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 5, MARCH 2025

HCU

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.

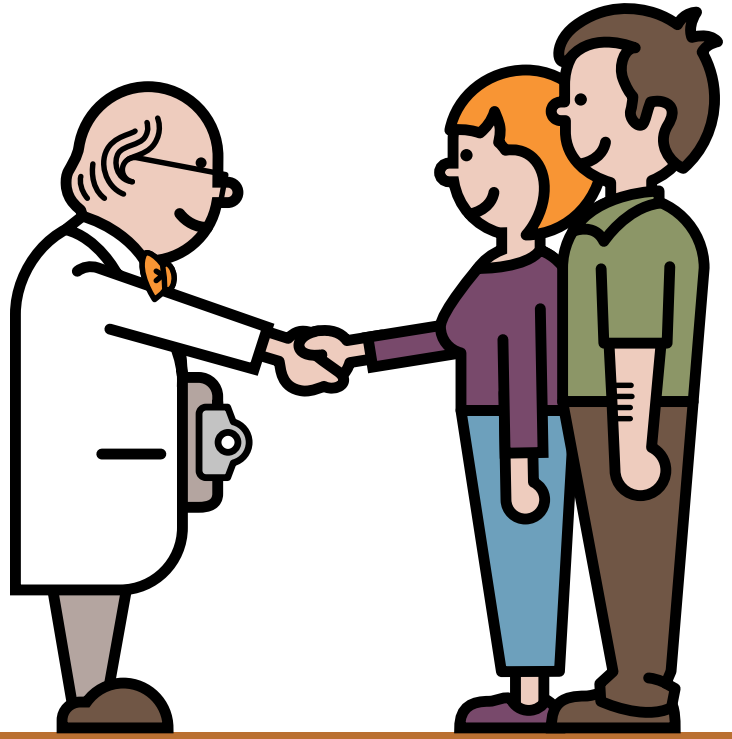
Each module produced is reviewed by a consultant clinician who is a member of the BIMDG.

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

HCU

Information for families following a positive newborn screening



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Tools Enabling Metabolic Parents LEarning

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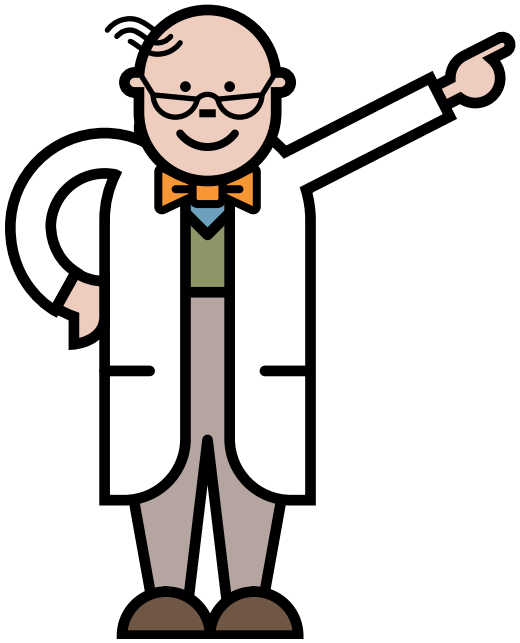
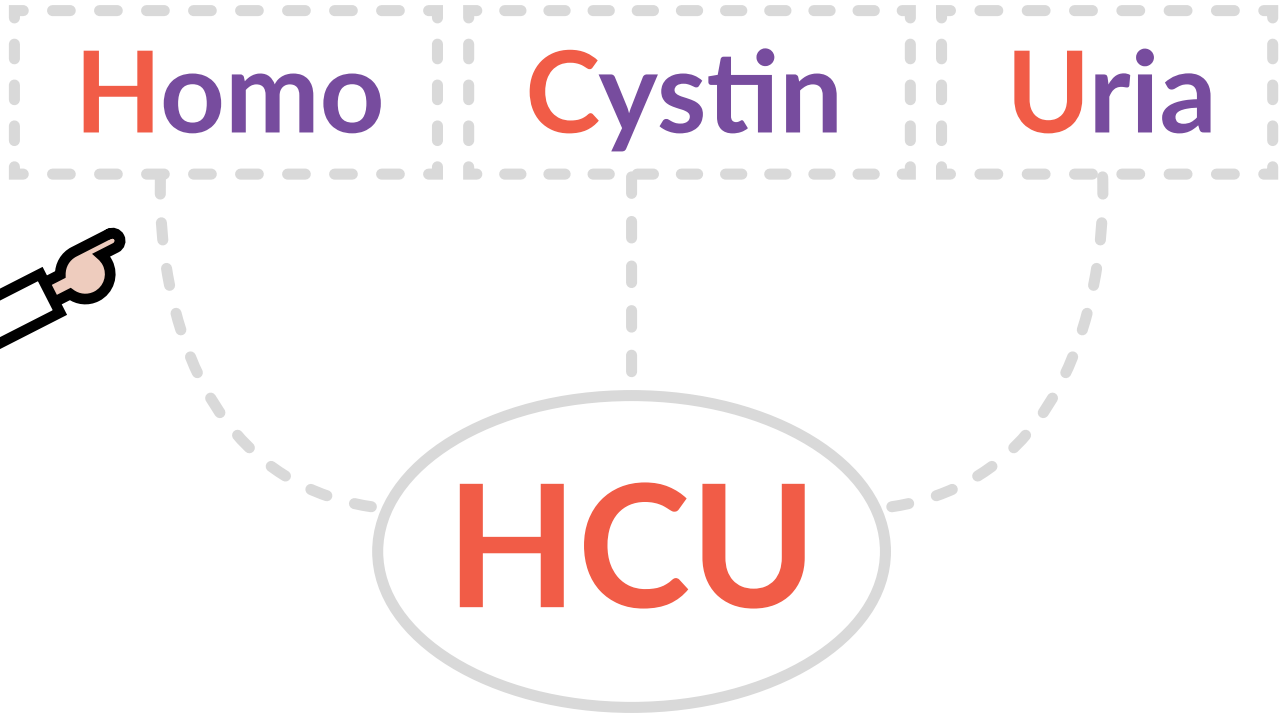
What is HCU?

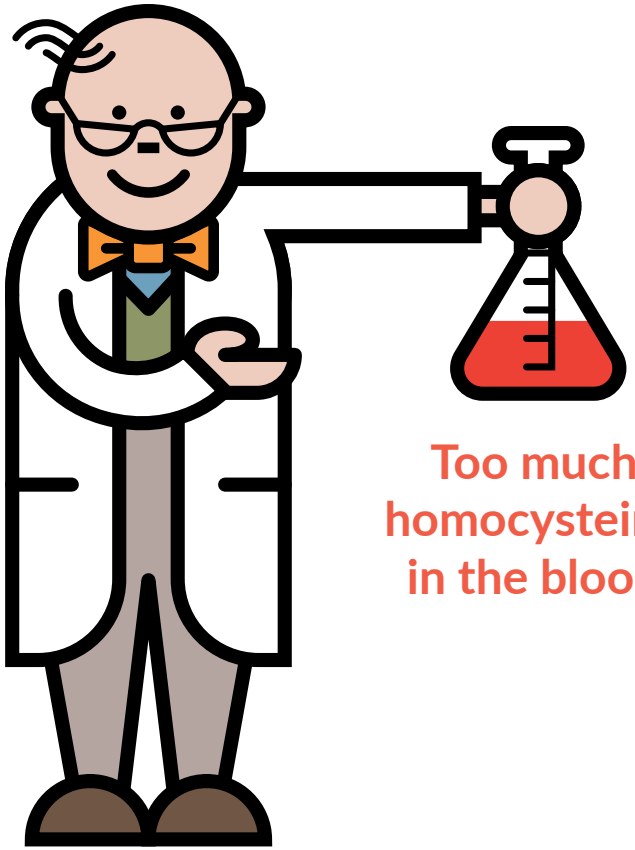
HCU stands for Homocystinuria

It is an inherited metabolic condition

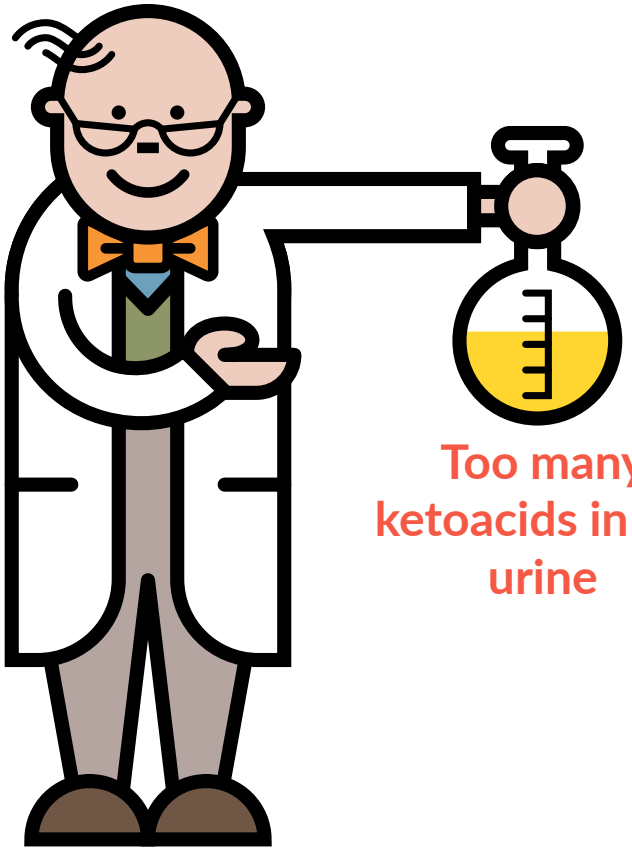




What is HCU?



Too much homocysteine in the blood



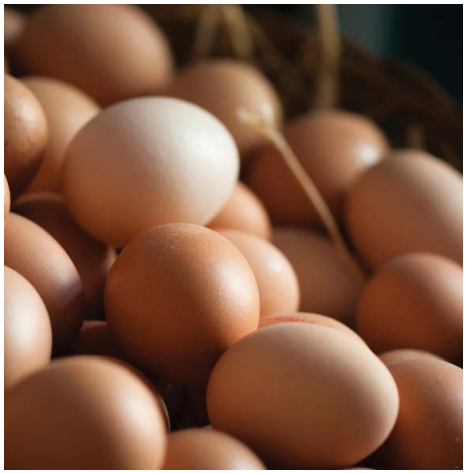
Too many ketoacids in the urine

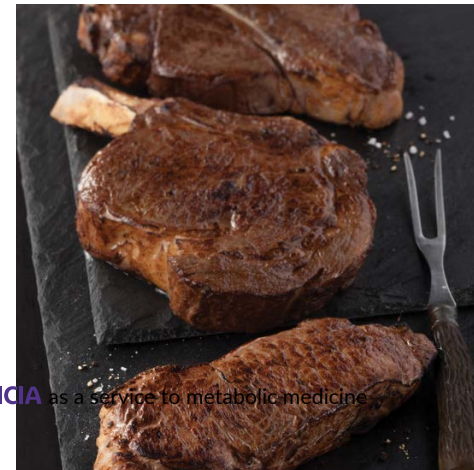
HCU and protein

HCU affects the way your baby breaks down protein

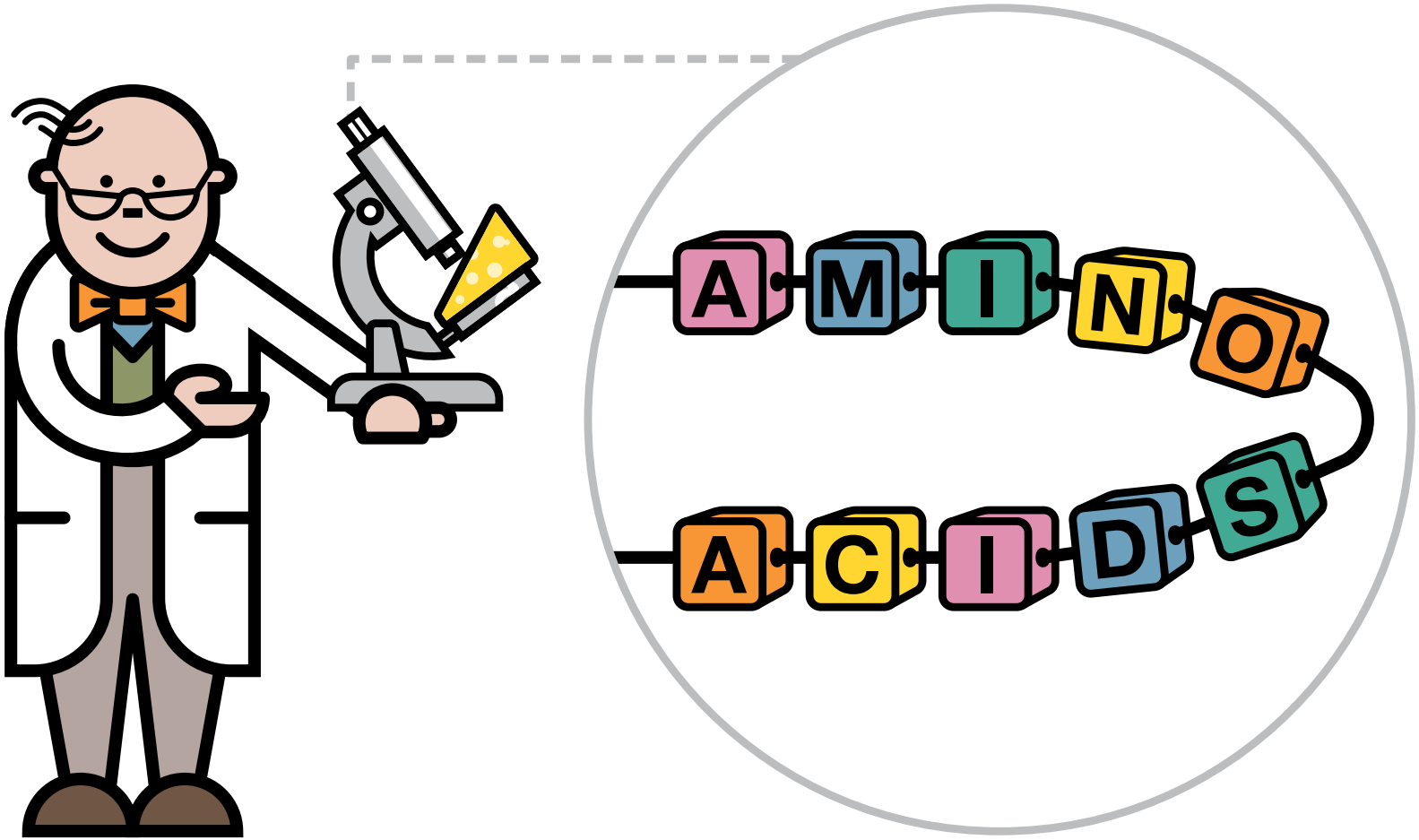
Many foods contain protein

The body needs protein for growth and repair





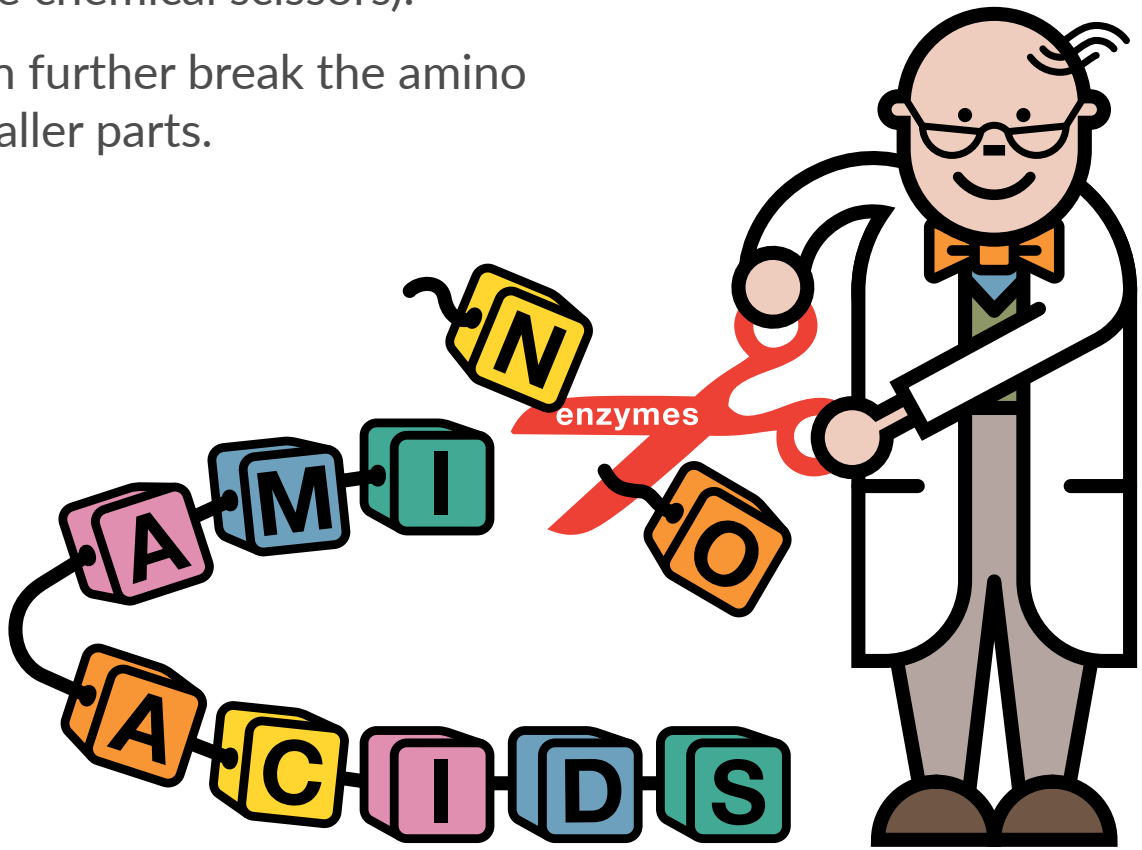
What is protein?



