Airway Malacia
Evaluation and Management

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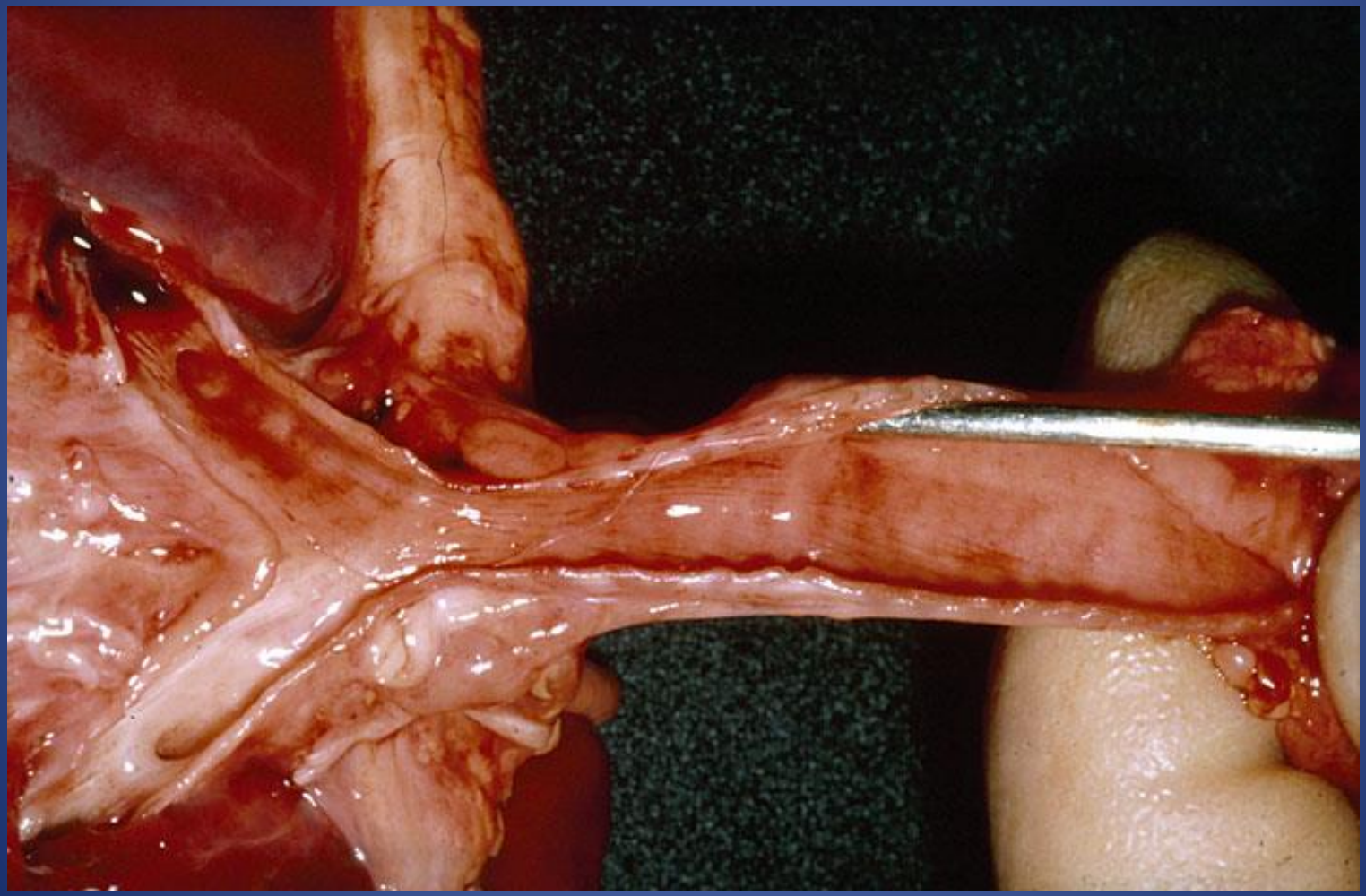
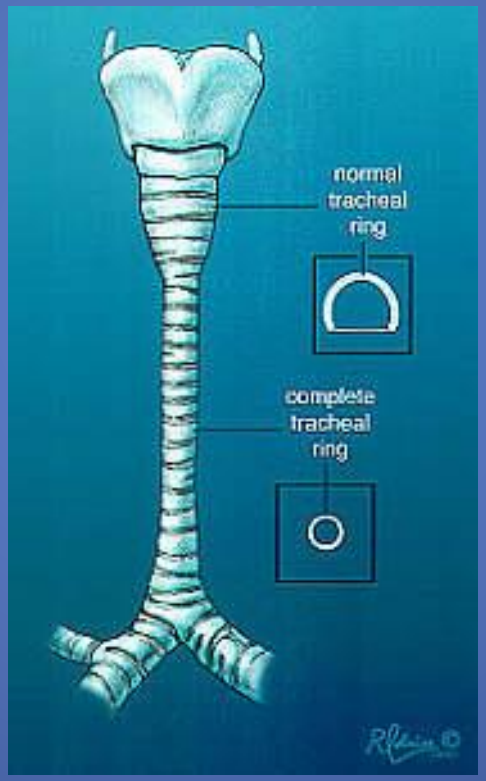
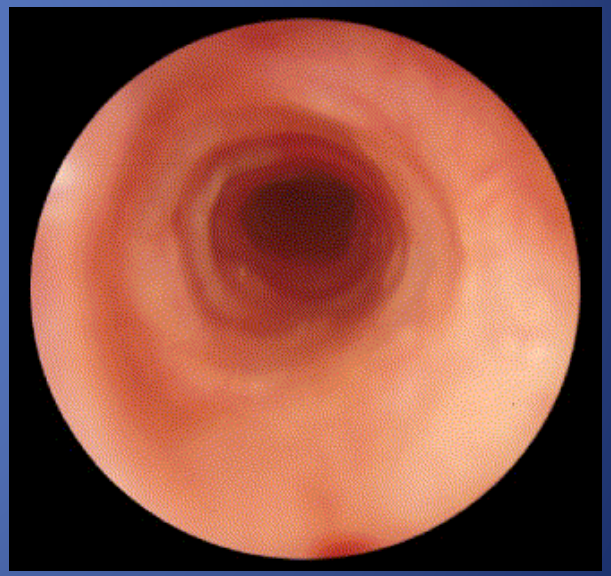
Tracheal Stenosis



What Are the Major Congenital Airway Abnormalities From Cricoid to Major Bronchi

- The narrow
- The deviant
- The floppy





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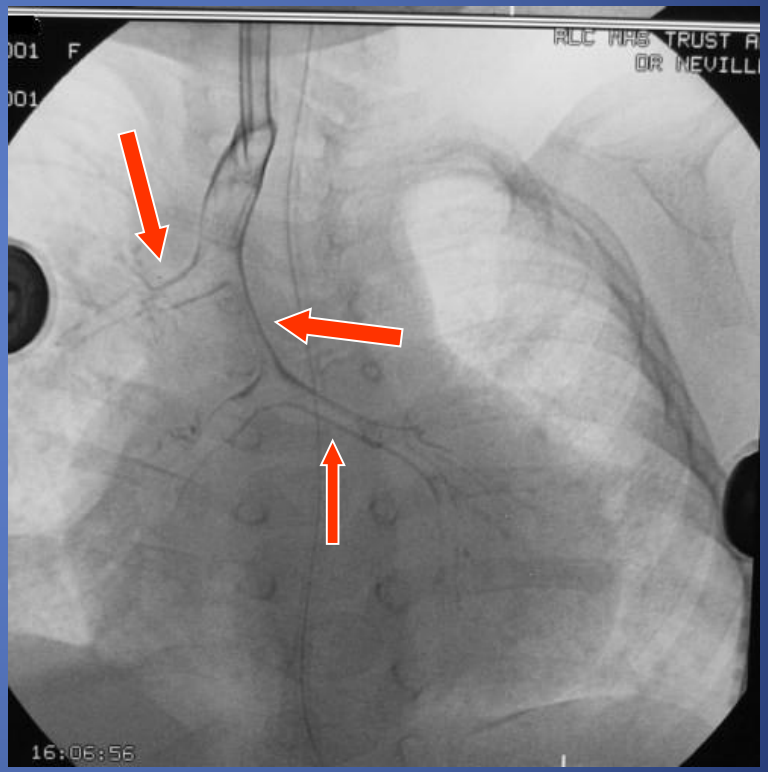

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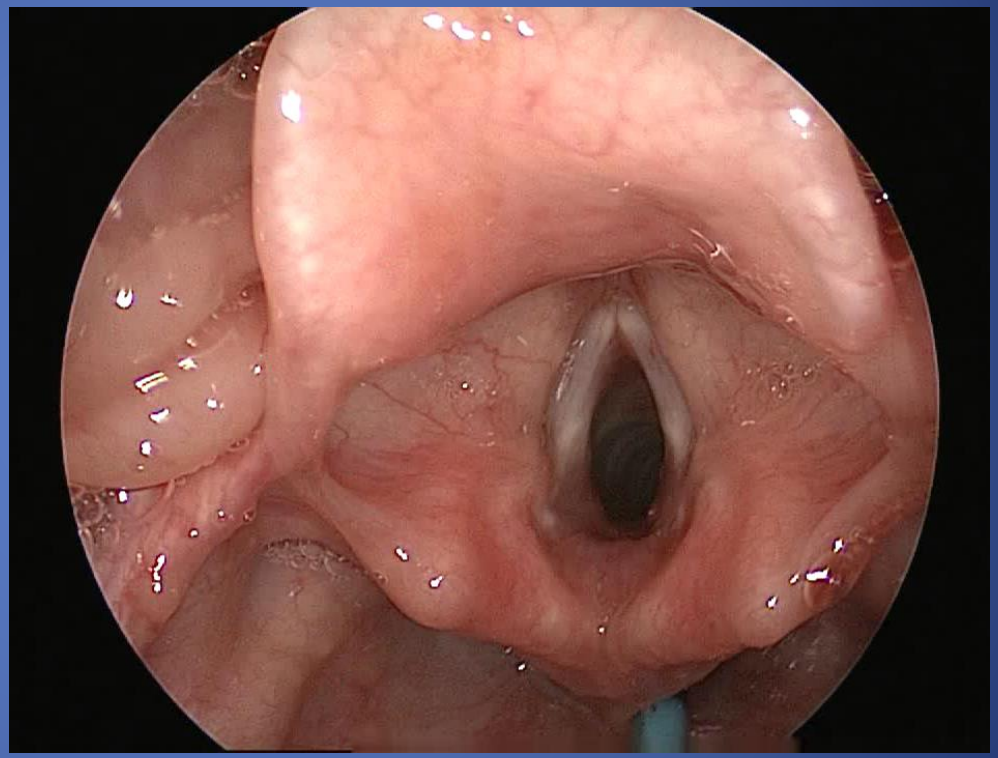

What Are the Major Congenital Airway Abnormalities From Cricoid to Major Bronchi

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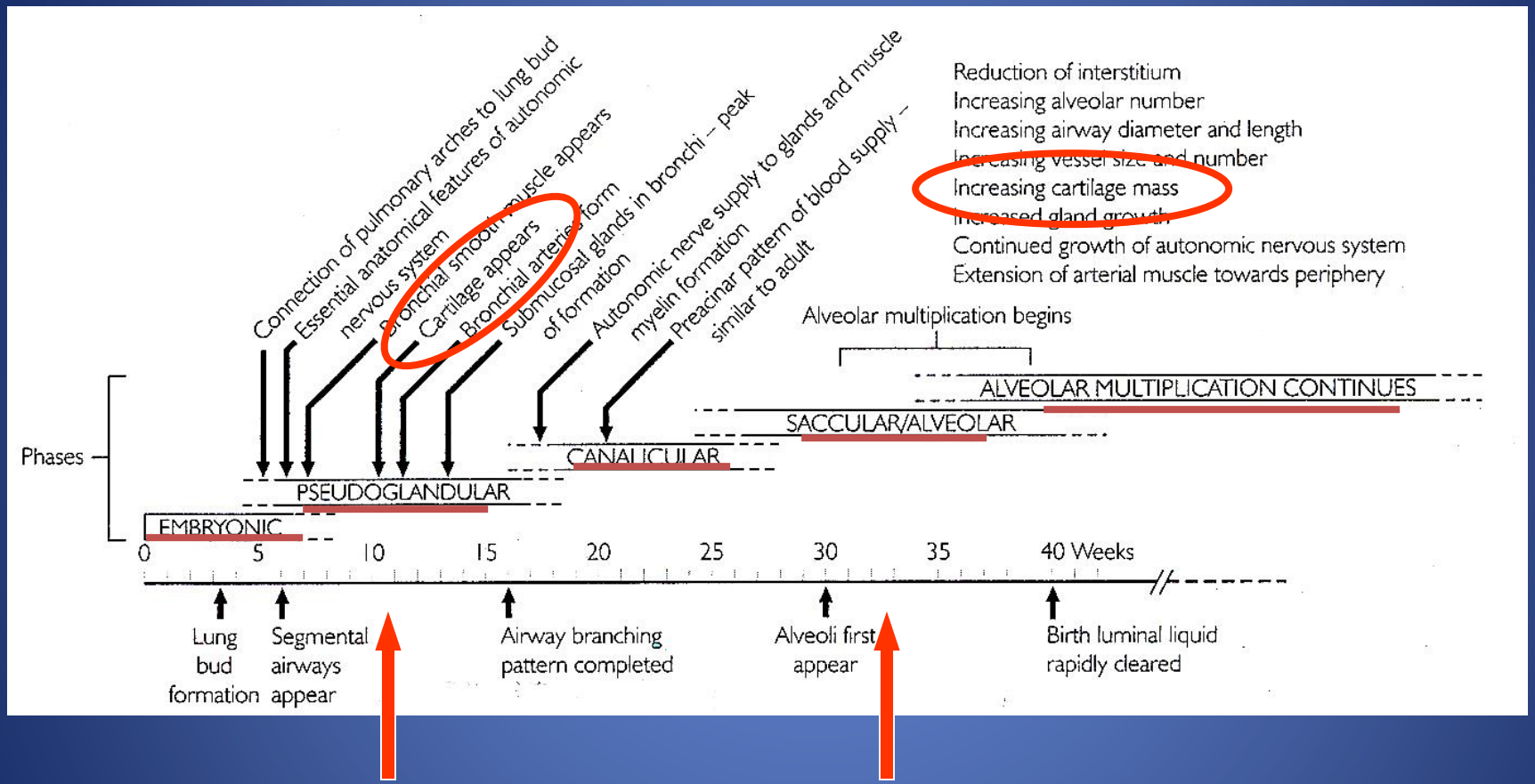
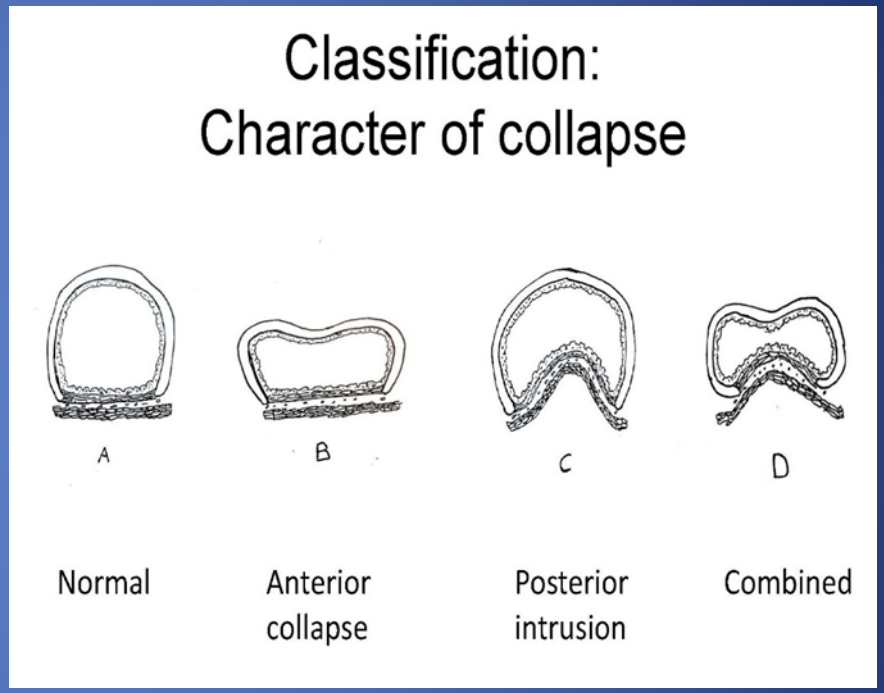
TRACHEO-BRONCHO- MALACIA

