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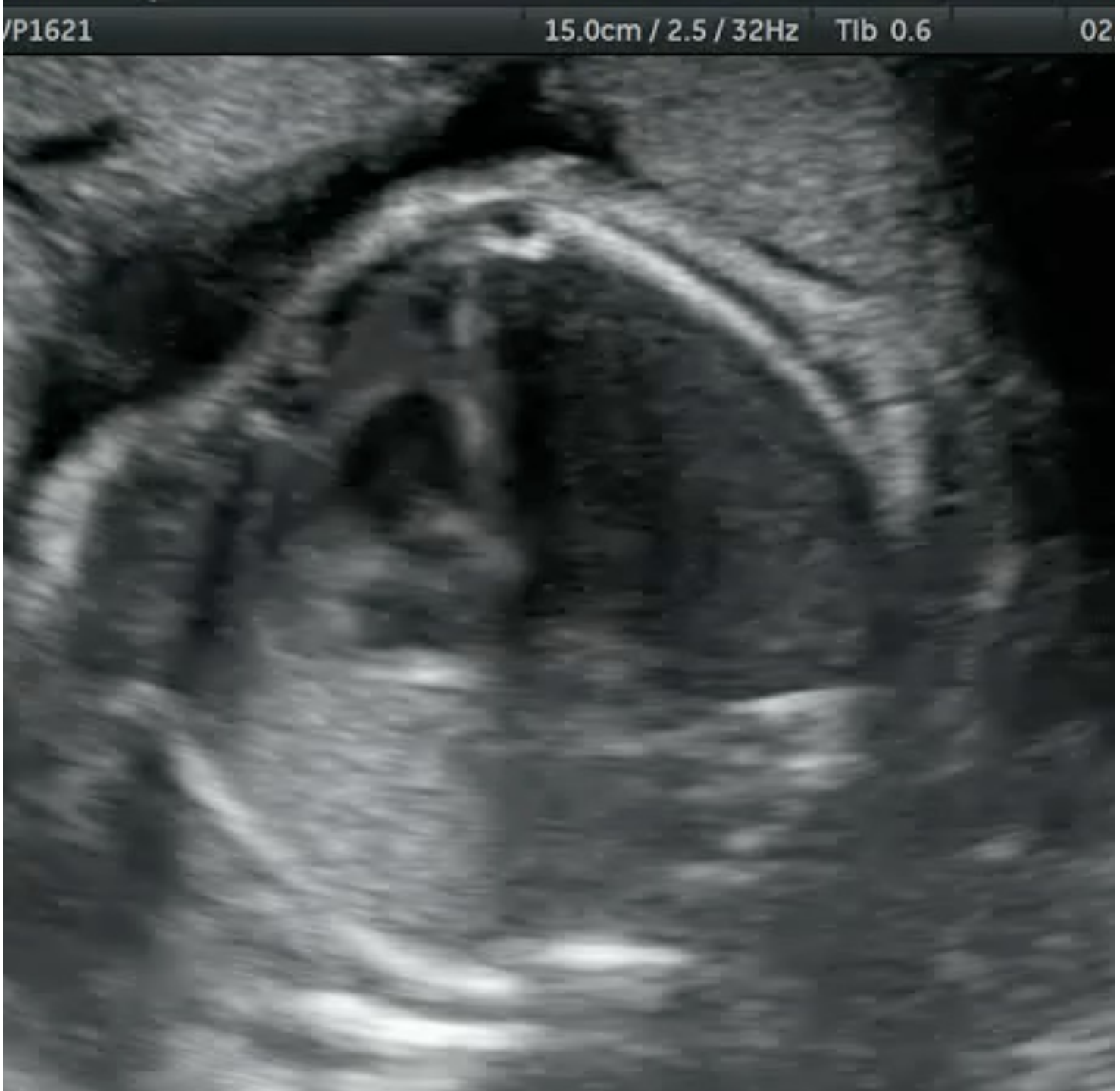
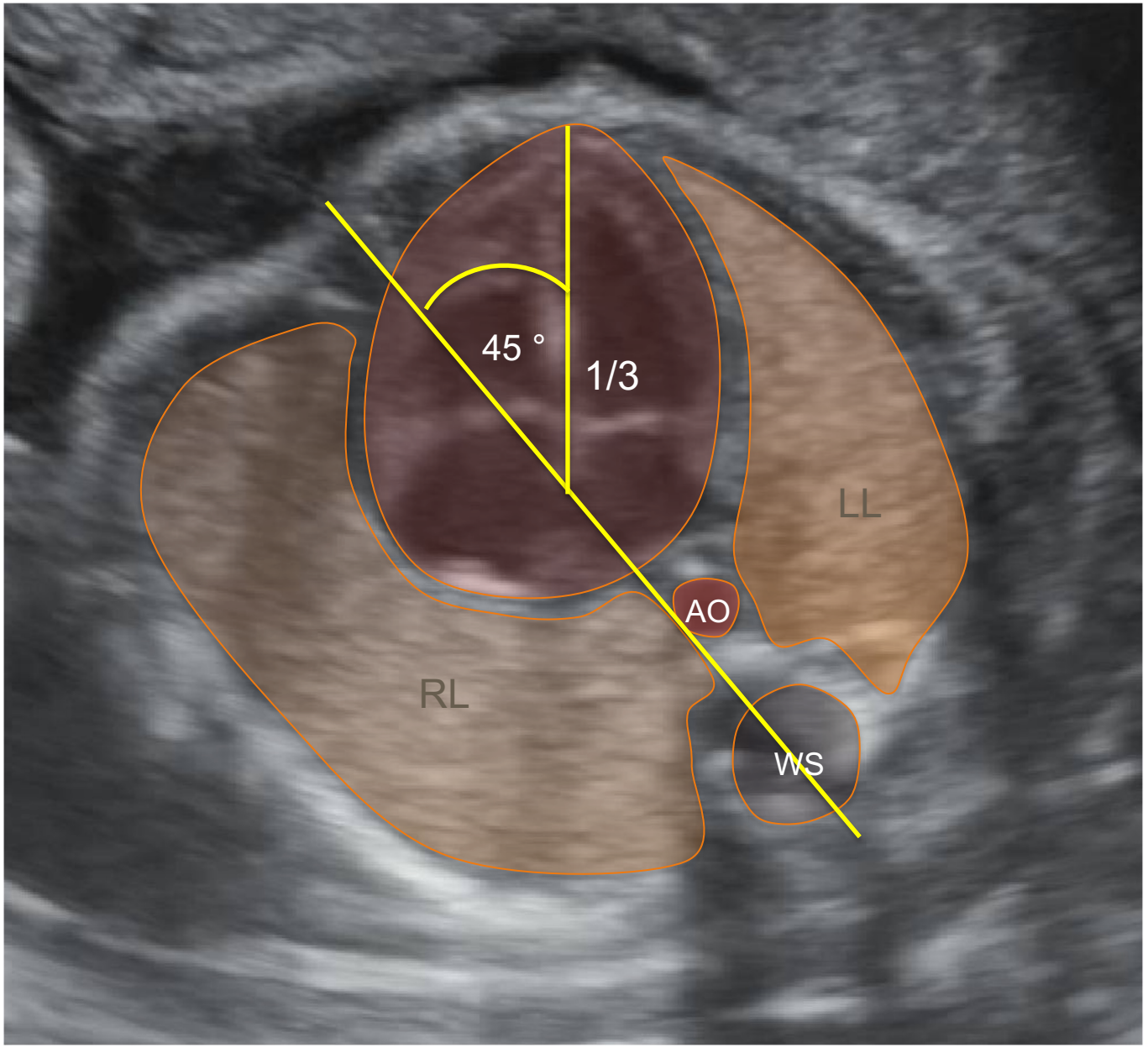
PRÄNATALDIAGNOSTIK

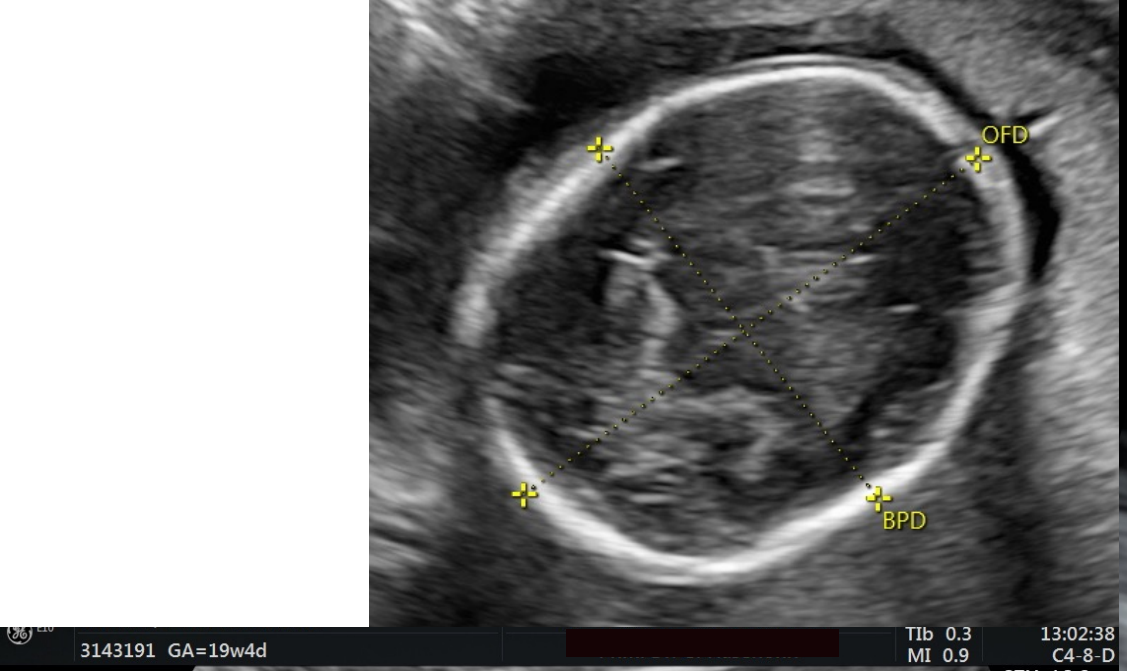
Fetale thorakale Probleme

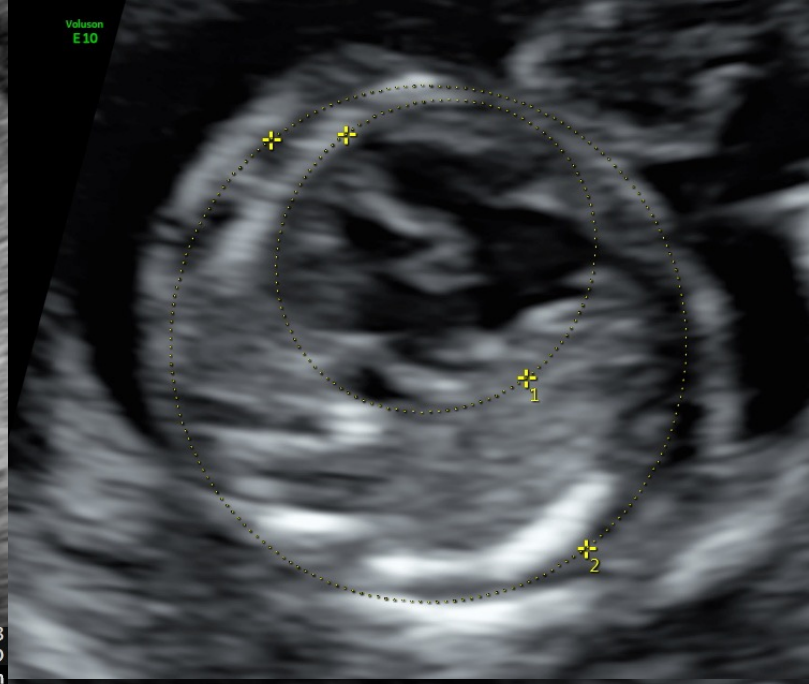
Fetale thorakale Probleme

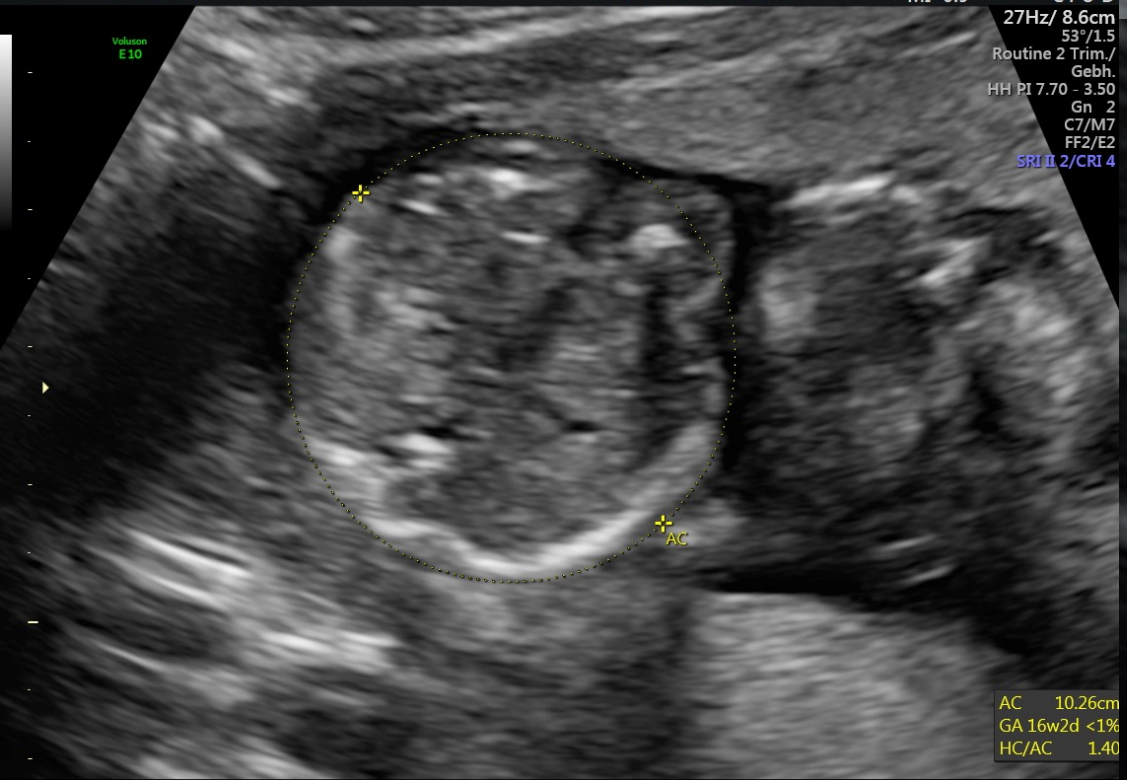
Horst Steiner
Praenamed Salzburg
Abteilung für Gynäkologie und Geburtshilfe A.ö. KH Braunau

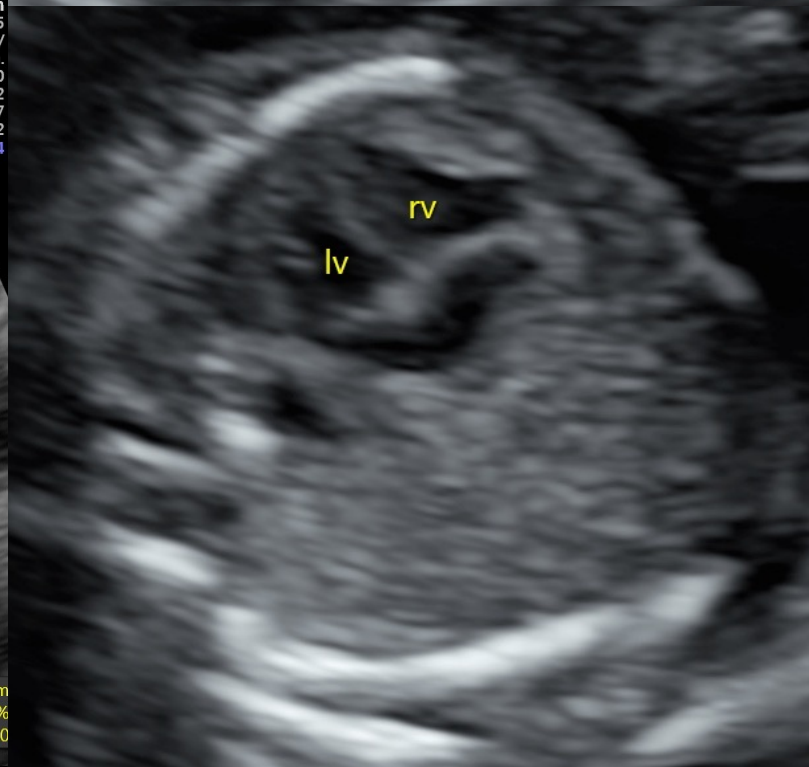

/P1621
15.0cm / 2.5 / 32Hz
TIb 0.6
02

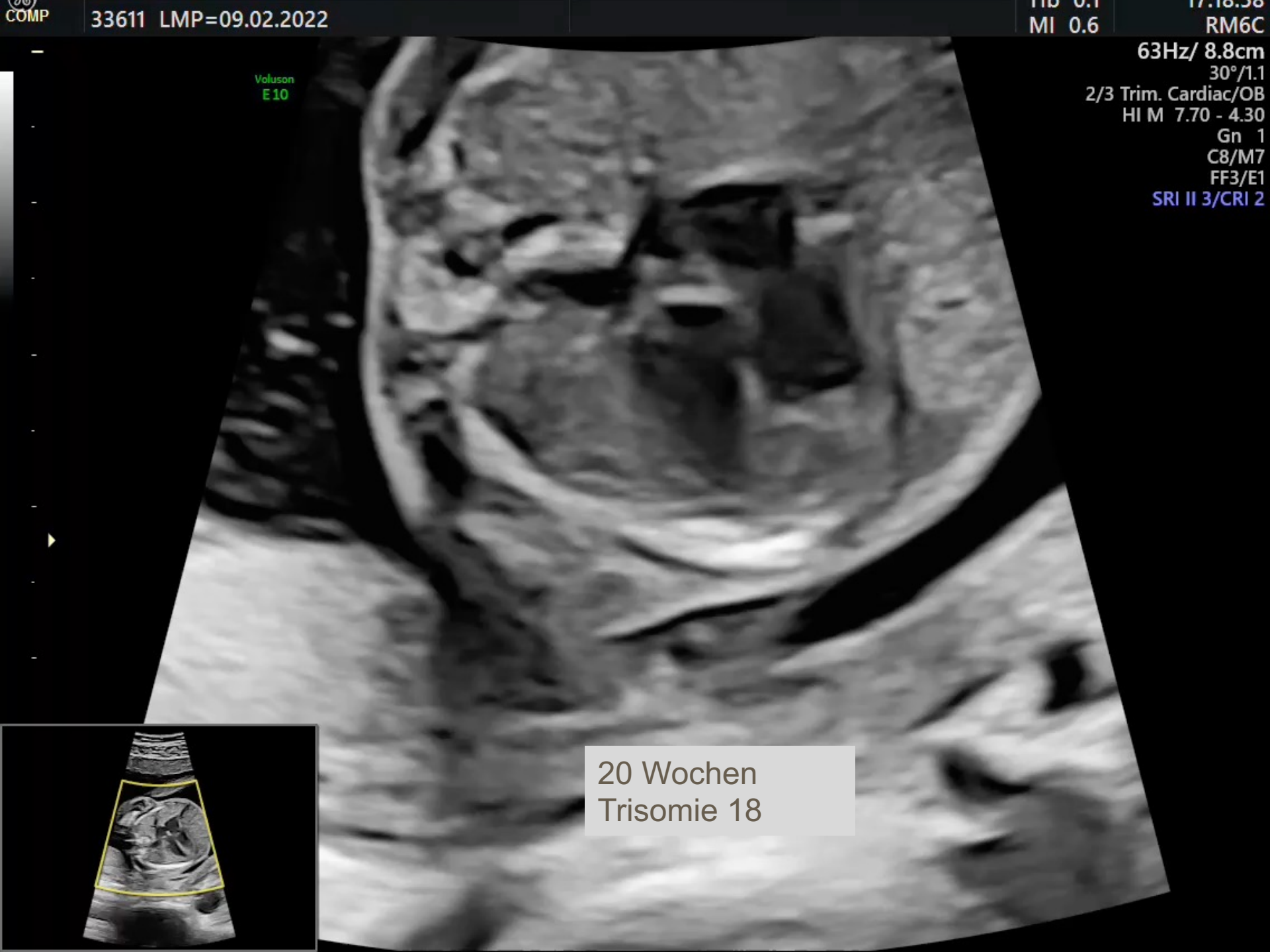

45 °
1/3
LL
AO
RL
WS


OFD
BPD
3143191 GA=19w4d
TIb 0.3
MI 0.9
13:02:38
C4-8-D
27Hz/ 8.6cm
53°/1.5
Routine 2 Trim./
Gebh.
HH PI 7.70 - 3.50
Gn 2
C7/M7
FF2/E2
SRI II 2/CRI 4


