More on Neonatal Lung Cysts

Leon Joseph and Elie Picard

Shaare Zedek Medical Center

A.B.

- 22yr old Gravid 1
- Natural pregnancy
- Normal OGTT
- Symptomatic UTI week 13 - zinnat
- URTI week 17 –azithromycin
- Triple test, early and late US normal

A.B.

- Presented week 41
  - Fever
  - PROM 26 hours
  - Failed trial spontaneous labour and induction
  - Fetal distress
  - Lower segment cesarean section

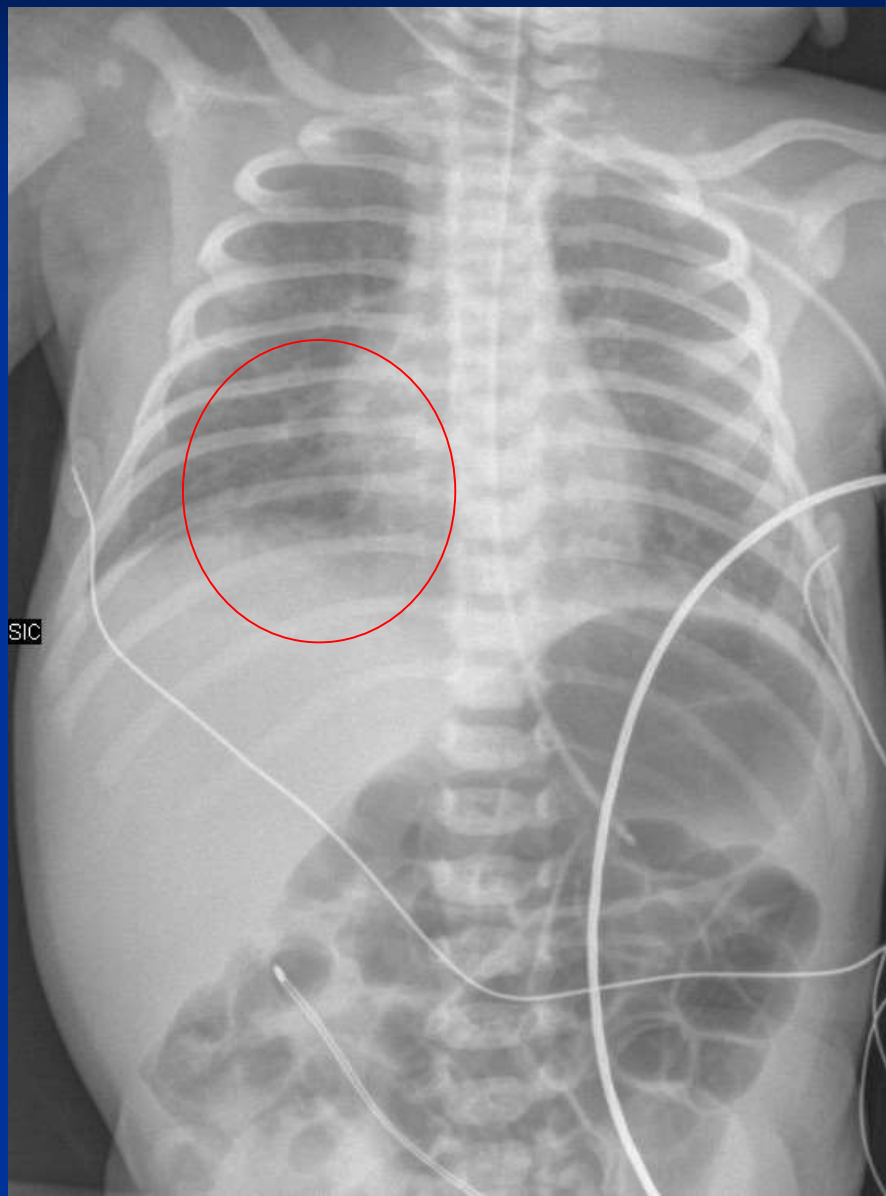
A.B.

- Foul meconium stained liquor
- Grunting and hypoxia – sats 88%
- NICU
- PH=7.16, PCO2=52, PO2=31, HCO3=15, BE=-9

A.B.

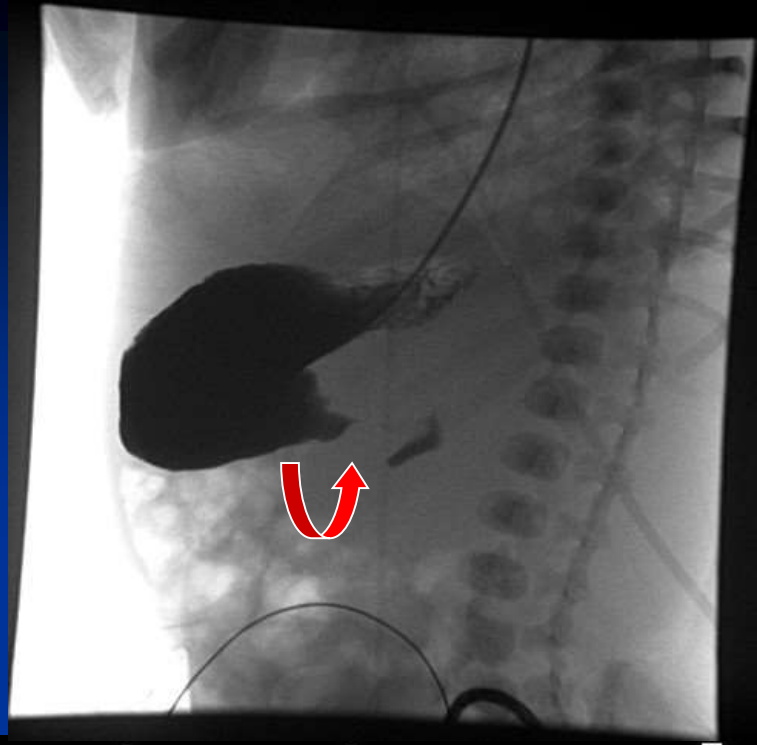
- NCPAP
- Negative sepsis screen
- Ampicillin & Gentamycin
- CXR

