Overcoming psychosocial challenges in cystic fibrosis: Promoting resilience

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Abstract
Individuals living with cystic fibrosis (CF), and their families, have experienced significant improvements in treatment and related research that have enhanced outcomes and survival. Despite such advancement, the burden of living with CF still exists. Many psychosocial stressors and risk factors are associated with the impact of CF. The identification and treatment of such risk factors are discussed throughout this review, with an emphasis on strategies to address psychosocial risk and the importance of promoting resiliency in those touched by CF.

Keywords
chronic illness, coping, cystic fibrosis, quality of life, resilience

1 | INTRODUCTION

Individuals with cystic fibrosis (CF), their families and caregivers, and health care providers have witnessed several decades marked by tremendous improvements in survival. In 2015, a major population milestone was achieved when the number of people living with CF into adulthood in the United States exceeded the number of children. No longer is CF a health condition of importance to predominantly pediatricians and pediatric specialists. Rather, the messaging to families of a newborn diagnosed with CF now addresses the eventualty of adulthood, and living with CF is a mantra that clinicians have embraced over terminology such as “suffering from CF.” With these advances, however, remains the reality that living with CF involves significant psychosocial challenges, some unique to the experience of CF and others common to most chronic disease populations. In this review, we describe key psychosocial determinants of health in CF, describe psychosocial risk factors for worse health outcomes in CF, and outline potential strategies to address these challenges.

2 | COMMON PSYCHOSOCIAL CHALLENGES ACROSS THE CF LIFESPAN

From the time of CF diagnosis, families, and caregivers face psychosocial challenges that may impact health outcomes. Now that newborn screening is the primary avenue by which new CF diagnoses are made, many caregivers may receive the diagnosis before their child develops any clinical symptoms. Pre-symptomatic diagnosis has clear advantages in improving early life outcomes in CF, but may increase the emotional burden on parents and families. Positive newborn screening tests result in high levels of emotional distress in parents, particularly during the waiting period between receiving results and confirmatory sweat testing or evaluation in a CF clinic. Primary care pediatricians also identify challenges in newborn screening diagnoses, including lack of CF-specific training and low confidence. Current quality improvement efforts in CF newborn screening are focused on reducing parental and provider anxiety by streamlining referrals to CF centers and minimizing time from an initial positive newborn screening result to a confirmatory sweat test.

During early childhood, parents/caregivers of children with CF are tasked with promoting normal childhood development in the setting of chronic disease. This intersection can result in unique psychosocial challenges for family functioning. For instance, the focus on nutritional health in CF, particularly for infants and preschool children with CF, can augment existing challenges that parents have in terms of childhood eating patterns. Behavioral feeding challenges, including mealtime struggles, have been associated with poor weight gain in CF, and many nutritional interventions for children with CF have identified behavioral strategies and therapies as key drivers of improvement. In addition, parents of children with CF experience disease-related “milestones” that could impact family functioning, adherence to...
treatment recommendations, and psychological health of parents and children alike. These sentinel events include: initiation of new therapies (particularly daily nebulized therapies), initial onset of CF-related infections such as Pseudomonas aeruginosa leading to treatments aimed at eradication, or first hospitalizations, procedures, or surgeries for CF-related complications. As CF care centers have embraced the philosophy of early, aggressive monitoring to improve health outcomes across the lifespan, parents and children are more likely to experience such disease-related milestones earlier in life, providing a more constant reminder to a family that they are living with a chronic health condition. This realization can lead to parental or child stress, depression or anxiety. In response to such stressors, health outcomes and adherence can suffer; for example, a recent study linked parental depression with significantly higher rates of non-adherence to pancreatic enzyme therapy in pediatric patients with CF.6

Challenges in CF care tend to be magnified during adolescence, a high risk period for both physical and psychological health. During adolescence, lung function decline is often accelerated, and the frequency of CF pulmonary exacerbations and hospitalizations often increases.7 Treatment complexity and burden also tend to increase, leading to significant challenges with adherence.6 Multiple studies of adherence rates in CF have identified adolescence and young adulthood as the periods of lowest adherence.8 Adolescence is also a period of heightened developmental challenges, as youth are normally expected to develop independence from their parents. This can lead to tensions within a family over chronic disease self-management, as parents who have been in charge of daily treatments are asked to cede responsibility to their adolescent child. Adolescents may not be prepared for such independence, may develop anger at their health condition and caregivers, and may not recognize long-term consequences of ignoring healthy behaviors. Adolescents with CF also may not feel comfortable disclosing their diagnosis to peers, and this may result in social isolation.11 Depression and anxiety among adolescents with CF is known to be higher than the general population.9 All of these issues form the cornerstone for discussions during transition programs in CF, and need to be addressed through multi-disciplinary interventions throughout adolescence.

