The Lungs in Congenital Heart Disease

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NO DISCLOSURES OR CONFLICT OF INTEREST
Pulmonary Changes Associated with Congenital Heart Disease

- Abnormality directly due to congenital heart disease
- Abnormality following treatment of the cardiac lesion
- Abnormality associated with congenital heart disease

Will focus on:
• Pulmonary Hypertension
• Lymphatic Abnormality
PULMONARY HYPERTENSION IN CHD
PULMONARY HYPERTENSION

- Common; 4-28% of adult survivors of CHD, many after repair
- Defined as pulmonary arterial pressure > 25mmHg at rest (left atrial pressure 15mmHg or less)
- Most commonly related to left to right shunts
- Increased pulmonary blood flow over time thought to cause endothelial dysfunction and progressive vascular remodeling leading to increased pulmonary vascular resistance
- Usually begins in early childhood and may be irreversible
- Genetic/developmental factors e.g. Down syndrome
Pulmonary H/T associated with left to right shunting

- Large unrepaired left to right shunt with or without flow reversal (Eisenmenger)
- Small shunt, likely incidental, form of idiopathic pulmonary hypertension
- Prior repair (?late) of shunt defect with PHT
PULMONARY HYPERTENSION

Important to evaluate other lesions or contributory factors

- Obesity, smoking, lack of physical activity
- Sleep apnea, chronic hypoxemia
- Pulmonary venous congestion or obstruction
- Chronic pulmonary thromboembolism
- Chronic/Interstitial lung disease
- Pulmonary AVM’s, liver disease, portosystemic shunting
- Fontan circulation- low cardiac output and increased pulmonary vascular resistance without overt pulmonary hypertension, may respond to PHT treatment
Increased incidence of PHT in Down’s

- Left to right shunts, faster progression of pulmonary vascular disease ? genetic
- Obstructive sleep apnea
- Pulmonary disease/hypoplasia

3mo SOB Down's
AVSD
Cardiomegaly, increased vascularity, congestion and hyperinflation
15yo prior ASD repair
severe pulmonary hypertension & sleep apnea

- Enlarged PA and central branches
- Peripheral pruning
- Scattered GGO and LLL hemorrhage
- RVH & decreased function
- Abnormal septal motion
14yo F repaired Scimitar syndrome
Rt pulmonary venous stenosis

PHT

Hypoplastic, congested Rt lung

30% flow to right lung
18mo F - Hypoxemia & Pulmonary Hypertension

Diffuse increased prominence of peripheral PA's

1yr post ASD repair & liver transplant for biliary atresia with heterotaxy/polysplenia
Congenital extrahepatic portosystemic shunt (Abernethy malformation)

MR 8dys, left splenorenal to hemiazygos shunt, no PV

CTA and NM scan age 18mos post OLTX. Still has S-R shunt, small PV

Rt/Lt shunt 39%
PORTOPULMONARY SYNDROME (HPS without liver disease)

• Congenital portosystemic shunt intra or extrahepatic results in portal flow bypassing the liver, lack of “hepatic factor” producing diffuse pulmonary vasodilatation and AV shunting
• May also cause Hepatic Encephalopathy
• Potentially reversible with flow rerouted through the liver - occlusion of the shunt (PV patent) or liver Tx + shunt occlusion (absent PV)
• Extrahepatic PS shunt associated with Polysplenia (Left Isomerism)
Percutaneous closure of shunt

- Decreased vascular prominence
- Improved oxygenation
- Decreased PHT
12YO POLYSPLENIA COMPLEX HEART DISEASE WITH DEXTROCARDIA, SINGLE VENTRICLE AND PERSISTENT HYPOXEMIA
POST LEFT KAWASHIMA (HEMIAZYGOS/LT SVC TO PA) & FONTAN (HV CONDUIT TO PA)
LYMPHATIC ABNORMALITY ASSOCIATED WITH CHD

- Lymphatic congestion
- Lymphangiectasia
- Chylothorax
- Plastic bronchitis
Lymphangiectasia
1/3 of cases associated with CHD (left sided obstruction)

34 week gestation infant with HPLH & restrictive atrial septum
Lymphangiectasia & pleural effusions on Fetal MR

NB HPLH emergency atrial stent
2yo- congenital pulmonary venous stenosis
Lymphatic distention/congestion
3wk old  28wk gestation F
-post successful arterial switch
-persistent chylothoraces
- Pulmonary lymphangiectasia
due to left sided venous thrombosis from CVL and
obstruction of thoracic duct
PLASTIC BRONCHITIS

• Obstructive bronchial casts form in the bronchial tree and are expectorated
• Vary in size from small pieces to complete casts of the lung
• Can cause cough, wheezing, dyspnea, hypoxia, airway obstruction, asphyxiation and death
• Rare in patients w/ severe asthma, cystic fibrosis, sickle cell acute chest syndrome, idiopathic (inflammatory casts – cellular, eosinophils)
• Most common in patients with CHD, Fontan physiology (4-14%) (hypocellular casts, fibrin, lymphatic material)
• Radiologic findings: CXR: Nonspecific – can include atelectasis or infiltrates
• Chest CT scan can demonstrate bronchial casts associated with atelectasis and consolidation
Fontan Procedure and Plastic Bronchitis

- Fontan a two staged repair
  - systemic venous return flows through the lungs w/o ventricular pump
- Increased risk of later developing plastic bronchitis associated with post operative chylothorax, ascites and presence of systemic collaterals to lung
- Hypothesized recently that PB results from protein leak into the airway from prominent peribronchial lymphatics that develop due to lymphatic congestion associated with elevated systemic venous pressure and decreased cardiac output
Treatment of Plastic Bronchitis

- Wide variety of treatments including bronchodilators, inhaled mucolytics, inhaled and systemic corticosteroids, aerosolized fibrinolytics & anticoagulants
- Bronchoscopy/lavage with extraction of obstructing airway casts
- Pulmonary vasodilators
- Optimization, fenestration or takedown of Fontan
- Cardiac transplant
- NEW- lymphatic imaging and embolization of peribronchial lymphatics
T2 MR prominent lumbar, supraclavicular and right peribronchial lymphatics
- Lymphangiogram-Retrograde flow to right hilum from a dilated lymphatic off the TD

- Inguinal lymphangiogram, TD catheterization with selective lymphatic branch embolization
- Excellent result, off Rx x 6mos

MR lymphangiogram (groin node injection of Gd)
Pulmonary Changes Associated with Congenital Heart Disease

- Focused on Pulmonary Hypertension and Pulmonary Lymphatic Abnormality
- Directly caused by CHD, associated with CHD or related to surgical procedure/s
- Complex with numerous interrelated factors
- Chronic and debilitating
- Surgical correction of CHD may not be “complete cure”
- Many procedures are palliative in complex CHD
- Patients should be followed long term
Thank You