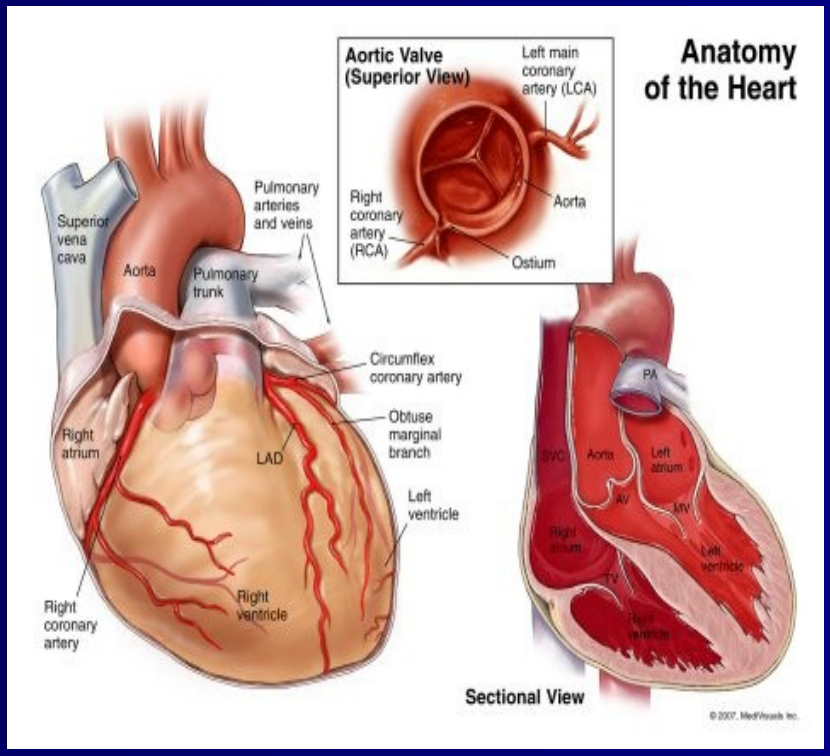
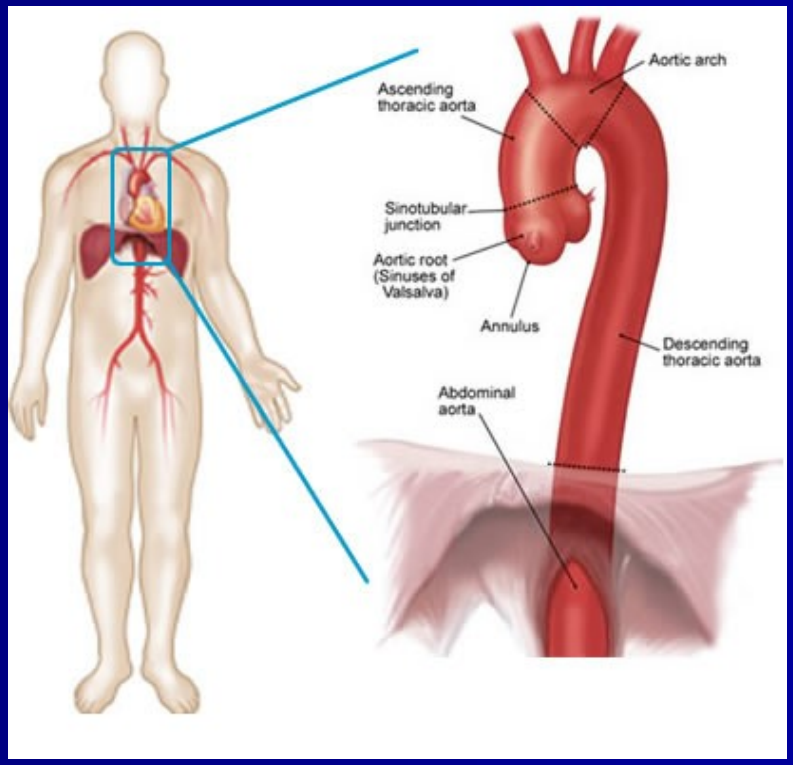
Thoracic aorta pathology

