

RESPIRATORY SYSTEM

The mission of BDSRA is:

To be an international support
and research networking
organization for families
of children and young
adults
with an
inherited
neurological
degenerative
disorder known
as Batten Disease.

“A light in a world of darkness...”

WELCOME

NOTICE TO THE READER

This Batten Disease Handbook is compiled with information from many sources concerning the topics included. Families in the organization have also contributed their specific situations that have been helpful in their own battle with Batten Disease. All material in this book is provided for information purposes only.

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Nancy Carney, RN
Batten Disease Support and Research
2002

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The Respiratory System

The pulmonary and circulatory systems are designed to provide the body with a continuous supply of oxygen and quick removal of carbon dioxide. The pulmonary system controls the exchange of gases between the atmosphere and blood, while the circulatory system transports these gases between the lungs and the cells. A dysfunction in either system disrupts homeostasis, can cause anoxia (an inadequate level of oxygen) and even cell death.

The organs used in the exchange of gases between the atmosphere and the blood are: the nose, pharynx, larynx, trachea, bronchi, and lungs. The trachea branches into primary bronchi, secondary bronchi, the bronchioles, terminal bronchioles, and finally the alveolar sacs, which are the functional units of the lungs (see diagram on page 6). Ventilation is the movement of air between the atmosphere and the alveoli. Diffusion is the process by which oxygen and carbon dioxide cross the alveolar capillary membrane and perfusion is the injection of blood into an artery that supplies blood to an organ or tissue. Lung problems go hand in hand with the progressive feeding problems that develop, in most children, with neuro-degenerative diseases. As these children begin to have difficulty swallowing, their gagging and coughing can lead to “mini” or “major” aspirations (inhaling liquid or solids into their lungs). This can cause wheezing, which a whistling sound is made by air going through a narrow passageway. These children develop increasing amounts of mucus in their lungs resulting in congestion or can lead to pneumonia. The lungs have a protective mucus lining that catches tiny pieces of dirt, dust and other particles in the air we breathe. These particles would irritate the lungs or cause infection if they stayed in the lungs. Then the cilia (tiny hairs), acting like an escalator, carry the mucus up the windpipe to be coughed up or swallowed. What color is mucus? Mucus can be yellow, green, rust colored, brown color, pink, or frothy white. What does the cough sound like? It can be dry, barking, hacking, or congested. When does he cough? Is it early morning, late afternoon, or evening? If the lungs become irritated or infected, a large amount of mucus is produced, because the lungs are working extra hard to get rid of the irritation or infection, and pneumonia likely develops.

Primary Controls

The following mechanisms are the primary controls of respirations: the nervous system adjusts the rate of respirations to satisfy physiological demands; and the respiratory center and the brain (which consists of the medulla oblongata, and the pons), direct the contraction and relaxation of respiratory muscles (see diagram page on page 8).

The Hering-Breuer reflex controls the depth of respiration and prevents over inflation of the lungs. This reflex occurs in response to nerve impulses transmitted from stretch receptors in the bronchi and bronchioles to the respiratory center in the brain. Carbon dioxide, oxygen, and hydrogen ion concentration determine the rate of respirations by acting directly on the respiratory center in the brain or on chemo

receptors located in the carotid arteries in the aorta. Together, these control mechanisms keep blood oxygen and carbon dioxide levels remarkably stable.

Acute Changes Demand Quick Detection

The two vital functions of the respiratory system are: maintenance of oxygen and carbon dioxide exchange in the lungs and tissues, and regulation of the acid base balance. Any changes in this system affect all the other body systems. In chronic respiratory disease, pulmonary changes (such as hypoxia) occur slowly, and a person's body has time to adapt. But with acute pulmonary changes, such as those from a pneumothorax (air in a lung which requires a chest tube to reinflate the lung) or aspiration pneumonia, the patient's other body systems do not have time to adapt to sudden hypoxia, which can cause death.

Changes in other body systems may reduce capabilities to provide oxygen. For example, a child's poor cardiac function results in decreased tissue oxygenation, which causes his lungs to work harder to provide oxygen. In fact, any acute disease state increases the body's oxygen demand and the lungs workload. Also, debilitation from acute disease makes the child more susceptible to secondary infections, which may affect his lungs.

A child with a less serious illness is not immune from pulmonary complications. For instance, simple post anesthesia or hypoventilation can develop into atelectasis, that, if untreated, can cause a devastating pneumonia or antibiotic therapy for a minor infection can alter the child's normal flora in the lungs and allow virulent pneumonia to develop. Regardless of their cause, changes in a child's lung function can result in acid base disturbances, tissues hypoxia, and even sudden death. Using correct techniques, you can detect changes in a child's respiratory system early, and intervene quickly, perhaps preventing serious complications.

The Upper Airways

The upper airway's including: the nose, the mouth, the nasopharynx, the oropharynx, the laryngopharynx, and the larynx. The two portals for air are the nose and the mouth. The nasal openings for air passage are the nares or nostrils. Divided by the nasal septum at the midline, the two separate nasal passages are formed anteriorly by cartilage walls and posterior by bony structures. Covered with mucus membranes, they form additional surface areas over which air is humidified and warmed as it passes into the nasopharynx. Anteriorly, the nose hairs trap dust and large particles; posterior, mucus traps finer particles, which the cilia carry to the pharynx to be swallowed. Posterior, the olfactory epithelium overlies the superior concha, an upper portion of the septum. The superior portion of the hard pallet forms the nasal floor. The paranasal sinuses provide speech resonance. The maxillary and the frontal sinuses are large, paired, air filled cavities. The sphenoid and the ethmoidal sinuses, also covered with mucus membranes, consist of several small air sacs in the nasal cavities bony posterior portions. The nasopharynx has lateral and posterior mucosal walls. Air passes from the nasal cavity into the nasopharynx, through the choanae, which are always open. The pharyngeal tonsils (adenoids) are located bilaterally near the choanae. In the lateral

walls above the soft pallet level are the eustachian tube openings. The eustachian tubes regulate the middle ear pressure during swallowing or yawning.

The oropharynx is the posterior wall of the mouth that connects the nasopharynx with the laryngopharynx, the lowest pharyngeal region, which stretches down to the esophagus.

The larynx, which contains the vocal cords, is a cartilaginous and muscular organ connecting the pharynx and trachea. Two of the nine principal cartilages, the large, shield shaped thyroid cartilage or the “Adams apple” and the cricoid cartilage, inferior to it, can be palpated in the neck.

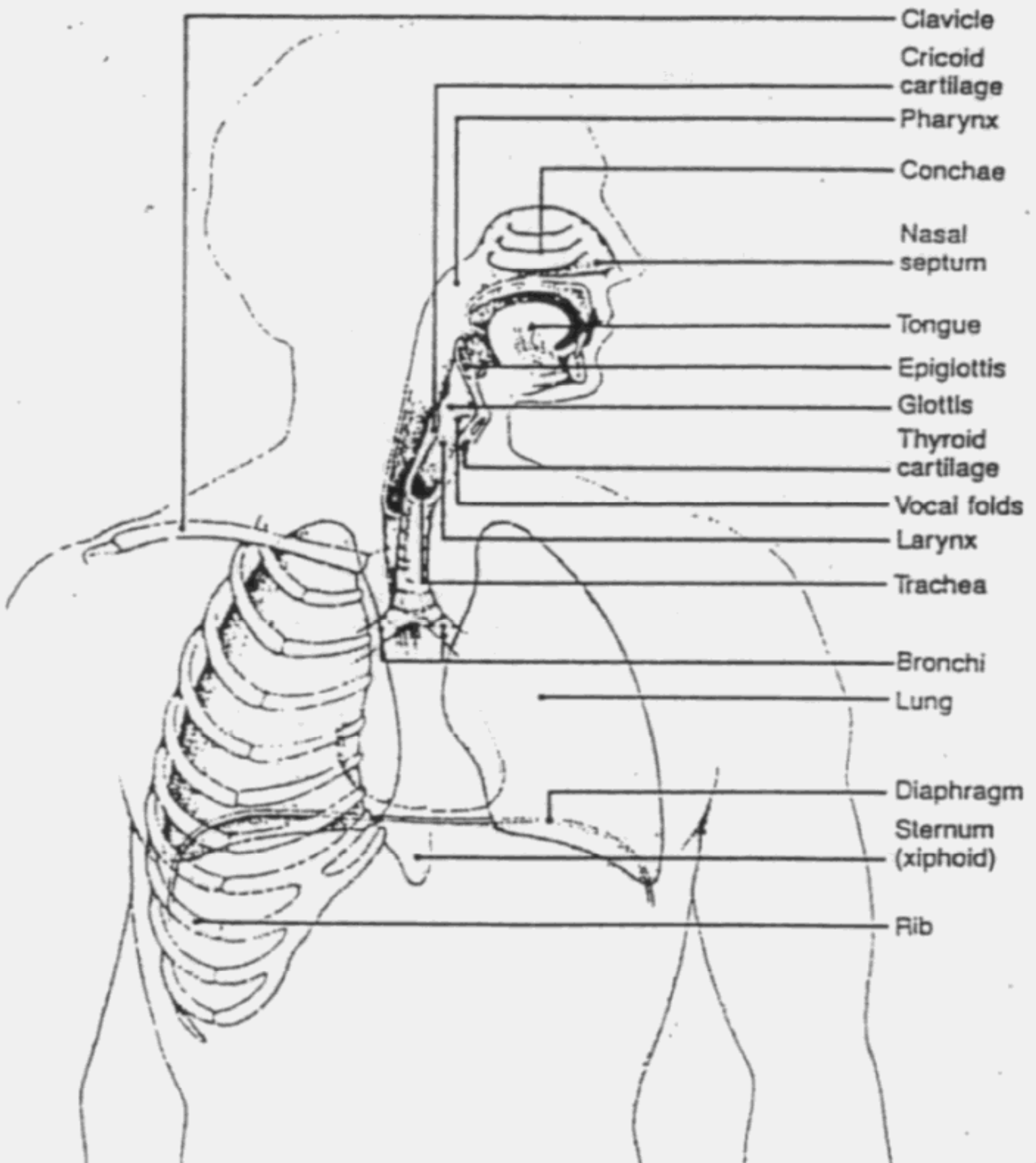
Formed by a flexible cartilage the epiglottis bends reflexively on swallowing, closing off the larynx to swallow substances. The inner laryngeal structure's, include the vestibule, the glottis, the vocal folds, and the infraglottic cavity. The vestibular folds (false vocal cords) are thick mucosal folds located just below the vestibule and superior to the true vocal cords. The opening between the folds is called the glottis. Located posteriorly, the small-paired arytenoid cartilages act as a fulcrum for vocal cord muscles (see diagram on page 4).

The Lower Airways

The lower airways begin at the top of the trachea, which is a tubular structure that extends about five inches from the only complete tracheal ring, the cricoid cartilage to the carina at the level of the sixth or seventh thoracic vertebrae. C-shaped cartilage rings with posterior openings, reinforce and protect the trachea, preventing its collapse. The isthmus of the thyroid covers the upper two or three rings. About half of the trachea is in the neck and half is in the thorax; the right and the left primary bronchi (main stem bronchi) begin at the carina, or tracheal bifurcation. Composed of tissues similar to that of the trachea, the walls of the primary bronchi are lined with epithelium containing cilia, goblets (mucus-producing cells), and microvilli that increase surface area absorption. The right primary bronchus is shorter, wider, and more vertical than the left.

The primary bronchi divide into the secondary or lobar branches and accompanied by blood vessels, nerves, and lymphatic, as they enter the lungs at the hilum. Each of the five secondary branches, the right upper, middle, and lower, and the left upper and lower, enters its own lung lobe. These branches further divide into the tertiary or segmental bronchi, which supply air to the eighteen bronchopulmonary segments. From the tertiary bronchi, branching continues and the airway becomes narrower, cartilage decreases, and smooth muscle increases. The functional units of the lungs, the lobules, are supplied with air from the small airways (the terminal bronchioles), with an average diameter of 0.5mm. The terminal bronchioles divide into two or more respiratory bronchioles, which are the entrance to the lobules. These passages continue to branch off, terminating in alveolar ducts, which lead to walled alveoli surrounded by a network of anastomosing capillaries. Alveoli cells are simple squamous epithelium, as well as, type II cells. Type II cells are capable of secreting surfactant, which reduces surface tension and enhances alveolar elasticity. Also,

THE UPPER AND LOWER AIRWAYS



elastic fibers surround the alveoli and allow them to recoil to preinspiratory size (see diagram on page 4).

Thoracic Cage, Lungs, and Pleurae

The anterior boundary of the thoracic cage, which contains the lungs and the pleurae, is the sternum. Its posterior boundary is formed by: the thoracic vertebra, the anterior boundary by the costal margins, the xiphoid process, and the diaphragm.

The right lung has three lobes and is larger than the left, which has two lobes. The medial inferior surface of the left lung curves slightly around and under the heart and forms a tongue like structure called lingula pulmonis sinistae. The inferior surfaces of both lower lobes are separated from the abdominal viscera by the diaphragm. The apices of the two upper lobes are slightly above the first rib about one inch above the clavicles. The lateral boundary of both lungs is the chest wall.

The visceral pleura, a serous membrane adhering closely to the lungs parenchyma, envelopes each lung and separates it from the mediastinal structures, the heart and its great vessels, the trachea, the esophagus, and the bronchi. The parietal pleura, which line the thoracic cavity from the hilum, cover all areas that contact the lungs. The visceral pleura meet the parietal pleura at the hilum, forming a narrow fold called the pulmonary ligament. These membranes consist of connective and epithelial tissue and a single layer of secreting epithelium. As they rub together during respiration, secretions minimize friction. The area between the two membranes is only a potential space and cannot be seen unless air or excess fluid occupies it. Normally pleural fluid maintains the surface tension forces.

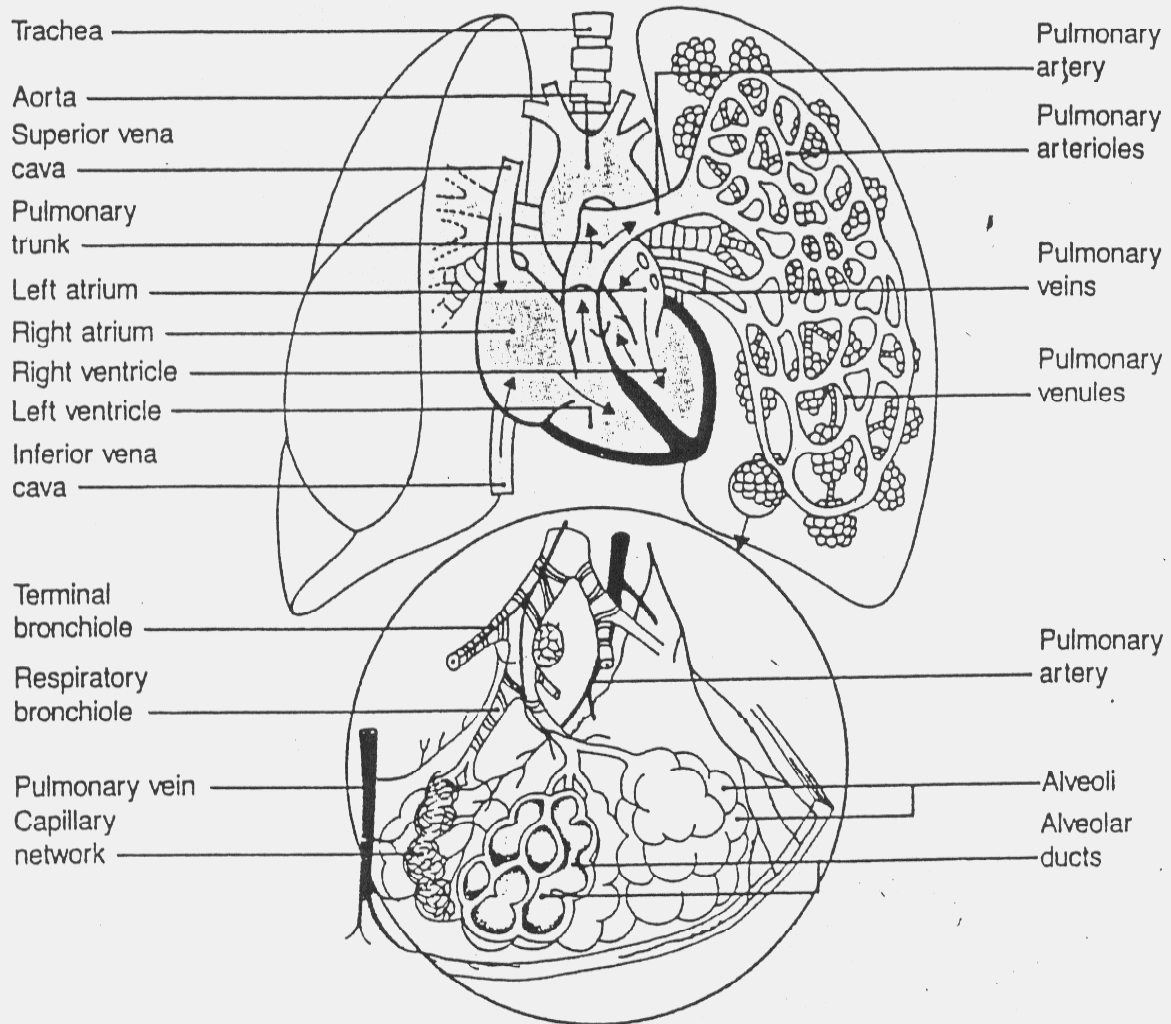
Pulmonary Blood Supply

Oxygen-deficient blood is pumped from the right ventricle of the heart to the pulmonary trunk. This trunk then branches laterally into the right and the left pulmonary arteries, which further divide into smaller arteries that closely follow the bronchial tree airways throughout the lungs. Eventually, pulmonary arterioles enter the lungs lobules, where arterioles and the venules form the capillary beds around the alveoli. This is where gas exchange occurs. The venous system then returns oxygenated blood to the left atrium at the heart to be pumped throughout the body. The tissues of the airways and pleura receive blood from bronchial arteries, which arise from the aorta and its branches (see the diagram on page 6).

Gas Exchange

Diffusion, also called external respiration, is the exchange of gases between the alveoli and the capillaries. In this process, oxygen diffuses across the alveolar epithelium, the epithelial basement membrane, the capillary basement membrane, and the capillary endothelial membrane. It then dissolves in plasma and passes through the red blood cell membrane. Carbon dioxide diffuses in the opposite direction. Oxygen moves from the alveolus into the venous end of the capillary because oxygen pressure (pO_2) is greater in the alveolus; carbon dioxide diffuses from the venous end of the capillary, where carbon dioxide pressure (pCO_2) is greater to the alveolus. Each gas acts

UNDERSTANDING THE PULMONARY BLOOD SUPPLY



independent of the other because of their distinctive partial pressure. A number of factors affect the rate of gas exchange, including drug use, geographic altitude, and the extent of the person's functional lung surface area.

Grape-like clusters of alveoli are the sites of gas exchange in the lungs. Each alveolus is served by two systems: the capillary network that transports mixed venous blood to the alveolar membrane and the tracheobronchial tree (trachea, bronchi, and bronchioles) that delivers air to the alveolar space. When the venous blood passes through the alveolar membrane, it releases carbon dioxide and takes in oxygen. Then, the venous blood pressure travels to the heart. From there, it circulates throughout the body, releasing oxygen and taking in carbon dioxide and cellular waste before returning to the lungs.

Hemoglobin, which is normally about 98 percent oxygen-saturated, transports oxygen to the tissues for gas exchange between body cells and red blood cells. This exchange is called internal respiration. Because increase temperature and decrease pH trigger oxygen release from hemoglobin, only tissues needing oxygen receive it. Blood also carries carbon dioxide to the lungs, primarily in the form of bicarbonate.

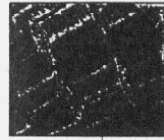
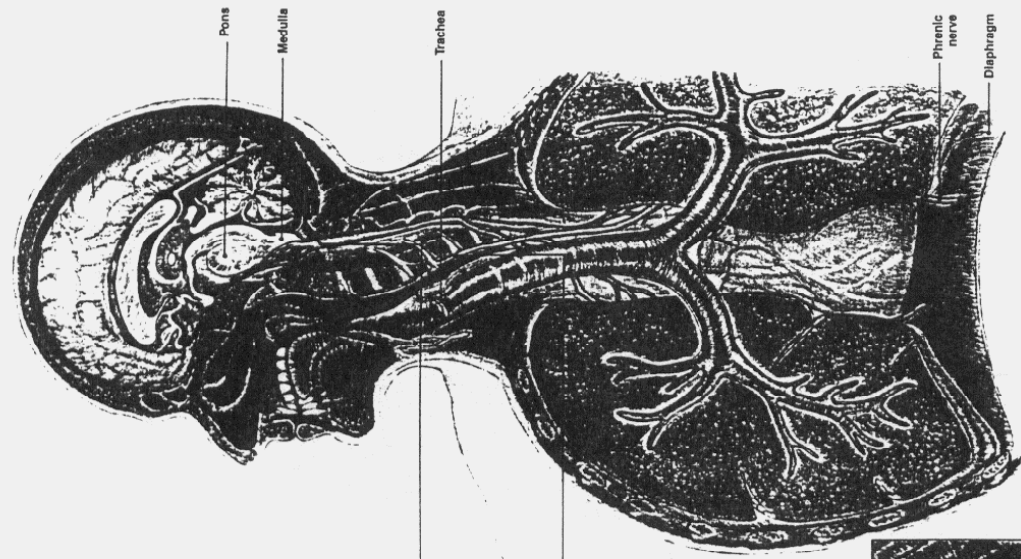
Mechanics of Respiration

Voluntary and intracostal muscles, working with the diaphragm, produce normal inspiratory and expiratory movement of the lungs and the chest wall. The lungs tendency to recoil inward, balanced by the chest walls tendency to spring outward, creates a subatmospheric pressure in the closed pleural cavity. This pressure, which is about -5 mm below atmospheric pressure, makes lung ventilation (gas volume movement) possible.

