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

HTI

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.

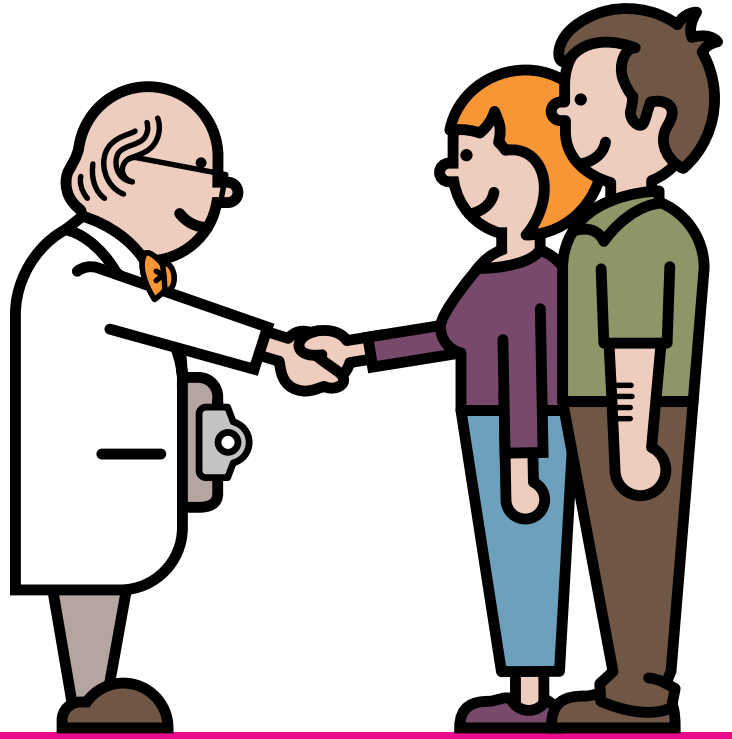
Each module produced is reviewed by a consultant clinician who is a member of the BIMDG.

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 I (HT I)

Information for families following a new diagnosis



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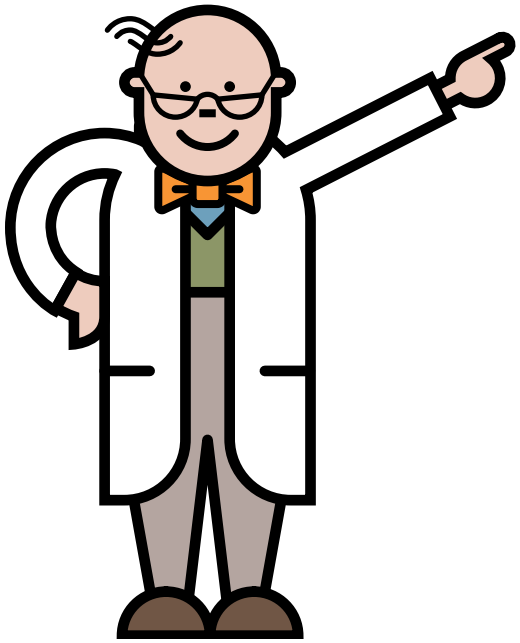
Tools Enabling Metabolic Parents LEarning

What is HT I?

HT I stands for hereditary tyrosinaemia Type I

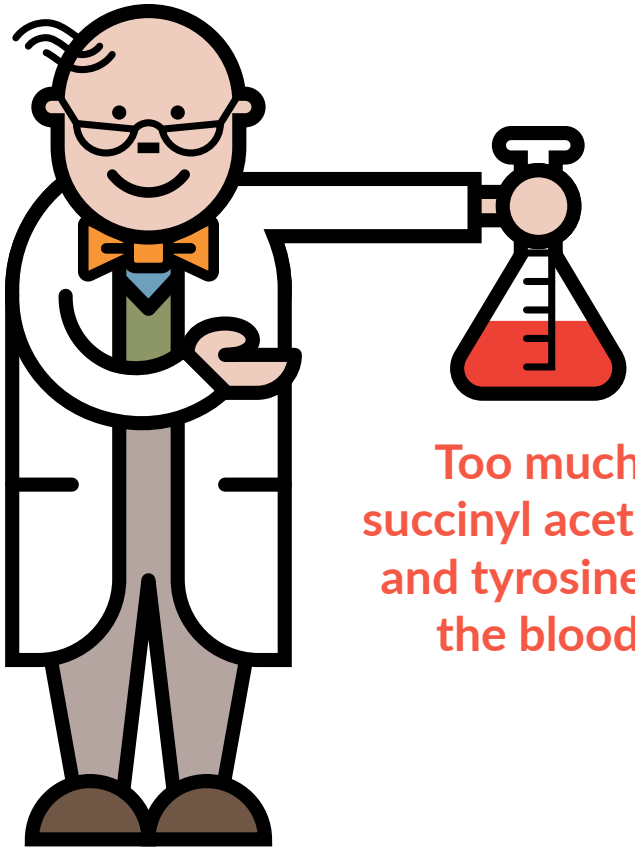
It is an inherited metabolic condition

Hereditary Tyrosinaemia Type I



HT I

What is HT I?



Too much succinyl acetone and tyrosine in the blood

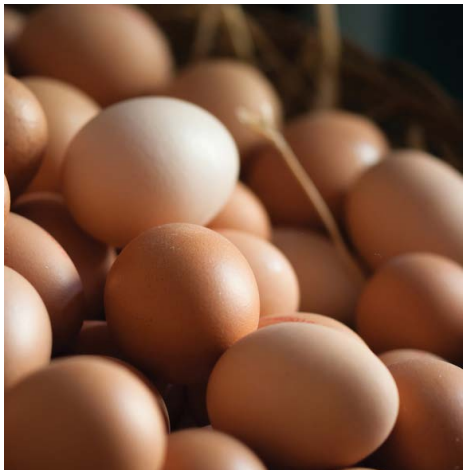
Too much succinyl acetone in the urine

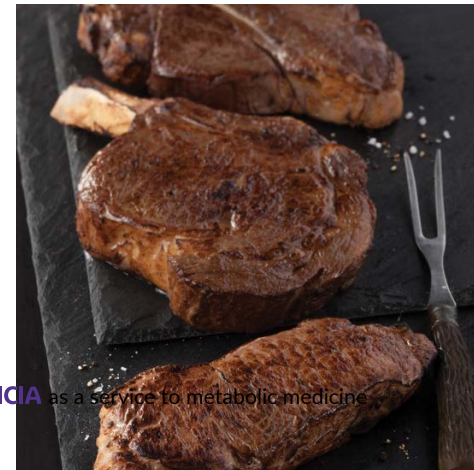
HT I and protein

HT I affects the way your baby breaks down protein

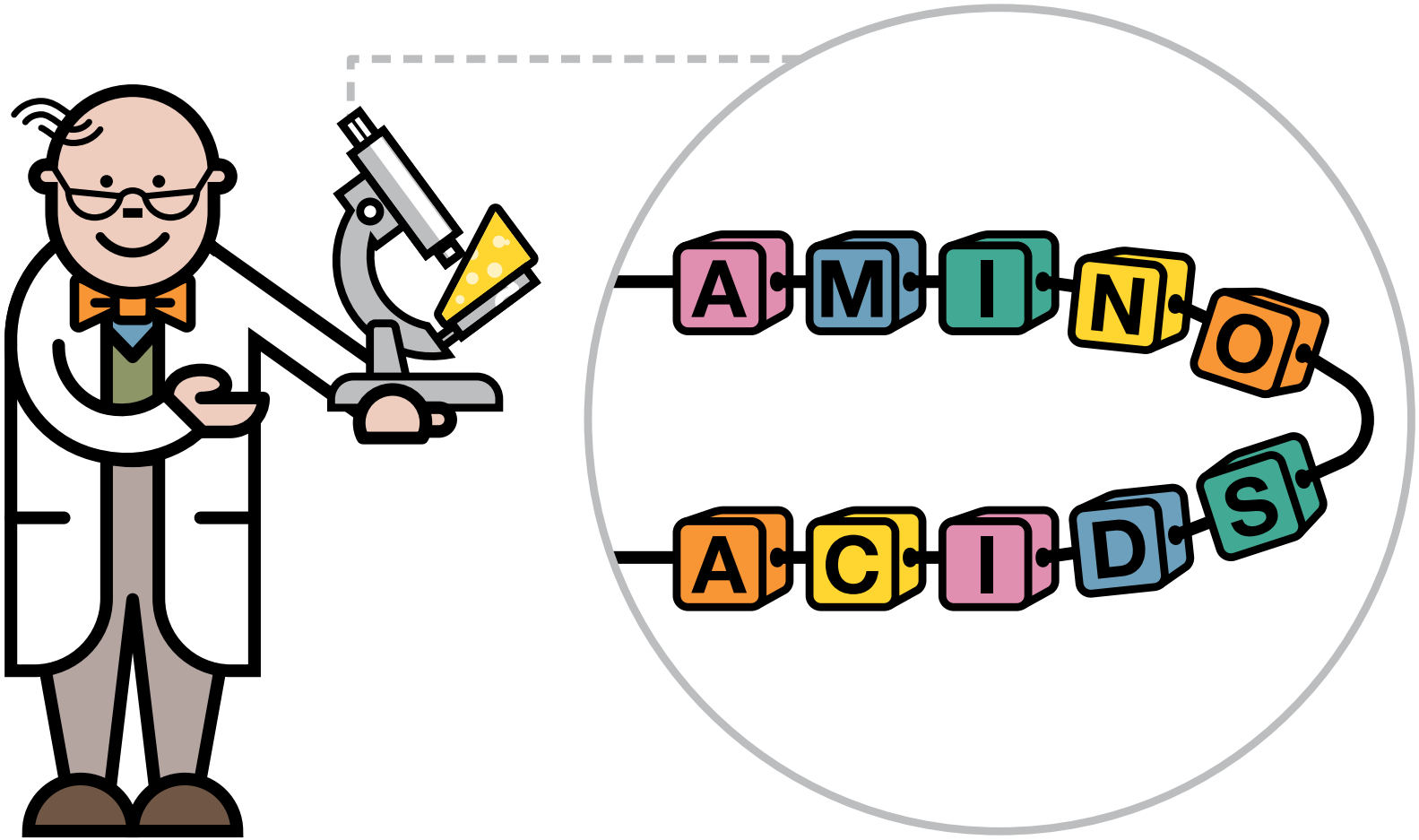
Many foods contain protein

The body needs protein for growth and repair





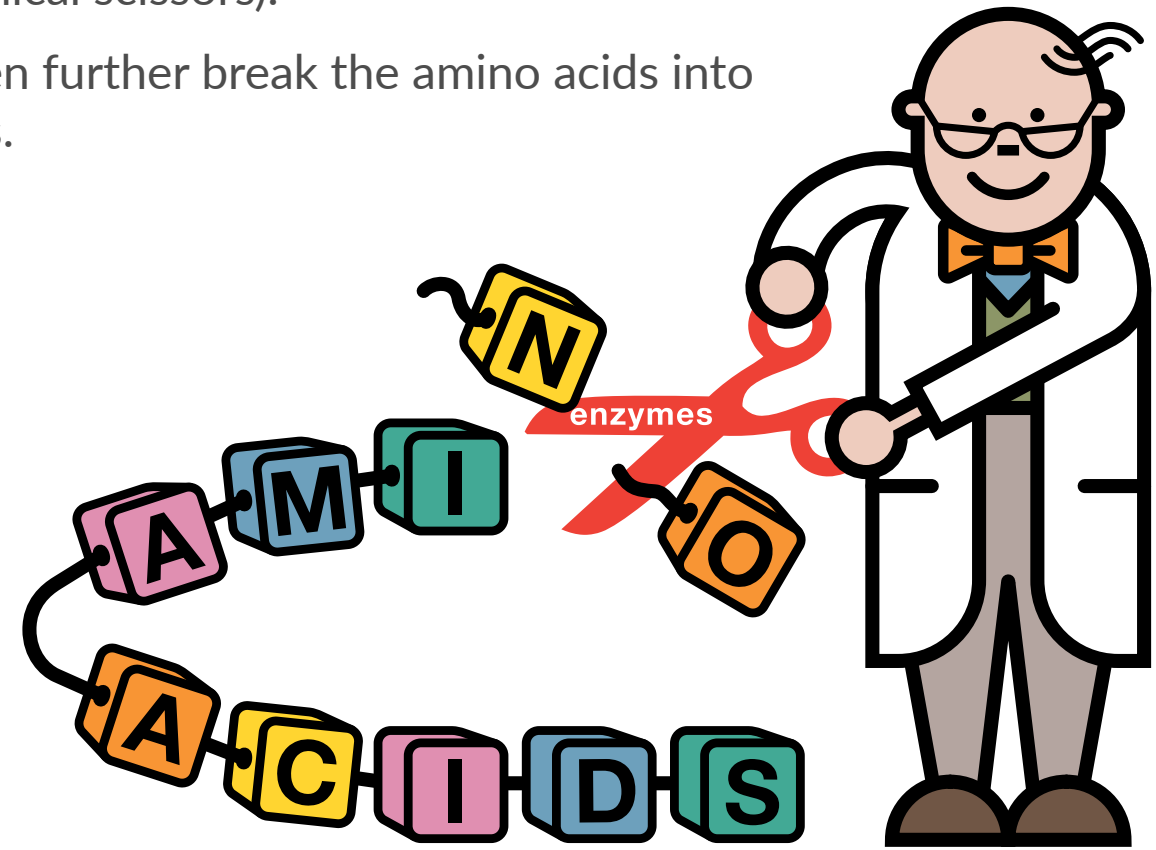
What is protein?



Protein and enzymes

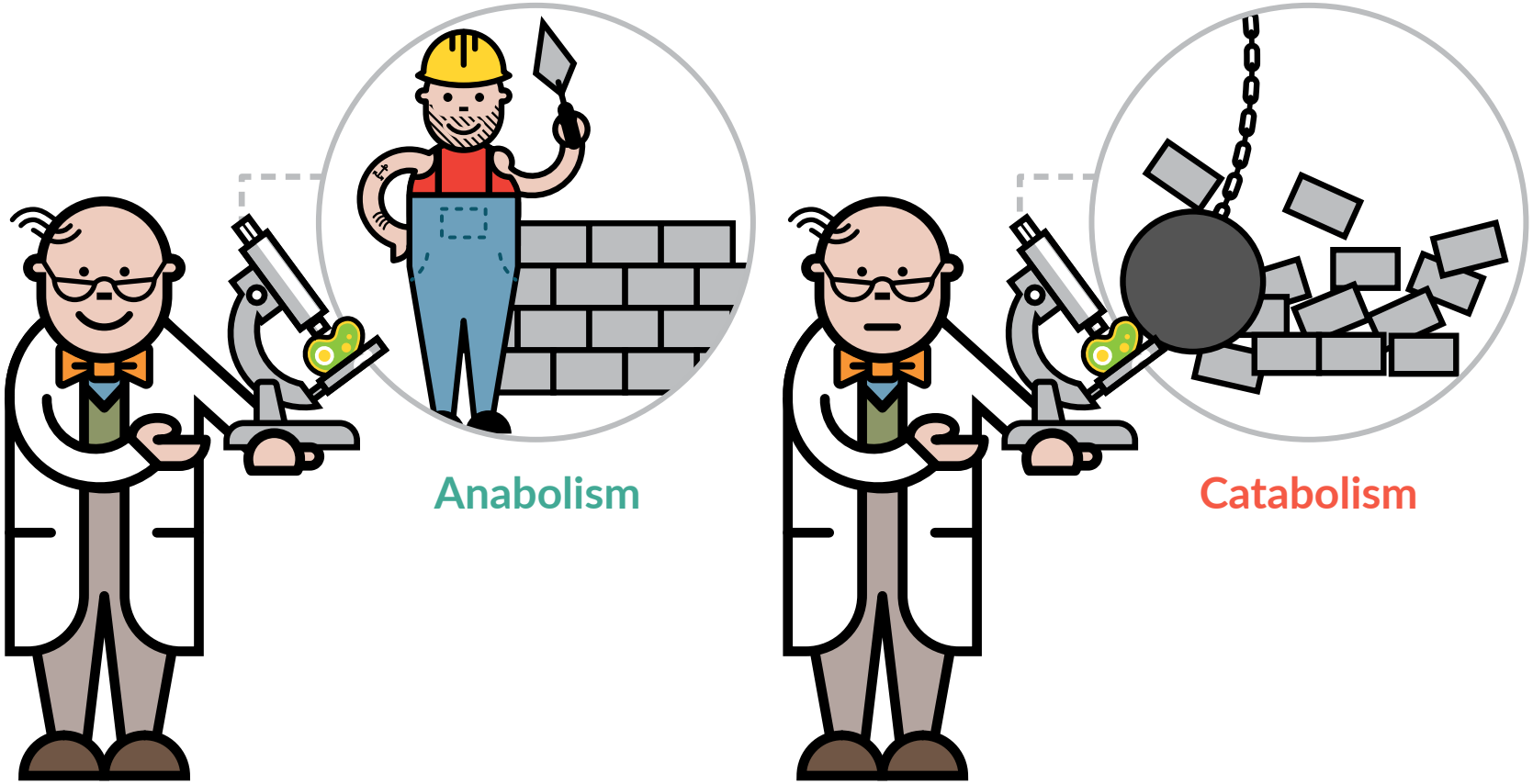
Protein is broken down into amino acids (building blocks of protein) by enzymes (which are like chemical scissors).

