CONGENITAL DIAPHRAGMATIC HERNIA

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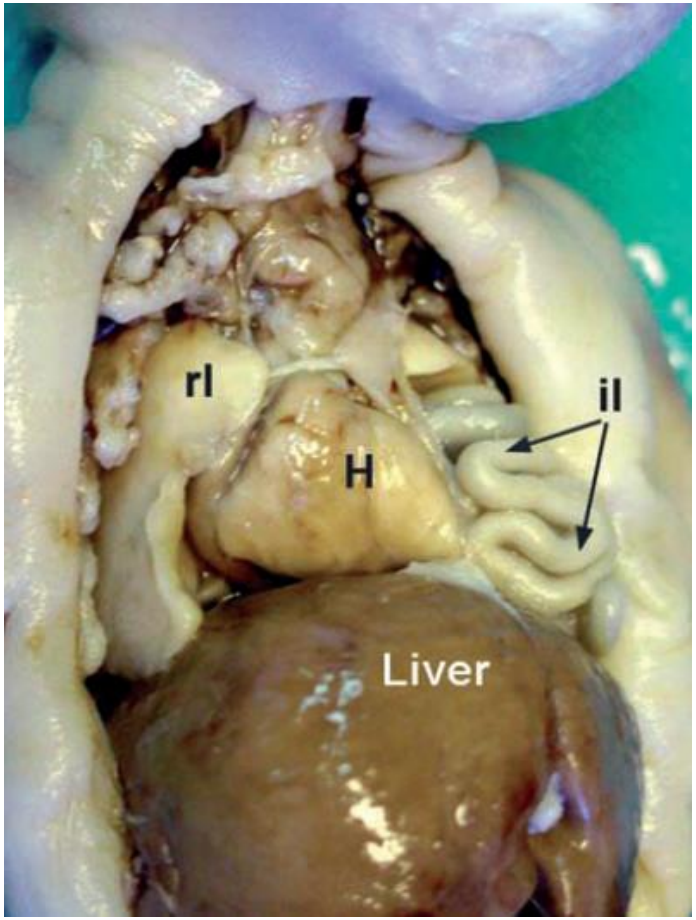
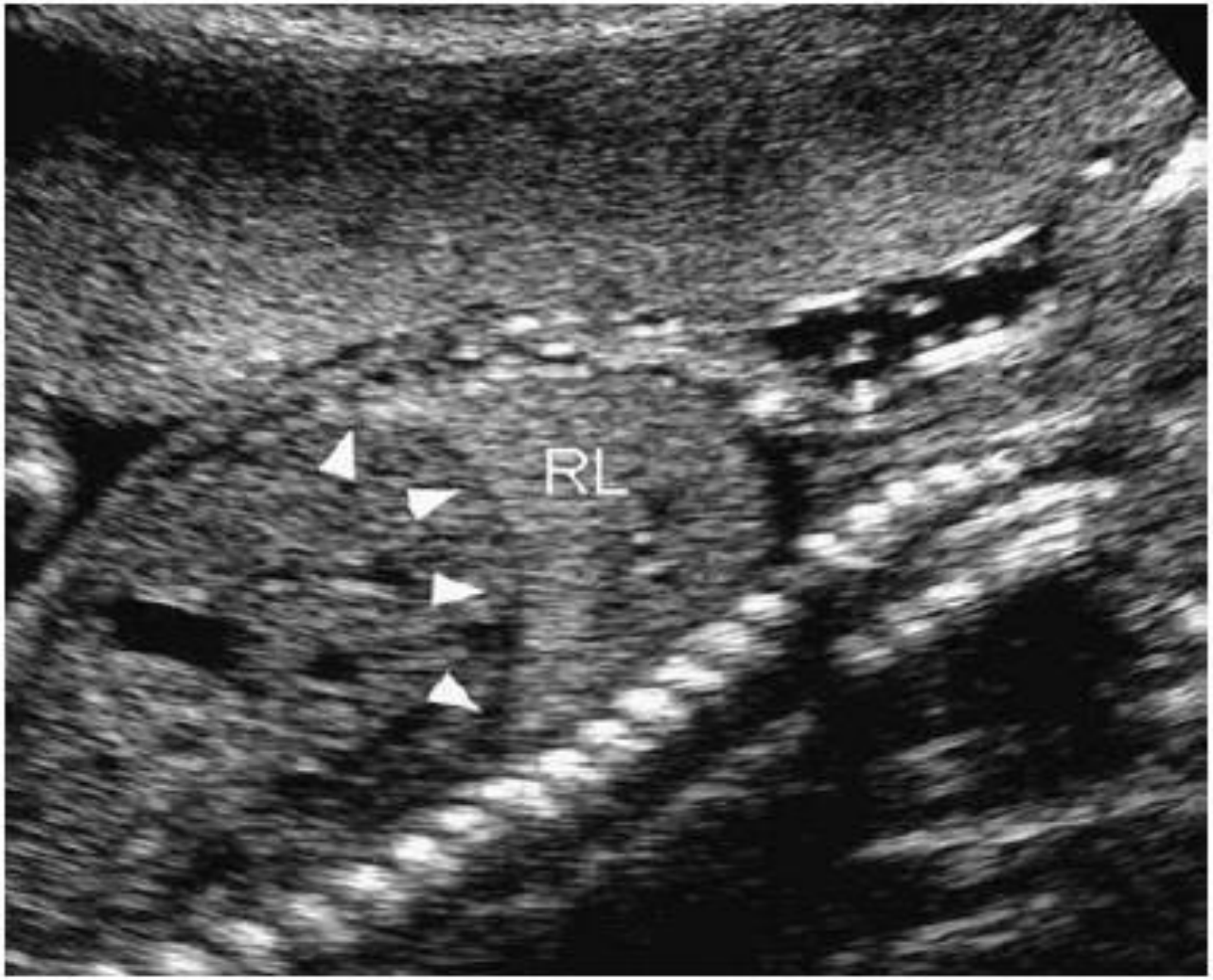


