FETAL LUNG LESIONS AND POSTNATAL CORRELATION

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I HAVE NO DISCLOSURES
Lecture Outline

- Normal fetal chest
- Imaging, prognosis, and treatment of fetal chest lesions:
  - Congenital Pulmonary Airway Malformation
  - Bronchopulmonary Sequestration
  - Congenital Lobar Overinflation (Overinflation Syndrome)
- Summary and approach to diagnosis
NORMAL CHEST
Normal Lung Development

- 26 days of gestation to 8 years
- Adequate thoracic space
- Adequate intrapulmonary fluid
- Adequate amniotic fluid
- Fetal breathing
T2W

17 weeks

37 weeks
Bronchopulmonary Malformations

- Parenchymal Lesions
  - CPAM
  - Sequestration
- Tracheal or Bronchial Obstruction
  - Congenital Lobar Overinflation (CLO)
  - Congenital High Airway Obstruction (CHAOS)
Why is prenatal diagnosis important?

- **Mass effect**
  - Hydrops
  - Pulmonary hypoplasia

- **Outcome**
  - Timing of secondary effects
  - Severity of pulmonary hypoplasia

- **Counseling** and possible fetal intervention
CONGENITAL PULMONARY AIRWAY MALFORMATION (CPAM)
CPAM

- Most common congenital bronchopulmonary mass
- ½ of BPMs
- Multiple variable sized cysts
- Lobar or segmental
- Communication with tracheobronchial tree
- Usually unilateral
- Vascularity - PA and PV
Stocker et al. Classification System

- **Type 0** (rare) – Acinar dysgenesis; incompatible with life
- **Type 1** (65%) – 1 or more large (1-10 cm) cysts
- **Type 2** (20%) – Numerous small (0.5 – 1.5 cm) cysts
- **Type 3** (<10%) – Microcysts (appears solid)
- **Type 4** (10%) – Large cysts with mass effect
CPAM - Prognosis

- 40% increase in size during pregnancy
- Most rapid growth during 20-26 weeks
- Low risk of hydrops after 28 weeks
- 15% regress and disappear in 3rd trimester
CPAM - Prognosis

- 9-21% risk of hydrops
- Risk factors
  - Absolute size
  - Rate of growth
  - Associated macroscopic cyst
- CPAM Volume Ratio (CVR)
  - $\geq 1.6 \rightarrow$ increased risk of hydrops
  - $\leq 1.6 \textit{with no dominant cyst} \rightarrow <3\%$ risk of hydrops
CPAM - Treatment

- Hydrops after 32 weeks, delivery is recommended
- Hydrops in the second trimester, piece of fetal lung is removed, and the remaining lung compensates by growing
- Type I CPAM with macrocysts, aspiration of cyst and/or thoracoamniotic shunting
- Maternal steroids
CPAM - Treatment

- Symptomatic infant \(\rightarrow\) Immediate resection
- Asymptomatic infant
  - CXR is usually normal
  - CT may show residual CPAM
  - Surgery advocated for potential infection or malignancy
BRONCHOPULMONARY SEQUESTRATION (BPS)
Bronchopulmonary Sequestration

- 2\textsuperscript{nd} most common BPM
- Commonly hybrid lesions
- Abnormal pulmonary tissue with \textit{no connection} to the normal tracheobronchial tree
- Systemic arterial supply
- 15\% - \textit{Infra-} or subdiaphragmatic
Bronchopulmonary Sequestrations

**Intralobar Sequestration**
- No separate pleural covering
- Systemic arterial supply
- Venous drainage via pulmonary veins

**Extralobar Sequestration**
- Separate pleural covering
- Systemic arterial supply
- Systemic venous drainage through azygous or IVC
- Higher association with congenital abnormalities
  - CDH
  - Eventration
  - Foregut anomalies
Bronchopulmonary Sequestration - Prognosis

- Majority regress during pregnancy
- Rare reports of hydrops
- If hydrops develops after 32 weeks, delivery is recommended
- If hydrops develops before 32 weeks, in utero resection or thoracentesis

Postnatal Issues
- Postnatal contrast CT to help identify feeding vessel
- Infection
- Elective Surgery
CONGENITAL LOBAR OVERINFLATION (OVERINFLATION SYNDROME)
Congenital Lobar Overinflation

- Overinflation of a lung segment
- LUL > RML > RUL
- 29% of BPMs
- Difficult to distinguish from CPAM
  - Blood supply from pulmonary artery
  - Lobar or segmental distribution
- Fetal and postnatal complications are rare
- Most cases are asymptomatic at birth
Approach to Bronchopulmonary Malformations

- Many appear as echogenic masses on US
- Discernable cysts → CPAM
- Color Doppler
  - Pulmonary arterial supply → CPAM
  - Systemic arterial supply → BPS or hybrid
- Hydrops fetalis and pulmonary hypoplasia → CPAM
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References: