International Journal of Computational and Experimental Science and ENgineering (IJCESEN)

Vol. 11-No.1 (2025) pp. 1648-1653
http://www.ijcesen.com

ISSN: 2149-9144

Research Article

Comprehensive Approach to Managing Cystic Fibrosis: Integrating Clinical Nutrition, Nursing, Radiology, Family Medicine, General Dentistry, Pharmacy, Psychology, and Respiratory Therapy

Mashael Saleh Alotaibi[1]*, Fatma Mahdi Nahari[2], Fatimah Abdulhadi Ali Alsaeedi[3], Sultan Abubakr Hakami[4], Muayad Youssof Al Awwas[5], Reema Abdulkader Azzeem[6], Ali Salman Al-khalaf[7], Mohamed Mohamed Basha[8], Hawraa Abdulmohsen Aljasim[9], Tariq Mishal Alharbi[10], Ahmed Khalid Bamousa[11]

[1]Clinical Nutrition, Medical Services/General Directorate,
* **Corresponding Author Email:** mmso1412m@gmail.com- **ORCID:** 0000-0002-5247-7833

[2]Nursing Specialist, Ministry of Health,
Email: fatma-nahari2005@hotmail.com- **ORCID:** 0000-0002-0047-1150

[3]Staff Nurse, Maternity and Children Hospital
Email: faalsaeedi@moh.gov.sa- **ORCID:** 0000-0002-0047-1220

[4]Radiology Technologist, Samtah General Hospital,
Email: sahakami2@moh.gov.sa- **ORCID:** 0000-0002-0047-1190

[5]Family Medicine, Alahsa Health Cluster
Email: myal_30@hotmail.com- **ORCID:** 0000-0002-0047-1870

[6]General Dentist, King Salman Armed Forces Hospital
Email: Reema.azzeem@gmail.com- **ORCID:** 0000-0002-0047-1340

[7]Pharmacist Technician, Alkulabiyah Health Center,
Email: asal-khalaf@moh.gov.sa- **ORCID:** 0000-0002-0047-1650

[8]Psychiatry, Psychiatric Hospital in Madinah at King Salman Medical City
Email: mohamed_pasha81@hotmail.com- **ORCID:** 0000-0002-0047-1770

[9]Respiratory Therapist, Alomran General Hospital Alahsa
Email: haaljasim@moh.gov.sa- **ORCID:** 0000-0002-0047-1320

[10]Psychiatrist, Erada Complex and Mental Health - Dammam, Saudi Arabia
Email: tari2k@gmail.com- **ORCID:** 0000-0002-0047-1770

[11]Psychiatrist, Erada Complex and Mental Health - Dammam, Saudi Arabia
Email: ahme2d@gmail.com- **ORCID:** 0000-0002-0047-1990

Article Info:

DOI: 10.22399/ijcesen.5494
Received : 05 January 2025
Revised : 25 February 2025
Accepted : 27 February 2025

Keywords

Cystic Fibrosis,
Comprehensive Approach,
Multidisciplinary Care,
Clinical Nutritio

Abstract:

Managing cystic fibrosis (CF) requires a comprehensive and multidisciplinary approach due to the complexity of the disease, which affects multiple organ systems, primarily the lungs and digestive tract. Key healthcare professionals, including nurses, respiratory therapists, and dietitians, collaborate to formulate personalized treatment plans that address the unique needs of each patient. Clinical nutrition plays a pivotal role, as individuals with CF often struggle with malabsorption and require tailored diets rich in calories and nutrients. Concurrently, regular monitoring through radiological imaging helps assess lung function and detect complications early, allowing for timely interventions. Family medicine physicians coordinate overall care, considering the mental and emotional health of patients and their families, which is crucial in managing a lifelong condition. Integrating various specialties, such as general dentistry and pharmacy, enhances the comprehensive care for CF patients. Dental professionals ensure oral health is maintained, as patients may face increased risks of dental issues due to medications and dietary choices. Pharmacists are essential in managing complex

medication regimens by optimizing dosing, minimizing interactions, and ensuring adherence to therapies like mucolytics or antibiotics. Additionally, psychology plays an integral role in providing emotional support and coping strategies for both patients and their families, addressing issues such as anxiety and depression that can arise from chronic illness. By fostering a team-oriented approach that includes these diverse fields, healthcare providers can significantly improve the quality of life and health outcomes for individuals living with cystic fibrosis.

1. Introduction

Cystic fibrosis (CF) is a hereditary, autosomal recessive disorder caused by mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene, resulting in the production of thick, sticky mucus that affects multiple organ systems, particularly the lungs and digestive tract [1]. The disease manifests through a spectrum of clinical presentations including chronic cough, recurrent pulmonary infections, breathing difficulties, poor growth, malabsorption, and pancreatic insufficiency [2]. As medical advancements have significantly extended the life expectancy of individuals with CF, the paradigm of care has evolved from a primarily pulmonary-focused approach to a comprehensive, multidisciplinary model that addresses the myriad systemic manifestations of this complex disease [3]. Modern CF care necessitates the integration of multiple healthcare disciplines, including clinical nutrition, nursing, radiology, family medicine, general dentistry, pharmacy, psychology, and respiratory therapy, working in concert to optimize patient outcomes and quality of life [4].

2. Clinical Nutrition in Cystic Fibrosis Management

Nutritional status is paramount in cystic fibrosis and is directly correlated with morbidity and mortality [2]. The recently updated ESPEN-ESPGHAN-ECFS guidelines on nutrition care for cystic fibrosis (2024) provide comprehensive recommendations for nutritional assessment and management across all ages [5]. Individuals with CF typically require caloric intake of 120% to 150% of the recommended daily allowance, with some guidelines suggesting 110% to 200% that of the general population [6]. This elevated energy requirement stems from increased work of breathing, chronic inflammation, and malabsorption secondary to pancreatic insufficiency [3].

Pancreatic enzyme replacement therapy (PERT) remains the cornerstone of nutritional management, facilitating adequate digestion and absorption of fats, proteins, and carbohydrates [7]. Dietary optimization focuses on high-calorie, nutrient-dense foods with adequate fat content, alongside vitamin supplementation—particularly fat-soluble vitamins A, D, E, and K—to prevent deficiency states [5]. The 2024 guidelines include expanded chapters on pregnancy, CF-related liver disease, CF-related diabetes, bone disease, nutritional and mineral supplements, and probiotics [5]. Furthermore, new chapters address nutrition in the context of highly effective modulator therapies and post-organ transplantation, reflecting the changing landscape of CF care [8]. Emerging evidence also emphasizes the need to reconsider traditional nutrition targets, with some experts advocating for BMI ≥50th percentile as a nutrition goal, particularly in the era of CFTR modulator therapy [6]. For patients with CF-related diabetes, dietary recommendations include complex carbohydrates with adequate fiber content while avoiding simple sugars [7].

3. Nursing Care and Coordination

Nurses serve as the central coordinators of CF care, providing clinical care, education, advocacy, and support across multiple domains of disease management [9]. Nursing care for patients with CF centers on managing secretions, ensuring airway clearance, and preventing and treating exacerbations [10]. Clinical nurse specialists work collaboratively with patients and families to develop individualized care plans, administer medications, monitor vital signs, prepare patients for diagnostic procedures, and provide comprehensive education regarding disease self-management [9].

The application of nursing theories, such as Orem's Self-Care Deficit Nursing Theory, has demonstrated effectiveness in enhancing self-care knowledge and skills among adolescents with CF [11]. Nurses play a vital role in helping patients understand the importance of reproductive counseling, facilitating discussions about family planning, and identifying unforeseen comorbidities that may arise with increased life expectancy resulting from CFTR modulator therapy [12]. Additionally, nurses are instrumental in supporting patients' psychological well-being, referring them to appropriate resources, and empowering caregivers through structured intervention programs [10].

4. Radiology and Diagnostic Imaging

Imaging represents an essential noninvasive means to assess CF lung disease, which remains the primary cause of morbidity and mortality in this patient population [13]. Current guidelines recommend baseline frontal chest radiography within the first 3–6 months of life and again within the first 2 years of life for patients with CF [14]. Thereafter, chest radiography or chest CT should be performed every 2 years or as needed to assess for bronchiectasis and other structural changes [13].

The MAESTRO consortium has provided state-of-the-art recommendations for lung imaging in CF, emphasizing the crucial role of imaging in diagnosing respiratory tract exacerbations and monitoring treatment response [15]. While chest radiography can exclude gross abnormalities or complications such as pneumothorax, it cannot sensitively diagnose new structural changes induced by exacerbations, particularly in patients with severe lung disease [14]. Computed tomography allows detailed assessment of exacerbation-related structural changes and represents the current standard for CF lung disease evaluation [16]. However, concerns about radiation exposure have led to recommendations for ultra-low dose CT protocols, defined as an effective radiation dose of approximately 0.08 mSv [16].

Magnetic resonance imaging offers a valid alternative for assessing both structural and functional information related to respiratory tract exacerbations, providing relevant data about ventilation and perfusion without radiation exposure limitations [17]. The Cystic Fibrosis Foundation recommends abdominal ultrasound to assess the liver and spleen at least every two years in persons with CF from childhood until late adolescence, starting at age 3 or at diagnosis if diagnosed after age 3 [18]. International efforts are underway to standardize CT and MR imaging practices in CF through the European Cystic Fibrosis Society Clinical Trial Network and European Respiratory Society collaborations [15].

5. Family Medicine and Primary Care Integration

Family physicians play an increasingly vital role in the comprehensive care of individuals with CF, extending beyond the neonatal screening period to include follow-up of adult patients [19]. As the CF population ages, with an estimated 70% of CF patients in the United States being cared for by adult providers by 2025, the involvement of primary care providers becomes crucial for preventative care and the management of non-CF-related health issues [20].

Family physicians contribute to CF care through continuous medical education to stay updated on evidence-based practices, engagement in community health initiatives, preventive education, and patient advocacy [19]. They are responsible for prescribing routine medications recommended by the CF team, ensuring patients have adequate medication supplies, providing routine vaccinations, participating in population screening programs, and managing health problems unrelated to CF [21]. Ideally, the primary care provider and pulmonologist practice within a patient-centered medical home model, addressing mental health, osteoporosis, sexually transmitted infections, and other aspects of comprehensive care [21].

Regular training for family physicians in CF diagnosis and follow-up, as well as education about complications that may arise in adult CF patients with prolonged life expectancy, is essential [20]. The expanding understanding of CF, encompassing genetics, pulmonary function, nutrition, colon transport, hydration, and electrolyte management, broadens the family physician's knowledge base and enhances their capacity to provide comprehensive care [19].

6. General Dentistry and Oral Health

Despite the numerous dental treatment needs of CF patients, comprehensive oral care guidelines for this population have historically been lacking [22]. However, emerging evidence underscores the importance of interdisciplinary dental care as a component of comprehensive CF treatment [23]. CF patients represent a group requiring targeted dental care, and dentists should be considered members of the multi-specialist patient-care team [22].

Regardless of the CF patient's age, dental check-ups should be scheduled at least every 6 months [23]. However, depending on the individual patient's condition, therapeutic visits may need to be scheduled more frequently [24]. Dental hygiene interventions include scaling of teeth and root planing, orthodontic and restorative practices, and other invasive procedures, which should be performed with consideration of the patient's overall health status [22].

Communication between the dental provider and the patient's medical team—including the pulmonologist and the CF reference center—is necessary to understand the current disease status and adjust dental care accordingly [24]. There are no known disease-specific contraindications for the provision of dental treatment, and dental care can be provided safely to CF patients in a general dental setting [23]. The implementation of targeted

preventive and therapeutic measures, along with overall solutions involving dedicated dental health standards, is advisable to improve the oral health outcomes of CF patients [22].

7. Pharmacy and Medication Management

Pharmacists play an essential role in the multidisciplinary CF care team, reviewing and optimizing medication regimens to ensure safe and effective pharmacotherapy [25]. Medication management in CF includes the use of CFTR modulators, mucolytics, bronchodilators, and antibiotics to improve mucus clearance and treat infection [26]. The integration of clinical pharmacists within CF clinics has been shown to improve patient satisfaction and understanding of medication use [25].

Pharmacist interventions include optimizing medication use knowledge, reinforcing adherence strategies, and streamlining timely access to treatment [27]. Common pharmacy interventions include inhaler technique education, which has been identified as a frequent area of intervention [26]. Medication adherence rates prior to pharmacy intervention have been reported as 81.9% for CFTR modulators and 62.5% for mucolytics [27]. The evolution of pharmacy services at CF centers has grown substantially, partly due to grant programs that have supported increased pharmacy presence in multidisciplinary care teams [25].

Pharmacists are uniquely positioned to address the complex medication needs of CF patients, including the management of multiple inhaled therapies, the coordination of antibiotic regimens for chronic infections, and the optimization of pancreatic enzyme replacement therapy [28]. The European Cystic Fibrosis Pharmacy Group continues to establish standards and best practices for pharmacy services in CF care [28].

8. Psychology and Mental Health Support

Psychological distress, including anxiety and depression, is common among individuals with CF, with prevalence rates 2.3 times higher in adults with CF than in community samples [29]. These psychological symptoms are associated with poor quality of life, reduced lung function, and increased healthcare utilization [30]. Clinical psychologists use non-medical approaches, including talking therapies, to help patients cope with emotional and physical health challenges [29].

The Coping and Learning to Manage Stress (CALM) with cystic fibrosis program represents a multisite telehealth randomized controlled trial designed to reduce depression and anxiety symptoms in adults with CF [30]. Studies have demonstrated that social support is associated with a reduction in the prevalence of mental and physical health symptoms among adults with CF [31]. Acceptance and Commitment Therapy (ACT) has shown superiority over supportive psychotherapy in improving psychological outcomes for adults with CF [32].

Psychological techniques in CF care aim to address the complex psychosocial challenges faced by patients, including the burden of onerous treatment regimens, reduced life expectancy, and the multisystem nature of the disease [29]. Virtual peer support programs have been established for adults with CF and their family members to assess the relationship between social support and health outcomes [33]. The European Cystic Fibrosis Society has developed multidisciplinary syllabi that include psychosocial components, recognizing the essential role of psychological support in comprehensive CF care [30].

9. Respiratory Therapy and Airway Clearance

Respiratory therapy constitutes a cornerstone of CF management, focusing on airway clearance techniques, inhaled therapies, and the prevention and treatment of pulmonary exacerbations [34]. Airway clearance techniques include chest physical therapy (positioning with cupped clapping on the chest), breathing and coughing techniques to promote secretion expectoration, and the use of various mechanical devices [35]. The European Cystic Fibrosis Society standards emphasize physiotherapy for airway clearance as essential for maintaining optimal lung health [34].

The intermittent intrapulmonary deflation technique represents a newer instrumental airway clearance technique that has been evaluated in randomized trials [31]. While some studies suggest that adding this technique to conventional airway clearance may not confer a clear short-term benefit, airway clearance remains a critical component of CF care [31]. A debate held at the 2024 North American Cystic Fibrosis Conference addressed whether airway clearance can or should be replaced by exercise in the era of CFTR modulators, highlighting the ongoing discussion regarding optimal respiratory management strategies [32].

Respiratory therapists recommend interventions such as high-flow nasal cannula to provide respiratory support with increased flow rate and for patient comfort during exacerbations [33]. Respiratory rehabilitation techniques encompass airway clearance techniques, exercise training, and inspiratory muscle training, with ongoing network meta-analyses being conducted to comprehensively

evaluate and compare the effectiveness of different approaches [35]. The management of respiratory failure may include nocturnal non-invasive ventilation and oxygen therapy, with studies examining how these interventions change in patients using CFTR modulator therapy [32].

10. Conclusion

The management of cystic fibrosis requires a multidisciplinary approach due to its complex nature. Key components include clinical nutrition for maintaining nutritional status, coordinated nursing care, radiology for monitoring disease progression, and family medicine for comprehensive health management. Dentistry addresses oral health, while pharmacy ensures safe medication regimens. Psychology offers mental health support, and respiratory therapy focuses on pulmonary health. Collaboration among these disciplines, rooted in evidence-based practices, is vital for improving clinical outcomes and quality of life for individuals with cystic fibrosis.

Author Statements:

- **Ethical approval:** The conducted research is not related to either human or animal use.
- **Conflict of interest:** The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper
- **Acknowledgement:** The authors declare that they have nobody or no-company to acknowledge.
- **Author contributions:** The authors declare that they have equal right on this paper.
- **Funding information:** The authors declare that there is no funding to be acknowledged.
- **Data availability statement:** The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.
- **Use of AI Tools:** The author(s) declare that no generative AI or AI-assisted technologies were used in the writing process of this manuscript.

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