

CHAPTER 6 NUTRITION INTERVENTIONS

N. Forgiione, N. Saxby, S. King, R. Cavanagh & T. Crowder

The historical focus of nutrition interventions for people with CF has centred on managing undernutrition¹. Today, practitioners are also faced with the emerging issue of overweight and obesity amongst the CF population^{199,278}. This chapter provides guidance for nutrition interventions based on the nutritional status categories as outlined in [Chapter 5](#) (see table 5a). Table 6a below shows recommended escalation of routine care that should be considered due to the risk of development of undernutrition or overweight and obesity.

Table 6a. Recommended nutrition intervention strategies based on nutrition status category

Nutrition status category	Recommendations for nutrition intervention
Optimal	Routine nutritional care and surveillance +/- education and preventative counselling
Acceptable	Routine nutritional care and surveillance +/- education and preventative counselling
Suboptimal: at risk of undernutrition	Nutritional counselling, behavioural management +/- oral nutrition supplements
Persistent undernutrition	As above, plus medical +/- psychological evaluation investigating factors contributing to undernutrition. Consider intensive nutritional support via enteral nutrition
High BMI : at possible risk related to overweight or obesity	Diet and activity assessment, plus consideration of medical and psychosocial contributing factors. Consider goal-directed nutritional counselling ⁷

6.1 Undernutrition

Despite rapid and continuous advances in CF treatments, suboptimal nutritional status amongst people with CF remains common¹⁸⁸. According to the 2014 Australian Cystic Fibrosis Data Registry report, between 2 to 11 percent of adults with CF have a BMI of less than 18.5 kg/m² (i.e. the International Classification of Diseases (ICD 10) criterion for malnutrition¹⁸⁸). Adult females are almost three times more likely to be undernourished than their male counterparts¹⁸⁸. Undernutrition can be present from birth and the Australian CF Data Registry reports 6.6% of children with CF have a BMI less than the 10th percentile¹⁸⁸.

Disease Aetiology

The drivers of undernutrition in CF are complex and multifactorial. They include:

- Increased resting energy expenditure which may arise from a combination of:
 - lung infection and inflammation (through oxidant injury and inflammatory mediators)^{224,227}
 - increased work of breathing and coughing²⁷⁹
 - acute respiratory exacerbations which cause transient periods of increased resting energy expenditure (REE) in many patients^{223,224,228}
- Decreased protein synthesis²⁸⁰
 - associated with a systemic inflammatory response, more severe pulmonary disease and nutritional deficits^{223,224,228}
- Reduced absorption of nutrients:
 - pancreatic insufficiency results in maldigestion and subsequent malabsorption of macronutrients⁷⁸, which is partially (although not completely) overcome with pancreatic enzyme replacement therapy (PERT)

- inadequate dosing or timing of PERT results in missed opportunities for optimisation of fat absorption²⁰²
- other conditions which have the potential to compound malabsorption are: CF-related liver disease, DIOS, short gut syndrome, intestinal inflammation, low bicarbonate output, delayed gastric emptying rate and small intestinal bacterial overgrowth²⁰²
- Increased energy losses and metabolic alterations:
 - vomiting precipitated by coughing¹
 - CF-related diabetes (particularly that which is poorly controlled), impaired insulin secretion and insulin resistance results in hyperglycaemia and glycosuria⁷⁸
- Inadequate dietary intake:
 - anorexia resulting from illness or symptoms such as bloating, nausea or GOR can impede the individual's ability to consume adequate amounts¹
 - inflammation may contribute to decreased appetite due to the release of cytokines by immunocompetent cells, particularly during pulmonary exacerbations where inflammation increases and appetite is suppressed. (This can improve following antibiotic treatment)^{281,282}
 - altered body image and dysmorphia may limit the desire to maintain a sufficient nutritional intake²⁸³
 - psycho-social issues, low mood, treatment burden and a lack of general well-being may mean the effort required to consume a nutritionally adequate diet is more difficult for some⁷⁸
 - children and infants may experience feeding problems, meal time behaviours or interactions with parents that challenge the achievement of adequate nutritional intake^{4,5,8-10}

It is also known that lung transplantation (Chapter 16) and pregnancy (Chapter 14) in CF place further stress on diet and nutritional status^{1,190}.

Assessment of undernutrition

As the differential diagnosis for undernutrition in CF is broad, it is important that the wider CF treating team (nurse, respiratory physician, gastroenterologist, endocrinologist, psychologist, social worker, physiotherapist) assist in exploring the multi-faceted nature of undernutrition²⁰². The specific nutrition expertise of the CF dietitian places them in the ideal position to lead a coordinated assessment approach.

