

# TEMPLE



Tools **E**nabling **M**etabolic **P**arents **L**Earning

ADAPTED BY THE DIETITIANS GROUP

**BIMDG**

British Inherited Metabolic Diseases Group



BASED ON THE ORIGINAL TEMPLE WRITTEN BY  
BURGARD AND WENDEL

VERSION 4, JANUARY 2025

**MMA**

Supported by **NUTRICIA**  
as a service to metabolic medicine

# TEMPLE foreword

TEMPLE (Tools Enabling Metabolic Parents LEarning) are a set of teaching slides and booklets that provide essential information about different inherited metabolic disorders that require special diets as part of their management. These teaching tools are aimed at parents who may have an infant or child that has been recently diagnosed with a disorder. They are also useful when teaching children, extended family members, child minders, nursery workers and a school team.

**This teaching tool is not designed to replace dietary information that may be given by a dietitian in clinic.**

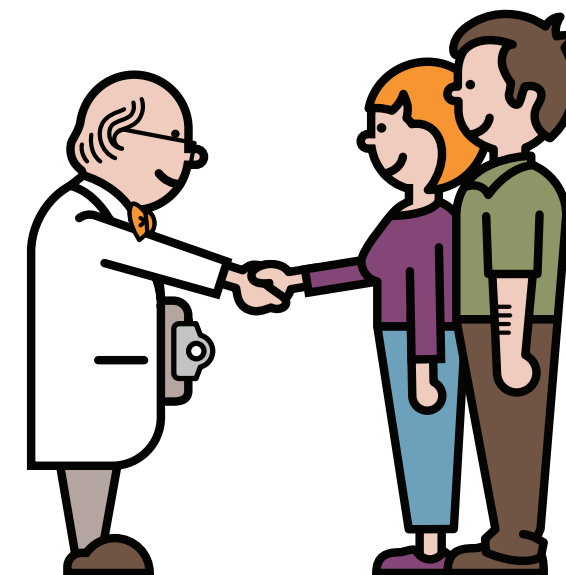
They have been developed by a team of experienced clinical and research metabolic dietitians from the UK who are members of the British Inherited Metabolic Disease Group (BIMDG).

The team are Rachel Skeath, Karen van Wyk, Pat Portnoi and Anita MacDonald. The group is facilitated by Heidi Chan from Nutricia.

Each module produced is reviewed by a consultant clinician who is a member of the BIMDG.

# MMA

Information for parents following  
a new diagnosis



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**TEMPLE**  
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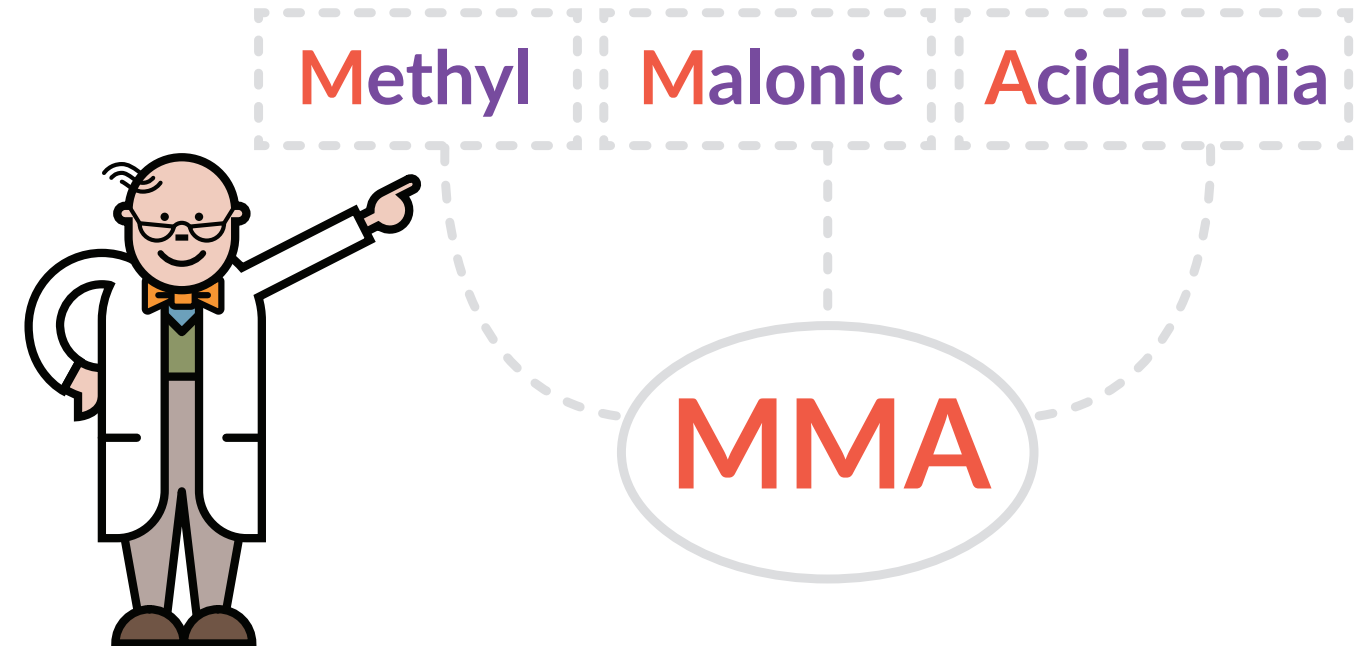


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# What is MMA?

MMA stands for Methyl Malonic Acidaemia

It is an inherited metabolic condition

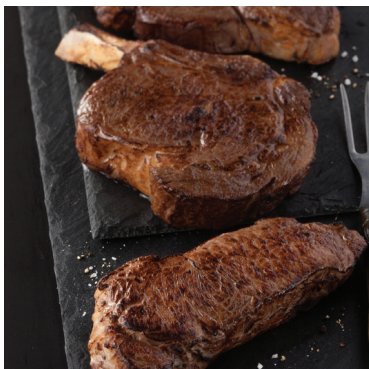


# MMA and protein

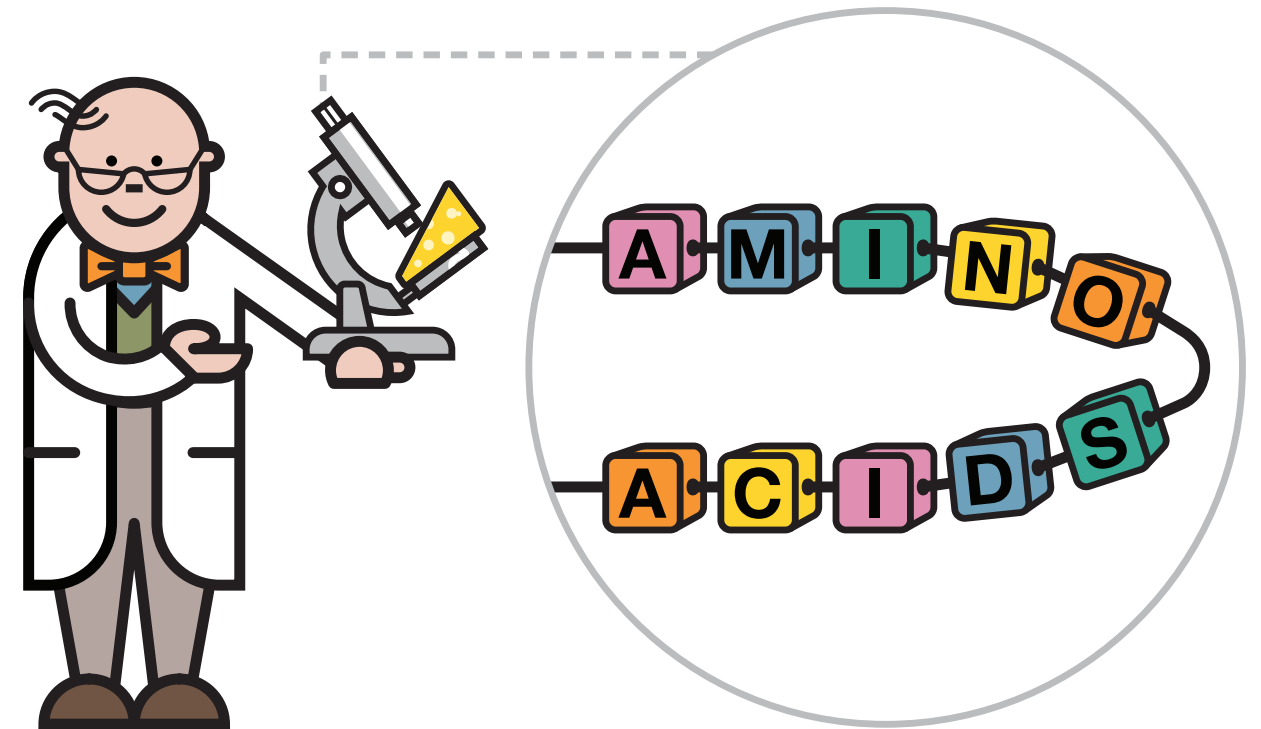
MMA affects the way your baby breaks down protein

