Congenital Cystic Pulmonary Airway Malformations

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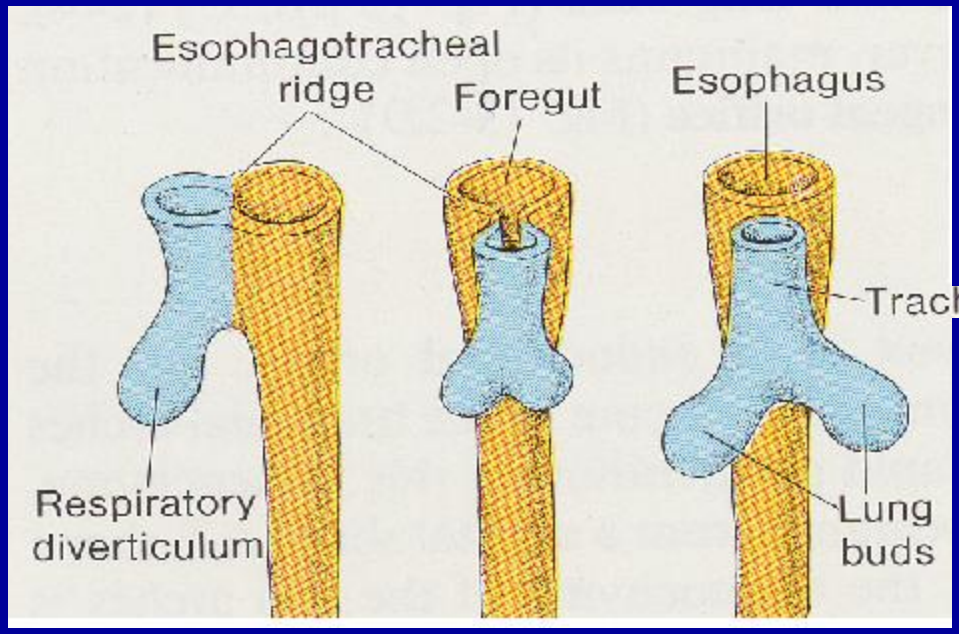
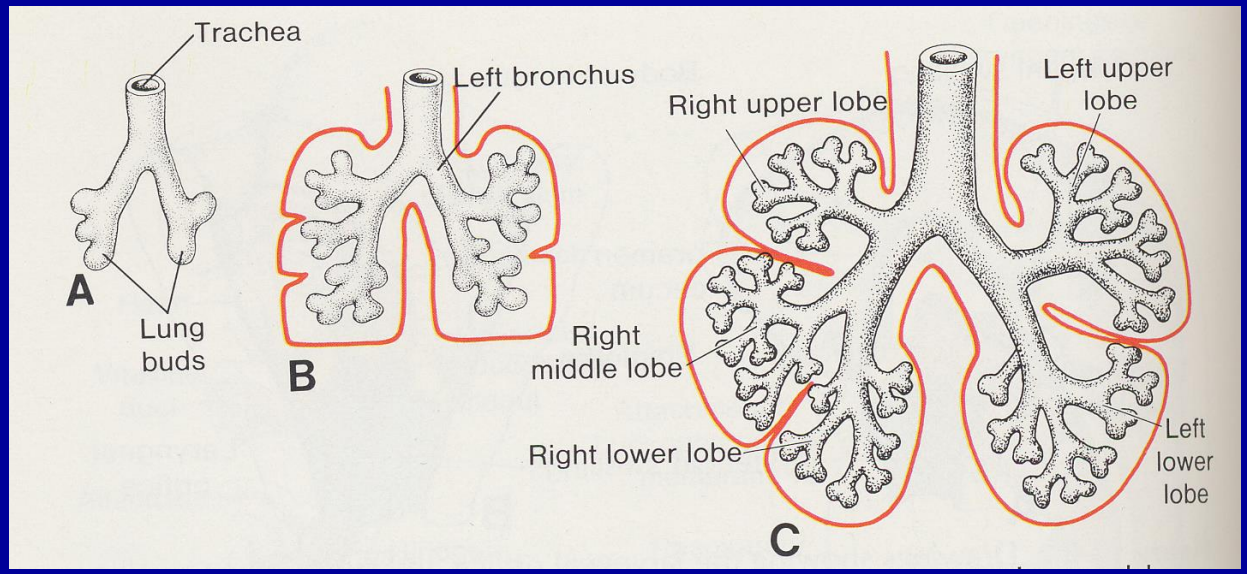
Embryology of the Respiratory Tract





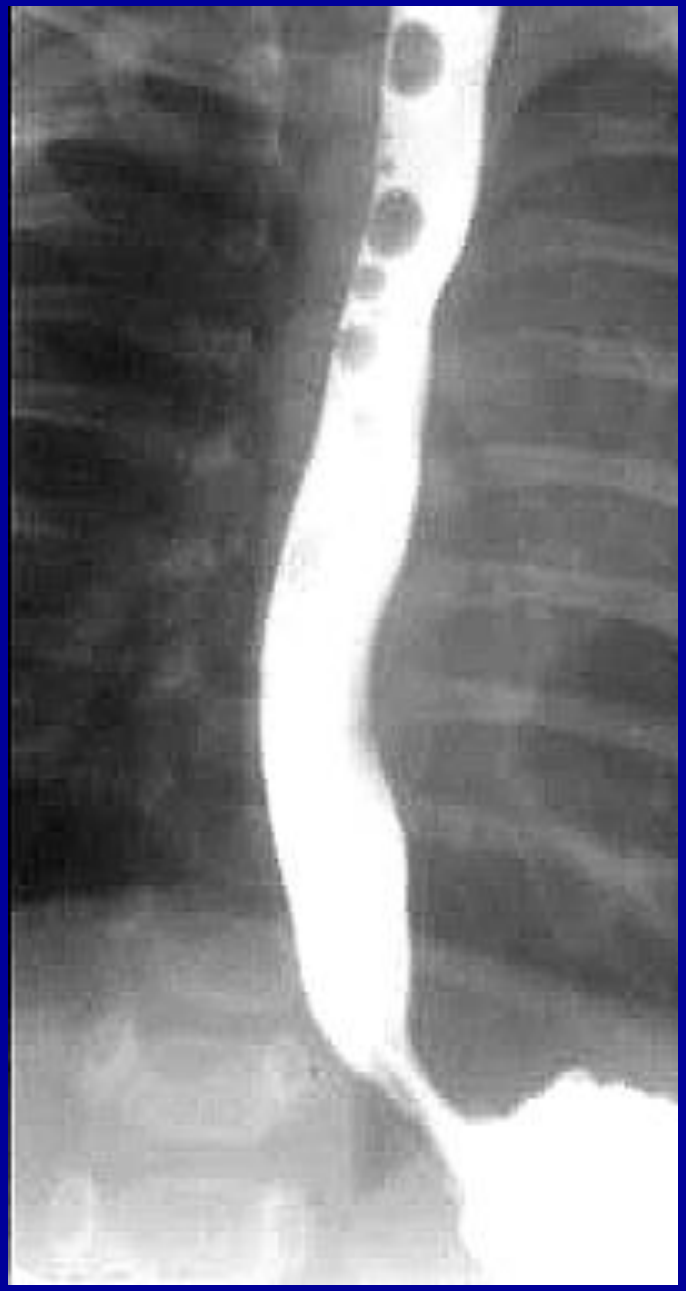
Anatomical list of congenital lung abnormalities

- cleft larynx/trachea
- tracheo-oesophageal fistula
- tracheal stenosis/malacia
- congenital cystic abnormalities
- hypoplasia/aplasia
- abnormal vascular connections/scimitar syndrome
- congenital diaphragmatic hernia

Post natal Presentation of 55 Cystic Lung Lesions from 1985 - 1995

(before the advent of widely available antenatal scanning)

18	Infection	- acute	10
		- recurrent	8
17	Compression / expansion effect		
11	Pneumothorax		
1	Malignancy		
1	Chest wall deformity		
7	Asymptomatic / incidental		



Antenatal diagnosis of congenital lung lesions

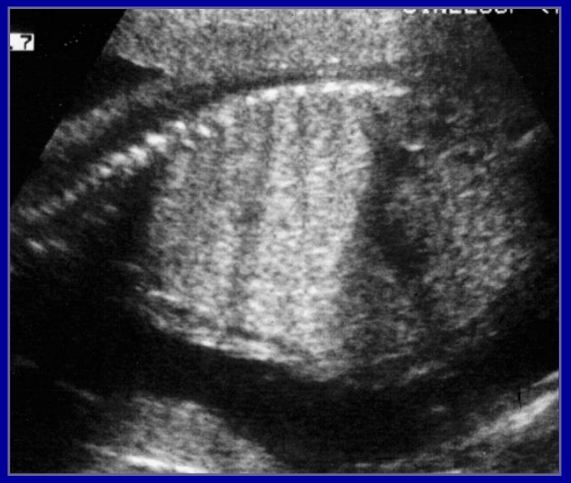
2019

Now most of the lesions
we will see are
asymptomatic

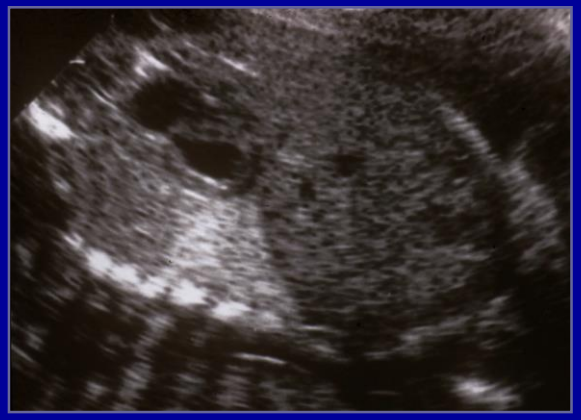
Antenatally diagnosed cystic adenomatoid malformation

- Unpredictable growth between 18-26 weeks.
- Growth plateau at 28 weeks gestation.
- After 28 weeks, regression may occur (15-30%) even disappear.
- Majority asymptomatic throughout pregnancy.
- Occasionally symptomatic

21 weeks



32 weeks



The big question for paediatricians: What happens to CPAMs after birth?

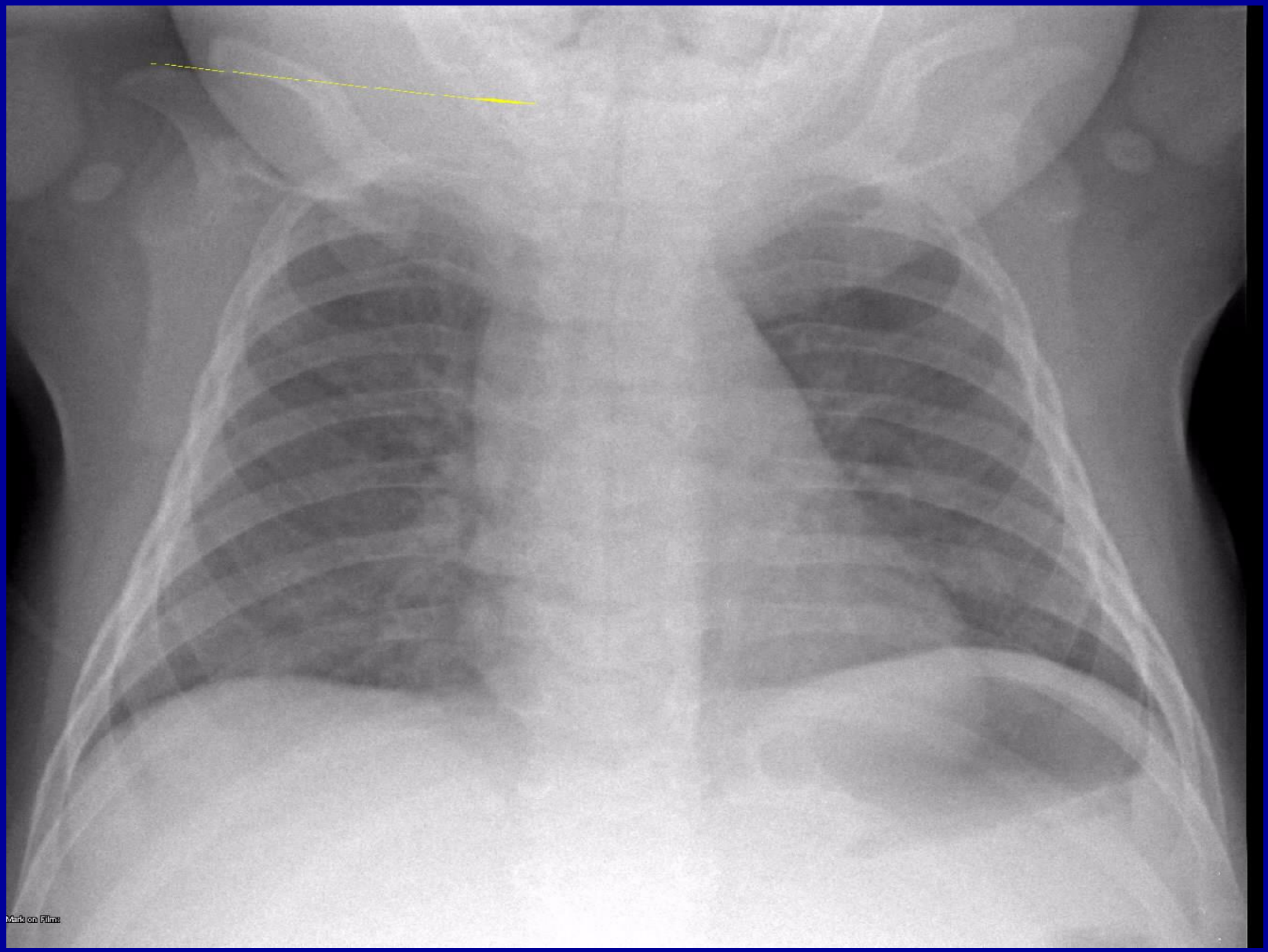
- Nothing
- Shrink
- Disappear
- Turn out to be something else
- Enlarge/shift/pneumo
- Become infected
- Malignant change

