Amjad Bani Hani

Associate Prof. of Cardiac Surgery and Intensive Care

Thoracic Aortic Disease

TAD

Thoracic Aortic Aneurysm TAA

Acute Aortic Syndrome AAS

Thoracic Aortic Aneurysm TAA

- What is a TAA?
- Who is at risk and what causes TAA?
- Is TAA inherited? What about genetic testing?
- What are the signs and symptoms of TAA?
- How is TAA diagnosed?
- What is the medical treatment for TAA?
- How often does TAA need to be monitored?
- When is surgery recommended for TAA?
- What are the surgical options for TAA?

What’s a TAA?

- The aorta is the ultimate conduit, carrying, in an average lifetime, almost 200 million liters of blood to the body

Through its elasticity, the aorta has the role of a ‘second pump’ during diastole

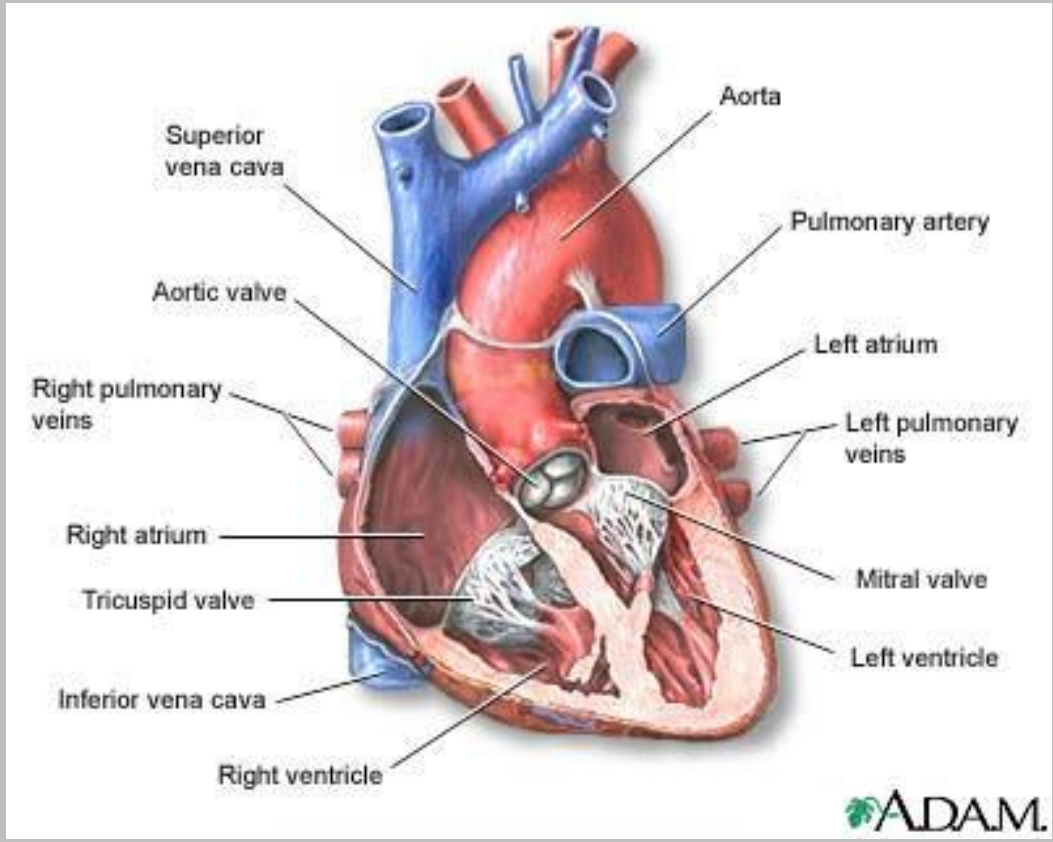
In healthy adults, aortic diameters do not usually exceed 40 mm and taper gradually downstream.

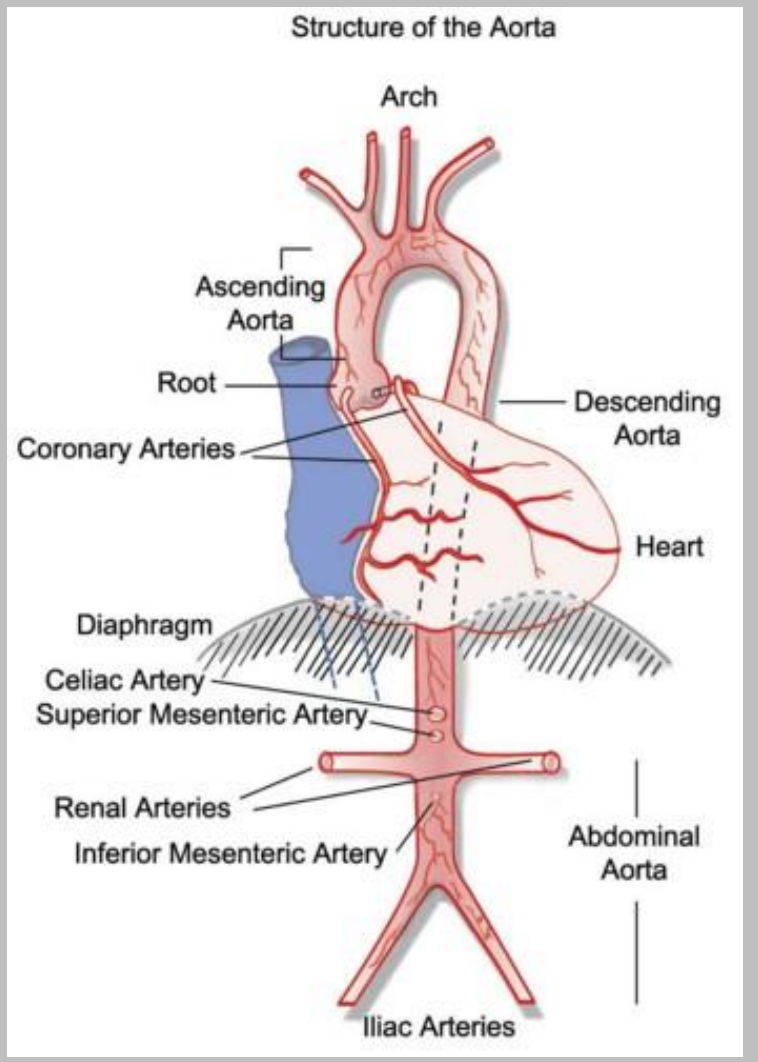
They are variably influenced by several factors including

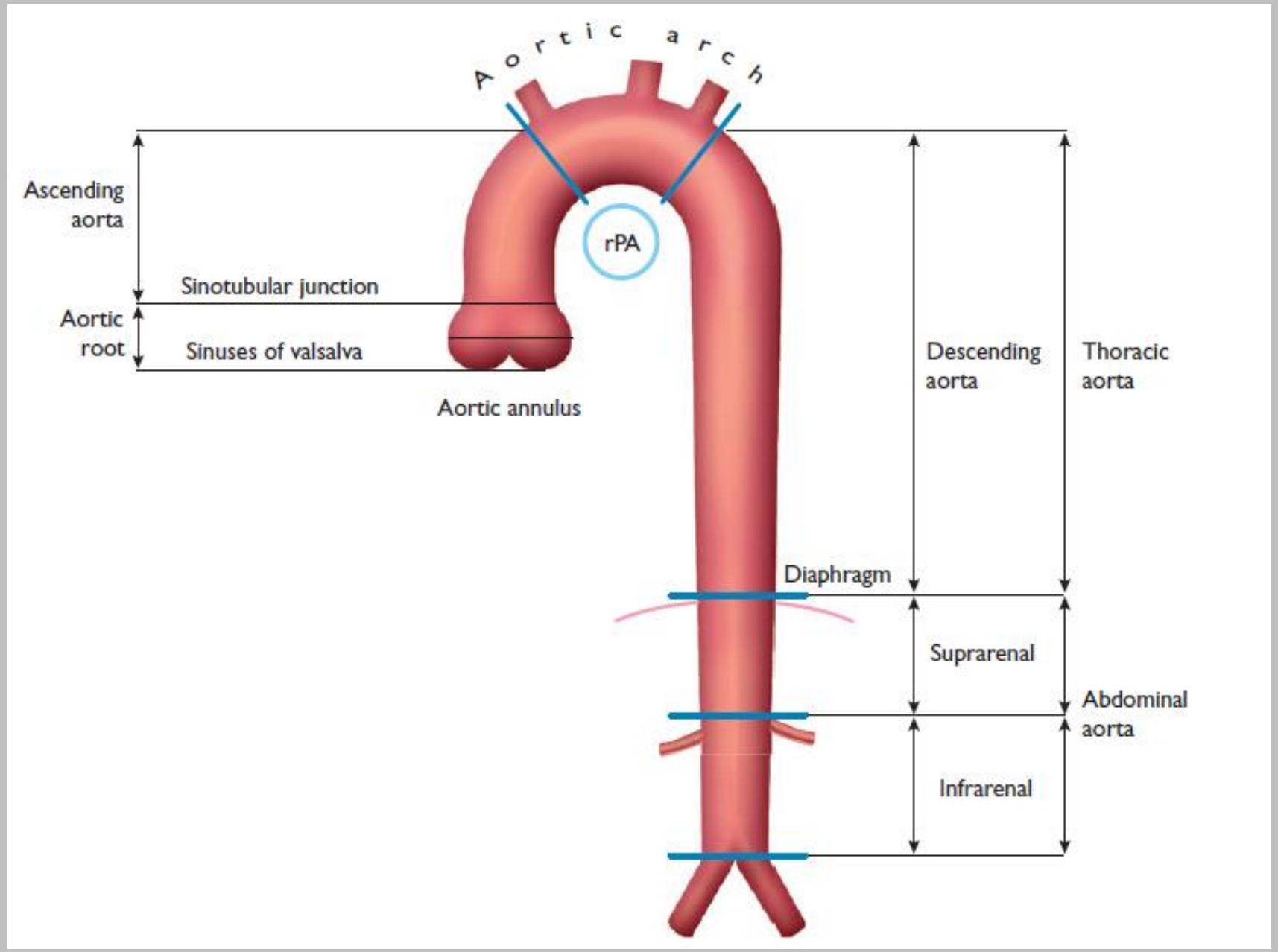
- age
- gender
- body size [height, weight, body surface area (BSA)]
- blood pressure

In this regard, the rate of aortic expansion is about 0.9 mm in men and 0.7 mm in women for each decade of life.

Anatomy


Superior vena cava
Aorta
Pulmonary artery
Aortic valve
Left atrium
Right pulmonary veins
Left pulmonary veins
Right atrium
Mitral valve
Tricuspid valve
Left ventricle
Inferior vena cava
Right ventricle
A.D.A.M.

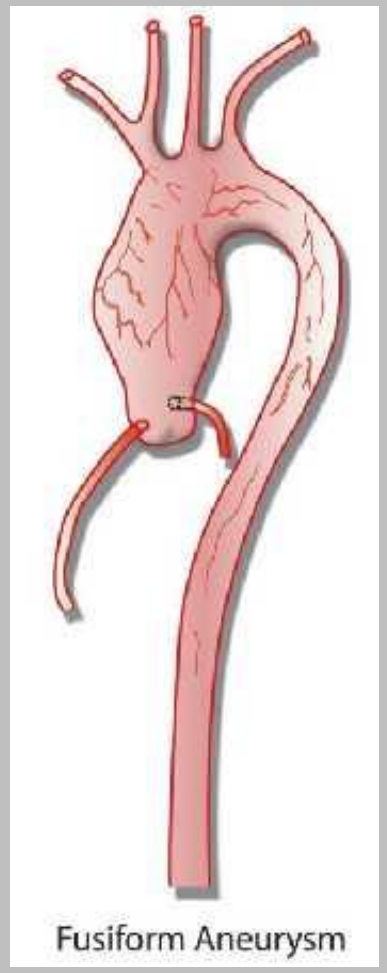

Structure of the Aorta
Arch
Ascending Aorta
Root
Descending Aorta
Coronary Arteries
Heart
Diaphragm
Celiac Artery
Superior Mesenteric Artery
Renal Arteries
Inferior Mesenteric Artery
Abdominal Aorta
Iliac Arteries

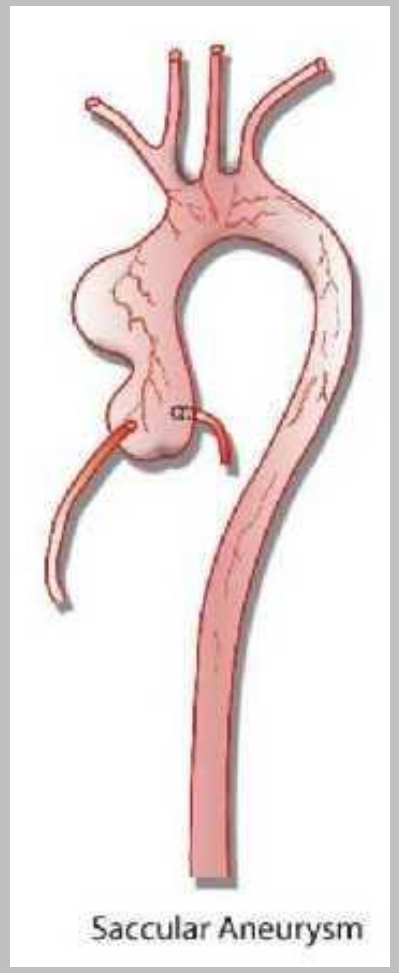

Aortic arch
Ascending aorta
Sinotubular junction
Aortic root
Sinuses of valsalva
Aortic annulus
rPA
Descending aorta
Thoracic aorta
Diaphragm
Suprarenal
Abdominal aorta
Infrarenal

Definitions

- Normal Dimensions
  - Mid-descending 26-28 mm
- Dilation (Ballooning, Bulging, Ectasia) up to 50%
- Aneurysm More than 50 %
    - **When the diameter exceeds 1.5 times normal**
  - Types
    - Saccular
    - Fusiform

