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, APRIL 2020

PKU

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.

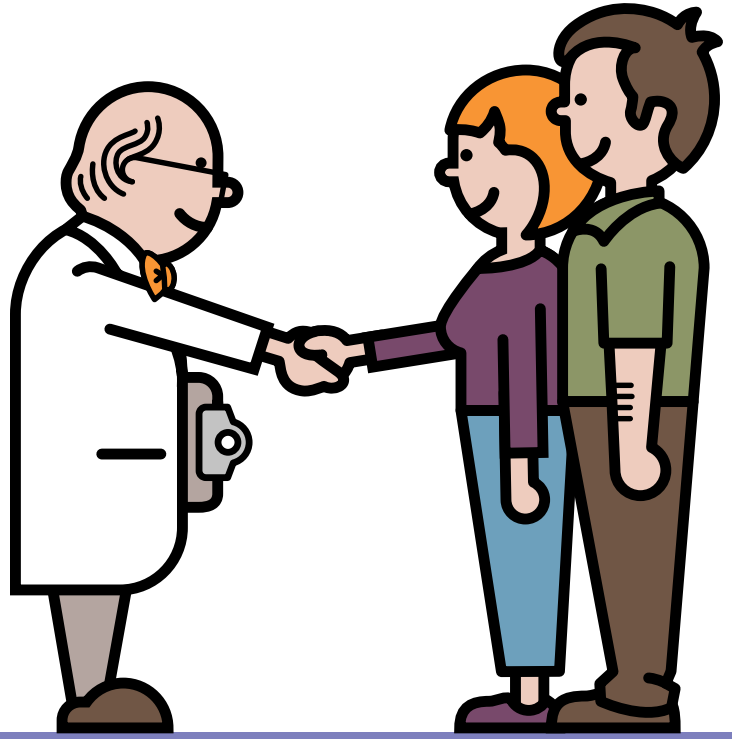
Each module produced is reviewed by a consultant clinician who is a member of the BIMDG.

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

PKU

Information for families following a new diagnosis



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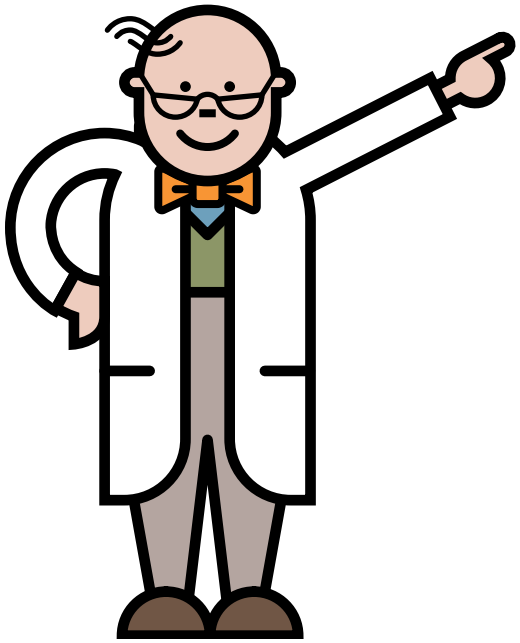
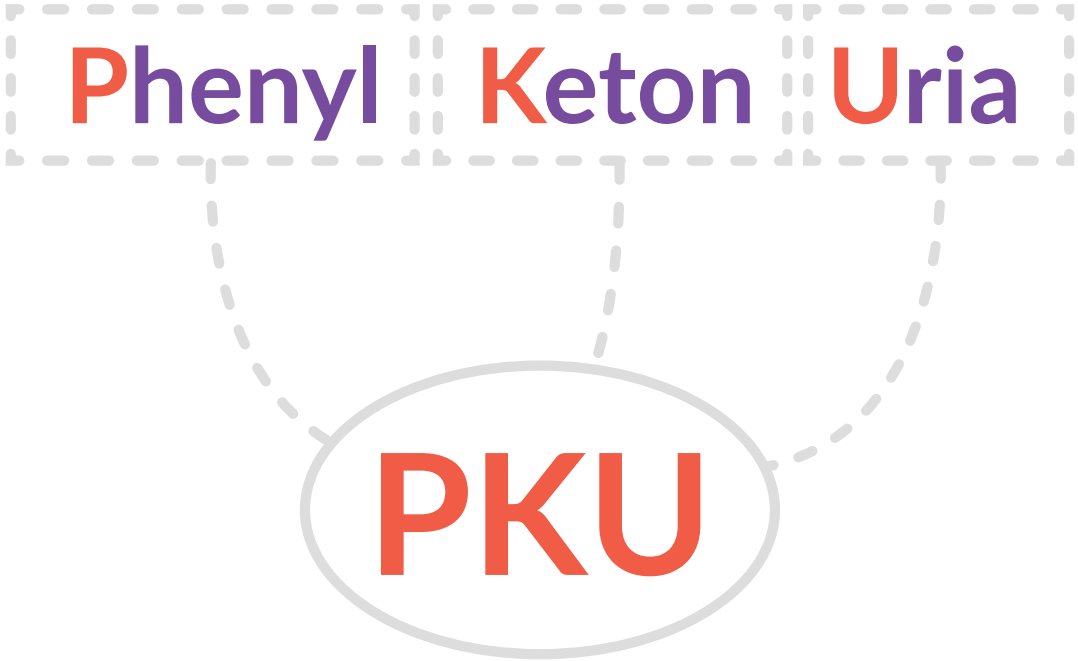
Tools Enabling Metabolic Parents LEarning

What is PKU?

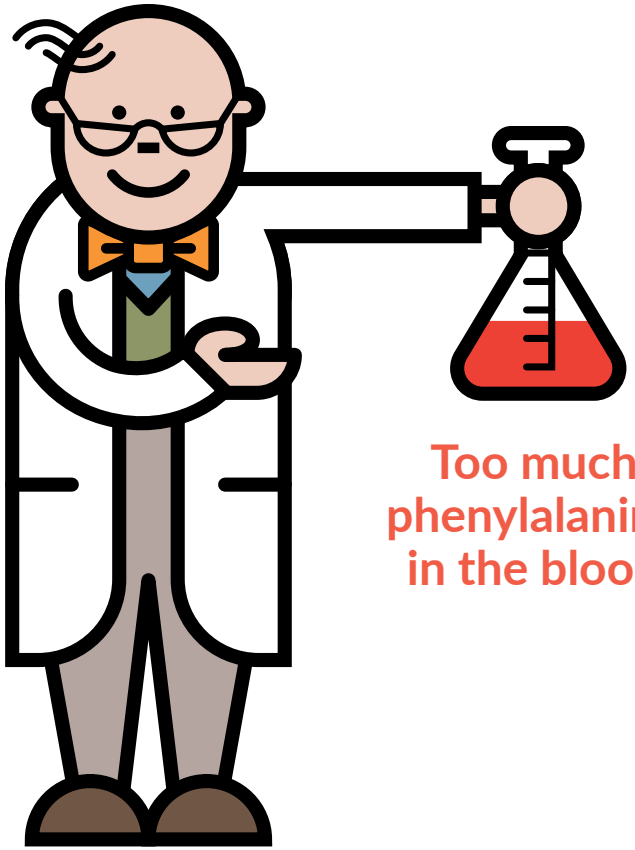
PKU stands for Phenylketonuria

It is an inherited metabolic condition

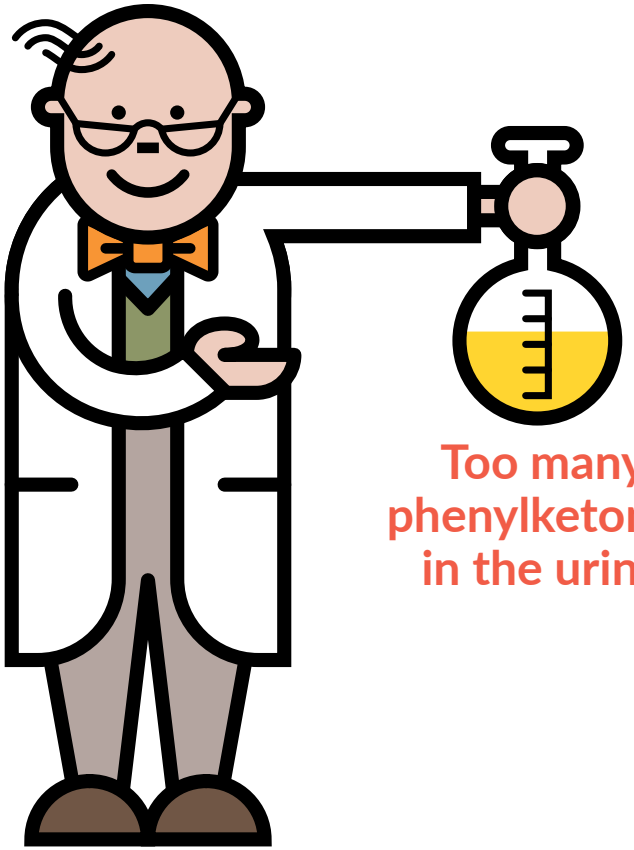




What is PKU?



Too much phenylalanine in the blood



Too many phenylketones in the urine

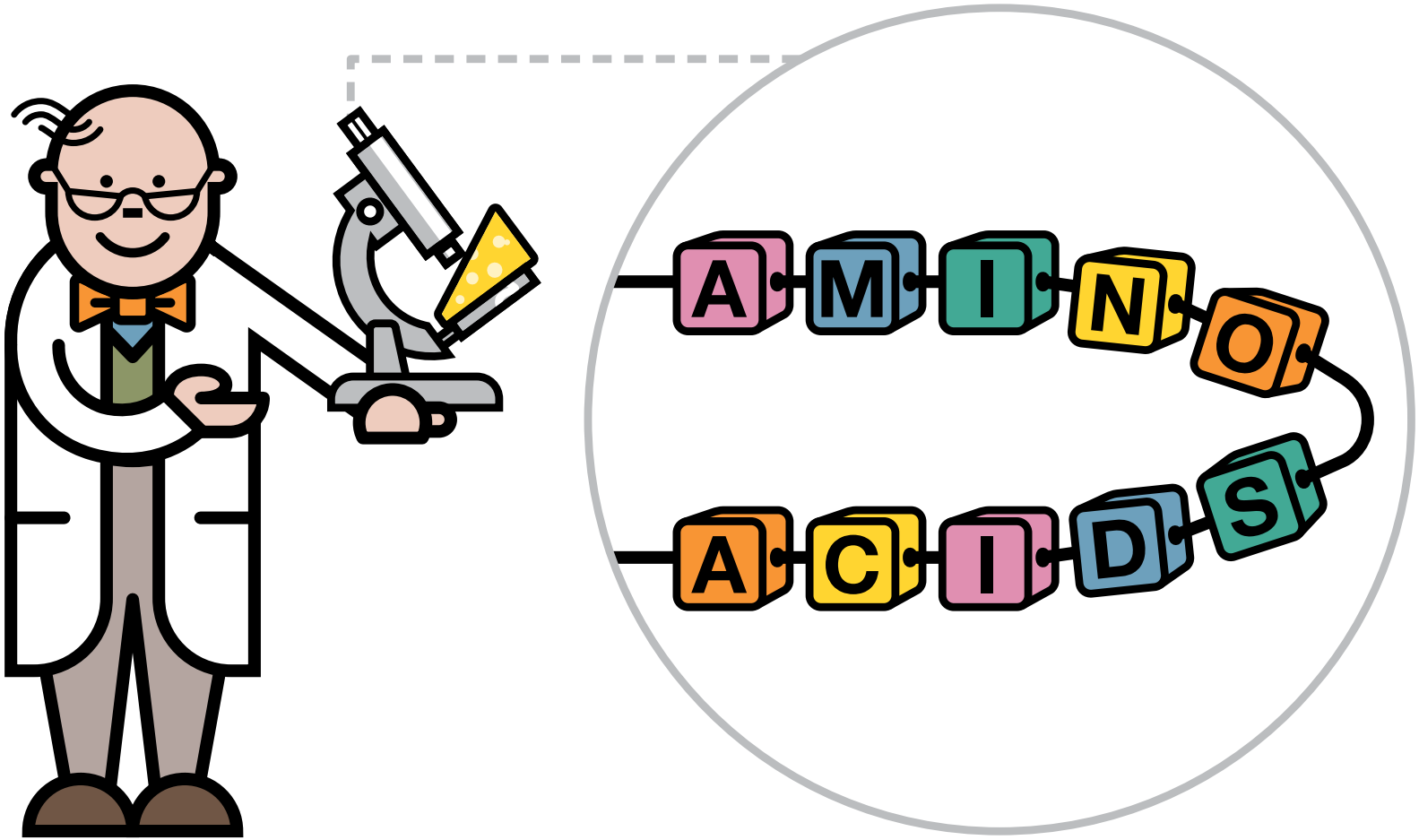
PKU and protein

PKU affects the way your baby breaks down protein.

Many foods contain protein.

The body needs protein for growth and repair.

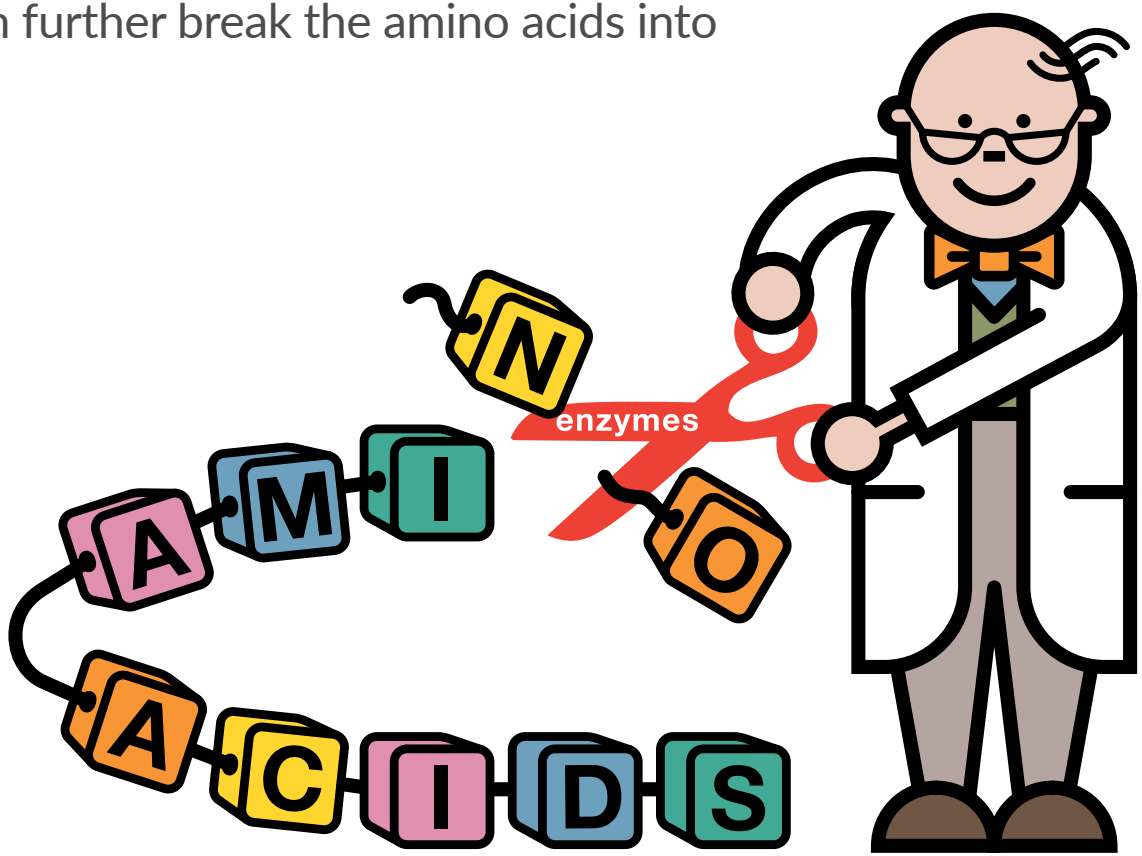
What is protein?



Protein and enzymes

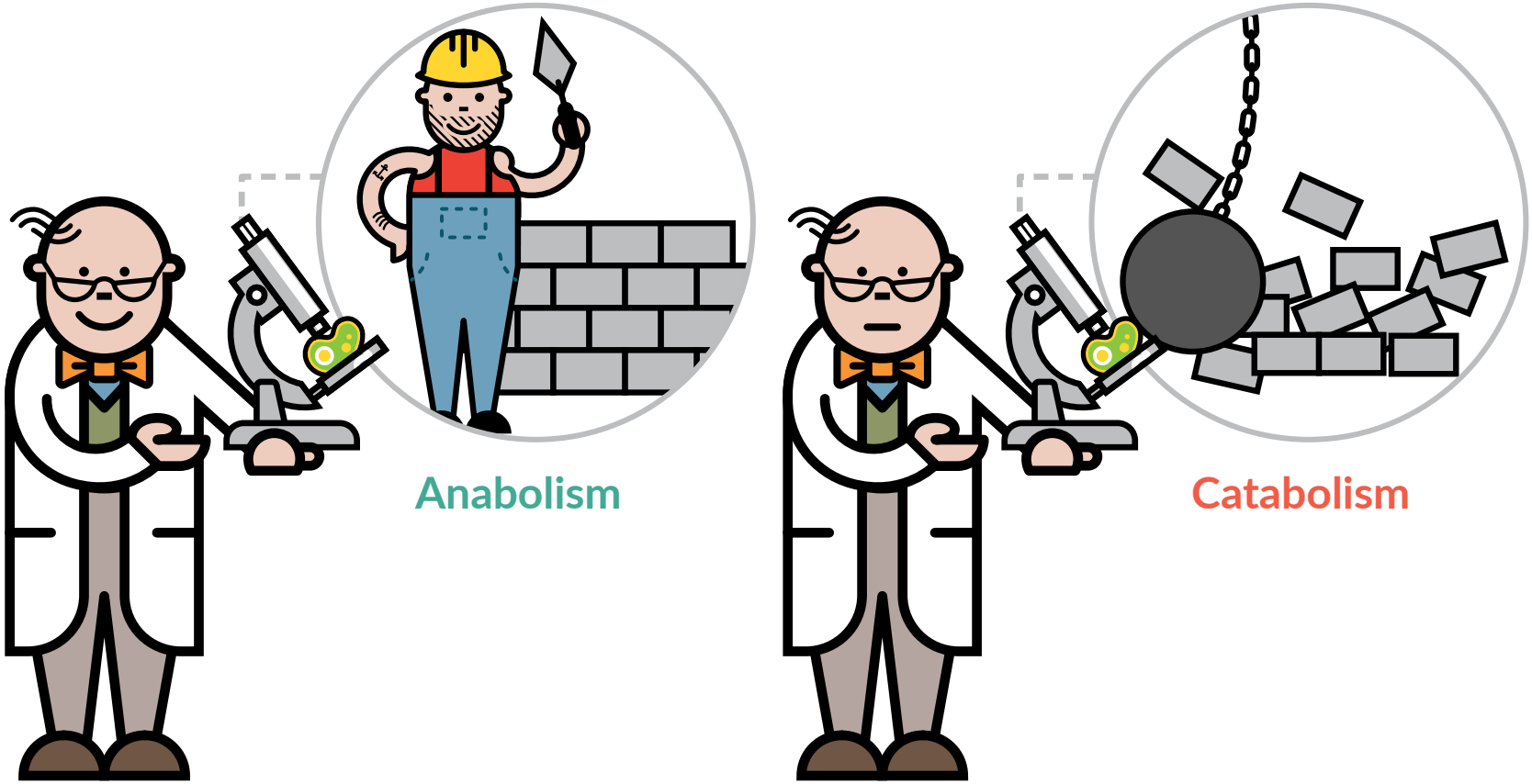
Protein is broken down into amino acids (building blocks of protein) by enzymes (which are like chemical scissors).