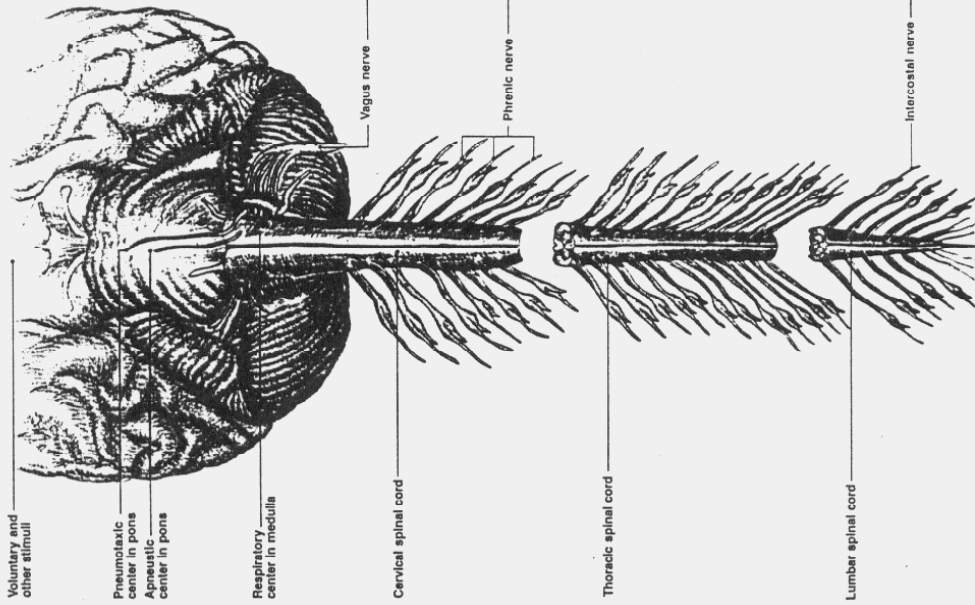
Neurologic Control

Innervation of pulmonary structures varies according to their function. The phrenic nerves, from the third to the fifth cervical vertebra, innervate the mediastinal and central diaphragmatic pleura and the diaphragm. Intercostal nerves innervate the costal and peripheral diaphragmatic pleura and intercostal muscles. Vagus nerve branches innervate the larynx. Visceral sensation through these vagal fibers is primarily limited to stretch, although some vagal (sympathetic) fibers are motor fibers of the bronchial trees smooth muscle. In addition, other sympathetic fibers regulate vasoconstriction of the pulmonary arterioles. The respiratory muscles respond to motor impulses from the respiratory center in the brain stem. The posterior medulla oblongata is functionally divided into inspiratory and expiratory centers. The apneustic center of the pons constantly stimulates neurons in the inspiratory center. The expiratory center inhibits inspiration, there by providing time for muscle relaxation. Impulses travel from the inspiratory center to the respiratory center (by way of the phrenic nerve) and the pneumotaxic center of the pons. After a slight delay, impulses move from the pons to the expiratory center. Thus, the pons is the pacemaker that regulates the rhythm of respiration; the medulla regulates rate and depth (note diagram on page 8). The Hering-Breuer reflex also regulates respiratory excursion. Lung expansion stimulates the stretch receptors that send impulses along the vagus afferent fibers to the expiratory

NEURAL CONTROL OF RESPIRATION



Cross section of intercostal muscle



center, thus inhibiting inspiration. When the lungs deflate, the stimulus stops and inspiration again dominates.

Central chemoreceptor, located in the anterior medulla responds to changes in pH, oxygen tension (pO₂), carbon dioxide tension (pCO₂), and peripheral chemoreceptors. These anterior neurons are particularly sensitive to the pH changes in the cerebral spinal fluid. A decrease in the cerebral spinal fluid quickly stimulates alveolar stimulation. The most active chemical stimulus is the CO₂ tension of arterial blood. A small increase in inspired CO₂ (or hydrogen ion concentration) decreases the pH causing quick medulla stimulation, which results in hyperventilation. The peripheral chemoreceptors (the aortic and carotid bodies) sense changes in the oxygen tension and directly stimulate the central chemoreceptors (Of course, respirations can also be partially controlled voluntarily).

Assessment of lungs-collecting appropriate history and data

Chief Complaints and History of Present Illness: The biographical data is important when accessing a child's respiratory system, remembering that his age and sex can affect his thoracic configuration. When you note the area where a child lives and what his lifestyle is, be alert of possible environmental or occupational hazards that can affect his lungs and breathing. The most common chief complaints for respiratory disorders are: cough (with or without sputum production or hemoptysis, which is blood in the sputum) dyspnea and chest pain. Next, you need to ask the child or parents to elaborate on his chief complaint. Then, depending on the complaint, explore the history of his present illness by asking the following type of questions.

Cough - does your cough usually occur at a specific time of day? How does it sound - dry, hacking, barking, congested? Try to determine whether the child's cough is related to second hand smoke or irritants. (The most common causes of coughing are smoke and chronic bronchitis.) Please describe any medication you are using for treatment of your child in receiving to clear the cough. How frequently does he take the medication or receive treatment? Have you recently been exposed to anyone with a similar cough? Was this person's cough caused by a cold or flu?

Sputum production: How much sputum are you coughing up per day? Remember, the tracheobronchial tree can produce up to three ounces of sputum, which is 90 cc or 3 ounces per day. What time of day do you cough up the most sputum? Smokers cough when they get up in the morning; non-smokers generally do not. Coughing from irritants occurs most often during exposure to it, for example at work. Is sputum production increasing? This may result in external stimuli or from such internal causes as chronic bronchial infection or a lung abscess. Excess production of sputum that separates into layers may indicate bronchiectasis. Does the sputum contain mucus or look frothy? What color is it? Has the color changed? Does it smell bad? Foul smelling sputum may result from an anaerobic infection, such as an abscess. Blood tinged or rust color sputum may result from trauma caused by coughing or from such underlying pathology

as bronchitis, pulmonary infection or infection, tuberculosis, and tumors. A color change from white to yellow or green indicates infection.

Dyspnea: Are you always short of breath or do you have attacks of breathlessness? Onset of dyspnea may be slow or abrupt. For example, a child with asthma may experience acute dyspnea intermittently. What relieves the attack: positioning, relaxation or medication? Do the attacks cause your lips and nail beds to turn blue? Does body position, time of day, or a certain type of activity affect your breathing? Paroxysmal nocturnal dyspnea and orthopnea are commonly associated with chronic lung disease, but may be related to cardiac dysfunction. How many stairs can your child climb or blocks can you walk before you begin to feel short of breath? Do such activities as taking a shower or shopping make you feel this way? Dyspnea from activities suggests poor ventilation or perfusion, or insufficient breathing mechanisms. Do you experience associated signs and symptoms, such as cough, diaphoresis, or chest discomfort? Does the breathlessness seem to be stable or getting worse? Is it accompanied by external sounds, such as wheezing or stridor? Wheezing sounds result from small airway obstructions (for example, from aspirated foreign body, a tumor, asthma, or congestive heart failure). Stridor results from tracheal compression or laryngoedema.

Chest pain: Is the pain localized? Is it consistent or do you experience attacks? Have you ever had a chest injury? Does a specific activity such as movement of the upper body or exercise produce pain? Chest pain may be associated with cardiovascular disorders, but respiratory disorders usually cause musculoskeletal chest pain (the lungs have no pain-sensitive nerves). However, the parietal pleura and the tracheal bronchial tree are sensitive to pain. Do other signs and symptoms such as coughing, sneezing, or shortness of breath accompany the pain? Does the pain occur when you breathe normally, or when you breathe deeply? This distinction is important in determining whether your child's pain is pleuritic. Does splinting relieve the pain?

As parents, try and fit these symptoms into what your child/children are expressing by verbalization, grimacing, amount of coughing or sputum he/she is producing and/or any choking, aspiration or shortness of breath that you may observe.

Past History

Focus on the following body systems, procedures, and conditions when reviewing your child's past history: *Respiratory system:* Ask the parents if their child has ever had pneumonia, pleurisy, asthma, bronchitis, emphysema, or tuberculosis. Ask how often he gets a cold. *Cardiovascular system:* Ask the parents if their child has ever had high blood pressure, heart attack, or congestive heart failure. A history of such a disorder is particularly important because of the close relationship between the cardiovascular system and the respiratory system. Ask the parents if their child has had any chest surgery, invasive medical procedures, such as a bronchoscopy, if there are any chest deformities (any congenital or trauma related or related deformities may distort cardiac and pulmonary structures), if he has had any lab tests and the results of his last chest x-ray, pulmonary function tests, blood gases, EKG, sputum cultures or skin tests for TB.

Ask the parents if their child is allergic to any medications, food, pets, dust, or pollen. Chronic allergies may predispose him to other respiratory disorders. Has he ever been treated for an allergy? Ask about medications. Does he take medications or over the counter drugs for cough control, expectoration, nasal congestion, chest pain, or dyspnea. And vaccinations, has the child ever been vaccinated against pneumonia, or flu.

FORMING NURSING DIAGNOSES

Chief Complaint

1. Dyspnea

Acute – diaphoresis; restlessness, nursing diagnosis – alterations in comfort

Chronic - Barrel chest; accessory muscle change, nursing diagnosis – ineffective breathing pattern

Acute and chronic – fatigue, exhaustion, emotional distress, hypoventilation/hyperventilation (may lead to respiratory acidosis/alkalosis), nursing diagnosis – fear of breathlessness, ineffective breathing pattern, impaired gas exchange, impaired physical mobility

2. Chest Pain

- Decreased ventilation (may lead to infection or pneumonia), nursing diagnosis – impaired gas exchange, ineffective breathing pattern
- Increased CO₂ retention and respiratory acidosis
- Discomfort, pain, nursing diagnosis – fear of chest pain alterations in comfort

3. Cough

Chronic and short term (less than 1 month)

- Hazardous elevation in intrathoracic pressure, intracranial pressure, and blood pressure (may lead to congestive heart failure, ruptured aneurysm, nursing diagnosis – alteration in comfort)
- Cough, syncope
- Musculoskeletal pain, fractured ribs – nursing diagnosis – alteration in comfort
- Fractured ribs

Chronic and long term (more than 1 month)

- Fatigue
- Weight loss, anorexia, nursing diagnosis – alteration in nutrition (less than body requirements)

Forced Cough

- Collapsed airways (atelectasis), nursing diagnosis – impaired gas exchange
- Rupture of thin-walled alveoli (may lead to pneumothorax)
- Hemoptysis, second-degree irritation of tracheobronchial tree, nursing diagnosis – fear of seeing blood

4. Increased and abnormal secretions

Increased sputum

- mucous plugs (may lead to airway obstruction, atelectasis, prevention of

alveoli gas exchange, hypoxia, respiratory acidosis, nursing diagnosis – ineffective airway clearance, impaired gas exchange

- increased secretions and/or abnormal fluids retained in lung (may lead to infection, tracheobronchitis, bronchopneumonia)

Hemoptysis

- Obstruction with blood (may lead to asphyxiation, atelectasis, pneumonia, nursing diagnosis – ineffective airway clearance, impaired gas exchange, alterations in tissue perfusion)
- Blood-streaked sputum, severe blood loss (may lead to shock), nursing diagnosis – fear of seeing blood

Family history

When reviewing the child's family history, ask if anyone in the family has ever had asthma, cystic fibrosis, or emphysema, all of which may be genetically transmitted. Other important disorders to ask about include lung cancer and infectious diseases such as tuberculosis. Also inquire about chronic allergies in the family, cardiovascular disorders (hypertension, myocardial infarction, heart attack), and congestive heart failure), and respiratory disturbances (such as frequent cold, flu, pneumonia, asthma, or emphysema). Disorders involving other body systems may be associated with pulmonary dysfunction, so ask about a family history as such other conditions as kyphosis, scoliosis, obesity, and neuromuscular dysfunction. Scoliosis is a lateral curvature of the spine that may be found in the thoracic, lumbar, or thoracolumbar spinal segment. The curve may be convex to the right (more common in thoracic curves), or to the left (more common in lumbar curves). Rotation of the vertebral column around its axis occurs and may cause rib cage deformity. Symptoms include backache, fatigue, and dyspnea. Untreated scoliosis may result in pulmonary insufficiency (curvature may decrease lung capacity), back pain, degenerative arthritis of the spine, disk disease, and sciatica. By the survey we are currently conducting, many of our children with Batten Disease have reported some degree of scoliosis. As we continue to receive more surveys, we will compile them and be more specific as to percentage of our children, etc.

Environmental and Psychological History Ask questions on home conditions as far as air pollutants, any chemical factories close by may develop respiratory disorders. Exposure to cigarette smoke in the home may aggravate respiratory symptoms. Crowded living conditions facilitate the transmission of communicable respiratory diseases. What about home or school exposure to cigarette smoke or other substances that may be irritating to the respiratory system may be significant? What about pets? Exposure to animals may precipitate an allergic or asthmatic attack. Hobbies, seemingly innocent past times, such as building model airplanes or refinishing old furniture may expose the patient to harsh chemical irritants. Stress: some respiratory conditions such as asthma or infection can be aggravated by stress.

Activities of daily living

When reviewing your child's activities of daily living, ask if he is around second hand smoke. Learning about a child's smoking environment is vital to completing a

comprehensive respiratory history. Smoking can be associated with numerous and varied pathology, such as lung cancer, chronic bronchitis, and emphysema. The risk of lung disease is higher among smokers exposed to respiratory irritants either near their homes or on the job. So when asking the child/parent about his daily activities, be especially alert to a history of exposure to chemicals, noxious fumes, chromium, and dust containing nickel, uranium, or asbestos. Your child's daily routine is also important, because respiratory signs and symptoms can interfere with such activities as climbing stairs, or other daily activities.

Child preparation and positioning for a physical assessment

You want to be able to have the child in an upright position so he can take a deep breath and you will be able to hear his lungs and the bases to know if there is any kind of pneumonia's particularly beginning.

Quick Observation of the Respiratory Status: You want to just take a quick observation of the respiratory status. Observe for signs and symptoms or maybe a severe hypoxia, which is not enough oxygen in the system and any acute labored breathing. Look at his level of consciousness; shortness of breath when speaking; rapid, very deep, very shallow or depressed respirations; use of accessory muscles when breathing, like the lower abdominal muscles; any sternal retractions, which means, does the chest bone in the middle go in instead of protruding out just a little bit; has he turned blue; is he crowing; wheezing; or having a stridor type sound in breathing; is he diaphoresing, which means is he sweating; does he have nasal flaring, or does it like flare out; does he seem extremely apprehensive or agitated? A child exhibiting most or all of these signs and symptoms require immediate intervention. Position him appropriately to relieve the stress and notify a doctor or a squad.

Inspection: The second step in assessment is inspecting the child's skin, nose, mouth, ear, trachea, and posterior and anterior chest. What you are looking for here is color of his face, possibly his nail beds, are they blue? Look when he is breathing, is each side of his lungs inflating and deflating at the same height and depth? Is he having any pain? Does he seem extremely anxious, in which you would be able to see in his facial expressions? Is there anything to indicate a possible obstruction of something? Is there something in his throat that is causing an obstruction for the increased air and labored respirations? Does he have a moist sounding cough, or is he wheezing?

Next, we are going to listen to his lung sounds if you have a stethoscope available. What you are going to be listening for are rales or crackles. This is going to be on inspirations, as they are taking a breath in, do you hear like little crackles or little bubbles or any sounds in his lungs? That most likely will indicate fluid in the lungs. Or are you hearing rhonchi, which sound more like a wheeze, but it is when they breathe out on expiration, you will hear a noisy sound and that does not usually indicate that there is any kind of fluid in there. Or is it specifically a wheeze, which can be heard on inspiration or expiration as what you would notice in an asthmatic child especially. Refer to the descriptions below.

Chest Sounds Heard on Auscultation

Rales – heard anywhere in the lungs, first with pulmonary edema (where there is fluid in the lungs), usually during the inspiratory phase, which are caused by air passing through moisture, especially in the small airways and alveoli – they sound like light crackling, popping, nonmusical, can be further classified by pitch –high, medium or low.

Rhonchi – heard in the larger airways, usually during the expiratory phase caused by fluid or secretions in the large airways or narrowing of the large airways – you hear coarse rattling, usually louder and lower pitched than rales; can be described as sonorous, bubbling, moaning, musical, sibilant, and rumbling.

Wheezes – may occur during inspiration or expiration, usually caused by narrowed airways, sounds like a creaking, groaning, always high – pitched, and musical squeaks

Pleural Friction Rub – at the anterolateral lung field, on both inspiration and expiration (with the patient in an upright position, caused by parietal and visceral pleural linings rubbing together – can hear superficial squeaking or grating.

Children's respiratory system vs. adults

The child's developing pulmonary system makes him more susceptible than an adult to certain respiratory disease. For instance, respiratory tract infections commonly occur in children because a child's respiratory tract is immature, and the mucous membranes often cannot produce enough mucus to warm and humidify the air. Also, a child's developing immune system cannot fight bacteria and viruses as well as adults can. Because the lower respiratory tract is small in infants and children, breath sounds are louder and more bronchial, expiration is longer, and vesicular sounds are harsher. The infant's thorax is round, with equal anteroposterior transverse diameter. The lateral diameter increases rapidly with growth, resulting in characteristic adult portions by age six. (Thin chest wall and lack of muscles allow palpitation of the floating ribs in young children).

Infants are nose breathers, which is one reason why colds are more serious for them. A child's breathing is primarily abdominal until the age of six or seven (longer if the child has done breathing exercises for singing or athletics). Abdominal breathing beyond this age may indicate pain or spinting of the chest walls as in pleuritis; respiratory movements before this age suggest pain or spinting of the abdomen as in peritonitis. A child's respiratory rate may double in response to exercise, illness, or emotion. Normally, the rate for newborns is 30-80 breaths per minute, for toddlers 20-40; and for children of school age and older, 15-25. Children usually reach the adult rate of 12-20 at about age 15.

Ask the parents how often the child has had upper respiratory tract infections.

Remember that a history of more than six nose or throat infections a year necessitates further evaluation of the child, because colds in preschool children are often a sign of streptococcal infection. Find out if the child has had other respiratory signs and symptoms, such as dyspnea, wheezing, runny nose, or a stuffy nose. Ask if these appear related to the child's activities or to seasonal changes.

Special pediatric history considerations

Ask if the child has had a cough that interrupts his sleep or causes vomiting. If so, does it produce sputum? Is the sputum blood-tinged? Ask if anyone in the family has ever had cystic fibrosis or other major respiratory diseases, such as asthma.

Examining a child's respiratory system

Positioning a child for a respiratory examination depends, of course, on his age, condition, and disposition. The sitting position offers you easiest access to his thorax, and usually a parent can help by holding the child in his lap. If the child is quiet, auscultate his lungs first. If you hear fluid, place the stethoscope's diaphragm over his nose to determine if the fluid is in the nose or upper respiratory tract. This is important in children, because the sound of fluid in the nose can be transmitted through the short distance between the nose and the lungs.

To examine the child's nostrils for patency, occlude one, put the stethoscope's diaphragm over the other, listen and watch for condensation on the diaphragm. With infants and young children, perform this procedure (which may provoke crying) after auscultating the lungs, because crying can cause an unnatural respiration rate and interfere with breath sound auscultation. Also, crying usually elicits mouth breathing, which can make determining the nostril's patency difficult. To quiet a crying child and relax his breathing, hand him a plastic windmill and ask him to blow on it, or have him pretend to blow out a candle.

The procedure you should use for inspecting the child's mouth and throat also depends on his age and disposition. Position the infant on his back and ask the parent to hold him still. A child age 6 or older will probably sit on an examining table without restraint (to ease his anxiety, you might allow him to handle the equipment).

Use a flashlight and tongue depressor to examine the child's mouth and throat. You can also use the tongue depressor to elicit the gag reflex in infants, but remember, you should never test this reflex or examine the pharynx in a child suspected of having epiglottitis, because these procedures can cause complete laryngeal obstruction, which could be fatal. While examining the posterior thorax of the older child, be sure to check for scoliosis. If you observe an abnormality, refer him to treatment.

Diagnosing childhood respiratory disorders

You may see croup – the most common cause of respiratory distress in children over age 3 – in a child with a history of upper respiratory tract infections, a hacking cough, fever, stridor, and diminished breath sounds with rhonchi. Signs and symptoms of this usually benign disease are similar to those for epiglottitis. Usually, a chest X-ray determines the cause of respiratory distress in children. Epiglottitis, a bacterial infection, preceded by a minor respiratory illness, sometimes may be present in a child with sudden respiratory distress and a high fever, so called seal bark, hoarseness, and anoxia. Remember: excitement or stress that causes the child to cry can produce immediate airway obstruction. Epiglottitis is more common in children between ages 3

and 8; croup, which has similar signs and symptoms, is more common in children between ages 2 and 5.

Intercostal, sub costal, and suprasternal retractions and expiratory grunts are always serious signs in children. Refer an infant or child with any of these signs for treatment immediately. He may have pneumonia, respiratory distress syndrome, or left-sided heart failure. An infant with untreated pneumonia can die within hours.

When a child's signs and symptoms include retractions, nasal flaring, cyanosis, restlessness, and apprehension – primarily on inspiration – the trachea or main stem bronchus may be obstructed. If these signs and symptoms occur on expiration, his bronchioles may be obstructed, as seen with asthma or bronchitis. Foreign body aspiration is another major cause of respiratory distress in children. These signs and symptoms indicate serious respiratory distress.

Respiratory Distress in Children

Ages 0 – 2

Common – acute lower respiratory tract infections (bronchitis, bronchopneumonia), bronchiolitis, aspiration pneumonia

Uncommon – congenital laryngeal web, laryngeal cyst, subglottic stenosis, congenital heart disease, laryngomalacia

Ages 3-18

Common – laryngotracheobronchitis (croup), acute pneumonia, atelectasis, foreign body (choking), asthma, acute epiglottitis

Uncommon – pneumothorax, congenital heart disease, neoplasms of laryngeal structure

Abnormal Pediatric Chest Anatomy

While you are examining the child, note any structural abnormalities of his chest. Chest abnormalities in children, and their significance, include the following:

- An unusually wide space between the nipples may indicate Turner's syndrome (the distance between the outer areolar edges should not be more than one quarter of the patient's chest circumference)
- Rachitic beads (bumps at the costochondral junction of the ribs) may indicate rickets
- Pigeon chest may be a sign of Marfan's or Morquio's syndrome or any chronic upper respiratory tract obstruction, funnel chest may indicate rickets or Marfan's syndrome, barrel chest may indicate chronic respiratory disease, such as cystic fibrosis or asthma
- Localized bulges may suggest underlying pressures, such as cardiac enlargement or aneurysm
- Multiple (more than five) café-au-lait spots may be associated with neurofibromatosis

Diagnostic Testing

The first thing that is usually done following a physical assessment is a **chest x-ray**, which depending on its results, may be followed by a **collection of sputum specimens** to determine if dysfunction results from cancer, bacteria, or parasites. **Arterial blood gases** can evaluate the child's ability to exchange a significant amount of carbon dioxide or oxygen. The chest x-ray visualizes the appearance and status of the entire respiratory system. A **lung scan** will visualize distribution of blood flow patterns inside the lungs, where a **ventilation scan** evaluates the ventilatory (respiratory) function.

A **CAT scan or MRI** locates a specific area of the lung to be x-rayed and it shows in more detail that particular area. **Pulmonary arteriogram** will visualize the pulmonary vascular system, or the blood supply to the lungs. A **bronchoscopy** is used to directly examine the larger airways or the tracheo-broncho tree and sometimes is necessary to remove the very thick secretions (or mucus plug) that the patient is unable to expectorate him or herself. **The mediastinoscopy** directly examines the mediastinum, which is the bone between the lungs for any kind of biopsies and they usually can be done with a bronchoscopy.

Pulmonary function studies (PFT'S) evaluate ventilation (respiratory) function of the lungs and the chest wall and can help in diagnosing pulmonary disorders. It helps classify these disorders as restrictive or obstructive and evaluates the severity of being able to measure how much volume is taken into the lungs, how much is taken out, expired volume, etc. **Sinus x-rays** are many times done to just make sure there is not just a simple explanation as a sinus infection and also a **thoracentesis** may be performed. This is when there is an increased amount of fluid in the pleural space which is the lining covering the lungs that can produce a lot of pain and difficulty in breathing. A thoracentesis can be done where they put a needle in between the pleuritic space and the lung wall and withdraw the amount of fluid so the patient is able to breathe much easier. The fluid is sent to the laboratory for diagnosing a specific organism if infection occurs.

