

Review

# Enteral tube feeding for individuals with cystic fibrosis: Cystic Fibrosis Foundation evidence-informed guidelines



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## Abstract

Nutrition is integral to the care of individuals with cystic fibrosis (CF). Better nutritional status is associated with improved pulmonary function. In some individuals with CF, enteral tube feeding can be useful in achieving optimal nutritional status. Current nutrition guidelines do not include detailed recommendations for enteral tube feeding. The Cystic Fibrosis Foundation convened an expert panel to develop enteral tube feeding recommendations based on a systematic review of the evidence and expert opinion. These guidelines address when to consider enteral tube feeding, assessment of confounding causes of poor nutrition in CF, preparation of the patient for placement of the enteral feeding tube, management of the tube after placement and education about enteral feeding. These recommendations are intended to guide the CF care team, individuals with CF, and their families through the enteral tube feeding process.

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**Keywords:** Cystic fibrosis; Enteral nutrition; Gastrostomy; Nasojejun tube; Feeding tube

**Abbreviations:** CF, Cystic Fibrosis; COPD, Chronic obstructive pulmonary disease; CFRD, Cystic Fibrosis Related Diabetes; FEV<sub>1</sub>, Forced Expiratory Volume; GER, Gastroesophageal Reflux; GERD, Gastroesophageal Reflux Disease; GT, Gastrostomy; NG, Nasogastric; NJ, Nasojejun tube; PEG, Percutaneous Endoscopic Gastrostomy.

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## 1. Introduction

Optimal growth in children and weight status in adults are important in maintaining best possible lung function in CF [1–3]. Parents are taught the importance of good nutrition soon after diagnosis, and best practices dictate frequent reassessment of growth and weight through childhood and adulthood [4,5]. Many factors impede achievement of nutritional goals in people with CF [6]. When nutritional progress sufficient to ensure optimal lung function cannot be achieved, the CF Team, patient and family may consider enteral tube feeding to provide adequate nutrition for growth and weight maintenance. Although this strategy has been used and reported since the early 1980's, there are no large-scale clinical trials of enteral tube feeding in individuals with CF. Nonetheless, small single center retrospective studies have demonstrated improved weight gain and some suggest improved pulmonary function [7–19].

The purpose of this guideline is to provide the CF Team with information critical to the use of enteral feeding tubes, including criteria for recommending enteral tube feeding, assessment of confounding causes of poor nutrition, preparation for placement of the enteral feeding tube, and management of the tube after placement. Education of patients and family caregivers on enteral feeding tubes is important throughout the lifespan, to ensure that they are active participants in this important choice.

Finally, the term “enteral feeding tube” may include gastrostomies (GT) (surgical, percutaneous endoscopic gastrostomy (PEG), radiographic insertion), gastrojejunal tubes, nasoenteral tubes (nasogastric (NG), nasojejunal (NJ)), or surgically-placed jejunal tubes for administration of feeding. Where the literature refers to a specific form of enteral feeding tube, that term is used.

## 2. Methods

The CF Foundation (CFF) invited a multidisciplinary team, including pediatric and adult dietitians, gastroenterologists, pulmonologists, a clinical psychologist, social worker, nurse, parent of a child with CF, and an individual with CF to participate in the development of these consensus guidelines. The committee met on March 24, 2015 at the CFF National Office in Bethesda, Maryland to determine the PICO (Population Intervention Control Outcome) questions, structured clinical questions that are used to guide a literature search. The committee was divided into three workgroups: shared decision making, enteral feeding tube management, and management after placement. These workgroups established the PICO questions

for their topics. The committee discussed and, when needed, revised the questions prior to accepting the PICO questions.

The workgroups determined MeSH (Medical Subject Headings) and key terms for the PICO questions. These terms were used to search Medline using the OVID database at Dartmouth.

The literature search resulted in 1080 unique citations, and committee members found an additional 77 titles. After de-duplication, the workgroups reviewed the 1138 unique titles and selected articles relevant to the PICO questions; 421 abstracts were selected for further review; 238 full articles were reviewed. An additional 3 articles were added after the public comment period. The workgroups developed draft recommendation statements to address the PICO questions.

The committee reconvened in September 2015 to revise and adopt the draft recommendation statements presented by the workgroup. The committee established an a priori voting threshold of 80% agreement. All statements reached 100% agreement.

In January 2016, the draft manuscript was distributed for a two week public comment period. The committee reviewed and responded to the public feedback and revised the manuscript as appropriate. One statement was revisited after public comment, and the committee re-voted on the revised statement.

## 3. Discussion of consensus statements

1. The CF Foundation recommends enteral tube feeding as a means to improve age-dependent anthropometrics in individuals with CF that are unable to consume adequate calories and protein to meet growth/weight maintenance goals, despite appropriate evaluation and intervention by a multidisciplinary team.

There are no randomized clinical trials to inform decisions regarding enteral tube feeding and CF, however, multiple retrospective studies suggest that its use can improve age-dependent anthropometrics [7–18,20]. Age-dependent anthropometrics include weight/age percentile, length/age percentile and weight-for-length percentile for 0–2 years; weight/age percentile, stature/age percentile and body mass index (BMI) percentile for 2–20 years; and BMI value for adults [21–23]. Enteral tube feeding can be offered as a means of meeting calorie goals to achieve optimal weight gain when other strategies are not sustainable or successful but should be presented when there is a chance of success; enteral tube feeding is generally not successful at end of life.