Voluson
E10
1
2


Voluson
E10
AC
AC 10.26cm
GA 16w2d <1%
HC/AC 1.40


rv
lv


COMP
33611 LMP=09.02.2022
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RM6C
63Hz/ 8.8cm
30°/1.1
2/3 Trim. Cardiac/OB
HI M 7.70 - 4.30
Gn 1
C8/M7
FF3/E1
SRI II 3/CRI 2
Voluson
E 10
20 Wochen
Trisomie 18

Herz-Lungen-Relation

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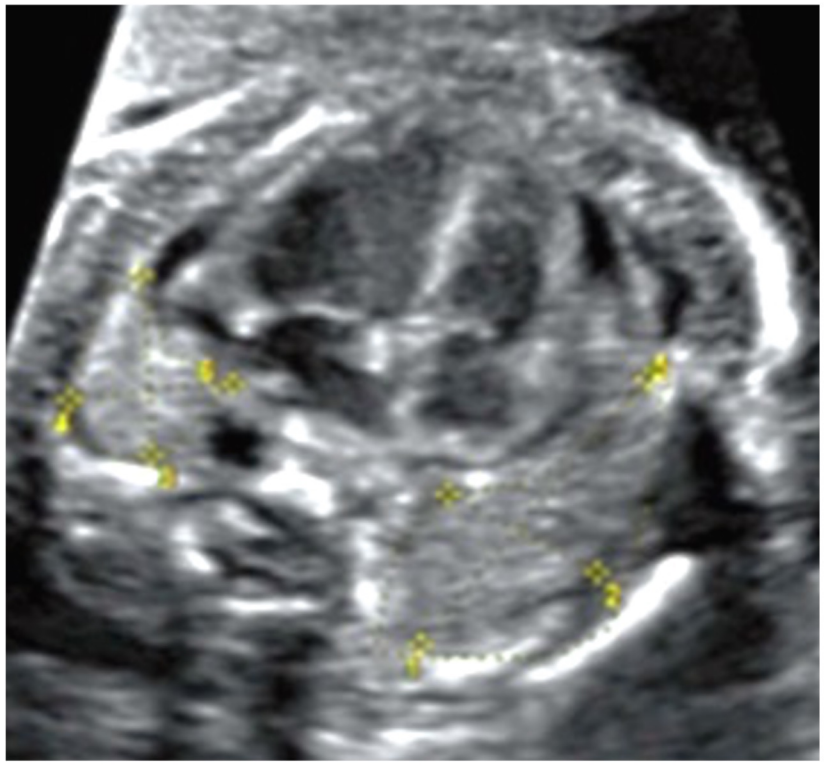
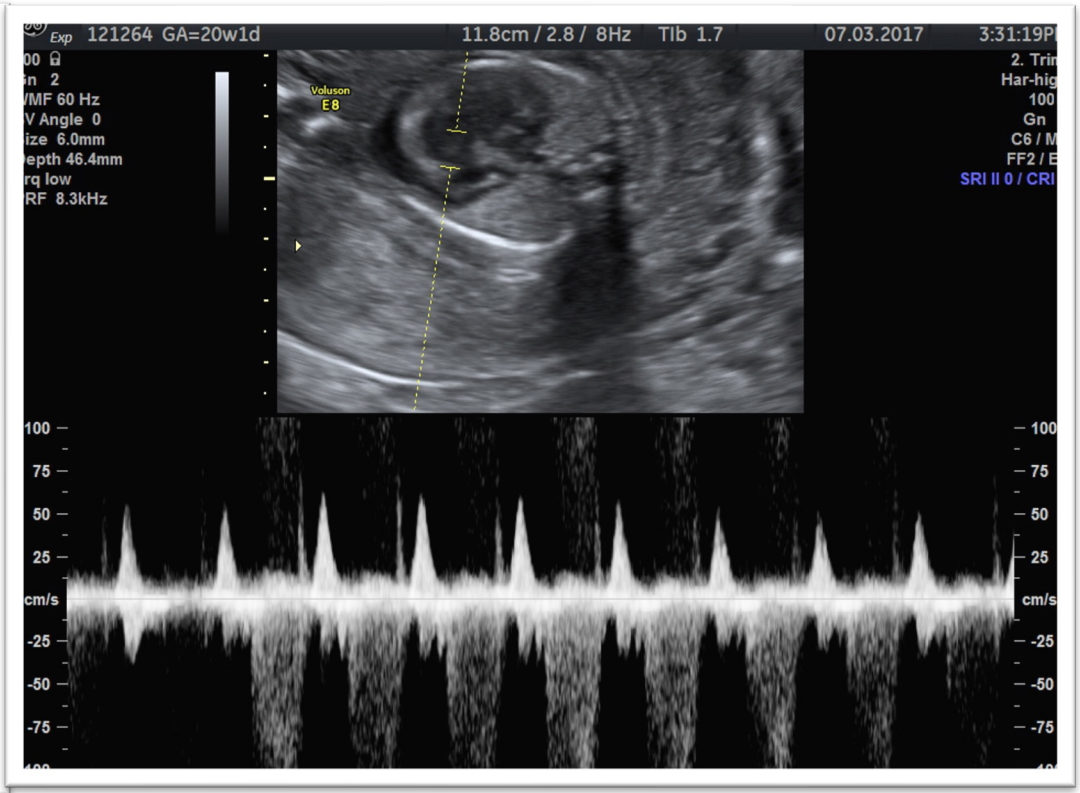
Primäre (bilaterale) Lungenhypoplasie

Dilatiertes Herz CMP





Thoraxhypoplasie

sagittaler Durchmesser

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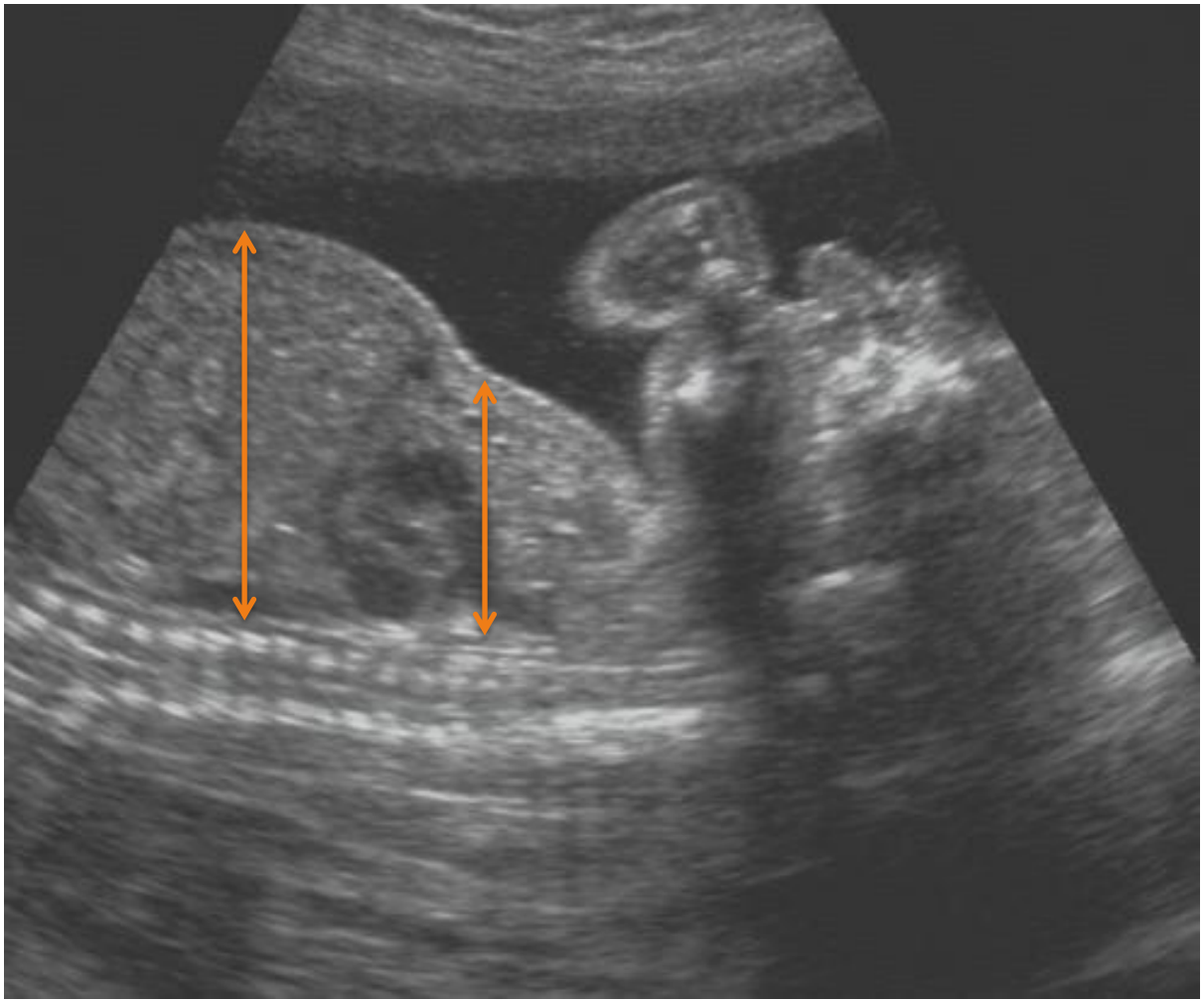
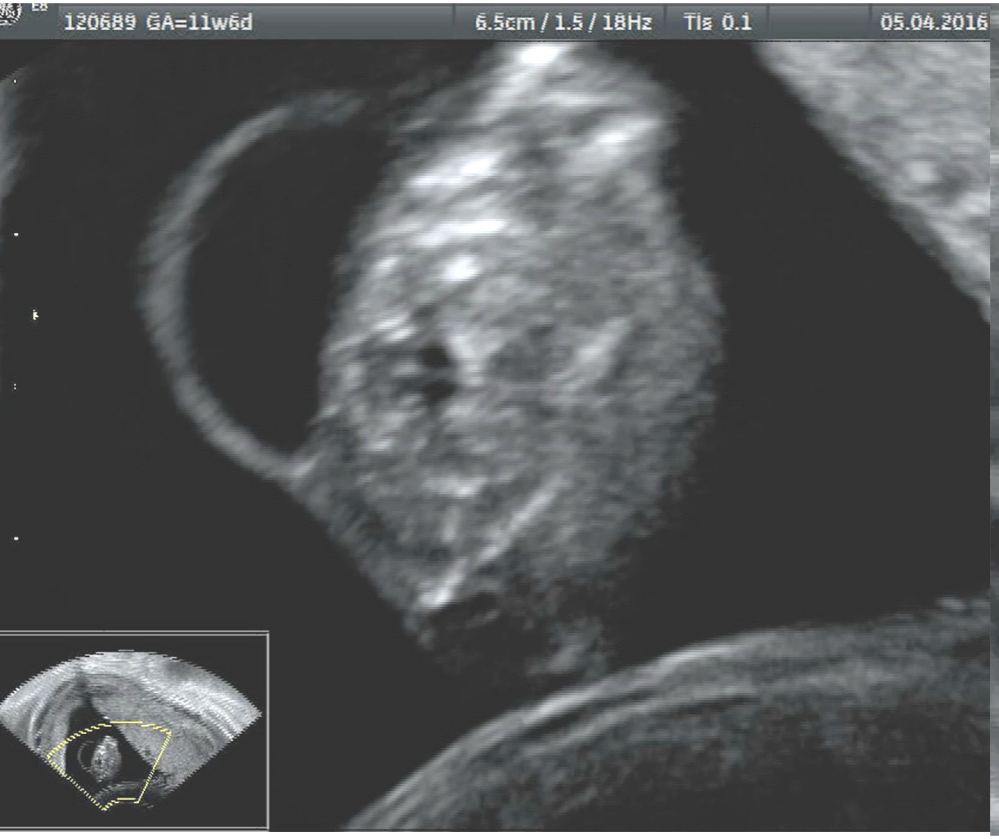
PRÄNATALDIAGNOSTIK

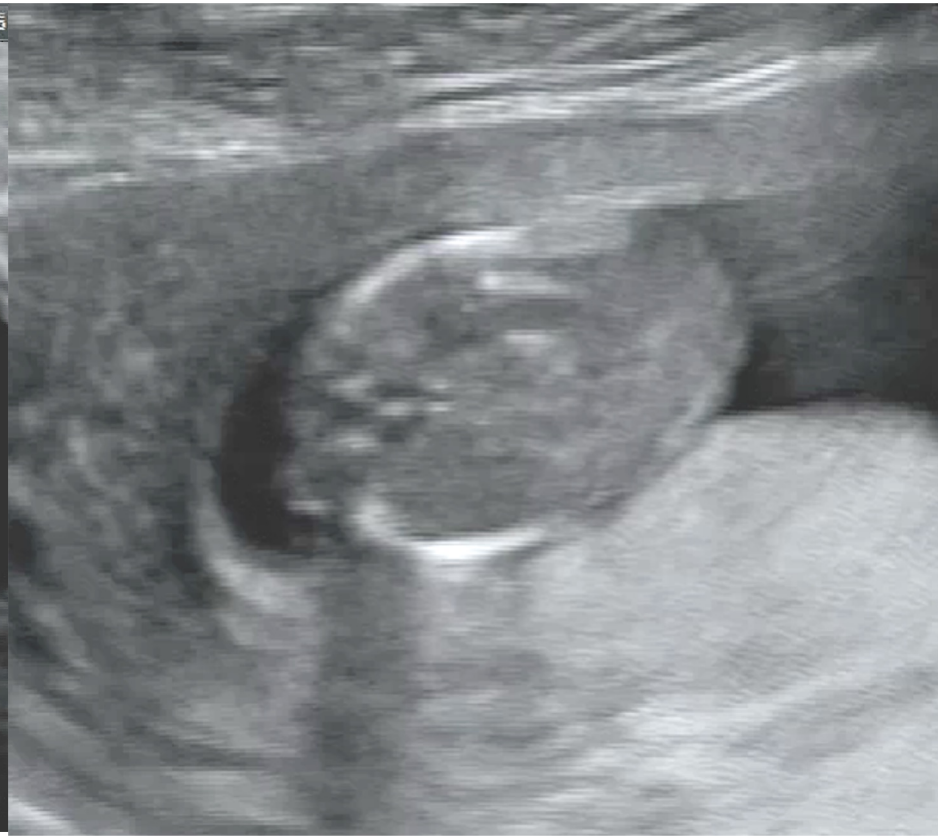


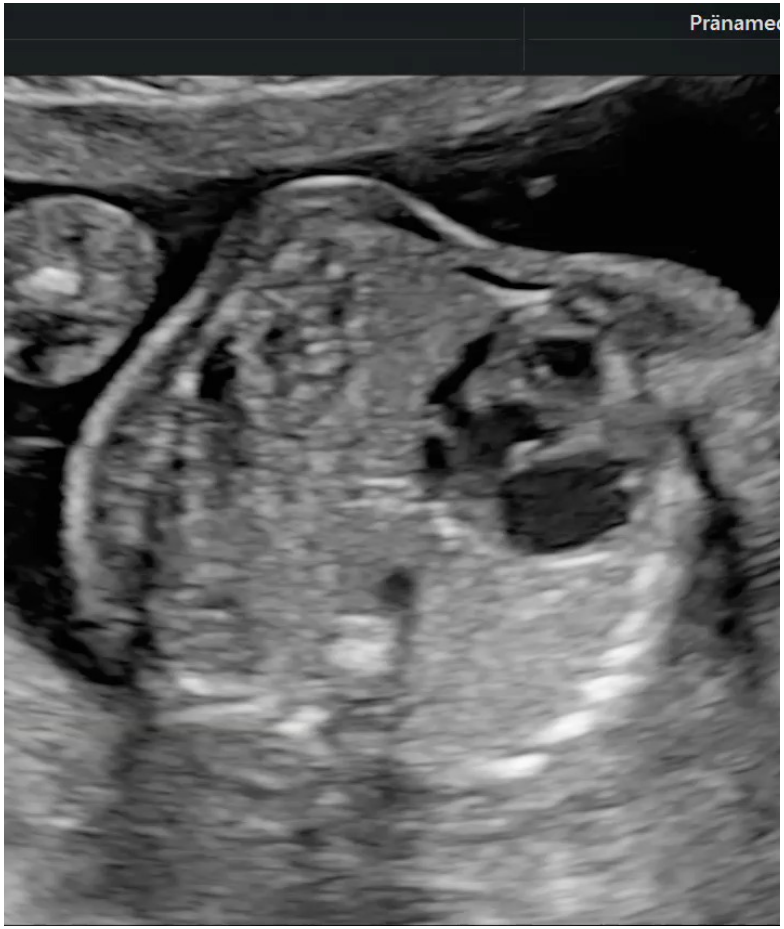
praena med
PRÄNATALDIAGNOSTIK

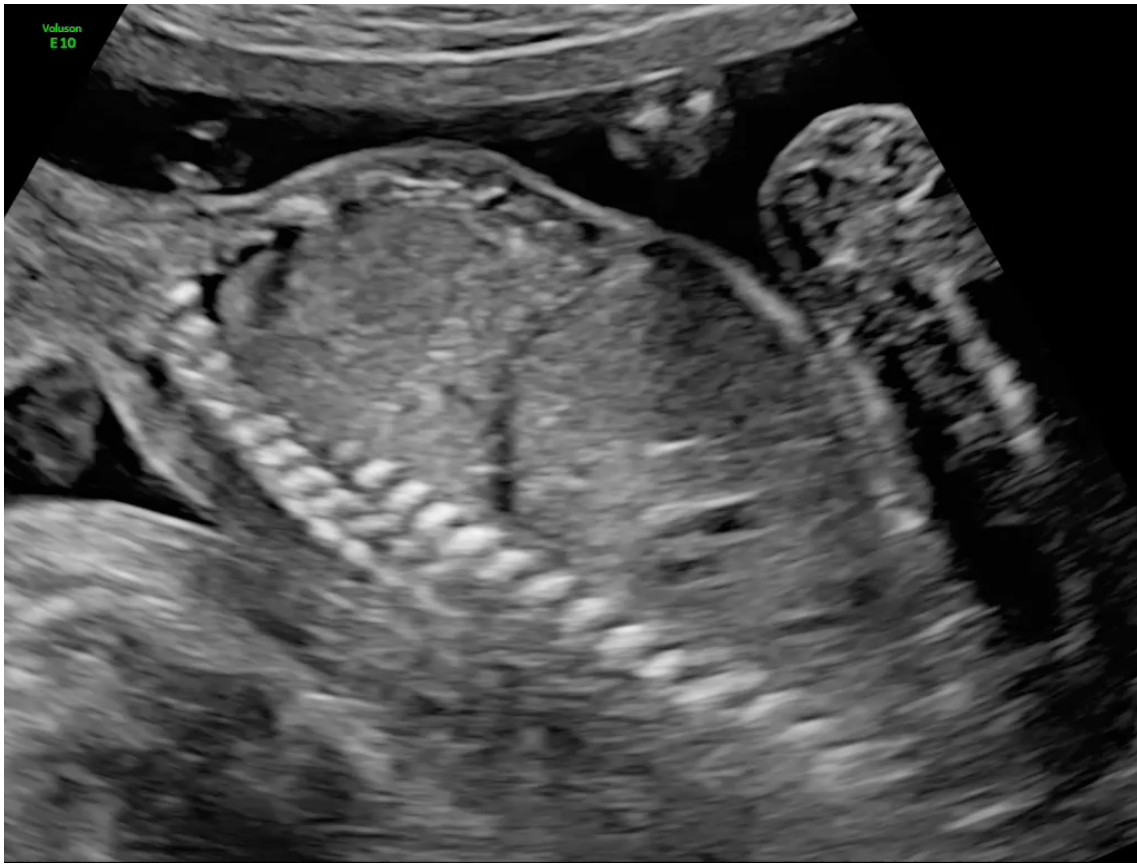
11+6

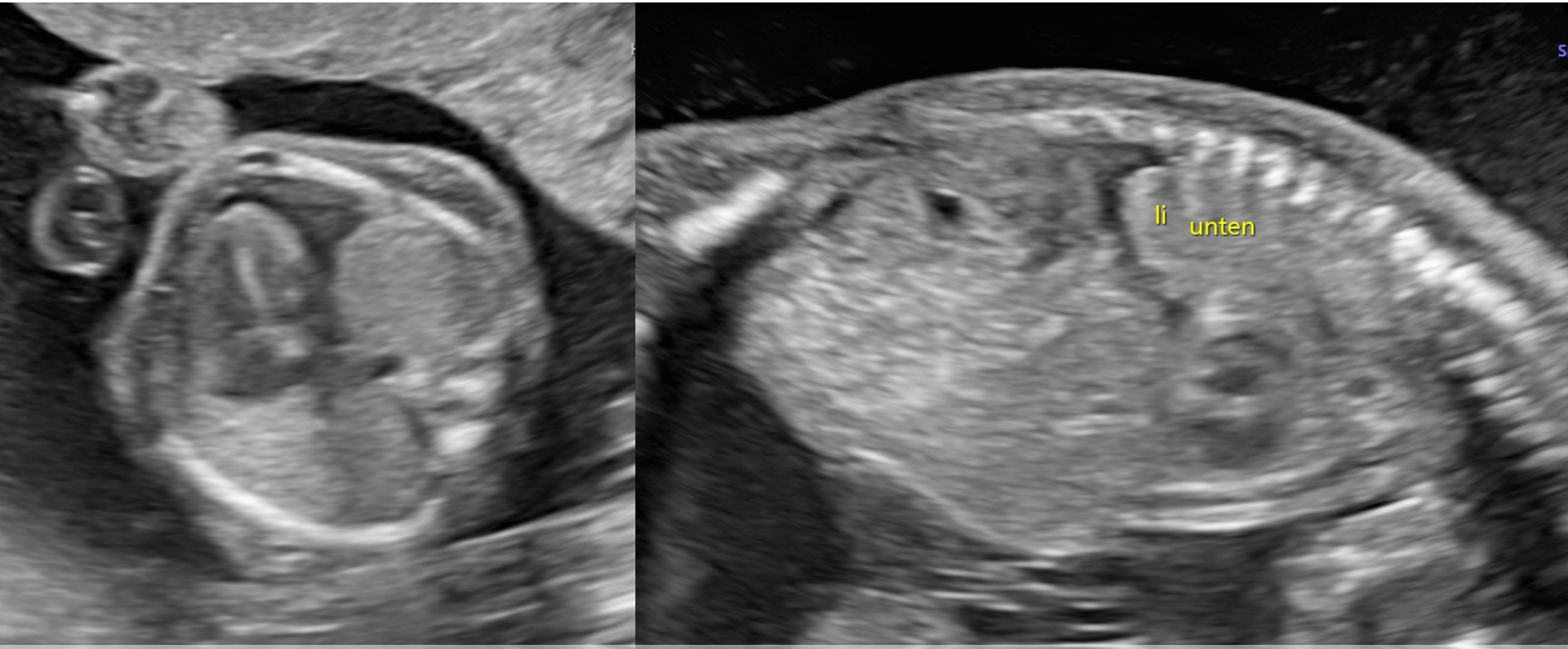


13+6




Pränamed


Voluson
E10


li
unten

Zwerchfellhernie (CDH)