Infections

Upper Respiratory Tract

Some of the infections that you will see from the upper respiratory tract are obstruction of the airways, which would include food or objects, and one should know the Heimlich maneuver. This is an emergency procedure for dislodging a bolus of food or other obstruction from the trachea to prevent asphyxiation. The rescuer, whose fist is placed under the sternum, grasps from behind the child and quick thrusts upward are given to dislodge the object). Also you might see colds with a runny nose or a head cold (watery to mucoid, sometimes nasal purulent discharge); pharyngitis (sore throat) a fever or swollen throat, not localized to the tonsils; tonsillitis - which you would see in a local infection of the tonsils, may be reddened, swollen, fever, sore throat, with exudate on the surface (bacterial tonsillitis is quite common); or you may see sinusitis and otitis media. Otitis media is an earache which can be a painful; inflammatory condition of the sinuses and the middle ear and drainage may occur leading to the bacterial infection

(usually secondary to viral infections of the nose and pharynx); or you may see flu like symptoms with fever, achiness, sore throat, headache, and cough (usually not much nasal discharge, compared to a cold).

Lower Respiratory Tract

Some of the problems you will see with lower respiratory tract infections are laryngotracheal bronchitis or croup, which many of us know it as, acute bronchitis; or pneumonia, which will include bronchopneumonia, aspiration pneumonia, or nosocomial pneumonia.

Croup

Laryngotracheal bronchitis or croup

Croup is an acute viral inflammation of the larynx and trachea in small children. Often preceded by a “cold”, accompanied by pyrexia, hoarseness, croaking cough, stridor, restlessness (respiratory insufficiency), can be fatal, and a life-threatening disease. Croup is a severe inflammation and obstruction of the upper airways occurring as acute bronchitis, (most common), laryngitis, and acute spasmodic laryngitis and most always can be distinguished from epiglottitis. Croup is a childhood disease affecting boys more often than girls, typically between ages 3 months and 3 years, occurs during the winter. About 15% of the children have a strong family history of croup. Recovery is usually complete. It is caused by a viral infection causing two thirds of the infection and bacteria, which are pertussis and diphtheria, account for the rest and this is why kids when they are little, are more susceptible due to lung development.

The signs and symptoms of croup usually follow an upper respiratory tract infection. You will see inspiratory stridor, hoarse or muffled vocal sounds, varying degrees of laryngeal obstruction and respiratory distress, and a characteristic sharp, bark-like cough. These symptoms may only last for a few hours or persist for a day or two. As it progresses, croup causes inflammatory edema and possibly spasms, which can obstruct the upper airways and severely compromise ventilation. It is usually diagnosed with a throat culture or a neck x-ray may show the areas that are swelling. A lot of times they will do a laryngoscopy to reveal the inflammation and obstruction in those areas. It is also necessary to consider foreign body obstruction, just because in children, especially, a common cause of croupy coughing can be masses and cysts.

Most kids will get better with just home care and rest, cool humidification during sleep and pyretics, such as aspirin or acetaminophen, will relieve the symptoms. However, respiratory distress also interferes with oral hydration, require hospitalizations and IV fluid and replacement to prevent dehydration. If it is a bacterial infection, then antibiotic therapy is necessary and oxygen may also be required. We need to monitor and support respirations and control the fever, because croup is so frightening to the child and the family, you have to also provide support and reassurance to them. We need to monitor the cough; breath sounds, to see how much hoarseness is there; the severity of

any kind of retractions; the respiratory rate, and the depth and character of the respirations.

We need to keep the child as quiet as possible, but not to the point of using sedation to do that. We need to keep other children with any kinds of flu, etc. away from our children. We need to control the fever with sponge baths and antipyretics; relieve the sore throat with soothing water based ices, such as fruit sugar to popsicles; you may want to avoid foods that are milk based, they produce a lot of mucus and there might be a lot more difficulty in swallowing. If the nose becomes sore, we can use ointments, etc. to keep it moist. You can use a cool humidifier or vaporizer, or we could take the child into the bathroom and turn on the hot water. Breathing in the warm moist air quickly eases an acute spell of croup. We need to warn parents that ear infections and pneumonia are complications of croup, which may appear about five days after the recovery. We need to tell parents to report earaches, productive cough, high fever, or increased shortness of breath, to their doctor immediately.

Acute Bronchitis

Bronchitis is produced by an excessive mucus production with a productive cough. Only a minority of children, with the clinical syndrome of chronic bronchitis, develops significant airway obstruction. It is caused by usually the amount of second hand smoke or respiration infection, which exacerbates the symptoms. Other signs and symptoms could just be colds associated with sputum production, hoarsening, and dyspnea, which take progressively longer to resolve. Copious amounts of grey, white, or yellow secretions, weight gain due to the increase edema, cyanosis, tachypnea, wheezing, prolonged expiratory time and use of accessory muscles of respirations are more signs and symptoms.

The diagnosis can also be confirmed with physical exam, chest x-ray, pulmonary function studies, arterial blood gases, sputum cultures, and an EKG. If it is an infection you want to use antibiotics, avoidance of smoking or air pollutants, or being around someone who is smoking, bronchodilators to relieve the bronchospasm to facilitate the mucosal clearance, adequate fluid intake and chest physiotherapy to mobilize the secretions, nebulizer treatments to loosen secretions and aid in mobilization.

Occasionally children respond to corticosteroids, diuretics for the increase in edema, and oxygen for the hypoxia if it is significant enough.

Pneumonia

Pneumonia, in general, is an acute infection of the lung tissue, which often impairs gas exchange. Prognosis is generally good for people who have normal lungs and adequate post defenses before the onset of the pneumonia. However, bacterial pneumonia is the fifth leading cause of death in debilitated children. Causes of pneumonia can be classified in several ways: microbiologic, where pneumonia can be a viral, bacterial, fungal, protozoal, microbacterial, microplasmal, or rickets in origin. The location in bronchopneumonia involves distal airways and alveoli, nebular pneumonia, part of the lobe or lobar pneumonia, which is an entire lobe and that is where we get the irregular pneumonia or double pneumonia where it includes both lungs as well. The

type of pneumonia (primary pneumonia) results from inhalation or aspiration of a pathogen. It is either pneumococcal or viral.

Secondary pneumonia may follow initial lung damage from a noxious chemical or other insult, which is a super infection or may result from spread of bacteria from a distant focus. Pre disposing factors to bacterial and viral pneumonia include chronic illness and debilitation, cancer particularly lung cancer, abdominal and thoracic surgery, atelectasis, common colds, or other respiratory infections. Chronic respiratory disease as in asthma, bronchiectasis, or cystic fibrosis, influenza, exposure to smoke, and malnutrition, alcoholism, tracheostomy, exposure to noxious gases, aspiration, and immunosuppressive therapy. Pre disposing factors to aspiration pneumonia include: older aged debilitation, nasogastric tube feeding, impaired gag reflex, poor oral hygiene, and decreased level of consciousness.

The five cardinal symptoms of early bacterial pneumonia are coughing, sputum production, thoracic chest pain, shaking chills, and fever. Physical signs vary widely ranging from diffuse, fine rales to signs of localized or extensive consolidation and pleural effusion. Complications include respiratory failure, pleural effusion, empyema, lung abscess, and bacteremia (which spread infection to other parts of the body, including possible meningitis). Clinical features such as chest x-rays will show infiltrates and sputum smears will show the acute inflammatory cells to diagnose pneumonia. Positive blood cultures with pulmonary infiltrates suggest pneumonia produced by the organism isolated from the blood cultures. Pleural effusion should be tapped and fluid analyzed for evidence of infection in the pleural space and this is when the doctor would consider performing a thoracentesis to relieve your child's breathing difficulties.

Treatment varies with the causative agent. Treatment should be re-evaluated early. Supportive measures include: humidified oxygen therapy for hypoxia, mechanical ventilation for respiratory failure, a high calorie diet and adequate fluid intake, bed rest and an analgesic to relieve pleuritic chest pain. Children with severe pneumonia, on mechanical ventilation, may require positive end expiratory pressure or what we call PEEP, to facilitate adequate oxygenation. That means that the ventilator just does not allow the lungs to completely exhaust all of the oxygen on expiration, it will keep the lung a little bit inflated to avoid the total exhaustion of the lung.

What we need to do as far as parents or nursing, we need to maintain a patent airway and adequate oxygen. We can check arterial blood gases and that will be our best bet to see how much oxygen is actually needed. We need to teach the child how to cough and perform deep breathing exercises, to be able to clear the secretions and encourage them to do so often (if the child is able). In severe pneumonia, an endotracheal tube or tracheostomy would most likely be present, we need to provide thorough respiratory care and suctioning often and using sterile technique to remove the secretions. Sputum specimens are needed for diagnosis and may require suctioning if the child cannot produce the secretions themselves. Administer antibiotics as ordered, pain medications, and record the child's response to the medications. Fever and dehydration may require IV fluids and electrolytes replacement.

Maintain adequate nutrition to offset high caloric utilization secondary to infection. Ask the dietary department to provide a high calorie, high protein diet, consisting of a soft easy to eat food. Encourage the patient to eat. As necessary supplement oral feeding with nasogastric tube feedings or G-tube feedings, if that route is available. Monitor fluid intake and output.

Provide a quiet, calm environment for the child with frequent rest periods. Get emotional support by explaining all procedures to the child and his family. Encourage family visits providing positionary activities appropriate to the child's age. To control the spread of infection dispose of secretions properly. Tell the child to sneeze and cough into a disposable tissue; tape a wax paper bag to the side of the bed for used tissues.

To prevent pneumonia, advise the parents to avoid using antibiotics indiscriminately during minor viral infections, since this may result in upper airway colonization with antibiotic resistant bacteria. If the child then develops pneumonia, the organisms producing the pneumonia, may require treatment with toxic antibiotics. Encourage annual flu vaccination and Pneumovax for high-risk children, which our children would be in that group. For bedridden and postoperative children, perform deep breathing and coughing exercises frequently. Position your child properly to promote full aeration and drainage of secretions.

To prevent aspiration during nasogastric tube feedings, elevate the child's head, check the position of tube, and administer tube feedings slowly. Do not give large volumes at one time since this can cause vomiting. If the child has an endotracheal tube inflate the tube cuff, keep his head elevated for at least a half-hour after feeding. Aspiration is a result from vomiting and aspiration of gastric or oropharyngeal contents into the trachea and lungs. Signs and symptoms of aspiration are pulmonary edema, which may follow damage to respiratory cells from contact with the stomach acid. You hear rales in the lungs, shortness of breath, color may change to a blue cyanotic color, blood pressure may drop, and his heart rate may increase. This may be a sub acute pneumonia with cavity formation or lung abscess, which may occur if foreign bodies are present. You need to do a chest x-ray, which will suggest the diagnosis and then treatment with penicillin or other antibiotics that are specific. We need to offer supportive care: oxygen therapy, suctioning, coughing, deep breathing, chest percussion, hydration, and IV steroids will be also effective.

Types of Pneumonia

Viral (prognosis poor even with treatment, 50% mortality)

Signs and Symptoms – cough (initially non-productive, later purulent sputum), marked cyanosis, dyspnea, high fever, chills, substernal pain and discomfort, moist rales, frontal headaches, myalgia; *death* results from cardiopulmonary collapse

Diagnosis - chest x-ray: diffuse bilateral bronchopneumonia radiating from hilus; WBC: normal to slightly elevated; sputum smears: no specific

organisms

Treatment – supportive: for respiratory failure, endotracheal intubation and ventilator assistance; hypothermia blanket or antipyretics to control fevers; Amantadine may be used for influenza A

Adenovirus (insidious onset: generally affects young adults)

Signs and Symptoms – sore throat, fever, cough, chills, malaise, small amounts of mucoid sputum, retrosternal chest pain, anorexia, rhinitis, adenopathy, scattered rales and rhonchi

Diagnosis – chest x-ray: patchy distribution of pneumonia, more severe than indicated by physical examination; WBC: normal to slightly elevated

Treatment – treat symptoms only, mortality low; usually clears with no residual effects

Respiratory syncytial virus RSV (most prevalent in infants and children)

Signs and Symptoms - listlessness, irritability, tachypnea with retraction of intercostal muscles, slight sputum production, fine moist rales, fever, severe malaise, and possibly cough or croup

Diagnosis – chest x-ray: patchy bilateral consolidation; WBC: normal to slightly elevated

Treatment – supportive: humidified air, oxygen, antimicrobials often given until viral etiology confirmed; complete recovery in 1-3 weeks

Measles/Rubeola

Signs and Symptoms – fever, dyspnea, cough, small amounts of sputum, coryza, skin rash and cervical adenopathy

Diagnosis – chest x-ray: reticular infiltrates, sometimes with hilar lymph node enlargement; lung tissue specimen: characteristic giant cells

Treatment – supportive: bed rest, adequate hydration, antimicrobials; assisted ventilation, if necessary

Chickenpox/varicella (uncommon in children, but present in 30% of adults with varicella)

Signs and Symptoms – cough, dyspnea, cyanosis, tachypnea, pleuritic chest pain, hemoptysis, and rhonchi 1-6 days after onset of rash

Diagnosis – chest x-ray: shows more extensive pneumonia than indicated by physical examination, and bilateral, patchy, diffuse, nodular infiltrates; sputum analysis: predominant mononuclear cells and characteristic intranuclear inclusion bodies, with characteristic skin rash confirm diagnosis

Treatment - supportive: adequate hydration, oxygen therapy in critically ill patients

Cytomegalovirus CMV

Signs and Symptoms – difficult to distinguish from other nonbacterial Pneumonias; fever, cough, shaking chills. Dyspnea, cyanosis, weakness and diffuse rales; occurs in neonates as devastating multisystemic infection; in normal adults resembles mononucleosis, in immunocompromised hosts,

varies from clinically inapparent to devastating infection

Diagnosis – chest x-ray: in early stages, variable patchy infiltrates; later, bilateral, nodular, and more predominant in lower lobes; *percutaneous aspiration of lung tissue, transbronchial biopsy,* or open lung biopsy: microscopic examination shows typical intranuclear and cytoplasmic inclusions; the virus can be cultured from lung tissue

Treatment – generally, benign and self-limiting in mononucleosis-like form, supportive: adequate hydration and nutrition, oxygen therapy, bed rest, in immunosuppressed patients, disease is more severe and may be fatal

Bacterial (Streptococcus (Diplococcus) pneumoniae)

Signs and Symptoms - A sudden onset of a single, shaking chill and sustained temperature of 102-104 degrees F (38.9-40C); often preceded by upper respiratory tract infection

Diagnosis: chest x-ray: areas of consolidation, often lobar; WBC: elevated, sputum culture: may show gram-positive *S. pneumoniae*; this organism not always recovered

Treatment – antimicrobial therapy: Penicillin G or a cephalosporin for 7-10 days. Such therapy begins after obtaining a culture specimen but without waiting for results

Klebsiella

Signs and Symptoms – fever and recurrent chills: cough-producing rusty, bloody, viscous sputum (currant jelly): cyanosis of lips and nail beds; likely in patients with chronic alcoholism, pulmonary disease, and diabetes

Diagnosis – chest x-ray: typically, but not always, consolidation in the upper lobe that causes bulging of tissues, WBC elevated; sputum culture and Gram's stain: may show gram-positive cocci *Klebsiella*

Treatment – antimicrobial therapy: Gentamicin, Tobramycin, Kanamycin, or a Cephalosporin

Staphylococcus

Signs and Symptoms – temperature of 102-104 degrees F (38.9-40C), recurrent shaking chills, bloody sputum, dyspnea, tachypnea, and hypoxemia; should be suspected with viral illness, such as influenza or measles, and in patients with cystic fibrosis

Diagnosis – chest x-ray: multiple abscesses and infiltrates; high incidence of Empyema; WBC: elevated; sputum culture and Gram stain: may show gram-positive staphylococci

Treatment – antimicrobial therapy: Nafcillin or Oxacillin for 14 days; chest tube drainage of empyema

Aspiration – results from vomiting and aspiration of gastric or oropharyngeal contents into trachea and lungs

Signs and Symptoms – noncardiogenic pulmonary edema may follow damage to the respiratory epithelium from contact with stomach acid; rales, dyspnea, cyanosis, hypotension, and tachycardia; may be subacute pneumonia with cavity formation, or lung abscess may occur if foreign body is present

Diagnosis – *chest x-ray*: locates areas of infiltrates, which suggest diagnosis

Treatment antimicrobial therapy: Penicillin G or Clindamycin; supportive: oxygen therapy, suctioning, coughing, deep breathing, adequate hydration, and IV steroids

Pseudomonas

Pseudomonas is motile, gram-negative rods that produce nosocomial infections very well. These bacteria are clinically important because they are resistant to most antibiotics and they are capable of surviving in conditions that few other organisms can tolerate. It is often encountered in hospitals and clinical work settings, because it is a major cause of hospital acquired nosocomial infections. Its main targets are those that are immunocompromised individuals and they are individuals who are debilitated, on respirators or with catheters in. The infection can occur at many sites and can lead to urinary tract infections, sepsis, pneumonia, pharyngitis and a lot of other problems. Rarely, will you find *Pseudomonas* as a cause of infection on healthy adults.

Pseudomonas usually carries with it a high fever, totally exhausting to the body, has green colored sputum and has a very foul odor. Along with the clinical picture of *Pseudomonas*, lab tests that need to be done are sets of blood cultures, because usually the temperature rises above 102-103 degrees, and also sputum specimen, which will totally identify the cause of the agent. Nursing interventions need to make sure we check for drug allergies, so a child is not allergic to penicillin and then administer the antibiotics as ordered. Usually, there will be at least two antibiotics, one is penicillin and one is a different strain to help rid the *pseudomonas*. One other thing that we can do other than washing our hands extremely well is (*pseudomonas* is an opportunistic infection that flourishes in most environments), do not allow water to stand around sink drains, or flower arrangements, make sure condensation around ventilator circuits does not drain back in to the reservoir. Because *pseudomonas* can cause a nosocomial infection especially in burned and immunocompromised children and those with catheters, use standard procedures and precautions and wash your hands thoroughly and often. Use proper aseptic technique when working with IV lines and catheters and when suctioning endotracheal or tracheostomy sites.

MRSA - Methicillin-resistant-staphylococcus-aureus

This organism is most commonly transmitted to people through direct contact with Healthcare workers who fail to wash their hands between patient contacts. MRSA lives on the hands for three or four hours after exposure. It can also survive for a considerable length of time on inanimate surfaces including furniture and equipment but they seldom are implicated in the spread of MRSA, except perhaps in burn units. The air is not believed to be an important route of transmission either, but again, burn units

are the possible exception. This form of pneumonia is very difficult to clear up, and in so doing, extreme caution needs to be used with using proper antibiotics after allergies are known, use good hand washing techniques and aseptic dressing changes, if that is the case. Gloves need to be worn and hands washed between patient contacts. You need to wash your hands twenty to thirty seconds, then dry them thoroughly, just rinsing ones hands with tap water and drying them can also reduce the contamination of MRSA. Gowns really are not necessary and foods on special disposable trays are really not necessary because this really has to do more with the secretions. Blood cultures and/or sputum cultures is the way to diagnose and to see how the antibiotics and the health care people are doing a repeat in those tests, are the only way to see if MRSA is being controlled. MRSA is here to stay unfortunately; simple infection control measures like hand washing, universal precautions, and contact isolation are usually effective in controlling its spread to the immunocompromised people, which our Batten children are in that group.

Diseases of the lungs

Asthma

Asthma is increased bronchial reactivity to a variety of stimuli, which produces episodic bronchospasms in airway obstruction. When asthma is associated with childhood onset, there can be an acute attack with severe bronchospasms that fail to clear with bronchodilators. More than half of asthmatic children become asymptomatic as adults and those that are acquired after the age of 15, occasionally have persistent disease and occasional severe attacks. Causes of asthma include: a possible mechanism of allergies, which are family tendency, seasonal occurrence, or allergic reactions, as a result of a cell vasoactive and bronchospastic mediators. An asthma attack may be precipitated by an upper airway infection, exercise, anxiety, and rarely coughing or laughing. Obstruction associated with nasal polyps may be seen and response to aspirin ingestion. Airway obstruction, from spasms of the bronchial smooth muscle, narrows airways and inflammatory edema of the bronchial walls, are also important particularly in status asthmaticus.

So what we need to do is get the history of these attacks as dyspneic and wheezing and you will see sometimes it progressing to severe dyspnea, audible wheezing, which is just what you can hear without even a stethoscope, tightness, and cough production of thick mucus. Other signs you might see are prolonged expiration, use of accessory muscles, flaring of the nostrils, fast heart rate, and labored breathing, along with perspiration and flushing of the face. If it is not treated, where someone is really in labored breathing, it can lead to respiratory failure. You can do all kinds of tests to determine if this is asthma from chest x-rays, to sputum cultures, to pulmonary function studies, arterial blood gasses, EKG, and skin tests. The prevention is to tell the parent to watch their children to avoid possible allergens and to use antihistamines, decongestants, inhalation of powder, oil, and aerosol bronchodilators as ordered. Explain the influence of stress and anxiety when asthma and frequent association of exercise, particularly running and cold air.

Acute respiratory failure

What happens in acute respiratory failure is when the child's lungs cannot maintain adequate arterial oxygenation or carbon dioxide elimination. This leads to hypoxemia, which is a decrease of oxygen in the blood and arises from decrease in ventilation or circulatory deficits. You can recognize it when their O₂ levels drop below 50 and which is going to be most likely a crisis situation. The carbon dioxide level will usually rise suddenly with a sharp decrease in the pH. Three main mechanisms may cause hypoxemia that leads to acute respiratory failure: hypoventilation, ventilation-perfusion, and right to left shunting. Alveolar hypoventilation commonly results from the chronic airway obstruction that reduces alveolar minute ventilation pO₂ levels in oxygen saturation decreases, while pCO₂ levels increase signaling hypercapnia, which is increase in carbon dioxide in the blood.

Ventilation-perfusion imbalances, the most common cause of hypoxemia, occur when conditions, such as massive pulmonary embolism or ARDS (adult respiratory distress syndrome) upset the ventilation-perfusion in a specific region of the lungs. Either too little ventilation with normal blood flow or too little blood flow with normal ventilation may cause an imbalance. Which ever happens, the results are the same; when the balance spreads over a large area, pO₂ levels decrease throughout the lungs. When hypoventilation and ventilation imbalances are not treated, they may lead to right to left shunting. When this happens, a large amount of blood passes from the right to the left lung without being oxygenated.

Hypoxemia, along with hypocapnia, triggers several compensatory mechanisms in the body. Hypoxemia stimulates the sympathetic nervous system, which in turn, produces tissue vasoconstriction, increases peripheral resistance, and increases the heart rate. Hypocapnia also works on local cellular and tissue function to cerebral depression, hypotension, and circulatory failure. Additionally, it stimulates the sympathetic nervous system to increase the heart beat and cardiac output. Eventually it may lead to acute respiratory acidosis.

