www.downstatesurgery.org

Hypoplastic Lung and Preoperative Evaluation for Lung Resection

Christopher Lau

Kings County Hospital Center

SUNY Downstate Department of Surgery

October 13, 2011

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

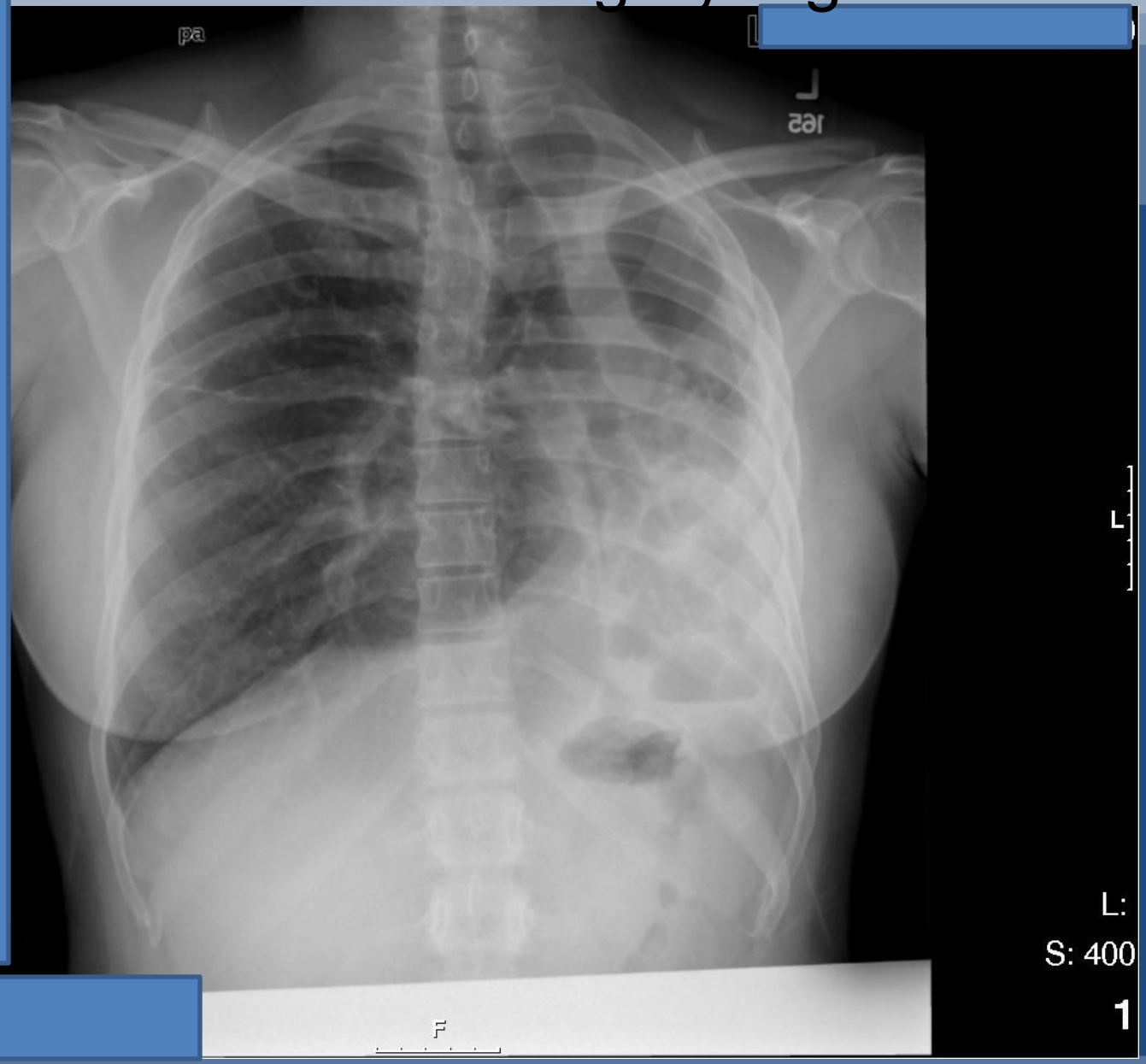
- T 97.8, BP 120/68, HR 69, RR 14
- Gen: AAOx3, NAD
- Neck: trachea midline
- CVS: S1S2 normal, no murmurs
- Chest: CTA on right, decreased breath sounds on left
- Abd: soft, NT, NT, normal BS
- Ext: no edema, cyanosis, clubbing

