Hypoplastic Lung and Preoperative Evaluation for Lung Resection
History

- xx year old female first presented in April 2011 with cough, SOB, left sided pleuritic chest pain
- Symptoms progressively worsening for 5-10 years
- Recurrent respiratory infection with productive sputum, improves with antibiotics
- Known “left lung collapse” since adolescence as per patient
- No history of TB, PE, tachypnea
- PPD negative in Feb, HIV negative per patient
Past Medical History

- Asthma
- “Left lung collapse”
- PSH: None
- Social Hx: No tobacco, etoh, drugs
- Family Hx: no hx of cancer or respiratory problems
- NKDA
- Meds: Albuterol PRN
Physical Exam

- T 97.8, BP 120/68, HR 69, RR 14
- Gen: AAOx3, NAD
- Neck: trachea midline
- CVS: S1S2 normal, no murmurs
- Chest: CTA on right, decreased breath sounds on left
- Abd: soft, NT, NT, normal BS
- Ext: no edema, cyanosis, clubbing
Labs

- PPD: positive 10mm
- Sputum AFB: negative x3
- CBC: 9.88>12.9/41.8<302
- BMP: 136/4.2/100/27/11/0.98/69/9.1
- Coag: 11.7/20.9/1.1
- RA ABG: 7.43/34.1/110/99/23.8/-1.2
<table>
<thead>
<tr>
<th></th>
<th>Pre-Rx</th>
<th></th>
<th>Post-Rx</th>
<th></th>
<th>% Change</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Best</td>
<td>% Predicted</td>
<td>Best</td>
<td>% Predicted</td>
<td></td>
</tr>
<tr>
<td>FVC</td>
<td>1.96</td>
<td>56</td>
<td>2.4</td>
<td>68</td>
<td>22</td>
</tr>
<tr>
<td>FEV1</td>
<td>1.08</td>
<td>35</td>
<td>1.12</td>
<td>37</td>
<td>4</td>
</tr>
<tr>
<td>FEV1/FVC</td>
<td>55</td>
<td></td>
<td>47</td>
<td></td>
<td></td>
</tr>
<tr>
<td>FEF25-75%</td>
<td>0.57</td>
<td>16</td>
<td>0.49</td>
<td>13</td>
<td>-15</td>
</tr>
<tr>
<td>PEF</td>
<td>2.59</td>
<td>36</td>
<td>3.42</td>
<td>47</td>
<td>32</td>
</tr>
<tr>
<td>FET100%</td>
<td>5.83</td>
<td></td>
<td>8.05</td>
<td></td>
<td>38</td>
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## PFT

<table>
<thead>
<tr>
<th>Lung Volumes</th>
<th>Best</th>
<th>% Predicted</th>
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</thead>
<tbody>
<tr>
<td>VC</td>
<td>2.23</td>
<td>57</td>
</tr>
<tr>
<td>TLC</td>
<td>4.58</td>
<td>86</td>
</tr>
<tr>
<td>RV</td>
<td>2.35</td>
<td>163</td>
</tr>
<tr>
<td>RV/TLC</td>
<td></td>
<td>51</td>
</tr>
<tr>
<td>FRC</td>
<td>3.6</td>
<td>129</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Diffusion</th>
<th>Best</th>
<th>% Predicted</th>
</tr>
</thead>
<tbody>
<tr>
<td>DLCO</td>
<td>15.8</td>
<td>61</td>
</tr>
<tr>
<td>DLCO/VA</td>
<td>5.3</td>
<td>110</td>
</tr>
</tbody>
</table>
Gross bronchiectasis of LUL and lingula with completely destroyed and shrunken left lung

No excess secretions, purulent discharge, fungal growth, or blood

BAL culture: pan-sensitive Pseudomonas
  - Negative for malignancy
  - Negative for viral inclusions
  - AFB and GMS stain negative for organisms

Treated with Levofloxacin 2 weeks

Cough, SOB, and chest pain resolved

Pt returned to baseline level of activity
**Operation**

- Bronchoscopy
- Left thoracotomy
- 5th rib resection
- Partially extrapleural pneumonectomy
  - Lung was dissected extrapleurally
  - Hilar structures identified intrapericardially and followed out to the pleural space and then divided
- Pericardial patch
POD 1: Extubated, chest tube removed, diet advanced

POD 3: Transferred to floor

POD 6: Started on zosyn for persistent leukocytosis and OR culture with pseudomonas

POD 7: Tachycardic to 115, SO2 85%
- CTA negative for PE, Transferred to SICU
- Improved with O2 face mask, chest PT, and continued abx
- CXR: RLL opacification
Post op

- **POD 12**: Abdomen distended
  - Disimpacted and enema given
  - CT: cecal volvulus
  - OR for ex lap, right hemicolecctomy

- **POD 25**: Discharged home

- **POD 32**: Seen in clinic, doing well.
Pathology

- Hypoplastic lung with marked cystic bronchiectasis and fibrosis
- Chronic active follicular bronchitis and bronchiolitis
Hypoplastic Lung
Pulmonary hypoplasia is rare in adults, usually diagnosed in childhood. Patients usually die before adulthood. Lung infections and other congenital anomalies may be present. Left side involvement is more common than the right. In utero, gas exchange is performed by the placenta. Substantial abnormalities may be present with minimal symptoms until the neonate is delivered.

