Genetic Education Materials for School Success

Cystic Fibrosis

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Acknowledgment:

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Disclaimer:

This is not intended to be medical or clinical advice. Any recommendations here should be reviewed for the appropriateness for your individual child and their unique situation. Also, please note that each document should include the date that it was last reviewed or updated. Research and guidelines that have been released after that date may represent more up-to-date recommendations.

Cystic Fibrosis

At a Glance

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Cystic Fibrosis (CF) is a common genetic condition that affects many of the body's systems, especially the lungs and digestion. It is most often diagnosed at birth through newborn screening. 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.

CF affects each child differently. 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. CF produces thick, sticky mucus that can clog the lungs and pancreas. This can make breathing difficult. It can also lead to digestive problems. CF is not contagious, and it does not affect intelligence or thinking. However, most children with CF may have symptoms that require special planning for success in school and life.

Common Features of CF:

- Chronic coughing, at times with phlegm
- Frequent lung infections, such as pneumonia or bronchitis
- Wheezing or shortness of breath
- Longer-than-normal recovery time from colds and viruses
- Fatigue
- Slow growth/weight gain despite good appetite
- Spikes and drops in blood sugar
- Gas and stomach pain from problems with digestion
- Salty sweat
- Depression or anxiety related to
    - Challenges of living with a chronic health condition
    - Concerns about "being different" than their peers

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In Depth

Medical and Dietary Considerations

What You Need to Know

Since each child is affected differently, be sure to learn about the specific needs of each child and ways to ensure a successful school year. Children with CF will have specialists supporting their care. The treatment varies depending on the child, but may include:

For the Lungs

To clear mucus and avoid infections

- *Airway clearance therapy* (ACT) which is performed one or more times each day
    - 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
- *Bronchodilators* to help open the airways
- *Anti-inflammatory drugs* to reduce swelling in the lungs
- *Antibiotics* to treat lung infections

For Nutrition and Digestion Support

To increase nutrient absorption

- *High calorie/high protein diets*
- *Frequent meals* or snacks for weight gain and growth
- *Pancreatic enzymes* and vitamins
    - To help with digestion and nutrient absorption
- *Insulin and glucose monitoring* to treat their diabetes

General Support

- *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

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, or as soon as possible in the event of a transfer. A child should not be kept from attending classes while special dietary and medical needs are determined. 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. Below are recommendations to consider when developing your plan.

Access to food and drink:

- Because even nutritious food is poorly digested, a high-calorie/high-protein diet is needed in order to gain weight and grow
- They may require
    - extra food at lunch and extra 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
        - Especially before, during, and after physical activity or when the temperature is hot indoors or outside

Digestive enzymes and vitamins to help with digestion:

- Some schools prefer a child to go to the school nurse's office for medications.
    - However, since most children with CF have been taking pancreatic enzymes since infancy, a student may request to handle the medications on their own.
    - Schools should honor such a request if possible
    - 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):

- Frequent monitoring of blood sugar levels may be required.
- Access to snacks or drinks should be permitted to maintain proper blood sugar levels
- Insulin may be required

Management of Coughing:

- 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.
    - Tissues should be easily available

Restroom privileges:

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

Staying healthy:

- Frequent handwashing is especially important to avoid or reduce exposure to germs

which could lead to dangerous lung infections
    - A sanitizing hand gel should be used when soap and water aren't available.
    - Classmates who are ill should be encouraged to wash their hands often and use tissues when sneezing, coughing, or blowing their noses.

Communicate with the parents if you notice changes in the child's energy levels, behavior, mood, or school performance.

Educational Supports

It is important to have high expectations in educating children who have CF, utilizing their strengths and interests as a foundation. Encourage use of the core educational curriculum and modify it to meet the child's individual needs.

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.

A child with CF may not need special education (an IEP) to participate in the regular school curriculum. However, the child may still need accommodation(s) to access regular classes and programs, and extracurricular activities. The child may be eligible for a Section 504.

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 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.
    - 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.

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 provide the child with permission to carry and take needed medications (digestive enzymes).
  - Adjust school rules to allow the child to take his or her own pancreatic enzymes, vitamins, and certain other medicines (as needed).
- Snacks
  - Ensure unrestricted access to food and drink, including salty snacks.
- 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
  - Arrange for the child to stay indoors if the temperature outside is too hot or too cold.
- Academics
  - Monitor daily progress and activities to help ensure that the student does not fall behind academically.
  - Have a plan to get homework or tutoring for the child when they are 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 textbooks at home.
    - Consider Web cameras or 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 them cope with the challenges of having 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).

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 understand 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 the child’s team is updated on any necessary changes.
- 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
        - 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.

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*** *e*xercise (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 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. For example, make sure the child has *extra* digestive enzymes, food, and drink available. 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

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 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.

School Absences and Fatigue

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

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.)
- Be aware of temperature extremes (indoor and outdoor) at all venues, e.g., school, field trips, events, on the bus
- Define the role of school staff in managing CF, which may also include specific assignments of emergency actions by teachers and staff (e.g., chest-clearing techniques when emergency measures should be taken)
- Establish 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.

Resources

Medline Plus

MedlinePlus is an online health information resource for patients and their families and friends. It is a service of the National Library of Medicine. This link will provide a good medical overview of the condition without being too technical.
https://medlineplus.gov/genetics/condition/cystic-fibrosis/

Cystic Fibrosis Foundation

The Cystic Fibrosis Foundation is a nonprofit organization that supports research to improve care and work towards a cure but also works to support families to get the tools they need. Click on the “Intro to CF” tab and scroll to the bottom for information for teachers.
https://www.cff.org/

Advice from a CF Mom

Here is a sample 504 Plan for a child with CF, written by a parent. There are other links on the website as well that you may want to review.
https://www.happyheartfamilies.com/articles/article/6447390/130282.htm

Info for School Nurse and Primary Care Staff

Cystic Fibrosis is a complex condition that will be best managed by good collaboration between the school nursing staff and primary care staff as part of the student's team. Work with the parents to get signed releases to share information at the school nurse's office and the primary care office.

Gene Reviews is a peer-reviewed point-of-care reference accessible via the National Library of Medicine. The entry for cystic fibrosis provides a comprehensive summary of the condition and describes some ongoing surveillance and management issues. It is a great resource for medical providers. https://www.ncbi.nlm.nih.gov/books/NBK1250/

HIPAA Concerns

Sometimes concerns by primary care staff about violating HIPAA regulations can hamper care by limiting access to necessary medical information. Please note that if care is intended to be delivered by school staff, clarifying medical orders or medication instructions represents continuation of medical care and is not a violation of HIPAA. To avoid any issues like this, remind the parents to sign a release of information form at the primary care office. The school nurse should keep a copy of that release in the student's file, if possible, to facilitate sharing of information in the event that there is difficulty obtaining needed information.

Example letter for physician to provide to school

"My patient______________________ has been diagnosed with Cystic Fibrosis. This is a genetic condition that affects multiple organs, most notably the lungs and digestive system. It can impact growth but does not generally lead to developmental delays. The child may miss more school than a typical student because of their increased risk of lung infections and their need to attend specialists' appointments.

This condition necessitates an Individualized Education Program (IEP) or 504 plan. The student can fully participate in school activities with appropriate modifications and accommodations."

Meet a Child with Cystic Fibrosis: *Insights about Rosie*

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 with 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 stomach ache. 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 sleepovers 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!

Thank you so much to Rosie and family for sharing their story! And thank you to the team at the Institute on Disability at UNH for assisting in collecting these stories.