EMERGENCY DIAGNOSIS AND TREATMENT OF AORTIC DISSECTION

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HISTORY

1761: Morgagni. Described dissecting aneurysm.

1804: Scarpa. Dissection related to "corrosion and rupture of the proper coats of the aorta." Hematoma resulting from blood dissecting through the arterial wall.

1826: Laennec. Introduced term "dissecting aneurysm."

1855: First antemortem diagnosis of dissection

1843-1863:

Thomas Peacock. Cadaver experiments.

1. Ruptured intima ("lacerable")
2. Blood penetrating the media
3. Distal reentry ("imperfect natural cure")

HISTORY cont'd.

1864: von Recklinghausen. Attributed dissection to molecular changes in elastic tissue

1910: Babes, Mironescu. Aortic dissection could occur secondary to hemorrhage in vasa vasorum "dissecting mesarteritis."

1910: Moriani. Microscopic changes in elastic tissue and connective tissue of media

1929: Erdheim. Cystic medial necrosis as underlying cause.

1934: Shennan. "Dissecting aneurysms"

- degeneration of media frequent
- neither atheroma nor lues were important

HISTORY cont'd.

1943: Oppenheimer and Taussig. First described ascending aortic *aneurysm* in the Marfan syndrome

Etter and Glover. First described ascending aortic *dissection* in the Marfan syndrome

1955: McKusick. Documented that aortic dissection is a common cause of death in the Marfan syndrome

1958: Hirst. Review of 505 cases.

Atherosclerosis not a related factor.

1973: Gore and Hirst. Rupture of vasa vasorum initiating event.

PATHOGENESIS

1. Intimal Tear as Primary Event
   Propagation of dissecting hematoma within media.

2. Intramural hemorrhage due to rupture of vasa vasorum in a defective media
   - Intimal tear is a secondary event.
   - Propagation of cleavage plane by pulsatile force of blood.

RISK FACTORS

ROLE OF <u>ABNORMAL AORTIC MEDIA</u>

1. Cystic medial necrosis (Erdheim)
   - Not always present in dissecting aorta
   - Also present in patients without dissection

2. Aortic dissection associated with connective tissue diseases
   - The Marfan syndrome, Ehlers-Danlos
   - Aortic dilation (thoracic or abdominal, often familial)

3. Aortic dissection associated with
   - Bicuspid aortic valve
   - Aortic coarctation
   - Turner syndrome

RISK FACTORS cont'd.

- **Marfan syndrome**
  (Present in 5-10% of ascending aortic dissections, .02% of adults)
- **Aortic dilatation**
- **Bicuspid aortic valve, coarctation**
- **Turner, Ehlers-Danlos (type IV), Noonan syndrome**
- **Family history of…**
  Marfan syndrome, aortic dissection, aortic disease
- **Hypertension**
  (Present in 70-90% of dissections, but 20-40% of adults)

MARFAN SYNDROME

FEATURES

- Very long arms, fingers, toes
- Pigeon breast or severe pectus carinatum
- Moderate or severe scoliosis
- Dislocated lenses
- Family history of the Marfan syndrome

1995 GHENT NOSOLOGY

INDEX CASE: Major criteria in two systems and involvement of a third system

FAMILY MEMBER: One major criterion in an organ system and involvement of a second organ system

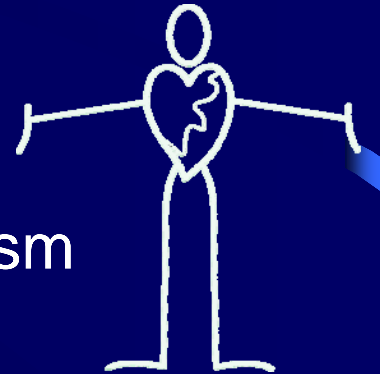
CARDIOVASCULAR SYSTEM

MAJOR CRITERIA

- dilation of ascending aorta
- aortic dissection

MINOR CRITERIA

- mitral valve prolapse
- dilation of a main pulmonary artery
- premature mitral anular calcification (<40yrs.)
- descending thoracic or abdominal aortic aneurysm (< 50 yrs.)



