Anticipatory Guidance: Infants

Q: What are my child's mutations?

The Cystic fibrosis transmembrane conductance regulator (CFTR) is the gene that is associated with cystic fibrosis (CF). There are many different mutations of this gene that can cause CF. Knowing your child's CF specific mutations can be important in helping treat their disease. Write your child's two gene mutations in the spaces below.

My child's mutations are: _____ & _____.

Q: How frequent are my appointments?

Every patient with CF should be seen at least once every 3 months (quarterly visits: 4 times per year) to check in on how their current treatments are working. These visits help assess lung function, nutritional status and current symptoms to provide early treatments to keep your child as healthy as possible. If your child is sick or trying to gain weight, they might be monitored more closely and require more frequent CF clinic visits. Babies are often seen more frequently (sometimes weekly) to make sure that they are growing well and getting all of the nutrition that they need.

Q: How long are my appointments?

Appointments at Rady's Children's Hospital CF Clinic typically last between 1.5-3 hours. Because of this extended appointment time, you should plan to bring enzymes, snacks, activities, diapers – anything needed for your child during this time frame.

Q: Can someone else bring my child to clinic?

Rady Children's Specialty Clinic requires that a legal guardian attend the clinic visit until the age of 18. The treatment and management of CF is very involved and requires a lot of attention from parents/guardians and caregivers. Therefore, it is preferred that at least one parent/guardian should attend clinic visits. It helps parents/caregivers understand their child's health and care if they attend clinic visits as frequently as possible. We also encourage you to bring grandparents, nannies and any other caretakers for your child to appointments so that they can receive education on your child's care.

We understand that life gets busy sometimes, and there may be times when parents can't make it to a scheduled CF appointment. In this case, another trusted adult may bring your child to clinic if you write and sign a permission form for them. Parents can also sign a release form to be kept on file at the CF clinic that allows another adult to bring their child to clinic. To have this form completed, please ask your clinic social worker.

During the COVID pandemic, the hospital has temporarily changed the policy and only 1 caregiver is permitted to attend appointments per patient. Siblings are not permitted to attend visits during this time as well. Please call ahead if you have questions or need to ask for a one-time exception so that we can ask for administrative clearance ahead of time.

Q: Does CF affect my child's immune system?

CF does not directly affect your child's immune system. In CF, the CFTR protein isn't functioning normally, which causes thick mucus to be made. This thick mucus traps germs in the lungs and creates an environment for germs to thrive. Your child's immune system then targets these germs and causes increased inflammation in the lungs, which causes more mucus to be produced. This excess mucus traps in more germs, and the cycle continues.

The thick, sticky mucus affects your child's ability to fight off infections:

1) Children with CF can experience more intense infections than children without CF who are infected by the same germs.
2) Some bacteria - such as Pseudomonas - can cause infection in people with CF, but not in individuals without CF. These germs are particularly concerning for people with CF and can have negative effects on lung function. People with CF can spread these germs to each other.

It is very important that all people with CF receive immunizations to protect them from disease. They should receive all of the recommended vaccinations, including the yearly flu vaccine starting at 6 months of age. It is also important for anyone who will be in contact with a child with CF to get their vaccinations. Family members and friends who are going to be around babies who are too young for the flu vaccine (under 6 months old) must get a flu vaccine.

Q: Does CF affect my child's development?

CF does not affect your child's ability to learn. It can, however, have an effect on your child's ability to grow. In people with pancreatic insufficiency (about 80% of CF'ers) due to CF, the pathways in the pancreas become blocked by sticky mucus which prevents pancreatic enzymes from entering the small intestine. Without the enzymes from the pancreas, protein and fat from food are not digested or absorbed well (malabsorption). This can lead to your child having problems gaining weight, which can cause poor growth and delayed puberty. As an infant, this can lead to impaired brain growth if severe. There are many strategies for dealing with malabsorption in CF patients, including enzyme replacement therapy, increased calorie intake, nutritional supplements, and feeding tubes. If your child begins to struggle with growth, your CF care team will help you decide which options are best for your child.

Q: When do I call clinic?

It is important to learn your child's baseline symptoms with CF. Once you know your child's health status on illness-free day, you have a comparison point for when their symptoms start to worsen. However, learning this about your child can take time. If things appear to be worsening for your child's lungs, sinuses, or stomach (all of which are affected by their CF), it is important to call the CF center. If it is a concern that does not relate to CF – for example, fevers after immunizations - you should call your child's primary care doctor.