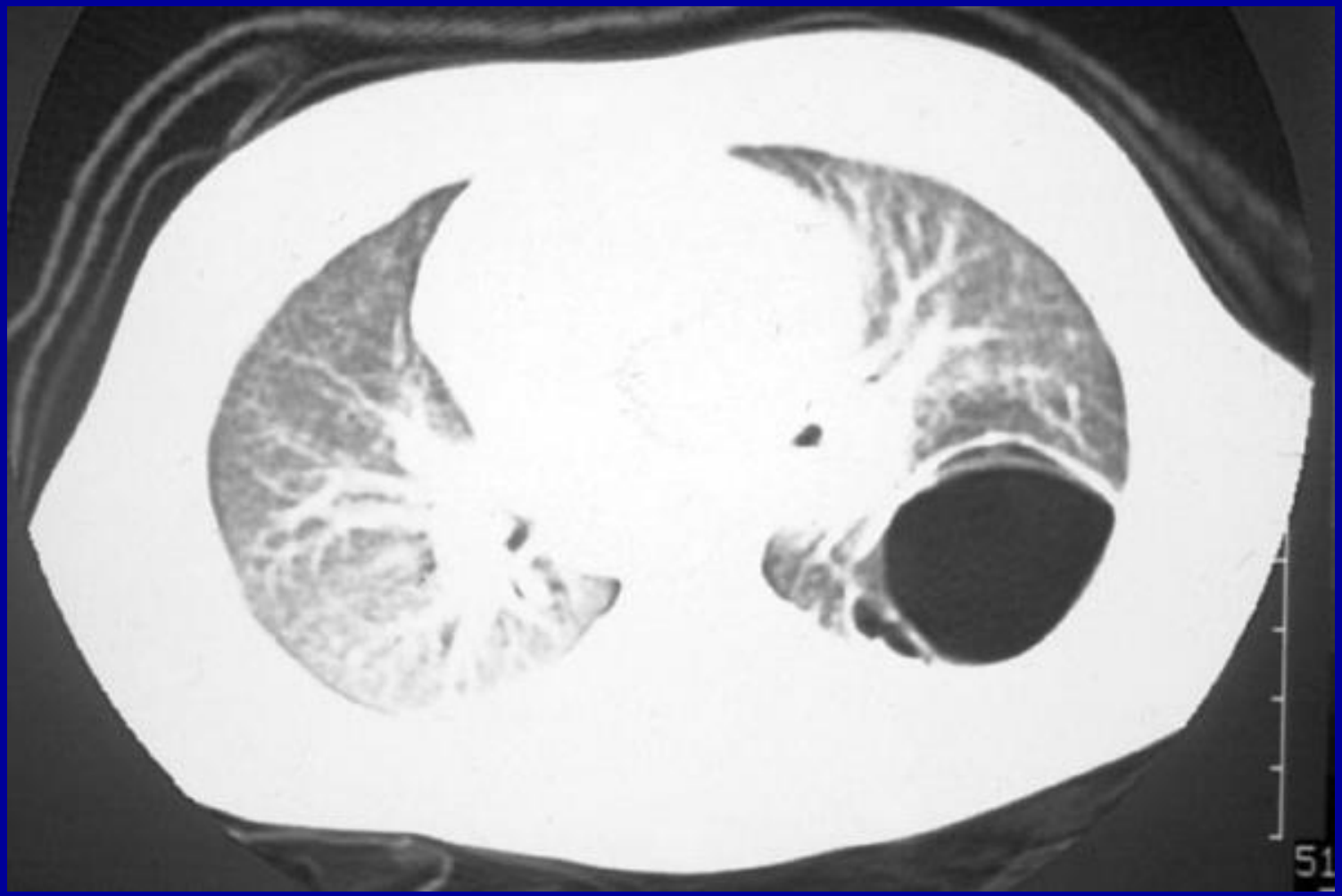
SURGERY

What are the chances?

What are the risks?

Post natal CXR: “ no evidence of ANTENATAL CCAM “

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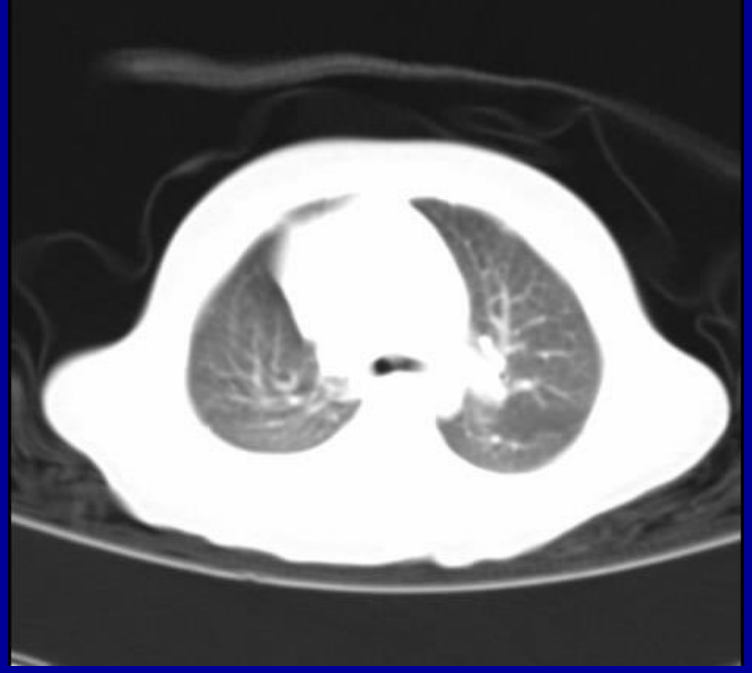
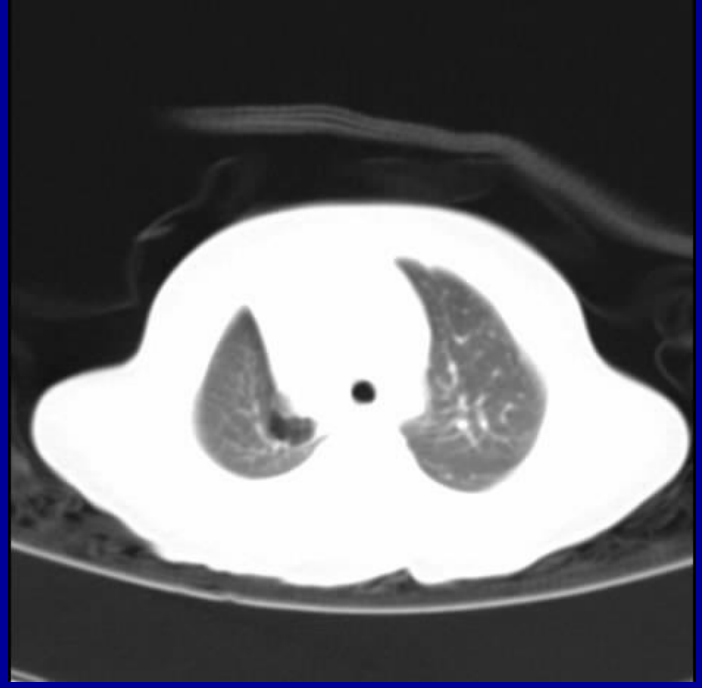
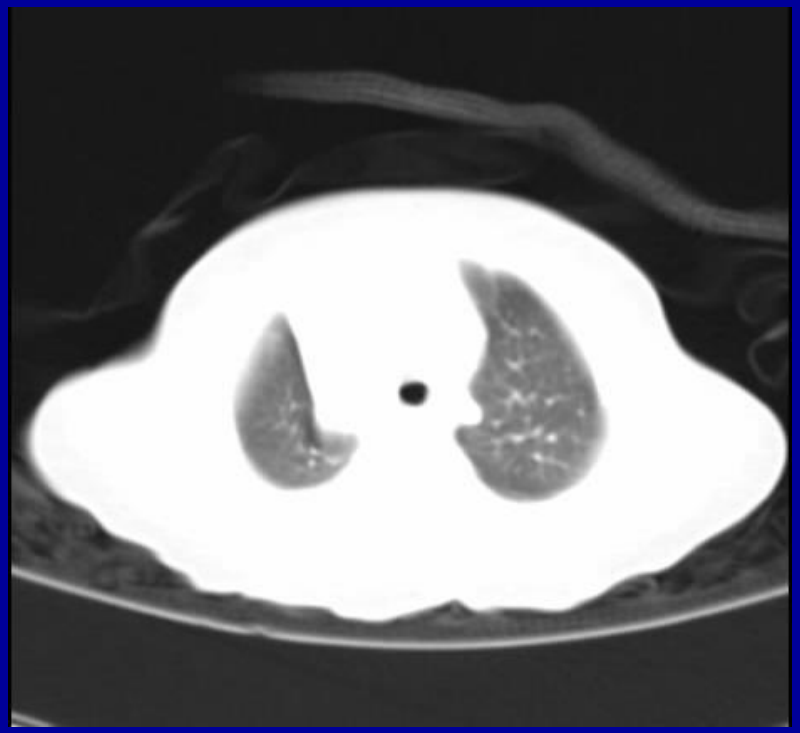
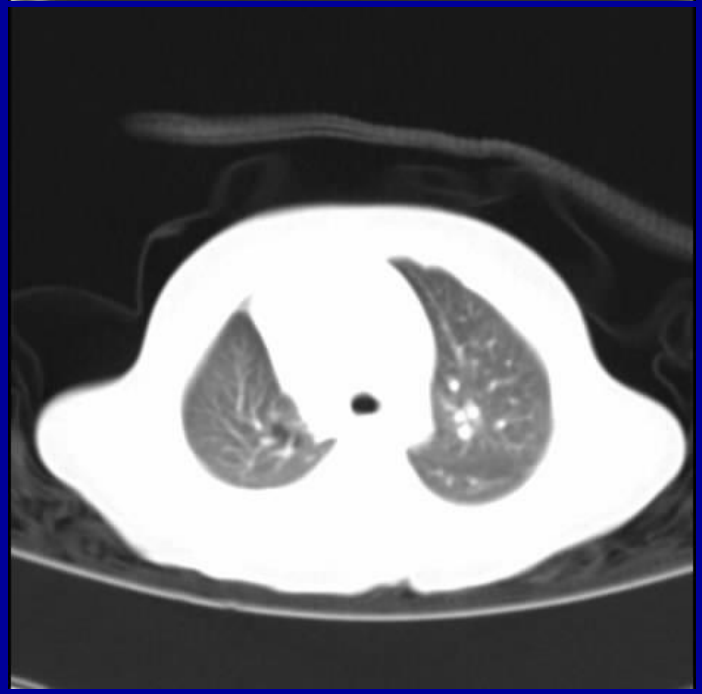

The big question for paediatric pulmonologists: What happens to CPAMs after birth?

- nothing
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- en[illegible]ift
- become infected
- malignant change

SURGERY

What are the chances?

What are the risks?

Congenital Cystic Lung Abnormalities diagnosed on antenatal ultrasound

- CPAM
  - 3 types
- Sequestration
- Lobar overinflation
- Bronchial agenesis
- Hybrid lesions
- Single cyst
- Bronchogenic cyst

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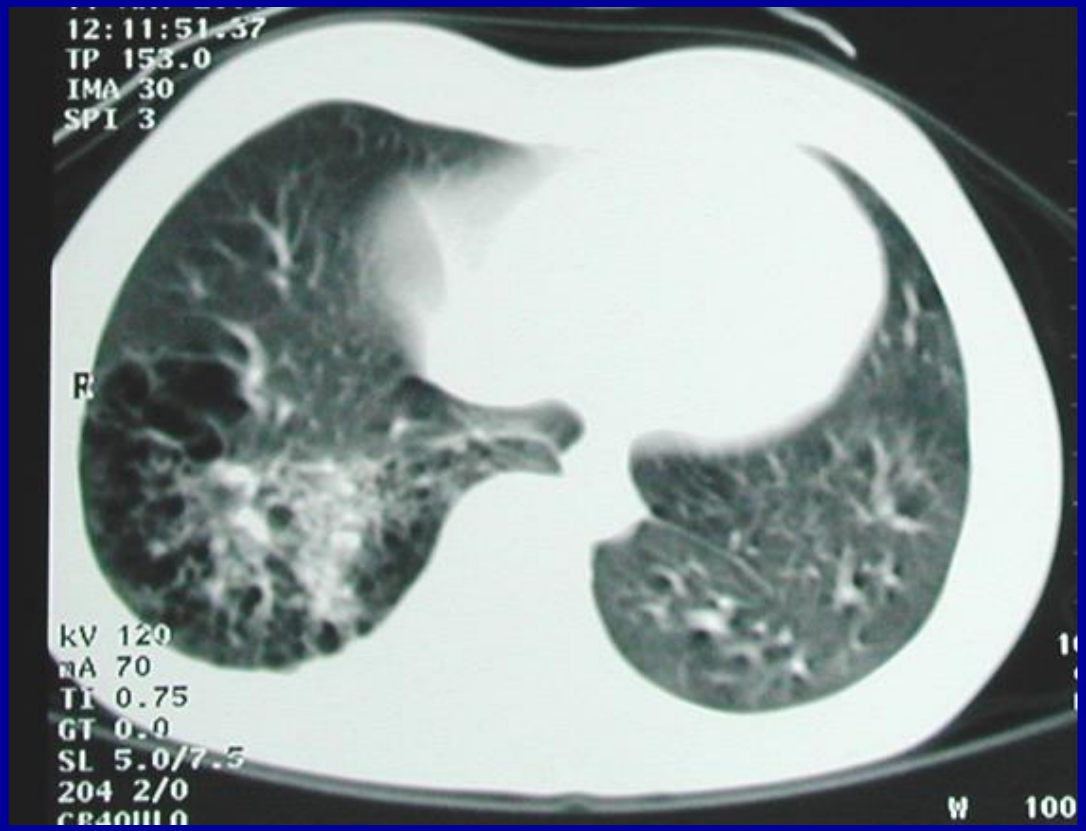
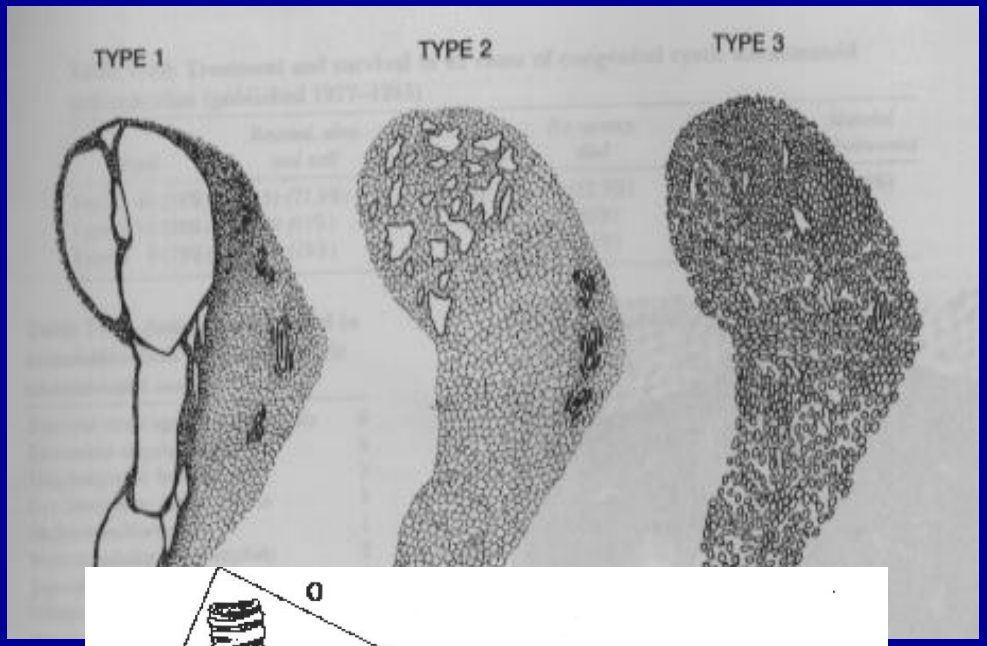
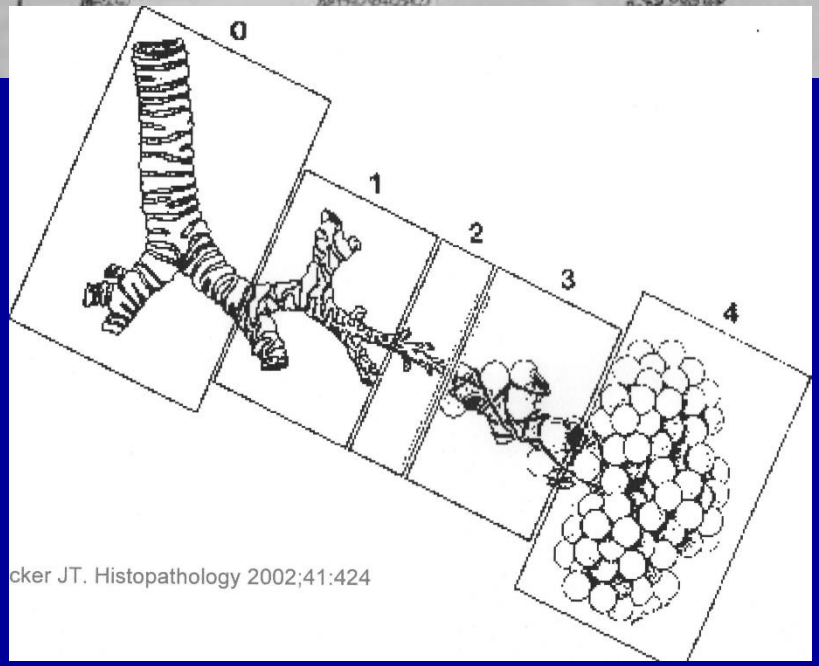


