Evidence Analysis Library
Cystic Fibrosis Systematic Review and Guideline (2020)

Relationship between Systematic Review PICO questions and CF Nutrition Recommendations

PICO Question(s)	Recommendation
MNT: Suggested Approach and Dietitian Full Time Equivalents (FTEs)	
In participants with CF, how does medical nutrition therapy (MNT or nutrition counseling) provided by a registered dietitian or international equivalent on nutrition-related outcomes? In participants with CF, what is the longitudinal relationship (at least 3 months) between weight and growth parameters and hard outcomes (FEV1, Quality of Life or mortality)?	RDNs or international equivalents should collaborate with all individuals with CF, their families, and interdisciplinary healthcare teams to co-produce individualized Medical Nutrition Therapy (MNT) based upon the individual's personal preferences, psychological and psychosocial factors, physiological needs, health status, and pharmacological interventions. MNT for individuals with CF should include comprehensive nutrition assessment and appropriate interventions, including individualized modification of diet, dietary supplements including micronutrient supplements, and pancreatic enzymes, in order to maintain or improve nutrition status and symptoms over time.
	It is reasonable for one FTE RDN or international equivalent to provide care for 75-150 individuals with CF. A caseload at the lower end of this range is appropriate for dietitians who work primarily with the pediatric population or adults with advanced disease and/or co-morbidities in order to deliver continuous, high-quality nutrition care that effectively manages nutrition challenges and prevents disease decline.
Nutrition Screening: Pediatrics	
In pediatric participants with CF, which nutrition screening methods are valid and reliable compared to reference standards, as measured by validity and/or reliability studies?	For infants and children with CF <2 years of age, it is reasonable to measure weight and length at each clinic visit and to screen for risk of impaired growth and other nutrition concerns at least monthly for the first six months of age, every other month from 6-12 months of age, and quarterly from 12-24 months of age to identify nutrition risk.
In participants with CF, how does medical nutrition therapy (MNT or nutrition counseling) provided by a registered dietitian or international equivalent on nutrition-related outcomes? In participants with CF, what is the longitudinal relationship (at least 3 months) between weight and growth parameters and and	Infants and children with CF <2 years of age should be screened for nutrition risk by comparing weight-for-age, weight-for-length and length-for-age z-scores or percentiles to birthweight and to growth norms using WHO growth charts for the general population, since these parameters are longitudinally associated with lung function. Children who are not maintaining birthweight, weight-for-length or length -for-age z-scores or who have depressed growth compared to the general population should be referred for full nutrition assessment by an RDN or international equivalent.
	For children and adolescents with CF ages 2-20 years, weight and height should be measured at each clinic visit and children should be screened for risk of impaired growth and other

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hard outcomes (FEV1, Quality of Life or mortality)?	nutrition concerns at least quarterly or more frequently based on clinical condition to identify nutrition risk.
	Pediatric individuals with CF 2-20 years of age should be screened for nutrition risk by comparing growth percentiles and z-scores to general population norms using CDC growth charts for the general population, since these parameters are longitudinally associated with lung function. Children and adolescents who have a BMI-for age <50th percentile and/or who have concerning trends in BMI-for-age, weight-for-age or height-for-age z-scores should be referred for full nutrition assessment by an RDN or international equivalent.
Nutrition Screening: Adults	
In adults with CF, which nutrition screening methods are valid and reliable compared to reference standards, as measured by validity and/or reliability studies?	In adults with CF > 20 years of age, it is reasonable to measure weight and height at each clinic visit and to screen for malnutrition and/or other nutrition concerns at least quarterly or more frequently based on clinical condition to identify nutrition risk.
In adults with CF, what is the longitudinal relationship (at least 3 months) between Nutrition Focused Physical Exam and hard outcomes (FEV1, Quality of Life or mortality)?	Adults with CF >20 years of age should be screened for nutrition risk by evaluating absolute values and trends in BMI, since BMI is longitudinally associated with lung function. Women who have a BMI <22 kg/m2, men who have a BMI <23 kg/m2, or adults who have concerning trends in BMI, either decreasing or increasing, should be referred for full nutrition assessment by an RDN or international equivalent.
Nutrition Assessment and Diagnosis of Nutrition Status: Pediatric and Adult	
In participants with CF, which nutrition assessment methods are valid and reliable compared to reference standards, as measured by validity and/or reliability studies?	In individuals with CF, a full nutrition assessment should be conducted by an RDN or international equivalent • at diagnosis; • when indicated by nutrition screening; • up to monthly for the first six months of life; up to every other month until one year of age; and up to quarterly until two years of age; • annually for individuals greater than two years of age; • when disease or treatment course changes
In participants with CF, how does medical nutrition therapy (MNT or nutrition counseling) provided by a registered dietitian or international equivalent on nutrition-related outcomes? In participants CF, what is the longitudinal relationship (at least 3 months) between nutrition parameters and hard outcomes (FEV1, Quality of Life or mortality)?	In individuals with CF, the RDN or international equivalent should diagnose nutrition status, including underweight and overweight, based on a comprehensive assessment of weight and growth history and stature, body composition, disease severity, laboratory values, drug-nutrient interactions/implications, and estimated energy expenditure compared to client/parent report of dietary intake and food security status, since CF nutrition pathology is highly individual and maintaining optimal nutrition status is a necessary component of preventing disease progression.
Role of CFTR Modulators in Nutrition Assessment	

