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| **The University of Arizona Pediatric Pulmonary Center** |
| Cystic Fibrosis and Mental Health |
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The University of Arizona

Pediatric Pulmonary Center

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Dear Provider:

I am a social work trainee with the University of Arizona’s Pediatric Pulmonary Center, working with children and adults diagnosed with Cystic Fibrosis (CF), and their families. In my clinical traineeship, I have identified the mental health needs of this population and the impact these issues have on a person's overall wellbeing. Mental disorders of anxiety and depression have been found to be highly correlated to patients diagnosed with CF and a mental health diagnosis of depression and/or anxiety can affect a patient's treatment adherence, quality of life, and overall health related outcomes. A needs assessment was created and distributed to CF patients to assess mental health needs. The majority (75%) of CF patients surveyed reported it would be very helpful if the providers had an awareness of the CF diagnosis and its correlation with mental health disorders.

My goal is to educate mental health providers on CF by providing an information packet with an overview of the CF diagnosis and research on CF and mental health. Additionally, the CF Foundation has noted the importance of mental health in providing care for our CF patients and their families, and has recommended annual mental health screening for patients and caregivers, so a referral list of mental health providers aware of CF and the mental health research with CF is extremely helpful for our patients and their families. Please take time to review this information packet and let myself, or our CF clinic social worker, Mary McGuire, MSW, know if you have any questions. Thank you for your time and collaboration with our CF patients and their families.

Sincerely,

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**What is Cystic Fibrosis?**

Cystic fibrosis is a life-threatening, genetic disease that causes persistent lung infections and progressively limits the ability to breathe. In people diagnosed with CF, a defective gene causes a thick, buildup of mucus in the lungs, pancreas and other organs. In the lungs, the mucus clogs the airways and traps bacteria leading to infections, extensive lung damage and eventually, respiratory failure. In the pancreas, the mucus prevents the release of digestive enzymes that allow the body to break down food and absorb vital nutrients.

**Symptoms of CF**

People diagnosed with CF can have a variety of symptoms, including:

* Very salty-tasting skin
* Persistent coughing, at times with phlegm
* Frequent lung infections including pneumonia or bronchitis
* Wheezing or shortness of breath
* Poor growth or weight gain in spite of a good appetite
* Frequent greasy, bulky stools or difficulty with bowel movements
* Male infertility

**Diagnosis and Genetics**

Cystic fibrosis (CF) is a genetic disease. People with CF have inherited two copies of a defective CF gene -- one copy from each parent. Both parents must have at least one copy of a defective gene. ­­People with only one copy of a defective CF gene are called carriers, but they do not have the disease. Each time two CF carriers have a child, the chances are:

* 25 percent (1 in 4) the child will have CF
* 50 percent (1 in 2) the child will be a carrier but will not have CF
* 25 percent (1 in 4) the child will not be a carrier and will not have CF

The defective CF gene contains a slight abnormality called a mutation. There are more than 1,800 known mutations of the disease. Most genetic tests only screen for the most common CF mutations.

**In the United States:**

* About 30,000 people are living with cystic fibrosis (70,000 worldwide)
* Approximately 1,000 new cases of CF are diagnosed each year
* More than 75 percent of people with CF are diagnosed by age 2
* Nearly half of the CF population is age 18 or older

**What to Expect**

CF is a complex disease and the types and severity of symptoms can differ widely from person to person. Many different factors, such as age of diagnosis, can affect an individual's health and the course of the disease. People with CF are at greater risk for lung infections because thick, sticky mucus builds up in their lungs, allowing germs to thrive and multiply. Lung infections, caused mostly by bacteria, are a serious and chronic problem for many people living with the disease. Minimizing contact with germs is highly important for people with CF.

The buildup of mucus in the pancreas can also stop the absorption of food and key nutrients, resulting in malnutrition and poor growth. In the liver, the thick mucus can block the bile ducts, causing liver disease. In males, CF often affects their ability to have children without in vitro fertilization. Breakthrough treatments have added years to the lives of people with cystic fibrosis. Today the median predicted survival age is close to 40. This is a dramatic improvement from the 1950s, when a child with CF rarely lived long enough to attend elementary school.

With tremendous advancements in research and care, many people with CF are living long enough to achieve their dreams of attending college, pursuing careers, getting married and having children. While there has been significant progress in treating this disease, there is still no cure so research continues to work toward extending the life span of individuals diagnosed with CF with medications and treatments.