Histology

Based on cyst size

- Type 1: Macrocystic
  Most common-60-70%
  cysts >2cm
- Type 2: Microcystic
  Cysts <2cm
  15-20%
- Type 3: Solid mass
  5-10%
- Type 4: (controversial)
  large cysts
  May be a precursor to PPB





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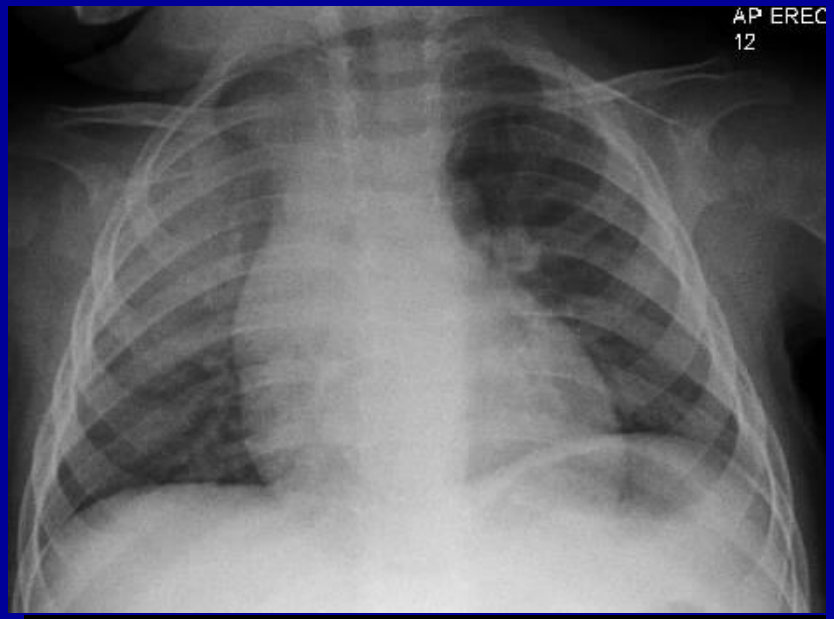
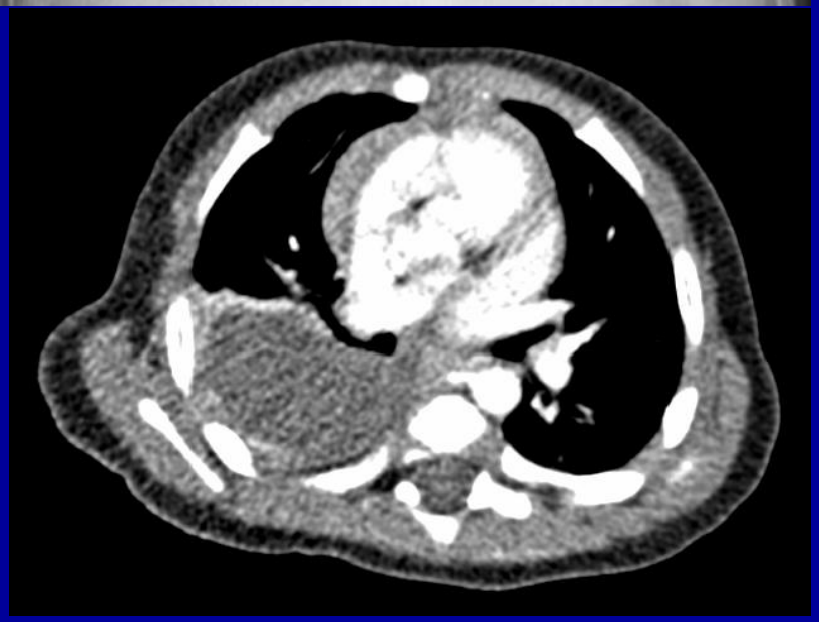
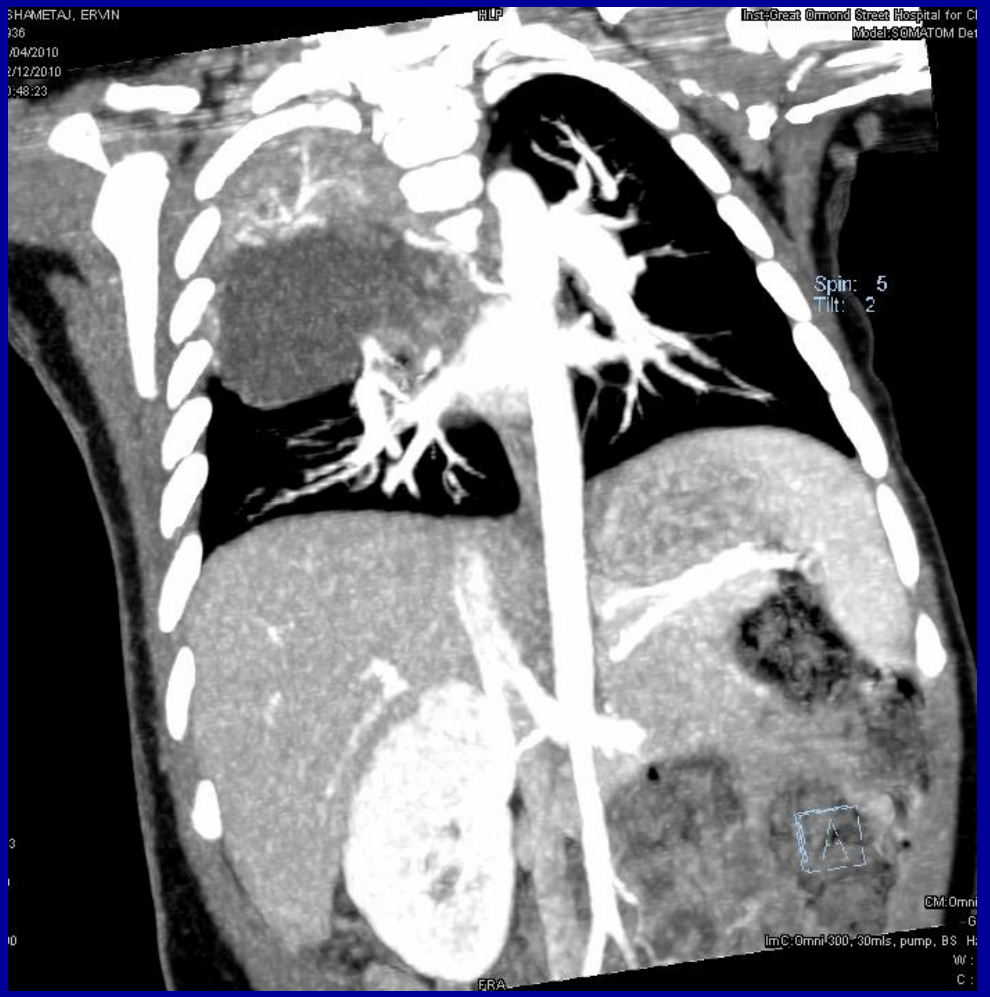
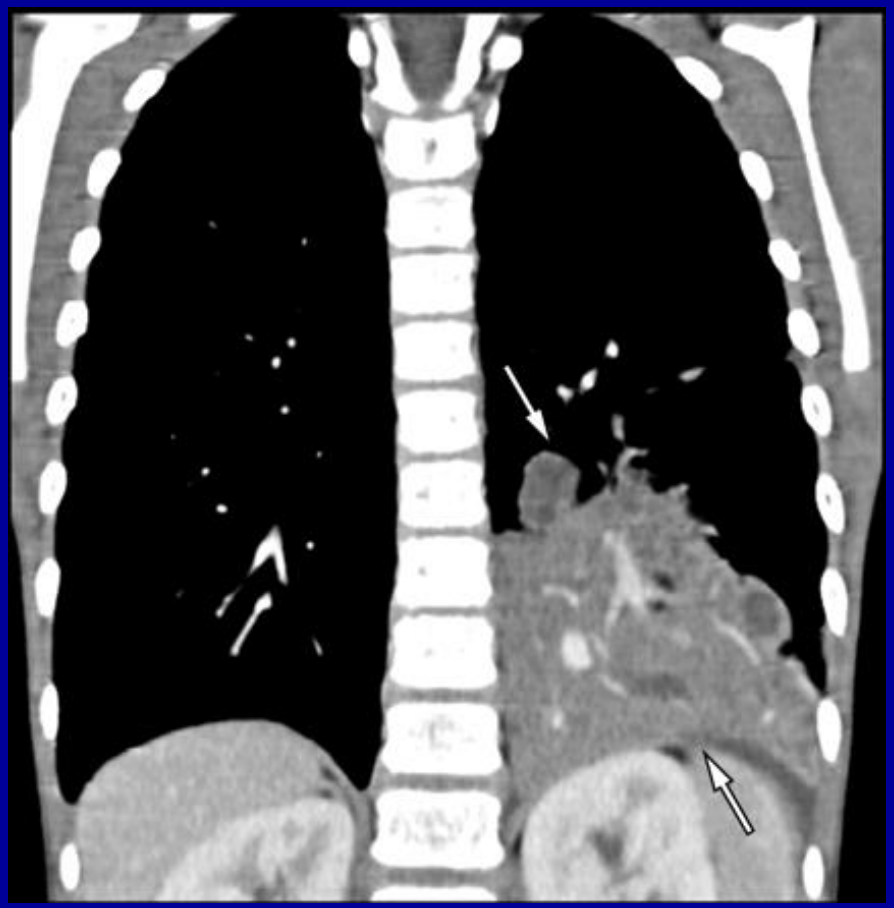
Pure sequestration – intra or extralobar

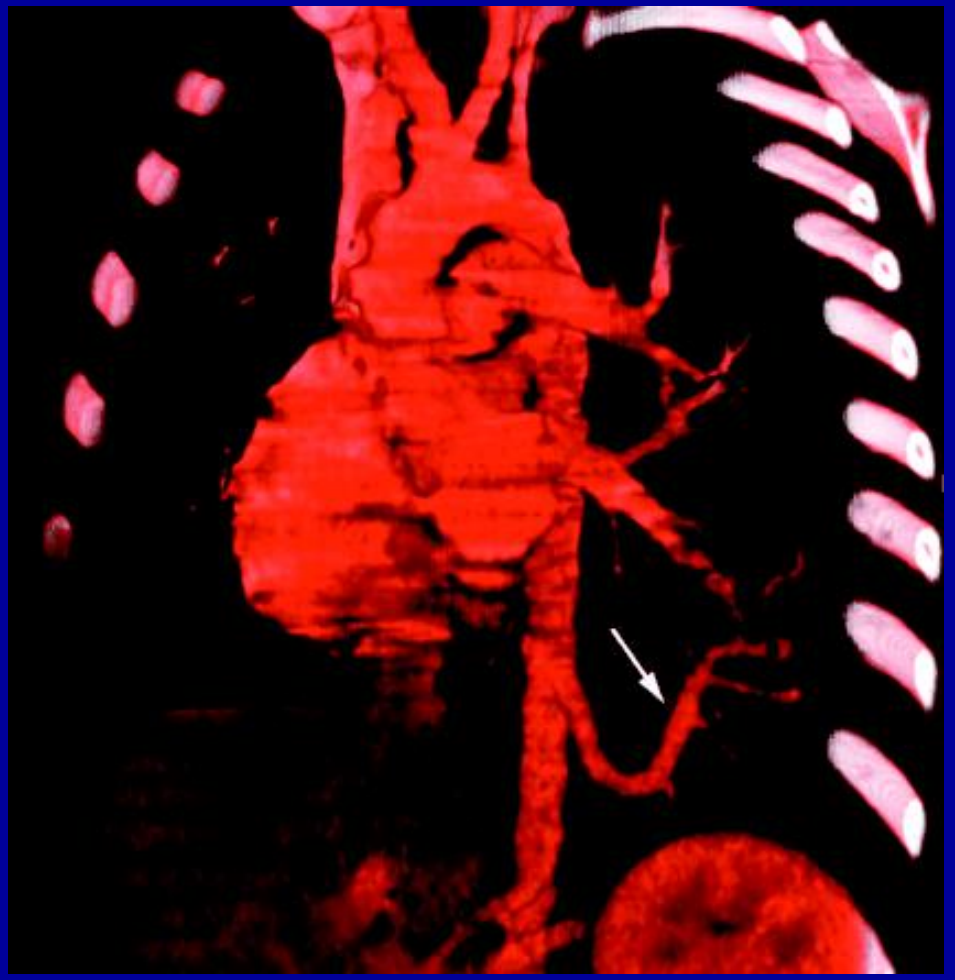






Intra lobar sequestration





Congenital Lung Abnormalities

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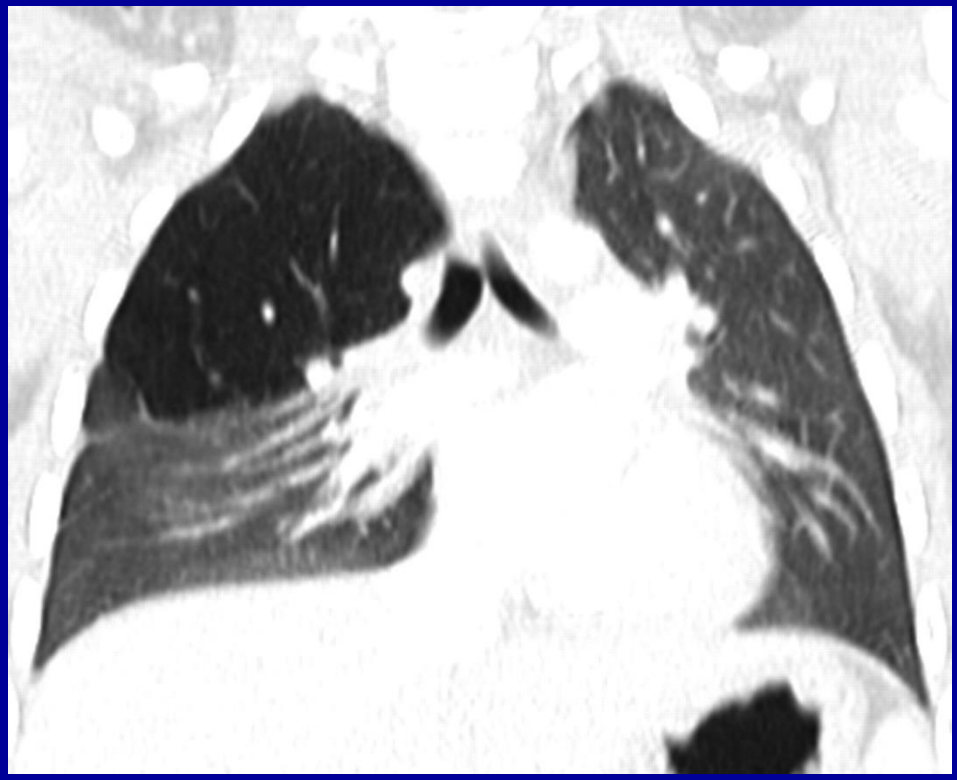
Congenital Lobar Overinflation (Emphysema)

