CF Guidelines - Nutrition & Oral Supplementation

Oral Nutritional Support in CF:

The aim of nutritional therapy for CF is to promote normal growth and development throughout childhood and on into adulthood.

Nutritional Requirements:

The nutritional requirements of patients with CF vary widely. Requirements are increase by poorly controlled fat absorption and increased energy expenditure. It is recommended that the diet of a CF patient should be 120% - 150% of the estimated average requirement - EAR - for energy. Although there has been limited research about protein requirements, it is generally accepted that protein intake should also be higher then the norm. This is to compensate for excessive los of nitrogen in faeces and sputum and to also ensure that there is adequate level in the system for growth. Fat restrictions are not recommended. Fat provides a useful source of energy and it is suggested that 35% - 45% of energy come from fat sources. A non CF patients healthy diet should be 25% - 30% but 100% of EAR.

Dietary Advice:

Principally, dietary requirements should be met by the patients diet. Patients should be reviewed by a CF dietician regularly to monitor growth and nutritional status. Advice should be on an individual basis. A high calorie and high protein diet should be promoted. If there is weight loss or poor weight gain, further advice should be provided to maximise calorie intake.

Dietary advice may include:

- Food fortification with cream, butter, oil, dried milk, jam and sugar etc.
- Promotion of full fat milk and dairy products.
- Avoidance of low fat food and drink products.
- Promotion of energy dense foods such as crisps, chocolate, cheese etc.
- Nourishing drinks.

Oral Nutritional Supplements:

	Paediatric	Adult
Sip Feeds - Milk-based sip feeds	Paedisure Plus, Fortini, Frebini Energy	Ensure Plus milkshake style, Fortisip, Fresubin Energy, Clinutren 1.5, Ensure Twocal, Fresubin, Resource 2.0
- Juice-based sip feeds	Enlive Plus*, Fortijuice*, Provide Xtra*, Clinutren Fruit*	Enlive Plus*, Fortijuice*, Provide Xtra*, Clinutren Fruit*
High Energy Powdered Milkshakes	Enshake, Scandishake, Calshake	
Pudding Style	Ensure Plus Crème, Forticreme, Clinutren Dessert	
High Energy Infant Milks	SMA High energy, Infatrini, Concentrated standard milks (requires supervision from Dietitian)	

Modules - Carbohydrate	Maxijul*, Polycal*, Caloreen*	
- Fat emulsions	Calogen, Liquigen (MCT fat source)	
- Carbohydrate & Fat mixtures	Duocal	
- Protein	Maxipro Procal Powder Procal shot	

The table above gives some examples of the variety available on prescription.

* Do not require enzymes.
** Check with manufacturers data for suitability of use of supplements with young children.

If a patient fails to meet their requirements through oral diet and are failing to grow and develop, loosing weight or have a low BMI, oral nutritional supplements should be considered. Supplementation with high energy milk based oral supplements has been demonstrated to be an effective way of improving nutritional status and weight of patients with CF.(Skypala et el, 1998) However, a Cochrane review concluded that they did not provide any more benefit in moderately malnourished children with CF than the use of dietary advice and monitoring. Whilst they may be used, they should not be considered as essential. (Smyth & Walters, 2000) There is no conclusion for there short term use for weight gain, or for use in the long term management in adult CF patients or those with advanced lung disease. (Smyth & Walters, 2007) There is an extensive range of supplements available to suit individual age and taste.

Supplements should be taken in addition to the patients normal diet to increase their daily energy intake and should not be used to replace meals. Consideration of timing and quantity is required to ensure appetite and intake of normal food is not reduced. The best way to take them is as snakes between meals, after meals or just before bed time.

Nutritional Intervention:

The table below shows the different stages of nutritional intervention - CF Trust, 2002:

	Age		
	<5 years	**5 - 18 years**	**>18 years**
Normal Nutritional Status	Weight/height 90-110%	Weight/height 90-110%	BMI 19-25 +/or no recent weight loss
Consider Supplements	Weight/height 85-89% Or weight loss over 4 months Or plateau in weight over 6 months	Weight/height 85-89% Or weight loss over 4 months Or plateau in weight over 6 months	BMI< 19 Or 5% weight loss over > 2 months
Growth Failure aggressive nutritional support	Supplements tried and either weight/ height<85% Or weight falling 2 centile positions	Supplements tried and either weight/ height<85% Or weight falling 2 centile positions	Supplements tried and either BMI <19 Or >5% weight loss over>2 months

References:

1, Leeds Teaching Hospital NHS Trust. Cystic Fibrosis in Children and Adults. The Leeds Method of Management, 2008.

2, Royal Brompton & Harefield NHS Trust. Clinical guidelines: Children with CF. http://www.rbht.nhs.uk/healthprofessionals/clinical-depts/paediatrics/childrencf/introduction
3, Skypala I.J, Ashworth F.A, Hodson M.E, Leonard C.H, Knox A, Hiller E.J, Wolfe S.P, Littlewood J.M, Morton A, Conwat S, Patchell C, Weller P, McCarthy H, Redmond A, Dodge J. Oral nutritional supplements to promote weight gain in Cystic Fibrosis patients. Journal of Human Nutrition and Dietetics, 1998: 11: 95-104
4, Smith R, Walters S. Oral calorie supplements for Cystic Fibrosis. The Cochrane Database of Systematic Reviews, 2000.

Document approved - December 2011
Document due for review - December 2013

Acknowledgements: *The Peninsula CF team acknowledges the use of guidelines produced by The CF Trust, Manchester, Papworth, Leeds and Brompton CF teams during development of these local Peninsula protocols and guidelines.*