Restrictive Lung Disease

Restrictive lung diseases are characterized by decreased total lung capacity and decreased lung compliance and may be broadly categorized into intraparenchymal causes, pleural causes and extrapulmonary causes (neuromuscular and extrinsic).

ANESTHETIC CONSIDERATIONS:

- Altered pulmonary mechanics may lead to difficulties with mechanical ventilation: decreased compliance, increased PIP/MAP, decreased FRC
- Rapid desaturation (secondary to decreased TLC, decreased FRC and potentially decreased DLCO)
- Pulmonary hypertension (RV dysfunction and cor pulmonale)
- Comorbid medical conditions (connective tissue disease, autoimmune disease, renal disease, obesity, neuromuscular disease)
- Increased risk of postoperative respiratory failure and need for postoperative disposition arrangement
- Medication considerations (stress dose steroids, immunosuppressants, drug interactions, increased sensitivity to respiratory depressants and NMBAs)

ANESTHETIC GOALS:

- Preoperative optimization (medications, investigations, medications - bronchodilators, steroids, home O2, pulmonary HTN therapies, preoperative consultation with pulmonary if applicable)
- Lung protective ventilatory strategies (Pplat <30cmH20; minimize PIPs; high frequency lower tidal volume ventilation)
- Consider regional techniques for analgesia to minimize respiratory depressants and maximize post operative respiratory function
- Avoid precipitants of pulmonary hypertension (hypoxemia, hypercapnia and acidosis)

HISTORY

- Etiology:
  - acute vs chronic
  - intrinsic pulmonary disease (autoimmune, connective tissue disease, radiation/chemotherapy)
  - extrapulmonary disease (obesity, ribcage [AS, scoliosis], pleural)
- Systemic implications of disease (esp cardiovascular, renal, msk)
- Functional capacity / exercise tolerance
  - ** most important risk factor in determining post operative pulmonary complications
  - Severe exertional dyspnea indicates risk of perioperative respiratory failure
- Symptoms
  - Cough: productive (superimposed URTI? Vs non productive chronic)
  - Dyspnea
  - Hemoptysis
  - Stridor
  - Cor pulmonale – leg swelling and edema, ascites, elevated JVP
- Home O_2
- Nocturnal ventilatory support (CPAP)
- Medications to manage disease especially **immune modulating, steroids**

