Congenital Pulmonary Airway Malformations

A Case Series

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Congenital pulmonary airway malformations (CPAM), previously known as congenital cystic adenomatoid malformation (CCAM) and before that cystic adenomatoid malformation of the lung (CAML), represent a spectrum of developmental abnormalities of the lung. The prognosis varies widely depending on the location and type of malformation. Sonography plays a essential role in the diagnosis and management of CPAMs. This case series will examine the different types of CPAMs, the role of sonography, and the prognosis and management of CPAMs.

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OBJECTIVES

- Know the pathogenesis of CPAMs
- Understand the different classifications of CPAMs and the sonographic appearance of each
- Discuss the role of sonography in the diagnosis and management of CPAMs

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EMBRYOLOGY AND ANATOMY

- 5 Stages of development
  - Embryonic (26 days – 6 weeks)
    - Laryngotracheal groove
    - Tracheal and lung buds
    - Primary bronchial buds and branches
  - Pseudo-glandular (6 – 16 weeks)
    - Formation of major airways
    - Respiration not possible yet
  - Canalicular (16 – 28 weeks)
    - Continued development and vascularization
    - Respiration possible late stage
  - Saccular Stage (28 – 36 weeks)
    - Number of terminal saccules increases
    - Surfactant production
    - Respiration and survival possible
  - Alveolar Stage (36 weeks – 8 years)
    - Continued development
    - Adequate surfactant production

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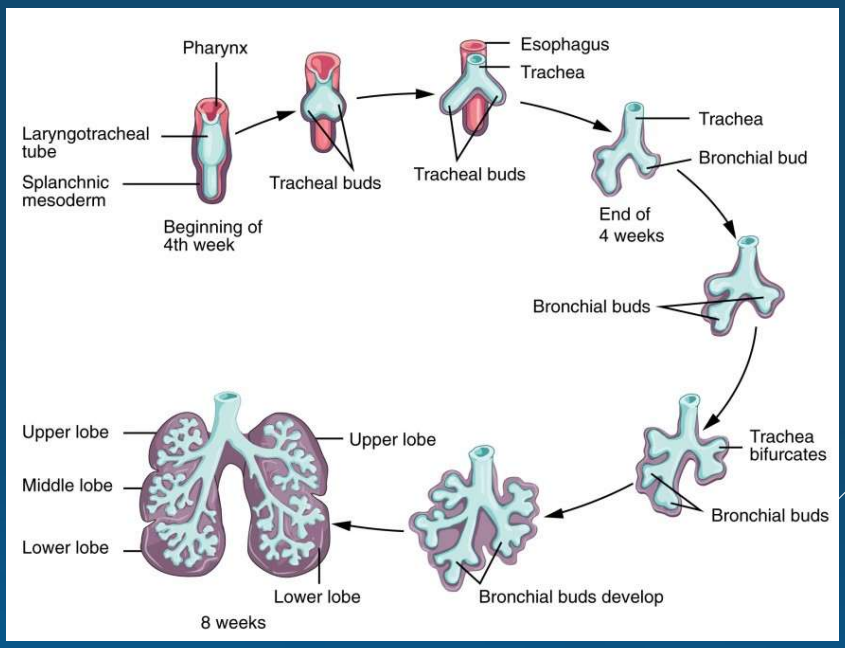

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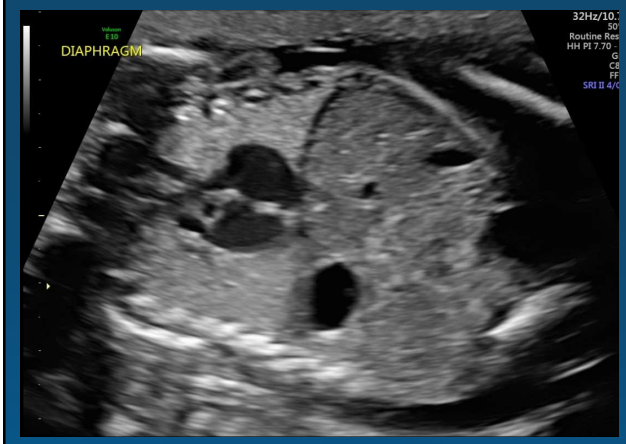
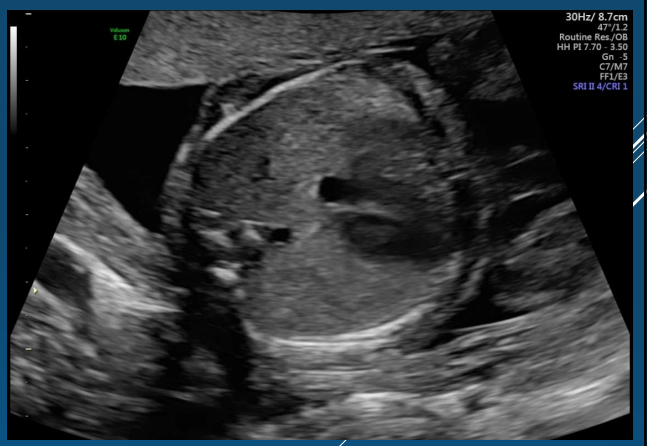
NORMAL SONOGRAPHIC APPEARANCE





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CONGENITAL PULMONARY AIRWAY MALFORMATION (CPAM)

