

CARING FOR YOUR INFANT

a parent's guide to healthy living with cystic fibrosis

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WHERE'S THE INSTRUCTION MANUAL FOR THIS KID?

Our child is born. We are excited and joyful. Then we get the diagnosis. We grieve. We might feel confused, overwhelmed, depressed or angry. Some of us reach out for support; others choose to be more private. Some devour information about cystic fibrosis and others choose not to.

Our reaction depends on our personality traits, background, and culture. Eventually, we reach a point of acceptance. This doesn't mean we are happy with the situation; it just means that we have come to terms with our new reality and have decided to pick ourselves up and move forward.

This also doesn't mean that we won't sometimes be overwhelmed by emotions. Life has its ebbs, flows and even some whirlpools that threaten to suck us under. But simply going with the current often gets us to calmer waters. The routines of daily life have a way of taking over.

Depending on the severity of your child's CF, you are managing doctors' visits, hospitalizations, insurance issues and multiple medications. You also have other responsibilities at home and work. But, with some organization and cooperation, family life can become enjoyable even with all of the demands that raising a child with CF can bring. People and families are amazingly adaptable.

The problem is that it doesn't take much to throw sand into the works and bring it all to a grinding halt: a long-term hospitalization, surgery, or adding another medical requirement to an already packed schedule. Life can get pretty stressful. With this extra stress comes more frustration! Kids pick up on stress and frustration and choose the worst moment to push our buttons.

We parents can easily feel overwhelmed and wonder, "Where's the instruction manual for this kid? What do I do *now*?" When a child has CF, this question is harder to answer without some outside guidance. Love for our child, coupled with fear, guilt and worry can cause us to act in ways that are more likely to put our child at risk including babying our child and being over-protective.

It helps to remember the ultimate goal of parenting: to *prepare our kids for the real world*. When we refer to a child "at risk," we mean a child who is not capable of functioning in the real world with all of its choices and consequences. And when a child has CF, their choices and consequences can have life-altering results, so they need to learn responsibility at a younger age than other kids.

There are no guarantees that our kids will always make the best decisions. However, using effective communication skills will increase the odds of raising confident, responsible kids with excellent coping skills; kids who take good care of themselves even when no one else is watching.

Wise parents realize that the process of transitioning their child to adulthood starts when their child is old enough to throw peas from the high chair!

Preparing our children for the real world is the biggest job we will face. That's what parenting is all about: getting our children ready to leave home, to live independently and to know they are not at risk. We want them to look at the real world and say:

"I recognize this world! We practiced for it at home!"

Excerpt from the booklet: *Parenting Children with Health Issues and Other Special Needs: Love and Logic Essentials for Raising Happy, Healthier Kids* by Foster W. Cline, MD and Lisa C. Greene. www.ParentingChildrenWithHealthIssues.com.



AGES AND STAGES: THE EARLY YEARS

By Lisa C. Greene - mom of two kids with CF and parent educator and Foster W. Cline, MD - child psychiatrist and co-founder of Love and Logic

If sleepless nights and dirty diapers has you down, there's good news! Children change quickly. As you might already know, each age and stage has its very own joys and challenges. The trick is to maximize the joy and minimize the challenges. And, with a handful of effective parenting skills and a basic understanding of child development, you'll find it much easier to enjoy your kids at all stages of life- including the teen years.

Let's start with the early years. The first six months are critical for bonding and attachment. Attachment takes place when an infant's needs for food, love, and comfort are consistently met. Each time a loving caregiver meets the child's needs, the child develops trust: "Whenever I need something, there is someone there for me." And the cycle goes around and around thousands of times in an infant's life.

Building a deep bond with your infant will help lay the foundation for good emotional and physical health in the years to come. Ways to bond with your baby include holding, rocking, talking, singing, eye-contact, smiles, touch, play and meeting needs for food, cleanliness, comfort, and medical requirements.

