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, OCTOBER 2020

TYR II

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.

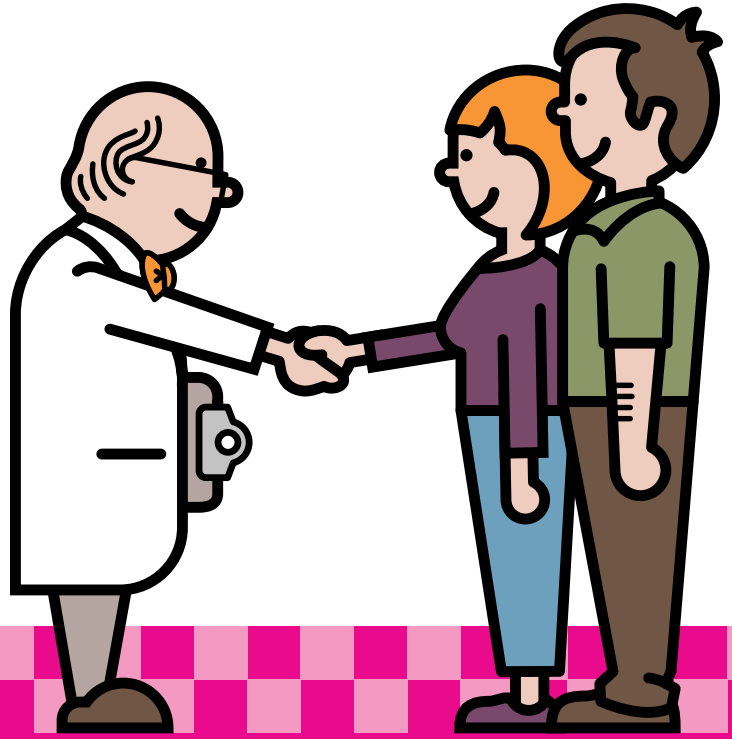
Each module produced is reviewed by a consultant clinician who is a member of the BIMDG.

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

Tyrosinaemia Type II (TYR II)

Information for families following a new diagnosis



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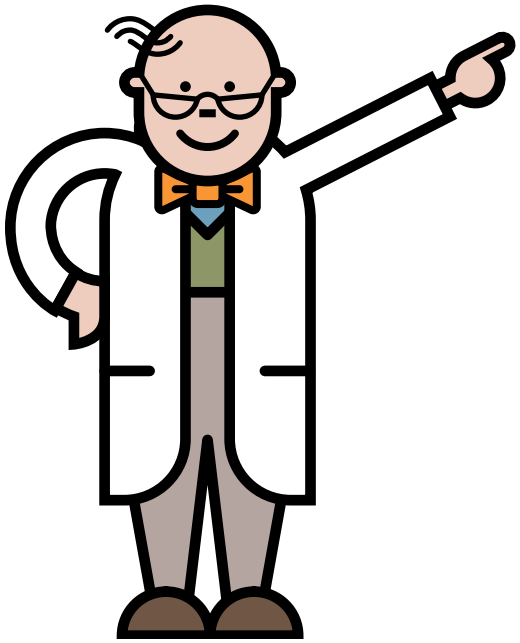
What is TYR II?

TYR II stands for Tyrosinaemia Type II

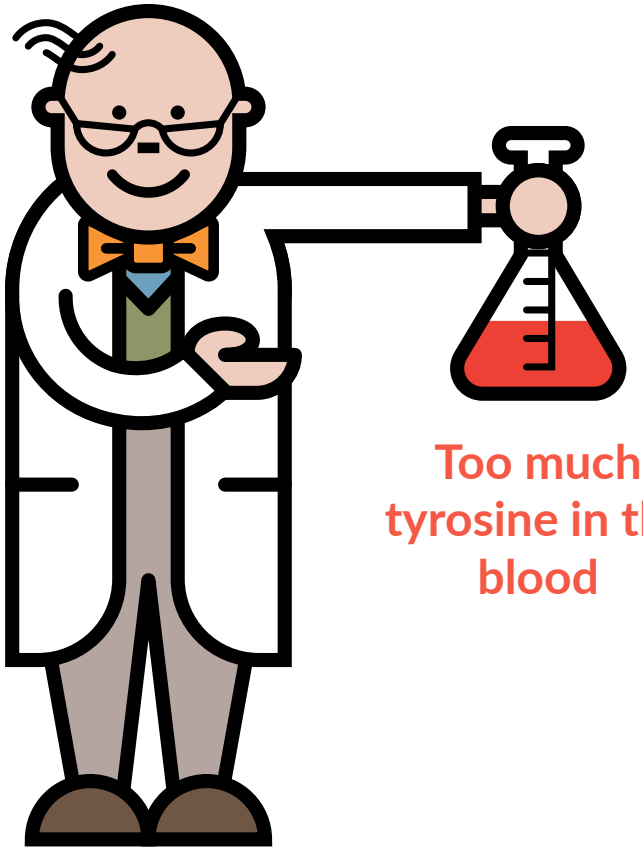
It is an inherited metabolic condition



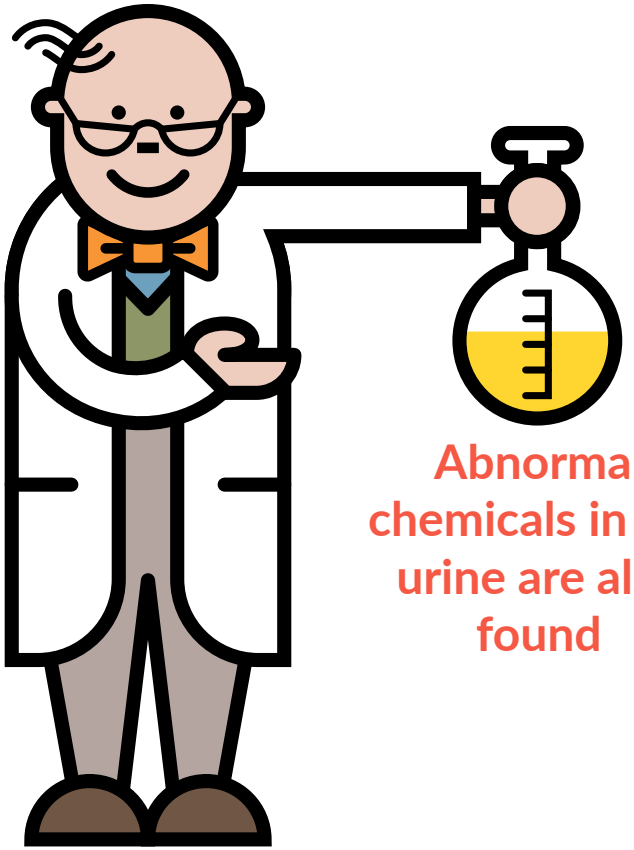
Tyrosinaemia Type 2

TYR II

What is TYR II?



Too much tyrosine in the blood



Abnormal chemicals in the urine are also found

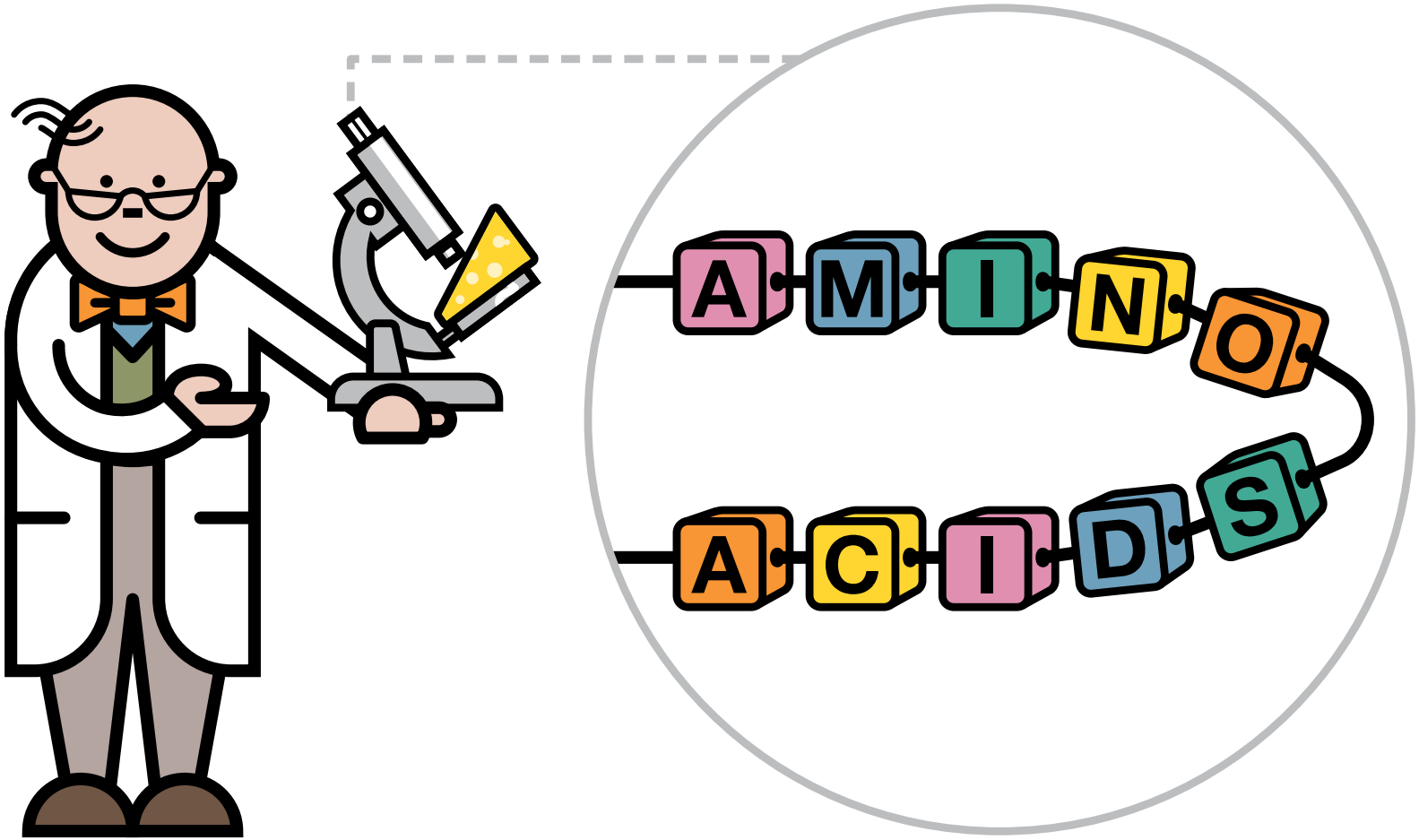
TYR II and protein

TYR II affects the way your baby breaks down protein

Many foods contain protein

The body needs protein for growth and repair

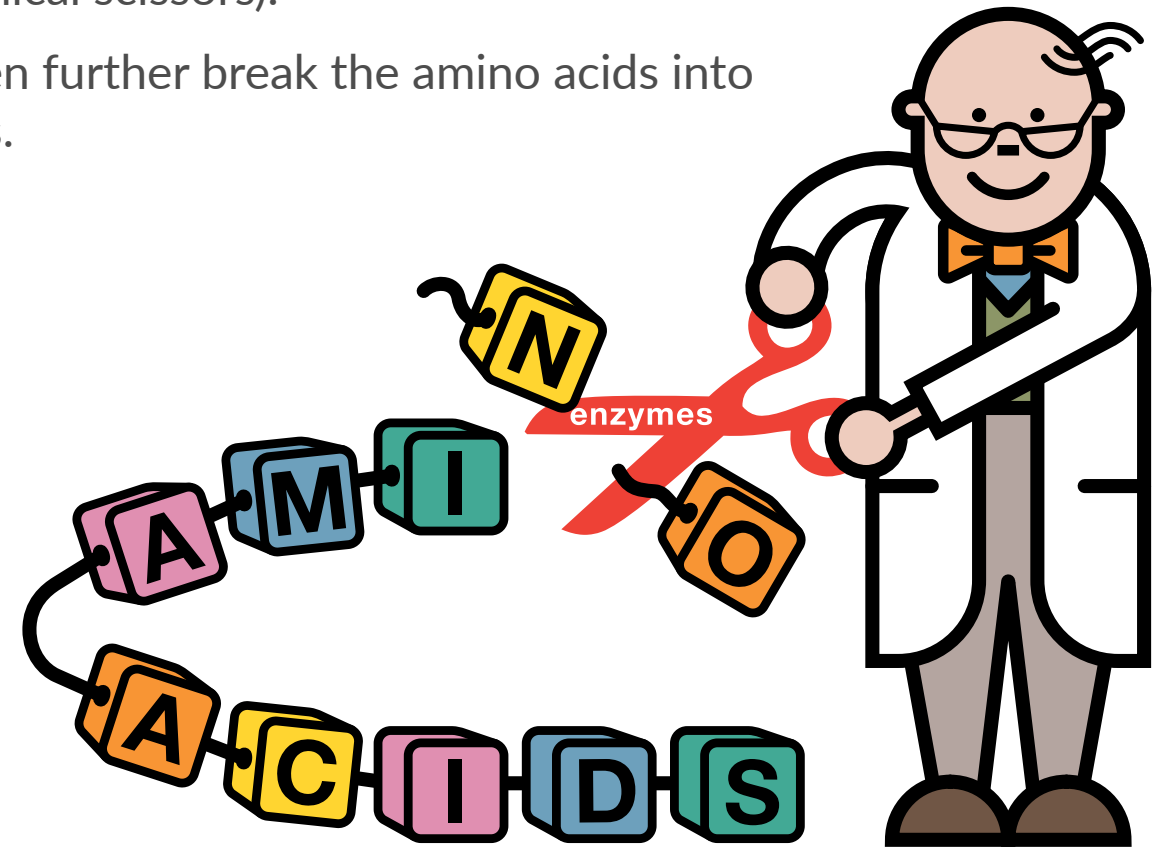
What is protein?



Protein and enzymes

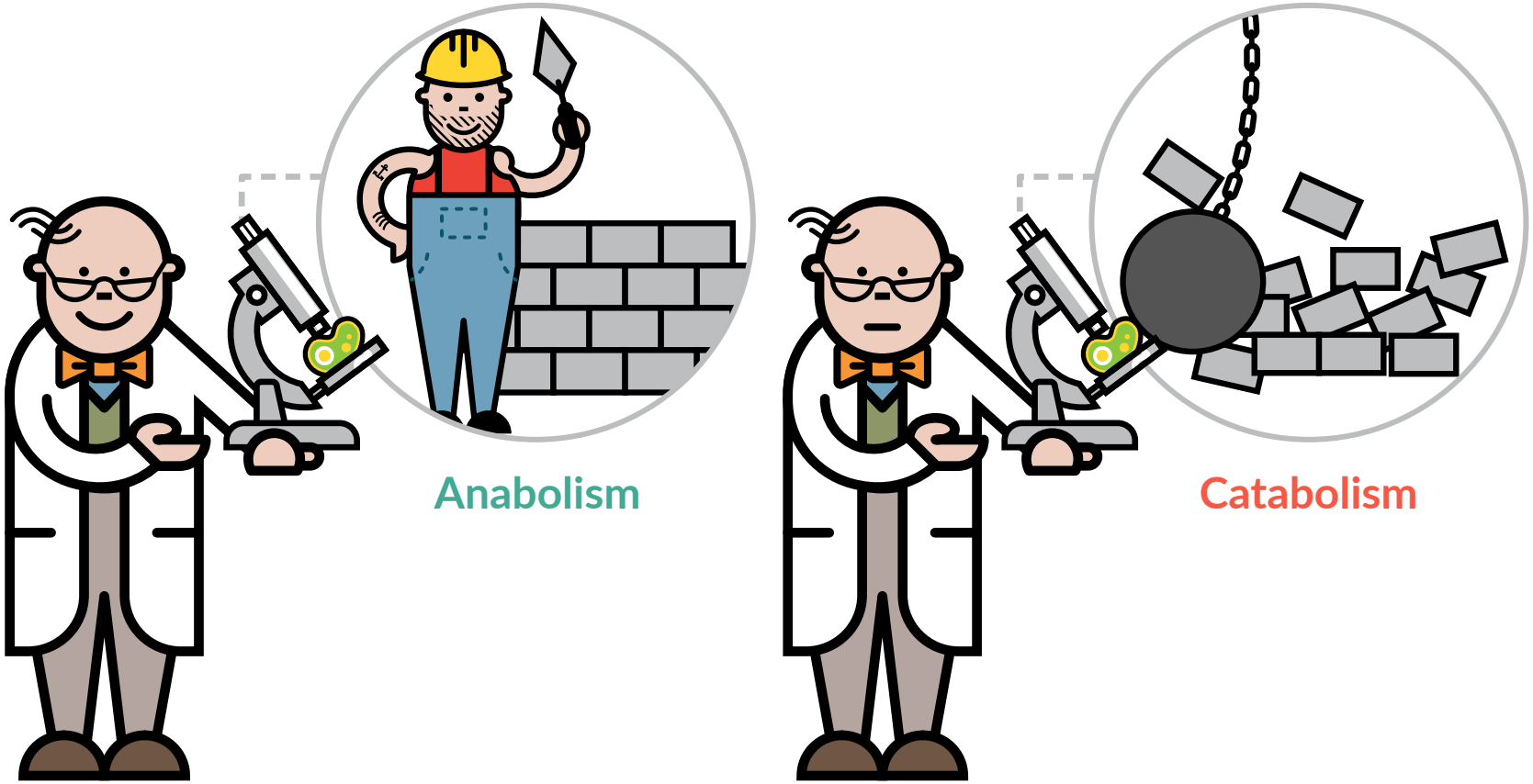
Protein is broken down into amino acids (building blocks of protein) by enzymes (which are like chemical scissors).

