PULMONARY DYSFUNCTION

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RESPIRATORY INFECTIONS

UPPER RESPIRATORY TRACT INFECTIONS (URI)

- Most are **viral**
  - **Rhinovirus, respiratory syncytial virus** (RSV), **adenovirus, parainfluenza**
  - Very contagious: hand-to-hand contact, inhalation upper respiratory tract

- Bacterial infections - certain organisms are quite common
  Streptococcus pneumoniae, other strep sp. **H influenzae**

- Pharynx
  - 60% viral; 30% bacterial
  - 5% **chlamydia** sp; 5-10% **mycoplasma** or **group A strep**

- Infections most frequent in fall and winter - indoors; enclosed spaces

- Pathophysiology and **clinical manifestations**

  - URI agents gain entry proliferate - initiate inflammatory reaction
  - Acute inflammation of upper airway structures
    Sinuses, nasopharynx, larynx, trachea
  - Pathogens trigger infiltration of mucus membrane
    - Inflammation
    - Infection-fighting cells
  - Cellular infiltration -> mucosal swelling and serous or **mucopurulent exudate**
    Clear discharge - viral
    Colored discharge - bacterial but viral can also be colored

  - Secondary bacterial infection - from obstruction of normal drainage pathways

VIRAL RHINITIS (common cold)

- Acute viral infection of upper airway: sinuses, pharynx
- Clinical presentation
  - **Rhinorrhea** (runny nose), sneezing
  - H/A, general **malaise**,
  - Scratchy/irritated throat
  - Colored **nasal discharge** (may indicate secondary bacterial infection)
  - **Redness**: nasal mucosal, oropharyngeal generalized redness

SINUSITIS

Usually bacterial infection of the sinuses/parasinus
Viral induced inflammation of sinus mucosa -> obstructs drainage
Obstruction results in ideal site for bacterial proliferation
Most common organisms
    Group A Streptococcus pyogenes
    Staphylococcus aureus
    Haemophilus influenzae
    Moraxella catarrhalis
Constitutional symptoms
    Purulent nasal discharge, sinus congestion, tenderness over sinus cavity, headache, post-nasal drip, cough, sore throat, sometimes fever
Chronic sinusitis: longstanding sinus infections

PHARYNGITIS - 'sore throat'

- **Usually viral** - can be bacterial invasion
- Clinical manifestations
    - Sore throat
    - Odynophagia: discomfort with swallowing
    - Hoarse voice (infection to larynx)
    - Tonsilitis can be present
    - Patchy **exudates**
        - **Group A beta hemolytic streptococcal infections**
        - **Mononucleosis**
        - Other bacterial infections
    - Sometimes **lymph nodes** swelling to neck
- Group A beta-hemolytic streptococcal infections - '**Strep throat**'
    - Untreated -> **rheumatic fever (**heart valve disease), **glomerular nephritis**
    - Requires antibiotic treatment - penicillin, erythromycin, others
- Most cases are **self-limiting**

LOWER RESPIRATORY TRACT INFECTION

One of three conditions needed to produce infection
    - Host defenses are weakened
    - Large enough inoculum
    - Organism must be sufficiently virulent

ACUTE BRONCHITIS

- Common - usually self-limiting
- Viral or bacterial infection of tracheobronchial tree

Etiologic agents

- **Usually viral**

  Rhinovirus, respiratory syncytial virus (RSV), parainfluenza virus, coronavirus, adenovirus, influenzae A and B

- Bacterial

  Mycoplasma pneumoniae, **chlamydia pneumoniae**, Bordetella pertussis

  - Fungal (occasional) usually with comorbid immunosuppression

Pathophysiology

- Inhalation or aspiration of secretions
- Pathology creates localized inflammatory reaction in airway mucosa
- Swelling and increased mucus production
- Significant inflammation and obstruction causes wheezing

Clinical manifestations bronchitis - similar to URI

ST, general malaise, chest congestion, cough (productive vs nonproductive), **chest tightness, retrosternal discomfort**, **wheezing** (sometimes), **fever** (sometimes)

- Individual not acutely ill - 3-10 days
- Residue **cough may persist for weeks**

PNEUMONIA

- Infection of lung where some/all of tissue becomes **edematous/fluid filled**
  - **Alveoli**
  - **Interstitial tissue**
  - **Bronchioles**

