Q: Helpful parenting techniques/websites?

Positive and/or differential reinforcement techniques have shown to be the most evidence-based and effective techniques for children and families. The techniques have been developed through decades of research. The Kazdin Method is one (from Yale University) that can provide a number of helpful techniques to shape positive behavior and help decrease frustration for children AND parents. http://alankazdin.com/portfolio-items/parenting-defiant-child/

Another helpful book that emphasizes healthy parent and parent/child communication: "How to Talk so Kids Will Listen…" The book includes easy to use and practice techniques with simple steps that are easy to learn. Results from the active listening techniques and reflection communication skills can often improve emotional interactions, even after a short time of implementing them.

https://www.amazon.com/How-Talk-Kids-Will-Listen/dp/0743525086

Love and Logic (available in clinic, please ask!)

The CFF has several articles and videos that give guidance, check CFF.org for more information. www.cff.org/Life-With-CF/Caring-for-a-Child-With-CF/Parent-and-Guardian-Guidance

Q: Do I give enzymes with milk?

Enzymes should be given with every meal and snack that contains fat, protein, or complex carbohydrates. This includes drinks (like milk or milkshakes), nutritional supplements, and tube feedings that contain these nutrients. The foods that don't require a dose of enzymes are those that contain only "simple carbohydrates" (sugars). These foods are easy for the body to digest without the help of pancreatic enzymes, and include fruit juice, flavored ice, hard candy, tea, fruit snacks, etc.

Q: What happens if we mess up an enzyme dose somehow?

Enzymes are very important to a child with CF's health, and we want them to have them with every meal or snack that requires them - but too many enzymes are not a good thing. Having too many enzymes in a day is only a problem if it happens repeatedly over many days or weeks. Having more than the maximum enzymes one day because you were at a birthday party and had pizza, ice-cream and cake is okay.

When a child takes enzyme beads, we want to make sure that they drink enough liquid afterwards to completely clear their mouth. If enzyme beads remain in the mouth for an extended period of time, they can cause irritation.

If a child forgets to take enzymes with one meal, they should continue to take their regular enzyme dose at the next meal. You should not alter or change your enzyme dose without first consulting your doctor - this means that you should not double up on enzymes in the subsequent meal to make up for the missed meal.

If you give enzymes to your child with CF and he or she refuses to eat, the enzymes travel to the intestines but don't have any food to work on. When the enzymes lack food materials to digest, they may

cause side effects, but usually there are no symptoms. This is usually not a problem if it only happens once in a while.

Q: Can I do a vest treatment while my child is sleeping?

The purpose of the vest is to loosen up mucus in the airways so that it can be coughed out easier. There will be pauses at certain intervals in the vest treatment for your child to huff cough to expel the mucus that has been loosened by the vest. This requires your child to be awake and participating in the airway clearance. If they do not cough out the mucus after the vest shakes it loose, the mucus will resettle into the airways, and the benefit of the procedure will likely be lost.

Additionally, your child will often be taking a nebulizer treatment while they have a vest on. While your child sleeps their breathing pattern is different than when they are awake. This sleep breathing pattern prevents medicine from getting where it needs to go in 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. You should not plan to do nebulizer and/or vest treatments while your child is sleeping.

Q: Information about alternative supplements/treatments?

Alternative CF medicines or supplements are things many CF families have questions about; however, there is a great need for extensive studying of these alternative treatments. Before any of them can be supported by the medical community, we need to know things like: how effective are they? what interactions do they have with other CF treatments? what is the right dose? what are the side effects in the short term and long term? Remember, supplement and alternative treatment should be evaluated and considered just like all the other medications you child takes for CF.

If you are looking at alternative CF medicines for your child, it's important to know that they may be processed by the liver, just like some of your child's other CF medications. This can change the effectiveness of your child's CF treatment plan, and has the potential to put strain on the liver.

Your ideas and beliefs are important to your CF care team. If you decide to use an alternative medicine product, even if your doctor advises against it, you should absolutely be open with them about your decision. They can help you formulate a plan to monitor the product's effects on your child's body. They will want to keep an eye on the body systems that could be affected by the alternative medicine – one way they might do this would be by doing regular kidney or liver tests. They also might want to set up a timeline for use of the supplement so that you can decide whether or not to continue it after observing its effects.

Q Can my child go to preschool? What should I ask about?

The goal with talking to your child's (future) teacher is to help them gain a better understanding of what CF is so that they can help keep your child as healthy as possible. The staff at the school should understand that CF is a genetic disease that doesn't delay your child's learning. They should also know that your child will need to cough frequently and that their cough does not mean that they have

something contagious that could be spread to other students. Your child might also need to eat snacks more often and have more frequent bathroom breaks than other children.

