TEMPLE

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

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

PA

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.

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

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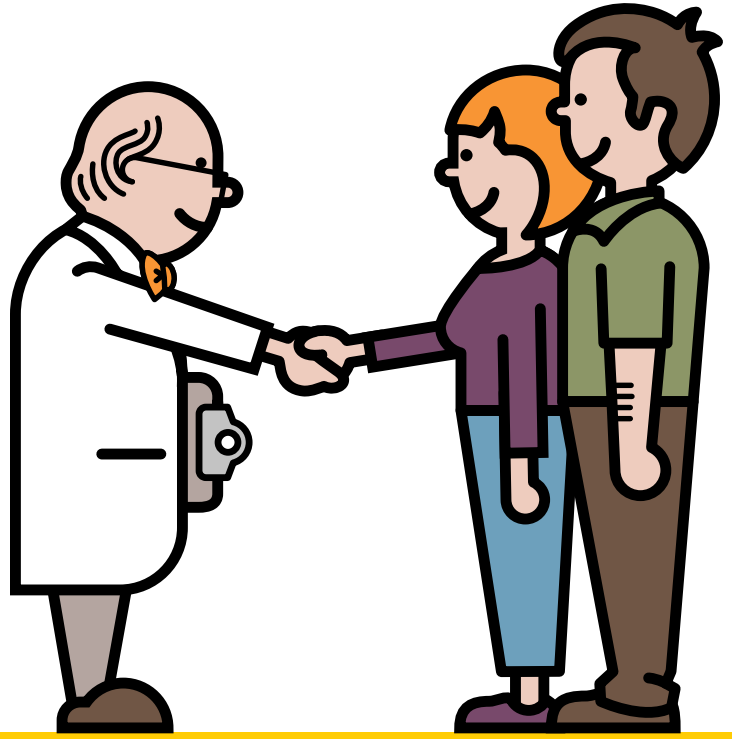
PA

Information for parents following a new diagnosis



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Tools Enabling Metabolic Parents LEarning

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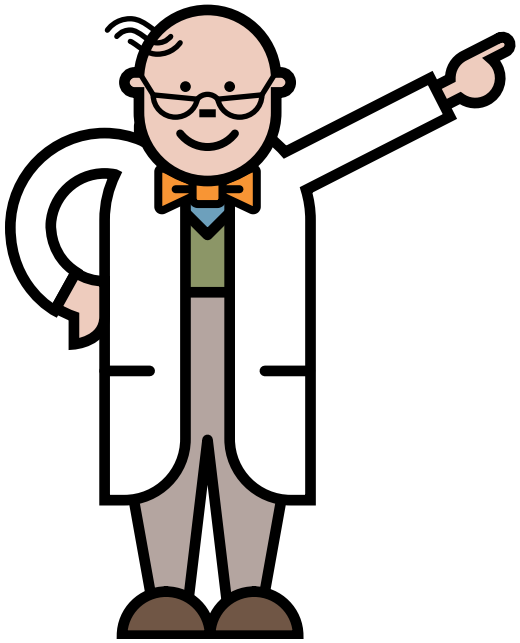
What is PA?

PA stands for Propionic Acidaemia

It is an inherited metabolic condition



Propionic Acidaemia

PA

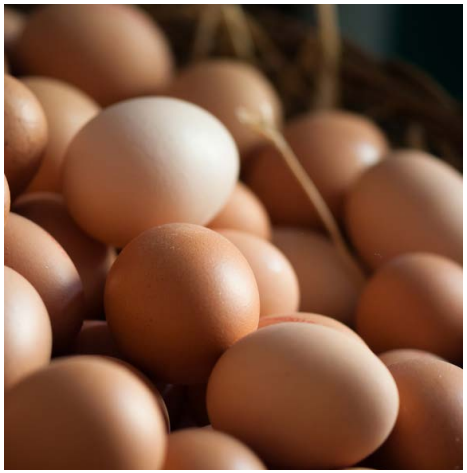
PA and protein

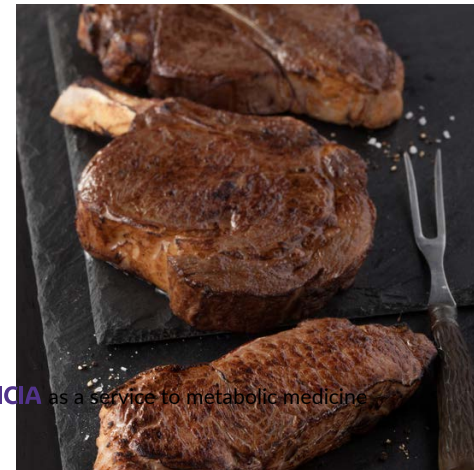
PA affects the way your baby breaks down protein

Many foods contain protein

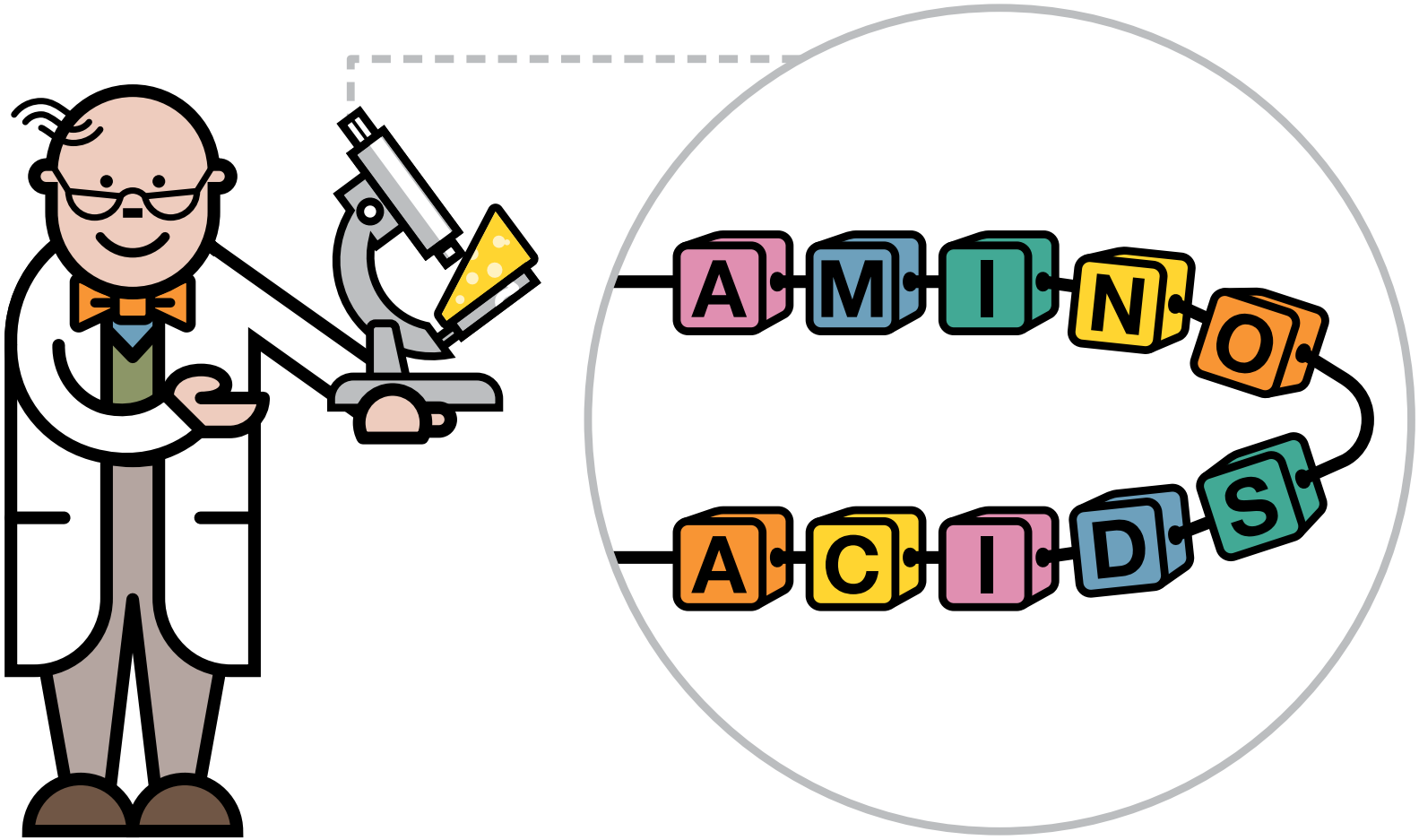
The body needs protein for growth and repair





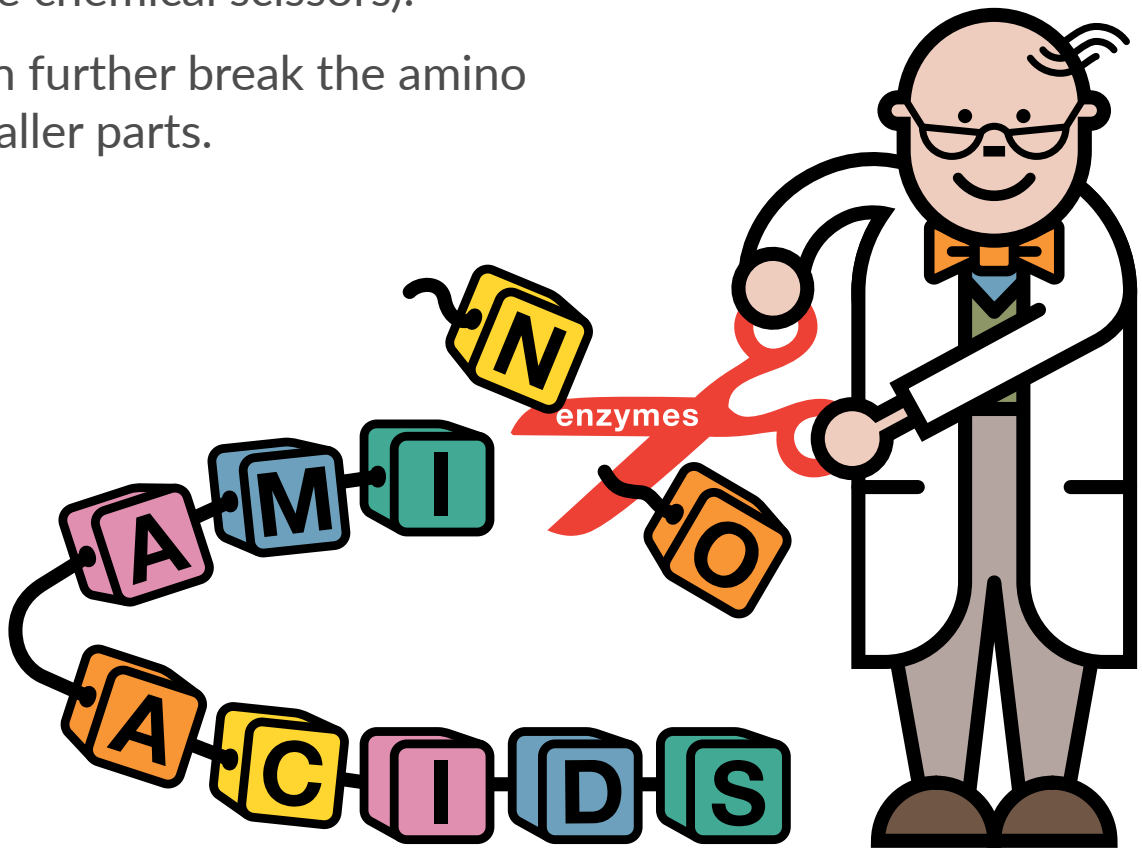
What is protein?



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.

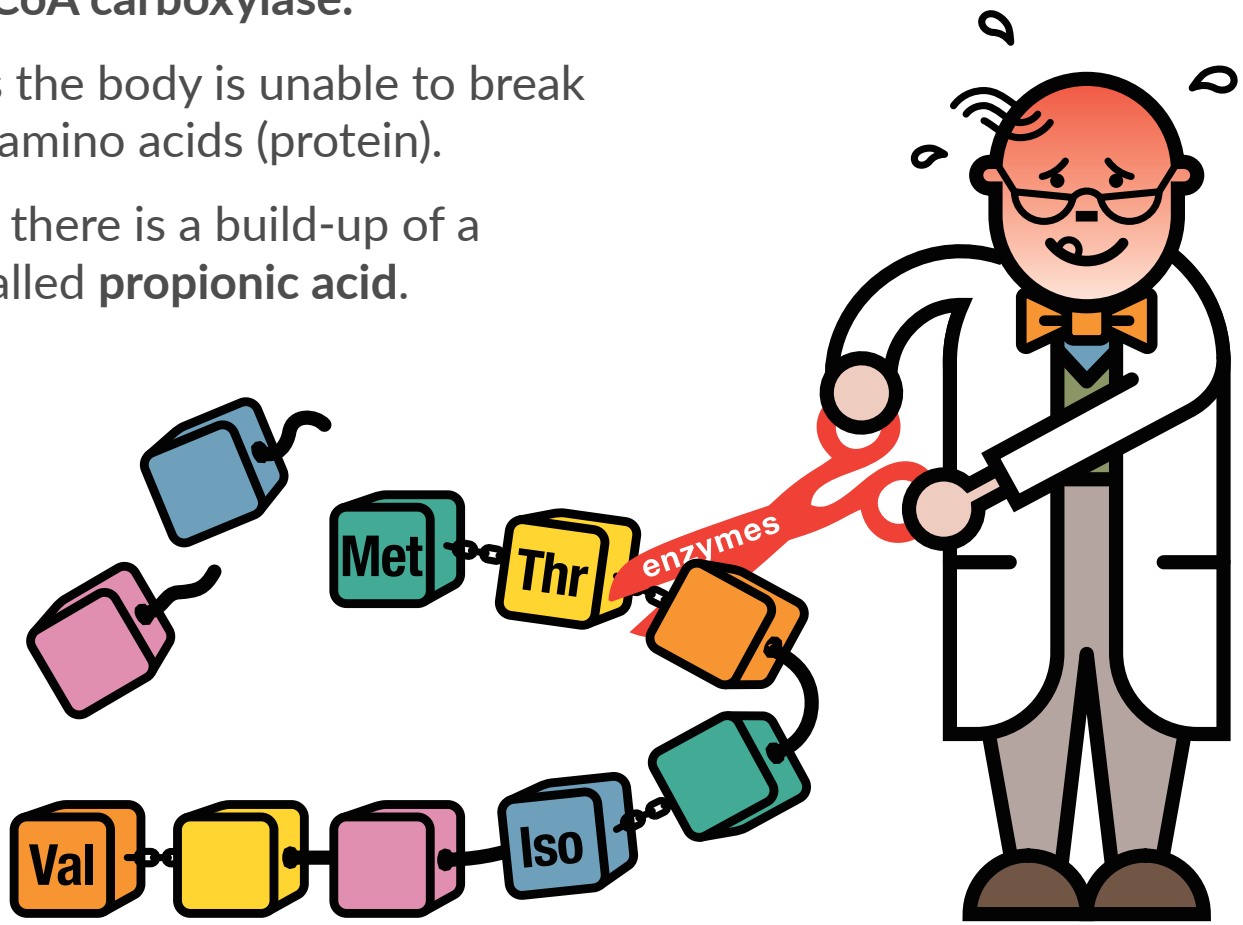


What happens in PA?

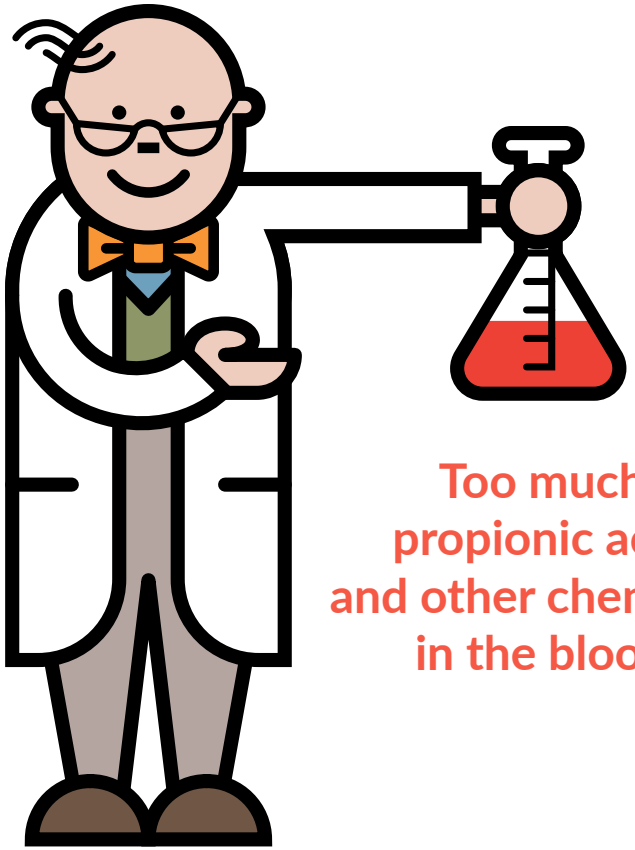
In PA, the body lacks an enzyme called **propionyl-CoA carboxylase.**

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

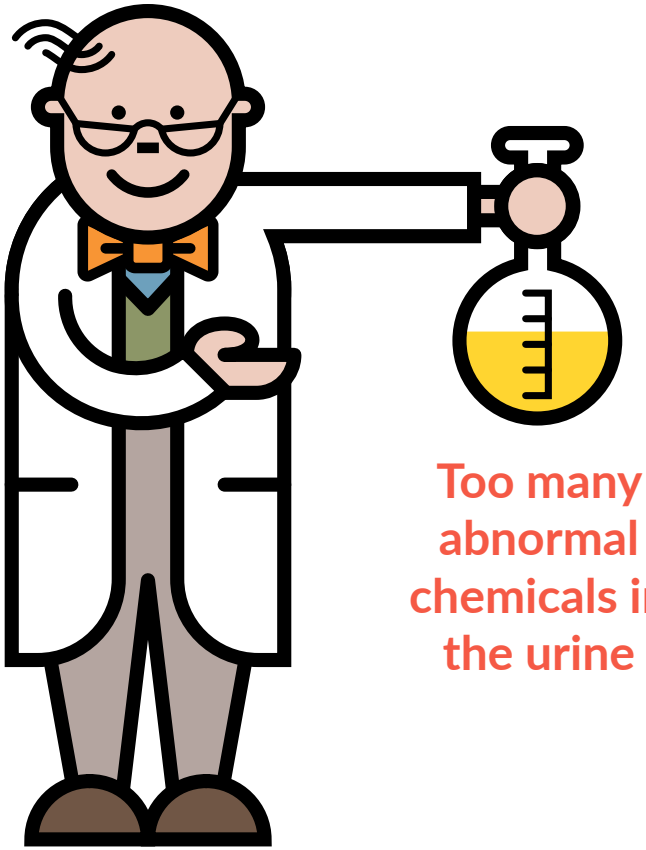
As a result, there is a build-up of a chemical called **propionic acid**.