DIET

In people with undernutrition, pay particular attention to:

- total energy intake:
 - energy, protein, fat and caloric density of foods
 - use of supplementary nutritional support
 - fat intake at each meal compared to dosing of PERT
- frequency of meals and snacks
- food and nutrition related knowledge and beliefs
- mealtime behaviors and environment
- food security and preparation skills
- energy demands of illness and exercise
- adherence to and impact of previous nutrition intervention

CLINICAL

GENERAL CONSIDERATIONS

- review an individual's lung function status and trends
- monitor pulmonary symptoms and frequency of respiratory exacerbations
- consider screening for the presence of, and review control of, comorbidities that may contribute to undernutrition:
 - **gastrointestinal system** (Chapter 11). Inadequately treated pancreatic insufficiency, constipation, motility issues (including delayed gastric emptying), small bowel bacterial overgrowth, GOR, irritable bowel syndrome and fermentable carbohydrate malabsorption, CF-related liver disease and coeliac disease



- **endocrine system** (Chapter 12). CF-related diabetes and lesser degrees of altered glucose metabolism, hypothyroidism, growth hormone deficiency, menstrual cycles and gonadotrophin deficiency secondary to low body weight. In females luteinising hormone (LH) and follicle-stimulating hormone (FSH) and testosterone blood levels
- consider the impact of anxiety, depression, stress, eating disorders, body dysmorphia or other behaviours on a person's ability to achieve adequate nutrition
- check available support systems (family, partner, friends, community links)

ANTHROPOMETRY

Routine anthropometric parameters include:

- height (length in infants <2 years),
- weight, and
- BMI.

It is also important to consider genetic growth potential in children and adults using the mid-parental height technique (Chapter 5).

BODY COMPOSITION

Body composition assessment may augment anthropometric data to identify the presence of depleted fat free mass (FFM) stores, fat stores or both ^{78,213,225} (Chapter 5).

PHYSICAL ASSESSMENT

Visually examine muscle and subcutaneous fat stores for signs of depletion (Chapter 5). While signs of oedema are not specific to undernutrition, they are useful to note as oedema may mask the true extent of weight loss. ²⁶⁷

BIOCHEMICAL AND LABORATORY DATA

There are no specific biochemical tests that indicate or validate the presence of undernutrition. General laboratory tests (e.g. routine measuring of urea, electrolytes and fat soluble vitamin levels, liver function tests and inflammatory markers) may contribute to characterising undernutrition and identifying contributing co-morbidities ⁷⁸.

Nutrition Diagnosis

MEDICAL DIAGNOSTIC CRITERIA AND DOCUMENTATION OF MALNUTRITION

Malnutrition is a more formal classification of undernutrition encompassing specific clinical, anthropometric and physical criteria in children and adults. There is considerable work currently in progress at an international level in the general clinical nutrition field to define malnutrition and related nutrition disorders including cachexia and sarcopaenia ^{261,267}.

An international working party is currently developing an international standard for defining malnutrition that will likely inform the next round of disease classifications (i.e. ICD-11). These definitions are important because they drive coding and thus influence hospital reimbursement. Some individuals with CF with undernutrition requiring intervention are likely to meet the generic clinical and coding criteria for the diagnosis of malnutrition.

Until the release of ICD-11, malnutrition status should continue to be documented for clinical coding where this is part of institutional requirements.

- adult malnutrition coding criteria: BMI <18.5 kg/m² and/or unintentional weight loss > 5% with evidence of suboptimal intake resulting in loss of subcutaneous fat and/or muscle wasting ²⁸⁴
- paediatric malnutrition coding criteria ²⁸⁴
 - Mild malnutrition BMI z-score -1 to -2
 - Moderate malnutrition BMI z-score -2 to -3
 - Severe malnutrition BMI z-score ≤-3

Intervention

It is important to realistically assess an individual's acceptance and commitment to any potential nutritional interventions, including review of the impact of previous nutrition interventions.

ROUTINE NUTRITION EDUCATION AND PREVENTIVE COUNSELLING

As energy requirements will vary throughout life stages and with disease progression, it is likely that a high protein, energy and fat diet may be required to assist in maintaining nutritional status in people with CF ²⁸⁵. Nutritional education to promote optimal weight status should be provided throughout the lifespan of those with CF. In order to achieve elevated nutrition requirements in the context of the daily diet, high protein and energy diet strategies are common practice in the nutritional management of CF ²⁰².