When a child with CF has a pulmonary exacerbation or their lung symptoms get worse, we want to act quickly to avoid lung damage. If you suspect a flare-up, you should contact your CF care team so that your

child can be evaluated. Parents of children with CF often report symptoms such as- their child has less energy than normal, increased mucus, nasal congestion, increased coughing, and signs of malabsorption (oily stools, more frequent stools or belly aches) that lead them to think their child is having a flare-up. When changes like this are observed, it is time to call the CF care team.

Q: What would lead to my child needing to come to the hospital?

Pulmonary exacerbations, or a worsening of your child's respiratory symptoms, are the most common cause for hospitalizations in children with CF. To avoid causing permanent lung damage, pulmonary exacerbations should be treated promptly. Some of the less severe exacerbations can be treated at home, but some require admission into the hospital because of the need for IV antibiotics and enhanced support with airway clearance techniques and nutrition.

Q: What should I expect if my child is hospitalized?

If your child needs to be hospitalized for a flare-up, it is likely that they will need intravenous (IV) medications. IV antibiotics are given through a plastic tube that is placed in a vein. Because the medicine is going directly into the patient's blood stream, it can be more effective in fighting off your child's infection than medicine taken by mouth. When a person requires IV medications for a longer time (more than a few days), a Peripherally Inserted Central Catheter (PICC) line is preferred. PICC lines have the benefits of staying in longer than the smaller IVs and can allow blood to be drawn for tests without additional needle pokes for your child.

The treatment regimen for CF that your child was taking at home will be continued in the hospital, but some aspects might be modified - such as more frequent airway clearance technique sessions (vest or chest physical therapy). Some other medications might be increased or added as well. We know how important nutrition and a healthy weight are in maintaining healthy lung function, so nutritional aspects of your child's care will also be assessed and monitored during your stay. All of the meals, supplements, snack and drinks that your child needs to meet his or her nutritional needs will be available to them in the hospital. You can bring in some favorite foods or snacks for them from home as well.

We may ask you to bring your home supply of certain medications (like the CFTR modulators) that the hospital does not stock, most of the CF medications though are available and will be provided by the hospital. When you are admitted, all medications must be administered under supervision of the inpatient nurse or respiratory therapist, even the ones you brought from home. Toys and entertainment can be brought in from home as well, check with your nurse about larger electronics.

When in the hospital, you will be using a disposable hospital nebulizer that is cleaned after each use, but it is discarded 24 hours after use. These are different from the nebulizers you use at home that can be sterilized daily and last for up to several months. The average hospital stay for a CF flare-up is one to two weeks as this is how long the typical IV antibiotic course lasts. Depending on the severity of the flare-up, hospital stays can be shorter or longer.

All hospital staff members who enter your room will be wearing a gown and gloves to avoid transmitting germs to your child. If you are coughing excessively or we think you have an active viral illness (like the flu) or if the staff member will be doing a "cough inducing procedure", the staff member will have on a gown, gloves and a mask. CF patients are required to wear a mask outside of their hospital room, which protects them from germ droplets that might be in the air throughout the hospital.

Q: Why do CPT if my child is not coughing?

Chest physical therapy (CPT) is an airway clearance technique that is used to free mucus in your child's lungs so that it can be coughed up. We know that babies have mucus in their lungs early in life, even before we can hear anything with our stethoscopes or see anything on a plain x-ray. When the thick mucus is removed, it is no longer blocking airways and germs can no longer grow in it. Removing mucus contributes a lot to keeping CF patients infection-free, so we want to do CPT even when the child does not seem ill or is not coughing.

Q: How does my child crying affect their treatments?

The goal of using nebulized medicine as a treatment for your child's CF is to get the medication far down into your child's lower airways. There are many factors that go into getting the dose of medicine to the lower airways - one of which is the fit of the mask to your child's face. If the mask does not fit snugly, regular air will be mixed in with the nebulizer treatment, and your child will not get the appropriate dose of their medicine. Another factor that affects the medicine's ability to get into the lower airways is the behavior of the child. More medicine gets to the lower airways when your child is calm and breathing at a steady pace. As you can imagine, crying disrupts a normal breathing pattern and causes your child to exhale for a long time with short bursts of inhalation. This means that more of the nebulized medicine stays in the upper part of the respiratory tract, and less reaches deep into the lower part where it needs to be. Crying can also disrupt the way the face mask fits on the child's face.

