Vascular Rings

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Outline

- Case report #1
- Anatomy
- Embryology
- Case report #2
- Epidemiology/presentation
- Work-up
- Operative Correction
- Pulmonary artery slings
Case report

- 2 year old girl
- Eating sausage
- Abrupt onset of drooling, vomiting and dysphagia
- Coughing, choking, wheezing
PMH

- Frequent respiratory tract infections since 1 year of age
- No stridor, no respiratory distress
- With introduction of solid food had increasing swallowing difficulties
- Failure to thrive
PMH

- 2 episodes pneumonia (middle lobe) treated with antibiotics, bronchodilators and physiotherapy
Physical Exam

- Stable respiratory status
- Small for age
- Drooling
- Gagging
Barium swallow

- Diagnosis?
- Foreign body of esophagus
- Plan?
Esophagoscopy

- Impacted sausage with complete occlusion of the esophageal lumen
- Foreign body removed in pieces
- No signs of mucosal lesion, granulation tissue or stenosis at the site of the impaction
Now what?
Repeat swallow

- Deep persistent impression on the posterior aspect of the esophagus
- now what?

CT scan
Anterior-lateral compression from the right side in the distal part of the trachea
Operation

- Ligamentum arteriosum divided, released encircled esophagus and trachea
Post-op

- Recovery uneventful
- No further dysphagia
- Decreased respiratory tract infections
- Repeat esophagram showed release of previously shown posterior dentation
Normal Anatomy

- Aortic arch left-sided
- Aorta descends on left
- Aortic branching
  - Innominate
  - Left carotid
  - Left subclavian
- Left-sided ductal ligament
  - From left PA to descending aorta

http://www.umm.edu/imagepages/19880.htm
Vascular Ring

- Abnormal combination of derivatives of the aortic arch system
- Results in complete encirclement of the trachea and esophagus
- May cause mass effect and obstruction
Double arch

http://www.umm.edu/imagepages/19880.htm
Right arch

- Right arch with mirror-image branching, left ductus from aortic diverticulum to pulmonary artery

Left arch

- Left arch with circumflex aorta and right ductus forms a ring

Hierarchy

- double aortic arch
  - Right arch dominant
  - Left arch dominant
  - Balanced
- Right aortic arch-left ligamentum
  - Mirror-image branching
  - Retroesophageal left subclavian artery
  - Circumflex aorta
- Innominate artery compression
- Pulmonary artery sling
Embryology

- 4th week of embryogenesis branchial arches form

- Arterial supply comes from the heart

- 6 paired arteries arise from the aortic sac and terminate in the ipsilateral dorsal aorta, forming embryonic aortic arches

Embryology

- Third pair of arches form the common carotid arteries
- LEFT fourth arch forms a portion of the adult aortic arch
- RIGHT fourth arch forms the right subclavian artery

http://php.med.unsw.edu.au/embryology/images/a/ae/Stage14_sem2l.jpg
Fifth arch degenerates
LEFT proximal sixth becomes left pulmonary artery
LEFT distal sixth becomes the ductus arteriosus
RIGHT proximal sixth becomes right pulmonary artery
RIGHT distal sixth involutes

Edward’s double arch

First surgical division of vascular ring by Dr. R Gross June 9, 1945 at Boston Children’s hospital

Dr. Neuhauser described radiographic diagnosis in 1946 by using barium esophagram
1931- autopsy on 5 month old child who had wheezing respirations since birth who had a ring of vessels compressing both the trachea and esophagus

Anomaly had been seen in some adult dissections and thought to have been asymptomatic
1944

- 4 month old child admitted with recurrent respiratory tract infections and respiratory distress
- CXR showed a widened mediastinum
  - Dx: enlarged thymus  Rx: radiation therapy
Barium swallow

- AP and lateral films of the esophagus
- Posterior indentation of esophagus

Tracheal imaging

- Lipiodol
  - Iodinated poppyseed oil
- Not standard work-up in 2013
- Showed bilateral compression of trachea

1944 postoperative care

- Steam room for 1 week
- Sulfadiazine and penicillin for 16 days
"with the ear held close to the chest, a faint inspiratory sound could be heard"
- Sounds increased during and immediately following deglutition
- Esophagram did not change
- Trachea had no compression on left; some remained on right
- Child contented and playful
- Free of respiratory embarrassment
Epidemiology

- Rare
- Estimates of 0.1-3%
- Slight male predominance in symptomatic rings 1.3-1.8:1
- Literature is descriptive, not experimental
Clinical presentation

- Exceedingly variable
- Severe presents shortly after birth
- Childhood wheezing, intermittent stridor, recurrent pneumonia, feeding difficulties, dysphagia, failure to thrive
- Adulthood - pregnancy
- Incidentally noted on imaging or at autopsy
TABLE 3. Symptoms leading to clinical presentation in patients with vascular rings*

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Double aortic arch (n = 80)†</th>
<th>Right aortic arch (n = 78)†</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stridor</td>
<td>46 (57%)</td>
<td>18 (23%)</td>
</tr>
<tr>
<td>Recurrent upper respiratory tract infections</td>
<td>22 (27%)</td>
<td>18 (23%)</td>
</tr>
<tr>
<td>Cough</td>
<td>17 (21%)</td>
<td>8 (10%)</td>
</tr>
<tr>
<td>Dysphagia</td>
<td>12 (15%)</td>
<td>12 (15%)</td>
</tr>
<tr>
<td>Respiratory distress</td>
<td>8 (10%)</td>
<td>13 (17%)</td>
</tr>
<tr>
<td>Ventilator preoperatively</td>
<td>7 (9%)</td>
<td>3 (4%)</td>
</tr>
</tbody>
</table>

*More than one symptom occurred in many patients. †Our records did not provide symptoms for the earlier patients in the series.
Chicago series

- 204 patients with vascular rings
- Two patients presented with a coin in their thoracic esophagus
- 3 others diagnosed at time of removal of foreign bodies from esophagus
- Physical exam findings variable
  - Rhonchi, stridor, retractions, wheezing, tachypnea, rales, opisthotonotic posturing with hyperextension of head
Many patients in the Chicago series were initially diagnosed by otolaryngologists.

