TEMPLE

Tools **E**nabling **M**etabolic **P**arents **LE**arning

LCHADD

ADAPTED BY THE DIETITIANS GROUP

BIMDG
British Inherited Metabolic Diseases Group

BASED ON THE ORIGINAL TEMPLE WRITTEN BY
BURGARD AND WENDEL

VERSION 4, MARCH 2025

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.

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).

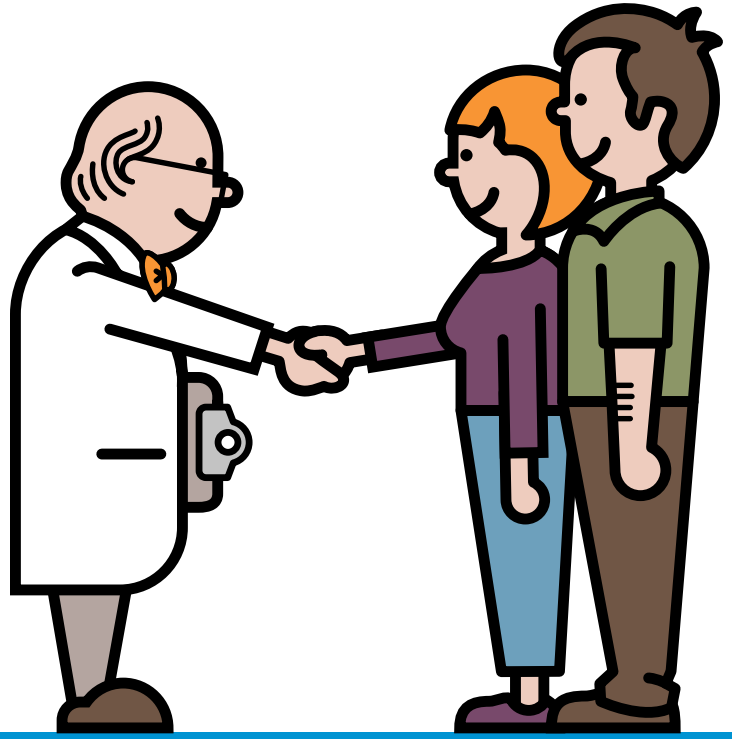
The team are Rachel Skeath, Karen van Wyk, Pat Portnoi and Anita MacDonald. The group is facilitated by Heidi Chan from Nutricia.

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

They are not designed to replace dietary information that may be given by a dietitian in clinic.

LCHAD Deficiency

Information for families following a new diagnosis



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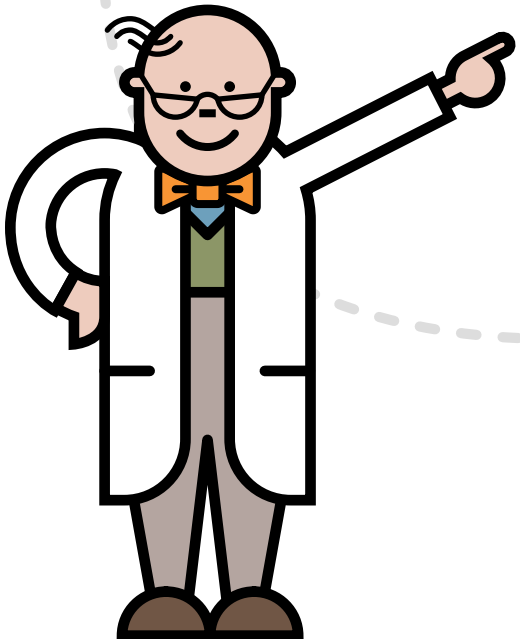
What is LCHADD?

LCHADD stands for long-chain 3-hydroxyacyl-CoA dehydrogenase deficiency

It is an inherited metabolic condition

Long-Chain | 3-Hydroxy | Acyl-CoA | Dehydrogenase deficiency

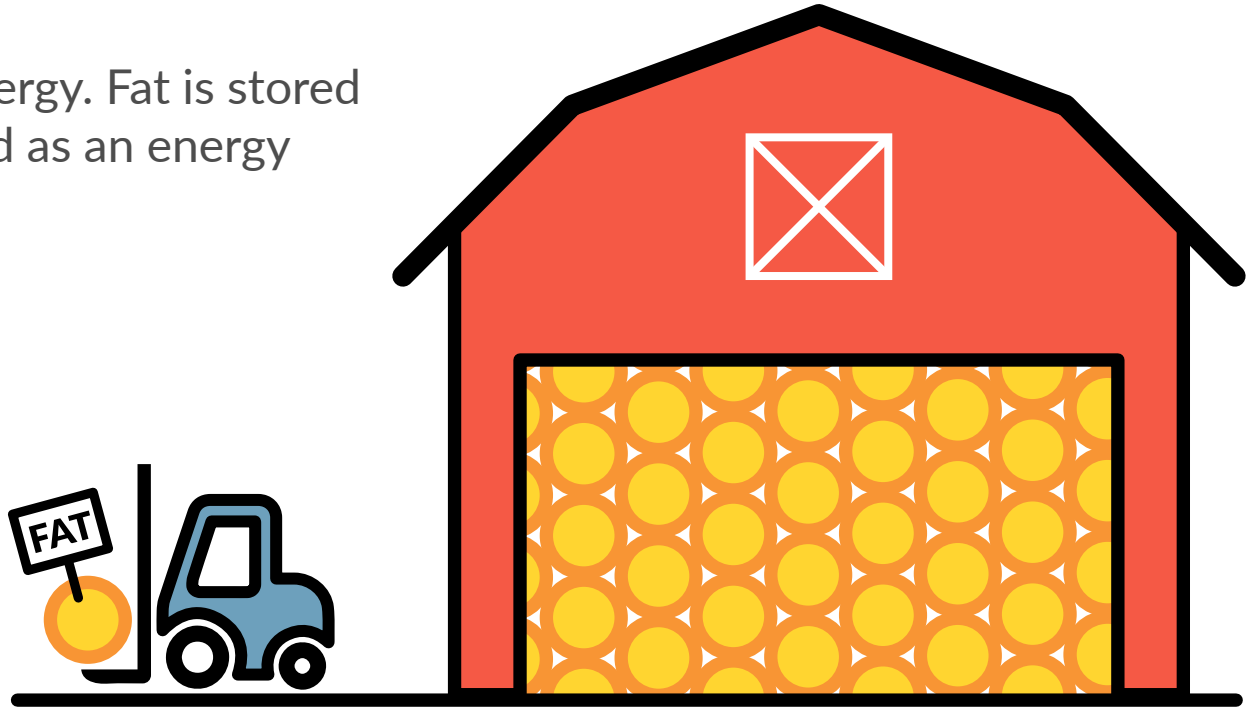


LCHAD Deficiency

Which foods supply the body with energy?

There are two main food groups that supply the body with energy:

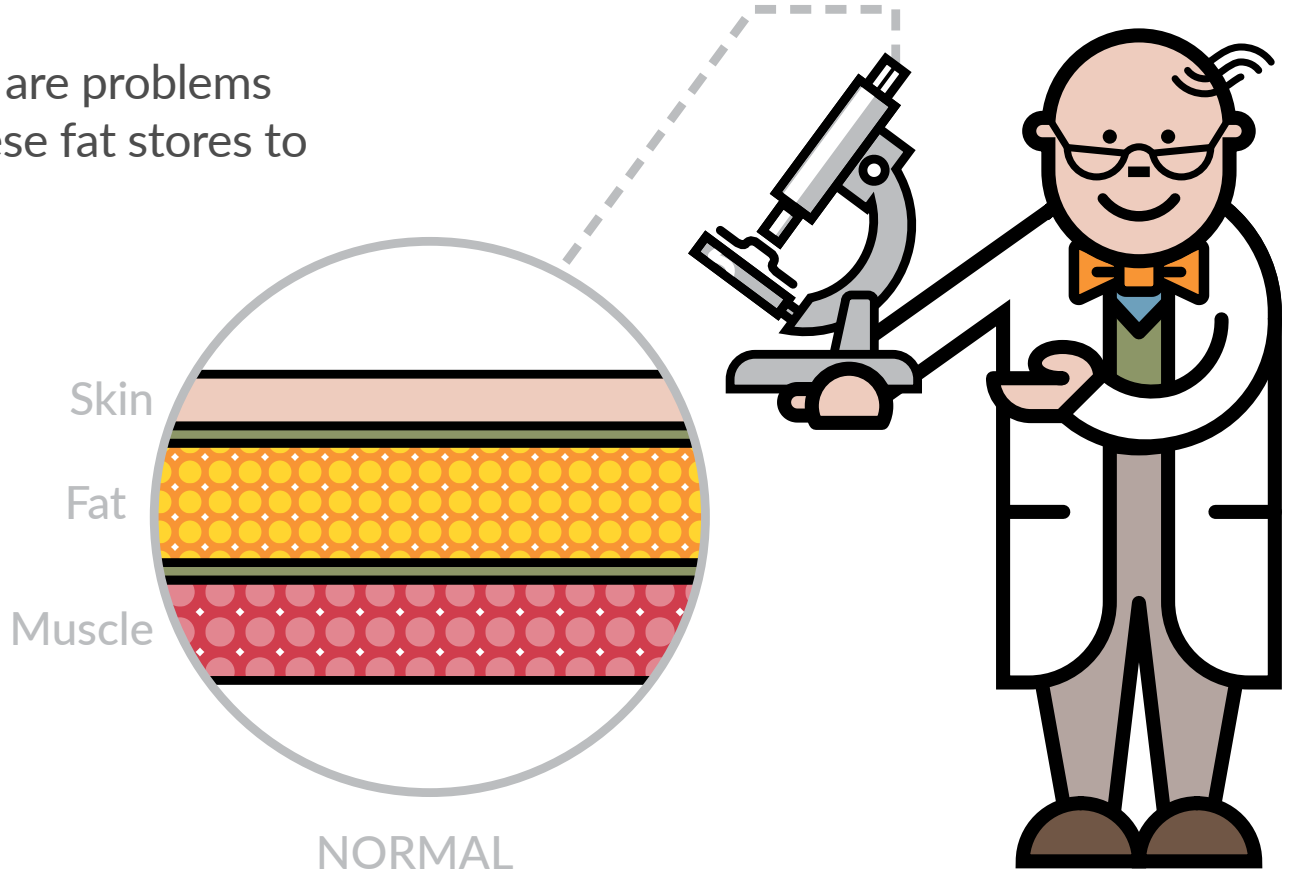
- Carbohydrates (starches and sugars) provide a readily available energy source
- Fats also provide energy. Fat is stored in the body and used as an energy reserve



LCHADD and fat

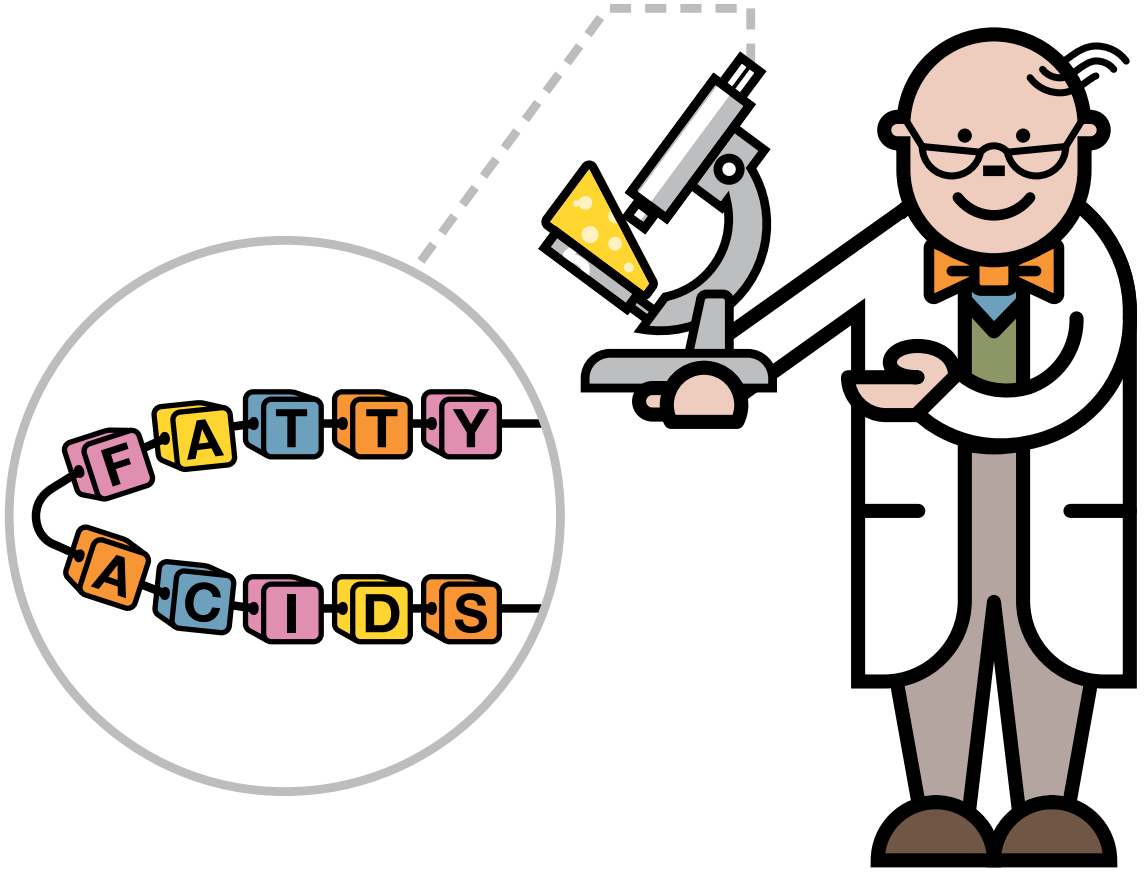
The body uses its own fat stores to provide energy when carbohydrate is depleted.

In LCHADD, there are problems breaking down these fat stores to release energy.



Breaking down fat stores for energy

Body fat stores are broken down into fatty acids.



What are fatty acids?

Fatty acids are made up of carbon atoms joined together to form chains of many different lengths.

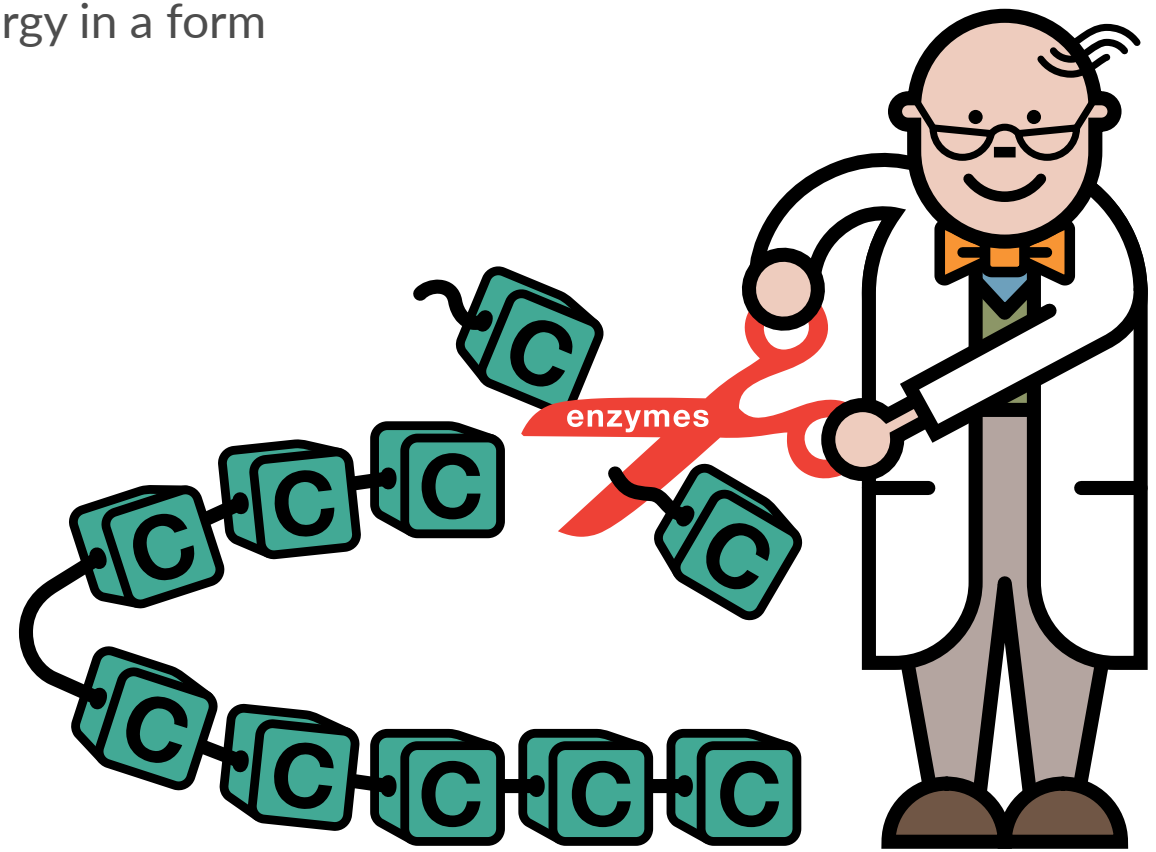
Short chain

Medium chain

Long chain

Fatty acids and enzymes

Fatty acid chains are then broken down into smaller units by enzymes (which are like chemical scissors). This enables the body to produce energy in a form which it can use.

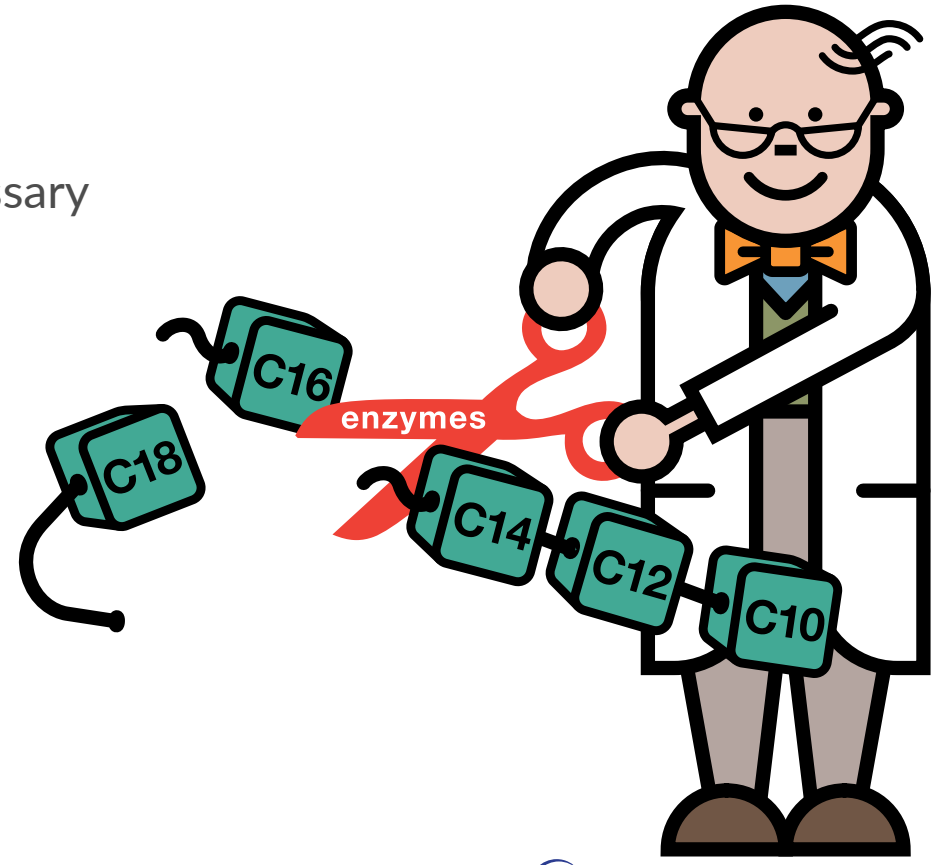


What happens in LCHADD?

