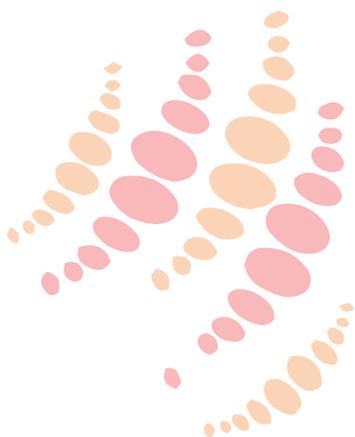




NUTRITION GUIDELINES FOR CYSTIC FIBROSIS

in Australia and New Zealand

August 2017



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The guidelines will not be published as a complete document. Instead, copies of the guideline can be downloaded from the websites of CFA (www.cysticfibrosis.org.au), CFNZ (www.cfnz.org.nz) and the TSANZ (www.thoracic.org.au).

Disclaimers

The primary custodian of the 'Nutrition Guidelines for Cystic Fibrosis in Australia and New Zealand Project' is the TSANZ. The following professional organisations were collaborators in the creation of this document:

- Dietitians Association of Australia (DAA)
- Dietitians New Zealand (DNZ)
- Cystic Fibrosis Australia (CFA)
- Cystic Fibrosis Association of New Zealand (CFANZ)

This document is written as a general guide to practice only and does not discount individualised assessment and advice, by a suitably qualified clinician. Content within this publication was accurate at the time of publication.

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PUBLICATION APPROVAL



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NHMRC is satisfied that the guideline recommendations are systematically derived, based on the identification and synthesis of the best available scientific evidence, and developed for health professionals practising in an Australian health care setting.

This publication reflects the views of the authors and not necessarily the views of the Australian Government.

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Additional information about the roles of the dietitian steering group and interdisciplinary clinical expert committee, including methods for addressing conflicts of interests and declarations of conflicts of interest, can be found in the accompanying [Administration Report](#).

IN MEMORIAM

We are honoured to dedicate this project to the memory of Anne-Maree Bosch. Every project has people on the sidelines – those people who cheer us on, and those who open doors to make things happen. For a generation of people working in CF, Anne-Maree was a wonderfully generous colleague and friend. Her enthusiasm, willingness to go above and beyond, and commitment, capture the spirit of this project.

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FOREWORD

Cystic Fibrosis Australia (CFA) whole-heartedly supports the development of nutrition guidelines for cystic fibrosis (CF) in Australia and New Zealand. In recent years nutrition has come to the forefront as a successful health strategy to improve the lives of people with cystic fibrosis. CFA's data registry shows the value of nutritional guidance and advice and therefore CFA is keen to see best practice 'rolled out' across Australia and New Zealand.

Australia and NZ are wide and diverse countries, and people with CF should have access to the best health professions and strategies where ever they live. The Nutrition Guidelines for Cystic Fibrosis in Australia and New Zealand will ensure the provision of quality, up to date information about nutrition management and importantly if people move to another CF Centre or state they will have consistency of care and guidance.

Cystic Fibrosis Nutrition - Australia New Zealand are a passionate group of dietitians, with representatives from all CF specialist centres and outreach services across Australia and New Zealand - they constantly impress care teams and the CF community with their innovative approach to CF nutrition. Cystic Fibrosis Australia thanks these dedicated dietitians for their commitment to best practice and we look forward to supporting their efforts to get exceptional national consistency in dietetics across Australia and New Zealand.

Nettie Burke
Chief Executive Officer
Cystic Fibrosis Australia



ABBREVIATIONS AND ACRONYMS

ALA	= alpha-linoleic acid
BMI	= body mass index
CCRS	= Clinical Care and Resources Subcommittee (TSANZ)
CDC	= Centers for Disease Control
CF	= cystic fibrosis
CFA	= Cystic Fibrosis Australia
CFNZ	= Cystic Fibrosis New Zealand
CRP	= C-reactive protein
DAA	= Dietitians Association of Australia
DHA	= docosahexaenoic acid
DIOS	= distal intestinal obstruction syndrome
DNZ	= Dietitians New Zealand
DXA	= dual x-ray absorptiometry
EAR	= estimated Average Requirement
FEV ₁	= forced expiratory volume in 1 second
FFM	= fat free mass
GOR	= gastro-oesophageal reflux
HbA1c	= glycated haemoglobin
HDL	= high-density lipoprotein
IV	= intravenous
LDL	= low-density lipoprotein
MUFA	= monounsaturated
NODAT	= new onset diabetes after transplant
NHMRC	= National Health and Medical Research Council
NZ	= New Zealand
ONS	= oral nutrition supplement
PERT	= pancreatic enzyme replacement therapy
PUFA	= polyunsaturated fatty acid
RDI	= recommended dietary intake
RDI	= recommended dietary intake
REE	= resting energy expenditure
SFA	= saturated fatty acid
TFA	= trans-fatty acid
TSANZ	= Thoracic Society of Australia and New Zealand
UK	= United Kingdom
UL	= upper Level
USA	= United States of America
WHO	= World Health Organization

EXECUTIVE SUMMARY

Optimising growth and nutrition in people with cystic fibrosis (CF) has been shown to positively influence lung function and survival. The '2017 *Nutrition Guidelines for Cystic Fibrosis in Australia and New Zealand*' (herein referred to as '2017 Guidelines') is a planned update of previously published guidelines - The '2006 *Australasian Clinical Practice Guidelines for Nutrition in Cystic Fibrosis*'. This edition acknowledges and incorporates new topic areas including the nutritional implications of new genetic modulatory therapies, the emergence of overweight and obesity in CF, and complementary nutritional therapies. An overview of dietitian prescribing in New Zealand (NZ) is also provided. This document was developed by a group of Australian and NZ dietitians experienced in CF care, with the assistance of an interdisciplinary clinical expert committee representing various other health professions and CF consumers. In total, 70 people from a total of 24 different CF centres in the 2 countries were involved in this project, see figure 1 below.

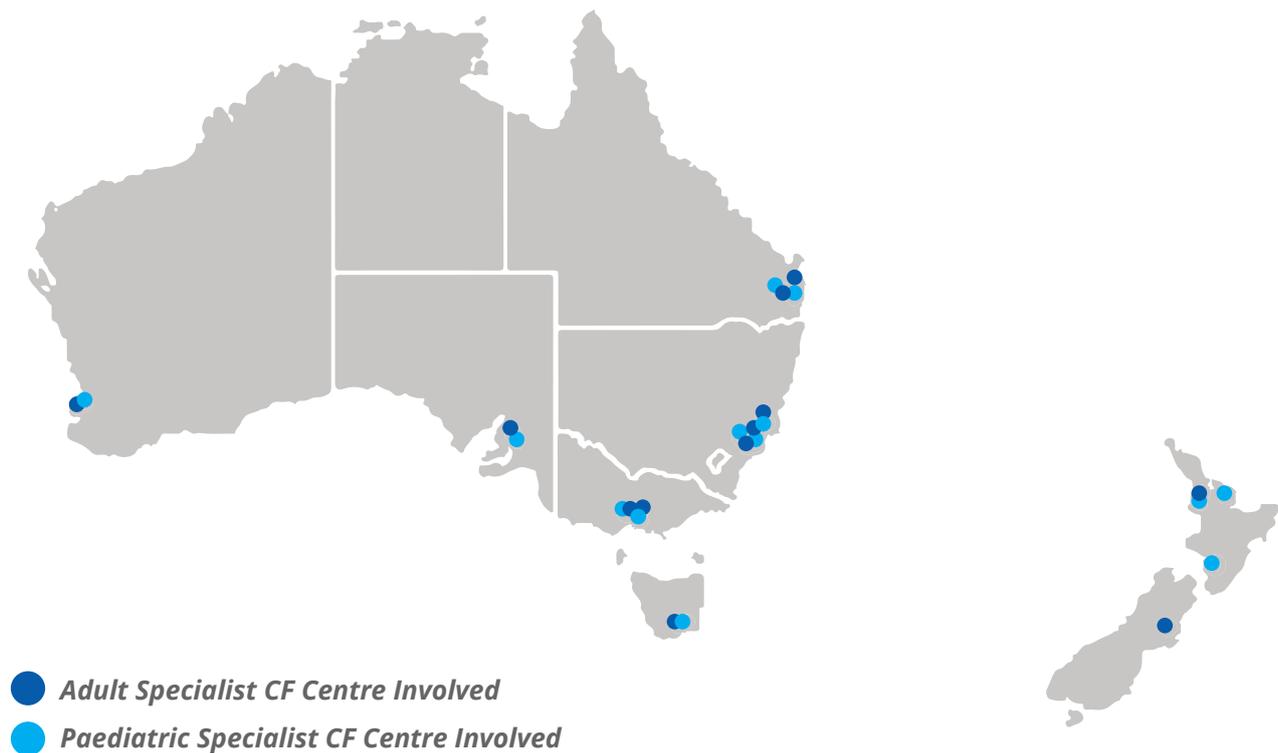


Figure 1. Specialist CF centres involved in the development of the guidelines

The key clinical question addressed is: ***“What is the role and scope of nutritional care in the management of CF?”***

Where possible, we have aimed to address specific clinical practice questions to help guide nutrition care. Details of the systematic review process used in the development of these guidelines are given in the technical report that accompanies this document.

While reading this guideline, it is important to remember the importance that people with CF and their families/carers play in attaining and maintaining optimal nutrition status. These are the people who live with the condition day-to-day and who are responsible for implementing nutrition interventions in the home environment. Individual needs and preferences need to be considered at all times.



MAJOR GUIDELINE THEMES

Below is a summary of the overarching recommendations made in the '2017 *Guidelines*'.

ASSESSMENT

- Assessment of anthropometric parameters should be conducted in children and adults at every clinic ([Chapter 5](#)).
 - It is becoming increasingly common to see people with CF who are overweight. Practitioners should screen for both undernutrition and overweight and obesity.
- Annual comprehensive nutrition assessments are strongly encouraged ([Chapter 5](#)).

