Cystic Fibrosis

Care Guidelines for Nutrition Management

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This document is based on a consensus of evidence and/or clinical expert opinion.

It is intended that this Care Pathway be reviewed and revised as needed by April 2020.

Table of Contents

General Age Range Guide

Infant	Preschool	Child	Adolescent	Adult
0 to 2 years	2 to 6 years	6 to 12 years	12 to 18 years	≥ 18 years

Abbreviation Guide

AC . . . . . Activity Coefficient
BMI . . . . . Body Mass Index
CFA . . . . . Coefficient of Fat Absorption
CFTR . . . . Cystic Fibrosis Transmembrane Conductance Regulator
COA . . . . Coefficient of fat absorption
CRP . . . . C-reactive protein
FE1 . . . . . Fecal pancreatic Elastase-1
DEE . . . . . Daily Energy Expenditure
DER . . . . Daily Energy Requirement
FEV_1 . . . . Forced Expiratory Volume in 1 second
GERD . . . . gastro-esophageal reflux disease
LGTH . . . . Length
mos. . . . . Months
PCTL . . . . Percentile
PERT . . . . Pancreatic enzyme replacement therapy
PI . . . . . Pancreatic Insufficiency
PS . . . . . Pancreatic Sufficiency
RBP . . . . . Retinol Binding Protein
Wt. . . . . Weight
CRFD . . . . Cystic Fibrosis-Related Diabetes

Introduction

Malnutrition is both a frequent feature and a comorbidity of cystic fibrosis, with nutritional status strongly associated with pulmonary function and survival. Nutritional management is therefore a standard of care in CF.

Best Practice Recommendations

The following six recommendations are taken from Section 4 of the European Cystic Fibrosis Society Standards of Care: Best Practice guidelines.[1]

Goals for Nutritional Status

- Infants and children should grow normally. By the age of 2 years, infants achieving normal weight and height percentiles similar to non-CF children.
- Older children and adolescents should achieve the 50th percentile for body mass index (BMI).
- In adults, absolute BMI should be maintained above 20 kg/m^2, ideally, 22 kg/m^2 (females) and 23 kg/m^2 (males).
- All patients should have normal fat-soluble vitamin and micronutrient status.
- Essential fatty acid status should be monitored, if the assay is available.

Routine Monitoring of Nutritional Status

- Until growth ceases, accurate measurement of weight (kg), length or height (cm), and head circumference (cm) (up to 2 years of age) should be made at each clinic visit.
- In adults, height should be measured annually.
- Measurements should be converted to BMI (> 2 years) and compared to national reference charts. Special attention is needed for toddlers and adolescents due to rapid growth velocity.

Exocrine Pancreatic Insufficiency and Adequate Pancreatic Enzyme Replacement

- Confirmation of exocrine Pancreatic Insufficiency (PI) is required. Coefficient of fat Absorption (COA) is the 'gold standard', but is cumbersome. Fecal pancreatic elastase-1 (FE1) is simple and reliable from two weeks of age in the absence of liquid stools.
- Patients with Pancreatic Sufficiency (PS) should be monitored by annual FE1 during infancy and childhood, and, if indicated, in adulthood. FE1 should also be checked during periods of failure to thrive, weight loss, or diarrhea.

- Adequate pancreatic enzyme replacement therapy (PERT) is determined clinically, monitoring nutritional status, signs and symptoms of malabsorption, and excessive appetite with poor weight gain. Inappropriate doses of PERT could result in abdominal pain and constipation.

Ongoing Preventative Nutritional Care

- CF centres should be familiar with the recommendations for age-appropriate dietetic advice directed by CF dietitians. This includes:
  - Assessment of PI and administration of PERT.
  - Selection of appropriate diet, with attention to a high fat intake.
  - Behavioural therapy to achieve positive mealtime experiences.
  - Providing sodium supplementation, when necessary, with special awareness in newborn screened infants.
  - Supplementing fat soluble vitamins, as indicated by laboratory testing.
- Women with CF who plan their pregnancies should receive pre-conception advice to improve their nutritional status.

