Clinical Guideline for the Dietary Management of Paediatric Medium Chain acyl-CoA Dehydrogenase Deficiency (MCADD) Clinical Support and Imaging CMG Nutrition and Dietetic Service	University Hospitals of Leicester NHS NHS Trust **Trust Reference Number C30/2016**

1. Introduction and Who Guideline applies to

These guidelines have been developed to ensure that dietetic advice given to patients with Medium chain acyl-CoA dehydrogenase (MCADD) and their families is consistent and follows current scientific evidence and consensus.

These clinical guidelines are intended for use by Senior Specialist Paediatric Dietitians and Senior Paediatric Dietitians within the Nutrition and Dietetic Service, University Hospitals of Leicester NHS Trust (UHL). Dietitians using the guideline should have had relevant clinical training and clinical supervision from the Senior SpecialistDietitian in Inherited Metabolic Disorders.

Medium chain acyl-CoA dehydrogenase (MCADD) deficiency is inherited as an autosomal recessive disorder with a 1 in 4 chance of a baby being affected from carrier parents. It is the most common fatty acid oxidation disorder with an estimated incidence of up to 1 in 10,000 in England (Dixon et al 2015) and is due to a deficiency of the enzyme medium chain acyl-CoA dehydrogenase which is necessary for theoxidation of medium chain fatty acids (C6-C12). Ketone body and energy production is reduced leading to build up of medium chain fats in particular C8, which are thought to be responsible for the cause for the clinical sequelae (Dixon et al 2015).

MCADD is part of the newborn screening programme (NBS). This is carried out on day 5 of life from a heel prick dried blood spot on the newborn blood spot card, (previously called the Guthrie card). Positive results are phoned through from Neonatal Screening Laboratory at Sheffield Children's Hospital after which further confirmatory tests are carried out.

In countries without NBS MCADD deficiency usually presents between 6 months and 4 years of age but neonatal onset can also occur. The typical picture is of encephalopathy with hypoketotic hypoglycaemia precipitated by metabolic stress such as fasting, gastrointestinal illness; particularly with vomiting or respiratory infections. Without prompt treatment with an appropriate Emergency Regimen patients will progress ultimately to coma with a high risk of mortality (Dixon et al 2015)

Between episodes of metabolic decompensation, patients are usually completely well on a diet of regular meals and snacks. This is because metabolism of medium chain fatty acids from energy stores is not relied upon until the body is in a state of prolonged fast or becomes stressed, as occurs during infection or illness.

2. Guideline Standards and Procedures

Normal diets contain predominantly fatty acids with a chain length of C16-C18. These long chain fats undergo □-oxidation to release energy and the carbon chain length is progressively shortened to medium, then short chain length. Children with MCADD deficiency have impaired oxidation of medium chain fatty acids (MCFA)

which are predominantly derived from long chain fatty acids. However, under basal conditions the oxidation of MCFA has been reported to be near normal due to overlapping enzyme substrate specificity (Heales et al 1994).

2.1 **Diet when well**

The well child can have a normal diet without restriction of long chain fat. It is nevertheless important to avoid prolonged fasts as fatty acid oxidation rates increase as the period of fasting is extended. In infants fatty acid oxidation rates are higher after a shorter duration of fast, therefore frequent feeding (3-4 hourly) is recommended, Both breast milk or standard infant formula are suitable. Weaning should commence as the usual time between 17 and 26 weeks.

Children should have regular healthy meals containing starchy carbohydrate, such as bread, potato, pasta, rice or cereals in normal age related quantities. A starch containing bedtime snack and breakfast are essential meals to minimise the duration of the overnight fast.

Missed meals should be replaced by a suitable snack or milky drink and if this is refused the child should be given their Emergency Regimen (ER), which is a glucose polymer-based drink. The appropriate amount and concentration of the ER is dependent on age (see Emergency Regimen, Appendix 1). As with the normal population, regular consumption of sugary drinks should not be encouraged without clear clinical indication as this can lead to overweight and obesity. This needs to be explained carefully to parents.

