


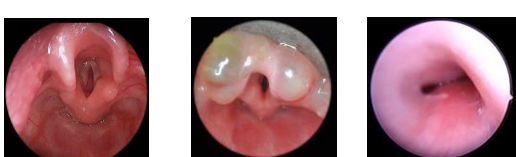
### Vascular Anomalies of the Airway



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 Children's National Medical Center  
 Pediatric Otolaryngology

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### Vascular Anomalies Affecting the Airway



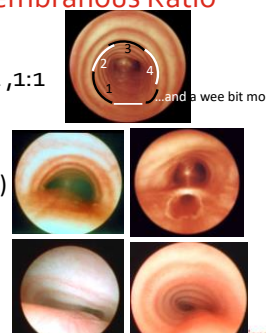
Subglottic Hemangioma    Lymphatic Malformation    Aortic Arch Anomalies

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### 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

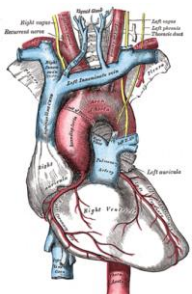


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### Secondary Tracheomalacia – Extrinsic Compression

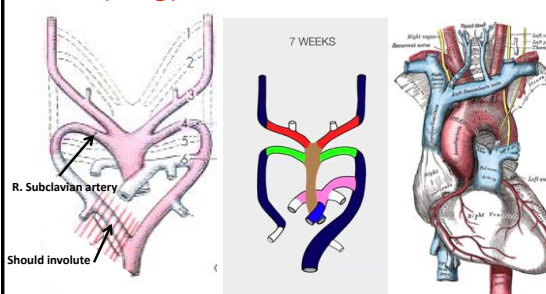
#### Anomalies of the Aortic Arch

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



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### Embryology of the Great Vessels



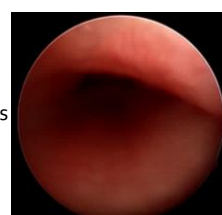
R. Subclavian artery  
Should involute

7 WEEKS

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### Symptoms of Tracheomalacia

- Stridor
  - Expiratory > inspiratory
- Chronic cough, barking
- Recurrent pneumonia or bronchitis
- Apnea, "dying spells"
- Dysphagia
- Exercise intolerance



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## Innominate Artery Compression Surgical Management

Pre Aortopexy  
Apnea

Post Aortopexy  
No apnea

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Contents lists available at ScienceDirect  
Journal of Pediatric Surgery  
journal homepage: www.elsevier.com/locate/jpedsurg

### Posterior tracheopexy for severe tracheomalacia

Hester F. Shieh <sup>a</sup>, C. Jason Smithers <sup>a</sup>, Thomas E. Hamilton <sup>a</sup>, David Zurakowski <sup>a</sup>, Lawrence M. Rhein <sup>b</sup>, Michael A. Manfredi <sup>c</sup>, Christopher W. Baird <sup>d</sup>, Russell W. Jennings <sup>a,d</sup>

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<sup>b</sup> Department of Pulmonology, Boston Children's Hospital, Harvard Medical School, Boston, MA 02115, United States  
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98 patients reported  
- median age 15 months  
- 80% with TEF

Posterior tracheal wall sutured to the anterior vertebral fascia

May be combined with anterior aortopexy or performed in isolation

Symptom resolution reported in 90% of patients

Generally performed for >2/3 intrusion

- Pexy from posterior tracheal membrane (PTM) to anterior longitudinal spinous ligament (ALSL)

June 2017; 52:951-55

## 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

Backer, J Thor Cardiovasc Surg 1992

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## 2. Pulmonary Sling

Complete tracheal rings  
10% of cases

Vascular compression BETWEEN trachea and esophagus

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## 3. Right Retroesophageal Subclavian Artery

Grant's atlas

1% of population  
"Dysphagia Lusorum"  
Right non-recurrent laryngeal nerve

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## 3. Retroesophageal subclavian artery

