 ± 1 mm/m²)
Diaphragm

Etiology

Degenerative/atherosclerosis

- Aging
- Cigarette smoking
- Hypercholesterolemia
- Hypertension
- Atherosclerosis

Genetic or developmental

- Marfan syndrome
- Loeys-Dietz syndrome
- Ehlers-Danlos syndrome type IV
- Familial
- Bicuspid aortic valve

Chronic aortic dissection

Trauma

Aortitis

- Vasculitis
  - Takayasu's arteritis
  - Giant cell arteritis
- Rheumatic
  - HLA-B27–associated spondyloarthropathies,
  - Behçet's syndrome,
  - Cogan's syndrome
  - Idiopathic aortitis
- Infective
  - Syphilis
  - Tuberculosis
  - Mycotic (Salmonella, staphylococcal, streptococcal, fungal)

Thoracic Aortic Aneurysm

- Cystic medial necrosis is the most common pathology associated with ascending aortic aneurysms, whereas atherosclerosis is the condition most frequently associated with aneurysms of the aortic arch and descending thoracic aorta.

- Risk of rupture is 2–3% per year for thoracic aortic aneurysms <4.0 cm whereas 7% per year for those >6 cm in diameter.

- Most thoracic aortic aneurysms are asymptomatic

- Most patients are hypertensive but remain relatively asymptomatic until the aneurysm expands

- Ascending aortic aneurysms tend to cause anterior chest pain, whereas arch aneurysms more likely cause pain radiating to the neck

- Descending thoracic aneurysms more likely cause back pain localized between the scapulae

- When located at the level of the diaphragmatic hiatus, the pain occurs in the mid back and epigastric region

- Large ascending aortic aneurysms may cause superior vena cava obstruction manifesting as distended neck veins

- Ascending aortic aneurysms also may develop aortic insufficiency, with widened pulse pressure or a diastolic murmur, and heart failure

- Arch aneurysms may cause hoarseness, which results from stretching of the recurrent laryngeal nerves

- Descending thoracic aneurysms and thoracoabdominal aneurysms may compress the trachea or bronchus and cause dyspnea, stridor, wheezing, or cough

- Compression of the esophagus results in dysphagia

- Erosion into surrounding structures may result in hemoptysis, hematemesis, or GI bleeding

- Erosion into the spine may cause back pain or instability

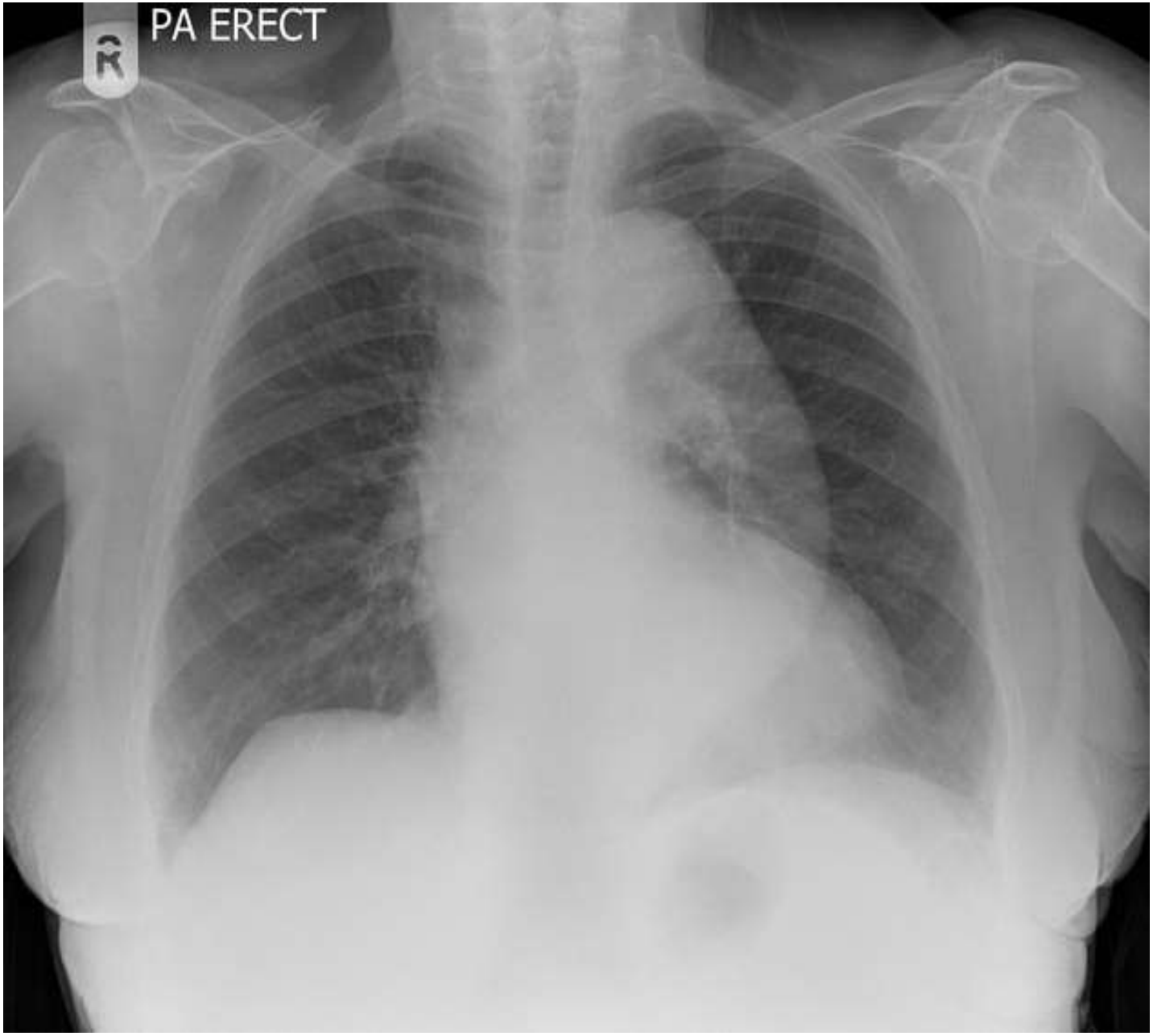
- Spinal cord compression or thrombosis of spinal arteries may result in neurologic symptoms of paraparesis or paraplegia