What does this cause?



Too much propionic acid and other chemicals in the blood

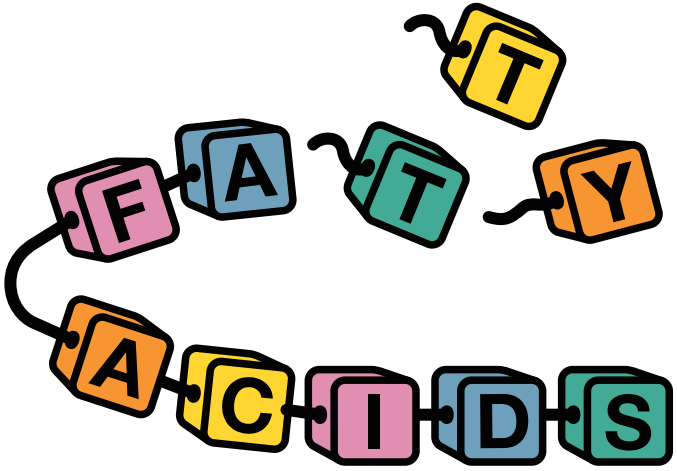


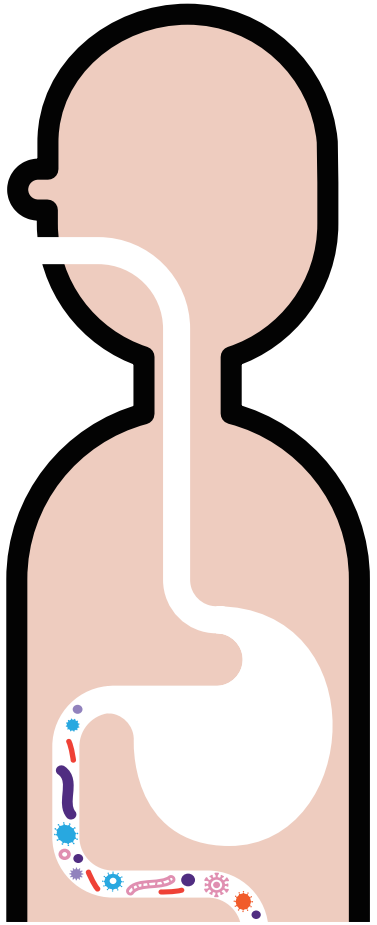
Too many abnormal chemicals in the urine

Other sources of propionic acid

Propionic 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 PA diagnosed?

PA is diagnosed by measuring chemicals containing propionic acid in the blood and urine. It can also be diagnosed by looking at enzyme levels and at the body's genes.

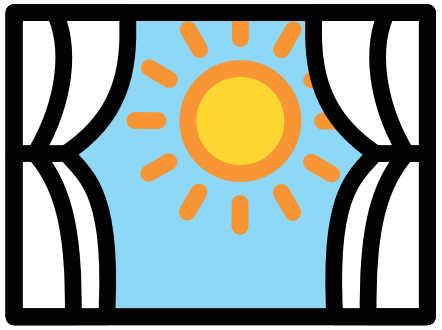
What are the symptoms in PA?

Symptoms commonly start 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 PA quickly become life-threatening if unmanaged



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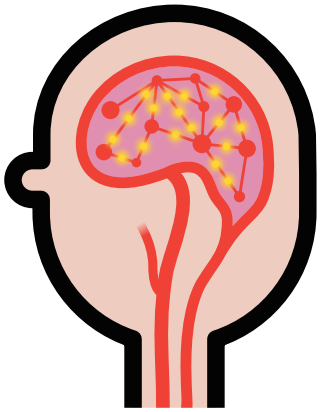
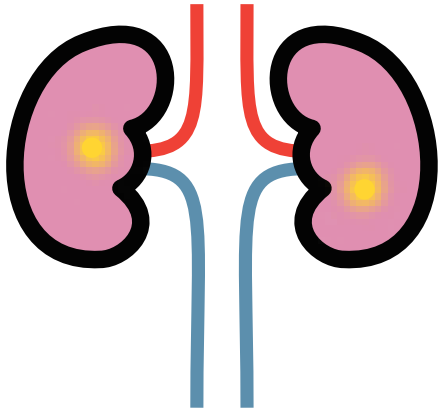
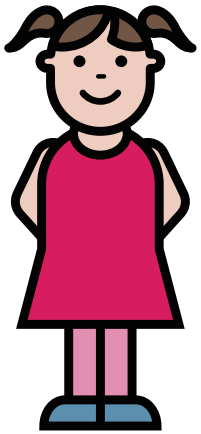
What can go wrong in PA?

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





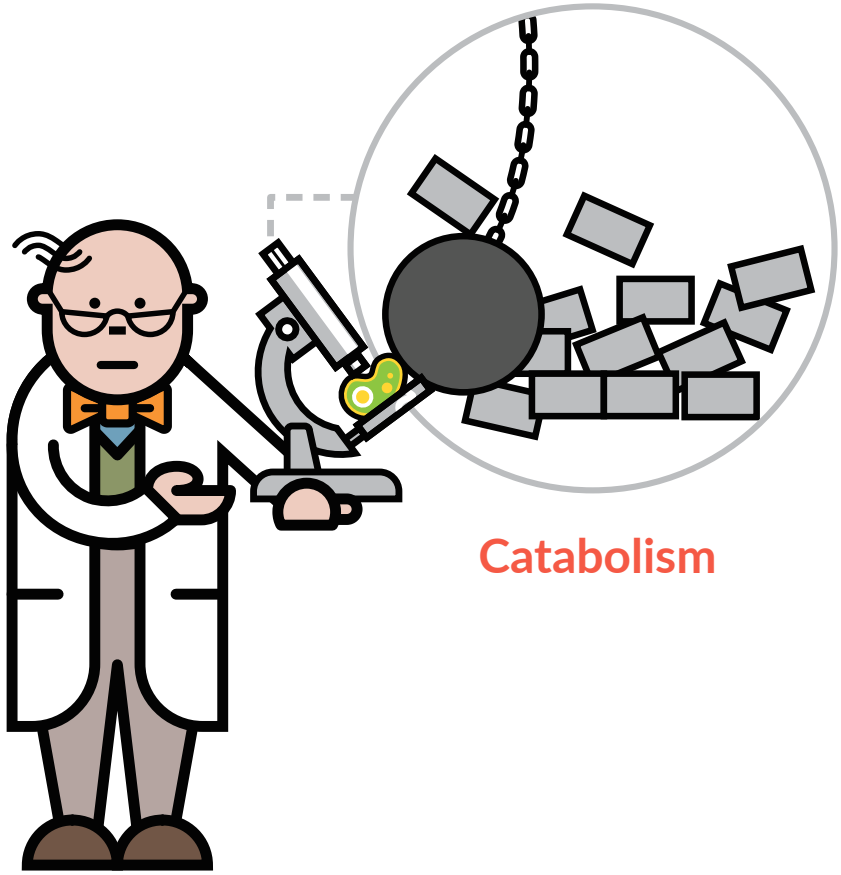
It may cause delays to normal development like walking and talking.



What else happens in PA?

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.

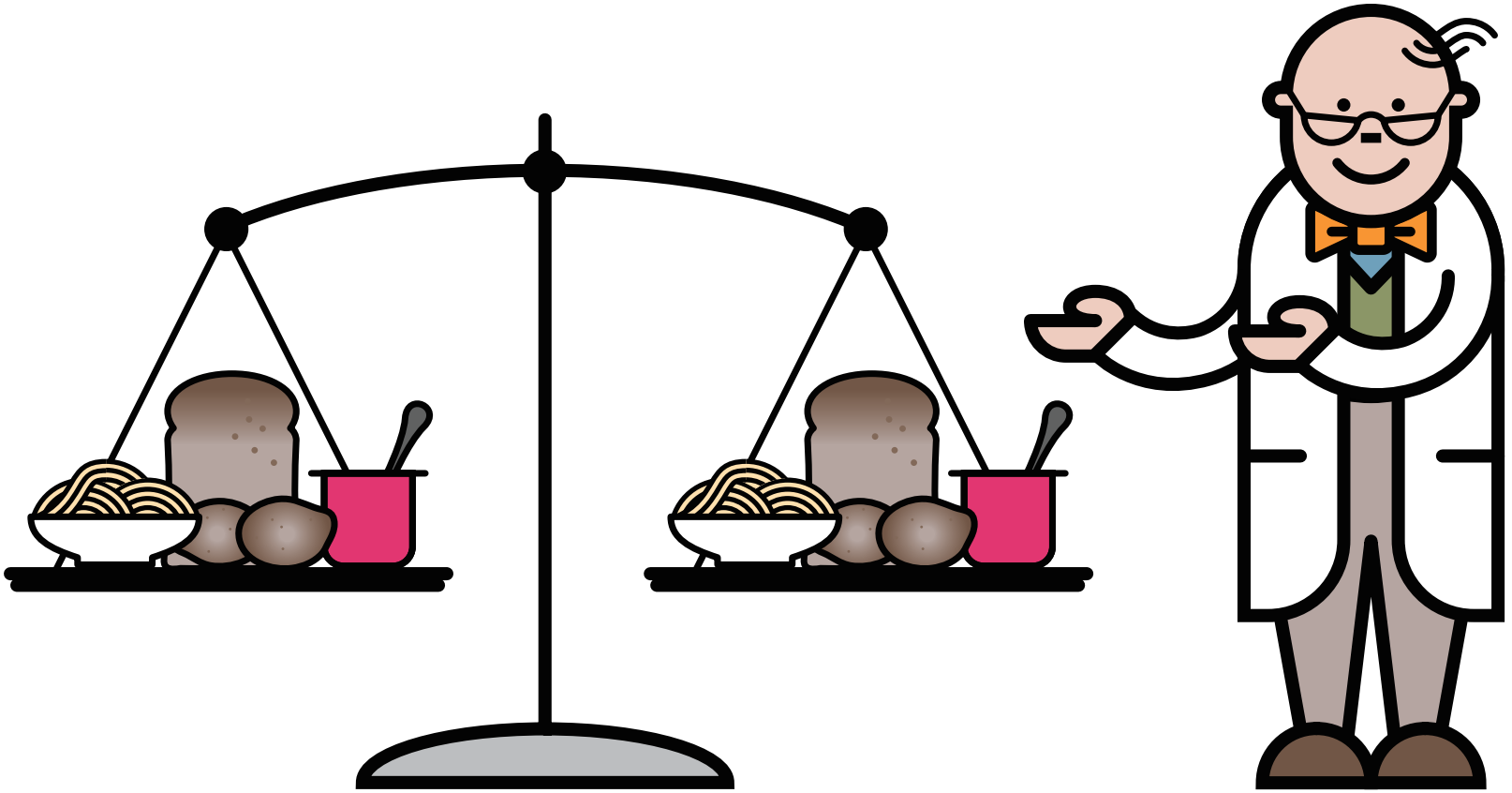


Metabolic crisis

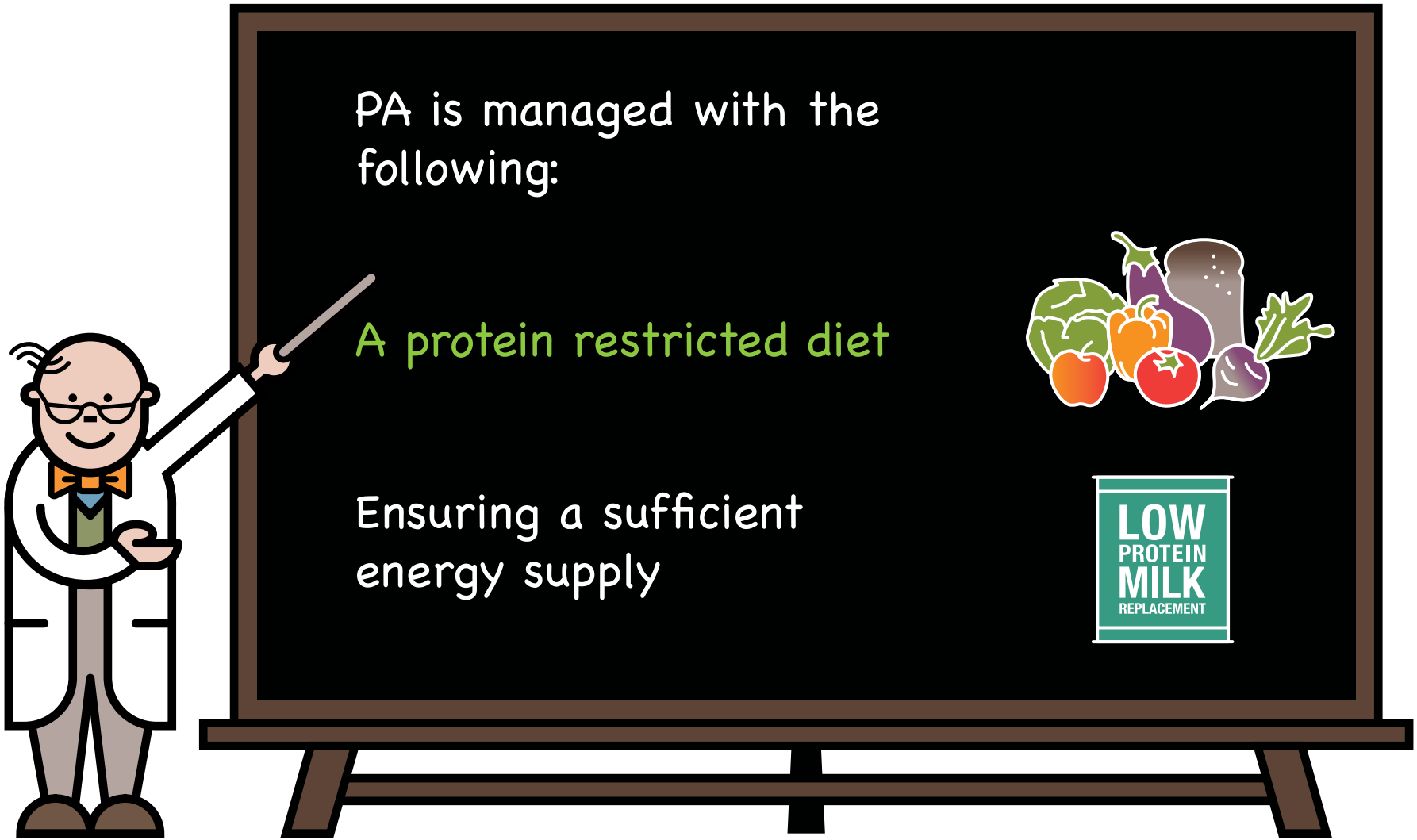
- In a **metabolic crisis** there is a build up of propionic acid and other toxic chemicals such as ammonia
- It is usually triggered by childhood illnesses e.g. vomiting and diarrhoea
  or fasting for too long
- There should be no delay in management
- Avoidance of a metabolic crisis is essential

Protein balance is needed in PA

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



How is PA managed day to day?



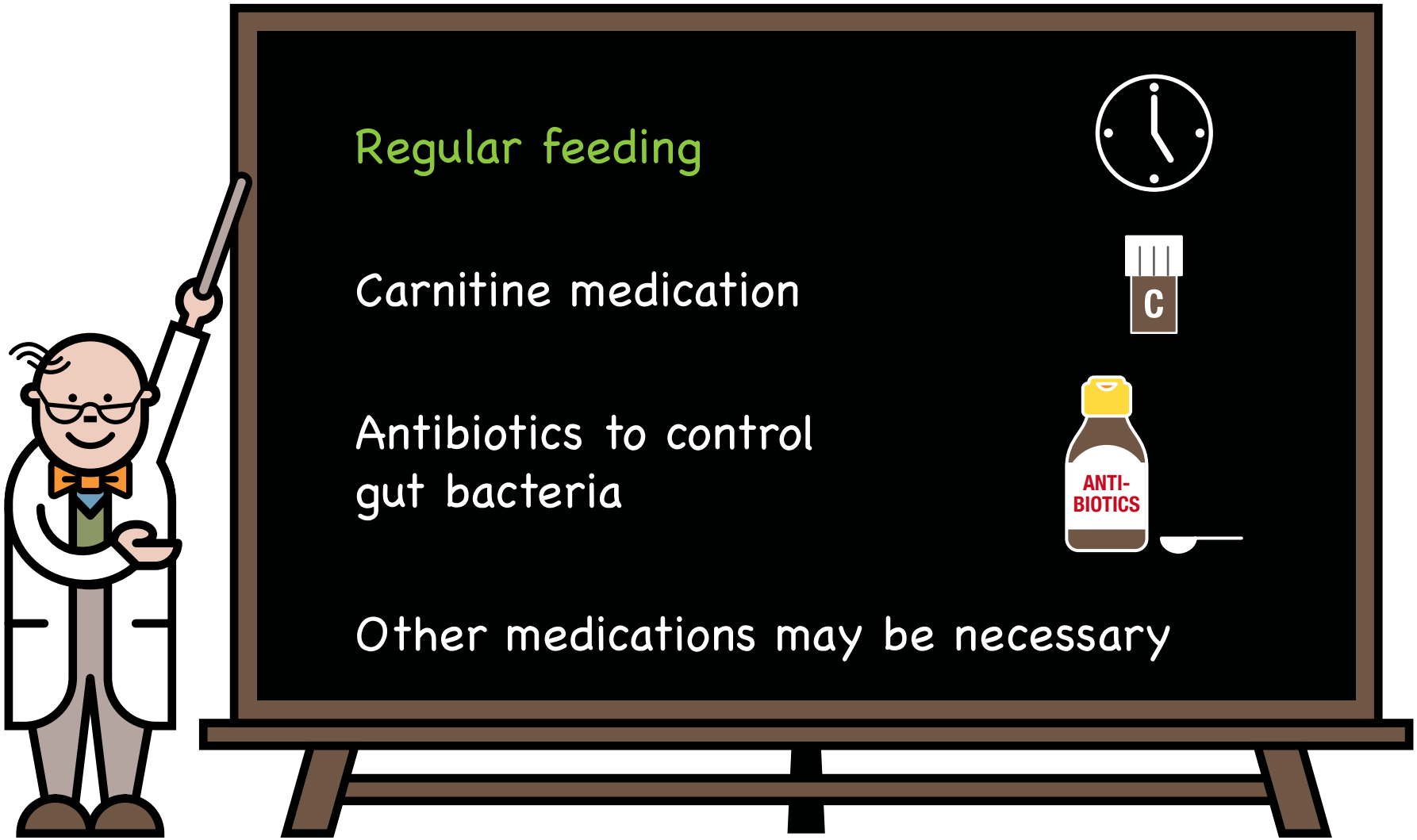
How is PA managed day to day?