Enzymes then further break the amino acids into smaller parts.



Protein metabolism

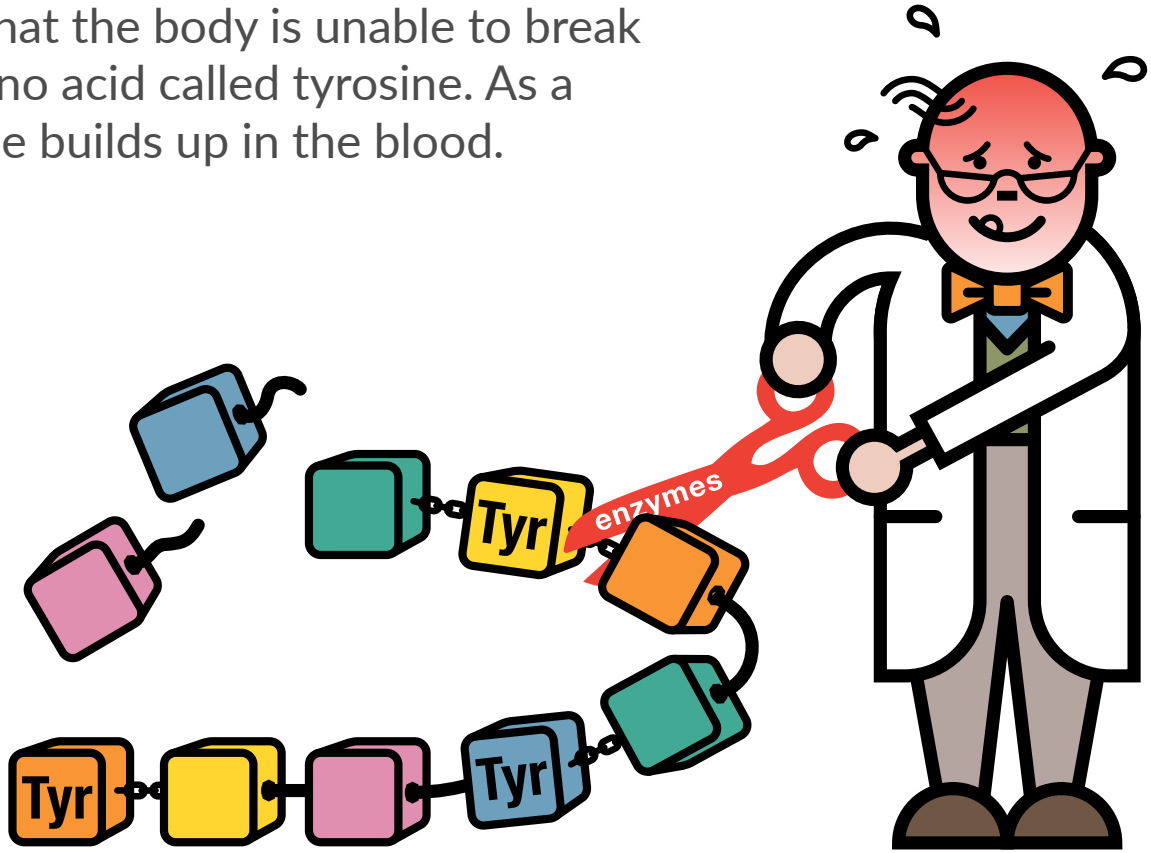
Metabolism refers to the chemical processes that occur inside the cells of the body.



What happens in TYR II?

In TYR II, the body lacks an enzyme called **tyrosine aminotransferase (TAT)**.

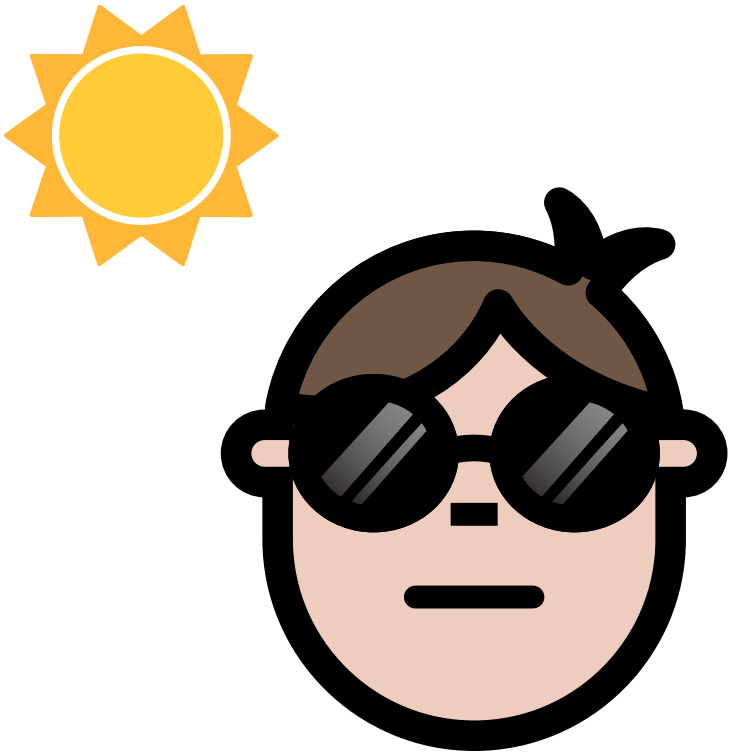
This means that the body is unable to break down an amino acid called tyrosine. As a result tyrosine builds up in the blood.



What are the signs in TYR II?

Signs of TYR II usually begin in the first year of life but can occur at any age. Signs may include:

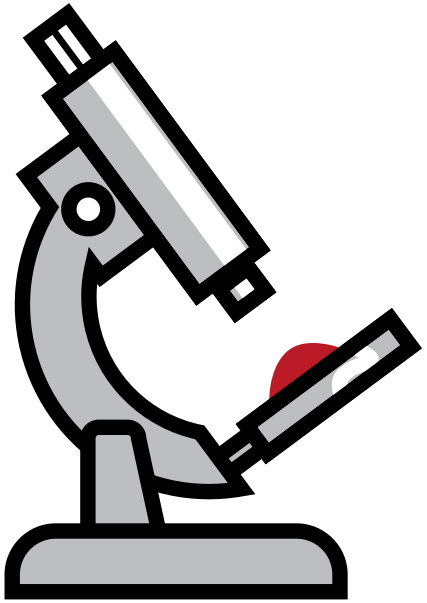
- Sensitivity to light (called photophobia)
- Eye redness
- Skin lesions on the hands and feet
- Behaviour problems
- Learning difficulties



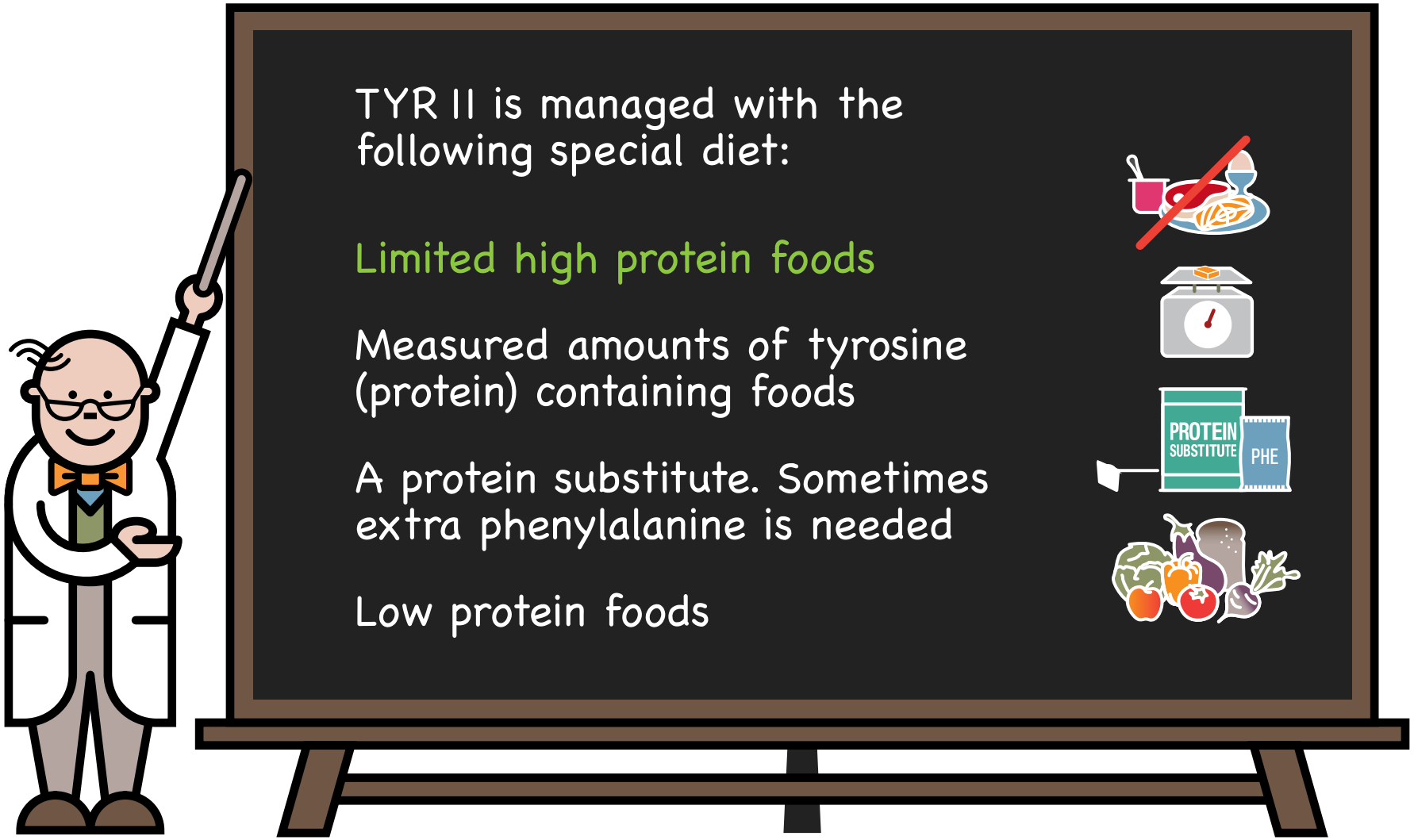
How is TYR II diagnosed?

TYR II is diagnosed by high levels of tyrosine in the blood and abnormal chemicals in the urine.

The diagnosis is confirmed by finding the mutation in the TAT gene.



How is TYR II managed?



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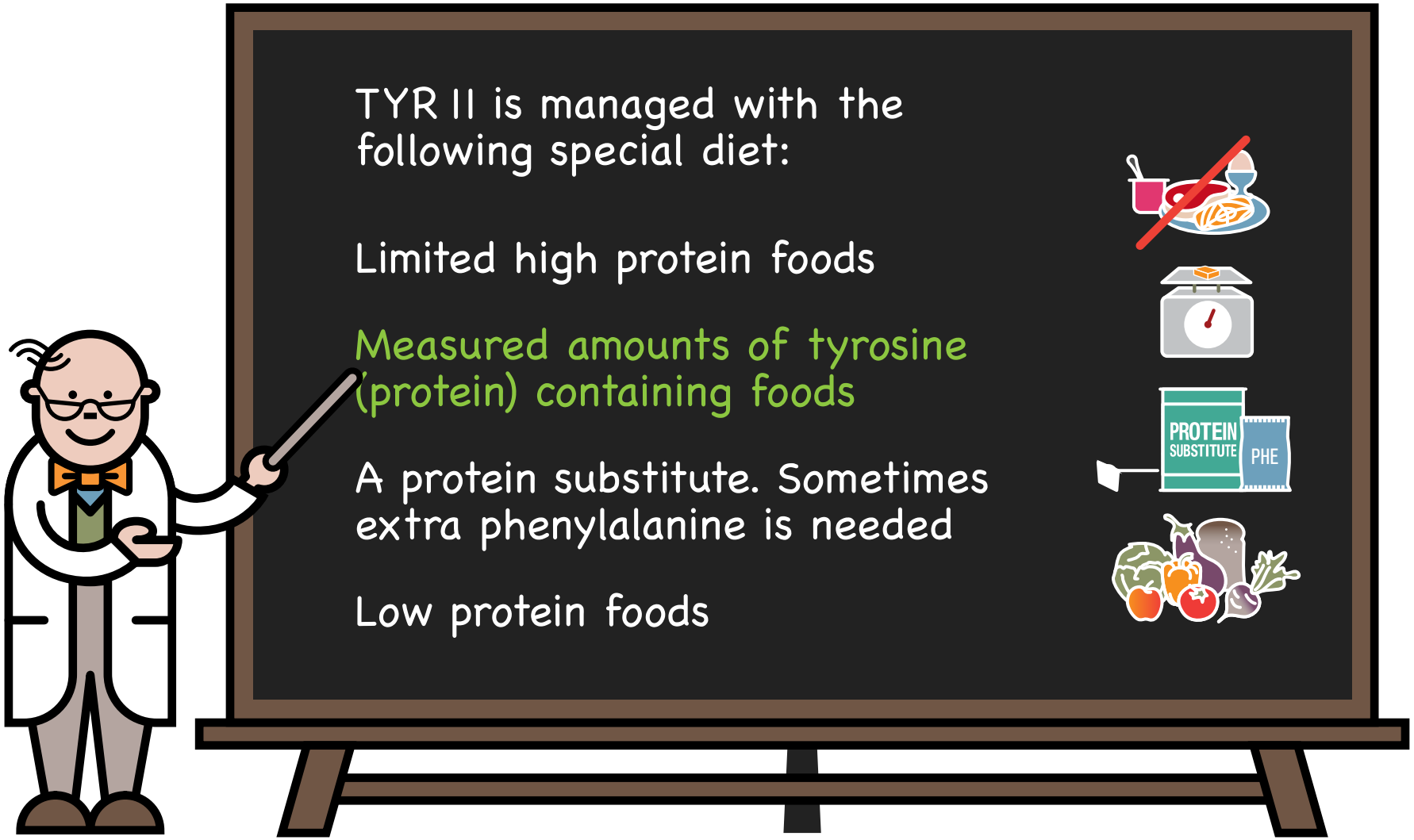