Many foods contain protein

The body needs protein for growth and repair



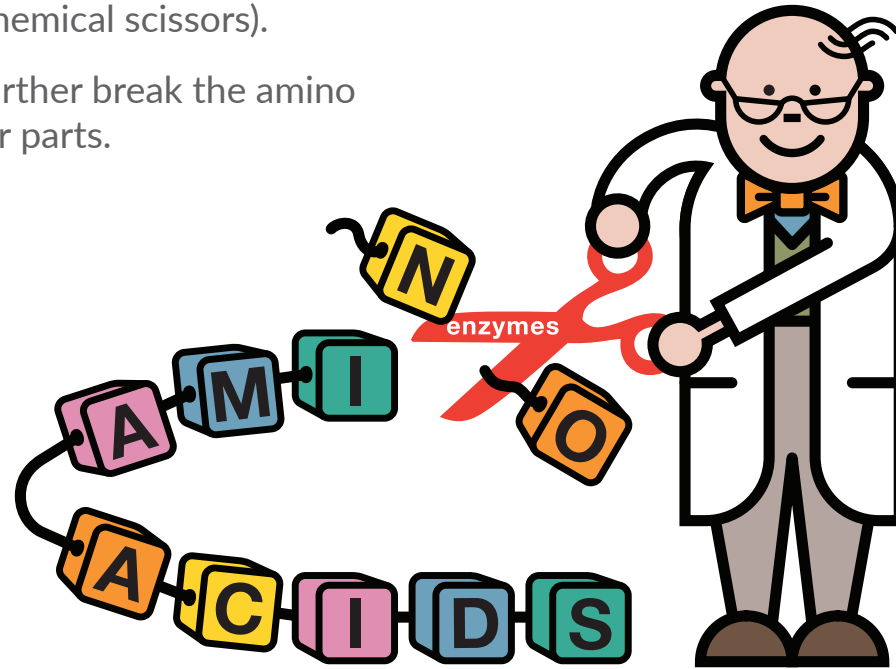
## What is protein?



# Protein and enzymes

Protein is broken down into amino acids (building blocks of protein) by enzymes (which are like chemical scissors).

Enzymes then further break the amino acids into smaller parts.

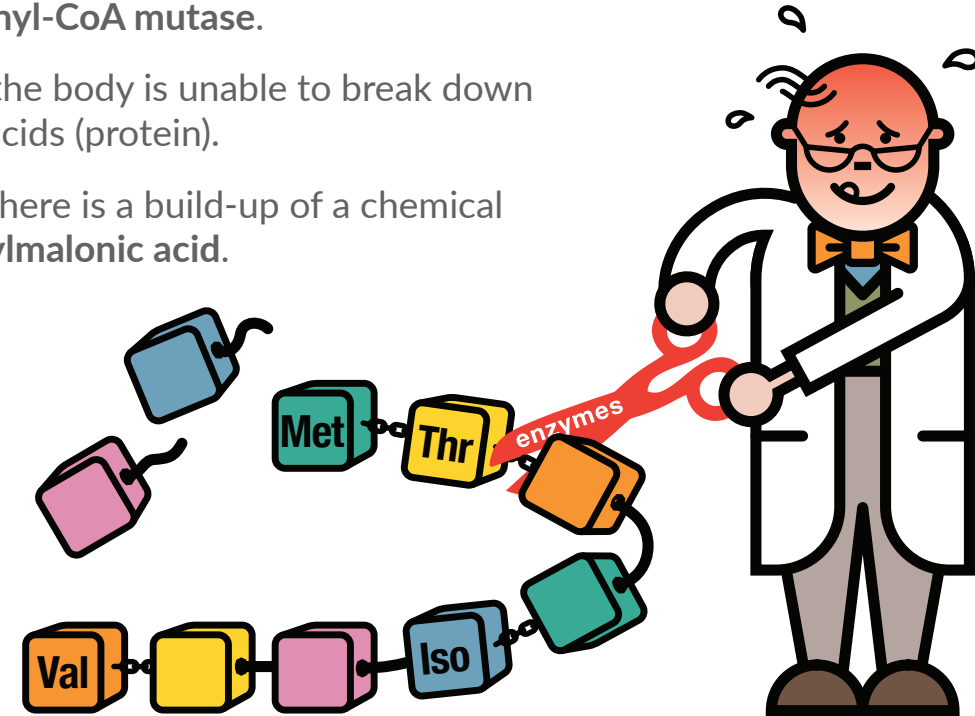


# What happens in MMA?

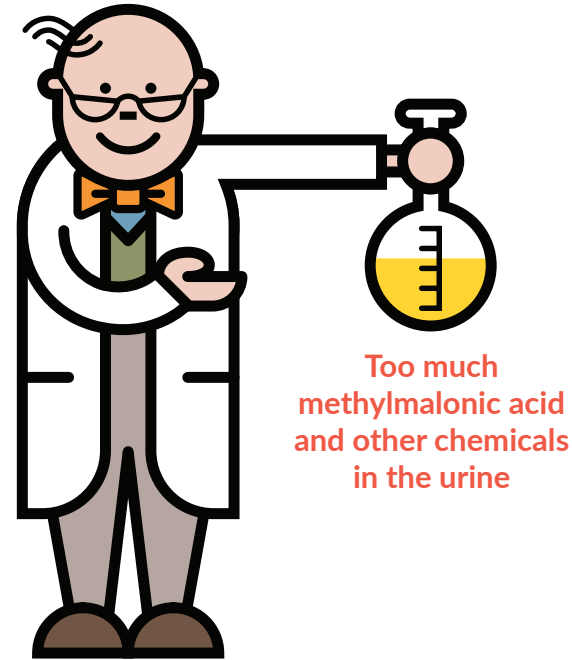
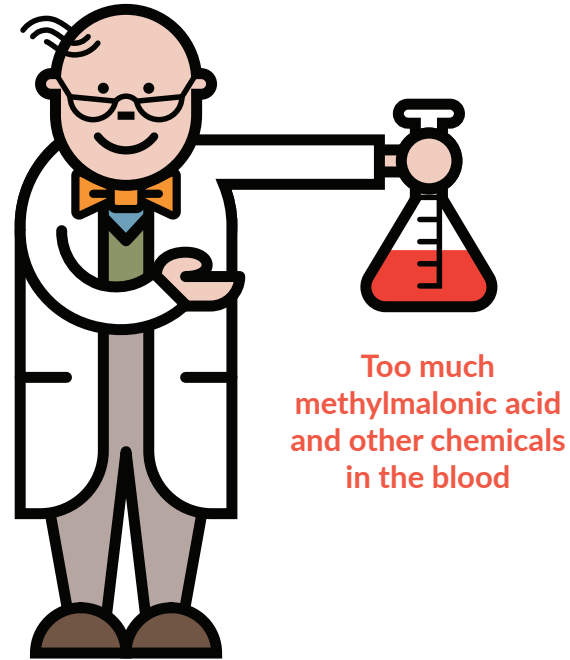
In MMA, the body lacks an enzyme called **methylmalonyl-CoA mutase**.

This means the body is unable to break down four amino acids (protein).

As a result, there is a build-up of a chemical called **methylmalonic acid**.



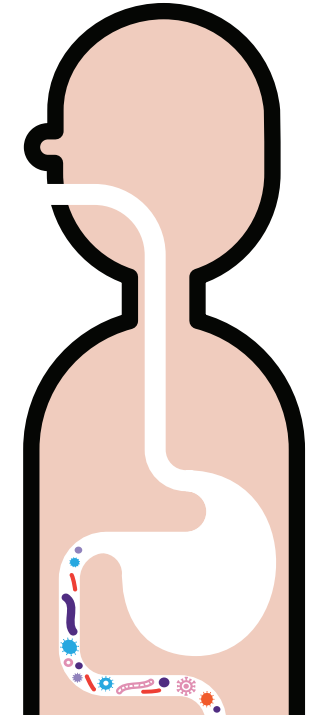
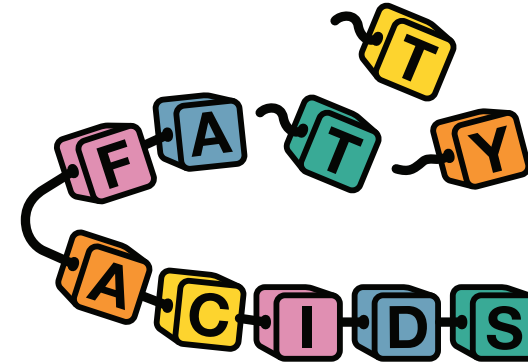
## What is does this cause?