Protein and enzymes

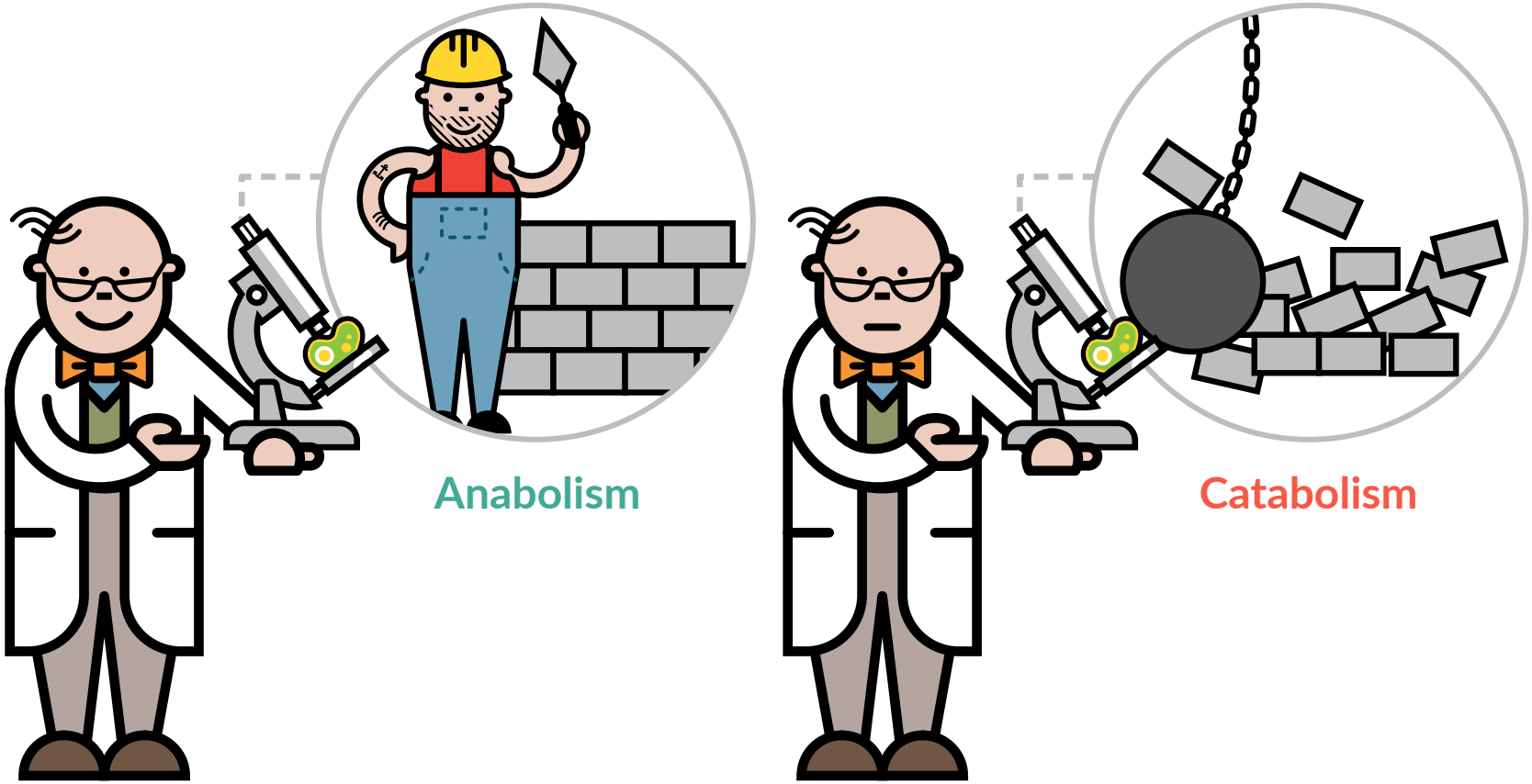
Protein is broken down into amino acids (building blocks of protein) by enzymes (which are like chemical scissors).

Enzymes then further break the amino acids into smaller parts.



Protein metabolism

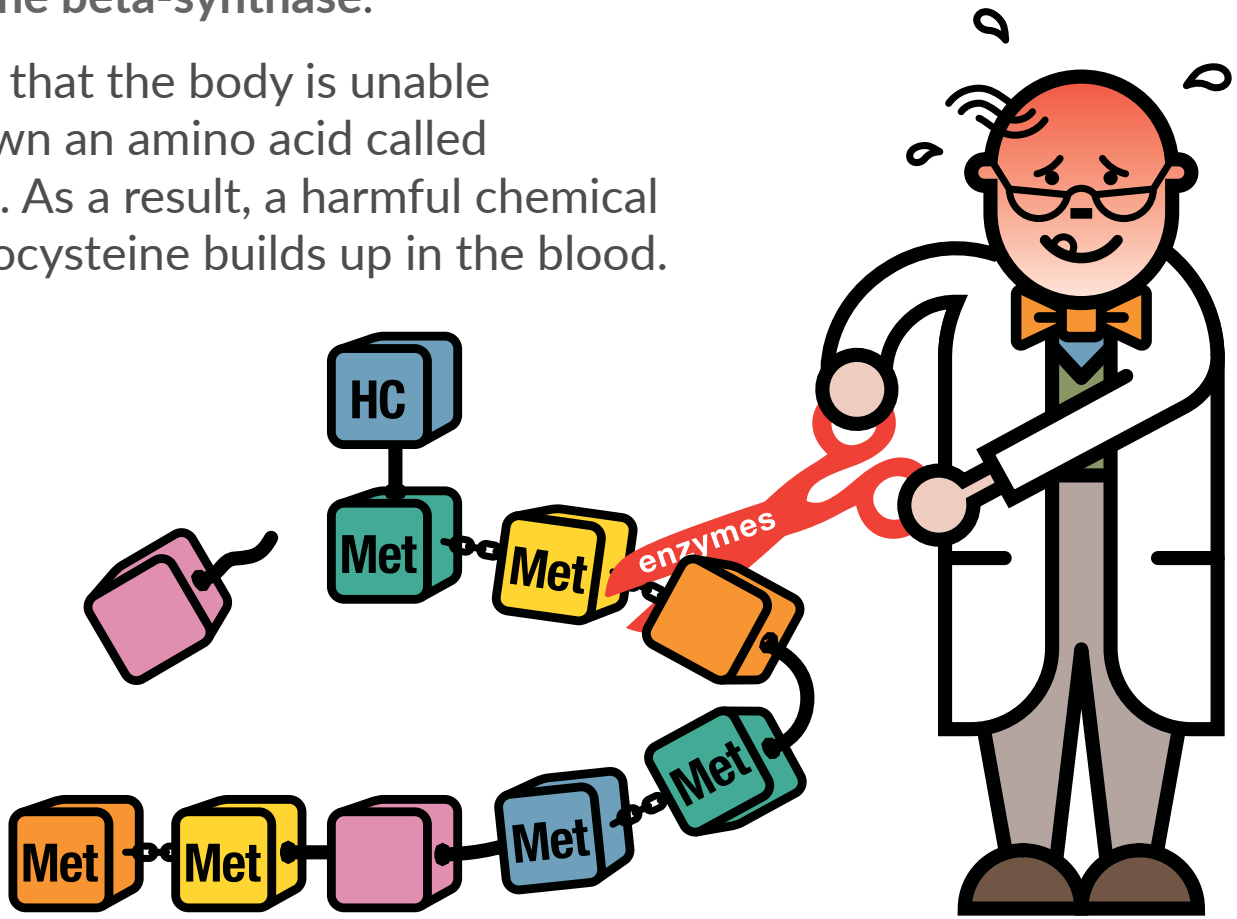
Metabolism refers to the chemical processes that occur inside the cells of the body.



What happens in HCU?

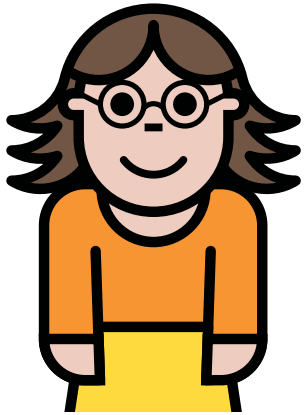
In HCU the body lacks an enzyme called **cystathionine beta-synthase**.

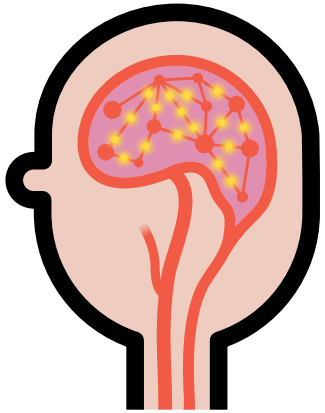
This means that the body is unable to breakdown an amino acid called methionine. As a result, a harmful chemical called homocysteine builds up in the blood.



What can go wrong in HCU?

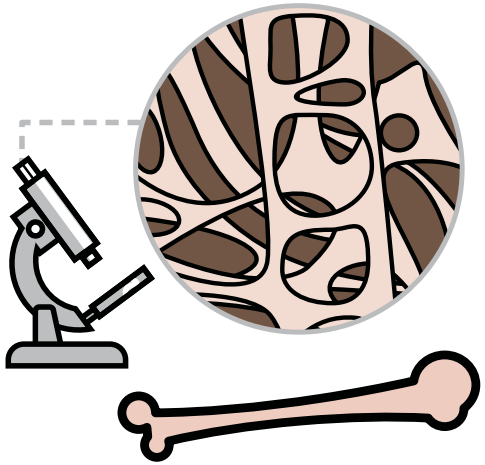
Short sightedness and dislocated eye lenses

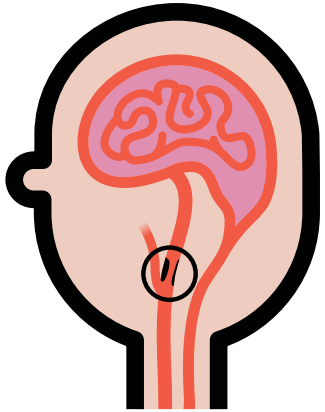




Difficulties with learning and behaviour

Long and thin bones



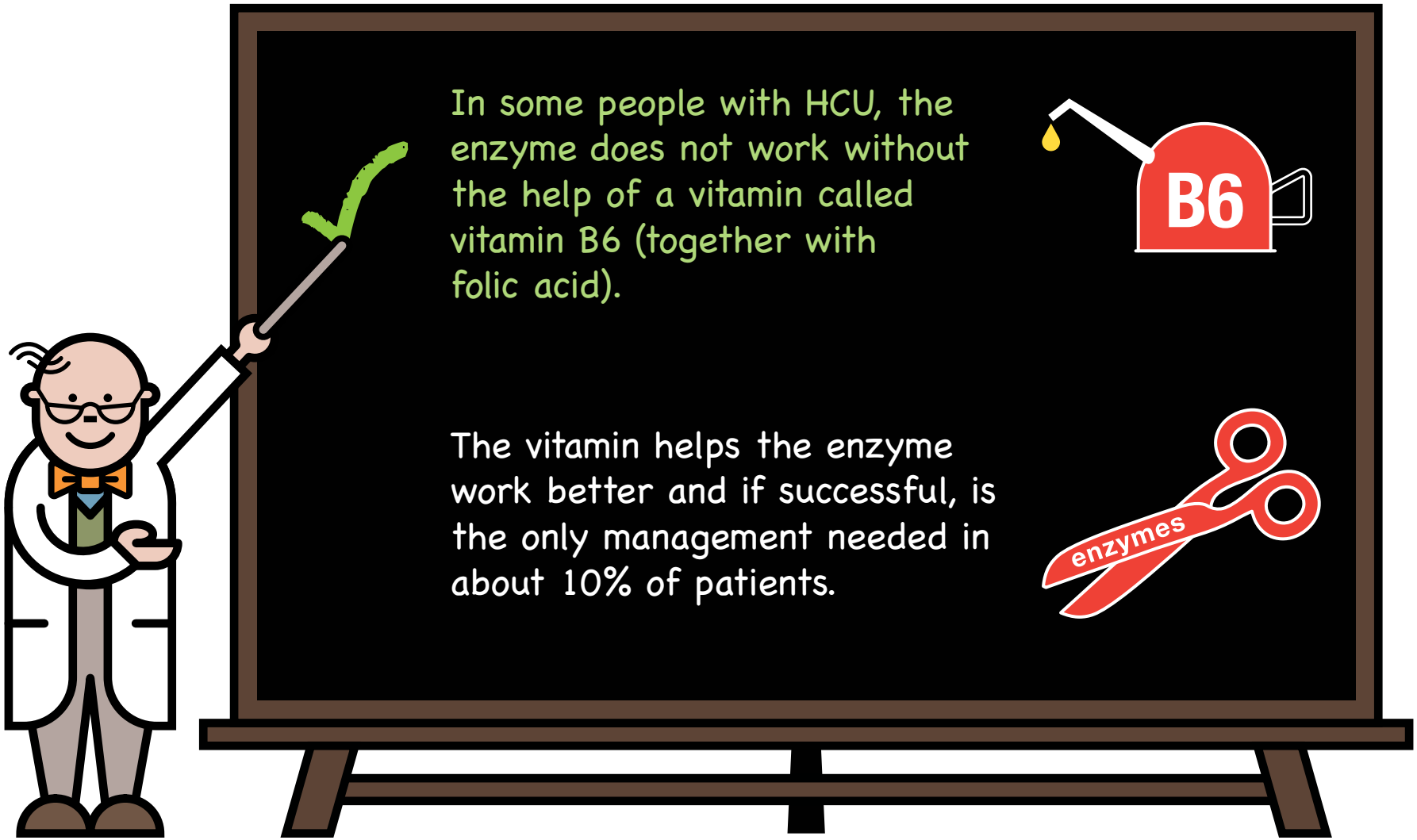


Blood clots and strokes

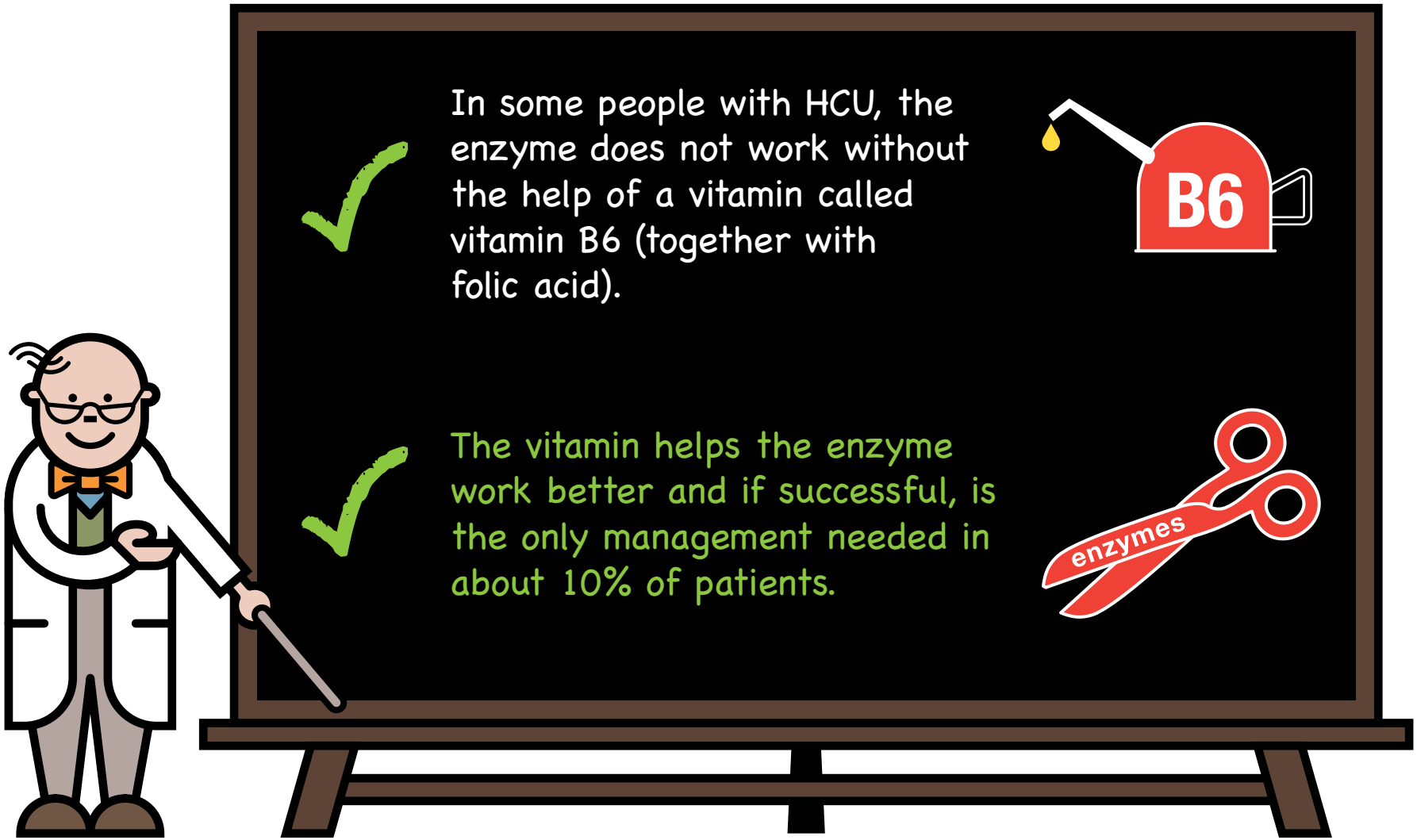
How is HCU diagnosed?

HCU is diagnosed by newborn screening. High levels of methionine and homocysteine are found in the blood.

How is HCU managed?



How is HCU managed?



