HOW TO IMAGE AND DESCRIBE CONGENITAL LUNG MALFORMATIONS

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I have no relevant disclosures
OBJECTIVES

• To present a general classification of the most commonly encountered congenital lung anomalies
• To present a simplified imaging algorithm
• To discuss clinically relevant imaging findings
CONGENITAL LUNG ANOMALIES

• A heterogeneous group of anomalies
• Wide variety of imaging and clinical manifestations
• Annual incidence of 32 to 42/100,000
• Can be detected across the age spectrum
  • From neonate through adulthood
GENERAL CLASSIFICATION SCHEME

- Many published ways of dividing these lesions
  - Some classified on a continuum, e.g. CPAM-sequestration sequence
  - Others based on vascular supply, other associated anomalies, etc.
- At is most basic and broad
  - Classify into mass-like versus non-mass-like lesions
Mass-like lesions

- CPAM
- Sequestration
- Hybrid lesions
- Bronchial Atresia
- Foregut duplication cyst
- Congenital lobar hyperinflation

Non-mass like lesions

- Pulmonary agenesis, aplasia, hypoplasia
- Interruption of a main pulmonary artery
- Anomalous origin of the left pulmonary artery from the right
- TAPVR
- PAPVR
- Pulmonary venous stenosis
- Tracheal bronchus
- Accessory cardiac bronchus
- Scimitar syndrome
- Pulmonary arteriovenous malformations
MASS-LIKE LESIONS

Vascular Lesions

• Congenital pulmonary airway malformations (CPAM)*
• Pulmonary sequestrations
• Hydrid lesions

Non-vascular Lesions

• Bronchial Atresia
• Foregut duplication cysts
• Congenital lobar hyperinflation
NON-MASS-LIKE LESIONS

Vascular Lesions
- Pulmonary agenesis, aplasia, hypoplasia
- Interruption (absence) of a main pulmonary artery
- Anomalous origin of the left pulmonary artery from the right
- TAPVR
- PAPVR
- Pulmonary venous stenosis
- Scimitar syndrome

Non-vascular Lesions
- Tracheal bronchus
- Accessory cardiac bronchus
PRACTICAL IMAGING EVALUATION

- Prenatal
  - Ultrasound
  - Fetal MRI
- Post-natal
  - Chest radiograph
  - Ultrasound
  - CT
  - MR
Prenatal Ultrasound

- Largely performed by our obstetric colleagues
  - However, useful for comparison in fetal MR or in postnatal imaging
- May show mass-lesions and some vascular malformations
- Often cannot distinguish among the mass-like anomalies
  - Even in the presence of cystic lesions can be difficult to distinguish between CLH, CPAM, foregut duplication cysts, and hybrid lesions
PRENATAL MR

- Experience has significantly increased over the last decade
- May be used to further characterize abnormalities seen on prenatal ultrasound
- Useful for assessing significant associated abnormalities such as pulmonary hypoplasia in large CPAM and hydrops
  - Probably where most of the benefit lies
- Of note, most mass-like lesions are T2 hyperintense and often indistinguishable (even if cysts are present)
POST-NATAL CHEST RADIOGRAPH

- Generally, the first line modality after birth
- Confirms the presence of an abnormality
  - Exception - certain non-mass-like vascular lesions
  - Do not be surprised if suspected CPAMS diagnosed prenatally are imperceptible
- Assess for other associated anomalies, e.g. vertebral anomalies in foregut duplication cysts
POST-NATAL ULTRASOUND

- Experience is increasing in usage for CLAs
- May be limited by poor acoustic windows in the older child or lesions surrounded by aerated lungs
- May demonstrate aberrant vascularity in sequestrations
  - In some centers, surgeons will operate on postnatal ultrasound alone
- Our surgeons want either CT or MR before proceeding
POST-NATAL CT/MR

- Both can well demonstrate pure vascular and mass-like lesions
- Both can show aberrant vascularity
- Obviously, MR has no radiation
  - However, patient may need sedation
  - Current sequences demonstrate lung parenchyma poorly in the absence of “fluid/soft tissue-increased” processes
  - Some surgeons uncomfortable with looking at MRI
- CT is faster, more widely available.
  - However, radiation is an issue
- Institutional/radiologist/surgeon preference over whether CT or MR is performed
HOW TO IMAGE

- Prenatal imaging review is key
  - Is this a mass-like or non-mass like lesion?
  - Generally, where is it located?
    - Lung parenchymal
    - Mediastinal
  - What is your best guess/differential diagnosis?
- This determines what you should recommend, i.e. postnatal CT, MR, US
- First step, postnatally is a conventional radiograph
  - Is the lesion still seen? Is it mass-like?
  - Confirmation or further definition of location
  - Are there other lung parenchymal or skeletal anomalies?
TO GIVE CONTRAST OR NOT?

- Lesions that need angiographic imaging
  - Mass lesions associated with aberrant vascularity
    - Sequestrations
    - Hybrid lesions
    - CPAMS (need to differentiate from hybrid lesions)
    - Bronchial atresia (often associated with small CPAMs, etc.)
  - Pure vascular anomalies
    - Interruption of a main pulmonary artery
    - Anomalous origin of the left pulmonary artery from the right
    - TAPVR
    - PAPVR
    - Pulmonary venous stenosis
    - Pulmonary AVMs
LESIONS THAT MAY NOT NEED CONTRAST

- **Congenital lobar hyperinflation**
  - Does not need contrast if the diagnosis is certain
  - Based on temporal evolution on serial postnatal radiographs
  - Still may not be able to tell from CPAM

- **Foregut duplication cysts**
  - Hyperdense cystic contents may be mistaken for enhancement
  - Accurate localization on prior imaging very helpful

- **Tracheal or accessory cardiac bronchi**
  - Often an incidental finding on cross-sectional imaging
  - May need contrast if infected or if an associated mass
HOW TO DESCRIBE/WHAT THE SURGEON/CLINICIAN WANTS TO KNOW

- First and foremost, where is it, i.e. lung parenchymal or mediastinal?
  - Can be difficult in large mass-like lesions
  - Two methods
    - Where is the epicenter?
    - What normal structures are displaced, i.e. fissures, vessels, bronchi?
- Is it confined to one lobe?
  - Many lesion such as CPAMs and CLH can be multilobar
- Is it central or peripheral in the lung parenchyma
- Is there aberrant vascularity?
IMAGING FEATURES/DESCRIPTION

- What does it look like?
  - Often just an exercise in artistic adjectives
  - I steal descriptors from the BIRADS lexicon
    - Size = three dimensions
  - Morphology
    - Shape = oval, circular, irregular
    - Margins = well-defined, partially well-defined, ill-defined, smooth, macrolobulated, wall-thickness (smooth versus irregular)
    - Cystic, solid, or mixed
  - Density = soft tissue, heterogeneous, etc.
  - Associated parenchymal distortion
    - Level and homogeneity of contrast enhancement
  - Obviously, this is adjusted for purely vascular anomalies
ASSOCIATED ABNORMALITIES

- Aberrant vessels
- Other unsuspected congenital anomalies of the lung parenchyma, heart, and mediastinal structures
- Vertebral and skeletal anomalies
- Compressed or absent normal structures such as lung hypoplasia
  - Very important in large lesions as it determines respiratory reserve
LASTLY

- What is it?
- Probably the number one area we can fail
- Doesn’t matter how eloquently we do with everything else
  - If we tell them we don’t know what it is or cannot give a plausible differential, everything prior is suspect (rightly or wrongly)
- Try your best to be definitive with your findings supporting your diagnosis
THANK YOU!

Feel free to contact me with comments/questions

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