Monitoring for Malnutrition

- Early intervention is essential to avoid significant loss of weight or growth.
- Evaluation should be triggered by
  - weight loss, or
  - decline in weight or length/height percentile (< 2 years of age), or
  - decline in BMI percentile for age and gender (> 2 years of age), or
  - poor linear growth (< 18 years), or
  - decline in BMI (> 18 years).
- Diagnosing the cause of malnutrition relies on a careful assessment and a multidisciplinary approach. Potential causes of malnutrition:
  - insufficient food intake
  - excessive stool energy losses (inadequate PERT or poor adherence)
  - Giardia infection
  - celiac disease
  - hypercatabolism from pulmonary disease
  - vomiting or gastroparesis
  - gastro-esophageal reflux disease (GERD)

- abnormal glucose metabolism (glycosuria, impaired glucose tolerance, CF related diabetes)
- distal intestinal obstruction syndrome
- psychological impacts of CF

Malnutrition and Options for Intervention

- Interventions should be tried stepwise for a limited period of time or until nutritional status is optimized, depending on the severity of malnutrition and the age of the patient.

Stepwise Intervention for Malnutrition	
Anticipatory guidance	Reinforce adherence to diet, sodium, and enzyme recommendations, using behavioral modification or motivational interviewing.
Moderate malnutrition	Oral supplements should be used as additional calories in a time-limited trial or temporarily as meal replacement for ill patients. Temporary nasogastric (NG)/nasojejunal (NJ) feeds may be useful.
Severe malnutrition	Enteral feeding via NG, gastrostomy, or jejunostomy tubes usually improves and maintains nutrition in a patient with CF.

- Other therapies:
  - Cyproheptadine and growth hormone are not part of routine management.
  - Parenteral nutrition is only appropriate when enteral nutrition is impossible or fails.
- Nutritional rehabilitation can take 3 to 6 months, so if being used pre-operatively should start well ahead of an anticipated operation (e.g. organ transplantation).

For more information on preventing, screening, diagnosing, and treating impaired bone health, see Cystic Fibrosis Care Guidelines for CF-Related Bone Disease

Ongoing Monitoring and Interventions

Overview Based on Nutritional Status

Wt. = Weight LGTH = Length PCTL = Percentile BMI = Body Mass Index mos. = months

	0 to 24 months			2 to 18 years			>18 years*		
	Weight for Length (PCTL)			Body Mass Index (PCTL)			Body Mass Index (kg/m2)		
	≥50th [2]	15 to 50th (z score <1)	<15th (z score <2)	≥ 50th [2]	15 to 50th (z score <1) wt. loss or no wt. gain x2 to 4mos [3]	< 15th	≥ 20 [2]	18.5 to 20 or wt. loss of 5% x2mos[3]	< 18.5
Education/Counselling									
education and behavior counselling for families, focusing on increasing fat and protein intake to prevent/ delay loss of muscle mass & function[3, 4, 5]	X	X	X	X	X	X			
nutrition review focusing on fat and protein intake to prevent/ delay loss of muscle mass and function [3]	X	X	X	X	X	X	X	X	X
psychology/social work consult [6, 7]		X	X		X	X		X	X
enteral nutrition support discussion to optimize growth velocity or nutritional status [8]		X			X			X	

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Nutrition Therapy									
CF fat soluble vitamin/ sodium +/- calcium/iron/ zinc supplementation[3, 9, 10] with repeat bloodwork 3 to 6 months after initiation or change in therapy	X	X	X	X	X	X	X	X	X
annual nutrition bloodwork to include CBC/ iron status/ zinc/ fat soluble vitamins, electrolytes, liver function, urine sodium and C-reactive protein[3]	X	X	X	X	X	X	X	X	X
oral supplement trial to increase growth[6] +/- food intake records		X							
oral supplement trial to increase BMI1 +/- food intake records					X			X	
+/- appetite stimulants[11]		X			X			X	
enteral feeding placement with close follow-up re: tolerance, weight gain, and behaviour issues[5, 8]			X			X			X
annual CFRD screening (>10years old) [12]					X			X	
insulin adjustment for enteral feeding if CFRD present[12]						X			X