## Other sources of methylmalonic acid

Methylmalonic acid also comes from:

- The breakdown of fatty acids. The body will use these for energy when it has gone a long time without food
- Gut bacteria



## How is MMA diagnosed?

MMA is diagnosed by measuring high levels of methylmalonic acid and other chemicals in the blood and urine.

It can also be diagnosed by looking at enzyme levels and at the body's genes.



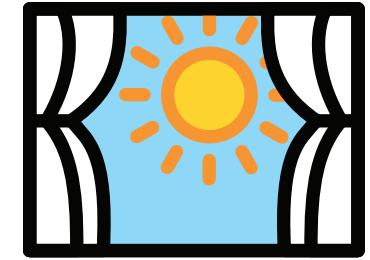
## What are the symptoms in MMA?

Some babies with MMA become ill in the first few days of life.

Symptoms include:

- poor feeding
- vomiting
- dehydration (lack of body fluids)
- floppy baby
- excessively sleepy
- rapid breathing
- seizures

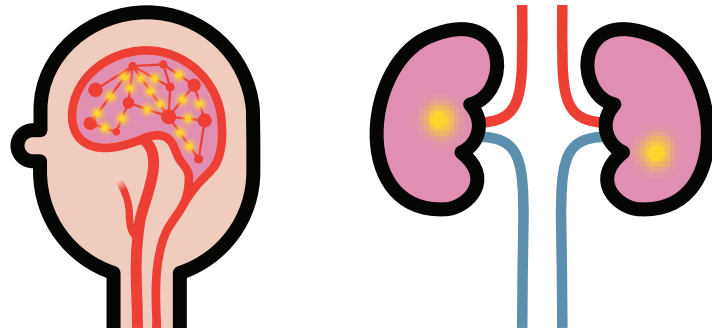
**The effects of MMA quickly become life-threatening if unmanaged**



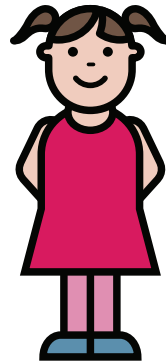


## What can go wrong in MMA?

The build up of harmful chemicals can damage the brain and kidneys and cause problems with other organs.



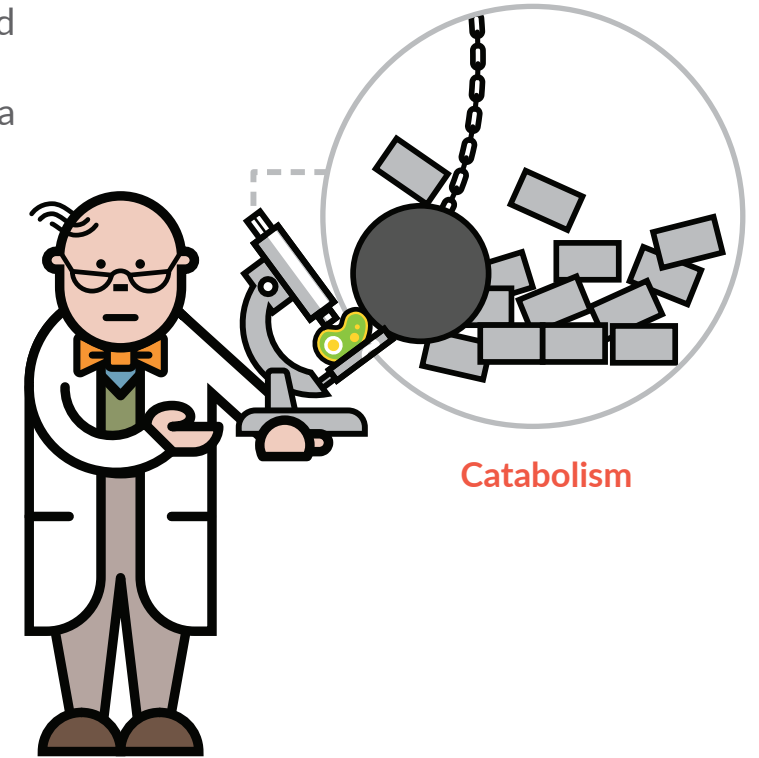
It may cause delays to normal development like walking and talking.



## What else happens in MMA?

If the body does not receive enough food e.g. during illness or the body has gone without food for too long, there may be a shortage of energy supply.

This causes **catabolism** which is a break down of body protein and can lead to a metabolic crisis.





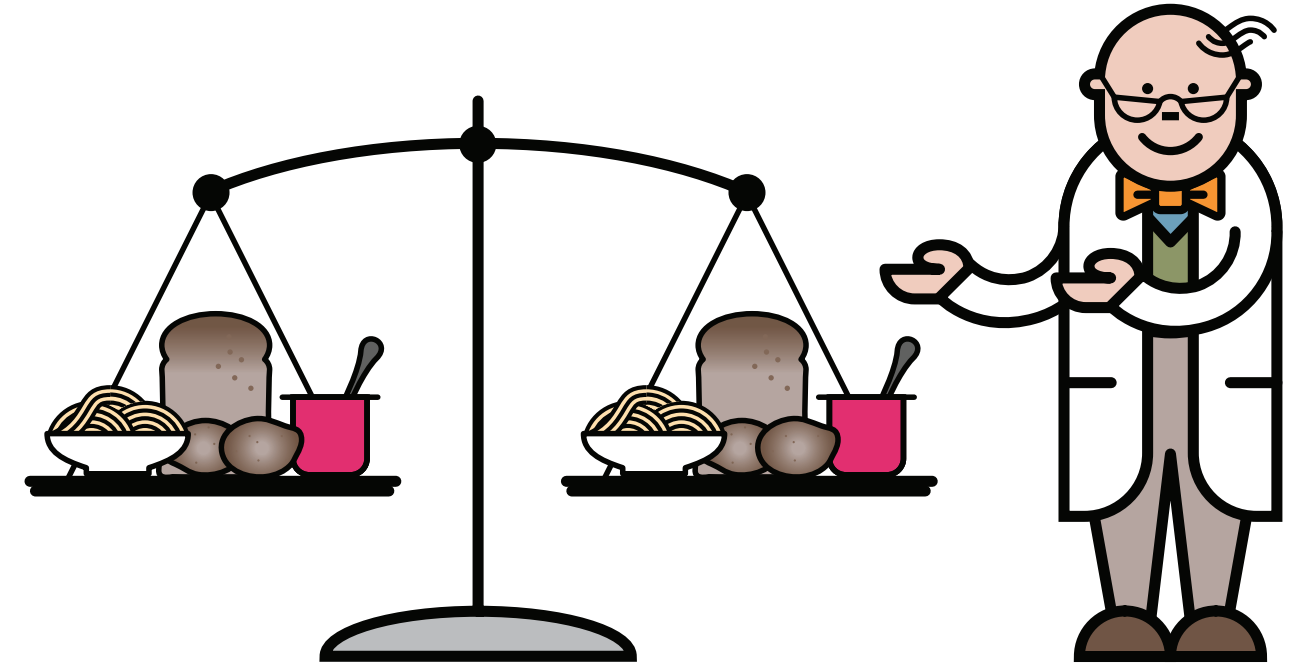
# Metabolic crisis

- In a **metabolic crisis** there is a build up of methylmalonic acid and other toxic chemicals such as ammonia
- It is usually triggered by childhood illnesses e.g. vomiting and diarrhoea, fasting for too long or not having enough energy from food
- There should be no delay in management
- Avoidance of a metabolic crisis is essential