- What is normal?
- How do you diagnose it?
- How do you treat it?
- Can malacic cartilage recover?
- Especially in the premature airway


Connection of pulmonary arches to lung bud
Essential anatomical features of autonomic nervous system
Bronchial smooth muscle appears
Cartilage appears
Bronchial arteries form
Submucosal glands in bronchi – peak of formation
Autonomic nerve supply to glands and muscle
myelin formation
Preacinar pattern of blood supply – similar to adult
Reduction of interstitium
Increasing alveolar number
Increasing airway diameter and length
Increasing vessel size and number
Increasing cartilage mass
Increased gland growth
Continued growth of autonomic nervous system
Extension of arterial muscle towards periphery
Alveolar multiplication begins
ALVEOLAR MULTIPLICATION CONTINUES
SACCULAR/ALVEOLAR
CANALICULAR
PSEUDOGLANDULAR
EMBRYONIC
Phases
0
5
10
15
20
25
30
35
40 Weeks
Lung bud formation
Segmental airways appear
Airway branching pattern completed
Alveoli first appear
Birth luminal liquid rapidly cleared

A Definition of Malacia

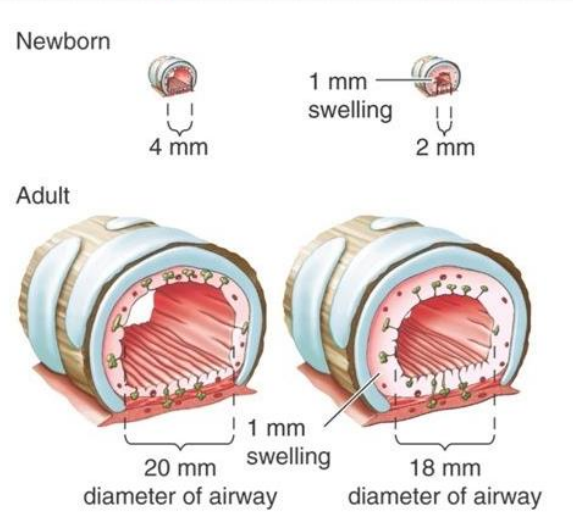
- ERS taskforce
- What is normal?



A Definition of Malacia

- ERS taskforce 2019
- What is normal?

The diameter of an infant's airway is approximately 4 mm, in contrast to an adult's airway diameter of 20 mm.



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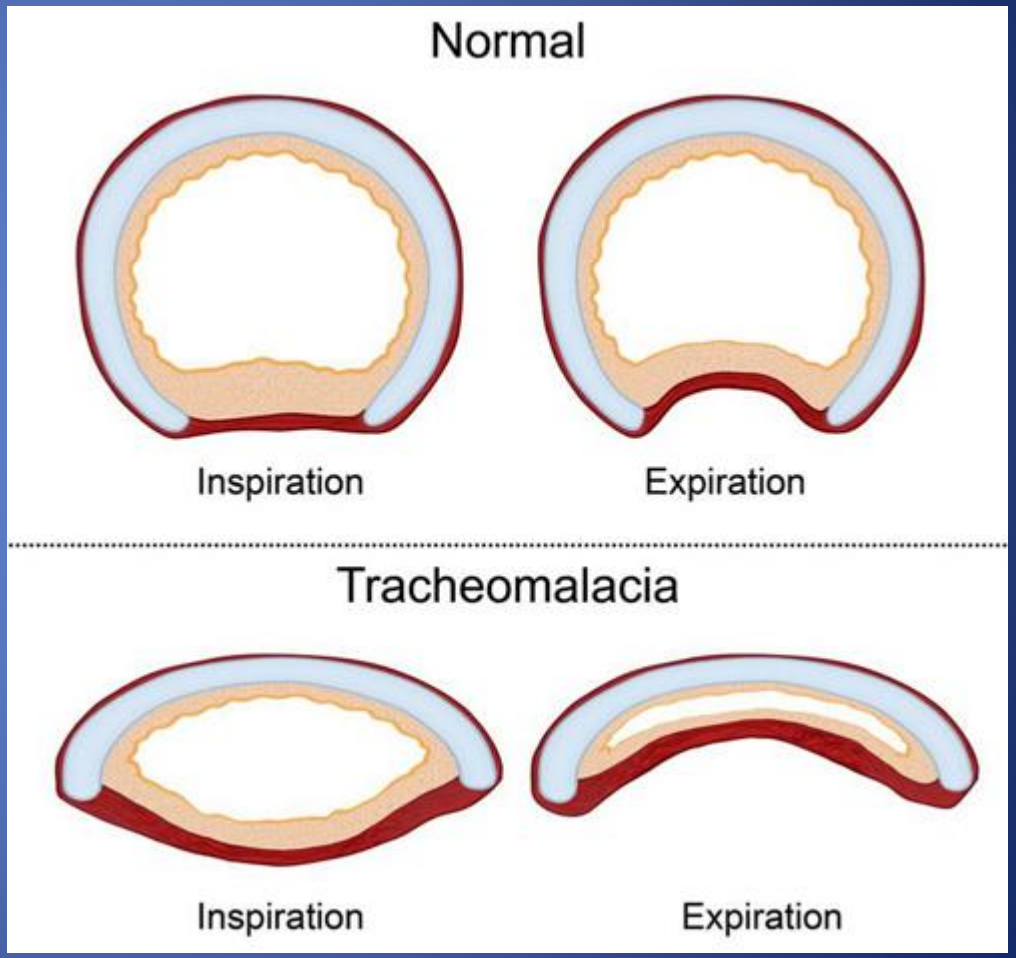

A Definition of Malacia

- ERS taskforce 2019
- What is normal?

- NORMAL:
  Up to 50% of internal lumen
- MILD MALACIA:
  50-75% loss
- MODERATE MALACIA:
  75% - 90%
- SEVERE MALACIA:
  >90%

Causes of Malacia

Two main groups

- Primary
- Secondary

Primary or Congenital

There is an intrinsic alteration of airway cartilage

Secondary or Acquired

Where the cartilage is embryologically normal but developmentally malformed because of external pressure on the airway wall or acquired from luminal infection, inflammation or surgical intervention.

Conditions Associated with Malacia

PRIMARY OR CONGENITAL

- Congenital idiopathic
- Congenital abnormalities of cartilage
- Congenital anomalies of aerodigestive tract
- Anomalies of respiratory tract development
- Syndromic conditions

SECONDARY OR ACQUIRED

- Cardiovascular anomalies
- Skeletal anomalies
- Infections and inflammatory
- Tracheobronchial injury
- Tumours and cysts
- Post surgical procedures

Clinical presentation

Brassy or barking cough
Stridor
Wheezing
Noisy breathing
Recurrent and/or prolonged respiratory infections
Dying spells
Feeding difficulties
Dyspnoea

- Intermittent
- Persistent
- From birth or later childhood
- Mild (intermittent stridor/wheeze)
- Severe:
  - dying spells,
  - apnoea,
  - sudden death,
  - unable to extubate

Making the diagnosis of malacia in children

- Flexible bronchoscopy
- Rigid bronchoscopy
- Plain xray
- CT scan
- Bronchography
- Lung function

