



MCADD (Medium-chain acyl CoA dehydrogenase deficiency)

What is MCADD?

Medium-chain acyl CoA dehydrogenase deficiency (MCADD) is a rare, inherited metabolic disorder where the body is unable to use medium chain fats for use as an energy source. There are different chain lengths of fat: long, medium and short. The main source of fat in our diet is long chain fat and when we are well this is the main fat fuel for our body. MCADD should not affect your everyday life.

It will only be a problem if you are unwell, have a hangover from drinking too much alcohol, are partaking in strenuous exercise, or you have not eaten for a long time. During these times the body may need to use medium chain fats as an extra energy source, but it has a problem with breaking them down for energy. This can lead to toxins building up in your body and making you unwell.

Do I need to be on a special diet?

You can eat a normal diet as the main source of fat in the foods we eat is long chain fat. There are only a few foods that contain medium chain fats, which include coconut and coconut oil. Small amounts in your diet as ingredients in food should be ok when you are well. As you get older your body can cope with not eating for long periods without a problem. When you are well you can fast for up to 12 hours overnight.

Treatment when unwell

When you are unwell your appetite decreases and your body needs more energy to help recover. The body will use stored long chain fats for energy, breaking them down to medium chain. We want to prevent the body having to use its medium chain fats for energy. This can be done by taking regular carbohydrate drinks when unwell to give the body glucose to use as energy instead. If you are unable to tolerate the drinks, then call your metabolic team or go straight to a hospital emergency department for an intravenous glucose drip. See emergency regimen section.

Follow these guidelines to ensure you stay safe:

- Eat regular meals – breakfast, lunch and an evening meal.
- Do not miss meals, especially breakfast. If you are going out and think you may miss a meal then take a snack with you e.g. cereal bar, fruit, sandwich or crackers.
- Each meal should contain a normal amount of carbohydrate for energy e.g. bread, potatoes, rice, pasta, cous cous, grains, breakfast cereals or crackers. Check the food packaging for standard portion size or speak to your dietitian if unsure.
- If needed, have a bedtime snack with carbohydrate as it is a long time until breakfast. For example milk and fruit, couple of biscuits or crackers, toast and jam, bowl of cereal or a currant bun or crumpet.
- If you are unwell and eating less than normal then start your emergency drinks. Contact your metabolic team for advice.

Information for Patients

Keeping well and being sensible with MCADD

1. Alcohol

If you get a hangover from drinking alcohol and you cannot eat then you must take your emergency drinks. If you are vomiting, and cannot keep the emergency drinks down then you must go to a hospital emergency department as soon as possible for IV 10% dextrose.

Stay safe by:

- Drink in moderation, no more than 14 units per week (this is equivalent to six pints of average strength beer or 10 small glasses of low strength wine)
- Units must be spread over the week with a couple of alcohol-free days
- Drink alcohol with a meal
- Have snacks/meal before and after drinking alcohol
- Space your alcoholic drinks out by drinking non-alcoholic drinks, water or having snacks in between your drinks
- If you are going out and won't be back until very late, then eat regular snacks, especially a bed time snack
- Ensure you have identification on you to let people know you have MCADD
- Tell a friend about your MCADD

2. Weight control

If you want to lose weight, then you must discuss this with your metabolic dietitian first. Weight loss must be slow and steady, avoid fad and crash diets. Losing weight quickly can cause your body to break down fat too quickly potentially, leading to a build-up of toxic chemicals and cause you to become unwell.

3. Surgery

If you are going for any surgery or medical procedure (especially if it requires fasting or a general anaesthetic), then contact your metabolic team for advice as soon as you know you are having it so they can liaise with your medical team about keeping you safe during and after the procedure.

4. Exercise

People with MCADD can do moderate intensity exercise without a problem. If you participate in competitive sport or endurance/prolonged sports, then please contact your metabolic dietitian for advice regarding carbohydrate intake.

5. Pregnancy

If you are female and planning a pregnancy, or become pregnant, then contact your metabolic team for a clinic appointment to discuss how to stay safe during pregnancy and birth. MCADD will only be a problem if you have morning sickness which reduces your appetite and/or causes vomiting and if you have a prolonged or difficult labour or a C section is needed (planned or unplanned).

Emergency regimen for illness

When should I start the emergency regimen?

When you are unwell and eating less than normal, start vomiting or have diarrhoea (two loose stools within two to three hours) then you must start your emergency regimen drinks.

What is an emergency drink?

High sugar drinks that should be taken during illness. They will help minimise body fat breakdown and the build-up of toxic chemicals during illness.

Information for Patients

What kind of drinks should be given?

Any drink that is high in sugar, easy to take and well tolerated.

Carbohydrate powders (on prescription) mixed with water can be used to make a 25% carbohydrate drink. Fruit juice or squash can be added for taste.

Your recommended 25% carbohydrate drink is:

.....

.....

.....

You can also buy high sugars drinks from the shops. Check the carbohydrate content of the drink by checking the carbohydrate content in 100mls.

For example Coco Cola is 10g of carbohydrate per 100mls = 10% carbohydrate drink.

Always check the label as diet, light or sugar-free drinks will be too low in sugar.

How much should I drink when I am unwell?

If you are still eating, but eating less than normal, then take a drink in between meals.

If you are managing very little food or none, then take 200mls of your 25% carbohydrate emergency drink continuously every two to three hours **day and night**.

If you choose a commercial high sugar drink then you will need to drink more as the carbohydrate content is lower. Discuss this with your dietitian.

What if I do not tolerate the emergency drink?

Contact a member of the metabolic team for help or advice. Alternatively, go to the emergency department at your local hospital. Remember to take your emergency letter about what MCADD is so you are started on a 10% IV dextrose drip quickly and take a supply of your emergency drinks.

Metabolic Team contacts

Dietitian	Monday – Friday 08:30-16:00	0121 371 6989 07799 038656
Clinical Specialist Nurse	Monday – Friday 08:00 -16:00	0121 371 6985/6986
Consultant (doctor) on call	Only for out of hours emergency (evening/weekend)	Call Switchboard 0121 627 2000 and ask to speak to the inherited metabolic disorders consultant on call.

Department of Inherited Metabolic Disorders

Queen Elizabeth Hospital Birmingham

Mindelsohn Way, Edgbaston, Birmingham B15 2GW

Telephone: 0121 371 6989

Information for Patients

Accessibility

To view this information in a different language or use the text-to-speech reader visit www.uhb.nhs.uk, click the yellow and black circular icon in the bottom right of the web page to open the ReachDeck toolbar and then use the search bar to search by the name of the leaflet. If you require this information in another format such as braille or audio please email interpreting.service@uhb.nhs.uk.



How did we do?

If you have recently used our services we'd love to hear about your experience. Please scan the QR code or follow the link to share your feedback to help us improve our services. Thank you! www.uhb.nhs.uk/fft

