Respiratory Care Services
Basic Care of the CF Inpatient
Overview:

• The Respiratory Therapist should explain all the equipment used to the patient in the hospital, for discharge and why it is important they take every treatment.
  
  • Pulmonary Hygiene is of the upmost importance the same as going to the dentist to get your teeth cleaned. You don’t want to wait until you need major work!!

• It is the RT’s responsibility to contact the physician to clarify orders.

• PFT’s should be schedule at the end of the day or be done in the patient’s room. All medications are to be given prior to PFT's.
WHAT IS CYSTIC FIBROSIS?

- **Cystic fibrosis** (usually called CF) is an *inherited* disease. It causes certain glands in the body to not work properly. These glands are called the exocrine (outward-secreting) glands.

- *Exocrine glands* normally make thin, slippery *secretions* including sweat, *mucus*, tears, saliva and digestive juices. These secretions move through *ducts* (small tubes) to the surface of the body or into hollow organs, such as *intestines* or *airways*. Exocrine glands and their secretions help the body function normally.

- In CF, exocrine glands (except sweat glands) make mucus that is too thick and sticky. This mucus plugs ducts and other passageways. *Mucous plugs* are most often in the lungs and intestines and can cause problems with breathing and digestion.

- CF also affects sweat glands. The levels of salt (sodium and chloride) and potassium in the sweat are too high. This may cause problems during times of increased sweating.

- CF does not affect *endocrine glands*. Endocrine glands and exocrine glands are different. Endocrine (inward-secreting) glands make *hormones* that pass into the blood.
Cystic fibrosis is an inherited chronic disease that affects the lungs and digestive system of about 30,000 children and adults in the United States (70,000 worldwide). A defective gene and its protein product cause the body to produce unusually thick, sticky mucus that:
• clogs the lungs and leads to life-threatening lung infections; and
• obstructs the pancreas and stops natural enzymes from helping the body break down and absorb food.

In the 1950s, few children with cystic fibrosis lived to attend elementary school. Today, advances in research and medical treatments have further enhanced and extended life for children and adults with CF. Many people with the disease can now expect to live into their 30s, 40s and beyond.

Symptoms of Cystic Fibrosis

People with CF can have a variety of symptoms, including:
• very salty-tasting skin;
• persistent coughing, at times with phlegm; frequent lung infections
• wheezing or shortness of breath;
• poor growth/weight gain in spite of a good appetite; and frequent greasy, bulky stools or difficulty in bowel movements.

Statistics
• About 1,000 new cases of cystic fibrosis are diagnosed each year.
• The predicted median age of survival for a person with CF is in the early 40s.
In CF, there are not enough working gates. Chloride can’t move and the airway surface is dehydrated, compromising patients’ ability to clear bacteria.
Why is Respiratory so Important?

• Cystic Fibrosis is a complex multiorgan disease.

• Lung disease accounts for nearly _____ of the mortality?

• 85%
2014 National Registry Data

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<tr>
<th>Primary Cause of Death in 2014</th>
<th>Number of Individuals</th>
<th>Percent</th>
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<tr>
<td>Respiratory/cardiorespiratory</td>
<td>325</td>
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<tr>
<td>Transplant-related: other</td>
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<tr>
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<td>Liver disease/liver failure</td>
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<td>2.8</td>
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<tr>
<td>Suicide</td>
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<tr>
<td>Trauma</td>
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85%
WILL RT REMAIN IMPORTANT?

Yes 80-85% of mortality is still Respiratory Related in the latest 2016 released Registry Data.
RESPIRATORY

- Although significant gains have been made in improving lung function and survival in cystic fibrosis (CF), ultimately respiratory failure is the leading cause of mortality in these patients.