Pulmonary Hypoplasia in Adults

Lung Development

- **Static lung expansion**
  - Epithelial cells secrete fluid into the lung lumen
  - Distends future air spaces to a fluid volume that approximates postnatal FRC
  - Inadequate production or excessive drainage leads to pulmonary hypoplasia

- **Dynamic lung expansion**
  - Fetal breathing movements
  - Absent or abnormal breathing leads to pulmonary hypoplasia

Pulmonary agenesis/aplasia is due to arrest of development at the embryonic stage

- Pulmonary agenesis: bronchial tree, pulmonary parenchyma, or pulmonary vasculature does not develop
  - Absence of carina; trachea into single bronchus

- Pulmonary aplasia: there is a rudimentary bronchial pouch with absence of distal lung
  - Secretions can pool in the stump and become infected
  - May involve one lobe or the entire lung

- Associated with other non-pulmonary anomalies

- Bilateral defects are rare and invariably lethal

Pulmonary Hypoplasia

- Pulmonary hypoplasia can occur at any time during gestation
- Hypoplastic lungs are small in volume
- Have decreased numbers of alveoli, bronchioles and arterioles
- Primary pulmonary hypoplasia is rare
- Usually occurs in conjunction with another abnormality (secondary pulmonary hypoplasia)

Causes of Secondary Hypoplasia

- Space occupying lesions
  - Diaphragmatic hernia
  - Massive pleural effusion

- Inadequate thoracic cage
  - Asphyxiating thoracic dystrophy
  - Achondrogenesis

- Oligohydramnios
  - Leakage (PROM)
  - Underproduction (renal dysplasia)

- Inadequate vascular supply
  - PA atresia
  - Hypoplastic right heart
  - Tetralogy of Fallot

- Lack of fetal breathing movements
- Chromosomal abnormalities
  - Trisomy 13 or 18

## TABLE 17-8. Causes of bilateral congenital small lungs

<table>
<thead>
<tr>
<th>System Fault</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lack of space</td>
<td>Abnormal thoracic, abdominal, or amniotic cavity contents (see Table 17-9)</td>
</tr>
<tr>
<td>Abnormal vascular supply</td>
<td>Pulmonary valve or artery stenosis</td>
</tr>
<tr>
<td></td>
<td>Tetralogy of Fallot</td>
</tr>
<tr>
<td>Neuromuscular disease</td>
<td>CNS, anterior horn cell, peripheral nerve, or muscle disease (particularly severe spinal muscular atrophy and myotonic dystrophy inherited from mother) reducing fetal breathing movements</td>
</tr>
</tbody>
</table>

## TABLE 17-9. Congenital small lungs due to extrapulmonary mechanical factors

<table>
<thead>
<tr>
<th>Extrapulmonary Mechanical Factor</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abnormal thoracic contents</td>
<td>Diaphragmatic hernia</td>
</tr>
<tr>
<td></td>
<td>Pleural effusion</td>
</tr>
<tr>
<td></td>
<td>Large CTM</td>
</tr>
<tr>
<td>Thoracic compression from below</td>
<td>Abdominal tumors</td>
</tr>
<tr>
<td></td>
<td>Ascites</td>
</tr>
<tr>
<td>Thoracic compression from the sides</td>
<td>Amniotic bands</td>
</tr>
<tr>
<td></td>
<td>Oligohydramnios (any cause)</td>
</tr>
<tr>
<td></td>
<td>Asphyxiating dystrophy/scoliosis or other chest wall deformity</td>
</tr>
</tbody>
</table>
**Signs and Symptoms**

- Infants generally have respiratory failure in the newborn period
- Reduced lung volumes impair ventilation and lead to hypercarbia
- Decreased surface area for gas leads to hypoxemia
- Decreased cross-sectional area of vasculature makes these infants susceptible to pulmonary hypertension

Treatment

- Supportive
- Outcome depends on severity of hypoplasia and associated anomalies
- Lungs may be extremely difficult to ventilate
- Pneumothorax is common due to high distending pressures
- HFV with low tidal volumes may be effective
- Treat infections with antibiotics

Summary

- Pulmonary hypoplasia is usually a disease of infants
- High mortality
- Usually associated with other anomalies
- Secondary pulmonary hypoplasia is more common than primary
- Treatment is supportive
- Can lead to recurrent infections
Preoperative Evaluation for Lung Resection
Incidence of pulmonary complications is directly related to proximity of procedure to diaphragm

Pulmonary, esophageal and other thoracic procedures are high risk for pulmonary complications

FRC declines by 35% after thoracotomy with lung resection and 30% after upper abdominal surgery

When FRC approaches closing volumes, atelectasis occurs

Evaluation for Lung Resection

- History and physical exam
- Labs: CBC, BMP, LFT, PT/PTT, T&C
- Imaging studies
- Blood gases
- Pulmonary function testing
- Quantitative V/Q scan if needed
- Exercise test if needed
- Echocardiogram/cardiac evaluation in at risk patients
Type of operation and incision have varying effects on pulmonary function.

