Dietetic Management of Infants Diagnosed With Cystic Fibrosis

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Background & Introduction

This protocol has been devised on behalf of the Scottish Paediatric Cystic Fibrosis MCN for use by dietitians and other health professionals working in cystic fibrosis (CF) care across Scotland. It is based on current evidence and best practice. It should be used in conjunction with the published CF Trust guidelines on Standards of Care and Nutritional Management of CF.

Paying close attention to nutrition in the first year of life is essential for infants diagnosed with CF. This is due to the rapid growth seen during this time and the benefits associated with good nutritional status and pulmonary outcomes (Borowitz et al, 2009).

Summary of Main Points

• An infant newly diagnosed with CF should see a specialist paediatric dietitian within two weeks of diagnosis.

• Treatment should be individualised depending on genotype, family circumstances and the clinical condition of the infant.

• The dietitian will advise on infant feeding, growth, pancreatic enzyme replacement therapy (PERT) and fat soluble vitamin supplements.

• Close attention should be paid to growth to ensure adequate weight gain in the first year of life.

• Regular follow up will be required, suggest minimum monthly review until six months old.

Aims of Dietetic Input

• To achieve normal growth and nutritional status.

• To provide optimal PERT to achieve maximum nutrient absorption.
Family Education

Once the diagnosis of CF is confirmed, the family are seen by CF medical staff and the CF nurse specialist to explain the diagnosis. The dietitian and the physiotherapist will usually see the family on the first or second visit to start therapy.

Education should include:
- Role of pancreas in digestion
- Importance of nutrition and growth
- Enzyme and vitamin therapy if required
- High energy, high protein diet

Please see attached dietetic check sheet for seeing a family of an infant newly diagnosed with CF (appendix 1).

Choice of Feed for Screened Infants

Nutritional requirements for infants with CF will vary depending on genotype, presentation and control of malabsorption. Requirements are estimated between 100-150% EAR (Australasian Clinical Practice Guidelines, 2009., Bines et al, 2002).

Breast Milk
Breast feeding should be encouraged due to the well documented benefits of breast milk. Breast milk also contains some lipase which will aid digestion. Creon Micro can be given from a spoon mixed with expressed breast milk, water or fruit puree and given as described below.

A mother who decides to breast feed will need lots of encouragement and support to ensure adequate PERT dosing for growth and confidence in breast feeding.

Standard Infant Formula
If breast milk is not available, the choice of infant formula will be a parental decision, but parents should be encouraged to choose a whey based formula and continue with this through the first year of life.

Infants should be fed on demand; however it is not unusual for infants with CF to consume between 150 – 200mls/kg/day (Shaw & Lawson, 2007). This will usually be for catch up growth and once adequate PERT is established, feed volumes may reduce.

Specialised Formula
High energy formulas such as Infatrini (Nutricia), Similac High Energy (Abbott) or SMA High Energy (SMA) may be considered for those who gain weight slowly. However, infants with CF will often consume large volumes of milk (150–200mls/kg) and therefore close attention to potassium and protein intakes will be required. See later section on slow weight gain.
Choice of Feed for Infants with Meconium Ileus

An infant born with meconium ileus will be transferred to the nearest neonatal surgical unit. Treatment required will vary greatly. The infant may require significant gut resection or conservative management only may be appropriate. In either case the infant may take some time to progress on to enteral feeds and a specialised formula may be required.

Breast Milk
Infants who have had surgery for meconium ileus may be able to have breast milk but it may need to be expressed and given via a naso-gastric tube.

Specialised Formula
If breast milk is not available, a protein hydrosylate should be used due to possible temporary lactose intolerance and/or liver damage. Examples would be Pepti junior (Cow & Gate) or Pregestimil (Mead Johnson). The length of time for which they remain on a hydrolysed formula will depend on weight gain, the presence of a stoma and when the stoma is going to be closed. The infant can be regraded on to breast milk/standard formula when appropriate and in discussion with surgical colleagues.

