Nutritional assessment and management in cystic fibrosis: a consensus report1,2

Bonnie W Ramsey, Philip M Farrell, Paul Pencharz, and the Consensus Committee3

ABSTRACT This report is a summary of a meeting convened by the Cystic Fibrosis Foundation to develop a consensus among nutrition specialists and cystic fibrosis care givers regarding optimal nutritional management of patients with cystic fibrosis. The first section of the report provides a rationale for emphasizing nutritional management of this genetic disorder. The multiple factors causing malnutrition and a negative energy balance are outlined. The second section provides guidelines for routine assessment of nutrition in these patients. Five categories of nutritional status are defined based on ideal weight for height, age, and gender. These categories are used to formulate a graded response for nutritional intervention. Recommendations are provided for routine dietary supplements, vitamin supplements, and pancreatic enzyme replacement. The primary aim of this report is to educate clinicians as to the importance of frequent assessments and early intervention. Am J Clin Nutr 1992;55:108–16.

KEY WORDS Nutrition, cystic fibrosis, malnutrition, pancreatic insufficiency, consensus report

Background

The importance of nutritional status in long-term survival and well-being of patients with cystic fibrosis (CF) is well documented (1, 2). There are multiple interrelated factors that affect the nutritional status of patients with this disorder, many of which are still not fully understood. Recent studies have supported the concept that genetic factors have an influence on the presence or absence of pancreatic insufficiency (PI) (2). Patients with PI and steatorrhea (based on a 3-d stool fat-balance study) as well as associated undernutrition have a worse prognosis in terms of growth, pulmonary function, and long-term survival than do pancreatic-sufficient patients (4). Although most patients with the most common genetic defect, the ΔF508 mutation, have steatorrhea, the present data indicate that there are no differences between PI patients who are homozygous or heterozygous for this defect. It appears that PI is a marker for a more severe genetic defect. Furthermore, among patients with PI, females have a worse prognosis than males.

Maldigestion and/or malabsorption of fat (and fat-soluble vitamins) is only one of many variables affecting energy balance and nutrition in patients with CF. Losses of bile salts and bile acids are frequently associated with steatorrhea and can exacerbate maldigestion and/or malabsorption. In addition, patients who have undergone intestinal resection secondary to bowel obstruction (because of meconium ileus) have reduced absorptive capabilities. The patient's ability to compensate for these nutritional losses is dependent on their appetite and the quantity and quality of food consumed. Although some children with CF have excellent eating habits, there are several factors that commonly reduce appetite and consumption, including recurrent vomiting from coughing and/or gastroesophageal reflux, chronic respiratory infections, and psychosocial stresses.

There is a clear association between malnutrition and deteriorating lung function (5). Chronic pulmonary infections (particularly with Pseudomonas aeruginosa) are associated not only with anorexia but also with increased metabolic rate and energy requirements.

Two other factors more common in adolescent and young adult patients are diabetes mellitus and cholestatic liver disease. Diabetes can increase caloric losses as a result of glucosuria. Liver disease with focal biliary cirrhosis may exacerbate the severity of malabsorption because of inadequate bile acid secretion.

Nutritional deficiency in CF ranges from mildly depleted fat stores to frank signs and symptoms of energy and protein malnutrition. Deficiencies are most likely to occur at times of rapid growth during pulmonary exacerbations and with increased severity of lung disease. Because CF has such a broad impact on nutritional status, clinicians must be vigilant in monitoring and properly instructing patients regarding the appropriate ingestion of the different dietary components, including total calories,

1 From the Cystic Fibrosis Foundation, Bethesda, MD.
2 Address reprint requests to BW Ramsey, Cystic Fibrosis Foundation, 6931 Arlington Road, Bethesda, MD 20814.
3 Contributing members of the Consensus Committee were Robert Beall, Cystic Fibrosis Foundation, Bethesda, MD; Scott Davis, Tulane University, New Orleans; Peter Durie, Hospital for Sick Children, Toronto; Van S Hubbard, NIDDK/NIH, Bethesda, MD; Angela Ibraham, Cystic Fibrosis Foundation, Bethesda, MD; J Nevyn Isenberg, University of Texas Medical Center, Austen; John Lloyd-Still, Children's Memorial Hospital, Chicago; Elisabeth Luder, Mt Sinai School of Medicine, New York; Russell J Merritt, Carnation Company, Los Angeles; Suzanne Michel, Hahnemann University, Philadelphia; Elaine Mischler, University of Wisconsin, Madison; Donna Mueller, Drexel University, Philadelphia; John Patrick, University of Ottawa; Ron Sokol, University of Colorado, Denver; Virginia Stallings, Children's Hospital of Philadelphia; Lori J Stark, Rhode Island Hospital, Providence; Robert Stone, Children's Hospital & Medical Center, Akron, OH; and William Zipf, Children's Hospital, Columbus, OH.