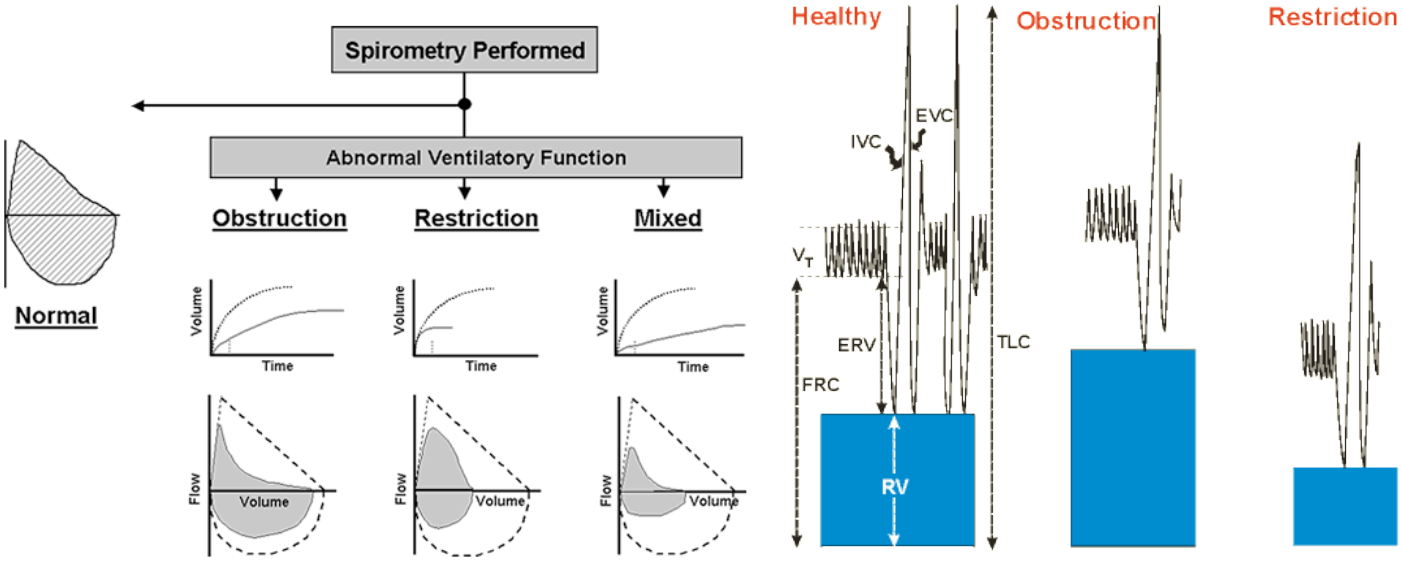
PHYSICAL

- **VITALS + General**
  - Skeletal deformities, kyphoscoliosis, obesity
- **HEENT**
  - tracheal deviation, airway impingement, signs of mediastinal mass
- **RESP**
  - Rapid shallow breathing
  - Number of words b/w breaths (can patient talk in full sentences?)
  - Cyanosis, clubbing late signs (remember – cyanosis associated with $SpO_2 < 80\%$, $PaO_2 < 50$ mmHg)
  - Paradoxical breathing pattern = diaphragmatic dysfunction
  - Asymmetry of lung expansion visible with phrenic nerve dysfunction, hemo- / pneumothorax, masses
  - Breath sounds: coarse, wet crackles with pulmonary edema, fine crackles with interstitial fibrosis
  - Dullness with effusions
- **CVS**
  - PHTN= loud + fixed (stiff RV unable to accommodate any increase in preload with inspiration therefore RV emptying is uniformly prolonged) + widely split S2, systolic murmur of TR (increased with inspiration), murmur of pulmonary regurgitation, JVD with prominent V wave
  - RVH = left parasteral heave due to RV enlargement, S4
  - RV failure = JVD, S3, hepatomegaly, pulsatile liver, ascites, edema
- **MSK**
  - evidence of skeletal and spinal abnormalities, obesity, abdominal distension causing restrictive lung pattern

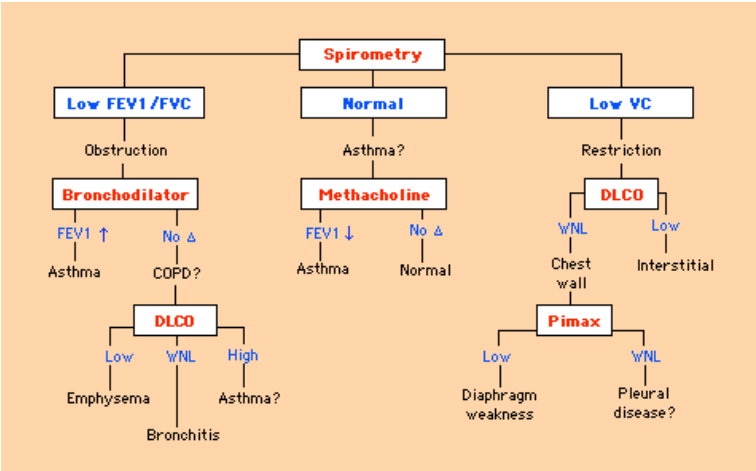
INVESTIGATIONS

- **Labs**
  - CBC, lytes, BUN, Cr, LFTs, INR, PTT (if liver dysfunction)
  - **ABG**
    - with chronic interstitial disease, expect normal to mild hypoxemia
    - arterial CO_2 remains normal until late in the disease
- **Imaging**
  - **CXR** – Interstitial pattern → Ground glass appearance, reticulonodular markings, honeycombing
  - **CT chest –** will give more information if a mass is involved
  - **ECG** – low-voltage QRS + poor R-wave progression due to lung hyperinflation

- Look for signs RA enlargement + RV hypertrophy and strain
  - A tall P wave (P pulmonale) in lead II is diagnostic of RA hypertrophy
  - R/S ratio of greater than 1.0 in lead V1 is found in RV hypertrophy
- **ECHO** – If evidence of PHTN, cor pulmonale, mediastinal mass, or severe lung disease



- **PFTs + Spirometry**
  - Reduced FVC with relatively normal FEV_1
  - Normal airway resistance, so FEV_1/FVC ratio preserved
  - TLC markedly reduced
  - MVV and FEF25-75% usually normal
  - Flow-volume curves are normal in shape, but lung volumes and peak flow rates are lower
  - **DLCO**
    - reduced with intrinsic lung disease (due to V/Q mismatch)
    - usually normal with extrinsic causes of restrictive disease
  - **Markers of severe disease and increased risk of postoperative ventilatory dependence**
    - VC < 15 mL/kg (normal = 70 mL/kg)
    - FVC < 50% or 500 mL



Approach to the patient with dyspnea An efficient stepwise method of determining the cause of chronic dyspnea using pulmonary function tests. WNL = within normal limits; VC = vital capacity; TLC = total lung capacity; DLCO = diffusing capacity; Pimax = peak inspiratory pressure.