- Descending thoracic aneurysms may thrombose or embolize clot and atheromatous debris distally to visceral, renal, or lower extremities

Investigations

- Chest xray:
  - Widening of the mediastinal shadow and
  - Displacement or compression of the trachea or left main stem bronchus

- Echocardiography: modality for imaging the aortic root, which is important for patients with Marfan syndrome, but it does not image the middle or distal ascending aorta well in many cases and is particularly limited in its ability to image the descending thoracic aorta


PA ERECT

- Contrast-enhanced CT, magnetic resonance imaging (MRI), and conventional invasive aortography are sensitive and specific tests for assessment of aneurysms of the thoracic aorta and involvement of branch vessels

Treatment

- Beta-Adrenergic blockers currently are recommended for patients with thoracic aortic aneurysms, especially in Marfan syndrome, with evidence of aortic root dilatation to reduce further expansion

- Additional therapy to control hypertension.

- Angiotensin receptor antagonists and angiotensin-converting enzyme inhibitors might reduce the rate of aortic dilation in patients with Marfan syndrome by blocking TGF- signaling

- Operative repair with placement of a prosthetic graft is indicated in symptomatic patients, in whom the ascending aortic diameter is >5.5cm or the descending thoracic aortic diameter is >6 cm, and those with an aneurysm that has increased by >1 cm per year.

- In patients with Marfan syndrome or bicuspid aortic valve, ascending thoracic aortic aneurysms of 4-5 cm should be considered for surgery

- Endovascular repair should be considered when feasible in patients with diameter >5.5 cm.

Abdominal Aortic Aneurysm

- M>F
- Incidence increases with age
- Abdominal aortic aneurysms 4.0 cm may affect 1–2% of men older than 50 years
- 90% of aneurysms >4.0 cm are related to atherosclerotic disease, and most of these are below the level of the renal arteries
- Risk of rupture increases with size: the 5-year risk of aneurysms <5 cm is 1–2%, whereas it is 20–40% for aneurysms >5 cm in diameter

- Mostly asymptomatic
- Usually detected on routine examination as a palpable, pulsatile, expansile, and nontender mass
- Incidental finding observed on an abdominal x-ray or ultrasound study
- Pain is the most frequent complaint and is usually located in the hypogastrium or lower part of the back
- The pain is usually steady, has a gnawing quality, and may last for hours to days at a time
- Patients may be more comfortable in certain positions, such as with the legs drawn up

- New or worsening pain, often of sudden onset, suggest expansion or rupture

- Pain is characteristically constant, severe, and located in the back or lower part of the abdomen, sometimes radiates into the groin, buttocks, or legs

- Actual rupture is associated with abrupt onset of back pain along with abdominal pain and tenderness

- Most patients have a palpable, pulsatile abdominal mass, and many are hypotensive when initially seen

