Case Based Fetal Lung Masses
Leann E. Linam, MD

1. Which of the following is true?
   A. Bronchopulmonary malformations are rarely encountered in relation to other perinatal chest masses.
   B. Pleuropulmonary blastoma, congenital pulmonary airway malformation, and pulmonary sequestration are the most common bronchopulmonary malformations.
   C. Bronchopulmonary malformation subtypes are believed to share a common etiology related to in utero airway obstruction, and are now considered a spectrum of disease.
   D. Bronchopulmonary malformations continue to grow throughout gestation.

Correct Answer: C

Reference:

2. Regarding foregut duplication cysts, which of the following is true?
   A. Prenatal US findings are a multilocular, vascular, cystic lesion.
   B. Fetal MRI demonstrates a circumscribed mass with hyperintense T2 signal and homogeneous hypointense T1 signal.
   C. Foregut duplication cysts arise from aberrant ventral budding of the tracheobronchial tree, and the spectrum includes esophageal duplication, bronchogenic, and neurenteric cysts.
   D. Infants are at risk for cyst superinfection, and airway compression is more common in older children.

Correct Answer: B

Reference:

Pulmonary Lymphangiectasia - More Common Than You Think
Dorothy I. Bulas, MD

1. Which of the following is NOT present in primary pulmonary lymphangiectasia prenatally?
   A. Bilateral pleural effusions
   B. Normal fetal echocardiogram
C. “Nutmeg lung”
D. Ascites

Correct Answer: D

Rationale:
Prenatal pleural effusions may be due to a variety of causes. It is important to first exclude the presence of fluid in other compartments, which may indicate hydrops fetalis. There should be no additional findings to suggest hydrops fetalis, including ascites, skin thickening or pericardial effusion.

If the pleural effusions are isolated, potential etiologies include pulmonary lymphangiectasia, congenital lung lesions such as pulmonary sequestration or congenital pulmonary airway malformation, and chromosomal anomalies such as Turner or Down syndrome. Additional imaging findings should help make the diagnosis and guide both prenatal counseling and postnatal management. The “nutmeg lung” appearance on fetal MRI has been shown to be relatively specific for pulmonary lymphangiectasia.

Option A is incorrect. Pulmonary lymphangiectasia is a congenital disease resulting in obstruction and dilation of the pulmonary lymphatic system. The lungs become enlarged and non-compliant, with effusions typically present which can lead to respiratory distress at delivery.

Option B is incorrect. Primary pulmonary lymphangiectasia is of unclear etiology, whereas the secondary form may be due to congenital heart disease causing poor venolymphatic return. A normal fetal echocardiogram in the setting of heterogeneous lung parenchyma and effusions suggests a primary etiology.

Option C is incorrect. The “nutmeg lung” appearance on fetal MRI has been shown to be relatively specific for pulmonary lymphangiectasia manifested by heterogeneous signal throughout both lungs, branching hyperintense structures emanating from the hila of both lungs and pleural effusions.

References:


2. Which of the following is true concerning primary pulmonary lymphangiectasia.
   A. It is a self-limited disease process and prognosis is generally favorable.
   B. On biopsy, both number and size of lymphatic channels are abnormally increased.
   C. Sclerotherapy is the treatment of choice.
   D. Restriction of dietary fats is a medical option for reducing effusions.

Correct Answer: D
Rationale:
Primary pulmonary lymphangiectasis, is often a fatal disease in the neonatal period characterized by marked congenital lymphatic channel dilation without an increased number of ducts. (In contradistinction to diffuse pulmonary lymphangiomatosis (DPL), a rare chronic interstitial lung disease most commonly seen in children characterized by excessive proliferation of dilated, anastamosing pulmonary and extrapulmonary lympatic channels lined by mature endothelium)

Strategies for postnatal management include surgical options such as thoracic duct ligation and medical options aimed at reducing the pulmonary lymph burden, such as drainage of pleural effusions, pleurodesis, and restriction of dietary fats. Recent experimental treatment using ethiodized oil to embolize the patulous pulmonary lymphatics has shown some success.