Labs

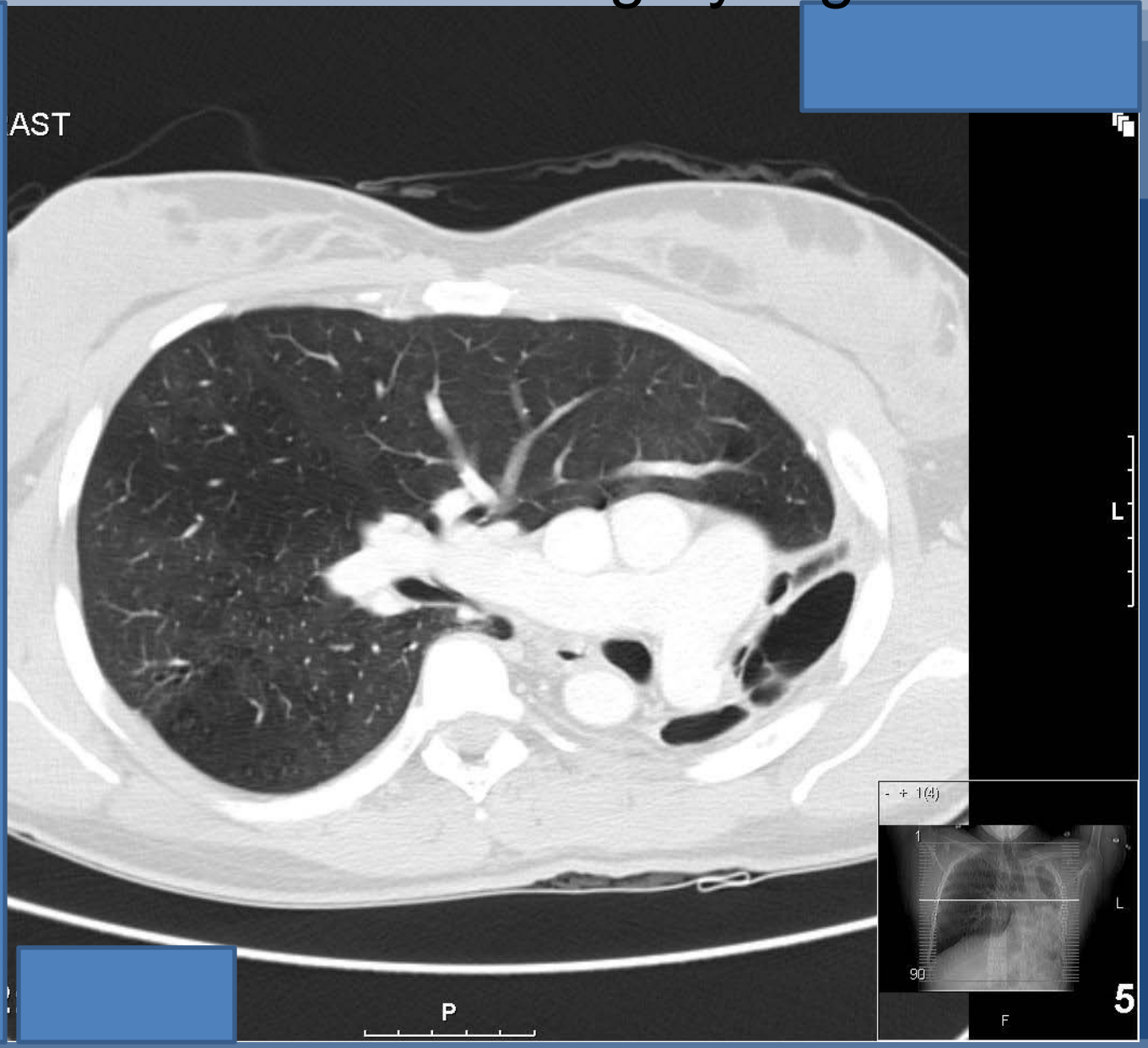
- PPD: positive 10mm
- Sputum AFB: negative x3

- 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

www.downstatesurgery.org



www.downstatesurgery.org



PFT

	Pre-Rx		Post-Rx		
	Best	% Predicted	Best	% Predicted	% Change
FVC	1.96	56	2.4	68	22
FEV1	1.08	35	1.12	37	4
FEV1/FVC	55		47		
FEF25-75%	0.57	16	0.49	13	-15
PEF	2.59	36	3.42	47	32
FET100%	5.83		8.05		38

PFT

Lung Volumes	Best	% Predicted
VC	2.23	57
TLC	4.58	86
RV	2.35	163
RV/TLC		51
FRC	3.6	129