2. The CF Foundation does not recommend for or against enteral tube feeding to improve or stabilize pulmonary function in individuals with CF.

Existing data are inadequate to recommend for or against enteral tube feeding as a means to improve or stabilize pulmonary function [7,8,10,11,13–18]. While some of the retrospective studies showed improvement in pulmonary function, this did not occur consistently through all studies. Some studies included very sick patients or those with other co-morbidities, which may have skewed the results [7,11,14,16,17].

3. The CF Foundation recommends evaluation by a multidisciplinary CF team prior to enteral feeding tube placement in individuals with CF, to identify and treat conditions that might be contributing to nutritional decline.

Achieving nutritional goals as defined by the CFF may be complicated by multiple factors [5]. Evaluation and treatment by a multidisciplinary team (Table 2) is recommended to assess for causes of inadequate nutrition achievement, including inadequate caloric intake, gastrointestinal and endocrine complications, pulmonary exacerbation, and behavioral and psychosocial factors. This avoids placement of an enteral feeding tube in an individual with a treatable condition [24–26]. The continued involvement of the multidisciplinary team can improve nutritional outcomes following initiation of enteral tube feeding [11,27–30].

4. The CF Foundation recommends that patient and family education about nutritional care, including the role of enteral tube feeding, be done throughout the lifetime of the individual with CF.

Nutrition education to promote optimal weight status, including enteral tube feeding, should be provided to every patient with CF throughout their lifespans (Table 3). Early introduction of enteral tube feeding as a treatment option allows the patient and family to become comfortable with all of the choices and to fully participate in the decision-making process [17,31]. Previous poor adherence is not an absolute contraindication to enteral feeding tube placement, but should be considered and discussed prior to deciding on enteral nutrition.

Once the decision to place the enteral feeding tube has been made, education should address a pre-, peri- and post-operative clinical plan, as well as discharge needs. After home enteral feeding is established, education should continue to include care of the enteral feeding tube, problem solving and required follow up. It is important to note that education is necessary but may not be sufficient to change behavior [32,33].

5. The CF Foundation recommends that the risks of certain conditions be considered and discussed with individuals with CF 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.

Individual risk factors should be assessed as part of the decision to intervene with an enteral feeding tube [14,34]. These risk factors should be discussed with the care team, patient, and family as part of the decision-making process. For lung transplant candidates and potential candidates, nutritional requirements vary by transplant center. For this reason, discussion with specific transplant centers is the best approach.

6. The CF Foundation recommends against using FEV<sub>1</sub> as an absolute contraindication to percutaneous or surgical enteral tube placement in individuals with CF.

Ten retrospective studies of gastrostomies in 267 individuals with CF report 17 deaths during follow-up of varying length. Two studies reported death in patients with lower FEV<sub>1</sub> [10,11], while another found no association of death with pre-operative FEV<sub>1</sub> [8]. While there is no absolute contraindication for an enteral feeding tube in a patient with low FEV<sub>1</sub>, careful consideration of the capacity for recovery should be made in patients with severe pulmonary dysfunction. Optimal lung function should be attained prior to anesthesia.

7. The CF Foundation recommends nasoenteral tube feeding in individuals with CF who require short-term (less than 3 months) nutritional repletion.

NG feeding avoids the risk of surgery, and placement is easily reversible. In selected patients NG feeds may be suitable for short-term nutritional rehabilitation during an acute illness or as a trial of feeding tolerance prior to GT placement. While suitable for short-term (less than 3 months) feeding, they may not be optimal for patients who require an indefinite period of supplemental feeding [30]. Some individuals with CF have success using long-term nasoenteral feeding, but others have experienced complications, including enteral feeding tube dislodgement, clogging, bleeding, nasal alae erosion, intestinal perforation, and transpyloric migration [34]. NG tubes may be challenging for individuals with nasal polyps [29,35,36].

8. The CF Foundation recommends discussion of third party/individual coverage of supplies and formula with individuals with CF prior to placement of an enteral feeding tube.

The financial burden of enteral nutrition is an important consideration in decision making for individuals with CF and their families. Insurance coverage and out of pocket costs related to enteral nutrition should be investigated well before enteral feeding placement. The CF team should work with the patient and family to identify insurance requirements and program eligibility criteria for enteral nutrition formula, durable medical goods, supplies, home care services, resources and assistance options to help in management of medical costs [37–40].

9. The CF Foundation recommends that a comprehensive history and physical exam, 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 by the medical team performing the procedure in individuals with CF.

In non-CF adults and children, careful pre-procedure assessment has been shown to reduce mortality after GT placement [29]. Ileus, mechanical obstruction, peritonitis, bowel ischemia, and perforation are contraindications to GT placement [30,41]. Relative contraindications to feeding GT include active gastritis, erosive esophagitis, peptic ulcer disease, gastrointestinal bleeding, previous abdominal surgery, intestinal dysmotility, intractable diarrhea, organomegaly, massive ascites, and esophageal or gastric varices. A pre-operative history and physical exam should be directed to assessment of these factors [30,34,41,42].

