DISEASE OF GREAT VESSELS

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DISEASE OF GREAT VESSELS

- Congenital
- Acquired

Congenital:
- PDA
- Coarctation of aorta
- Aortopulmonary window
- Anomalies of aortic arch
- Aneurysm of the sinuses of Valsalva

Acquired:
- Thoracic aortic aneurysm
- Aortic dissection
- Occlusive disease of branches of the aorta

Patent ductus arteriosus:
- Derived from 6th aortic arch and normally extends from the main PA or left PA to descending aorta just below the origin of left subclavian a.
- Rarely may be right side PDA or bilateral PDA
- It is essential during intrauterine life

Closure of PDA occurs at birth
- Functional closure occurs in 10-15 hrs. due to decreased pulmonary vascular resistance and increase in oxygen saturation, causing constriction of the smooth muscle layer
- Anatomical closure in 3 wks.
- Delayed closure is uncommon after 6 mo.
- High oxygen tension causes constriction of ductus
- Prostaglandin E dilated ductus
- Persistent patency of ductus results in L>R shunt

Incidence
- Once in 2500-5000 live births
- Increase in premature, 80% PDA in B.W < 1000 gm.
- Related with maternal rubella
- M.R. of untreated PDA ~ 30%
  - Average age of death: 24 years
  - Pt die of bacterial endocarditis
  - Congestive heart failure
Clinical manifestation
- depends on the size of PDA, PVR, age and associated anomalies
- can present with - congestive heart failure
  - failure to thrive
  - irritable
  - tachycardia
  - take feeding poorly
- pt. usually become symptomatic after 6-8 wk of life due to decreased pulmonary vascular resistance at this time

Diagnosis - continuous murmur LUSB
- CXR - cardiomegaly
  - pulmonary congestion
- EKG - LVH
- Echocardiogram: demonstrate PDA
  if LA/RA ratio > 1.5 : 1-L>R shunt
- cardiac cath indicated in adult or pt with atypical findings & suspicion of associated anomalies or pulm. HTN
- CHF
- bacterial endocarditis
- aneurysmal dilatation and rupture of PDA
- Eisenmenger complex

Management:
- medical : in premature infant try indomethacin success rate ~ 70%
- non operative therapy:
  - transcatheater closure with Ivalon plug
  - double umbrella device (Rash kind), coil
- Surgical therapy
  - multiple ligations
  - division
  - calcified ductus in older patients required cardiopulmonary bypass and patch closure

Result:
- Surgical closure is safe
- pt with PDA closure have a normal life expectancy
- Increased mortality and poor results occur in older patient with calcified ductus, and severe pulmonary hypertension and reverse shunting
Aortopulmonary window

- rare congenital defect due to abnormal septation of truncus into the aorta and PA
- other terminology include
  - aorto pulmonary fistula
  - aortic septal defect
  - aortopulmonary septal defect
  - aortopulmonary fenestration
related defect
- anomalous origin of right PA from Aorta

Embryology & Pathologic Anatomy
- failure of fusion or malalignment of the conotruncal ridges leads to AP septum defect
- abnormal migration of 6th aortic arch may result in aortic origin of PA
* Aortic & pulmonic valve are normally formed distinguishing from persistent truncus arteriosus
- lesion begins a few millimeters above aortic valve on the left lateral wall of the aorta
- mostly single defect, multiple defects are rare
- defect may be found distally overlying the origin of right PA

natural history:
- pt with large AP window usually do not survive infancy
- CHF & pulmonary hypertension occur early in life
- clinical course similar to pt with large VSD

Diagnosis
- CHF early in life
- Growth retardation
- recurrent pulmonary infections
- systolic or continuous murmur similar to PDA
- CXR - cardiomegaly
  - pulm. vascular engorgement
  - CHF
- DDX - PDA
  - persistent truncus arteriosus
  - VSD with AR
  - ruptured aneurysm of the sinus of valsalva
- 2-D echocardiogram
  - show the defect
- Cardiac cath
  - O2 sat. step up at the level of PA
  - Retrograde aortography confirmed Dx
Surgical treatment
- Trans aortic closure
  - Direct closure
  - Patch closure
- Simple ligation, division & primary closure should be discouraged
- Transpulmonic closure can also be done but transaortic is preferred because of better visualization of the defect as well as the coronary ostia.
- Aortic origin of the Rt PA is best repaired by division of the right PA and direct reanastomosis to the main PA.