- Proliferation of infectious agents results in pathology and clinical features
- Certain individuals at greatest risk
  - Very young, very old
  - Immunosuppressed

Classification - numerous systems of classification

- Causative agents: bacterial, viral, etc.
- Virulent vs opportunistic infection
- "Typical" vs "atypical" infection
- Community acquired vs hospital acquired pneumonia

Etiology

Infectious organisms - most common - presentation varies with agent
Aspiration pneumonia: inhalation of secretions or inert substances
Chemical irritation-inflammation
Secondary bacterial infection often results
Particularly common in elderly or altered level of consciousness
Comatose patients, inebriated patients (ETOH)
Anesthetized patients (hence NPO requirement)

Pathophysiology

- Most causative organisms colonize oropharynx and nasopharynx
- Pathogens gain entry vis aspiration of oropharyngeal secretions

- **Mucociliary escalator** (ciliary action of epithelium removes secretions)
  - Normal means of clearing secretions
  - Compromised in COPD - chronic bronchitis

- **Hematogenous spread** via extra-pulmonary site
  - Blood seeded organisms to lungs
  - **IVDA** -> septic embolic with ***S. aureus***

Mechanism: Organisms -> inflammation -> infiltration -> changes to epithelial cells -> enhanced bacterial adherence

S. Pneumoniae: pneumococci causes inflammation which results in inflammation with response of neutrophils and congestion

S. aureus: multi small and large abscesses - necrotizing pneumonia

Pathogens destroy defense mechanism (alveolar macrophages) causing damage to alveoli - enter interstitium via terminal bronchioles -> lymphatic drainage lodges organisms on pleural surface

Pathological processes with pneumonia

Focal pneumonia: lobar, bronchopulmonary, bronchial, bronchiolar
Spread of infection varies: segment to segment; lobe to lobe
Bronchopulmonary: process not confined by anatomic barrier
Lobar pneumonia: spreads alveoli to alveoli until confined
Necrotizing pneumonia - lung abscesses
- Proteolytic, elastolytic enzymes from bacteria/inflamed cells
- S aureus, S pyogenes, gm neg bacteria, pseudomonas

Complications of pneumonia

Pleuritis: inflamation of the pleura
Pleural effusion: fluid in the lung
Pyothorax: pus in the pleural cavity
Empyema: pyothorax organizes and has fibrous wall
Bacteremia: circulating bacteria -> endocarditis, meningitis

Clinical manifestation pneumonia

- **Inflamation** occurs 5-10 days after bacterial infiltration
    - 70% of cases infiltration coincides with symptoms
    - Inflammations can extend 2-3 weeks
    - Complications can cause degeneration

- Typical presentation for community-acquired pneumonia
    Abrupt onset **chills**, **sweats**, **cough**, **purulent** and/or **rust colored sputum**, **pleuritic pain, fatigue, dyspnea, fever**

- **Elderly** presentation may be subtle
    Change in mental status, poor appetite, deterioration in underlying COPD disorders

Hospital-acquired pneumonia

- Symptom onset at least 48 hours after admission
- Infectious process not present on admission
- Temp > 38C; leukocytosis, CXR shows new pulmonary infiltrates
- Purulent endotracheal secretions
- High mortalities (as high as 70%) - M/M increases with comorbidity
    Cardiac disease, COPD, cirrhosis of liver, malignant disease and asplenia

Aspiration pneumonia

- Occurs when secretions-inert substances inhaled into lung
- Healthy individuals commonly aspirate secretions during sleep
- Oropharyngeal-neurologic dysfunction -> more frequent aspiration
- Medications, ETOH or altered LOC increase incidence
- Aspiration syndromes:
    - Chemical pneumonitis
    - Aspiration of bacterial pathogens
    - Aspiration of inert substances

Chemical pneumonitis

- Toxic substances introduced into lung
- Examples: gastric acid, bile, hydrocarbon fats, mineral oil
- Acute lung injury/inflammation -> necrosis/fibrosis airways
- Secondary bacterial infection in 50% cases
- Symptom onset is rapid: 2-5 hours
    Cyanosis, tachypnea, dyspnea, tachycardia, hypotension, bronchospasm, congestion, frothy sputa

Aspiration of bacterial pathogens

- Results in same kind of infections as discussed below
- Oropharyngeal cavity is most common source of pathogens