10. The CF Foundation recommends that clinical assessment of gastroesophageal reflux be performed prior to enteral feeding tube placement in individuals with CF.
11. The CF Foundation recommends against routine pH/impedance or radiographic procedures to assess gastroesophageal reflux in individuals with CF prior to percutaneous or surgical enteral feeding tube placement.

Evidence from the non-CF population shows pre-operative evaluation of gastroesophageal reflux (GER) by pH/impedance or radiographic studies does not predict post-operative reflux, [42,43]. A radiographic study can outline anatomy prior to the procedure, and may be requested by the individual placing the enteral feeding tube. There is no evidence that GT placement induces gastroesophageal reflux disease (GERD) [43–45]. In the non-CF population, approximately 10% of children required an anti-reflux procedure after GT placement [46,47]; clinical assessment was sufficient to determine if a child would do well with GT alone, compared to GT combined with an anti-reflux procedure [48]. In non-CF adults, GER after PEG was more likely in individuals with hiatal hernia or severe reflux esophagitis [49].

Individuals with CF may be at increased risk of GER, bile reflux, and delayed gastric emptying [50–53]. Retrospective studies in CF show that reflux and/or vomiting may occur in up to 50% of those followed for a year or more after GT placement [8–14,17,18,54].

If clinical assessment suggests that the individual with CF has active GERD pre-operatively, this can be investigated, and the patient can be considered for fundoplication at the time of GT placement. Guidelines for evaluation of GER/GERD are available [55,56]; a gastroenterologist is helpful in making these assessments.

12. The CF Foundation recommends that, to mitigate perioperative risk, the CF provider managing the pulmonary care of individuals with CF determine timing, based on pulmonary status, for percutaneous or surgical enteral feeding tube placement.

Pulmonary function in patients requiring an enteral feeding tube ranges from normal lung function to severe obstructive lung disease. For the best outcomes, patients should be at their optimal respiratory status prior to undergoing the procedure.

This may require admission to the hospital for increased airway clearance and intravenous antibiotics. Individuals with better lung function may be able to prepare as outpatients. Consider instructing the patient on a mode of airway clearance that does not require physical manipulation of the abdominal wall such as a Positive Expiratory Pressure device to be utilized immediately after enteral feeding tube placement.

13. The CF Foundation recommends that platelet count and international normalized ratio (INR) be measured in individuals with CF prior to percutaneous enteral feeding tube placement.

Although routine measurement of coagulation and platelets before placement of percutaneous enteral feeding tubes in the general population is not considered necessary [41,42], individuals with CF are at risk of both portal hypertension and fat soluble vitamin deficiency [57,58], and therefore the risk of post-operative bleeding may be higher.

14. The CF Foundation recommends against the placement of a percutaneous or surgical enteral feeding tube during acute illness.

Increased mucus production, airway inflammation, and localized infection occur in the presence of a pulmonary exacerbation and other respiratory illnesses. In chronic obstructive pulmonary disease, these factors increase the risk of immediate post-operative pulmonary complications including pneumonia, atelectasis, bronchopneumonia, hypoxia, and respiratory failure [59,60]. If the need for nutritional support is urgent, consider a nasoenteral tube (see recommendation #7).

15. The CF Foundation recommends consultation with an anesthesiologist and the consideration of more intensive pulmonary therapy prior to placement of a percutaneous or surgical enteral feeding tube in individuals with CF and moderate to severe lung disease.

Options for anesthesia and pain management include general anesthesia with intubation, anesthesia with non-invasive ventilation, epidurals, and conscious sedation [59,61,62]. There is no evidence supporting a specific mode of anesthesia/pain management in this population. Prior to the procedure, the anesthesiologist, procedural physician, and primary pulmonologist should consult to review patient preferences, alternative modes of anesthesia, baseline respiratory status of the individual, and the length of the procedure.

16. The CF Foundation recommends that enteral feeding tubes be placed by percutaneous endoscopic, laparoscopic, or radiologic technique when possible as opposed to open surgical techniques in individuals with CF.

There is no evidence for superiority among PEG, laparoscopic, and radiographic GT placement, in terms of adequacy of placement or complication rate [41,63,64]. Single center

studies may demonstrate superiority of a procedure [65,66], but it is not clear if these are the result of local expertise or of true superiority of a specific method. There is a significant increase in complications associated with open surgical GT placement [67]. This may be due to both the indications for open placement and the invasiveness of the operation, but suggest that open placement should be performed only when there are clear contraindications to less invasive placement techniques.

17. The CF Foundation recommends that individuals with CF who are intolerant of gastric feeding receive jejunal feeding.

Patients with gastroparesis, severe GER, and pancreatitis may benefit from transpyloric feeding (either NJ or gastro-jejunal feeding) to reduce gastric distention and reflux [29]. Jejunal tubes are associated with risk of dislodgment, clogging, leaking, and perforation, and placement may require a radiographic procedure, increasing radiation exposure [68]. Jejunal feeds must be delivered by continuous drip, which may be inconvenient. Consultation with a gastroenterologist may be helpful.

18. The CF Foundation recommends that airway clearance be re-initiated within 24 h of percutaneous or surgical enteral feeding tube placement in children and adults with CF.  
19. The CF Foundation recommends optimal post-operative pain management to facilitate re-initiation of airway clearance in adults and children with CF who receive an enteral feeding tube.

