American Pediatric Surgical Association

Prenatal Counseling Series

Congenital Pulmonary Airway Malformation

from the Fetal Diagnosis and Treatment Committee of the American Pediatric Surgical Association

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Differential Diagnosis

Differential diagnosis of a prenatally diagnosed cystic lung lesion includes: congenital pulmonary airway malformation (CPAM), bronchopulmonary sequestration (BPS), hybrid lesions, bronchial atresia, congenital diaphragmatic hernia and bronchopulmonary foregut malformations.
How to Measure CVR

The CPAM volume is estimated using the formula for prolate ellipse

\[
\text{CPAM volume} = \text{length} \times \text{height} \times \text{width} \times 0.52
\]

\[
\text{CPAM volume ratio} = \frac{\text{CPAM volume}}{\text{head circumference}}
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Source: Colorado Fetal Care Center
Initial Evaluation

Obstetrical Ultrasound
Fetal echocardiography
Fetal magnetic resonance imaging

Obstetrical Ultrasound
• For anatomy, growth and fluid
• Document whether the lesion is microcystic (multiple cysts, solid <5mm) or macrocystic (>5mm)
• Search for a systemic feeding vessel: BPS/hybrid lesion
• Measure the CPAM Volume Ratio (CVR)
• Mediastinal shift
• Signs of hydrops: pleural fluid, ascites, pericardial fluid, skin edema, placentamegaly

Echo
• Cardiac structure and function
• Vascular supply of the lesion
• Pulmonary veins
• Signs of hydrops

Fetal MRI
• Delineate anatomy
• Vascular supply
Prenatal Counseling

Low-risk lesions (CVR < 1.6)
- Weekly CVR and growth Ultrasounds every four weeks
- Expectant management in low-risk cases without a dominant cyst
- In the presence of a dominant cyst (> 2cm), consider an MRI at 34 weeks GA to evaluate for hyperinflation/mediastinal shift and need for delivery at a tertiary center with ECMO capability

High-risk lesions (CVR > 1.6)
Referral to a fetal center
- Weekly or twice weekly ultrasounds depending on severity
- Betamethasone 12mg IM to be repeated in 24-48 hours
- Consider fetal MRI at 34 weeks
- Hydrops < 32 weeks GA
  - Macro cystic lesion: thoracoamniotic shunt
  - Micro cystic lesion: open fetal resection
- Hydrops > 32 weeks GA
  - EXIT-to-fetal CPAM resection
- Emergent Cesarean section with ECMO standby and postnatal resection
Prenatal Considerations

- Usually diagnosed on a prenatal screening ultrasound
- Prenatal natural history characterized by progressive growth until 26-28 weeks GA
- May regress in up to 50-60% of cases
- CVR at first presentation is prognostic. CVR < 1.6 = good prognosis.
- Pulmonary hypoplasia may occur secondary to mass effect
- Polyhydramnios may occur secondary to esophageal obstruction
- Lesions with a CVR > 1.6 have a 75% risk of hydrops

Postnatal Considerations

- Perinatal air trapping within the cysts may occur, resulting in respiratory distress
- Communication with the airways may result in recurrent infections
- Malignant degeneration to pleuro-pulmonary blastoma (PPB) is reported