Cystic Fibrosis (CF) is a genetic condition that thickens mucus in a variety of organ systems. Patients with CF will often have problems with their lungs and digestive systems. Thickened mucus in the lungs blocks airways and leads to infections. Thickened mucus also blocks ducts and causes scarring in the pancreas, intestines, and liver. Men and women with CF can have decreased fertility or infertility.¹

According to the Cystic Fibrosis Foundation Patient Registry, in the United States:

- More than 33,000 people are living with CF (more than 70,000 worldwide).
- Approximately 1,000 new cases of CF are diagnosed each year.
- More than 75 percent of people with CF are diagnosed by age 2.
- More than half of the CF population is age 18 or older.²

Treatments

While there is no cure, treatments for CF have come a long way. In the 1950s, a child with CF rarely lived long enough to go to elementary school. Today — thanks to advances in treatments — the predicted survival age is close to 40.² Each person’s CF symptoms vary widely, and there is no one set treatment that works for everyone. Your doctor will create an individualized treatment plan for you or your child.

Airway Clearance Therapy (ACT)

One of the first goals of treatment is to keep the airway clear. Airway clearance techniques loosen the mucus in order for you to clear it. ACT can be done by clapping a cupped hand on the front and back of the chest, or via a mechanical device such as the vibrating vest. Older kids and adults can do ACTs on their own, while infants and toddlers will need help from a parent or caregiver.³

Medications

Medication therapy is an important part of your CF treatment plan. Your doctor may prescribe some or all of these medications:⁴

- **Antibiotics**: kill bacteria and prevent/control lung infections
- **Drugs to thin the mucus**: makes coughing up mucus easier and improves lung function
- **Bronchodilators**: open your airways
- **Oral pancreatic enzymes**: help you digest and absorb nutrients

Adherence

It's important to take your medications as instructed! Failing to do so can increase the risk of hospitalization, increase the length of hospitalization, lead to pulmonary episodes, and lower lung function. Non-adherence is also associated with high mortality rates.⁵

Following the doctor’s treatment plan can literally be a matter of life or death. If you need help, talk to your stePS nurse. We will help you identify what barriers are in the way, and work with you to help you overcome them.

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Nutrition

Remember, CF doesn’t just affect the lungs. CF can affect other areas in the body, too. When mucus builds up in the pancreas, the pancreas isn’t able to produce the enzymes the body uses to digest and absorb protein and fats. Therefore, it is recommended that people with CF eat high-calorie and high-protein foods throughout the day.

The U.S. National Library of Medicine makes the following recommendations for adding protein and calories to your diet:6

- Add grated cheese to soups, sauces, casseroles, vegetables, mashed potatoes, rice, noodles, or meat loaf.
- Use whole milk, half and half, cream, or enriched milk in cooking or beverages. Enriched milk has nonfat dry milk powder added to it.
- Spread peanut butter on bread products or use it as a dip for raw vegetables and fruit. Add peanut butter to sauces or use on waffles.
- Skim milk powder adds protein. Try adding two tablespoons of dry skim milk powder in addition to the amount of regular milk in recipes.
- Add marshmallows to fruit or hot chocolate. Add raisins, dates, or chopped nuts and brown sugar to hot or cold cereals, or have them for snacks.
- A teaspoon of butter or margarine adds 45 calories to foods. Mix it into hot foods such as soups, vegetables, mashed potatoes, cooked cereal, and rice. Serve it on hot foods; hot breads, pancakes, or waffles absorb more butter than cool ones.
- Use sour cream or yogurt on vegetables such as potatoes, beans, carrots, or squash. It can also be used as a dressing for fruit.
- Breaded meat, chicken, and fish have more calories than broiled or plain roasted.
- Add extra cheese on top of frozen prepared pizza.
- Add coarsely chopped hard cooked egg and cheese cubes to a tossed salad.
- Serve cottage cheese with canned or fresh fruit.
- Add grated cheeses, tuna, shrimp, crabmeat, ground beef, diced ham, or sliced boiled eggs to sauces, rice, casseroles, and noodles.