Pneumonia, atelectasis, and pulmonary infection are well-established complications of anesthesia and surgical procedures. Abdominal operations may carry higher risk of pulmonary complications due to pain with cough and deep breathing [69,70]. For this reason, individuals with CF must receive post-operative pain management and airway clearance within 24 h after the procedure to recruit atelectatic lung. Pain is often undertreated both postoperatively and when a patient is managing a chronic condition [71].

20. The CF Foundation recommends initiation of a bowel regimen to prevent post-operative constipation or distal intestinal obstruction syndrome in individuals with CF, especially those receiving narcotic pain management.

Delayed gastrointestinal motility secondary to anesthesia and pain medications may place the individual with CF at high risk for constipation and/or distal intestinal obstructive syndrome in the post-operative period. Initiation of a bowel regimen immediately post-operatively is recommended. A gastroenterologist may be helpful in managing post-operative bowel conditions.

21. The CF Foundation recommends adherence to the 2010 clinical care guidelines for cystic fibrosis-related diabetes in individuals with CF who are using enteral feeding tubes.

Regardless of formula selected or method of delivery, enteral feeding may contribute to hyperglycemia in individuals

with CF. When enteral tube feeding is initiated, individuals with CF should be screened with mid- and immediate post-feeding plasma glucose measurement. These measurements should be repeated monthly by self-monitoring blood glucoses. Suspicion of cystic-fibrosis related diabetes developing during enteral tube feedings should prompt referral to an Endocrinologist [72,73].

22. The CF Foundation recommends the use of supplemental enteral nutrition for pregnant or lactating women with CF who are unable to consume adequate calories and protein to meet nutritional goals despite appropriate evaluation and intervention by a multidisciplinary team.

No data is available to guide management of enteral feeding during pregnancy in the woman with CF. Data from the general population shows that optimizing nutrition in the preconception period is important for optimal pregnancy outcome; malnourished pregnant women are more likely to deliver low birth-weight infants with increased perinatal morbidity [74]. If intake is not adequate to maintain maternal and fetal health, enteral feeding can be discussed. Limited evidence shows that both NG and PEG tube placement have been safely performed in pregnancy [75]. With the limited evidence for specific methods, the least invasive form of supplemental nutrition that meets the needs of the mother and baby is best. Careful discussion of risks and benefits with the care team and the patient is crucial to making the best choice for mother and baby.

23. The CF Foundation recommends the use of Cystic Fibrosis 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 children with CF under 2 years of age.

In infants with enteral feeding tubes, the CFF evidence-based guidelines for the management of infants with CF should be followed with respect to type of feed used [4]. CF infants with additional gastrointestinal diagnoses such as intestinal failure or formula protein intolerance may benefit from an evaluation by a pediatric gastroenterologist with expertise in CF.

24. The CF Foundation recommends continuous nocturnal infusion for individuals with CF who are receiving supplemental enteral tube feeding.

The published reports of enteral feeding in CF describe overnight continuous feeding; bolus feeding was used in 5% or less [9,11,12,76]. Although intermittent feeding is more physiological, the advantages of continuous overnight feeding include the ability to provide more nutrients, potentially improve absorption, and allow daytime consumption of an oral diet [30,77]. Families and patients should be educated on advantages and disadvantages of different types of feeding administration and work with the CF team to develop an individualized plan that accommodates nutrition and lifestyle needs.

25. The CF Foundation does not recommend for or against the use of a specific type of formula (polymeric, semi-elemental, elemental) for enteral tube feeding in individuals with CF

In published reports of enteral feeding in CF, the majority of patients received polymeric formula; a minority received semi-elemental formula [9,11,12,76]. For each study, enteral feeding

Table 1  
Consensus statements.