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protein, fat, fat-soluble vitamins, and several minerals. In contrast to the usual textbook description of the voracious appetite of children with CF, many dietary surveys indicate that CF patients often eat less than do nonaffected children (6, 7). Children with CF may also have increased total energy requirements (as described above), so that the net effect is a negative energy balance. The traditional tendency to restrict fat consumption in these patients has aggravated this energy deficiency. Dietary fat is the highest density source of calories, improves palatability of foods, and is needed to maintain normal essential fatty acid (EFA) status. Individuals with CF should whenever possible follow a normal diet pattern with no specific restrictions.

Protein poses less of a nutritional problem than does fat in this population. Protein deficiency is primarily seen in the first year of life when the average requirements are three times as great as those in adulthood. Human milk, which is relatively low in protein (7% of calories), and soy-based formula have been associated with hypoproteinemic edema and growth retardation before instituting enzyme therapy.

Deficiencies in vitamins A and E occur commonly in patients with CF (8–11). Vitamin K deficiency is most likely to occur during infancy, or in association with cholestatic liver disease (5). Vitamin D deficiency has been reported to occur rarely (12).

The following consensus report is the compilation of recommendations generated by a working group that was assembled by the Cystic Fibrosis Foundation to provide nutritional guidelines for the care of patients with CF. The group, which met in April 1990, consisted of nutritional specialists, CF caretakers, and child psychologists. The primary aim of this consensus report is to educate clinicians about the importance of frequent nutritional assessments and early intervention.

There is no reason to accept nutritional failure and/or impaired growth in any individual with CF. The goal of every CF center should be to support normal nutrition and growth for patients of all ages.

**Guidelines for assessment of nutritional status**

**Introduction**

It is important to monitor growth and nutritional status at each clinic visit, thus ensuring early detection of any deterioration and prompt, appropriate nutritional intervention. Present Cystic Fibrosis Foundation (CFF) guidelines suggest that patients with CF be seen on a routine follow-up basis every 3–4 mo. Thus, at a minimum, growth and nutritional status should be monitored at these intervals, at diagnosis, and during the more frequent follow-up of patients whose clinical progress is unsatisfactory. The frequency at which the different indices of nutritional status monitoring should be measured is given in Table 1.

**Growth measurements**

*Length, height, weight, and head circumference.* Accurate and sequential measurements of length (in an infant and toddler aged < 2 y), height, and weight are essential. Similarly, occipitofrontal circumference must be sequentially measured until the age of 2 y. These values should be plotted on a standard growth chart. For patients with suboptimal growth, growth-velocity charts provide additional sensitivity in assessing the patient's progress. A description of standardized techniques for making these measurements have been published (13–15).

It is useful to express weight as a percentage of ideal weight for height, age, and gender. The method for calculating this index is described in Appendix A. The following classification of nutritional status is a modification of that by Waterlow and Rutishauser (16). It is a working classification on which the rec-

![Table 1: Nutritional-status assessment](https://academic.oup.com/ajcn/article-abstract/55/1/108/4715278)
ommendations for nutritional intervention in this document (see Nutritional Management) are based. The range of body size for normal nutritional status is between 90% and 110% of ideal weight-for-height. Eighty-five to 89% of ideal weight-for-height is defined as underweight (may indicate early malnutrition), 80–84% is defined as mild malnutrition, 75–79% is defined as moderate malnutrition, and < 75% is defined as severe malnutrition. A significant fall in the percentage of ideal weight-for-height is reason for concern and the patient should be seen by a dietitian for evaluation. Active dietary intervention should be initiated when a CF patient declines below their established weight curve or becomes underweight (85–89% of ideal weight-for-height). A further deterioration below 85% should provoke consideration of more aggressive clinical assessment and nutrition support measures (see Nutritional Rehabilitation).

Special consideration should be given to the child whose height is less than the third percentile on the standard growth grid. In these cases of stunting, the ideal height-for-weight index may not be appropriate. Waterlow and Rutishauser (16) have published a stunting classification that may be helpful. Stunting or nutritional dwarfism may reflect events in early infancy and as such may not be relevant to nutritional intervention at a later age. A child who has a normal weight-for-height and normal triceps skinfold thickness and midarm muscle circumference should not require nutritional intervention. On the other hand, a child who has had a recent decline in linear growth resulting in stunting (compared with midparental height) and demonstrates an unfavorable energy balance should receive nutritional intervention.

_Midarm indices._ The measurement of midarm circumference in the right arm and triceps skinfold thickness is a useful way of screening and following sequentially the progress of a patient’s body fatness and muscle. The triceps skinfold thickness gives an estimate of energy status whereas the arm-muscle circumference (or area) gives an assessment of lean body mass. A description of standardized techniques for these measurements (13–15) and normative data (17) were published previously. (Muscle mass may decrease from disuse as well as from nutritional problems. Please note this possible effect on midarm-muscle measurements in patients with limited capacity to exercise.)

_Assessment of energy requirements._ Energy requirements in patients with CF are affected by the presence and degree of malabsorption and hypermetabolism. At diagnosis it is essential to assess the dietary intake of the patient and their degree of malabsorption. Similarly, patients who show signs of growth faltering should undergo an assessment of intake and net absorption.

_Assessment of dietary intake._ Assessment of dietary intake requires that each CF center has a dietitian-nutritionist with special interest and training in the care of patients with CF. A 3- or 5-d prospective dietary record is the best way of obtaining an assessment of dietary pattern and energy intake in the outpatient setting. This dietary assessment should be used to determine where energy density and intake may be increased and how enzyme use may be optimized.

A patient who is growing satisfactorily should be seen by a dietitian once a year (minimum). The dietitian may assess dietary habits, using a 24-h dietary recall with an assessment of dietary pattern. The 24-h recall is a qualitative assessment of dietary patterns. A prospective 3–5-d record must be collected for any quantitative assessment of energy intake. The dietitian should provide anticipatory dietary guidance. Times at which this anticipatory guidance is particularly important are infancy and adolescence.

**Assessment of malabsorption.** At the time of diagnosis, every patient should ideally be evaluated for the presence or absence of malabsorption. This assessment is best achieved by a 3-d stool collection combined with a 3-d dietary record. Stool collections can be performed at home after parents have been properly instructed in how to obtain complete fecal specimens and to measure and record food intake. For practical purposes a coefficient of fat absorption < 93% (< 85% in infants) can be used to define steatorrhea. This stool collection must be analyzed for its fat content by using appropriate methods. The laboratory must be informed if the patient is on a formula containing medium-chain triglycerides (MCTs) because a special solvent system must be used in determining stool fat.

It is frequently not possible to judge the degree of steatorrhea by the number or the nature of stools. Many patients with PI will continue to have a significant degree of steatorrhea even when receiving enzyme-replacement therapy. It is therefore important to reevaluate the degree of fat malabsorption in patients with impaired growth.

_Calculation of energy requirements._ Energy requirements are best determined by calculating the basal metabolic rate. Details on how to determine energy requirements are given in Appendix B. Starting from basal metabolic rate, daily energy requirements are estimated taking into account the patient’s activity, pulmonary status and degree of malabsorption.

**Laboratory measurements._**

At diagnosis and when the patient shows clinical deterioration, the following should be determined: electrolytes and acid-base status, complete blood count (CBC), serum albumin, and plasma or serum retinol and α-tocopherol. Annually a CBC should be made; if there is any evidence of iron deficiency, then iron status must be assessed (ie, serum iron, serum iron-binding capacity, and serum ferritin); plasma or serum retinol and α-tocopherol should also be measured.

Routine measurements of vitamin D (25-hydroxycholecalciferol) and vitamin K status are not presently regarded as necessary. There is a potential concern in steatorrheic patients with regard to their absorption of calcium and phosphorus, and bone mineralization. However, routine measurements of plasma, calcium, phosphorus, and alkaline phosphatase may not reflect bone mineralization, which is best assessed by dual-photon absorptiometry (18). This technique is currently used mainly in research.

_Evaluation of patients with nutritional failure._

Nutritional failure is defined as follows: patients aged < 5 y (preschool)—a weight-for-height index < 85% of ideal weight and standard height, loss of weight for > 2 mo, and/or a plateau in weight gain for 2–3 mo; patients aged 5–18 y (school age)—a weight-for-height index < 85% of ideal weight and standard height, loss of weight for > 2 mo, and/or a plateau in weight gain for 6 mo; and patients aged > 18 y (postpubertal)—a weight-for-height index < 85% of ideal weight and standard height and/or weight loss ≥ 5% of usual weight for > 2 mo.

Nutritional assessment must include measurements of height (length), weight, head circumference (for those aged < 2 y), and
midarm indices; 3-d dietary records to quantitate energy intake and to determine distribution of calories by macronutrient categories (record must account for losses due to vomiting); (and a 3-d fat-balance study in conjunction with a dietary record both while the patient is consuming enzymes).

Pulmonary disease evaluation includes pulmonary-function tests (if child can properly perform them), a chest roentgenogram, and oximetry (arterial blood gases, if indicated). Other medical disorders should be considered in the evaluation of nutritional failure: diabetes mellitus, progressive liver disease (multifocal biliary cirrhosis), and other gastrointestinal complications (eg, lactose intolerance, Crohn’s disease, chronic abdominal pain, distal intestinal obstruction, esophagitis). A behavioral assessment should also be done. Family dynamics, eating patterns, recent stressors, and depression should be considered.

Guidelines for nutritional management

Introduction

Many CF patients will adjust their dietary intake in response to acute illnesses and changes in illness severity. However, they are not always successful in maintaining normal nutritional status. At these times of nutritional failure, further medical intervention will be needed. All nutritional-management decisions should be based on a graded response that is appropriate for the needs of each patient. Five response categories are defined in Table 2 and will be described in detail in the following section.

It is important that nutritional intervention be initiated early in the course of the illness when patients have mild or moderate pulmonary involvement. Aggressive nutritional rehabilitation in terminally ill patients is not medically indicated (unless they are awaiting organ transplantation). A prognostic index was published, which may be helpful in determining those malnourished CF patients who are not likely to benefit from intensive nutritional support (19).

Routine management

Diagnosis of cystic fibrosis. The time of diagnosis is a crucial period for beginning nutritional education, dietary counseling, and therapeutic interventions. Extensive discussion of nutritional management with patients and/or parents is just as important as are other aspects of CF care and should be reviewed on an ongoing basis. By concentrating on nutrition during the family’s first few visits to the CF Center, care givers will be able to stress the importance of establishing and maintaining good nutrition.

Infancy to 2 y. The first 2 y represent the phase of life with the highest growth rate and relative energy needs. Infants with CF should be evaluated weekly or every other week until normal weight gain has been established, and every 2–3 mo thereafter. Clinical and/or laboratory assessment of pancreatic function should be completed as described in the Nutrition Assessment section.

Pancreatic-enzyme-replacement therapy should be initiated with each feed if malabsorption and/or malabsorption has been identified. Enzymes should be given with all types of milk products, including predigested formulas and human breast milk (see below). Microspheres are preferable to granules because they cause less excoriation of the mouth and perianal region. Microspheres can be made palatable by administering them with a vehicle such as applesauce or rice cereal. Pancreatic-enzyme supplementation must be determined on an individual basis. A guideline dose for initiation of therapy would be 1000–2000 IU lipase per 120-mL feeding, which can be adjusted according to growth velocity and stool pattern. If an infant requires more than 1 capsule (providing ≥4000 IU lipase) per 120-mL feeding, further evaluation may be indicated and other etiologies of malabsorption (eg, lactose intolerance) should be considered.

Infants who have short-gut syndrome or residual cholestasis should be considered to be in a special risk category and may require, depending on the extent of bowel resection, nasogastric continuous feeds as well as predigested formula with enzyme supplementation for adequate growth. Some infants (and older patients) do not produce adequate pancreatic bicarbonate to activate enteric-coated enzymes. These patients may benefit from hydrogen-receptor blockers or nonenteric-coated enzymes.

Some infants with CF will sustain normal growth on human milk when receiving pancreatic-enzyme supplementation. However, special attention should be given to two potential metabolic complications in breast-fed infants: hypoproteinemia

<table>
<thead>
<tr>
<th>Category</th>
<th>Target group</th>
<th>Goals</th>
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<tbody>
<tr>
<td>Routine management</td>
<td>All CF patients.</td>
<td>Nutritional education, dietary counseling, pancreatic-enzyme replacement (for patients with PI), vitamin supplementation (for patients with PI).</td>
</tr>
<tr>
<td>Anticipatory guidance</td>
<td>CF patients at risk of developing energy imbalance (ie, severe PI, frequent pulmonary infections, periods of rapid growth), but maintaining a weight-height index ≥ 90% of ideal weight.</td>
<td>Further education to prepare patients for increased energy needs; increased monitoring of dietary intake; increase caloric density in diet as needed; behavioral assessment and counseling.</td>
</tr>
<tr>
<td>Supportive intervention</td>
<td>Patients with decreased weight velocity and/or a weight-height index 85–90% of ideal weight.</td>
<td>All of the above plus oral supplements as needed.</td>
</tr>
<tr>
<td>Rehabilitative care</td>
<td>Patients with a weight-height index consistently &lt; 85% of ideal weight.</td>
<td>All of the above plus enteral supplementation via nasogastric tube or enterostomy as indicated.</td>
</tr>
<tr>
<td>Resuscitative and palliative care</td>
<td>Patients with a weight-height index &lt; 75% of ideal weight, or progressive nutritional failure.</td>
<td>All of the above plus continuous enteral feeds or total parenteral nutrition.</td>
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and hyponatremic alkalosis. These infants should have appropriate monitoring of protein and electrolyte status as well as growth velocity at 1–3 mo intervals. Breast-fed infants will generally require supplementation with sodium chloride, especially during summer months. An appropriate and safe dose is 2–4 mmol·kg body wt·d⁻¹.

Milk products serve as the predominant source of nutrients for infants with CF during the first year of life. Lactose intolerance does not appear to be more common in the CF population. In some instances supplementary fat or carbohydrate might be needed or a caloric density > 2.8 kJ/L (>20 kcal/30 mL) if concentrated formulas are used. Although the duration of formula feeding varies among different CF centers, it is prudent to continue formula for ≥12 mo and up to 24 mo. If an infant with CF is growing at a normal rate, a change to whole cow milk may be recommended between 12 and 18 mo of age; however, an adjustment of the pancreatic-enzyme-supplement dose might be needed during that interval.

Complementary foods should be introduced according to the guidelines of the American Academy of Pediatrics (AAP), namely between 4 and 6 mo of age or earlier if necessary. A source of iron and fluoride needs to be introduced during the first year as for any infant (see AAP guidelines, AAP, Elk Grove Village, IL). Infants with CF can be referred by the CF Center to Women, Infant, and Children (WIC) programs in each state. These programs provide predigested formulas with proper documentation of need.

**Toddlers to preschool (age 2–6 y).** Children in this age group have developed self-feeding behaviors and express individual food preferences. Dietary intake and degree of physical activity vary from day to day. For these reasons, close monitoring of dietary habits, caloric intake, and growth velocity are important. Dietary counseling and anticipatory guidance by CF team members (eg, RN, RD, and MSW) are very beneficial during this period when long-term feeding habits are developing.

**School age (age 6–12 y).** Children in this age group are exposed to significant degrees of peer pressure and are challenged to self-manage their disease. Compliance with prescribed medications such as pancreatic enzymes and fat-soluble vitamins can become a major problem during this period. In addition, acceptance and understanding by teachers and fellow students may be lacking, further stressing a child with CF.

**Adolescence and puberty (age 13–18 y).** This developmental stage is associated with high nutrient requirements because of accelerated growth, endocrine development, and high levels of physical activity. Pulmonary infections are often more common in this period, increasing caloric needs. Growth failure and pubertal delay are common and come at a time of extreme social pressures and psychosocial stresses. Females are at greatest risk for nutritional failure because of their lower lean body mass.

Frequent monitoring of dietary intake, growth velocity, and nutritional status are important: every 3 mo is optimal. Nutritional counseling must be directed toward the patient rather than the parents. The adolescent will benefit from a team effort (eg, RN, RD, and MSW).

**Adulthood.** CF patients reaching adulthood are usually responsible for the entire management of their disease, as well as for the financial burden of a chronic illness. They are facing the challenge of supporting themselves and providing optimal nutrition. While in college or working, adults with CF are constantly adapting to new schedules and stresses. Nutritional counseling must be practical and pragmatic to help adults adjust to these changes.

**Pregnancy and lactation.** Widespread experience in recent years has demonstrated that pregnancy and lactation can be accomplished successfully by some women with CF (20). In addition to the usual principles of nutritional care, it is recommended that pregnant women follow the guidelines of the RDA Committee of the Food and Nutrition Board, National Academy of Sciences (21). Special attention should be given to adequate weight gain, particularly during the last trimester of pregnancy. Multivitamins should be taken as before and, in addition, one prenatal vitamin should be consumed daily.

During lactation the marked increase in energy needs must be taken into account. Breast-feeding a 3-mo-old infant requires assimilation of 3150 J (750 kcal) dietary energy/d. This means that markedly increased energy intake is needed by a lactating CF patient to avoid weight loss. Some women with CF will not be able to maintain adequate nutrition during lactation. They must be closely monitored and if their nutrition suffers, they should be reassured that their infants will be adequately nourished with commercial formulas.

**Vitamin supplementation for CF patients.** Every patient with CF and PI will eventually require supplementation with fatsoluble vitamins. Vitamins A and E are of particular concern. Vitamin D deficiency occurs primarily in CF patients with inadequate sunlight exposure or cholestatic liver disease. More research is needed to define the frequency of vitamin K deficiency in this population. Fat-soluble vitamins are absorbed most effectively when taken in the morning with fat-containing meals and pancreatic enzymes. If vitamin A or E concentrations are low despite the doses prescribed below, it is important to evaluate patient compliance. In addition, it is conceivable that some individuals may need higher supplements depending on their degree of malabsorption and whether cholestatic liver disease is present.

Most CF patients will receive adequate supplementation from a multiple-vitamin preparation. Among CF clinical centers there are varied recommendations for multiple-vitamin supplementation. Until further information is available, the following is recommended. Infants aged ≤2 y should take 1 mL Polyvisol (Mead Johnson Nutritional, Evansville, IN) or similar liquid multivitamin preparation daily. Children aged 2–8 y need a standard multiple vitamin containing 400 IU vitamin D and 5000 IU vitamin A in a dose of 1 tablet/d. Older children, adolescents, and adults need a standard adult multiple-vitamin preparation, 1–2 tablets/d. For patients not maintaining adequate serum vitamin A concentrations with a multivitamin preparation, water-miscible vitamin A preparations (eg, Aquasol A, Rover Pharmaceuticals, Fort Washington, PA) are available.