Tracheo – oesophageal fistula

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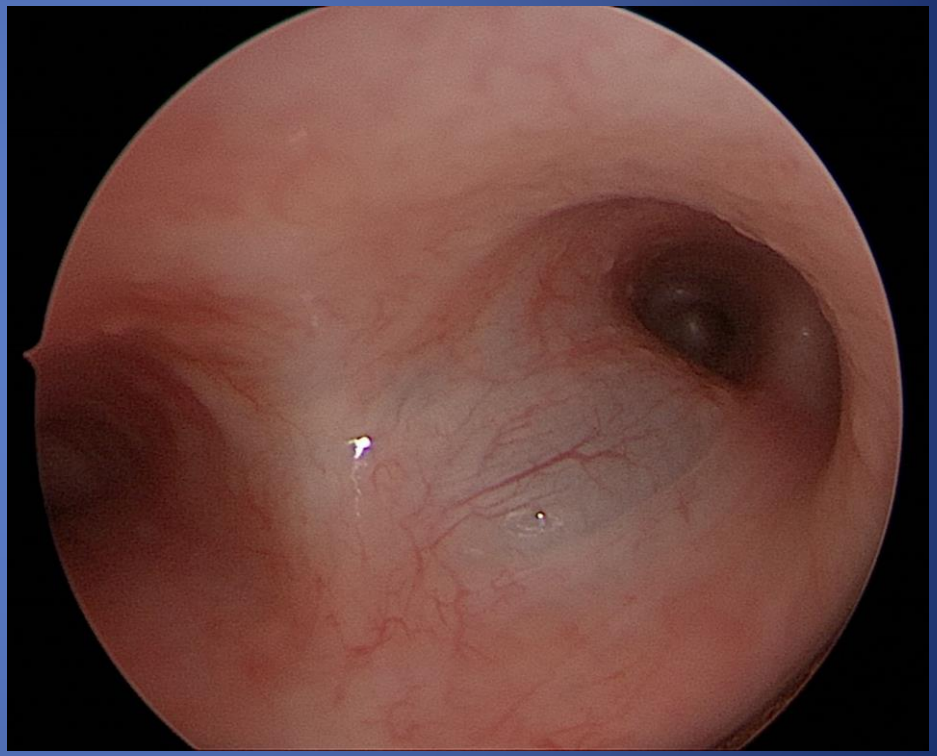
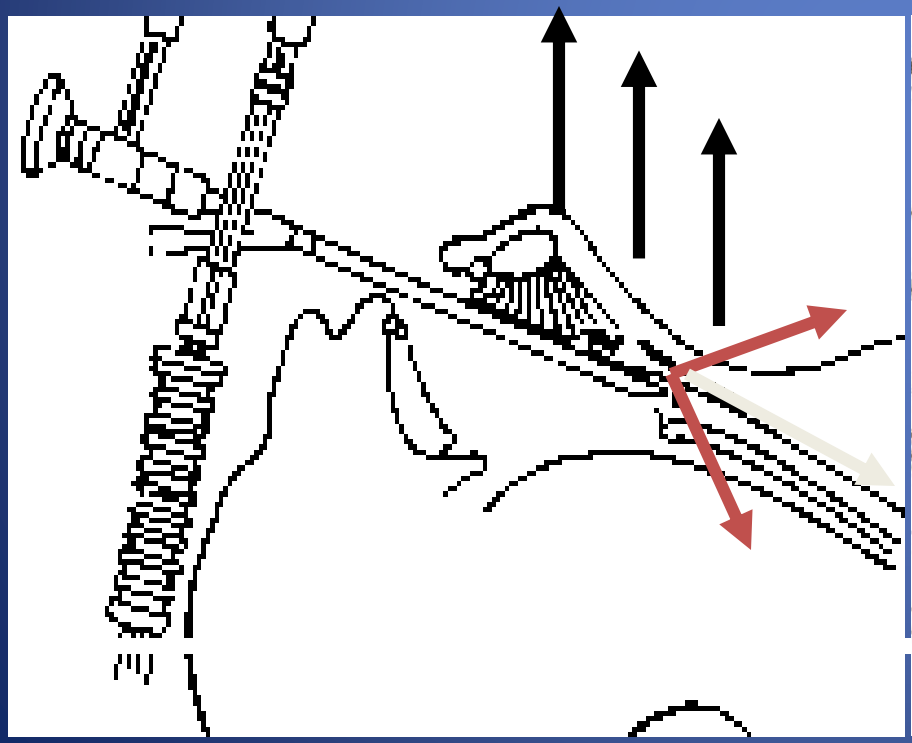
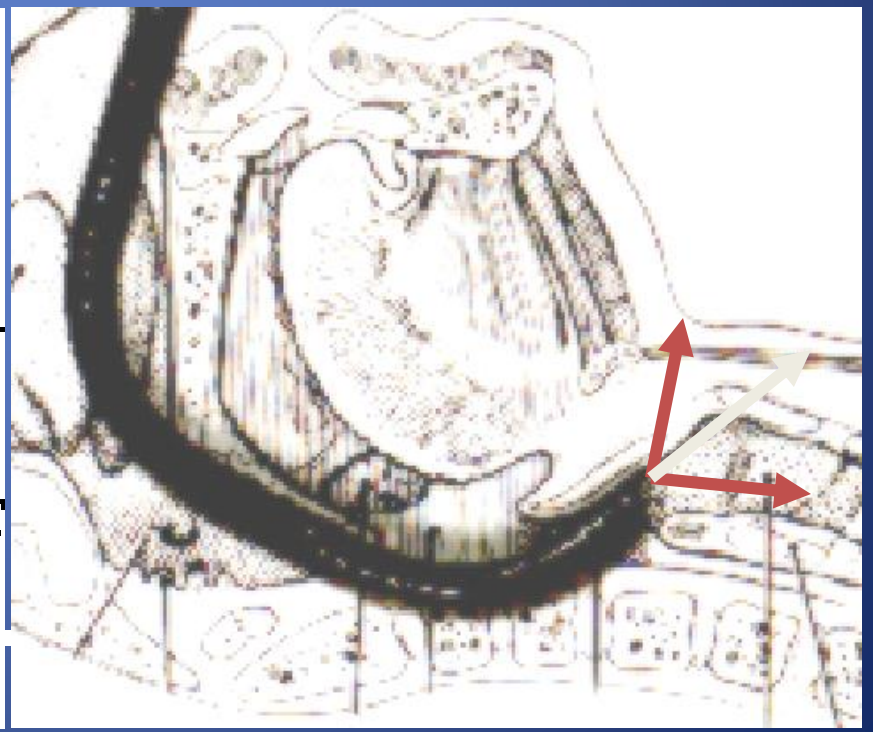
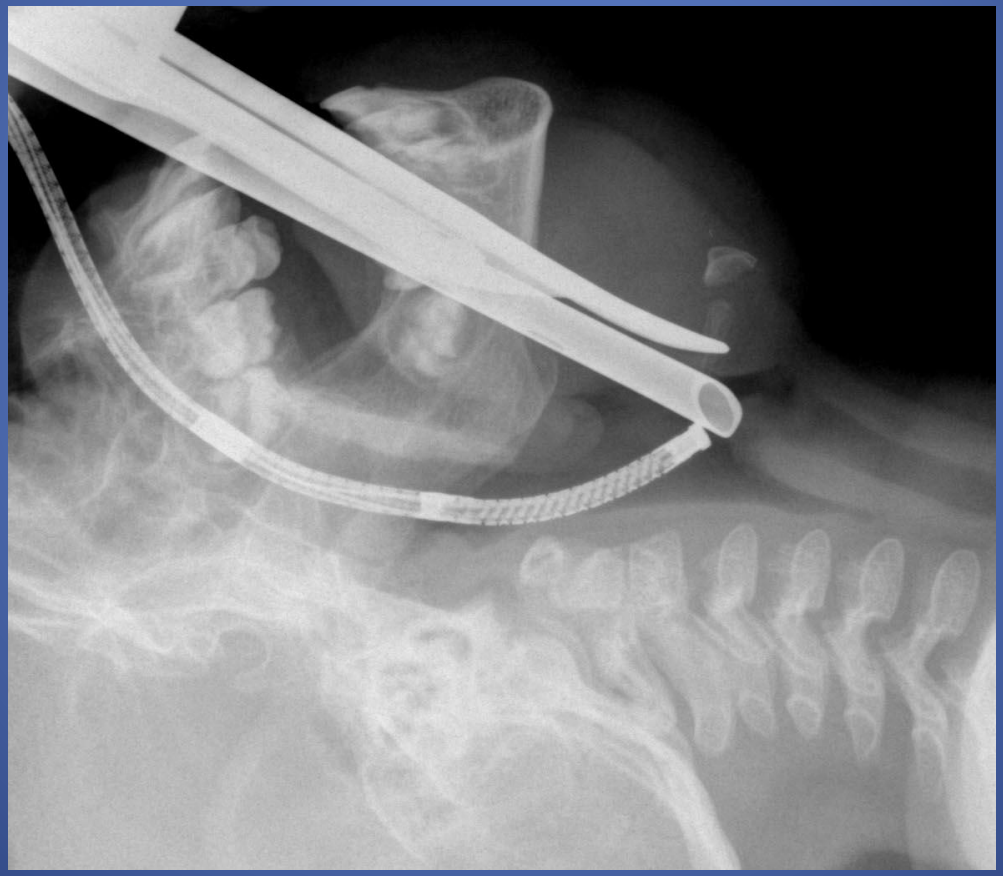


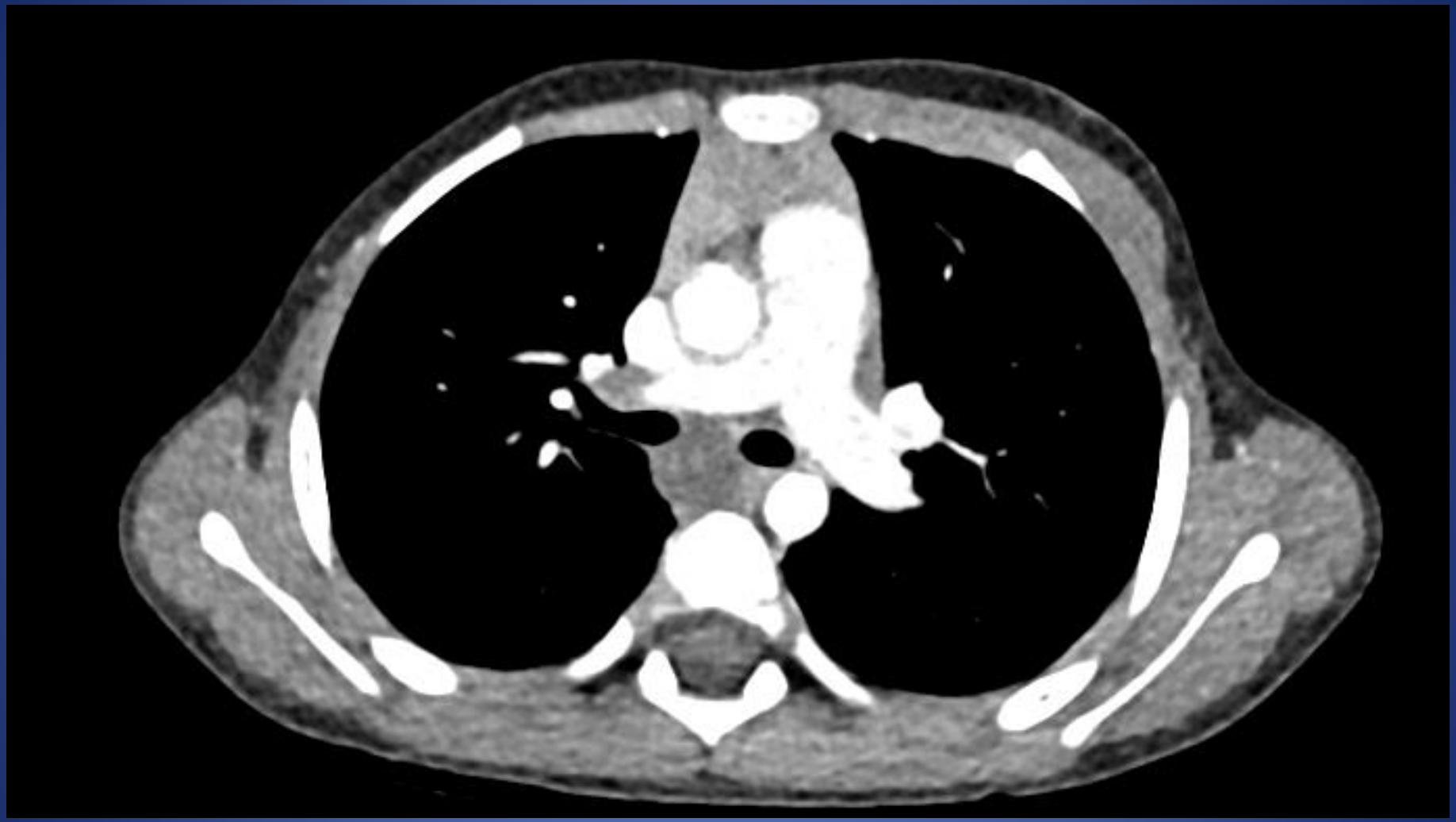
Approach to the airway...