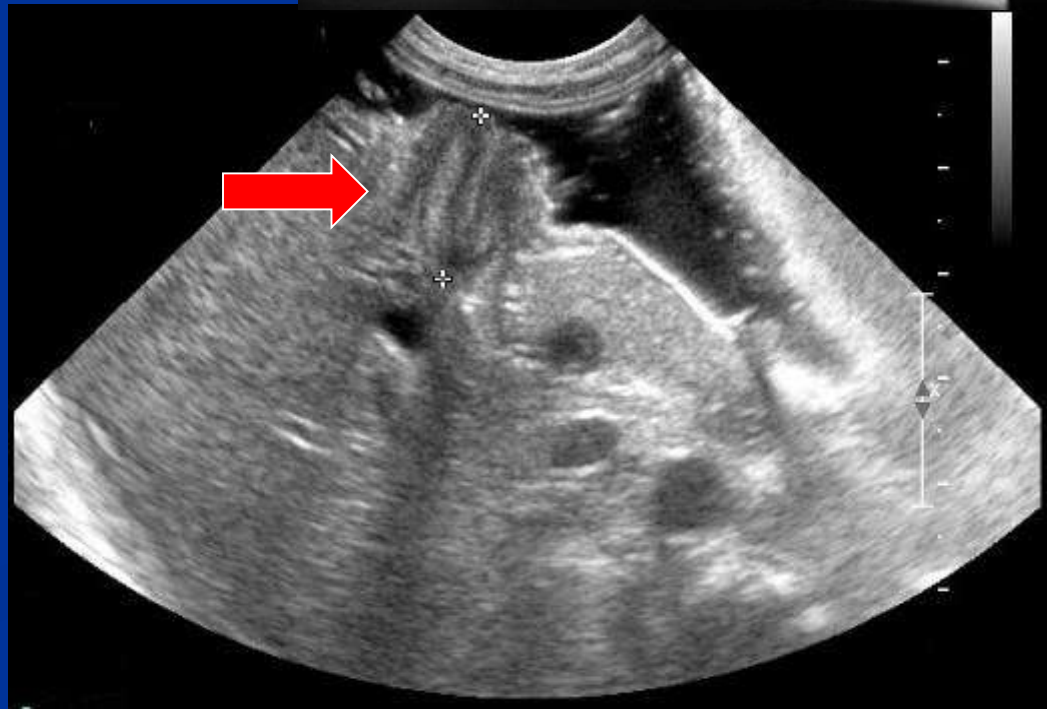
- Weaned off CPAP



A.B.

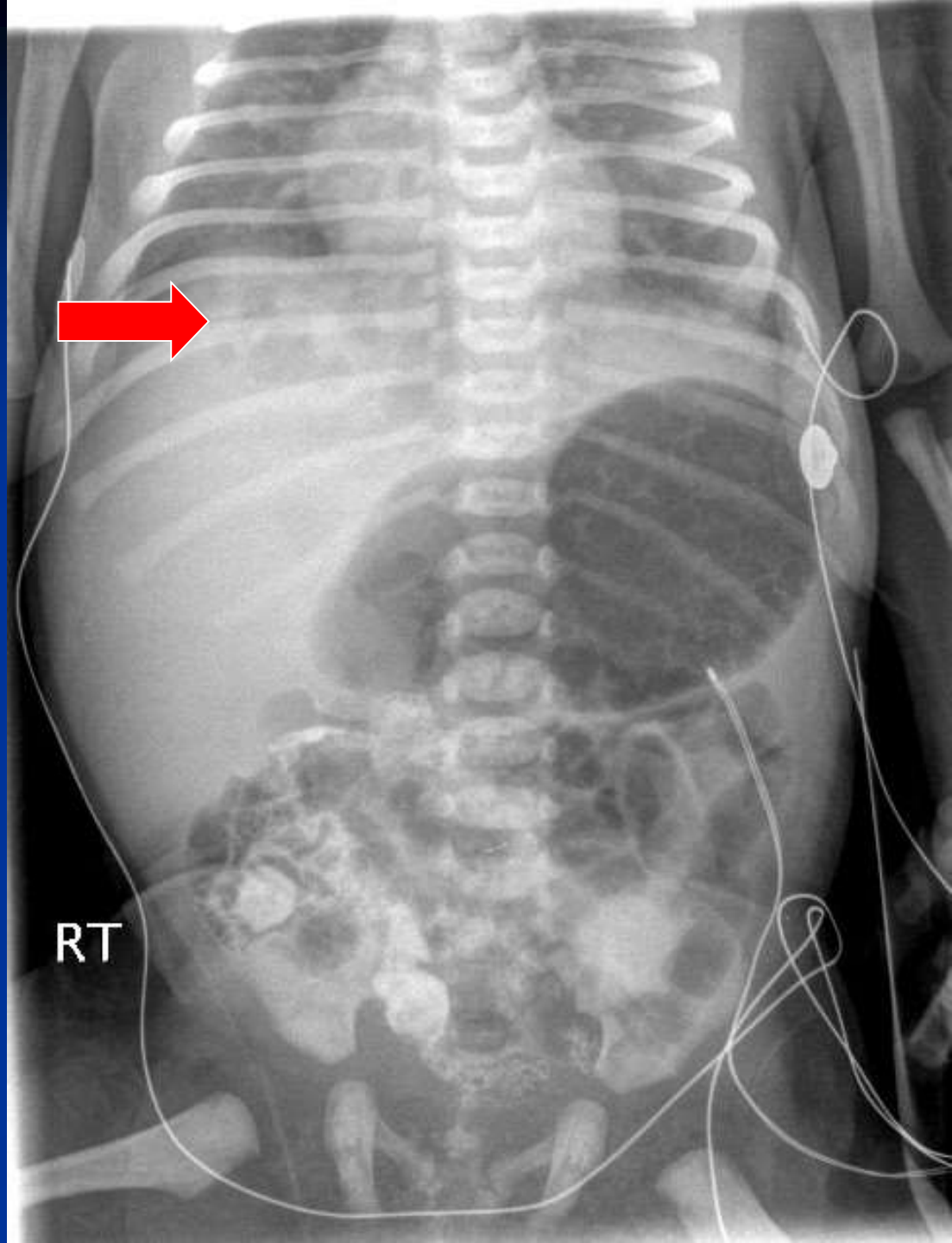
- Day 2 introduced feeding
- Recurrent vomiting
- AXR unremarkable
- UGI – suspected pyloric stenosis
- US – confirmed