Contrary to what you might read in some parenting books, you can't spoil an infant. Letting an infant "cry it out" isn't good advice, especially for babies with medical issues. In the case of CF, pancreatic insufficiency and malabsorption can cause discomfort so it's important to do what you can to comfort your baby. When my (Lisa's) son was a baby, he had a lot of tummy aches from both food allergies and CF. I found that rocking, holding him upright, and walking around with him really helped.

When babies and toddlers have CF, we have to get very creative about getting the pancreatic enzymes down the old hatch. Be sure the enzyme beads aren't sitting in the folds of your baby's skin under the neck or in the diaper area. They can cause a painful rash. Also watch for severe diaper rashes that can be common with CF.

We also have to get creative about how to effectively handle breathing treatments and chest physical therapy. Getting ideas from your CF Team as well as other parents is helpful both to learn proper techniques and to know you are not alone.

Caring for your baby can be exhausting so you'll need support from others. It's also important that you take good care of yourself. Infants mirror the parent's emotions: when daddy smiles, baby smiles. So if you're happy, odds are good your baby will be, too.

Research shows that the parents' emotional state is vitally important. Children of depressed mothers have been shown to end up with more behavioral and developmental problems than non-depressed moms. And, when a baby is diagnosed with CF, many moms become depressed. If you are depressed for more than a month, it is important to get help. Your CF clinic has people who are trained to help you with this. Hang in there- things will get easier as your baby gets older.

As babies get older, they start to have both needs and wants. Maybe your baby is crying in her bed at night and you can't tell whether she really needs you because she has a tummy ache or just wants you to pick her up because she doesn't want to miss any of the action!

When a child is younger than one year of age, we suggest that if you can't distinguish between a need or a want, then meet the need. It is much easier to "course correct" a spoiled toddler than one who has attachment issues.

That being said, letting your baby cry for a few minutes every now and then is not going to cause a problem. It's chronically unmet needs that can cause challenges in an infant's development. So if you are responding to your infant's cries and giving food, love and comfort most of the time, then you are building a strong foundation that will help in the stages ahead.

ADVICE FROM OTHER PARENTS

Insight and Encouragement From Parents Who Have Been There



Other parents of children with CF can help you put together the pieces of the CF puzzle. They are always willing to share what they know, so you can master all that comes along with this stage - especially the challenges that come with caring for your child's CF. However, it's important to remember that what works for some, doesn't work for everyone. Here are some experiences, tips, and encouraging words from other CF parents on Chest Physical Therapy, breathing treatments, and pancreatic enzymes:

Chest Physical Therapy

Chest Physical Therapy, CPT for short, is often started in infancy. Most start with manual CPT, a steady "thumping" of the hands on the chest, back and sides. Occasionally, a medical vest system will be used, but there are conflicting opinions in the medical community about the safety and effectiveness of these systems for infants. CPT can be overwhelming for both babies and parents at first, but can become a very enjoyable time; full of bonding and games. Here's what other parents have to say about CPT:

We do manual CPT while listening to my son's favorite nursery rhymes and songs. We play the songs on a CD and sing along as we keep the beat. We love "nursery" time at our house! - *Ryan, dad of 2 year old with cystic fibrosis.*

At first, my daughter cried during CPT. But now, she finds the rhythmic thumping soothing and often can't fall asleep without a steady tap on her back. So hang in there, CPT does get easier. - *Vince, dad of 18 month old with cystic fibrosis.*

CPT is a great bonding experience for parents and children. While it may seem like a daily battle now, it will bring you and your child closer. I'm positive it's a big factor in why my son and I have the relationship we do today. - *Chris, mom of 30 year old with cystic fibrosis.*

Your arms may get tired when you first start doing CPT - mine sure did! You can avoid some of the fatigue by lessening your arm movement and using more just wrist movement. It makes the treatment a lot easier! - *Amber, mom of 15 month old with cystic fibrosis.*