This may include ²⁴⁹:

- the inclusion of high protein foods at each meal
- encouraging the consumption of energy dense foods, particularly those that are also nutrient dense
- the fortification of foods with additional sources energy
- encouraging frequent meals or the addition snacks in between meals, and
- the consumption of drinks naturally high in protein or energy to supplement intake outside of meal times

During periods of illness, progressing CF lung disease or malnutrition, dietary intervention strategies may need to be intensified and increased. This may require additional support, re-enforcement of strategies and perhaps the progression to commercial nutritional supplements ²⁴⁹. Similarly, during exacerbations requiring inpatient hospital admission, the CF dietitian should ensure the provision of a nutritionally sufficient hospital diet. This may require higher protein or energy items to be offered to CF patients above and beyond the usual hospital menu items and depending on nutritional risk, may progress on to commercial oral or enteral supplement provision.

BEHAVIOURAL MODIFICATION STRATEGIES

The literature regarding behavioural modification strategies around food and mealtimes for people with CF is currently limited to children.

Compared to standard nutritional care, do behavioural interventions around food and mealtimes improve behaviours, diet variety, and weight or nutrition status in children with CF? PICO 6.1.1

[Grade B] Offer behavioural modification strategies to children at risk of/or with identified undernutrition. Conduct behavioural modification strategies in combination with nutrition education. ⁴⁻⁸

Behaviour modification strategies are a valuable component of standard paediatric CF care, especially for children with or at risk of undernutrition ⁴⁻⁸. The following behaviour modification strategies have been found to be effective:

- differential attention (praise and ignoring),
- contingency management (child only receives a desired reward after they have eaten their meal and/or performed desired mealtime behaviours), and
- self-monitoring of food intake (parents and/or child) and parental limit setting (establishing clear expectations and consequences).

It has been shown that the best results are achieved when behavioural strategies are conducted in combination with nutrition education ^{5,8,10}.

When should behavioural interventions around food and mealtimes be considered for children with CF?

PICO 6.1.2

[Grade C] Commence behavioural modification strategies early in life (i.e. during infancy or toddlerhood) and potentially continue throughout childhood. Offer the following strategies:

- Differential attention (praise and ignoring)
- Contingency management (child only receives a desired reward after they have eaten their meal and/or performed desired mealtime behaviours)
- Self-monitoring of food intake (parents and/or child)
- Parental limit setting (establishing clear expectations and consequences) ^{7,9,10}

Research indicates that behaviour modification strategies are to be considered at a young age, before disruptive eating and mealtime behaviours become an ongoing issue. There is also evidence to support the ongoing use of these strategies throughout childhood ^{7,9,10}.



The Behavioral Pediatrics Feeding Assessment Scale (BPFAS) is a validated questionnaire for children aged 7 months to 7 years that has been used to assess the frequency of child and parent behaviours during mealtimes ²⁸⁶. This tool has been used successfully with the CF population ²⁸⁷.

Easy to implement behavioural management strategies include ⁴⁻⁸:

- normalise food and eating by making mealtimes enjoyable experiences;
- encourage a structured approach to meal and snack times;
- set rules and clear expectations for desired behaviours (e.g. limit meal times to 15 minutes for toddlers, 20 minutes for young children);
- set energy goals for meals and snacks;
- encourage new foods;
- praise child for eating, and complement appropriate eating behaviours (e.g. verbal “nice job with eating everything on your plate, non-verbal high five, hug);
- ignore child complaints or disruptive behaviors incompatible with eating (e.g. turn away from child, leave the room, silence, get up from the table);
- remove distractions at mealtimes (e.g. television, computers, electronic tablets, toys);
- provide extra fluids only after a meal is eaten; and
- provide regular reward incentives for meeting caloric goals (e.g. star charts, trophies)

Many paediatric hospitals have practice guides and education resources to help clinicians further explore the use behavioural modification strategies in their practice.

APPETITE STIMULANTS AND GROWTH HORMONE

The use of appetite stimulants to improve nutritional status in CF is limited due to concern over adverse side effects and insufficient evidence to support routine use in patients with CF.

Megesterol acetate was first used to treat breast cancer and is chemically similar to progesterone ²⁸⁸. One of its side effects is stimulation of appetite. The exact mechanism as to how megesterol acetate stimulates appetite is not known ²⁸⁹. It is proposed that it may have an effect on the inflammatory process (cytokines) whereby lowering cytokine levels have been shown to have an effect on the treatment of anorexia and cachexia in patients with cancer ^{290,291}.

Cyproheptaine (Periactin®) is an anti-histamine that has a secondary effect of stimulating appetite. Its mechanism for stimulating appetite is not known however it is thought to interfere with serotonin levels ^{292,293}.