As far as taking care of a child in acute respiratory failure, the doctor will order an IV and be given either IV bronchodilators or oral ones, if you detect any signs or symptoms of infections, a sputum culture will be obtained and antibiotics will be prescribed according to the type of infection. You want to avoid sedating a child, because that can just reduce his respiration effort as well. But if he is anxious and his oxygen levels are okay, the doctor may prescribe a mild sedative or antianxiety agent such as Valium or something to help him relax. Checking the ABG's or checking the amount of oxygen or carbon dioxide in the blood will be a good factor in determining how the child is progressing. During this phase, he will also need aggressive chest physiotherapy and postural drainage to remove the secretions and improve pulmonary function. If the child is trached or has an endotracheal tube you will want to do meticulous trach care and also suctioning which we will get into in a little bit.

SIDS - Sudden Infant Death Syndrome

SIDS is sudden infant death syndrome or crib death. It is a medical mystery of early infancy; sudden infant death syndrome kills apparently healthy infants, usually between age's four weeks to seven months for reasons that remain unexplained, even after an autopsy. Typically, the parents put the infant to bed and later find him dead, often with no indications of a struggle or distress of any kind. Some infants may have signs of a cold, but such symptoms are usually absent. SIDS has occurred throughout the history all over the world and in all climates. SIDS accounts for 6 to 7 thousand deaths annually in the United States making it one of the leading causes of infant death. Most of these deaths occur during the winter, and among underweight babies and those to mothers under age 20. Although infants who die from SIDS often appear healthy, research suggests that many may have had undetected abnormalities, such as immature respiratory system and respiratory function. In fact, the current thinking is that SIDS may result from an abnormality in a control of ventilation, which causes prolonged apneic periods with profound hypoxemia and serious cardiac arrhythmias. The use of bottle-feeding instead of breast-feeding in advanced parental age does not cause SIDS.

Atelectasis

Atelectasis is an incomplete expansion of the lobules (clusters of alveoli) or lung segments, which may result in partial or complete lung collapse. This causes the loss of regions of the lungs for gas exchange; unoxygenated blood passes through these areas, unchanged, thereby producing hypoxia. Atelectasis may be chronic or acute, and occurs to some degree in many children undergoing upper abdominal or thoracic surgery. Prognosis depends on prompt removal of any airway obstruction, relief of hypoxia and reexpansion of the collapsed lung. Atelectasis often results from bronchial occlusion by mucus plugs (a special problem in persons with chronic obstructive pulmonary disease and where the secretions are just so thick that they are unable to get them coughed up). Atelectasis may also result from occlusion of foreign bodies in the lungs and any inflammatory lung disease.

Atelectasis of a small area of the lung may produce only minimal symptoms, which subside without specific treatment; however, massive collapse can produce severe dyspnea, anxiety, cyanosis, diaphoresis, peripheral circulatory collapse, tachycardia, and substernal or intercostal retractions. Also, atelectasis may result in compensatory hyperinflation of unaffected areas of the lungs, mediastinal shift to the affected side and elevation of the diaphragm. The chest x-ray will show as well as physical examination that atelectasis is the problem.

The treatment for atelectasis is incentive spirometer, chest percussion, postural drainage, frequent coughing and deep breathing exercises or intermittent positive pressure breathing which is done with a machine. If these measures fail a bronchoscopy may be helpful in relieving secretions. Humidity and bronchodilators can improve mucociliary clearance and dilate airways; they are sometimes used with IPPB treatments (intermittent positive pressure breathing).

Postoperative thoracic and abdominal surgery patients require analgesic to facilitate deep breathing, to minimize the risk of atelectasis. We need to encourage post op

patients and other high-risk patients to cough and deep breath every one to two hours. To minimize pain you might get them to hold a pillow tightly over the incision, gently reposition these children often and help them walk as soon as possible. Use incentive spirometry to increase his inhalations to 800-1500 CC's on the machine and that should be adequate volume to have a normal respiratory state. To promote mobilization and clearance of secretions, use chest percussion and postural drainage. Assess breath sounds frequently and report changes immediately. Teach the child about respiratory care including what he has been experiencing, so he can continue at home with postural drainage as much as possible, coughing and deep breathing. Encourage anyone around him who is smoking to stop. Provide reassurance and emotional support, as the child will undoubtedly be frightened by limited breathing capacity and difficulty in speaking.

Respiratory Acidosis

Respiratory acidosis is an acid based disturbance characterized by reduced alveolar ventilation and manifested by hypercapnia, which is an increase in carbon dioxide. Respiratory acidosis can be acute (due to a sudden failure in ventilation) or chronic (as in long term pulmonary disease). Prognosis depends on the severity of the underlying disturbance, as well as the child's general clinical condition. Acute respiratory acidosis produces CNS (central nervous system disturbances), which reflect changes in the pH rather than increased carbon dioxide level and cause the child to become restless, confused, apprehensive or somnolent and to develop a fine or flapping tremor or to slip into a coma. He may complain of headaches, exhibit dyspnea, tachypnea with edema and depressed reflexes; unless the child is receiving oxygen (hypoxemia accompanies respiratory acidosis). This disorder may also cause cardiovascular abnormalities, such as tachycardia, hypertension, atrial and ventricular arrhythmias and in severe acidosis, hypotension and vasodilatation (bounding pulses and warm periphery).

Blood gases will confirm the respiratory acidosis and treatment is designed to correct the underlying cause of alveolar hypoventilation. It may require mechanical ventilation until an underlying condition can be effectively treated. Sometimes all it requires is a change on the ventilator settings where better oxygenation will occur. Be alert for critical changes in the patients respiratory, CNS, and cardiovascular functions. Report any such changes immediately as well as any variations in the blood gases and electrolyte status. Maintain adequate hydration. Make so an adequate airway is provided and also adequate humidification if acidosis requires mechanical ventilation. Perform tracheal suctioning and regularly and vigorous chest physiotherapy if ordered.

Respiratory Alkalosis

Respiratory alkalosis is a condition marked by a decrease in the carbon dioxide levels, which is due to alveolar hyperventilation. Uncomplicated respiratory alkalosis leads to a decrease in hydrogen ion concentration, which causes elevated blood pH, hypocapnia occurs when the elimination of carbon dioxide by the lungs see the production of carbon dioxide at the cellular level which will fall into one of two categories: one is pulmonary, which can be due to pneumonia, interstitial lung disease, pulmonary vascular disease

and acute asthma; where the non-pulmonary one can be caused by anxiety, fever, aspirin toxicity, metabolic acidosis, central nervous system disease, like in an inflammation of a tumor, gram negative septic cases and liver failure. The cardinal sign for respiratory alkalosis is deep, rapid breathing, possibly above 40 respirations per minute. Such hyperventilation usually leads to central nervous system and neuromuscular disturbances, causing lightheadedness or dizziness, agitation, excitement, peripheral paresthesias, spasms, twitching (possibly progressing to tetany) and muscle weakness. Characteristic affects of severe respiratory alkalosis include hyperpnea and cardiac arrhythmia (that may fail to respond to conventional treatment).

Again, diagnosis is with arterial blood gases and you want to treat them to irradiate the underlying condition; removal of the ingested toxins, treatment of the fever or sepsis, and treatment of the central nervous system disease. Parents may have their child be instructed to breathe into a paper bag, which helps relieve acute anxiety, and increases the carbon dioxide levels. Prevention in hyperventilation receiving mechanical ventilation requires monitoring of the arterial blood gases and adjusting minute ventilation volume. Nursing intervention is to observe the child carefully for subtle changes in neurological, neuromuscular, or cardiovascular functions. Report any such changes immediately. Remember twitching and cardiac arrhythmias may be associated with alkalemia and electrolyte imbalances. Monitor arterial blood gasses and serum electrolytes closely reporting any variations immediately. Explain all diagnostic tests and procedures to reduce anxiety.

Medications

Make sure we check the child's allergies before administering antibiotics.

Antihistamines can lower seizure threshold. Refer to the medication book for the section on antibiotic therapy. There is a wide range of medications that can be used with the respiratory system, but one thing to keep in mind is that too much sedation can cause slower, shallow breaths, which in turn will increase the congestion already present in most children with Batten Disease. Be sure to talk with the pharmacist to make sure the combination of medications that your child is receiving is OK. Give antibiotics for the full length of the prescription. Just because he may be "better" after 7 days, give the full 10 days to prevent a reoccurrence of the infection. Milk or dairy products thicken secretions, so you might want to avoid them during a case of pneumonia. Nebulizer treatments also are medications. Be sure to give your child the number of treatments per day as ordered. If you have reached a point where most bronchodilators are not working very well, you might want to ask your doctor about using morphine as a bronchodilator in a small dose, I hear it works well. Also, postural drainage and percussion is also very important in the care of your child with pneumonia. Refer to the pictures and procedures for chest physiotherapy under the treatment section (pages 35-39).

Treatment

Using Oxygen Safety and effectively

Whether the oxygen is being used at home or in a hospital setting, especially at home, is where we are going to concentrate our thinking today. They are going to be having extra oxygen at home, using an oxygen concentrator, a liquid oxygen unit or an oxygen tank and the prescribed amount of flows per liter per minute for so many hours a day will be prescribed and that will be written on a prescription. When you get the oxygen set up at home from the medical equipment suppliers they will teach you how to set it up, check for problems, and clean it properly. The oxygen system will include a humidifier to warm and add moisture to the prescribed oxygen, and nasal cannula or a facemask to breathe the oxygen. Make sure you keep the suppliers phone number handy in case the system does not work properly. Also arrange to get a back up system - a small portable oxygen tank is usually best. An emergency, for example in a power failure, will protect you from being without needed oxygen.

The general guidelines for oxygen equipment at home are: when using an oxygen tank, oxygen concentrator, or liquid oxygen, be sure to follow these important guidelines: check the water level in the humidifier bottle often, if it is near or below the refill line, pour out any water remaining in the bottle and refill it with sterile or distilled water; if your nostrils become dry or cracked, apply a water-soluble lubricant such as KY jelly - avoid using a petroleum - based lubricant such as Vaseline; always reorder a new supply of oxygen two to three days in advance or when the register reads $\frac{1}{4}$ full so that you do not run out; maintain your oxygen flow at the prescribed rate; if you are not sure whether oxygen is flowing through the system, first check the tubing for kinks or other obstructions, then, double check that the system is turned on, if you are still unsure try this test, invert the nasal cannula in a glass of water, if bubbles appear, oxygen is flowing through the system; be sure to shake off excess water before you reinsert the cannula in your nose.

Safety Tips

Remember that oxygen is highly combustible. Alert your local fire department that oxygen is in the house, and keep an all-purpose fire extinguisher on hand. If a fire does occur, turn off the oxygen immediately and leave the house. Do not smoke or allow others to smoke near your oxygen system. Keep the system away from open flames and heat. If you have a gas range, stay out of the kitchen when it is on. Do not run oxygen tubing under clothing, bed covers, furniture, or carpets. Keep the oxygen system upright. Make sure the system is turned off when it is not in use.

When to call your doctor

To use oxygen effectively, you need to know when you are not getting enough oxygen- and when you are getting too much. You may not be getting enough oxygen if you notice these symptoms:

Difficult irregular breathing
Restlessness
Anxiety

Tiredness or drowsiness
Blue fingernail beds or lips
Confusion or inability to concentrate.

You may be getting too much oxygen if you notice these signs:

Headaches
Slurred speech
Sleepiness or difficulty waking up
Shallow, slow breathing

If any of these signs develop call your doctor immediately. Above all, never change the oxygen flow rate without checking with your doctor first. Oxygen administration is by cannula, catheter, or mask. Oxygen is administered by cannula which is a nasal prongs, catheter or mask to prevent or reverse hypoxia and improve tissue oxygenation. Hypoxia can result from a disorder in respiration - ventilation distribution, diffusion, or perfusion. Unchecked, it affects the brain, adrenal gland, heart, kidneys, and liver.

The nasal cannula delivers low flow oxygen between 22% and 30% concentration to enrich the respiration when accuracy is not crucial. Inexpensive and easy to use it interferes less with the child's movement and offers other functions than any other device. If the oxygen mask is needed depending on the type it can deliver concentrations of up to 100% of oxygen. It is used when the child requires high humidity and precise amounts of oxygen or can breathe only through his mouth. Because the mask is confining, it prevents eating, and hampers speech, it may reduce compliance in the child, he may remove his mask when not in crisis.

Humidifiers

Humidifiers add water vapor to inspired air to prevent drying and irritation of the respiratory mucosa and to help loosen respiratory secretions for easier removal. Some humidifiers also heat the water vapor, thereby enhancing the moisture - carrying capabilities of the gas and increasing the humidity delivered to the child. Supplemental humidity must accompany delivery of such medical gases as oxygen, because the gases are totally dry and extremely irritating to mucus membranes. It is used with every oxygen delivery device except the Venturi mask. If a child is using a Venturi mask and requires humidification, the room air is humidified, not the oxygen. Supplemental humidity is also used when secretions are particularly thick and tenacious and the child needs relief from croup or tracheitis. Humidity may be added to a room by using a room humidifier, or combined with inspired gas lines and delivered directly to the child. In line devices include, the cold bubble - diffusion humidifier for the child with an intact upper airway and a cascade humidifier, which can deliver a 100% of needed body humidity to the child when heated and connected to a ventilator.

Pediatric and young children nebulizer therapy

If you are caring for an infant or a child with breathing difficulties that stems from a condition such as asthma, bronchitis, or fibrosis you need to teach his/her parents how

to administer pulmonary medications via nebulizer. Each treatment takes ten to twenty minutes depending on the medication and the child's condition. Parents have three options for administering a nebulizer treatment, the preferred method for infants and young children is to use an aerosol mask, but the child may not tolerate the mask. An alternative is the blow - by method, which the parent directs the aerosol medication towards the child's nose and mouth. Though this method is less effective than using a mask, it is preferable to struggling with a child who already has breathing troubles. The third method, often used for children older than five year who can cooperate, is to use a mouthpiece and reservoir tube. You will learn about all three methods. Steps may differ slightly depending on the equipment model, so refer to the manufacturers instructions before proceeding. Gather all the equipment needed: your flow meter, the compressor, the nebulizer equipment, the medications, syringes if needed and sodium chloride solution if necessary to mix the medications.

Mucus is 95% water; it thickens so its water content evaporates in an attempt to counter the humidity deficit. The dense mucus makes it more difficult for the slow - like cilia to move the mucus. That hinders the process of mucociliary clearance the movement of mucus and contaminates up and out of the lungs. As the mucus moves more slowly, the pulmonary system becomes less able to fend off infections. Eventually, the mucus becomes thick and inelastic, the cilia immobilized, and mucociliary clearance stops all together. This further decreases pulmonary defenses and puts the child at even greater risk for infection. This will lead to cell damage – the lack of moisture causes the cilia to break off from the cells in the airway causing damage to the mucosal lining which causes more stress to the mucosal lining – atelectasis occurs.

The steps for initiating a nebulizer treatment as ordered by your doctor are as follows:

Wash your hands and fill the medication cup with the appropriate medication, if necessary mix the medication with the 0.9% sodium chloride solution, then screw the cap onto the cup.

Screw the cap to the t-piece. For the blow-by method, tape one end of the t-piece so that no aerolized medication will escape from it. For the aerosol mask method, screw the cap directly onto the mask (you will not use the t-piece). For the mouthpiece method, attach the mouthpiece to one end of the t-piece and the corrugated reservoir tube to the other end, so exhaled air can escape.

For all treatments, attach one end of the oxygen tubing to the compressor and the other end to the bottom of the medication cup. Plug the compressor into an electrical outlet. Administer the treatment, have the child sit upright, whenever possible, for the treatment. Always stay with the child until all of the medication is gone and the treatment is complete.

- If using the blow-by method, direct the aerosolized medication toward the child's nose and mouth by placing the nebulizer as close as he/she will allow without touching the skin. You may have to follow a toddler as he/she moves

about. With good inhalation, you will see the mist disappear directly into his/her nose and mouth.

- If using the mask, fit it over the child's nose and mouth. Adjust the elastic strap so it fits comfortably, but snugly enough to prevent aerosolized medication from escaping. Turn the compressor on so aerosolized medication comes from the nebulizer and ask the child to breathe deeply every few breaths.
- If using the mouth-piece method, ask the child to close her lips tightly around the mouthpiece, during the treatment, she should breathe through her mouth only. Every few breaths, she should take a deep breath to "make the mist disappear"

Take note of the child's medication type, dose, frequency, and method of delivery. Parents do not need to record this information for individual treatments, but they should be able to inform a health care provider of these basics.

Wash the equipment as directed in its product insert.

Wait until it is dry before putting it away.

How to make treatments fun and easier to give

With treatments lasting 10-20 minutes and some children needing two to four treatments a day, getting a child to take a nebulizer treatment can be challenging. Here are some tips to improve the odds of success:

- Distract the child with age-appropriate toys, videos, and games or by reading or singing to him/her.
- Let him/her help assemble the equipment and give a pretend treatment to a doll or bear.
- Allow the child to decorate the mask with stickers.
- Buy a mask in the shape of a dragon or other creature.
- Comfort the child, especially if he/she is an infant or toddler, by rocking or holding him/her.

Inhalers

Inhalers are useful forms of medications for quick relief of an asthma attack and today, inhalers are available of a preventative nature, but they need to be used properly.

The step-by-step instruction of "Using An Oral Inhaler" are as follows:

1. Remove the mouthpiece and cap from the inhaler. Then, remove the cap from the mouthpiece. Next, turn the mouthpiece sideways. On one side of the flattened tip you will see a small hole. Fit the metal stem on the inhaler into the hole. After it is assembled, shake the inhaler five times (the assembly of different inhalers may be slightly altered from one inhaler to another).
2. Exhale fully through pursed lips. Hold the inhaler upside down and close your lips and teeth around the mouthpiece.
3. Tilt your head back slightly. Take a slow deep breath. As you do, firmly press the inhaler against the mouthpiece – one time only – to release one dose of medication. Continue inhaling until your lungs feel full.

4. Take the mouthpiece away from your mouth and hold your breath for three to five counts of one-1000; two-1000; three-1000. Exhale slowly through pursed lips.
5. If your doctor wants you to take more than one dose, wait a couple of minutes before repeating steps 2 through 4.
6. Rinse your mouth, gargle, and drink a few sips of fluid.
7. Remember to clean the inhaler once a day; take it apart and rinse the mouthpiece and cap under warm running water for 1 minute. Shake off the excess water. Allow the parts to dry, and then reassemble them.

Pulse oximetry

Here is how pulse oximetry works: approximately 3% of the total amount of oxygen in the body is dissolved in the plasma and is measured by the pressure of arterial oxygen or pAO₂. The rest of the oxygen is bond to hemoglobin as oxyhemoglobin, or HbO₂. It is this large component that is reflected in the arterial saturation value.

To measure arterial saturation, pulse oximeters use a sensor that is connected to a main unit housing a microprocessor. You attach the sensor to a finger, toe, or earlobe. The sensor has two components - a light source called a light emitting diode or LED and a light detector called a photo detector.

The LED gives off alternating bursts of red and infrared light. As the light passes through the vascular bed, the oxyhemoglobin absorbs more of the infrared light, while the unoxygenated hemoglobin absorbs more of the red light.

The photo detector, which is positioned directly across from the LED, measures how much of each light passes through the finger, toe, and earlobe, to the other side. This information is then sent to the microprocessor in the monitor, which calculates the SpO₂ by measuring how much light was absorbed, and averages the ratios of red light to infrared light absorption.

That in turn, determines the oxygen saturation value, which is displayed as a digital readout. Most units also display the child's pulse rate. (When measured by pulse oximetry, oxygen saturation is noted as SpO₂ instead of SAO₂ to differentiate it from a value obtained from ABG analysis).

Since pulse oximeters monitor light absorption only in tissue with pulsatile flow, they eliminate false readings from fat, bone, connective tissue, and venous blood; thus good arterial flow is required to achieve a reliable reading. While pulse oximeters are generally reliable for oxygen readings between 70% and 100% there are still a number of technical factors that can affect accuracy. One of those is motion artifact and that is if the child moves or partially dislodges the sensor affects the ability of the light to travel from the light emitting diode or the LED to the photo detector. Another thing that might cause an irregular reading is ambient light and that means because pulse oximeters measure the amount of light transmitted through arterial blood, bright light, that shines directly on the sensor - whether from the sun or an overhead exam light can alter the readings. Just simply move the sensor or cover it with something opaque like a

washcloth. The third thing is abnormal hemoglobin. Pulse oximeters cannot differentiate between different forms of saturated hemoglobin. A child with carbon monoxide poisoning, due to smoke inhalation, bonds with the carbon monoxide instead of the oxygen producing carboxyhemoglobins. Because of its absorption it is similar to oxyhemoglobin, the pulse oximeter will provide a false elevated O₂ reading and you will have to do ABGs in order to see what the actual oxygen level is. Another one is low perfusion state. There must be sufficient perfusion in the monitored area. If the child has a weak or absent peripheral pulse, pulse oximetry readings will not be precise. In fact, a weak pulse may even make the sensor more susceptible to motion artifact.

Children who are most at risk for low perfusion states that affect pulse oximetry are those with hypotension, hypovolemia, and hypothermia and of course, in cardiac arrest. Children who are cold but not hypothermic may have vasoconstriction in their fingers and toes that can also compromise arterial flow. Again, you can put the sensor on a different area like the earlobe or a toe. Warming the extremity may also enhance the peripheral perfusion.

Another factor is finger nail polish. It has been said that fingernail polish does affect the accuracy of the pulse ox reading, generally however the darker the polish the more likely the oxygen reading is inaccurate. Blue, black and green polish seems to cause the most problems.

Mechanical Problems

Pulse oximeters are not full proof that is why you should periodically compare the pulse reading from the monitor with the child's actual heart rate to be certain they match. Also, you could do blood gases to correlate the findings.

In today's society, children with more complex illnesses are increasingly being treated at home and this complexity requires more sophisticated monitoring by home care nurses. At risk children include those possibly on mechanical ventilation and those with respiratory diseases that could effect oxygenation. Pulse oximetry can identify changes in a child's pulmonary status before life-threatening symptoms occurs which means early, life saving interventions can be implemented.

Postural Drainage and Percussion

Percussion Techniques and Respiratory Therapy

This is a technique used to assist in the removal of excessive secretions from the airways of the lungs. The technique uses a combination of positioning and percussion so that gravity and vibrations allow secretions to flow toward the mouth where they can be removed with coughing or suctioning.

Indications: This type of therapy should be used whenever secretions in the airways make a continuous audible sound with breathing. These secretions can accumulate from production within the airways or from aspiration of liquids or upper airway

secretions. In small children, the airways are very narrow and a build up can easily block them with these secretions, making breathing difficult. This difficulty in breathing can become evident by attempts at coughing, unusual chest movement during inspiration, and sometimes a bluish color around the lips indicating inadequate oxygen to the tissues.

Frequency: Chest percussion therapy can be delivered as often as necessary to control the build up of secretions, although typically it is administered three or four times daily. The therapy usually lasts from ten to fifteen minutes. Each area of the lung should be percussed from one to three minutes.

