IF YOU THINK cystic fibrosis (CF) is strictly a childhood disease, think again. Because of scientific discoveries and improvements in clinical care, 40% of the estimated 30,000 patients with CF in the United States are 18 or older.¹

In the 1950s, children born with CF rarely survived to first grade. Today, the median life span for Americans with CF is 36.5 years.¹

Cystic fibrosis affects all races but primarily whites. About 1,000 patients are diagnosed each year, with males and females being equally affected. Although more than 80% of the newly diagnosed patients are under age 3, nearly 10% are 18 or older when first diagnosed.¹ Some may have been misdiagnosed earlier in life as having asthma or chronic bronchitis.

In this article, I’ll discuss what you need to know about assessing your patient and helping her manage complications and maintain her health.
Cystic fibrosis is a genetic disease of the exocrine glands. Because of a defective gene, the body produces abnormally thick and tenacious mucus, primarily in the respiratory, gastrointestinal, and reproductive systems. (See Behind the scenes with CF.)

Currently, genetic testing for CF isn’t recommended for the general population. The American College of Obstetricians and Gynecologists recommends that whites be tested for the gene if they are pregnant or considering pregnancy. In infants or young children, common signs and symptoms of CF include meconium ileus (a small-bowel obstruction in newborns caused by thick, tenacious meconium), chronic respiratory infections, and failure to thrive.

The first sign of CF may appear in the first 48 hours of life when the newborn fails to pass a meconium stool. Subsequent stools may be large and bulky, fatty or greasy (steatorrhea), and foul smelling. Large, difficult-to-pass stools may cause rectal prolapse in infancy or early childhood.

Even with a healthy appetite and diet, an affected child may have trouble gaining or maintaining weight due to problems with nutrient absorption. She’s likely to have a distended abdomen, with thin extremities. She’ll bruise easily because her ability to absorb fat-soluble vitamins A, D, E, and K is impaired.

Respiratory signs and symptoms include a chronic moist, productive cough caused by accumulated secretions. The child may have chronic purulent nasal discharge and recurrent episodes of bronchitis and bronchopneumonia. Air trapping sometimes causes a barrel-shaped chest. If chronic fibrotic changes in the lungs impair gas exchange, she may have cyanosis and clubbing of her fingers and toes.

Diagnostic evaluation

The Centers for Disease Control and Prevention recommends that states include testing for CF in newborn screening blood work. Newborns with the disease have an elevated serum level of the pancreatic enzyme trypsinogen and are referred for further testing.

The gold standard for diagnosis of CF is the sweat chloride test (piocarpine iontophoresis). The test stimulates the production of sweat, then measures the amount of sweat electrolytes. A sweat chloride concentration is normally less than 40 mEq/L; a level greater than 60 mEq/L indicates CF. For an infant younger than 3 months, a chloride content over 40 mEq/L is considered borderline for diagnosis.

Behind the scenes with CF

In CF, the abnormal gene interrupts the transport of sodium and water across the cell membranes. Instead of forming freely flowing secretions, the mucous glands produce an unusually thick and sticky mucoprotein that affects numerous organs. Inflammation and lung changes have been found in infants as young as 4 weeks old. Eventually, all mucous ducts become obstructed and damaged.

Clearing the air

To maintain gas exchange and prevent infection, clearing mucus from the lungs is an important part of a patient’s daily CF treatment regimen. A patient may use some or all of the following:

- Chest physiotherapy is the cornerstone of respiratory therapy. It’s usually performed twice a day (morning and evening) or more often as needed (for example, when the patient has a respiratory infection).
- Postural drainage and percussion of the lungs loosen secretions to encourage coughing and expectoration. A family member, respiratory therapist, or nurse percusses or applies vibration over each portion of the lung for 3 to 5 minutes while the patient is repositioned to promote drainage. Electric percussors are available for older children and adults.
- Positive expiratory pressure involves a breathing device with a special expiratory valve that creates a positive pressure in the airways during exhalation. The patient breathes in and out about 15 times, then removes the mouthpiece and performs two or three forced but gentle exhalations like breathing on a mirror to steam it up (called huffing), followed by a cough (huff cough technique). This method, which is less tiring than just coughing, stabilizes small airways, increases aeration to the alveoli, and improves pulmonary functioning.
- The flutter mucus clearance device is a small handheld plastic pipe with a mouthpiece and stainless-steel ball inside. As the patient exhales, the weight of the ball provides intermittent positive expiratory pressure, promoting movement of secretions in the lungs and expectoration. Most patients can clear their airway in 5 to 15 minutes when using this technique.