INTERVENTION

- Newborn screening is standard practice in Australia and NZ; promotion of good nutrition should be commenced at diagnosis ([Chapter 4](#)).
- Interdisciplinary nutrition management is essential for people with CF ([Chapter 4](#)).
 - Liaise with a gastroenterologist, ideally with CF experience, for the management of common gastrointestinal co-morbidities ([Chapter 11](#)) e.g. distal intestinal obstruction syndrome (DIOS), constipation, gastro-oesophageal reflux (GOR) and liver disease.
 - Liaise with an endocrinologist, ideally with CF experience, for the management of CF-related diabetes ([Chapter 12](#)) and bone disease ([Chapter 13](#)).
- Diet recommendations for people with CF are moving closer to general population guidelines.
 - Aim to achieve and maintain optimal weight status and encourage people with CF to be physically active and choose amounts of nutritious foods and drinks to meet individual energy needs
 - There is a wide inter-individual range of energy requirements about 1.1 to 2 times reference intakes of the general population ([Section 7.1](#)).
 - A high energy/high fat diet will be especially beneficial for those people who are undernourished ([Section 6.1](#)).
 - Promote enjoyment of a wide variety of nutritious foods from all five food groups every day.
 - Encourage intake of foods containing unsaturated fats and omega-3 fatty acids.
 - Encourage, support and promote breastfeeding of infants diagnosed with CF. ([Section 3.1](#)).
- A high salt diet is recommended for most people with CF ([Chapter 9](#)).
- Routine supplementation of fat soluble vitamins (A, D, E, K) is encouraged, particularly in those who are pancreatic insufficient ([Chapter 8](#)).
- Pancreatic Enzyme Replacement Therapy (PERT) is recommended for all people who are pancreatic insufficient and should be taken with fat-containing foods ([Chapter 10](#)).
- Behavioural modification strategies (children) and nutrition education (children and adults) are evidence-based components of standard CF care ([Chapter 6](#)).
- Consider the use of oral supplements on an individual basis, the evidence does not support their routine use ([Chapter 6](#)).
- For people who are underweight long-term, enteral feeding should be considered by the interdisciplinary team. ([Chapter 6](#)).
- There is insufficient evidence to recommend routine supplementation of any complementary nutritional therapies (including probiotics, garlic, ginseng, curcumin, coconut oil) ([Chapter 15](#)).

MONITORING AND EVALUATION

Conduct regular and lifelong nutrition surveillance, with all aspects of nutrition and gastrointestinal status being reviewed ([Chapter 5](#)).

RECOMMENDATION TABLES

KEY: Q = question, R = recommendation (evidence based), PP = practice point (consensus)

Chapter 4 Service Delivery	
Q 4.1	What is the level of dietetic service required for people with CF?
R 4.1	Insufficient evidence to make a recommendation
PP 4.1	Dietetic staffing levels should follow the most recent country specific Standards for CF Care ^{2,3} .

Chapter 5 Nutrition Assessment
No PICO questions were formulated for Chapter 5 . This is an area of limited evidence specific to CF, with most international recommendations being formed by expert opinion.
Key Points
<ul style="list-style-type: none"> • Assessment of anthropometric parameters should be conducted in children and adults at every clinic – practitioners should screen for both under and over nutrition. • Nutrition review by a dietitian is recommended at least four times a year as per the Australia and New Zealand Standard of care documents^{2,3}. Deterioration in nutrition parameters should be detected early, before growth and pulmonary function are compromised. • Annual comprehensive nutrition assessments are strongly encouraged. These should encompass a collation of anthropometric, dietary, biochemical and relevant clinical data. • Levels of fat soluble vitamins (with associated tests to aid interpretation) should be routinely tested together as a group to aid interpretations of abnormal findings • The following criteria are suggestive of optimal weight status: <ul style="list-style-type: none"> ○ Infants (0 to 24 months): weight-for-length $\geq 50^{\text{th}}$ percentile using WHO growth charts ○ Children and Adolescents (2-18 years): BMI 50-85th percentile (if using CDC growth charts) or 50-91st percentile (if using WHO growth charts) ○ Adults: males BMI 23 - 27 kg/m², females BMI 22 - 27 kg/m² • The transition from WHO to CDC growth charts at 2 years can be difficult to interpret as weight and height percentiles do not correspond precisely (see Appendix B). • Body composition measurements can further aid nutrition status evaluation. Practitioners should consider using body composition methods in people who are underweight, overweight or in those with unexplained weight changes. • It is important to raise nutrition concerns with the interdisciplinary team early. • Telehealth models of care are emerging and may augment face to face interdisciplinary reviews when more frequent dietetic monitoring and/or education is required. • Where there is a shared care model, the responsibility for integrating nutrition assessment data and identifying nutrition diagnoses/nutrition problems must be clearly identified to ensure that early signs of undernutrition are detected. • Food diary apps, allowing for electronic documentation of intake, are becoming more readily available. The Australia based app, <i>Easy Diet Diary</i>[®] allows people to manually enter intake or scan barcodes of packaged foods. When shared with the CF dietitian, intake can be quantified using computerised food composition software.



Chapter 6 Nutrition Interventions	
Behaviour Modification Strategies & Nutrition Education	
Q 6.1.1	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?
R 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
Q 6.1.2	When should behavioural interventions around food and mealtimes be considered for children with CF?
R 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: <ul style="list-style-type: none"> • 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}
PP 6.1.1 and 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.
Appetite Stimulants & Growth Hormone	
Q 6.1.3	Do appetite stimulants, megestrol acetate and cyproheptadine, improve nutritional status in CF?
R 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
Q 6.1.4	Does the use of recombinant growth hormone improve nutritional status in pre-pubertal people with CF?
R 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
PP 6.1.3 and 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). <ul style="list-style-type: none"> • 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. <ul style="list-style-type: none"> • 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: <ul style="list-style-type: none"> • 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 • Potential side effects of each appetite stimulant. • The role of other factors which may impact oral intake including (but not limited to) reflux, intestinal dysmotility and early satiety.

Oral Nutritional Supplements	
Q 6.1.5	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?
R 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
PP 6.1.5	<p>Where possible, avoid using oral supplements as a meal substitution</p> <ul style="list-style-type: none"> • 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. <p>Regularly review oral supplement tolerance, adherence and nutritional status response.</p> <p>The most commonly used oral supplements in CF are dairy based and usually 1-1.5kcal/ml.</p> <p>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.</p>
Enteral Feeding	
Q 6.1.6	Should enteral feeding be considered to improve nutrition outcomes for people with CF?
R 6.1.6	GRADE B. Consider enteral feeding as a means of improving markers of nutritional status (including weight, BMI and BMI z-score) in children and adults with CF who have been assessed as being undernourished. ²⁰⁻²⁹
Q 6.1.7	Should enteral feeding be considered to improve pulmonary status in people with CF?
R 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}
PP 6.1.6 and 6.1.7	<p>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.</p> <ul style="list-style-type: none"> • Benefits on nutrition outcomes, particularly weight and BMI are well documented • There is no conclusive evidence to support beneficial effects on pulmonary function <p>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.</p>
Q 6.1.8	When should enteral feeding be introduced for people with CF?
R 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.
PP 6.1.8	<p>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:</p> <ul style="list-style-type: none"> • 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



Q 6.1.9	What is the ideal enteral feeding regimen for people with CF?
R 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.
PP 6.1.9	<p>Enteral feed regimens should be devised on an individual basis</p> <p>The following considerations should be assessed in relation to the individual:</p> <ul style="list-style-type: none"> • 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. <p>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.</p>
Q 6.1.10	What are the risks associated with enteral feeding in CF compared to the general population?
R 6.1.10	GRADE C. There is no evidence that people with CF are at increased risk of major complications and mortality as a result of enteral feeding. Manage minor side effects of enteral feeds, including stoma site issues and GOR, as for the general population. ^{20-27,29}
PP 6.1.10	<p>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.</p> <p>Safety considerations around enteral feeding include, but are not limited to the following:</p> <ul style="list-style-type: none"> • 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 <ul style="list-style-type: none"> ○ 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 <ul style="list-style-type: none"> ○ 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. <p>Dietetic department protocols should guide the use of gastrostomy tube and care education prior to discharge home with a feeding tube.</p>

Chapter 7 Macronutrients	
Energy	
Q 7.1.1	Are energy requirements increased in the CF population compared to the general population?
R 7.1.1	GRADE D. Limited evidence to guide determination of energy requirements for people with CF of all ages. Until further evidence is available, health professionals should be guided by the consensus recommendation of 110-200% of the general population energy target. Use clinical reasoning and an individualised approach to setting energy targets. ³⁰⁻⁴²
PP 7.1.1	<p>Energy requirements are likely to be elevated for people with CF. Aim for 110-200% of the recommended daily energy intake for age and gender when setting energy targets for the CF population. Take into account the following when setting individualised energy targets for people with CF:</p> <ul style="list-style-type: none"> • Nutritional status • Dietary intake • Growth pattern – aiming to achieve normal growth (avoid both undernutrition and overweight/obesity) • Clinical status (including pulmonary function) • Pancreatic function • Physical activity • Any additional requirements for weight gain/growth and nutritional repletion • Pregnancy and lactation • Transplantation
Protein	
Q 7.1.2	Are protein requirements increased in the CF population compared to the general population?
R 7.1.2	UNGRADED. Insufficient evidence to make a recommendation about protein requirements
PP 7.1.2	<p>Aim for 15-20% energy from protein. Take into account the following when setting individualised protein targets for people with CF:</p> <ul style="list-style-type: none"> • Protein intake generally increases as energy intake increases <p>A mixed high energy diet should provide adequate protein for people with CF Vegans, vegetarians, fussy/restrictive eaters, people with allergies i.e. cow's milk protein allergy and the obese CF population on an energy restricted diet, will require specific dietary advice regarding protein intake</p> <p>Further research is needed into the impact of protein quality on health outcomes in CF. Protein requirements may be elevated with malabsorption and catabolism. It is particularly important to consider the adequacy of protein intake for people with CF who have signs/symptoms of malabsorption, are unwell e.g. with a respiratory exacerbation or poorly controlled diabetes.</p> <ul style="list-style-type: none"> • Upper Limit for protein: The NHMRC recommends an upper limit for protein of 25% of energy intake for the general population,⁴³ however there is no evidence to guide a CF-specific upper limit. Evaluate individual dietary practices contributing to protein intake when intake is above 25% of energy intake; to identify if the high protein intake is contributing to a specific nutritional goal, or if other sources of energy and nutrients can be substituted without compromise to overall nutritional intake and status.
Fat	
Q 7.1.3	What is the evidence to support the routine recommendation of a high fat diet for people with CF?
R 7.1.3	GRADE D. There were no new studies included in this systematic literature review (2002-2016) to make changes to the existing recommendation for fat intake in CF from the '2006 Australasian Clinical Practice Guidelines for Nutrition in CF'. Continue to recommend an unrestricted diet that contains adequate fat to meet energy requirements. Target an intake of 100g/day if over five years of age based on the premise that a diet high in fat is less bulky and energy targets are more achievable than a diet that is low in fat.



