



Metabolic.ie

National Centre for Inherited Metabolic Disorders

MCADD

**Medium Chain Acyl CoA
Dehydrogenase
Deficiency**

WHAT is MCADD?

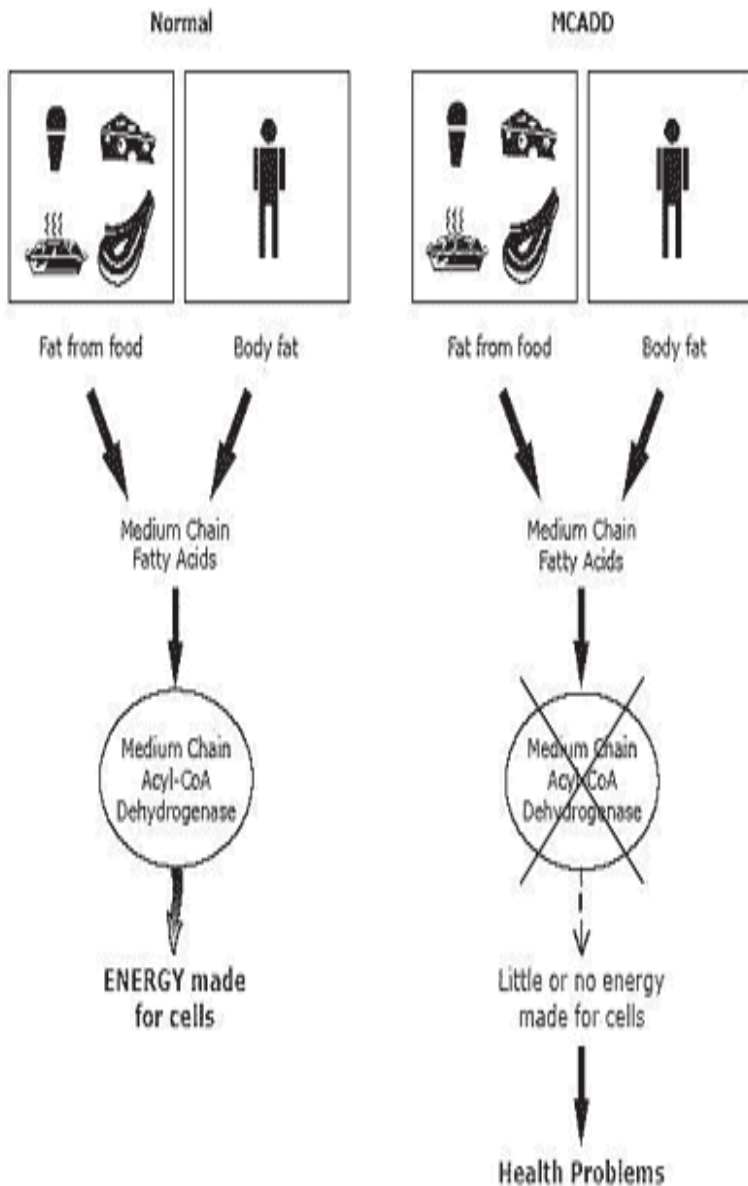
MCADD occurs due to a deficiency of the enzyme “medium chain acyl-CoA dehydrogenase” (MCADD). This enzyme breaks down certain fats in the food we eat into energy. It is also used to break down fat already stored in the body when we need energy.

In MCADD the body cannot breakdown medium chain fats. 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 needed. (**Note: Coconut and coconut containing foods are the exception—see page 4**)

If this condition is not diagnosed early or is ignored and not treated appropriately it could lead to serious illness and possibly death. Fortunately, once diagnosed, MCADD is usually easy to manage and children and adults with this condition lead healthy normal lives.



Medium Chain Acyl-CoA Dehydrogenase Deficiency MCADD



MANAGEMENT of MCADD

1. You / 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 carbohydrate or glucose have been used up, the body breaks down fat to provide an alternative form of energy. In people with MCADD it is essential to prevent this from happening to avoid becoming unwell. Eating frequent carbohydrate containing meals and snacks prevents the body running out of glucose and breaking down fat stores.

The Doctors / Dietitians will tell you how long it is safe for you / your child to fast for and this will increase as you / your child gets older until adulthood.

