

## Cystic Fibrosis At a Glance

### What is Cystic Fibrosis (CF)?

Cystic Fibrosis (CF) is an inherited, chronic condition that affects many of the body's systems, especially the lungs and digestion. About 30,000 children and adults in the U.S. have CF. It is most often diagnosed at birth through newborn screening, or before age 2. However, some people with CF have even been diagnosed in their twenties or older. With recent advances in treatment and the right supports, children with CF can be part of regular school programs and activities from preschool to college and beyond.

### What are the effects of CF?

CF affects each child differently and in varying degrees. Because CF is a progressive condition, the effects may change over time, relatively healthy children with CF may face more challenges as they enter middle or high school. It is important to understand the condition and how it may uniquely affect each child.

CF produces thick, sticky mucus that can clog the lungs and pancreas. This can cause respiratory problems and make breathing difficult. It can also lead to digestive problems.

CF is not contagious. It does not affect cognitive ability. However, most children with CF may have a number of symptoms that require special planning for success in school and life.



Meet Rosie on page 12.

## 1. Medical/Dietary Needs

### What you need to know

Children with CF have a variety of needs, which may include:

#### ***Access to food and drink:***

- A high-calorie/high-protein diet in order to gain weight and grow
- Extra food at lunch

- Time to finish lunch
- Frequent opportunities during the day for snacks and drinks
  - These may include high-calorie nutrition supplements
- Fluids and salty snacks such as pretzels or potato chips or salt tablets
  - Needed before, during, and after physical activity or when the temperature is hot indoors or outside (see section 4).

***Digestive enzymes and vitamins:***

- To help with digestion, they may need to take pancreatic enzymes along with meals and snacks.
- They may also need vitamins and other supplements.
- Some schools expect a child to go to the school nurse’s office for medications.
  - However, since most children have been taking pancreatic enzymes since infancy, a student may prefer to handle the medication and vitamins on his or her own.
  - When the child and parents prefer this approach, it should be supported and encouraged.
  - Enabling children to manage certain medications is an important step in building confidence and minimizing feelings of “being different”

***Managing Cystic Fibrosis Related Diabetes (CFRD):***

- Children with CFRD will need to monitor their blood sugar.
- They need ongoing access to snacks or drinks to maintain their blood sugar.
- They may also need to take insulin.

***Coughing:***

- CF can cause a lot of coughing, sometimes producing mucus.
  - Coughing helps the body to clear the lungs, so it should be encouraged.
  - If it is disruptive or embarrassing, the child should be allowed to leave and get a drink of water.
  - The child should have tissues nearby.

***Restroom privileges:***

- Although enzymes help with digestion, children may still experience abdominal pain.
- They may take frequent trips to the bathroom.
  - Access to a private bathroom, such as in the nurse’s office, may help prevent embarrassment.

**Staying healthy:**

- In order to reduce exposure to germs, children should wash their hands frequently.
  - A cleansing hand gel should be used when soap and water aren't available.
  - Other children who are ill should be encouraged to wash their hands often and use tissues when sneezing, coughing, or blowing their noses.

**Physical characteristics and/or symptoms:**

*Not all people with Cystic Fibrosis have all of these characteristics.*

**What are the symptoms of CF?**

*Children with CF may experience some or all of the following symptoms:*

- Gas and stomach pain from problems with digestion
- Fatigue
- Persistent coughing, at times with phlegm
- Frequent lung infections, such as pneumonia or bronchitis
- Wheezing or shortness of breath
- Slow growth/weight gain in spite of good appetite
- Salty sweat
- Spikes and drops in blood sugar from Cystic Fibrosis Related Diabetes (CFRD)
- Longer-than-expected recovery time from colds and viruses
- Depression or anxiety
  - Related to the challenges of living with a chronic health condition
  - Concern about “being different” than their peers