# Anxiety and CF

Anxiety is one of the most common emotional issues that people face as a CF patient or parent/caregiver, due to high levels of stress with the disease. Making time for daily treatments, remembering to take medications, missing out on activities and being hospitalized for an infection can all cause stress and anxiety, which directly impacts emotional health. Studies show that people with CF, as well as parents who take care of children diagnosed with CF, are more likely to experience anxiety than people in the general population. These CF patients and caregivers experiencing anxiety are also more likely to experience [depression](https://www.cff.org/Living-with-CF/Emotional-Wellness/Depression-and-CF/).

An untreated anxiety disorder affects both the patient and caregivers’ physical and emotional health and how they care for themselves. For example, CF patients with untreated anxiety:

* Are less likely to manage their treatment plans
* Have lower body mass index (BMI)
* Tend to have worse lung function
* Experience more hospitalizations
* Often have higher health care costs
* Experience a lower quality of life

**Depression and CF**

People with chronic diseases, such as CF, are at greater risk for developing clinical depression. When left untreated, depression can interfere with the ability to manage their CF effectively and experience a better quality of life.  Researchers also found that individuals diagnosed with CF and caregivers who take care of children with CF, are more likely to experience depression than individuals in the general population.

# Living with Cystic Fibrosis Cystic Fibrosis Foundation (CFF) Tucson a Day in the Life



This is an educational video of the CFF interview with Dr. Cori Daines and patient.

**YouTube video link:** <https://www.youtube.com/watch?v=_6GBE4uLThM>

**International Committee on Mental Health in Cystic Fibrosis: Cystic Fibrosis Foundation and European Cystic Fibrosis Society consensus statements for screening and treating depression and anxiety**

**Alexandra L Quittner, Janice Abbott, Anna M Georgiopoulos, Lutz Goldbeck, Beth Smith, Sarah E Hempstead, Bruce Marshall, Kathryn A Sabadosa, Stuart Elborn, and the International Committee on Mental Health**

**Abstract:**

Studies measuring psychological distress in individuals with cystic fibrosis (CF) have found high rates of both depression and anxiety. Psychological symptoms in both individuals with CF and parent caregivers have been associated with decreased lung function, lower body mass index, worse adherence, worse health-related quality of life, more frequent hospitalizations and increased healthcare costs. To identify and treat depression and anxiety in CF, the CF Foundation and the European CF Society invited a panel of experts, including physicians, psychologists, psychiatrists, nurses, social workers, a pharmacist, parents and an individual with CF, to develop consensus recommendations for clinical care. Over 18 months, this 22-member committee was divided into four workgroups: Screening; Psychological Interventions; Pharmacological Treatments and Implementation and Future Research, and used the Population, Intervention, Comparison, Outcome methodology to develop questions for literature search and review. The committee reviewed 344 articles, drafted statements and set an 80% acceptance for each recommendation statement as a consensus threshold prior to an anonymous voting process. Fifteen guideline recommendation statements for screening and treatment of depression and anxiety in individuals with CF and parent caregivers were finalized by vote. As these recommendations are implemented in CF centers internationally, the process of dissemination, implementation and resource provision should be closely monitored to assess barriers and concerns, validity and use.

**Key points:**

* A recent study in nine countries screened 6088 patients with CF ages 12 years through adulthood and 4102 parents.
* Elevated symptoms of depression were found in 130 adolescents (10%), 913 adults (19%), 1165 mothers (37%), and 305 fathers (31%).
* Anxiety was reported by 281 adolescents (22%), 1503 adults (32%), 1496 mothers (48%), and 343 fathers (36%).
* High rates of comorbidity were found between depression and anxiety across patient and parent samples.

**Eating Disorders and Disturbance in Children and Adolescents with Cystic Fibrosis**

**Mandy Bryon, Joanna Shearer, and Helen Davies**

**Abstract:**

People with cystic fibrosis are considered at risk for developing anorexia (Raymond et al., 2000), but studies have used methodologically flawed measures. Using improved methodology, the current study examines the prevalence of eating disorders/disturbance in adolescents with CF. **Method:** 55 adolescents with CF, age range 11–17 years (mean 14.2 years) randomly selected were administered the Child Eating Disorder Examination (Bryant-Waugh, Cooper, Taylor, & Lask, 1996). **Results:** No participant met full criteria for a diagnosis of anorexia or bulimia. Of those with a BMI 17.5, 5% avoided weight gain. Fifty-three percent demonstrated disturbed eating attitudes and 16% disturbed eating behaviors. **Discussion:** The study finds that gold standard diagnostic methods indicate the prevalence of disturbed eating attitudes and behaviors in CF.