While your child sleeps their breathing pattern is different as well. This sleep breathing also prevents medicine from getting into the lower respiratory tract. Attempting to place a mask on a sleeping child's face will likely wake them, and can cause future anxiety about using the mask at all. Thus, it is not advised to plan to do nebulizer treatments while your child is sleeping.

Q: Why do I give salt and how much should I give?

People with CF lose a lot of salt in their sweat. Baby foods, formula, and breast milk contain low amounts of salt. Because salt balance in the body is very important, extra salt should be added to the diet of an infant with CF.

- For children less than 6 months old, 1/8 teaspoon of table salt should be added to their food every day.
- For children older than 6 months but younger than 2 years old, 1/4 teaspoon of table salt should be added to their food every day.

The salt can be added to formula or milk and fed to the child over the course of one full day. Salt can also be added to cereals or baby foods. Morton lite salt can be used as well but at double the amount as regular table salt.

When people sweat more, they lose more salt. For this reason, on particularly hot and sweaty days, extra salt may need to be added. Your CF health care team can help you make a plan for how much salt to add in these situations, as adding too much salt can have health consequences as well. Always give your child the amount of salt that your CF care team recommends.

Q: Stools, poops, bowel movements, #2?

Because of the effects that CF can have on food digestion, paying attention to your child's stool is important as changes in stool can be a sign of malabsorption. Notify your CF care team if you notice any of the following changes to your child's stool habits:

1) Larger or more frequent stools: Most children have one to three stools per day, but babies can have more (after every feed) or less (every 2-3 days). The size and frequency of stools can vary from child to child, so pay attention to what's normal and usual for your child.

2) Change in consistency and appearance of stools: With malabsorption, stools can start to look soft, but they don't often look watery. Because the child is not absorbing fat well, the stool may look like it has excess oil in it or appear "greasy." The stools may also be more smelly than normal.

Other changes: When undigested food goes from the small intestine to the large intestine, bacteria in the large intestine break it down and create gas. With malabsorption, more undigested food is making it to the large intestine, so more gas is created. This increase in gas also can lead to bloating, which is when the abdomen swells out. If your child is experiencing increased gas and bloating, it may be a sign of malabsorption.

Q: How do I get medication refills?

For your child's CF treatment plan to work, they must take all their medications as prescribed. Ensuring that you always have medicine when you need it requires a lot of organization. Many people find that using phone calendars that send alerts or an easily visible calendar with the medication re-order date on it can aid in remembering. Order the medication refills 7 days before you run out of medication. This will give you room for error in case something happens, and you can't get to the pharmacy for a few days. To order refills, look for a web address or a telephone number on the label of the medication containers. You will want to have the prescription number ready when you order. If you are using a mail-order service, verify your address and phone number with them regularly so there is no delay in getting your medicines to you.

Q: How do I give enzymes, and do I give them with every feeding?

Enzymes should be given with every meal and snack that contains fat, protein, or complex carbohydrates. This includes breast milk, formula, nutritional supplements, and tube feedings. Remember, without the help of the enzymes, your child is not able to get the energy from food that they require to grow and be healthy.

There are certain foods that don't require enzymes because they contain only "simple carbohydrates" (sugars). These foods are easy for the body to digest without the help of pancreatic enzymes. Some examples include: fruit juice, flavored ice, hard candy, tea and fruit snacks.

If a child cannot yet swallow capsules that contain enzymes, open the capsule and pour the enzyme beads onto a spoonful of a soft, acidic food. The typical suggestion is applesauce, but once your baby is greater than 6 months of age you could also use different pureed baby fruits, tomato paste, jelly, or even ketchup. We use an acidic food because the enzyme beads are covered in a coating that dissolves away in basic/alkaline environments. This is helpful because the small intestine is basic/alkaline and will dissolve the coating so that the enzymes can work. We do not want the coating to dissolve until it reaches the small intestine. Therefore, the enzymes should be given with acidic food.

You also don't want to pre-mix the enzymes before giving them to your child. It is best for the enzymes to first contact the food substance right before it is swallowed. If your baby spits the enzymes out, just spoon them up and gently put them back into their mouth. Enzymes don't have a taste, so it is the new texture that the baby is unsure about.

The enzymes should be swallowed whole, and not chewed. If the enzyme beads are crushed, there is a risk that the enzymes won't function well or will irritate the child's mouth. Have the child drink something after taking enzymes to make sure that the mouth is clear. This also applies to the mother's breast when breast-feeding a child who has just had enzymes. Make sure that the child's mouth does not have beads in it before he or she latches on the breast to avoid breast irritation.