**Vitamin E supplementation should follow these guidelines:**

- ages 0–6 mo, 25 IU/d; 6–12 mo, 50 IU/d; 1–4 y, 100 IU/d; 4–10 y, 100–200 IU/d; and >10 y, 200–400 IU/d. Vitamin E is currently given as Aquasol E (Rover Pharmaceuticals, Fort Washington, PA) or Liqui-E (Twin Labs, Ronkonkoma, NY) for the first year or two. After the first year or two, capsules (chewed up, squeezed out, or swallowed) of α-tocopherol or d-α-tocopheryl acetate may be used. Note that excessive doses of vitamin E (>1000 IU/d) may exacerbate the coagulopathy associated with vitamin K deficiency.

More research is needed to define the optimal supplementation regimen for vitamin K in patients with CF. Until further infor-
Anticipatory guidance

It should be the goal of CF care givers to educate and prepare families and patients for periods of increased caloric needs (eg, rapid growth) or decreased appetite (eg, pulmonary infections). Special attention should be directed to the families of females patients during the preadolescent and adolescent years. With proper guidance and training, families and patients can minimize nutritional decline during these periods. Two areas of anticipatory guidance, 1) oral supplementation (22) and 2) behavioral assessment and modification, are outlined below.

The first level of oral nutrition support is adequate daily food intake with adequate pancreatic-enzyme-replacement and vitamin-mineral supplementation. The second level is boosted oral intake. The booster concept considers the person’s usual food preferences and habits and then increases the total nutrient and caloric intake without dramatically increasing the amount of food consumed. For example, margarine or butter may be added to potatoes, vegetables, rice, and cereal. Instead of tomato soup made with water [1.74 kJ/L (50 kcal/120 mL)], it can be prepared with half-and-half (milk and cream) and garnished with grated cheese and croutons [6.97 kJ/L (200 kcal/120 mL)]. In addition, socially acceptable, nutrient-dense foods such as nuts, cheese and crackers, cold cuts, whole milk, peanut butter and jelly sandwiches, or pizza may be added as snack items instead of soda or fruit juice.

The third level of nutritional support is adding homemade or proprietary oral supplements to the usual diet. Homemade supplements such as milk shakes are often more palatable, less expensive, and can be prepared from readily available ingredients. Recipes have been formulated to satisfy numerous individual taste preferences and medical conditions. However, some CF centers recommend (and patients select) commercial beverages, powders, and puddings because of their convenience. New products are continuously being manufactured and marketed and may provide an adjunctive source of nutrient supplementation. Because of growth and development issues, taste fatigue, and changes in health status, a combination of approaches to oral nourishment must be offered and taught throughout the entire life span of the person with CF.

Oral intake in children with CF may be influenced by many psychosocial and environmental factors. These factors include parent-child mealtime interactions, child eating behaviors, and the temporal sequencing of eating and abdominal discomfort. CF care providers should assess the amount of time a CF child spends at meals, whether the child consistently eats or whether the child dawdles, and how the parent handles the situation. Long mealtimes (> 20–30 min), slow eating, and reports by parents that they must constantly nag the child to eat are indicative of problem areas that can be addressed through behavioral-management strategies.

Families should be encouraged to have regular mealtimes in which the family sits together, without distractions such as television. Parents should establish clear rules and consequences for mealtimes and these should apply to their healthy child and CF child alike. Expansion of food choice is important and best done gradually. Similarly, to increase energy consumption, the child’s baseline caloric intake should be established and small increments of 418–836 J/d (100–200 kcal/d) should be added every 1–2 wk with specific recommendations for foods and meals to be targeted. In conjunction with the gradual increase of calories, parents should be taught to encourage eating by complimenting their child and paying attention when their child is eating.

Parents should be discouraged from nagging and keeping their child at the table beyond when other family members leave the meal. A child’s refusal to eat should be managed by consistent consequences that may be applicable to children in general, such as no dessert unless dinner is eaten. For problematic refusals, parents may need to implement a more structured program, such as sticker charts as a reward for adequate food consumption and eating within reasonable time limits. To promote child self-selection of appropriate foods, parents should be instructed to offer food choices in which both alternatives are acceptable choices.

As the child progresses into adolescence, nutritional issues may be best addressed directly between the physician and adolescent via discussion and formal contracting. In these cases, reasonable caloric-intake goals should be set that will gradually increase at successive clinic visits. Emphasis should be placed on increasing strength and/or enhancing body image rather than on weight gain.

Consistency is a key factor in the success of behavioral programs. Inclusion of both parents in the program, including divorced households as well as significant-other adults who may care for the CF patient during mealtimes (eg, grandparents), is necessary. Referral for more intensive behavioral intervention should be considered when problematic eating behaviors have been identified and/or when the parents have been unable to implement the above strategies successfully. It is important to clarify to families that even with optimal feeding patterns, some children will not be able to sustain adequate nutrition because of their very high caloric needs. Families and patients must never feel that they have failed or that we blame them when feeding strategies fail and growth is impaired.