PA is managed with the following:

A protein restricted diet

Ensuring a sufficient energy supply

How is PA managed day to day?



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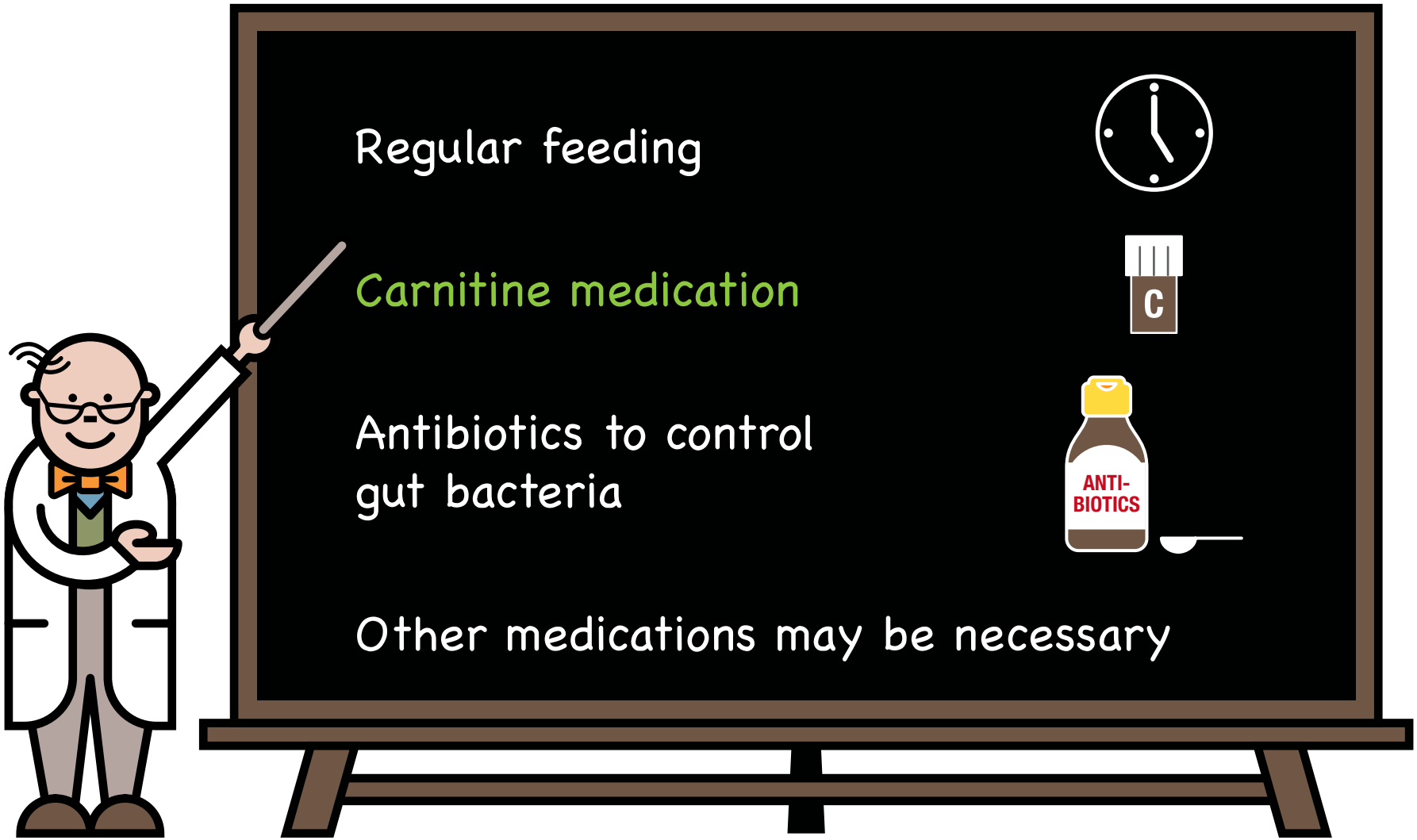

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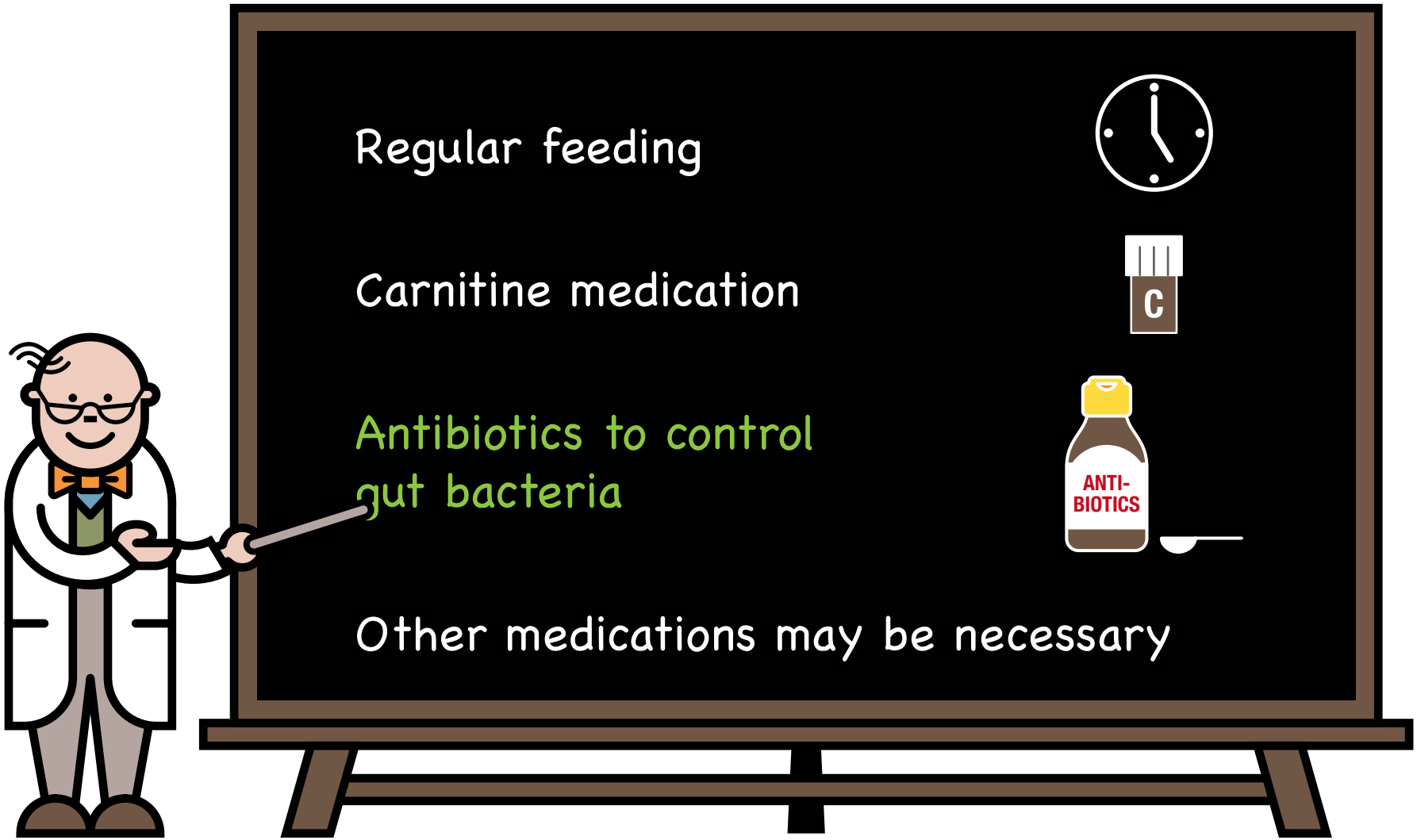

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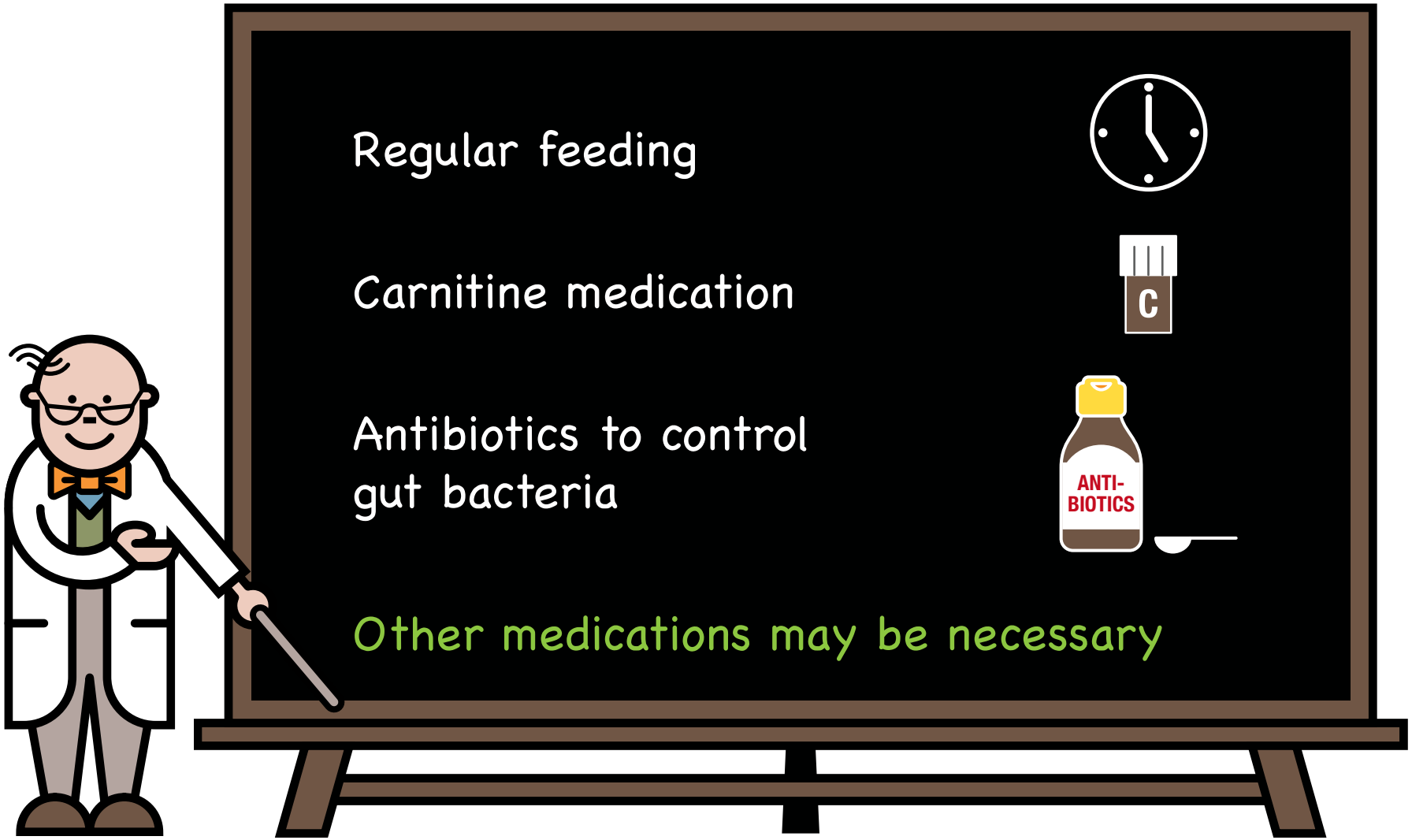
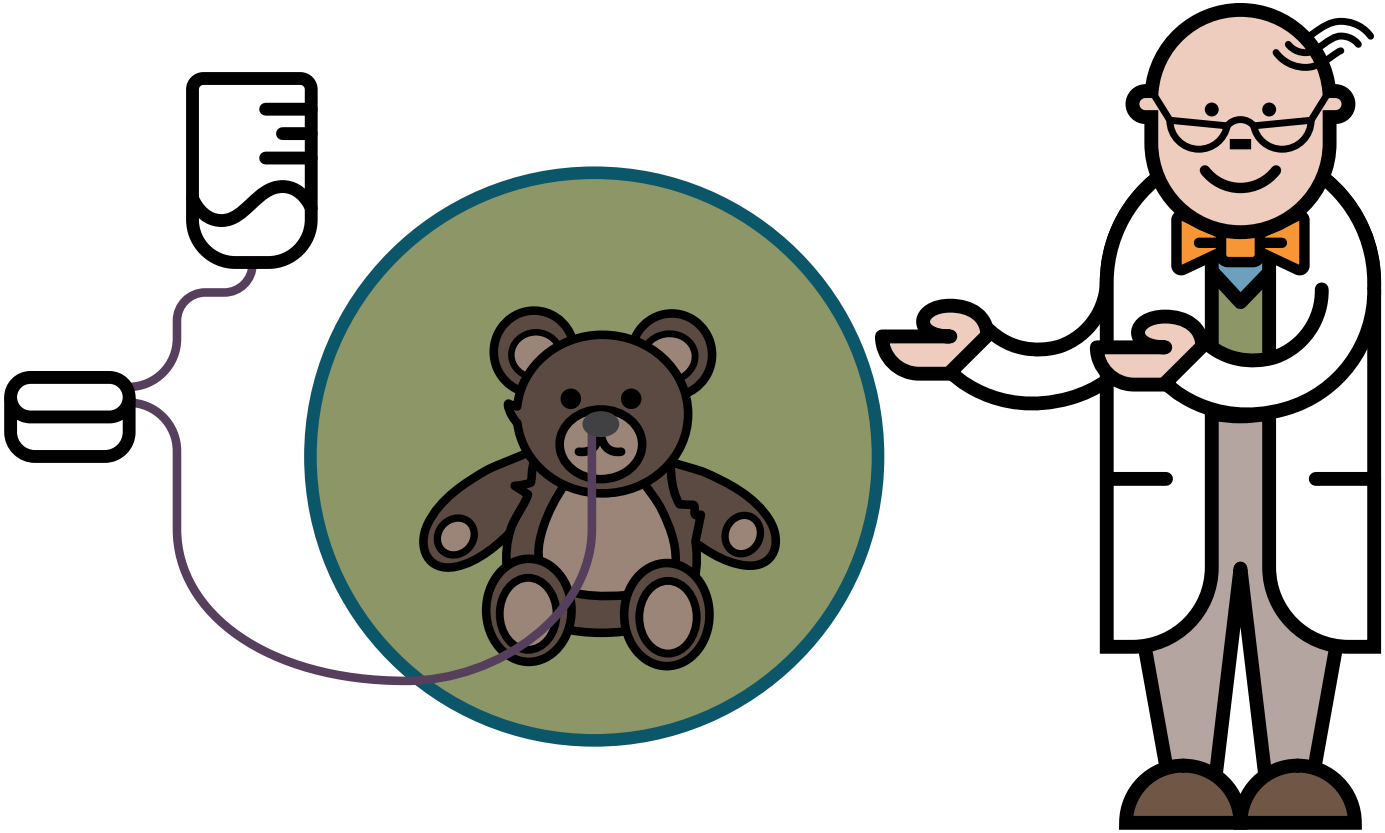


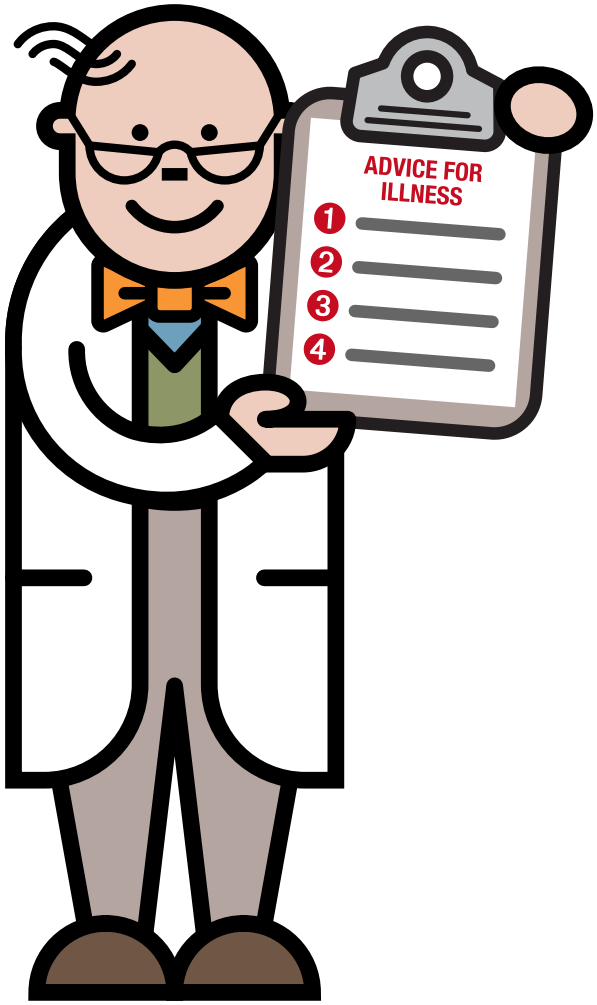
Is tube feeding needed?

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.