Technique: Position the child according to segmental drainage. Using cupped hands or appropriate device, percuss rapidly over lung area creating a low frequency “clapping sound”. Use the wrists and forearms only, avoiding force or slapping. The effectiveness is derived from the vibrations and sounds created within the chest. Have the child attempt to cough, or suction the mouth if necessary.

Precautions: Although quite safe, chest percussion should not be delivered right after feeding, as this may cause vomiting. The percussion may be painful over tender skin areas. A gown, thin shirt, or towels should protect the skin during percussion. Percussion should be confined to lung areas only, avoiding the areas of organs such as the kidney or stomach.

The specific instructions for Chest Physical Therapy for infants & toddlers (on the left), and the older child (on the right) is on page 39.

Chest Physical Therapy (CPT), also called postural drainage, is a way for you to help your child get rid of extra mucus in his lungs. This is important because too much mucus can block the air passages in the lungs. Giving a CPT treatment to your child at home helps keep extra mucus from building up.

HOW DO THE LUNGS WORK?

We breathe in air (inhale) through the nose and mouth. The air goes through the windpipe into the large airways in the lungs. Then the air goes into the small airways and into the air sacs.

The air sacs in the lungs do important work. The oxygen from the air, which we need to live, goes into the blood through the air sacs. The used oxygen is changed into carbon dioxide in the blood. The carbon dioxide goes from the blood into the air sacs and into the air we breathe out (exhale).

WHAT IS MUCUS?

All parts of the lungs have a protective mucous lining. The mucus that covers the lining catches tiny pieces of dirt, dust, and other particles in the air we breathe. These particles would irritate the lungs or cause infection if they stayed in the lungs.

HOW DOES THE MUCUS GET OUT OF THE LUNGS?

All parts of the air passages are lined with tiny hairs called cilia. The cilia act like an escalator and carry the mucus and particles up to the windpipe to be coughed out or swallowed.

WHAT HAPPENS IF THERE IS TOO MUCH MUCUS?

Normally, there is just the right amount of mucus in the lungs. But, when the lungs become irritated or infected, a large amount of thick mucus is produced. This happens because the lungs are working extra hard to get rid of the infection or irritation.

Extra mucus can slow down or stop the cilia from working. If the cilia do not work well, we have to help the lungs to get the mucus out. This is why CPT is done.

This extra mucus can block the air passages. If air passages are blocked, the air cannot move in and out of the air sacs. Then the child does not get enough oxygen into his blood and does not get enough carbon dioxide out of his blood.

HOW IS CPT DONE?

CPT helps to move the extra mucus into the windpipe where it can be coughed up more easily. The doctor will give you the specific orders as to what medication to use; the length of time spent giving CPT in each area, for how many minutes, and how often. Give the treatment before the child eats. (The head-down position may cause vomiting or stomach discomfort if there is food in the stomach).

1. Positioning

- The child should be positioned so that the part of the lung to be drained is higher than any other part of the lung.
- It is important for *you* to be in a comfortable position because this makes the treatment more effective and easier for both you and your child. You may use a pillow to make your child more comfortable.
- Place your child on your lap for the head-down positions.
- Always have your child's knees and hips bent to help him relax and to make coughing easier.

- For the older child – all of the above plus – your child can lie on a padded board and you can get the needed slant for head-down positions by placing one end of a bed or board on blocks

2. **Clapping** – before you begin, explain to your child that the clapping will make a noise like a galloping horse, or like drums in a parade.

Place a lightweight towel or blanket over the child's chest or back.

- Cup your hands by bending them at the knuckles. Hold your thumb against your index finger. Keep your fingers together to form a cup
- Clap your hands, first one and then the other, on the area of the child's chest or back.
- The clapping should be done just firmly enough so that your child's head bobs.
- Do the clapping in a regular rhythm.
- The rate of clapping should be comfortable, and not so fast that you get too tired.
- Clapping, when done properly, does not hurt. If your child cries, it may be because he does not like being held in the position. It is very important that your child does not think of it as a punishment.
- For the older child – do the clapping fairly fast. The rate of clapping should be comfortable, and not so fast that you get too tired.
The clapping should be firm so the mucus in the lungs will be moved.
During the clapping, the child should breathe normally.

HOW IS CPT DONE?

3. **Vibrating** – after the clapping, vibrating is done over the same area of the lung.

- To do the vibrating, place your hand flat on the area to be vibrated. Stiffen your shoulder and arm so that your whole shoulder area, arm, and hand vibrates (like shivering). Make sure not to use fingertips.
- The vibrating should be done with gentle, downward pressure on the area.
- Start each vibration at the outside edge of the chest or back and move slowly towards the center.
- Have your child take a regular breath and then vibrate as he exhales (breathes out).
- Vibration should be repeated for 5 breaths out.
- If the child can, have him say "SSS" when he breathes out.

4. **Coughing**

- After the mucus has been loosened by clapping and vibrating, encourage your child to cough and spit out as much mucus as possible.

If you see any blood or blood streaks in your child's mucus, call your doctor or













health care nurse

PUTTING IT ALL TOGETHER

- Refer to the pictures on page 40 for the infant, toddler and then for the older child.
- Place the child in the first position.
- Clap for 1 minute and vibrate 5 breaths out.
- Then clap for another minute in this same position, vibrate 5 times again
- Encourage coughing (your child may not be able to cough up something after each position).
- Repeat the steps for each position marked or do as prescribed by your doctor.
- You may want to purchase a percussor for an older child to perform CPT on his own. Several types of percussors are available for home use – ask your home health nurse or doctor for more information.

The “Vest”

It is called the airway clearance system “The Vest” and it consists of an inflatable vest connected by hoses to air pulse generator. The generator rapidly inflates and deflates the vest compressing and releasing the chest wall. This process is called high frequency chest wall oscillation. Cleared to market by the FDA in 1988, the vest has been used in more than 14,000 patients with a variety of pulmonary complications. It has just recently been used by several of our families with children of Batten Disease

	<p style="text-align: center;">Upper lobes Place child in an upright position.</p> <p><input type="checkbox"/> Front - Percuss between the clavicle (collarbone) and the nipple on each side of the chest.</p>	
	<p><input type="checkbox"/> Back - Percuss between the clavicle (collarbone) and the shoulder blade on each side of the back.</p>	
Middle lobes		
	<p style="text-align: center;">Position child with the head down.</p> <p><input type="checkbox"/> Left lingula - Place child on the right side. Percuss on the left side below the armpit.</p>	
	<p><input type="checkbox"/> Right middle - Place child on the left side. Percuss on the right side below the armpit.</p>	
Lower lobes		
	<p style="text-align: center;">Position child with the head down.</p> <p><input type="checkbox"/> Front - Place child on the back. Percuss on the front of the chest in the nipple area and just below.</p>	
	<p><input type="checkbox"/> Back - Place child on the abdomen. Percuss below the shoulder blades and above the lower edges of the rib cage on both sides of the back.</p>	

More than 1500 physicians across the country have prescribed the vest for airway clearance. It offers a number of advantages over CPT and other technique dependent therapy modality.

The Vest:

- Provides consistently effective airway clearance therapy.
- Mobilizes secretions without postural drainage or complicated breathing techniques.
- Eliminates the need for intensive physical involvement by a caregiver.

The Vest includes:

- Training by a clinician from American Biosystems.
- Complete reimbursement services from initial submissions to appeals.
- A comprehensive patient follows up program.
- A warranty for lifetime use.

Impaired airway clearance

- Complications contributing to airway clearance needs:
- Restrictive lung disease
- Dysphasia and aspiration
- Hyperproduction of secretions
- Immobility
- Ineffective cough
- Recurrent respiratory infections
- Mucus plugging and atelectasis
- Respiratory muscle weakness and/or dysfunction

BIPAP, CPAP

BiPAP can be used in the home setting where it can be used as an intermittent positive airway pressure or can be used at night for children who are apneic. CPAP is used with mechanical ventilation but is built into the machine. When BIPAP is used, it is an apparatus that covers the nose or the nose and mouth, to force air into the lungs at a specified rate. It provides positive pressure in the airways throughout the entire respiratory cycle. It maybe delivered through nasal prongs, a mask, or mechanical ventilation. It can be a demand system or a continuous flow system. In the demand system, a valve opens in response to the patient's inspiratory effort. In the continuous flow system, an air oxygen blend flows through a humidifier and a reservoir bag into the system. This requires less work for the patient. CPAP is useful in: pneunonitis and viral pneumonia by improving oxygenation in acute respiratory disorders, sleep apnea by preventing closure of the upper airways, and in the assistance in weaning from the mechanical ventilator, and to prevent alveolar and airway collapse in neonates with respiratory distress syndrome. BIPAP/CPAP increases the functional residual capacity by distending collapsed alveoli. This increases oxygen and decreases the work of breathing. It can cause nausea and vomiting. One should be careful if the child is unresponsiveness or at risk for aspiration.

Suctioning

Suctioning of the child (with or without an artificial airway like a tracheostomy) in the home includes nasal, oropharyngeal and endotracheal which means through the nose, through the mouth, or through a trach site. Suctioning is a component of bronchial hygiene, which involves the mechanical aspirations of secretions from the nose, the mouth, and the trachea. The airway may be in its natural state or artificial or surgically altered like in a laryngectomy. The child may or may not be receiving mechanical ventilation. The procedure includes child preparation, the actual suctioning event, and the follow up care and observation of the child. Child preparation, whenever possible, should be encouraged to clear the airway by directive cough or other airway clearance techniques. Whenever possible, the child/parent should be taught to perform this procedure for himself/or their child. Preoxygenation or hyperinflation prior to the suctioning event will not be routinely indicated for all children cared for in the home. Whenever possible, the child's response to suctioning during his stay in acute care or long term care facility should be made apart of the discharge summary and the Healthcare professional establishing the child in the home should request this information.

The Suctioning Event – Oral, Nasal or Per Trach

Actual introduction of the suction device, (catheter oral suction tip) into the nose, the mouth or into a trach should be in accordance with the clinical practice guidelines. It is common and accepted practice to use "clean" rather than sterile technique during suctioning in the home environment, although scientific evidence to support or discount either technique in home care is lacking. Clean "non-sterile" gloves should be used when endotracheal suctioning is performed. Gloves reduce the risk of inoculants to the child's airway, the risk cutaneous infection in the caregiver, and transmission of organisms to others. Gloves may not be necessary when suctioning through the mouth is performed. At the conclusion of the suctioning event, the catheter or tonsil tip should be flushed by suctioning recently boiled or distilled water to rinse away mucus, followed by the suctioning of air through the device to dry the internal surface and, thus, discourage microbial growth. The outer surface of the device may be wiped with alcohol or hydrogen peroxide. The suction catheter or tonsil tip should be allowed to air dry and then stored in a clean dry place. Suction catheters treated in the manner described may be reused. We recommend the catheters be discarded after twenty-four hours, although no evidence for or against this can be found. Tonsil tips may be cleaned, boiled, and reused indefinitely. If it is feasible to clean the suction device and subject it to high-level disinfection, it may be reused until its integrity is lost. The importance of mechanical cleaning cannot be overemphasized (removal of mucus and other organic material). Following the suctioning event the patient should be monitored for adverse reactions.

Indications

The primary indications for suctioning in a home care setting are a child's inability to adequately clear the airway by coughing. The need for airway clearance is evidenced by:

- More frequent or congested sounding cough

- Coarse rhonchi and expiratory breathing audible to the child and/or caregiver with or without auscultation by stethoscope
- Visible secretions
- Indication by the child that suctioning is necessary
- Suspected aspiration of gastric or upper airway secretion
- Your child is restless or irritable and cannot be calmed
- Your child's breathing is too rapid
- The hollow in your child's neck may pull in
- The skin below the breast bone may pull in
- Your child's face may have a frightened look
- Your infant has difficulty sucking
- Mucus bubbles form at the trach opening
- The mouth looks pale, bluish, or dusky
- Nostrils flare out
- You feel a vibration when you touch your child's chest or back with the palm of your hand
- You hear the chest rattling

Equipment

You will need is a suction machine, suction catheters, normal saline, a basin, and water. While your child is in the hospital, sterile gloves and sterile water will be used for suctioning. At home you may use tap water and freshly washed hands. You want to gather the equipment in one place and turn on your machine. Make sure you wash your hands, have your child be as comfortable as possible in a position that allows you to see the trach clearly, his mouth or his nose. Determine the length of the catheter needed for suctioning by estimating the trach tube length and add a quarter of an inch. You can do this basically the same just by measuring the mouth. Instill normal saline if the secretions are thick (some debate about this issue). Using the thumb and index finger, insert the catheter down the trach opening or through his mouth or nose. Use intermittent suctioning while blowing the catheter between the thumb and index finger when removing. This should take ten seconds or less and then let the child rest. You may need to reinsert the catheter for additional suctioning to clear the airway. Rinse the catheter with water between insertions. Observe your child before, during, and after suctioning for color and ease of breathing.

Helpful Hints

Be thorough when you suction, whether it be oral, nasal, deep suctioning, or per trach. Make sure your child breathes freely before you stop. Suction only as needed. As your child adapts to a tracheostomy he or she will usually need suctioning first thing in the morning, before meals, and the last thing at night before going to bed. This again will depend on each child individually and if there is a presence of an infection.

Cleaning of your suction catheters

What you need to do is wash your hands. Wash and scrub your suction catheters in mild soap. Rinse with tap water. Soak catheters in solution of ½ cup white vinegar and

one quart tap water. Soak for thirty to forty five minutes, rinse with tap water, swing to dry, and store in a clean towel until ready to use.

Tracheal Suctioning or deep nasal suctioning

The proper method of tracheal suctioning. When suctioning, stay within the trach tube as much as possible to suction secretions. Suction beyond the trach tube only if necessary to reach secretions in the lower areas. This precaution is taken to limit the frequency of direct exposure of the lining to the friction and sucking action of the catheter. Also suctioning deeper with the catheter exposes the airways to the possibility of infection. Please take note that the shallow suctioning technique is based on someone having normal lungs. This is a technique to be used in the absence of pneumonia, bronchitis, emphysema or any other lung infection, all of which would require deep suctioning. Do not apply suction while inserting the catheter; only apply suction when pulling the catheter out, with a twisting motion in 10 seconds or less. Use a gentle up and down motion while turning or gently spinning the tube to get mucous on all sides of the tracheal wall. The person suctioning should hold their own breath, when they apply suction to limit the amount of unnecessary time for the procedure since you cannot breathe when suction is being applied. You would want to keep sterile technique as good as you can in the home setting, since suctioning is probably one of the most common sources of contamination and infection in a person with a trach. Sterile gloves and sterile catheters and sterile normal saline should be used with each suctioning, although due to the cost factor, that might be impossible and to do the cleaning techniques may be OK with your physician. Be sure to ask him. The amount of supplies required varies since the need to be suctioned varies from hour to hour and day to day. There are times when suctioning is required 5 – 10 times an hour and other times when it is not needed at all. Since Medicaid has limits on the amounts of supplies it will pay for such as allowing 3 suction catheters a day, sometimes you have to reuse suction supplies while maintaining their sterility.

Endotracheal tubes and ventilators

The ventilator is a machine that helps you breathe. It is also called a respirator or a “vent”. The ventilator helps you by getting oxygen and pushing air into your lungs. To use a ventilator, you must have an ET tube (endotracheal tube) in your nose or throat (for short term use) or a tracheostomy tube in your neck (for a longer period of time). An ET tube is put into your nose or mouth into your trachea (windpipe). The ventilator then attaches to the end of the ET tube. This will allow air to pass from the ventilator into your lungs. The ET tube is held in place with tape or a soft strap. Sometimes the ET tube is called the breathing tube.

Things to know while you have an ET tube:

- An ET tube goes through your vocal cords
- You cannot talk while the ET tube is in your throat
- We will find another way for you to communicate
- Writing notes or hand gestures may help

- After the ET tube is taken out, you will be able to talk again, although you may be hoarse for a short time
- While the ET tube is in place, you may not eat or drink. This is to prevent choking
- The nurse/caretaker will clean your mouth with a wet swab to keep you from having a dry mouth
- The ET tube makes it hard to cough up mucus from your lungs
- Therefore the nurse must suction the mucus out for you
- During suctioning, the nurse/caretaker puts a small tube into the ET tube, then pulls it back out, removing the mucus with it
- Usually, suctioning will make you cough hard.
- You may also feel short of breath
- The ventilator has an alarm system, which alerts the nurse to possible problems. Do not be afraid of the alarms. A nurse will respond and correct any problems.
- The alarm may mean that you need to be suctioned
- The ventilator controls are set to give you the amount of breaths and oxygen that you need
- The ventilator settings change as your condition improves
- This is done by checking the amount of oxygen and carbon dioxide in your blood called ABG's (arterial blood gases). Blood is usually taken from your wrist or elbow.

Weaning from the ventilator and the ET tube:

- Weaning is a process of gradually having you breathe more and more on your own
- Weaning will begin when you are strong enough to begin breathing on your own
- Weaning can be done many different ways
- The nurse/caretaker will explain the method chosen by your doctor
- For most people, weaning takes from 1 day to 1 week
- Try to stay relaxed and breathe deeply during this time
- Oxygen will be given during weaning
- You should be able to cough most of the mucus out of your lungs without suctioning before you are completely off of the ventilator
- When you can breathe without the ventilator
- The ET will be taken out and the weaning will be finished

Prevention

How to detect and prevent respiratory infections.

Because of the debilitated state of a child with Batten Disease, the condition makes them a easy target for respiratory infections, that's why the doctor wants you to be alert for warning signs and take steps to prevent infections.

Warning signs:

Respiratory infections can worsen quickly.

Call the doctor immediately if your child develops any of these signs:

- Fever
- Worsening cough or wheezing
- Increased difficulty breathing
- Changes in the mucus: thicker, increase or decrease in amount, foul smelling or appearing green, yellow, brown, pink, or red
- Unusual fatigue or weakness
- Swollen ankles
- Gain or loss of 5 pounds or more in a week
- Confusion, decreased alertness, or memory loss

Preventive Steps

To help prevent infections, follow these steps:

- Eat a well-balanced, nutritious diet
- Drink 6 glasses of water a day unless the doctor directs otherwise
- Get at least 7-8 hours of sleep a night, and take frequent short naps during the day
- Perform chest physiotherapy and use supplemental oxygen as directed
- Take your medications exactly as prescribed
- Rinse your oral inhaler after each use
- Avoid people who have a cold or the flu. If you can not avoid them, wear a disposable surgical mask when around them
- Check with your doctor about the pneumonia and flu vaccine
- Avoid exposure to inhaled pollutants, such as cigarette smoke, car exhausts, and noxious industrial fumes
- Carefully wash your hands before taking the prescribed medications or setting up oxygen equipment. Also wash them after handling soiled tissues with mucus and after using the bathroom
- Watch areas of water that are standing in sinks, drains or flowers
- Provide good oral hygiene
- Use proper technique when performing oral or deep suctioning is used
- Know how to watch for swallowing safety to avoid aspiration
- Know how to check residuals (aspirate contents from the stomach), listen for bowel sounds before scheduled feedings if a Gtube is in place
- Teach all caregivers proper techniques for treatments

In the latter stages of Batten Disease, children cannot breathe deeply enough to take in enough oxygen to meet their needs. Deep breaths also function to expand both lung tissue and the chest wall. Without occasional expansion, the lungs and chest wall gradually stiffen, with serious consequences for lung health. With the diaphragm to weak and the chest wall to stressed to permit intake of a deep breath, the lungs cannot hold enough air to generate a cough. Coughing plays a vital role in the maintenance of pulmonary health. Forceful expulsion of air from the lungs during a cough, works to loosen, localize, and clear mucus other secretions from the airways.

In Batten Disease, however, as the abdominal muscles become progressively weaker, the ability to expel air with enough force to produce an effective cough is lost; likewise, weakness in the throat muscles gradually impairs the ability to cough. For a cough to take place, the flap of the tissue at the top of the throat, called the glottis, must close in order for an expulsive pressure to build up inside the chest. As throat muscles weaken, the glottis ceases to close tightly. As a consequence, secretions are not cleared effectively from the airways. Normal respiratory function requires open airways to permit the sufficient exchange of oxygen and carbon dioxide, and an effective mechanism to keep airways free from harmful pathogens.

However, with each breath of air, potentially harmful substances such as dust, pollen, viruses, fungi, and bacteria may be inhaled along with vital oxygen. Respiratory muscles play an essential role in maintaining good respiratory health. To prevent inflammation or infection, specialized cells in the airways secrete a steady supply of mucus. Mucous is slightly viscous with mild microbial properties. The chemical and physical properties of normal mucus function to prevent or fight infection and to provide appropriate humidification in the respiratory tract. With the aid of airway cilia (small hair-like structures that move in a sweeping fashion), contaminated mucus is constantly carried from small airways and moves toward central airways. It is then eliminated from the respiratory tract by coughing or swallowing. When mucus is cleared effectively, respiratory infections are usually prevented.

Pulmonary clearance therapy should:

- Clear secretions effectively and consistently
- Preserve lung function
- Reduce infectious exacerbations
- Dependence on antibiotics and other medications
- Reduce need for hospitalization and auxiliary medical treatment
- Delay the disease progression
- Reduce the burden of care
- Enhance the quality of life

Six beliefs and practices in respiratory nursing need to be evaluated.

- Thick secretions that make endotracheal suctioning difficult should be distilled with normal saline into patient's lungs. Two reasons to abandon saline in suctioning routine. One it can lead to a significant decrease in arterial saturation, in some cases, the saturation never returns to presuctioning levels. And two, it increases the likelihood of pulmonary infection by carrying bacteria into the lower airways.
- Changing O2 lines daily. It has been shown that there are no more bacteria present after a week versus changing it daily.
- O2 given via nasal cannula should always be humidified. This was to prevent drying of the nasal sinuses. There is no scientific evidence that O2 at flow rates of 1-4 liters per minute require humidification.
- Pneumonia patients should be turned from side to side to maximize oxygenation by compressing the chest wall, preventing proper ventilation and raising the risk

of atelectasis. A research showed that with a patient lying down on the affected side, may lead to intrapulmonary shunting which is the return of oxygenated blood to the left side of the heart which would result in decreased arterial oxygenation. If the good lung is being lied on, it maximizes oxygenation by using gravity to enhance blood flow to the healthy lung thus creating the best possible match between ventilation and perfusion

- Asthma results from bronchi constriction – It is caused by an inflammatory response to various stimuli, like viral infections, allergens, cold air and exercise and airway hyper responsiveness. Treatment therefore has shifted from bronchodilators to antiinflammatory medications – particularly inhaled corticosteroids like Alupent, Ventolin, Proventil, for acute need for treatment. These are characteristic: shortness of breath, and the force with which air can be expelled from the lungs.
- Cyanosis is a reliable indicator of hypoxia. We need to look at cyanosis in the content of the patient's total blood count

As nurses and parents we have a responsibility to support the research that examines the safety and efficacy of our practices and gives them a sound scientific basis. But that is only half of the battle. We also have to put those research findings into practice. The economics of health care demand it and our children deserve no less.