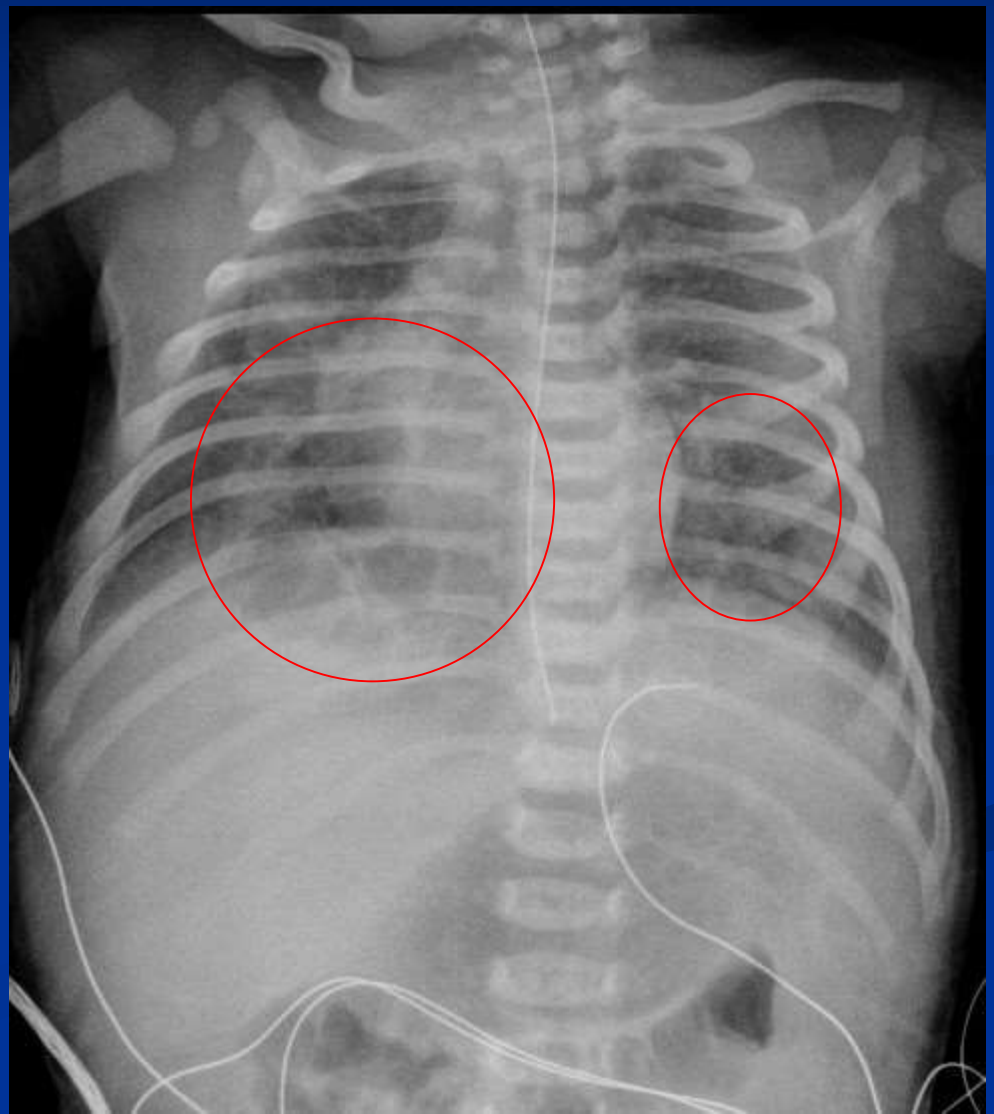
A.B.

- Day 6 – Pyloromyotomy
- Mild post – operative wound infection
- Full recovery
- CXR – peri-operatively