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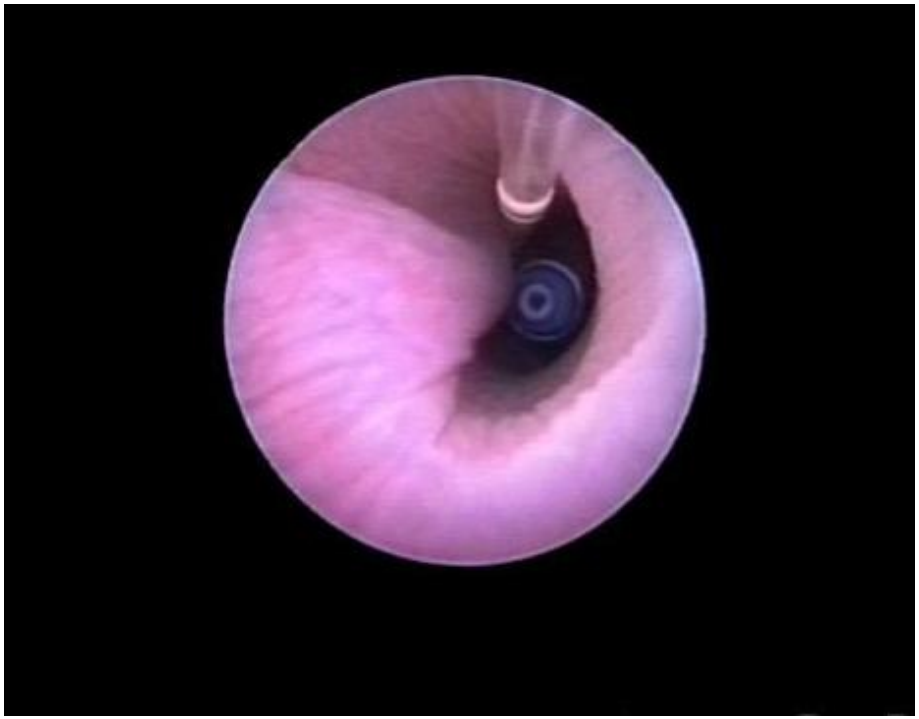
PRÄNATALDIAGNOSTIK

- Praenatal diagnostizierte CDH nehmen zu (Europa 50-70 %)
- Prognose:
  - Isolierte: Survival <15% für schwere Formen
  - 50% für moderate Erkrankungen
  - >90% für milde Formen
- Prognose hängt vom funktionellen Lungenvolumen ab
- Fetale Operationen (Plugs – Fetendo)

Zwerchfellhernie
Fetoskopische Plugs



Geipel und Hecher, Ultraschalldiagnostik in Geburtshilfe und Gynäkologie, Springer 2018

Prenatal management of the fetus with isolated congenital diaphragmatic hernia in the era of the TOTAL trial

Jan Deprest [a, b, *], Paul Brady [c], Kypros Nicolaides [b, d], Alexandra Benachi [b, e], Christoph Berg [b, f], Joris Vermeesch [c], Glenn Gardener [g], Eduard Gratacos [b, h]

Seminars in Fetal & Neonatal Medicine 19 (2014) 338–348

percutaneous fetal endoscopic tracheal occlusion (FETO) under local anesthesia. The Tracheal Occlusion To Accelerate Lung growth trial (www.totaltrial.eu) is an international randomized trial investigating the role of fetal therapy for severe and moderate pulmonary hypoplasia. Despite an apparent increase in

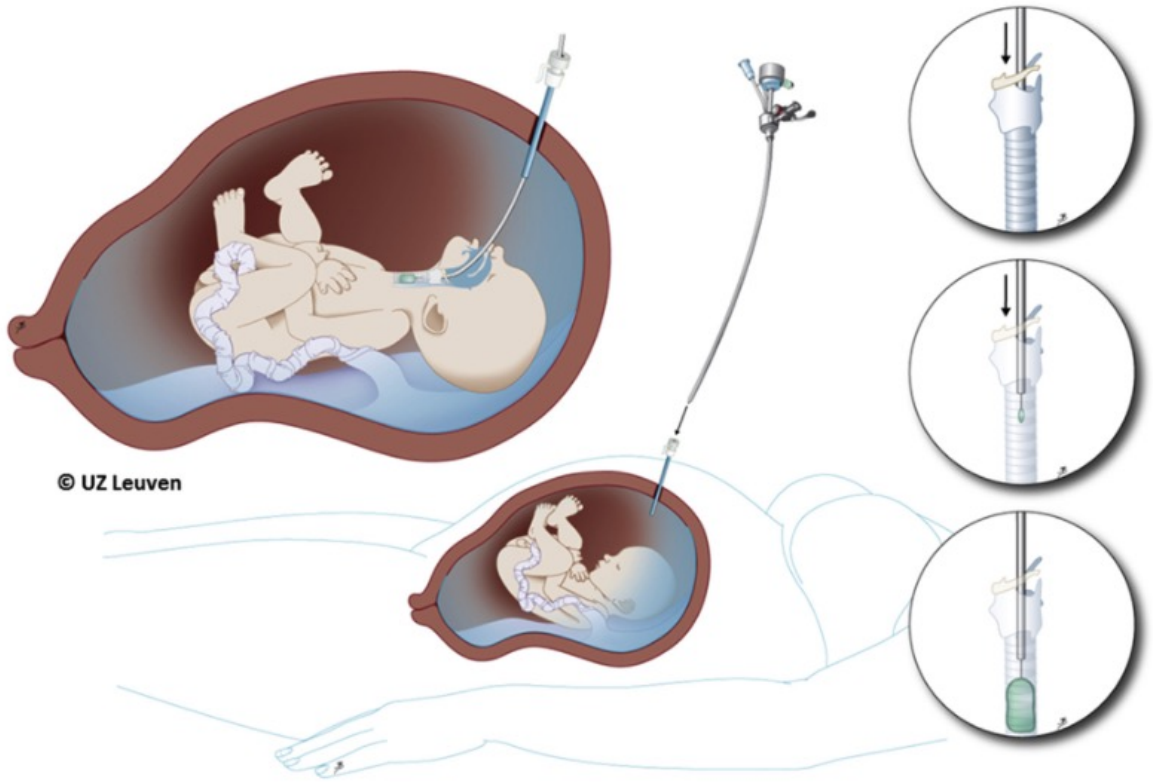


Fig. 3. Fetoscopic endoluminal tracheal occlusion (FETO). A. schematic drawing. Inserts: steps in balloon delivery. Reproduced with permission from UZ Leuven, Belgium. Video available as additional information.

Prenatal management of the fetus with isolated congenital diaphragmatic hernia in the era of the TOTAL trial

Jan Deprest [a, b, *], Paul Brady [c], Kypros Nicolaides [b, d], Alexandra Benachi [b, e], Christoph Berg [b, f], Joris Vermeesch [c], Glenn Gardener [g], Eduard Gratacos [b, h]

Seminars in Fetal & Neonatal Medicine 19 (2014) 338–348

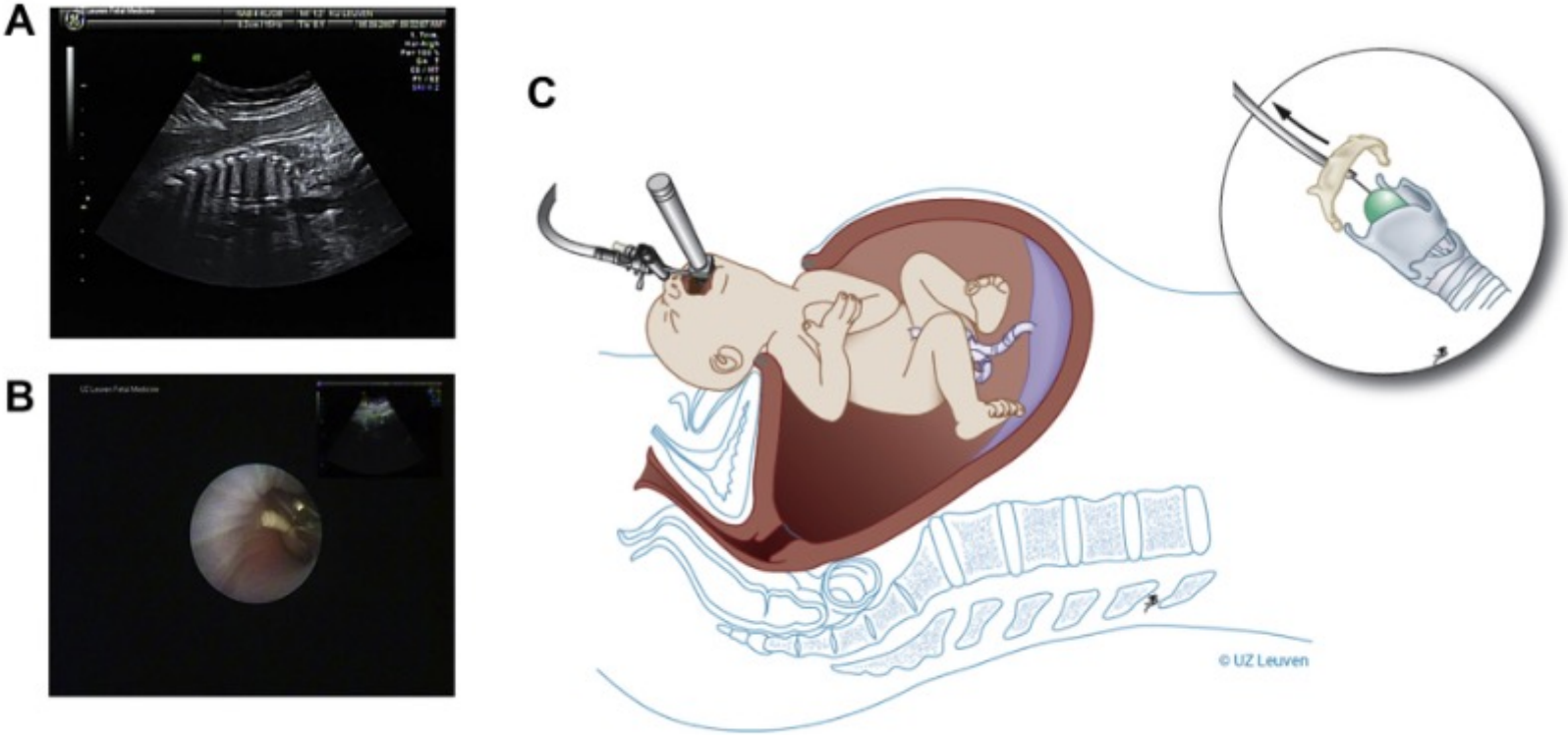


Fig. 5. Balloon removal: (A) ultrasound guided puncture of the balloon; (B) fetoscopic balloon retrieval and (C) schematic drawing of a retrieval on placental circulation (reproduced with permission of the UZ Leuven, Belgium). Videos available as additional files.

ORIGINAL ARTICLE

July 8, 2021
N Engl J Med 2021; 385:107-118

Randomized Trial of Fetal Surgery for Severe Left Diaphragmatic Hernia

Jan A. Deprest, M.D., Ph.D., Kypros H. Nicolaides, M.D., Alexandra Benachi, M.D., Ph.D., Eduard Gratacos, M.D., Ph.D., Greg Ryan, M.D., Nicola Persico, M.D., Ph.D., Haruhiko Sago, M.D., Ph.D., Anthony Johnson, M.D., Mirosław Wielgoś, M.D., Ph.D., Christoph Berg, M.D., Ph.D., Ben Van Calster, Ph.D., and Francesca M. Russo, M.D., Ph.D. for the TOTAL Trial for Severe Hypoplasia

CLINICAL PROBLEM

Observational studies have shown that fetoscopic endoluminal tracheal occlusion (FETO) is associated with increased survival among infants with severe pulmonary hypoplasia due to isolated congenital diaphragmatic hernia on the left side, but data from randomized trials are lacking.

CLINICAL TRIAL

Design: An open-label, randomized, controlled trial was conducted to compare FETO with expectant care among women carrying singleton fetuses with isolated severe congenital diaphragmatic hernia on the left side.

Intervention: 95 women underwent randomization; 47 were assigned to undergo FETO at 27 to 29 weeks of gestation, and 48 were assigned to expectant care. The primary outcome was infant survival to discharge from the neonatal intensive care unit.

RESULTS

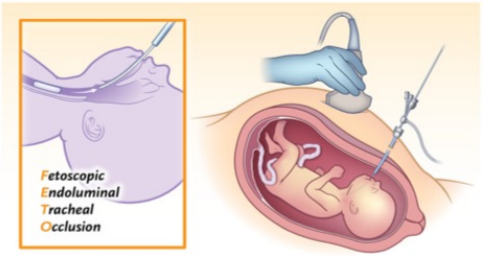
Efficacy: As compared with expectant care, FETO improved survival to discharge, and the benefit was sustained to 6 months of age. The trial was stopped early for efficacy after the third interim analysis.

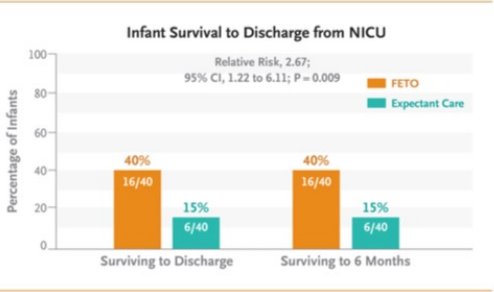
Safety: FETO increased the risks of preterm, prelabor rupture of membranes and preterm birth.

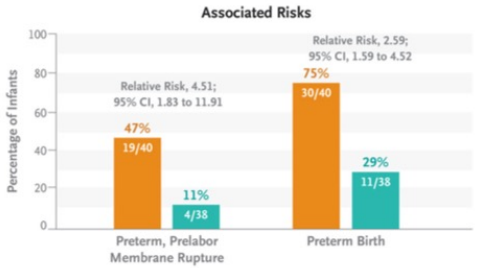
LIMITATIONS AND REMAINING QUESTIONS

- Data on longer-term outcomes are needed, including neurodevelopmental outcomes.
- These results should not be generalized to centers without extensive experience in fetoscopy and FETO.

Links: Full article | NEJM Quick Take | Editorial







CONCLUSIONS

The use of FETO in fetuses with isolated severe congenital diaphragmatic hernia on the left side resulted in increased survival to hospital discharge but an increased risk of preterm, prelabor rupture of membranes and preterm birth.

Severe:
defined as a quotient of the observed-to-expected lung-to-head ratios of less than 25.0%, irrespective of liver position

ORIGINAL ARTICLE

July 8, 2021

N Engl J Med 2021; 385:119-129

Randomized Trial of Fetal Surgery for Moderate Left Diaphragmatic Hernia

Jan A. Deprest, M.D., Ph.D., Alexandra Benachi, M.D., Ph.D., Eduard Gratacos, M.D., Ph.D., Kypros H. Nicolaides, M.D., Christoph Berg, M.D., Ph.D., Nicola Persico, M.D., Ph.D., Michael Belfort, M.D., Ph.D., Glenn J. Gardener, M.D., Ph.D., Yves Ville, M.D., Ph.D., Anthony Johnson, M.D., Francesco Morini, M.D., Ph.D., Mirosław Wielgoś, M.D., Ph.D., et al., for the TOTAL Trial for Moderate Hypoplasia Investigators*

CONCLUSIONS

This trial involving fetuses with moderate congenital diaphragmatic hernia on the left side did not show a significant benefit of FETO performed at 30 to 32 weeks of gestation over expectant care with respect to survival to discharge or the need for oxygen supplementation at 6 months. FETO increased the risks of preterm, prelabor rupture of membranes and preterm birth. (Funded by the European Commission and others; TOTAL ClinicalTrials.gov number,

Ultraschallbefunde die mit schlechtem Outcome bei isolierter linksseitiger CDH assoziiert sind

Fehlbildungen, Chromosomenanomalien

- Leber im Thorax
- Ausmaß des Mediastinalshifts
- Vorhandensein eines Polyhydramnions
- Lung to head ratio < 1.00
- Frühe praenatale Diagnose
- Erhöhte NT beim ETS 11-14 Wochen
- Kleine Lungen im MRI

- Deprest, NEJM 2021:

- Leber im Thorax
- Lungengröße: Quotient of the observed-to-expected lung-to-head ratios (< 25%)

Ultrasound Obstet Gynecol 2021; 57: 378–385
Published online in Wiley Online Library (wileyonlinelibrary.com). **DOI:** 10.1002/uog.23115

Fetal endoscopic tracheal occlusion reverses the natural history of right-sided congenital diaphragmatic hernia: European multicenter experience

F. M. RUSSO[1,2], A.-G. CORDIER[3,4], D. BASURTO[2], L. SALAZAR[5], E. LITWINSKA[6], O. GOMEZ[5], A. DEBEER[2,7], J. NEVOUX[8], S. PATEL[9], L. LEWI[1,2], A. PERTIERRA[10], M. AERTSEN[11,12], E. GRATACOS[5], K. H. NICOLAIDES[6], A. BENACHI[3,4] and J. DEPREST[1,2]

CONTRIBUTION

What are the novel findings of this study?
In right-sided congenital diaphragmatic hernia (RCDH), survival can be predicted by prenatal measurement of lung size and, in cases with the worst prognosis, is improved by fetal endoscopic tracheal occlusion (FETO).

What are the clinical implications of this work?
In cases with RCDH and severe lung hypoplasia, defined as observed-to-expected lung-to-head ratio $\leq 45\%$ or $\leq 50\%$, FETO is associated with significantly improved survival without increased neonatal morbidity.