CPR

Cardio Pulmonary Resuscitation

The respiratory system plays a huge roll in the life of a child with Batten Disease. Because of their immobility and progressive decline, the respiratory system is probably most affected, due to eating, swallowing difficulties to lung problems, which we have discussed in this packet on the respiratory system. A child's life with Batten Disease will be shortened due to the nature of the disease, therefore the family or parent needs to make a decision on their own. It is a very difficult issue to discuss. At what point in time, will the decision be made to *Do Not Resuscitate*? You need to discuss this issue with your family, extended family, physicians, nurse, hospice nurse, attorney, minister, or whom ever you feel comfortable talking with. BDSRA has many support individuals who are more than happy to be there for you in your time of need. Many have already lost children to Batten Disease and can help you get through those hard times. In the pages, which follow this one, are the CPR guidelines for doing CPR on your child if you so wish. Absent pulses, absent respirations and lifeless appearance are the obvious initial signs of collapse of cardiopulmonary function. When these conditions are present, resuscitative measures in order to be effective must be begun immediately and carried out vigorously according to the comprehensive preconceived and practiced plan. However, not every person with presumptive evidence of death should be subjected to such efforts. Hopefully, by understanding what is actually done during CPR, you can better make a decision that will be acceptable for you, your child, and you family. Later on, I will be incorporating some DNR issues in one of the further topics in a packet on Batten Disease for your handbook.




CPR for infants (up to 1 year)

Quick reference

<p>A</p> <p>Arouse:</p> <ul style="list-style-type: none">• rub or tap, do not shake <p>Alert:</p> <ul style="list-style-type: none">• turn on back on hard surface• remove clothing from infant's chest• yell "Help - call 911" <p>Airway:</p> <ul style="list-style-type: none">• head-tilt / chin-lift	
<p>B</p> <p>Check for breathing:</p> <ul style="list-style-type: none">• look, listen, and feel <p>Give 2 breaths</p> <ul style="list-style-type: none">• if chest rises, go to C <p>If chest does not rise:</p> <ul style="list-style-type: none">• reposition head and try again• if chest still does not rise, give 5 chest compressions• look in mouth• repeat until chest rises	
<p>C</p> <p>Check for signs of circulation.</p> <p>If no circulation, do chest compressions:</p> <ul style="list-style-type: none">• place 2 fingers on the breast bone about 1 finger-width below the nipple line• press breastbone down 1/3 to 1/2 the depth of the chest• 5 compressions to 1 breath• 100 compressions per minute <p>After 20 cycles (about 1 minute):</p> <ul style="list-style-type: none">• call 911 if you are alone• check for breathing and circulation• continue CPR as needed• check for breathing and other signs of circulation every few minutes	




CPR for children (1 to 8 years)

Quick reference

<p>A</p> <p>Arouse: rub or tap, do not shake</p> <p>Alert: turn on back on hard surface remove clothing from child's chest yell "Help - call 911"</p> <p>Airway: head-tilt / chin-lift</p>	 A line drawing showing a person's hands performing a head-tilt/chin-lift maneuver on a child. The person is on the left, and the child is on the right, lying on their back. The person's hands are positioned to lift the child's chin and tilt their head back.
<p>B</p> <p>Check for breathing: look, listen, and feel</p> <p>Give 2 breaths: if chest rises, go to C</p> <p>If chest does not rise: reposition head and try again if chest still does not rise, give 5 chest compressions look in mouth repeat until chest rises</p>	 A line drawing showing a person performing rescue breaths on a child. The person is on the left, and the child is on the right, lying on their back. The person's mouth is over the child's mouth, and they are blowing air into their lungs.
<p>C</p> <p>Check for signs of circulation.</p> <p>If no circulation, do chest compressions: place the heel of 1 hand between the nipples on the breastbone press breastbone down 1/3 to 1/2 the depth of the chest 5 compressions to 1 breath 100 compressions per minute</p> <p>After 20 cycles (about 1 minute): call 911 if you are alone check for breathing and circulation continue CPR as needed check for breathing and other signs of circulation every few minutes</p>	 A line drawing showing a person performing chest compressions on a child. The person is on the left, and the child is on the right, lying on their back. The person's hands are positioned on the child's chest, and they are pressing down.

CPR (age 8 and older)

Quick reference

<p>A</p> <p>Arouse: rub or tap, do not shake</p> <p>Alert: call 911 turn on back on hard surface remove clothing from person's chest</p> <p>Airway: head-tilt / chin-lift</p>	 A line drawing showing a person's head being tilted back and their chin being lifted upwards. An arrow points to the chin, and another arrow points to the neck area, indicating the direction of movement.
<p>B</p> <p>Check for breathing: look, listen, and feel</p> <p>Give 2 breaths if chest rises, go to C</p> <p>If chest does not rise: reposition head and try again if chest still does not rise, give 5 chest compressions look in mouth repeat until chest rises</p>	 A line drawing showing a person's head tilted back with their mouth open. A rescuer is shown blowing into the person's mouth. An arrow points to the mouth, and another arrow points to the neck area, indicating the head position.
<p>C</p> <p>Check for signs of circulation.</p> <p>If no circulation, do chest compressions: place the heel of 1 hand between the nipples on the breast bone; put other hand over the first, keeping your fingers off the chest press breastbone down 1/3 to 1/2 the depth of the chest 15 compressions to 2 breaths 4 cycles per minute</p> <p>After 4 cycles (about 1 minute): check for breathing and circulation continue CPR as needed check for breathing and other signs of circulation every few minutes</p>	 A line drawing of a person lying on their back. A rescuer's hands are shown on the person's chest, with arrows pointing to the location of the hands (between the nipples on the breastbone). The rescuer's hands are positioned one on top of the other, with fingers interlocked.

I am hopeful that this packet on the respiratory system has been helpful to you and your family. I have tried to include all of the different aspects of the system, and what happens as a child with Batten Disease begins having respiratory complications. Remember, each child is different. They react to situations just like any other child. They are growing and will go through all of normal childhood illnesses: colds, flu, early puberty, early menstruation, behavioral problems, etc. But they are your children – you know them best – let instincts guide you to the right decisions. And again, we are here to help you and answer any questions you may have in the care of your child and your family.

Thank You!

Nancy Carney RN
Medical Liaison/Educator
BDSRA

RESPIRATORY SYSTEM GLOSSARY

ABG's - Arterial blood gases; blood taken from an artery to measure the pH (acid-base system); the oxygen and the carbon dioxide levels of the body; very important in crisis situations, when the buffer system is put to a test; added oxygen or bicarbonate can be added to the body through IV access and stabilize the body

Abscess - a localized collection of pus in any part of the body - symptoms usually are redness, swelling, warm to the touch, and pain

Accessory muscles - auxiliary; assisting; muscle which assists other muscles to do its function

Acid-base - the mechanisms by which the acids and alkalies are kept in a state of equilibrium so that the hydrogen ion concentration of the arterial blood is maintained between pH of 7.35 and 7.45, this is accomplished by action of the buffer system (alkali reserve) of the blood, and the functioning of the respiratory and urinary systems; disturbances in acid-base balance results in acidosis or alkalosis

Acute - having rapid onset, sharp, severe symptoms and a short course; not chronic

Adenoids - lymphatic tissue forming a prominence on the wall of the pharyngeal recess of the nasopharynx; usually referred to as tonsils and adenoids

Adrenal glands - triangular shaped body adjacent to and covering the superior surface of each kidney; it is a gland of internal secretions producing hormones essential for life

Afferent - carrying impulses toward the center, as when a sensory nerve carries a message toward the brain

Alveolar sacs - pertaining to the alveolus which are air sacs (cells) in the lungs

Anaerobic - a microorganism which thrives best or lives only without oxygen; having the power to use oxygen for metabolism from oxygen compounds

Anastomosis - an opening; a communication between two vessels; the surgical or pathologic formation of a passage between any two normally distinct spaces or organs; an end-to-end union or joining together or intercommunication of parts of any network or set of fibers such as nerves, or connective tissue fibers

Anesthesia - partial or complete loss of sensation, with or without loss of consciousness; as result of injury, disease, or administration of a drug or gas

Anoxia - deficiency of oxygen-carrying power of the blood; diminished oxygen in the arterial blood despite normal ability of the blood to contain and carry oxygen (oxygen capacity); may be due to reduced oxygen supply, respiratory obstruction, reduced surface area in lungs for exchange of gases (as in pneumonia) or inadequate respiratory movements

Anterior - before or in front of as the front side of the body compared to the posterior or backside of the body

Anti-inflammatory - pertaining to a substance or procedure that counteracts or reduces inflammation; an anti-inflammatory drug or agent

Antipyretic - an agent that reduces elevated temperatures like Tylenol, Aspirin, or Ibuprofen

Aorta- the main trunk of the arterial system of the body, it is about 3 cm in diameter at its origin; it arises from the upper surface of the left ventricle, passes upward as the

ascending aorta, turns backward and to the left (aortic arch) at about the level of the fourth thoracic vertebrae and then passes downward as the descending aorta which is divided into the thoracic and abdominal aorta, the abdominal aorta terminates at its division into the two common iliac arteries, at its exit from the ventricle; the aortic opening is guarded by three semi-lunar valves

Apices - the apex or tip of anything as in the (heart which can be felt at the fifth left intercostal space), approximately $3 \frac{1}{2}$ inches from the middle of the sternum (bone in between the ribs or the breast bone)

Apnea - cessation of breathing, usually of a temporary nature; may result from a reduction in stimuli to the respiratory center as in over breathing in which the carbon dioxide content of the blood is reduced, or from failure of the respiratory center to discharge impulses as occur when the breath is held voluntarily

Apneustic - a sustained respiratory, inspiratory effort, due to surgical removal of the upper portion of the pons

ARDS - adult respiratory distress syndrome - like SIDS in children, where a patient has difficulty maintaining respirations; a set of symptoms including decreased compliance of the lung tissue, pulmonary edema, and acute hypoxemia; may be caused by chest trauma, shock, hemorrhage, repeated blood transfusions, drug overdose, aspiration of gastric contents, or near drowning; may require mechanical ventilation during the acute phase

Arrhythmia - irregular heartbeat, caused by disturbances, either pathological or physiological; in discharge of cardiac impulses from the SA node or their transmission through conductile tissue of the heart

Artery - one of the vessels carrying blood from the heart to the tissues - frequently is nearly empty after death; the arteries carry the blood from the right and left ventricles of the heart to all parts of the body; there are two sets, the pulmonary and the systemic - the pulmonary artery carries the venous blood from the right ventricle to the lungs, the systemic system begins as the aorta from the left ventricle to all parts of the body

Asbestos - a fibrous form of magnesium and calcium silicate, a lung disease due to protracted inhalation of asbestos particles

Aseptic - a condition free from germs, free from infection; sterile; free from any form of life

Aspiration - to draw in or out as by suction; foreign bodies may be aspirated into the nose, throat, or lungs on inspiration; seen in aspiration pneumonia when food or fluids are inadvertently inhaled into the lungs usually due to poor swallowing ability

Assessment - an evaluation of a patient's condition; the process of gathering data needed to formulate a nursing diagnosis; in a problem-oriented medical record, an examiner's evaluation of a disease or condition based on the patient's subjective report of the symptoms and course of the illness or condition and the examiner's objective findings, including data obtained through laboratory tests, physical examination and medical history

Asthma - paroxysmal dyspnea accompanied by the adventitious sounds caused by a spasm of the bronchial tubes or due to swelling of their mucous membrane; status asthmaticus is a more or less continuous asthmatic state which may last for hours or days; no age is

exempt but occurs most frequently in childhood or early childhood - may be caused by allergens or infections of the respiratory system; treatment is with antibiotics if infection, inhalers, expectorants, steroids, or sedatives

Atelectasis - condition in which lungs of a baby at birth remain unexpanded, may be partial or complete; a collapsed or airless condition of the lungs; may be caused by obstruction by foreign bodies, mucous plugs or excessive secretions or by compression from without as by tumors, aneurysms, or enlarged lymph nodes; it sometimes is a complication following abdominal surgeries due to the fact to take a deep breath hurts on inspiration, so the patient does not fully expand his lungs and the air sacs at the bottom of his lungs become stagnate

Atrium - a cavity or sinus - the upper chambers of the half of the heart, the right atrium receives deoxygenated, dark red blood from the entire body (except the lungs) through the superior and inferior vena cava and coronary sinus, the left atrium receives oxygenated red blood from the lungs through the pulmonary veins, blood passes from the atria to the ventricles through the atrioventricular openings

Audible - being able to hear as in an audible wheeze without the use of a stethoscope

Auscultation - the process of listening for sounds produced in some of the body cavities, especially the chest and abdomen, in order to detect or judge some abnormal condition

B

Bacterial - unicellular, plant-like microorganisms, lacking chlorophyll, so it cannot carry on photosynthesis; if they live on living organisms they are called parasites; if bacteria produce disease in their host they are called pathogenic; bacteria, if causing disease need to be treated quickly

Bicarbonate - a salt resulting from the incomplete neutralization of carbonic acid or from the passing of an excess of carbon dioxide into a solution of a base, an alkali - you need to have an equal acid-base balance in the blood to function properly; part of the buffer system

Bifurcation - a separation into two branches; the point of forking

Brainstem - all of the brain except the cerebrum and cerebellum, where all of the functions of the body stem from, including respirations, heat and cold, and all of the motor skills where the brain tells a toe to move for example, and many more

Bronchi - the primary divisions of the trachea, divides opposite the 3rd dorsal vertebrae; the right bronchus is shorter and more vertical than the left; they penetrate the lungs, one for the right and the other one for the left lung and terminate in the bronchioles or bronchial tubes

Bronchiectasis - dilatation of a bronchus or bronchi, usually secreting large amounts of offensive pus; dilatation may be in an isolated segment or spread throughout the bronchi; acquired or congenital; on one or both sides of the chest; acquired usually secondary to obstruction or infections, as bronchopneumonia, chronic bronchitis, tuberculosis, or whooping cough; symptoms include cough, dyspnea, expectoration of foul secretions

especially in the morning or when changing position; on standing, sputum separates into 3 layers, a very thick layer, a greenish colored layer and a frothy layer; antibiotics for treatment and postural drainage, or aerosols for bronchodilatation or bronchospasm

Bronchioles - one of the smaller subdivisions of the bronchi, they lack cartilage; the last division of the bronchial tree, they are branches of terminal bronchioles and lead to alveolar ducts leading to the alveoli

Bronchitis - inflammation of the bronchial mucous membranes; infectious agents, especially of the flu virus or strep, staph, or pneumococcus is often preceded by the common cold; predisposing factors are exposure, chilling, fatigue and malnutrition, physical and chemical agents such as dust, fumes, etc.; also allergic reactions may also be of concern

Bronchodilators - medications that dilates the bronchus to ease breathing, by use of pills or inhalers

Bronchoscopy - examination of the bronchi through a bronchoscope, which examines the interior of the bronchus

Bronchospasm - a spasm of the bronchus

C

Capillary - minute blood vessels, finer than a hair, carrying blood and forming the capillary system; they connect the smallest arteries (arterioles) with the smallest veins (venules)

Carbon monoxide - a colorless, odorless, poisonous gas produced by the combustion of carbon or organic fuels in a limited oxygen supply; it combines irreversibly with hemoglobin, preventing the formation of oxy-hemoglobin and reducing the oxygen supply to the tissues; prolonged exposure to high levels result in asphyxiation

Cardiac - pertaining to the heart or to the heart opening into the stomach; having heart disease

Cardiac arrest - the complete cessation of contractions of the heart, resulting in circulatory failure, unconsciousness, respiratory arrest and death; it requires immediate CPR, open cardiac massage, or stimulation of the heart with an electric current

Cardiac output - the volume of blood expelled by the ventricles of the heart, equal to the amount of blood ejected at each beat (the stroke output) multiplied by the number of beats in the period of time used in the computation; a decreased output at rest is usually indicative of a late stage in abnormal cardiac performance; its failure to increase during exercise occurs much earlier in a malfunctioning heart

Cardiovascular - the network of structures, including the heart and the blood throughout the body; the system includes thousands of miles of vessels, capillaries and veins and is vital in maintaining a stable atmosphere; numerous control mechanisms of the system assure that the blood is delivered to the structures where it is most needed and at the proper rate; the system delivers nutrients and other essential materials to the fluids surrounding the cells and removes waste products, which are conveyed to excretory organs, as the kidneys and the intestine; the cardiovascular system functions in close

association with the respiratory system, transporting oxygen inhaled into the lungs and conveying carbon dioxide to the lungs for expiration

Carina - a keel-like structure; a ridge at the lower end of the trachea separating openings of the two bronchi

Carotid artery - the principal artery of the neck, it divides into the right and left branches; pertaining to any carotid part, carries blood to the brain only

Cartilage - a type of dense connective tissue consisting of cells embedded in a ground substance or matrix, which is firm and compact rendering it capable of withstanding considerable pressure or tension; having a white or gray color, is semi-opaque and is nonvascular; the cells lie in cavities and may be single or in groups of two, three, or four; it consists of a part of the skeleton occurring in the costal cartilages of the ribs, the nasal septum, in the external ear and lining the eustachian tube, in the wall of the larynx, in the trachea and bronchi, between bodies of vertebrae, and covering the articular surfaces of bones

Cartilaginous - pertaining to or consisting of cartilage

Cavities - a hollow space, such as a body organ or the hole in a tooth

Cerebral spinal fluid - a watery cushion protecting the brain and spinal cord from shock; shrinking or expanding of the cranial contents is usually quickly balanced by an increase or decrease of this fluid; possibly cell nourishment and the removal of waste are minor functions

Chemoreceptor - a sense organ or sensory nerve ending (as a taste bud), which is stimulated by a chemical substance

Chest physiology - a group of techniques including postural drainage, chest percussion, and vibration, and coughing and deep breathing maneuvers; used together to mobilize and help eliminate lung secretions; help re-expand lung tissue, and promote efficient use of respiratory muscles

CHF - congestive heart failure - an abnormal condition characterized by circulatory congestion caused by cardiac disorders, especially myocardial infarction (heart attack) of the ventricles; this condition usually develops chronically in association with the retention of sodium and water by the kidneys - symptoms include shortness of breath, swelling of the ankles up to the hips sometimes, blood pressure elevation, and not able to breath very deeply due to the increased amount of fluid in the lungs - will hear rales (crackling sounds) in the lungs; may need mechanical ventilation for the first few days

Chromium - a hard metallic element; may be important in human nutrition, especially in carbohydrate metabolism

Chronic - long drawn out; applied to a disease that is not acute

Cilia - hair-like processes projecting from epithelial cells (as in the bronchi), which wave mucous, pus, and dust particles upward

Circulatory - movement in a circular course, as in the blood; the blood leaving the left ventricle entering the aorta, from which it escapes into the various large arteries; it thus reaches the coronary arteries of the heart itself and the arteries of the head, body wall, abdominal viscera, and extremities, passing through the various capillary systems; it is

gathered into veins of which there are two systems, the vena cava - inferior and superior; once the blood enters the veins including the coronary veins, the blood enters the right atrium, passes through the right ventricle and is forced out into the pulmonary artery; the pulmonary capillary system (reoxygenates the blood,) drains by way of the pulmonary veins into the left atrium and thence into the left ventricle into the aorta and throughout the body again

Circumoral - encircling the mouth; becomes white or cyanotic (blue) around the mouth; contrasting vividly with color of the face; especially seen in scarlet fever

Clavicles - the collarbone; a bone curved like the letter f, which articulates with the sternum and the scapula

CNS - central nervous system - one of the two main divisions of the nervous system of the body, consisting of the brain and the spinal cord; it processes information to and from the peripheral nervous system and is the main network of coordination and control for the entire body; the brain controls many functions and sensations, as sleep, muscular movement, hunger, thirst, memory, and emotions; the spinal cord extends various types of nerve fibers from the brain and acts as a switching and relay terminal for the peripheral nervous system; there are 12 pairs of cranial nerves emerging directly from the brain; sensory and motor nerves of the peripheral nervous system leave the spinal cord separately between the vertebrae but unite to form 31 pairs of spinal nerves containing sensory fibers and motor fibers

CO₂ - carbon dioxide - a colorless, pungent and acid-tasting gas; heavier than air, generally produced in the combustion, decomposition or fermentation of carbon, or its compounds, and found in the air and exhaled by all animals; the final product of combustion of carbon in food, which the body exhales through the lungs, or eliminates through the kidneys in urine, or in perspiration through the skin; although a waste product, in small quantities in inspired air, it stimulates respirations in greater quantities; it produces an uncomfortable degree of hyperpnea with mental confusion, although not supposed to be poisonous, it will cause death by suffocation

Colonization - a mass of microorganisms in a culture that originates from a single cell; examples-smooth, rough, and dwarf colonies

Communicable - any disease transmitted from one person to another, either directly by contact with excretions from the body or indirectly via substances or inanimate objects, as contaminated drinking glasses, toys, or water, or as vectors like flies, mosquitoes, ticks, or other insects; need to identify the organism, prevent its spread, protect others against it, and treat the infected person

Compensatory - making up for a defect, as cardiac circulation competent to meet demands made upon it, regardless of valvular defect

Compliance - fulfillment by the patient of the prescribed course of treatment; the quality of yielding to pressure

Concha - the outer ear or the pinna; one of the three nasal conchas

Congestion - the presence of an excessive amount of blood in an organ or in tissue

Connective tissue - one of the four main tissues of the body, concerned primarily with supporting bodily structures and binding parts together; they also are involved in other functions such as food storage, blood formation, and defensive mechanisms of the body

Consolidation - the combining of separate parts into a single whole, a state of solidification; in medicine, the process of becoming solid, as when the lungs become stiff or firm and inelastic in pneumonia

Contamination - the introduction of disease germs or infectious materials into or on sterile objects

Copious - large amounts; said of copious amounts of secretions (mucous) of the lungs

Corticosteroids - any one of the natural or synthetic hormones associated with the adrenal cortex, which influences or controls key processes in the body, as carbohydrate and protein metabolism, electrolyte and water balance, and the functions of the cardiovascular system, skeletal muscle, kidneys, and other organs; can prevent or reduce inflammation by inhibiting swelling

Cricoid - shaped like a signet ring, the lowermost cartilage of the larynx; the broad portion or lamina being posterior; the anterior portion forming the arch

Croup - disease characterized by suffocative and difficult breathing, laryngeal spasm which we hear as wheezing, and sometimes by the formation of a membrane, spasm of the glottis; occurs in children - where inhalation of steam usually helps the most

Crowing - children with croup usually are referred to as crowing with their wheezy bark-like cough

Cyanosis - slightly bluish, grayish, slate-like or dark purple discoloration of the skin due to presence of abnormal amounts of reduced hemoglobin in the blood; when the entire body is affected, the color is dusky leaden; caused by deficiency of oxygen and excess of carbon dioxide in the blood; caused by gas or any condition interfering with entrance of air in the respiratory tract; also by overdoses of certain drugs, or any form of asphyxiation

Cystic Fibrosis - an inherited disorder of the exocrine glands, causing those glands to produce abnormally thick secretions of mucus; the glands most affected are those in the pancreas, the respiratory system, and the sweat glands; usually recognized in infancy or early childhood; occurring chiefly among Caucasians; no none cure; treatment is directed at the prevention of respiratory infections, which are the most frequent cause of death

D

Debilitation - feebleness, weakness, or loss of strength

Diaphoresis - the profuse secretion of sweat associated with an elevated body temperature, physical exertion, exposure to heat and mental or emotional stress

Diaphragm - a dome-shaped musculofibrous partition that separates the thoracic and the abdominal cavities; the convex cranial surface of the diaphragm forms the floor of the thoracic cavity, and the concave surface forms the roof of the abdominal cavity; the diaphragm aids respiration by moving up and down; during inspiration it moves down and

increases the volume of the thoracic cavity, during expiration it moves up, decreasing the volume

Diffusion - the process in which ions and molecules move from an area of greater concentration to an area of lower concentration, resulting in an even distribution of the particles in the fluid

Diphtheria - an acute contagious disease caused by a bacteria characterized by the production of a systemic toxin and a false membrane lining of the mucous membrane of the throat; damaging to the tissues of the heart and central nervous system, and the dense pseudo membrane in the throat may interfere with eating, drinking, and breathing; if untreated many times are fatal, patient needs isolation, rest, fluids, antibiotics

Diuretics - a drug tending to promote the formation and excretion of urine

Dysphagia - difficulty in swallowing, commonly associated with obstructive or motor disorders of the esophagus

Dyspnea - a shortness of breath or a difficulty in breathing that may be caused by certain heart conditions, strenuous exercise, asthma or anxiety

E -

Efficacy - the maximum ability of a drug or treatment to produce a result, regardless of the dosage.