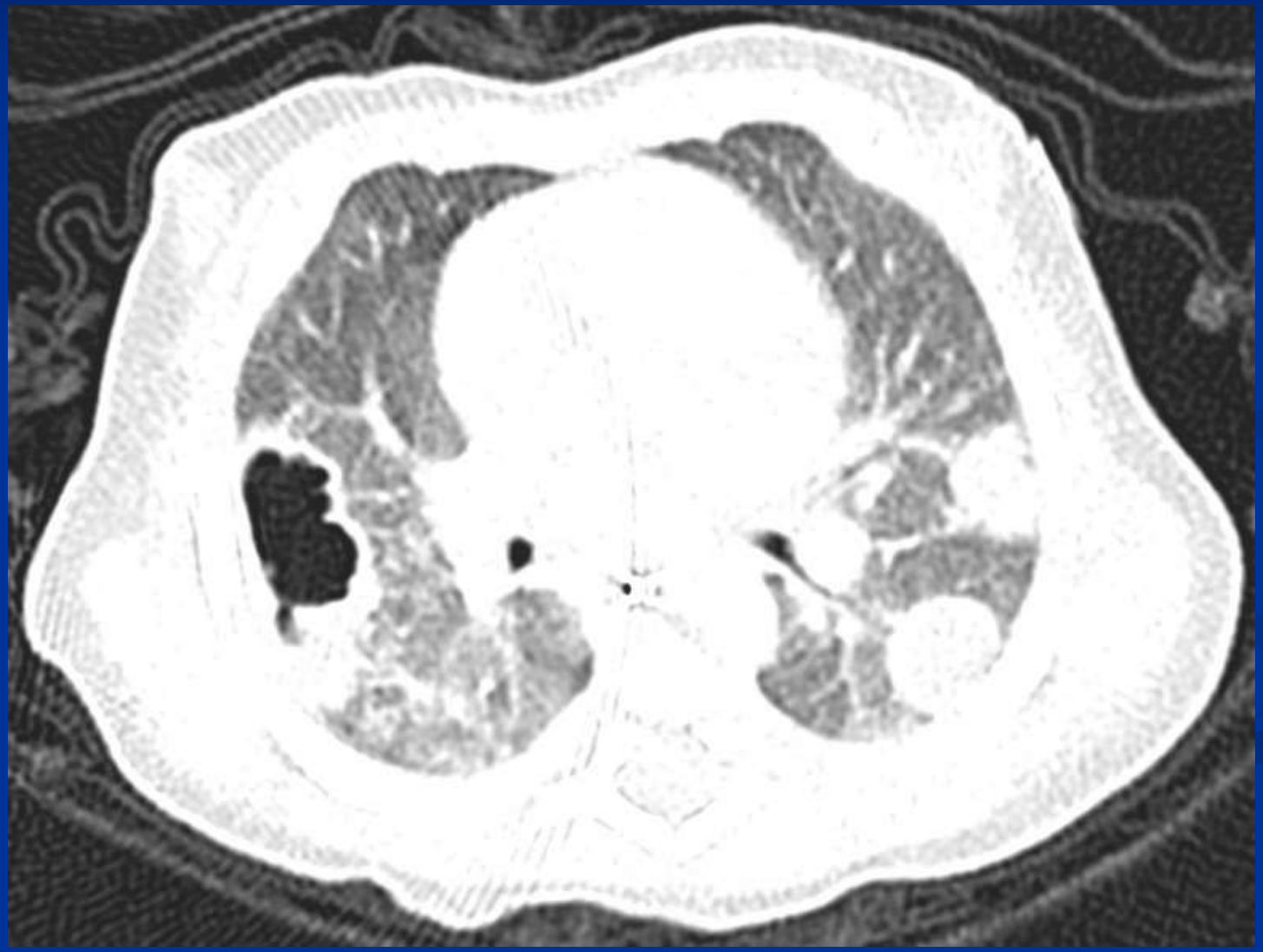


A.B.

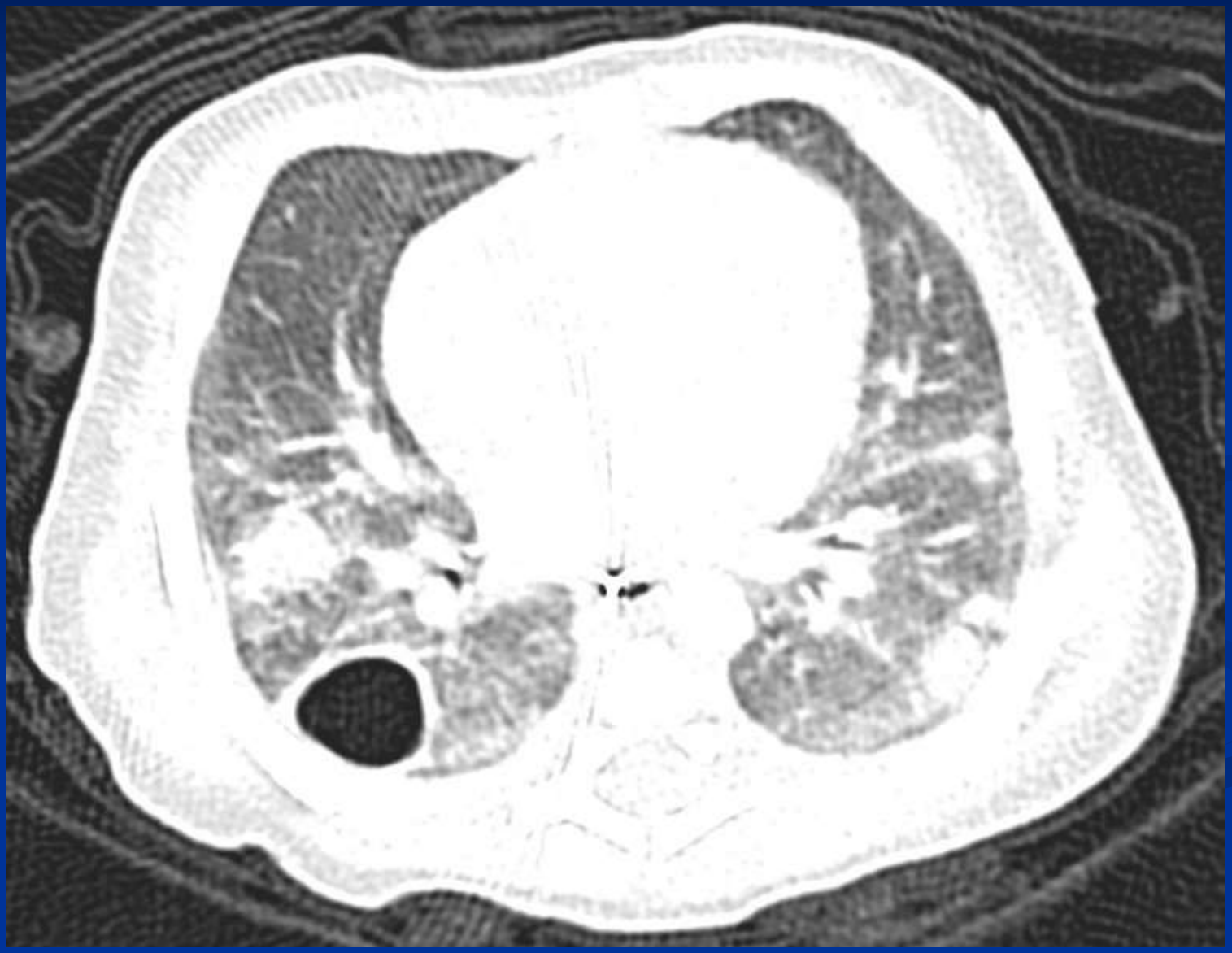
- Improving clinically
- Good air entry
- No O2 requirement
- No tachypnea or dyspnea