Enzymes, vitamins, and salt:

- Most people with CF must take pancreatic enzymes. These enzymes help your body absorb fat and protein. Taking them all the time will decrease or get rid of foul-smelling stools, gas, and bloating.
- Take enzymes with all meals and snacks.
- Talk to your doctor about increasing or decreasing your enzymes, depending on your symptoms.
- Ask your doctor about taking vitamins A, D, E, K, and extra calcium. Special formulas exist for patients with CF.
- People who live in hot climates may need a small amount of extra table salt.
Eating patterns:

- Eat whenever you are hungry. This may mean eating several small meals throughout the day.
- Keep a variety of nutritious snack foods around. Try to snack on something every hour, such as cheese and crackers, muffins, or trail mix.
- Make an effort to eat regularly, even if it is only a few bites, or include a nutritional supplement or milkshake.
- Be flexible. If you are not hungry at dinner time, make breakfast, mid-morning snacks, and lunch your main meals.

The Academy of Nutrition and Dietetics provides a sample 1-Day Menu for people with CF:

<table>
<thead>
<tr>
<th>Meal</th>
<th>Menu</th>
</tr>
</thead>
<tbody>
<tr>
<td>Breakfast</td>
<td>1 – 2 large eggs scrambled in 1 tablespoon of butter</td>
</tr>
<tr>
<td></td>
<td>2 slices of whole wheat toast with butter</td>
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<tr>
<td></td>
<td>6 oz orange juice</td>
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<tr>
<td>Snack</td>
<td>Instant pudding made with evaporated milk</td>
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<tr>
<td>Lunch</td>
<td>Tuna salad (tuna canned in oil, hard-cooked egg, onion, pickle relish, and mayonnaise)</td>
</tr>
<tr>
<td></td>
<td>6 club crackers</td>
</tr>
<tr>
<td></td>
<td>2 canned peach halves with 2 tablespoons cottage cheese and 4 walnut halves</td>
</tr>
<tr>
<td>Snack</td>
<td>Fruit smoothie (apple juice, banana, frozen strawberries, and 1/4 cup nonfat dry milk)</td>
</tr>
<tr>
<td>Evening Meal</td>
<td>3 oz ground beef patty with gravy</td>
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<tr>
<td></td>
<td>Baked potato with butter</td>
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<tr>
<td></td>
<td>Broccoli with cheese sauce</td>
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<tr>
<td></td>
<td>2 slices bread with butter</td>
</tr>
<tr>
<td>Snack</td>
<td>1 scoop of ice cream with chocolate syrup</td>
</tr>
</tbody>
</table>
Exercise

CF patients who exercise regularly feel better than those who don’t.\(^7\) There are studies that show lung function declined more slowly in CF people who exercised than those who did not.\(^7\) However, it’s important to talk to your doctor before starting any exercise plan. If you are going to a CF Care Center, they will also provide advice and support.

The CF Foundation Education Committee suggests three things to consider when choosing an exercise activity:

1. Do you like the activity?
2. Does the activity connect you to friends or family?
3. Are the activities cardiovascular exercise (do they work out your heart and lungs)? Can you do them for longer than 20 minutes at a time?

If you have a physical therapist, ask them to design an exercise program that will work best for you. Also, keep these things in mind when you are looking for a new exercise routine:

- Think of exercises that use multiple major muscle groups at the same time like walking, jogging, swimming, and/or dancing. Remember, the more you enjoy it the more likely you will stick with it.
- Look for moderate intensity. Moderate intensity means that you should be able to carry on a conversation comfortably while your heart rate climbs, your breathing rate increases, and you sweat a little.

Stay hydrated

Because of the amount of salt lost when you sweat, dehydration can be dangerous for CF patients. This is especially important during the summer months when you may be playing sports or in a place that is hot and humid.

Stay on top of your hydration! Drink more fluids before, during, and after you exercise or spend a lot of time outside. Here are some other things you can do to help:

1. Drink sports drinks; they are a good source of electrolytes.
2. Increase your salt intake by snacking on something salty: chips, pretzels, nuts, crackers.
3. Try to avoid caffeine, which can make you dehydrated from fluid loss.\(^8\)

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