FRC is important lung volume measurement associated with pulmonary complications.

Reduction in FRC results in premature airway closure and atelectasis.

Timed measurements (e.g. FEV1) have better predictive value for morbidity and mortality.
PFT

- Unlikely to contribute for mediastinoscopy, pleural effusions, pleural biopsy, esophageal surgery with no hx of lung disease

- Appropriate in patients with dyspnea, significant functional limitation, prior pulmonary resection, COPD with change in functional capacity

- Mandatory in patients being considered for pulmonary resection

- Two tests with best predictive value for post op M&M
  - FEV1 and DLCO

Predicting Post op Lung Function

- Simple calculation
  - \( \text{ppo-FEV}_1 = \text{FEV}_1[1 - (\text{number of segments resected} \times 0.0526)] \)
  - Similar for DLCO

- Regional assessment of lung function
  - Quantitative V/Q scan is the current standard
  - Reported as percent function contributed by 6 regions
  - \( \text{ppo value} = \text{baseline value} \times \frac{(100 - \text{percent ventilation or perfusion in the region of planned resection})}{100} \)

Assessment of Functional Capacity

- Lung function and calculation of post op function can reliably identify patients at low risk.

- They do less well at defining high risk patients.

- For refinement of risk, assessment of functional capacity is needed.

Table 4-3. Risk Assessment for Pulmonary Surgery

<table>
<thead>
<tr>
<th>Higher Risk</th>
<th>Lower Risk</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age &gt;70</td>
<td>FEV\textsubscript{1} &gt;2 L for pneumonectomy; &gt;1 L for lobectomy; &gt;0.6 L for segmentectomy</td>
</tr>
<tr>
<td>Higher extent of resection</td>
<td>Predicted postoperative FEV\textsubscript{1} &gt;30–40% of predicted (pneumonectomy &gt; lobectomy &gt; wedge resection)</td>
</tr>
<tr>
<td>Poor exercise performance</td>
<td>Stair climbing &gt;5 flights for pneumonectomy; 3 flights for lobectomy</td>
</tr>
<tr>
<td>Low predicted postoperative FEV\textsubscript{1}</td>
<td>Cycle ergometry &gt;83 W</td>
</tr>
<tr>
<td>Low predicted postoperative DLCO</td>
<td>Predicted postoperative DLCO &gt;40% of predicted</td>
</tr>
<tr>
<td>High P\textsubscript{CO\textsubscript{2}} (controversial)</td>
<td>Maximal oxygen uptake &gt;15–20 mL/kg/min</td>
</tr>
</tbody>
</table>
Performance Tests of Functional Capacity

- Stair climbing
- Incremental cardiopulmonary exercise testing
  - Measures maximal oxygen uptake rate (MVO2)
- Predicted post op exercise capacity (ppo-MVO2)

- There is no consensus to the sequence of testing
- Whether exercise testing or quantitative V/Q scan is done first is a matter of local practice and availability
Assessment of Operability

History
Physical exam
Pulmonary function tests

ppoFEV1% >40
and
ppoDLCO% >40
Resect
pO2 >45
and
pCO2 <60
and
VO2max >10

ppoFEV1% <40
or
ppoDLCO% <40
Arterial blood gas
Exercise test
pO2 <45
or
pCO2 >60
or
VO2max <10

Quantitative perfusion scan
Recalculate predicted values

Average risk:
ppoFEV1% >40
ppoDLCO% >40
pO2 >60
pCO2 <45
VO2max >15

High risk:
ppoFEV1% 20 - 40
ppoDLCO% 20 - 40
pO2 45 - 60
pCO2 45 - 60
VO2max 10 - 15

Prohibitive risk:
ppoFEV1% <20
ppoDLCO% <20
pO2 <45
pCO2 >60
VO2max <10
**Review**

- **FEV1 > 2L**: proceed with pneumonectomy
- **FEV1 > 1L**: proceed with lobectomy
- Need ppo-FEV1 > 0.8 (40% pred.)
- Need ppo-DLCO > 11-12 ml/min/mmHgCO (40% pred.)
- If borderline then get quantitative V/Q scan and recalculate
- Still unsure? Get exercise testing
  - Need ppo-VO2Max > 10 ml/kg/min
- Need ppo-FVC > 1.5L
- No resection if pCO2 > 45 or PO2 < 50
References