Do appetite stimulants, megesterol acetate and cyproheptadine, improve nutritional status in CF? PICO 6.1.3

[Grade C] There is some evidence to suggest that appetite stimulants may improve weight and appetite for people with CF. However, the potential risk of adverse side effects and insufficient evidence means that routine use of appetite stimulants to improve nutritional status is not recommended. ¹¹

The 2014 Cochrane systematic review regarding the use of appetite stimulants in CF included three small randomised and quasi-randomised controlled trials (n=47 patients total) that look specifically at the use of megesterol acetate and cyproheptadine hydrochloride in CF ¹¹. The results of the meta-analyses found that ¹¹:

- weight z-score significantly improved across all trials after 3 months of use
- weight significantly improved after 6 months with megesterol acetate use in one study
- no significant impact on pulmonary outcomes (FEV₁ percent predicted)
- a statistically significant increase in the proportion of patients with an increased appetite.

Despite the potential for appetite stimulants to improve weight and appetite in the short term (6 months), there is inadequate evidence to support their use in CF at this point in time ¹¹. There is also concern regarding their safety as potential adverse side effects of appetite stimulants use are not well documented ¹¹. Potential side effects of appetite stimulants include ¹¹:

- impaired blood glucose control
- fatigue
- mood
- fluid retention
- shortness of breath
- elevated liver transaminases.

Growth hormone is an anabolic agent that promotes the synthesis of protein, optimises fat utilisation and reduces the oxidation of glucose.

Does the use of recombinant growth hormone improve nutritional status in pre-pubertal people with CF?

PICO 6.1.4

[Grade C] There is some evidence to suggest that growth hormone may improve height, weight and lean tissue mass for pre-pubertal people with CF. Routine use of growth hormone to improve nutritional status in people with CF is not recommended.¹²

The 2015 Cochrane systematic review regarding the use of recombinant growth hormone therapy in CF includes 4 randomised and quasi-randomised controlled trials (n=161 patients total, study durations 6-12 months). The review specifically focuses on the use of growth hormone therapy on lung function, quality of life and clinical outcomes in children and young adults¹². The results of the meta-analyses found that growth hormone use¹²:

- Modestly improves height, weight and lean tissue mass;
- Improvement in lean tissue mass;
- Has no consistent impact on lung function, muscle strength, clinical condition and/or quality of life; and
- Has no effect on glucose metabolism and doesn't increase the chance of developing CF-related diabetes.

ORAL NUTRITIONAL SUPPLEMENTS

Is there any rationale for the use of commercial oral nutritional supplements in addition to food and mealtime strategies to improve nutritional intake, weight or pulmonary function in CF?

PICO 6.1.5

[Grade B] Consider the use of oral nutrition supplements on an individual basis. There is no clear evidence that their routine use in addition to food and behavioural modification strategies will result in improvements to nutritional intake, weight or pulmonary function in CF.¹³⁻¹⁹

Oral nutrition supplements (ONS) are unlikely to result in improvement in outcomes such as BMI, nutritional intake or pulmonary function in adults and children with CF over and above the use of routine dietary advice and monitoring alone^{13,14,16-19,249}.

ONS may replace some of the energy taken as food and their potential effect on overall total energy intake may be either reduced or eliminated. This does not mean that these products may not be efficacious in some individuals, but that clinicians should balance potential benefits against potential adverse effects¹⁴. ONS should not be regarded as essential in the long term care of all individuals with CF who are malnourished^{13,14,249}. Further randomised controlled trials are needed to establish the role of short-term ONS in people with CF and acute weight loss and also for the long-term nutritional management of individuals advanced lung disease²⁴⁹.

If it is thought that ONS may aid in achieving nutritional adequacy, the choice of supplement should be assessed on an individual basis dependent on the patient's nutritional requirements, age and taste preferences. At the commencement of ONS, clear and measurable outcomes should be set to assist in evaluating effectiveness. This can guide the need for continuation, reduction or cessation of ONS depending on whether outcomes are met. ONS should complement the normal food intake and are best taken after a meal or in between meals and snacks so as not to replace the appetite for normal food⁷⁸.

There is a wide variety of ONS available on the market. They commonly come in the form of milk or fruit flavoured drinks, custards, puddings, protein bars and macronutrient powders or liquids. They contain energy ranging from 4.2-8.4kJ/ml (1-2cal/ml), additional protein from whey or plant sources and are sometimes fortified with vitamins and minerals.

ENTERAL FEEDING

Enteral tube feeding may be considered in undernourished patients or those with declining nutritional status where oral nutrition support interventions have proven insufficient to meet nutritional goals. However, it should be noted that the quality of the evidence base for the use of enteral feeding in CF is poor; hence outcomes may vary on an individual basis in the clinical setting. The decision to place an enteral feeding tube should be made on a case-by-case basis.



