It’s a tight spot!!

Imaging of airways and esophagus in aortic arch abnormalities: Beyond airway compression

Robert J. Fleck, Jr., M.D.
Cincinnati Children’s Hospital Medical Center

Introduction

Much of medicine is empiric

Original Report

Robert J. Fleck[1,2]
Preeyacha Pacharn[1,3]
Bradley L. Fricke[1]
Matthew A. Ziegler[1]
Robin T. Cotton[4]
Lane F. Donnelly[1]

Imaging Findings in Pediatric Patients with Persistent Airway Symptoms After Surgery for Double Aortic Arch

AJR;178, May 2002

Objectives

- Discuss the current methods of assessing the aorta, airway and esophagus.
- Propose a greater role for the consideration of tracheomalacia for persistent symptoms
- Present dynamic imaging as a method of problem solving

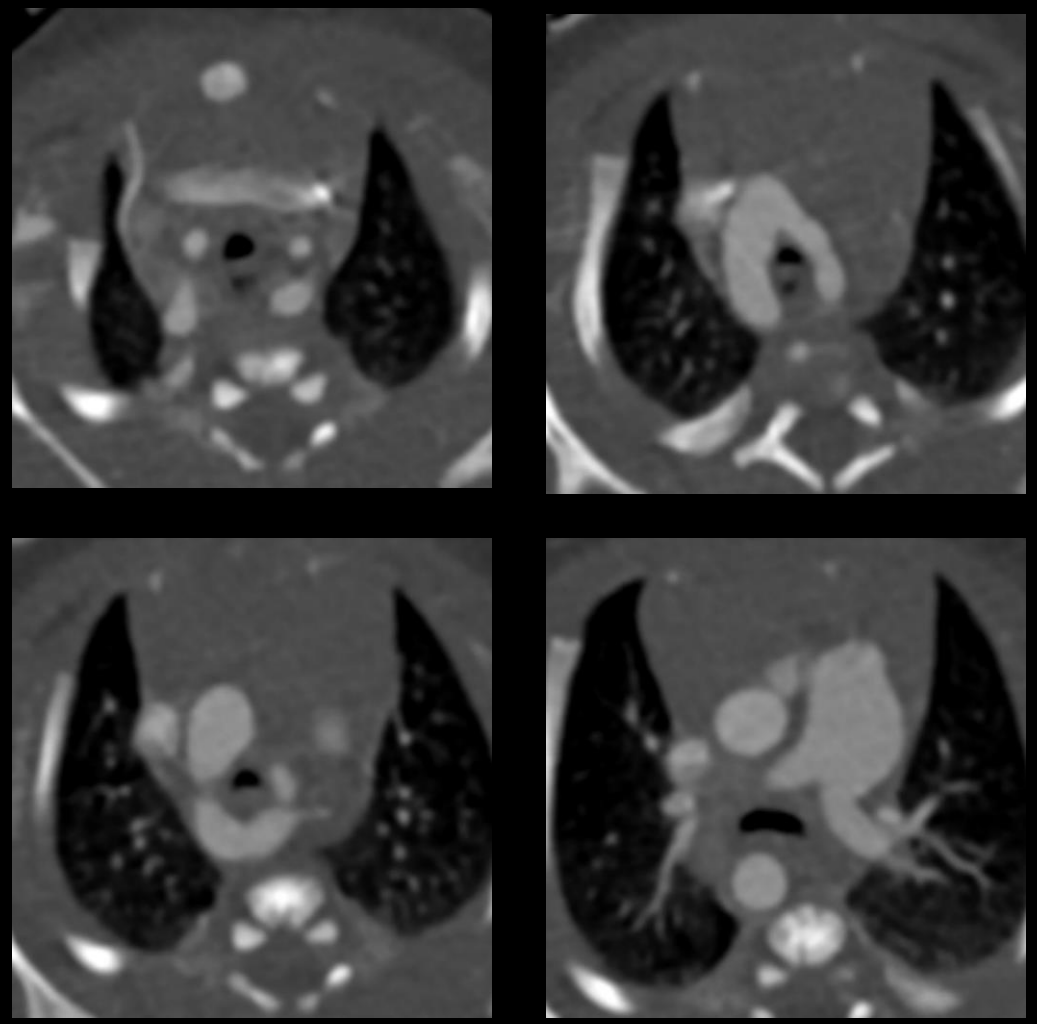
How patients come to imaging…

- Symptomology
- Echocardiography
- Bronchoscopy
- Fluoroscopy

Cincinnati Children's
changing the outcome together

Initial Assessment of Aorta

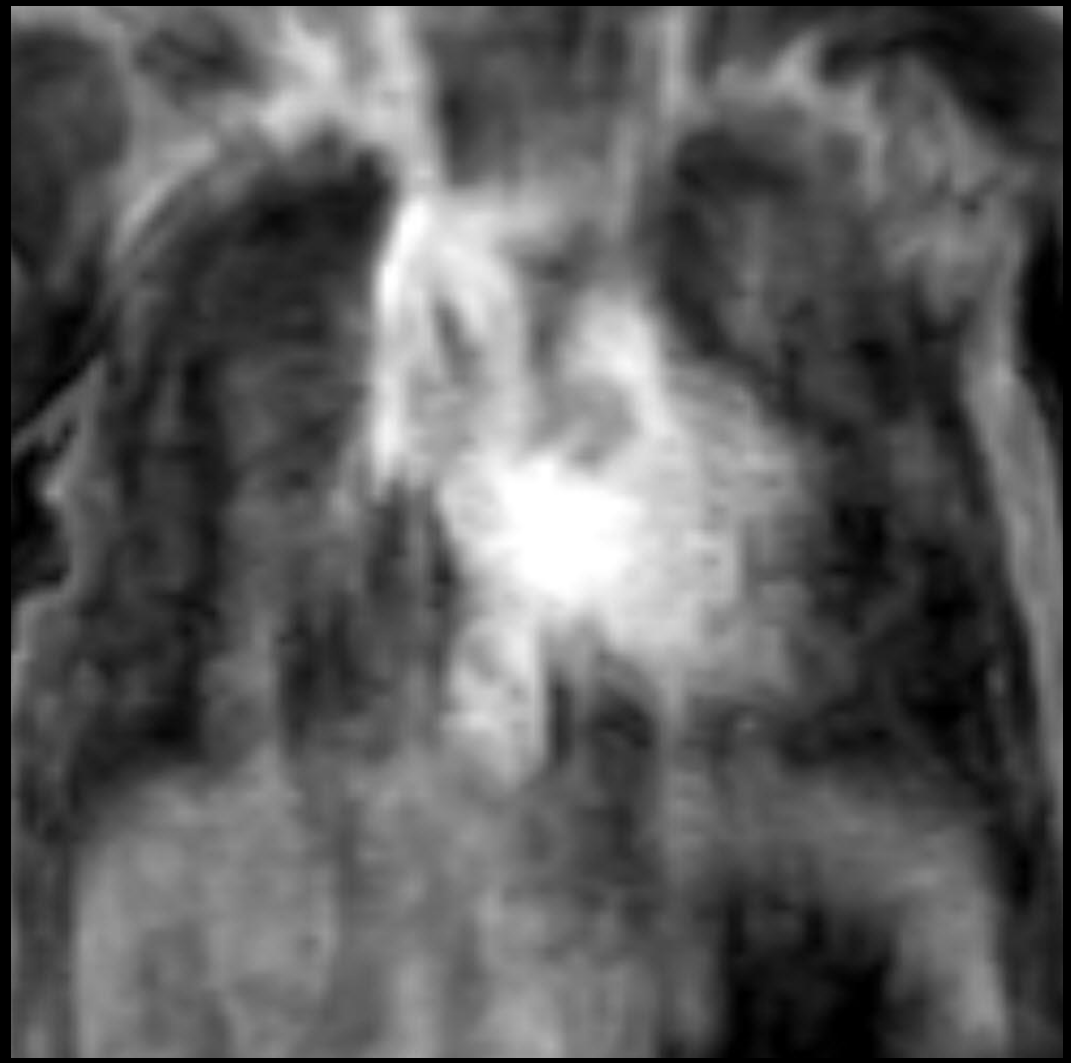
- CT
  - CT angiography
  - Dynamic Airway with or without contrast