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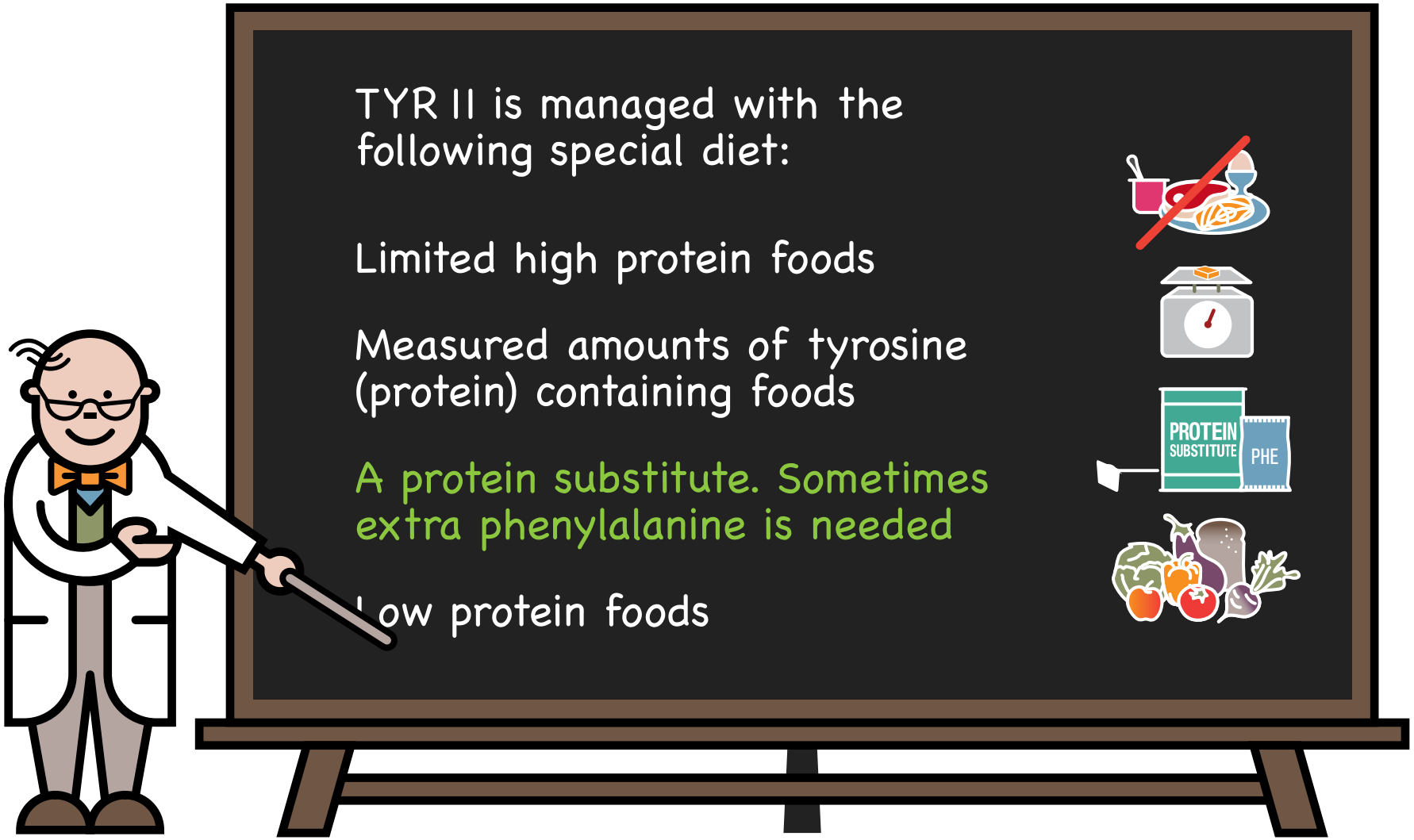

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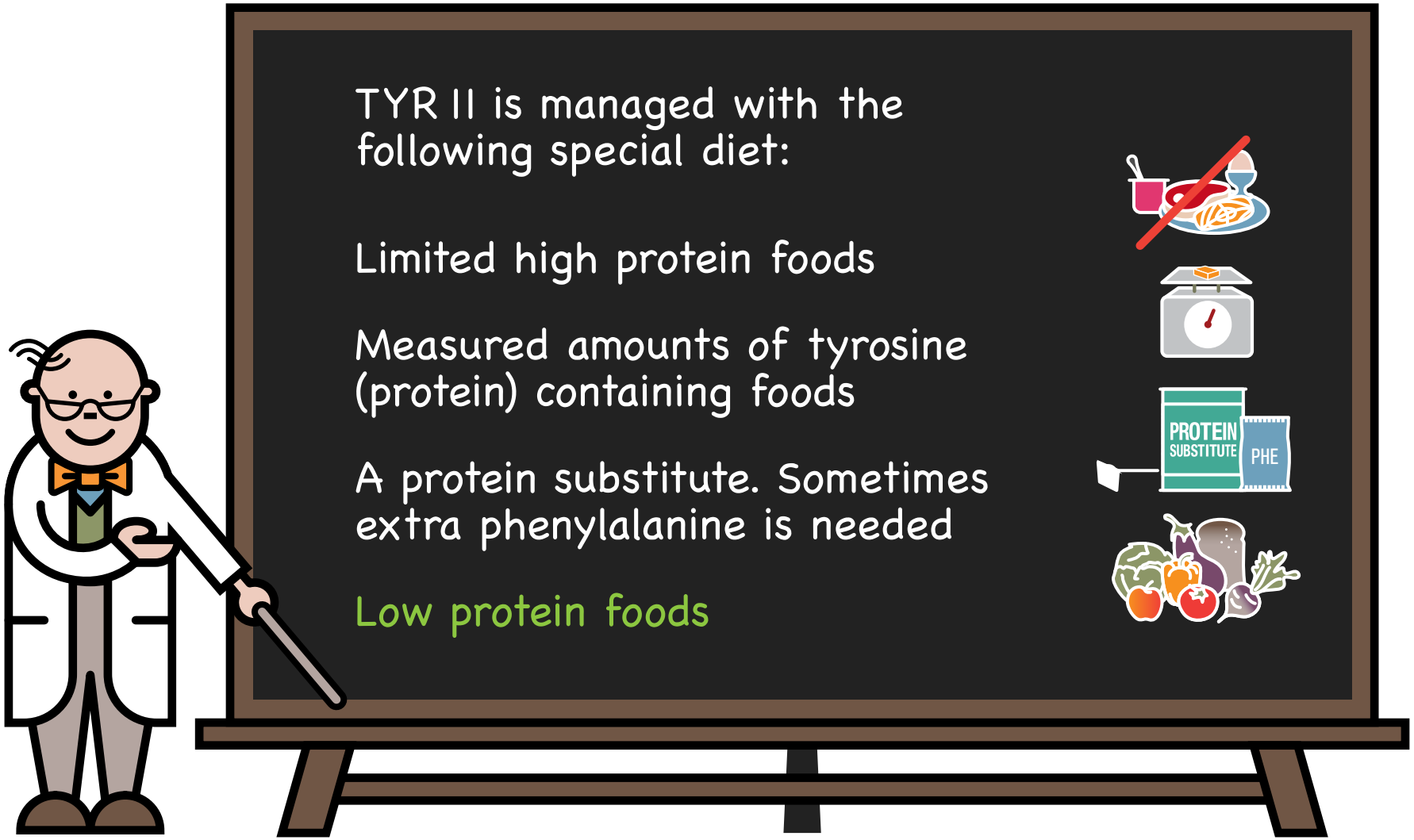


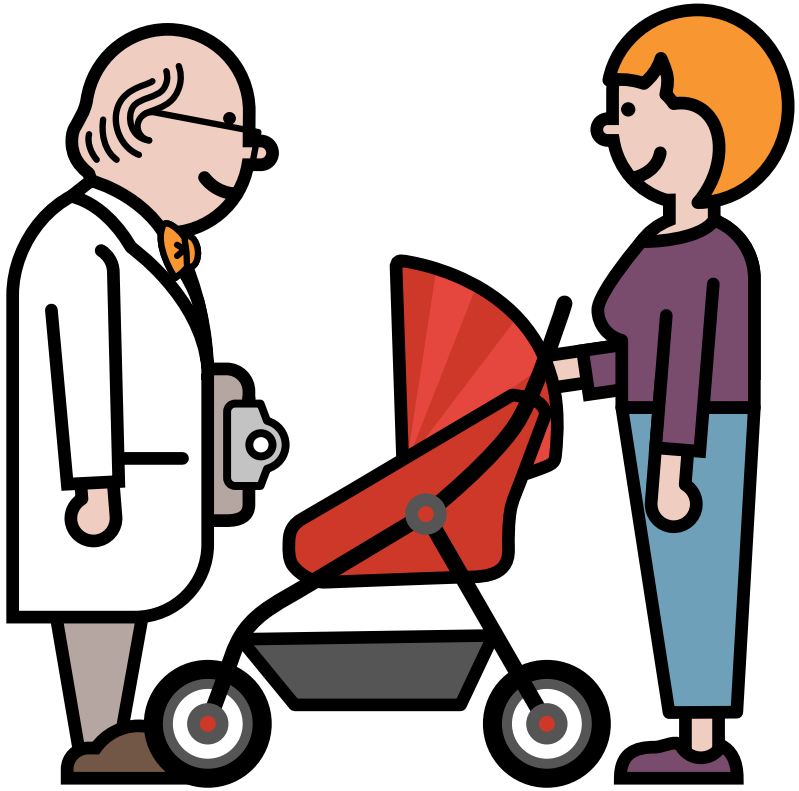
High protein foods

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

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.

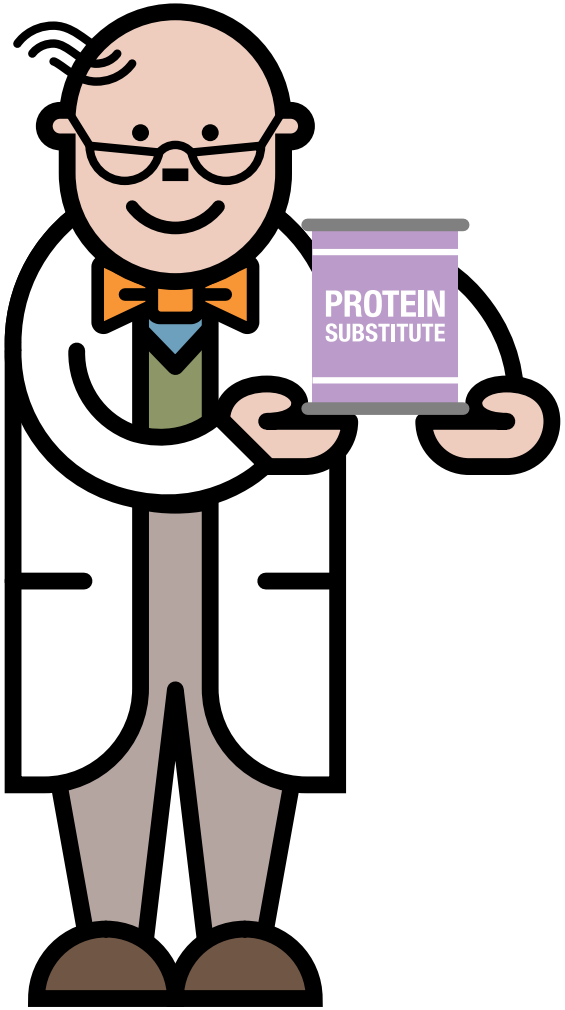


Protein substitute

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.

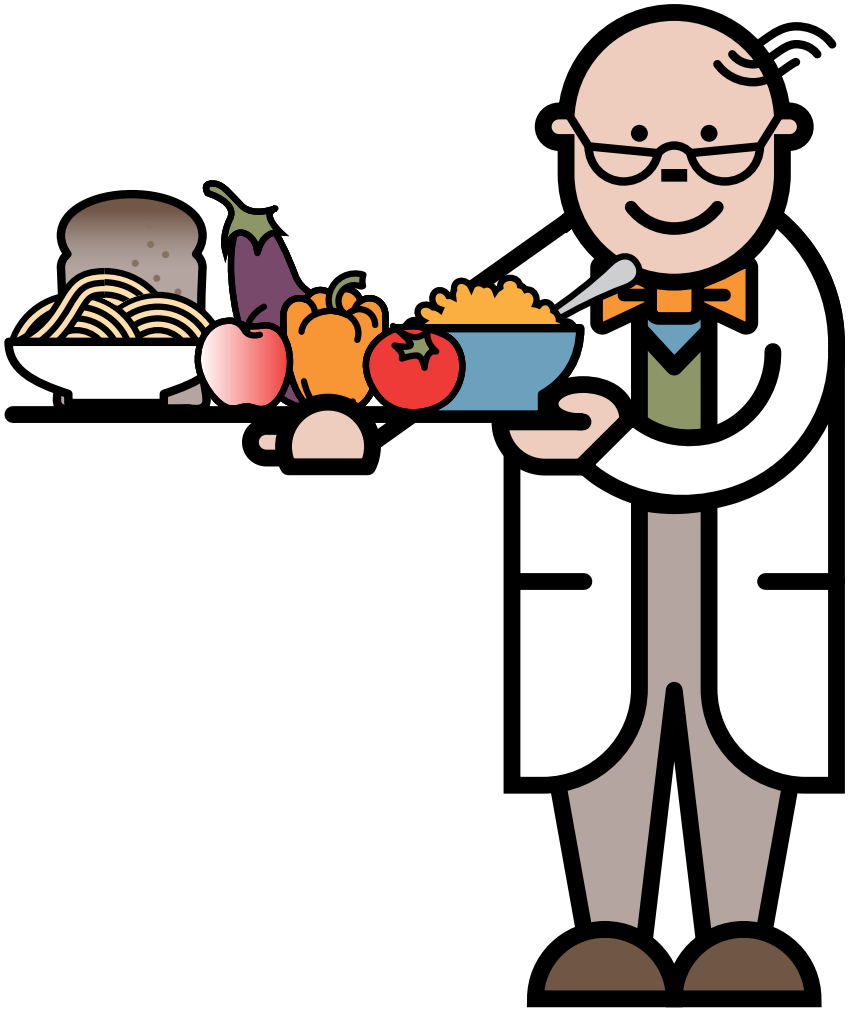


Low protein foods

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

They provide:

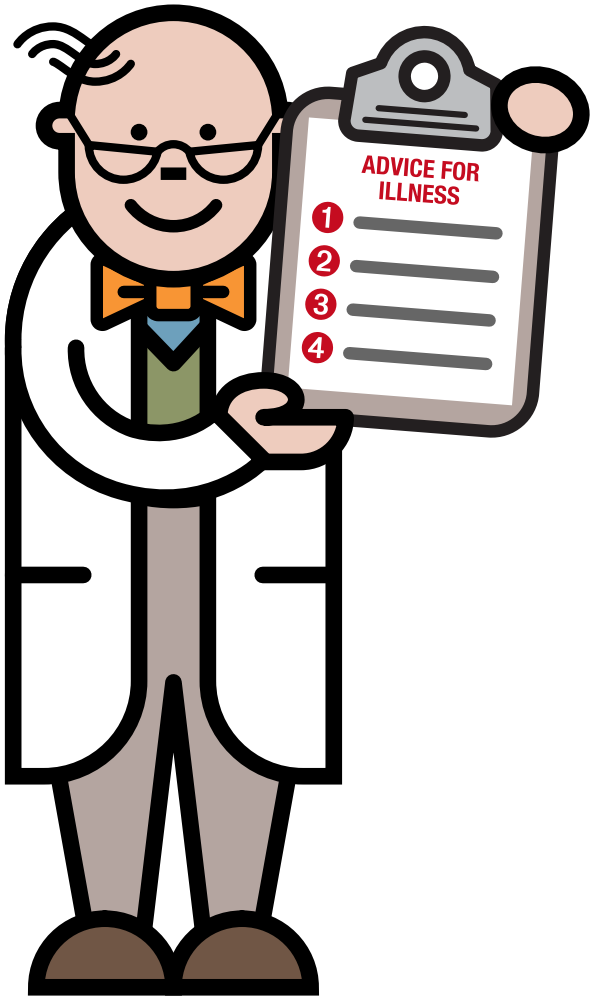
- a source of energy
- variety in the diet