Elasticity - the ability of tissue, as muscle tissue; to regain its original shape and size after being stretched, squeezed, or otherwise deformed

Emphysema - an abnormal condition of the pulmonary system, characterized by over inflation and destructive changes of the alveoli walls resulting in a loss of lung elasticity and decreased gases; symptoms include shortness of breath, dyspnea, cough, cyanosis, orthopnea, unequal chest expansion, tachypnea, tachycardia and an elevated temperature; anxiety, carbon dioxide narcosis with a decreased pH, increased pco₂, restlessness, confusion, weakness, anorexia, congestive heart failure, pulmonary edema, respiratory failure are common in advanced cases; treatment may include airway maintenance, humidified oxygen, bronchodilators, antibiotics, expectorants, corticosteroids, postural drainage (cupping and vibration), and IPPB (incentive positive pressure breathing); sedation is to be avoided, as more sedatives depress respiratory function; patient is taught breathing exercises and encouraged to drink plenty of fluids; activity to the patients tolerance, fatigue, constipation, and upper respiratory tract infection and irritation are to be avoided; may have home oxygen and breathing treatments

Empyema - an accumulation of pus in a body cavity, especially the pleural space; as a result of a bacterial infection, as pleurisy or tuberculosis; usually removed by a surgical incision, aspiration and drainage; antibiotics are administered to correct the underlying infection

Endothelial - the layer of squamous epithelial cells that lines the heart, the blood and the lymph vessel, and the serous cavities of the body; it is highly vascular, heals quickly and is derived from the mesoderm

Endotracheal - within or through the trachea; used in mechanical ventilation of an endotracheal tube put in the mouth, nose or throat to provide adequate oxygen for the body

Epiglottis - the cartilaginous structure that overhangs the larynx like a lid and prevents food from entering the larynx or the trachea while swallowing

Epithelium - the covering of the internal and external organs of the body, including the lining of arteries; it consists of cells bound together by connective material and varies in the number of layers and the kinds of cells

Esophagus - the muscular canal extending from the pharynx to the stomach; it begins at the neck at the inferior border of the cricoid cartilage, opposite the sixth cervical vertebra, and descends to the cardiac sphincter of the stomach in a vertical path with two slight curves; it is the narrowest part of the digestive tube and its most constricted at its origin and at the point where it passes through the diaphragm; the esophagus is composed of fibrous, muscular, and sub mucous coats, and is lined with mucous membrane

Ethmoidal - one of the numerous small, thin walled cavities in the ethmoid bone of the skull, rimmed by several small bones; they are lined with mucus membrane continuous with that of the nasal cavity and lie between the upper part of the nasal cavities and the orbits; they are divided bilaterally into anterior, middle, and posterior cavities; the anterior and the middle cavities open into the middle meatus (opening) of the nose, the posterior opens into the superior meatus

Eustachian - a tube lined with mucous membranes that joins the nasopharynx and the tympanic cavity, allowing equalization of the air pressure in the inner ear with atmospheric pressure

Exacerbation - an increase in the seriousness of a disease or disorder as marked by greater intensity of the patient's signs and symptoms

Expectorant - a substance that promotes the ejection of mucus or other exudates from the lungs, bronchi, and trachea; helps reduce the viscosity of pulmonary secretions or by decreasing the force with which exudates adhere to the lower respiratory tract

Expiration - breathing out, normally a passive process, depending on the elastic qualities of lung tissue and the thorax

Expulsion - having a tendency to drive something out

F

Facilitates - the enhancement or reinforcement of any action or function so that it is carried out with increased ease; the phenomenon whereby two or more afferent impulses that individually are not strong enough to elicit a response in a neuron can collectively produce a reflex; discharge greater than the sum of the separate responses; the process of lowering the threshold potential of a neuron by the repeated passage of an impulse along the same pathway

Fatigue - a state of exhaustion or a loss of strength or endurance, as many follow strenuous physical activity, loss of ability of tissues to respond to stimuli that normally

evoke muscular contraction or other activity; muscle cells generally require a refractory or recovery period after activity, during which time cells restore their energy supplies and excrete metabolic waste products

Flora - plant life occurring or adapted for living in a specific environment as flora in the intestine

Fulcrum - the stable point or the position on which a lever, as the ulna and the femur, turns in moving an object; numerous common movements of the body, as raising the arm and walking, are combinations of lever actions involving fulcrums

Fungi - a simple parasitic plant that, lacking chlorophyll, is unable to make its own food and is dependent on other life forms; a simple fungus reproduces by budding; multi-cellular fungi reproduce by spore formation

G

Glottis - a slit-like opening between the true vocal cords

GT feedings - gastrostomy feeding - the introduction of a nutrient solution through a tube that has been surgically inserted in to the stomach through the abdominal wall

H

Heimlich maneuver - an emergency procedure for dislodging a bolus of food or other obstruction from the trachea to prevent asphyxiation

Hemoglobin - a complex protein - iron compound in the blood that carries oxygen to the cells from the lungs and carbon dioxide away the cells to the lungs

Hemoptysis - coughing up of blood from the respiratory tract, blood streaked sputum often occurs in minor upper respiratory

Hering-Breuer reflexes - inhibitory and excitatory impulses that maintain the rhythm of respiration and prevent the over distention of alveoli; the impulses originate in stretch receptors of the bronchi and bronchioles, travel via afferent fibers of the vagus nerve, to the medullary respiratory centers, and back to the motor neurons to the respiratory muscles of the chest; they are stimulated by distention of the airway increased intra-tracheal pressures; or pulmonary inflation; the inflation reflex stops inspiration and stimulates expiration, the deflation reflex inhibits expiration and brings on inspiration; these reflexes are hyperactive in conditions of restrictive ventilatory insufficiency

Hilum - a depression or pit at that part of an organ where vessels and nerves enter

Hydrogen - a gaseous univalent element; the simplest and lightest of the elements and is normally a colorless, odorless, highly inflammable gas; a component of numerous compounds, many of them produced by the body; is crucial in the metabolic interaction of acids, bases, and salts within the body and in the fluid balance necessary for the body to survive

Hydrogen ion - the positively charged nucleus of a hydrogen atom; the relative proportion of hydrogen ions in a solution; the factor responsible for the acidic properties of a solution

Hypercapnea - greater than normal amounts of carbon dioxide in the blood

Hypertension - a blood pressure elevation of greater than 140/90

Hyperthermia - a higher than normal body temperature

Hyperventilation - a pulmonary ventilation rate that is greater than that metabolically necessary for the exchange of respiratory gases; it is the result of an increased frequency of breathing; an increased tidal volume, or a combination of both, and causes excessive intake of oxygen and the blowing off of carbon dioxide, hypocapnia and respiratory alkalosis then occur leading to chest pain, dizziness, faintness, numbness of the fingers and toes, and psychomotor impairment; causes of hyperventilation include asthma, or early emphysema, increased metabolism owing to exercise, fever, hyperthyroidism or infections, lesions of the central nervous system, as in cerebral thrombosis, encephalitis, head injuries or meningitis, drugs such as salicylates, difficulties with mechanical respirators, and psychogenic factors, as acute anxiety or pain

Hypocapnea - not enough carbon dioxide in the blood

Hypotension - a blood pressure decrease below 100/60 - not adequate for normal perfusion and oxygenation of the tissues

Hypothermia - an abnormal and dangerous condition in which the temperature of the body is below 95 degrees usually due to prolonged exposure to the cold, respirations are slow and shallow and the heart rate is faint and slow; very pale and appears dead

Hypoventilation - abnormally reduced rate and depth of inspiration; it occurs when the volume of air which enters the alveoli and takes part in gas exchange is not adequate for the metabolic needs of the body; may be caused by uneven distribution of inspired air, as in bronchitis, morbid obesity, neuromuscular or skeletal disease affecting the thorax, decreased response of the respiratory center to carbon dioxide, and by reduced functional lung tissue, as in atelectasis, emphysema, and pleural effusion; the result of unresolved hypoventilation is hypoxia, hypercapnia, pulmonary hypertension with cor pulmonale and respiratory acidosis; treatment aims to correct underlying cause

Hypovolemia - low amount of plasma in the blood, which can cause circulatory dysfunction and inadequate tissue perfusion

Hypoxemia - an abnormal deficiency of oxygen in the arterial blood; symptoms of acute hypoxemia are cyanosis, restlessness, stupor, coma, Cheyne-Stokes breathing, or apnea, increased blood pressure, tachycardia, and an initial increase in cardiac output which later falls, resulting in hypotension, ventricular fibrillation, asystole; it stimulates red cell production by the bone marrow

Hypoxia - oxygen deficiency caused by reduced oxygen-carrying capacity (anemic hypoxia), insufficient oxygen in inspired air (hypoxic hypoxia), or inadequate blood flow to transport oxygen (stagnant hypoxia), infections or in bronchitis, more profuse bleeding may occur with specific infections, lung abscess, tuberculosis, or cancer

I

Immune - the quality of being in susceptible to or unaffected by a particular disease or condition

Immuno compromised - even though a person's tissues and immuno system is deficient in some areas, it has compromised into functioning adequately

Impulses - the electrochemical process involved in neural transmission

Incentive spirometer - a device which the patient initiates a deep breath, to increase lung volume of air for better oxygen/carbon dioxide exchange

Infarction - the development and formation of a localized area which becomes necrotic (dead) in a tissue, organ, or vessel or a part resulting from tissue anoxia - caused by the interruption of a blood supply to the area, or less frequently by circulatory stasis produced by an occlusion of a vein that ordinarily carries blood away from the area

Inflammation - the protective response of the tissues of the body to irritation or injury; may be acute or chronic; signs are redness, heat, swelling and pain, accompanied by loss of function

Influenza - a highly contagious infection of the respiratory tract transmitted by airborne droplet infection; will see sore throat, cough, fever, muscle pain, and weakness, incubation period is 1-3 days and the onset is sudden with chills, fever and general malaise, treat symptomatically with bed rest, fluids, and aspirin

Innervate - distribution and action of the nervous system, nerve supply and nerve stimulation of a part

Inoculate - to introduce a substance into the body to produce or to increase immunity to the disease or condition associated with the substance; it is introduced by making multiple scratches in the skin after placing a drop of the substance on the skin; by puncture of the skin with an implement bearing multiple short tines or by intra-dermal, subcutaneous or intra-muscular injection

Inspiration - the act of drawing air into the lungs in order to exchange oxygen for carbon dioxide; the end product of tissue metabolism; the major muscle of inspiration is the diaphragm, the contraction of which creates a negative pressure in the chest, causing the lungs to expand and air to flow inward

Invasive - characterized by a tendency to spread, infiltrate and intrude

IPPB - incentive positive pressure breathing - a machine which forces air into the lungs, medication like broncho dilators can be added to improve the patients breathing; used with patients with asthma, emphysema, and other respiratory problems

Isthmus - a narrow connection between two larger bodies or parts as the isthmus of the auditory tube in the ear which connects the bony and the cartilaginous parts of the tube

J

K

Kyphosis - an abnormal condition of the vertebral column, characterized by increased convexity in the curvature of the thoracic spine as viewed from the side; may be caused by

tuberculosis or rickets; may be moderate back pain; often undiagnosed; conservative treatment is spine-stretching exercises, sleeping without a pillow and a board under the mattress; a brace may be used or rare surgery is required

L

Laryngeal - pertaining to the larynx, as in reflex which is a cough as a result of irritation of the larynx

Laryngectomy - removal of the larynx to treat cancer of the larynx; in a partial laryngectomy only the vocal cords are removed and the patient has a temporary tracheostomy; in a total laryngectomy the larynx is also removed and the tracheostomy is permanent

Laryngitis - inflammation of the mucous membrane lining the larynx accompanied by swelling of the vocal cords with hoarseness and or loss of voice

Laryngopharynx - one of the three regions in the throat, extending from the hyoid bone to its termination in the esophagus

Laryngoscopy - an lighted instrument for examining the larynx

Larynx - the organ of voice that is part of the air passage connecting the pharynx with the trachea; it produces a large bump in the neck called the Adam's apple, and is larger in men than in women; lined with mucous membranes that is continuous with that of the pharynx and the trachea

Lateral - pertaining to the side or lateral aspect

Lingula pulmonis sinispr - a tongue-like process of the left lung

Lobar - a bronchus extending from a primary bronchus in a segmental bronchus into one of the lobes of the right or left lung

Lobules - a small lobe as a soft, lower, pendulous part of the external ear

LOC - level of consciousness - awareness of one's surroundings

Lungs - one of a pair of light spongy organs the thorax; constituting the main component of the respiratory system; are elastic, and the main mechanisms of the body for inspiring air from which oxygen is extracted for the arterial blood system and for exhaling carbon dioxide dispersed from the venous system; the right lung has three lobes and the left lung with two lungs

Lymphatic - the lymphatic system of the body, consisting of a vast network of tubes transporting lymphs which is a thin opalescent fluid originating in many organs and tissues that is circulated through the vessels and filtered by the lymph nodes; lymph enters the blood stream at the junction of the internal jugular and subclavian veins; similar to plasma

M

Mechanical ventilation - sometimes called the vent, or ventilator, used in respiratory therapy to provide assisted respiration and positive pressure breathing; levels of oxygen

and number of breaths per minute, etc., can be altered according to ABG's to assist the patient while he improves in his condition

Mediastinal shift - where an increase in the amount of fluids in the lung cavity, causes the mediastinum to shift to the left or right depending on which side the fluids are more increased

Mediastinoscopy - an examination of the mediastinum, which is a portion of the thoracic cavity in the middle of the thorax, between the pleural sacs containing the two lungs; it extends from the sternum to the vertebral column and contains all of the thoracic viscera, except the lungs

Medulla - the most vital part of the brain, continuing as the bulbous portion of the spinal cord just above the foramen magnum and separated by the pons by a horizontal groove; one of three parts of the brain stem and contains mostly white substance with some mixture of gray substance; contains the cardiac, the vasomotor and the respiratory centers of the brain; medullary injury or disease often proves fatal

Meningitis - an infection or inflammation of the membranes covering the brain and spinal cord, usually purulent and involves the fluid in the subarachnoid space in the brain; characterized by severe headache, vomiting and pain and stiffness in the neck; causes usually are a strep pneumonia; nursing care includes watching for signs of increased intracranial pressure (ICP), prevent aspiration in the case of convulsive seizures, and to avoid airway obstruction; first day or two - need isolation, after that just IV fluids and feedings (NG) if necessary for a prolonged period of time

MI - myocardial infarction - heart attack - where damage to the heart muscle has occurred

Micro-villa - one of the many tiny projections, barely visible to the naked eye, clustered over the entire surface of the small intestine, they diffuse and transport fluids and nutrients; each has a core of delicate aerolar and reticular connective tissue supporting the epithelium, various capillaries and usually a single lymphatic lacteal, which fills with milky white secretions during the digestion of a fatty meal

Modality - a method of application or the unemployment of any therapeutic agent; limited to usually physical agents; any state that modifies the action of a drug

MRSA - methicillin resistant staphylococcal aureus - an infection which is resistant to most kinds of penicillin, therefore need to be treated with more potent antibiotics to rid the organism; difficult to treat; takes along time of antibiotics, not 7-10 days worth to improve the condition; seen when patients are debilitated, on mechanical ventilation or chronic respiratory problems; have thick tenacious secretions, sometimes mucous plugs etc.

Mucous membrane - any one of four major kinds of thin sheets of tissue that cover or line various parts of the body; lining cavities or canals of the body that open to the outside, as the linings of the mouth; they consist of a surface layer of connective tissue and protect the underlying structure, secrete mucous, and absorb water, salts, and other solutes

Mucus - the slippery, viscous secretions of the mucous membranes and glands, containing mucin, white blood cells, water, inorganic salts, and exfoliated cells

Mucus plug - a collection of thick secretions usually seen in the respiratory system, that the patient is unable to expectorate (cough up) and needs to be suctioned; it can occlude the trachea and can cause death if the "wind pipe" is totally obstructed

Musculoskeletal - of or pertaining of the muscles and the skeleton

N

Nares - the pairs of anterior and posterior openings in the nose that allow the passage of air from the nose to the pharynx and the lungs during respiration

Nasal flaring - where you see a sudden intensification of a disease in the respiratory system

Nasal septum - the partition separating or dividing the nostrils; composed of bone and cartilage covered by mucous membrane

Nasopharynx - one of the three regions of the throat, situated behind the nose and extending from the posterior nares to the level of the soft palate, on the posterior nasopharynx, opposite the posterior nares, are the pharyngeal tonsils

Neoplasm - an abnormal growth of new tissue, benign or malignant

Nervous system - the extensive, intricate network of structures that activates, coordinates, and controls all of the functions of the body; divided in to the central nervous system composed of the brain and the spinal cord, and the peripheral nervous system which includes the cranial and spinal nerves; these subdivision include the somatic and visceral parts of the body with the afferent (carry sensory impulses to the CNS) and efferent (carries motor impulses from the CNS to the muscles and other organs) nerve fibers, somatic fibers are associated with bone, muscle and skin, the visceral fibers associated with the internal organs, blood vessels, and mucous membranes

Neuro-degenerative - deterioration or impairment of the neuro system

Neuromuscular - pertaining to the nerves and the muscles

NG - nasogastric - pertaining usually to a tube that is inserted in the nose, down into the stomach for purposes to keep the stomach empty if vomiting is a problem; give the stomach a rest if stomach or bowel surgery; feedings or administration of medications, to give extra fluids, and also an outlet for a lot of gas

Nocturnal - pertaining to or occurring at night, someone who is active at night and sleeps in the day

Nosocomial - also called hospital acquired infection - an infection acquired during hospitalization often candida, ecoli, hepatitis, herpes zoster (chicken pox virus), pseudomonas or staphylococcus

Nostrils - anterior nares - the ends of the nostrils that open anterior into the nasal cavity and allow inhalation and exhalation of air

O

O₂ - symbol for oxygen

Obesity - an abnormal increase in production of fat cells in the subcutaneous tissues of the body

Olfactory - the part of the brain responsible for subjective appreciation of odors; a complex group of neurons located near the junction of the temporal and parietal lobes

Oropharynx - one of the three anatomical divisions of the pharynx, it extends behind the mouth from the soft palate above to the level of the hyoid bone below and contains the palatine, tonsils and the lingual tonsils

Orthopnea - an abnormal condition in which a person must sit or stand in order to breathe deeply or comfortably; occurring in many cardiac and respiratory disorders, as asthma, pulmonary edema, emphysema, pneumonia, and angina pectoris (chest pain)

Oscillation - a swinging pendulum like movement; a vibration

Otitis Media - inflammation or infection of the middle ear; a common affliction of children; mostly caused by influenza or strep pneumonia; usually preceded by an upper respiratory infection; gain access to the ear through the eustachian tube; signs include fullness of the ear, diminished hearing, pain or fever; treat symptomatically

Oximeter - apparatus for continuously determining the amount of oxygen in the blood, usually done by measuring the amount of light transmitted through the skin

Oxygenation - the process of combining or treating with oxygen

P

PO₂ - symbol for partial pressure oxygen of the blood

Palate - a structure that forms the roof of the mouth; it is divided into the hard and soft palates

Palpitations - a pounding or racing of the heart associated with normal emotional responses or with certain heart disorders; some people may complain of pounding hearts and display no evidence of cardiac disease, while others with serious cardiac disease may not detect associated abnormal palpitations; some patients complain of palpitations after receiving digitalis medication because it increases the force of heart contractions; many healthy individuals become concerned and describe as palpitations the normal sounds of their hearts against their pillows when they lie down to sleep

Paranasal sinus - the air cavities in various bones around the nose, as the frontal sinus in the frontal bone lying deep to the medial part of the superciliary ridge and the maxillary sinus within the maxilla between the orbit, the nasal cavity and the upper teeth

Parenchyma - the tissue of an organ as distinguished from supporting or connective tissue

Parietal - pertaining to the outer wall of a cavity, the parietal bone of the skull such as the parietal lobe of the brain

Paroxysmal - a marked usually episodic increase in symptoms, like heart rate, pain, cough, etc.