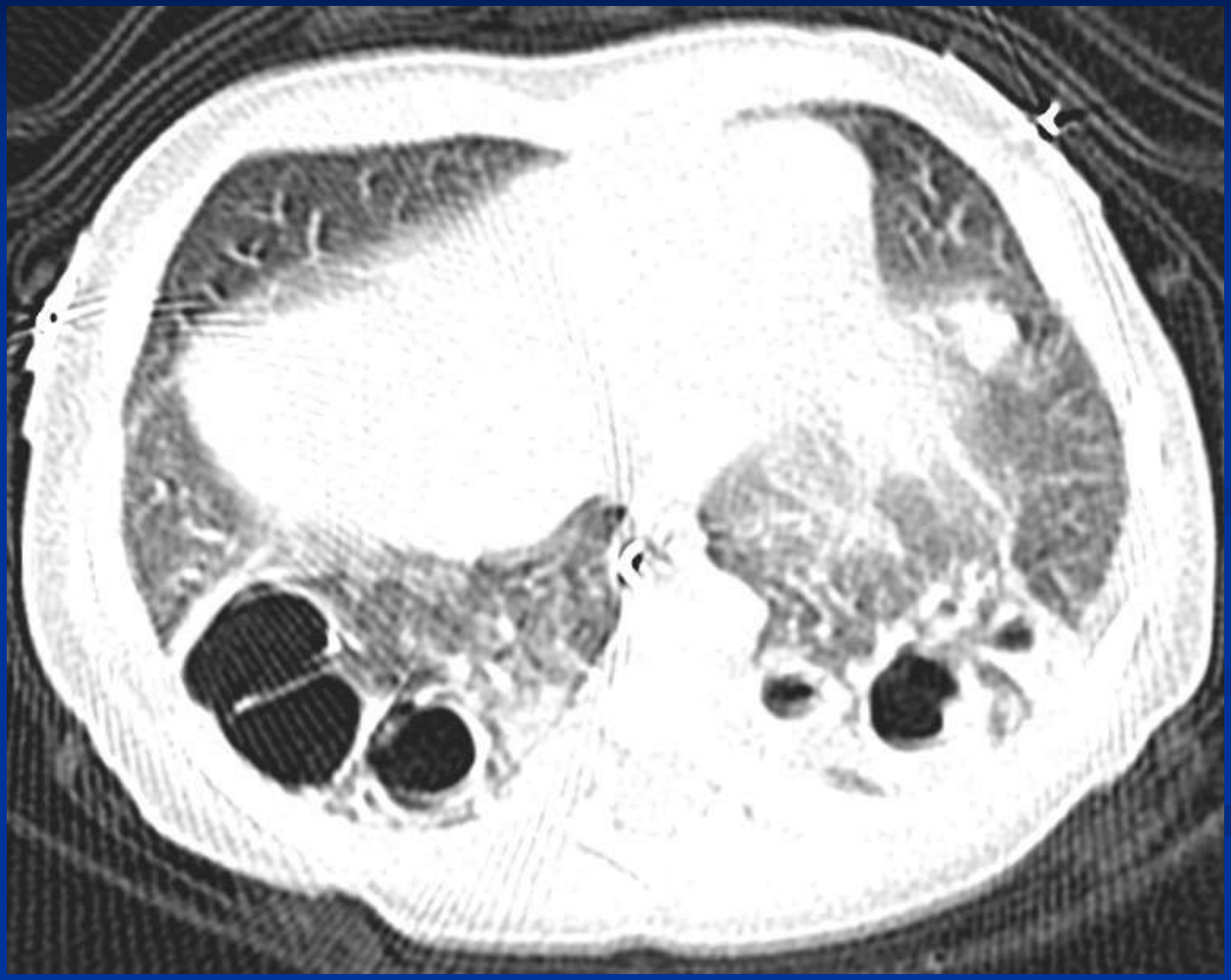
CT chest



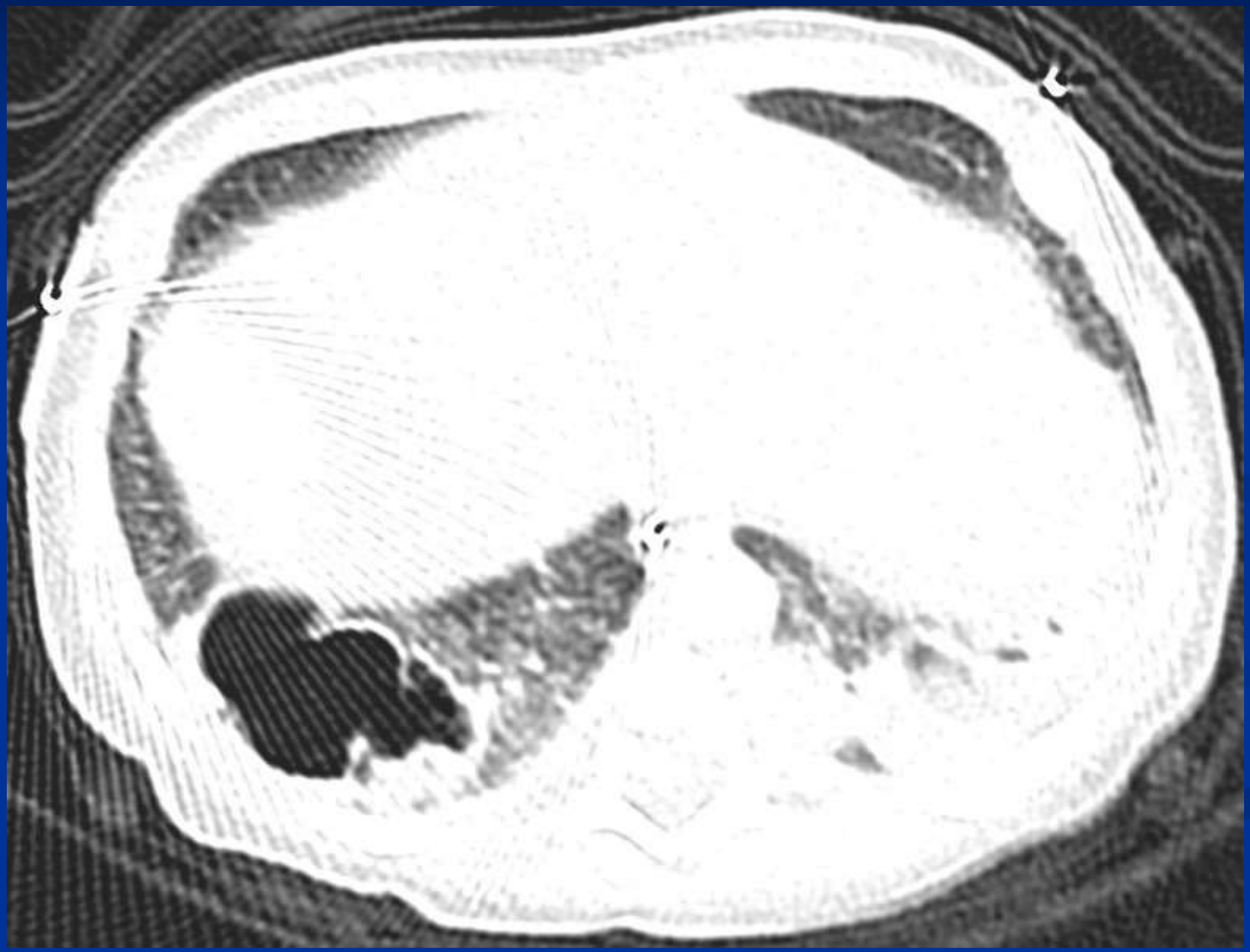
CT chest



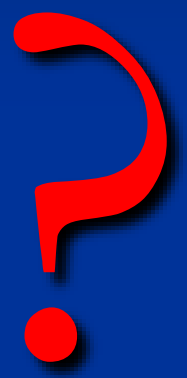
CT chest



CT chest



Differential Diagnosis



Differential Diagnosis

- Congenital CPAM: (PPB?)
- Infectious – Bacterial
  CMV or other viral
  fungal / TB
- PIE
- Langerhan's cell Histiocytosis

Not in keeping with improving clinical picture

State of the Art

Pulmonary Cysts in Early Childhood and the Risk of Malignancy

John R. Priest, MD,[1]* Gretchen M. Williams, BS,[2] D. Ashley Hill, MD,[3] Louis P. Dehner, MD,[3] and Adam Jaffé, MD, FRCP, FRCPCH, FRACP[4]

Pediatric Pulmonology 44:14–30 (2009)

CPAM

- Congenital Cystic malformation:
  - CCAM/CPAM 0-4
  - 1,2,4 = 85% - these are air filled
  - 2 are small up to 2.5cm – may exist with other extrapulmonary malformations – not associated with malignancy.
  - 1&4 are large up to 10cm cysts – associated with malignancy

CPAM

TABLE 1—Nomenclature and Features of Congenital Cystic Adenomatoid Malformation (CCAM) and Congenital Pulmonary Airway Malformation (CPAM)[1]

CCAM name	N/A	Type I	Type II	Type III	N/A
CPAM name	Type 0	Type 1	Type 2	Type 3	Type 4
Descriptive name	"Acinar dyplasia or dygenesis"	"The large cyst lesion"	"The small cyst lesion"	"The adenomatoid lesion"	"The unlined cyst lesion" (actually lined by flattened respiratory epithelium)
Postulated airway origin	Tracheal/bronchial	Bronchial/bronchiolar	Bronchiolar	Bronchiolar/alveolar duct	Distal acinar