Cardiovascular involvement: one minor criterion

OCULAR SYSTEM

MAJOR CRITERION

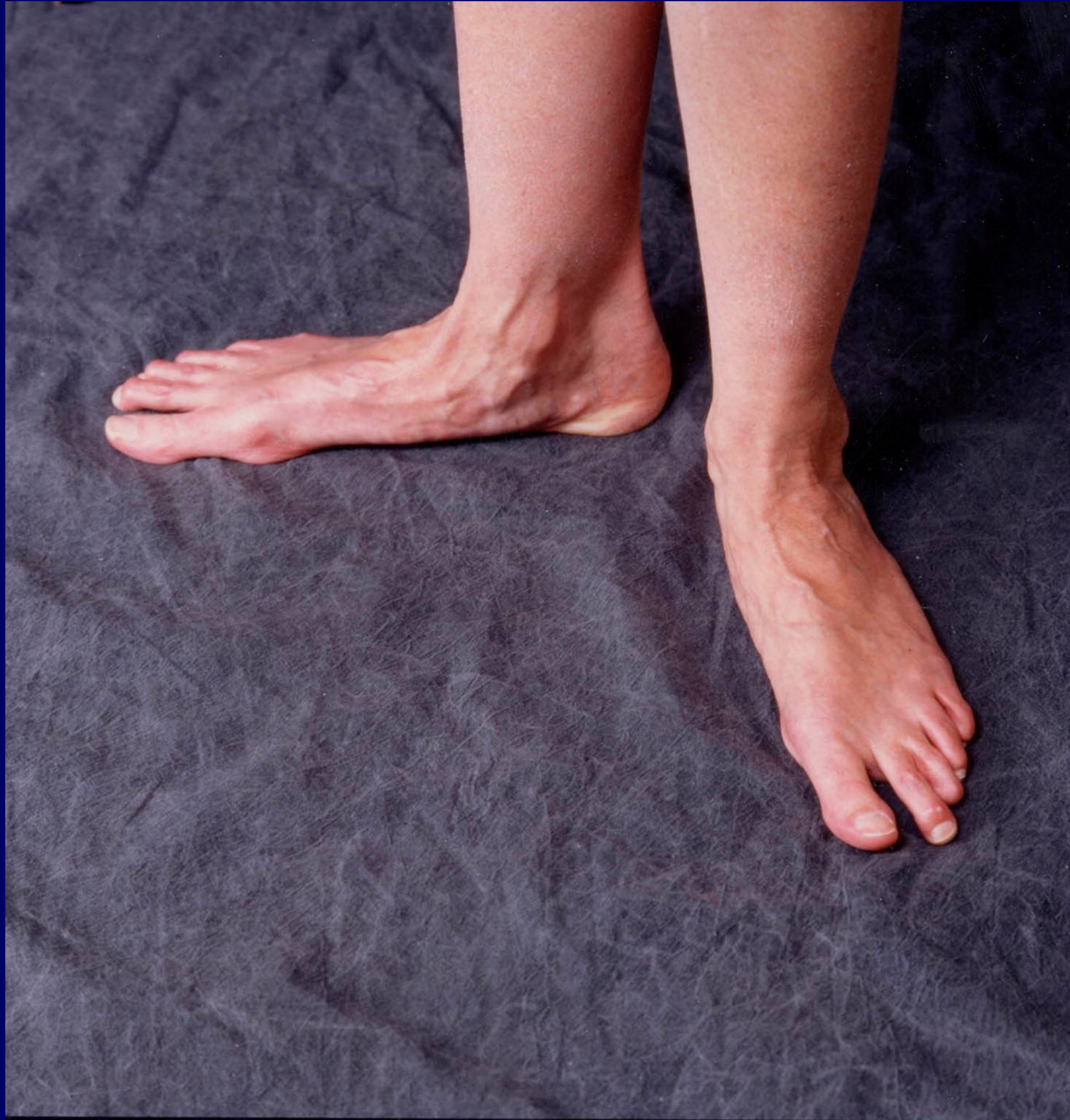
- Ectopia lentis

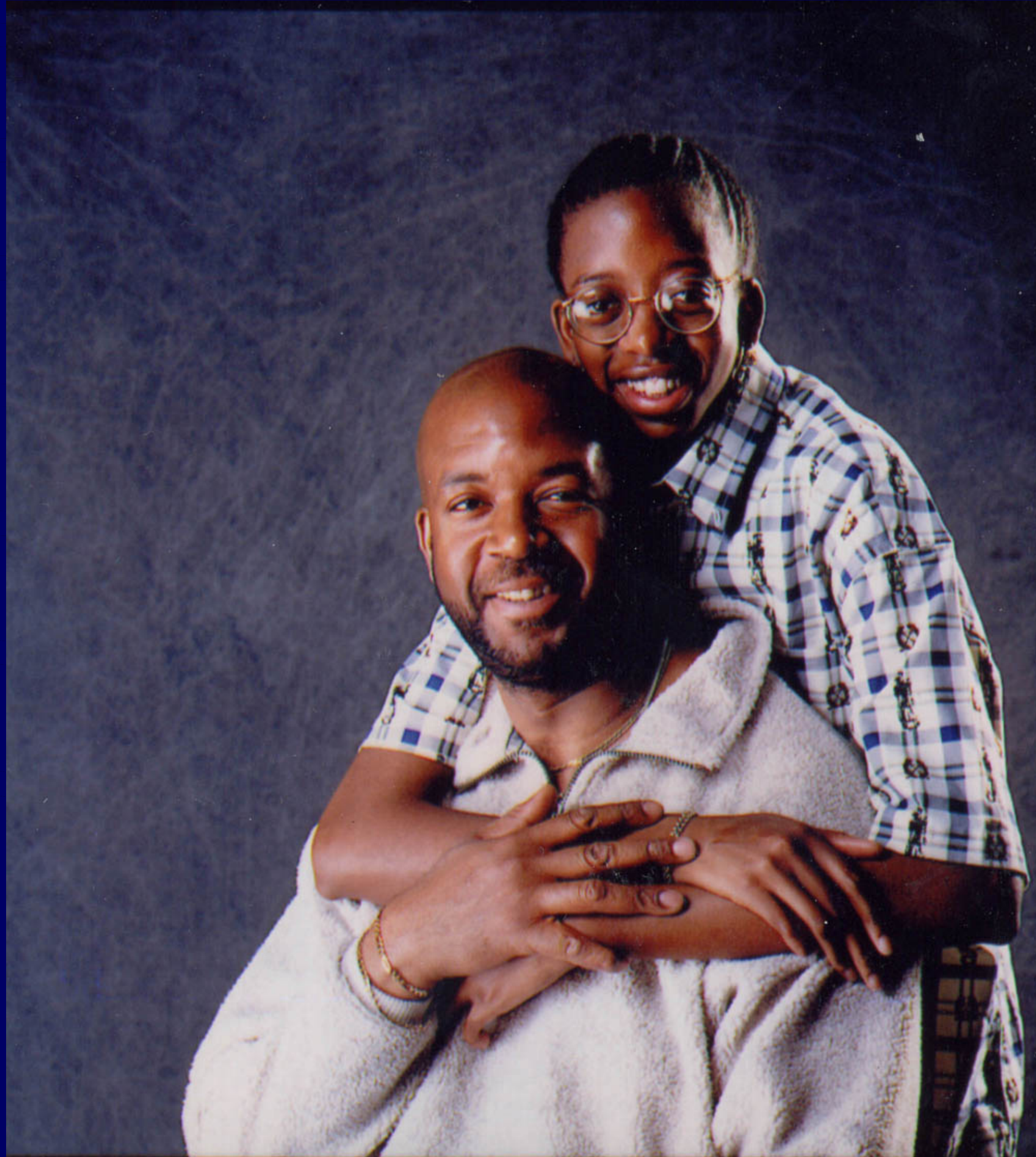
SKELETAL SYSTEM

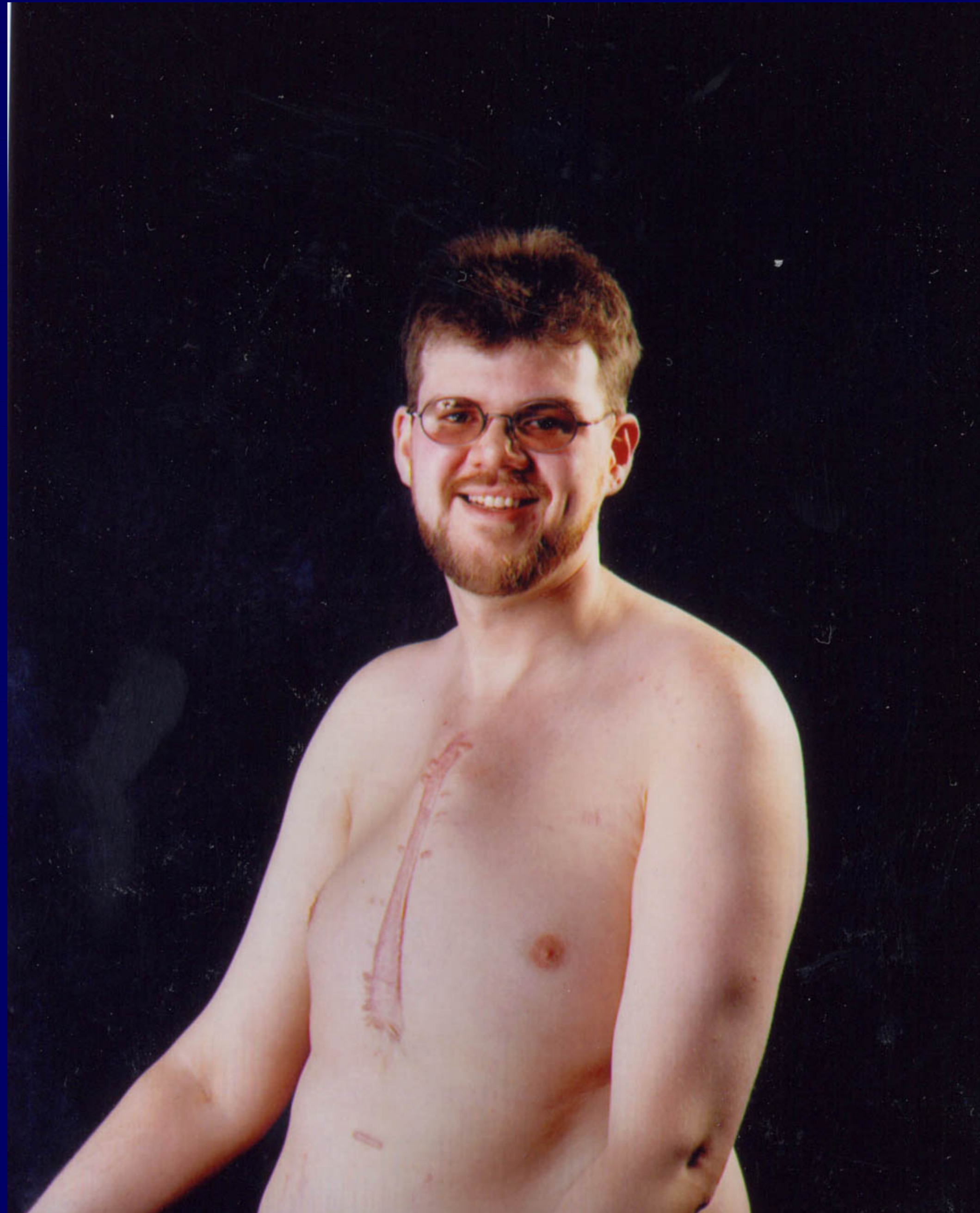
MAJOR CRITERIA

Presence of at least four of the following:

- pectus carinatum
- pectus excavatum requiring surgery
- reduced upper to lower segment ratio or arm span to height ratio > 1.05
- wrist and thumb signs
- scoliosis > 20° or spondylolisthesis
- reduced extension at the elbows (<170°)
- medial displacement of the medial malleolus causing pes planus
- protusio acetabulae of any degree







PROLONGING LIFE

- Correct Diagnosis
- Medical Management
- Surgical Repair

DIAGNOSIS

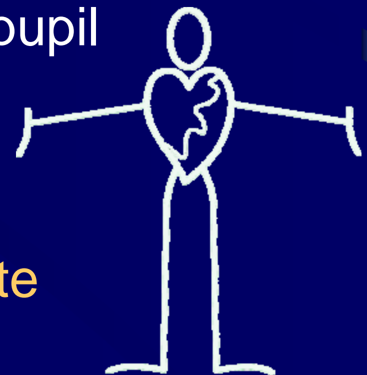
<u>Clinical Evaluation</u>

1. Complete physical examination, including a careful musculoskeletal and skin examination

2. Detailed medical and family history

3. Electrocardiogram (EKG) and echocardiogram, looking for cardiovascualr involvement

4. Eye examination by an ophthalmologist, who dilates the pupil and uses a slit lamp to assess for lens dislocation



This series of steps is important, not only in determining the diagnosis, but also in detecting problems that require immediate attention or long-term management.

MANAGEMENT

<u>UNDERLYING PRINCIPAL:</u>
Weak connective tissue cannot withstand normal tension on aorta.

Since T~P x R

1. Avoid strenuous exercise that increases BP.

2. Use medicine to lower BP and dP/dT.
   - ß-blockers
   - Verapamil
   - Other calcium blockers or ACE inhibitors that lower BP

3. Replace aortic root/valve when aortic diameter $\geq$ 6.0 cm without AR or $\geq$ 5.0 cm with 3+ - 4+ AR.

ETIOLOGIC MECHANISM

- Intimal tear with secondary extension into media
- Hemorrhage into media precipitating secondary intimal tear
- Intramural hematoma
- Penetrating atherosclerotic ulcer