Supportive intervention

When a CF patient’s weight-for-height ratio falls between 85% and 90% of ideal, or his weight velocity declines, reevaluation is necessary. Treatment should focus on increasing the frequency and energy density of meals, optimization of enzyme therapy, and identification of any remediable psychological stressors.

The initial management for this level of nutritional decline is the same as that outlined in the previous section, Anticipatory guidance. Emphasis should be placed on optimizing feeding behaviors and adding oral supplements. If these measures are not successful within 3 mo or if the patient’s weight-height ratio declines to < 85% of ideal, more aggressive nutritional management should be considered, as outlined below.

The onset of nutritional failure necessitates an immediate nutritional and medical evaluation of the patient. The definition of nutritional failure and an appropriate assessment are outlined under Evaluation of patients with nutritional failure.

Enteral feeds. When the use of noninvasive techniques (eg, oral supplementation and behavioral modification) fails to improve the nutritional status of CF patients, within a 3-mo period, further intervention is needed with enteral feedings. Although there has not been a controlled clinical trial of nutritional support by enterostomy techniques, the following potential benefits have
been observed in studies performed thus far (23–25): 1) improved status of body composition, 2) increased strength, 3) increased sense of well-being, 4) increased sense of control over body weight, 5) improved body image, 6) development of secondary sexual characteristics, 7) less weight loss during exacerbations of pulmonary infections, and 8) improved nutritional status during the terminal phase of CF.

Until more results of clinical investigation have been published on the use of supplemental tube feedings, the following approach and guidelines are suggested.

Before placement of an enterostomy tube, a family assessment should be done, the patient and family should be educated about the technique, and glucose tolerance should be assessed. Complete information should be provided as to the anticipated duration of tube feedings and the possible need for insulin. The acceptance and long-term commitment of both the patient and family are necessary for a successful intervention. The choice of an enterostomy tube and technique for its placement should be based on local factors and the expertise of the CF center involved. Nasogastric tubes, gastrostomy tubes, and jejunostomy tubes have all been used successfully. Nasogastric tubes are acceptable for short-term nutritional support in highly motivated patients followed in centers committed to this approach. Gastrostomy tubes can be placed percutaneously in sedated patients as well as under general anesthesia by a surgeon. Gastrostomy tubes carry the risk of gastroesophageal reflux (GER) and vomiting. Most patients with GER can be controlled by adjustment of the feeding rate, appropriate positioning during sleep, and in some cases use of prokinetic agents (eg, domperidone). Jejunostomy tubes potentially increase the problem of nutrient malabsorption but have been used very successfully in some CF centers. Patients with significant lung disease will generally require a pulmonary tune-up before surgical placement of an enterostomy tube.

The choice of a nutritional supplement (formula for tube feeding) should be based on gastrointestinal and nutrition principles. There are advantages and disadvantages associated with each option (eg, elemental formulas, high-fat mixtures). Furthermore, there is no compelling evidence to support any particular option although the supplements should be nutritionally balanced with respect to micronutrients. There is no ideal method for enzyme delivery at night. Until further research has been completed on this issue, it is generally recommended that the patients take pancreatic enzymes as the tube feedings begin at bedtime. Elemental diets do not require enzymes.

Initiation of enteral tube feedings usually requires approximately 1 wk of hospitalization for education, evaluation, and for advancing the feedings. This applies to nasogastric tubes as well as to gastrostomy and jejunostomy tubes. The principle for advancing feedings involves starting slowly and increasing the nutrient density of the formula first, and then the volume. It is advisable to start with 50–75% strength formula for the first 24 h and advance to full strength over 48 h. After full caloric density is reached, hourly volume may be increased slowly to a maximum rate tolerated by each patient.

The recommended nutritional supplementation regimen should be individualized and flexible, with adjustments made to achieve nutritional goals as indicated. Generally, tube feedings involve nocturnal infusions of additional energy with an increment of one-fourth to one-third the previously consumed oral caloric intake. This is a good starting point but some patients can be repleted with less and some require more daily caloric supplements. In all instances a carefully planned home-care system is needed with follow-up contact, infusion pumps, etc. Attention needs to be given to the patient’s position during nocturnal tube feedings. It may be necessary to raise the head of the bed. If vomiting occurs, medical assessment is needed. Monitoring for glucose intolerance and fluid and electrolyte imbalance must be a routine part of follow-up care.

**Parenteral nutrition support.** Parenteral nutrition supplements and total parenteral nutrition per se are only indicated for short-term support in CF patients with specific problems, such as short-gut syndrome, pancreatitis, severe gastroenteritis, and for the postoperative management of patients who have had intestinal surgery or similar procedures. Patients with short-gut syndrome may require long-term parenteral nutrition support. There are other special situations in which CF patients might require parenteral nutrition, eg, patients who refuse the enteroctomy approach and patients who are candidates for heart-lung-transplant surgery.