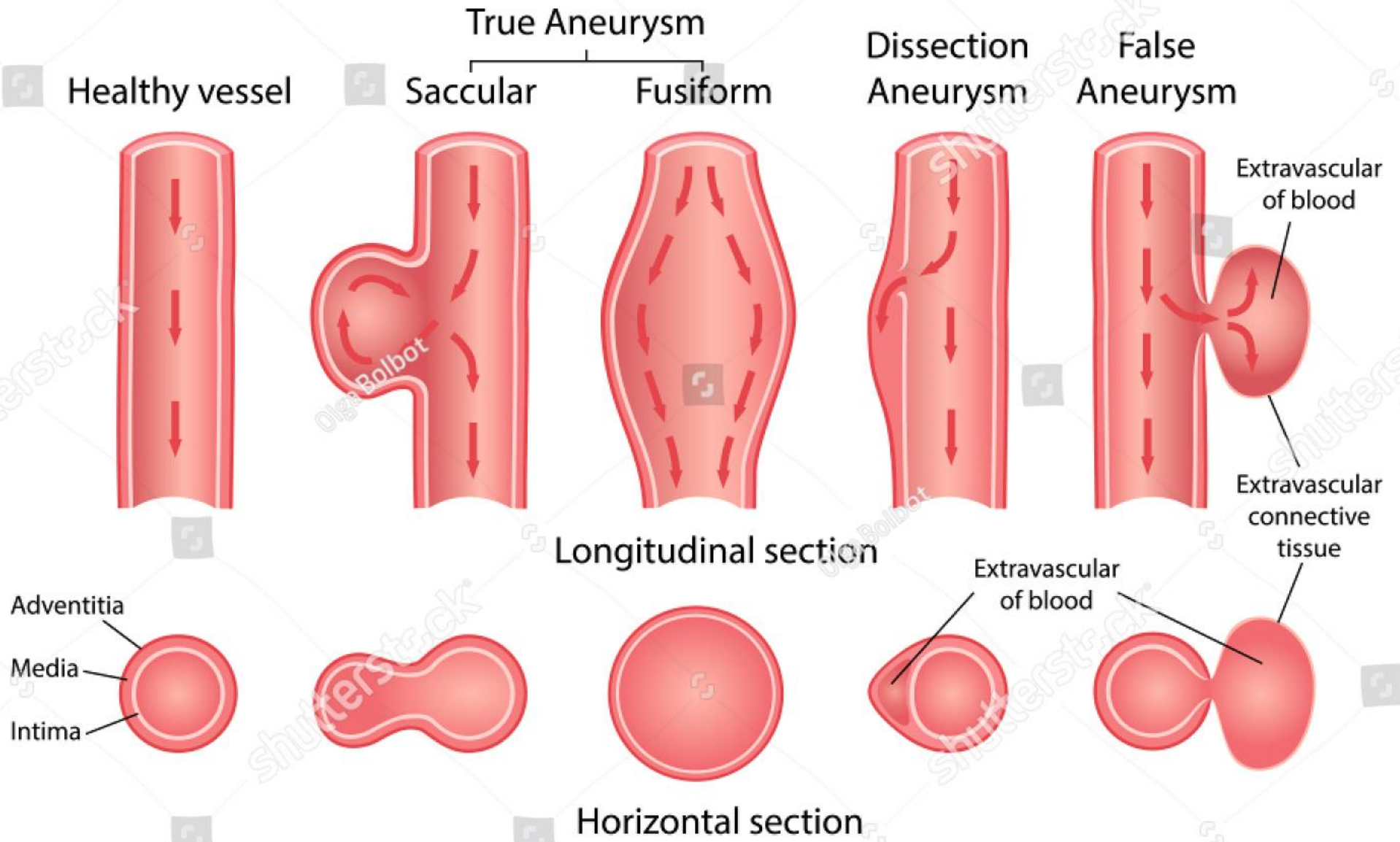
Aortic Aneurysm





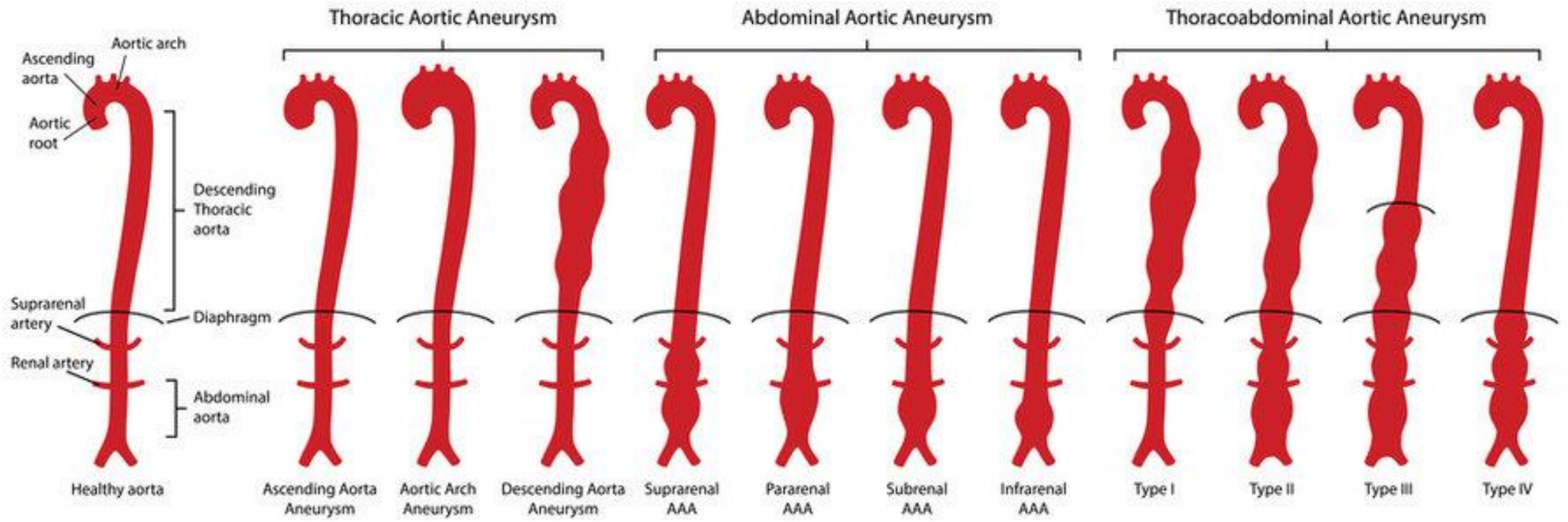
- true aneurysm means enlargement of the inner lumen caused by vessel wall expansion
- false aneurysm (also called pseudoaneurysm) means lumen enlargement caused by perforation (penetration) of all parts of the vessel wall forming an outer sack in communication with the inner lumen of the aorta

Types of Aneurysm




Aortic Arch
Ascending Aorta
Aortic Root

Classification of Aortic Aneurysm



Who is at risk and what causes TAA?

- The ageing of the aorta is accompanied by a loss of compliance, and an increase of wall stiffness caused by structural changes including an increase in the collagen content and formation of intimal atherosclerosis with calcium deposits.

- cystic medial degeneration, in which the elastic fibers in the wall of the aorta degenerate, weakening the wall of the aorta and causing it to dilate and form an aneurysm

- In younger patients, TAA is often due to a genetic cause

Table 1. Causes of TAA.

Acquired	Degenerative • Age • Hypertension • Smoking Autoimmune (inflammatory) • Takayasu's arteritis • Giant cell arteritis Infectious • Syphilis Traumatic
Genetic	Connective tissue disorders • Marfan syndrome • Loeys–Dietz syndrome • Ehlers–Danlos syndrome Familial TAA syndrome
Congenital	Bicuspid aortic valve

TAA, thoracic aortic aneurysm.

Table 1. Causes of TAA.

Risk Factors

- Smoking
- COPD
- HTN
- Male gender
- Older age
- High BMI
- Abnormal aortic valve (e.g., bicuspid valve)
- Family history

Epidemiology

- Thoracic aneurysms
  - Prevalence greater than 3-4% of those over 65
  - 6 cases per 100,000 person-years
  - Incidence increasing
  - In the top 15 causes of death
  - Thoracic aortic aneurysm – rupture 3.5/100,000 persons

Is TAA inherited? What about genetic testing?

- If Marfan syndrome is suspected based on physical features, then testing for the *FBN1* gene can be performed. Genetic testing can help to diagnose or rule out other conditions such as Loeys–Dietz and vascular Ehlers–Danlos syndromes

- First-degree relatives (parents, brothers, sisters, and children) of patients with TAA should be screened because family studies have found an approximately 20% chance of another first-degree relative having a TAA.

What are the signs and symptoms of TAA?

- Most people with TAA have no symptoms. TAAs are typically found incidentally when the patient is undergoing an imaging study for another reason. Aneurysms of the aortic root may lead to leakage of the aortic valve, so a heart murmur may be heard on physical examination.

Presentation

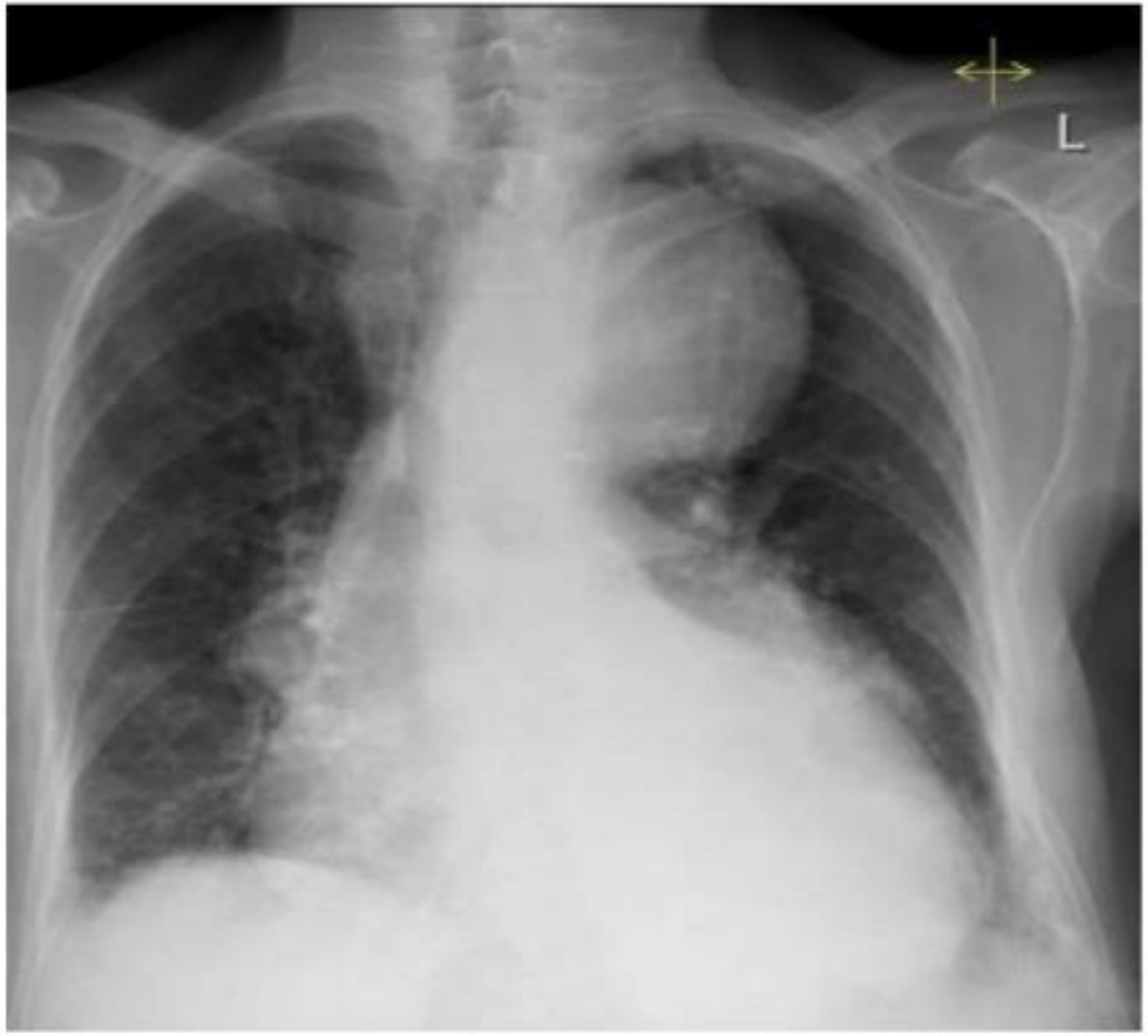
- Aneurysm
  - Most asymptomatic
  - Superior vena cava syndrome
  - Hoarseness
  - Bronchial obstruction
  - Dysphagia
  - Hemoptysis
  - Paralysis/paraplegia
  - Lower extremity embolism

Diagnosis

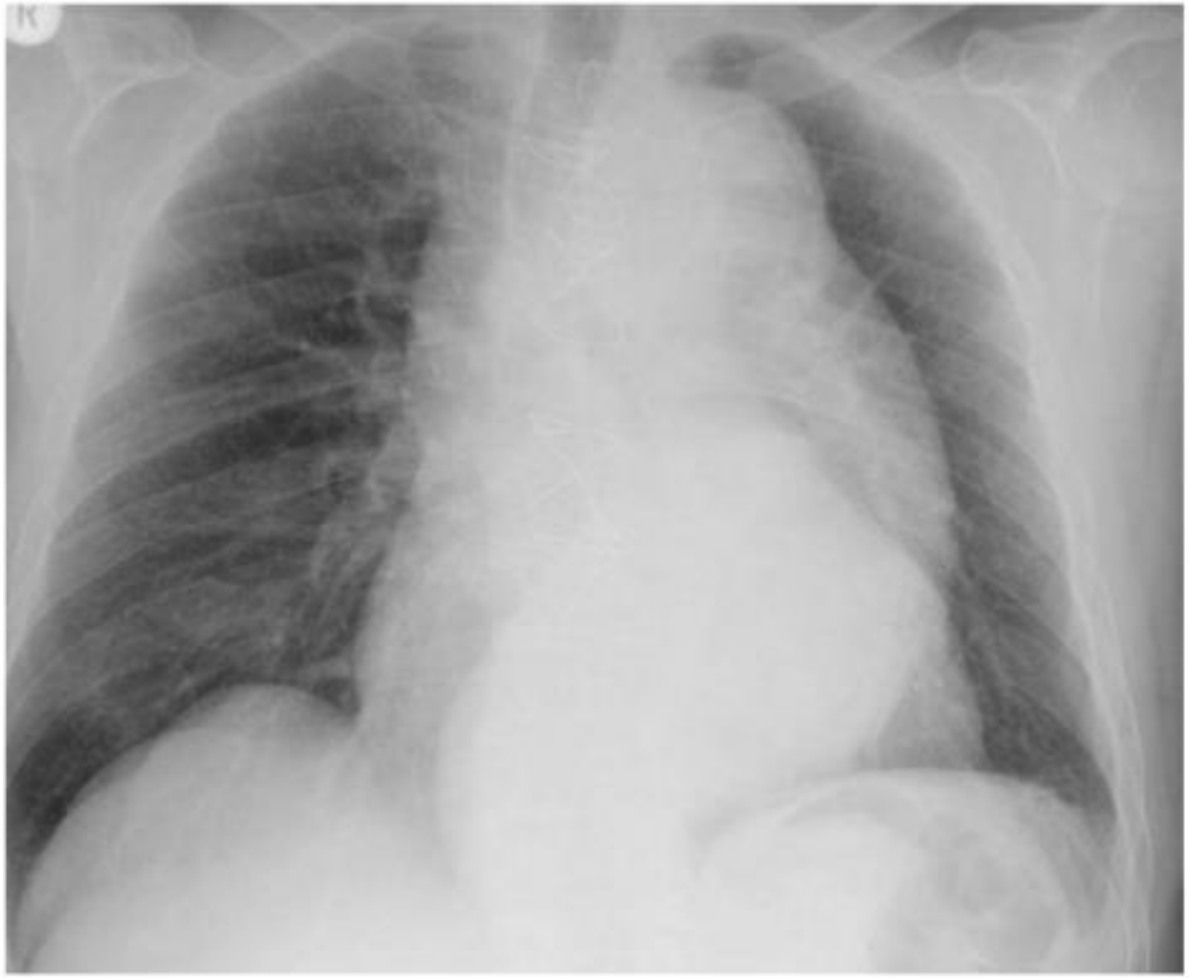
- Chest x-ray
  - Widened mediastinum
- Echocardiogram
  - Transthoracic – aortic root
  - Transesophageal – ascending and descending
- Aortography
  - Delineates the lumen
- CT scan
  - Most widely used diagnostic tool
- MRI
  - Avoids contrast dye

How is TAA diagnosed?

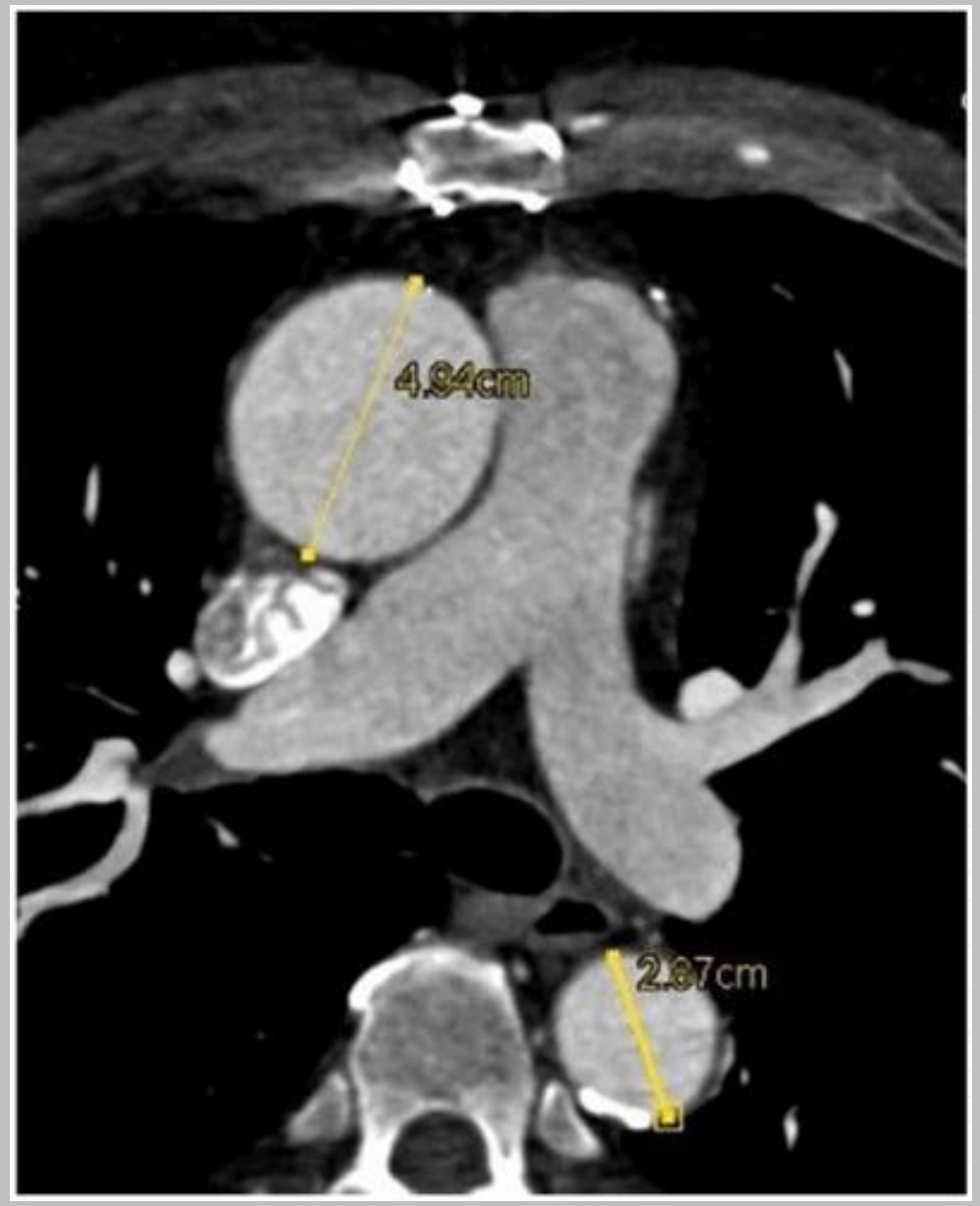
- (CTA)
- (MRA) are the imaging tests of choice for diagnosing and measuring TAAs.
- Disadvantages of CTA include radiation exposure and the need for using intravenous (IV) contrast dye
- Disadvantages of MRA include the length of the test (45–60 minutes), the use of gadolinium for contrast,