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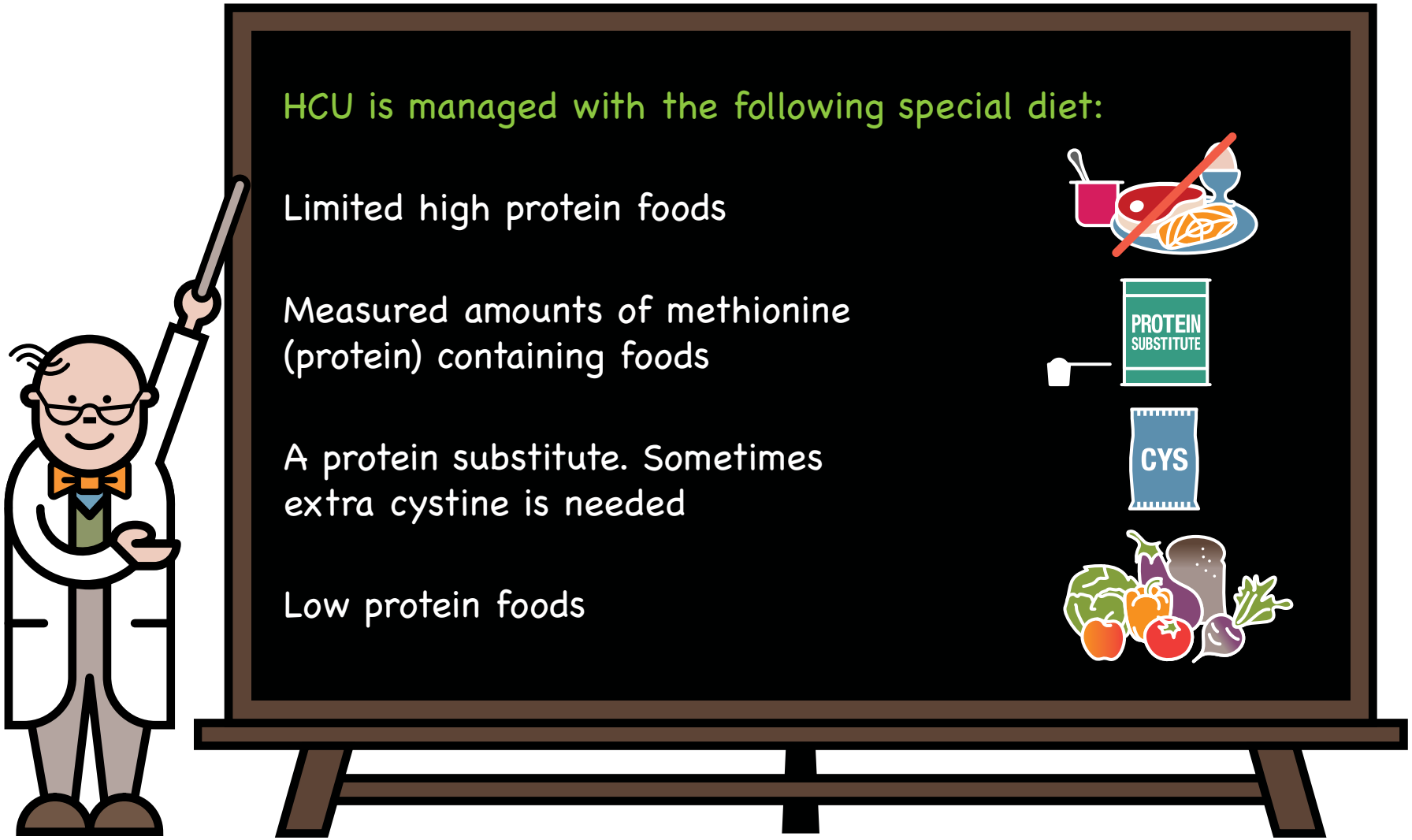

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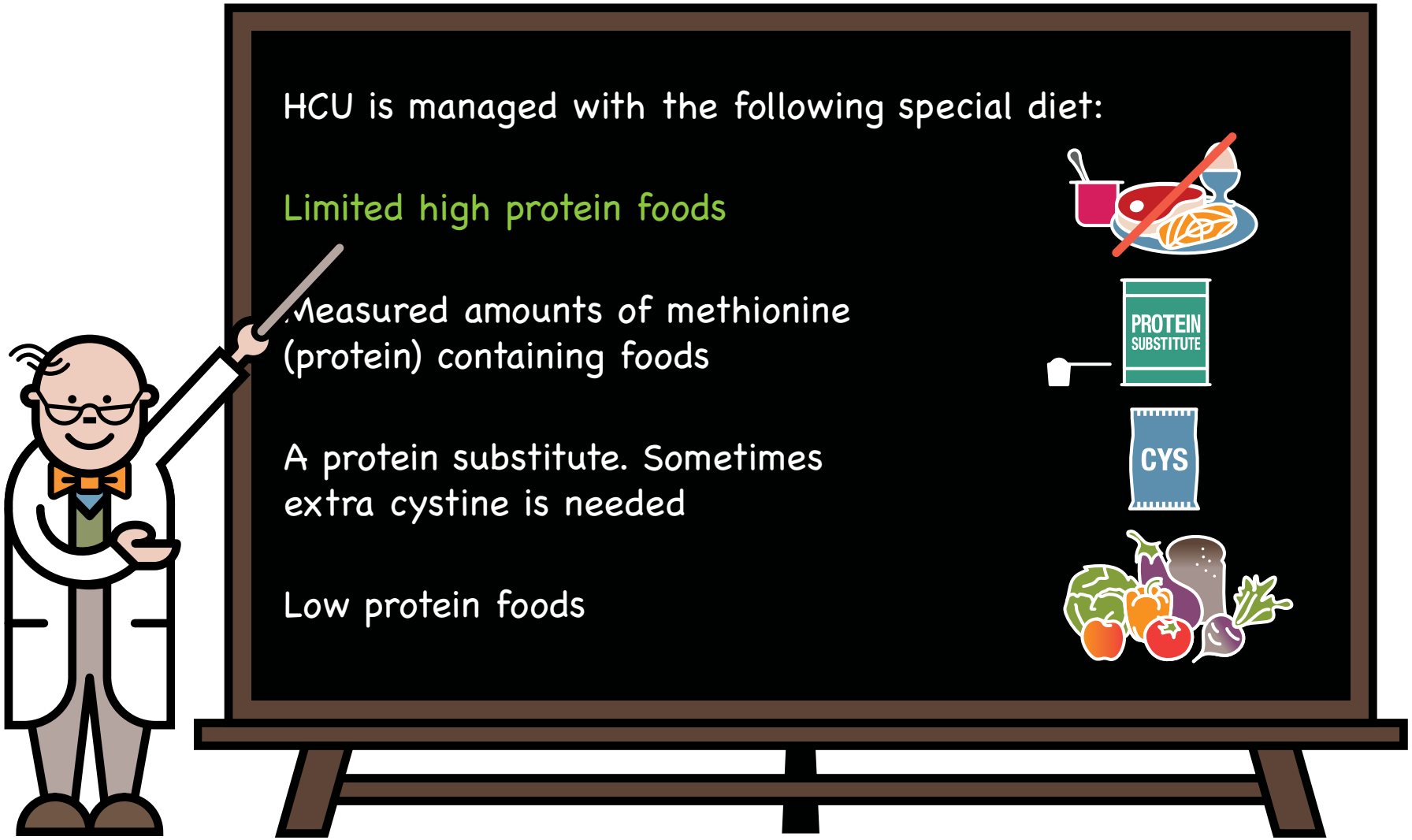

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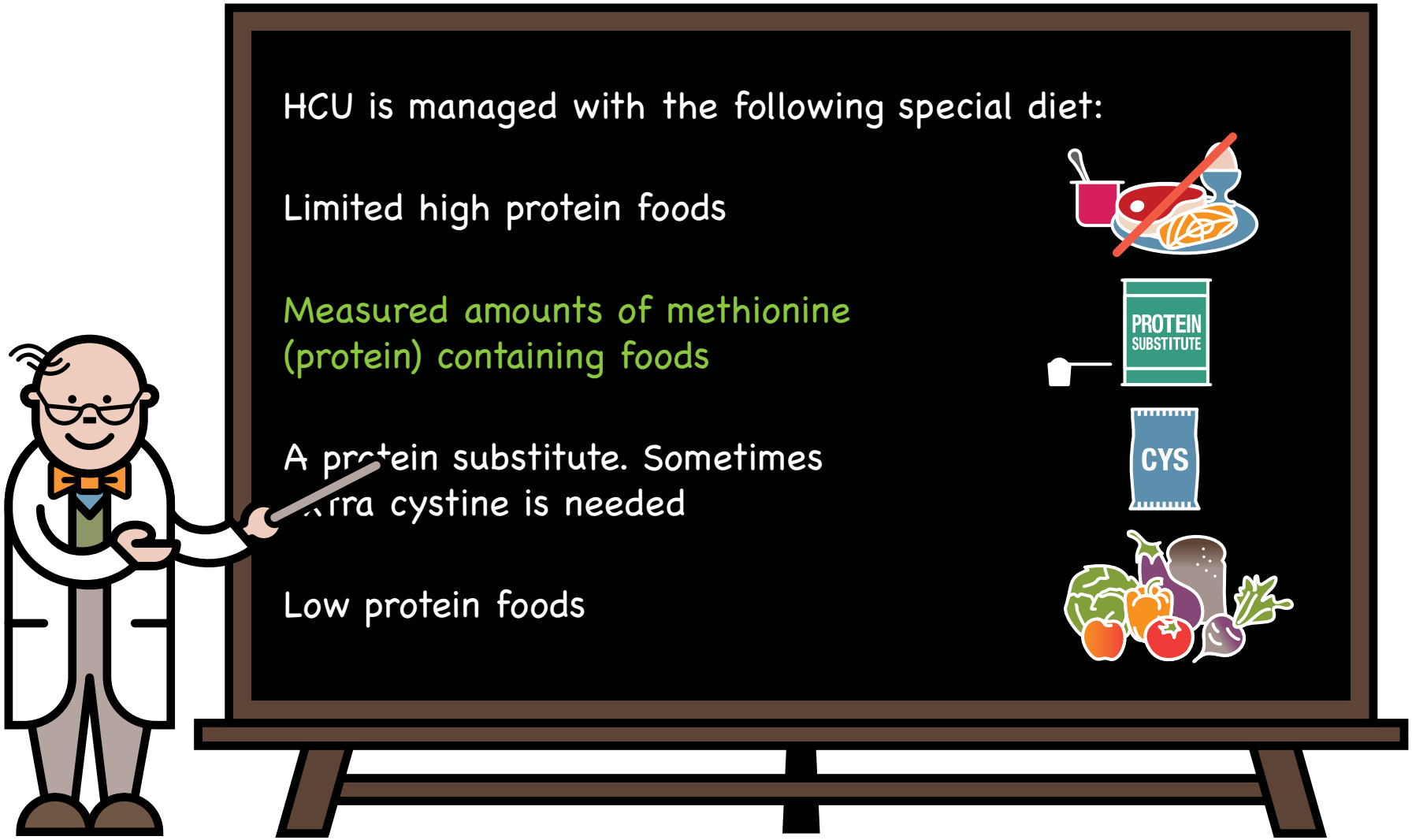

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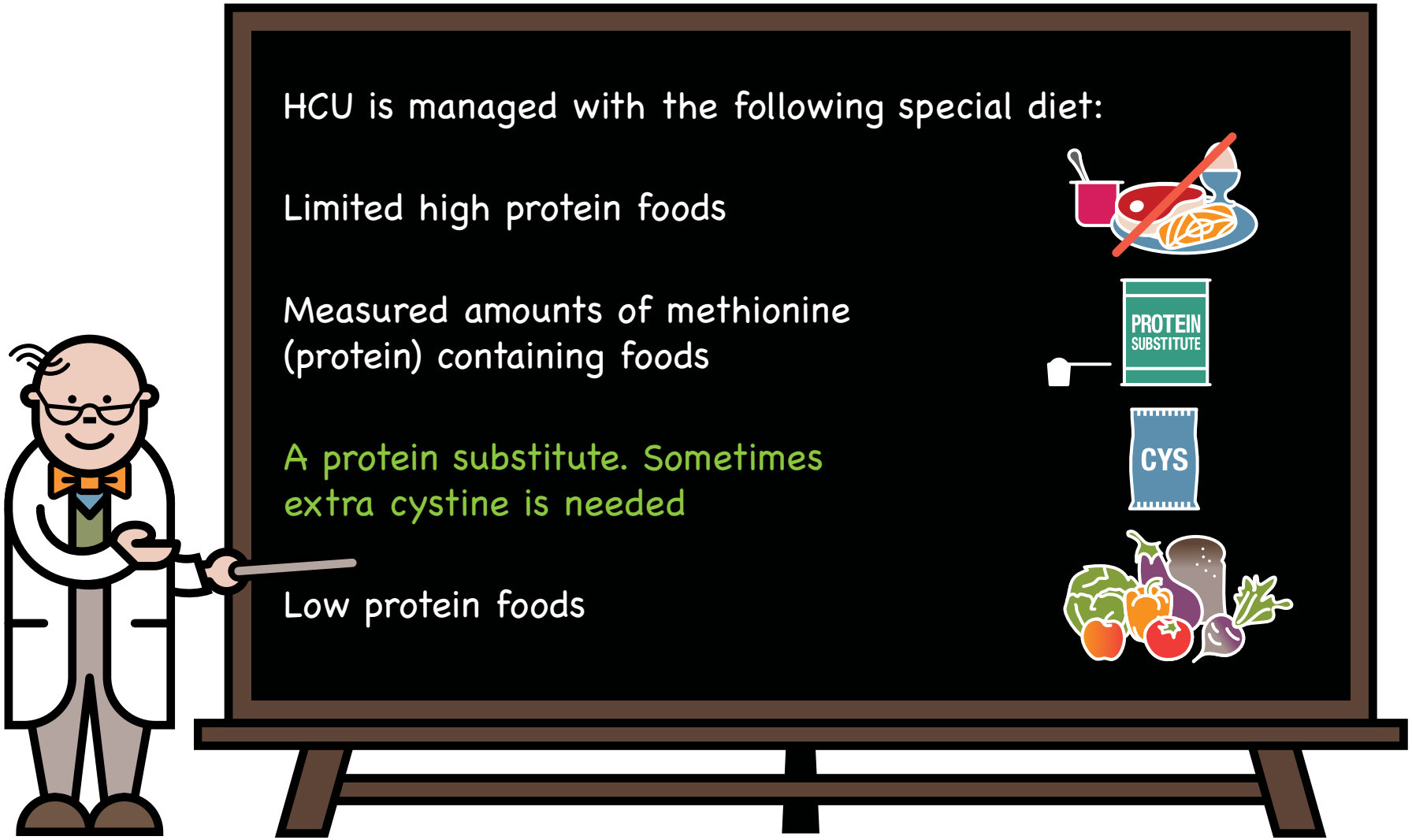

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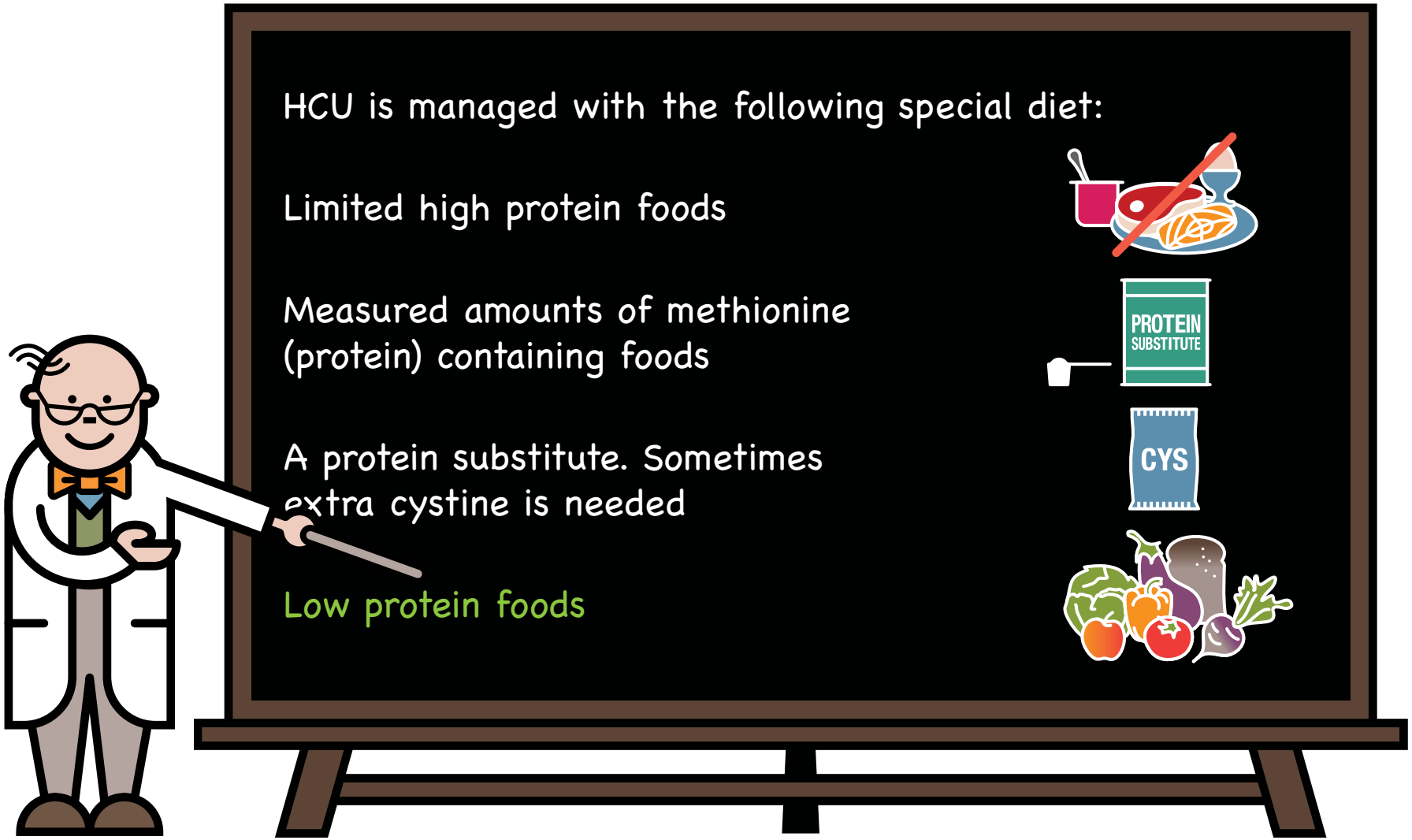