Nevertheless, it should be recognized that long-term parenteral nutrition support is not appropriate for energy supplementation because of its high cost and increased risks. Furthermore, it has been well-established during the past decade that the enteral approach of tube feeding is generally satisfactory. The well-established principle that the gut should always be used first for nutritional support needs to be followed.

**Anabolic agents, appetite stimulants, and growth hormone.** The use of appetite stimulants and/or androgens to attempt to enhance growth in children with CF has not been shown to have significant advantages and is not recommended. The potential advantages and/or complications of growth hormone as an anabolic or growth promoting agent in CF are not known. The possibility that growth hormone might have negative effects on the increased incidence of glucose intolerance and diabetes mellitus in CF is of concern. Until additional information is available from carefully controlled trials, the use of growth hormone in CF is not recommended.

**Treatment of the CF patient with diabetes mellitus.** The patient with both CF and sufficient glucose intolerance to warrant directed therapy needs special attention. The CF care provider should consult the Report of the Consensus Conference on CF-Related Diabetes Mellitus (26).

**References**

APPENDIX A

Calculation of weight as a percentage of ideal weight for height (1)

Step 2: Determine the ideal weight-for-height. Is weight on the same centile as that for height, age, and gender, eg, for a 6-y-old girl with a height on the 25th centile, the ideal weight should be on the 25th centile of the weight chart for a 6-y-old girl. Note that growth charts only go up to 18 or 19 y of age. Experience has shown that calculation of ideal weight-for-height in older patients is satisfactory when the values at the oldest end of the growth chart (ie, 18 or 19 y of age) are used.

Step 3: Express actual weight as a percent of ideal weight-for-height: (actual weight/ideal weight-for-height) × 100.

Reference

APPENDIX B

Determination of energy requirements

For patients with CF who are growing normally and whose steatorrhea is under good control, the total daily energy requirement (DER) is consistent with recommended dietary allowances (RDAs) (1) for age and sex. It is reasonable to assume that these patients do not need greater than the RDA to achieve normal growth.

If a patient fails to grow adequately while receiving caloric intake, based on RDAs, the following formula should be used to calculate the DER:

Step 1: Calculate the basal metabolic rate (BMR) by using the World Health Organization equations for predicting BMR from weight (2; see Table 1B).

Step 2: Calculate daily energy expenditure (DEE) by multiplying the BMR by activity plus disease coefficients as listed below: activity coefficients (ACs) —confined to bed (BMR × 1.3); sedentary (BMR × 1.5); and active (BMR × 1.7). Disease coefficients—for patients with essentially normal lung function, ie, forced expiratory volume in 1 s (FEV1) of that predicted (BMR × (AC + 0)); for patients with moderate lung disease, ie, FEV1 of that predicted (BMR × (AC + 0.2)); for patients with severe lung disease, ie, FEV1 of that predicted (BMR × (AC + 0.3)). If pulmonary-function tests (PFTs) are not available, assess severity of lung disease clinically.

Sample calculation: For patients with an FEV1 of 42% of that predicted and who attend school but are relatively sedentary:

\[
DEE = BMR \times (1.5 + 0.2) = BMR \times 1.7
\]

* May range up to 0.5 with very severe lung disease.

**TABLE 1B**

Equations for predicting BMR (in kcal) from body weight (in kg)

<table>
<thead>
<tr>
<th>Age range</th>
<th>Females</th>
<th>Males</th>
</tr>
</thead>
<tbody>
<tr>
<td>0–3 y</td>
<td>6.1wt−51</td>
<td>6.0wt−54</td>
</tr>
<tr>
<td>3–10 y</td>
<td>22.5wt+499</td>
<td>22.7wt+495</td>
</tr>
<tr>
<td>10–18 y</td>
<td>12.2wt+746</td>
<td>17.5wt+651</td>
</tr>
<tr>
<td>18–30 y</td>
<td>14.7wt+496</td>
<td>15.3wt+679</td>
</tr>
<tr>
<td>30–60 y</td>
<td>8.7wt+829</td>
<td>11.6wt+879</td>
</tr>
</tbody>
</table>
Step 3: Calculate DERs from DEE, taking into account the degree of steatorrhrea. For pancreatic-sufficient patients (including patients on enzymes with a coefficient of fat absorption ≥ 93% of intake): DER = DEE. For pancreatic-insufficient patients [the coefficient of fat absorption (CFA) must be determined as a fraction of fat intake]: DER = DEE(0.93/CFA). If a stool fat collection is not available to determine the fraction of fat intake, an approximate value of 0.85 may be used in the calculation.

Sample calculation for a typical patient: If fat absorption on enzymes is equivalent to 78% of intake, CFA = 0.78.

† As measured by a 72-h fecal fat collection.

Daily energy expenditure with average activity = 8360 J, or 2000 kcal

Daily energy requirement = 8360 J (0.93/0.78), or 2000 kcal (0.93/0.78)

= 9968 J/d, or 2384 kcal/d

References