Medium chain triglycerides (MCT) occur in only a few foods. Coconut and coconut oil contain a small quantity of MCT which in **normal** quantities are not sufficient to be harmful to a **well** child. All standard infant formulas contain a small amount of coconut oil and are safe to use. Some specialist dietetic products such as Pepti-Junior, Monogen, Paediasure Pepdite and some low birthweight infant formulas contain a high percentage of MCT and must not be given. It is always advisable to check up to date product information available from companies before advising on a specialist feed. A child requiring parenteral nutrition should **not** be given “SMOF” lipid.

Table 1 “Safe” fasting times for the well child

Age	Time in hours
Positive screening to 4 months	6
From 4 months	8
From 8 months	10
From 12 months	12

Source British Inherited Metabolic Disease Group (BIMDG)

a) **At risk neonate with a family history of MCADD**

Neonatal deaths have been reported in MCADD deficiency (Patterson and Henderson 2010). 25% of first metabolic crisis in previously undiagnosed

MCADD was fatal.

If an infant is at risk of MCADD deficiency for example first degree relative, they will betreated as having MCADD from birth and screening will take place on day 2 of life. It isessential that the infant receives adequate feeds and are not starved during the first few days of life or whilst under investigation for MCADD. This particularly important for the breast fed infant who may take longer to establish feeding than the formula fed infant. Supplementary feeds will be required until breast feeding is established or MCADD deficiency excluded.

There is a separate neonatal UHL medical guideline available for this group of patients "Guidelines for Management of Newborn Infants Born to a Family Affected with MCADD" (Trust Ref C4/2012). This contains full details about feeding

Principally

- High risk babies should be screened at 24-48 hours and results made availablepromptly.
- The baby should be fed regularly; term babies at least 3 hourly and pre-term at least 2 hourly day and night until a diagnosis is confirmed or excluded.
- Breast fed babies should receive top up bottle feeds for the first 72hrs until breast feeding is established.
- A volume of 50-60ml/kg/day of standard infant formula is advised for the first 24 hours, increasing to 90ml/kg/day in the next 24 hours building up gradually to 150ml/kg/day by day 6 at the latest.
- If the baby is unable to meet the volumes above then they should receive "top ups" by a nasogastric tube. If there are any problems with toleration then an intravenous infusion of 10% glucose should be given and feeds reviewed.

b) **Dietary management of infants**

Infants can be breastfed or bottle fed with a standard infant formula and weaned on to a normal diet without restriction of fat. Specialist formulas containing a high proportion of MCT should be avoided examples include; Monogen, Infatrini Peptisorb and Pepti-junior, however it is prudent to check up to date product information before changing to a specialist feed.

Principles of feeding

- Allow breast or bottle feeds 3-4 hourly
- Avoid prolonged fasts, particularly at night – refer to table 1 for guidance onage appropriate times
- Commence weaning at the normal age (minimum of 17 weeks for term babiesand 17 weeks corrected for a pre-term baby)
- Wean onto a normal diet with inclusion of regular meals containing starchycarbohydrate
- Avoid specialist infant feeds containing a high proportion of MCT (always checkup to date composition).

c) **Principles of Dietary Management for toddlers and children**

- Avoid prolonged fasts - maximum 12 hours overnight fast
- Avoid missing breakfast
- Give regular meals containing starchy carbohydrate
- Give a late night snack containing starchy carbohydrate
- Replace missed meal with an alternative snack or milky drink or a suitableglucose drink (see Emergency Regimen-Appendix 2)
- Do not give feeds that contain a high proportion of MCT
- Encourage additional carbohydrate snack pre-exercise for those who take part in prolonged exercise. These may need an individual plan. A further snack maybe required post exercise to replenish glycogen stores.

d) **Principles of Dietary Management for adolescents**

The principles in 2.1c apply to this patient group but in addition adolescents should be educated regarding the dangers of alcohol intoxication; vomiting associated with this is particularly dangerous for the MCADD patient. Alcohol inhibits gluconeogenesis, therefore both this and fat oxidation, which provide fuel during fasting are impaired. Patients should be encouraged to have as sensible to alcohol consumption If they choose to consume it then alcohol I should be taken in moderation and alongside food.

