Introduction to Pulmonary Impairments

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- Respiratory System
  - Gas exchange system
    - Inhale oxygen
      - Oxygen delivery to the blood
    - Exhale carbon dioxide
  - Upper airway system
  - Lower airway system
  - The upper and lower airways are anatomically distinct, but interrelated in many ways.
Introduction to Pulmonary Impairments

Upper Airway System (UAS)

- **UAS**: Above the glottis (7 components)
  - The **SINUSES** (frontal, maxillary, and sphenoidal) are hollow spaces in the bones of the head. Small openings connect them to the nose. The functions they serve include helping to regulate the temperature and humidity of air breathed in, as well as to lighten the bone structure of the head and to give resonance to the voice.
  - The **NOSE** (nasal cavity) is the preferred entrance for outside air into the respiratory system. The hairs that line the wall are part of the air-cleaning system.
  - Air also enters through the **MOUTH** (oral cavity), especially in people who have a mouth-breathing habit or whose nasal passages may be temporarily obstructed, as by a cold or during heavy exercise.
  - The **ADENOIDS** are lymph tissue at the top of the throat. When they enlarge and interfere with breathing, they may be removed. The lymph system, consisting of nodes (knob of cells) and connecting vessels, carries fluid throughout the body. This system helps to root body infection by filtering out foreign matter, including germs, and producing cells (lymphocytes) to fight them.

- **TONSILS** are lymph nodes in the wall of the throat (pharynx) that often become infected. They are part of the germ-fighting system of the body.
- The **THROAT** (pharynx) collects incoming air from the nose and mouth and passes it downward to the windpipe (trachea).
- The **EPIGLOTTIS** is a flap of tissue that guards the entrance to the windpipe (trachea), closing when anything is swallowed that should go into the esophagus and stomach.
- UAS treated primarily by a **otolaryngologist**
Introduction to Pulmonary Impairments
Lower airway system (LAS)

- LAS: Below the glottis (14 components)
  - The VOICE BOX (larynx) contains vocal chords. It is the place where moving air being breathed in and out creates voice sounds.
  - The ESOPHAGUS is the passage leading from the mouth and throat to the stomach.
  - The WINDPIPE (trachea) is the passage leading from the throat (pharynx) to the lungs.
  - The LYMPH NODES of the lungs are found against the walls of the bronchial tubes and windpipe.
  - The RIBS are bones supporting and protecting the chest cavity.
  - They move in a limited degree, helping the lungs to expand and contract (Chest Wall).
  - The windpipe divides into the two main BRONCHIAL TUBES, one for each lung, which subdivide into each lobe of the lungs. These, in turn, subdivide further.
  - The right lung is divided into three LOBES, or sections. Each lobe is like a bullpen filled with sponge-like tissue. Air moves in and out through one opening – a branch of the bronchial tube.

Introduction to Pulmonary Impairments
Lower airway system (LAS)

- The left lung is divided into two LOBES.
- The PLEURA are the two membranes, actually one continuous one folded on itself, that surround each lobe of the lungs and separate the lungs from the chest wall.
- The bronchial tubes are lined with CILIA (like very small hairs) that have a wave-like motion. This motion carries MUCUS (sticky phlegm or liquid) upward and out into the throat, where it is either coughed up and swallowed. The mucus catches and holds much of the dust, germs, and other unwanted matter that has invaded the lungs. You get rid of this matter when you cough, sneeze, clear your throat.
- The DIAPHRAGM is the strong wall of muscle that separates the chest cavity from the abdominal cavity. By moving downward, it creates suction in the chest to draw in air and expand the lungs.

Introduction to Pulmonary Impairments
Lower airway system (LAS)

- The smallest subdivisions of the bronchial tubes are called BRONCHOLES, at the end of which are the air sacs or alveoli (plural of alveoli).
- The ALVEOLI are the very small air sacs that are the destination of air breathed in. The CAPILLARIES are blood vessels that are imbedded in the walls of the alveoli. Oxygen from the alveoli crosses the capillaries, brought to them by the PULMONARY ARTERIES and carried away by the PULMONARY VEINS. While in the capillaries, the blood gives off carbon dioxide through the capillary wall into the alveoli and takes up oxygen from the air in the alveoli.
- LAS treated primarily by a pulmonologist.
Introduction to Pulmonary Impairments