	0 to 24 months			2 to 18 years			>18 years*		
	Weight for Length (PCTL)			Body Mass Index (PCTL)			Body Mass Index (kg/m2)		
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Pancreatic Enzyme Replacement Therapy									
optimize dosing for pancreatic insufficiency[13] based on fecal elastase[4, 14] <200 mcg/g stool with yearly monitoring if pancreatic sufficient, sooner with growth failure	X	X	X	X	X	X	X	X	X
adjust dosing for enteral feeding9			X			X			X
Bone Density									
encourage high impact weight bearing activity for bone health[15]				X	X	X	X	X	X
encourage weight bearing and resistance activities[15]							X	X	X
Bone Mineral Density screening using DEXA (≥ 8 to 10 years old)[15]					X	X		X	X
Follow-up									
weekly then monthly follow-up anthropometrics & CF team review with prompt pulmonary exacerbation treatment to lessen adverse nutrition effects[3, 15]	X								
quarterly follow-up anthropometrics & CF team review, including FEV_1 for children ≥6 y (consider testing age 3 to 5) with prompt pulmonary exacerbation treatment to lessen adverse nutrition effects[3,5,15]				X					

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	Weight for Length (PCTL)			Body Mass Index (PCTL)			Body Mass Index (kg/m2)		
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regular follow-up anthropometrics & CF team review[3, 15] including FEV_1 with prompt pulmonary exacerbation treatment to lessen adverse nutrition effects							X		
malnutrition assessment re: nutrition support goals[3]		X	X		X	X		X	X
GI/ endocrine consult if indicated		X			X			X	
endocrine consult			X			X			X
GI/surgical consult if surgical placement of feeding tube indicated			X			X			X

	0 to 24 months			2 to 18 years			>18 years*		
	Weight for Length (PCTL)			Body Mass Index (PCTL)			Body Mass Index (kg/m2)		
	≥50th [2]	15 to 50th (z score <1)	<15th (z score <2)	≥ 50th [2]	15 to 50th (z score <1) wt. loss or no wt. gain x2 to 4mos [3]	< 15th	≥ 20 [2]	18.5 to 20 or wt. loss of 5% x2mos[3]	< 18.5
annual assessment for pubertal status[16] females ≥ 9 years: • physician exam for breast and pubic hair (Tanner Stage development) • inquire as to menarcheal status • record month and year of menarcheal age on growth chart males ≥ 10 years: • physician exam for genital development and pubic hair (Tanner Stage development)				X	X	X			

*CFF recommendation: BMI women ≥22 men ≥23[1]

Energy Intake

- Percentage of energy level requirements for patients compared to those of the same age in the healthy population:[3]

Age	Energy Level	Detail
0 to 24 months	110 to 200%	Adapt energy intake to achieve weight for length ≥50th PCTL
2 to 18 years	110 to 200%	Adapt energy intake to achieve target BMI ≥50th PCTL
>18 years	110 to 200%	Adapt energy intake to achieve BMI targets*

*CFF recommendation: BMI women ≥22 men ≥23[1]

- Methods for determining energy level requirements according to the US Cystic Fibrosis Foundation[16,17, 18]

1. Calculate BMR(kcals) from weight (kg) using World Health Organization equations

Age	Females	Males
0 to 3 years	61.0 x wt - 51	60.9 x wt - 54
3 to 10 years	22.5 x wt + 499	22.7 x wt + 495
10 to 18 years	12.2 x wt + 746	17.5 x wt + 651
18 to 30 years	14.7 x wt + 496	15.3 x wt + 679

2. Calculate Daily Energy Expenditure (DEE) by multiplying BMR by Activity Coefficient (AC) Plus Disease Coefficients

Activity (AC)		Disease Coefficients		DEE
Confined to bed:	BMR x 1.3	FEV_1 >80% predicted:	0	BMR x (AC + 0)
Active:	BMR x 1.7	FEV^1 40 to 79% predicted:	0.2	BMR x (AC + 0.2)
Sedentary:	BMR x 1.5	FEV^1 <40% predicted:	0.3 to 0.5*	BMR x (AC + 0.3)

*May range up to 0.5 with very serious lung disease

3. Calculate total Daily Energy Requirements (DER) from DEE and Degree of Steatorrhea (COA)
   - COA of 1.1 can be used for pancreatic insufficient patients.
   - No COA needs to be used for pancreatic sufficient patients.

Stool collection for fat is no longer being done in B.C. due to inaccuracy and expense.

