



# Metabolic.ie

National Centre for Inherited Metabolic Disorders



Children's Health Ireland  
at Temple Street



## **Medium Chain Acyl CoA Dehydrogenase Deficiency**

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# Chapter 1: What is MCADD?



Medium Chain Acyl CoA Dehydrogenase Deficiency (MCADD) is an inherited metabolic disorder. It occurs due to a deficiency of the

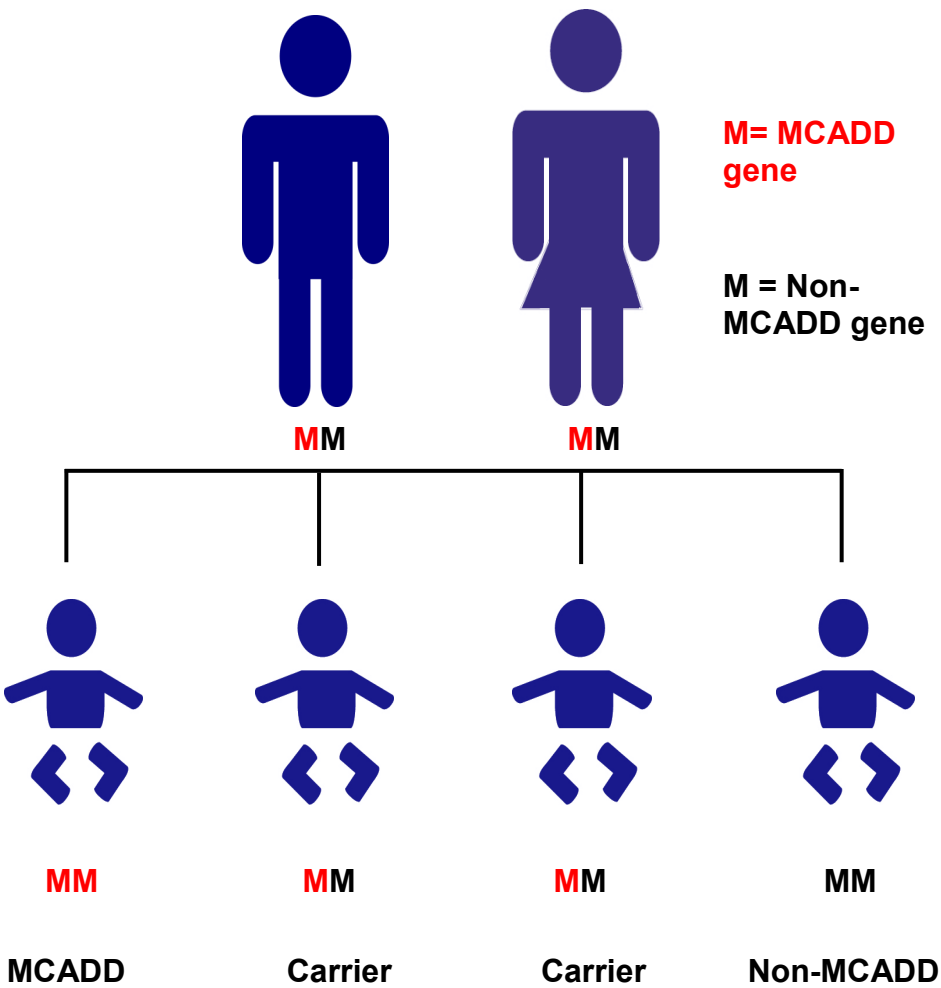
enzyme 'medium chain acyl-CoA dehydrogenase'. This enzyme breaks down certain types of fat in the food we eat to provide energy. It is also used to break down fat already stored in the body when we need energy.

## How is MCADD diagnosed and what causes it?

All babies born in Ireland are screened for MCADD by the heel prick test shortly after birth. Around one in 10,000 babies born in Ireland have MCADD. Some babies may be diagnosed before newborn screening if they develop symptoms of MCADD

Children inherit two sets of genes, one from each parent. As a parent of a child with MCADD, you have one MCADD gene and one non-MCADD gene. This is known as being a carrier. People who are carriers for MCADD do not have MCADD themselves. It is important to remember MCADD is inherited (although there is often no known family history of it) and so it is not because of something you did. There is a 1 in 4 chance that another child born to the same parents will have MCADD.