<p>PP 7.1.3</p>	<p>Providing the person is not overweight or at risk of overweight/obesity, avoid restricting fat intake in people with CF</p> <ul style="list-style-type: none"> • Aim for 100g/day of fat for people with CF aged >5 years <p>Take into account the following in setting individualised fat targets for people with CF:</p> <ul style="list-style-type: none"> • Source and quality of fat i.e. polyunsaturated and monounsaturated versus saturated fat <ul style="list-style-type: none"> ○ Potential implications of a high saturated fat diet on cardiovascular health ○ Potential benefits of long chain polyunsaturated fatty acids on inflammation • Macronutrient distribution <ul style="list-style-type: none"> ○ Initially aim for 20-30% energy from fat, according to the recommendations for the general population ○ Up to 35-40% energy from fat is considered acceptable for the paediatric CF population and for those requiring a high energy density diet (e.g. for nutritional repletion).
<p>Q 7.1.4</p>	<p>What are the recommendations for fibre in people with CF?</p>
<p>R 7.1.4</p>	<p>UNGRADED. Insufficient evidence to make a recommendation about fibre intakes.</p>
<p>PP 7.1.4</p>	<p>Aim for a moderate fibre intake in line with the general population recommendations of 14-30g dietary fibre per day. Refer to the Nutrient Reference Values for Australia and New Zealand webpage, https://www.nrv.gov.au/nutrients/dietary-fibre for specific age and gender recommendations.</p>
<p>Essential Fatty Acids</p>	
<p>Q 7.2. 1</p>	<p>Does dietary supplementation with omega-3 essential fatty acids improve health outcomes in people with CF?</p>
<p>R 7.2.1</p>	<p>GRADE C. Dietary supplementation with omega-3 fatty acids may improve health outcomes for people with CF, however, the evidence is insufficient to recommend routine use of omega-3 supplementation. ⁴⁴⁻⁴⁶</p>
<p>PP 7.2.1</p>	<p>People with CF may be at risk of EFA deficiency</p> <ul style="list-style-type: none"> • The prevalence of EFA deficiency in CF is unknown <p>Omega-6 fatty acids - Includes linoleic acid (LA), a precursor of arachidonic acid (AA)</p> <ul style="list-style-type: none"> • LA occurs in seed oils (sunflower, safflower and corn) • AA occurs in meat, poultry and eggs • Can exert a pro-inflammatory effect <p>Omega-3 fatty acids - Includes alpha linolenic acid (ALA), a precursor of the long-chain PUFAs (Eicosapentaenoic acid (EPA), docosahexaenoic acid (DHA) and docosapentaenoic acid (DPA))</p> <ul style="list-style-type: none"> • ALA occurs in legumes, canola oils, margarine, linseed oils and nuts (walnuts) • Long-chain omega-3 PUFAs occur in fish oils • Known for their anti-inflammatory properties <p>Source of supplementation</p> <ul style="list-style-type: none"> • Insufficient evidence to suggest that any type or combination of omega-3 EFA (dietary or commercial) is superior. <p>Take into account the following prior to recommending omega-3 supplements to people with CF:</p> <ul style="list-style-type: none"> • Safety – intakes above 5000mg/d have been associated with an increase in oxidative stress and gastrointestinal discomfort • Efficacy – long term efficacy of omega-3 supplementation in CF is unknown • Cost of commercial omega-3 supplements • Impact on burden of treatment and adherence • May be better tolerated with meals (and PERT)

Chapter 8 Fat Soluble Vitamins	
Vitamin A	
Q 8.1.1	How should vitamin A be assessed for people with CF?
R 8.1.1	<p>UNGRADED. There is insufficient evidence to make a CF-specific recommendation about assessing vitamin A status.</p> <p>In the absence of evidence health professionals should continue to use serum retinol when assessing vitamin A status.</p> <p>Explore the addition of tests to assist with the interpretation of vitamin A status including, zinc, retinol binding protein (RBP), an inflammatory marker and retinol : RBP molar ratio, though there is limited evidence for their use in CF.</p> <p>Serum retinyl esters may be tested for the assessment of vitamin A toxicity.</p> <p>There is no evidence to recommend the routine assessment of β-carotene levels.</p>
PP 8.1.1	<p>Serum retinol is the most common and readily available measure of vitamin A deficiency status, however there is significant variability in what is used to define deficiency, adequacy and excess.</p> <ul style="list-style-type: none"> • Interpret results using reference ranges provided by the laboratory doing the test. • Where possible measure levels when clinically stable. Acute illness may result in decreased serum retinol. • Ideally measure when fasting. Non-fasting levels may reflect recent intake of vitamin A. <p>If low serum retinol levels despite recommended supplementation consider:</p> <ul style="list-style-type: none"> • Adherence with recommended vitamin supplementation • Whether supplements are being taken with PERT and fat containing meal <p>Measure the following to assist in the interpretation of serum retinol;</p> <ul style="list-style-type: none"> ○ A marker of inflammatory status such as CRP ○ Zinc ○ Retinol binding protein ○ Other fat soluble vitamin levels <ul style="list-style-type: none"> • Acute illness / increased inflammation will result in increased inflammatory markers and decreased levels of serum retinol, zinc and RBP. • If retinol, zinc and RBP are all low in the setting of raised inflammatory markers, then results most likely reflective of acute illness not vitamin status. Recommend re-assess levels when patient clinically stable. • If serum retinol is still low despite normal inflammatory markers, serum zinc and RBP, measure the molar ratio of RBP : retinol. This will assist in the interpretation of retinol levels and the adequacy or excess of supplementation. A ratio <0.8 suggests deficiency of vitamin A. Supplement vitamin A as per recommendation 8.1.3 and practice point 8.1.3. • If serum zinc is low or zinc status assessed as likely deficient, supplementation of zinc may be beneficial. Note that serum zinc is not a sensitive or specific test of zinc status and zinc may be normal even with subclinical zinc deficiency (Chapter 9). • If RBP is low in the setting of normal inflammatory markers, be cautious with high dose supplementation, particularly in those with CF-related liver disease. Gastroenterologist advice is recommended (Chapter 11). <p>Enquire about symptoms such as poor night time vision if deficiency is suspected. Poor night vision will manifest prior to xerophthalmia.</p> <p>If high serum retinol;</p> <ul style="list-style-type: none"> • Consider if fasting levels. High levels may reflect recent intake of vitamin A. • Assess Retinol : RBP ratio. A ratio >1.0 may indicate excess intake and toxicity. <p>If the person with CF is on high dose supplementation and at risk of vitamin A toxicity;</p> <ul style="list-style-type: none"> • Assess Retinol : RBP ratio. A ratio >1.0 may indicate toxicity <p>Measure serum retinyl esters as a function of total serum retinol. Serum retinyl esters $>10\%$ of the total vitamin A pool are usually considered abnormal.</p>



Q 8.1.2	What is the role for routine supplementation of vitamin A in people with CF and pancreatic insufficiency?
R 8.1.2	<p>GRADE D. Routine supplementation of vitamin A in all people with CF and PI is recommended (table 8e) (unchanged from the '2006 Australasian Clinical Practice Guidelines for Nutrition in CF'¹).</p> <p>There is inadequate evidence at this time for the routine adjunctive supplementation of β-carotene as an antioxidant.⁴⁷⁻⁶¹</p>
PP 8.1.2	<p>For people who are pancreatic insufficient fat soluble vitamin supplementation should be commenced at diagnosis and, if indicated, continued throughout life. Aim to achieve serum retinol levels within the normal population reference ranges.</p> <p>'2006 Australasian Clinical Practice Guidelines for Nutrition in Cystic Fibrosis':</p> <ul style="list-style-type: none"> • Infants: 1500-2000 IU vitamin A /day • Young children: 1500-5000IU vitamin A /day • Older children, adolescents and adults: 2500-5000IU vitamin A / day <p>Evaluate the need for supplementation on an individual basis for the pancreatic sufficient population.</p>
Q 8.1.3	What vitamin A supplementation dose should be prescribed to treat vitamin A deficiency in people with CF?
R 8.1.3	<p>GRADE D. In the absence of quality evidence for supplementation to treat subclinical vitamin A deficiency in people with CF, it is recommended to follow the doses recommended in the '2006 Australasian Clinical Practice Guidelines for Nutrition in CF'¹ as outlined in Table 8e (Chapter 8).</p> <p>There is no evidence specific to people with CF for supplementation to treat severe deficiency. Assess supplementation on an individual basis with interdisciplinary input and referral to relevant disciplines outside of CF as appropriate.</p>
PP 8.1.3	<ul style="list-style-type: none"> • In cases of subclinical deficiency supplement as per table 8e (Chapter 8) above guidelines aiming to achieve serum retinol levels within the normal population reference ranges. • If RBP : retinol ratio indicates deficiency i.e. <0.8, increase supplementation to upper limits of recommended supplementation e.g. adults to 10 000IU. • Low RBP may occur in those with severe liver disease. Be cautious with high dose supplementation in these circumstances. Recommend consultation by gastroenterologist. See chapter 11 for supplementation recommendations in CF-related liver disease. • For vitamin A deficiency refractory to upper level of recommended supplementation, think about an empiric trial of zinc supplementation. See chapter 9 • Severe vitamin A deficiency with overt symptoms will require high dose supplementation (>10,000IU). An individualised approach with interdisciplinary input including CF physician, dietitian and appropriate other health professionals such as gastroenterologist is recommended.
Q 8.1.4	What is the safe upper limit for vitamin A supplementation in people with CF?
R 8.1.4	<p>UNGRADED. There is insufficient evidence available regarding the safe upper limit for vitamin A supplementation in CF. In the absence of evidence specific to CF, health professionals should be guided by recommendations for the general population.</p>