Most of your questions for a preschool will be about infection prevention. You will want to know about:

- The procedure for disinfecting the classroom and toys
- Hand hygiene practices for staff and chilren
- The policy regarding sick children
    - People with CF are at risk for more intense versions of common colds or the flu, so to help lower the number of illnesses a child with CF is exposed to, any child that is sick should not attend school. Most daycares and schools have this policy but how it is enforced should be asked.

Q: When do we start pulmozyme and hypertonic saline?

At age 6 it is recommended for all patients with CF. We will start it earlier on a case by case basis if it is needed.

Pulmozyme (dornase alfa) is a mucolytic. This means that it breaks up or thins the mucus in the lungs, making it easier to cough out. It is an inhaled medication that requires refrigeration and is delivered using a nebulizer and compressor.

Hypertonic saline is highly-concentrated salt solution that is inhaled into the airways. The salt collects in the mucus, which helps draw more water into the mucus from the cells. This increase in water content thins the mucus and allows it to be removed easier during airway clearance techniques. It is also delivered by nebulizer and compressor but does not require refrigeration.

The CF medicines should be taken in a specific order to help them work most effectively. You should have received this “order of therapies” handout if you child is on more that 1 inhaled medication. If not, please ask us for one at your next clinic visit or through MyChart.

Q: When should we switch to a vest?

High-frequency chest wall oscillation (HFCWO) vests can start to be used as an airway clearance technique as soon as the child with CF is big enough to fit properly into the smallest size. The decision of whether or not your child should change to using a vest is based on their size, not their age. Your child should also be able to sit up independently to be able to use this device appropriately. The RT in clinic can measure your child’s chest circumference to see if they are big enough to consider this therapy.

Q: What is pseudomonas?

Pseudomonas aeruginosa is a type of bacteria that is found in many places in the environment. Pseudomonas grows well in areas that are moist, like CF equipment, sinks, drains in bathrooms and kitchens, and hot tubs and pools that are not adequately chlorinated. It can also be found in the soil and in areas of natural water, especially stagnant ones.

Importantly, if a person has Pseudomonas in their lungs, they are able to spread the disease to other people who have CF or weakened immune systems (like people on chemotherapy or who have an immune dysfunction). The spreading can occur by direct contact with an infected person or by indirect contact with an object that the infected person has shed germs on. This sharing of germs between people with CF is the basis for a lot of the guidelines that have been made about keeping CF patients a safe distance apart from one another.

Q: Can my child have playdates with other kids with CF?

As we have learned, some germs - like Pseudomonas aeruginosa and Burkholderia cepacia complex - have a higher risk of affecting people with CF. The infections that these germs cause can be spread between people with CF while not posing a risk to healthy individuals. For this reason, guidelines have been developed regarding contact between people with CF. Researchers have found that when a person coughs, droplets from the cough can travel as far as 6 feet. The CFF uses this information in recommending that people with CF remain 6 feet (2 meters) apart from each other at all times. These guidelines don't apply to family members with CF.

As you can imagine, these safety guidelines mean that certain activities involving more than one person with CF should be avoided. Close contact activities such as playdates, handshakes, sharing hotel rooms, being in the same classroom, or car rides involving more than one person with CF can contribute to the sharing of germs and an increase in CF exacerbations. Similarly, things like CF-specific camps are not a good idea.

We advise against having in-person meet ups with other children (or adults) with CF. Despite these guidelines, it is understood that forming relationships with other people who have CF can have a very positive impact. If you or your family is searching for advice or support from other people with CF, there are virtual ways to connect with other families that have a child with CF.

Q: Can my child take swimming lessons?

Because Pseudomonas has been found in aquatic places, people with CF must approach activities that involve water with some caution. Hot tubs and spas should be avoided by people with CF because Pseudomonas grows well there, and has been traced back to them. Stagnant water should also be avoided for this same reason.

Swimming pools that have been chlorinated adequately to kill Pseudomonas are acceptable for CF patients to swim in. This means that your child should be allowed to take swimming lessons as long as the facility is following public health guidelines for maintaining appropriate chlorine levels.

The CFF does not have a formal recommendation for or against people with CF swimming in other aquatic places such as oceans, rivers, lakes, and ponds that are not stagnant. More research is needed about these locations before formal recommendations can be made.

Q: Can we have pets?

Zoonotic infections are infections that can be passed back and forth between animals and humans. There are many different types of germs that fall into this category, all of which we want to avoid if possible. People with CF should still be allowed to be around animals, have pets, and participate in pet therapy in health care settings if they would like; however, the CFF recommends certain things to prevent illnesses from spreading from the animal to a person with CF.