- UA and LA systems
  - Common and often co-morbid conditions
  - Common risk factors
  - Share many pathophysologic processes
  - Allergen challenge in either the upper or lower airway may have an impact on the other portions of the airway

Introduction to Pulmonary Impairments

- Dyspnea: Shortness Of Breath (SOB)
  - Grade 1: SOB with severe exertion
  - Grade 2: SOB with climbing
  - Grade 3: SOB with prolonged walking and/or climbing
  - Grade 4: SOB from ambulating several minutes on even ground
  - Grade 5: SOB with activities of daily living (ADLs) and/or at rest

- Classifications of Respiratory Impairment

<table>
<thead>
<tr>
<th>Class</th>
<th>Impairment</th>
<th>Limitations</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>0%</td>
<td>None-severe FEV1, within 85%</td>
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<tr>
<td>2</td>
<td>10-20%</td>
<td>Mild FEV1, 70-85%</td>
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<tr>
<td>3</td>
<td>25-39%</td>
<td>Moderate FEV1, 50-70%</td>
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<tr>
<td>4</td>
<td>30-50%</td>
<td>Severe FEV1, &lt;50%</td>
</tr>
</tbody>
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Introduction to Pulmonary Impairments

- Traumatically induced Impairments
  - Burns
    - Smoke Inhalation Injury Complex
    - Systemic Effect of Heat (Hot Air) Injuries
    - Restrictive Chest Wall Burn
  - Spinal Cord Injury
  - Brain Injury
  - Chest Trauma
Introduction to Pulmonary Impairments
Burns

- Smoke inhalation injury complex
  - Carbon Monoxide, Cyanide
  - Upper Airway Inhalation (UAI) injury above the glottis (vocal cords) with edema
  - Lower Airway Inhalation (LAI) injury below the glottis (vocal cords) with a chemical burn
  - Lung injury
  - Impaired chest wall compliance

Introduction to Pulmonary Impairments
Burns

- Systemic Effect of Heat (Hot Air) Injuries
  - Upper Airway Injuries (UAI)
    - Cause upper airway/laryngeal obstruction from tissue inflammation
      - Hot Air
      - Noxious gases
  - Lower Airway Injuries (LAI)
    - Smoke/irritant gases
    - Hot particulates
    - Aspiration
    - Lungs causing pulmonary edema and/or alveolar-capillary defect (a-c defect)

Introduction to Pulmonary Impairments
Burns

- Systemic Effect of Heat (Hot Air) Injuries
  - Inflammatory response
    - Local (site of injury)
    - Regional (unjured respiratory tissue)
    - Distant (systemic effects)
  - Injuries
    - Cell damage
    - Cell surface
    - Vasodilation
    - Vascular permeability
    - Bronchospasm and/or constriction
Introduction to Pulmonary Impairments

**Burns**

- **Restrictive Chest Wall Burn - External**
  - Loss of elasticity for inspiration
  - Treatment
    - Residuals to grafting
    - Residuals to escharotomies
  - Consequences
    - Dyspnea
    - Fatigue
    - Atelectasis (collapse)
    - Impaired gas exchange

**Introduction to Pulmonary Impairments

**Burns**

- **Upper Airway Burn Injuries**
  - Facial burns causing external compression of the larynx and/or non-elastic burn tissue preventing expansion
  - Airway obstruction
    - Oral Burns: Injury to tongue and airway mucosa causing edema, erythema, and ulceration
    - Supraglottic Edema
    - Cord and Intraglottic Edema

**Introduction to Pulmonary Impairments

**Burns**

- **Lower Airway Burn Injuries**
  - Mucosal damage with slough
  - Increased secretions
  - Airways inflammation
  - Bronchoptasis
  - Airway plugging impairing gas exchanges
  - Bacterial colonization
  - Airway infection
  - Continued upper airway obstruction
  - Decreased chest wall compliance
  - Tracheobronchitis from inhalation injury
  - Cardiogenic or high pressure pulmonary edema
  - Impaired immune function
  - Cardiogenic pulmonary edema
  - Respiratory failure
  - Impaired particulate and bacterial clearance
Introduction to Pulmonary Impairments

