

CPAM and other lung lesions of fetus

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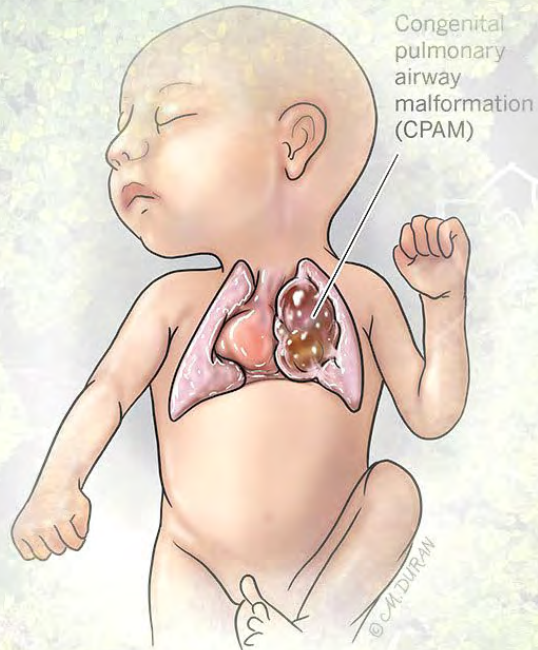


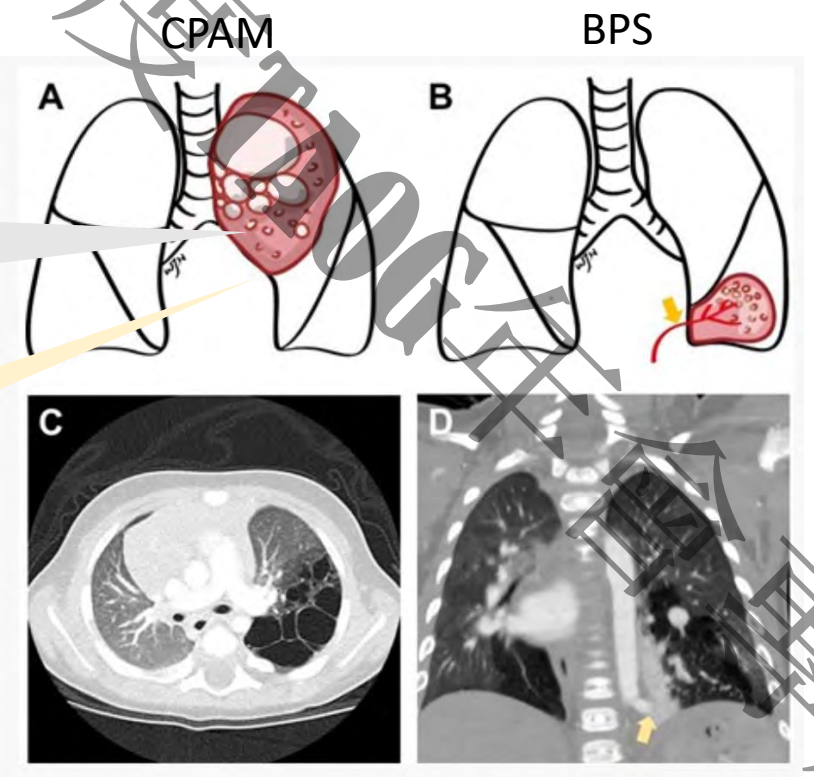
Photo by Michael Olsen on Unsplash and Myriams-Fotos on Pixabay
https://partnersincare.health/conditions/congenital-pulmonary-airway-malformation

CPAM

congenital pulmonary airway malformation

Also known as congenital cystic adenomatoid malformation (CCAM) previously

the **most common (ca. 59-95%)** type of fetal lung lesion.



Mostly asymptomatic, although looks terrible on prenatal ultrasound.

Large lesions can cause serious complications, including heart failure (***fetal hydrops**) or maternal mirror syndrome.

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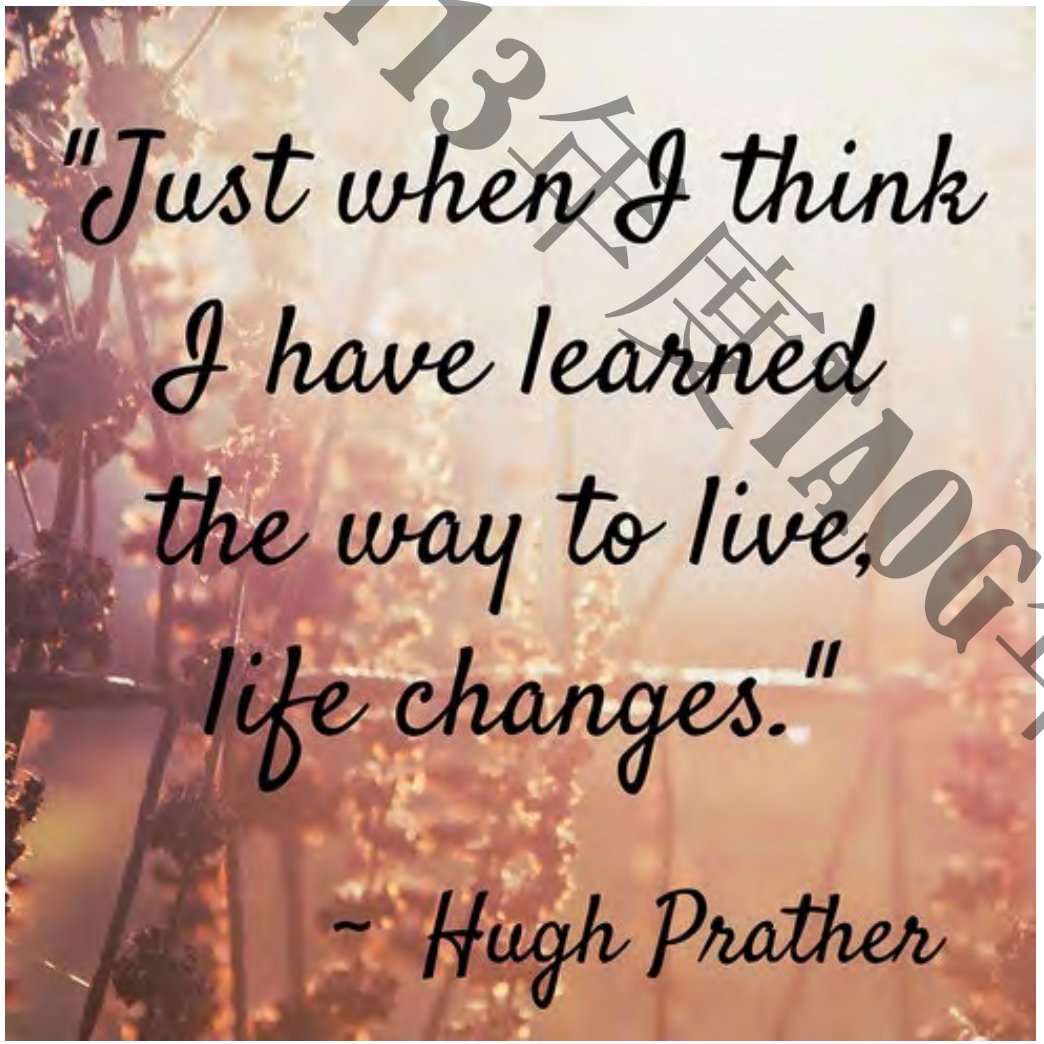

Just when I think

I have leaned

every facts about

CPAM,

they changes.

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Incidence

Rare -> not so rare

1/35,000 (earlier report)

To

1/2,500 (current data)

Type

Diverse

two

(macro- and microcystic)

three

(Stocker's classification,
Type I-III)

or Five

(pathological classifications
Type 0-4)

Outcome

Poor -> excellent
(survival > 95%,
98% for non-hydrops)

Microcystic type

was thought be the worst, but

now was considered **the best.**

Macrocystic – vice versa

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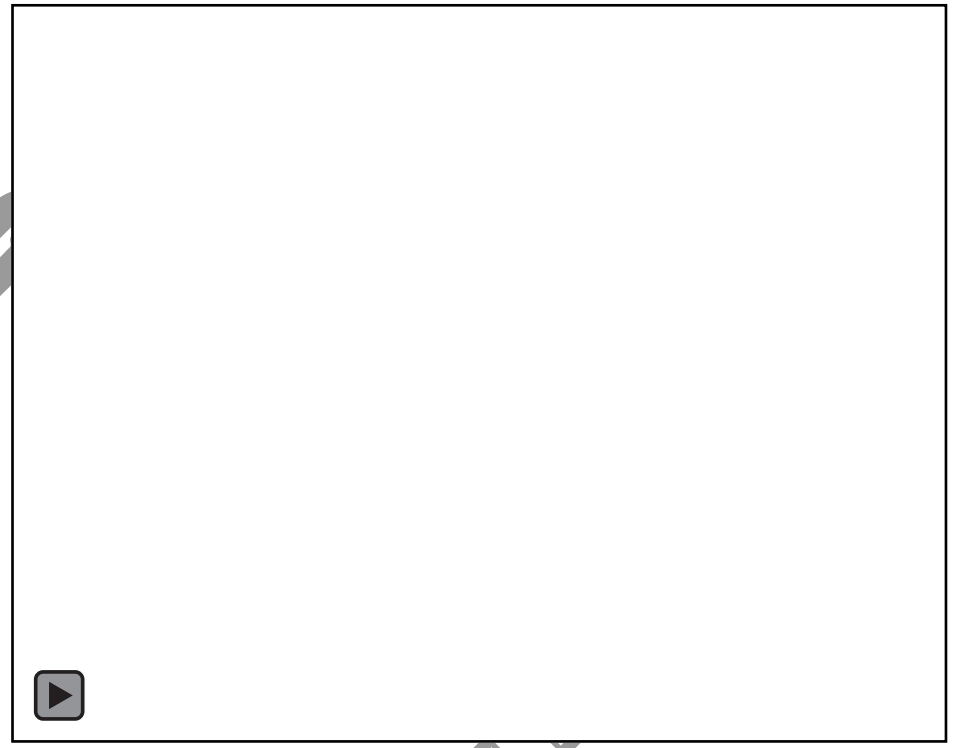
Ultrasound

- predominantly cystic (single or multiple variable-size cysts)
- hyperechoic (solid); with or without scattered small cysts



23w, CVR 2.34 (pleural effusion +)



22w, CVR 0.75, pleural effusion (+)

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Left lur
with m

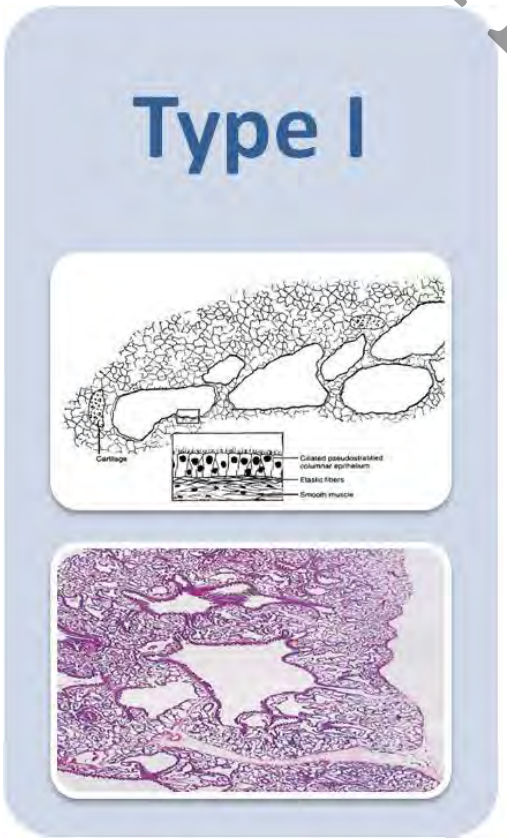
CVR 1.3
CVR 1.0
*Not ap
Deliver



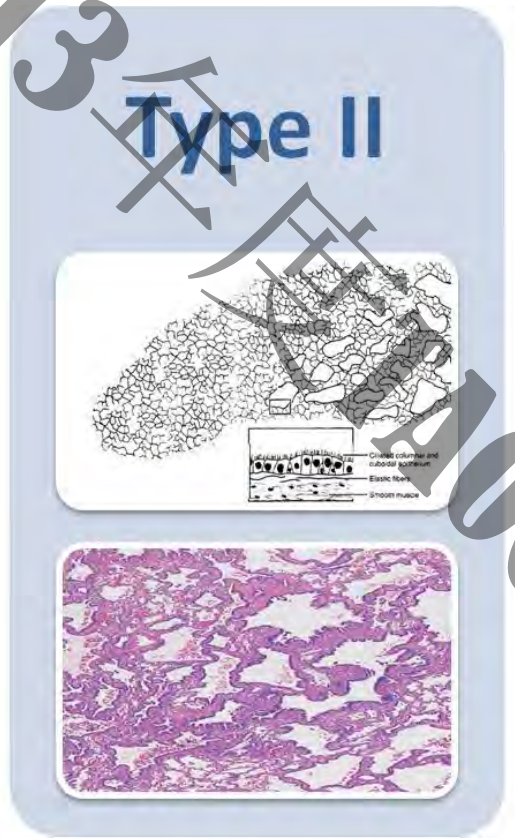


Stocker's classification

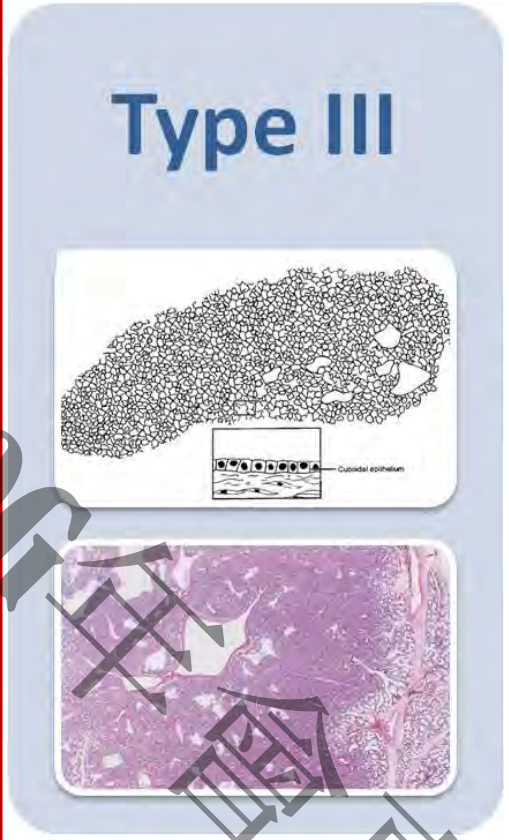
Cystic ⟷ solid



Ciliated psudostratified columnar epi.



Ciliated columnar and cuboidal epi.



Cuboidal epi.

TABLE 2.