In LCHADD, the body lacks an enzyme that helps convert fat stores into energy.

This **stops** the long chain fats from being used for energy.

There is a problem when it is necessary to break down fats quickly.



What can go wrong in LCHADD?

There can be a shortage of energy supply and a build up of harmful chemicals during illnesses or with lack of food.

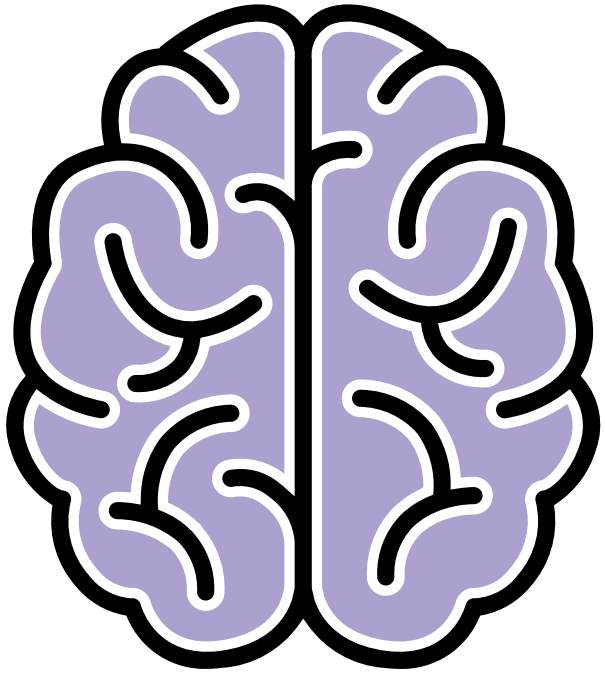
Symptoms include:

- poor feeding
- excessive sleepiness
- rapid breathing
- seizures
- low blood sugar
- floppiness
- enlarged heart
- heartbeat abnormalities
- liver problems

What can go wrong in LCHADD?

If there is a shortage of energy and this is not corrected, it can lead to coma and brain damage and it may be life threatening.

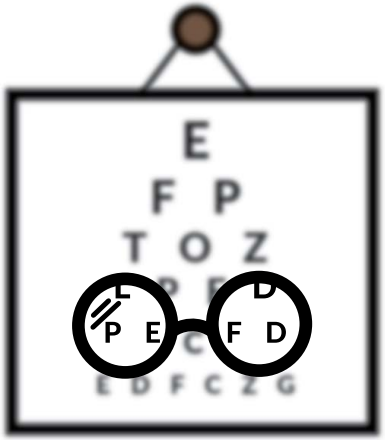
However, please remember, this can all be prevented with timely and correct management.

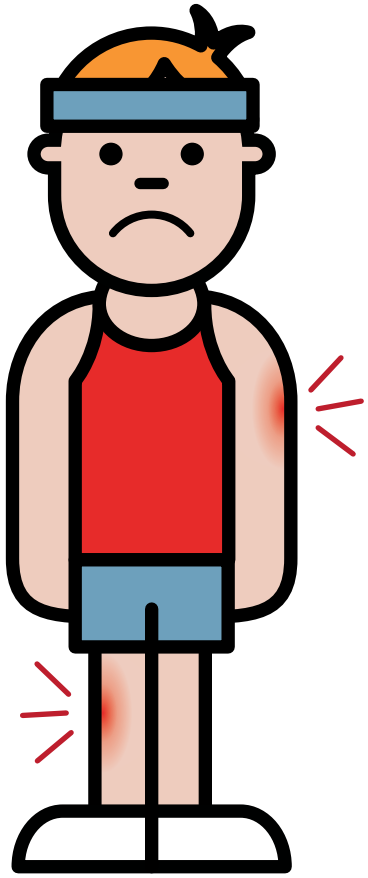


What else happens in LCHADD

It may also cause

- learning difficulties
- delays in walking
- muscle pain especially with exercise
- vision loss
- nerve problems in arm and legs
- enlarged heart and breathing difficulties





Metabolic crisis

- A **metabolic crisis** triggers the LCHADD symptoms
- There is a lack of energy and build up of toxic chemicals
- It is usually triggered by childhood infections causing high temperatures, vomiting, and diarrhoea
- It can also be caused by going for a long time without food
- Avoidance of a metabolic crisis is essential

How is LCHADD diagnosed?

LCHADD is suspected because of the pattern of chemicals (acylcarnitines and organic acids) found in the blood and urine.

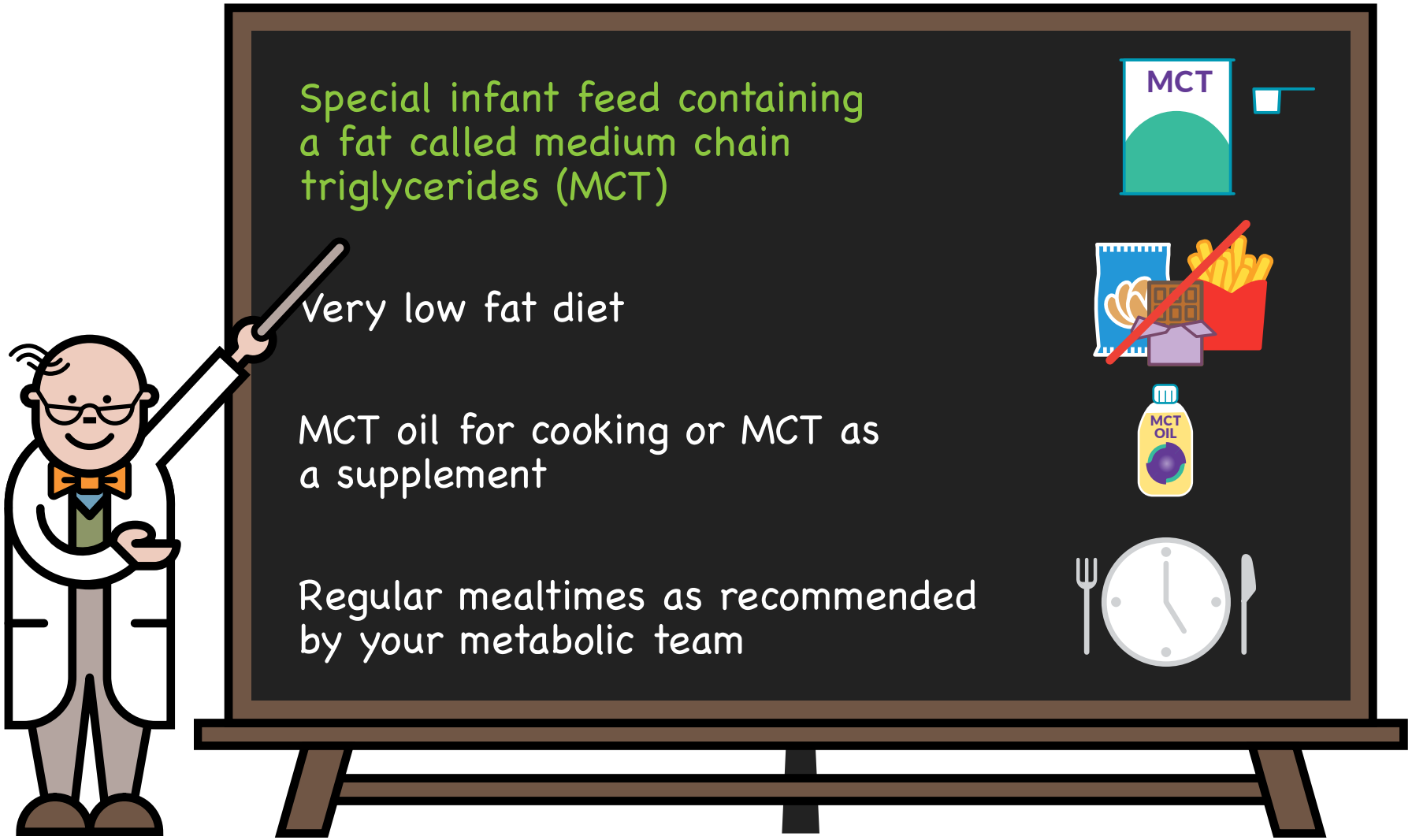
The diagnosis is confirmed by finding mutations in the genes for LCHAD.

How is LCHADD managed?

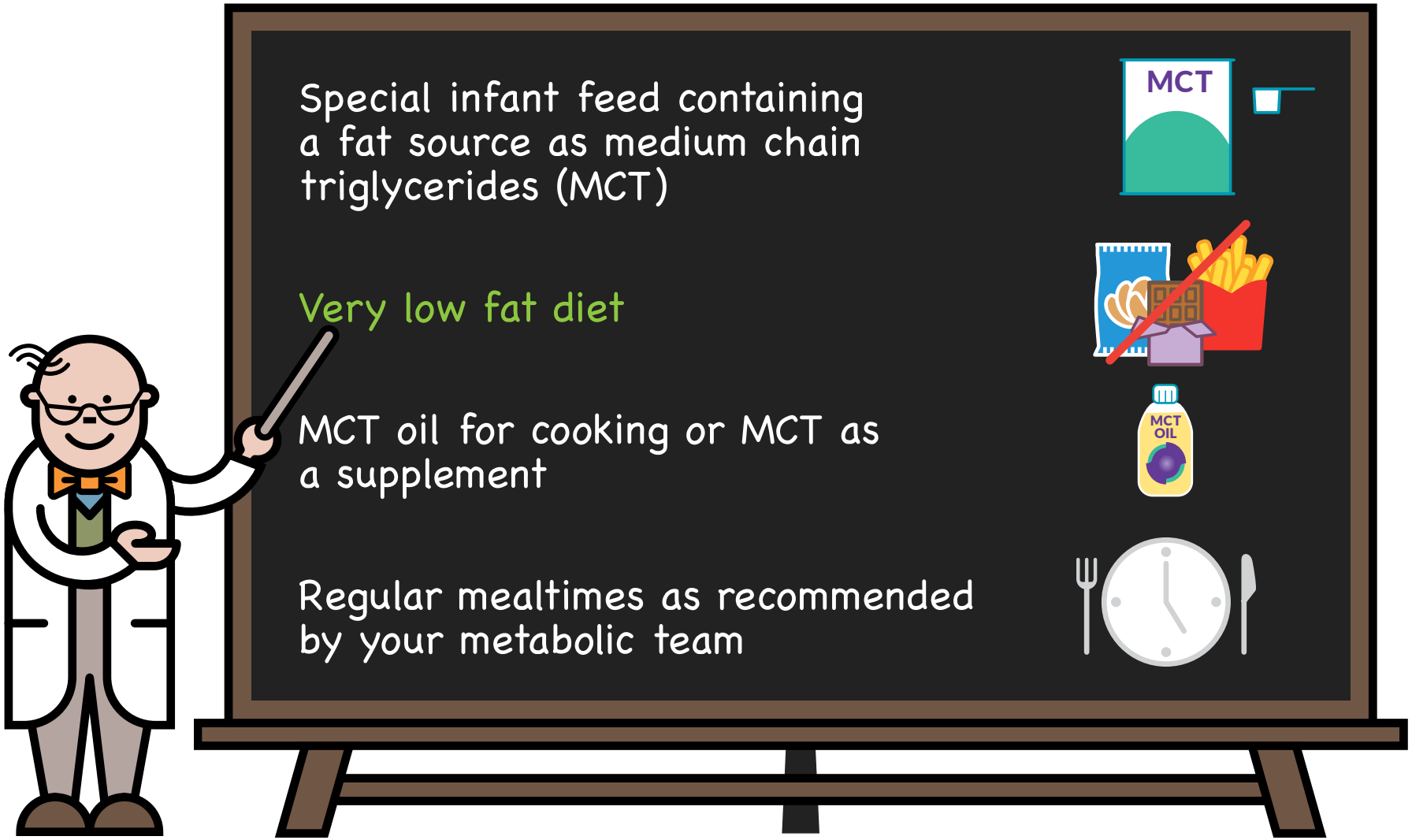
In babies, it is managed day to day by **avoiding** long periods without feeding, even when well.

The length of time babies can go without feeds is known as **the safe fasting time**.

How is LCHADD managed day to day?



How is LCHADD managed day to day?



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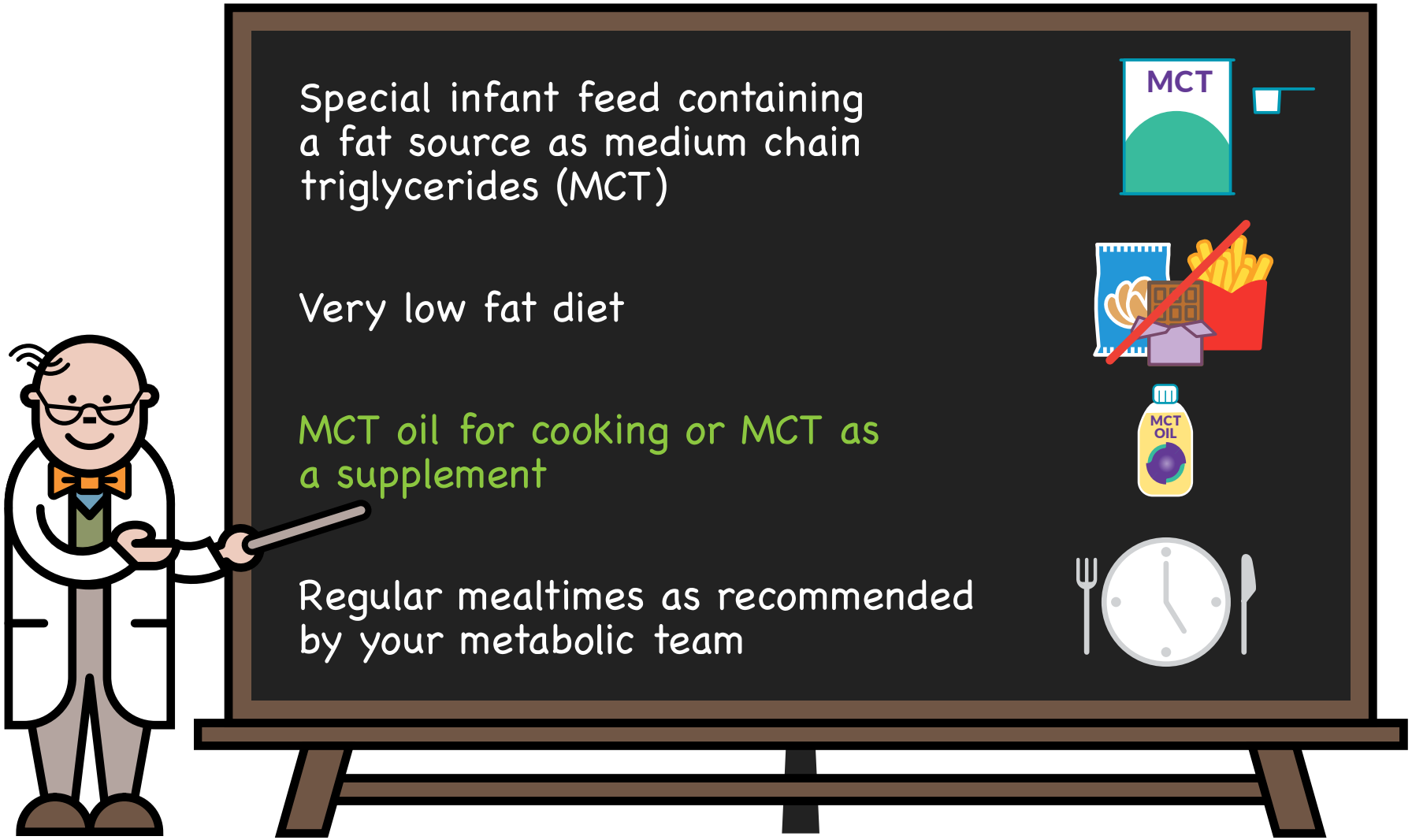

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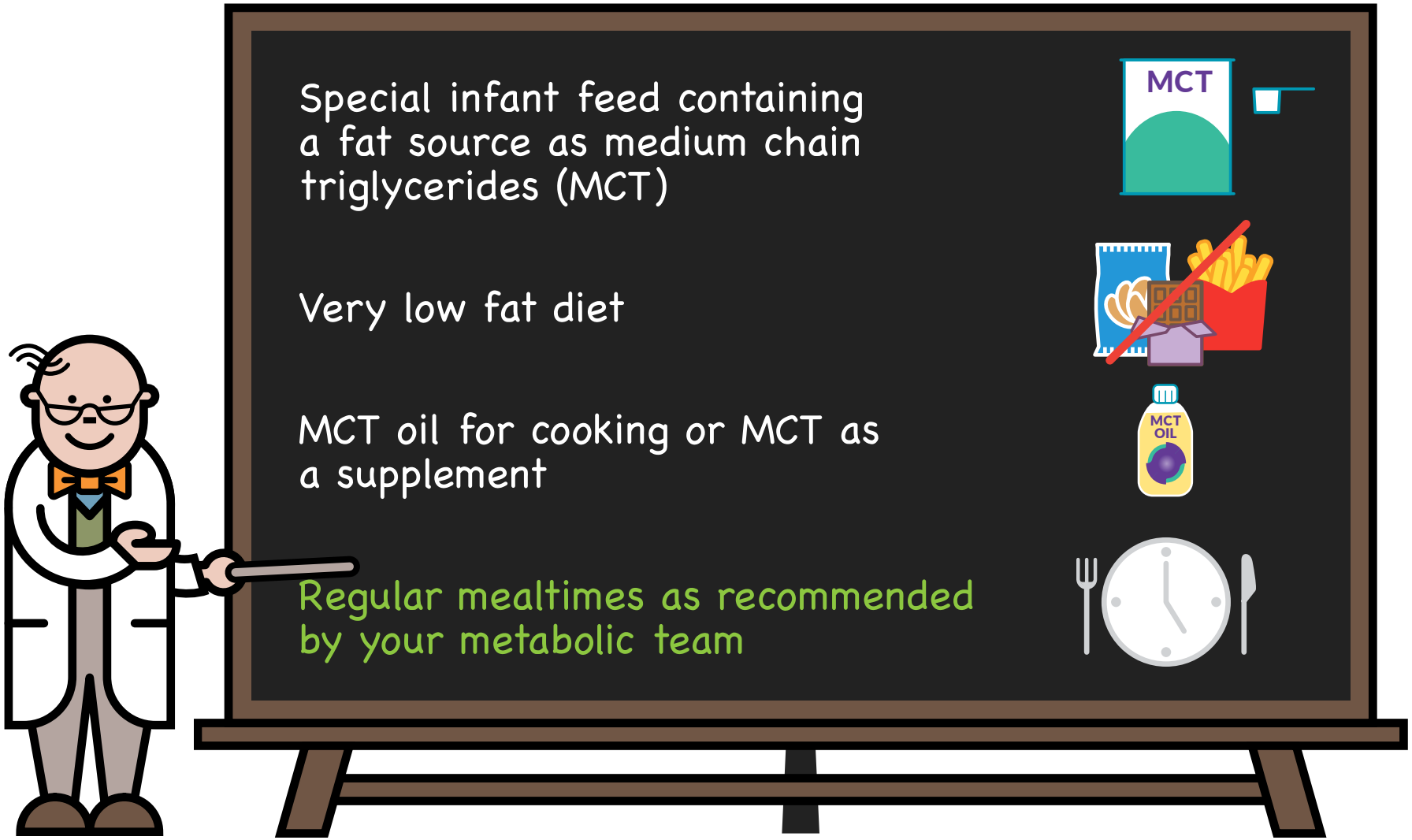
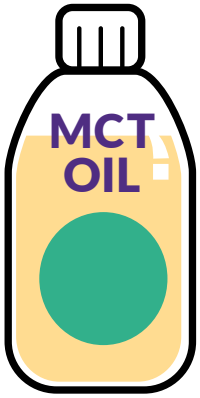


LCHADD and fat

The diet needs to be very low in fat.