Pancreatic Enzyme Replacement Therapy

Stool samples will be required to test for pancreatic sufficiency. Stool samples should be fresh or frozen and sent to biochemistry to be tested for chymotrypsin or faecal elastase levels.

If the infant is found to be pancreatic insufficient, PERT should be started as soon as possible. If the infant is on low enteral feed volumes post gut resection, it would usually be started when the infant is on about 80mls/kg/day of enteral feed (i.e. half of their normal requirements met enterally).

In some geographical areas the stool results may take some time to come back which may result in growth failure while waiting. For some infants, for example likely insufficient due to genotype, presentation, frequent oily stools, it may be appropriate to start PERT once samples have been collected but before results are available.

Creon Micro is the enzyme preparation of choice for infants. The initial Creon Micro dose will be ¼ to ½ scoop of Creon Micro per feed. The dose may be gradually increased depending on stool output, growth and fat microscopy if available. Pancrex V powder may be used for those who are tube fed and those who have a poor swallow. It may also be considered for preterm infants.

Creon Micro should be given from a spoon and mixed with either expressed breast milk or infant formula, water or fruit puree and given immediately prior to feeds.
If an infant is on continuous feeds, Creon Micro should be given 2-3 hourly. Creon Micro should never be added to feeds or put down a nasogastric tube, as it will block the tube. It should not be crushed or added to warm milk, as this will denature the enzymes.

Please refer to separate protocol on PERT for more details.

**Vitamin Therapy**

When the infant is known to be pancreatic insufficient, fat soluble vitamins can be commenced. Please refer to the separate vitamin supplements protocol for recommendations. Preferred products are Ketovite liquid and Ketovite tablets, Dalivit or Abidec. Consideration should be given to the higher vitamin A content of Dalivit. Vitamin E is usually provided as a liquid suspension.

**Slow Weight Gain**

Nutritional requirements for infants with CF will vary depending on genotype, presentation and control of malabsorption. Requirements are estimated between 100-150% EAR (Australasian Clinical Practice Guidelines, 2009., Bines et al, 2002). Satisfactory weight gain with careful plotting on an appropriate growth chart (UK-WHO 0-4years) is the best determinant of an adequate energy intake.

If stools are described as loose, oily and frequent and weight gain is not satisfactory,

- Reassess the parent’s technique for administering enzymes.
- Check the enzyme dose given is as prescribed.
- Check the enzymes are not being added to warm milk.
- Check enzymes have been stored correctly and remain in date.
- Stools may be sent for fat microscopy if available to determine if sufficient PERT is being given to control malabsorption.
- If the baby is on a standard infant formula, stools may be sent for reducing substances to assess carbohydrate tolerance.

Some infants may benefit from the use of H₂ antagonists such as Ranitidine or a proton pump inhibitor such as Omeprazole. These products reduce gastric acid levels and therefore optimise artificial pancreatic enzyme function. This may be useful if the infant is on a large dose of enzymes but fat malabsorption is still not controlled. Introduction of these medicines should always be discussed with medical staff first.

Frequent stools can be common in infants who have had surgery for meconium ileus and have previously had a stoma. Enzyme doses should not be increased without evidence of fat malabsorption and discussion with the CF medical staff.
Sodium depletion may result in slow weight gain. Infants with stomas are more at risk from sodium depletion. Breast milk and standard infant formula may not provide enough sodium for the infant with CF (Laughlin et al, 1981). The best measure of sodium depletion is in the urine. A sample of urine can be sent to biochemistry for measure of urinary electrolytes. Refer to individual unit's policy for more information, example policies are listed in appendix 2.

If weight gain remains poor, despite adequate enzyme therapy, good feed volumes, normal biochemistry and adequate sodium levels, then manipulation of feed may be required. If the baby is breast fed, breast milk fortifiers may be useful or a high energy infant formula can be given as one feed per day.