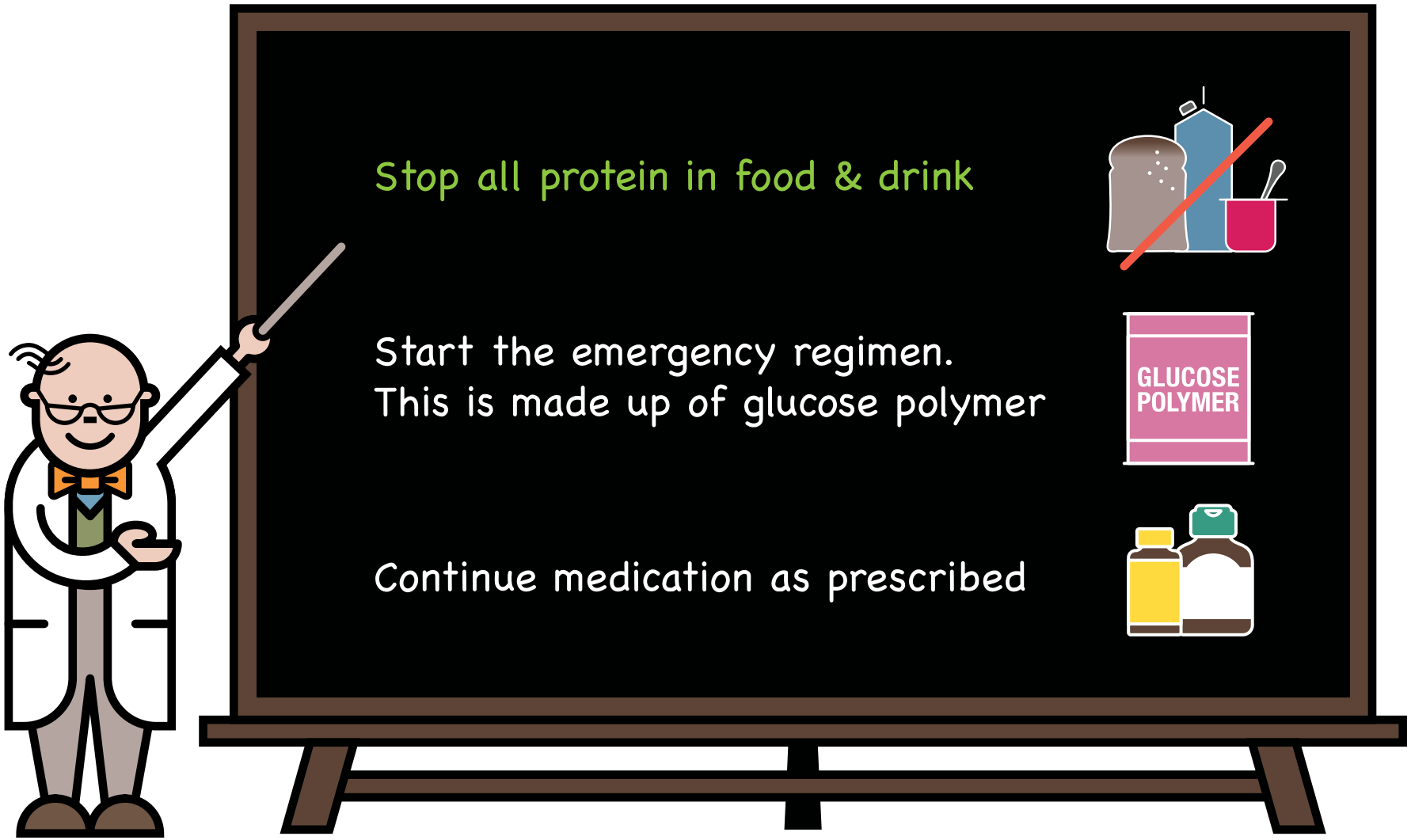


How is PA managed during illness?

- 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 PA managed during illness?



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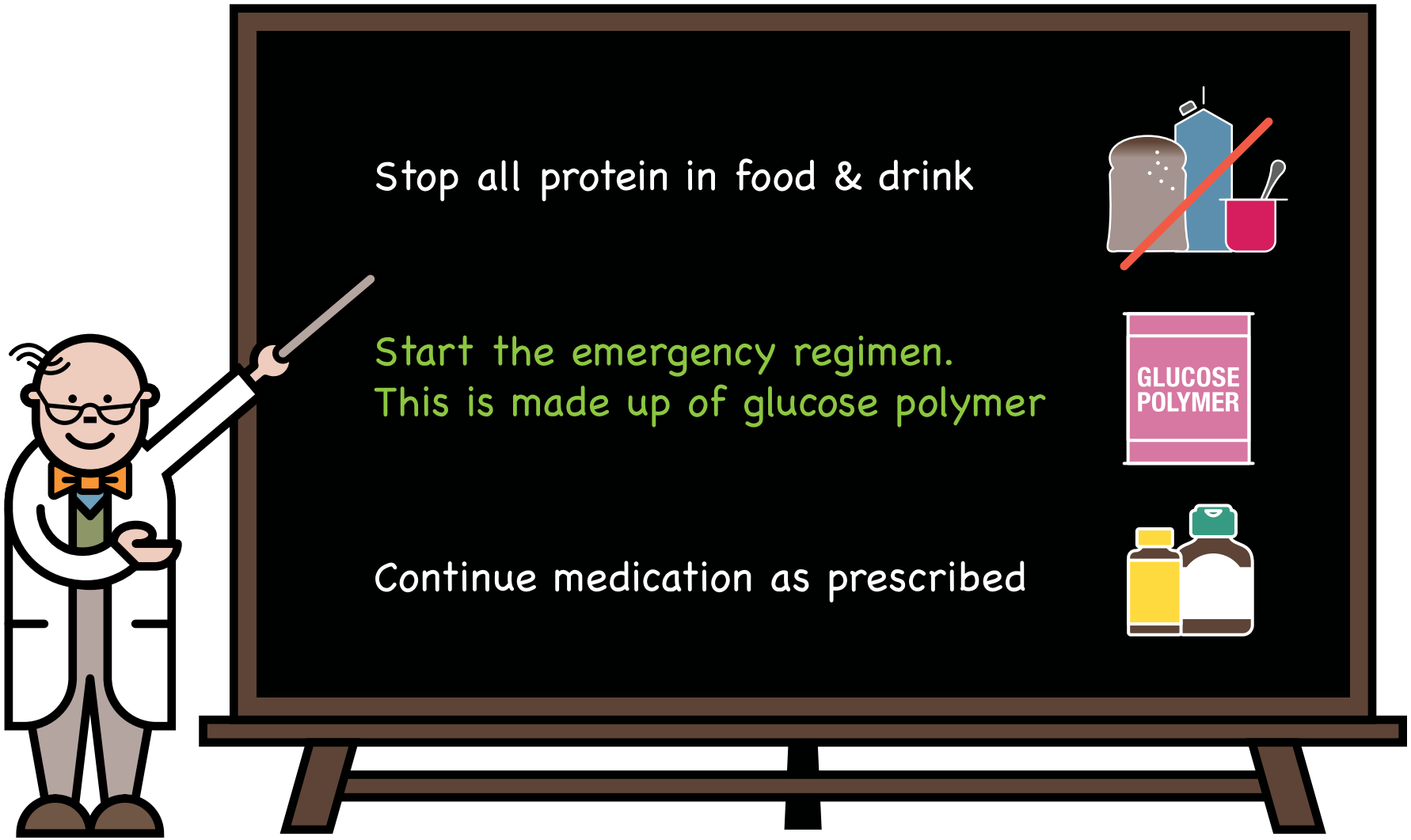

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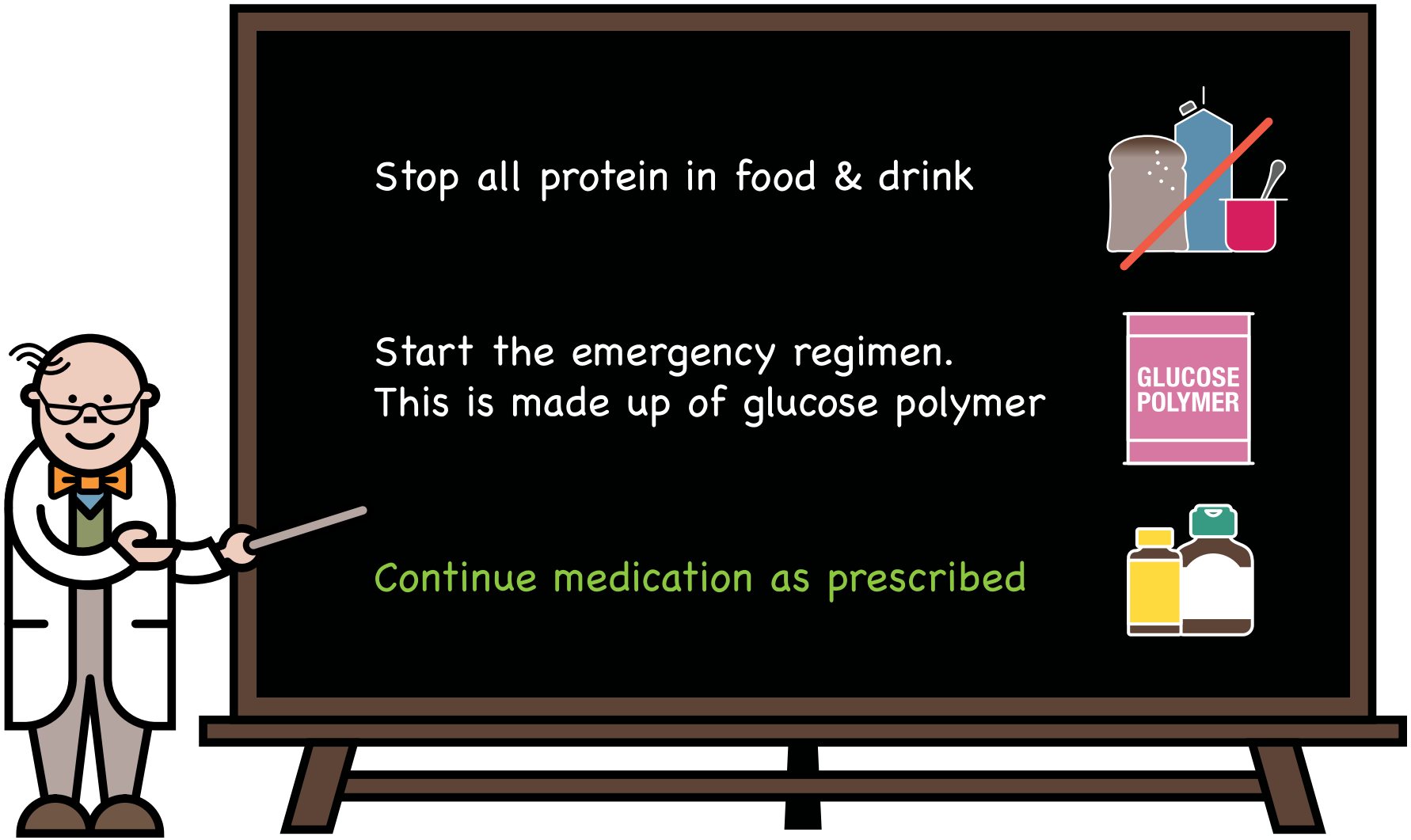
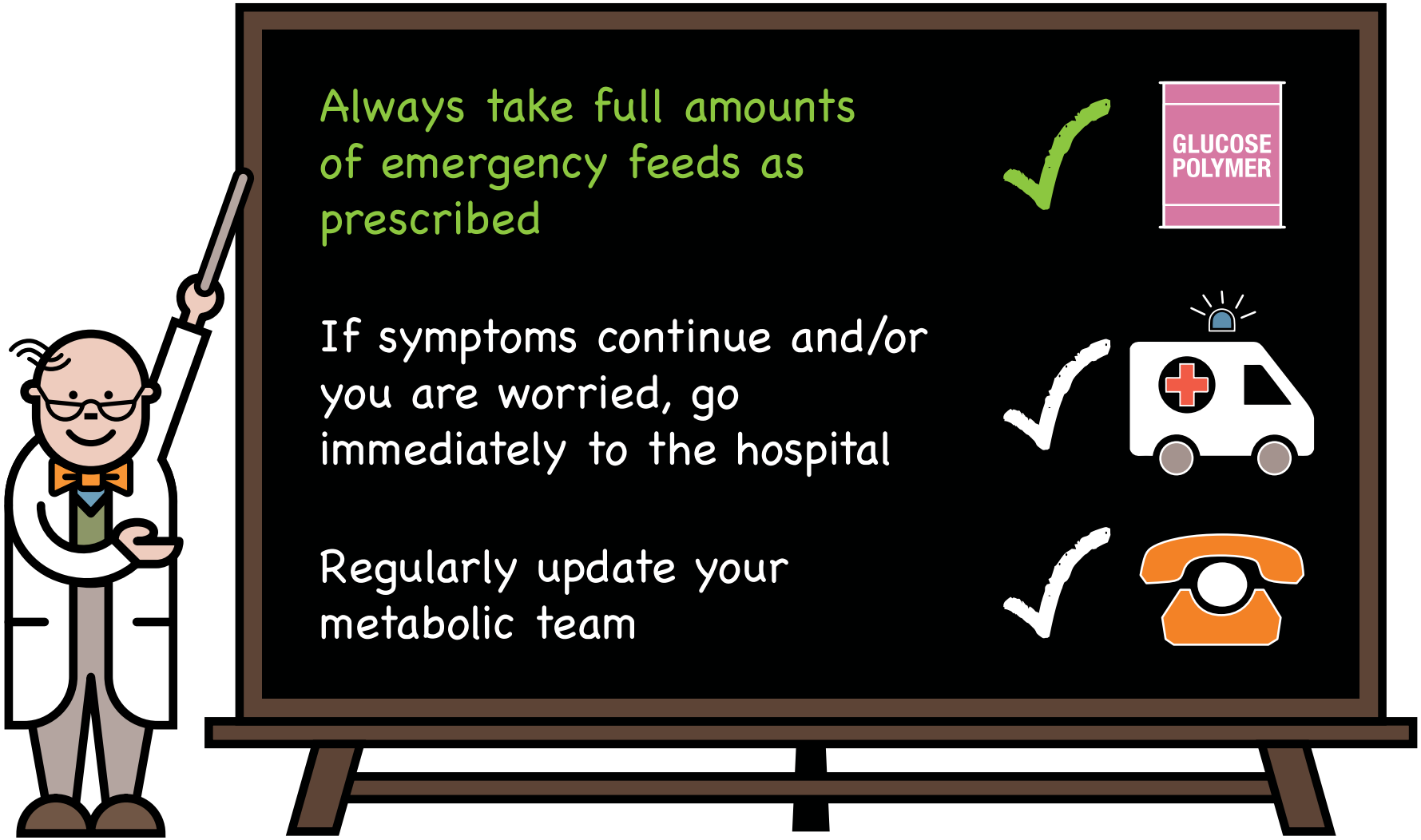


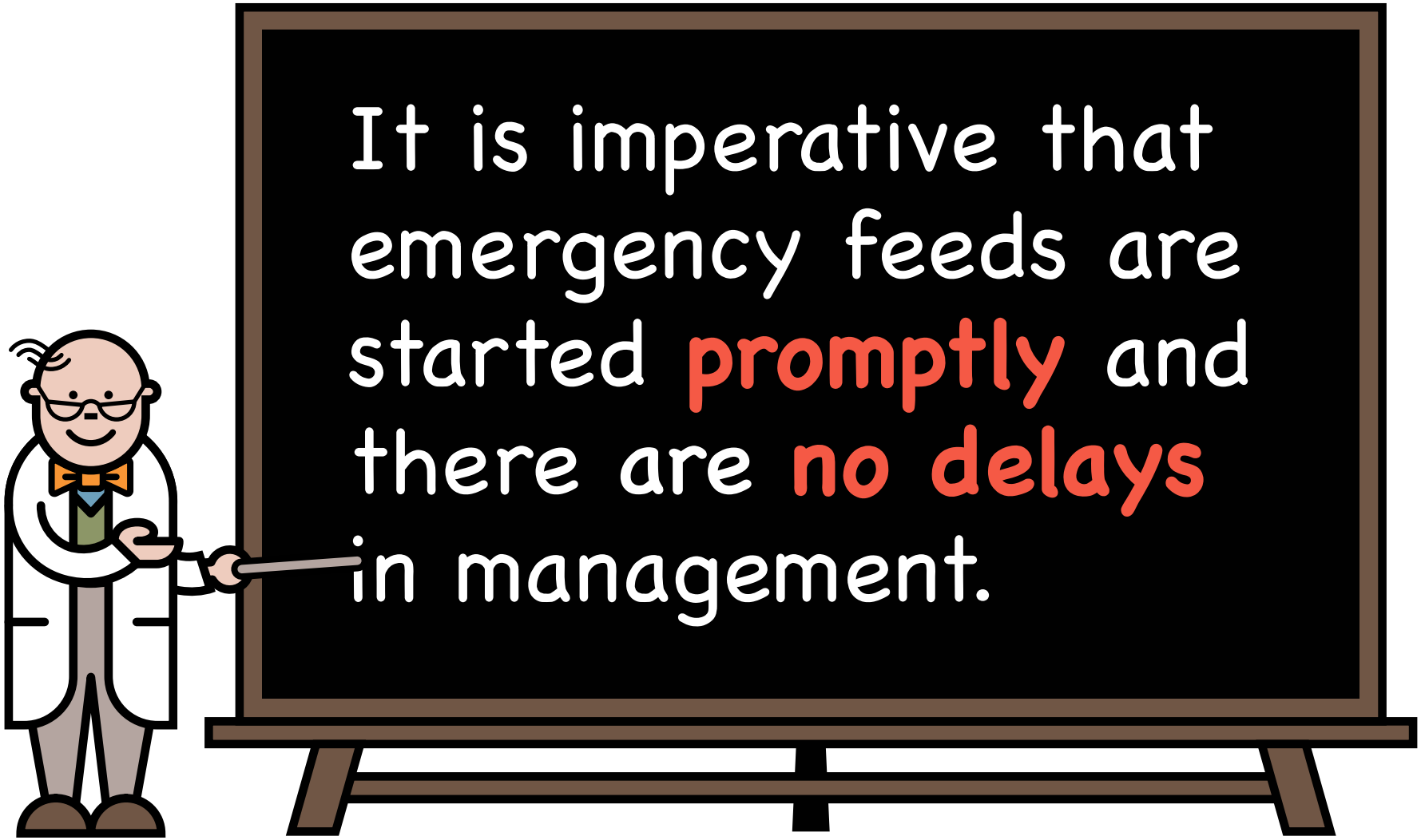
Checklist for illness



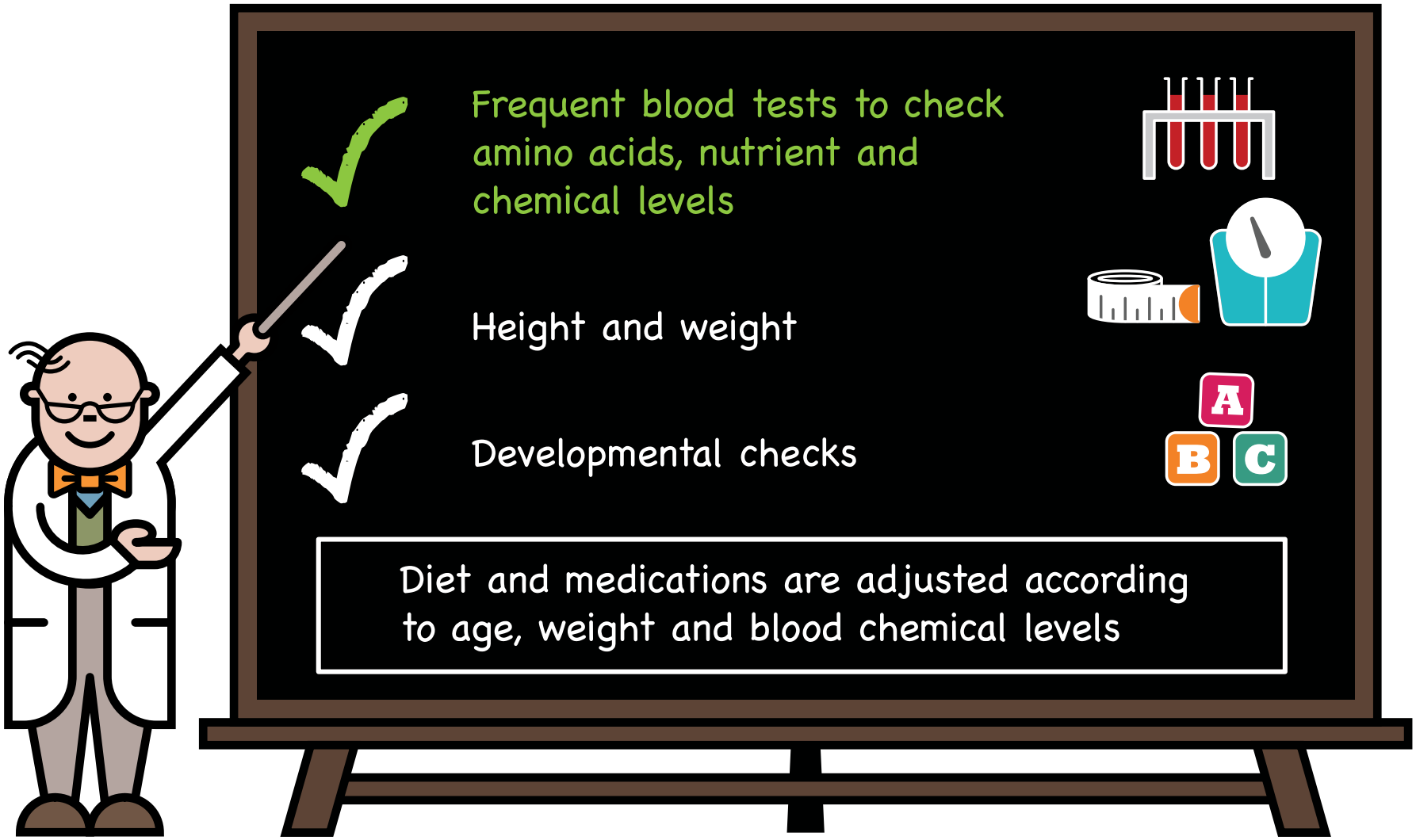
Checklist for illness

Checklist for illness

Key message



How is PA monitored?