Cystic?	No	Yes	Yes, multiple	No (or scattered)	Yes
Adenomatoid?	No	No	No	Yes	No
Proportion of CPAM	<2%	60–70%	15–20%	5–10%	10%
Unique features	All lobes involved; incompatible with life			More common in males	Tension pneumothorax
Typical age at presentation	Birth	In utero, if large, to many years of age, if small	1st month of life	In utero; or at birth	Newborn to 6 years, or rarely much later
Presentation	Lungs will not aerate	*Newborn*: respiratory distress, mediastinal shift; or *later*: incidentally, or cough + fever + infection, or emergence of tumor	Non-pulmonary anomalies may supercede lung abnormalities	May be stillborn, or severe neonatal respiratory distress	Respiratory distress ± tension pneumothorax, or infection/pneumonia, or incidental finding, or emergence of tumor
Lobar involvement	All lobes involved	One lobe in 95%; rarely bilateral	Usually one lobe	Entire lobe or lung	Usually one lobe
Associated anomalies	CV anomalies, renal hypoplasia, focal dermal hypoplasia	None	CV anomalies, diaphragmatic hernia, extralobar sequestration, renal agenesis or dysgenesis	N/A	Patient or familial childhood neoplasia and dysplasia suggests PPB (see Table 3)
Lesion and cyst size	Lungs small	1–10 cm	0.5–2.0 cm	Entire lobe or lung	Large multilocular cysts
Malignancy risk	No	BAC	No	No	PPB

[1]Sources: Stocker et al.,[42] Stocker,[41] Bush,[1] Langston,[39] and International PPB Registry unpublished data.