2.2 Diet during illness

In contrast to the diet advised for the well child a much stricter dietary regimen is needed during illness (see Appendix 2). It is critical to inhibit the mobilisation of fatty acids, which would result in the production of toxic fatty acid metabolites and precipitate overwhelming illness.

If the child is unwell and has a reduced appetite, the standard Emergency Regimen (ER) appropriate for age should be given without delay. This consists of a regimen of very frequent feeds, day and night, of a specific concentration and volume of glucose polymer dependent on age (Appendix 1). This is a solution of short acting carbohydrate, which helps to prevent the body from breaking down its fat stores.

The ER should be used for up to a maximum of 48 hours at home. As the child improves, the normal diet can be resumed but extra ER drinks should be given, particularly during the night, until the child is fully recovered and eating well.

If the ER is not tolerated i.e. the child will not take the ER, or the child vomits on two successive occasions, they should be taken immediately to The Children's Emergency Department (ED), Leicester Royal Infirmary in order that either a nasogastric tube can be placed to administer the ER (if refused or if target volumes are not met). If the child is vomiting a 0.45% saline 10% glucose with additives must be started immediately by the medical team.

Patients are advised to bring their oral ER and any written information to hospital with them to avoid any delays- all children with MCADD have a

medical alert card provided by the Metabolic Team. Medical staff must follow the BIMDG guidelines that can be accessed from the BIMDG website www.bimdg.org.uk

Monitoring of blood glucose at home is not recommended because hypoglycaemia is a relatively late finding during illness in MCADD deficiency and the child will already be metabolically decompensated at this point.

2.3 **Newly Diagnosed Patients**

Newly Diagnosed Infants should be seen by the Metabolic Team either the same day or the next working day following a presumptive positive screening result (BIMDG 2013). The medical team will take samples for confirmatory tests Further information is available from the newborn screening section of the BIMDG website www.bimdg.org.uk

First Meeting with the family

- Establish method of feeding and frequency
- If bottle fed check that feed does not contain added MCT
- Advise plan for frequency of feeding, feed volumes and maximum fasting times. Provide written information on MCADD
- Explain the necessity for the Emergency Regimen and give a written copy (Appendix 2).
- Provide a supply of glucose polymer.
- Provide TEMPLE information booklet on MCADD

- Advise the family to bring to Children's ED if the baby is unwell or not feeding whilst waiting for confirmation of the diagnosis.
- Arrange for GP letter re diagnosis and arrange for prescription and supply of glucose polymer (Appendix 3)
- Arrange for follow up usually within 7-10 days when confirmatory tests available
- Provide contact details of Metabolic Team

2.4 **Monitoring**

Patients under 1 year of age are usually seen by face to face review on a 3 monthly basis and by telephone as necessary. Thereafter patients are seen in clinic on a 6 monthly basis. Patients are invited to attend an annual group education session run by the Specialist Dietitian and Specialist Nurse, the aim of which is to empower patients and their carers to have a better understanding of their condition and its management, as well as be able to meet with other families whose children have the same condition.

2.5 **Use of Levocarnitine** (L-carnitine)

The use of L-carnitine in MCADD is controversial. Studies looking at the beneficial effect on exercise tolerance are conflicting Lee et al, 2005 and Huidekoper et al, 2006). It is not used routinely in UHL. If a child is experiencing symptoms of muscle weakness and pain in conjunction with low serum carnitine, a trial may be considered under medical supervision, see British National Formulary (BNF) for children for dosing advice (Levocarnitine).

3. Education and Training

Senior Specialist Paediatric Dietitians/ Senior Paediatric Dietitians with appropriate training ideally having undertaken the British Dietetic Association Masters Level Module 4 Dietetic Management of Inherited Metabolic Disorders, Plymouth University.

Ongoing clinical supervision (1:1 and group) should be accessed regularly n.b. contract of employment states x 4 times per rolling 12 months. Dietitians should also ensure that they meet the Health and Care Professional Council (HCPC) standards for continuing professional development (CPD).