- Previously known as Congenital Cystic Adenomatoid Malformation (CCAM), and before that Cystic Adenomatoid Malformation of the Lung (CAML)
- Account for 1/3 – 1/2 of all lung lesions
- Occurs in 1:11,000 - 1:35,000 pregnancies
- 80-95% are unilateral
- Grow rapidly from 20 – 26 weeks, usually peaking at 25 weeks, and 15% subsequently decrease in size in the late 2nd and 3rd trimesters
- Males are affected slightly more often than females
- Stocker classification has 5 types based on histologic examination
- Sonographically described as either microcystic (< 5mm) or macrocystic (> 5mm)

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FIVE TYPES OF CPAM
STOCKER CLASSIFICATION

- Type 0
  - Trachea/Bronchus
  - Rarest
  - Severe/Lethal
- Type 1
  - Most common (50-70%)
  - Distal bronchus/proximal bronchiole
  - Small number of large cysts (3-10cm)
  - May cause mass effect
  - May lead to hydrops

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FIVE TYPES OF CPAM
STOCKER CLASSIFICATION

- Type 2
  - 15-20%
  - Arises from terminal bronchiole
  - Smaller cysts, 0.5-2 cm
  - Highest incidence of associated anomalies (up to 60%)
- Type 3
  - 5-10%
  - Small cysts, appears solid
- Type 4
  - 5-15%
  - Large cysts, up to 10cm
  - Associated with malignancy

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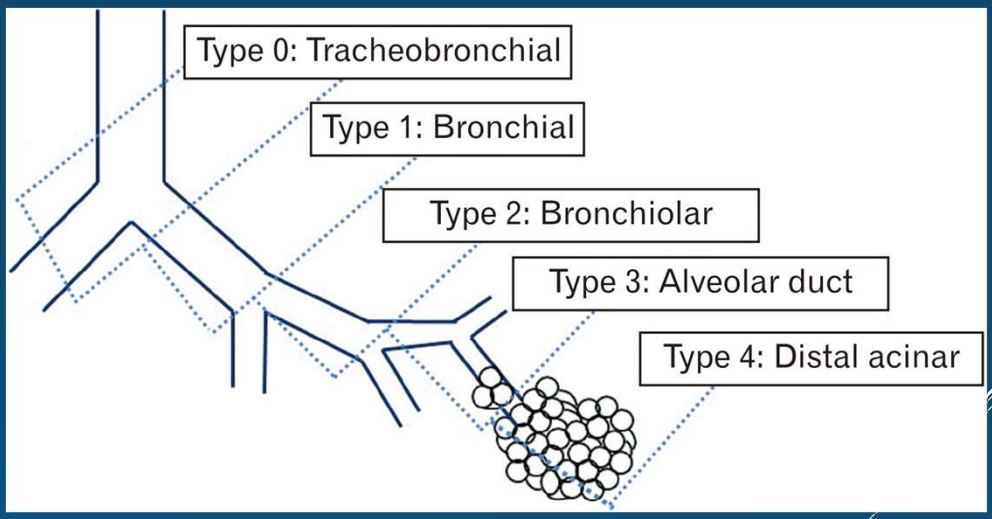

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OTHER CONSIDERATIONS WITH CPAM

- Other findings
  - Mediastinal shift
  - Pleural effusion
  - Hydrops
- Large lesions may increase the risk of pulmonary hypoplasia
- CPAM Volume Ratio (CVR)
  - CPAM volume/Head circumference
  - Evaluation of risk of hydrops and need for intervention

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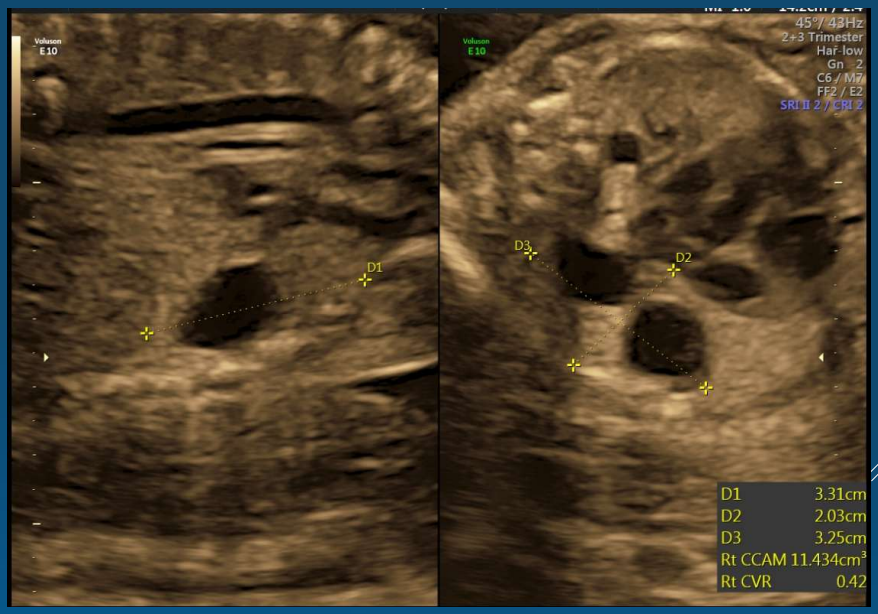

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CPAM VOLUME RATIO (CVR)

- A CVR ≤ 1.6 is at less than 3% risk of developing hydrops
- A CVR > 1.6 predicts increased risk of developing hydrops
- If left untreated, CPAMs with hydrops are almost uniformly fatal

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CASES

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CASE 1

- Diagnosed at 20 week anatomy scan
- Followed weekly
- CVR never exceeded 1.6
- Delivered at 39 weeks 1 day
- No respiratory distress
- Doing well, CT planned for 4-6 months
- Type 1
- Macrocystic

