Parents' Guide To Cystic Fibrosis (CF)

California Department of Public Health
Genetic Disease Screening Program
Newborn Screening Branch
www.cdph.ca.gov/nbs

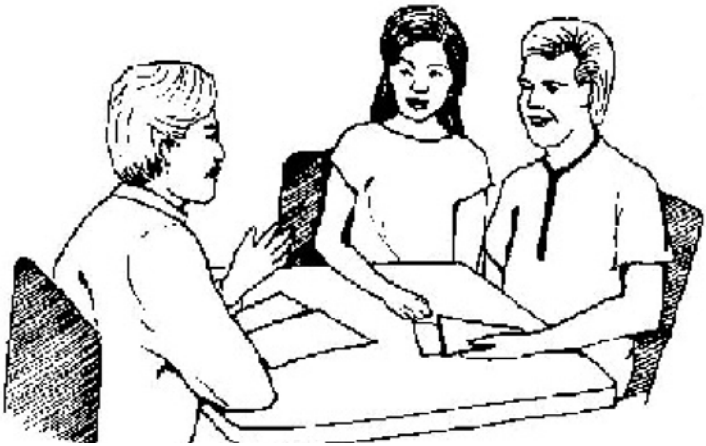
To Parents:

California State Law requires that all babies have the newborn screening test before leaving their hospital of birth. A few drops of blood were taken from your baby's heel. One of the tests was for cystic fibrosis.

Your health care provider may have told you your baby has cystic fibrosis (CF). Babies can look healthy at birth and still have this disorder, but babies, who are not treated, often have serious and permanent health problems. With early and ongoing care, your baby should develop normally both physically and mentally.

This booklet was written to help parents learn more about this disorder. Use this booklet to learn how to care for your child.

For more information about the Newborn Screening Program or CF, visit our website at www.cdph.ca.gov/nbs.



Discuss this booklet with your doctor.

What Is CF?

CF stands for cystic fibrosis. It is an inherited disease that can affect many body organs including the lungs and the digestive system. Babies born with this disease do not have a fully functional chloride channel in the cell membranes of many body organs. This disease is genetic so your child cannot outgrow it.

What Causes CF?

A special protein called "cystic fibrosis transmembrane conductance regulator" (CFTR) is not working well or is missing. People with CF have too much chloride in their sweat and lower amounts of fluid outside the cells that line the lungs and other body organs.

What Causes the CFTR To Be Missing Or Not Working?

Genes tell the body to make various proteins. Everyone has a pair of genes that provide instructions to make the CFTR protein. Babies with CF have a pair of genes that do not work correctly. They inherited one non-working gene from each parent. Because of the changes in this pair of genes, the CFTR protein does not work properly or is not made at all.

Parents of babies with CF rarely have CF. They each have a single non-working gene for CF. They are called carriers. Carriers do not have CF because the other gene of the pair is working correctly.

How Is CF Diagnosed?

The "sweat chloride" test is usually used to confirm CF. A substance that produces sweat is put on a small section of an infant's arm or leg. A tiny electrical current is used to make that part of the skin sweat. This is painless. The sweat is collected and the amount of chloride is measured. A high level of chloride in the sweat confirms CF. This test does not tell how mild or severe CF will be. The sweat test is usually done after a baby is a few days or weeks old because newborns often do not have enough sweat to do the test properly.

What Are the Symptoms of CF?

In the first few months of life, a baby with CF can have:

- poor digestion of breast milk or formula
- failure to gain weight normally
- frequent coughs
- recurrent lung infections
- salty sweat
- frequent runny stools that are greasy and smelly
- dehydration and
- life-threatening salt imbalance.

The symptoms of CF can be variable. Some will have fewer symptoms while others may have more serious health problems. About 15-20% of newborns with CF will have a blockage of their intestines soon after birth. This is caused by the thick stool that gets stuck in the intestines. About 15% of children with CF have lung but not digestion problems. About 85% of children with CF will have both lung and digestion problems, to a variable degree.

What Is the Treatment for CF?

Early treatment along with ongoing health care by a team at a specialty care center can alleviate many of the CF problems. One goal is to provide the right amount of nutrients to keep your child growing to his or her potential. Another goal of treatment is to keep your child's lungs clear of infection. Treatment can include pancreatic enzymes, dietary supplements, chest percussion, antibiotics and other medicines. The following are typical treatments for children with CF.

Diet and Vitamins

Vitamin Supplements - People with CF have a hard time absorbing some vitamins such as vitamin A, D, E, and K. The CF center staff may suggest these supplements for your child.

High Calorie Diet - Many babies with CF need more food than usual to stay healthy. Some infants will need twice the average number of calories to grow properly. The CF center staff can help you come up with a good nutrition plan for your child.

Pancreatic Enzymes - Most people with CF need to take digestive enzymes before or during each meal and snack. The enzymes will help your child digest and absorb food properly and allow him or her to gain weight and grow at a healthy rate. The CF center staff will explain how to use these preparations.

Airway Clearance

This treatment is done to break up and move mucus that has settled in the lungs so it can be coughed up. It is done several times a day and can take up to 20-30 minutes each time. As the parent or caregiver, you will be involved in chest percussion. This technique involves clapping on the chest and back to break up and move the mucus. The CF center staff will show you how to do this. As your child gets older, a special vest can be worn or a machine can be used. These methods cause vibrations on the chest and back that loosen the mucus.

Medicines

Anti-inflammatory medicines are used to reduce the inflammation (swelling) that is common in the lungs of children with CF. Antibiotics are used to fight off bacteria that infect the lungs of children with CF. Some antibiotics are taken orally, while others are inhaled.

