Title	Nutrition & Growth
Version	2.0

1. Introduction

Good nutrition is important for optimising life-expectancy in CF. It is strongly linked to improving lung function.

2. Aims

2.1. To achieve normal growth
2.2. Children growing along appropriate centiles for height & weight, BMI 25th – 75TH centile
2.3. Adult BMI: Males 23kg/m^2, BMI Female 22kg/m^2
(Aim for a health BMI avoiding BMI greater than 25kg/m2)

3. Standards

- A Registered Dietitian is available at CF clinics and inpatient admissions
- To ensure nutritional adequacy of dietary intake
- To assess nutritional adequacy annually by a 24-hour recall or 3-day Food Diary as appropriate
- If nutritional status declines, appropriate nutritional management will be advised.
- To provide up-to-date nutritional advice based on evidence-based research.
- In pancreatic insufficient patients, increased doses of Vitamin A, Vitamin E, Vitamin K and Vitamin D should be supplemented according to blood levels.
- Vitamins A, E and D levels should be checked annually, and vitamins titrated with results.
- If vitamins levels are low adherence should be addressed before adjusting dose and levels rechecked.
- If pancreatic insufficient, aim to maintain a dose no greater than 10,000iu lipase/kg/day (unless in certain circumstances where maldigestion cannot be controlled otherwise – Consultant to be aware)

4. Nutritional Requirements

4.1 Energy

This is dependent on the severity of the chest involvement and degree of maldigestion/ malabsorption. If a high fat diet is recommended for nutrition support purposes, mono and polyunsaturated fats should be encouraged.
Energy levels can be 110% to 200% above the normal estimated average requirement. Those on modulator therapy may have more normalised nutritional requirements and therefore healthy eating and weight maintenance of a healthy BMI would be recommended.

4.2 Protein

A high protein intake is advised.

Although there is no specific evidence, an intake 1.5-2 times above reference intake is recommended.

To monitor protein status the serum albumin and urea are good indicators (if the liver is functioning normally). These are done annually when there is no infection present.

4.3 Suggested Daily Energy & Protein Requirements for Cystic Fibrosis

Table 1: Suggested Daily Energy & Protein Requirements for Cystic Fibrosis

Age	**Estimated Average Requirements**		**Increased Energy to 150% Protein x 2**	
Infants protein /req	2.0 –1.5 g protein/kg		4.0 – 3. 0g protein/kg	
Infants kcal /req				
0 - 3 months	120-96 kcal / kg		144-180 kcal / kg	
4 months – 1years	96 - 82/ kg		144 - 123kcal / kg	
Male	Kcal	Protein	Kcal	Protein
1-3years	980	14.5g	1470	29g
4 - 6 years	1480	20g	2220	40g
7 - 10 years	1750	28g	2625	56g
11 - 14 years	2355	42g	3530	84g
15 - 18 years	3010	55g	4515	110g
Adults	2605	55g	3910	110g
Female				
1-3 years	910	14.5g	1365	29g
4 - 6 years	1380	20g	2070	40g
7 - 10 years	1705	28g	2560	56g
11 - 14 years	2165	42g	2800	84g
15 - 18 years	2430	45g	3250	90g
Adults	2080	55g	3120	110g

Adapted from The Scientific Advisory Committee on Nutrition Dietary Reference Values for Energy SACN London 2011 Dietary Reference Values HMSO, 1991 (DoH publication report no 41).

4.4 Fat soluble vitamins (See separate guideline)

4.5 Water soluble vitamins

These are generally met by normal balanced diet and are not supplemented.

4.6 Minerals

4.6.1 Salt supplements

Babies are routinely supplemented with salt solution. Urinary sodium is checked if there are concerns with growth. Children are encouraged to add additional salt to foods and eat foods containing salt.

To be used in hot weather >21°C / 70°F/ vigorous exercise (i.e. causing sweating) below is a rough guide

Age	**Sodium mmol**
<6 mths	2 mmol/kg NaCl solution
6 - 12 months	1 mmol/kg NaCl solution
1 - 5 years	10 mmol/day (2 x 300 mg NaCl tab)
6 - 10 years	20 mmol/day (2 x 600 mg NaCl tab)
>11 years	30 - 40 mmol/day (3-4 x 600 mg NaCl tab)

4.6.2 Fluoride

Discuss with Dentist toothcare and management

4.6.3 Calcium

Needs monitoring due to osteoporosis risk in CF. Recommendations are:

<u>Calcium recommendation for CF prevention of osteoporosis</u>

Children	
1-3 years	500mg/d
4-8 years	800mg/d
9-18	1300mg/d
Adults	
19-50	1000mg/d
>50	1200mg/d

Maximum dose 2500 mg calcium/day

4.6.4 Magnesium

Serum Magnesium is reviewed annually. If hypomagnesaemia is seen supplementation will be assessed.
Higher risk of hypomagnesaemia with frequent aminoglycoside therapy.

4.6.5 Iron

Normal requirements are needed, consider supplements if iron deficiency anaemia is diagnosed, or if symptomatic.

4.6.6 Zinc

Zinc deficiency can cause poor growth and low Vitamin A levels. If concerns with poor growth. Zinc levels will be reviewed. Zinc levels needs to be done in a fasting state.

4.7 Antioxidants: Carotene, Vitamin C, Vitamin E, Carotenoids, Selenium

These are protective to oxidative damage of cell walls. Research in CF suggests there may be benefit in increasing these substances, but optimal doses have not been determined. Vitamin E will be monitored annually to maintain normal levels.

4.8 Polyunsaturated Fats

Role: Essential fatty acids and anti - inflammatory response
This area has little strong evidence for supplementation, as an imbalance of these may be harmful.