Aspiration of inert substances - may cause obstruction/pneumonitis

- Foreign bodies: teeth, food, etc
- Large amounts of water: near drowning
- S/S airway obstruction: coughing, cyanosis, wheezing
- Water dilutes surfactant -> atelectasis and ARDS

INFECTIOUS ORGANISMS CAUSING PNEUMONIA

Bacterial pneumonia - most common type of pneumonia

- Consolidated over lobe vs scattered over one or several lobes
- Scattered: "patchy infiltrates" - common with 'atypical' organisms
- ***Streptococcus pneumoniae*** - most common
  - 30%-50% of all community acquired pneumonia (CAP)
  - Outpatient mortality 1%-5%
- ***Mycoplasma pneumonia*** - common 'atypical pneumonia'
- Chlamydia pneumonia - 'atypical' pneumonia
- ***Staphylococcus aureus*** - purulent abscesses
  Common with IVDA induced hematogenic septic emboli

Viral pneumonia - 8% adult pneumo; 16% of children (incl bronchiolitis)
Common: influenzae, adenovirus, herpes, RSV
Adults: viral pneumonia affects alveolar epithelial cells
- Interstitial inflammation
- Intra-alveolar edema

Mononuclear cells characteristic
Typically rapid course -> acute respiratory distress; +/- fever
Bronchiolar damage -> secondary bacterial infection to alveoli

Atypical pneumonia - common

Mycoplasma pneumoniae is *main causative organism*
Develops gradually with prolonged course
Rarely fatal - common in young people (college, recruits)
Affects age 45 or lower most commonly
Patchy intracellular infiltrates on CXR - persists 6-8 weeks

Chlamydia pneumoniae is common
Also causes upper respiratory tract infection
Obligate intracellular organism (grows within host cells)
Eradication requires long-term broad spectrum antibiotic treatment

OTHER LESS COMMON ATYPICAL ORGANISMS

Legionella pneumophila
First noted: American Legion Convention 1976 (Philadelphia)
Fastidious bacteria living in aquatic environment
Outbreaks traced to air conditioners, cooling towers, condensers
Rapid growth in lungs -> alveolar fibrin and inflammation
Clinical picture complicated by empyema
Fever, cough, chest pain; mortality 10%-20%

Chlamydia psittaci - psittacosis
Small intracellular bacterium
Transmitted via birds and sheep
Flu-like disease -> irregular consolidation, interstitial pneumonia

Rickettsia - "Q-fever" - ***Coxiella burnetii***
- Spread via animals or infected dust particles
- Grows in intracellular macrophages
  Lung, liver, bone marrow, spleen
- Stimulates formation of granulomas

PCP - pneumocystis carinii pneumonia - very common **HIV infections**

- "Opportunistic" pathogen
- Infection implies a weakened immune system
- Most common in **AIDS/HIV infections**
- PCP does not typically cause disease with normal immune function
  - Organism has low virulence with immune competent host
  - May cause clinically inapparent illness in childhood
  - Altered T-cells of HIV+ patients may reactivate old infection

- Clinical manifestations are dependent on immune function
- Presentation varies according to HIV status

- HIV negative - immunocompromised - host has rapid deterioration
  - **Fever, nonproductive cough, dyspnea, congestion**
  - Diffuse alveolar and interstitial infiltrates on CXR
  - Hypoxia and respiratory failure in 4-15 days

- HIV+ host - progression is more insidious but with similar symptoms
  - **Low grade fever, weight loss, mild cough**
  - CXR shows intersitial infiltrates
  - Tachypnea with eventual hypoxemia
  - Severe respiratory alkalosis on ABG esp in terminal stage

Less common pathogens causing pneumonia

Fungi - various potential pathogens exist in soils/environment in US
- Organisms introduced via inhalation of infected dust particles
- **Coccidioidomycosis, histoplasmosis, cryptococcus**, others
- Chronic infection; lesions similar to Ghon complex of TB

Non-tubercular Mycobacterium
- Cause infections resembling TB
- Particularly immunosuppressed immunocompromised hosts
- ***Mycobacteria avium*** intracellulare (**MAC**)
- ***Mycobacteria kansasii***

PULMONARY TUBERCULOSIS

Epidemiology

- Major cause of death from infectious disease worldwide
- Rate TB declined 1950-1985
- Rate TB increased annually beginning 1985
    - Emergence of HIV
    - Decrease in federal funding for TB-control programs
    - Increase in number of immigrants from areas where TB is endemic