If formula fed, consider concentrating the infant formula, for example up to 15g powder per 100mls or using one of the high energy formulas. However, infants with CF will often consume large volumes of milk (150–200mls/kg) and therefore close attention to potassium and protein intakes will be required.

**Weaning**

Infants with CF should be weaned as per current recommendations for all infants, which is that weaning should begin by 6 months of age and not before 17 weeks of age. Each infant should be managed individually. Families should be advised on standard weaning practices but with an emphasis on higher calorie options, if appropriate. Close attention to PERT dosing will be required during weaning.

Initial weaning foods will not require PERT. However once significant amounts of fat and protein containing foods are introduced, PERT will be required. Referring to the infant’s current PERT dose for milk feeds will guide the dose for solids. Amounts of solids are usually very small, ½ scoop creon Micro may be enough initially. Families should be advised on varying the dose depending on the fat content of foods. Significant dietetic support is usually required during the weaning process. Families require reassurance and guidance on getting PERT doses right along with adequate intake and good growth.

Refer to the PERT protocol for more information.

**Follow Up**

Parents should be seen by the dietitian frequently to provide education on the administration of enzymes, vitamin therapy and the importance of nutrition and weight gain in CF.

Parents should be given written information, for example the CF Trust Factsheet, “Eating well with Cystic Fibrosis – A guide for feeding infants”. A contact telephone number for the Dietitian should be provided. Screened infants should be reviewed 1 – 2 weekly by the dietitian to ensure expected
growth is achieved. Once weight gain is established this can reduce to monthly.

If the infant has been an inpatient after surgery for MI and is discharged on special feeds or supplements added to their feeds, they should be given written instructions on making up the feeds from the dietitian. A letter for ACBS prescribable items should be sent to the GP. The dietitian will usually need to see these infants more regularly for weight checks and monitoring compared to other team members. In these cases, home visits may be appropriate. If the child is brought up to hospital to see the dietitian only, strict adherence to the CF unit’s segregation policy is required.
References


Further Reading


Resources

Appendix 1  
Dietetic Check Sheet for seeing Newly Diagnosed Infant with CF

Info from Medical Notes:

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<th>Genotype</th>
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<tr>
<td>Presentation</td>
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<td>Gestation and D.O.B</td>
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From Parents:

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<td>Stools - frequency</td>
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<tr>
<td>Stools – any samples sent/? need stool collection kits for home</td>
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Info to give:

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<td>Why is Nutrition Important?</td>
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<td>PERT – why?</td>
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<td>PERT – how?</td>
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<td>Vitamins</td>
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<td>Sodium</td>
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<td>Need for high energy/high protein diet</td>
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Appendix 2 – Examples of Sodium Supplementation Protocols.

Sodium Supplementation for Infants with CF.
Yorkhill Children’s Hospital, Glasgow.

BACKGROUND

Sodium is an important growth factor which stimulates cell proliferation & protein synthesis. Reported salt loss causes growth failure with subsequent salt repletion improving growth (Haycock, 1993).

Infants with CF lose large amounts of sodium in their sweat. Breast milk and standard infant formulas do not meet the increased sodium requirements for infants with CF (Borowitz et al, 2009). Also for the older infant, baby foods contain no added salt putting them at risk of inadequate sodium intake.

Serum sodium levels do not accurately reflect total body sodium as when hyponatraemia is present this is significant depletion (Borowitz et al, 2009). Urinary levels are more sensitive as they will fall before serum levels drop.

Low urinary sodium and high urinary potassium has been described in CF patients with sodium chloride deficiency (Ozelik et al, 1994) and in preterm infants with poor weight gain (Vanpee et al, 1995). Normal urine sodium to urine potassium ratio in children is generally reported as 2 (Satlin & Schwartz, 1992).

Haycock (1993) describes a urinary sodium of <10mmol/L in 6 infants who failed to grow. Normal growth occurred in each case when urinary sodium concentration rose to >10mmol/L.

This protocol is based on the small amount of evidence available in the literature and historical practice and experience in the neonatal and gastroenterology units at Yorkhill.