- Overinflation not ‘emphysema
- Over distension of a lobe or lobule following a partial bronchial obstruction.
- Bronchial obstruction only clearly defined in less than a quarter of cases
- ’



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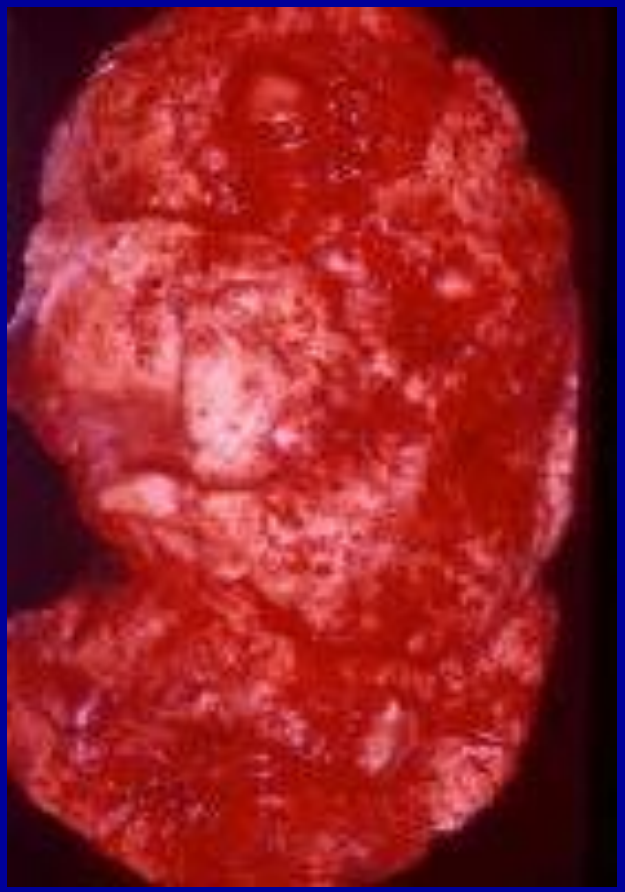

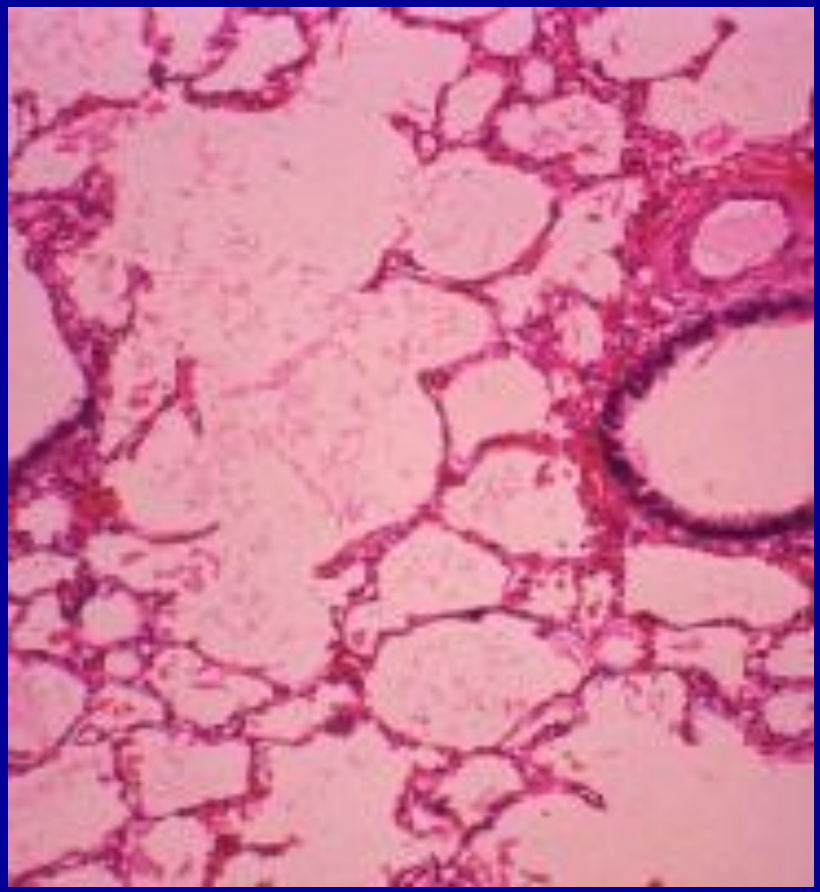

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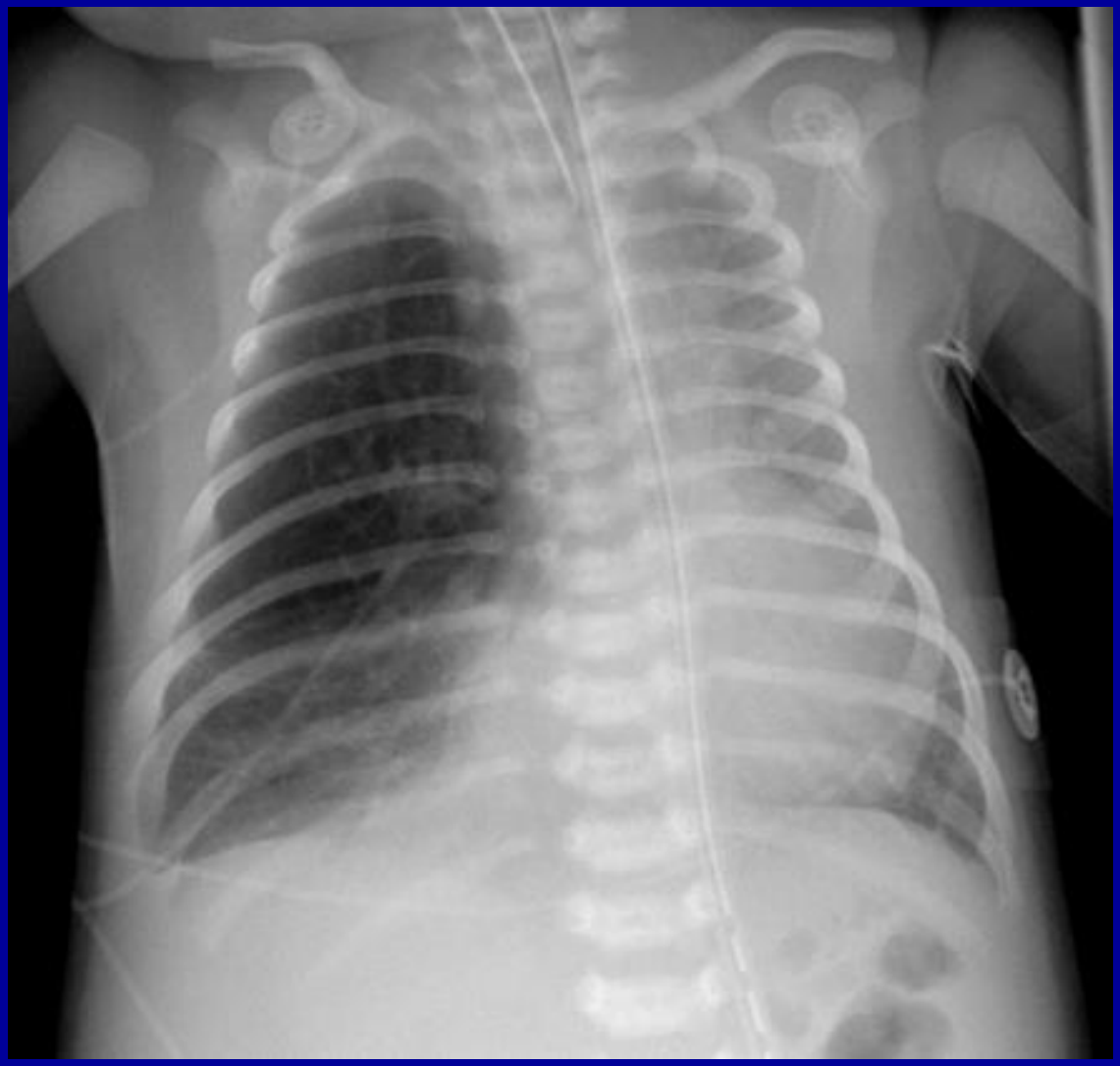

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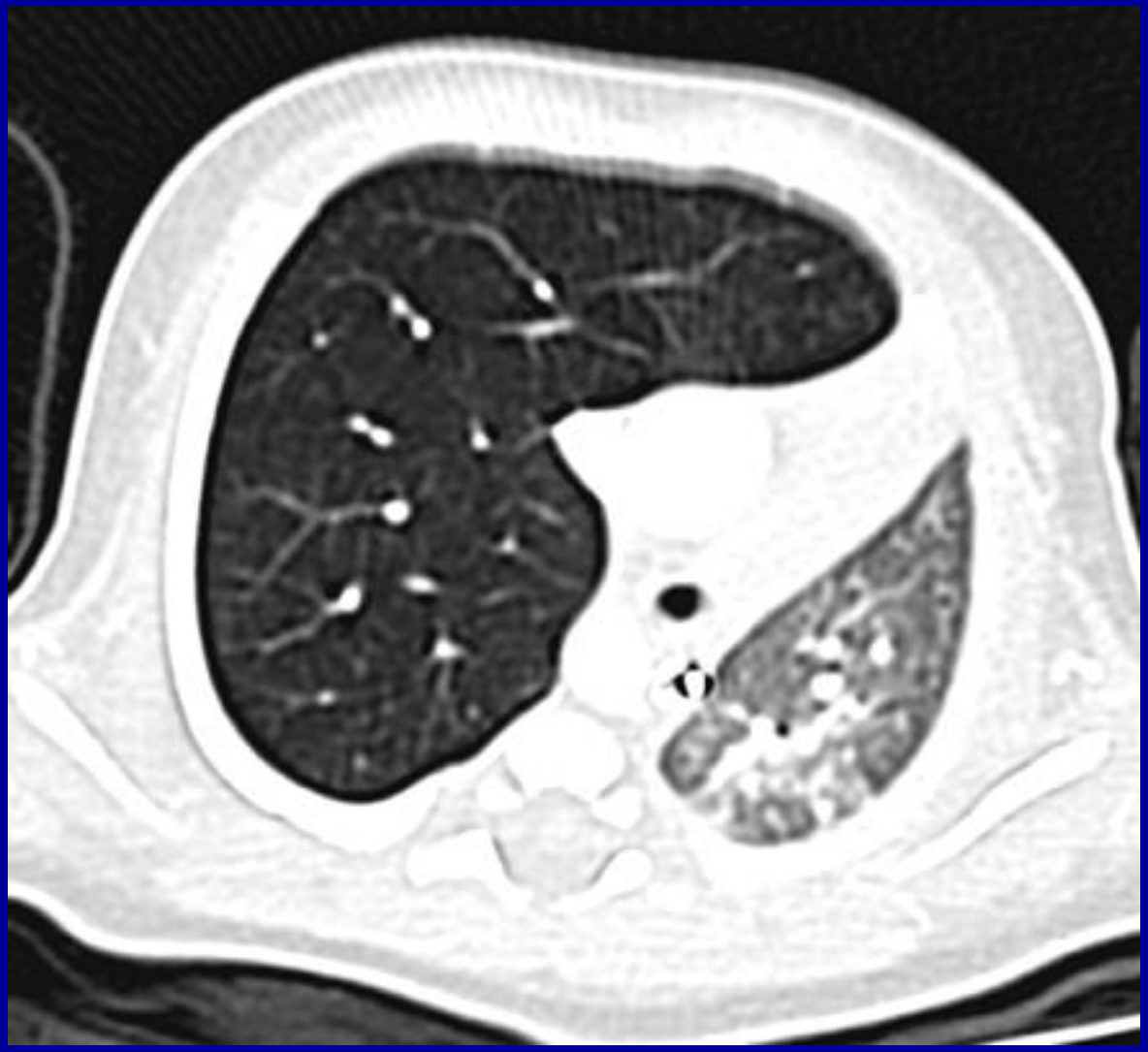

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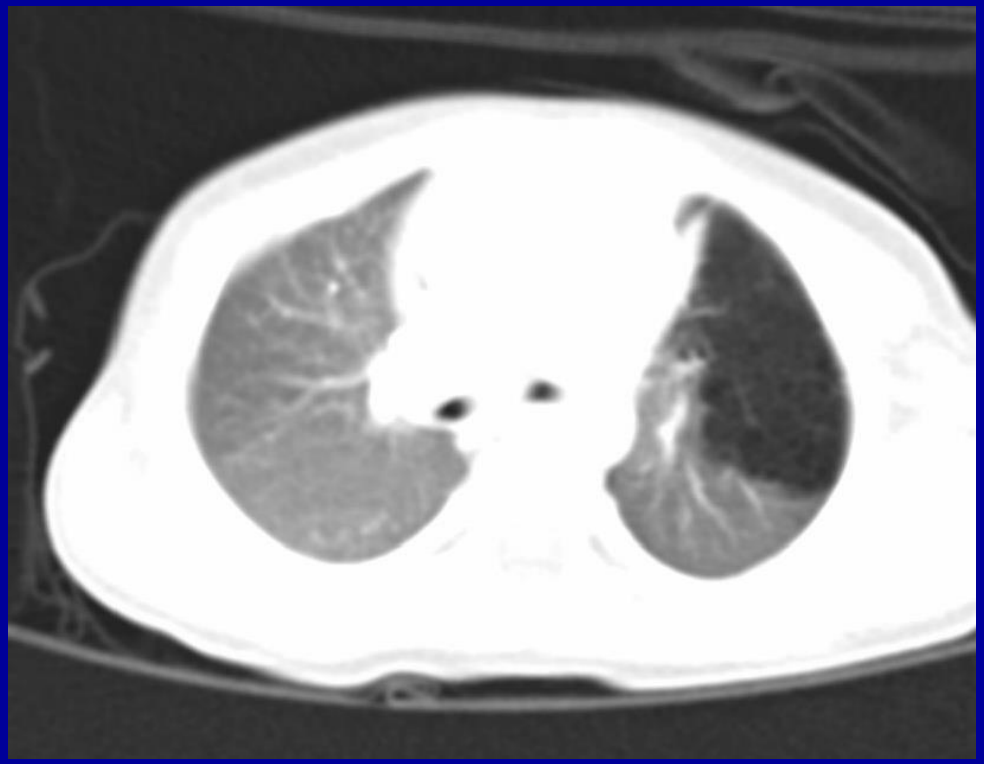

