Q: Parenting resources –

Parenting Children with Health Issues by Foster Cline and Lisa Greene. Parenting Teens with Love and Logic.

https://www.cff.org/CF-Community-Blog/Posts/2015/5-Ways-Parents-Can-Empower-Hope-in-CF-Teens/

Q: Information about Make-A-Wish?

Most individuals with Cystic Fibrosis qualify for Make-A-Wish. The criteria for wish eligibility has changed over the years and may continue to change. The CF social work clinic team works closely with Make-A-Wish. The social workers have a good understanding of when your child might qualify, based on Make-A-Wish guidelines. The social workers will discuss Make-A-Wish with your family when/if it is appropriate and will refer your child directly. The wish referral must be made before age 18 years of age but does not need to be completed before then..

Please note, there are specific limitations on wishes! The criteria have changed, as have the decisions that determine IF a child qualifies. MAW has its own panel of physicians who review the medical records and determine if a child qualifies (SL Based on medical status). So far, all children we have referred have qualified. We prefer to refer a child during high school (middle school at the earliest), because most preteens/teens present with increased difficulty managing their CF during these ages. They also wish for something very different at this age than at a younger age. Most patients who completed a wish at a very young age report they would have preferred to do a different wish as a teen.

Q: How often should I get a new nebulizer kit? How often should I get a new compressor? How do I go about obtaining these? Who should I contact if I have nebulizer issues?

When it comes to replacing nebulizer parts, you should follow the manufacturer's recommendations. The plastic parts of a nebulizer – such as the cup and tubing - can be broken down with use and decrease the effectiveness of the treatment.

A disposable neb cup will need to be replaced every week, and a non-disposable cup will need to be replaced every 6 months. The compressor is the machine that pushes air through your medicine to transform it into a mist that you breathe in. The compressor has a longer lifespan than the nebulizer kits and may be used until it stops working effectively. To extend the life of your compressor, change the filter in your nebulizer on the recommended schedule.

If the compressor is broken or replacement parts need to be ordered, you should first contact the Durable Medical Equipment (DME) provider that supplied the machine. If it is under warranty, usually 1-2 years, the DME provider should be able to repair or replace it and will provide a loaner machine in the meantime. However, often times a prescription from your physician is needed for it to be covered by insurance. In this case, you should contact the nurses in the CF clinic. If you're having an issue with your nebulizer compressor or with the nebulizer supplies, you should report it right away to help minimize missed treatments.

If you are interested in purchasing additional compressors or nebulizers that are not covered by your insurance, we can provide prescriptions for those products. Often families purchase addition supplies when they are going on vacation in places that it may be difficult to perform sterilization on a daily basis. Portable compressors are also available but are not recommended for long term use as they do not hold up well with extended use times and are not generally covered by insurance companies.

Q: Help with medication/pharmacy issues? Are there mail order pharmacies?

Many families with CF find that using a mail-order pharmacy can help simplify the process of obtaining medications. There are many mail-order pharmacies available for you to use but may depend on your insurance company. If you're interested in a mail-order pharmacy for your medications, talk with your CF care team pharmacist and they can help find a program that will work for you.

If you have questions about your medications or working with your pharmacy, call or message us through MyChart. Your CF care team pharmacist can also communicate directly with the pharmacy or your insurance company if you are having trouble filling any of your prescriptions. Prior authorization is often needed for CF specific medications and our care team can make sure all the necessary paperwork is done.

There are also many copay assistance programs and foundations that offer grant assistance for patients with CF. The pharmacist and pharmacy technician can help you enroll in these programs and make sure that these claims get submitted correctly. People on state insurance (Medi-Cal, CCS) are usually not eligible for most of the copay assistance programs.

Q: How do I know if my vest still fits?

Your vest is intended to be worn over top of a thin cotton t-shirt. The vest should cover most of your chest without being too snug under the armpits. The bottom of the vest should reach the top of the hip bones. There should be room for a flat hand (approx. 3/4 inch) to fit between the chest and the vest when you take a full inhalation. Your CF care team will check to make sure your vest fits properly by having you bring it into a clinic appointment once a year. You may also contact your vest provider to help with assessing the fit. To request a new size garment, please contact your vest provider directly. There should be a phone number on the equipment but if it has worn off, call or message the care team for assistance.