**How is CF treated?**

*Treatment varies depending on the child, but may include:*

- Airway clearance therapy (ACT) which is performed one or more times each day
  - These are treatments that help people with cystic fibrosis (CF) stay healthy and breathe easier.
  - ACTs loosen thick, sticky lung mucus so it can be cleared by coughing or huffing.
    - Clearing the airways reduces lung infections and improves lung function.
    - For infants and toddlers, ACTs can be done by almost anyone.
    - Older kids and adults can do their own ACTs.
- Mucus thinner to make it easier to cough mucus out
- Bronchodilator to help open the airways
- Anti-inflammatory drugs to reduce swelling in the lungs
- High calorie/high protein diet
- Frequent meals or snacks for weight gain and growth

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- Pancreatic enzymes and vitamins
  - To help with digestion and nutrient absorption
- Antibiotics to treat lung infections
- Insulin and glucose monitoring to treat their diabetes
- Regular exercise
- Counseling to help children and their families cope with the stress of a chronic health condition
- Good oral health care to ensure healthy teeth and gums
  - Medications may affect oral health

Since each child is affected differently, be sure to learn about specific needs of each child and ways to ensure a successful school year.

## What you can do

- **Meet with the child, parents, and the school nurse or health coordinator *before* the child first enters your class** to learn about his/her specific dietary and medical needs.
  - If a child transfers in later, schedule this meeting as soon as possible.
  - A child must not be kept from attending classes while special dietary and medical needs are determined.
- **Create a plan to address needs**
  - If the child does not already have an Individualized Education Plan (IEP) under IDEA or an accommodation plan (Section 504 of the Vocational Rehabilitation Act), consider making a referral.
  - An IEP or a Section 504 plan requires a clearly defined outline for how the school will make accommodations for the child's health condition, including managing dietary and medical needs.
  - Section 2, Educational Supports, includes more information on IDEA and Section 504 and examples of possible dietary and medical accommodations for a child with CF.
  - At a minimum, a child with CF should have an emergency plan that all staff (including subs/volunteers) are prepared to follow (see section 6).
- **Notice if the child seems extra tired, is coughing more, and/or shows other medically related needs.**
  - Talk to the child, school nurse, and child's parents about your observations.

## 2. Educational Supports

## What you need to know

A child with CF may be eligible for special education (an Individualized Education Program or IEP) under the Individuals with Disabilities Education Act (IDEA). If he or she requires specially designed instruction to address the unique needs that result from physical and/or mental challenges, an IEP may be needed.

- The IDEA is a federal law that requires public elementary and secondary schools to provide a free appropriate education to children with disabilities.
- Children with CF may be eligible under the “Other Health Impaired” category of IDEA.
  - Services must meet the child’s needs and cannot be determined by the child’s eligibility category.

A child with CF may not be in need of special education (an IEP) in order to participate in the regular school curriculum. However, the child may still need accommodation(s) to access regular classes and programs, as well as extracurricular activities.

- Then, the child may be eligible for a Section 504 plan under the Rehabilitation Act of 1973.
- Section 504 is a federal law that prohibits discrimination against anyone because of a disability by any group (including public schools) that receives federal funds.
- A child is eligible for a Section 504 plan if he or she has a “physical or mental impairment that substantially limits a life activity.”
  - Life activities include: caring for oneself, performing manual tasks, walking, seeing, hearing, speaking, breathing, learning, and working.

## What you can do

Have a clear understanding of the need for educational supports.

- Whether through an IEP, Section 504 Plan and/or emergency plan, the team should ensure that the child has the right interventions and accommodations to be successful.
- When creating the plan, ensure that the child’s school day is as typical as possible. View him or her as a child first who just happens to have a chronic health condition.
- Ensure that everyone who comes in contact with the child (including bus drivers, cafeteria staff and monitors, substitute teachers, faculty and administrators, coaches, specialists, etc.) understands and supports his/her accommodations.
  - This understanding is particularly critical regarding:
    - Access to food/drink
    - Access to bathroom
    - Carrying/taking approved medications such as digestive enzymes.