CPAM Prenatal Screening Classification

Type	Description
1	Macrocystic: cysts >5 mm in size
2	Microcystic: cysts <5 mm in size (appear solid on ultrasound)

Abbreviation: CPAM, congenital pulmonary adenomatoid malformation.
Adapted from Sockrider[12] and Adzick et al.[14]

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CPAM Type	Type 0	Type 1	Type 2	Type 3	Type 4
Stoker classication		Type I	Type II	Type III	
Developmental origin	tracheal/ bronchial	bronchial/ bronchiolar	bronchiolar	bronchiolar/ alveolar	acinar
Proportion of CPAM	2%	60-65%	15-25%	5-10%	10%
Cyst size	none	2-10 cm	0.5-2 cm	microcystic/solid	large multilocular
Timing of presentation	birth	prenatal	postnatal	prenatal	postnatal
Clinical presentation	lethal pulmonary hypoplasia	asymptomatic, respiratory distress or infection	asymptomatic, respiratory distress or infection	prenatal (hydrops), postnatal respiratory distress	incidental finding

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malignancy change

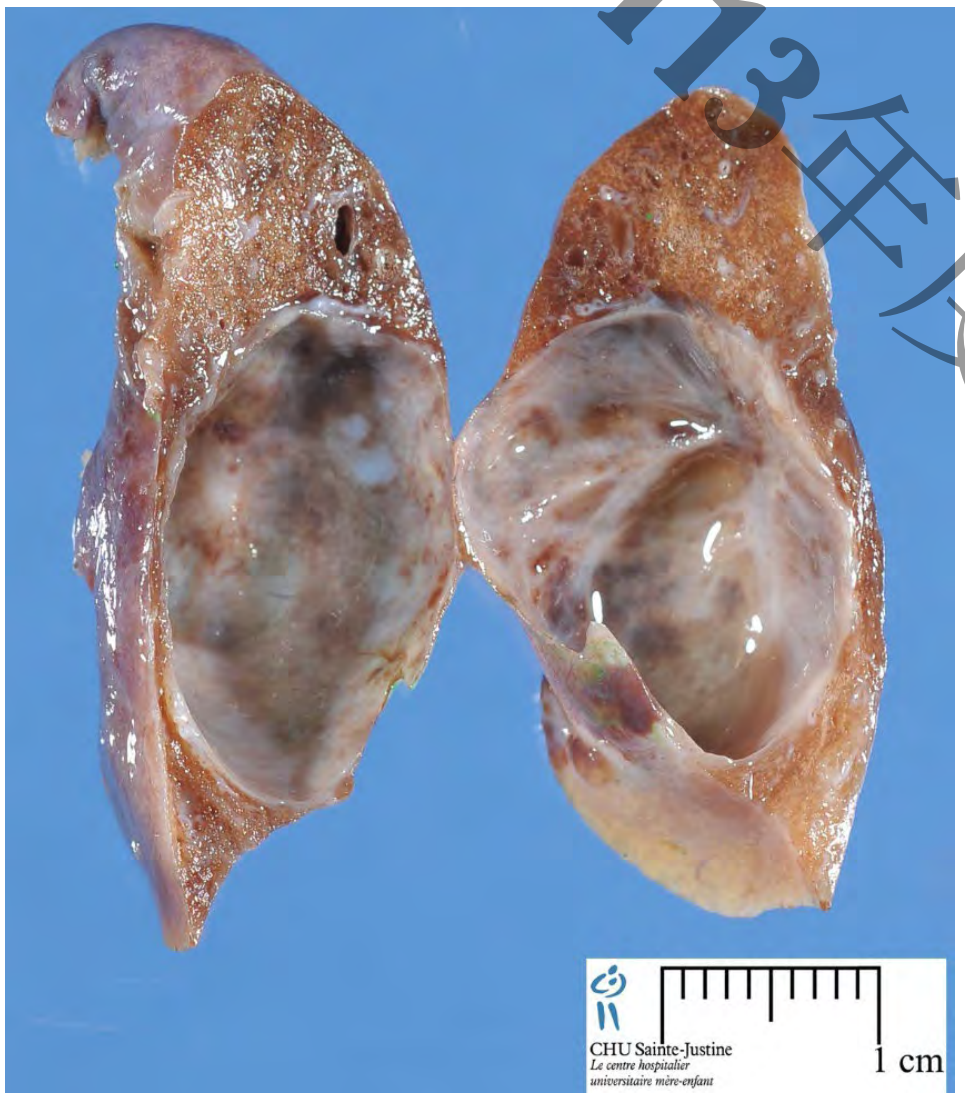
malignancy change

solid

Large cystic



Type I



Ciliated psudostratified columnar epi.

Type III



Cuboidal epi.

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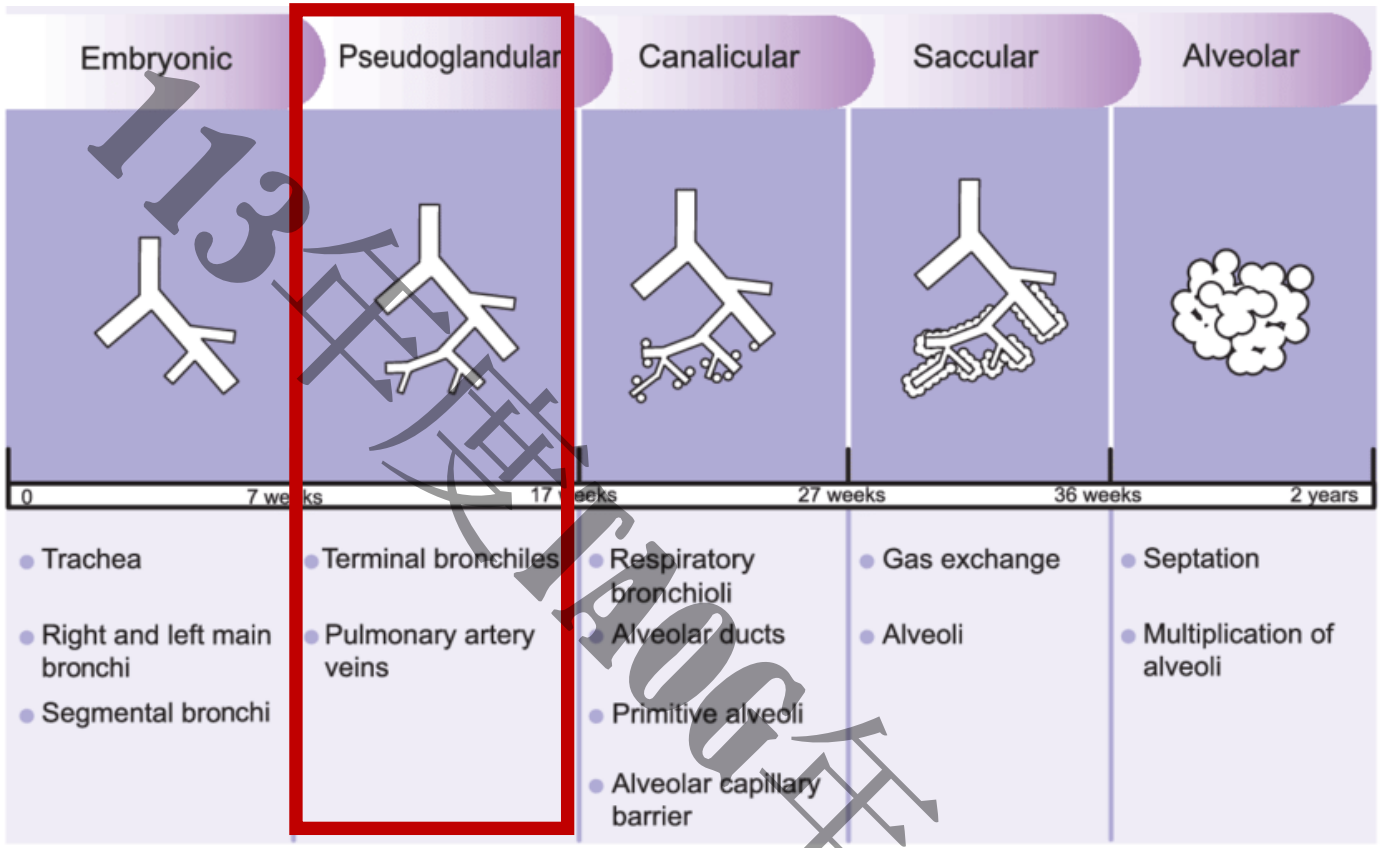

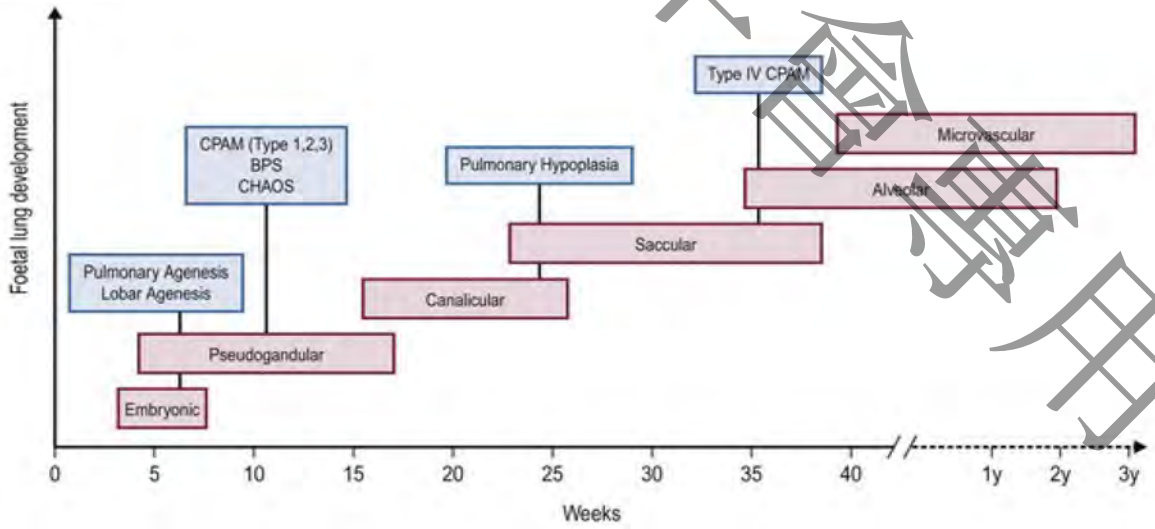

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Subtype distribution

Past (patho)-

Type I (macro-) : 65%

Type II (mixed): 15%

Type III (solid): 5%

Others: 10-15%

Current data (2018, prenatal) **-**

Macro – 26%

Micro (type III) – 74%

Outcome predictor

CPAM volume ratio

(**CVR>1.6**)

Only for prediction of hydrops

Not truly for all parameters

Management

Expectant (past)

to

Maternal steroid injection

Shunting or OK-432

(only for macrocystic)

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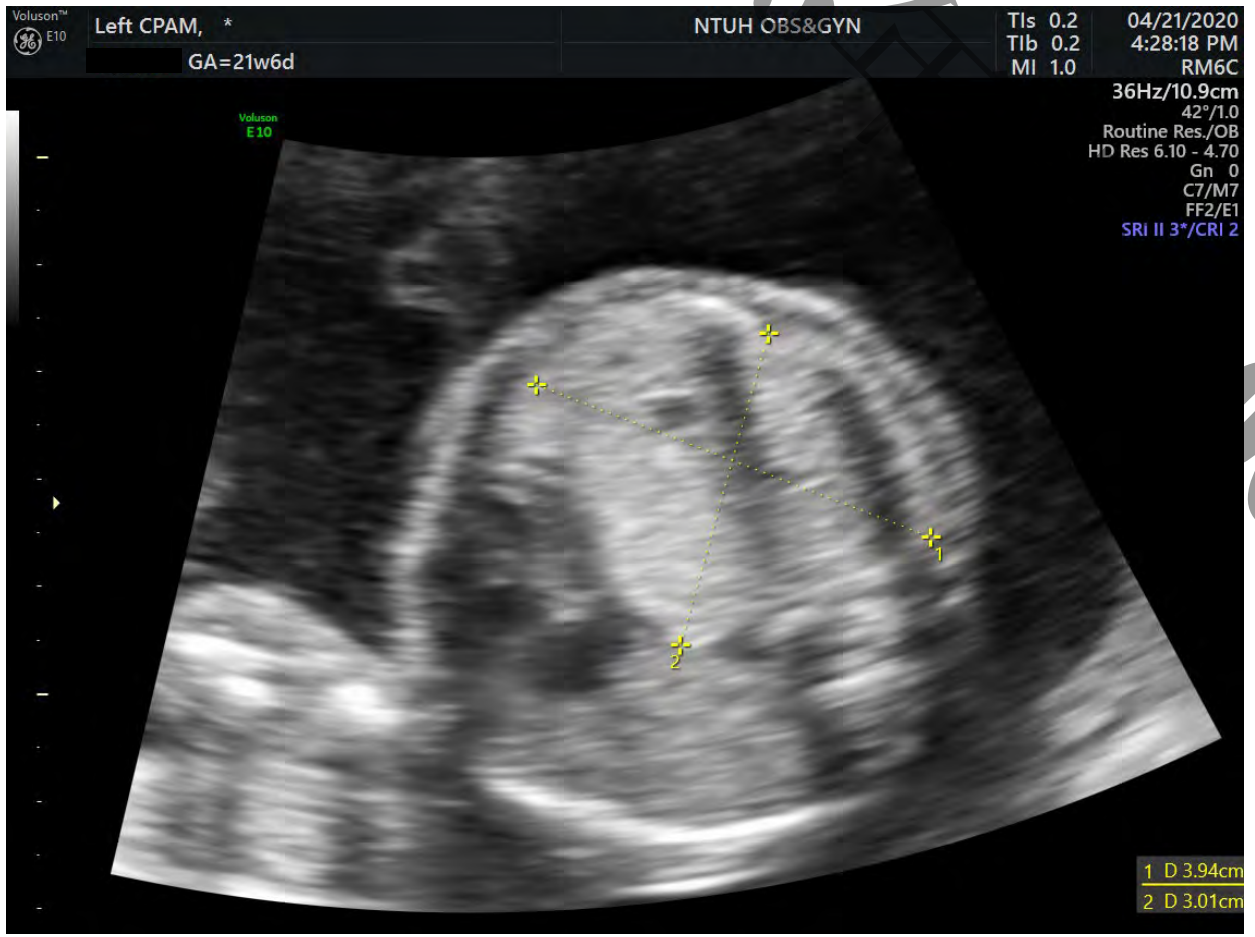
CVR 1.05

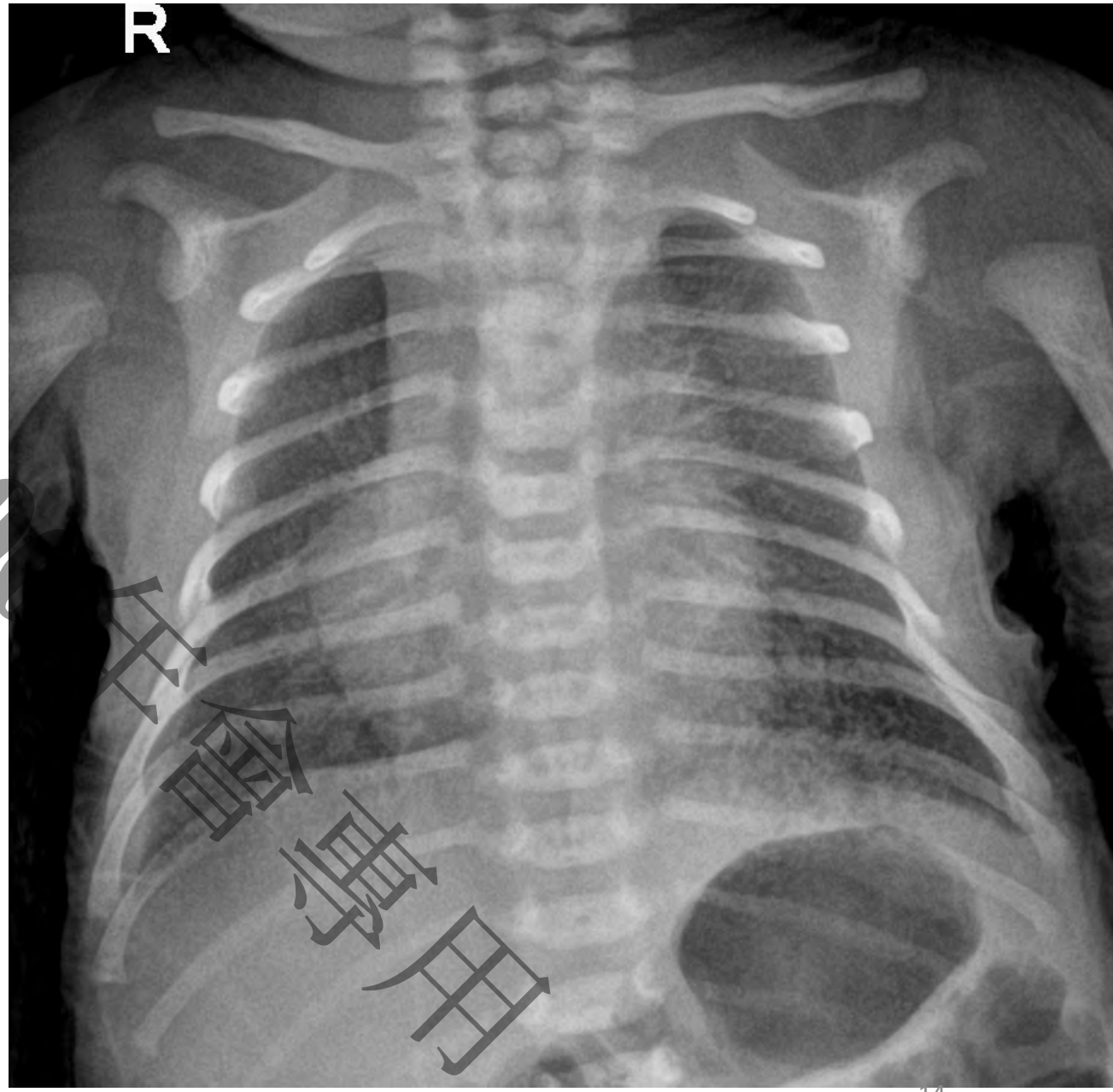
Type I macrocystic

CVR 1.16

(Type II Mixed)



