

Diagnostic dilemmas of prenatal imaging in children with congenital thoracic lesions

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Use of fetal ultrasound for prenatal diagnosis of lung lesions

- Widespread screening prenatal US >20y

Close collaboration between:

- Ultrasound Unit, Helen Schneider Hospital for Women, Rabin Med Center (director: Prof. Israel Meizner) and Pulmonary Institute, Schneider's

CCAMs, sequestration, CLE, CDH, hydrops etc.

If need more information: refer to fetal MRI

The Use of MRI in antenatal diagnosis of fetal lung abnormalities

(in: Williams HJ, Paediatr Resp Rev 2002)

- Avoided during active organogenesis in 1st trimester
 - Effect of strong magnetic field?
- Ultra-fast MR sequences **eliminate movement artifacts**
- Homogeneous moderately high signal intensity on **T2 weighted MR sequences** (>chest wall muscle, < amniotic fluid)

● MRI Depicts

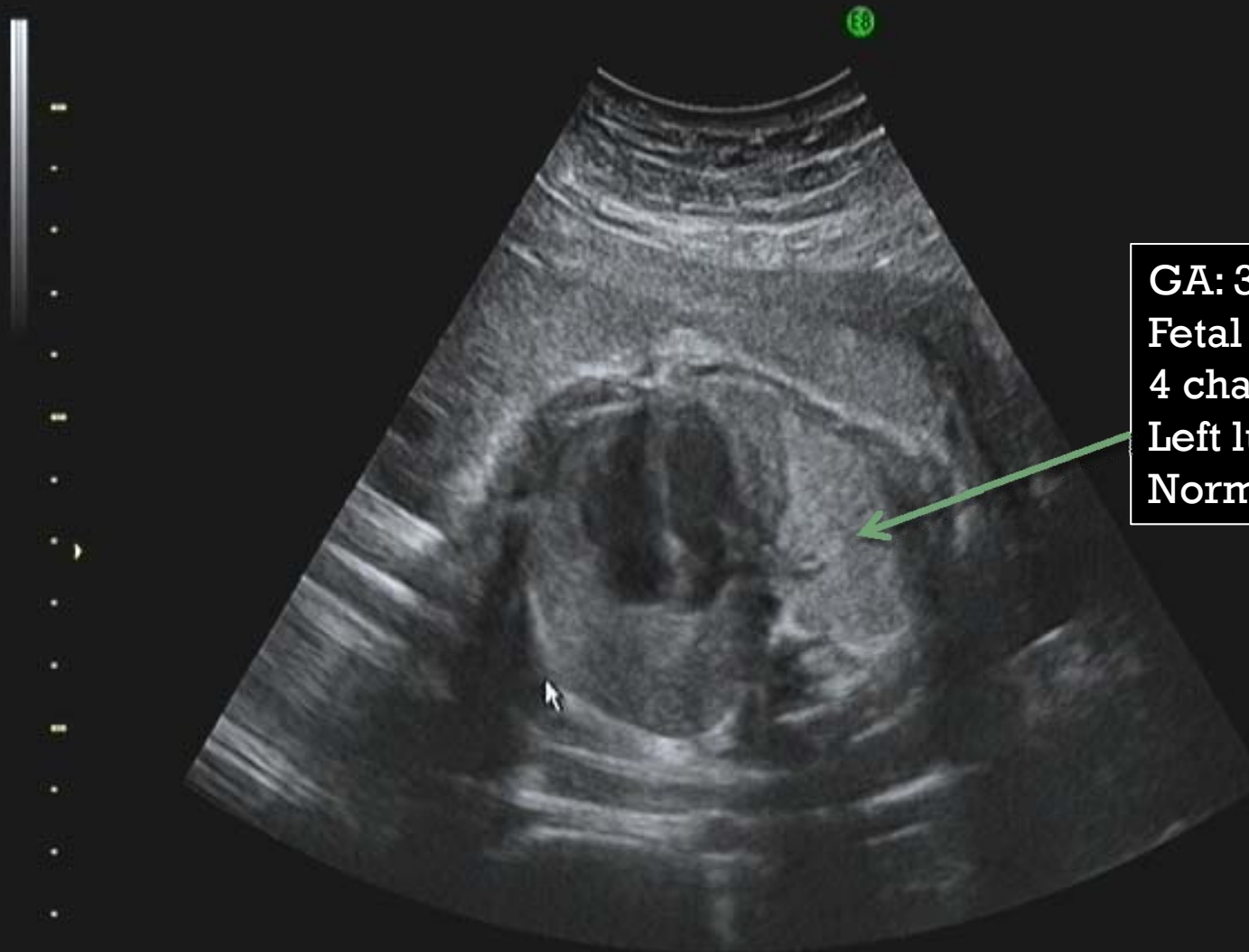
- Size and nature of lesion – CCAM? Sequestration? CLE? Hydrops?
- Relationships to other chest structures
- Lung volume assay – quantify hypoplasia
- Assay of lung maturation?

● Appearance

- Hypoplasia: lower intensity
- Lesions: often increased intensity
- Compare to liver

Case 1:

- VK, age 32y
- 31 wks pregnant, IVF
- Past termination of pregnancy- hydrocephalus
- Current pregnancy: Since 16wks, hyperechogenic lesion in LUL with deviation of mediastinum to the right
- LLL also hyperechogenic but less so.



GA: 31+1 wks
Fetal ultrasound:
4 chamber heart view
Left lung echogenic
Normal lung volumes

Referred
for MRI

Coronal Section



GA: 32+4 wks

“Left hemithorax much larger than right with marked hyperinflation of the left upper lobe. It is hyperintense, similar to amniotic fluid.

The mediastinum is pushed to the right
The right lung and LLL are hypo-intense similar to soft tissues and the placenta.

The trachea and bronchi are deviated to the right and filled with fluid.

The right lung may be hypoplastic.

The only part of the lung which appears normal although hyperintense and hyperinflated may be the LUL”

Voluson



D03457-11-08-03-4

RAB4-8-D/OB

MI 1.1

Belinson Medical Center

15.4cm / 1.1 / 29Hz Tls 0.1

03.08.2011 13:17:46

2+3 Trim.

Har-low

Pwr 100 Q

Gn 15

C6 / M7

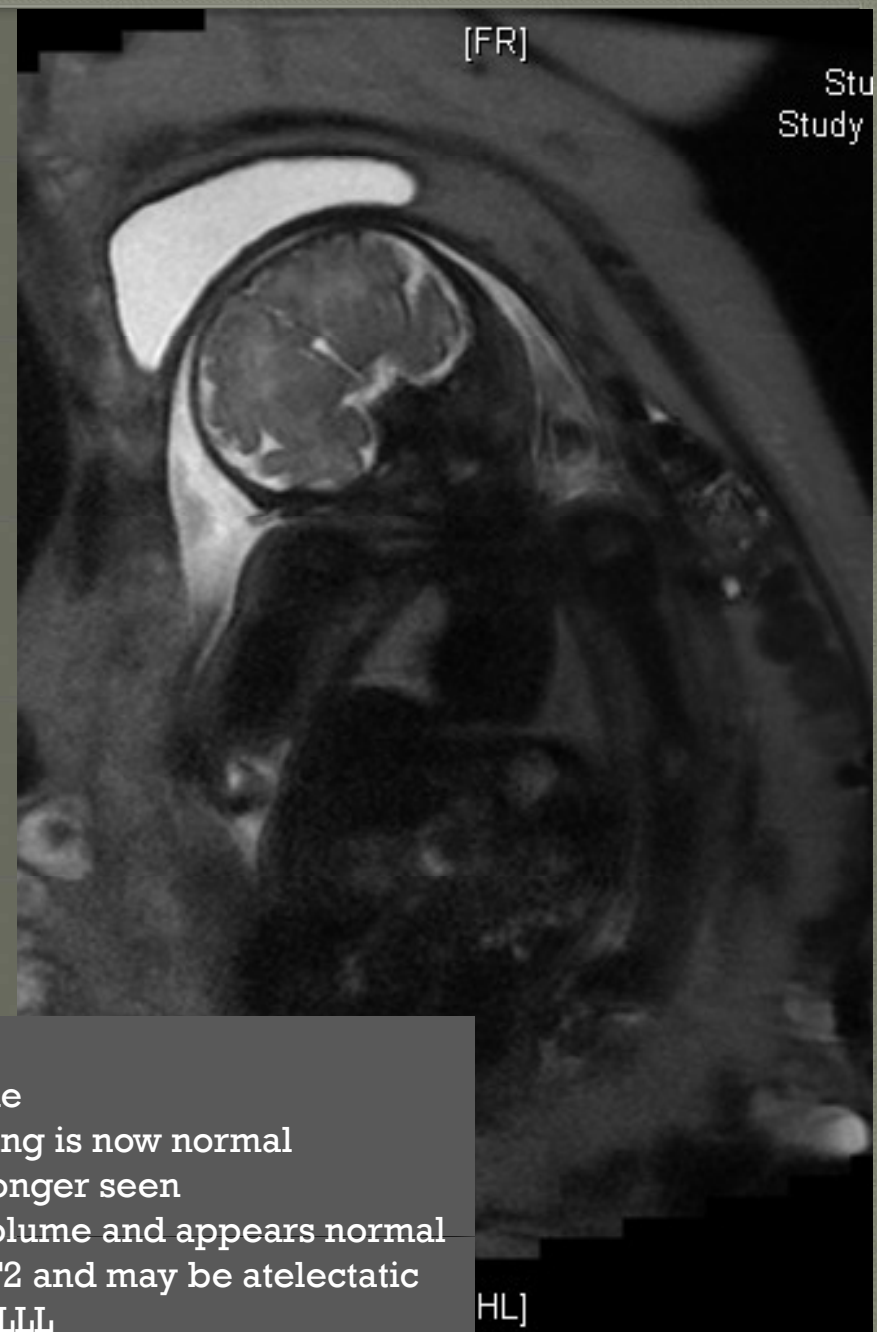
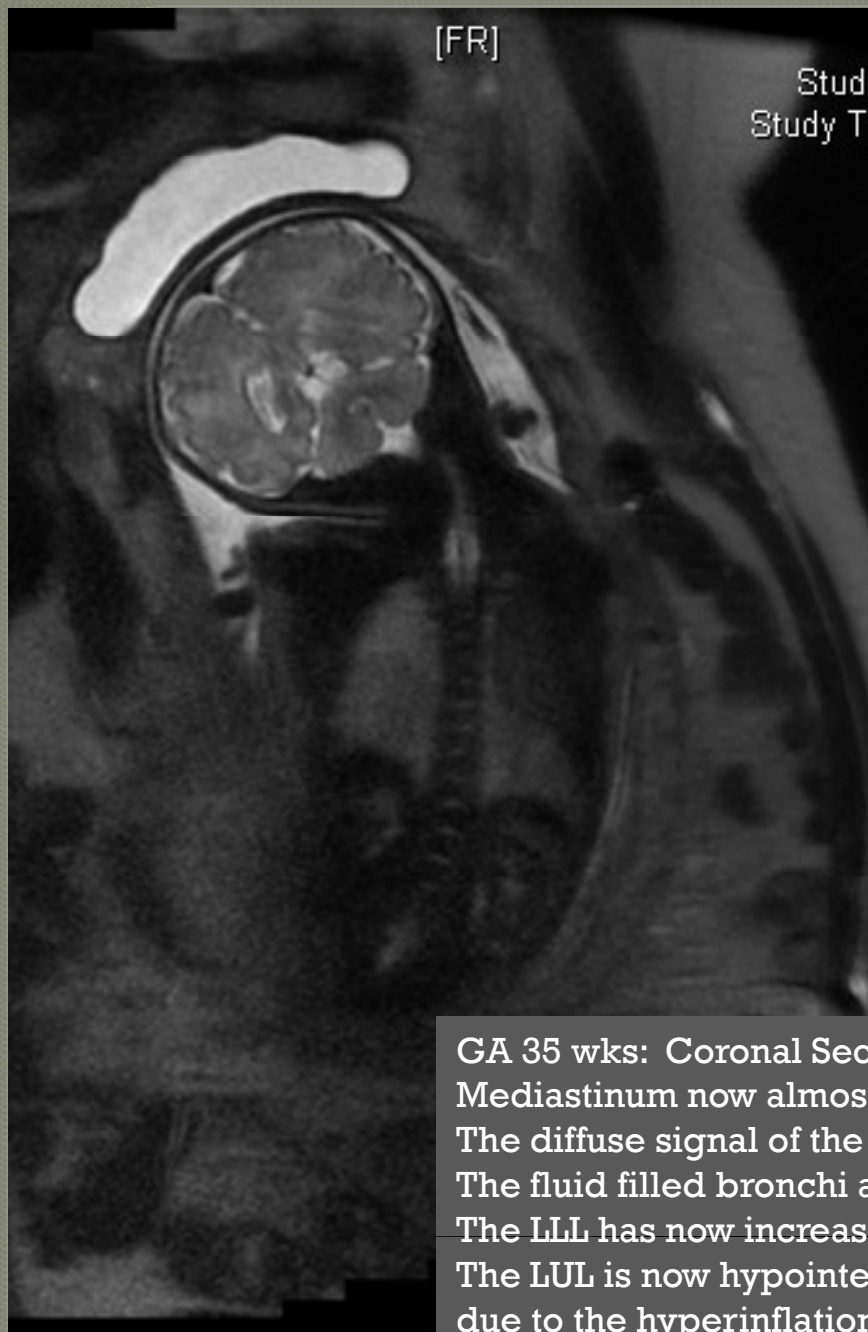
P2 / E2

SRI II 3



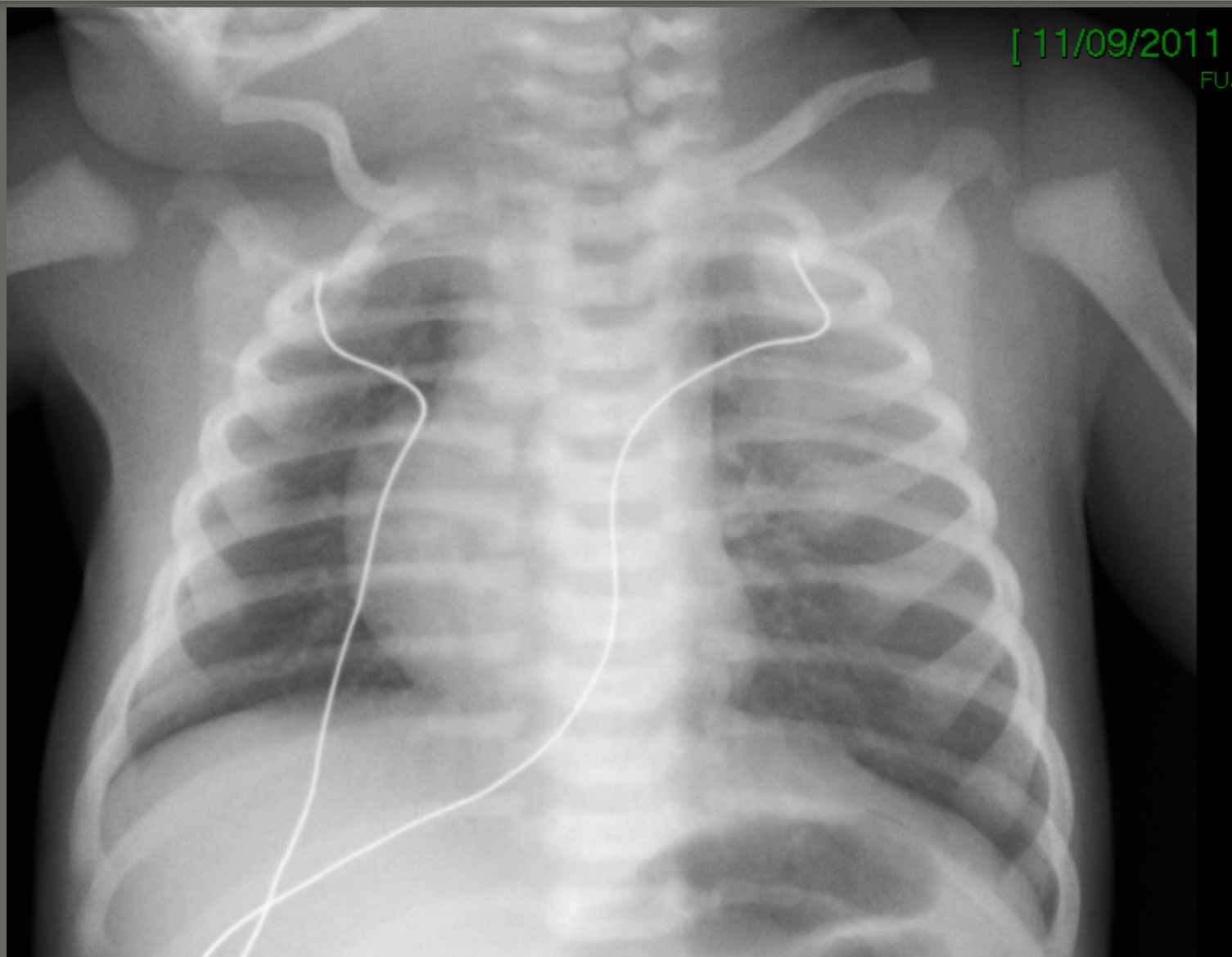
GA: 34 wks
Lung echogenicity
Almost normal

What is the next step?