Rigid and flexible bronchoscopes





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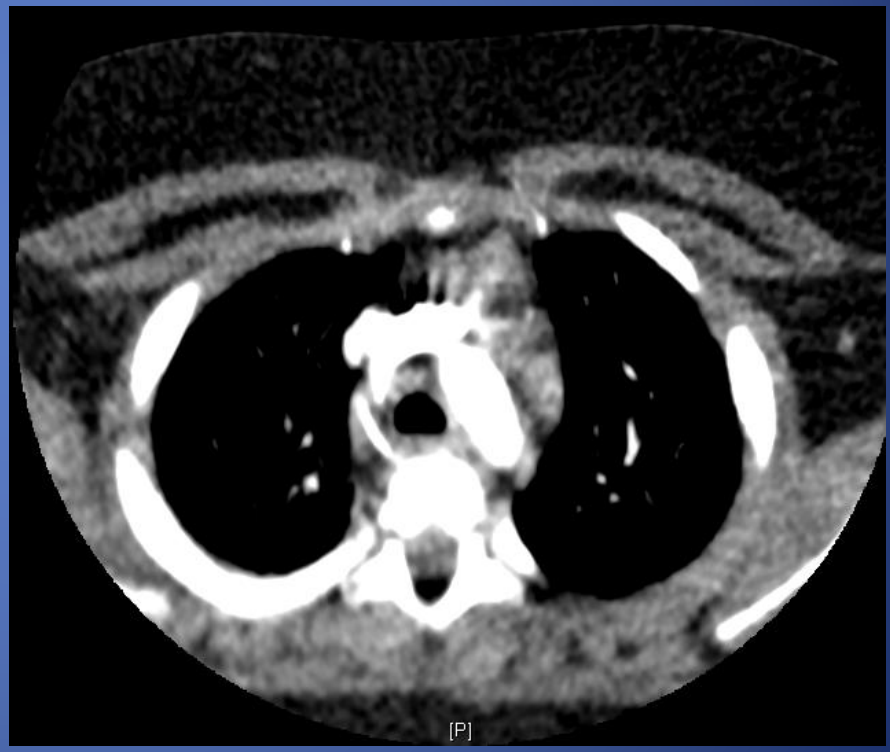
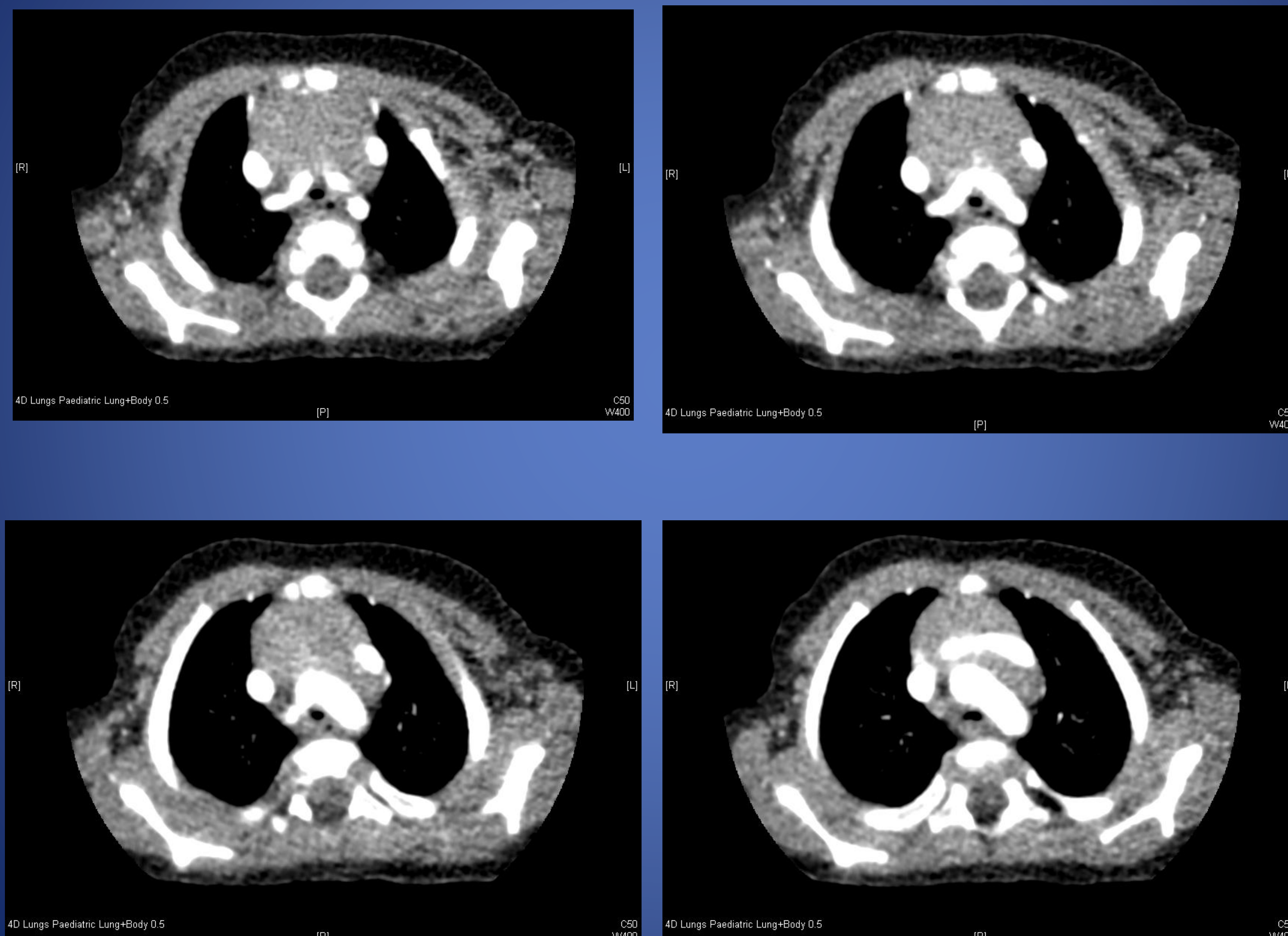
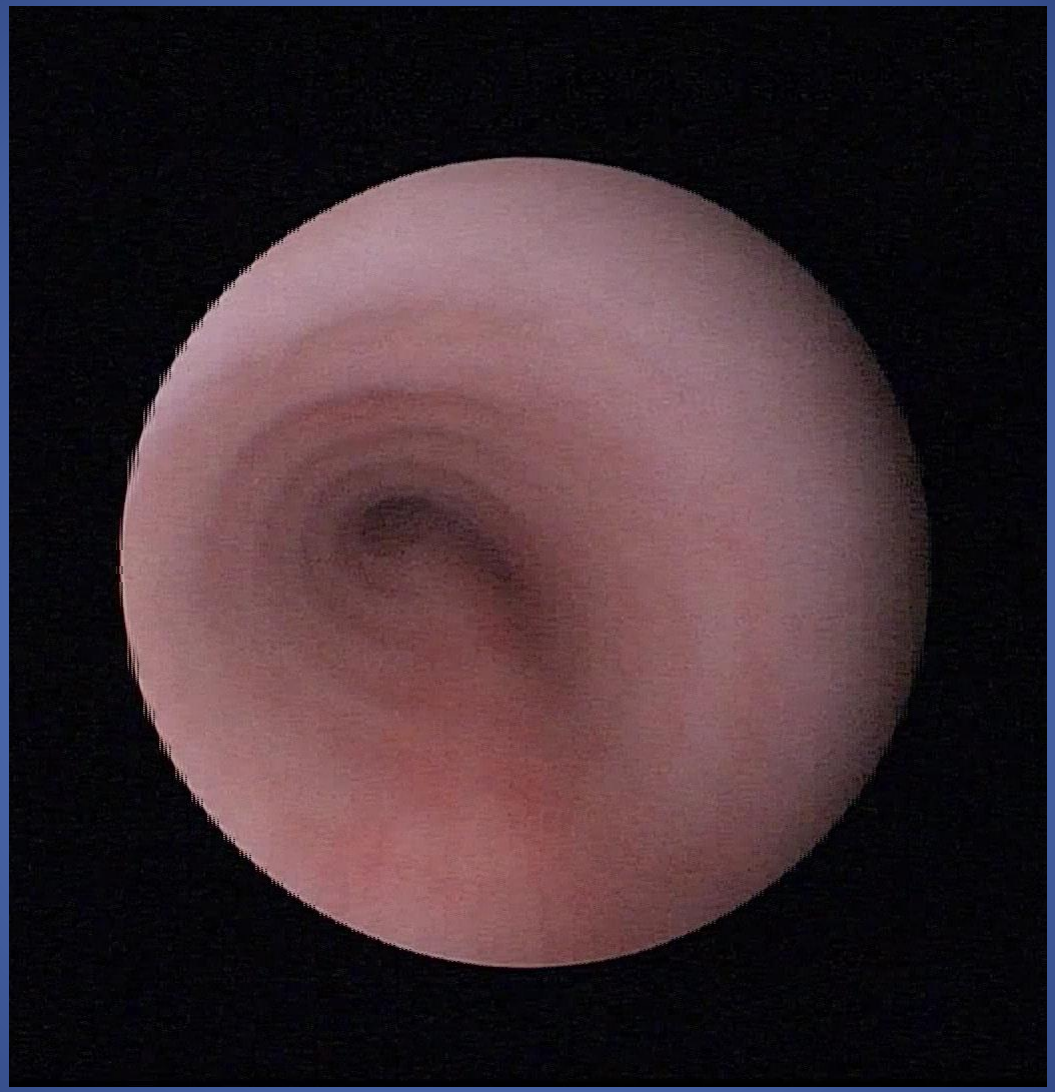
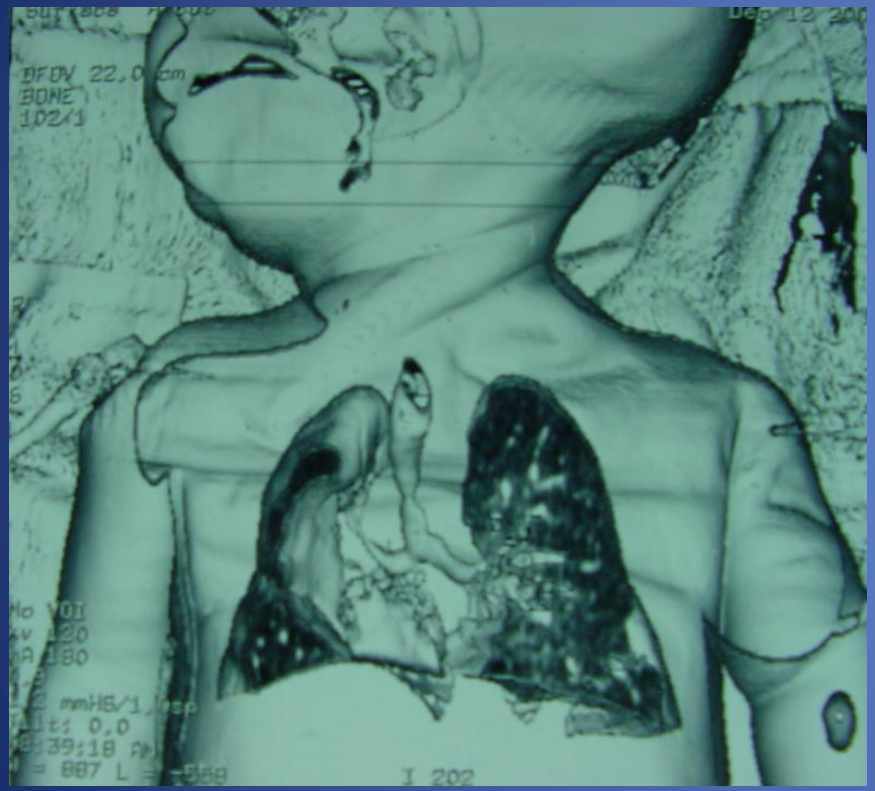


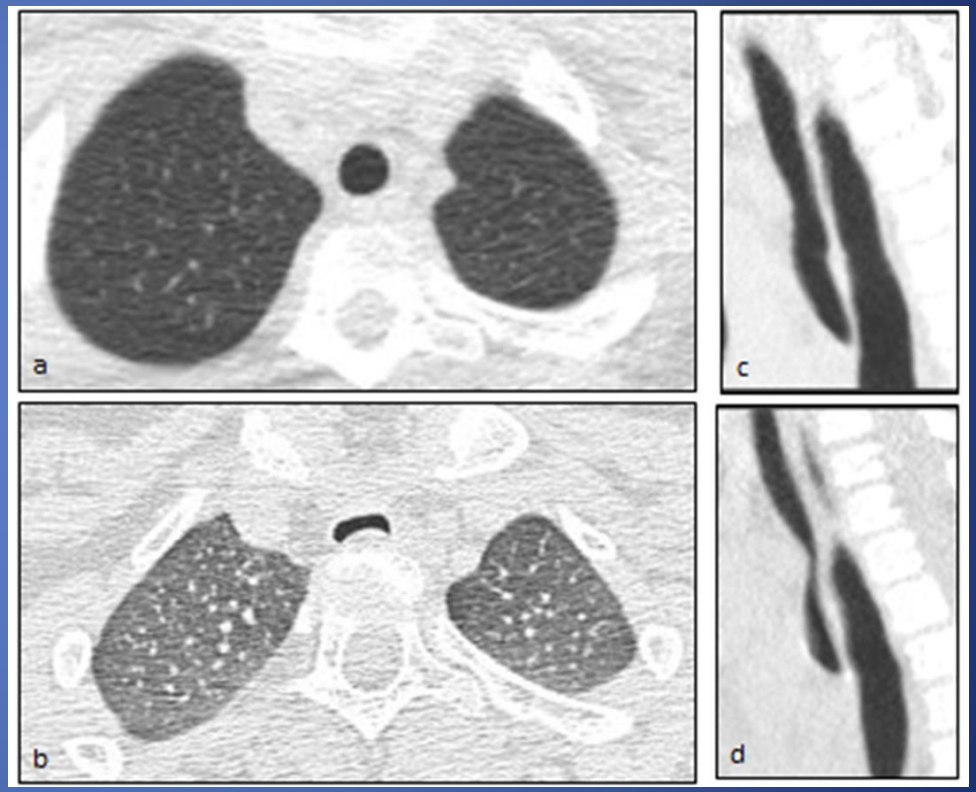
TRANSVERSE COURSE OF THE INNOMINATE ARTERY





CT technology is improving





3-D reconstuction

4-D CT scanning

?MRI

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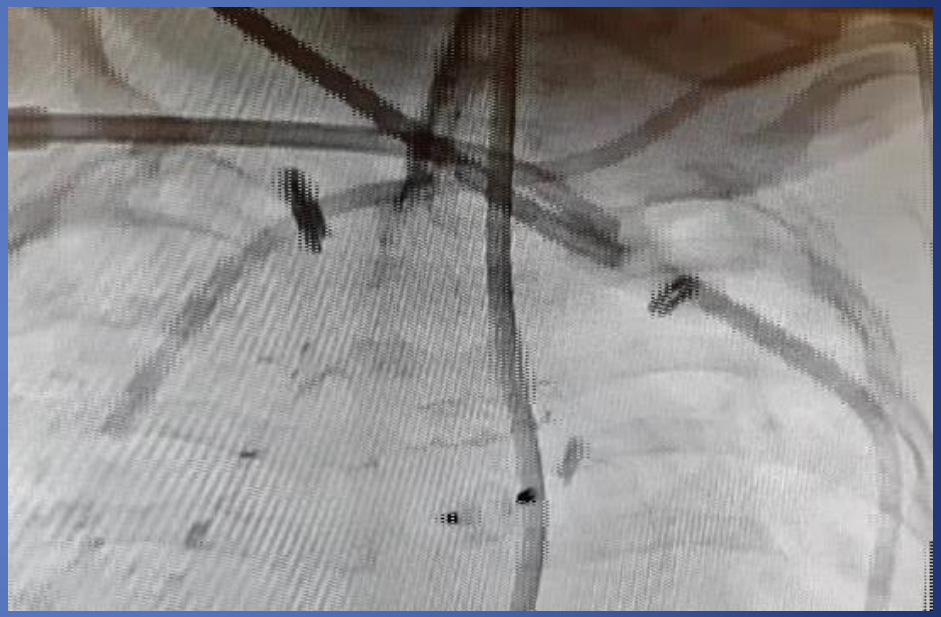
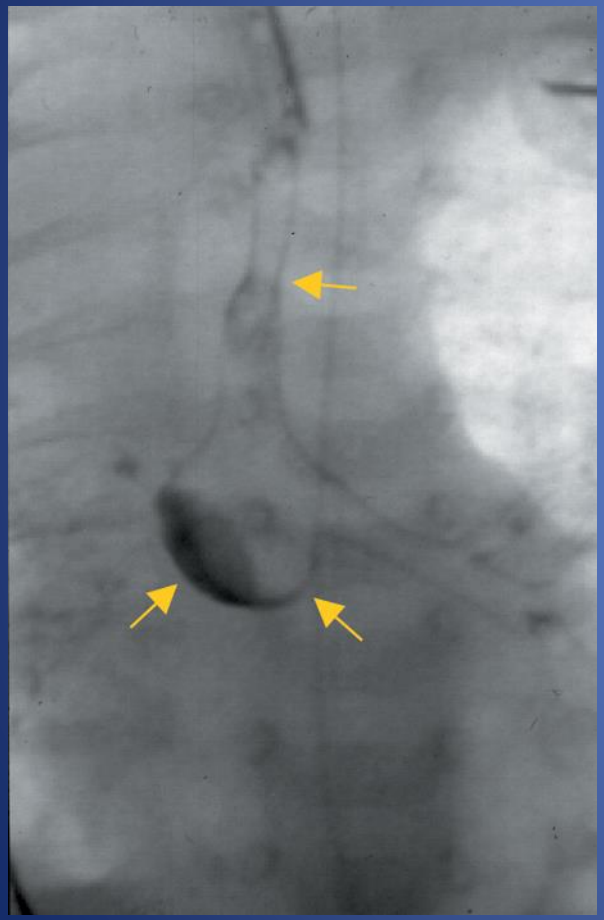
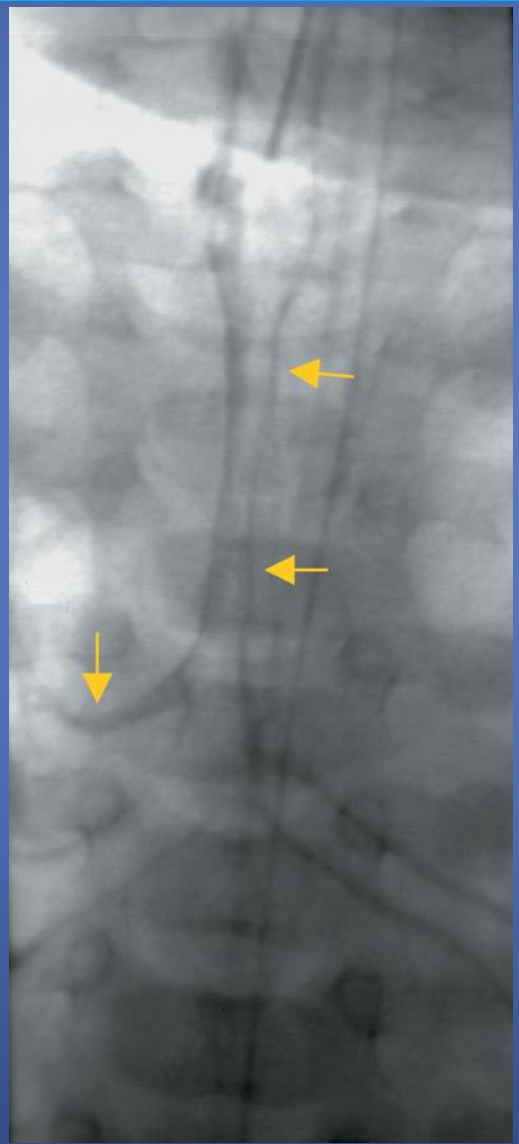
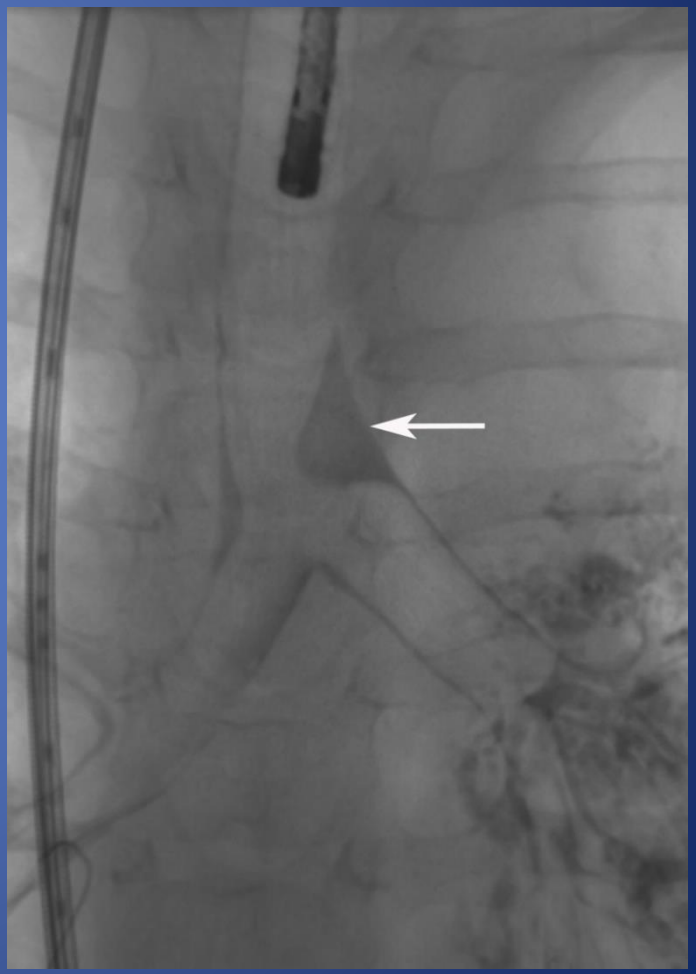