Enzymes then further break the amino acids into smaller parts.



Protein metabolism

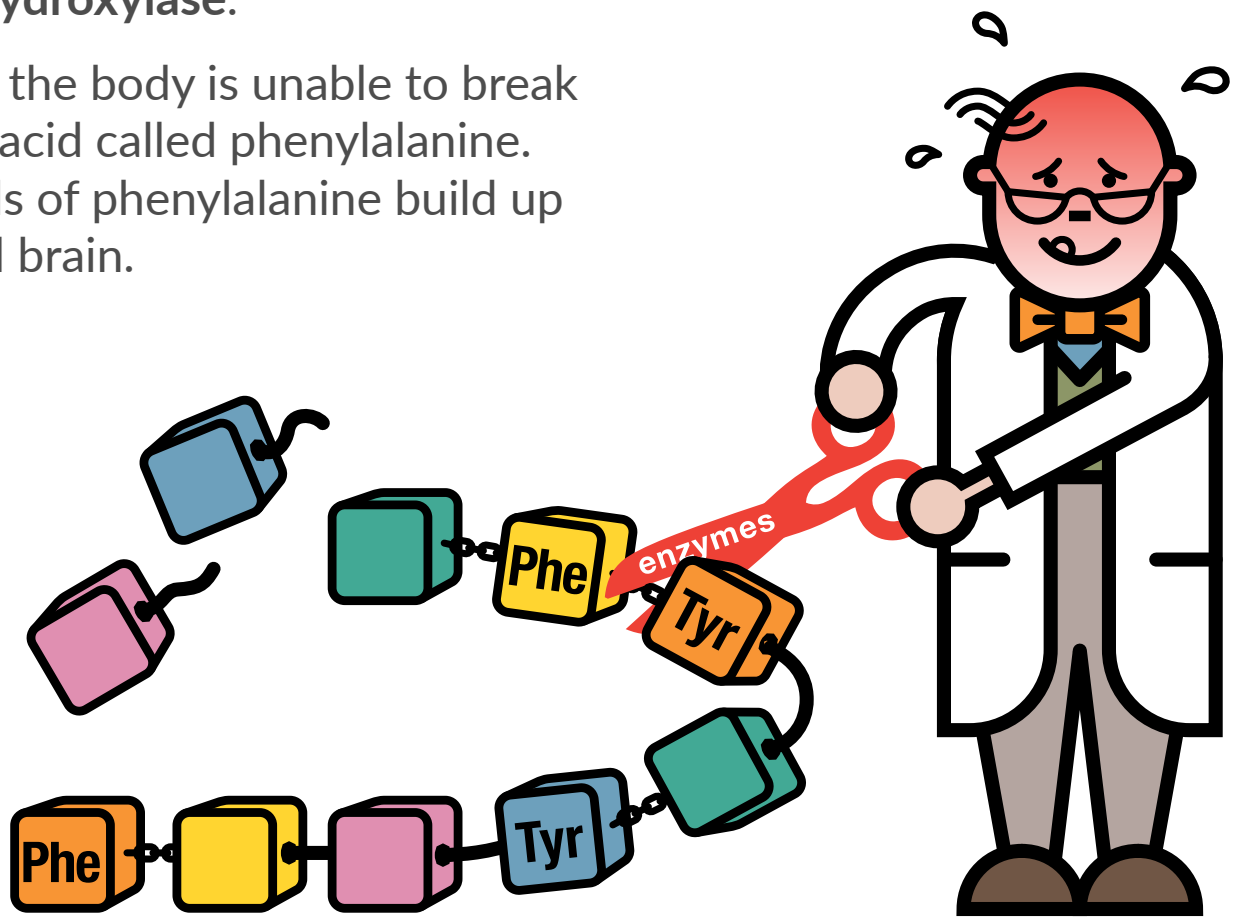
Metabolism refers to the chemical processes that occur inside the cells of the body.



What happens in PKU?

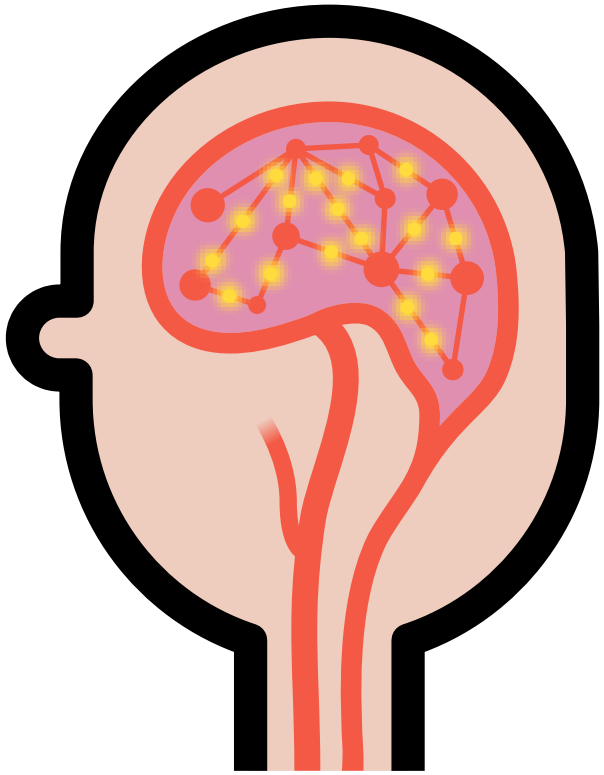
In PKU, the body lacks an enzyme called **phenylalanine hydroxylase**.

This means that the body is unable to break down an amino acid called phenylalanine. As a result, levels of phenylalanine build up in the blood and brain.



What can go wrong in PKU?

The build-up of phenylalanine can lead to brain damage, with learning and behavioural difficulties.

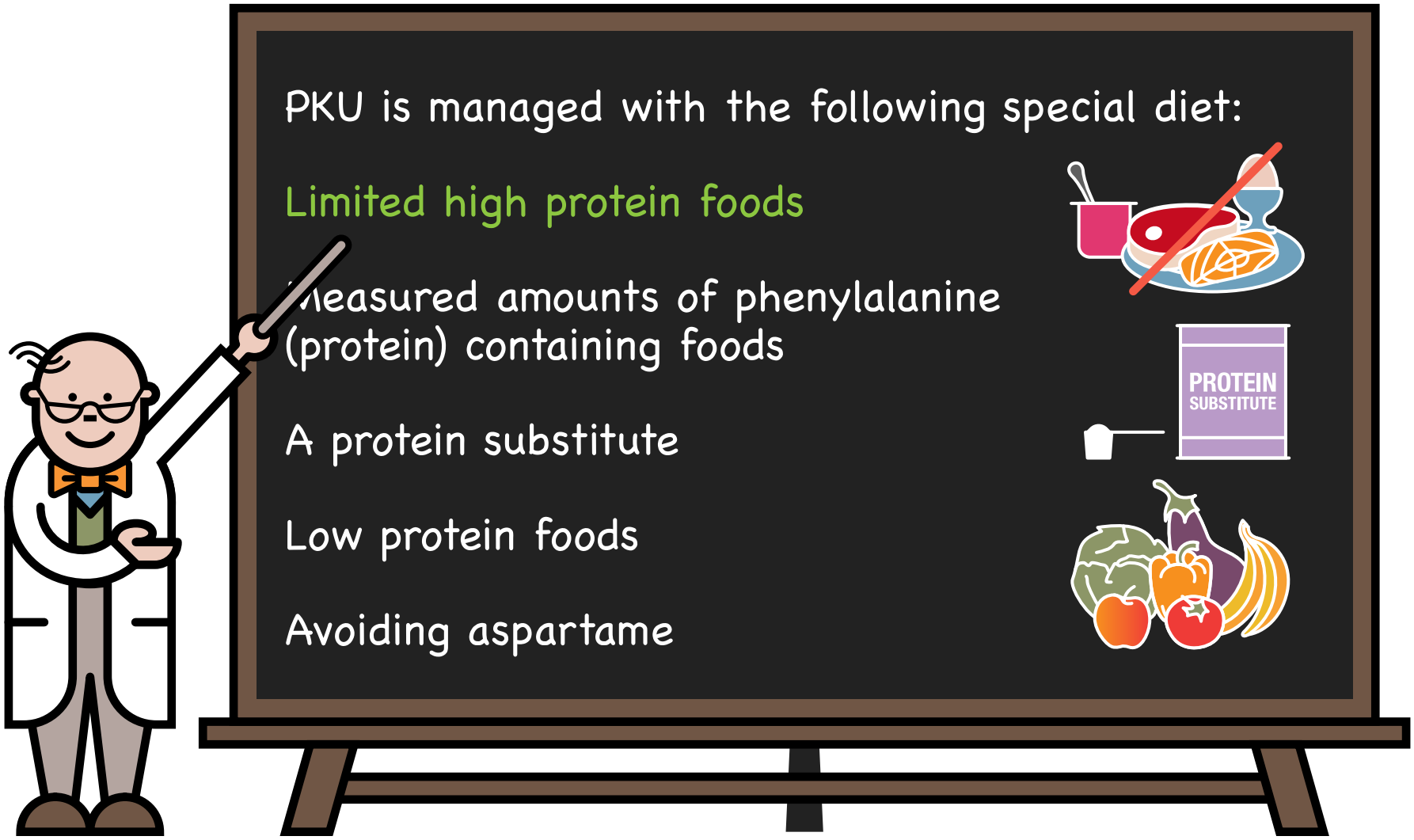


Early management can prevent brain damage and learning difficulties

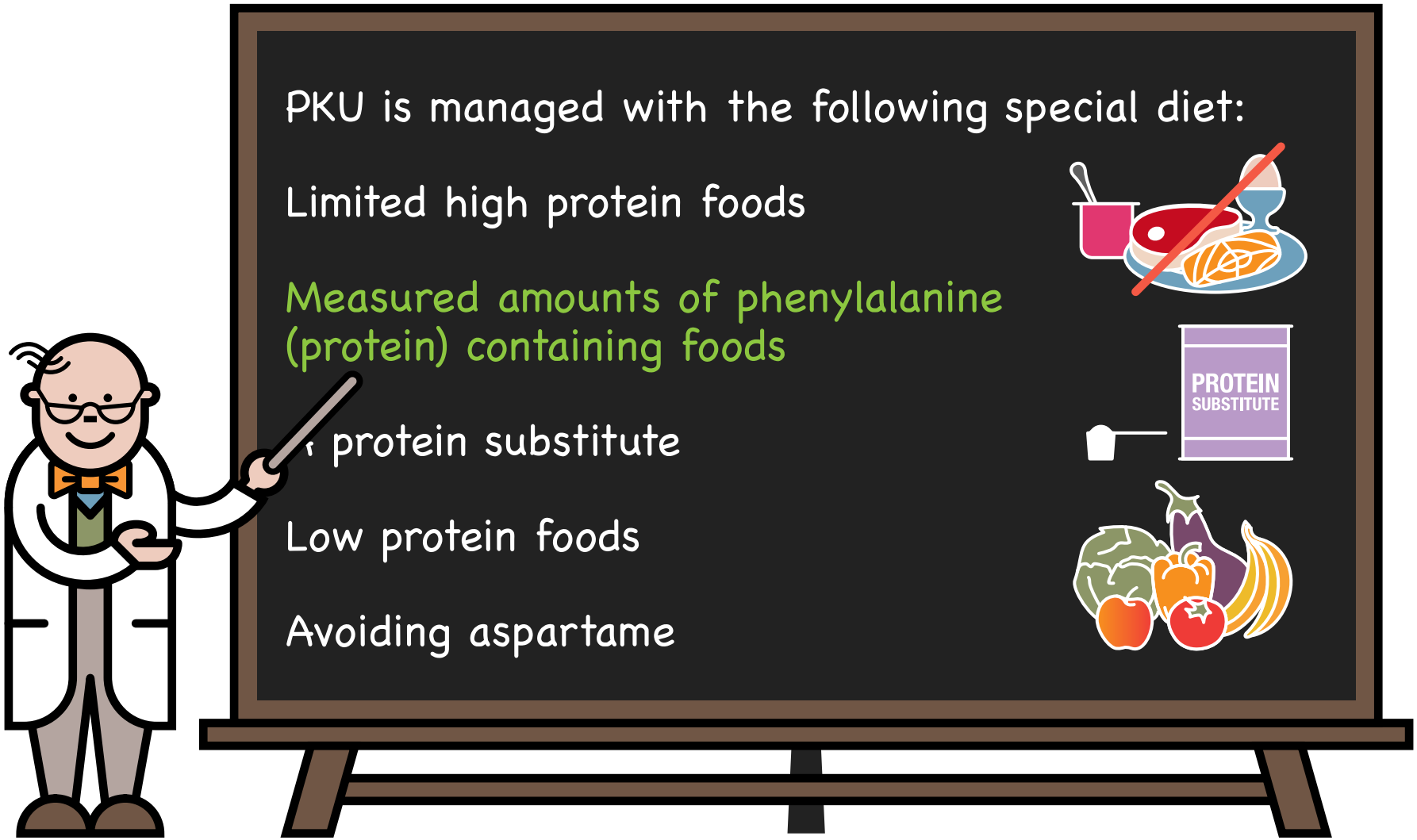
How is PKU diagnosed?

PKU is diagnosed by newborn screening. High levels of phenylalanine are found in the blood.

How is PKU managed?



How is PKU managed?



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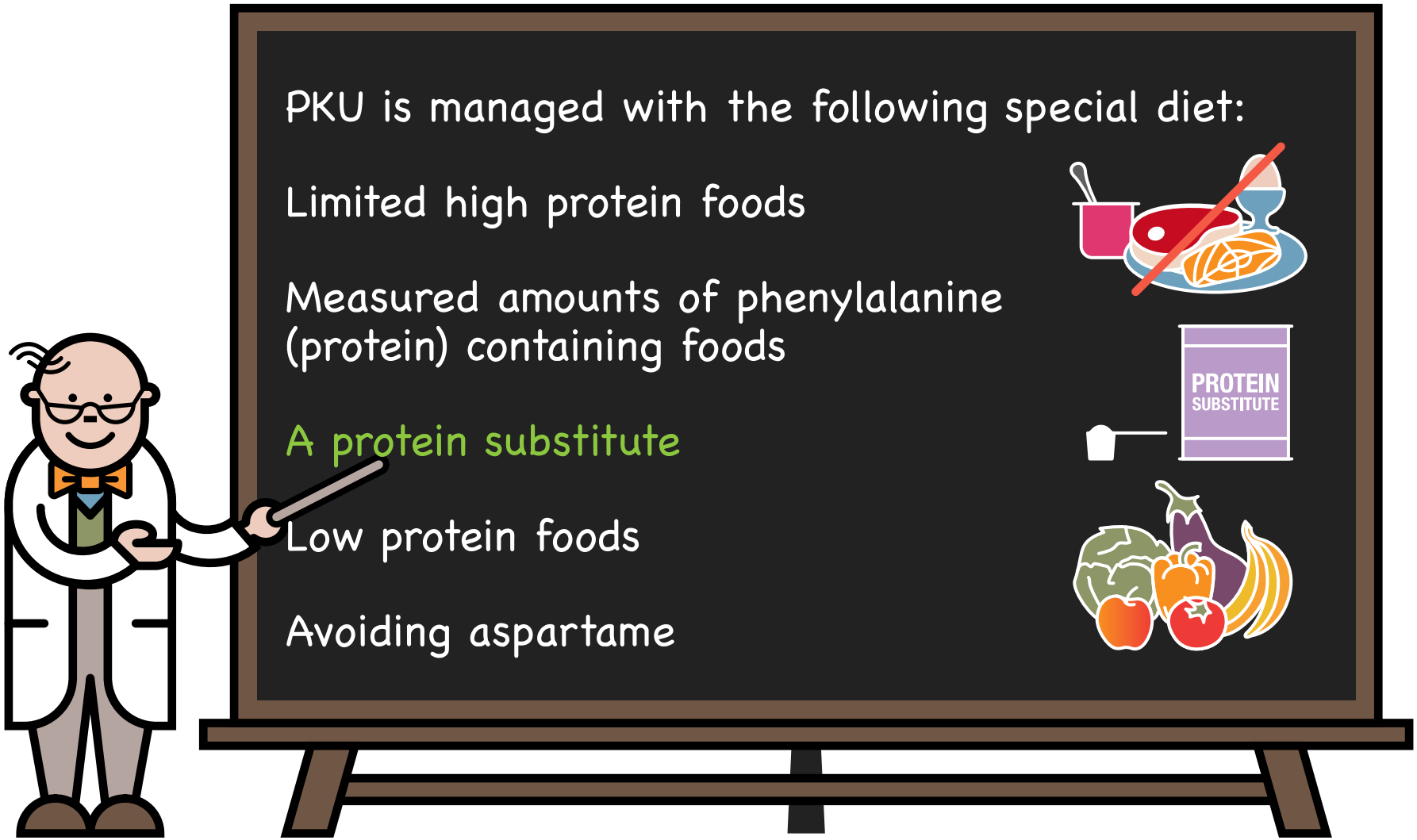