<p>PP 8.1.4</p>	<p>Be guided by upper limits for the general population. Note upper limits are for preformed vitamin A as retinol. Australian and New Zealand Nutrient Reference Values (2006) ⁴³;</p> <ul style="list-style-type: none"> • Infants: 2000 IU vitamin A • Young children: 2000 – 3000 IU vitamin A • Older children: 5667 IU vitamin A • Adolescents: 9333 IU vitamin A • Adults: 10 000 IU vitamin A <p>Supplementation for severe vitamin A deficiency may require doses greater than the above recommended upper limits. Supplementation doses in excess of these upper limits should be advised with caution and only following a thorough interdisciplinary assessment of potential risks and benefits. Referral to a gastroenterologist with CF experience is recommended.</p> <ul style="list-style-type: none"> • Where vitamin A toxicity is a concern, consider additional supplementation in the form of β-carotene, as excessive ingestion of this form is generally considered safe. • In the absence of an available β-carotene supplement, a regular multivitamin with greater proportion of vitamin A as β-carotene may be considered. However additional supplementation of other fat soluble vitamins may be required, with an increase cost and burden to person with CF. <p>Monitor serum retinol, RBP, retinol : RBP ratio and if available serum retinyl esters for those on high dose supplements.</p>
<p>Q 8.1.5</p>	<p>How often should vitamin A levels be measured in people with CF?</p>
<p>R 8.1.5</p>	<p>UNGRADED. There is insufficient evidence available to recommend specific monitoring and evaluation protocols for vitamin A levels in CF. Health professionals should continue to follow recommendations in the '2006 Australasian Clinical Practice Guidelines for Nutrition in CF', to assess annually and monitor more frequently in those at high risk of deficiency or toxicity¹.</p>
<p>PP 8.1.5</p>	<p>Monitor vitamin A status annually. More frequent monitoring (e.g. 3-6 monthly) is suggested in the following scenarios:</p> <ul style="list-style-type: none"> • After changing supplementation doses, especially after high dose supplementation • For people with CF-related liver disease or history of intestinal resection or malabsorption • In people with poor adherence to PERT and fat soluble vitamin supplementation • After changes to treatment for malabsorption where a change in the level of fat absorption has occurred • On some drugs such as acne medications e.g. Roaccutane®
<p>Vitamin D</p>	
<p>Q 8.2.1</p>	<p>Is vitamin D status associated with measures of respiratory health (lung function, pulmonary exacerbations, markers of inflammation) in people with CF?</p>
<p>R 8.2.1</p>	<p>Grade D. At this stage there is insufficient evidence that vitamin D status is associated with measures of respiratory health in people with CF. ⁶²⁻⁶⁸</p>
<p>PP 8.2.1</p>	<p>There is no evidence to support a causal role between vitamin D and respiratory health, however it is reasonable to assume that individuals with severe lung disease may be more likely to be vitamin D deficient due to spending more time indoors.</p>
<p>Q 8.2.2</p>	<p>Is there an ideal serum 25-hydroxyvitamin D level to aim for in people with CF?</p>
<p>R 8.2.2</p>	<p>UNGRADED. Insufficient evidence specific to CF for the ideal serum 25-hydroxyvitamin D level. It is suggested that the general Australian and New Zealand goal of ≥ 50 nmol/L⁶⁹ be used with a caveat for the time of year at which testing occurs.</p>
<p>PP 8.2.2</p>	<p>It is suggested that the general Australasian goal of ≥ 50 nmol/L serum 25(OH)D be used if measuring vitamin D at the end of winter or in early spring⁶⁹. If testing at other times of year, aim for a level 10-20nmol/L higher (i.e. ≥ 60-70nmol/L).</p>
<p>Q8.2.3</p>	<p>Is the time of year, specifically the season, important when measuring and interpreting an individual's serum vitamin D level?</p>



R8.2.3	GRADE C. Aim to measure serum vitamin D at the end of the winter months and adjust supplementation accordingly. If not feasible, take into account the season of assessment when interpreting results and prescribing supplementation. ^{51,64,65,70-74}
PP8.2.3	<ul style="list-style-type: none"> • Aim to measure serum vitamin D at the end of winter/early spring • Take into account the following when interpreting results: <ul style="list-style-type: none"> • ≥50nmol/L = adequate at the end of winter or in early spring • ≥60-70nmol/L = adequate at other times of the year • Specific considerations for the Australian and NZ context: <ul style="list-style-type: none"> • Seasonal variations may differ according to geographic location <p>People from far north of Australia who spend time outdoors during the winter months, may not see as much seasonal variation in serum vitamin D levels</p>
Q8.2.4	Should supplemental vitamin D be given to people with pancreatic sufficient cystic fibrosis as part of routine care?
R8.2.4	GRADE C. There is inconsistent evidence to support routine vitamin D supplementation for all people with CF, regardless of pancreatic status. It is recommended that all people with CF undergo annual serum vitamin D testing and be supplemented accordingly. ^{51,64,65,71,72,74-76}
PP 8.2.4	Consider all individuals, including those with pancreatic sufficiency at risk of vitamin D deficiency and screen annually. Supplement as required.
Q 8.2.5	What doses of vitamin D are needed to prevent deficiency in people with CF?
R 8.2.5	UNGRADED. There is insufficient evidence available to recommend evidence-based routine supplementation doses for people with CF
PP 8.2.5	<p>Base routine supplementation of vitamin D on the US and European consensus documents^{77,78}:</p> <ul style="list-style-type: none"> ○ Infants = 400-1000 IU ○ Young children = 800-2000 IU ○ Older children, adolescents and adults = 800-4000 IU <p>Additional points:</p> <ul style="list-style-type: none"> • Take into account medication adherence and cost when prescribing supplementation • Ergocalciferol and cholecalciferol are the two forms of vitamin D available • Cholecalciferol should be used as it most effective in increasing serum 25OHD levels. • Cholecalciferol is the major form of supplemental vitamin D currently available in Australia and New Zealand.
Q 8.2.6	What doses of vitamin D are needed to correct deficiency in people with CF?
R 8.2.6	<p>GRADE C. There is a lack of evidence on conventional, daily doses of vitamin D needed to correct vitamin D deficiency.</p> <ul style="list-style-type: none"> ○ Health professionals should be guided by consensus based guidelines^{77,78} <p>There is some evidence to support the use of high dose cholecalciferol ("STOSS therapy") in CF. Use with caution due to risk of toxicity in those who are unable to convert excess cholecalciferol to its inactive form. ^{79,80}</p>
PP 8.2.6	<p>Base supplementation of vitamin D to correct deficiency on the US and European consensus documents^{77,78}:</p> <ul style="list-style-type: none"> • Infants = 400-2000 IU • Young children = 1000-5000 IU • Older children, adolescents and adults= 1000-10 000 IU <ul style="list-style-type: none"> • Before escalating treatment of vitamin D deficiency, check adherence to PERT and prescribed vitamin supplementation. • Use of high dose vitamin D supplementation (STOSS) should be carefully evaluated and done in conjunction with an endocrinologist with experience in CF. Some people are unable to convert excess cholecalciferol to its inactive form and are therefore at increased risk of toxicity. • Refer to an endocrinologist if people are unresponsive to maximal treatment doses.

Vitamin E	
Q 8.3.1	How should vitamin E status be assessed for people with CF?
R 8.3.1	<p>UNGRADED. There is insufficient evidence to make a CF-specific recommendation about assessing vitamin E status.</p> <p>In the absence of evidence health professionals should continue to use serum α-tocopherol when assessing vitamin E status. Lipid adjustment may provide a more accurate assessment of vitamin E status and its use may be explored where available.</p>
PP 8.3.1	<p>Serum or plasma levels of α-tocopherol are the most common measure used to assess vitamin E status. However there is significant variability in what is used to define deficiency, adequacy and excess.</p> <ul style="list-style-type: none"> Interpret results using reference ranges provided by the laboratory doing the test. <p>Measurement of α-tocopherol to total lipid ratio will aid in the interpretation of serum vitamin E status in the following situations:</p> <ul style="list-style-type: none"> Abnormal serum α-tocopherol levels Abnormal lipid levels (common in CF and liver disease) <ul style="list-style-type: none"> Normal ratio in children = 0.6mg/g Normal ratio in adults = > 0.8mg/g <p>When not available, the α-tocopherol to cholesterol ratio can be used in place of total lipid ratio although;</p> <ul style="list-style-type: none"> Overall a less sensitive and specific test Aim for a ratio >5.4mg/g in CF Where possible measure fasting levels of total lipid ratios. <p>There is no evidence for the assessment of other tocopherols including γ-tocopherol in CF</p>
Q 8.3.2	What is the role for supplementation of vitamin E in people with CF?
R 8.3.2	<p>GRADE C. Routinely supplement vitamin E in people with CF who are pancreatic insufficient.</p> <p>For pancreatic sufficient individuals commence supplementation on an individual basis.</p> <p>There is inadequate evidence to establish recommendations for supplement dose specifically for CF. Overall the evidence is insufficient to recommend change from current practice as per the '2006 Australasian Clinical Practice Guidelines for Nutrition in CF' (Table 8j) (Chapter 8).¹ 47-54,81-86</p>
PP 8.3.2	<p>Routine supplementation of vitamin E is recommended for all pancreatic insufficient people with CF. Aim to achieve the normal population reference ranges. At this time, continue to follow the '2006 Australasian Clinical Practice Guidelines for Nutrition in CF' when supplementing vitamin E in CF':</p> <ul style="list-style-type: none"> Infants: 40 - 80IU/day Children 1 to 3 years: 50 - 150IU/day Children 4 to 7 years: 150 - 300IU/day Children > 8 years & Adults: 300 - 500IU/day <p>Consider supplementation on an individual basis for the pancreatic sufficient population.</p> <p>Higher supplementation doses may be required if significantly increased dietary and/or supplemental polyunsaturated fatty intakes.</p> <ul style="list-style-type: none"> If ongoing deficiency despite recommended levels of supplementation, consider adherence to prescribed supplement and PERT before increasing the dose further. Water-miscible vitamin E preparations are generally more bioavailable than fat soluble preparations.



Q 8.3.3	What is the safe upper limit for vitamin E supplementation in people with CF?
R 8.3.3	UNGRADED. There is insufficient evidence available regarding the safe upper limit for vitamin E supplementation in CF. Health professionals should be guided by the upper limit of the age-specific supplementation ranges recommended in the '2006 Australasian Clinical Practice Guidelines for Nutrition in CF' for routine supplementation of vitamin E (table 8j) ¹ . Make a thorough assessment of risks and benefits before considering higher supplement doses as required to correct deficiency states, and closely monitor the response to supplementation.
PP 8.3.3	A safe upper limit has not yet been determined for vitamin E supplementation in CF. No evidence of vitamin E toxicity in CF. Supplementation above the upper recommended dose should only be considered following thorough dietary and clinical assessment and based on serum levels.
Q 8.3.4	How often should vitamin E levels be measured in people with CF?
R 8.3.4	UNGRADED. Insufficient evidence to make a recommendation. Health professionals should continue to follow recommendations in the '2006 Australasian Clinical Practice Guidelines for Nutrition in CF', to assess annually and to monitor more frequently in those at high risk of deficiency or toxicity ¹ .
PP 8.3.4	Assess vitamin E status annually. Monitor more frequently in the following groups: <ul style="list-style-type: none"> • Infants: 2 months post first commencing supplementation ⁸⁷ • After changing supplementation doses i.e. 3-6 months post changes • After changes to treatment for malabsorption where a change in the level of fat absorption has occurred. • People with CF-related liver disease or history of intestinal resection or malabsorption • People with poor adherence to PERT and fat soluble vitamin supplementation
Vitamin K	
Q 8.4.1	How should vitamin K status be assessed for people with CF?
R 8.4.1	UNGRADED. There is insufficient evidence to make a recommendation about methods for assessing vitamin K status. At this time, it is recommended that health professionals assess vitamin K status using the best readily available biochemical measure together with a thorough diet and clinical assessment.
PP 8.4.1	<ul style="list-style-type: none"> • There is no readily available direct measure of vitamin K status (sufficiency or deficiency). • Serum vitamin K is unreliable and should not be used to assess vitamin K status in CF. • PIVKA-II (protein induced vitamin K absence-II) and uc-OC (undercarboxylated osteocalcin) are considered the most accurate measures of vitamin K status. <ul style="list-style-type: none"> ◦ Not readily available in clinical practice in Australia and New Zealand • Prothrombin Time (PT) is a measure of coagulation and is often used as a more readily available surrogate measure of vitamin K status. Consider the following prior to use: <ul style="list-style-type: none"> ◦ Insensitive and non-specific ◦ Marker of advanced vitamin K deficiency. ◦ Not recommended in infants, other than those with CF-related liver disease as it requires large amount of blood for collection ◦ Ideally assess biochemical status using a more accurate measure such as PIVKA-II, however where not available, use surrogate measure of PT.
Q 8.4.2	Should vitamin K supplementation be recommended for all people with CF and pancreatic insufficiency?
R 8.4.2	GRADE C. Routinely supplement vitamin K in all people with CF and pancreatic insufficiency as outlined in table 8l (Chapter 8). In practice supplementing at these doses will require an increase in vitamin K supplementation doses that are routinely provided and currently available in Australia and NZ. There is insufficient high quality evidence available to recommend an optimal dose. ^{85,88-95}