Diffusion	Best	% Predicted
DLCO	15.8	61
DLCO/VA	5.3	110

Bronchoscopy

- 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

Post op

- 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 hemicolectomy
- 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

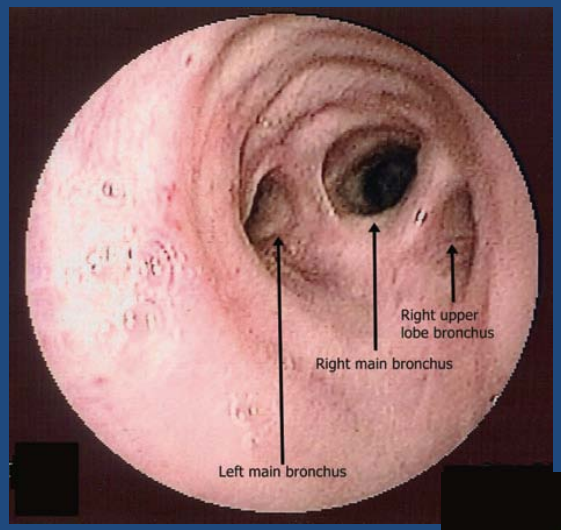
Hypoplastic Lung

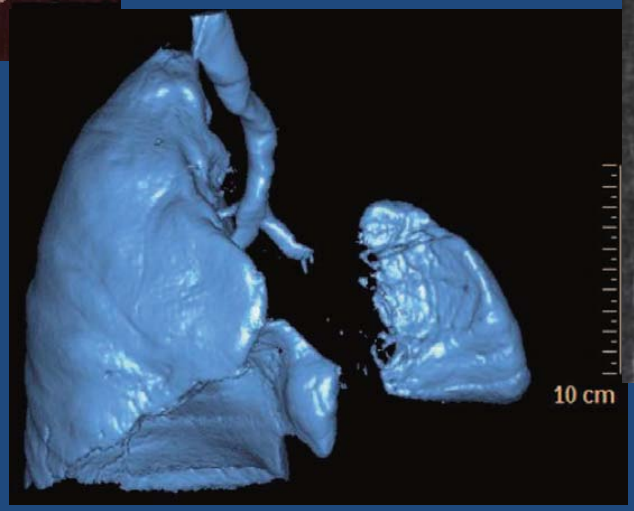
Introduction

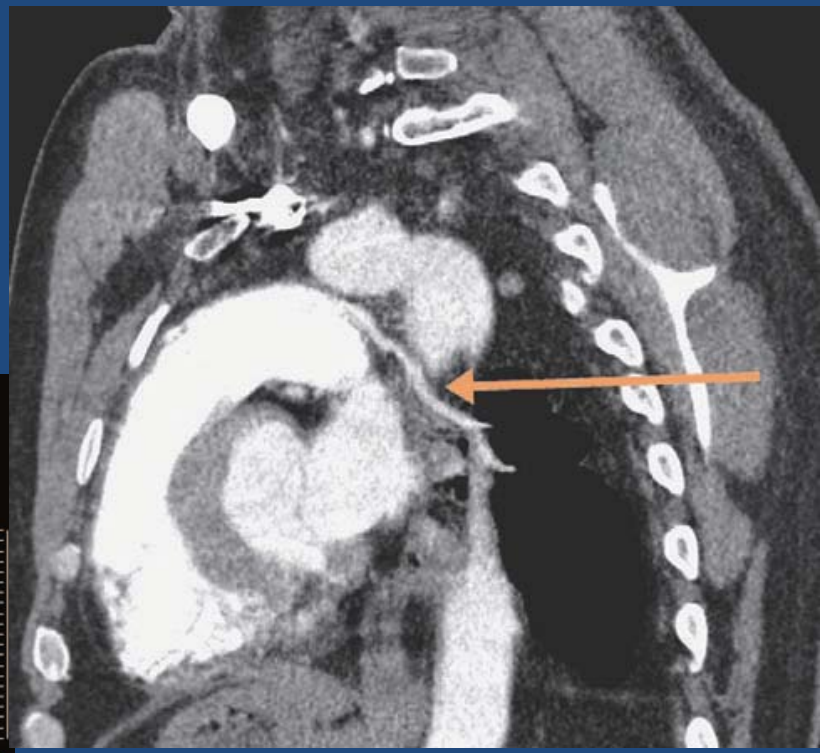
- Pulmonary hypoplasia is rare in adults, usually diagnosed in childhood
- Patients usually die before adulthood
  - Lung infections
  - Other congenital anomalies
- Left side is involved more often than right
- In utero, gas exchange is performed by the placenta
- Substantial abnormalities may be present with minimal symptoms until the neonate is delivered

Mármol E, Martínez S, Baldo X, Rubio M, Sebastián F. [**Pulmonary hypoplasia** in the adult]. Cir Esp. 2010 Oct;88(4):274-6. Epub 2010 Mar 4.
Fuhrman B, Zimmerman J. Pediatric Critical Care, 4th ed. Neonatal Respiratory diseases: pp 596-597, 599-600.