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In participants with CF, what is the effect of CFTR modulation therapy on weight/growth parameters and body composition?	For individuals with CF of all ages who receive CFTR modulation therapy, the RDN or international equivalent should continue to conduct nutrition screening with nutrition assessment as indicated based on age, since these medications may change nutrient needs for some individuals with CF.
Assessment of Energy Requirements	
In participants with CF, what is the accuracy of using energy requirement formulas to determine resting energy expenditure (REE) compared to indirect calorimetry or doubly labeled water?	In pediatric (≤20 years of age) and adult individuals with CF, it is reasonable for the RDN or international equivalent to measure energy needs using indirect calorimetry, when feasible and indicated, since indirect calorimetry is the gold standard for measuring energy expenditure in clinical settings.
	In pediatric individuals with CF ≤20 years of age, the RDN or international equivalent may estimate energy needs at each nutrition assessment using the RDA or IOM active lifestyles formulas, since these formulas were the most accurate compared to indirect calorimetry in this population. Energy needs should be individualized based on growth history, nutrition status and medications, physical activity and disease severity.
	In adults with CF >20 years of age, the RDN or international equivalent may estimate energy needs annually or with unintentional weight changes using standard energy expenditure equations x 1.25, since estimated energy requirements for the general population may underestimate needs in adults with CF. Energy needs should be individualized based on nutrition status and medications, physical activity and disease severity.
Body Composition Assessment	
In participants with CF, which body composition parameters are valid and reliable compared to reference standards, as measured by validity and/or reliability studies? In participants with CF, what is the longitudinal relationship (at least 3 months) between body composition and hard outcomes (FEV1, Quality of Life or mortality)?	In individuals with CF >8 years of age, it is reasonable for the RDN or international equivalent to assess bone mineral density, fat mass and lean mass using dual-energy X-ray absorptiometry (DEXA), when feasible and indicated, since DEXA is the gold standard for assessing these measures in clinical settings.
	In all individuals with CF, when body composition assessment with dual-energy X-ray absorptiometry (DEXA) is not feasible or indicated, it is reasonable for the RDN or international equivalent to assess mid-upper arm circumference with single site skinfold measures and/or Nutrition-Focused Physical Exams at each nutrition assessment in order to aid in the classification of nutrition status over time.
	In individuals with CF, when dual-energy X-ray absorptiometry (DEXA) is not feasible or indicated, the RDN or international equivalent may use age-appropriate tests including skinfold measures or bioelectrical impedance analysis (BIA) with caution when tracking body composition over time, understanding that prediction equations using these methods may over- or under-estimate absolute fat and fat-free mass.
Nutrition Assessment of Biochemical Values	