- Triad of abdominal/back pain, a pulsatile abdominal mass, and hypotension recognized as pathognomonic of a ruptured abdominal aortic aneurysm and is seen in as few as one-third of cases

- Hemorrhagic shock and its complications may ensue rapidly

- Retroperitoneal hemorrhage may be signaled by hematomas in the flanks and groin

- Rupture into the abdominal cavity may result in abdominal distention, whereas rupture into the duodenum is manifested as massive gastrointestinal hemorrhage

- Many aneurysms can be detected on physical examination, although even large aneurysms may be difficult or impossible to detect in obese individuals

- When palpable, a pulsatile mass extending variably from the xiphoid process to the umbilicus may be appreciated

- Aneurysms are often sensitive to palpation and may be quite tender if rapidly expanding or about to rupture

- Although tender aneurysms should be examined cautiously, no risk is known to be associated with palpation of the abdominal aorta.

- Associated occlusive arterial disease is sometimes present in the femoral pulses and distal pulses in the legs and feet

- Bruits arising from associated narrowed arteries can be heard over the aneurysm

- Occasionally, an arteriovenous fistula is formed by spontaneous rupture into the inferior vena cava, iliac vein, or renal vein and can cause a syndrome of hemodynamic collapse and acute high-output cardiac failure

Imaging

- Abdominal radiography may demonstrate the calcified outline of the aneurysm; however, about 25% of aneurysms are not calcified and cannot be visualized by x-ray imaging

- An abdominal ultrasound can delineate the transverse and longitudinal dimensions of an abdominal aortic aneurysm and may detect mural thrombus

- Abdominal ultrasound is useful for serial documentation of aneurysm size and can be used to screen patients at risk for developing an aortic aneurysm

- CT with contrast and MRI are accurate noninvasive tests to determine the location and size of abdominal aortic aneurysms and to plan endovascular or open surgical repair

- Contrast aortography may be used for the evaluation of patients with aneurysms, but the procedure carries a small risk of complications such as bleeding, allergic reactions, and atheroembolism

Treatment

- Operative repair with insertion of a prosthetic graft or endovascular placement of an aortic stent graft is indicated for abdominal aortic aneurysms of any size that are expanding rapidly or are associated with symptoms

- For asymptomatic aneurysms, repair is indicated if the diameter is >5.5 cm

- Serial noninvasive follow-up of smaller aneurysms (<5 cm) is an alternative to immediate repair

- Long-term surveillance with CT or MR aortography is indicated after endovascular repair to detect leaks and possible aneurysm expansion

- Medical management:

- Risk factor modification
- Smoking cessation
- Adequate control of hypertension
- Hypercholesterelemia if present should be adequately treated
- Beta blockers

Aortic Dissection

- Aortic dissection is caused by a circumferential or, less frequently, transverse tear of the intima.
- It often occurs along the right lateral wall of the ascending aorta where the hydraulic shear stress is high.
- Another common site is the descending thoracic aorta just below the ligamentum arteriosum.
- The initiating event is either a primary intimal tear with secondary dissection into the media or a medial hemorrhage that dissects into and disrupts the intima.

- The pulsatile aortic flow then dissects along the elastic lamellar plates of the aorta and creates a false lumen.

- The dissection usually propagates distally down the descending aorta and into its major branches, but it may propagate proximally

- Two important pathologic and radiologic variants of aortic dissection: intramural hematoma without an intimal flap and penetrating atherosclerotic ulcer

- Intramural hematoma result from rupture of the vasa vasorum with hemorrhage into the wall of the aorta which may progress to dissection and rupture

- Most of these hematomas occur in the descending thoracic aorta

- Penetrating atherosclerotic ulcers are caused by erosion of a plaque into the aortic media, are usually localized, and are not associated with extensive propagation

- They are found primarily in the middle and distal portions of the descending thoracic aorta and are associated with extensive atherosclerotic disease

- The ulcer can erode beyond the internal elastic lamina, leading to medial hematoma, and may progress to false aneurysm formation or rupture

Classification

- DeBakey

- type I, tear occurs in the ascending aorta but involves the descending aorta as well;
- type II, in which the dissection is limited to the ascending aorta; and
- type III, in which the intimal tear is located in the descending aorta with distal propagation of the dissection

- Stanford

- type A, in which the dissection involves the ascending aorta (proximal dissection), and
- type B, in which it is limited to the descending aorta (distal dissection).