Coarctation of the Aorta
- Localized or diffused obstruction of the segment of the aorta
- Most common location: below left subclavian take off

Etiology: Two theories
1. Extension of ductal tissue into the aortic wall causing localized narrowing
2. Abnormal fetal blood flow patterns

Incidence: 5-10% of congenital heart dis.

Associated anomalies:
- Bicuspid aortic valve
- VSD
- PDA
- Mitral valve disorders

Clinical manifestation:
- Arm leg systolic pressure gradient
- Hypotension, oliguria & severe metabolic acidosis may be presented in severely ill infants
- Differential cyanosis
- Unexplained hypertension
- Complication of hypertension
- Headache, epistaxis
- Visual disturbance
- Exertional dyspnea
- Aortic rupture
- Dissecting aneurysm

Diagnosis:
- Hypertension
- Systolic pressure gradient between arms and legs
- Systolic murmur over the left precordium & between scapulas
- Diminish or absent femoral pulses with a delayed upstroke

EKG: RVH, LVH, LV strain
CXR: cardiomegaly, LVH
  - Rib notching
  - "3" sign
- Angiogram - pressure gradient across the lesion
- 2-D Echocardiography
- CT scan
- MRI
- DSA
Natural history
- 74% die by 40 years of age
- average age at death = 32 years (Abbott)

Causes of death:
- Spontaneous rupture of aorta
- Bacterial endocarditis
- CHF
- CVA

Campbell (1970)
- 25% die by 20 years of age
- 50% die by 20 years of age
- 75% die by 20 years of age
- 90% die by 20 years of age

Pseudocoarctation
- buckling or kinking of the aorta without obstruction to flow, considered benign
- CXR reveals abnormal aortic contour mimicking a left superior mediastinal mass
- Dx confirmed by aortogram
- Ruptured of aorta due to pseudocoarctation has been reported
- Surgical intervention should be done in pt developing aortic dilatation

Physiology of Hypertension
- Pathogenesis of hypertension
  - mechanical: due to narrowed segment
  - renal factor: Goldblatt’s model
  - Renin angiotensin activation

Management
- Prostaglandin E. to open PDA for perfusion of lower extremities
- Correct acidosis
- Surgical correction as soon as pt is stabilized and in optimal condition in symptomatic patients
- in elective repair, it should be done in the early age ≡ (1-6 yr. of life) to prevent persistent hypertension

Surgical treatment
- Resection with end to end anastomosis
- Resection with interposition graft
- Patch repair
- Subclavian flap

Non operative therapy
- Percutaneous balloon angioplasty

Complications:
- Hemorrhage
- Chylothorax
- Recurrent nerve palsy
- Infection
- Suture line thrombosis
- Paradoxical hypertension
- Post coarctectomy syndrome
- Paraplegia

**Result**
- Operative mortality ≈ 5-10% in neonates
- Recoarctation are more or less the same for end to end anastomosis, subclavian flap or extended resection with anastomosis
- Pt operated between 2-4 years had lowest risk for restenosis and persistent hypertension

**Interruption of the Aortic Arch**

- Complete absence of a segment of the aortic arch without any anatomic connection between the proximal & distal segments
- Incidence: < 1.5% of congenital heart dis.

**Classifications**: 3 types:
- Type A: Interruption distal to left subclavian a. (43%)
- Type B: Interruption between left subclavian (53%) and left common carotid arteries
- Type C: Interruption between left common carotid and innominate arteries (4%)

If anomalous origin of right subclavian a. is present they are designated as A1, B1, C1

**Etiology**: - unclear
  - may be abnormal fetal blood flow pattern

**Natural history**: - uncorrected interruption of the aortic arch carried a poor prognosis
  - mean age of death = 4-10 days
  - 90% die in the first year of life

**Dx and management**
- mostly present with CHF secondary to L → R shunt
- lower body perfusion maintained by R → L shunt thru PDA
- when ductus closes, perfusion of the lower body essentially ceases and infants become anuric, severely acidotic and femoral pulses become nonpalpable
- CHF & acidosis are resistant to medical therapy
- Diagnosis made by cardiac cath with contrast injection both proximal & distal segments to define anatomy adequately
Surgical treatment consisted of
- direct end to side anastomosis
- interposition graft
- primary repair may be done using CPB

