

M C A D D

MEDIUM CHAIN ACYL CoA DEHYDROGENASE DEFICIENCY

Dietary management guidelines for dietitians

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Adobe Acrobat Reader downloadable PDF file of this document available from:

www.bimdg.org.uk or www.newbornbloodspot.screening.nhs.uk

(for links to other downloadable files)

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MCADD dietary management guidelines

These practical guidelines are designed for dietitians and doctors in the dietary management of patients with Medium chain acyl-CoA Dehydrogenase Deficiency (MCADD). They form a series of suggested dietary management guidelines for use at specified ages, from a positive newborn screening result throughout childhood.

The dietary guidelines have been compiled in conjunction with clinician and dietitian members of the British Inherited Metabolic Disease Group (BIMDG) with the aim of giving the best advice using the limited evidence available.

The dietary treatment is twofold: when well there is no specific dietary management apart from avoiding prolonged fasting; during illness an emergency regimen of very frequent glucose polymer drinks is implemented.

It was considered important to set 'safe fasting times' for well infants despite unscreened cases of MCADD rarely presenting in the first few months of life, even though they will have been demand fed with no restrictions on time between feeds.

The times are based on the few single patient reports of controlled fasting studies in infants and the feeding practices that have proven successful in avoiding symptomatic episodes in screened populations elsewhere in the world. The times given are maximum recommended fasting times, although the true fasting tolerance is likely to be longer.

**No liability can be taken as a result of using this information.
New evidence at any time could invalidate these recommendations.**

How to use the guidelines

The recommended frequency of dietetic reviews are based on the child's age. There are age appropriate, dietary management guidelines for all such reviews.

Guidelines (PDF file) for dietitian and patient use can be downloaded as needed from:

www.bimdg.org.uk or www.newbornbloodspot.screening.nhs.uk
(for links to other downloadable files).

The dietary guidelines are in two complementary parts:

1. Guidelines for dietitians

Each guideline states the child's review age and is colour coded for this.

Each guideline is divided into three sections :

- a checklist of important questions to ask parents/carers about usual feeding practices and the emergency regimen
- information sheets to give parents/carers and copies to professionals (refer to point 2 below)
- guideline on maximum 'safe fasting time' for the well child.

Each item has a tickbox to be used as a completion checklist.

2. Information sheets for parents/carers

These are divided into sections according to the child's review age and colour coded as per dietitians' guidelines, with age appropriate information sheets to give to parents/carers as listed.

M C A D D

MEDIUM CHAIN ACYL CoA DEHYDROGENASE DEFICIENCY

Guidelines for dietitians

Around 2 weeks old

1. Face to face review

Within 24 to 48 hours of presumptive positive screening result

Checklist of important questions

- method of feeding (breast or bottle)
- if bottle fed, check feed does not contain added MCT
- usual feeding frequency and maximum overnight fasting time
- weight and length

Information sheets to give parents/carers and copies to professionals

- ① Contact names, telephone and bleep numbers – to be completed by dietitian
- ② Infant feeding: age under 1 year
- ③ Emergency Regimen: for infants age under 1 year
- ④ Emergency Regimen recipes: for infants age under 1 year – 10% Carbohydrate (if necessary demonstrate making of Emergency Regimen feed)
- GP prescription letter for glucose polymer (not provided in guidelines)
- copies of all information sheets to local dietitian, consultant, GP and HV
- give supply of glucose polymer and scoops

Guideline on maximum 'safe fasting time' for well infant

- from presumptive positive screening result to 4 months of age = 6 hours

2. Face to face or telephone review

2-7 days after presumptive positive screening review

(This review is mainly medical to discuss biochemical test results for MCADD. A dietetic review is optional but may be beneficial for some families to reinforce the initial dietary advice given.)

Checklist of important questions

- if feeding well
- usual feeding frequency and maximum overnight fasting time
- understanding of Emergency Regimen instructions
- weight

4 to 6 months old

Face to face review

Checklist of important questions

- method of feeding (breast or bottle)
- if weaning commenced
- weight and length
- usual feeding frequency and maximum overnight fasting time
- if needs to be wakened for night feed
- if used Emergency Regimen and if any problems
- understanding of Emergency Regimen instructions
- supplies of glucose polymer and scoops
- parents to ensure glucose polymer is 'in date' and open tins to be discarded according to manufacturers' instructions

Information sheets to give parents/carers and copies to professionals

- ① Contact names, telephone and bleep numbers – to be completed by dietitian
- ② Weaning diet
- copy of weaning diet information sheet to local dietitian, consultant, GP and HV