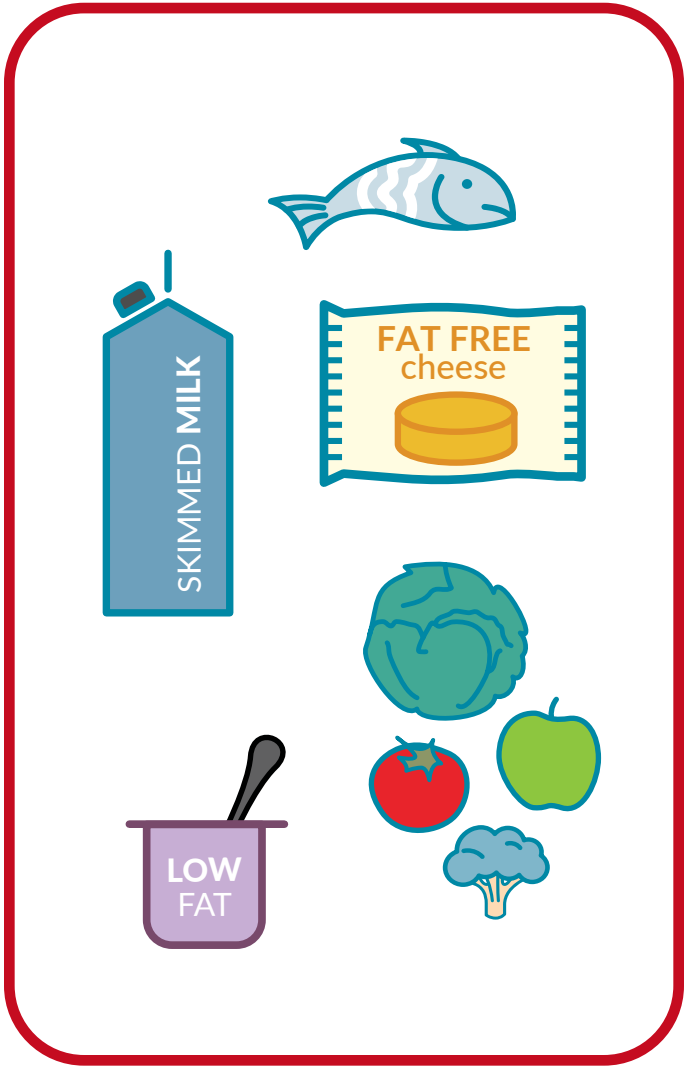
Foods high in fat are avoided.
Many foods are high in fat e.g. full fat milk, full fat cheese, fatty meat, eggs, ice cream, chips, crisps and chocolate.



Very low fat foods

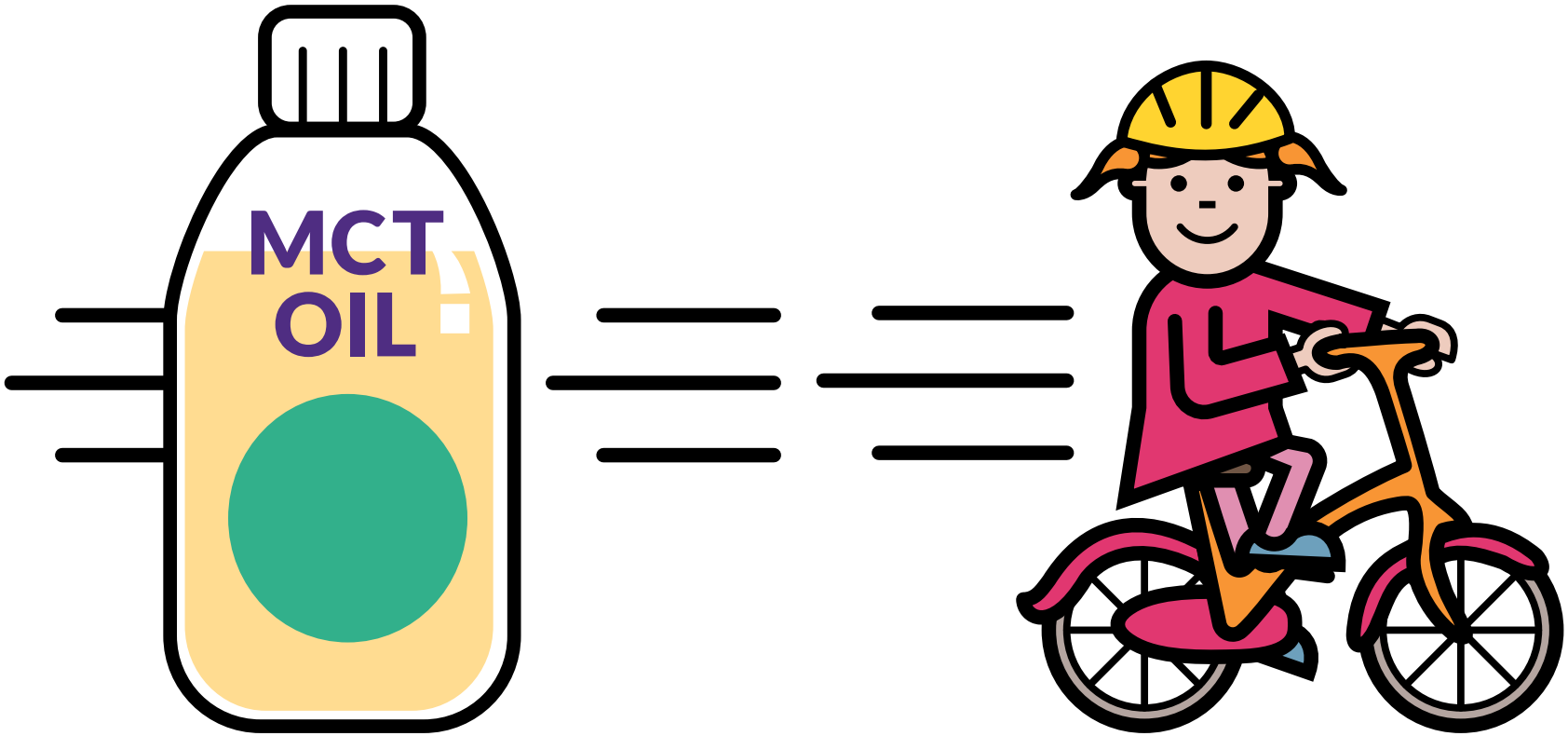
The diet needs to be very low in fat.

Very low fat foods are permitted.
There is a wide range of very low fat foods available e.g. skimmed milk, very low fat yoghurt/cheese, white fish, fruit and vegetables.



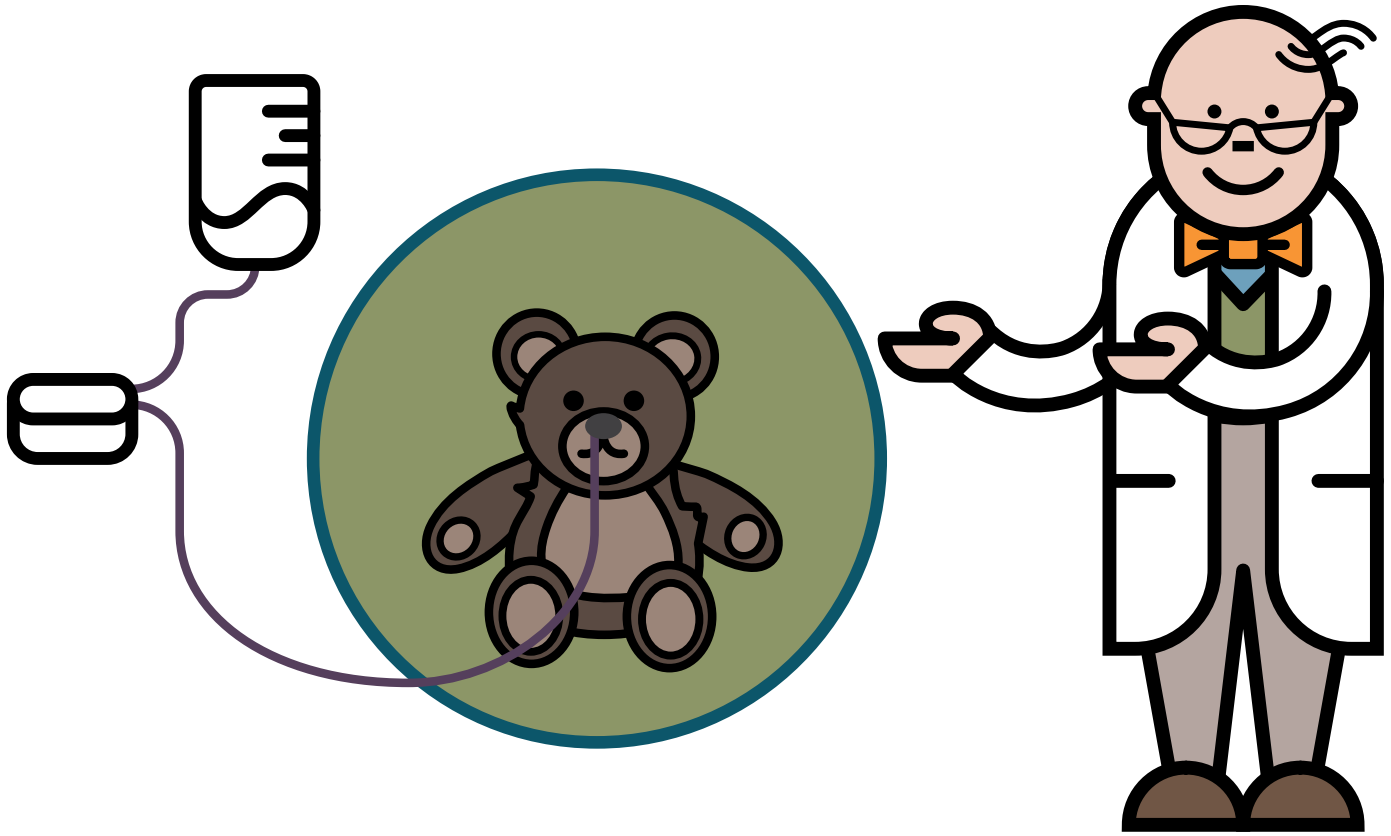
LCHADD and exercise

It is advisable to take a source of MCT, or a high sugar snack or drink before exercise.
This will supply an extra source of energy.



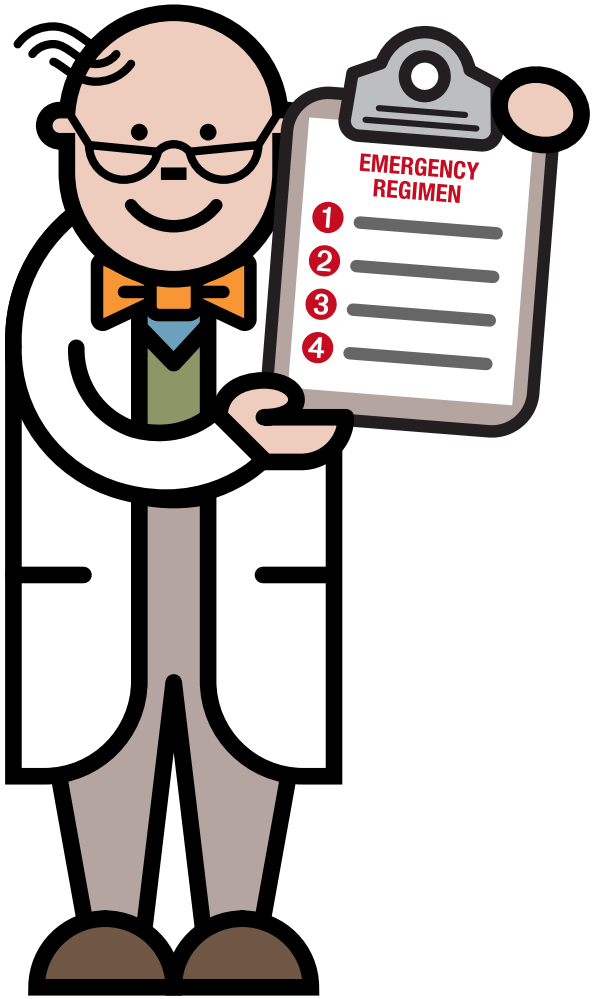
Is tube feeding needed?

In babies with the most severe forms of LCHADD, tube feeding may be necessary. This will ensure energy, nutrient and fluid needs are met.

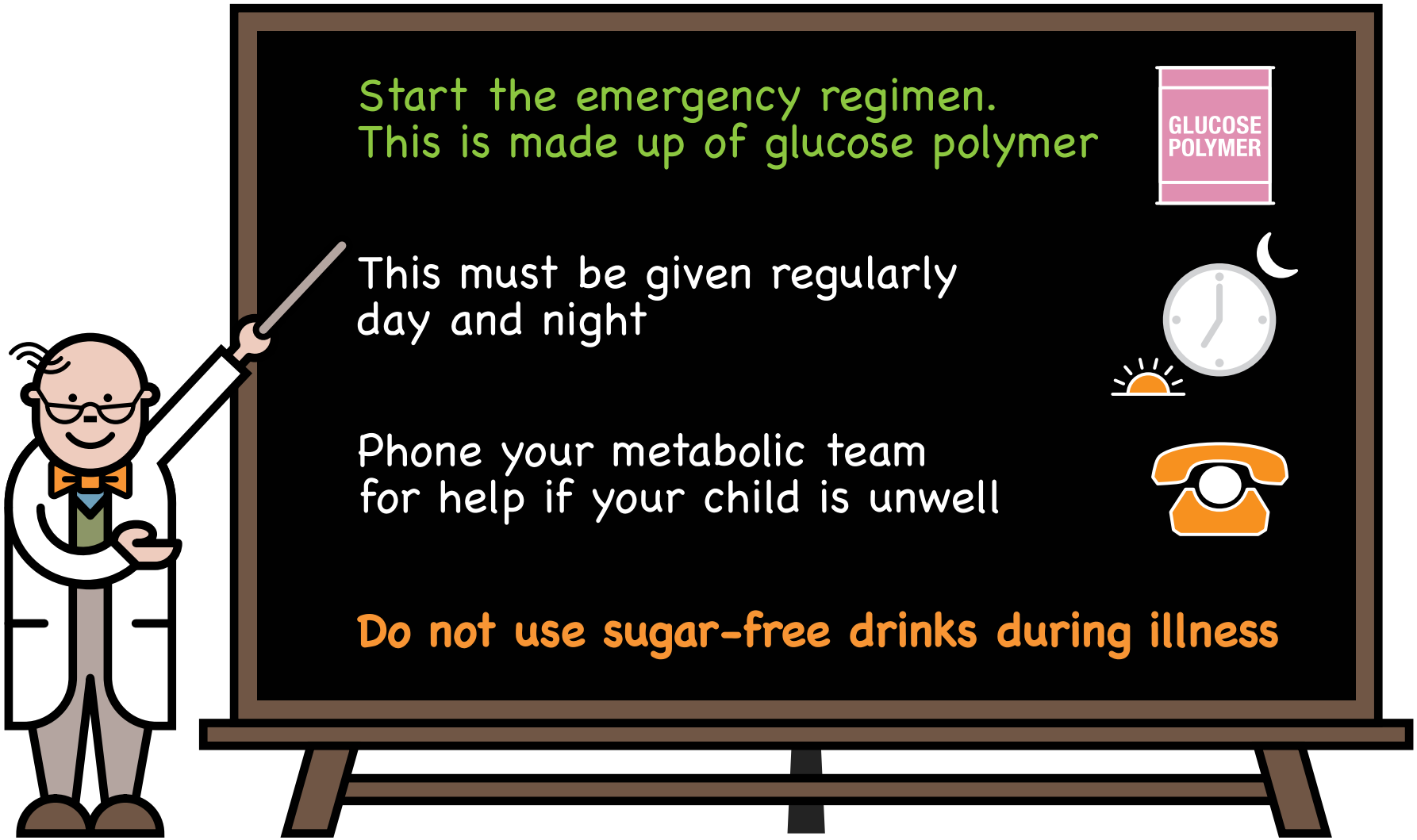


How is LCHADD managed during illness?

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



How is LCHADD managed during illness?