The patient should include daily physical exercise in her regimen to stimulate mucus expectoration and promote a sense of well-being. Encourage her to do any type of aerobic exercise that she enjoys.

Special dietary needs

Meeting nutritional needs is an important goal for a patient with CF. Because her body can’t readily absorb nutrients and vitamins from food, she needs a well-balanced, high-calorie diet plus supplemental vitamins. Taking exogenous pancreatic enzymes will help her absorb nutrients. As her pulmonary disease progresses, she’ll have chronic weight and nutrition problems because of her increased pulmonary energy requirements.

Encourage her to get calories from complex carbohydrates and fats.
Dietary fat is high in calories, improves the taste of food, and provides essential fatty acids. Salt depletion from sweating can be a problem during hot weather or exercise, so she may need to consume extra salt in hot weather or when she’s febrile.

Pulmonary exacerbations are characterized by shortness of breath and increased work of breathing, and the patient may experience anorexia, nausea, or early satiety. She may need short-term or long-term supplemental enteral or parenteral nutrition to meet her nutritional needs.

Complications throughout life
As the patient ages, some new problems may arise or existing problems may worsen. Here’s a look at some common complications.

Respiratory problems. Almost all patients with CF have respiratory complications. Impaired airway mucus clearance is the primary cause of chronic respiratory inflammation and periodic infections. The copious abnormal secretions, which are difficult to expectorate, trap air in the bronchioles, leading to alveolar hyperinflation and atelectasis. An older child or adult may develop pneumothorax or hemothorax. Secretions also impair her ability to exhale carbon dioxide, which can cause respiratory acidosis.

A pulmonary infection, once established, is difficult to eradicate because of the thick, tenacious mucus. As the disease progresses, the airway walls thicken due to increased mucus, inflammation, and bacteria, leading to dilation of the airways and enlarged bronchial arteries. Lung function usually continues to decline. More than 95% of patients with CF eventually die of pulmonary disease.

Gastroesophageal reflux disease (GERD). In many cases, chronic coughing and forced respirations during chest physiotherapy can lead to GERD. Some respiratory medications increase reflux and worsen GERD. (See Tips for avoiding GERD.)

Pancreatic problems. Most patients with CF suffer pancreatic damage from blocked pancreatic ducts. The pancreatic enzymes that are needed to digest fats and proteins aren’t secreted, so food is digested poorly. More than 90% of patients with CF have pancreatic insufficiency, sometimes leading to pancreatitis. Signs and symptoms include severe epigastric pain, nausea and vomiting, abdominal distension, tachycardia, and fever. Diagnosis and treatment of pancreatitis is the same as for patients without CF.

Intestinal obstruction. Thick intestinal secretions, which usually continue to be a problem for patients with CF throughout their life, can lead to intestinal obstruction. Signs and symptoms include abdominal distension and pain, vomiting, constipation, dehydration, and electrolyte imbalance.

Distal intestinal obstruction syndrome is a chronic problem of the small bowel related to abnormal intestinal mucus, prolonged gastrointestinal (GI) transit time, and abnormal fluid and electrolyte transport. The small-bowel obstruction causes right lower quadrant pain, abdominal distension and cramping, flatulence,