- More recent data - decline again beginning 1997

- Specific populations at risk (CDC 1995 *Morbidity and Mortality* 44 (No RR-11)
    Close contacts of TB cases
    HIV infected
    Homeless, medically underserved or low-income groups
    Substance abusers (ETOH or street drugs)
    Residents-employees of medical institutions, shelters or correctional facilities
    Recent immigrants where TB is prevalent; high risk ethnic groups
    Infants, children and adolescents in contract with high risk adults

Etiology

- ***Mycobacterium tuberculosis*** - aerobic, rod-shaped, acid-fast bacilli
- Spread via aerosolized droplet nuclei from infected person
    - Droplets expelled into environment from infected host
    - Laughing, sneezing, coughing, singing
- Droplets gain entry into airway and proliferate -> new TB infection
- Immune system determines extent and nature of infection
    - Normal hosts - immune system contains infection -> **inactive/dormant**
    - Progression to **primary TB occurs** where immune system cannot contain
    - **Reactivation -> active TB** - risk is 5% to 10% over lifetime

Pathophysiology

- M TB can **escape** destruction by **macrophage** -> induces **Type IV hypersensitivity**
- Elude destruction from phagocytes via avoiding lysosomal destruction
- Staging: primary infection, secondary/disseminated infection

Primary Infection

- Inhaled into alveoli; evades protective mechanism of lung via small size
- Organism lodges in lung periphery; phagocytosed by **alveolar macrophages**
- Macrophages transport organisms to hilar lymph nodes
- Establishing infection depends on two factors
    - Number of organisms
    - Alveolar macrophage microbicidal activity
        Macrophage may not kill M TB but contain them in giant cells
        T lymphocytes interact with macrophage to form **granulomas**
        Granulomas may sometimes kill organisms

- **Ghon complex** - well healed calcified lesion with tissue necrosis and scarring

- T-cell mediated response takes 4-6 weeks - seen via **positive PPD**
    - Old infection may require two doses of PPD to show positive
    - First dose stimulates immune system; 2nd dose reacts

- Minority of patients - progressive disease follows primary infection
    - **Cavitation, tubercular pneumonia, miliary TB**
    - Infants and immune-deficients adults are vulnerable

Secondary or Reactivation Tuberculosis

- Most cases due to reactivation of primary infection
- Disseminated organisms which did not produce a clinical infection
- Formulation of many **granulomas** and extensive tissue necrosis
- **Tubercles** - central areas of caseous necrosis
    - May heal or they may erode bronchus and drain infective material
    - Cavities **are large; tend to be situated in apices of lung**
- **Tuberculous** bacillemia - organisms cross alveoli into lymphatics
    - Extra-pulmonary infections occurs: bone, gut, urinary tract, etc.
    - Known as **miliary TB**

Clinical manifestations

Fever, *weight loss*, night sweats, malaise,
Cough*, sputum production, vague chest pains and *hemoptysis
Classic picture: hemoptysis and weight loss
Physical exam frequently normal
Common: active lung infiltrates, pleural effusion, wasting

Diagnostics for TB

PPD positive (purified protein derivative)
- Indurated (raised) reaction 48 hours after intradermal injection
- Suggests presence of memory T cells for M Tb
- 10 mm or larger needed to be consider positive
- Less than 10 mm may be positive for some groups esp HIV+
- Age, geography, medications affect whether <10 mm is positive
- Test not 100% reliable; can't distinguish recent vs past infection

Acid fast bacilli (AFB) on initial **sputum smear**
- Termed positive smear
- Positive smear does not confirm that organism is tuberculosis
- Positive smear suggests heavy bacterial load -> strong potential for contagion

Sputa culture positive - *M TB* grows out in culture

Radiographic manifestations
Infiltrates - certain areas particularly common
- Upper lobes or superior segments
- Superior segments of middle or lower lobes

Cavities or **calcified nodular** lesions for old infection
Immunocompromised host may show normal CXR even with active infection

OBSTRUCTIVE DISORDERS

REVERSIBLE OBSTRUCTIVE DISORDERS

ASTHMA

- Chronic inflammatory disorder of airways
- **Reversible** airway **bronchospasm**, mucus hypersecretion, airway edema
- Reversibility differentiates asthma from other COPD
    - Asthmatics can have symptom free periods
    - Key to control is in combating/preventing inflammation