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In participants with CF, what is the longitudinal relationship (at least 3 months) between CFRD and hard outcomes (mortality, FEV1 and QoL)?	For individuals with CF ≥10 years of age who have not previously been diagnosed with diabetes, oral glucose tolerance testing (OGTT) is recommended annually or as indicated by glucose levels and clinical signs and symptoms (weight loss, increase in pulmonary exacerbations and/or loss of lung function) during nutrition assessment, since progression to CFRD is a risk factor for pulmonary decline and mortality.
In participants with CF, what is the longitudinal relationship (at least 3 months) between fat-soluble vitamin levels and hard outcomes (mortality, FEV1 and QoL)?	For all individuals with CF, regardless of exocrine pancreatic function, it is reasonable for the RDN or international equivalent to assess fat-soluble vitamin levels at least annually, since there may be high risk of fat-soluble vitamin abnormality due to pancreatic insufficiency and malabsorption.
In participants with CF, what is the longitudinal relationship (at least 3 months) between lipid profile and hard outcomes (mortality, FEV1 and QoL)?	For individuals with CF, it is reasonable for the RDN or international equivalent to evaluate fasting lipid profile at least once between the ages of 10 and 20 years and every 4-6 years thereafter, or more frequently if the individual has multiple risk factors for cardiovascular disease, in order to detect and prevent dyslipidemia.
General Guidance for Food Intake	
In participants with CF, what is the relationship between dietary intake of food groups, dietary patterns and meal frequency and nutrition-related outcomes?	For all individuals with CF, it is reasonable for the RDN or international equivalent to advise an age-appropriate, healthy diet that emphasizes culturally appropriate foods associated with positive health outcomes in the general population, including vegetables, fruits, whole grains, seafood, eggs, beans and peas, nuts and seeds, dairy products, and meats and poultry, as tolerated and preferred by the individual with CF, since there is no evidence to suggest that routine modification from a well-balanced, healthy diet is associated with improved outcomes. It is reasonable to advise supplementation with energy and/or protein dense foods or oral or enteral supplements, as needed to achieve or maintain normal growth (pediatrics) or BMI status (adults).
	For all individuals with CF, it is reasonable for the RDN or international equivalent to consider advising a dietary pattern, individualized for dietary preferences and nutrient needs, that promotes consumption of nutrient-dense foods, including healthy fats and micronutrients.
	For all individuals with CF, it is reasonable for the RDN or international equivalent to suggest frequent food intake throughout the day, including at least three meals with snacks in between, as needed, in order to meet energy and protein needs and achieve or maintain optimal weight/growth and nutrition status.
In participants with CF, what is the relationship between dietary	For all individuals with CFRD, it is reasonable for the RDN or international equivalent to consider advising a diet consistent

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intake of food groups, dietary patterns and meal frequency and nutrition-related outcomes? In participants with cystic fibrosis-related diabetes (CFRD), what is the relationship between refined carbohydrates, including juice, soda and candy, and glycemic control? In participants with CF, what is the longitudinal relationship (at least 3 months) between CFRD and hard outcomes (mortality, FEV1 and QoL)?	with general, age-appropriate healthy dietary recommendations and individualize as needed according to CFRD pathology. It is reasonable for the RDN to emphasize limiting high-sugar foods and beverages with low nutrient density, due to adverse effects on blood glucose levels. For individuals with CF who are overweight or obese, it is reasonable for the RDN or international equivalent to advise an age-appropriate diet that emphasizes foods associated with positive health outcomes in the general population, including vegetables, fruits, whole grains, seafood, eggs, beans and peas, nuts and seeds, dairy products, and meats and poultry, as tolerated and preferred by the individual with CF, with energy needs adjusted to achieve or maintain normal growth (pediatrics) or BMI status (adults).
Macronutrient Distribution	
In participants with CF, what is the relationship between dietary macronutrient distribution and nutrition-related outcomes?	For individuals with CF who are not at risk of malnutrition, RDNs or international equivalents may suggest consuming macronutrients (carbohydrates, protein and fat) in the same percentage distribution as is recommended for the typical, age-matched population, since there is no current evidence to suggest benefits from modified macronutrient distribution.
Fiber Intake	
In participants with CF, what is the relationship between fiber intake and nutrition-related outcomes?	For individuals with CF, the RDN or international equivalent may suggest dietary fiber intake in line with the dietary reference intake for the general population, as tolerated on an individual basis, since evidence suggests fiber intake at the recommended level does not increase risk of constipation, DIOS or other gastrointestinal symptoms.
Infant Feeding	
In infants with CF, what is the relationship between exclusive breastfeeding, mixed feeding or formula feeding and nutrition-related outcomes? In participants with CF, what is the longitudinal relationship (at least 3 months) between weight and growth parameters and hard outcomes (FEV1, Quality of Life or mortality)?	In infants diagnosed with CF, RDNs should recommend breastfeeding to the greatest extent possible, with breast milk fortification and formula supplementation as necessary to regain birthweight z-score and achieve normal growth for age. Breastfeeding is associated with improved FEV1% predicted and decreased antibiotic use, but supplementation may be needed for infants with high nutrient requirements.