PP 8.4.2	<p>Vitamin K supplementation is recommended for all pancreatic insufficient people with CF. At this time, it is recommended to follow the most recently released international guidelines for vitamin K supplementation dosing in CF.</p> <ul style="list-style-type: none"> • <i>ESPEN-ESPGHAN-ECFS guidelines on nutrition care for infants, children and adults with cystic fibrosis</i>⁷⁸ <ul style="list-style-type: none"> ○ Infants: 300 - 1000µg ○ Children & Adults: 1000 - 10 000 µg <p>Higher doses may be required for those with CF-related liver disease, intestinal resection or when on longer term antibiotic or steroid regimens.</p> <p>Daily administration of vitamin K is preferred due to the body's low storage capacity.</p> <p>Vitamin K preparations in Australia and New Zealand all contain vitamin K1 (phytomenadione), however in amounts well below recommended supplementation doses.</p> <p>Additional supplementation of vitamin K may be required particularly for those considered at high risk of deficiency and/or with bone disease, however, evaluate the availability, cost and treatment burden for people with CF.</p>
Q 8.4.3	How often should vitamin K levels be measured in people with CF?
R 8.4.3	UNGRADED. Insufficient evidence to make a recommendation. Aim to assess vitamin K status at diagnosis and annually in all people with CF.
PP 8.4.3	<p>Aim to assess vitamin K status annually.</p> <ul style="list-style-type: none"> • Additional screening for vitamin K deficiency should be considered for newly diagnosed patients and those with haemoptysis/haematemesis, CF-related liver disease or recent intestinal resection. <p>Vitamin K status should ideally be re-checked 3 months after any change to vitamin K supplementation or treatment for malabsorption.</p>

Chapter 9 Minerals

Iron

Q 9.1.1	How should iron status be assessed in people with CF?
R 9.1.1	GRADE C. The level of evidence to guide practice for assessing iron status in CF is insufficient. Further research or expert consensus is required. Until further evidence is available, it is suggested that iron status in CF be assessed as per guidelines for the general population. ^{96,97}
Q 9.1.2	How should iron deficiency be treated in people with CF?
R 9.1.2	UNGRADED. Insufficient evidence to make a recommendation
Q 9.1.3	Is iron supplementation contraindicated for people with CF who are chronically colonised with pseudomonas aeruginosa (PA)?
R 9.1.3	GRADED. There is insufficient evidence from clinical trials to suggest that iron supplementation is contraindicated in adults and children with CF who are chronically colonised with <i>Pseudomonas aeruginosa</i> . When indicated, an iron supplement should be prescribed for adults and children with CF who are chronically colonised with <i>Pseudomonas aeruginosa</i> . ⁹⁸
PP 9.1.1, 9.1.2 and 9.1.3	<p>Increased risk of iron deficiency in CF due to chronic inflammation, inadequate dietary intake, gastrointestinal comorbidities and haemoptysis. Iron studies are difficult to interpret in CF due to chronic inflammation. Aim to assess iron status during clinical stability.</p> <p>Interpretation of biochemical markers in CF - Serum ferritin is an acute phase protein (rises during periods of inflammation) and may be unreliable. Inflammatory markers, including C-reactive protein (CRP) should be taken into consideration.</p> <ul style="list-style-type: none"> • Serum transferrin receptor (sTfr) is not readily available but should be considered as it is not affected by inflammation. A raised sTFR may be a useful indicator of functional iron deficiency in CF.



<p>PP 9.1.1, 9.1.2 and 9.1.3</p>	<p>Absolute iron deficiency - Iron stores are depleted, as indicated by serum ferritin (low), serum iron (low), transferrin (high), transferrin saturation (low), sTfR (high), CRP (normal)</p> <ul style="list-style-type: none"> • Oral iron supplementation is recommended <p>Functional iron deficiency - Iron stores are normal-high but not available at the site of erythroblast production. Serum ferritin (low – normal), serum iron (low), transferrin (normal-high), transferrin saturation (low), sTfR (high), CRP (high)</p> <ul style="list-style-type: none"> • Oral iron supplementation may be required <p>Dietary considerations Increasing dietary iron intake is often inadequate in the treatment of iron deficiency in CF. Iron is available in food as haem iron (more bioavailable) and non-haem iron (less bioavailable).</p> <ul style="list-style-type: none"> • Meat, seafood and poultry are good sources of haem iron. • Plant-based foods (wholegrain cereal and green leafy vegetables) and iron-fortified foods (infant rice-cereal) are good sources non-haem iron. • Foods high in vitamin C improve the absorption of iron. • Foods high in calcium, phytates (legumes, rice and other grains) and tannins (tea) can inhibit the absorption of iron. <p>Oral iron supplement considerations Prescribe an iron supplement, in addition to dietary change, for 2-3 months to treat diagnosed iron deficiency. Suggested treatment doses:</p> <ul style="list-style-type: none"> • Children: 3-6mg/kg/d elemental iron for 2-3 months after serum Hb normalises • Adolescents & adults: 100-200mg elemental iron daily for 3-6 months after Hb normalises • Gastrointestinal complaints (including constipation and epigastric pain) are common side effects of oral iron supplements. <p>Multivitamins are not recommended in the treatment of iron deficiency. Concerns regarding potential drug-nutrient interactions should be discussed with the CF pharmacist.</p> <p>Iron supplementation is not contraindicated for people with CF with chronic <i>Pseudomonas aeruginosa</i> infection.</p> <p>Intravenous iron supplementation Iron (ferric carboxymaltose) 500mg/10ml injection is associated with fewer adverse events than other IV iron supplements. It is also available on the pharmaceutical benefits scheme (PBS) in Australia and via PHARMAC in New Zealand (District Health Board hospitals only)</p>
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Magnesium

Q 9.2.1 Does supplementing magnesium above the RDI improve nutrition and/or respiratory outcomes in people with CF?

R 9.2.1 **GRADE D.** There is insufficient evidence to support that routine magnesium supplementation above the RDI improves health outcomes in people with CF. Explore the use of oral magnesium supplementation only when dietary intake is unable to meet the RDI.⁹⁹

PP 9.2.1 Encourage people with CF to achieve adequate consumption of magnesium as part of a varied diet.

- Foods high in magnesium include green leafy vegetables, unrefined cereals, legumes, nuts & shellfish
- Magnesium deficiency is likely to co-exist with other micronutrient deficiencies. A nutrition review should therefore consider overall micronutrient adequacy.

Oral magnesium supplementation is considered safe and cost-effective

High dose magnesium supplementation may result in gastrointestinal side effects, especially diarrhoea. This is most commonly seen in patients on higher dose magnesium supplements after lung transplantation.

Calcium

No PICO questions were formulated for section 9.3. Refer to [Chapter 9](#), section 9.3 Calcium for narrative text.

Sodium	
Q 9.4.1	How do environmental factors and exercise impact on sodium requirements for people with CF compared to those without CF?
R 9.4.1	GRADE C. Climate (heat and humidity) is thought to have an impact on salt requirements in CF. There is also some evidence to support an altered thirst drive for people with CF. However, at this time, there is insufficient evidence available to conclude how environmental factors and exercise impact on sodium requirements for the wider CF population. ¹⁰⁰⁻¹⁰²
PP 9.4.1	Take into account the following when evaluating sodium requirements: <ul style="list-style-type: none"> • Infants & people with CF exposed to hot / humid environments or illness are at high risk of sodium depletion & hyponatraemia. • Signs & symptoms of sodium depletion include nausea, vomiting, muscle cramps, deposition of salt crystals on the skin, fatigue, poor growth (especially in infants) and/or hyponatraemia • Sweat sodium losses vary amongst individuals (with and without CF) • Dietary intake, sweat rate, hydration and heat acclimation can impact on sodium losses. • Sweat rates/sodium losses are elevated and thirst drive potentially diminished for people with CF in hot / humid conditions and during exercise. Dehydration/hyponatraemia is a risk under these conditions.
Q 9.4.2	What is the recommended daily sodium requirement for people with CF compared to those without CF?
R 9.4.2	GRADE D. There is a lack of research available to guide sodium requirements for people with CF. As a result, recommendations vary in international consensus and review documents. Recommendations for daily sodium requirements in CF are: <ul style="list-style-type: none"> • Infants - 500-1000mg • Children - 1000-4000mg • Adolescents and adults - 6000mg (unchanged from the '2006 Australasian Clinical Practice Guidelines for Nutrition in CF')
PP 9.4.2	<ul style="list-style-type: none"> • Clinicians should continue to use nation-specific guideline/consensus documents (including the '2006 Australasian Clinical Practice Guidelines for Nutrition in Cystic Fibrosis' in addition to a thorough nutrition assessment and clinical judgment as a guide when recommending sodium supplementation to people with CF ¹. • Serum sodium is not a sensitive marker for salt depletion in CF. • Undertake a spot urine sodium analysis if sodium depletion is suspected or supplementation significantly changes. • Clinicians should be guided by local climate-based recommendations and clinical judgment when individually tailoring sodium supplementation to people with CF.
Zinc	
Q 9.5.1	How should Zinc status be assessed for people with CF?
R 9.5.1	Grade D. The level of evidence to guide practice for assessing zinc status in CF is insufficient. Further research or expert consensus is required. ¹⁰³