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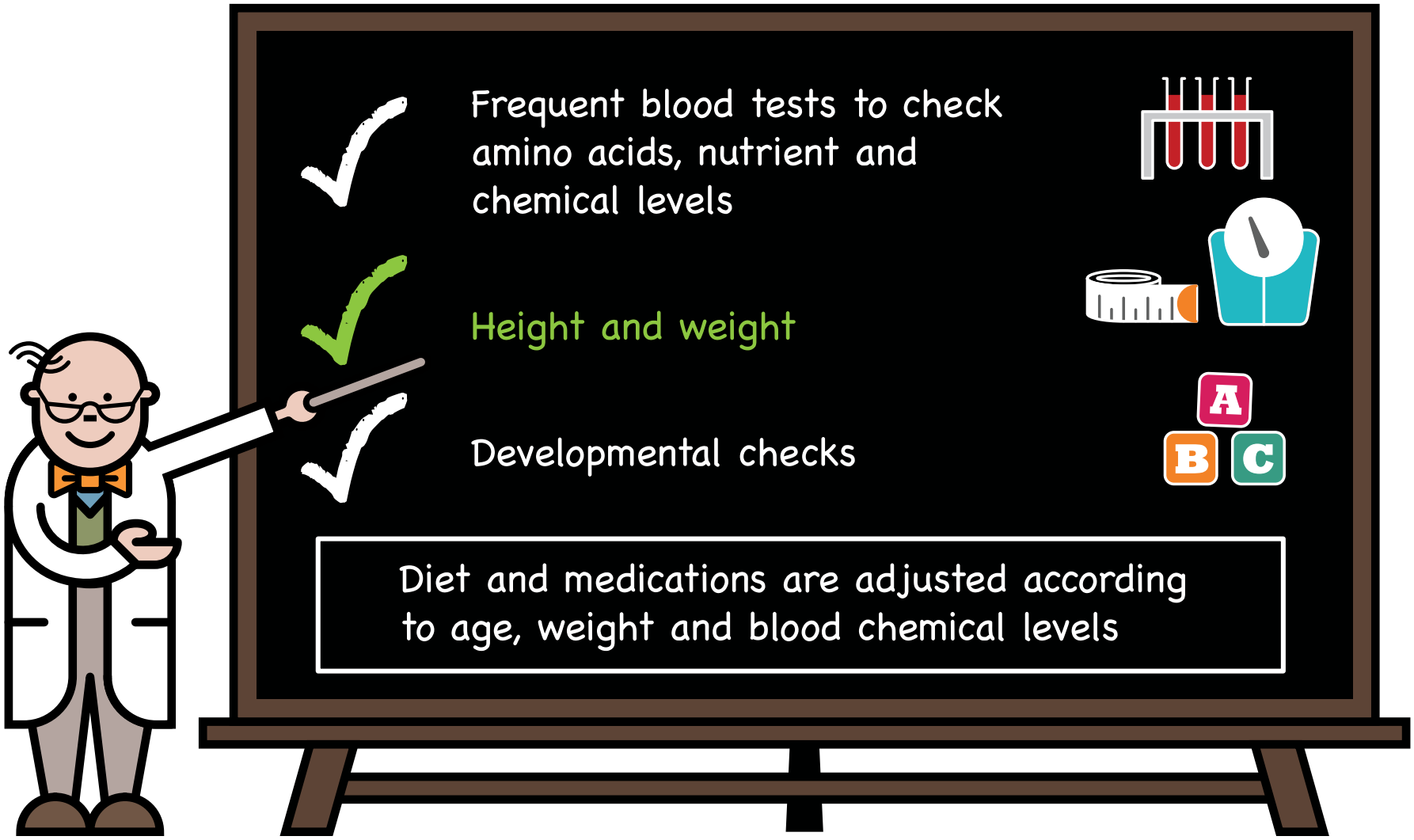

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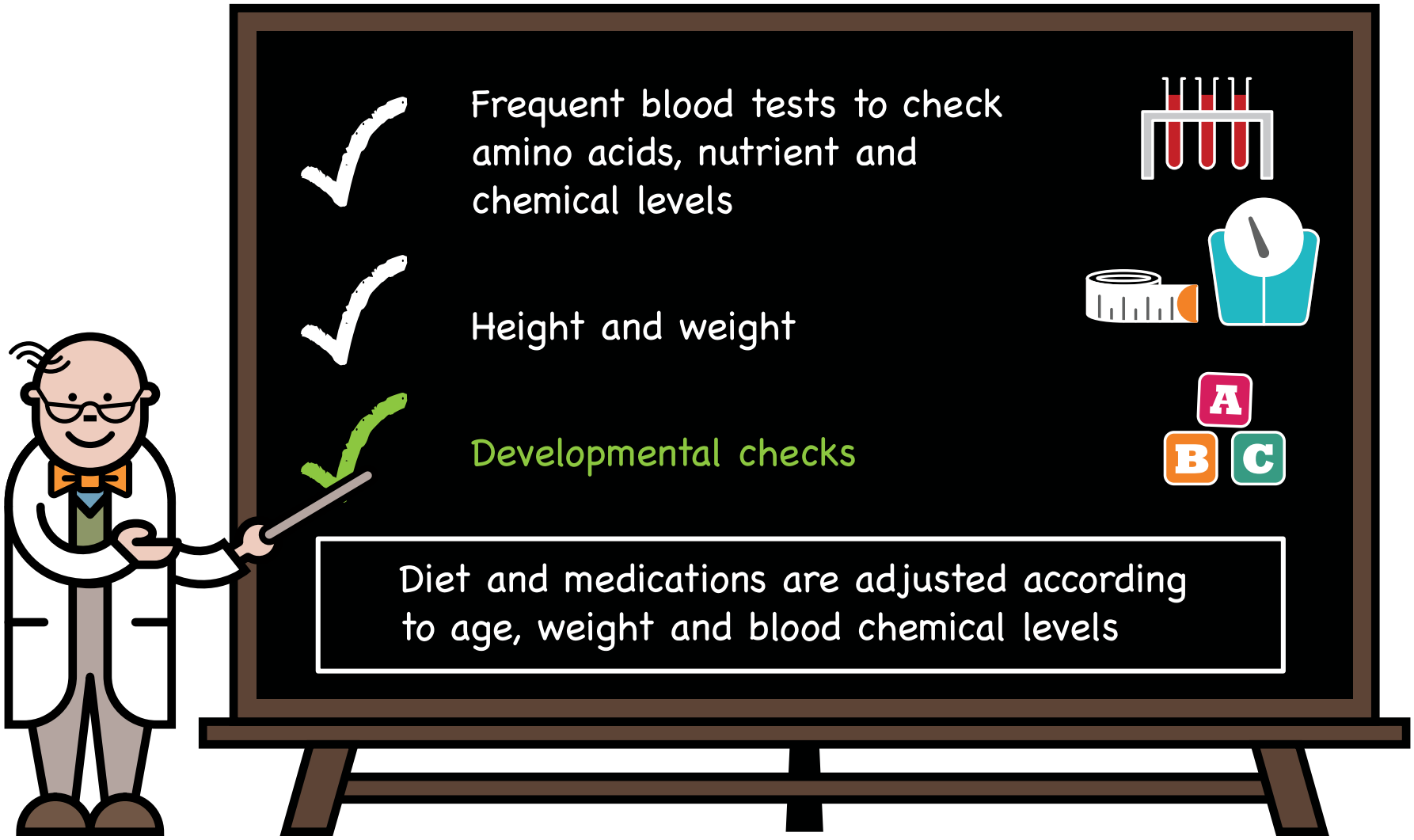

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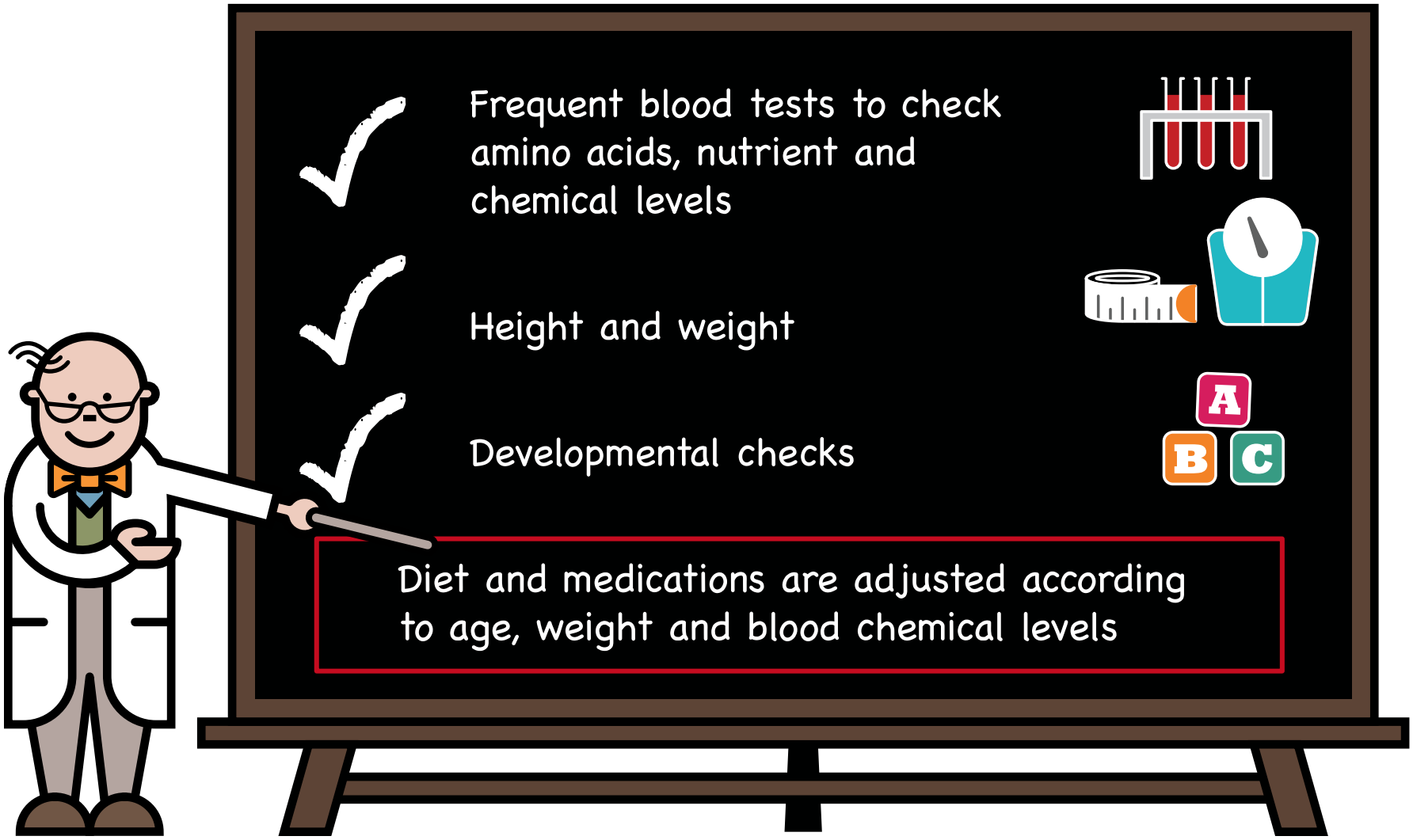
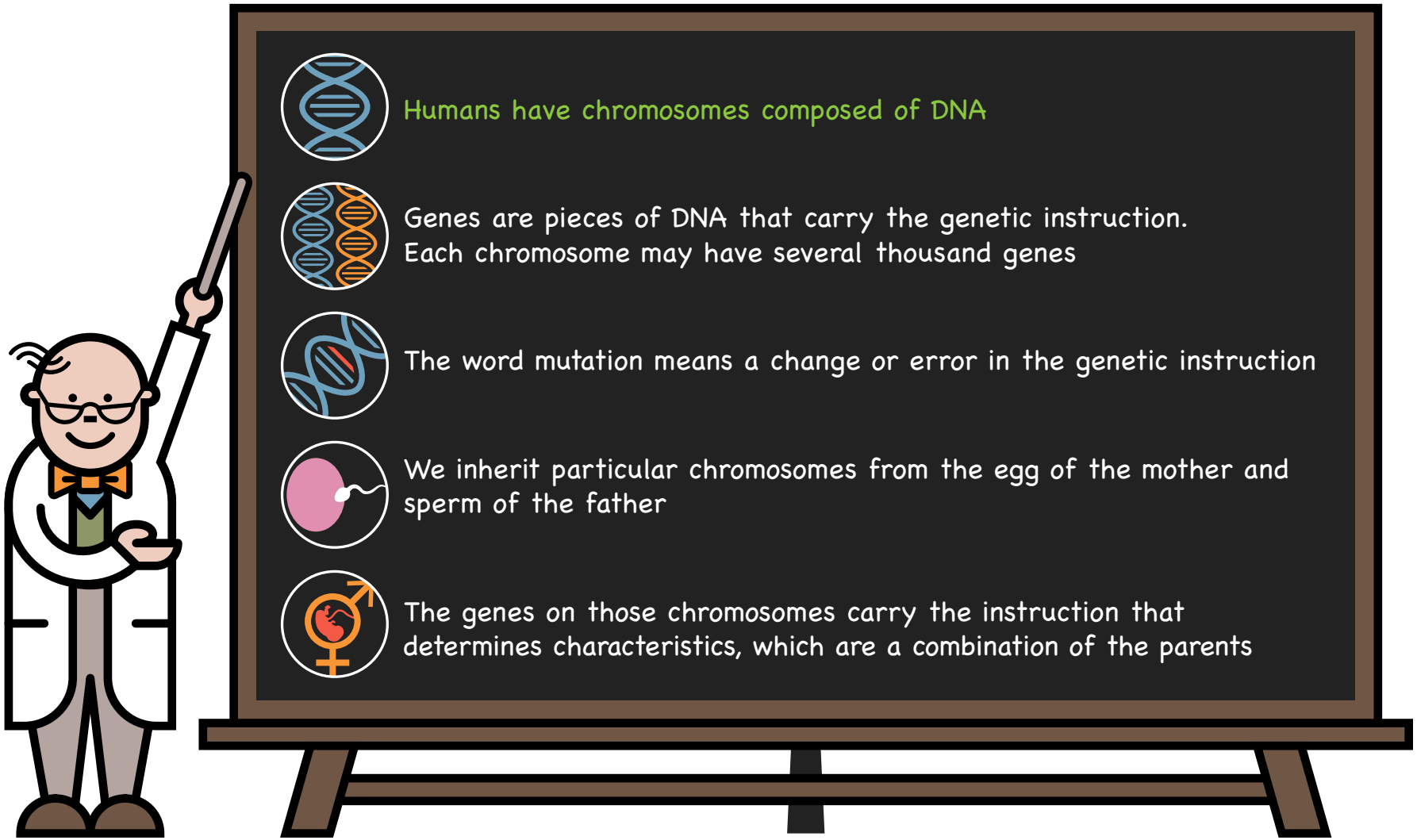