Most postoperative complications were due to airway issues.

Bronchoscopy important preop to assess for possible complications.
Radiographic work-up

- Barium esophagram supplemented by angiography was former standard.
- X-ray
- CT angiography
- Ultrasound (prenatal)
Chest X-rays

- Pickhardt et al 1997
- 41 children with symptomatic vascular rings
- Retrospectively assessed pre-op chest x-rays
- No symptomatic patient had a normal CXR
- 92% showed anterior tracheal bowing
- 77% showed anterior tracheal narrowing
Esophagram

- Shows posterior indentation
- Often valuable for your work-up
- Normal study excludes vascular ring
Posterior indentation of the esophagus
Esophagram

- Bilateral impressions
- Suggestive of bilateral aortic arches

Ultrasound

- Can define structure and vascular flow
- Very useful for prenatal diagnosis
CT or MRI

- Cross-sectional imaging imperative prior to OR
- Define precise anatomy
- Thoracotomy performed on side of nondominant arch
CT vs. MRI

- **CT**
  - Rapid
  - Requires contrast
  - Radiation exposure

- **MRI**
  - Requires sedation - can be a problem with airway obstruction
CT scans

- CT angio can define anatomy very well
- Various cuts and reconstructions
- A: axial C: coronal G: oblique showing complete ring

Treatment

- Only symptomatic rings require treatment
- Effect on the trachea and esophagus paramount
Pre-op work-up

- **Echocardiogram**
  - 12-30% have associated congenital heart disease

- **Bronchoscopy**
  - 3% have associated airway anomalies, e.g., tracheomalacia, bronchomalacia, complete tracheal rings
  - Blind intubation not recommended
Operative correction

- muscle-sparing thoracotomy
- For double arch - divide lesser of 2 arches
  - Clamp first and monitor pulses prior to division
  - If equal arches, clamp and measure blood pressure in lower legs
Operative correction

- For right arch and left ligamentum
  - Divide ligamentum
- Recurrent laryngeal nerve identified and preserved
- Phrenic nerve identified and preserved
- Divide any and all fibrous bands adjacent to the esophagus and trachea
- Rarely need a chest tube
- Mean hospital stay 3 days
Persistent Respiratory distress

- 64-81% of patients have complete resolution of symptoms
- Often due to tracheomalacia
- Residual compression by the aorta

Aortopexy

- Displacement of vessels away from trachea
- Sutured at the posterior surface of sternum
Pulmonary sling

- Left pulmonary artery comes off the right pulmonary artery
- Usually travels between trachea and esophagus

Trachea malformations

- 50-65% of patients have complete tracheal rings
- Length of stenosis frequently extensive
- External and internal narrowing

Clinical presentation

- Stridor, wheezing, respiratory distress
- Often intubated
- More severe associated anomalies
  - Frequent VATER syndrome
  - ASD, VSD
  - Agenesis of right lung
Work-up

- Echocardiogram
- Bronchoscopy
  - Essential, define length and extent of tracheal stenosis
  - Operative planning for tracheal reconstruction
  - Extent of stenosis not always apparent when viewing trachea externally
Operative technique

- Median sternotomy
- Resect thymus
- Harvest pericardium
- Place on cardiac bypass or ECMO
- Resect left pulmonary artery and sew it to the main pulmonary artery
Repair

Trachoplasty

- Often required
- Pericardial patch tracheoplasty
- Slide tracheoplasty
Pericardial patch tracheoplasty

- Pericardium harvested
- Trachea incised
- Sewn in place

Slide tracheoplasty

- Divide stenosis transversely in the midportion
- Incise the upper segment vertically posteriorly
- Incise the lower segment vertically anteriorly
- Two ends placed over each other
Advantages

- Less tension
- Decreased granulation tissue
- Avoidance of graft materials
- Decreased tracheal mobilization
- Good success rate
Fiore et al.

- 14 patients with pulmonary artery sling from 1983-2003
- 6 days to 27 months
- 8 with long segment congenital tracheal stenosis associated with it and underwent pulmonary artery sling and tracheal surgery
- 6 had mild tracheal narrowing and had repair of pulmonary artery sling only
Postoperative morbidity

- 14 patients- 12 hospital survivors
- Plication of the left hemidiaphragm in 2 patients
- Pacemaker in 1 patient
- Lobar emphysema requiring resection of right upper and middle lobes in 1 patient
- Mediastinal bleed requiring reexploration
- Superficial wound infection
- Tracheostomy after multiple resections of granulation tissue at the carina on vent
- 1 late death from fungal sepsis

Yong et al


- Slide tracheoplasty
- Improved critical care
References