Anomalies of the Aortic Arch

Embryology & Pathologic anatomy
- 6 pairs of aortic arches with normal regression and persistence results in normal pattern of aorta, pulmonary artery, great vessels
- In normal development
- 3rd arch forms parts of common carotid a.
- Left 4th arch forms aortic arch
- proximal of right 4th arch persist as the innominate artery
- proximal right and left sixth aortic arches form pulmonary arteries
- distal left 6th arch → ductus arteriosus
- distal right sixth arch regresses (if persist, it will result in a vascular ring)

Anomalies of the aortic arch
1. Double aortic arch
2. Aberrant right subclavian artery
3. Anomalies of the innominate artery and brachiocephalic arteries
4. Subclavian steal syndrome
5. Circumflex retroesophageal aortic arch
6. Right aortic arch
   - right aortic arch with mirror image branching
   - right aortic arch with aberrant left subclavian artery
   - right aortic arch with isolated left subclavian artery

7. Cervical aortic arch
8. Anomalous left pulmonary artery
9. Interrupted aortic arch
10. Pseudocoarctation of aorta
11. Anomalous origin of a pulmonary artery from ascending aorta
12. Absent pulmonary artery
13. Persistent fifth aortic arch

1. Double aortic arches

Pathology:
- Splitting of ascending aorta into two limbs
- Pass on either side of trachea & esophagus and join as a single descending aorta
- Left arch tends to be smaller than right
- Persistent of right & left 4th aortic arch
Diagnosis:
- Echocardiogram
- CT scan
- DSA
- Barium swallow, lateral esophagogram shows an oblique posterior impression coursing upward from left to right and arterial pulsation may be seen

Treatment:
- asymptomatic pts require no treatment
- ligation and division in children
- In adult division alone can cause subclavian steal, and reanastomosis to the aorta may be necessary

3. Anomalies of the innominate artery
- abnormal course may compress the anterior trachea
- can cause reflex apnea (respiratory arrest following stimulation of the compressed area of the trachea
- less than 14% required surgical intervention

Clinical manifestation
- respiratory distress
- stridor
- respiratory arrest
- dysphagia does not occur because the esophagus is not obstructed
- recurrent pneumonia

Diagnosis:
- Physical exam is not helpful
- CXR - only pneumonia
- atelectasis
- Barium swallow - normal
- angiogram: abnormally left ward origin of the innominate artery but not diagnostic of tracheal compression
- Bronchoscopy - demonstrates tracheal compression

Treatment:
- usually pt will become progressively, less symptomatic with time
- syndrome rarely seen past 1 year of age
- Surgical technique include:
  - aortopexy via median sternotomy or right anterior thoracotomy
  - or reimplantation of the innominate artery

4. Subclavian steal syndrome
- normally, vertebral a. originates from subclavian a. If there is pathologic interruptions of proximal subclavian a. flow, which create low pressure distal to the obstruction, and encourage retrograde flow from the vertebral a into the subclavian a. which results in reduction of flow in to the basilar system and precipitate cerebral ischemia. Such reduction of basilar flow depends on either exercise of the affected arm or in adequate collateral circulation to the circle of Willis.
- Example in children. Coarctation of aorta or interrupted arch with aberrant right subclavian a. it will serve as a site for diversion of blood from vertebral basilar system if the origin is distal to coarctation of aorta
- Infant with large VSD or PDA may remain asymptomatic due to high pressure flow from the PDA

**Clinical manifestation**
- Symptom of cerebral ischemia are rare in infants due to the commonly associated findings of VSD or PDA
- Symptoms include - arm claudication
  - syncope

**Diagnosis**
- History & physical exam.
- CXR & Barium swallow nonspecific
- Angiogram is diagnostic, subclavian a. will be noted to fill late in its distal portion by retrograde flow from the vertebral a.

**Treatment**
- cerebral symptoms usually appear after age 20
- surgical intervention
- ascending aorta to junction of subclavian & vertebral a. bypass.
- In case of coarctation of aorta or interruption of aorta, correction of primary congenital defect is indicated

5. **Circumflex retroesophageal aortic arch**
- Combination of left aortic arch and right descending aorta create potential vascular ring due to retroesophageal course of aorta
- Ring is completed by PDA or ligamentum arteriosum
- Compression of esophagus in more significant than the trachea

**Clinical manifestation**
- usually no symptoms due to mild compression
- incidental finding during investigative W/U for congenital heart defect
- may present with - dysphagia
  - regurgitation of feedings
  - occasional resp. symptoms