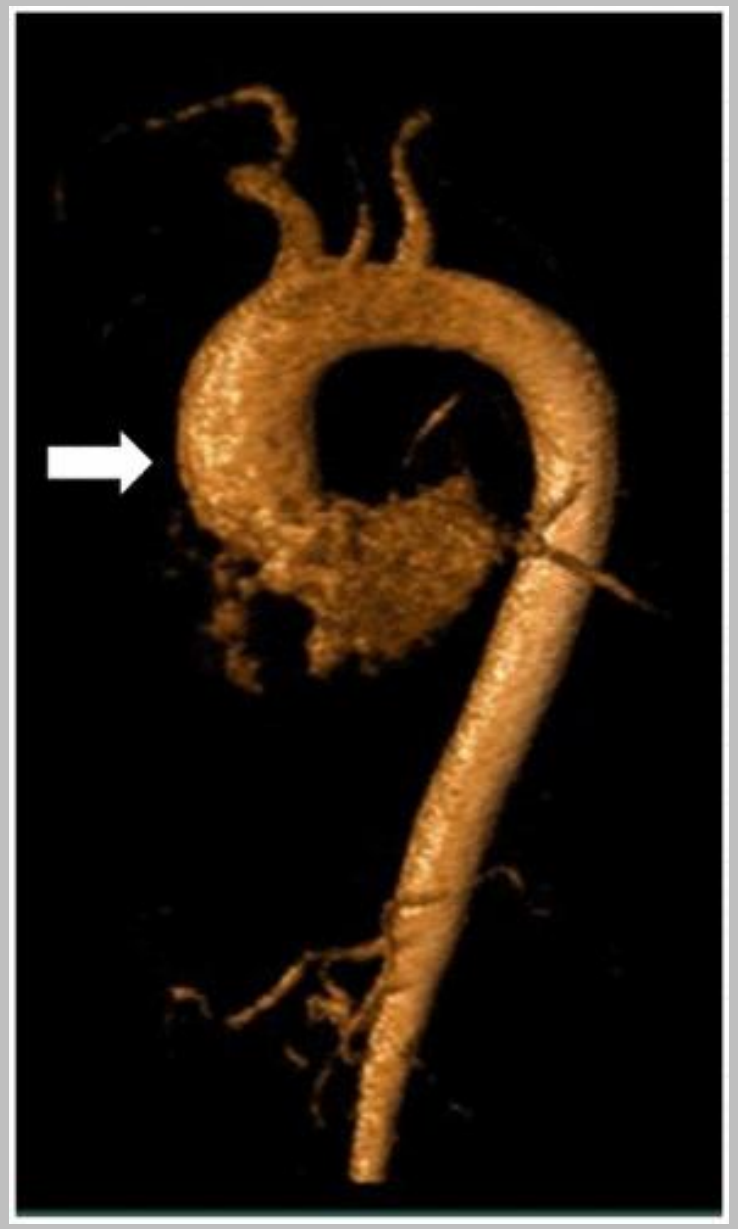


TAA. This huge aneurysm of the aortic arch and descending thoracic aorta was an incidental finding on this chest radiograph

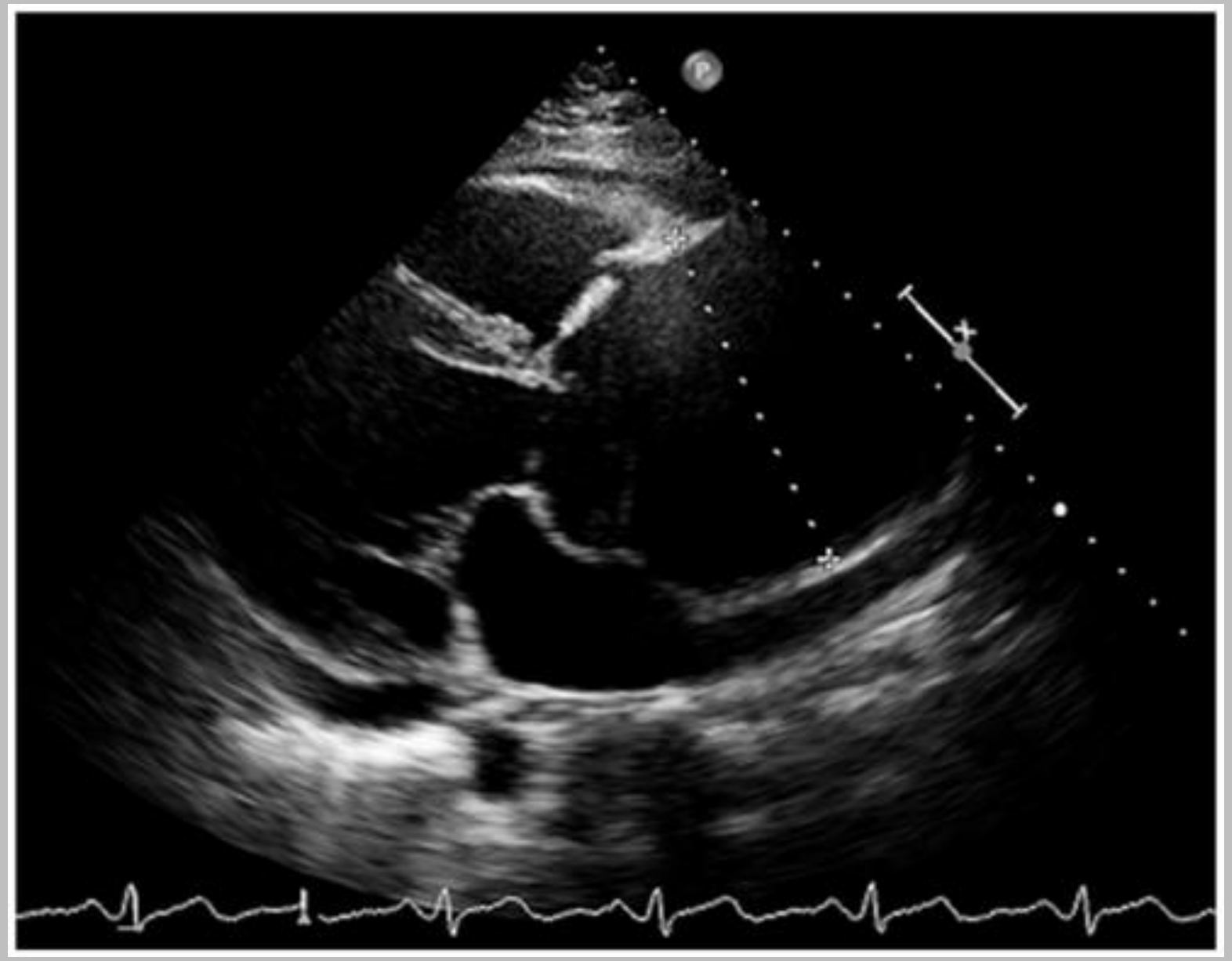


Chest X-ray (postero-anterior projection) showing that the descending thoracic aorta is extremely dilated and tortuous.


4.94cm
2.87cm



- Echocardiography may help in the diagnosis of TAA.
- Echo is useful to look at the aortic root but may miss aneurysms farther away from the heart in the ascending aorta and aortic arch.



What is the medical treatment for TAA?

- In general, medications to slow the growth of TAAs and prevent dissection and rupture are limited

Treatment - Aneurysm

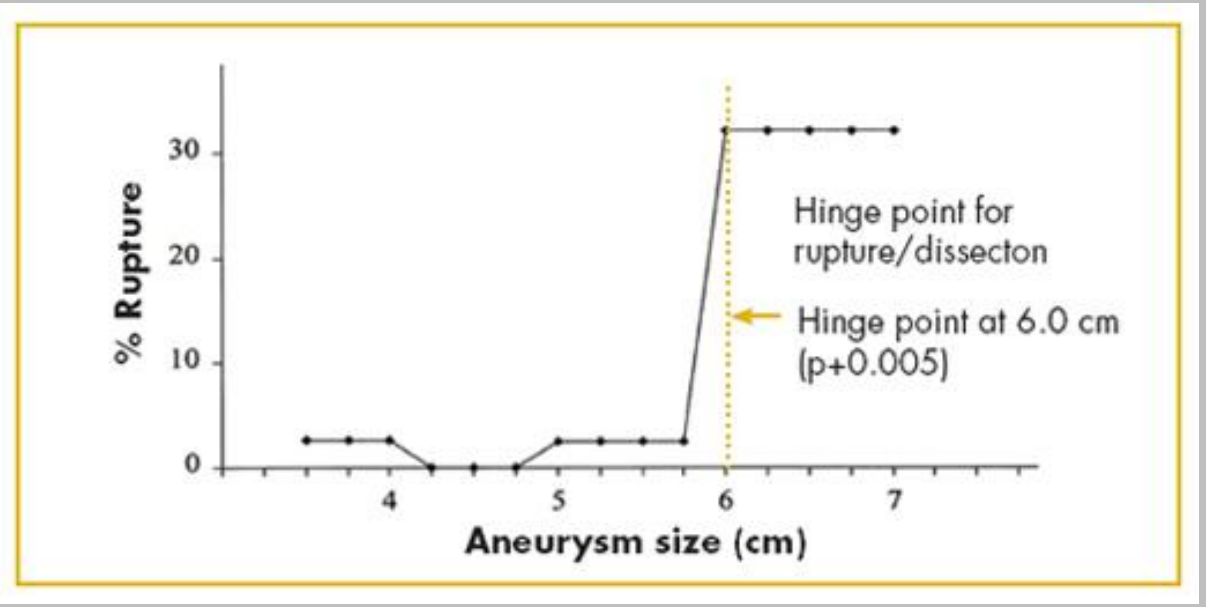
- Medical
  - BP control
  - Smoking cessation
  - No heavy lifting
- Surgical
  - Dacron tube graft
  - Ascending – may need to replace valve
  - Arch – graft
  - Descending – graft, stent grafts

Treatment – Indications for Intervention

- Aortic size
  - Ascending diameter ≥5.5 cm
  - Growth rate ≥1 cm/yr (avg ascending 0.07 cm/yr; descending 0.19 cm/yr)
- Symptomatic aneurysm
- Traumatic rupture
- Pseudoaneurysm
- Large saccular aneurysm
- Mycotic aneurysm
- Aortic coarctation
- Bronchial compression
- Aortobronchial or aortoesophageal fistuala

The annual risk of rupture or dissection

- 2% for TAAs < 5 cm in diameter
- 3% for TAAs 5–5.9 cm
- 7% for TAAs > 6 cm
- surgery is recommended for TAAs ⩾ 5.5 cm in most cases.



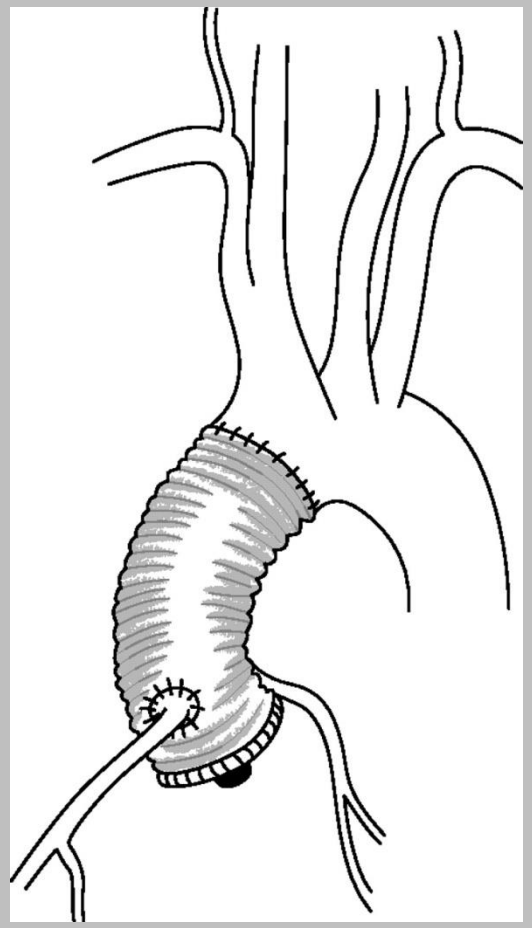
When is surgery recommended for TAA?

- 4.5 cm for patients with Marfan syndrome and 4.0 cm for patients with Loeys–Dietz syndrome.
- For patients with bicuspid aortic valve, lower threshold of 5.0 cm

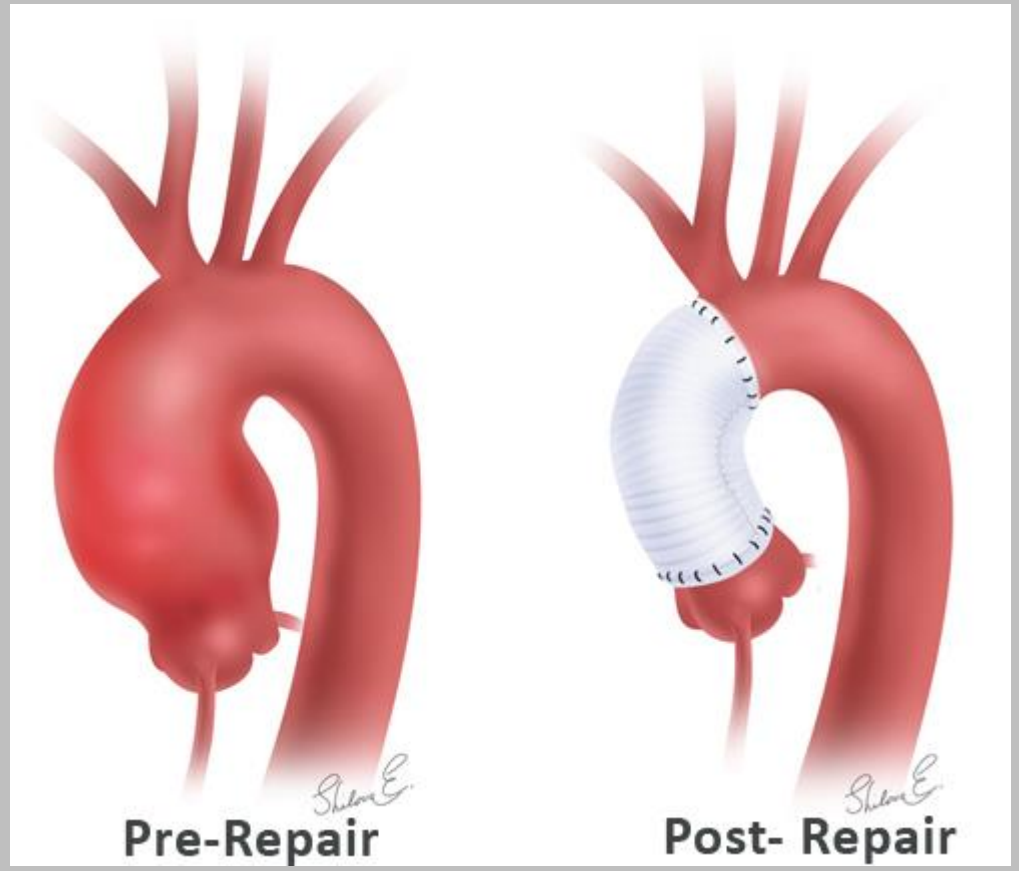
Treatment - Surgical

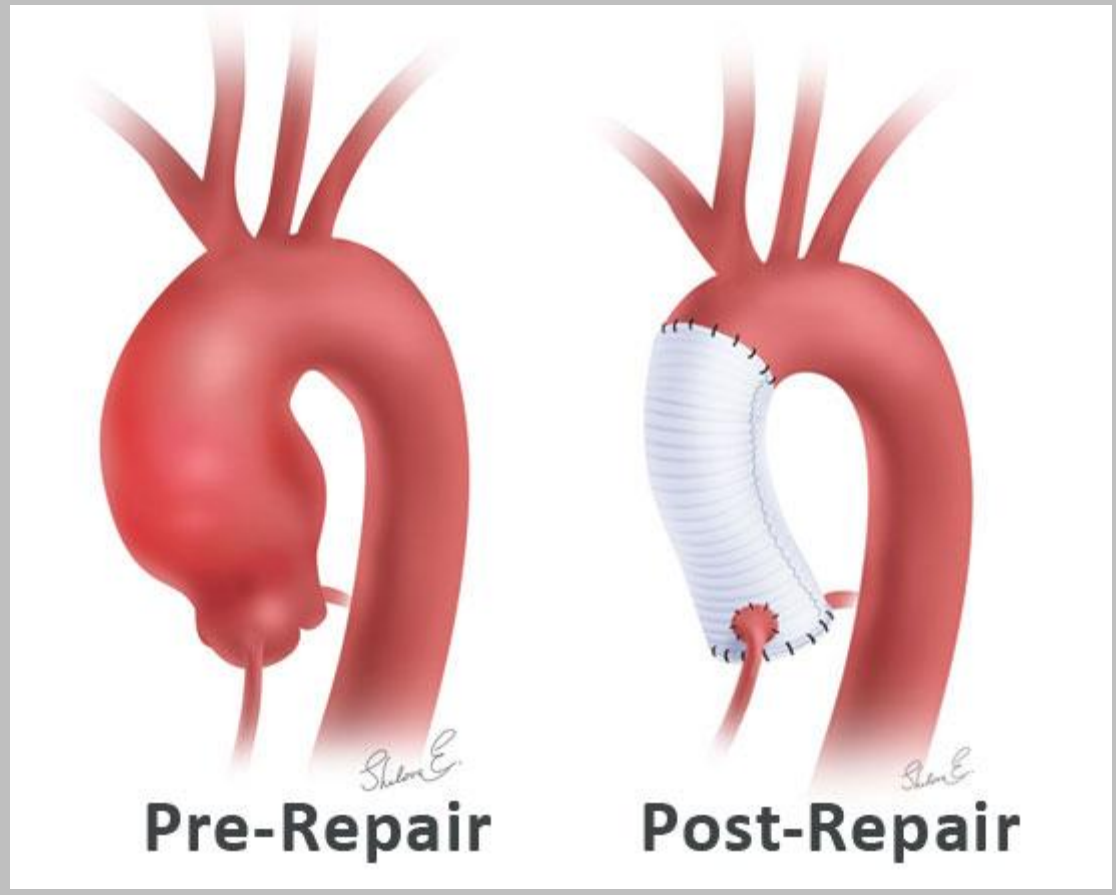
.