# Protein balance is needed in MMA

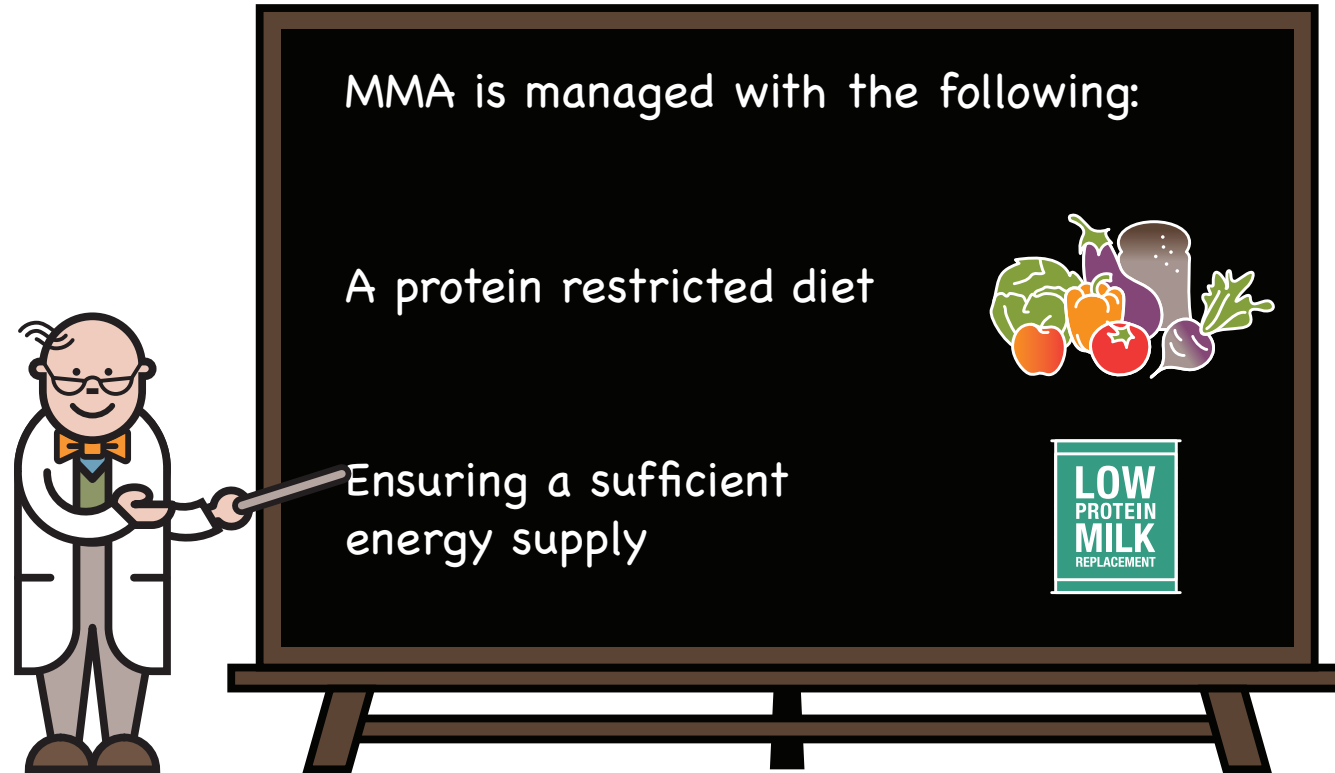
In MMA, it is important that enough protein is given for growth ... but not too much as toxic chemicals will be made.



## How is MMA managed day to day?

MMA is managed with the following:

- A protein restricted diet
- Ensuring a sufficient energy supply



The illustration shows a cartoon doctor with glasses and a bow tie pointing to a chalkboard. The chalkboard lists two management strategies for MMA. To the right of the text, there is an illustration of various fruits and vegetables, and below that, a box labeled 'LOW PROTEIN MILK REPLACEMENT'.

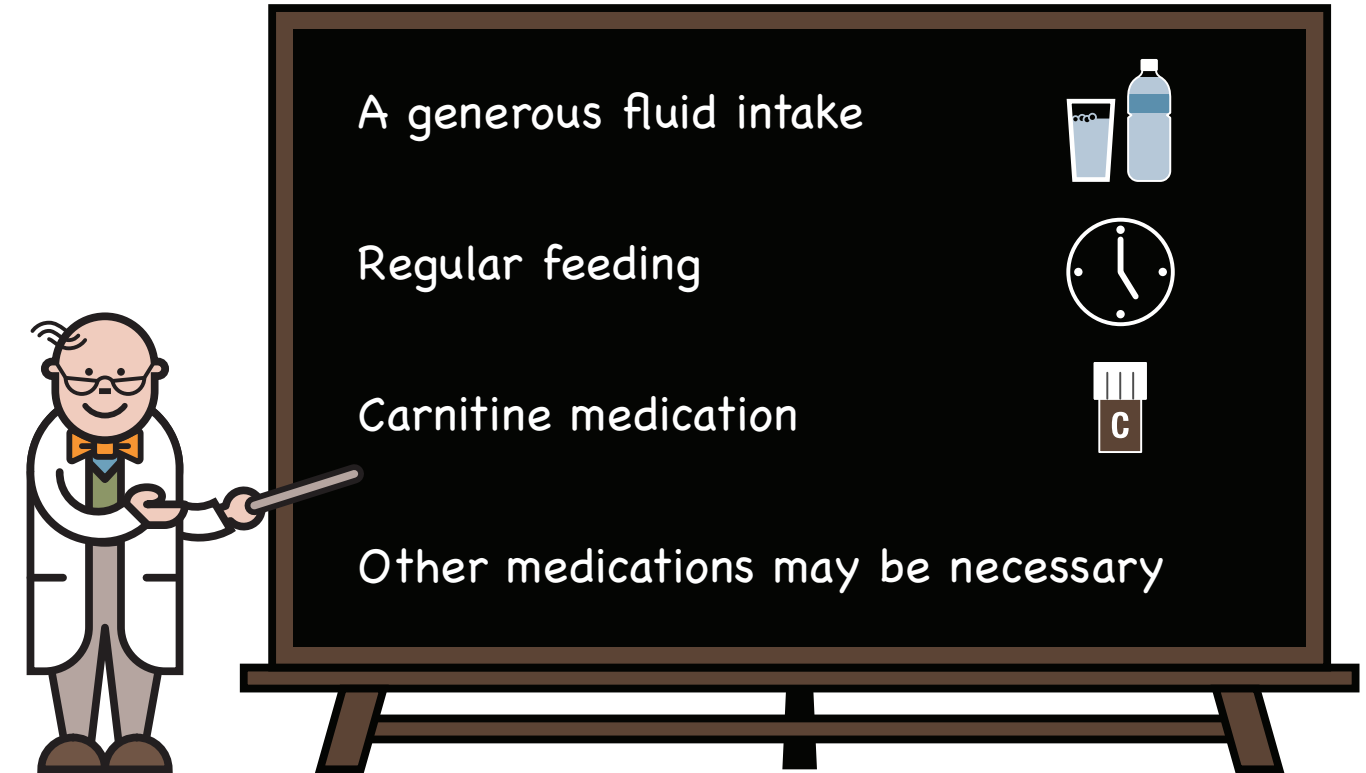
## How is MMA managed day to day?

A generous fluid intake

Regular feeding

Carnitine medication

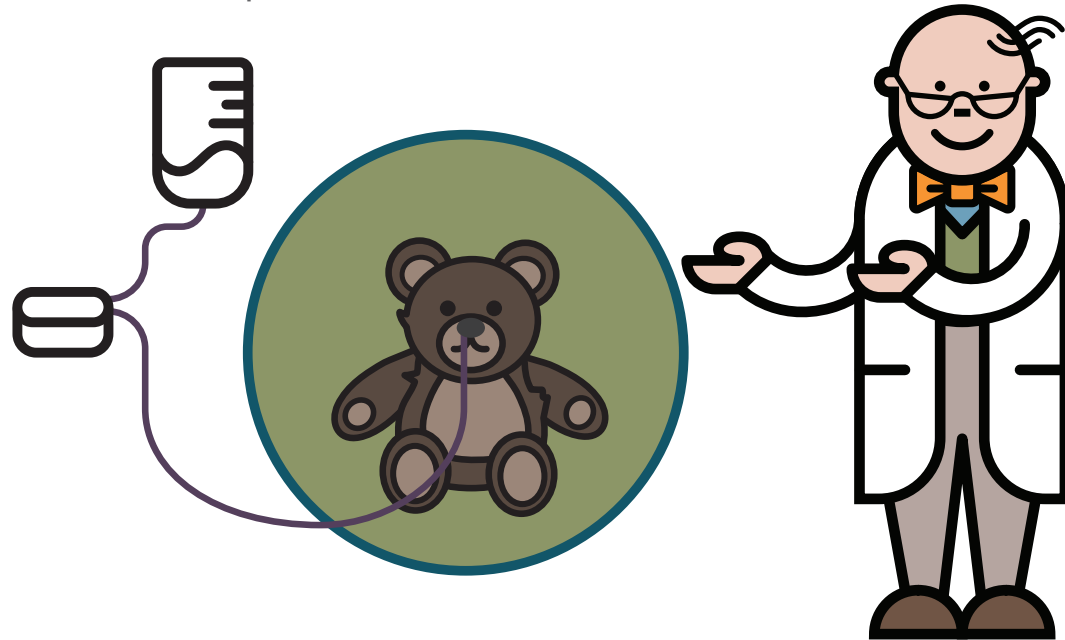
Other medications may be necessary



The illustration shows a cartoon doctor with glasses and a bow tie pointing to a chalkboard. The chalkboard lists four management strategies for MMA. To the right of the text, there are three icons: a glass and a water bottle for fluid intake, a clock for regular feeding, and a pill bottle labeled 'C' for carnitine medication.