4 day old male with in utero meningomyelocele repair and double aortic arch

Initial Assessment of Aorta

- CT
  - CT angiography
  - Dynamic Airway with or without contrast
- MRI
  - Out of favor in US
  - Sedation/General Anesthesia
  - New techniques avoid sedation and are quiet helpful



3 week old infant with cough, shortness of breath

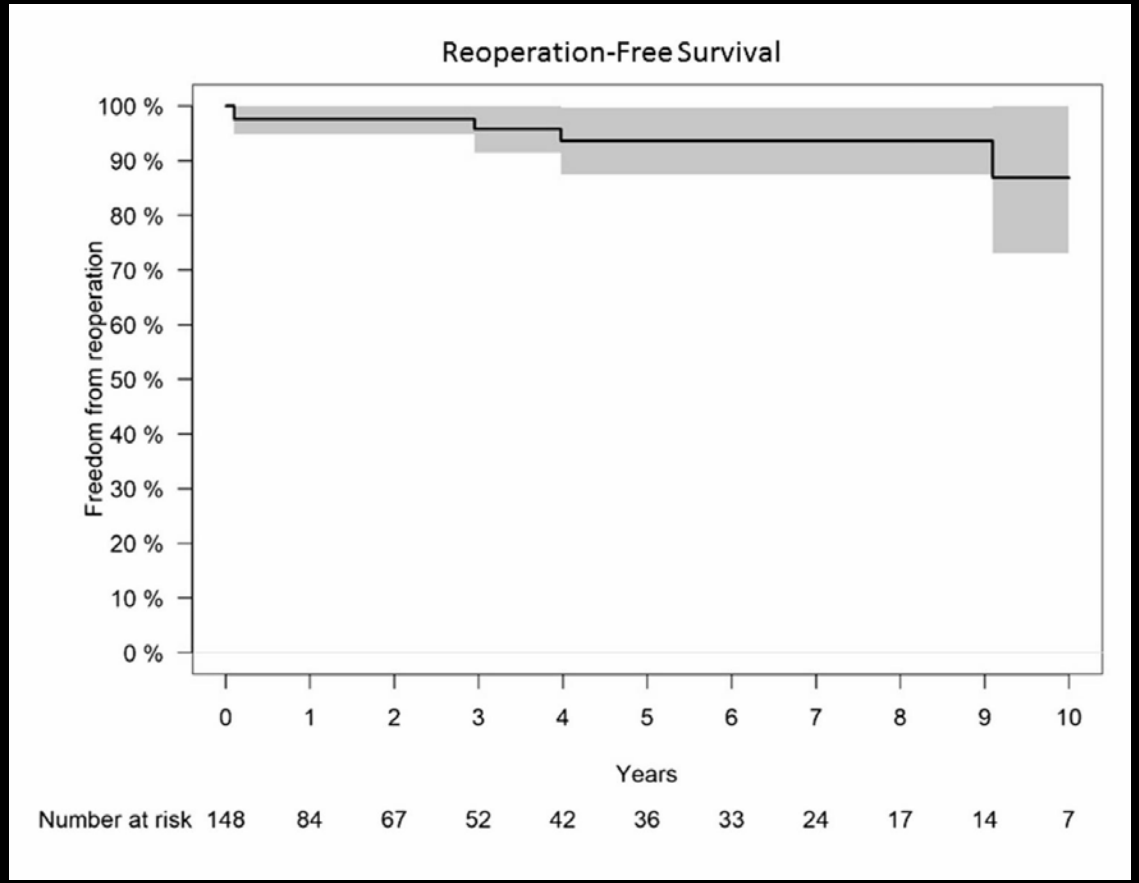
Goals of Initial Assessment

- Define the anatomy of the heart and vascular structures
- Visualize the effect on the airway
- Evaluate the lung
- Other chest anomalies

Newborn with biphasic stridor and respiratory distress

Outcomes

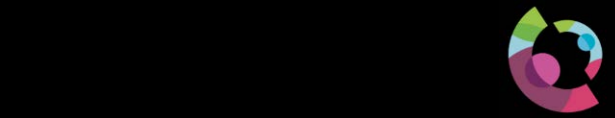
- Reoperation
  - 6 cases
    - 2 with dysphagia
    - 3 Thoracic duct ligations
    - 1 translocation of descending aorta
  - Median follow-up 1.72 years
- 10% had persistent symptoms



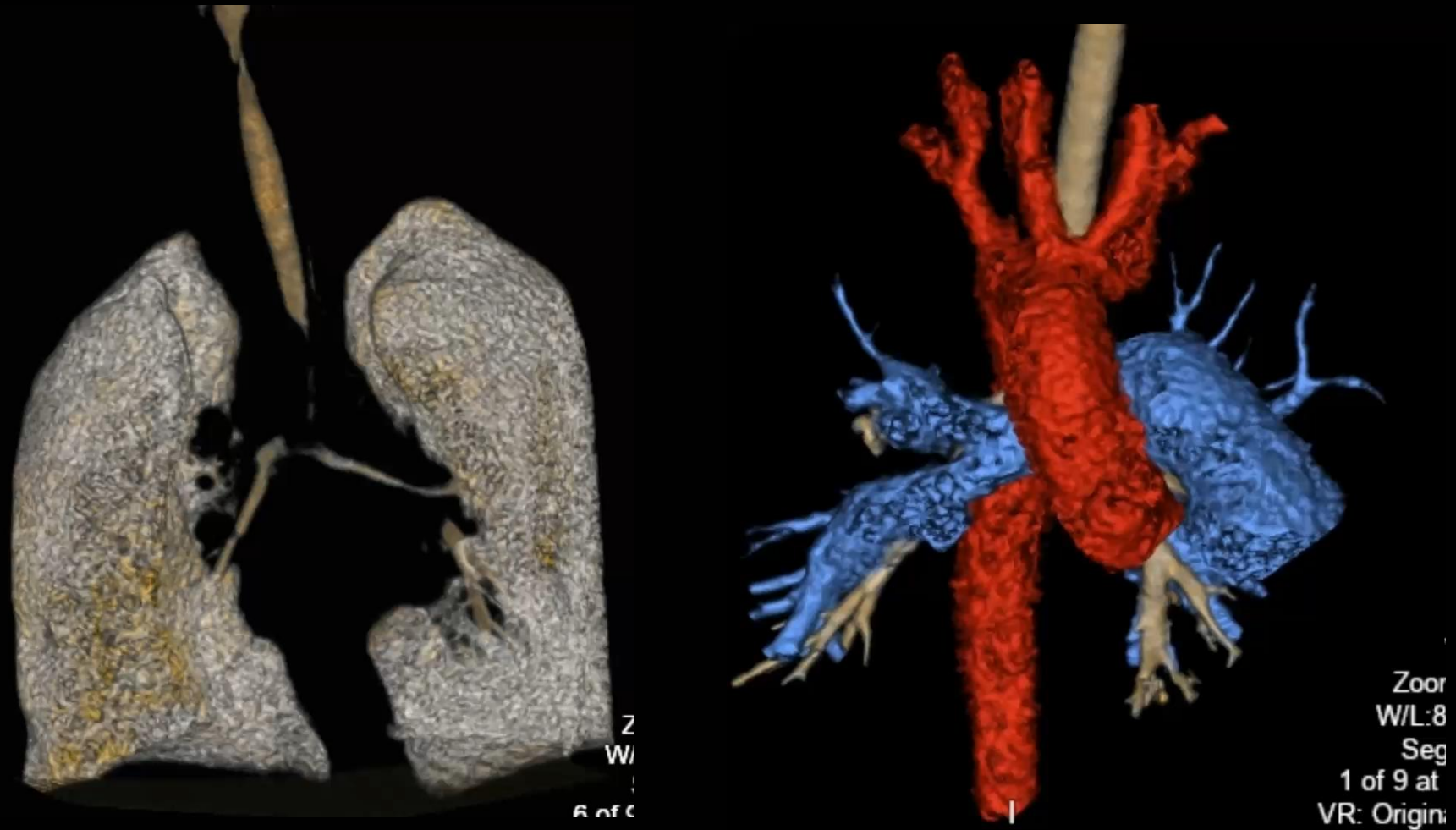
Binsalamah, Ann Thorac Surg 2019

Outcomes – Really, they are very good!

- What are other outcomes to take into account?
  - Hospitalization for respiratory illness
  - Persistent respiratory symptoms
  - Additional bronchoscopies or CT scans
  - Aspiration
- Large majority do very well but a substantial number are seen in follow-up by pulmonary and otolaryngology

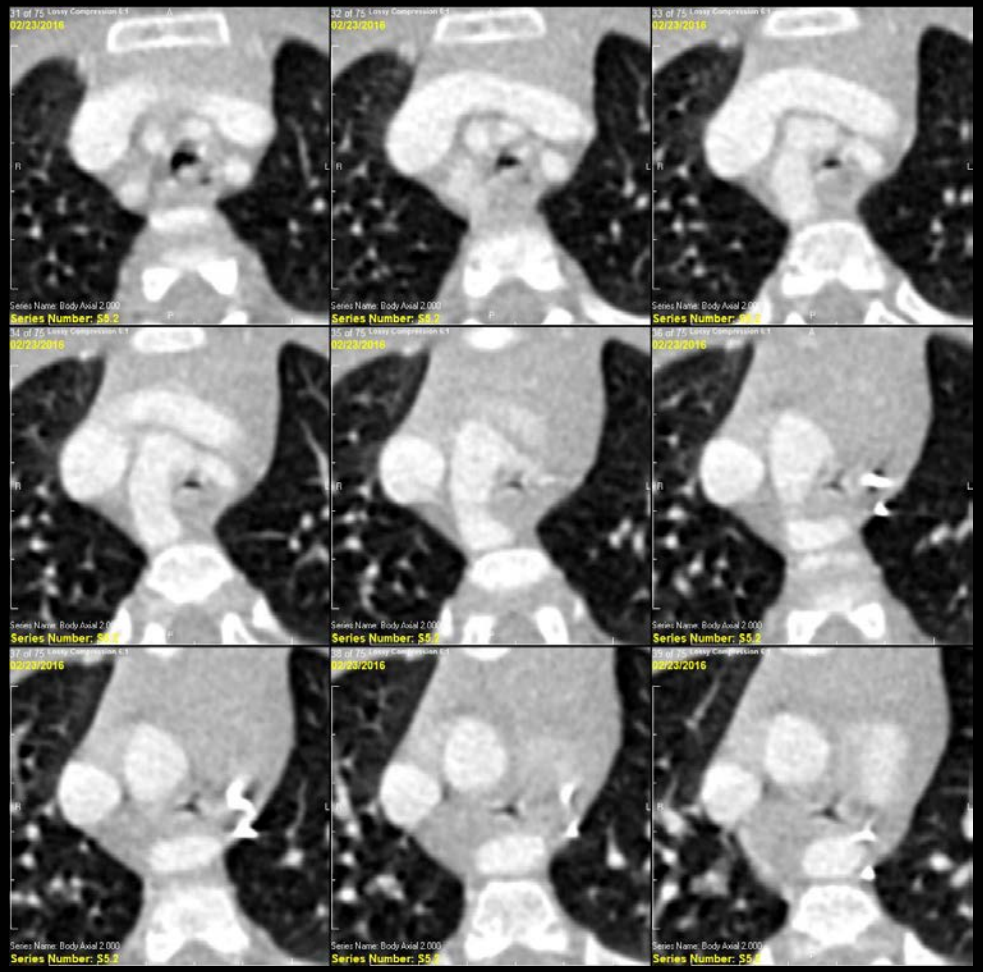


14 month old with history of double aortic arch with repair at 7 months referred for persistent symptoms



apneic spells, cyanotic spells, increased work of breathing with retraction during exertion, 6 hospitalizations for respiratory illness, apnea monitor triggers twice per night

Cincinnati Children's
changing the outcome together



Division of ligamentum arteriosum, lateralization and aortopexy and lysis of adhesions

