Nutrition and Growth in Cystic Fibrosis

Sarah Lusman, MD[a], Jillian Sullivan, MD, MSCS[b,*]

KEYWORDS

• Cystic fibrosis • Nutrition • Growth • Body mass index • Anthropometrics
• Enteral feeding • Fat-soluble vitamins

KEY POINTS

- Close monitoring of nutrition and growth is essential to the care of children with cystic fibrosis (CF). Growth and nutrition should be assessed at every visit to the CF care center.
- Body mass index (BMI) percentile is the most commonly used marker of nutritional status. Nutritional status is directly associated with pulmonary function.
- It is recommended that children with CF achieve growth and nutritional status comparable with that of well-nourished children without CF. Specifically, this means weight-for-length greater than the 50th percentile in children less than 2 years of age and BMI greater than the 50th percentile in children older than 2 years.
- Barriers to attaining and maintaining nutritional status include decreased caloric intake, gastrointestinal dysfunction, increased caloric expenditure, and psychosocial issues.
- Methods used to optimize nutritional status include intensive dietary and behavioral counseling, use of oral supplements, and enteral tube feedings.

IMPORTANCE OF ADEQUATE NUTRITION AND GROWTH

Close attention to nutrition and growth is integral to the care of patients with cystic fibrosis (CF).[1] The Cystic Fibrosis Foundation (CFF) recommends that children with CF achieve nutritional status comparable with that of healthy children. Growth and nutritional status should be assessed at every visit. Despite multiple advances in the digestive care of patients with CF, appropriate nutritional status remains difficult to attain. In this article the authors discuss the assessment of growth and nutrition, nutritional goals and challenges, and optimization of nutritional status.

Dr Dorothy Andersen wrote the original reference to CF as a new entity in 1938. She described CF causing early death in infants because of extreme malnutrition.

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[a] Department of Pediatrics, Columbia University Medical Center, 622 West 168th Street, PH 17 East 105L, New York, NY 10032, USA; [b] Department of Pediatrics, University of Vermont College of Medicine, 111 Colchester Avenue, Burlington, VT 05401, USA
* Corresponding author.
E-mail address: jillian.sullivan@uvmhealth.org

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Autopsies revealed destruction of the pancreas, and eventually the role of maldigestion as a cause of malnutrition in CF was understood. Dr Harry Shwachman encouraged patients to follow a low-fat or fat-free diet to minimize the symptoms of steatorrhea, resulting in worsening malnutrition and a characteristic physical appearance including a swollen abdomen, lack of subcutaneous fat and muscle mass, and overall wasting. Later, comparisons of 2 large CF programs with differing approaches to dietary fat restriction showed that those on a high-fat diet had better growth and survival contributing to the current recommendations for a high-calorie, fat-unrestricted diet.[2] Important advances in the nutritional management of patients with CF include the development of pancreatic enzyme replacement therapy, the availability of nutritional supplements, and the use of enteral tube feedings.[2,3]

Body mass index (BMI) and BMI percentile for age are important measures of nutritional status in children with CF. It is recommended that height and weight be measured and BMI calculated by dividing the weight in kilograms by the height in meters squared at least every 3 months (**Table 1**). The 2000 edition of the growth charts issued by the Centers for Disease Control and Prevention (CDC) should be used to compare each patient's BMI percentile to age- and sex-matched norms. The goal is a weight-for-length at or greater than the 50th percentile in children less than 2 years of age and a BMI at or greater than the 50th percentile for children older than 2 years, meaning that nutritional status is comparable with that of well-nourished healthy children. The rationale for this goal is that an association exists between lung function, generally measured by forced expiratory volume in 1 second (FEV_1) percent predicted, and nutritional status (**Fig. 1**).[2,4–6]

BMI is the most widely accepted measure of nutritional status in patients with CF. In the past, percentage of ideal body weight was often used; however, BMI is a more accurate measure of nutritional status.[7,8] Even so, it is essential to remember that some children with short stature may have a BMI at or greater than the 50th percentile but still have suboptimal nutritional status. In fact, it is thought that height is related to lung volume; therefore, the importance of achieving adequate linear growth cannot be overstated.[9,10]

The 2002 consensus report on nutrition for pediatric patients with CF[1] notes that extra attention should be paid to nutrition and growth at 3 specific times: the first 12 months after diagnosis, the first 12 months of life for infants diagnosed prenatally or at birth, and the peripubertal growth period. The establishment of a pattern of normal growth (ie, similar to children without CF) during early childhood sets the stage for continued growth for the remainder of childhood and adolescence. As more and more patients are diagnosed prenatally or at birth (see section on newborn screening and diagnosis), increasing emphasis is being placed on the care of toddlers and preschoolers. New guidelines that pertain to all aspects of care in this age group are forthcoming and are discussed later. In addition, recent research based on data from the CF Patient Registry has shown the importance of early childhood nutrition for pulmonary health and overall outcomes later in life. One important study demonstrated that greater weight at 4 years of age is associated with greater height, better pulmonary function, fewer complications of CF, and better survival through 18 years of age.[11]

ASSESSMENT OF GROWTH AND NUTRITIONAL STATUS

The frequency at which growth parameters should be assessed is described in **Table 1**. Growth is measured in children with CF by the same standards that are used for healthy children. For children less than 2 years of age, the World Health