Sotiris Papaspyros

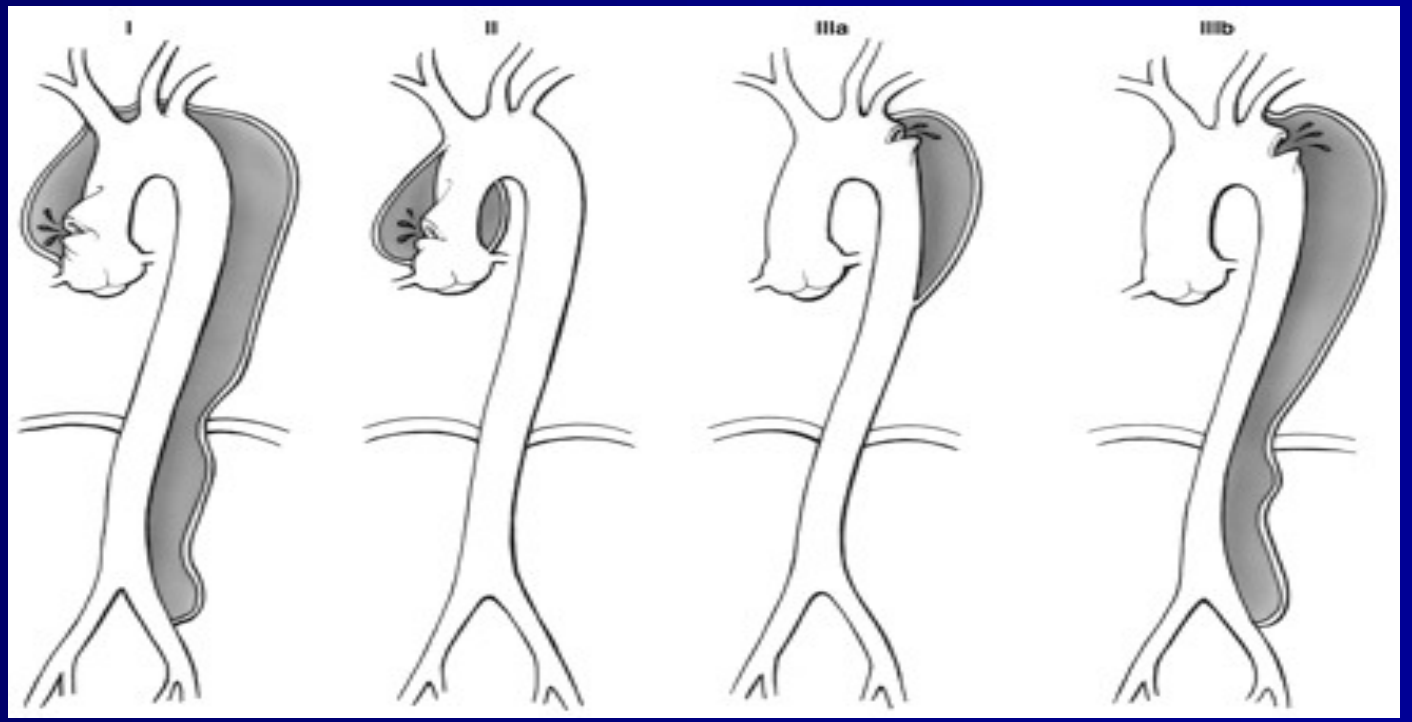
- Aortic Dissection
- Aneurysms of the Thoracic Aorta

Anatomy


Anatomy of the Heart
Aortic Valve (Superior View)
Left main coronary artery (LCA)
Right coronary artery (RCA)
Aorta
Ostium
Superior vena cava
Aorta
Pulmonary arteries and veins
Pulmonary trunk
Circumflex coronary artery
Obtuse marginal branch
LAD
Right atrium
Left ventricle
Right ventricle
Right coronary artery
PA
Aorta
Left atrium
AV
MV
Left ventricle
TV
Sectional View


Aortic arch
Ascending thoracic aorta
Sinotubular junction
Aortic root (Sinuses of Valsalva)
Annulus
Descending thoracic aorta
Abdominal aorta

Classification



Classification of aortic dissection. DeBakey type I and Stanford type A include dissections that involve the proximal aorta, arch, and descending thoracic aorta. DeBakey type II only involves the ascending aorta; this dissection is included in Stanford type A. DeBakey type III and Stanford type B include dissections that originate in the descending thoracic and thoracoabdominal aorta regardless of any retrograde involvement of the arch. These are subdivided into a and b depending on abdominal aortic involvement.

Risk factors

- Hypertension
- Connective tissue disorders Ehlers-Danlos syndrome Marfan disease Turner's syndrome
- Cystic medial disease of aorta
- Aortitis
- Iatrogenic
- Atherosclerosis
- Thoracic aortic aneurysm
- Bicuspid aortic valve
- Trauma
- Pharmacologic
- Coarctation of aorta
- Hypervolemia (pregnancy)
- Congenital aortic stenosis
- Polycystic kidney disease
- Pheochromocytoma
- Sheehan's syndrome,Cushing's syndrome

