PULMONARY LYMPHANGIECTASIA
MORE COMMON THAN YOU THINK!

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Objectives

1. Understand the etiology of primary and secondary lymphangiectasia
2. Review the prenatal findings by US and MRI “nutmeg lung pattern”
3. Discuss how prenatal diagnosis can affect counseling and management
Congenital Pulmonary Lymphangiectasia (CPL)

- Dilatation of lymphatics draining the pulmonary interstitial and subpleural spaces
- Lungs become enlarged and non-compliant
- Effusions
- May lead to respiratory distress at delivery.
Etiology

**Primary** - congenital, assoc genetic syndromes (FOXC2)
- Noonan classified CPL into 3 groups. Primary developmental defect of pulmonary lymphatics (group 3) normal regression of connective tissue elements fails to occur after 16th wk of fetal life
- Often poor prognosis with pleural effusions, non-immune hydrops.

**Secondary** - acquired pulmonary lymphatic or venous obstruction – congenital heart disease causing poor venolymphatic return.
Differential Diagnosis

• Exclude the presence of fluid in other compartments - hydrops fetalis due to other etiologies.

• If effusions confined to lungs
  – congenital lung lesions (pulmonary sequestration, CPAM)
  – chromosomal anomalies (Tri 21, Turner syndrome).
Diagnosis - US

- Small bilateral pleural effusions
- Slightly heterogeneous parenchyma.
28 wk GA
Fetal MRI can provide a more specific dx by detecting a “nutmeg lung pattern“ and excluding other lung masses.

Victora T, Andronikou S Ped Rad 2014

The nutmeg lung pattern = heterogeneous signal with tubular structures radiating peripherally from the hila

8 fetal cases – 2 primary, 6 secondary CHD 1 hydrops
28 wks GA
GA bilateral effusions, radiating tubular lymphatics
small bilateral pleural effusions and heterogeneous signal = nutmeg lung pattern
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Day 2 of life
Day 3 CT - bilateral pleural effusions, thick septa
2 yo  Chronic lung disease, PDA treated medically
Secondary Lymphangiectasia due to Congenital Heart Disease

- Exclude Fetal hydrops due to elevated systemic venous pressures
  - ascites,
  - pleural effusions,
  - pericardial effusions
  - skin edema.
Obstructed pulmonary venous drainage

- Hypoplastic left heart syndrome (HLHS)
- Totally anomalous pulmonary venous drainage (TAPVR)
- Can result in pulmonary lymphangiectasia - high signal branching structures through interstitium.
Antenatal MR imaging of pulmonary lymphangiectasia secondary to hypoplastic left heart syndrome.
Mike Seed Toronto Pad Rad 2009;39:747

Pathology - dilated lymphatics.
CT – thick interlobular septae
Hypoplastic Left Heart with intact septum 31 wks

- 44 fetal MR w HLHS - 4 (9%) nutmeg lung pattern – 3/4 restrictive lesions
- Mortality in nutmeg group 100% by 5 mos.
- 40 w/o nutmeg lung, mortality/orthotopic heart transplant (OHT) 35%.
- 5 restrictive - 3/5 died/had OHT before 5 mos (60% w restriction / non-nutmeg lung).
- Higher incidence of restrictive lesions & mortality/OHT in pts w/nutmeg

CONCLUSION:
- Nutmeg lung in HLHS assoc w/ inc mortality/OHT (100% by 5 mos vs 35% w/ HLHS alone).
- Not all restrictive lesions develop nutmeg lung, outcome not as poor when only restriction present.
- Evaluation for nutmeg lung by MR useful to guide prognostication & counsel parents of fetuses with HLHS.
22 weeks GA. – low signal slightly heterogeneous lungs
Hypoplastic Left Heart  Duplication 12p12.1
Progressed to nutmeg lung pattern by 32 weeks GA.

Hypoplastic Left Heart  Duplication 12P12.1
Tetralogy of Fallot w/ absent pulmonary valves

Asymmetric fluid trapping due to bronchial compression by dilated pulmonary arteries

Clinical utility of fetal MRI in TOF w absent pulmonary valve
Chelliah, Berger, Blask, Donofrio  Circulation 2013 12;127
Tetrology of Fallot with absent pulmonary valve

These early MR findings may help identify cases with severe pulmonary vascular disease that could benefit from early intervention.
Fetal MRI correlates w postnatal CTA assessment of pulm anatomy in TOF w absent pulm valve
Sun HY, Boe J, Rubesova E, Barth RA, Tacy TA. Congenit Heart Dis. 2014;9(4

- Significant pulmonary AA dilatation compressed the tracheobronchial tree, causing fluid trapping prenatally, air trapping/atelectasis after birth.
- Distribution of lobar fluid trapping on MR correlated with air trapping on postnatal CTA.
- Prenatally can predict postnatal pulmonary AA size & visualize airway compression & lung lesions.
TOF dysplastic pulmonary valve, pulmonary stenosis
22q 11 deletion  35 wk  GA
TOF dysplastic pulmonary valve, pulmonary stenosis
TOF dysplastic pulmonary valve, pulmonary stenosis
TOF dysplastic pulmonary valve, pulmonary stenosis
SSFSP – aneurysmal dilatation
Postnatal cases of congenital lobar obstruction have been reported with TOF w/ absent pulmonary valve and aneurysmal dilatation of PA’s.

Associated unilateral left pulmonary hypoplasia if absent or hypoplastic left PA.
Treatment

Strategies for postnatal management

• Surgical options (thoracic duct ligation)
• Medical options – reduce pulmonary lymph burden (drainage of pleural effusions, pleurodesis, restriction of dietary fats).
• Interventional options - ethiodized oil to embolize the patulous pulmonary lymphatics has shown some success.
Conclusions

1. Lymphangiectasia can be diagnosed prenatally.
2. MR is a useful adjunct in identifying “nutmeg lung pattern.”
3. While primary lymphangiectasia is rare, assessing for lymphangiectasia and/or fluid entrapment secondary to CHD prenatally can be useful for counseling and guiding postnatal management.
References