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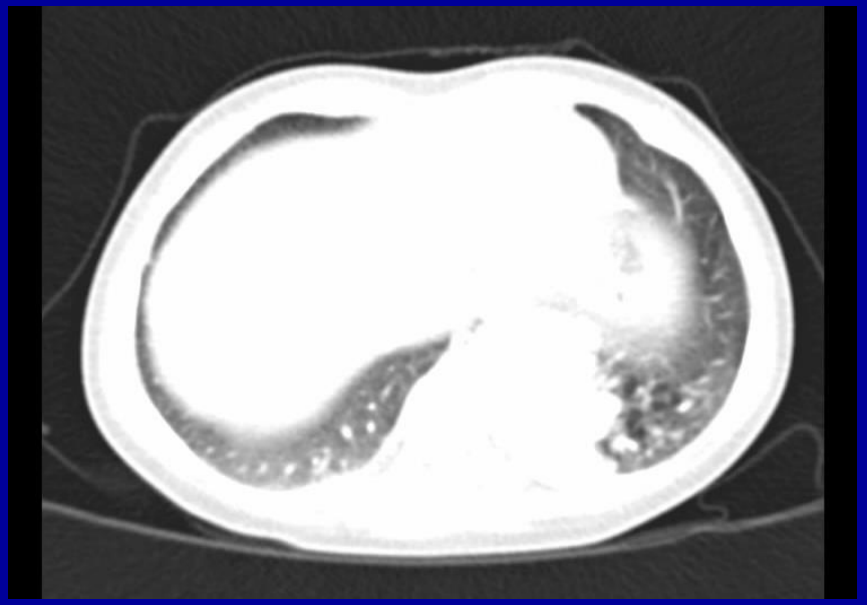
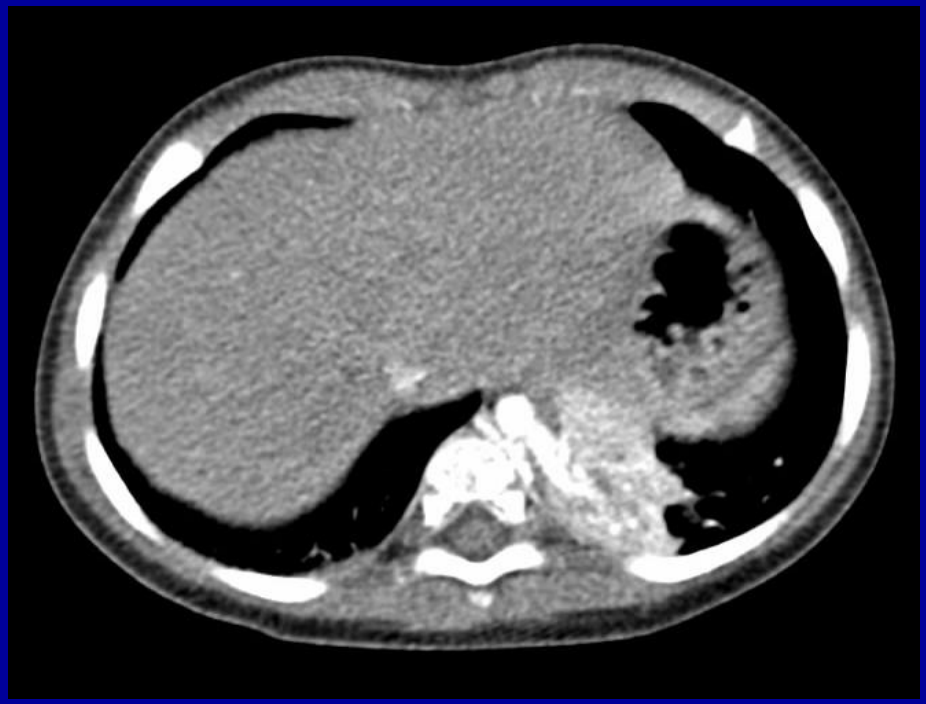
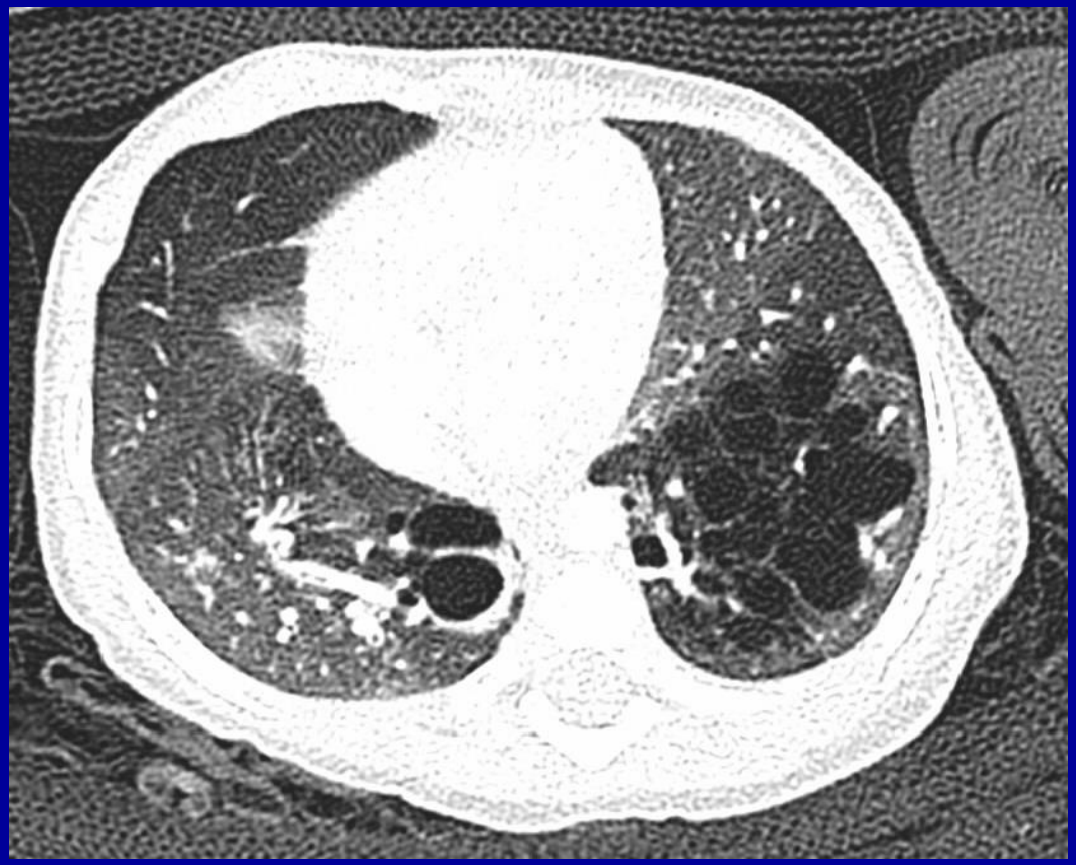
The hybrid lesion - 1

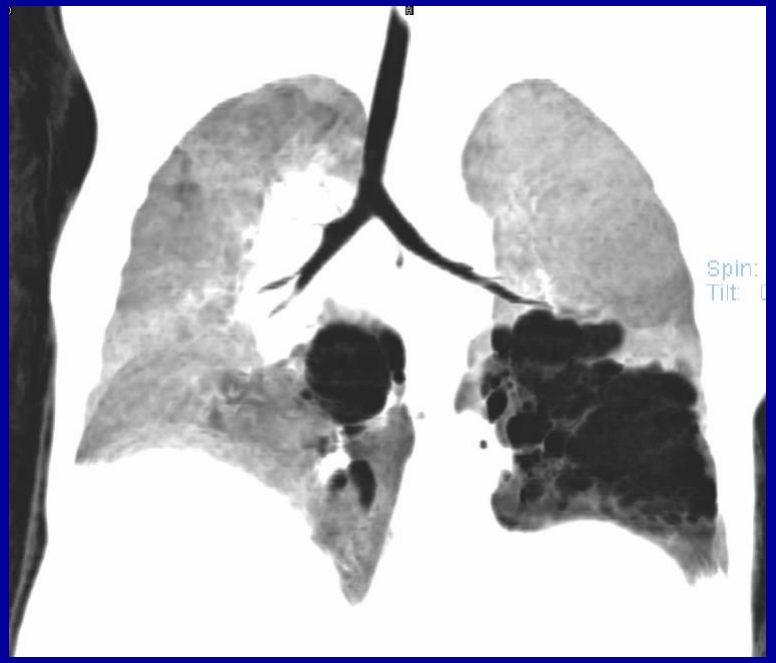
- Routine antenatal scan at 24 weeks
- Dense cystic mass involving the left hemithorax
- Delivered at term
- Healthy and thriving
- CT scan done at 3 months

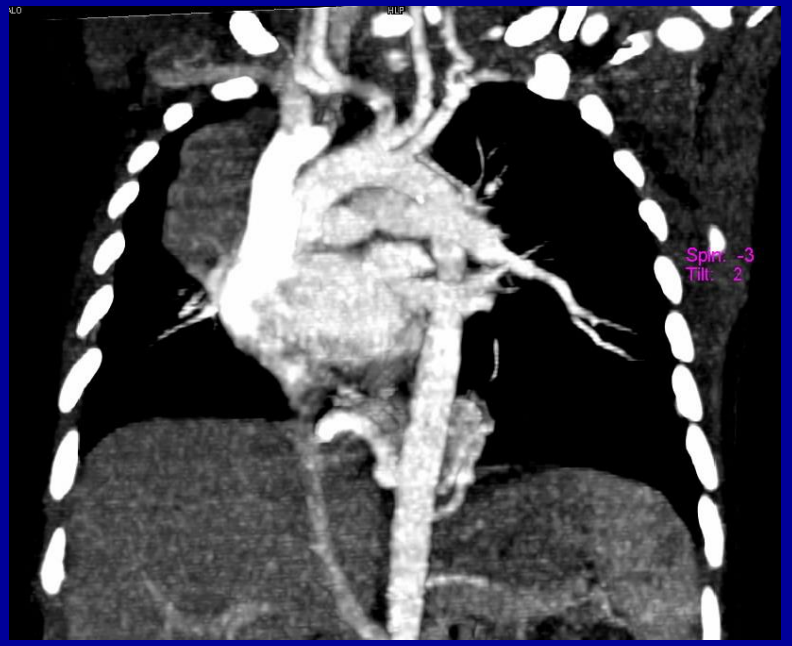




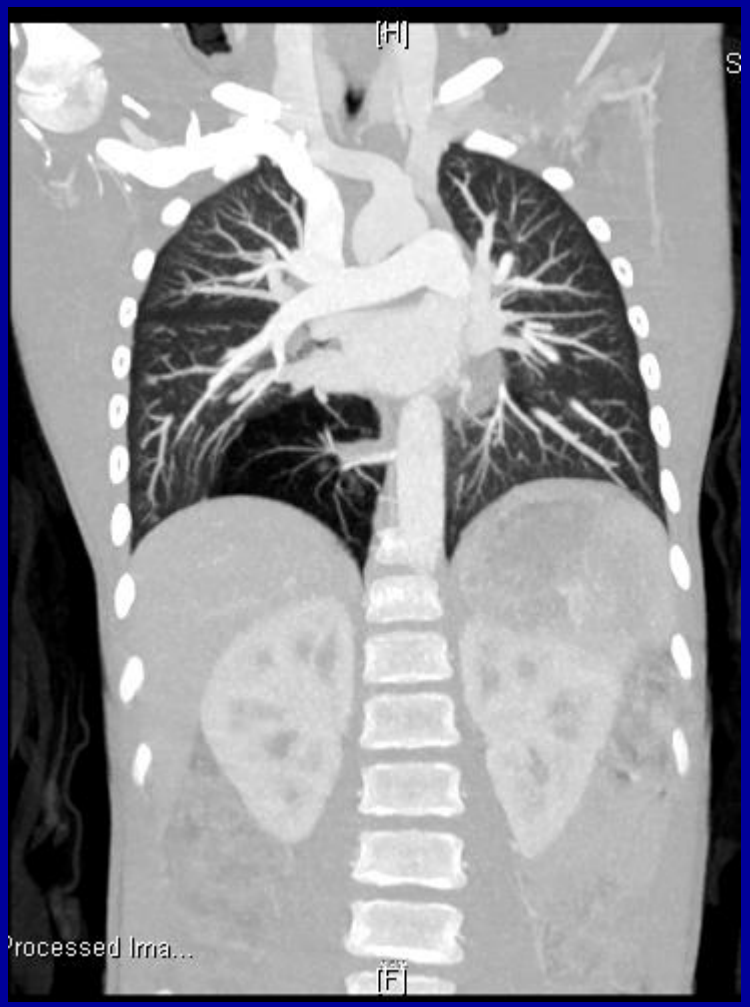
Hybrid lesions -2

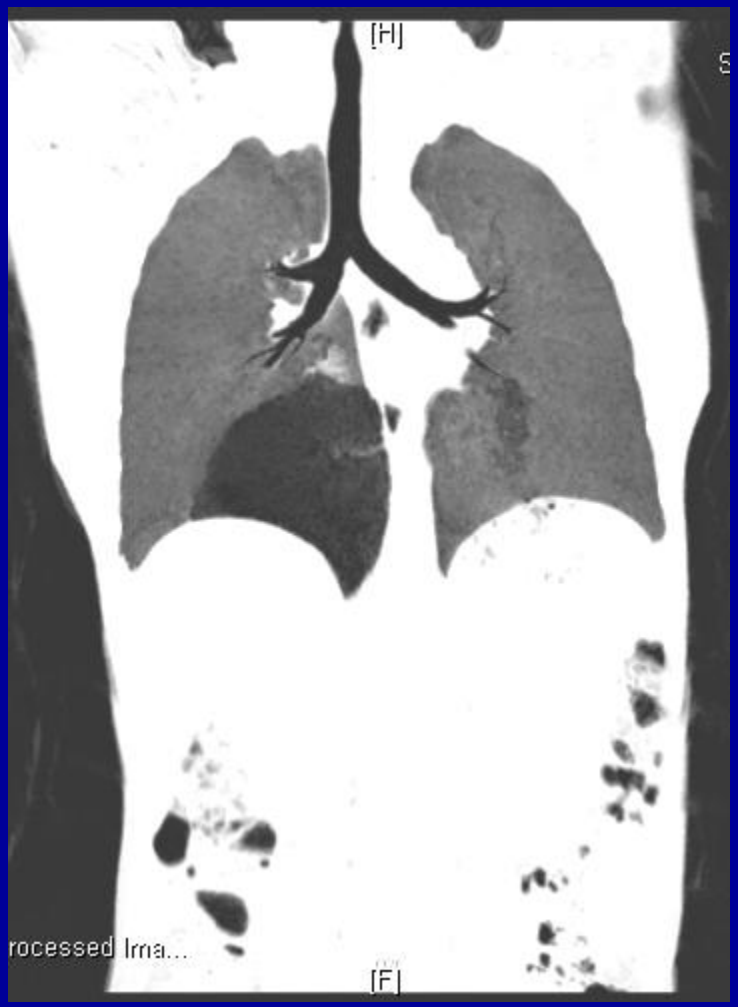






Hybrid lesion – 3: sequestration / bronchial atresia





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Study Time:11:47:02

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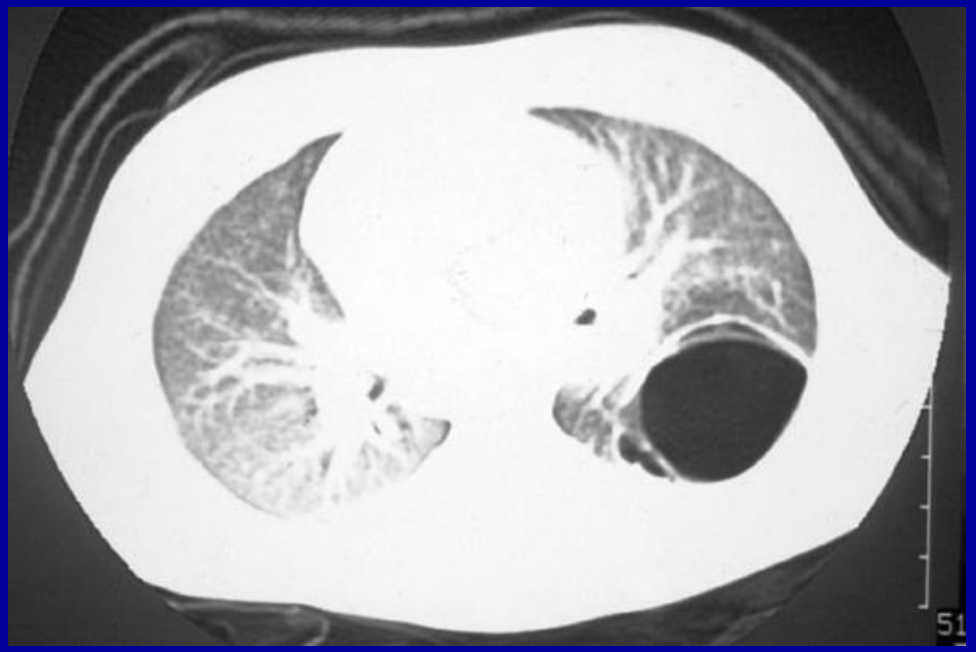