CLASSIFICATION

DEBAKEY

Type I

Ascending aorta extending beyond arch

Type II

Ascending aorta only

Type III

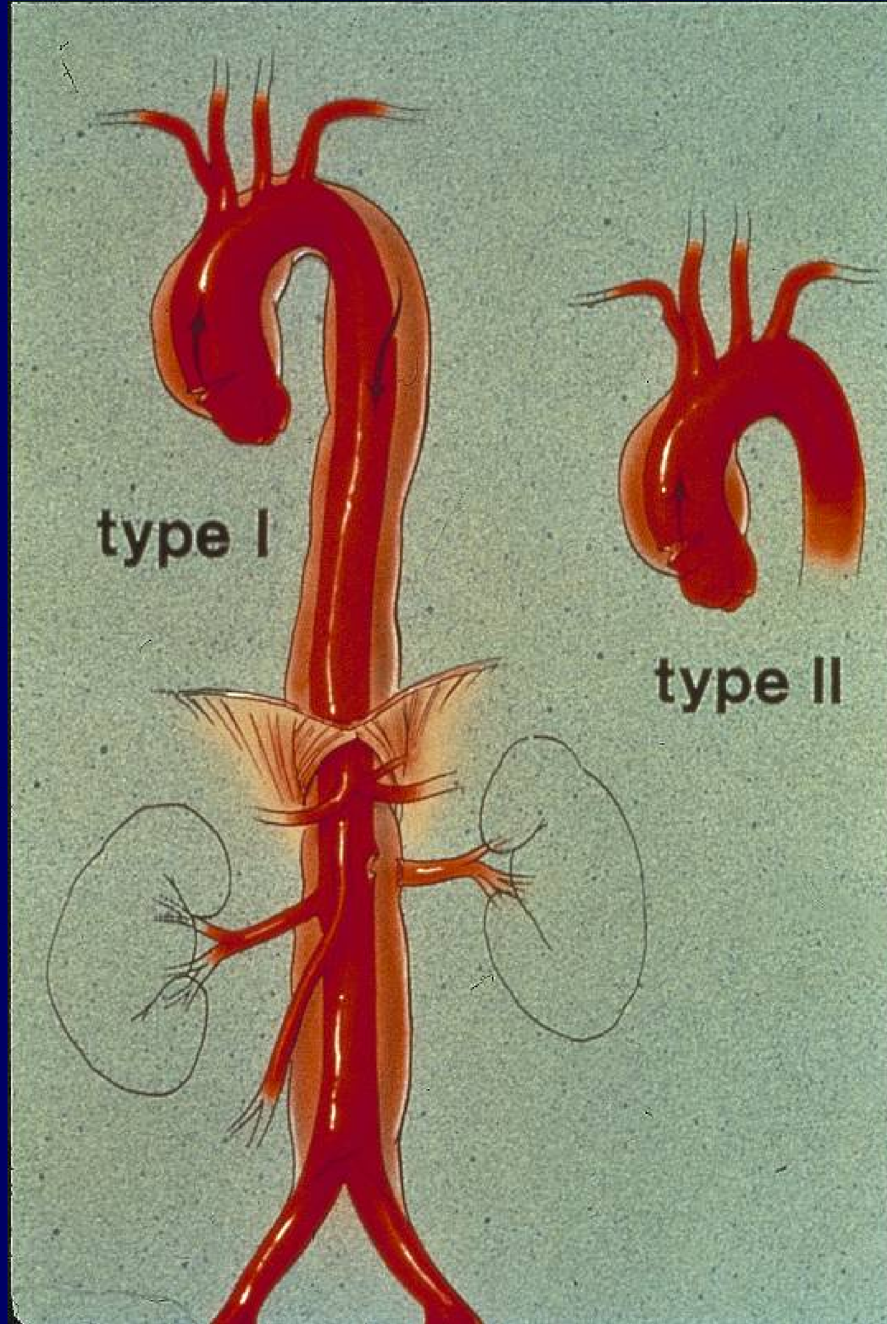
Desending aorta

Type IIIb

Descending aorta extending below diaphragm

STANFORD

- A – Ascending aorta
- B – Descending aorta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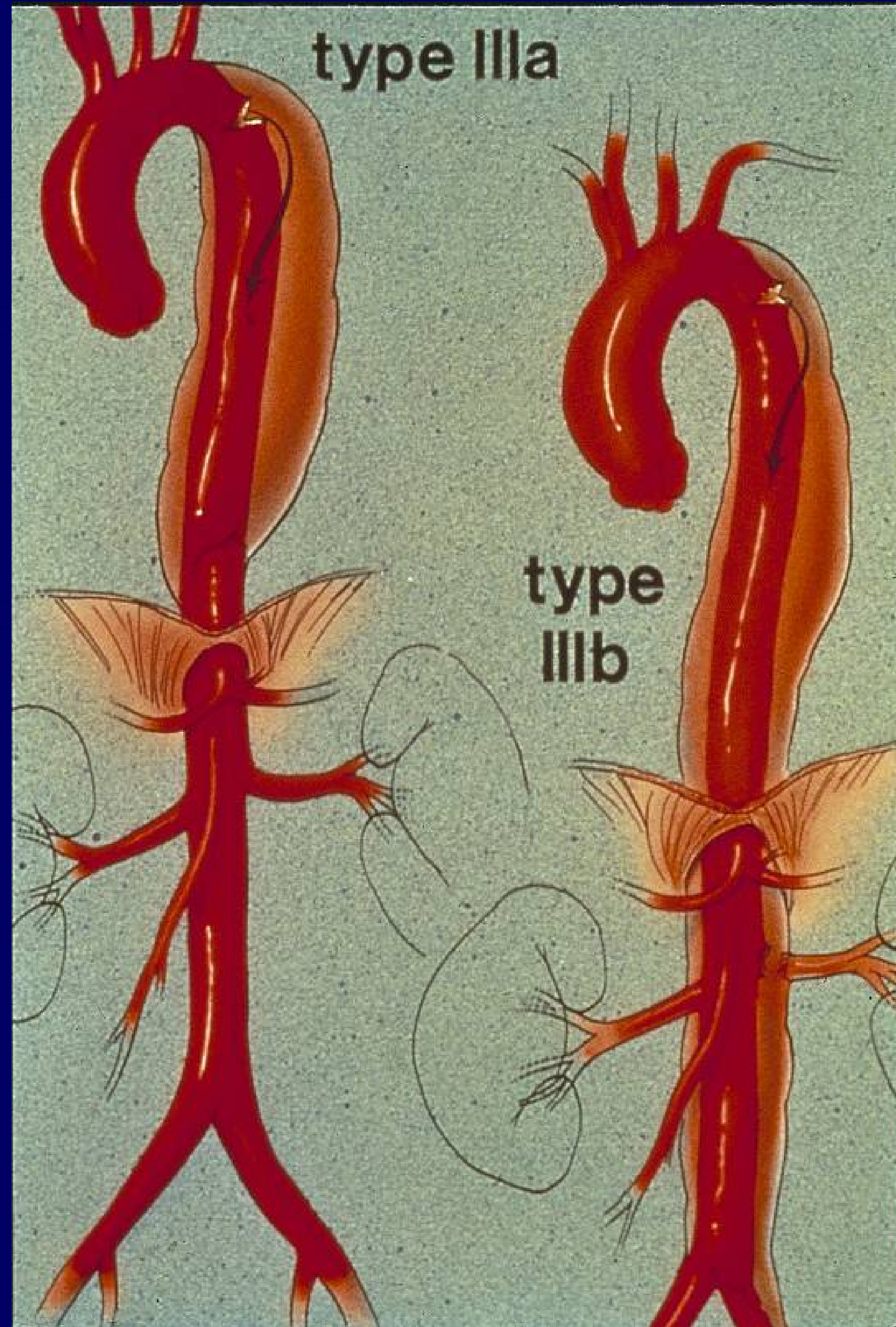
type II



type IIIa
type
IIIb

CLASSIFICATION cont'd.