Dissection sites

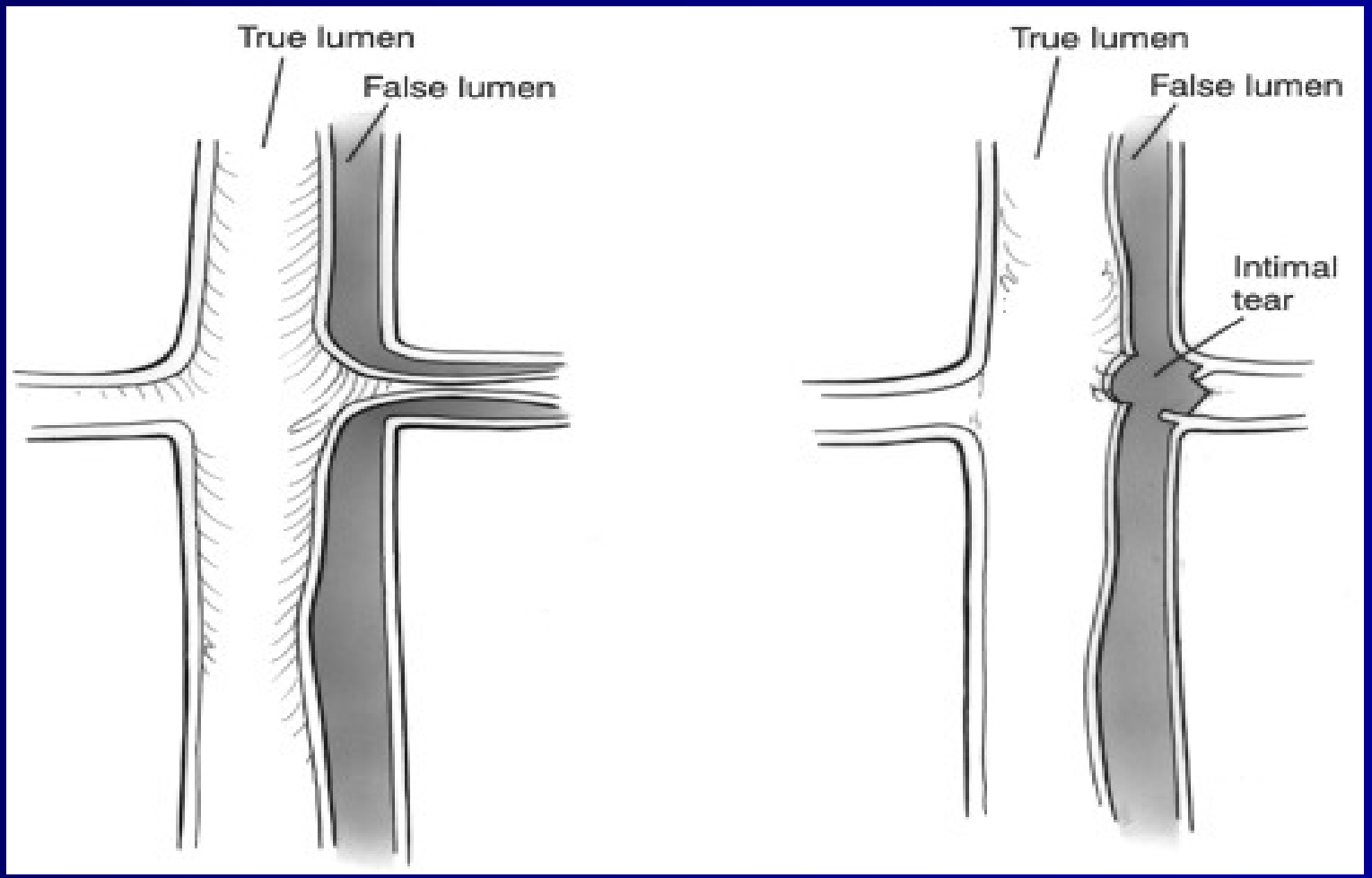
- The most common site of dissection is the first few centimeters of the ascending aorta, with 90% occurring within 10 centimeters of the aortic valve.
- The second most common site is just distal to the left subclavian artery.
- Between 5% and 10% of dissections do not have an obvious intimal tear. These often are attributed to rupture of the aortic vasa vasorum as first described by Krukenberg in 1920.

Incidence/Mortality

- Aortic dissection is the most common catastrophe of the aorta, 2-3 times more common than rupture of the abdominal aorta. When left untreated, about 33% of patients die within the first 24 hours, and 50% die within 48 hours. The 2-week mortality rate approaches 75% in patients with undiagnosed ascending aortic dissection.
- Race: Aortic dissection is more common in blacks than in whites and less common in Asians than in whites.
- Sex: The male-to-female ratio is 3:1.
- Age: Approximately 75% of dissections occur in those aged 40-70 years, with a peak in the range of 50-65 years.

History

- ". . . spontaneous tear of the arterial coats is associated with atrocious pain, with symptoms, indeed, in the case of the aorta of angina pectoris and many instances have been mistaken for it" William Osler, 1910.


True lumen
False lumen
True lumen
False lumen
Intimal
tear

Presenting signs and symptoms

- Chest pain is the most common presenting symptom, ripping or tearing, maximal at onset.
- sudden onset of chest pain has been shown to have a sensitivity of 84%.
- some patients present with only mild pain, often mistaken for musculoskeletal conditions, located in the thorax, groin, or back.
- painless in about 10% of patients

Presenting signs & symptoms

- Anterior chest pain - Ascending aortic dissection
- Neck or jaw pain - Aortic arch dissection
- Interscapular tearing or ripping pain - Descending aortic dissection
- Chest pain
- Myocardial infarction
- Neurologic symptoms
- Syncope
- Stroke symptoms
- Altered mental status
- Limb paresthesias, pain, or weakness
- Hemiparesis or hemiplegia
- Horner syndrome
- Dyspnea by congestive heart failure or tracheal or bronchial compression
- Dysphagia from compression of the esophagus
- Orthopnea
- Anxiety and premonitions of death
- Flank pain if renal artery is involved
- Dyspnea and hemoptysis if dissection ruptures into the pleura

BP

- Blood pressure may increase or decrease.
  - Hypertension may result from a catecholamine surge or underlying essential hypertension.
  - Hypotension is an ominous finding and may be the result of excessive vagal tone, cardiac tamponade, or hypovolemia from rupture of the dissection.
  - A blood pressure differential of greater than 20 mm Hg was an independent predictor of aortic dissection. A pressure differential of greater than 20 mm Hg should increase the suspicion of aortic dissection, but it does not rule it in. Significant interarm blood pressure differentials may be found in 20% of people without aortic dissection.