DD Diaphragmahernie

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JOGC Journal of Obstetrics and Gynaecology Canada
Journal d'obstétrique et gynécologie du Canada

CASE REPORT · ÉTUDE DE CAS | VOLUME 43, ISSUE 8, P993-997, AUGUST 01, 2021

Fetal Diaphragmatic Eventration: A Case Report

Bahauddin Sallout, MBBS • Danya Alshebli, MBBS • Luai Sallout, MBBS • Badi Al Baqawi, MBBS • Majed S. Faden, MD

Published: January 04, 2021 • DOI: https://doi.org/10.1016/j.jogc.2020.12.014

Case Reports

Congenital Cystic Diaphragm with Diaphragmatic Eventration in a Fetus: A Case Presentation

Li Zhen, Cong-Min Gu, Lv-Yin Huang & **Dong-Zhi Li**

Pages 335-339 | Received 15 Jan 2019, Accepted 21 Feb 2019, Published online: 26 Mar 2019

Download citation https://doi.org/10.1080/15513815.2019.1588440 Check for updates

Hydrothorax

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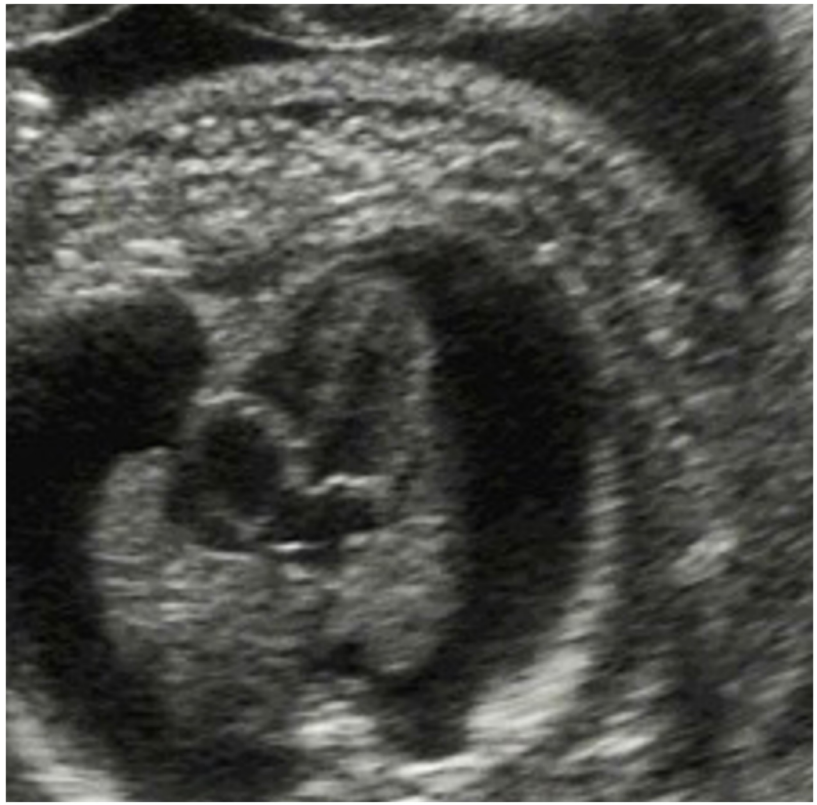

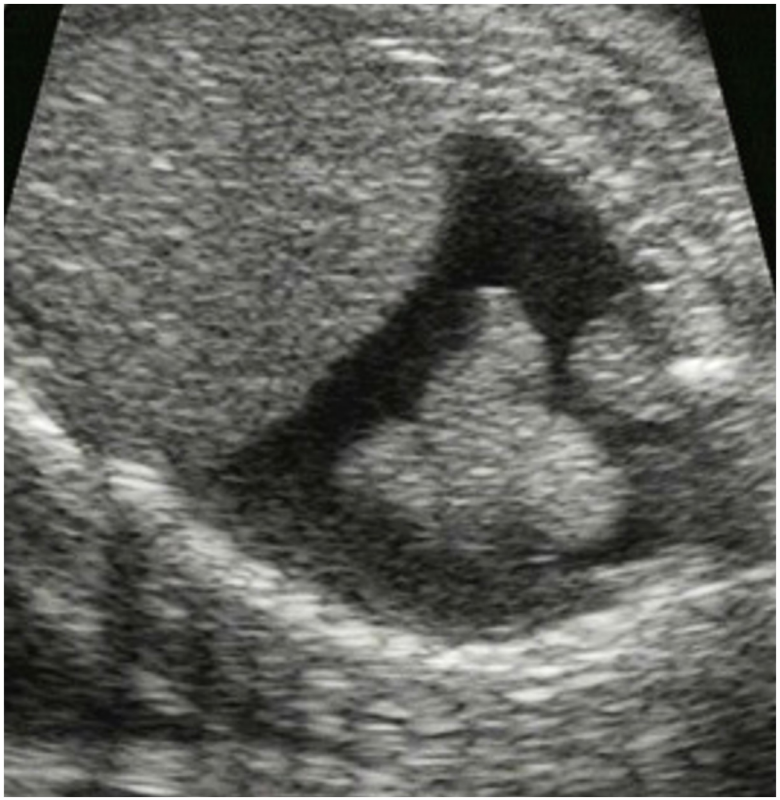

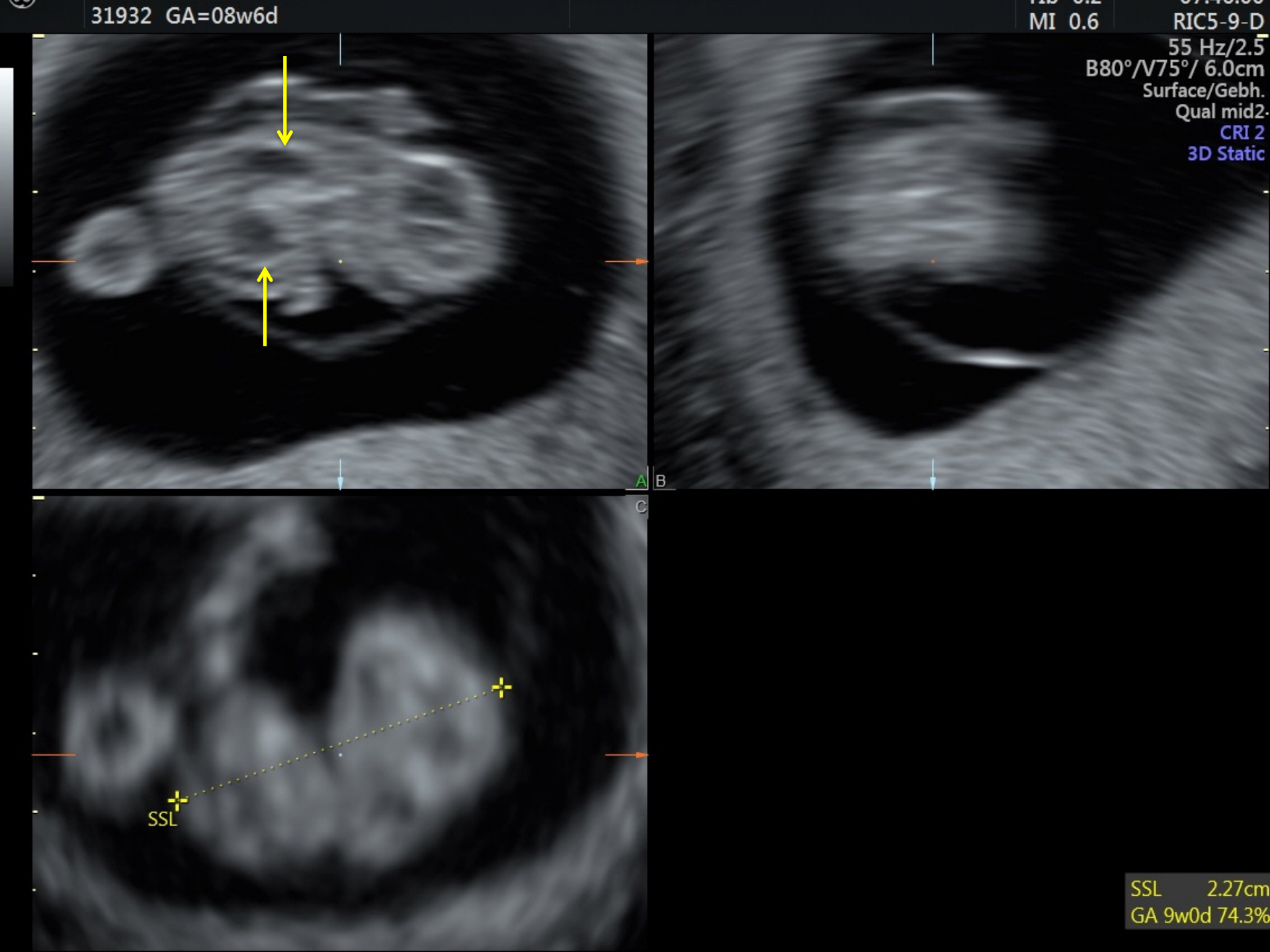
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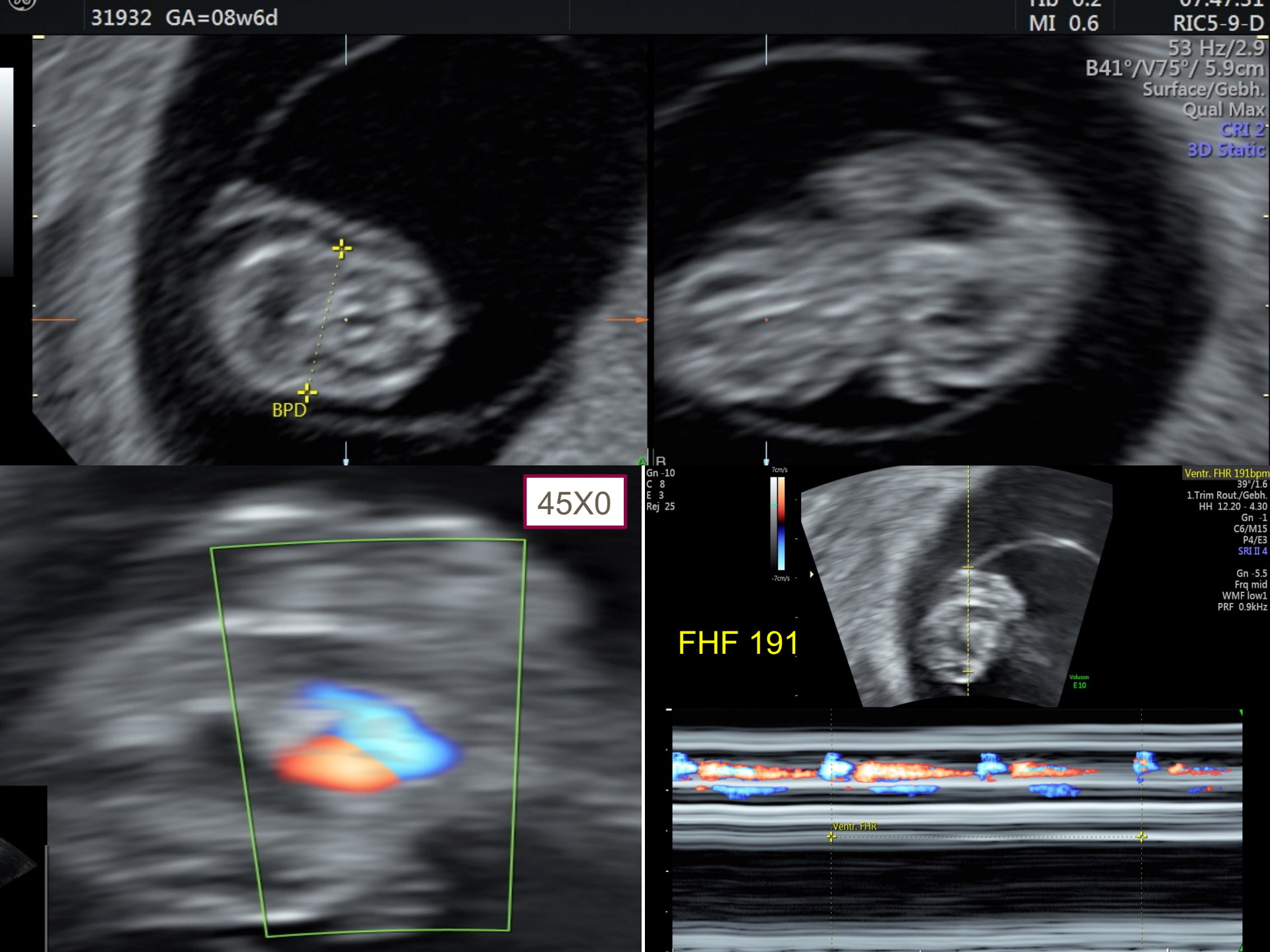
Surface/Gebh.
Qual mid2
CRI 2
3D Static
A
B
C
SSL
SSL 2.27cm
GA 9w0d 74.3%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RIC5-9-D
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B41°/V75°/ 5.9cm
Surface/Gebh.
Qual Max
CRI 2
3D Static
BPD
45X0
Gn -10
C 8
E 3
Rej 25
7cm/s
-7cm/s
Ventr. FHR 191bpm
39°/1.6
1.Trim Rout./Gebh.
HH 12.20 - 4.30
Gn -1
C6/M15
P4/E3
SRI II 4
Gn -5.5
Frq mid
WMF low1
PRF 0.9kHz
FHF 191
Ventr. FHR

Praenatales Management Hydrothorax

- Ausschluss IHF, Infektionen (Parvovirus) und fetomaternale Transfusion, fetale Anämie sowie komplexe Fehlbildungen
- Syndrome (z.B. Noonan Syndrom, T21, Turner Syndrom)
- Drainage des Pleuraergusses durch Punktion oder (besser) Shunting erhöht overall survival rate auf 60-70%

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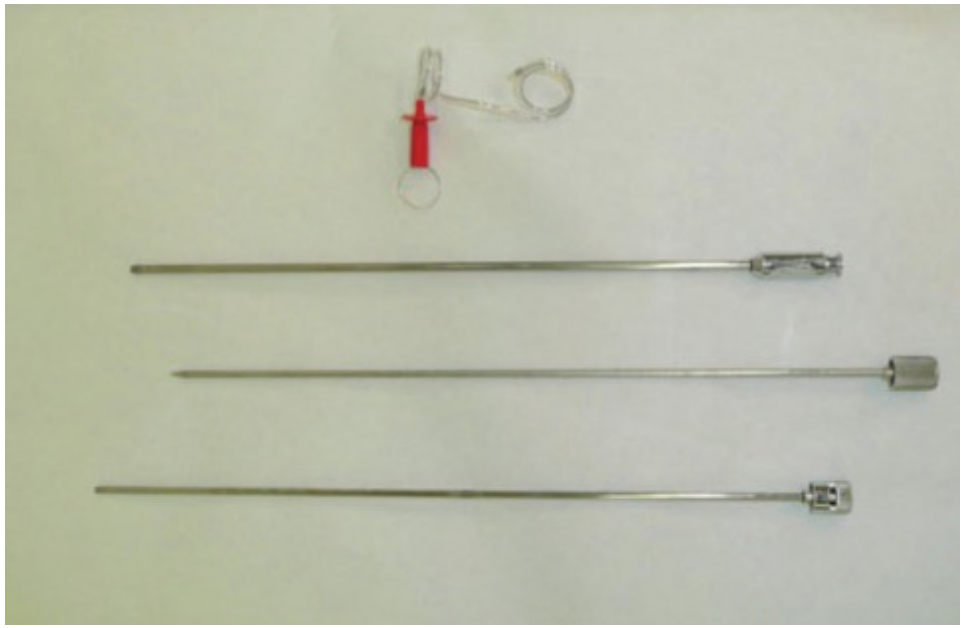
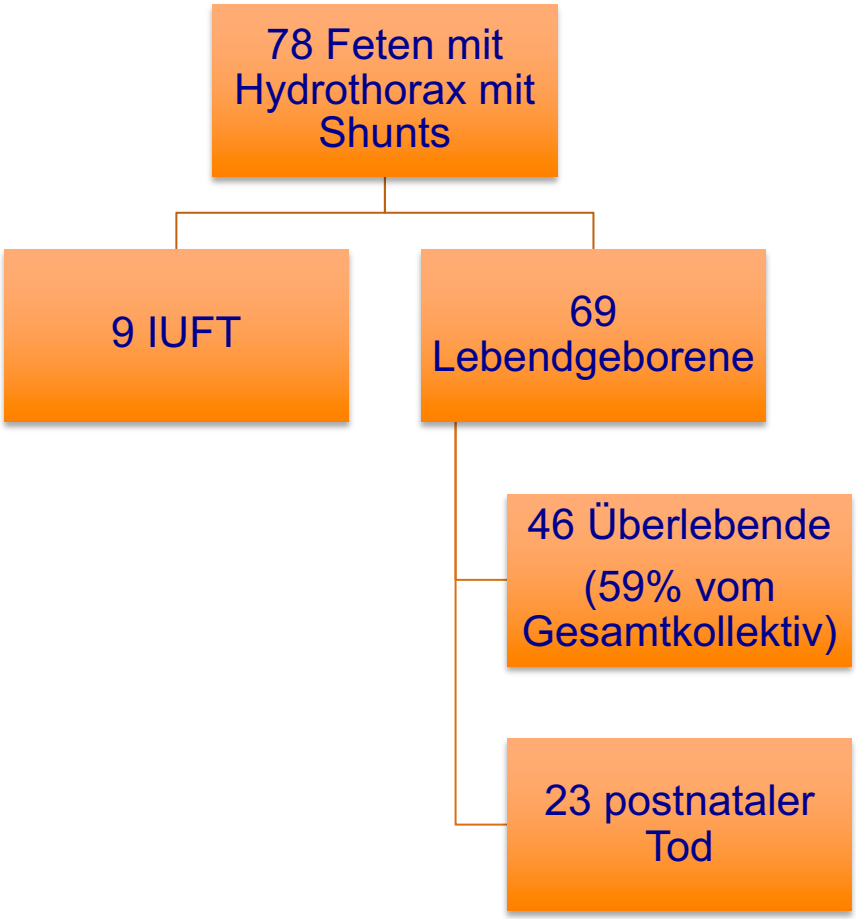
Shunts



Somatex

Thoracoamniotic Shunting for Fetal Hydrothorax: Predictors of Intrauterine Course and Postnatal Outcome

Michael R. Mallmann[a] Viola Graham[a] Bettina Rösing[a] Ingo Gottschalk[c]
Andreas Müller[b] Ulrich Gembruch[a] Annegret Geipel[a] Christoph Berg[a, c]
Fetal Diagn Ther 2017;41:58–65



- Zeitraum 2002 – 2011
- GA bei Diagnose (MW): 25,6 SSW(12-34)
- GA bei Shunteinlage (MW): 26,5 SSW (16-33)
- Im Mittel 2.53 shunts eingelegt (1-7) pro Fet

Thoracoamniotic Shunting for Fetal Hydrothorax: Predictors of Intrauterine Course and Postnatal Outcome

praena PRÄNATALDIAGNOSTIK med

Michael R. Mallmann[a] Viola Graham[a] Bettina Rösing[a] Ingo Gottschalk[c]
Andreas Müller[b] Ulrich Gembruch[a] Annegret Geipel[a] Christoph Berg[a, c]
Fetal Diagn Ther 2017;41:58–65

- Neg. prognostische Marker (für Nicht-Überleben)
  - Polyhydramnion
  - Hydrops placenta
  - Mediastinalshift beim ersten Schall
  - Hydropsentstehung nach Shunteinlage
  - Vorz. Blasensprung
  - Shunt-Geburt Intervall < 4 Wochen
  - Niedriges Gestationsalter
- T 21 Feten hatten höhere Überlebensrate

Short- and long-term outcome following thoracoamniotic shunting for fetal hydrothorax

E. N. Kelly ✉, G. Seaward, X. Y. Ye, R. Windrim, T. Van Mieghem ... See all authors

Ultrasound Obstet Gynecol April 2021

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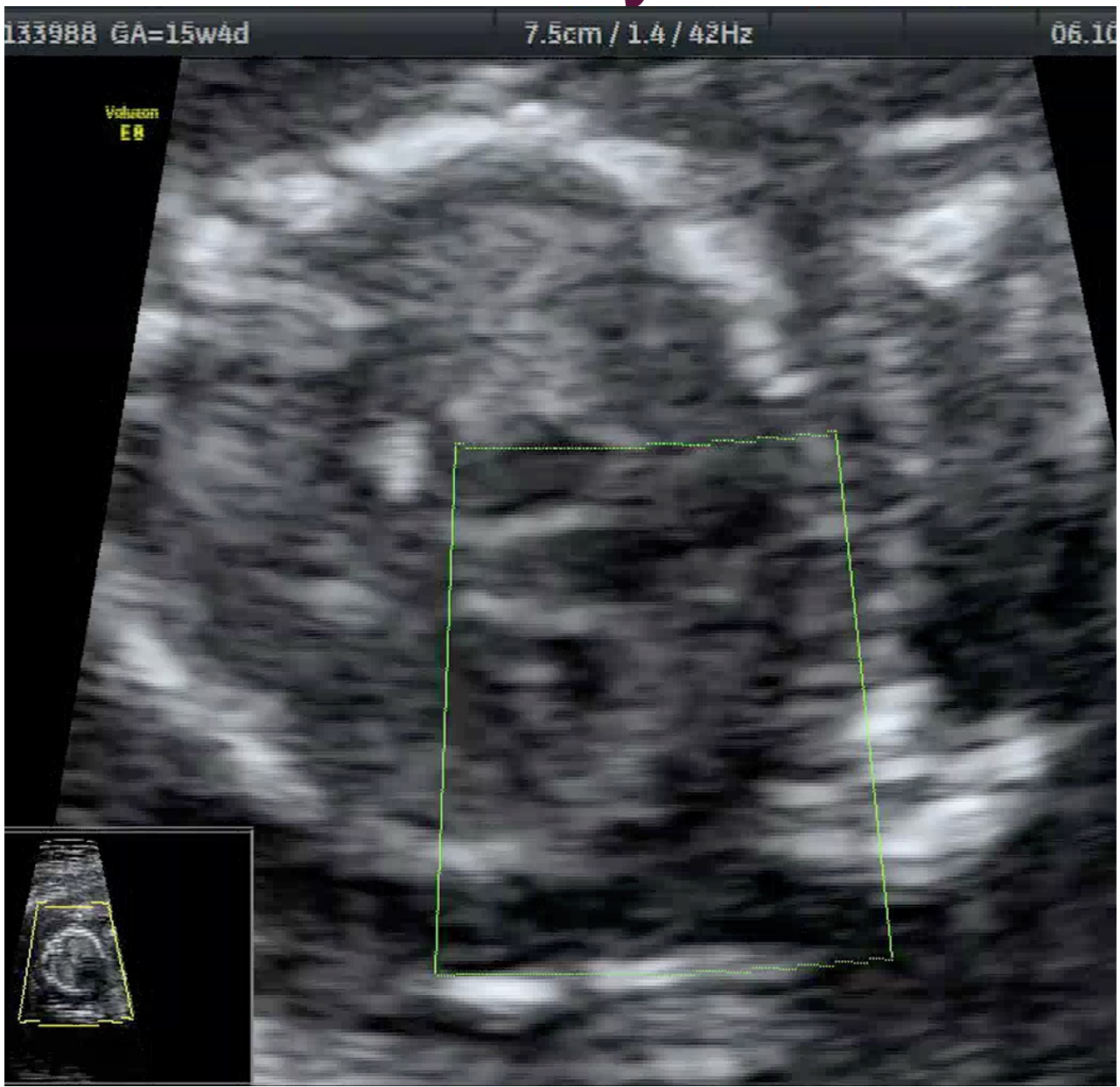
- 132 Feten mit Shunts, 1991 – 2014, single center
- Mittleres GA beim Shunt 25,6 SSW, bei Geburt 35.4 SSW
- 61 % hydropisch, 69% bilaterale Ergüsse
- 35 % intrauteriner oder neonataler Tod
- 65 % Überlebende:
  - 84% normale Entwicklung nach 18 Monaten
  - 92% der isolierten Hydrothoraces neurologisch unauffällig
  - 89 % ohne pulmonale Komplikationen
- Schlechte Prädiktion des Outcomes zum Zeitpunkt der Diagnosestellung

Hydrothorax



Hydrothorax
SGA
DPH ?