Q: What are 504 Plans and Individualized Education Plans (IEPs)?

Public schools that receive federal funding are required to provide education and services to children with disabilities under federal laws such as the Individuals with Disabilities Education Act (IDEA) and the Rehabilitation Act of 1973. Individualized Education Plans (IEP) exist within the Individuals with Disabilities Education Act (IDEA), and the Rehabilitation Act of 1973 contains Section 504 Plans. Both IEP's and 504 Plans can be very useful in helping your child with CF get the assistance that they need to be successful at school.

IEP's and 504 Plans are slightly different from one another, but some schools and school districts might treat them interchangeably. You will want to check with your school to see which one they will accept for

your child with CF. IEPs are used for adjusting a child's required work load to fit more reasonably with their capabilities given a health condition that has affected their learning abilities. Even though CF does not affect your child's cognitive abilities, their ability to learn can be affected if they have to miss a lot of school due to hospitalizations or sick days. The workload adjustments that an IEP covers can include less work assigned, extended deadlines, note-taking assistance, extra time for tests, etc. IEPs also can be used for advanced planning of hospitalizations to determine how children will get tutored during their absence from school.

Some schools require 504 Plans instead of IEPs because CF does not affect a child's cognitive abilities. 504 Plans differ from IEPs in that they address the physical aspects of your child navigating school each day. With CF, your child might need to do CF treatments or take medicines while at school. A little extra time getting to classes might be required, or they might need to use the bathroom more often than other children. To reach calorie goals, they might need to have snacks and water available to them throughout the day, or to have different high calorie choices at the school cafeteria. There may be days where they need to remain inside when the air quality is poor. All of these things, if addressed ahead of time in a 504 Plan, can help you advocate for the needs of your child.

To initiate your child's IEP or 504 Plan, you will need to make a request to the school in writing. Your CF social work team care team can help you identify some things that would be helpful to your child to request. A meeting will be scheduled with the school team and you and your child. Together with the school team, a plan will be developed to assure your child has accommodations in place so he/she can succeed at school. Your IEP or your 504 Plan will need to be renewed and revised each year. More information about IEP and 504 Plan can be found here: https://www.cff.org/Life-With-CF/Caring-for-a-Child-With-CF/Working-With-Your-Childs-School/Individualized-Education-504-Plans/

Private schools that do not receive federal funding are not obligated to follow the IDEA or the Rehabilitation Act of 1973. If you are interested in a private school, you will want to see that they are willing and able to meet your child's needs before agreeing to attend.

Q: How will my child keep up with school work if they are admitted to hospital?

If your child has to be hospitalized due to CF, keeping up with their school work can be a challenge. When you learn that your child will need to go to the hospital, ask your doctor to estimate how long the stay will be and share this information with your child's teachers. A typical admission lasts for 2 weeks. Encouraging your child to be involved in the communication with their teachers can help them take ownership of the work they will need be doing while in the hospital. Your child might require a reduced work load or extended due dates for their assignments. Agreeing on a plan for this with your teachers is an important first step.

While you're in the hospital, keep your teachers updated about how the work is coming along. Don't be afraid to let them know if your child's progress on school work has been slower than was originally planned. Teachers are more likely to be understanding if they know ahead of time that a deadline is not going to be met.