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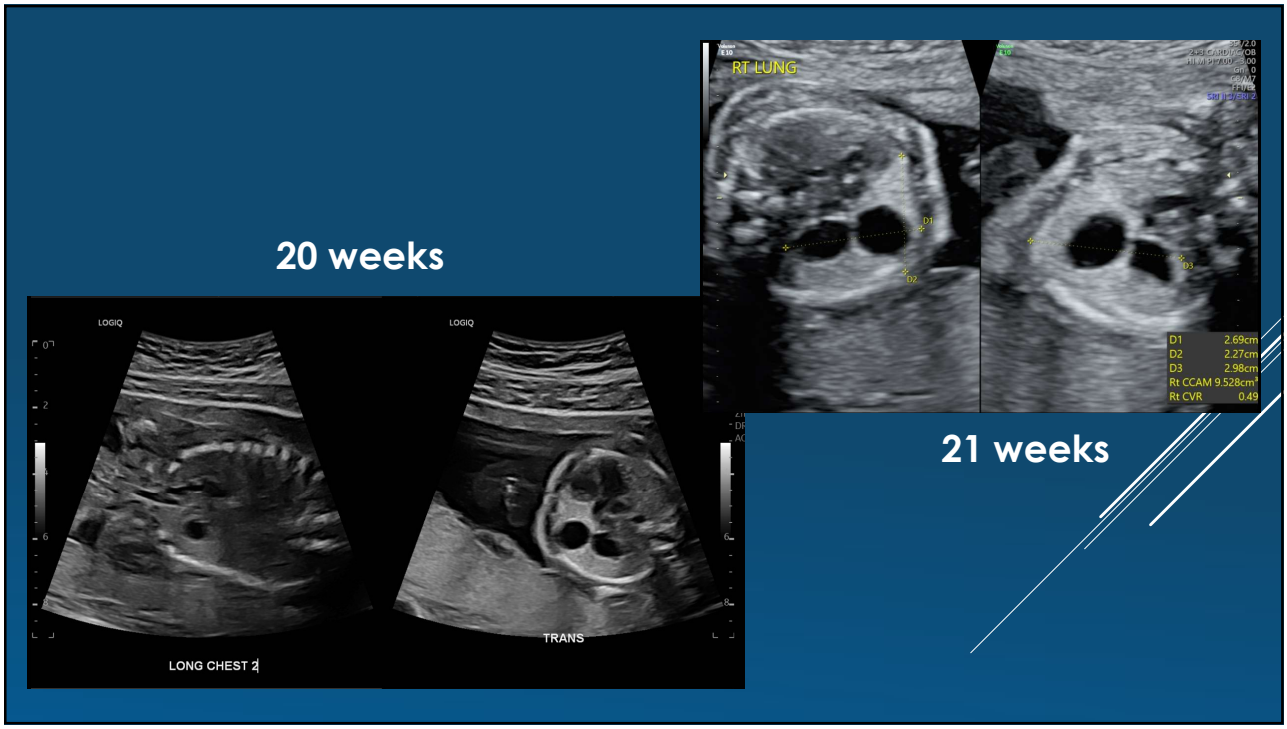

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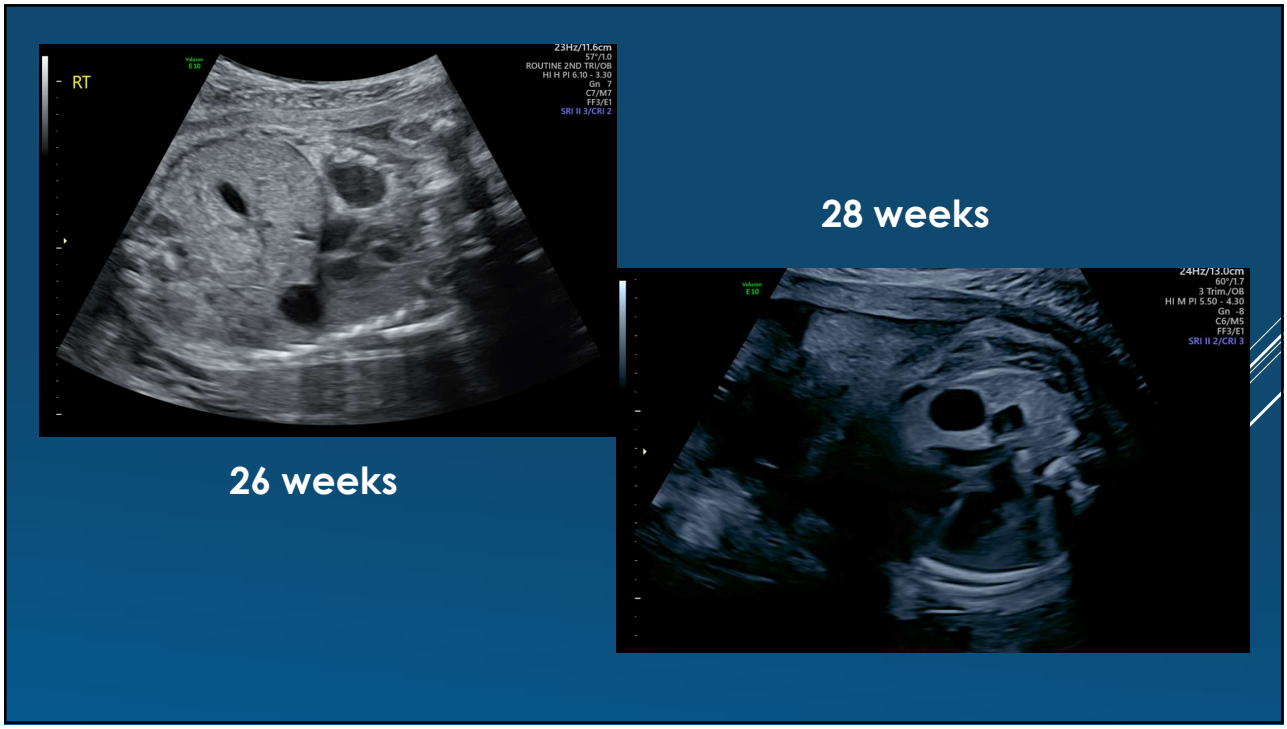