Pathogen - any microorganism capable of producing disease

Pathophysiology - the study of the biological and physical manifestations of the disease as they correlate with the underlying abnormalities and physiologic disturbances; it does not

deal directly with the treatments of the diseases rather it explains the processes within the body that results in the signs and symptoms of the disease

PCO₂ - symbol for partial pressure of carbon dioxide of the blood

PEEP - positive end expiratory pressure - in respiratory therapy - ventilation controlled by a flow of air delivered in cycles of constant pressure through the respiratory cycle; the patient is intubated and a respirator cycles the air through an ET tube (endotracheal tube) or tracheostomy to keep the lungs from totally deflating; PEEP is used for the relief of respiratory distress secondary to shock, pulmonary edema, trauma, surgery, or other conditions in which arterial levels of oxygen are deficient; during PEEP therapy close observations and monitoring of ABG's and vital signs are necessary

Percussion - a technique in physical examination used to evaluate the size, borders and consistency of some of the internal organs and to discover the presence and evaluate the amount of fluid in a cavity of the body; direct or immediate percussion refers to percussion performed by striking the fingers directly on the body surface; finger indirect or mediate percussion involves striking a finger of one hand on a finger of the other hand as it is placed over the organ

Perfusion - passing of a fluid through spaces; the pouring of a fluid; supplying an organ or tissue with a fluid by injection into an artery; the treatment of putting fluids, antibiotics, dye into the body as a lung perfusion test to examine the lungs to check for blood clots

Pertussis - an acute infectious disease characterized by inflammation of a mucous membrane followed by a paroxysmal cough, ending in a whooping inspiration; begins as a common cold - slight temperature, sneezing, rhinitis and dry cough; after 7-10 days the paroxysmal stage begins with more violent cough, consists of a series of several short coughs followed by long drawn inspiration during which time the whoop is heard - patient gets a worried expression, face appears cyanotic, veins distended, vomiting is now common due to the violent cough; may see epistaxis, sub conjunctival hemorrhages, or hemorrhages in other parts of the body; the number of paroxysm may vary from 3-4 or 40-50; after several weeks symptoms begin to diminish slowly, the disease may be prevented by immunizing infants beginning at 3 months of age

PFT - pulmonary function tests - a series of tests for determining the capacity to exchange oxygen and carbon dioxide efficiently; two major types of tests: one to measure ventilation or the ability of the bellows action of the chest and lungs to move gas in and out of the alveoli, the other to measure the diffusion of gas across the alveolar capillary membrane and the perfusion of the lungs by blood

pH - a scale representing the relative acidity or alkalinity of a solution; the numerical pH value indicates the relative concentrations of hydrogen ions in the solution compared to that of a standard solution

Pharyngitis - inflammation or infection of the pharynx, usually causing symptoms of a sore throat; causes may be diphtheria, herpes simplex virus, infectious mononucleosis and streptococcal infection; specific treatment depends on the cause; symptoms may be relieved by analgesic medications, drinking warm or cold liquids, or saline irrigations of the throat

Pharynx - a tubular structure that extends from the base of the skull to the esophagus and is situated just in front of the cervical vertebrae; serves as a passageway for the respiratory and digestive tracts and changes shape to allow the formation of various vowel sounds; composed of muscle, lined with mucous membranes and is divided into the nasopharynx, oropharynx, and the laryngopharynx; contains the openings of the right and left auditory tubes; those of the two posterior nares, fauces, and those into the larynx and the esophagus; contains the pharyngeal tonsils, the palatine tonsils, and the lingual tonsils

Phrenic nerve - one of a pair of muscular branches of the cervical plexus, arising from the fourth cervical nerve; it contains about half as many sensory as motor fibers and is generally known as the motor nerve to the diaphragm; although the lower thoracic nerves also help to innervate the diaphragm; it lies on the ventral surface of the scalenus anterior, crossing from its lateral to its medial border, continuous down between the subclavian artery and subclavian vein, enters the thorax, along the anterior aspect of the pericardium and reaches the diaphragm, where it divides into terminal branches; the right phrenic nerve is deeper and shorter than the left; the pleural branches of the phrenic nerve are very fine filaments supplying the mediastinal pleura; the pericardial branches are delicate filaments passing to the upper pericardium; the terminal branches diverge after passing separately through the diaphragm and are distributed on the abdominal surface of the diaphragm

Pinna - the external ear

Plasma - the watery colorless, fluid portion of the lymph and the blood in which the leukocytes, erythrocytes, and platelets are suspended; it contains no cells and is made up of water, electrolytes, proteins, glucose, fats, bilirubin, and gases; is essential for carrying the cellular elements of the blood through the circulation, transporting nutrients, maintaining the acid-base balance of the body, and transporting wastes from the tissues, plasma and interstitial fluid correspond closely in content and protein concentration, therefore plasma is important in maintaining the osmotic pressure and exchange of fluids and electrolytes between capillaries and tissues

Pleurae - a delicate serous membrane enclosing the lung, composed of a single layer of mesothelial cells resting on a delicate membrane of connective tissue; beneath the membrane is a stroma of collagenous tissue containing yellow elastic fibers; the pleura divides into the visceral pleura, which covers the lung, dipping into the fissures between the lobes and the parietal pleura, which lines the chest wall; covers the diaphragm and reflects over the structures in the mediastinum; the parietal and visceral pleura separated from each other by a small amount of fluid that acts as a lubricant as the lungs expand and contract during respiration

Pleural effusion - an abnormal accumulation of fluid in the interstitial and air spaces of the lungs, characterized by fever, chest pain, dyspnea, and non-productive cough; the fluid involved in an exudate or transudate from inflamed pleural surfaces; a transudate that accumulates in pulmonary edema is commonly aspirated; an exudate may result from pulmonary infarction, trauma, tumor, or infection as tuberculosis; the specific cause of the

exudate is treated and the exudate may be aspirated or surgically drained; may also give corticosteroids, diuretics, vasodilators and oxygen therapy

Pleural space - the potential space between the visceral and parietal layers of the pleurae; it contains a small amount of fluid that acts as a lubricant, allowing the pleurae to slide smoothly over each other as the lungs expand and contract with respiration

Pleurisy - inflammation of the parietal pleura of the lungs, characterized by dyspnea and stabbing pain, leading to restriction or ordinary breathing with spasm of the chest on the affected side; a friction rub may be heard on auscultation; simple pleurisy with undetectable exudate is called fibrinous or dry; pleural effusion indicates extensive inflammation with considerable amounts of exudate in the pleural spaces; causes may include bronchial cancer, lung or chest wall abscess, pneumonia, pulmonary infarction, and tuberculosis; pleurisy may result in permanent adhesions between the pleura and adjacent surfaces; treatment consists of relief of pain and therapy for the primary disease

Pneumonia - an acute inflammation of the lungs usually owing to inhaled pneumococci, causing the alveoli and the bronchioles of the lungs to become plugged with a fibrous exudate; may be caused by other bacteria as well; signs - severe chills, high fever, cough, and chest pain, inflammation of the right lower lobe may produce a pain suggesting appendicitis; as the disease progresses the sputum may become thicker and more purulent and the patient may experience painful coughing spells, breathing more difficult, painful shallow and rapid, pulse increases (120 or more per minute), profuse sweating and cyanosis; in children, may see convulsions; as the alveoli become filled with exudate, the affected area of the lung become increasingly firm, consolidated and stiff; x-rays, blood cultures and sputum cultures are done to identify the specific organism; treatment includes bed rest, fluids, antibiotics, analgesics, antipyretics and if necessary, oxygen, expectorants, postural drainage and suctioning as needed is usually necessary

Pneumothorax - a collection of air in the pleural space causing the lung to collapse; may be the result of an open chest wound that permits the entrance of air into the pleural space or of the rupture of a vessel on the surface of the lung; possibly without apparent cause; a tension pneumothorax results when air enters the pleural space with each inspiration and becomes trapped causing pressure buildup, this increased pressure may cause a mediastinal shift; onset of a pneumothorax is sudden sharp chest pain, followed by difficult rapid breathing, cessation of normal chest movements on the affected side, tachycardia, weak pulse, hypotension, diaphoresis, elevated temperature, pallor, dizziness, and anxiety; patient is urged to remain calm, head of the bed is elevated (semi-fowler's position), O₂ administration; the air in the affected area is aspirated by placing a chest tube and attached to a water seal unit and left there until all of the air is gone and the lung is again fully expanded, patient is taught how to cough, turn, and deep breathe

Pneumovax - an immunization against pneumonia indicated for the elderly or those in debilitated states

Polycythemia - an abnormal increase in the number of erythrocytes (red blood cells) in the blood; it may be secondary to pulmonary or heart disease or to prolonged exposure to high

altitudes; in the absence of a demonstrable cause, it is considered idiopathic (without a known cause)

Polyp - a tumor with a stem, commonly found in vascular organs such as the nose, uterus, or rectum, they bleed easily and need to be removed surgically

Pons - any slip of tissue connecting two parts of a structure or an organ of the body, a prominence on the vertebral surface of the brain stem, between the medulla and the cerebral peduncles of the mid brain, the pons consists of white matter and a few nuclei and is divided into a ventral portion (consists of transverse fibers separated by longitudinal bundles and small nuclei) and a dorsal portion (comprises the tegmentum which is a continuation of the reticular formation of the medulla)

Posterior - pertaining to or situate in the back part of a structure, as of the dorsal surface of the human body, the back part of something

Postural drainage - the use of positioning to drain secretions from specific segments of the bronchi and the lungs into the trachea; coughing normally expels secretions from the trachea; positions are selected that promote drainage from the affected parts of the lungs; pillows and raised sections of the hospital bed support or elevated parts of the body; effectiveness of the procedure depends on positioning that allows drainage by gravity, ciliary action and effective breathing

Pulmonary - pertaining to the lungs

Pulmonary embolus - (PE) - the blockage of a pulmonary artery by foreign matter, as fat, air, tumor tissue, or a thrombus that usually arises from the pulmonary vein; causes an alteration of blood constituents with increased coagulation, damage to blood vessel walls, and stagnation or immobilization, especially when associated with prolonged bed rest, childbirth, CHF, or surgery; it is difficult to distinguish from pneumonia or heart attack; symptoms include dyspnea, sudden chest pain, shock and cyanosis, pulmonary infarction usually occurs 6-24 hours after a pulmonary embolus, usually also pleural effusion, hemoptysis, leukocytosis, fever, tachycardia, atrial arrhythmias, and a striking distention in the neck veins are seen

Pulmonary ligament - a white, shiny, flexible band of fibrous tissue in the lung, slightly elastic and composed of parallel collagenous bundles

Q

R

Rales - a common respiratory sound heard on auscultation of the chest during inspiration, characterized by continuous bubbling noises; fine rales have a crackling sound produced by air entering distal bronchioles or alveoli that contain serous secretions, as in congestive heart failure; medium rales are medium pitched bubbling or gurgling sounds caused by air passing through secretions in the bronchioles or by separation of bronchiolar walls previously adhered by exudate; coarse rales originate in the larger bronchi or trachea and have a lower pitch

Receptors - molecular group of cells which have a special affinity for toxins, ambo-receptors, etc.; group of cells functioning in reception of stimuli; a sense organ endings of afferent (sensory) nerves; many kinds of receptors, - examples, chemo-receptors, photoreceptors, presso-receptors, tango-receptors

Resonance - an echo or other sound produced by percussion of an organ or cavity of the body during a physical examination

Respiratory acidosis - a condition characterized by increased arterial PCO_2 , excess carbonic acid, and increased plasma hydrogen-ion concentration, caused by reduced alveolar ventilation; the hypoventilation associated with this condition inhibits the excretion of carbon dioxide which consequently combines with water in the body to produce excessive carbonic acid and thus reduce blood pH; signs include lethargy, headache, shallow and irregular respirations, fine tremors, tachycardia hypertension and vasodilatation

Respiratory alkalosis - a condition characterized by decreased PCO_2 , decreased hydrogen-ion concentration and increased blood pH; caused by pulmonary and non pulmonary problems; the hyperventilation associated with respiratory alkalosis most commonly stems from extreme anxiety; may also be induced by excessive mechanical ventilation, deep and rapid breathing at rates of 40 or higher per minute is a major sign, as well as lightheadedness, dizziness, peripheral paresthesia, spasms of the hands and feet, muscle weakness, tetany, and cardiac arrhythmia, treat by breathing into a brown bag or sedatives to reduce respiratory rate

Retractions - the visible sinking in of the soft tissues of the chest between and around the firmer tissue of the cartilaginous and bony ribs as occurs with increased inspiratory effort; retraction begins in the inter costal spaces, if increased effort is needed to fill the lungs, the supra clavicular and infraclavicular retraction may be seen; in infants, sternal retractions occurs with only slight increase in respiratory effort, owing to the pliability of their chests

Rhonchi - abnormal sounds heard on auscultation of a respiratory airway obstructed by thick secretions, muscular spasms, neoplasm, or external pressure; the continuous rumbling sounds are more pronounced during expiration and they characteristically clear on coughing; sibilant rhonchi are high pitched and are heard in the small bronchi as in asthma where sonorous rhonchi are lower pitched and are heard in the large bronchi as in tracheo bronchitis

S

Saturation - unable to absorb, dissolve or accept any more of a given substance

Scoliosis - lateral curvature of the spine, a common abnormality in children; causes include congenital malformations of the spine, poliomyelitis, skeletal dysplasias, spastic paralysis and unequal leg length; early recognition and orthopedic treatment may prevent progression including braces, casts, exercises and corrective surgery

Segmental - a section or portion of the body, organ; object occurring naturally or having its borders artificially established

Sepsis - infection, contamination, usually seen throughout the entire body not just a localized area

SIDS - sudden infant death syndrome-also called crib death - the sudden and unexpected death of an apparently normal and healthy infant that usually occurs during sleep and with no physical or autopsic evidence of disease; usually occurs between 2 weeks and 1 year of age; may be caused by mechanical suffocation, a deficit in respiratory mucosal defense, prolonged apnea, an unknown virus, anatomical abnormality of the larynx and immunoglobulin abnormalities; occurs more often in the winter months in infants 10-14 weeks old, especially those born prematurely, and in males more than females

Sinus - a canal or passage leading to an abscess, a cavity within a bone, dilated channel for venous blood; any cavity with a relatively narrow opening, lined with epithelium, filled with air and communicate with nasal cavities through their various openings, function of the sinuses are warming, moistening, filtering of the air

Sinusitis - inflammation or infection of one or more of the paranasal sinuses, it may be a complication of an upper respiratory infection, dental infection, allergy, a change in atmosphere, or a structural defect of the nose; with swelling of the nasal mucous membranes the openings from sinuses to the nose may be obstructed resulting in an accumulation of sinus secretions, causing pressure, pain, headache, fever, and local tenderness; treatment includes steam inhalations, nasal decongestants, analgesics, and if infection is present, antibiotics

Somnolent - the condition of being sleepy or drowsy, tending to cause sleepiness

Sphenoidal - pertaining to the sphenoid bone, which is at the base of the skull anterior to the temporal bones and the basilar part of the occipital bone, it resembles a bat with its wings extended

Splinting - supporting the ribs or abdomen after a severe coughing attack or abdominal surgery with a pillow or just your arms or hands

Sputum - material coughed up from the lungs and expectorated from the mouth, it contains mucous, cellular debris, microorganisms and possibly blood or pus; the amount, color and constituents of the mucous are important in the diagnosis of many illnesses, including tuberculosis, pneumonia and lung cancer

Sternal retractions - the visible sinking in the soft tissues of the chest between and around the firmer tissue of the cartilaginous and bony ribs, as occurs with increased inspiratory effort; retraction begins in the inter costal spaces, if increased effort is needed to fill the lungs; super ventricular and infraclavicular retraction may be seen; in infants sternal retractions occurs with only slight increase in respiratory effort, owing to the pliability of their chests

Sternum - the elongated flattened bone forming the middle portion of the thorax; it supports the clavicles, articulates with the first seven pairs of ribs and comprises the manubrium (one of the small bones at the end of the sternum comprising the xiphoid

process); it is composed of highly vascular tissue covered by a thin layer of bone, longer in men than women

Stridor - an abnormal, high-pitched musical respiratory sound; caused by an obstruction of the trachea or larynx; it is usually heard during inspiration, may indicate several neoplastic or inflammatory conditions

Super infection - an infection occurring during antimicrobial treatment for another infection, usually a result of change in the normal tissue flora favoring replication of some organisms by diminishing the vitality and then the number of competing organisms

Surfactant - an agent, as soap or detergent dissolved in water to reduce its surface tension or the tension at the interface between the water and another liquid; certain lipoproteins that reduce the surface tension of pulmonary fluids, allowing gas exchange in the alveoli and contributing to the elasticity of pulmonary tissue

Susceptible - the condition of being more than normally vulnerable to a disease or disorder

Sympathetic - also called the autonomic nervous system - the part of the nervous system that regulates involuntary vital function, including the activity of the cardiac muscle, smooth muscle and the glands; it has two divisions, the sympathetic nervous system accelerates heart rate, constricts blood vessel and raises blood pressure, the parasympathetic nervous system slows the heart rate, increases intestinal peristalsis and gland activity and relaxes sphincters

T

Tachycardia - a circulatory condition in which the myocardium contracts regularly but at an accelerated rate of 100-150 beats per minute

Tachypnea - an abnormally rapid rate of breathing

TB - tuberculosis - a chronic granulomatous infection caused by an acid-fast bacillus generally transmitted by inhalation or ingestion of infected droplets and usually affecting the lungs; listlessness, vague chest pain, pleurisy, anorexia, fever and weight loss are early symptoms; night sweats, pulmonary hemorrhage, expectoration of purulent sputum and dyspnea develop as the disease progresses; the lung tissues react by producing protective cells that engulf the disease organism forming tubercles; the patient needs to be isolated to prevent the spread of the disease, bed rest and good nutrition and to adhere to the medication regime

Tenacious - holding onto; adhering to; adhesive, as in tenacious secretions (thick-unable to cough up) due to the adherence in the trachea, bronchus or respiratory tree

Terminal - of a structure or process near or approaching its end, as a terminal bronchiole or a terminal disease

Tertiary - belonging to the third level of sophistication of development; third in frequency or in order of use

Tetanus - an acute potentially fatal infection of the central nervous system caused by an exotoxin; the bacillus is a common resident of the superficial layers of the soil and a

normal inhabitant of the intestinal tracts of cows and horses; may enter the body through a puncture wound, laceration or burn, via the uterus into the blood stream; it occurs in two forms - one with an abrupt onset, high mortality and a short incubation period (3-21 days), the other with less severe symptoms, a lower mortality and a longer incubation (4-5 weeks); wounds of the face, head and neck are most likely to result in fatal infection; as the bacillus may travel rapidly to the brain; disease characterized by irritability, headache, fever, and painful spasms of the muscles resulting in lockjaw, eventually every muscle in the body is in toxic spasm; the motor nerves transmit the impulses from the infected CNS to the muscles; no lesions; prompt and thorough cleansing and debridement of the wound are essential to avoid the tetanus infection, immunizations of tetanus toxoid are given to prevent the condition

Tetany - a condition characterized by cramps, convulsions, twitching of the muscles and sharp flexion of the wrist and ankle joints, sometimes may be accompanied by stridor; caused by an abnormality of calcium metabolism which can occur in association with Vitamin D deficiency, hypoparathyroidism, alkalosis, or the ingestion of alkaloid salts

Thoracentesis - a surgical procedure that makes a perforation into the chest wall and the pleural space with a needle to aspirate fluid for diagnostic or therapeutic purposes or for the removal of a specimen for biopsy; performed under local anesthesia with the patient in the upright position; may be used in the treatment of pleural effusion

Thoracic - pertaining to the chest, the cavity, which includes the lungs and the heart

Thyroid - a highly vascular organ at the front of the neck, consists of bilateral lobes connected in the middle by a narrow isthmus; slightly heavier in women than men and enlarges during pregnancy; secretes the hormone thyroxin directly into the blood and is part of the endocrine system of ductless glands; essential to normal body growth in infancy and childhood; its removal greatly reduces the oxidative processes of the body, producing a lower metabolic rate characteristic of hypothyroidism; activated by the pituitary hormone and requires iodine to elaborate thyroxin

Tonsillitis - inflammation or infection of the of a tonsil; acute tonsillitis is usually caused by a strep infection characterized by severe sore throat, fever, headache, malaise, difficulty in swallowing, earache, and enlarged tender lymph nodes in the neck; may accompany scarlet fever

Trachea - a nearly cylindrical tube in the neck, composed of cartilage and membrane, that extends from the larynx at the level of the sixth cervical vertebrae to the fifth thoracic vertebrae, where it divides into the bronchi; it conveys air to the lungs; the ventral surface of the tube is covered in the neck by the thyroid gland, isthmus and various other structures; dorsally, the trachea is in contact with the esophagus

Tracheostomy - an opening through the neck into the trachea with an indwelling tube inserted; the incision and stoma are created surgically to establish an airway when the pharynx is obstructed by a foreign body, tumor, or edema of the glottis; local or general anesthesia is used

U

Universal precautions - used in health care where gloves, masks and gowns are worn to protect the patient and the care giver from infection such as respiratory secretions or AIDS

V

Vaccinations - any injection of attenuated or killed microorganisms, as bacteria; to induce immunity or to reduce the effects of associated infectious diseases; historically, the first vaccinations were administered to immunize against smallpox; vaccinations are now available to immunize against many diseases

Vagus nerve - either of the longest pair of cranial nerves essential for speech, swallowing and the sensibilities and functions of many parts of the body

Vaporizer - device for converting liquids into a vapor spray, used with asthmatics, croup or other respiratory conditions

Vascular - composed of or pertaining to blood vessels, the vascular system includes the heart, blood vessels, lymphatics; and their parts considered collectively, includes the pulmonary and portal systems

Vasoconstriction - a narrowing of the lumen of any blood vessel, especially the arterioles and the veins in the blood reservoirs of the skin and the abdominal viscera; it depends on the stimulation of the vasomotor constriction center in the medulla-where impulses travel from along the sympathetic nerve fibers and contract the smooth muscle layers of the arteries, the arterioles, the venules and the veins, causing the constriction of these vessels; also induced by vasomotor pressure reflexes, chemical reflexes, the medullary ischemic reflex and vasomotor impulses from the cerebral cortex and the hypothalamus

Vasodilatation - widening or distention of blood vessels, particularly arterioles usually owing to nerve impulses or to certain drugs that cause relaxation of smooth muscle in the walls of the blood vessels

Venous - pertaining to a vein which carries blood back to the lungs to be re-oxygenated and pushed back into the body through arteries

Ventilation - the process by which gases are moved into and out of the lungs

Ventilator - any of several devices used in respiratory therapy to provide assisted respirations and positive pressure breathing; can assist the patient short term or long term; can also be used in home care with portable units, keeps the oxygen levels adequate when ABG's are monitored

Ventricle - a small cavity, as one of the cavities filled with cerebrospinal fluid in the brain or the right and left ventricles of the heart, which push the blood out into the body with every beat; the left ventricle is many times damaged during a heart attack

Venturi Mask - one of several masks, which cover the nose and mouth for respiratory therapy, adequate oxygen, especially for mouth breathers

Venules- any one of the small blood vessels that gather blood from the capillary plexuses and anastomose to form the veins

Vertebrae - any one of the 33 bones of the spinal column, 7 cervical, 12 thoracic, 5 lumbar, 5 sacral, and 4 coccygeal vertebrae, they are alike except the 1st and 2nd cervical vertebrae which if traumatized can and usually will be fatal because they are at the level of the most vital functions of the body

Vesicular - vesicle - a small sac or bladder containing fluid, a small blister like elevation on the skin from the size of a pinhead to that of a split pea containing serous fluid; vesicles may be round, transparent; opaque or dark elevations of the skin, sometimes containing seropurulent or bloody fluid

Vestibular - pertaining to the vestibule, as the vestibular portion of the mouth, which lies between the cheeks and the teeth

Viral - pertaining to or caused by a virus (200 pathogenic to mankind)

Virulent - pertaining to a highly pathogenic or rapidly progressive condition

Visceral - pertaining to the viscous, any one of the large internal organs contained in the abdominal, the thoracic, or the pelvic cavities of the body

Vocal cords - either of two strong bands of yellow elastic tissue in the larynx enclosed by membranes called vocal folds and attached ventrally to the angle of the thyroid cartilage and dorsally to the arytenoid

W

Wheezing- a form of rhonchi characterized by a high-pitched musical quality, it is caused by a high velocity flow of air through a narrowed airway and is heard both in inspiration and expiration; associated with asthma and chronic bronchitis; unilateral rhonchi are characteristic in bronchogenic cancer, foreign bodies, and inflammatory stenosing lesions; an asthmatic wheeze is caused by an obstruction in the trachea or bronchus

X

Xiphoid process - the smallest of three parts of the sternum, articulating with the body of the sternum and laterally with the seventh rib; several muscles of the abdominal wall are attached to the xiphoid process including the rectis abdominis and the linia alba

Y

Z

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