  - Woo MS. Overview of lung transplantation 2008
SURVIVAL TRENDS

Median Predicted Survival Age, 1986–2016 In Five Year Increments

Year

24.9 - 90.91 98.91 99.91 92.93 94.95 96.97 99.99 0.01 0.02 0.03 0.04 0.05 0.06 0.07 0.08 0.09 0.10 0.11 0.12 0.13 0.14 0.15 0.16

Median Predicted Survival Age (Years)

24 28 32 36 40 44 48
PROGRESS CONTINUES

LUNG FUNCTION

MEDIAN FEV₁ PERCENT PREDICTED IN 1996 AND 2016

For 10 year olds

1996: 86%
2016: 96%

For 18 year olds

1996: 66%
2016: 83%

For 30 year olds

1996: 49%
2016: 61%
PROGRESS CONTINUES

LIFE EXPECTANCY AT BIRTH

31 YEARS
1992-1996

43 YEARS
2012-2016
Airway Clearance Therapies/Techniques
HFCWO the vest remains the most popular form of ACT in the US
Mucociliary Clearance and Obstruction

- Tenacious Mucus
- Periciliary Liquid (PCL)
- Surface Epithelial Cells

Normal Lung vs. CF Lung

CFTR
Common Nebulized Medications

(GIVEN IN THIS ORDER)

- Bronchodilator

- Hypertonic Saline usually 7% 3-5ml
  - HS and BD With a full vest treatment for 20 minutes sitting straight up using a mouthpiece always (when pt. is able) nice slow deep breaths.
  - 10 minutes then HUFF cough for 1-3 minutes (3 to 5 cycles of huffing and resting) then 10 more minutes on the vest.
  - Remember Hypertonic can cause bronchospasm so always pre-medicate with bronchodilator

- Pulmozyme:
  - Used to break down extra cellular DNA left by white blood cells when fighting bacteria trapped in the lungs. Which cause thick and sticky mucus.

- Wait 30 minutes (if possible see another pt.) and then give antibiotic:
  - Tobramycin
  - Cayston
Cleaning of Nebulizers  

- Cleaning Respiratory Equipment

- Germs can get on respiratory equipment and cause lung infections. Clean and disinfect all equipment used for inhaled medicines or airway clearance by using the guidelines by the Cystic Fibrosis Foundation and in the manufacturer’s instructions. It is vital to clean and disinfect equipment such as nebulizers that have been in contact with mucous membranes, sputum or phlegm. To clean and disinfect your nebulizer, follow these steps:
Cleaning of Nebulizers Inpatient

- Hand Held Nebulizers or Small volume nebulizers, should be air-dried (RT may run flowmeter through nebulizer to aid in drying)

  Rinse all nebulizers with sterile water after treatments and air dry between treatments. Nebulizers should not be rinsed with tap water. Recommended to wipe mouthpiece or mask off with alcohol pad

- Rinse the nebulizer with sterile water and shake excess water into the sink

- Place the nebulizer on clean paper towels to dry inside the plastic bin

- Bin should be stored in an area in the room that is away from the sink

- Cover the drying nebulizer parts with a clean paper towel

- Leave the tubing at the flowmeter

- Mesh nebulizers should be changed out per with entire ventilator every 30 days per manufactures instructions.

- Sterile waters should contain the Date, Time, RT Initials on all Sterile waters opened by Respiratory and disposed of after 24 hours.

- CF Nebulizers should be changed out daily
Cleaning of Nebulizers at home

- **IF the patient ask -AT HOME:** The nebulizer parts must be cleaned before they can be disinfected. With a new paper towel, wash the inside and outside of the nebulizer parts with clear liquid dish soap and hot water. Be careful not to damage any of the parts.

- Throw the paper towel away, then rinse the nebulizer parts with water. Clean the nebulizer right after it is used to keep the medicine and debris from drying. Once debris dries, it is difficult to wash off.