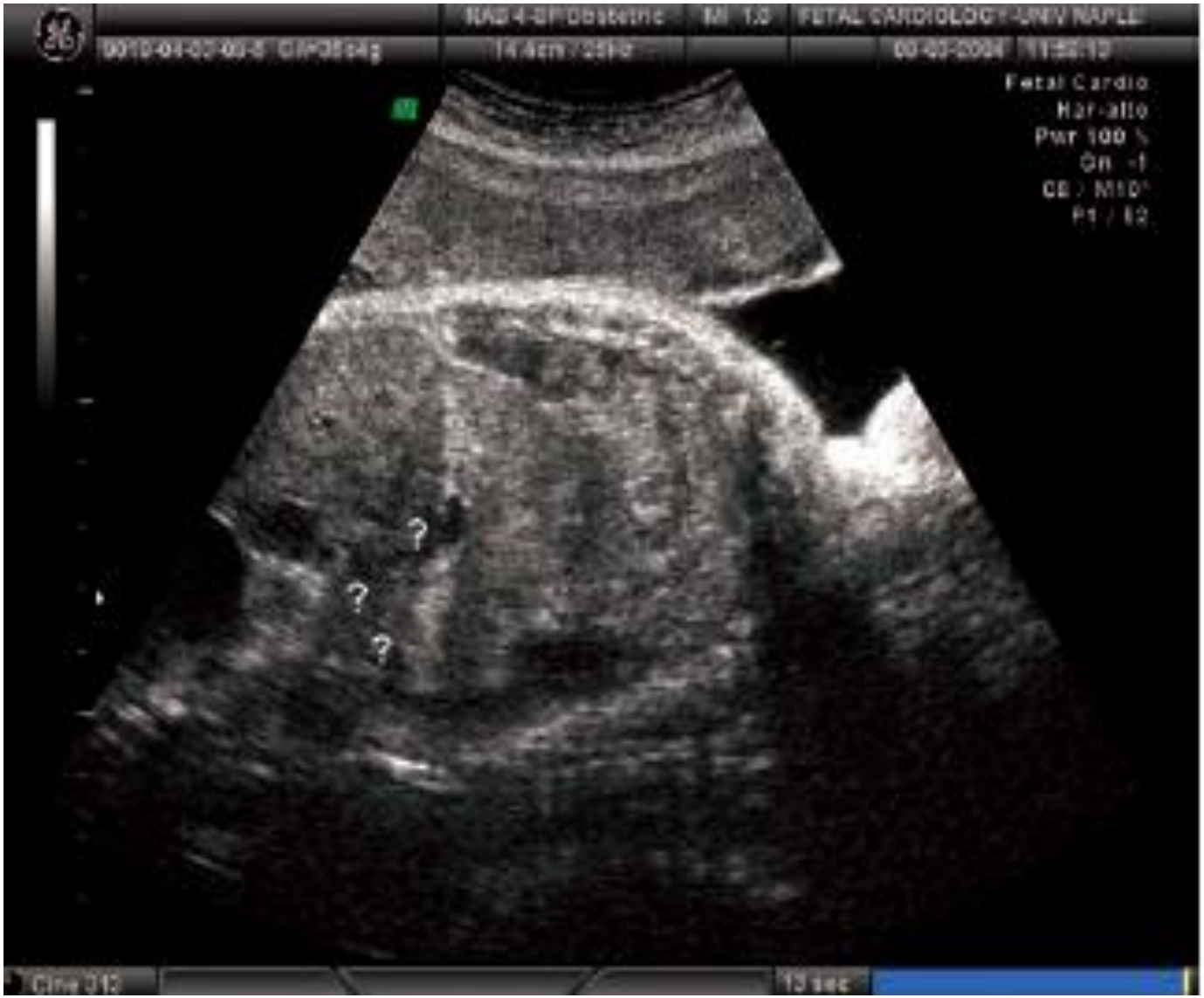
- *2.4-4.9 in 10000*
- *Most Sporadic*
- *Male=female*
- *Left>>>>Right> Bilateral*
- *Associated with smoking, alcohol, vitamin A deficiency, thalidomide, anticonvulsants*
- *Survival for isolated CDH :60%*

- *Results from abnormal diaphragm development at 6–10 weeks with incomplete closure of the pleuroperitoneal folds*
- *Herniated viscera cause decreased bronchial branching, alveolar number, and pulmonary vascularization, and over muscularization of pulmonary arterial tree, leading to:*
- *Pulmonary Hypoplasia*
- *Pulmonary Hypertension*

Diagnosis of CDH

- *Displaced heart (to the right)*
- *Ectopic stomach in the chest*
- *Scaphoid abdomen*
- *Very small ipsilateral fetal lung*
- *Liver in chest (poorer prognosis)*
- *Longitudinal views: a tortuous aspect of the inferior vena cava and the absence of the hypoechoic contour of the diaphragm*
- *Polyhydramnios*
- ****Defect in 1st trimester but herniation in different time (50-60% prenatal diagnosis)*