Chromosomes, genes, mutations



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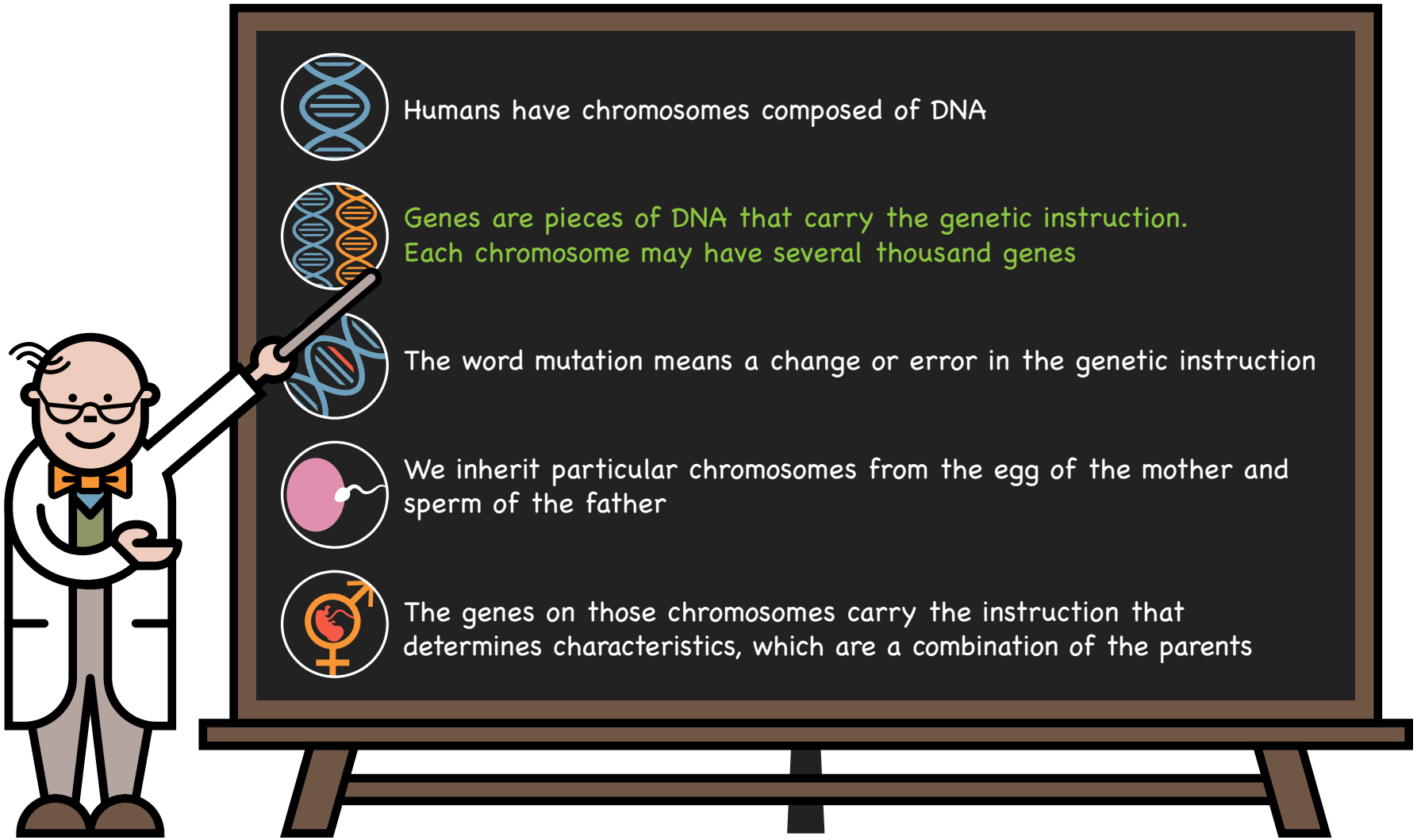
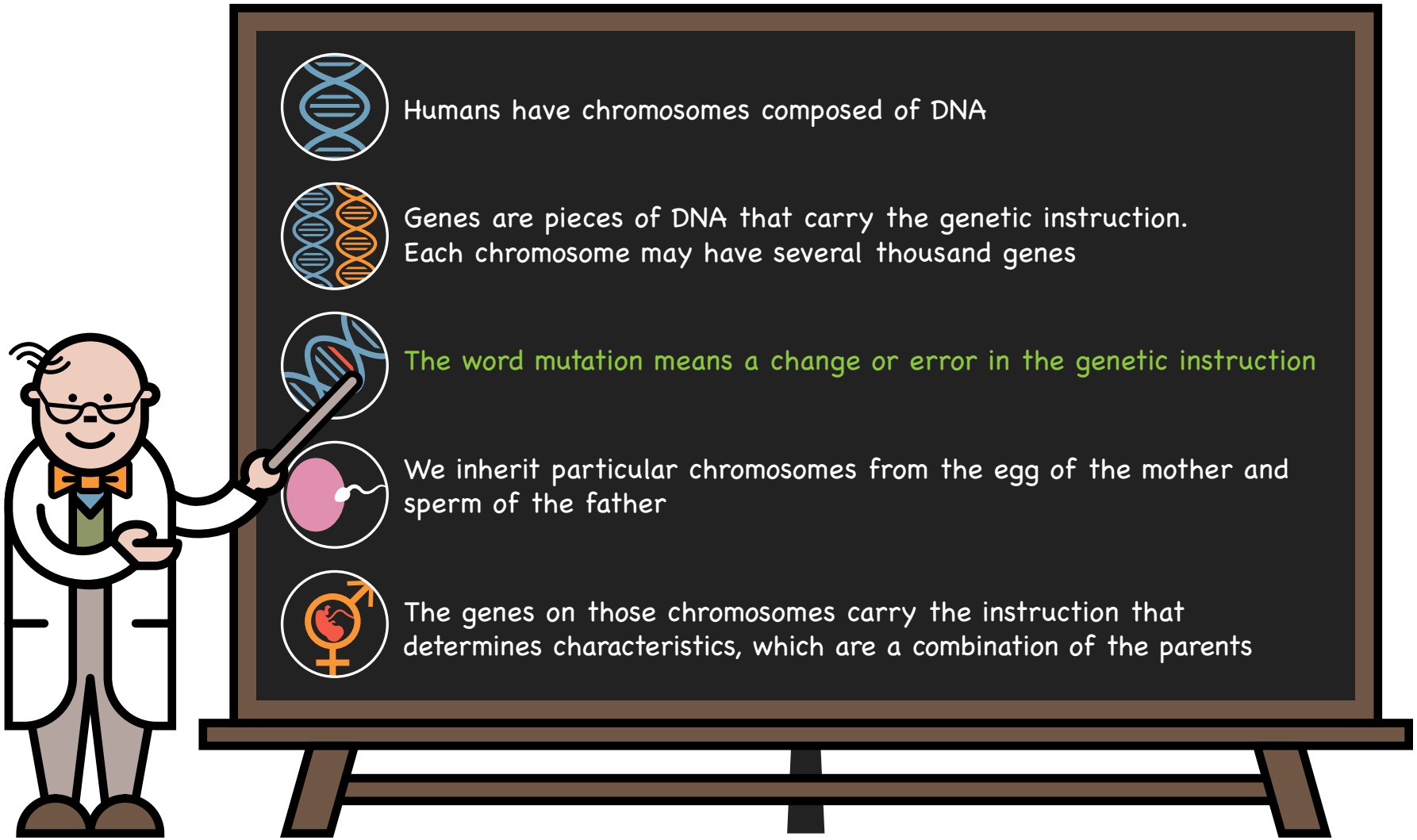


Chromosomes, genes, mutations



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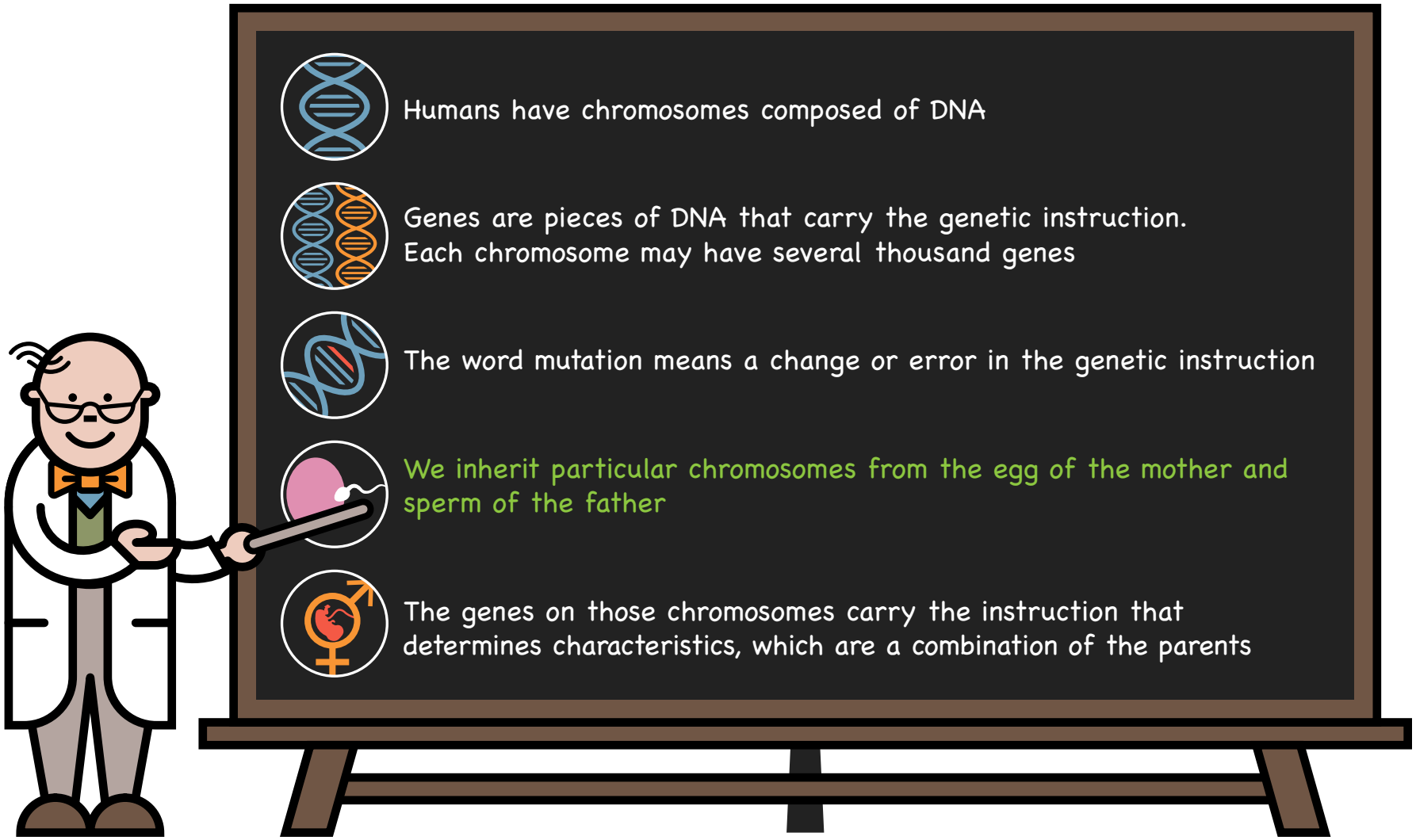

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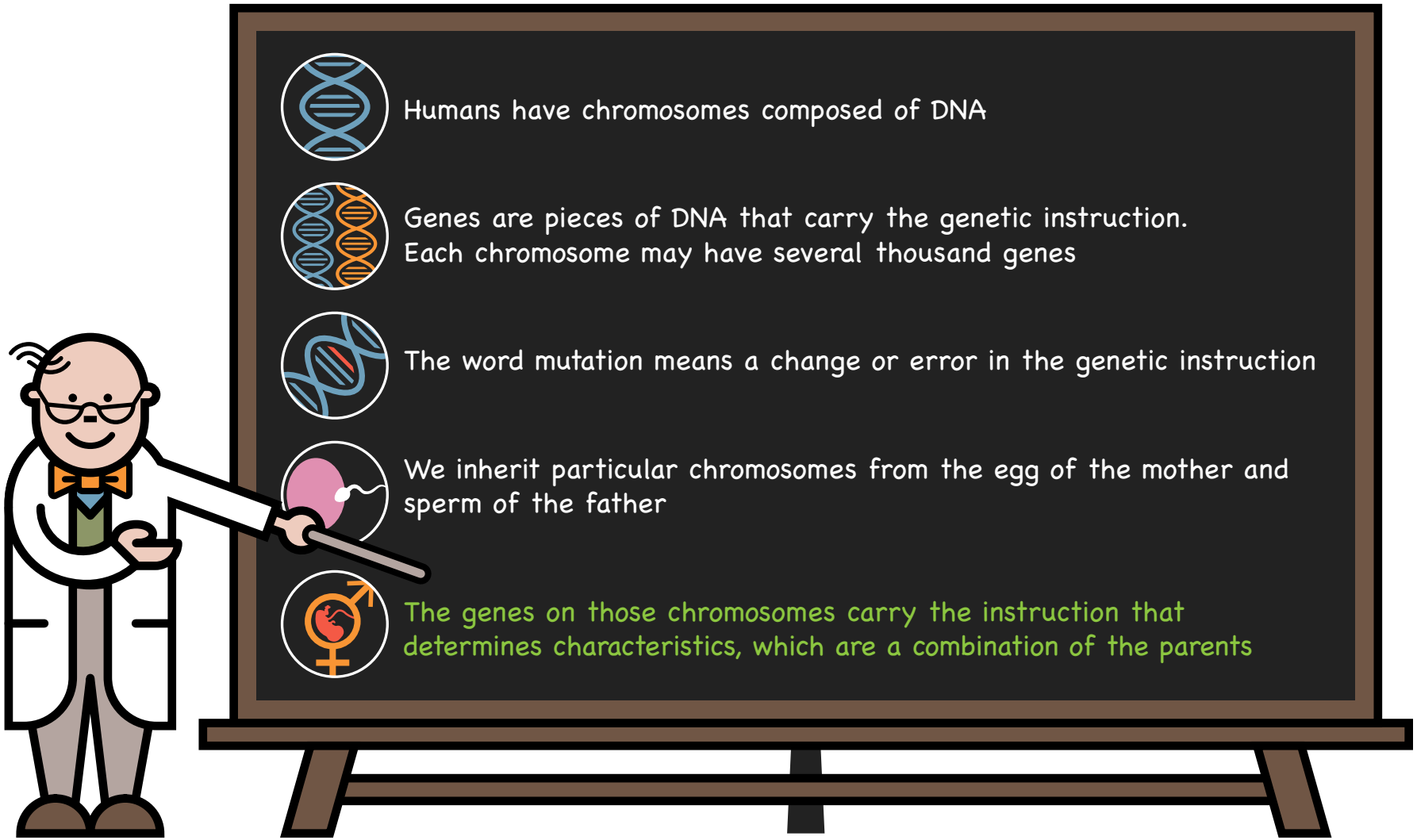
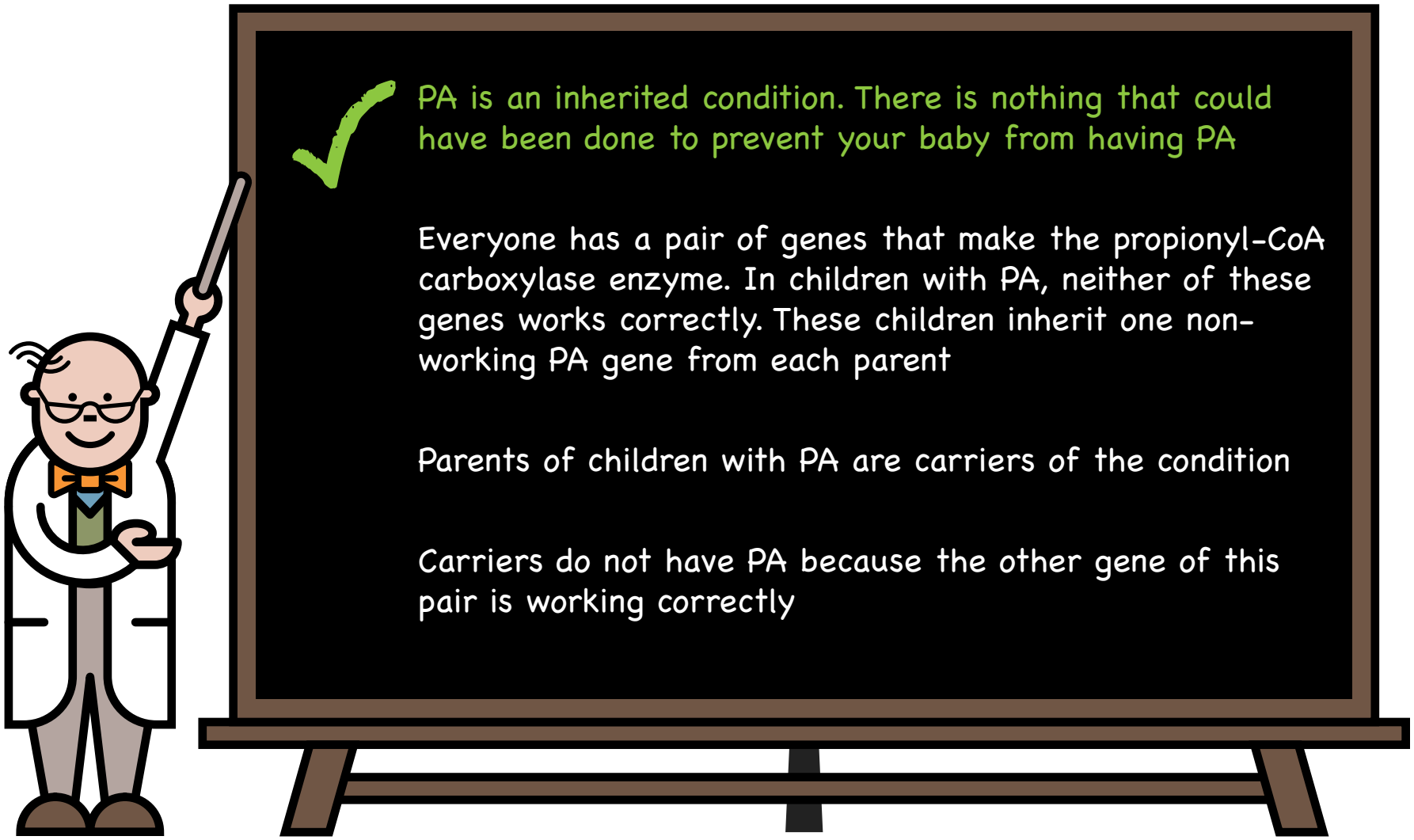