Fortunately, once diagnosed MCADD is usually easy to manage and your child will go on to lead a healthy normal life. If it was not diagnosed, or not treated it can lead to serious illness, coma and possibly death. Therefore it is important to take it seriously to avoid these severe complications.



## Chapter 2: Overview of the Management of MCADD

### Fasting

Your child must not go without food for extended periods of time. Carbohydrate (or glucose) is the main form of energy used in the body. When glucose has been used up after prolonged fasting, illness or during times of high energy needs such as illness or surgery, the body breaks down body fat stores into long chain fats, medium chain fats and short chain fats to provide an alternative form of energy. Having MCADD means your child's body cannot use medium chain fats effectively for energy once their blood glucose levels drop. This causes medium chain fats and other harmful substances to rise in the body and very low blood glucose levels, which is extremely serious.

The Doctor/Dietitian will tell you how long it is safe for your child to fast for and this will increase as your child gets older until adulthood (See page 13)

### Unwell management

During illness, it is necessary that your child eats carbohydrate foods and drinks sugary fluids more frequently, even if your child does not feel hungry.



If your child is sick, their appetite may be reduced, so they may need to be admitted to hospital for intravenous fluids (drip) to prevent any problems (See page 16)

## Dietary restriction

With MCADD your child's body cannot breakdown medium chain fats (also known as medium chain triglycerides or MCT). However, other types of fats in the diet (long chain and short chain fats), can be eaten normally. Most fats in our diet are long chain fats and can be broken down without any difficulty so no fat restriction in the diet is required.

In general, you should avoid coconut and MCT coconut oil in everyday cooking and avoid foods where coconut is listed as *one of the first three ingredients*.

It is important to be able to read food labels in order to avoid MCTs. The following are two examples of foods contain coconut. Read the ingredients and determine if it is safe to give your child.



## Bounty bar

### Ingredients:

Sugar, **Desiccated Coconut** (21%), Glucose Syrup, Cocoa Butter, Cocoa Mass, Skimmed Milk Powder, Emulsifiers (Soya Lecithin, E471), Lactose, Milk Fat, Whey Powder (from Milk), Humectant (Glycerol), Salt, Natural Vanilla Extract, Milk  
Chocolate contains Milk Solids 14% minimum

Your child should avoid this food as coconut is listed in the top three ingredients.



## Flahavan's Oaty Flapjacks Fruit and Nut

### Ingredients:

Wholegrain Rolled Oats (43%), Vegetable Margarine (Palm Fat\*, Rapeseed Oil, Water, Salt, Natural Flavour), Sugar, Partially Inverted Sugar Syrup, **Desiccated Coconut**, Oat Flour (5.5%),  
\*RSPO Certified Sustainable Palm Oil

This is safe for your child to have as coconut is not in the top three ingredients and therefore the amount in the food is very small.



## Carnitine:

This may be prescribed which helps the body to the fat in the diet more efficiently. The Doctor will advise on the required dosage at each clinic visit if it is prescribed for your child.

# Chapter 3: Infant Feeding When Well

## Does my baby need a special feed?

No, you can give your baby breast milk or standard infant formula. If you are considering changing your baby's infant formula, please contact the metabolic dietitians to make sure it is suitable.

## Breastfeeding

Breast milk is the ideal food for babies. It contains all the nutrients needed for growth and health. It also helps protect babies from bacterial and viral infections.

If your baby has a small appetite or is gaining weight slowly, your dietitian or doctor may suggest top up feeds after breastfeeds. There will be either expressed breast milk or formula milk.

While breastfeeding, you do not need to alter your diet in any way because your baby has MCADD.

## Formula feeding

If you choose not to, or are unable to breastfeed, standard infant formula is an appropriate alternative. Some specialised infant formulas contain





added medium chain fats. If your baby is prescribed a specialised formula, check with your dietitian if it is suitable.

If you are advised to change the infant feeding formula you are giving to your baby by, for example, your local GP or Public Health Nurse and are unsure if the formula is suitable, you should contact the metabolic dietitians for advice before commencing it.