Enzymes then further break the amino acids into smaller parts.



Protein metabolism

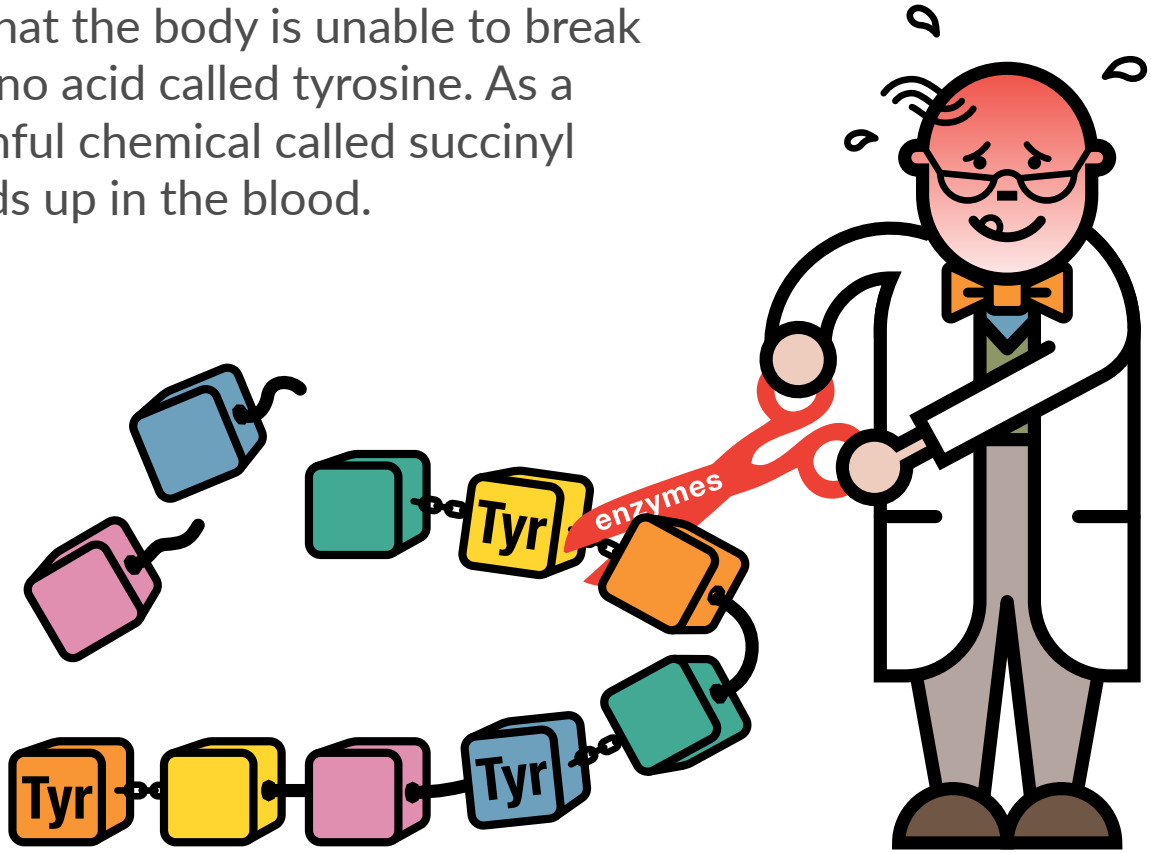
Metabolism refers to the chemical processes that occur inside the cells of the body.



What happens in HT I?

In HT I, the body lacks an enzyme called **fumarylacetoacetate hydrolyase**.

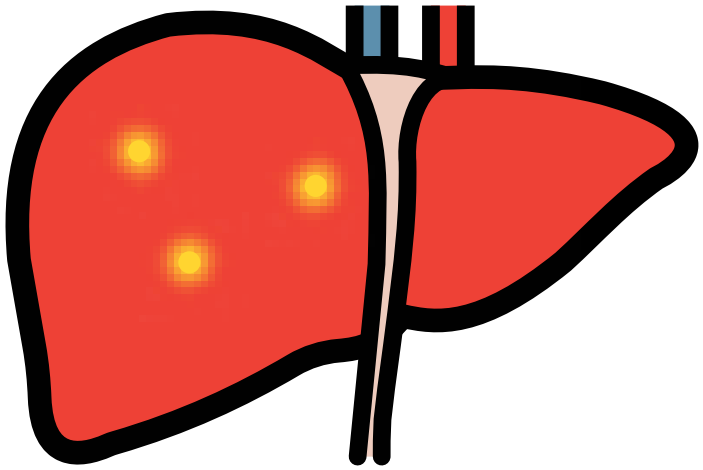
This means that the body is unable to break down an amino acid called tyrosine. As a result, a harmful chemical called succinyl acetone builds up in the blood.

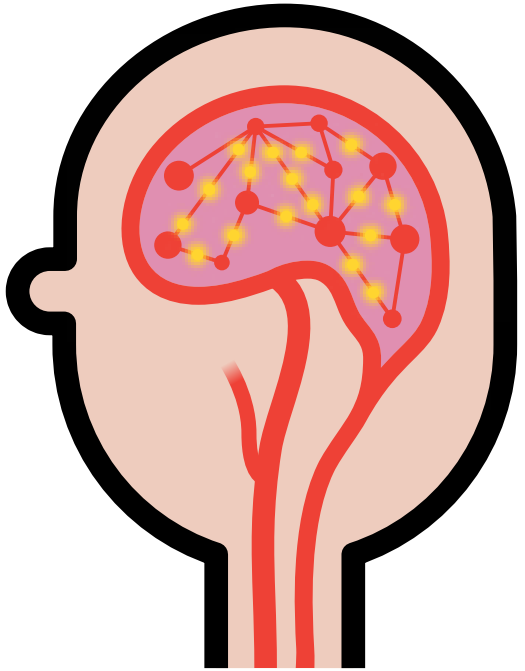


What can go wrong in HT I?

When unmanaged HT I can cause liver failure and lead to liver cancer.

Some children may have lower school achievement.

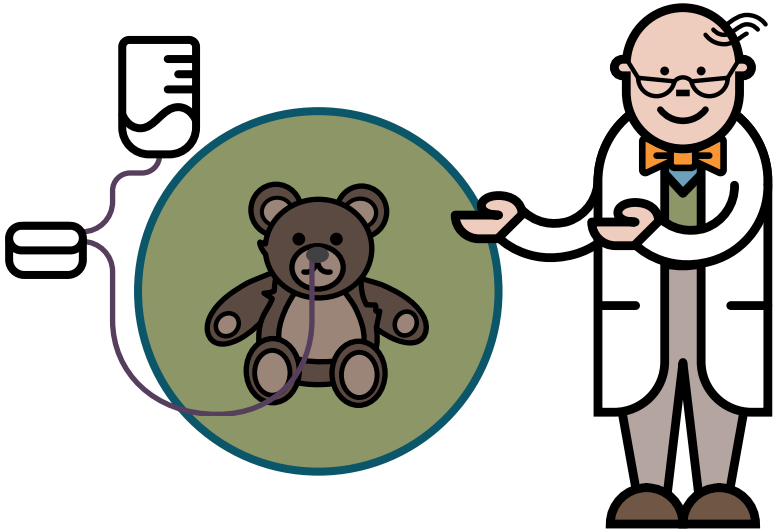




What are the symptoms in HT I?

Most babies become unwell in the first few months of life. Symptoms include:

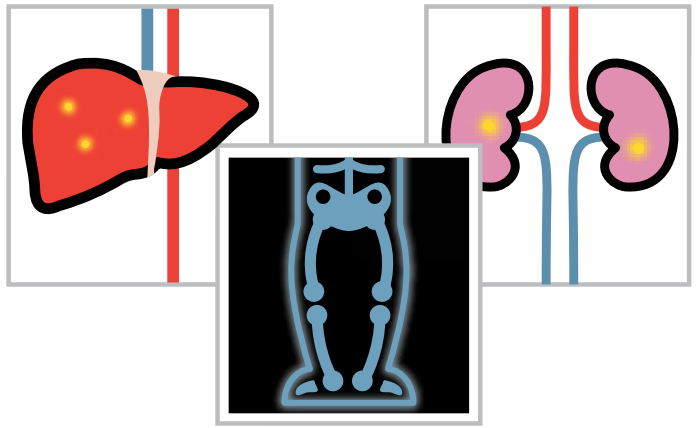
- Poor weight gain
- Liver failure



Other children have a gradual onset of symptoms such as:

- Large liver
- Rickets
- Kidney problems

Some children may develop liver cancer.



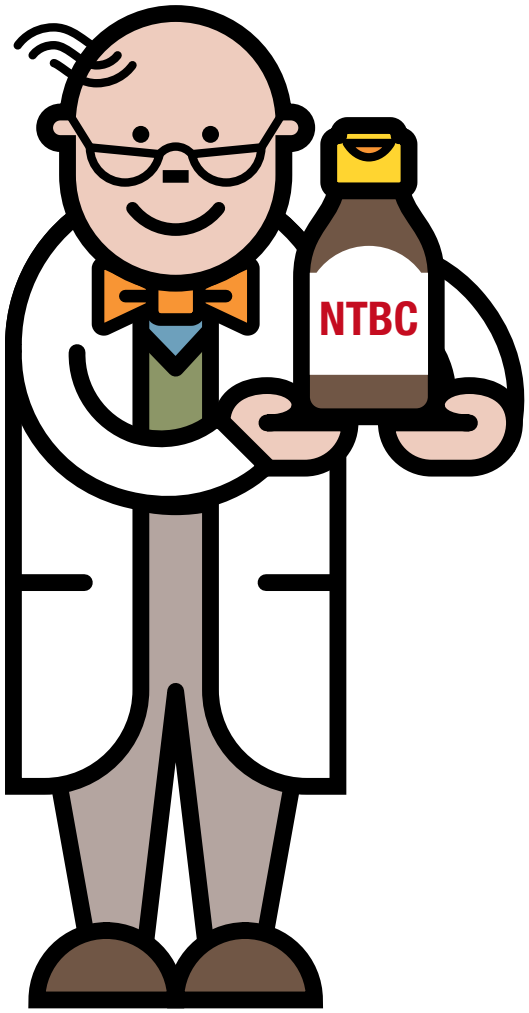
How is HT I diagnosed?

HT I is diagnosed by high levels of succinyl acetone in the blood and urine.

Management with nitisinone

This medication is also called NTBC.

- Your child will start taking NTBC/ nitisinone as soon as possible
- NTBC/nitisinone helps prevent liver and kidney damage and lowers the risk of liver cancer



How is HT I managed?

HT I is managed with the following special diet and medication:

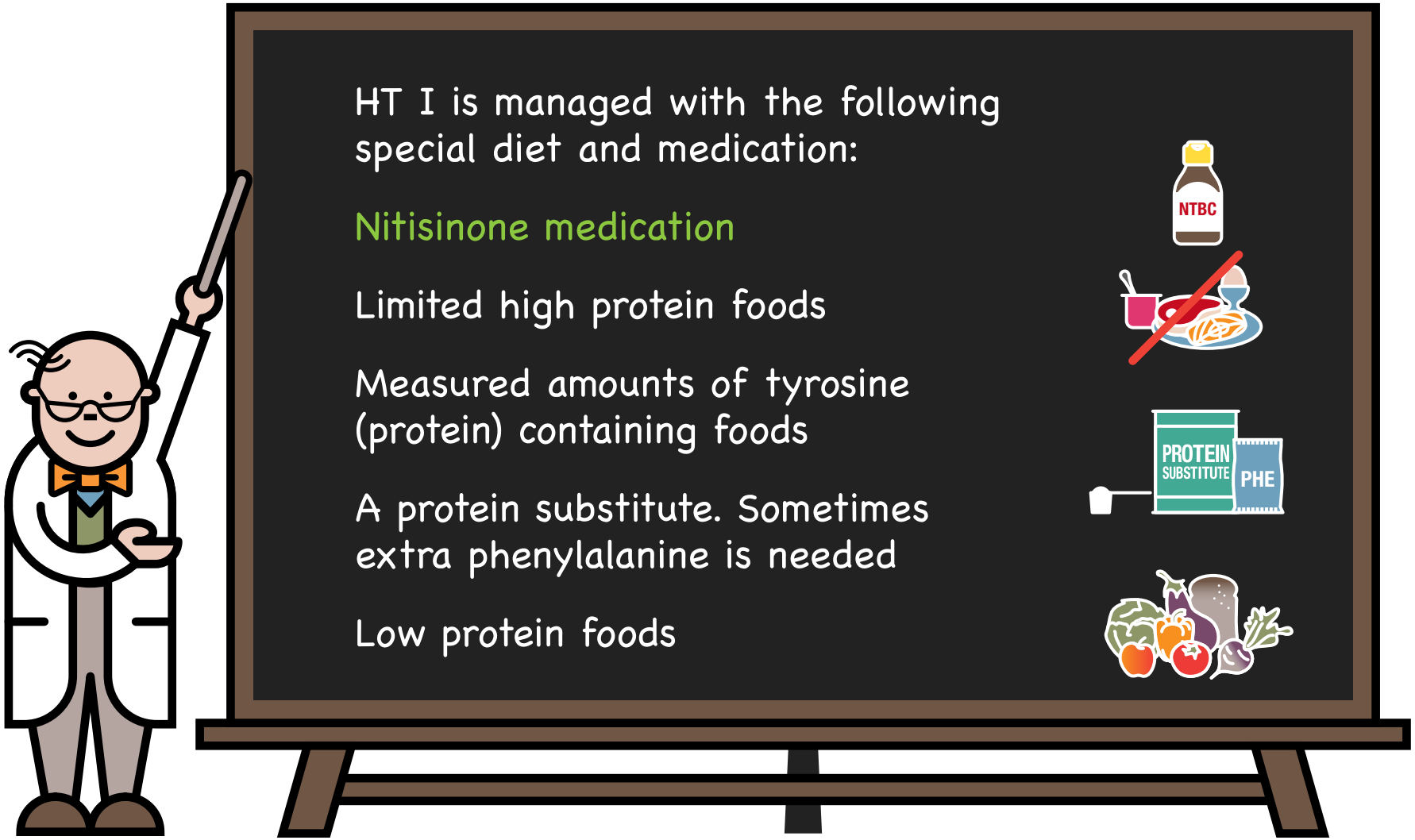
Nitisinone medication

Limited high protein foods

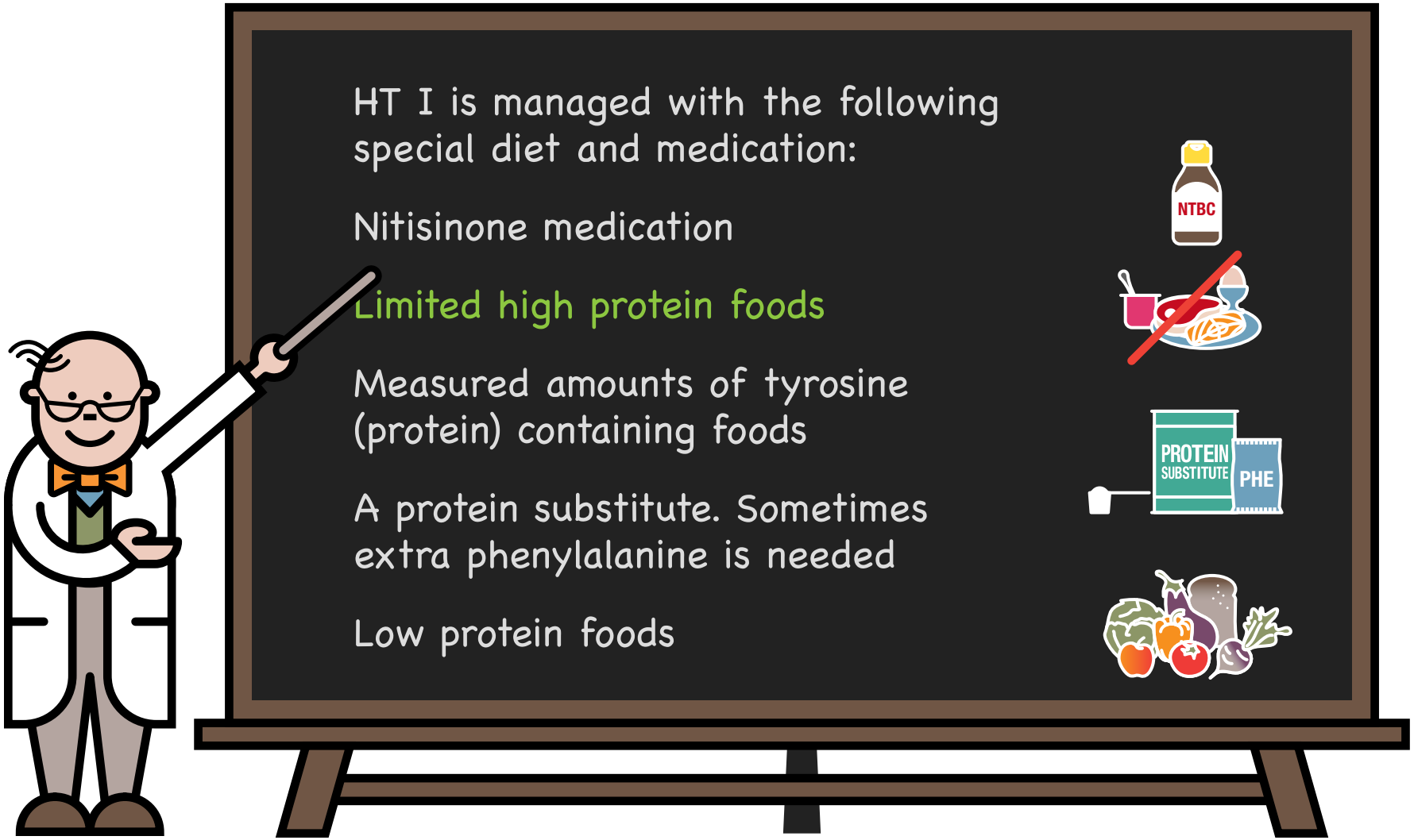
Measured amounts of tyrosine (protein) containing foods

A protein substitute. Sometimes extra phenylalanine is needed

Low protein foods



How is HT I managed?