Examples of what an IEP or a Section 504 Plan might include for a child with CF:

- Medication/Therapy
  - Allow for time during the school day for airway clearance therapy and taking medicines as needed.
  - Adjust school rules to allow the child to take his or her own pancreatic enzymes, vitamins, and certain other medicines (as needed).
  - The school plan should ensure that all who may come in contact with the child (including bus drivers, cafeteria staff and monitors, substitute teachers, faculty and administrators, etc.) understands and supports his/her accommodations.
    - For example, the child may need to carry a note in his/her backpack with permission to carry and take medication such as digestive enzymes so that uninformed adults do not take the pills away from the child such that he or she cannot eat.
- Snacks
  - Ensure unrestricted access to food and drink, including salty snacks (see section 1, Dietary/Medical Needs).
- Bathroom
  - Provide unrestricted access to a bathroom, perhaps a private bathroom in the nurse's office.
- Attendance
  - Adjust or waive attendance guidelines to provide for the child's individual accommodation needs (e.g., illness, multiple medical appointments).
- Shorter Day
  - Allow for a later start or early ending for a child's school day
    - This can help with time-consuming therapies and accommodate fatigue
- Recess
  - Make arrangements for the child to remain indoors if the temperature outside is too hot or cold.
- Academics
  - Monitor daily progress and activities to help ensure that he or she does not fall behind academically.
  - Have a plan to get homework or tutoring to the child when he or she is ill or in the hospital.
    - Children with CF may be absent periodically to receive IV antibiotics for lung infections, among other reasons.
    - Allow the child to have a second set of text books at home.
    - Consider Web cameras or DVD recordings of classes the child may miss.
- Home/School Communication

- Have a communication plan within the school and between the school and the child's parents.
- Counseling
  - Provide counseling to the child
    - This can begin in early elementary school
    - It can help him or her cope with the challenges of a chronic health condition.
- Emergency Planning
  - Prepare a medical emergency plan for the school day, to include:
    - Extracurricular activities
    - School transportation
    - Fire or other drills
    - Actual emergencies as needed (see section 6. Emergency Planning).

### 3. Behavioral and Sensory Support

#### What you need to know

Like many children and families, those living with CF typically are resilient in the face of numerous challenges.

- Children with CF usually want to participate in school life as much as possible along with their peers.
- However, it is helpful when school personnel have an understanding of the stress and demands associated with a complex chronic health condition and are as supportive as possible.
  - For example, the child and family may need to invest a great deal of time and energy every day just to manage the condition.
    - Examples of issues: medications, airway clearance therapies, nutritional needs, health care coordination, medical appointments, hospitalizations, etc.

Children may experience anxiety, depression and pain or discomfort (e.g., deep, chronic cough; gas and stomach pain) related to CF.

- Some worry that peers or teachers will perceive them as “different.” They may try hard to fit in like everyone else.
  - Pre-teens or teens may skip their digestive enzymes and risk stomach pain rather than take pills in front of their friends during lunch.

## What you can do

- Have a clear understanding of the need for any behavioral and sensory supports
- Whether through an IEP, Section 504 Plan and/or emergency plan, work with the team to ensure that the child has the interventions and accommodations needed for school success.
- Ensure that everyone who comes in contact with the child (including bus drivers, cafeteria staff and monitors, substitute teachers, faculty and administrators, coaches, specialists, etc.) understands and supports his/her accommodations.
- Discuss with the team how they prefer to address the issue of informing classmates about their health condition.
  - Young children may want to share some information
    - Such as why they need to take pills when they eat
    - Or the need to stay inside during recess on a very hot day
  - Older children often are very private about the condition and prefer not to tell others.

## 4. Physical Activity, Trips, Events

### What you need to know

A child with CF can safely participate in many physical activities, school sponsored events, and field trips when the right supports and services are provided.