## **Why is regular feeding necessary?**

Babies with MCADD need to avoid using energy from their body fat stores and fasting (going without a feed) for too long.

## **How long can my baby fast for?**

Newborn and young infants normally demand feed every 3-4 hours. As your baby gets older, they will be able to fast for longer. Fasting times are based on age (see page 13).

## **What should I do if my baby will not wake for a night feed?**

If your baby will not wake to take a feed during the night and is fasting for longer than the recommended time (see page 13 ), contact your hospital doctor for further advice.

## **What should I do if my baby becomes unwell and /or feeds poorly?**

Follow your emergency regimen (ER) plan provided by your Dietitian. The ER is a special feeding plan that is only to be used if your baby becomes unwell e.g. poor appetite, temperature, vomiting and/or diarrhoea or has vaccinations (see page 16)

## **When can I wean my baby?**

As per HSE weaning recommendations, babies can be weaned from the age of 17 weeks. As weaning progresses, try to give your baby complex carbohydrate foods at each meal (See page 20)

## **When can I give my baby cow's milk to drink?**

From one year. As solid food increases, your baby will naturally take fewer milk feeds.

# Chapter 4: Healthy Eating from 1 Year Onwards

Children should follow the HSE food pyramid healthy eating guidelines, paying particular attention to including regular complex carbohydrates and avoiding medium chain fats..



For more information see: <https://www.safefood.net/healthy-eating/guidelines>

## Chapter 5: When your Child is Well

In general the fasting times for babies based on age are:

Age	Fasting Times When Well
0-1 month	3 hours
1-3 months	4 hours
3-6 month	4 hours during the day and 6 hours overnight
6-12 months	4 hours during the day and 6-8 hours overnight
1 year	5 hours during the day and 10-12 hours overnight

### Older Children

In general the maximum fasting times for older children when well are:

Age	Fasting Times When Well
2-10 years	12 hours overnight



Overall it is important that your child:

- Avoid prolonged fasts as carbohydrate is important to provide energy so that their fat stores are not broken down for energy.
- Eat regular meals based on complex carbohydrate foods, aiming for 3 meals and 2-3 snacks daily (See page 20 for examples of complex carbohydrates).
- Include a complex carbohydrate food at supper time before bed
- Not miss meals. If your child misses a meal, ensure a carbohydrate snack or carbohydrate containing drink is eaten before the next meal time
- Has no need for additional sugary foods such as fizzy drinks or sweets when well.

## Chapter 5: When Your Child is Unwell

For times of illness, your child's Consultant will advise on an **overnight fasting time**. The following points should be adhered when your child is sick:



- Follow the Emergency regime feeding plan (see page 16)
- Regular carbohydrate drinks appropriate for age must be encouraged in the volumes recommended (see page 20 for examples of sugary carbohydrates)
- If your child is not eating much, you may need to give them additional carbohydrate drinks
- Be aware that they will have reduce fasting times both day and night when they are unwell
- It is likely you will need to wake your child during the night for a carbohydrate rich drink
- If necessary, you may need to bring them to your local hospital, they should be sipping on their sugary drink en route
- It is important that you contact a member of the Metabolic team in Temple Street for advice

### **Metabolic Medical Team in Temple Street**

01 8784317 (9am-5pm Mon-Fri) ask for Metabolic Nurse Specialist  
01 8784200 (other times) ask for the Metabolic Doctor on-call

## Chapter 6: Emergency Regimen

The Emergency Regimen(ER) is a special feeding plan used if your child is unwell. Very regular feeds of carbohydrate are needed to provide energy during the day and overnight to reduce the length of time your child fasts.

The ER should be commenced at home if your child is unwell i.e.

- has a temperature above 38 degrees celsius,
- is vomiting,
- has diarrhoea,
- Seems unwell with poor energy levels or they are off their food.

It is important to start the carbohydrate drinks promptly without delay. These drinks are

called 'SOS' and come in different sizes based on your child's age.