I often squeeze in 'extra' CPT, since I can do it anywhere. I start patting my daughter's back while riding in the car; walking down the street; laying on the couch; you name it. A little 'extra' every day can go a long way. - *Mary, mom of 2 year old with cystic fibrosis.*

Talk with your CF Team about the best way to do CPT on your baby. The CF Foundation guidelines show only "upright" infant CPT (baby sits upright on a caregiver's lap). For details, visit www.CFF.org under Therapies/ Clearing Airways/ Postural Drainage. - *Lisa, mom of two kids with cystic fibrosis.*

Breathing Treatments

Breathing treatments are done by inhaling medicines using a nebulizer compressor. For infants, medicines are typically administered through a mask attached to the nebulizer cup. The length of breathing treatments varies based on the number and type of medications, as well as the nebulizer compressor being used. Here's how other parents do breathing treatments with their children and what they do to make them fun:

We turn them into games. We'll count how long he can inhale or the number of breaths he takes; or we'll put on an extra mask and pretend we're snorkeling or scuba diving. - *Adam, dad of 18 month old with cystic fibrosis.*

We've found the trick is to keep our daughter entertained with different activities every few minutes. I always have a basket of toys next to us so I can hand her different toys when she starts losing interest. - *Jessica, mom of 1 year old with cystic fibrosis.*

We carefully time our breathing treatments. We wait until our son is winding down and getting sleepy, but not to the point of being cranky. We make sure he's fed, changed and happy. It takes a little practice to get the timing just right, but you know your baby's mood and patterns. You'll get it down. - *Courtnie, mom of 2 year old with cystic fibrosis.*

I've tried several different methods to secure the mask on my daughter's face. Some days the elastic head strap does the trick. Others, I hold it by hand. But it's important to know that there will be times that you'll see a little mist escaping from the mask. Just do the best you can! - *Sara, mom of 10 month old with cystic fibrosis.*

Start the process with fun and love, singing, reading a book, playing with a baby toy. Create a calm, pleasant routine- just like you do at bedtime. Ease into the treatment gently. Put the mask on your baby for a few minutes each day without the nebulizer running to get him or her used to it. If it's okay with your doctor, start with short treatments and then build up to the full time slowly. - *Lisa, mom of two kids with cystic fibrosis.*

Pancreatic Enzymes

Pancreatic enzymes, also referred to as digestive enzymes or, simply, enzymes, are taken by individuals with CF to help absorb nutrients in the foods they are eating. It is not uncommon to start enzyme replacement therapy in infancy. Often parents of infants and toddlers administer enzymes by opening the capsules and mixing the enzyme beads with applesauce, but each parent has to find what works for their child. All it takes is a little practice and creativity to master the art of getting those enzymes down the hatch. Here are a few pointers from other parents:

Our son started refusing enzymes with applesauce, but we found that you can put the beads in any soft, acidic food that isn't milk-based. As soon as we started switching it up, he started taking them like a champ. - *Sue, mom of 1 year old with cystic fibrosis.*

When our son was an infant, we administered the enzyme/applesauce mixture with a medical syringe. We'd open the capsules, pour the microspheres into applesauce, mix, and scoop the mixture into the syringe. Be sure to cut off the tip of the syringe so the enzyme/applesauce mixture won't clog it. - *Tricia, mom of 8 year old with cystic fibrosis.*

At around one year old, my daughter became fascinated with her enzymes, so I would give her empty ones to play with. By 14 months she was swallowing them whole and now at 2 years old she takes all 7 at once! - *Kacie, mom of 2 year old with cystic fibrosis.*

Newborns have a tongue thrust reflex which protects them from choking. Enzymes may pop back out of your baby's mouth! Try dipping a clean finger into applesauce, then dip into the enzymes, and place towards the side back of your baby's tongue (without gagging!). - *Julie, mom of 3 year old with CF*

When our son was very young, he would spit the enzyme/applesauce mixture out. Our nutritionist told us to just scoop the mixture back into his mouth until he would finally swallow it. He eventually got used to the new texture. - *Tracy, mom of 3 year old with cystic fibrosis.*