SCI

- Impact of SCI on pulmonary function by level of impairment
  - T5-T12
    - Progressive loss of abdominal and chest wall function that impairs expiration and cough
  - T1-T5
    - Intercostal function is impaired affecting inspiration and expiration
  - C4
    - Phrenic nerve and diaphragm involvement
    - May possibly be weaned from ventilator
  - C3-Above
    - Total ventilator dependence
  - High level attendant care
  - Frequent physician visits
  - May consider phrenic pacers (fewer complications, greater quality of life, improved longevity)

Introduction to Pulmonary Impairments

Brain Injury

- Loss of Autonomic Function
  - Much the same as that of a high level SCI
- Physical Fatigue
- Mental fatigue
- Limited or inability to direct own care
- High level attendant care
- Frequent physician visits

Introduction to Pulmonary Impairments

Chest Trauma

- Physiology of Injuries
  - Chest Wall
  - Airways
  - Diaphragm
  - Lungs
- Types of Injuries
  - Penetrating wounds
  - Lacerations
  - Blunt trauma
  - Soft tissue – pulmonary contusions
  - Fractures
  - Electrical Shock
- Broad Range of Medical and Therapeutic Implications and Interventions
Introduction to Pulmonary Impairments

Occupational/Acquired Conditions

• Reactive Airway Dysfunction Syndrome (RADS)
  – Most common new cause of lung disease
  – Occupational Asthma (OA)
    • Occupational asthma is a disease characterized by variable air flow limitation and/or airway hyperresponsiveness due to causes and conditions attributable to a particular occupational environment and not to stimuli encountered outside the workplace (Bernstein et al 1993).
  – Pneumoconiosis
    • Greek for “Dusty Lungs”, a.k.a. “Black Lung”
      – Permanent and/or progressive alteration of lung structure caused by the reaction of lung tissue to inhalation of mineral dust particles
        • Nodular fibrosis (silicosis)
        • Diffuse fibrosis (asbestosis)

• Malignant Mesotheliomas
  – 20 - 40 years after exposure.
  – Risks associated with age, duration, intensity and type of exposure
  – Not associated with smoking
  – No known effective therapy but may remain static or progress over time
  – Occupational Sources:
    • Mining
    • Milling
    • Transporting
    • Manufacture of asbestos-containing products
    • Construction
    • Shipbuilding
    • Plumbing
    • Pipefitting
    • Insulating
    • Electrical

• Silicosis
  – Inhalation of crystalline silica
  – Acute and Accelerated Silicosis: Within 5 - 15 years of exposure
  – Chronic: after 20 years exposure
  – Occupational Sources:
    • Mining
    • Foundry
    • Drilling
    • Quarrying
    • Construction
Introduction to Pulmonary Impairments
Occupational/Acquired Conditions

- Mycobacterial Disease (Hypersensitivity Pneumonitis)
  - Hot tubs and indoor swimming pools
  - Water damaged buildings
- Chronic Idiopathic Interstitial Lung Disease
  - AKA: Hard Metal Disease or Hard-Metal Pneumoniosis
  - Caused by recurrent lung injury or autostimulating processes
  - Exposure to Cobalt used as a catalyst for bonding of tungsten to carbon
  - Treated acutely with corticosteroids
  - Exposure to metal fume: 6-12 weeks of steroid therapy (e.g., 0.5 to 1.5 mg/kg/day of prednisone) tapering with improvement with no expectation of return to normal pulmonary function
  - Cytotoxic in combination with steroids (e.g., Azathioprine 100 mg q.d. inflating by 50 mg every two weeks to a maximum of 200 mg q.d.) requiring hepatic and lymphopenia monitoring
  - Occupational Sources
    - Users of high speed tools (dental labs, sharpening services, polishing services)
- Beryllium-Induced Lung Disease (CBD or berylliosis: Granulomatous Lung Disease)
  - Delayed hypersensitivity reaction
  - Genetic predisposition
  - Systemic involvement
  - Treatment
    - No cure
    - Treated acutely with corticosteroids
    - Treated end stage with methotrexate
    - Lung Transplant may be indicated
  - Occupational Sources
    - Manufacturing of cutting and boring tools
    - Nuclear power, aerospace and electronic industries
- “Popcorn Workers Lung
  - Bronchiolitis Obliterans
  - Diacetyl fumes
  - Fixed airway obstruction
  - Lung scarring
  - Lung inflammation
  - Dry cough
  - SOB
  - Diminished lung capacity
  - Only known treatment may be lung transplantation
  - Some evidence of possible improvement over time
Introduction to Pulmonary Impairments
Lung Cancer

