Guideline for the Dietetic management of Cystic Fibrosis in newly diagnosed Cystic Fibrosis babies.	University Hospitals of Leicester NHS NHS Trust **Trust Reference Number C51/2023**

1. Introduction and Who Guideline applies to

This policy sets out the UHL Trust Policy and procedures for a newly diagnosed baby with Cystic Fibrosis from a dietetic point of view.

These guidelines have been developed to ensure that dietetic advice given to patients with Cystic Fibrosis and their families is consistent and follows current scientific evidence. This guideline has been written to reflect the current UHL practice in addition to the evidence base, including 'European Cystic Fibrosis Society Standards of Care: Framework for the Cystic Fibrosis Centre' and 'European Cystic Fibrosis Society Standards of Care: Best Practice guidelines'.

Cystic Fibrosis is a genetic disorder which impacts 1 in 2500 births in the UK. In the UK newborn screening (NBS) has been available for all babies since 2007, the heel prick test screens for common mutations of Cystic Fibrosis. However some of the rarer mutations may not be picked up by NBS. Appropriate nutrition is essential for the long term health of a child with CF. 85% of the patients with Cystic Fibrosis are pancreatic insufficient and therefore require pancreatic enzyme replacement therapy (PERT) in addition to fat soluble vitamins and sodium supplementation.

The guidelines are intended for use by Senior Specialist Dietitians (Paediatric) and Senior Dietitians (Paediatric) within the Dietetic and Nutrition Service, University Hospitals of Leicester NHS Trust. They may be used as a point of reference for other Health Care Professionals involved in the care of CF, but detailed assessment and advice should only be provided by a Senior Specialist Dietitian or Senior Dietitian.

2. Guideline Standards and Procedures

1. Aims of Dietetic Treatment

The aim of the dietetic treatment is to assess the nutritional status of the patient. The dietitian should consider their current symptoms, weight and feeding to make an assessment and a plan to support appropriate growth for the baby.

2. Dietetic Management of newly diagnosed patients

The dietitian will be contacted by the CF specialist nurse if a new baby is highly suspected or has a diagnosis of CF.

Usually baby's with CF will have been picked up via newborn screening, however they can present with meconium ileus (MI) which can be detected antenatally. MI causes intestinal obstruction, this occurs in up to 20% of newborns with CF. Meconium Ileus is usually associated with pancreatic insufficiency (Nutritional management of CF., 2016).

85-90% of patients with CF are pancreatic insufficient (Nutritional management of CF., 2016). This means they require Pancreatic enzyme replacement therapy to digest and absorb the fat and protein within food. Without PERT this can lead to malabsorption. The main aim of PERT is to; promote growth and development, control symptoms of malabsorption, achieve sufficient fat soluble vitamin levels and achieve appropriate bowel movements. Within UHL pancreatic insufficiency is detected via a faecal elastase (FE1) test.

Table 1: Defining exocrine pancreatic insufficiency as per faecal elastase results

Pancreatic Function	Faecal Elastase (FE1) microgram µg E1/g
Normal	>200 µg E1/g
Mild PEI	100-200 µg E1/g
Severe PEI	<100 µg E1/g
CF PEI	<15 µg E1/g

Normal FE1 levels are expected by day 3 in term infants and by 2 weeks of age in those born before 28 weeks gestation. Tests should be performed after this time. It should be noted that watery samples may not be accurate and may need to be re-checked. In addition if the sample is too small then it may not be able to be run.

It can take between 2-3 weeks for faecal elastase results to come back, therefore in some cases if the patient has symptoms of malabsorption and or has a CF genotype associated with pancreatic insufficiency, PERT can be started. However if should be stopped if faecal elastase is found to be normal.

For Infants born with Meconium Ileus PERT will be started post-surgical interventions.

Indications of pancreatic insufficiency:

- Genetics
- Large feeding volumes (above 200ml/kg/day)
- Poor weight gain
- Frequent large/ smelly/greasy stools (steatorrhea)
- Bloating or abdominal discomfort

Initiation of pancreatic insufficiency

Oral feeding

Creon Micro is the usual PERT for babies used within UHL. Creon micro provides 5000IU/lipase per scoop. The max dose is 10,000 IU/kg/day or 2500 IU/kg/feed.

1 x Scoop of Creon Micro = 5000iu = 2.5-5g of fat. Fat content of standard formula/ breast milk is ~3.5g/100mls.

Table 2: A guide for appropriate Creon starting dose in newly diagnosed CF babies.

Volume of milk	Creon micro for Breast milk/ standard formula	Creon for High energy formula
<20mls	Nil	Nil
20-40mls	¼ scoop	½ scoop
41-70mls	½ scoop	¾ scoop
70-100	¾ scoop	1 scoop
101-140mls	1 scoop	1 ¼ scoop

Creon doses are individual and the table above is a guide. Creon will be adjusted by a Senior Specialist Dietitian based on symptoms, feed volumes and weight gain.

When introducing Creon do not start with the full dose. Initially start with a few granules then increase by a ¼ scoop per feed until reaching desired dose or until symptoms of malabsorption improve.