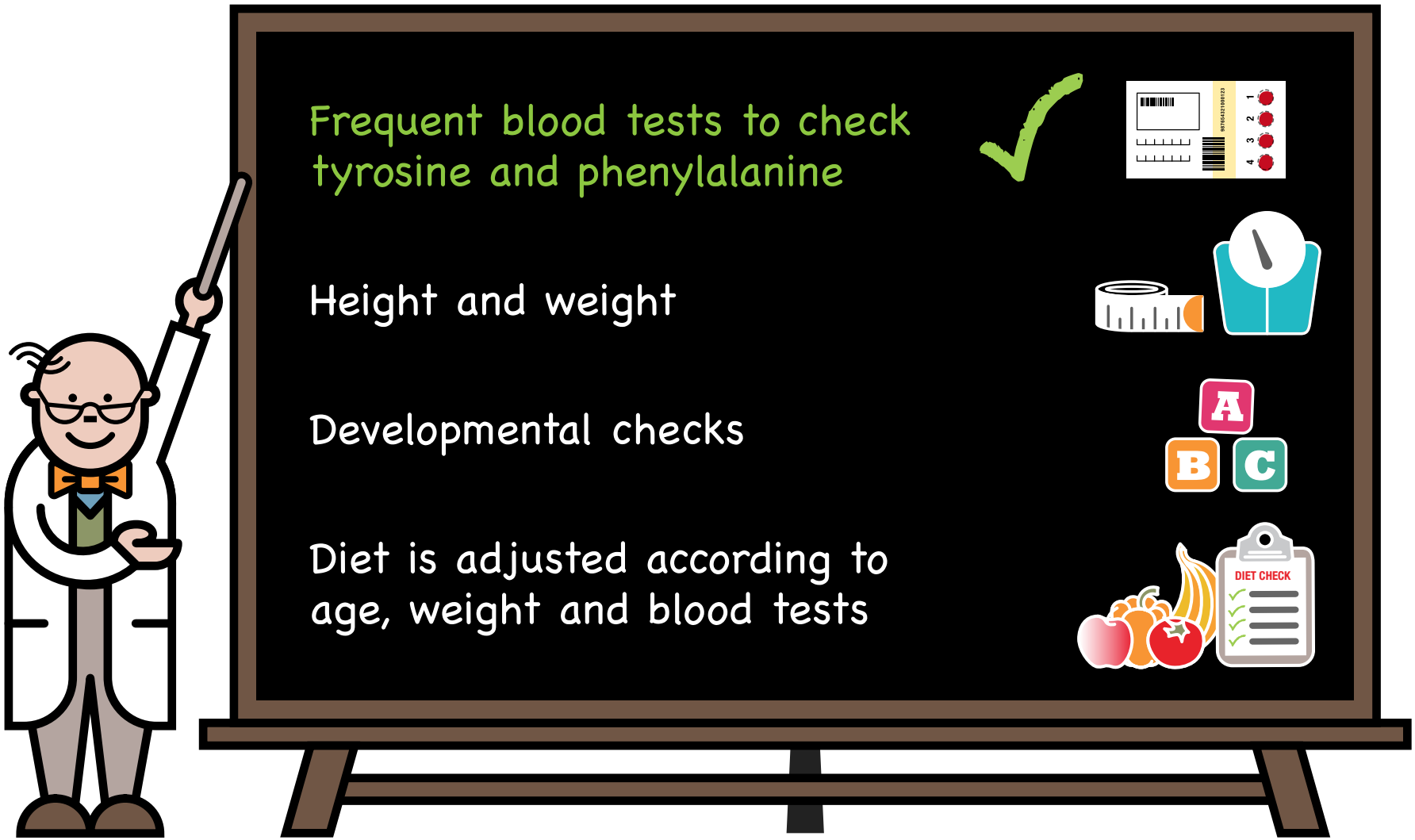
How is TYR II managed during illness?

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

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



How is TYR II monitored?



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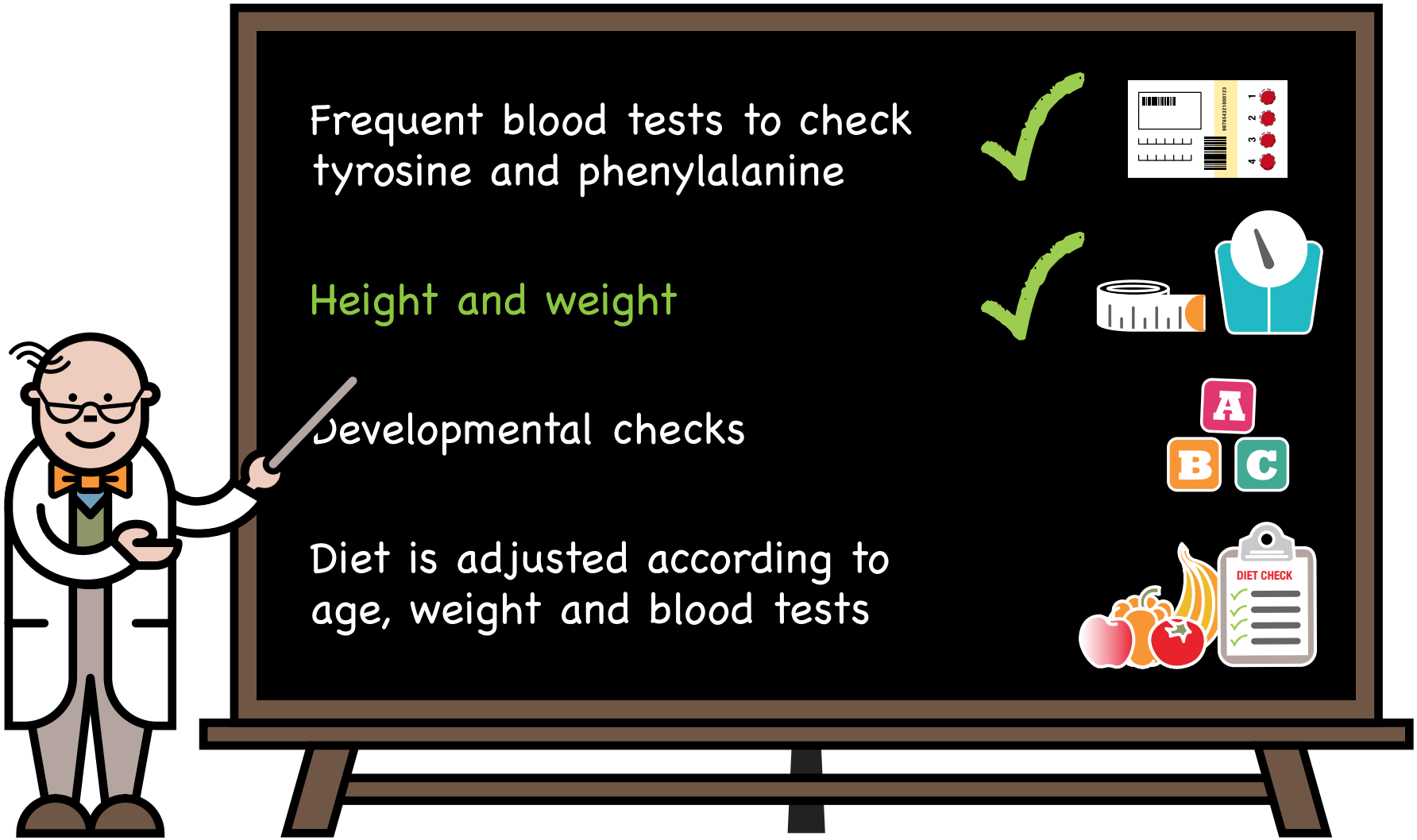

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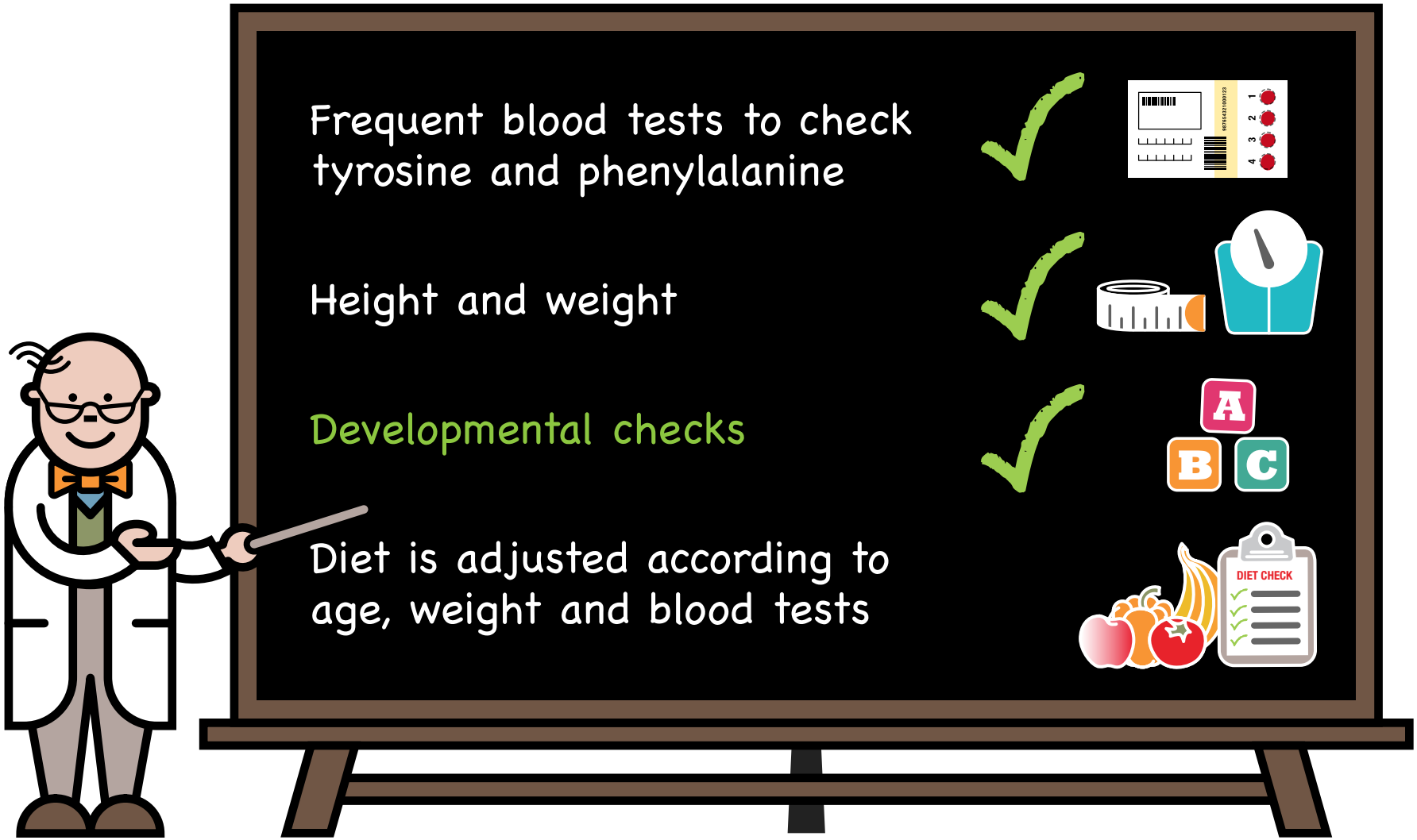

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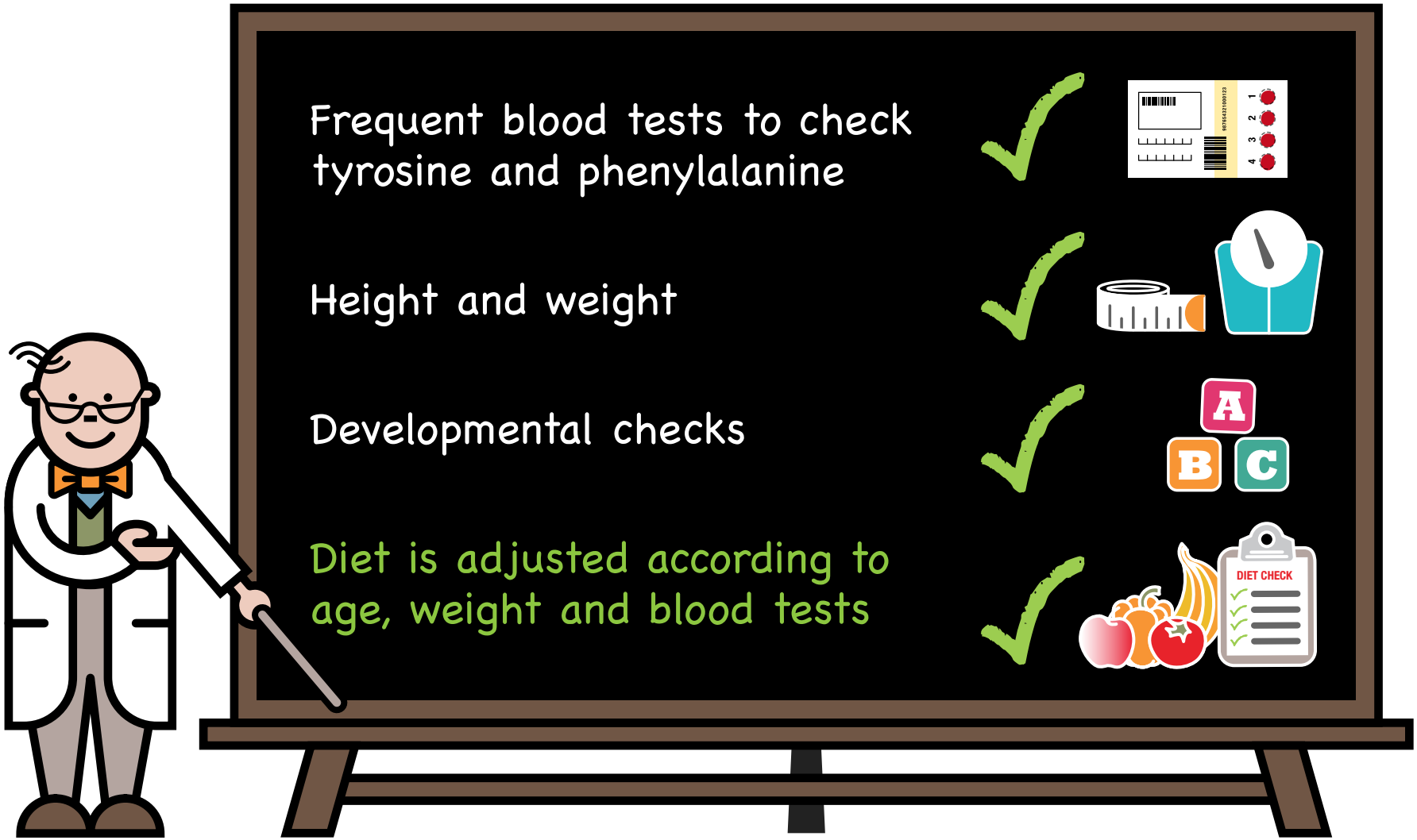
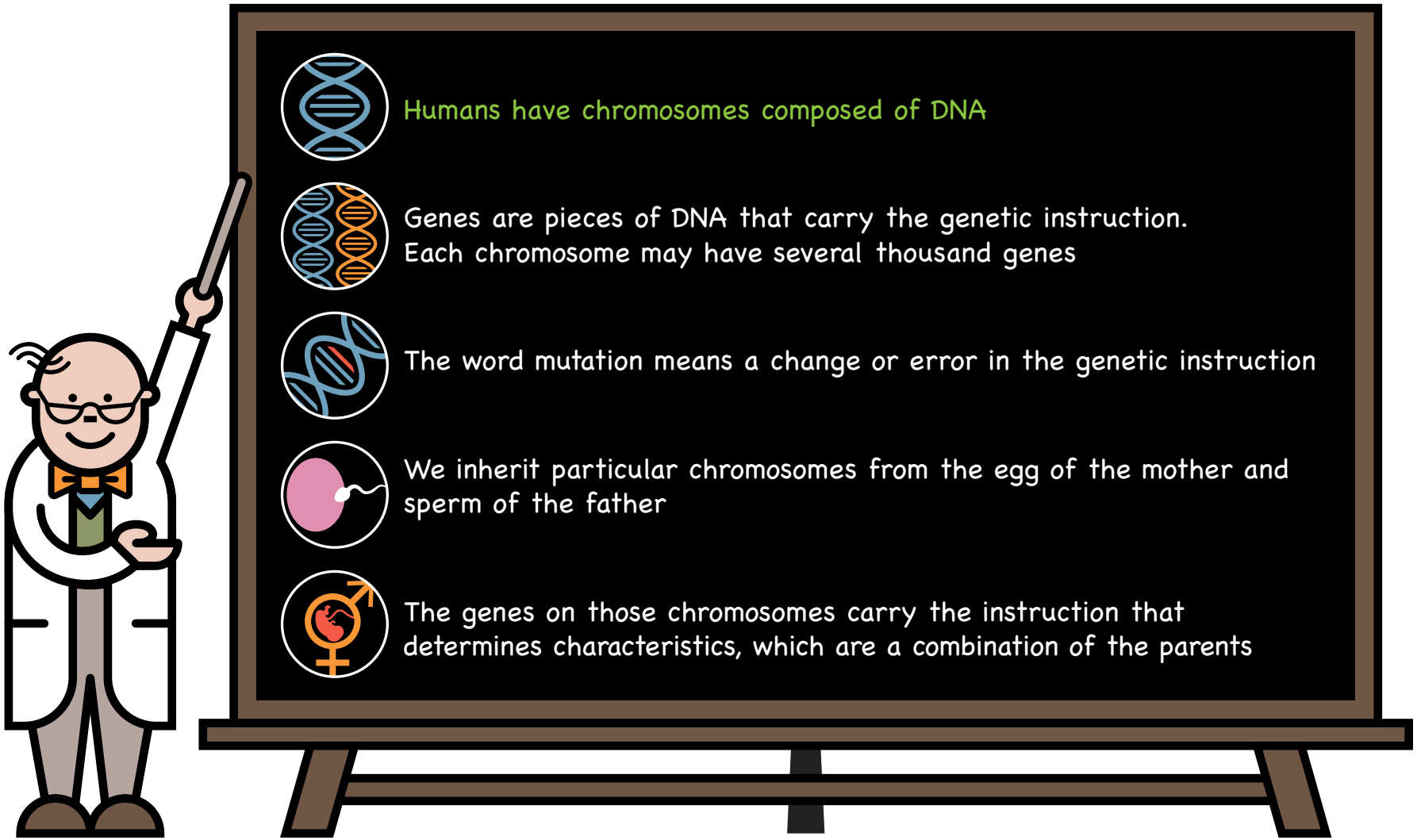