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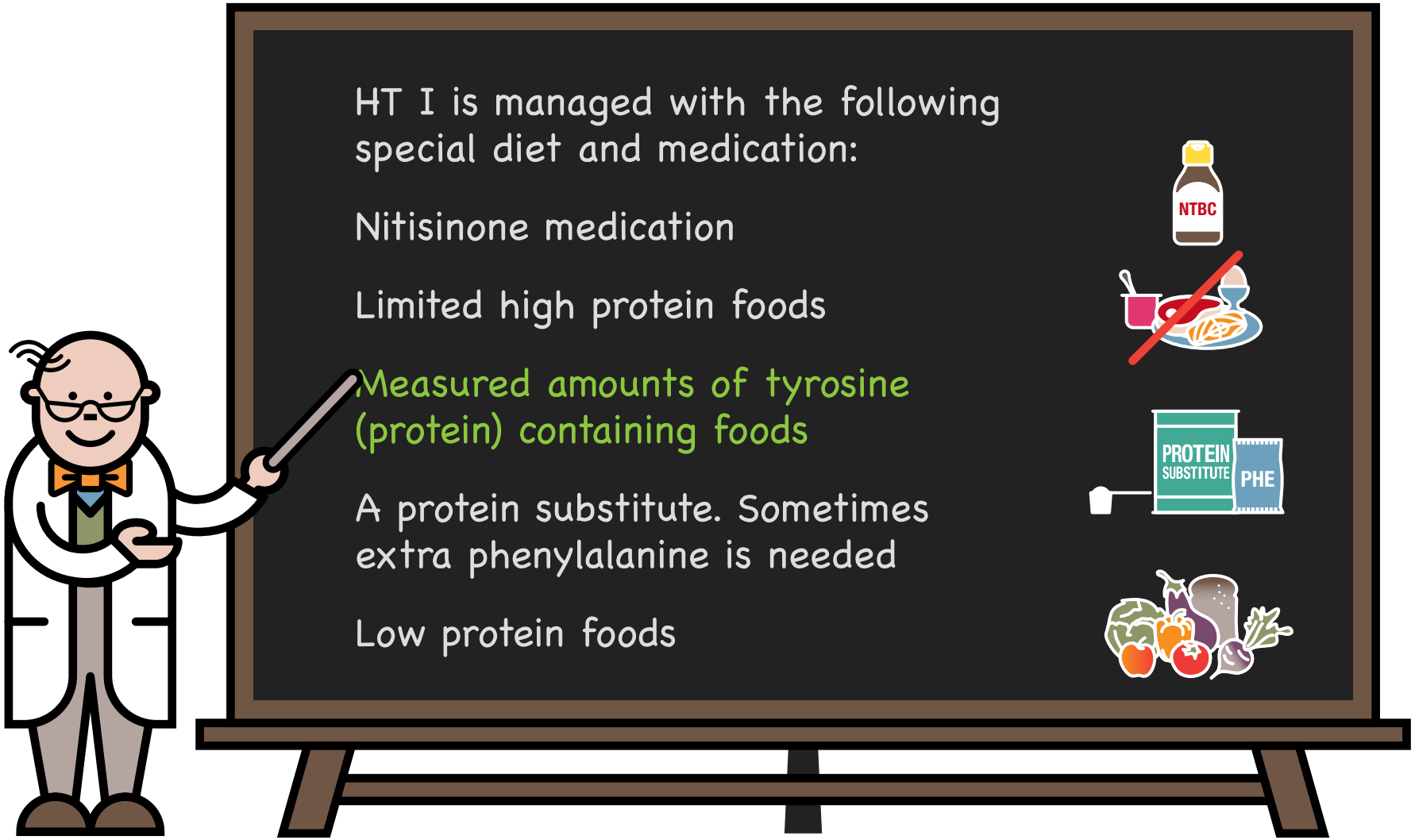

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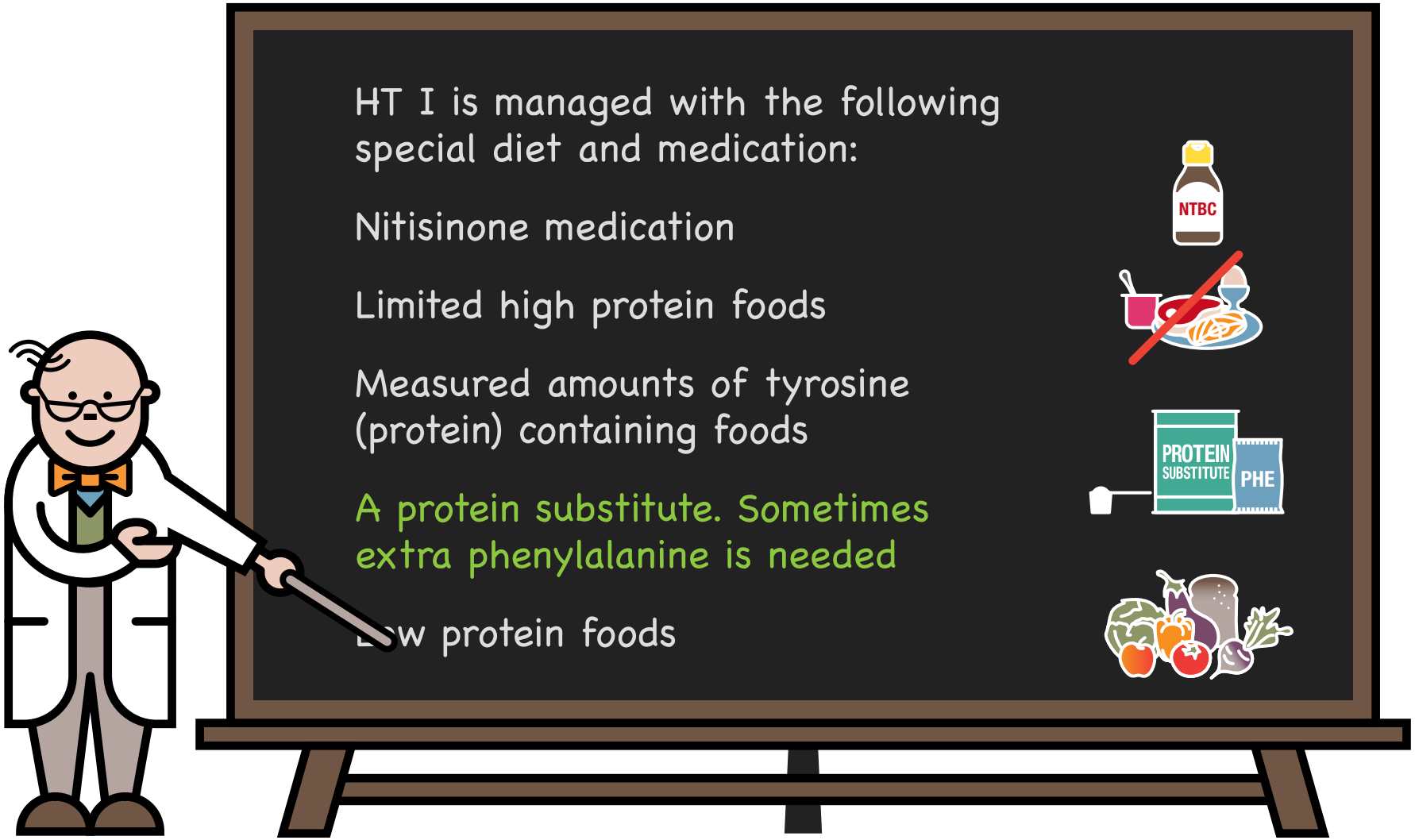

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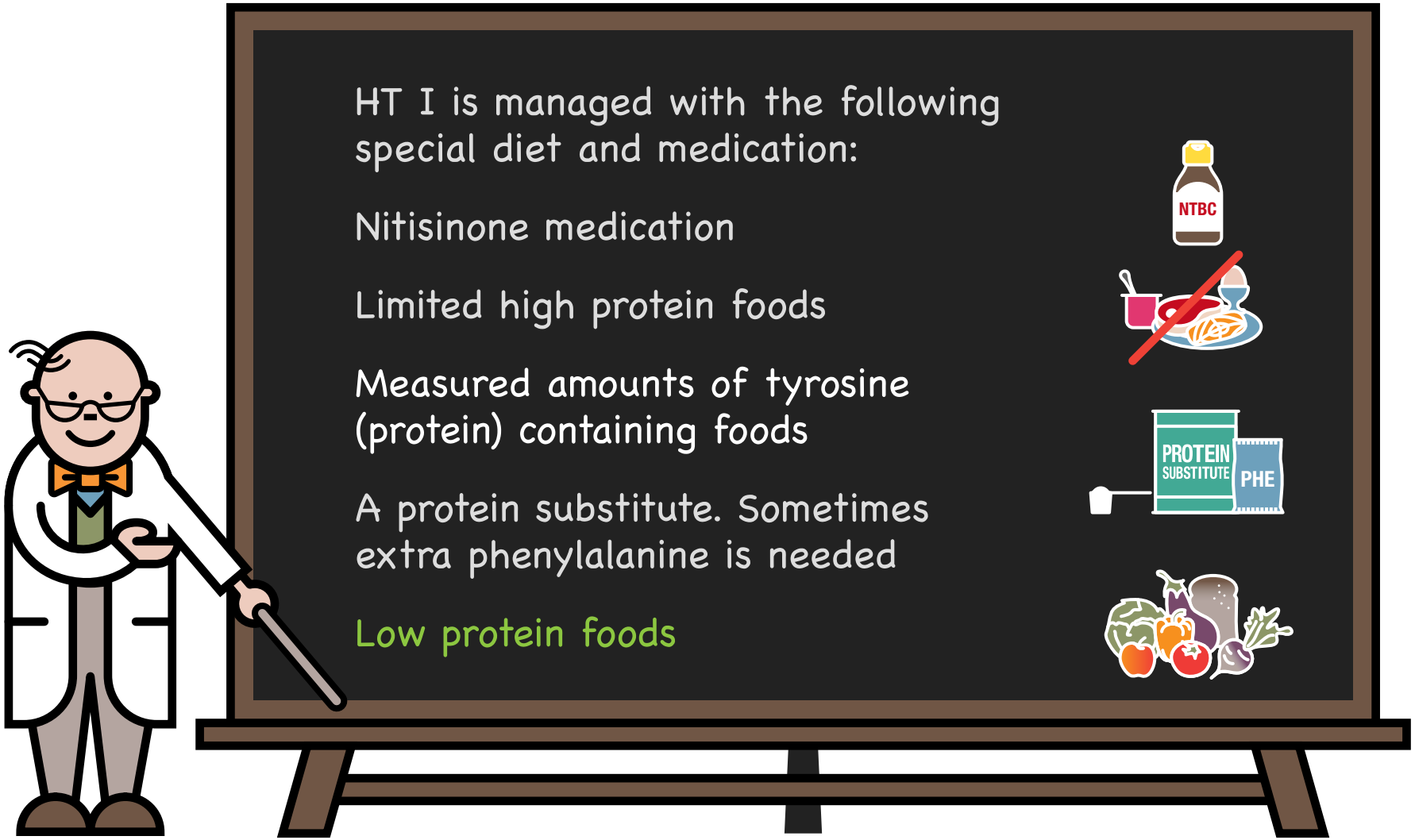


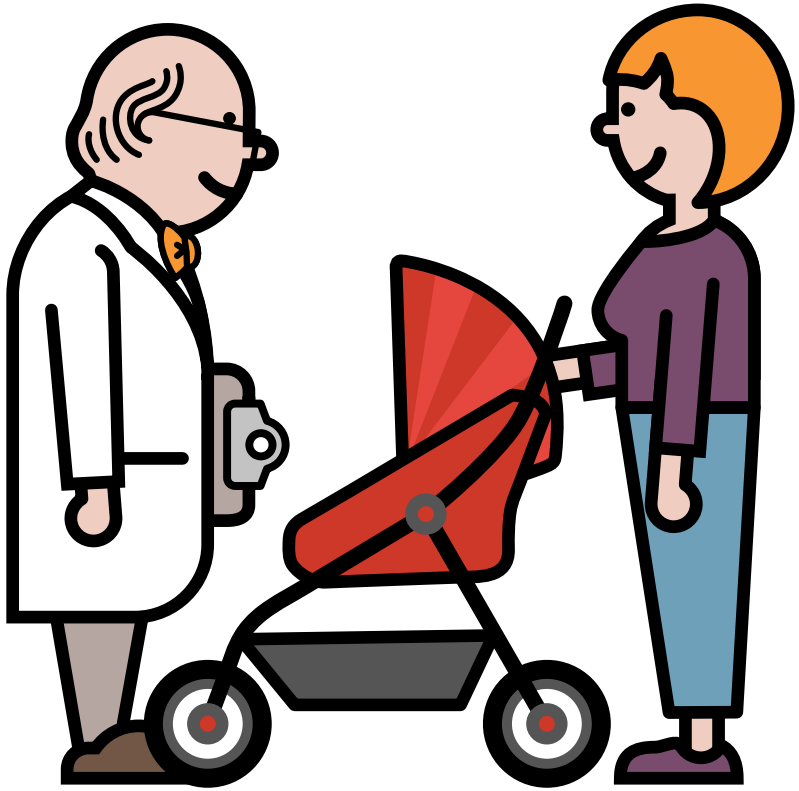
High protein foods

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 tyrosine intake

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

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

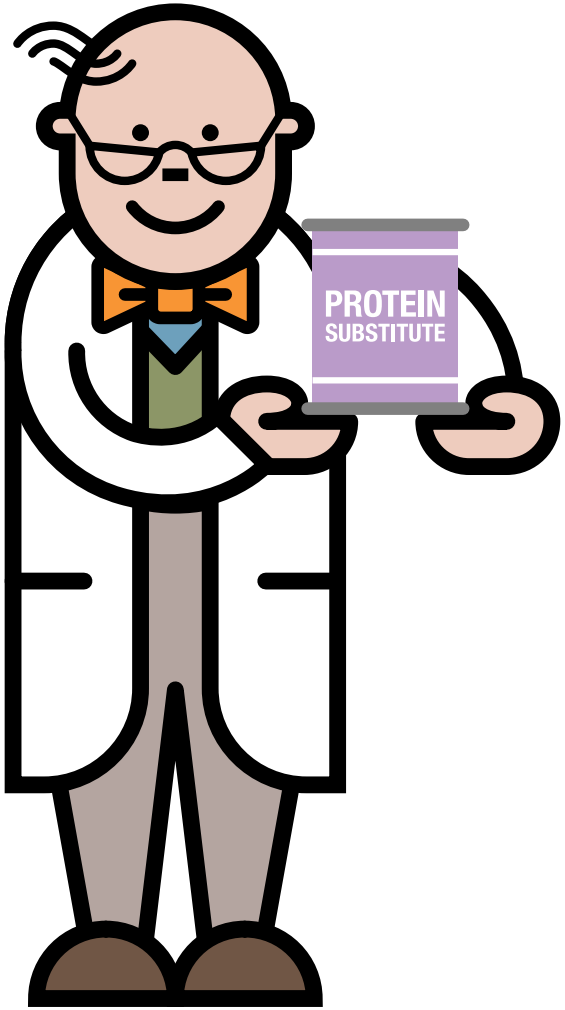


Protein substitute

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.