**Radiographic features**
- CXR
- Barium swallow
- Angiogram
- CT scan may be helpful

**Treatment**
- in pt with severe symptoms
- surgical division of PDA or ligamentum
- mobilization of the esophagus
- Rt thoracotomy is preferred
6. Right aortic arch
- aorta ascends anterior to the right bronchus and arch to the right of the trachea and descends on the right
- classification of right side aortic arch
  6.1 right aortic arch with mirror image branching
  6.2 right aortic arch with aberrant left subclavian artery
  6.3 right aortic arch with isolated left subclavian artery

6.1 Right aortic arch with mirror image branching
- mirror image branching is the most frequent arrangement of the arch in pt with right arch
- 1st branch is left innominate a.
- 2nd branch is right common carotid a.
- 3rd branch is right subclavian a.
- high association with congenital heart dis (98%)
- TOF most common (48%)
- PA with VSD
- DORV
- Truncus arteriosus
- agenesis of the left PA

Clinical manifestation
- discovered during evaluation of CHD
- rarely symptomatic
- bronchomalacia or tracheomalacia may occur in severe cases which cause respiratory failure & hyperinflation of the lung
- bronchial rather than tracheal compression should alert the physician to look for atypical aortic arch anomalies

Diagnosis:
- CXR
- Brain swallow
- 2-D Echocardiography
- CT scan
- Angiogram

Treatment:
- Division of PDA
- Suturing the aortic stump to the prevertebral fascia

6.2 Right aortic arch with aberrant left subclavian a.
- give off branches in the following orders
- left common carotid a.
- right common carotid a.
- right subclavian a.
- left subclavian a. from descending aorta crossing behind the esophagus to reach the left arm

Clinical manifestation
- only those with left side PDA (50%) have vascular ring
Radiographic Features
- Barium swallow: small indentation of posterior esophagus

Treatment: In severe symptoms
- Division of PDA or ligamentum

6.3 Right aortic arch with isolated left subclavian a.
- 1st three branches
  - left common carotid
  - right common carotid
  - right subclavian a.
  - left subclavian a. originates from the ductus arteriosus
  - associated with TOF

Manifestations
- present as subclavian steal syndrome
- or discovered as part of evaluation for congenital heart disease or subclavian steal syndrome

Treatment
- directed toward underlying congenital heart disease or subclavian steal syndrome

7. Cervical aortic arch
- Rare
- Comprise of an ascending aorta that extends high in the mediastinum - before turning sharply to descend through the thorax
- Tracheoesophageal compression can be associated
- Ligamentum arteriosum complete the ring

Clinical features
- pulsatile mass in the neck
- may present with symptoms of vascular ring

Diagnosis: - CXR
  - widening of the mediastinum
  - absent aortic knob
  - anterior displacement of trachea
  - Barium swallow - posterior compression of the esophagus
  - Echocardiography
  - Angiography

Treatment: - Surgical division of PDA or ligamentum arteriosum in symptomatic patients.

8. Abnormal left pulmonary artery

Pathology: - left PA may be absent
  - left PA - arise from Rt PA
  - form ascending aorta extend from the arch via ductus arteriosus
  - usually abnormal left PA means pulmonary artery sling in which left PA originates from the right PA cross the right main bronchus and courses between the trachea and esophagus to the left lung
- hypoplasia of distal trachea or right mainstem bronchus, stenosis of left mainstem bronchus, proximal stenosis of trachea, occur in 54% of cases

**Clinical manifestation**
- Respiratory symptoms due to airway compression
- Differential points from vascular ring
  - lower frequency of apnea
  - stridor expiratory phase
  - more emphysema and lung collapse
- Dysphagia & vomiting are rare

**Diagnostic Method**
- CXR - air trapping
  - Collapsed lung
  - Rt lung hyperlucent
  - Trachea deviates to the right
  - Lateral film show mediastinal mass at the carina
- Barium swallow
- oval mass between trachea & esophagus
- Bronchography - abnormality of tracheobronchial tree
- Cardiac cath & pulmonary angiogram

**Treatment**:
- Surgical division & reimplantation of the anomalous left PA
- Symptoms may not be relieved due to associated abnormalities of the tracheobronchial tree
- M.R. ≥ 50%

**Anomalous origin of a PA from ascending aorta**
* PA originates just distal to the aortic valve
* Causes by defect in fusion of opposing conotruncal ridges
* Elevated pulmonary vascular resistance in older children

