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Nutrition: Prevention and management of nutritional failure in Cystic Fibrosis

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Abstract

Close monitoring of nutritional status is critical to the overall health of a patient with CF. As part of routine CF care, measurement of weight and height (and calculation of weight/length or BMI as appropriate) should be performed and analyzed at each visit. Early recognition of nutritional risk is imperative and evaluation with a multidisciplinary team should be performed to assess for caloric intake, caloric malabsorption, and other causes of poor weight gain and growth. Many tools are available to use for intervention, including oral supplementation, behavioral interventions, medications, nutritional therapies, and enteral tube feeding.

Keywords: Nutrition; Cystic fibrosis; Fat soluble vitamins; Enteral feeding

1. Background

Close monitoring of nutrition and growth is essential in caring for pediatric and adult patients with Cystic Fibrosis (CF) [1]. The Cystic Fibrosis Foundation (CFF) recommends that nutritional status be monitored as part of routine CF care, and that both children and adults with CF achieve nutritional status comparable to healthy children and adults, as optimal nutritional status is associated with better clinical outcomes [2,3]. Despite this knowledge, optimal nutritional status may be difficult to achieve and maintain.

Nutritional status may assessed by weight, height, weight-for-length percentiles (in children <2 years of age), and Body Mass Index (BMI; used in children ≥2 years of age and adults). The CFF recommends performing these parameters every month during the first year of life and then every three months as part of routine CF care, for patients meeting nutritional goals [1]. The frequency of nutritional assessments should increase in clinical scenarios where nutritional status is suboptimal (to every 2 weeks in infants and every 6–8 weeks in children >2 years of age and adults) [1]. Recent guidelines from the European Society for Clinical Nutrition and Metabolism (ESPEN) discuss limitations of using only weight, length, and BMI or weight-for-length, and suggest supplemental methods to analyze body composition including dual-energy X-ray absorptiometry (DEXA), anthropometrics, and bioelectrical impedence for all patients with CF [4]. Specifically, DEXA is recommended for all patients from 8 to 10 years of age [4].

Focusing on and achieving optimal nutritional status early in life is important. Identifying children with CF early in life through newborn screening leads to improved nutritional status, improved growth, fewer pulmonary exacerbations, and fewer hospital days [5–7]. By age two, weight-for-length >50%ile correlates with improved lung function between ages six

Abbreviations: BMI, Body Mass Index; CDC, Centers for Disease Control and Prevention; CF, Cystic Fibrosis; CFF, Cystic Fibrosis Foundation; CFTR, cystic fibrosis transmembrane conductance regulator; DEXA, dual-energy X-ray absorptiometry; EFA, essential fatty acids; ESPEN, European Society for Clinical Nutrition and Metabolism; FDA, Food and Drug Administration; FEV, forced expiratory volume; GERD, gastroesophageal reflux disease; IBW, ideal body weight; WHO, World Health Organization.

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through fifteen [8], and children who recovered their birth weight z-score by two years of age had improved lung function at six years of age [9]. Weight-for-age percentile at four years of age has been associated with improved height and growth velocity as well as improved survival and fewer pulmonary exacerbations at age eighteen [2]. Continuing this effort into adulthood is essential, as higher BMI in adults with CF has been associated with improved pulmonary function [8].

Current recommendations for nutritional goals vary depending on the age of the patient. For infants and children younger than two years of age, achieving a weight-for-length percentile of ≥ 50% is recommended [8]. This recommendation was based on growth points recorded on the 2000 Centers for Disease Control and Prevention (CDC) growth charts [10]. However, since that recommendation, the American Academy of Pediatrics and the CDC have recommended the use of the 2006 World Health Organization (WHO) growth charts for routine assessment of nutritional and growth status for children less than two years of age [11]. These charts vary significantly, with the WHO growth charts demonstrating higher percentiles for the same weight compared with the CDC growth charts. As such, achieving a weight-for-length of 50th percentile using WHO standards was associated with a lower (but adequate) forced expiratory volume (FEV_1) compared with using CDC standards [12].

For children two years of age and older, achieving BMI percentile ≥ 50% using the CDC growth charts is recommended [8]. For adults older than 20 years of age, the CFF recommends attaining a BMI ≥ 22 kg/m^2 in females and attaining a BMI ≥ 23 kg/m^2 in males [8]. While these recommendations note the importance of achieving an adequate BMI, it is also important to evaluate the overall growth and nutritional status in children. A slowed growth velocity may result in a higher BMI despite the presence of suboptimal nutritional status, and monitoring BMI alone may result in overestimating nutritional status in stunted children [13]. In addition, height is related to lung volumes, and is an important factor to consider in the overall health of the child and adolescent with CF [14,15]. Lastly, the composition of body weight should be considered, as an increased Lean Body Mass Index (LBMI) is more closely associated with improved pulmonary function than BMI, particularly in undernourished children [16]. Thus, achieving a goal BMI by gaining muscle mass (and not by gaining visceral fat mass) is important.

The CFF has published several guidelines with critical information and guidance on the diagnosis and management of malnutrition. In 2002, the CFF and the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition, convened a team of experts to determine guidelines for the timing and composition of nutritional assessments in children with CF, including anthropometric measurements and macro- and micronutrient requirements, the evaluation of patients with nutritional failure, and strategies for improving nutritional status [1]. In 2008, the CFF revised these recommendations to improve classification of nutritional risk through analysis of the CFF Patient Registry and reviewing current literature [8]. The 2009 infant care guidelines, developed by the CFF, recommend increasing caloric intake, considering micronutrient deficiencies, and using behavioral interventions to improve weight gain in infants and young children with suboptimal nutritional status [17]. In 2016, clinical practice guidelines for preschoolers with CF were developed through the CFF [18]. While these recommendations inform the management of pulmonary disease, much of the recommendations are focused on nutrition and gastrointestinal complications of CF. Specifically, these guidelines recommend an algorithm for the management of nutritional status (including anthropometric assessment, determination of nutritional risk, further evaluation for co-morbidities, and treatment of nutritional failure). Most recently, also in 2016, evidence-informed guidelines for the use of enteral tube feedings in children and adults with CF were published, with emphasis on indications and contraindications for placement of feeding devices, perioperative management of feeding devices, and long-term follow up for enteral devices [19]. Internationally, the Dietitians Association of Australia National Cystic Fibrosis Interest Group published Australasian Clinical Practice Guidelines for Nutrition in Cystic Fibrosis [20], and ESPEN (with collaboration from the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition and the European Cystic Fibrosis Society) published guidelines in 2016 on nutrition care for pediatric and adult patients with CF [4].

2. Clinical presentation and differential diagnosis

In the 2002 consensus report on nutrition for pediatric patients, nutritional failure was defined as height <5%ile for age, ideal body weight (IBW) <90%ile for age, weight-for-length <10%ile for age (in children and infants younger than two years of age), and BMI <10%ile for age (in children two years and older) [1]. At-risk nutritional status was defined as height %ile less than genetic potential, IBW 10–25%ile for age, weight-for-length 10–25%ile for age (in children and infants younger than 2 years of age), and BMI 10–25%ile for age (in children two years and older) [1]. Based on evidence linking BMI and FEV_1, nutritional guidelines were updated in 2008 [8]. In addition to these BMI goals, the most recent nutritional guidelines for preschoolers with CF recommend attaining a weight for-age ≥ 10%ile, accounting for evidence that achieving weight ≥ 10%ile at age four is associated with improved survival at age 18 [2,18]. Table 1 summarizes current CFF recommendations for nutritional goals.

Patients with CF and their families should be educated about the importance of nutritional care throughout the life span of the patient with CF [19]. Reviewing diet history every 3–6 months is recommended [4]. However, when a patient is not able to attain the nutritional goals recommended by the CFF, interventions should begin early, and all options should be

Table 1
Summary of Nutritional Goals as Recommended by the Cystic Fibrosis Foundation [2,8,18].

Age	Guideline
<2 years	Weight-for-length ≥ 50%ile for age
2–5 years	BMI ≥ 50%ile for age
	Weight ≥ 10%ile
>5–18 years	BMI ≥ 50%ile for age
>18 years (female)	BMI ≥ 22 kg/m^2
>18 years (male)	BMI ≥ 23 kg/m^2