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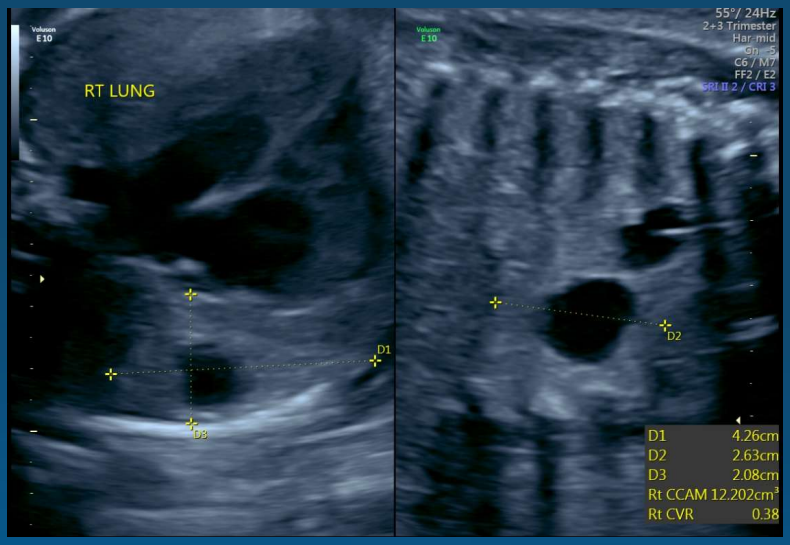

34 weeks

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CASE 2

- Diagnosed at 20 week anatomy scan
- Followed every 1-2 weeks
- CVR never exceeded 1.6
- Delivered by cesarean at 39 weeks
- 3050g female, apgar 8/8
- CT performed at 5 months
- Left lower lobe lobectomy performed at 9 months
- Type 2

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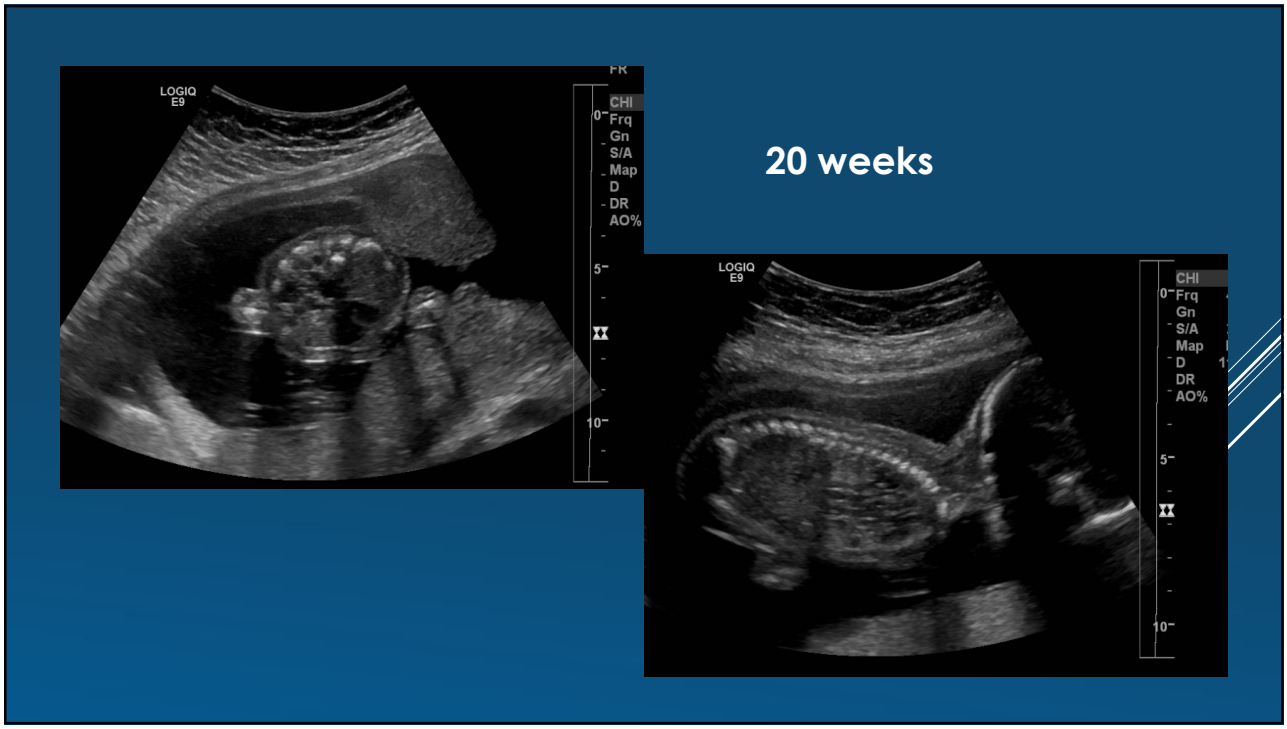