- You also can clean the nebulizer parts in an automatic dishwasher if the nebulizer’s manufacturer’s instructions allow.
3. **Disinfect the nebulizer parts.**
   - **DO NOT USE VINEGAR.** Vinegar is not strong enough to kill the germs a person with CF might get.
   - Instead, ask your CF care team what is the best way to disinfect the nebulizer parts. Some options are:
     - Using an electronic steam sterilizer (e.g., used for baby bottles)
     - Boiling (in water) for 5 minutes
     - Microwaving (in water) for 5 minutes
     - Washing in dishwasher, if the water is hotter than 158°F, for 30 minutes
     - Soaking in 70 percent isopropyl alcohol for 5 minutes
     - Soaking in 3 percent hydrogen peroxide for 30 minutes
   - Read the manufacturer’s instructions to learn about cleaning and disinfecting your nebulizer. Do not use a nebulizer that cannot be disinfected.

4. **Rinse the nebulizer parts.**
   - If you disinfect with isopropyl alcohol or hydrogen peroxide, rinse all parts well with sterile water.
   - **DO NOT USE WATER FROM THE FAUCET, BOTTLED WATER OR DISTILLED WATER.** You can make water sterile by boiling it for 5 minutes. Use this water once, then throw it out. If you disinfect by other methods, you do not need to rinse the nebulizer.

5. **Air-dry the nebulizer parts.**
   - After the final rinse, drain the parts on a clean surface covered with new paper towels. Replace wet paper towels with dry ones and fully air-dry all parts. Germs will grow on anything that stays wet. Store the dry nebulizer in a clean, dry bag in a clean, dry place.
   - Some respiratory equipment may need to be cleaned but not disinfected. These items can be cleaned often with liquid soap and hot water. Ask the respiratory therapist, the nurse or physician at your CF care center, how often to clean your equipment and the best way to do so.
Airway Clearance Therapies (ACTs)

- HFCWO – Vest Therapy
- Metaneb
- IPV
- Huff Coughing
- Autogenic Drainage
- PEP Therapy
- Aerobika
- VibraLung
ACT’s

• A number of different ACT’s are provided or taught to all of our patients with acute or chronic pulmonary diseases
  • There are only 2 over-riding physical principles to airway clearance techniques: first, there must be airflow; and, second, for the patient to have air flow, the patient must be able to get air behind the mucus.

• With Cystic Fibrosis:
  • All ACTs should involve coughing and huffing. Many of them use percussion (clapping) or vibration to loosen mucus from airway walls.
  • In an era of personalized medicine it is important to choose the best device possible for your patient. The best ACT’s are the ones that your patient is most likely to perform as part of their daily treatment plan.
Breath holds

• Why do we recommend Breath holding for 3-4 seconds with ACT’s and treatments

• The theory is a pause allows for equalization of ventilation and allows pressure within the lung to equalize.

• It has been demonstrated in multiple clinical trials human lungs can be re-expanded using collateral ventilation channels.
Breath holds allow for collateral ventilation to take effect and get airflow behind collapsed airways or obstructing mucus.
Coughing

• A cough is our natural reflex that protects your lungs. Coughing helps clear your airways of lung irritants, such as smoke and mucus.

• This helps prevent lung infections.
Cough
When patients cough alone the glottis closes and you limit airflow. Coughing alone brings mucus from the 5th to 16th generation of lung.

Huff Coughing and other forms of ACT must be utilized to bring mucus up from airways > generation 16 (The Respiratory ZONE)
Coughing is your body’s own way to remove mucus from the lungs. However, coughing is not always enough to clear the mucus. Reflex coughing can leave you worn-out. Hard coughing can also lead to airway collapse making it harder to clear mucus. The Huff Cough is a gentle way of coughing, which speeds airflow while you keep the throat open. To save energy and oxygen, you must practice and master the method of controlled "Huff Cough".

