Lymphatic Anatomy and Flow

Lung Lymphatic System

Disclosure

None
Lymphatic Imaging

- T2W MRI
- DCMRL

Pre and Postnatal Lung Lymphatic Dysfunction

- Lung function shortly after birth is dependent on:
  1. Lung surfactant ➔ lung compliance
  2. Lymphatic function to adsorb fluid

- Surfactant deficiency ➔ RDS
- Lymph dysfunction ➔ lung edema ➔ lung compliance ➔ RDS

- CCBE1 = lymphangiogenic factor
- CCBE1-/- mice lack a lymphatic system but have otherwise normal lung anatomy

- Prenatal:
  - Normal CV system
  - Tissue edema
- Postnatal:
  - Death from RDS
  - Unable to inflate the lungs
Patterns of central lymphatic flow in CLFD:

- Abnormal conduction
  - ↑PLPS
- TD outlet obstruction
  - ↑PLPS, ↑Dermal flow
- Functional TD outlet obstruction
  - ↑PLPS, ↑Dermal flow
- Secondary obstruction
  - ↑PLPS, ↑Dermal flow
Congenital Lymphatic Flow Disorder (CLFD)

D-TGA s/p Switch with CLFD

CLFD - External Lymphatic Drainage

Before After 24 hours

TD Outlet Occlusion
PLPS In Patients with SV

- Glenn with chylothorax
- Fontan with chylothorax
- Fontan with PB

PLPS Anatomic Categories

- Type 1
- Type 2
- Type 3
- Type 4
- Type 5

Plastic Bronchitis (PB)

Blue Dye Injection in PB
Contrast Lymphangiogram and SLDE

6 yo HLHS s/p Fontan with severe PB

Pre and Post DCMRL

DCMRL

Outcome

Discharged home 2 weeks post procedure
No complications
Adults with Idiopathic PB

- 76 yo with severe idiopathic PB

Idiopathic PB Versus Neonatal Chylothorax

Chyloptysis

- 57 yo with chyloptysis

Chyloptysis
Overall Survival in Gorham Stout and GLA

GSD patients complicated by effusion 20 94 ± 6
GLA patients complicated by effusion 77 84 ± 5
GSD/GLA patients without effusion 55 100 (N/A)

Chylothorax - GLA

11 yo with GLA and chylothorax
Chylothorax - GLA

Chylothorax - KLA

Lymphatic Embolization

♥ 4 yo with KLA and persistent chylothorax

Lung Disease
Summary

♥ Lymphatic flow disorders are serious, sometimes life threatening diseases

♥ PLPS can lead to abnormal lung mechanics and disease

♥ Imaging is the key first step in diagnosis and to guide therapy

♥ New lymphatic treatments are now available