Type III, CVR 1.10



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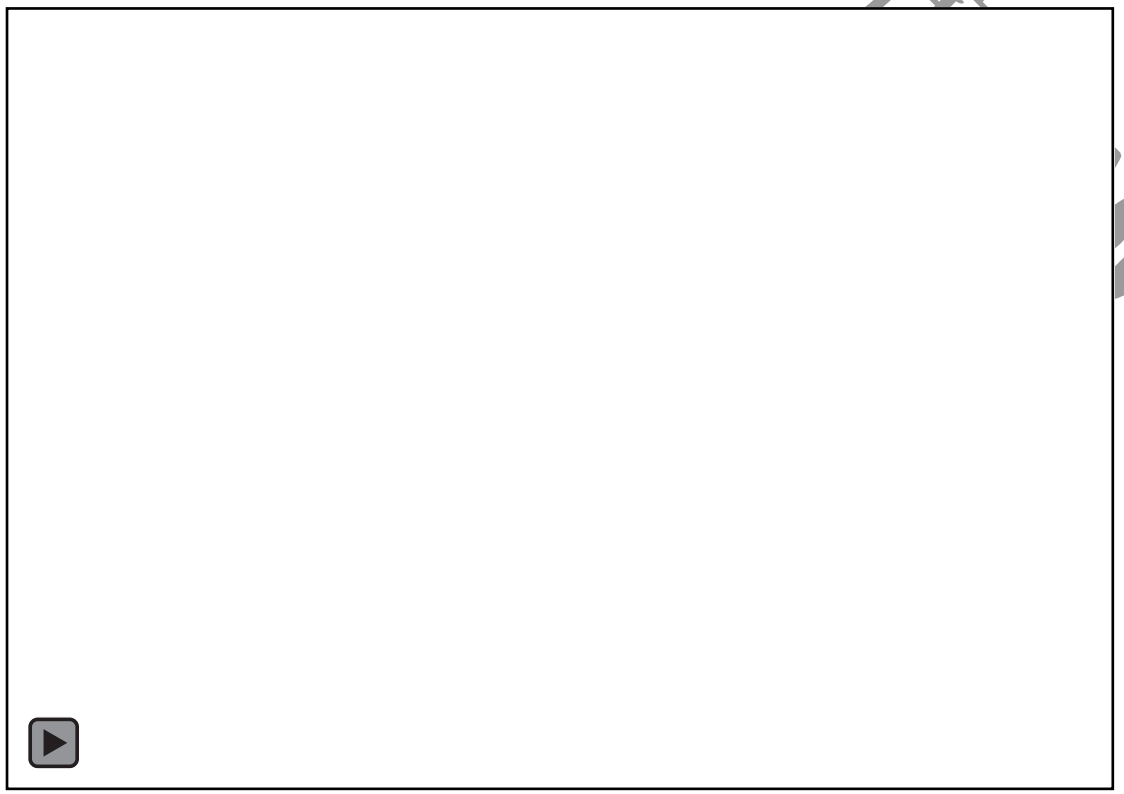
Comparison of the Prognostic Factors of Fetuses With Congenital Pulmonary Airway Malformations According to Type

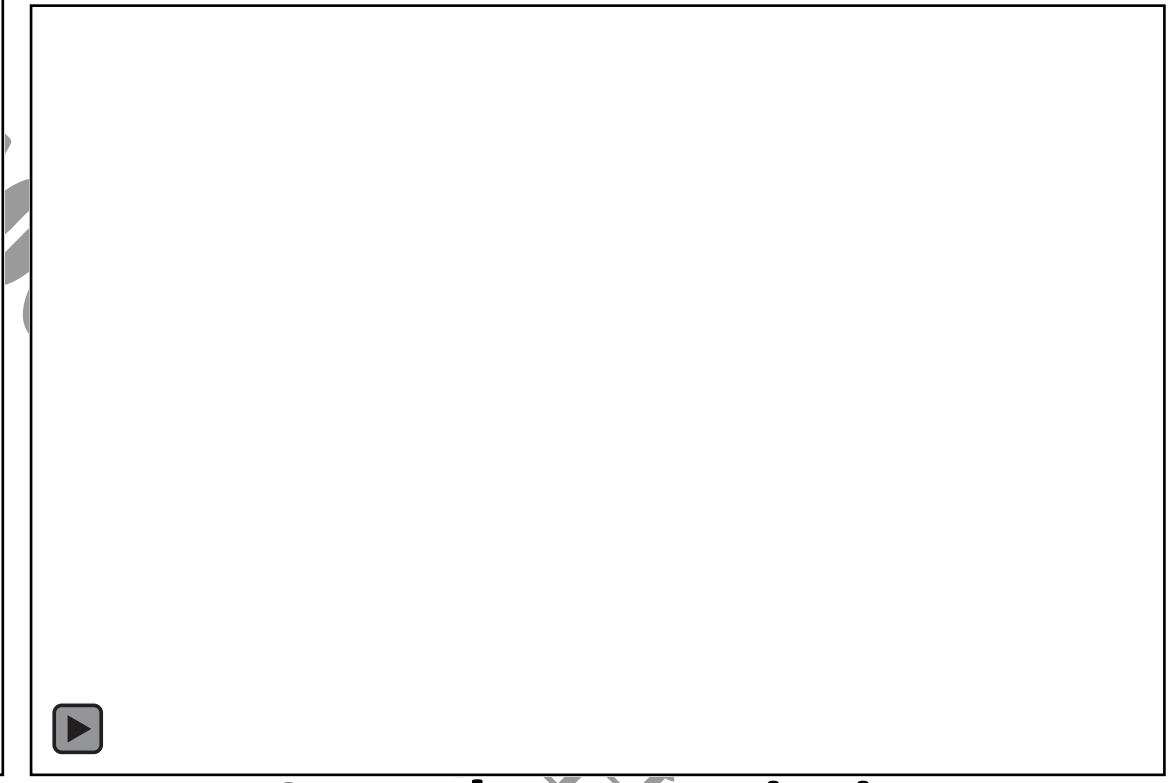
Ba-Da Jeong, MD, Su-ah An, MD*, Mi-Young Lee, MD, PhD, Hye-Sung Won, MD, PhD, Minkyu Han, PhD, Heemang Yoon, MD, PhD, Ji-Hoon Lee, MD, Yoon-Jung Cho, MD*

N = 118 fetuses

J Ultrasound Med 2020; 39:2243–2252



Hydrops



Complete remission

Table 1. Prenatal Characteristics of the Fetuses With CPAMs

Characteristic	Study Population (n = 118)		Microcystic CPAM (n = 46)		Macrocystic CPAM (n = 72)		*P*
GA at diagnosis, wk	23.2	(17.4–32.3)	23.2	(17.6–32.3)	23.2	(17.4–28.5)	.334
GA at last follow-up, wk	32.1	(21.1–38.6)	32.2	(26.6–38.6)	32.1	(21.1–38.4)	.383
Location							
Unilateral (left/right)	116	(45/71)	45	(16/29)	71	(29/42)	.569
Bilateral	1	(0.8)	0		1	(1.4)	
Extrapulmonary	1	(0.8)	1	(2.2)		0	
CPAM size							
Initial volume, cm^3	16.7	(0.4–110.8)	10.7	(0.4–73.8)	18.4	(2.3–110.8)	.037
Initial CVR, cm^2	0.76	(0.02–5.8)	0.5	(0.02–3.5)	0.85	(0.11–5.8)	.039
Maximum volume, cm^3	18.8	(0.4–119.4)	13.3	(0.4–97.3)	21	(3.1–119.4)	.012
Maximum CVR, cm^2	0.80	(0.02–5.9)	0.58	(0.02–4.10)	0.95	(0.16–5.9)	.008
Maximum cyst size, mm					12.4	(5.0–47.3)	
Mass effect							
Mediastinal shift	89	(75.4)	33	(71.7)	56	(77.8)	.457
Pleural effusion	5	(4.2)	0		5	(6.9)	.155
Ascites	10	(8.5)	1	(2.2)	9	(12.5)	.086
Polyhydramnios	12	(10.2)	6	(13.0)	6	(8.3)	.534
Fetal hydrops	7	(5.9)	0		7	(9.7)	.042
Changes in CVR on follow-up scans							.567
Increase	29	(24.6)	10	(21.7)	19	(26.4)	
Decrease	89	(75.4)	36	(78.3)	53	(73.6)	
Nonvisualization	8	(6.8)	7	(15.2)	1	(1.4)	.006
Intrauterine fetal death	2	(1.7)	0		2	(2.8)	.520

Data are presented as median (range) and number (percent).

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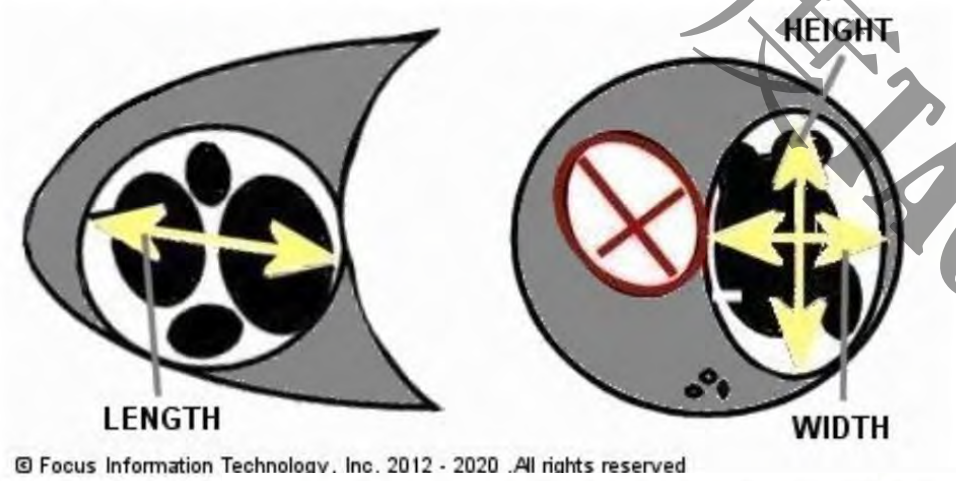
Congenital Pulmonary Airway Malformation Volume Ratio (CVR)

CPAM CVR calculator

https://perinatology.com/calculators/CVR.htm

- CVR = (Length X Height X Width X 0.52)/Head Circumference



The calculator below may be used to estimate the CVR.

Enter in centimeters:
Mass Length
Mass Width
Mass Height
Fetal Head Circumference

Calculate Reset

A **CVR > 1.6;**
or with **a dominant large cyst***
increases risk of developing hydrops.

*** cyst > 1/3 the size of the CPAM.**

1.2 < **CVR** < 1.6 - monitoring twice a week
CVR > 1.6 - monitoring 2 to 3 times per week

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Credit：Nav Photography: https://www.pexels.com/zh-tw/photo/5458389/

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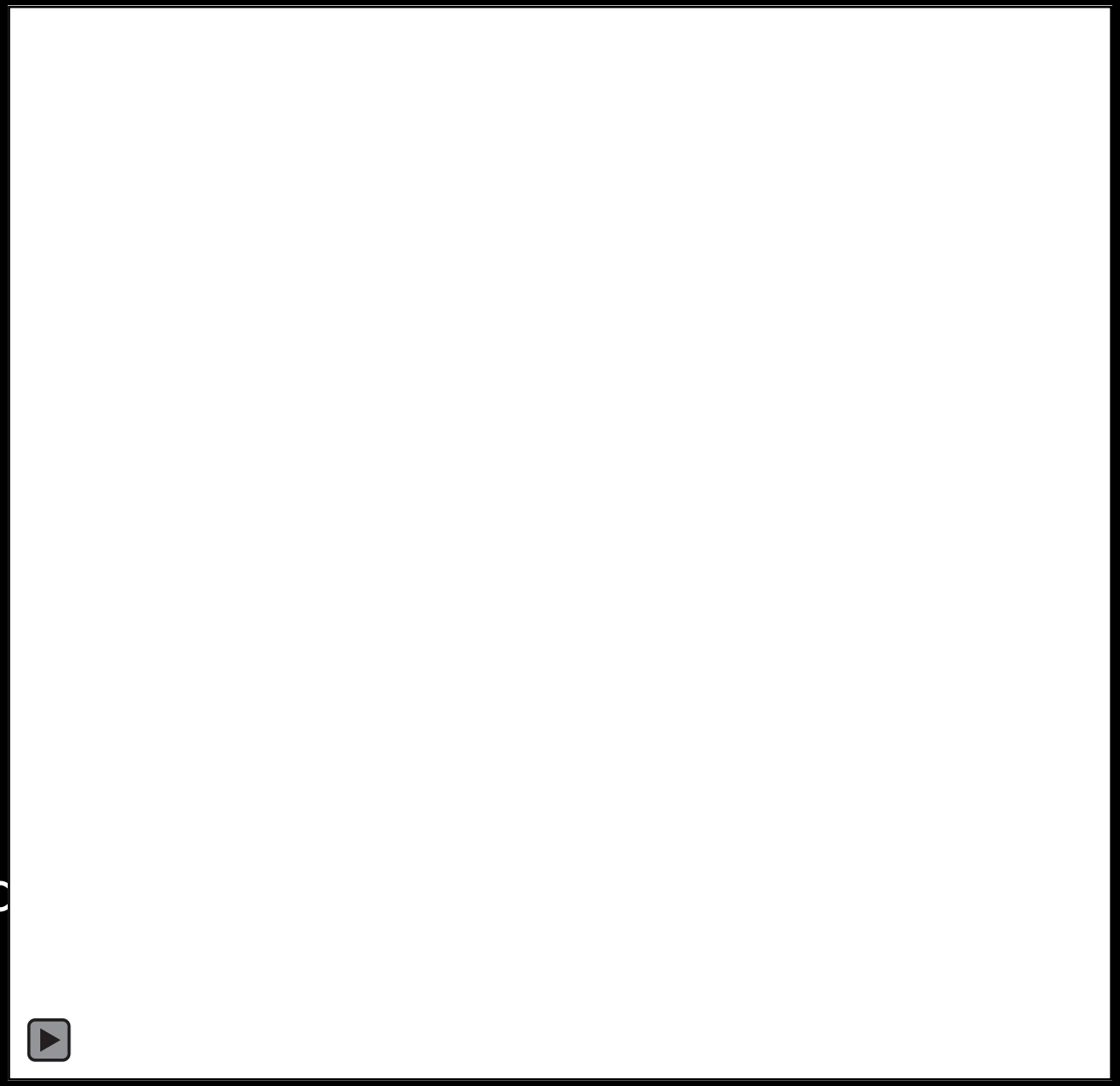
- grow rapidly at 20–28w, subsequent growth typically plateaus
- with 15% of CPAMs decreasing in size during the 2nd and 3rd trimesters