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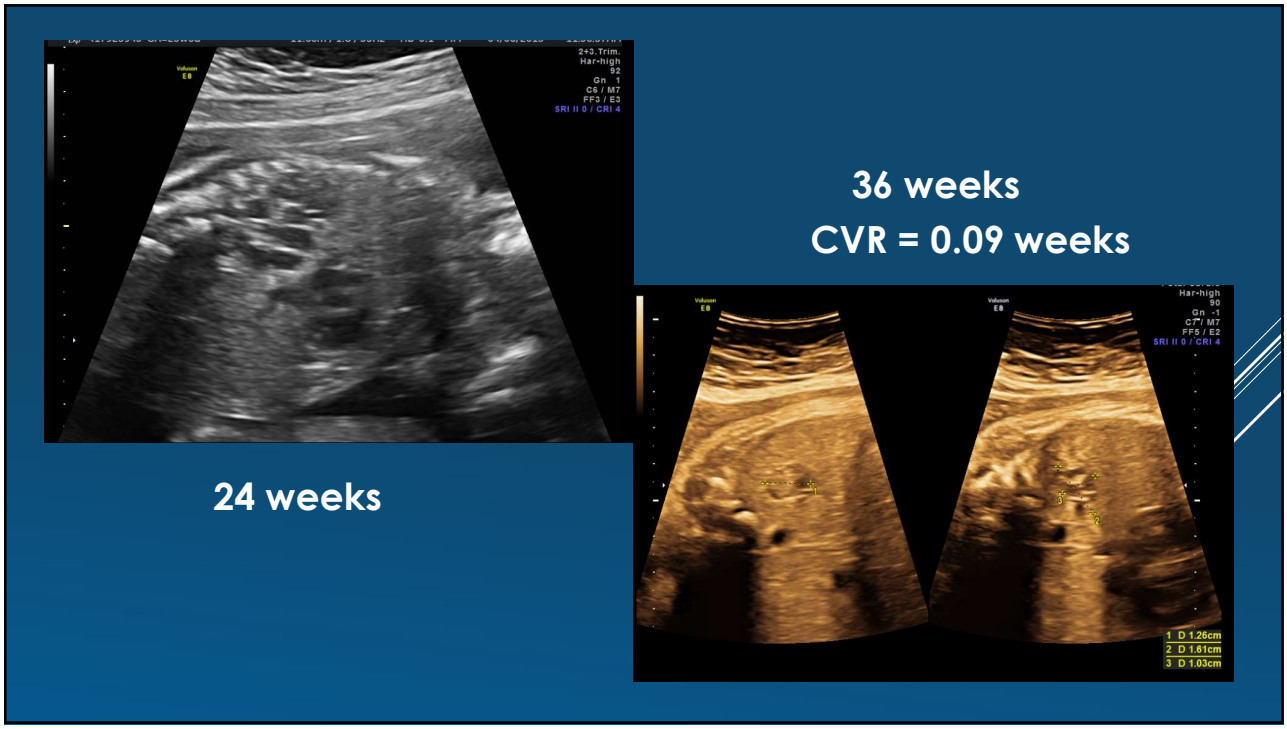

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CASE 3

- Diagnosed at 20 week anatomy scan
- Followed every 1-2 weeks
- CVR ranged between 0.25 and 0.32
- Induction at 39 weeks
- 2977g male, apgar 8/9
- CT performed at 6 months
- Resection performed at 7 months
- Type 3

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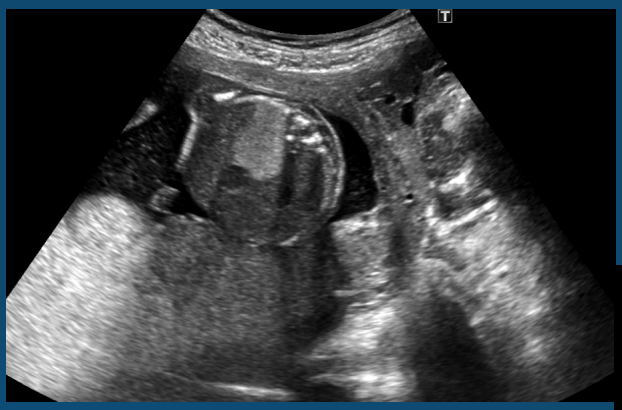
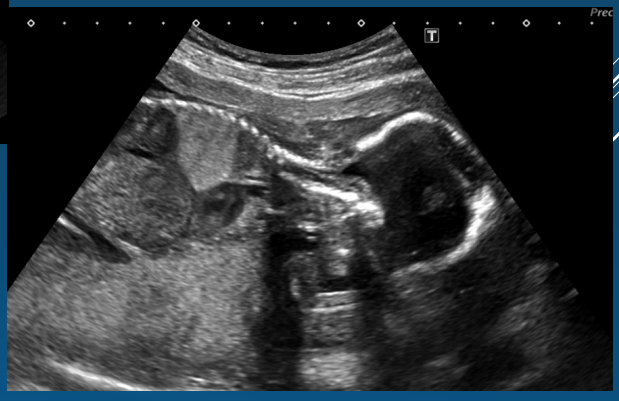