**Clinical features**
- Congestive heart failure
- Cyanosis
- Respiratory distress
- Accentuated P2

**Radiologic features**
- CXR * moderate cardiomegaly
  * increased pulm. blood flow
- Aortography is diagnostic

**Treatment**
- Surgical anastomosis of the anomalous PA to the PA
- Early surgery is desirable to prevent pulmonary vascular obstructive disease
**Absent Pulmonary Artery**

- Unilateral absence of PA, usually on the side opposite the arch
- Associated with diverticulum of the innominate a.
- TOF is the most common associated CHD (ass. with absent left PA)
- PDA mostly associated with absent right PA
- Other anomalies
  - Truncus arteriosus
  - D-transposition of great vvs.
  - Coarctation of aorta
  - VSD
  - Aberrant subclavian a.
- Blood supply is generally from bronchial collaterals or from aortic arch, these vvs. have histologic characteristic of a ductus
- Pulmonary hypertension occur in normal lung but uncommon in the affected lung

**Clinical features**
- Heart murmur
- Cyanosis
- Respiratory symptoms
- Shortness of breath
- Smaller hemithorax & lung on the affected side
- Shift of the heart to the affected side

**Radiographic features**
- CXR
  - * discrepant blood flow between the two side
  - * shift of the heart and mediastinum
  - * absent or aberrant hilar comma

**Angiography** - confirm the diagnosis

**Treatment**
- minimize the progression toward severe pulm. hypertension
- associated shunt lesion should be corrected in early age

**Persistent Fifth Aortic Arch**

- made up of a band of arterial tissue that divides the aortic arch into a larger superior channel and a smaller, inferior channel
- The fourth arch give rise to the superior channel
- The inferior lumen connects the innominate artery and left subclavian a.
- Associated congenital heart disease, including
  - TA
  - Cortriatriatum
  - VSD
  - PS
  - PDA
  - Bicuspid aortic valve
  - Single right coronary artery
Clinical manifestation
- usually discovered during work up for congenital heart defect
- aortography - parallel horizontal channels on the lateral view
- Barium swallow - negative

Occlusive disease of branches of the aorta
- Ischemic disturbance of the head, neck, upper extremities with absent or decreased pulses
- 5-15% of extracranial lesion producing cerebral symptoms

Etiology:
- Atherosclerosis
- Nonspecific arteritis (Takayasu's disease)
- Syphilis
- Tuberculosis
- Periarteritis nodosa
- Collagen - vascular disease
- Rheumatic fever
- Trauma
- Congenital malformations
- Radiation injury
- Fibromuscular dysplasia
- Aneurysmal disease
- Aortic dissection
- Tumor and scar

Clinical manifestation
- Sign & Symptoms depend on
  - the location
  - nature of the lesion (ulcerative or obstructive)
  - extent of obstruction
  - degree of collateral circulation

Pathophysiology
1. diminished forward flow thru the involved vessels
2. reverse of cerebral blood flow from the brain
3. ulcerative lesions that discharge emboli into the involved vessels

Symptoms of inadequate cerebral circulation
- unilateral impairment of motor or sensory function
- syncope
- headache
- confusion
- speech disorder
- tinnitus
- impaired vision
- convulsion & paralysis
Cerebellar symptoms from vertebral a lesion
- vision loss
- vertigo
- ataxia
- dizziness
- syncope
- visual hallucination

Upper extremities ischemic symptoms
- Claudication
- Weak or tire easily
- embolism

Diagnosis:
- History
- Bruits
- Upper extremity pressure gradients
- Pulse deficit
- Arch aortogram
- MRI
- Non surgical therapy
  - percutaneous transluminal angioplasty (PTA)

Surgical therapy:
1. Endarterectomy
2. Bypass

Result: 95% relief of symptoms
Aortic Dissection

1. **Definition**
- not an aneurysm
- Acute < 2 wk.
- Chronic > 2 wk.
- DeBakey Classification
  - Type I - from ascending aorta to descending Ao.
  - Type II - tear & dissection limited to Asc Ao.
    (Type I + Type II = type A)
  - Type III or type B - tear & dissection originated in descending Ao.

![Type A and Type B Diagram]

Fig 1. Classification of aortic dissections and sites of the intimal tear.