Example: 10 y old boy

wt = 32kg, AC = active, FEV_1% predicted = 85%, COA 1.1

a. BMR calculation: 17.5 x 32 + 651 = 1211
b. DEE calculation: 1211 x (1.7 + 0) = 2058
c. DER calculation: 2058 x 1.1 = 2265 kcals/ day

Behaviour Strategies[4, 19]

- Early bonding with infant being held for breast/ formula feeding, responding to baby's cues, limiting noise, light and other distractions facilitates growth.
- Introduction of solids when infant is developmentally ready, between 4 to 6 months of age, with repeated exposure to foods before determining a child dislikes food decreases neophobia.
- Eating with child at regularly scheduled meal and snack times helps with food intake.
- Limit mealtime to 15 minutes for toddlers and use snack times as mini meals.
- Compliment appropriate eating behaviours (e.g. trying new food, taking a bite), and pay minimal attention to behaviour not compatible with eating (refusing food).
- Encourage families to monitor food and energy intake for reassurance of their child's nutrition well-being.
- Discuss nutrition or behaviour issues of concern with dietitian or behaviour counsellor.

Pancreatic Enzyme Replacement Therapy (PERT)

a. PERT Oral replacement therapy

- Enzyme administration guidelines:
  - Enzyme capsules should be swallowed whole.
  - For pediatric patients who cannot swallow pills, enzyme beads can be mixed in a small amount of acidic food (pH <4.5) that does not require chewing (e.g. applesauce).
  - Enzymes are given before and/or during all meals and snacks, including milk and oral supplements.
- Enzyme Replacement Therapy is indicated for all patients under 2 years of age with:
  - 2 CFTR mutations associated with pancreatic insufficiency
  - Fecal elastase <200 mcg/g or CFA <85% (in patients <6 months of age) or other objective evidence of pancreatic insufficiency
  - Unequivocal symptoms of malabsorption while awaiting confirmatory test results

Based on North American CF Foundation Consensus Statement[4,13]

Refer to the 'Guidelines for Management of the Infant Diagnosed with Cystic Fibrosis' from the British Columbia Provincial Newborn Screening Program

- Recommended dosages:

Age	Dose
0 to 12 months	2000 to 4000 U lipase/120 mL infant formula or breast milk
12 months to 4 years	1000 U lipase/kg/meal initially, then titrate per response
> 4 years and adults	500 U lipase/kg/ meal initially Up to maximum of 2500 U lipase/ kg meal **or** 10, 000 U lipase/kg/day or 4000 U lipase/gram fat ingested/day **Plus** one half of the standard meal dose to be given with snacks
Low or phthalate-free enzymes may be preferred	

b. PERT via enteral feeding[8]

- Suggested dosing options:
  - Per gram fat dose (1000 to 4000 U lipase/ gram fat; mean 1800 U lipase/g fat)
  - Per meal dose 500 to 2500 U lipase/kg/meal
- Suggested PERT timing:
  - Oral administration prior to bolus or continuous enteral feeding
  - Oral administration after continuous enteral feeding
  - Oral administration mid continuous enteral feeding
  - Combination of above
- Crushing enzymes to add to enteral feeding is not FDA approved.

Vitamin Supplementation

Vitamin supplementation needs to be individualized. Pancreatic sufficiency may permit use of a standard multivitamin in place of a CF specific brand. The vitamin comparison chart from MVW Nutritionals (MVWnutritionals.com) highlights vitamin content differences between CF vitamin formulations in North America. In British Columbia, MVW Nutritionals is the current CF vitamin supplement approved under Pharmacare Plan D.

MVW Complete Formulation		
Form	**Dose**	**Age**
Drops	0.5 mL	0 to 12 months
	1 mL	1 to 3 years
Chewables	1 tablet	4 to 8 years
	2 tablets	≥9 years
Softgels	1 capsule	4 to 9 years
	2 capsules	≥10 years

Following NBS (newborn screening) diagnosis, multivitamins containing vitamin A are started once serum albumin normalizes (> 33), due to risk of pseudotumor cerebri.

CF vitamins should be taken with pancreatic enzymes and fatty food to increase vitamin absorption.