Chromosomes, genes, mutations



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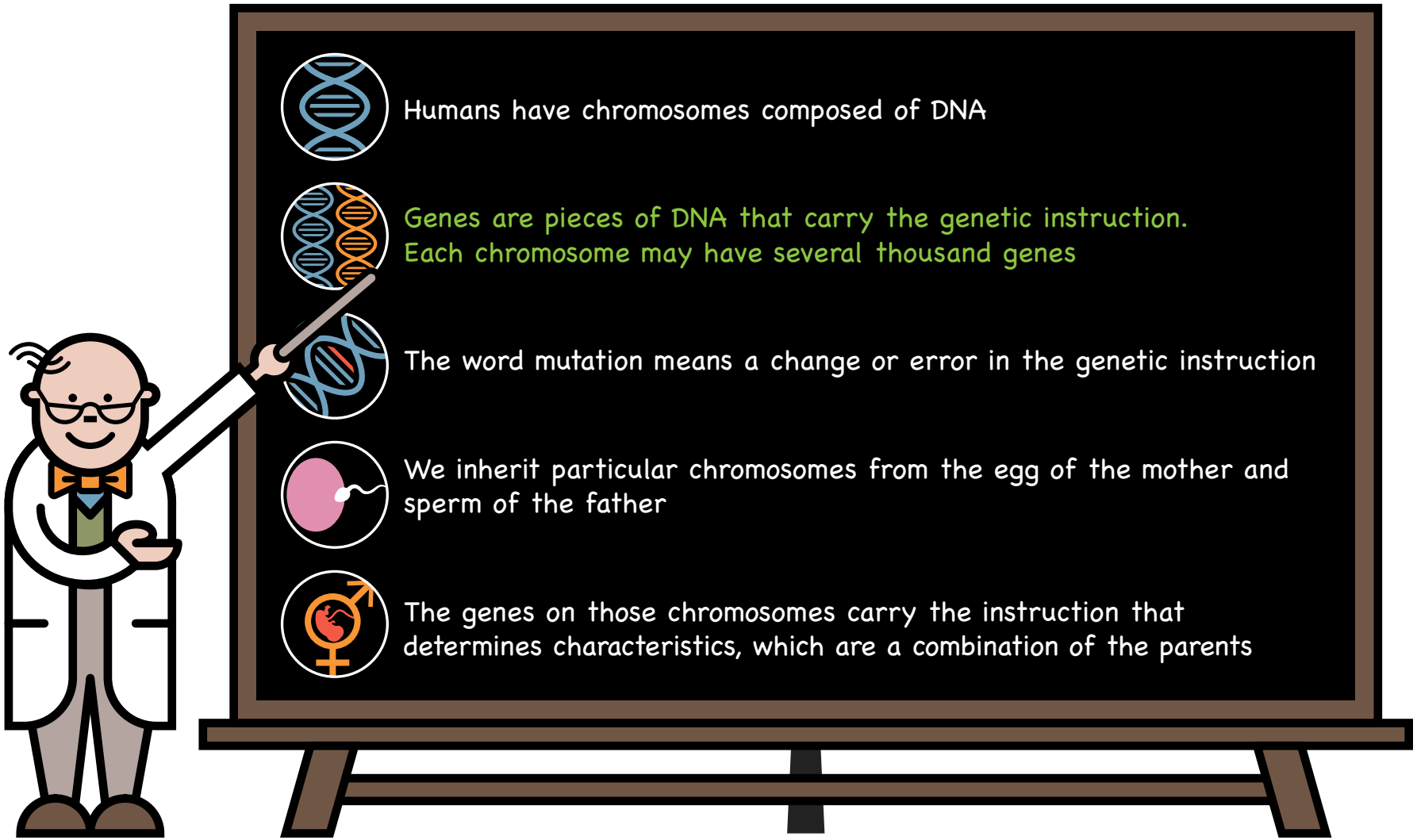
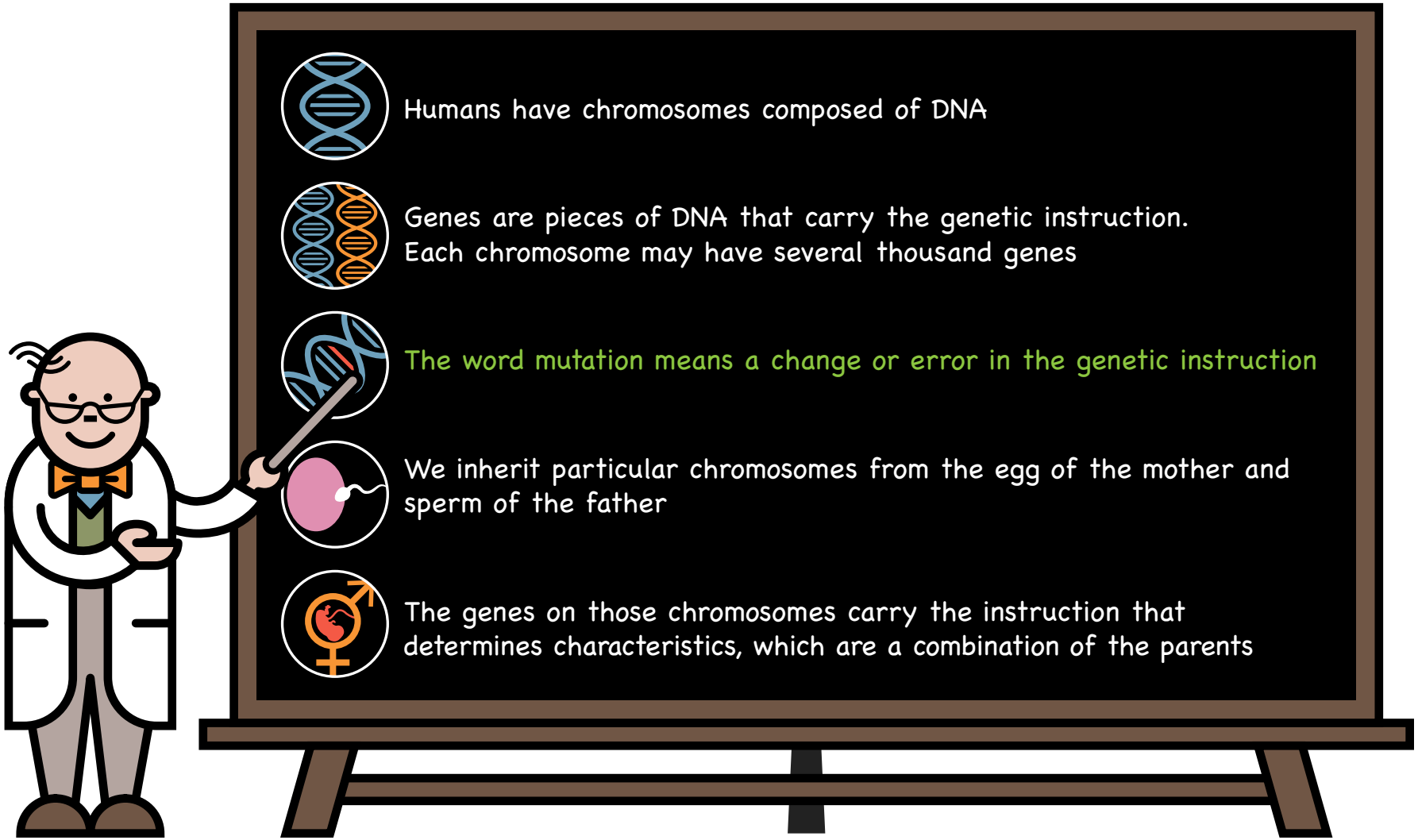
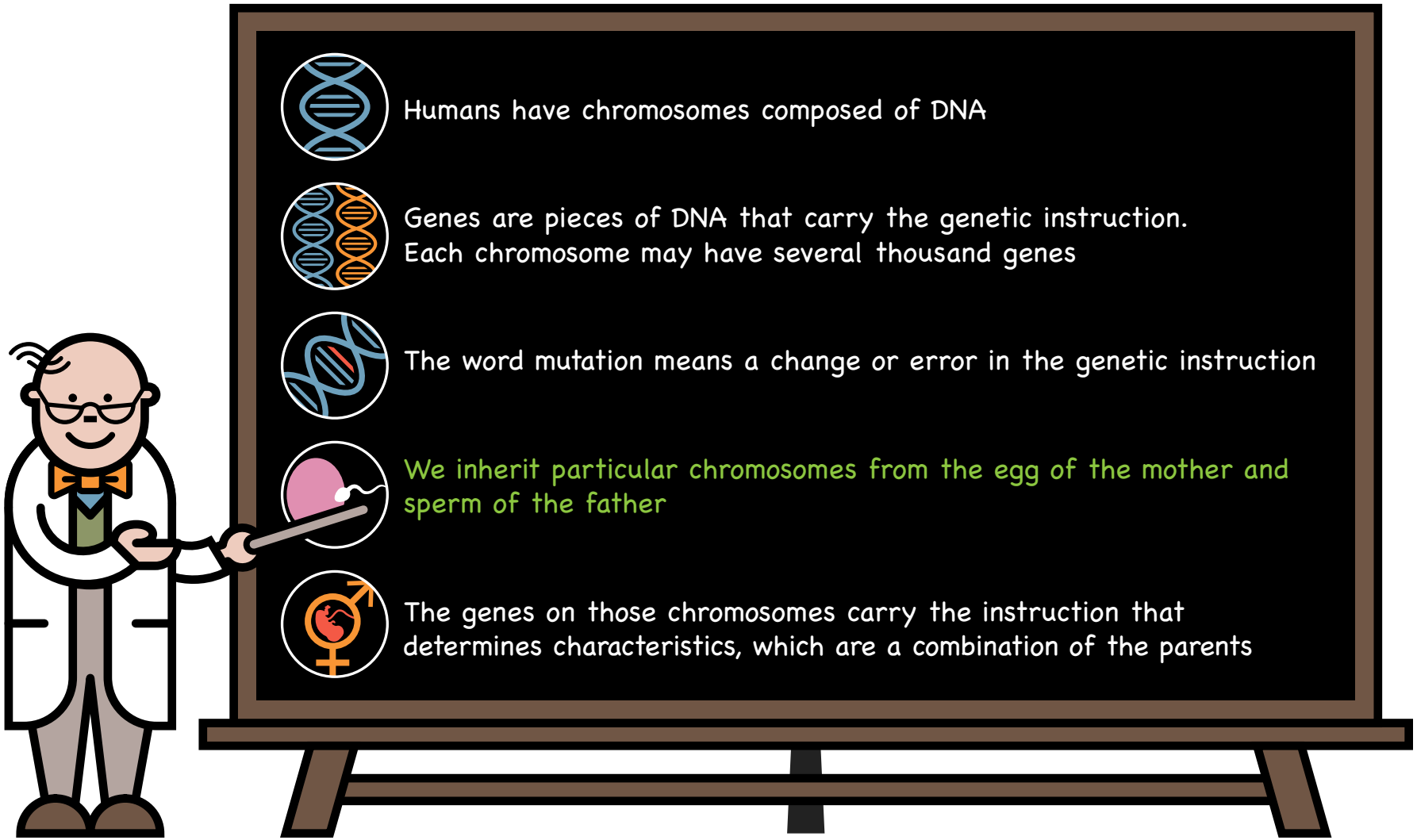