Cincinnati Children's
changing the outcome together

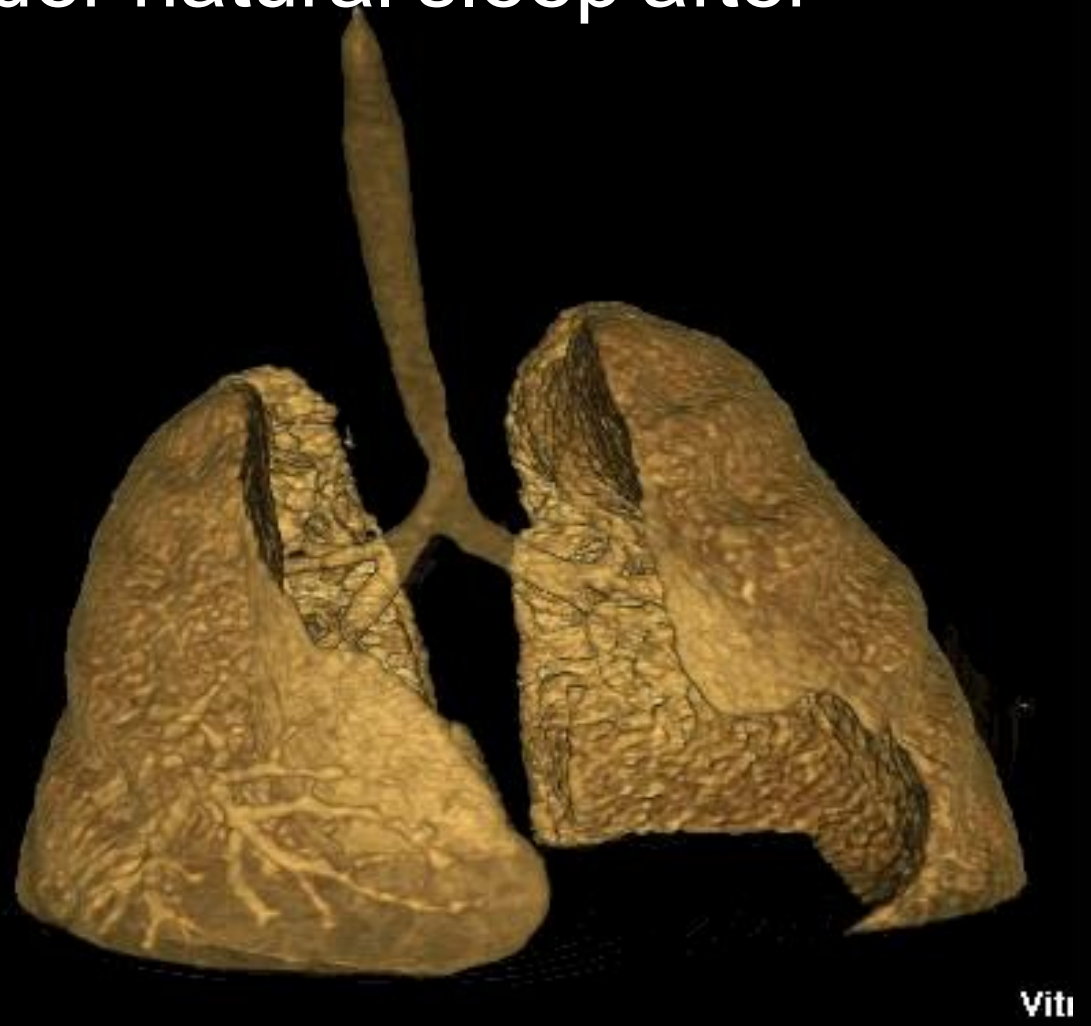
Another example – closer to home

- Division of left arch with a right arch dominant.
- Division and ligation of patent ductus arteriosis
- Persistent symptoms and illnesses
- 6 scopes “mild to moderate tracheomalacia”

Newborn with biphasic stridor and respiratory distress

Dynamic Cine CT performed under natural sleep after feeding, no contrast

- Advantages
  - No sedation/anesthesia
  - Evaluates the trachea in natural breathing
    - If collapsing, likely bad tracheobronchomalacia
    - If flattened - compression and malacia
  - Noninvasive
- Disadvantage
  - Not sensitive for airways that may collapse during coughing, exercise or other dynamic airway movement
    - These airways will still appear “oval” in shape which is very sensitive for trachomalacia



Cincinnati Children's
changing the outcome together

Esophageal Atresia and Tracheoesophageal Fistulae (TEF)

Pathophysiology

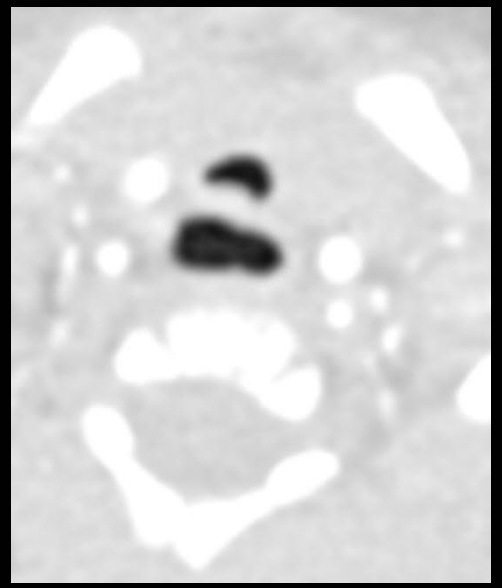
- Embryologically – common lumen that develops a septum
- Failure results in TEF and clefts
- Trachea cartilages and membrane do not develop normally
- Esophageal plexus does not develop normally
- Dilated esophagus compresses

Potential Problems

- Tracheobronchomalacia - 33%
- Esophageal reflux and aspiration
- Recurrent stricture of esophagus
- Compression by normal vascular structures due to soft trachea
- Other associated congenital anomalies

Esophageal Atresia (EA) and Tracheoesophageal Fistulae (TEF)

4 m.o. female with EA/TEF repair with aortopexy presents vomiting and feeding intolerance.



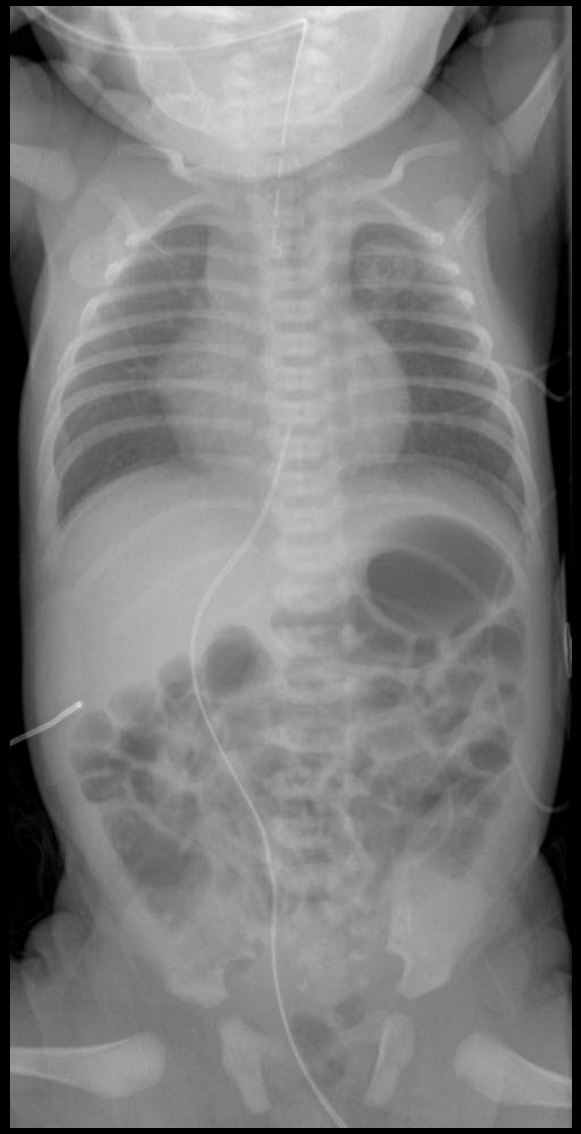
CTA performed without sedation or airway pressure support.