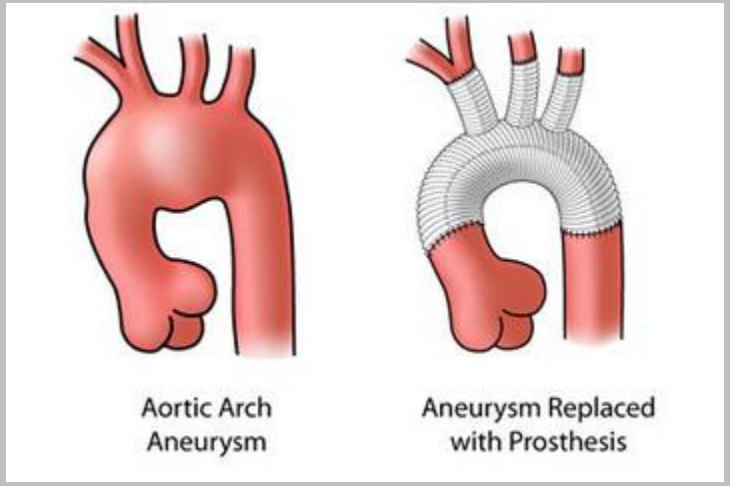
- Composite valve and graft replacement

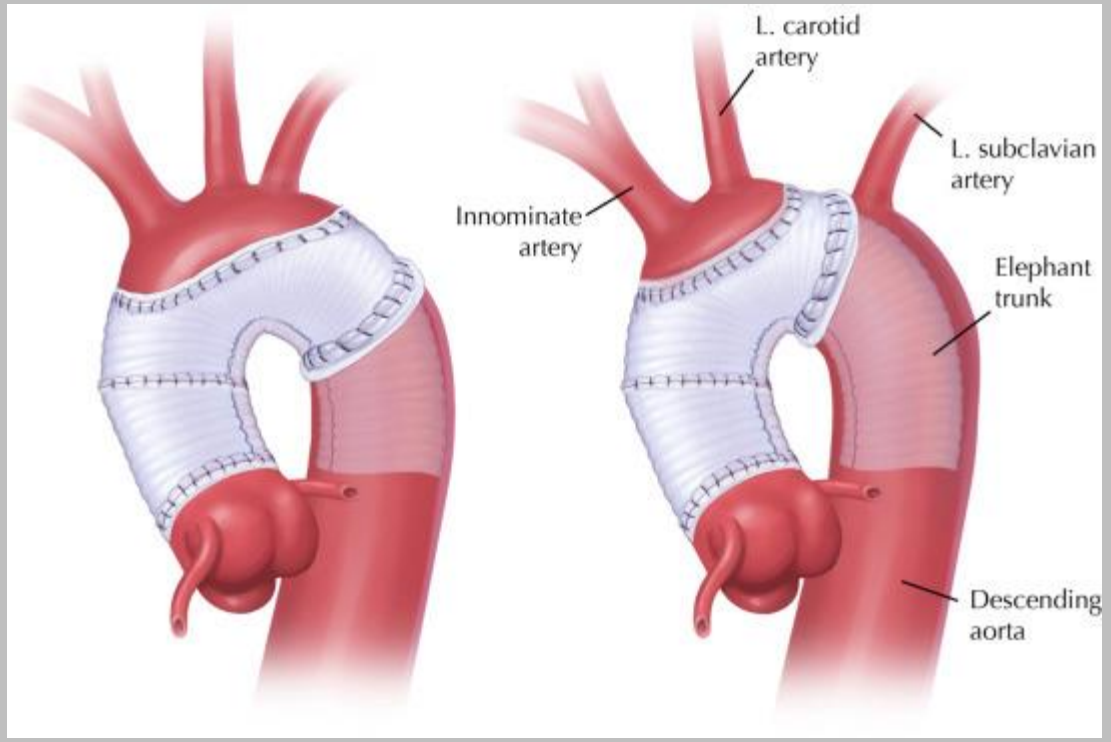


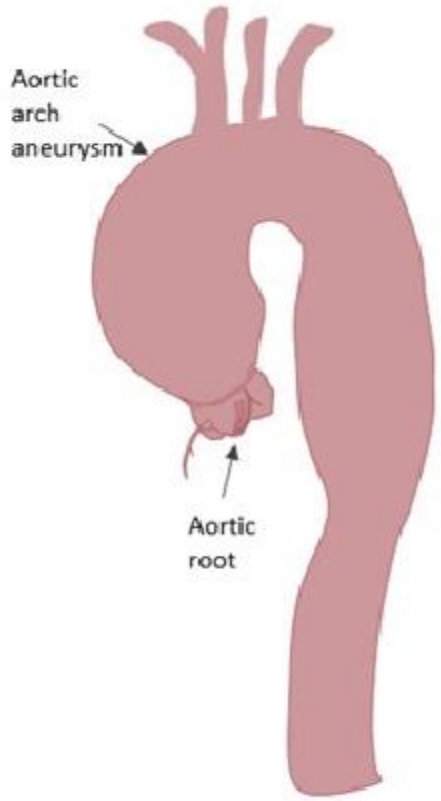
Nataf P , Lansac E Heart 2006;92:1345-1352