46 xx
Array CGH:
Del 7p21.3
Del 10q23.1
(In der Lit. bis dato nicht beschrieben)

Abbruch

Echogene und zystische Läsionen

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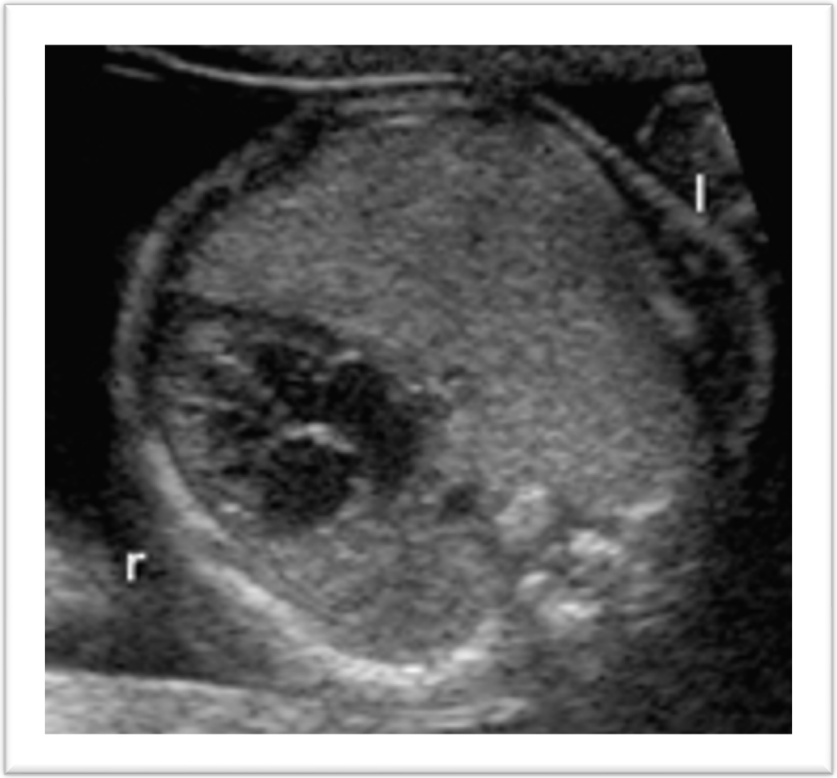
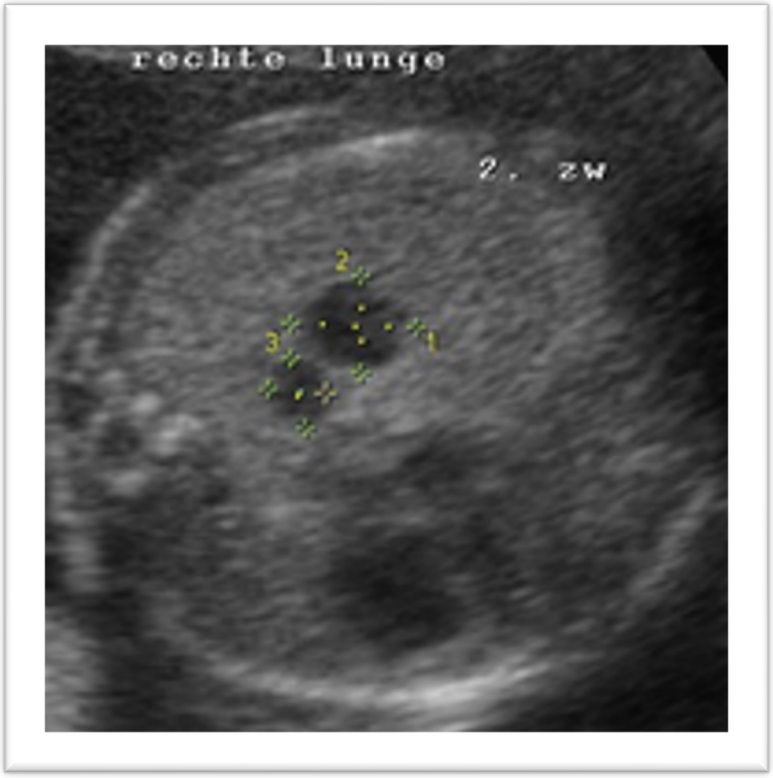
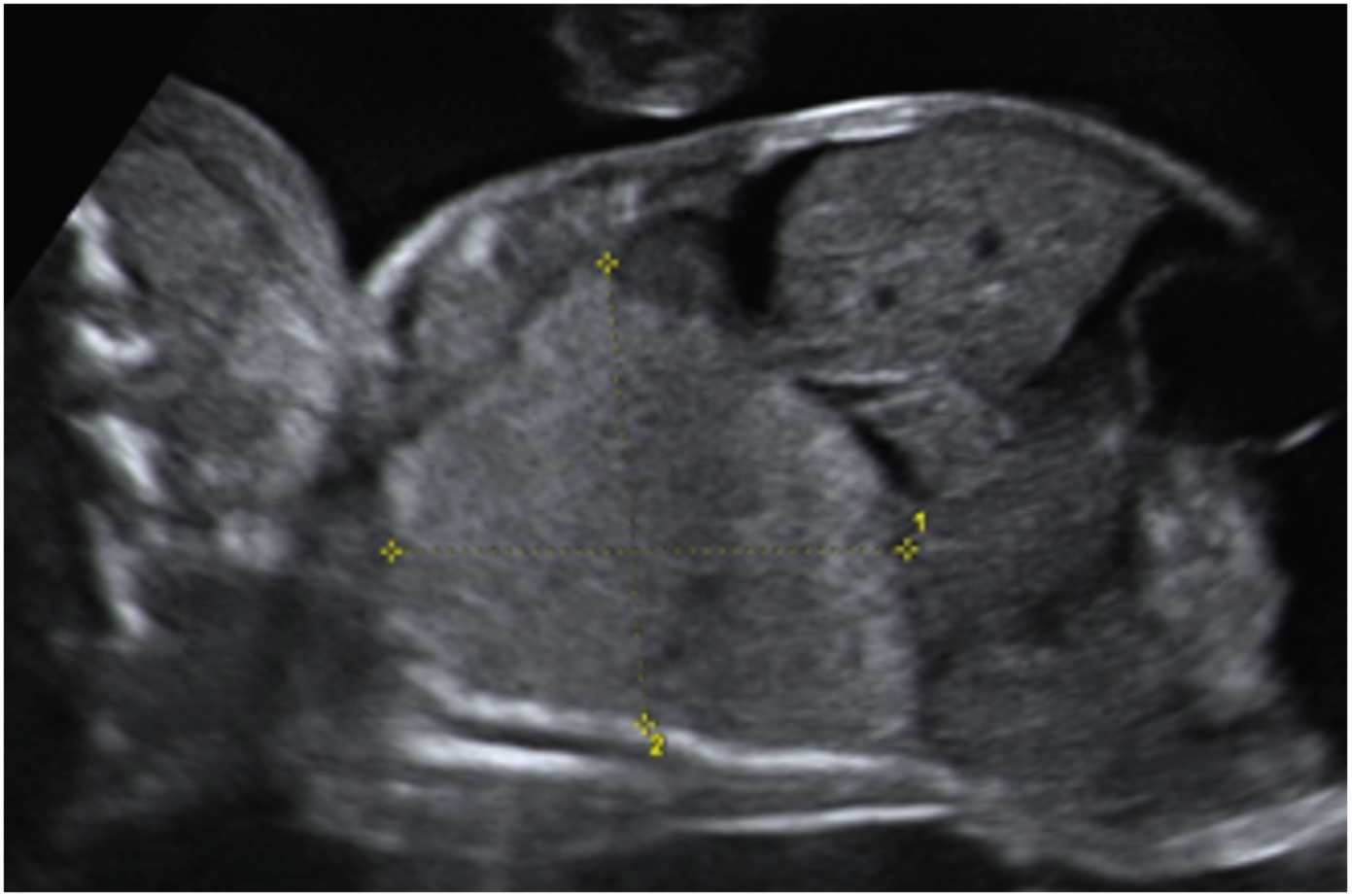
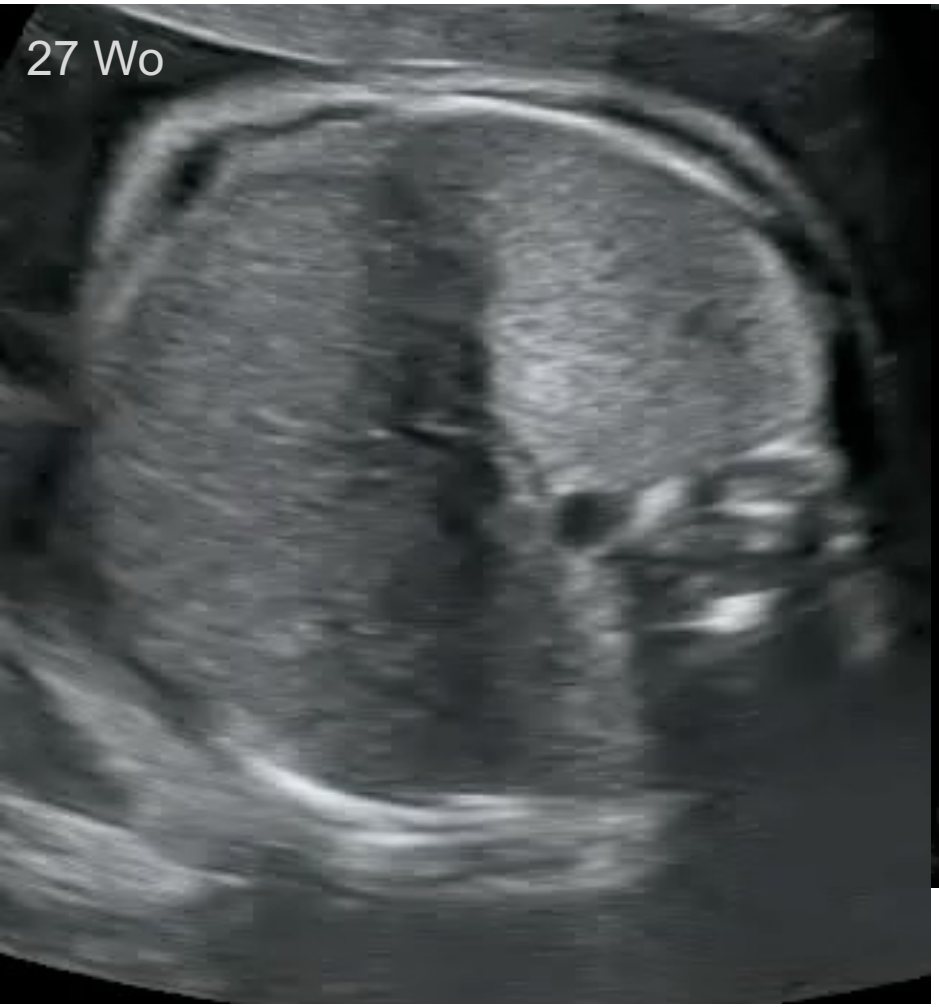
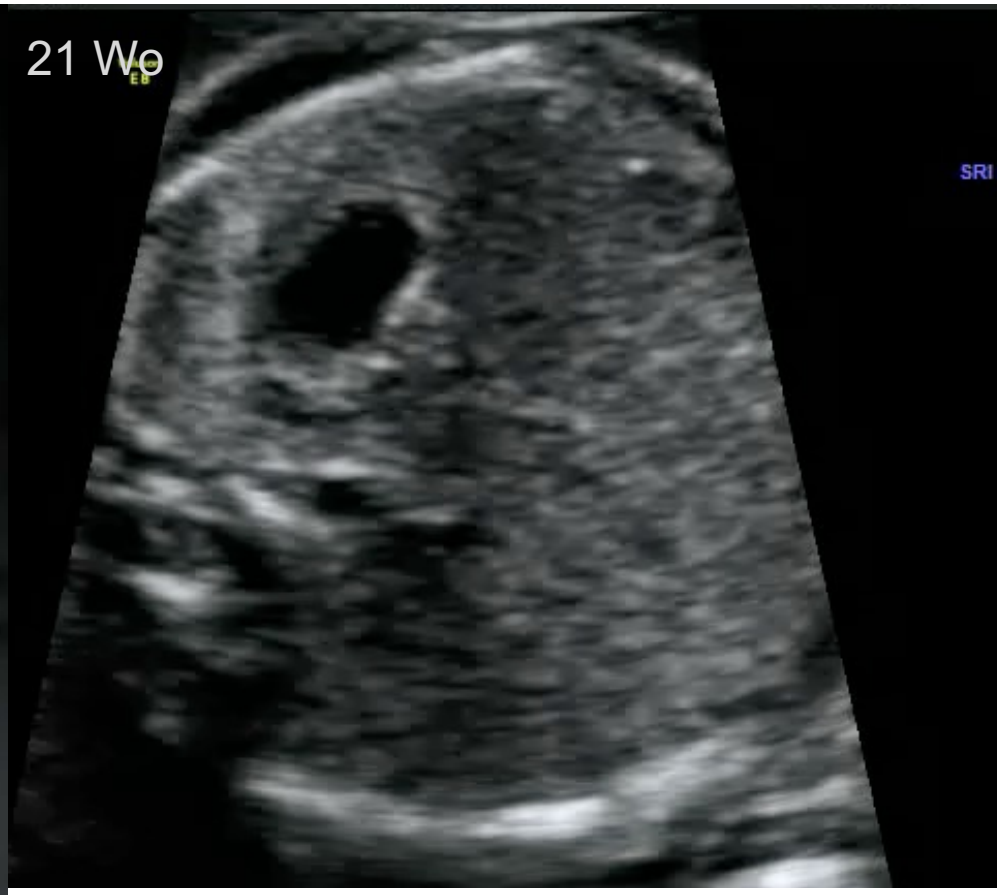