Bronchoscopy: posterior pulsatile mass  
Esophagoscopy: posterior pulsatile mass

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### 3. Right Retroesophageal Subclavian Artery

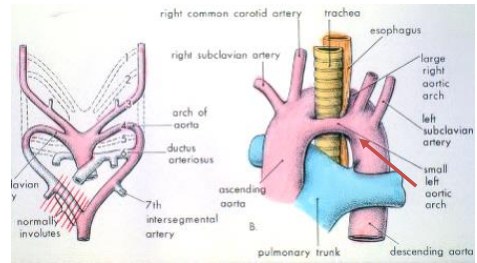


Air-filled Esophagus

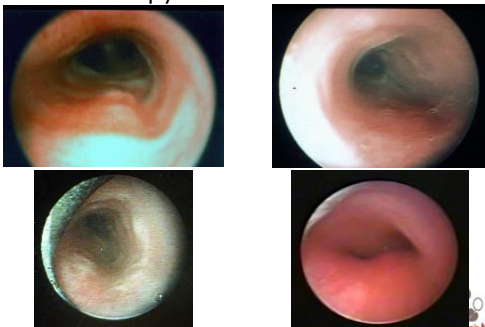
Right Subclavian Artery



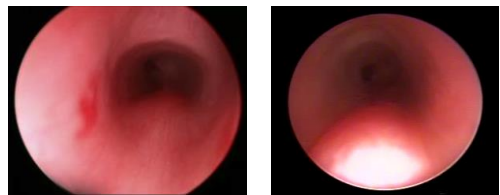
### 4. Vascular Ring Double Aortic Arch



### 4. Vascular Ring Bronchoscopy



### 4. Vascular Ring Bronchoscopy



### 4. Vascular Ring

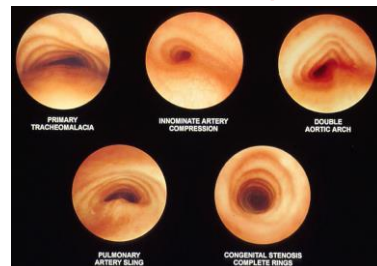
- Usually symptomatic during infancy or early childhood
- May sometimes be relatively asymptomatic

- 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)

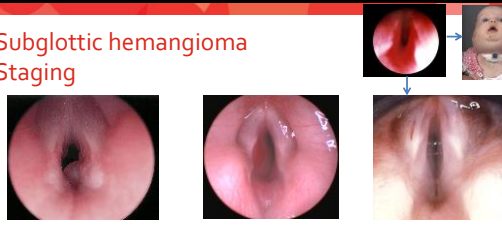


## Subglottic hemangioma-SGH

- 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 Staging



Stage*	Unilateral airway hemangioma	Circumferential or bilateral airway hemangioma	Percent laryngeal airway obstruction	CTA or MRI extralaryngeal hemangioma volume (mm <sup>3</sup> )
1	Yes	No	≤50	<4000
2	Yes	Yes	>50-90	4001-10,000
3	No	Yes	>90	>10,000



CTA, CT angiography; IH, infantile hemangioma.  
\*Stage based on lowest stage in a row with two or more positive findings.

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

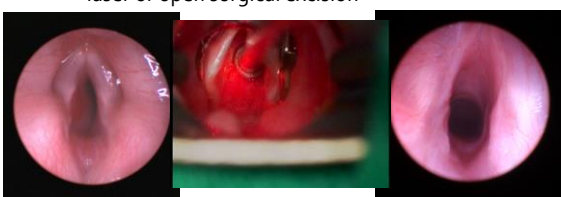
- Propranolol first line of therapy, efficacy: 75%
- +/- 48 hours corticosteroids and/or intubation
- Surgery for non -responders or those not wanting prolonged use of medications

Denoyelle F, Garabedian EN. Propranolol may become first-line treatment in obstructive subglottic infantile hemangiomas. Otolaryngol Head Neck Surg. 2010;142:463-4.  
Elluru, Priess, Richter, Gimmer, Danow, Shih, Perkins. Multicenter Evaluation of the Effectiveness of Systemic Propranolol in the Treatment of Airway Hemangiomas. OHSU, 2015.