Many of the psychosocial challenges faced by adults with CF are similar to those experienced in childhood and adolescence. These include a high treatment burden, with estimates that routine CF care can take upwards of 2 h daily.12 Adherence to a complex, burdensome CF treatment regimen remains a concern throughout adulthood. Mental health co-morbidities such as depression and anxiety are common among adults with CF and may negatively impact respiratory and nutritional health outcomes.5 Unique to adulthood are some of the adult milestones that intersect with chronic disease. These include educational and vocational achievement, family planning, social support outside of the family, disease disclosure to peers and employers, and sexual and reproductive health concerns. As CF lung disease accelerates, symptom management, disability, and end-of-life decision making also augment the psychosocial burden of living with CF.

3 | PSYCHOSOCIAL RISK FACTORS THROUGHOUT THE LIFESPAN

Living and growing into adulthood with CF has become a reality for most people impacted by the illness. Yet, the symptoms and treatments required to maintain health have remained complex, time consuming and result in a debilitating effect on the daily life of the individual and often their family.12 Individuals typically benefit physically as a result of improvements in their psychological health.13 Given the heavy burden of CF, psychosocial functioning is an integral component of one’s overall health and wellbeing and has led to a need for increased understanding of the risk factors associated with living with CF. A clear understanding of these risk factors leads to a more sustainable approach to implementing interventions and strategies to overcome the identified risks.

3.1 | Mental health

Over the last decade there has been a better grasp of the increased prevalence of mental health symptoms in individuals living with chronic health conditions. As the link between mental health and physical health in those living with medical conditions has become clearer, so too has the risk of increased psychological symptoms and the negative impact on health and wellbeing. Original studies, meta-analyses and systematic reviews have all shown that adults and children with chronic conditions, as well as caregivers, are at a higher risk for depression and anxiety as compared to similar community samples.14–16 Several recent studies have measured psychological distress in individuals with CF and their caregivers, and symptoms of anxiety and depression have been shown to be 2-3 times higher than the general population.9,17–19

Detrimental mental health symptoms in both individuals living with CF and their caregivers have been associated with decreased lung function,20 lower body mass index,21 poorer adherence to medical care,19,22 worse health-related quality of life,23 more frequent hospitalizations and increased healthcare costs.21 The International Committee on Mental Health in CF (ICMH) has developed recommendations for the identification and treatment of mental health risk factors in CF Centers internationally.16 These recommendations include annual screening of depression and anxiety in adolescents and adults with CF, and at least one parent caregiver of children under 18 years, along with preventative psychoeducational support, formal assessment, and evidence-based psychological and pharmacological intervention when indicated. The CF Foundation (CFF) has formed a national steering committee to oversee implementation of the mental health guidelines and continue to identify the best practices for screening, education, and training of CF care team members.24 Further, inception of the CFF Mental Health Coordinator program has afforded numerous CF care centers the ability to enlist mental health professionals in their care teams. Arising from disciplines such as social work and psychology, CF care team mental health coordinators further the scope of management by performing guideline-based screening for depression and anxiety, and undertaking novel research and quality
improvement programs to improve mental health care delivery in CF. Still, further work is needed to better understand triggers for onset of mental health symptoms in people with CF and their caregivers, plus implementation of flexible and effective interventions to treat them.