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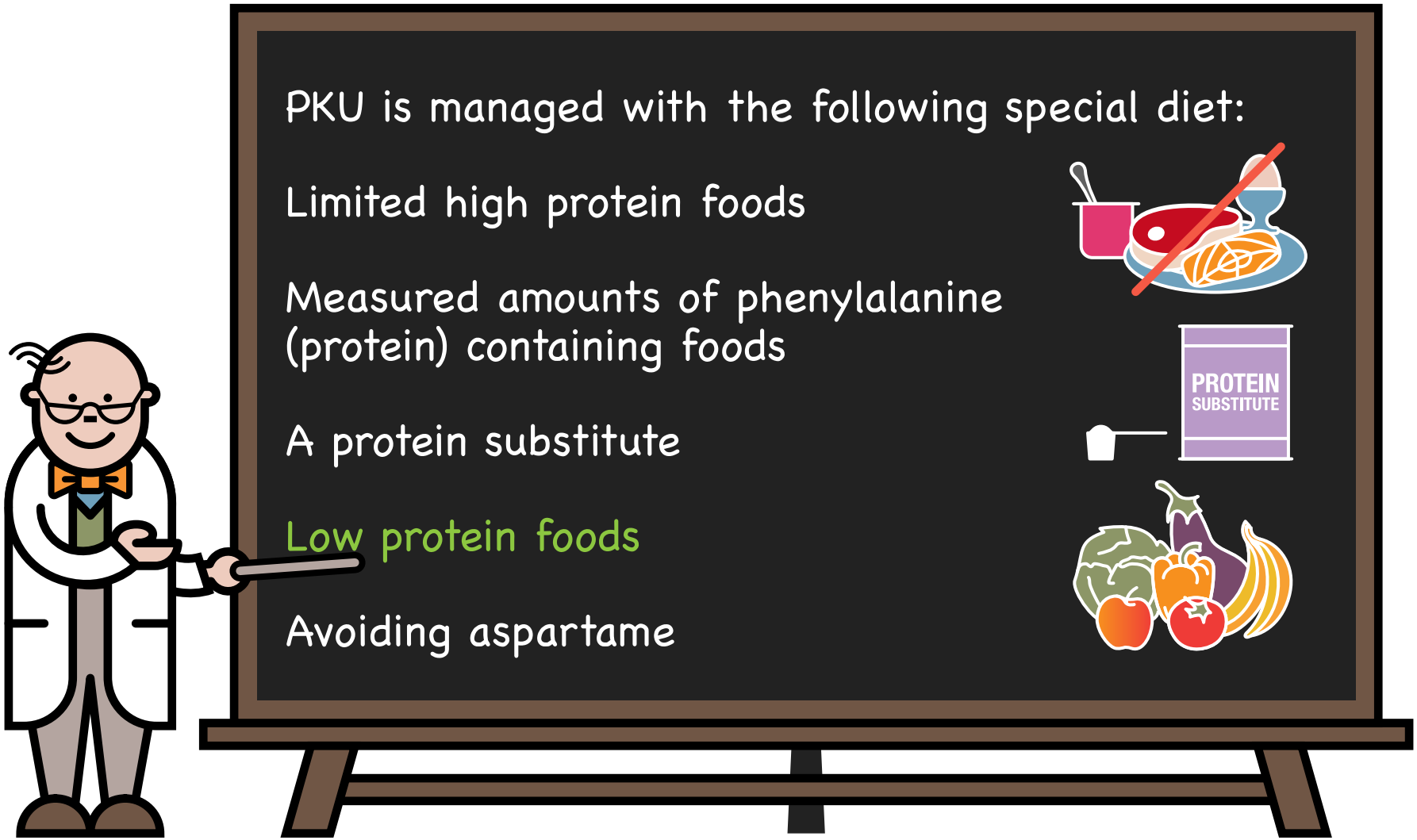

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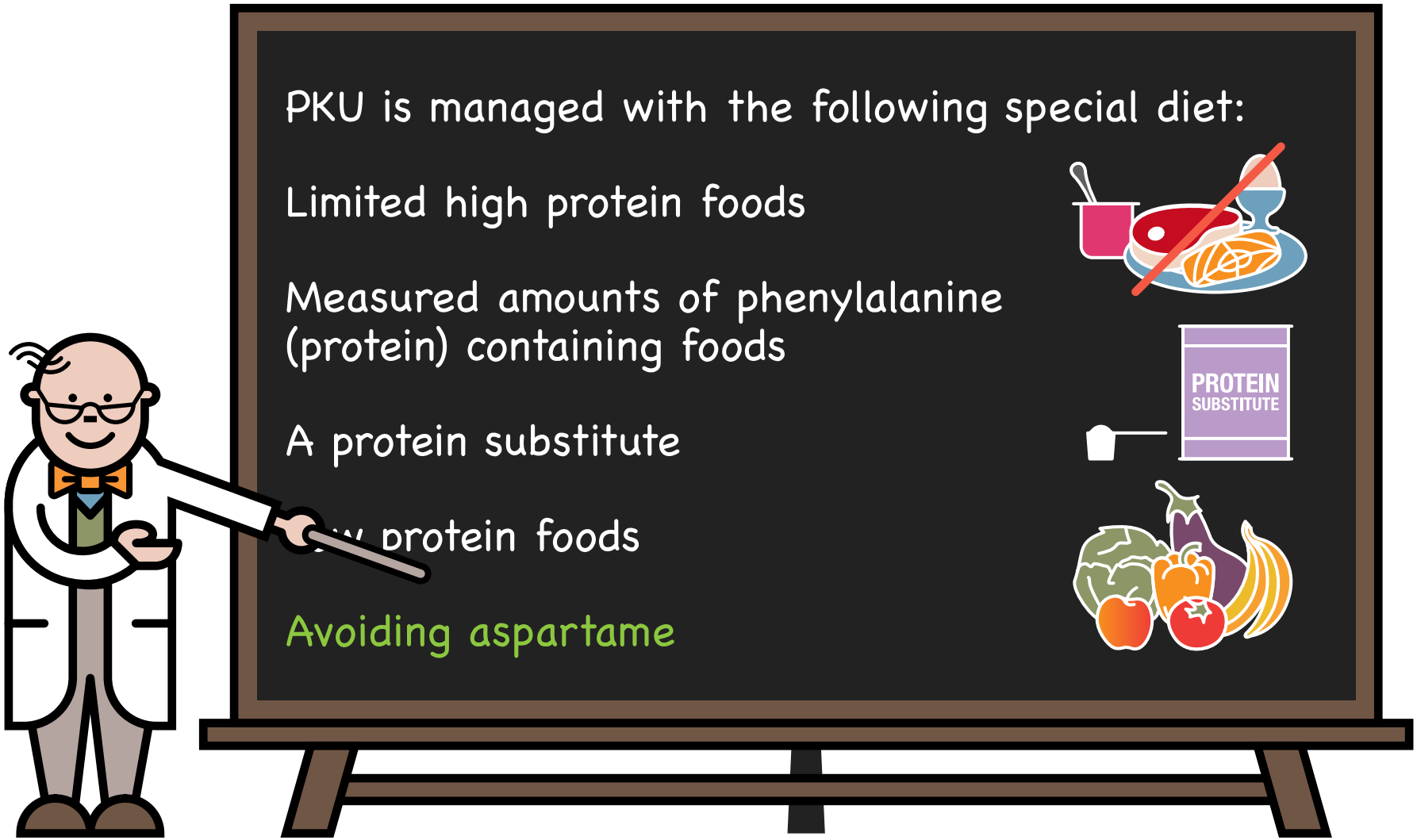


High protein foods

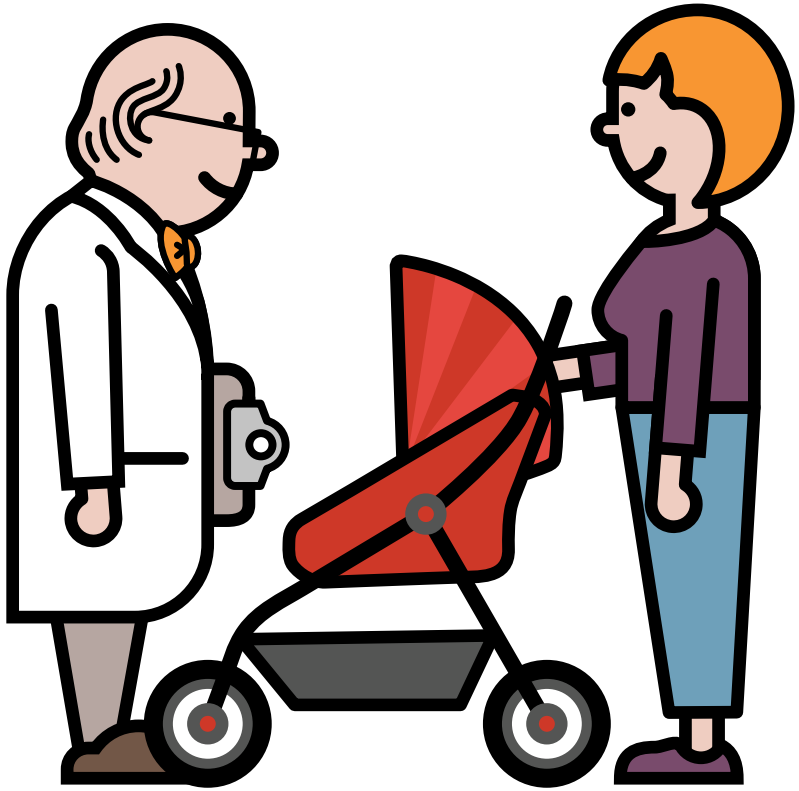
These foods are high in phenyalanine (protein) and must be avoided: **meat, fish, eggs, cheese, bread, pasta, nuts, seeds, soya** and **tofu**.

Any food or drink containing **aspartame** should also be avoided.

Measured phenylalanine intake

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

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

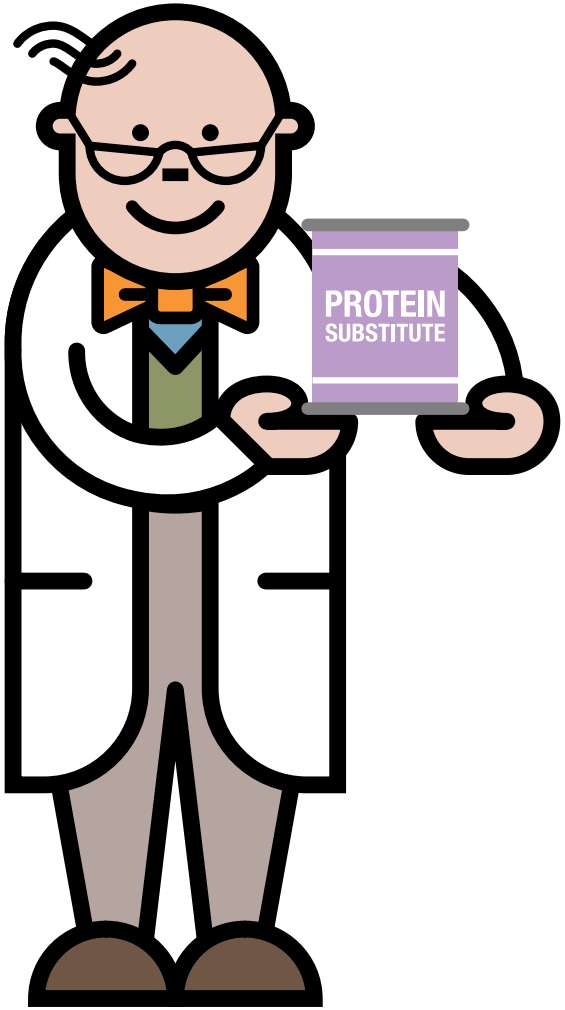


Protein substitute

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.

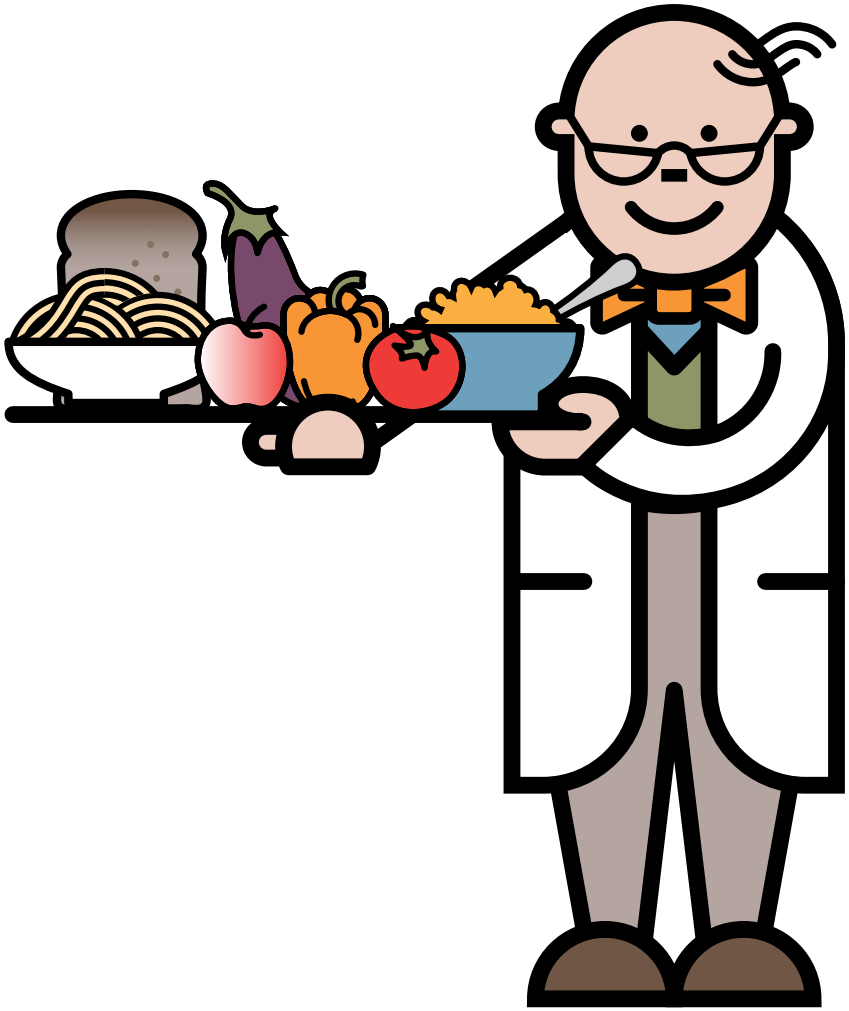


Low protein foods

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

They provide:

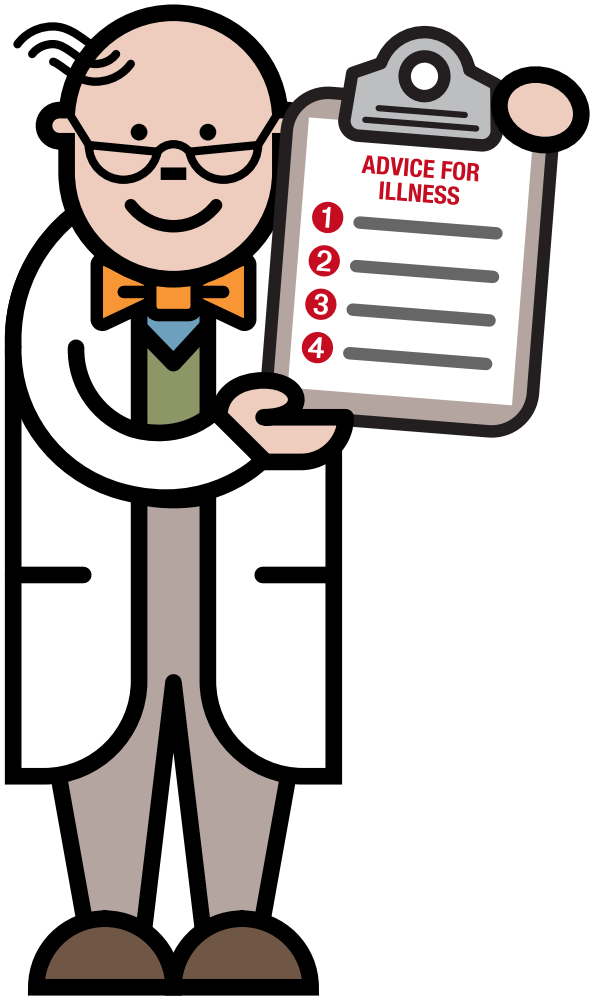
- a source of energy
- variety in the diet