Option A is incorrect. Primary pulmonary lymphangiectasis, is often a fatal disease in the neonatal period due to respiratory insufficiency.

Option B is incorrect. While the lymphatic channels are dilated there is no increase in number of ducts histologically.

Option C is incorrect. Sclerotherapy is used more typically for lymphatic malformations.

References:


Measuring Lung Volumes: How, Why, When
Amy R. Mehollin-Ray, MD

1. A fetus with a left congenital diaphragmatic hernia has biometric measurements consistent with fetal growth retardation. Which fetal lung volumetric method would be the best for this fetus?
   A. Observed TFLV divided by expected for gestational age
   B. Observed TFLV divided by expected for fetal body volume
   C. Observed LHR divided by expected for gestational age
   D. Observed LHR divided by expected for fetal body volume

Correct Answer: B

Rationale:
Answer: B. Several studies support the use of fetal body volume to normalize measured fetal lung volume. Fetal body volume can be measured reliably at MRI and can be used in the setting of small- or large-for-gestational-age fetuses. However, since measuring fetal body volume is time-consuming, it is typically reserved for outliers, with the gestational age-based expected values being more convenient for normally-grown fetuses. Answer A and C are still valid
representations of pulmonary hypoplasia, but in the setting of fetal growth retardation, the expected values for gestational age may overestimate fetal lung size and result in the appearance of more severe pulmonary hypoplasia. Answer D has not been investigated, so there is no data to support representing LHR as a ratio with fetal body volume.

References:
  iii. Nawapun K et al. Comparison of matching by body volume or gestational age for calculation of observed to expected total lung volume in fetuses with isolated congenital diaphragmatic hernia. Ultrasound Obstet Gynecol 2014; 44: 655-660.

2. The measurement of fetal lung volumes has been validated for predicting the development of chronic lung disease in which one of these fetal anomalies?
   A. Skeletal dysplasia
   B. Cervical teratoma
   C. Giant omphalocele
   D. PPROM

Correct Answer: C

Rationale:
Answer: C. All of these fetal conditions have been studied with regards to the development of pulmonary hypoplasia, but for Answers A, B and D, fetal lung volumes have been used in an attempt to predict mortality. Total fetal lung volumes have only been linked to the development of chronic lung disease for omphalocele patients, with the O/E TFLV less than 50% being associated with chronic lung disease.

References:
Updates on Fetal Chest Surgery
Holly A. Hedrick, MD

1. Fetoscopic endoluminal tracheal occlusion (FETO) is being offered as a part of a clinical research study for congenital diaphragmatic hernia. Which of the following is criteria for FETO inclusion?
   A. O/E LHR <25%
   B. Gestational age greater than 30 weeks
   C. Isolated Right CDH
   D. Infradiaphragmatic liver location

Correct Answer: A

Rationale:
Ultrasound O/E LHR of < 25% is consistent with SEVERE pulmonary hypoplasia and thus is an inclusion criteria.

Other inclusion criteria include Pregnant women age 18 years and older, who are able to consent, Singleton pregnancy, Normal Karyotype, Diagnosis of Isolated Left CDH with liver up, Gestation at enrollment prior to 29 weeks plus 5 days

Exclusion criteria include Maternal contraindication to fetoscopic surgery or severe maternal medical condition in pregnancy, Technical limitations precluding fetoscopic surgery, Rubber latex allergy, Preterm labor, cervix shortened (<15 mm at enrollment or within 24 hours of FETO balloon insertion procedure) or uterine anomaly strongly predisposing to preterm labor, placenta previa, Psychosocial ineligibility, precluding consent, Fetal Diaphragmatic hernia: right-sided or bilateral, major associated anomalies, isolated left-sided with the O/E LHR ≥ 25%, Inability to remain at FETO site during time period of tracheal occlusion, delivery and postnatal care

Option B is incorrect. Enrollment needs to be prior to 29 weeks 5 days gestation

Option C is incorrect. Right and bilateral CDH meet exclusion criteria.

Option D is incorrect. Isolated left CDH with liver up are inclusion criteria.

References:
II. https://www.clinicaltrials.gov/ct2/show/NCT02549820?term=Hedrick&rank=1