Cincinnati Children's
changing the outcome together

Esophageal Atresia and Tracheoesophageal Fistulae

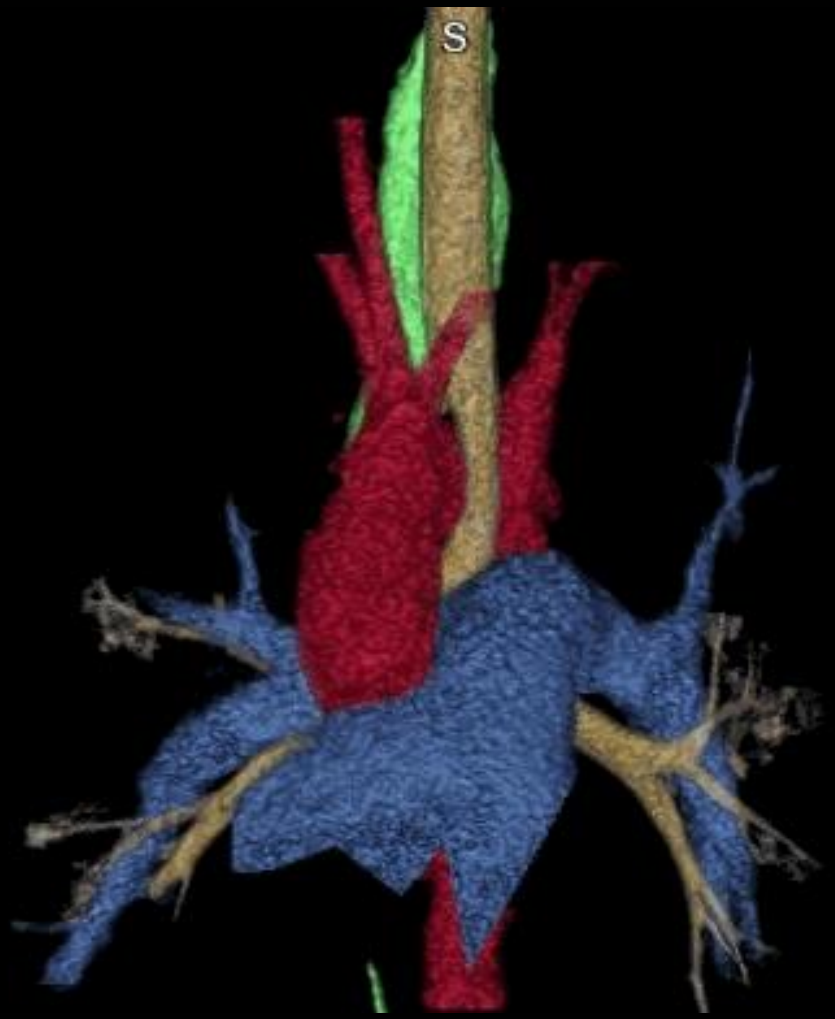
Newborn with EA and TEF

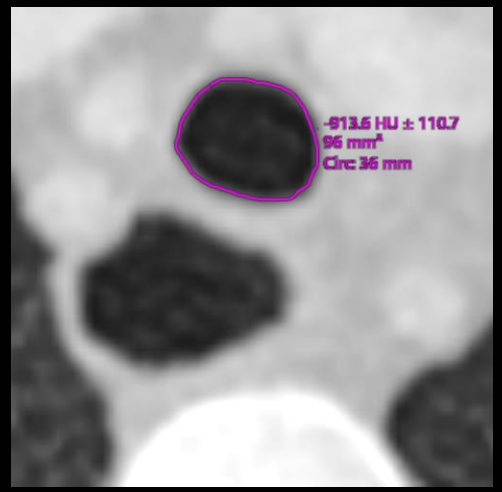
- Right approach discovered right arch, repair performed
- Age 7 – dysphagia
- Age 15 – dysphagia of solids and worsening dyspnea on exertion

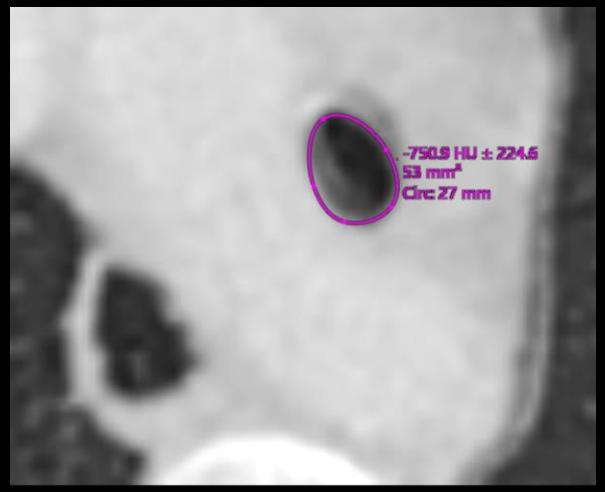


Cincinnati Children's
changing the outcome together

Esophageal Atresia and Tracheoesophageal Fistulae





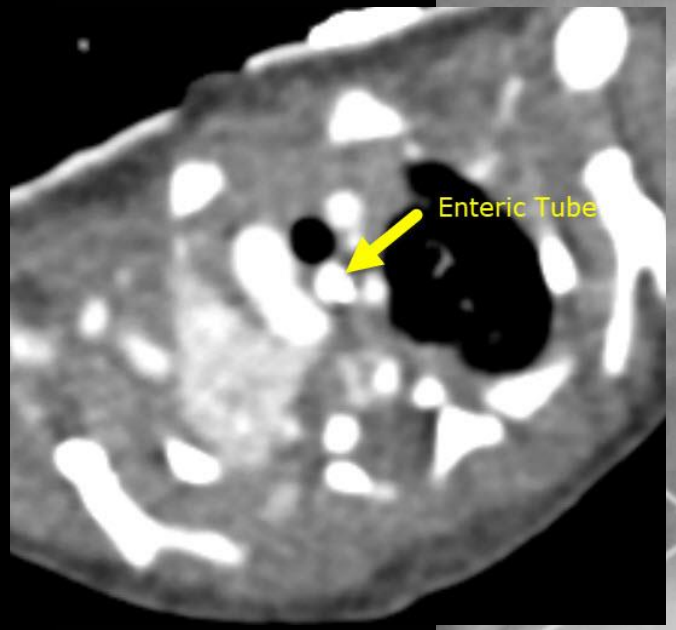


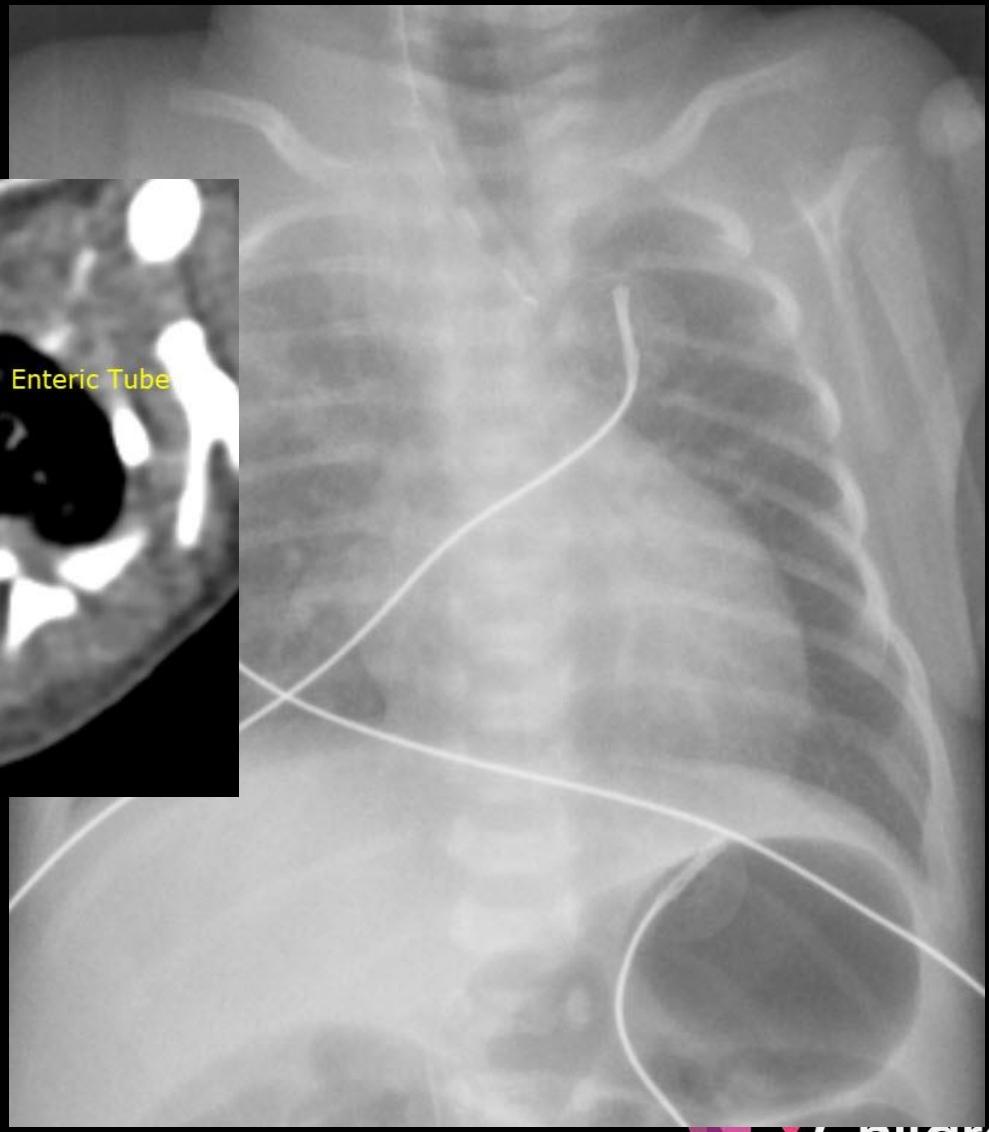
Treatment of GERD and newly diagnosed asthma resulted in marked improvement