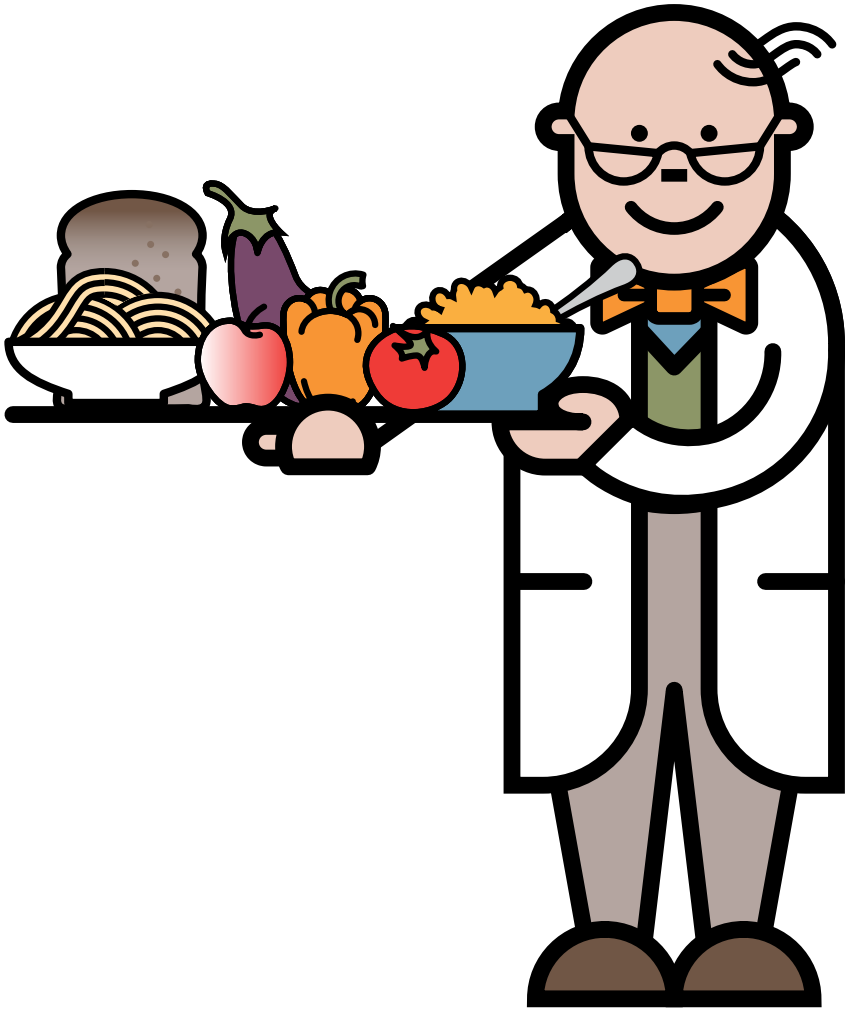


Low protein foods

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

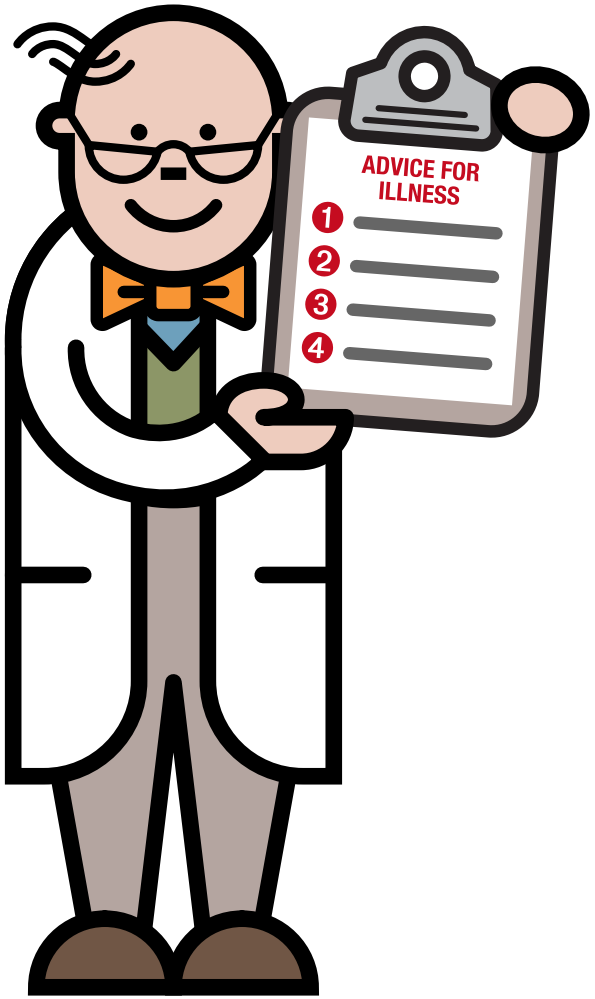


How is HT I managed during illness?

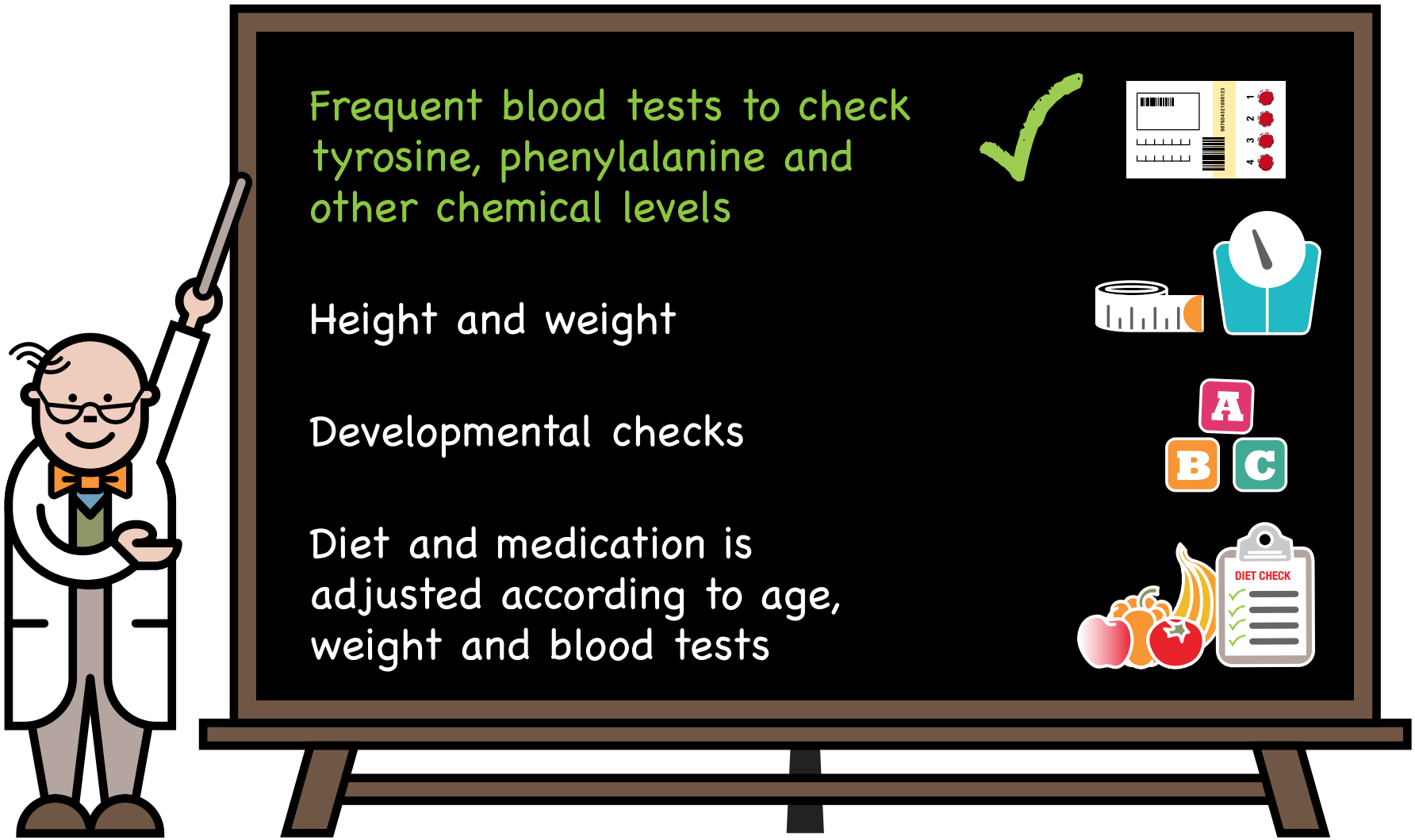
During any childhood illness, catabolism or protein breakdown occurs, causing blood tyrosine levels to increase.

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

The drug nitisinone should be given at all times including illness.



How is HT I monitored?



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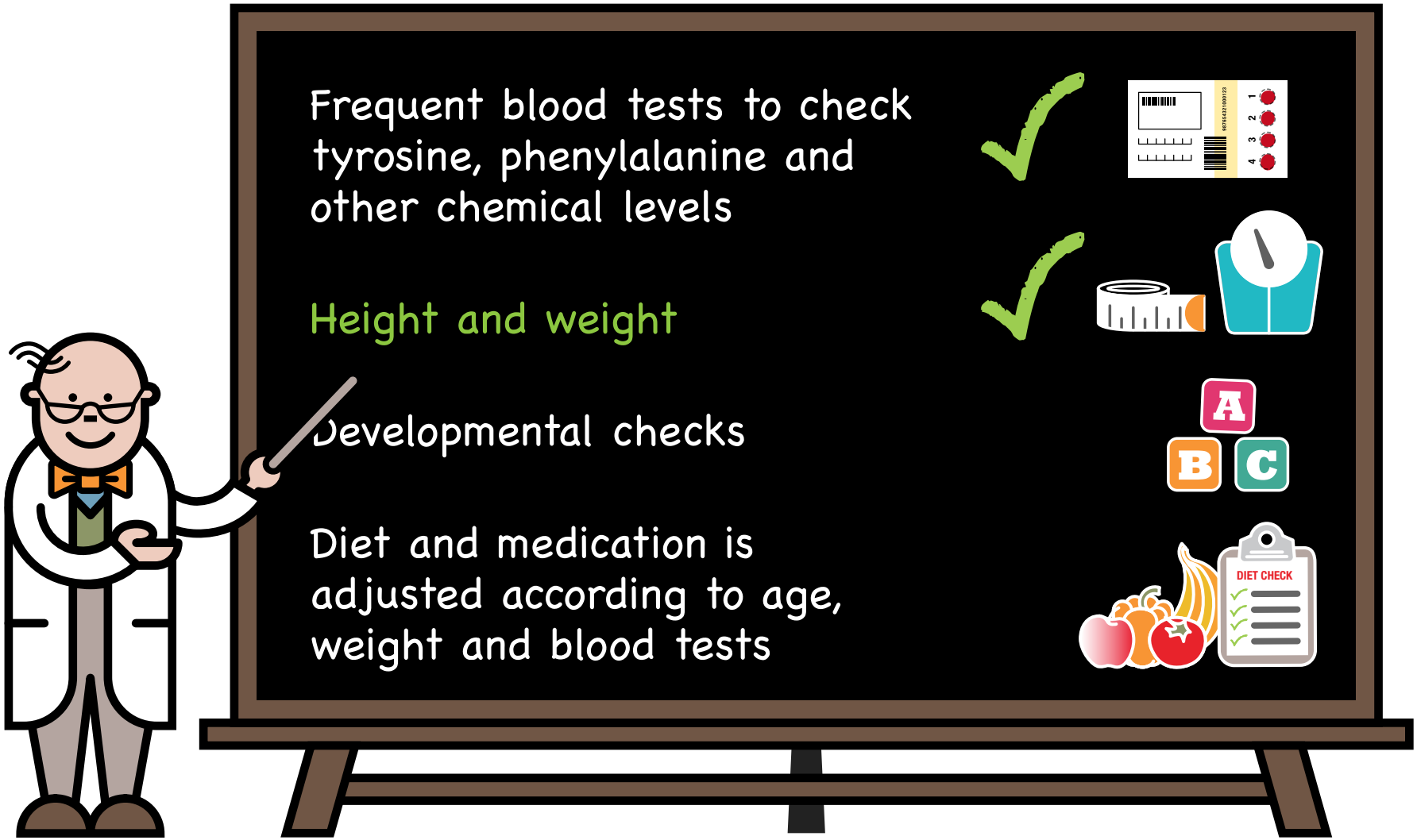

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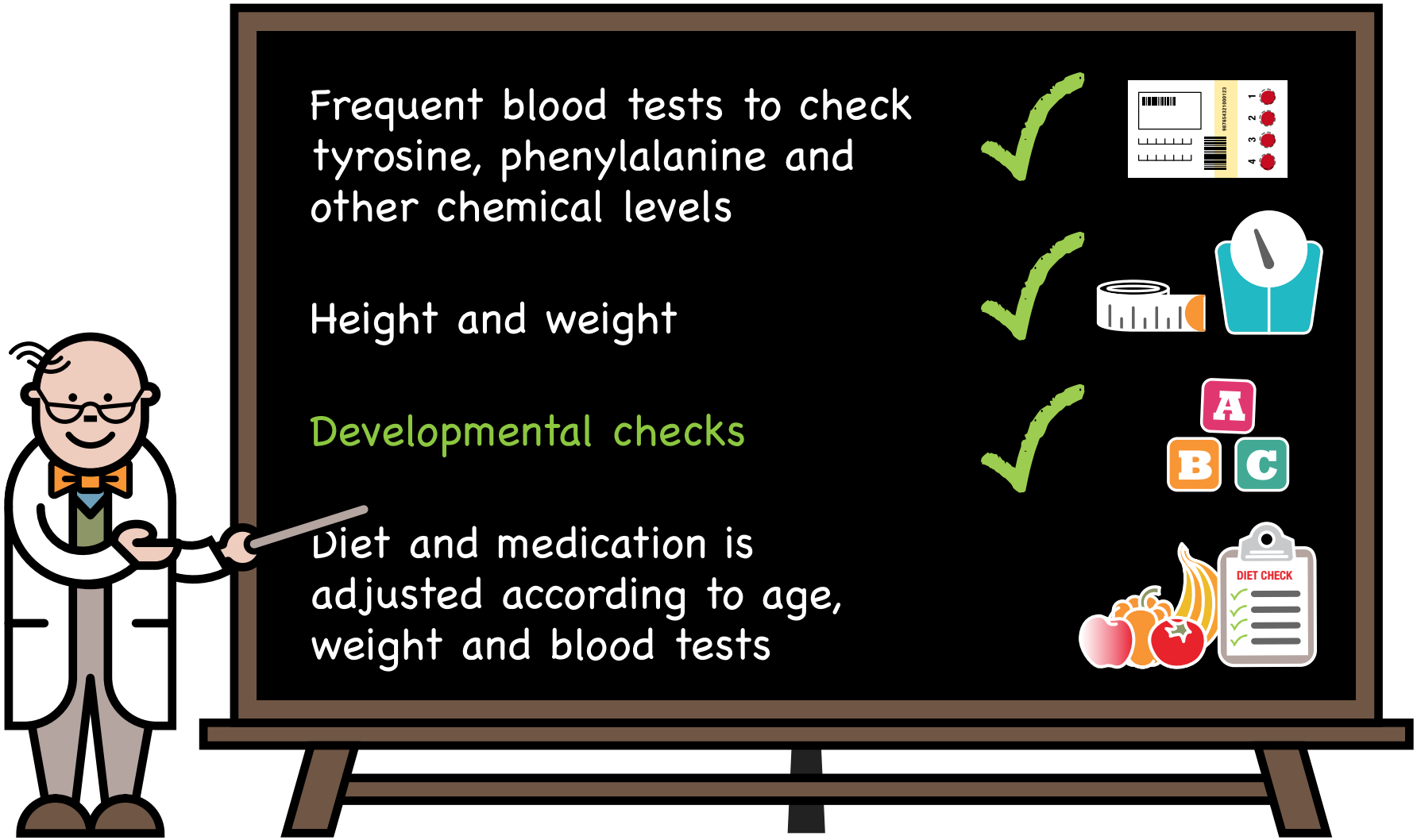