2. Prompt action when unwell

During illness it is necessary that you or your child eat carbohydrate foods and drink fluids more frequently, even if you or your child does not feel hungry. You will be provided with recipes appropriate for you or your child's age that will need to be used during illness. These recipes use a glucose powder to provide the additional carbohydrate necessary when unwell. During illness our appetite can be reduced and so it may be necessary for you or your child to be admitted to hospital for intravenous fluids to prevent any problems.

3. Carnitine

This may be prescribed which helps the body to use long chain fats more efficiently. Your doctor will advise on the required dosage at each review if it is prescribed.

WHEN WELL:



Overnight fasting time when WELL.....HOURS

- Avoiding prolonged fasts.
- Eat regular meals based on starchy foods: **see list on page 7**
- Ensure you / your child includes a starchy carbohydrate food at breakfast **and** supper times.
- If you / your child misses a meal, ensure some starchy snack or carbohydrate containing drinks are taken until the next meal time e.g. toast, cereal.

It is not necessary to avoid fat in the diet when well. **However, coconut and products derived from coconut, such as coconut milk, coconut oil etc, contain significant amounts of medium chain fat and should be avoided.**



WHEN UNWELL:



Overnight fasting time when **UNWELL.....HOURS**

- Regular carbohydrate drinks appropriate for age must be encouraged in the volumes recommended
- Particular attention should be taken if oral intake is poor from ordinary foods — give additional carbohydrate drinks.
- Reduce fasting times as advised by your Doctor/ Dietitian.
- The night fast should also be included, so it may be necessary to wake yourself or your child during the night for a carbohydrate rich drink.
- You will be provided with a list of suitable drinks which can be used appropriate for you / your child's age.
- **Remember most diet, sugar free or no added sugar drinks or squashes contain no carbohydrate and so are not suitable.**

If your child is vomiting or has persistent diarrhoea, they may require a hospital admission for IV (intravenous) dextrose supplementation. In this case it is vital to contact a member of the Metabolic Medical Team in Temple Street for advice:

01 8784317 (9am-5pm Mon-Fri)

01 8784200 (other times)

and ask for metabolic doctor on-call.

CARBOHYDRATE FOODS

The following are suggestions for carbohydrate foods to include in your diet. The starchy carbohydrates should be included at all meals when well and the sugary carbohydrate foods are usually necessary when unwell. Sugary drinks should not be used on a regular basis when well. If you are concerned about your child's appetite contact your dietitian.

Starchy Carbohydrate

- Bread & Bread rolls
- Potatoes
- Pasta
- Rice and Noodles
- Popcorn
- Breakfast Cereals
- Scones
- Pizza Bases
- Crackers/Crispbread



Sugary Carbohydrates

- Lucozade/Coke/Orange/Fruit Juice/
Ribena
- Sugar/Jam/Honey/ Marmalade
- Fruit
- SOS 10, 15, 20, or 25
- Drinks augmented with Maxijul/SOS
- Boiled Sweets
- Jelly
- Milk and Milk puddings
- Yoghurts



CARBOHYDRATE DRINKS USED DURING the EMERGENCY REGIMEN

The Emergency regimen is a special feeding plan used if you or your child is unwell. Very frequent feeds of carbohydrate are given to provide energy.

The table below advises on the appropriate carbohydrate concentration of drinks for different age groups.

Standard Carbohydrate concentrations for age

AGE	% CHO CONCENTRATION	SUITABLE PRODUCTS
0-12 months	10	S.O.S. 10 or Maxijul*
1 year	15	S.O.S. 15 or Maxijul*
2-9 years	20	S.O.S. 20 or Maxijul*
10+ years	25	S.O.S. 25 or Maxijul*

*Please ensure appropriate sheet for age and recipes are provided.



TABLE of APPROPRIATE VOLUMES for AGE

Suggested feed volumes for age to meet requirements:

Age	2 hourly volumes	3 hourly volumes	Total volume over 24 hours
0-3 months	45-80ml	70-120ml	540-960ml
4-6 months	85-100ml	130-150ml	1020-1200ml
7-9 months	90-100ml	130-150ml	1040-1200ml
10-12 months	100ml	150ml	1200ml
1, 2 years	100ml	150ml	1200ml
3, 4 years	110ml	170ml	1320-1360ml
5, 6 years	130ml	200ml	1560-1600ml
7, 8 years	140ml	210ml	1680ml
9, 10 years	150ml	220ml	1760-1800ml
11, 12, 13 years	170ml	250ml	2000-2040ml
14, 15 years	180ml	270ml	2160ml
16 years and over	200ml	300ml	2400ml



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