Neurologic deficits

- are a presenting sign in up to 20% of cases
  - The most common neurologic findings are syncope and altered mental status.
  - Syncope is part of the early course of aortic dissection in about 5% of patients and may be the result of increased vagal tone, hypovolemia, or dysrhythmia.
  - Other causes of syncope or altered mental status include strokes from compromised blood flow to the brain or spinal cord and ischemia from interruption of blood flow to the spinal arteries.
  - Peripheral nerve ischemia can manifest with numbness and tingling in the extremities.
  - Hoarseness from recurrent laryngeal nerve compression also has been described.
  - Horner syndrome is caused by interruption in the cervical sympathetic ganglia and presents with ptosis, miosis, and anhidrosis.
  - Superior vena cava syndrome, caused by compression of the superior vena cava from a large distorted aorta, may occur.

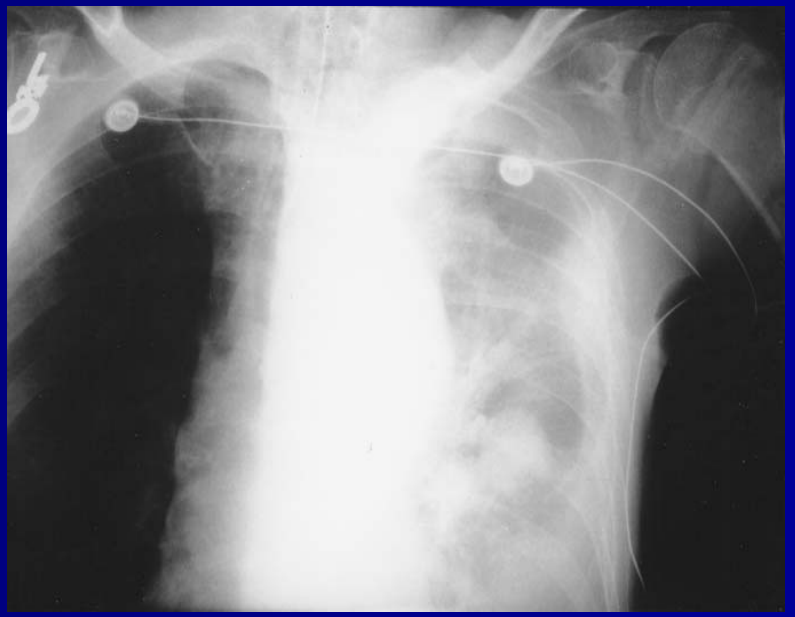
- Findings suggestive of cardiac tamponade, such as muffled heart sounds, hypotension, pulsus paradoxus, jugular venous distension, and Kussmaul sign, must be recognized quickly.
- Pay careful attention to carotid, brachial, and femoral pulses on initial examination and look for progression of bruits or development of bruits on reexamination.
- Physical findings of a haemothorax may be found if the dissection ruptures into the pleura.

Lab Studies

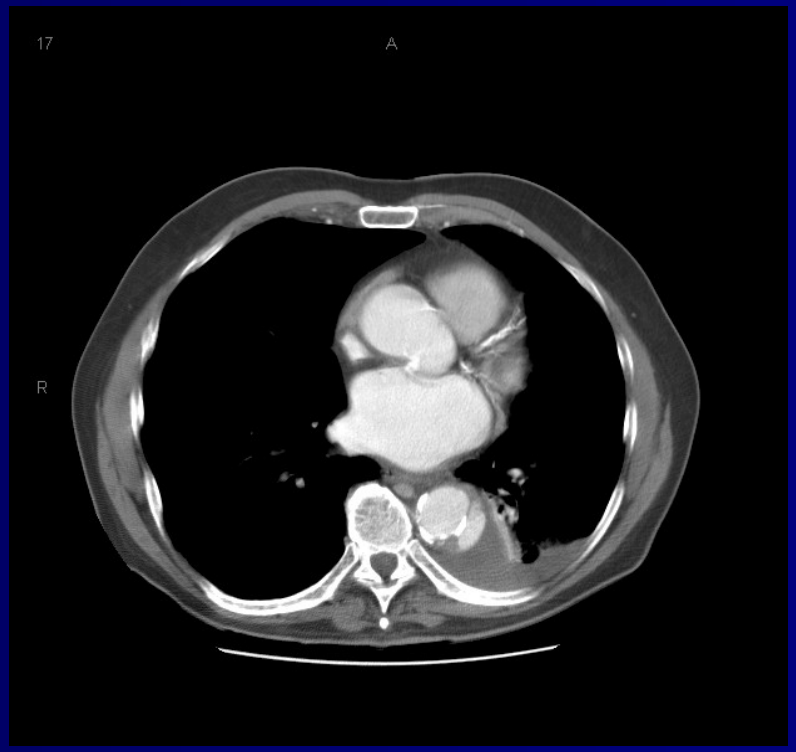
- Blood studies
  - Usually, the diagnosis is made before the blood work is returned; however, leukocytosis may be present.
  - BUN and creatinine are elevated if the dissection involves the renal arteries.
  - Troponin and creatine kinase (CK) can be elevated if the dissection has caused myocardial ischemia.
  - Decreases in the hemoglobin and hematocrit are ominous findings suggesting the dissection either is leaking or has ruptured.
  - Some studies suggest that D-dimer should be a part of the initial workup if aortic dissection is suspected. A negative result makes the presence of the disease unlikely.
  - Hematuria, oliguria, and even anuria (<50 mL/d) may occur if the dissection involves the renal arteries.