Statement
1. The CF Foundation recommends enteral tube feeding as a means to improve age-dependent anthropometrics in individuals with CF that are unable to consume adequate calories and protein to meet growth/weight maintenance goals, despite appropriate evaluation and intervention by a multidisciplinary team.
2. The CF Foundation does not recommend for or against enteral tube feeding to improve or stabilize pulmonary function in individuals with CF.
3. The CF Foundation recommends evaluation by a multidisciplinary CF team prior to enteral feeding tube placement in individuals with CF, to identify and treat conditions that might be contributing to nutritional decline.
4. The CF Foundation recommends that patient and family education about nutritional care including the role of enteral tube feeding be done throughout the lifetime of the individual with CF.
5. The CF Foundation recommends that the risks of certain conditions be considered and discussed with individuals with CF 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.
6. The CF Foundation recommends against using FEV <sub>1</sub> as an absolute contraindication to percutaneous or surgical enteral tube placement in individuals with CF.
7. The CF Foundation recommends nasogastric tube feeding in individuals with CF who require short-term (less than 3 months) nutritional repletion.
8. The CF Foundation recommends discussion of third party/individual coverage of supplies and formula with individuals with CF prior to placement of an enteral feeding tube.
9. The CF Foundation recommends that a comprehensive history and physical exam, 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 by the medical team performing the procedure in individuals with CF.
10. The CF Foundation recommends that clinical assessment of gastroesophageal reflux be performed prior to enteral feeding tube placement in individuals with CF.
11. The CF Foundation recommends against routine pH/impedance or radiographic procedures to assess gastroesophageal reflux in individuals with CF prior to percutaneous or surgical enteral feeding tube placement.
12. The CF Foundation recommends that, to mitigate perioperative risk, the CF provider managing the pulmonary care of individuals with CF determine timing, based on pulmonary status, for percutaneous or surgical enteral feeding tube placement.
13. The CF Foundation recommends that platelet count and international normalized ratio (INR) be measured in individuals with CF prior to percutaneous enteral feeding tube placement.
14. The CF Foundation recommends against the placement of a percutaneous or surgical enteral feeding tube during acute illness.
15. The CF Foundation recommends consultation with an anesthesiologist and the consideration of more intensive pulmonary therapy prior to placement of a percutaneous or surgical enteral feeding tube in individuals with CF and moderate to severe lung disease.
16. The CF Foundation recommends that enteral feeding tubes be placed by percutaneous endoscopic, laparoscopic, or radiologic technique when possible as opposed to open surgical techniques in individuals with CF.
17. The CF Foundation recommends that individuals with CF who are intolerant of gastric feeding receive jejunal feeding.
18. The CF Foundation recommends that airway clearance be re-initiated within 24 h of percutaneous or surgical enteral feeding tube placement in children and adults with CF.
19. The CF Foundation recommends optimal post-operative pain management to facilitate re-initiation of airway clearance in adults and children with CF who receive an enteral feeding tube.
20. The CF Foundation recommends initiation of a bowel regimen to prevent post-operative constipation or distal intestinal obstruction syndrome, in individuals with CF, especially those receiving narcotic pain management.
21. The CF Foundation recommends adherence to the 2010 Clinical Care Guidelines for Cystic Fibrosis-Related Diabetes in individuals with CF who are using enteral feeding tubes.
22. The CF Foundation recommends the use of supplemental enteral nutrition for pregnant or lactating women with CF who are unable to consume adequate calories and protein to meet nutritional goals despite appropriate evaluation and intervention by a multidisciplinary team.
23. The CF Foundation recommends the use of Cystic Fibrosis 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 children with CF under 2 years of age.
24. The CF Foundation recommends continuous nocturnal infusion for individuals with CF who are receiving supplemental enteral tube feeding.
25. The CF Foundation does not recommend for or against the use of a specific type of formula (polymeric, semi-elemental, elemental) for enteral tube feeding in individuals with CF.
26. The CF Foundation does not recommend for or against a specific method of providing pancreatic enzyme therapy during enteral tube feeding in individuals with CF.
27. The CF Foundation does not recommend for or against the routine use of acid blockade during enteral tube feeding in individuals with CF.
28. The CF Foundation recommends a comprehensive planning approach with a multidisciplinary CF care team including the managing gastroenterologist, case manager and home care agency prior to discharge.
29. The CF Foundation recommends evaluation by a CF-trained Registered Dietitian Nutritionist (RDN) to calculate energy needs and assess optimal enteral tube feeding supplementation from enteral tube feeding in individuals with CF.
30. The CF Foundation recommends monitoring growth or BMI and tolerance of enteral tube feeding to allow changes if the individual with CF is not meeting goals or tolerating the current regimen.
31. The CF Foundation recommends monitoring for the development of an oral aversion, disordered eating, or other related behavioral concerns in individuals with CF receiving enteral tube feeding.
32. CF Foundation recommends that enteral feeding tube removal follow careful consideration of medical and psychosocial goals for individuals with CF.
33. The CF Foundation recommends that individuals with CF 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 center visit.

Table 2  
The CF multidisciplinary team.

Healthcare professional	Role
*Pulmonologist	Assess pulmonary health, determine preoperative pulmonary preparation; consult with anesthesia, initiate airway clearance port-procedure
*Registered Dietitian/ Nutritionist	Assess individual nutritional needs and dietary intake Recommend nutritional interventions including enteral tube feeding (type of formula/nutrition) given individual patient needs Monitor nutritional intervention and recommend adjustments to feeding schedule (timing, rate and volume) and formula based on weight change and patient/family preferences
*Gastroenterologist	Assess for other treatable causes of poor appetite or malabsorption Recommend enteral tube feeding (type of formula/nutrition, mode of providing enteral nutrition) given individual patient needs; Manage enteral feeding tube and complications long-term
*Endocrinologist	Assess for other causes of poor growth or malabsorption
Clinical Psychologist	Assess behavioral, family, and psychosocial contributors to problems with attaining adequate energy intake and optimal growth Participate in evidence-based behavioral therapy Provide support with adherence to enteral nutrition therapy
*Nurse	Educate family, patient, other care providers on use of enteral nutrition May be involved in case management (coordinating home health, insurance approval)
*Social worker	Provide support with adherence to enteral nutrition therapy May be involved in case management (coordinating home health, insurance approval)
*Respiratory Therapist/Physical Therapist	Educate and assist with airway clearance options and techniques prior to enteral feeding tube placement and in the post-operative period to optimize lung function

Ideally, all members of the team have training in CF management. Each member of the team has both unique and overlapping knowledge and responsibilities. The final recommendations are the sum of their assessments. \* Mentoring and/or training program available through CFF.

volumes were adjusted to individual patient needs and provided between 25 and 60% of total nutritional needs [9,11,12,76]. A single study using a crossover design in 16 patients aged 4–20 years determined a polymeric formula with enzyme replacement was absorbed as well as a semi-elemental formula in individuals with CF who are pancreatic insufficient [78].