Bronchodilators are drugs that are inhaled and used to prevent and treat wheezing, difficulty in breathing and chest tightness caused by lung diseases. They are also used to prevent breathing difficulties during exercise. They work by relaxing and opening air passages in the lungs to make breathing easier. They come as a solution (liquid) to be inhaled by mouth using a nebulizer (machine that turns medicine into a mist that can be inhaled) and as an aerosol to be inhaled by mouth using an inhaler. Another inhaled drug your doctor may prescribe thins mucus in the lungs for your child.

How Will I Know My Child Is Doing Well On Treatment?

Your CF care center, the baby's doctor and you as the parents are a team to help care for your child and prevent problems from occurring. Check on the following:

- Extra Fluid. Children with CF lose more salt than others. Your child may need to drink more water and liquids to stay hydrated, especially during hot weather and exercise.
- Growth. Making sure nutrition is good and managing digestive problems through pancreatic enzymes will help your child gain weight and grow properly.
- Managing Respiratory Problems. Follow the respiratory care guidelines consistently and recognize the early signs of infection so treatment can start as early as possible.
- Doctor Visits. Visits to the CF care center should be at least quarterly and in addition to your regular baby doctor visits. These visits will help monitor your child's health for changes so early preventive treatments can be started.

How Do I Keep My Child Healthy?

Since children with CF tend to get more lung infections than others, it is best to try to limit as much contact as possible with germs by:

- Careful Hand Washing - This is one of the best ways to avoid catching or spreading germs.
- Immunizations and Vaccines - Be sure to get all the recommended shots for your child including the flu vaccine. Other household members should be vaccinated as well. Talk with your doctor about protection from RSV infection with monthly shots of Palivizumab during the winter.
- Cross Infections - Avoid others with CF and those with a compromised immune system until you talk with your CF care team.
- Smoke - Keep your child away from all forms of smoke. It can increase lung infections and damage.

What If My Child Gets Sick?

If your baby has a lung infection and is too sick to eat or follow regular health habits, call your doctor right away. During some illnesses, your child may need to be seen in the hospital for treatment.

If you are on vacation away from home and need to see a doctor, ask your doctor to give you a letter stating:

- Your child has CF
- The medicines she or he takes
- Other special problems
- Who to call in case of an emergency

A sample of such a travel letter is on page 10 for your reference.

Will My Child Grow Like Others?

Recent improvements in research and therapeutic treatments in CF have lead to improved outcomes for infants, children and adults with CF. CF is a chronic but treatable disease. Many with CF now have a very good quality of life well into adulthood. The key is to learn about CF, to follow your doctors recommendations, to adhere to your child's treatment plan, and to have regular baby visits and scheduled visits with your CF care team. These steps will optimize your child's health, growth potential and development.

For more information about CF, please contact your doctor or your CF care center.

Resources

Cystic Fibrosis Research, Inc.
2672 Bayshore Parkway, Ste. 520
Mountain View, California 94043
(650) 404-9975
www.cfri.org

Cystic Fibrosis Foundation
6931 Arlington Road
Bethesda, Maryland 20814
(800) 344-4823 or (800) Fight CF
www.cff.org

Notes/Questions

Medication prescription for my baby:

Anti-inflammatories:

Antibiotics:

Bronchodilators:

Sample Travel Letter

To whom it may concern:

_____ is under the care of Dr._____ at the CF care center in _____. Cystic Fibrosis (CF) is an inherited disease that affects the lungs and the digestive system.

As a part of their medical care, people with CF need to take the following medications.

_____
_____
_____
_____

In addition, some other problems to be aware of include:

_____
_____
_____
_____
_____

Please call with questions.

Sincerely,

_____
Physician Signature

_____
Print Name

_____
Phone Number Date

California Children's Services And/Or CF Foundation Approved CF Centers

Northern California

Children's Hospital & Research Center at Oakland
Oakland, CA 94609
(510) 428-3305

Lucile Salter Packard Children's Hospital at Stanford
Palo Alto, CA 94301
(650) 723-5191

UCSF Pediatric Pulmonary Center
Walnut Creek, CA 94598
(925) 280-8131

Sutter Memorial Hospital
Sacramento, CA 95819
(916) 453-1454

UC Davis Medical Center
Sacramento, CA 95817
(916) 734-3189

UC San Francisco Medical Center
San Francisco, CA 94143
(415) 476-2072

Kaiser Permanente No. California*
Oakland, CA 94611
(510) 752-6596

Southern California

Children's Hospital Central California
Madera, CA 93638
(559) 353-5587

Children's Hospital of Los Angeles
Los Angeles, CA 90027
(323) 669-4539

Children's Hospital of Orange County
Orange, CA 92868
(714) 289-4059

Rady Children's Hospital, San Diego
San Diego, CA 92123
(858) 966-6790

Naval Medical Center**
San Diego, CA 92134
(619) 532-6896

Loma Linda University
Loma Linda, CA 92354
(909) 558-2301

Miller Children's at Long Beach Memorial Medical Center
Long Beach, CA 90801
(562) 933-8567

Pediatric Diagnostic Center
Ventura County Medical Center
Ventura, CA 93003
(805) 641-4490

Kaiser Permanente So. California*
Panorama City, CA 90033
(818) 375-2909

* Kaiser patients only
** Military patients only

Acknowledgments

We would like to thank the following people for their help in providing input into this booklet:

* California Children's Services Cystic Fibrosis Centers staffs including medical directors, other physicians, dieticians, genetic counselors, nurses, social workers, and other members of the CF center team.

* California Newborn Screening Area Service Centers staffs including medical consultants, project directors, coordinators, program specialists, community liaisons and administrative assistants.

California Department of Public Health
Genetic Disease Screening Program

© CDPH, GDSP, Newborn Screening Branch, February, 2008 - web version 2/14