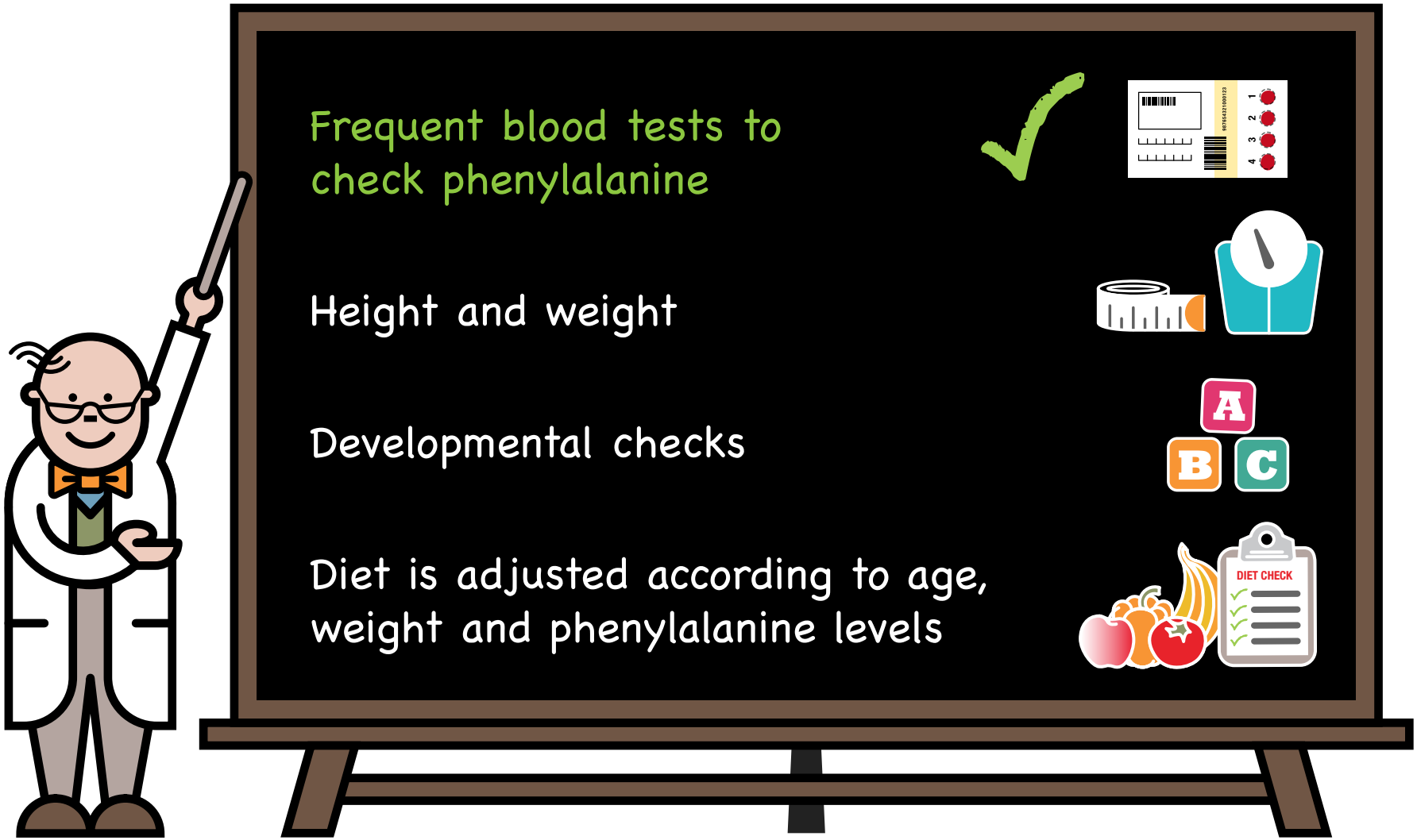
How is PKU managed during illness?

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

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



How is PKU monitored?



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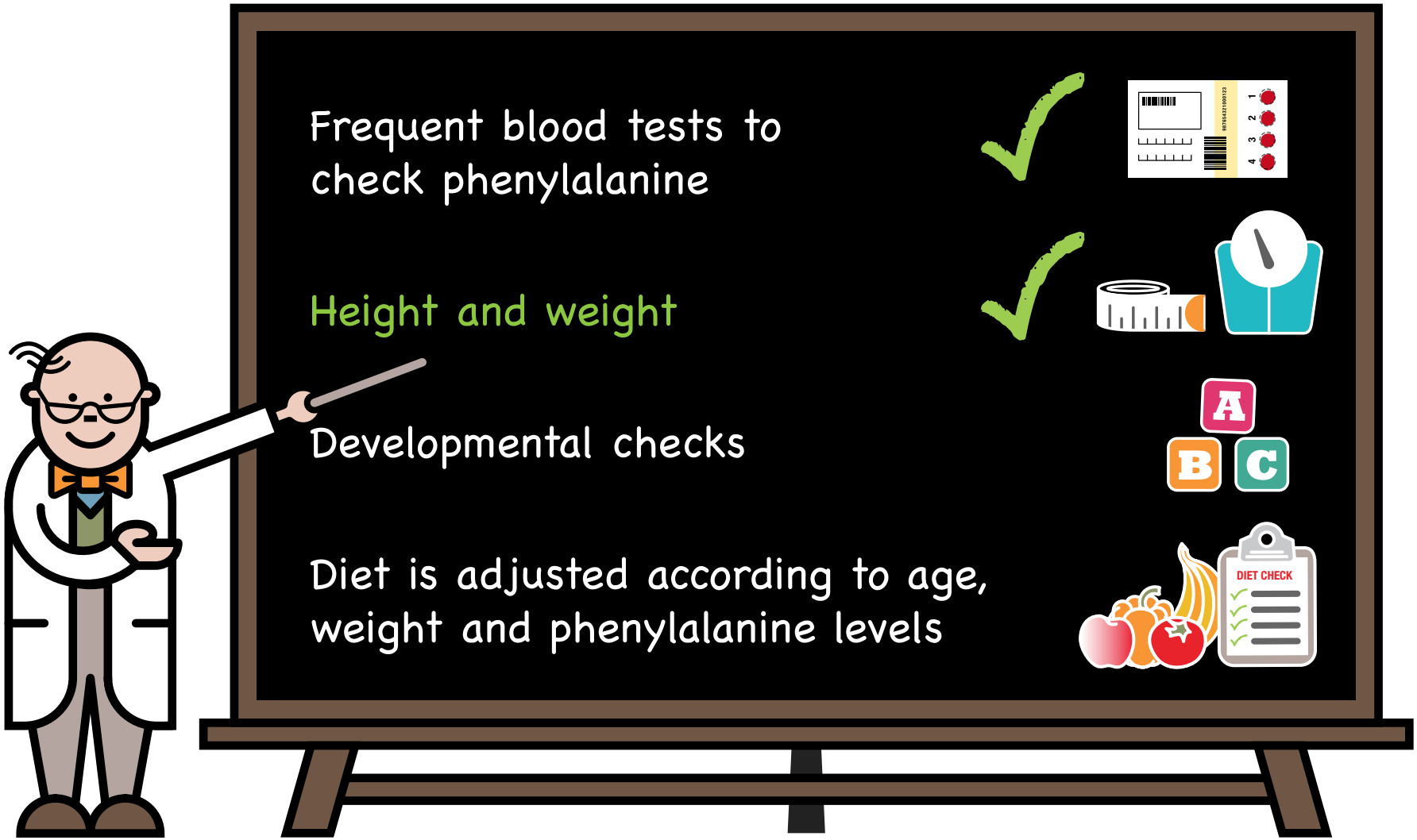

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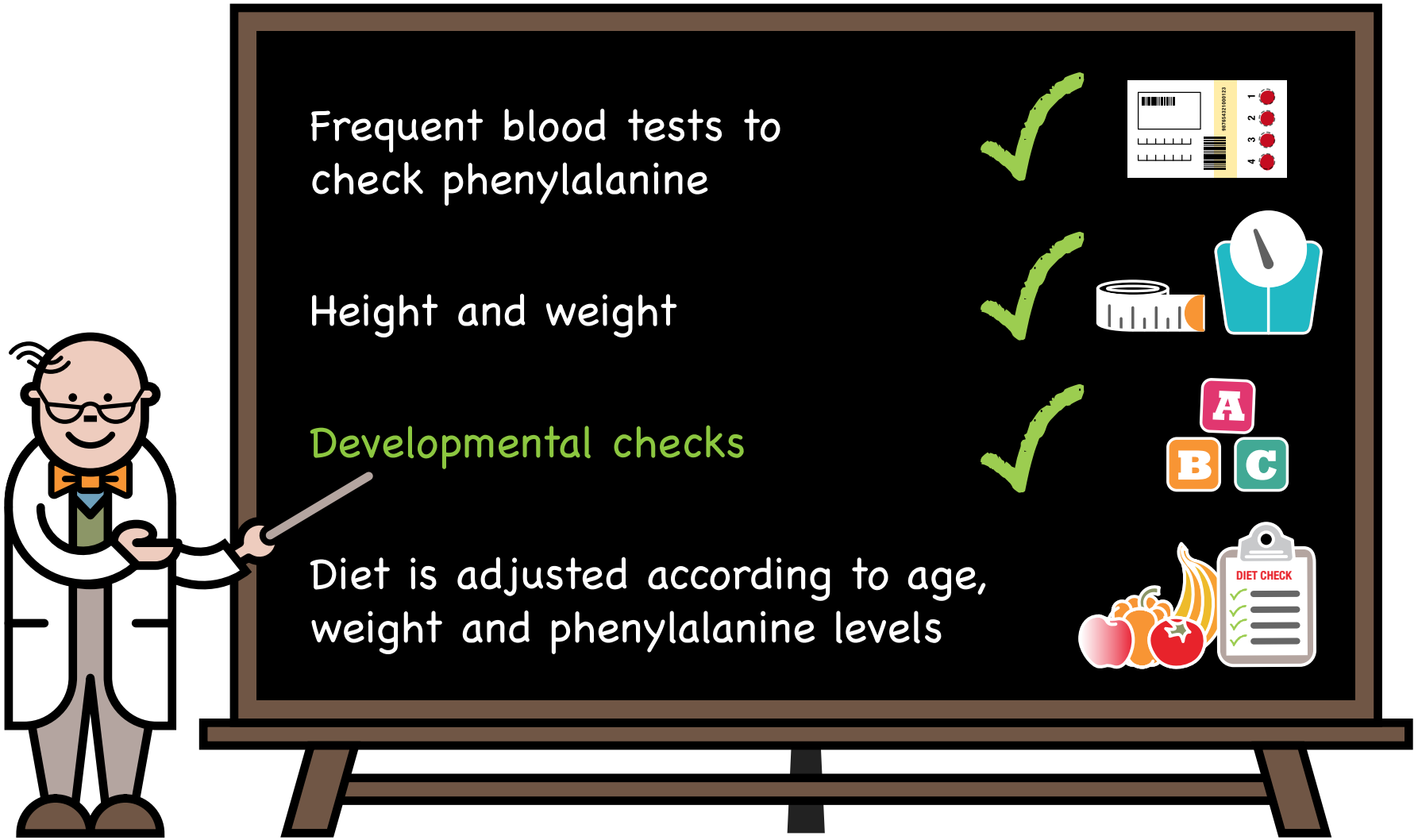

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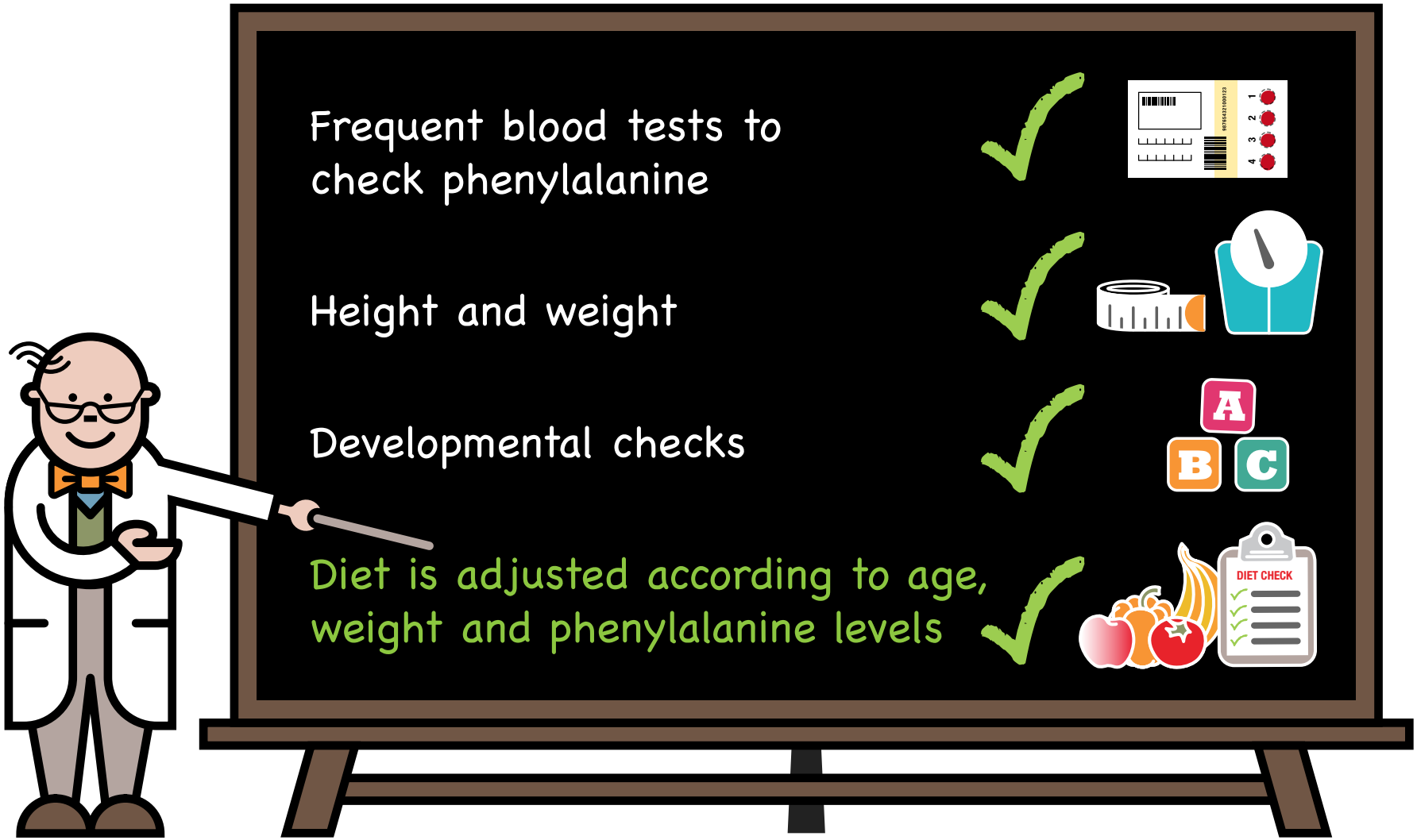
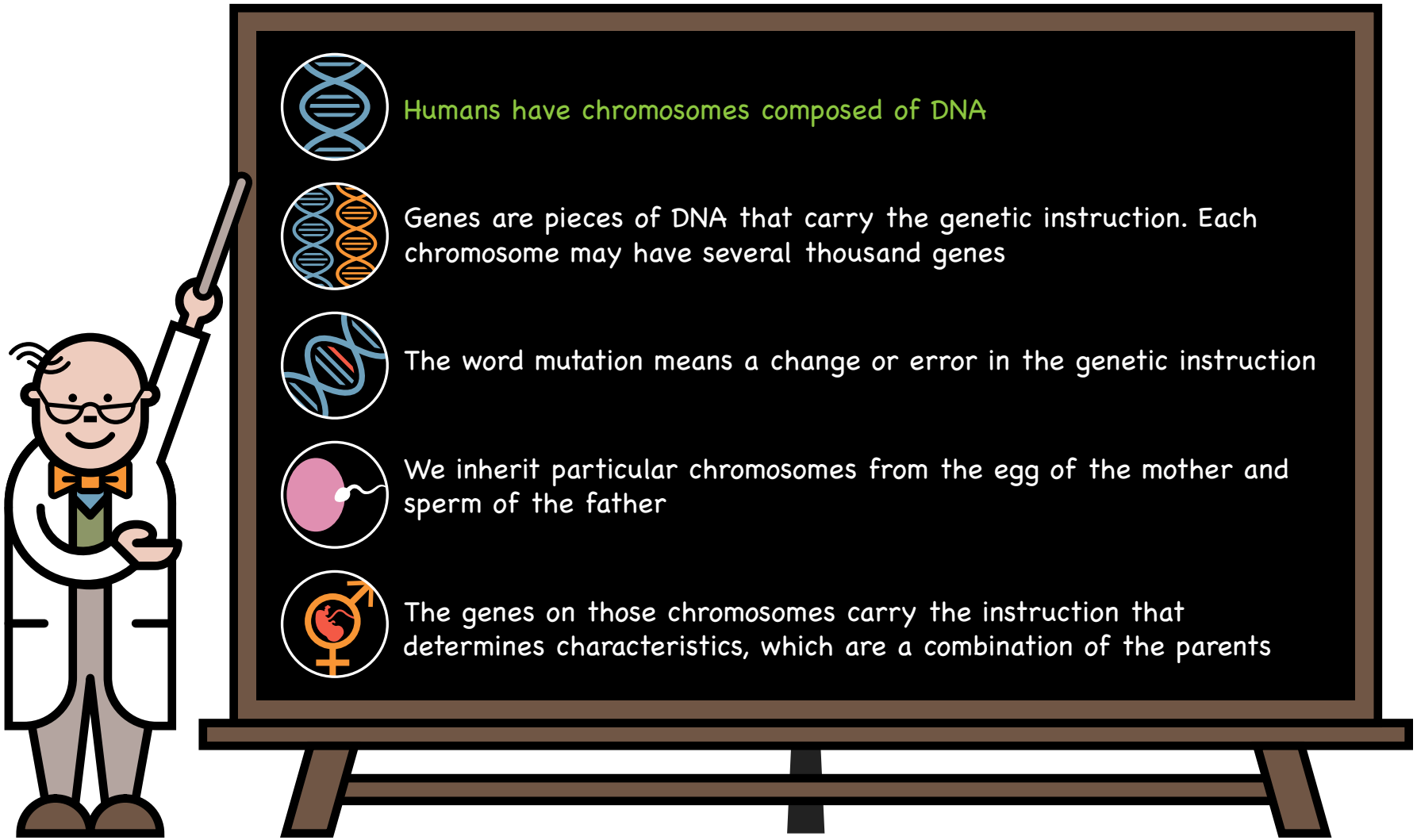
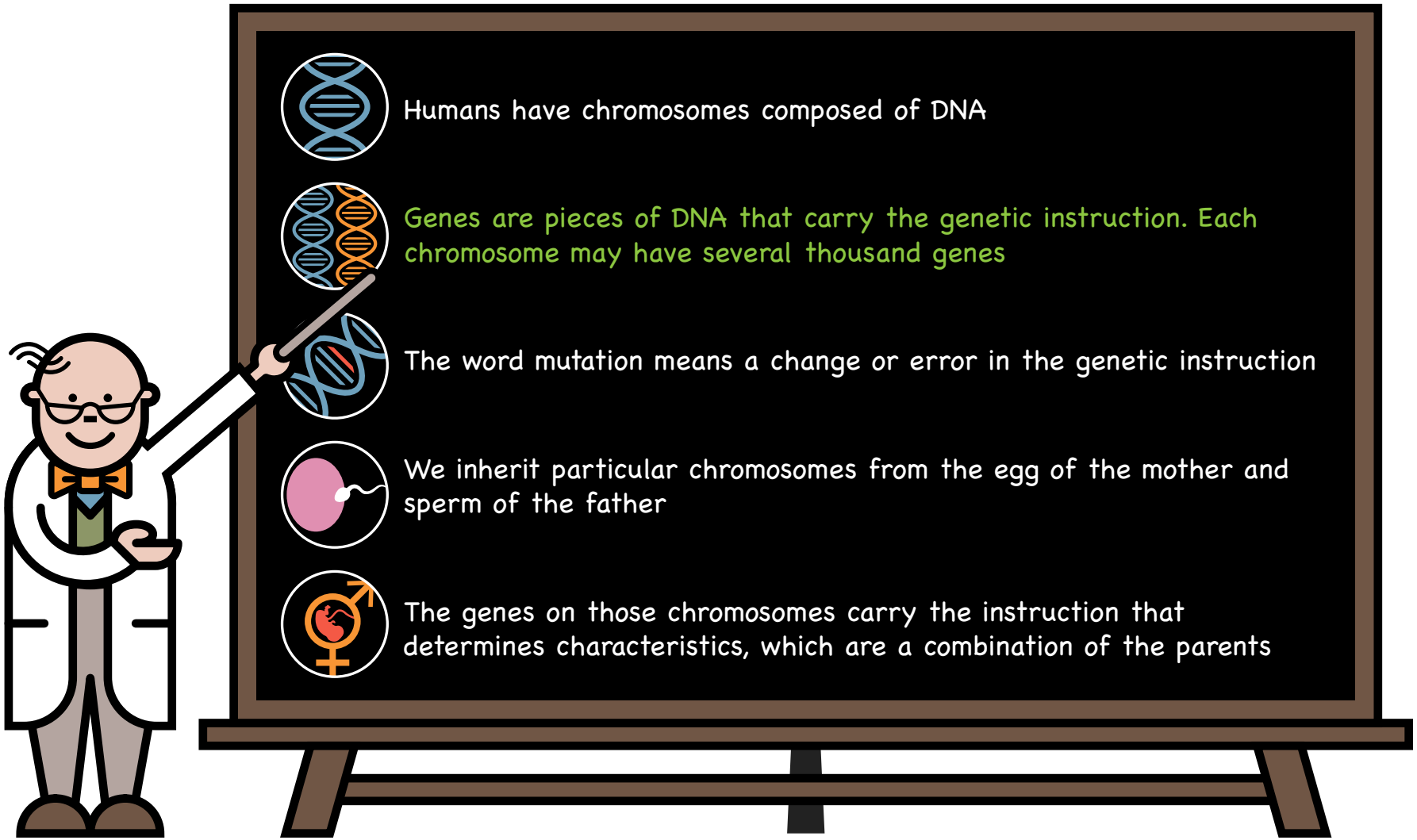