Pulmonary Hypoplasia in Adults







Faruqi S, Varma R, Avery G, Kastelik J. Pulmonary Hypoplasia. Intern Med. 2011;50(10):1129. Epub 2011 May 1.

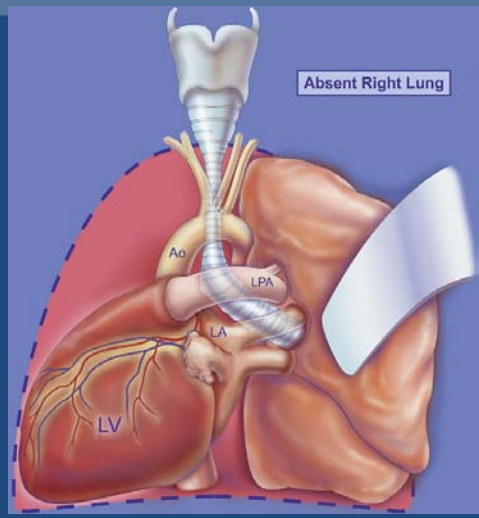
Lung Development

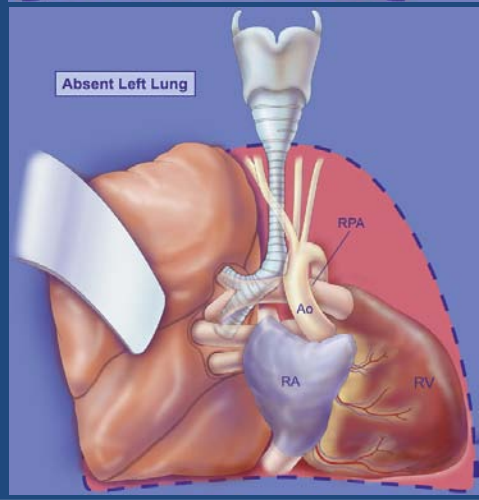
- Static lung expansion
  - Epithelial cells secrete fluid into the lung lumen
  - Distends future air spaces to a fluid volume that approximates postnatal FRC
  - Inadequate production or excessive drainage leads to pulmonary hypoplasia
- Dynamic lung expansion
  - Fetal breathing movements
  - Absent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

Fuhrman B, Zimmerman J. Pediatric Critical Care, 4th ed. Neonatal Respiratory diseases: pp 596-597, 599-600.

Pulmonary Agenesis and Aplasia

- Pulmonary agenesis/aplasia is due to arrest of development at the embyonic 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





Fuhrman B, Zimmerman J. Pediatric Critical Care, 4th ed. Neonatal Respiratory diseases: pp 596-597, 599-600.
Wilmott R, Boat T, Bush A, Chernick V. Kendig's Disorders of the Respiratory Tract in Children, 7th ed. Congenital Lung Disease: p 297.
Backer CL, Kelle AM, Mavroudis C, Rigsby CK, Kaushal S, Holinger LD. Tracheal reconstruction in children with unilateral lung agenesis or severe hypoplasia. Ann Thorac Surg. 2009 Aug;88(2):624-30; discussion 630-1.

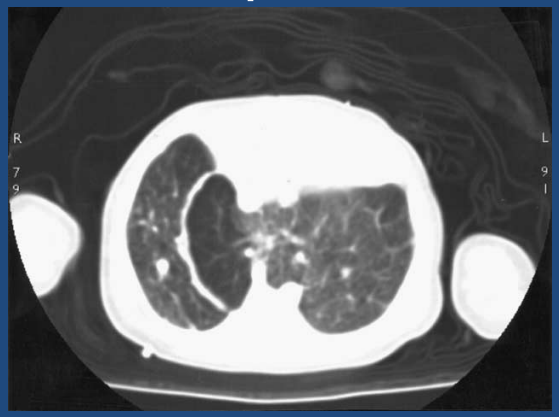
Pulmonary Hypoplasia

- 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)

Fuhrman B, Zimmerman J. Pediatric Critical Care, 4th ed. Neonatal Respiratory diseases: pp 596-597, 599-600.