ASCENDING DISSECTIONS

65-75% of dissections

Entry tear within a few centimeters of aortic valve

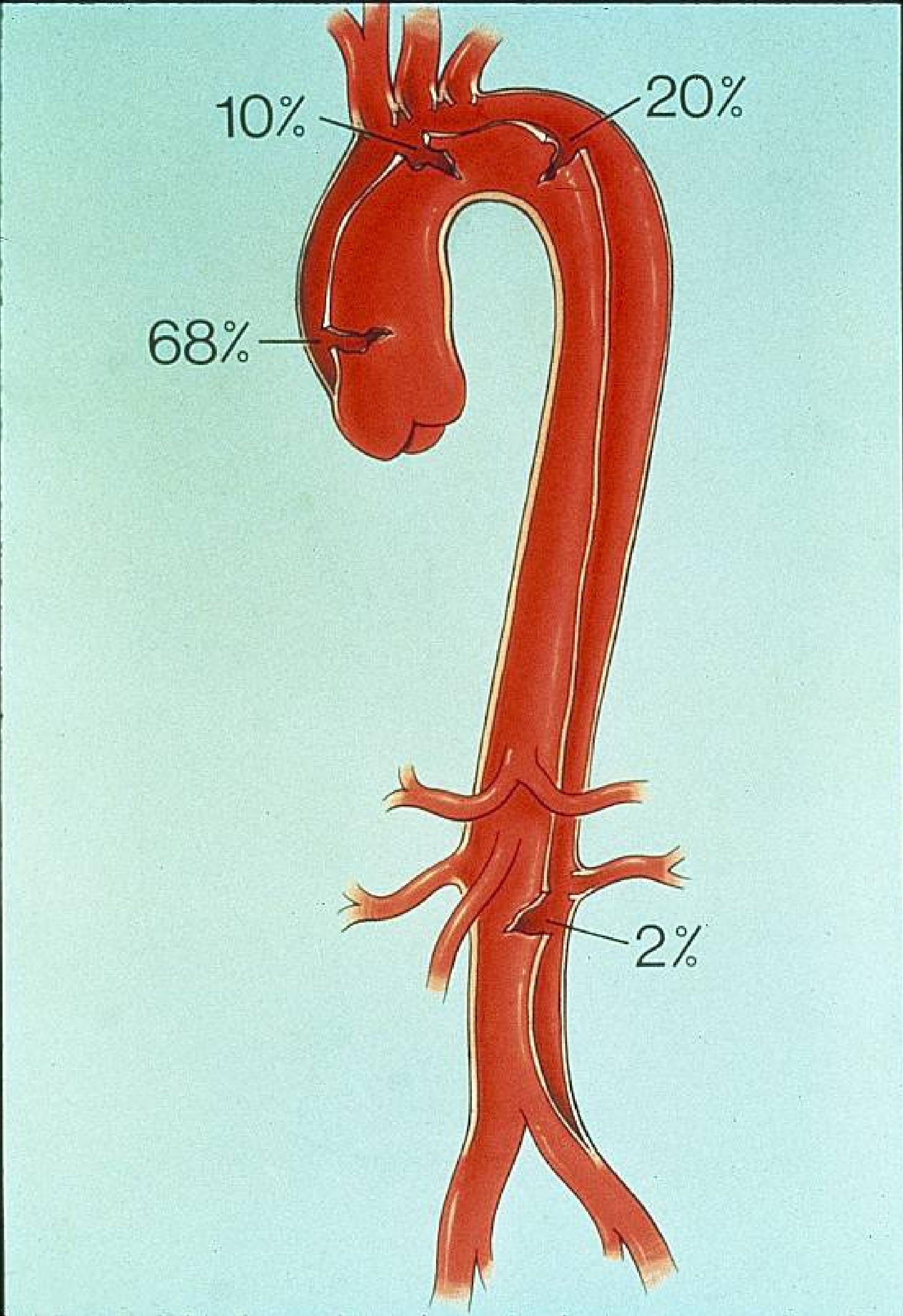
50% extend to iliac bifurcation

DESCENDING DISSECTIONS

25-35% of dissections

False channel begins distal to (L) subclavian

Variable extension



10%
20%
68%
2%

COMPLICATIONS

1. Rupture through outer wall of false channel

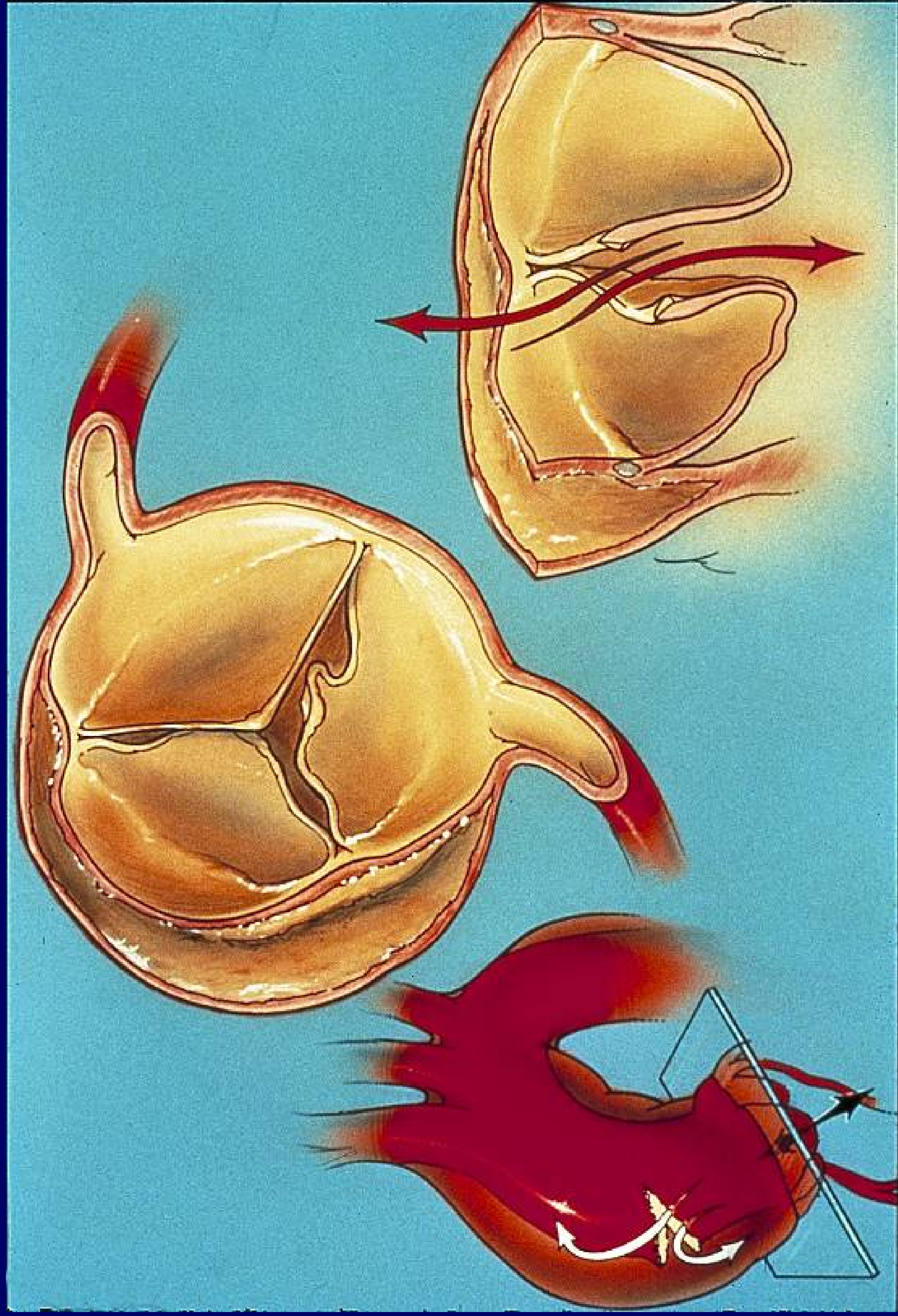
   *typically directly across from entry tear

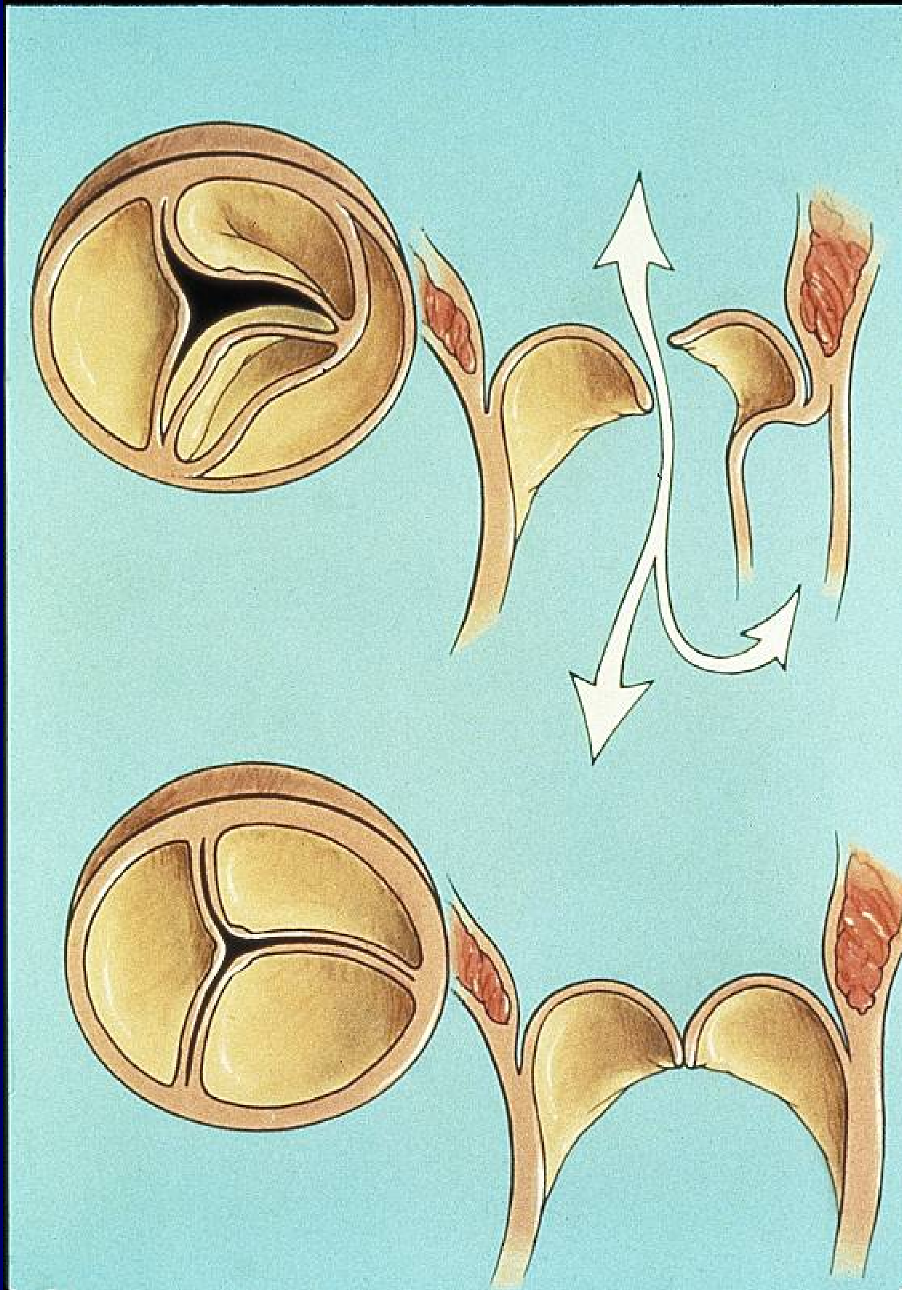
   - pericardial tamponade
   - mediastinal or pleural rupture
   - exsanguination