Should enteral feeding be considered to improve nutrition outcomes for people with CF? PICO 6.1.6

[Grade B] Consider enteral feeding as a means of improving markers of weight, BMI and BMI z- score in adults and children with CF who have been assessed as being undernourished. ²⁰⁻²⁹

Should enteral feeding be considered to improve pulmonary status in people with CF? PICO 6.1.7

[Grade C] Practitioners should refrain from commencing supplementary enteral feeding for the sole purpose of improving or stabilizing pulmonary outcomes. ^{20-26,28}

Practitioners should expect the most favourable nutritional gains to take place in the first six to 12 months of enteral tube feeding ²⁰⁻²⁹. However, poor compliance with feeding regimens, CF-related diabetes, and young age at tube insertion may not predict success of nutritional rehabilitation with enteral feeding ²¹. In regards to the effect of enteral tube feeding on pulmonary outcomes, there is weak and inconsistent evidence that enteral feeding helps to improve pulmonary function or stability ^{20-26,28}.

When should enteral feeding be introduced for people with CF? PICO 6.1.8

[Ungraded] There is insufficient evidence to make a recommendation regarding when to introduce enteral nutrition in CF. Evaluate appropriate timing on an individual basis.

Despite the lack of conclusive evidence, it is suggested that gastrostomy feeding should be used earlier to optimise growth in CF children. In adults, enteral feeding is likely to be more successful if initiated before advanced lung disease is irreversibly established ^{25,27}. Aggressive nutrition support by way of tube feeding should be planned ²¹. Practitioners should aim to optimise pulmonary status prior to gastrostomy insertion ²⁹⁴.

What is the ideal enteral feeding regimen for people with CF? PICO 6.1.9

[Ungraded] There is insufficient literature to suggest the ideal enteral formula or regimen in the CF population. Select enteral formulas and devise enteral feeding regimens on an individual basis.

What are the risks associated with enteral feeding in CF compared to the general population? PICO 6.1.10

[Grade C] People with CF are not at increased risk of major complications and mortality as a result of enteral feeding. Minor side effects of enteral feeds, including stoma site issues and GOR should be managed as for the general population.

Enteral feeding in CF is safe, including in people with low lung function and in those prone to pulmonary exacerbation. No major complications or mortality have been reported in the literature. Studies describe a range of minor complications associated with enteral feeding in CF, such as stoma site issues and GOR. Minor complications may not warrant the cessation of feeding, instead requiring change in feeding management or a non-invasive treatment ^{20-27,29}. Documented risks associated with enteral feeding in CF include:

- Gastrostomy site, itchiness, redness and infection ²⁴:
 - Regular stoma monitoring and involvement of a gastrostomy credentialed dietitian or stoma specialist nurse may help in preventative treatments for local site irritation ¹³²;
- Increasing symptoms of GOR:
 - May result in less favourable nutritional outcomes if not symptomatically controlled ²⁷. Positioning at a 30-45% angle during feeding ²⁹⁵. A slower feed rate and post pyloric feeding may be of assistance in GOR ¹.
- Bloating or nausea during enteral feeds:
 - May benefit from the use of prokinetic or antiemetic agents pre feed ¹³²
 - Review PERT dosing strategies as outlined in [chapter 10](#).
- Increased incidence of hyperglycaemia or CF-related diabetes ²²:

- Review CF-related diabetes management prior to tube insertion ²⁹⁴.
- Pre, mid-way and post feeding blood glucose monitoring is indicated when enteral tube feeding is commenced ²⁹.
- A small dose of insulin pre feeding may be required in some cases ⁷⁸.

In 2016, the Cystic Fibrosis Foundation (CFF) released evidence-informed guidelines for enteral tube feeding individuals with CF. These guidelines are based on a thorough review and critique of 241 papers addressing a number of PICO questions specifically relating to enteral tube feeding in CF ²⁹⁴. A total of 33 recommendations, all with 100% agreement, were established by an interdisciplinary working group ²⁹⁴. This may be an additional useful reference for clinicians exploring enteral feeding. These American guidelines are compatible with the recommendations within this chapter.

Considerations for nutrition support funding:

- Facilities around Australian and NZ have varying centre-based protocols for the provision and funding of oral and enteral supplements and equipment to people with CF.
- Many Australian states are able to provide patients with hospital funded enteral and/or oral formula and supplies. However, in some states, and the private sector, people with CF may be required to fund their own products.
- In NZ all enteral feeds and equipment are supplied at no cost to the patient and ONS are either fully or partly subsidized by PHARMAC^{1*}.