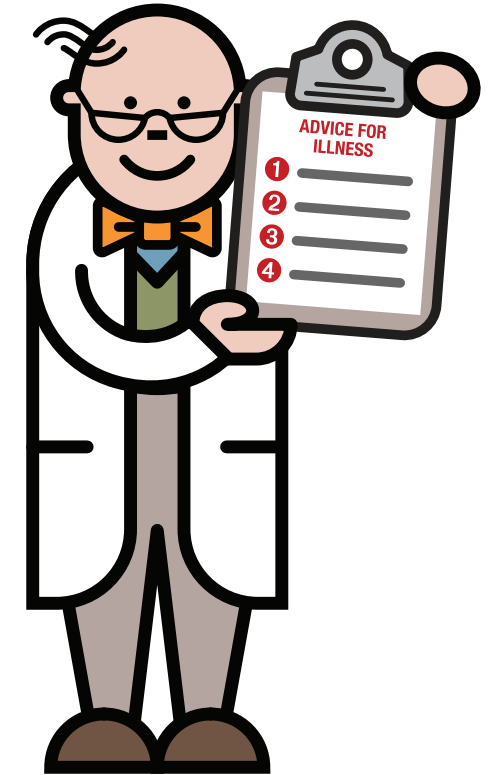
## Is tube feeding needed?

Tube feeding may be necessary to give regular feeds. This will ensure energy, nutrient and fluid needs are met and can help to reduce the production of abnormal chemicals.

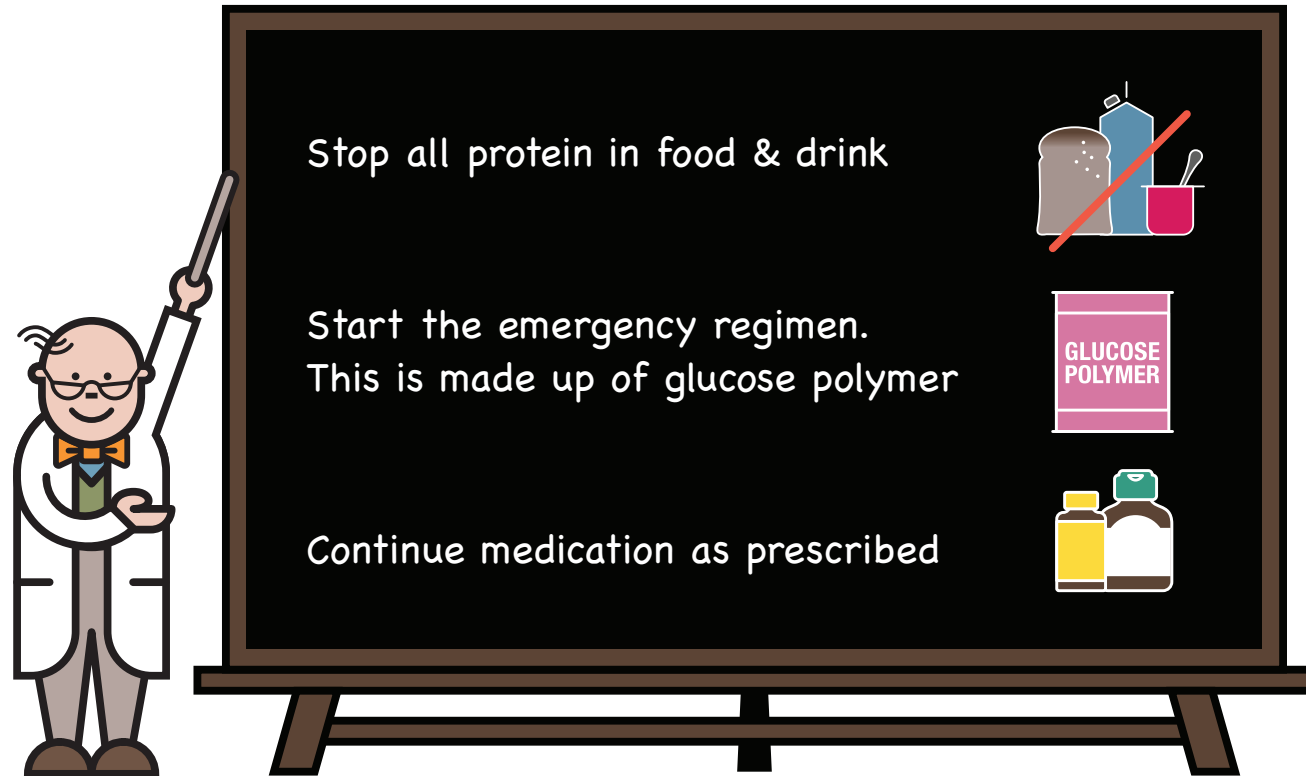


## How is MMA managed during illness?

- During any childhood illness, an emergency regimen is given
- This is to avoid a lack of energy supply and build-up of harmful chemicals that cause a metabolic crisis