PPB

TABLE 2—Malignancies Associated With Lung Cysts in Children

Malignancy	Associated cyst type	Number of cases in patients <20 years of age	Gross morphology	Age at diagnosis		Common presentations[1]	Prognosis
				Median	Range		
Type I PPB	Type I PPB (CPAM type 4)	~450	Cystic	9 mo	0–114 mo	Dyspnea, pneumothorax in 40%, or incidental discovery	85–90%[4]
Type II PPB			Cystic and solid	36 mo	6–236 mo[3]	Dyspnea, pneumothorax in 28%, "pneumonia," or incidental discovery	45–60%[4]
Type III PPB			Solid	43 mo	18–147 mo	Dyspnea, "pneumonia"	
Bronchiolo-alveolar carcinoma (BAC) in CPAM	CPAM type 1	<10	Cystic and solid	20 yr[2]	0.5–75 yr	Recurrent infection, hemoptysis, cough, chest pain, pleural effusion	Too few cases
Primary pleuropulmonary synovial sarcoma	Unknown	<5	Cysts (blebs), cystic and solid, solid	30 yr	15–72 yr	Dyspnea, cough, pneumothorax, chest pain, hemoptysis, hemothorax	~60% alive[85]

[1] "Pneumonia:" dyspnea, cough, fever, chest and/or abdominal pain, anorexia and/or malaise.
[2] BAC *not* associated with CPAM has median diagnosis age 68 years.
[3] Excludes an unusual patient diagnosed at 36 years.[94]
[4] Five-year Kaplan–Meier overall survival.

PPB

- CPAM is 98% unilateral and 95% in one lobe vs PPB – 10% bilateral and unilateral multifocal 5%.

- IPPBR data (estimated) – (www.ppbregistry.org)
  - PPB: 0.35-0.65/100,000 births
  - CPAM: 2-8/100,000 births

מה השלב הבא?

Aspiration of cyst

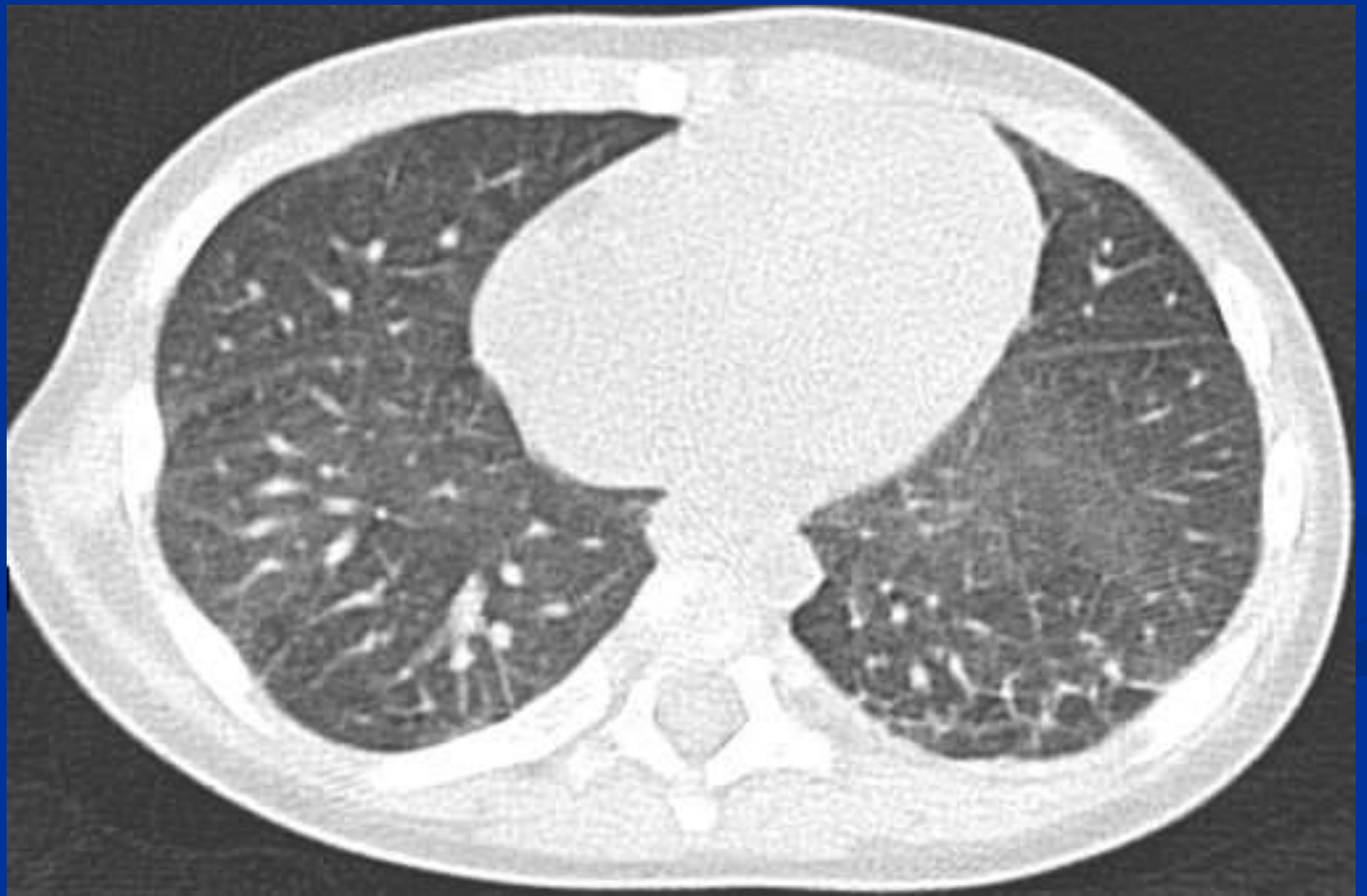
- בדיקה מיקרוסקופית -
- תפליט דלקתי חריף, פרורי רקמת גרעון מודלקת ופרורי רקמת ריאה.
- בצביעה ל CD68-נראו מאקרופגים מפוזרים, כולל תאי ענק רב גרעיניים.
- צביעות גרם, ציל- נילסן PAS ו CMV-שליליות.
- מתאים ל.acute pneumonia-
- לא זוהו סימני ממאירות.