Age	% CHO Concentration	Suitable SOS Product
0-12 months	10	S.O.S. 10
1 year	15	S.O.S. 15
2-9 years	20	S.O.S. 20
10+ years	25	S.O.S. 25

If your child dislikes 'SOS', there are alternative drinks available such as 'Maxijul' or 'Polycal'. Recipes for the use of these drinks can be obtained from your child's dietitian. It is vital that you contact us if your child doesn't like SOS.

It is essential that you understand the emergency regimen, keep it in an accessible location and have a supply of SOS that are in date. A prescription will be provided for the SOS.

**For older children:** Give regular complex carbohydrates meals, sips of SOS/sugary drinks during the day. Overnight, give the recommended volume of SOS at the times as per the Emergency Regimen. If they are unable to tolerate the full volume, you may need to wake your child and offer the SOS drink more often.

## **What about oral rehydration solutions such as Dioralyte?**

Oral rehydration solutions such as Dioralyte are often recommended by health care professionals to treat vomiting and diarrhoea. These solutions alone do not contain sufficient glucose for an unwell patient with MCADD and therefore **if used, they must be fortified with SOS**. Each emergency regimen will have an oral rehydration solution recipe which is fortified with glucose. If vomiting and/or diarrhea persists, you should bring your child to your local hospital for a medical assessment and intravenous dextrose.



## Example of an Emergency Regimen:

### Emergency Regimen: for 1-2 year olds

#### 15% Carbohydrate Recipes

*(once made, store at the back of the fridge and use within 24 hours; shake before use)*

#### SOS recipe\*

1 sachet of SOS 15 (31 g)

Add water to 200ml

Or

#### Oral rehydration solution\*\* recipe (12 % Carbohydrate Recipe)\*

1 sachet of Dioralyte (4.1 g)

5 white scoops of SOS 15 powder (21.5 g)

Add water to 200 ml

\*Sugar-free squash can be added to flavour these drinks but it must be added before the water (add SOS 15 powder, then add sugar free squash and then add water to the amount advised).

\*\*If your child is vomiting and/or has diarrhoea, the oral rehydration solution recipe should be used. Please note that this recipe contains less carbohydrate than the other recipes in the Emergency Regimen so if vomiting and/or diarrhoea persists, you should bring your child to the local hospital for medical assessment and intravenous Dextrose.

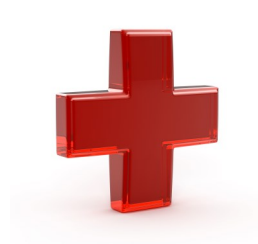
#### Recommended minimum feed volumes:

Age	Recommended minimum feed volumes:
1-2 years	100ml every 2 hours or 150ml every 3 hours day and night

If the above are refused please see the alternative 15 % Carbohydrate drink recipes overleaf.

When your child is well and starts eating again, you can give less carbohydrate drinks but continue some carbohydrate drinks at night. Carbohydrate drinks can be stopped once your child is eating normally again.

## An admission to hospital is needed if your child is:



- Refusing or vomiting the emergency regimen
- Not taking the full amount of recommended carbohydrate drinks
- Has diarrhoea (unless very mild)
- Is not improving on the emergency regimen
- Refusal of food and drinks for more than 6 hours (baby) or 8 hours (over 1 year old)
- Drowsiness, floppiness, not responding normally

Your Metabolic Consultant will provide you with an emergency letter which will have a plan for when you take your child to the hospital that will prompt review in A&E and guide your child's initial treatment. **You should also contact the Metabolic Doctor/ Dietitian using the details on page 14**

### **If you need to go to your local hospital, you need to bring:**

- Your child's emergency medical letter
- Copy of the emergency regimen
- Supply of SOS
- Beakers and scoops required for the emergency regimen

# Chapter 7: Carbohydrate Foods

Complex carbohydrates should be included at all meals when well and the sugary carbohydrate foods are only necessary when unwell.