#### ***Physical Education/Activity:***

- Exercise, is particularly good because it strengthens the lungs and helps clear mucus
  - Consider involvement in games, activities, organized sports, and other physical activities
- Children with CF may not have as much stamina as children without CF.
- Children with CF lose much more salt when they sweat than children without CF. They also are more at risk for dehydration.
  - A child with CF should be encouraged to drink fluids directly ***before, during and after*** exercise (PE, playground, etc.) and when it is hot indoors or outside.
  - They need to replace salt by eating high-salt foods like pretzels or potato chips or by taking salt tablets.

- It may be difficult for children with CF to regulate their body temperature during when it is very hot or cold.
  - Make alternative arrangements for recess or physical education, whether outdoors or indoors (e.g., very hot gym) as needed.

#### ***Field Trips & School Sponsored Events:***

- For many children with CF, joining field trips and sponsored school events requires planning and back up planning. For example, making sure the child has *extra* digestive enzymes, food, and drink available is critical. Plan ahead!
  - Running out of enzymes hours away from school on a trip could pose a significant problem
  - Ensure a drink is handy to help swallow the enzymes
- Under the IDEA and Section 504, if a child with CF needs accommodations or support services to be part of a school program or event, they must be provided by the school district.

#### **What you can do**

- Understand the need for support or accommodations for physical activity, trips, and school-sponsored events.
- Whether through an IEP, Section 504 Plan and/or emergency plan, work with the team to ensure the right interventions and accommodations. This will allow the child to safely and successfully join physical activities, trips, and events.
  - Plan ahead. Have extra food, drinks, medications, etc. on hand.
- Ensure that everyone who comes in contact with the child (including bus drivers, cafeteria staff and monitors, substitute teachers, faculty and administrators, coaches, specialists, etc.) understands and supports his/her accommodations.

## **5. School Absences and Fatigue**

#### **What you need to know**

- A child with CF may experience fatigue and have frequent absences related to the condition.

#### **What you can do**

Have a clear understanding of the need for support or accommodations relative to school absences and fatigue.

- Whether through an IEP, Section 504 Plan and/or emergency plan, work with the child and family to ensure that the child has the right interventions and accommodations to address absences and fatigue and succeed in school.
- Ensure that everyone who comes in contact with the child (including bus drivers, cafeteria staff and monitors, substitute teachers, faculty and administrators, coaches, specialists, etc.) understands and supports his/her accommodations.

## 6. Emergency Planning

### What you need to know

Every child with CF should have an emergency plan

- It may be simple and just describe how medication and treatments are to be managed in case of emergency
- It may be much more extensive and complex to address significant health issues.

### What you can do

In collaboration with the child, family, school nurse, child's doctor, and CF team, create a written, individualized emergency plan based on the child's needs. Include:

- All relevant issues (e.g., dietary/medical, behavioral, etc.)
- Possible emergency situations (child medical emergency, fire drills, actual fire, natural disasters, school lock downs, etc.)
- Extreme temperatures (indoor and outdoor)
- Multiple settings (school, field trips, events, on the bus, etc.)
- The role of school staff in managing CF, which may also include specific assignments of emergency actions by school teachers and staff (e.g., chest-clearing techniques when emergency measures should be taken)
- Strategies for communicating with the parents

For children with significant health needs, you may want to alert the local fire/emergency department ahead of time regarding the child's needs in an emergency.

## 7. Resources

There are many on-line supports for schoolteachers and staff to assist in identifying, accommodating, and supporting children with CF in the schools. The following will get you started. Take note of related links to other helpful information as you search these and other sites:

### **Cystic Fibrosis Foundation**

The Cystic Fibrosis Foundation is a nonprofit donor-supported organization dedicated to attacking cystic fibrosis from every angle. Our focus is to support the development of new drugs to fight the disease, improve the quality of life for those with CF, and ultimately to find a cure. [www.cff.org](http://www.cff.org)

"Living with CF at School", produced by the CF Foundation, is intended to help individuals manage CF at school.