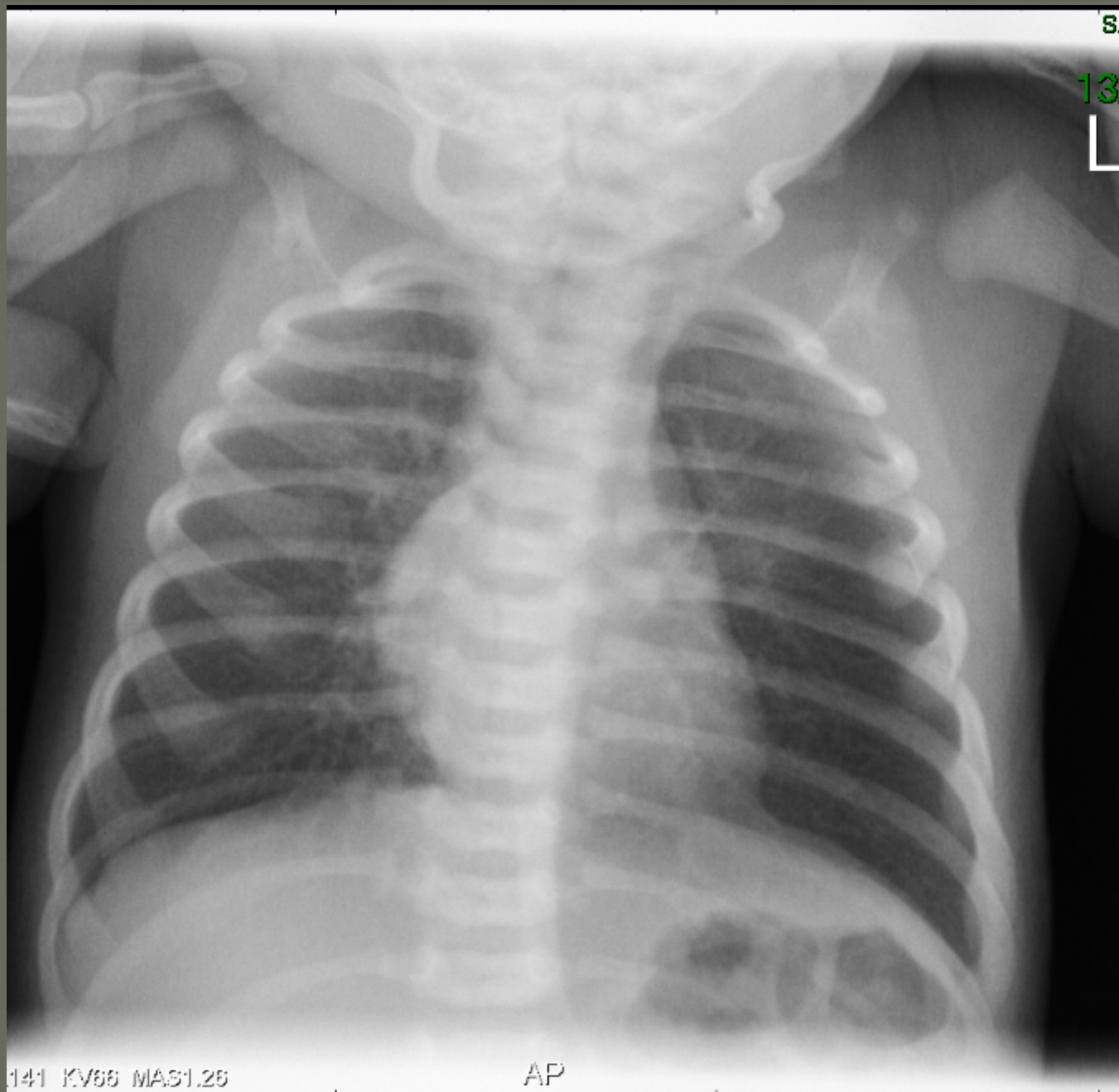


GA 35 wks: Coronal Sections:
Mediastinum now almost midline
The diffuse signal of the right lung is now normal
The fluid filled bronchi are no longer seen
The LLL has now increased in volume and appears normal
The LUL is now hypointense at T2 and may be atelectatic
due to the hyperinflation of the LLL

[HL]



At age 1 day: normal delivery. Asymptomatic



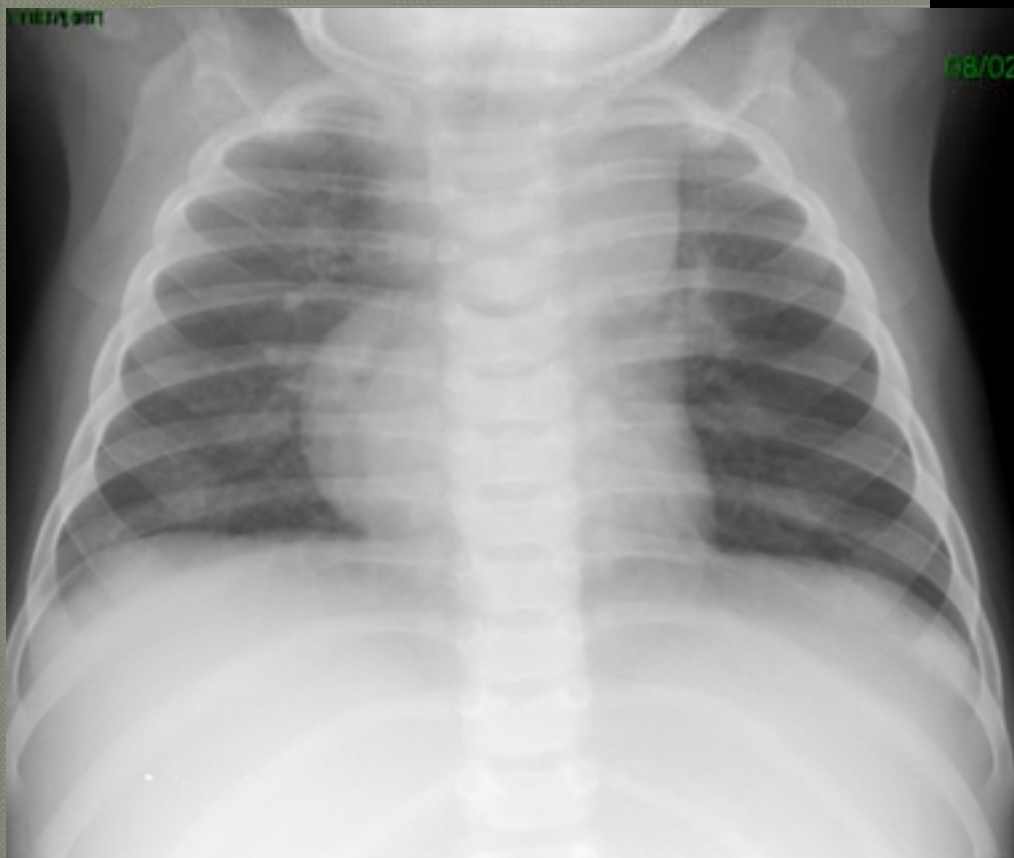
S.O.M.C.I - DIAGN

13/09/2011

L

141 KV66 MAS1.26

AP

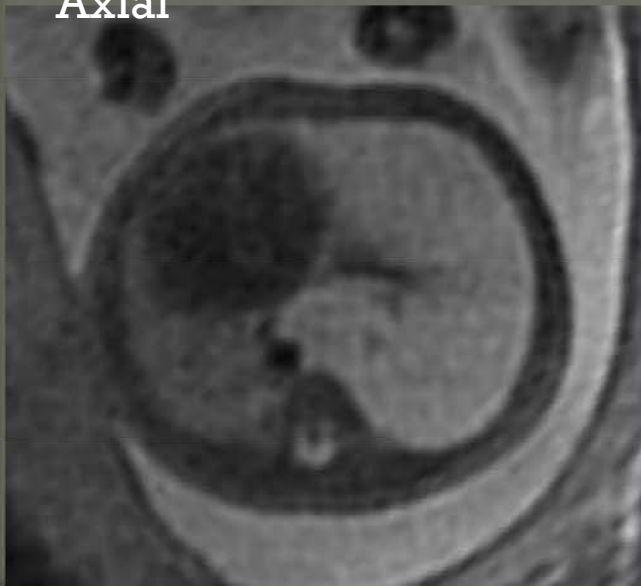


At age 5 months: Asymptomatic. No dyspnea. Thriving
Trachea and mediastinum deviated to the right
Hyperinflation, more on the left

Congenital lobar emphysema: appearance on fetal MRI. Yu-Peng Liu, Pediatr Radiol 2008

A water-overloaded lung lobe prenatally. MRI should differentiate from CPAM and sequestration – intact lung structure and stretched hilar vessels.

Axial



Coronal

Case 2: SL

11-14-2

9.4cm / 1.0 / 2.1Hz TIs 0.1

14.11.2010

Th



2944-10-10-14-5

17.2cm / 17Hz

TIs 0.1

14.10.2010 1

GE

1

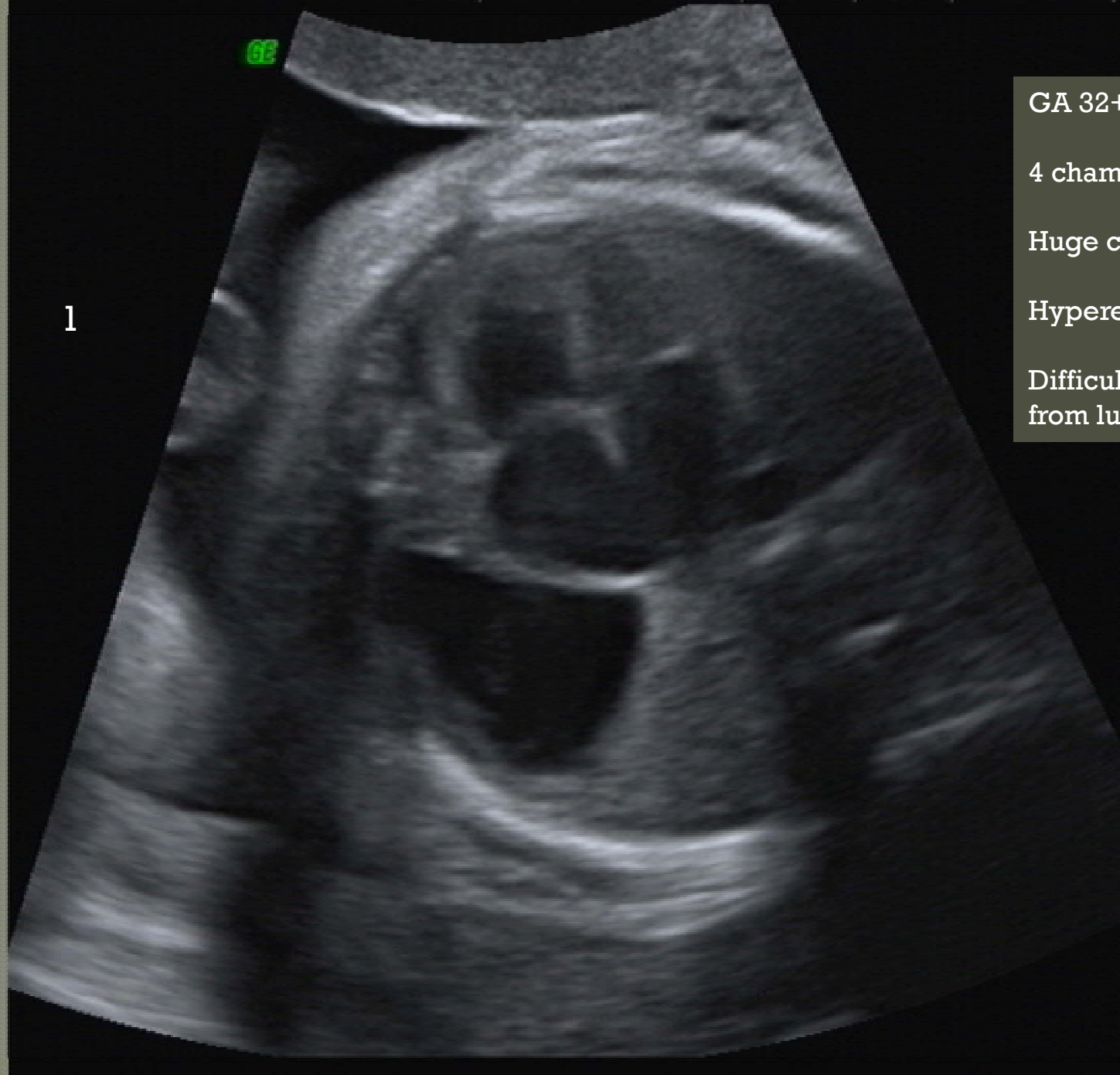
GA 32+1 wks

4 chamber view of heart

Huge cystic lesion, on right

Hyperechogenic lung

Difficult to separate lesion
from lung

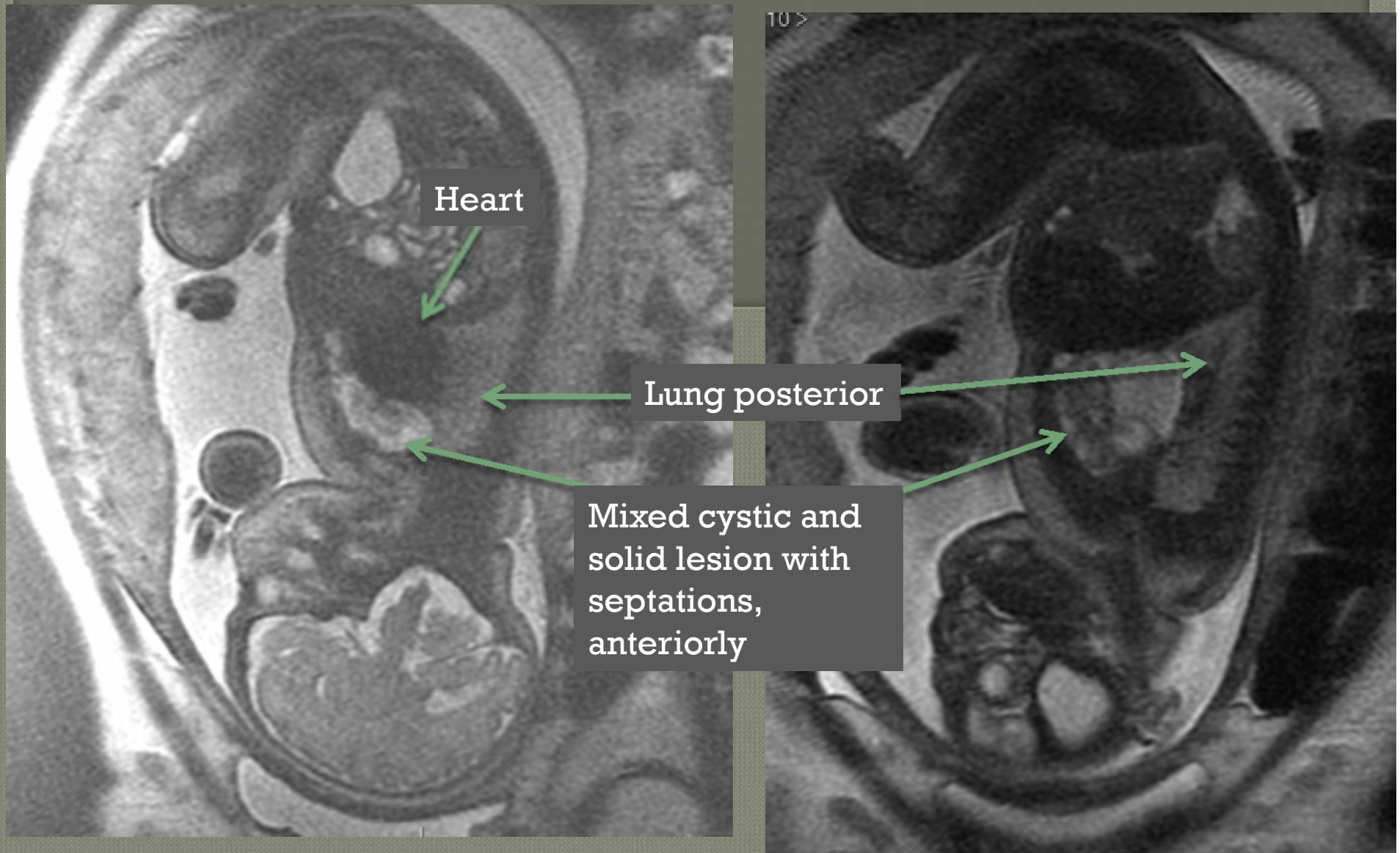


Heart shifted to the left by massive, cystic and mixed lesion

Lung is echogenic and may be compressed

GA 34 +4 wks

T2 Sagittal sections (fluids appear white)



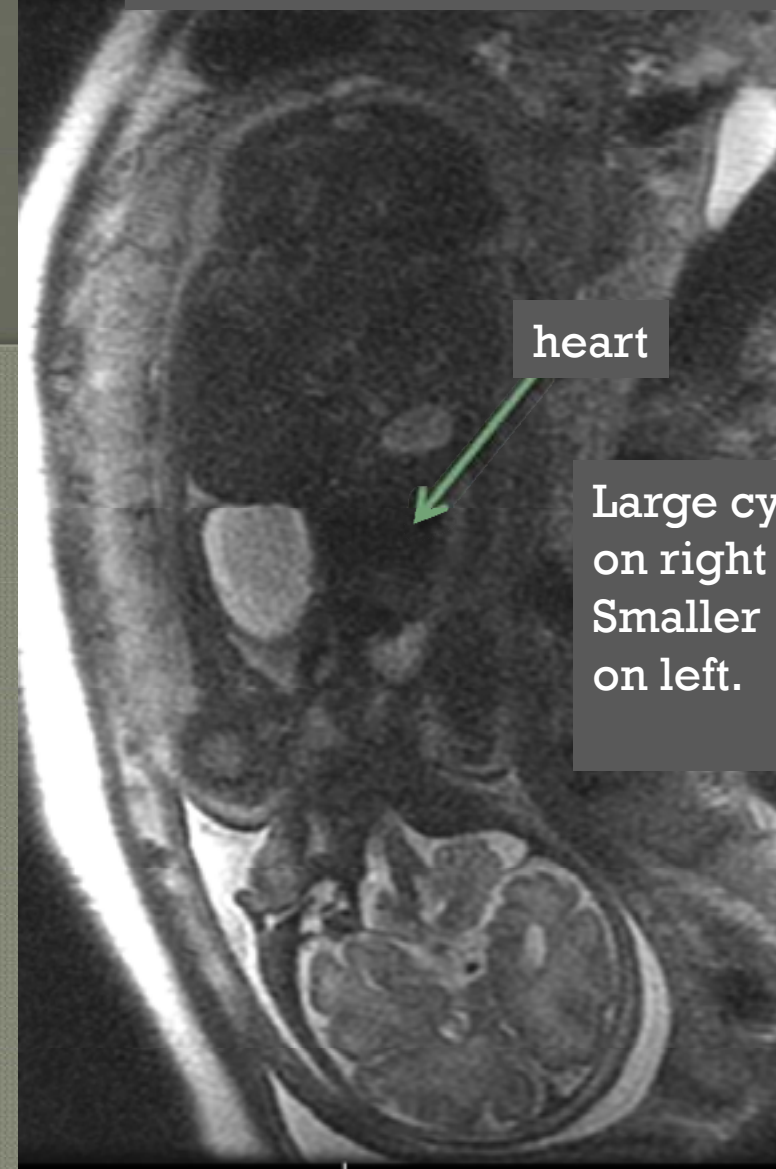
“Suspected CCAM”