Pre-Repair
Post- Repair

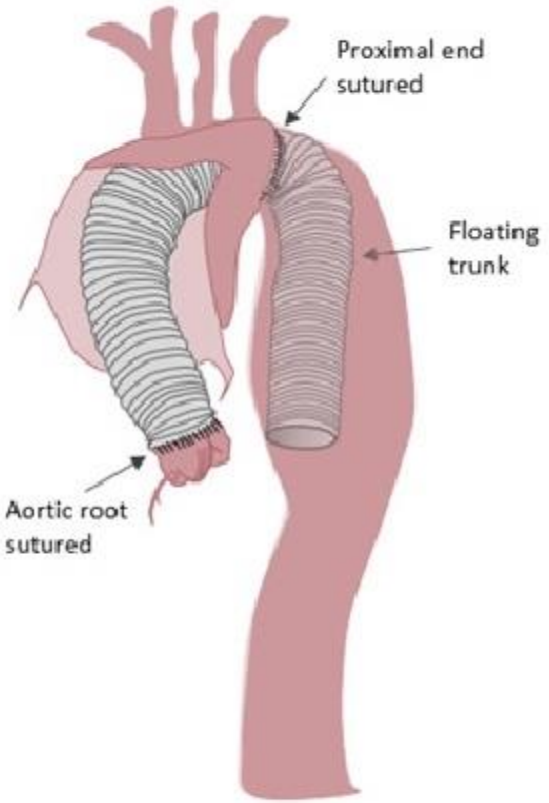

Pre-Repair
Post-Repair


Aortic Arch
Aneurysm
Aneurysm Replaced
with Prosthesis

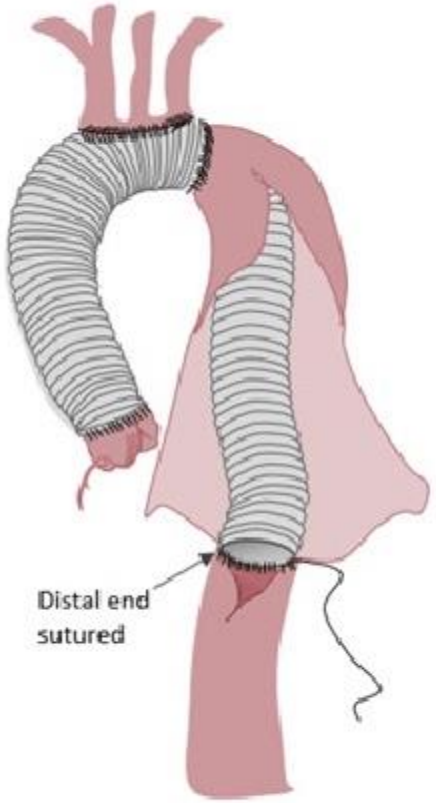

L. carotid
artery
L. subclavian
artery
Innominate
artery
Elephant
trunk
Descending
aorta



COMPLEX ANEURYSM

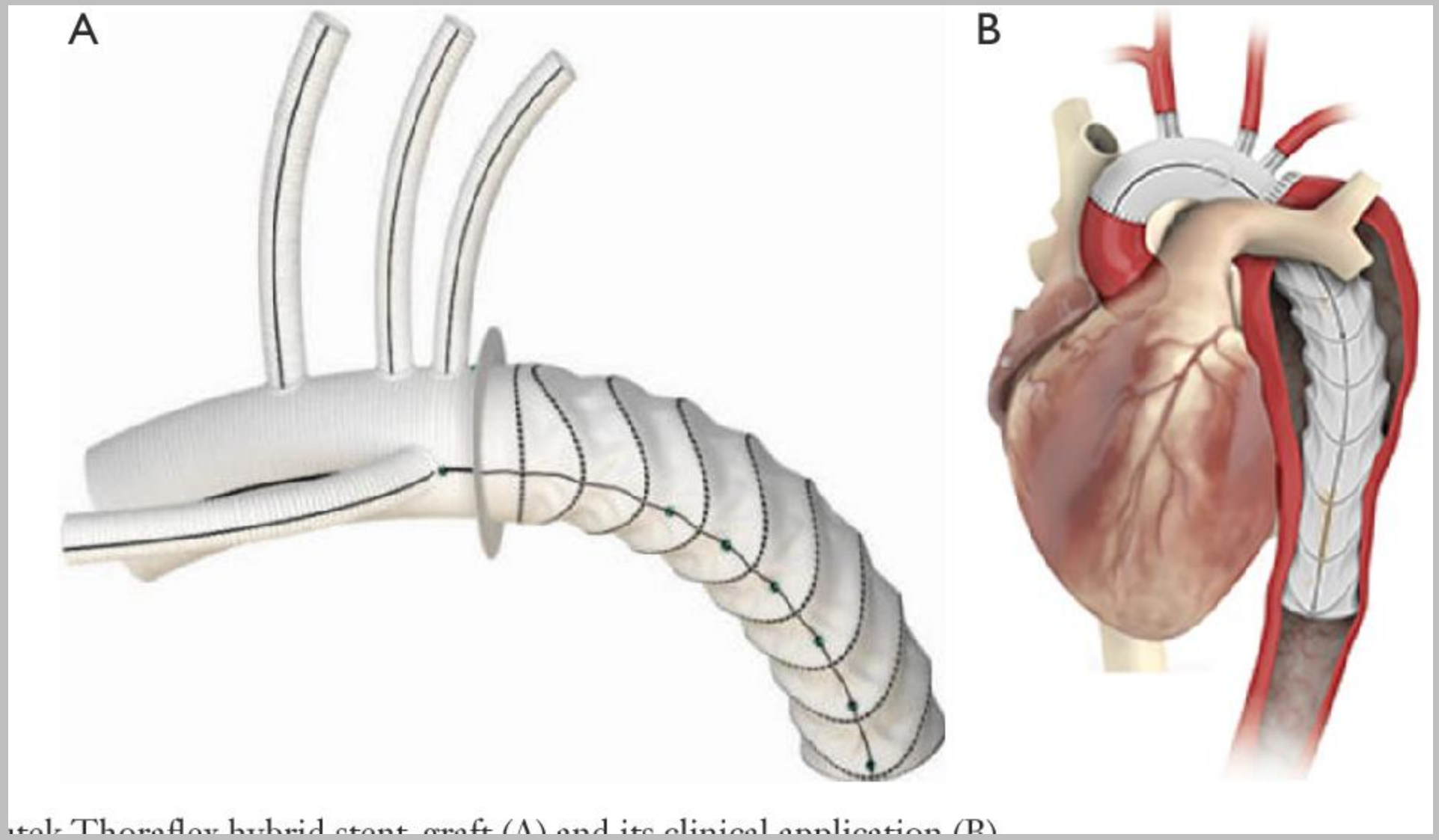


STAGE - 1

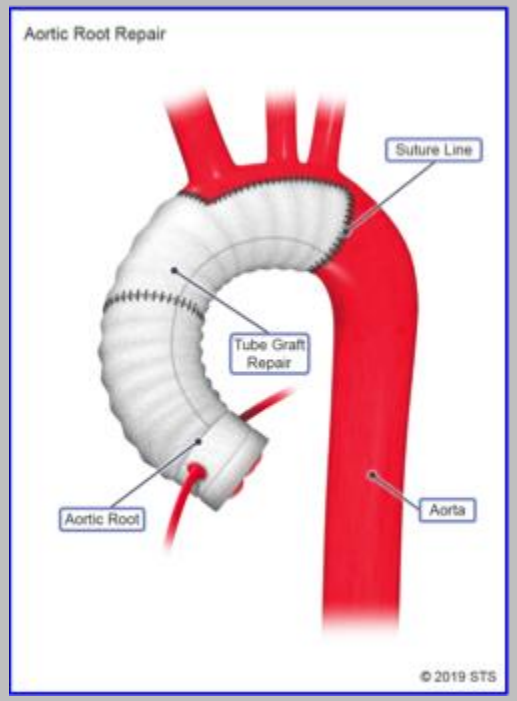


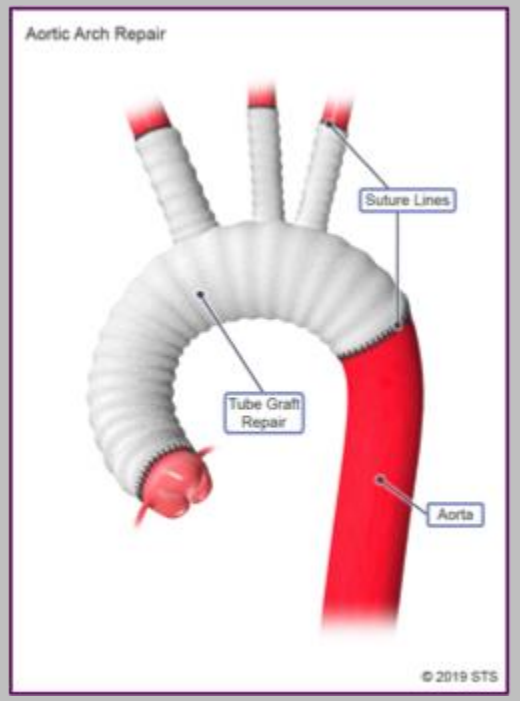
STAGE - 2

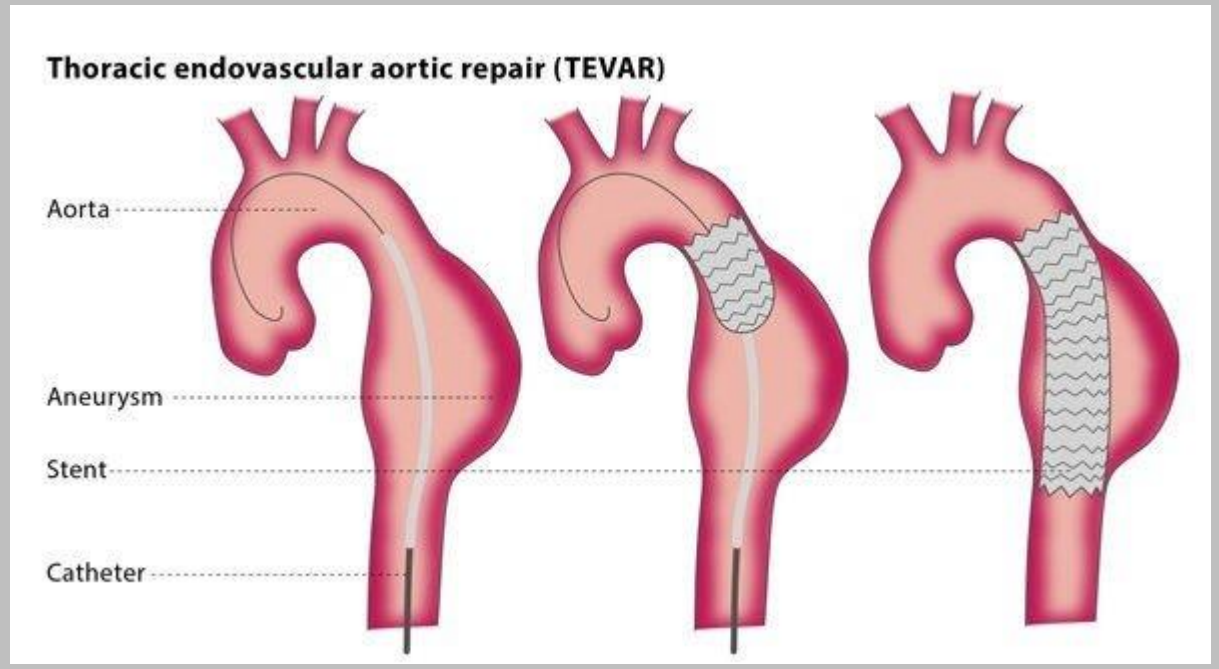
COMPLETED REPAIR

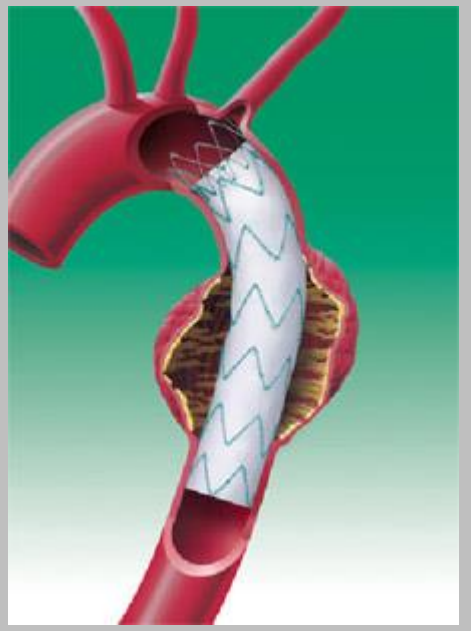


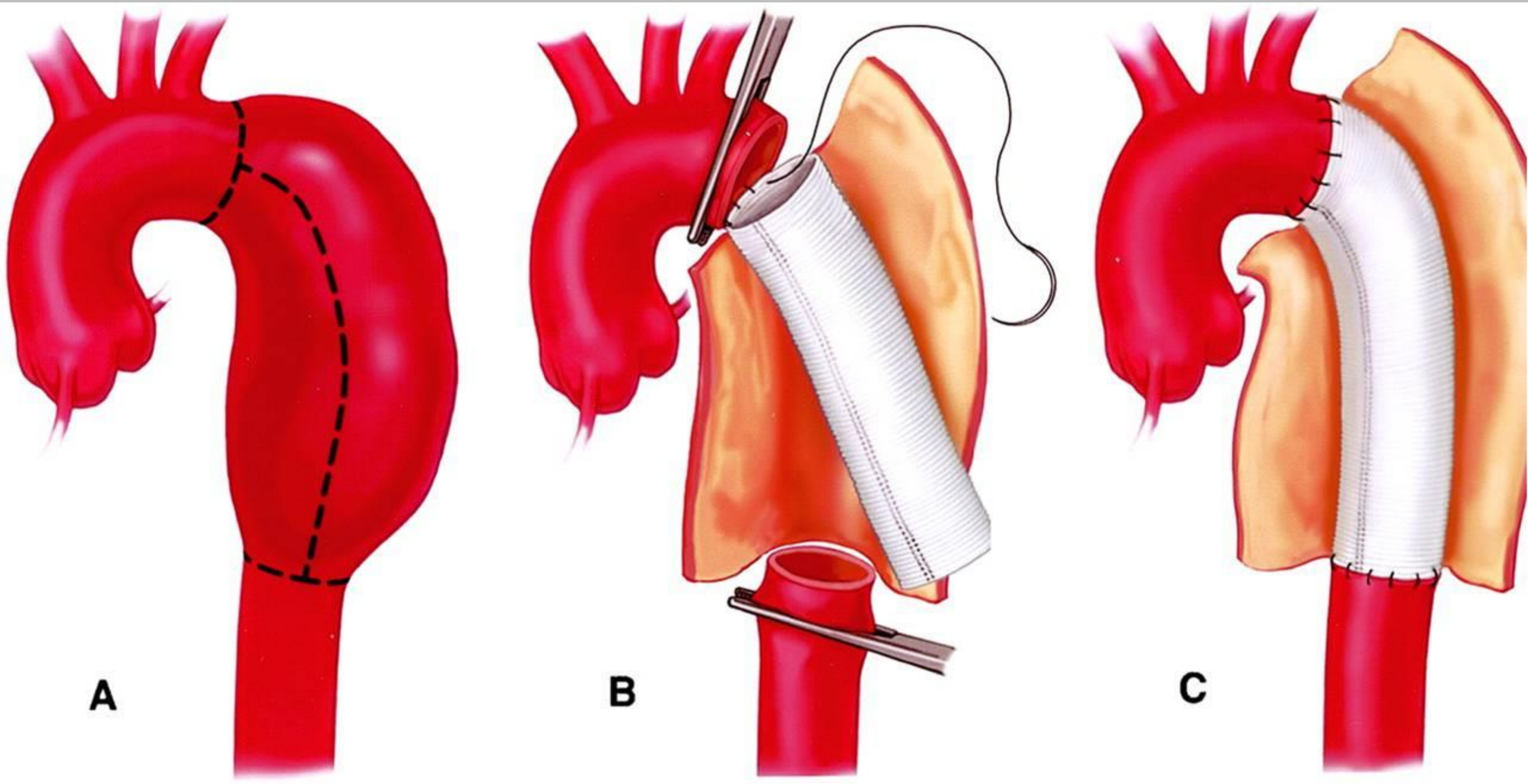
tek Thoraflex hybrid stent-graft (A) and its clinical application (B)

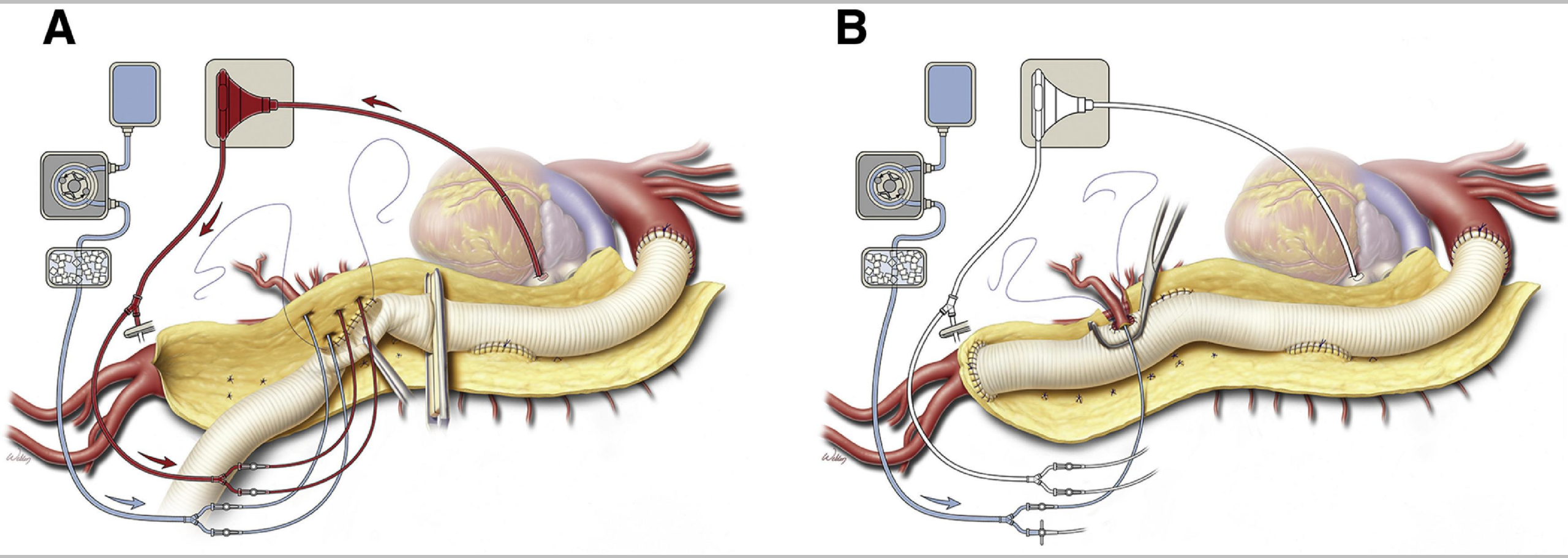

Aortic Root Repair
Suture Line
Tube Graft Repair
Aortic Root
Aorta
© 2019 STS


Aortic Arch Repair
Suture Lines
Tube Graft Repair
Aorta
© 2019 STS


Thoracic endovascular aortic repair (TEVAR)
Aorta
Aneurysm
Stent
Catheter

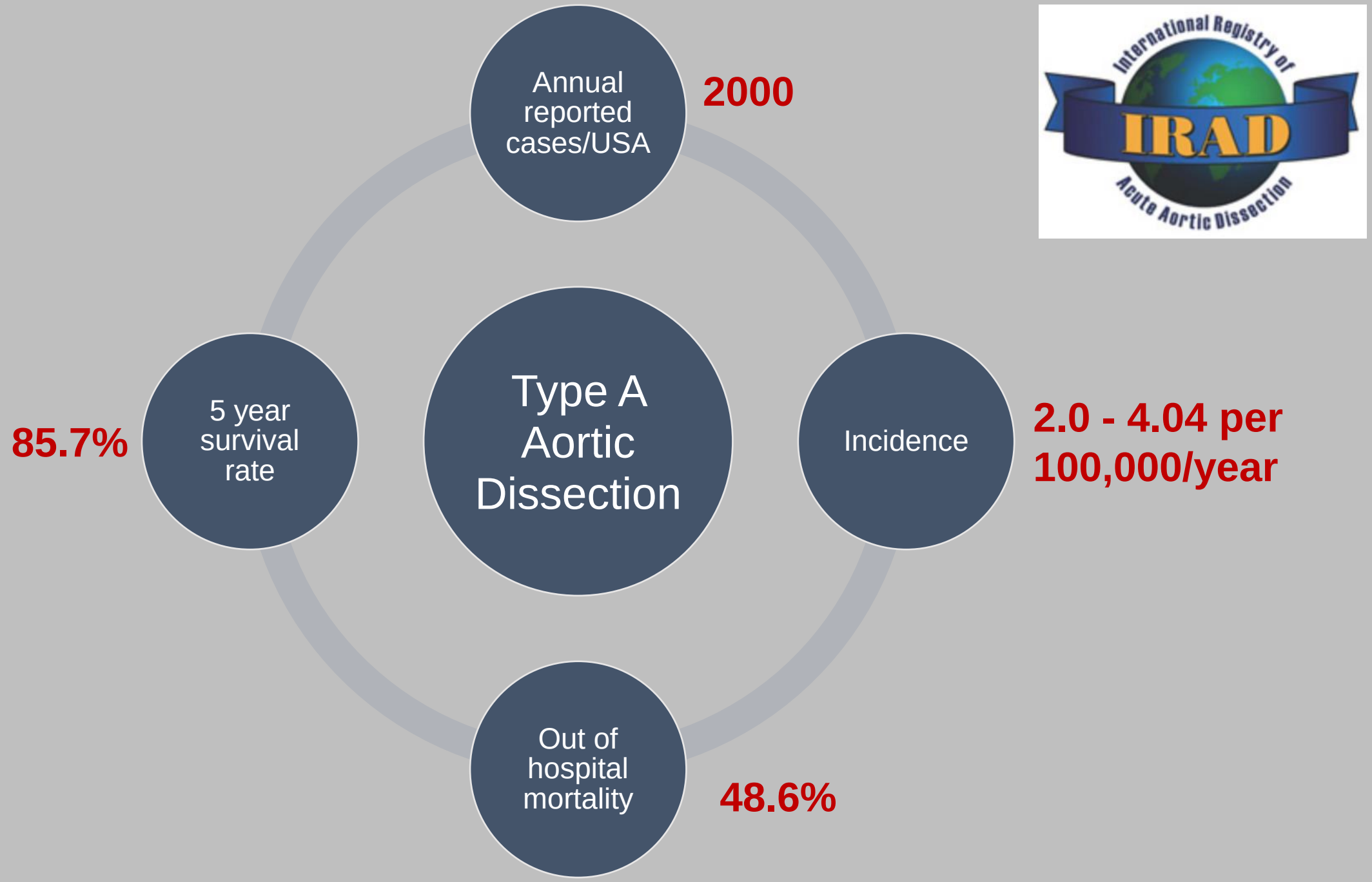



A
B
C

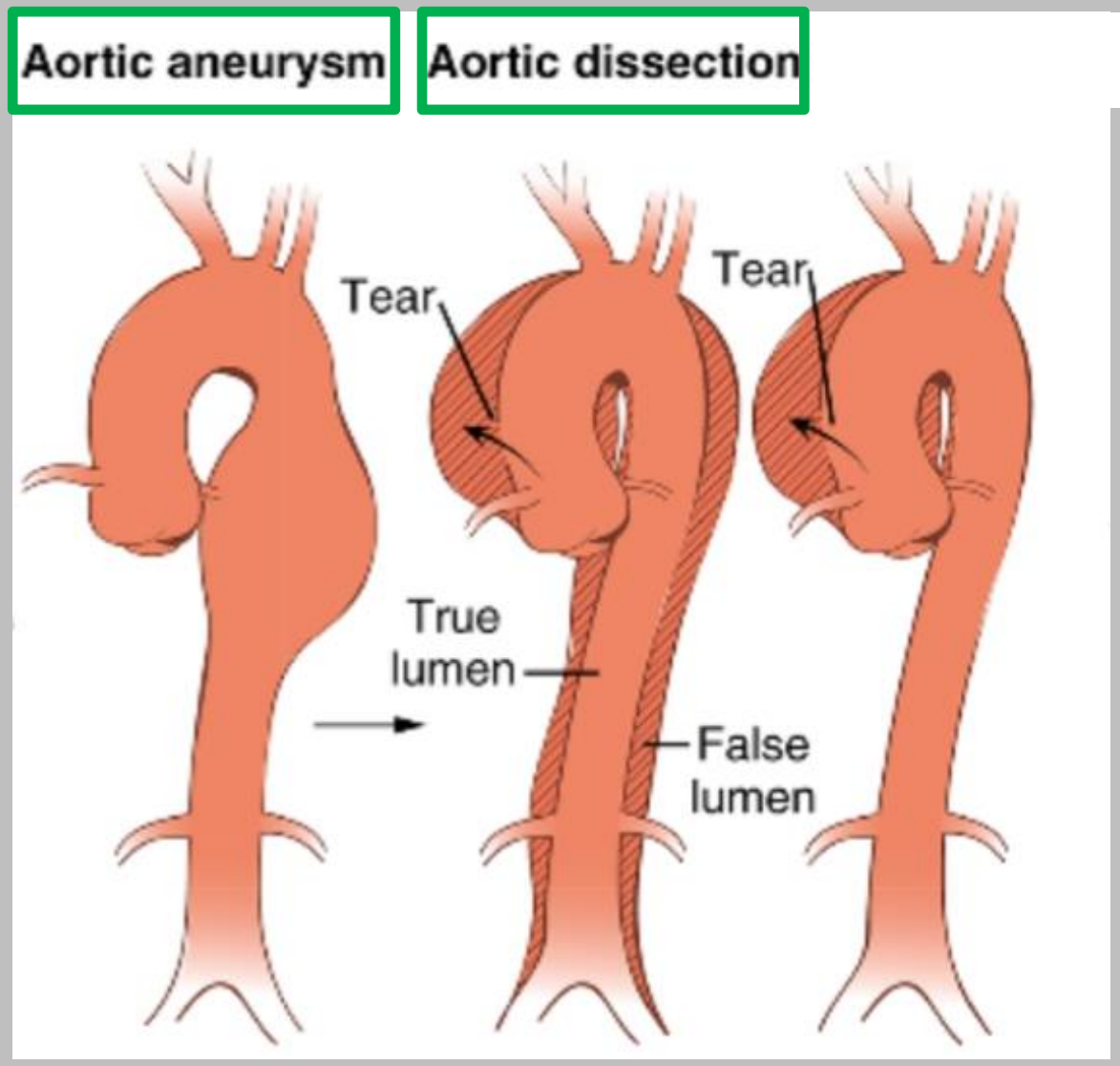

A
B

Acute Aortic Syndrome (AAS)

