

ROBERT WOOD JOHNSON MEDICAL SCHOOL

## **Recommended Medications**

#### **IDEAL** recommended order of inhaled therapy by CF Foundation:

Step One: Albuterol – Opens the airways Step Two: Mucolytics - any in any order

Step Three: Inhaled Antibiotic Step Four: Inhaled steroids

### **AIRWAY OPENERS**

**Nebulizer: The Cocktail- Inhaled medications (Albuterol intal and mucomyst)** 

Albuterol – usual dose is .5 ml, with infants till about age 2 we usually use .25 ml

Albuterol -is a bronchodilator, relaxes the smooth muscle around the airways –helps to open up the airways

Also comes in a premix vile .083% - with normal saline (unit dose)

**Intal** (Cromolyn Sodium) – this medication helps stabilize the mast cells in the airway lining- Mast cells release histamine, the release of histamine can trigger asthma like episode 2 ml vials. No associated side effects with CF. Alternative are 2 ml of normal saline.

# **Mucolytics**

**Mucomyst** -1 or 2 mls comes in 2 strengths 10 % and 20% we are able to increase the dose as needed depending on mucous in the lungs- it helps dilute the mucous so that it can be coughed out easily.(breaks it down)thereby helping prevent the growth of bacteria. -4 ml and 10 ml vial - good for 48 hr in fridge

Normal Saline- used as a diluents for the mucomyst and albuterol if cromolyn is unavailable – it is tolerated well in the lung tissue. (unit dose albuterol contains NS or 2 ml of NS)

**Hyper saline- 7%-** comes in 4 ml vial, the increased amount of sodium in it helps dilute the mucous, has been shown to improve FEV1 if productive.

#### **Pulmozyme**

Used as a mucolytic to help thin down the mucous – used 1 or 2 x a day if needed. Pulmozyme acts like "scissors" in the mucus, cutting up the DNA strands that make the mucus associated with CF thick and sticky. This thick, sticky mucus blocks airways, promotes harmful bacteria that cause infection, and starts



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inflammation, resulting in permanent lung damage and loss of lung function and slough off the cells, thinning mucous in the process.

## **ANTIBIOTICS**

**TOBI -** Targets Pseudomonas Aeruginosa in the lungs decreasing the bacteria load, and hopefully improving symptoms and lung function

Cayston (Inhaled Aztreonam) - Inhaled antibiotic via e-flow nebulizer, acts similarly to TOBI

**Colistin -** Another inhaled antibiotic that can help with PA- if TOBI is not tolerated or PA is resistant to other medications

# **Airway Inflammation and Reversibility**

**Flovent:** Inhaled steroid used to reduce the inflammation in the lungs with associated asthma reversibility- 3 strengths to methods, MDI with spacer, Dry powder discus- last 12 hours taken 2 xs daily

**ADVAIR:** Combination of Flovent and long acting bronchodilator Serevent; used for asthma symptoms associated with CF, MDI with spacer, and discuss.