Chromosomes, genes, mutations



Chromosomes, genes, mutations



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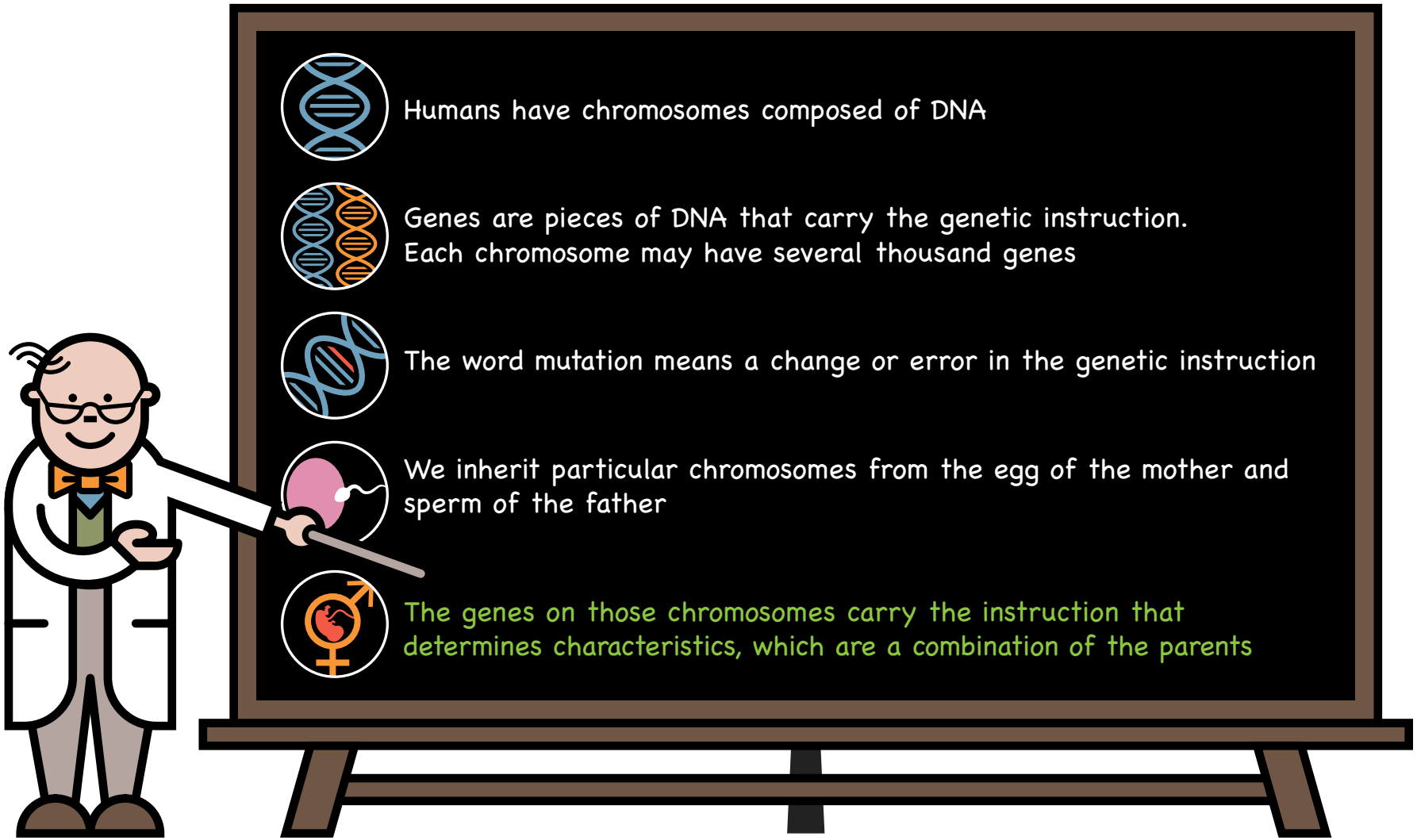
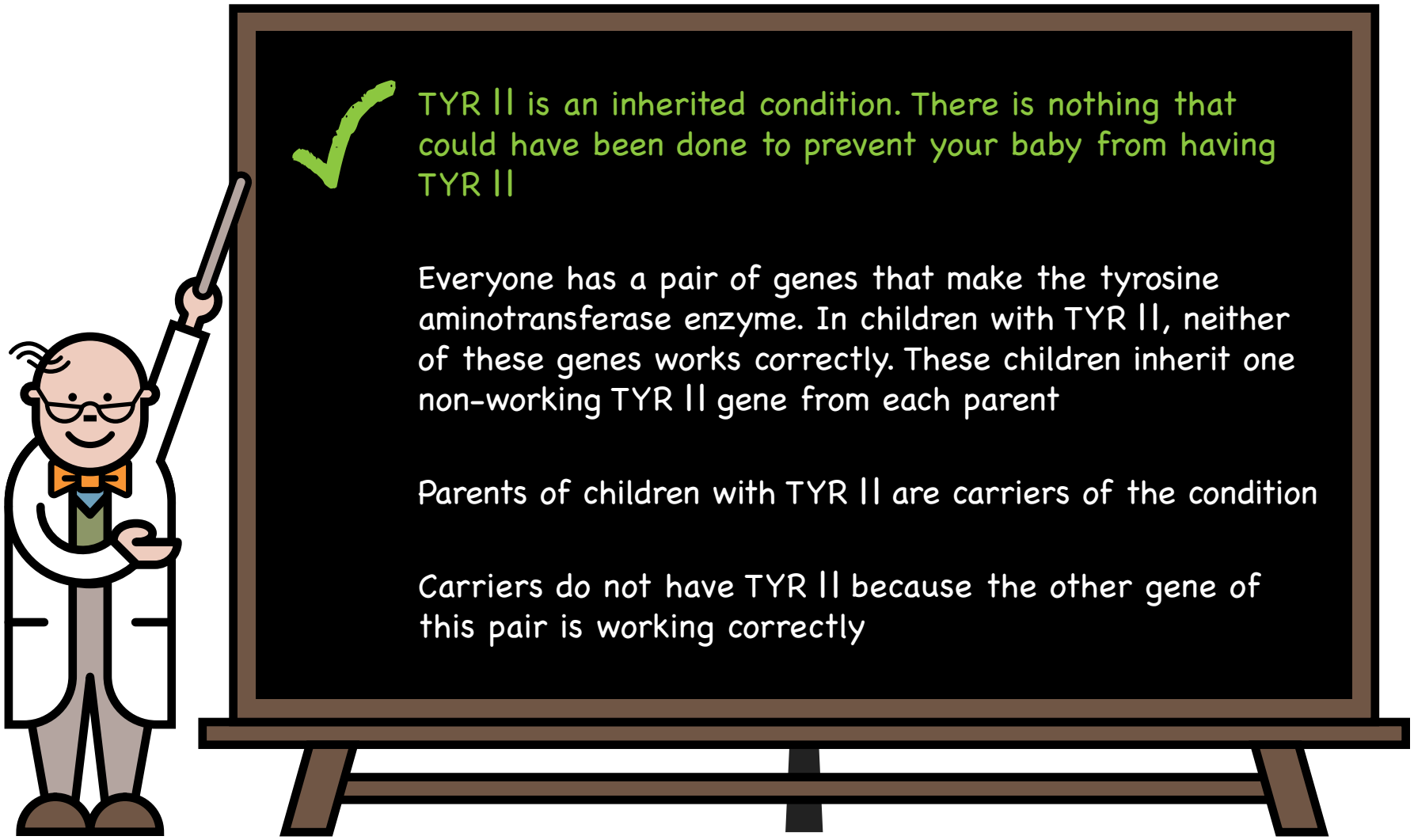


Inheritance



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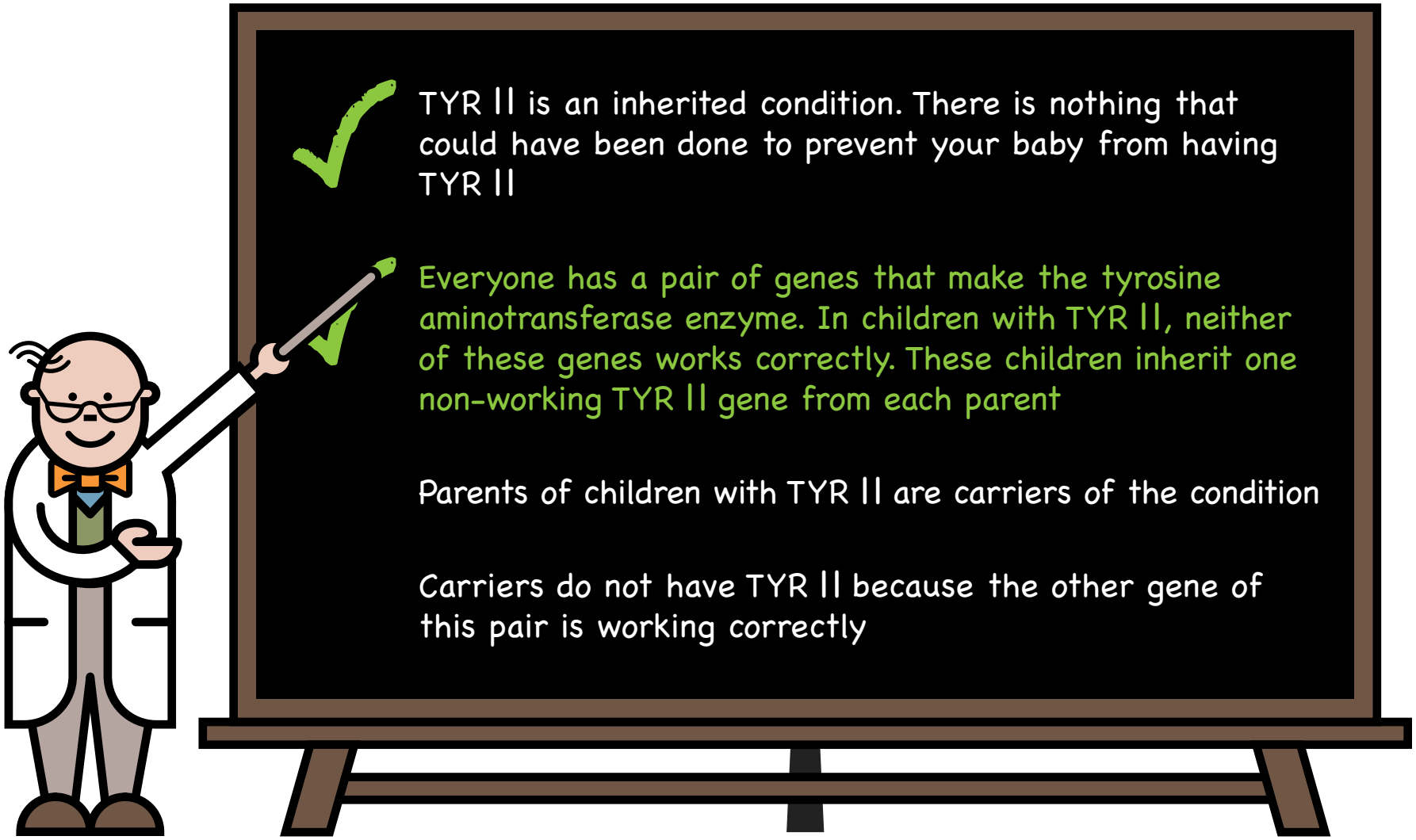

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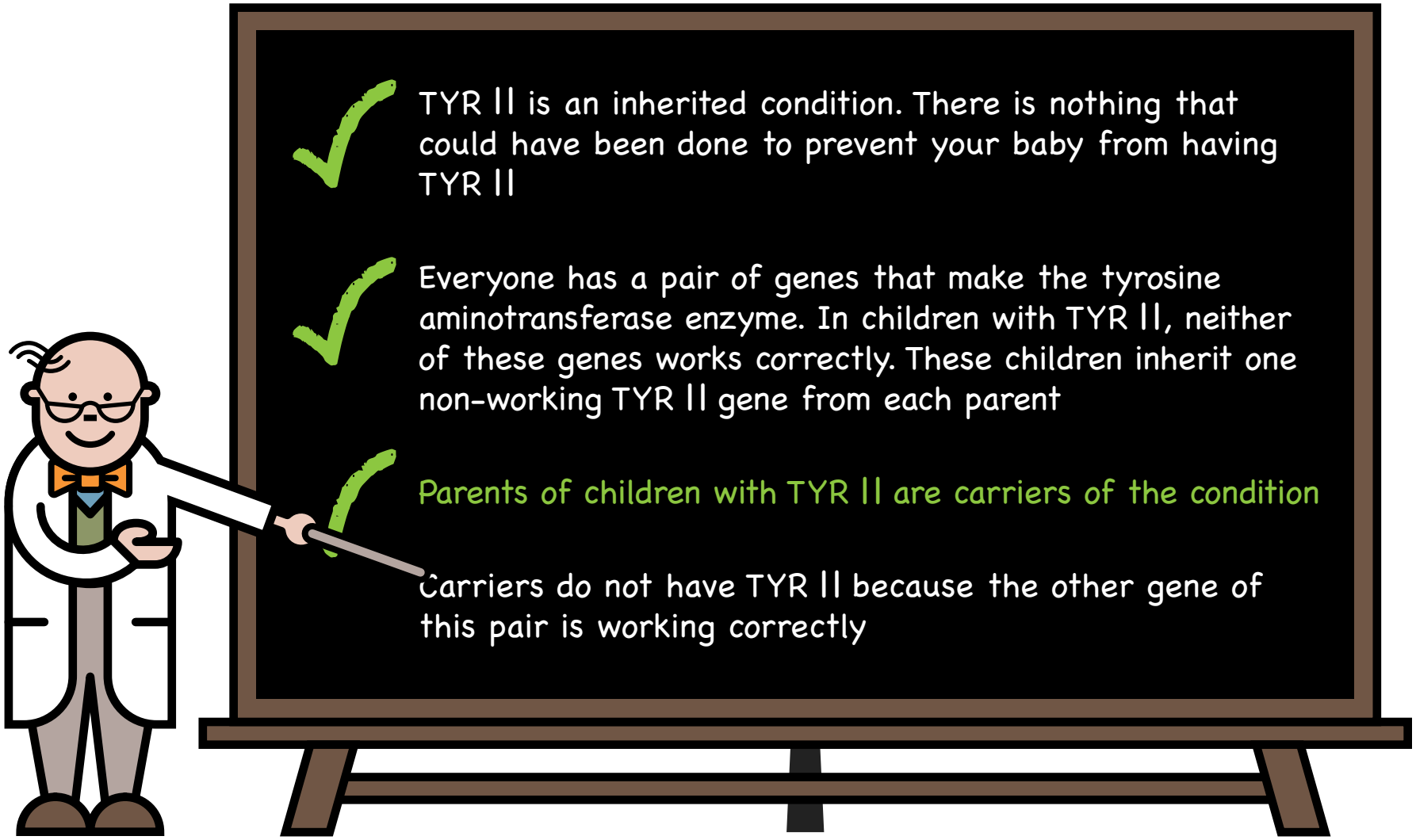