26. The CF Foundation does not recommend for or against a specific method of providing pancreatic enzyme therapy during enteral tube feeding in individuals with CF.

In the absence of clinical trials, no specific recommendations can be made regarding the use of pancreatic enzyme therapy with enteral feeding. It is common for individuals with CF on enteral feeding to take enzymes at the beginning and end of a nocturnal feed, and if possible, in the middle of the feed [72,78–82]. Some centers suggest crushing or dissolving pancreatic enzymes in the formula; there is no evidence this is effective, and it is against manufacturer guidelines. See Table 4.

27. The CF Foundation does not recommend for or against the routine use of acid blockade during enteral tube feeding in individuals with CF.

Acid suppression to improve fat absorption in individuals with CF and persistent steatorrhea has produced conflicting results. Some studies show improvement, [83–87] and others show no improvement in coefficient of fat absorption [77]. No recommendation can be made regarding routine addition of acid suppression or whether there is an advantage of proton pump inhibitors over histamine blockers. In patients with refractory steatorrhea, the currently available data may suggest a trial of acid suppression [83–87].

28. The CF Foundation recommends a comprehensive planning approach with a multidisciplinary CF care team including the managing gastroenterologist, case manager and home care agency prior to discharge.

Evidence suggests that home enteral feeding is relatively safe with few reported adverse events [7–10]. Clinical decisions, including initiation and advancement, preparation and hang-time of formula, drug-nutrient interactions, and monitoring, should comply with peer-reviewed, evidence-based recommendations [29,30]. Risk for refeeding syndrome should be evaluated prior to initiation of enteral nutrition [29,30,88]. Tolerance of enteral feeding at goal rate should be assessed; if goal rate is not reached prior to discharge, consider follow up with CF team within 2–4 weeks.

Care coordination among hospital discharge planners, home care agencies, and the primary ordering provider can mitigate logistical complications in home care such as incomplete home care orders, delayed formula deliveries, missing equipment, and reimbursement obstacles. A single, coordinated discharge plan including recommendations from all participating services is essential in preparing instructions after GT placement. Clinical care-givers should observe patient/family performing enteral nutrition management tasks to ensure competency prior to discharge. Ongoing communication between the primary managing provider and the home nutrition support team is necessary for addressing complications that may develop in the home setting and preventing disruption in home care services [89–91]. Support to achieve optimal outcomes is of utmost importance to patients and families.

29. The CF Foundation recommends evaluation by a CF-trained registered dietitian/nutritionist (RDN) to calculate energy needs and assess optimal enteral tube feeding supplementation from enteral tube feeding in individuals with CF.

30. The CF Foundation recommends monitoring growth or BMI and tolerance of enteral tube feeding to allow changes if the individual with CF is not meeting goals or tolerating the current regimen.

Patients utilizing enteral tube feeding should meet with a CF-trained RDN regularly to ensure goals are achieved [12]. Individuals should have age-appropriate anthropometric

Table 3

Clinical and educational care pathway for enteral tube feedings.