How Long:
- You should never Huff Cough to the point of exhaustion!
- Perform 2 or 3 huff breaths
- Cough when you feel the mucus collected in your breathing tubes
- Rest for a while (5 to 10 breaths)
- Repeat the huffs until you feel you have cleared mucus or you become tired
- Try to do 3 to 5 cycles of huffing and resting
Huff Coughing Cont…

• To Do Huff Cough:
  • Begin in a sitting position with your chin slightly upward
  • Use your diaphragm (stomach muscle) to breathe in slowly
  • Hold the breath for 2 to 3 seconds
  • Force the breath out your mouth in one quick burst of air
  • Make sure the back of your throat is kept open

• Adjusting the therapy:
  • The length and force of the breath will change where mucus is cleared
  • A normal size breath out clears the larger breathing tubes
  • A longer breath out clears the smaller breathing tubes
  • You should do both to get the best therapy
Vest Therapy

- Next, the practitioner prescribes the Frequency (Hz) and Pressure settings for the treatments:
  - Frequencies between 10 and 14 Hz are commonly used.

- For toddlers and young children, use 10 Hz
- For older children, adolescents, and adults: start at 10 Hz then ramp up to 14 Hz

- Depending on the type of Vest, the common settings for Pressure are:
  - Chest vest = 1 – 4
  - Full vest = 5 – 6
  - Wrap vest = 1 – 4

- As for the Frequencies, younger children/infants should start out at the lower Pressures, but the older children, adolescents, and adults should start at the lower-mid range of pressures and then go quickly to the highest Pressures!

- Patients should receive HFCC vest therapy for 20 minutes at least twice a day. Have them use Vest with aerosol treatment for 10 minutes, pause to huff-cough, then complete the final 10 minutes of treatment.
Breathing Techniques

• Active Cycle of Breathing Technique (ACBT)

- Active cycle of breathing technique (ACBT) involves three phases of breathing techniques. The first phase helps you relax your airways. The second phase helps you to get air behind mucus and clears mucus. The third phase helps force the mucus out of your lungs.
Active Cycle of Breathing Technique

• 1. Breathing control

• Breathing control helps relax the airways. You should breathe in through your nose and out through your mouth with very little effort. Use normal, gentle breathing with the lower chest while relaxing the upper chest and shoulders.

• A good way to do this is to place one hand on your stomach as you breathe. Remember to breathe gently so you relax the airways. If breathing out through the mouth is difficult, you can use the "pursed lips" technique. Repeat breathing control for six breaths before moving to chest expansion exercises.
ACBT cont.

• 2. Chest expansion exercises

• Breathe in deeply. (Some people use a three-second hold of breath to get more air into smaller airways and behind the mucus.) Then breathe out without forcing the air out. Keeping your hands on your ribs will help you reach the level of expansion we are looking for.

• 3. Huffing or huff coughing

• Also called forced expiration technique, huff cough at different, controlled lengths to move mucus up to the larger airways. This huffing should be repeated until all mucus has been huffed out of the body.
• **Autogenic Drainage**

• A breathing exercise using controlled breathing at different lung volumes to move secretions from the small airways to larger airways so that they are easier to expel. Utilizing the expiration for audio feedback for volume and secretions.

• There are three stages:

  1. Unsticking - tidal breathing at low volumes.
  2. Collecting - tidal breathing at mid-lung volumes.
  3. Evacuation - tidal breathing at large lung volumes.
Breathing Techniques cont.

• **Autogenic Drainage (AD)**

  Autogenic drainage (AD) means “self-drainage” and uses different speeds of breathing to move mucus. To do it, your respiratory therapist can teach you how to control your breaths. You will learn how fast and deeply to breathe to fill certain parts of your lungs with air. AD can take some practice and is usually recommended for people age 8 and older.

  • Each phase should take two to three minutes to complete. Completing all three phases is one cycle, which should take about six to nine minutes to complete. Repeat the cycles until you have cleared your lungs as much as possible, which should take between 20 and 45 minutes.
Autogenic Drainage

Phase 1 | 2 | 3
---|---|---
ERV | FRC | $V_t$
RV | Unsticking | Collecting | Evacuating
PEP Therapy

• Positive expiratory pressure (PEP) therapy gets air into the lungs and behind the mucus hopefully utilizing collateral ventilation.