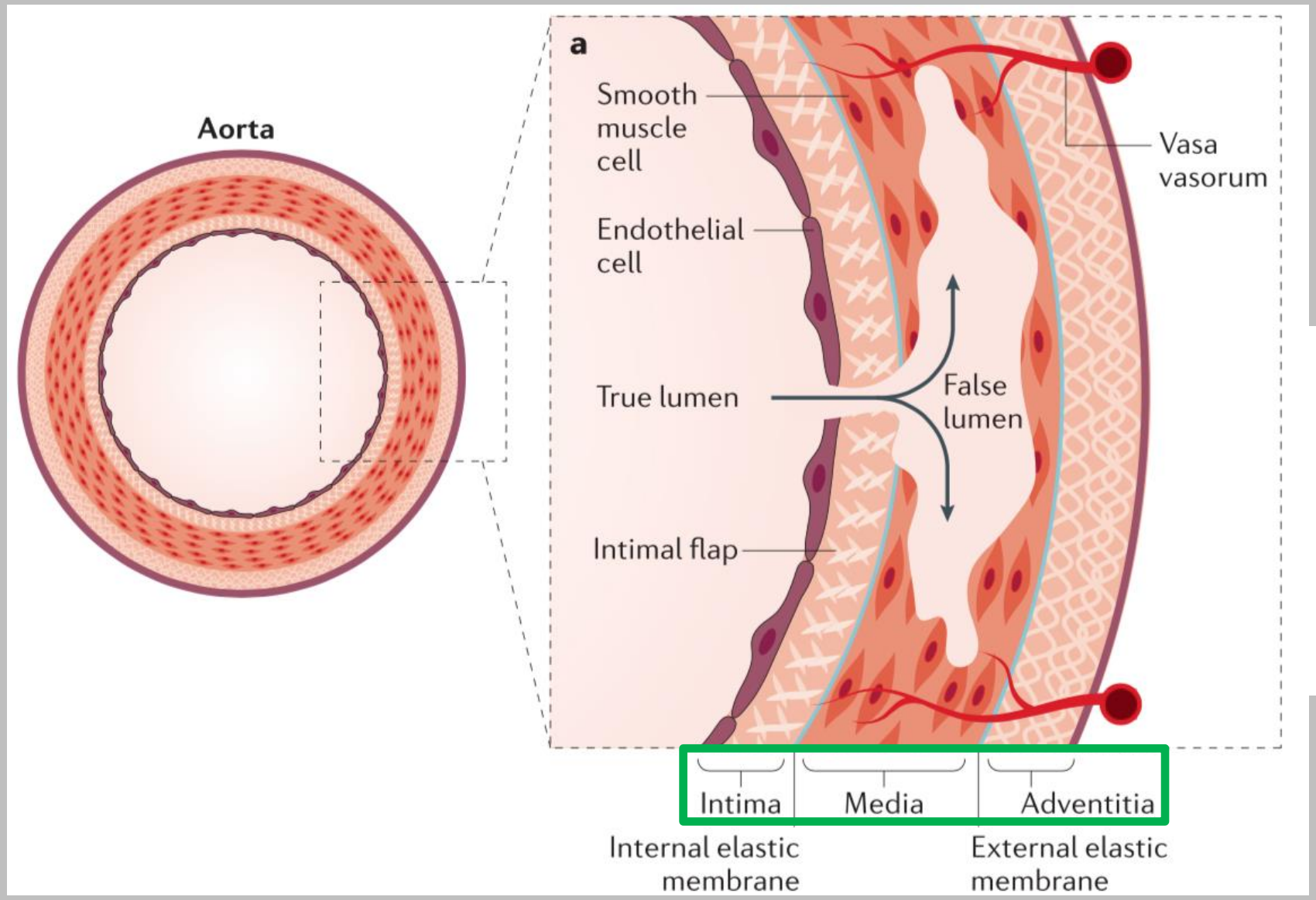
- Aortic dissection (AD)
- Intramural hematoma (IMH)
- Penetrating atherosclerotic ulcer (PAU) and
- Traumatic aortic injury (TAI)

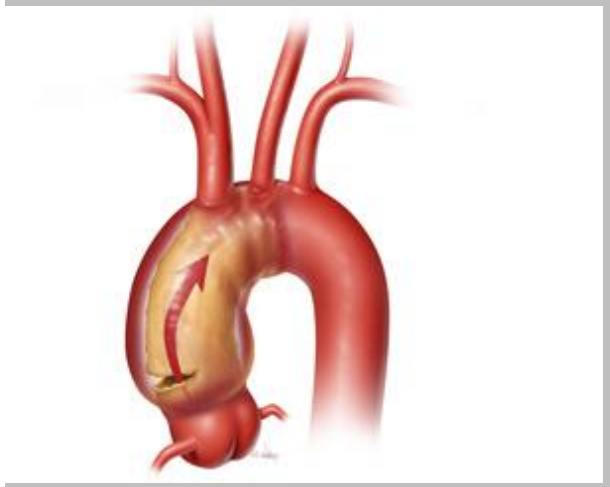

Annual reported cases/USA
2000
5 year survival rate
85.7%
Type A Aortic Dissection
Incidence
2.0 - 4.04 per 100,000/year
Out of hospital mortality
48.6%

Aneurysm vs dissection



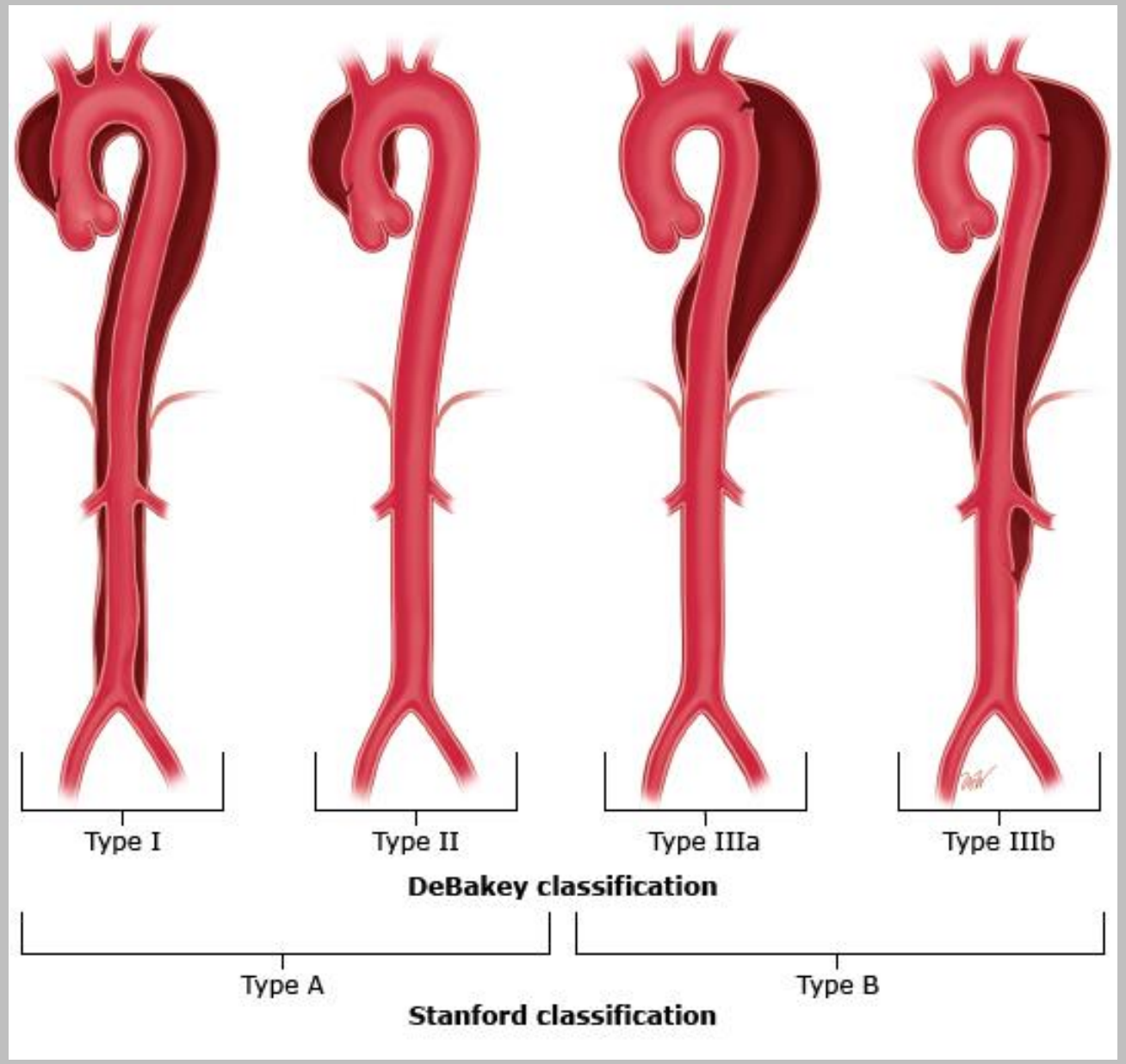
Aortic anatomy and dissection





Background

- Aortic dissection is a tear in the wall of aorta
  - Tear involves inner and middle layers of the aortic wall with propagation of a false lumen within the middle layer
- Aortic dissections are classified by the area of aortic involvement
  - Type A: involving the ascending aorta
  - Type B: all other dissections


Type I
Type II
Type IIIa
Type IIIb
DeBakey classification
Type A
Type B
Stanford classification

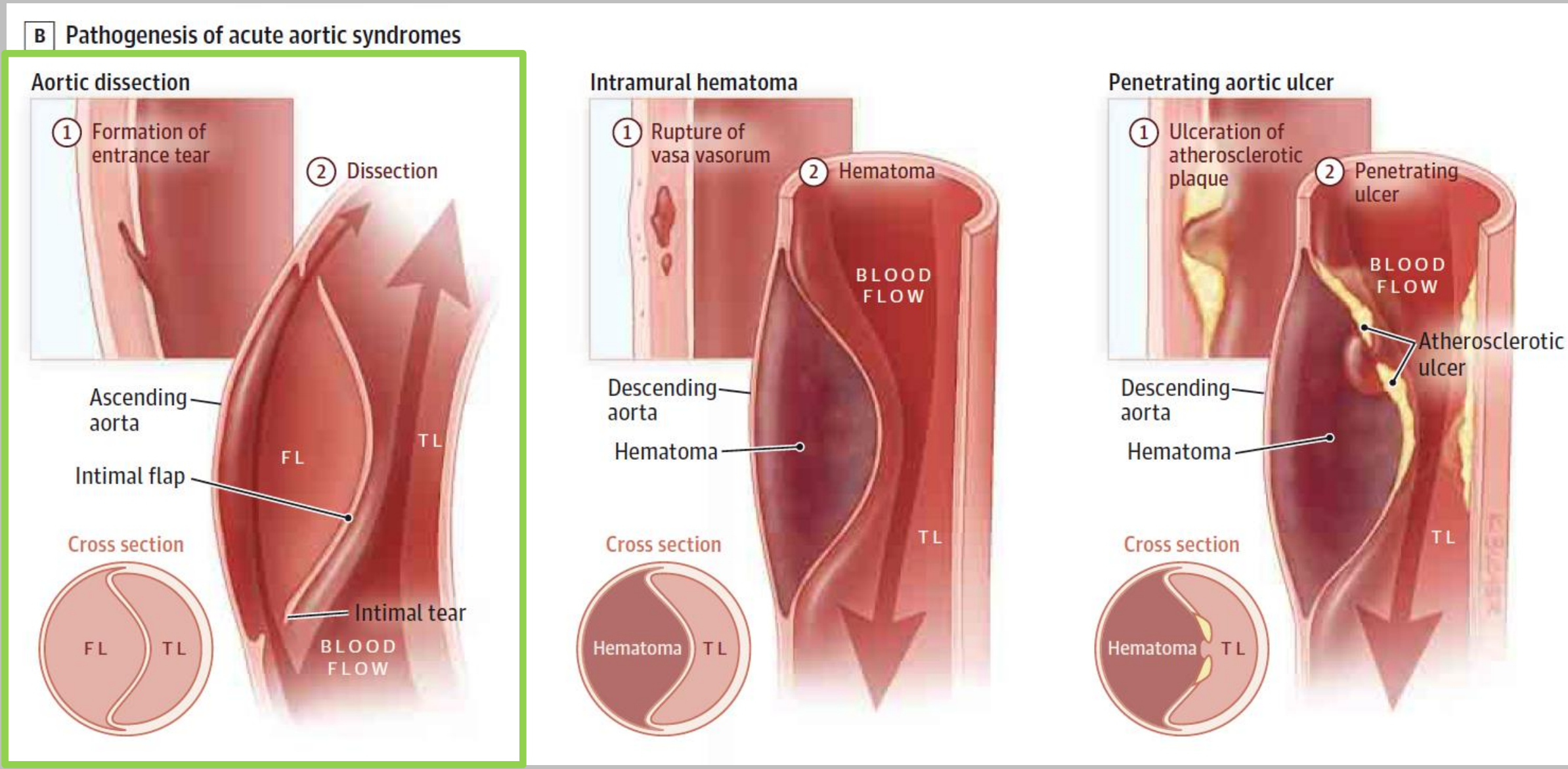
Epidemiology of Aortic Dissection

- Acute aortic dissection is rare
- The true incidence is hard to define because aortic dissections can be instantly fatal in the pre-hospital setting; death is often attributed to other causes
- The incidence of acute aortic dissection is higher in men and older adults

- Immediate death following a short period of symptoms, or no symptoms occurs in an overwhelmingly high percentage of patients with acute aortic dissection.
- Some investigators report the immediate mortality to be as high as 40%.
- Mortality results from aortic rupture, pericardial tamponade leading to cardiogenic shock, acute aortic valve regurgitation, and acute myocardial ischemia in the case of coronary ostia involvement.

- The diagnosis of acute type A aortic dissection requires a high index of suspicion. Up to 30% of patients are initially misdiagnosed
- In-depth history taking and a detailed physical examination are essential in the diagnosis of type A aortic dissection.

Acute aortic syndrome



Risk factors for aortic dissection

Lifestyle and cardiovascular risk factors

- Long-term hypertension
- Old age
- Dyslipidemia
- Pregnancy-induced hypervolemia
- Weight-lifting
- Smoking
- Cocaine abuse

Congenital and connective tissue disorders

- Bicuspid aortic valve
- Marfan syndrome
  - FBN1 & FBN2
- Loeys-Dietz syndrome
- Ehlers-Danlos syndrome
- Turner syndrome

Trauma

- Aortic transection
- Motor vehicle deceleration injury
- Falling from height

Iatrogenic

- Cardiac catheterization
- Arterial cannulation for cardiopulmonary bypass
- Aortic cross-clamping during valvular or aortic surgery
- Intra-aortic balloon pumps

Vascular inflammation

- Autoimmune disease
  - Giant cell arteritis
  - Takayasu arteritis
  - Bechet disease