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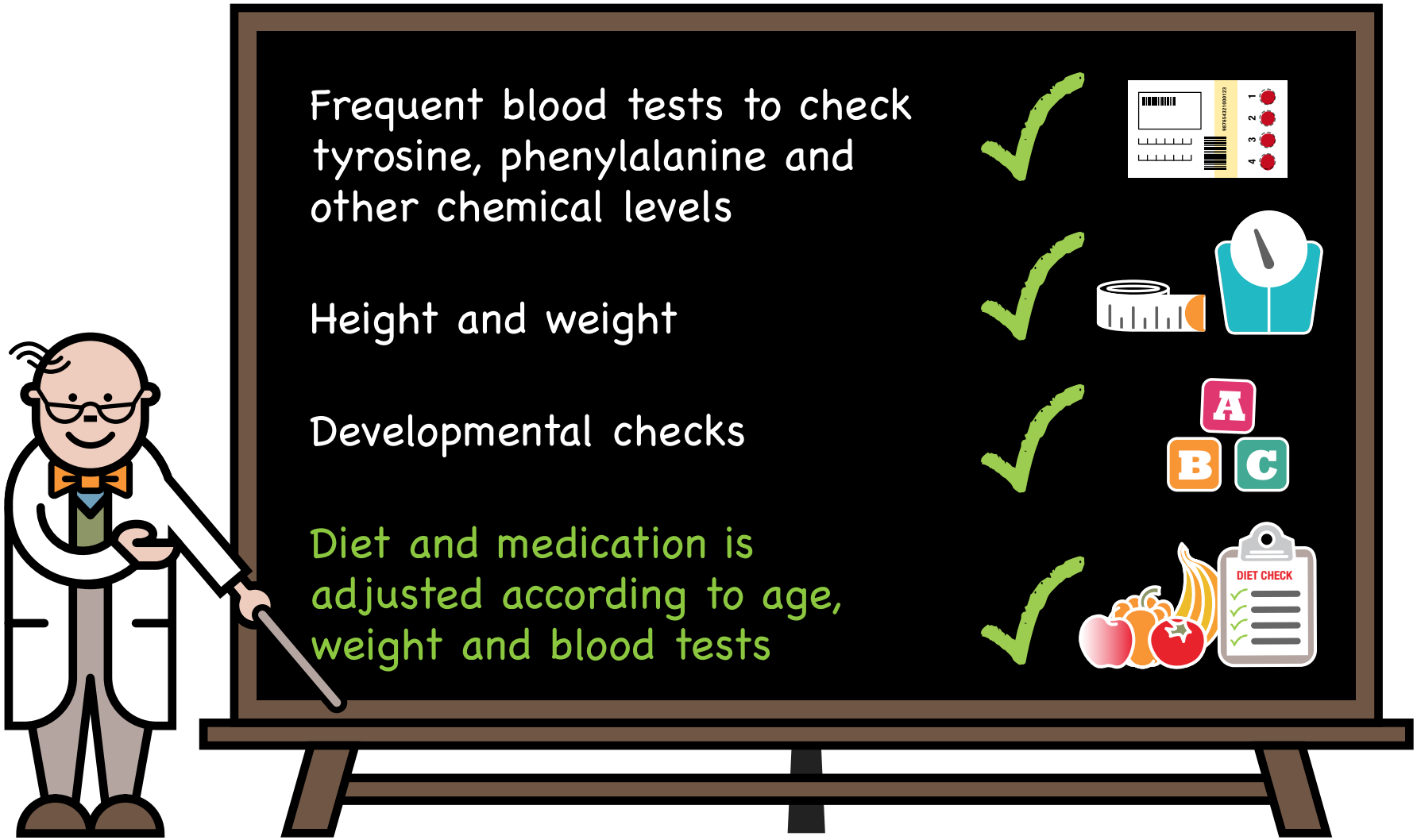
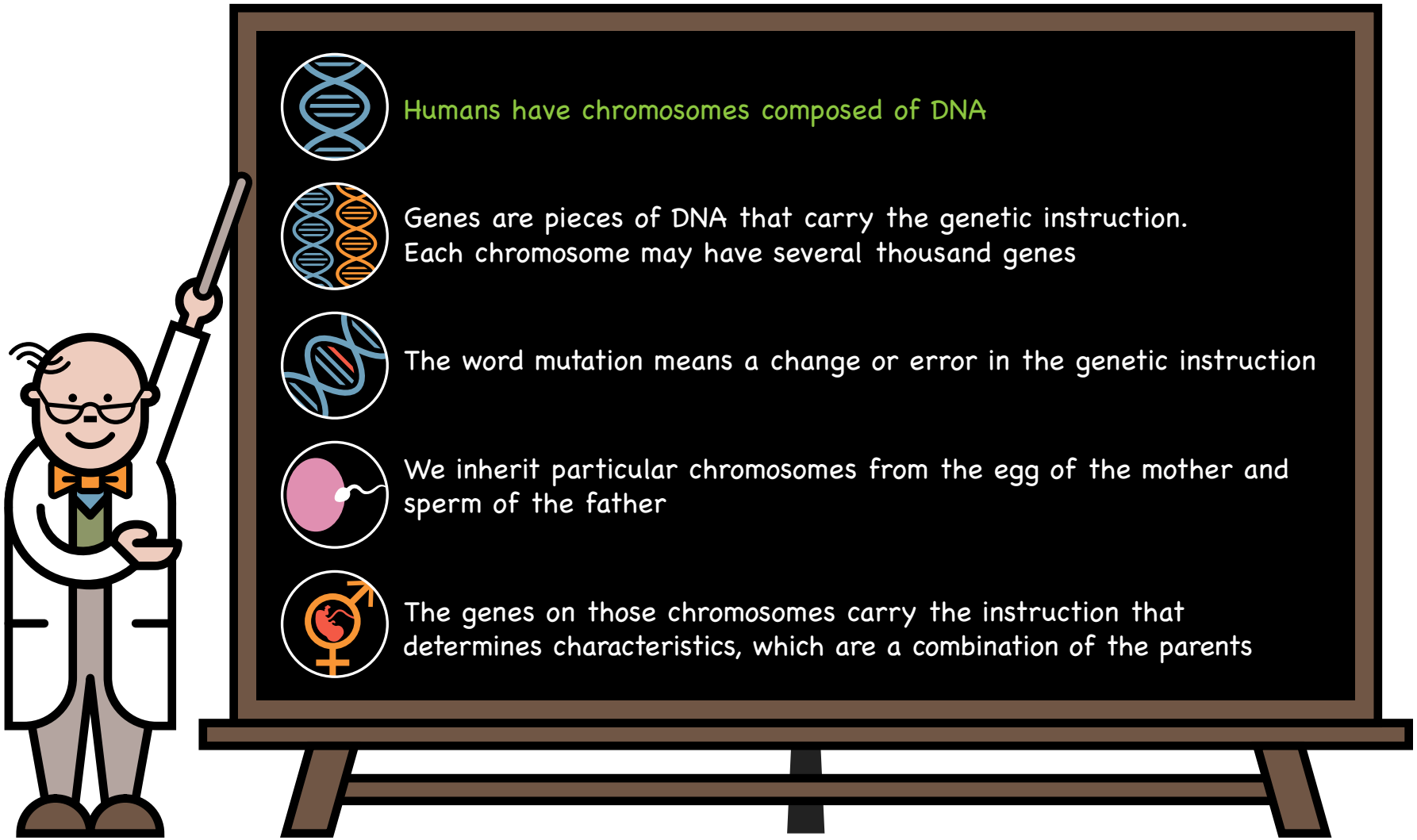
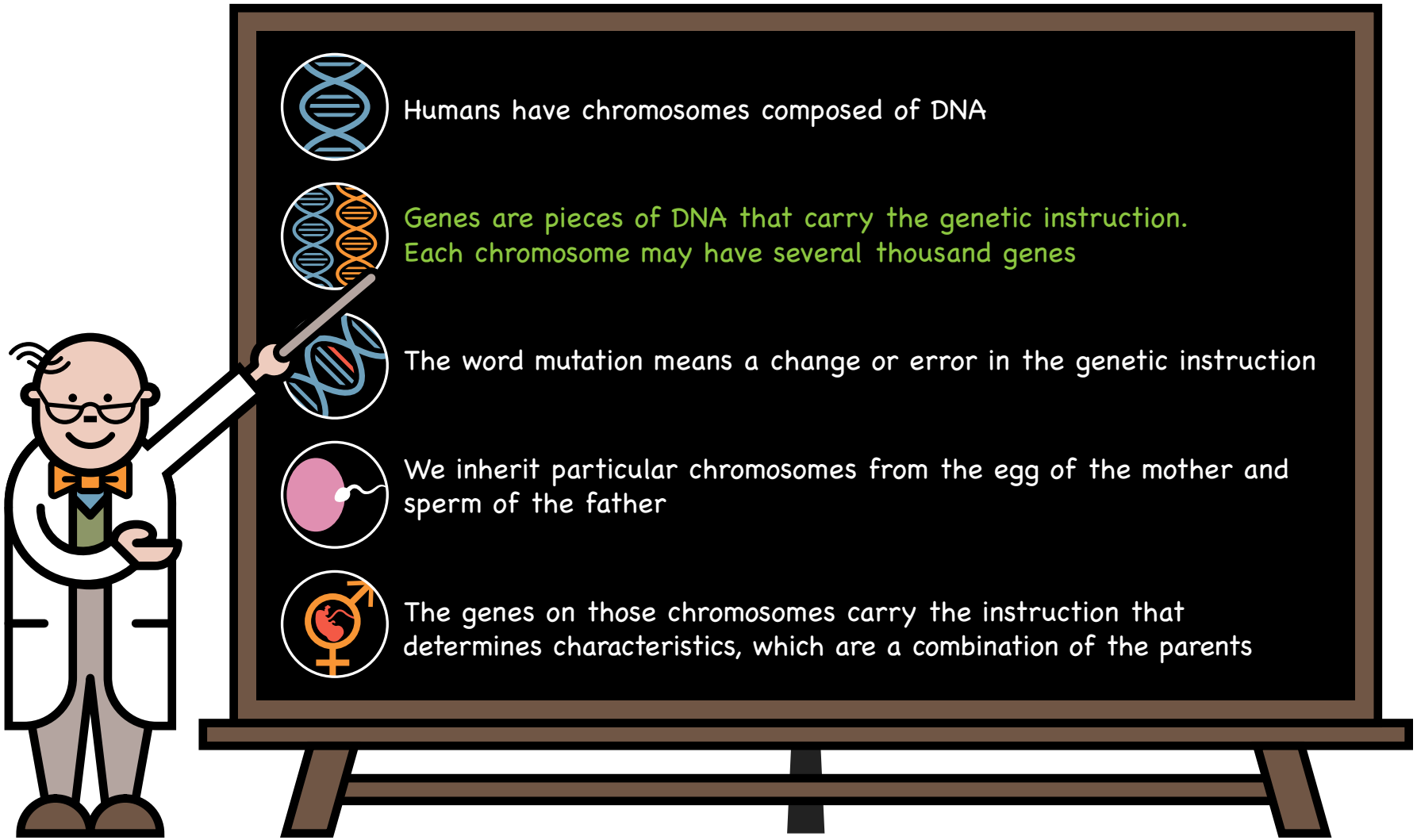