T2 Posterior Coronal Section



Lungs well-developed bilaterally

T2 Anterior Coronal Section



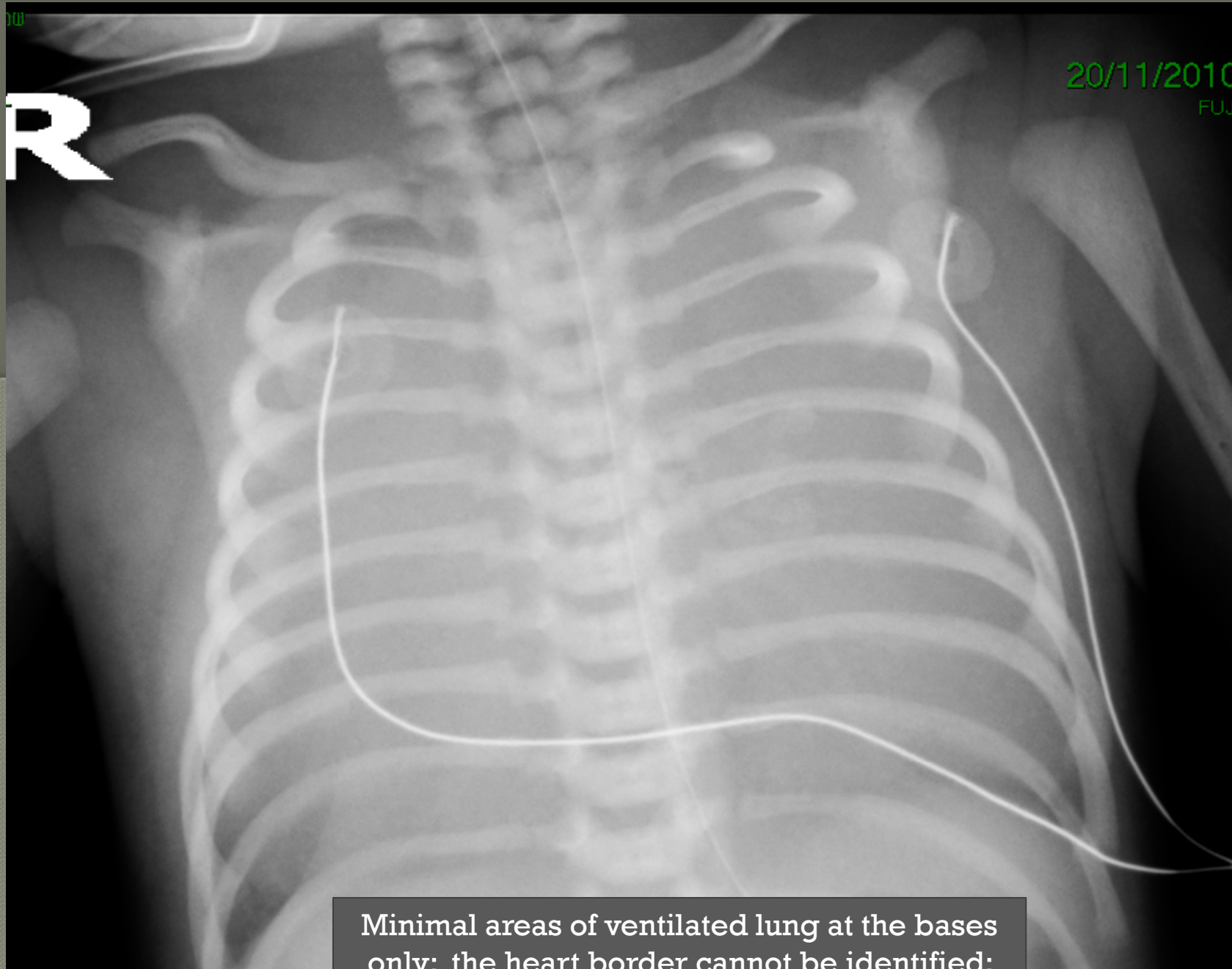
heart

Large cyst on right
Smaller on left.

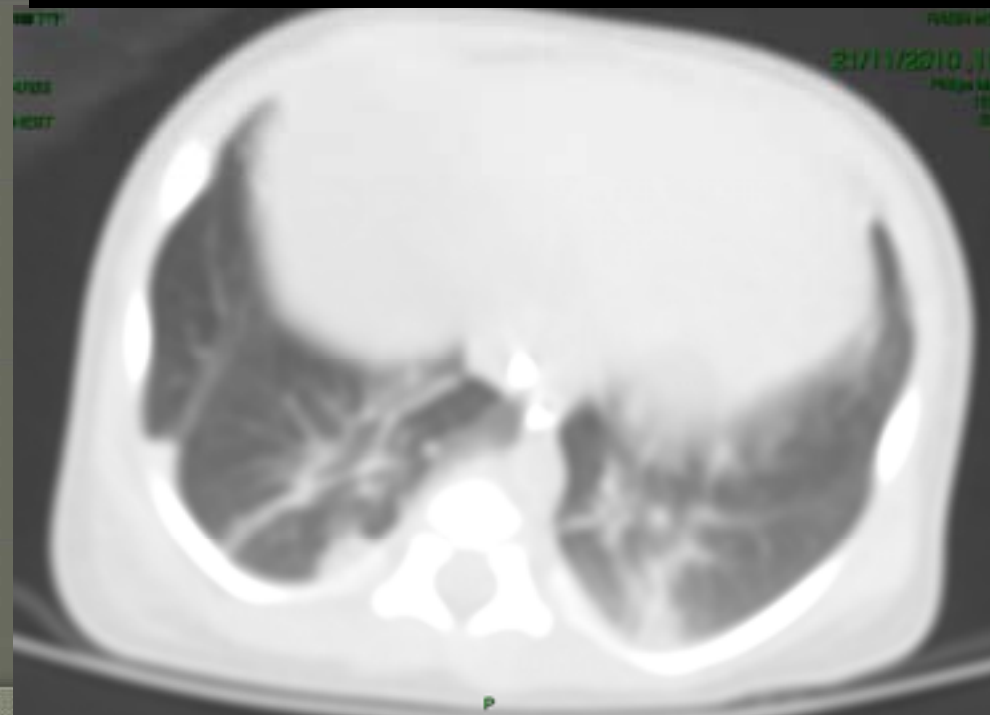
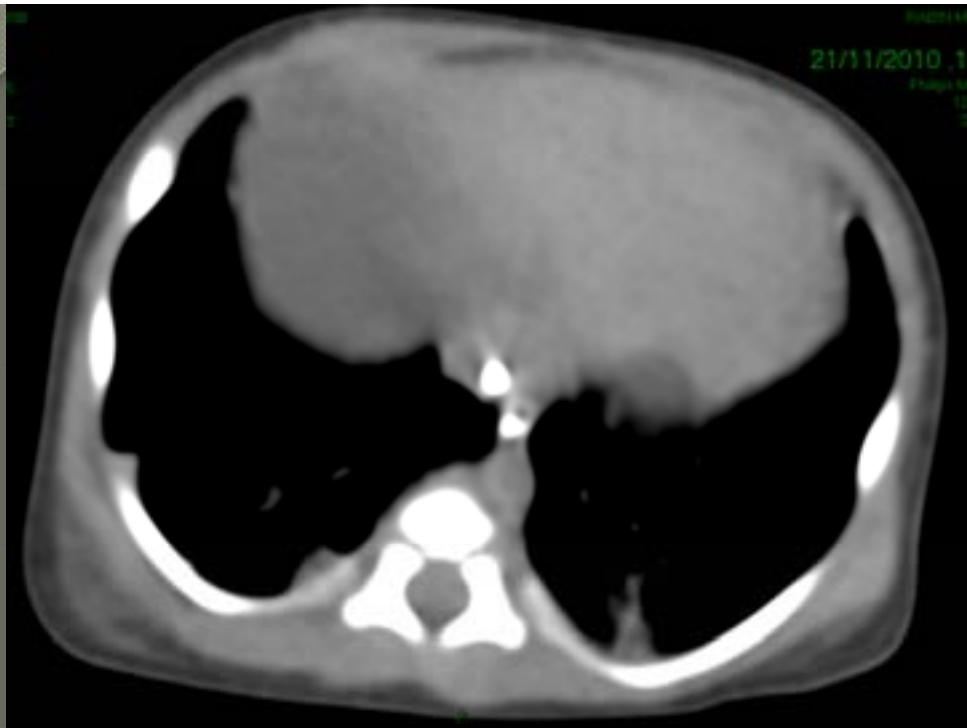
● Diagnosis?

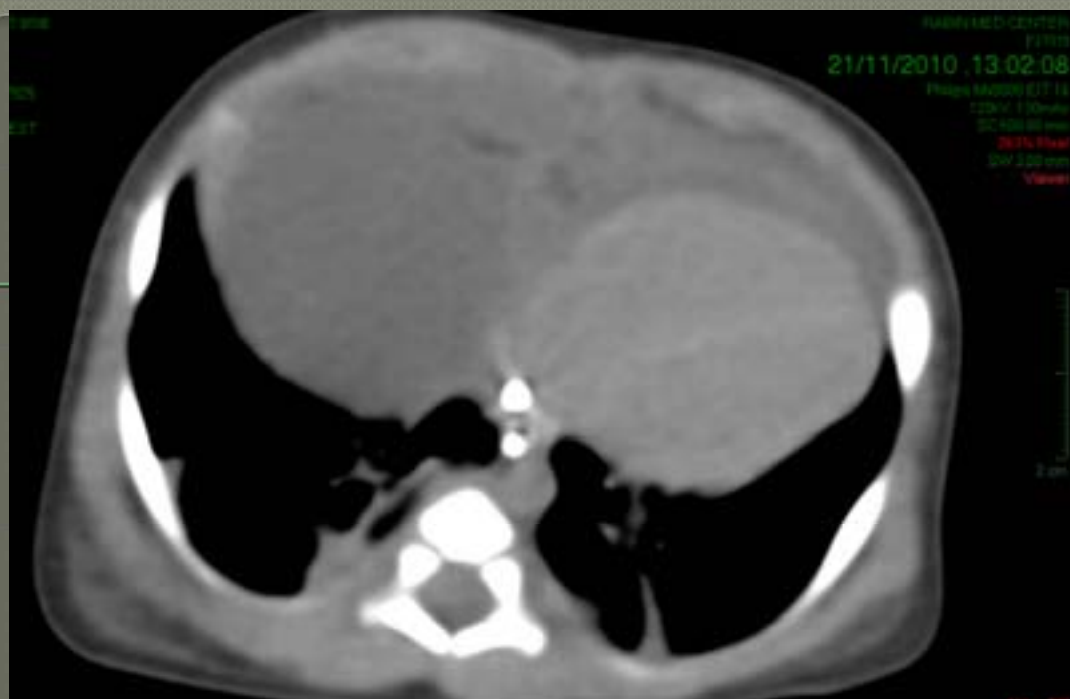
At birth:

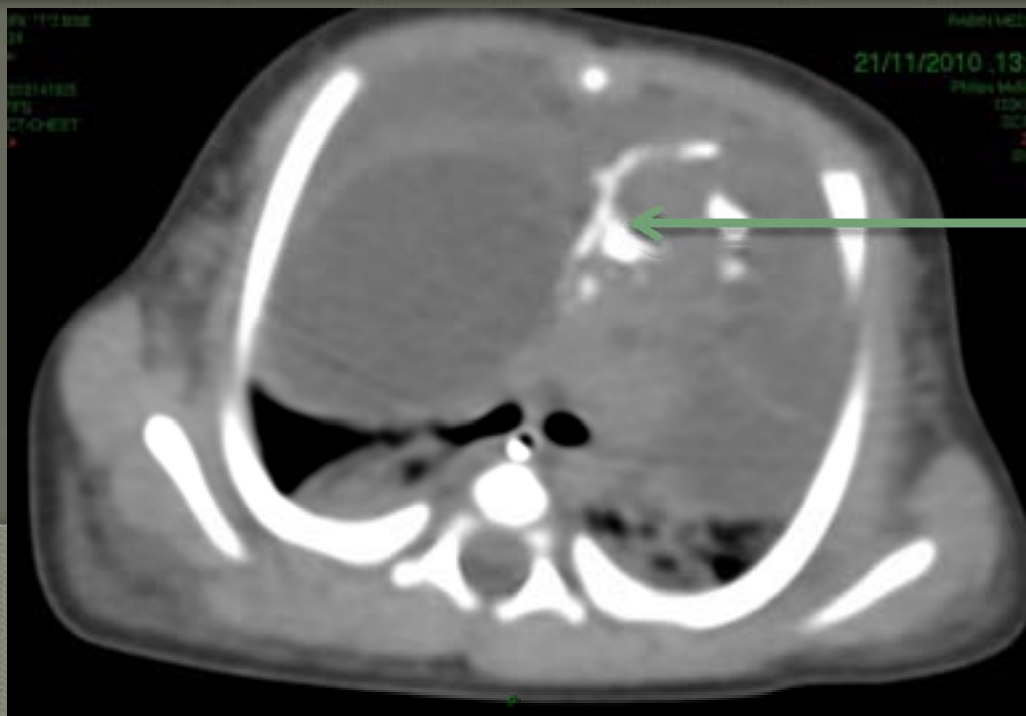
- Dyspnea, respiratory acidosis (pH 7.16, PCO₂ 68)
- Intubated and ventilated



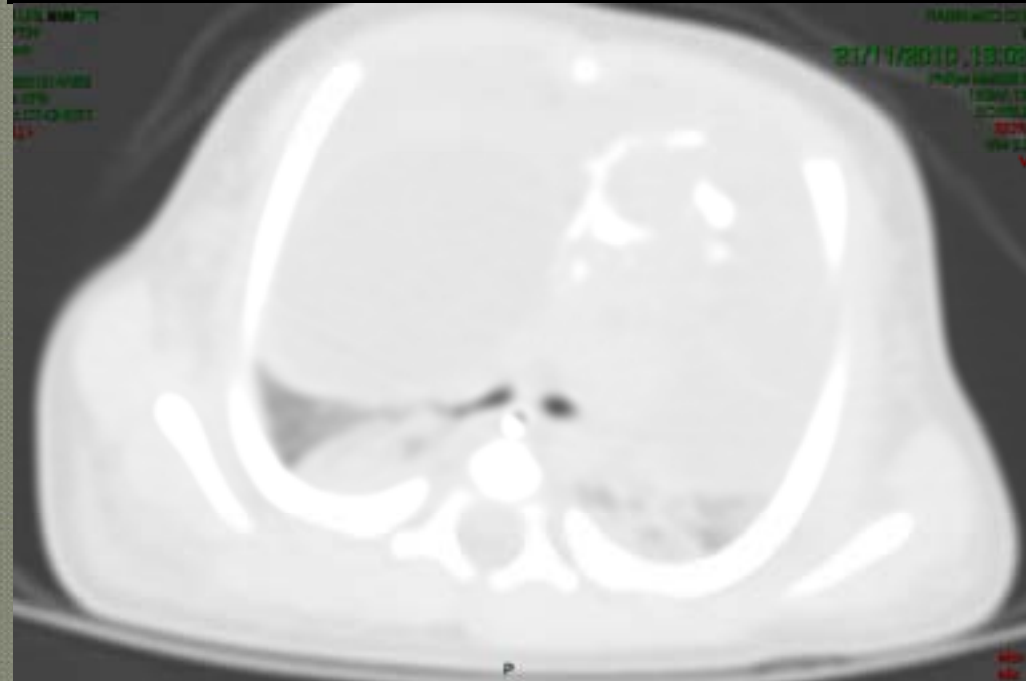
Minimal areas of ventilated lung at the bases only; the heart border cannot be identified; no pressure on the trachea