- Good handwashing skills should be practiced when interacting with animals, especially when cleaning up poops or cages/tanks.
- Any pet that is around a person with CF should receive regular preventative care from a veterinarian and be assessed right away if they seem ill.
- Farm animals' living spaces often have fecal matter spread throughout them, and their feed is a common place for molds like Aspergillus to grow. It's safer for a person with CF to avoid the types of chores that would keep them in these living spaces for too long, such as cleaning the stalls or shoveling feces.

Q: What if my child is not cooperating with treatments or medications?

CF treatments and medications can take time away from playing and other activities that children would rather be doing, so it's not surprising that they might be resistant at times; however, CF is not going to go away. To help them be as healthy as possible, parents should help them get into a normal routine with their CF care. If a child doesn't want to do a CF care task and the parent allows them to skip it, they will learn that this is an option, and the parent will be met with more resistance in the future.

When a child fights against CF treatments or does something that the parent doesn't want, the best thing to do is to pay as little attention to the bad behavior as possible. To a child, any attention from a parent is desirable, whether the attention is positive or negative. The child will want to repeat whatever behavior earned them the attention. A parent should be very positive and encouraging when the child is doing good things, and respond very little when the child is misbehaving.

For example, when a child with CF is protesting an airway clearance treatment, rather than fighting with him, the parents should ignore his resisting and verbally repeat that it is time for the treatment to begin. If the misbehavior continues after a set amount of time - say 5 minutes - the parents should put the vest on him and not allow him to take it off. It will take a few days of ignoring the resistance before some improvements might be seen. As soon as you see less struggling, you should praise their cooperation and offer a treat, such as a book to read or a video to watch during the treatment. Even if you see improvement, there will undoubtedly be slip ups where the child begins to protest again. These should be handled with a simple statement such as, "Your treatment is not finished yet," while gently restraining the child. Follow this with a quick return to the treatment. With this, you are successfully getting the treatment done while not showing attention to his protesting.

Q: How much should my child be eating?

We know that people with CF (with pancreatic insufficiency) can have trouble with the process of digestion and obtaining energy from their food. We also know that fighting off lung infections requires a lot of energy. The inability to get all of the nutrients out of food plus the increased energy requirements of a person with CF leaves them requiring more food intake than other people their age. A person with CF often needs to eat 1.5 to 2 times as many calories as a person without CF. The CF care team will determine how many calories your child needs to be consuming by tracking his or her growth.

Your CF care team will calculate your child's weight for length or BMI and use it as the value to track growth. The goal is for your child to have a BMI in the 50th percentile or higher. You can help them achieve this by sticking to the diet recommended by our dietician.

Getting your child with CF to eat a higher amount of food can be challenging. It is really important to have structured meal times and snack times every day. If a child is allowed to graze all day, it is hard to manage enzymes and can make it harder to get all of the calories they need, because they will consume less at meal times. This is important to help establish hunger and fullness cues in all children, even those without CF.

There are a lot of ways to add calories without necessarily increasing the amount of food the child with CF has to eat. These include adding extra cheese or butter, using high calorie liquid supplements, using salad dressings, adding bacon to sandwiches, adding nuts to baked goods and salads, using avocados, making foods that come with gravies or sauces, making high-calorie milkshakes or smoothies, adding whipped cream to desserts, and grilling sandwiches in butter.

Q: Why is my child a picky eater? What can I do?

It is not unusual for any toddler to be picky about what they eat. This has the potential to be a big stressor for parents of children with CF because of how important it is to keep their child's weight up. Many kids need to see a food on their plate 6 or more times before they are comfortable enough to try it. Many parents will have given up long before this sixth attempt of presenting a food. Even if a child refuses to eat a new food, the food should be left on their plate so that they can become accustomed to it even if they won't try it yet. Praise the child when they show any interest in the new food like touching, smelling, or tasting it.

Just as when a child resists CF treatment, when a child refuses to eat or try new foods we should also not pay attention to the misbehavior. This goes back to understanding how children react to emotion. We know that attention is a reward for children – whether it is good or bad attention. They will continue to do whatever behaviors we pay the most attention to. Even though it can be challenging, parents should try to ignore the mealtime behaviors that are undesirable, such as complaining about having to eat, disliking the food, extended chewing time, extended talking time, etc. Instead, parents should emphasize the things that the child is doing correctly. This praise should be as specific as possible so the child knows which behaviors you would like him or her to continue. Examples of this are, "I really like the way you are

chewing and swallowing"; "I noticed that you took 2 big bites of your taco in a row and I am proud of you"; "It was very brave of you to try that new food".