1
2

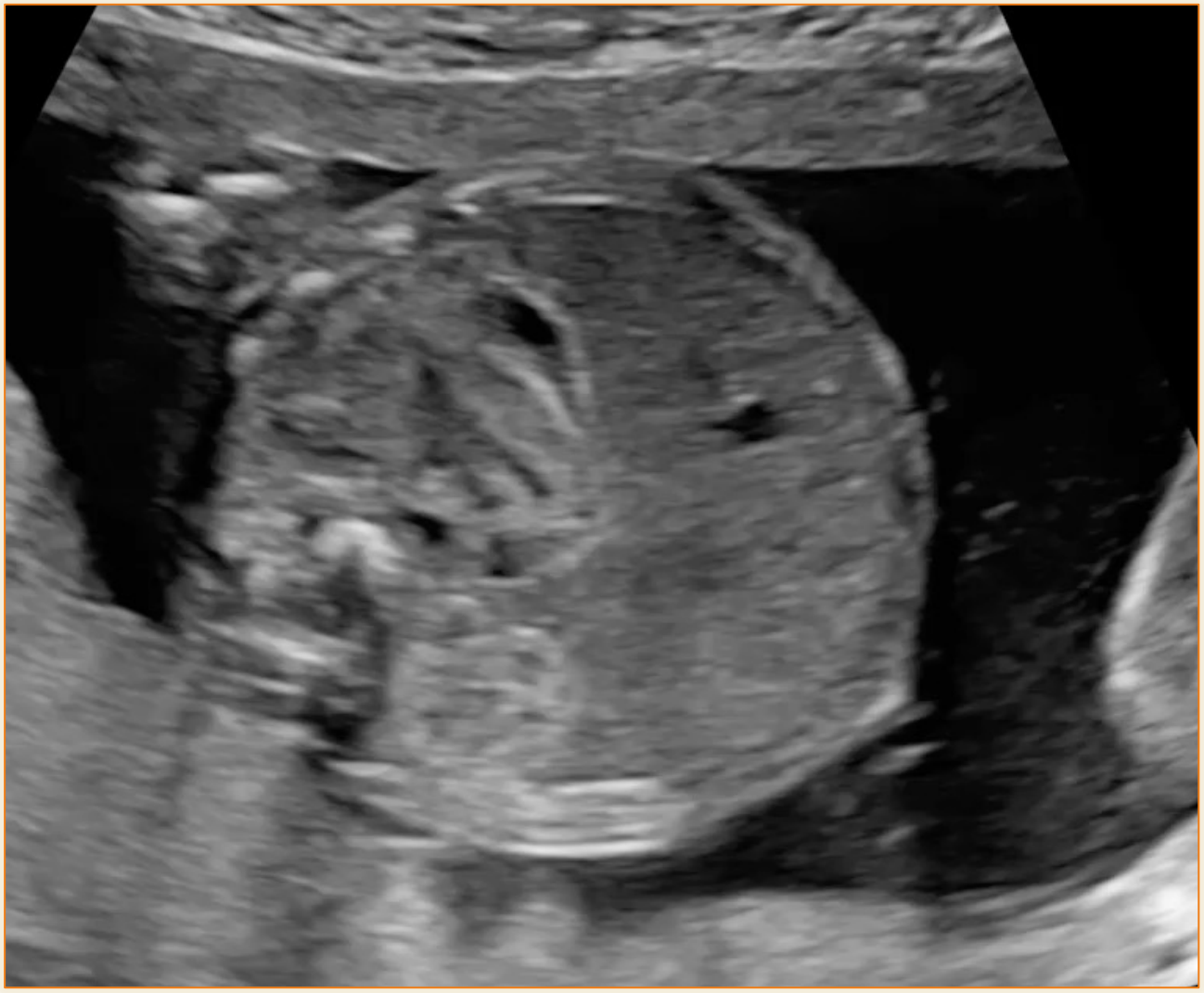

27 Wo


21 Wo
SRI

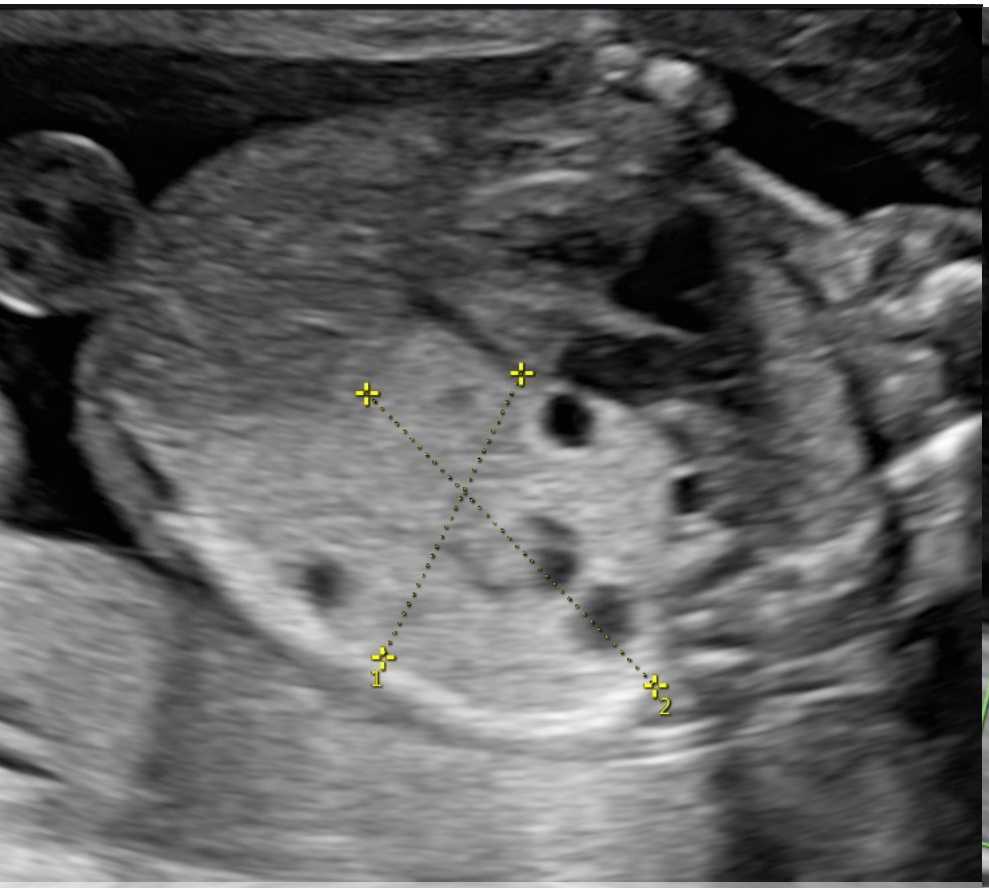
Fetale echogene Lungen

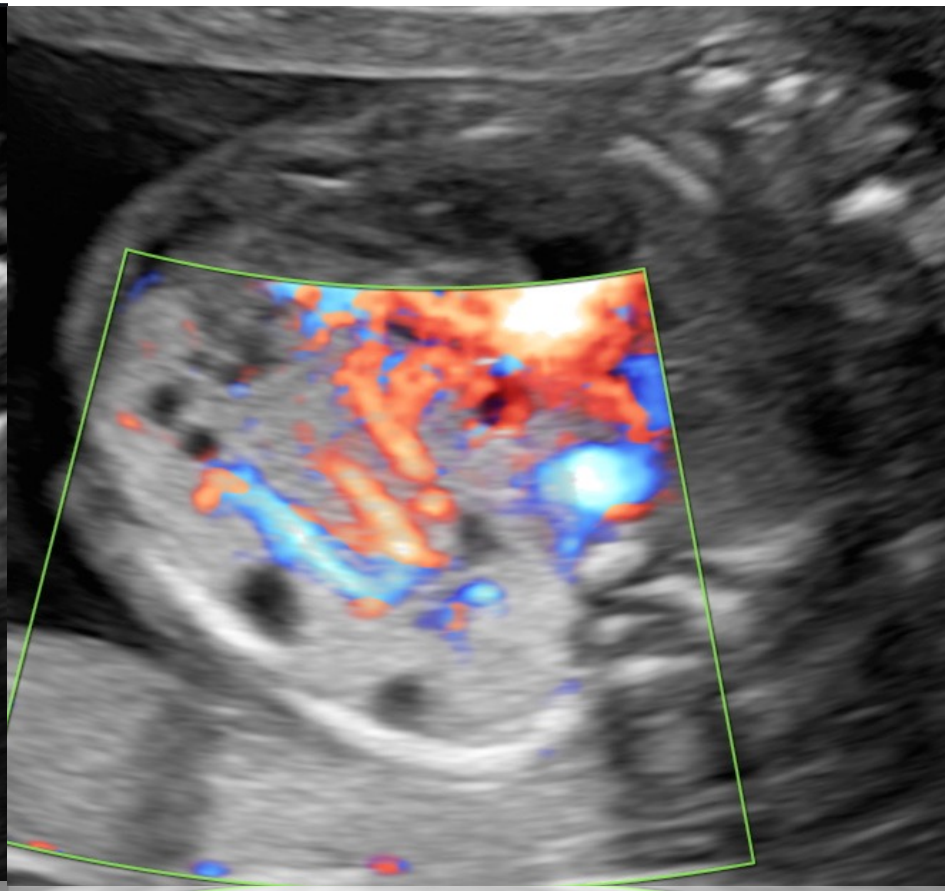
praena med
PRÄNATALDIAGNOSTIK

- Echogenität und Vergrößerung der Lungen resultiert durch die vermehrte “gefangene” Flüssigkeit
- Differentialdiagnosen:
  - Congenitale Cystische (Pulmonale) Adenomatoide Malformation (CCAM - CPAM)
  - extralobäre Lungensequestration (ELS)
  - Trachealatresie / Larynxatresie - CHAOS
  - Congenitales lobares Emphysem (CLE)
- Pathophysiologie und Histologie können überlappen (bronchiale Obstruktion/Atresie);
- Größe nimmt mit zunehmendem GA im 3. Trimenon oft ab



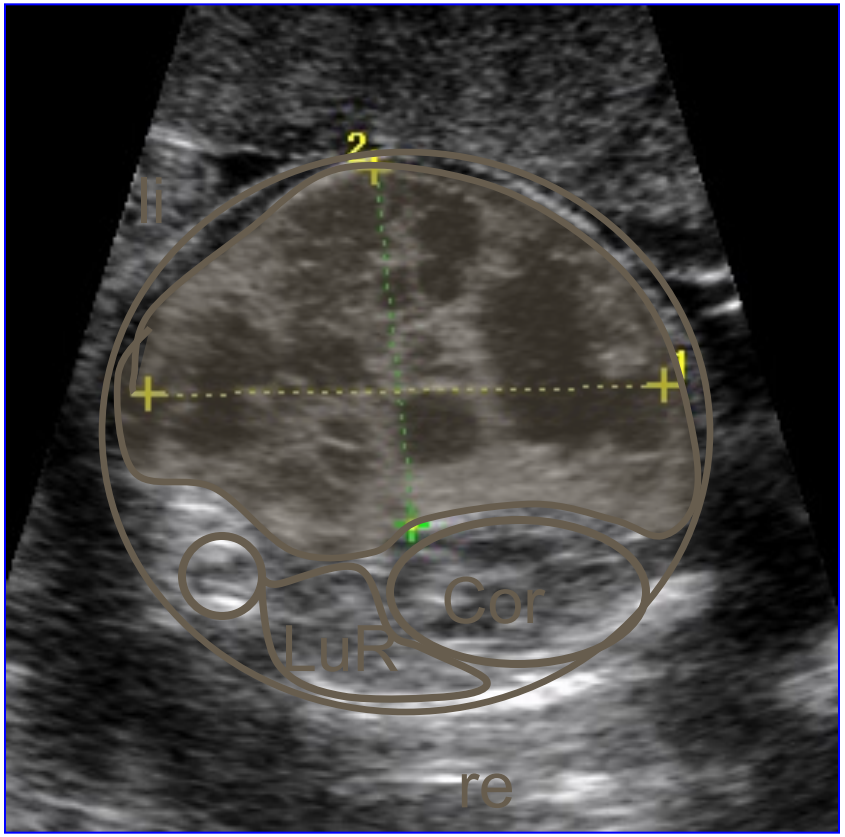
20 Wochen


1
2

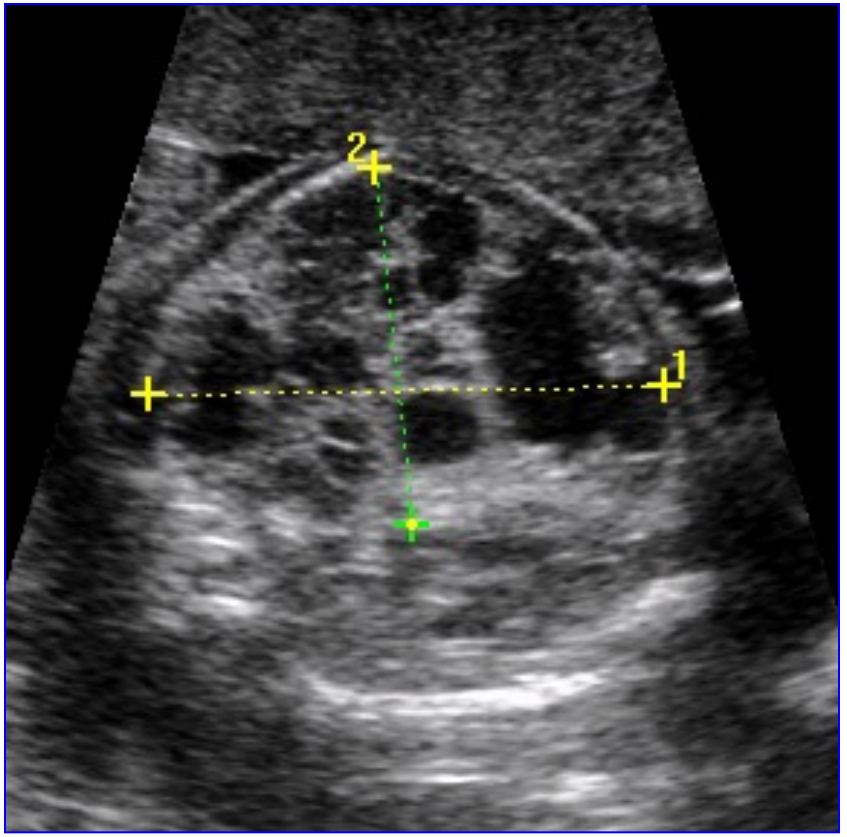


Zuweisung 22+1 SSW

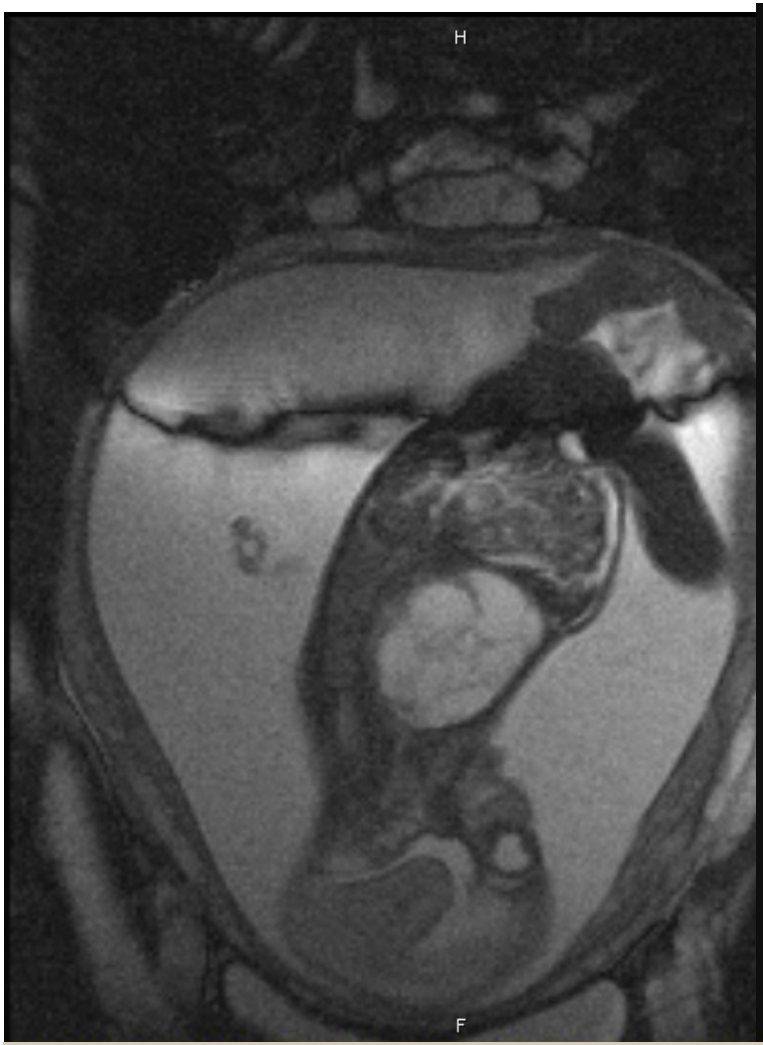
CCAM linke Lunge

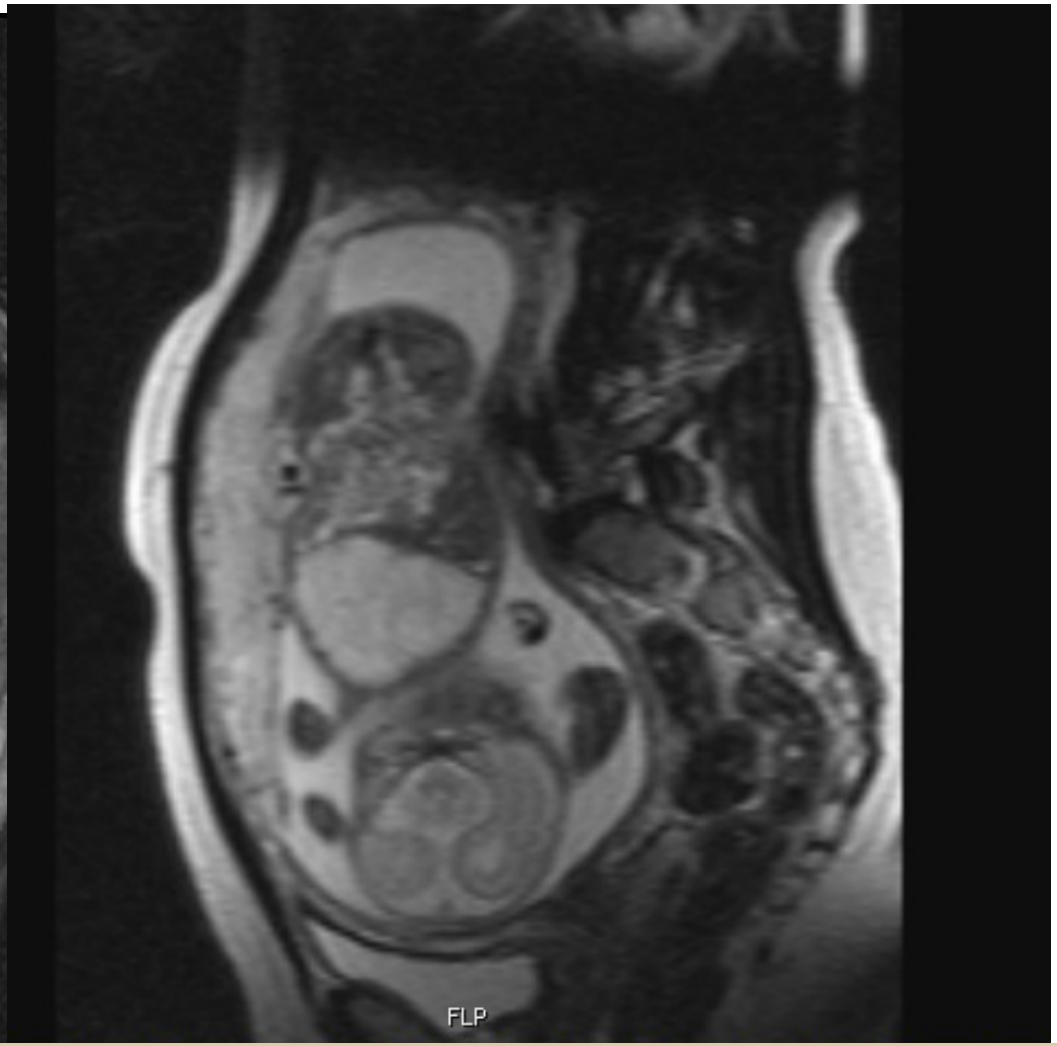


Steiner et al., UiM 2007



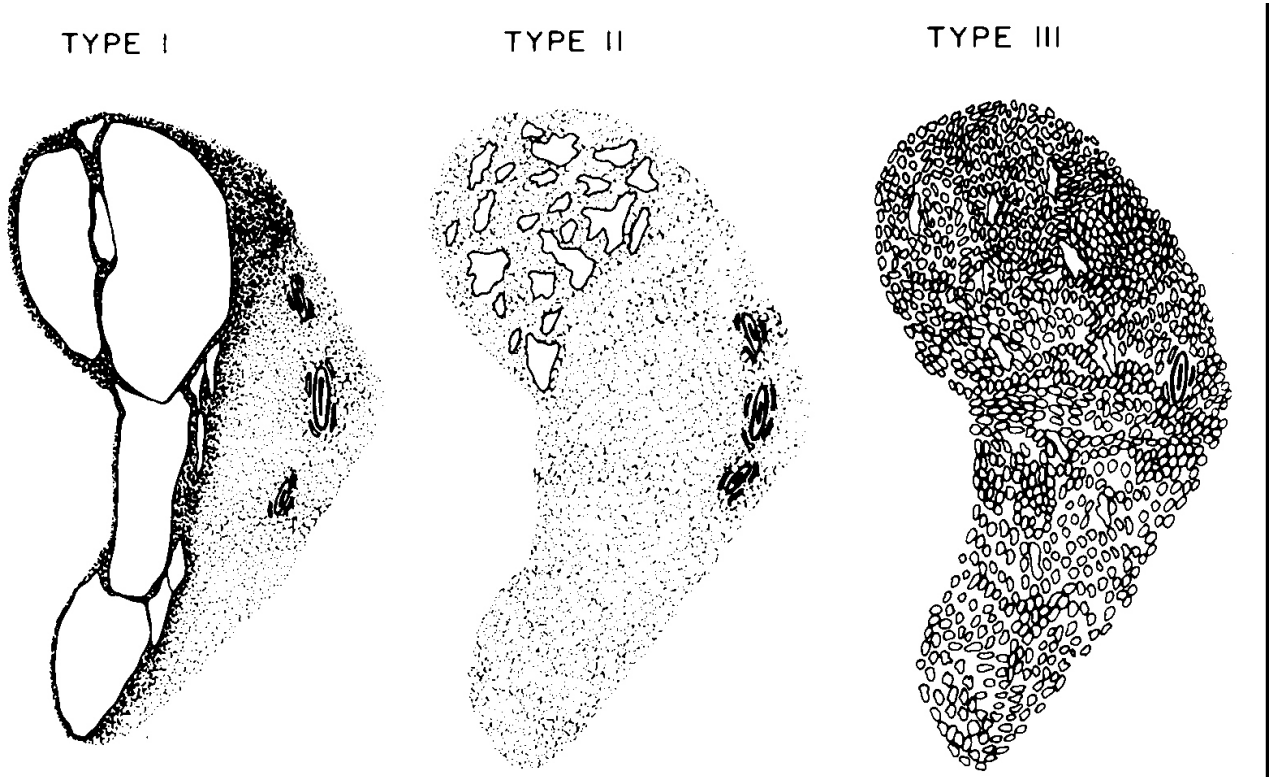
Pränatales MRI 23+4 SSW





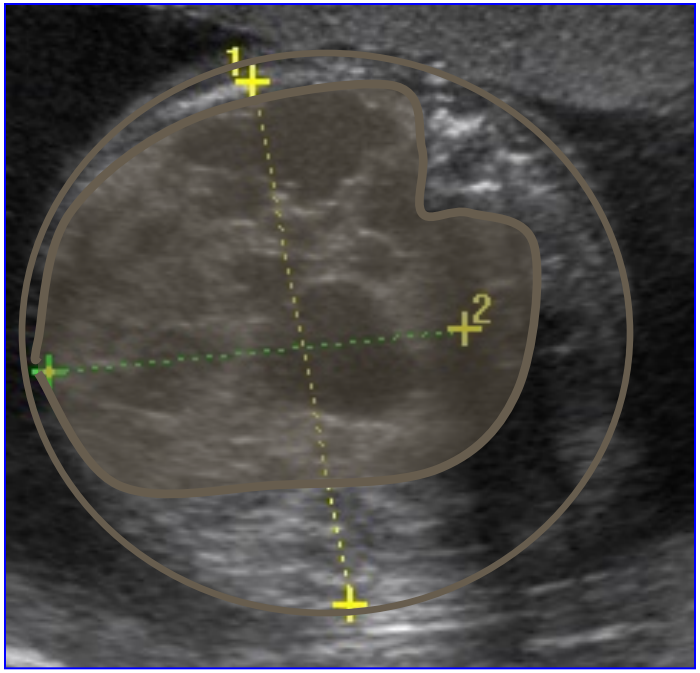
CCAM Klassifikation

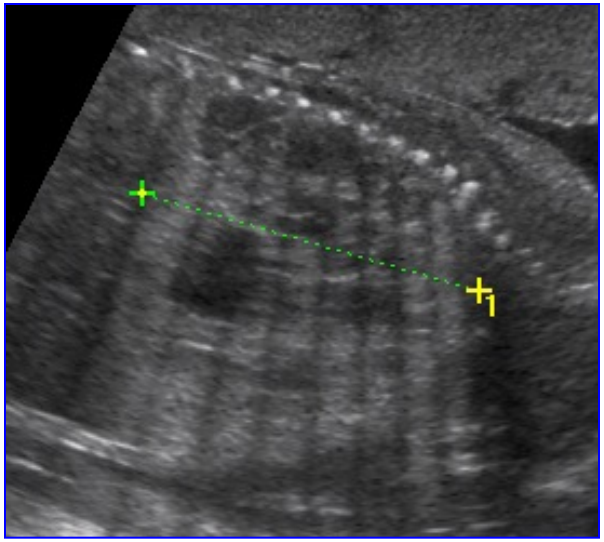
Stocker JT et al., Hum Pathol 1977



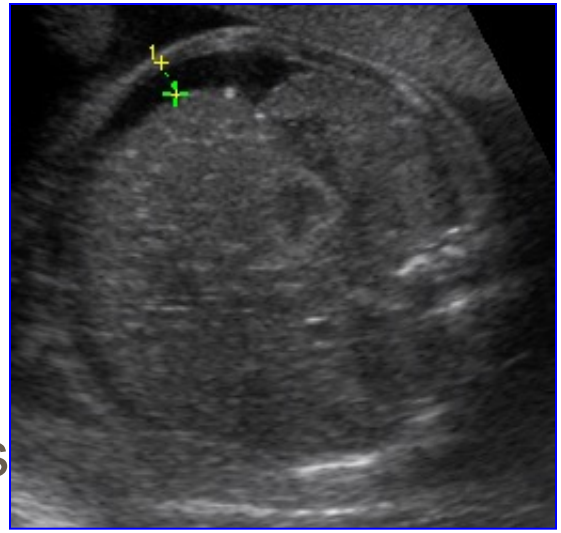
Histomorphologie	einzelne grosse Zysten	viele Zysten < 1,2cm	„nicht-zystische“ Läsion mit Mediastinalshift
Assoziierte Anomalien	selten	häufig	selten
Prognose	+/-	-	---