15 years-old with chief complaint of dysphagia with solids and sometime large volumes of reflux. Dysmotility on esophagram but no stricture. Dyspnea on exertion increasing over several months with walking tolerance decreasing from one mile to 100 yards.

Cincinnati Children's
changing the outcome together

Esophageal Atresia and Tracheoesophageal Fistulae





Non sedated CTA, tidal breathing, no airway or pressure support: No tracheal narrowing prior to repair

BPD MRI phenotype: fibro-inflammatory, simplified, hyperinflated, possible airway malacia

Moderate/severe

MRI parenchyma score: 11.5/14

12-35% by volume

Fibrosis, atelectasis, inflammation

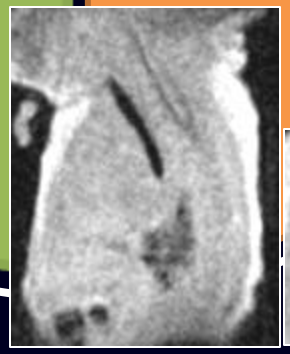
2-10% by volume

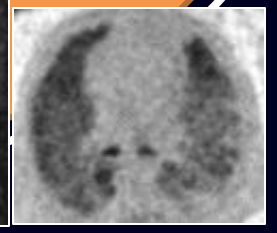
Alveolar simplification, hyperinflation, cysts

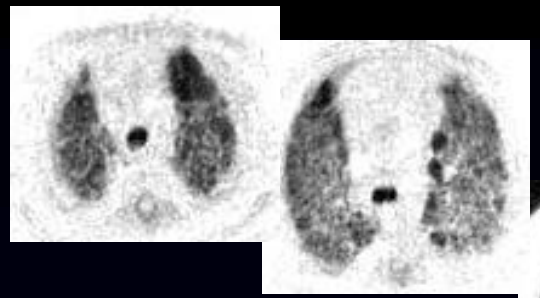
Pulmonary hypertension, vascularity, flow

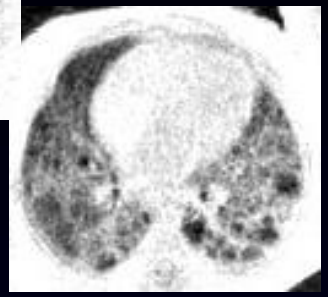
Airway Malacia

intubated at MRI





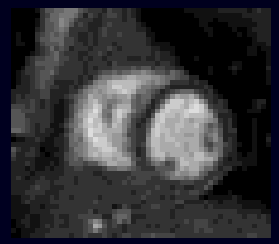




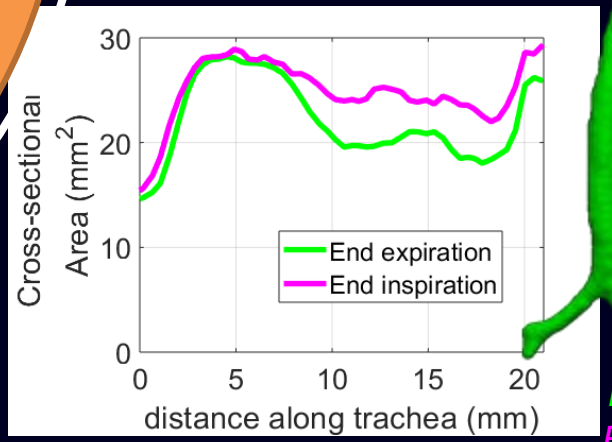
Moderate low-density tissue
Moderate hyperinflation