One tactic for dealing with picky eaters is to use choices to give the child a sense of control over meals. For example, at lunch time you could ask, "Do you want a peanut butter and jelly or a ham sandwich? Do you want two slices of cheese on the ham sandwich or three? Do you want mustard or mayonnaise or both? Carrots or celery? Ranch or Italian dip? Orange or apple slices? White milk or chocolate?" Using choices and involving them in the planning and preparation of a meal can help make kids more interested in eating and less resistant at meal times.

Making your child swallow the food once they have it in their mouth is not a good idea, because it can make them afraid to taste new foods in the future. Instead, they should be allowed to spit the food out and praised for at least tasting the food. It's also a good tactic to avoid labeling foods that the child has rejected as foods that they don't like. You can encourage them to try again when they're older by calling such foods "foods that they are not big enough for yet." You can also use rewards like reading a book, or a trip to the park to encourage them to eat their meal or taste a new food. This can be part of a sticker chart if you are using that at home.

People with CF have an increased calorie demand compared to their healthy peers. Having to eat so much can be impactful on a child. At a time other than mealtime, ask your child how they are feeling about their enhanced diet with CF. Help them understand the reasons they should eat healthily, so that they can spend more time playing with siblings and friends and less time recovering from CF illnesses.

The social worker, nurse, and dietician are all excellent resources within your CF care team to help you improve your child's eating habits. They can help you figure out the best way to get your child to eat, and they can also connect you with other CF parents who have successfully navigated the "picky eater toddler stage" of their child with CF.

Q: What if we are having potty training issues?

Toilet training can sometimes be seen as a dreaded adventure, and parents of a child with CF might find it even more intimidating because of the effects that CF can have on their child's bowel movements. Because children with CF often have to poop more frequently and can have smellier and softer stools, it can make toilet training for them a bit harder. Typically, children start to be ready for potty training between 1.5 and 2.5 years old. Any given child could take longer to become ready, especially the children with CF who have the additional complication of messy and frequent stools. Your child with CF may take until they are 3 or 4 years old before they are ready to become potty trained, and this time extension shouldn't be a source of concern for you.

Awhile before you think your child is ready to begin toilet training, some parents have found it helpful to get a few children's books on the subject so the child can become familiar with the idea. Some ways to know if your child is ready to start potty training are if they can go for 2 hours without wetting their diaper, if they can walk, sit up on a toilet, pull their pants down and up, and communicate enough to respond to small commands and tell you when they need to go to the bathroom.

Once you have determined that your child might be ready to try potty training, show them the child-sized potty that you will use. At first, you can have them sit on it with their clothes on to gain comfort with it. You might consider taking a dirty diaper and emptying it into the small potty and then full-sized toilet to demonstrate what they are used for. Show them praise whenever they show interest in the potty or sit on it for any amount of time. When you have them sit on the potty without a diaper, do so at pre-arranged times at first. Having them sit on it in the morning, after naps, and for a few minutes every couple of hours throughout the day will help them begin to think about going to the bathroom regularly. Make their time spent on the potty chair a positive thing by letting them choose a book to read or a game to play. Accidents happen – if they forget to tell you, it's best to avoid being overly negative or to associate shame and bad feelings with potty training. Neutral statements like, "You didn't tell me you had to go to the bathroom this time. Next time you'll remember," are more helpful.

If you have given potty training a try but your child doesn't seem interested in learning about it or is putting up a lot of resistance, it might be best to give it a break and try again a few months later. Potty training goes best when the child is emotionally and physically ready for it.

Q: Can we travel? What do I need to bring with me?

When traveling, it is important to have all of your CF medicine with you. It's best if you have the amount you need for each day of your trip, plus a little extra in case your travel gets extended for some reason. Depending on the length of your travel and whether or not you use a mail-order pharmacy, you may need to order refills in advance to make sure that you have enough of your CF medicines. Share your trip plans with your CF care team. Your CF physician can give you a written prescription for your medications to carry with you if you are ever in need of more. Keep in mind that if your traveling is taking you outside of the U.S., the prescriptions might not be valid, or the medications might not be available. In these cases, you will want to make sure that you carry enough medicines on your person.

If your travel plans involve flying, you should carry the CF medicines onto the plane – do not check them. This allows you to always have them accessible and ensures that they do not get lost. It also allows you to be in control of the temperature that they are kept at. Enzymes need to be kept at room temperature - keeping them with you prevents them from getting too hot or too cold while flying.