• Non-small cell lung cancer is staged using the TNM system
  – "T" for extent of primary tumor
  – "N" for regional lymph node involvement
  – "M" for metastases
• Primary Tumor (T) Descriptors
  – T1: A small tumor not locally advanced or invasive
  – T2: A larger, minimally advanced or invasive
  – T3: Any sized tumor locally advanced or invasive up to but not including major intrathoracic structures
  – T4: Any sized tumor advanced or invasive into the major intrathoracic structures

Introduction to Pulmonary Impairments
Lung Cancer

• Regional Lymph Node (N) Involvement
  – N1: Metastatic disease to nodes within ipsilateral lung
  – N2: Metastatic disease to nodes beyond ipsilateral lung but not contralateral to primary tumor
  – N3: Metastatic disease to nodes distant to those of N2
• Metastases (M)
  – M0: Local or regional without distant metastases
  – M1: Disseminated with distant metastases
• Staging
  – IA: T1N0M0
  – IB: T2N0M0
  – IIA: T1N1M0
  – IIB: T2N1M0, T3N0M0
  – IIIA: T4N(0-3)M0, T1-4N3M0
  – IVA: T(Any)N(Any)M1

Introduction to Pulmonary Impairments
Lung Cancer

• Treatment by Stage
  – Non-Small Cell
    – Stages IA, IB, IIA, IIB:
      – Surgical resection if able to tolerate
      – Limited resection if unable to tolerate larger resection
      – Radiography if unable or unwilling to undergo resection
      – Adjunct radiotherapy if resection has occurred
      – Adjunct chemotherapy a consideration
    – Stage IIIA
      – Concurrent chemoradiotherapy using a platinum-based regimen
      – Induction chemoradiotherapy followed by resection as part of study protocol
    – Stage IIIB
      – Concurrent chemoradiotherapy using a platinum-based regimen
      – Concurrent chemotherapy followed by resection as part of study protocol
    – Stage IV
      – Concurrent chemoradiotherapy using a platinum-based regimen
Introduction to Pulmonary Impairments
Lung Cancer

- **Treatment by Stage**
  - **Small Cell**
    - **Limited Stage**
      - Combination chemotherapy with concurrent hyperfractionated radiotherapy
      - Prophylactic cranial radiation following complete response to chemotherapy
    - **Extensive Stage**
      - Combination chemotherapy

Introduction to Pulmonary Impairments
Chronic Obstructive Pulmonary Disease

- “Air flow obstruction that is chronic, progressive, and for the most part fixed.”
- “A diseased state characterized by airflow limitation that is not fully reversible. Airflow limitation is usually both progressive and associated with an abnormal inflammatory response of the lungs to noxious particles or gases.”
- **A.K.A.** Emphysema
  - “alveolar wall destruction with irreversible enlargement of the air spaces distal to the terminal bronchioles and without evidence of fibrosis”
- 4th leading cause of death in US behind heart disease, cancer and cerebrovascular disease
- Only disease worldwide in which prevalence and mortality continue to rise
- Cigarette smoke is the main causative agent

Introduction to Pulmonary Impairments
Chronic Obstructive Pulmonary Disease

- **Chronic Bronchitis**
  - “productive cough that is present for a period of 3 months in each of 2 consecutive years in the absence of another identifiable cause of excessive sputum production”
- **Diagnosis**
  - Supportive risk factors:
    - Family history
    - Smoking exposure history
  - Laboratory tests
    - Spirometry demonstrates a decreased forced expiratory volume in 1 second (FEV1) and FEV1/FVC, forced vital capacity (FVC) ratio
    - Diffusing capacity for carbon monoxide (DLCO) may differentiate between emphysema and chronic bronchitis
  - Chest radiograph to exclude other etiologies of symptoms
  - High-resolution computed tomography (CT) imaging
### Introduction to Pulmonary Impairments