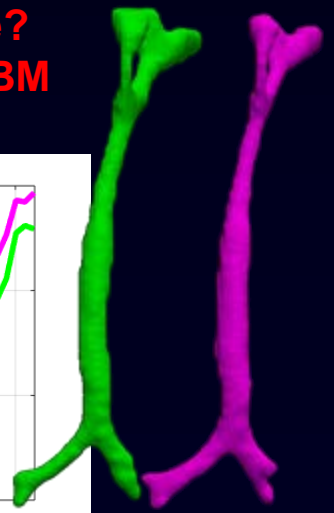
Mild

Short-axis cine



Collapsing to tube?
Potential TM and BM



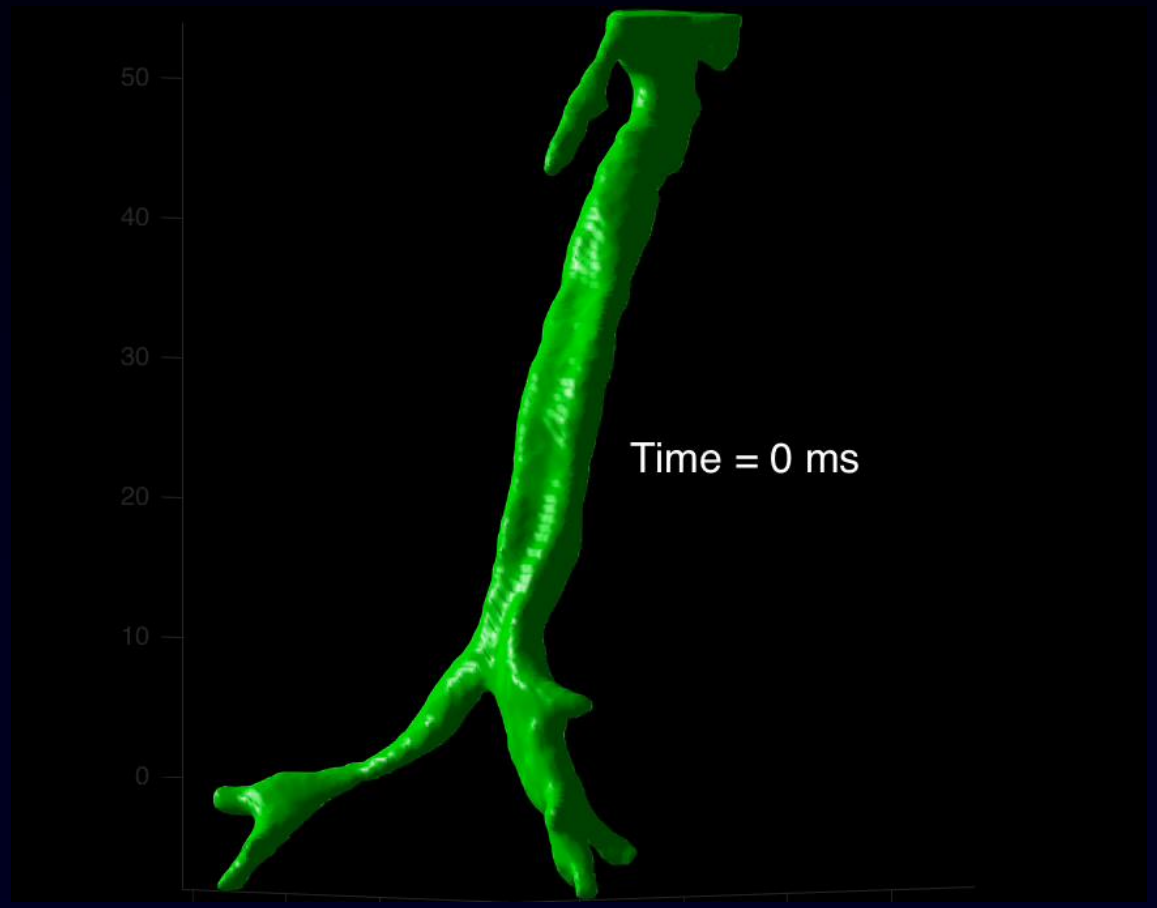


End-expiration
End-inspiration

Flow [/BSA] (L/min [/m^2])	Anatomy ratios
MPA: 0.75 [3.5]	LV EI (d) = 0.91
RPA: 0.46 [2.2]	LV EI (s) = 0.81
LPA: 0.48 [2.3]	MPA/AO = 1.13

Tracheomalacia in Prematurity and Bronchopulmonary Dysplasia

- Very common association – 36% in a 27 NICU cohort with >900 bronchoscopies
- Results in much greater morbidity and longer hospital stays, more pneumonias, gastroesophageal reflux and more tracheostomies
- Increases work of breathing 4-5 times

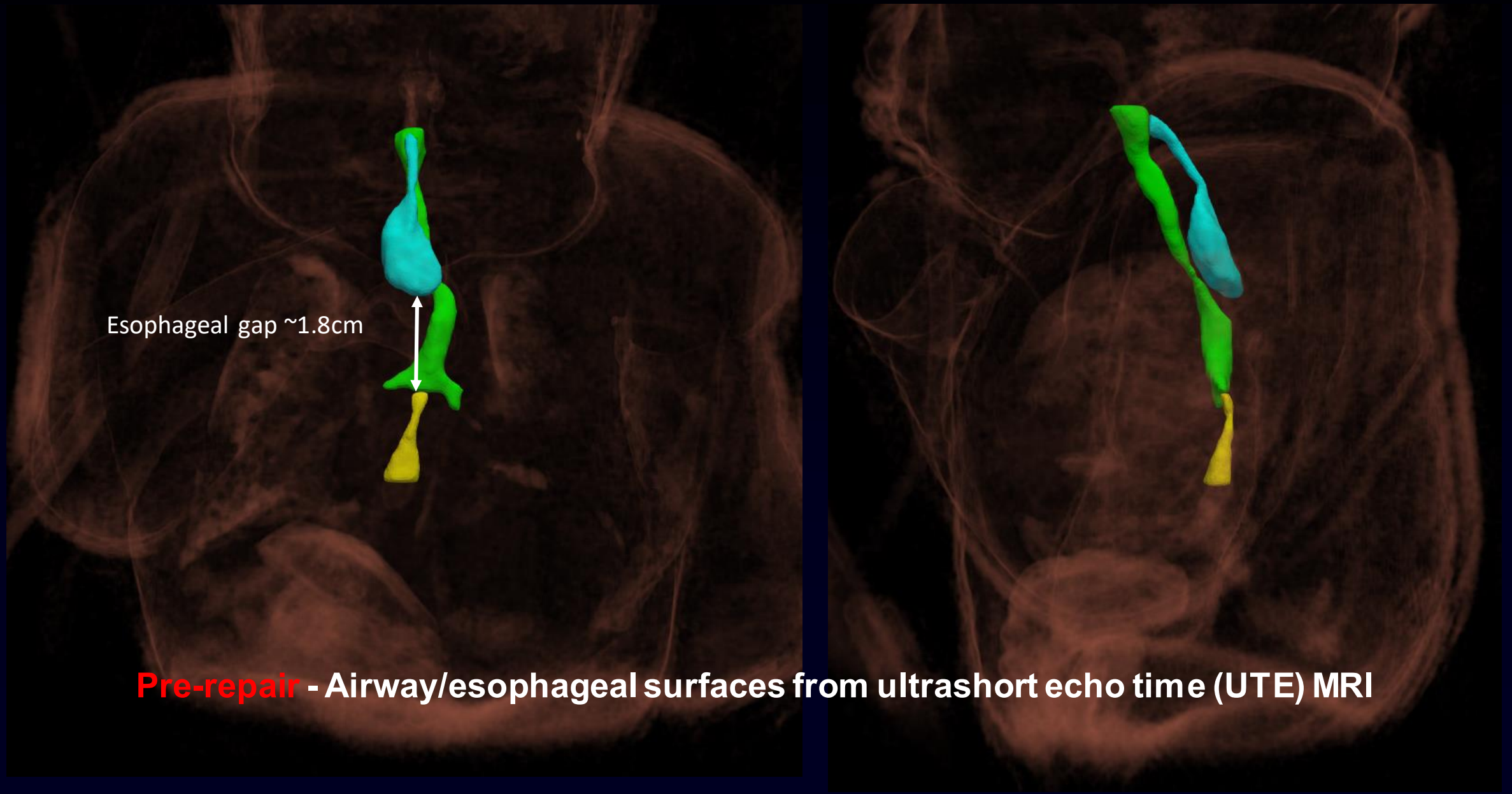


“Good luck with that!”

- Trachea Esophageal Disorders (TED) Grant
- Largely a pulmonary biology grant to look at genetics and developmental aspects of the TEDs
- They wanted to image patients pre-operative
- My experience with CT of TED patients was not good and I described our problems and frustrations
- But I agreed to do my best in reading UTE MRI images

Coronal (view from posterior)