## Unilateral Subglottic Hemangioma

Non-pharmacologic treatment:  
laser or open surgical excision

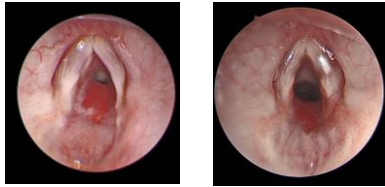


Pre-Op      Post 2nd laser




## Subglottic Hemangioma

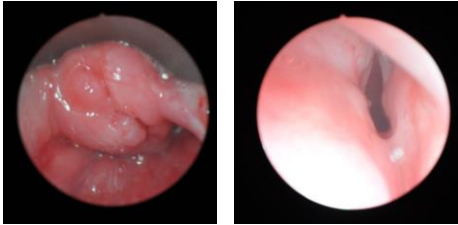
3.5 weeks propranolol




11/27      12/20



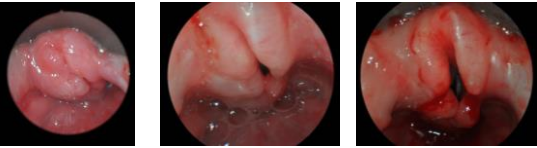
### Microcystic LM of Supraglottis



4 year old female with microcystic LM of supraglottis, Left > Right  
LMs spare glottis and trachea




### 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


- Christina Daniel, BA, Andrea L. Tichy, PhD, Umar Tariq, MBBS et al. An open-label study to evaluate sildenafil for the treatment of lymphatic malformations. J Am Acad Derm 2014;70:1040-7.  
- Ozeki M, Kanda K, Kawamoto N. et al. Propranolol as an alternative treatment option for pediatric lymphatic malformation. Tohoku J Exp Med 2013; Vol. 229:61-6



### Bleomycin Sclerotherapy A word of caution!





### Bleomycin Lymphatic Malformation of Tongue





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

3 weeks post injection

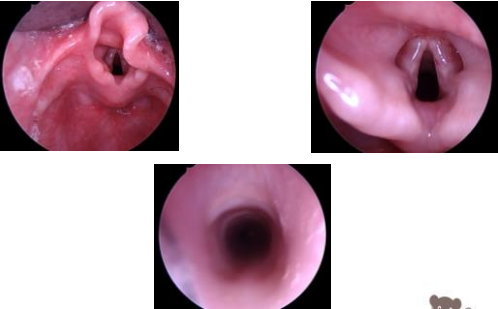



### Vascular Anomaly Case Report

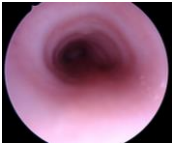
- 4 month old female with isolated left upper lip hemangioma
- Feeding well
- "Barky cough" noted at 2 months of age, worsening, particularly at night
- Management?

### Laryngoscopy

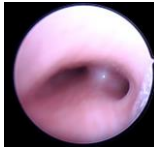
## Bronchoscopy



Subtle narrowing and abnormal shape of distal trachea



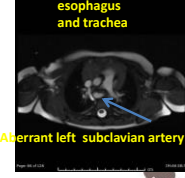
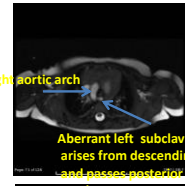
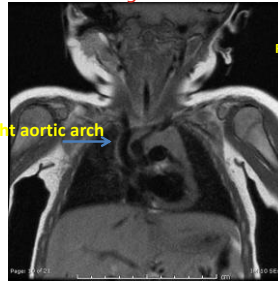
Posterior pulsatile area limiting view of right superior lobe bronchus



Upper lobe bronchus normal once area passed.



## RAA with Ring



The End  
Thank you!



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