4. Monitoring Compliance

What will be measured to monitor compliance	How will compliance bemonitored	Monitoring Lead	Frequency	Reporting arrangements
Appropriate ER for age	Check with parents/child and documenton clinic proforma . Encourage attendance at structured education session	Senior Specialist Paediatric Dietitian	At each Appointment	Clinical notes
Knowledge of appropriate volume ofER	Check with parents/child at each clinic appointment and document on clinic proforma. Encourage at structured education session	Senior Specialist Paediatric Dietitian	At each appointment	Clinical note
Knowledge of safefasting times	Check with parents/child at each clinic appointment and document on clinical notes	Senior Specialist Paediatric Dietitian	At each appointment	Clinical note Discussion with MDT

5. Supporting References

Guidelines for Management of Newborn Infants Born to a Family Affected with MCADD (Trust Ref C4/2012).

Dixon M, Stafford J, White F, Clayton N and Gallagher J (1995) 'Disorders of Mitochondrial Energy Metabolism, Lipid Metabolism and Other Disorders', in Shaw (ed.) Clinical PaediatricDietetics 4th Edition, London, Wiley Blackwell

Heales SJ, Thompson GN, Massoud AF, Rahman S, Halliday D, Leonard JV. Production and disposal of medium- chain fatty acids in children with medium-chain acyl-CoA dehydrogenase deficiency. *J Inherit Metab Dis*, 1994, **17** 74−80
Heales SJ, Thompson GN,Massoud AF, Rahman S, Halliday D, Leonard JV

Huidekoper HH, Schneider J, Westphal T, Vaz FM, Duran M, Wijburg FA. Prolonged moderate-intensity exercise without and with L-carnitine supplementation in patients with MCAD deficiency. J Inherit Metab Dis. 2006 Oct;29(5):631-6. Epub 2006 Aug 2

Lee PJ, Harrison EL, Jones MG, Jones S, Leonard JV, Chalmers RA. 'L-carnitine and exercise tolerance in medium-chain acyl-coenzyme A dehydrogenase (MCAD) deficiency: apilot study.' J Inherit Metab Dis. 2005;28 (2):141-52

Patterson A and Henderson M Early MCADD deaths: 5 cases*. J Inher Metab Dis*, 2010, **33** S59

Useful websites:

British Inherited Metabolic Disease Group http://www.bimdg.org.uk

British National Formulary for Children https://bnf.nice.org.uk/guidance/prescribing-in-children.html

6. Key Words

List of words, phrases that may be used by staff searching for the Policy on

SharePointMedium Chain Acyl-coA Dehydrogenase Deficiency,

MCADD,

Emergency Regimen,

ER

CONTACT AND REVIEW DETAILS	
Guideline Lead (Name and Title) Moira French Senior Specialist Paediatric Dietitian	**Executive Lead**
Details of Changes made during review: Section 1 Reference to incidence in England only Change to paragraph order Information regarding coconut has changed Information regarding at risk neonate with a family history of MCADD matches with the Guideline produced after this document Information regarding first meeting with the family has been updated Contact details of where to present when unwell has changed Generic bimdg website has been included for ease Updated oral Emergency Regimens included in the appendices ICE changed to DIT3 in Appendix 3	