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

Fuhrman B, Zimmerman J. Pediatric Critical Care, 4th ed. Neonatal Respiratory diseases: pp 596-597, 599-600.
Lutterman J, Jedeikin R, Cleveland DC. Horseshoe lung with left lung hypoplasia and critical pulmonary venous stenosis. Ann Thorac Surg. 2004 Mar;77(3):1085-7.

www.downstatesurgery.org

■ **TABLE 17-8.** Causes of bilateral congenital small lungs

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

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

Extrapulmonary Mechanical Factor	Examples
Abnormal thoracic contents	Diaphragmatic hernia Pleural effusion Large CTM
Thoracic compression from below	Abdominal tumors Ascites
Thoracic compression from the sides	Amniotic bands Oligohydramnios (any cause) Asphyxiating dystrophy/scoliosis or other chest wall deformity

Wilmott R, Boat T, Bush A, Chernick V. Kendig's Disorders of the Respiratory Tract in Children, 7th ed. Congenital Lung Disease: p 297.

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

Fuhrman B, Zimmerman J. Pediatric Critical Care, 4th ed. Neonatal Respiratory diseases: pp 596-597, 599-600.

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

Fuhrman B, Zimmerman J. Pediatric Critical Care, 4th ed. Neonatal Respiratory diseases: pp 596-597, 599-600.

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

www.downstatesurgery.org

Preoperative Evaluation for Lung Resection

General Considerations

- 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

Sugarbaker D, Bueno R, Krasna M, Mentzer S, Zellos L. Adult Chest Surgery, 1st ed.
Shields T, LoCicero J, Reed C, Feins R. General Thoracic Surgery, 7th ed.

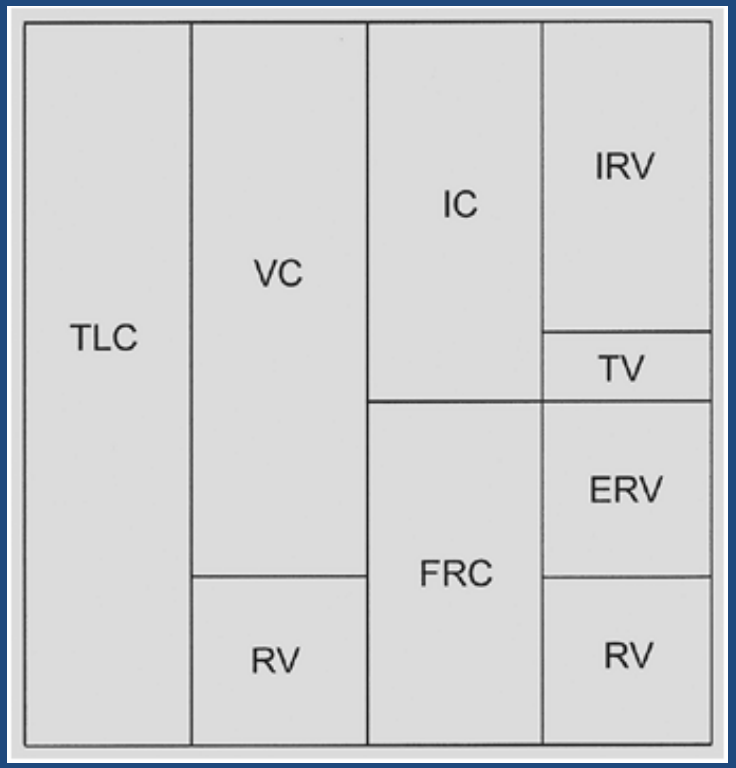
Evaluation for Lung Resection

- 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

Sugarbaker D, Bueno R, Krasna M, Mentzer S, Zellos L. Adult Chest Surgery, 1st ed.
Shields T, LoCicero J, Reed C, Feins R. General Thoracic Surgery, 7th ed.

Lung Volumes

- 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



Sugarbaker D, Bueno R, Krasna M, Mentzer S, Zellos L. Adult Chest Surgery, 1st ed.
Shields T, LoCicero J, Reed C, Feins R. General Thoracic Surgery, 7th ed.

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

Sugarbaker D, Bueno R, Krasna M, Mentzer S, Zellos L. Adult Chest Surgery, 1st ed.
Shields T, LoCicero J, Reed C, Feins R. General Thoracic Surgery, 7th ed.

