Vascular Anomalies of the Airway

The Trachea
Cartilaginous to Membranous Ratio

- Normal : 4.5:1
- Tracheomalacia: 3:1, 2:1, 1:1
  - Primary (Intrinsic)
    - Often with TEF
  - Secondary (Extrinsic)
    - Vascular Compression

Secondary Tracheomalacia – Extrinsic Compression
Anomalies of the Aortic Arch

- 1. Anomalous innominate artery
- 2. Vascular sling
- 3. Retroesophageal subclavian artery
- 4. Double aortic arch

Symptoms of Tracheomalacia

- Stridor
  - Expiratory > inspiratory
- Chronic cough, barking
- Recurrent pneumonia or bronchitis
- Apnea, “dying spells”
- Dysphagia
- Exercise intolerance
Tracheomalacia from Aortic Arch Anomalies

**Diagnosis**
- Bronchoscopy
  - Defines severity, location and extent of dynamic obstruction
  - Spontaneous ventilation
- Magnetic Resonance Angiography
- Computed Tomography with Angiography
- Barium Swallow

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**Management**

**Medical Management: The Sick Plan**
- Chest physiotherapy
- Nebulized saline treatments
- Inhaled steroids
- Antibiotics as indicated

**Surgical Management**
- Surgical correction – cause specific
- Tracheotomy

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1. “Anomalous” Innominate Artery Compression

Most common vascular anomaly
Innominate arises more distally along arch

Age of diagnosis - Bimodal
- Infancy or early childhood – most cases
- Adolescence
  - Chronic cough
  - Exercise intolerance

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**Bronchoscopy**

**Anomalous Innominate Artery**

Mild
Severe

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**Innominate Artery Compression**

Surgical Management: Aortopexy

**Absolute indications**
- Reflex apnea
- Recurrent episodes of respiratory distress

**Relative indications**
- Recurrent pneumonia
- Exercise intolerance

Intra operative bronchoscopy very helpful in determining vectors to maximize expansion!!
Innominate Artery Compression
Surgical Management

Pre Aortopexy
Apnea

Post Aortopexy
No apnea

2. Vascular Sling
• Left pulmonary artery arises from right pulmonary artery
• Vessel courses posterior to trachea, anterior to esophagus to approach left lung
• Symptoms arise from tracheal compression


2. Pulmonary Sling
Complete tracheal rings
10% of cases

Vascular compression BETWEEN trachea and esophagus

3. Right Retroesophageal Subclavian Artery
1% of population
“Dysphagia Lusorum”
Right non-recurrent laryngeal nerve

Grant’s atlas

3. Retroesophageal subclavian artery

Bronchoscopy: posterior pulsatile mass
Esophagoscopy: posterior pulsatile mass
3. Right Retroesophageal Subclavian Artery

- Air-filled Esophagus

4. Vascular Ring

- Double Aortic Arch
- Bronchoscopy
- Failure of right dorsal arch involution
- Arches encircle esophagus and trachea
- Ring may be vascular or ligamentous

- Usually symptomatic during infancy or early childhood
- May sometimes be relatively asymptomatic
- 80 year old male who failed extubation post bypass surgery
- Relatively asymptomatic but "had trouble keeping up with the grandkids"

Summary

Professor Bruce Benjamin
Vascular Tumors and Malformations
Larynx

- Subglottic hemangioma
- Lymphatic malformation (LM)

Subglottic hemangioma-SGH

- Symptom onset: 6 weeks
- Mean age of diagnosis: 3.6 months
- 50% of patients have cutaneous hemangiomas
- Beard distribution higher likelihood of SGH
- Unilateral, bilateral, circumferential
- MR imaging for extralaryngeal extension
- Watchful waiting unacceptable!

Subglottic hemangioma

**Subglottic hemangioma-Stage**

<table>
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<th>Stage 1</th>
<th>Stage 2</th>
<th>Stage 3</th>
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<td>Percent extralaryngeal hemangiomas</td>
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Table 1: Proposed airway III staging system and treatment protocol

Perkins JA, Chen EY, Hoffer EA, Manning SC. Oto H and N Surg 2009;141:516-21

Subglottic Hemangioma

**Treatment**

- Propranolol
- Corticosteroids
- Intubation—short term
- Laser Excision
- Open resection
- Tracheotomy

Pre propranolol

6 months post propranolol

- Propranolol first line of therapy, efficacy: 75%
- +/- 48 hours corticosteroids and/or intubation
- Surgery for non-responders or those not wanting prolonged use of medications


Unilateral Subglottic Hemangioma

Non-pharmacologic treatment:
- Laser or open surgical excision

Pre-Op  | Post 2nd laser

Unilateral Subglottic Hemangioma

3.5 weeks propranolol

11/27  | 12/20
4 year old female with microcystic LM of supraglottis, Left > Right LMs spare glottis and trachea

Microcystic LM of Supraglottis

Bleomycin Sclerotherapy

- Tracheotomy dependent
- 12 CO2 laser procedures
- Systemic therapy
  - Sildenafil
  - Sirolimus
  - Propranolol

- Post 2 Bleomycin injections
  - Judicious!!!!
  - 0.1-0.2 ml/site
  - 3-4 sites max
  - Lifetime max: 300 units (100mls)

Post supraglottoplasty
Decannulated 3 months later

Bleomycin Sclerotherapy
A word of caution!

Vascular Anomaly Case Report

- 4 month old female with isolated left upper lip hemangioma
- Feeding well
- "Barky cough" noted at 2 months of age, worsening, particularly at night
- Management?

Bleomycin Lymphatic Malformation of Tongue

- Embolization therapy
- CO2 laser therapy
- Coblation
- Bleomycin injection
- Surgical excision

3 weeks post injection

Laryngoscopy
Subtle narrowing and abnormal shape of distal trachea

Posterior pulsatile area limiting view of right superior lobe bronchus

Upper lobe bronchus normal once area passed.

Nancy M. Bauman MD FACS FAAP
Professor
George Washington University
Children’s National Medical Center
Pediatric Otolaryngology

The End
Thank you!