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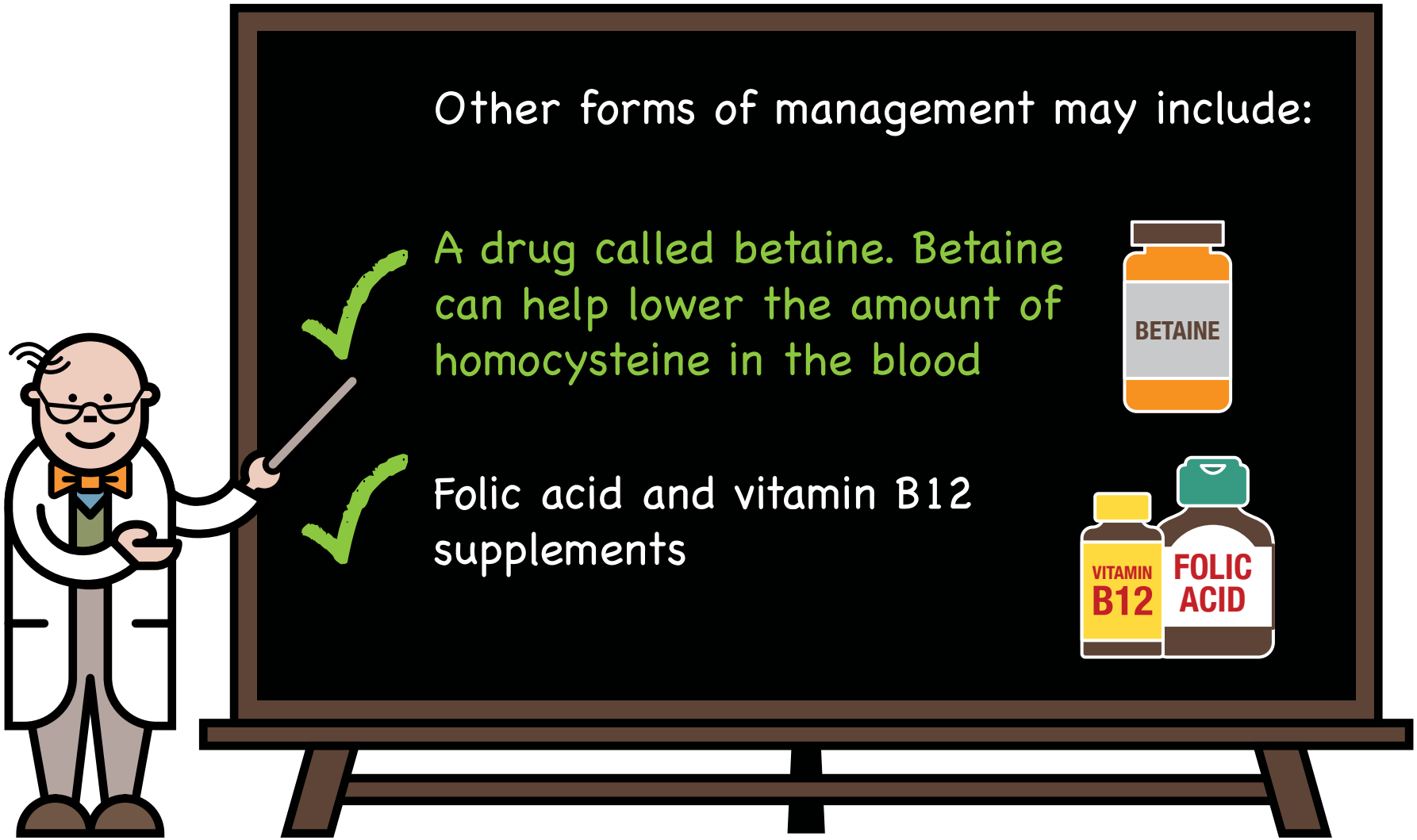

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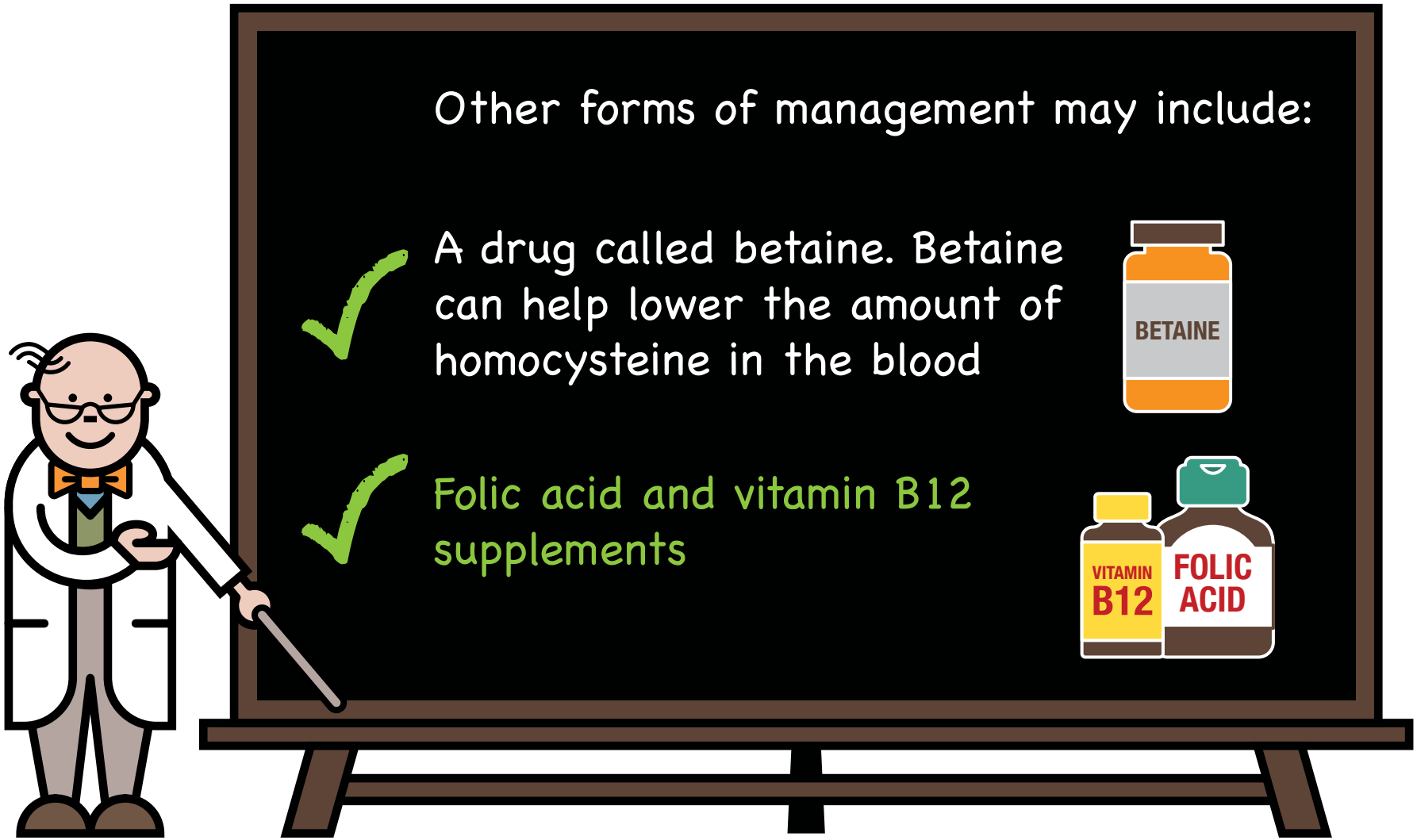


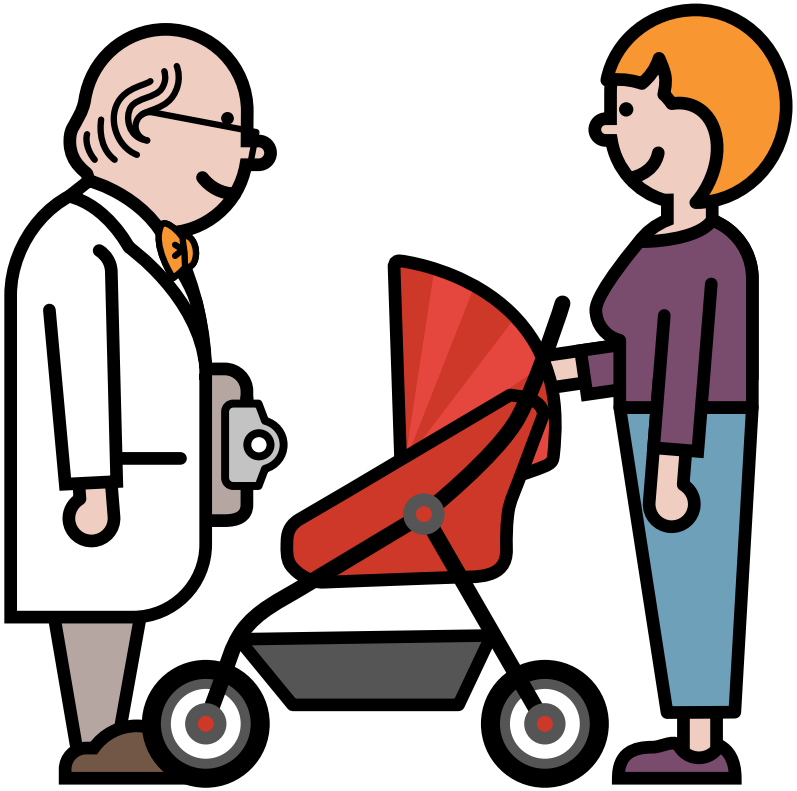
High protein foods

These foods are high in methionine (protein) and must be avoided: **meat, fish, eggs, cheese, milk, bread, pasta, nuts, seeds, soya, Quorn** and **tofu.**

Measured methionine intake

In babies, a restricted amount of methionine (protein) is given from breast milk or measured amounts of infant formula.

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

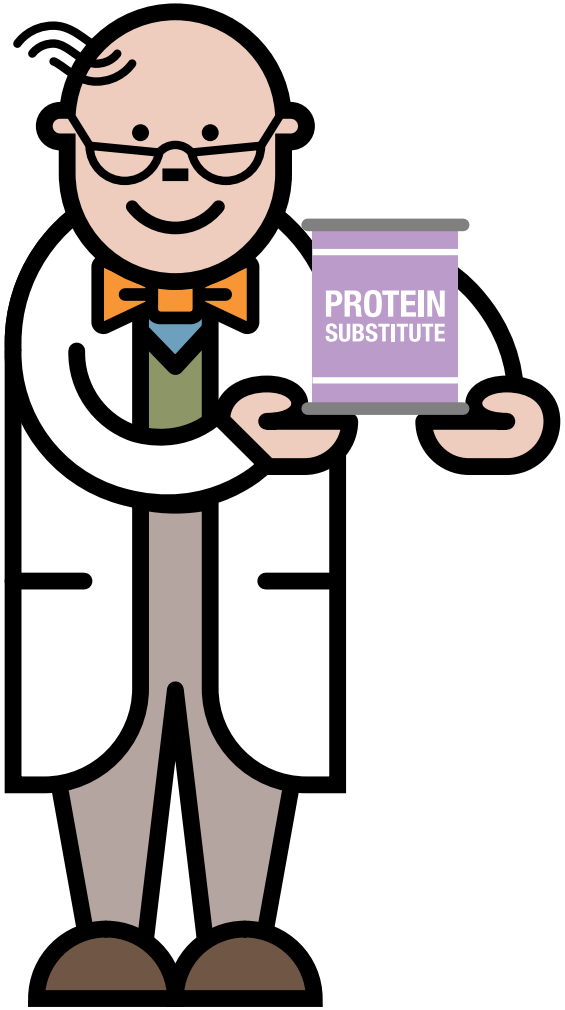


Protein substitute

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.

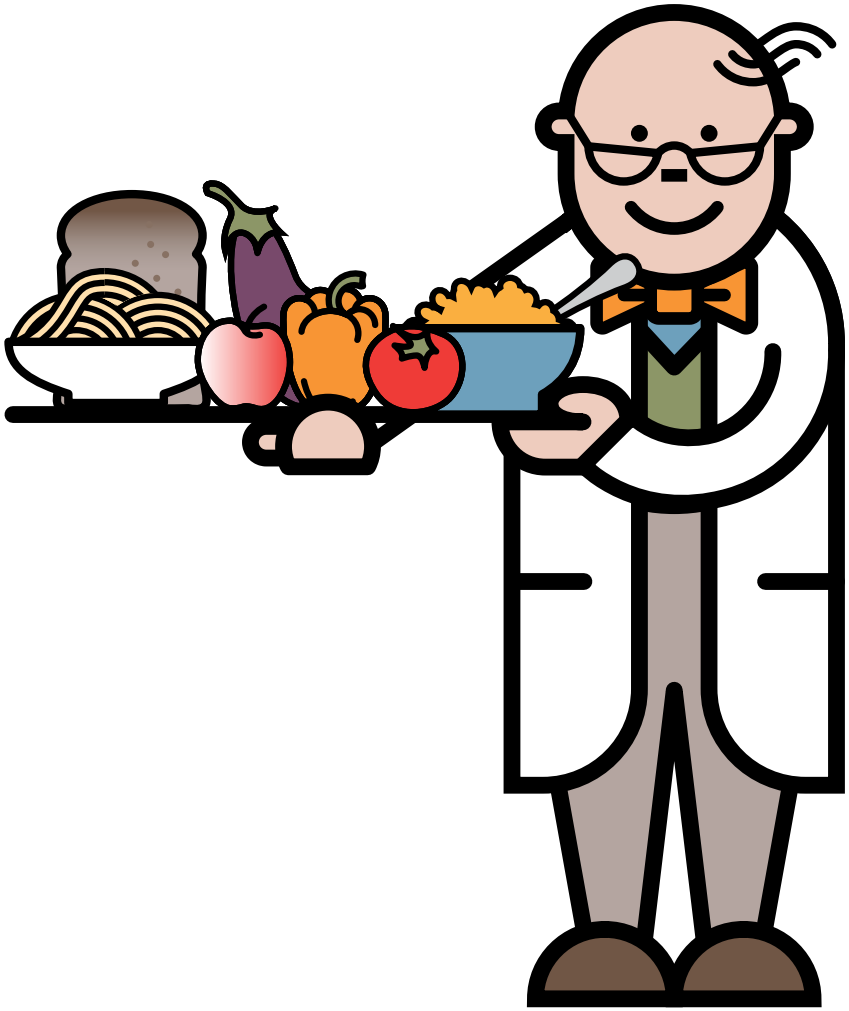


Low protein foods

There are many low protein foods. These include fruit, many vegetables and prescribable low protein foods such as bread and pasta.

They provide:

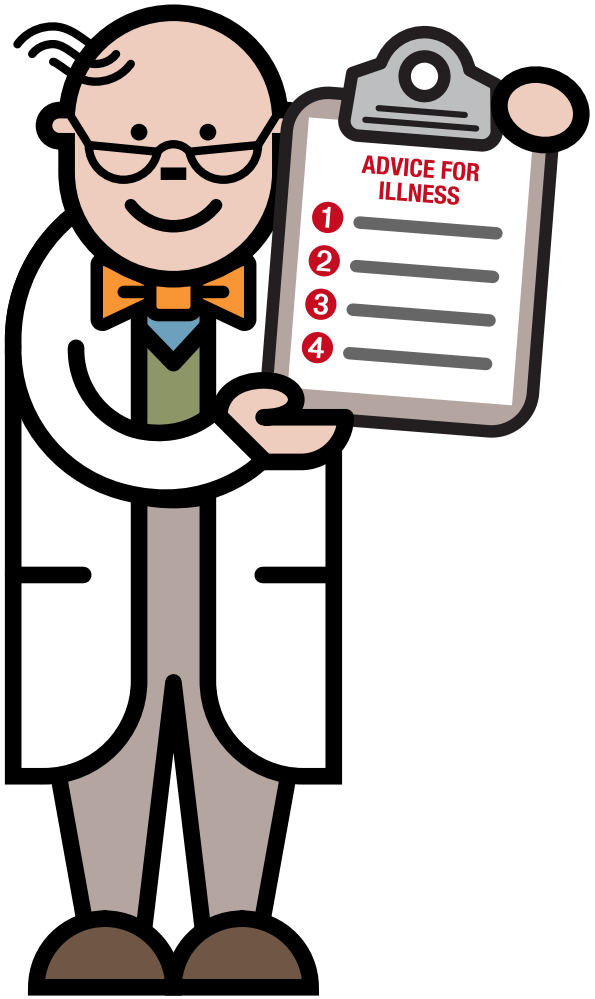
- a source of energy
- variety in the diet



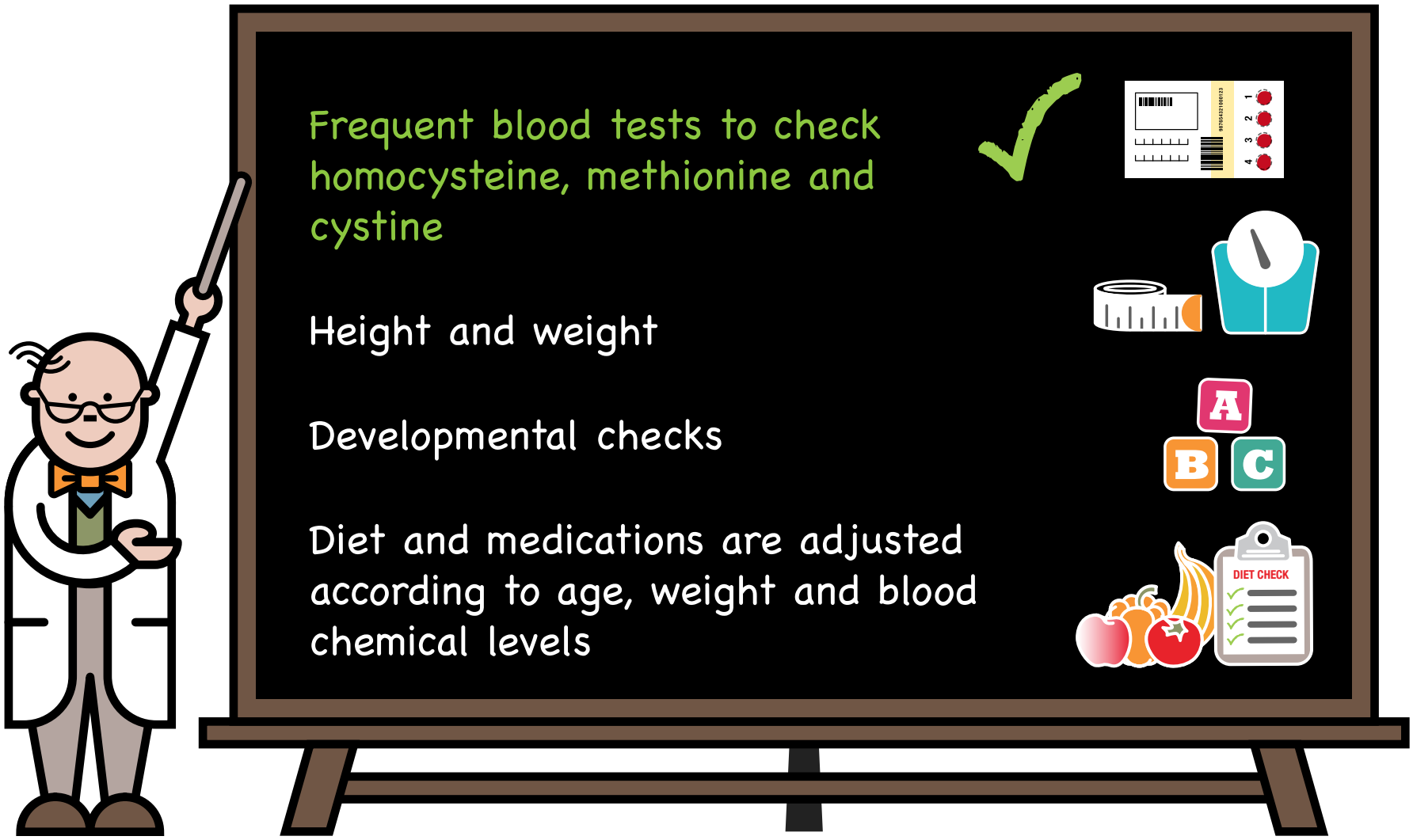
How is HCU managed during illness?