2. Acute Aortic Regurgitation

   *50% of ascending dissections

   - medial split undermines support of aortic valve
   - may lead to severe CHF



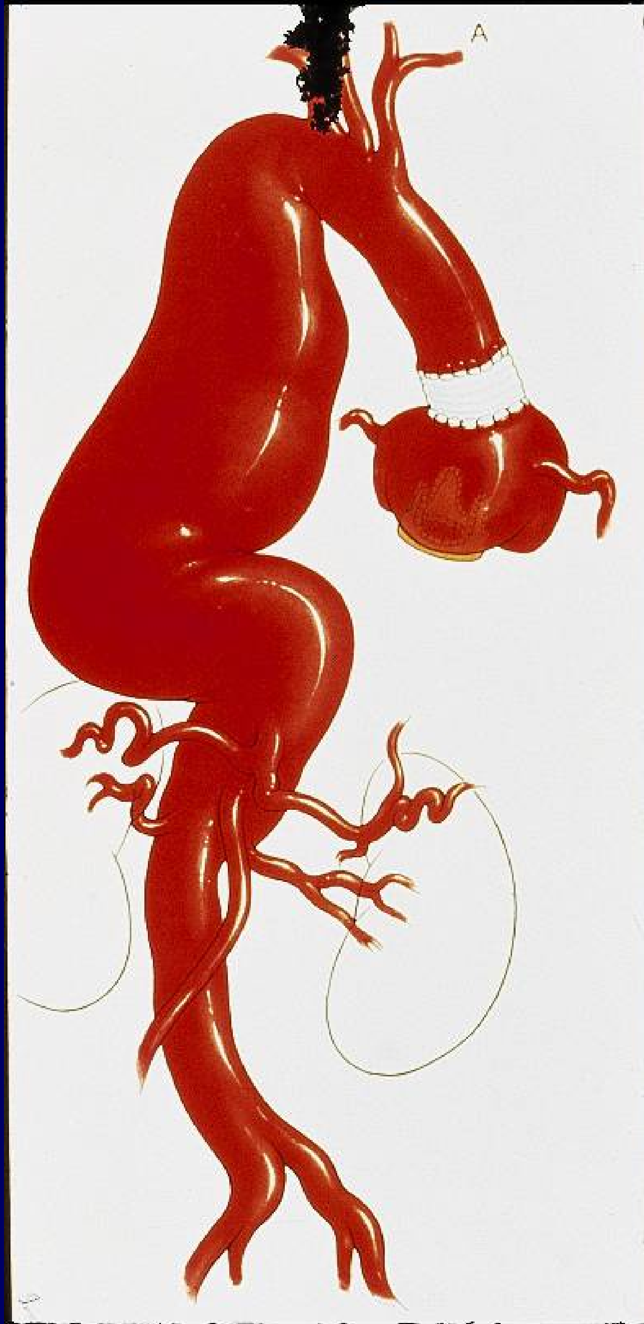


COMPLICATIONS cont'd.

3. Branch Vessel Compromise
   - extension of dissection into a branch vessel
   - compression of orifice by intimal flap

Clinical Scenarios
- stroke
- paraplegia
- HTN-renal failure
- visceral ischemia
- MI

4. Aneurysmal Dilation and Subsequent Rupture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PRIMARY TEAR AND SITE OF RUPTURE

Location of the Tear	Site of Rupture
Ascending	intrapericardial 70% left pleura 6%
Arch	intrapericardial 35% left pleura 32%
Descending	intrapericardial 12% left pleura 44%

CLINICAL FEATURES

1. Incidence uncertain.

   Available information: 5,000 – 10,000 cases/year in U.S.

   Number may be higher (not reportable condition, few autopsies now)
2. Autopsy series – 0.2% autopsies
3. Males 2-5 times more frequent than females
4. Ascending dissections: 50-55 years old

   (<40 years: Marfan, pregnancy, AV disease)
5. Descending dissections: 60-70 years old

CLINICAL FEATURES cont'd.

Most important factor
leading to a correct diagnosis is
a high clinical suspicion!

CLINICAL FEATURES cont'd.

1. Sudden Onset Severe Pain
   - May or may not be catastrophic
   - *Ripping, tearing*
   - Migratory
   - Never experienced before
   - Restless, sense of doom

2. Look for underlying connective tissue disorder

3. Hypertension (especially moderate or severe) or known aortic aneurysm

PAIN CHARACTERISTICS

SEVERE PAIN (90%)

- Most Severe at Onset
  - Anterior Pain: Proximal Dissection
  - Posterior Pain: Distal Dissection
  - Migratory Pain

- Pain in these locations usually due to other more common disorders (MI, pneumonia, pleurisy, pulmonary embolism, pneumothorax, ulcer, cholecystitis, pancreatitis)

- Must consider aortic dissection in cases without other confirmed cause of pain.

CLINICAL FEATURES

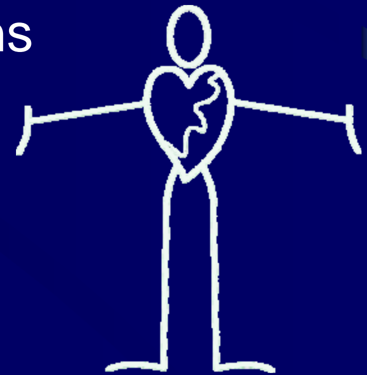
A. Syncope 10% Ominous external rupture (hemopericardium)	B. Stroke Carotid artery
C. Paraparesis, Paraplegia Spinal artery	D. Horner Syndrome

PHYSICAL EXAMINATION

- Acutely ill
- Hypertension (catecholamines, renal ischemia)
- Hypotension (20%): acute complications
- Aortic insufficiency: (50-60% ascending dissections)
- Pulse deficits: (60% ascending dissections)
  - May change over time
  - Be wary the pt with acute (L) leg ischemia and negative embolectomy
- Other
  - Sternoclavicular joint pulsation, high JVP, bruits, abdominal mass

NATURAL HISTORY

- Autopsy Series: >50% of people with untreated aortic dissections are dead within 48 hours.
- 1934 Shennan: >300 cases reviewed.
  40% acute ascending dissections died suddenly.
  None lived > 5 weeks
- Anagnostopoulos et al. Am J Card 1972
  973 pts with untreated proximal and distal dissections
  50% died with 48 hours
  84% died within 1 month
- 1% per hour risk of death in first 48 hours



• Acute	0-14 days
• Chronic	14+ days

DIAGNOSTIC EVALUATION

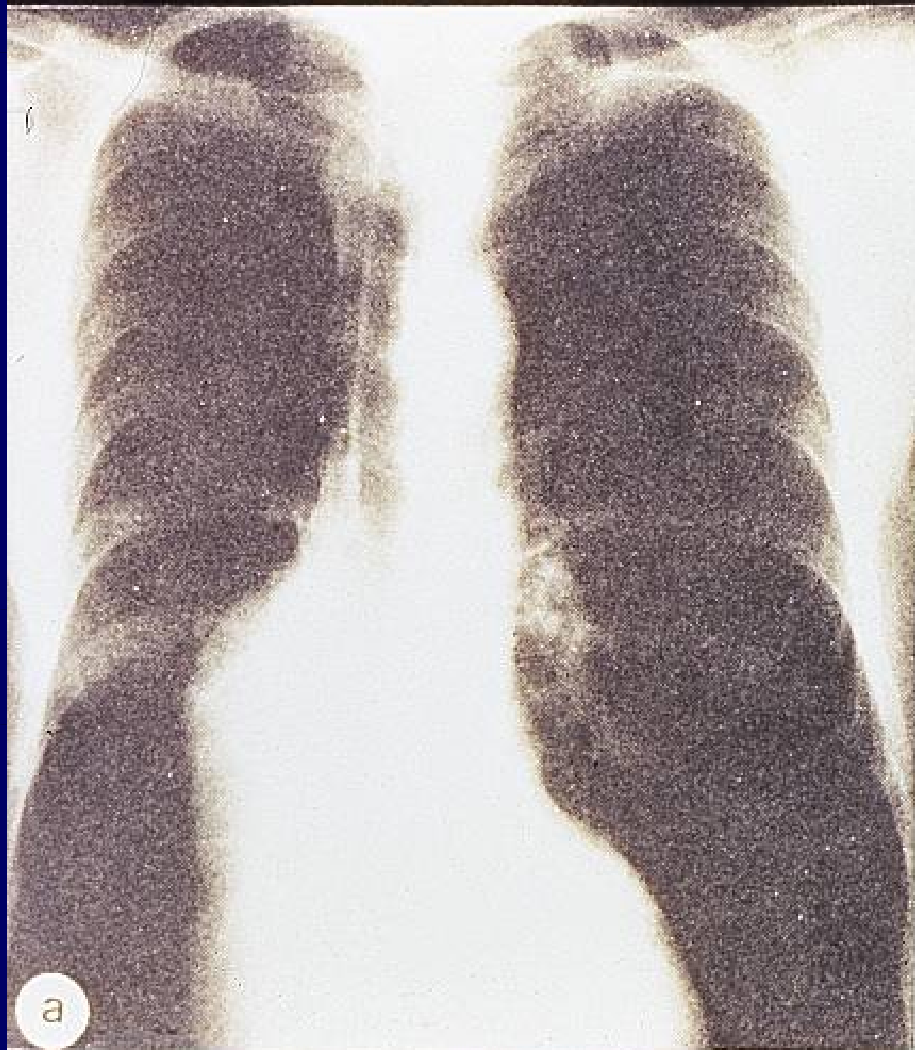
- Chest radiograph
- Transthoracic echocardiogram
- Transesophageal echocardiogram*
- Computed tomography*
- Magnetic resonance imaging*
- Aortography