## How is MMA managed during illness?



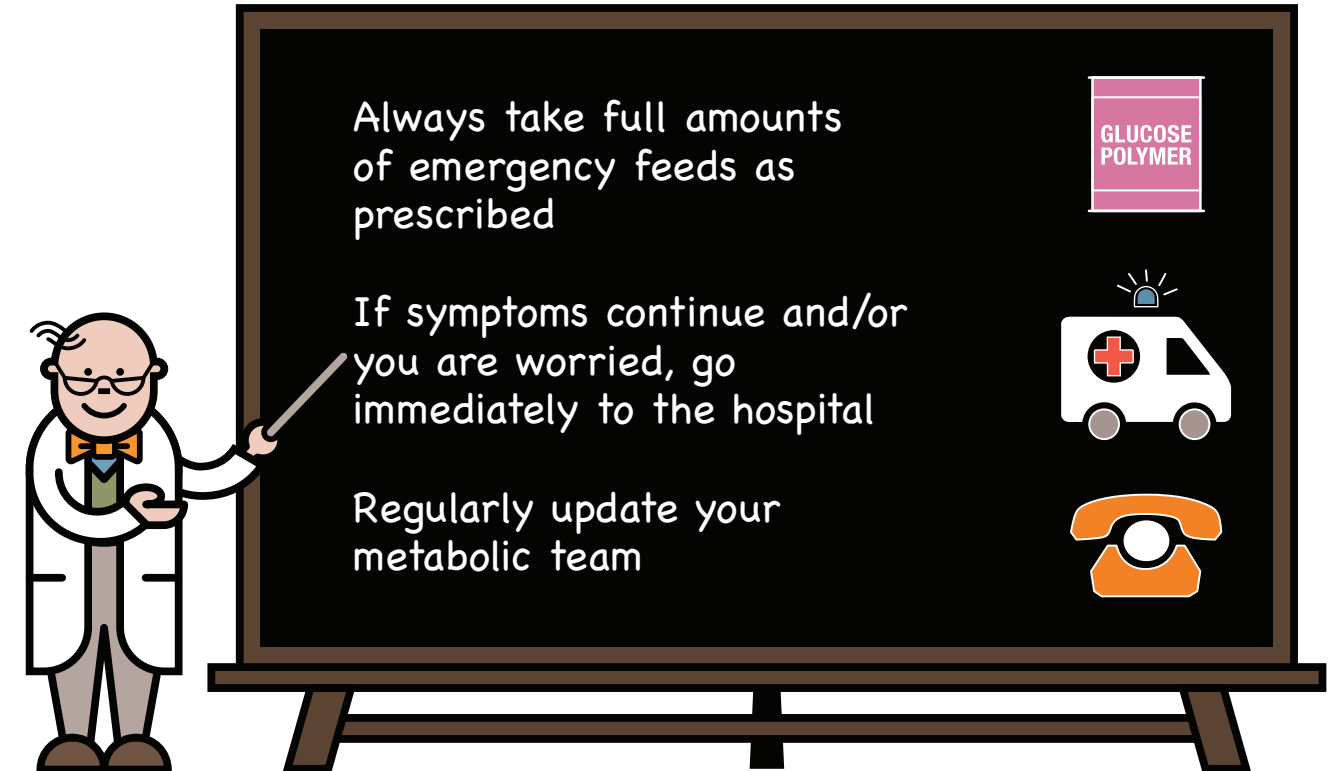
Stop all protein in food & drink

Start the emergency regimen.  
This is made up of glucose polymer

Continue medication as prescribed






## Checklist for illness



Always take full amounts  
of emergency feeds as  
prescribed

If symptoms continue and/or  
you are worried, go  
immediately to the hospital