PP 9.5.1	<p>Assess zinc status and monitor empiric trials of zinc supplementation using a combination of dietary, biochemical and clinical/functional indicators.</p> <p>Serum/Plasma Zinc</p> <ul style="list-style-type: none"> • Measure using the local laboratory reference ranges. • Analyse levels in the context of dietary and clinical information. Consider; <ul style="list-style-type: none"> ○ Zinc is an insensitive marker of deficiency however may be helpful diagnostically in severe zinc deficiency ○ Levels are best measured when person with CF is clinically stable. Where acute phase response and inflammation is suspected, check CPR. ○ There is high individual biological variation in zinc levels ○ There is diurnal variation in zinc levels and levels may reflect recent dietary intake. Recommend where possible to measure fasting levels. <p>Clinical Indicators</p> <ul style="list-style-type: none"> • Marginal zinc deficiency may be diagnosed in some patients via a positive response to zinc supplementation e.g. improved growth. • Evaluate differential diagnoses as other conditions may present with similar signs and symptoms of zinc deficiency. <p>Diet</p> <ul style="list-style-type: none"> • Attention should be given to those at high risk of inadequate zinc intakes/absorption; <ul style="list-style-type: none"> ○ Strict vegetarian diets with high intake of phytates ○ Infants older than 6 months exclusively breastfed or consuming limited high bioavailable zinc foods such as fortified cereals or meat ○ High iron supplementation <p>Assess adequacy of protein and essential fatty acids because deficiency may manifest similarly to zinc deficiency.</p>
Q 9.5.2	What are the recommendations for zinc supplementation in people with CF?
R 9.5.2	<p>Grade D. There is insufficient evidence to make recommendations for routine supplementation or supplementation for suspected zinc deficiency in CF. Until further evidence is available, it is suggested that zinc supplementation be guided by recommendations in CF consensus guidelines. As per the 2016 ESPEN-ESPGHAN-ECFS CF nutrition guidelines⁷⁸, CF people at high risk of deficiency should receive the following supplementation doses for 6 months; infants (1mg/kg/d), children (15mg/d), adults (25mg/d).¹⁰³⁻¹¹⁰</p>
Q. 9.5.3	What is the safe upper limit for zinc supplementation in CF?
R 9.5.3	UNGRADED. Insufficient evidence to make a recommendation.
PP 9.5.2 and 9.5.3	<p>Suggested supplementation doses.</p> <ul style="list-style-type: none"> • Infants (<2yrs) with persistent failure to thrive and/or those with severe steatorrhea: <ul style="list-style-type: none"> ○ Consider a trial of zinc supplementation ○ 1mg elemental zinc/kg/d in divided doses for 6 months (max 15mg/day). • Children with suspected zinc deficiency: <ul style="list-style-type: none"> ○ 15mg/d for 6 months • Adults with suspected zinc deficiency: <ul style="list-style-type: none"> ○ 25mg/day for 6 months • In vitamin A deficiency refractory to Vitamin A supplementation, consider an empiric trial of zinc supplementation. • The amount of zinc in the CF-specific multivitamin, VitABDECK®, is not adequate to correct zinc deficiency and additional zinc is likely required. • The main dietary source of zinc are animal foods as well as zinc fortified cereals • Where practical, zinc is best tolerated if given in divided doses

Chapter 10 Pancreatic Enzyme Replacement Therapy (PERT)	
Q 10.1.1	Does gastric emptying rate impact PERT efficacy in people with CF?
R 10.1.1	GRADE C. Gastric emptying rate may impact PERT efficacy and should be considered in people with CF. ^{111,112}
Q 10.1.2	Does the timing of PERT administration in relation to a meal impact PERT efficacy in people with CF?
R 10.1.2	GRADE D. Limited evidence suggests PERT is equally effective when taken before or after a meal in individuals with CF. It also suggests that for some individuals, changing PERT timing in relation to a meal may improve PERT efficacy. A change of PERT timing can be considered for people with symptoms of fat malabsorption or poor growth once other treatment strategies such as adherence have been taken into account. ¹¹²
PP 10.1.1 and 10.1.2	<p>Prior to changing the timing of PERT in patients with symptoms of fat malabsorption and poor growth evaluate:</p> <ul style="list-style-type: none"> • Is the patient compliant with PERT? • Is PERT distributed appropriately according to fat content? <p>Explore the role of gastric emptying rate and referral to a gastroenterologist for consideration (± formal gastric emptying assessment). People with fast gastric emptying may benefit from taking PERT before a meal.</p>
Q 10.1.3	How should PERT be dosed for people with CF to support optimal fat absorption?
R 10.1.3	GRADE D. There is inconsistent and insufficient evidence to recommend specific doses of PERT required to support <i>optimal</i> fat absorption in individuals with CF. A wide range of doses have been shown to be effective. ¹¹³⁻¹³¹
PP 10.1.3	<p>PERT Dosing Recommendations</p> <p>Dosing recommendations adapted from international recommendations:</p> <ul style="list-style-type: none"> • 2006 Australasian Clinical Practice Guidelines for Nutrition in Cystic Fibrosis¹ • 2008 Evidence-Based Practice Recommendations for Nutrition-Related Management of Children and Adults with Cystic Fibrosis and Pancreatic Insufficiency¹³² • 2009 Cystic Fibrosis Foundation evidence-based guidelines for the management of infants with cystic fibrosis⁸⁷. • 2016 ESPEN-ESPGHAN-ECFS guidelines on nutrition care for infants, children, and adults with cystic fibrosis⁷⁸ <p>Infants</p> <ul style="list-style-type: none"> • Breastfeeds and Infant Formula <ul style="list-style-type: none"> ○ Initiate at 2500-5000 IU lipase per breastfeed or formula feed and adjust up according to weight gain and bowel symptoms to a maximum of 10 000 IU lipase/kg/day* • Solids <ul style="list-style-type: none"> ○ Approximately 2000 IU lipase/g fat ○ Maximum 10 000 IU lipase/kg/day* <p>Children and Adults</p> <ul style="list-style-type: none"> • 500-4000 IU lipase/g fat. Maximum 10 000 IU lipase/kg/day* <p>General PERT recommendations</p> <ul style="list-style-type: none"> • Aim for the lowest effective dose • Use an individualised approach • Distribute the enzymes throughout the day according to the fat content of food and drinks consumed • Monitor weight, growth and bowel symptoms • Individuals should be encouraged to discuss PERT with clinic staff before increasing dose • Branded PERT preparations should be used



	<p>Distribution Ensure PERT is correctly distributed over the day's meals based on the fat content of food and drinks consumed</p> <p>Administration</p> <ul style="list-style-type: none"> • Capsules should be swallowed whole or the granules mixed in with an acidic fruit puree – e.g. apple puree. Granules should not be chewed • PERT should be given with all meals, snacks and food containing fat • PERT may be given before, during or after a meal** <p>Physical Storage Store capsules in an airtight container in a cool, dry place – see specific product information for more information on storage. Ensure capsules have not exceeded the expiry date</p> <p>* In some situations the upper limit may need to be exceeded in the short term, such as infants feeding frequently – this should be done with caution and with regular review by a gastroenterologist and dietitian</p> <p>**new recommendation</p>
Q 10.1.4	Is there evidence to support the use of acid suppression medications to improve PERT efficacy for people with CF?
R 10.1.4	GRADE C. There is inconsistent and limited evidence to support for or against the use of acid suppression medication to improve PERT efficacy by increasing fat absorption for individuals with CF. Further research is required. ¹³³⁻¹³⁵
PP 10.1.4	Take into account the following prior to commencing acid suppression medication in people with CF with symptoms of steatorrhoea and on high dose PERT: <ul style="list-style-type: none"> • Is the patient adherent with PERT? • Is PERT distributed appropriately according to fat content? • Role of gastric emptying rate and referral to a gastroenterologist for consideration (± formal gastric emptying assessment) • Increasing the dose by stepwise increments to a maximum of 10 000 IU lipase/kg/day • If a trial of acid suppression medication is commenced review its effect regularly, e.g. 3-6 monthly
Q 10.1.5	What are the risks and long-term health implications associated with phthalate exposure via PERT to people with CF?
R 10.1.5	UNGRADED. Insufficient evidence to make a recommendation
PP 10.1.5	Provide people with CF and their families with the latest information regarding phthalates and PERT on request. <p>Phthalate polymers, including hypromellose phthalate (HMP), are non-active ingredients in the enteric coating of many medications, including all Creon® products available in Australia and NZ.</p> <ul style="list-style-type: none"> • Unlike other phthalates that degrade to potentially harmful monoesters, phthalate polymers are considered to be of low or no known toxicity risk. • In Australia the Therapeutic Goods Administration (TGA) lists phthalate polymers as an ingredient for use in prescription medications without any restriction (www.tga.gov.au). <p>All PERT products in Australia are approved by the TGA for use, therefore concern about phthalates is not an indication on its own for changing the choice of PERT preparation. Exploring such a change for an individual should be based on factors such as adequacy of control of malabsorption, and/or the occurrence of side effects.</p> <p>Panzytrat 25 000® does not contain phthalates or the HMP polymer.</p>

Chapter 11 Gastrointestinal and Hepatobiliary Considerations

Gastro-oesophageal Reflux

Q 11.1.1

What are the nutrition considerations for the management of gastro-oesophageal reflux (GOR) in CF?

R 11.1.1

GRADE D. Specific dietary factors that influence the occurrence, severity and management of GOR in CF have not been identified. Further research into the impact of dietary factors on reflux in CF is warranted. ¹³⁶⁻¹³⁹

PP 11.1.1

- GOR is common in children and adults with CF and can present as either symptomatic or asymptomatic.
- The Mayo GER questionnaire (GERQ) and the Gastroesophageal Reflux Disease Symptom Assessment Scale (GSAS) are both validated self-administered questionnaires that have been used to record reflux symptoms and severity in the general population.
- There are no established guidelines for the diagnosis and treatments of GOR specific to CF. Use clinical judgment when applying GOR guidelines for general population to individuals with CF.
- Pharmacological therapy options to reduce the symptoms of GOR are often first choice of treatment. These include histamine receptor antagonists (H₂ antagonists) and protein pump inhibitors (PPIs).
- If dietary interventions are considered, take an individualised approach whereby nutritional adequacy is not compromised or unnecessarily restricted.
- Supine positioning may exacerbate GOR. Review bed head elevation, feed volumes and rate of feeding to optimise tolerance and reduce the risk of symptoms. Assess reflux symptoms prior to enteral feeding.
- Surgical intervention (fundoplication) may be explored if symptoms have not been controlled by pharmacological, dietary or lifestyle interventions. It is important for the interdisciplinary team to evaluate the potential risks and benefits of the surgery for each individual.

DIOS and Constipation

Q 11.2.1

What are the nutrition considerations for the prevention and management of Distal Intestinal Obstruction Syndrome (DIOS) in CF?

R 11.2.1

GRADE C. Inadequate PERT, including poor adherence and under-dosing, is unlikely to play a role in DIOS but should still be assessed as part of an overall dietetics review in CF.

The impact of diet, particularly fibre and fluid intake on DIOS is unclear.

Further research in the Australian and New Zealand context, particularly in regards to the impact of hydration on DIOS is required. While not examined in the current body of evidence, the impact of sodium intake on hydration and DIOS is warranted in future research. ¹⁴⁰⁻¹⁴²

Q 11.2.2

What are the nutrition considerations for the prevention and management of constipation in CF?