Guideline on maximum 'safe fasting time' for well infant

- from 4 to 8 months of age = 8 hours

8 months old

Face to face or telephone review

Checklist of important questions

- method of feeding (breast, bottle, solids)
- usual feeding frequency and maximum overnight fasting time
- weight and length
- weaning is progressing and starchy foods being given
- if used Emergency Regimen and if any problems
(if never used Emergency Regimen suggest make a feed for baby to try)
- understanding of Emergency Regimen instructions
- supplies of glucose polymer and scoops
- parents to check glucose polymer is 'in date' and open tins to be discarded according to manufacturers' instructions
- confirm has up-to-date contact sheet

Guideline on maximum 'safe fasting time' for well infant

- from 8 to 12 months of age = 10 hours

1 year old

Face to face review

Checklist of important questions

- weight and height
- diet history, any feeding problems
- usual feeding frequency and maximum overnight fasting time
- can change to cows milk instead of infant formula, if appropriate
- if used Emergency Regimen and if any problems
- understanding of Emergency Regimen instructions
- supplies of glucose polymer and scoops
- parents to check glucose polymer is 'in date' and open tins to be discarded according to manufacturers' instructions

Information sheets to give parents/carers and copies to professionals

- ❶ Contact names, telephone and bleep numbers – to be completed by dietitian
- ❷ Children's diet: from 1 year
- ❸ Emergency Regimen: for children age from 1 year
- ❹ Emergency Regimen recipes: for children age 1 to 2 years – 15% Carbohydrate
- ❺ Alternative Emergency Regimen drinks: for children age 1 to 2 years – 15% Carbohydrate (show samples of commercial drinks)
- copies of all information sheets to local dietitian, consultant, GP and HV

Guideline on maximum 'safe fasting time' for well child

- from 12 months of age onwards = 12 hours

2 years old

Face to face review

Checklist of important questions

- weight and height
- diet history, any feeding problems
- usual feeding frequency and maximum overnight fasting time
- if used Emergency Regimen and if any problems
- understanding of Emergency Regimen instructions
- supplies of glucose polymer and scoops
- parents to check glucose polymer is 'in date' and open tins to be discarded according to manufacturers' instructions

Information sheets to give parents/carers and copies to professionals

- ① Contact names, telephone and bleep numbers – to be completed by dietitian
- ② Emergency Regimen recipes: for children over 2 years – 20% Carbohydrate
- ③ Alternative Emergency Regimen drinks: for children over 2 years – 20% Carbohydrate (shows samples of commercial drinks)
- copies of all information sheets to local dietitian, consultant and GP

Guideline on maximum 'safe fasting time' for well child

- from 12 months of age onwards = 12 hours

3-9 years old

Face to face review

A yearly dietetic review is recommended for this age group.

Checklist of important questions

- weight and height
- diet history, any feeding problems
- usual feeding frequency and maximum overnight fasting time
- if used Emergency Regimen and if any problems
- understanding of Emergency Regimen instructions
- supplies of glucose polymer and scoops
- parents to check glucose polymer is 'in date' and open tins to be discarded according to manufacturers' instructions

Information sheets to give parents/carers and copies to professionals

There are no new information sheets for this age group. The information sheets given at age 1 year (number 2 and 3 only) and all at age 2 years are appropriate for this age group, as needed.

- check parents/carers have copies of all relevant information sheets.

Guideline on 'maximum safe fasting time' for well child

- 12 hours provided child is well and eating normally

10 years old

Face to face review

Older children and teenagers should continue to eat regularly and avoid prolonged overnight fasts, by having a starchy bedtime snack and not to miss or have a late breakfast.

It is very important advice about alcohol is given. They need to be informed that drinking large amounts of alcohol can be dangerous, its intake needs to be limited and must always be taken in combination with food.

If a teenager is vomiting due to drinking too much alcohol they must go to hospital immediately.

Wearing a Medic Alert bracelet may be useful for some teenagers.

Checklist of important questions

- weight and height
- diet history, any feeding problems
- usual feeding frequency and maximum overnight fasting time
- if used Emergency Regimen and if any problems
- understanding of Emergency Regimen instructions
- supplies of glucose polymer and scoops
- parents to check glucose polymer is 'in date' and open tins to be discarded according to manufacturers' instructions

Information sheets to give parents/carers and copies to professionals

- ① Contact names, telephone and bleep numbers
- ② Emergency regimen recipes: for children age over 10 years – 25% carbohydrate
- ③ Alternative Emergency Regimen drinks: for children age over 10 years – 25% carbohydrate

The information sheets given at age 1 year (number 2 and 3 only) are also appropriate for this age group, as needed.

Guideline on maximum 'safe fasting time' for well child

- 12 hours provided child is well and eating normally