Other medicines may need to be refrigerated. To accomplish this while traveling you will want to have a cooler bag with ice packs in it. The temperature requirements of each medicine can be found on their drug packaging. Keep your medicines in their original containers when traveling. This helps you make sure that you are taking the right drugs at the right time. It also helps prove to security that the medicines belong to you.

For the medicines that need to be refrigerated, you can call your hotel room ahead of time to request that a refrigerator be there for your use. If your hotel does not offer in-room refrigerators, label your medicines with your name, hotel room number, and phone number. Ask the hotel staff to use the hotel refrigerator and freezer to keep your medicines and re-freeze your ice packs.

Don’t forget to keep your child’s calorie intake high while traveling. Pack some high calorie snacks to take with while you are on the road.

It is a good idea to look up CF centers that are near your destination, so in the event that you need to seek care, you will already know where to go. If you’re traveling within the U.S., you can find a list of CF centers on CFF.org. If you’re traveling internationally, you can find a list of CF centers on CFWW.org. You should have a copy of your insurance card and information with you in case you need receive medical care.

You can work with your CF care team ahead of time to ask for a note from your doctor that explains your diagnosis and your need for the medicines and treatment supplies that you carry with you. This can help streamline things like getting through airport security. You’ll also want to bring a copy of the instructions for your treatment supplies in case they stop working while you are traveling.

Infection prevention is a main concern while traveling as well. Make sure you have plenty of alcohol-based hand sanitizer so that you can clean your hands often, and to stay 6 feet away from anyone who appears to be sick. If you are on a plane and the person next to you is sneezing or coughing, ask to be moved to a different seat.

You will need to continue your airway clearance techniques and nebulizer treatments while traveling. You should bring whatever equipment you use at home with you for this. This also means that you will need to have the necessary tools to properly clean your nebulizer equipment. The place you’re staying may have a microwave so that you can disinfect your nebulizer parts between uses. If this is not an option, you could pack your own sterilizer device. Like your medicines, you should carry your nebulizer and other CF treatment equipment onto planes with you. This is to be sure that it doesn’t get lost or damaged in the checked luggage. If you’re traveling outside of the U.S., you will want to check with the manufacturer of your equipment to make sure that it will be able to function at your destination. If you’re traveling by car, look into getting a car adapter so that you can do nebulizer treatments on the road.

Lastly, having checklists readily available that are easy to edit as your child ages help ensure you keep track of all necessities. It also helps to add your needs to the list as parents can forget to take care of their own packing needs while focusing on their child’s daily medication and treatment needs on a trip. Due to the increased baggage you will likely be bringing on your trip, when flying, it is good to know medical equipment is free to check on any airline (for equipment approved to be stowed and that will not be readily needed). Additionally, 2 baby/child items – a stroller and a car seat – are free to check. If you use a stroller or a car seat bag, you can usually add additional items in the bag to check and still check them for free. This will help reduce checking costs. While strollers are convenient for moving about the terminal, they can also be bulky and hard to pack and unpack at the gate. Given the large number of carry-on baggage for medication and treatment needs, CF parents have found it easier to check as many items as possible. And, as size allows it, carrying a child in a body worn carrier can help free up hands for carry-on luggage.

Q: When should I call CF clinic?

It is important to learn your child's baseline symptoms with CF. Once you know how they are on a healthy, illness-free day you have a comparison point for when their symptoms start to worsen; however learning this about your child can take some time. If things appear to be worsening for your child's lungs, sinuses, stomach, or GI tract (all of which are affected by their CF), it is a good time to call the CF center. If it's a concern that doesn't relate to CF – for example, a skin rash due to poison oak - 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 on that 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 like their child having less energy than normal, increased mucus, nasal congestion, increased coughing, and signs of malabsorption 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: Help with insurance, payment for medications, financial hardship?

All of the medicines, office and hospital visits, and treatment equipment that CF requires can add up and become a financial hardship for many families. The CFF has a program called Compass that exists to help you find financial assistance. Compass does not have funds to disperse, but its case managers can help you find resources in your national, state, and local communities that may be able to help you cover the costs of the CF care your child is receiving. Compass can help you find grants or assistance that you are eligible for and can also give you instructions for how to get enrolled in a program.

In addition to finding ways to cover the medical costs of your child with CF, Compass can help find assistance for other life costs as well. They can work with you to find out how much you owe on things like monthly bills, and search for grants to help you cover those costs. They also can point you in the direction of local resources for discounted food programs, aid in affordable home repair, filing taxes, and transportation to and from appointments.

To find out how Compass can help you navigate your financial assistance options, call 844-COMPASS (844-266-7277), Monday through Friday, 9 a.m. until 7 p.m. ET, or email compass@cff.org.