PARENTERAL NUTRITION

Parenteral nutrition is not recommended for routine use or long-term treatment in CF due to the requirement for centrally placed catheters, the risk of complications (including line sepsis and new onset diabetes) and the challenges associated with administering parenteral nutrition outside the hospital setting. However, parenteral nutrition may be useful for short term support during bowel obstruction, meconium ileus, major gastrointestinal surgery and in the severely ill person with CF awaiting lung or liver transplantation ²⁷⁹. The initiation, management and monitoring of parenteral nutrition should be guided by the specialist interdisciplinary team, hospital protocols and evidence-based parenteral nutrition guidelines.

Monitoring & Evaluation

People with CF who are taking ONS or enteral feeding should be reviewed regularly to assess tolerance, adherence, progress towards objectives and ongoing need ¹. It is important to monitor for disordered eating, oral intake aversion and other behavioural concerns in adults and children ²⁹⁴.

Enteral feeding could be withdrawn at any time deemed necessary or appropriate by the individual with CF or the treating team. If goals of feeding are met, the individual is nutritionally stable and the gastrostomy is not likely to be required again in the near future consideration could be given to its removal. Similarly, if the tube is inhibiting quality of life or psychosocial health and nutritional goals are not being met, removal of feeding tubes should be considered.

Practice Points ^{PICO 6.1.1 & 6.1.2}

Behavioural modification strategies are a valuable component of standard paediatric CF care

Strategies should be considered at a young age, before disruptive eating and mealtime behaviours become an ongoing issue.

For best results, strategies should be conducted with nutrition education.

¹ *PHARMAC (Pharmaceutical Management Agency) is the NZ Crown agency that decides on which medicines are subsidised for use in the community and public hospitals.



Practice Points PICO 6.1.3 & 6.1.4

The decision to commence an appetite stimulant should be made as an interdisciplinary team and in consultation with the individual with CF and their family or carers, following evaluation of potential benefits and risks in the individual with CF.

The most commonly used appetite stimulants in CF are megestrol acetate and cyproheptadine (peractin).

- They may improve weight and appetite but evidence is inconclusive.
- Some concerns regarding side effects and therefore safety with longer term use.

Growth hormone may improve height, weight and lean tissue mass for pre-pubertal individuals with CF, however, longer term randomized controlled trials are required.

- Until more studies are done looking at the longer term use of growth hormone, it is not recommended for routine use in CF

Prior to commencing a trial of appetite stimulants in CF, issues to explore include:

- Identification of other factors that may be contributing to a poor appetite and subsequently poor weight gain or growth and where possible, treat the underlying cause first.

Practice Points PICO 6.1.5

Where possible, avoid using oral supplements as a meal substitution

- Oral supplements should complement usual intake
- Best taken after a meal or as a snack
- A maximum of three oral supplements daily is often recommended to avoid a reduction in appetite around mealtimes.
- Particularly important for the paediatric population where normalised eating is still developing.

Regularly review oral supplement tolerance, adherence and nutritional status response.

The most commonly used oral supplements in CF are dairy based and usually 1-1.5kcal/ml.

Evaluate individual cost versus benefit because oral supplements can be a financial burden because funding for nutrition support varies across the health systems in Australia and New Zealand.

Practice Points PICO 6.1.6 & 6.1.7

The decision to commence either short or long term enteral nutrition support should be made by an interdisciplinary team and in consultation with the individual and their family, including discussion of risks and benefits.

- Benefits on nutrition outcomes, particularly weight and BMI are well documented
- There is no conclusive evidence to support beneficial effects on pulmonary function

The decision can be emotionally challenging for some people with CF. Where possible, appropriate psychosocial support should be provided and the individual's decision should be respected. An anaesthetist should be consulted prior to surgical or endoscopic gastrostomy tube insertion in people with moderate to severe CF lung disease.

Practice Points PICO 6.1.8

No evidence to support best timing for enteral nutrition support in CF. The following considerations should be noted in regards to timing of enteral nutrition:

- The person is unable to meet nutritional requirements via oral intake alone
- Conduct Interdisciplinary review with investigation of reasons for any decline in nutrition status and interventions commenced as appropriate
- Explore the role for behavioural modification strategies (In the paediatric population)
- Whilst many patients will have had a trial of oral nutritional supplements (ONS) prior to the need for enteral nutrition being assessed, there is no evidence that favours assessing the impact of ONS first, over proceeding to enteral nutrition. Evaluate whether to trial ONS prior to considering enteral nutrition on an individual basis

Enteral nutrition should be commenced prior to the onset of significant disease progression and FEV₁ decline for more favourable nutritional outcomes