Because babies feed frequently and their diet is high fat, they are unlikely to exceed maximum doses per feed but often need to exceed the 10,000IU/kg/day. Where this is necessary, regular review is required to ensure clinical improvement in symptoms. If an infant's dose needs to exceed 10,000IU/kg/day then Omeprazole should be used to optimise absorption (Proesmans, M., & De Boeck, K., 2003). Creon micro should be given on a pea size amount of apple puree (other fruits are not the correct pH). Sprinkle the Creon granules on top so they stick. Creon should be given immediately before feeds. Parents should check the child's gums and face for residual Creon as it can cause damage. If a child takes a long time to feed, the Creon dose can be split. Creon works best within the first 20-30 minutes after administration (Creon pancreatin., 2023). For breast feeding mothers, ensure no granules are on the skin.

It is common for the dose of Creon to change based on symptoms and growth.

Signs of malabsorption if the dose of Creon is not sufficient:

- Poor weight gain/ dropping centiles
- Feed volume increased
- Foul smelling stools or wind
- Increased frequency of stools
- Change in bowel habits

Signs of malabsorption resolving:

- Feed volumes reducing
- Stools becoming more formed
- Stools becoming less greasy and smelly
- Weight gain improving

It is unusual to see signs of giving too much Creon, however consider this if symptoms such as…

- Sore bottom (barrier creams can be used)
- Constipation (can be due to other causes)
- Granules in the stools (may suggest a quick transit time)

Enteral feeding

If the patient is nil by mouth (NBM) then the PERT should be given via the enteral tube. Powdered PERT such as Pancrex V powder should be used.

If giving bolus feeds, PERT should be administered within 30 minutes of giving the feed. If the bolus will take longer than 30 minutes then give PERT during the feed.

Giving PERT as flushes: mix 1 g scoop pancreatin powder (Pancrex V Powder, Essential Pharmaceuticals, UK) with 50 mL sterile water. Shake well and immediately flush via a feeding tube. Do not give with other medication. Do not flush between the feed and the enzyme as this will reduce the mixing of the feed with the PERT. Administer every 2 hours hours throughout enteral feeding, increase dose of PERT if needed.

Table 3: The different enzyme preparations available and the enzyme units per capsule.

Enzyme preparations	Lipase (Units per capsule)	Amylase (Units per capsule)	Protease (Units per capsule)
*Creon Micro®	5000	3600	200
Pancrex V powder®	25,000 in 1g of powder	30,000 in 1g of powder	1400 in 1g of powder

Those denoted with a * are the most commonly used PERT in UHL

Follow up of babies started on Creon

After the initial consultation when Creon has been started, the Dietitian will have regular contact with the families. It would be usual for the dietitian to contact daily initially, once the family are more settled this will progress to weekly contact. Going forward the dietitian will see the baby at least once a month during the first year of life. The dietitian will be monitoring weight, feed volumes, stools, creon dosing and gastrointestinal symptoms.

Salt

Children with Cystic Fibrosis lose more salt in their sweat (Turck et al., 2016). Therefore they need to be supplemented. A lack of salt can impact growth, muscle weakness and cramps and impacts of moods (Leonard et al., 2015). Salt is prescribed by the medical team.

In babies salt should be given throughout the year, whereas in children over one salt is usually only given in the warmer months (Children's Hospital Cystic Fibrosis ORAL Drug Formulary., 2022).

<u>0-2 years</u>

Salt 2mmol/kg/day as bd dose

(30% NaCL contains 5mmol sodium per ml)

Vitamins

Children who are pancreatic insufficient do not have the enzymes to digest and absorb fat from their diet. Therefore there may be some risk of fat soluble vitamin deficiencies. Therefore all children who are pancreatic insufficient should be started on fat soluble vitamins. The medical team will prescribe them.

Table 4 – Doses of Paravit CF required for different ages in children with CF who are pancreatic insufficient (Children's Hospital Cystic Fibrosis ORAL Drug Formulary., 2022)**.**

Age	Dose
Birth-1 year	0.1ml OD

<u>3. Education and Training</u>

Senior Specialist Paediatric Dietitians/ Senior Paediatric Dietitians with appropriate training.

To adjust Creon doses Extended scope training should have been completed by the Senior Specialist Dietitian.

4. Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Nutritional status of CF infants	Full nutritional assessment including assessing growth over the past 12 months.	Hannah Harding	Annually	Annual review reports

5. Supporting References (maximum of 3)

Cystic Fibrosis Trust. 2016. Nutritional management of Cystic Fibrosis. [online] Date accessed 25.05.23. Available from: Nutritional Management of cystic fibrosis Sep 16.pdf

Turck D., Braegger C.P., Colombo C., Declercq D., Morton A., Pancheva R., Robberecht E., Stern M., Strandvik B., Wolfe S., et al. ESPEN-ESPGHAN-ECFS guidelines on nutrition care for infants, children, and adults with cystic fibrosis. Clin. Nutr. 2016;35:557–577. doi: 10.1016/j.clnu.2016.03.004.

Leonard, A. Cystic Fibrosis Nutrition: Outcomes, Treatment Guidelines, and Risk Classification. In: Diet and Exercise in Cystic Fibrosis. San Diego, CA: Academic Press; 2015:27-34.

6. Key Words

Cystic Fibrosis

PERT- Pancreatic enzyme replacement therapy

CONTACT AND REVIEW DETAILS	
Guideline Lead (Name and Title)	**Executive Lead**
Details of Changes made during review:	