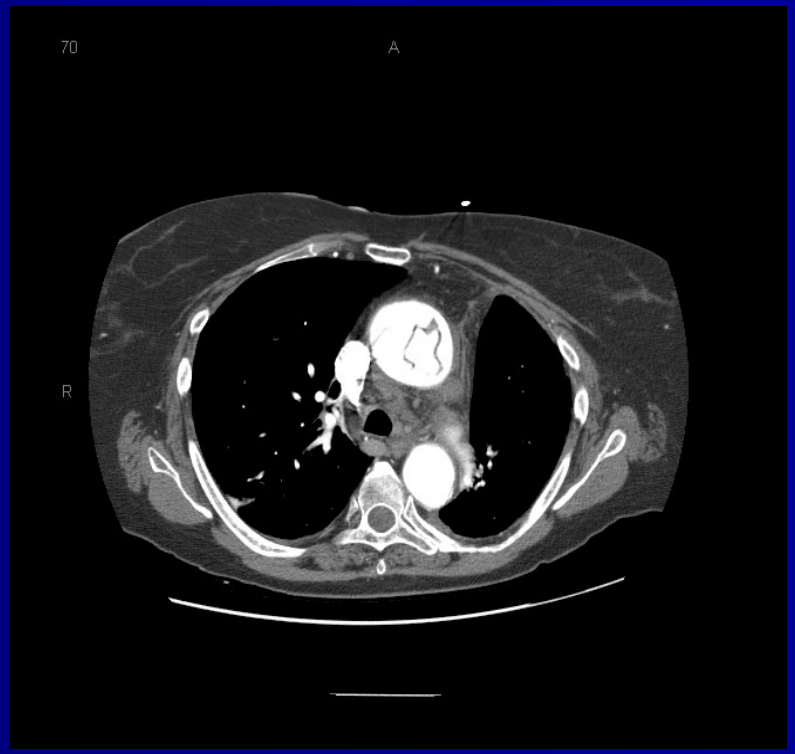
Imaging Studies





Imaging Studies





- Angiography: Still considered by some as the diagnostic criterion standard test for aortic dissection, it is being replaced by newer imaging modalities.

Echocardiography

- With its increasing acceptance and use in the ED, ultrasonography is becoming a valuable diagnostic aid, although transthoracic echocardiography (TTE) has a much lower sensitivity (80%) and lower specificity (90%) than angiography.
- Benefits include its rapid simple bedside use in the ED and its noninvasive nature.
- Drawbacks include the lack of sensitivity and specificity, especially with arch and descending aortic dissections, and dependence on operator experience.
- Transesophageal echocardiography (TEE) has greater sensitivity and specificity than TTE (in the range of 97-99% and 97-100%, respectively).

ECG

- ST elevation can be seen in Stanford type A dissections because the dissection interrupts blood flow to the coronary arteries.
- The incidence of abnormal ECG findings is greater in Stanford type A dissections than in other types of dissections.
- In one study, 8% of patients with type A dissections had ST elevation, while no patients with type B dissections had ST elevation.
- More commonly, the ECG abnormality is ST depression.
- If the dissection involves the coronary ostia, it is the right coronary artery that is most commonly involved, leading to inferior ST segment elevation pattern

Emergency Department Care

- The mortality rate of patients with aortic dissection is 1-2% per hour for the first 24-48 hours. Initial therapy should begin when the diagnosis is suspected. This includes 2 large-bore intravenous lines (IVs), oxygen, respiratory monitoring, and monitoring of cardiac rhythm, blood pressure, and urine output.
- Clinically, the physician should reassess the patient frequently for hemodynamic compromise, mental status changes, neurologic or peripheral vascular changes, and development or progression of carotid, brachial, and femoral bruits.
- Aggressive management of heart rate and blood pressure should be initiated.
- Beta-blockers should be given initially to reduce the rate of change of blood pressure (dP/dt) and the shear forces on the aortic wall.
- The target heart rate should be 60-80 beats per minute.
- The target systolic blood pressure should be 100-120 mm Hg.
- End organ perfusion should be evaluated. Balancing the risks of dP/dt on the aortic wall versus the benefits of acceptable end organ perfusion may be a difficult clinical decision.

Treatment

- Urgent surgical intervention is required in type A dissections.
  - The area of the aorta with the intimal tear usually is resected and replaced with a Dacron graft.
  - The operative mortality rate is usually less than 10%, and serious complications are rare with ascending aortic dissections.
  - The development of more impermeable grafts has greatly enhanced the surgical repair of thoracic aortic dissections.
  - With the introduction of profound hypothermic circulatory arrest and retrograde cerebral perfusion, the morbidity and mortality rates associated with this highly invasive surgery have decreased.
  - Dissections involving the arch are more complicated that those involving only the ascending aorta because the innominate, carotid, and subclavian vessels branch from the arch. Deep hypothermic arrest usually is required. If the arrest time is less than 45 minutes, the incidence of central nervous system complications is less than 10%.
  - Aortic stent grafting is a challenging technique. It may prove feasible and has offered good results in a small series of patients. It may be a reasonable alternative in high-risk patients in the near future.