Complex when well	Sugary when unwell
<ul style="list-style-type: none"><li>• Bread &amp; Bread rolls</li><li>• Pasta, Rice Noodles</li><li>• Potatoes</li><li>• Popcorn</li><li>• Breakfast cereals such as Weetabix, Porridge, Ready Brek,</li><li>• Scones</li><li>• Crackers</li><li>• Crispbread</li><li>• Pancakes</li></ul>	<ul style="list-style-type: none"><li>• SOS 10, 15,20 or 25 (see page 10)</li><li>• Lucozade/Coke/7up/Fruit Juice/Ribena (remember not diet, 'zero', sugar free or no added sugar drinks)</li><li>• Sugar/Jam/Honey/Marmalade</li><li>• Sweets such as jellies, and boiled sweets</li><li>• Milk and milk puddings</li><li>• Yoghurts (not sugar free versions)</li></ul>



**Complex carbohydrates include potatoes, pasta and rice**

## Chapter 8: Exercise and Sport

Carbohydrates are the most important form of fuel for exercise and sport activity. During exercise and sport the body uses a lot of energy. When you use a lot of energy you use a lot of glucose (carbohydrates). Therefore, it is important that you make sure you eat regular meals and snacks containing carbohydrate foods on days that you are more active. They may need to bring a sugary drink with them for long periods of exercise.



## Chapter 9: Preparing for Childhood, Pre-school and School

It is important to let anyone looking after your child that they have MCADD and alert you if:

- They are not eating well, especially the carbohydrate part of their meal
- They become unwell
- They are unable to focus at school, appear overly fatigued, or are acting different than usual.
- They are participating in sport or will be outside in hot weather for a long period of time

## Chapter 10: Travel

Make sure that you are prepared if your child becomes unwell while you are away and requires their emergency regimen.

You will need:

- SOS sachets- make sure you have enough as these are not available to buy over the counter
- Medical letter from metabolic team
- Customs letter for security at the airport from metabolic team
- Any other medication such as Carnitine if prescribed



# Chapter 11: Teenage and Adult Considerations

## Alcohol

Binge drinking is dangerous, particularly if you have MCADD. You may forget to eat after drinking, which can mean prolonged fasting. It can also cause blood glucose levels to drop which can be very dangerous leading to confusion, coma or death.

## Healthy Eating guidelines

Children and adults with MCADD do not require any additional calories when well therefore, it is important to follow the general healthy eating guidelines.

In particular MCADD patients should:

- Choose wholegrain cereals, bread, pasta and rice
- Aim for 5-7 portions of fruit and vegetables a day. Include fruit and vegetables with each meal and as snacks throughout the day
- When buying vegetable oils, avoid MCT oil

## The Food Pyramid

For adults, teenagers and children aged five and over

Not needed for  
good health.

Foods and drinks high in  
fat, sugar and salt

NOT  
every  
day

!

Maximum once or twice a week

Fats, spreads and oils

In very  
small  
amounts

Meat, poultry, fish, eggs,  
beans and nuts

2  
Servings  
a day

Milk, yogurt and cheese

3  
Servings  
a day  
5 for  
children age  
9-12 and  
teenagers  
age 13-18

Wholemeal cereals  
and breads, potatoes,  
pasta and rice

3-5\*  
Servings  
a day  
Up to 7\*  
for teenage  
boys and  
men age  
19-50

Vegetables,  
salad and fruit

5-7  
Servings  
a day

Needed for good health. Enjoy a variety every day.

\*Daily Servings Guide - wholemeal cereals and breads, potatoes, pasta and rice

Active	Children (6-12)	Teenager (13-18)	Adult (19-50)	Adult (51+)
♂	3-4	4	4-5	3-4
♀	3-4	4	4-5	3-4
♂	3-5	5-7	5-7	4-5
♀	3-5	5-7	4-5	4-6

There is no guideline for inactive children as it is essential that all children are active.

Source: Department of Health, December 2016.

Drink at least 8 cups  
of fluid a day -  
water is best



Get Active!



To maintain a healthy weight adults need at least 30 minutes  
a day of moderate activity on 5 days a week (or 150 minutes a  
week); children need to be active at a moderate to vigorous level  
for at least 60 minutes every day.



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**Children's Health Ireland at Temple Street**  
**Dublin 1**  
**01 878 4317**

**Email: [metabolic.dietitians@cuh.ie](mailto:metabolic.dietitians@cuh.ie)**  
**[www.metabolic.ie](http://www.metabolic.ie)**



Author: Metabolic Dietitians

Version: 3

Review date: April 2024

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