At diagnosis of CF and throughout the life-span	<p><i>Clinical</i></p> <ul style="list-style-type: none"> <li><input type="checkbox"/> Establish and maintain care with dietitian/nutritionist (RDN) trained in the care of persons who have CF.</li> </ul> <p><i>Education</i></p> <ul style="list-style-type: none"> <li><input type="checkbox"/> Introduce enteral tube feeding as a potential treatment option to help maximize nutrition (growth in children/weight in adults)</li> <li><input type="checkbox"/> Frame with a positive attitude – not a failure, but rather a tool to ensure maintenance of a nourished state to promote normal growth in children and weight in adults</li> </ul>
When growth/weight maintenance is faltering	<p><i>Clinical</i></p> <ul style="list-style-type: none"> <li><input type="checkbox"/> Nutrition assessment by RDN</li> <li><input type="checkbox"/> Implement nutrition support strategies               <ul style="list-style-type: none"> <li>- Oral nutrition supplements</li> <li>- Appetite stimulants</li> <li>- Behavioral modification</li> <li>- Management of GI symptoms</li> <li>- Financial/social support</li> </ul> </li> <li><input type="checkbox"/> CF care team assess potential contributing factors               <ul style="list-style-type: none"> <li>- Medical, psychosocial, &amp; financial</li> </ul> </li> <li><input type="checkbox"/> Referral to Gastroenterology, Psychology, Endocrinology as indicated to identify and treat conditions contributing to nutritional decline</li> <li><input type="checkbox"/> Increase frequency of visits or anthropometric measurements to determine if new strategies are improving growth</li> </ul> <p><i>Education</i></p> <ul style="list-style-type: none"> <li><input type="checkbox"/> Revisit enteral tube feeding as a potential treatment option to help maximize nutrition</li> <li><input type="checkbox"/> Discuss purpose of enteral tube feeding, benefits and potential risks</li> <li><input type="checkbox"/> Begin education early in the “enteral feeding tube decision process” to inform and empower patients and families to fully participate in the decision making process</li> <li><input type="checkbox"/> Discuss logistics/specifics/daily routine of enteral feeding to help alleviate potential stress</li> <li><input type="checkbox"/> Connect patient/family with a CF patient/family experienced with an enteral feeding tube</li> <li><input type="checkbox"/> Review/discuss third party/individual coverage of supplies and formula. This information should be explained to the patient as part of the decision on enteral feeding.</li> <li><input type="checkbox"/> Discuss potential psychological issues (i.e. different than peers, people may stare, body image)</li> <li><input type="checkbox"/> Include “real world” information (i.e. feeding initially time consuming but become routine; activity not impacted once site heals, impact on quality of life)</li> <li><input type="checkbox"/> Utilize available education materials (print, online videos, blogs) that discuss the challenges related to an enteral feeding tube.</li> </ul>
Pre-op (after decision to get an enteral feeding tube has been made)	<p><i>Clinical: These actions may be undertaken simultaneously</i></p> <ul style="list-style-type: none"> <li><input type="checkbox"/> Perform comprehensive medical history and physical examination to identify factors that represent potential contraindications or increase risk of complications with enteral feeding tube placement.</li> <li><input type="checkbox"/> Assess for co-morbidities which can complicate the procedure including- coagulopathy, severe obstructive lung disease, ascites, portal hypertension, history of abdominal surgery, peritoneal dialysis, substance abuse</li> <li><input type="checkbox"/> Determine timing of procedure determined based on pulmonary status</li> <li><input type="checkbox"/> Perform platelet count and INR prior to procedure</li> <li><input type="checkbox"/> Insure patient at baseline state of health and without acute illness.</li> <li><input type="checkbox"/> Consult anesthesia prior to procedure for patients with moderate to severe lung disease</li> <li><input type="checkbox"/> Intensify pulmonary treatments just prior to procedure in patients with moderate to severe lung disease.</li> <li><input type="checkbox"/> Secure third party/individual coverage of supplies and formula</li> <li><input type="checkbox"/> Determine feeding plan: formula, timing of feeding, rate of feeding, enzyme use</li> </ul> <p><i>Education</i></p> <ul style="list-style-type: none"> <li><input type="checkbox"/> Determine a pre-operative clinical plan to include the indication for the enteral feeding tube, proposed length of treatment, enteral feeding tube options, risks and benefits (including psychological issues and increased burden of treatment), and a plan for feeding and long-term follow-up</li> <li><input type="checkbox"/> Review/discuss insurance coverage and how feeding supplies will be obtained</li> <li><input type="checkbox"/> Discuss management plan for immediate post op discomfort</li> <li><input type="checkbox"/> Discuss wound care and healing</li> <li><input type="checkbox"/> Review modified airway clearance techniques for post op period</li> <li><input type="checkbox"/> Patient/family should demonstrate understanding</li> </ul>
Immediate post-operative assessment and care	<p><i>Clinical</i></p> <ul style="list-style-type: none"> <li><input type="checkbox"/> Optimal pain management immediately after the procedure</li> <li><input type="checkbox"/> Re-initiation of airway clearance within 24 h post-operative</li> <li><input type="checkbox"/> Bowel regimen to prevent constipation/DIOS</li> <li><input type="checkbox"/> Initiate feeding regimen with nursing support to assist education of patient and family</li> </ul>

(continued on next page)

Table 3 (continued)

Discharge care	<p><i>Clinical</i></p> <ul style="list-style-type: none"> <li><input type="checkbox"/> Teach blood sugar monitoring per the 2010 CFRD guidelines</li> <li><input type="checkbox"/> Ensure optimal pain management</li> <li><input type="checkbox"/> Review home airway clearances techniques</li> <li><input type="checkbox"/> Insure patient and family can administer and troubleshoot enteral tube feeding</li> </ul> <p><i>Education</i></p> <ul style="list-style-type: none"> <li><input type="checkbox"/> Education topics to include ostomy tube site care, hand hygiene and use of sterile techniques, formula handling and storage according to manufacturer instructions, appropriate operation and cleaning of equipment (i.e. pumps, tubing), recognition of potential problems, and designation of a point of contact if complications develop</li> <li><input type="checkbox"/> Review/discuss insurance coverage and how feeding supplies will be obtained</li> <li><input type="checkbox"/> Comprehensive planning with interdisciplinary staff, hospital case management and receiving home care agencies prior to discharge. This should include identification of financial factors that may further delay care.</li> <li><input type="checkbox"/> Designate contact for problems or complications: Home care company, RDN, Surgeon, CF team, or Gastroenterologist</li> </ul>
Post discharge care and follow up	<p><i>Clinical</i></p> <ul style="list-style-type: none"> <li><input type="checkbox"/> Ongoing follow up with RDN and CF team to track progress toward nutrition goals</li> <li><input type="checkbox"/> Ongoing follow up with Gastroenterologist to monitor for complications of the enteral feeding tube (at least yearly) and other gastrointestinal disease</li> <li><input type="checkbox"/> Monitor for complications requiring referral, including behavioral feeding problems and glucose dysregulation</li> </ul>
When growth/weight maintenance goals are achieved	<ul style="list-style-type: none"> <li><input type="checkbox"/> Discuss any changes to enteral feeding regimen in maintenance period and long-term goals of therapy</li> <li><input type="checkbox"/> Discuss whether enteral feeding tube removal is an option and possible timing of enteral feeding tube removal</li> </ul>

parameters measured at each visit [12]. During visits current enteral feeding use, ad-libitum oral intake, signs and symptoms of gastrointestinal complications, and glucose tolerance should be assessed. Referral to other members of the multidisciplinary team may be valuable during this period [12,92].