R 11.2.2

GRADE D. There is inadequate evidence to recommend nutrition considerations in the prevention and management of constipation in CF.

Until further evidence is available, complete a thorough diet history, including assessment of hydration (fluid and sodium intake) as well as fibre intake. Review PERT to optimise absorption.

Further research in the Australian and New Zealand context is warranted, particularly in regards to hydration (including sodium intake) and constipation in CF. ¹⁴³



<p>PP 11.2.1 and 11.2.2</p>	<ul style="list-style-type: none"> • Medical treatment is a priority, particularly for the diagnosis and management of DIOS. • Surgical intervention is rarely required. • Polyethylene glycol (PEG) laxatives are usually used for the treatment of constipation and are often a first line treatment for DIOS. Many patients continue on laxatives after the resolution of DIOS. • Optimise fluid and salt intake as well as fat absorption in those who present with or have a history of constipation and DIOS. • Patients with complete DIOS may be fasted during initial treatment. In most cases this is short term, however, monitor patients for risk of malnutrition. Total parenteral nutrition (TPN) may be required for more complex cases that do not resolve in a few days. • The Bristol stool chart can be used when assessing bowel patterns in CF. A paediatric version is also available when trying to engage younger children. <p>Laxatives:</p> <ul style="list-style-type: none"> • Insufficient evidence to assess the relative effectiveness or tolerability of different classes of laxatives. Considerations when choosing an agent should include hardness of the stool, potential adverse effects, effectiveness of previous treatments and patient preference. • Adjust laxative regimens according to response and tolerability. • Osmotic laxatives draw water into the stool to help soften the stool and washout the colon. The active ingredient in most osmotic laxatives is usually Polyethylene glycol (PEG). Lactulose, a non-absorbable sugar, is another osmotic laxative used in CF but is often associated increased abdominal pain/cramping. • Stool lubricants help lubricate the bowel wall & soften faecal mass to allow the faecal mass to transit through the colon. <p>NOTE - Mineral oils should not be used for the treatment of DIOS & constipation in children less than 12 months of age. Potential side effects of longer term use in children greater than 12 months of age include:</p> <ul style="list-style-type: none"> • Possibility of reduced fat soluble vitamins. • Abdominal cramps • Risk of aspiration if used during period of respiratory exacerbation or if the child is fighting the dose.
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Colon Cancer Screening

<p>Q 11.3.1</p>	<p>What are the nutrition considerations for colon cancer screening in CF?</p>
<p>R 11.3.1</p>	<p>UNGRADED. Insufficient evidence to make a recommendation.</p>
<p>PP 11.3.1</p>	<p>People with CF have a greater risk of developing colon cancer than the general population. Individual centres need to develop local guidelines with regard to screening older adults to assess risk of colorectal cancer.</p> <ul style="list-style-type: none"> • More extensive colonoscopy preparation may be required and further research is required.

Liver Considerations

<p>Q 11.4.1</p>	<p>Should vitamin K supplementation be recommended for all people with CF-related liver disease?</p>
<p>R 11.4.1</p>	<p>UNGRADED. Insufficient evidence to make a recommendation – follow recent consensus guidelines. ¹⁴⁴</p>
<p>PP 11.4.1</p>	<p>The risk of vitamin K deficiency is increased in people with CF-related liver disease. Clinicians should base supplementation on the most recent recommendations for vitamin K supplementation in CF ^{78,144}.</p> <ul style="list-style-type: none"> • Routine daily supplementation for all PI patients • Infants: 300 – 1000µg/d • Children (>1year) and adults: 1000 - 10 000 µg/d

Q 11.4.2	What are the requirements for effective supplementation in episodes of vitamin A deficiency in peoples with CF-related liver disease?
R 11.4.2	UNGRADED. Insufficient evidence to make a recommendation – follow recent consensus guidelines. ¹⁴⁴
PP 11.4.2	<p>Supplement with high oral doses between 5000 – 15 000IU/day (1500ug - 4500ug/d) with the aim of achieving the normal range of serum retinol for healthy individuals.</p> <p>Use caution when giving doses above 20 000 IU/d (6000ug/d) preformed vitamin A if RBP is low. A low retinol : RBP molar ratio may indicate deficiency but further increase in supplementation of preformed vitamin A may be toxic to the liver. Although not routinely available we recommend increased supplementation with β-carotene in these circumstances.</p> <p>Monitor serum retinol and retinol binding protein to ensure the adequacy of therapy to prevent deficiency as well as fasting serum retinyl ester concentration to assess for toxicity.</p>

Chapter 12 CF-related Diabetes

No PICO questions were formulated for [Chapter 12](#). This is an area of limited evidence specific to CF, with most international recommendations being formed by expert opinion.

Key Points

- CF-related diabetes shares features of type 1 and type 2 diabetes but is a distinct form of diabetes classified as “other forms of diabetes” or pancreatogenic diabetes. ^{145,146}
- CF-related diabetes is at the end of a spectrum of progressive glucose tolerance abnormalities; it may occur intermittently, and few people with CF demonstrate completely normal glucose tolerance ^{147,148}.
- Annually screen for CF-related diabetes by 2 hour oral glucose tolerance test and commence insulin early.
- Energy, protein and fat intake targets remain the same as for those without CF-related diabetes (see Recommendations 7.1.1, 7.1.2, 7.1.3 and the corresponding practice points, [Chapter 7](#))
- Assess the quantity (eg. grams of carbohydrate consumed at usual meals / snacks / supplements) and quality of carbohydrate intake (the glycaemic index (GI)) of the carbohydrates at meals and snacks.
- The blood glucose levels pre-meal and 2 hours post-meals should be assessed to assist with planning an insulin regimen in conjunction with the Endocrinologist and planning diet modifications.
- For most people with CF-related diabetes in Australia and New Zealand the HbA1c treatment goal is <7% or <53 mmol/mol, individualise goal as required.
- Annually screen for diabetic complications including hypertension, hyperlipidaemia and neurologic assessment.
- From 5 years after CF-related diabetes diagnosis, screen for complications including nephropathy and retinopathy

Chapter 13 Bone Health

Q 13.1.1	How and when should bone mineral content and density be assessed for people with CF?
R 13.1.1	UNGRADED. Insufficient evidence to make a recommendation specific to CF. Health professionals should follow consensus document recommendations for assessing bone mineral density for people with CF. ¹⁴⁹



<p>PP 13.1</p>	<p>2006 Australasian Clinical Practice Guidelines for Nutrition in CF: Assess bone mineral density periodically in people with CF who are more than eight years of age¹. Dual energy X-ray absorptiometry scanning is the current gold standard assessment tool.</p> <p>Follow up: Frequency of follow-up scanning is dependent on previous bone mineral density results, type of treatment that was initiated to improve bone mineral density and the emergency of additional risk factors of bone disease. When clinical status is stable, follow-up scanning should be conducted at least:</p> <ul style="list-style-type: none"> • every three to five years if bone mineral density was normal; Z or T scores > -1 • every two years if bone mineral density was moderately reduced; Z-score between -1 and -2; or T-score between -1 and -2.5, and • annually if bone mineral was severely reduced; Z-score <-2 or T-score <-2.5. <p>More frequent DEXA scanning is suggested if significant new risk factors emerge (e.g. prolonged corticosteroid exposure)</p>
<p>Q 13.1.2</p>	<p>Is there an ideal serum 25-hydroxyvitamin D level to aim for in people with CF?</p>
<p>R 13.1.2</p>	<p>UNGRADED. Insufficient evidence specific to CF for the ideal serum 25-hydroxyvitamin D level. It is suggested that the general Australian and New Zealand goal of ≥50 nmol/L be used with a caveat for the time of year at which testing occurs.</p>
<p>PP 13.1.2</p>	<p>It is suggested that the general Australasian goal of ≥50 nmol/L serum 25(OH)D be used if measuring vitamin D at the end of winter or in early spring. If testing at other times of year, aim for a level 10-20nmol/L higher (≥60-70nmol/L).</p>
<p>Q 13.1.3</p>	<p>What are the calcium requirements in CF to reduce the risk of low bone mineral density?</p>
<p>R 13.1.3</p>	<p>GRADE D. Calcium requirements to reduce the risk of low bone mineral density in CF are unknown. At this time, health professionals should aim for the RDI when making calcium recommendations in CF. ¹⁵⁰</p>
<p>Q 13.1.4</p>	<p>Does supplementing calcium above the RDI improve bone mineral density in CF?</p>
<p>R 13.1.4</p>	<p>GRADE D. There is insufficient evidence to support that routine calcium supplementation above the RDI will improve bone mineral density in CF. Consider calcium supplementation only when dietary intake is unable to meet the RDI. ¹⁵¹</p>
<p>PP 13.1.3 and PP 13.1.4</p>	<p>Foods high in calcium include dairy foods (e.g. cow's milk, cheese & yoghurt), fortified dairy alternatives (i.e. soy milk), firm tofu & bony fish. Legumes, nuts and some green vegetables also contain small amounts of calcium. More information regarding the RDI for calcium can be found via the Nutrient Reference Values for Australia & New Zealand webpage - http://www.nrv.gov.au/</p>

Chapter 14 Special Considerations

Pregnancy

<p>Q 14.1.1</p>	<p>What are the nutrition considerations of the management of pregnancy in CF?</p>
<p>R 14.1.1</p>	<p>UNGRADED. Insufficient evidence to make a recommendation</p>
<p>PP 14.1.1</p>	<ul style="list-style-type: none"> • Before pregnancy a BMI greater or equal to 22kg/m² is recommended. • Undertake a comprehensive nutrition assessment prior to conception and ongoing during pregnancy and post-partum, including standard pregnancy counselling around food safety, alcohol, caffeine and fish consumption recommendations as per Australian ¹⁵² and NZ recommendations ¹⁵³.