<https://www.cff.org/Living-with-CF/CF-and-School/>

### **Genetics Home Reference**

Consumer-friendly information about human genetics from the U.S. National Library of Medicine

<http://ghr.nlm.nih.gov/condition/cystic-fibrosis>

### **National Heart Lung and Blood Institute**

*"What is Cystic Fibrosis?"*

<http://www.nhlbi.nih.gov/health/health-topics/topics/cf/>

### **Sample 504 Plan for a child with CF**

Here is a sample 504 Plan for a child with CF, written by a parent.

<http://www.happyheartfamilies.com/articles/article/6447390/130282.htm>

### **Sample emergency plan**

Here is a sample emergency plan and forms for a child with special health needs (including CF).

<http://opi.mt.gov/pdf/health/healthcareneeds.pdf>

### **Individuals with Disabilities Education Act (IDEA)**

IDEA is a law ensuring services to children with disabilities throughout the nation. IDEA governs how states and public agencies provide early intervention, special education and related services to more than 6.5 million eligible infants, toddlers, children and youth with disabilities.

<http://idea.ed.gov/>

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## 8. Meet a Child with Cystic Fibrosis

### *Insights about Rosie*

*GEMSS would like to thank Alex and his mother for their generosity in sharing this story with us. You have made the site come to life with the addition of your thoughts and feelings. Thank you so much!*



Rosie is a strong willed, very determined, and independent fourteen year old who has a diagnosis of Cystic Fibrosis. She is a 'typical teenager' and a 'tough character' according to her mother Paula. Rosie is doing well in high school, academically, athletically, and socially. She 'knows what she wants' and is taking good care of her medical needs and challenges, balancing her health and school life well.

As an avid athlete, Rosie can be found playing field hockey or on the track team working very hard! Because CF causes her to sweat more, she has to take great care to hydrate, especially on warm days because her brain doesn't process the feeling of thirst typically. The demands of a travel team, night games and events, and the temperature changes all create challenges for her to manage her food and fluid intake carefully. Eating well, not fast foods, creates better outcomes for her. And staying well-hydrated is so important, especially during athletic events.

Her mother feels that Rosie learned a great deal from listening to an adult woman who also experiences CF when Rosie was thirteen years old. It was like a light bulb went off, and this woman had a profound effect on Rosie in that she began to realize she could take control her own destiny.

School has been very supportive of Rosie and, although she is the only student who has CF in the system, they will do whatever needs to be done for her. When she was younger, she knew the nurses 'very well' according to Paula. "If I had it to do over" she warns, "I would have kept her out of the nurse's office more, as they can be great places for germs."

Her issues are mainly digestive and if she is not feeling well, she will often text her mom and they will figure out how to manage the issue. For example, if she forgot to take her enzymes the

day before, she might have a stomachache. Adherence to her medication is a huge issue and the school allows her to carry the meds she needs so she can be in control. Rosie has a 504 plan to help her have extra time when needed, to be able to go to the bathroom whenever needed, to have water and snacks at any time, and to carry her meds with her.

When she was younger, she had to go to the nurse to get her own snack when other children brought in special snacks. As a preschooler, a bit of extra care was needed from her teacher learning some self care skills and they were 'very fortunate to have a supportive preschool,' says Paula.

As a high school student, Rosie enjoys her friends and get-togethers. Large sleep-overs can be a 'recipe for disaster' as far as breaking up the routines that work to keep her healthy and also can be high risk for infection. Rosie is very busy managing the demands on time that CF creates. She has to work in two nebulizer treatments, fit in meds and vest therapy, prioritize sleep, manage her stress, and get plenty of exercise. It takes a level of maturity to make sure all this is done well. Rosie goes to a CF clinic at least every 3 months (more if she isn't feeling well). The fact that the clinic is less than an hour from her home is a huge advantage, as it does not interfere too much with her school schedule.

As Rosie ages, with more transitions and perhaps college in her future, they will take on the challenges as a team and make sure Rosie has all the supports she needs to be successful!