



**CHOC Children's Hospital**  
*Best Evidence and Recommendations*

**Increasing Cystic Fibrosis Treatment Plan Adherence**

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**PICO:** In 11 to 21-year-old adolescents with Cystic Fibrosis, does an individualized patient care approach better identify patient barriers and enhance treatment plan adherence?

**P (Population/problem):** In 11 to 21-year-old adolescents with cystic fibrosis

**I (Intervention/issue):** does an individualized patient care approach

**C (Comparison):** compared to standard of care

**O (Outcome):** better identify patient barriers and enhance treatment plan adherence?

**Background:**

Cystic Fibrosis (CF) is a progressive genetic disease, which causes a thick buildup of mucus in the lungs, pancreas and other organs. It often results in persistent lung infections and may limit the ability to breathe over time (Cystic Fibrosis Foundation, 2010). There are significant variations in the incidence and morbidity and mortality of CF around the world. It is the most common, life-limiting autosomal recessively inherited condition in Caucasian populations (Williams, 2010). Over the past several decades, advances in CF treatments and medications have led to an increased life expectancy for this patient population. The current average lifespan of CF patients in the U.S. is 37.5 years. (Cystic Fibrosis Foundation, 2010).

Due to the complexity of the treatment regimens, patients with pediatric pulmonary diseases such as cystic fibrosis encounter numerous challenges in adhering to their treatment. Research has demonstrated that poor adherence can have potential serious consequences including increased morbidity and mortality, decreased quality of life, and increased cost of care. Noncompliance is linked to poor clinical outcomes such as pulmonary exacerbations, decreased lung function and increased hospitalizations (Goodfellow, Hawwa, Reid, Horne, Shields, & McElnay, 2014).

Several factors have been shown to influence adherence including: patient and family knowledge of the disease and the importance of treatments; multidisciplinary team communication; and the complexity of the treatment. Additionally, studies suggest that noncompliance increases with patient age, independence, and disease severity (Llorente, Garcia, & Martin 2008).

Consistent with the literature, treatment adherence in the cystic fibrosis population has been identified as an opportunity for improvement at CHOC Children's. Despite our multidisciplinary team approach, gaps in patient-provider communication and the complexity of treatment plans appear to be contributing factors. Even under optimal conditions, compliance with the plan of care can be challenging for a CF patient and their family.

The purpose of this evidence-based practice project was to conduct a comprehensive review of the literature to explore common factors that influence treatment adherence in the CF population and



determine if an individualized patient care approach will better identify patient barriers and enhance treatment plan compliance, leading to better patient outcome.

### **Search Strategies and Databases Reviewed:**

Databases searched for this review included: CINAHL, PubMed, and Cochrane. Key search terms included: treatment, adherence, barriers, compliance, motivation. A total of 16 research articles were reviewed; 11 articles were of a qualitative design, 3 studies were both quantitative and qualitative (mixed) design, and 2 were systematic reviews.

### **Synthesis of the Evidence:**

The literature review yielded primarily qualitative studies. The patient's perspective was the confounding variable that influenced the dependent variables of compliance, participation (independent variables – barriers, addressing the variables, interventions etc.)

- Patient's think that they are more compliant than they actually are (Llorente, et. al., 2008).
- Common barriers to adherence include: time management, competing priorities, not fully understanding disease process and benefits of treatment, depression and psychological factors, privacy and independence, transfer of responsibilities (Dziuban, Saab-Abazeed, Chaudhry, Streetman, & Nasr, 2010)
- Addressing identified barriers directly with the patient increases compliance (Sawicki, Heller, Demars, & Robinson, 2014).
- Age and disease severity increases non-compliance, suggesting that education beginning early on and throughout is needed (Hilliard, Eakin, Borrelli, Green, & Riekert, 2015).
- Educating patients and families helps to facilitate the transition of the CF patient into independent care as young adults (Goodfellow, et al, 2015).
- Education and shared decision making improves satisfaction and treatment adherence. Improved partnerships between patient and multidisciplinary team encourages exchange of information, which allows children to make meaningful contributions to their health and health care decisions; hence promotion of treatment compliance (Malone, Biggar, Javadpour, Edworthy, Sheaf, & Coyne, 2017).
- There is a higher rate of compliance with personalized care plans (Weiland, Schoettker, Byczkowski, Britto, Pandzik, & Kotagal, 2003).
- Treatment compliance is vital to the management and outcomes of CF patients. It is clear that health care professionals need to be knowledgeable about barriers to compliance. Studies suggest that identifying patient barriers and motivating patients to remain compliant does improve overall outcomes for CF patients, and has the potential to increase their lifespan (Santer, Ring, Yardley, Geraghty, & Wyke, 2014).
- Studies have also found that comprehensive, multidisciplinary inpatient programs have a positive impact on patients with CF, such as improved pulmonary function tests, improved BMI, and a decrease in repeat admissions (Alison, Donnelly, Lennon, et al, 1994).

### **Practice Recommendations:**

After a comprehensive review of the literature, it is evident that patients with cystic fibrosis would benefit from an individualized patient care approach that identifies what barriers they are specifically experiencing and focuses multidisciplinary efforts to minimize these barriers to support treatment plan adherence. Our practice recommendations therefore include developing an inpatient cystic fibrosis incentive program, utilizing a multidisciplinary approach to partner with adolescents CF patients and their family to:

- Develop a tool to identify patient-specific barriers



- Implement an individualized plan of care that addresses identified barriers
- Create a personalized schedule utilizing shared decision making (SDM) to promote adherence
- Implement a rewards system based on patient designated goals and compliance. Patients to receive a piggy bank in which they can collect tickets as they complete treatments on time and correctly. Tickets earned will be tallied at time of discharge and a gift card will be given based on goals. Our goal is behavior modification that will be carried on after a patient is discharged, the patient's reward is an incentive to be compliant while inpatient and to motivate positive behavioral change when it comes to treatment compliance in daily life.
- Provide education to the patient and families on the disease and disease process, developments, and resources.
- Develop and educate multidisciplinary team including (but not limited to): Nursing, Respiratory Therapy, Pulmonologists, Hospitalists, Physical Therapy, Nutrition, and Psychology.
  - Educate about the program and process
  - Educate on the evidence regarding motivation and rewards

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