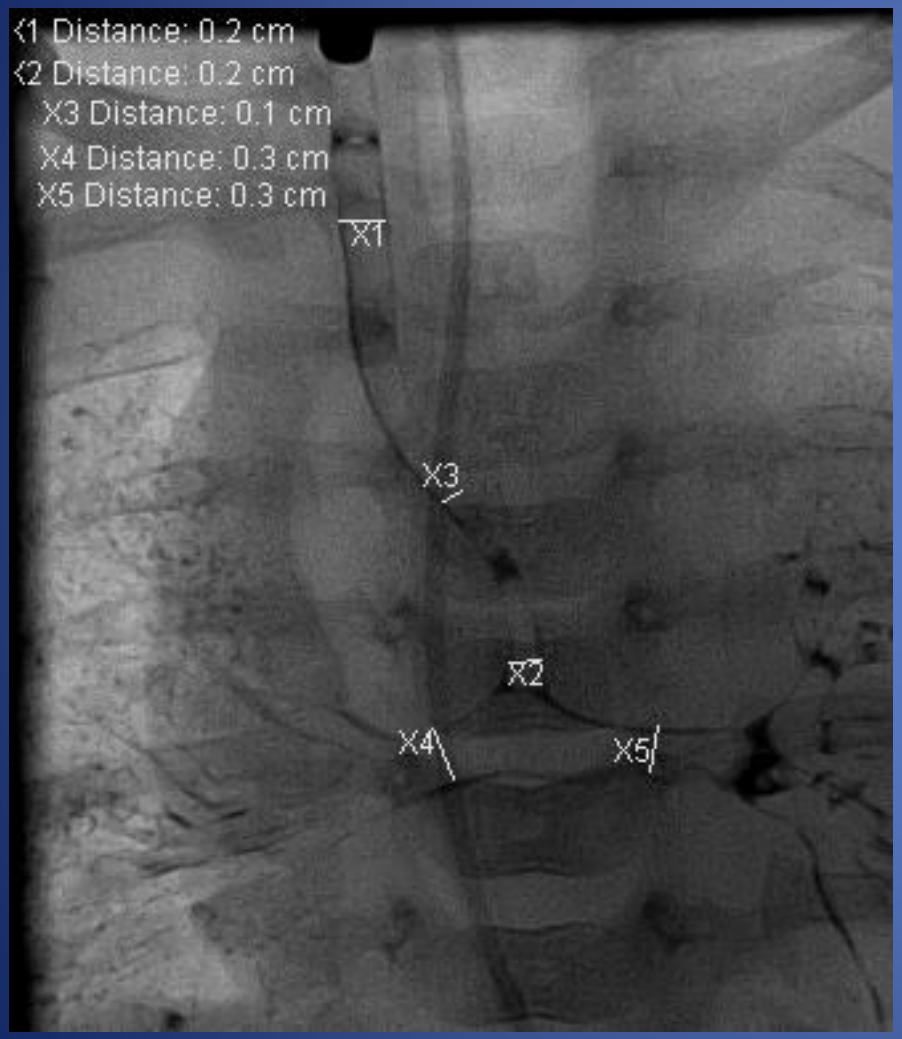
Investigating Tracheal Pathology
Bronchoscopy and bronchogram

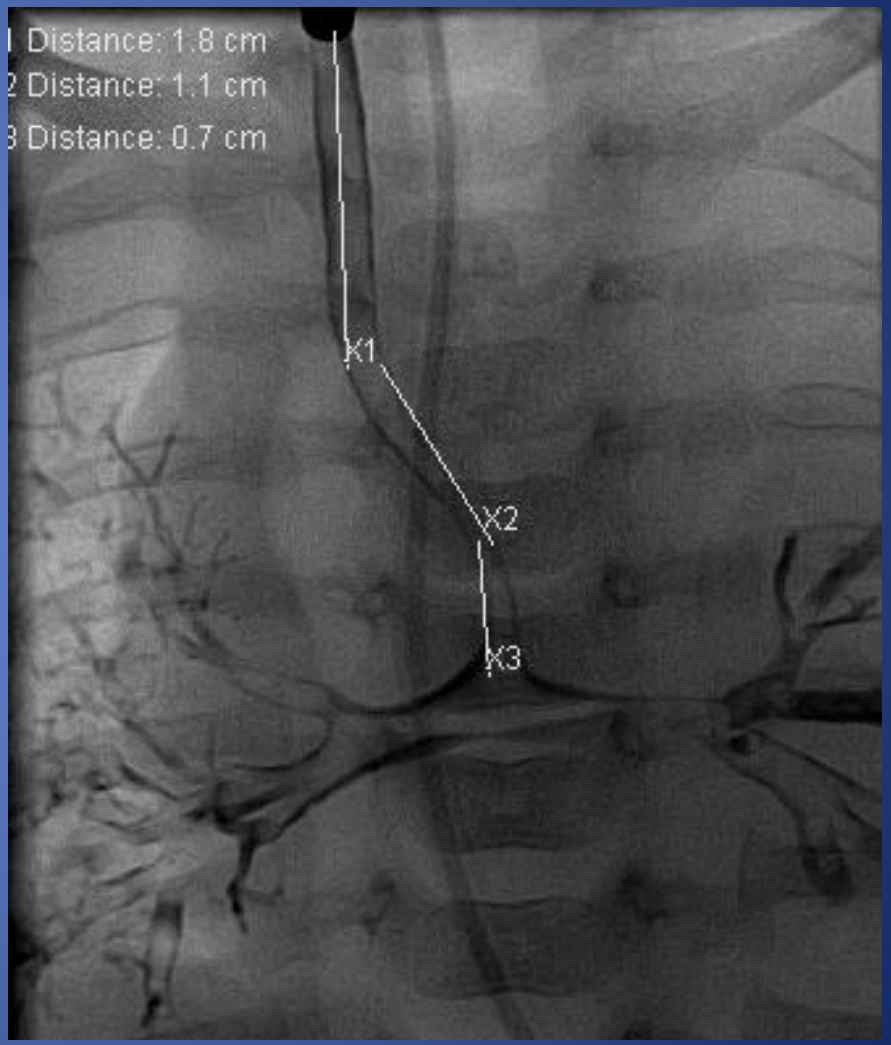


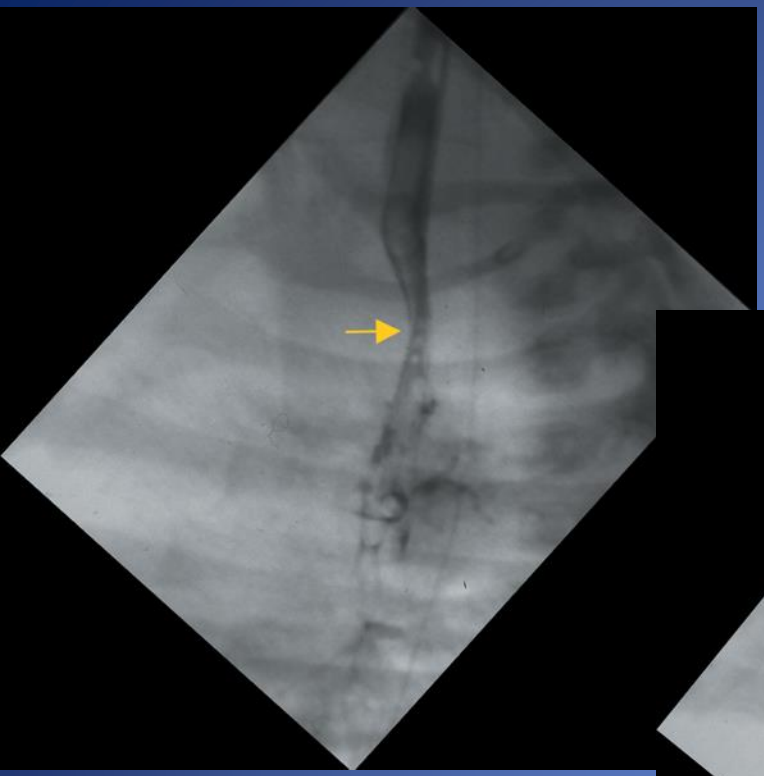




Bronchography







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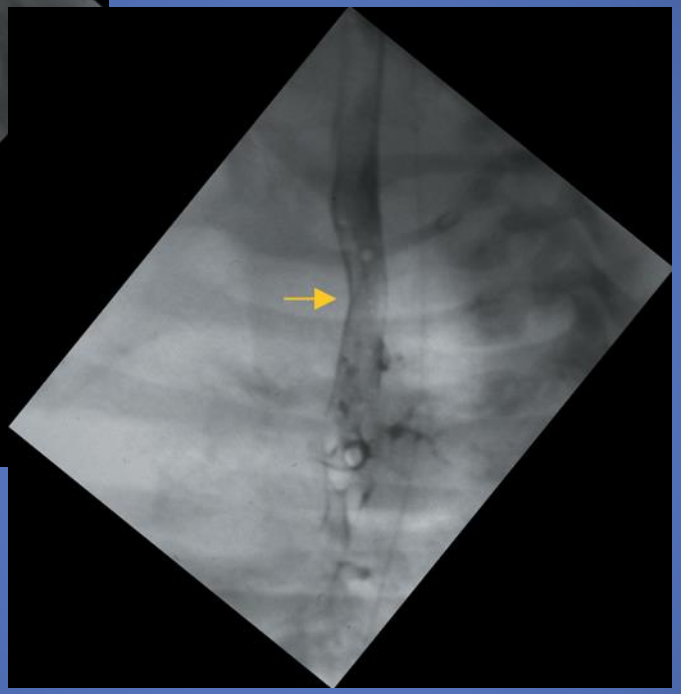

20 PEEP

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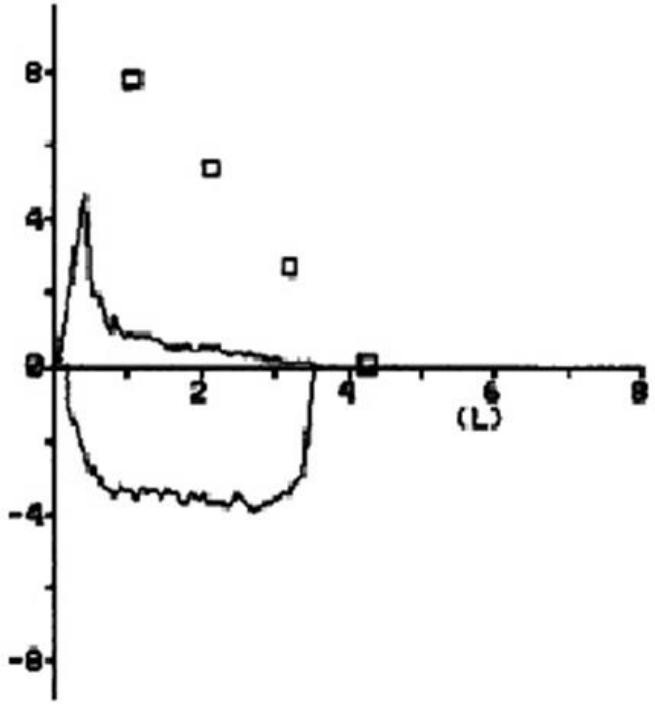
DIAGNOSIS

Pulmonary Function Tests

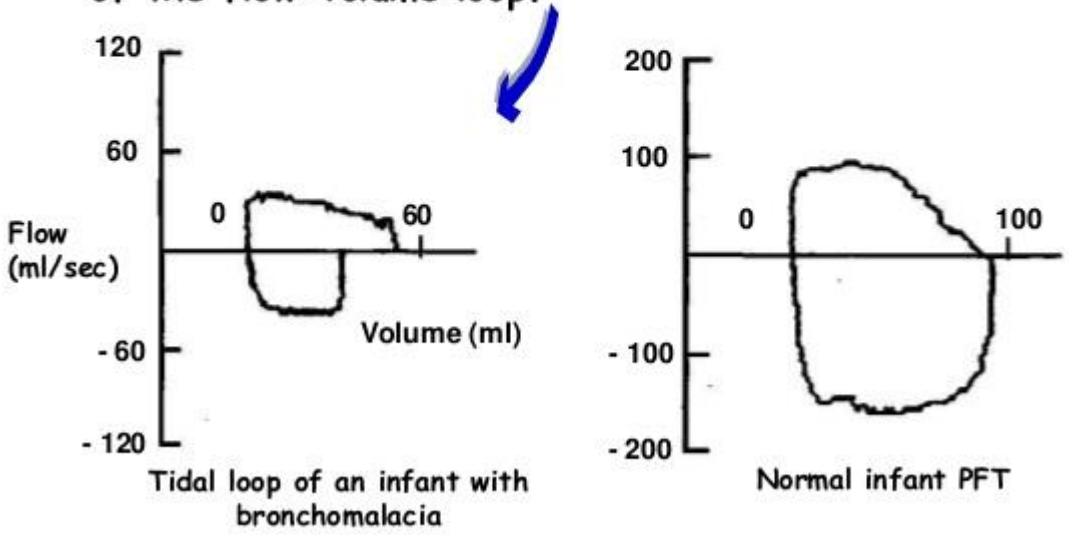
- Supportive but not diagnostic
- The obstruction on the spirometry usually is proportional to the severity of the airway collapse
- Rapid decline in the maximal expiratory flow after a sharp peak (collapse of central airways due to negative transmural pressure)



BRONCHOMALACIA: LUNG FUNCTION TESTING

Large airway obstruction can generally be differentiated by the pattern of the flow-volume loops in spirometry

The patient with bronchomalacia demonstrates flattening of the flow-volume loop.