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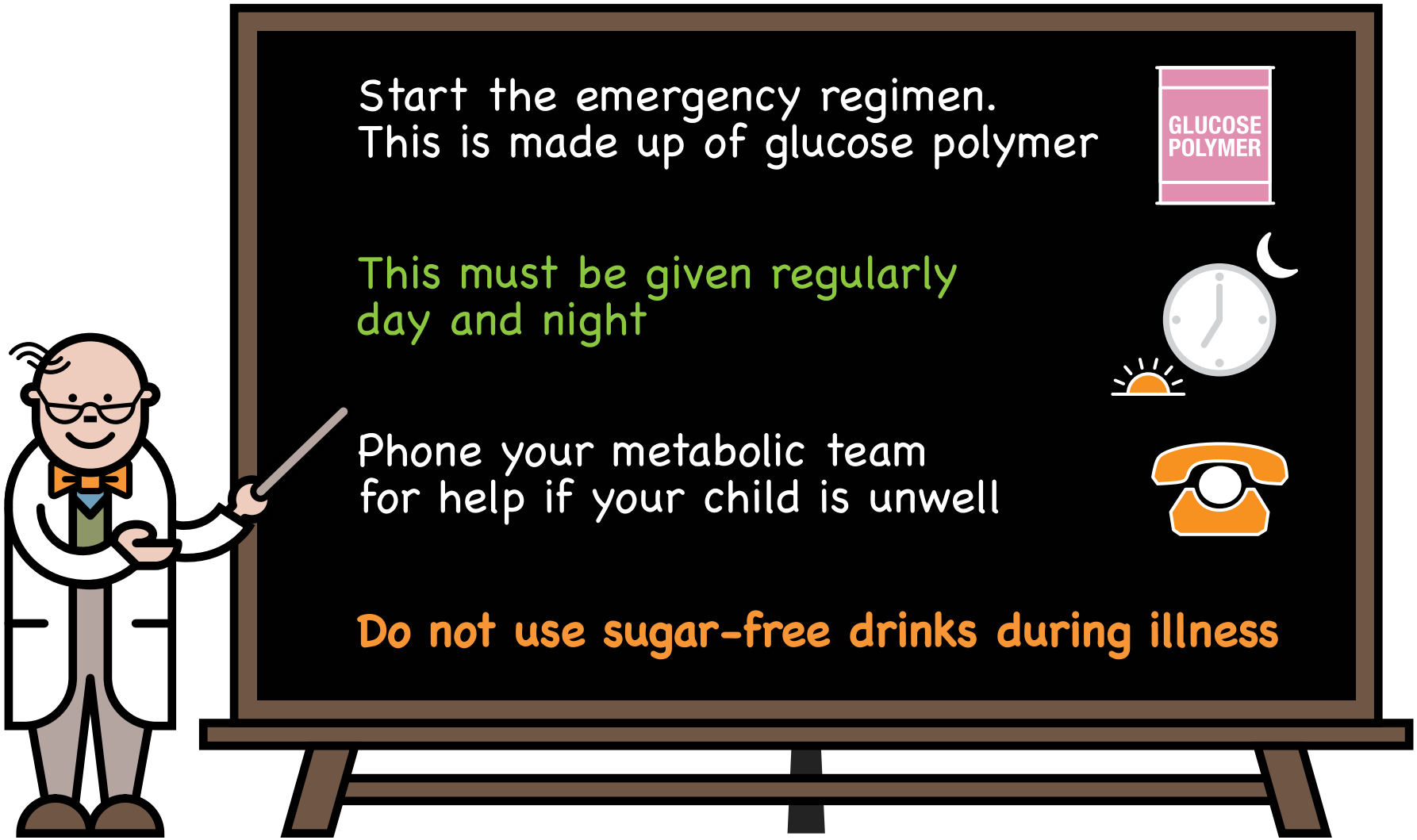

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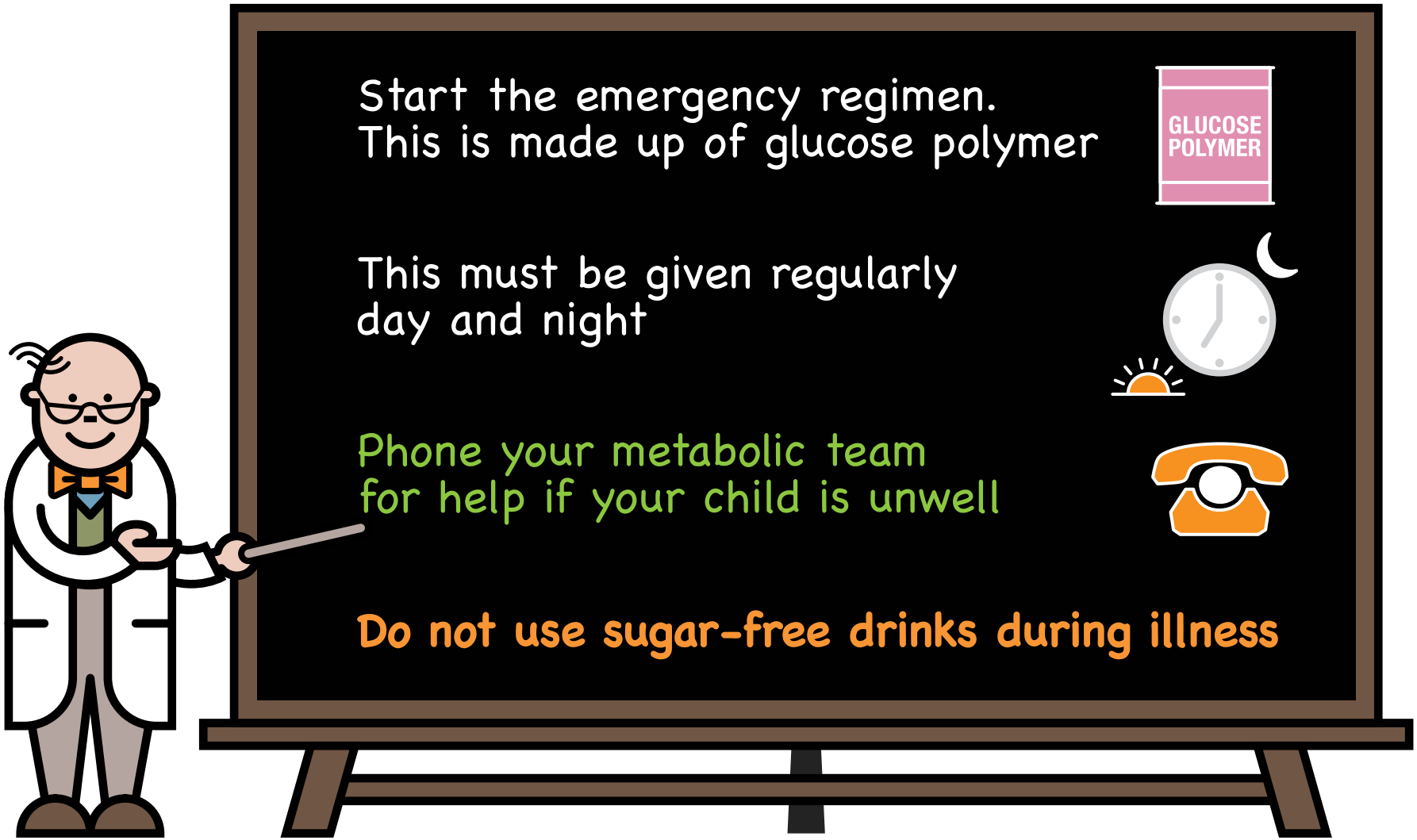
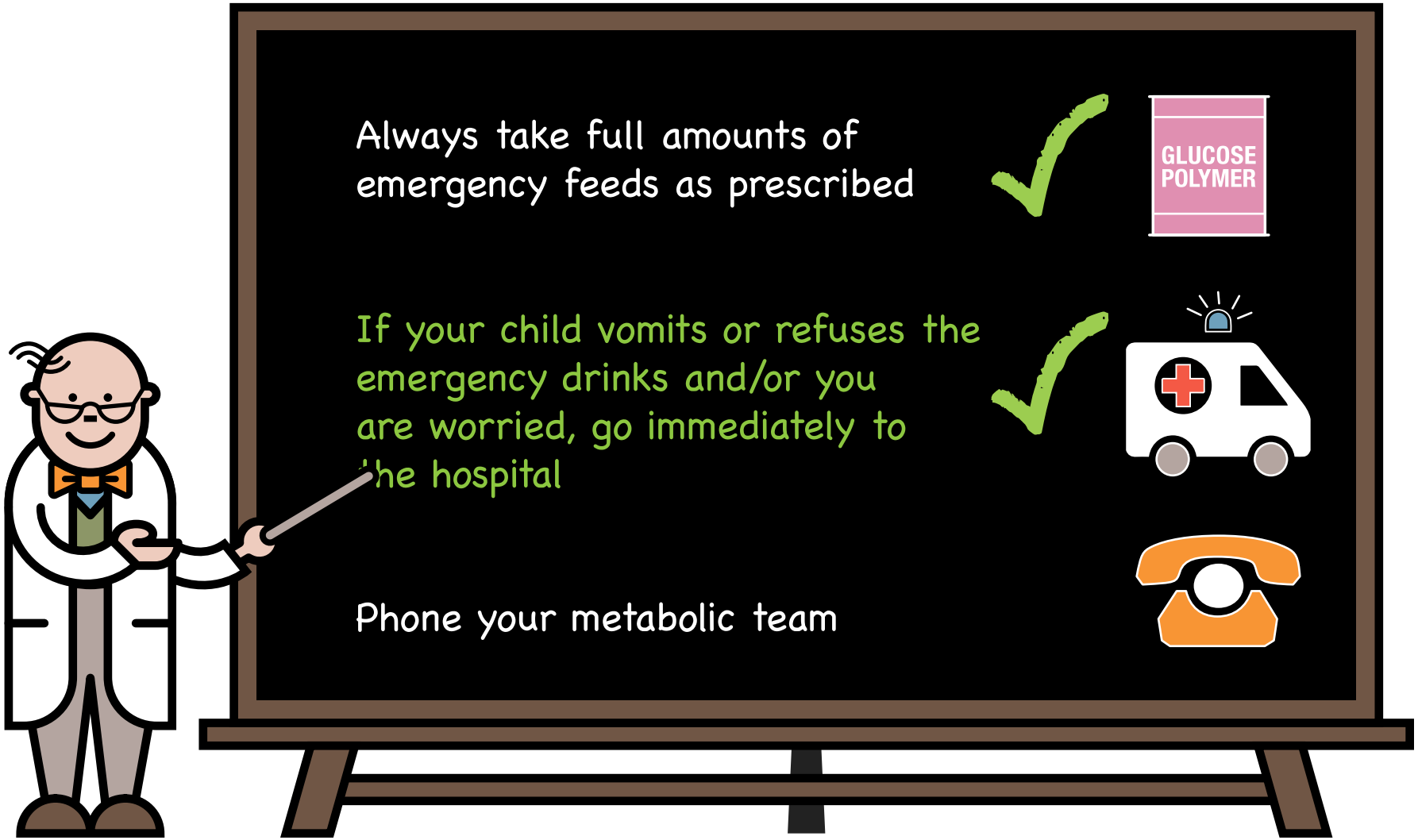


Checklist for illness

Checklist for illness



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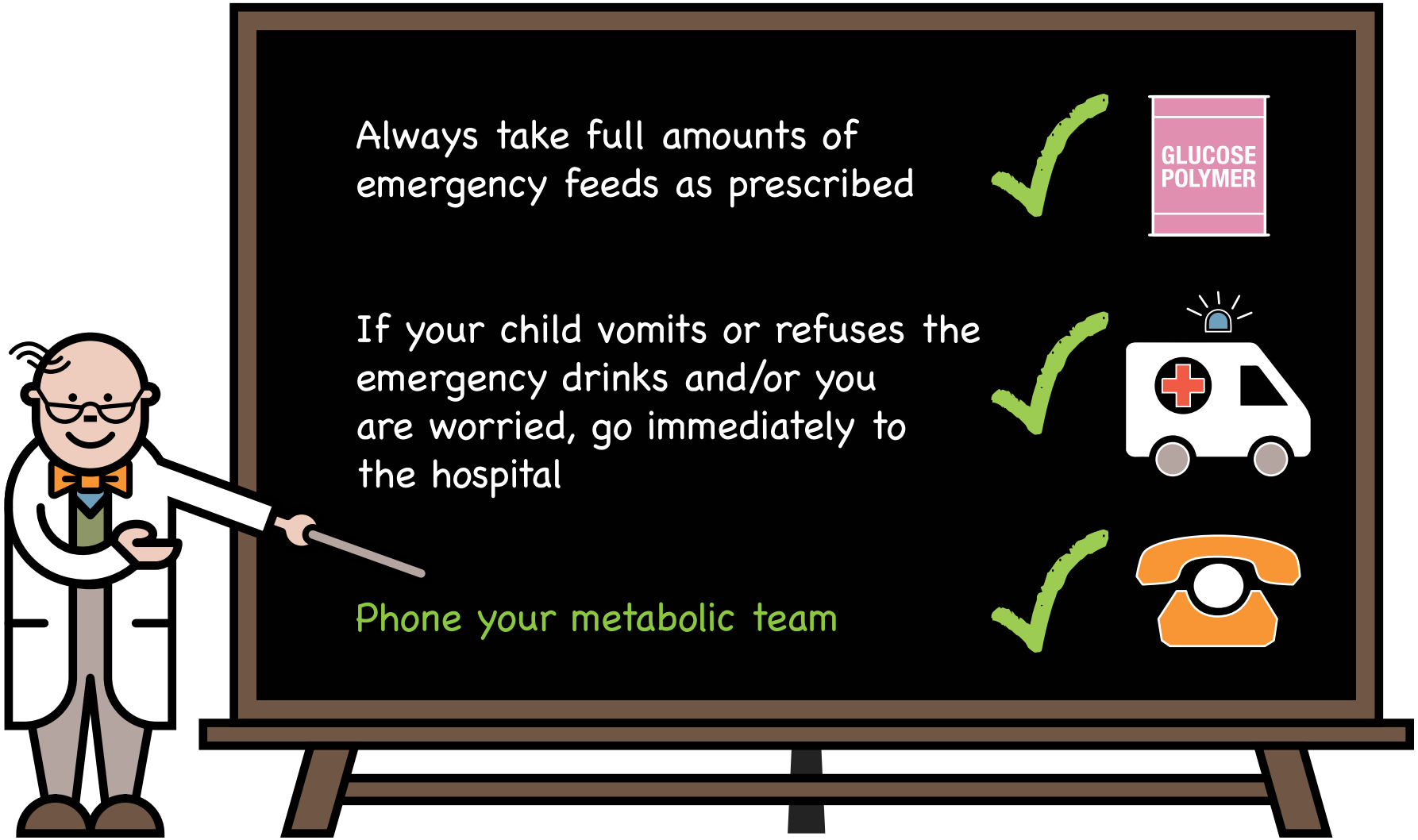
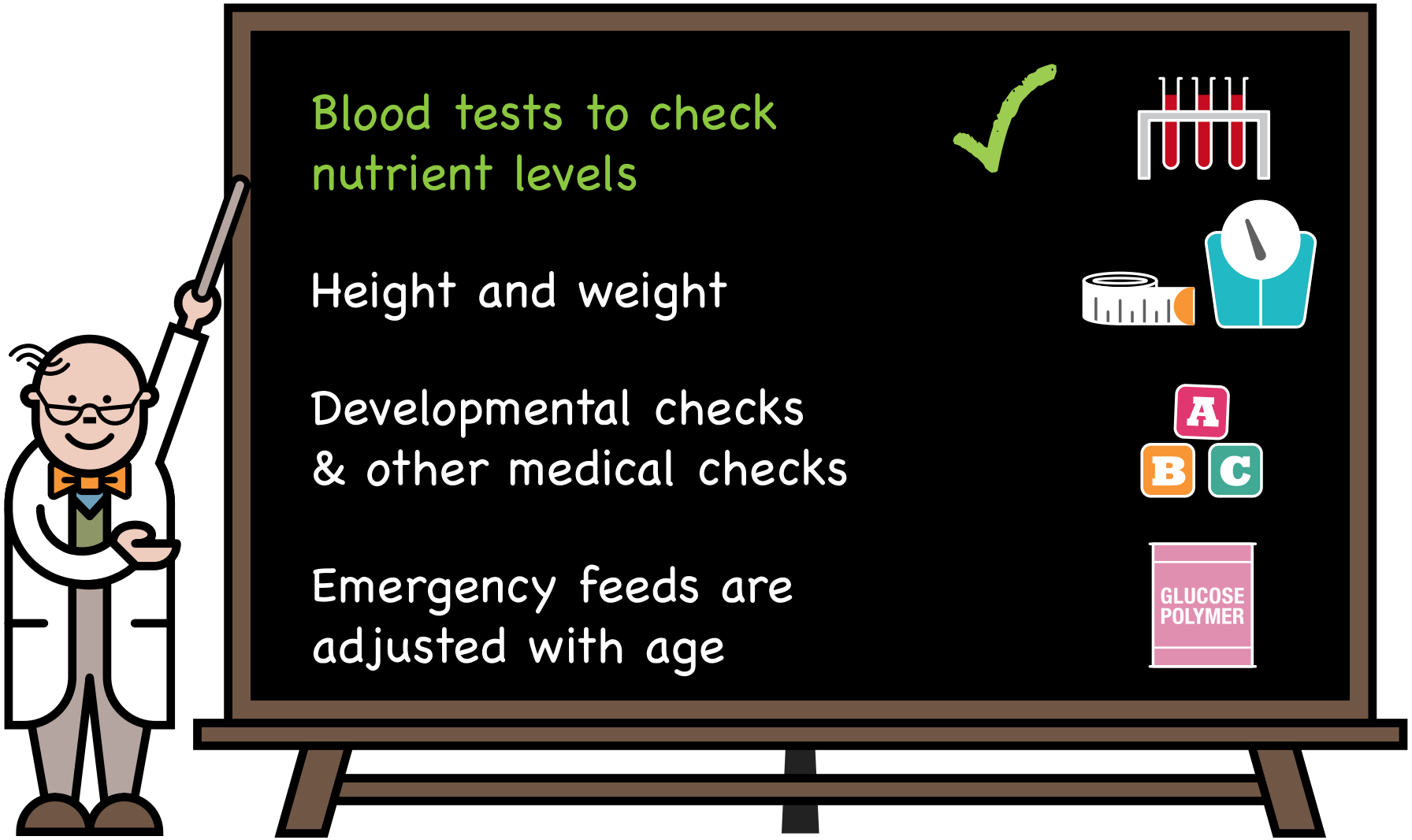


Key message

How is LCHADD monitored?