**Choose based on rapid availability and quality of performance*

CHEST X-RAY

1. Signs of dissection are indirect
   - Abnormal aortic knob, widened mediastinum, pleural effusion
2. Insensitive with variable inter-observer agreement
3. Look for changes compared with old films
4. Displaced intimal calcification (>5 mm) – useful in older patients
5. <u>Normal in 18%</u>

<u>A Normal CXR Should Not Deter Further Evaluation.</u>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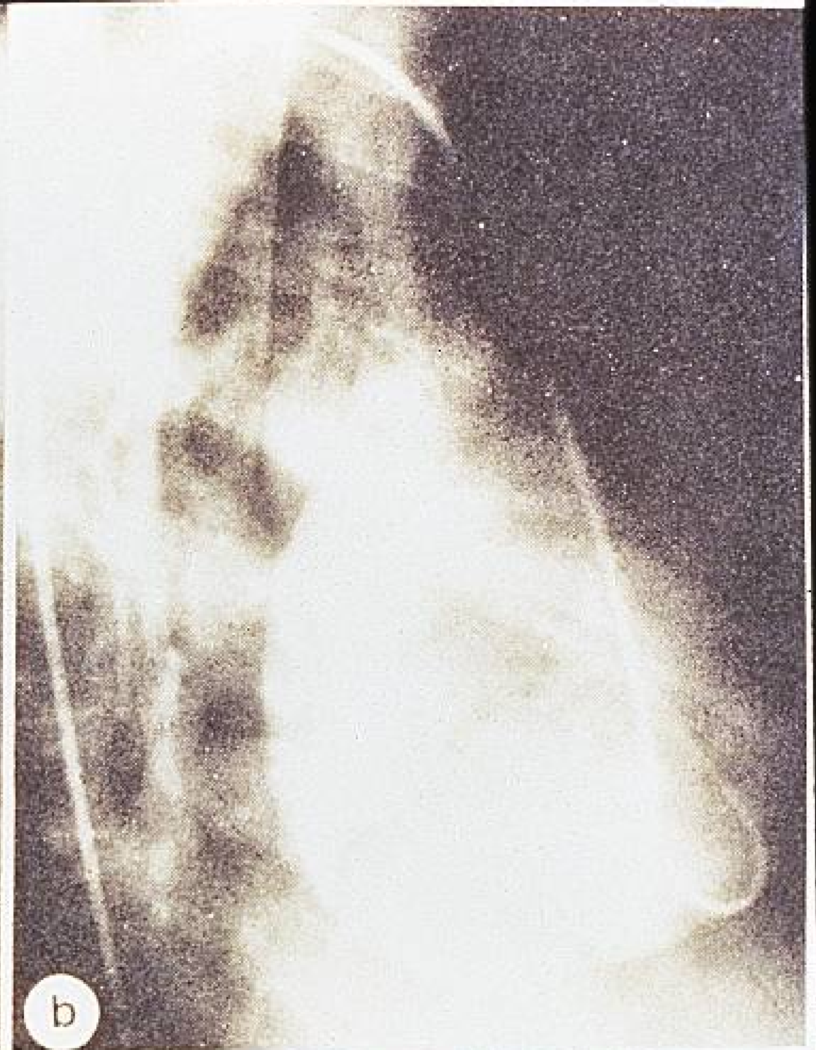
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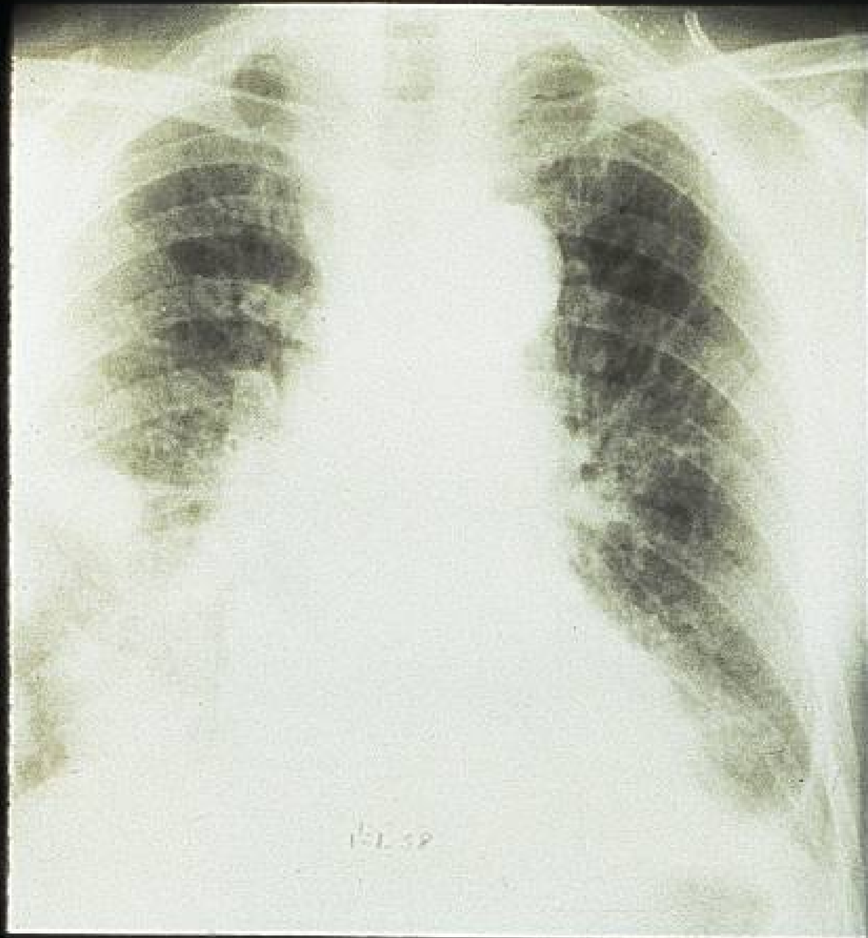
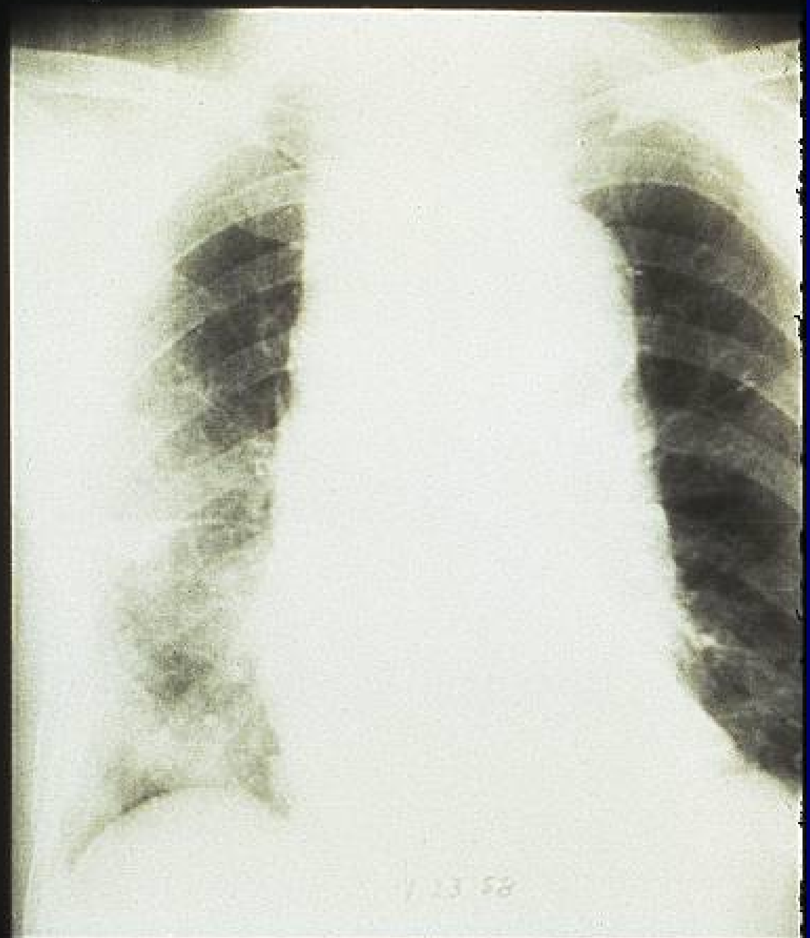
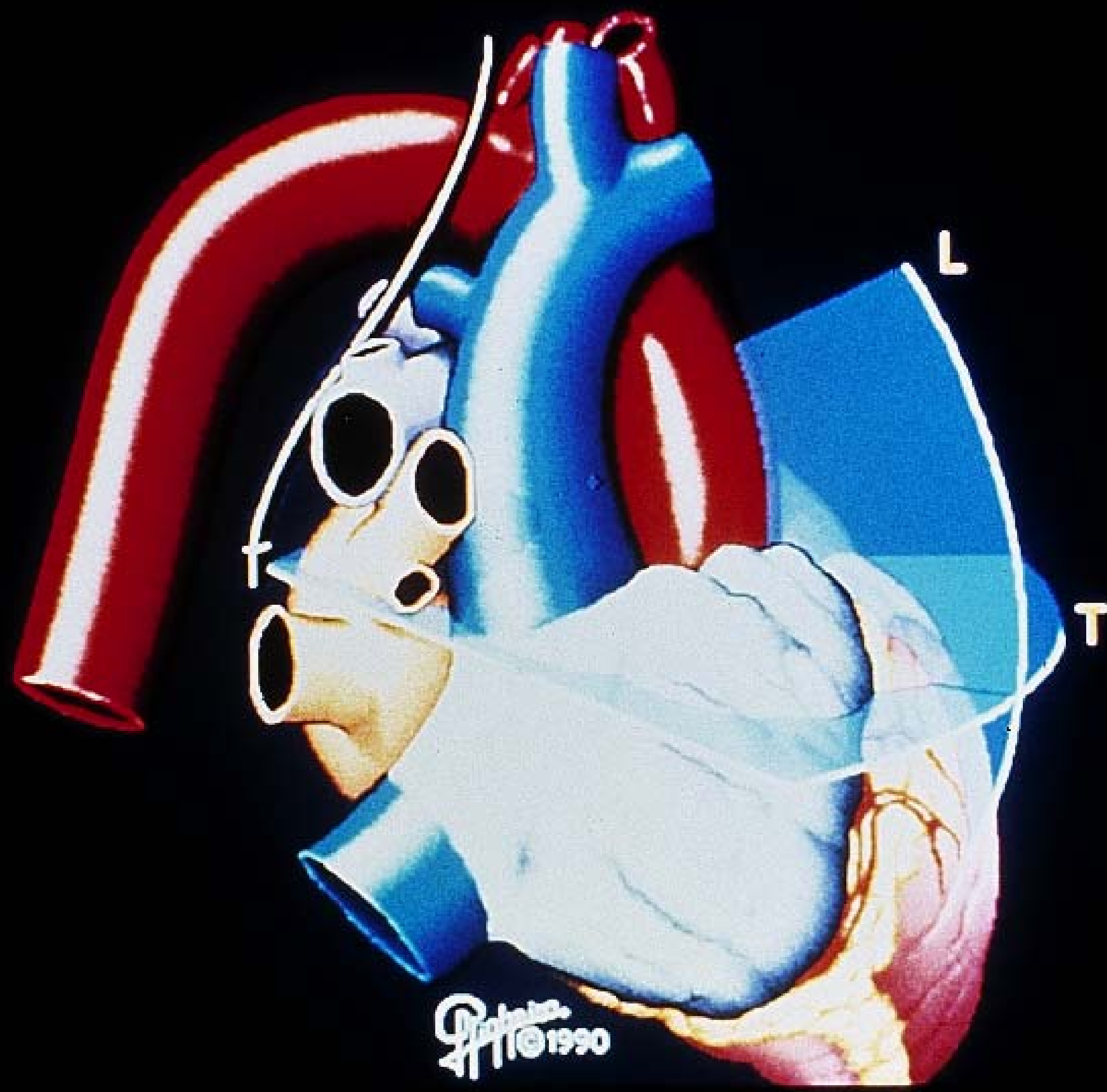