21w CVR = 2.37

4.86*4.80*3.85c





23w CVR = 2.0
4.84*4.67*3.90 cm



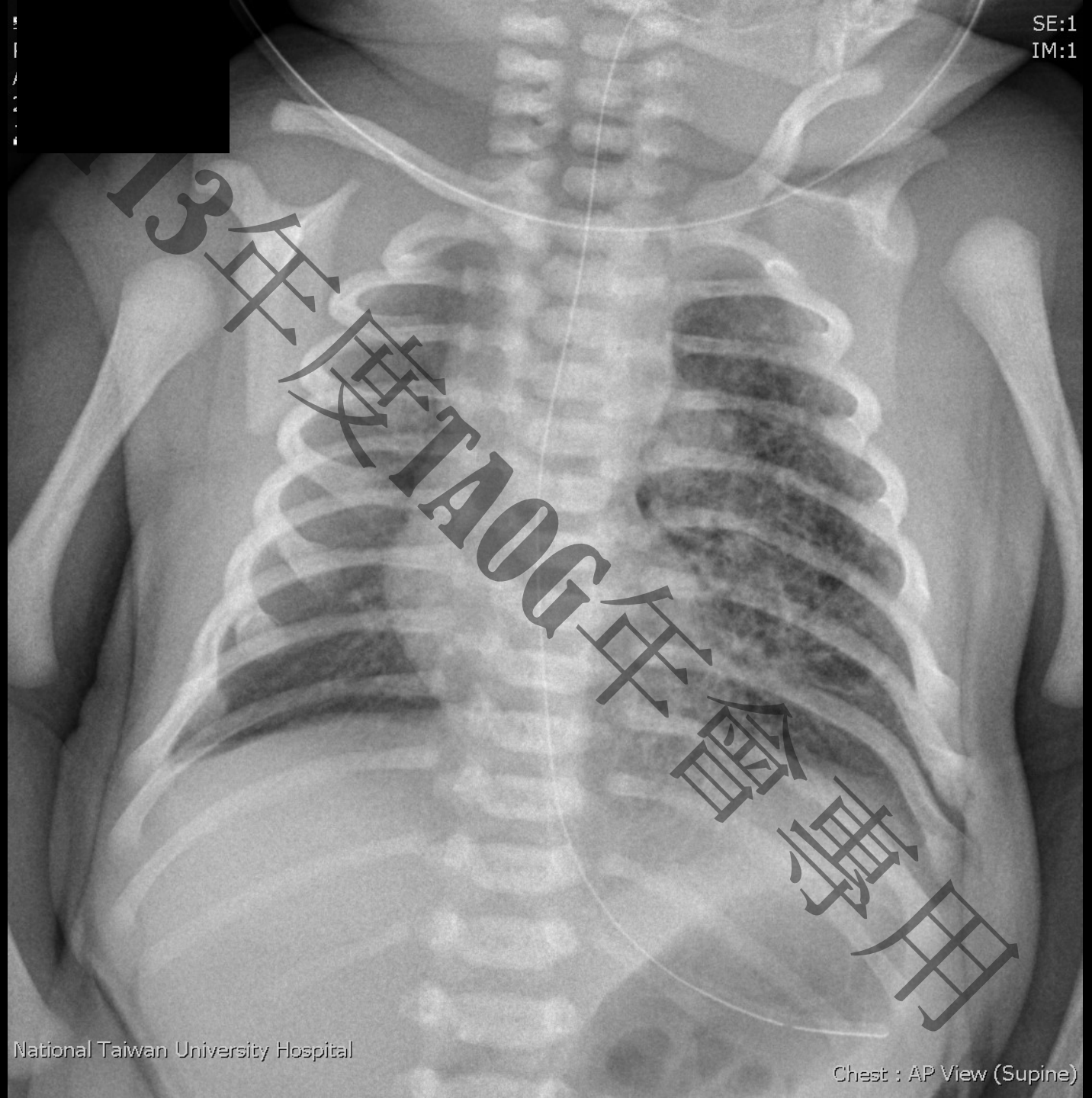
31w CVR= 0.87
3.56*3.5*3.82 cm,











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Hydrops fetalis

- The main outcome determinant
- hydrops (-): survival rate of nearly 95%

  whereas mortality rate was nearly 100% in hydrops (+) .



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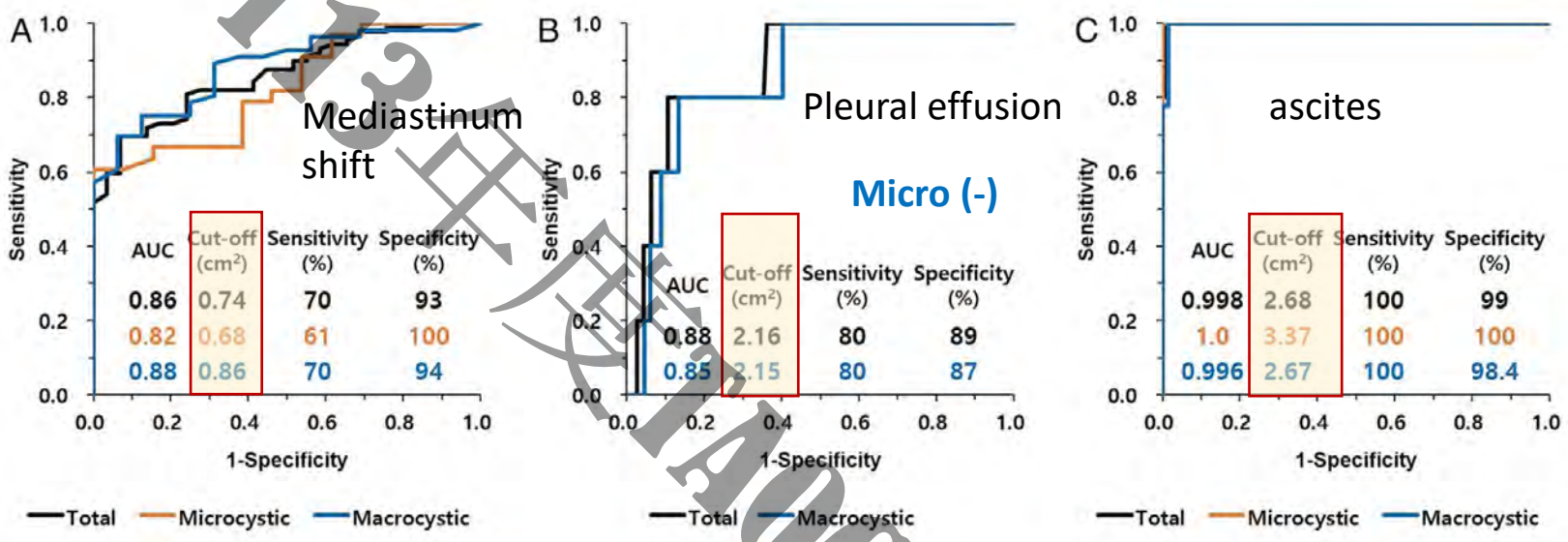
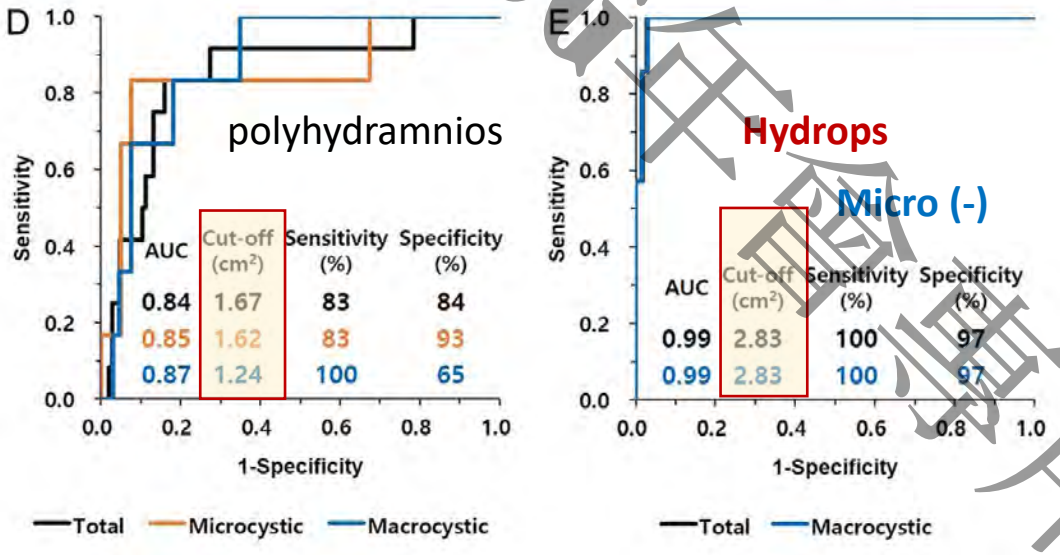
Figure 3. Receiver operating characteristic curves for the maximum values of the CVR and antenatal mass effects according to type. The mass effects are mediastinal shift (**A**), pleural effusion (**B**), ascites (**C**), polyhydramnios (**D**), and fetal hydrops (**E**). AUC indicates area under the curve.



CVR > 1.6 is only a high risk factor for hydrops fetalis, but not for other clinical parameters.

And It may be **higher** than 1.6 in other series to predict hydrops.



CVR 2.83 for hydrops

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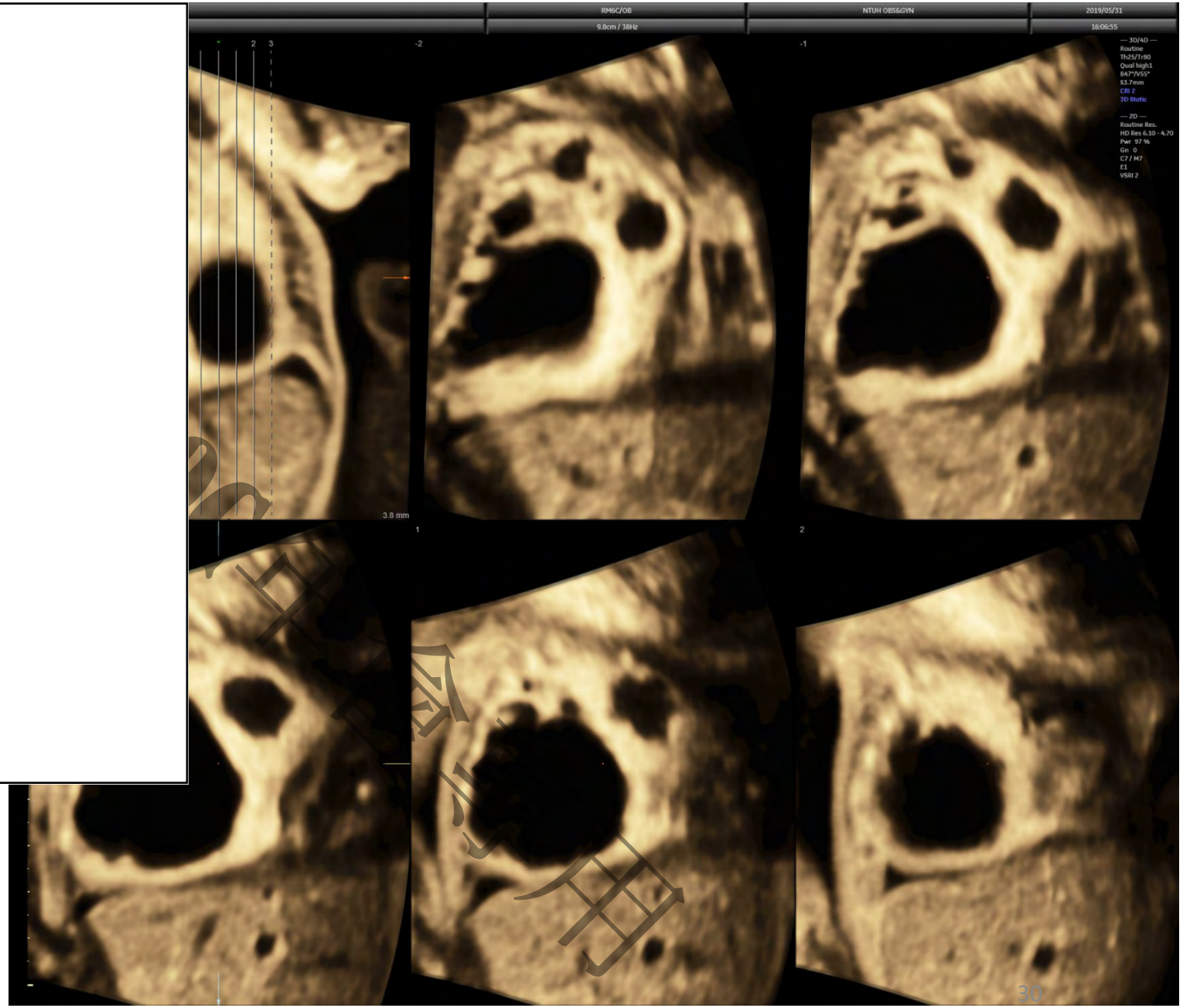
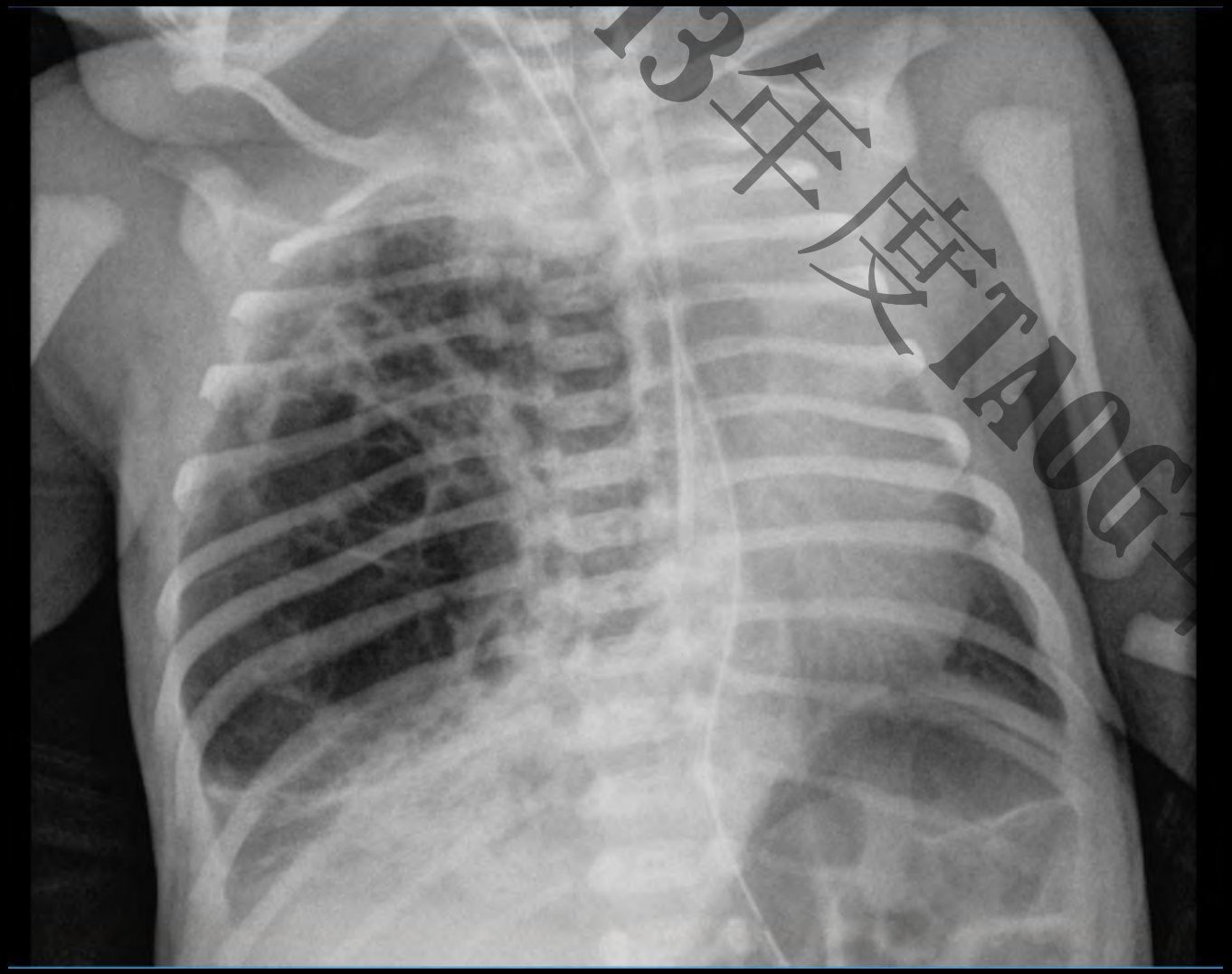
J Ultrasound Med 2020; 39:2243–2252

