N.B TYR is also referred to as Hereditary Tyrosinaemia Type 1 (HT-1).

TYR-0619-V2
August 2019

# An introduction to Tyrosinaemia Type 1 (TYR)

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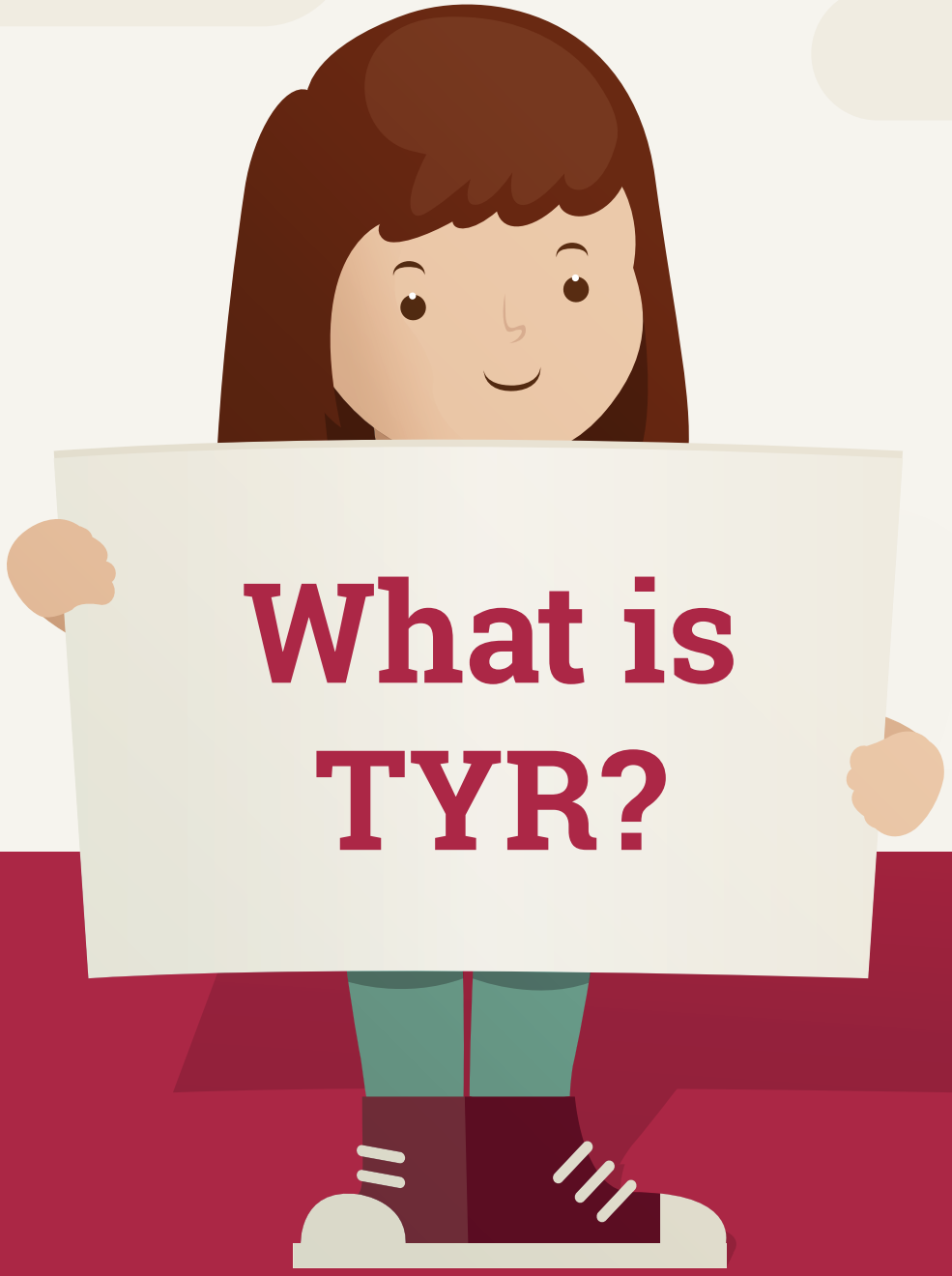
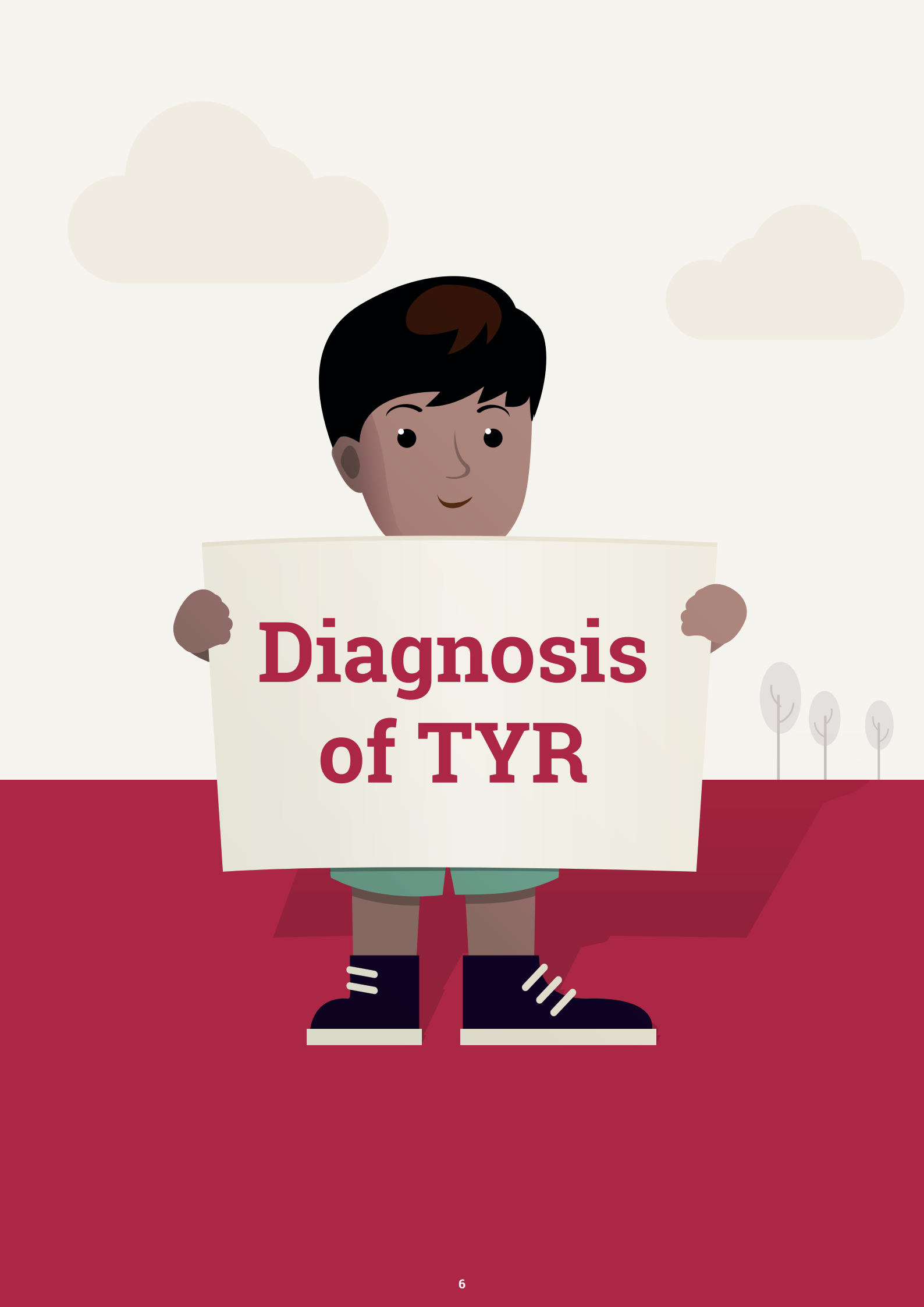
# Contents