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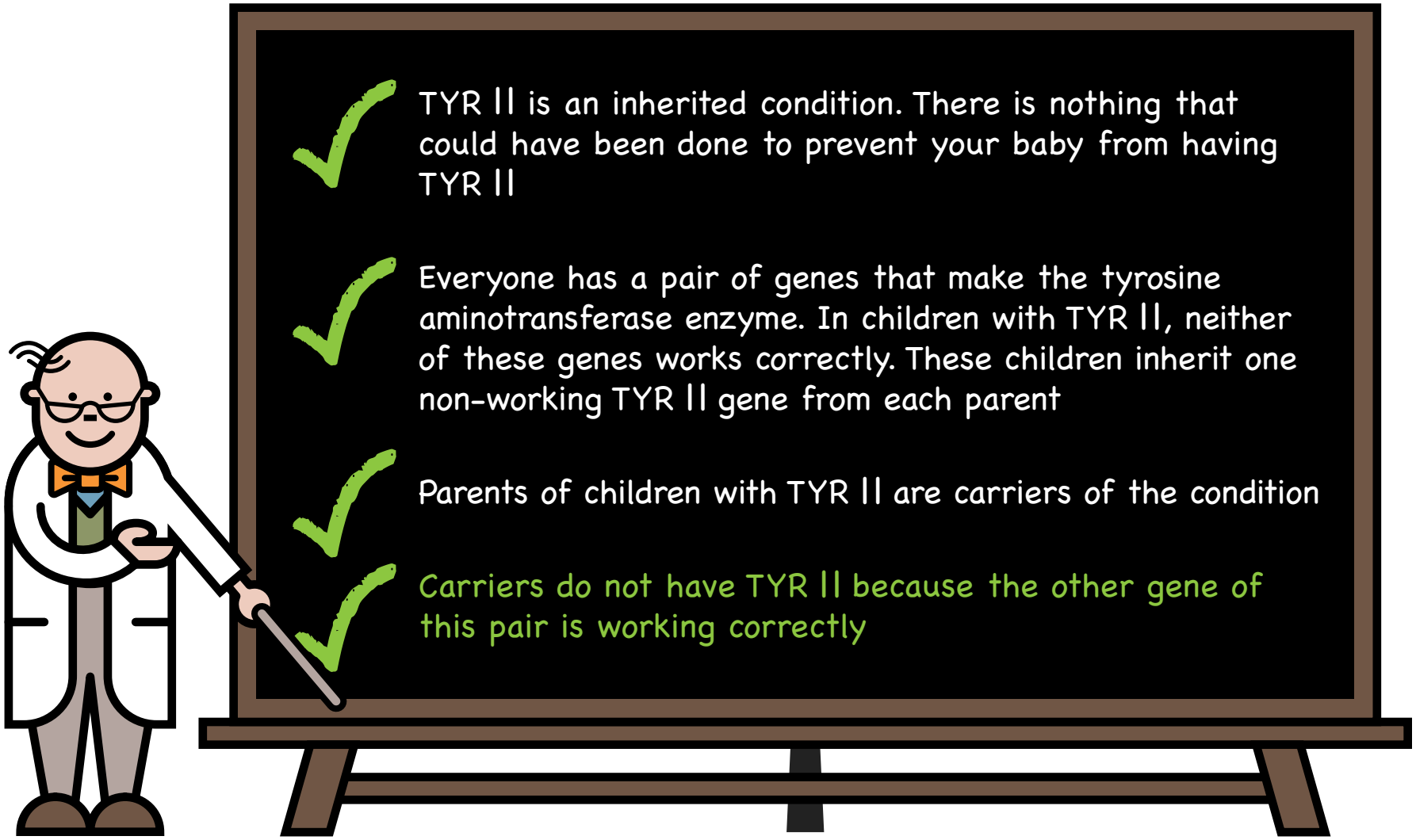
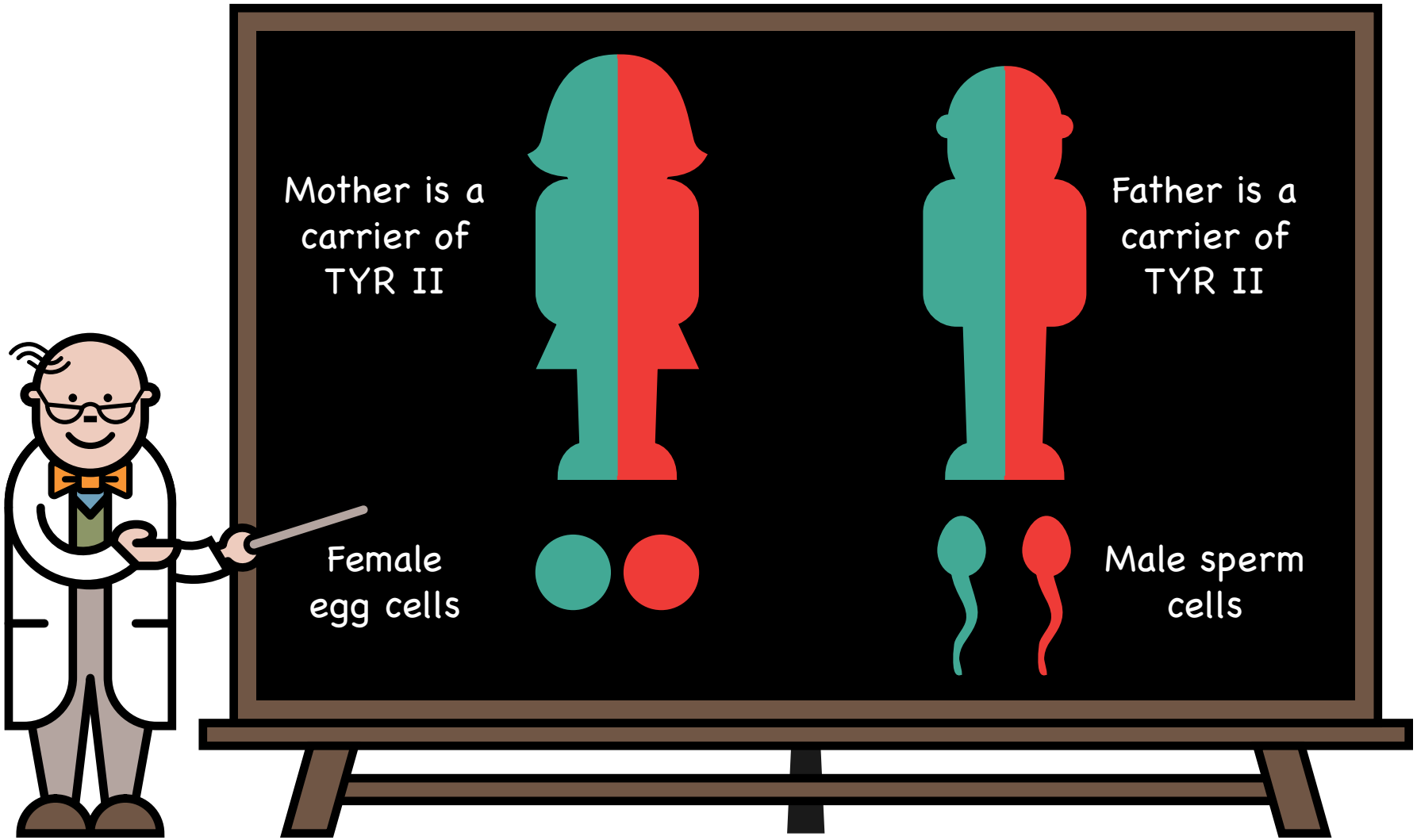
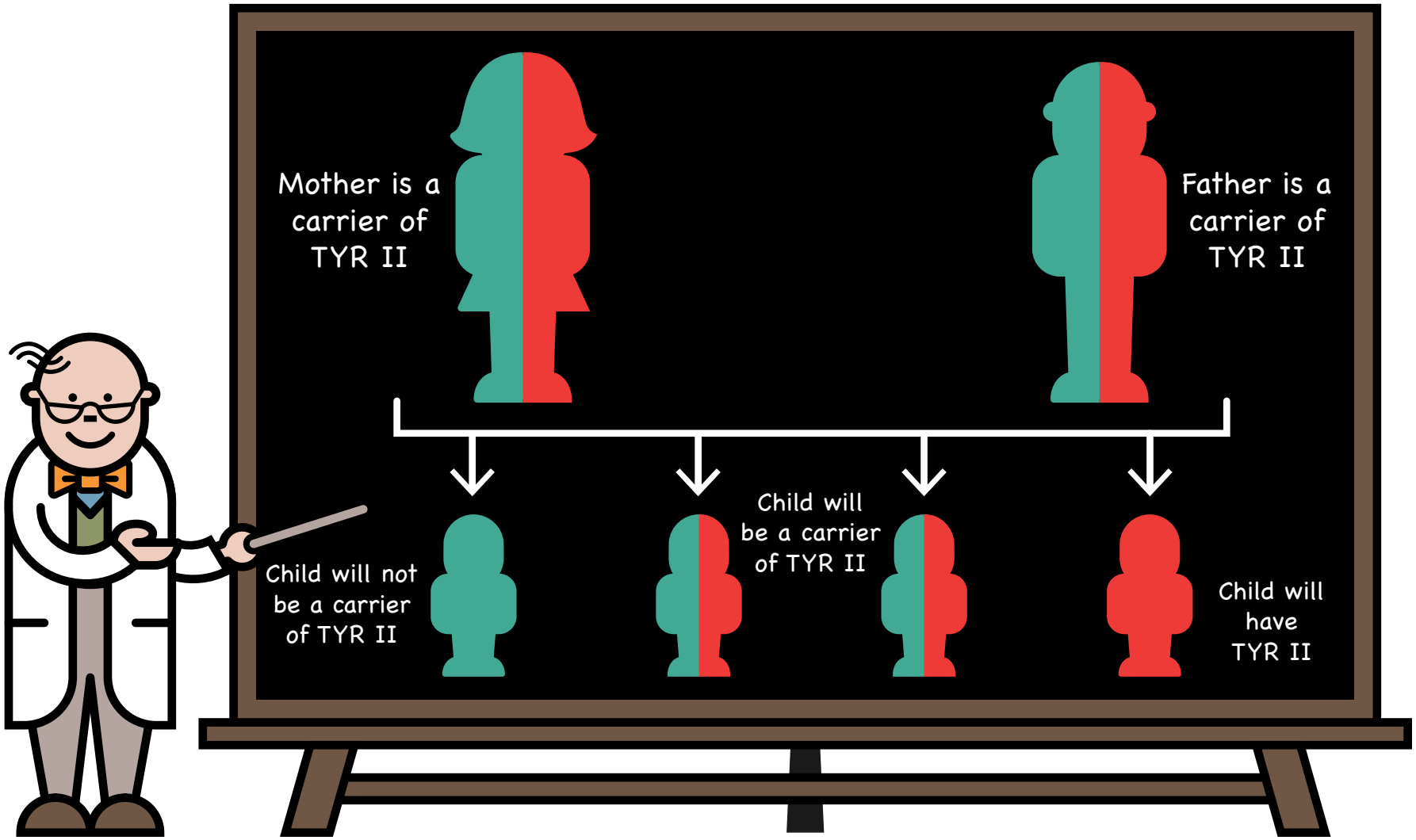


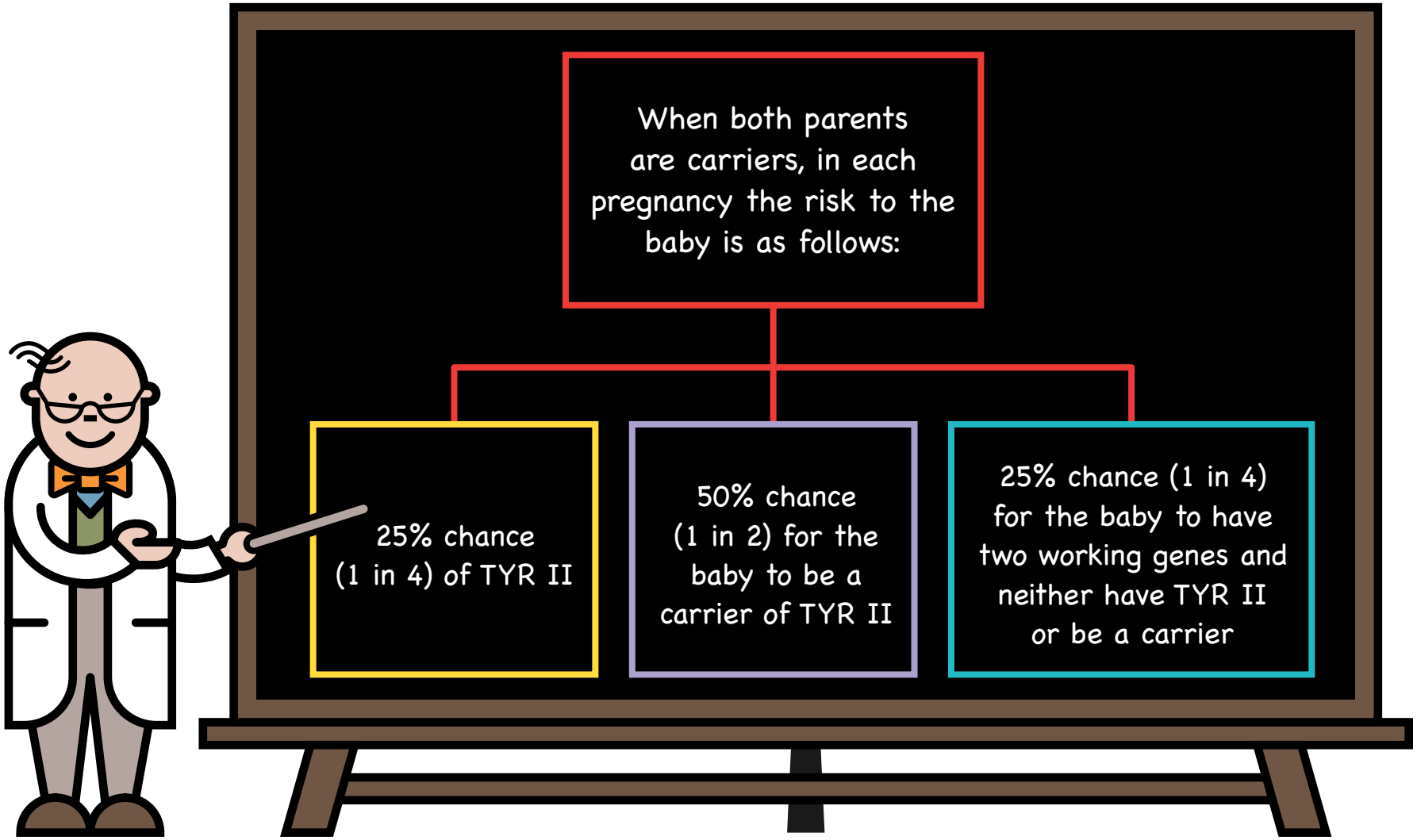
Inheritance – Autosomal recessive (carriers of TYR II)



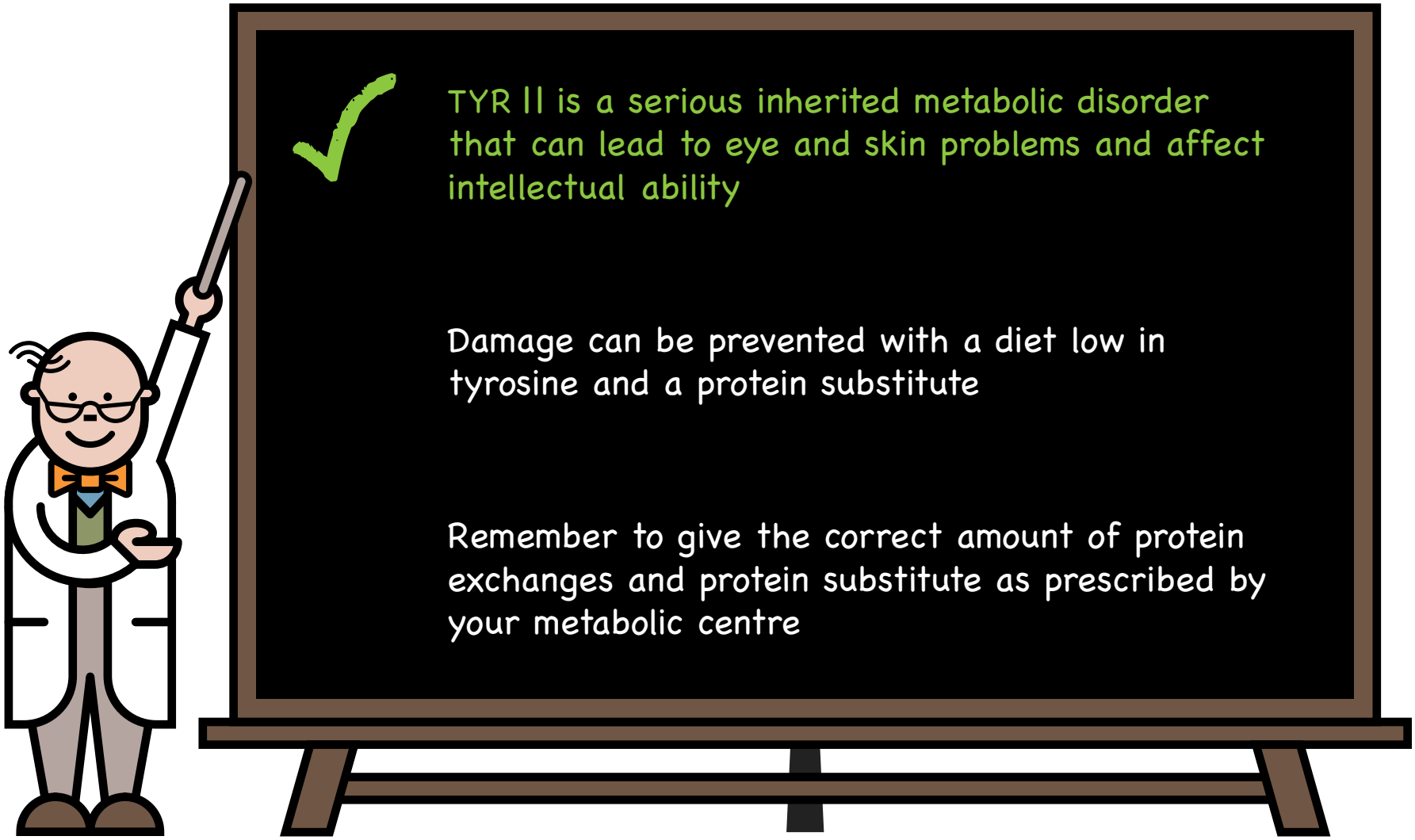
Inheritance – Autosomal recessive – possible combinations