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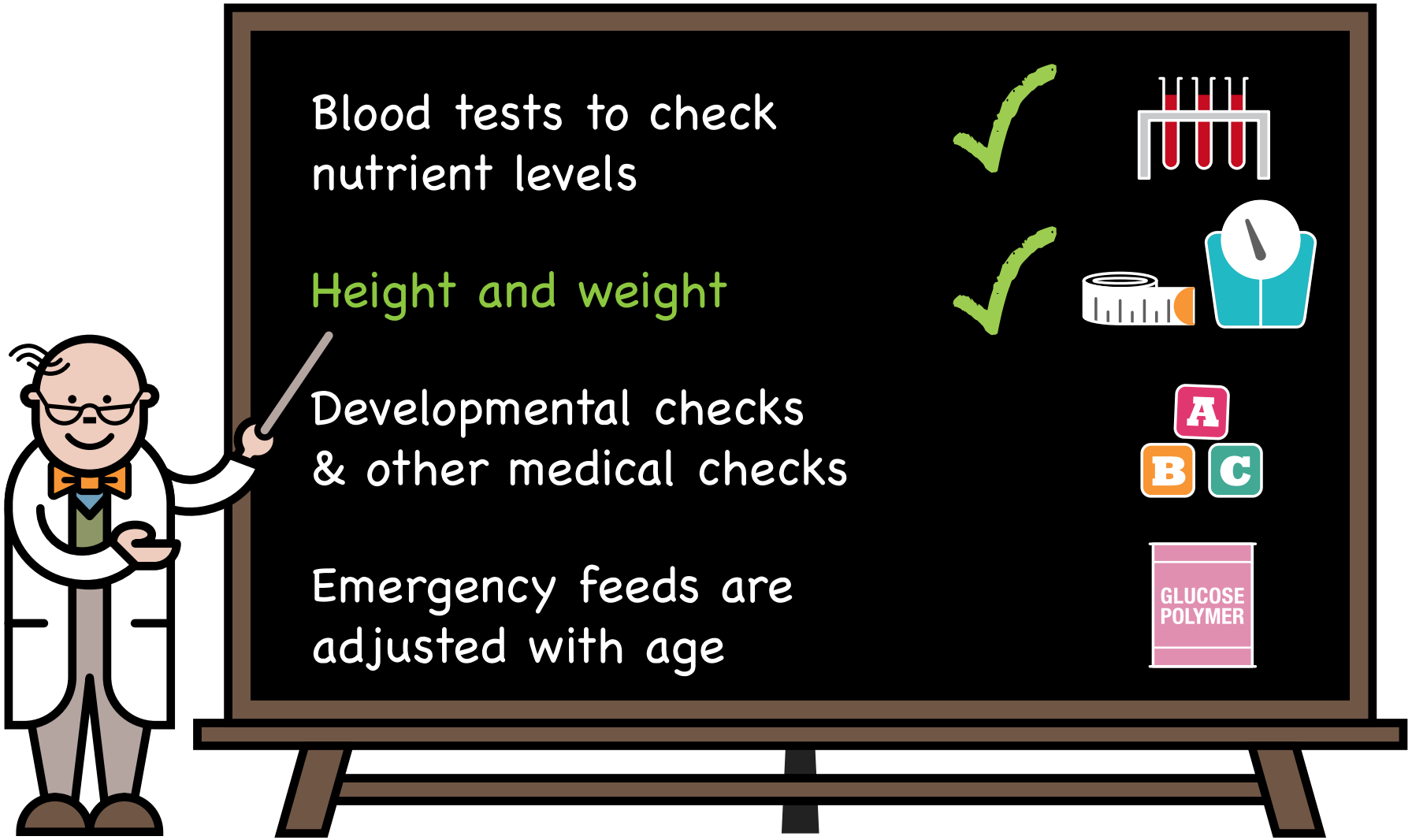

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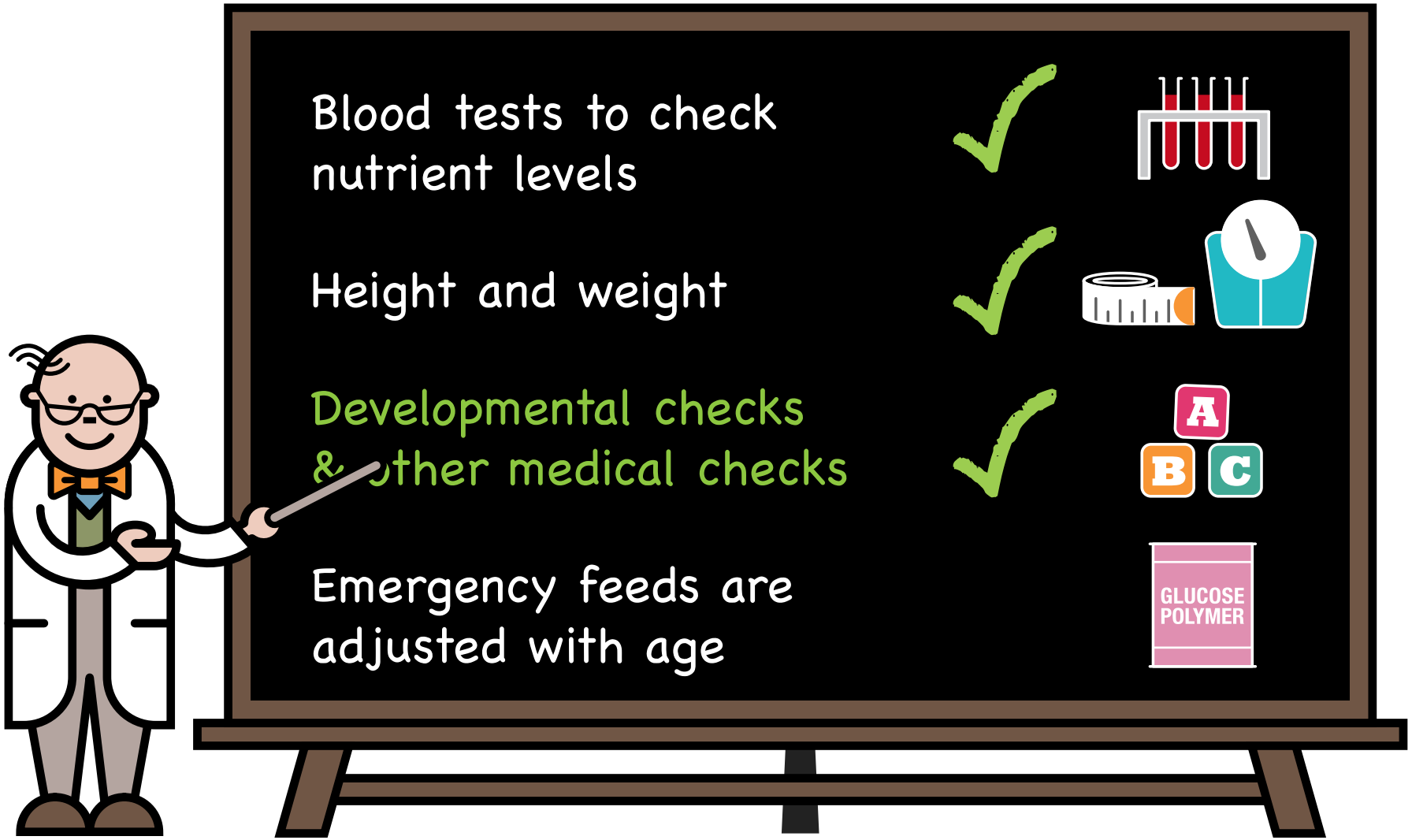

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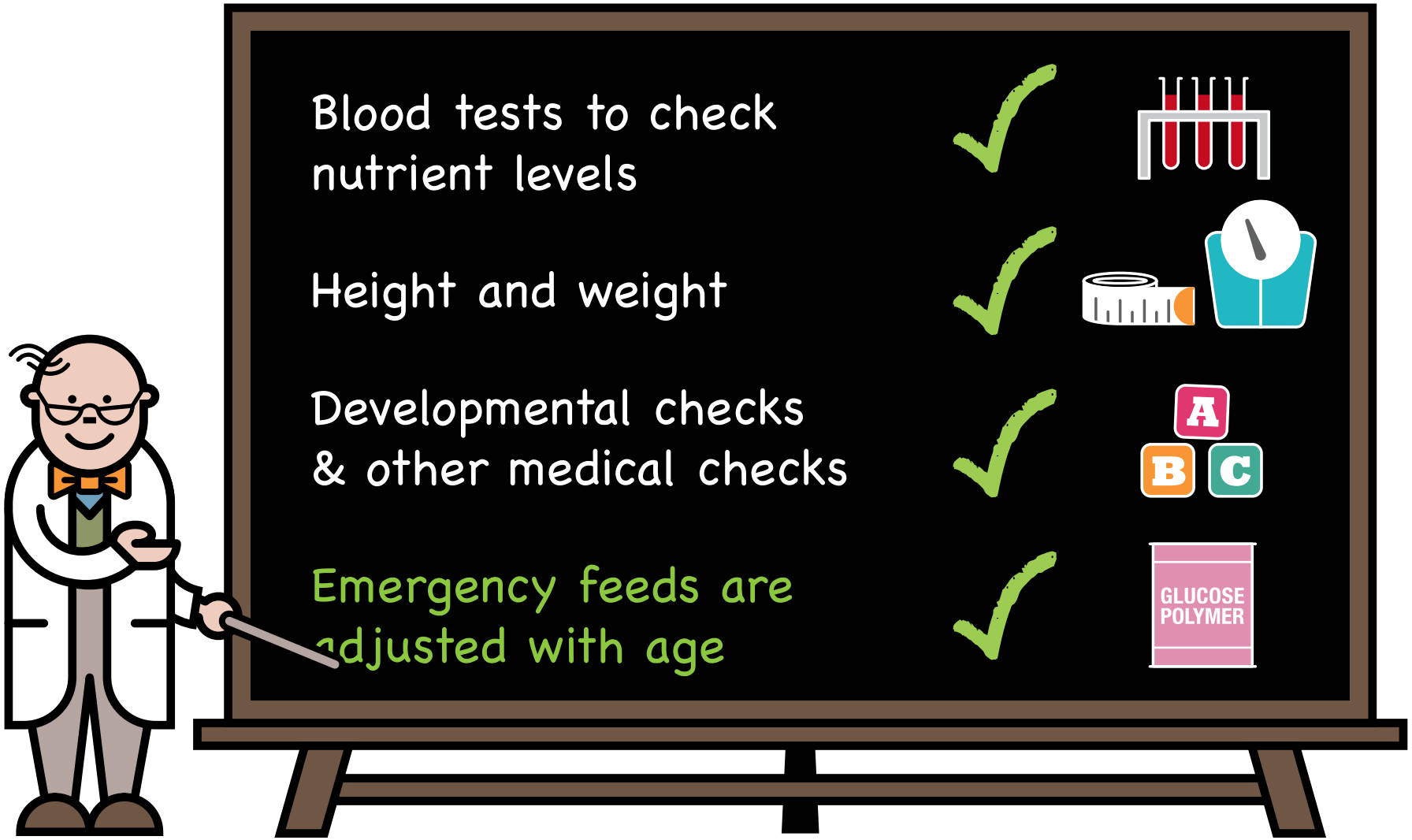
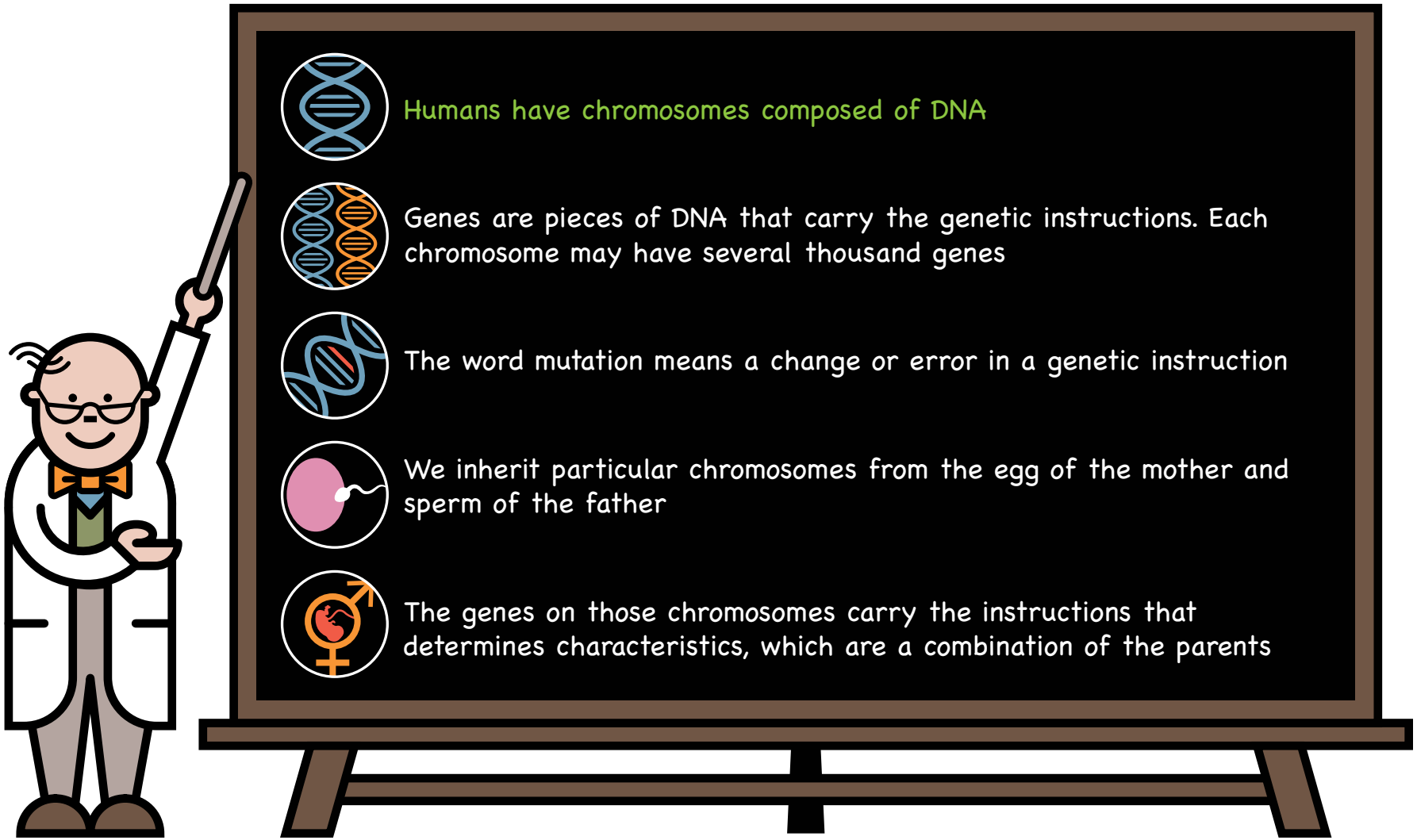


Chromosomes, genes, mutations



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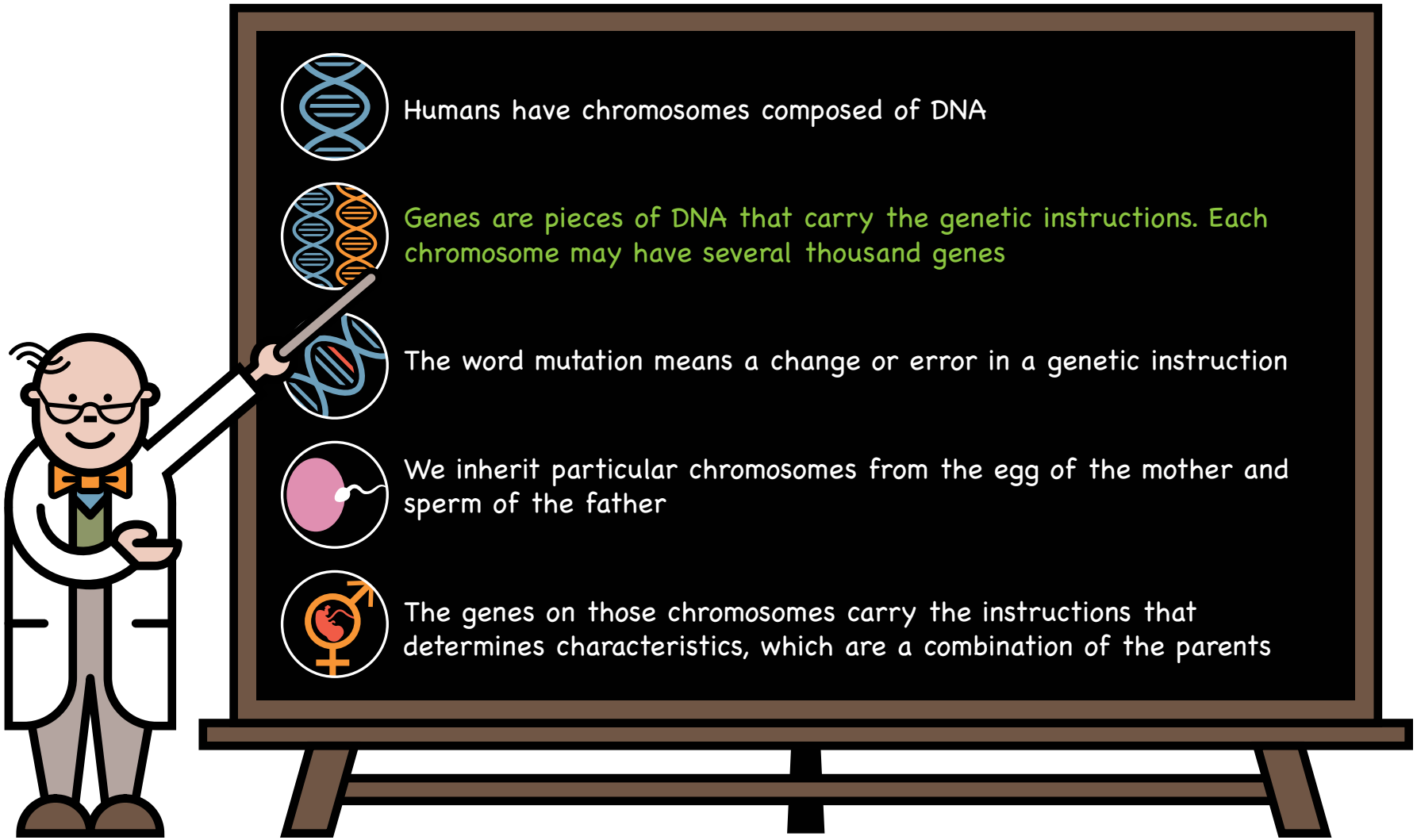
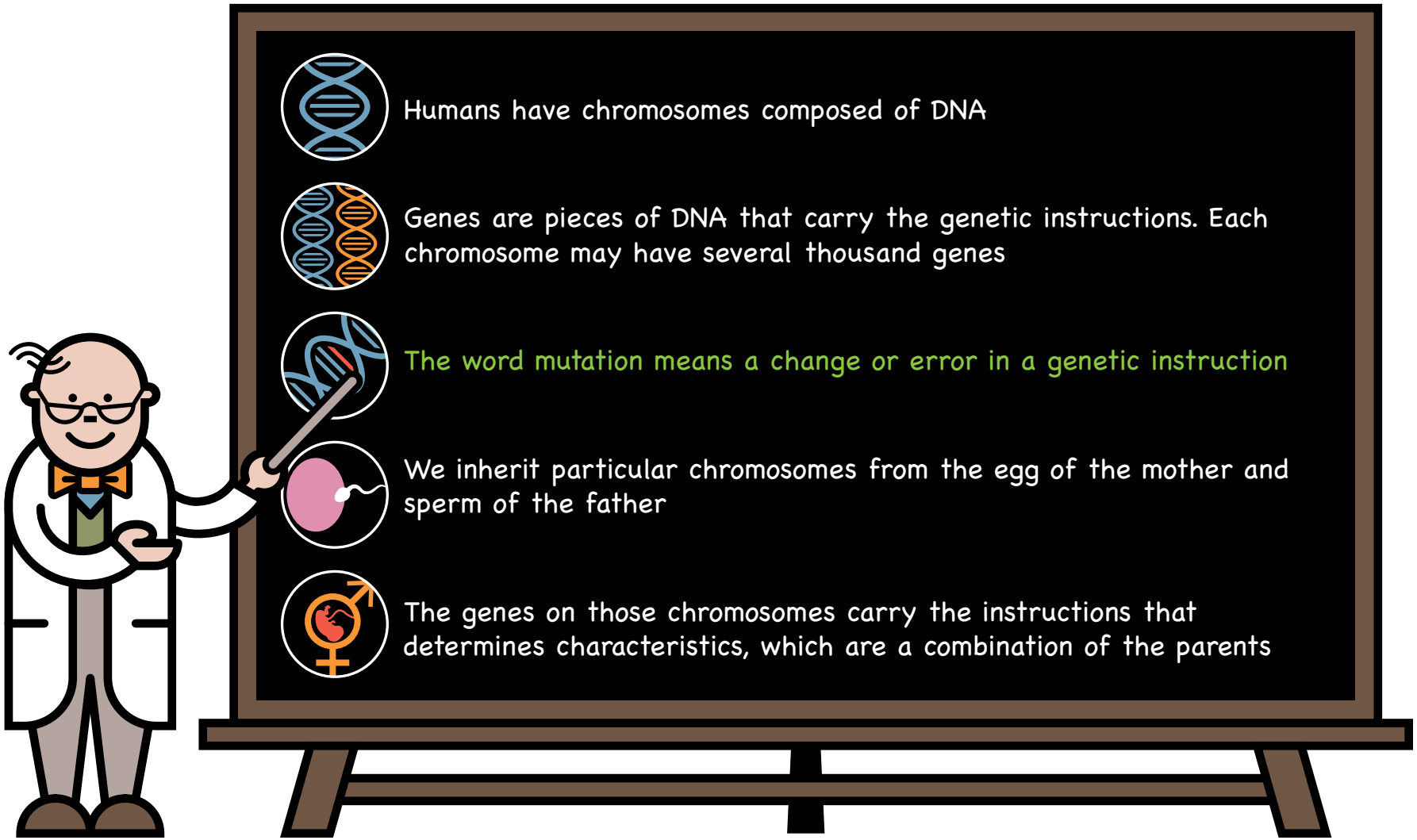