If biochemical deficiency is detected despite adequate vitamin supplementation, assess for poor adherence or poor absorption before adjusting dosage.[3]

a. Guidelines for Pancreatic Insufficiency

Vitamin	Supplementation	Serum Reference Values Monitoring Frequency
Fat-soluble vitamins[3]		
Vitamin A	Amounts dependent on serum values and supplement form **Retinol (preformed)** ■ Start low ■ Adapt rapidly to target normal serum reference range **Beta carotene (provitamin A)** ■ Prescribe 1 mg/kg/day (maximum 50 mg/day) for 12 weeks ■ Follow with maintenance dose (maximum 10 mg/day)	■ Normal reference range provided by laboratory processing sample ■ Monitor annually ■ Check 3 to 6 months after a dosage change ■ Test when pregnancy is considered ■ Consider Retinol Binding Protein (RBP) measurement when liver disease present

Vitamin	Supplementation	Serum Reference Values Monitoring Frequency
Vitamin D	Dependent on serum values, which vary with dietary intake and sun exposure **Vitamin D3 (cholecalciferol)** ■ Starting dose ■ Infants: 400 IU/day (advance to upper limit of 1000 IU/day) ■ All others: 800 IU/day (advance to upper limit of 2000 for children 1 to 10 years, and 4000 IU/day for older) ■ Maintenance dose: adapt to annual serum values (see Recommendations for Vitamin D Deficiency)	■ Serum 25-hydroxyvitamin D minimum 20ng/mL (50nmol/L) ■ Monitor annually, preferably at the end of winter
Vitamins E (tocopherols)	**α-tocopherol** ■ 100 to 400 IU/day ■ 50 IU/day for infants <12 months (1 mg=1.49 IU)	■ Plasma α-tocopherol:cholesterol ratio >5.4mg/g ■ Monitor annually ■ Check 3 to 6 months after a dosage change
Vitamin K	**Vitamin K_1** ■ Infants: 0.3 to 1.0 mg/day ■ Older children and adults: 1 to 10 mg/day	Routine biochemical measurement not widely available
Water-soluble vitamins[3]		
Folic Acid	Women planning pregnancy and during 1st trimester: 400 mcg/day	Check CF vitamin dosage to confirms is contains sufficient folic acid
Vitamin B12	Following ileal resection: 1000 mcg/month IM injections if deficient	

b. Recommendations for Vitamin D Deficiency

- The North American CF Foundation recommends that all patients.[10]
  - have serum 25-hydroxyvitamin D measured to assess Vitamin D status annually, preferably at the end of winter
  - be treated with Vitamin D_3 (cholecalciferol) to achieve and maintain serum 25-hydroxyvitamin D levels of at least 30 ng/ mL (≥75 nmol/L)
  - serum 25-hydroxyvitamin D level below 30 ng/mL (<75 nmol/L) be assessed for adherence to the prescribed regimen

- have serum 25-hydroxyvitamin D levels re-checked 3 months after the dose of Vitamin D_3 has been changed
- with difficult-to-treat Vitamin D deficiency be seen by a specialist with expertise in vitamin D therapy

- Supplementation and Treatment of Deficiency

Dosing with CF-specific vitamins				
Age	Routine dosing	Step 1 Dose increase	Step 2 Dose Titration Maximum	Step 3
0 to 12 months	400 to 500 IU	800 to 1000 IU	Not more than 2000 IU	Endocrine referral may be needed
1 to 10 years	800 to 1000 IU	1600 to 3000 IU	Not more than 4000 IU	
10 to 18 years	800 to 2000 IU	1600 to 3000 IU	Not more than 10,000 IU	
> 18 years	800 to 2000 IU	1600 to 6000 IU	Not more than 10,000 IU	

Sodium Replacement[3, 4, 20]

a. Monitoring

- Assess urine and serum electrolytes annually for inadequacy:
  - urine sodium <10 mmol/L
  - sodium:creatinine ratio <17 to 52 mmol/mmol[20]

b. Supplementation

- Supplement as sodium chloride (5 mL table salt = 100 mmol sodium)

Age	Sodium Supplementation	Detail
Infants		
0 to 6 months	■ 2 to 4 mmol/kg/day ■ (12.5 mmol Na/ day)	For infants at risk of sodium deficiency, give salt in small portions throughout the day, diluted in water or fruit juice
6 to 12 months	■ 25 mmol Na/ day	Infants beginning to eat can have salt sprinkled on food
Infants in special situations	Up to 4 mmol/kg/day	■ Increase sodium Intake in these situations: ■ infants living in hot ambient temperatures ■ infants with increased fluid loss due to vomiting, fever, diarrhea, or tachypnea ■ infants with ostomies