20 weeks



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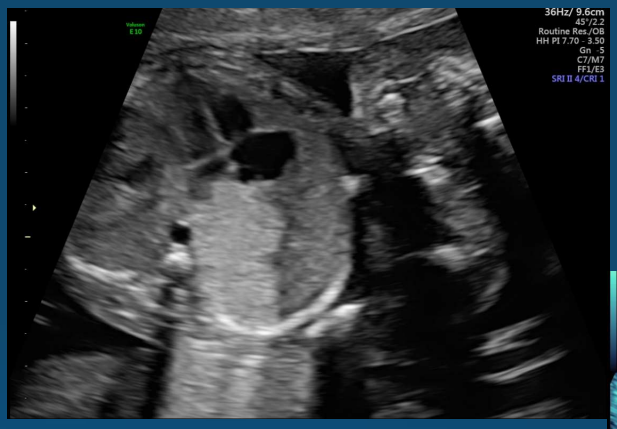
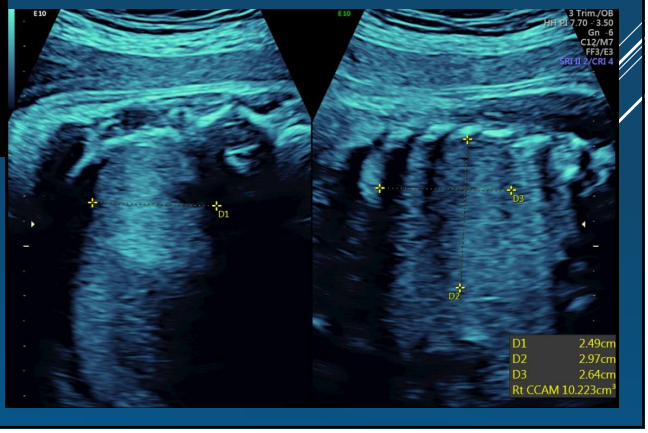


22 weeks

32 weeks



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CASE 4

- Diagnosed at 20 week anatomy scan
- Followed every 1-2 weeks
- CVR ranged between 0.25 and 0.32
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- 2977g male, apgar 8/9
- CT performed at 6 months
- Resection performed at 7 months
- Type 1

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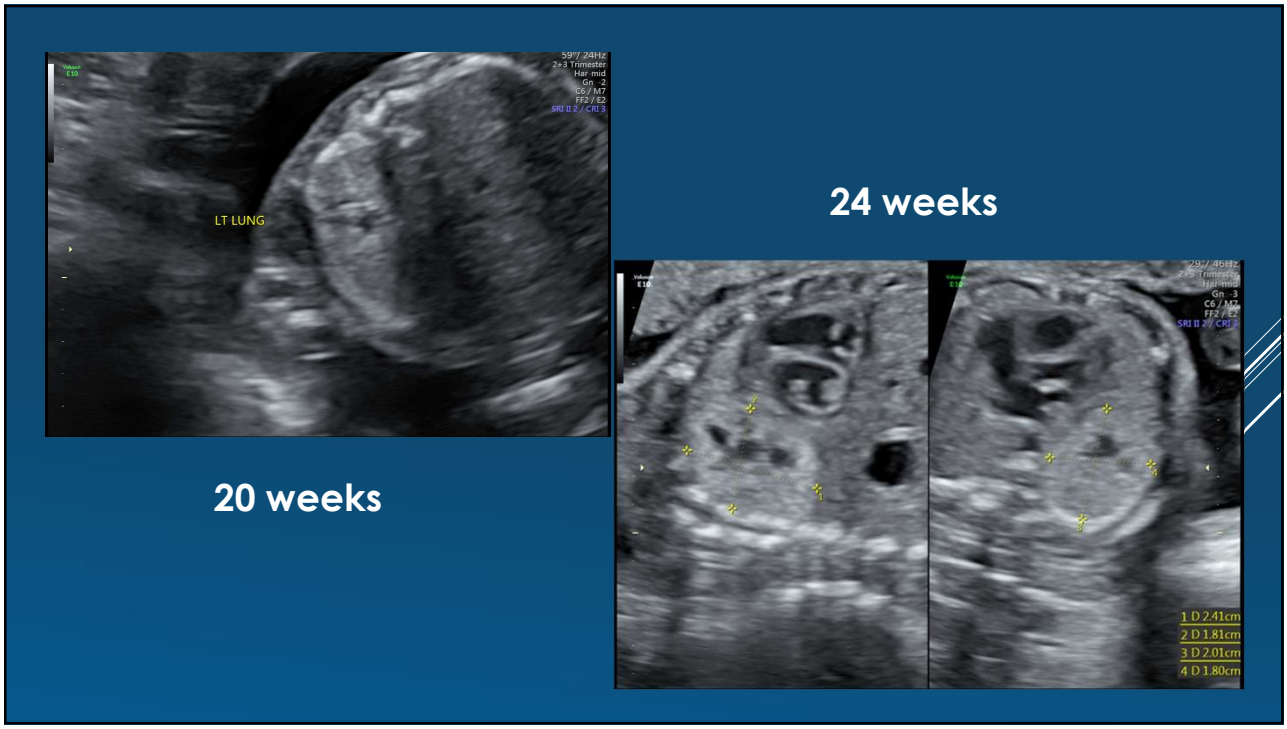