Once you've agreed on a schedule for deadlines and due dates, write them on a calendar that your child can access easily. Start working on study skills with your child by helping them plan ahead for big projects and tests. Teach them how to schedule a realistic amount of time beforehand to prepare for assignments. It also helps to break down assignments or homework into small portions of dedicated time over the course of several days (e.g. 30 minutes a day for current work and 30 minutes a day for makeup work). Check with your child's school teacher and school district to determine what/if school support services are available when your child is in the hospital. A 504 School Plan can be helpful to assure that your child receives additional time to complete assignments and tests during hospital admissions. The CF clinic social work team can explain typical 504 accommodations, and how to request a 504 Plan for your child.

https://www.cff.org/Life-With-CF/Caring-for-a-Child-With-CF/Working-With-Your-Childs-School/Individualized-Education-504-Plans/

https://www.cff.org/Life-With-CF/Caring-for-a-Child-With-CF/Working-With-Your-Childs-School/When-There-s-More-Than-One-Person-With-CF-in-the-Same-School/

Q: What do I do if my child doesn't like taking enzymes or coughing in front of their friends?

Having a chronic disease like CF requires a lot of maintenance and can set your child apart from their friends. It can be especially challenging in certain developmental stages, such as middle school. Some kids decide that they would prefer to keep their CF diagnosis a secret from their friends at school. While having to do CF treatments can make your child feel different, the treatment plan needs to be followed for your child to stay healthy. This is an opportunity for you to help your child accept CF as part of who they are. You can encourage them to take steps toward acceptance by guiding them on how to talk about CF to their friends and school mates. This doesn't mean that your child has to share the information of their CF diagnosis with everyone. Help them decide who to tell about their CF, and work with them to come up with some simple answers they can give if friends have questions. You can also explain to your child that skipping enzyme doses or holding their cough will make them sick more often. Help them learn to view CF medications and treatments as helpful therapies that will make them feel well enough to have time for friends and fun activities.

Q: What if my child doesn't want to use the bathroom at school?

Children with CF who have pancreatic insufficiency are not able to absorb nutrients well. As a result, children with CF may have more frequent stools that can be softer and have a stronger smell. It can also cause increased gas and bloating. This could potentially cause an embarrassing situation for a kid who is shy about using the restroom in public. If your child is experiencing this, check with the school to see if there is a private restroom your child could use. Many schools have a bathroom in the nurse's office or teachers' lounge that your child could use to help alleviate stress regarding bathroom visits.

The school might not have any accommodations to offer. In this case, help your child become more comfortable with talking about their bowel habits. By using a matter-of-fact tone and medical terms instead of common ones, you can help your child normalize this for your child. Encourage your child to talk about their concerns with the CF team.

Q: What do I do if my child has concerns about getting fat?

As children grow up, they naturally become more aware of their bodies and appearances. They might encounter pressure from peers or the media to look or eat a certain way. They also might learn about the dangers of eating unhealthily in school or health class. These influences, combined with the CF requirement that their diet be high in calories and fat, can be very confusing for a child at this age. Further education about the topic can be a big help in getting your child on board with their CF health plan.

Use this opportunity to check in on your child's understanding of why they have to eat a high-calorie, high-protein diet. Make sure they understand that it's because their bodies don't break down fat and protein as well as they should. Even with the help of the enzymes they take, some food does not get digested. Your child's body also requires more energy for them to be able to fight off lung infections. Being at a healthy weight can help them to continue living their lives as fully as possible. Additionally, puberty can be delayed if a child does not weigh enough or does not have enough body fat. This can be a powerful motivator for your child to eat well because most children want to develop at the appropriate time and along with their peers.

Q. What do I do if my child is overweight?

We are seeing a rise in obesity in all kids in the US, even those with CF. Increasing activity and working on portion sizes are the recommendations for this health issue. Being overweight can put CF kids at increased risk for diabetes and lung exacerbations. There are resources at Rady and in the community for weight management.

Q: Why do I get asked about depression and anxiety?

Having a chronic illness like CF can add responsibility, worry and stress to your child's daily life. This can impact the way your child feels from day to day and as well as overall. It is important to check in with your child to see if these stressors are causing depression and/or anxiety.