31. The CF Foundation recommends monitoring for the development of an oral aversion, disordered eating, or other related

behavioral concerns in individuals with CF receiving enteral tube feeding.

For most individuals with CF, enteral tube feeding supplements rather than replaces oral intake; the balance of intake from oral and enteral tube feeding should be monitored to maintain oral intake while still meeting weight goals. Regular evaluation by the CF-trained RDN should allow early identification of decreasing or

Table 4

Community-derived practical considerations for enteral feeding management after G-tube placement.

Calorie goal	30–65% of total estimated calorie and nutrient needs provided by enteral feeding
Delivery regimen	<p>Overnight continuous feeds (to allow for daytime oral intake)*</p> <p>Intermittent bolus feeds to replace some or all meals</p> <p>Bolus feeds after meals (if oral intake inadequate)</p> <p><i>Note: disadvantages of overnight feeds include enuresis and inadvertent enteral feeding tube disconnections</i></p>
Considerations in formula selection/delivery regimen	<p>Patient medical/surgical history*</p> <p>Anticipated tolerance*</p> <p>Patient/family preferences</p> <p>Formula cost</p> <p>Insurance reimbursement</p> <p>Institution-specific protocols</p>
Formula selection	<p>Standard, polymeric age appropriate formula*</p> <p>Calorically dense formula (1.5 or 2.0 cal/ml) in patients &gt;1 year of age*</p> <p>Semi-elemental, age-appropriate products</p> <p>Home prepared or blenderized formulas (less common)</p>
Pancreatic enzyme replacement therapy dosing	<p>Per gram of fat dose (1000–4000 units lipase/g fat; mean 1800 units lipase/g fat)</p> <p>Meal dose (500–2500 units lipase/kg/meal)</p> <p>Above doses follow CFF guidelines for enzyme administration</p>
Pancreatic enzyme replacement therapy timing	<p>Oral administration prior to bolus or continuous enteral feeding*</p> <p>Oral administration after continuous enteral feeding*</p> <p>Oral administration mid continuous enteral feeding</p> <p>Combination of above methods<sup>#</sup></p> <p>Use of an inline cartridge enzyme delivery system for enteral feeding**</p>

The table represents a query of an online community for CF healthcare professionals conducted in October 2015 to determine current practice for the initiation of enteral feeding in CF patients. Responses from 93 adult and pediatric practitioners indicated widespread practice variation, reflecting the lack of evidence upon which to make management decisions. \*denotes majority of respondents agreed on this. <sup>#</sup>Anecdotally, some centers crush enzymes into the formula prior to delivery. No data supports this practice and crushing enzymes is not FDA approved.

\*\*An inline cartridge enzyme (lipase) delivery system for enteral feeds was approved by the FDA for adults during the development of these guidelines; evaluation of its benefits and limits should be considered before use.

disordered oral intake during enteral tube feeding. The placement of an enteral feeding tube does not preclude continued therapy to address behavioral and/or psychological challenges to improve oral intake [93,94]. Referral to the appropriate therapists may be helpful even after enteral tube feeding placement. Changes should be made to the enteral feeding regimen in a timely manner if the individual is not meeting nutritional or quality of life goals.

32. CF Foundation recommends that enteral feeding tube removal follow careful consideration of medical and psychosocial goals for individuals with CF.

No studies have explored criteria for enteral feeding tube removal. Retrospective studies of individuals with CF reporting outcomes before and after GT placement show improvements in nutritional parameters up to 4 years after placement [12,17]. The decision for removal should include individual tolerance of enteral feeding as well as quality of life factors. Anecdotal practice patterns include demonstration of maintained BMI over a designated period of time without the use of the enteral feeding tube prior to permanent removal.

33. The CF Foundation recommends that individuals with CF 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 center visit.

There are minor complications that may occur at any time after placement. Individuals with enteral feeding tubes need reassessment to ensure the tube is appropriate in size, and they may develop local skin infection or irritation, or leaking at the site. Occasionally, more serious late complications may occur, including gastrointestinal perforation and systemic infection. Patients and families should be provided with training and a resource to manage both feeding and tube-related problems should they occur. While CF-trained RDNs generally monitor the effectiveness of the feeding, they are not trained to manage medical complications of enteral feeding tubes. A gastroenterologist or affiliated provider trained to manage enteral feeding tubes should be involved from the beginning and provide at least annual follow up thereafter.

#### 4. Conclusions and next steps

While this document provides guidance on the use of enteral tube feedings in individuals with CF based on the literature and expert opinion, there are many unanswered questions. Some will only be resolved with prospective multicenter studies. Future research should address the impact of enteral tube feeding on pulmonary function and survival in CF. Studies to determine who benefits most from enteral tube feeding, behavioral therapy, or a combination of the two is an important goal. Multicenter trials may help understand patient characteristics that predict failure of enteral tube feeding. Practitioners would benefit from

evidence-based methods for providing digestive enzymes during enteral tube feeding. The recommendations presented in this guideline (Table 1) serve as a starting point for future studies.

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