

The Psychosocial Effects of Cystic Fibrosis

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Abstract

Cystic Fibrosis (CF) is a chronic genetic multi-systemic disease that affects water and chloride transport. As a result, mucous production is increased which leads to a mucous build up in organs, airway obstruction, reduced digestive function, and vulnerability to infections. Most patients with CF are diagnosed by 2 years of age, and the current median predicted survival rate is 37.4 years old, with 95% of patients dying from complications related to pulmonary infection.³ This disease can impact the psychosocial development of those who suffer from CF. This paper explores the psychosocial development and adjustments of those living with CF.

Keywords: Cystic Fibrosis, adherence, quality of life, coping, morbidity, mortality

Abbreviations: CF = Cystic Fibrosis, CPT = Chest Physical Therapy, ACT = Airway-Clearance Therapy, ACP = Advance Care Planning, ESLD = End Stage Lung Disease, CFRD = cystic fibrosis related diabetes

Introduction

At this time, there is no cure for CF; however, there are preventative treatments that help reduce the chance of premature death. These treatments include special diets, daily medication therapy, daily chest physical therapy (CPT), daily airway clearance therapy (ACT), and routine doctors' checkups and tests. CF is an exhausting and time consuming disease that can interfere or impair typical daily life activities and social interactions. As a result, interactions and relationships with members of the community, intimate partners, friends, and family can be strained. Dealing with chronic illness and societal pressures can lead to stress, anxiety, and depression. There is a high prevalence of psychiatric disorders and distress in individuals with chronic physical illness.¹ There is evidence of CF affecting the psychosocial development of individuals throughout their lifespan.

Early Childhood

From birth to age 6, children with CF undergo a change in lifestyle to accommodate survival. During this time, children begin to learn and form an identity through psychosocial development. CF has a direct influence on a child's psychosocial development and health. During the preschool period, developmental issues related to expanding language skills, cognitive development in areas such as understanding causality and assessment of ability to control environment, and emotional/behavioral regulation development may be of particular relevance to the experience of CF.³

In the first 2 years, children develop a sense of trust or mistrust and autonomy or shame/doubt. The parent-child relationship is a crucial context for the developing infant, and the quality of attachment predicts important child psychosocial factors such as emotional and behavioral regulations, social skills and the ability to cope with stress management.³ Many factors of CF and the treatment of CF affect whether a child will develop healthy forms of these psychosocial aspects. Early treatment of CF include CPT using percussion cups, ACT using nebulizers, and enzymes for digestion. A child struggles with developing autonomy for certain activities due to their dependence on the parent to provide care. They form trust, yet their trust can be questioned during the necessary treatments. Mistrust of parents can develop as a result of underdeveloped attachment from not being breast fed, being held down during CPT, being forced to inhale nebulized medications through a mask and ingest enzymes. In addition, the parent-child relationship can be damaged by parental stress. Parental distress can have negative implications for attachment, particularly when coupled with parental depression and anxiety.³

Between age 3-6, children continue their psychosocial development with the addition of initiative or guilt. During these ages, their understanding of their disease is improved, but limited. Many understand that they were born with the disease; however, few actually understand what the daily medications and treatments, or frequent doctors visits were for. This has a direct impact on the development of initiative. Mealtime and treatment time can become problematic as children begin to manifest behaviors that demonstrate a struggle to exercise choice and independence. Nearly half of the children with CF were found to have moderate to large sleep and/or eating problems, and 40% of the children had poor compliance with ACT.³ The conflict between parent and child over adherence of treatments and nutrition can have a negative effect on the emotional health of the parent and child. As parent levels of depression, anxiety, and stress become elevated, the child's emotional development and health is effected as well. During these ages, children struggle to comprehend the relationship between treatments and survival. Children between the ages of three and five years simply have no comprehension of the idea of death; they cannot conceived of lifelessness.¹² As a result, children have difficulty balancing their want for initiative through choice and independence and their need for treatment adherence.

It is important to aid children with CF in healthy psychosocial development. It is crucial to give accurate information about their illness according to their age and level of understanding. Maximizing a child's ability to cope with stress and anxiety is also important for the development of emotional health. Also, assisting families in maintaining a healthy family dynamic can enhance biological, emotional, and psychosocial health of the CF child and family.