Treatment

- The definitive treatment for type B dissections is less clear.
  - Uncomplicated distal dissections may be treated medically to control blood pressure.
  - Distal dissections treated medically have the same or a lower mortality rate as those treated surgically.
  - Surgery is reserved for distal dissections that are leaking, ruptured, or compromising blood flow to a vital organ.
  - Acute distal dissections in patients with Marfan syndrome usually are treated surgically.
  - Inability to control hypertension with medication is also an indication for surgery in those with a distal thoracic aortic dissection.
  - Patients with a distal dissection are usually hypertensive, emphysematous, or older.
  - Long-term medical therapy involves a beta-adrenergic blocker combined with other antihypertensive medications.
  - Aortic stent grafting may play a role in how type B dissections are treated in the near future

Prognosis

- On the basis of his experience, Crawford has stated that "no patient should be considered cured of the disease."
- The 5-year survival rate is about 75% whether the patient is treated medically or surgically.
- The 10-year survival rate is between 40% and 69% for both surgically and medically treated dissections.
- In the pretreatment era, the 1-year survival rate was 5-10%.
- Reoperation may be necessary for late complications.

Aortic Aneurysms

- Aneurysm is defined as dilatation of the aorta of greater than 150% of its normal diameter for a given segment. For the thoracic aorta, a diameter greater than 3.5 cm is generally considered dilated, whereas greater than 4.5 cm would be considered aneurysmal.
- Aneurysms may affect one or more segments of the thoracic aorta, including the ascending aorta, the arch, and the descending thoracic aorta. As many as 25% of patients with TAA also have an abdominal aortic aneurysm.

Pathophysiology

- Degenerative changes in the wall of the aorta lead to cystic medial necrosis. This causes damage to collagen and elastin, loss of smooth muscle cells, and increased amounts of basophilic ground substance in the medial (elastic) layer of the aorta.
- The ascending thoracic aorta is generally most affected by cystic medial necrosis, whereas a descending thoracic aneurysm is primarily a consequence of atherosclerosis.
- In Marfan syndrome, abnormalities of the gene encoding for the synthesis of fibrillin have been implicated in the predisposition to form aneurysms. Mutations in the gene responsible for this structural lipoprotein found in the aortic wall have been found in patients who do not have Marfan syndrome but have aneurysms.

- The cumulative risk of rupturing a TAA is related to aneurysm diameter. In a recent series of 133 patients with TAA, risk of rupture at 5 years was 0% for diameter less than 4 cm, 16% for diameter 4-5.9 cm, and 31% for aneurysms greater than 6 cm in diameter.
- **Race:** Thoracic aortic aneurysm is most common among whites.
- **Sex:** Men are affected 2-4 times more frequently than women.
- **Age:** The mean patient age at diagnosis is 60-70 years.

History

- Patients with TAA may be asymptomatic. Forty percent may be found incidentally during workup for other processes. Symptoms vary according to the size, location, and changes in the aneurysm. Chest, back, and abdominal pain are common symptoms in patients who are symptomatic.
- Aortic root dilatation may lead to symptoms of congestive heart failure (CHF) due to aortic insufficiency.
- Hoarseness may signify vagus or recurrent laryngeal nerve compression.

Physical

- The physical examination findings are usually normal.
- Ruptured thoracic aneurysm may cause hypotension, tachycardia, and shock.
- An early diastolic murmur may be heard in patients with aortic root dilatation causing aortic insufficiency.
- Wheezing or cough suggests compression of the trachea, and hemoptysis may be a sign of aneurysm erosion into the trachea.
- Dysphagia, hematochezia, or hematemesis may be caused by esophageal compression or aortoesophageal fistula.

Causes

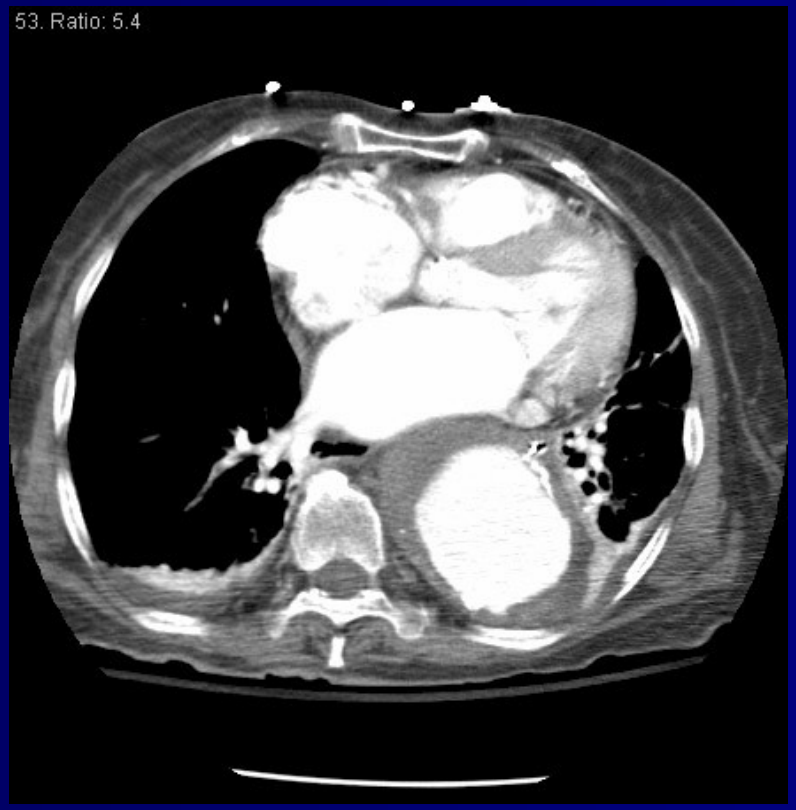
- Although atherosclerotic disease is often present in patients with TAA, it may only play a minor causal role in the pathogenesis of aneurysm development.
- Aortic aneurysm is often associated with smoking and hypertension.
- Marfan syndrome and Ehlers-Danlos syndrome are associated with an increased incidence of TAA and dilatation of the aortic root.
- Aortic aneurysm has been associated with a number of rheumatologic disorders, such as giant cell arteritis, Takayasu arteritis, and psoriatic arthritis.
- Syphilitic aortitis is an increasingly uncommon cause of thoracic aneurysm.

