CF AND MENTAL HEALTH: WHAT HEALTH PROVIDERS NEED TO KNOW

A GUIDE FOR CLINICIANS

WHAT IS CYSTIC FIBROSIS FROM A BIOLOGICAL STANDPOINT?

Cystic fibrosis (CF) is caused by mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene, leading sticky mucus to build up in the lungs and cause chronic inflammation, infection, and damage. There are significant differences from one person to the next in how CF impacts the body. Several medications (CFTR modulators) targeting these gene mutations are available, but they do not cure CF or eliminate all symptoms. Approximately 10 percent of people with CF are still waiting for a medication that is matched for their genes. As the disease progresses, individuals face increasing symptoms. Those with end-stage lung disease may pursue lung transplantation to extend their life. Comprehensive information about CF can be found at cff.org.

BODY SYSTEMS THAT MAY BE AFFECTED

- Respiratory system
  - Lungs and sinuses
- Gastrointestinal (GI) system
  - Poor digestion
  - Constipation and obstruction
  - Liver disease
- Endocrine system
  - CF-related diabetes
- Reproductive system
  - 98% of males are infertile
  - Most females can become pregnant

THE BURDEN OF CF DAILY CARE

In addition to typical life stressors, people living with CF manage distinct challenges. Staying healthy involves a complex treatment regimen that can take hours per day and be difficult to sustain. Airway clearance therapy is needed to clear mucus from the lungs. Along with other medications, most people with CF take oral enzymes with meals and snacks to aid digestion and decrease GI discomfort. Many individuals need double the calories required by someone without CF to achieve optimal weight and growth, which might require supplemental tube feedings. During acute illnesses (e.g., pulmonary exacerbations), individuals may stay in the hospital for intravenous antibiotics and miss important activities like school and work. Acute and chronic pain can also impact important daily activities.

WHAT IS CF FROM A PSYCHOSOCIAL STANDPOINT?

The diagnosis of CF is most often made in infancy, after newborn screening results are confirmed by a positive sweat test. This can be a stressful time, as parents plan for their child's complex medical needs. Adjusting can also be difficult for those diagnosed later in life, and as people with CF and their loved ones navigate new challenges during childhood, adolescence, and adulthood. Individuals with CF and their caregivers are at elevated risk for symptoms of depression and anxiety. CF can be a uniquely isolating disease. Symptoms are often invisible to others. In addition, people with CF must avoid close physical contact with each other to avoid cross-infection.

SOME STRESSORS ASSOCIATED WITH CF DIAGNOSIS AND DISEASE

- (Unexpected) diagnosis of lifelong genetic condition
- Keeping up with burdensome daily care regimen
- Social isolation, feeling "different," and disclosure of illness
- Impact on body image and eating behaviors
- Disruption of personal life goals, activities, and relationships
- Financial stress
- Complexity of health care and insurance systems
- Coping with difficult physical symptoms, medical procedures, and pain
- Existential concerns including the uncertainty of illness course and survival

THE KEY ROLE OF THE MENTAL HEALTH PROVIDER

Mental health providers can deliver empirically supported treatments to manage conditions such as depression, anxiety, ADHD, trauma, substance misuse, chronic pain, and distress from medical procedures. There are many ways mental health providers can support people living with CF and their caregivers to cope with stressors and to pursue their life goals.

HOW TO SUPPORT PEOPLE LIVING WITH CF AND THEIR CAREGIVERS

PROMOTE lifelong wellness through healthy sleep, physical activity, nutrition, and understanding mind-body connections.

ENHANCE motivation and skills to sustain complex self-care, with increasing transfer of treatment responsibility as children mature.

ADDRESS the impact of CF throughout the lifespan, especially at developmental milestones such as toilet training, peer relationships and school, transition to college/workplace, planning a family, and disease progression.

BUILD effective communication and trusting relationships within the family, with peers, and with the health care team.

TO LEARN MORE

For questions, call 800-FIGHT-CF (800-344-4823) or email info@cff.org.

Content adapted with permission from:

Mueller AE, Georgiopoulos AM, Smith BA, Quittner, AL, Roach CM, Reno KL, Lomas P, Kvam CM, Filigno SS. (2020). Introduction to Cystic Fibrosis for Mental Health Care Coordinators and Providers: Collaborating to Promote Wellness. *Health and Social Work.*

Comprehensive mental health guidelines for individuals 12+ years with cystic fibrosis can be found:

Quittner, AL, et al. (2014). Prevalence of depression and anxiety in patients with cystic fibrosis and parent caregivers: Results of the international depression epidemiological study across nine countries. *Thorax*, 69(12), 1090-1097.

CYSTIC FIBROSIS FOUNDATION®