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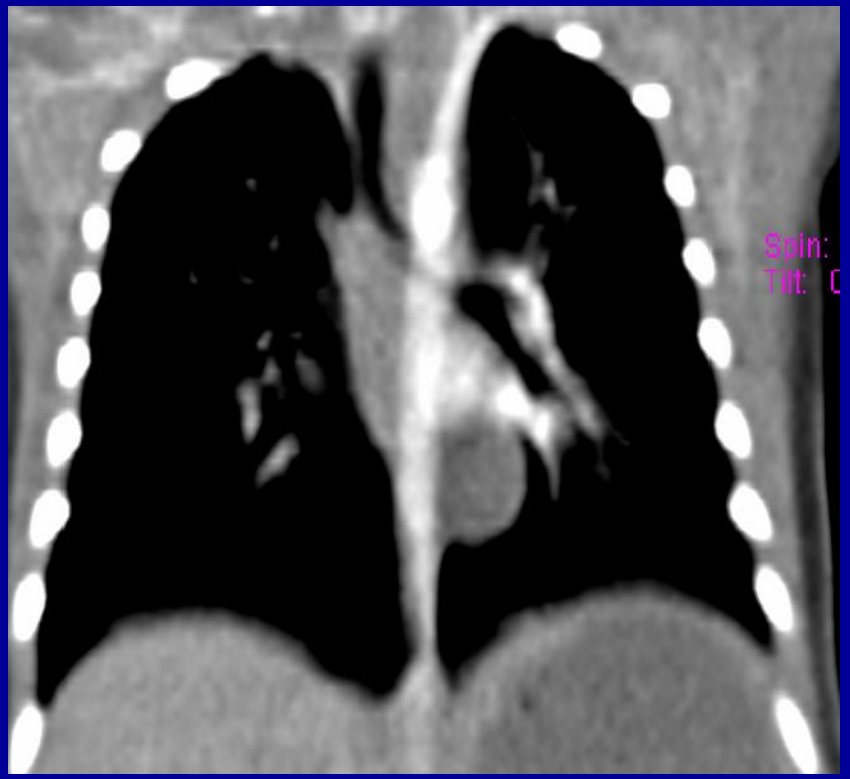
Bronchogenic Cysts

- Abnormal budding of the bronchial tree.
- 2/3 are mediastinal masses
  - not communicating with the major airways.
- 1/3 lung parenchyma.
- Unilocular cysts filled with fluid or air.



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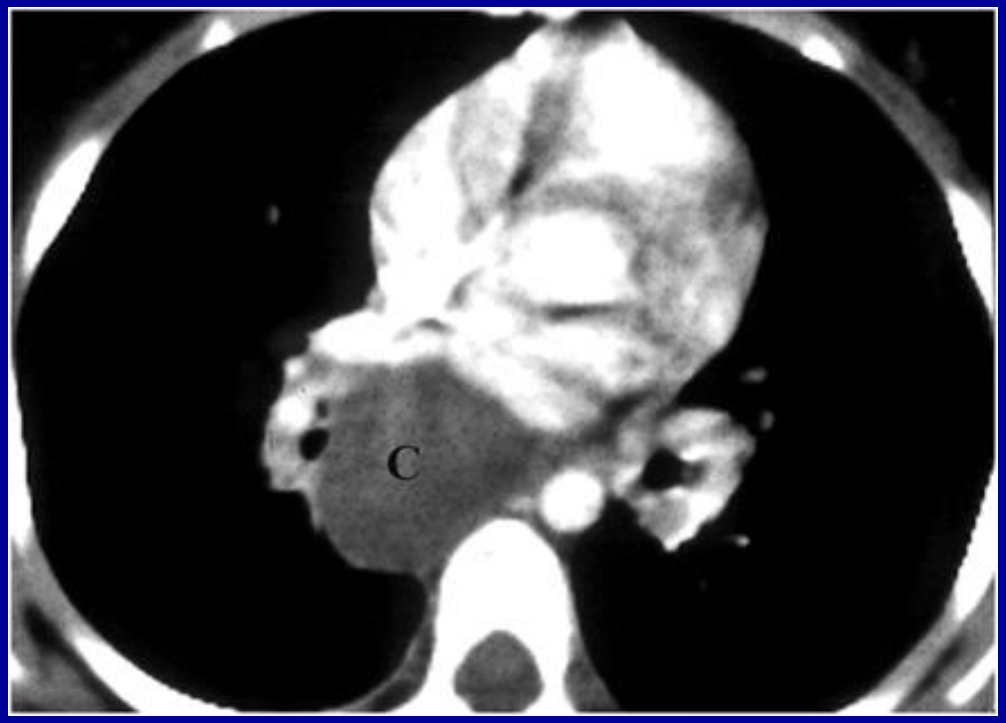

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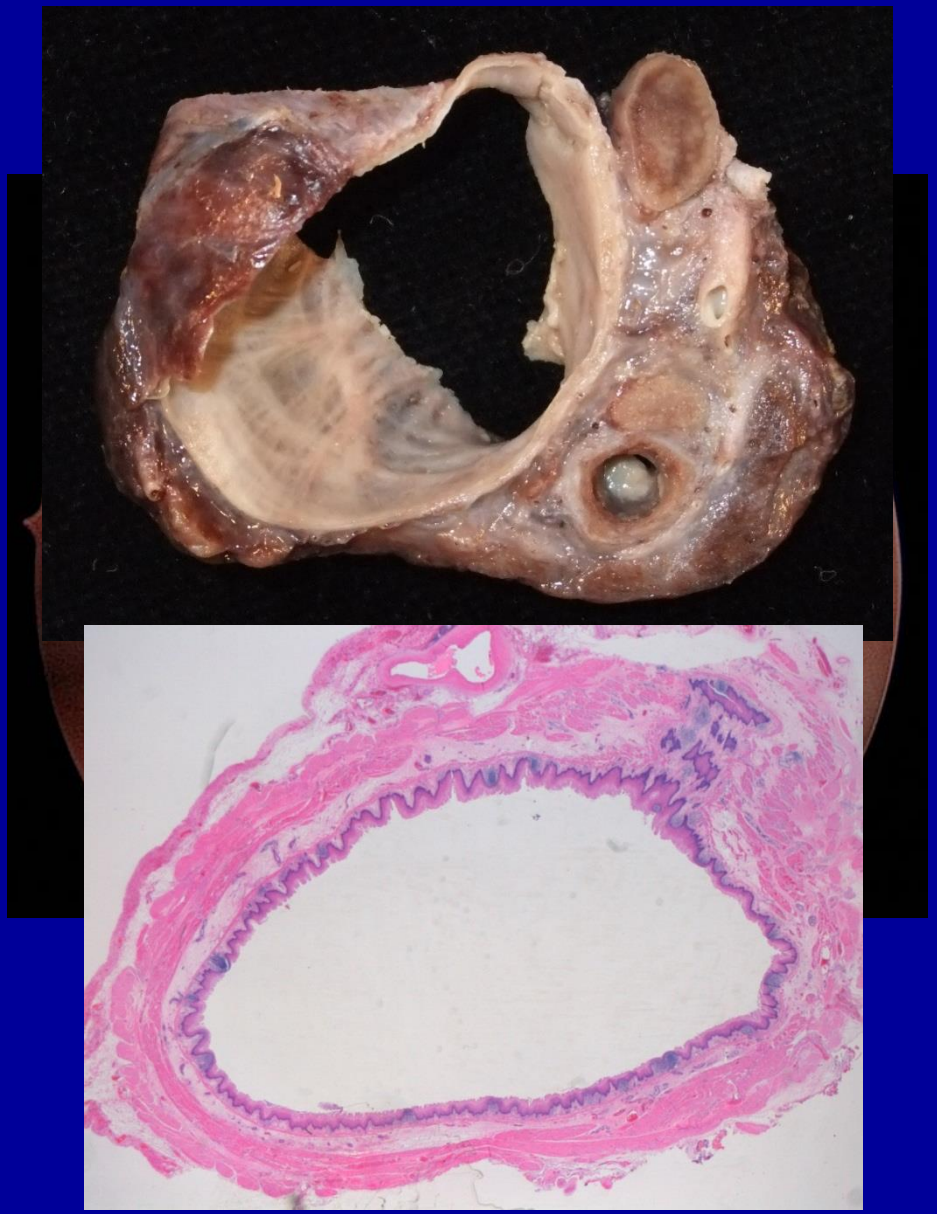
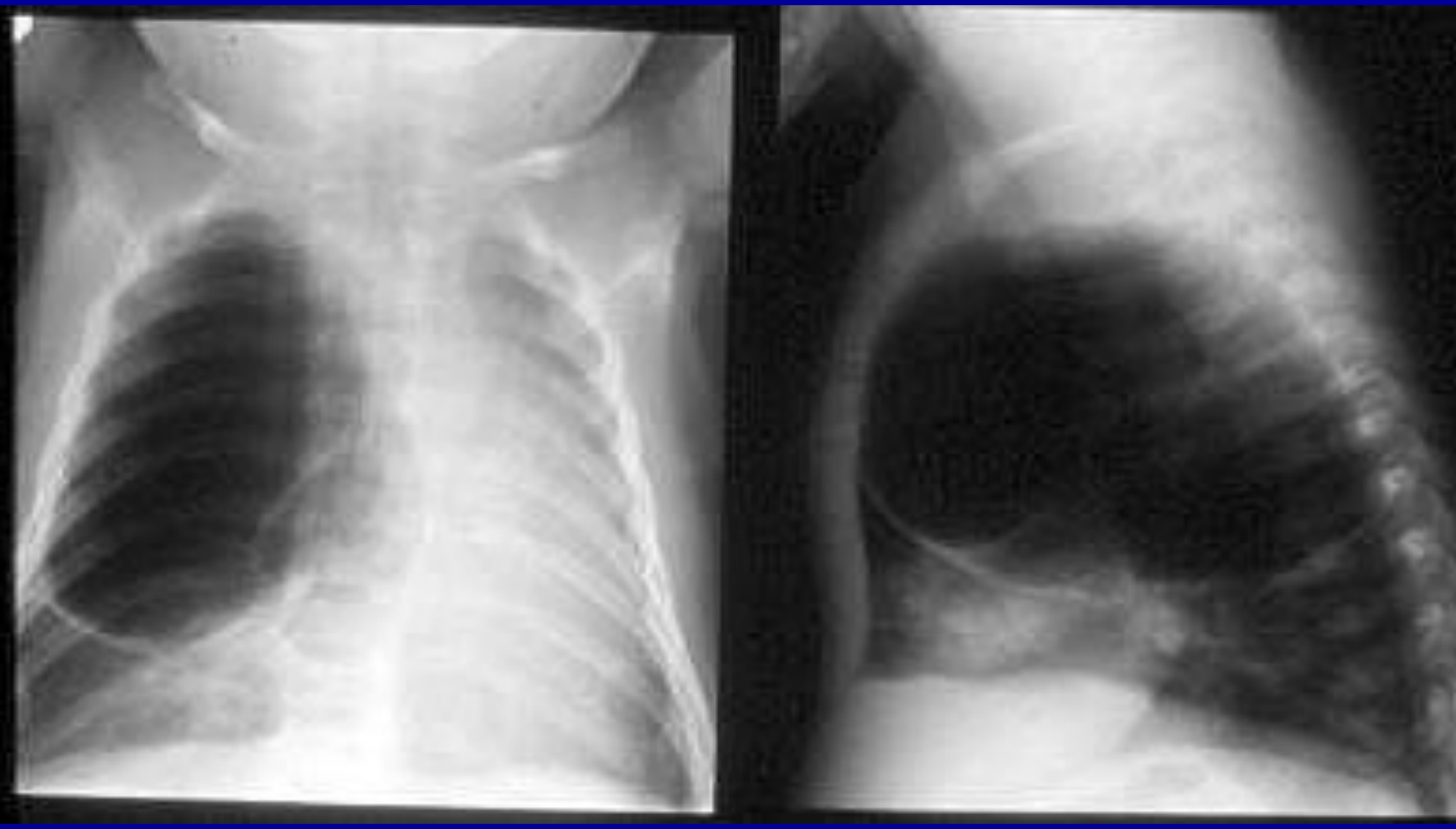