Emergency Regimens Appropriate to Age

Age	Concentration ofglucose polymer	Volume
0-6months	10 %	Aim 150-200ml/kg in 24 hours split into 8 feeds 3hourly or 12 feeds 2 hourly Average amount 75ml every 2 hours or 115mlevery 3 hours
6-12months		Aim 120-150ml/kg in 24 hours split into 8 feeds 3hourly or 12 feeds 2 hourly Average amount 85ml every 2 hours or125ml every 3 hours
12-18 months	15 %	Aim 100ml/kg in 24 hours split into 8 feeds 3hourly or 12 feeds 2 hourly Average amount 85ml every 2 hours or 125mlevery 3 hours
18-24 months		Aim 100ml/kg in 24 hours split into 8 feeds 3hourly or 12 feeds 2 hourly Average amount 100ml every 2 hours or 150mlevery 3 hours
2-6 years	20 %	Aim 1200-1600ml in 24 hours; offer 100 – 130mlevery 2 hours or 150 – 200ml every 3 hours
7-9years		Aim 1500-1800ml in 24 hours; offer 135-150mlevery 2 hours or 210- 220ml every 3 hours
10 and 11 years	25 %	Aim 1900ml in 24 hours; offer 160ml every 2 hoursor 240ml every 3 hours
12 and 13 years		Aim 2000ml in 24 hours; offer 170ml every 2 hoursor 250ml every 3 hours
14 and 15 years		Aim 2250ml in 24 hours; offer 190ml every 2 hoursor 280ml every 3 hours
>16 years		Aim 2500ml in 24 hours; offer 210ml every 2 hoursor 310ml every 3 hours

University Hospitals of Leicester Nutrition and Dietetic Service

EMERGENCY REGIMEN (ILLNESS PLAN) FOR CHILDREN – FACT SHEET
Give with the appropriate age information

What is the Emergency Regimen (ER) (Illness Plan)?
This is a special plan whereby drinks that are high in sugar (glucose polymers) are given regularly during the day and night when your child is unwell and unable to eat properly. The purpose of giving these drinks is to keep your child safe by providing them with energy to help prevent them from breaking down their fat stores. Glucose polymers, e.g. Polycal, Maxijul, Vitajoule and S.O.S are available on prescription from your GP.

When should I give the Emergency Regimen (Illness Plan)?
Anytime your child is not eating well, e.g. during illness.

Step1: If you are unsure if your child is unwell, continue with normal medicines, give the sugary drink and continue to review.

Step 2: If your child is definitely unwell, start the full illness plan of frequent sugary drinks. If your child wants to eat this is fine – try to give them starchy foods such as pasta, potatoes or cereal.

Step 3: If you child is not getting better, not tolerating or refusing to take the sugary drinks you should bring your child to the Children's Emergency Department.

If you go to hospital take your information pack, glucose polymer and scoops if you have them.

Vomiting - Use this rule to assess when to start the emergency regimen:

1 vomit - Start the full Emergency Regimen and monitor (you may find that your child is able to take the 2 or 3 hourly amount as small frequent sips). If your child is able to eat a little but not the usual amount then continue to top with regular sugary drinks.

2 vomits - Continue the Emergency Regimen and bring your child to the Children's Emergency Department immediately.
Your child will need IV glucose if they are unable to tolerate their Emergency Regimen orally or by a nasogastric tube.

How do I give the sugary drinks?
Give glucose polymer drinks every 2 or 3 hours, both **day and night**. If you child is vomiting give the drinks volume needed over the 2-3 hours as small frequent sips.

When can I stop giving the sugary drinks?
When your child starts eating again you can give less sugary drinks but continue some night drinks. The drinks can be stopped once your child is eating normally again. Try to do this within 48 hours or starting the sugary drinks.

Diarrhoea

If your child is feeding normally then do not worry but make sure you give him/her plenty of fluids. You may need to use an oral rehydration solution such as dioralyte particularly if they have profuse diarrhoea.
If you are using the Emergency regimen then add the sachet of dioralyte to the glucose polymer before making up (see the recipes overleaf).

Should I contact the Hospital doctor if my child is on the Emergency Regimen (Illness Plan)?
If you are concerned and want advice then please telephone a member of the Metabolic Team during normal working hours. You should contact the Metabolic Team if you feel that your child may need their Emergency Regimen for more than 48 hours because they are not eating.

