Cystic Fibrosis (CF)
For Healthcare Providers

This is a customized health care provider version of our website. Please visit the main website to find more comprehensive information for families and schools (www.negenetics.org).

Physical characteristics and/or symptoms

Note: not all people with CF will have all of these features.

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.

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.
    - ACTs loosen thick, sticky lung mucus so it can be cleared by coughing or huffing.
- 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
- Pancreatic enzymes and vitamins
- 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

Recommended Routine Surveillance

- Frequent visits to CF care providers and/or a CF clinic to monitor for changes in symptoms
- Cultures of respiratory tract secretions at least four times yearly
- As needed: pulmonary function studies, chest radiographic examination, annual electrolytes, fat-soluble vitamin levels and IgE levels; bronchoscopy and chest CT examination
- Monitor weight gain and caloric intake in infants until age six months
- Annual oral glucose tolerance test in individuals older than age ten years

- Evaluation of bone mineral density starting in adolescence
- Annual liver function tests and liver ultrasound to monitor progression of liver disease

Emergency Protocols

There are no specific emergency protocols for this particular condition as it is not typically associated with episodes of sudden and serious medical decompensation.

- Emergencies should be handled as with any child.
- 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

Specialists Who May Be Involved

Follow up is need on a case-by-case basis. A multidisciplinary team approach to best meet the child's individual needs is recommended.

- Endocrinologist
  - Pancreatic insufficiency
  - CF-related diabetes mellitus
  - Nutritional support
  - Liver disease
- Gastrointestinal
  - Meconium ileus
  - Distal obstructive syndrome
- Geneticist / Genetic Counselor:
  - Diagnosis
  - Coordination of care
  - Genetic risk for family
  - Clinical trials
- Pulmonologist
  - Respiratory disease

Sample Forms

- Sample paragraph to be used for Letters of Medical Necessity or Letters to the school:

My patient_____ has been diagnosed with Cystic Fibrosis (CF). This condition is characterized by respiratory and digestive disease. Medical complications with CF

included breathing problems, difficulty digesting food and susceptibility to developing lung infections from germs. Because of these, _____ needs the following accommodations.

Please see www.cff.org/Life-With-CF/ for a sample letter.

- Sample health plan for school: https://cfsmart.org/wp-content/uploads/2017/01/CFF035I-CF-Health-support-plan-Primary.pdf

Seven Important Aspects of School Life

"Cystic Fibrosis at a Glance" will help you talk with parents and schools about:

- Medical / Dietary Needs
- Education Supports
- Behavior & Sensory Supports
- Physical Activity, Trips, Events
- School Absences & Fatigue
- Emergency Planning
- Transitions

Resources

GeneReviews: Cystic Fibrosis
https://www.ncbi.nlm.nih.gov/books/NBK1250/

Medline Plus
https://medlineplus.gov/genetics/condition/cystic-fibrosis/

Clinical Care Guidelines
https://www.cff.org/Care/Clinical-Care-Guidelines/

Additional resources including support group information can be found on the main website.