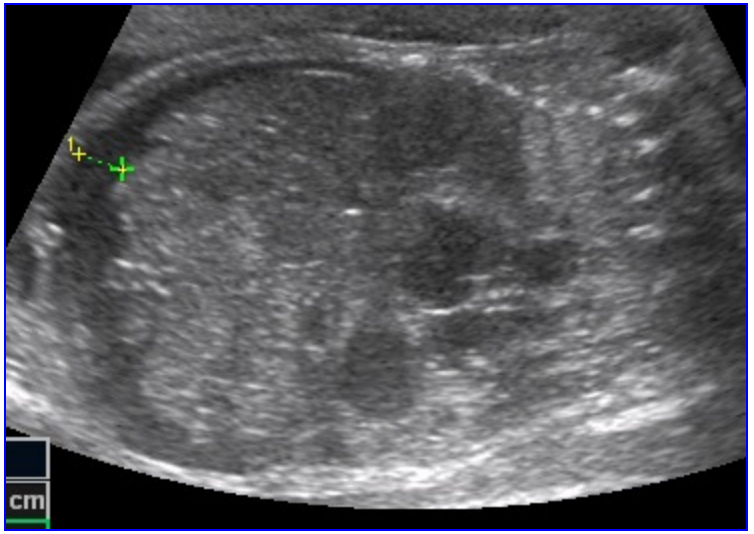
CCAM 24+5 SSW





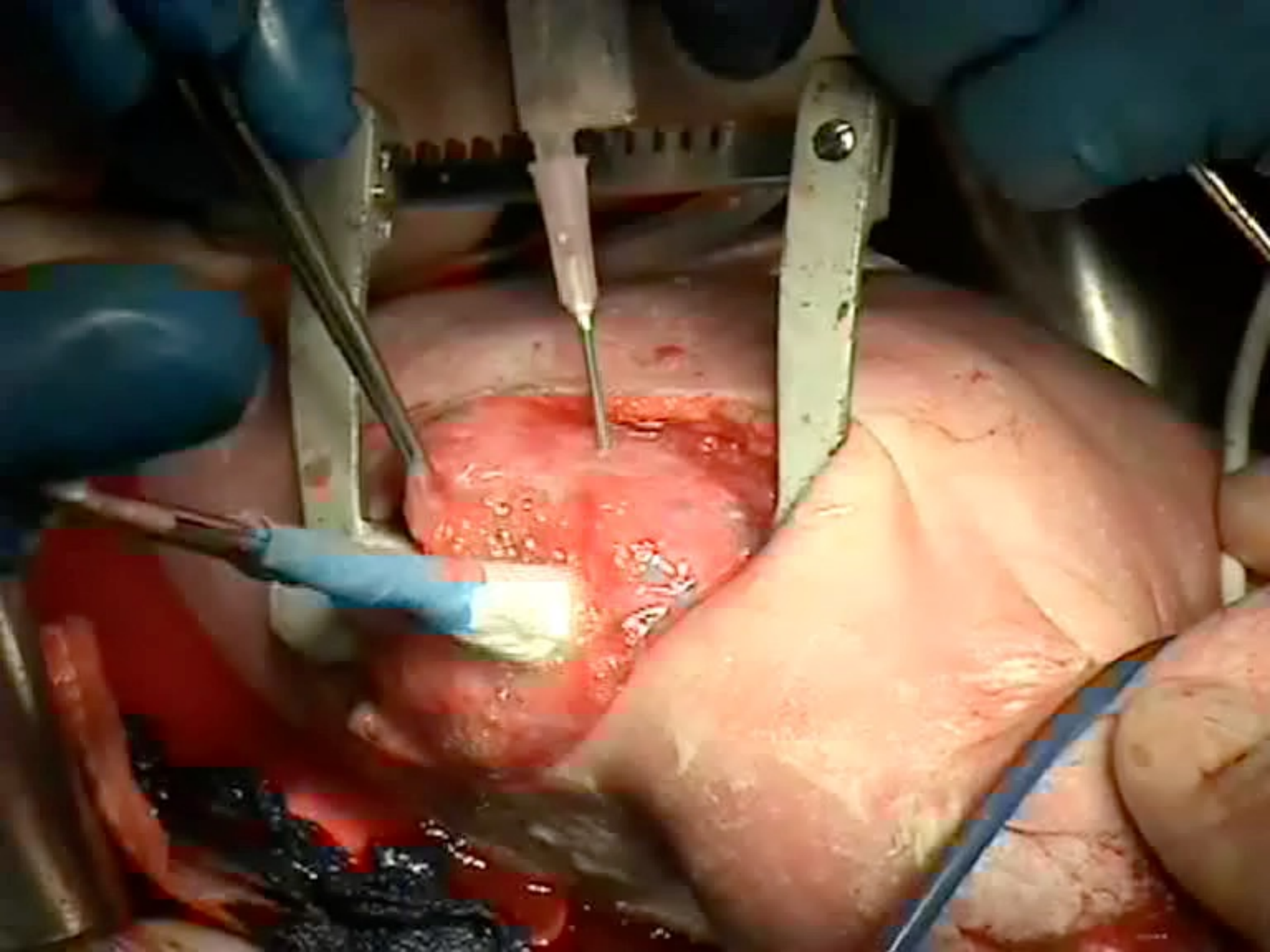
Aszites

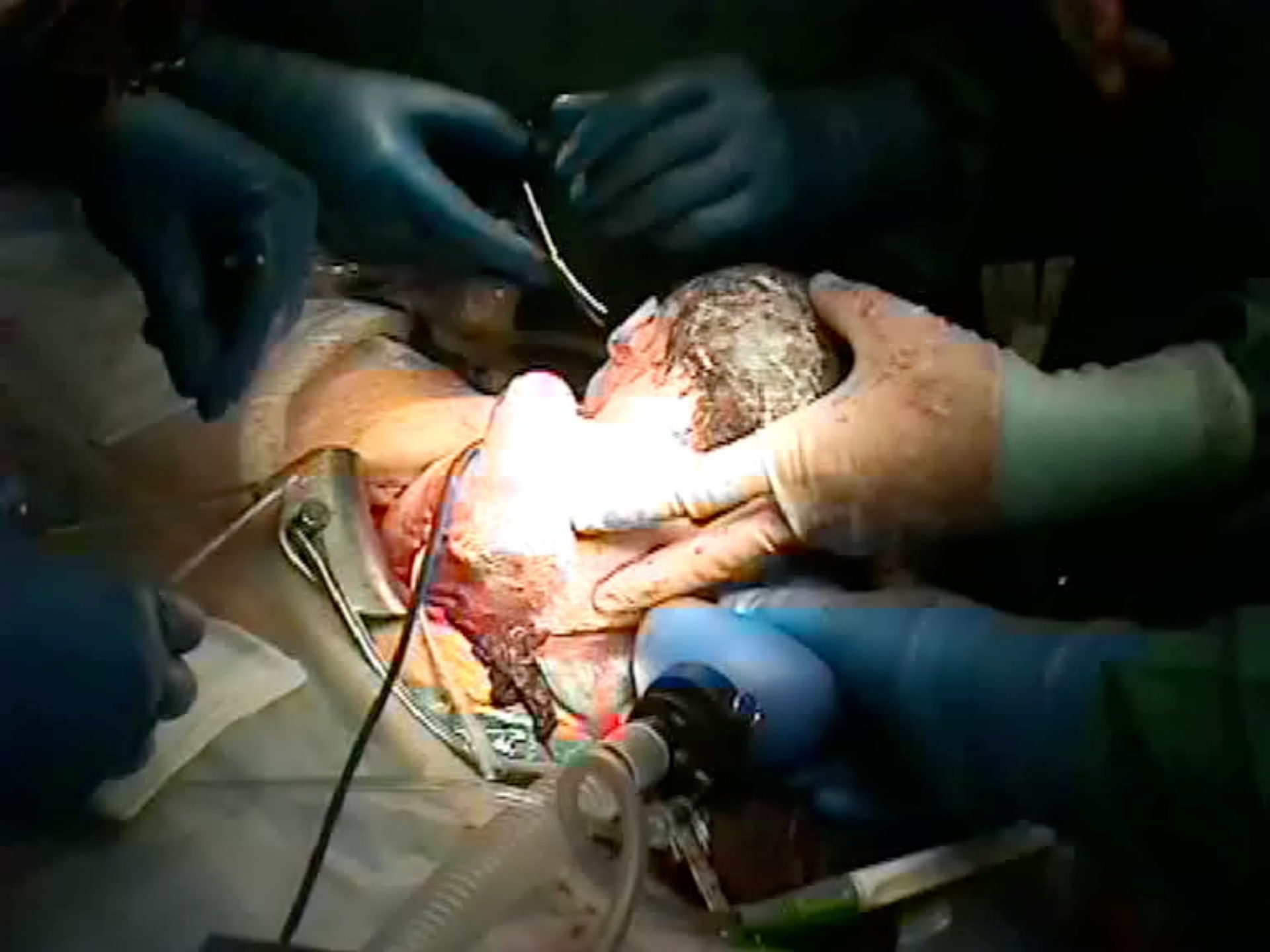


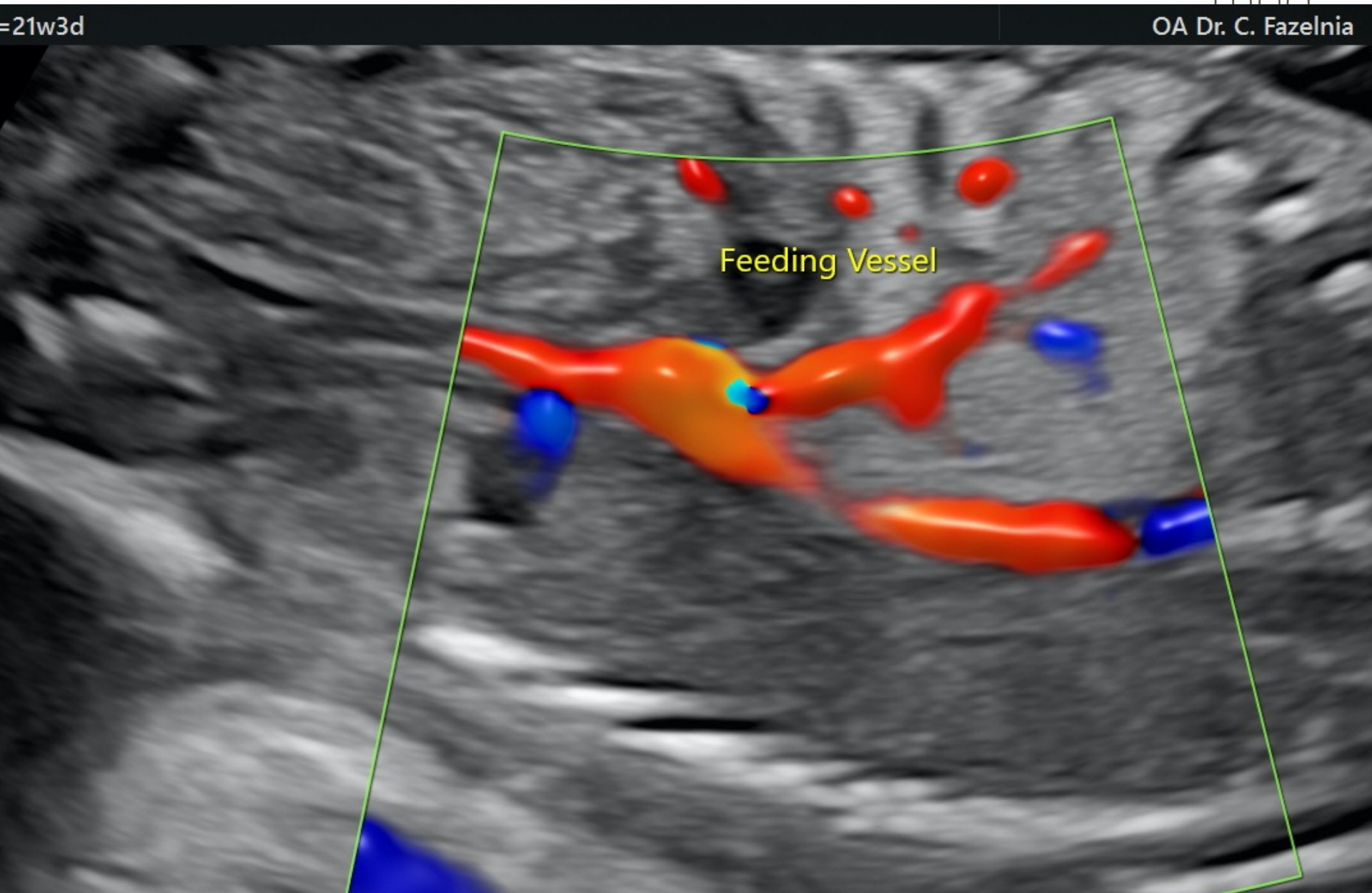


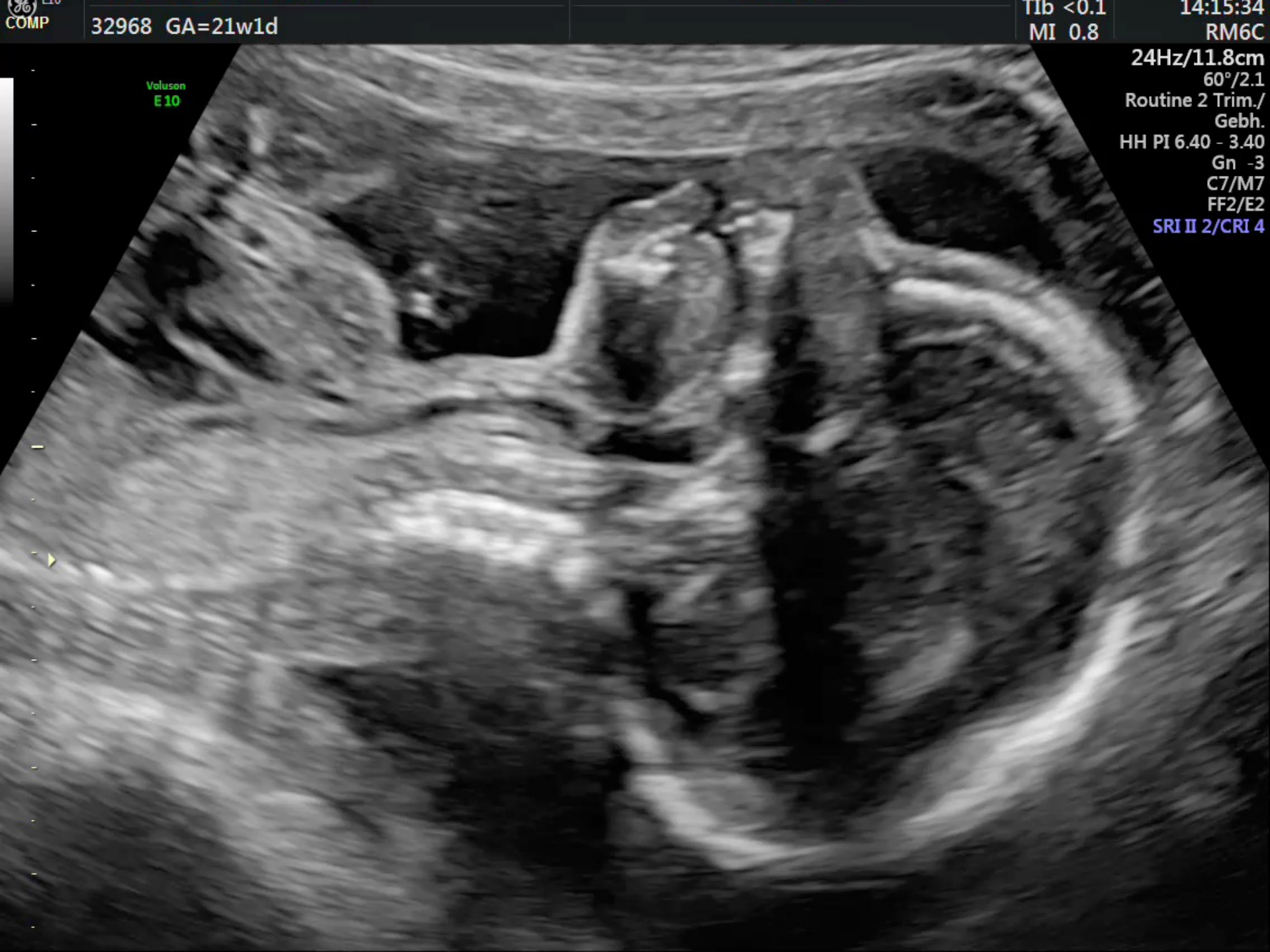
- Gewährleistung der Plazentaperfusion während fetaler OP
- Uterusrelaxation
- „tiefe“ Narkose – keine „Sektio-Narkose“
- Tokolyse Nitro, Atosiban
- Uterotomie mittels Staplertechnik nach intraoperativer sonographischer Kontrolle bei VW-Plazenta

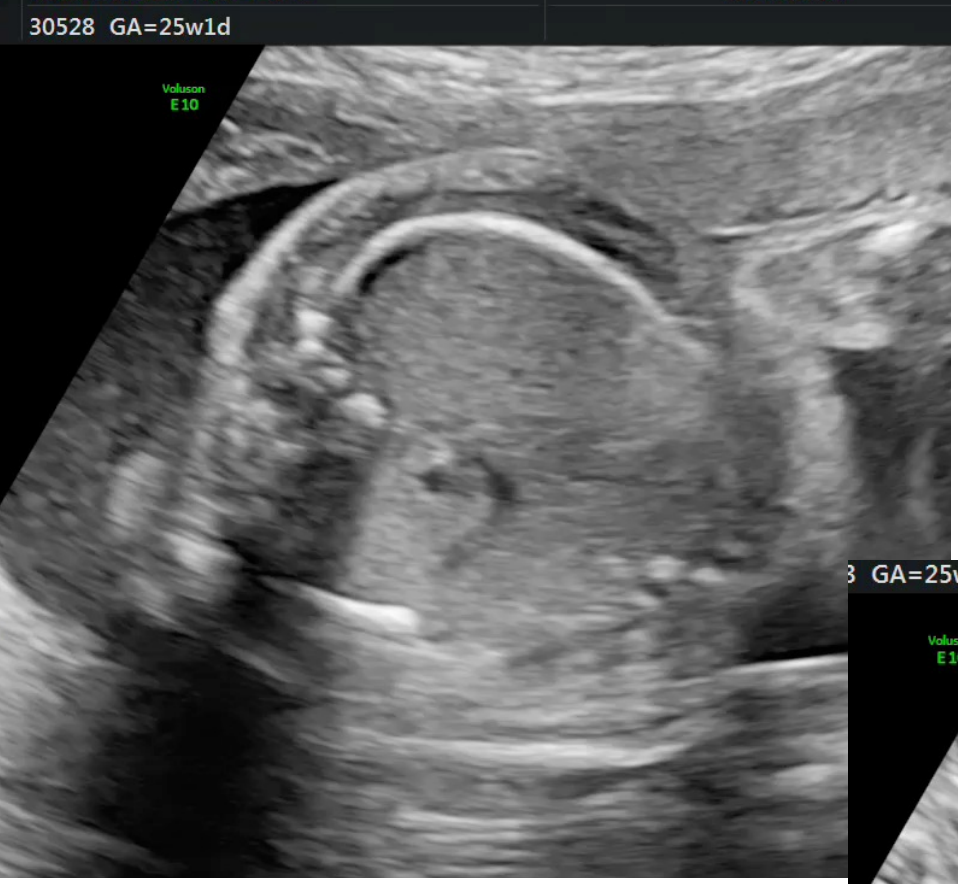
- Entscheidung zu EXIT
  - Keine Spontanatmung
  - Enorm hoher Beatmungsdruck
  - Pulsoximetrie nach O2-Gabe kein Anstieg der Sättigung, bleibt bei Werten um 50-60