During any childhood illness, catabolism or protein breakdown occurs, causing blood homocysteine levels to increase.

It is important to continue with the usual diet as much as possible.



How is HCU monitored?



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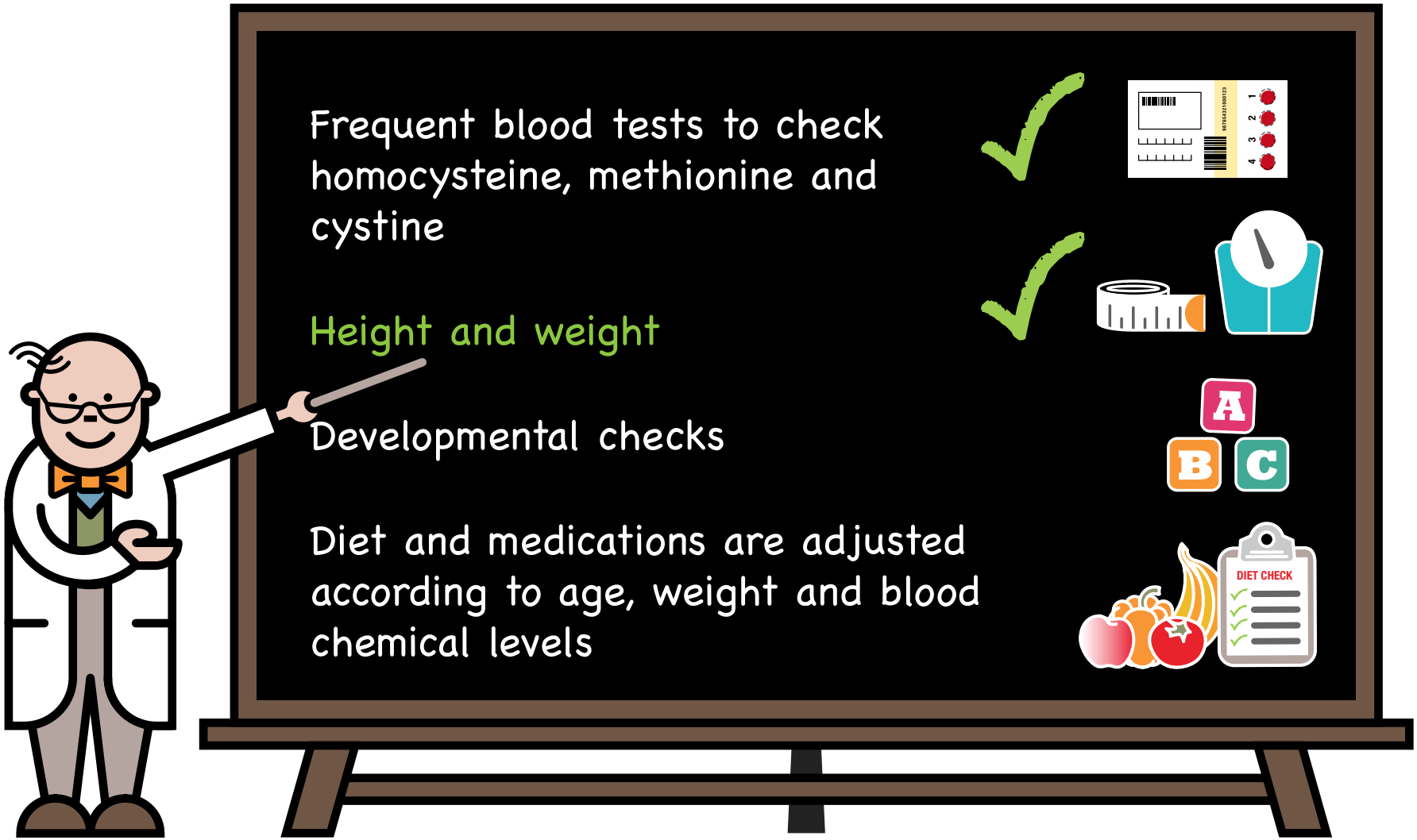

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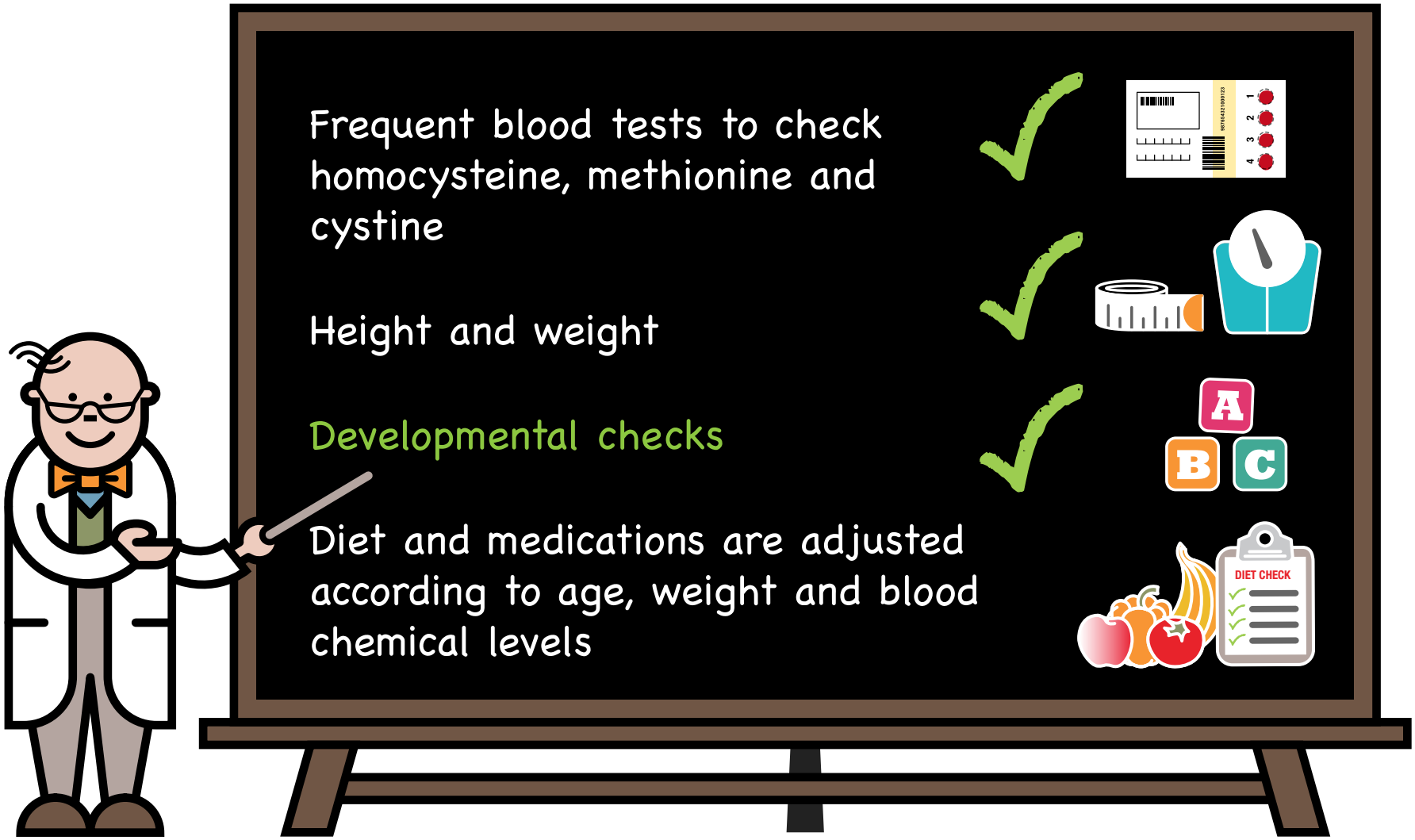

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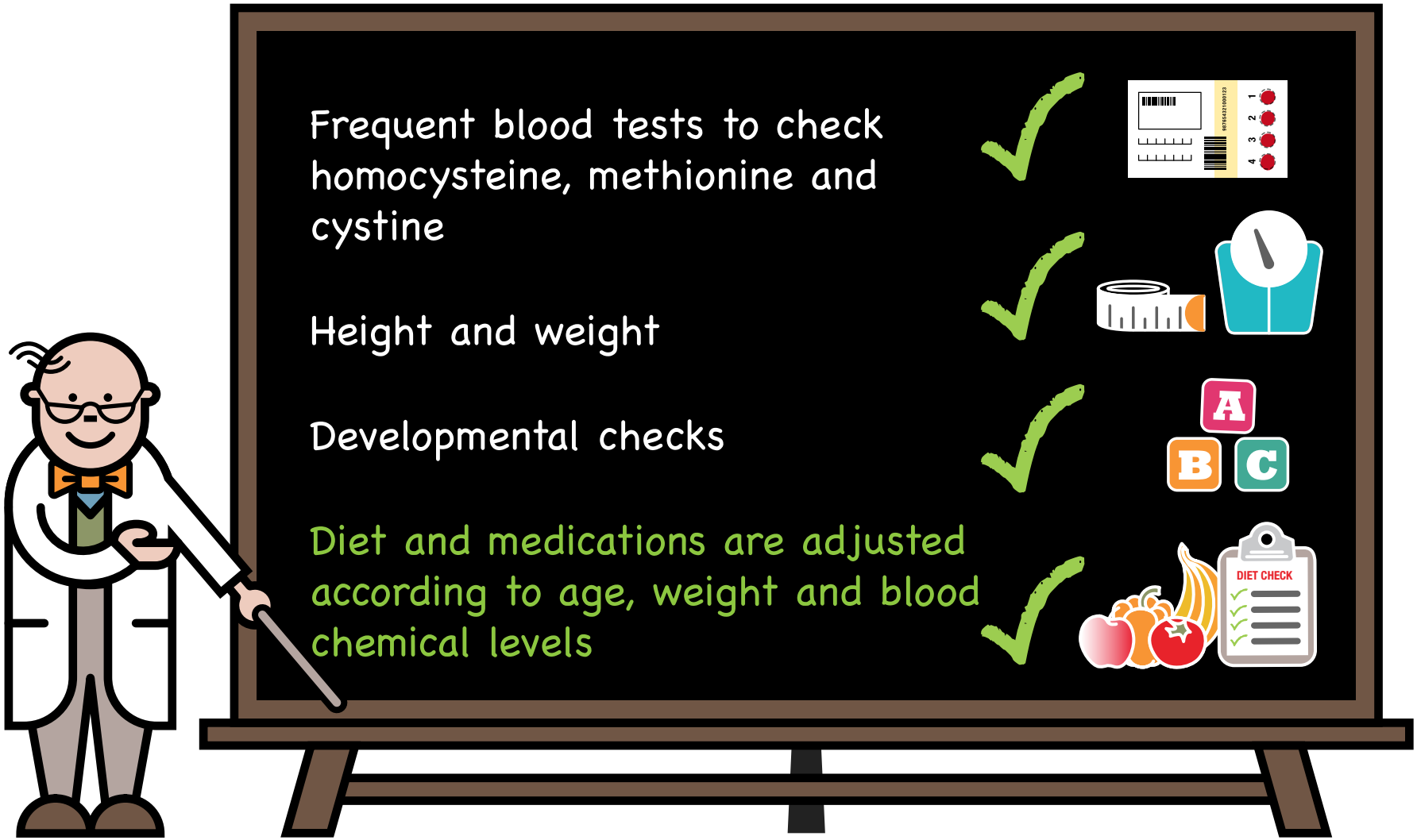
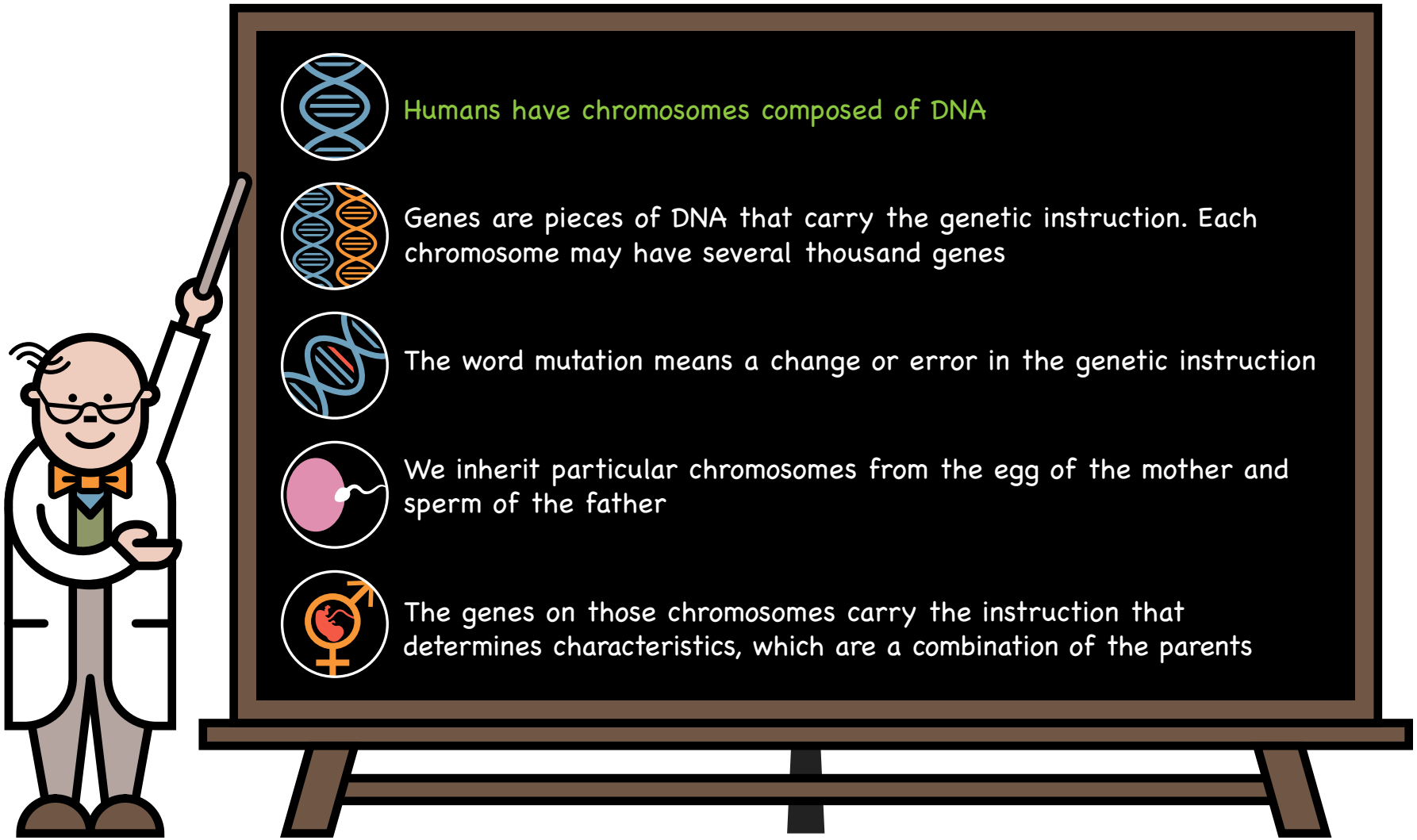
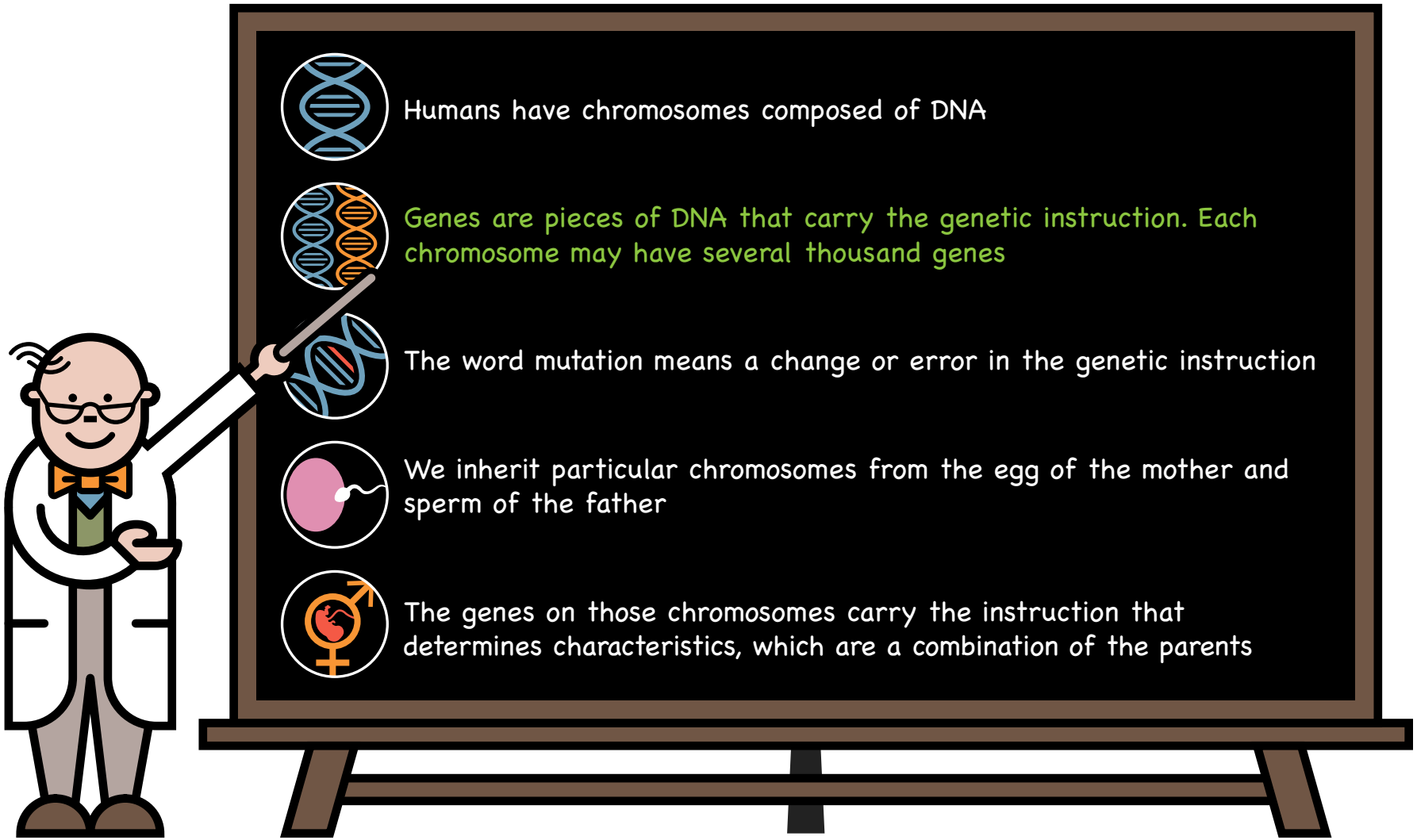