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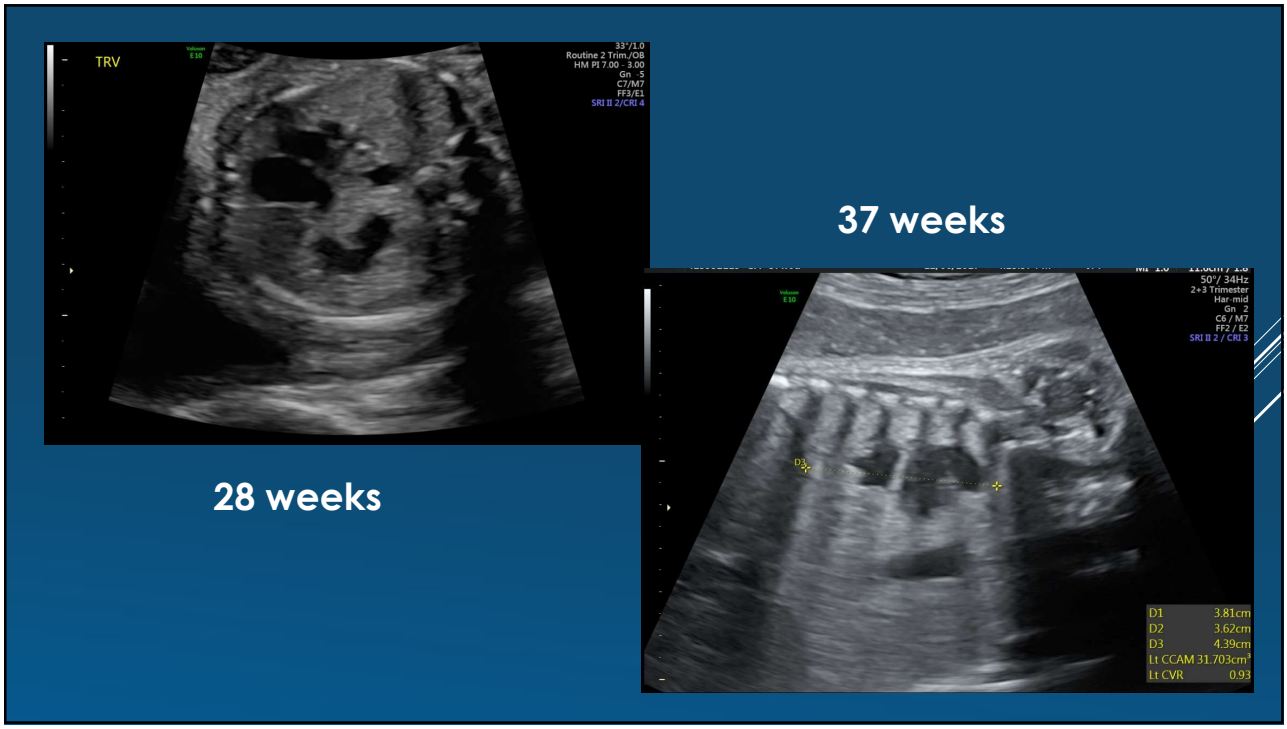

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CASE 5

- CPAM with Congenital Diaphragmatic Hernia (CDH)
- Diagnosed at 20 week anatomy scan
- Fetal MRI left sided CDH containing stomach, small bowel, colon, spleen, and left hepatic lobe as well as a left CPAM
- PPROM at 35 weeks
- Infant was placed on ECMO and had surgery to correct the CDH
- Died at 95 days of life

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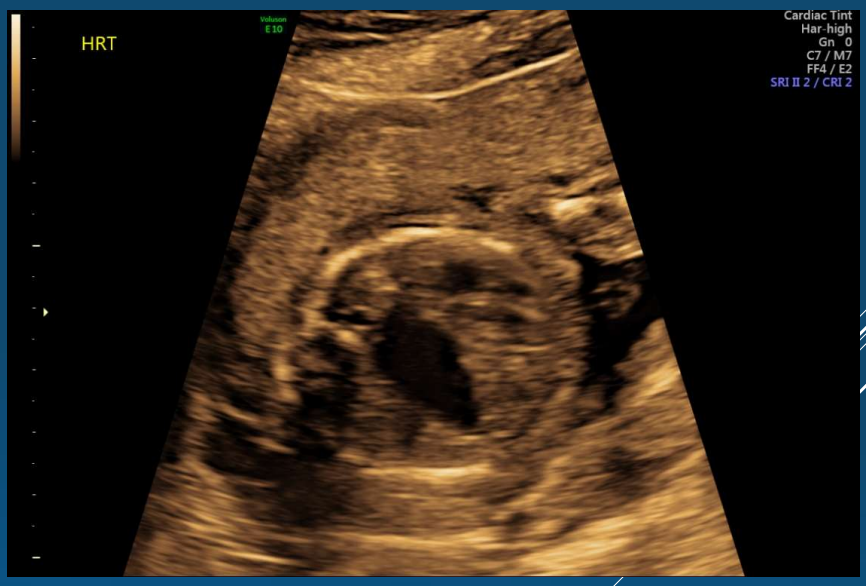
20 weeks