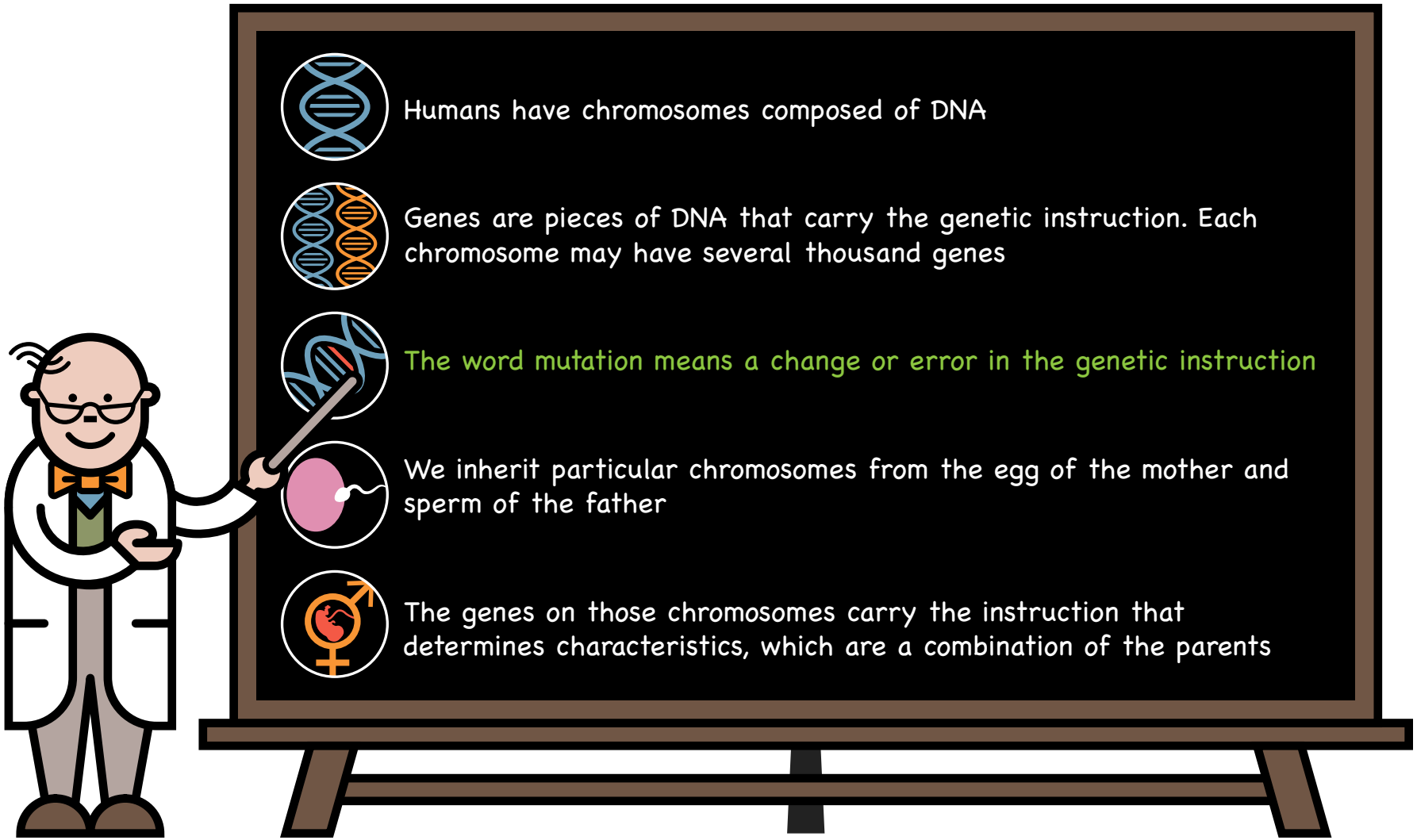
Chromosomes, genes, mutations



Chromosomes, genes, mutations



Chromosomes, genes, mutations



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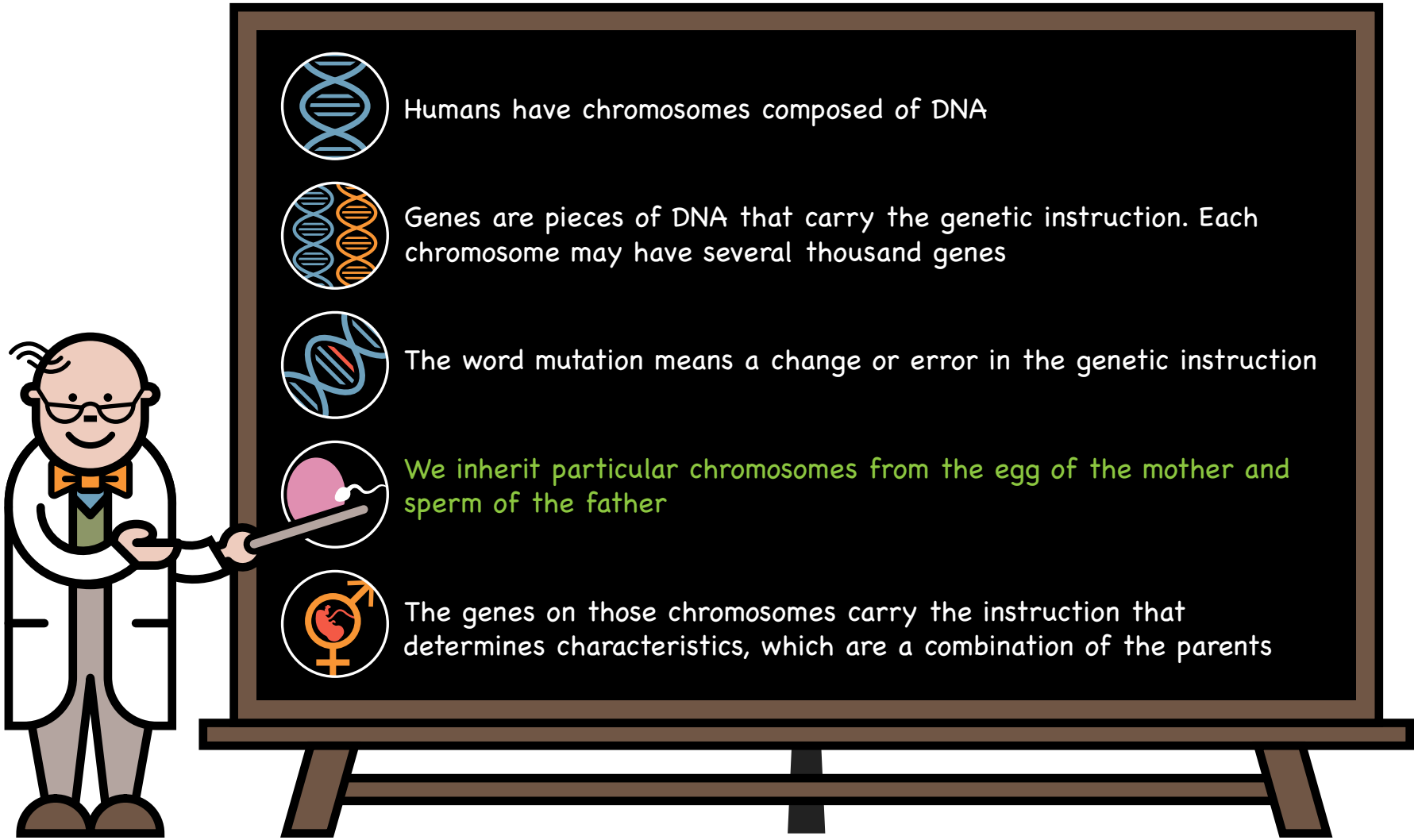

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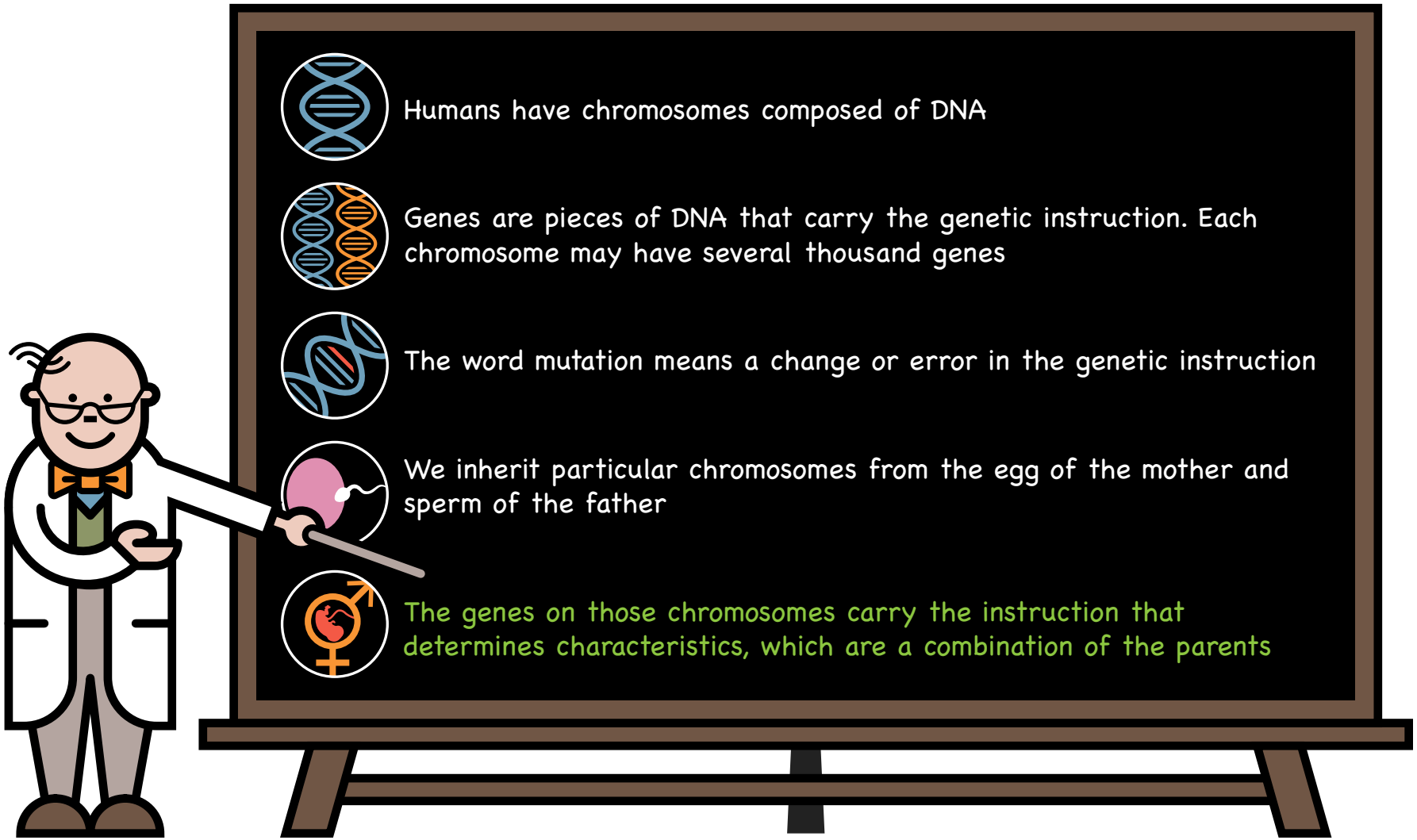
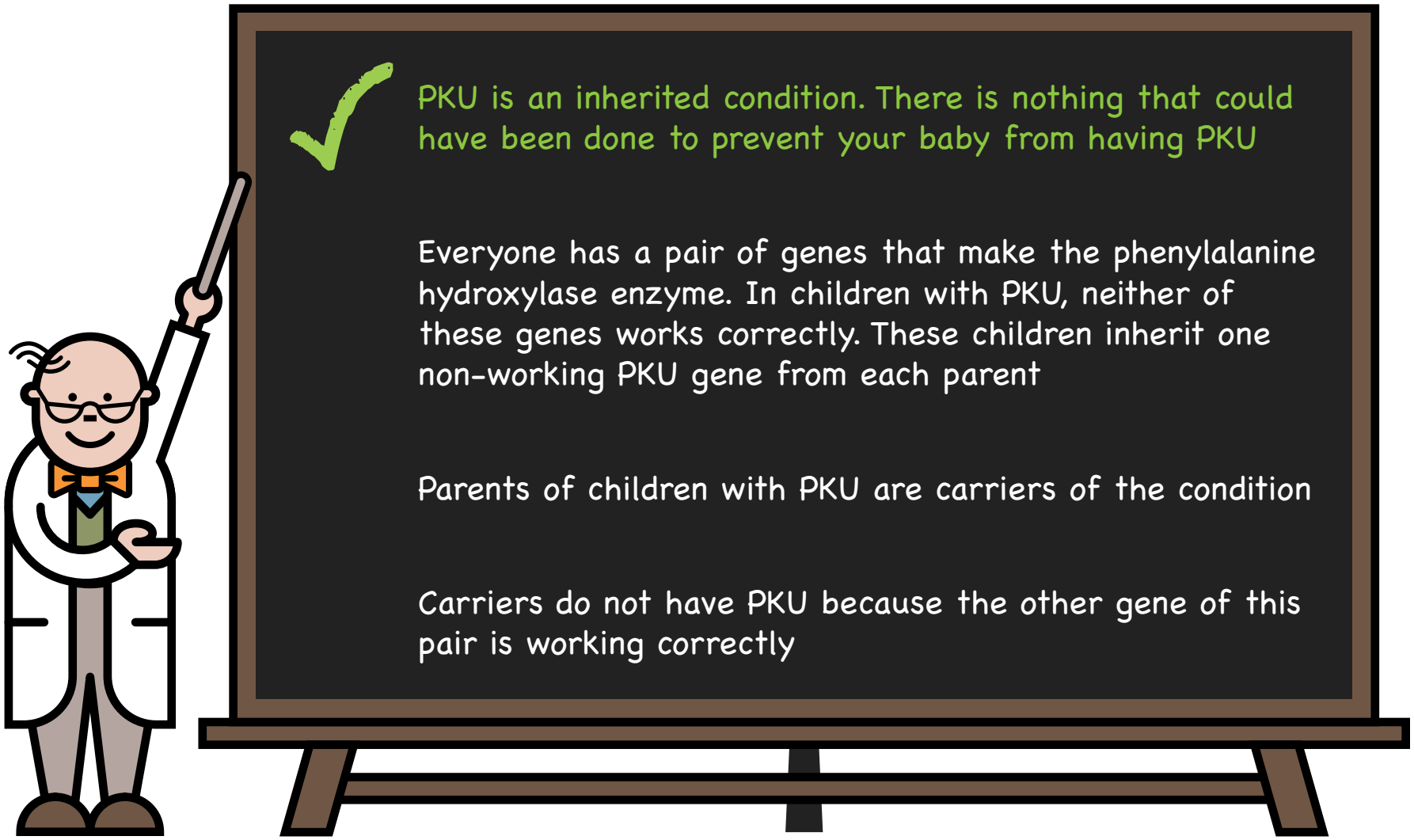