#### Chronic Obstructive Pulmonary Disease

- **Disease Staging of Disease Severity (FEV1 % Predicted)**
  - Stage 0: At Risk Normal
  - Stage 1: Mild >80 FEV1 % Predicted (but FEV1/FVC <70)
  - Stage 2: Moderate 30 – 79 FEV1 % Predicted
  - Stage 3: Severe <30 FEV1 % Predicted

- **Bronchodilators**
  - Stage 1
    - Short-acting ß-adrenergic agonist (e.g., albuterol)
  - Anticholingergic prn
  - Stage 2 and 3
    - Use of one or more bronchodilators (combined albuterol and ipratropium bromide is recommended)
    - Use of inhaled once daily for long-term treatment of bronchospasm
    - Corticosteroids
      - Oral prednisone (e.g., 30 mg q.d. 2 weeks)
      - Inhaled steroid (e.g., beclomethasone 500 mg b.i.d. for 6 weeks)
      - Corticosteroids (e.g., 0.4 to 0.6 mg/kg/day for 2 to 4 weeks)

- **Other Treatments**
  - Pulmonary rehabilitation
  - Immunizations
    - Prophylactic annual influenza vaccine
    - Prophylactic 23 polyvalent pneumococcal vaccine 1 X/5 10
    - Antibiotics
      - No prophylactic benefit
    - Mucokinetics
      - No prophylactic benefit
    - Others are contraindicated
      - Narcotic antitussives
        - Inhaled nitric oxide

- **Non-pharmacologic Treatments**
  - Pulmonary rehabilitation (at all stages)
  - Long-term oxygen therapy (LTOT)
  - Ventilatory support
  - Lung volume reduction surgery (LVRS)
  - Nocturnal non-invasive ventilatory support
  - Unproven
  - Lung transplantation for severe airflow obstruction and function impairment
    - Single lung transplant actuarial 5 year survival rate 43.2%
Introduction to Pulmonary Impairments
Pediatric Respiratory Conditions

- Anatomy:
  - Airway size
  - Airway maturity
- Mechanics:
  - Chest structure
  - Dependence on diaphragmatic breathing during infancy
  - Nose breathing for first few months of life
  - Tongue size in relationship to mouth effecting airway interventions such as intubation
  - Bronchi expansion during inspiration and contraction during expiration increases risk of air trapping
  - Larynx and glottis is higher in the airway increasing risk of aspiration
  - Cartilagic cricoid ring provides a natural intubation cuff until about age 8
  - Lower airways epithelial lining more susceptible to edema and airway narrowing during respiratory illness
  - Anatomically adult lungs by age 8 with continued remodeling into adolescence.

Introduction to Pulmonary Impairments
Pediatric Respiratory Conditions

- Congenital Disorders
  - Cystic fibrosis
  - Tracheoesophageal fistula
  - Diaphragmatic hernia
  - Congenital central hypoventilation syndrome
- Acquired disorders
  - Diseases of prematurity
  - Bronchopulmonary Dysplasia (BPD)
- Chronic pulmonary/immunologic obstructive airway disorders
  - Asthma

Introduction to Pulmonary Impairments
Pediatric Respiratory Conditions

- Congenital Disorders - Life Care Planning Implications
  - Most common lethal congenital disorder for Caucasians
  - Thickened mucus gland secretions
  - Increased susceptibility to bacterial colonization of the airways and parasympathy
  - Pulmonary damage secondary to inflammation
  - Median survival rate 32 years (2003)
  - Multi-organ involvement – Expected complications
    - Pancreatic insufficiency and pancreatitis
    - Pseudomonas cultures
    - Diabetes mellitus
    - Recurrent respiratory infections
    - Newborn neonatal cyanosis
    - Failure to thrive/ malnutrition
    - Renal failure (between ages of 6 and 24 months)
    - Liver cirrhosis (0-7%)

Most common lethal congenital disorder for Caucasians
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Introduction to Pulmonary Impairments
Pediatric Respiratory Conditions

Cystic fibrosis - Life Care Planning Implications
- Multi-organ involvement – expected complications
  - Multinodular cirrhosis with portal hypertension progressing from biliary cirrhosis
  - Esophageal varices
  - Liver failure (rare)
  - Gallbladder disease
    - Cholangitis risk
    - Delayed puberty
    - 50% males infertile
    - 75% females infertile
    - Birth control counseling