Chromosomes, genes, mutations



Chromosomes, genes, mutations



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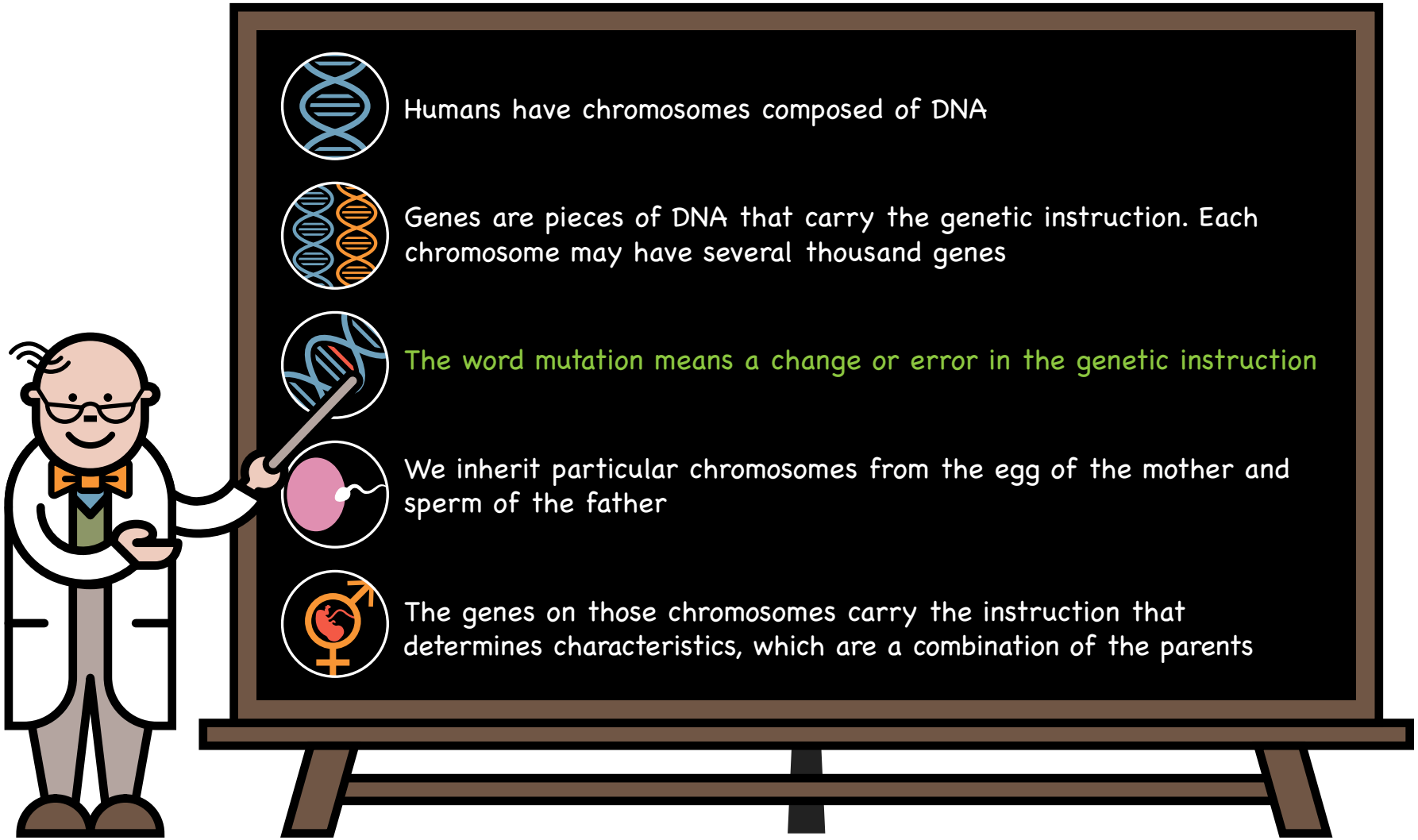

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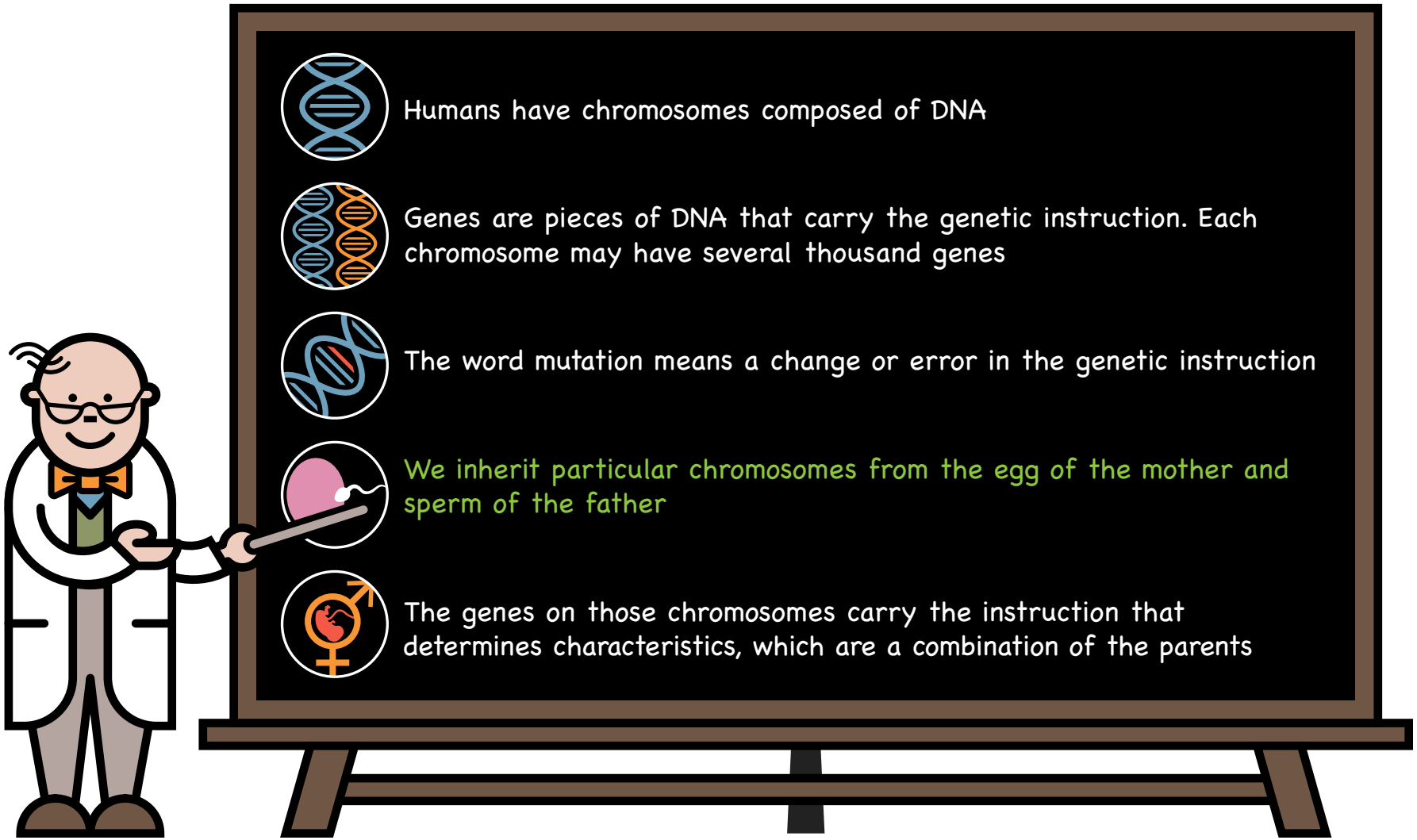

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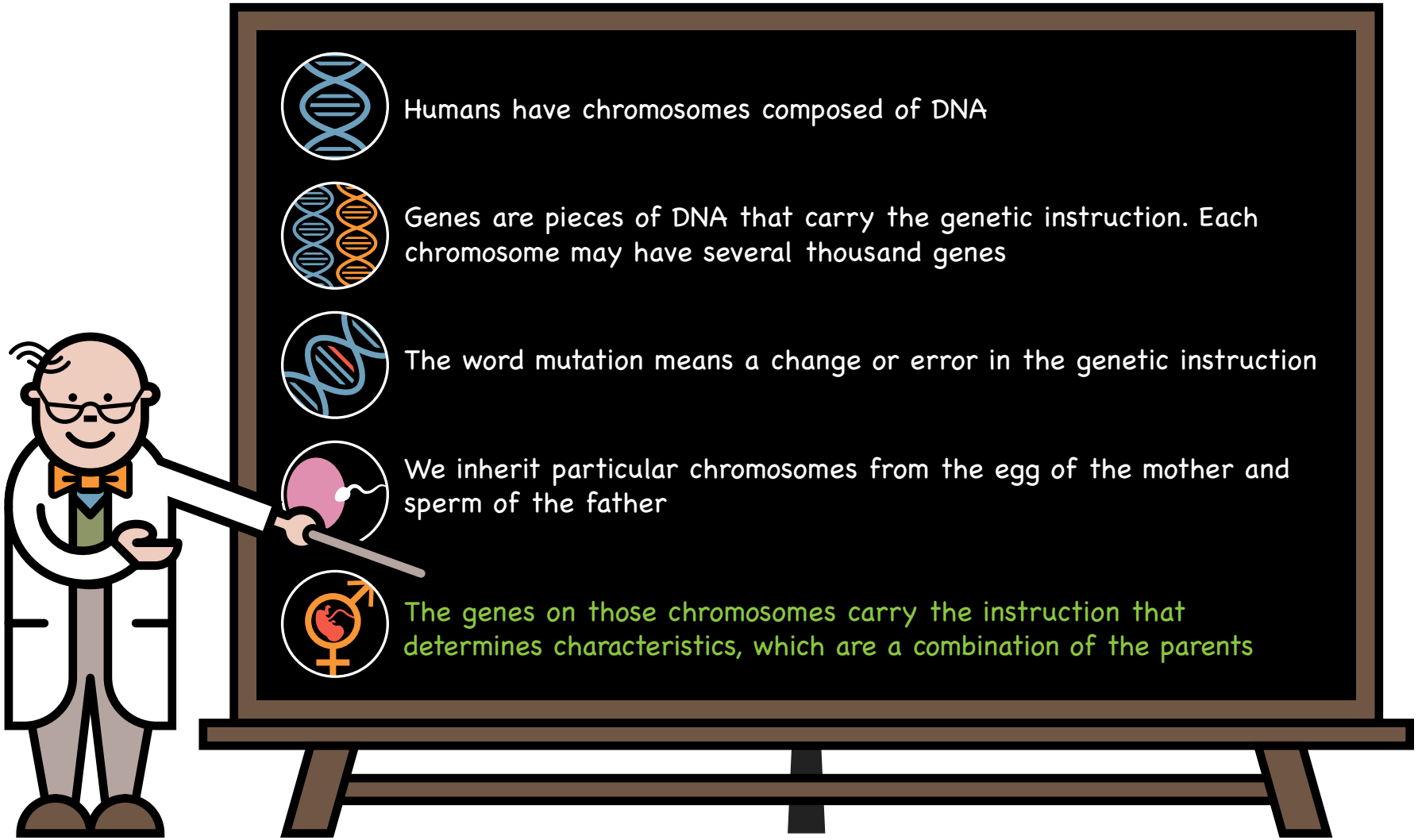
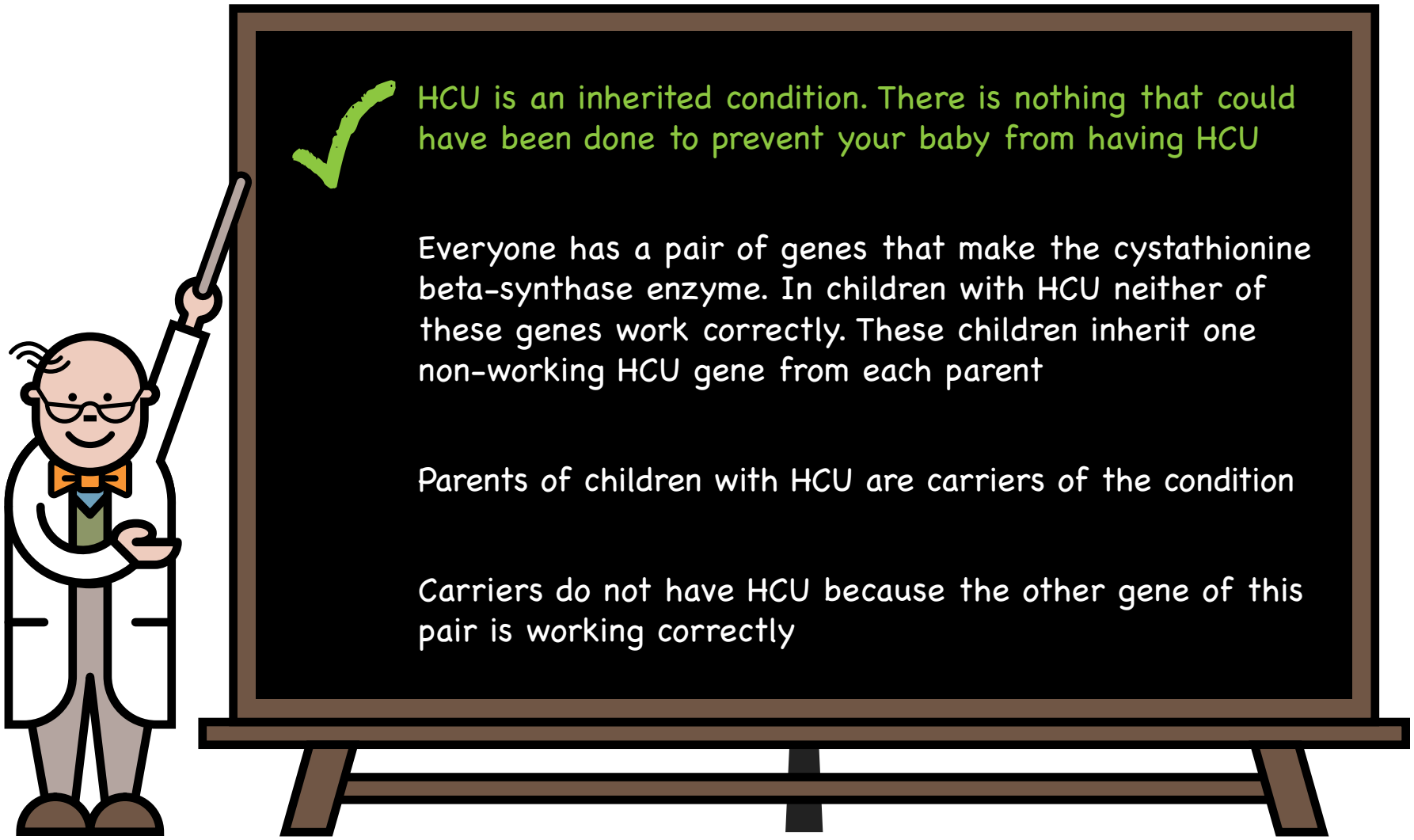