PROTOCOL

Urinary electrolytes should be measured using spot urine analysis in

• all infants with a stoma following a gut resection for meconium ileus.
• all infants diagnosed via newborn screening once established on feeding.

Low result classed as

• Urinary sodium <10mmol/L and/or
• Sodium:potassium ratio <2:1

Supplementation should be given in divided doses using 30% NaCl solution, providing 5mmols Na/1ml. Solution can be added to bottle feeds or given mixed with a little EBM via syringe/spoon.
Starting dose is 2mmols Na per Kg body weight.

Urinary electrolytes should be rechecked at next review and an increase in dose may be required if levels remain low. Aiming for sodium levels above 20mmols/L and sodium to potassium ratio of 2:1.

Ideally urinary electrolytes should be checked at each review and adjustments made as infants gain weight or at least if weight gain begins to slow.

Sodium supplements should continue until 1 year of age. Supplements can then stop if diet is adequate, weight gain is good and a urine sample confirms adequate sodium levels.

**Sodium supplements should be temporarily stopped if infant has diarrhoea and vomiting.

References


Devised by CF Dietitians, Yorkhill Children’s Hospital, Glasgow.
CF Infants Salt Supplement Policy

Summary:
Check urinary sodium and urinary creatinine at each clinic visit
Calculate ratio:  \[
\text{Urinary sodium (mmol/l) / Urinary creatinine (mmol/l)}
\]
Aiming for Ur Na:Cr ratio between 17 – 52

Sodium supplement to be prescribed: 1 mmol / ml sodium chloride solution
Starting dose: 1-2 mmol Na / kg body weight / day

Bottle fed babies – add NaCl solution to feeds
Breast fed babies – give as medication mixed with small amount of EBM or formula milk in 3 – 4 doses per day (baby medicine dispensers can be useful). Can be added to solids once taking weanings.

Parents to be provided with information sheet advising of sodium requirement, available from dietitian. (See below)

Frequency of monitoring: Monthly at each clinic visit

Dietitian will usually review results and advise parents.
Confirmation of advice given to be emailed to CF nurses and CF secretary for copy to be filed in patient notes.

Adjustment of doses on review:
As a baseline, if ratio is low, increase Na supplement by around 1 mmol/kg body weight / day
If ratio is high, reduce by around 1 mmol /kg
If urine is very dilute (ie. urinary creatinine less than 0.5), repeat sample in following week.

Additional information:
Most infants require 1-2mmol sodium / kg/day but some may require more, particularly in first 6 months of age.
The amount of sodium supplement added per feed needs to be increased regularly in line with increases in body weight. Also need to take into account if baby is taking larger but less frequent feeds.
If results are unusual, check dilution of NaCl solution being used at home, consider if gaviscon is being used, check how NaCl is being added at home (for example, parents may be making up 180 ml feeds but baby is mainly taking 100 ml feeds)
If baby is unwell with diarrhoea or vomiting, NaCl supplements should be stopped until baby is rehydrated and condition improved.

Newly diagnosed infants
Check urine results a few days / 1 week after starting enzymes
Pancreatic sufficient babies may still require sodium supplements.
Information included in parental advice sheet:

- NaCl dose to be added
- Emphasis to always double check dilution of NaCl solution when opening new bottle
- NaCl solution should be measured accurately with syringe
- If baby develops diarrhoea and vomiting, NaCl supps should be stopped until baby rehydrated and condition improved.
- There may be a delay in their pharmacist obtaining supply of NaCl solution, (pharmacy give sheet on supplier to parents to give to their pharmacy) parents need to plan ahead

Stopping NaCl Supps
NaCl supps are usually stopped at 12 months of age. Usual advice for CF children regarding generous use of salt in diet should be followed, particularly in hot weather.

Check urine sample for urinary Na and creatinine 1 month after stopping Na supps.

Reference:

Policy compiled by Alison Coates, Clinical Specialist Dietitian for CF 9th October 2009