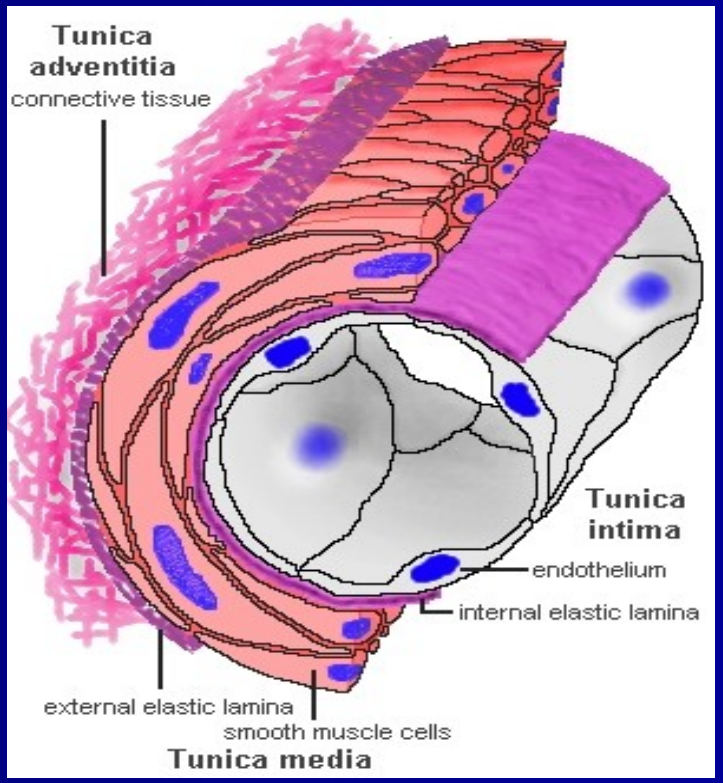
Lab Studies

- Hematocrit may be lowered in patients with a ruptured aneurysm.
- Coagulation studies may demonstrate coagulopathy.
- BUN and creatinine levels may be elevated in patients with shock and renal hypoperfusion.
- A blood bank sample should be ordered.
- Creatine kinase (CK) and troponin levels may be measured to assess for myocardial infarction.

Imaging Studies

- Chest radiography
  - mediastinal widening, blurring of the aortic knob, and tracheal displacement. Pleural effusion is usually associated with aortic dissection rather than with a stable aneurysm.
  - An elevated hemidiaphragm may suggest phrenic nerve compression from mass effect, but this finding is exceedingly rare compared with the other findings listed.
- Thoracic CT scanning
  - Intravenous contrast-enhanced CT scanning is the procedure of choice for diagnosis.
  - Its sensitivity is 96-100%, and its specificity is 99% for detecting aneurysms.
  - CT scanning is useful in evaluating aneurysm size, proximal and distal extension, presence or absence of dissection, and in seeking other pathology within the chest.
  - Use caution in patients with an allergy to the contrast agent or in those with renal failure.
  - Use caution in moving patients who are potentially unstable to the CT scanner.