3.2 Family functioning

CF is one of the most challenging pediatric illnesses for families to manage. As children become adolescents and move into adulthood, the environment in which they live is known to play an important role in their health outcomes and wellbeing. There has been a well-established association between family functioning and treatment adherence in many pediatric chronic conditions, including diabetes and asthma. While less work has been done in CF, there is encouraging evidence to suggest the same link exists between adherence and family functioning in CF. An exploratory, mixed-methods study to determine the level of family functioning specific to mealtime among families with a child with CF found results consistent with other studies, suggesting that family functioning was poor across almost all types of functioning except that which measures task-oriented types of behaviors. This finding suggests that families may feel required to be more structured in their interactions to complete all the necessary aspects of caring for their child with CF, at the expense of more emotional and social aspects of family functioning. In the study by Everhart and colleagues, lower levels of family functioning were present among families in lower SES categories. Socioeconomic status has been shown to be associated with parenting behaviors, psychological stress, and disease burden, which may all impact family functioning.

Family functioning has been found to be less of a risk factor for long-term health outcomes and well-being in CF as the child matures and grows into adulthood. It is important to incorporate family-centered care in the diagnosis and treatment of children with CF to reduce the long-term impact of poor family functioning across the developmental trajectory. Early family-based intervention, including the identification of significant parental mental health needs, during the toddler and preschool years may enhance family interactions over time and mitigate this risk.

3.3 Self-efficacy

Self-efficacy is defined as an individual's belief in his or her ability to perform the actions needed to accomplish a given task. Research has shown that self-efficacy beliefs are related to whether a person will attempt to perform a task and to how long he or she will persevere. This is relevant when considering the rigorous, daily and time-consuming aspects of care that are asked of those living with CF. Those individuals with low self-efficacy are less likely to tolerate the indefinite expectation of care and disruption in their life caused by CF. Self-efficacy is closely linked with self-management, defined as the necessary skills and activities an individual learns to control symptoms of a chronic disease and improve their quality of life. Self-efficacy has been directly linked to adherence to one's medical regimen, health behaviors, effective pain management, and chronic disease management in adolescents living with illness. Self-efficacy is highly correlated to self-management and the performance of routines and tasks to maintain health in CF.

Because it is possible to enhance a person's self-efficacy beliefs via a variety of behavioral interventions, it is important to reliably assess this construct to promote better self-management and resiliency. Self-efficacy beliefs are often measured by asking individuals to estimate their confidence in being able to perform specific behaviors across multiple contexts. For example, someone with CF may feel more confident in their ability to manage all aspects of their daily care during the weekend, but would judge less confidence in completing treatments during the week, when experiencing more stress and feeling busy with demands of work/school. Education and support of patients with CF should include an emphasis on promoting confidence, motivation, and problem solving barriers in order to maintain or improve self-management via one's sense of self-efficacy.

3.4 Social isolation

Individuals living with CF experience inherent social isolation due to an inability to receive in-person support and connection from others living with the disease. Infection control guidelines prohibit close (ie, less than six-foot distance) in-person contact between individuals with CF, posing a barrier for those living with the illness from the shared experience of developing relationships and interactions with others with CF. Thus social support, social connectedness, and validation of experience, known to be beneficial across many illness populations, is severely reduced in CF. Newer technologies such as internet-based forums, and social media, circumvent existing infection control barriers to social contact in CF, and may prove critically important as social isolation in people living with CF has been linked to an increase in many risk factors, including mental health problems. On the contrary, research across chronic illness populations has shown greater levels of social support are associated with fewer depressive symptoms, higher levels of self-efficacy, greater adherence to medical regimen, and better overall quality of life.

Adolescents living with CF have reported feelings of social isolation from peers, including feeling ostracized due to peers avoiding contact because of increased coughing and differences in physical appearance (eg, finger clubbing). Children and adolescents with CF experience frequent absences from school due to illness and hospitalizations, which makes it harder to establish friendships and increases feelings of loneliness and not fitting in. Individuals living with CF are also asked to avoid environmental situations (eg, environmental contaminants, hot tubs) that can cause increased health risks, further separating them from the common settings and experiences of peers. Addressing social isolation in individuals living with CF is critical. Social support experienced from friends, or others with a perceived ability to understand one's experience, is known to positively impact adherence to medical care and quality of life, Healthy development and functioning across the lifespan is characterized by social connection and perceived support by others.
As CF can disrupt this experience, reducing the isolation frequently experienced by individuals living with CF is critical.