To help keep you or your child safe and healthy, your CF care team will periodically ask you questions about depression and anxiety. The goal of this is to identify people who have mild, moderate or severe symptoms of depression or anxiety, and help connect them to appropriate services so they can feel better and have a better quality of life. Depression and anxiety can also make a person less likely to stick to their CF treatment plan, causing a negative impact on their health. The good news is that depression and anxiety can be treated. Once a person gets the help they need to feel better emotionally, they are more likely to have the energy to give to their life and CF treatments.

https://www.cff.org/Life-With-CF/Daily-Life/Emotional-Wellness/Depression-and-CF/

https://www.cff.org/Life-With-CF/Daily-Life/Emotional-Wellness/Substance-Misuse/

https://www.cff.org/Life-With-CF/Daily-Life/Emotional-Wellness/Anxiety-and-CF/

Adolescent symptoms of depression can present in a variety of ways. The most common symptom of depression in children (and adults) is *irritability*. Despite popular media portrayals of overwhelming sadness, the most regularly described symptom of depression in youth is chronic irritable mood, including increased anger and verbal aggression. While irritability and mood changes are common in teens and preteens, there are also noticeable changes in daily functioning that can signal their mood swings reflect more than just normal adolescent mood fluctuations. The following observed behaviors in addition to recent changes in your child's functioning (lower grades in school, recent change/decrease in hygiene, dropping out of activities, etc.) would warrant a consultation with a mental health provider and your CF team. Symptoms to look for are:

- Loss of enjoyment in activities, especially activities they used to love
- Less social engagement with friends or family members or isolating for increasing amounts of time
- Expressing feelings related to worthlessness or hopelessness such as "This is never going to get better," "This always happens to me," "I wish this would all just go away," or comments expressing not wanting to be a bother or a burden to others
- Expressing guilt, such as feeling that negative events are "their fault" or that they "always mess things up"
- Changes in sleeping and eating patterns
- Recent difficulties with attention and concentration
- Increased thoughts of wanting to go to sleep and never wake up, death, or "wanting it all to go away"
- Decreased energy or moving more slowly than usual

Adolescent anxiety symptoms are also highly varied. There are many types of anxiety that can impact children and adults alike. Most children and teens report anxiety surrounding everyday worries and social situations. While there are several anxiety symptoms that can be observed (panic attack, skin flushing, etc.); the majority of anxiety symptoms are less obvious. As with all mental health symptoms the following observed behaviors in addition to recent changes in your child's functioning (lower grades in school, recent change/decrease in hygiene, dropping out of activities, etc.) would warrant a consultation with a mental health provider and your CF team. Symptoms to look for are:

- Recent increase in physical symptoms that have no medical correlation or cause
- Increased embarrassment in situation that used to be more comfortable
- Avoiding specific places, situations or crowds
- Checking in with parent more frequently about past and future events, such as "Did I do that okay?" or frequently asking about upcoming plans or plan changes
- Increased fears or phobias not observed before
- Expressing worry they are "losing it"
- Restlessness or increased irritability
- Easily exhausted or tired and not associated with physical activity
- Child appears preoccupied more than usual and often "goes blank" or "zones out"
- Changes in sleeping and eating patterns
- Muscle tension

- Child appearing shyer than before

Mental health issues for kids and adults have been worsened by the COVID-19 pandemic and are likely to persist even as things "return to normal". Please reach out to the CF team or your primary care doctor if you have any of the symptoms listed above.

Q: Can I meet other kids with CF?

As we have learned, some germs – such as Pseudomonas aeruginosa and Burkholderia cepacia have a higher likelihood of affecting people with CF. The infections that these germs cause can be spread between people with CF while not posing a risk to other individuals. Researchers have found that when a person coughs, droplets from the cough can travel as far as 6 feet. For this reason, there are guidelines regarding contact between people with CF. The CFF recommends that people with CF remain at least 6 feet (2 meters) apart from each other at all times and that contact be limited to outdoors environments. These guidelines do not apply to family members with CF.

These safety guidelines mean that certain activities involving more than one person with CF should be avoided. Close-contact activities such as hanging out with another person with CF, being in the same classroom, sharing hotel rooms, car rides, or handshakes can contribute to the spread of germs and an increase in CF exacerbations. Similarly, things like summer camps that are specifically for CF people are not safe and are not recommended by the Cystic Fibrosis Foundation. .