I had trouble remembering his enzymes at first, so we started stashing them everywhere. We have a bottle in the kitchen, diaper bag, my purse and at his grandparent's house. The applesauce hasn't been as easy, but I make sure our fridge is stocked and we always have a few containers in the diaper bag. - *Rachel, mom of 2 year old with cystic fibrosis.*

NUTRITION IN THE FIRST YEAR

Nutrition and CF

Normal growth and development are important goals for your child throughout his or her childhood. Excellent growth and good nutritional habits from the very beginning are extremely beneficial in CF. As you will learn, cystic fibrosis can affect nutrition in many ways. The recommended diet is high in calories and protein with liberal amounts of fat and salt. This “Dream Diet” is contrary to the health recommendations for the general public! Your CF clinic will incorporate growth and nutrition monitoring at every visit, and nutritionists are available at many clinics for each visit.

Formula or Breast Milk?

Breast milk is excellent nutrition for babies with CF and we encourage families who wish to breast-feed. Commercial infant formula can also support good growth and nutrition. Whether your baby is breast-fed or bottle fed, the CF team will ask about his or her feeding pattern, stool habits, and sleep and nap schedules as part of their overall nutrition assessment.

Pancreatic Status: Pancreatic Insufficiency versus Pancreatic Sufficiency

By far, most individuals with CF are “pancreatic insufficient”. This means that the digestive enzymes that are made by the pancreas are not able to help with digestion. If untreated, this would result in malnutrition. Fortunately, these individuals can take digestive enzymes in the form of a medication that is commonly referred to as simply “enzymes”.

A stool elastase test can tell us if your child’s pancreatic insufficient or pancreatic sufficient. This requires a quarter-sized sample of stool; results are known in about 7-10 days. The test may be repeated later in infancy if the first test was borderline or normal, as pancreatic function can change over time.

Pancreatic Enzyme Replacement Therapy, also known as “enzymes”

There are several brands of prescription enzymes used to treat pancreatic insufficiency. At the present time these include Creon, Zenpep, and Pancreaze. Never accept a generic substitution for name brand enzymes because they don’t work as well. Enzymes come as capsules and are available in different strengths. The dose depends on many factors and will be determined by your care team. Inside the capsules are the digestive enzymes in the form of tiny beads. Each bead is coated to protect the enzymes from being destroyed by the strong acid in the stomach. The beads are designed to release their activity in the small intestine where digestion and absorption occur.

When the enzymes are in contact with formula, breast milk or food in the body, they start to break down the protein and fat that were eaten. Enzymes should be given at the start of each formula feeding or breastfeeding session, so they can travel with the food as it passes through the intestine. For this reason, enzymes are given every time your baby eats. Because infant formula and breast milk contain a lot of fat, your baby will need to have enzymes with it in order to benefit from it.

Giving enzymes to a newborn baby: Open the capsule and pour the beads onto a small amount of an acidic food, such as baby applesauce, and immediately feed it to your baby on a spoon, at the start of every bottle feeding or nursing session. Even newborns can safely take enzymes this way. Your CF team will provide you with additional guidance and instruction.

Additional tips for using enzymes: Don’t allow the beads to sit in the applesauce – exposure to the moisture will start to activate the enzymes too early. Also, take care to store your enzymes at room temperature.

CF Vitamin Therapy

Individuals with CF have trouble absorbing the fat-soluble vitamins A, D, E and K. To prevent vitamin deficiencies, special vitamins designed for CF are prescribed (AquADEK®, SourceCF® or Vitamax®). Because enzymes help the body absorb these vitamins, it is best to give vitamins at the time of a meal or snack when enzymes are also given.

Salt

People with CF lose more salt through sweat than people without CF. Salt depletion can lead to serious health problems. Your nutritionist will recommend adding a specific amount of table salt to your baby's diet every day to prevent this. The salt is usually added to formula; for breast fed babies, it can be added to the applesauce given with the enzymes. Liberal salt intake is encouraged through life and it should be added to baby foods and table foods as your baby gets older. All people with CF should increase their salt intake during the summer months and when sweating from exercise.