Practice Points PICO 6.1.9

Enteral feed regimens should be devised on an individual basis

The following considerations should be assessed in relation to the individual:

- Caloric targets should be calculated by the specialist dietitian.
- Overnight continuous feeds are usually recommended to preserve appetite and oral intake during the day, though supplementary bolus feeds may also be useful for some people.
- Feed composition
- The choice between polymeric, semi-elemental and elemental feeds should be made on an individual basis.
- Many people with CF will tolerate polymeric feeds well and more specialised formulas are not usually required.
- Choose energy dense feeds i.e. 1.5-2kcal/ml where possible
- Evaluate individual cost/financial burden versus benefit because funding for enteral nutrition varies across the health systems in Australia and New Zealand.
- Feed tolerance should be reviewed regularly
- Co-morbidities such as reflux may play a role in feed regimens and enzyme dosing strategies ([Chapter 10](#)).
- Feeding route
- Nasogastric feeding is usually recommended when feeds are required for < 3 months.
- Gastrostomy insertion should be considered when feeds are required for > 3 months.

For supplementary feeding, aim to meet 30-60% of the individual's calculated energy requirements, or to meet a specifically calculated energy deficit in the diet.

Practice Points PICO 6.1.10

Enteral feeds are considered safe for the CF population. However, as with any intervention, potential risk factors should be evaluated and investigated prior to feed commencement. Safety considerations around enteral feeding include, but are not limited to the following:

- Nasogastric tubes
- Insertion may be difficult and uncomfortable for people with nasal polyps
- Tubes may be dislodged with significant coughing and/or vomiting
- Gastrostomy tubes
 - Aim to optimise pulmonary to health prior to placement of gastrostomy tube
 - Plan for postoperative pain management with the goal of initiating airway clearance within 24hrs
 - Gastrostomy site, itchiness, redness and infection are common. Regular stoma monitoring is recommended.
- GOR
 - Positioning: ensure the person is elevated to a 30-45% angle during feeding, reducing feed rate and post pyloric feeding may be of assistance
- Bloating or nausea during enteral feeds may benefit from the use of prokinetic or antiemetic agents prior to feeding.
- Potential risk of hyperglycaemia or CF-related diabetes. Blood glucose monitoring is indicated prior to, mid-way through and at the end of feeding.

Dietetic department protocols should guide the use of gastrostomy tube and care education prior to discharge home with a feeding tube.



6.2 Overweight and Obesity

N. Saxby, T. Crowder, N. Forgiione & S. King

Recent data from around the world show that the proportion of people with CF who are overweight/obese is increasing^{199,278,296}. In one study, based at an American CF centre, it was identified that 23% of children and adolescents with CF had an average BMI percentile greater than 90²⁷⁸. Similar rates of overweight and obesity BMI were identified among a Canadian adult CF population in 2011¹⁹⁹. Overweight/obese individuals (all ages) with CF tended to be older, have better lung functions, have milder genotypes, and were more often male and pancreatic sufficient¹⁹⁹.

Whilst the significance of overweight and obesity in people with CF is unknown, recent research suggests that coronary artery disease, hypertension and obstructive sleep apnoea are emerging areas of concern for this sub-population group²⁷⁸. There also appears to be minimal benefit to lung function when adults with CF have a BMI greater than 25 kg/m² and thus a high BMI needs to be balanced against the known health risks of obesity¹⁹⁹.

Disease Aetiology

Overweight and obesity can be attributed, at least in part, to continuously advancing treatment options for CF (e.g. genetic modulator therapies), longer life expectancies, and increased prevalence of obesity in the general population¹⁹⁰. There is no research, as yet, into metabolic risks of excess adiposity in CF. However, with increased longevity, people with CF may potentially be at risk of some of the metabolic complications seen in the general population.

Assessment

DIET

Attention should be given to:

- Total energy intake and nutrient density of foods; and
- Diet quality to ensure that micronutrient requirements are being met.

CLINICAL

GENERAL CONSIDERATIONS

- Lung function, stage of disease and goals of care
- Assess a person's willingness to make the behavioural changes needed to maintain and/or lose weight (readiness for change)
- Psychological issues associated with weight (especially in younger women)
- Lung transplant - some lung transplant centres accept people with a BMI up to 29 kg/m² while others have a lower cut-off
- The risks of metabolic disease (eg type 2 diabetes, cardiovascular disease) should be considered particularly in those with mild lung disease and who are over the age of 30 years. Diagnosing metabolic disease/s, however, is particularly challenging in CF.