Inheritance



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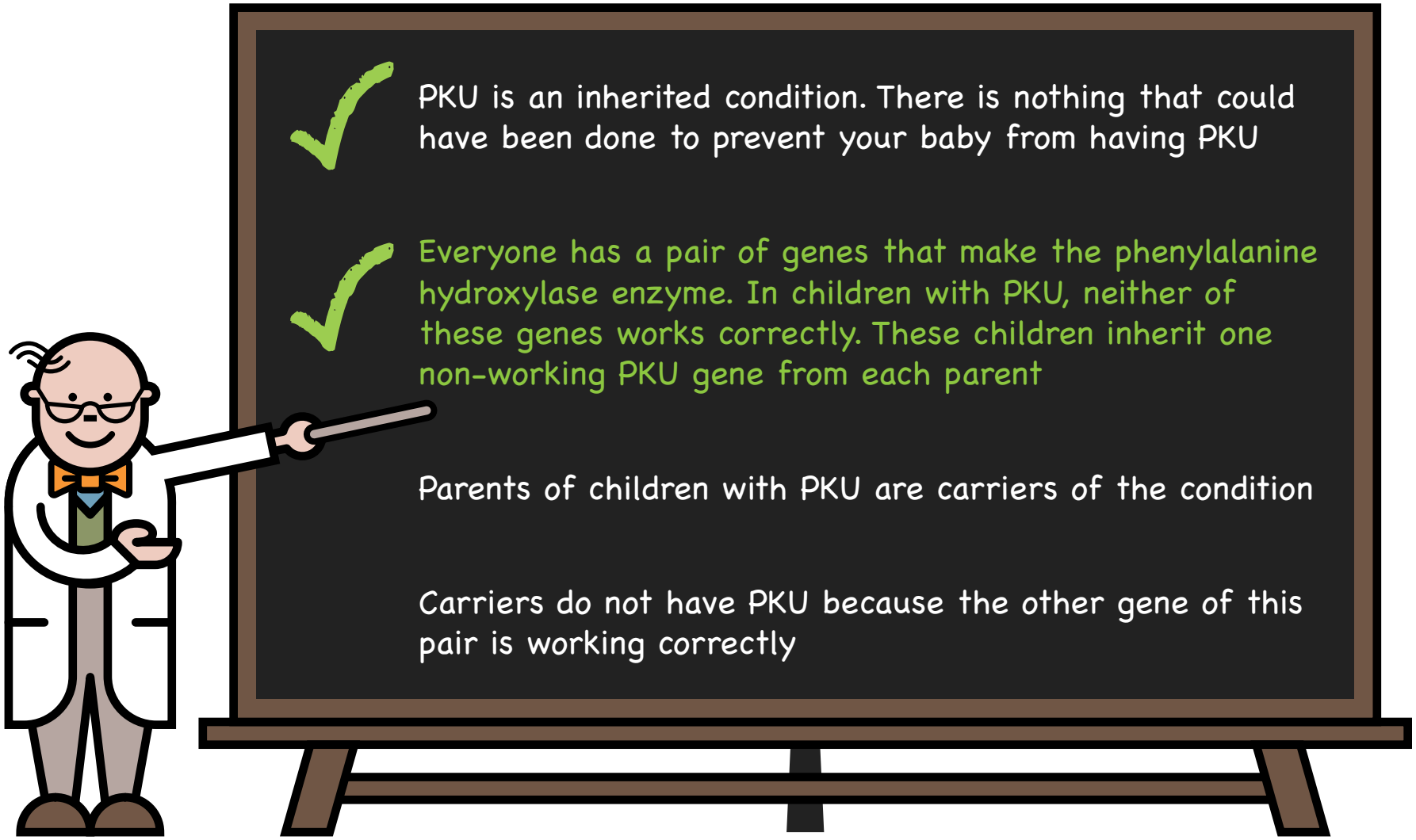

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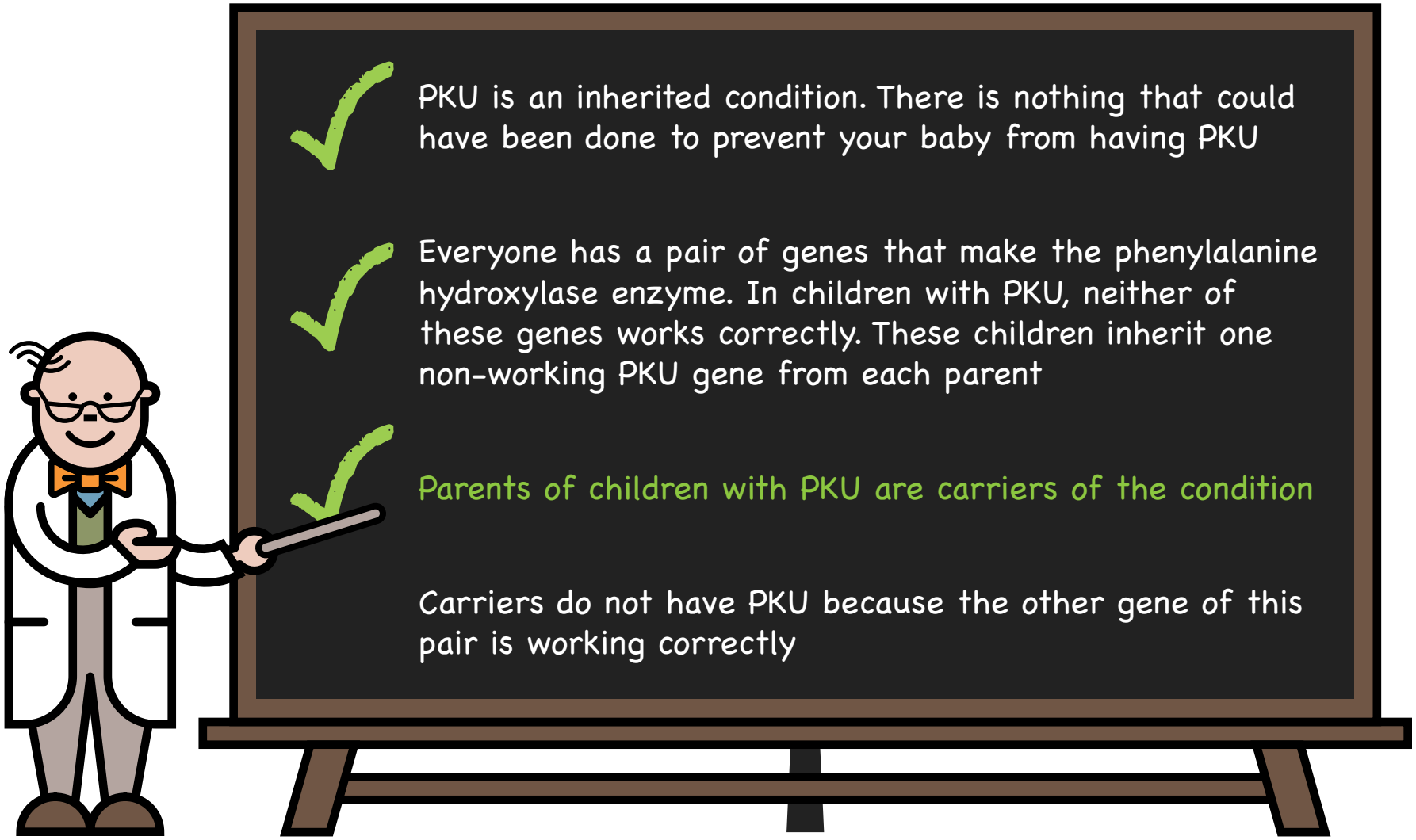

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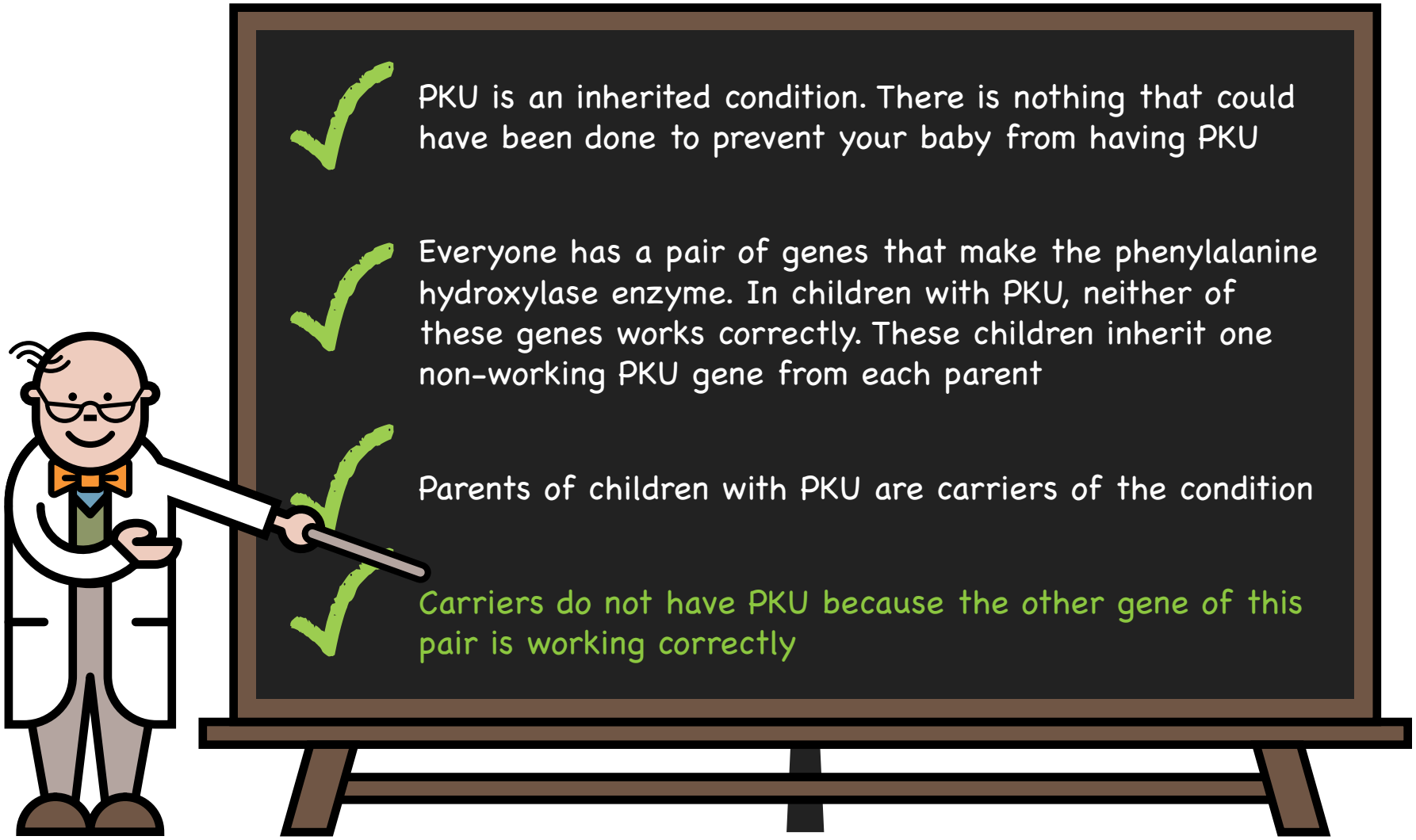
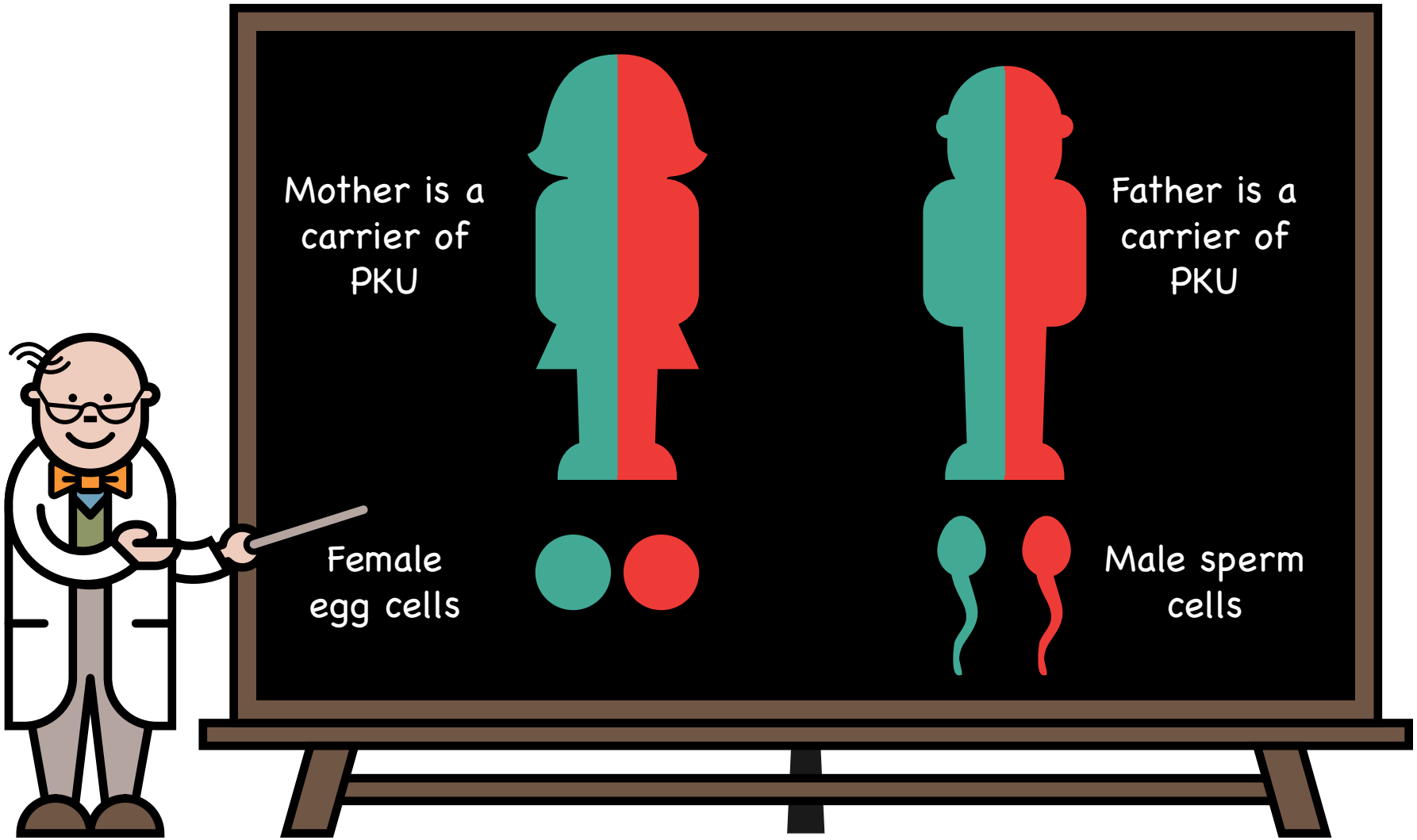
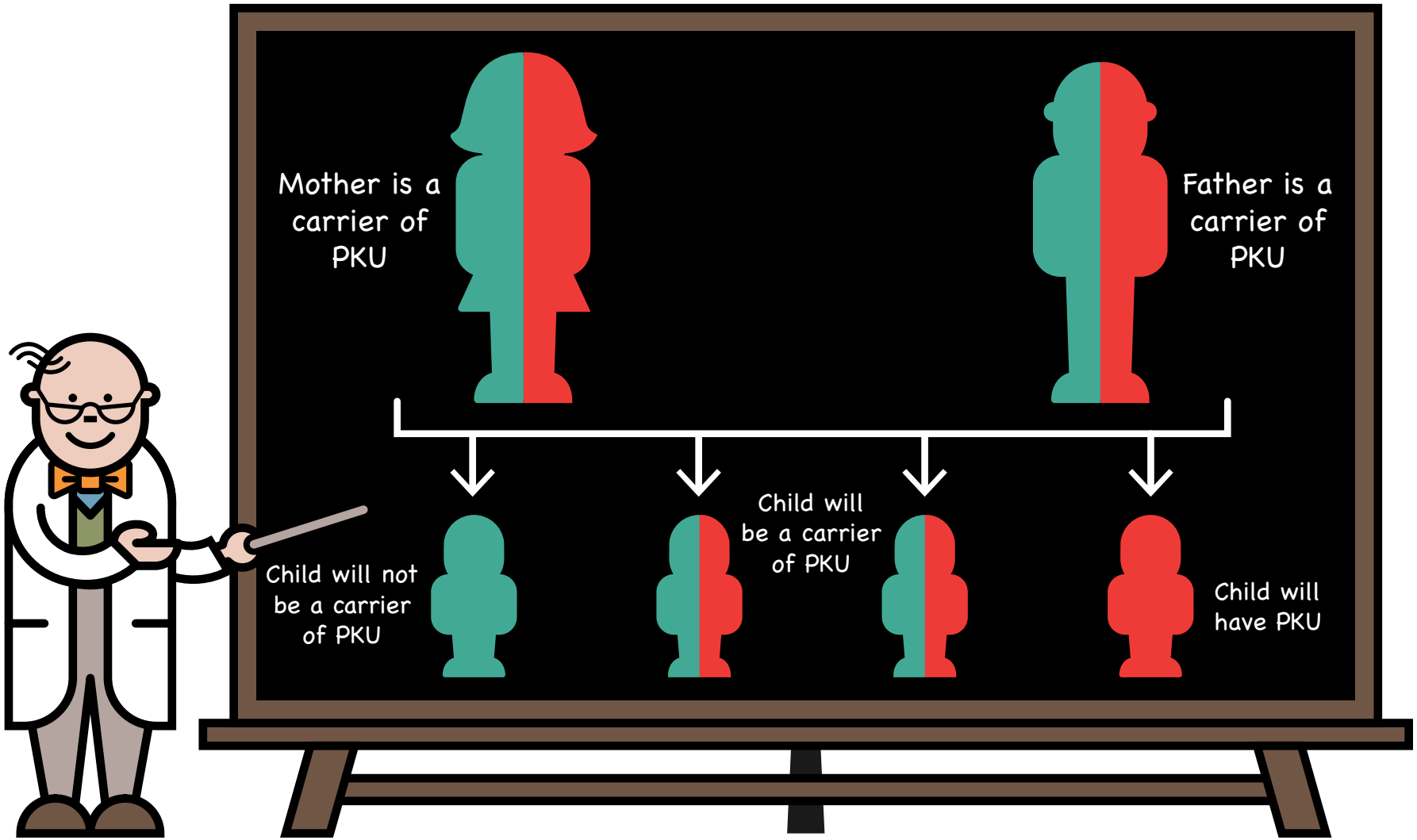