RL


Fetal Cardio
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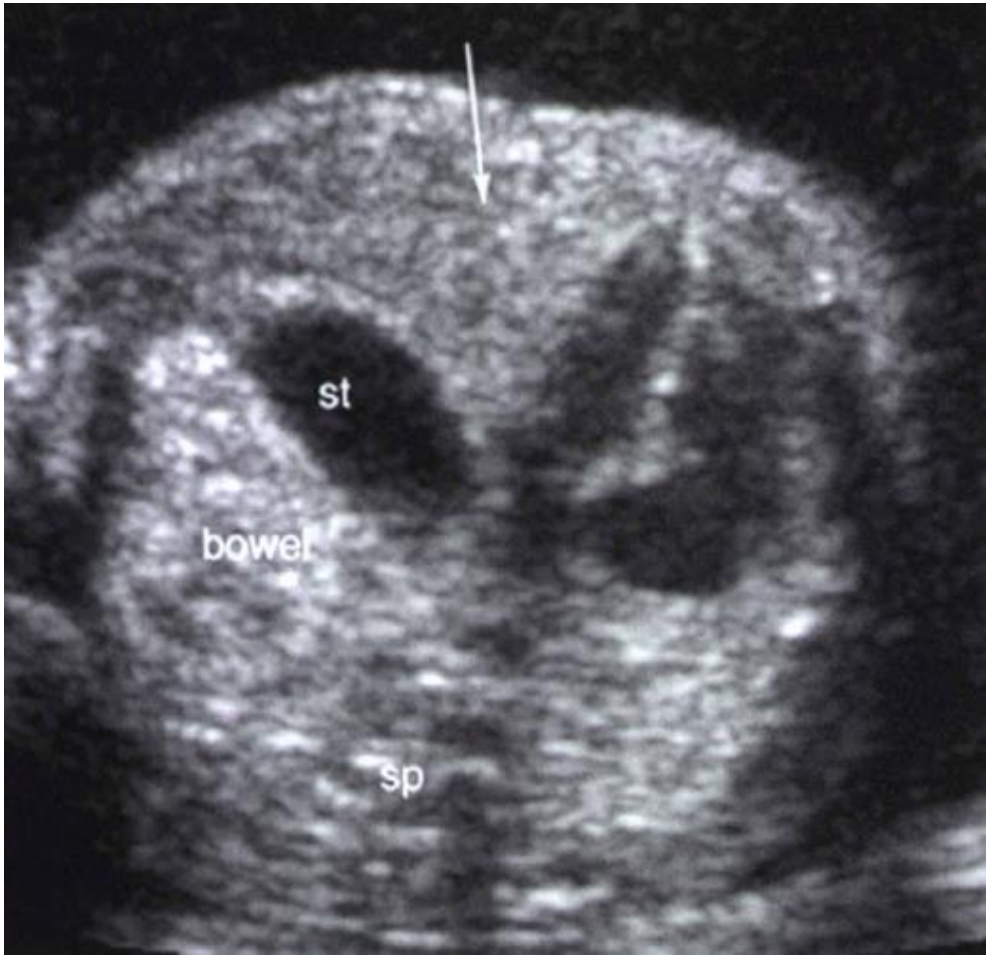
?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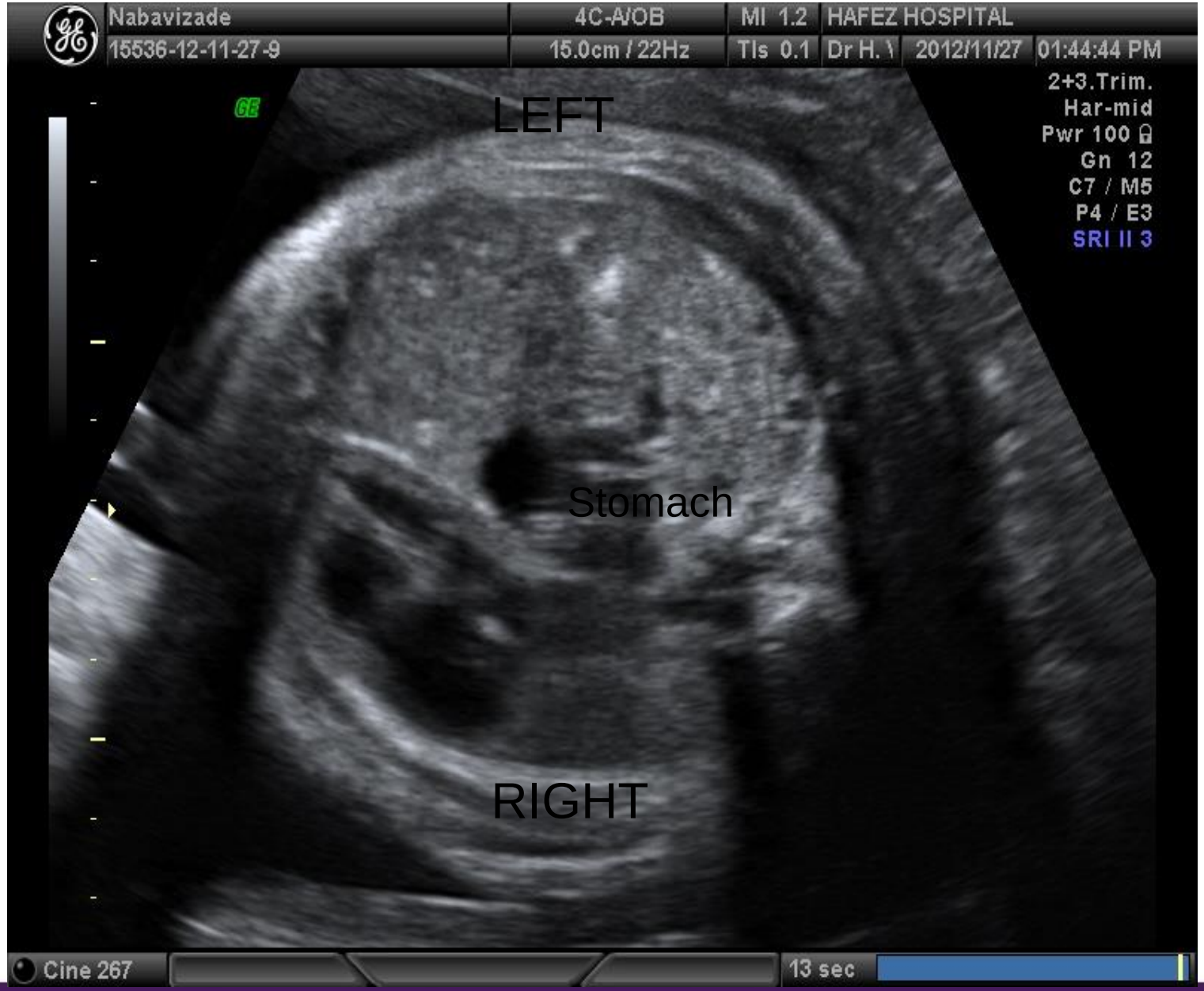
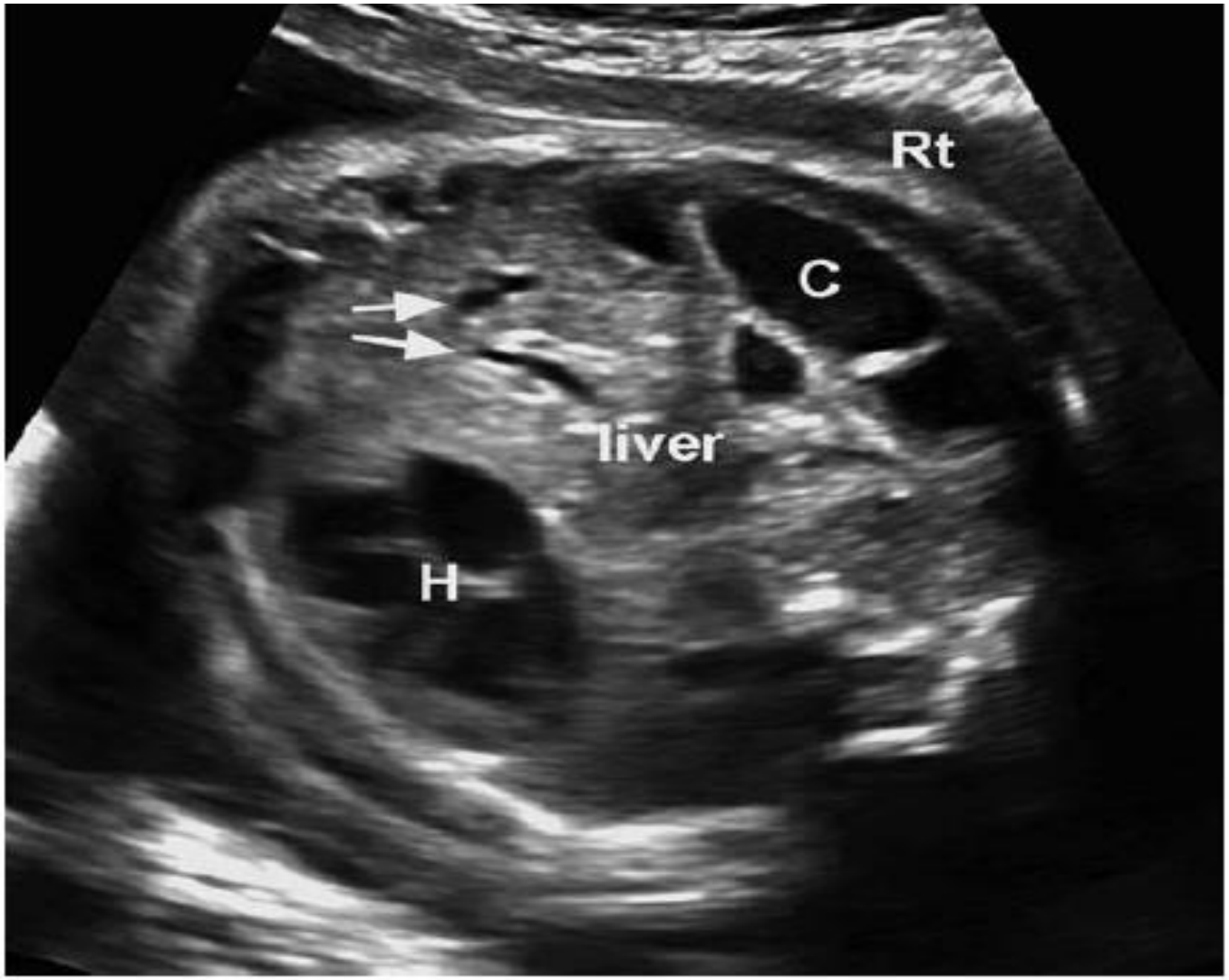
bowel
sp