OPTIMIZATION

- Pulmonary consult
- Treat infections
- Bronchodilators, steroids (stress dose if needed)
- Treat sleep apnea, nocturnal hypoventilation
- For acute processes:
  - Treat fluid overload with diuretics
  - CHF may require inotropes / afterload reduction
  - Drain pleural effusions
  - Place NG if increased abdominal distension secondary to bowel obstruction
  - Drain ascites

ANESTHETIC OPTIONS

- None
- Regional (may be appropriate for peripheral surgeries and avoidance of general anesthetic/LMA/ETT)
- Neuraxial (may be appropriate for peripheral surgeries and/or for postoperative pain control)
- General
  - Consider pros/cons of minimally invasive surgery
    - Decreased burden of post operative pain and analgesia
    - Increased risk of complications of pulmonary hypertension (pneumoperitoneum, hypercapnia, decreased VT, sympathetic stimulation, etc.)
  - Consider post operative observation or ICU for gradual weaning and extubation

ANESTHETIC SETUP

- **Drugs**
  - Standard emergency drugs +/- epinephrine for resuscitation in case of cardiac arrest
    - Preferential use of vasopressin as vasopressor as no effects on pulmonary vascular resistance
  - PHTN meds if required (NO, inhaled PGE2, sildenafil, milrinone)
  - Stress dose steroids if indicated
- **Equipment**
  - CAS +/- 5-lead
  - Arterial line

MANAGEMENT OF ANESTHESIA

- **Induction**
  - Preoxygenate
  - Rapid desaturation due decreased FRC + potential difficult BMV (decreased compliance of lung or chest wall)
- **Maintenance**
  - Limit FiO_2 to maintain a goal saturation of 88-92% in patients previously exposed to Bleomycin
  - Lung protective ventilation strategies
    - PCV to avoid high peak a/w pressures and minimize risk of pneumothorax (PAP<35 mmHg)
    - tidal volumes of 4-8 mL/kg x RR 14-18/min to minimize barotrauma
      - increased risk of atelectasis
    - Consider permissive hypercapnia
    - PEEP increases FRC but decreases cardiac output and may worsen hypoxemia
    - Consider using ICU ventilator and HFOV for critically ill patients
  - Minimize long-acting CNS depressants in high risk patients
- **Emergence**
  - Usual extubation criteria
    - high risk of postoperative respiratory failure
    - failure to wean from ventilator with severe disease
  - FRC will plummet once PEEP / CPAP removed
    - highest incidence of postop pulmonary complications are with upper abdominal surgery
    - FRC recovers in 3-7 days spontaneously, in 72 hrs with CPAP
  - Incentive spirometry rarely used correctly but consider
  - Encourage early mobilization

DISPOSITION & MONITORING

- Risk for post-op ventilation (especially if VC <15mL/kg)
- Resume pre-operative pulmonary care (e.g. O_2 therapy, nocturnal O_2, meds)
- Analgesia – for upper abdominal / thoracic procedures, epidural indicated to optimize pulmonary mechanics

COMPLICATIONS

- Perioperative hypoxia / hypercapnia
- Barotrauma – PTX

PATHOPHYSIOLOGY

- Typically decreased TLC, FEV_1 may be decreased as well, but FEV_1/FVC ratio will be normal
- **Gas exchange generally well preserved until severe disease is present (especially CO2 exchange)**
  - Arterial hypoxemia results from V/Q mismatch and decreased DLCO
- Caused by acute and chronic intrinsic pulmonary disease as well as extrinsic processes:

1. **EXTRINSIC**: (may be presented with the condition but not specifically told they have restrictive lung disease)
   - **Pleura and mediastinum**
     - Effusion
     - Pleural fibrosis
     - Hemo/ pneumothorax
     - Mediastinal tumors
     - Acute mediastinitis
     - Pneumomediastinum
     - Bronchogenic cysts
   - **Chest wall**

        - Kyphoscoliosis (increased risk of resp failure if VC < 45% predicted and scoliotic angle (cobb angle) greater than 110 degrees)
        - Ankylosing Spondylitis
        - Pectus excavatum / Pectus carinatum
        - Flail chest
    - **Intra-abdominal**
        - Obesity
        - Ascities
    - **Neuromuscular or Diaphragm**
        - Muscular dystrophies
        - Myasthenia gravis
        - Guillain-Barre
        - ALS
        - Quadriplegia
        - Diaphragmatic dysfunction (diaphragmatic paralysis, phrenic nerve dysfunction, eventration of diaphragm from previous trauma)
        - Neoplastic invasion of phrenic nerve