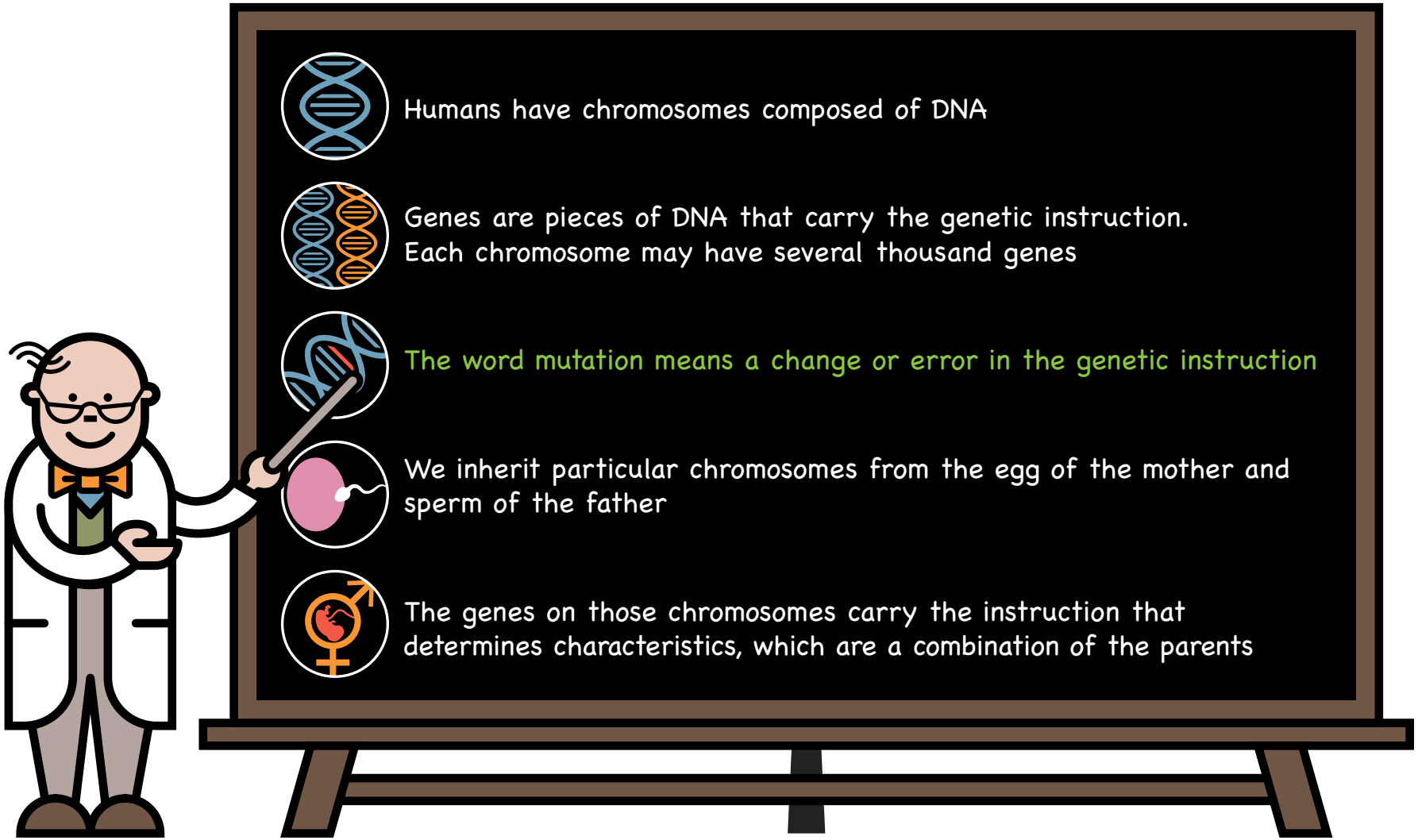
Chromosomes, genes, mutations



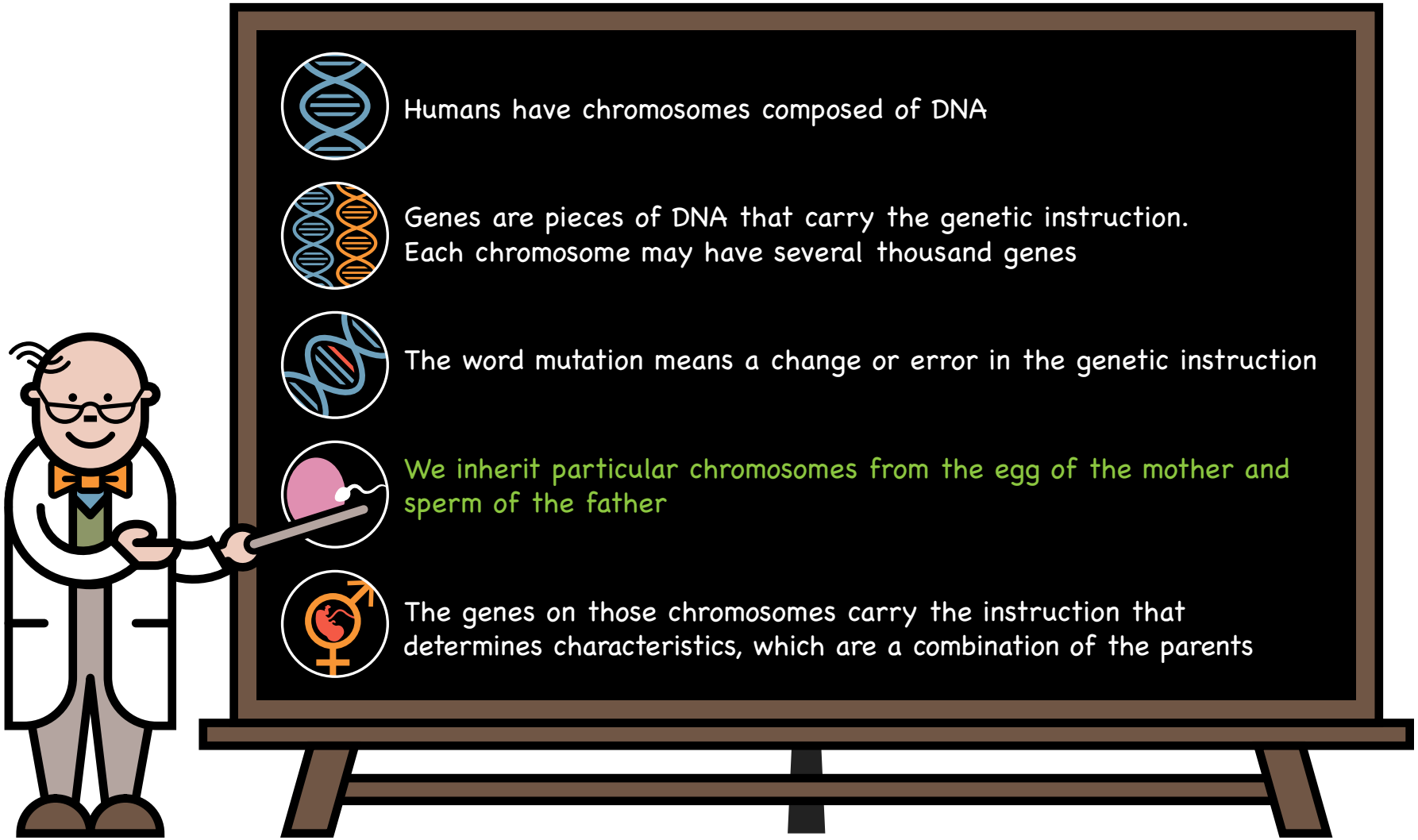
Chromosomes, genes, mutations



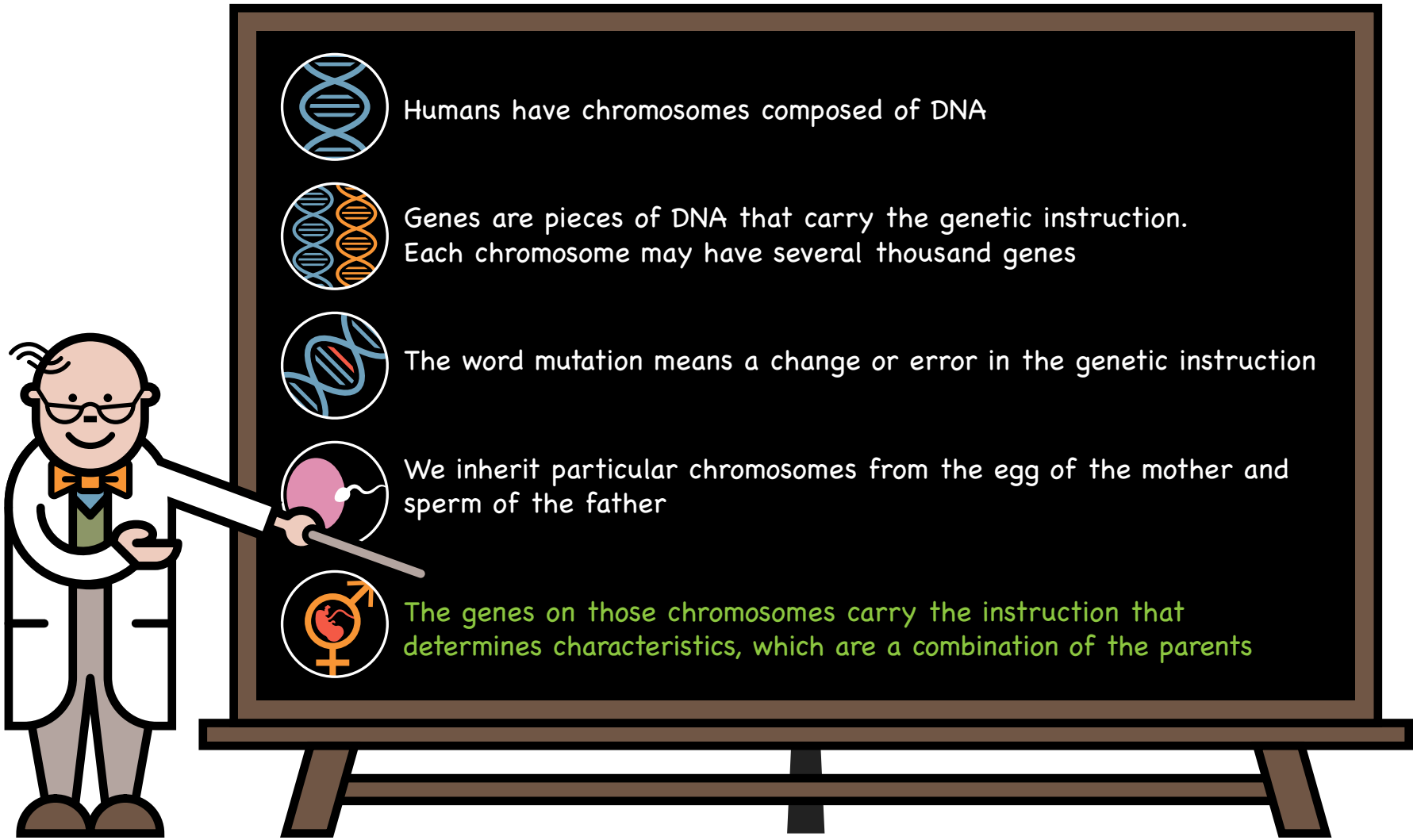
Chromosomes, genes, mutations



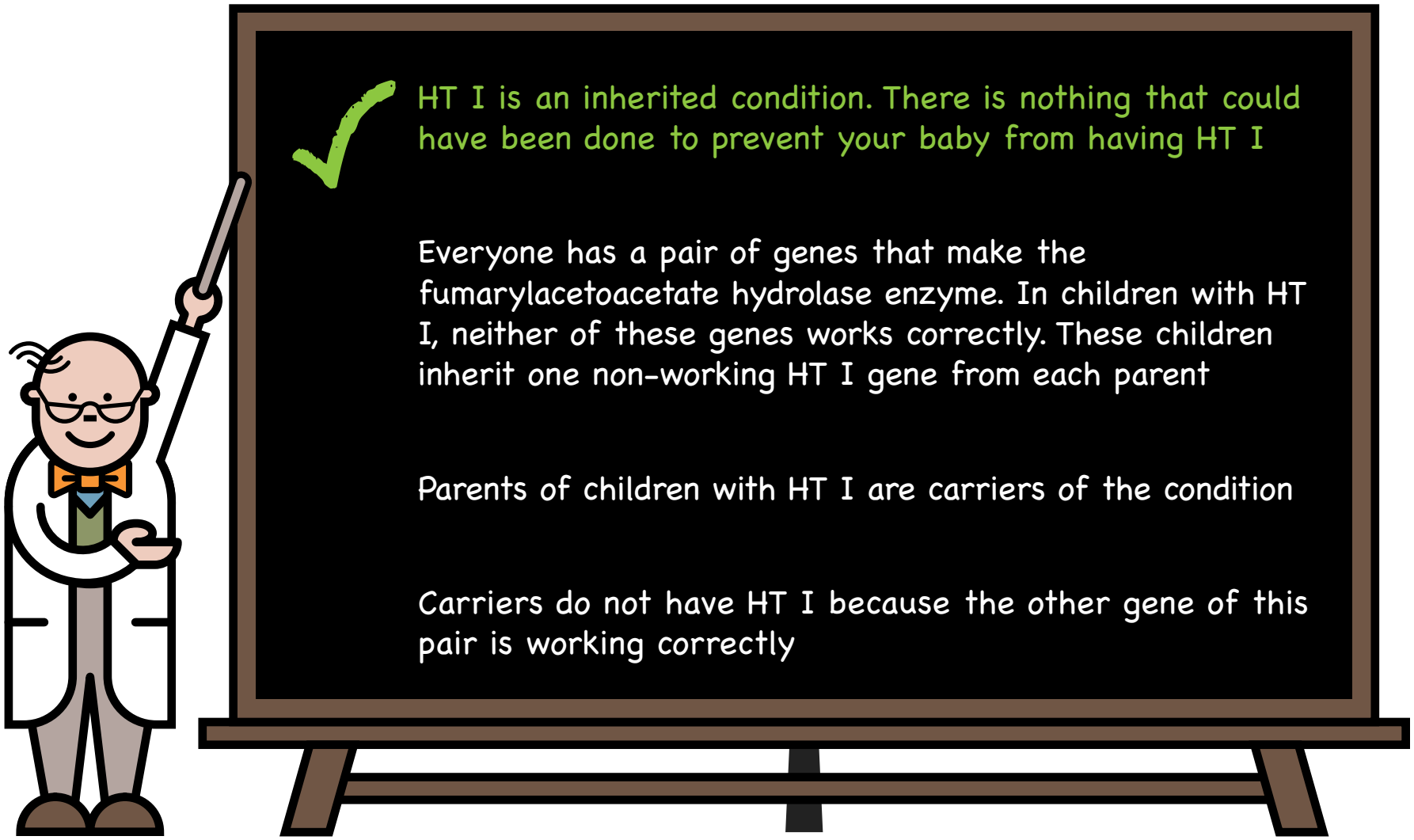
Chromosomes, genes, mutations



Chromosomes, genes, mutations



Inheritance



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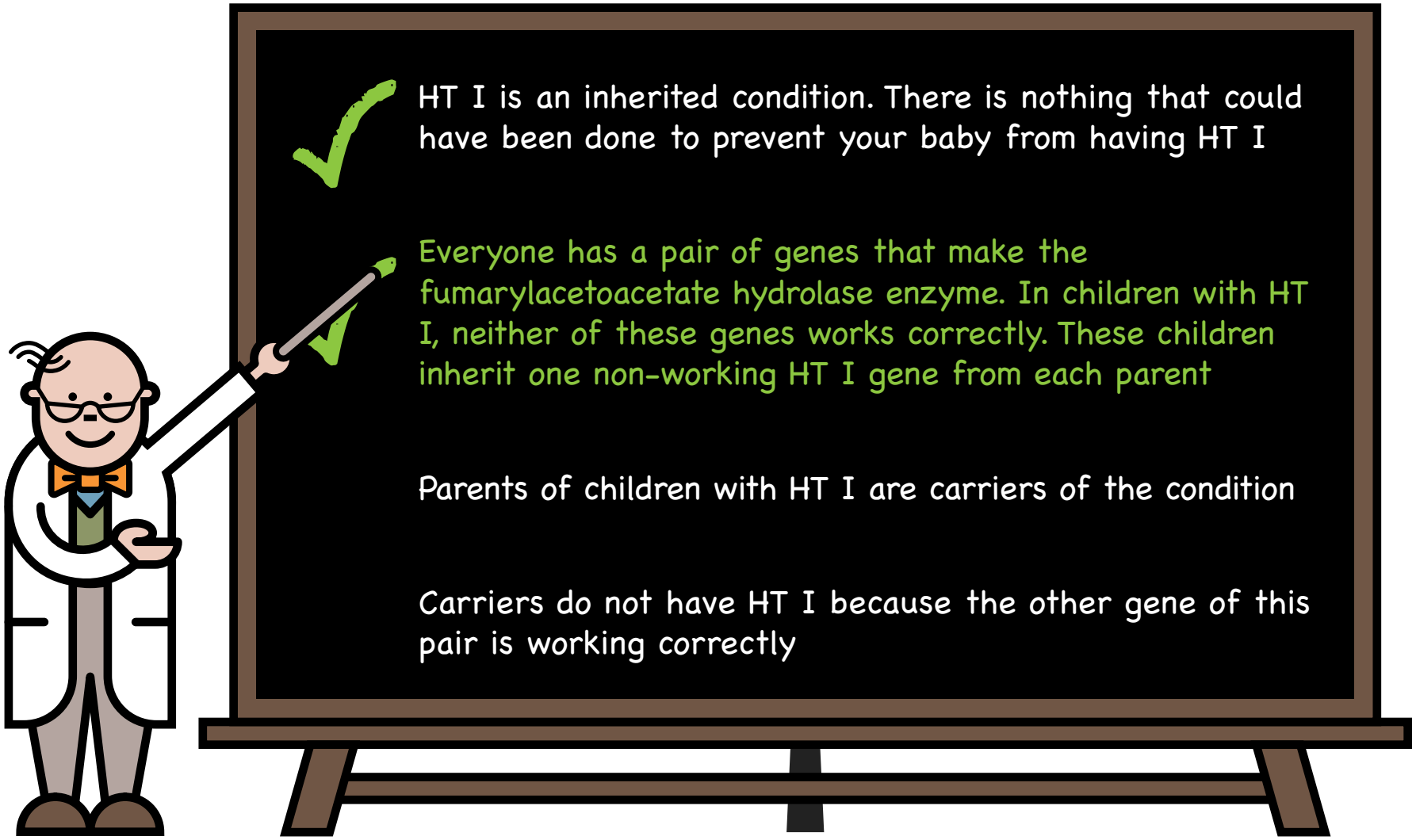