4 | STRATEGIES TO PROMOTE RESILIENCE AND OVERCOME CHALLENGES

Efforts from the CFF targeting assurance of mental health guideline implementation for people with CF, partnering between patients and care teams, and research strategies for improving treatment adherence, are all aimed toward overcoming psychosocial challenges in CF. As described above, the addition of the mental health coordinator role in care teams is implicitly aimed toward improving screening for depression and anxiety in CF and embedding the treatment of comorbid psychosocial factors into the overall care of individuals with CF. However, the challenge of chronically addressing psychosocial challenges and mental health conditions in CF remains primarily in the hands of individuals and families with CF. Discussions in coping and strategies for promoting resilience in individuals and families with CF should be addressed in comprehensive CF care.

4.1 | Coping

Broadly, the coping mechanisms and styles employed by individuals living with chronic illness are believed to serve either as a buffer or risk factor for psychological problems and poor health-related quality of life.47–49 Passive or avoidant coping strategies (eg, disengagement, self-distraction, substance use) have been shown to be associated with increased risk of mental health symptoms and lower quality of life for individuals with CF.48 These coping styles result in efforts to divert attention away from the illness and promote poorer self-efficacy.50 In contrast, more active coping styles such as acceptance, seeking, and maintaining social support, and planning have all predicted better quality of life and improved self-management in adolescents and young adults with CF.48,51–52 These more adaptive coping styles have also been associated with other positive characteristics such as hope and optimism and an ability to relieve the risk for depression and anxiety, often implicated in psychological, and medical morbidity risk.52,53

4.2 | Promoting resilience

Resilience is defined by the American Psychological Association (APA) as the process of adapting well under adverse circumstances or significant stress.54 Because of the continuous need to confront uncertainty that comes with living with CF, many individuals experience emotion regulation strategies that contribute to resilience and acceptance of one’s health.49 However, as we describe, the psychosocial stressors of living with CF over an improved lifespan are increasingly complex. Strategies to promote resilience and disease management are vital to helping individuals navigate their disease burdens. In this section, we outline key strategies to promote resilience, and discuss how some of these strategies can benefit individuals with CF. By incorporating these strategies into comprehensive care, patients with CF and their families may experience improved quality of life and better disease management.

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<th>TABLE 1</th>
<th>Strategies to promote resilience</th>
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<td><strong>Strategy</strong></td>
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| Make connections | • Good relationships with close family members and friends are important.  
• Accept help and support from others.  
• Joining and being active in groups (faith-based, civic groups, etc.).  
• Help others in their time of need. |
| Avoid seeing crises as insurmountable problems | • Focus on what you can control.  
• You can change how you interpret and respond to stress.  
• Look to how things may improve in the future.  
• Acknowledge subtle ways you might already be dealing with stress. |
| Accept that change is a part of living | • Accepting circumstances that cannot be changed allows you to focus on things you can control. |
| Move toward your goals | • Develop realistic and measurable goals.  
• Do something regularly, even if it seems small.  
• “What is one thing I know I can accomplish today that helps me move in the direction I want to go?” |
| Take decisive actions | • Act on problems and stressors rather than detaching or wishing they would just go away. |
| Look for opportunities of self-discovery | • People learn from and find evidence of growth as a result of stress or loss (eg, better relationships, new sense of strength, increased self-worth, heightened appreciation for life, spirituality). |
| Nurture a positive view of yourself | • Develop confidence in your ability to solve problems.  
• Trust your instincts. |
| Keep things in perspective | • Even in the face of pain and significant stress, try to keep a long-term perspective. |
| Maintain a hopeful outlook | • Optimism allows you to expect good things will happen in your life.  
• Visualize what you want rather than worry about what you fear. |
| Take care of yourself | • Pay attention to your needs and feelings.  
• Engage in activities that you enjoy.  
• Exercise regularly. |

American Psychological Association. The Road to Resilience.  
self-management need to be considered for sustainment of health and quality of life in people living independently with CF. Promoting care-provider resilience is further important to ensure optimization of care for less autonomous patients with CF such as severely ill persons, children, and aging patients. Numerous methods for building resilience are recommended by the APA including: self-care, outlook modification, realistic goal setting, and decisive action, amongst others (See Table 1).54