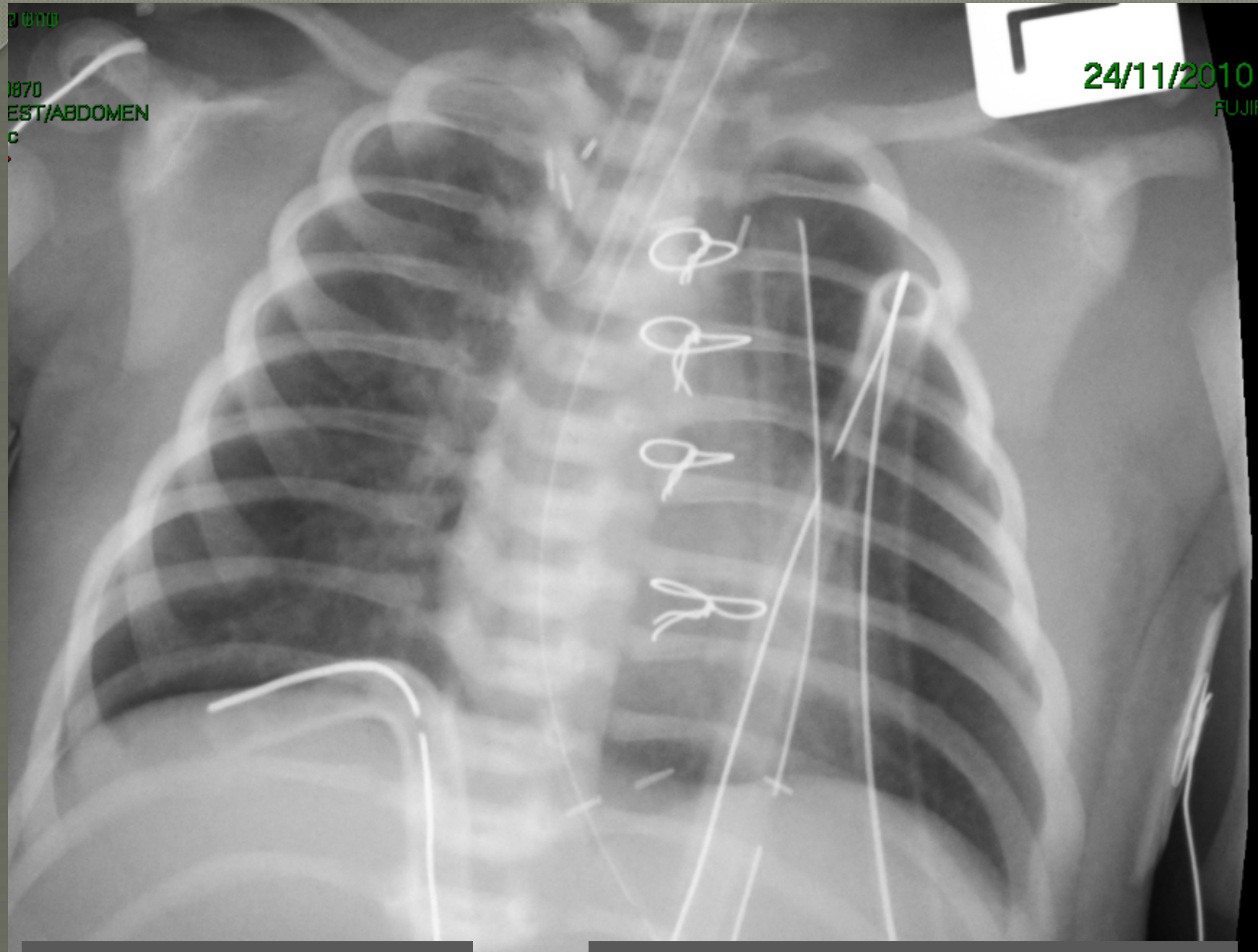






calcifications



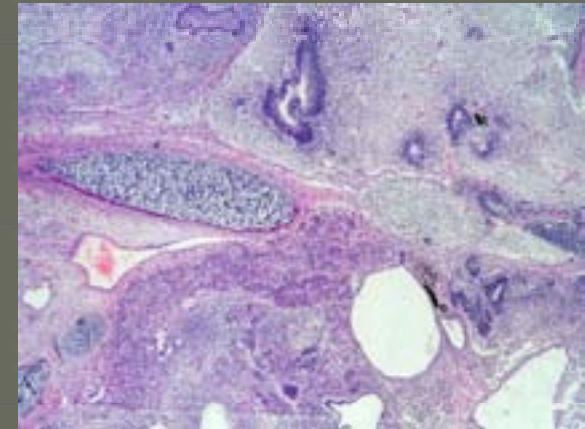


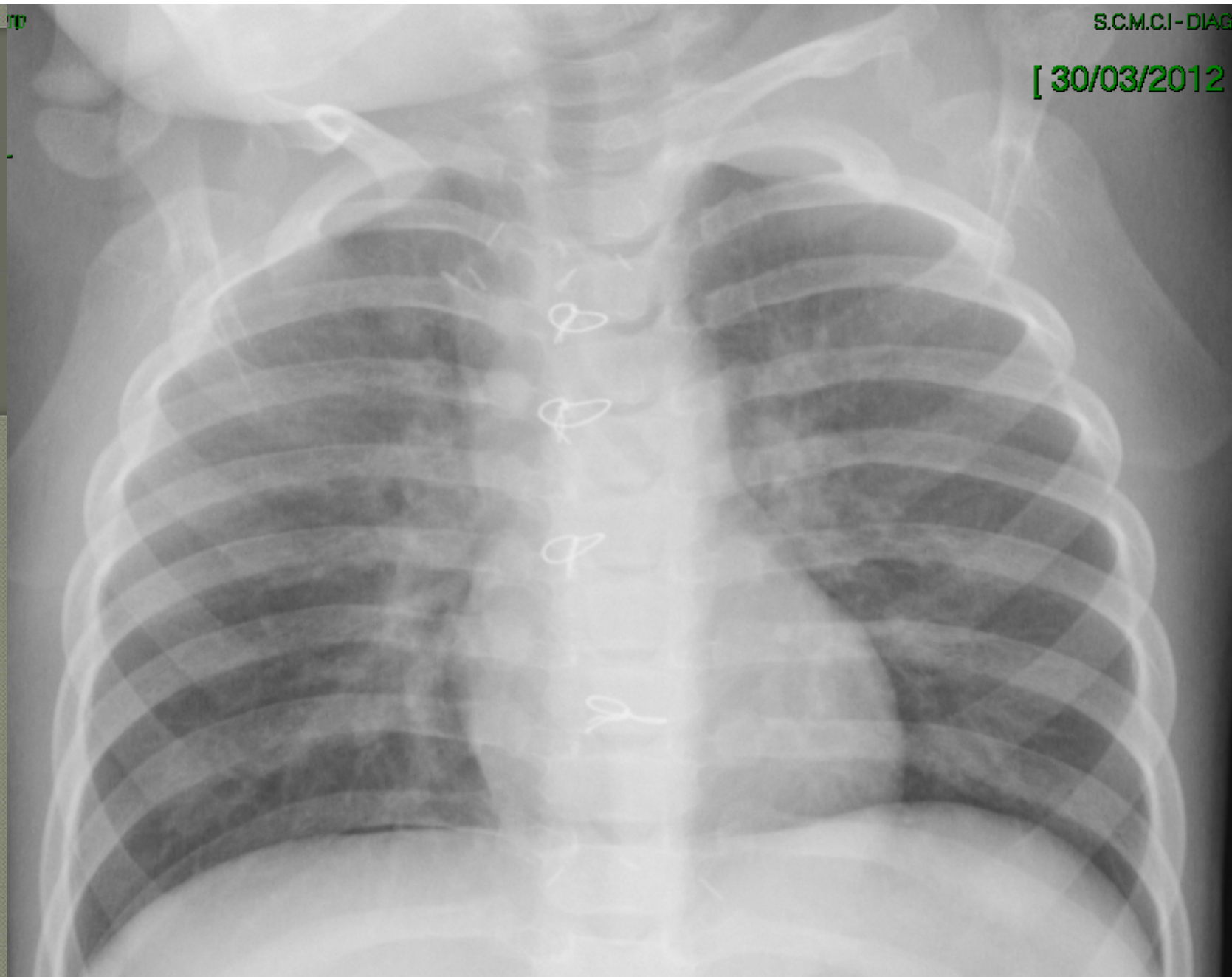
Lesion resected at 5 days

Mild pulmonary hypertension till 9wks

Pathology report: Immature teratoma

- diameter 7.8 cm
- Consists of all 3 elements:
- ecto, endo and mesodermal
- Ectoderm:
 - mainly immature neuroglial tissue
- Mesoderm: mature: embryonal ratio, 3:1
- Endoderm: mainly mature.
- No evidence of yolk sac tumor, embryonal carcinoma or other malignant tissue





At 18 mths: asymptomatic; normal pulmonary pressure; normal alpha-feto protein;
No evidence pulmonary hypoplasia or tracheomalacia. Continued oncology follow up

Neonatal Teratomas

- Multiple tissues from totipotent primordial germ cells?
- Neonatal: most are benign
- Histology: mature – contains mature elements e.g. skin, hair, fat, cartilage
- Immature: immature neuroectodermal elements
- Presence of microscopic foci of yolk sac tumor, rather than histological grade of immaturity predict recurrence
- Complete surgical excision is the treatment of choice
- Alpha feto protein is the tumor marker of choice re. residual or recurrent disease

Acute Respiratory Failure associated with Intrathoracic
Masses in Neonates. Takahiko S et al, J Pediatr. Surg 1999

- Operated at 8d. Giant mediastinal grade 2 immature teratoma. Post op uneventful

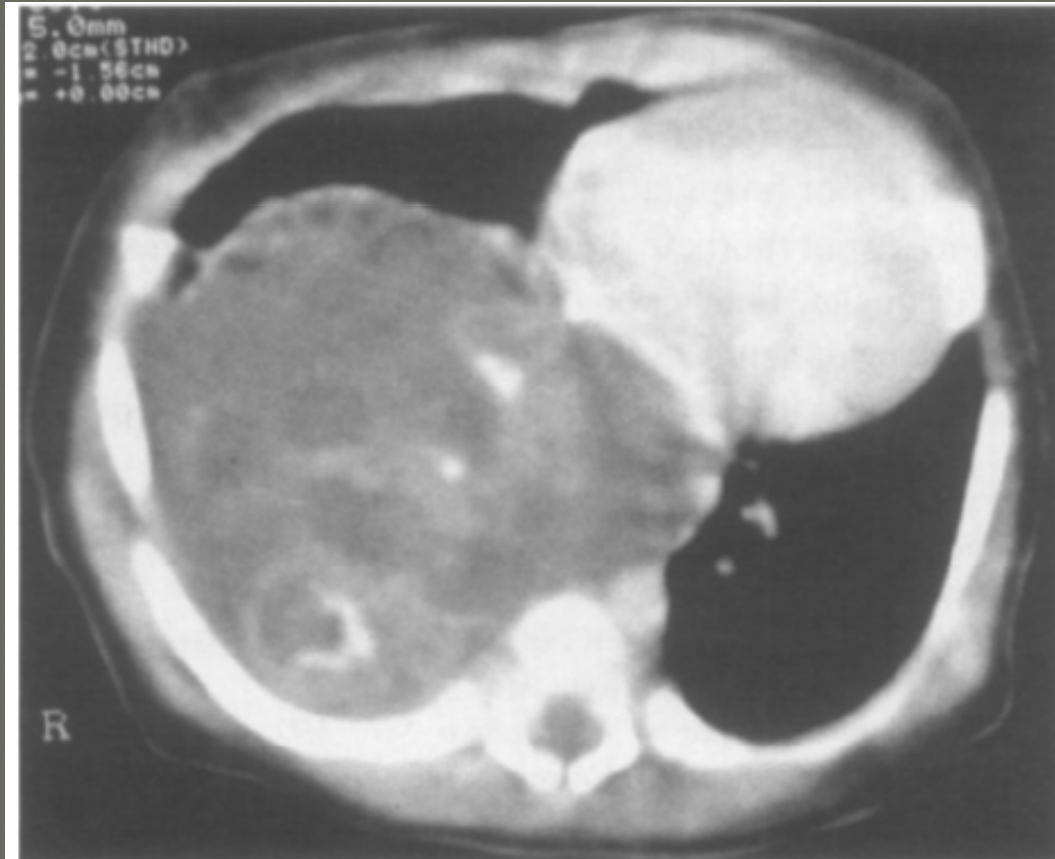
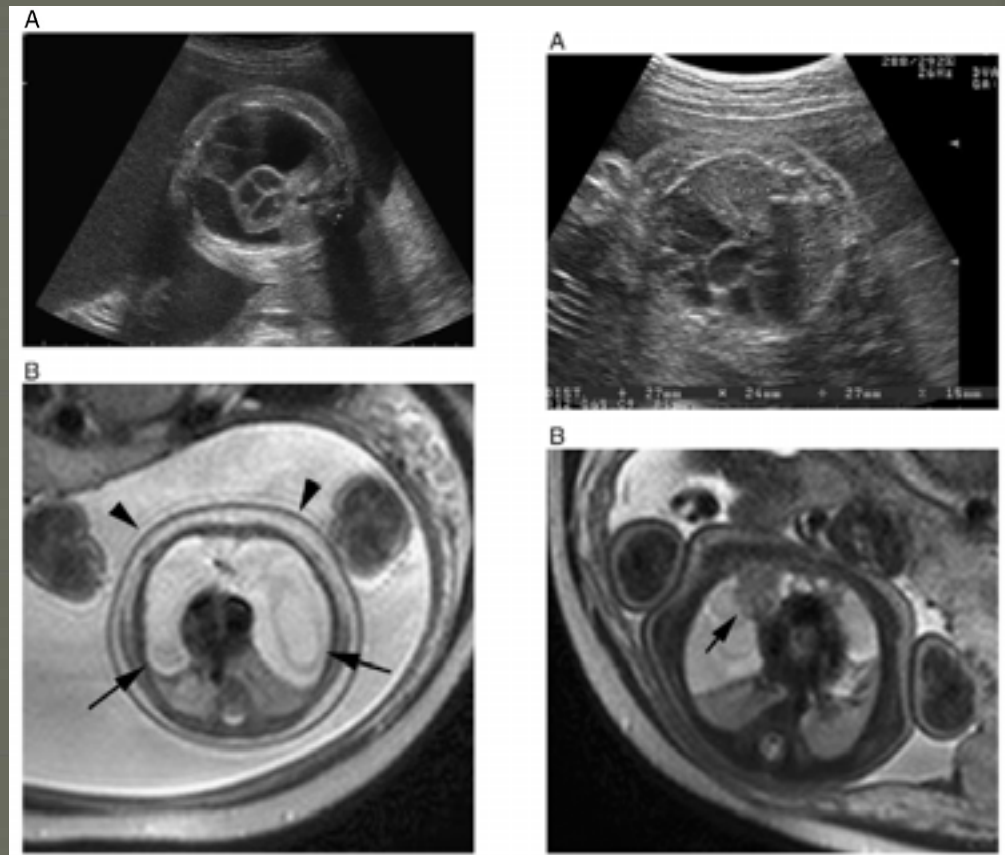


Fig 1. Chest plain CT in case 2 shows multiple cysts and calcification, which were indication of a mediastinal teratoma.

Mediastinal teratoma

- 20% of mediastinal masses
 - usually anterior
- 10% of teratomas
- If prenatal:
 - Often hypoplasia lungs, heart
 - hydrops
- This case: large cyst from 20w, hydrops
 - fetal cyst aspiration at 29w
 - (Takayasu H. J Ped Surg 2010)



Uneventful birth. Resected at 30d

Case 3: TN

4-09-01-11-2

15.4cm / 28Hz

Tls 0.1

11.01.2009 09:45:53 AM

1

2+3 Trim.

Har-low

Pwr 97 %

Gn -1

C7 / M7

P3 / E2

SRI II 3

63

GA 31+2 wks

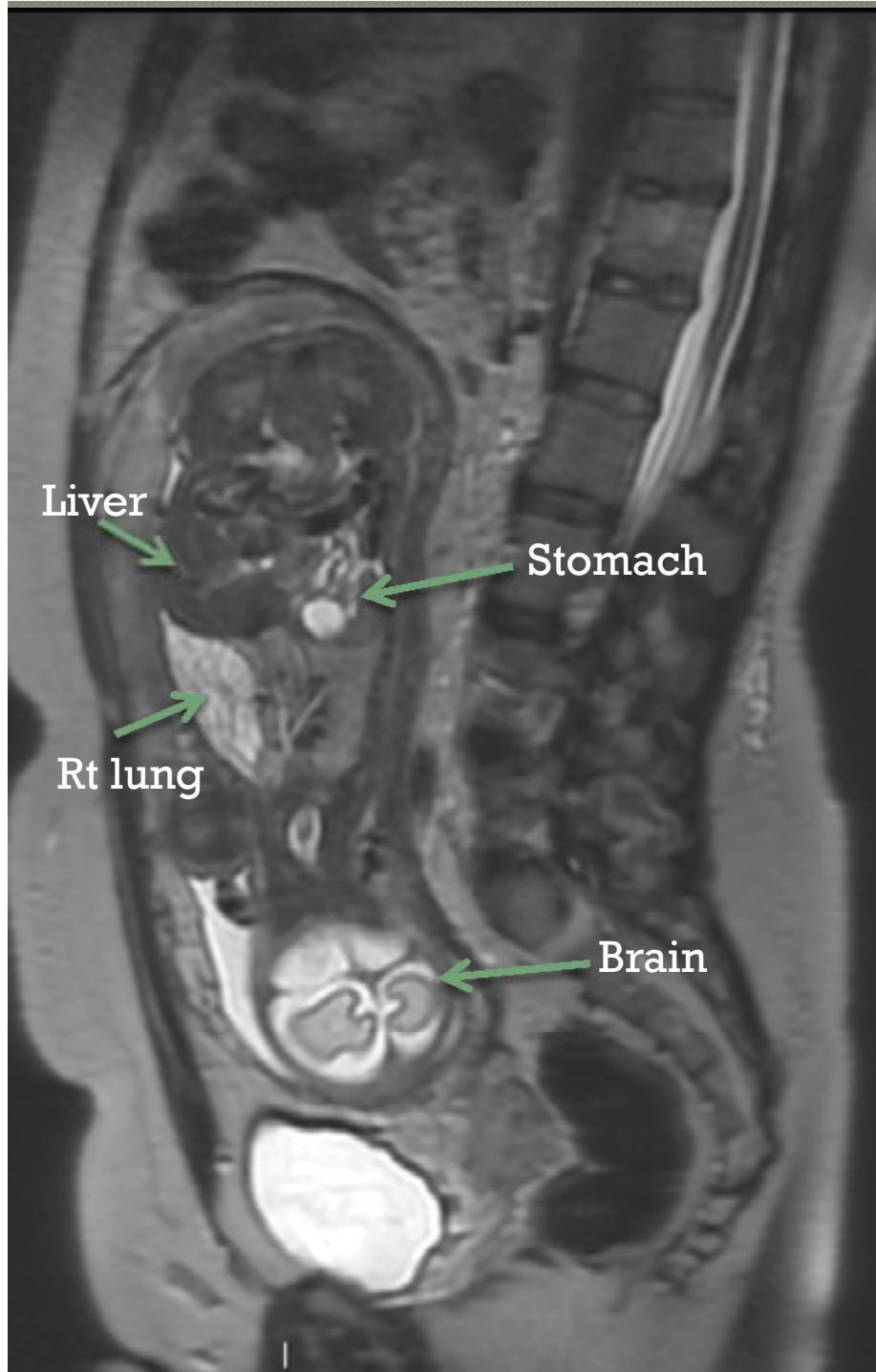
Hyperechogenic
right lung with
cysts



1 D 2.58cm

2 D 1.93cm





Central coronal section:

- main bronchi (white)
- Large hyperintensive lesion on right lung- many small fluid attenuated cysts, all 3 lobes; some foci of normal lung tissue on the right

No evidence of mediastinal deviation

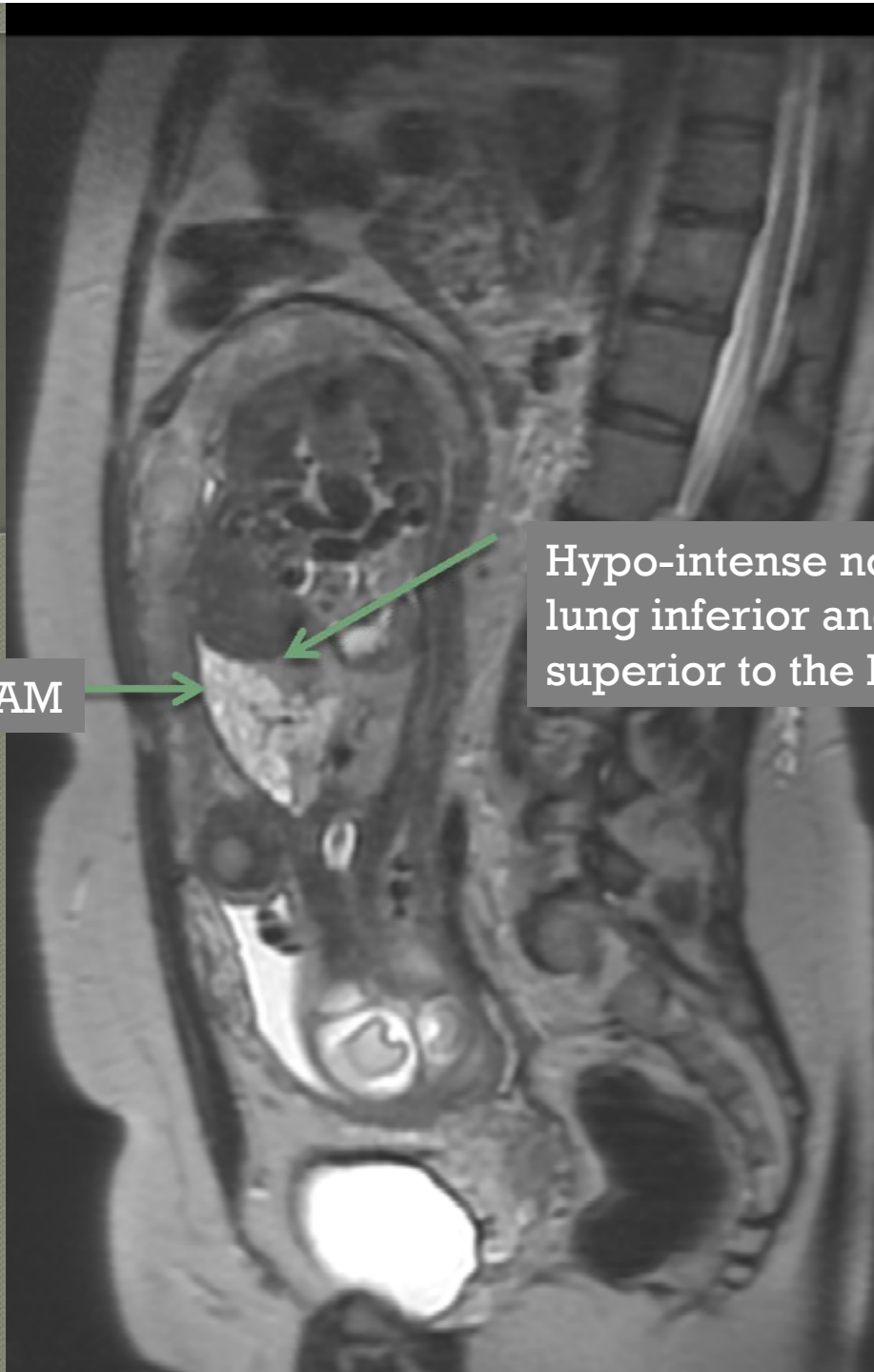
The left lung signal is slightly lower than usually seen but still within normal limits.

M/P Large CCAM on rt

Coronal section

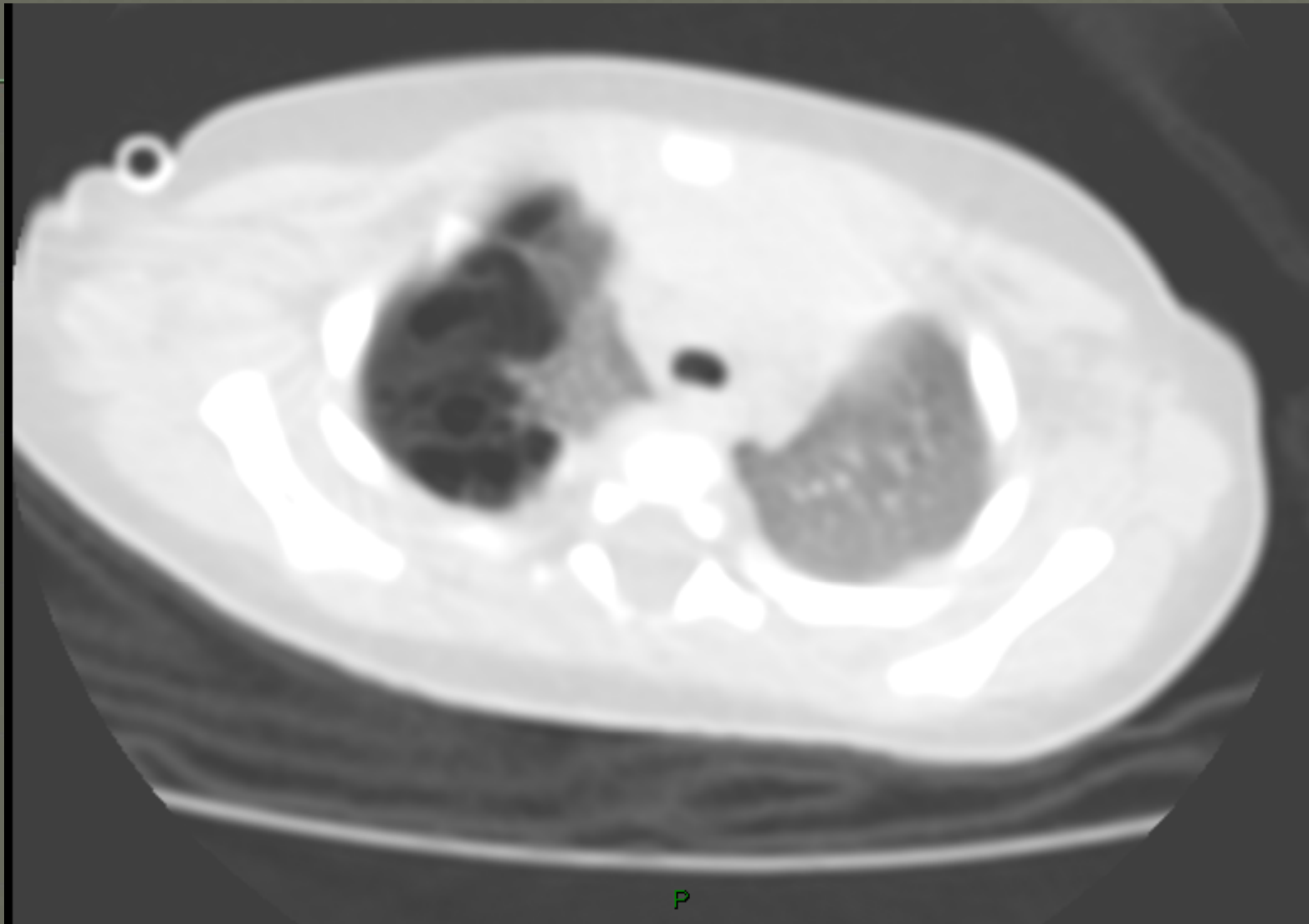
Type II CCAM

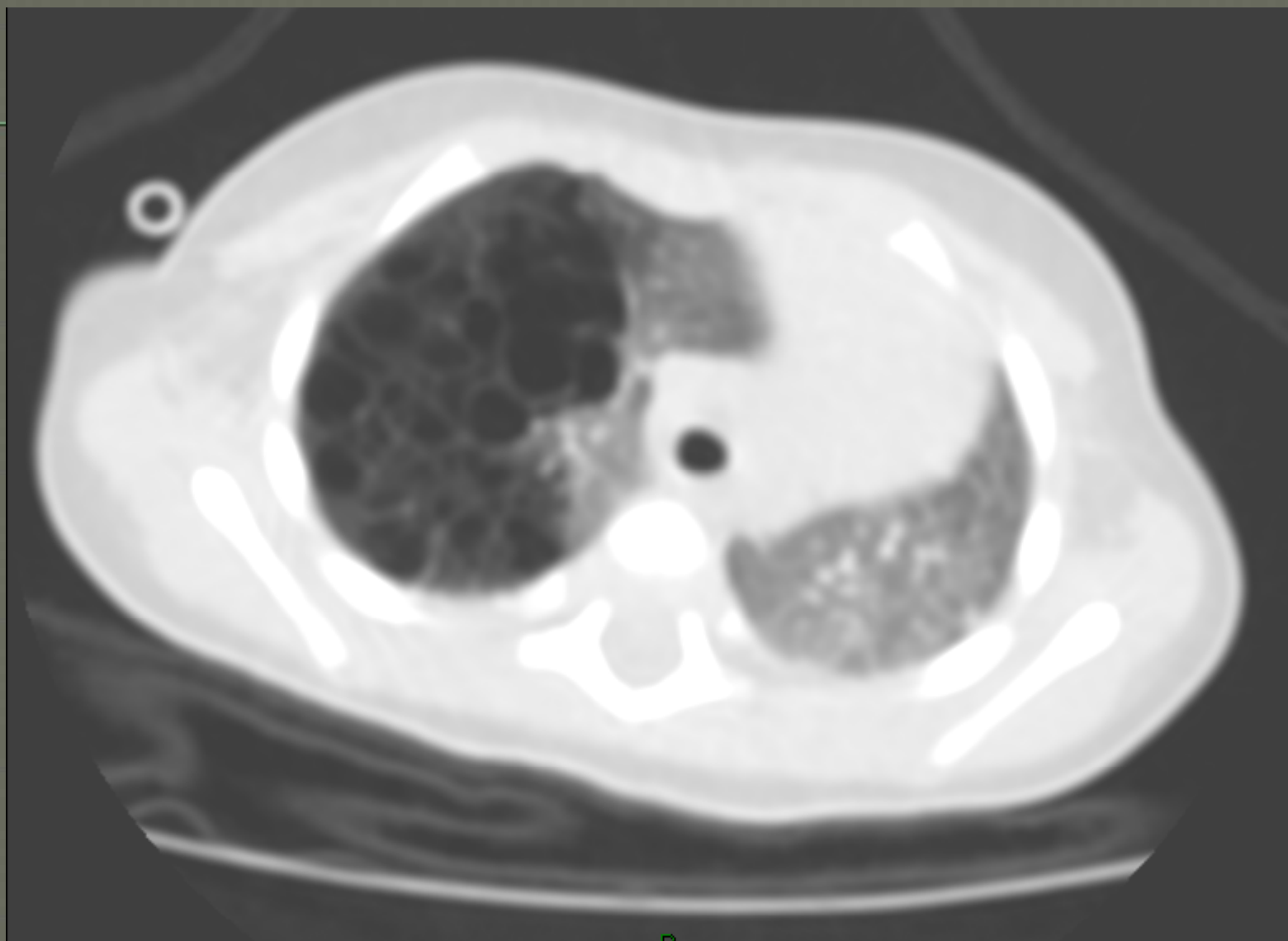
Hypo-intense normal
lung inferior and
superior to the lesion

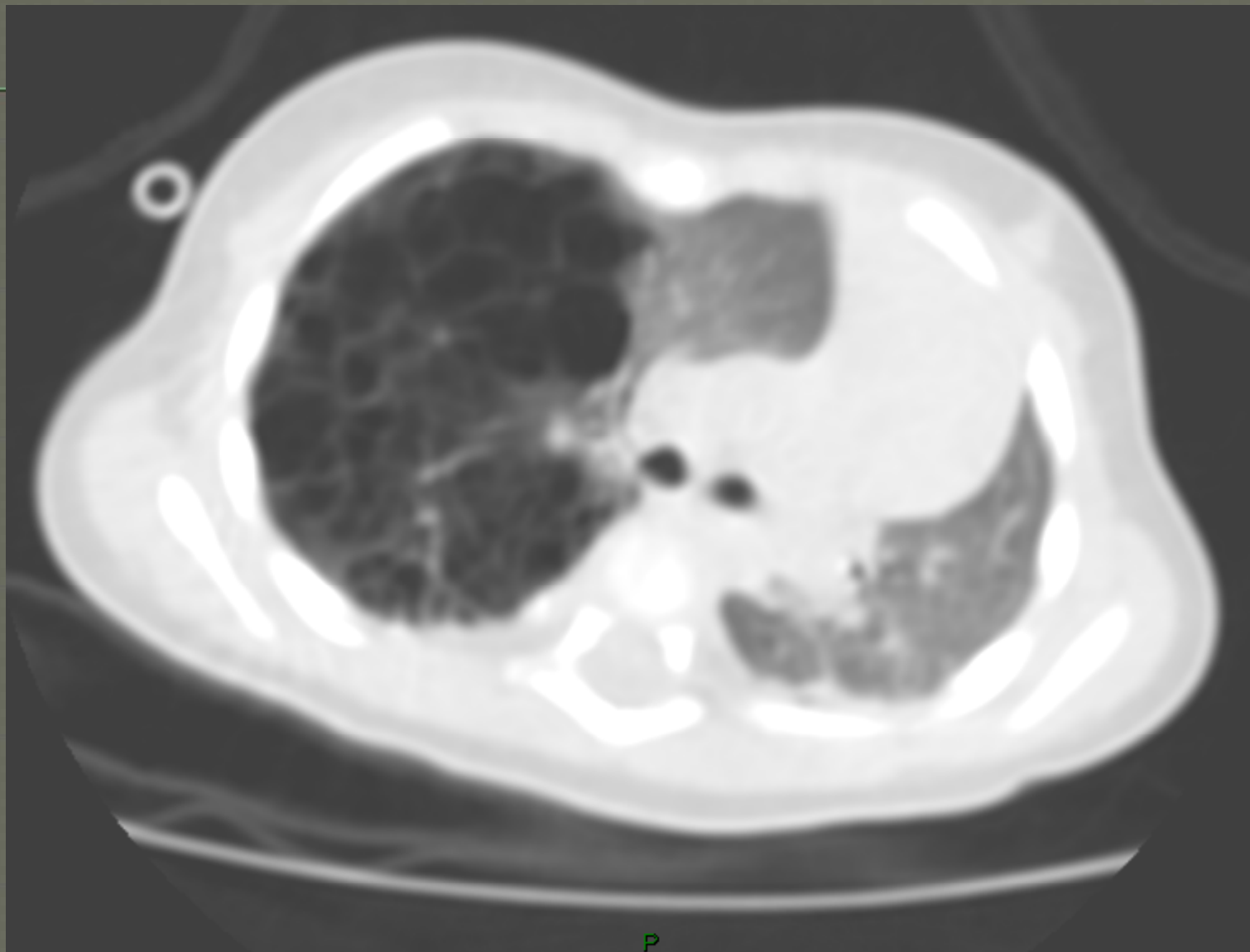


TN

- Born 37w:
- large cystic lesion rt lung.
- Very mild tachypnea till 4mths. Thriving. Normal saturation and cardiac echo
- Gradually increasing dyspnea and hypoxia
- At 5ths: removal of large right CCAM
- (RMPA cut and anastomosed with decreased perfusion; dilated at catheterization)