Treatment options

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- Intraluminal stents
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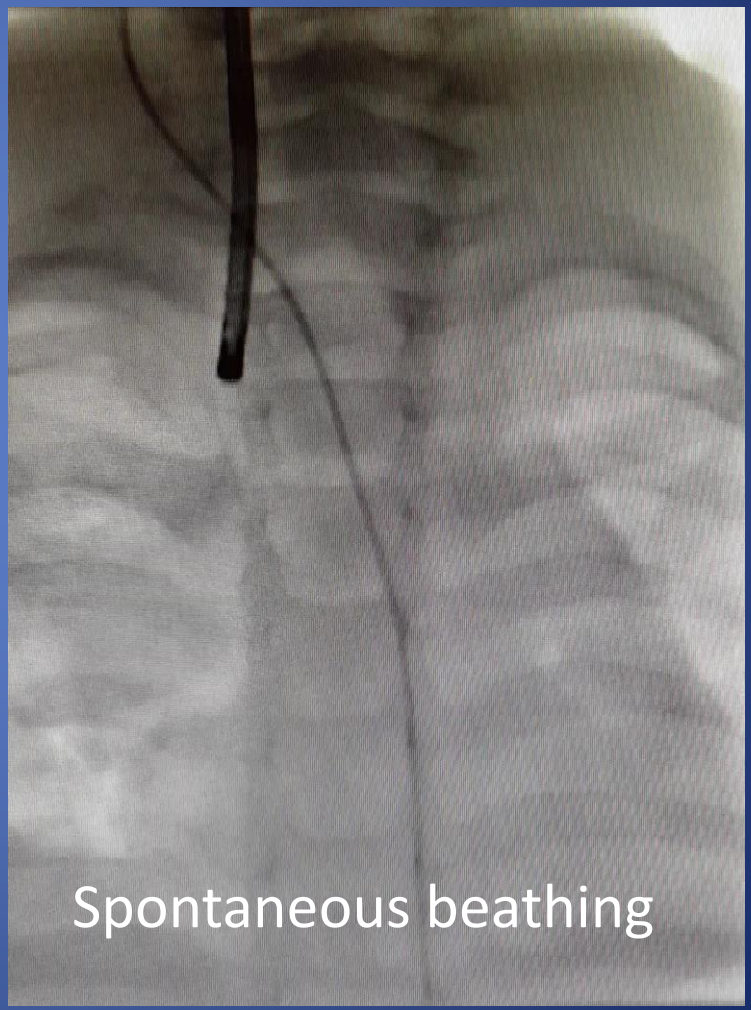

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- Beta 2 Agonists
- Ipratropium bromide
- Methacoline (trachealis constriction)
- Mucolytics (DNAse)
- Antibiotic therapy (Azithromycin)
- Physiotherapy
- Treatment of co-morbidities:
  - GOR
  - Eosinophilic oesophagitis
  - General respiratory health

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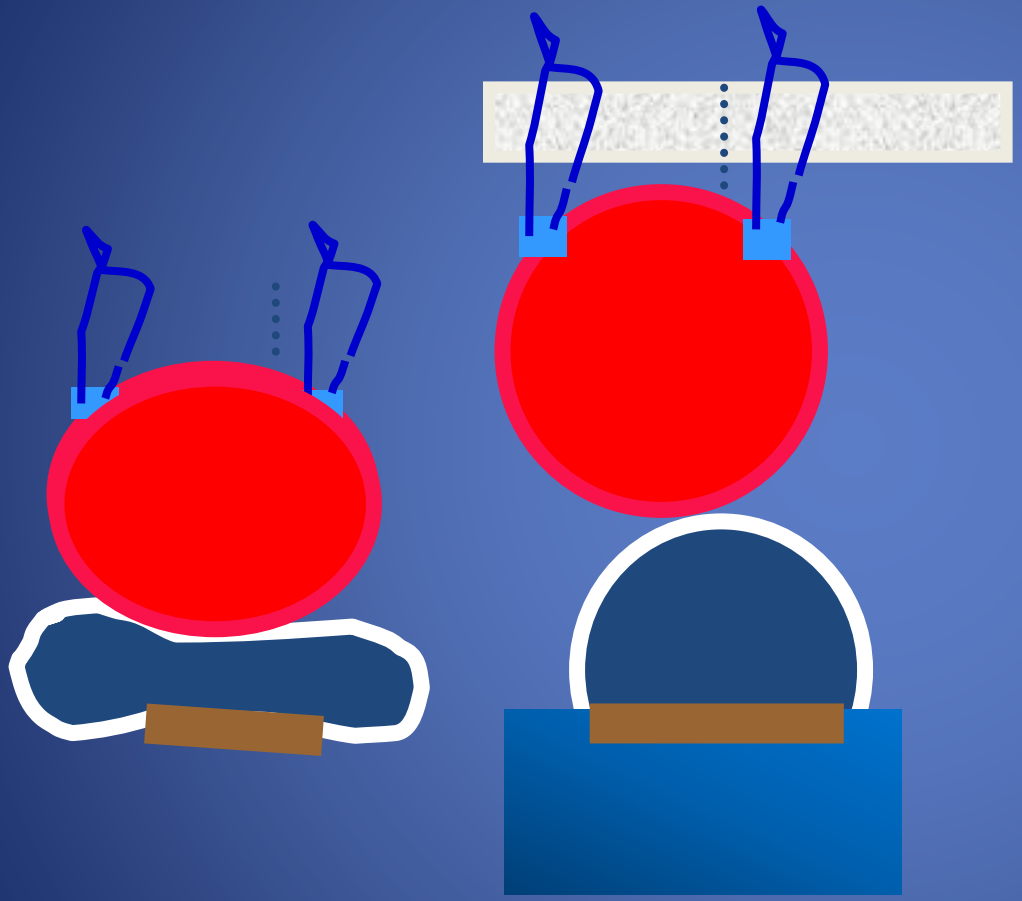
- Aortopexy
- Tracheal resection
- Tracheostomy
- Posterior tracheopexy
- External stents

Surgical Mx of Tracheomalacia: Aortopexy



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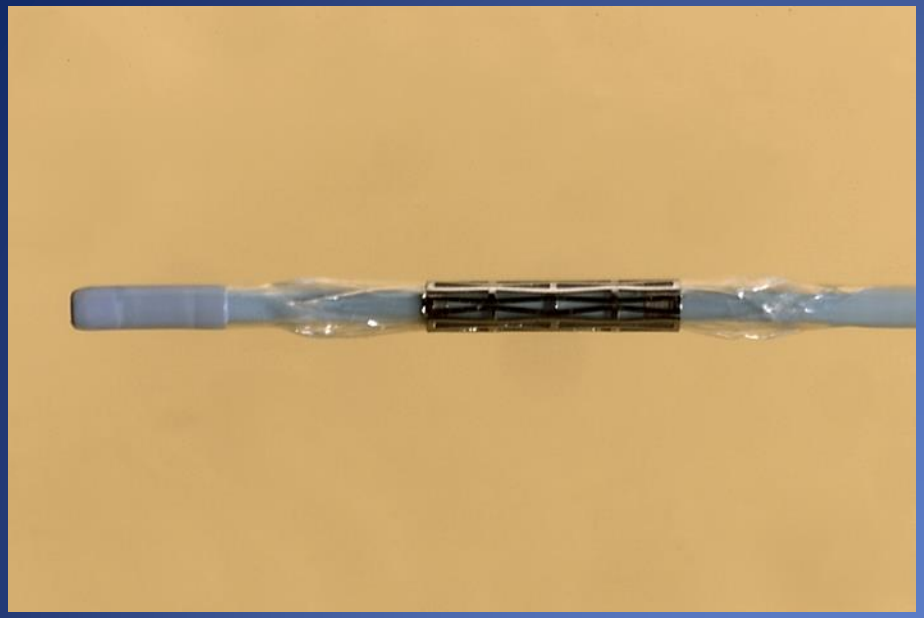