- Recurrent nasal polyps (late childhood and early adolescence)
- Facial sinusitis (facial sinus malformation from bacterial colonization)
- Bronchiectasis (progressive dilation of bronchi)
- Atelectasis (lung collapse from obstruction)
- Pneumothorax (lung collapse from pressure change)
- Hemoptysis - Caused by damage from progressive bronchiectasis
  - Exacerbated by chronic coughing
    - Some medications known to interfere with coagulation until symptoms subside
    - May require embolization of involved blood vessels
    - 84% success rate
    - 32% recurrence
    - More serious in adults with risk of death

- Respiratory failure
  - Requiring mechanical ventilation
    - Complicated by heart failure
- Gastroesophageal reflux (GERDS)
  - Requiring barium studies
- Right sided cardiac failure
  - Survival rate generally < 1 year

Diagnostic and Long Term Preventive Treatment Considerations
- Sweat chloride testing (best)
- Antibiotic therapies
- Steroid therapies
- Bronchodilator therapy (beneficial despite study results)
- Aerosolized mucolytics (beneficial despite study results)
- Daily pulmonary physiotherapy
### Introduction to Pulmonary Impairments

#### Pediatric Respiratory Conditions

- **Cystic fibrosis** - Life Care Planning Implications

  - **Long Term Treatment Considerations**
    - Aggressive nutritional support due to increased metabolic demands by failing respiratory system and risk of anorexia
    - High protein, high calorie meals with numerous snacks
    - Oral supplementation
    - Nasogastric tube feedings
    - Intravenous feedings
    - Gastrostomy tube feedings
    - Jejunal tube feedings
    - Pancreatic enzyme supplementation (to metabolize fat and protein) prior to meals and snacks
  - **Daily respiratory treatment**
    - Inhaled medications
    - Chest physiotherapy (CPT)
    - 2 times daily for 30–60 minutes during periods of good health
    - Up to 4 times daily for 30–60 minute duration during periods of illness

- **Travel**
  - Per diem
  - Caregiver travel and per diem
  - Caregiver compensation

- **Regular exercise**
  - Recreation and leisure activities
  - Camps
  - Home equipment

- **Pneumothorax** (progressively common in adults)
  - Typically treated conservatively
  - Large pneumothoraces may require chest tube for draining
  - Chest tube drainage greater than 5–7 days may require chemical pleurodesis to seal the leak
  - Chemical pleurodesis may contraindicate future lung transplant due to risk of hemorrhage during anticoagulation

- **Lung transplantation**
  - Referral when 2 year survival rate is less than 50%
  - 51% survival at 1 year
  - 29% survival at 5 years
  - 38% survival at 10 years
Introduction to Pulmonary Impairments

Pediatric Respiratory Conditions

- **Cystic fibrosis - Life Care Planning Implications**
  - Routine Tests/Labs/Diagnostic Imaging
    - Pulmonary function studies (PFT) every 6-12 months (if old enough)
    - Pulse oximetry
    - Qualitative sputum cultures
    - Annual imaging (e.g., chest x-ray)
    - Chest x-rays every 6-12 months
    - Other diagnostic imaging (e.g., CT scans)
    - Liver function tests
    - Renal function tests
    - Nutritional/vitamin levels
    - Albumin levels
    - Fecal fat studies
    - Oral glucose tolerance annually

- Prescriptions
  - **RX** cromolyn (anti-asthmatic agents) or nedocromil (anti-allergic agents) and oral steroids more frequently
  - Pancreatic enzymes
  - GERD medications
  - Bronchodilators
  - Anti-inflammatory agents
  - Moisturizing agents
  - Oral antibiotic agents
  - Intravenous antibiotic therapy for longer duration and greater frequency (inpatient 10-14 days with exacerbations)
  - Oxygen therapy during exacerbations
    - American Academy of Pediatrics Practice Guidelines
      » On-line: http://aapgrandrounds.aappublications.org/cgi/content/extract/5/4/37
      » http://aappolicy.aappublications.org/online