2247

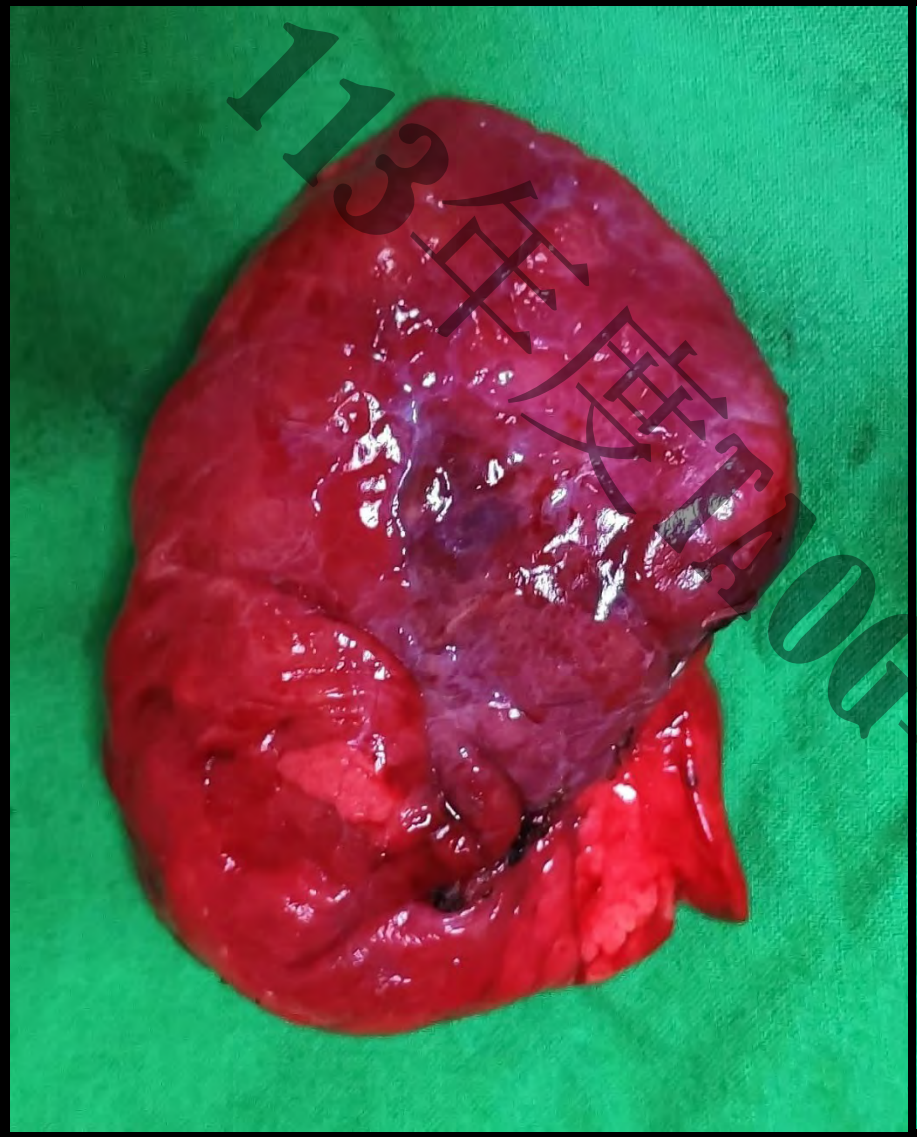


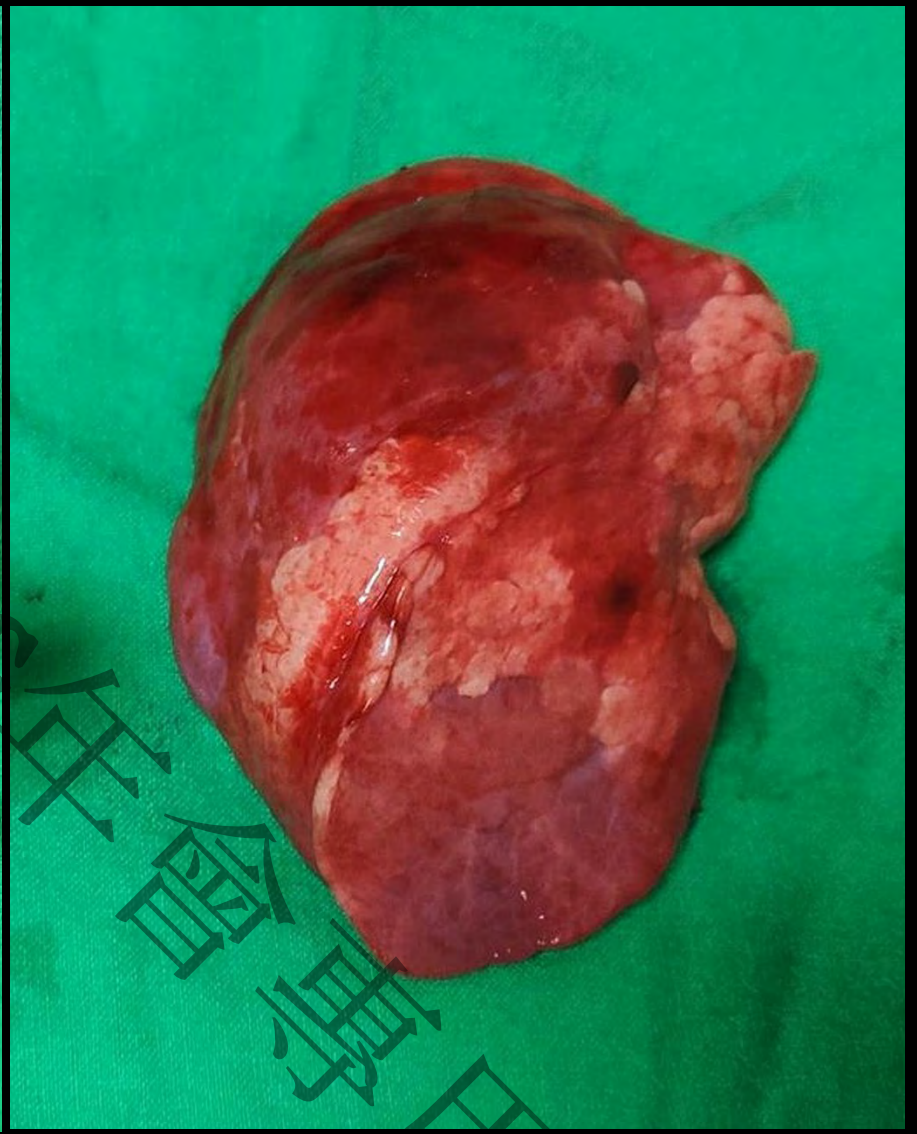
26w, CVR 5.17, Single dominant cyst
Ascites (+) and hydrocele
Chest tapping 3 times (100ml -> 50 ml -> 40 ml)
Rinderon 12 mg IM * 2 full courses
Shunt procedure was not performed due to license.





PROM @ 36w, vaginal birth. Baby – presented with respiratory distress after birth





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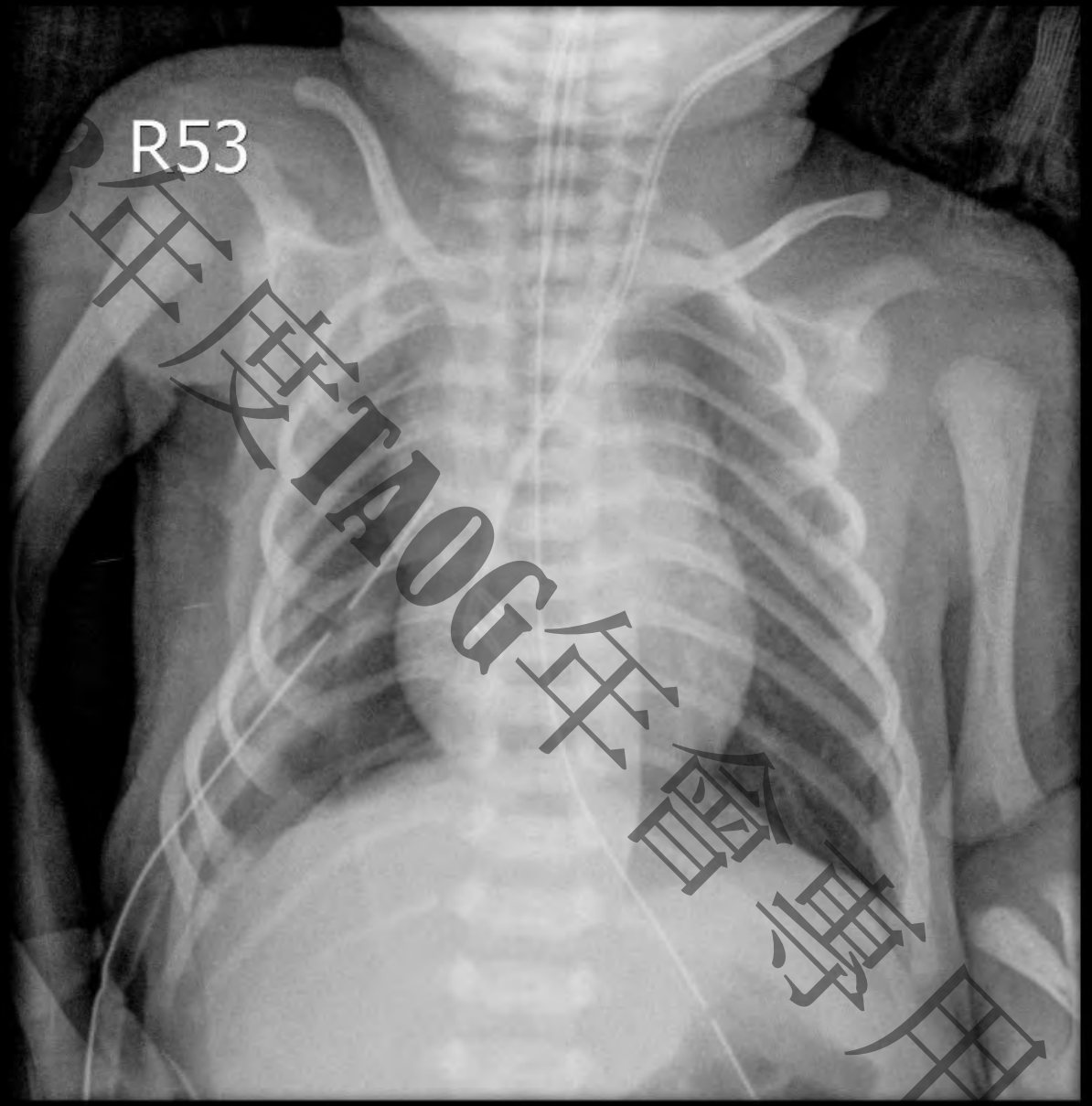

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Life is beautiful and a miracle

when you recognized

the evidence from the literature

is not always the truth.

* With parent's permission

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Association for Academic Surgery

Maternal Steroids in High-Risk Congenital Lung Malformations

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[a] *Department of Surgery, Baylor College of Medicine, Houston, Texas*
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Single vs. **multiple** courses?

Multiple steroid courses have a potential positive impact on **congenital lung malformation (CPAM and non-CPAM lung lesions)**.

Given the low risk-benefit ratio of maternal steroids, physicians could consider use of **multiple steroid courses** for CLM **refractory** to a single course.

仅供ITOR年会演讲用

Initial CVR = 0.62 @22
Macrocystic type (Type
OK 432 injection since

Delivery at term by CS



*Antenatal administration of steroid

vs.

open fetal surgery

Journal of Pediatric Surgery (2009) 44, 60–65

ELSEVIER

Journal of Pediatric Surgery

www.elsevier.com/locate/jpedsurg

High-risk fetal congenital pulmonary airway malformations have a variable response to steroids

Lee M. Morris[a], Foong-Yen Lim[a], Jeffrey C. Livingston[b], William J. Polzin[c], Timothy M. Crombleholme[a,*]

2004-2008

Table 1 Primary and secondary endpoints comparing steroid versus open fetal surgery in fetuses with predominantly microcystic CCAM

	Steroids	Open fetal surgery	*P*
Mean gestational age at delivery (wk)	34	31	<.05
% Survival to birth	12/13 (92%)	9/11 (82%)	.58
% Survival to neonatal discharge	10/12 (83%)	5/9 (56%)	.33
% Resolution of hydrops	10/13 (77%)	2/11 (18%)	<.05
CVR (cm^2) total	2.68 ± 0.29	2.95 ± 0.31	.55
CVR (cm^2) non-survivors	3.66 ± 0.66	3.02 ± 0.40	.42
Mean gestational age at treatment (wk)	23 (20-26)	24 (20-29)	.49
Ventilator requirement	1/12 (8%)	8/9 (89%)	<.001

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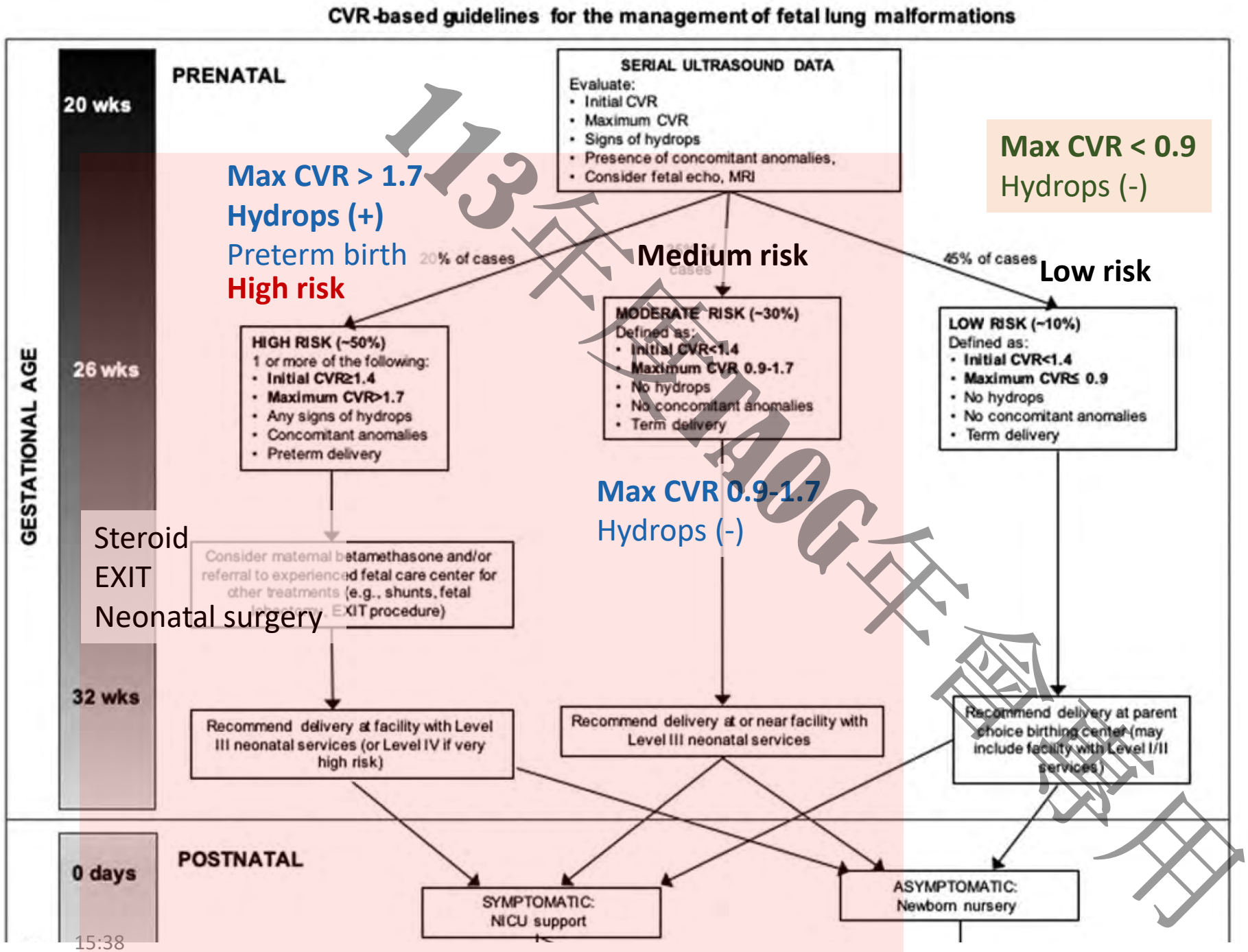

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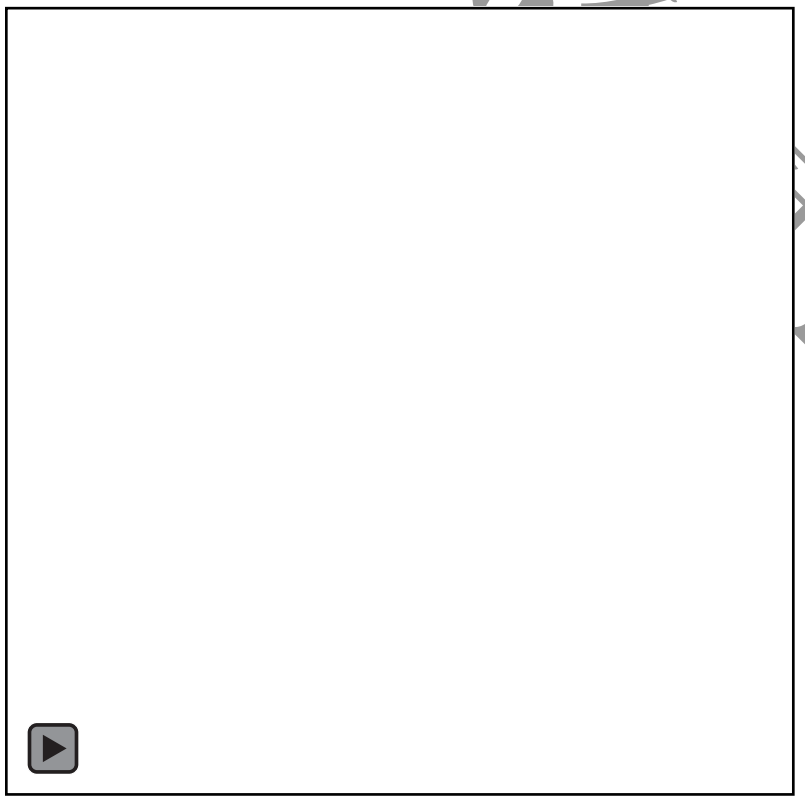
Initial CVR: measured at first visit