Chromosomes, genes, mutations



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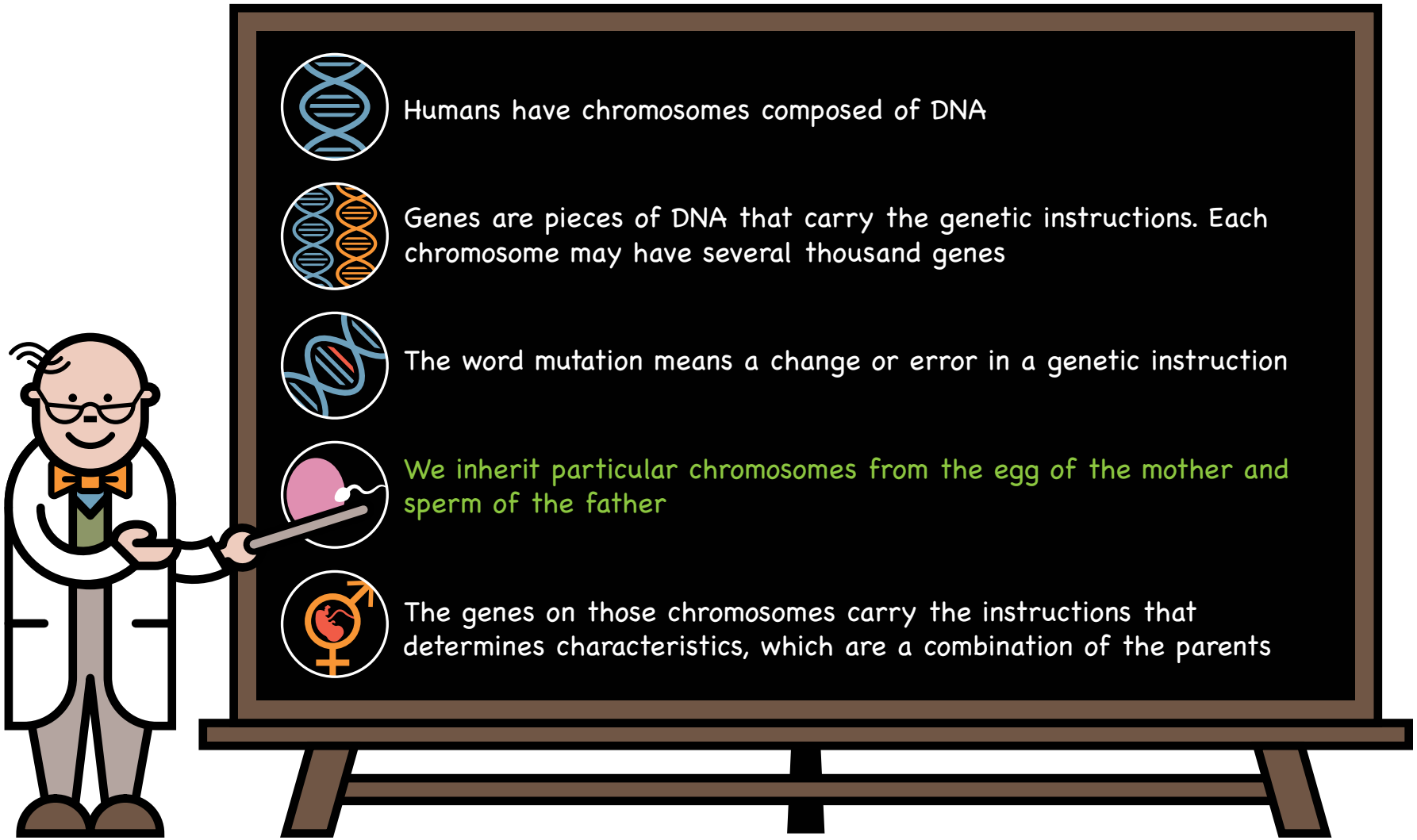

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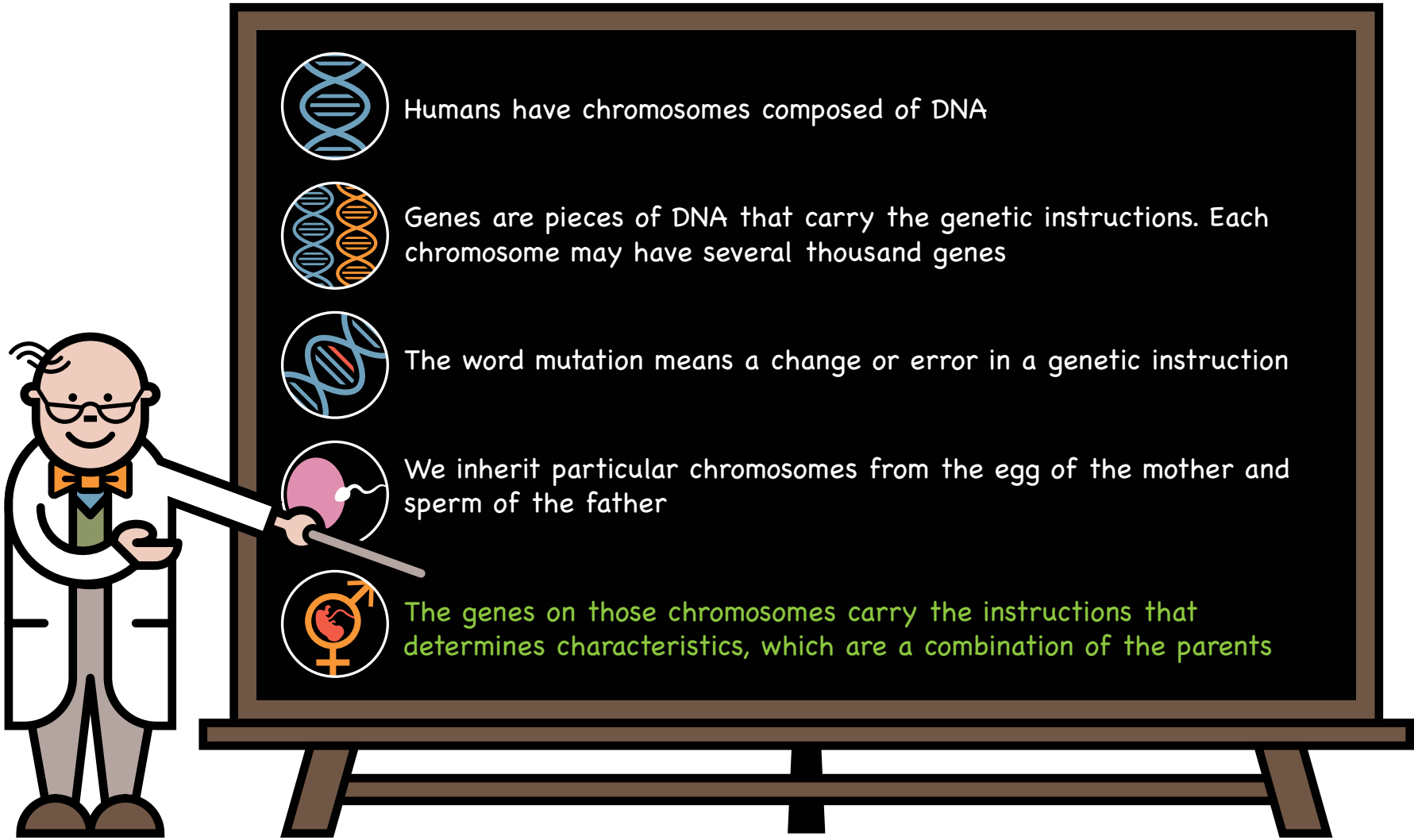
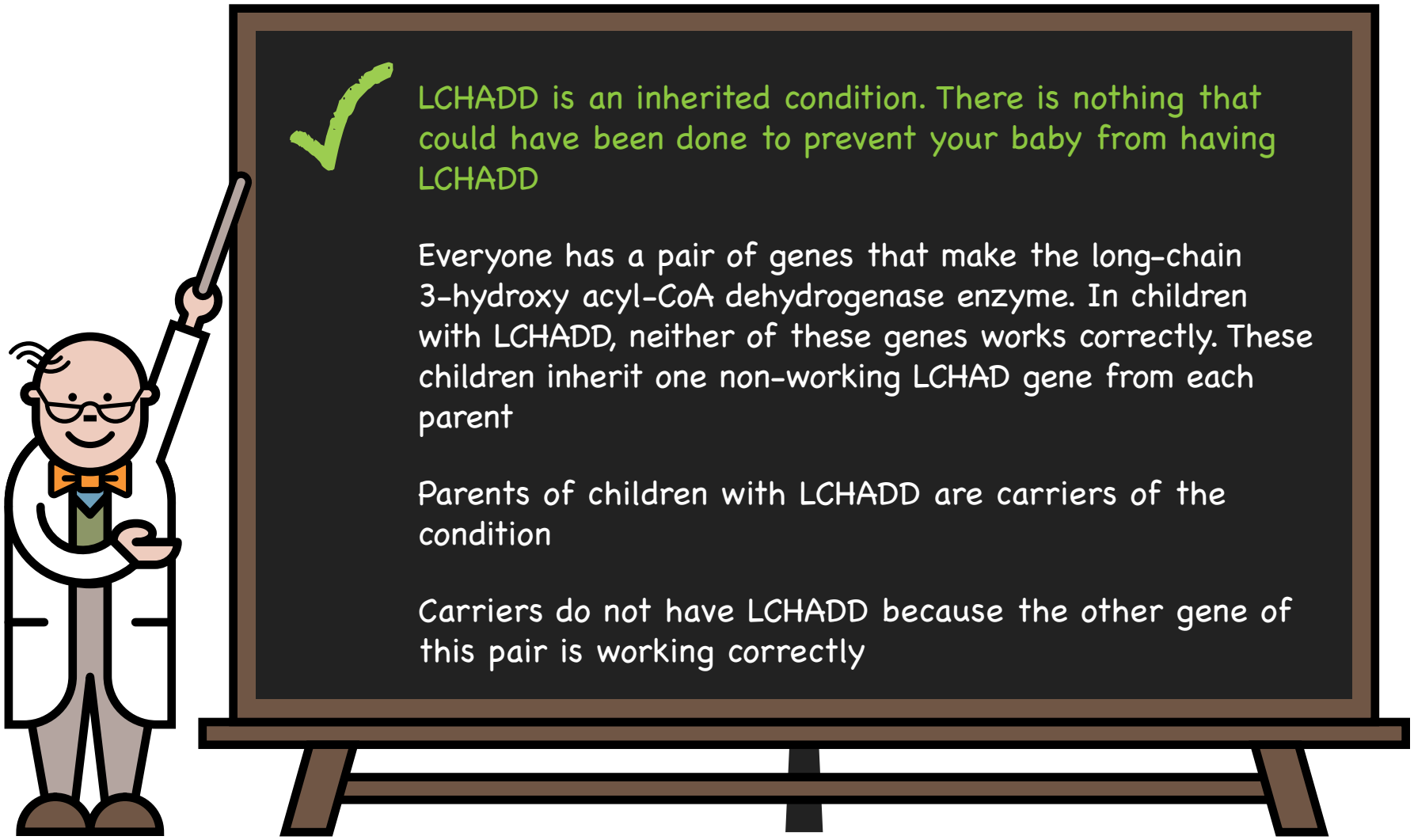


Inheritance



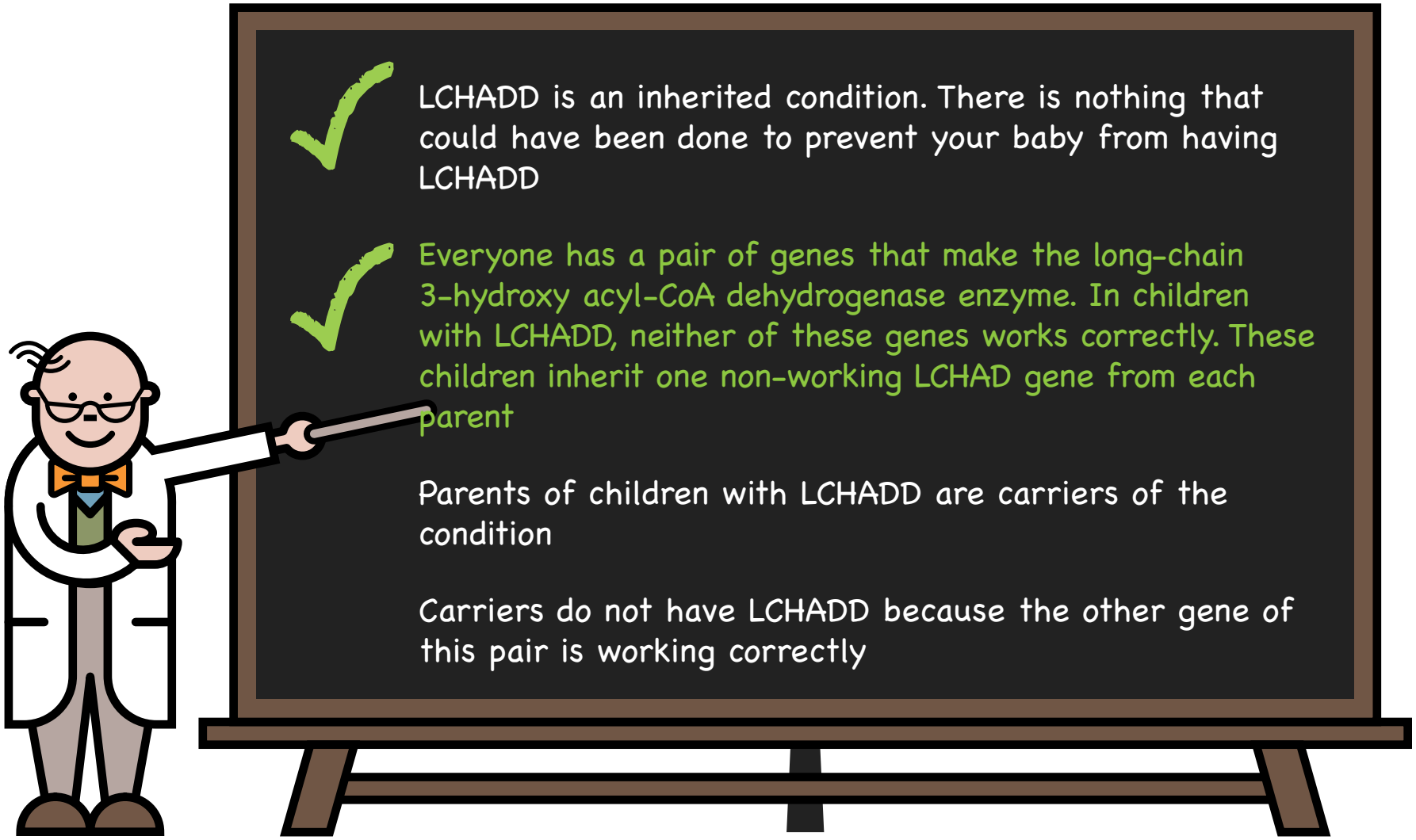
LCHADD is an inherited condition. There is nothing that could have been done to prevent your baby from having LCHADD

Everyone has a pair of genes that make the long-chain 3-hydroxy acyl-CoA dehydrogenase enzyme. In children with LCHADD, neither of these genes works correctly. These children inherit one non-working LCHAD gene from each parent

Parents of children with LCHADD are carriers of the condition

Carriers do not have LCHADD because the other gene of this pair is working correctly