Predicting Post op Lung Function

- Simple calculation
  - ppo-FEV_1 = FEV_1[1 – (number of segments resected x 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
  - ppo value = baseline value x (100 – percent ventilation or perfusion in the region of planned resection)/100

Sugarbaker D, Bueno R, Krasna M, Mentzer S, Zellos L. Adult Chest Surgery, 1st ed.
Shields T, LoCicero J, Reed C, Feins R. General Thoracic Surgery, 7th ed.

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	
Higher Risk	**Lower Risk**
Age >70	FEV_1 >2 L for pneumonectomy; >1 L for lobectomy; >0.6 L for segmentectomy
Higher extent of resection	Predicted postoperative FEV_1 >30–40% of predicted (pneumonectomy > lobectomy > wedge resection)
Poor exercise performance	Stair climbing >5 flights for pneumonectomy; 3 flights for lobectomy
Low predicted postoperative FEV_1	Cycle ergometry >83 W
Low predicted postoperative D_{LCO}	Predicted postoperative D_{LCO} >40% of predicted
High P_{CO_2} (controversial)	Maximal oxygen uptake >15–20 mL/kg/min

Sugarbaker D, Bueno R, Krasna M, Mentzer S, Zellos L. Adult Chest Surgery, 1st ed.
Shields T, LoCicero J, Reed C, Feins R. General Thoracic Surgery, 7th ed.

Performance Tests of Functional Capacity

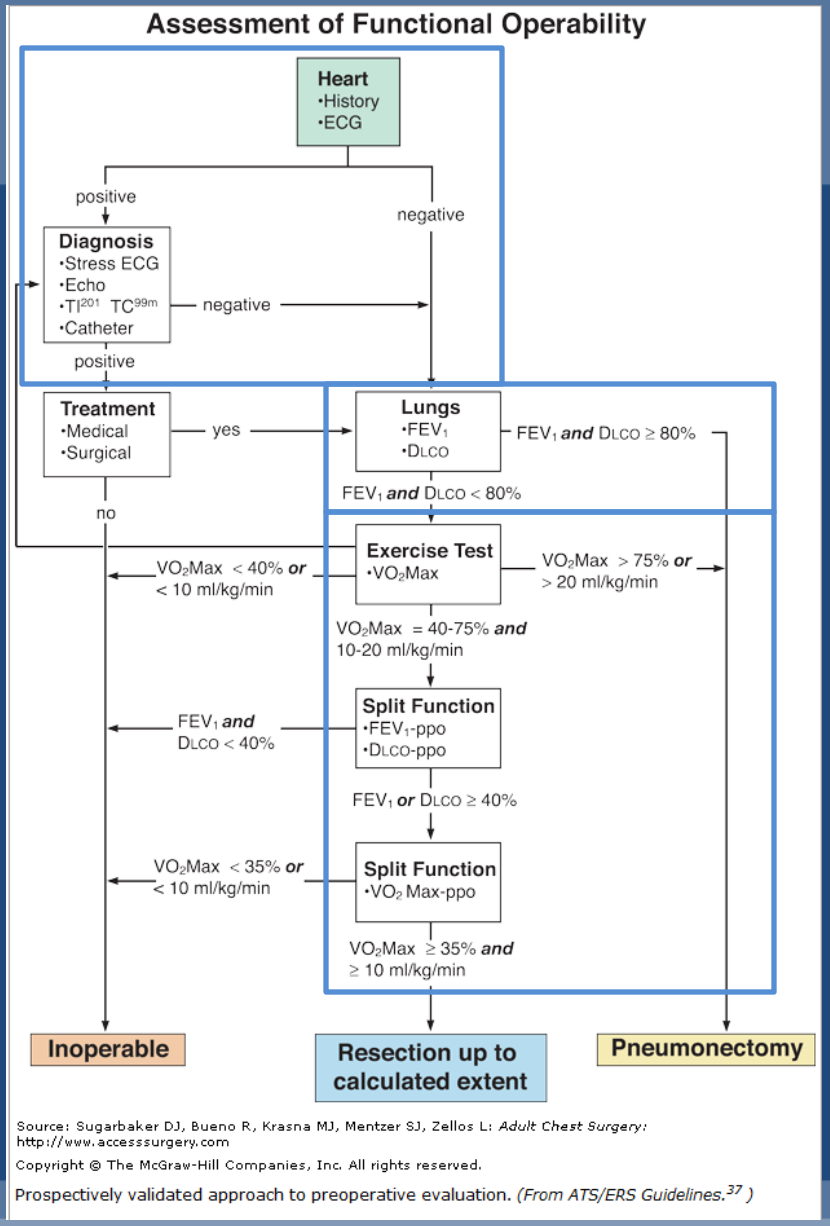
- Stair climbing
- Incremental cardiopulmonary exercise testing
  - Measures maximal oxygen uptake rate (MVO2)
- Predicted post op exercise capacity (ppo-MVO2)