Etiology

- Multifactorial; dependent on age of presentation
- **Atopy** frequently associated with child-onset asthma
    Atopy is genetic propensity to produce IgE proteins
    IgE towards common environmental antigens
    House-dust mites, fungi, animal proteins

Adult-Onset Asthma

- Factors which increase likelihood of developing asthma
    - Childhood history allergy or wheezing with viral infections
    - Family history of allergies - less common vs childhood allergies
- Allergies play a significant role but not to same extent as with children
    - IgE antibodies less common vs childhood asthma
    - Common: respiratory tract infections, nasal polyps, sinusitis

Occupational Asthma

- Disorder occurring with work environment exposures
- Environmental pollution may contribute
- Triggers: **dusts, fumes, animal dander, molds**

Drug-Induced Asthma

- Asthma-like symptoms due to hypersensitivity to various drugs
- Aspirin common trigger; also propranolol, NSAIDS

Exercise-Induced Asthma

- Occurs in individuals with no other trigger for asthma
- Exercise may provoke a response in a known asthmatic
- Heat or water loss from airway epithelium

Emotional Triggers - seen in approximately half asthmatics

Pathophysiology

Airway inflammation most important pathophysiologic factor
Inflammation may occur in response to allergen or infection exposure

Inflammatory-mediated pathophysiologic changes
- Airway hyperresponsiveness ('twitchiness")
    - Airflow restriction, symptoms, chronic disease
- Bronchospasm - involuntary tightening airway smooth muscles
- Airway edema, airway wall remodeling, mucus plug formation
- Immunologic responses
    - Mast cell activation
    - Infiltration of inflammatory cells - PMN, eosinophils, lymphocytes
- Denudation of airway epithelium
- Collagen deposition below basement membrane

Classification

Classified according to severity:
Mild intermittent
Mild persistent
Moderate persistent
Severe persistent

Symptom assessment
- Frequency of symptoms
- Frequency and character of attacks
- Use of medication
- Presence or absence of night-time symptoms
- Pulmonary function values

Clinical manifestations

- Acute, abrupt onset cough, **wheezing, chest tightness**, **tachypnea**, tachycardia
- Increased work of respiration - use of **accessory muscles**
- "Late-phase reaction" - occurs 4-8 hours after exposure to triggering antigen
    - Coincides with inflammatory response
- Atopic disease comorbidity in some cases
    - Concurrent allergic symptoms
    - Rhinitis, nasal polyps, sinusitis, eczema
- **Hypoxemia** and **respiratory fatigue** in severe cases
- **Respiratory failure** if accompanied by hypercarbia

- **Pulmonary Function Testing**
    - Airway obstruction from inflammation, swelling, mucus, bronchospasm
    - **FEV1** (forced expiratory volume in one second) - decreased during attack
    - **PEF** (peak expiratory flow) - decreased during attack

CLASSIFICATION OF ASTHMA

INTERMITTENT

- Symptoms no more than twice a week
- Nocturnal symptoms no more than twice a month
- Exacerbations are brief (few hours to few days);
- Normal between episodes
- Peak expiratory flow (PEF) is normal
- Forced expiratory volume in 1 second (FEV1) or PEF
    - At least 80% of predicted normal value
    - Varies by less than 1 second

MILD PERSISTENT

- Symptoms more than twice per week but not every day
- Nocturnal symptoms more than twice per month
- Exacerbations curb pt's daily activities
- FEV1 or PEF is 80% predicted value
- PEF varies by 20-30%

MODERATE PERSISTENT

- Daily asthma symptoms are th rule
- Daily use of inhaled, short acting B2 agonist
- Asthma episodes
    - Disrupt pt's activities
    - Occur at least twice a week
    - Continue several days

- Nocturnal symptoms typically occur once per week
- FEV1 or PEF between 80% and 61% of predicted value
- PEF varies by more than 30%

SEVERE PERSISTENT

- Symptoms are continual and restrict physical activity
- Exacerbations are frequent
- Nocturnal symptoms frequent
- FEV1 or PEF is no more than 60% predicted value
- PEF varies by more than 30%

Adapted from the National Institutes of Health: National Heart and Lung and Blood Institute (1997) Expert Panel Report II: Guidelines for the diagnosis and management of asthma. NIH Publication No 97-4051 p 8 Washington, DC: NIH