Nabavizade
4C-A/OB
MI 1.2
HAFEZ HOSPITAL
15536-12-11-27-9
15.0cm / 22Hz
TIs 0.1
Dr H. \
2012/11/27
01:44:44 PM
2+3.Trim.
Har-mid
Pwr 100
Gn 12
C7 / M5
P4 / E3
SRI II 3
LEFT
Stomach
RIGHT
Cine 267
13 sec


Rt
C
liver
H

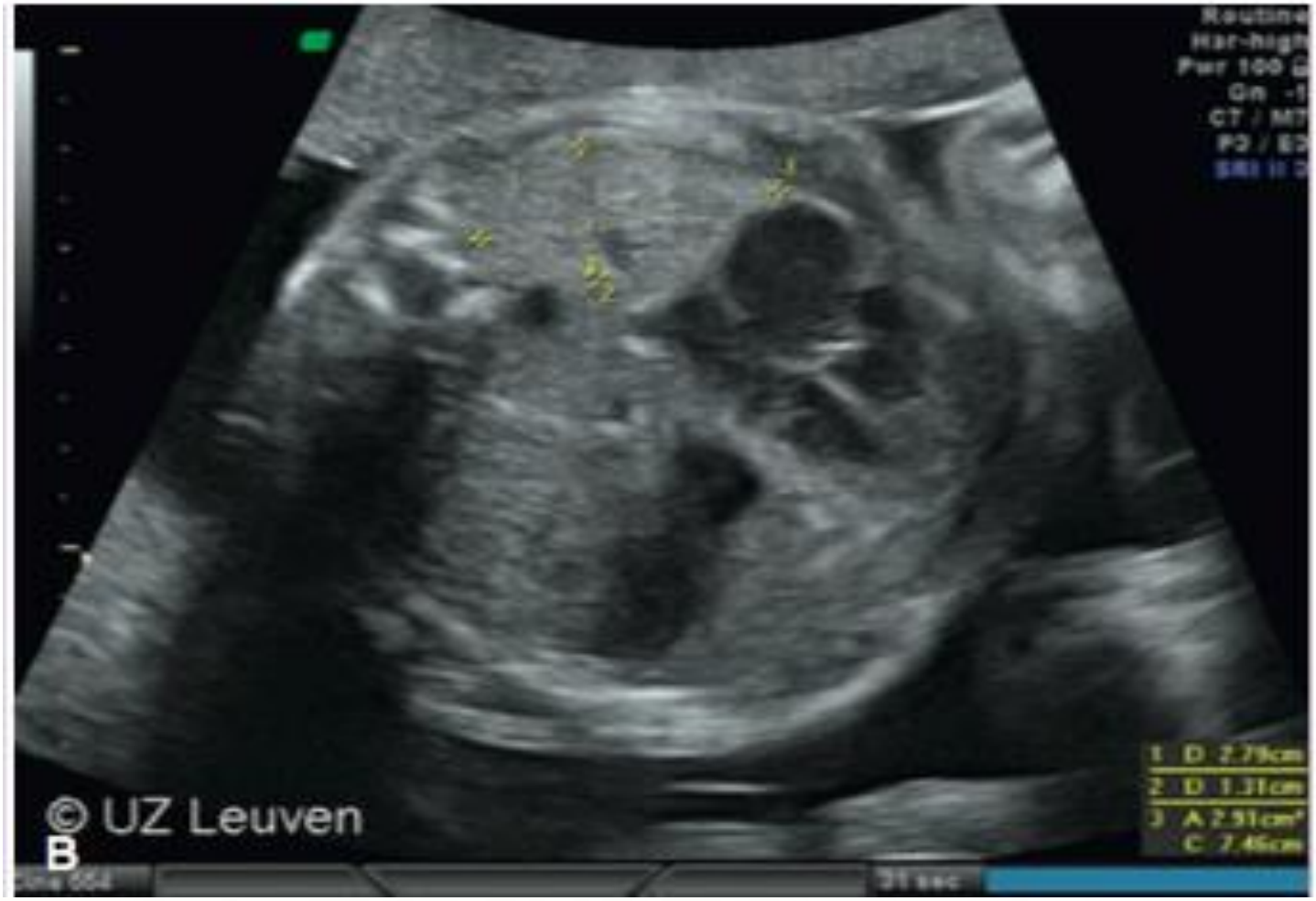
D.DX of CDH

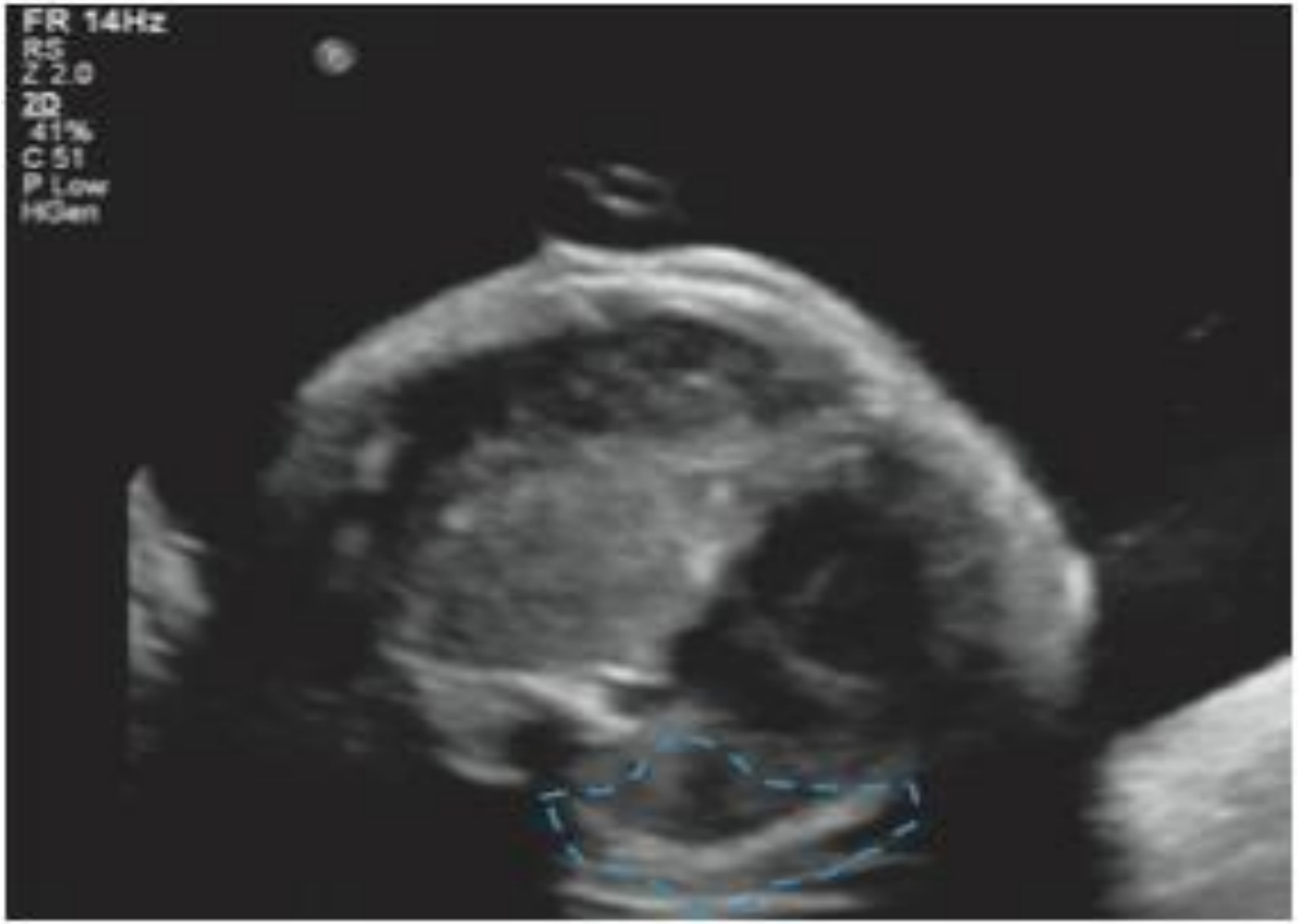
- Congenital pulmonary airway malformation
- Congenital diaphragmatic hernia
- Tracheoesophageal fistula
- Pulmonary sequestration
- Cysts
  - Bronchogenic
  - Foregut
- Tumors
  - Neuroblastoma
  - Mediastinal teratoma
  - Rhabdomyoma
- Atresia
  - Bronchial with distal degeneration
- Congenital lobar emphysema
- Congenitally small lungs
- Lung agenesis
- Vascular abnormalities
  - Vascular rings
  - Pulmonary artery slings