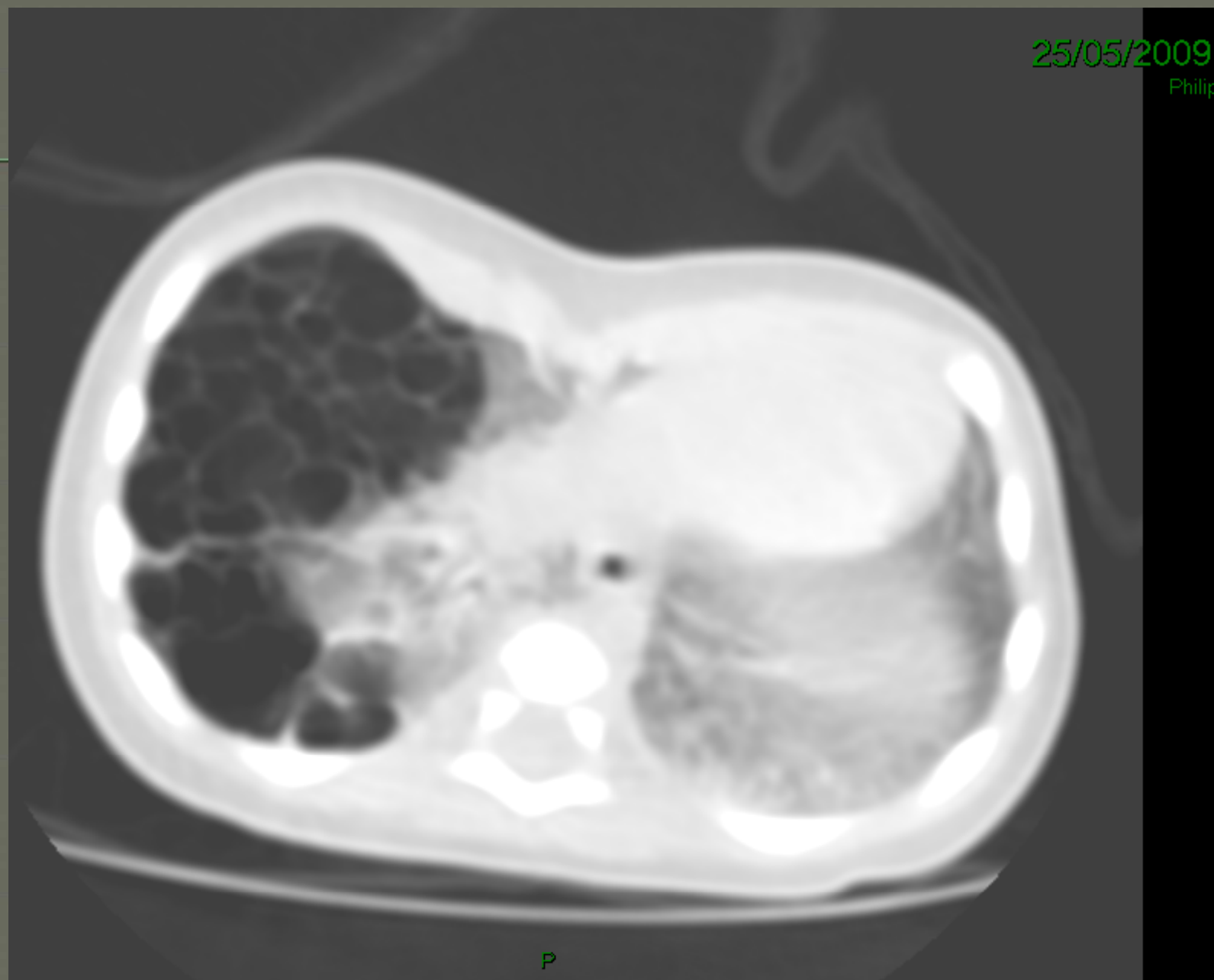




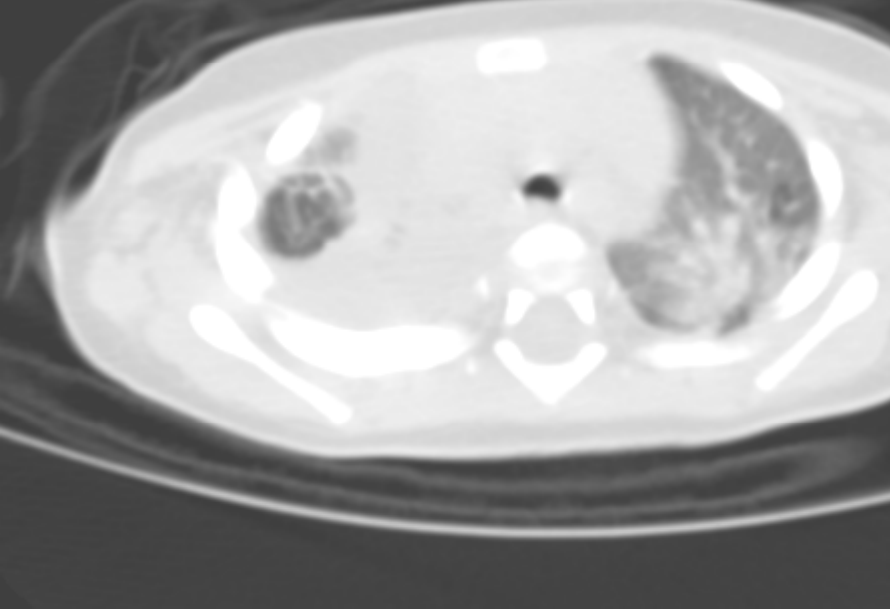
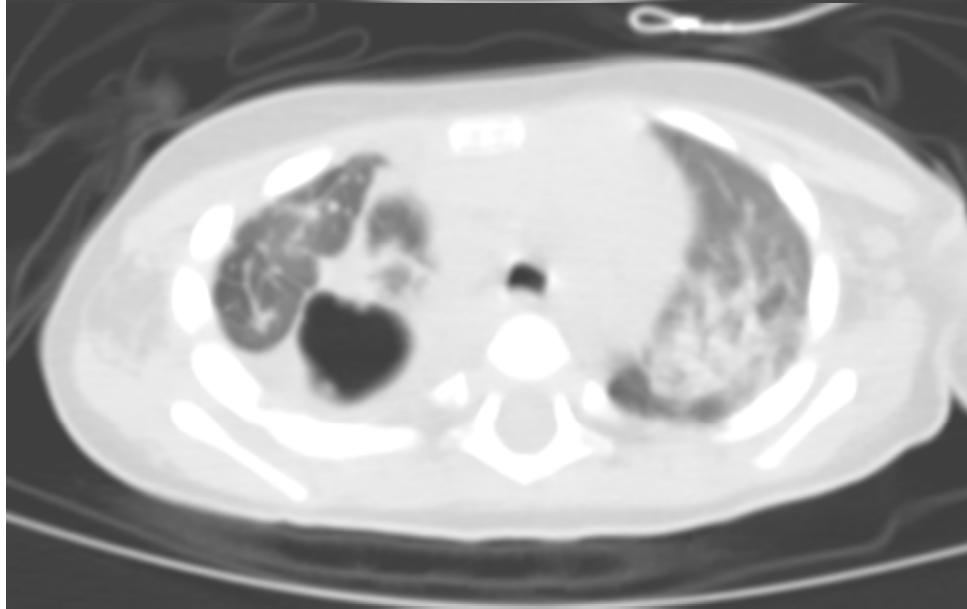
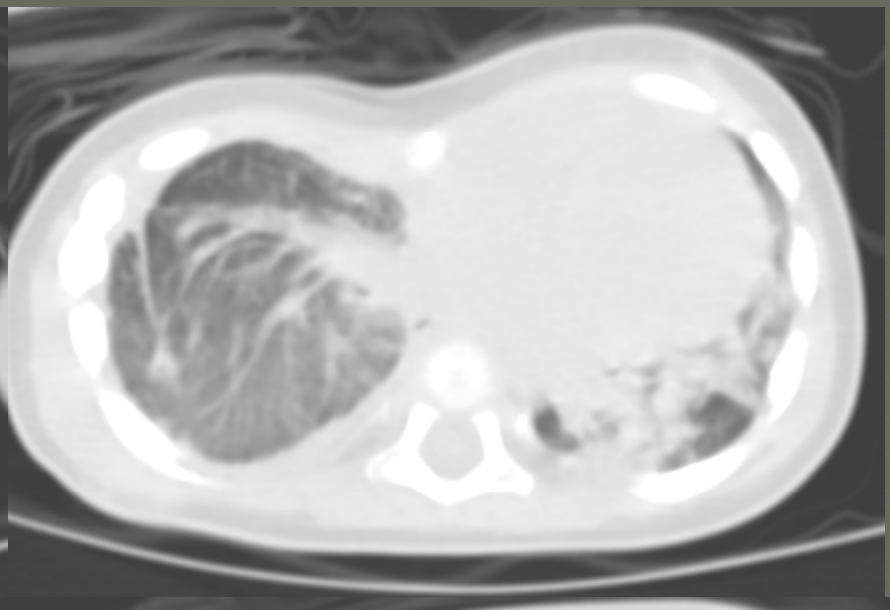
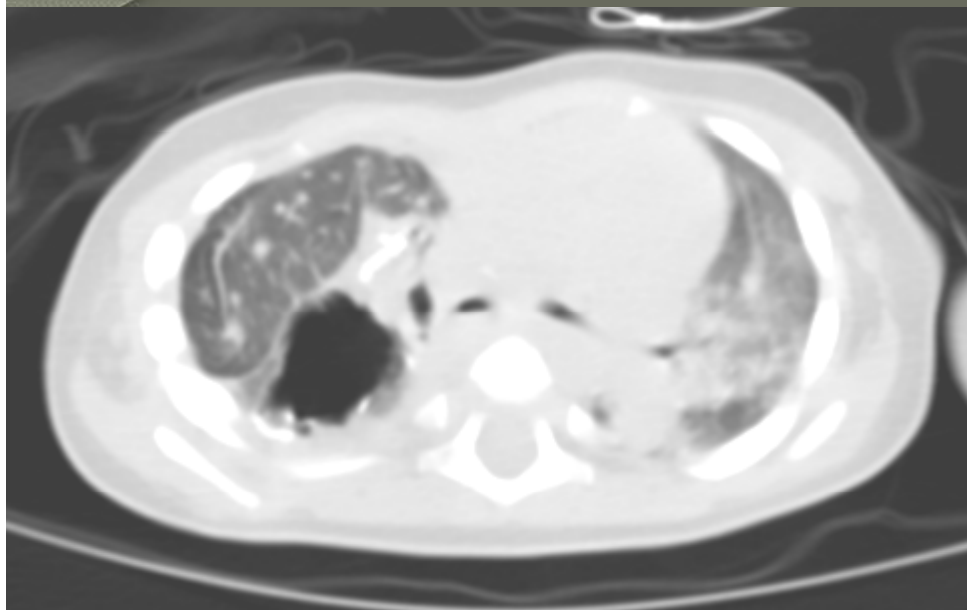


25/05/2009

Philips



P



Lossless

SCHNEIDER HOSPITAL
Marconi Medical Systems, NM Division IRIX
[29/10/2009 ,08:38:31]

258% Pixel
Viewer

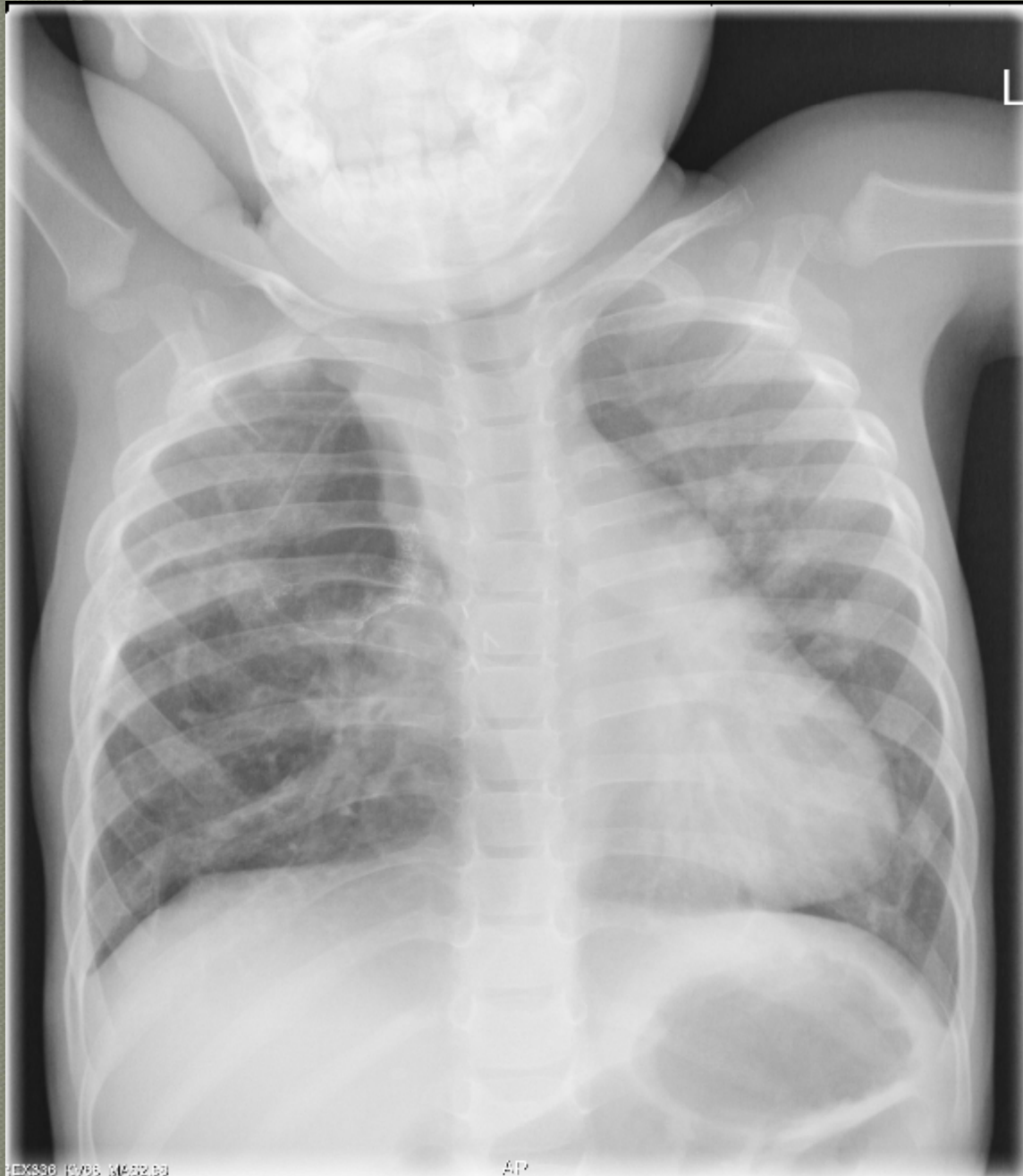


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Marconi Medical Systems, NM Division IRIX
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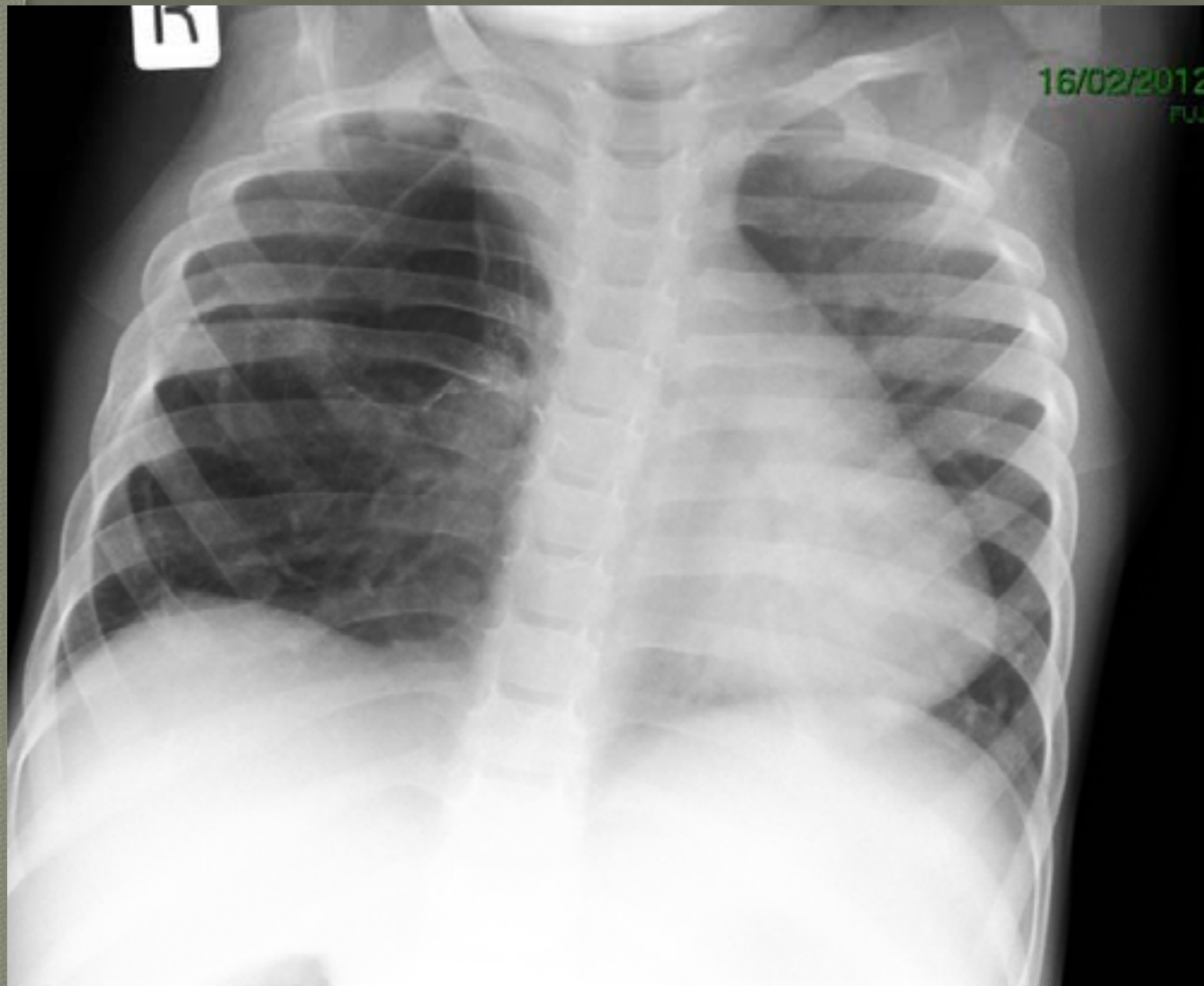




Left lung remained small
– hypoplastic!