Inheritance



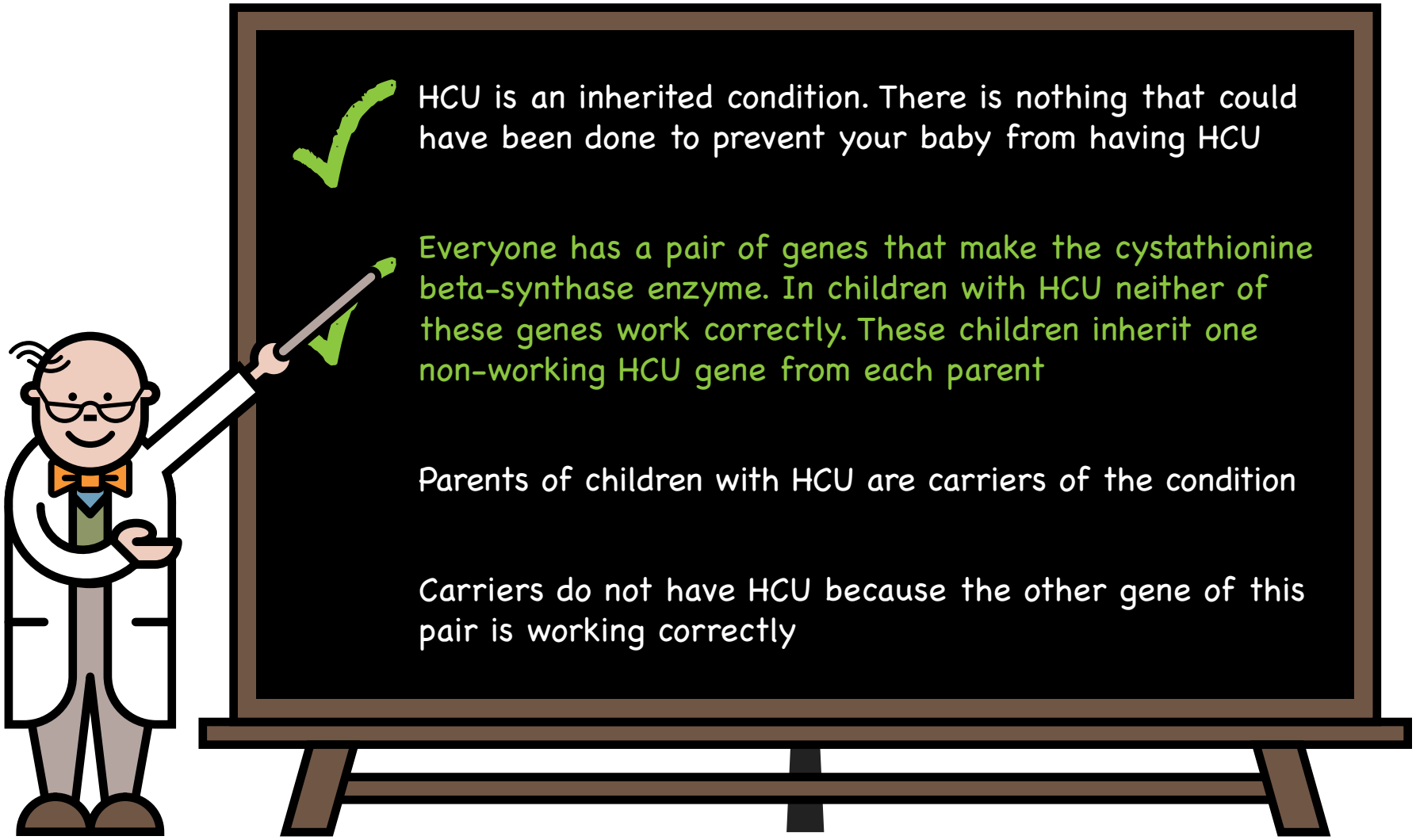
HCU is an inherited condition. There is nothing that could have been done to prevent your baby from having HCU

Everyone has a pair of genes that make the cystathionine beta-synthase enzyme. In children with HCU neither of these genes work correctly. These children inherit one non-working HCU gene from each parent

Parents of children with HCU are carriers of the condition

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

Inheritance



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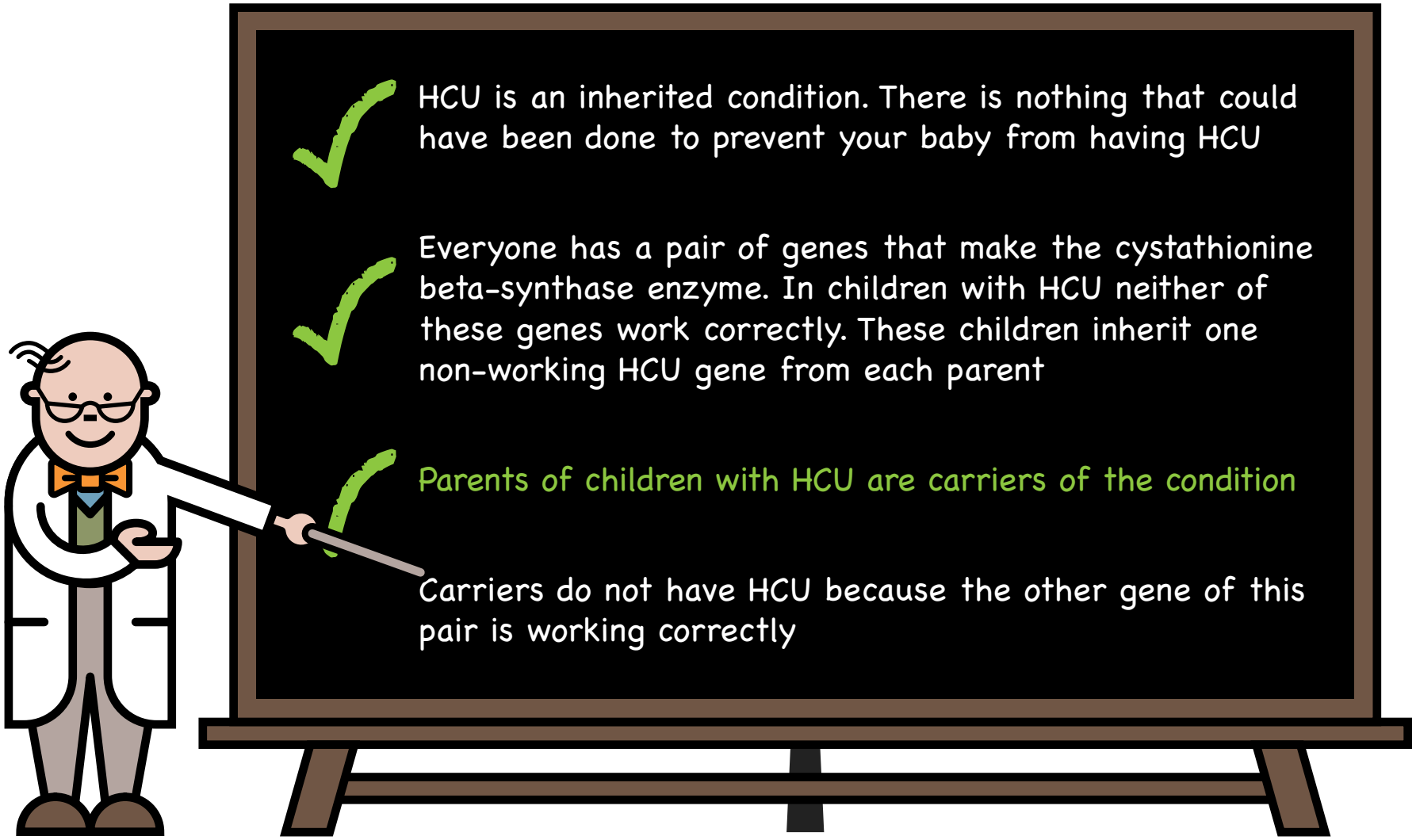

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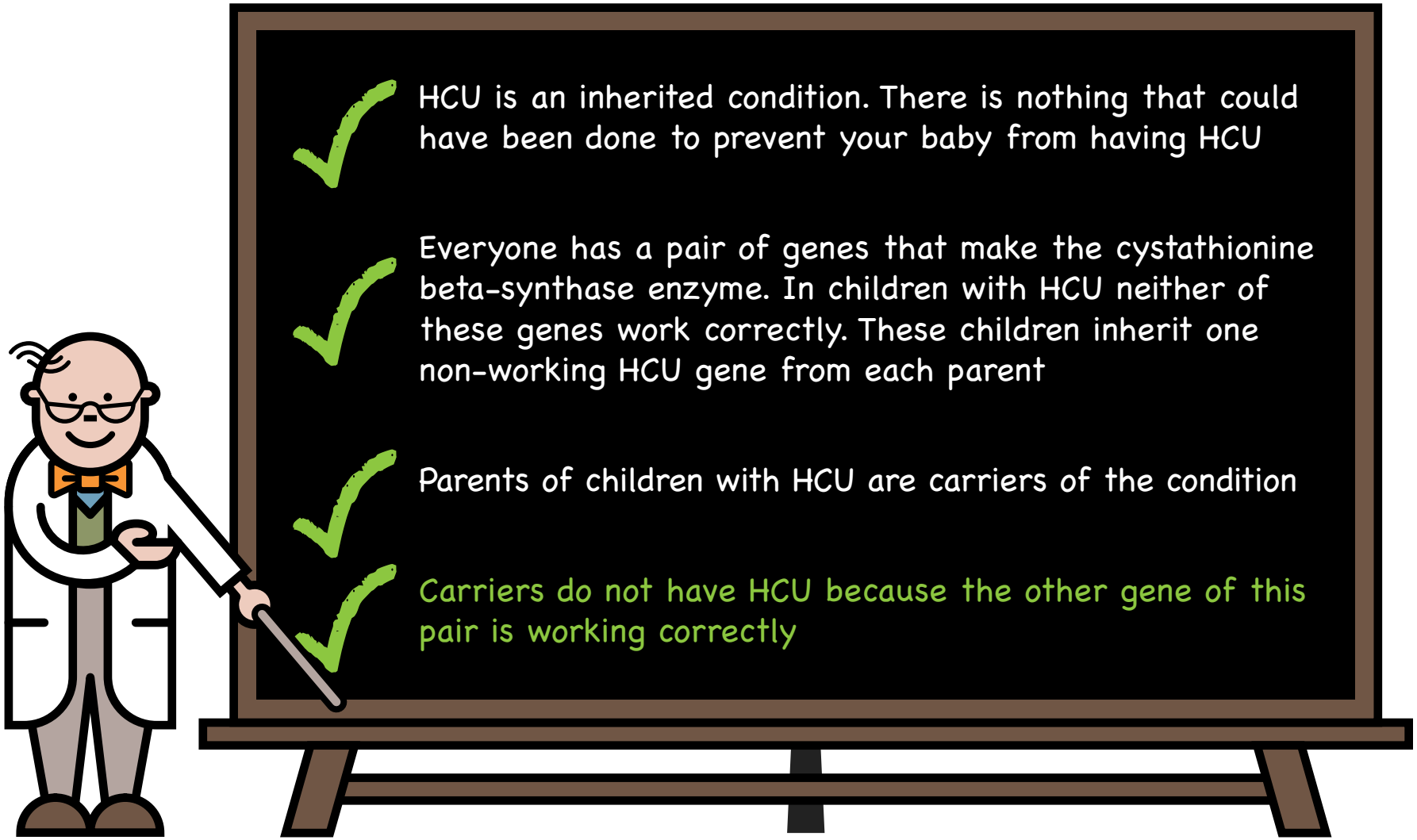
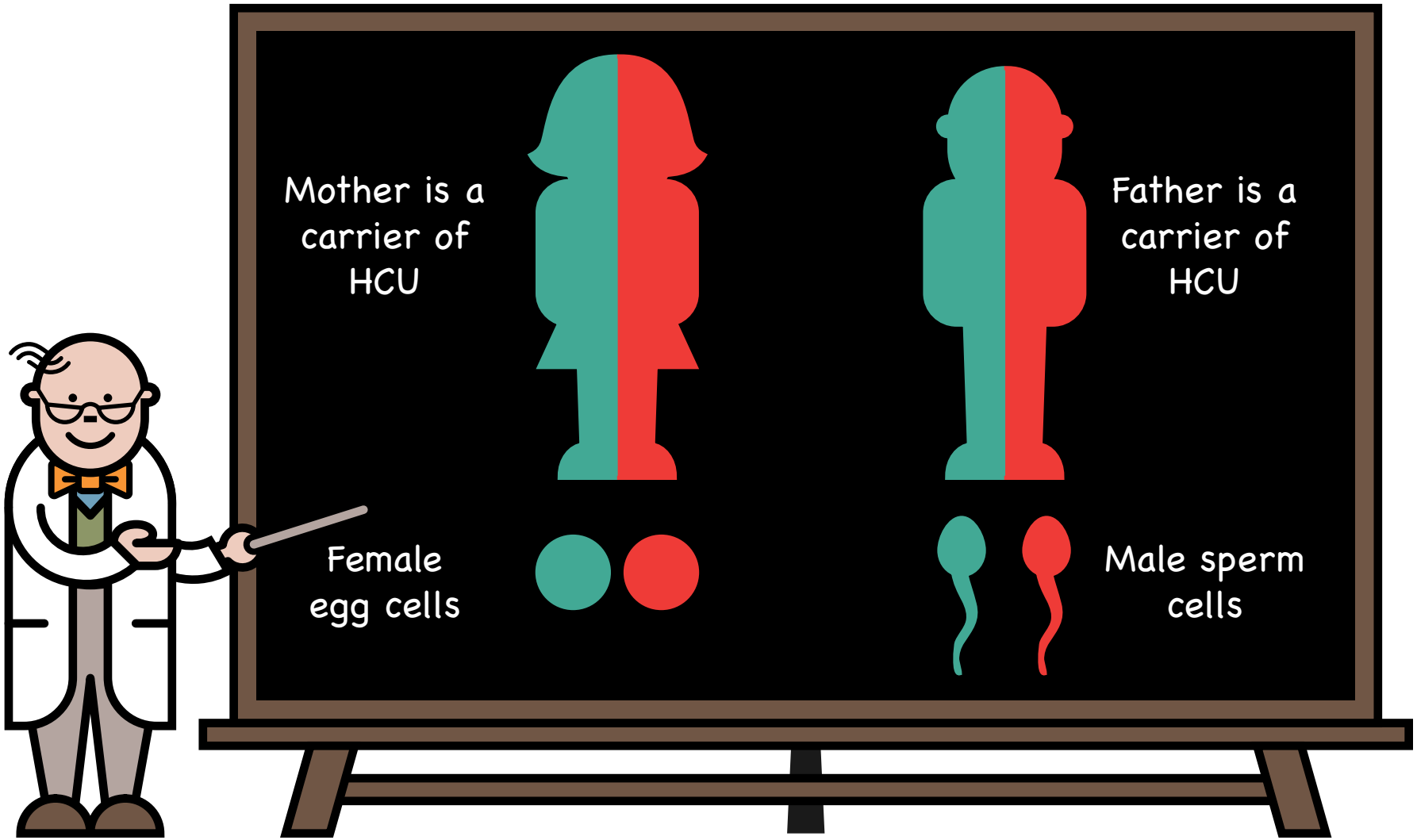
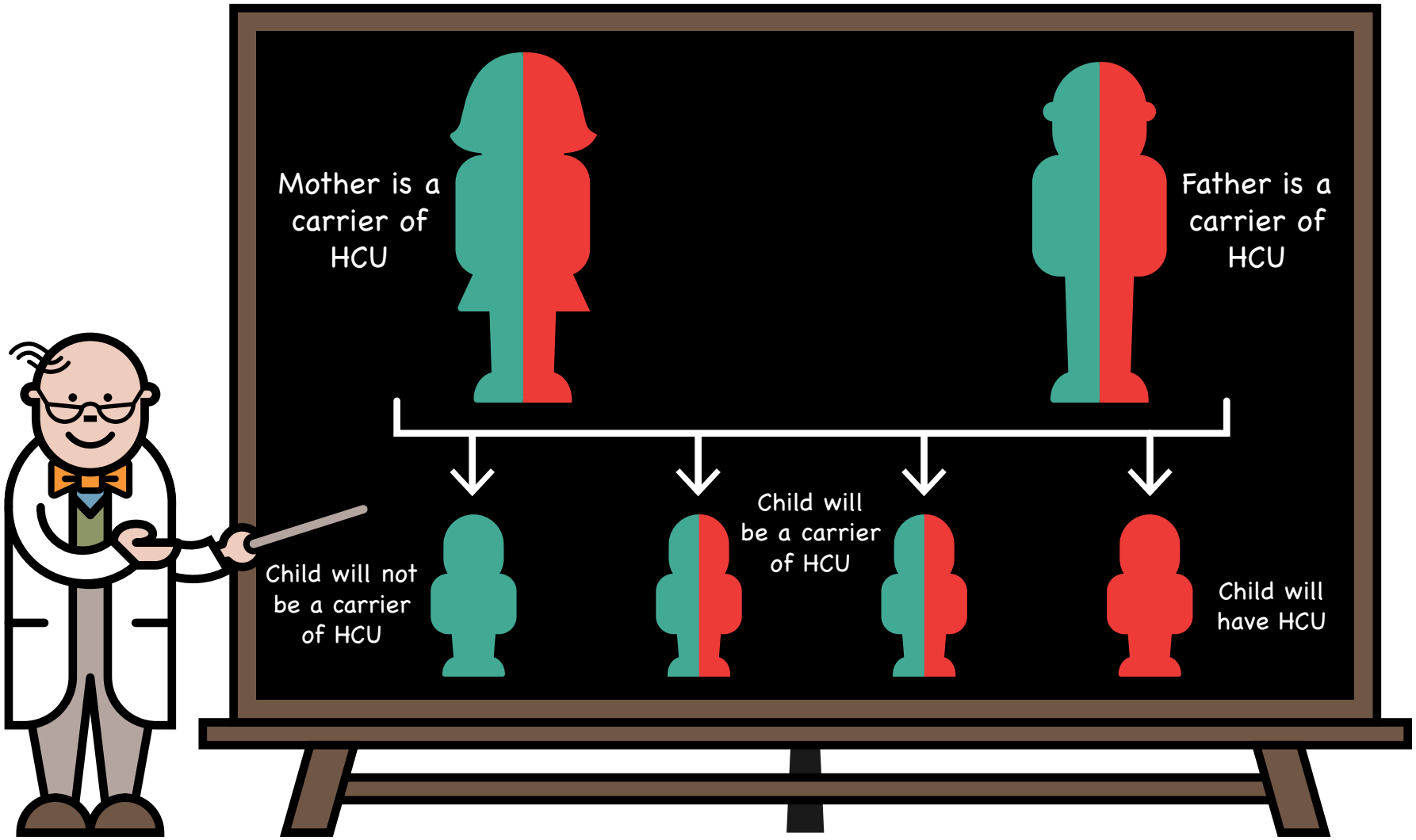