PROGNOSTIC FEATURES OF LEFT CDH

- *Chromosomal abnormality or Other anomalies*
- *Early diagnosis (less than 24 wk gestation) and early delivery*
- *Lung – Head Ratio*
- *Intrathoracic stomach*
- *Intrathoracic liver*
- *Pul HTN*
- *Bilateral*

Lung to Head Ratio




FR 14Hz
RS
Z 2.0
ZD
41%
C 51
P Low
HGen

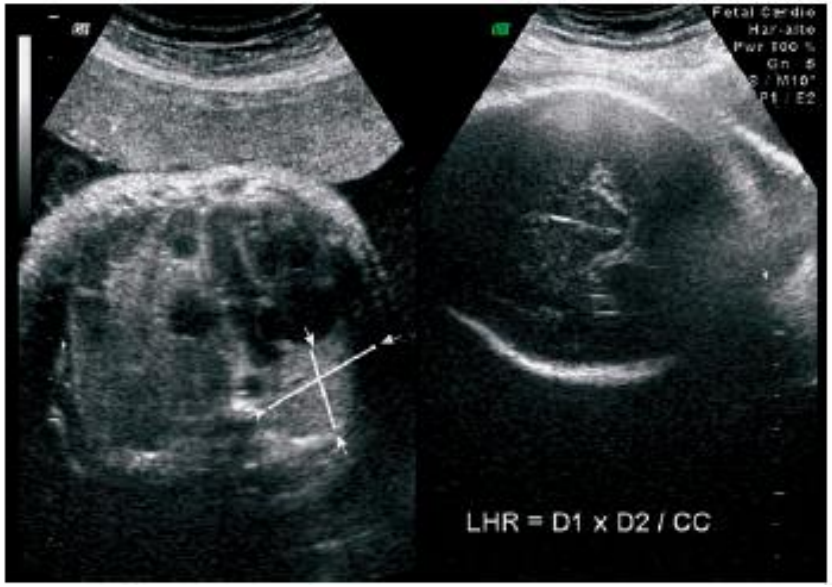


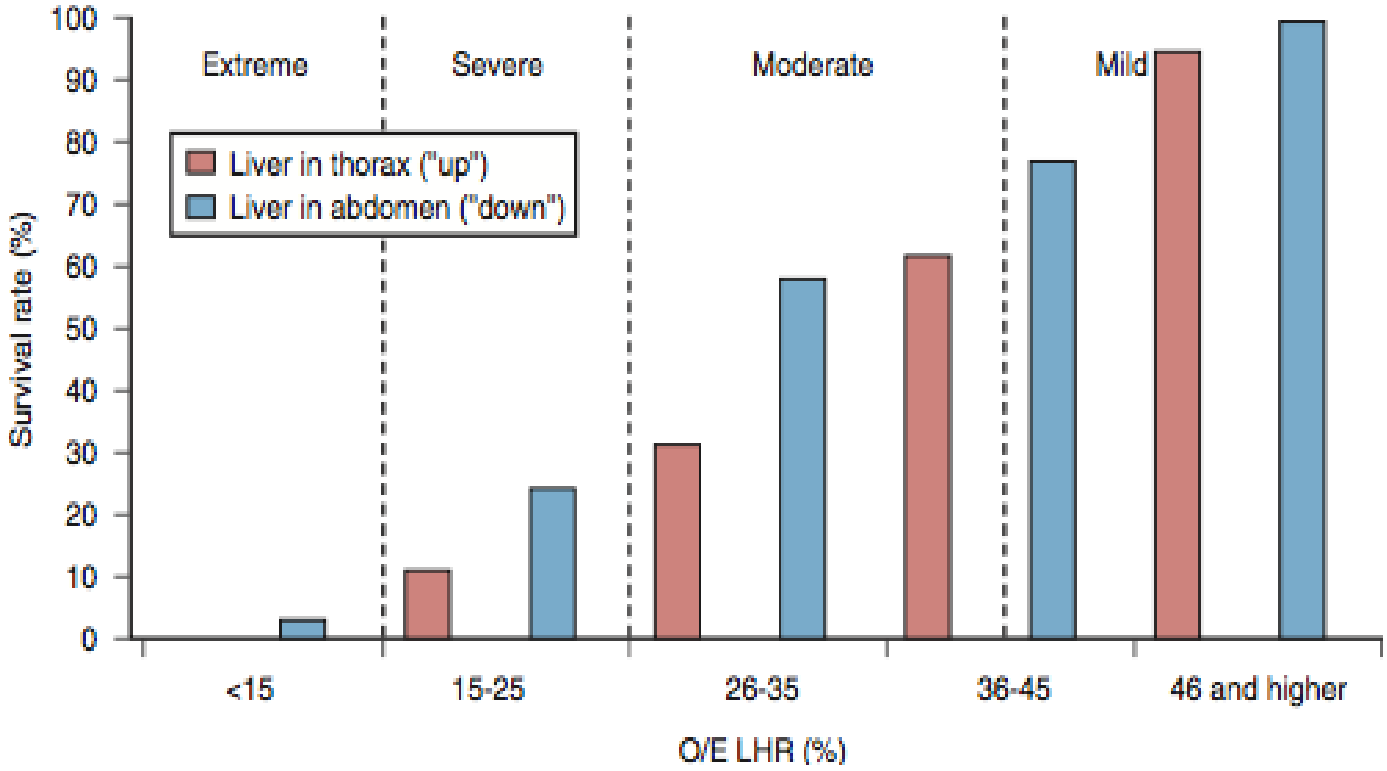
TABLE 37.9 Neonatal Outcome as a Function of Lung-to-Head Ratio in Fetuses With Left-Sided Isolated Congenital Diaphragmatic Hernia and Liver Herniation

