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 3, OCTOBER 2020

GA1

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.

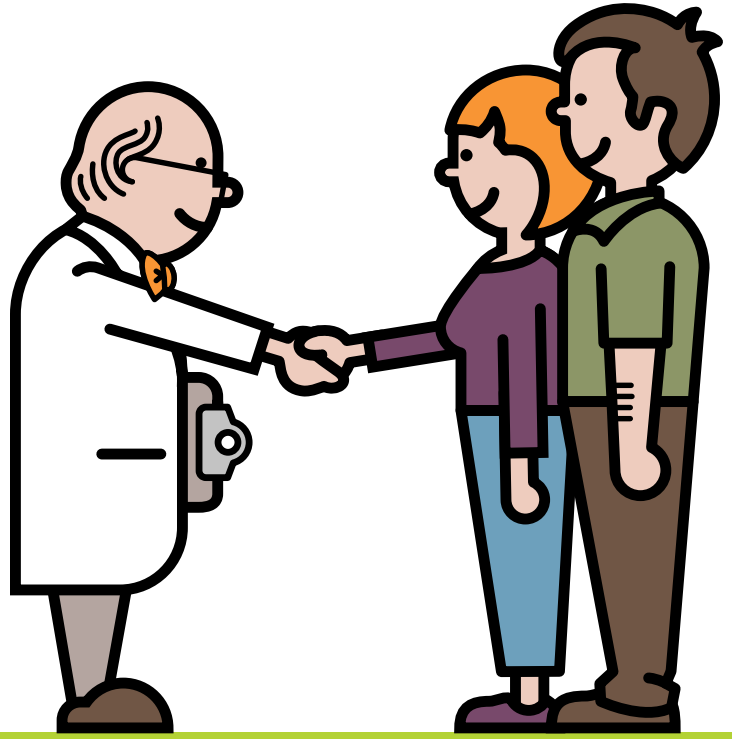
Each module produced is reviewed by a consultant clinician who is a member of the BIMDG.

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

Supported by NUTRICIA as a service to metabolic medicine

GA1

Information for families following a positive newborn screening



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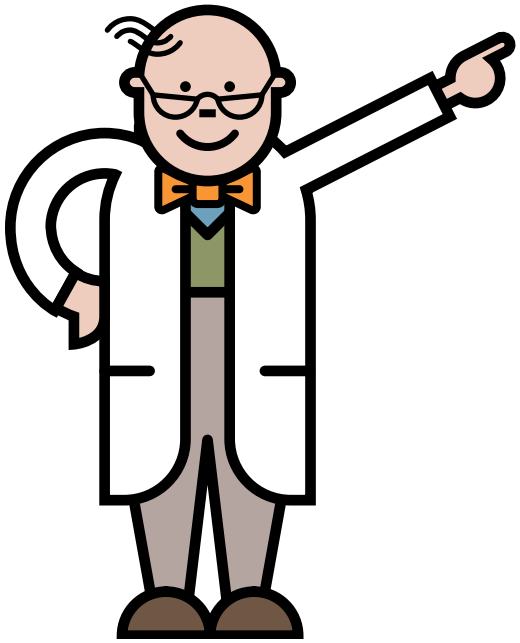
Tools Enabling Metabolic Parents LEarning

What is GA1?

GA1 stands for Glutaric Aciduria Type 1

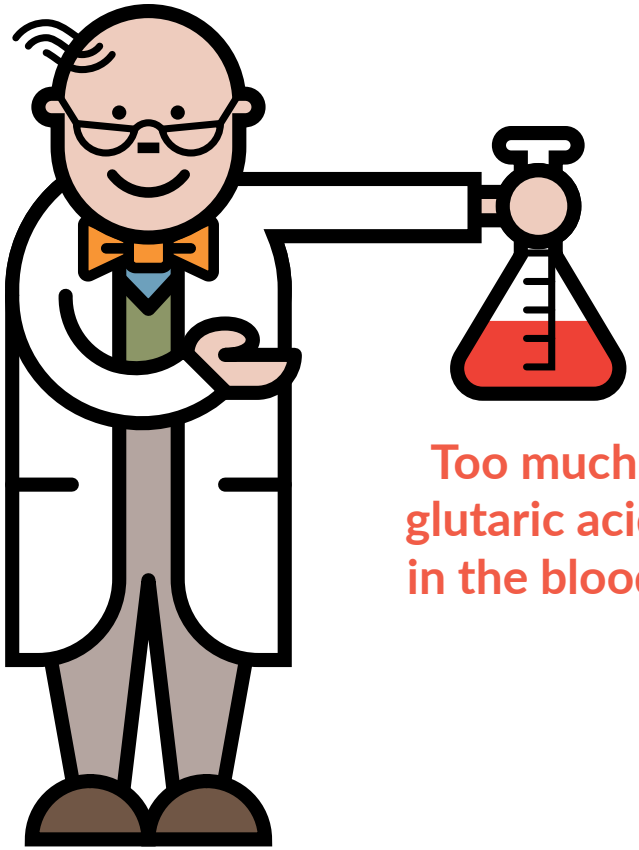
It is an inherited metabolic condition



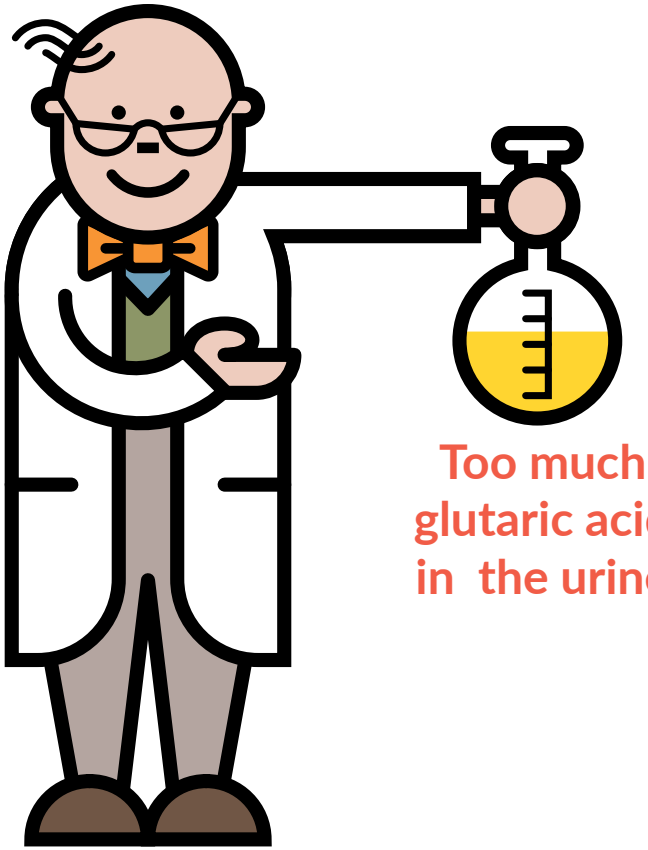
Glutaric Aciduria Type 1

GA1

What is GA1?



Too much glutaric acid in the blood



Too much glutaric acid in the urine

GA1 and protein

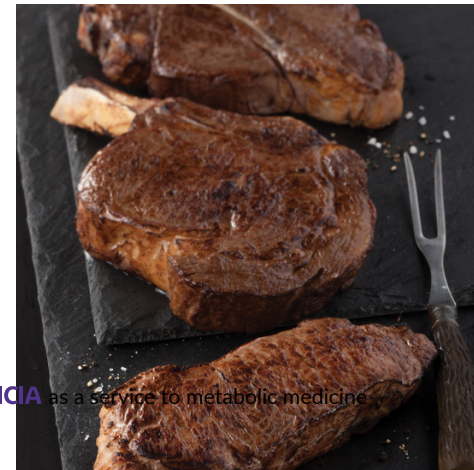
GA1 affects the way your baby breaks down protein

Many foods contain protein

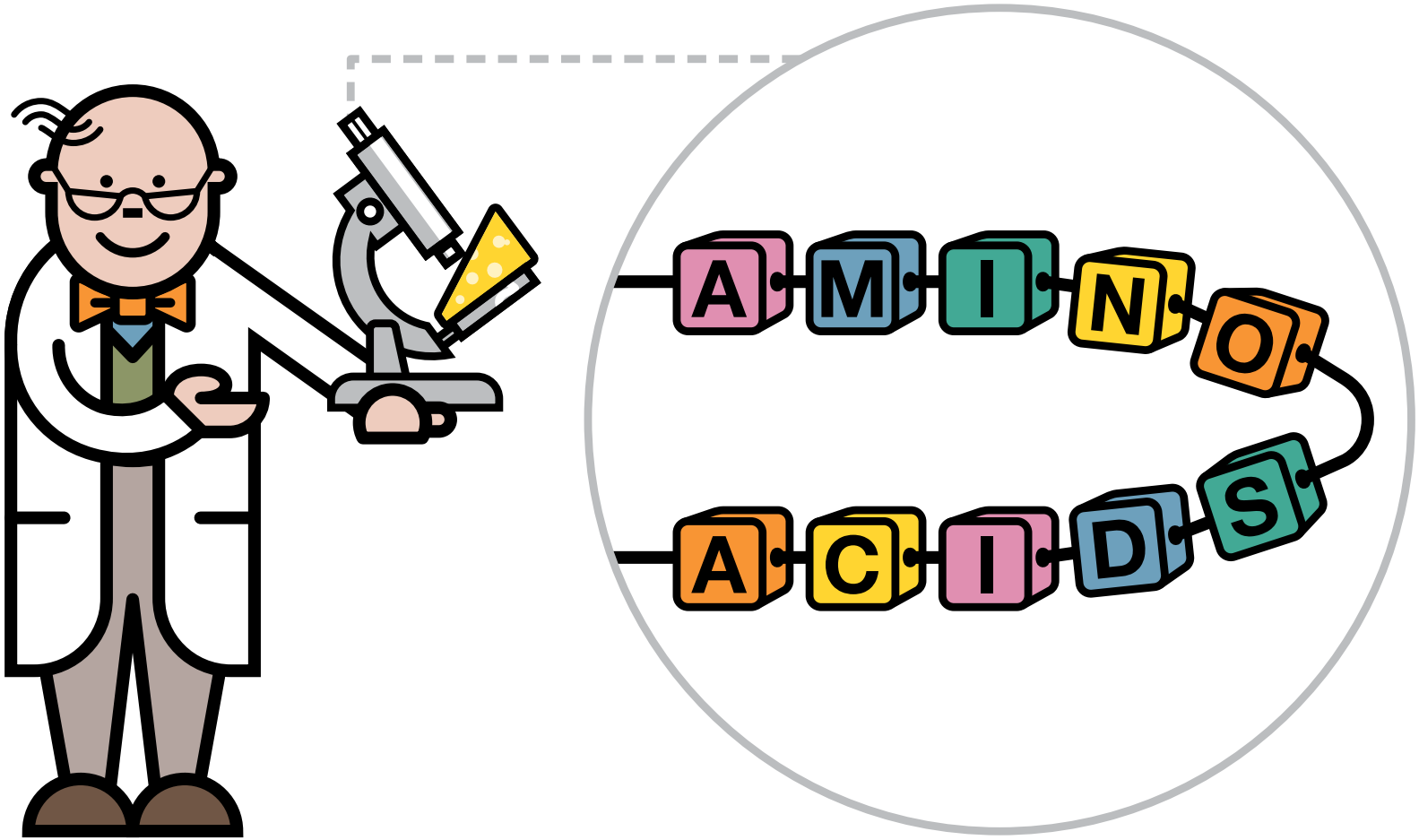
The body needs protein for growth and repair





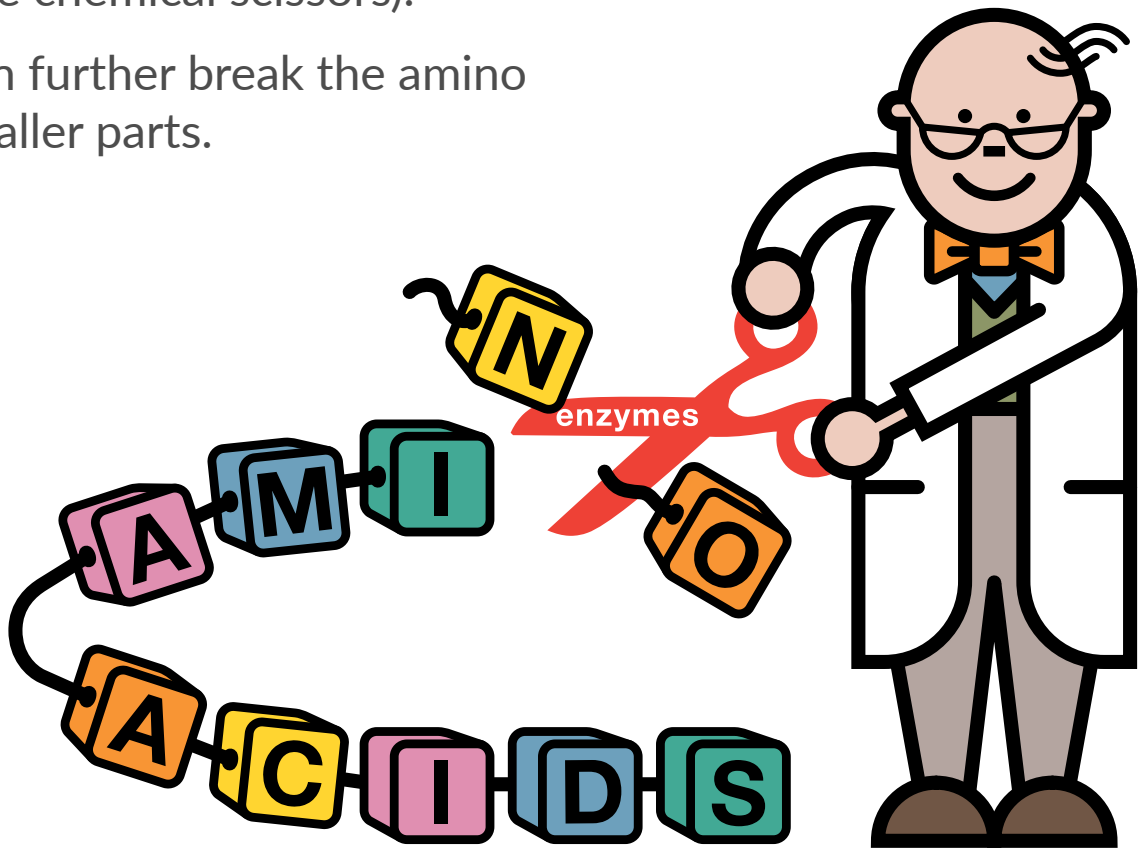
What is protein?



Protein and enzymes

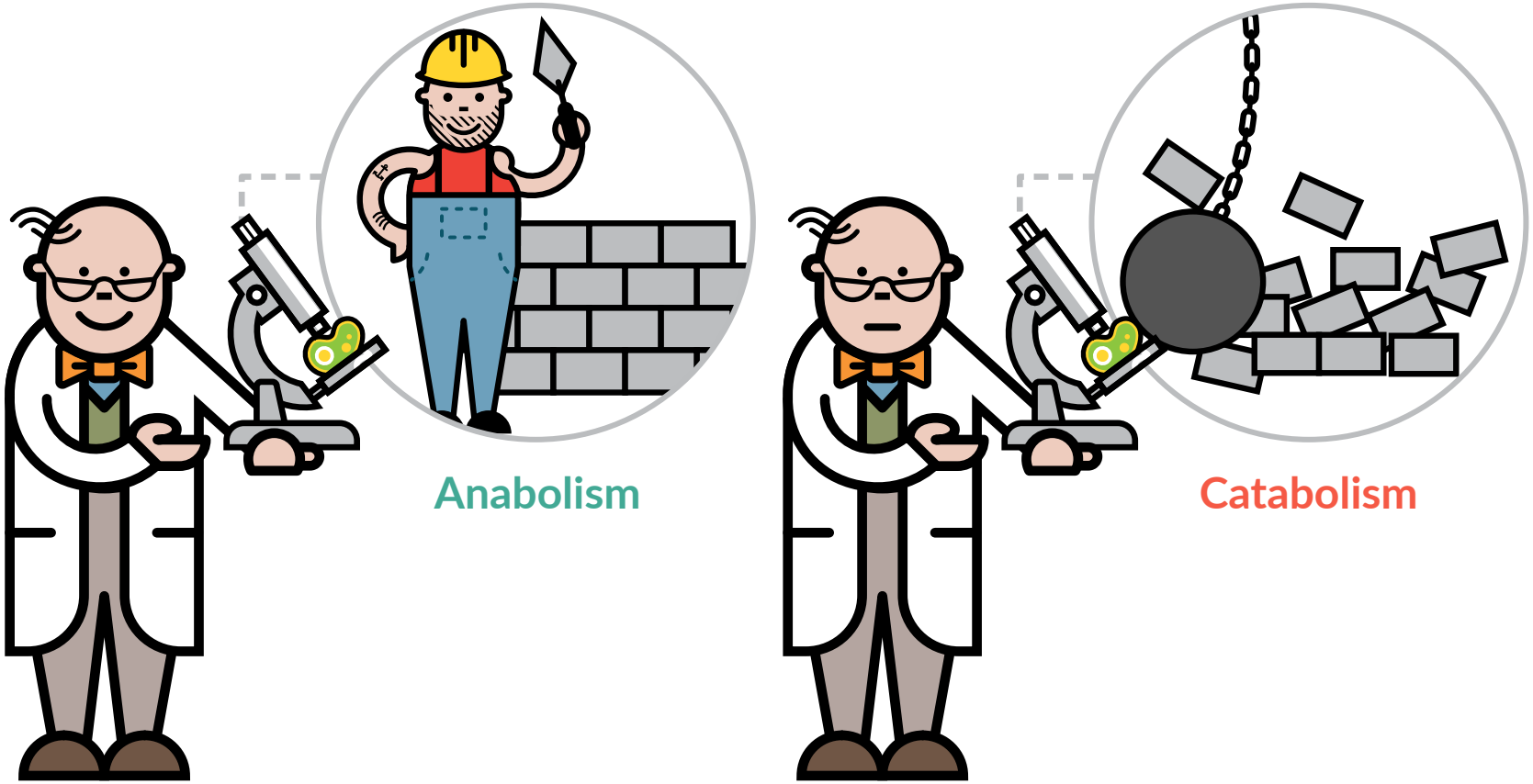
Protein is broken down into amino acids (building blocks of protein) by enzymes (which are like chemical scissors).

Enzymes then further break the amino acids into smaller parts.



Protein metabolism

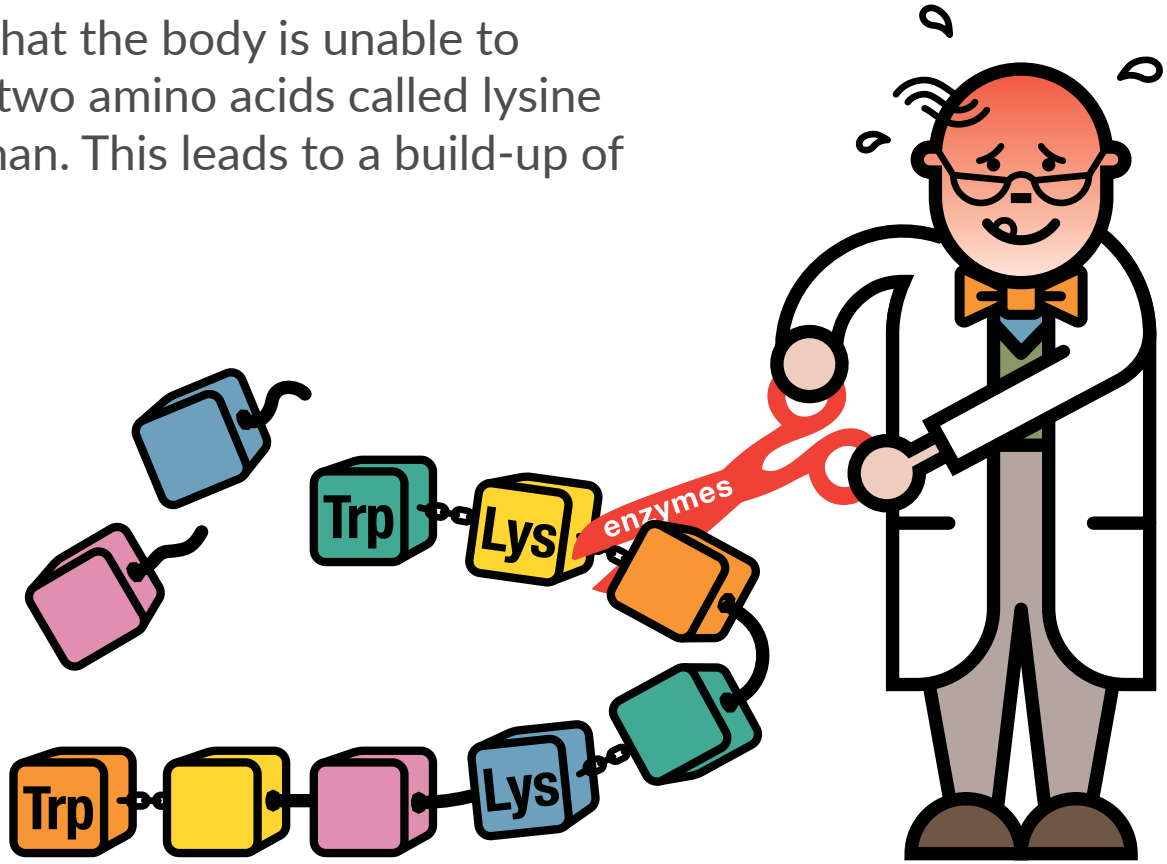
Metabolism refers to the chemical processes that occur inside the cells of the body.



What happens in GA1?

In GA1, the body lacks an enzyme called **glutaryl-CoA dehydrogenase.**

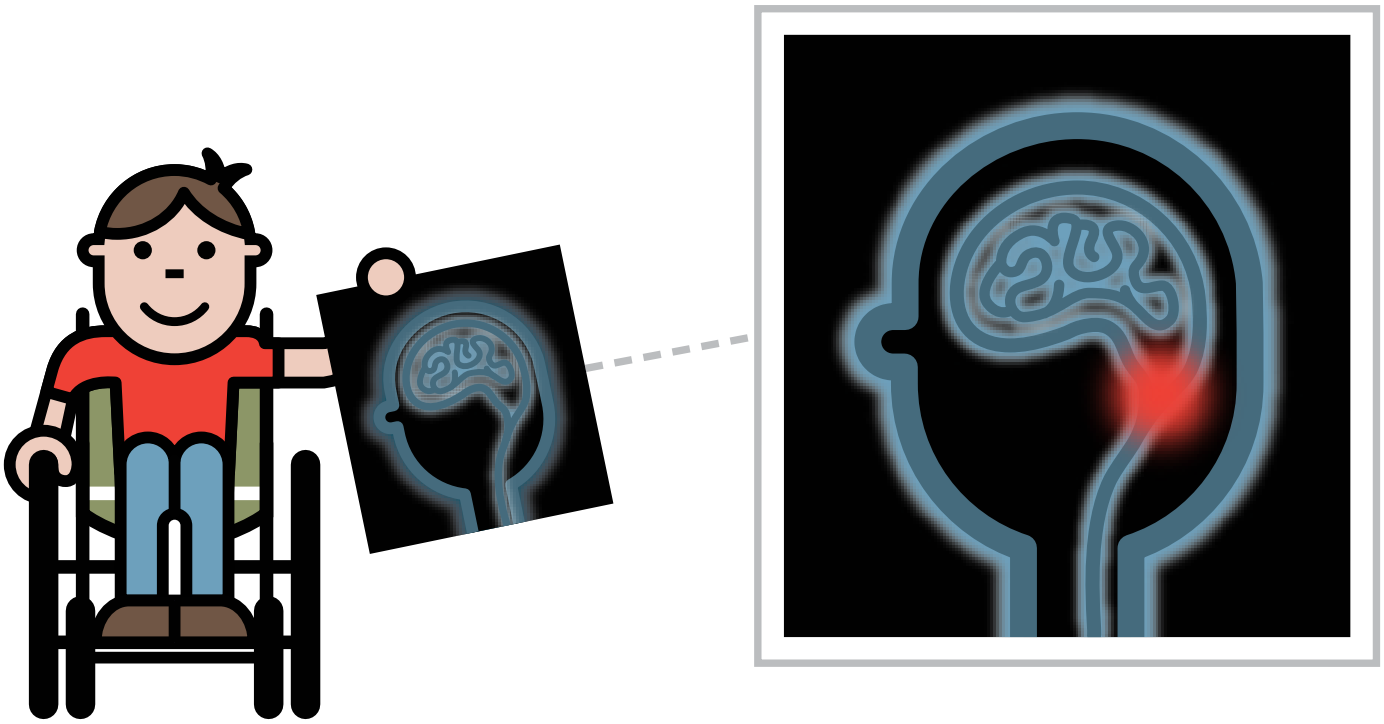
This means that the body is unable to break down two amino acids called lysine and tryptophan. This leads to a build-up of glutaric acid.



What can go wrong in GA1?

The basal ganglia in the brain controls movement.

The build up of glutaric acid damages the basal ganglia and causes movement problems.

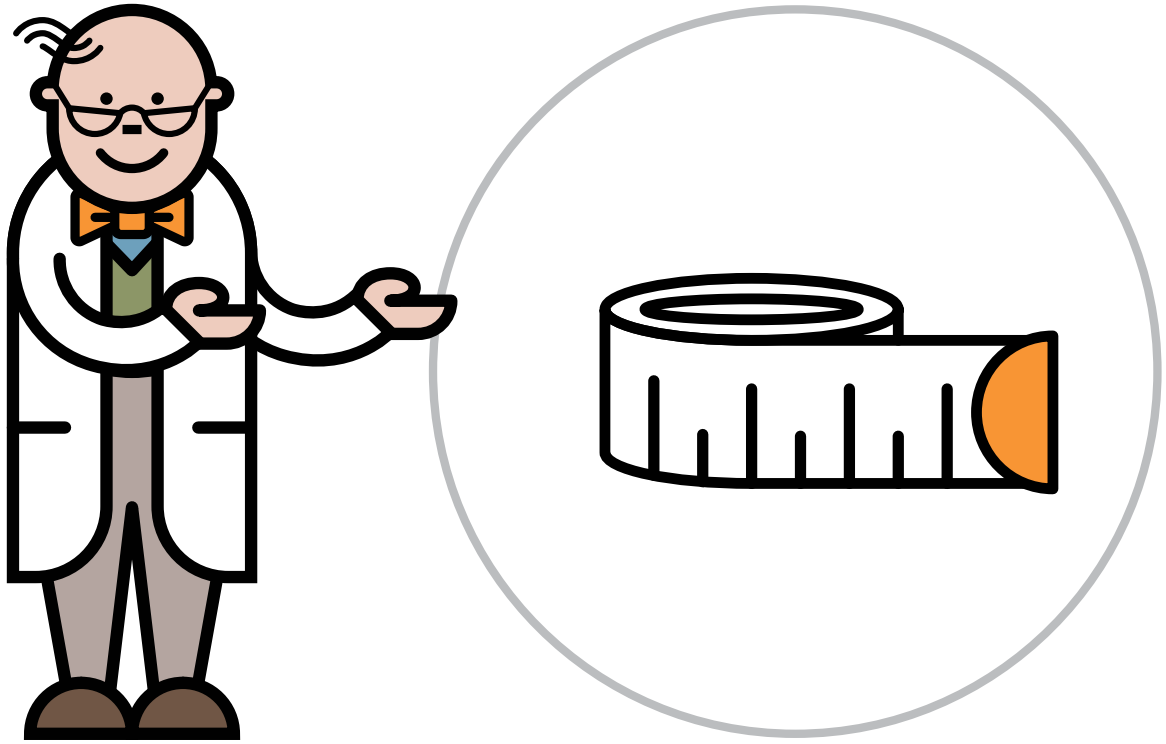


Metabolic crisis

- A **metabolic crisis** can trigger the **movement problems**. This is because there is a build-up of glutaric acid and other toxic chemicals
- It is usually caused by childhood infections or viruses causing high temperatures, vomiting and diarrhoea
- Avoidance of a metabolic crisis is essential

What about other symptoms in GA1?

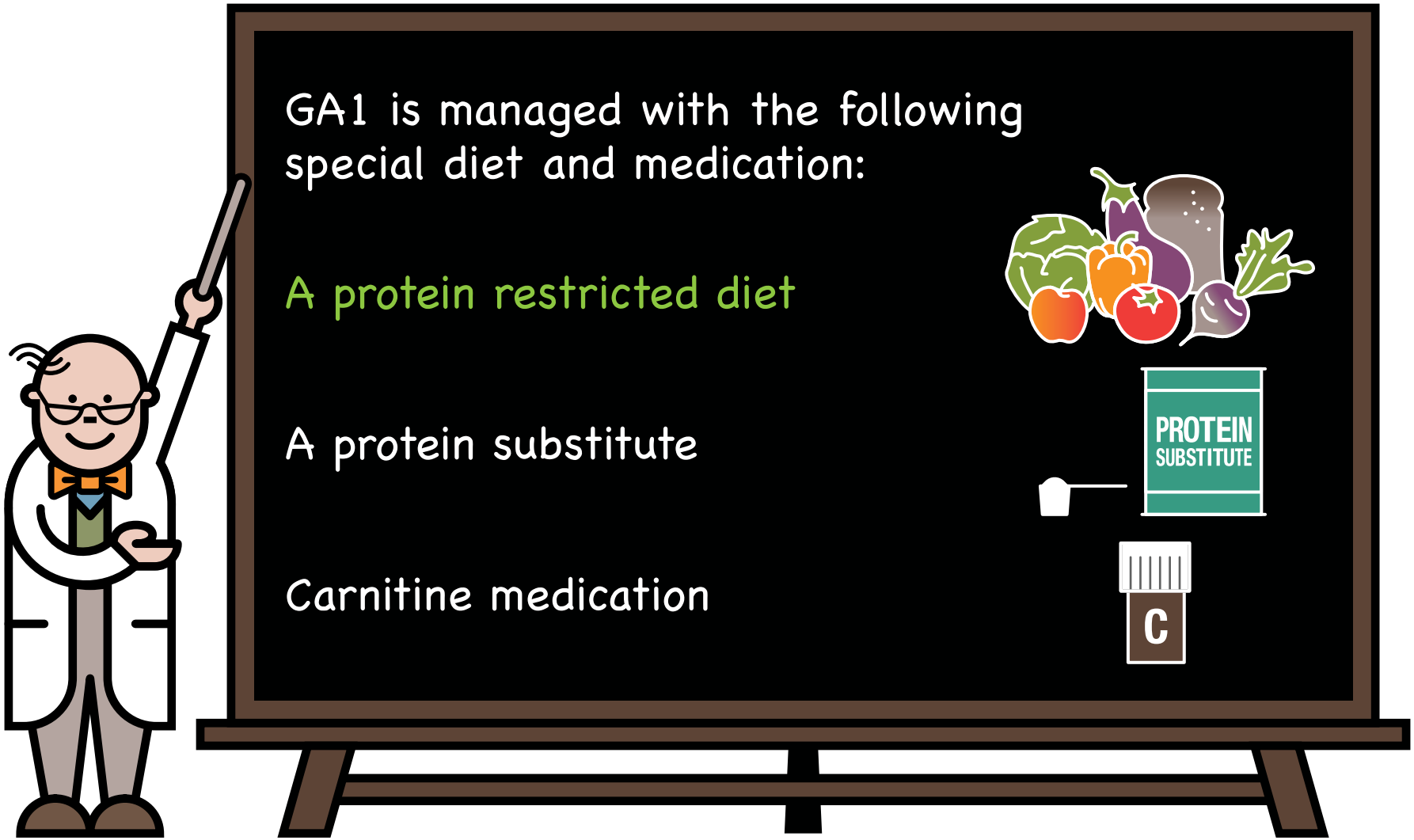
Babies with GA1 are usually healthy at birth although many are born with a larger than average head size.



How is GA1 diagnosed?

GA1 is diagnosed by newborn screening.
High levels of glutaric acid are found in the blood.

How is GA1 managed day to day?



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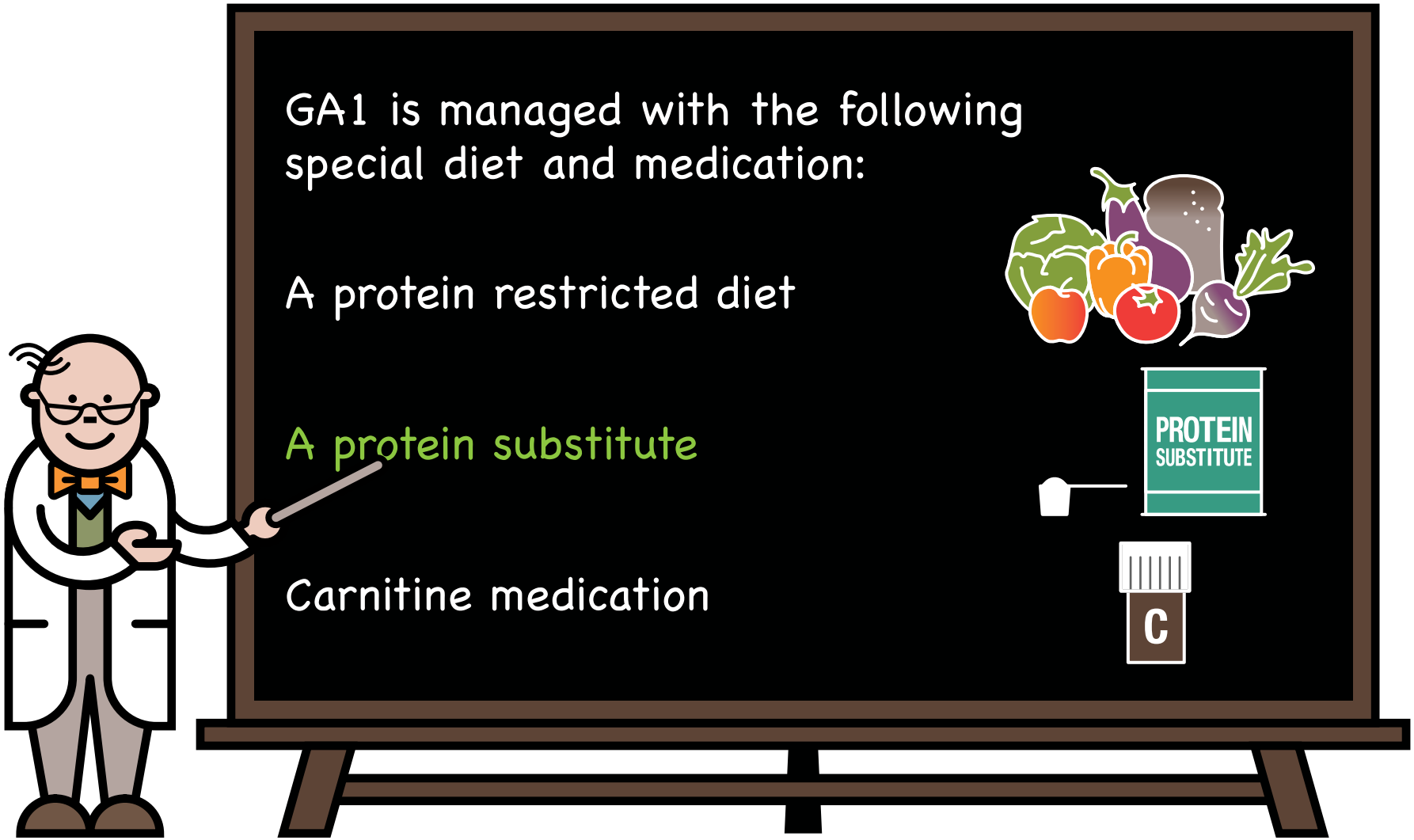

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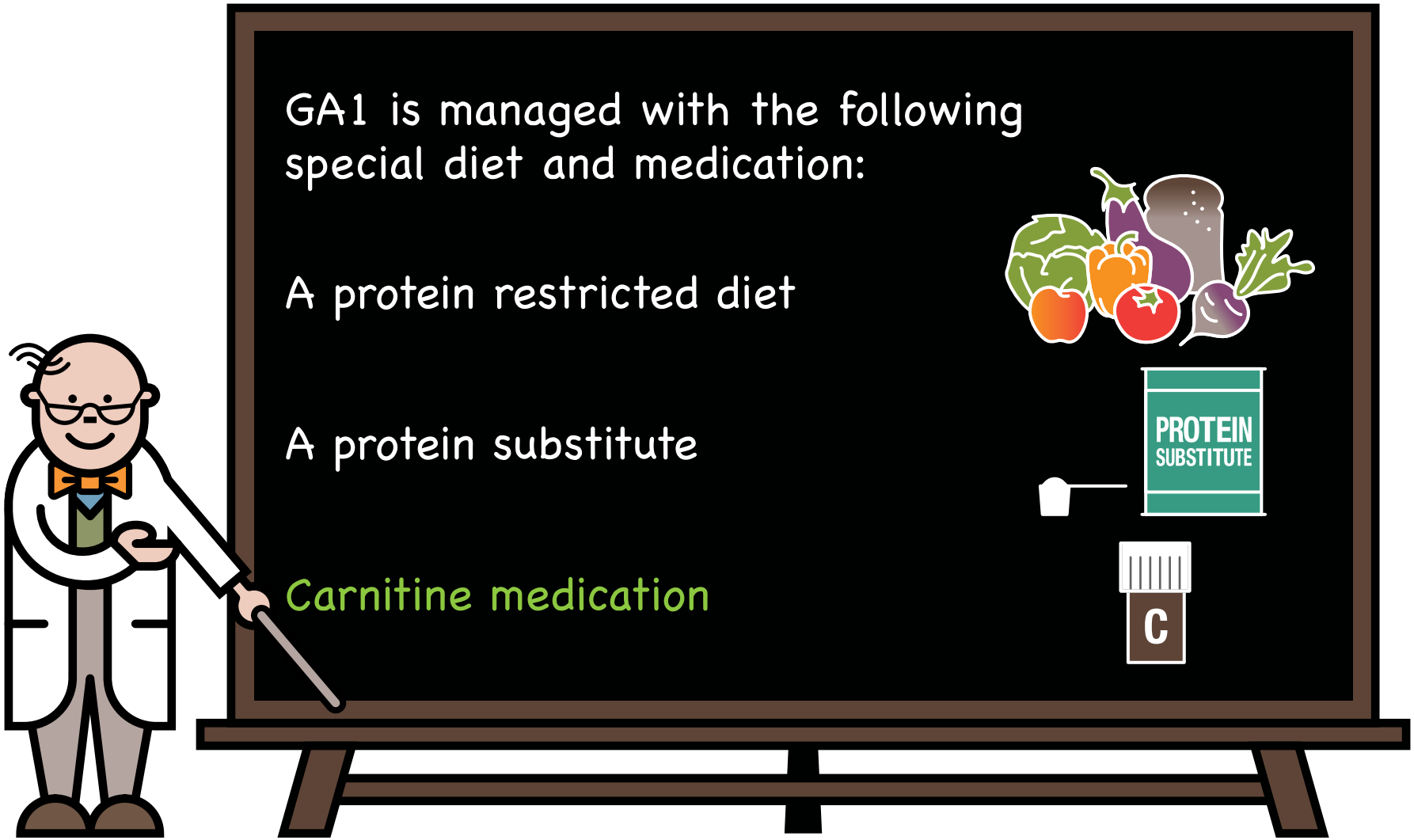
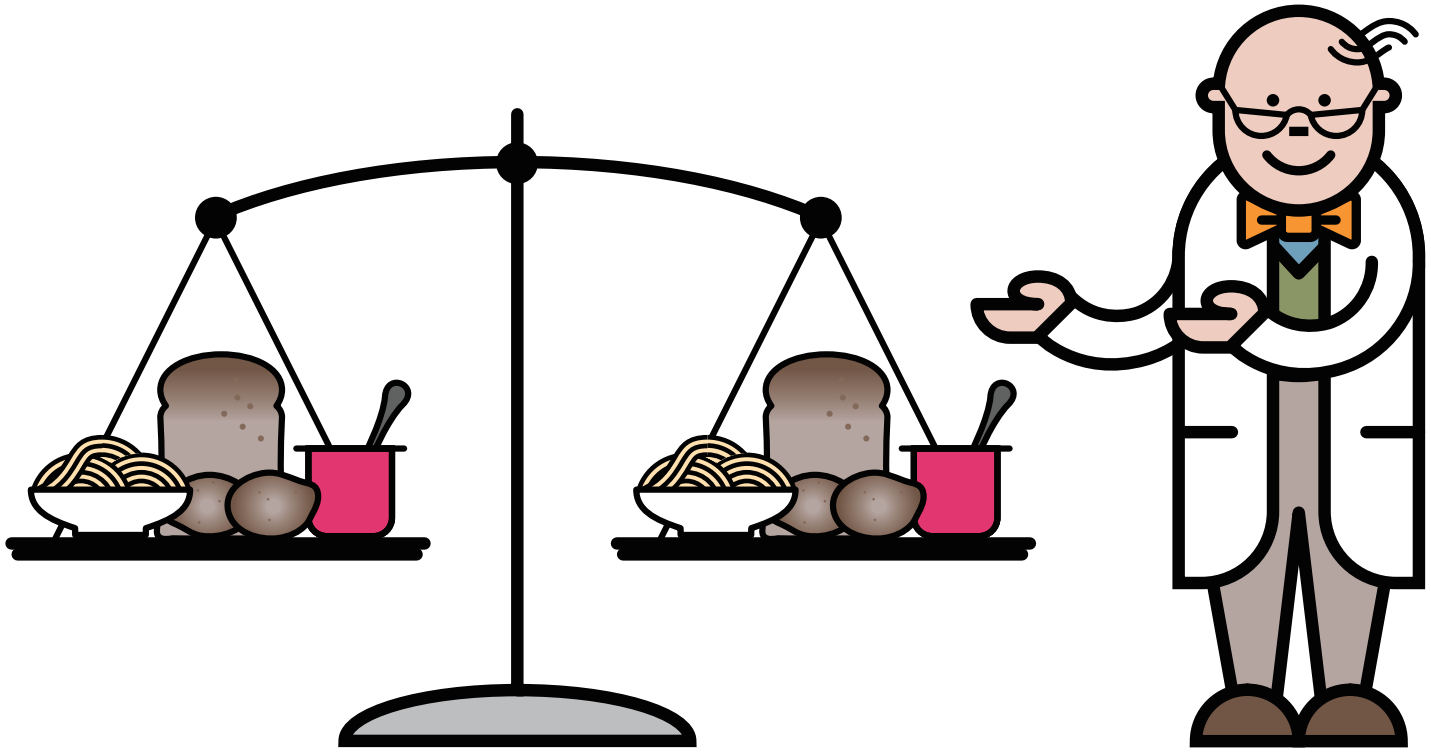


Protein balance is needed in GA1

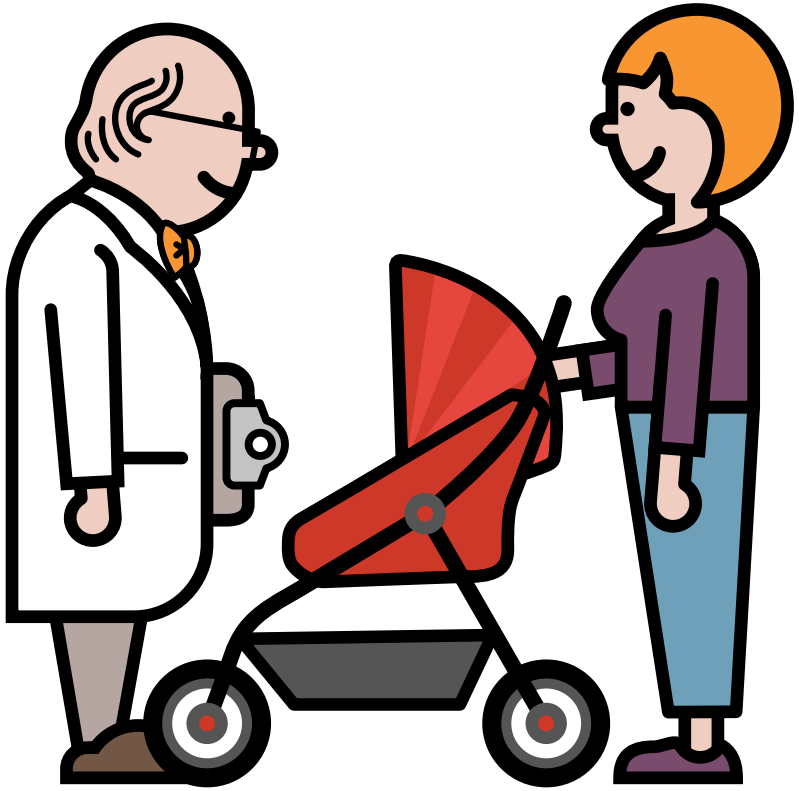
In GA1, it is important that enough protein is given for growth
... but not too much as toxic chemicals will be made.



Measured protein intake

In babies, a restricted amount of protein is given from breast milk or measured amounts of infant feed.

The amount given will be monitored regularly by your specialist metabolic dietitian.

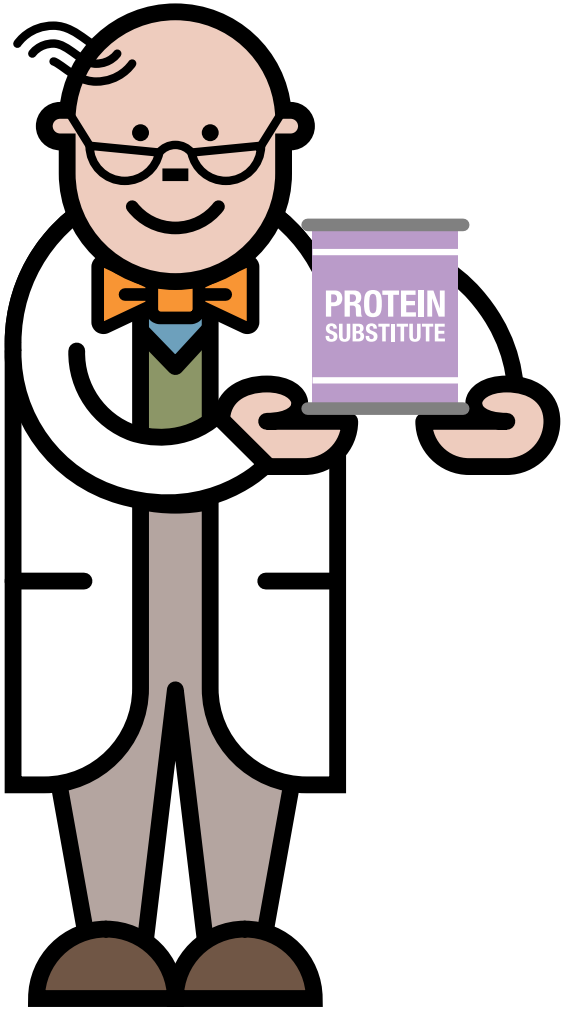


Protein substitute

Protein substitute is essential for metabolic control.

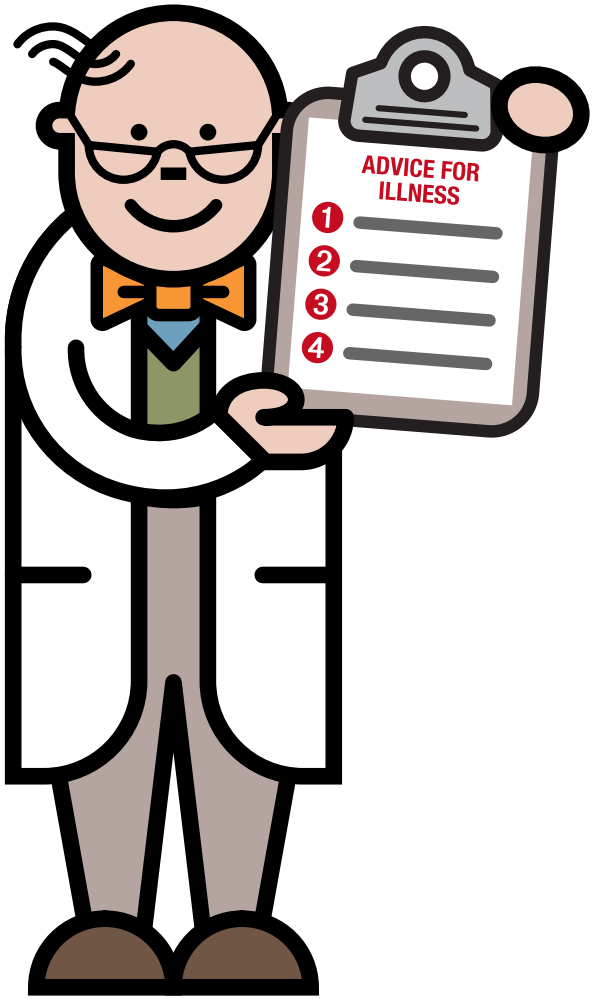
It will help to meet your baby's protein, energy, vitamin and mineral requirements.

It is available on prescription.



How is GA1 managed during illness?

- During any childhood illness, an emergency regimen is given
- Illnesses can cause catabolism or protein breakdown
- This will lead to a build up of glutaric acid and cause a metabolic crisis



How is GA1 managed during illness?

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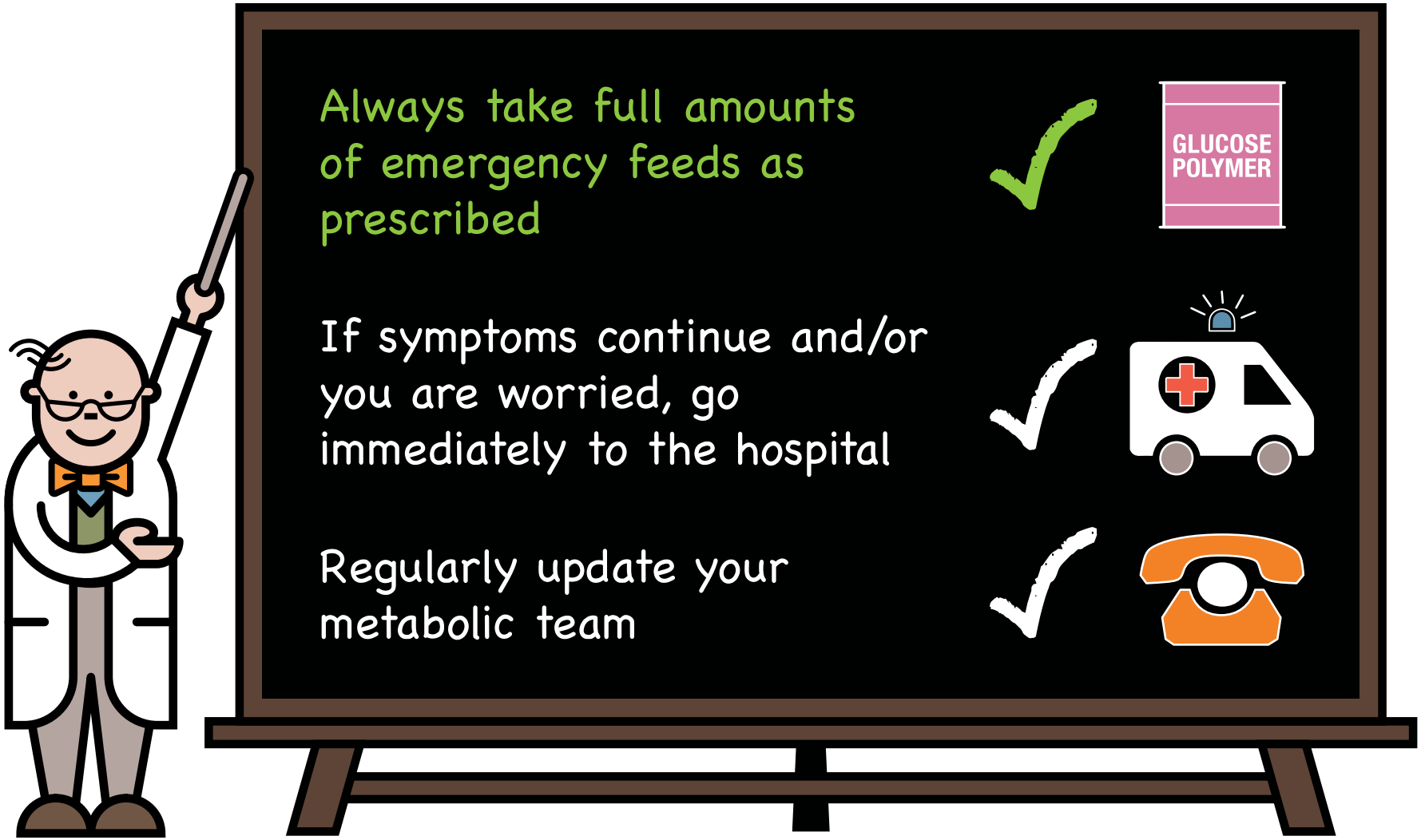
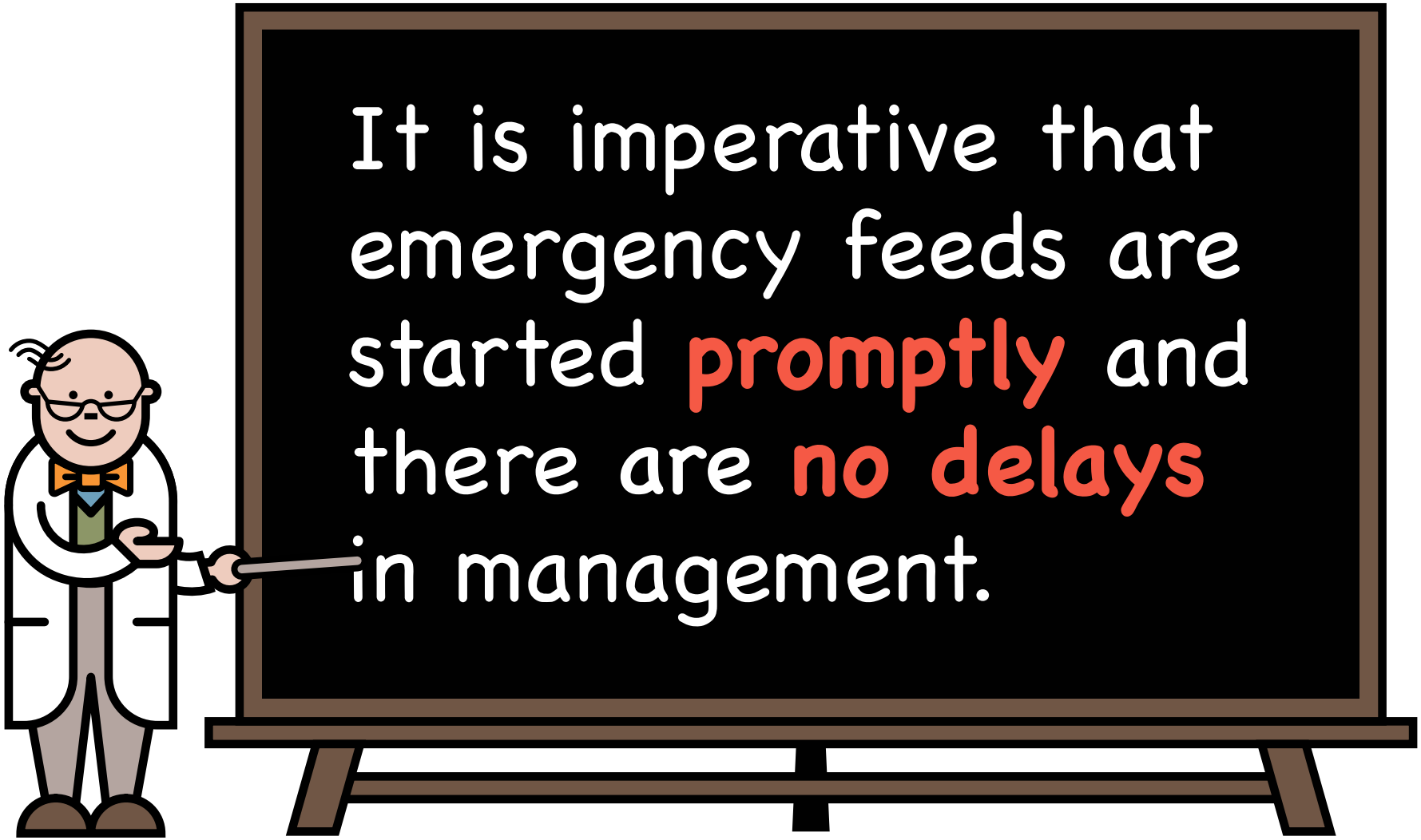
Checklist for illness



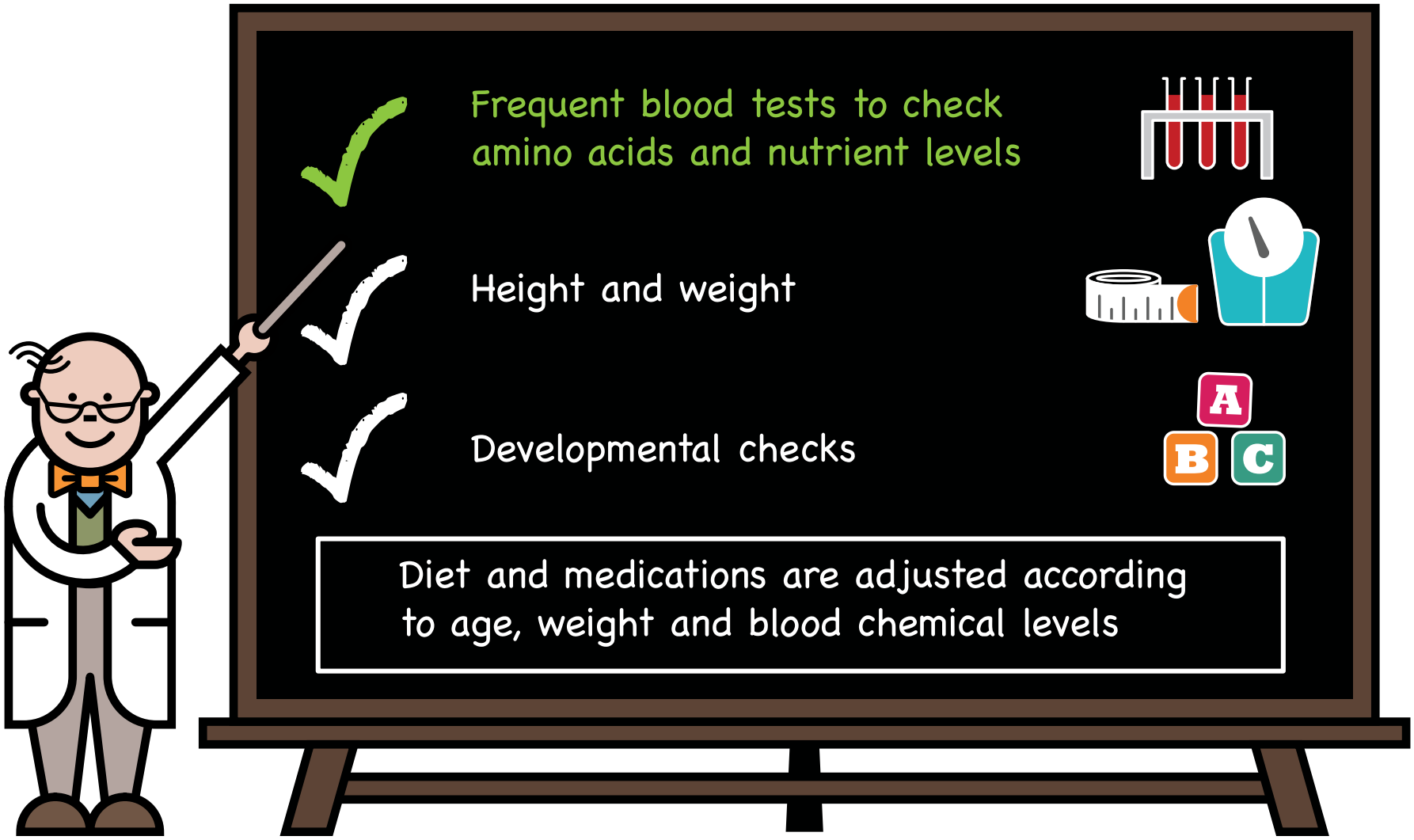
Checklist for illness

Checklist for illness

Key message



How is GA1 monitored?



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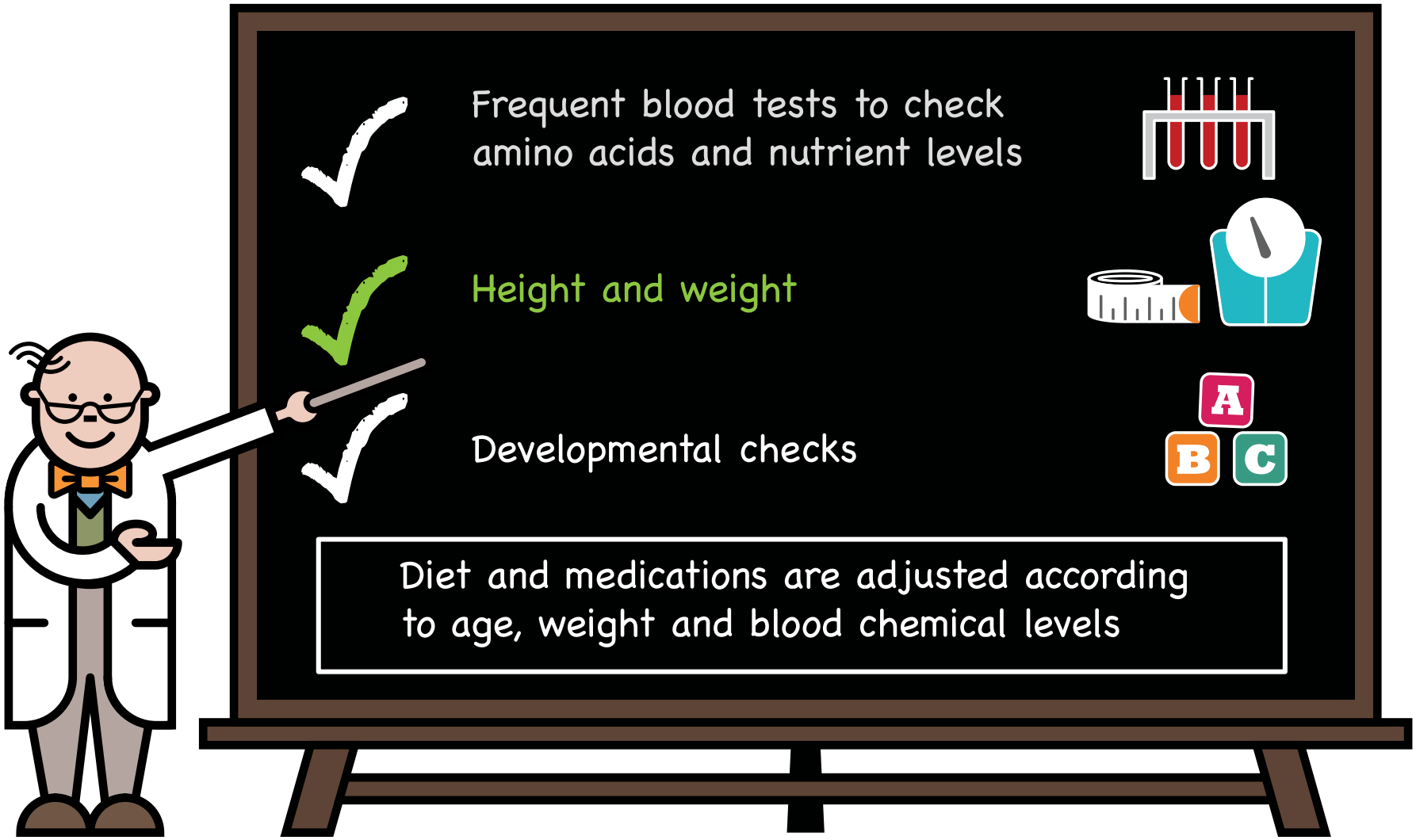

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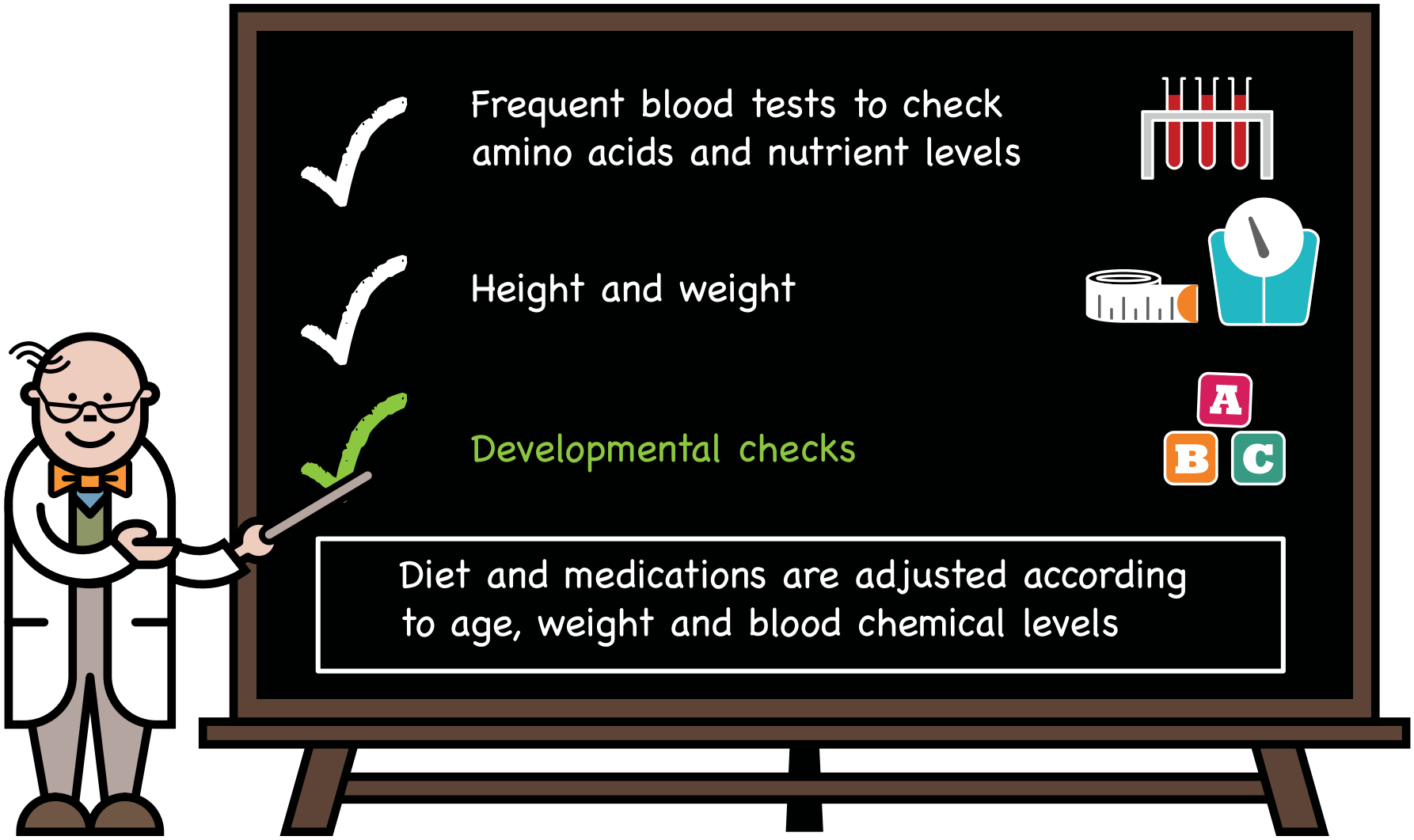

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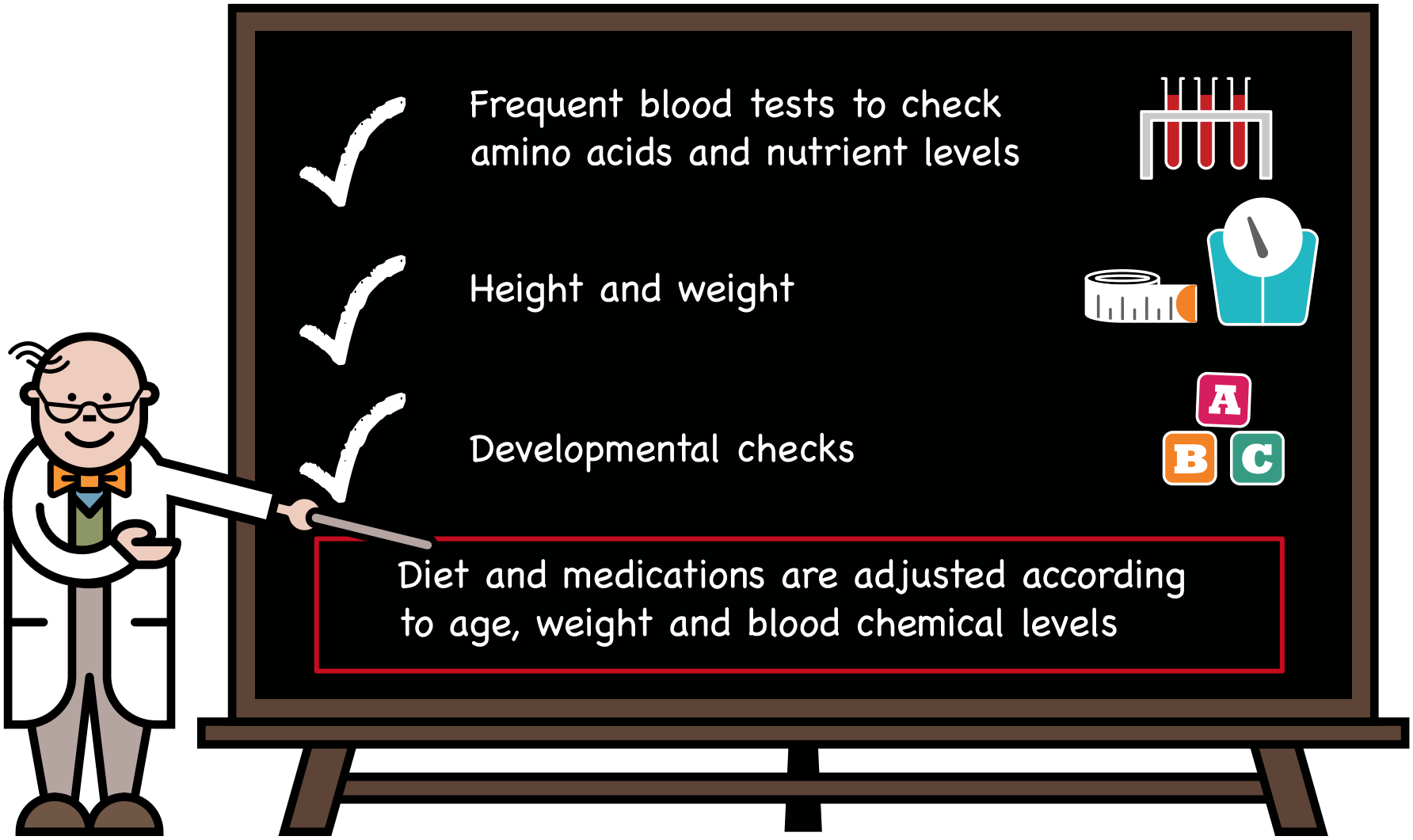
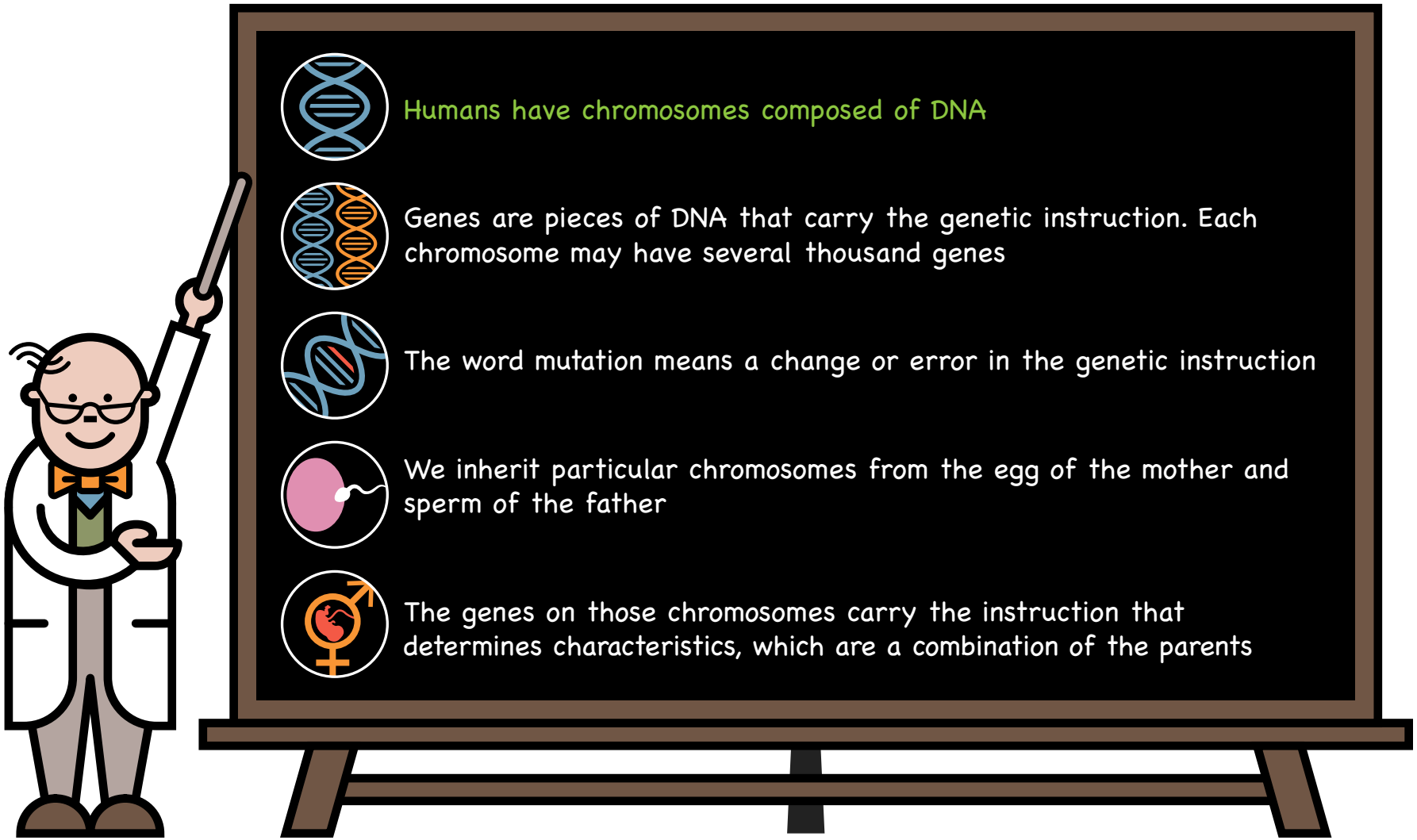
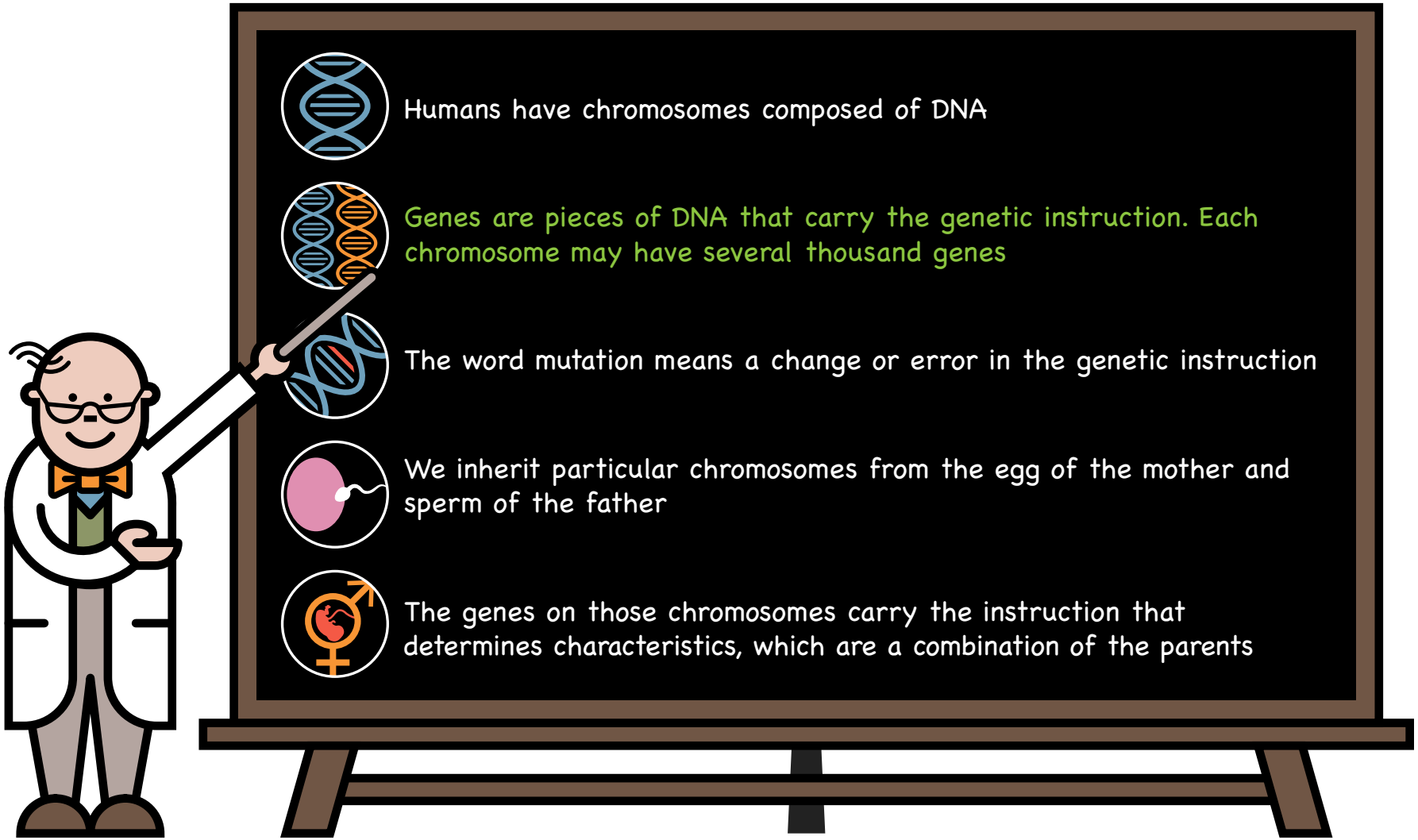


Chromosomes, genes, mutations



Chromosomes, genes, mutations



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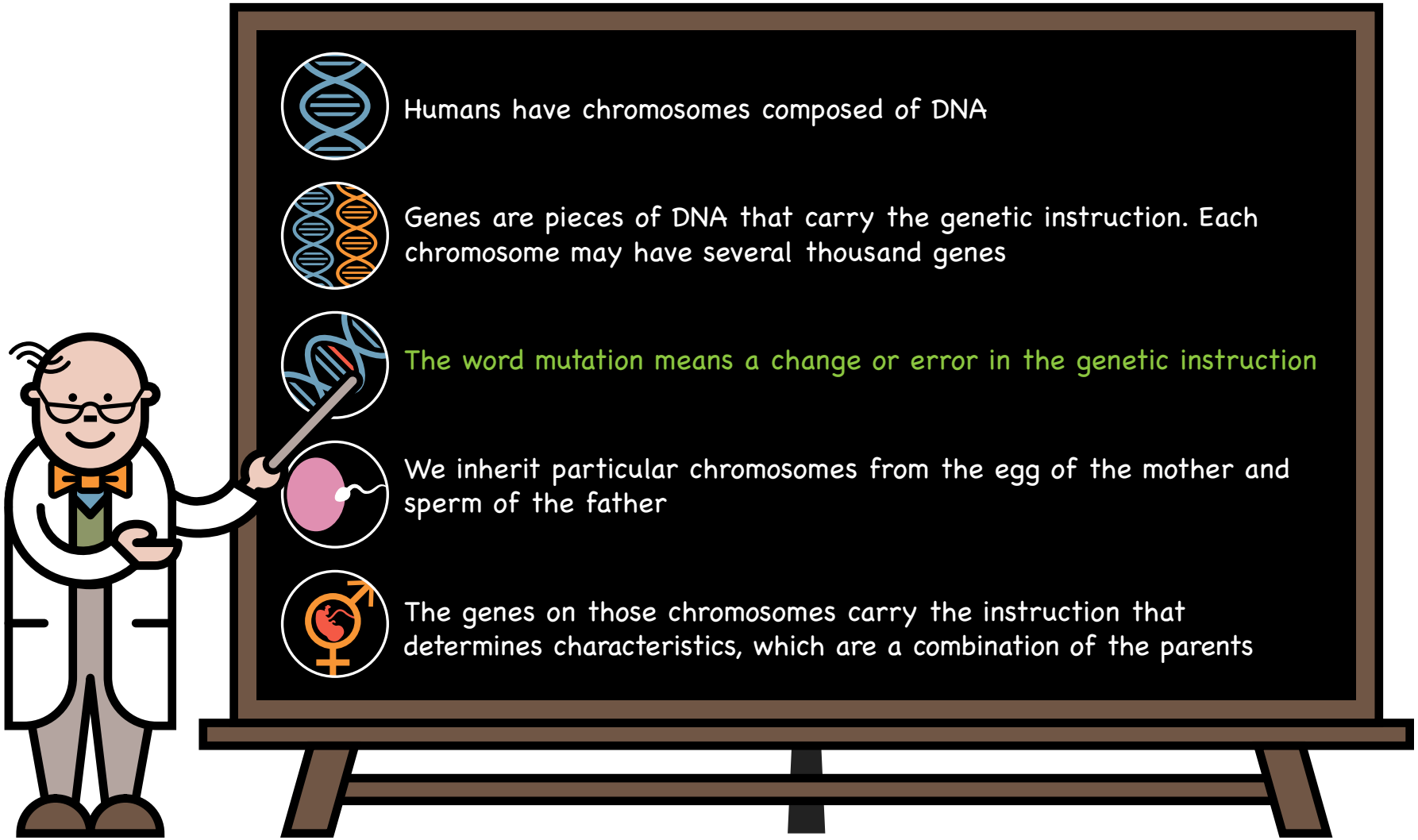

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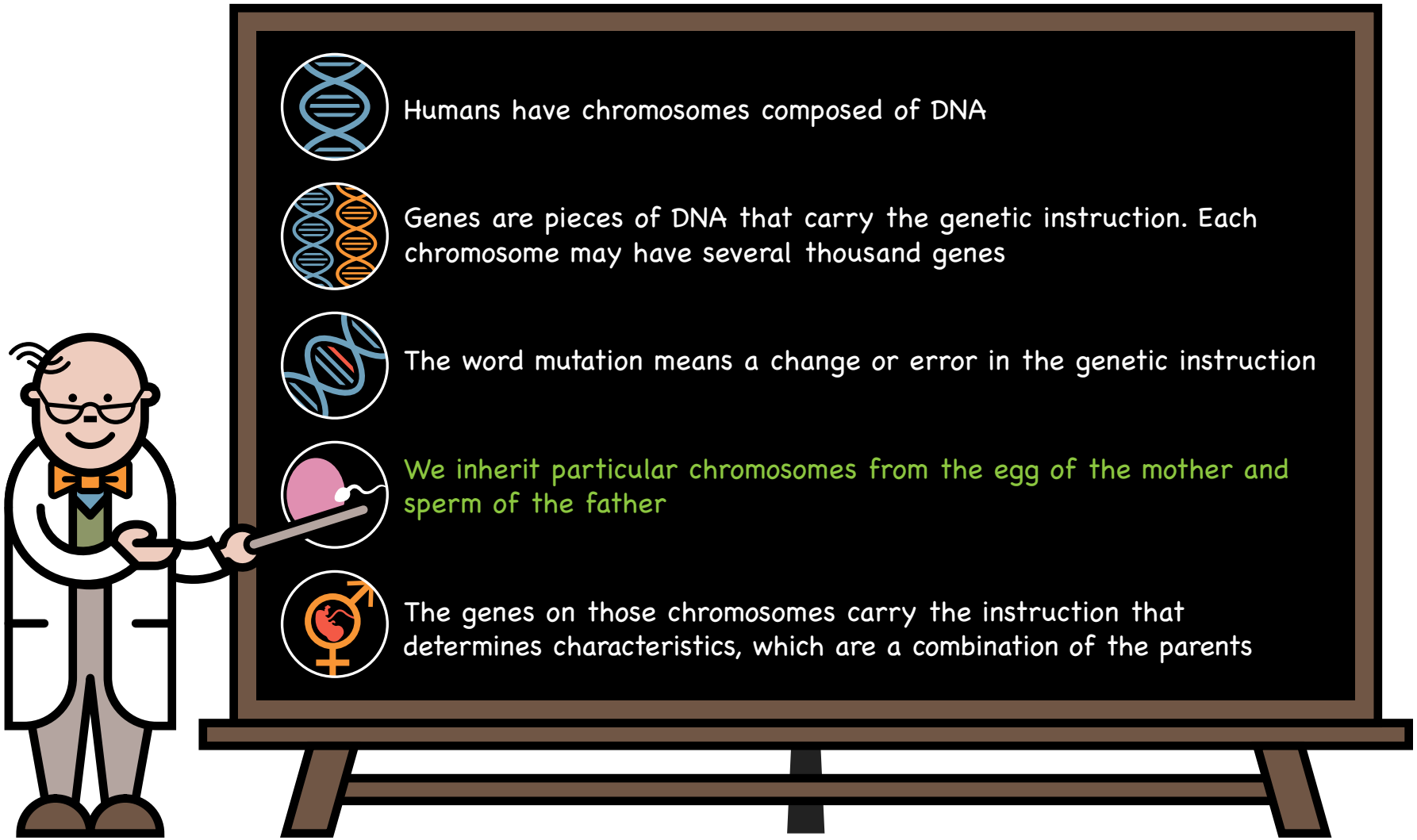

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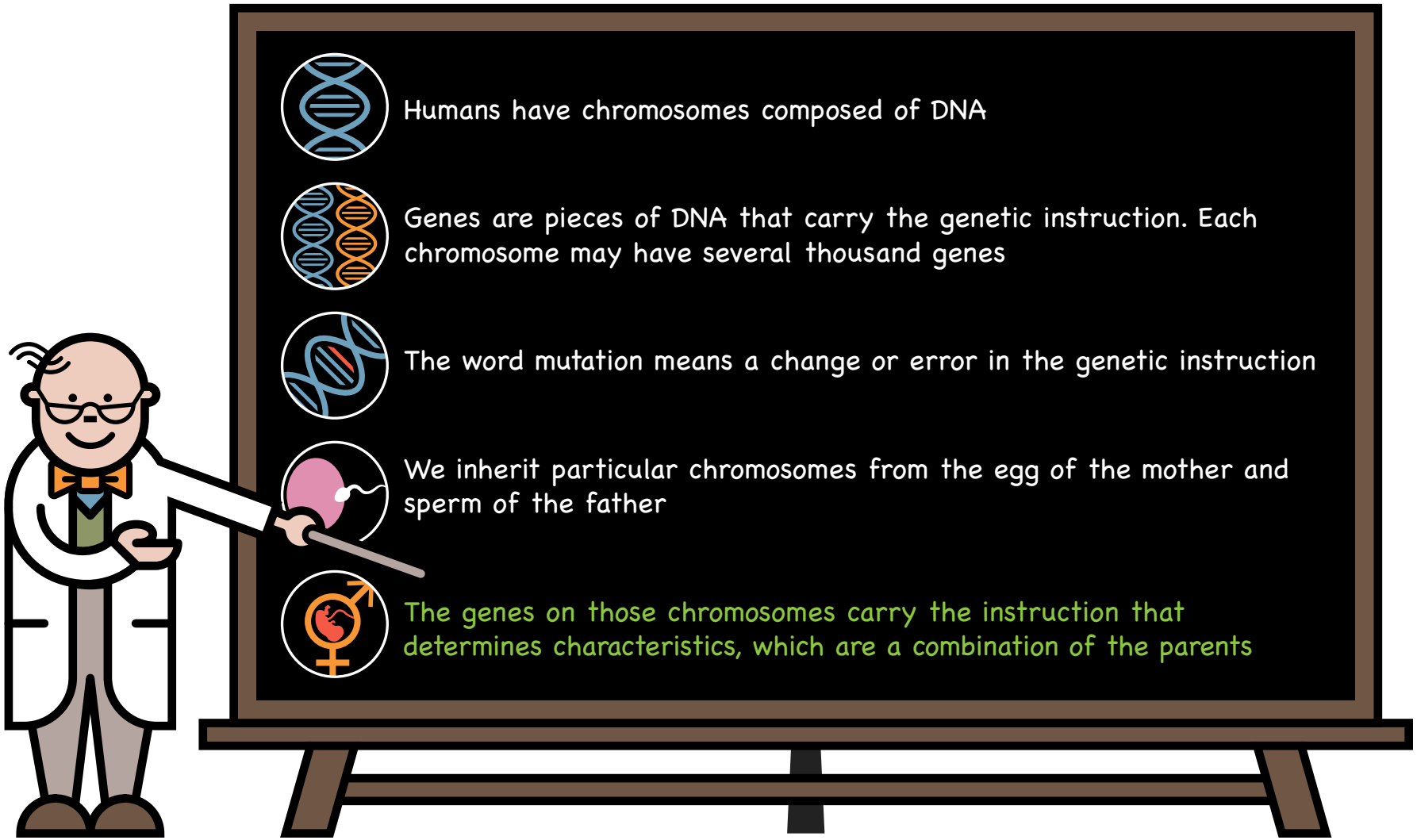
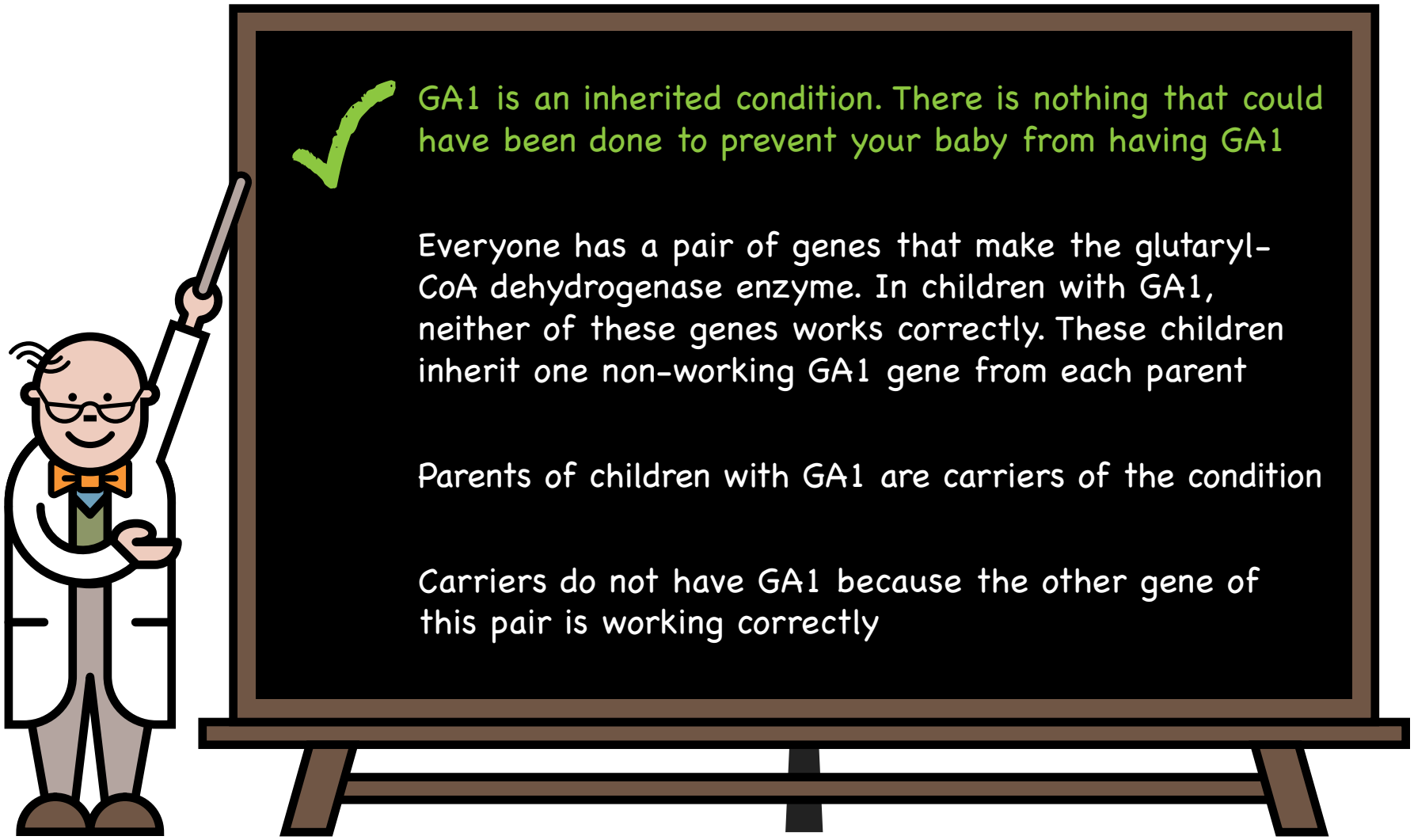


Inheritance



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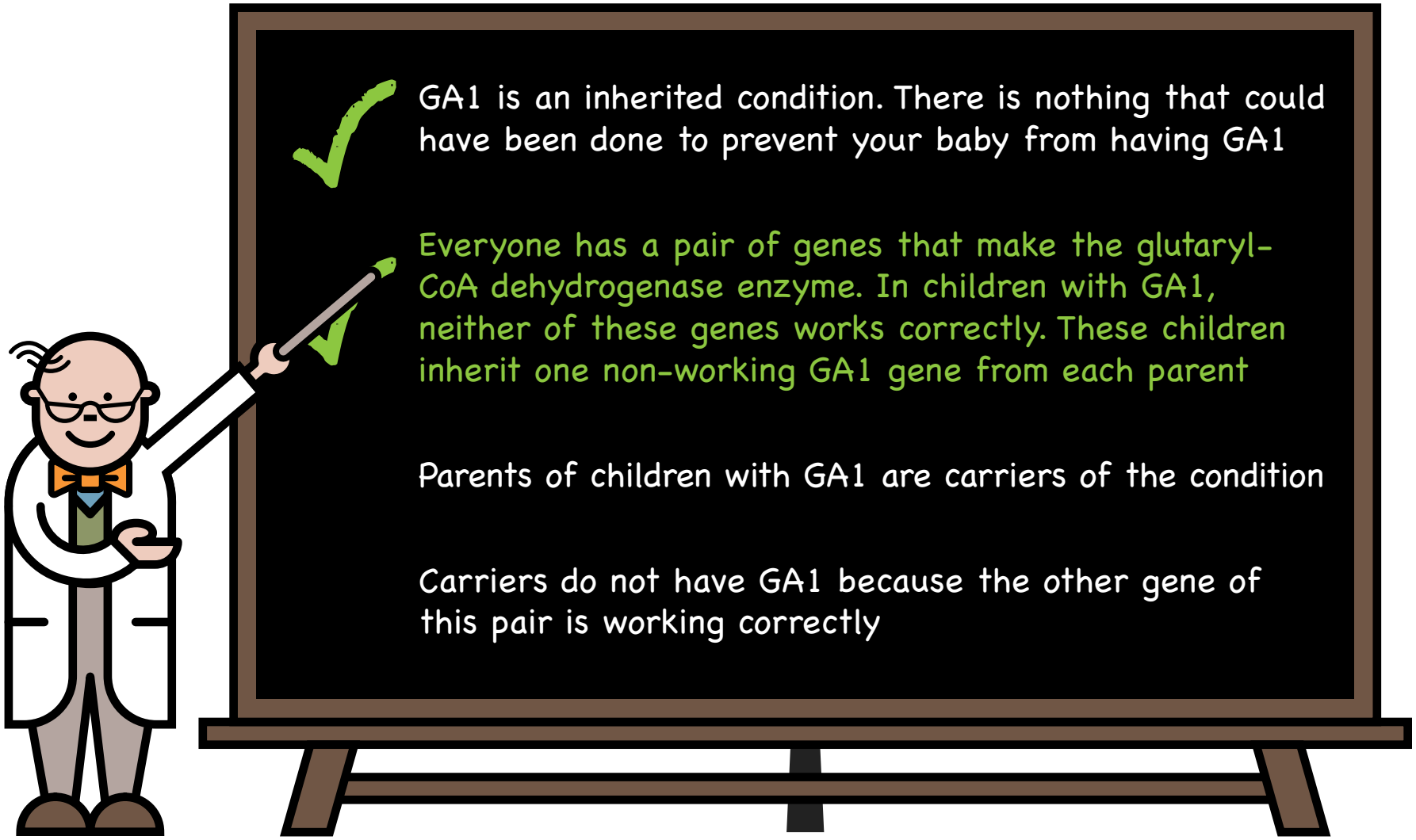

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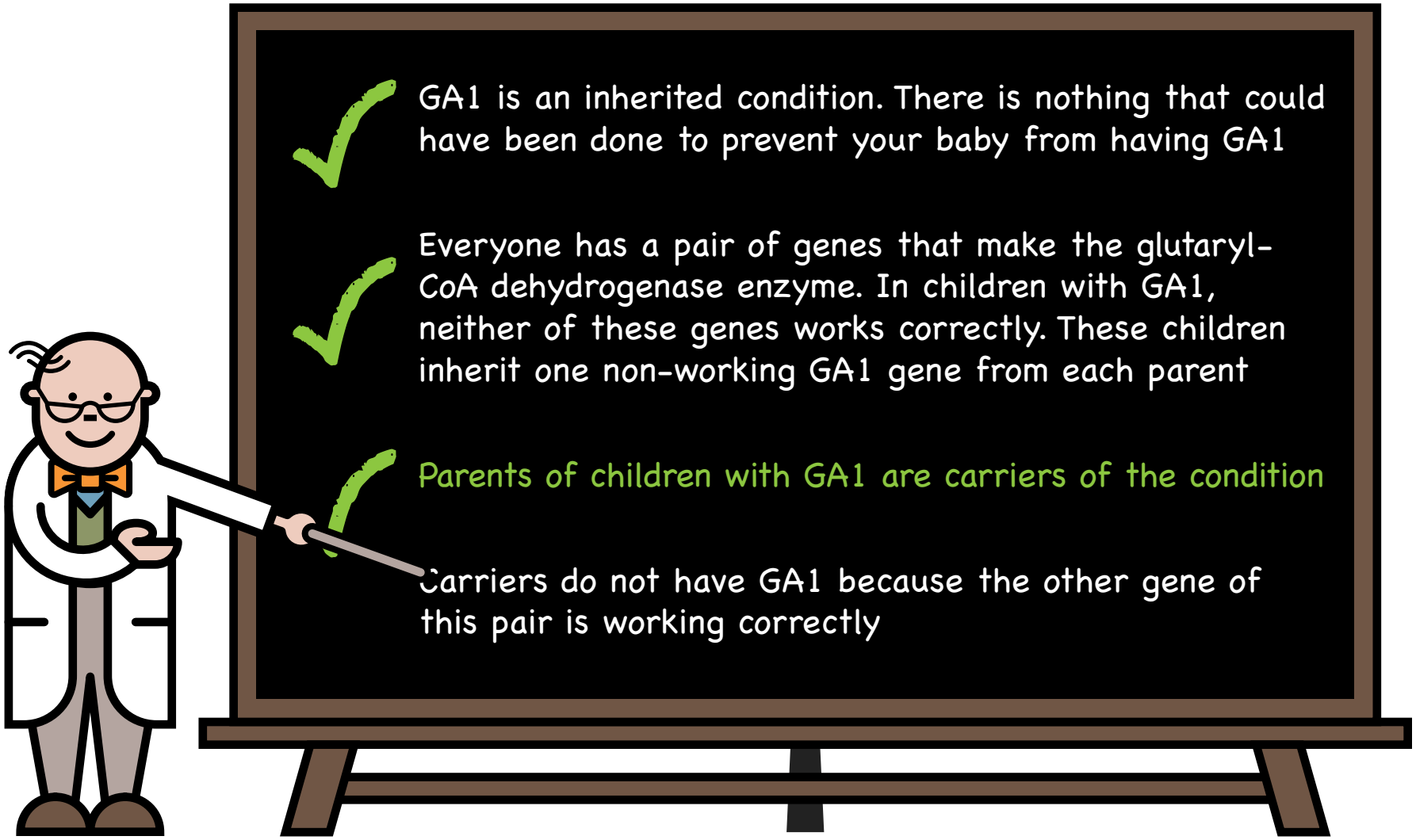

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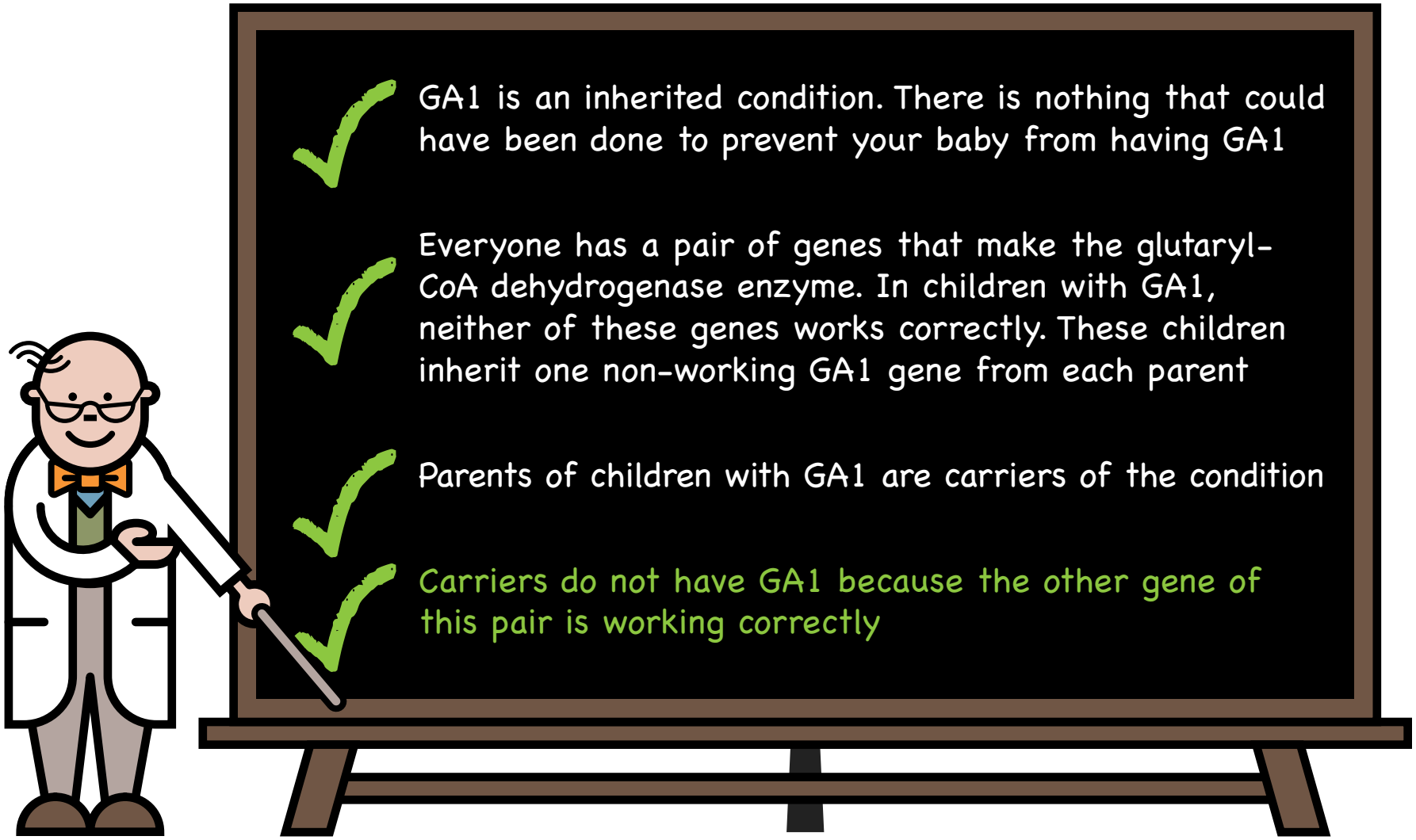
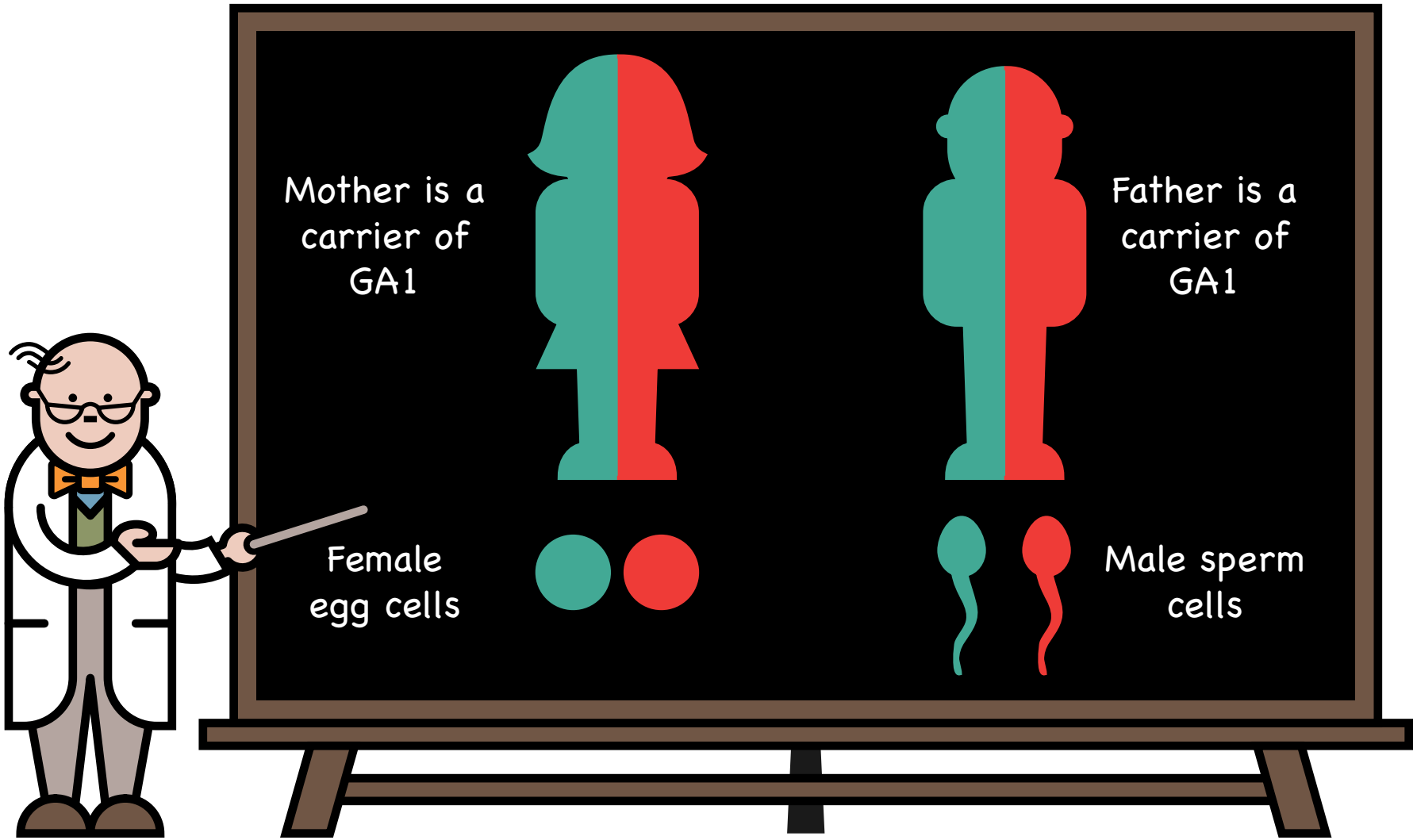
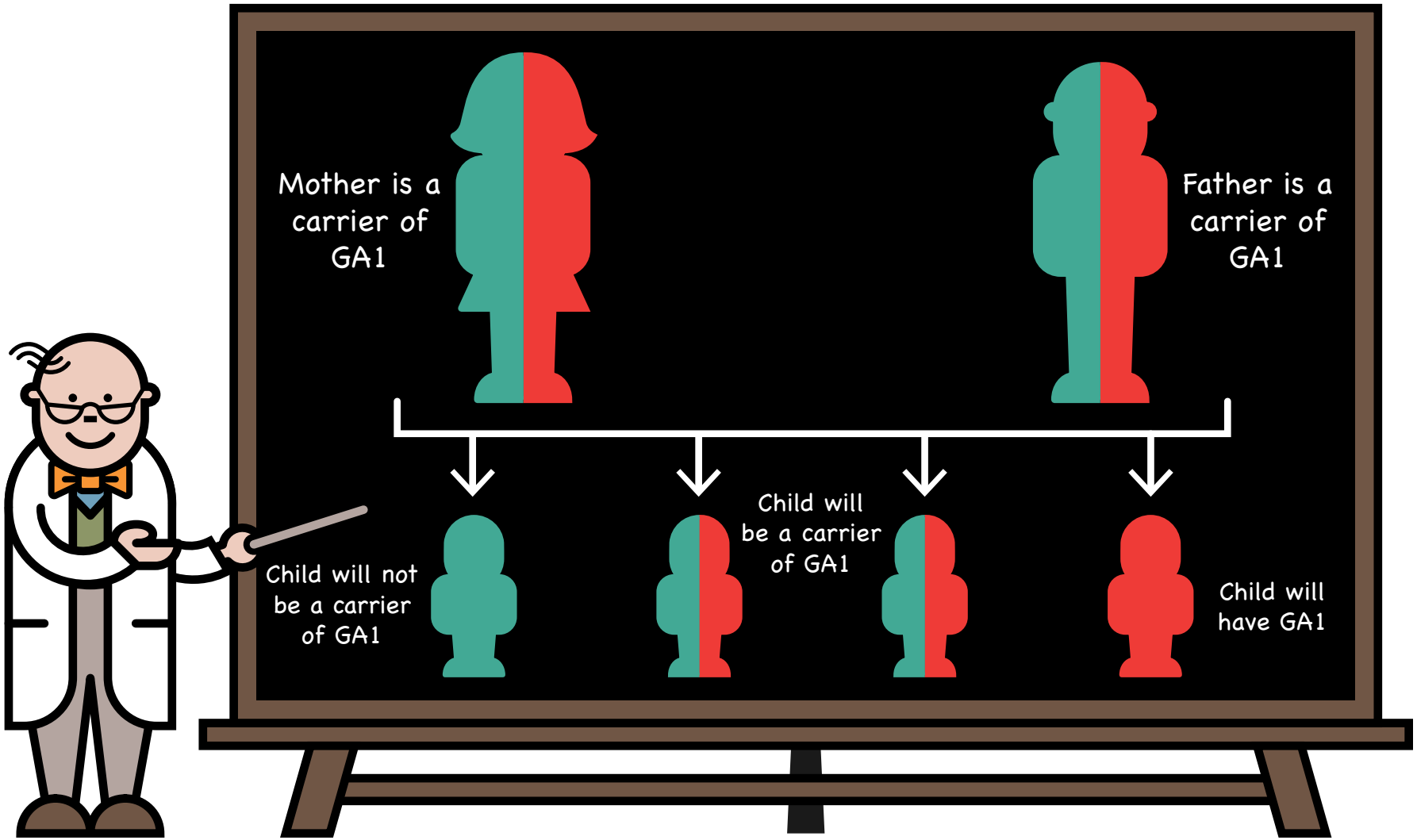


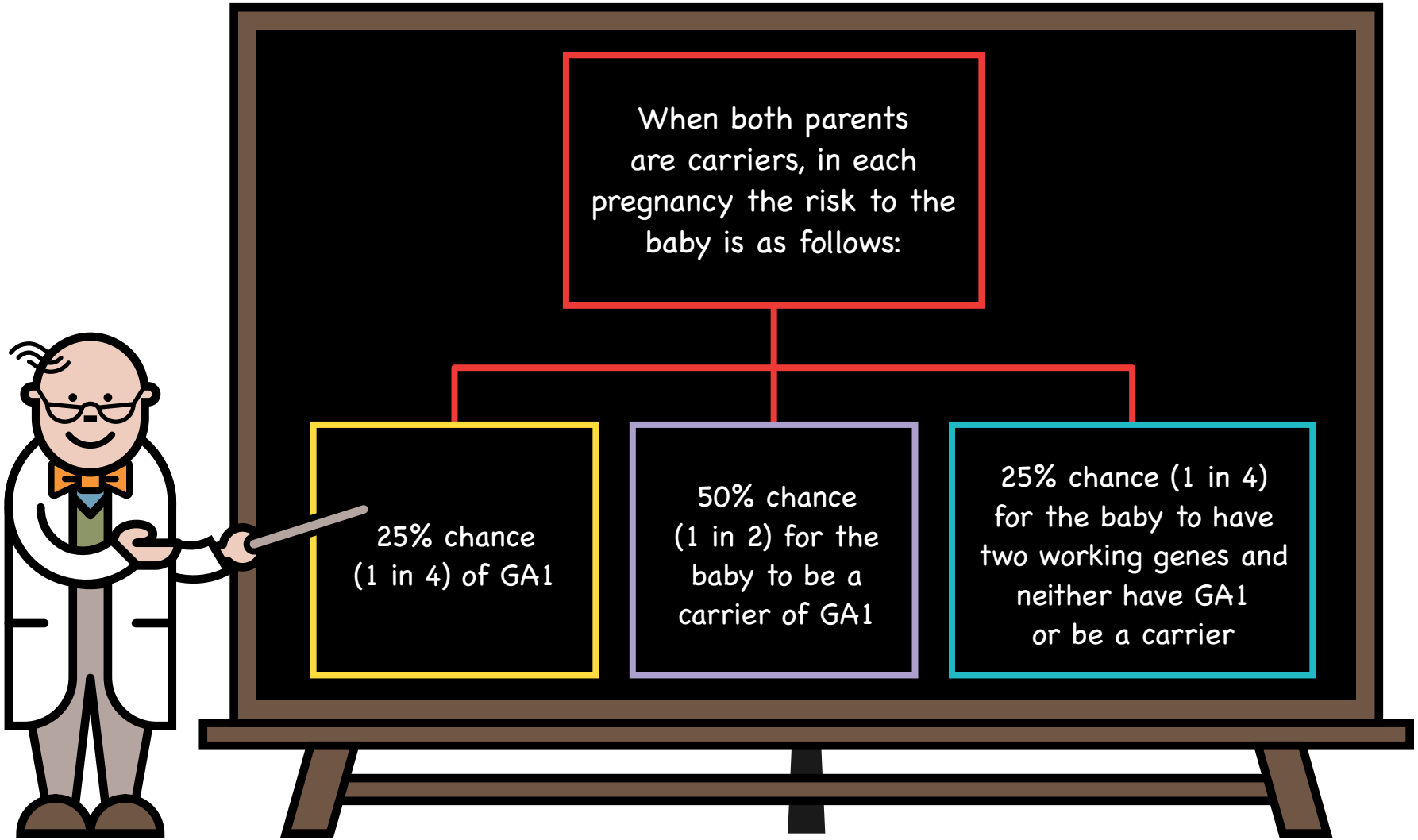
Inheritance – Autosomal recessive (carriers of GA1)



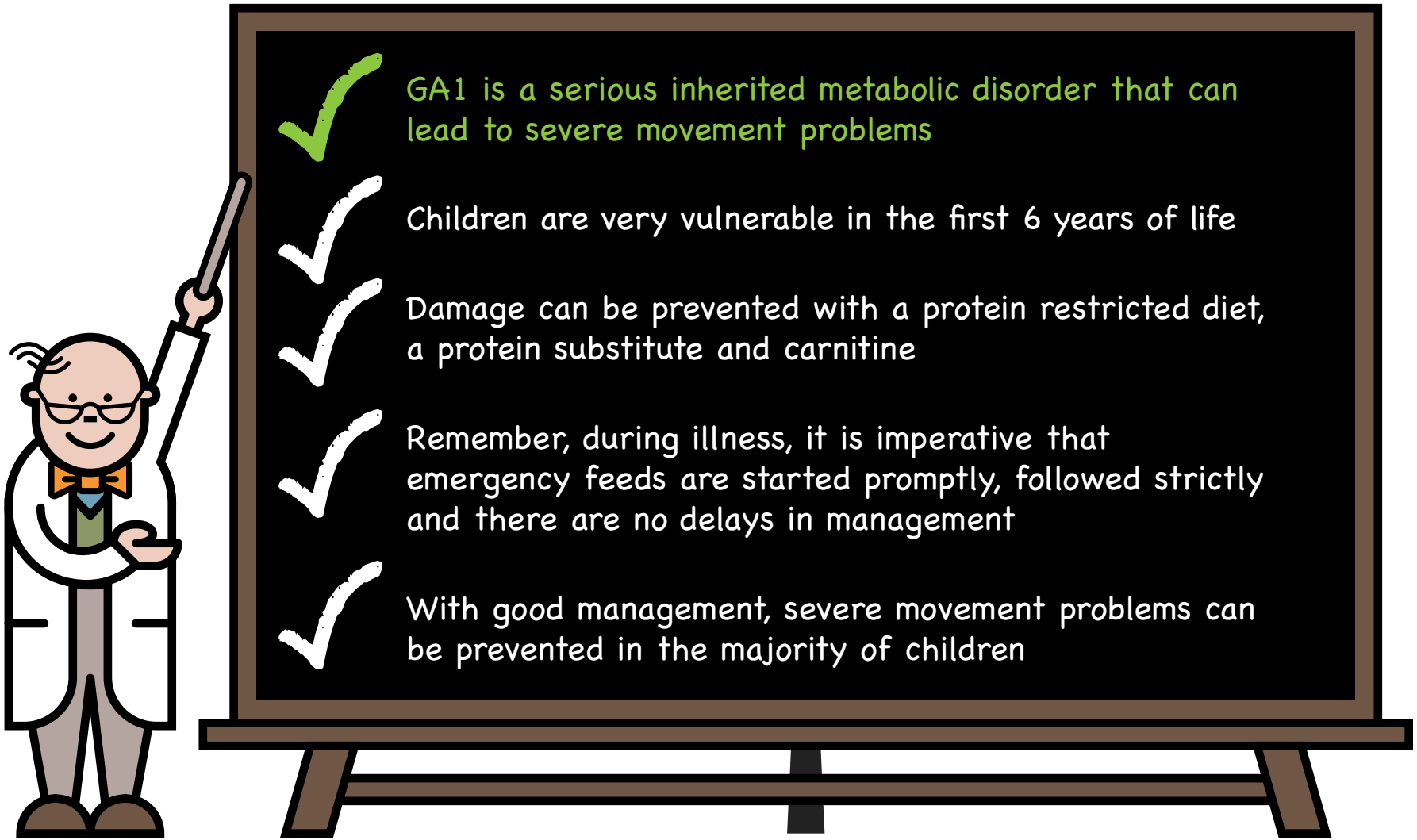
Inheritance – Autosomal recessive – possible combinations



Future pregnancies



Take home messages



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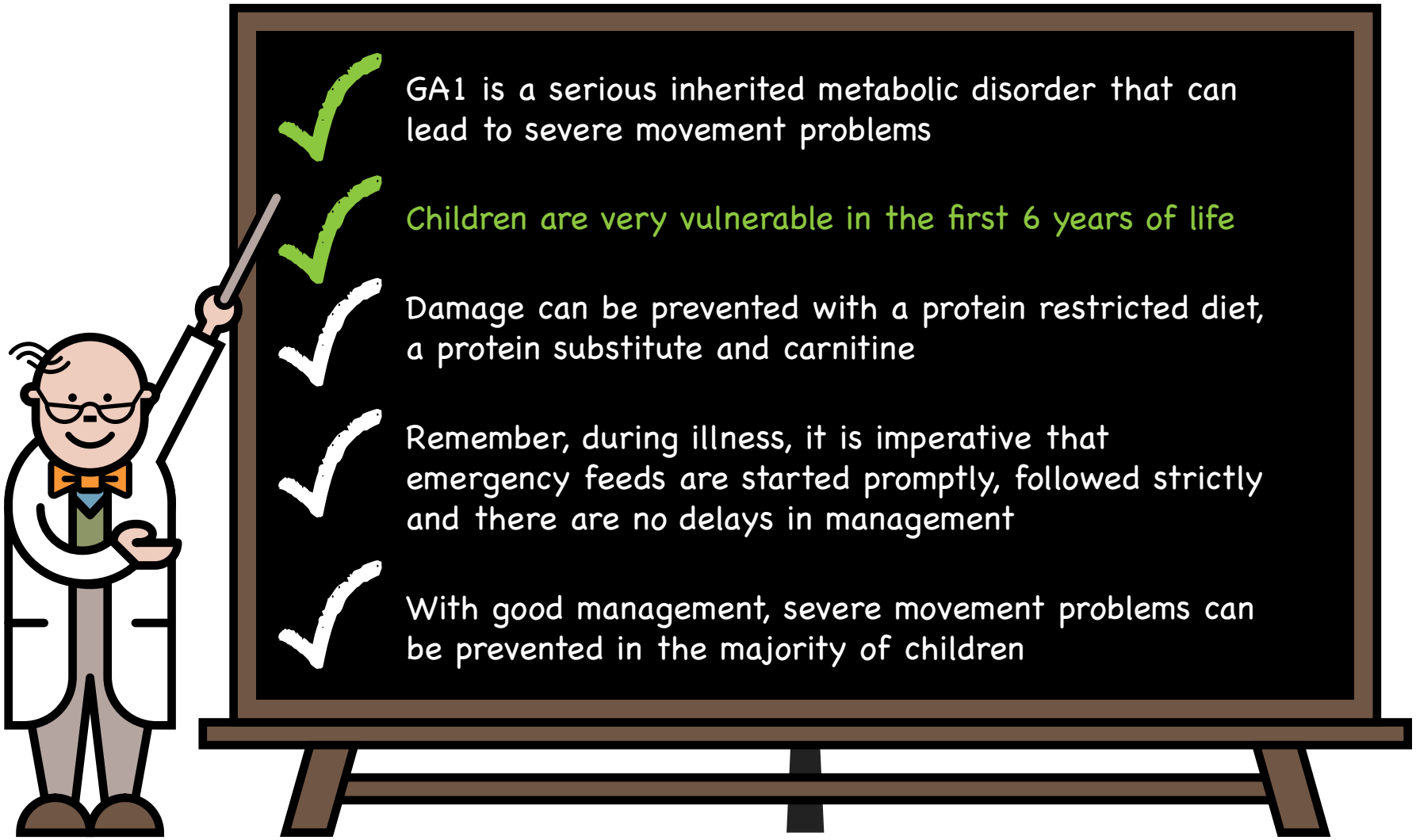

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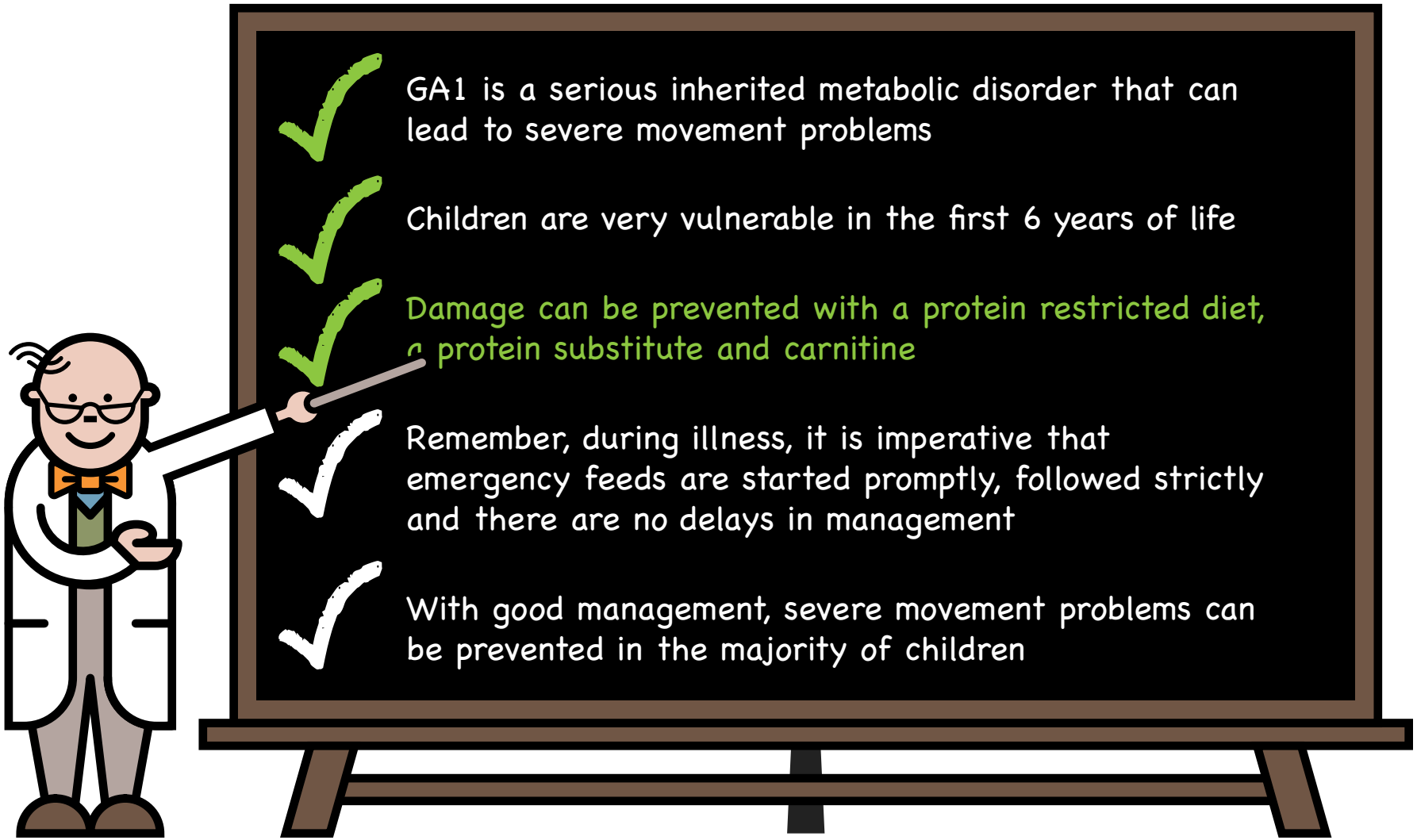

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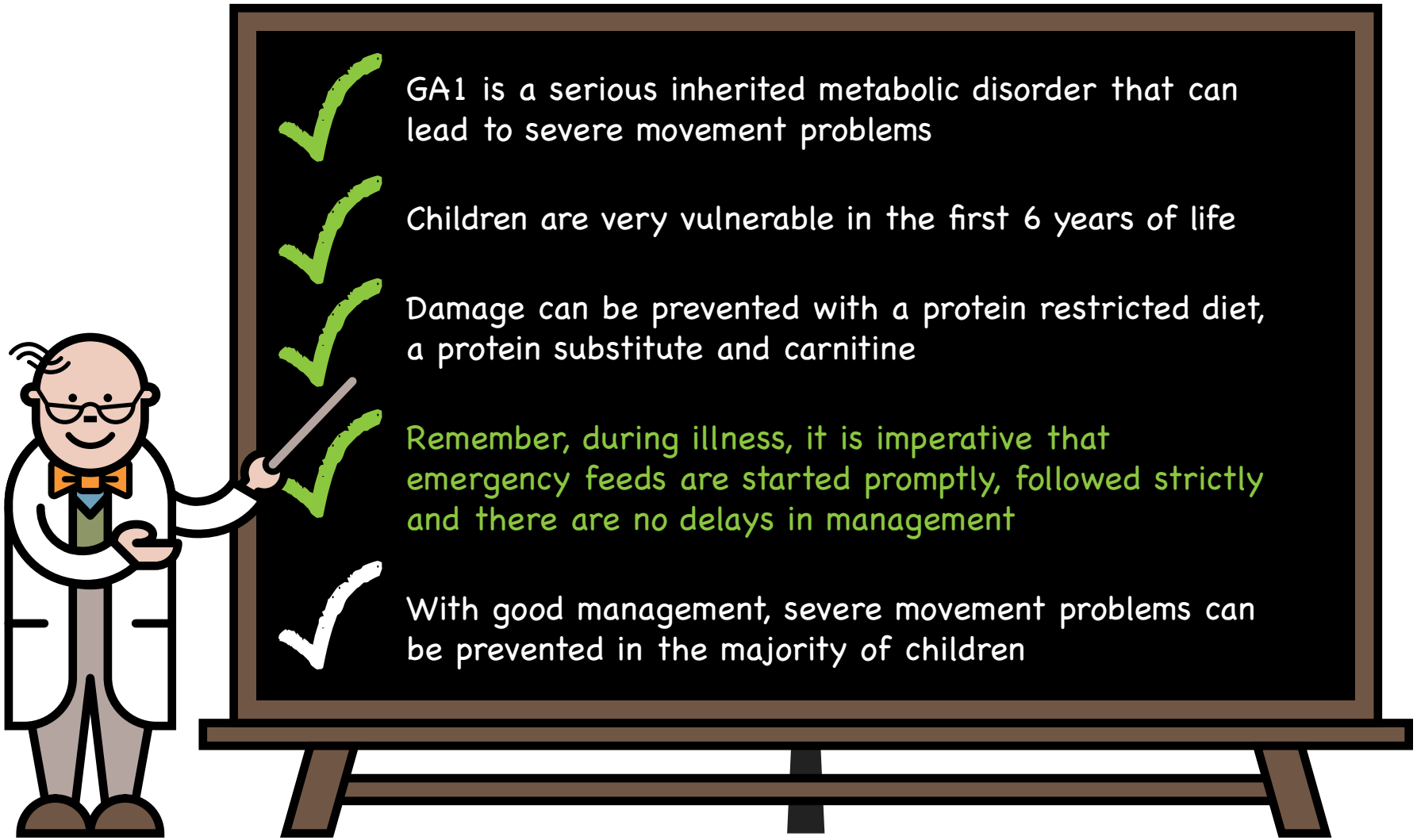

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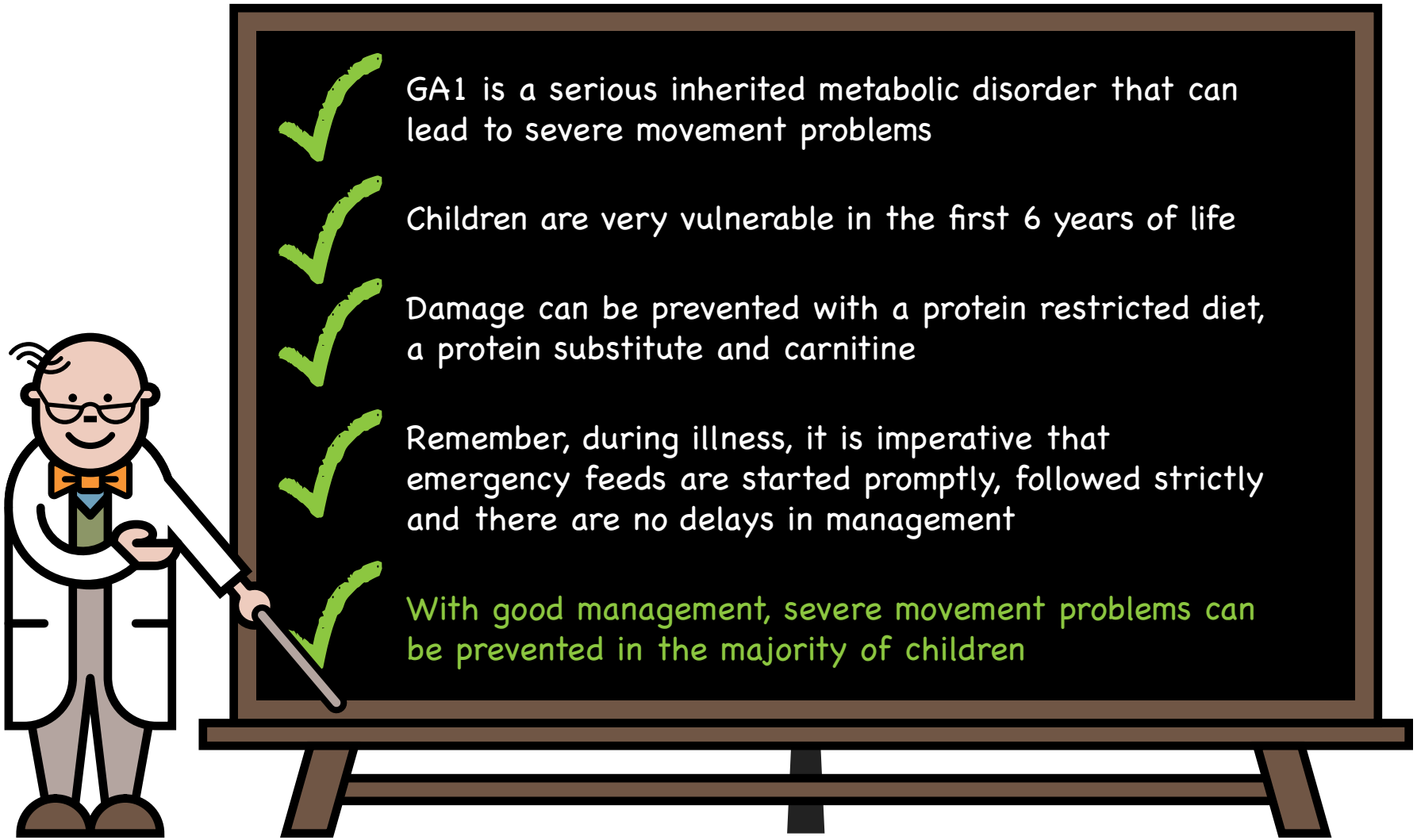

Helpful hints

Helpful hints

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Helpful hints

Who's who

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

Visit **www.lowproteinconnect.com** 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.

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