School Age

From ages 7-11, children begin to evolve in physical, cognitive, emotional, and psychosocial development. At this time, psychosocial development of industry or inferiority will take shape. It is essential that children's sense of ownership and control over their chronic illness be

encouraged during this period in order to develop skills and self-efficacy related to self-management and collaboration throughout the lifespan.³ During this period, children have a better understanding of their illness and mortality. Thus, new stressors are introduced that ultimately influence the psychosocial health of a CF child.

At this age, children's autonomy improves as they strive to master essential life skills. Children also begin to develop industry or inferiority psychosocial identities. Children attempt to take more control in the decision of their care, but are limited by parental/physician requests. Ernst reported that diet consultants were focused toward the parent and emphasized weight gain, whereas children reported that their primary desired outcome for increased nutrition was more energy.³ This hinders a child's ability to develop industry as their opinions and desires are not included in their treatment plan. They can feel incompetent in managing aspects of their treatment.

At this stage in life, children learn to balance daily treatments with school, friends, and home life. Peer relationships are essential to psychosocial development and identity. Much of what is learned is through the process of comparison. For the child with CF, this peer comparison process may highlight their CF-related differences.³ Children begin to observe noticeable difference in physical size, endurance, eating habits, taking enzymes with food/drink, school achievement and attendance, and physical indicators of CF such as frequent coughing. Children begin to manifest signs of anxiety and stress as they make attempts to prove that they are not inferior to their peers. Negative peer reactions to the disease, such as teasing or over-protectiveness, tend to result in CF children suppressing coughs and keeping their illness a secret. Children with CF express concerns of 'being different' and have the desire to appear as 'normal' as possible. This effects a CF child's ability to develop close friendships with peers.

Children with CF struggle to develop industry when it relates to their treatment regimen. Disease management adherence tends to be low due to the increased demands of school, peer-related activities, and the lack of desire to comply. As a result, treatments are purposely skipped, forgotten, or neglected altogether. Earnst reported that only 51% of the recommended ACT completed, and of those completed, 64% lasted the recommended duration.³ Data indicated that enzyme and nebulizer medications were below 50%.³ Some indicators of the lack of adherence is attributed to forgetting, lack of time management, complexity of treatments, non compliance, disagreement with recommended treatment plans, and/or psychological issues. Low adherence interferes with the child's ability to master treatment adherence skills and develop a strong sense of industry.

There are ways to help CF children and families cope with the new stressors of the school-age years. Helping children prepare for social situations that may be awkward, can improve their confidence and enhance their ability to traverse through social settings. In addition, using behavioral methods to enhance adherence and track outcomes can increase children's sense of

involvement, control of self-efficacy, and essential components of self-management across the lifespan.³ Providing knowledge and support to school-aged children with CF will allow them to maintain healthy psychosocial development.

Adolescence

From ages 12-17, adolescents experience many physical, emotional, cognitive, and psychosocial changes. During this stage in life, it is difficult for many to navigate through these changes. As for an adolescent with CF, the difficulty is magnified. The interface between biopsychosocial development and disease can make this time period particularly challenging for the adolescent with CF.³ These new challenges help adolescents shape identity or role confusion.

Many societal issues impact whether an adolescent with CF will develop psychosocial health. Social isolation and reduced independence have a negative influence on forming an identity. CF interferes with maintaining academic achievement, sustaining energy, and participating in social activities. School time may be reduced due to disease management, lack of energy, a sense of 'not fitting in,' and socializing may also be affected by the need to avoid smoked-filled rooms, which exacerbate CF symptoms such as coughing.¹¹ In addition, adolescents with CF have to avoid certain social settings in order to maintain health. They have to avoid hot tubs, saunas, and other socially accepted areas in order to avoid exposure to harmful bacteria. Further social isolation occurs when an adolescent with CF does not participate in experimenting with drugs and alcohol with their peers.

Social isolation not only effects the development of social identity, but also the ability to form healthy sexual identity. Those with CF have difficulty forming relationships, not only with members of the same sex, but especially with those of the opposite sex. CF typically worsens as adolescents grow older. Puberty is delayed up to two years. The manifestations of symptoms are more evident which inhibit their ability to form intimate relationships. The outcome may be reduced social inclusion and hence effects on peer relationships and later sexual relationships, as well as potential vocational failure.¹¹ Anxiety and embarrassment begin to shape how an adolescent will act in social settings. Many adolescents suppress their cough in social settings, skip treatments while sleeping over at a friend's house, and refuse to take enzymes while eating with friends. The lack of adherence due to social pressures ultimately affects the overall health of the adolescent.