- **Congenital Disorders**
  - Bronchopulmonary Dysplasia (BPD)/Chronic Lung Disease of Prematurity
    - Neonatal chronic lung disease (CLD)
    - Continued oxygen treatment at 28-days to 36 weeks after birth
    - Very low birth weight infants (VLBW)
    - Severe respiratory distress syndrome
    - Impaired surfactant (surface-active agent) production
  - Treatment
    - Requires high concentration of oxygen administered via positive pressure mechanical ventilation with high inspiratory pressures
    - Mechanical ventilation
    - Diuretic therapy
      - Furosemide (Lasix) most common
      - Requires monitoring for renal function for calcification
Introduction to Pulmonary Impairments
Pediatric Respiratory Conditions

- Bronchopulmonary Dysplasia (BPD)/Chronic Lung Disease of Prematurity
  - Treatment (continued)
    - Requires hearing monitoring for ototoxicity (auditory - VIII cranial nerve)
    - Requires monitoring of serum electrolytes with potential for potassium supplementation
    - Spironolactone (Aldactone®) in less severe cases (diuretic)
    - Cromolyn Sodium (Intal®) has anti-inflammatory effects
    - Sodium bicarbonate
    - Intravenous fluids
  - Nutrition therapy
    - Increased metabolic rate requires up to 200 kcal/kg per day
    - Calculated using the Harlan formula
    - Caloric intake must be concentrated to 30 kcal/ounce to comply with diuretics
  - Gastrostomy tube feeding for supplementary enteral feeding

Introduction to Pulmonary Impairments
Pediatric Respiratory Conditions

- Bronchopulmonary Dysplasia (BPD)/Chronic Lung Disease of Prematurity
  - Risk of hypoxemia, hypoxia, then anoxia
  - Treatment Complications
    - Oral oxygen
    - GERD
    - Subglottal stenosis
    - Tracheal scarring
    - Paralyzed vocal cords
    - Stoma infection
    - Respiratory infection
    - Tracheal tube obstruction
    - Accidental decannulation

Introduction to Pulmonary Impairments
Pediatric Respiratory Conditions

- Congenital Disorders
  - Idiopathic Congenital Central Hypoventilation Syndrome (CCHS)
    - Insensible (Child’s) Cough
    - Brain-tum involvement
    - Hypoventilation and shallow breathing during sleep
    - Able to breathe when told to do so
    - Typically able to breathe during waking hours
  - Diagnostic Work-up
    - Chest x-ray
    - Diaphragm fluoroscopy
    - Bronchoscopy
    - Electrocardiogram
    - Echocardiogram
    - Holter monitor recording
    - Brain and Brainstem MRIs
    - Serum carnitine (Vitamin B7) levels
    - Urine carnitine levels
    - Sleep (pediatric) respiratory physiology lab studies
    - Awake (pediatric) respiratory physiology lab studies
Introduction to Pulmonary Impairments
Pediatric Respiratory Conditions

- Idiopathic Congenital Central Hypoventilation Syndrome (CCHS)
  - **Treatment**
    - Positive pressure ventilation
    - Intermittent monitoring due to lack of ability to present with a normal physiological response to illness especially if mechanically ventilated
    - Telemetry
    - Heart rate
    - Hypoxemia (more CO in Blood)
    - Hyperventilation

Introduction to Pulmonary Impairments
Pediatric Respiratory Conditions

- Chronic pulmonary/immunologic obstructive airway disorders
  - **Asthma**
    - Acute inflammatory and bronchospastic response to environmental triggers within 10 to 20 minutes of exposure
    - Treated with bronchodilators
    - Delayed cyclical inflammatory and bronchospastic responses hours after exposure to environmental triggers
    - Treated with anti-inflammatory agents and bronchodilators
  - **Classifications**
    - Mild
    - Moderate
    - Severe
    - Life Threatening

Introduction to Pulmonary Impairments
Pediatric Respiratory Conditions

- Mild Asthma occurs with brief, rapidly diminishing symptoms (50%)
  - Mild intermittent asthma
  - Mild persistent asthma
  - Exacerbations are rare
  - Use of bronchodilators is infrequent
  - Rarely miss school/work days
  - Rarely limited in activities