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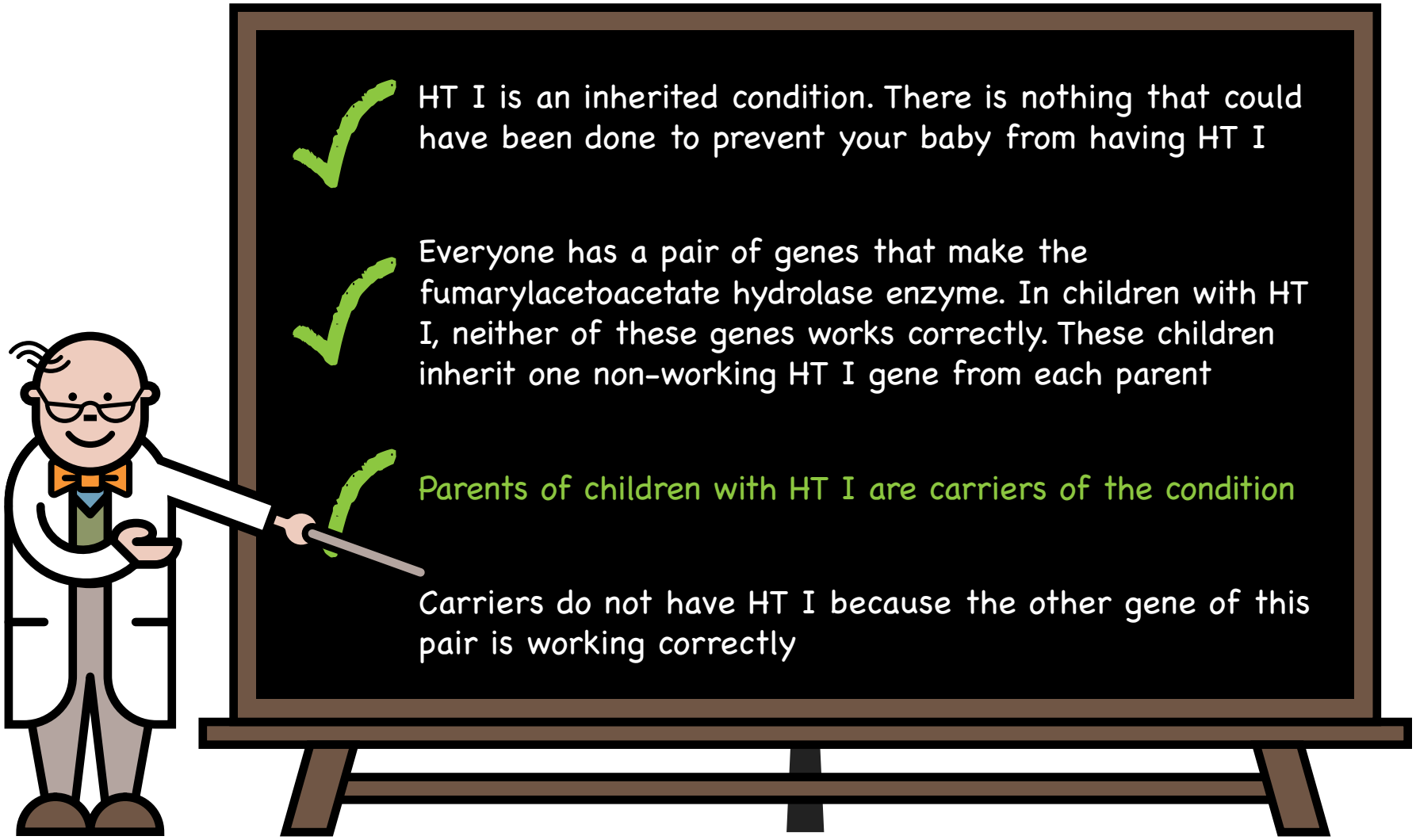
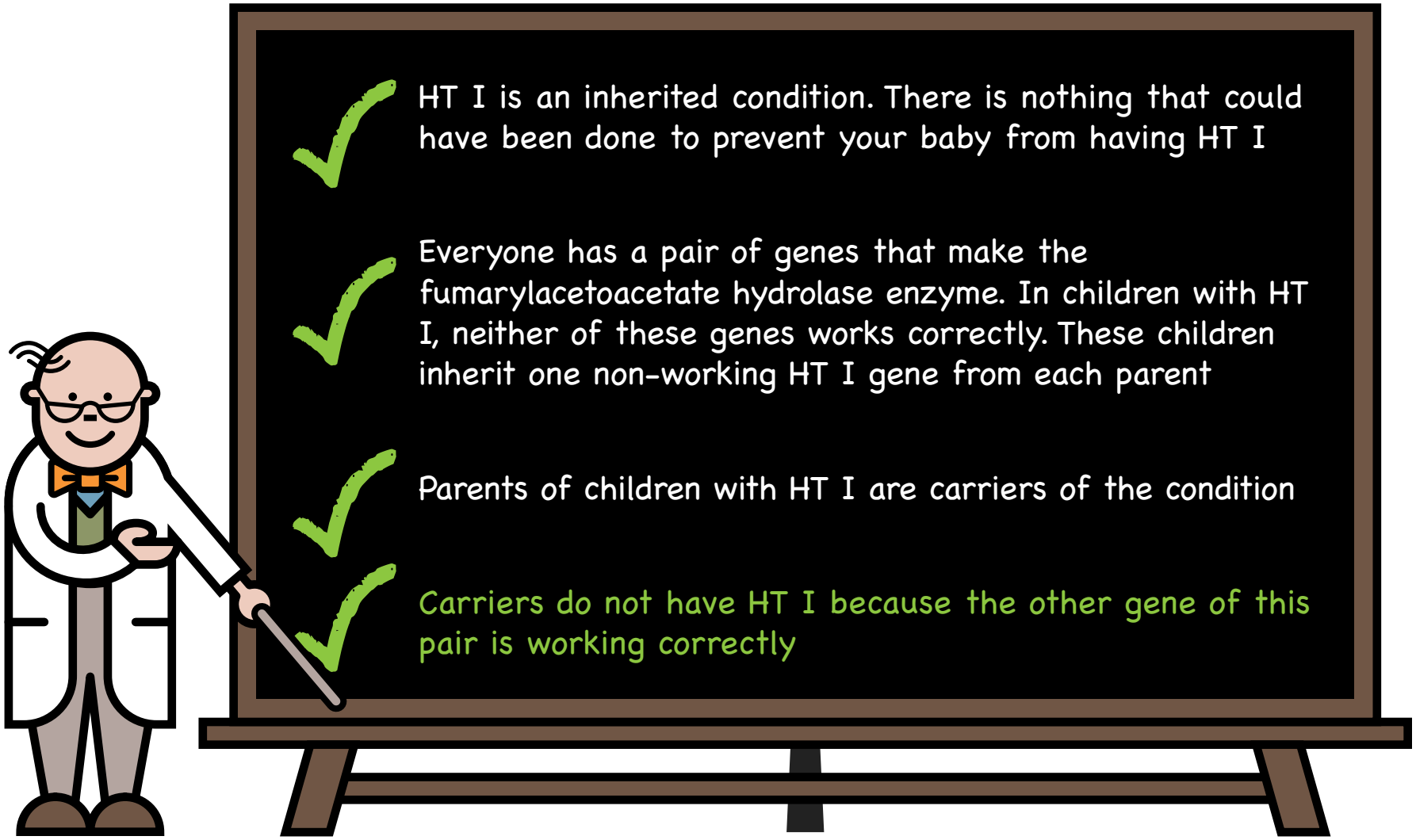
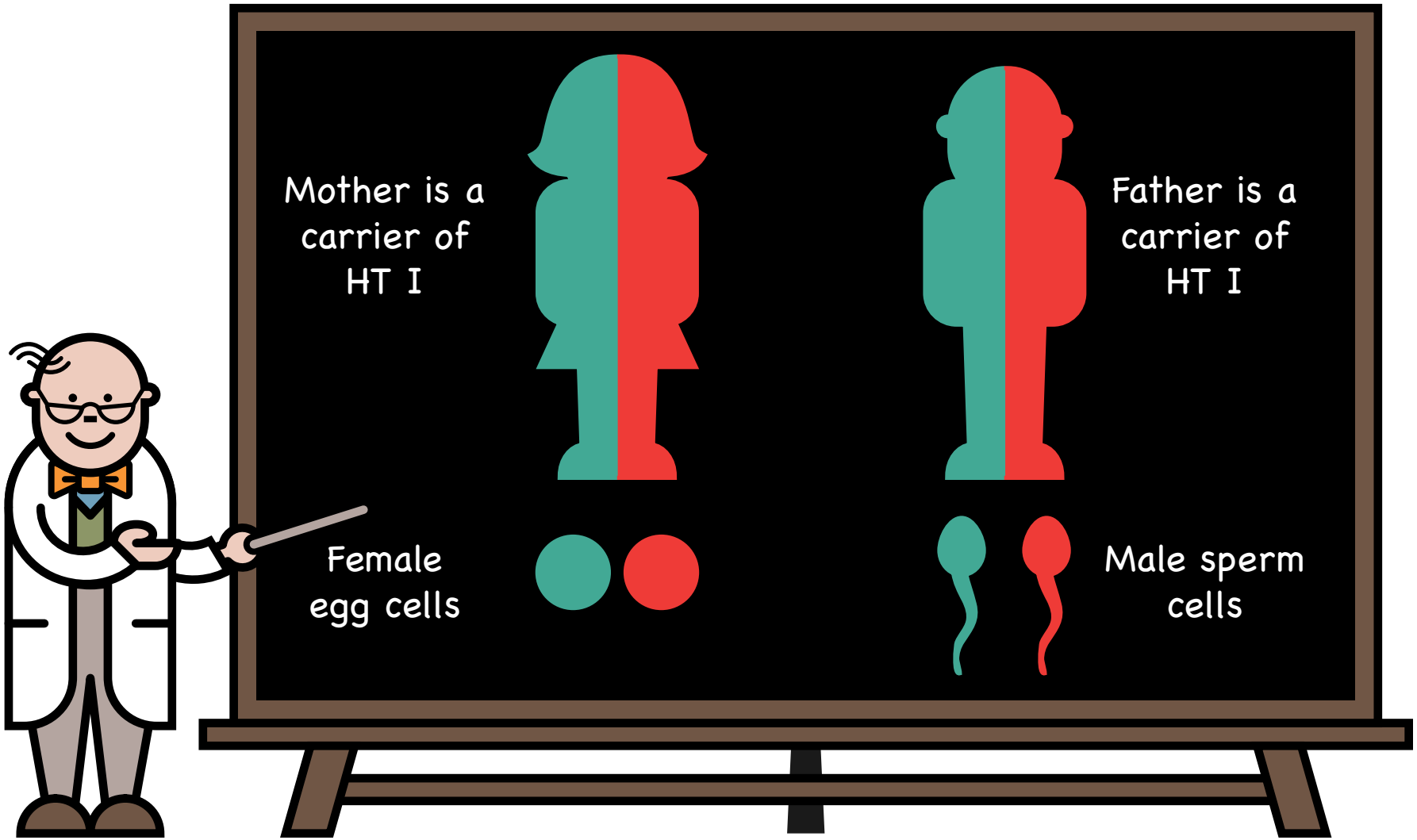


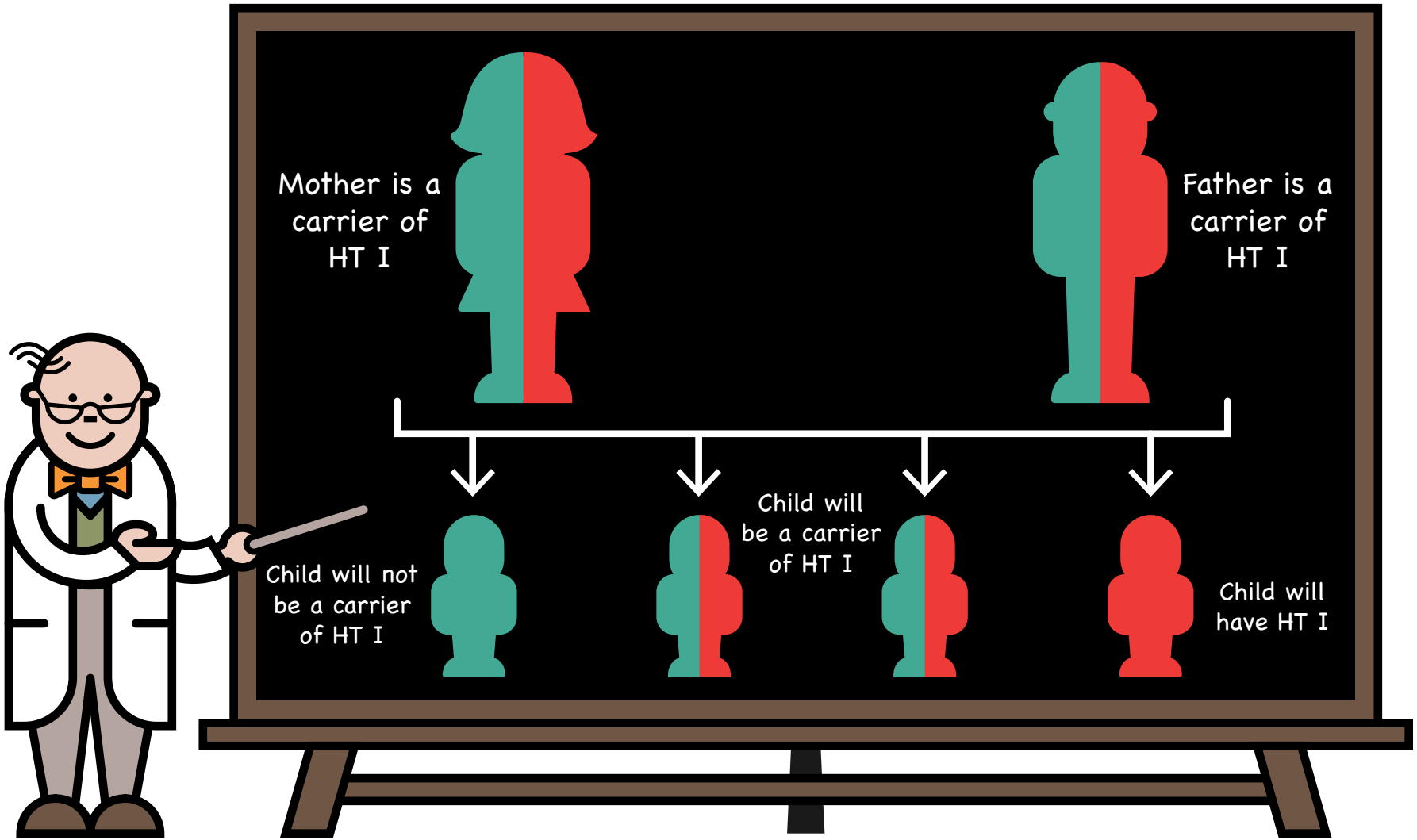
Inheritance



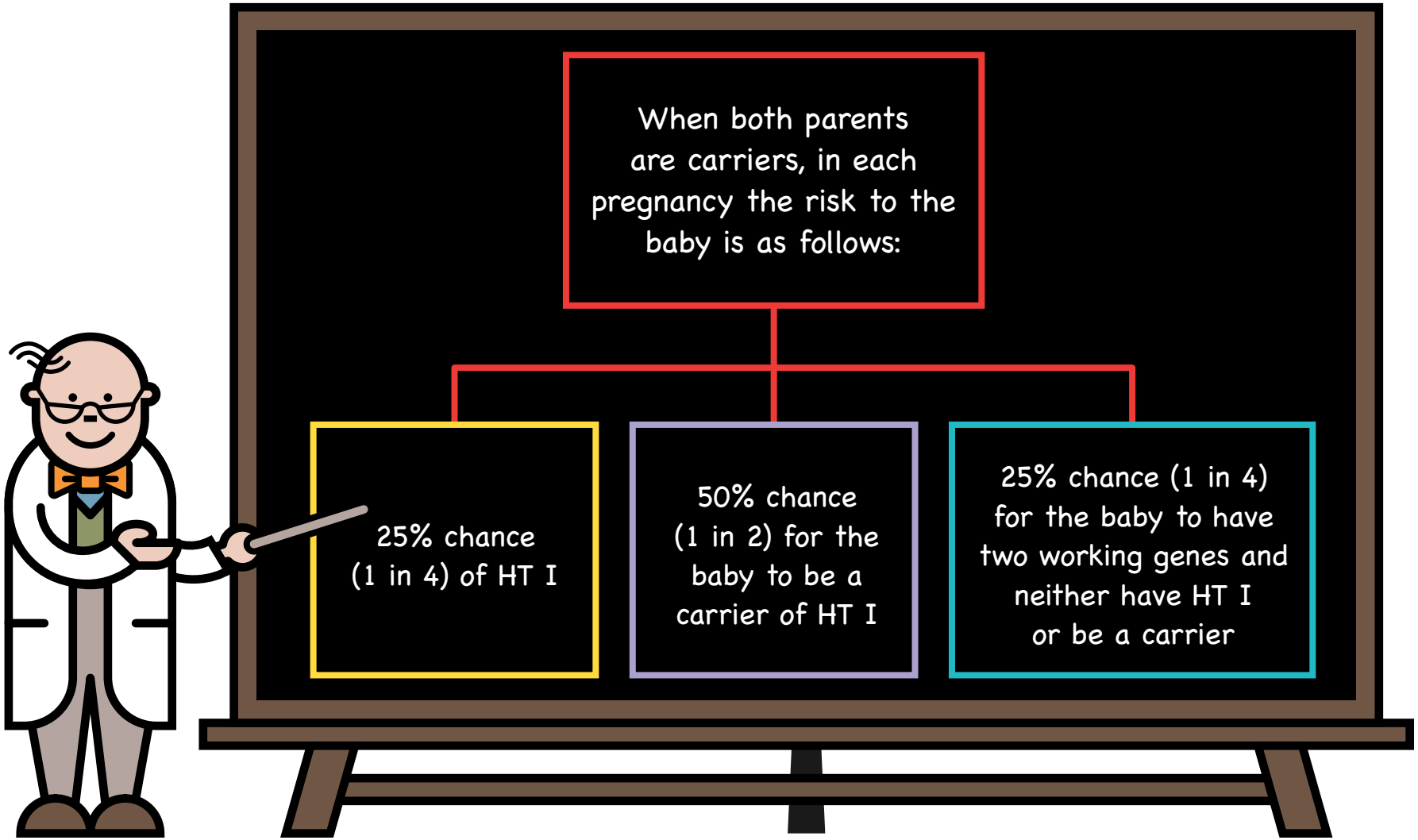
Inheritance – Autosomal recessive (carriers of HT I)