Microbiology

- PCR based bacterial identification
  - Gram negative bacteria
  - Consistent with E Coli
- E. Coli on maternal placental culture
- IV Augmentin 10 days.

Follow up

- No respiratory symptoms or signs



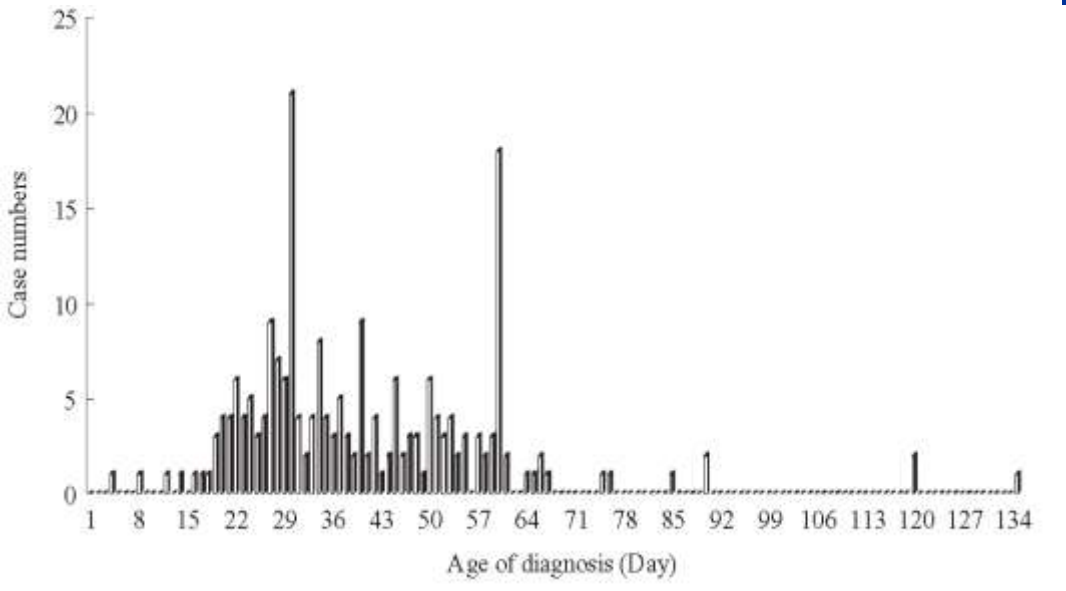
PEDIATRICS INTERNATIONAL Official Journal of the Japan Pediatric Society
Pediatrics International (2011) **53**, 18–23 doi: 10.1111/j.1442-200X.2010.03185.x

Original Article

Infantile hypertrophic pyloric stenosis before 3 weeks of age in infants and preterm babies

Results: A total of 214 patients were enrolled into the study; the mean age of diagnosis was 40 days of age; the average duration of hospital stay was 6.27 days. Eighteen (8.41%) patients were diagnosed before 3 weeks of age. A significantly shorter timeframe of diagnosis, a higher rate of jaundice, a lower daily body weight gain and longer duration of hospital stay were noted in the IHPS group prior to 3 weeks compared with those in IHPS group after 3 weeks. Eighteen were preterm infants. A significantly older age of symptom onset, a lower body weight at admission, more cases diagnosed by barium meal study and higher postoperative complication rates were noted in the preterm group versus full-term infants with IHPS.

- Rare cases neonatal IHPS



Pathophysiology

Editorials

New Insights in Infantile Hypertrophic Pyloric Stenosis

Raphael Udassin MD

Department of Pediatric Surgery, Hebrew University-Hadassah Medical Center, Jerusalem, Israel

Key words: pyloric stenosis, laparoscopy, atropine

IMAJ 2004;6:160–161

The last decade shed some light on the pathogenesis of infantile hypertrophic pyloric stenosis. IHPS is a developmental abnormality (erroneously called "congenital" in the past) in which the pyloric muscle hypertrophies after birth. There are sound data showing that the dimensions of the pyloric muscle at birth are the same in normal infants as in those destined to develop IHPS [6]. The hypertrophied muscle causes a time-dependent increase in mechanical obstruction; hence the clinical symptoms.

Pathophysiology

- Reversible developmental condition
  - Substance P ↓
  - Somatostatin ↓
  - Gastrin ↑
  - Atropine treatment prevents need for surgery in Osaka Japan in 21/23 infants. Normalization of pylorus at 4-12 months.

Macrolides

ADC

Are young infants treated with erythromycin at risk for developing hypertrophic pyloric stenosis?

Nitin Maheshwai

Arch Dis Child 2007 92: 271-273
doi: 10.1136/adc.2006.110007

At this stage, on the basis of published evidence, it can be concluded that young infants exposed to erythromycin in the first few weeks of life are at greater risk for developing hypertrophic pyloric stenosis. The highest risk seems to be in the first 2 weeks of life in term or near-term infants, and with courses of >14 days.

Azithromycin

Prenatal Prescription of Macrolide Antibiotics and Infantile Hypertrophic Pyloric Stenosis

William O. Cooper, MD, MPH, Wayne A. Ray, PhD, and Marie R. Griffin, MD, MPH

VOL. 100, NO. 1, JULY 2002

- Third trimester exposure

CONCLUSION: The hypothesized association between erythromycin and infantile pyloric stenosis was not seen. Causal inference from the association between prenatal nonerythromycin macrolides and infantile hypertrophic pyloric stenosis is limited by the small number of affected children and the evidence of other differences between users of nonerythromycin macrolides and controls. (Obstet Gynecol 2002;100:101–6. © 2002 by The American College of Obstetricians and Gynecologists.)

Conclusion

- We have presented a case of neonatal bilateral pulmonary cysts most probably due to very early aspiration of infected meconium liquor due to pyloric stenosis.

- To the best of our knowledge this is the first time that a possible association between maternal macrolide therapy in the second trimester and IHPS has been reported.

- **Not all lung cysts in a neonate are CPAM.**