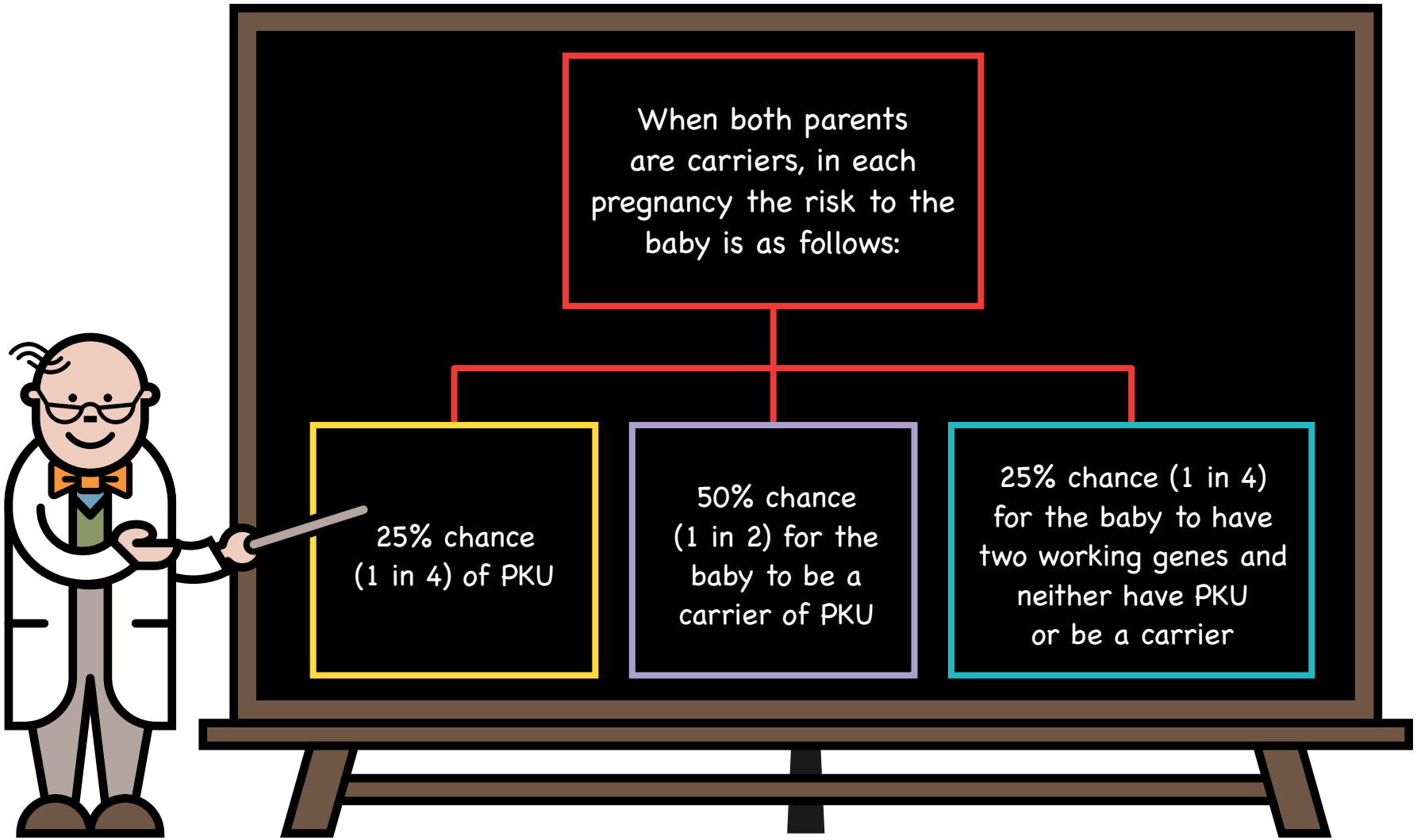
Inheritance – Autosomal recessive (carriers of PKU)



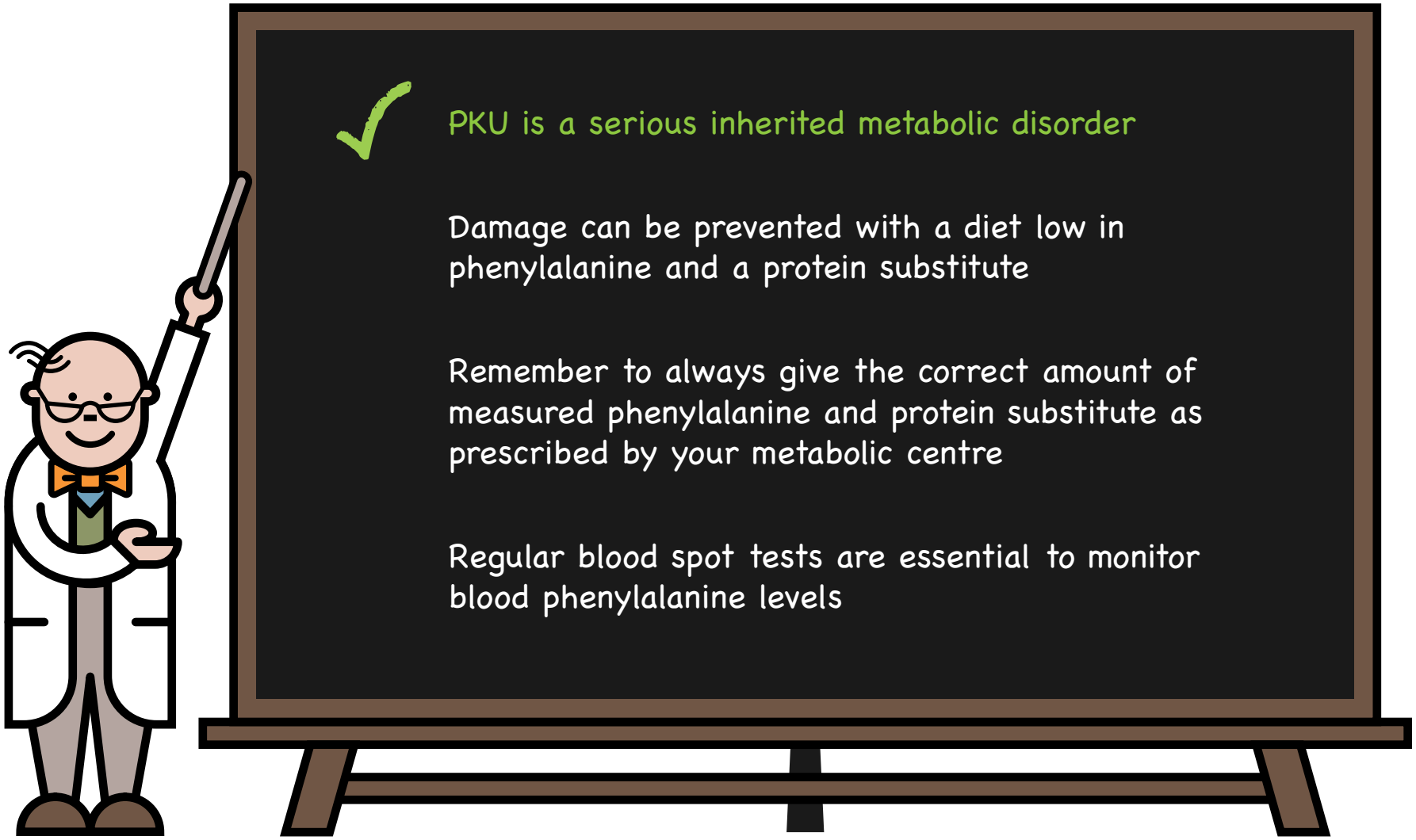
Inheritance – Autosomal recessive – possible combinations



Future pregnancies



Take home messages



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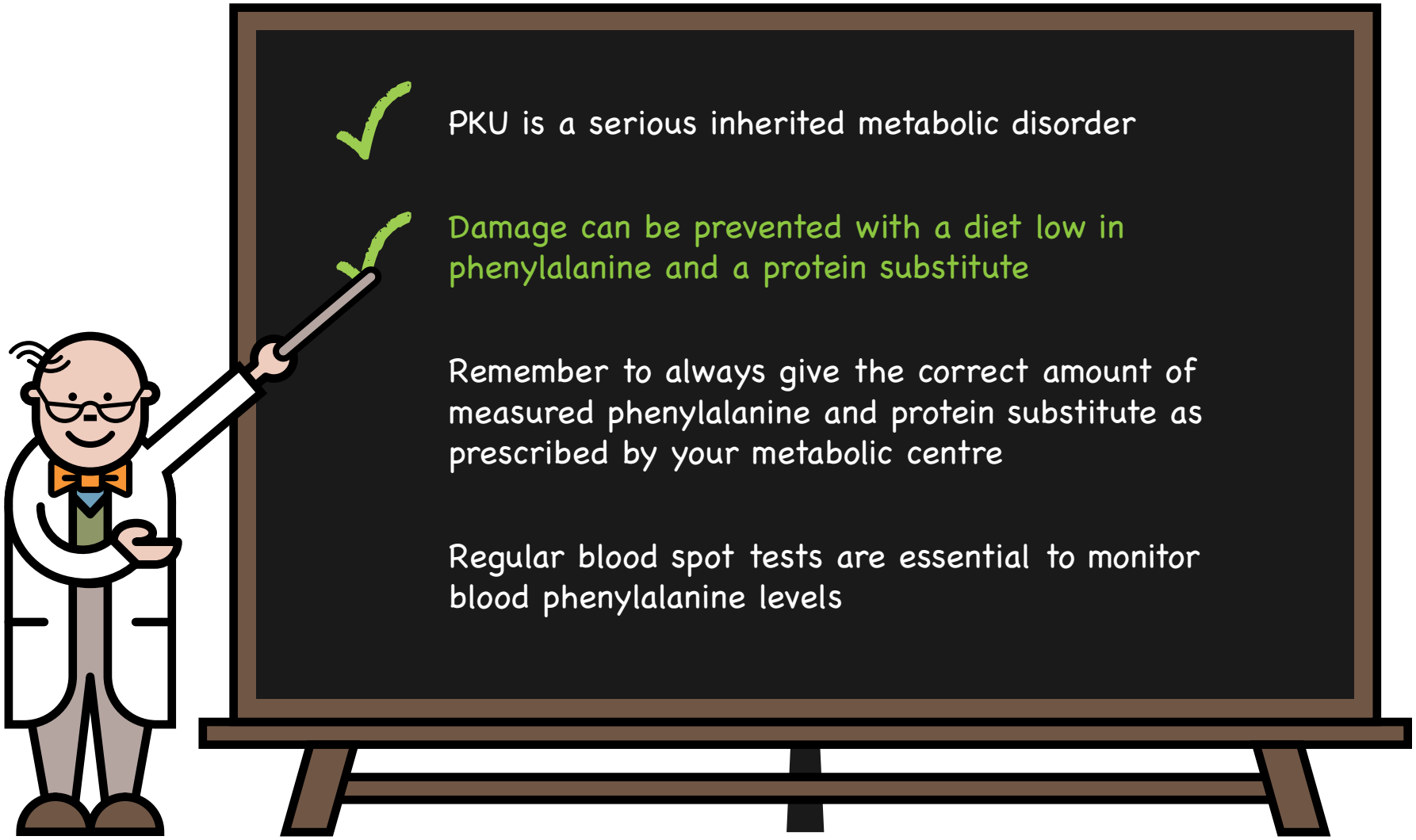

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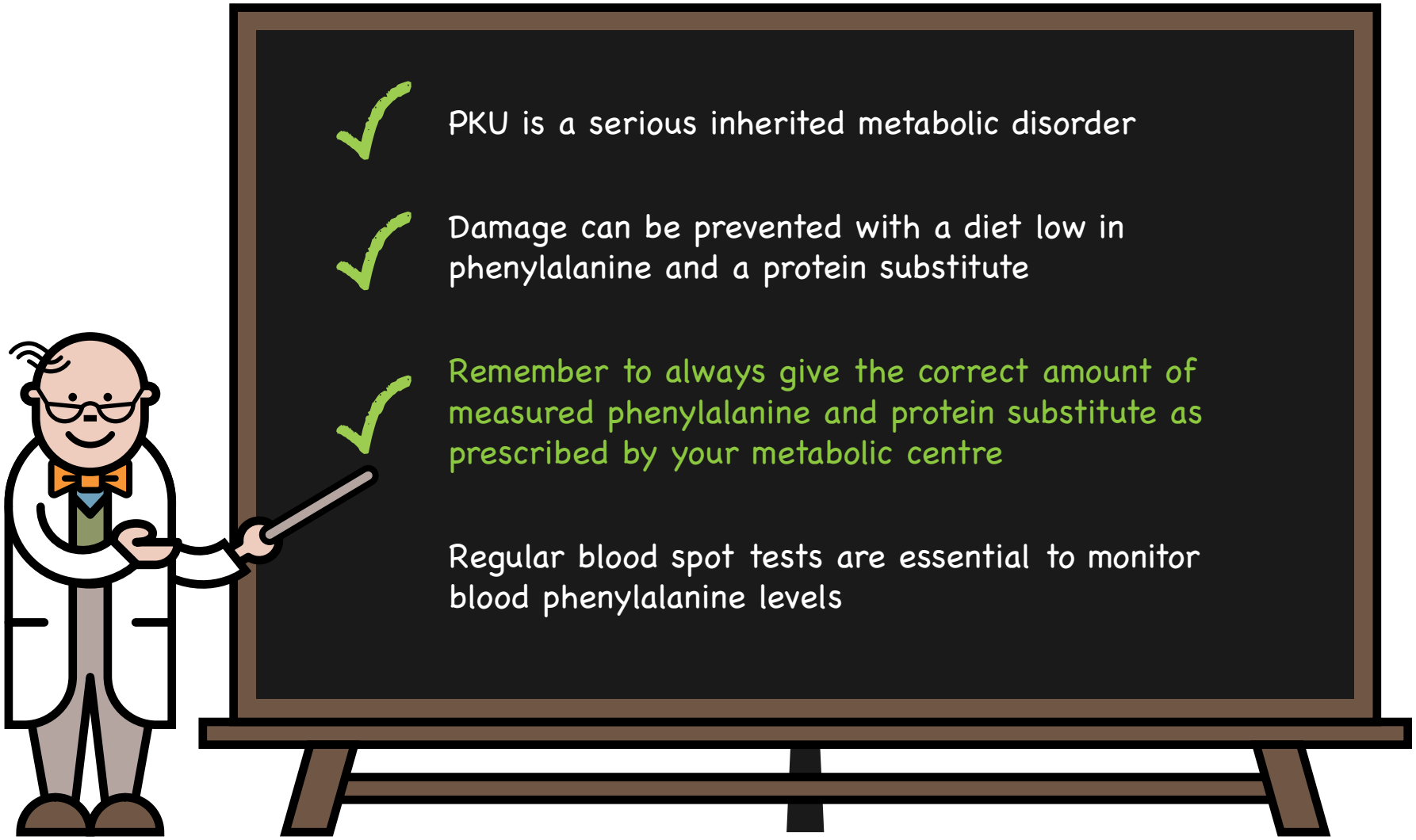