Max CVR: measured at the beginning of 3rd trimester

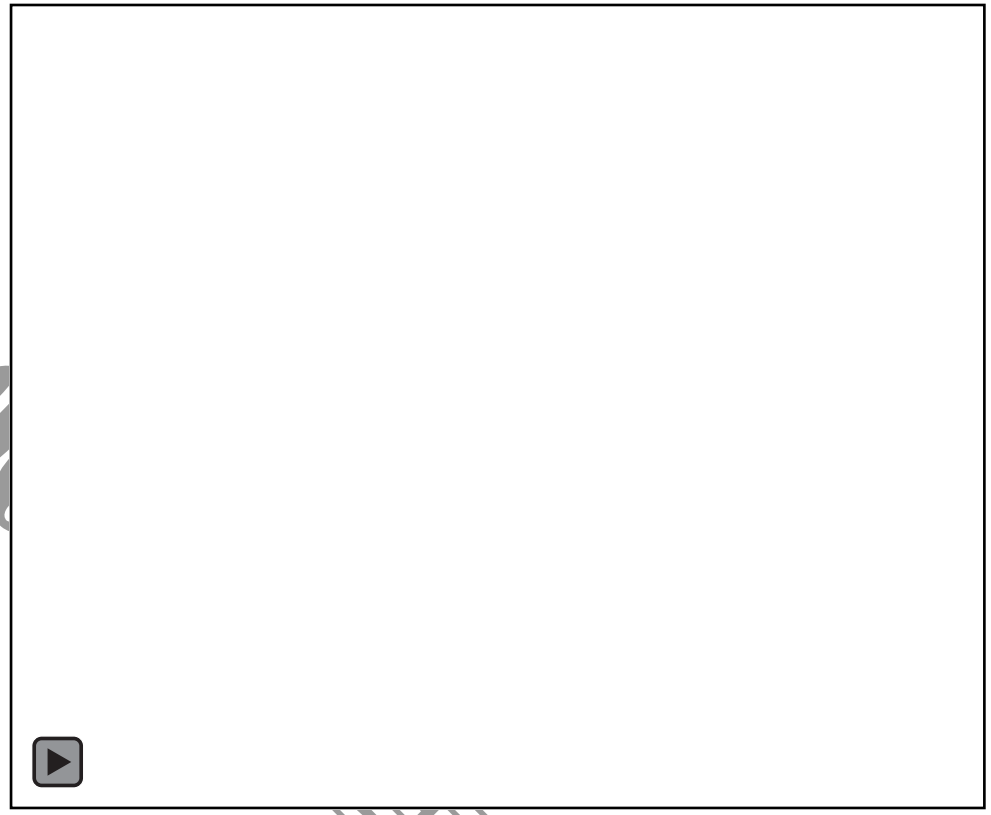
*High risk: respiratory distress at birth

Mortality cases

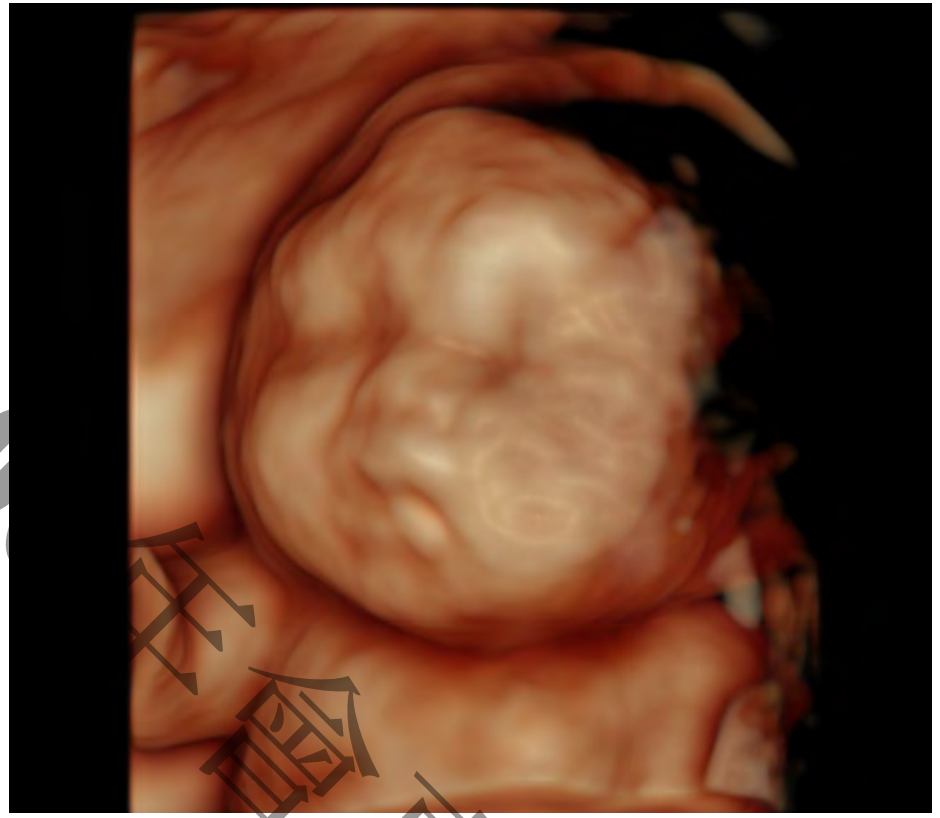
Congenital Pulmonary Airway Malformation, type I
(macrocystic type)



21w CVR 0.40



24w CVR 0.76



28w Hydrops (+), polyhydramnios
CVR 1.05

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30w
EXIT surgery



13年度TAOS年會專用



NND at D7

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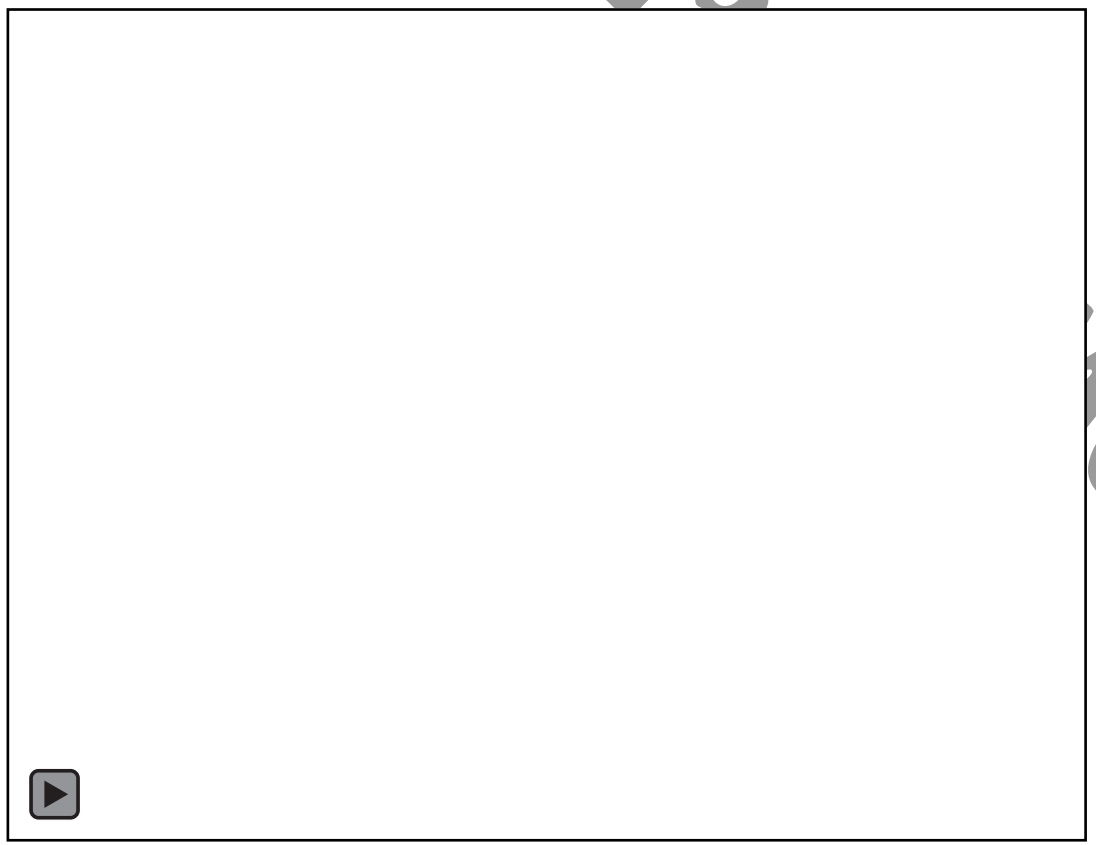
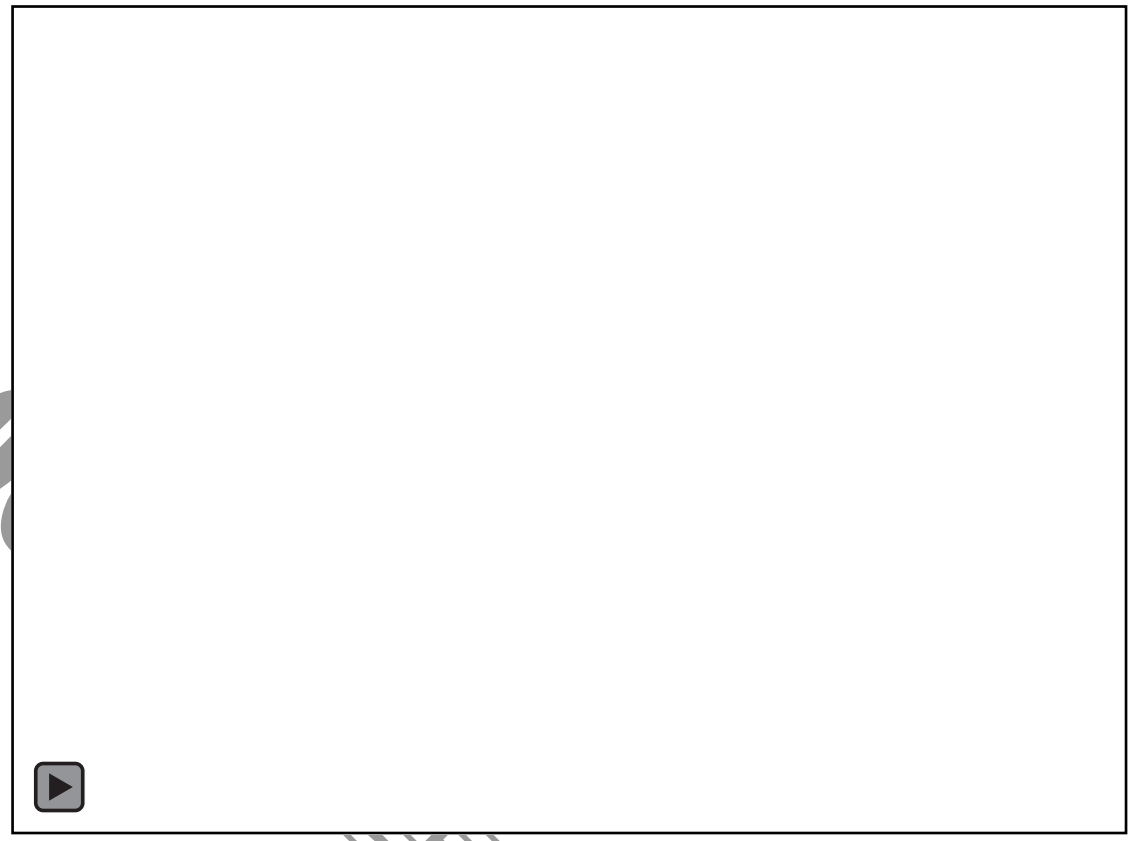
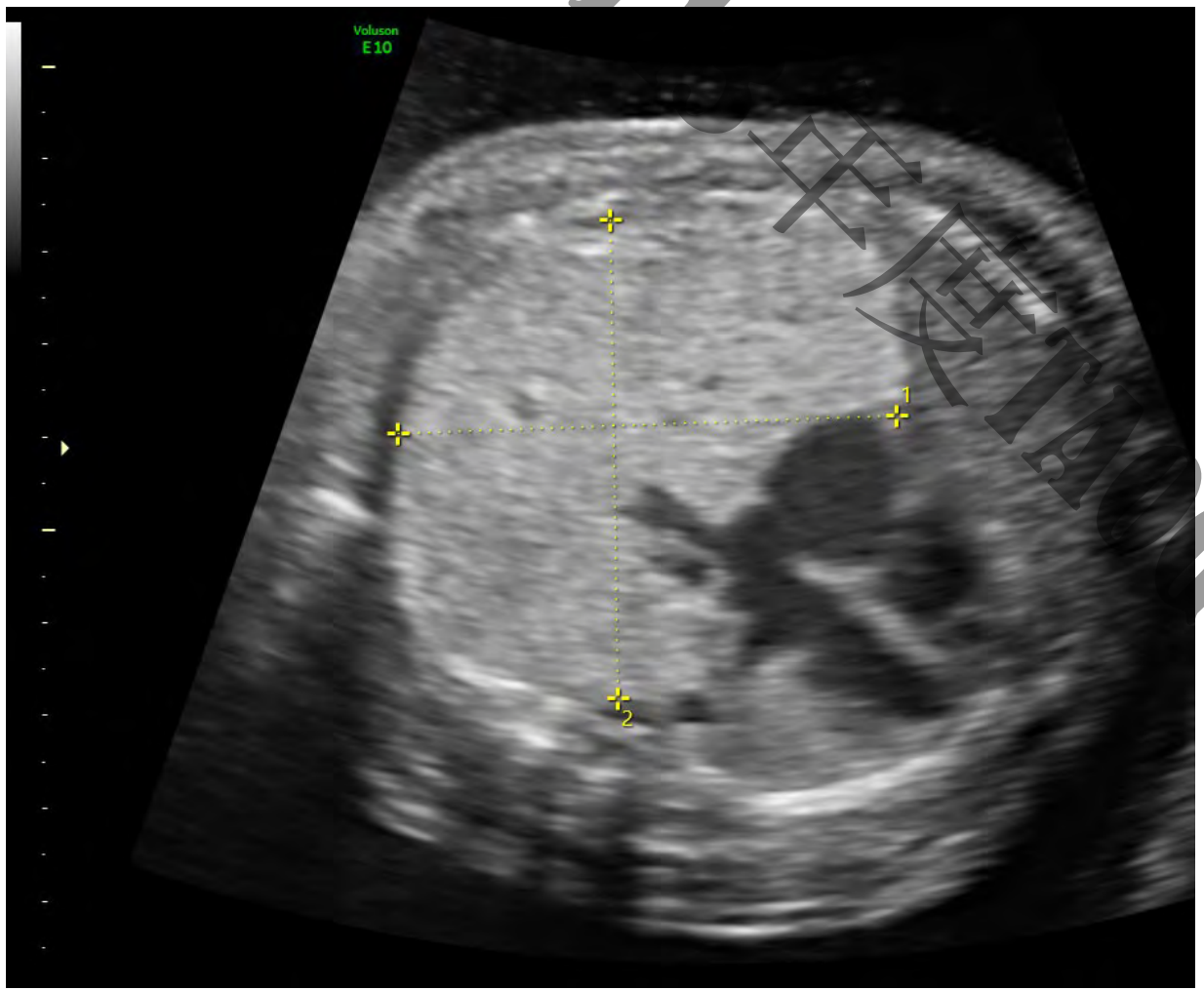