Degree of Pulmonary Hypoplasia	LHR	Expectant Management[a] N	Expectant Management[a] Survival	FETO[b] N	FETO[b] Survival
Extreme	0.4–0.5	2	0 (0%)	6	1 (16.7%)
Severe	0.6-0.7	6	0 (0%)	13	8 (61.5%)
	0.8–0.9	19	3 (15.8%)	9	7 (77.8%)
	Total LHR <1.0	27	3 (11.1%)	28	16 (57.1%)
Moderate	1.0–1.1	23	14 (60.9%)		NA
	1.2–1.3	19	13 (68.4%)		NA
Mild	1.4–1.5	11	8 (72.7%)		NA
	≥1.6	6	5 (83.3%)		NA
Total		86	43 (50%)		

[a]Jani J, Keller RL, Benachi A, et al; Antenatal-CDH-Registry Group. Prenatal prediction of survival in isolated left-sided diaphragmatic hernia. *Ultrasound Obstet Gynecol.* 2006;27:18–22.

[b]Jani JC, Nicolaides KH, Gratacos E, et al. Fetal lung-to-head ratio in the prediction of survival in severe left-sided diaphragmatic hernia treated by fetal endoscopic tracheal occlusion (FETO). *Am J Obstet Gynecol.* 2006;195:1646–1650.

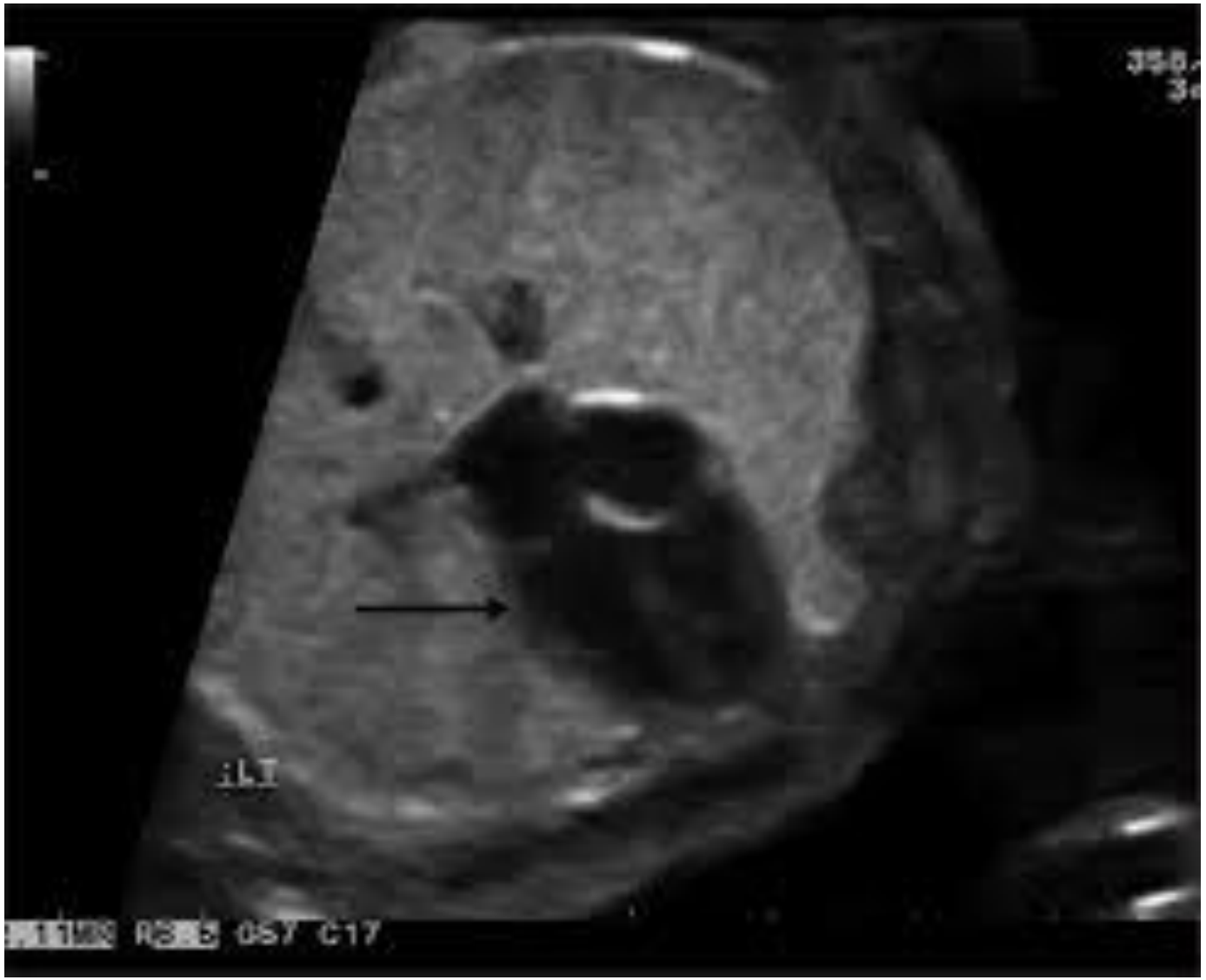
FETO, Fetoscopic endoluminal tracheal occlusion; *LHR*, lung-to-head ratio; *NA*, not applicable (fetuses were not eligible for FETO).


Extreme
Severe
Moderate
Mild
Liver in thorax ("up")
Liver in abdomen ("down")
Survival rate (%)
100
90
80
70
60
50
40
30
20
10
0
<15
15-25
26-35
36-45
46 and higher
O/E LHR (%)

Management of CDH

- ***Diagnosis of CDH should prompt referral to a tertiary center
- Detailed morphologic survey, 3D US, MRI
- Fetal echocardiography
- Genetic testing
- parental counseling:
- Termination of the pregnancy
- Repair of the defect after birth
- Fetal surgery
- *** Repair after birth does not reverse the pulmonary hypoplasia and pulmonary hypertension caused by the CDH***

*** Prenatal intervention for CDH will be justified if improvement in survival or morbidity can be demonstrated when compared to planned delivery and postnatal management with gentle ventilation strategy*****