COPD - *CHRONIC OBSTRUCTIVE PULMONARY DISEASE*

- Gradually progressive airway obstruction
- Sometimes with hyperactive airway - possibly partially reversible

- COPD is global term - used to define group of overlapping heterogenous disorders
  - **Emphysema**
  - **Chronic Bronchitis**
  - **Bronchiectasis**

Epidemiology - extremely prevalent (14 million Americans)

Death rate is 18.6 per 100,000 people
Tobacco smoke is primary cause
Typical person has at least 20 pack-year history and presents in 5th decade
Pack year: number of packs per day x number of years smoking

Typically presents in context of acute or recurrent respiratory illness or cough
Sixth decade - **dyspnea** is constant feature
Persons typically present with **mixed picture** emphysema, chronic bronchitis, etc.
Rarely does person present with only a single entity
One form may predominate

Virtually all smokers will develop the disease to some extent
Contrasts with lung cancer wherein some smokers may never be affected
COPD esp emphysema may develop in absence of smoking
Alpha-1 antitrypsin disease
Environmental pollutants, asbestos exposure, etc.

Risk factors

Cigarette smoking - Passive smoke exposure
Male sex - nonwhite race
Low socioeconomic status
Alpha 1-antitrypsin deficiency
Occupational exposure
Hyper-responsive airways

EMPHYSEMA

- Abnormal and permanent enlargement of airspaces distal to terminal bronchioles
- Resultant destruction of walls of alveoli
- Classified according to anatomic changes from destruction
  Centrilobular emphysema: dilation/destruction involves central part of acinus
  Panacinar emphysema: dilation/destruction involve entire acinus
    - Results from genetic deficiency of alpha 1-antitrypsin
    - Early onset emphysema including non-smokers
- Most severe types occur in men who smoke heavily

Pathophysiology

- Permanent destruction of air spaces
- **Alveolar walls destroyed** without evidence of fibrosis
    - Significant **hyperinflation**
    - Decrease in functional alveolar capillary bed surface area
    - Inefficient gas exchange

- Destruction causes unsupported small airways -> distorted and deformed
    Deformity results in **air-trapping**
    Premature small airway closure during exhalation

Clinical manifestations - evident with destruction of 1/3 functioning parenchyma

- **Dyspnea** is first symptom - > steadily more pronounced
- **Cough** and **wheezing** common esp with associated bronchitis
- Cachetic body habitus
- **Increased anterior-posterior** diameter (barrel-chested)
- **Prolongation of expiratory** phase of respiratory cycle
- Use of **accessory muscles** in **forced expiration**
- **Florid** skin color - "**Pink Puffer**"
    ABG only moderately hypoxemic until late-stage disease
    Skin color pink even in face of severe pulmonary damage
- **Erythrocytosis** (high hematocrit) from hypoxemia
- **SOB** with activity - **little sputa** production
- Breath sounds distant
- Respiratory failure from long-term energy cost
- **CXR - hyperinflation**

CHRONIC BRONCHITIS

- Defined as presence of **continual productive cough**
- Criteria: **cough more than half the time over period of 2 years**
- Smoking is the cause in over 90% of cases
- Increase in **mucus secretion by goblet cells** of bronchial mucous glands

Pathophysiology

- Involve **airways** rather than alveoli
- **Goblet cells** (airways) multiply and secrete **excessive mucus**
- **Squamous metaplasia** of bronchial epithelium

- **Hypertrophy** of airway smooth muscle
    - Basal cells become hyperplastic
    - Basement membrane thickens

- **Chronic inflammation-infection** attracts lymphocytes and macrophage

- **Small airways**: distorted and plugged with secretions
    - Loose structural integrity from supporting cartilage damage
    - **Close prematurely** during exhalation -> **airway trapping**

- **Sustained hypoxia** -> **erythropoietin** from kidney -> stimulates **RBC**
- **Erythrocytosis** attempts to increase O2 delivery to offset hypoxia
- Excessive RBCs **increase blood viscosity** -> interfere with circulation

Clinical Manifestations

- Initial sputa is mucoid -> **increase quantity-purulence** during respiratory infections
- Expectoration mostly during **morning**
- **Acute exacerbation chronic bronchitis (AECB)**
    - Acute respiratory illness intermittently superimposed
    - Increase in frequency in later stages of disease