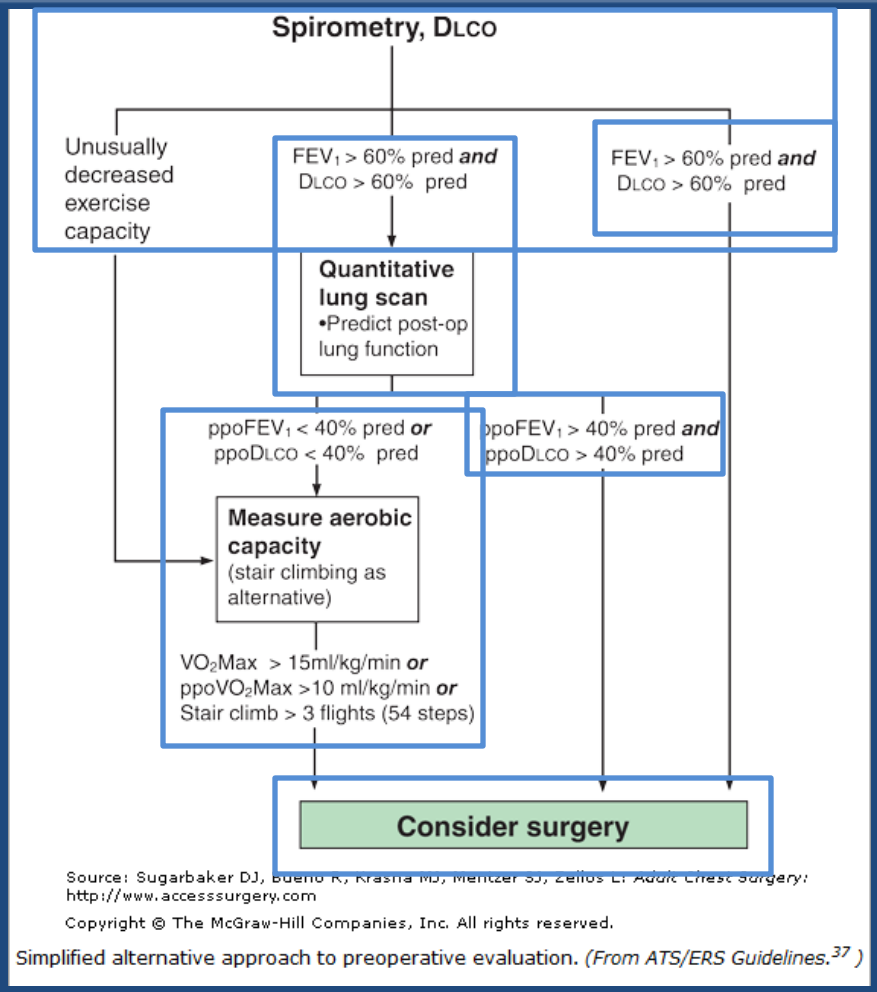
- There is no concensus to the sequence of testing
- Whether exercise testing or quantitative V/Q scan is done first is a matter of local practice and availability

Sugarbaker D, Bueno R, Krasna M, Mentzer S, Zellos L. Adult Chest Surgery, 1st ed.
Shields T, LoCicero J, Reed C, Feins R. General Thoracic Surgery, 7th ed.