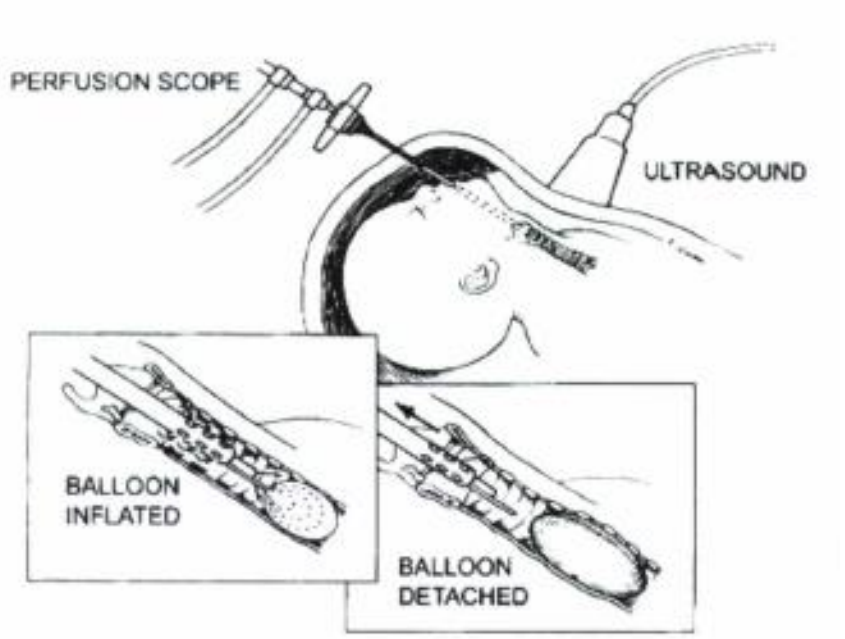


Prenatal surgical intervention

- ***Open fetal surgical repair***
- ***Open surgical tracheal occlusion***
- ***Endoscopic external tracheal occlusion***
- ***Endoscopic endoluminal tracheal occlusion***

Fetoscopic Endoluminal Tracheal Occlusion (FETO)

- Prevents egress of lung fluid
- Increasing airway pressure,
- Promotes pulmonary tissue proliferation
- Increases alveolar airspace
- Encourages maturation of pulmonary vasculature

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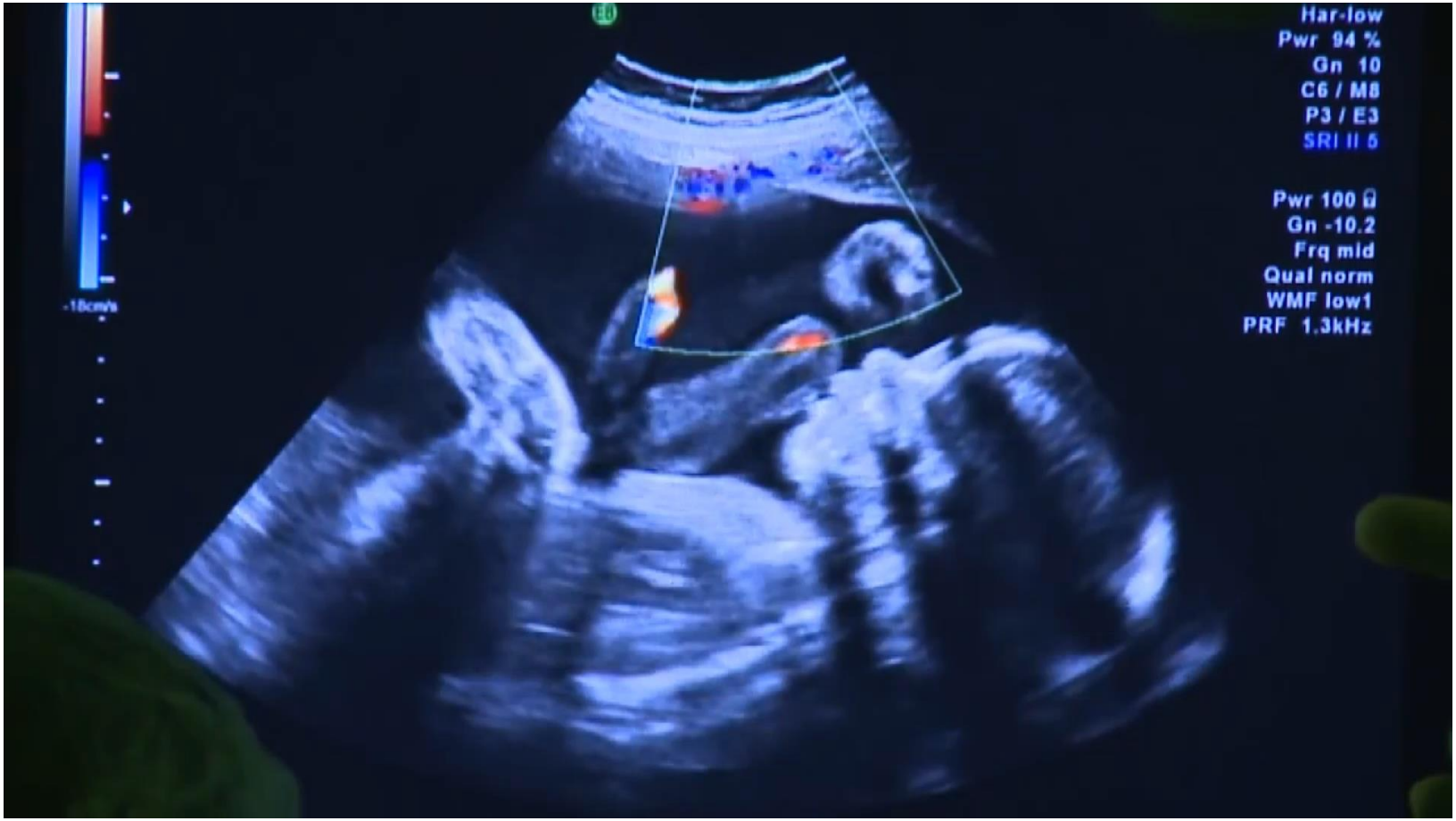
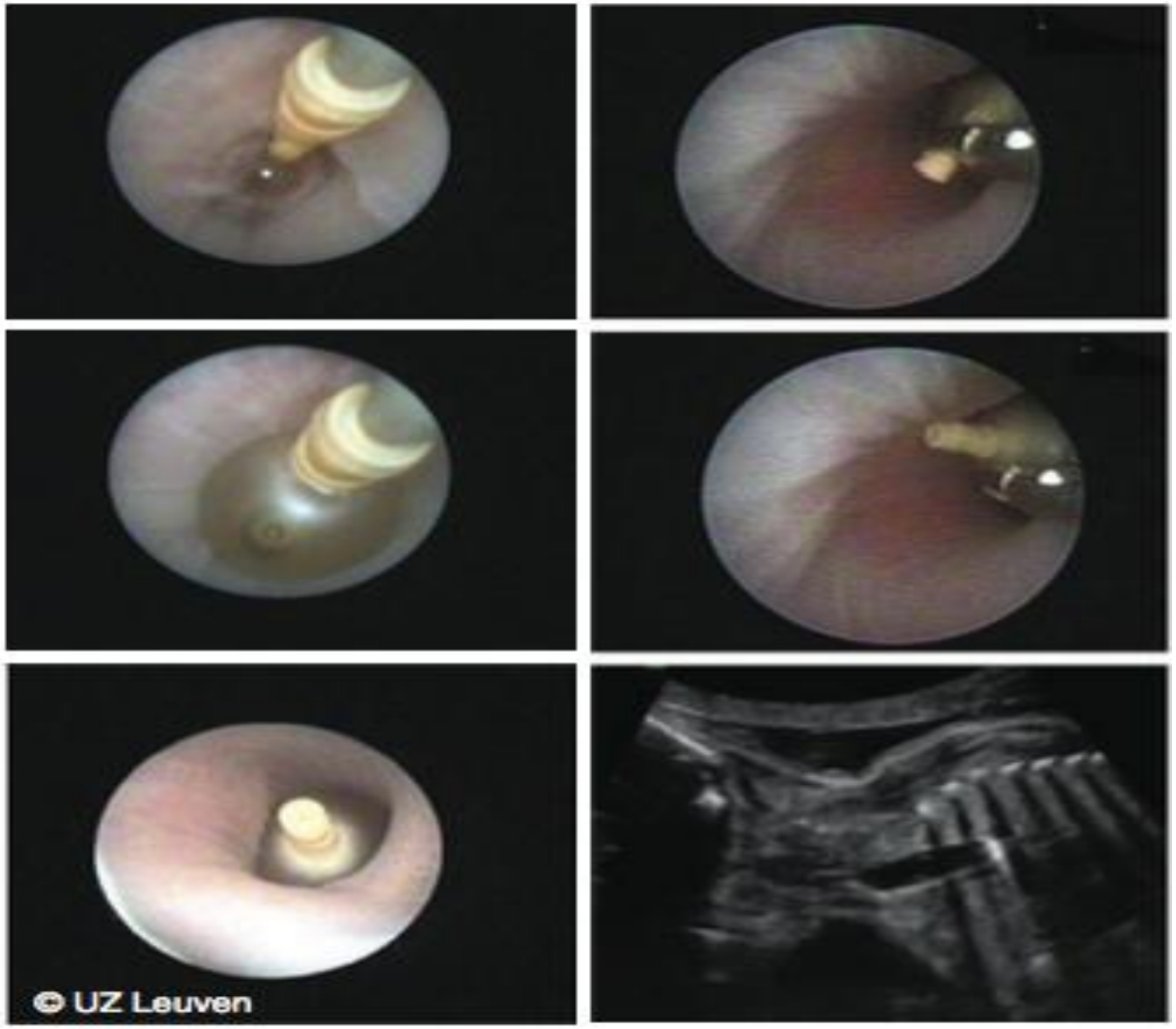

