American Pediatric Surgical Association

Prenatal Counseling Series

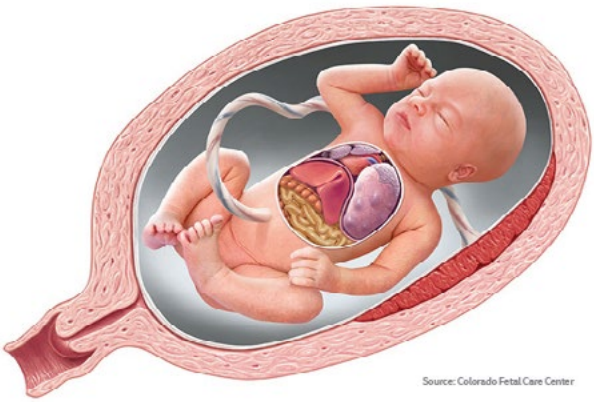
Congenital Pulmonary Airway Malformation

APSA
American Pediatric Surgical Association
Saving Lifetimes™

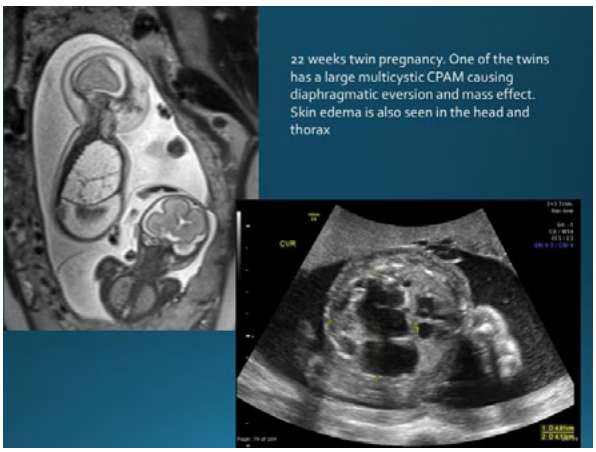
from the
Fetal Diagnosis and Treatment Committee
of the American Pediatric Surgical Association
Editor-in-Chief: Ahmed I. Marwan, MD

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.



Source: Colorado Fetal Care Center



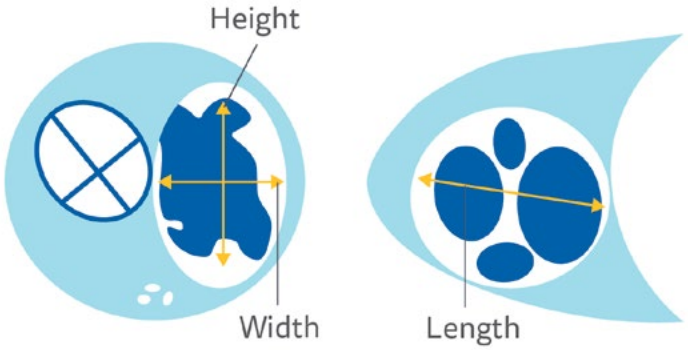
Courtesy of Mariana Meyers, MD, Colorado Fetal Care Center

How to Measure CVR

The CPAM volume is estimated using the formula for prolate ellipse

CPAM volume =
(length x height x width x 0.52)

CPAM volume ratio =
CPAM volume/head circumference



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

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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
    - Macrocystic lesion: thoracoamniotic shunt
    - Microcystic lesion: open fetal resection
- Hydrops > 32 weeks GA
    - EXIT-to-fetal CPAM resection
- Emergent Cesarean section with ECMO standby and postnatal resection

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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