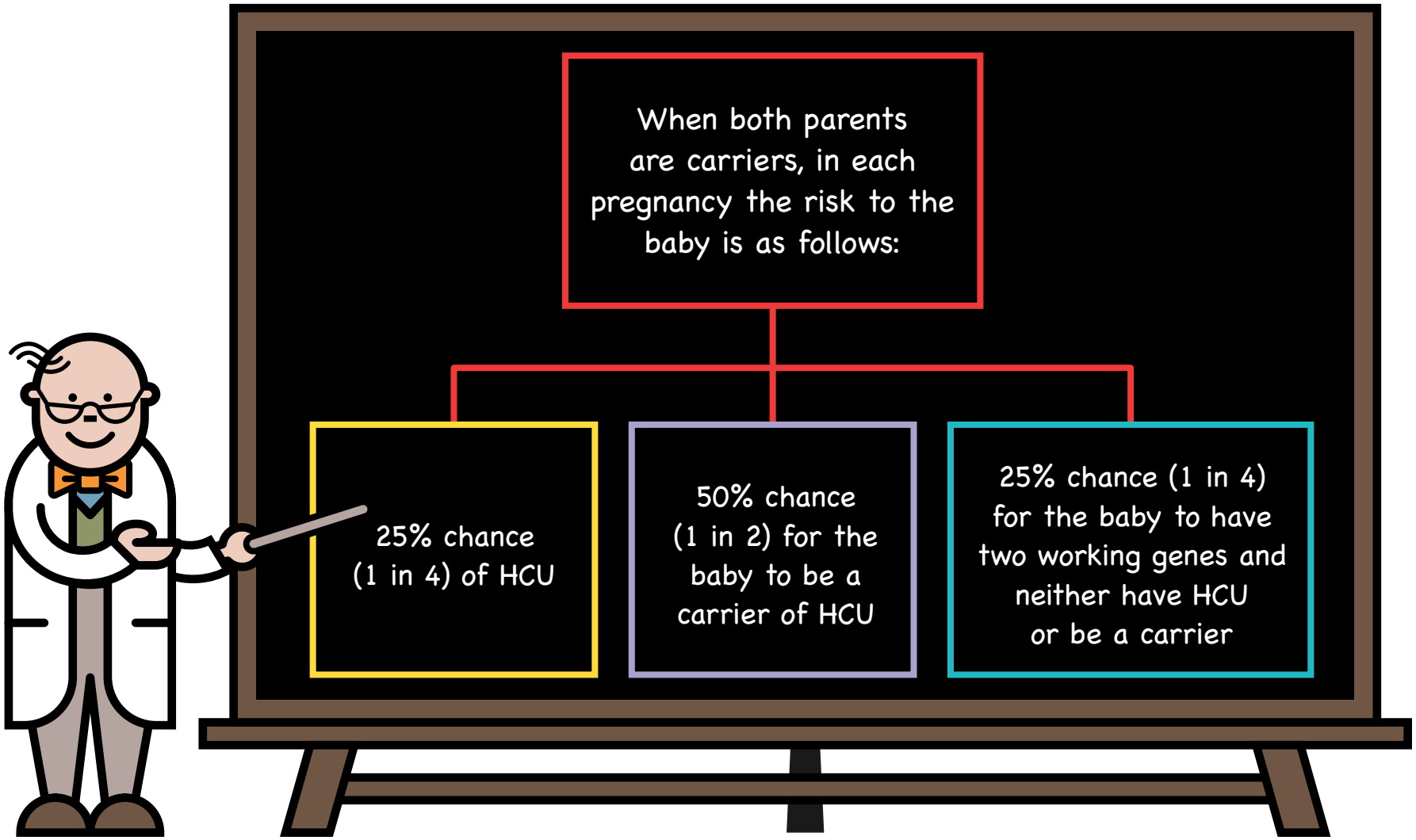
Inheritance – Autosomal recessive (carriers of HCU)



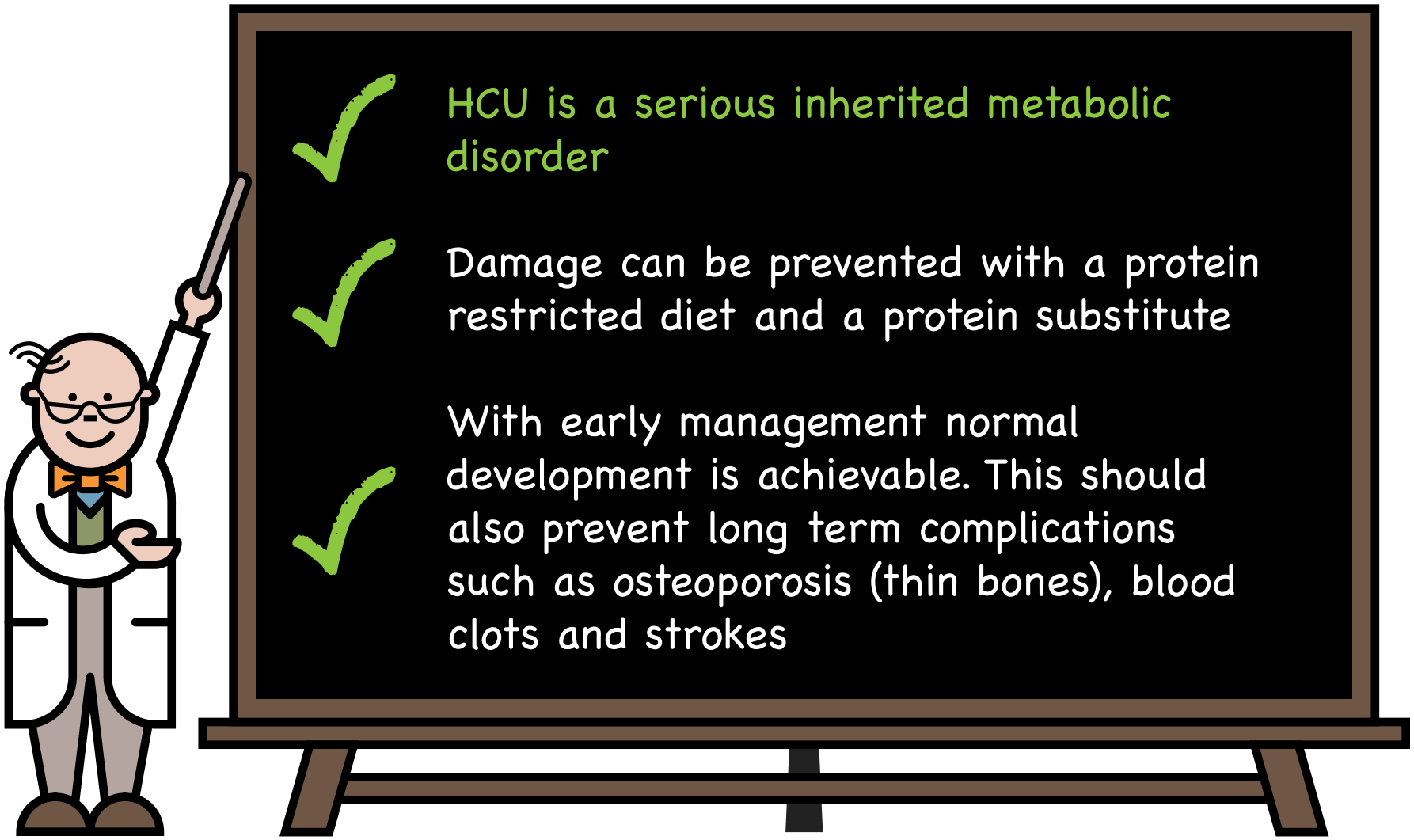
Inheritance – Autosomal recessive – possible combinations



Future pregnancies



Take home messages



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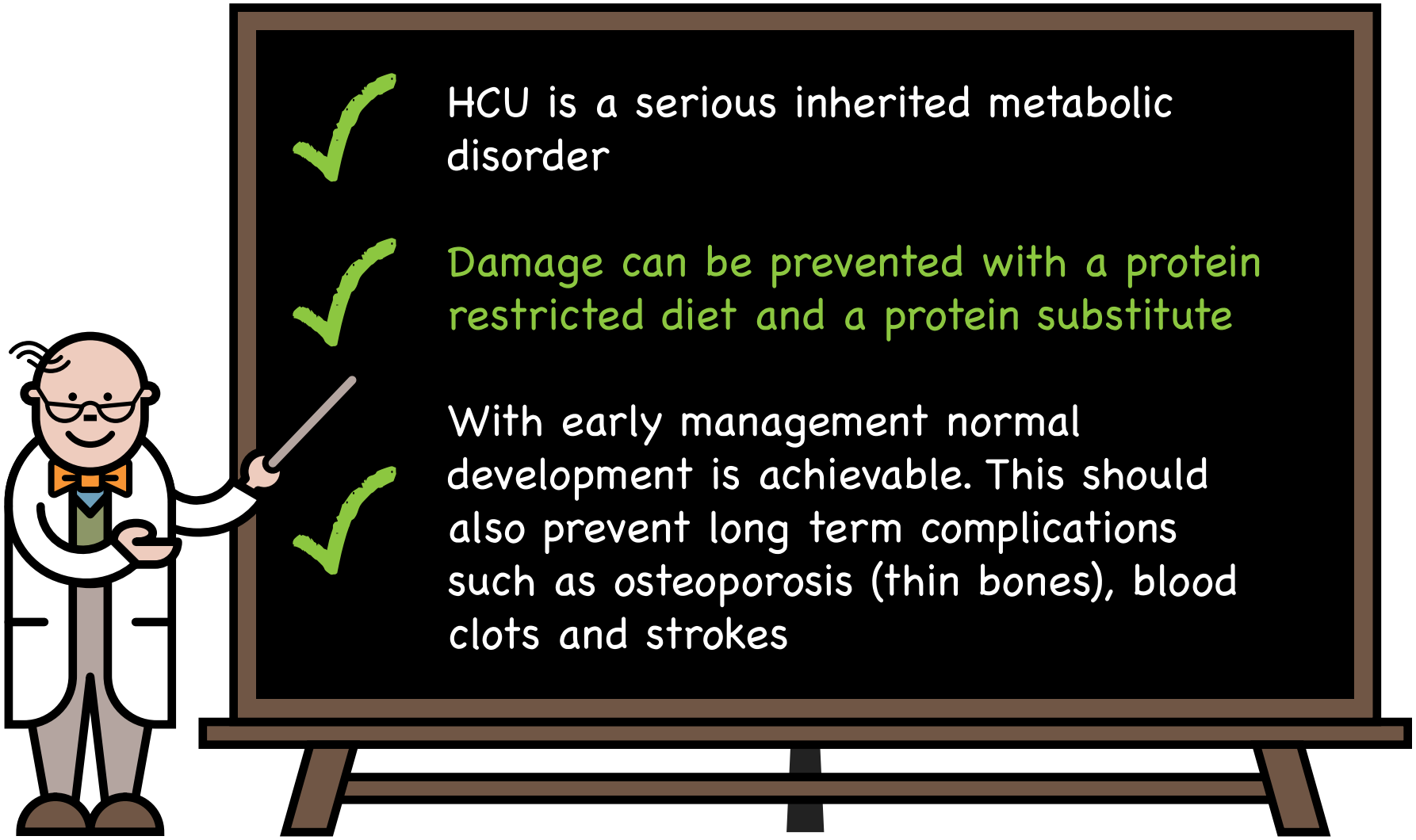

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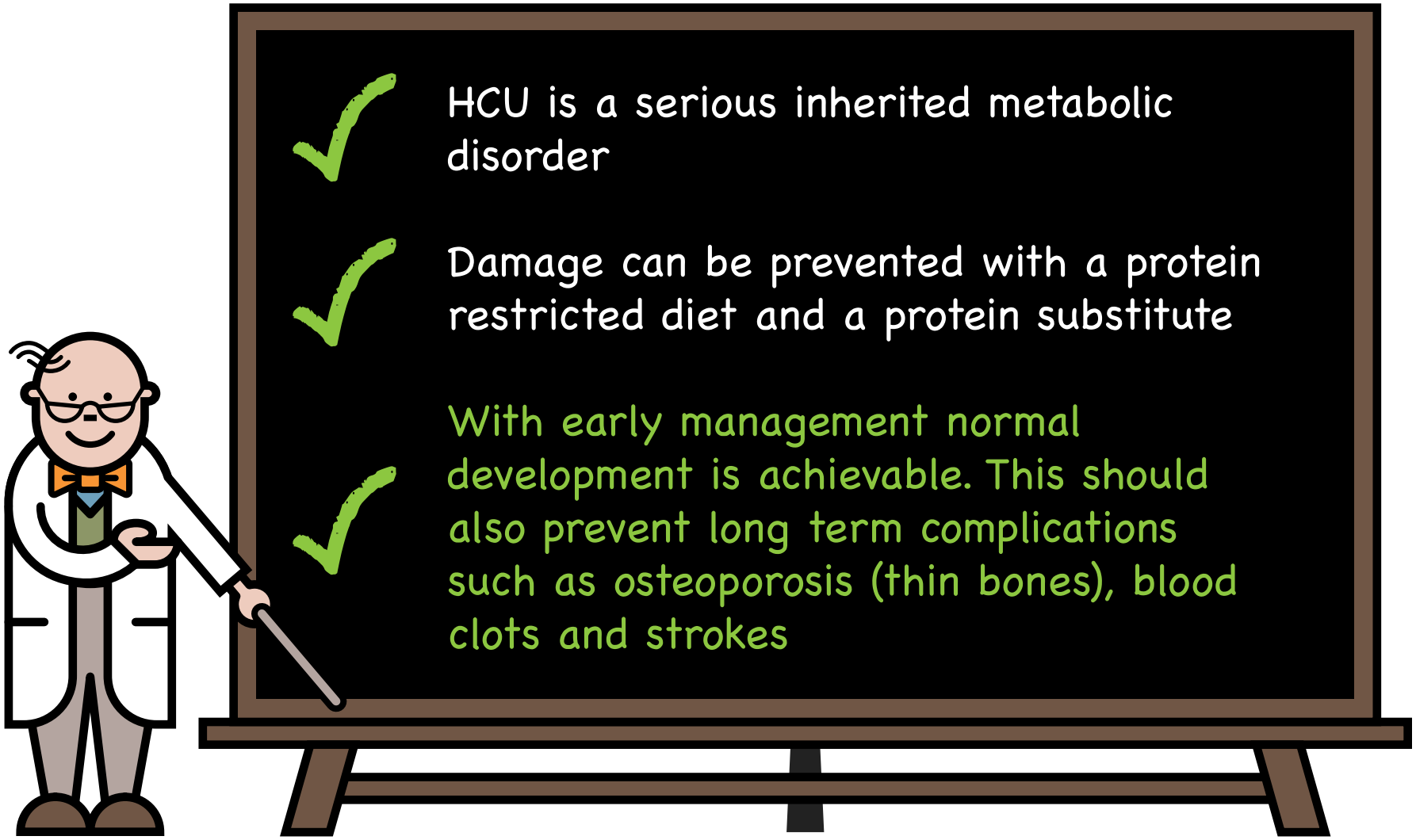

Helpful hints

Helpful hints

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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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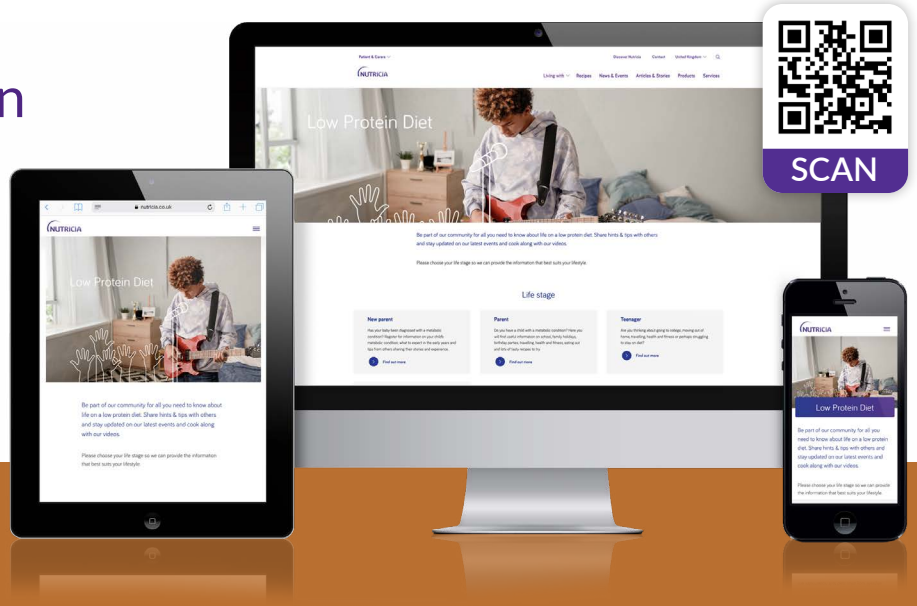
LowProteinConnect



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