Hypoxia and pulmonary
hypertension developed

required continuous
oxygen and viagra
till age 3 years



Now age 3y

Gradual improvement

Viagra stopped

Oxygen sat normal
on room air

Still FTT

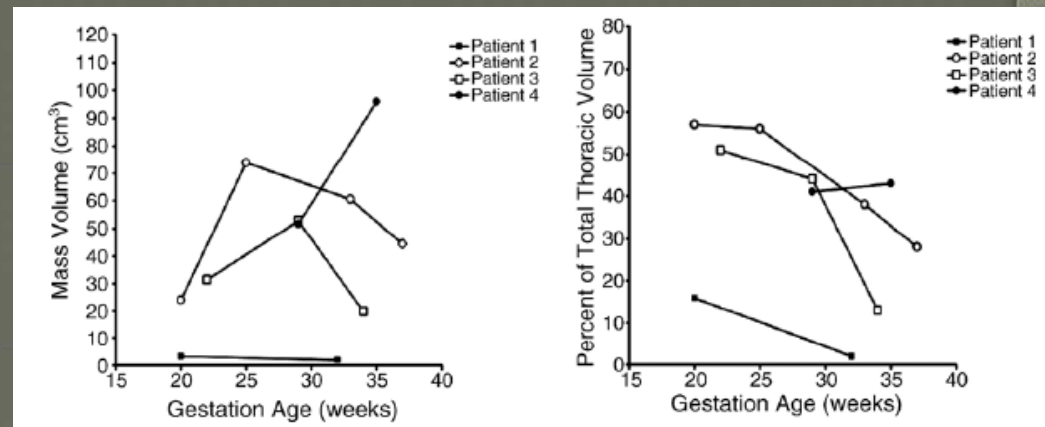
Large CPAMS

- ◉ For most CPAM's excellent prognosis
- ◉ Large CPAM: significant mass effect on adjacent thoracic structures
- ◉ Hydrops – high risk in utero death
- ◉ But: other outcome measures?
- ◉ prognosis?

Large fetal CCAMs: growth trends and patient survival

Kunisaki SM, J Pediatr Surgery 2007 (Harvard)

- 5y , 59 chest lesions
- 12 large CCAM (>3cm, shift of mediastinum/ diaphragm)
 - 6 hydrops/ascites
- US
- Serial MRI volumetry
- Peak at 25w
- Postnatal:
 - 5 – emergency:
 - ECMO 1,
 - cyst aspiration 1,
 - lung resection – 5



3 deaths (all severe hydrops <30wks)

But what about lung hypoplasia?

LHR = lung to head circumference ratio

- By fetal Ultrasound
- The traditional prognostic indicator for fetuses with CDH (O/E <25% = poor prognosis).
- More accurate earlier in gestation
- measurement of contralateral LHR:
- Right lung area measured by 2-D:
 - product of the 2 longest perpendicular linear measurements (with A-P axis parallel to sternovertebral body axis)
- Ratio of right lung area (mm²) to head circumference (mm)
- Disadvantages: measures only the contralateral lung. The ipsilateral lung may contribute 22% lung volume

Use of MRI in prenatal prognosis of the fetus with isolated left CDH.

Victoria T et al. Prenatal Diagnosis 2012 (CHOP)

- 85 cases.
- Prognosis correlated with:
- Lung-to head ratio (LHR) at diagnosis ($p=.009$)
- O/E LHR ($p=.01$)
- Total FLV ($p=.03$)
- O/E FLV ($p=.002$)

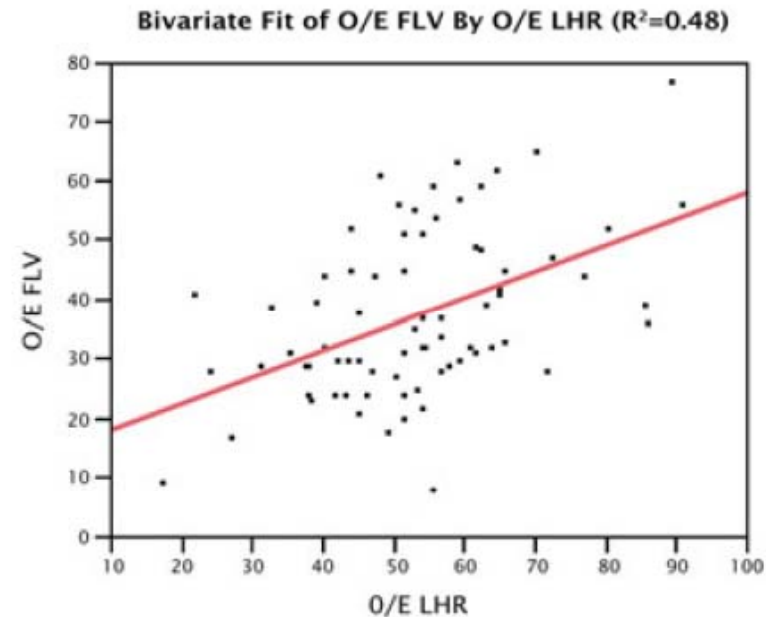


Figure 7 Scatter plot demonstrating that as O/E LHR increases, the ratio between MRI observed and expected lung volumes also rises. An O/E LHR value of 25 correlates with an MRI O/E FLV value of 25%. $LHR = 14.0755 + 44.317895 \times (O/E \text{ MRI total lung volume})$. $R^2 = 0.48$

Assessment of Lung Volume by 3-D sonography and MRI in fetuses with Congenital Diaphragmatic Hernia

Kehl S et al. J Ultrasound Med 2011

Pulmonary hypoplasia determines morbidity and mortality
Total fetal lung volume: predictive

Previous method: 2D ultrasound: lung to head ratio

81 pts, 18-39wks

3D sonographic volume of contralateral fetal lung

Rotational technique, at 6, 15 and 30 degrees

Ref. method: Transverse multiplanar T2-weighted MRI

Antepartum prediction of pulmonary hypoplasia: an elusive goal?

Harstad TW, Am J Perinatol. 1993

“....we were unable to predict accurately those fetuses with pulmonary hypoplasia. Moreover, MRI appears not to offer any advantages over sonographic evaluation of fetal pulmonary hypoplasia.”

Fetal lung volume: estimation at MR imaging-initial results. Rypens F. Radiology. 2001

- 336 fetuses, 21-38wks gestation.
- Normal FLV increases with GA as a power curve;
- spread of values increases with age.
- Normal FLV in all fetuses with CCAM; Lowest FLV with CDH
- potential for comparing FLV in fetuses at risk of lung hypoplasia with normative values

Fetal Body MRI-

Lisa Raviv-Zilka M.D

Diagnostic Imaging, Pediatric Imaging Unit
The Edmond and Lily Safra Children's Hospital
Sheba Medical Center

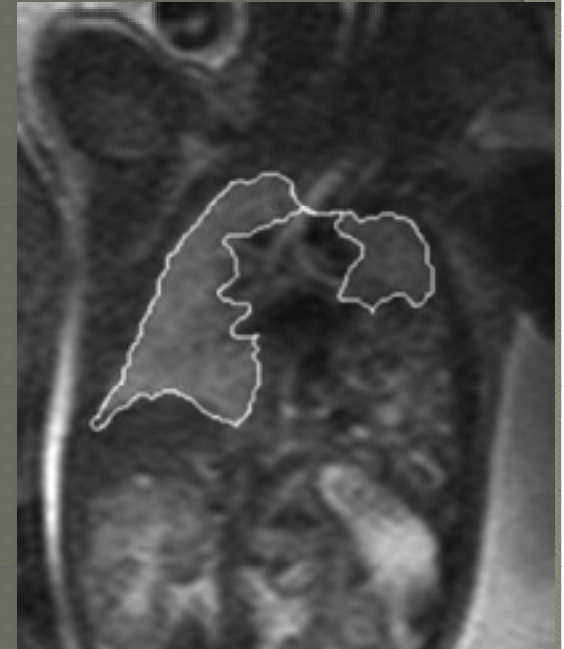
>200 fetal MRI's to date (June 2012)



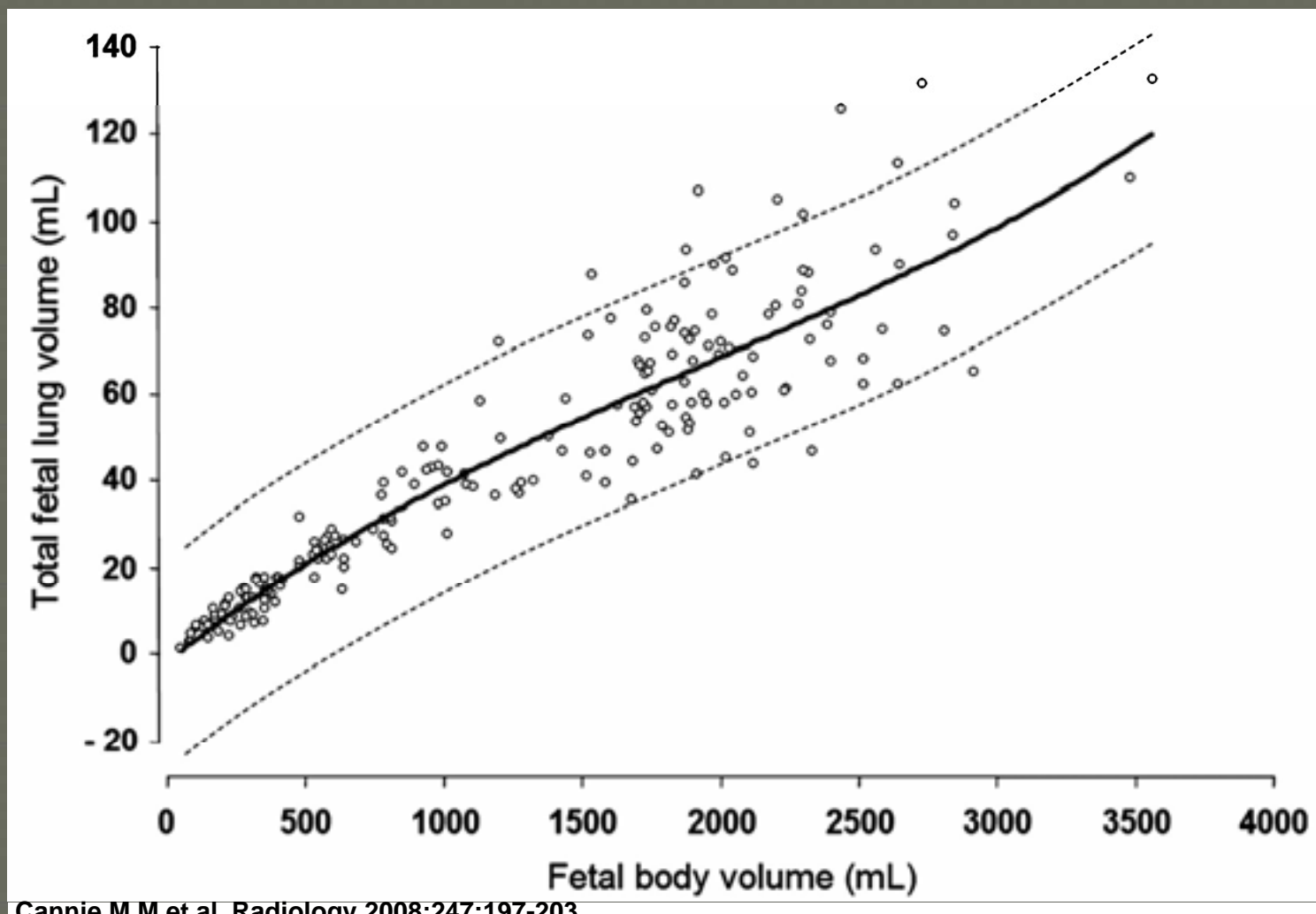
Fetal Body Volume at MR Imaging to Quantify Total Fetal Lung Volume: Normal Ranges

Cannine et al Radiology, 2008

- 200 fetuses, 16 - 40w
- MR protocol: T2. 38 adjacent sections
- free-form regions of interest.
- added and multiplied by the section thickness to determine the entire volume of the right and left lungs (allowing calculation of TFLV)



Plot of TFLV with FBV in fetuses with normal lungs shows strong correlation between these two parameters, with mean (solid line) and 95% confidence intervals for prediction (dashed lines).



Cannie M M et al. Radiology 2008;247:197-203

Radiology

MR Relative Fetal Lung Volume (FLV) in Congenital Diaphragmatic Hernia (CDH): Survival and Need for ECMO

Busing KA, Radiology 2008 Jul;248(1):240-6 (Heidelberg)

Aim:

FLV at MRI - absolute and as % predicted lung vol.

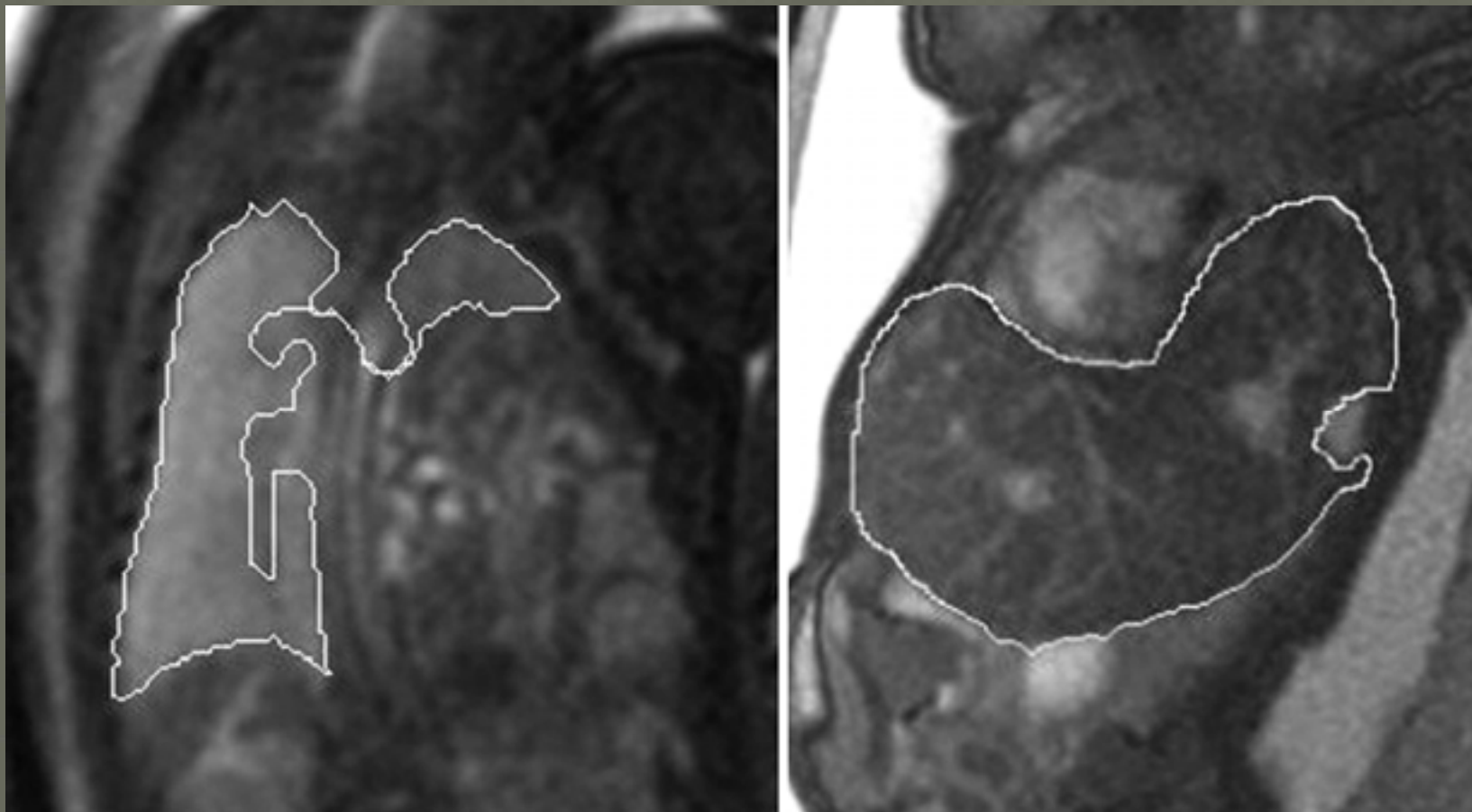
to predict neonatal survival and need for ECMO

68 fetuses with CDH (23-29wks) – using 7 diff. formulas, AUC

RESULTS:

- **FLV was sig. different in neonates who survived and neonates who did not survive** ($p = .001 - < .001$) with each method (AUC, 0.800-0.900).
- **FLV differences for predicting need for ECMO were smaller but still significant** ($p = .05 - < .009$)