Fluids

Generally, babies that drink enough breast milk or formula to grow well are getting enough fluid. For most infants with CF, we do not recommend routinely giving water or juice because the nutrition content is poor. Occasionally, there are situations that require more attention to fluid intake, such as vacationing in hot climates. Your nutritionist will guide you.

Introducing Solid Foods

For a baby with CF, the guidelines for starting baby food and table food may sometimes be different from the child without CF. In general, babies without health concerns are ready to start solids between 4-6 months of age; soft table foods between 6-8 months; and whole cow's milk at 12 months. The timing for babies with CF is individualized. The factors to consider are your baby's growth, appetite and interest in eating. While your pediatrician may talk to you about normal infant feeding stages, your CF nutritionist will guide you on the timing of solids, enzyme and vitamin dose adjustments, and high calorie strategies as needed.

Reaching Nutrition Goals

Calorie and protein needs are generally higher in those with CF than the non-CF population. Despite this, many babies grow well with regular infant feeding practices and proper dosing of enzymes. If a baby has trouble gaining weight, there are many strategies you can try with the guidance of your CF Team. These include:

- Add more calories to formula, breast milk, baby food or table food.
- Fine-tune enzymes and absorption.
- Adjust times of feedings, meals and/or snacks.
- Review common causes of slow weight gain.

Sometimes, even with the best nutrition plan, there can still be a large "calorie gap". That is, the calories consumed are not enough to match the calories used, and growth slows. In this case, tubefeeding can be used to supplement the food a child eats. Tubefeeding is a method of delivering liquid nutrition through a tube directly into the stomach. In CF care, tubefeeding is not a "last resort" treatment, nor a sign of failure to feed a child well. Instead, by supporting robust growth, it is one of many "tools" available to fight the disease.

Living the Dream Diet, not the Heart Healthy Diet

Dietary fat is a concentrated source of calories and it plays a very important role in CF nutrition. It may be hard to get used to the idea of feeding your child a diet that is high in fat and calories when other parents are focused on avoiding childhood obesity and heart disease! You may find it necessary to explain to family members, friends and other caregivers that restricting fat intake can actually be harmful to your baby. For individuals with CF of all ages, we encourage full fat yogurts and ice creams, whole milk or even half and half, and generous amounts of butter or margarine, just to name a few!

HEALTH MAINTENANCE RECOMMENDATIONS

Regular clinic visits:

Frequent visits with the CF Team when your infant is young are needed for close monitoring of growth and nutrition. These visits also allow time for you to learn all the different aspects of care involved in keeping your infant healthy. Anticipate that visits will be scheduled based on your infant's needs and your clinic's policy. Here is a rough guideline for your reference:

- Every 2-4 weeks until about 3 months of age.
- Every 1-2 months until about one year of age.
- Every 2-3 months from one year of age forward.

Laboratory Evaluations:

While we know having blood drawn can be stressful for both infants and parents, there are certain tests that are important to do to help your CF Team best care for your child. These are usually done between 3 and 6 months of age and routinely every year as determined by your CF Center's guidelines.

•Blood tests:

•**Vitamin levels:** Fat soluble vitamins can be poorly absorbed in babies with CF, but are very important for infant development. Vitamin A, E and D levels will be checked. Vitamin K is also a fat soluble vitamin but there is not a test for Vitamin K levels.

•**Kidney function:** Many medications are processed in the kidneys. In order to make sure the kidneys are working well and can process any medications that your child is taking, kidney function should be checked.

•**Liver function:** The liver can have a back-up of thick secretions that can affect how well it works. Many medications are also processed in the liver, so it's important to check liver function.

•**Nutrition tests:** Your CF Team will monitor your child's growth closely but there are also blood tests to check that your child has enough protein to build muscle and grow normally.