2. **ACUTE INTRINSIC LUNG DISEASE**
    - Due to **increased left heart pressures** (increased hydrostatic pressure) or **increased permeability** of the lung vasculature (increased reflection coefficient – starlings law)
    - **Pulmonary edema**
        - ARDS
        - Neurogenic
        - Cardiogenic
        - Post traumatic (pulmonary contusion)
        - Drug-induced
        - Re-expansion pulmonary edema
        - Negative-pressure pulmonary edema
    - **Pneumonia**
    - **ARDS**
    - **Aspiration pneumonitis**
3. **CHRONIC INTRINSIC LUNG DISEASE**
    - **Idiopathic pulmonary fibrosis**
        - **UIP (usual interstitial pneumonia)**
            - Typical pathologic finding in patients with suspected IPF (in fact, IPF is defined by the American thoracic society as a specific form of chronic fibrosing interstitial pneumonia limited to the lung and associated with the characteristic histologic apperance of UIP on lung biopsy)
            - Significant decrease in lung volumes and diffusion capacity on PFTs
            - Prevalence and incidence increase with increasing age, M>F
            - Most cases are sporadic, familial cases rare
            - CT findings: subpleural (peripheral) and bibasilar reticulonodular opacities with architectural distortion, traction bronchiectasis and honeycombing
            - Prognosis is poor: no real treatment available and most patients die of respiratory failure 5-10 years from diagnosis
        - **DIP (desquamative interstitial pneumonia)**
            - Associated with smokers and relatively uncommon (3-8% of all IPF diagnoses)
            - Lung function studies: mild reduction in lung volumes and moderate decrease in diffusing capacity
            - Less severe radiographic abnormalities vs UIP, and lacks subpleural reticulonodular changes of UIP; ground glass opacities present on HRCT
            - On pathology alveolar septa are lined with thick cuboidal pneumocytes and thickened by inflammatory infiltrate including plasma cells and eosinophils
            - Minimal fibrotic changes and no honeycombing
            - Treatment is with smoking cessation and DIP is steroid responsive
            - 10 year survival is 70-88%
            - may be similar to respiratory bronchiolitis associated interstitial lung disease (RB-ILD); occurs in 4-5th decade of life, M>F (2:1), associated with heavy smokers (generally >30PY hx), generally mild symptoms and mild radiographic changes (interstitial reticular pattern on CXR) and defined pathologically as presence of pigmented macrophages within lumen of bronchial wall; good prognosis overall
        - **NSIP (non specific interstial pneumonia)**
            - Subacute onset of respiratory symptoms, associated fever, lack of male predominance, no clubbing, features suggestive of connective tissue disease
            - Pathological findings lack findings specific for the other IPs (variable histological findings)
            - Can be subdivided according to relative amount of fibrosis (associated with worse outcome)
            - Generally has better prognosis than UIP
            - Treatment: removing inciding exposure, pulse steroids followed by maintenance steroid therapy, if refractory or need for steroid sparing azathioprine or cyclophosphamide, at end stage may be considered for lung transplant
        - **AIP (Acute interstitial pneumonia – Hamman Rich syndrome)**
            - Characterized by acute onset of rapidly progressive respiratory failure and bilateral pulmonary infiltrates (similar to ARDS without systemic or catastrophic inciting component)
            - Poor prognosis
            - Histologically similar to organizing or proliferative stage of diffuse alveolar damage (interstitial fibroblast proliferation, edematous stroma and thickened alveolar septa lined with hyperplastic Type II pneumocytes

        - May be confused with accelerated IPF – but those patients tend to be older, have clubbing and have evidence of UIP on pathology and HRCT
    - **LIP (lymphoproliferative interstitial pneumonia)**
    - **COP (cryptogenic organizing pneumonia – formerly BOOP)**
        - Features more suggestive of pneumonia versus primary airway disorder
        - Can be associated with connective tissue diseases, drug exposure, malignancy and other interstitial pneumonias
        - On pathology: patchy and peribronchial distribution, uniform recent appearance to lung changes without major architectural disruption, foamy macrophages in alveoli, absence of granulomas or vasculitic changes.
        - 5-6th decades of life, M=F, no association with smoking, usually 2-3 month onset
        - leukocytosis, ESR, CRP elevated
        - bilateral diffuse alveolar opacities that may be migrating or recurrent, generally peripheral distribution
        - mild-moderate restrictive defect with severe diffusion defect
        - on BAL: lymphocytic predominance with macrophages, foam cells, mast cells, plasma cells – mixed cellular appearance is characteristic
        - generally steroid and cyclophosmamide responsive; recovery occurs in 2/3 of patients
- **Hypersensitivity pneumonitis**
- **Drug toxicity** (bleomycin, nitrofurantoin, amiodarone can lead to pulmonary fibrosis)
- **Radiation pneumonitis**
- **Autoimmune**
- **Sarcoidosis**

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