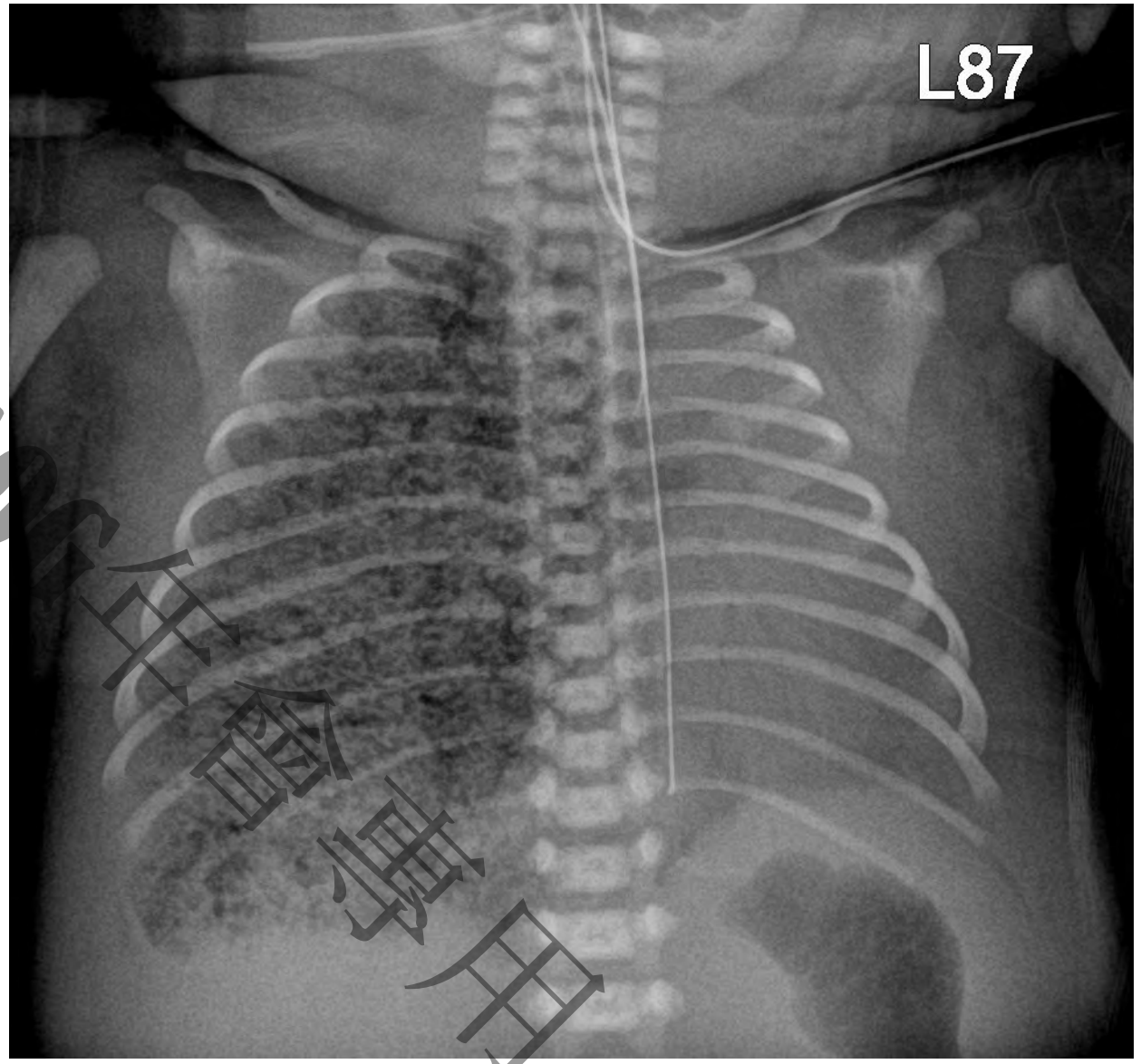
19w CVR 2.4

Type III CPAM



CVR 4.0 (27w) hydrops





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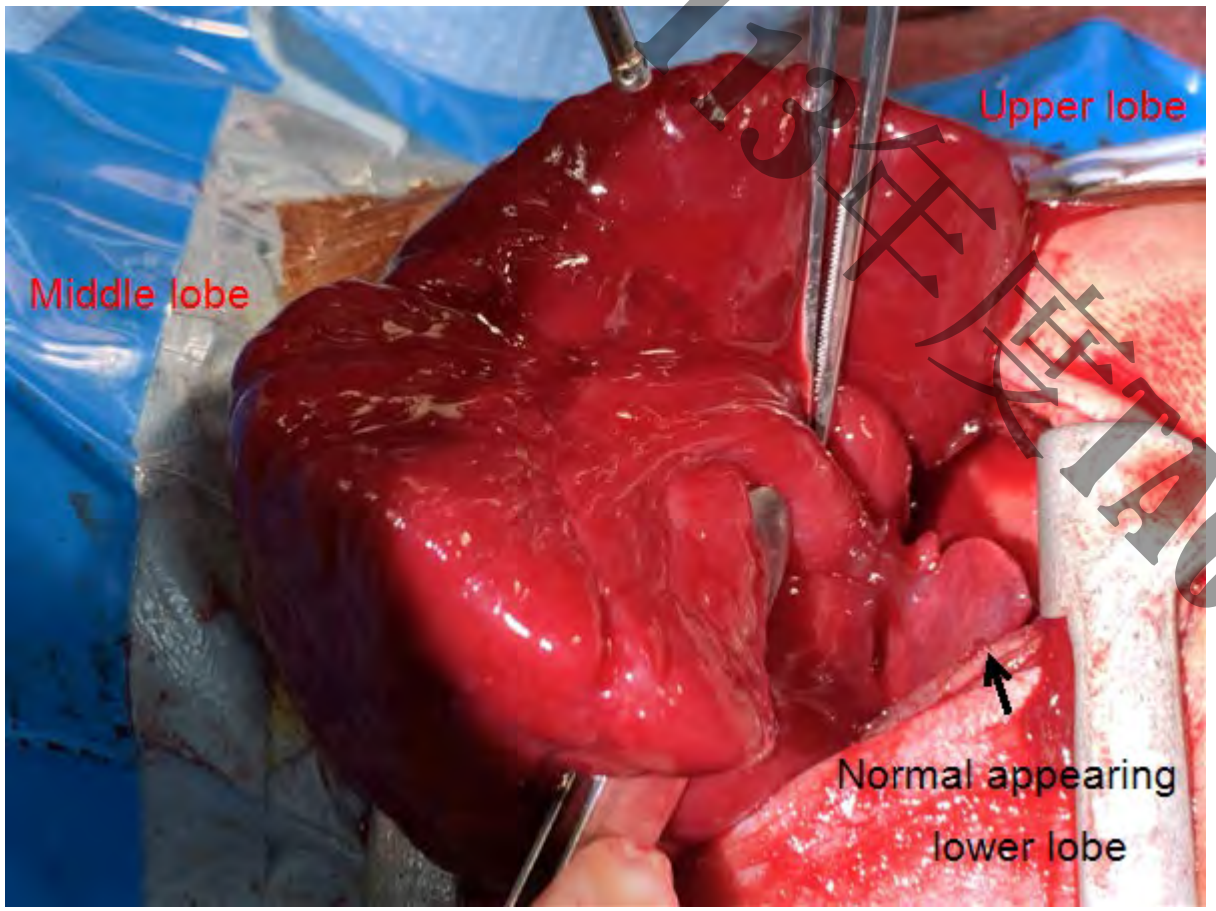
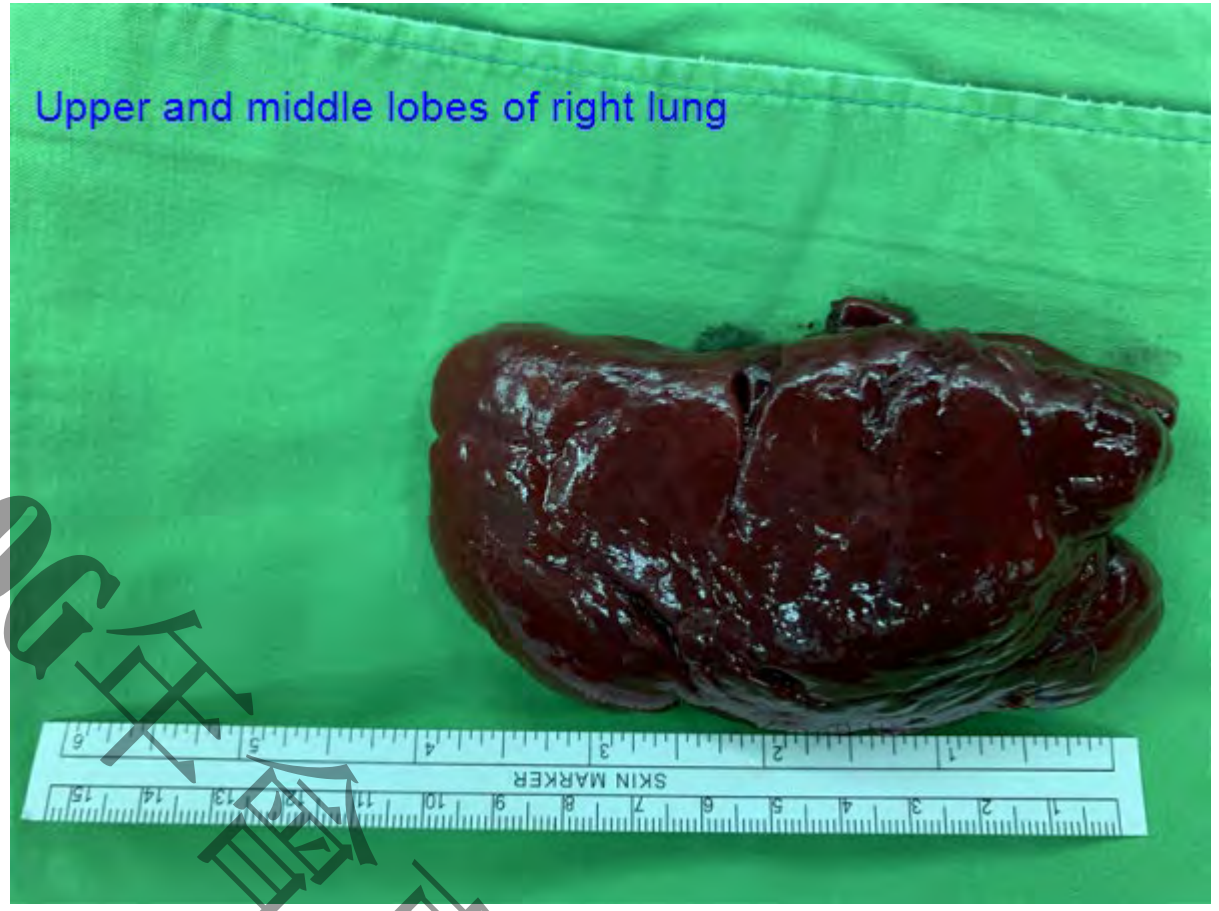




113年度TAOG年會專用



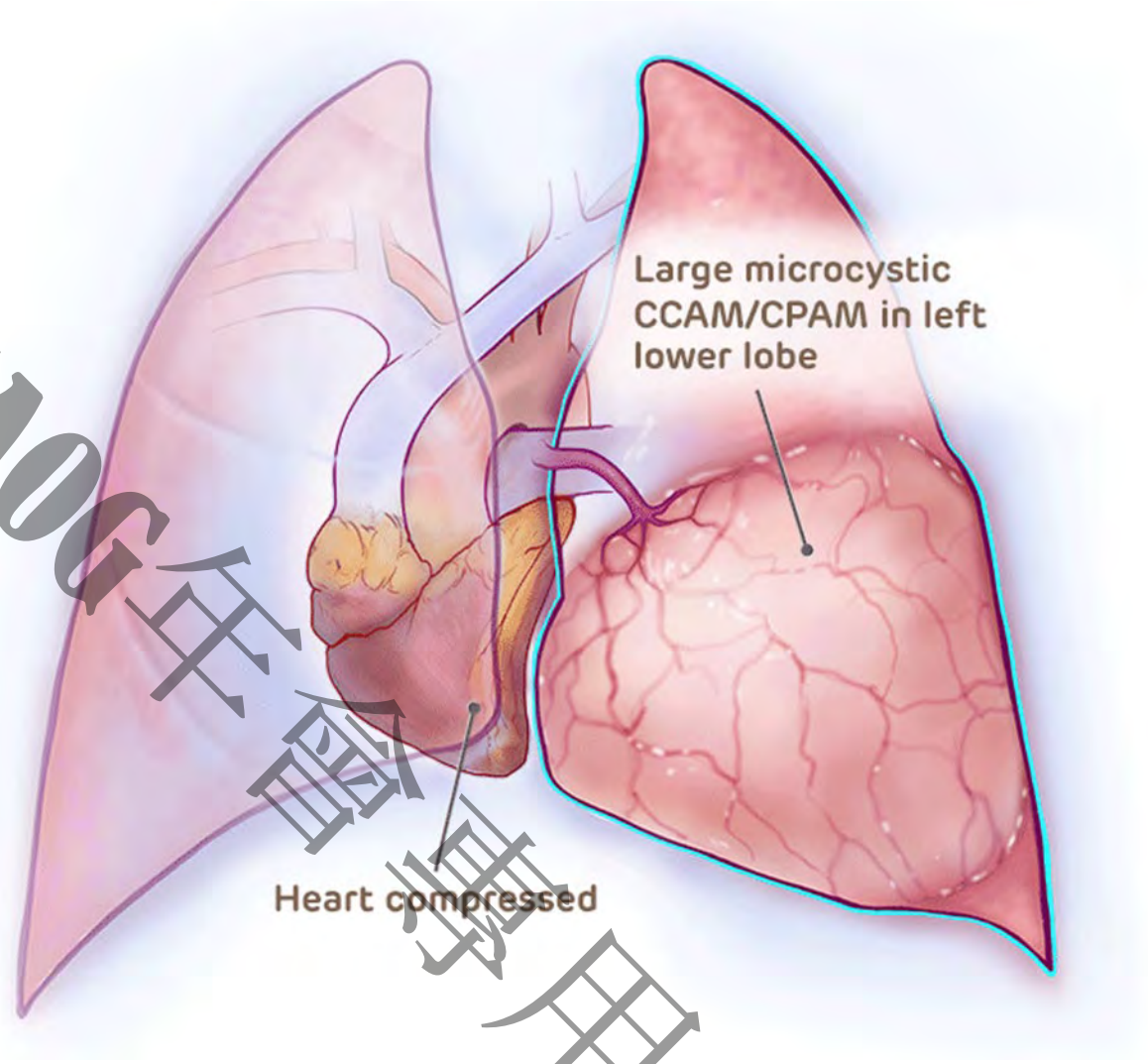
113年度T10C年度畢業

Lung mass

Not always CPAM

DDx

1. Broncho-pulmonary sequestration (BPS)
2. Hyperinflation (Lobar emphysema; no need for surgery)
3. Bronchogenic cyst
4. CHAOS (lethal)