Har-low
Pwr 94 %
Gn 10
C6 / M8
P3 / E3
SRI II 6
Pwr 100
Gn -10.2
Frq mid
Qual norm
WMF low1
PRF 1.3kHz
-18cm/s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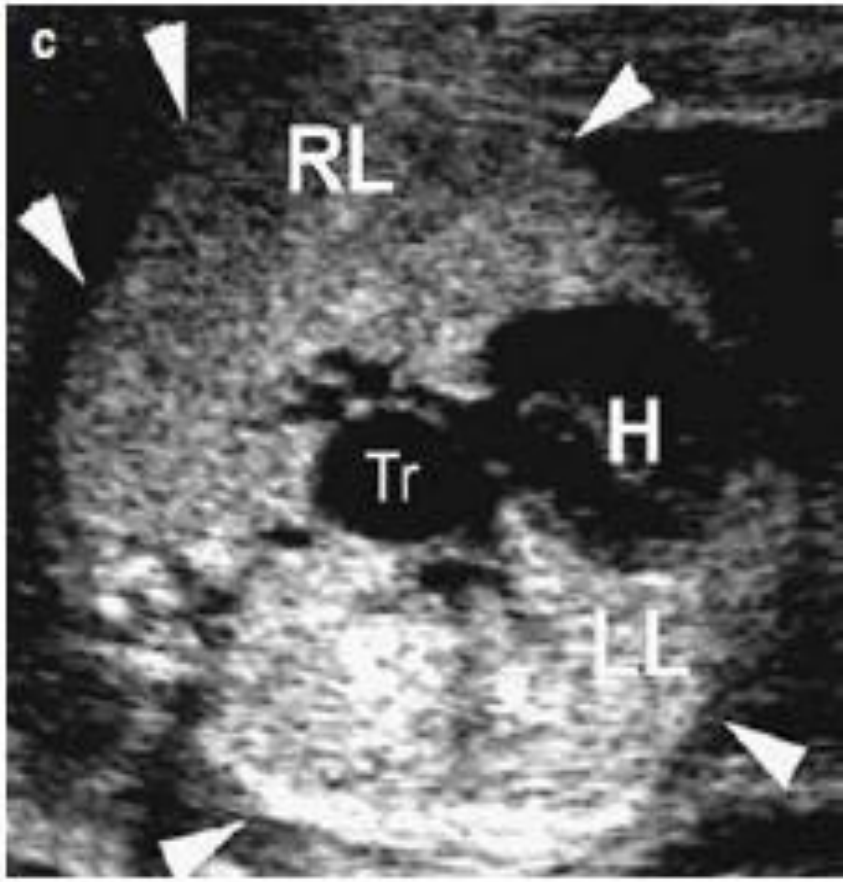
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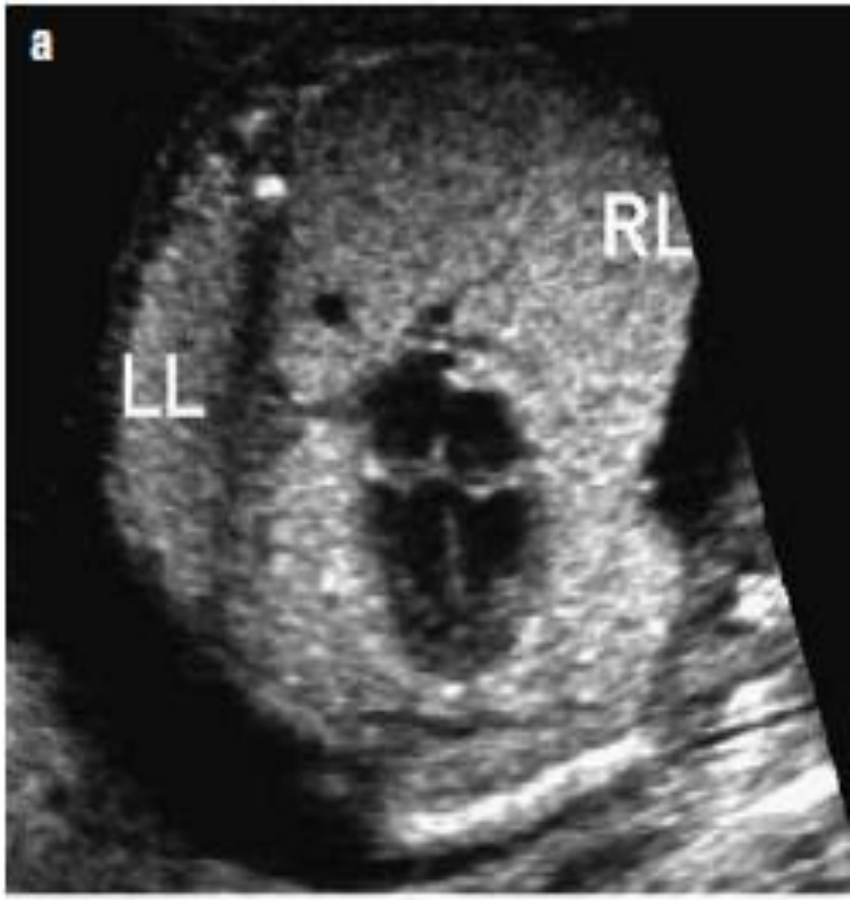
a
RL
LL

Many Thanks for your attention

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