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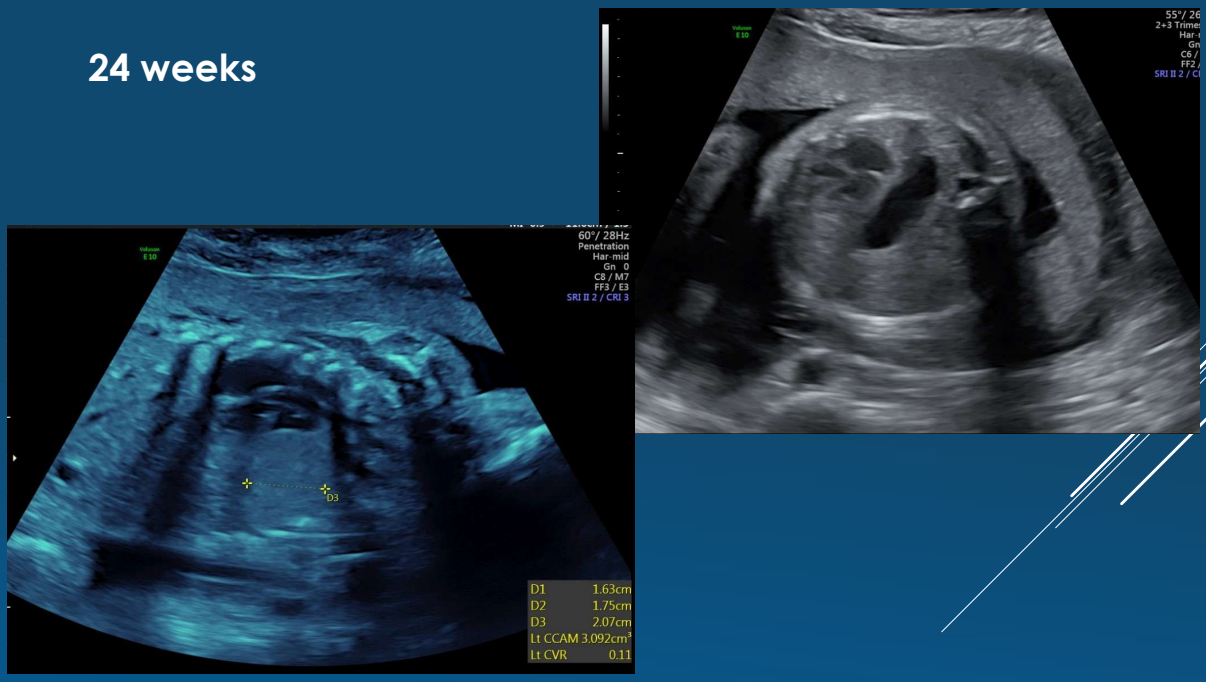
24 weeks



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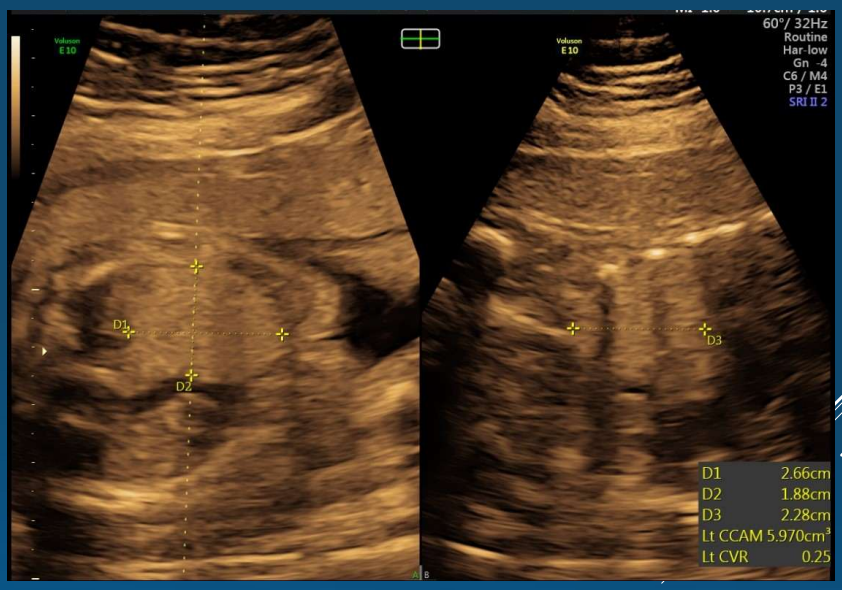
26 weeks



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REVIEW

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QUESTIONS

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REFERENCES

- Puligandla P, Laberge J-M. Congenital Lung Lesions. Clin Perinatol. 2012; 39:331-347
- Hellmund A, Berg C, Geipel A, et al. Prenatal Diagnosis and Evaluation of Sonographic Predictors for Intervention and Adverse Outcome in Congenital Pulmonary Airway Malformation. PLoS ONE 11(3): e0150474.
- Kane S, Da Sliva Costa F, Crameri J, et al. Antenatal assessment and postnatal outcome of fetal lung lesions: a decade's experience at a tertiary referral hospital. Journal of Maternal-fetal and Neonatal Medicine. 2019; 32(5):703-709
- Sfakianaki A, Copel J. Congenital Cystic Lesions of the Lung: Congenital Adenomatoid Malformation and Bronchopulmonary Sequestration. Reviews in Obstetrics and Gynecology. 2012; 5(2):85-93
- Hardee S, Tuzovic L, Silva C, et al. Congenital Cystic Lung Lesions: Evolution From In-utero Detection to Pathology Diagnosis – A Multidisciplinary Approach. Pediatric and Developmental Pathology. 2017; 20(5):403-410
- Sintim-Damoa A, Cohen H. Fetal imaging of congenital lung lesions with postnatal correlation. Pediatric Radiology. 2022; 52:1921-1934
- Hegde B, Tsao K, Hirose S. Management of Congenital Lung Malformations. Clin Perinatol. 2022; 49:907-926
- Zobel M, Gologorsky R, Lee H, Vu L. Congenital lung lesions. Seminars in Pediatric Surgery. 2019; 28(4):150821
- Stocker J, Madewell J, Drake R. Congenital cystic adenomatoid malformation of the lung: Classification and morphologic spectrum. Human Pathology. 1977; 8(2): 155-171