Age	Sodium Supplementation	Detail
> 1 year to adult	Salty foods or sodium chloride capsules, tablets or vials	Supplement in stress situations when excessive sweating is expected (e.g. fever, exercise, sports, hot weather)

Iron Replacement[3]

a. Monitoring

- Monitor children, adolescent, and adult patients annually using serum iron determination, differentiating between iron deficiency and anemia of chronic inflammation.
- If iron deficiency is suspected, increase frequency of monitoring.
- Laboratory assessment of anemia using normal reference range provided by the laboratory processing the sample.

	Indicators of Anemia (based on normal reference range)		
Test	Iron Deficiency Anemia	Anemia of Chronic Inflammation	Both forms of anemia
Serum iron	Below normal	Below normal	Below normal
Serum ferritin	Below normal	Above normal (levels <100 could also reflect iron deficiency)[21]	Varies
Total iron binding capacity	Above normal	Below normal **or** Normal	Varies
% transferrin saturation	Below normal	Below normal	Below normal

b. Oral Supplementation[22]

- In cases of iron deficiency, resolve underlying inflammation first. Supplement with iron only if deficiency persists. If ongoing evidence of deficiency persists despite supplementation, cause of iron deficiency anemia need to be further investigated.

Iron Salt supplements	Form (Elemental) and Dose
Ferrous Gluconate	Tablet: 300 mg (35 mg)
Ferrous Sulfate	Tablet: 300 mg tab (60 mg) Liquid: (15 mg/mL), (6 mg/mL)
Ferrous Fumarate	Tablet: 300 mg (100 mg) Liquid: (20 mg/mL)
Polysaccharide Iron Complex (Feramax)	Capsule: 150 mg (150 mg) Powder: (15 mg/1.25 mL)
Iron Polypeptide (Proferrin)	Tablet: 398 mg (11 mg)

	Dosage for Repletion	
Iron Salt	(Of elemental iron) Ferrous Gluconate Ferrous Sulfate Ferrous Fumarate	Iron Polypeptide (Proferrin)
Pediatric	3 to 6 mg/kg	Not indicated
Adult	100 to 200 mg/day or 2 to 3 mg/kg/day in 2 to 4 divided doses	1 tablet BID to TID
Elderly	15 mg/day	1 tablet OD
Pregnant	30 to 120 mg/day	1 tablet BID to TID

c. Intravenous supplementation

- Intravenous supplementation may be indicated for intestinal malabsorption or intolerance of oral iron supplement. GI tolerance needs to be evaluated during iron supplementation. Smaller, less frequent doses in adults enhance absorption and GI tolerance.[23] If iron IV supplementation required use hospital based protocols for infusion and dosage.

Zinc Replacement[3]

a. Monitoring

- Plasma zinc level is not a sensitive marker of zinc status as it is an acute phase reactant.[24]
- Current CF vitamin therapy provides appropriate zinc supplementation. Before considering additional supplementation, check adherence to the recommended vitamin dosage.

b. Supplementation[3, 4]

- Consider zinc supplementation for people with CF who are at risk of zinc insufficiency (e.g. growth retardation, increased susceptibility to infections, delayed sexual maturation, eye problems associated with Vitamin A insufficiency, and anorexia).
- Divided doses are recommended for supplementation.

Age	Zinc Supplementation*	Recommended dosing period
0 to 2 years and at risk of zinc insufficiency	1 mg/kg/day (maximum 15 mg/day)	6 months
2 to 18 years and at risk of zinc insufficiency	15 mg/day	6 months
> 18 years and at risk of zinc insufficiency	25 mg/day	6 months

*Recommended total maximum dosing/day including dosage obtained from CF Vitamin Therapy

Enteral Tube Feeding (CF Foundation Statements)[8]

a. CF Foundation recommends:

- Enteral tube feeding as a means to improve age-dependent anthropometrics in patients that are unable to consume adequate calories and protein to meet growth/weight maintenance goals, despite appropriate evaluation and intervention by a multidisciplinary team.
- Evaluation by a multidisciplinary CF team prior to enteral feeding tube placement to identify and treat conditions that might be contributing to nutritional decline.
- Patient and family education about nutritional care including the role of enteral tube feeding done throughout the lifetime of the patient.
- The risks of certain conditions to be considered and discussed with patients prior to the placement of an enteral feeding tube including but not limited to: coagulopathy, severe obstructive lung disease, ascites, portal hypertension, history of abdominal surgery, peritoneal dialysis, or alcohol and/or substance abuse.
- Nasoenteral tube feeding for patients who require short-term (less than 3 months) nutritional repletion.
- Discussion of third party/individual coverage of supplies and formula with patients prior to placement of an enteral feeding tube.
- A comprehensive history and physical exam by the medical team performing the procedure, with specific attention to factors that represent potential complications, be performed in advance of scheduling the placement of the percutaneous or surgical enteral feeding tube.
- Clinical assessment of gastroesophageal reflux be performed prior to enteral feeding tube placement.
- To mitigate perioperative risk, the CF provider managing the pulmonary care of the patient determine timing, based on pulmonary status, for percutaneous or surgical enteral feeding tube placement.
- Platelet count and international normalized ratio (INR) be measured prior to percutaneous enteral feeding tube placement.
- Consultation with an anesthesiologist and the consideration of more intensive pulmonary therapy prior to placement of a percutaneous or surgical enteral feeding tube in patients with moderate to severe lung disease.
- Enteral feeding tubes be placed by percutaneous endoscopic, laparoscopic, or radiologic technique when possible as opposed to open surgical techniques.
- Patients who are intolerant of gastric feeding receive jejunal feeding.
- Airway clearance be re-initiated within 24 hours of percutaneous or surgical enteral feeding tube placement in both pediatric and adult patients.

- Optimal post-operative pain management to facilitate re-initiation of airway clearance in both pediatric and adult patients who receive an enteral feeding tube.
- Initiation of a bowel regimen to prevent post-operative constipation or distal intestinal obstruction syndrome, especially those receiving narcotic pain management.
- Adherence to the 2010 Clinical Care Guidelines for Cystic Fibrosis-Related Diabetes in individuals with CF who are using enteral feeding tubes.
- The use of supplemental enteral nutrition for pregnant or lactating female patients who are unable to consume adequate calories and protein to meet nutritional goals despite appropriate evaluation and intervention by a multidisciplinary team.
- The use of CF Foundation Evidence-based Guidelines for Management of Infants with Cystic Fibrosis to choose the best feeding type, breastmilk or formula, for enteral tube feeding in pediatric patients less than 2 years of age.
- Continuous nocturnal infusion for patients who are receiving supplemental enteral tube feeding.
- A comprehensive planning approach with a multidisciplinary CF care team including the managing gastroenterologist, case manager, and home care agency prior to discharge.
- Evaluation by a CF-trained Registered Dietitian (RD) to calculate energy needs and assess optimal enteral tube feeding supplementation from enteral tube feeding.
- Monitoring growth or BMI and tolerance of enteral tube feeding to allow changes if the patient is not meeting goals or tolerating the current regimen.
- Monitoring for the development of an oral aversion, disordered eating, or other related behavioral concerns in patients receiving enteral tube feeding.
- Enteral feeding tube removal follow careful consideration of medical and psychosocial goals.
- Patients who have had enteral feeding tube placement be monitored at least annually by a gastroenterologist, preferably with enteral device experience, in addition to their quarterly CF care centre visit.

b. CF Foundation recommends against:

- Using FEV_1 as an absolute contraindication to percutaneous or surgical enteral tube placement
- Routine pH/impedance or radiographic procedures to assess gastroesophageal reflux prior to percutaneous or surgical enteral feeding tube placement.
- The placement of a percutaneous or surgical enteral feeding tube during acute illness.

c. CF Foundation does not recommend for or against:

- Enteral tube feeding to improve or stabilize pulmonary function in individuals with CF.
- The use of a specific type of formula (polymeric, semi-elemental, elemental) for enteral tube feeding.
- A specific method of providing pancreatic enzyme therapy during enteral tube feeding.
- The routine use of acid blockade during enteral tube feeding.

Endnotes

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