Future uncertainties also have an impact on the development of vocational or political identity. The adolescent's awareness of a limited lifespan influences their decision on adherence and future activities. For those who cannot continue to cope with the stress and requirement of disease management, coping mechanisms may break down, manifested by poor adherence, risk-taking, withdrawn from developmental tasks and depression.¹¹ As a result, role confusion can occur. Adolescents choose to live for now and not plan for their future. In a study conducted by

George, out of 25 participants ranging from 16-35 years of age, 36% had some high school or less, 8% had a high school diploma or GED, 8% attended vocational school or some college, 32% had a college degree, and 16% had a professional or graduate degree.⁴ It is evident that future plans for post secondary school is affected by CF. Marriage and a family can seem implausible. The adolescent struggles to gain full independence due to the nature of the CF disease.

As the adolescent strives to achieve autonomy, it is important that parents stay involved in the adherence of their adolescent's ACT and CPT treatments. The lack of adherence can interfere with the achievement of autonomy due to the fact that as their health declines, their dependence on their parents will increase. As health declines, identity achievement also suffers. Improved adherence and health increases survival rates and the opportunity to plan for future endeavors. Acceptance-based skills, such as learning to do treatment recommendations even though they are evasive or staying engaged with friends even when fatigued, may help adolescents with CF to achieve important developmental tasks and meaningful goals as they move into adulthood.³

Early Adulthood

From ages 18-25, the young adult with CF experience further growth in cognitive, emotional, and psychosocial development. The transition into young adulthood can reveal new stressors as the young adult gains more autonomy and industry, and begins to explore the psychosocial realms of intimacy or isolation. Some studies have suggested elevated levels of psychosocial impairment, including anxiety, depression, and eating disorders.¹ As a young adult, they have to learn to balance daily treatments with work, school, and social activities without the supportive promptings of their parents.

Adolescents face many challenges as they transition into young adulthood. First of all, they must change from pediatric care to adult care. This transfer can be stressful on many adolescents as they leave their 'comfort zone' and discover a new reality of their illness. The lack of money or the lack of disability allowances can overwhelm the young adult. To help in this transition, social workers need to review all documents to ensure that disability allowances are maintained. The financial burden of their illness can distress the young adult, however other aspects of the transition from pediatrics to adult care can add to the mounting stress. Perhaps, the most difficult aspect for the younger CF patient on being admitted to the adult ward is the sudden realization that significant numbers of the adult population are chronically unwell, needing a transplant, and each year, some, with whom they may have become good friends, will die.¹³ It is important that the young adult have support from their health team, friends, and family during this transition.

Many young adults find it difficult to comply with treatment adherence due to social pressures. Balancing time-intensive treatments with the demands of work, school, and social life proves to be a challenge for young adulthood. Out of a study of 24 adults ranging from 16-35, 64%

admitted that treatment burden reduced their medical adherence, 60% admitted that treatments were replaced by social events, and 60% admitted that work demands trumped treatments. “I work full-time, probably 50 hours a week right now, so wanting to have a ‘normal’ life, where you go out to get drinks with friends, go to spend time with friends, or go for a bike ride, or a run or do something to try to maintain a normal life. The most difficult part is fitting the treatments into that.”⁴ The key phrase is ‘wanting to have a normal life.’ This desire tends to be stronger than their desire to maintain a higher quality of life. The struggle to find a balance for treatments and social demands can increase stress for young adults.

During this stage in life, young adults look to find intimacy. CF tends to complicate relationships with those of the same sex and those of the opposite sex. Coffman studied 48 single patients with CF over 19 years of age and reported that the single female patients with cystic fibrosis evidenced several signs of social/sexual developmental delays, while the single male patients did not demonstrate delayed sexual developmental landmarks.² Coffman compared the sexual adaptation of those with CF to those without CF. The results reported that women with CF had a lower level of sexual desire, while men with CF were sexually healthy. There are many factors that contribute to the sexual health of a young adult with CF. Both men and women with CF feel unattractive. Typically, they point to their thinness, short stature, clubbed fingers, or stained teeth.² Not only does the physical aspects of CF inhibit the development of healthy intimacy, the daily routines of medical treatments take shape as an elephant in the room. “When I was married, I wouldn’t do my treatments around my wife...like my nebulizer, after I use it, I push it under my bed, I guess maybe I am hiding it...it is more my concern how they are going to react seeing their sick friend, sick husband, sick boyfriend.”⁴ George reported that out of 25 CF participants ranging from ages 16-35, 4% were with a partner, 8% were married, 16% were divorced, and 72% were single/never married.⁴ With the burden of living with CF, young adults tend to take comfort in isolation rather than intimacy.