**Tyrosinaemia Type 1**
(sounds like Ty-ro-sin-nee-mia)
or **TYR** for short.

# TYR is a manageable condition which affects the way the body breaks down protein.

- Children with TYR have a defect in the process which breaks down part of a protein called Tyrosine.
- If left untreated, products in the blood build up to toxic levels which can lead to organ failure.
- This can be avoided by following a special diet alongside drug treatment.
- With management, people with TYR can attend university, build successful careers and enjoy a happy family life the same as everyone else.

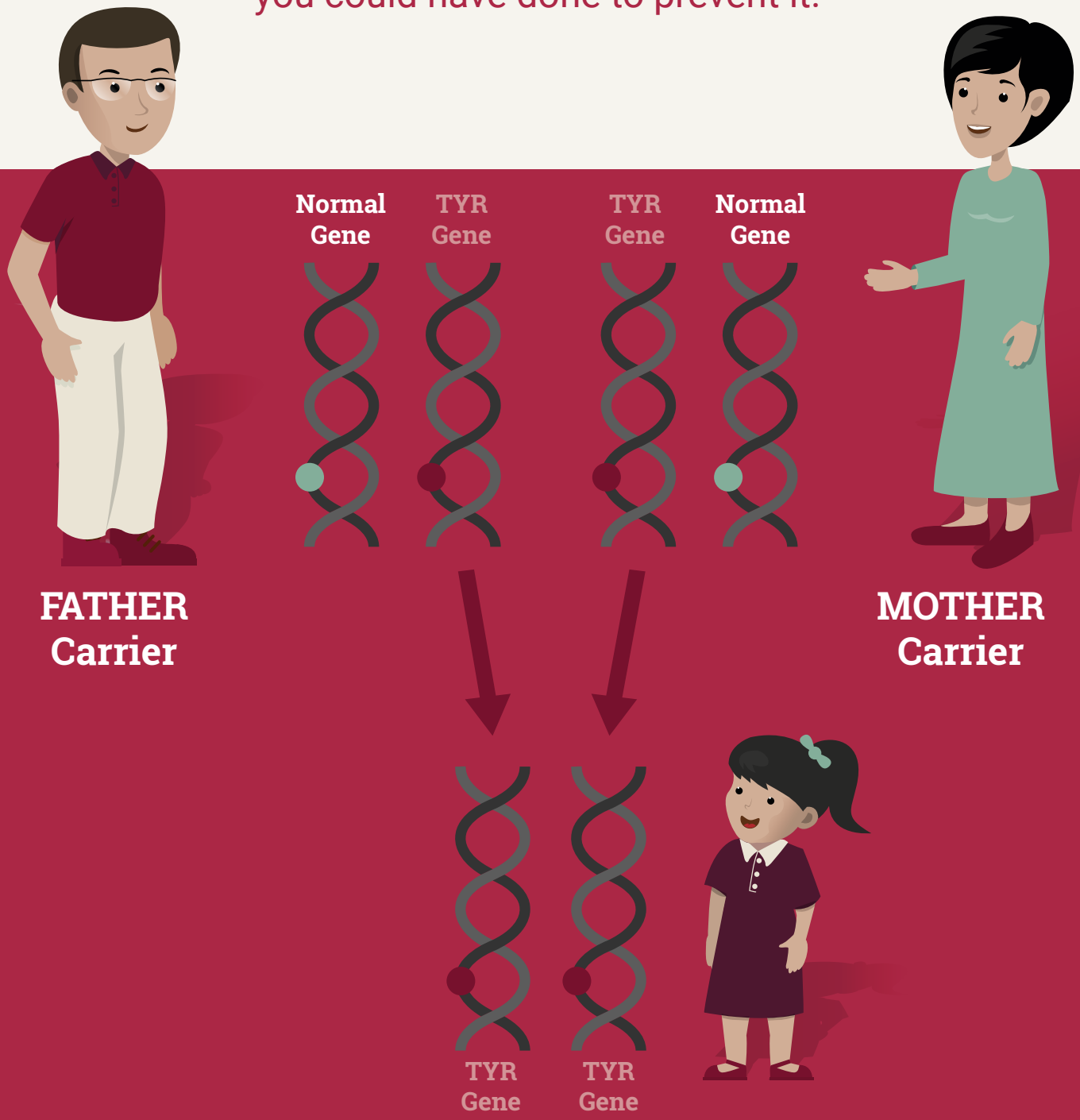

Diagnosis
of TYR

Children with TYR are identified through a blood test. Special dietary management and careful monitoring can improve long term outcomes. If treated effectively and early, children with TYR can do well.