Inheritance



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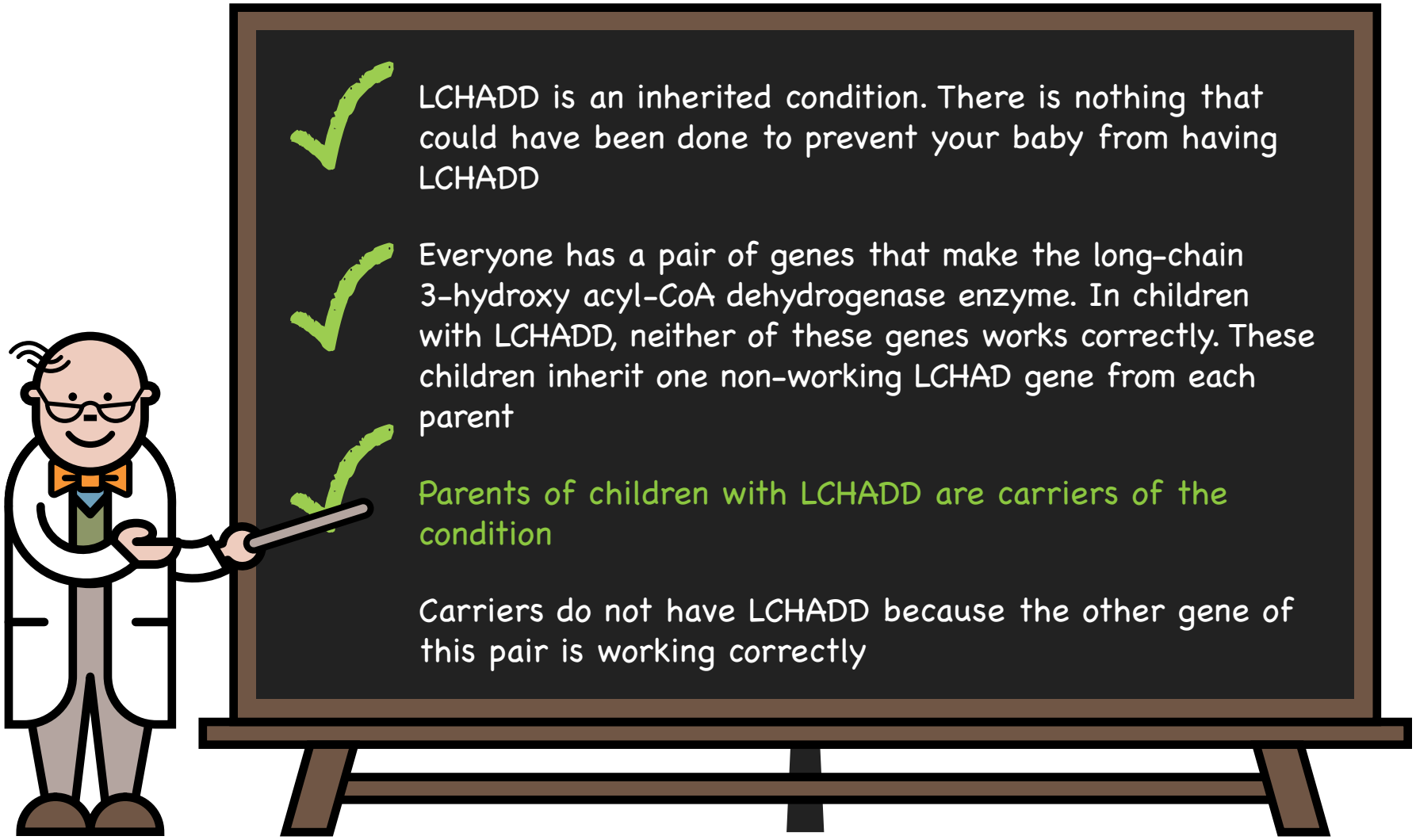
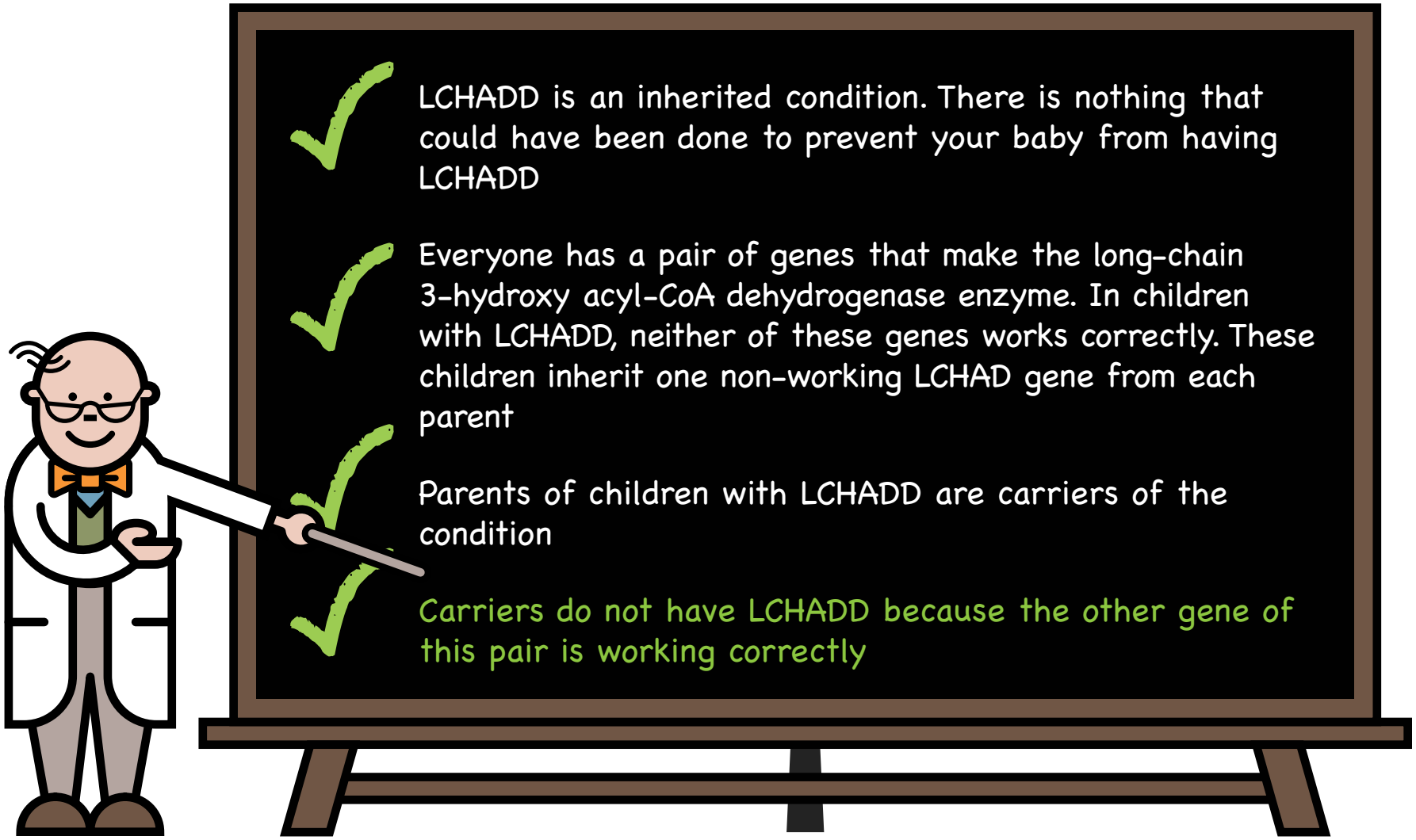
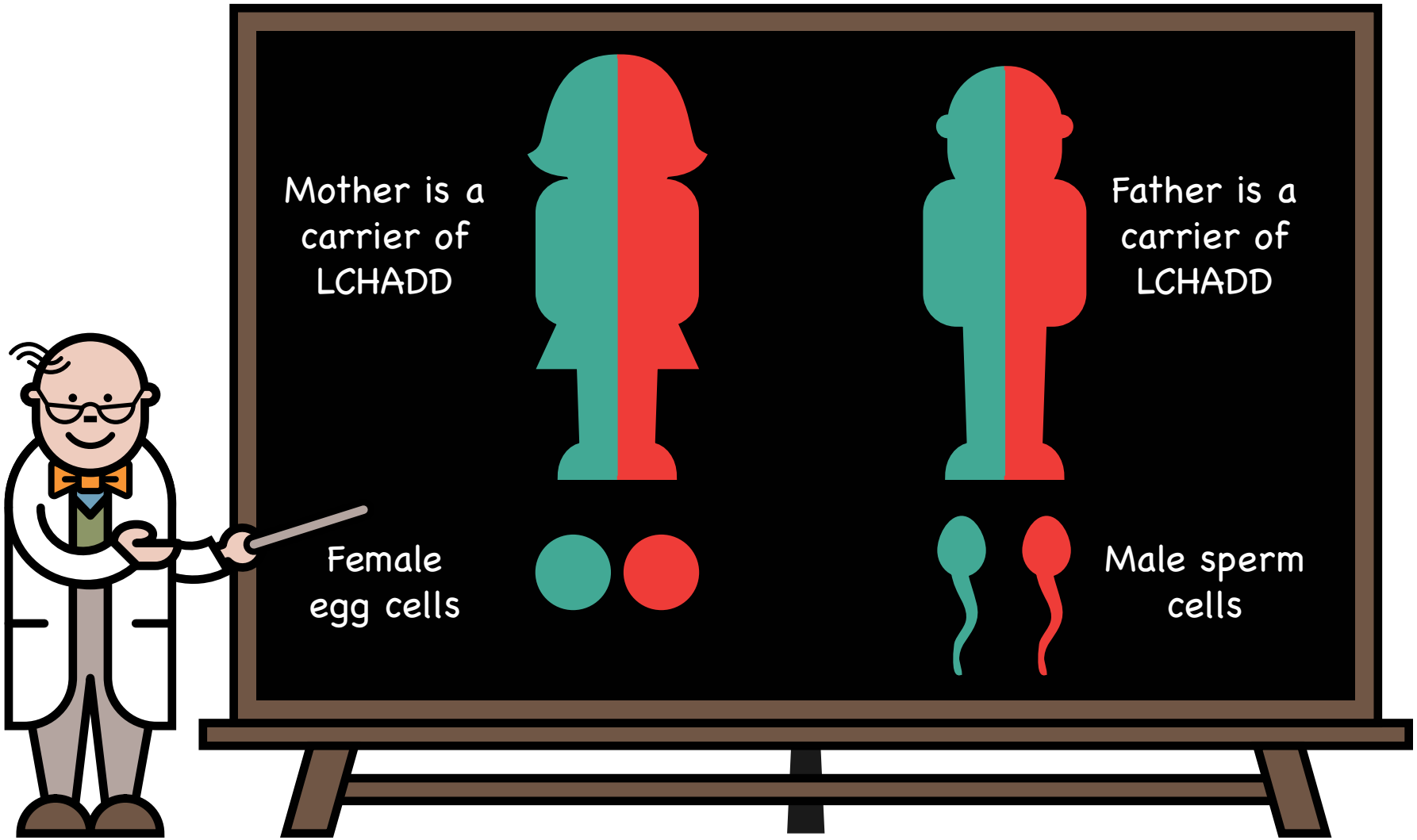


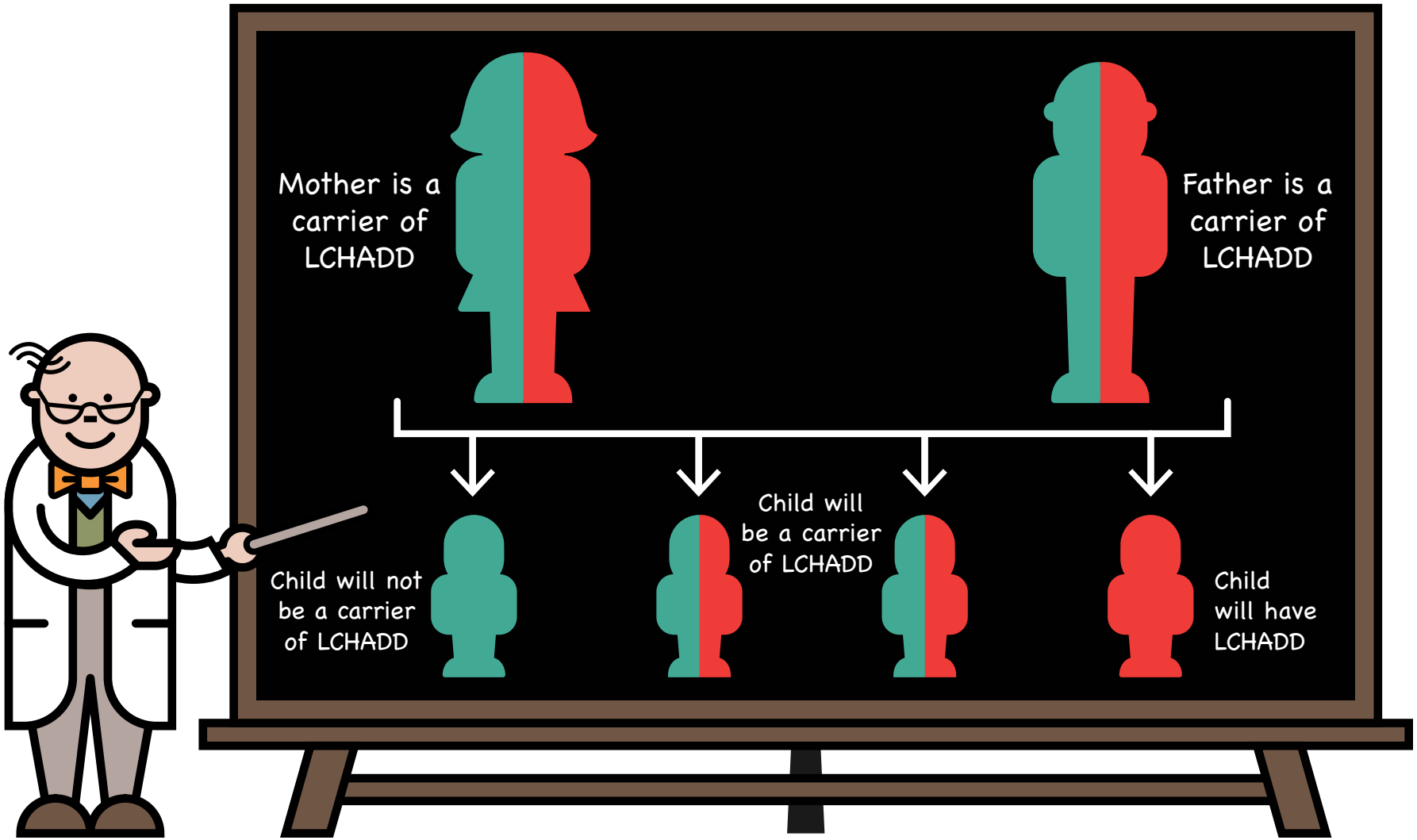
Inheritance



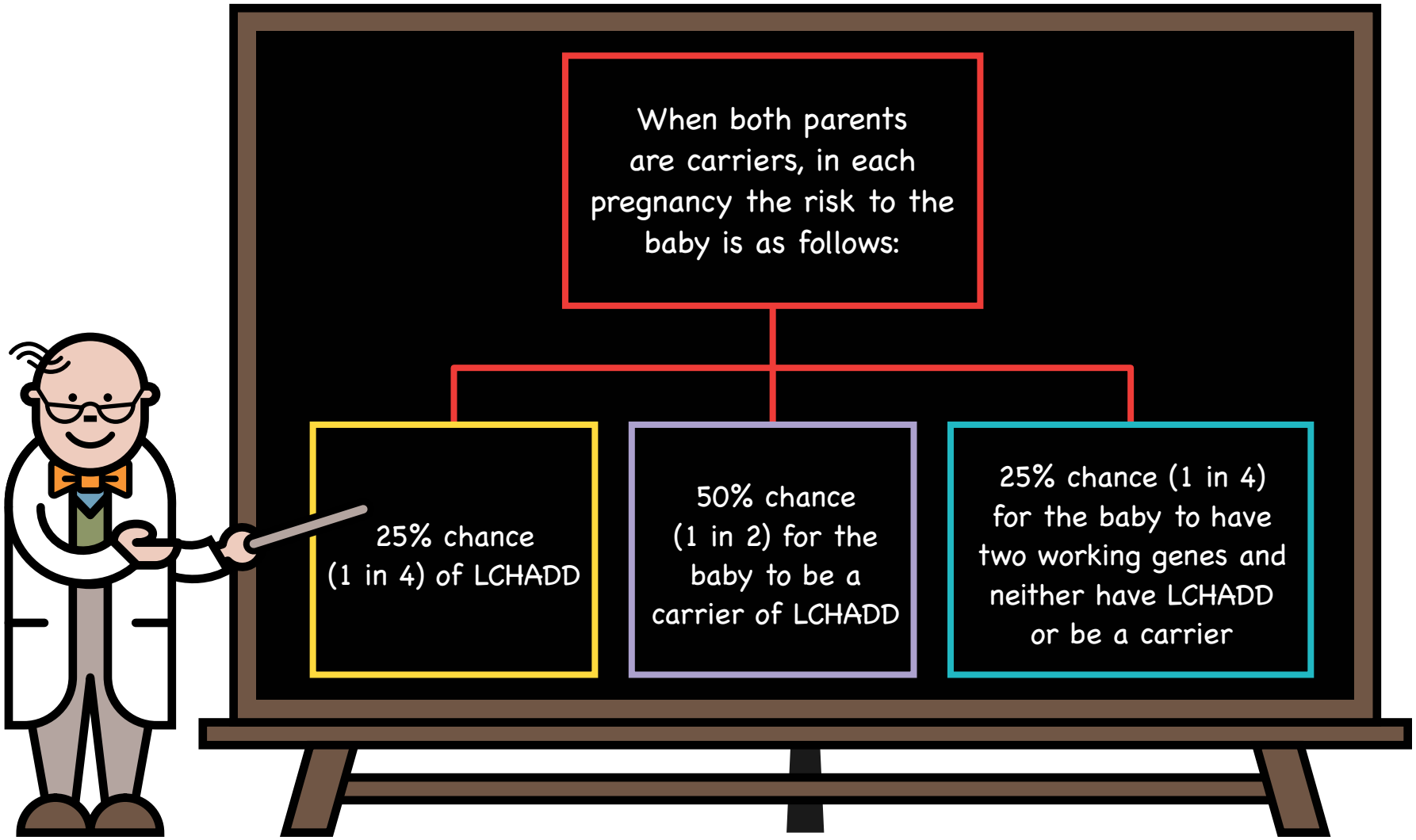
Inheritance – Autosomal recessive (carriers of LCHADD)



