PULMONARY DISEASES YOU RARELY HEAR ABOUT (BUT STILL NEED TO TAKE CARE OF)

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The Usual Suspects

Chronic Bronchitis
Emphysema
Pneumonia
Pleural effusions (Cancer, CHF, others)
Lung Cancer

The Unusual Suspects

- Chronic Bronchiectasis
- Cystic Fibrosis
- Lymphangioleiomyomatosis (LAMM)
- Idiopathic Pulmonary Fibrosis
- Pulmonary Hypertension (ES Lung disease, CHF, OSA, Idiopathic, Many other causes)
- Restrictive Lung Diseases (Chest wall, diaphragm, obesity)
- Respiratory Failure (ALS, CVA, Head Trauma)
The Common Final Pathway
- Shortness of Breath
- Cough/Sputum
- Chest discomfort
- Wheezing
- Peripheral edema/ascites
- Orthopnea
- Hemoptysis
- Fatigue/Low Energy

Bronchiectasis
- Widening of the airways usually due to destruction by infection, often in childhood
- Much less common with the antibiotic era
- Daily cough & purulent sputum
- Detected by HRCT Scanning
- End stage patients have cough, sputum, hemoptysis, dyspnea, mucus plugging, wheezing, Chest pain

Causes of Bronchiectasis
- Cystic fibrosis
- Airway obstruction (foreign body aspiration)
- Defective host defenses, mucociliary transport
- Allergic Bronchopulmonary aspergillosis
- Cigarette smoking
Sputum culture, rotating antibiotics
Mucolytics, Guafenesin, Mucomyst
Mechanical means: chest percussion, postural drainage, Theravest, Intermittent Percussive Ventilation, In-exsufflator, Flutter valves
Lung resection if localized

Treatment of Bronchiectasis

Most common life-shortening autosomal recessive disease in Caucasian, 1 in 2000-3000 live births
Median survival 37 years
Findings: persistent pulmonary infections, pancreatic insufficiency, chronic sinusitis, bronchiectasis, elev sweat chloride, ABPA, cough, wheezing, dyspnea, air trapping, digital clubbing, Pseudomonas infection

Cystic Fibrosis
Treatment of CF

- Antibiotics based on culture
- Mucolytics
- Mechanical secretion removal techniques
- Mechanical ventilation or BiPAP for respiratory failure
- Lung Transplantation
- Nutritional supplementation, pancreatic enzymes
- Genetic counselling (although males often sterile)

Lymphangioleiomyomatosis (LAMM)

- Rare disease affecting mostly women of childbearing age
- Related to tuberous sclerosis
- A “benign” tumor of the lung that progresses and causes respiratory failure
- Produces cyst formation and multifocal nodules of immature smooth muscle and epithelioid cells (LAM cells)
- Complications include pneumothorax, hemoptysis, chylothorax,
Treatment of LAMM
- Organ transplant anti-rejection drugs: Sirolimus, everolimus
- Supplemental O2, bronchodilators, pulmonary rehab
- Avoid estrogens
- Possibly progesterone, Tamoxifen, Lupron
- Lung transplantation

Idiopathic Pulmonary Fibrosis
- Chronic, progressive, fibrotic disorder
- Mostly adults > 40 y.o.
- No specific known cause
- Associated factors: smoking, environmental pollutants, chronic microaspiration
- Possible genetic predisposition
- Inflammation may be inciting stimulus, or aberrant epithelial cell & fibroblast responses to injury
- End result is irreversible lung scarring (fibrosis)
Dyspnea on exertion or at rest
Non-productive cough
Fine rales on chest exam
Digital clubbing

No sufficient evidence anything works
Steroids often tried but little evidence to support
Supportive care: influenza & pneumococcal vaccination, pulmonary rehab, oxygen
Pirfenidone or nintedanib may be available in mild-moderate IPF
Sildenafil may help in those with Pulm HTN
GERD prophylaxis
Lung Transplantation
Pulmonary Hypertension (PAH)

Usually the end result of chronic lung diseases, heart failure, valvular disease, but can be idiopathic, due to genetic defects, drugs, toxins, connective tissue diseases, HIV/AIDS, portal HTN, congenital HD, schistosomiasis

Also, Thromboembolic disease, ILD, Left heart disease, OSA & other sleep disorders

Involves all ages, racial groups, genders

Pathology shows proliferative vasculopathy of the muscular pulmonary arterioles

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PAH Findings

- Exertional dyspnea, atypical chest pain, syncope, fatigue, peripheral edema that is progressive
- CXR is normal or shows big PA’s
- Right heart cath shows MAP >25mm Hg
- Exam is often normal or may show jugular venous distention, edema, hepatomegaly, right heart gallop or tricuspid regurgitation murmur

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Treatment of PAH

- Rx with oxygen, anticoagulation, diuretics, CPAP, treatment of underlying disease
- Embolectomy if due to chronic PE
- Calcium channel blockers for those with vaso-reactive PH on right heart cath
- Endothelin receptor antagonists: ambrisentan, bosentan, macitentan
- Oral phosphodiesterase inhibitors: sildenafil, tadalafil
- Oral guanylate cyclase inhibitors: riociguat
- IV epoprostenol, inhaled iloprost, IV treprostinil, or SQ treprostinil
- Atrial septostomy & Lung transplantation for those refractory to medical Rx
Extrapulmonary Restrictive Lung Diseases

- Kyphoscoliosis, scoliosis, ankylosing spondylitis
- Rib fractures, thoracotomy, pectus excavatum, flail chest
- Pleural diseases, mesothelioma, asbestosis, chronic pleural effusions, hemothorax, fibrothorax
- Obesity, ascites, diaphragmatic paralysis
Respiratory Failure

- ALS
- Stroke
- Head Trauma
- Brain Tumors
- Spinal Cord Injury
- Myasthenia gravis

Features of Respiratory Failure

- Inability to maintain adequate oxygenation or removal of carbon dioxide
- Small, shallow breaths or apnea
- Diminished breath sounds
- Atelectasis, poor inflation, pneumonia
- Somnolence due to elevated CO2 or dyspnea
Treatment of Respiratory Failure
- BiPAP
- Mechanical Ventilation
- Trilogy ventilator
- Diaphragmatic pacing

Nebulizer Therapy: Albuterol, Xopenex,
Atrovent, Duoneb, Budesonide, Brovana

Metered Dose Inhalers: Albuterol, Xopenex,
Combivent (Respimat), Advair, Symbicort, etc

Steroids: Prednisone, Dexamethasone, Medrol

Narcotics: Short & Long Acting

Mucolytics: Guafenesin, Mucomyst, Hydration

Mechanical Devices: Oxygen, Fan, Theravest,
BiPAP, Trilogy, Ventilator, IPV, IPPV
Cor Pulmonale

- Edema, Ascites, Liver Distention, Anorexia
- Diuretics: Lasix, Demadex, Zaroxolyn, Aldactone, Bumex
- Potassium & Magnesium supplementation
- Elevation, Lymphapres Wraps, Hose
- BiPAP, Oxygen

Hemoptysis

- Good Lung Up!
- Dark Towels!
- Stop any anticoagulants
- Narcotics
- Anxiolytics (Usually Lorazepam)
- Inhaled epinephrine

Fatigue/Low Energy

- Antidepressants: All are about equally effective, the problem is time for effect
- Dextroamphetamine, Adderall, Provigil, Ritalin (Methylphenidate), but all have side effects of tachycardia, anorexia, sleep disturbance
- Good sleep hygiene