<table>
<thead>
<tr>
<th>Medications to treat....</th>
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<tr>
<td><strong>the gastrointestinal system</strong></td>
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<tr>
<td>Lactulose</td>
<td>Given to prevent constipation</td>
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<tr>
<td>Multivitamins, iron; vitamins A, E, D, and K when deficient</td>
<td>Given because the production and absorption of vitamins are decreased by the thick secretions. Give water-soluble vitamins for better absorption.</td>
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<tr>
<td>Pancreatic enzyme concentrates such as pancrélipase (Viokase)</td>
<td>Given to decrease the fat and bulk of stools. Give immediately before or with meals and snacks to increase the absorption of nutrients.</td>
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<td>Ursodeoxycholic acid or ursodiol</td>
<td>Used to treat and possibly prevent progressive liver disease in CF. It improves the viscosity of biliary secretion.</td>
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<td><strong>the respiratory system</strong></td>
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<td>Bronchodilators (albuterol, terbutaline)</td>
<td>Used to open airways. They’re used before chest physiotherapy (CPT) and to relieve symptoms; administered via nebulizer or metered-dose inhaler.</td>
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<tr>
<td>Dornase alfa (Pulmozyme)</td>
<td>Used with standard CF therapies to improve pulmonary function and to decrease risk of respiratory tract infections requiring parenteral antibiotics; administered via nebulizer</td>
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<tr>
<td>Aerosol antibiotics (gentamicin, ticarcillin, tobramycin)</td>
<td>Administered via nebulizer after CPT. Aerosolized antibiotics decrease systemic absorption.</td>
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<tr>
<td>Oral and I.V. antibiotics</td>
<td>Prolonged administration with high doses given to treat infection; specific drug based on culture sensitivity</td>
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<tr>
<td>Corticosteroids and ibuprofen</td>
<td>Decreases inflammation from infection. Corticosteroids and ibuprofen are given on alternating days to decrease adverse reactions to corticosteroids.</td>
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anorexia, and weight loss. Surgery is usually needed to treat a complete GI obstruction. A partial obstruction can be treated by administering a balanced polyethylene glycol-electrolyte solution (Golytely) by mouth or nasogastric tube. **Diabetes.** Glucose intolerance and CF-related diabetes (CFRD) can become a problem for adults with CF. Rare in children, it becomes more common with advancing age, affecting 20% to 30% of patients older than age 20. Patients diagnosed with CFRD usually have more severe pulmonary disease, poorer nutritional status, and shorter life expectancy than other CF patients. The primary cause of CFRD is insulin deficiency, but glucose metabolism is also impaired. Most patients with CFRD need insulin to manage it.

**Reproductive problems.** Cystic fibrosis affects the reproductive system in both men and women. Most men are sterile because abnormal secretions block sperm in the vas deferens. Women have normal, functioning ovaries and fallopian tubes, but becoming pregnant may be difficult because thick secretions may block sperm from entering the uterus. Artificial insemination and in vitro fertilization are options for women who want to become pregnant.

**Is transplant an option?** When a patient has severe lung disease, a lung transplant may be an option. (Other criteria for having a transplant depend on the transplant center.) It’s estimated that 120 to 150 patients with CF have lung transplants each year, with about 90% of them surviving 1 year after the transplant and 50% surviving at least 5 years. The surgery can be a double-lung transplant from a cadaver or, much less commonly, lower lobe transplants from two living donors. Transplanted lungs won’t have the defective gene that produces thick, tenacious mucus, but the condition of other organs won’t be improved by the transplant.

If a patient decides to have a transplant, she may have a long wait for donated lungs. Disadvantages of transplant include possible surgical complications and having to take immunosuppressant drugs to prevent organ transplant rejection, increasing her risk of infections and malignancies.

**Psychosocial support**

The Cystic Fibrosis Foundation supports research and provides educational materials and services. You can help your patient’s family contact groups that may provide equipment and medications for low or no cost. When CF is diagnosed during childhood, the patient and her parents need strong emotional support. Parents must adjust to the stress of raising a child with a chronic, life-threatening disease. Other issues that impact the family are the demands of many treatments and hospitalizations and the financial burden. Help the parents teach the child and other family members how to manage the disease.

When the patient is 10 to 12 years old, she’ll be able to understand that she can die of the disease. Be sure to give her hope for future breakthroughs in treating the disease. When your patient is a teenager or adult, she’s likely to know more about her condition and treatments than you do, so collaborate with her on her care when she’s hospitalized.

**End-of-life issues**

Your patient may have to face complex end-of-life and ethical issues. For example, if she’s an adult, how will she make informed decisions about advanced directives and choosing do-not-resuscitate status? Or if she’s still an adolescent, how can her family make decisions with her input?

A patient with CF and her family may have tremendous physical, psychosocial, and educational needs, requiring comprehensive, multidisciplinary care. If you work with patients with CF, stay up-to-date on diagnostic and evidence-based treatment options. Be sure to convey to them that advances are being made in treatment and survival rates all the time, with more likely to come. Refer them to Web sites (see Resources) so they can keep up with research and advances.

**Tips for avoiding GERD**

Teach your patient to:

- avoid large meals and tight clothing
- perform postural drainage before meals
- remain in an upright position for 30 minutes after meals
- not eat or drink fluids within 2 hours of bedtime
- elevate the head of her bed on 8- to 12-inch (20- to 30-cm) blocks.

**REFERENCES**


**RESOURCES**


Web sites last accessed on June 5, 2007.

Janelle Gardner is an associate professor of maternal-child nursing at the school of nursing at California State University in Chico.