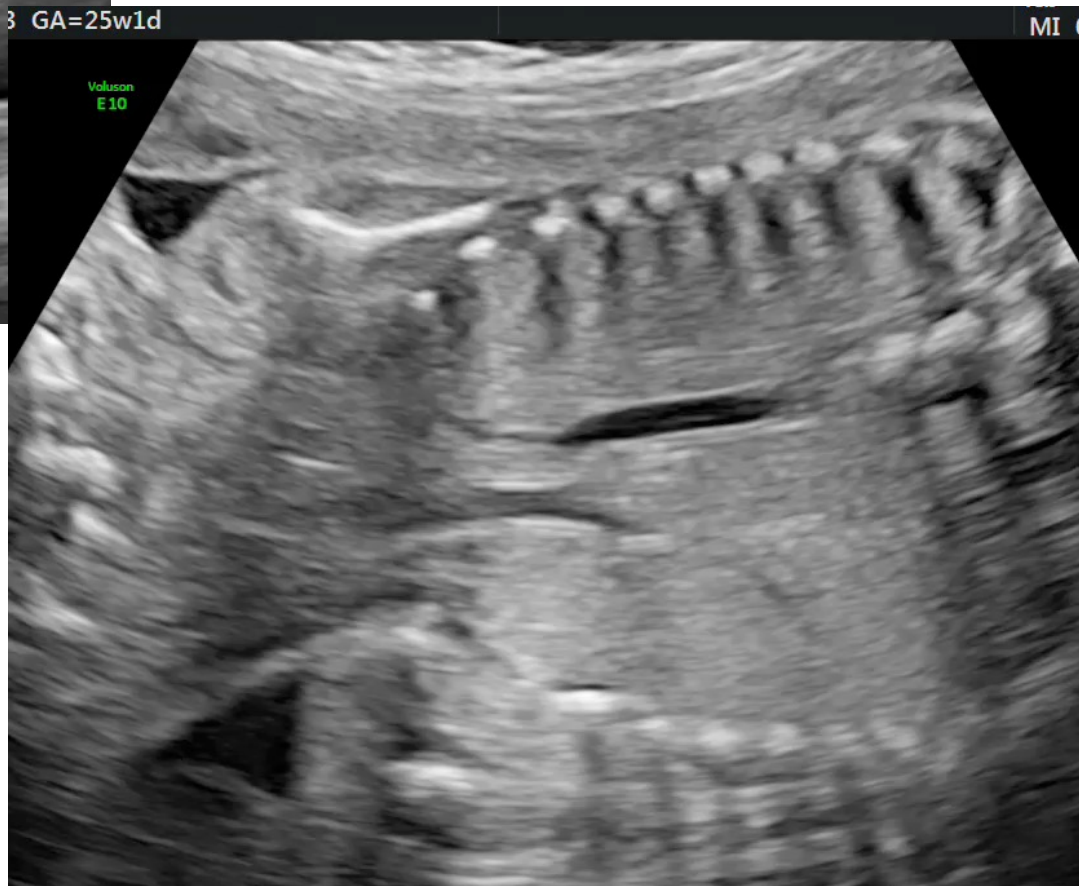


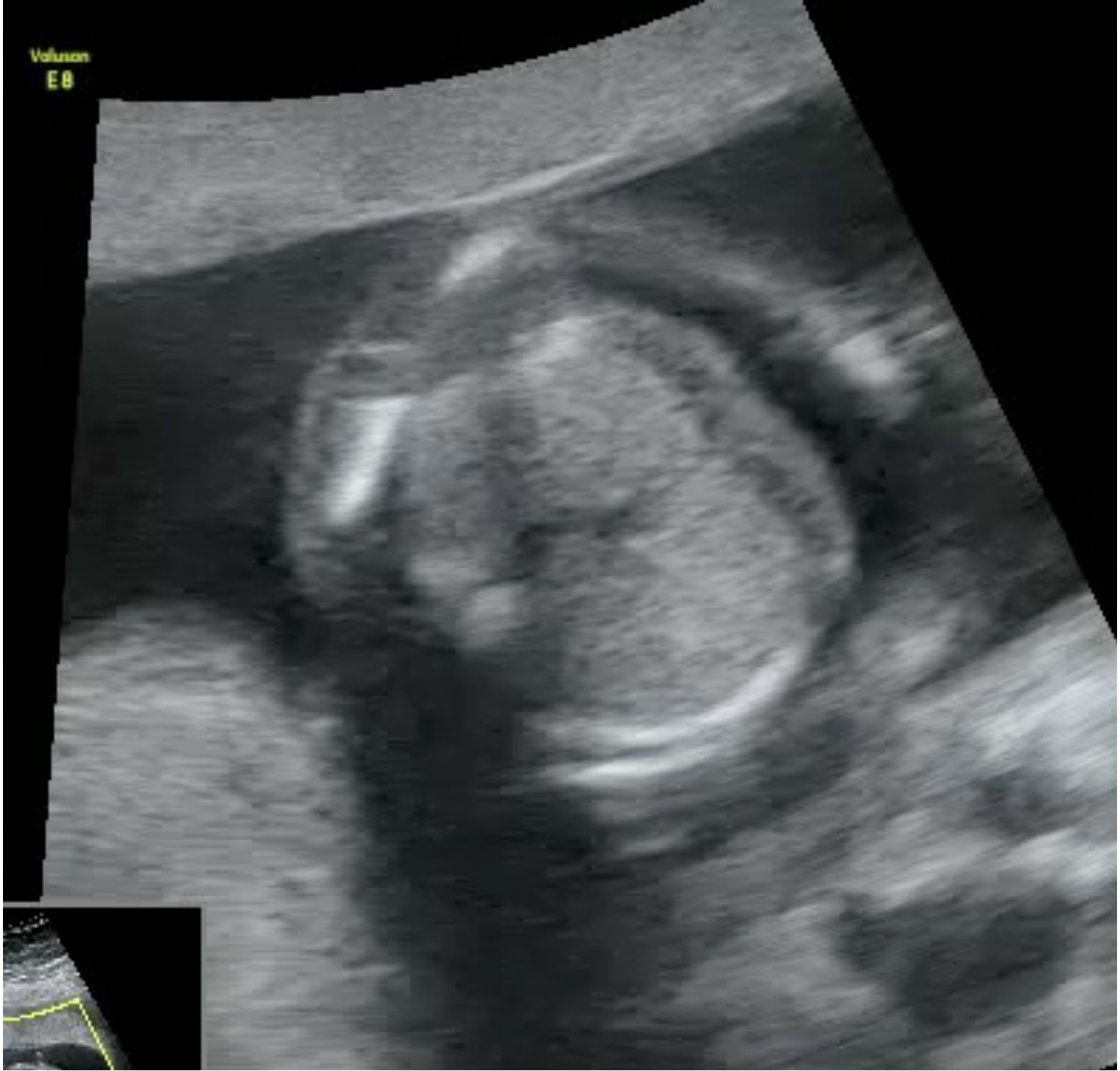

=21w3d
OA Dr. C. Fazelnia
Feeding Vessel

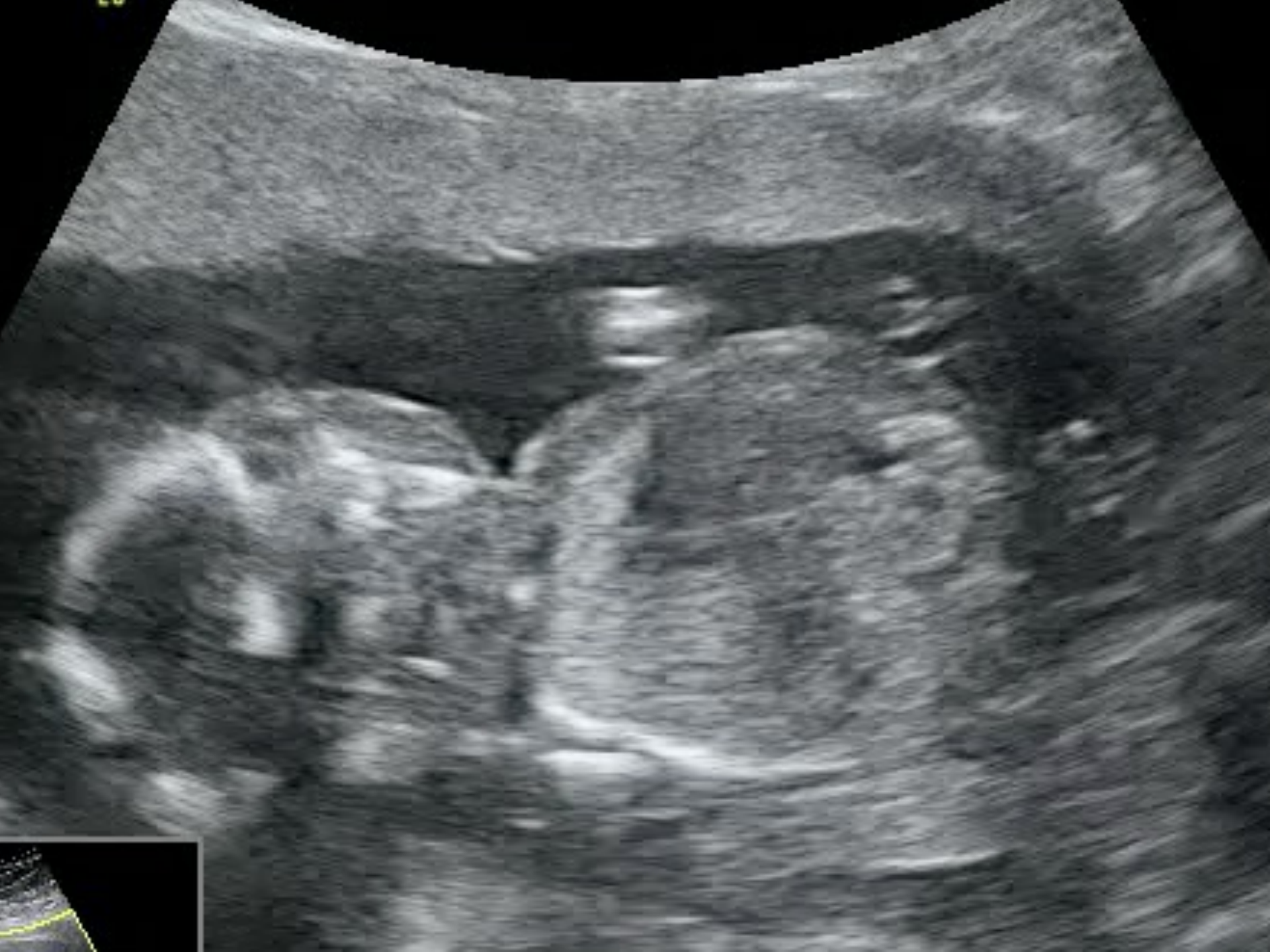

COMP
32968 GA=21w1d
TIb <0.1
MI 0.8
14:15:34
RM6C
Voluson
E 10
24Hz/11.8cm
60°/2.1
Routine 2 Trim./
Gebh.
HH PI 6.40 - 3.40
Gn -3
C7/M7
FF2/E2
SRI II 2/CRI 4

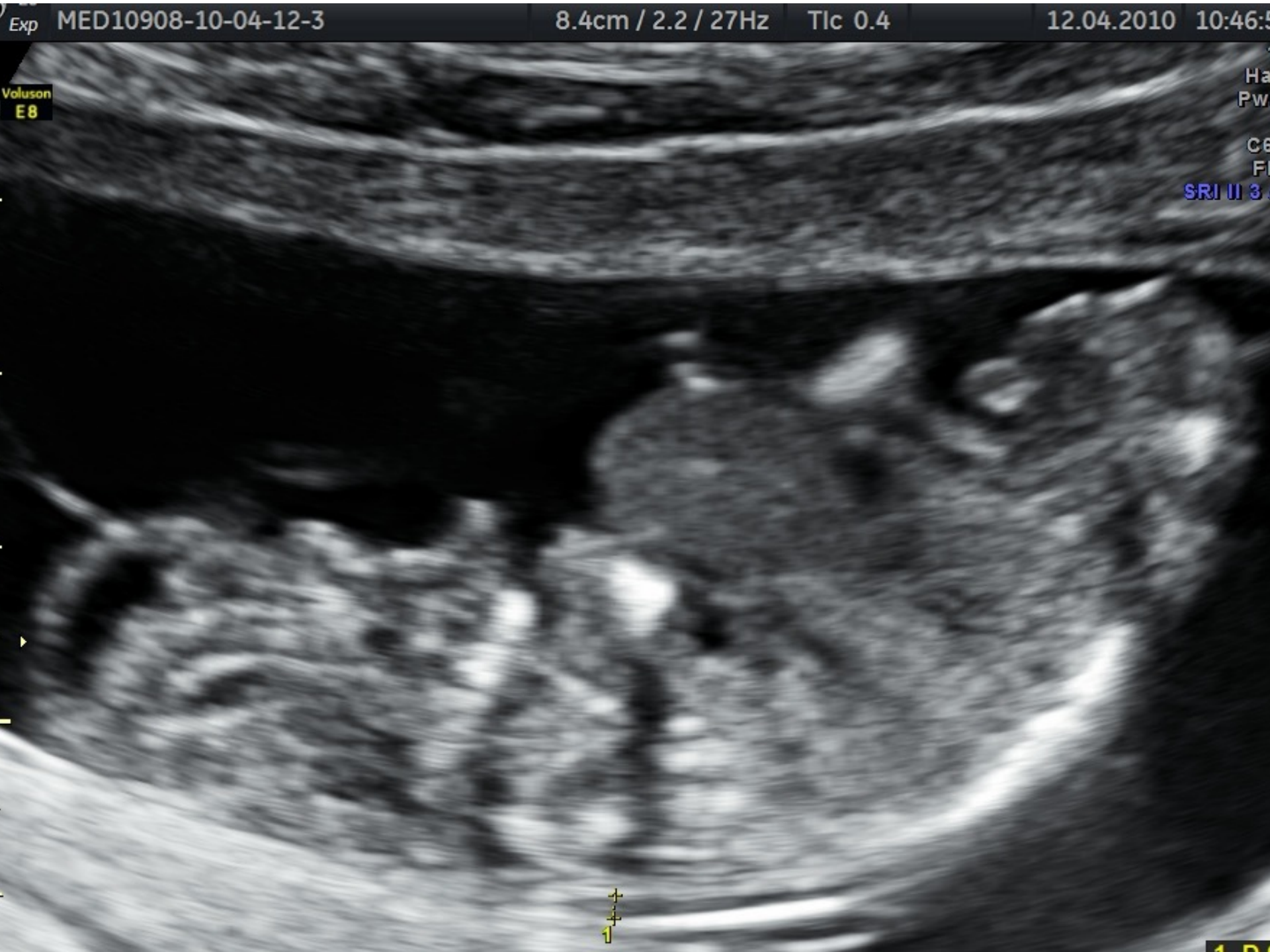


Trachea




Voluson
E8



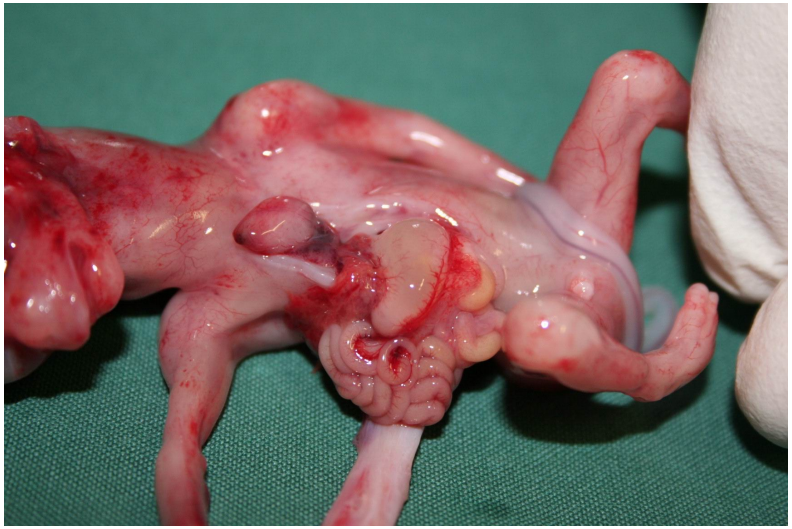

Exp
MED10908-10-04-12-3
8.4cm / 2.2 / 27Hz
TIc 0.4
12.04.2010
Voluson
E8
SRI II 3

Prevalence of body fold defects

Defect	Prevalence
Omphalocele (simple)	1/5500 births
Pentalogy of Cantrell	1/100,000 births
Cloacal exstrophy	1/200,000 births
Body stalk defect	3/700,000 births

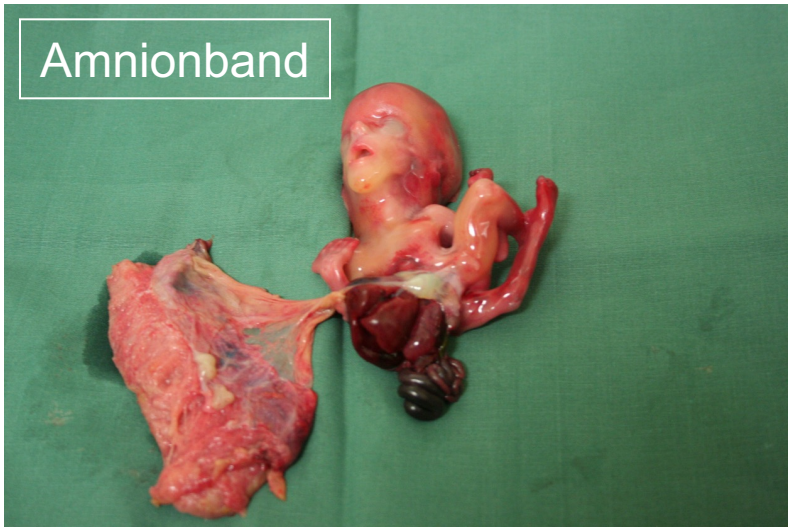
Cantrell-Pentalogie

Epigastrischer Bauchwanddefekt
Defekt des unteren Sternums
Defekt des diaphragmatischen Perikards
Defekt des vorderen Diaphragmas
Intrakardiale Anomalie



Body Stalk Anomalie

Große komplexe craniale Defekte (Encephalocelen)
Gesichtsspalten
Große thorakale und Bauchwanddefekte
Extremitätendefekte



Original article

Diagnostic accuracy of imaging studies in congenital lung malformations

Rodrigo A Mon [1, 2], Kevin N Johnson [1], Maria Ladino-Torres [2, 3], Amer Heider [4], George B Mychaliska [1, 2], Marjorie C Treadwell [2, 5], Shaun M Kunisaki [1, 2]

Arch Dis Child Fetal Neonatal Ed.
2019 Jul;104(4):F372-7

- US-Detektionsrate
  - der Läsionen insgesamt: 82 %
  - Korrekte Diagnose CPAM 75%
- MR-Detektionsrate vergleichbar
  - etwas besser in der Diagnose der systemischen Gefäßversorgung

Journal of Pediatric Surgery
Volume 55, Issue 5, May 2020, Pages 844-847

Neonatal Condition

Accuracy of prenatal and postnatal imaging for management of congenital lung malformations

Candace C. Style MD [a, b], Amy R. Mehollin-Ray [a, c], Mariatu A Verla MD [a, b], Oluyinka O. Olutoye MD, PhD [a, b, d], Patricio E. Lau MD [b], Brittany L. Johnson MD [a, b], Alice King MD [a, b], Sundeep G. Keswani MD [a, b], Timothy C. Lee MD [a, b]

- Evtl. postnataler CT wichtig für Operationsindikationsstellung

Fetale Thoraxprobleme Zusammenfassung

praena PRÄNATALDIAGNOSTIK med

- Den Thorax bewusst untersuchen !!
- CDH = schwere Fehlbildung mit hoher Mortalität, besonders wenn nicht pränatal diagnostiziert. Ultraschall hilft bei prognostischer Einschätzung

  Pränatale Chirurgie = anzubieten
- Echogene und zystische Lungen haben großteils gute Prognose
- Hydrothorax kann unbekannte Atiologie haben, Outcome ist dann unsicher zu prognostizieren, Behandlung /Shunting bei Persistenz
- Thoraxfehlbildungen im Rahmen von Syndromen und Sequenzen
- 1.Trimesterdiagnostik !