•**Blood counts:** Red blood cells are checked because children with CF can have anemia (not enough red blood cells to carry oxygen). White blood cells are checked because a change in them can mean infections. Platelets are also checked as a marker of inflammation and to see how well your child's blood clots.

•Stool studies:

•**Fecal Elastase:** This checks to see how well the pancreas is working to absorb nutrients. Most infants with CF do not have the ability to absorb all nutrients because their pancreas does not work correctly. Usually this happens shortly after birth. However, some children do not lose the ability of their pancreas to work correctly until later in the first year of life. Depending on your CF Center, this test will be done when your child is diagnosed and repeated again in the first year of life if it is normal the first time it is checked.

Evaluations of Lung Health:

•**Respiratory cultures:** Children with CF can have many different bacteria (germs) in their airways. It is not easy to get a culture from the lungs so a cotton throat swab is used to see what bacteria might be in the airways. It is important to know what bacteria are in your child's airways so they can get the right medications when they are sick. Respiratory cultures are repeated every 3 months and/or with any new respiratory symptoms.

•**Pulmonary Function Testing (PFTs):** This is a test of lung volume and function. A special machine is used for infants. Typically, the first test is done at 4 months of age and then repeated every 6 months depending on your CF Center's guidelines and equipment availability. Some centers do not do infant testing.

•**Chest X-ray:** An x-ray of your child's lungs are generally taken in the first year of life. This helps your CF Team monitor any changes in the lungs over time.

This information was kindly provided by the Division of Pulmonary Medicine, Children's Memorial Medical Center in Chicago. It is based on the Current Clinical Practice Guidelines as established by the CF Foundation for accredited CF care centers as of 2010 and is subject to revision. Check with your CF Team for the most current guidelines.

ADDITIONAL RESOURCES

Valuable Online Resources for Information and Encouragement

There are many useful and encouraging web sites that you can visit to get information about cystic fibrosis and living with CF. These are sites that we recommend you visit to find information, comfort and friendship. Be sure to check out any medical advice with your doctors *before* putting it into practice.

www.CysticLife.org - A social network providing the CF community with peer-generated tips, information and encouragement with access to blogs, forums, reviews and user profiles, as well as the ability to directly communicate with other members.

www.ParentingChildrenWithHealthIssues.com - Discover the skills to raise happy, healthier children with CF. Free video, audio, and other resources provide effective, practical tools to empower, motivate and communicate with children of all ages. Based on the popular Love and Logic ® parenting program (www.loveandlogic.com).

www.TipsForCFParents.com - Provides tips and resources for parents of children with CF. Lisa C. Greene is the mom of two kids with CF, a parent coach and co- author with Foster Cline, MD of the award-winning *Parenting Children with Health Issues*.

www.BloomingRoseFoundation.org - The Foundations' mission is to provide social services, positive contacts and hope to families immediately after diagnosis.

www.cff.org - The Cystic Fibrosis Foundation (CFF) provides information about living with CF, treatments and research. The CFF assures the development of the means to cure and to control CF and improve patient's quality of life.

www.CFLiving.com - An educational site that offers information and support for those living with or caring for someone with cystic fibrosis. It is designed to help you work more closely with your Care Team, learn about treatment options, and provide interactive educational resources.

www.cfri.org - CFRI's mission is to fund research, provide educational and personal support, and spread awareness of cystic fibrosis.

www.CFvoice.com - An online community for those living with cystic fibrosis. A place for motivation, inspiration and connection to the CF community.

www.CysticFibrosis.com - An online community offering tools and resources for Cystic Fibrosis.

www.cystic-L.org - A free email service dedicated to the exchange of information and support specific to cystic fibrosis.

www.Esiason.org - The Boomer Esiason Foundation is a dynamic partnership of leaders in the medical and business communities joining with volunteers to heighten awareness, education and the quality of life for those affected by CF.

www.Facebook.com - This internationally known social network is used by many CF-related organizations and community members to share information and encouragement.



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