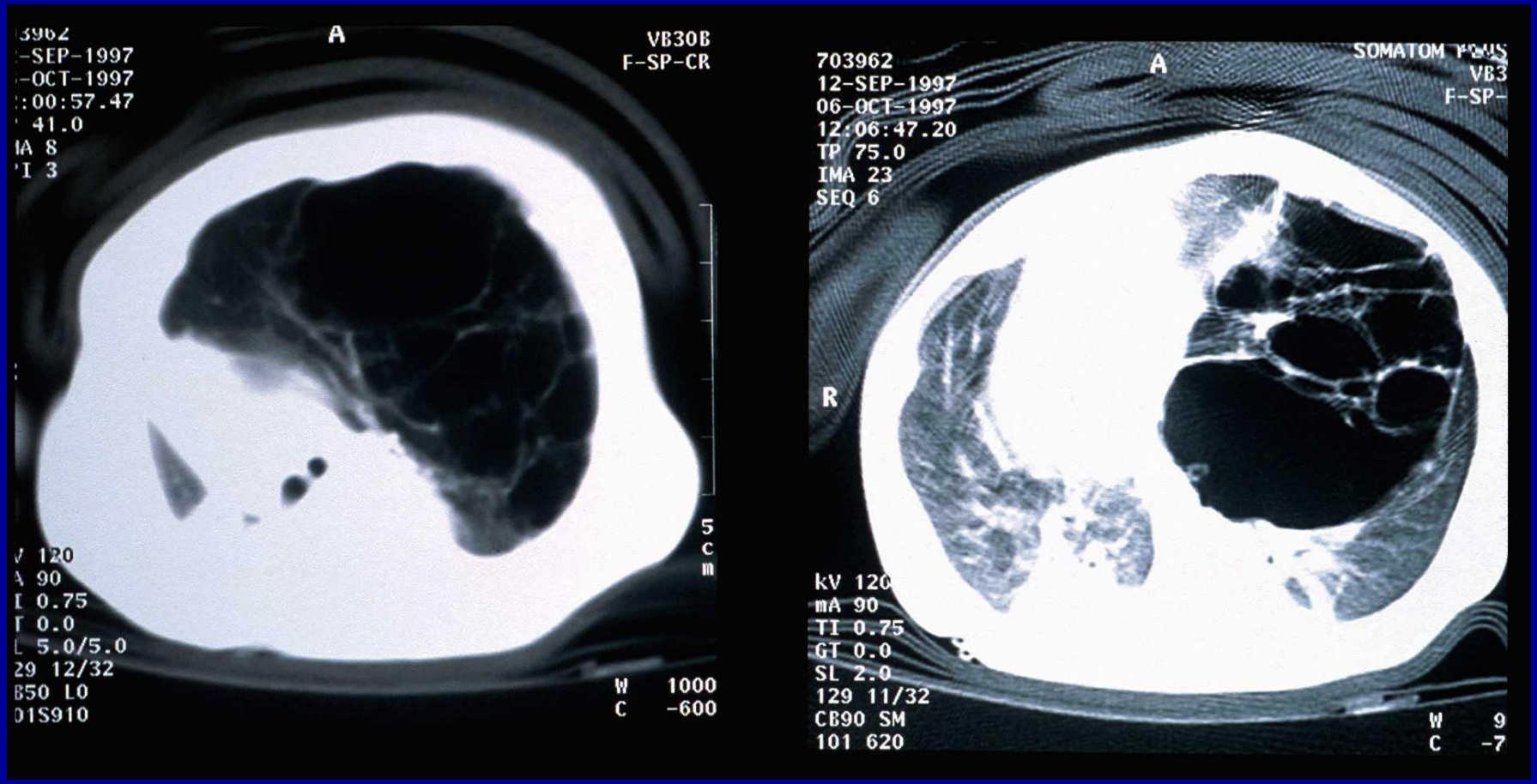
Antenatal diagnosis of a CCAM Post delivery – what happens next?

Child is unwell:

- Admit to NICU
- Imaging to include CT
- Determine type of lesion
- Usually expanding CLO
- Or large CPAM
- Plan for surgery

Child is well:

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Antenatal diagnosis of a CCAM Post delivery – what happens next?

Child is unwell:

- Admit to NICU
- Imaging to include CT
- Determine type of lesion
- Usually expanding CLO
- Or large CCAM
- Plan for surgery

Child is well:

- Baseline chest xray
- Planned non urgent CT scan with contrast
- MDT to determine diagnosis
- Meeting with family to discuss surgery vs conservative approach

Asymptomatic Congenital Lung Abnormalities

- CPAM
  - 4 types
- Pure Sequestration
- Lobar overinflation
- Bronchial agenesis
- Hybrid lesions
- Single cyst
- Bronchogenic cyst

Pure Sequestration → Probably watch and monitor & ECHO

Lobar overinflation, Bronchial agenesis → Probably watch and monitor

Single cyst, Bronchogenic cyst → Probable surgery

Probably watch and monitor & ECHO → Three options:

- Do nothing
- Consider coiling feeder arterial vessel
- Surgically remove

Asymptomatic Congenital Lung Abnormalities

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Divided opinion

Probably watch and monitor & ECHO

Probably watch and monitor

Divided opinion

Probable surgery

Three options:

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The asymptomatic CPAM
The management controversy

- Stanton, Davenport et al, J Pediatr Surg 2009. Systematic review and meta-analysis of the postnatal management of congenital cystic lung lesions

PRO SURGERY	WATCH AND WAIT
Davenport, J Pediatr Surg 2004	Aziz, J Pediatr Surg 2004
Calvert, Arch Dis Child Fetal Neonatal 2005	Hsieh, Int J Gynaecol Obstet 2005
Parikh, Pediatr Pulmonol 2005	Jaffe, Arch Dis Child 2006
Azizkhan, Pediatr Surg Int 2008	Chetcuti, Arch Dis Child Fetal Neonatal 2006
	Hammond, Eur J Pediatr Surg 2010

THE CASE FOR SURGERY

- Because they all become infected
- Because they are space occupying lesions
- Because they become malignant
- Because surgery is safe

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Pediatr Surg Int (2009) 25:479–485
DOI 10.1007/s00383-009-2371-5

ORIGINAL ARTICLE

Long-term outcome of asymptomatic patients with congenital cystic adenomatoid malformation

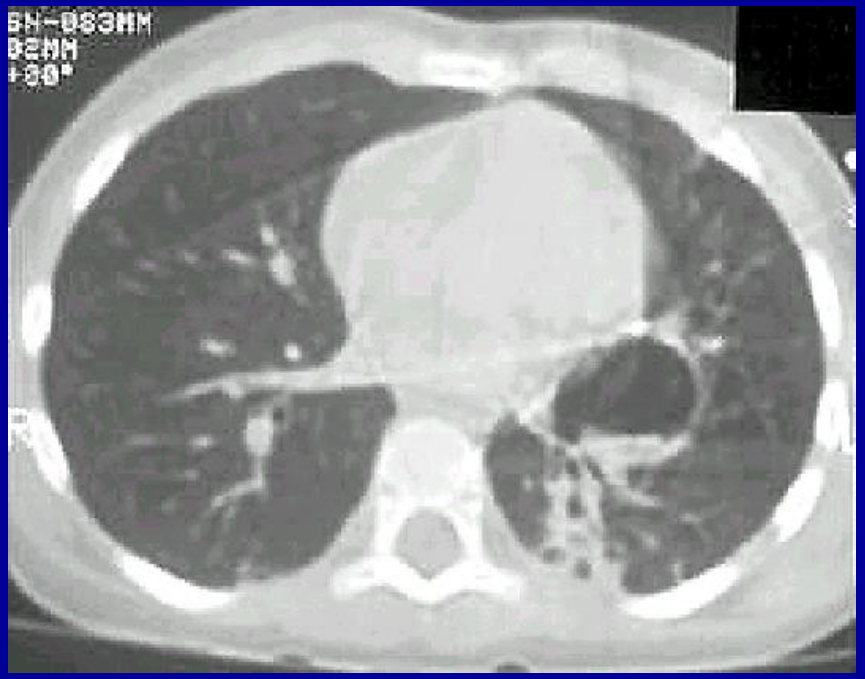
A. Wong · D. Vieten · S. Singh · J. G. Harvey · Andrew J. A. Holland

86% of asymptomatic patients eventually become symptomatic

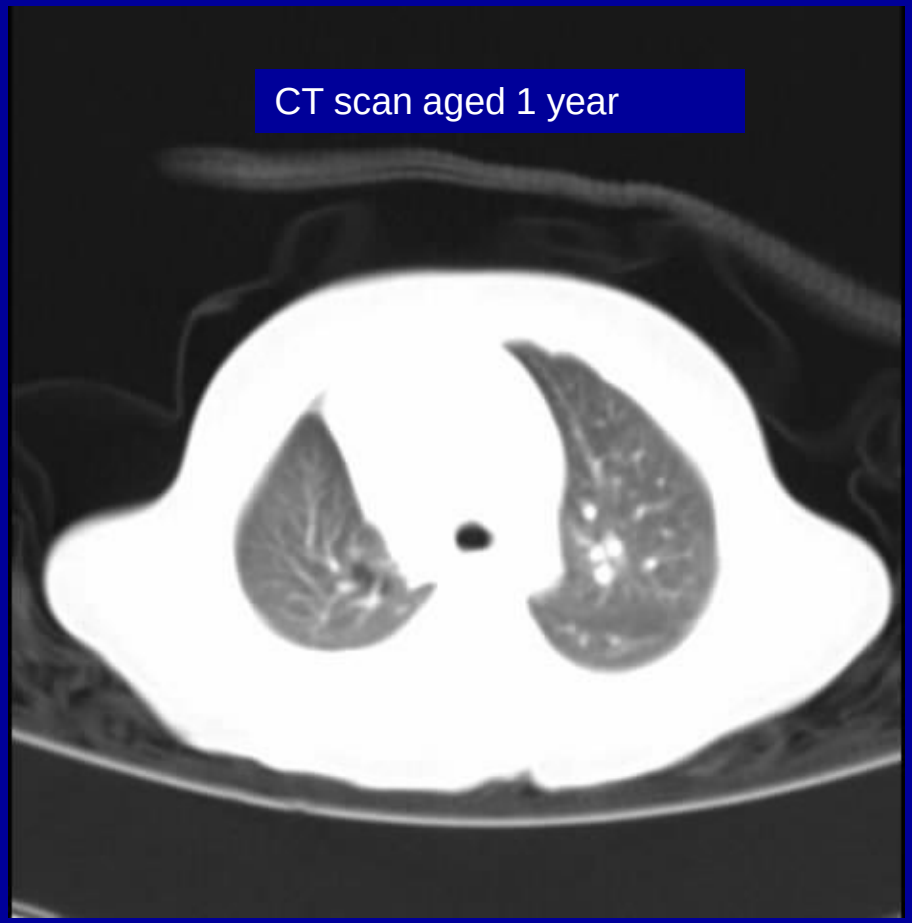
Conclusion This study suggests patients who present with asymptomatic CCAM will subsequently become symptomatic. Early surgical referral and intervention may be beneficial to avoid the development of complications.

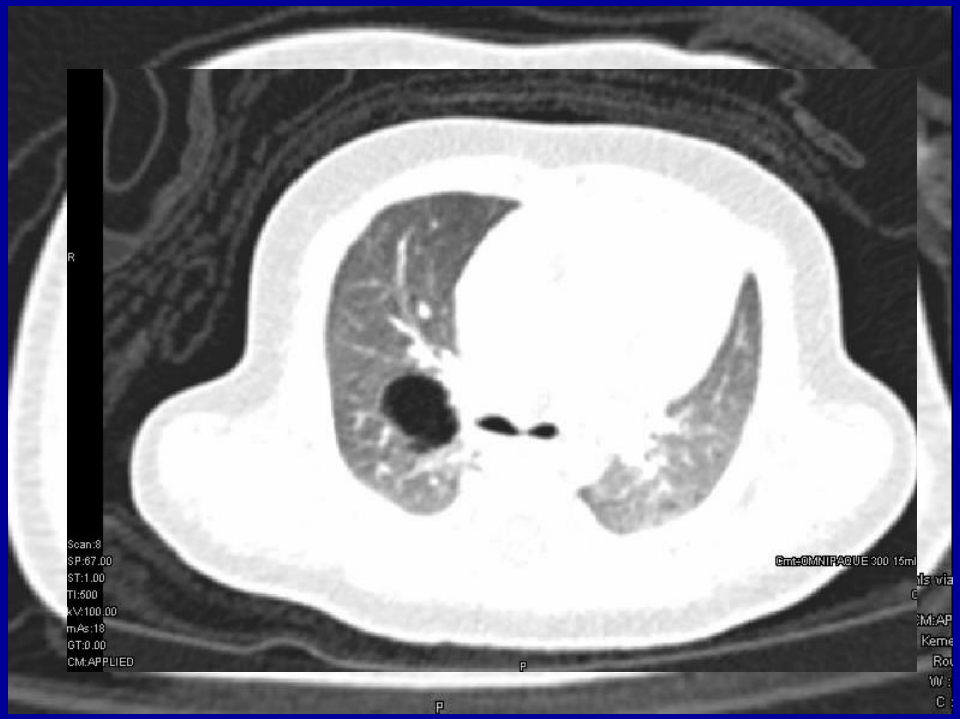
A REVIEW OF INFECTION

- Denominator to determine risk is unknown
- Reviews indicate a 10% risk **[Ref: AZIZ 2004]**
- 55% of incidental CCAMS in adults had no history of infection
  [Ref: PAPGIANNOPOULOS 2002]



Are all CPAMS at equal risk of infection?

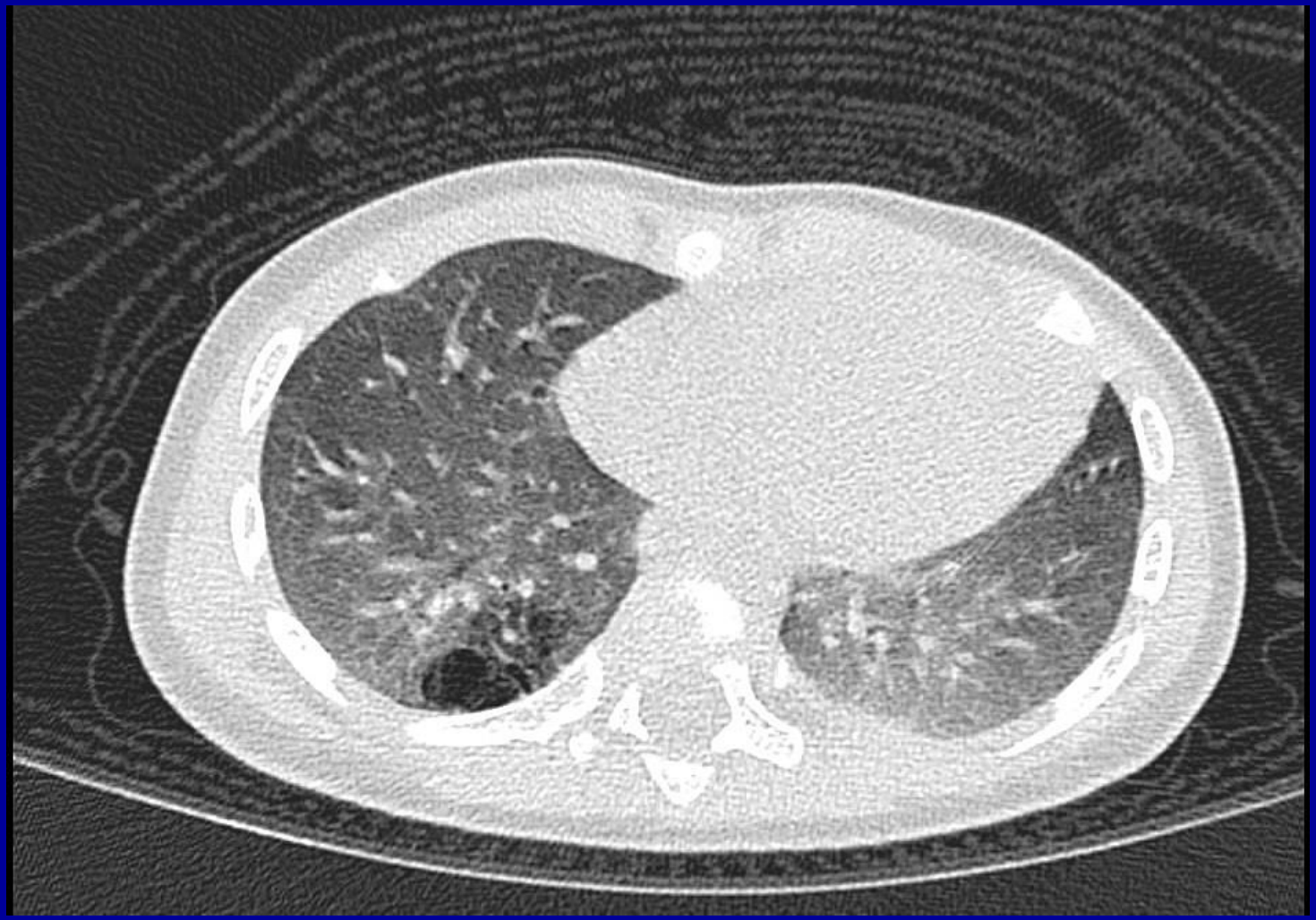




THE VERY SMALL

THE CASE FOR SURGERY

- Because they all become infected
- Because they are space occupying lesions
- Because they become malignant
- Because surgery is safe