**Key Points:**

* Nutritional management involves providing enzyme replacement supplements, increased calorie intake (125–140% of recommended daily allowance [RDA] of calories), and vitamin supplements. Adherence to dietary recommendations has been found to be poor and parents frequently struggle to provide adequate nutrition in young children.
* Eating disturbance had been found to be significantly prevalent particularly in the adolescent population with rates around 40–47% and studies have indicated a continuum between eating disturbance and later developed eating disorder.
* Truby and Paxton (2001) found girls with CF at or below 50th BMI percentile, 54% wanted to stay the same or be thinner; for girls with CF body dissatisfaction was a significant predictor for BMI; for boys with CF body dissatisfaction and body esteem were significant predictors of BMI.
* Willis, Miller, and Wyn’s (2001) study of children with CF found that girls are generally happy with their slender body shape and do not wish to gain any weight; a number of girls wished to lose weight irrespective of current weight percentile; boys reported wanting to gain weight and become more muscular and strong.

**Pain in Cystic Fibrosis: Review of the literature**

**Trudy Havermans, Kristine Colpaert, Kris De Boeck, Lieven Dupont, and Janice Abott**

**Abstract:**

Over the past 10 years’ studies have shown that pain assessment and pain treatment in Cystic Fibrosis (CF) are important, especially as pain has been associated with survival. Pain reduces the quality of life of chronically ill patients and may negatively impact the ability to participate in disease-related daily care. Consequently, the assessment and treatment of pain are imperative. **Method:** A proforma was used to record the rationale for the study, characteristics of the sample, pain assessment tools, pain location, frequency and severity, treatment/self-management, coping and the impact on daily activities and quality of life. **Results:** All studies (n = 13) were retrospective. Chest and abdominal pains were most commonly reported. Pain was negatively associated with pulmonary exacerbations, quality of life and treatment adherence. Approximately 50% of patients do not consult their GP or CF team about pain, with many patients reporting self-management. **Conclusion:** A high incidence of pain is reported in CF although there is little standardization of CF pain measurement. The way forward is to develop guidelines on how to assess pain and provide adequate treatment for pain in CF. Efficient pain management will improve patients HRQoL and help patients to perform essential interventions and treatments that prevent daily deterioration.

**Key points:**

* Festini et al. found that 30% of adults suffered pain more than 10 times over a two-month period. Hubbard et al. found that 10 out of 18 adult patients reported daily pain.
* Sermet-Gaudelus et al. found that 40% of the children and 50% of the adults with pain reported using analgesics. Acetaminophen was used most frequently (59%) followed by NSAIDs (10%) and aspirin (5%).
* Sixty-five percent of the children and 60% of the adults reported significant relief after treatment whereas 25% of the children and 10% of the adults reported either very limited or no effectiveness.
* Many patients did take some action to alleviate the pain, other than medication. These actions included homeopathic products or non-pharmacological remedies.

# When Addiction Takes Over: Managing Dependence Issues in the Patient with Cystic Fibrosis

**Nicola J. Felicetti**

**Abstract:**

Comparable to CF, substance abuse and addiction can be classified as chronic diseases. The World Health Organization (WHO) defines a chronic disease as one that is not passed from person to person and is of long duration and generally slow progression. The American Society of Addiction Medicine (ASAM) states that addiction is a primary, chronic disease of brain reward, motivation, memory and related circuitry and further discusses that dysfunction in these circuits’ leads to characteristic biological, psychological, social, and spiritual manifestations. While little is known about the long-term effects of substance addiction in people living with CF, it is generally believed to have a negative impact on health, well-being and longevity of any individual who struggles with a substance use disorder. This subsequently presents as a challenge for the CF healthcare team to assist in the management of two very complex chronic diseases simultaneously.

**Key Points:**

* The National Survey on Drug Use and Health (NSDUH) in 2013 identified that an estimated 21.6 million persons in the U.S. aged 12 or older were classified with a substance dependence or abuse in the past year.
* Overall, 17.3 million had alcohol dependence or abuse, and 6.9 million had illicit drug dependence or abuse.
* While it has been recognized pain is associated with chronic disease, there are limited studies that discuss the long-term management of pain in CF, to include addressing the management of chronic pain medication use. Pain, alone, has been identified as an independent risk factor for inappropriate analgesic use.
* The Centers for Disease Control (CDC) identifies that enough prescription pain killers were prescribed in 2010 to medicate every American adult around-the-clock for a month.
* Most of these pills were prescribed for a medical purpose, but many ended up in the hands of people who misused or abused them.
* When addiction becomes the focus for an individual, the balance and attention towards maintaining health as it pertains to their CF care, is directly affected and is evident in the decline in their overall health status.
* Establishing routine and standardized assessments for those patients receiving care at a CF center, beginning in early adolescence, could identify at-risk patients and assist in the management of existing dependence issues going forward.

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