TRANSESOPHAGEAL ECHO

- Procedure of first choice for dissection, if readily available
- Portability of equipment facilities emergency management by cardiologist in ER or ICU
- High sensitivity and specificity
- Additional information:
  - aortic regurgitation
  - pericardial effusion
  - ostia of coronary arteries
  - LV function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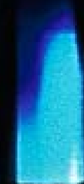

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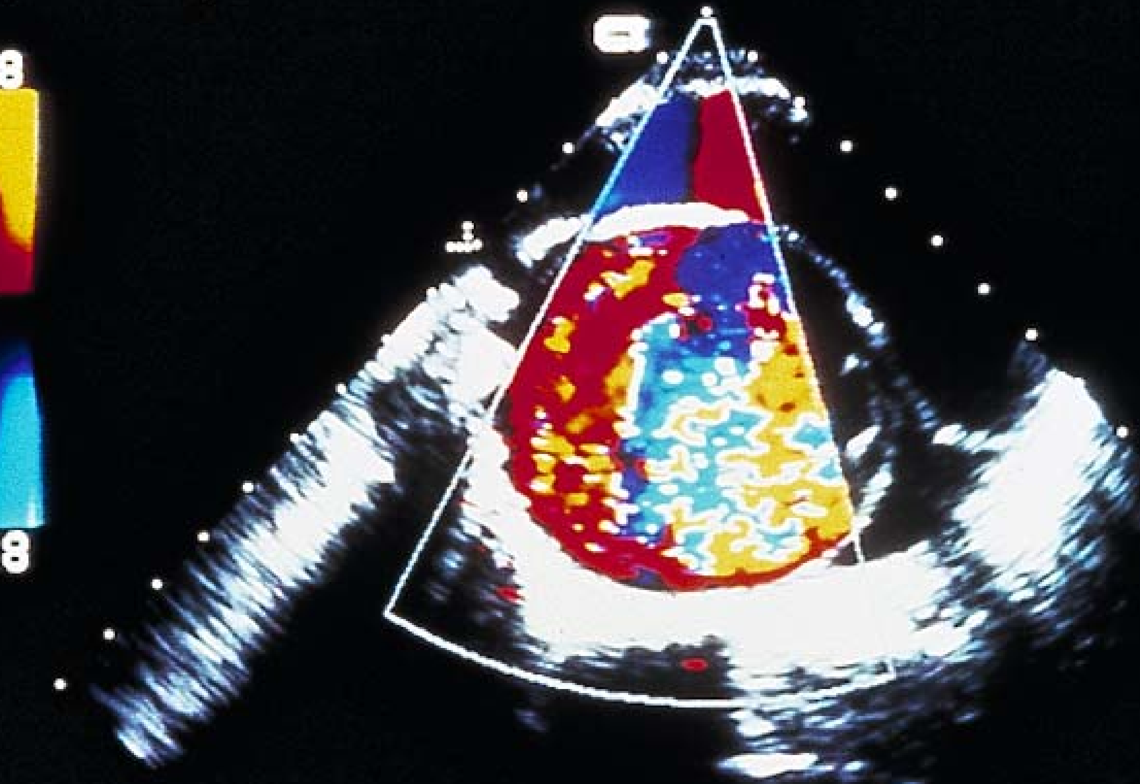

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ST.JOES /V

PWR<50FET
A/0/VV5
1/1
MAX SCALE

COMPUTED TOMOGRAPHY

- Rapid IV bolus and sequential imaging
- 88-100% accuracy
- Limitations:
  - no evaluation of aortic regurgitation
  - Limited information on branch vessels
  - "streak artefacts" may cause false (+)
    (Bone-air interface may simulate flap)
  - False (-) from poor bolus of contrast
- Useful for follow-up of dissections

MRI

- Good alternative to TEE or CT, if readily available
- High sensitivity and specificity
- Can detect slow blood leaks
- Non-invasive; neither x-rays nor contrast needed
- 90-100% accuracy
- Limitation: requires patient to be in claustrophobic apparatus without standard ECG monitoring
- Useful for follow-up of dissections

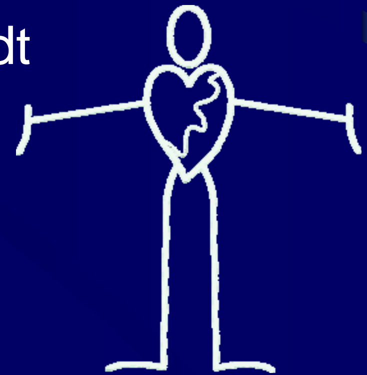
AORTOGRAPHY

- Identify intimal flap, true and false lumen
- Thickened wall (thrombosed false lumen)
- Aortic insufficiency, branch vessel involvement
- Diagnostic accuracy 90-95%

  5-10% false (-) rate
  - thrombosed false lumen
  - simultaneous opacification of both lumens
  - misses intramural hematoma
- Risks of procedure (time delay in type A)
- Coronary angiography usually not necessary in acute ascending dissection (surgeon can inspect coronaries)

TREATMENT

1. ICU admission in a tertiary center
2. Immediate cardiothoracic surgical consultation
3. Close observation of BP, urine output, neurologic status
4. Prompt blood pressure control is critical
   - Can reduce propagation of dissection
   - Decrease BP and LV contractility to decrease dP/dt
     - Sodium nitroprusside + b – blocker
     - α and β - blocker, Calcium channel blocker (heart rate slowing)