53. Ratio: 5.4

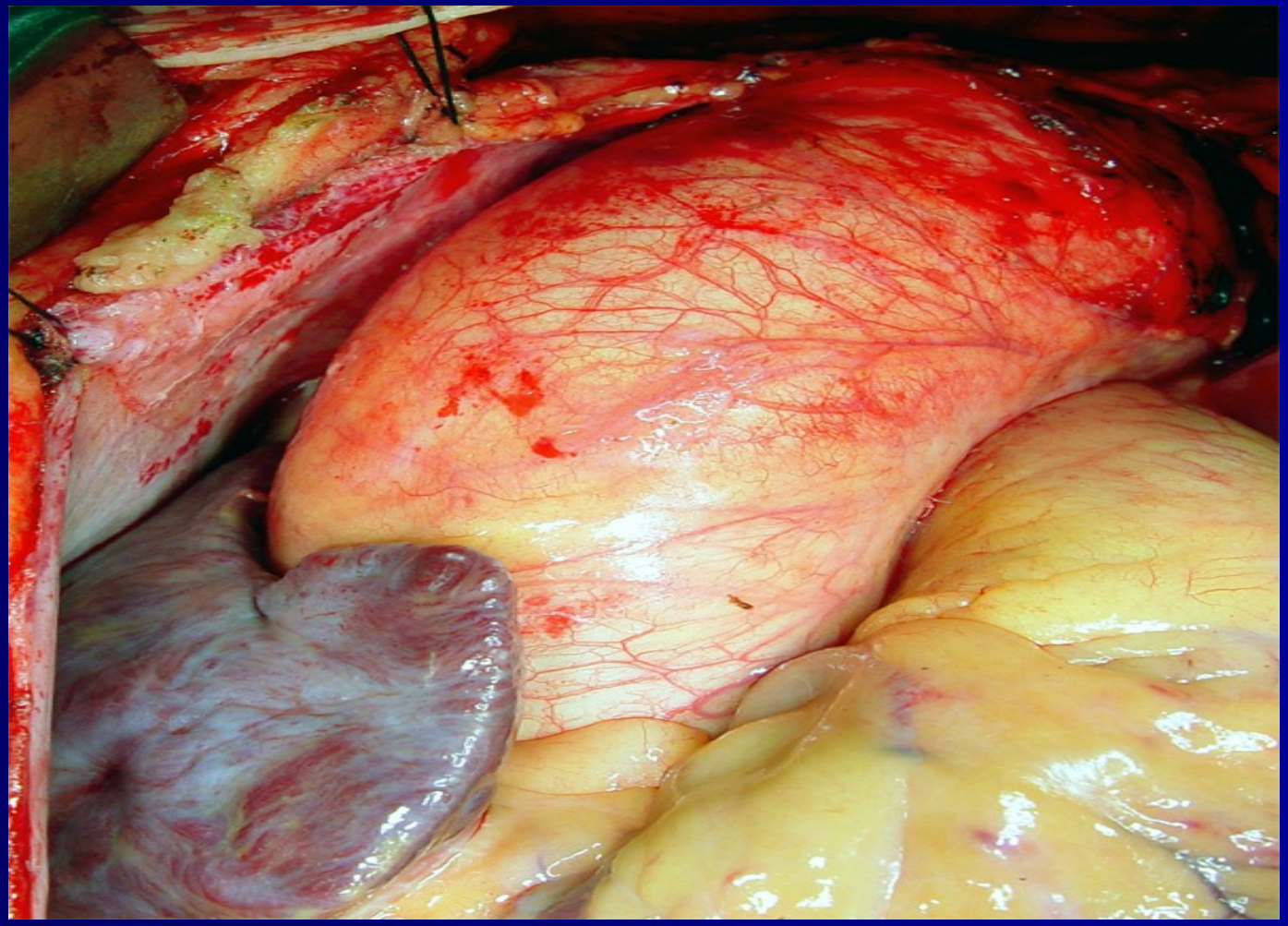

Tunica adventitia
connective tissue
Tunica intima
endothelium
internal elastic lamina
external elastic lamina
smooth muscle cells
Tunica media

Imaging Studies

- Contrast angiography
  - Contrast angiography is useful in assessing complex aortic pathology and identifying anatomy of branch vessels.
  - Its sensitivity is 85% and its specificity is 95% in detecting aneurysms.
- Magnetic resonance angiography
  - Magnetic resonance angiography is useful in assessing the aortic anatomy, the size of the aneurysm, the dissection, and the branch vessels.
  - Its sensitivity is 100% and its specificity is 100% in detecting aneurysms.
  - Magnetic resonance angiography does not require the administration of iodinated radiologic contrast material.
  - This study requires longer image acquisition times than other modalities.
- Transesophageal echocardiography
  - Transesophageal echocardiography is increasingly used to assess the anatomy of the aorta and its valves and the presence of dissection.
  - Its sensitivity is 98% and its specificity is 99% in depicting aneurysms.
  - Transesophageal echocardiography may be performed rapidly at the bedside.
  - The results are operator dependent.

Emergency Department Care

- Initial stabilization includes the following:
  - Placing 2 large-bore intravenous lines, administering 100% oxygen, and providing a cardiac monitor
  - Monitoring urine output
- Consider alternate diagnoses. Until the diagnosis of TAA is established, be vigilant for other causes of symptoms, such as myocardial infarction (MI), aortic insufficiency, CHF, or pulmonary embolus.
- Provide aggressive blood pressure control. Beta-blockers and nitrates are commonly used.
- For patients who are hemodynamically unstable, provide the following:
  - Aggressive fluid resuscitation (including blood products)
  - Placing an arterial line
  - Correction of coagulopathy
  - Immediate surgical consultation



Treatment

- Indications for surgical repair include the following:
  - Rupture
  - Acute dissection (ascending requires urgent intervention, whereas descending is managed medically or surgically, if vascular complications arise)
  - Symptomatic states, including pain, mediastinal organ compression, or aortic insufficiency severe enough to cause CHF or a dilated hypokinetic left ventricle
  - Rapid aneurysm growth rate
  - Absolute size (5.5 cm for ascending aortic aneurysm, 6.0 cm for descending aortic aneurysm; in patients with Marfan syndrome, 5.0 cm for ascending aortic aneurysm, 6.0 cm for descending aortic aneurysm)
- Surgical and other interventional options for TAA repair include the following:
  - Open approaches using cardiopulmonary bypass, hypothermia, and grafting
  - Endovascular stent grafting may be an option when TAA is limited to the descending aorta.
  - Complications of repair include paraplegia, renal failure, and intraoperative mortality.

Prognosis

- In a series of 370 patients with TAA, survival at 1, 5, and 10 years were found to be 88%, 69%, and 56%, respectively.