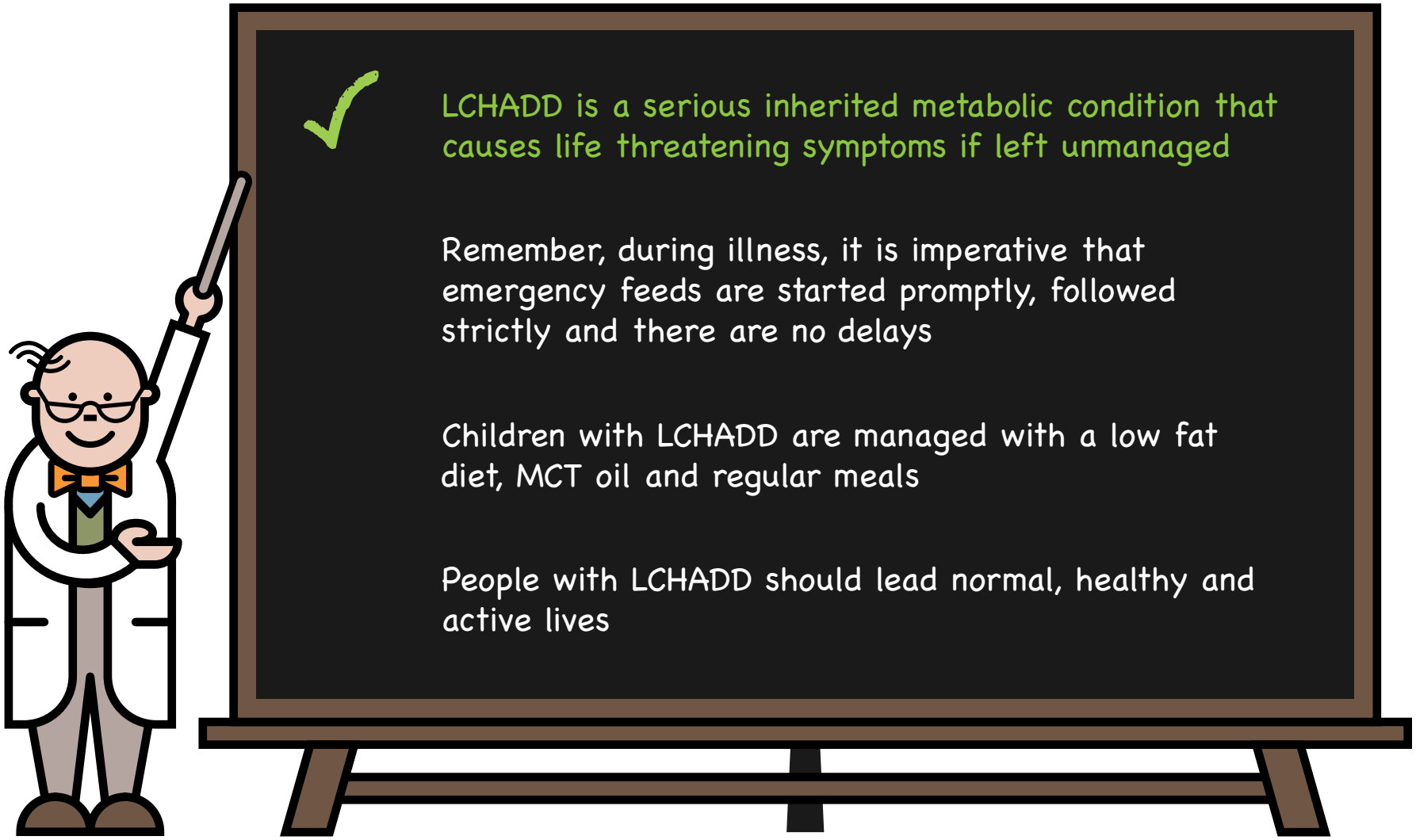
Inheritance – Autosomal recessive – possible combinations



Future pregnancies



Take home messages



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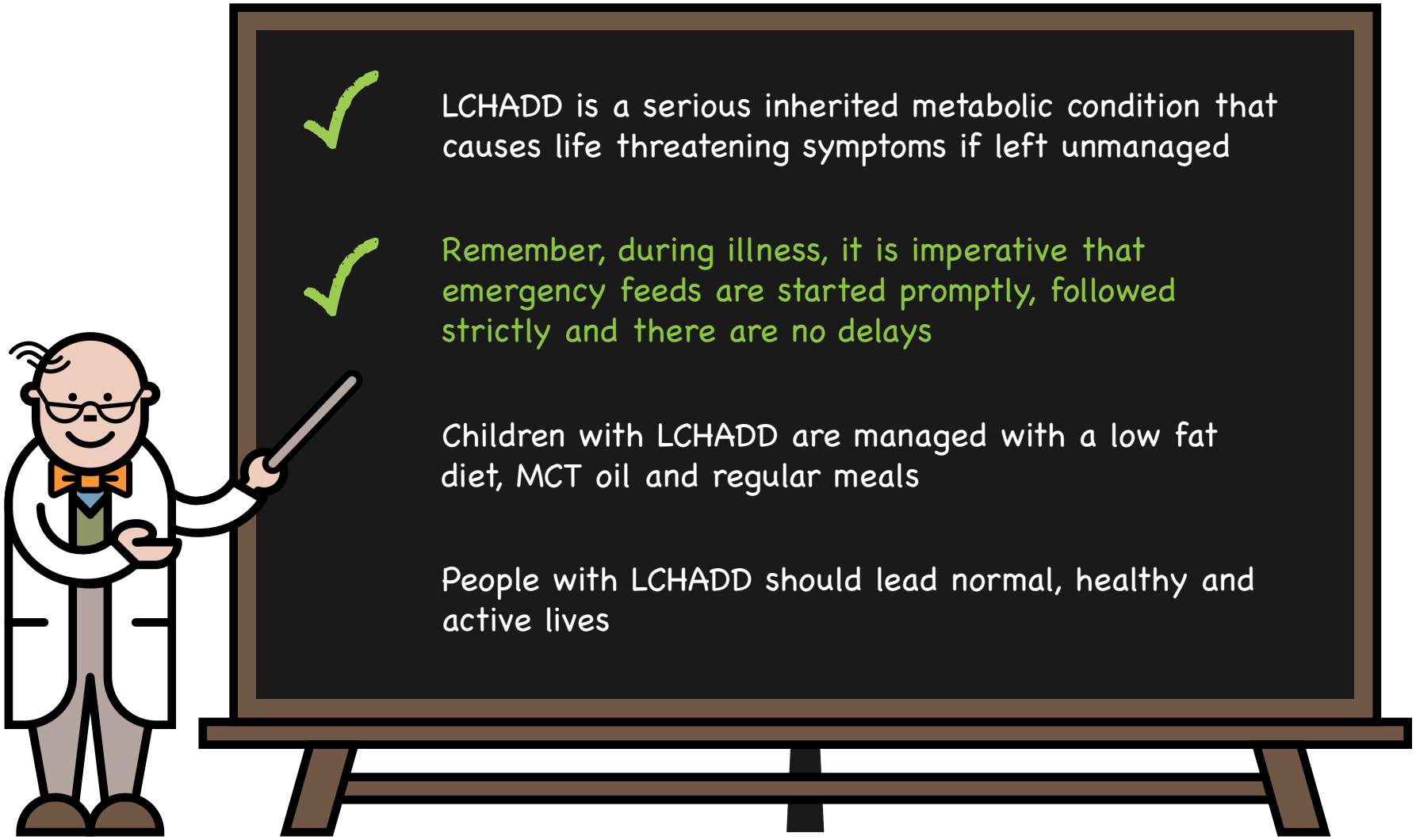

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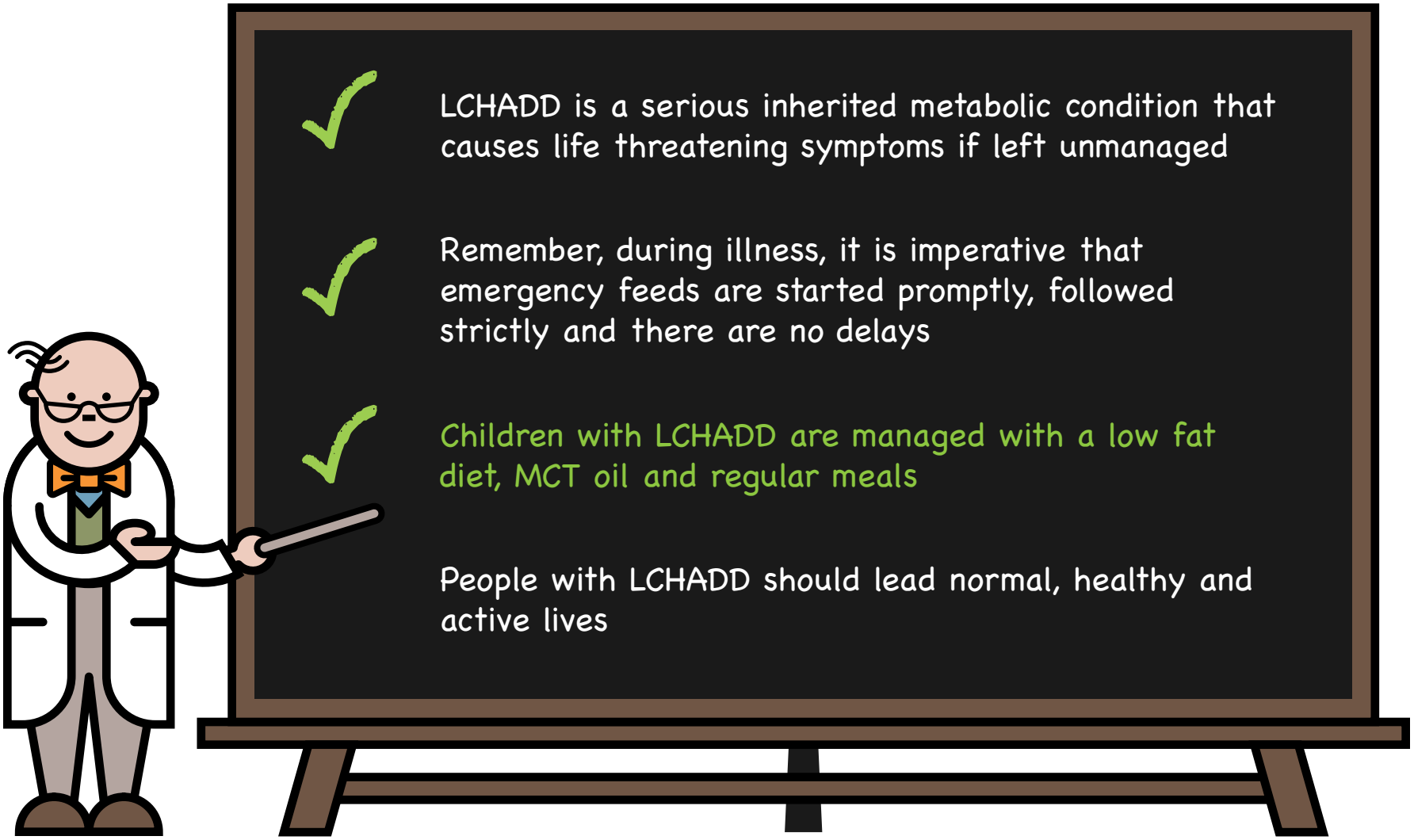

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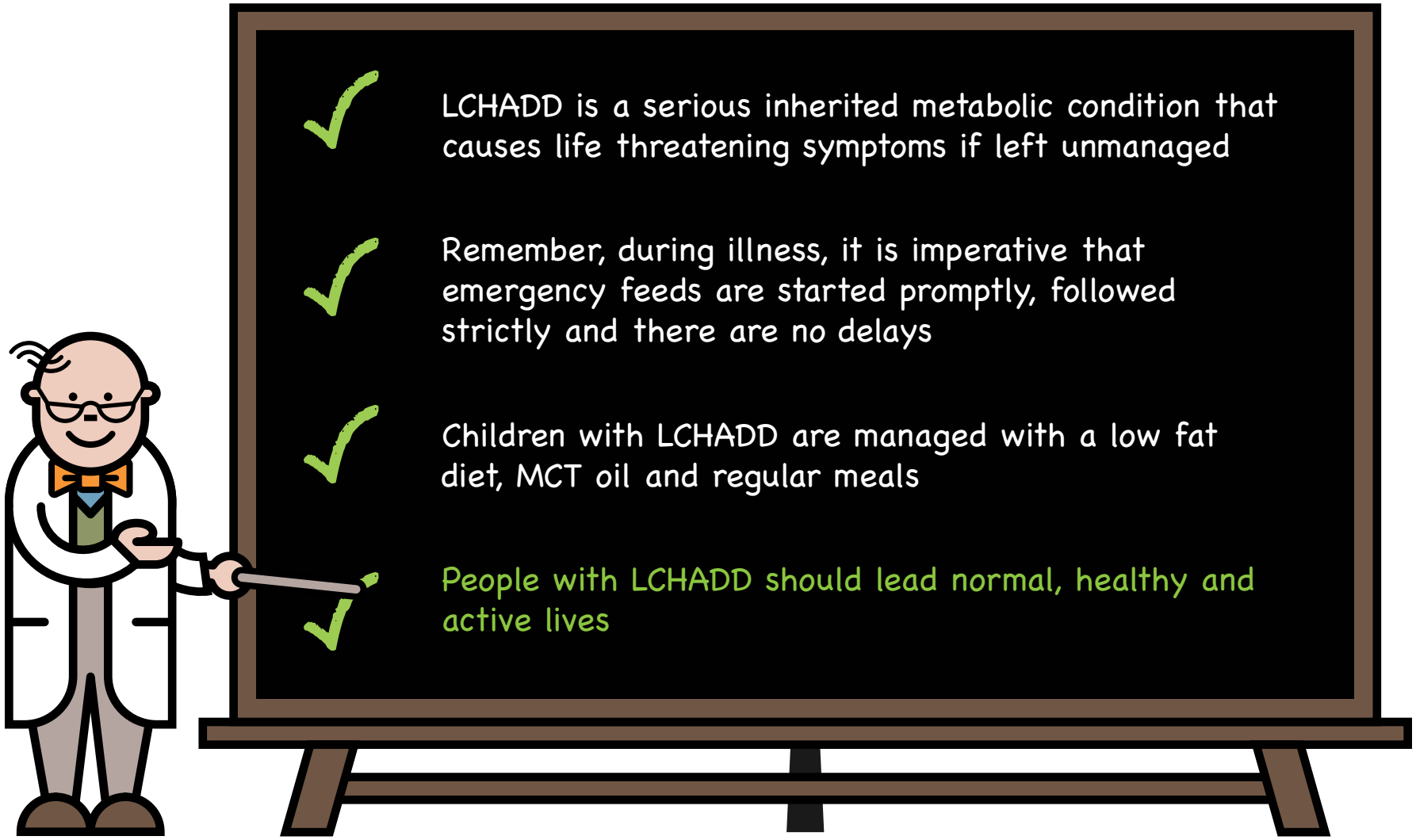
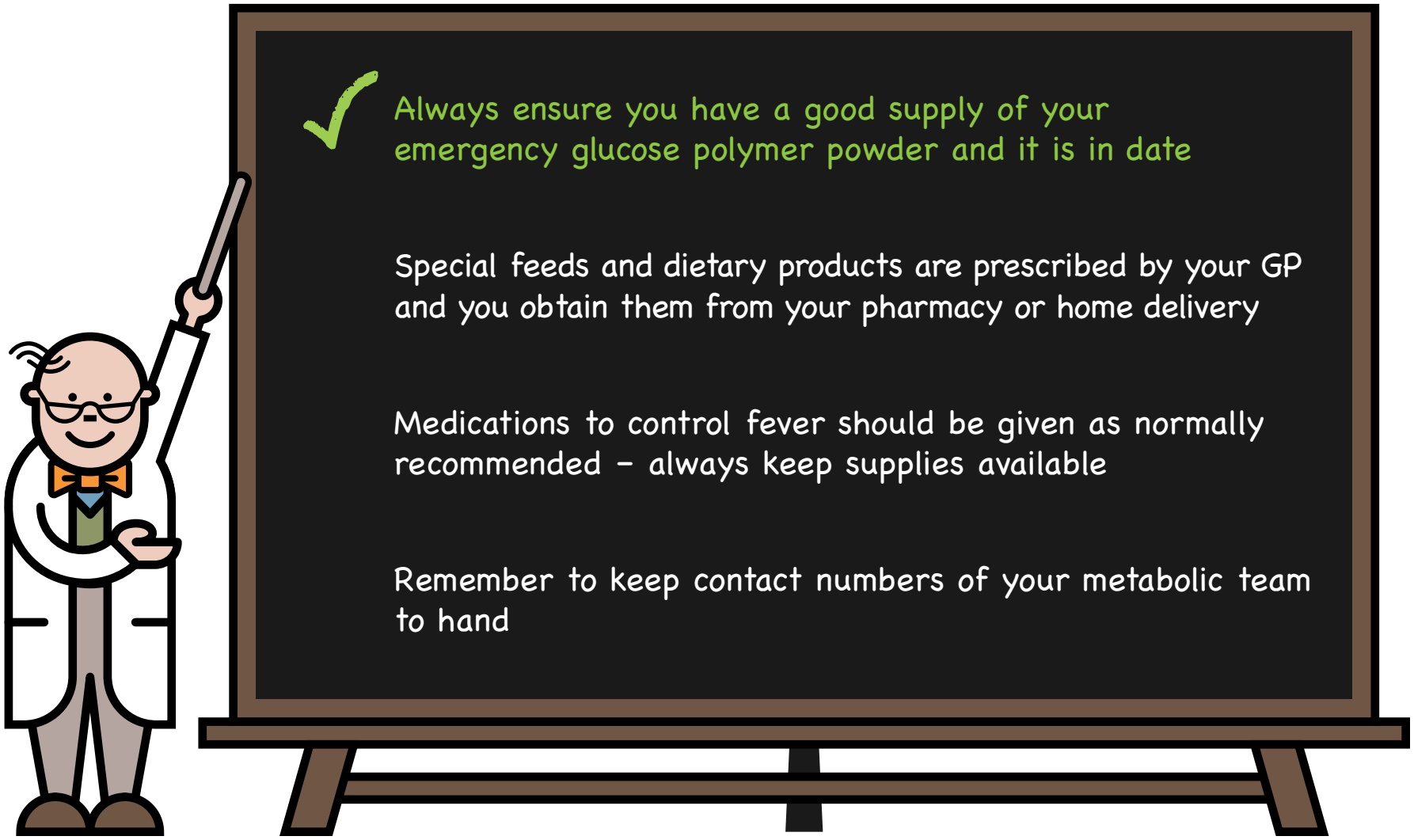


Helpful hints



Helpful hints

Helpful hints

Helpful hints

Who's who

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

MET1843UKIR

Visit **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.

Low Protein Connect

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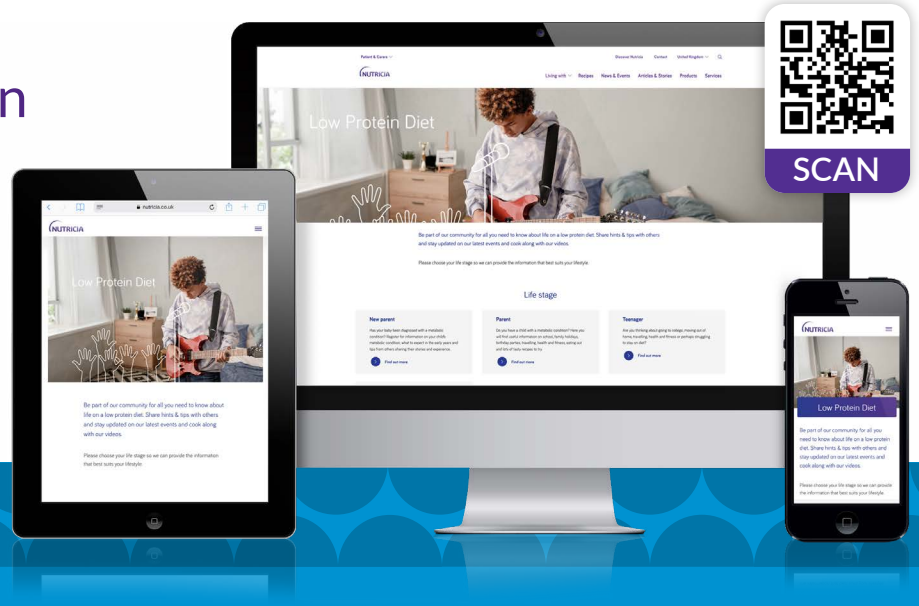

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METABOLIC SUPPORT UK
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