ANTHROPOMETRY AND BODY COMPOSITION CONSIDERATIONS FOR OVERWEIGHT/OBESE INDIVIDUALS

- Measuring abdominal circumference may be useful for identifying excess central adipose tissue in adults and for monitoring the effect of interventions in individuals identified as suitable for weight loss. As yet there is no CF-specific evidence to determine if the general population cut-offs for abdominal obesity apply to stratification of risk for people with CF.
- Other body composition measurement techniques may be useful to assess relative contribution of FFM (especially in males)

Table 6c. Summary of anthropometric criteria to diagnosis of overweight and obesity in CF*

Classification criteria - High BMI - at possible risk related to overweight and obesity	
Infants	n/a
Children & Adolescents	<p>Overweight: BMI 85-95th percentile if using CDC growth chart (Australia)</p> <p>OR BMI 91-98th percentile if using NZ-WHO growth chart</p> <p>Obese: BMI >95th percentile if using CDC growth chart (Australia)</p> <p>OR BMI >98th percentile if using NZ-WHO growth chart</p> <p>High risk of developing overweight or obesity Unintentional weight gain from previously acceptable BMI percentile: gain of more than 2 BMI percentile bands</p>
Adults	<p>BMI >27kg/m²</p> <p>OR Unintentional weight gain from previously acceptable BMI of >5kg within one year</p>

* Refer to table 5a for more information

Intervention

There is a lack of evidence available to substantiate interventions for the management of overweight and obesity in people with CF. Leveraging the recommendations for the general population, when appropriate, can provide some guidance to practitioners. It is important to note however, that general population recommendations are intended for use in people with stable health, milder genotypes and good lung function and thus they may not always be appropriate for the CF population. In adults with severe CF lung disease and a BMI >27kg/m², achieving reduction in fat mass while preserving lean mass is likely to be challenging. In such individuals, who may have limited exercise capacity, intentional or unintentional weight loss is likely to be predominantly lean mass. This may be associated with reductions in strength and function. Advice regarding nutritional status and metabolic risks should take into account body composition and include monitoring for signs of sarcopaenic obesity and potentially increased mortality²⁹⁷.

For people with CF consider the appropriateness of the current clinical practice guidelines for the management of overweight and obesity in children, adolescents and adults in Australia²⁹⁸ and NZ^{299,300}.

Advise people about the potential health benefits of lifestyle change and weight loss²⁹⁸.

- [Grade A general population, CF unknown] Adults who are overweight or obese can be advised that modest weight loss reduces cardiovascular risk factors
- [Grade B general population, CF unknown] Adults with sleep apnoea can be advised that improvements in this condition are associated with a 5% weight loss
- [Grade C general population, CF unknown] Adults with musculoskeletal problems, GOR or urinary incontinence can be advised that weight loss of 5% or more may improve symptoms



Assist people to lose weight through lifestyle modification ²⁹⁸.

- [Grade A general population, CF unknown] Adults who are overweight or obese can be strongly recommended lifestyle change – including reduced energy intake, increased physical activity and measures to support behavioural change
- [Grade B general population, CF unknown] Utilise the expertise of the interdisciplinary team for overweight adults and children, and multicomponent approaches as these work better than single interventions (e.g. allied health)
- [Ungraded general population – consensus, CF unknown] Assist adults children and adolescents to get help for disordered eating, poor body image, depression and anxiety and weight-related bullying where these are present
- [Grade C general population, CF unknown] For children and adolescents, focus lifestyle programs on parents, carers and families
- [Grade B general population, CF unknown] For children and adolescents who are overweight or obese, recommend lifestyle change – including reduced energy intake (e.g. encourage drinking of water) and sedentary behaviour (e.g. reduce screen time), increased physical activity and measures to support behavioural change
- [Grade D general population, CF unknown] For children who are managing overweight or obesity, advise that weight maintenance is an acceptable approach in most situations

Additional intensive interventions such as bariatric surgeries should only be considered on an individual basis in people with CF, as the risks may outweigh the possible benefits and nutritional status greatly compromised.

Monitoring & Evaluation

Regular monitoring of people with CF who have a high BMI is essential. Ideally anthropometric monitoring should take place every three months at routine CF clinic appointments (or more frequently if able). Reviews should include monitoring of eating behaviours, sedentary times, physical activity habits and psychosocial factors. Practitioners may also choose to monitor for additional co-morbidities that may be related high BMI in individuals older than 30 years.

Translating into Practice

- Transition to adult care can sometimes enable a change of focus from the high fat high energy diet to a more nutrient dense, moderate energy diet, particularly if the individual has a BMI >27 kg/m² or BMI percentile in the overweight or obese range.
- Assessment of nutritional status, anthropometry and dietary intake should occur as for those who are overweight or obese as those who are at nutritional risk or undernourished.