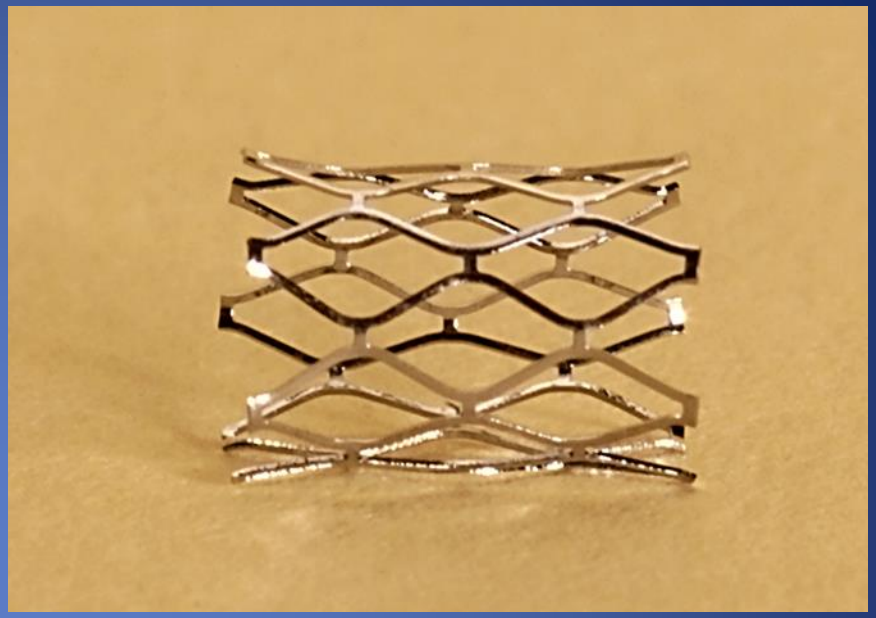

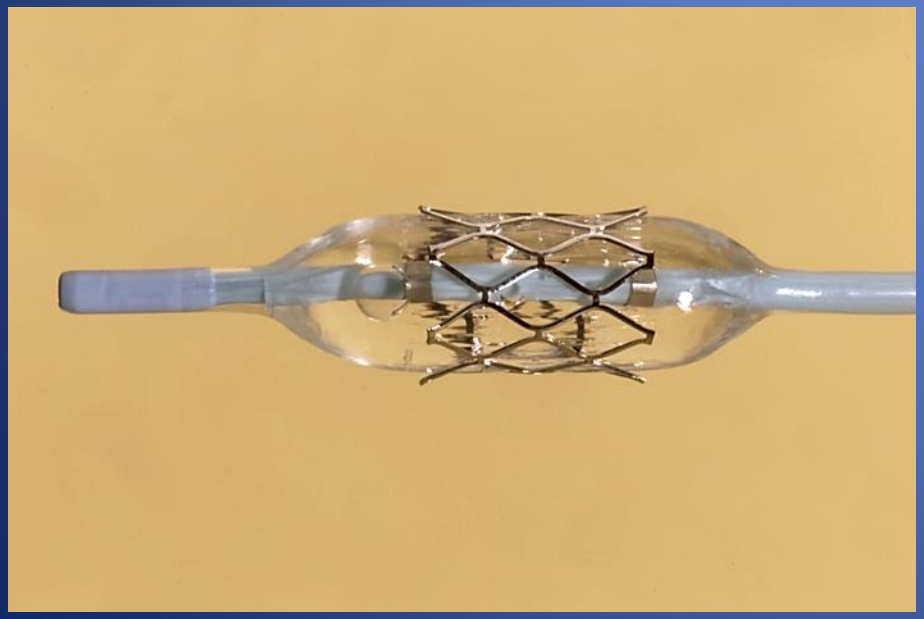

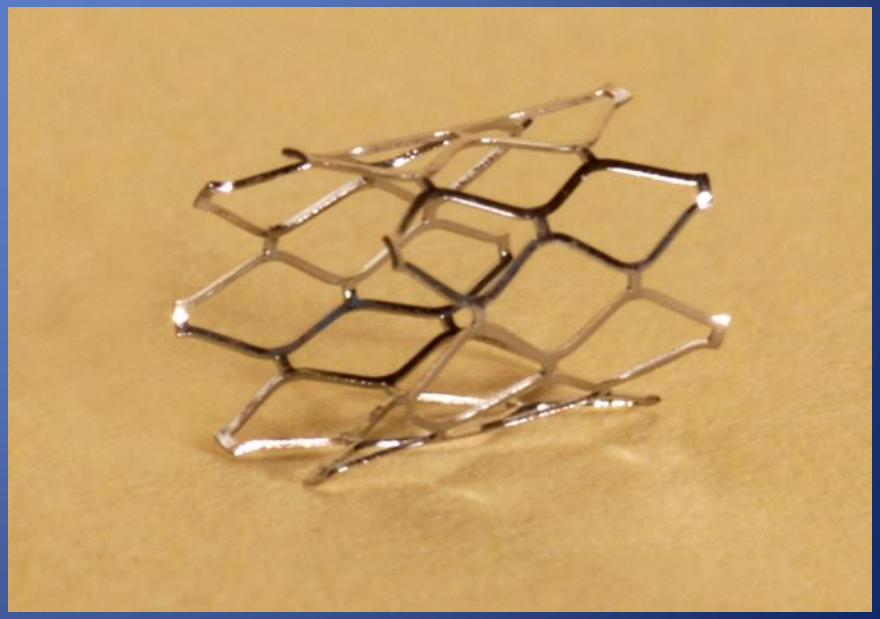

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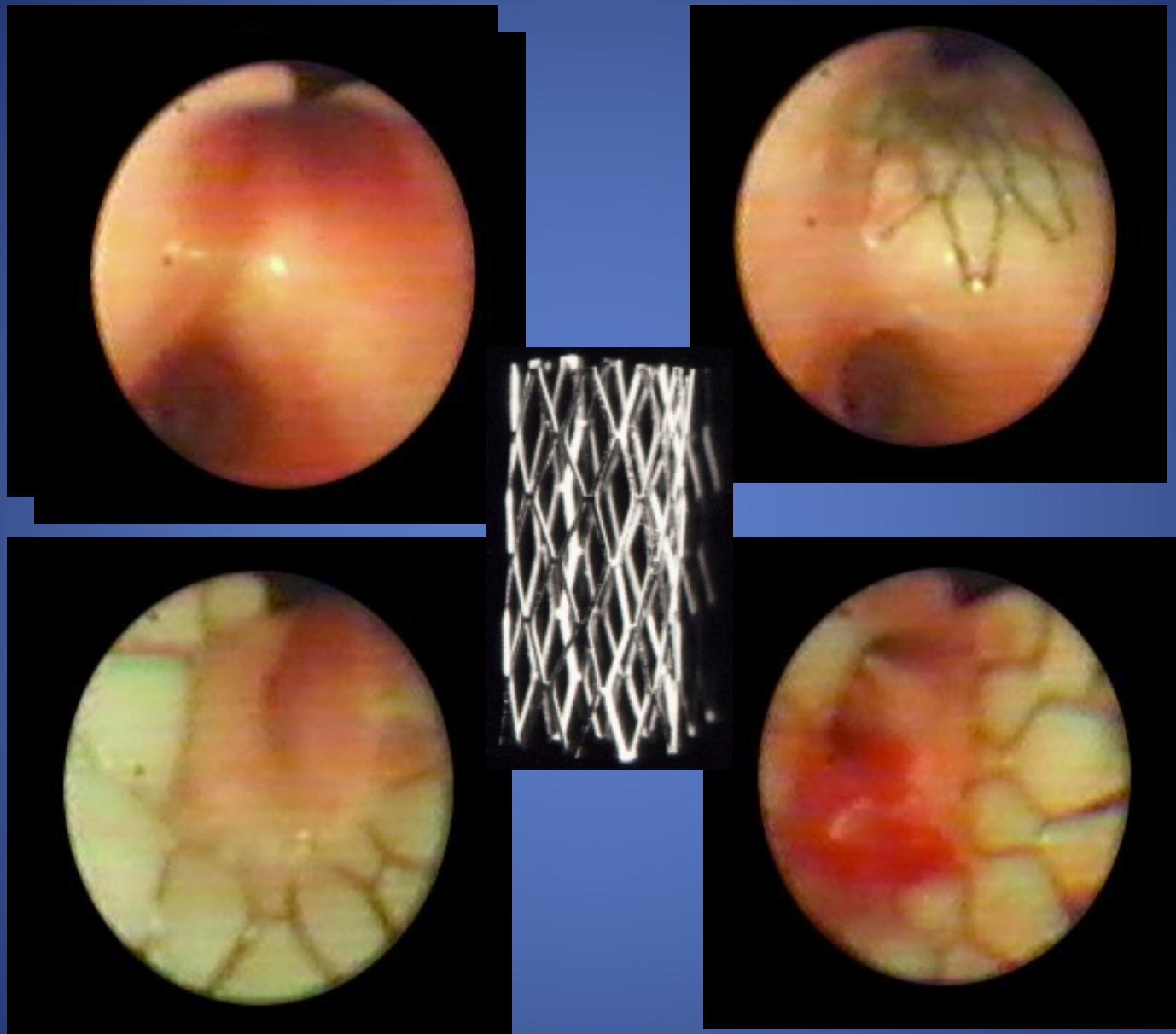

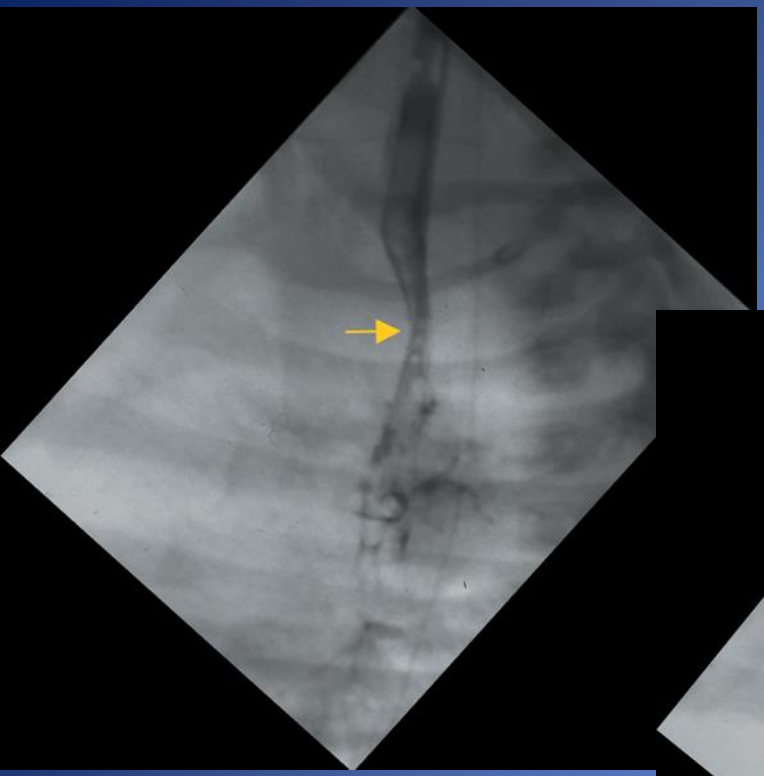

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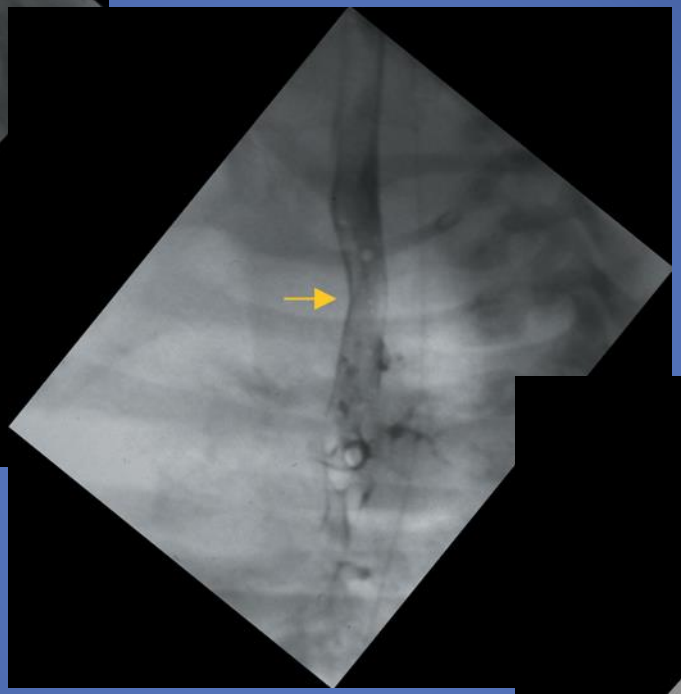

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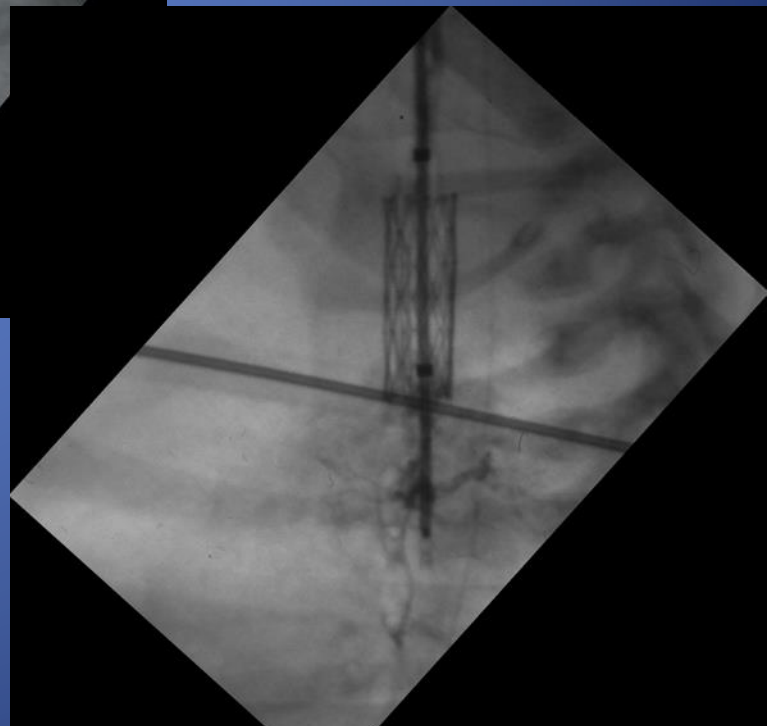
STENT



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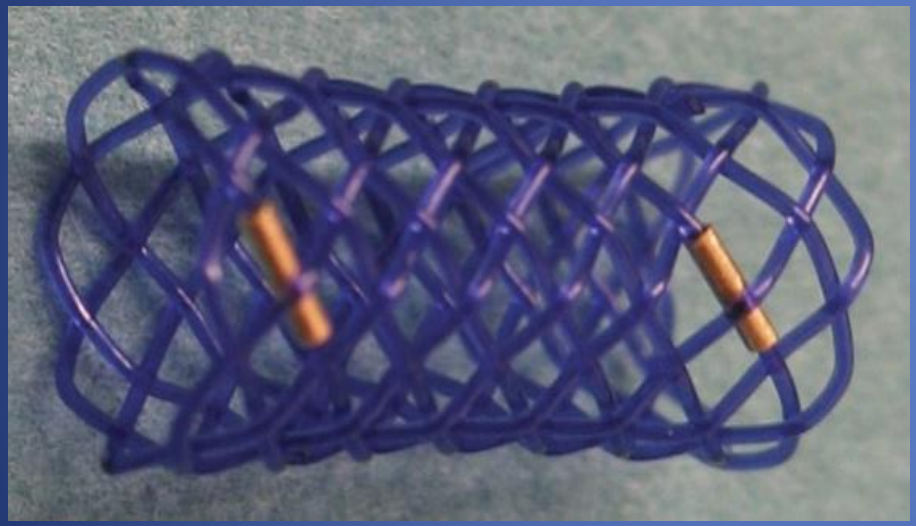
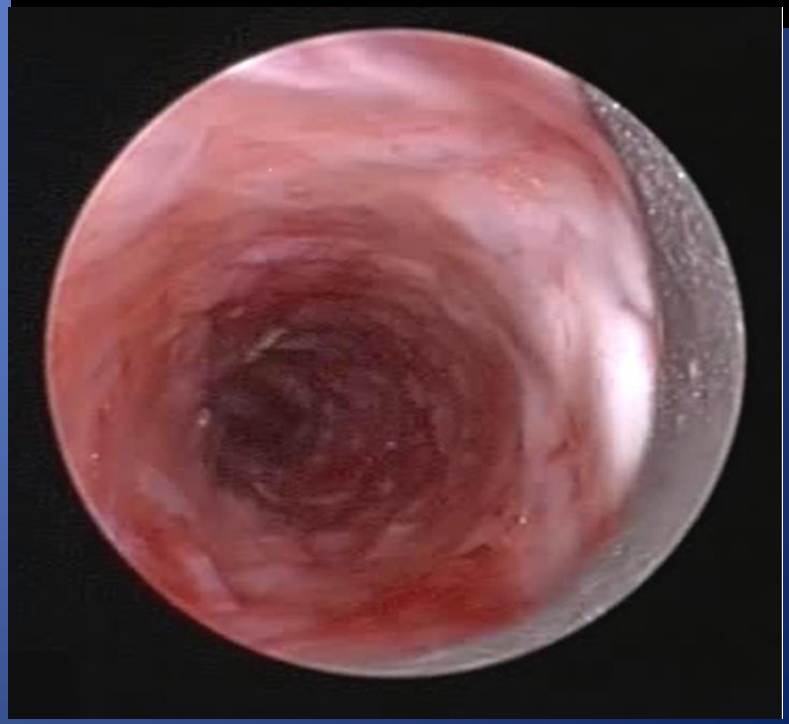
PROBLEMS WITH STENTS

- Granulation formation
- Infection
- Bridge to mucociliary clearance
- Migrate
- Won't come out!
- Don't grow with the child
- Obstruct the lumen
- Erode into the wall
- Erode into surrounding structures





Wallis C, McLaren CA.
Tracheobronchial stenting for airway malacia.
Paediatr Respir Rev 2018:

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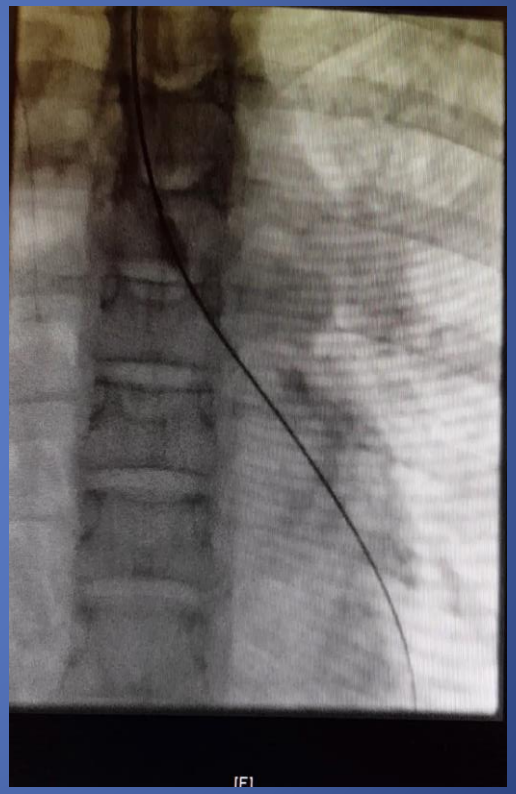

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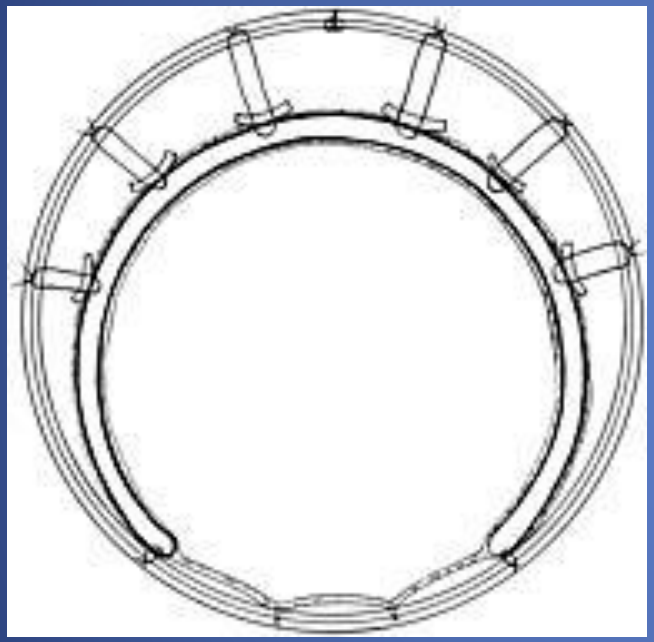
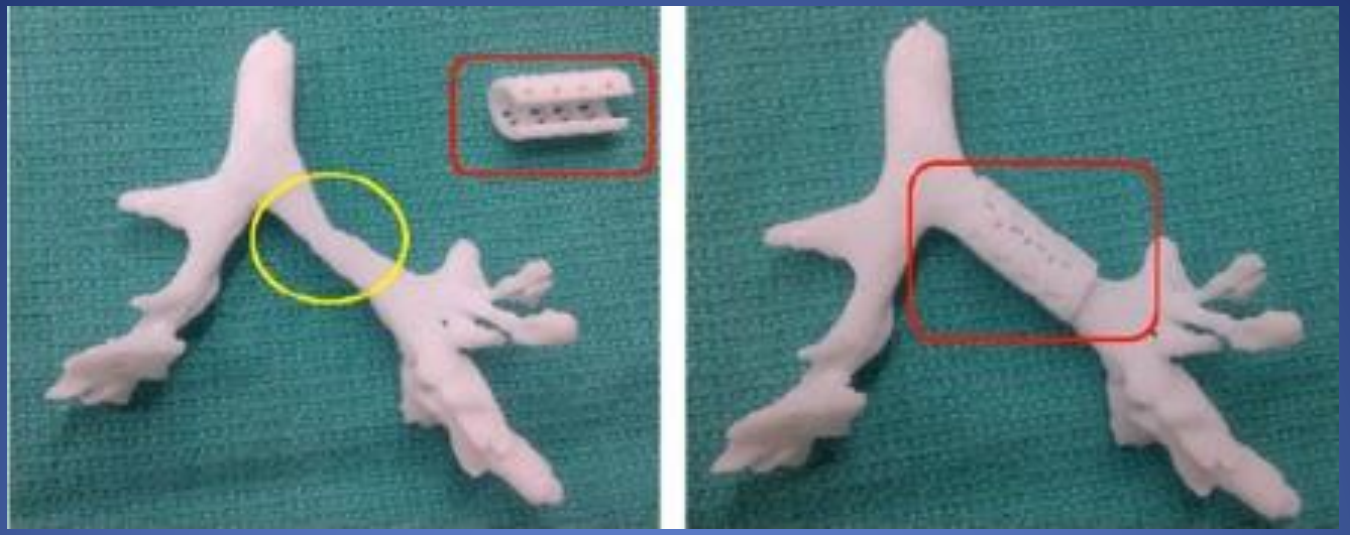
Surgical Mx of Tracheomalacia: External Hagl stent

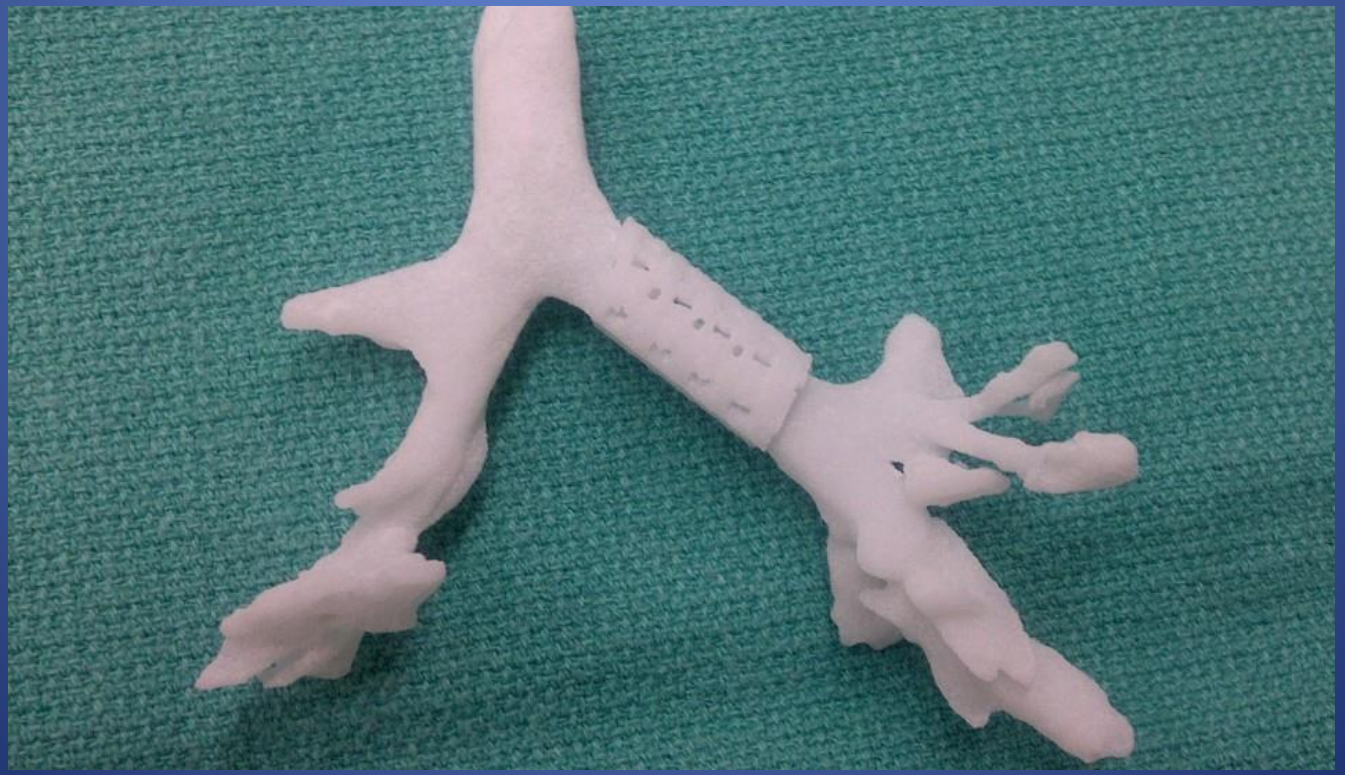


- **7 children**
- **extubated 1 - 12 days**
- **1 unrelated late death**
- **excellent at F/U (21 - 54 m)**

GOSH
7 patients, no deaths,
1 oesophageal perforation
1 airway penetration

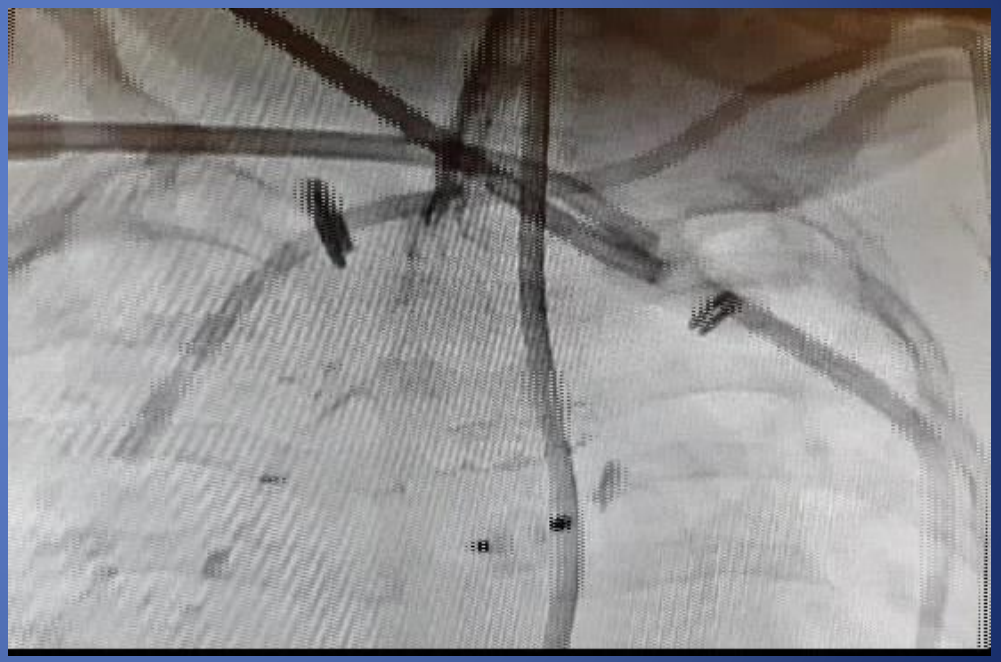
Hagl S et al *Ann Thorac Surg 1997;64:1412--21*





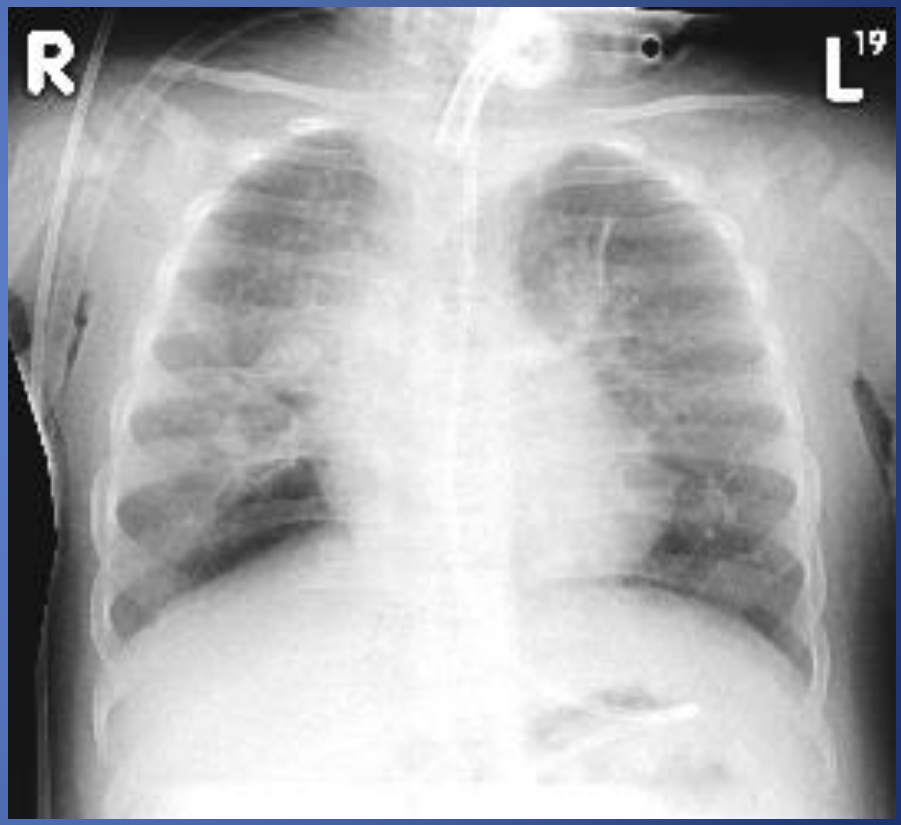
Long term positive pressure support

- Premature 32 weeks
- Complex congenital heart disease
- Neonatal cardiac repair
- Stuck on ventilator
- Unable to extubate
- High PEEP requirements



Long term positive pressure support

- Prem; post cardiac surgery
- Born at 32 weeks
- Complex congenital heart disease
- Neonatal surgery
- Stuck on ventilator
- Underwent tracheostomy and positive pressure
- Home and thriving
- Slowly weaning off support



Outcomes for Tracheostomy LTV in BPD

- 10 children with BPD and TBM
- 1 died at age 9 months
- 8 discharged home with 24 hour CPAP via trache
- 5 children weaned off ventilation by age 2 –4 years
- 1 weaned at age 7 years
- 2 children still under 2 years but weaning steadily

Summary

- Malacia is a common respiratory problem
- It presents with a wide range symptoms and signs
- Diagnostic techniques are improving
- Treatments are personalised with no perfect solution
- The long term outcome for any of the interventions is unknown with scant knowledge on natural history

THE END