Many young adults who have intimacy issues due to poor self image and low self esteem were aware of their illness and its impact on their life concerning social and sexual development. However, parental treatment of males and females with CF can also impact the young adult’s social and sexual development. Parents tend to overprotect their daughters resulting in inhibiting their sense of autonomy and self worth. It is important for family and friends to provide a healthy environment and support in respect to a young adults intimacy needs. Therapy can also provide help to those young adults who identify themselves as asexual.

Adulthood

From ages 26-65, each year of adulthood is considered a milestone. Many adults with CF face more troublesome manifestations of their illness. Cystic fibrosis related diabetes (CFRD) and end-stage lung disease (ESLD) can manifest in adolescence, young adulthood, or adulthood. The current age of death from CF is approximately 25 years old and the majority die from ESLD.³

With the sense of having cheated death, adults continue to learn and master the balancing act of medical adherence and social norms. The majority of adults with CF do not live to the age of 65; therefore they experience generativity or stagnation and integrity or despair earlier in life. During this stage in life, adults focus on careers, family, and preparation for death.

Adults with CF continue to struggle with the development of psychosocial aspects from their school-age years. Many have improved their abilities to trust, have autonomy, have initiative, be industrious, form an identity, and manage to have intimacy. As the survival rate increases for those with CF, they reach adulthood and disclosure of their medical status becomes more relevant in work and social settings. Most adults with CF have already disclosed their disease to their relatives and close friends. However, disclosing their illness in situations involving employment and dating can be complicated. Fewer patients reported disclosure to dating partners (73%), bosses/supervisors/teachers (51%), co-workers (39%), neighbors (25%), and acquaintances (20%).⁷ The severity of the adults' disease influenced whether they would disclose their illness or not. Those with a normal to mild case of CF were less likely to disclose their illness to people at work, than those with a moderate to severe case of CF. Overall, disclosure of having CF had a positive effect. However, reported negative effects were from intimate partners and bosses/teachers. Disclosure of the illness has the tendency to have a negative effect on achieving intimacy/marriage. As a result, it is difficult for a CF adult to achieve generativity. Those who do not have the opportunity to have a family and children participate in volunteer work with organizations related to CF. They are able to mentor parents and children who have been affected with CF.

Psychosocial health of a CF adult is dependent on the severity of their illness and how their health is maintained. The health of an adult declines, as they get older. If their health declines, they may become dependent on family or medical staff for care. At that time, it is important to provide the emotional support needed to help the CF adult with the transition. There is a direct correlation between an increase in emotional disturbance in adults with CF and the decline in health. It is important to note that considering such problems in our patients as psychological dysfunction may adversely affect their health.¹ The onset of depression is most prevalent with adults who lose independence. Consistent with previous studies, our study found that depressive symptoms are common among adults with CF, with 30% of participants exhibiting signs of depressive symptoms and 11% of participants screening positive for moderate-to-severe levels of depression.⁸ Social support systems and focusing on medical intervention will improve the physical and emotional health of the CF adult.

It is possible that an adult with CF has already confronted end-of-life issues several times throughout their lifetime. As an adult, it is important that they prepare for premature death especially if they have a spouse and children. Advanced care planning (ACP) is an important tool to promote alignment of the patient's care with his or her needs, goals, and values and is particularly useful if the patient's ability to make decisions becomes compromised.⁹ Creating an

ACP can help adults with CF achieve integrity and can come to accept the morbidity of their illness.

Depression therapy and periodic screening for depression and help improve the quality of life of CF adults. Honest communication maximizing provision of care options and emotional support, family involvement, attending to spiritual and psychological needs of patients and comprehensive symptom management are the cornerstones of end of life care.³

Conclusion

The psychosocial effects on those with CF vary. Experiences of anxiety, depression, and stress are evident throughout much their lifespan, but the degree of their distress and psychosocial health is highly influenced by the severity of their health condition and the support of friends, family, and community.

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