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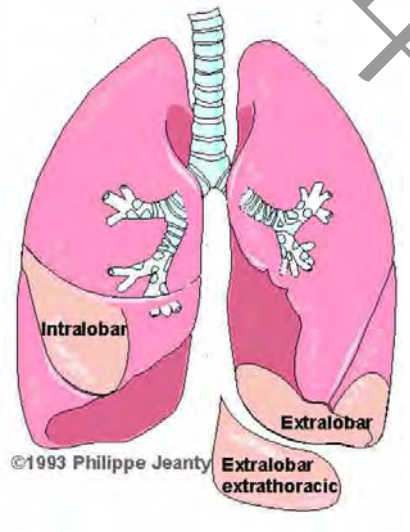
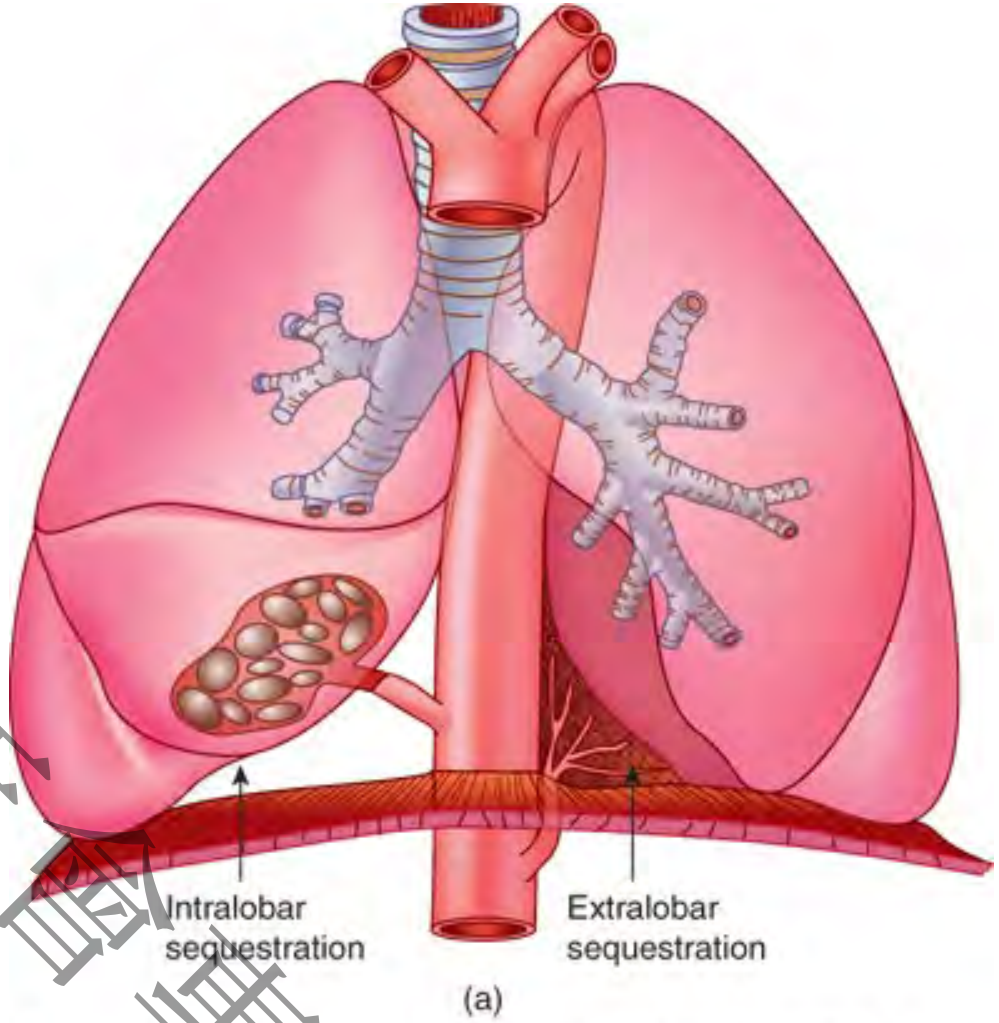
II. Bronchopulmonary sequestration (BPS)

1. abnormal, non-functional pulmonary tissue without connection to the tracheobronchial tree.
2. systemic artery supply (mostly descending aorta)
3. Intralobar (75%) and extralobar (25%).
4. Sub-diaphragmatic extralobar pulmonary sequestration vs. neuroblastoma





Source: Andrew J. Lechner, George M. Matuschak, David S. Brink: Respiratory: An Integrated Approach to Disease
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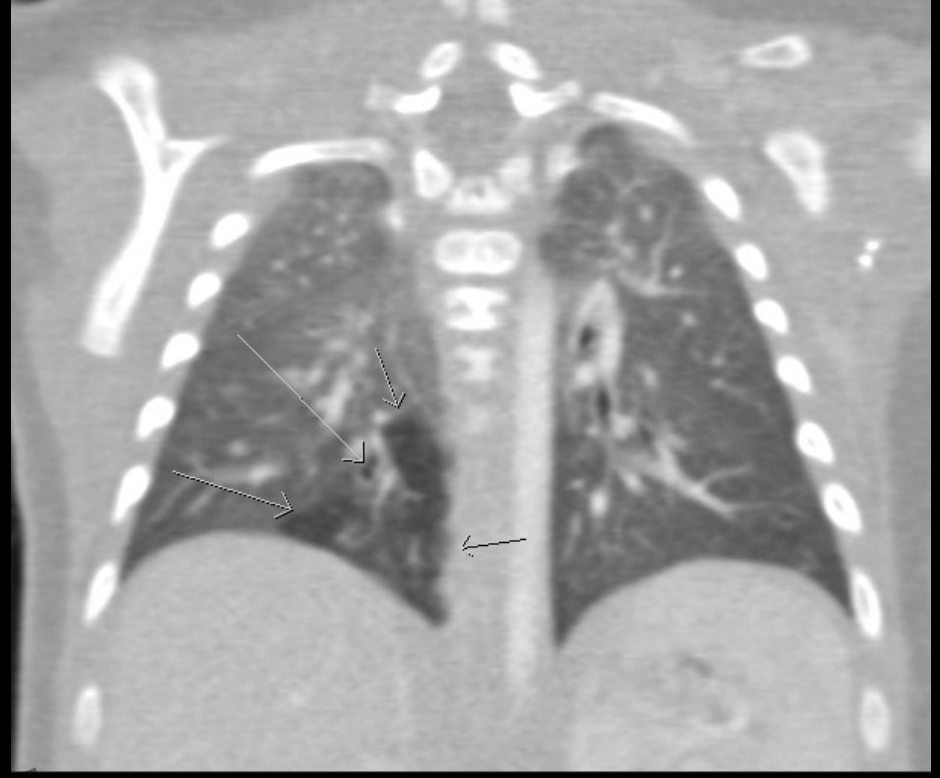
III. Congenital lobar emphysema (overinflation)





CPAM or BPS?

CP
@2
Rir

Lu

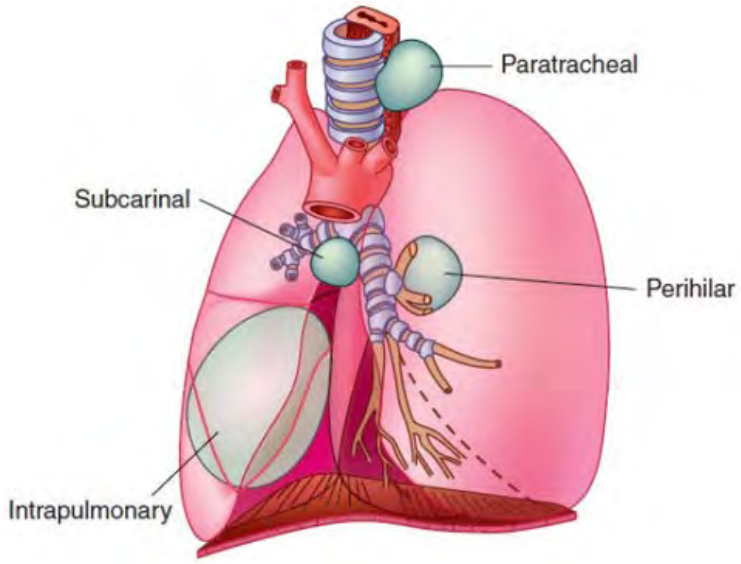


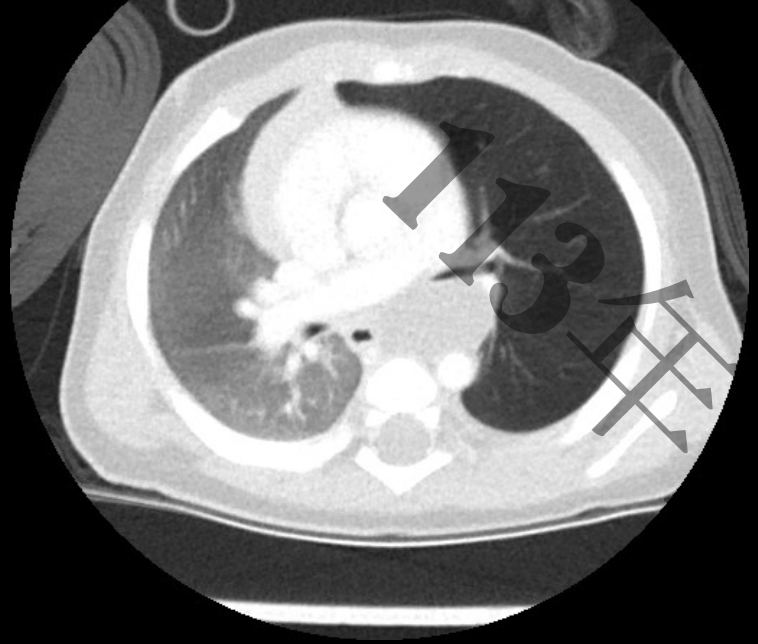
ysema

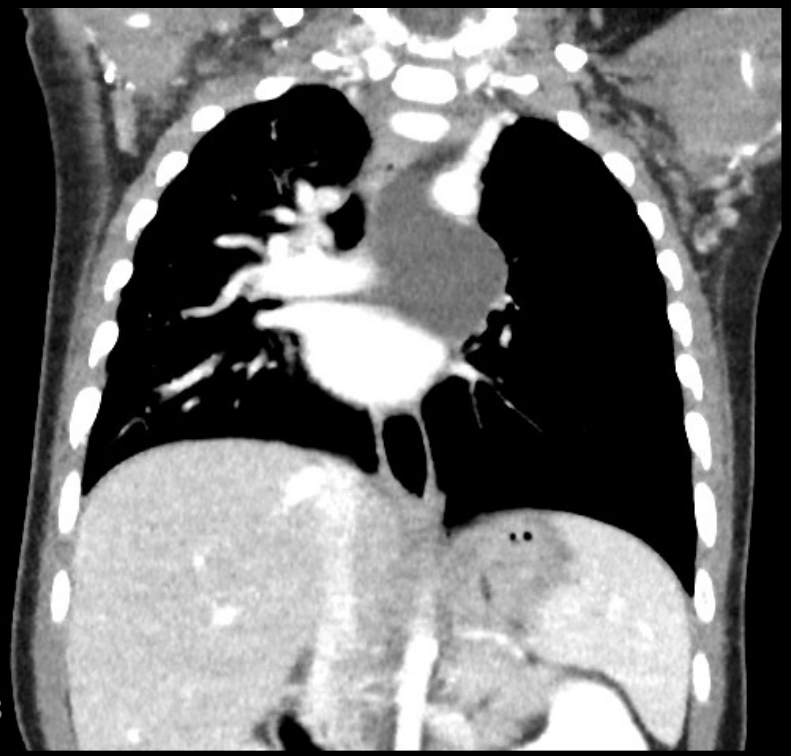
III. CHAOS: congenital high airway obstruction syndrome






Paratracheal
Subcarinal
Perihilar
Intrapulmonary





R



L

I

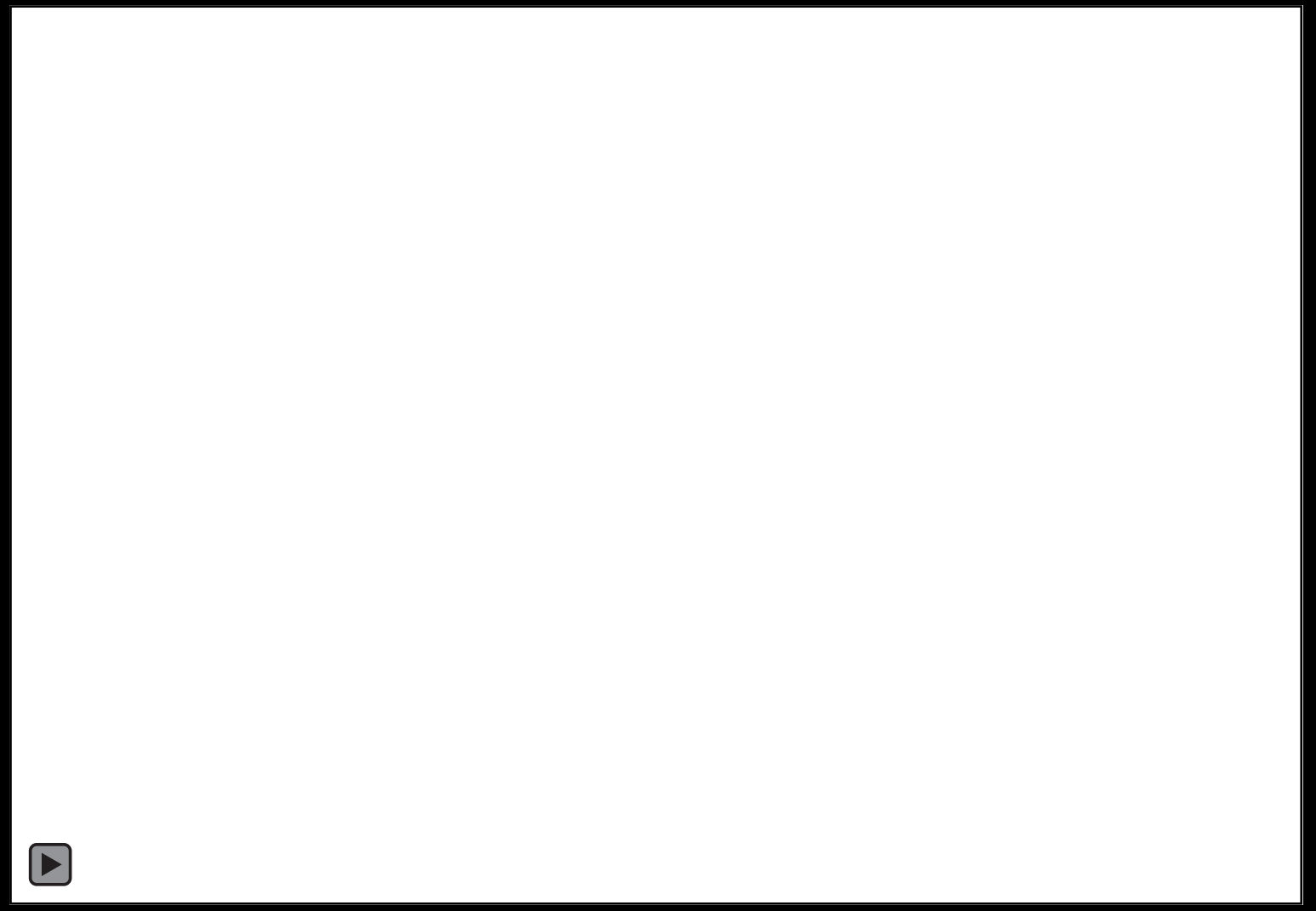
Standard 3D
VR: Pulmonaries
Segmented

15:38

59

Congenital diaphragmatic hernia

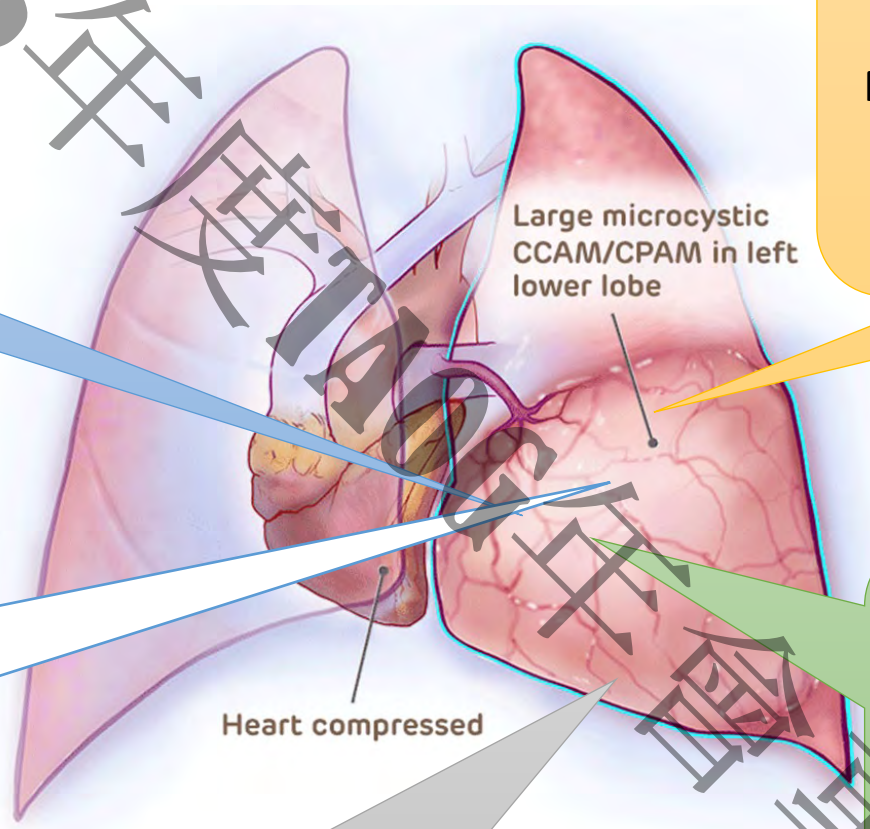
21w



Take home message

Incidence
more **common** than you thought

Subtype
Microcystic (solid) type constitutes the majority form of CPAM
Best outcome subtype



Outcome
Much **better** than you assumed

Risk-based management
Betamethasone injection (CVR> 1.6?)
or
shunt/OK-432 injection (macrocystic)

CVR> 1.6 is not a gold standard for all risk assessment

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