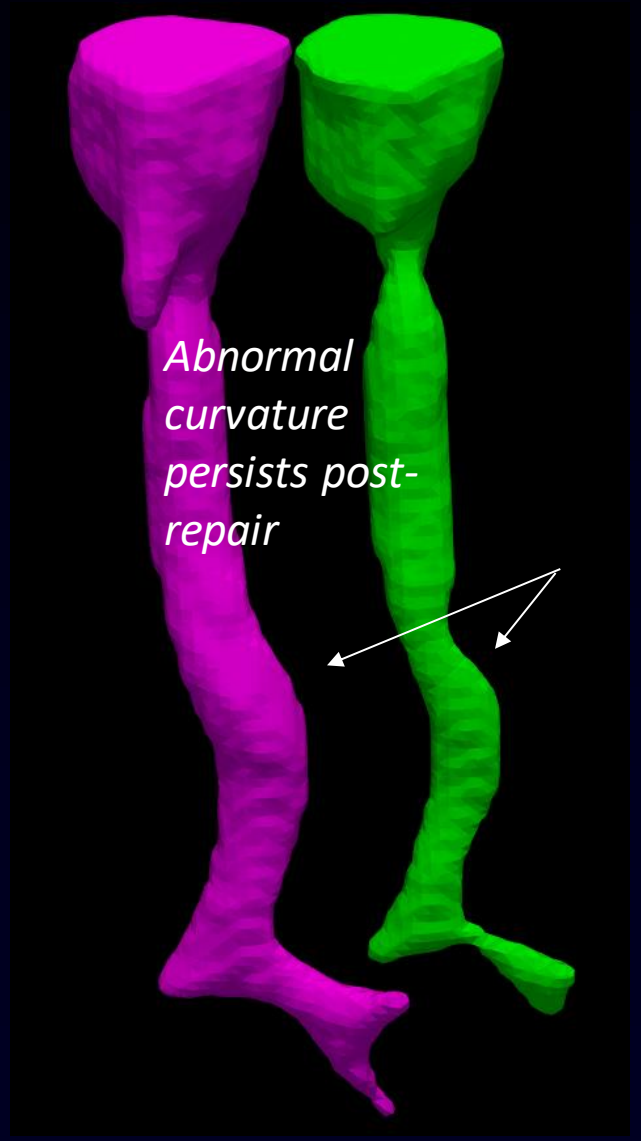
Sagittal



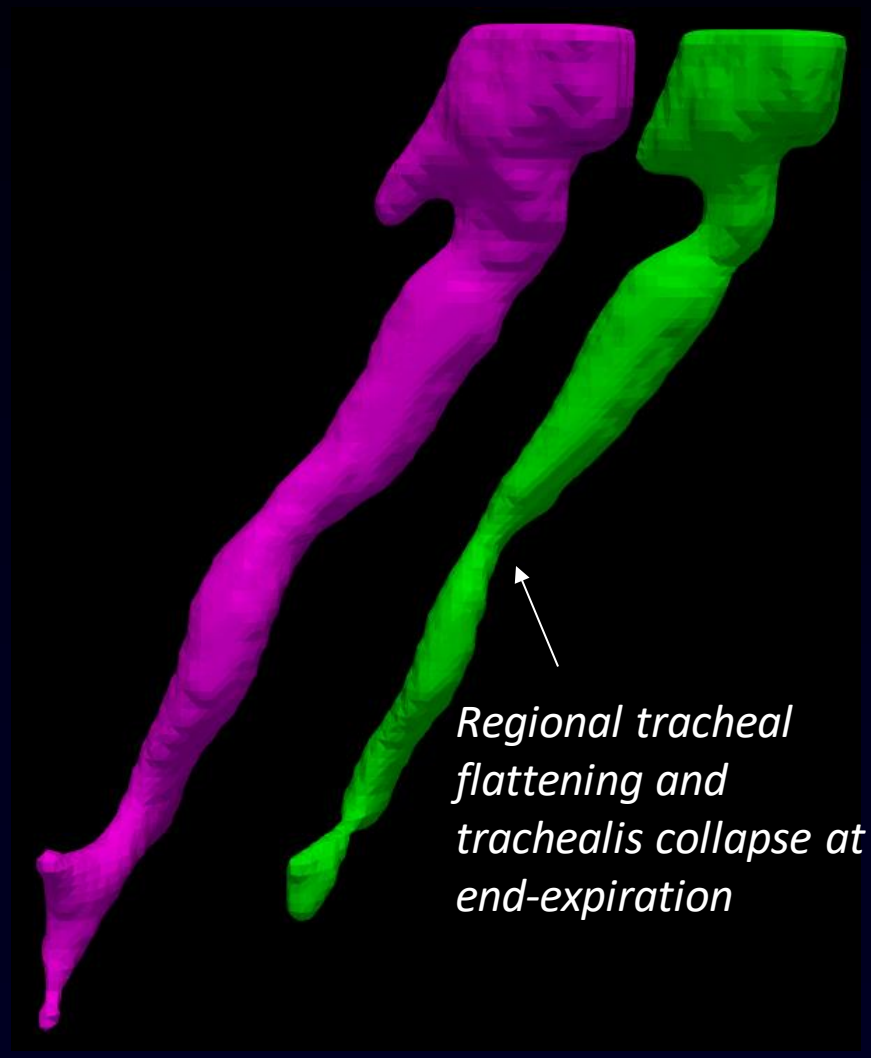
Pre-repair - Airway/esophageal surfaces from ultrashort echo time (UTE) MRI

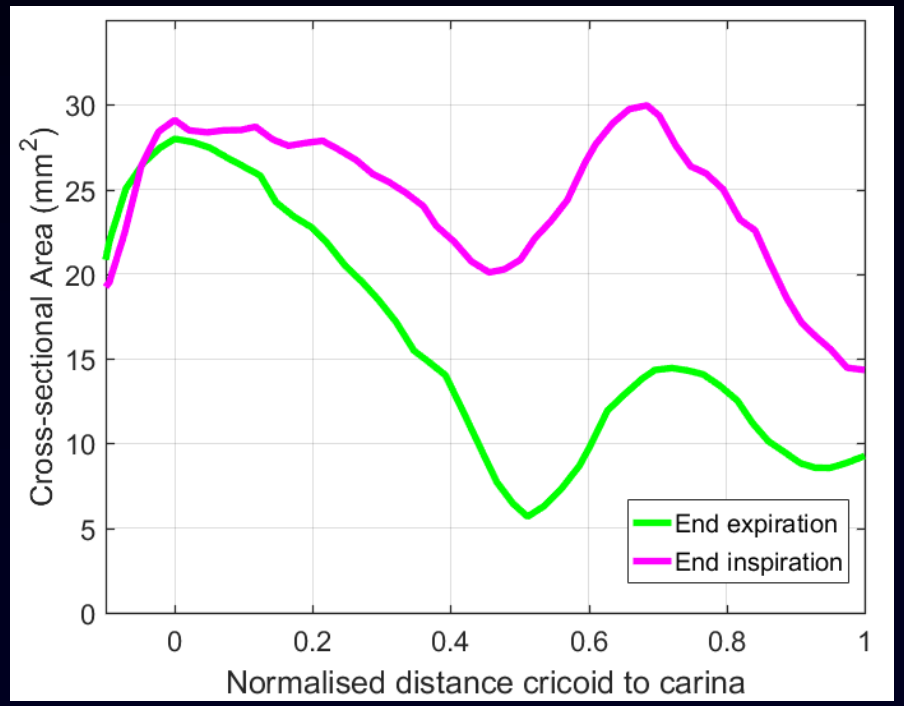
Post-repair Airway/esophageal surfaces from ultrashort echo time (UTE) MRI

View from posterior (minorly tilted)



View from sagittal





Can we prospectively predict and prevent “adverse” outcomes with either persistent symptoms or need for reoperation?

- Pragmatic
  - No, we can never anticipate everything
  - Since outcomes are very good – 90%
    - Added risk and effort may not result in a net gain
    - We work in the real world….

Cincinnati Children's
changing the outcome together

Can we prospectively predict and prevent “adverse” outcomes with either persistent symptoms or need for reoperation?

- Blue Sky Thinking
  - Measure airflow and mask pressure
  - Measure esophageal pressure
  - Apply computational fluid mechanics and flow structure interaction
- Voila! – Airway compliance and compression
  - Then the surgeons need to figure out how to address the specific issue

Cincinnati Children's
changing the outcome together

Cincinnati Children's™
changing the outcome together