Inheritance



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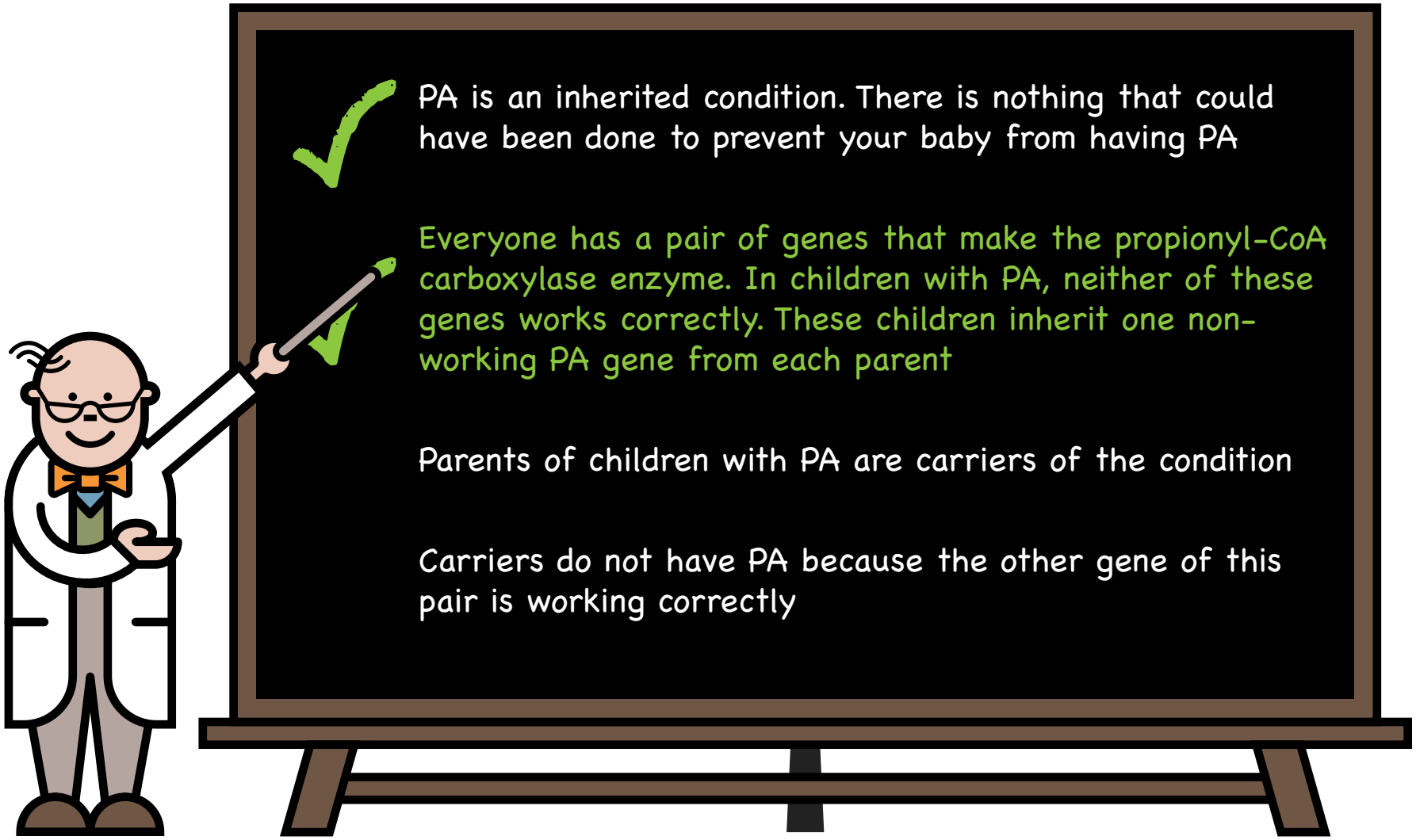

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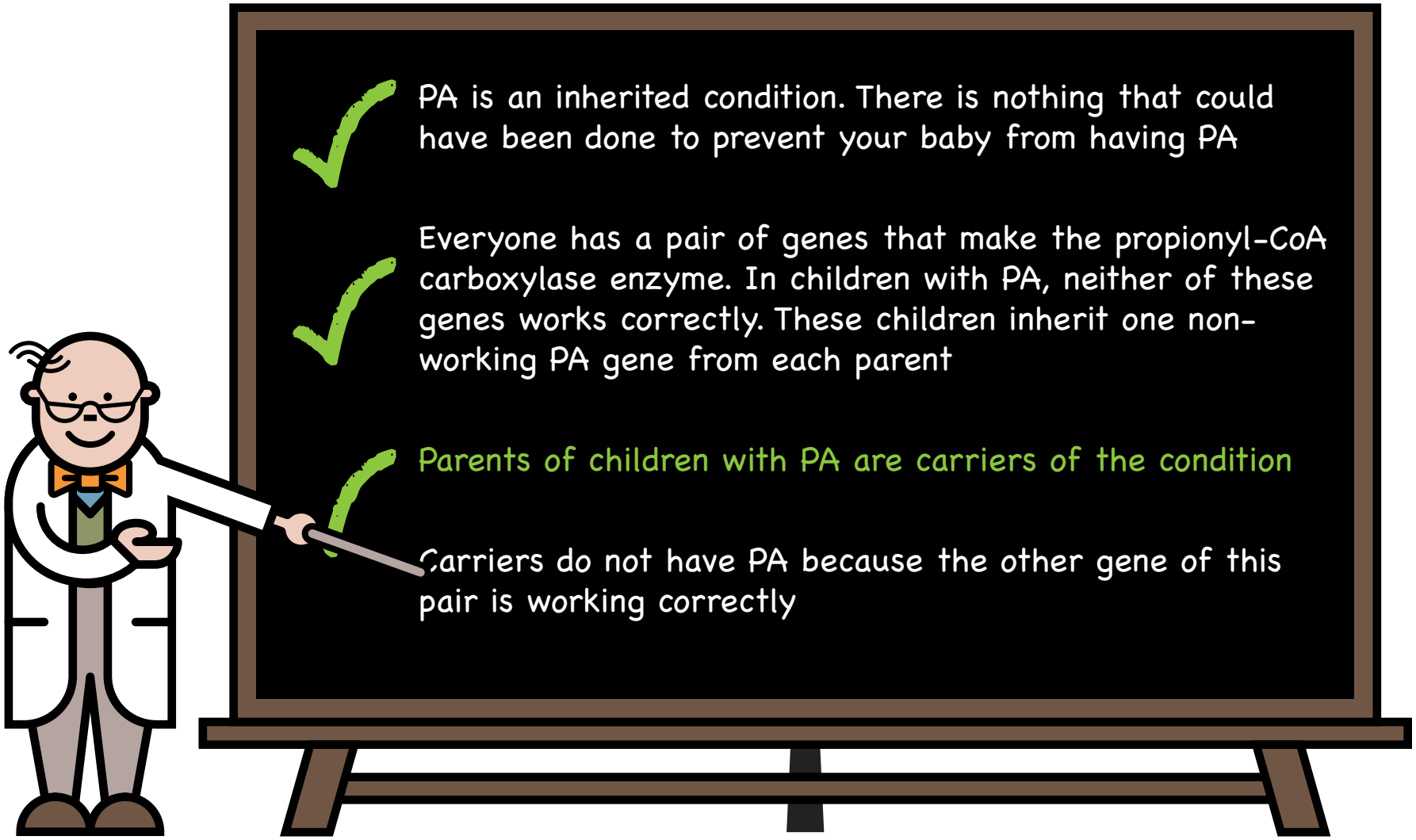

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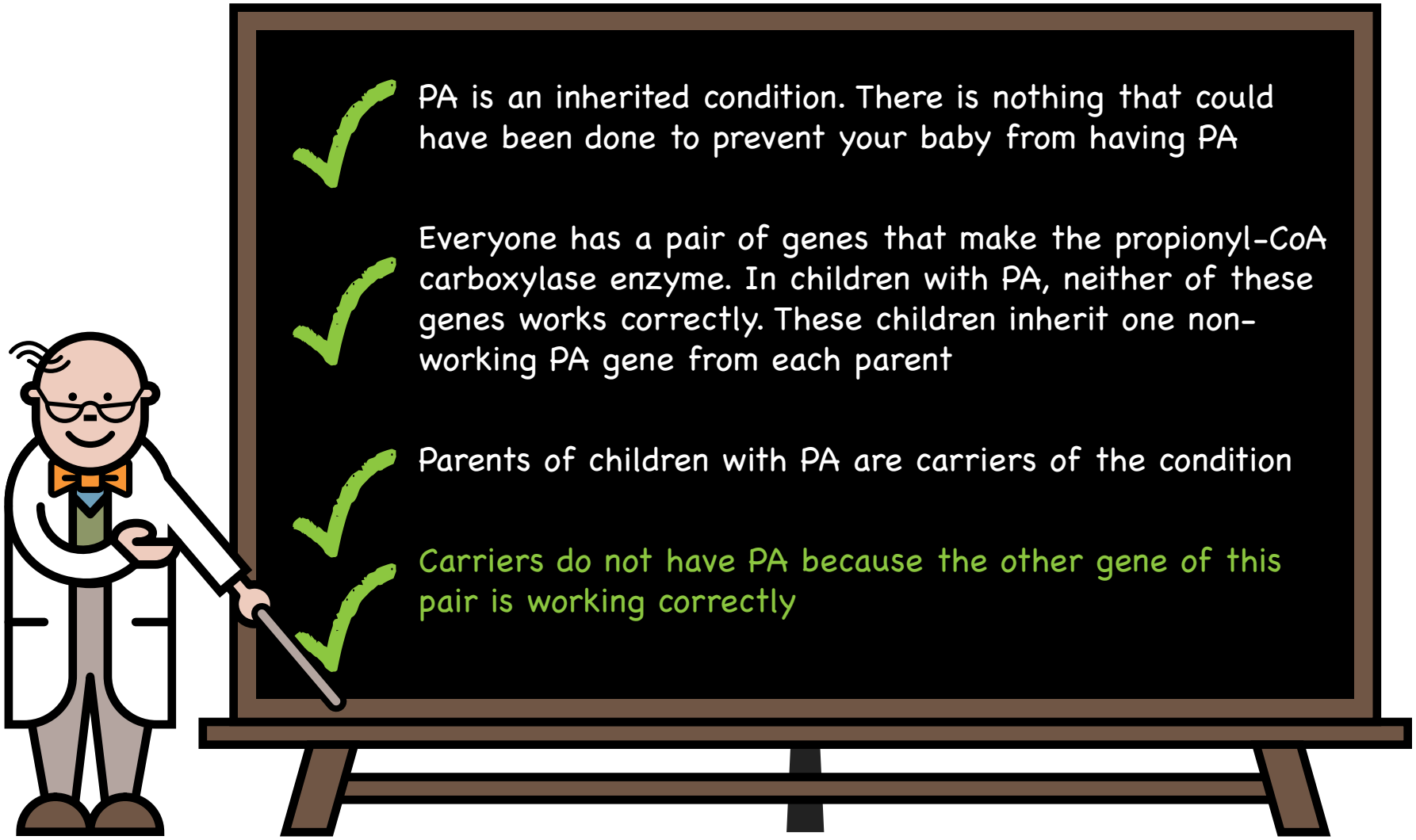
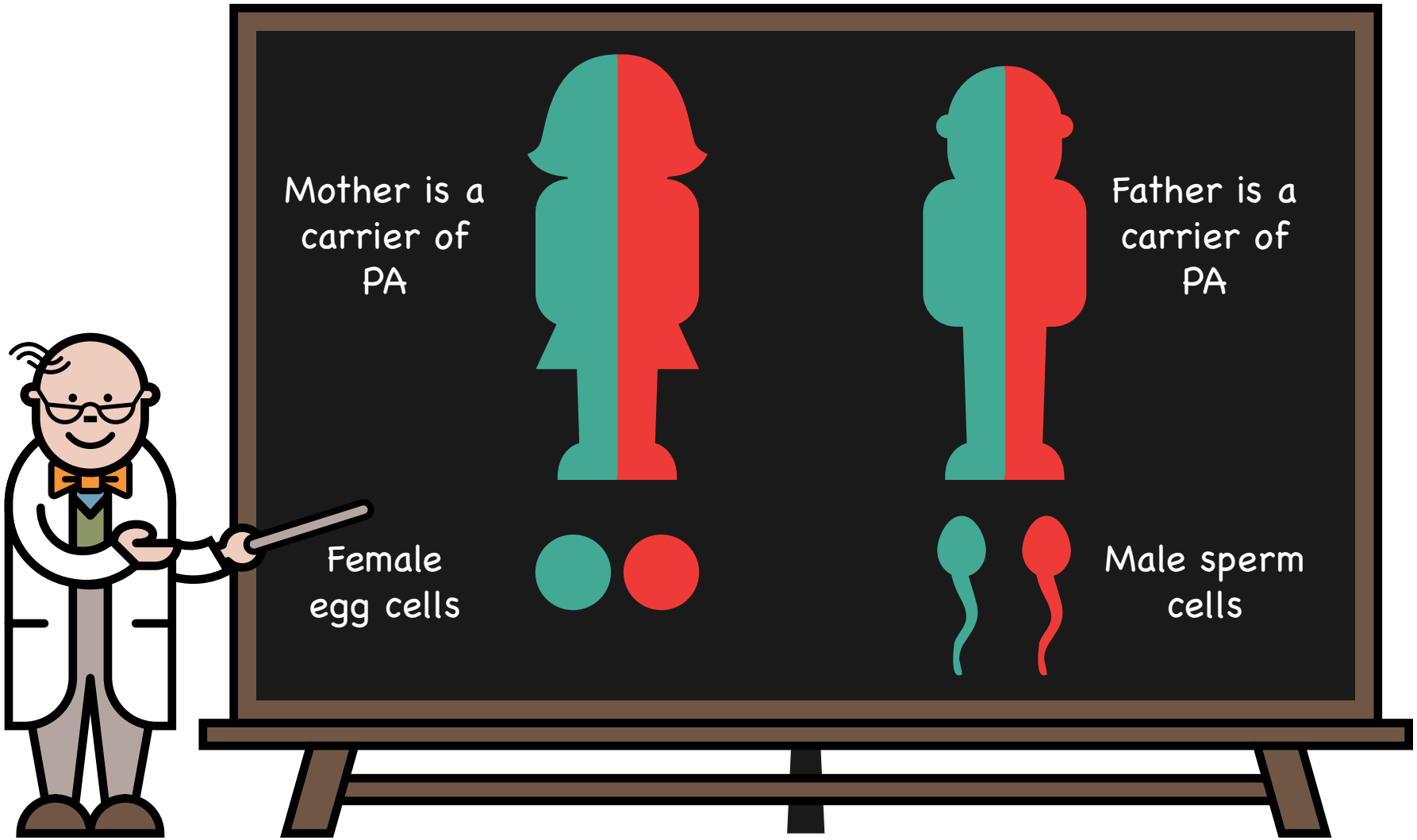
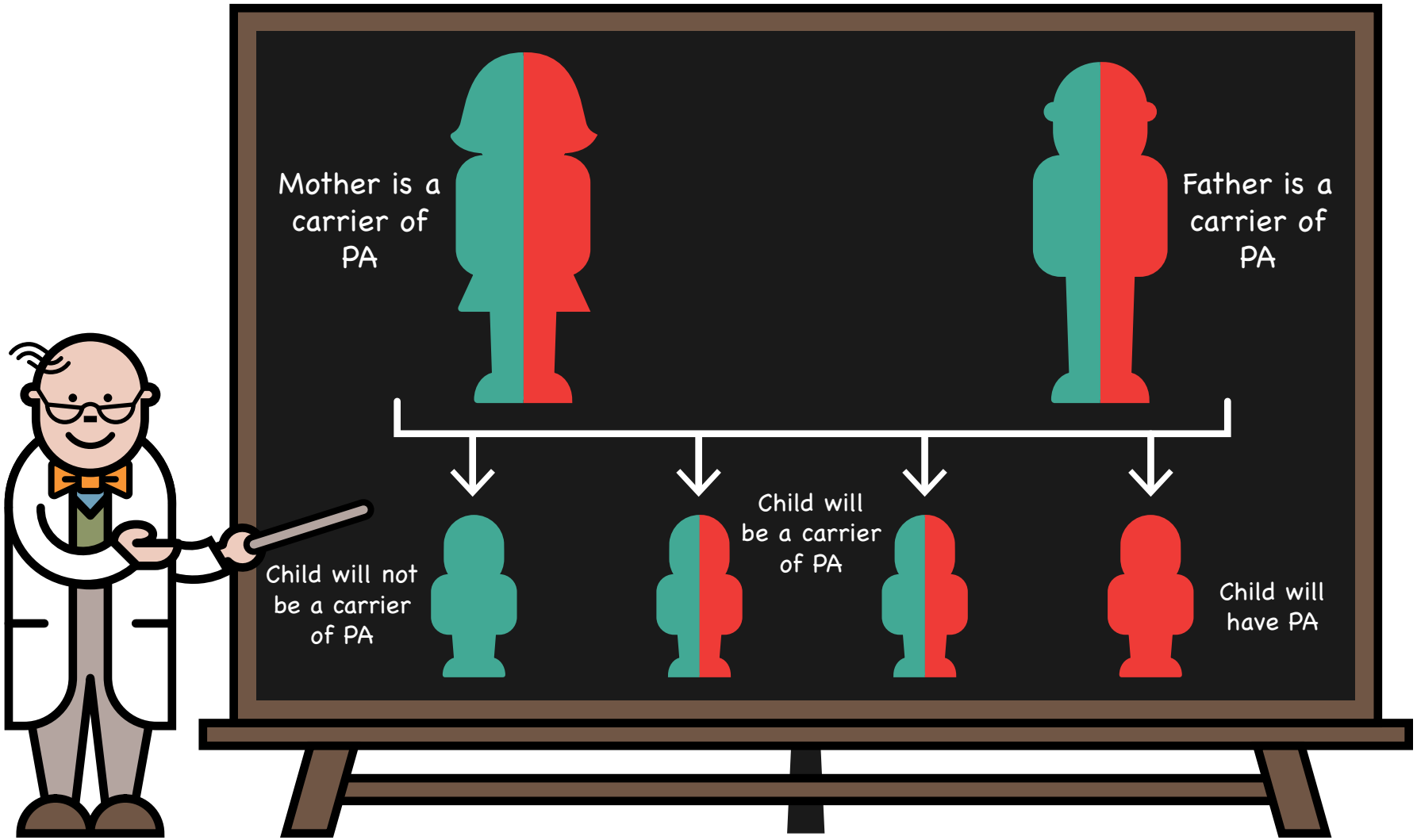


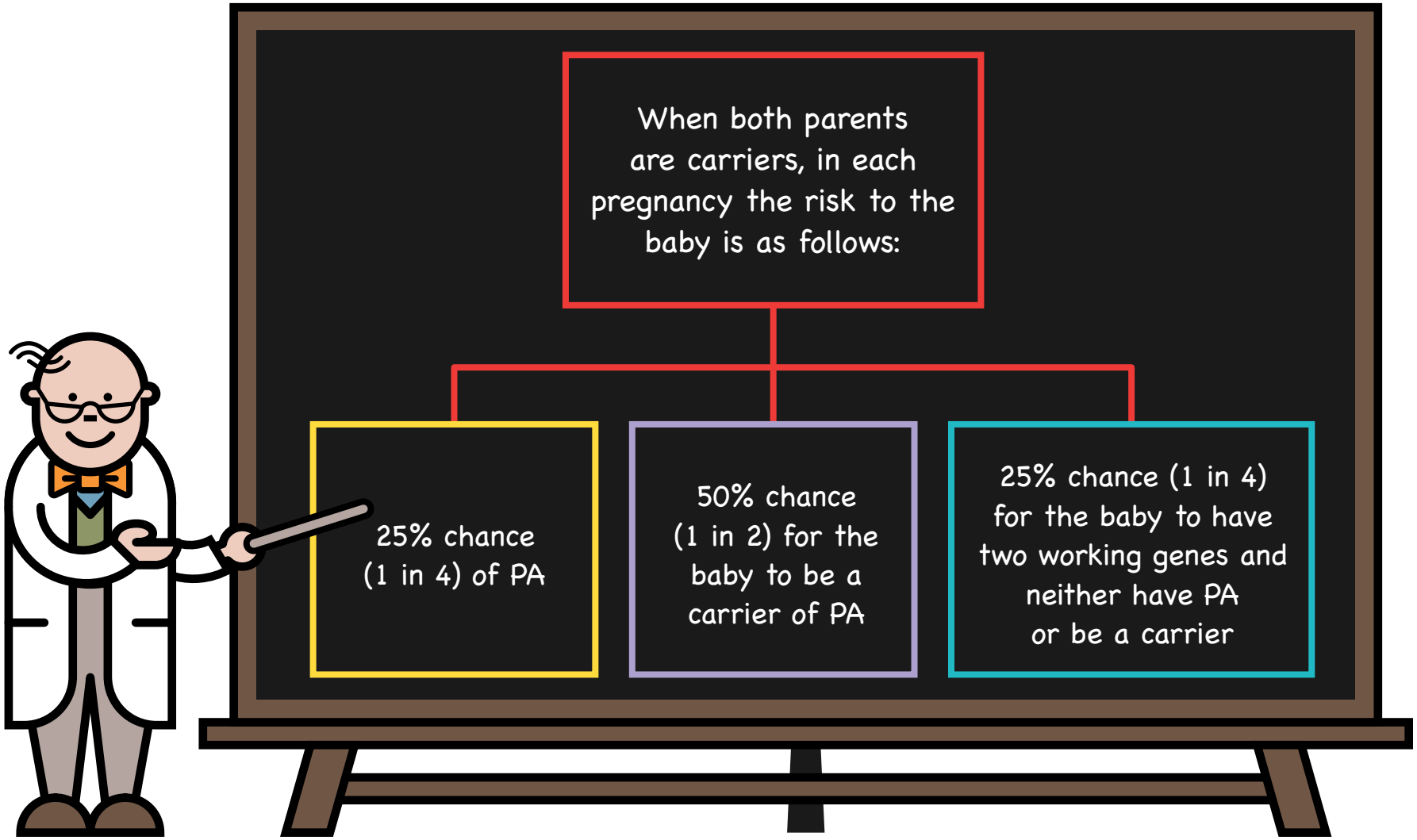
Inheritance – Autosomal recessive (carriers of PA)