Future pregnancies



Take home messages



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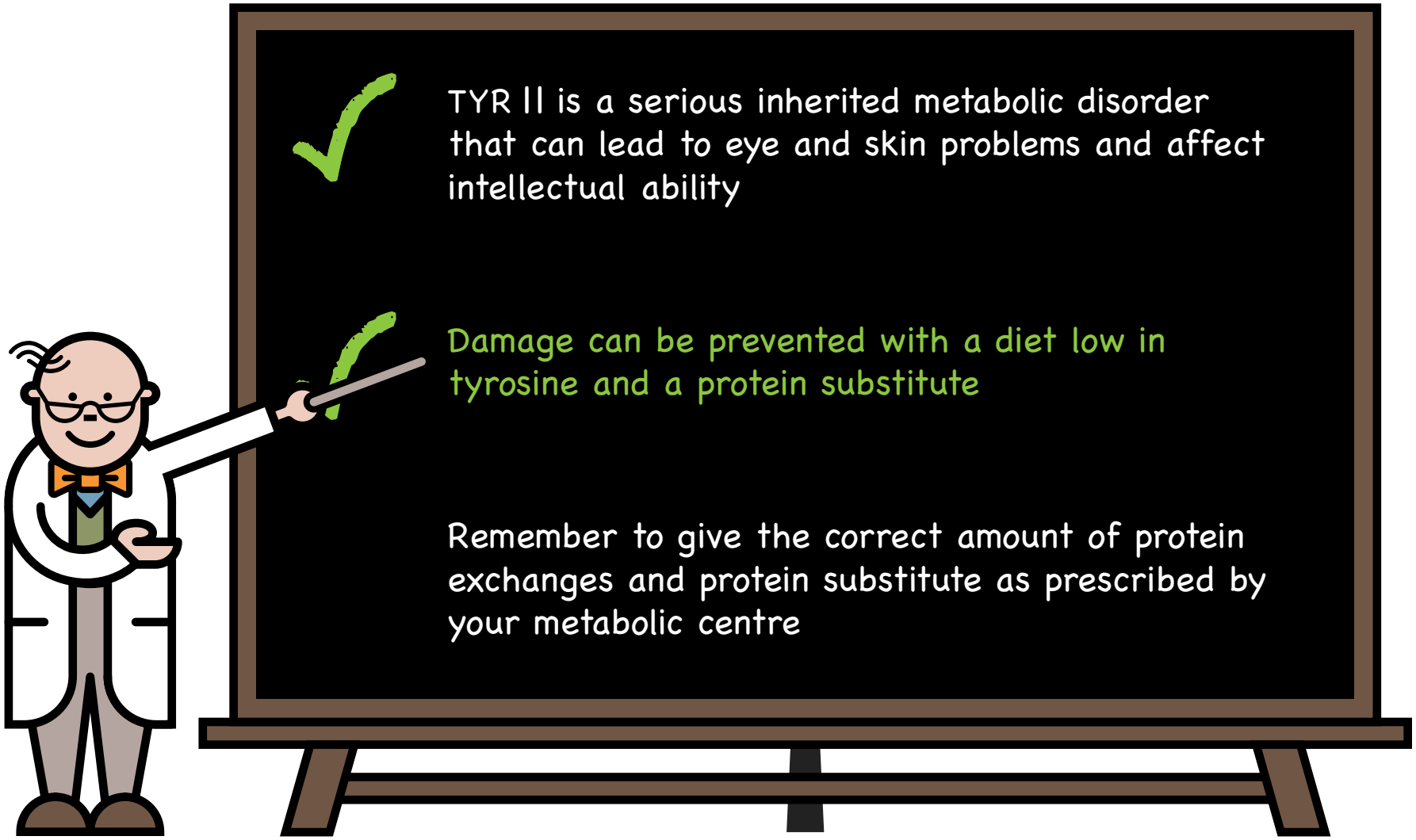

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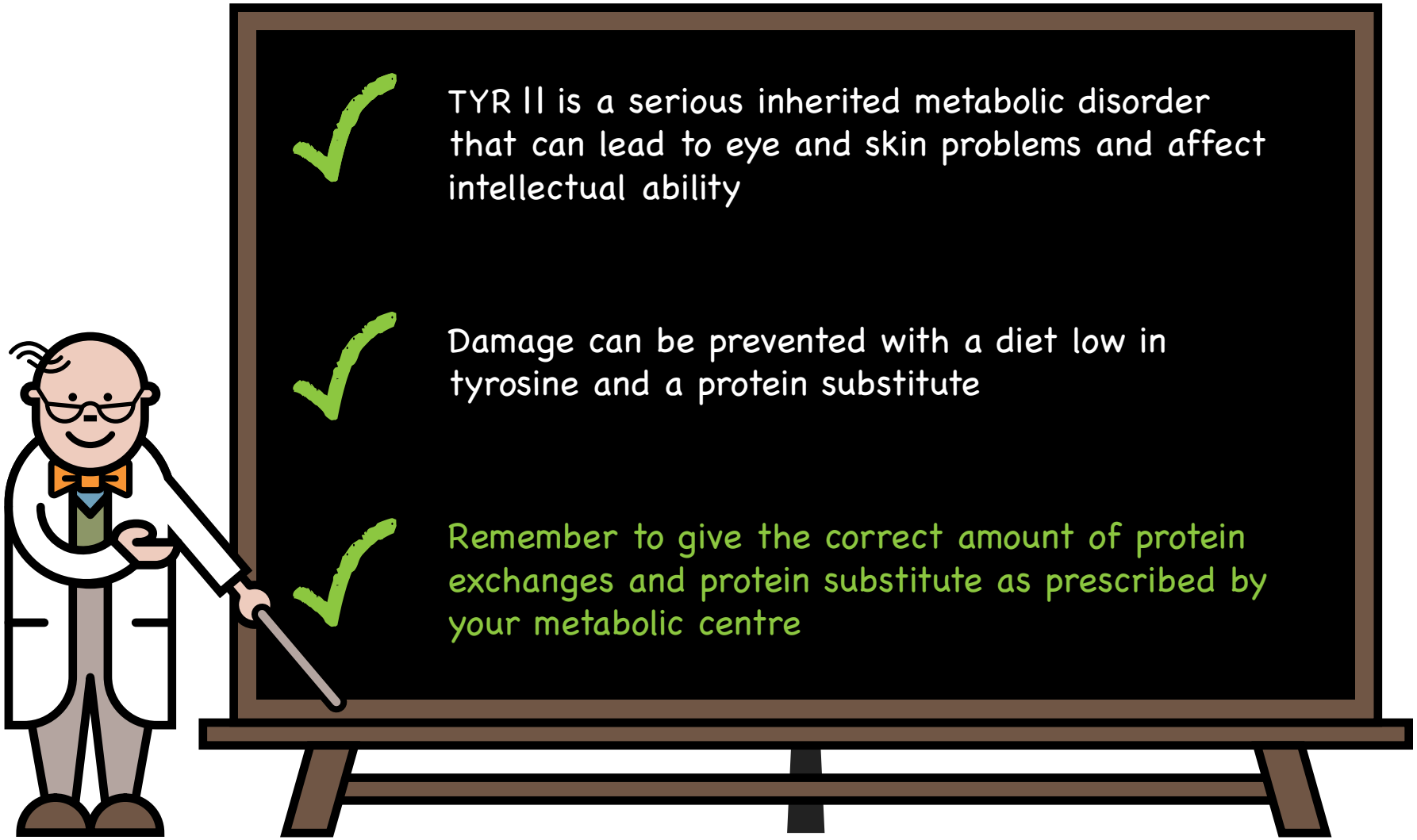

Helpful hints

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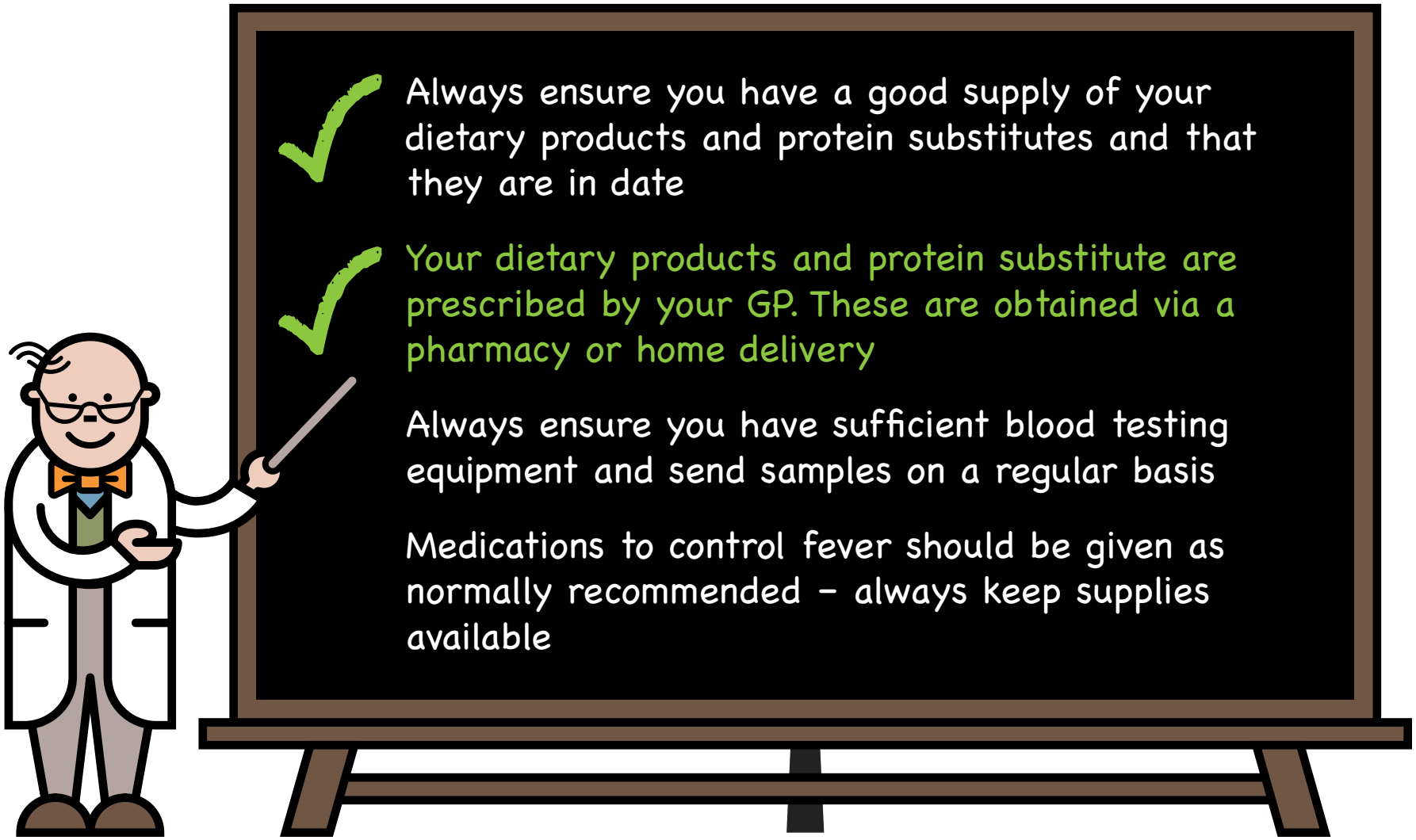

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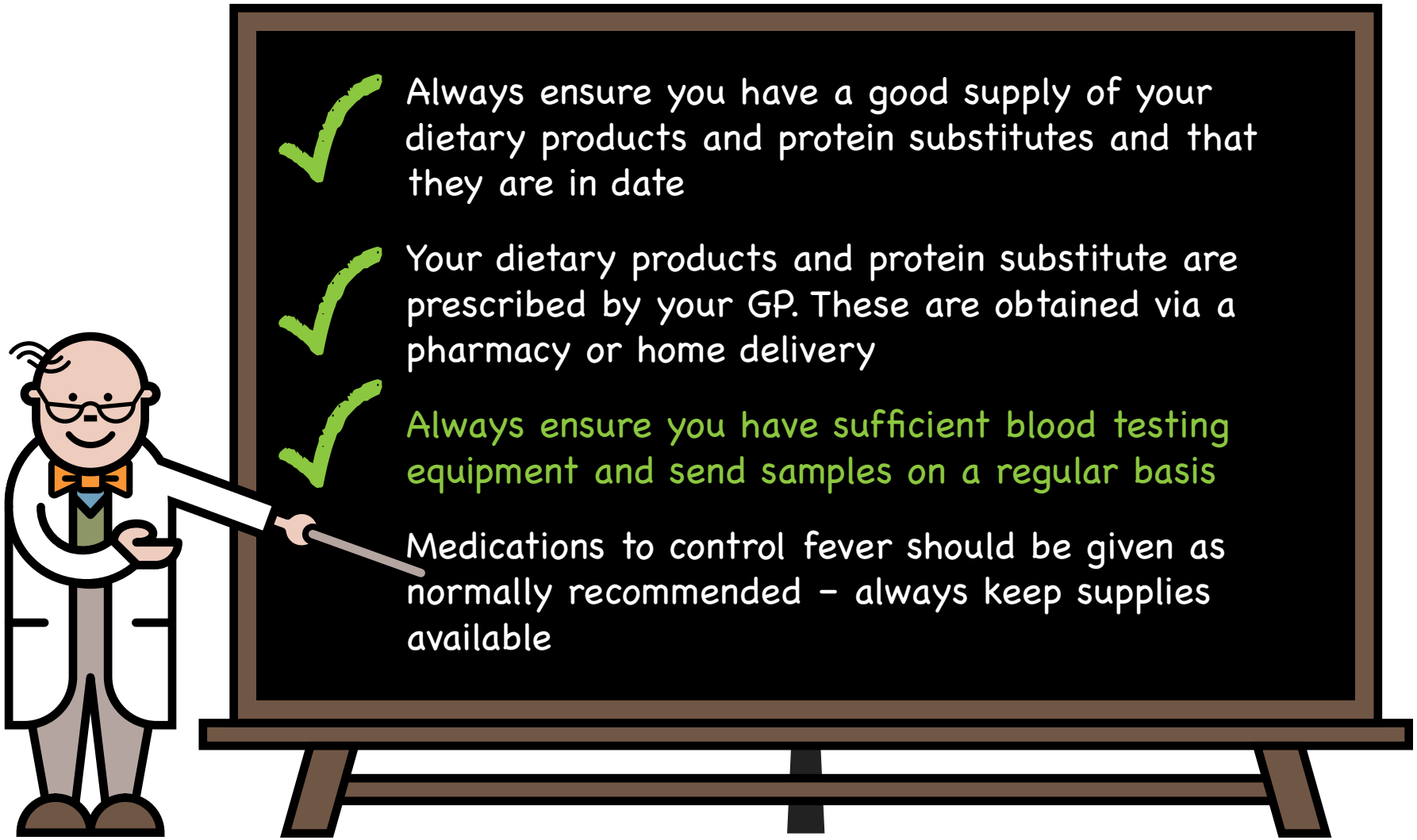

Helpful hints

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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