Glucose Polymer recipes for children age under 1 year (10% carbohydrate)

20g or 4 scoops* of Polycal (glucose polymer)
Make up to 200ml with cooled boiled water

*always use level unpacked scoops

OR 1 S.O.S 10 sachet (Vitaflo) made up to 200ml with cooled boiled water

How to give the drinks

Age	Carbohydrate solution	Polycal + Water Dilution	S.O.S10	Daily Volume
0-3 months	10%	2 scoops up to 100ml	1 sachet made up to 200ml	60mls every 2 hours or 90mls every 3 hours **Aim for 700-750 in 24 hours**
3-6 months	10%	2 scoops up to 100ml		75ml every 2 hours or 115ml every 3 hours **Aim 800 – 1000ml in 24 hours**
1 year	10%	2 scoops up to 100ml		85ml every 2 hours or 125ml every 3 hours **Aim 1000 – 1100ml in 24 hours**

Dioralyte (oral rehydration solution) recipe

1 sachet of Dioralyte
Add 20g **or** 4 scoops* of Polycal **or** one sachet of S.O.S 10
Make up to 200ml with cooled boiled water

Fluid volume as in the table.

*always use level unpacked scoops

Please show this information to any Doctor who has to see your child.

For further information contact the Metabolic Team:

Moira French Specialist Dietitian Leicester Royal InfirmaryLeicester LE1 5WW	Tel:	0116 2585400
Siobhan Felix Children's Metabolic Specialist NurseLeicester Royal Infirmary Leicester LE1 5WW	Tel:	07921545407
Dr J. Forster Consultant PaediatricianLeicester Royal Infirmary Leicester LE1 5WW	Tel:	0116 2585564 (Secretary) or air page
Childrens Emergency Department Leicester Royal Infirmary Leicester LE1 5WW	Tel:	0116 258 6923

Glucose Polymer recipes for children age over 1 year (15% carbohydrate)

30g or 6 scoops* of Polycal (glucose polymer)
Make up to 200ml with water

*always use level unpacked scoops

OR 1 S.O.S 15 sachet (Vitaflo) made up to 200ml volume with water.

How to give the drinks

Age	Carbohydrate solution	Polycal + Water Dilution	S.O.S 15	Daily Volume
1 year	15%	3 scoops up to 100ml	1 sachet made up to 200ml	85ml every 2 hours or 125ml every 3 hours **Aim 1000 – 1100ml in 24 hours**
18-24 months	15%	3 scoops up to 100ml		100ml every 2 hours or 150ml every 3 hours **Aim 1100 - 1200ml in 24 hours**

Your child may tolerate the 2 or 3 hourly volume given as small frequent sips rather than the full volume given in one go.

Dioralyte (oral rehydration solution) recipe

1 sachet of Dioralyte
Add 30g **or** 6 scoops* of Polycal **or** one sachet of S.O.S 15
Make up to 200ml cooled boiled water

Fluid volume as in the table.

*always use level unpacked scoops

Please show this information to any Doctor who has to see your child.

For further information contact the Metabolic Team:

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Glucose Polymer recipes for children age over 2 years (20% carbohydrate)

40g or 8 scoops* of Polycal (glucose polymer)
Make up to 200ml with water

*always use level unpacked scoops

OR 1 S.O.S 20 sachet (Vitaflo) made up to 200ml with water

How to give the drinks

Age	Carbohydrate solution	Polycal + Water Dilution	S.O.S 20	Daily Volume
2 years	20%	4 scoops up to 100ml	1 sachet made up to 200ml	100ml every 2 hours or 150ml every 3 hours **Aim 1200ml in 24 hours**
3 and 4 years	20%	4 scoops up to 100ml		110ml every 2 hours or 170ml every 3 hours **Aim 1300-1400ml in 24 hours**
5 and 6 years	20%	4 scoops up to 100ml		125ml every 2 hours or 190ml every 3 hours **Aim 1500ml in 24 hours**
7 and 8 years	20%	4 scoops up to 100ml		140ml every 2 hours or 210ml every 3 hours **Aim 1700ml in 24 hours**
9 years	20%	4 scoops up to 100ml		150ml every 2 hours or 220ml every 3 hours **Aim 1800ml in 24 hours**

Your child may tolerate the 2 or 3 hourly volume given as small frequent sips rather than the full volume given in one go.

Dioralyte (oral rehydration solution) recipe

1 sachet of Dioralyte
Add 40g **or** 8 scoops* of Polycal **or** one sachet of S.O.S 20
Make up to 200ml with water

The daily fluid volume is the same as in the table.