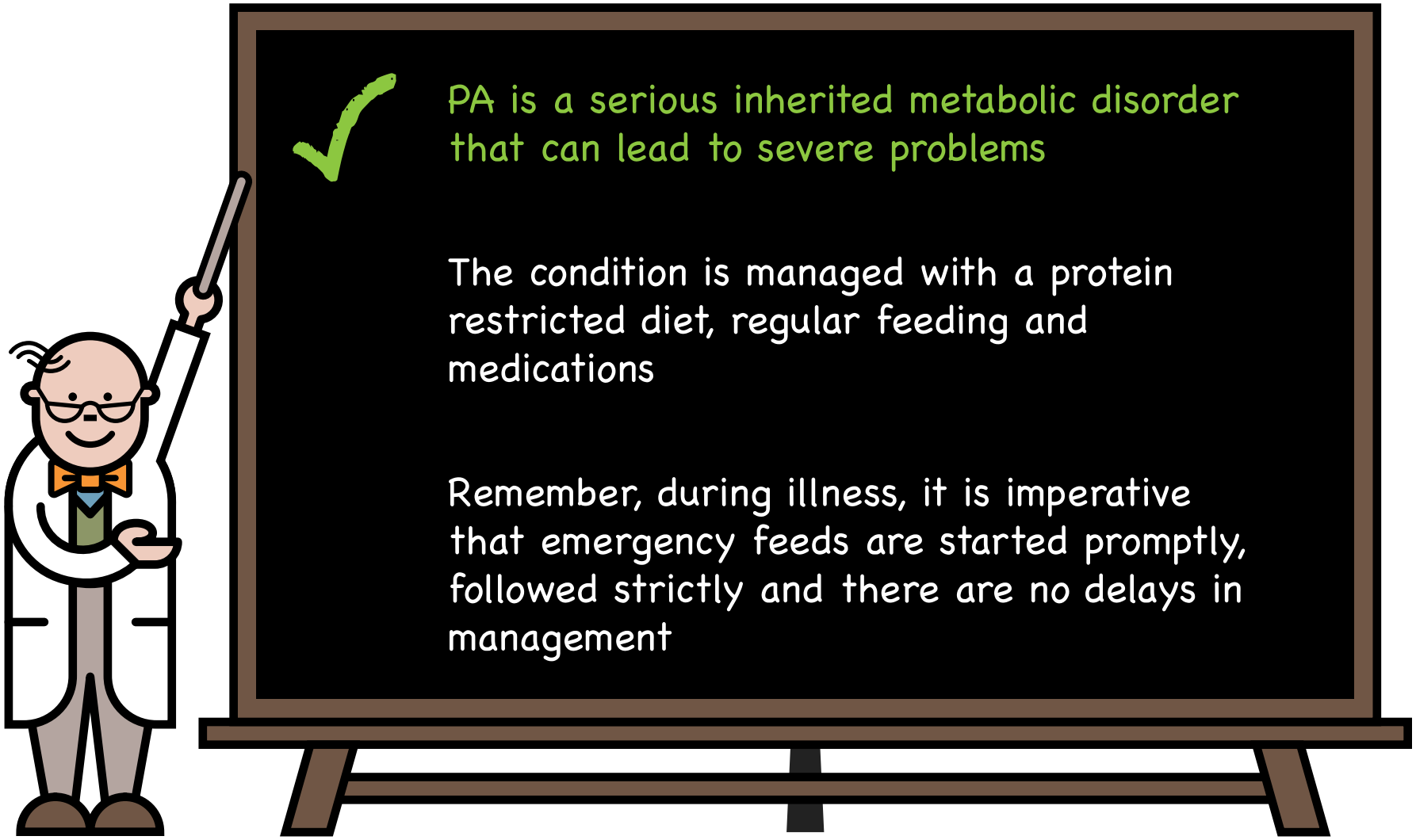
Inheritance – Autosomal recessive – possible combinations



Future pregnancies



Take home messages



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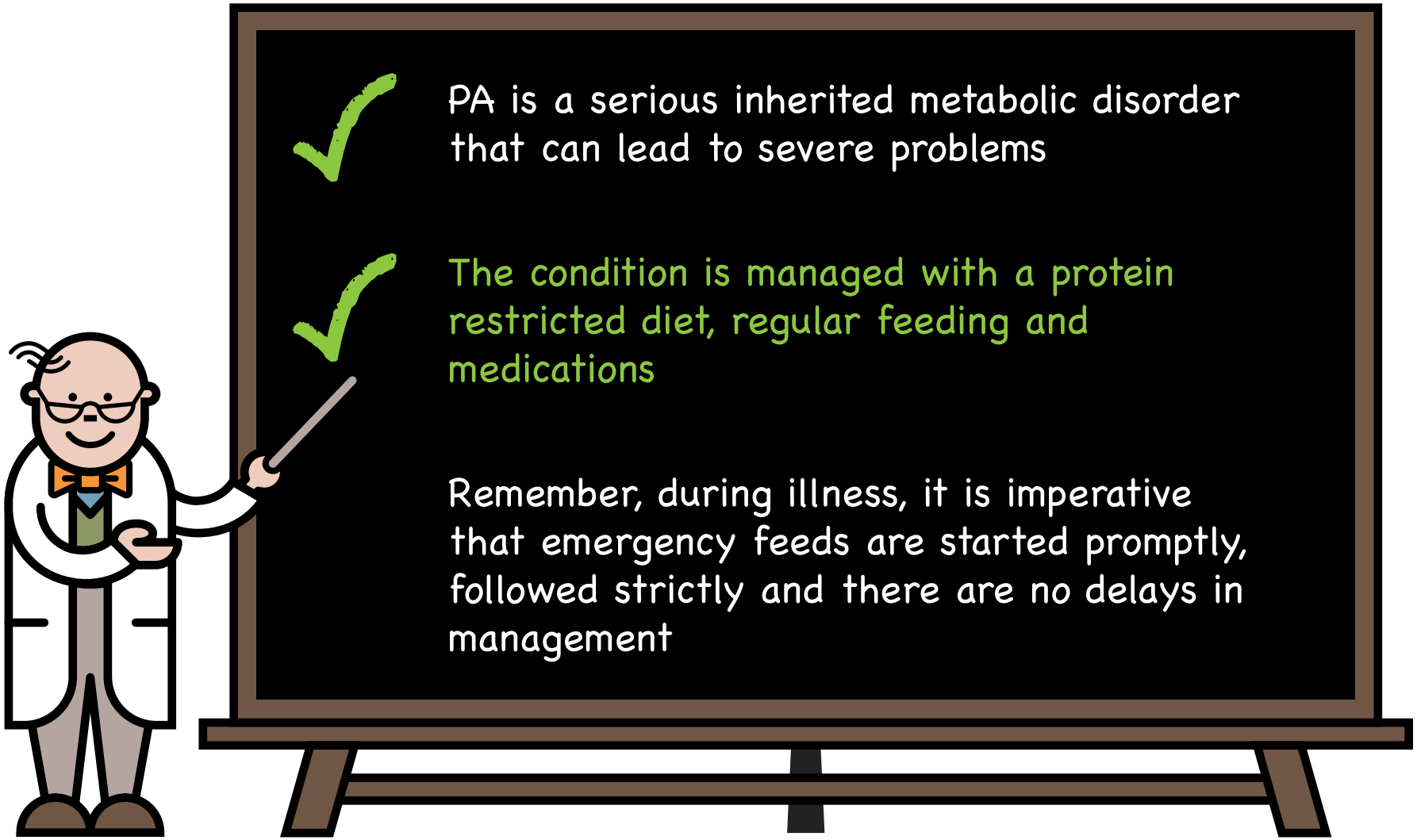

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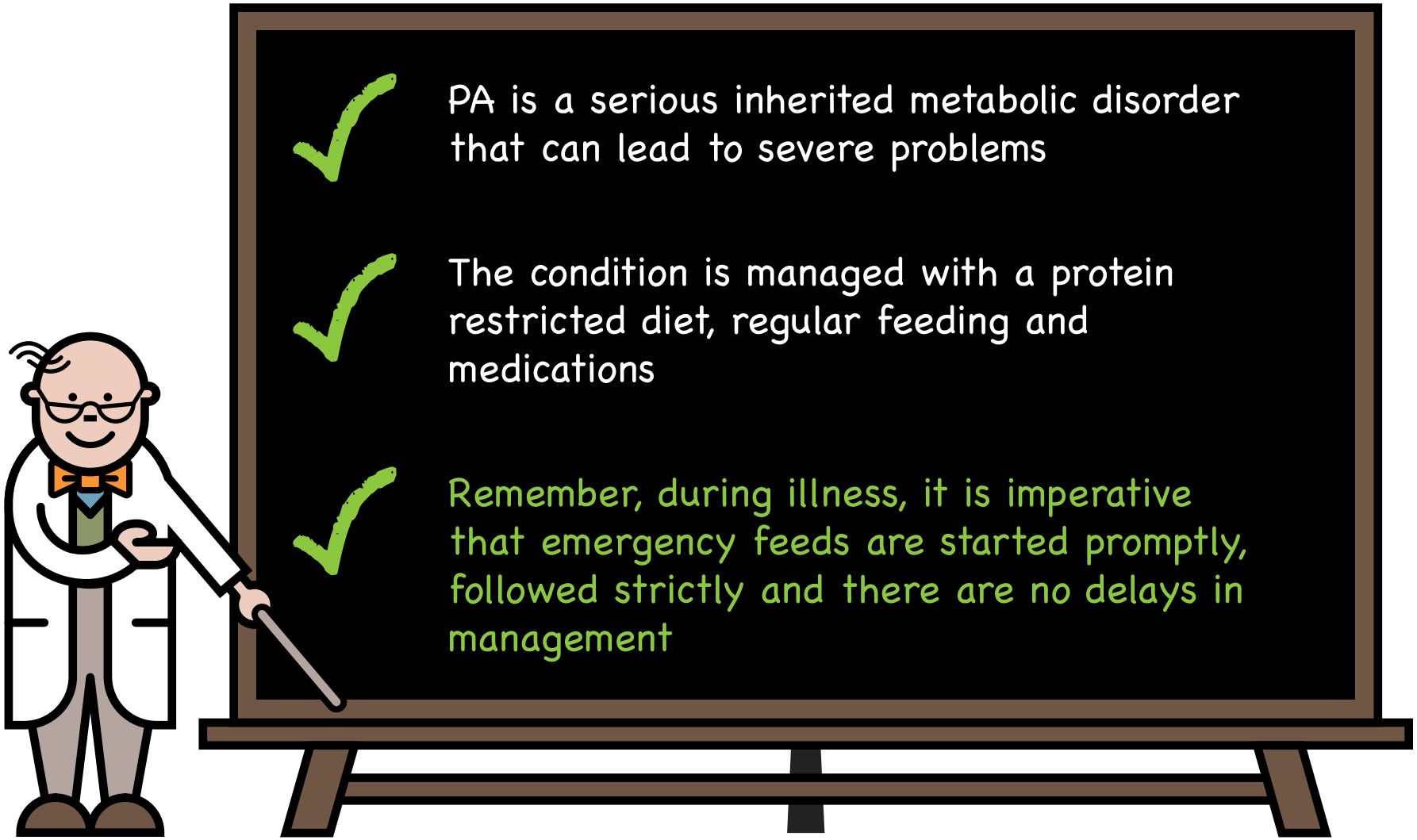
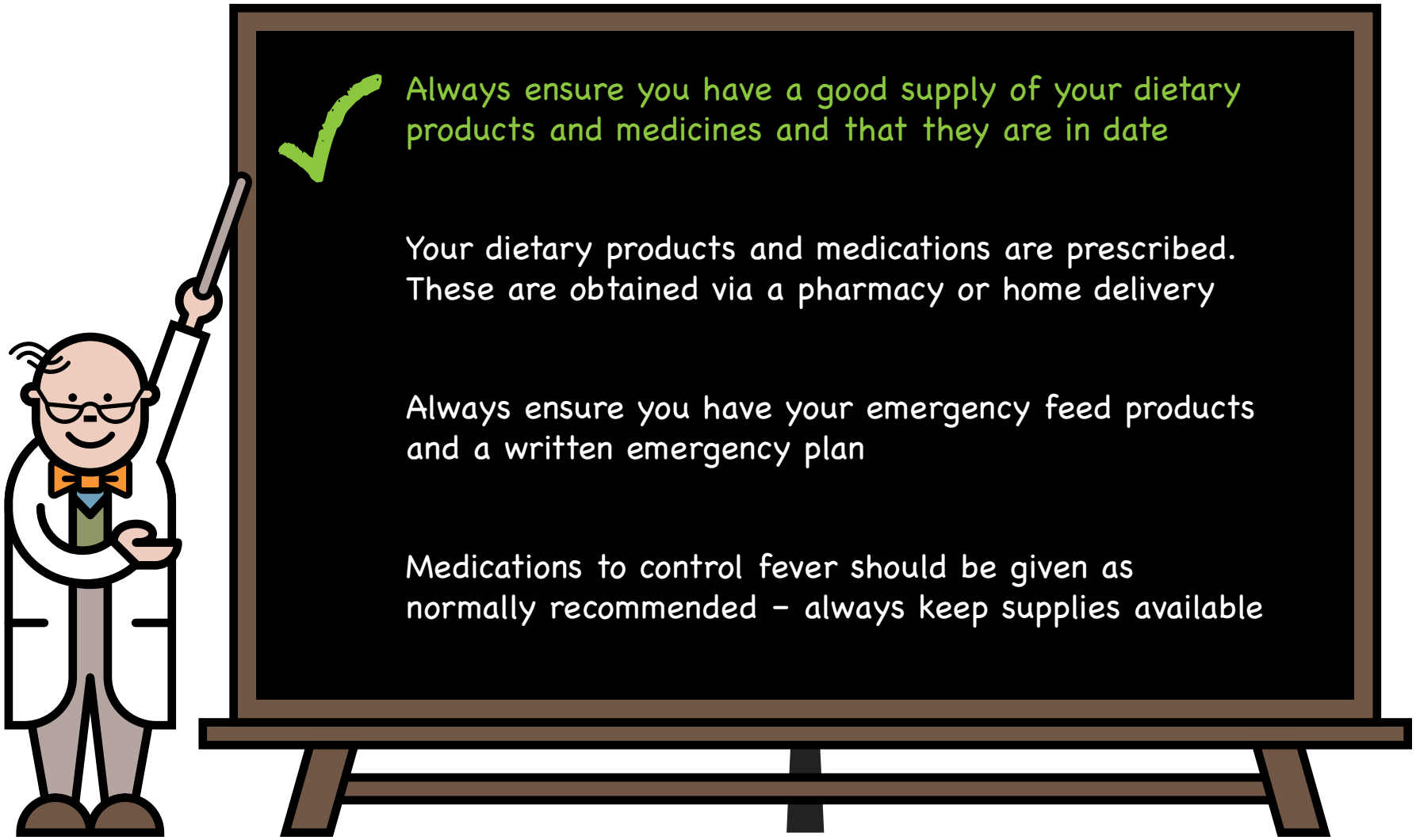


Helpful hints



Helpful hints

Helpful hints

Helpful hints

Who's who

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

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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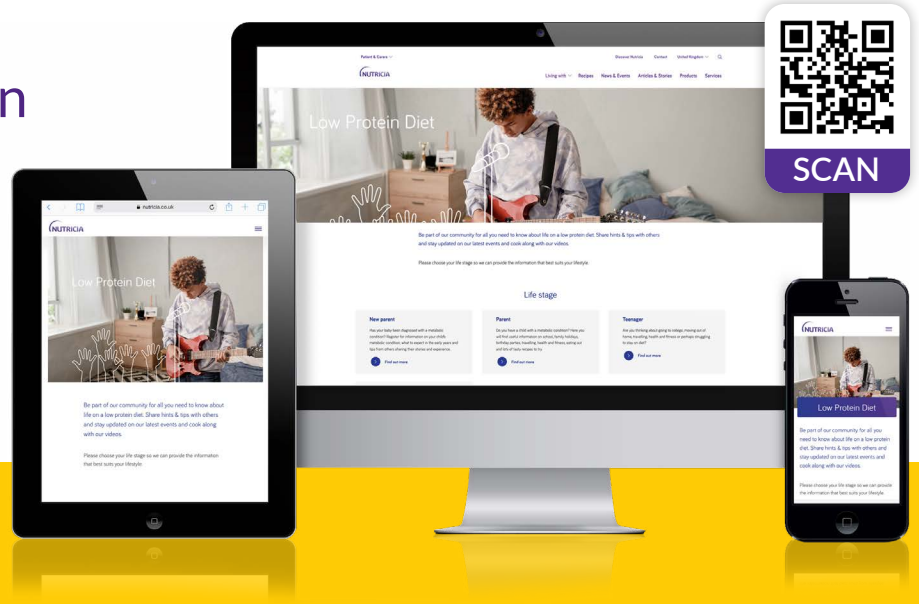

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