THE CASE FOR SURGERY

- Because they all become infected
- Because they are space occupying lesions
- Because they become malignant
- Because surgery is safe

A REVIEW OF MALIGNANCY

- Pleuropulmonary Blastoma
- Bronchioalveolar Carcinoma
- Rhabdomyosarcoma

A REVIEW OF MALIGNANCY

- Pleuropulmonary Blastoma
- Bronchioalveolar Carcinoma
- Rhabdomyosarcoma

- **Among the rarest tumours of childhood**
- **Considerable histological overlap with CCAM IV**

  [Ref: Miniati 2006]
- **May be present antenatally**
- **25% have a predisposition to dysplastic or neoplastic disease**

  [Ref:Priest 1996]
- **Can appear in healthy lung away from previous CCAM resection**

  [Ref: Payagiannopoulos 2001; Kazlovsky 1997; Indolfi 2000]

A REVIEW OF MALIGNANCY

- Pleuropulmonary Blastoma
- Bronchioalveolar Carcinoma
- Rhabdomyosarcoma

- **Hypothesis: cysts associated with PPB are not CPAM but may be separate diagnosable pathologic entities (Type I “cystic” PPB).**
- **PPB is not a pre-existing CPAM which has undergone “malignant transformation”.**
- **Cystic PPB is the earliest manifestation of a sequence in which low-grade cystic PPB evolves over 2-4 years to a high-grade, solid sarcomatous disease.**

A REVIEW OF MALIGNANCY

- Pleuropulmonary Blastoma
- Bronchioalveolar Carcinoma
- Rhabdomyosarcoma

Be suspicious if:

- **Large cyst/s, usually multilocular, peripheral lung**
- **± Respiratory distress**
- **± Pneumothorax (43%)**
- **10% bilateral cysts (vs 2% CPAM)**
- **5% multifocal cysts**
- **Typically presents within the first 2 yrs of life**

REF: Dehner Ped Surg Int 2005;21:123-4

Paediatric Respiratory Reviews 19 (2016) 62–68

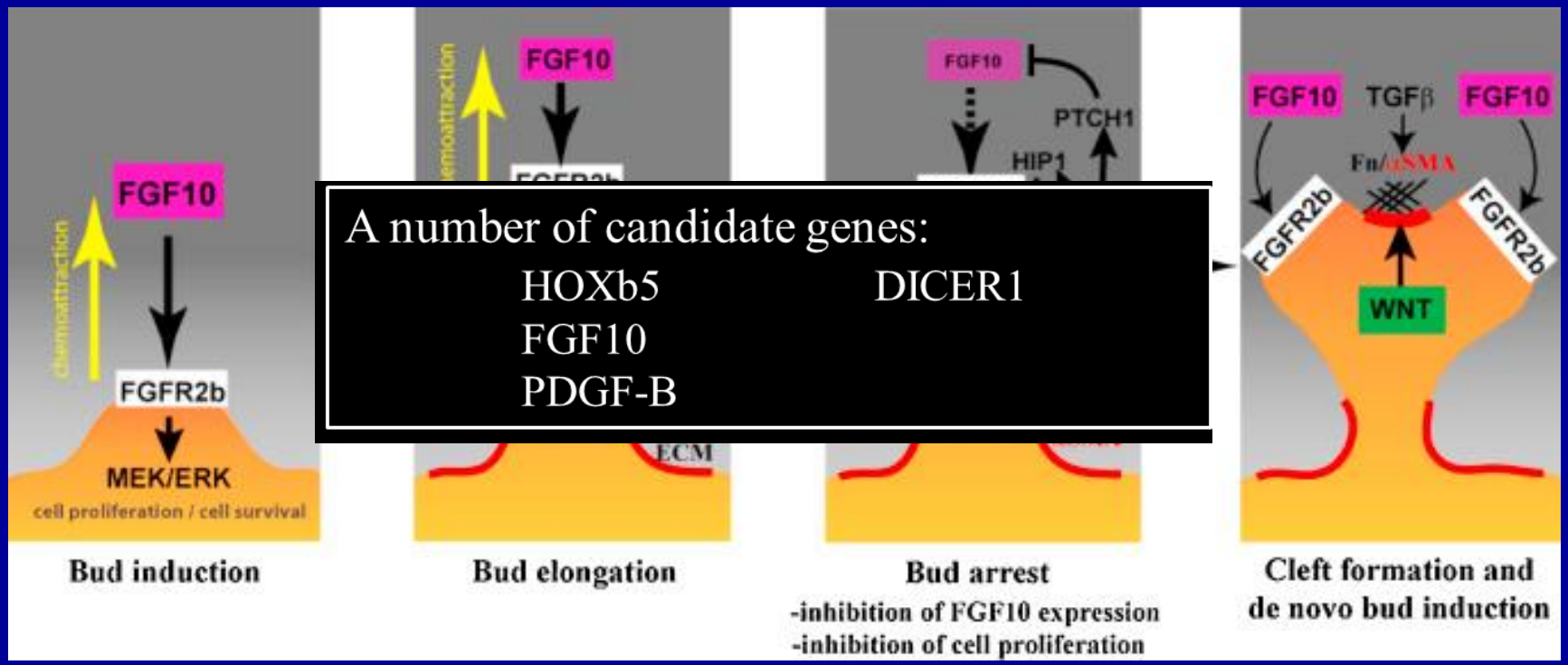
Contents lists available at ScienceDirect

Paediatric Respiratory Reviews

Review

Pathomechanisms of Congenital Cystic Lung Diseases: Focus on Congenital Cystic Adenomatoid Malformation and Pleuropulmonary Blastoma

Olivier Boucherat [1,*], Lucie Jeannotte [2,3], Alice Hadchouel [4,5,6], Christophe Delacourt [4,5,6], Alexandra Benachi [7,8,*]



A number of candidate genes:

HOXb5 DICER1
FGF10
PDGF-B

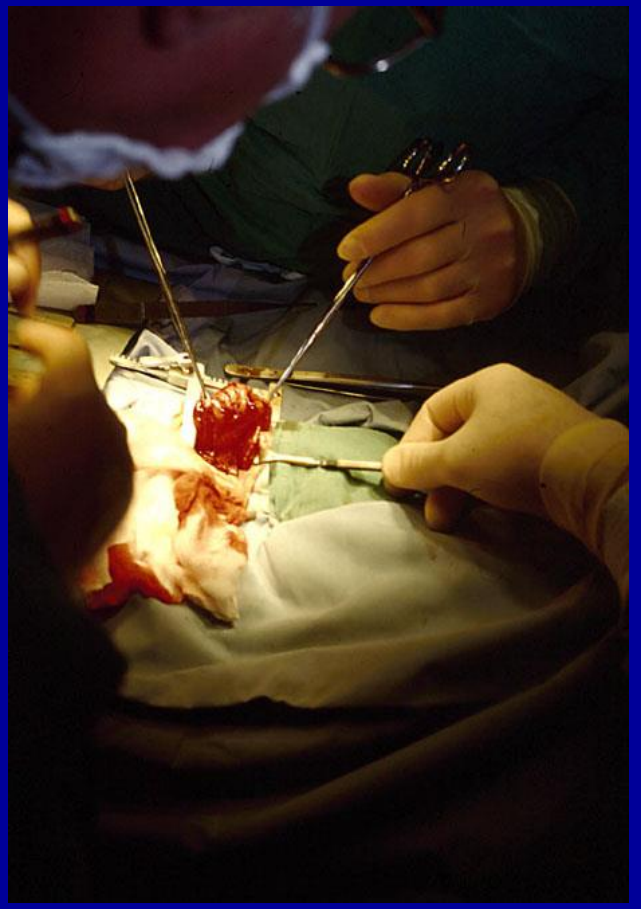
THE CASE FOR SURGERY

- Because they all become infected
- Because they are space occupying lesions
- Because they become malignant
- Because surgery is safe

IS SURGERY SAFE?

COMPLICATIONS:

- Prolonged air leak
- Wound infection
- Pneumonia
- Empyema
- Haemorrhage
- Portal vein thrombosis
- Residual cysts (30%)
- (Removal of normal lung)



[REF Keidar S; 2001]

GOSH outcome of conservative lesions – a prospective study

- We will watch the small ones
- We will watch those who do not have a large cystic component
- We will watch those who do not have sinister features
- We will watch when parents are happy for a conservative approach in a completely asymptomatic child

CPAM / PS 119

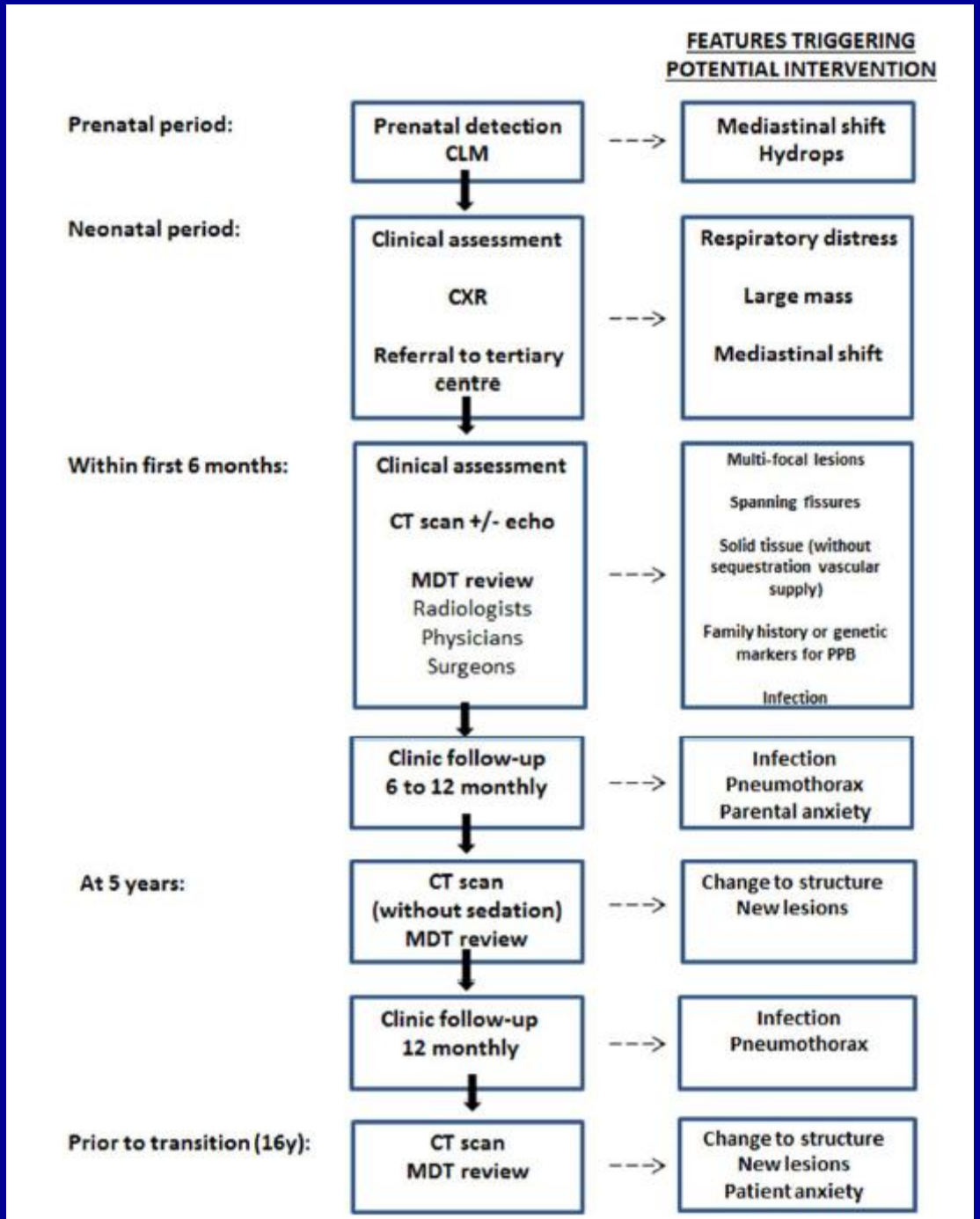
Conservative 68

- Median follow up for 9.9 years [range 5 – 18]
- 4 spontaneously resolved
- 8 lost to follow up
- 52 remain well

Surgery 51

- 8 had emergency surgery
- 6 concerning features on CT
- 20 Elective (parental decision)
- 12 Chest infections
- 5 unknown
- 0 malignant change in resected specimens

REF: Cook et al ADC 2017


FEATURES TRIGGERING POTENTIAL INTERVENTION
Prenatal period:
Prenatal detection CLM
Mediastinal shift Hydrops
Neonatal period:
Clinical assessment
CXR
Referral to tertiary centre
Respiratory distress
Large mass
Mediastinal shift
Within first 6 months:
Clinical assessment
CT scan +/- echo
MDT review
Radiologists
Physicians
Surgeons
Multi-focal lesions
Spanning fissures
Solid tissue (without sequestration vascular supply)
Family history or genetic markers for PPB
Infection
Clinic follow-up 6 to 12 monthly
Infection
Pneumothorax
Parental anxiety
At 5 years:
CT scan (without sedation) MDT review
Change to structure
New lesions
Clinic follow-up 12 monthly
Infection
Pneumothorax
Prior to transition (16y):
CT scan MDT review
Change to structure
New lesions
Patient anxiety

In Summary

- Parents arrive with high anxiety even though the child is well
- Contrast CT scan in all – not all will be CPAMs / CCAMs
- Discuss the little that is known
- No rush to act on asymptomatic lesions
- Consider a repeat CT at 12 – 18 months

- Surgery may not be:
  - indicated (bronchial atresia)
  - desirable (very small)
  - necessary (pure sequestration)
  - definitive (residual cysts, multiple lobes, malignancy in distant sites)

- Surgery on a case by case basis
  - the very big or multicystic,
  - those with unusual features, family history of malignancy
  - parental request / anxiety,
  - infection concerns

THE

END