Vascular Rings and Slings

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Vascular Rings

- Double aortic arch
- Right arch with aberrant LSCA
- Pulmonary sling
- Anomalous innominate artery
Evaluation of Stridor/Dysphagia

- PA and lateral chest and high kV airway films – determine side of arch
- Esophagram with airway fluoroscopy
- MRI/MRA or CTA if vascular ring suspected
Clinical Presentation

• Respiratory symptoms – tight complete rings
  • Stridor
  • Recurrent respiratory difficulties
  • Apnea
• Feeding difficulties
  • Choking with feeds
  • Failure to thrive
  • Solid food dysphagia
Embryology of the Aortic Arch
Aortic Arch Vessel Development

Human Embryo Week 5

Human Embryo Week 8
Edwards’ Theoretical Double Arch
Edwards’ Theoretical Double Arch
Normal Left Arch
Normal Left Arch

- 4th arch
- 8th DA
- 4th arch
- P
- A
Normal aorta and branches
Aberrant RSCA

- Most common – incidence 0.5%
- Asymptomatic in children – normal variant
- 4 branches of aortic arch
- RSCA originates distal to LSCA
- Posterior impression on the esophagus
- Left arch
Normal left arch

Aberrant right subclavian artery
Left Arch Aberrant RSCA

A3 - Development of variant left aortic arch with aberrant right subclavian artery

A4 - Left Aortic Arch with aberrant right subclavian artery

Abnormal obliteration of 4th right arch
Theoretical Aberrant RSCA
Oblique posterior impression on the esophagus
Abnormal contour of aortic knob
Determining Side of Arch

- Tracheal deviation, buckling or impression
- Descending aorta can be on right or left with right arch
Normal tracheal buckling
Right-sided impression on trachea due to right arch
Right Arch

- Branching patterns
  - Mirror image
  - Aberrant LSCA
  - Isolated LSCA – congenital subclavian steal
- High association with congenital heart disease
Mirror Image Right Arch

- Association with CHD 98%
- 25% of patients with Tetralogy of Fallot have mirror image right arch
- 35% of patients with truncus arteriosus have mirror image right arch
- 90% of patients with mirror image right arch have tet
Right arch
Mirror image branching

Normal left arch
Right arch with mirror image branching and left ductus
Right Arch with Mirror Image Branching
Truncus arteriosus
Aberrant LSCA

- Mirror image of aberrant RSCA – right arch
- 12-25% of symptomatic rings
- Left ductus arteriosus completes the ring
- Symptomatic patients typically have a tight ductus or large diverticulum of Kommerell (dilation of origin of aberrant artery)
Aberrant LSCA

- Association with congenital heart disease 5-12%
- Dysphagia lusoria
- Posterior impression on the esophagus
- Right-sided impression on the trachea
- Ddx: double arch
Aberrant left subclavian artery

Aberrant right subclavian artery
Aberrant LSCA
A3 - Development of variant left aortic arch with aberrant right subclavian artery
Right arch with aberrant LSCA and left ductus
Right arch aberrant LSCA with posterior impression on esophagus
Large oblique posterior impression
Diverticulum of Kommerell
Cervical Aortic Arch

- Arch above the clavicle
- May have pulsatile mass in supraclavicular fossa
- 80% are right arches
- Half are symptomatic rings
- Most common variant - right arch that descends on the right then crosses to left behind esophagus and gives off aberrant left subclavian artery and left ductus
Circumflex Aorta

- Rare cause of tracheal compression
- Right arch with left descending aorta
- Retroesophageal portion of the aorta
Double Aortic Arch

- Persistence of both left and right fourth arches which arise from single ascending Ao and join to form single descending Ao
- Most common symptomatic vascular ring
- Right is usually larger, higher and posterior
- Bilateral lateral impressions on the trachea
- Posterior and bilateral lateral impressions on esophagus
- Treatment is to ligate and divide the nondominant arch
Bilateral aortic impression
3-sided impression on esophagus
Double arch atretic left
Double arch ant to post
Double arch ant to post
Double arch ant to post
Pulmonary Sling

- AKA anomalous pulmonary artery
- Left PA originates from right
- Anterior impression on esophagus at level of carina
- Posterior impression on trachea
- Ductus passes from origin of RPA to aorta forming a complete ring around the trachea only
Pulmonary Sling
Pulmonary Sling

- Compression of bronchus intermedius by anomalous artery
- Associated tracheal abnormalities
  - Tracheomalacia
  - Complete tracheal rings – long segment stenosis
  - T-shaped trachea
- Other associated anomalies – abnormal pulmonary lobation, bronchus suis, CHD
Abnormal aeration RML/RLL
Complete tracheal rings

Courtesy Geoffrey Agrons, MD
Philadelphia, PA
Innominate Artery Compression Syndrome

- Normally the innominate artery passes in front of the trachea just below the thoracic inlet
- In infants it arises more to the left than in adults and there is also thymus in this region, so it may cause symptomatic compression of the trachea
- Increased incidence of symptomatic compression in patients with dilated esophagus
- Compression decreases with advancing age
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