- Chronicity results in progressive deterioration
    - **Chronic productive cough**
    - **Chest congestion**
    - **SOB at rest**

- Late signs of chronic bronchitis
    - **Fluid retention** in the periphery -> **edema**
    - **Cor pulmonale** (right-sided heart failure)
    - **Cyanotic** appearance of skin - **"Blue Bloater"**

- **CO2 retention** -> **hypercarbia** and **hypoxemia** on ABG
- Auscultation: **crackles, wheezes, rhonchi**

COMPARISON BRONCHITIS AND EMPHYSEMA

	PREDOMINANT BRONCHITIS	PREDOMINANT EMPHYSEMA
Age	40-45	50-75
Dyspnea	Mild; late	Severe, early
Cough	Early; copious sputum	Late; scanty sputum
Infections	Common	Occasional
Respiratory insufficiency	Repeated	Terminal
Cor pulmonale	Common	Rare, terminal
Airway resistance	Increased	Normal or slightly increased
Elastic recoil	Normal	Low
Chest radiograph	Prominent vessels, large heart	Hyperinflation, small heart
Appearance	*Blue Bloater*	*Pink Puffer*

Adapted from Cotran, R.S., Lumar V and Collins T (1999) Robbins pathologic basis of disease (6th ed.) Philadelphia: W.B. Saunders

BRONCHIECTASIS

- Permanent **dilation** and **destruction** of cartilage-containing **airways**
- Multiple pulmonary insults vs a specific disease entity
- Very common prior to antibiotic therapy for respiratory infections

Etiology

- Overtime **chronic inflammation** - > **impaired mucociliary clearance** -> airway abnormalities
  - **Repeated infection**
  - **Toxic exposure** or **foreign body**
- Other causes of bronchiectatic airway
  - **Pulmonary TB, fungal infections**
  - Genetic disorders e.g. **cystic fibrosis**

Pathophysiology

- **Mucus secretion** by **goblet cells** (bronchial mucous glands)
- Secretions cause **deformity and dilation of distal airways**
- Bronchiectatic changes confined to one or two neighboring lobes
- Left lower lobe most common site
- Inflammation and denuding of airway epithelium common
- Supportive cartilage and elastic tissue damaged
- **Peribronchial pneumonia** or **atelectasis** common around bronchiectasis
- **Obstruction** from **inflammatory infiltration** of small airways
- **Fibrosis** in severe cases
- Bacteria proliferate in bronchiectatic deformations -> pneumonia

Clinical manifestations

- **Persistent cough** with **copious purulent sputum** (can be > 100 cc/day)
- Congenital disorders: symptoms may start as early as 2- 7 years
- Post-infective bronchiectasis -> symptom onset is more insidious
- Individual may present history of childhood infections
- Systemic or genetic disorders may present with additional symptoms
  - Sinus disease, malabsorption
  - Example: Cystic fibrosis

- **Hyperresponsive airways** and **wheezing** are common
- **Coarse inspiratory** and **expiratory crackles** - clear with coughing
- CXR: abnormally enlarged airways
- PFT and ABG: similar to those associated with chronic bronchitis

CYSTIC FIBROSIS

- Inherited **autosomal recessive** exocrine disorder
- Affects 1 in 1500 to 4000 live births
- Pulmonary manifestations due to **viscous mucus secretions** - **obstructed/infected airways**
- Different combinations of gene abnormalities present inconsistently
    - Varying disease characteristics of organ involvement in a given individual
    - Not all individuals with CF manifest significant pulmonary involvement

Pathophysiology

Pathology due to dysfunction of epithelial **chloride ion channels**
- Channels are closed or absent
- Results in excessive sodium reabsorption -> decreased chloride excretion

Mucus abnormalities secondary to ion abnormality
- Dehydration of mucous layer
- Defective mucociliary action
- Mucus plugging of airways

Pseudomonal colonization - *Pseudomonas aeruginosa*
- Particularly mucoid form which resists antibiotics
- Bronchiectasis results in response to infection

Clinical manifestations

- Cough, chest congestion, copious sputum, shortness of breath
- Symptoms begin in infancy and are progressive
    - **Obstruction, deterioration** of pulmonary function
    - **Dyspnea, hemoptysis, hypoxemia**
    - Complex **bacterial infections**
- Death secondary to **respiratory failure** - 95% cases