*always use level unpacked scoops

Please show this information to any Doctor who has to see your child.

For further information contact the Metabolic Team:

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Childrens Emergency Department Leicester Royal Infirmary Leicester LE1 5WW	Tel:	0116 258 6923

Glucose Polymer recipes for children age over 10 years (25% carbohydrate)

50g or 10 scoops* of Polycal (glucose polymer)

made up to 200ml with water.

*always use level unpacked scoops

OR 1 S.O.S 25 sachet (Vitaflo) made up to 200ml with water

How to give the drinks

Age	Carbohydrate solution	Polycal + Water Dilution	S.O.S 25	Daily Volume
10 and 11 years	25%	5 scoops up to 100ml	1 sachet made up to 200ml	160ml every 2 hours or 240ml every 3 hours **Aim 1900ml in 24 hours**
12 and 13 years	25%	5 scoops up to 100ml		170ml every 2 hours or 250ml every 3 hours **Aim 2000ml in 24 hours**
14 and 15 years	25%	5 scoops up to 100ml		200ml every 2 hours or 300ml every 3 hours **Aim 2400ml in 24 hours**
16 and 17 years	25%	5 scoops up to 100ml		210ml every 2 hours or 310ml every 3 hours **Aim 2500ml in 24 hours**

Dioralyte (oral rehydration solution) recipe

1 sachet of Dioralyte

Add 50g **or** 10 scoops* of Polycal **or** one sachet of S.O.S 25
Make up to 200ml with water

The daily fluid volume is the same as in the table.

Please show this information to any Doctor who has to see yourchild

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Developed and produced by University Hospitals of Leicester Nutrition and Dietetic Service – April 2021

Initial letter to be copied to GP via DIT 3

[Date]

Re: [name of child], [address of child], [NHS number]

[Name of child] has been detected on newborn screening to have a positive (abnormal) test for medium-chain acyl-CoA dehydrogenase deficiency (MCADD). Confirmatory testing is in progress.

MCADD is a rare inherited disorder causing a block in the metabolism of fat into energy. A child with this condition is at risk from hypoglycaemia, coma and death with fasting and particularly during intercurrent illnesses when the body's demand for energy increases and food intake is often reduced. S/he may appear drowsy or lethargic, vomit, have seizures or have a deteriorating conscious level.
Hypoglycaemia is a late sign, and therefore treatment must be initiated if [name of child] is unwell even if the blood sugar is normal.

MCADD requires no special dietary treatment when the child is well apart from avoiding long periods without feeds or food. Parents/carers are instructed on maximum 'safe fasting times' for age (these times get longer with age). Babies can be breast or bottlefed. Specialised infant formula or enteral feeds with added medium-chain triglycerides (MCT) should be avoided. When intercurrent illnesses occur, MCADD is treated with an Emergency Regimen (ER) of high energy feeds/drinks (glucose polymer), please prescribe 1 box of SOS 10 (Vitaflo) PIP Code 353- 59031 (30 x 21g sachets) on repeat as required, if unable to tolerate these for whatever reason such as vomiting or diarrhoea, or refusing, or the child's condition deteriorates, then urgent admission to hospital should be arranged for an intravenous 10% glucose infusion. The child will have open access to the children's admissions unit. When the child is well, they should return to their usual feeds.

The family have been instructed on maximum 'safe fasting times' when well and preparation and use of the Emergency Regimen (ER), details of which are enclosed. Oral Rehydration Solutions (ORS) do not contain sufficient energy to avoid decompensation, and therefore require fortifying with glucose polymer. The recipe for this is contained in the ER. The Dietary Management Guidelines for MCADD are available at www.bimdg.org.uk/mcadd.asp.

The long-term prognosis for MCADD is very good once the diagnosis is known as treatment is straightforward. The condition is inherited in an autosomal recessive fashion, with a 1 in 4 risk of recurrence in each pregnancy.

If you have any questions, please do not hesitate to contact the metabolic team on [contact number/details].

Kind regards