Older children and adults with CF can take the enzymes in capsule form. Enzymes may also be taken with tube feedings. There are many ways to accomplish this, so work with your CF care team to figure out what works best for you.

Q: In what time interval should enzymes be given? How long are they good for?

Enzymes come in capsules with small beads inside them. These beads are the enzymes covered in a coating that dissolves in the small intestine, where the enzymes help our body digest food. Enzymes are most effective for 45 to 60 minutes after they are swallowed, so they should be taken immediately before meals or snacks. Enzymes need to be stored at room temperature (59 degrees F to 86 degrees F) and not allowed to get too hot or too cold. For example, they should not be stored too close to a stove or in the refrigerator. They also should only be taken if they are within their expiration date. When picking up enzymes at the pharmacy, it's important to get the specific brand that your CF care team prescribed. Check with your care team before making any changes to your enzyme regimen.

Q: Hot tubs—what are off limits?

Hot tubs have been found to be places where bacteria called Pseudomonas aeruginosa can thrive. Pseudomonas is one of the bacteria that specifically causes illness in people with CF, so hot tubs should be avoided. Additionally, any pools or water that are not adequately chlorinated to kill Pseudomonas can pose the same risk to people with CF. You can contact your health department to see what levels of chlorine are required to kill Pseudomonas and whether or not your local pool meets the requirements.

Some communal play areas, such as those found in malls and restaurants, should be avoided if the cleanliness level is not satisfactory. Many children pass through these areas and spread germs onto toys - ball pits in particular are very hard to fully disinfect.

Q: How do I cope with roller coaster feelings?

Receiving a CF diagnosis is a major life event for a family. If you are struggling with where to begin, try sitting down with your calendar and finding out how to make the new responsibilities of CF care a part of your schedule every day. Once you've figured out what times CF treatments need to be done, break up the tasks among different family members. Dividing the workload will prevent any single person from feeling that they have to shoulder all of the responsibility. When you're having a down day, reach out to trusted individuals about what you're feeling. If you're searching for advice from someone who has been through this before, look for people in your online CF network. Even with all of the best intentions, things will not go perfectly every time. Be patient with yourself as you learn what works best for your family.

Q: What resources should I use for information about medications and supplements?

Learning about all of the different CF medications and how they work can seem like a daunting task. The Cystic Fibrosis Foundation has created a helpful section on their website that provides information on the different types of CF medications.

https://www.cff.org/Life-With-CF/Treatments-and-Therapies/Medications/

If your questions regarding CF medicines are more specific, DailyMed is a useful online source of information about the different medicines - including how to take them and possible side effects. To use this resource, go to the following link and search for the drug in the search bar at the top of the page.

https://dailymed.nlm.nih.gov/dailymed/

Q: How can I get help with insurance problems?

Dealing with insurance can be intimidating for many people, but the good news is that there are many different people available to help you navigate your claims. Talking with the social worker that is part of your CF care team is a great place to start, as they can help you figure out what kind of benefits you qualify for. The hospital has financial representatives who can explain the bills you receive for your child's hospital stays. They can also help you come up with a payment plan and give you reports on what has been paid so far.

When speaking with your insurance company, it is a good idea to ask to be assigned a case manager. Once assigned, this person can help you have full knowledge about what will be covered by your current plan. They can also help you with making an appeal if you disagree with a decision your insurance company has made regarding your coverage.

The Cystic Fibrosis Foundation has an assistance service of its own, called *Compass*. *Compass* can help you work with your insurance company, and can help you do comparisons between different insurance plans to find one that works best for you. Compass also has the ability to find local resources for you, such as mental health resources, peer mentoring, and locating clinical trials.

You can find more information about Compass at:

https://www.cff.org/Assistance-Services/How-Compass-Helps-People-With-CF-and-Their-Families/Understanding-Insurance/Your-Insurance-Plan/The-Insurance-Basics/

You also can contact *Compass* at:
844-COMPASS (844-266-7277)
Monday - Friday, 9 a.m. - 7 p.m. ET
compass@cff.org

Q: What are reliable websites to learn more?

Cystic Fibrosis Foundation
www.cff.org

MedlinePlus
http://www.nlm.nih.gov/medlineplus/cysticfibrosis.html

PubMed Health
https://www.ncbi.nlm.nih.gov/pubmed/