

Treatments for Cystic Fibrosis

In conversations with patients, case managers can help patients and caregivers understand their CF treatments.

The Cystic Fibrosis Foundation (CFF) recognizes the importance of treating CF early and provides guidelines based on the latest research, medical evidence, and consultation with experts on best practices.¹⁻³

According to the Cystic Fibrosis Foundation, treatment plans for people with CF may include⁴:



Quarterly visits to their CF care center



Airway clearance techniques



Oral, inhaled, and nebulized medications



Nutrition plans



Fitness plans

There are many CF treatments

Individuals with CF have personalized treatment regimens that often include several medications and other modalities.^{1, 5-23}

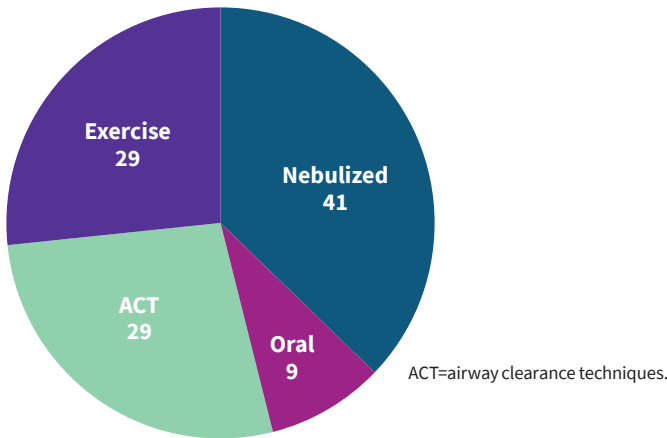
Manifestations	Example treatments
Pulmonary disease	<ul style="list-style-type: none"> • Antibiotics for chronic lung infections and pulmonary exacerbations • Airway clearance, including chest physical therapies, and mucolytic and osmotic agents, as well as anti-inflammatory agents and aerobic exercise for maintenance of lung function, cough with or without sputum, and difficulty breathing • Oxygen, noninvasive ventilation, and lung transplantation for severe lung disease
Pancreatic insufficiency	<ul style="list-style-type: none"> • Pancreatic enzyme supplementation • Nutritional supplementation (eg, fat-soluble vitamins) for weight loss/poor weight gain, fat-soluble vitamin deficiency, flatulence, and abdominal discomfort
CF-related diabetes	<ul style="list-style-type: none"> • Insulin
Intestinal blockage	<ul style="list-style-type: none"> • Acute: meconium ileus (a type of bowel obstruction) requires medical and surgical evaluation; DIOS (a type of bowel obstruction) treatment usually requires hospitalization, surgery only in very severe cases • Chronic: preventative treatment (eg, polyethylene glycol, prokinetic agents)
Liver disease	<ul style="list-style-type: none"> • Ursodeoxycholic acid (to help dissolve gallstones as an alternative to surgery) • Nutritional therapy • End-stage liver disease: portal hypertension management, liver transplant
GERD	<ul style="list-style-type: none"> • Proton pump inhibitors, H₂-receptor antagonists
Bone/joint disease	<ul style="list-style-type: none"> • Exercise (weight-bearing) • Vitamin D and calcium supplements • Bisphosphonates
Psychosocial effects of CF	<ul style="list-style-type: none"> • Psychotherapy, cognitive behavioral therapy, family therapy • Antidepressants

Burden of treatment

Treatment of CF is perceived as time-consuming and burdensome by both patients and caregivers. Patients typically take a median of 7 medications, plus chest physical therapy and treatment sessions daily.^{24,25}

This graph shows the breakdown of time spent on therapy reported in a study of adult patients with CF.

Mean patient-reported time spent on therapy (minutes/day)²⁴



- Therapy consumes an average of 108 (±58) minutes a day for adults and 75 (±57) minutes for children^{24,26}
- Treatment times for children take up an adult's time as well as the child's²⁶

Adapted from Sawicki 2009.

Complex disease requires a CF care team

Because of the complex nature of the disease, people with CF do best when care is comprehensive, coordinated, and done in partnership with their CF care teams. Some care centers are accredited by foundations like the Cystic Fibrosis Foundation to ensure people with CF can receive high quality, specialized care.²⁷

Key members of CF care team²⁷



CF nurse

Primary contact to address patient concerns, answer questions, or direct to the right CF care team member



CF physician

Works with team to develop a patient's medical plan, make changes to care, and apply best clinical practices



CF respiratory therapist

Performs pulmonary function tests and helps with pulmonary therapies, ACTs, and nebulizers



CF dietitian

Helps achieve optimal weight via nutritional plan with recommended calories and nutrients to promote better health



CF social worker

Assesses and counsels on the emotional, social, and financial aspects of living with CF

Patients with CF work with their care team to manage all aspects of their disease.

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