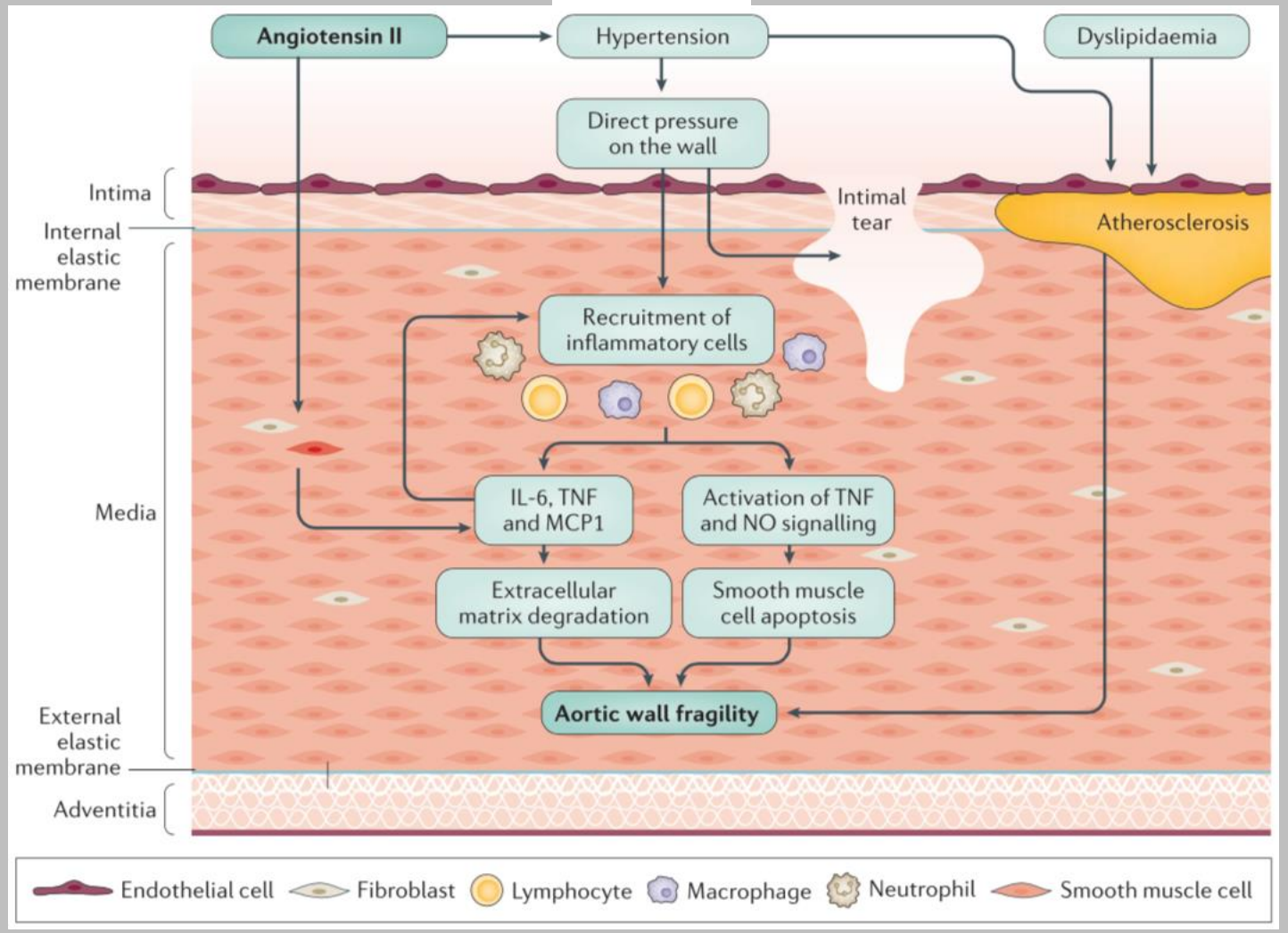
Aortic aneurysm

Infectious disease

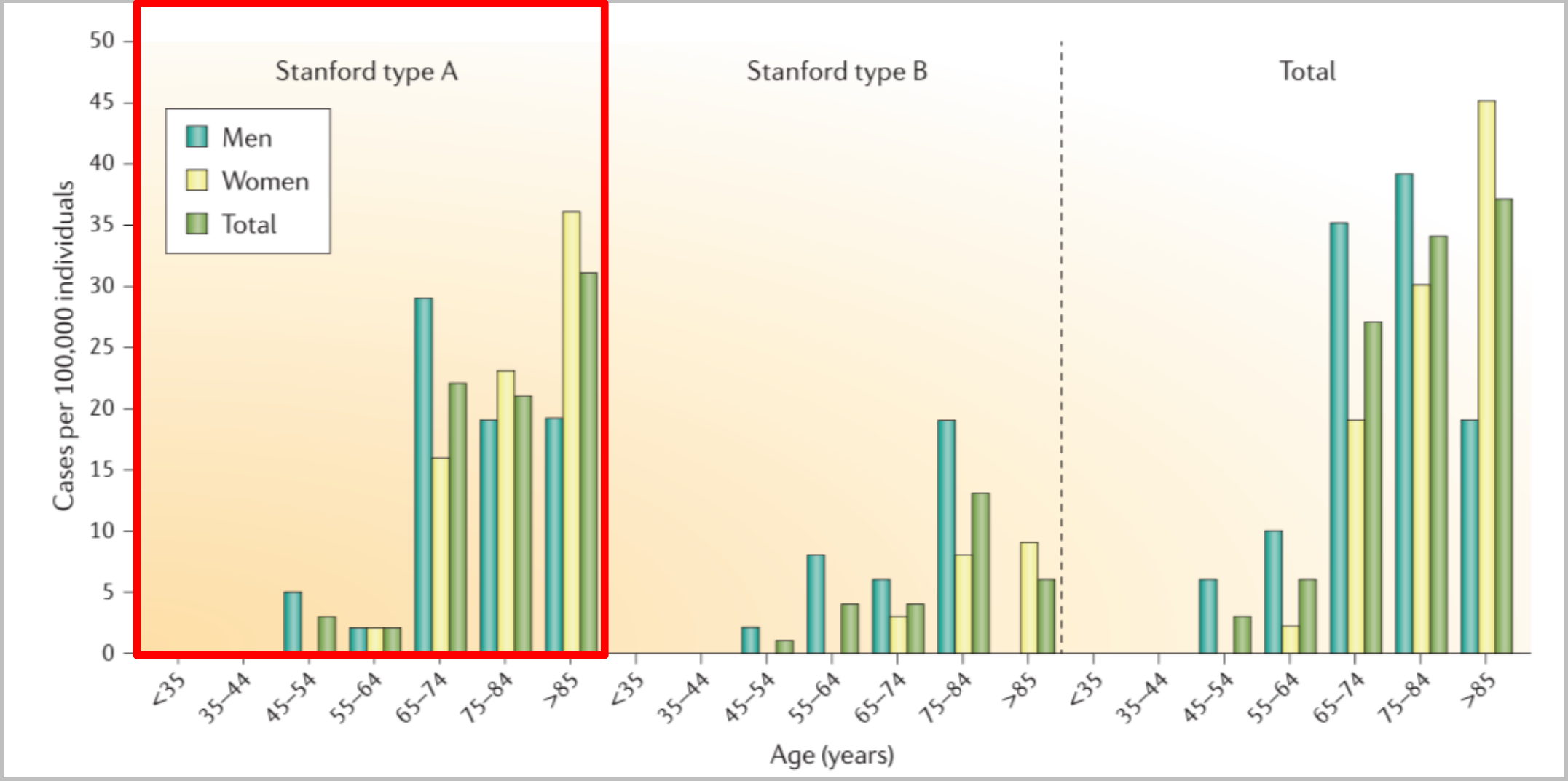
- Syphilis
- Tuberculosis

Aortic intimal pathophysiology

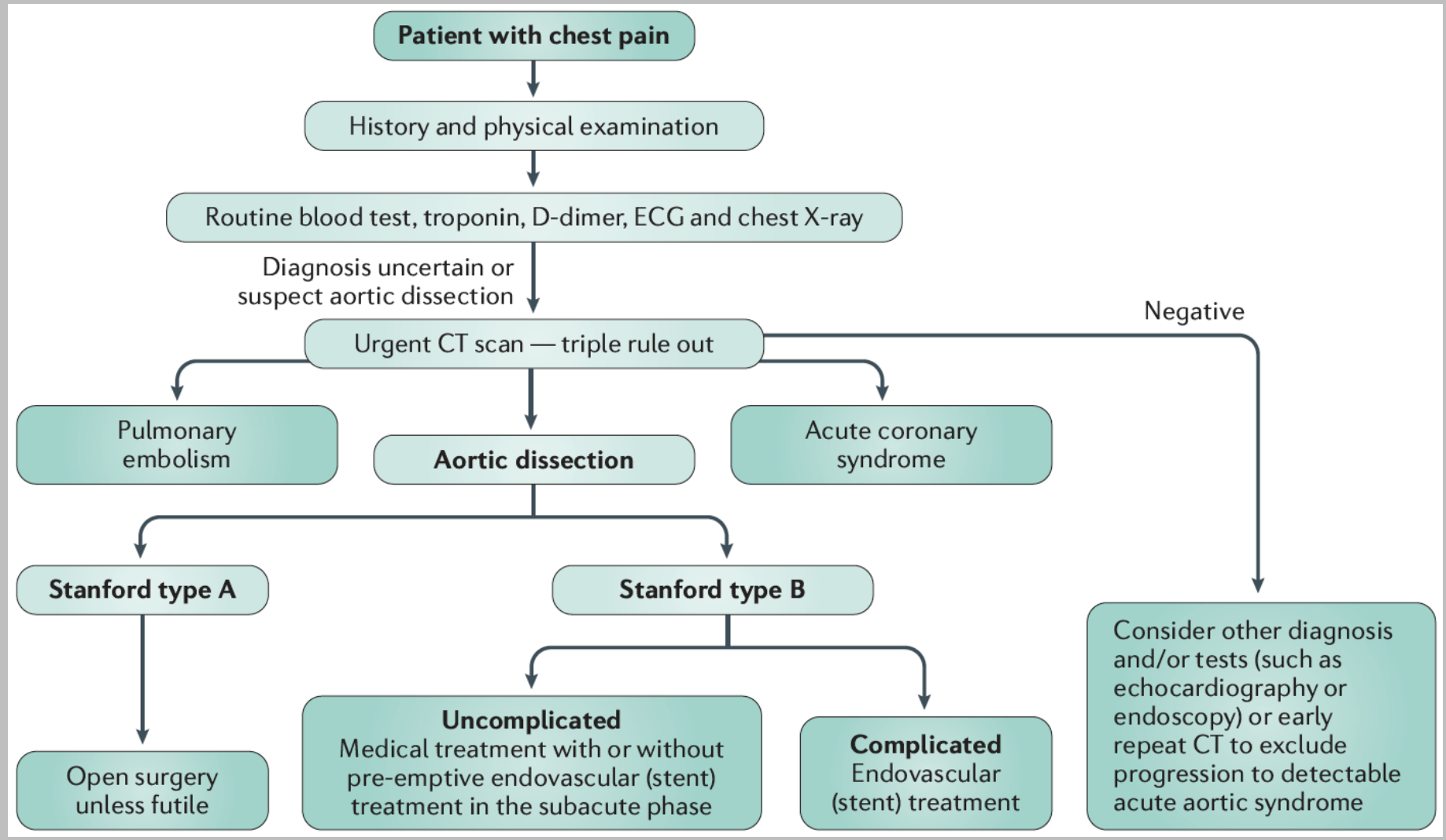
$$F = \frac{\Delta P}{R}$$



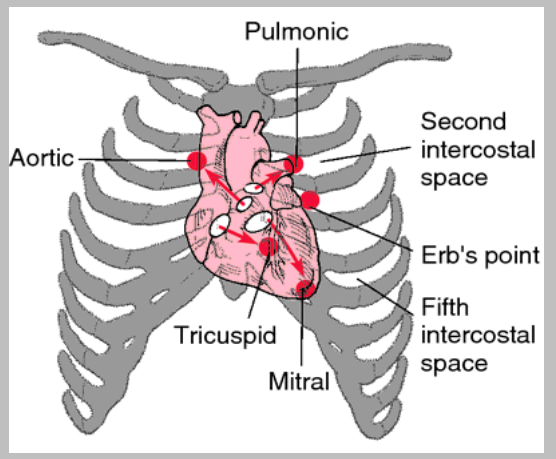
Aortic dissection epidemiology



Diagnostic algorithm



Physical exam / preop evaluation



- Anterior chest or interscapular pain of extreme intensity
  - Accompanied sweating, vomiting, fainting
- Diastolic murmur (if aortic valve insufficiency)
- Pulsus paradoxus (if tamponade)
- Cardiogenic shock (if aortic insufficiency, tamponade, major coronary occlusion)
- Hypovolemic shock (aortic rupture)
- Abdominal bruit
- Loss of peripheral pulses / bilateral BP differential of >20mmHg
- Hemiplegia, hemiparesis, or paraplegia
- Evaluation of preoperative state of all organ systems

Clinical Presentation

- If undiagnosed and untreated, acute aortic dissection can have a very high mortality
  - For untreated type A dissection, mortality is estimated to be 1%−2% per hour for the first 48 hours
  - Type B dissections can also have a high mortality, up to 70% at 30 days for high-risk groups

Clinical Presentation

- Common presentations :
  - Chest pain (90%)
  - Sudden, severe, sharp, stabbing, or tearing chest pain (40%−50%)
  - Pain radiating to the back (47%−64%)
  - Chest pain with a widened mediastinum on chest radiograph (60%)
  - Pulse deficits or differences in blood pressure between the arms (19%−34% for type A)
  - Chest pain with new aortic regurgitation (32%−76%)

- As many as 33% of acute type A aortic dissection patients present with symptoms of end organ malperfusion, which substantially impacts outcomes.

- Cerebral, peripheral, and visceral malperfusion can occur separately or in combination and have been shown to be independent predictors of postoperative outcomes

Clinical Presentation

Less common presentations for dissection include:

- Pain that radiates to the abdomen and lower extremities (17%)
- Ischemic complications such as renal infarction (14%), mesenteric ischemia (5%), spinal cord ischemia (3%)
- Inferior myocardial infarction (1%–7%)

Physical Examination

- The classic physical examination findings for acute aortic dissection (e.g., diastolic murmur, blood pressure differential between arms, focal neurologic deficit) are seen in fewer than half of all patients with acute aortic dissection

Diagnostics - Laboratory

- Current Tests:
  - Inflammatory markers:
    - C-reactive protein
    - IL-6
  - Cardiac stress or damage:
    - Troponin
    - Creatinine kinase
    - Pro-BNP
  - Thrombosis / fibrinolysis
    - D-dimer

Diagnostic Testing for Dissection

- Conventional chest radiographs show widening of the aorta in 63% of type A dissections, while 11% show no abnormality
- The comparable values in type B dissections were 56% and 16%

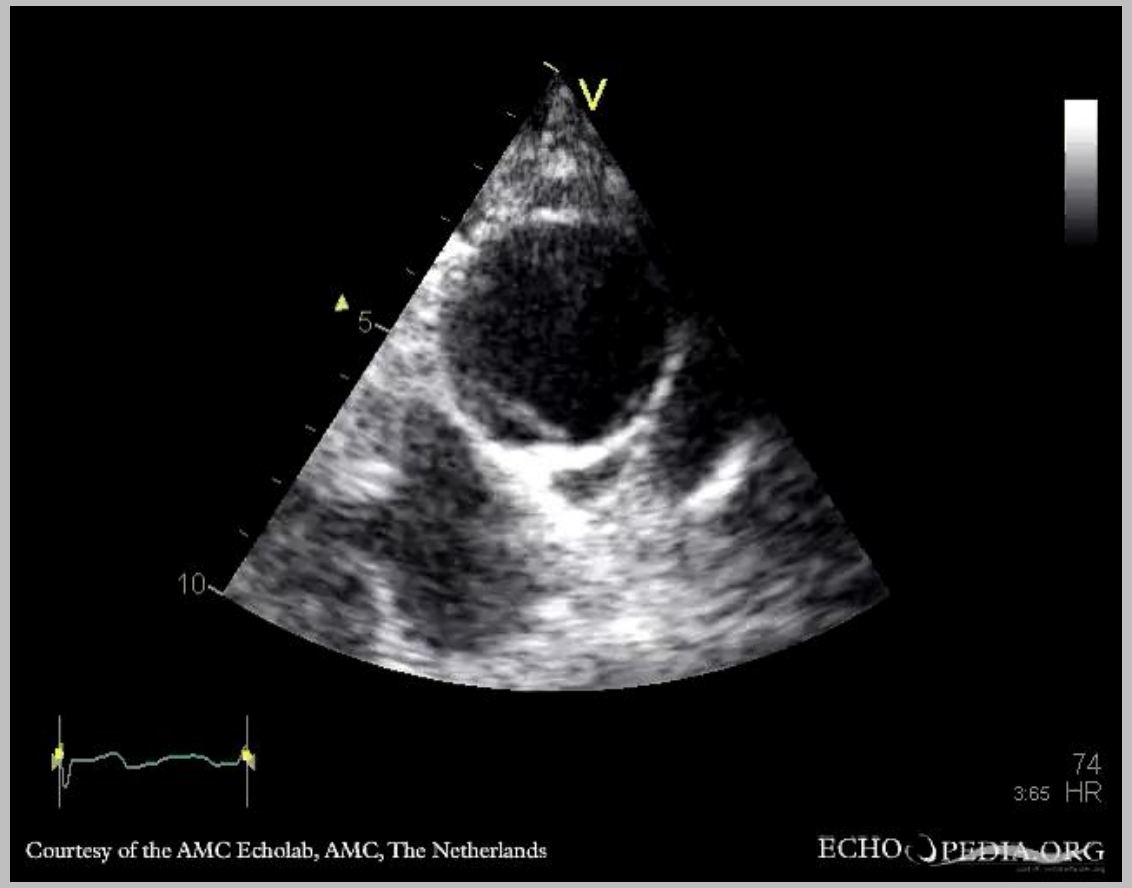
Diagnostic Testing for Dissection

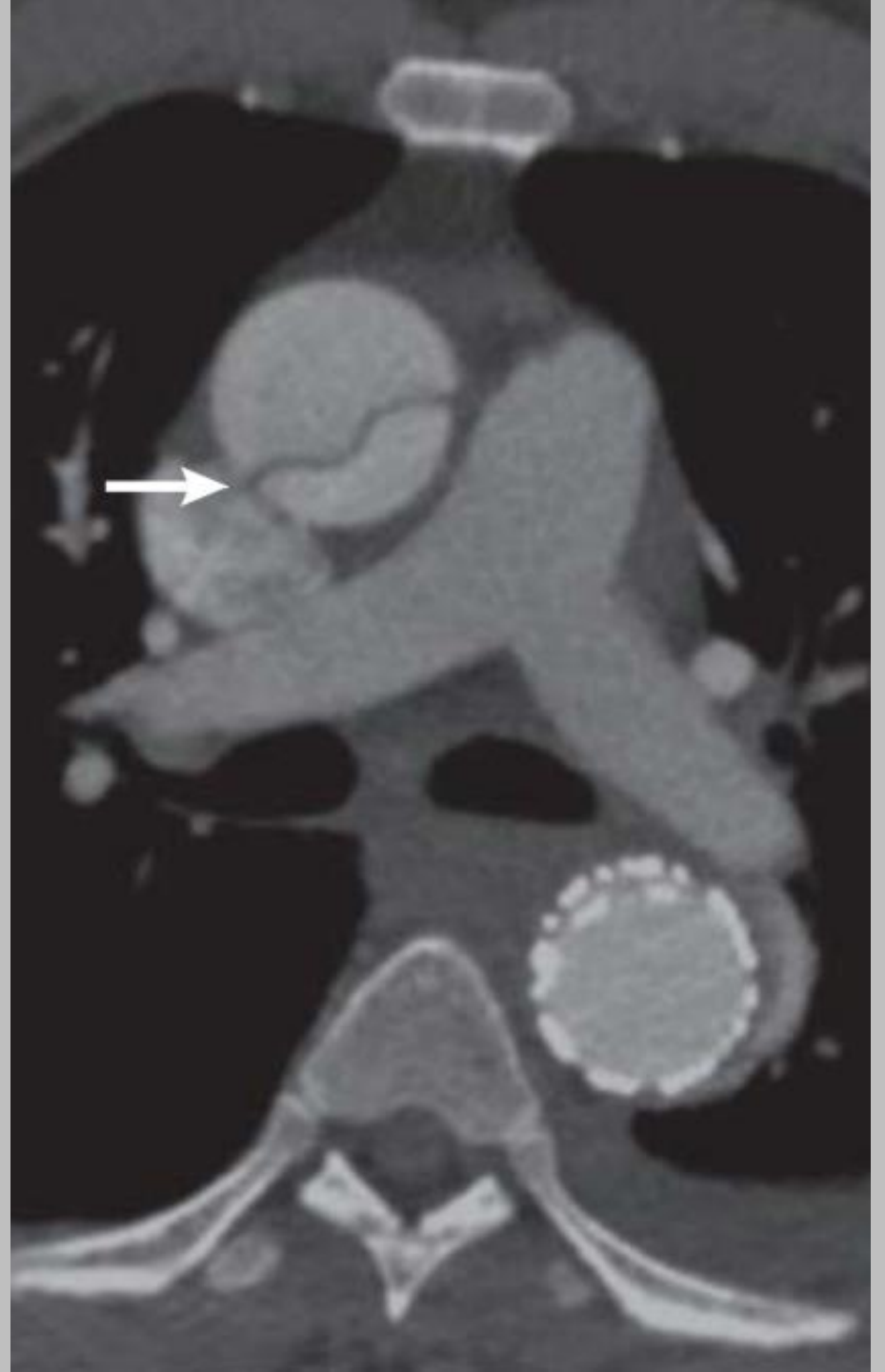
- Due to the limited sensitivity of chest radiographs, especially in type B dissections, a normal chest radiograph should not be used to rule out acute aortic dissection
- In this patient, a right hilar fullness was noted. Without access to the actual chest radiograph, it is difficult to comment on how or whether this finding should have influenced the clinicians in this case