	<ul style="list-style-type: none"> • Clinicians should be guided by local country recommendations for supplementation amounts of folic acid. Assess the need for additional supplementation of 5mg folic acid per day in women with risk factors such as family history of neural tube defects, taking certain medications or with insulin dependent diabetes. • Screening for gestational diabetes mellitus is recommended via a 2hr 75g fasting OGTT when pregnancy is confirmed, at 12-16 weeks and 24-28 weeks gestation. Screen for CF-related diabetes at 6-12 weeks post-partum. • Measure levels of fat soluble vitamins A, D and E at first review after pregnancy confirmation and the beginning of the second and third trimesters. Monitor levels and supplement to maintain in the reference range (refer to PP 14.2 for specific information about vitamin A supplementation). • Undertake iron studies at 20 weeks' gestation and assess the need for supplementation if deficiency is developing. Tolerance of supplementation can be problematic in pregnancy further aggravating gastrointestinal symptoms especially constipation. Preventative management strategies including use of stool softening agents can be helpful. • Weight gain of at least 11 kg has been recommended for women with CF. If nutritional status cannot be optimised by a high energy diet alone, explore oral nutrition supplements or enteral nutrition support. In those requiring tube feeding for the first time; it is best commenced early in pregnancy when best tolerated • It is important to discuss infant feeding options during pregnancy with women with CF. Breast feeding in the mother with CF can be successfully undertaken, however close monitoring of nutritional status and fatigue should be undertaken • It is important to monitor the weight of the woman with CF post-partum. Significant weight loss due to breastfeeding, and potential time burden that may compromise self-care can impact on overall health. Optimising nutrition at this time is vital. • For any complex issues in the pregnant woman with CF, consult a CF specialist adult centre.
Q 14.1.2	What recommendations around vitamin A supplementation and monitoring should be provided to women with CF who are pregnant or planning a pregnancy?
R 14.1.2	UNGRADED. Insufficient evidence to make a recommendation
PP 14.1.2	<ul style="list-style-type: none"> • Measure fat soluble vitamin A level at first review after pregnancy confirmation and at the beginning of the second and third trimesters. • If normal vitamin A levels, supplementation should continue at a dose <10,000 IU/day of retinol. These levels are in line with recommendations for the healthy population in pregnancy. • Reassure the woman that supplements are being prescribed to prevent vitamin A deficiency which like vitamin A excess, is also teratogenic. • If vitamin A levels are high, it is recommended to reduce vitamin A supplementation. A different multivitamin supplement may be required with lower vitamin A (particularly preformed vitamin A, retinol). Assess adequacy of other fat soluble vitamins if the CF-specific multivitamin is ceased. • More frequent monitoring of vitamin A levels may be required following changes to supplement formulation and/or dose. • Review dietary intake of vitamin A including oral and enteral supplements with particular attention to high retinol sources. • Review all non-prescription, over the counter supplements with particular consideration for high retinol supplements (e.g. cod liver oil).
Genetic Modulator Therapies	
Q 14.2.1	What are the implications of Ivacaftor on nutritional status in children >2 years and adults with cystic fibrosis who have at least one G551D or other gating mutation allele?
R 14.2.1	GRADE A. There is evidence to suggest that continued use of Ivacaftor therapy leads to significant improvements in weight and BMI in adults and children > 2 years. ¹⁵⁴⁻¹⁶¹



Q 14.2.2	Are there any other nutritional considerations (energy, salt intake) that practitioners should take into consideration for people on Ivacaftor therapy?
R 14.2.2	GRADE D. Well-nourished individuals on Ivacaftor therapy may benefit from a diet more in line with the general healthy population recommendations, although at this stage there is insufficient evidence to recommend routine changes of energy and salt intake for people with CF receiving this medication. ¹⁵⁴⁻¹⁶³
PP 14.2.1 and 14.2.2	<ul style="list-style-type: none"> • Practitioners need to proactively monitor weight gain patterns throughout the first few years of Ivacaftor therapy so that nutritional recommendations can be tailored to the rapidly changing body composition. • People with CF-related diabetes are at risk of experiencing hypoglycemia, especially upon commencement of Ivacaftor – monitor blood glucose levels closely • The relationship between sodium intake and sweat chloride levels is currently unknown. There is not yet clear evidence for a change in salt supplementation requirements, but provide advice on sodium requirements based on the person’s signs and symptoms of salt depletion. • Genetic modulators should be taken with a fat containing meal or snack <ul style="list-style-type: none"> ○ People who are PI should also take their PERT at this time • Children: <ul style="list-style-type: none"> ○ After establishment of Ivacaftor ensure that catch up growth is achieved before considering altering a child’s diet in terms of energy and/or salt. • If the course of CF lung disease is altered (e.g. a reduction in exacerbation frequency), an individual’s nutritional status or dietary intake pattern may also change if overall nutritional requirements are lowered or appetite/intake becomes more stable. A reduction in overall energy intake may not be a concern if adequate nutritional status is maintained, however attention to diet quality may be required.
Q 14.2.3	What is role of gastrointestinal and/or other nutritional outcome measures in individuals with CF receiving Ivacaftor therapy?
R 14.2.3	UNGRADED. Insufficient evidence to make a recommendation
PP 14.2.3	<p>Gastrointestinal outcome measures such as faecal elastase and intestinal fat absorption are appropriate to use in this population group. Use of these tests may help guide practitioners in what are appropriate concurrent nutritional therapies (i.e. PERT and protein pump inhibitors).</p> <p>As genetic modulator therapies are relatively new, it is possible that a range of clinical and symptomatic observations will be made, about which more evidence to guide practice recommendations may emerge in the future in relation to modulation or restoration of physiological functions affected by CFTR. There is limited evidence to date of the impact of other CFTR modulator therapies on gastrointestinal function or nutritional outcome measures. Until such evidence is available, identify and evaluate any changes in gastrointestinal or other symptoms in people taking CFTR modulator therapies.</p>

Chapter 15 Complementary Nutrition Therapies

Probiotics

Q 15.1.1	Does dietary supplementation with probiotic genus <i>Lactobacillus</i> improve nutritional and/or respiratory status in people with CF?
R 15.1.1	GRADE C. Dietary supplementation with a <i>Lactobacillus</i> genus probiotic (single or as part of a mixture) may have health benefits for people with CF, particularly in regards to intestinal inflammation, the intestinal microbiota and risk of pulmonary exacerbation. ¹⁶⁴⁻¹⁷¹
Q 15.1.2	Should routine or targeted use of probiotic supplements be recommended for people with CF?
R 15.1.2	GRADE C. The body of evidence to support health benefits from probiotic supplementation in CF is growing, however there is insufficient high quality evidence to support the <i>routine</i> or <i>targeted</i> supplementation of probiotics in individuals with CF. ¹⁶⁴⁻¹⁷¹

<p>PP 15.1 and 15.1.2</p>	<p>Mechanism of action</p> <ul style="list-style-type: none"> Probiotics are used as a therapeutic option to modulate the composition and actions of the gut microbiota. <p>Probiotic species, strain and dose</p> <ul style="list-style-type: none"> Mechanism of action and efficacy are strain specific. Insufficient evidence to recommend any particular probiotic species or strain, single or mixed, as being superior for beneficial health outcomes in CF. The beneficial dose of probiotics varies depending on the particular species and strain used and the reported benefit – in most cases dose and duration should be based on manufacturer’s recommendation. <p>Other considerations</p> <ul style="list-style-type: none"> Impact on burden of treatment and medication adherence. Cost – probiotics can be expensive and not subsidised in Australia or New Zealand. Concerns regarding reported variability in quality control, efficacy and viability of probiotic microbes in different products. Storage – probiotics are sensitive to temperature, air, light and moisture and often require refrigeration. Probiotics are often marketed via their trade name (e.g. <i>Lactobacillus rhamnosus</i> is marketed as <i>Lactobacillus GG</i>). Recommend that if trialled, probiotics are taken for at least 4 weeks. If after this time they have no impact on symptoms, cease or trial an alternative preparation. Potential health benefits are thought to subside shortly after supplementation is ceased. Use probiotics with caution with high risk patients such as those with severe respiratory function. Care should also be taken administering probiotics to those with venous catheters due to risk of sepsis.
<p>Glutathione</p>	
<p>Q 15.2</p>	<p>Does antioxidant supplementation with oral glutathione or N-acetylcysteine improve nutritional and/or respiratory status in CF?</p>
<p>R 15.2</p>	<p>GRADE C. Dietary supplementation with oral antioxidant glutathione or N-acetylcysteine may improve nutritional status in individuals with CF. There is inconsistent evidence to suggest that dietary supplementation of either of these constituents improves respiratory status. The currently available evidence does not support the use of glutathione therapy in people with CF ^{78,172-176}</p>
<p>PP 15.2</p>	<p>Mechanism of action</p> <ul style="list-style-type: none"> Glutathione is a water-soluble antioxidant. N-acetylcysteine provides the amino acid cysteine (non-essential amino acid) for systemic glutathione replenishment. <p>Sources of supplementation</p> <ul style="list-style-type: none"> Dietary sources of cysteine include meat, dairy, poultry, fish and soy-based products. There is insufficient evidence to support the use of glutathione therapy in CF. <p>Other considerations</p> <ul style="list-style-type: none"> Impact on burden of treatment and medication adherence



Coconut Oil	
Q 15.3	Is there evidence that dietary supplementation with coconut oil improves nutritional status in pancreatic insufficient people with CF?
R 15.3	UNGRADED. Insufficient evidence to support a recommendation.
PP 15.3	<p>Coconut oil composition:</p> <ul style="list-style-type: none"> • Lauric acid (45 - 48%) – medium chain triglyceride (MCT) • Myristic acid (14 - 18%) – long chain triglyceride (LCT) <p>Lauric acid is considered a MCT, however, it is metabolised differently. In digestion, lauric acid behaves more like a long chain fatty acid.</p> <p>Commercially manufactured MCT oils are generally derived from coconut or palm oils and contain approximately 95% MCT.</p>
Herbal Supplements	
Q 15.4	Is there evidence that dietary supplementation with specific herbal products or their components improves health outcomes in individuals with CF?
R 15.4	GRADE D. There is no evidence that dietary supplementation with specific herbal products or their components improve health outcomes in individuals with CF ¹⁷⁷
PP 15.4	<p>People with CF should be encouraged to discuss herbal and complementary therapies with their interdisciplinary CF team prior to commencing any form of supplementation. Specific enquiry by the CF pharmacist or dietitian may be helpful.</p> <p>Limited evidence surrounding dosing, safety or efficacy of most herbal supplements.</p>

Chapter 16 Lung Transplantation

No PICO questions were formulated for [Chapter 16](#). This is an area of limited evidence specific to CF, with most international recommendations being formed by expert opinion.

Key Points

Pre-transplantation

- Care should be overseen by the local CF team with the aim of maintaining nutritional status (i.e. BMI \geq 18.5 kg/m² to 35 kg/m²)
- Once listed for transplantation close liaison between the CF team and the transplant team is essential

Post-transplantation

- Post-transplant care is generally coordinated by the specialised transplant unit.
- Nutritional management in the immediate post-operative period should focus on attaining adequate protein and energy intake ¹⁷⁸
- Post transplantation there is potential for additional nutrition related issues such as bowel management, GOR, delayed gastric emptying, diabetes, lipid abnormalities, bone disease and renal disease
- Medications used to prevent lung transplant complications (e.g. antibiotics, immunosuppressants and anti-fungal medications) can have marked nutrition-related side effects, including taste changes, nausea, vomiting and diarrhoea
- Longer term post transplantation energy and vitamin requirements (i.e. vitamin A and E) are often reduced, thus regular biochemistry and monitoring by a dietitian is essential