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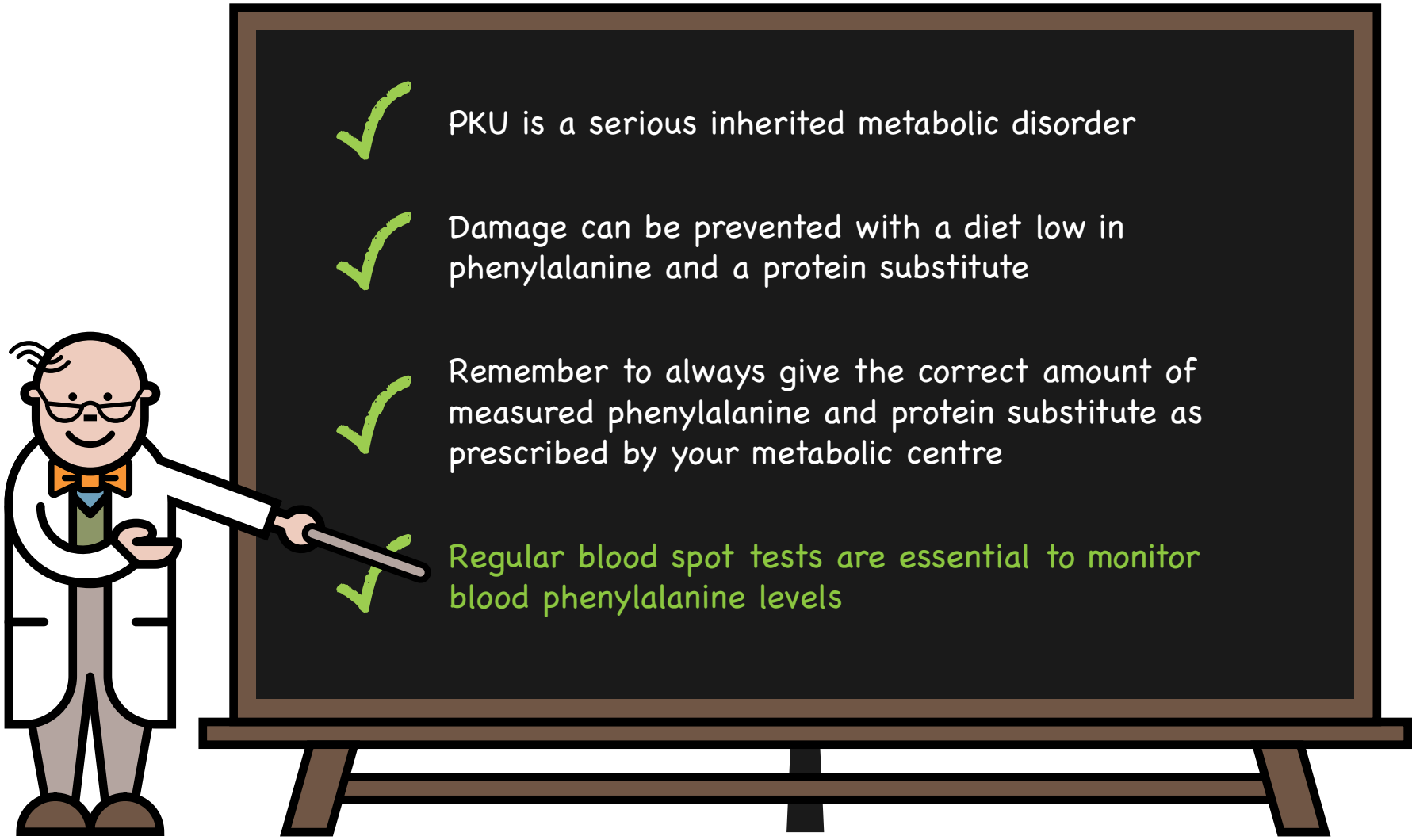
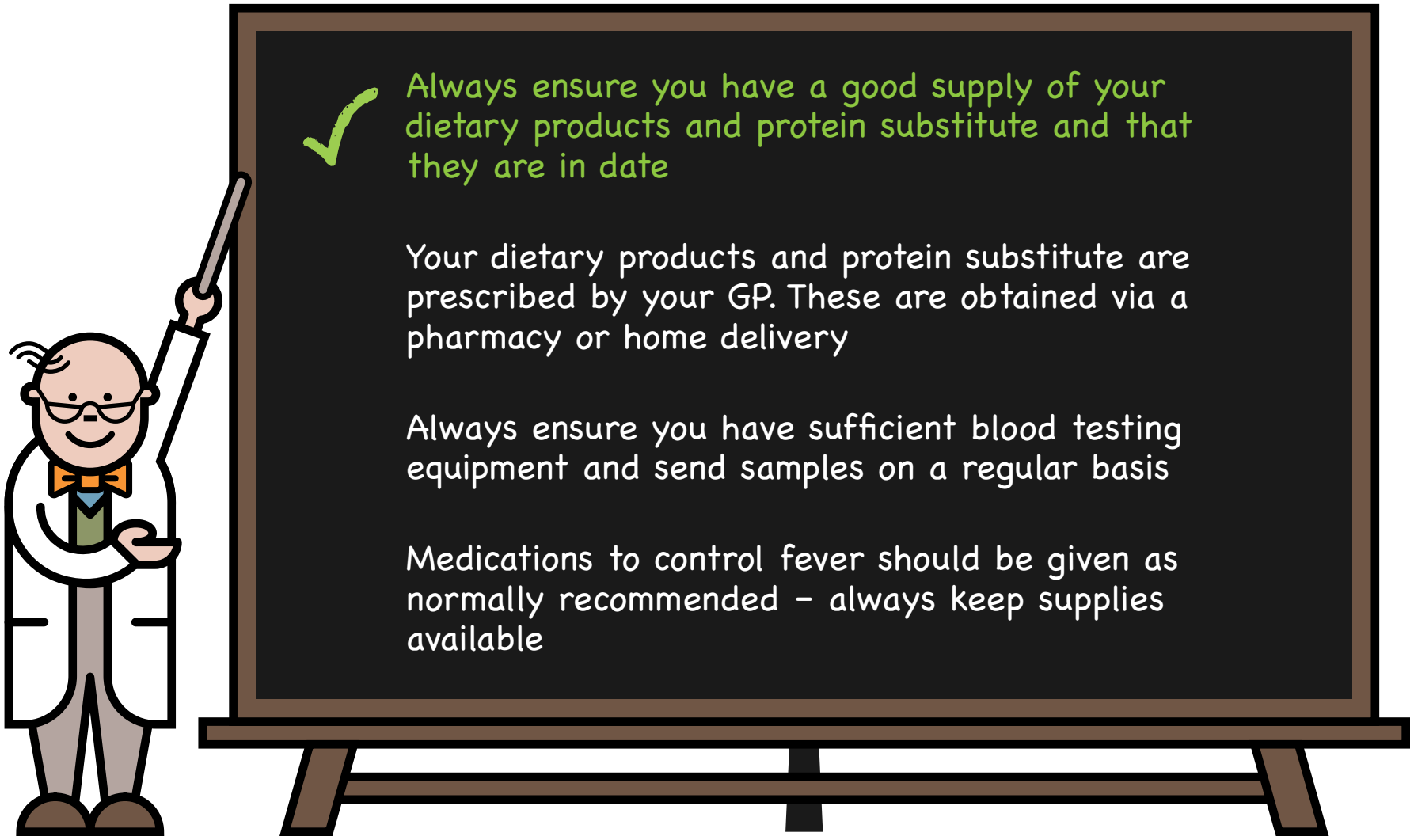


Helpful hints



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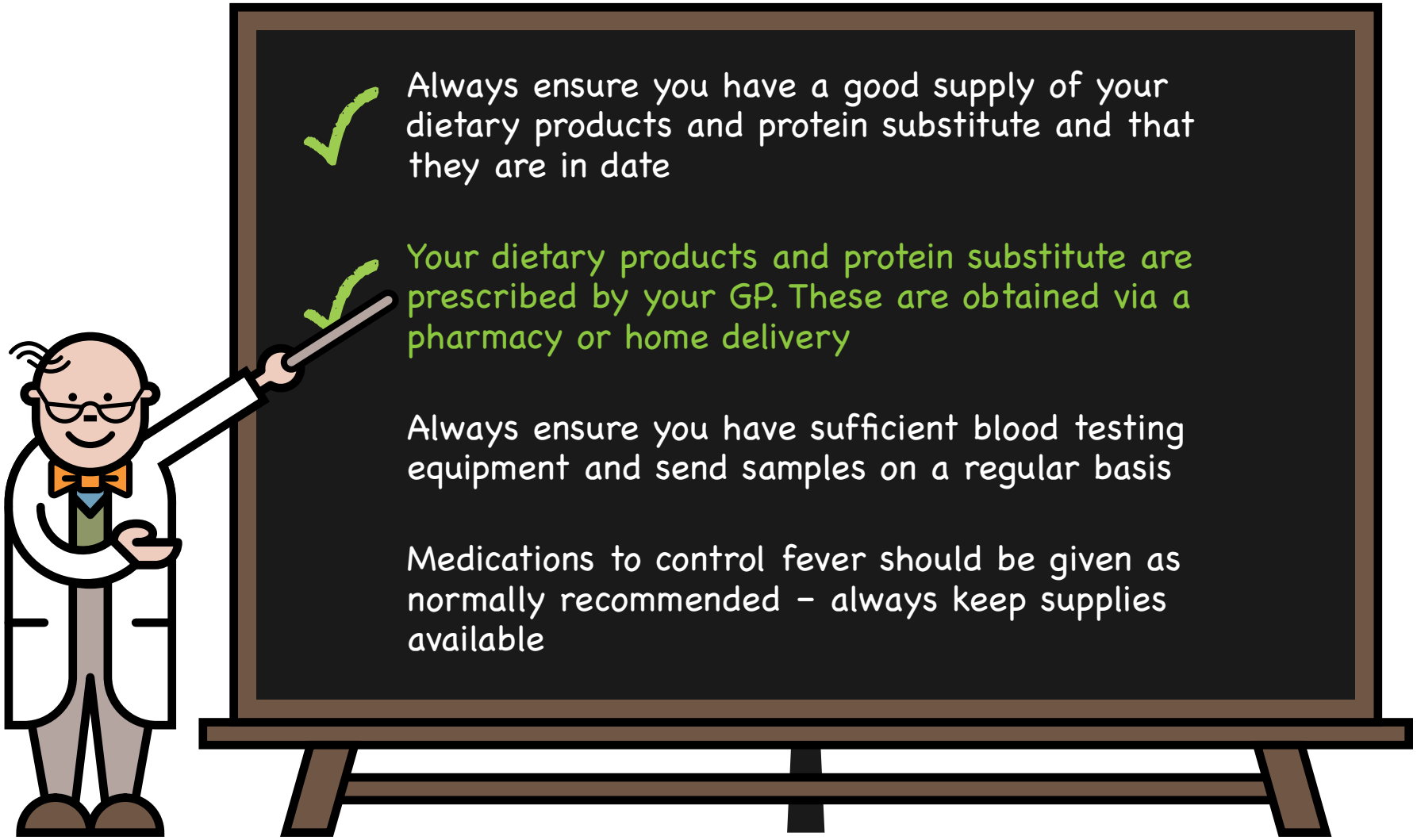

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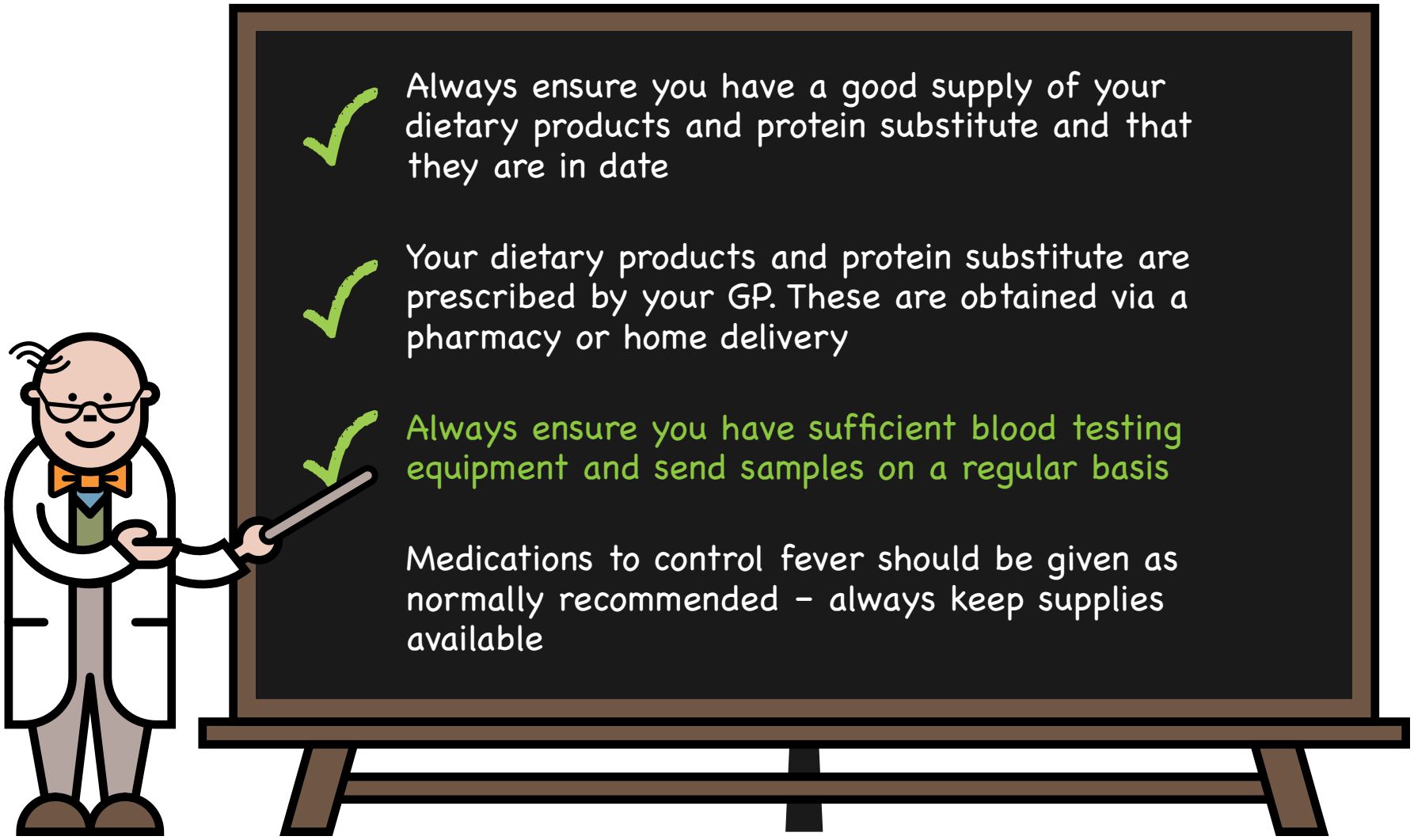

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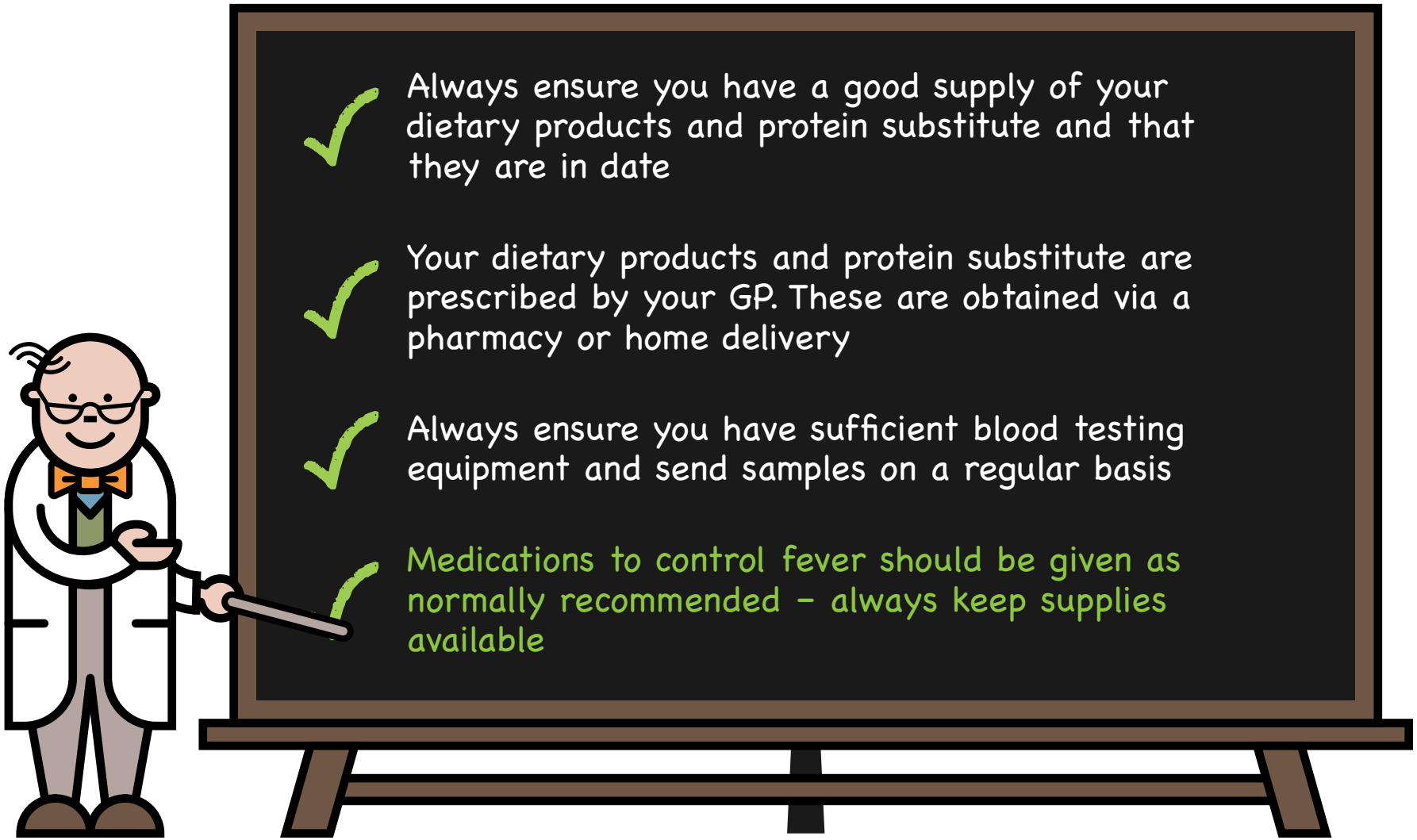

Who's who

- 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.

MET1540UKIR

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

BIMDG
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