Classification of aortic dissection

			
Percentage	60%	10–15%	25–30%
Type	DeBakey I	DeBakey II	DeBakey III
	Stanford A (Proximal)		Stanford B (Distal)

- Factors predisposing to aortic dissection

- Systemic hypertension
- Cystic medial necrosis
- Marfan syndrome
- Ehlers-Danlos syndrome
- Takayasu’s arteritis
- Giant cell arteritis
- Congenital aortic valve anomalies (e.g., bicuspid valve, coarctation of the aorta
- Aortic trauma
- Third trimester of pregnancy

Clinical Manifestations

- The peak incidence is in the sixth and seventh decades
- M:F ratio 2:1
- Acute aortic dissection presents with the sudden onset of pain, which often is described as very severe and tearing and is associated with diaphoresis
- The pain may be localized to the front or back of the chest, often the interscapular region, and typically migrates with propagation of the dissection
- Other symptoms include syncope, dyspnea, and weakness

- Physical findings include hypertension or hypotension, loss of pulses, aortic regurgitation, pulmonary edema, and neurologic findings due to carotid artery obstruction (hemiplegia, hemianesthesia) or spinal cord ischemia (paraplegia)

- Bowel ischemia, hematuria, and myocardial ischemia have all been observed reflecting complications resulting from the dissection occluding the major arteries

- Furthermore, clinical manifestations may result from the compression of adjacent structures (e.g., superior cervical ganglia, superior vena cava, bronchus, esophagus) by the expanding dissection, causing aneurysmal dilation, and include Horner's syndrome, superior vena cava syndrome, hoarseness, dysphagia, and airway compromise

- Hemopericardium and cardiac tamponade may complicate a type A lesion with retrograde dissection

- Acute aortic regurgitation is an important and common (>50%) complication of proximal dissection

Investigations

- Chest xray:

- Widened mediastinum
- Calcium Sign: Separation of the intimal calcification from the outer aortic soft tissue border by more than 1 cm
- Left sided pleural effusion

- Electrocardiography: One third of electrocardiograms show changes consistent with left ventricular hypertrophy, whereas another one third are normal

- D-dimer level

Diagnostic testing

- The diagnosis of aortic dissection can be established by noninvasive techniques such as echocardiography, CT, and MRI
- Aortography is used less commonly because of the accuracy of these noninvasive techniques
- Transthoracic echocardiography can be performed simply and rapidly and has an overall sensitivity of 60–85% for aortic dissection
- Echocardiography also provides important information regarding the presence and severity of aortic regurgitation and pericardial effusion

- CT and MRI are both highly accurate in identifying the intimal flap and the extent of the dissection and involvement of major arteries; each has a sensitivity and specificity >90%

- They are useful in recognizing intramural hemorrhage and penetrating ulcers

- MRI also can detect blood flow, which may be useful in characterizing antegrade versus retrograde dissection

Treatment

- Intensive Care Unit Admission for hemodynamic monitoring
- Unless contraindicated, beta adrenergic blockers should be administered parenterally using intravenous propranolol, metoprolol, or short acting esmolol to achieve a heart rate of 60 beats/ min
- This should be accompanied by iv nitroprusside infusion to lower systolic BP to <120 mmhg
- Labetalol may also be used

- Verapamil or Diltiazem may also be used

- Enalaprilat plus beta blocker can also be considered

- Emergent or urgent surgical correction is the preferred treatment for acute ascending aortic dissections and intramural hematomas (type A) and for complicated type B dissections, including those characterized by propagation, compromise of major aortic branches, impending rupture, or continued pain

- Surgery involves excision of the intimal flap, obliteration of the false lumen, and placement of an interposition graft.

- A composite valve-graft conduit is used if the aortic valve is disrupted

- Overall in-hospital mortality rate after surgical treatment of patients with aortic dissection is reported to be 15–25%

- The major causes of perioperative mortality and morbidity include myocardial infarction, paraplegia, renal failure, tamponade, hemorrhage, and sepsis

- For uncomplicated and stable distal dissections and intramural hematomas (type B), medical therapy is the preferred treatment

- The in-hospital mortality rate of medically treated patients with type B dissection is 10–20%

- Long-term management consists of control of hypertension and reduction of cardiac contractility with the use of beta blockers plus other antihypertensive agents, such as ACE inhibitors or calcium antagonists

- Patients with chronic type B dissection and intramural hematomas should be followed on an outpatient basis every 6–12 months with contrast-enhanced CT or MRI to detect propagation or expansion

- The long-term prognosis for patients with treated dissections is generally good with careful follow-up; the 10-year survival rate is approximately 60%

References

- Harrison's Principles of Internal Medicine 19th edition

- Braunwald's Heart Disease 8th edition


Thank you