4.9 Probiotics

There is currently not enough evidence to use routinely in CF. These can be used safely except when the immune system is supressed.

4.10 Fibre

In normal health, bowels function better with adequate fibre in the diet.
Aim to give the following amounts:

Age Group	Amount of recommended fibre (grams per day)
Children (2-5 years)	15g
Children (5-11 years)	20g
Children (11-16 years)	25g
Adolescents (16-18 years)	30g
Adults	30g

4.11 Fluids

Hydration is very important in CF this will help prevent constipation and DIOS.
A guide for adults and children with cystic fibrosis over 14 years of age is to try and have at least 2000ml/day. Younger children will need less than this between 1000-1700ml. Aim to have at least 6-8 drinks a day.

5. Monitoring and Optimal Growth

In babies head circumference is measured under 12months .
Routine measurements length and weight are monitored for all patients at each clinic visit.
Optimal growth:

Children

- Achieve expected centiles for height and weight
- BMI 25th -75th centile

Adults

- Males BMI 23-25kg/m2
- Female BMI 22-25kg/m2

6. Nutrition and Modulator therapy

Modulator therapy, KAFTRIO, SYMKEVI, ORKAMBI have been seen to impact on patients weight. These medications may have led to an effect on nutritional requirements and therefore patients' weight. It is therefore important to take into account modulator prescriptions, adherence to these and patient weight and trends to tailor the nutritional advice appropriately. It is important that modulator treatments are taken with a fat containing snack and Pancreatic Enzyme Therapy to optimise absorption and effect. Adherence should be rechecked should there be a decline in lung function / weight.

7. Nutritional Support

7.1. If nutritional status declines, nutritional support is needed. Refer to nutritional status in chart below and to the flowing guidance:

	<12 months	1–2 years	2–18 years	>18 years
Normal nutritional status Preventative nutritional counselling	Weight, length and head circumference increasing at expected rates for age	Weight and length increasing at expected rates for age	BMIp 25–75th **or** Weight and length increasing at expected rates for age	BMI 20–25kg/m2
Additional nutritional support Further strategies to enhance energy content of diet +/-use ONS	Correction of nutritional deficits present at diagnosis **or** Weight gain less than expected for age over 2–8 weeks **or** Acute disease related reduction in appetite	Weight loss or no weight gain over 6– 8 weeks **or** Acute disease related reduction in appetite	BMIp <25th **or** Weight loss or no weight gain over 4–6 months **or** Acute disease related reduction in appetite	BMI< 20kg/m2 **or** Weight loss over 4–6 months **or** Acute disease related reduction in appetite
Intensive nutritional support Discuss benefits and use of enteral tube feeding	Sustained deviation from previous weight and/or length percentile	Sustained deviation from previous weight and/or length percentile	Persisting BMIp<25th **or** Sustained deviation from previous weight and/or height percentile	Persisting BMI <20kg/m2 **or** Continuing weight loss
Overweight Consider strategies to reduce rate of weight gain or in significant cases weight reduction		Significantly increased weight velocity or weight over 2 percentile positions above height percentile	BMIp >91st	BMI >25 kg/m2

7.2. Fortify Family Foods

Try additional supplements.
Milk based and fruit-based drinks can be prescribed.
Aim for additional calories of:

- 200 kcal 1-2 years)
- 400 kcal 3-5 years) kcal supplied by
- 600 kcal 6-11 years) supplements
- 800 kcal over 12 years)

Monitor and reassess.

7.3. Behavioural Therapy

Research has found that behavioural therapy works best to enhance weight-gain.
The clinical psychologist should work with the dietitian to improve intake.
Enteral Feeding: if poor weight gain/growth, or BMI < 17, with no improvement over 6 months a discussion on enteral feeding will be arranged with carers/patients/CF team.

7.4. Nasogastric /Gastrostomy /Jejunal feeding are used when nutritional interventions fail to optimise weight and or growth. Normal tube feeding:

- 4 - 12 hours overnight feeding.
- 30-50% of energy requirements given.
- Enzyme dose assessed on fat and given at beginning and during the feed and at the end of feeds.

- Enteral nutrition used:
  - Whole protein feeds 1 – 2.0 kcal/ml either paediatric or adult feed.
  - Semi-Elemental /Elemental feeds can be used if problem with whole protein feed.

8. CF Diabetes, Liver & Bone Disease

8.1. CF Related Diabetes

(See also separate guidelines)

Dietary modification

- Insulin regimen titrated with diet
- Regular carbohydrate at meals and snacks
- Allow sugars with meals and snacks to avoid high blood glucose levels
- Allow high fat use mono & polyunsaturated
- Allow fibre rich foods if indicated
- Avoid sugary drinks and Maxijul supplements
- In severe malnutrition consider simple sugars in all drinks
- Consider carbohydrate counting with a basal bolus regimen

8.2. CF Related Liver Disease

(See also separate guideline)

- Aim for normal growth/BMI.

8.3. CF Bone Disease

(See also separate guideline)

- Review biochemistry and osteomalacia/osteoporosis status.
- Ensure vitamin D and dietary calcium intake are adequate.
- Ensure growth/weight/BMI are within normal range.

9. References

- Nutritional Management of Cystic Fibrosis CF Trust Second edition – September 2016
- Nutrition Guidelines for Cystic Fibrosis Nutrition in Australia and New Zealand 2017 www.cysticfibrosis.org.au
- ESPEN-ESPGHAN-ECFS guidelines on nutrition care for infants, children, and adults with cystic fibrosis, by Turck D, et al., Clinical Nutrition (2016), http://dx.doi.org/10.1016/j.clnu.2016.03.004