www.downstatesurgery.org

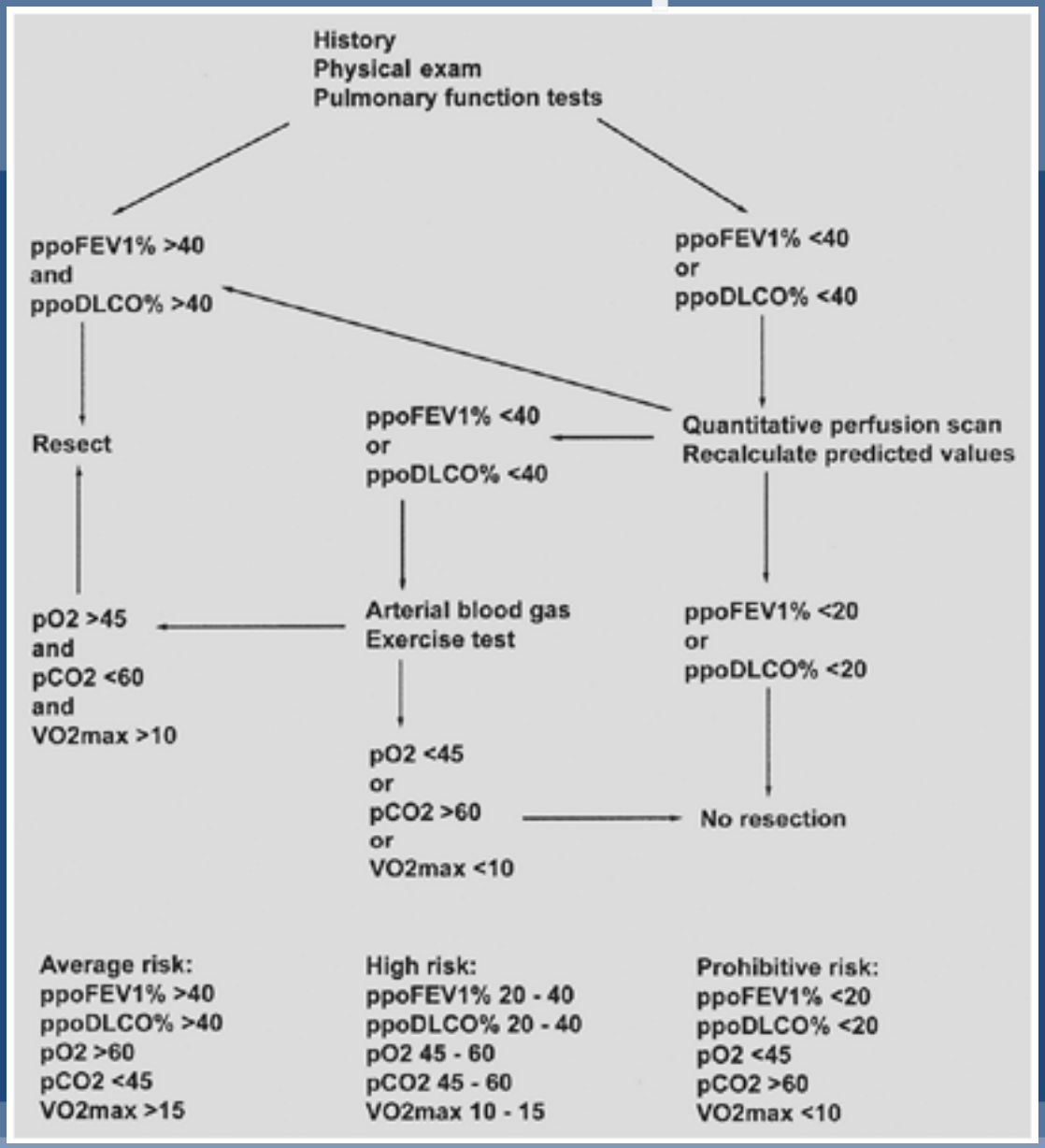
Assessment of Operability





Sugarbaker D, Bueno R, Krasna M, Mentzer S, Zellos L. Adult Chest Surgery, 1st ed.

Assessment of Operability



Shields T, LoCicero J, Reed C, Feins R. General Thoracic Surgery, 7th ed.

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

- Mármol E, Martínez S, Baldo X, Rubio M, Sebastián F. [Pulmonary hypoplasia in the adult]. Cir Esp. 2010 Oct;88(4):274-6. Epub 2010 Mar 4.
- Fuhrman B, Zimmerman J. Pediatric Critical Care, 4th ed. Neonatal Respiratory diseases: pp 596-597, 599-600.
- Wilmott R, Boat T, Bush A, Chernick V. Kendig's Disorders of the Respiratory Tract in Children, 7th ed. Congenital Lung Disease: p 297.
- Lutterman J, Jedeikin R, Cleveland DC. Horseshoe lung with left lung hypoplasia and critical pulmonary venous stenosis. Ann Thorac Surg. 2004 Mar;77(3):1085-7.
- Faruqi S, Varma R, Avery G, Kastelik J. Pulmonary Hypoplasia. Intern Med. 2011;50(10):1129. Epub 2011 May 1.
- Backer CL, Kelle AM, Mavroudis C, Rigsby CK, Kaushal S, Holinger LD. Tracheal reconstruction in children with unilateral lung agenesis or severe hypoplasia. Ann Thorac Surg. 2009 Aug;88(2):624-30; discussion 630-1.
- Sugarbaker D, Bueno R, Krasna M, Mentzer S, Zellos L. Adult Chest Surgery, 1st ed.
- Shields T, LoCicero J, Reed C, Feins R. General Thoracic Surgery, 7th ed.