- Moderate asthma manifested with persistent airway obstruction between acute episodes
  - Exacerbations occur at least twice weekly with persistent intervening symptoms
  - Bronchodilators are used more than twice weekly
  - Usually miss 9 school/work days per year for asthma related symptoms
  - Disrupt developmental activities (recreational)
Introduction to Pulmonary Impairments
Pediatric Respiratory Conditions

- Severe asthma manifested with persistent airway obstruction (5%)
  - Symptoms most of the time
  - Bronchodilators are used daily up to 6 months per year
  - Usually miss 20 or more school/work days per year for asthma-related symptoms

- Life threatening asthma manifested as severe asthma
  - Medications
    - Bronchodilators
    - Steroids
    - Leukotriene modifiers
    - Omega-3 fatty acids
  - Significant risk of dying from a severe episode

- Treatment: Identify and control exposure to triggers
  - Allergens
  - Exercise
  - Irritants
  - Weather changes
  - Infections
  - Gastroesophageal reflux
  - Endocrine factors
  - Emotional factors
  - Age <5 and >5

Introduction to Pulmonary Impairments
Pediatric Respiratory Conditions

- Chronic pulmonary/immunologic obstructive airway disorders
  - Medications
    - Long Term Control
      - Corticosteroids
      - Cromolyn sodium
      - Nedocromil (Tilade)
      - Long acting beta2 agonist
      - Methylxanthines
      - Leukotriene inhibitors (inhalation anti-allergens)
      - Montelukast (Singulair)
    - Quick Relief
      - Short acting beta2 agonist
      - Anticholinergics
      - Systemic corticosteroids

Pulmonary Rehabilitation

- A "multi-disciplinary program of care for patients with chronic respiratory impairment that is individually tailored and designed to optimize physical and social performance and autonomy.”
  - American Association for Respiratory Care (AARC). AARC clinical practice guideline: pulmonary rehabilitation. Dallas (TX): American Association for Respiratory Care (AARC); 2002. 9 p.
  - In-patient
  - Out-patient
  - Minimum of two months
Pulmonary Rehabilitation

• Based on the individualized assessment the following areas of education and training should be considered:
  - Pulmonary anatomy and physiology including the pathophysiology of lung disease
  - Description and interpretation of medical tests
  - Bronchial hygiene techniques
  - Exercise conditioning and techniques which include:
    - Breathing retraining
    - Endurance, strength, and flexibility training
    - Upper extremity
    - Lower extremity
  - Ventilatory muscle training (its role is still undetermined, since no evidence exists that it contributes to functional improvement when added to a traditional upper and lower extremity exercise training program)
  - Energy conservation as it applies to activities of daily living
  - Indications, actions, and side-effects of medications including non-prescription products, such as vitamins, over-the-counter medications, and herbal remedies
  - Functional self-management

Pulmonary Rehabilitation

- Self-assessment and symptom management
- Infection control with emphasis on avoidance, early intervention, and immunization
- Environment control
- Indications for seeking additional medical resources
- Sleep disturbances, e.g., insomnia and sleep apnea as they relate to chronic lung disease
- Sexuality and intimacy
- Nutrition
- Smoking cessation
- Psychosocial intervention and support
- Available community services, including patient/family support groups
- Advance care planning
- Travel issues
- Recreation/leisure activities
- Stress management
- Indications for oxygen, and methods of delivery

Pulmonary Rehabilitation

• Program schedules vary according to staff, facilities, resources, budget, and patient needs.
  - Pulmonary rehabilitation services are commonly provided over a period of 12 hours per week for 6 or more weeks, governed by the patients individual needs.
  - Patients are encouraged, when possible, to participate in an ongoing maintenance exercise program to sustain the training effect.
Pulmonary Rehabilitation

- Treatment team composition
  - CFCC
  - Pediatrician/PCP for routine preventive care and monitoring
  - Routine vaccinations (including influenza and pneumococcal)
  - Pediatric/adult pulmonologist and/or intensivist
  - Pediatric/adult otorhinolaryngologist
  - Pulmonary nurse clinician
  - Respiratory therapist
  - Registered dietitian/nutritionist
  - Physical therapist
  - Exercise physiologist
  - Occupational therapist
  - Medical social worker
  - Home health care agency
  - Case manager
  - DME dealer

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Bibliography


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• The American Academy of Pediatrics Practice Guidelines Respiratory syncytial virus (RSV) prophylaxis: American