• Oscillating PEP
PEP MASK (Used in Canada and the UK)
Non mask Pep devices
If possible use with an Ambu mask when tolerated by a patient. When done with a regular mouthpiece pressure may escape from the oropharynx and nasopharynx.
Hill Rom vest

- Wrap Vest
- Chest Vest
- Full Vest
Other vest companies
Fitness

• Research shows that regular physical activity or exercise provides multiple benefits for people with cystic fibrosis. These benefits go beyond better lung function keeping fit also helps you strengthen your bones, manage diabetes and heart disease and improve your mood.

• There is not sufficient evidence to conclude that exercise is as effective as other ACT’s.

• It can also help with overall mental health which is extremely important when battling a chronic disease.
AffloVest
AffloVest

• A light truly portable High Frequency Chest Wall Oscillation (HFCWO) vest, now available in the U.S. Approved for Medicare and private health insurance reimbursement,

• AffloVest provides treatment of respiratory diseases like Chronic Obstructive Pulmonary Disease (COPD), Cystic Fibrosis, Chronic Bronchitis, and similar related ailments.
AffloVest

Weight:
Approximately 5-10 lbs depending on size.

Available Sizes:
XXS, XS, S, M, L, XL

Battery Life:
Approximately 2 hours of use depending on intensity.

Treatment Settings:
Nine total setting variations; Three modes of oscillation treatment and three adjustable intensity levels.

Frequency Range:
The AffloVest ranges from 5Hz to 20 Hz. The intensity levels dictate the frequency which generally runs at 5 Hz for the lowest setting, 13 Hz for the medium setting and 20 Hz for the highest setting.
The Monarch™ Airway Clearance System is a high frequency chest wall oscillation (HFCWO) therapeutic device with revolutionary new technology. The therapy combines mobility with targeted kinetic energy and airflow to thin and mobilize secretions from the airways. By allowing patients to move about freely during therapy, it empowers them to take control of their therapy – and their lives.

**Powered by POD technology.**

The Monarch™ System is used to aid mobilization of secretions from the airways to help improve airway health. This is achieved by the placement of eight pulmonary oscillating discs (PODs) containing magnets, on the upper and lower lobes of the lungs. The PODs oscillate and provide a targeted kinetic energy to the lungs. This therapy generates airflow to help thin and mobilize mucus from the small airways to the large airways, where it can be coughed out or suctioned.
Monarch™ Airway Clearance System
SOUND CLEARANCE
The Frequencer™ is a digitally controlled acoustic airway clearance device that uses sound and sound only.
A transducer targets treatment in specific areas of the lungs.
Frequencer

The control unit generates acoustic waves at different frequency levels and treatment intensities.
• **Clinical Application and Intended Use**

• The Vibralung® Acoustical Percussor is intended for use in the hospital or at home for patients with respiratory diseases and related conditions that involve:

  • Increased mucus production
  • infection and inspissation of respiratory secretions,
  • defective mucociliary clearance.
Vibralung

- As a generalization, long and wide airways tend to have a lower RF while short and narrow airways have a higher RF.

- The Vibralung® Acoustical Percussor utilizes frequencies from 5 to 1,200 Hz to achieve resonance in all airways, large and small.
Vibralung
Metaneb

Provides three different modes of therapy.

- Lung Expansion
- Secretion Clearance
- Aerosol Delivery
CPEP Continuous positive expiratory Pressure

CPEP

Flow during both inspiration and expiration helps to prevent or reverse atelectasis
CPEP cont.