Despite these guidelines, forming relationships with other people who have CF can have a very positive impact. If you or your family knows another person with CF that you would like to get to know, consider using computers or phones for messaging and video chatting.

The CF Foundation has developed a program called Teen Connections for CF teens aged 13 to 18 who are searching for a way to virtually connect with others their age. The program uses methods of communication including online chats and video calls, and each event will be facilitated by an adult who has CF and a member of the CF Foundation staff. If you think your child might benefit from being a part of this community, visit cff.org/teensignup to help them register. Parental consent is required for all participants under the age of 18. There are separate groups for younger teens and older teens. Contact Aimee Jeffrey at ajeffrey@cff.org with any questions

Q: When should my child start taking over their treatments?

The goal with CF care is that your child will grow into a young adult who is able to manage their CF once they leave home; however, becoming independent doesn't happen overnight. You want to gradually start handing off CF care tasks as soon as you think your child is mature enough to handle them. The CF social work team will discuss appropriate developmental CF goals for your child, and will help you and your child determine which tasks to focus on for success. Around the time that your child starts having other responsibilities around the house – such as helping in the kitchen or doing household chores - they are old enough to start taking on some CF care tasks. When your child is in middle school, the CF team would expect that they would be able to answer some questions about their CF, name their medications and the reason for taking them, be independent with airway clearance, and take their enzymes independently.

Q: How often should I clean my respiratory equipment? Do I have to clean my Aerobika (OPEP device)?

The four steps to maintaining respiratory equipment are clean, disinfect, rinse, and air dry. Reusable nebulizers, Positive Expiratory Pressure (PEP), and Oscillatory Positive Expiratory Pressure (OPEP) devices should be cleaned after each use as small amounts of mucus and phlegm can be left behind. This could cause germs in the saliva or mucus to be breathed in during the next treatment.

Clean

Clean your hands first to avoid introducing any new germs to the equipment. Use dish soap and hot water – tap water or well water are suitable - to clean inside and outside all of the parts of the cup, mouthpiece or mask. This needs to be done after each treatment. Let the equipment air dry fully before reusing.

Disinfect

There are different options for disinfection involving both heat and cold methods. Check with the manufacturer of your equipment to learn what the suggested method of disinfection is. You should disinfect equipment once a day.

<u>To disinfect with heat:</u>

Tap water or well water are acceptable to use for disinfection options that involve heat.

Option 1) Boil water on the stove. Once the water is boiling, put your nebulizer parts in it and continue to boil for 5 minutes.

Option 2) Put the nebulizer parts in a microwavable-safe bowl and add enough water to completely cover the parts. Put this in the microwave and microwave for 5 minutes.

Option 3) If your dishwasher gets to 158 degrees F / 70 degrees C for at least 30 minutes, you can put the nebulizer parts in the dishwasher

Option 4) Electric steam sterilizer (commonly used for baby bottles)

<u>To disinfect with cold:</u>

Option 1) Soak the parts in 70% isopropyl alcohol for 5 minutes

Option 2) Soak the parts in 3% hydrogen peroxide for 30 minutes

Rinse

If you used a heat method for disinfection, there is no need for a rinse step.

If you used a cold method for disinfection (isopropyl alcohol or hydrogen peroxide), you must rinse all of the nebulizer parts with sterile water. To make sterile water, boil water on the stove for 5 minutes. Each

time you need to rinse with sterile water, you should freshly boil a new pot to avoid any contamination. Bottled water and tap water can all contain germs and should not be used in place of sterile water. Purchased distilled water can also be used for the rinse step.

Air Dry

To air dry the nebulizer parts, place them on clean, dry paper towels. The drying step is extremely important, because any environment that has water is a place where germs can grow. Once all of the parts are dry, store them in a clean container that will keep them dry until their next use.

Additional information: Vinegar (acetic acid), bleach, and benzalkonium chloride are not recommended for use in cleaning or disinfecting equipment, because they are not effective enough at killing all of the CF pathogens.