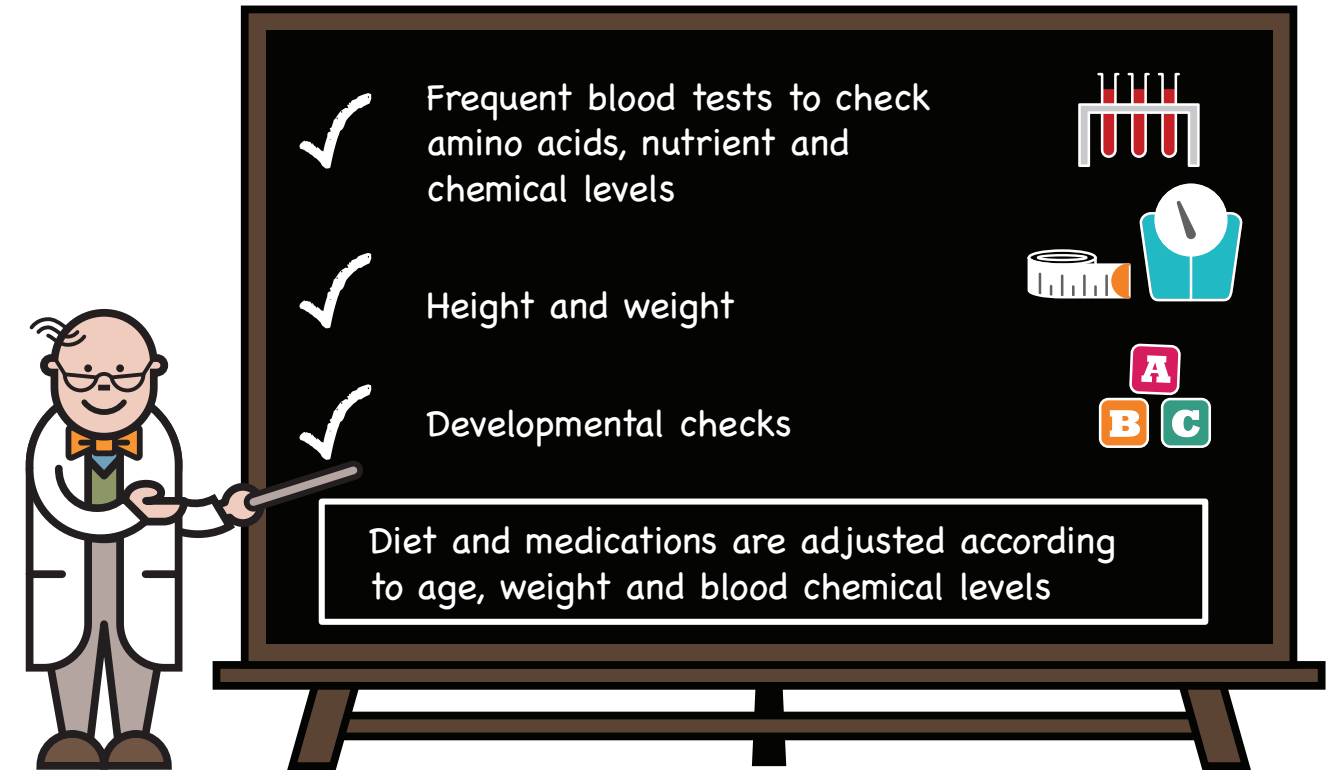
Regularly update your  
metabolic team



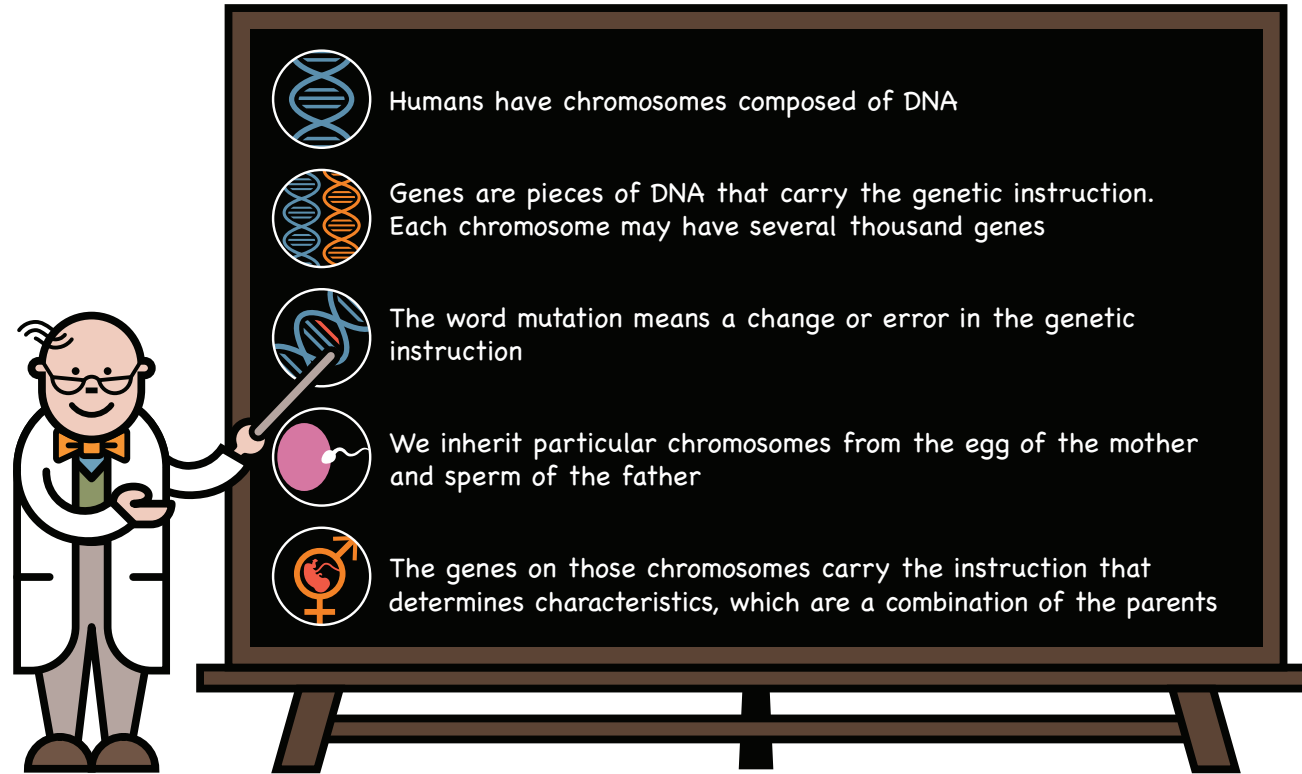
## Key message



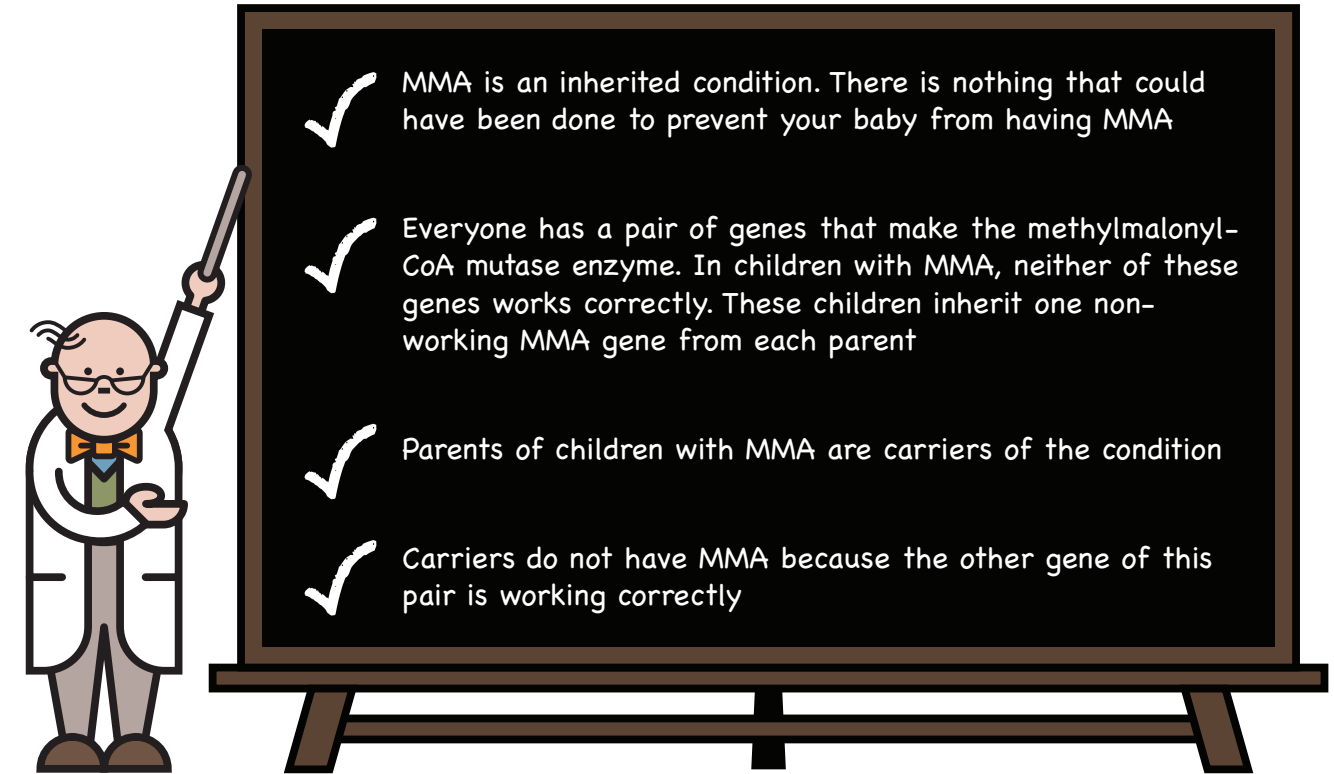
## How is MMA monitored?



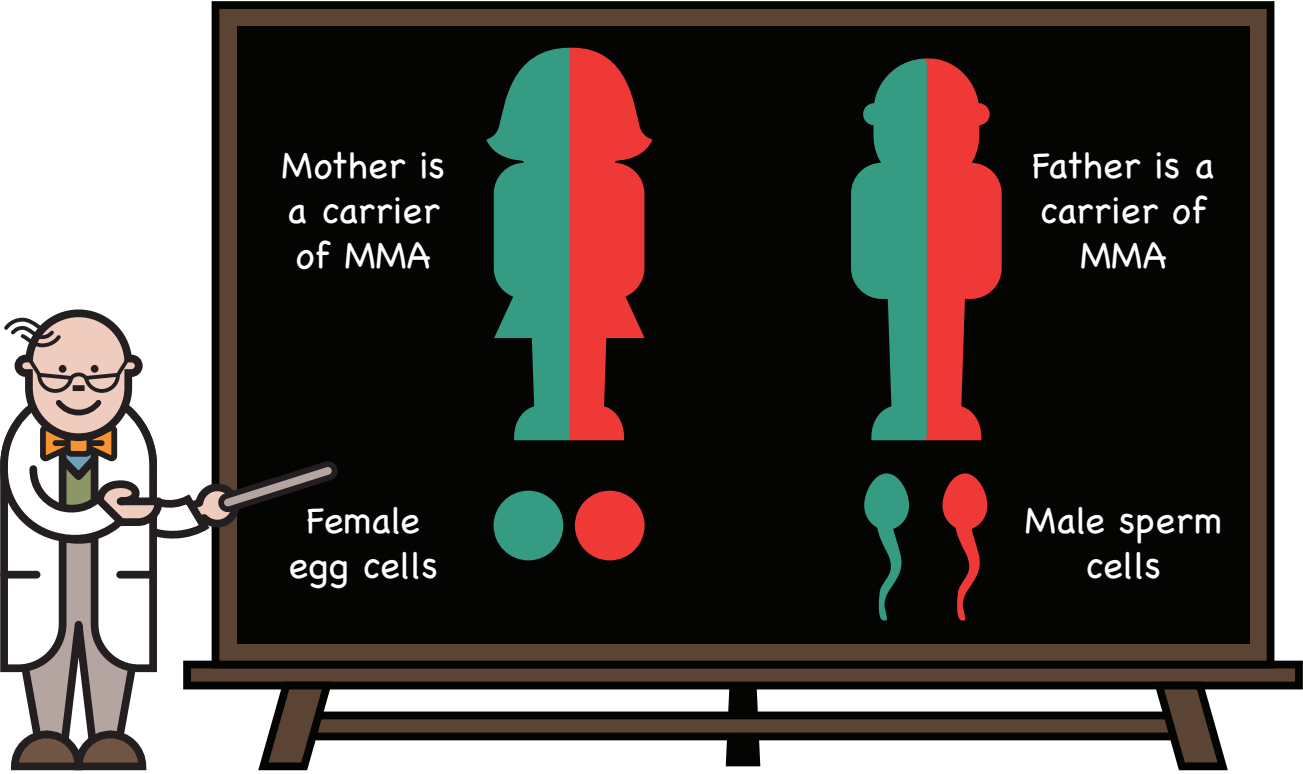
# Chromosomes, genes, mutations



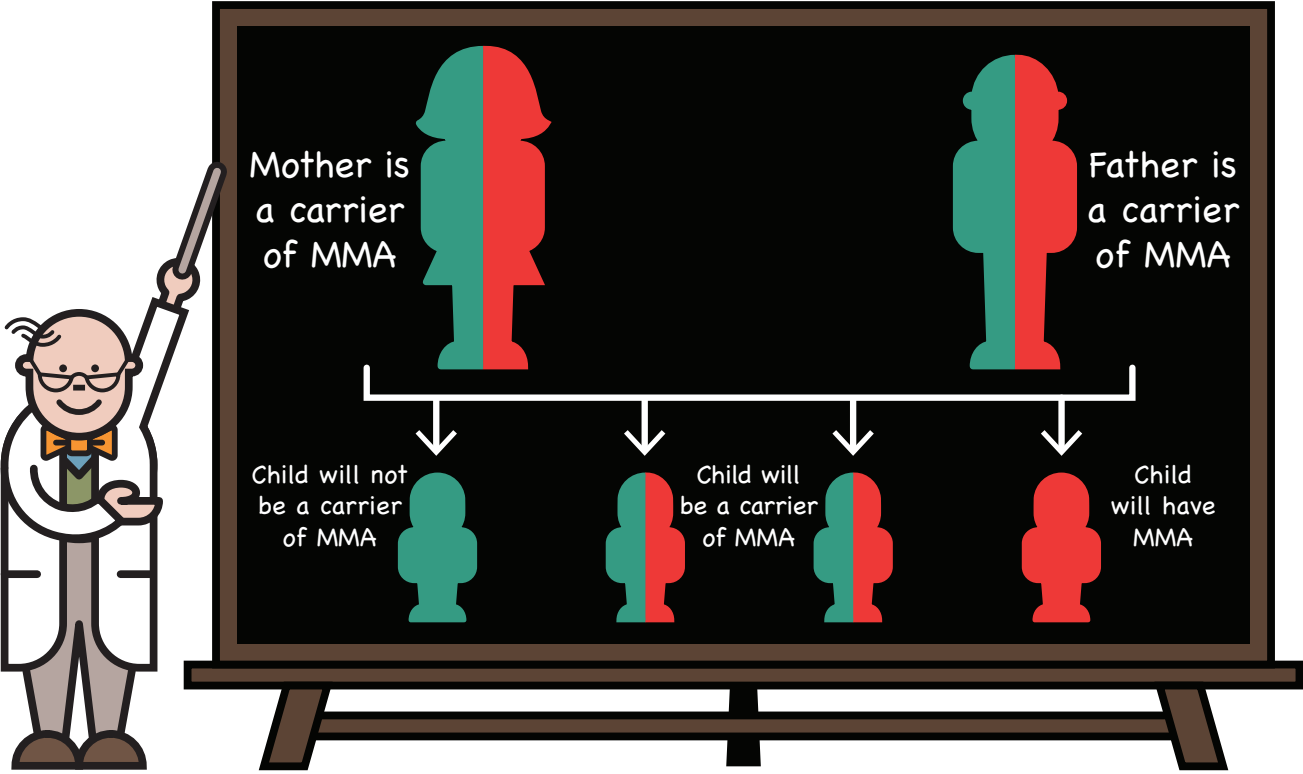
# Inheritance



Inheritance — Autosomal-recessive (carriers of MMA)