INDICATIONS FOR SURGERY

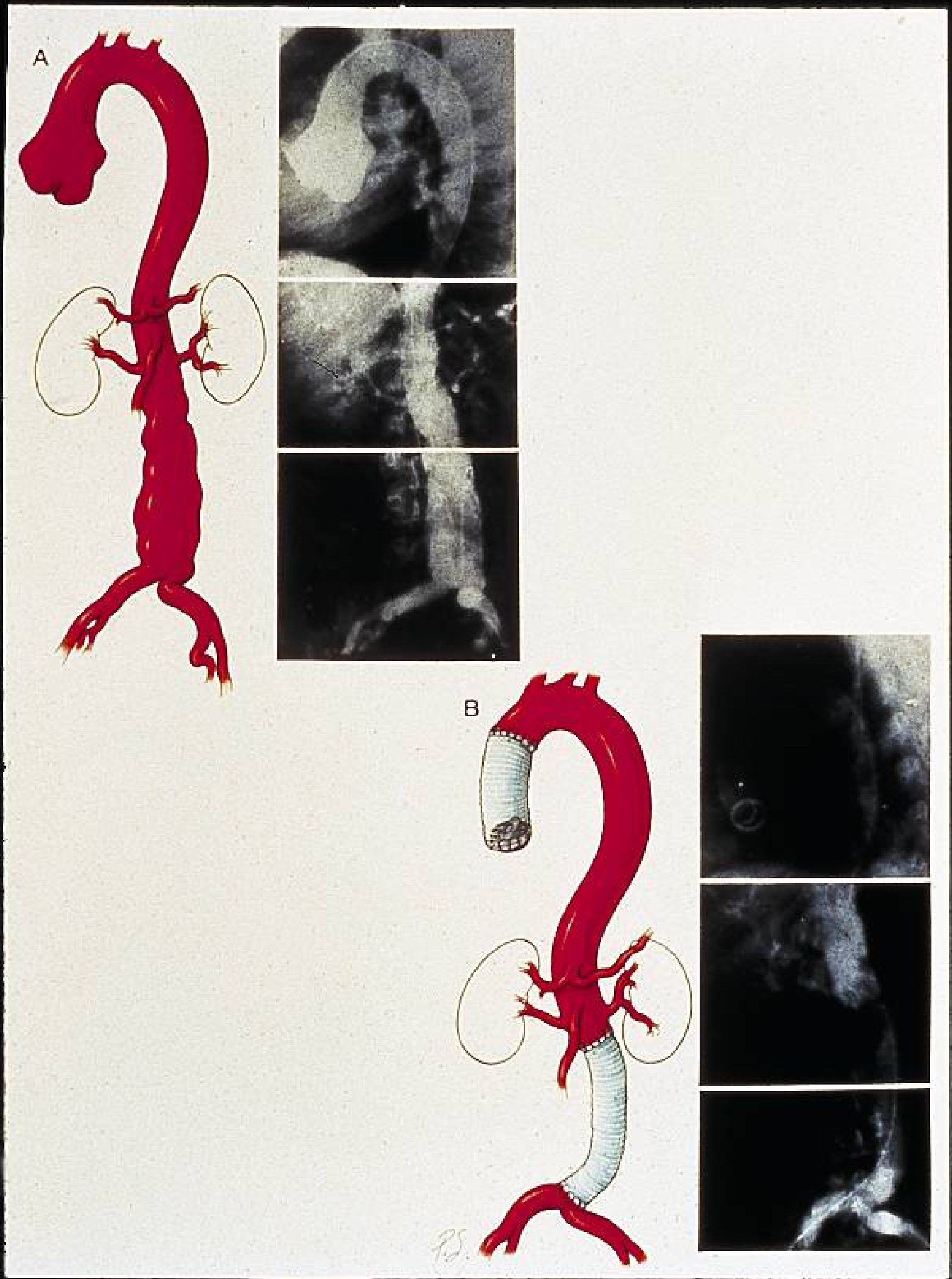
- Consider operative treatment for all patients
- Hypotension: Emergency surgery
- Ascending Dissection: Emergency surgery
- Descending Dissection:
    - Operation in acute phase no difference in survival compared to medical therapy 35-75% mortality
    - Higher risk of surgery with renal failure, visceral ischemia, age > 70 years
- Risk of surgery inversely related to experience with dissection surgery

GOALS OF SURGERY

1. Excise the intimal tear
2. Obliterate entry into false lumen proximally and distally
3. Reconstitute the aorta (Dacron graft)

GOALS OF SURGERY cont'd.

- If aortic regurgitation complicates ascending dissection:
    - Surgical decompression of false lumen and resuspension AV leaflets
    - Aortic valve replacement required if annular supports of leaflets damaged (composite graft or homograft)
    - Aortic valve replacement required if aortic root >5 cm (likely to progress)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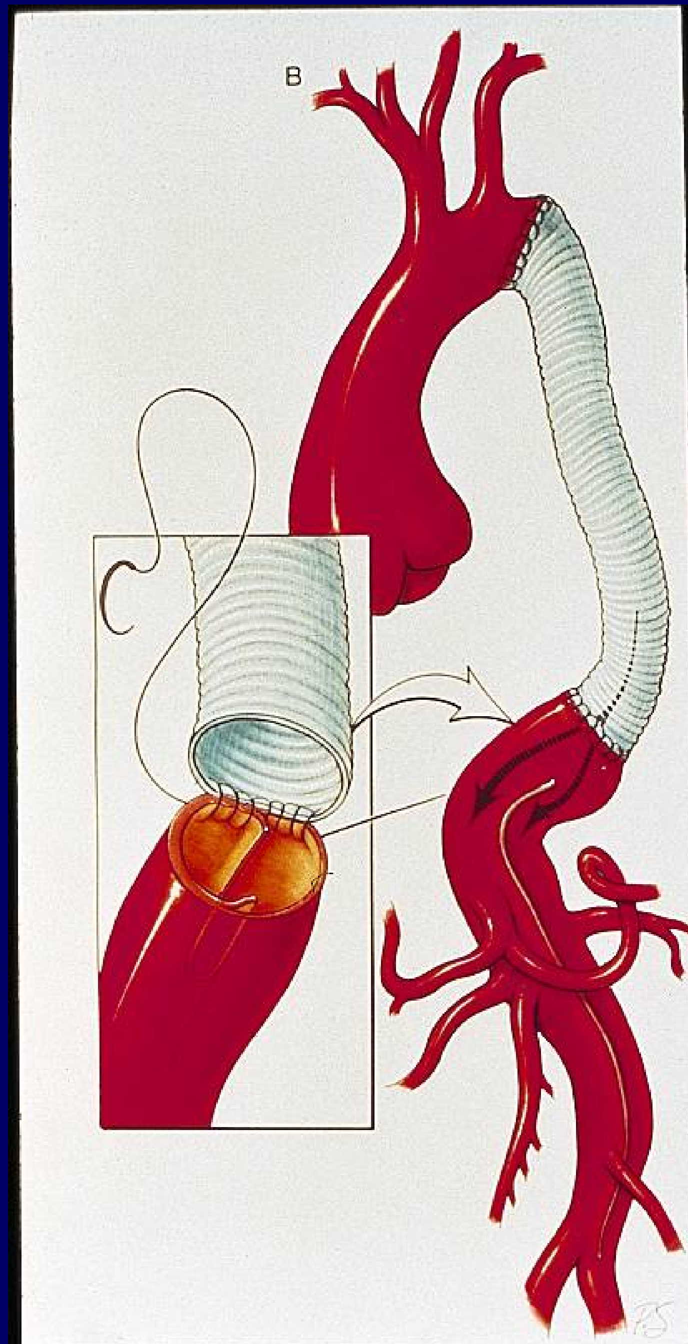
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INDICATIONS FOR SURGICAL THERAPY

- Hemorrhage or rupture
- End-organ ischemia
- Continued pain
- Rapid expansion ($\geq$5mm in 6 months) of diameter of any segment $\geq$6 cm (less in some centers)
- Uncontrolled HTN
- Younger patients at relatively good operative risk

FOLLOW-UP

- Aneurysmal dilation and rupture are leading causes of late death
- 1982 DeBakey. 527 pts operated for aortic dissection: 30% of late deaths due to rupture of post-dissection aneurysms
- 1990 Crawford. Death from rupture occurred in 12/130 (9%) cases with a dilated but unrepaired residual aorta
- After extensive aortic dissection, many patients will eventually require surgical therapy (especially if on anticoagulants after composite graft repair)

FOLLOW-UP cont'd.

1. Before hospital discharge: CT scan or MRI

2. Initial evaluation after discharge:
   CT scan or MRI at 3, 6 and 12 months

3. Reimage aorta every 6-12 months thereafter

4. Meticulous control of blood pressure dP/dt
   (starting with b-blocker or heart rate
   slowing calcium channel blocker)

   Avoid isometric exercise.

SURGICAL THERAPY

1. Early operations attempted to create reentry passage or restoration of circulation to ischemic branches.
   High failure rate.

2. 1935 Gurin: Fenestrated dissecting membrane in iliac artery.

3. 1948 Paullin and James: Wrapped chronic dissection of descending aorta with cellophane.

SURGICAL THERAPY cont'd.

4. 1955 Michael DeBakey: Modern treatment of aortic dissection

   Collegues: Denton Cooley and O' Creech

   1st case: Descending thoracic dissection

   - Excision of aneurysmal dilation
   - Oversewed distal entry into the false channel
   - End-to-end anastomosis of aorta

   Later: Dacron graft replacement of descending aorta

5. 1962 Spencer and Blake: First successful repair of chronic ascending dissection with resuspension of aortic valve commissures

SURGICAL THERAPY cont'd.

6. 1960's: Importance of sandwiching the friable aortic layers between strips of Teflon felt

7. Bentall procedure

8. Evolution of the composite graft approach

9. Aortic homograft

10. Aortic conduit with AV sparing

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