- Computed Tomography
- The sensitivity and specificity of this imaging modality approach 100% (82%-100% and 90%-100%, respectively)
- Transesophageal Echocardiography
- TEE is especially useful in patients wherein contrast is absolutely contraindicated. The sensitivity of TEE in acute type A aortic dissection approaches 100% with a lesser specificity of 70% to 100% due to reverberation artifacts.48 TEE is less useful for the definitive diagnosis of acute type A aortic dissection

- The use of MRI in the diagnosis of acute type A aortic dissection is limited because it is not as readily available as CTA and is time consuming, although it can definitely be diagnostic and provide all the information needed to plan operative repair

Diagnostics - Imaging





Estimation of Pretest Risk of Thoracic Aortic Dissection

High Risk Conditions

1

- Marfan Syndrome
- Connective tissue disease*
- Family history of aortic disease
- Known aortic valve disease
- Recent aortic manipulation (surgical or catheter-based)
- Known thoracic aortic aneurysm
- Genetic conditions that predispose to AoD†

* Loeys-Dietz syndrome, vascular Ehlers-Danlos syndrome, Turner syndrome, or other connective tissue disease.

†Patients with mutations in genes known to predispose to thoracic aortic aneurysms and dissection, such as *FBN1*, *TGFBR1*, *TGFBR2*, *ACTA2*, and *MYH11*.

Estimation of Pretest Risk of Thoracic Aortic Dissection

High Risk Pain Features

2

Chest, back, or abdominal pain features described as pain that:

- is abrupt or instantaneous in onset.
- is severe in intensity.
- has a ripping, tearing, stabbing, or sharp quality.

Estimation of Pretest Risk of Thoracic Aortic Dissection

High Risk Examination Features 3

- Pulse deficit
- Systolic BP limb differential > 20mm Hg
- Focal neurologic deficit
- Murmur of aortic regurgitation (new or not known to be old and in conjunction with pain)

Acute AoD Management Pathway

STEP 1: Immediate post-diagnosis management and disposition considerations

- Arrange for definitive management:
  - Appropriate surgical consultation
  - Inter-facility transfer if indicated based on institutional capabilities

- If transfer required, initiate aggressive medical management until transfer occurs.

Acute AoD Management Pathway

STEP 2: Initial management of aortic wall stress

- Obtain accurate blood pressure prior to beginning treatment.
- Measure in both arms.
- Base treatment goals on highest blood pressure reading.

Acute AoD Management Pathway

STEP 2: Initial management of aortic wall stress

Intravenous rate and pressure control

Rate/Pressure Control

1

Intravenous beta blockade
(If contraindicated,
diltiazem or verapamil)

Titrate to heart rate <60

+

Pain Control

2

Intravenous opiates

Titrate to pain control

Hypotension
or shock state?

No

Yes

Anatomic based management

Systolic BP >120mm HG?

Secondary pressure control

BP Control

3

Intravenous vasodilator

Titrate to BP <120mm HG (Goal is lowest possible BP that maintains adequate end organ perfusion)

Acute AoD Management Pathway
STEP 2: Initial management of aortic wall stress

Anatomic based management

Type A dissection

1. Urgent surgical consultation
   +
   Arrange for expedited operative management
2. Intravenous fluid bolus
   •Titrate to MAP of 70mm HG or Euvolemia
   (If still hypotensive begin intravenous vasopressor agents)
3. Review imaging study for:
   • Pericardial tamponade
   • Contained rupture
   • Severe aortic insufficiency

Type B dissection

1. Intravenous fluid bolus
   •Titrate to MAP of 70mm HG or Euvolemia
   (If still hypotensive begin intravenous vasopressor agents)
2. Evaluate etiology of hypotension
   • Review imaging study for evidence of contained rupture
   • Consider TTE to evaluate cardiac function
3. Urgent surgical consultation

Acute AoD Management Pathway

STEP 3: Definitive management

- Depending on the results from the pressure control or anatomic based management, continued treatment will involve either:
  - ongoing medical management, or
  - operative or interventional management.

Acute AoD Management Pathway

STEP 3: Definitive management

Based on results from intravenous rate and pressure control:

Based on results from anatomic based management:

Dissection involving the ascending aorta?

No

Etiology of hypotension Amenable to operative management?

Ongoing medical management

Close hemodynamic monitoring
Maintain systolic BP < 120mm Hg
(Lowest BP that maintains
end organ perfusion)

Complications requiring operative or interventional management?

Limb or mesenteric ischemia
Progression of dissection
Aneurysm expansion
Uncontrolled hypertension

Yes

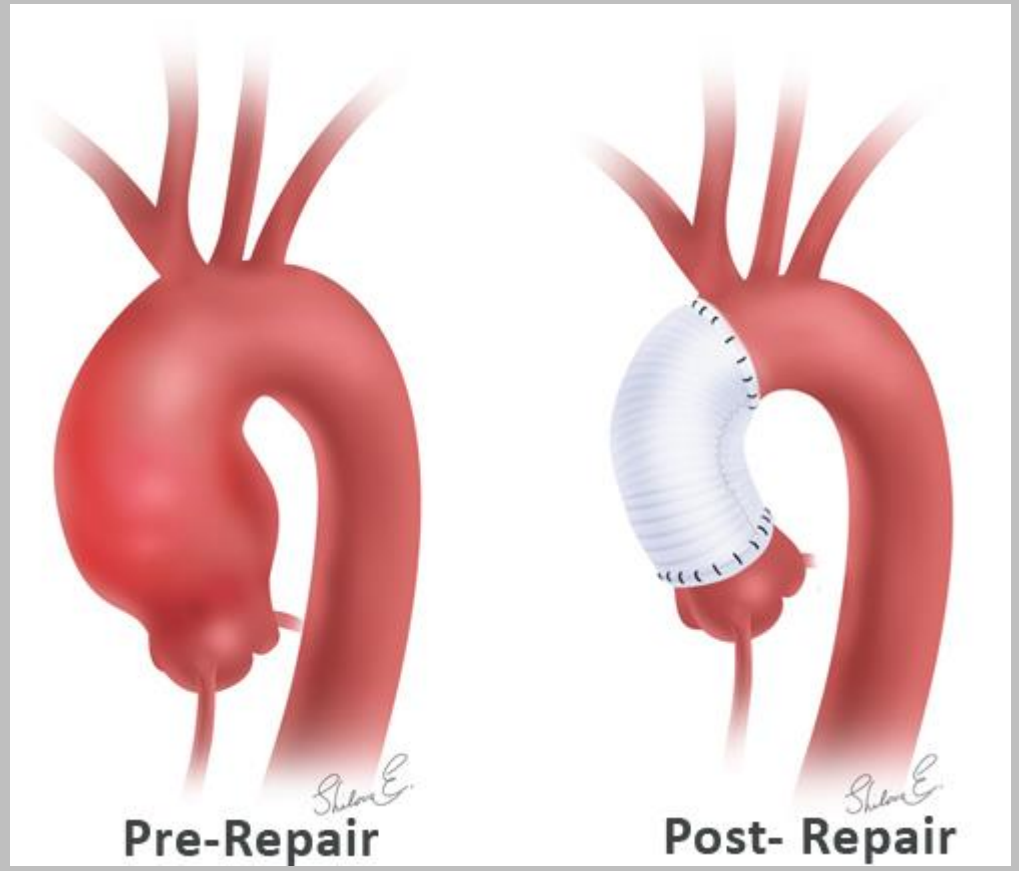
Operative or interventional management

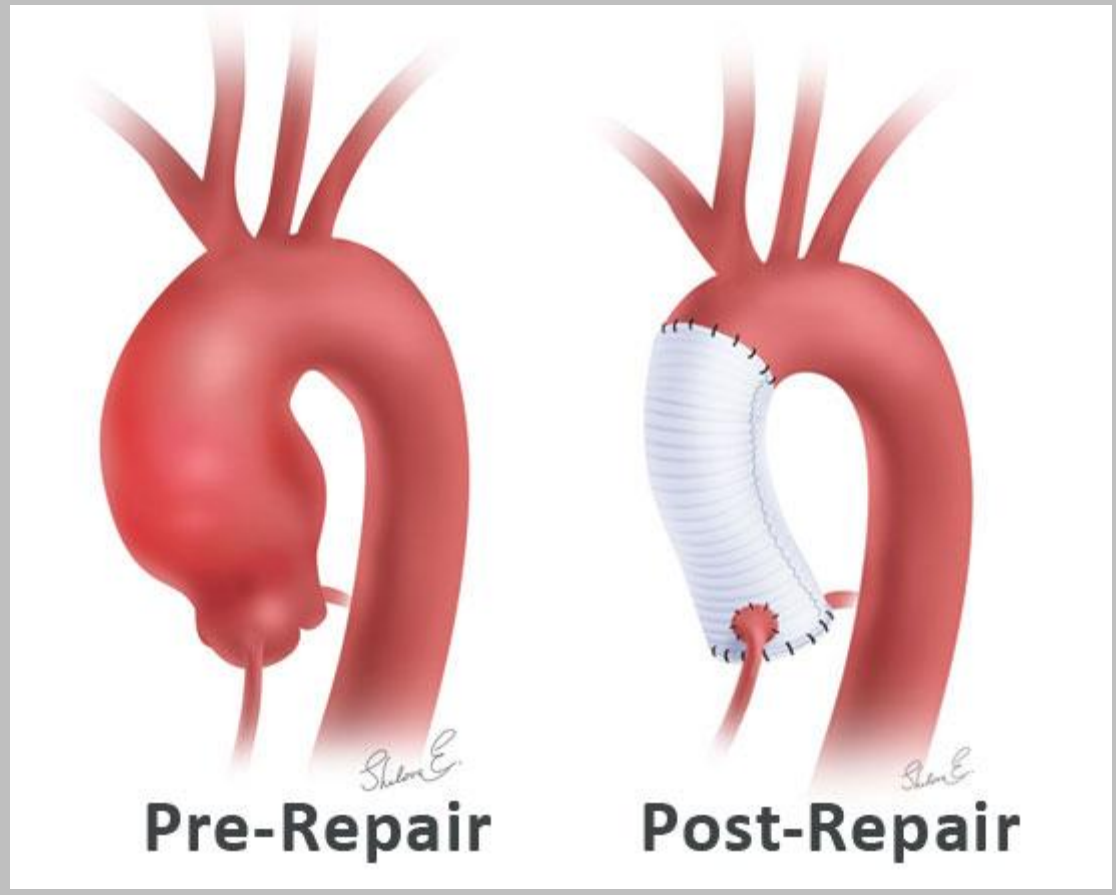
Yes

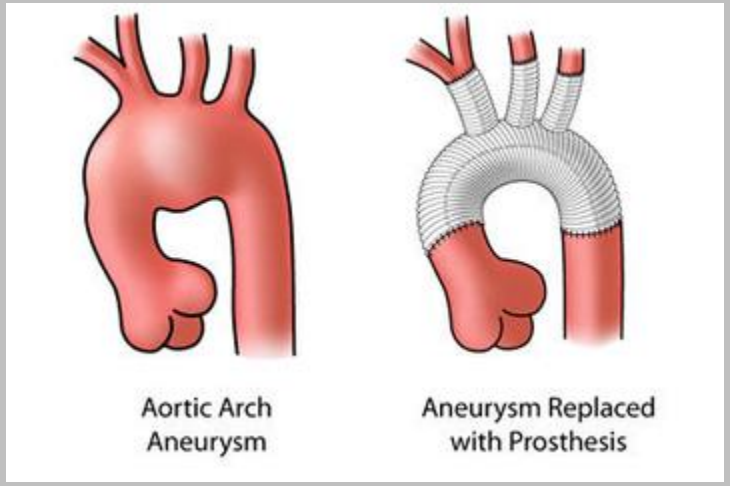
Operative or interventional management

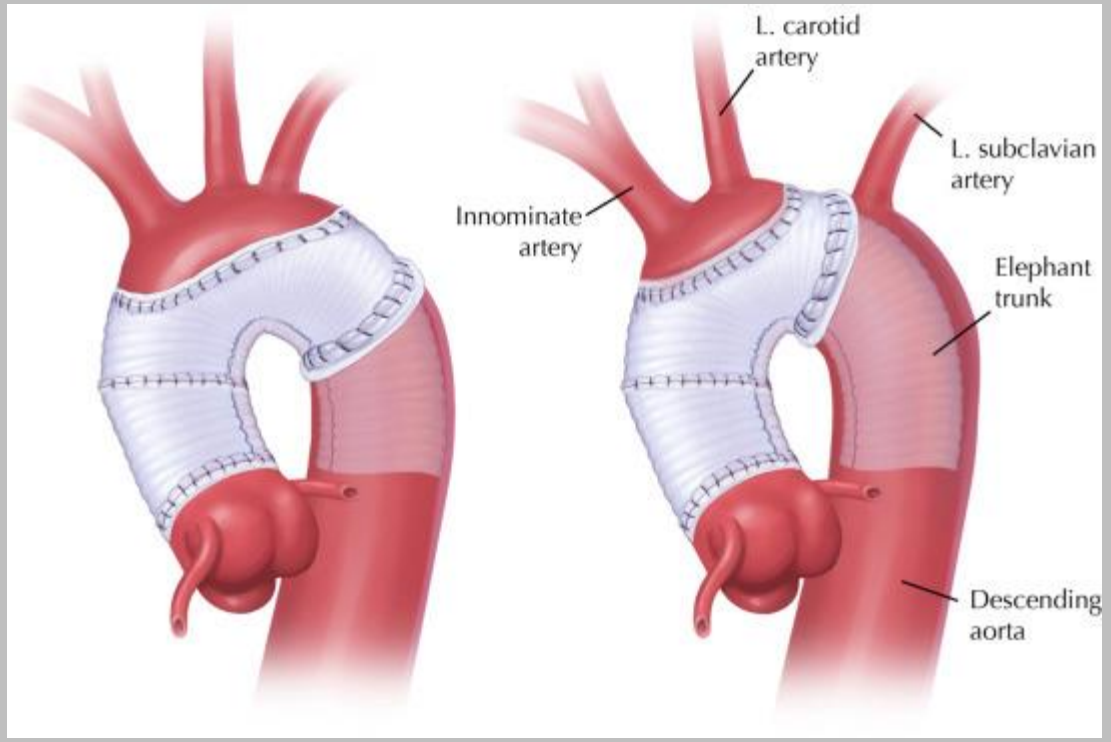
Treatment - Dissection

- Type A
  - Surgical
- Type B
  - Medical
  - Surgical
    - Acute with rupture, leak or distal ischemia.


Pre-Repair
Post- Repair


Pre-Repair
Post-Repair


Aortic Arch
Aneurysm
Aneurysm Replaced
with Prosthesis


L. carotid
artery
L. subclavian
artery
Innominate
artery
Elephant
trunk
Descending
aorta

Thank You for your Attention