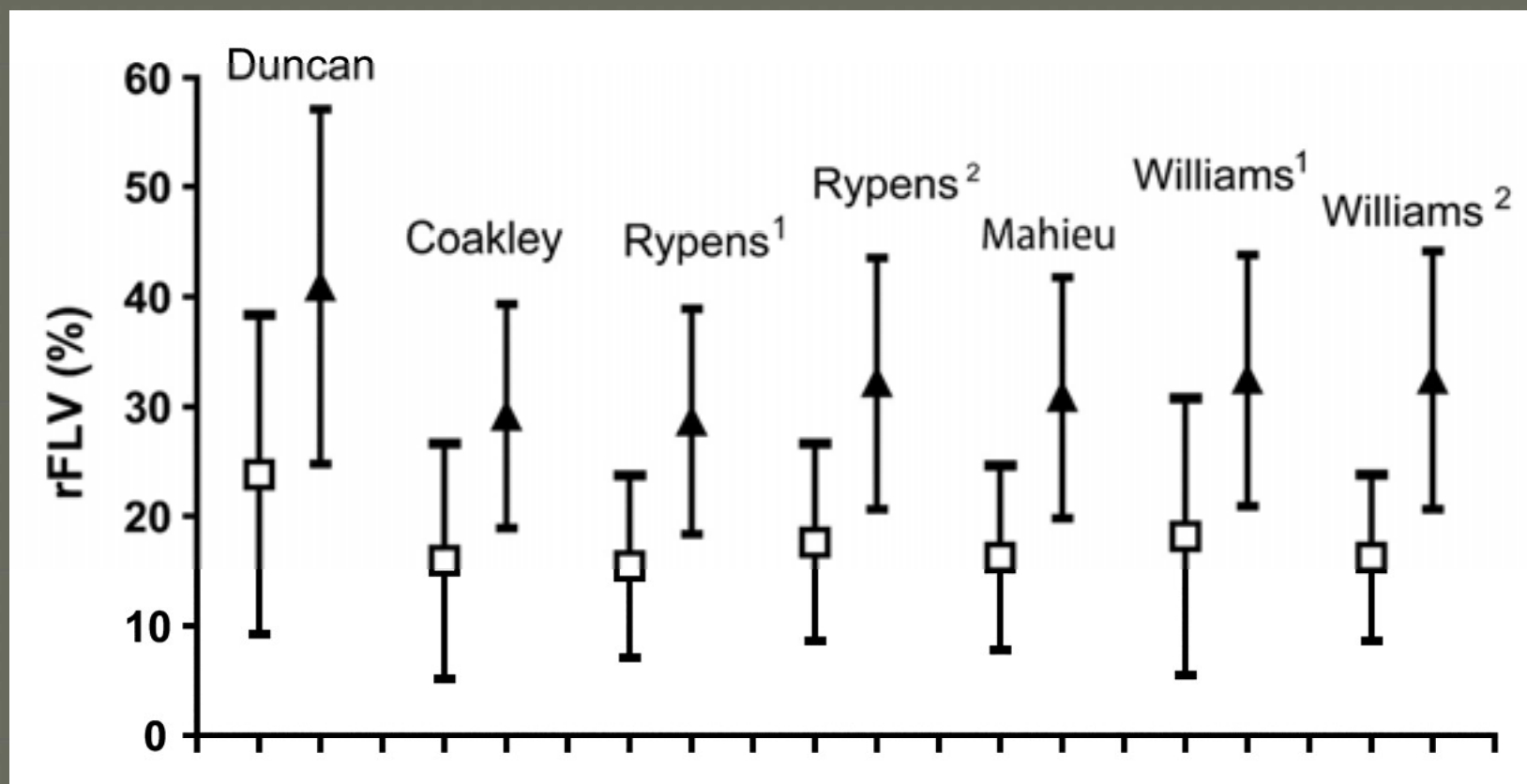
Coronal true fast imaging with steady-state precession MR images (3.6/1.5; flip angle, 59°) of fetal lung (left) and liver (right) at 30 weeks gestation.



Büsing K A et al. Radiology 2008;248:240-246

Radiology

Graph shows relative FLVs in fetuses with CDH assessed with different formulas.



Büsing K A et al. Radiology 2008;248:240-246

Radiology

Fetal Lung-to-Liver Signal Intensity Ratio (LLSIR) at MR Imaging: **Development of a Normal Scale and Possible Role in Predicting Pulmonary Hypoplasia in Utero**

Brewerton LJ et al, Radiology, 2005

Lung
Liver

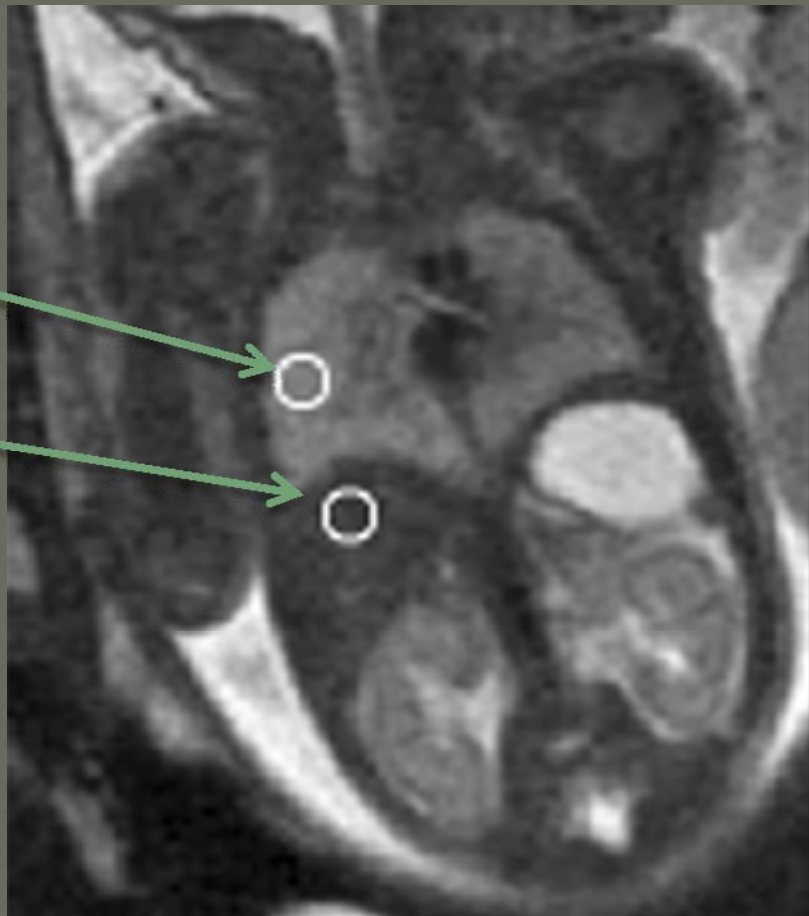
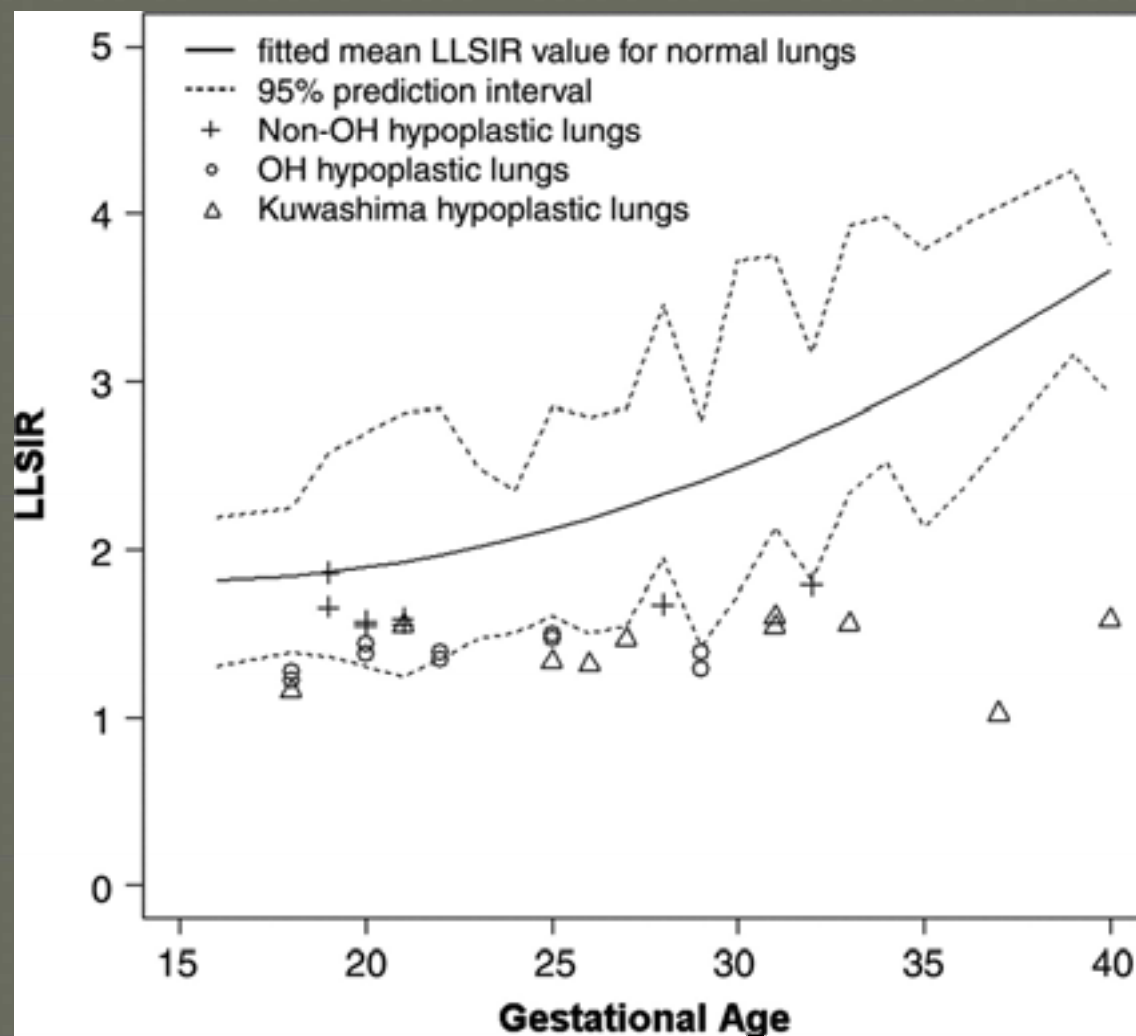


Figure 1. Coronal half-Fourier single-shot turbo spin-echo MR image (1100/68, 4-mm section thickness, 30 × 30-cm field of view, 218 × 256 matrix) of fetus at 28 weeks gestation shows regions of interest in lung (upper region of interest) and liver (lower region of interest) from which LLSIR was calculated.

Figure 3. Graph shows LLSIR for hypoplastic lungs compared with normal LLSIR range. ○ = hypoplastic lungs associated with oligohydramnios. + = hypoplastic lungs not associated with oligohydramnios.



Brewerton L J et al. Radiology 2005;235:1005-1010

Radiology

Effect of antenatal corticosteroid treatment on the fetal lung: a magnetic resonance imaging study

M. SCHMID*, G. KASPRIAN†, L. KUESSEL*, A. MESSERSCHMIDT‡, P. C. BRUGGER§
and D. PRAYER†

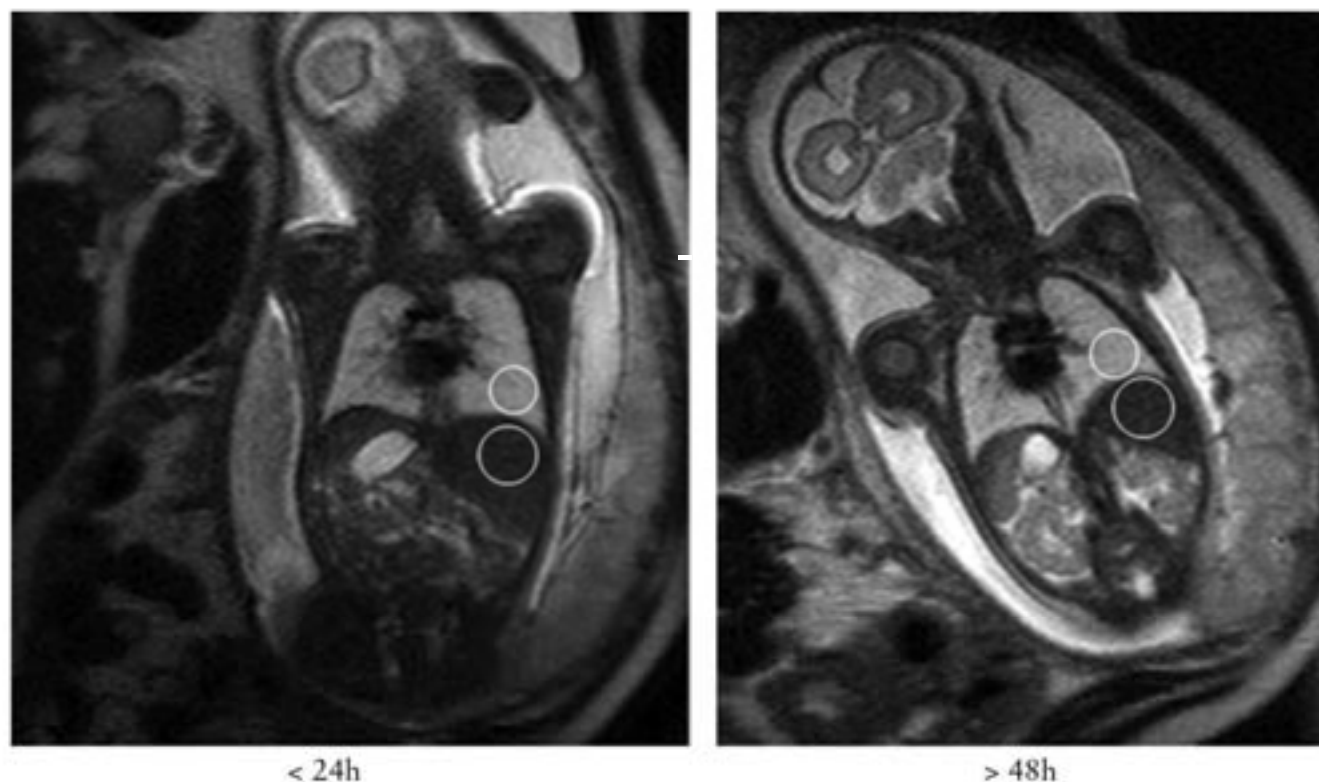


Figure 1 T2-weighted magnetic resonance images of a fetus at 27 gestational weeks. Time point '< 24 h' was within 24 h after the first dose of betamethasone. Time point '> 48 h' was after the second dose of betamethasone, more than 48 h after the first dose. For the fetus shown, the time spans for '< 24 h' and '> 48 h' were 2 h 38 min and 77 h 23 min, respectively. The regions of interest for signal intensity measurement in the lung and the liver are depicted (white circles).

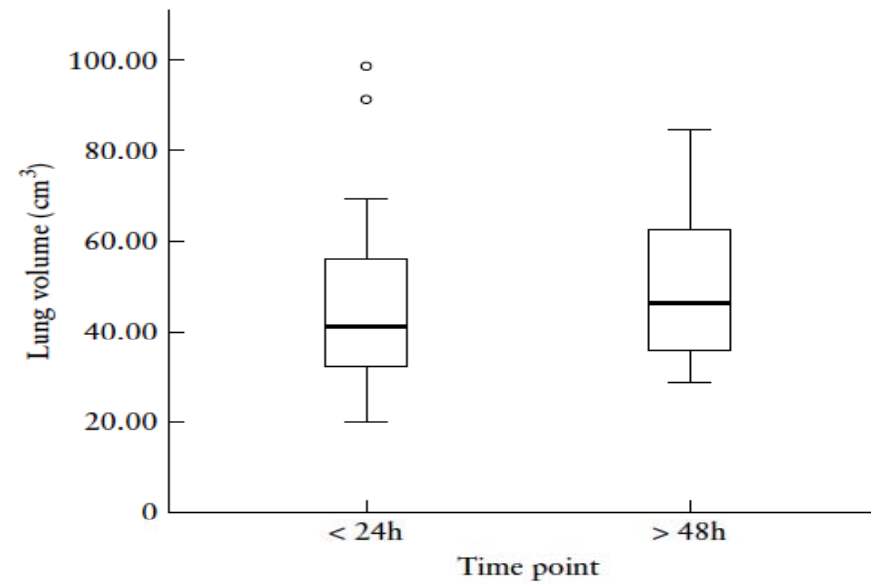


Figure 3 Box plot of fetal lung volumes at the two evaluated time points. Median, interquartile range, range and outliers are shown.

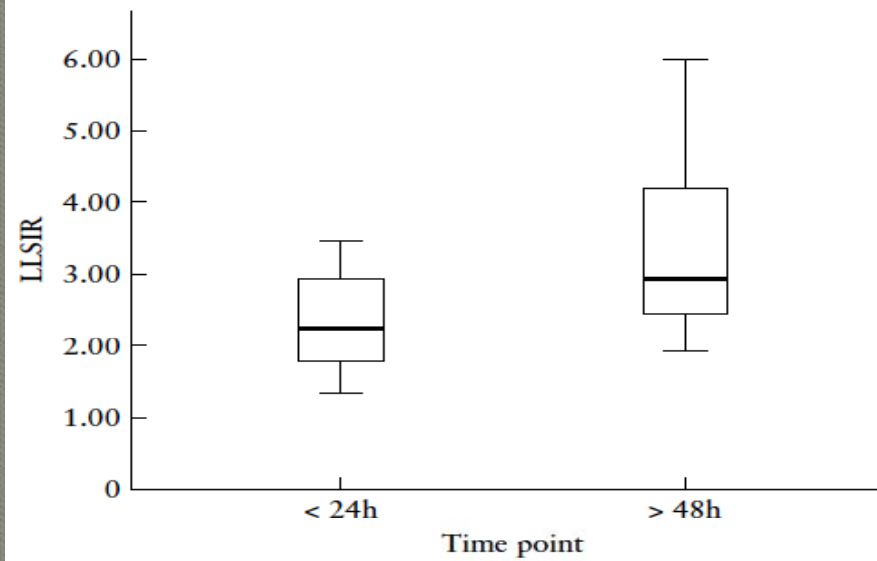


Figure 4 Box plot of fetal lung and liver signal-intensity ratios (LLSIR) at the two evaluated time points. Median, interquartile range and range are shown.

With thanks to:

Prof. Israel Meizner and all the team at the Ultrasound Clinic, Helen Schneider Women's Hospital, Rabin Med Cr

Dr. Lisa Raviv-Zilka, Pediatric Radiology department, Sheba Med Cr, Tel Hashomer

Dr. Liat Ben Sira, Pediatric Radiology department, Sourasky Med Cr, Tel Aviv

Pulmonologists, Schneider Children's – prenatal consults regularly