2. **Incidence**
- most common catastrophe involving Ao.
- 5-10 cases per million each year
- 2-3 times that of ruptured Abd. aortic aneurysm
- Mean age - type A = 49 (16-77)
- type B = 60 (38-83)
- M:F = 3:1

<table>
<thead>
<tr>
<th></th>
<th>Type A</th>
<th>Type B</th>
</tr>
</thead>
<tbody>
<tr>
<td>Frequency</td>
<td>85-70%</td>
<td>30-35%</td>
</tr>
<tr>
<td>Male-female ratio</td>
<td>2:1</td>
<td>3:1</td>
</tr>
<tr>
<td>Average age</td>
<td>60-70</td>
<td>60-70</td>
</tr>
<tr>
<td>Associated hypertension</td>
<td>50%</td>
<td>80%</td>
</tr>
<tr>
<td>Hypertension on admission</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Associated atherosclerosis</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Aortic regurgitation</td>
<td>50%</td>
<td>10%</td>
</tr>
<tr>
<td>Intimal tear</td>
<td>Always present</td>
<td>Absent in 5-10% of patients</td>
</tr>
<tr>
<td>Acute mortality</td>
<td>90-95%</td>
<td>40%</td>
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</tbody>
</table>

Table 1. Characteristics of Type A and Type B
3. **Etiology**
- HTN in 75-85%
- Connective tissue abnormality e.g. Marfan's Syndrome
- Associated Condition, Coarctation of Aorta, pregnancy, kyphoscoliosis
- Atherosclerosis & syphilis - not proven

4. **Pathologic mechanisms**
- Degeneration of cellular components of Aortic media
- Intimal tear resulting from degeneration of and excessive tension within the media
- Propagation of the dissecting hematoma
- Related to magnitude and/or velocity of arterial pulse i.e. \( \frac{dp}{dt_{\text{max}}} \)
- may also be related to absolute level of blood pressure

5. **Natural History**
- mortality
  - 3% sudden death
  - 28% dead with in 24 hrs.
  - 50% dead with in 48 hrs.
  - 90% mortality at 3 mo.
- morbidity & mortality related to propagation of the dissection
- pericardial tamponade
- left hemotherax
- Aortic valve incompetence
- right atrial communication
- major arterial compromise
- involvement of coronary arteries
6. **Clinical manifestation**
- Pain, sudden, tearing or ripping in character usually in chest
- Signs of acute aortic incompetence
- Acute neurologic change
- Hypotension or shock (tamponade, hemothorax)
- Change in peripheral pulse characteristics
- Acute SVC. compression

<table>
<thead>
<tr>
<th></th>
<th>Type A</th>
<th>Type B</th>
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</thead>
<tbody>
<tr>
<td>Pain</td>
<td>Anterior substernal</td>
<td>Posterior midscapular, Abdominal</td>
</tr>
<tr>
<td>Syncope</td>
<td>+</td>
<td>Rare</td>
</tr>
<tr>
<td>Dipse</td>
<td>+</td>
<td>80%</td>
</tr>
<tr>
<td>Arterial pressure on admission</td>
<td>Normal or elevated—50%</td>
<td>Hypotensive—20%</td>
</tr>
<tr>
<td>Asymmetric pulses</td>
<td>Upper extremity (30–50%)</td>
<td>Lower extremity</td>
</tr>
<tr>
<td>Diastolic murmur</td>
<td>50%</td>
<td>10%</td>
</tr>
<tr>
<td>Pericardial effusion</td>
<td>+ +</td>
<td>Rare</td>
</tr>
<tr>
<td>Pleural effusion</td>
<td>+</td>
<td>+ (Left pleura)</td>
</tr>
<tr>
<td>Hemiparesis or hemiplegia</td>
<td>+</td>
<td>—</td>
</tr>
<tr>
<td>Paresis or paraplegia</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Renal–intestinal infarction</td>
<td>+ (2–3%)</td>
<td>+</td>
</tr>
<tr>
<td>Myocardial infarction</td>
<td>+</td>
<td>Rare</td>
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Table 2. Symptoms and signs of acute aortic dissection

7. **Diagnosis**
- CXR widened mediastinum, abnormalities of the aortic knob,
  increased aortic diameter, displacement of calcified intima, deviation of trachea
- EKG - LVH, must rule out AMI
- Mild leukocytosis, acute anemia
- Aortography
- Locate intimal tear
- Define extent of dissection
- Examine deformation of the true lumen
- Determine presence, absence or degree of aortic valve incompetence
- Determine major branch involvement
- CT scan
- Echocardiogram
8. Treatment
- Type A - Emergent surgery
- 88% MR with medical treatment
- 23% MR with surgical treatment
- Type B - medical treatment
- anti-impulse
- antihypertensive
- endpoint of medical therapy is lack of evidence of propagation of dissection
- dissection treated medically have 32% MR, with surgical treatment 36% MR