Personal practices to improve health and resilience may also include techniques such as spiritual coping, meditation and/or meditative movement (eg, tai chi). Of these, spiritual coping has been most researched in CF. Spiritual beliefs can help provide a framework for making meaning of experiences, increase social support and sense of belonging, and can play an important role as a coping strategy to manage aspects of chronic illness.55,56 The presence of positive spiritual coping, such as praying to get better, is associated with better emotional functioning in youth with pulmonary and other disorders.57 Among adults with medical illness, positive spiritual coping has also been associated with better health outcomes, including improved postoperative cardiac functioning58 and lower rates of mortality.59 A study evaluating the relationship between spiritual coping and subsequent changes in pulmonary function, malnutrition, and hospitalizations over a 5-year period in adolescents with CF found that positive spiritual coping was associated with slower decline in pulmonary function, stable nutritional status, and fewer days hospitalized.55

Spiritual coping is believed to play an important role in buffering against the risk factors associated with living with CF and helps maintain long-term health and wellbeing. The impact of spiritual coping has been shown to extend beyond the patient and to parental caregivers as well. A qualitative study examining parental use of religious coping strategies in the first year after their child's illness found that a sample of parents of youth with CF used more positive religious coping styles (eg, meaning making, seek comfort/closeness with God, seeking spiritual support) than negative (eg, feeling punished by God).60 Spirituality has also been shown to be an important factor in treatment adherence in CF. Parents of children with CF who use negative spiritual coping have been found to be less adherent to their child’s prescribed care.61 An association between positive spiritual coping and better adherence to medications and treatments has been demonstrated in patients with HIV.62 Individuals with CF who report being more engaged in spiritual practices have demonstrated increased adherence to their medical regimen and more adaptive coping behaviors.63

While personal practices such as meditation and meditative movement have been researched in other chronic diseases, few studies have been conducted in CF. Mindfulness meditation has been well demonstrated to subjectively reduce stress, and recent studies demonstrate that mindfulness may further have a beneficial impact on decreasing inflammation. One recent randomized clinical trial of mindfulness meditation training versus general relaxation, in 35 community adults under the increased stress of job-seeking during unemployment, showed reduced levels of circulating levels of interleukin 6, a biomarker of systemic inflammation.64 Meditation, and meditative movement exercises (eg, tai chi, yoga) may provide patient-autonomous and safe methods for reducing stress, coping with psychosocial challenges, and promoting resilience.

4.3 | Overcoming challenges: Improving treatment adherence

Improving and sustaining treatment adherence remains a marked obstacle as individuals increasingly transition from pediatric to adult CF care, or as they strive to meet the demands of daily adult life while living with chronic disease. In 2014, the CFF Success with Therapies Research Consortium (STRC) was created not only to identify barriers in treatment adherence, but also to develop interventions improving the same. Currently, five diverse research studies supported by the STRC are underway. Broadly, the purposes of these STRC studies include: development and standardization of survey instruments to identify and characterize adherence barriers; validation of “smart” devices to track adherence (eg, electronically monitored medication bottles, nebulizer devices, and airway clearance vests); feasibility testing of video-conference based focus groups (thereby overcoming infection control limitations in CF); and, stakeholder engaged development of interventions such as nutrition focused web-based tools, tele-coaching sessions, and a web-based mobile application to augment self-management. No single best solution will exist to resolve the complex factors that constitute individuals’ psychosocial challenges or even barriers in treatment adherence. However, research studies, such as these, aim to ameliorate treatment burden and improve disease self-management.

5 | CONCLUSION

Individuals with CF and their families, along with care teams, are experiencing an ever-changing horizon, with improving survival. While there is much to be celebrated in this progress, there remains critical need to address psychosocial challenges and comorbid health complications including mental health concerns. Care providers and individuals with CF alike will benefit from strategies promoting resilience and improving treatment burden. Continued research should be focused toward overcoming psychosocial challenges in CF using stakeholder engagement, by developing sustainable interventions that may be disseminated into individual, care-provider, and care-team practices; and integrating effective preventative approaches to buffer against risk factors before they have impacted the health and wellbeing for those most impacted by CF.

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