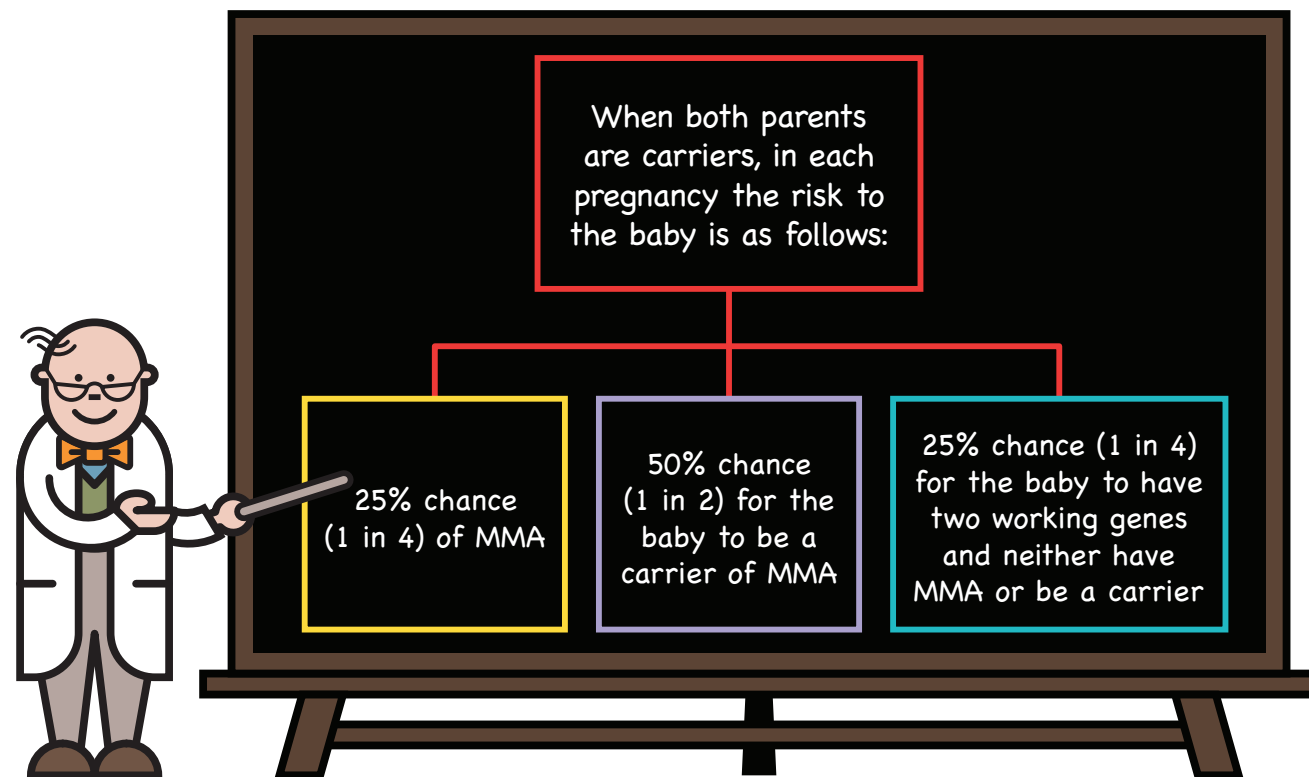


Inheritance — Autosomal recessive – possible combinations

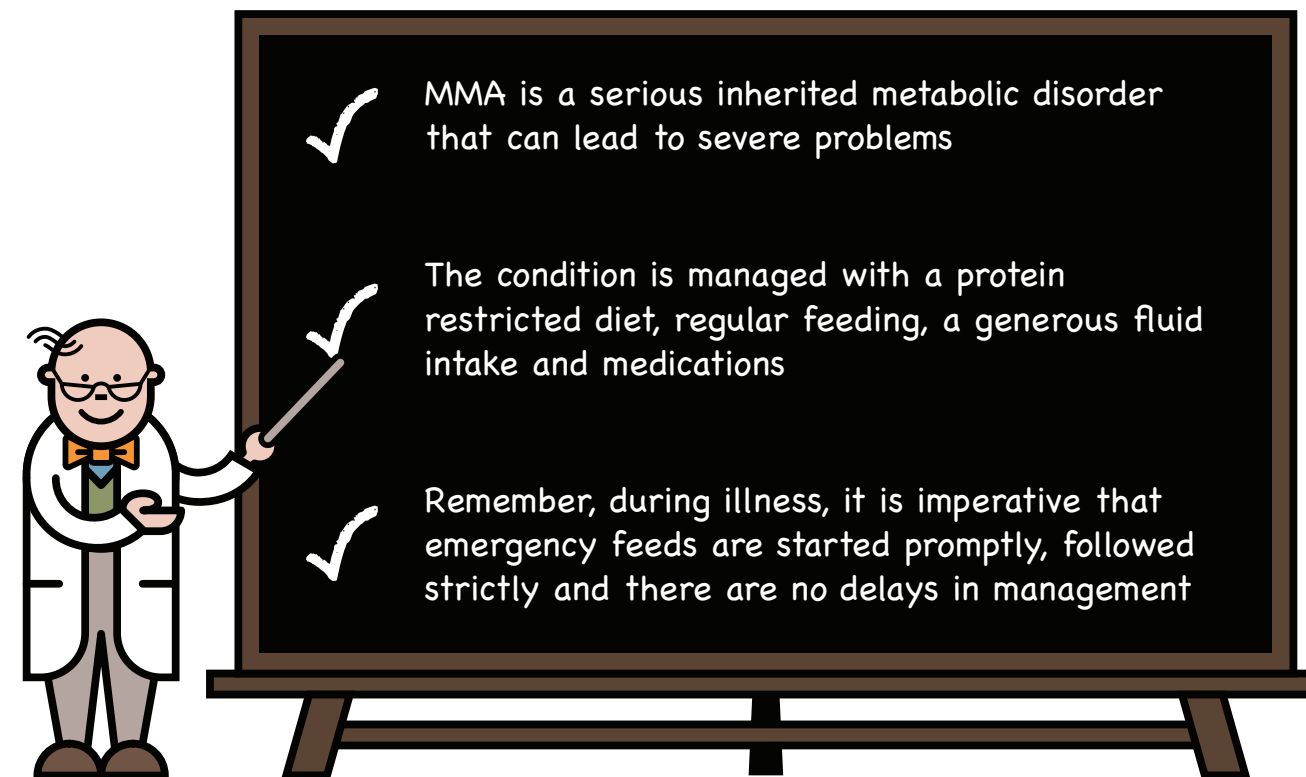




## Future pregnancies



## Take home messages



## Helpful hints



## Who's who

- My dietitians
- My nurses
- My doctors
  - Contact details, address, photos

## Notes

This image shows a blank sheet of white paper designed for handwriting practice. It features two vertical columns of horizontal dashed lines. Each column contains ten rows of these lines, providing a guide for letter height and placement. The lines are evenly spaced and extend across the width of each respective column.

## Notes

[illegible]

Visit [www.nutricia.co.uk/patients-carers/living-with/low-protein-diet.html](http://www.nutricia.co.uk/patients-carers/living-with/low-protein-diet.html) and register to get access to support and practical advice for those living on a low protein diet.

The site also provides information on upcoming events and personal stories from others on a low protein diet.

MET1855UKIR



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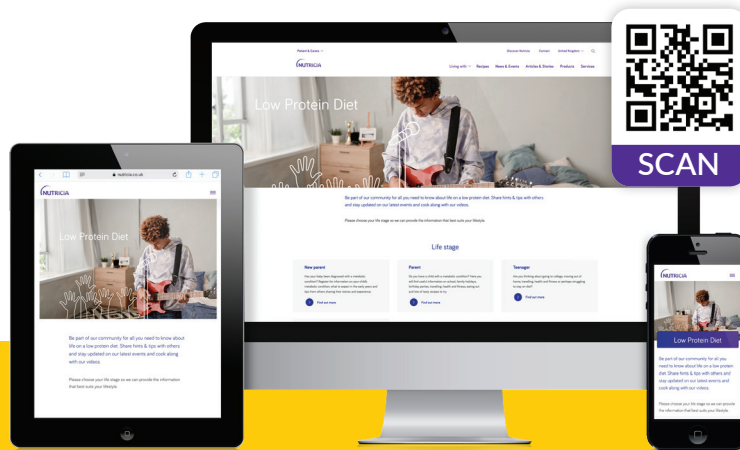
LowProteinConnect



LowProteinConnect



SCAN



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