



# THE SUPPLEMENT

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## WEIGHT AND CYSTIC FIBROSIS

The relationship of weight to overall well-being of individuals who have CF is well documented. From the time Andersen identified CF in 1938, [1] to the present, caregivers have been challenged to help patients optimize their weight. Although aggressive medical and nutritional care has resulted in improved weight, the median BMI percentile (BMI%) for children in the United States (US) remains less than the 50%. [2] This issue of The Supplement focuses on current knowledge of weight and CF.

### FEATURED PAPERS:

#### **Evidence-based practice recommendations for nutrition-related management of children and adults with cystic fibrosis and pancreatic insufficiency: Results of a systematic review.**

Stallings VA, Stark LJ, Robinson KA, Feranchak AP, Quinton H. Clinical Practice Guidelines on Growth and Nutrition Subcommittee, Ad Hoc Working Group. J Am Diet Assoc 2008;108:832-39. Besides other aspects of nutrition, this paper includes the recommendations of the Ad Hoc Working Group on weight and growth.

**Objective:** To update recommendations for weight and growth monitoring. **Subjects:** Data of 22,700 pancreatic insufficient (PI) patients from the 2005 US CF Foundation (CFF) Patient Registry. **Methods:** Review of available literature and analyses of registry data to determine the association between FEV<sub>1</sub>% predicted with weight-for-stature. **Results:** The BMI% method had a stronger association with FEV<sub>1</sub>% predicted and was more sensitive with changes in FEV<sub>1</sub>% predicted when compared to percent ideal body weight (%IBW). **Conclusions:** BMI% for children and adolescents and BMI for adults are the recommended assessment methods.

Recommendations for optimal weight:length, BMI%, and BMI are provided. (See Review below.)

**Improved pulmonary and growth outcomes in cystic fibrosis by newborn screening.** Collins MS, Abbott MA, Wakefield DB, Lapin CD, Drapeau G, Hopfer SM, Greenstein RM, Cloutier MM. Pediatr Pulmonol 2008;43:648-55. **Objective:** To determine if newborn screening (NBS) results in improved longitudinal growth and maintenance of normal

pulmonary function. **Subjects:** 55 children diagnosed with CF between 1983 and 1997. By NBS: 34; 18 male; 28 PI. By traditional methods: 21; 12 male; 18 PI. **Methods:** Growth and pulmonary function data were compared for those diagnosed by NBS to those diagnosed by sweat test after symptom appearance.

**Weight and Growth Results:** Patients identified through NBS demonstrated greater weight and height for age at diagnosis and through 15 years of age.

**Weight and Growth Conclusions:** Those diagnosed by NBS had improved weight and growth.

#### **Validation of a nutrition risk screening tool for children and adolescents with cystic fibrosis ages 2-20 years.**

McDonald CM. J Pediatr Gastroenterol Nutr 2008;46:438-46. **Objectives:** To validate a nutrition risk screening tool to identify children with CF who may benefit from more intensive nutrition intervention. **Methods:** A screening tool was developed based upon CF and non-CF data describing weight and height, as well as the rate of weight and height gain. The tool was field tested by US CF Center RDs and modified based on their comments. Two tests of sensitivity and specificity were performed. **Results:** The two tests indicated a satisfactory level of reliability. **Conclusions:** Based on this study, the tool may be a useful component of nutrition screening.

### SPECIAL POINTS OF INTEREST:

- *Newborn screening allows diagnosis and treatment prior to the occurrence of failure to thrive and results in better weight and height attainment.*
- *Weight loss, or gain, is best evaluated within the context of the patient's overall health status, including weight and height history.*
- *Weight is only one component of a comprehensive nutrition assessment.*

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## REVIEW

This review focuses on three areas related to weight and CF: 1. Evolution of knowledge about the importance of an individual's actual weight attainment and about weight calculation methods; 2. Impact of NBS on weight at diagnosis and beyond; and 3. Recent recognition of the occurrence of overweight and obesity in people who have CF. Inadequate rates of weight gain and weight attainment, along with growth failure in children, are unfavorable prognostic indicators in CF. [3,4,5] Therefore, the issue of weight has been, and continues to be, a major focus in CF.

Initial descriptions of people with CF highlighted thin, frail children who often succumbed very early in life due to both pulmonary and gastrointestinal complications. [1,6] In 1988, Corey, et al. [7] published a landmark paper pivotal to the topic of weight and nutritional management. By comparing the survival rates of patients in CF Centers in Canada and the US and with a focus on the CF Centers in Toronto and Boston, these investigators noted major differences. Although patients at these two centers had similar FEV<sub>1</sub> scores, the Toronto patients had a better survival rate, were encouraged to consume a high fat, high calorie diet, and were prescribed as many nonenteric-coated pancreatic enzyme replacement capsules (enteric not yet available) as needed to control symptoms of fat malabsorption. The authors postulated that, "It is possible that the height and weight differences presented in this report are the result of these different treatment regimens, and that improved growth and nutrition have contributed to better survival in the Toronto patients." In 1999, Lai, et al. revisited the question of weight and growth status of patients with CF in the US and Canada and found substantially smaller differences in life expectancy. The authors hypothesized the results might be related to the improved weight of patients due to changes in dietary advice and the development and use of enteric-coated enzymes. [8]

To address the need for optimal nutrition management, in 1991, the CFF convened a Consensus Committee to develop nutrition care guidelines. The Committee's report [9] acknowledged weight as a crucial component in the overall care of persons who have CF. The Committee recommended the use of %IBW as the method of categorizing weight. Ten years later, in 2001, the CFF revisited nutrition care. [10] Height or %IBW or BMI% were used as evaluation methods to classify children's nutritional status as acceptable, at-risk, or in failure. For adults, in 2004, the CFF Adult Consensus Committee recommended using %IBW or BMI. [11]

In other parts of the world, CF weight policies were established. The European Nutrition Consensus Report [12] recommended the use of %IBW for children and BMI for adults. The authors of the Australasian Nutrition Guidelines, 2005, recommended using both %IBW and BMI% for children and BMI for adults, with the caveat that %IBW may not be the best method of assessing weight adequacy. [13]

Subsequent research demonstrated that using %IBW as a weight evaluation method for children and adults to be problematic. Zhang, et al. observed that %IBW was an invalid marker because the "basis for determining the ideal weight for age is flawed." [14] Later, Lai [15] reported that the %IBW for children underestimates the severity of underweight in short patients, and overestimates it in tall patients. Importantly, %IBW and BMI% are not equivalent. A child may be identified as underweight by one method and not the other. Analysis of data from the German CF Quality Assurance Project, revealed similar discrepancies. [16] For adults, using the Metropolitan Life Insurance reference weights overestimates the severity of underweight. [15]

To address these and other challenges, the CFF convened an Ad Hoc Working Group to provide weight recommendations. [Stallings, featured paper] The Group recommended: 1. Infants: Reach a weight-for-length of  $\geq 50$  percentile by 2 years of age; 2. Children and adolescents (2 to 20 years of age): Use BMI% and maintain a BMI%  $\geq 50$ th percentile; and 3. Adults (20 to 40 years of age): Maintain a BMI  $\geq 22$  for women and  $\geq 23$  for men. Based on work by Lai and Shoff, discontinuation %IBW and using BMI% alone resulted in changes in individual pediatric CF Center ranking in the CFF Registry and a 6% reduction in the prevalence of "nutritional failure" as defined by the 2002 CFF guidelines. [17]

Newborn screening has allowed for the diagnosis and treatment of babies with CF prior to the development of the hallmark CF signs and symptoms, including malabsorption with subsequent failure to thrive. [18] Ongoing data from the Wisconsin newborn screening program, started in 1984, has provided evidence that, when compared to traditional diagnostic methods, early diagnosis through NBS prevents failure to thrive. [19] Additionally, both the weight and height of these patients identified through NBS remained higher into adolescence. [20, Collins, featured paper]

Overweight and obesity associated with CF appear to be rare, but do occur. With the epidemic of overweight and obesity in the general public, Kastner-Cole, et al.

## REVIEW (CONT.)

[21] questioned the prevalence of obesity in the United Kingdom's CF population. Of the 2,987 subjects in the UK CF registry database, 9.1% were considered overweight and 1.1% obese. The authors suggested that these conditions might result from intensive clinical interventions, genetic background, and changes in eating and exercise habits common in the general public. In adults diagnosed with CF as adults, overweight and obesity perhaps may be attributed to

milder CF genotypes and lower incidences of PI. [22,23,24] Unknown in children and adults who have CF are the occurrences of overweight and obesity associated comorbidities, such as pulmonary (e.g., sleep apnea, asthma), endocrine (e.g., type 2 diabetes), and cardiovascular (e.g., hypertension). [21] For adults with any pulmonary disease seeking lung transplant, severe obesity (defined as a BMI >30) is considered a relative contraindication. [25]

## CLINICAL APPLICATIONS

RDs are aware that weight is only one component of a comprehensive nutrition assessment. [26] In addition to measures of weight and height, the diagnosis of malnutrition is based on full array of anthropometric, laboratory, clinical, and dietary data. As the authors of the Australasian Practice Guidelines so eloquently stated, "Growth charts, changes over time and BMI% (%IBW in children <2 years) should be used together to form an anthropometric-based clinical decision regarding the nutritional status of children who have CF." [13] Chronic underweight may impact growth potential, with negative impact on genetic potential for height and is considered an independent risk factor for poor survival. [5]

Rates of gain for both weight and height are crucial when assessing adequacy of growth. As with children with a BMI <50%, children with a BMI ≥50%, may require nutrition intervention if rates of weight or height gain are inappropriate. McDonald's [featured paper] method to classify the growth of children is a clinical tool for RDs to use in prioritizing care specific to weight and/or height. For adults, it is suggested that unintentional weight loss be evaluated in the context of

the patient's usual weight and health status. [Stallings, featured paper]

Adolescent girls and women may be of particular concern for the RD. Similar to the general population, teen girls and women who have CF may prefer to be slender. [27, 28] In their study group, Kastner-Cole, et al. noted a near absence of obesity in women, which may be related to overall concern for body size often seen in females. [21]

In summary: 1. Optimal weight is critical in the continued well-being of persons who have CF; 2. Weight is best evaluated within the context of the patient's overall health, weight, and height history; 3. Weight assessment is dependent upon accurate measurement and, for infants, children, and adolescents, accurate plotting on growth charts; 4. Weight is not a surrogate for the assessment of body composition; 5. Weight does not supplant a full nutrition assessment, and should not be the sole determinant of nutritional status classification. The RD is pivotal in guiding individuals to reach and maintain their optimal weight through a healthy diet. A good rule of thumb for RDs might be: Start weight intervention early, and often.

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