Cystic Fibrosis (CF) Survival Guide

Admission:

- Admission labs:
  - Consider obtaining CBC with differential, CMP, PT-INR.
  - Consider obtaining CF sputum culture, particularly if this was not done recently.
- Admission order set: there is a pediatric cystic fibrosis admission order set. Use this with all admitted CF patients
- Contact isolation: all admitted CF patients should be on contact isolation
- Airway clearance:
  - All admitted CF patients:
    - RT consult for airway clearance
    - PT consult for airway clearance
  - Airway clearance frequency:
    - All CF patients admitted for CF pulmonary exacerbation require increased airway clearance (4 times a day).
    - CF patients that are not admitted for pulmonary exacerbation require airway clearance 2 times a day
  - Airway clearance modalities:
    - Manual chest physiotherapy, vest therapy, Metaneb, Vibralung, Aerobika
      - **Manual chest Physiotherapy**: manual percussion
      - **Vest Therapy**: wearable vest
      - **Metaneb**: providing a simultaneous combination of positive pressure, continuous high frequency oscillations and aerosol delivery
      - **Vibralung**: vibratory sound waves during the breathing cycle over a wide range of frequencies to loosen mucus in the air passages making it easier to expectorate (cough up)
      - **Aerobika**: Oscillating Positive Expiratory Pressure (OPEP) device is a drug-free, easy to use, hand-held device
    - Some CF patients change their airway clearance modality during hospitalizations, so ask the patient and family about preferred modality
  - Airway medications: not all CF patients are on all of the following medications
    - Albuterol: usually 2-4 puffs
    - Hypertonic saline (HTS): helps to hydrate secretions
      - It comes in 3%, 5%, 7% concentrations
      - This is typically done 2 times a day
    - Pulmozyme (dornase alfa): helps to liquefy sputum
      - This is typically done 1-2 times a day
    - Inhaled antibiotics: this can include TOBI (inhaled tobramycin), Cayston (inhaled aztreonam), and Colistin
    - Inhaled corticosteroids: examples are Flovent, Symbicort, Advair, etc.
- CFTR-modulators: some CF patients are on CFTR-modulators. These drugs target the underlying CFTR defect.
  - CFTR Modulators: patients qualify based on genotype
    - **Kalydeco** (ivacaftor)
    - **Orkambi** (lumacaftor/ivacaftor)
    - **Symdeko** (tezacaftor/ivacaftor)
    - **Trikafta** (elexacaftor/tezacaftor/ivacaftor)—newest medication

- The dose of these medications may need to be modified if patients are on certain antibiotics and antifungals. This should be discussed with the pediatric pulmonary pharmacists.
- These medications need to be brought from home and relabeled by the pharmacy for hospital use.

- PFTs:
  - PFTs are typically done every week on CF patients admitted for pulmonary exacerbations.
  - PFTs should be ordered by using spirometry order. The procedure is performed at UNC Childrens. The date and time can be adjusted on the order, you do not have to schedule them once order is placed. PFTs should ideally be done after airway clearance has been completed.
- Gastrointestinal and Nutrition:
  - All admitted CF patients:
    - Nutrition consult
  - Diet: high calorie, high protein.
    - Ideal diet for CF patients is high calorie, high protein, high fat, and high salt
    - Nutrition supplementation: Ensure, etc.
      - Order these for patients if they are on them at home.
  - Pancreatic enzyme replacement:
    - Continue the admitted CF patient's home enzyme replacements
    - Types:
      - PO options: **Creon**, **Zenpep**, **Pertzye**
      - **Relizorb** is used for patients with gastrostomy tube feeds
    - CF patients will have different enzyme dosing for meals and snacks.
      - Dosing is calculated as ***mg/kg/dose and ***mg/kg/day.
  - Multivitamins: CF patients who are pancreatic insufficient have difficulty absorbing fat soluble vitamins (A, D, E, and K), so they require CF-specific vitamins.
    - Continue the admitted CF patient's home multivitamin
      - There are multiple brands, including Aquadeks, MVW complete, and Dekas-Plus.
    - All admitted CF patients on antibiotics
      - **Need additional vitamin K (phytonadione)**
        - Typical dosing: vitamin K 5mg twice weekly while on antibiotics
      - Does adjusted for PT/INR
  - Appetite stimulant: some CF patients with poor appetite are on appetite stimulants, including **Periactin, Megace, Marinol**
  - Reflux medications: help activate pancreatic enzymes
- Antibiotics:
  - Review previous sputum cultures, past antibiotic regimens
  - Pseudomonas and Burkholderia
    - ALWAYS double-covered
      - Two antibiotics in DIFFERENT classes
  - Review the CF antibiotic dosing guide
  - Review the antibiotic screening labs (ask Pharmacy)
  - Nephrotoxic Antibiotics: Follow the **NINJA AKI prevention pathway** which will include daily creatinine and monitoring hydration status
    - No NSAIDs while on nephrotoxic medications
  - Vitamin K supplementation needed while on antibiotics
  - Azithromycin is often used for anti-inflammatory effects, rather than antimicrobial effects
- IV access:
  - Some CF patients have ports, while others need PICC line placement

- Review previous admissions to determine if PICC line placement can be done by PICC team versus VIR.

Discharge:

- On the discharge summary, please use the discharge diagnosis "CF bronchopneumonia caused by *** (whichever organisms). This is the way its needs to be documented for billing purposes
- Follow Home IV Antibiotic discharge pathway