Fig 2. Surgical treatment of acute type A dissection
Fig 3. Partial arch replacement for acute arch dissections

Fig 4. Surgical treatment of type B. dissections
Fig 5. Surgical treatment of chronic type A dissections

**Prognosis**

- 75% survival for 5 years with medical treatment
- Saccular aneurysm 15%
- AR 10%
1. **Etiology**:
- Congenital anomalies
- Cystic medial necrosis
- Trauma
- Aortitis
- Infection
- Arteriosclerosis

2. **Pathology**:

**Arteriosclerosis**: most common in Abd. aorta but 1/4 developed above diaphragm
- Weaken aortic wall from medial degeneration
- may be sacular or fusiform
- Spontaneous rupture occurs

**Cystic medial necrosis**: mostly involved ascending aorta
- associated with AI from aortic annulus dilatation
- may be termed annuloaortic ectasia
- present in Marfan's syndrome
- necrosis, loss of muscle cells in the elastic laminae and often cystic spaces filled with mucoid material

**Trauma**: from blunt trauma
- usual sites are in proximal descending Aorta, Isthmus, (2/3 of the tears)
- tears can also occur in the ascending aorta, aortic arch, distal descending aorta
- Tears are related to points of attachment of the Aorta, ligamentum arteriosum
- 90% die in 1 hr.
- 90% die in 2 wks.
- 5% develop chronic post traumatic aneurysm

**Aortitis**

**Syphilis**: now rare
- ascending aorta predominates
- periarteritis & mesarteritis of aorta lead to destruction of media

**Giant cell Aortitis**: affect large, medium & small arteries
- associated with dissection

**Takayasu's Arteritis**: 5% developed aortic aneurysm

**Mycotic aneurysm**: bacterial infection
- sepsis, embolization
- significant threat of rupture

most common organisms: staph aureus, E.Coli, strep. viridans, Pseudomonas

**Congenital aneurysms** associated with Cong. defect of aorta as Coarctation of Aorta
3. Clinical presentation
   - asymptomatic, accidental finding
   - symptoms of pressure or obstruction of the adjacent structures
   - pain, dyspnea, wheezing, Cough, hemoptysis, recurrent pneumonia
   - Stridor, phrenic n. stretching
   - Hoarseness
   - large ascending aorta may cause SVC syndrome
   - AR symptoms in asc. aortic aneurysm
   - Death . 44% due to rupture
   .33% due to cardiovascular dis.

Diagnosis
   - CXR
   - Aortogram
   - CT scan
   - Transesophageal Echocardiogram

Surgical Treatment
   - Surgery indicated in aneurysm size 5-6 cm.
   - Graft replacement is the standard treatment
   - Ascending aorta and aortic arch always require cardiopulmonary bypass with or without profound systemic hypothermia and circulatory arrest
   - ascending aortic replacement
   - aortic root replacement
   - aortic arch replacement
   - For descending thoracic aortic aneurysm simple resection and graft replacement
Fig 1. Surgical treatment of ascending aortic aneurysm using composite graft
Fig 2. Aortic root replacement (cont.)
Fig 3. Reimplantation of coronary arteries
Fig 4. Distal anastomosis
Fig 5. Completion of anastomosis

Fig 6. Survival of 127 patients with aortic root replacement
Fig 7. Ascending aorta and aortic arch replacement
Fig 8. Aortic arch and aortic root replacement
Most feared complication of descending aortic aneurysm resection is paraplegia from spinal cord ischemia which accounts for ~5-10% of cases. Several techniques have been proposed to prevent the spinal cord ischemia:

- Simple cross clamp
- Gott's shunt
- Left atrial femoral bypass using centrifugal pump
- Ascending Aorta to Descending aorta or femoral a. bypass
- Femoro-femoral bypass
- CSF drainage

So far none has been shown to be superior to the others. Somatosensory evoked potential response monitoring has been used to monitor the spinal cord ischemia during the procedure and for post op follow up.

Fig 9. Techniques of clinical SEP monitoring

Fig 10. Type 1 SEP response: loss of SEP secondary to spinal cord ischemia after aortic cross-clamping without distal perfusion