Air moves beyond the retained secretions to aid in secretion mobilization and reduces the incidence of air trapping.
RC Cornet

- Silicone hose and mouthpiece
- Easy to use
- Several settings with PEP with pressure Fluctuations

**Asymmetrical wave:**
Unlike symmetrical PEP waves that increase and decrease in exactly the same frequency with no phase difference, the RC-Cornet produces an asymmetrical wave. Viscous mucus cannot follow these fast movements and is shed from the bronchial walls.
Hailing in a setting

Tune OPEP therapy for maximum efficacy. Twisting the mouthpiece from the starting position to position five changes the pressure and flow characteristics.

- Positions 1, 2 and 3 create combined PEP typical in successfully treating patients with COPD and emphysema.
- Positions 4 and 5 create dynamic PEP used in patients with tenacious secretions, bronchiectasis and cystic fibrosis.

The RC Cornet can deliver combined PEP therapy or dynamic PEP therapy based on the clinical indications.

- Combined PEP is characterized as continuous positive pressure above baseline with applied pressure changes. The RC-Cornet promotes collateral ventilation by stabilizing and enlarging the airway.

- Dynamic PEP is characterized by a pressure increase from zero to maximum with a drop back to zero. The RC-Cornet produces a continuous positive pressure with a slow increase from zero to maximum with an abrupt return to zero.
Aerobika

• Easy to use
• Several settings with PEP with pressure Fluctuations
• To USE:
  • Inhale through the device inspiratory pause 3-5 seconds
  • Use Active (but not forceful) exhalation 3 to 4 times longer than inhalation.
  • Describe the effort required as blowing up a balloon but not as forceful as blowing out birthday candles
  • Keep your cheeks tight as you exhaled (no vibrating or “chipmunk cheeks”)
  • Repeat 10 to 15 times and rest Huff Cough before resting
  • Repeat 10 to 20 minutes as tolerated
  • Increase to QID during exacerbations
Aerobika - PEP stents open the airways and the oscillations should assist in moving the mucus.
• Patient’s are responsible if on Cayston in the hospital

• Rinse under water for approx. 10sec

• Wash in warm water and clear liquid soap

• Rinse under warm water
• Air dry on lint free towel or clean paper towel

• You can disinfect the equipment using an electronic baby bottle steam sterilizer

• Do NOT use a MICROWAVE steam sterilizer with the Altera handset!
PARI Nebulizer for Pulmozyme and Tobramycin

Change Daily with all nebulizers
What is the best modality for CF??

- No Single Modality alone is the answer.
- Cystic Fibrosis is a complicated disease that requires a multi-disciplinary approach and teamwork in order fit the needs of the patient and the disease.
- Different techniques and devices may be more or less effective at varying times in different individuals during baseline function vs. pulmonary exacerbations.
- Each person should talk to their clinician to help choose which method of airway clearance is best for them and which they will adhere to, so as to provide the best quality of life and long-term outcomes.
Prevent infection transmission UCLA HOSPITALIZED CYSTIC FIBROSIS (CF) PATIENT GUIDELINES

- Place all patients on contact precaution. Other isolations may be ordered as necessary.
- Place all patients in a private room with private shower/bath
- If two family members who cohabitate at home are admitted concurrently, they may share a semi-private room.
- **Burkholderia cepacia** (*B. cepacia*). This organism is multidrug resistant, highly contagious, and associated with high morbidity and mortality for CF patients. **Pseudomonas aeruginosa** is a commonly found pathogen in these patients. **Staphylococcus aureus** is also a relatively common organism.
- Evaluate activity outside the room based on age, developmental level, and ability to adhere to the following:
  - Hand hygiene immediately before leaving the room
  - Avoid direct contact with other CF patients including in activity room, cafeteria, and other common areas
  - Pediatric patients require MD/designee’s order to leave room; consult Child Life Services for scheduled personal playroom time
  - Perform all respiratory interventions including aerosol therapy, airway clearance and specimen collection inside the patient room
  - Dedicate airway clearance devices to single patient use
  - Encourage patients to bring their own vests from home while in the hospital