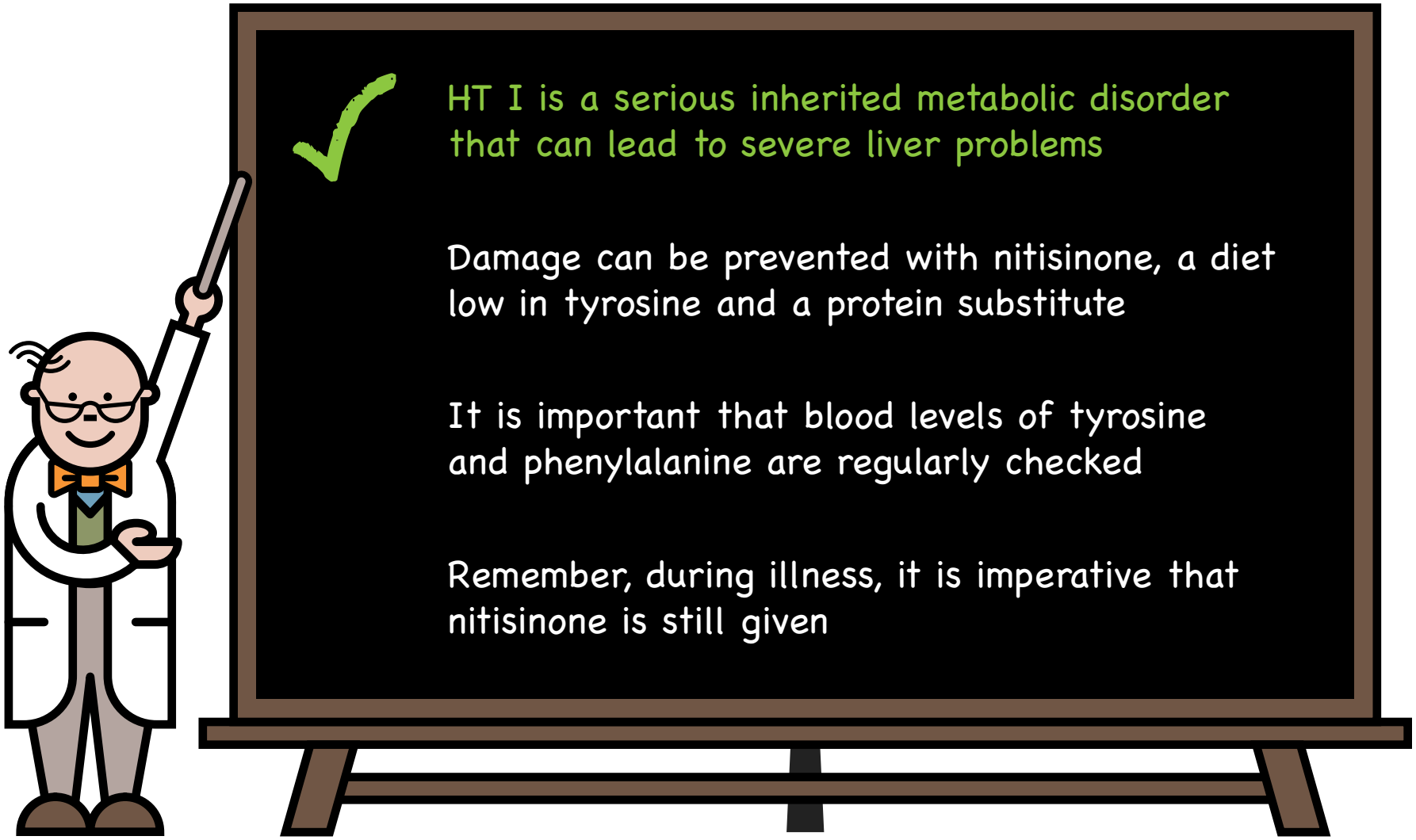
Inheritance – Autosomal recessive – possible combinations



Future pregnancies



Take home messages



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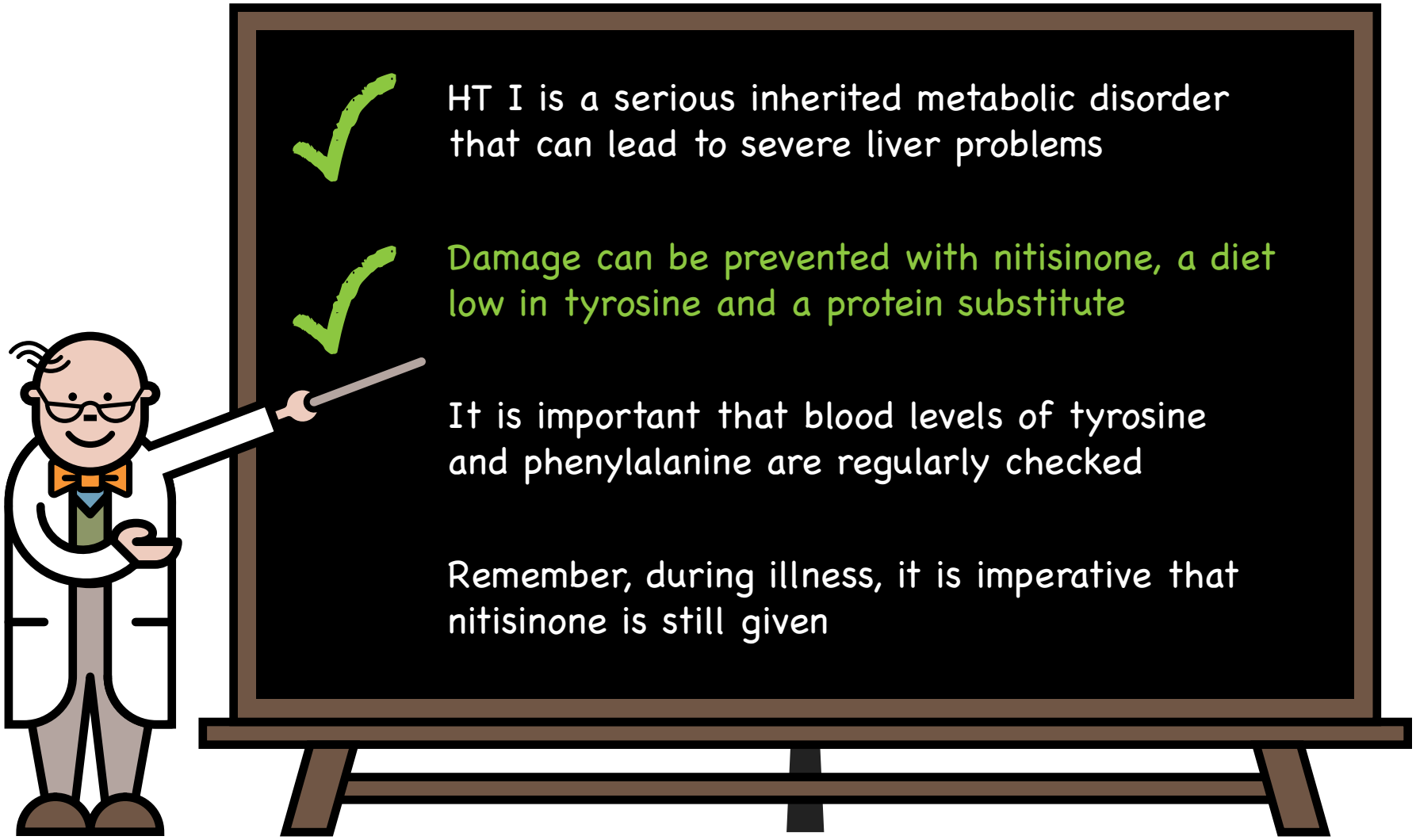

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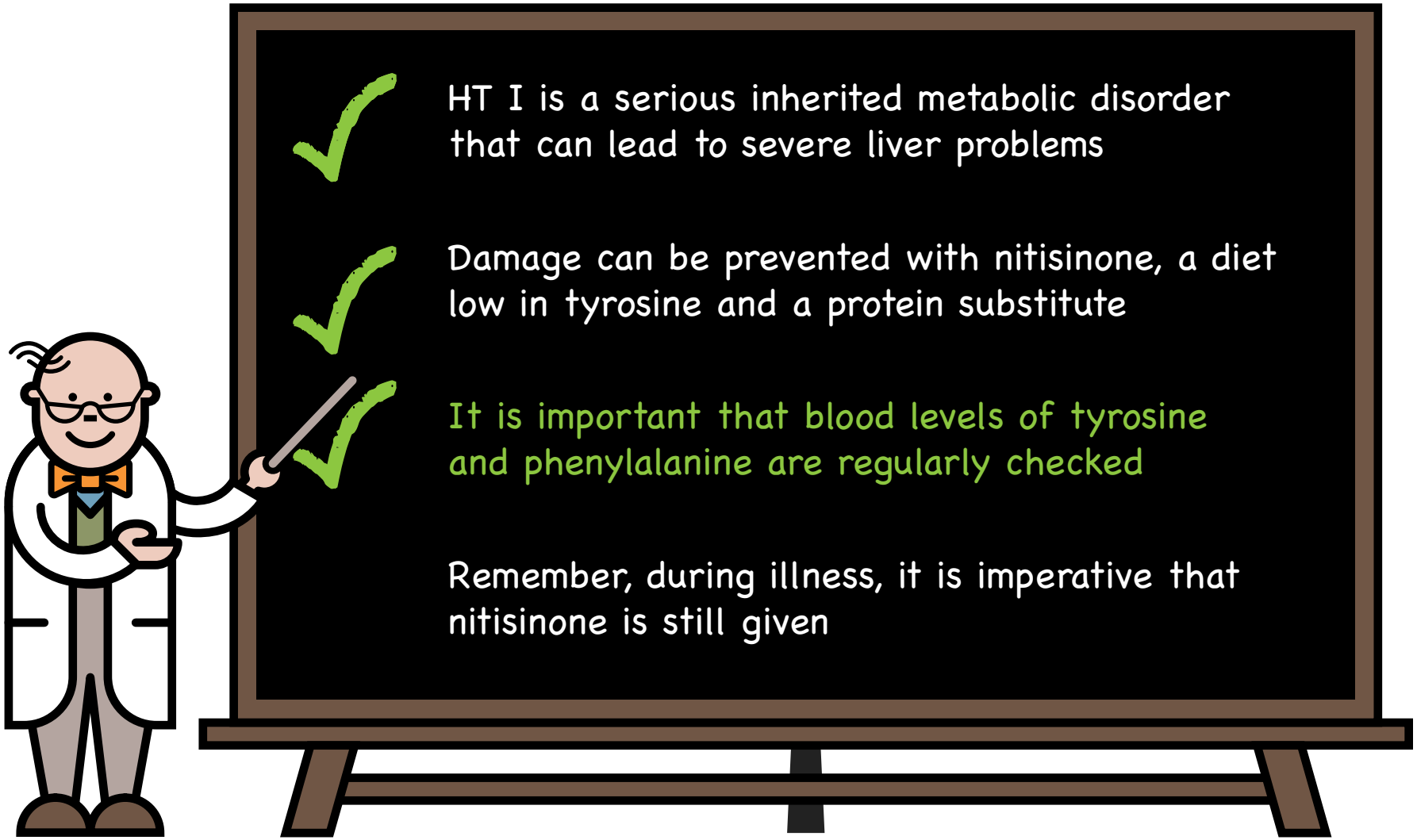

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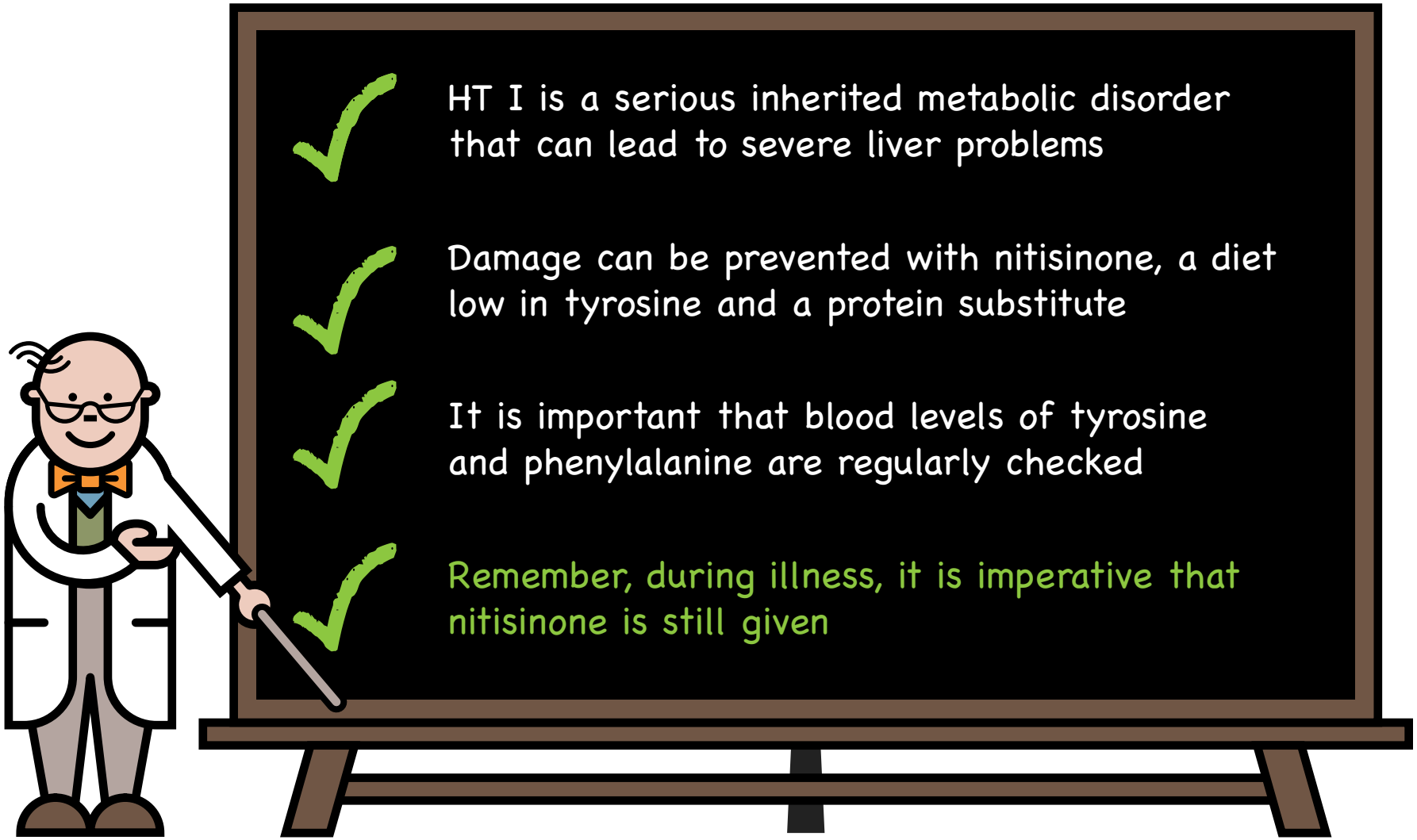

Helpful hints

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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LowProConnect
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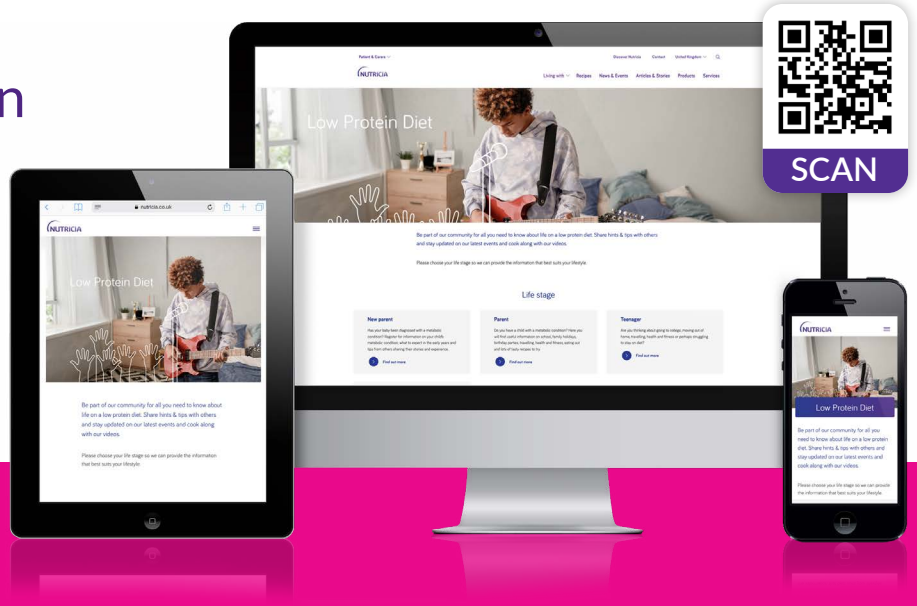

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