Why does my child have TYR?

# TYR is an inherited condition

It's nobody's fault and there is nothing you could have done to prevent it.



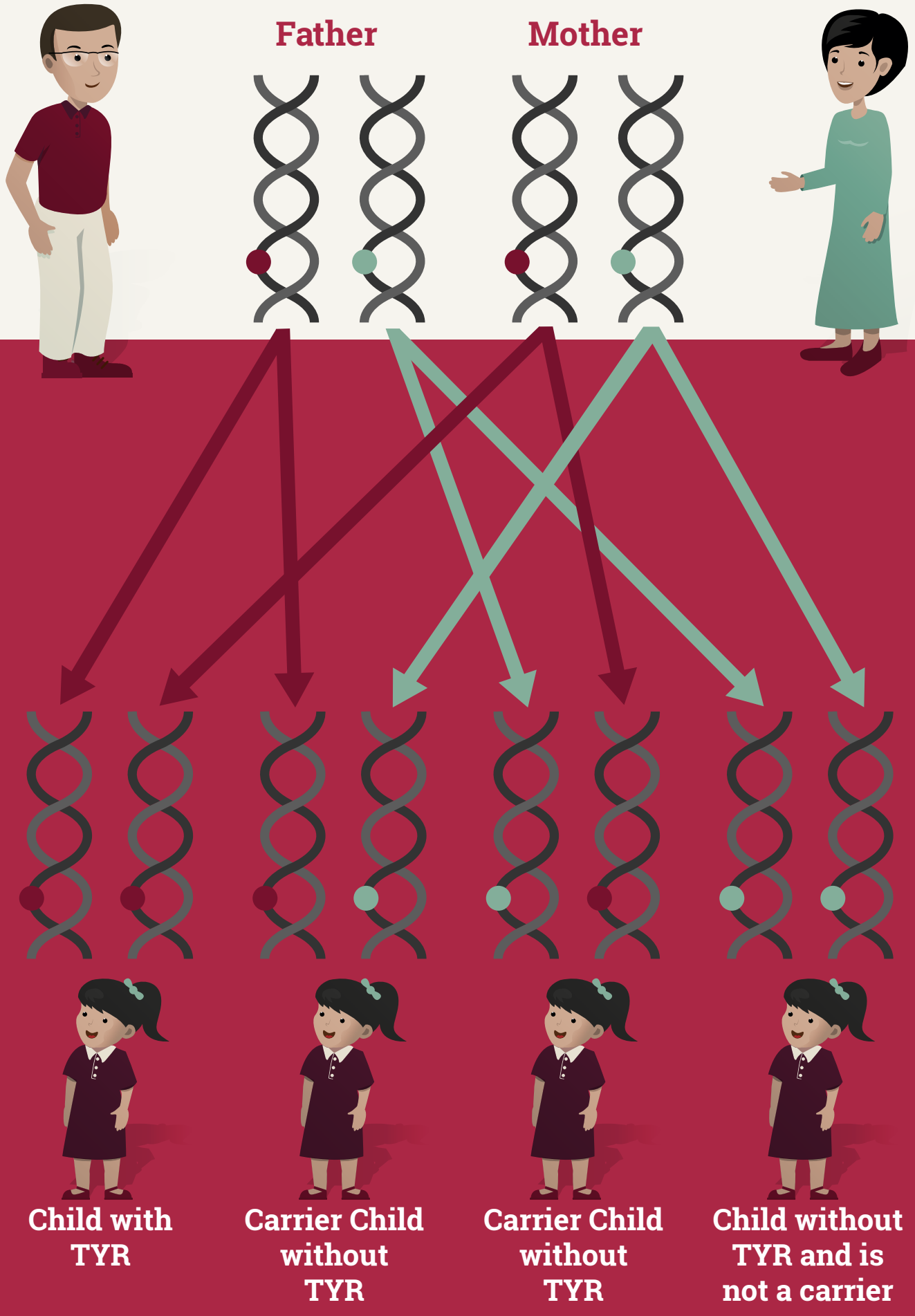

## Child with TYR

As a parent of a child with TYR, you have one TYR gene and one normal gene. This is known as being a carrier.

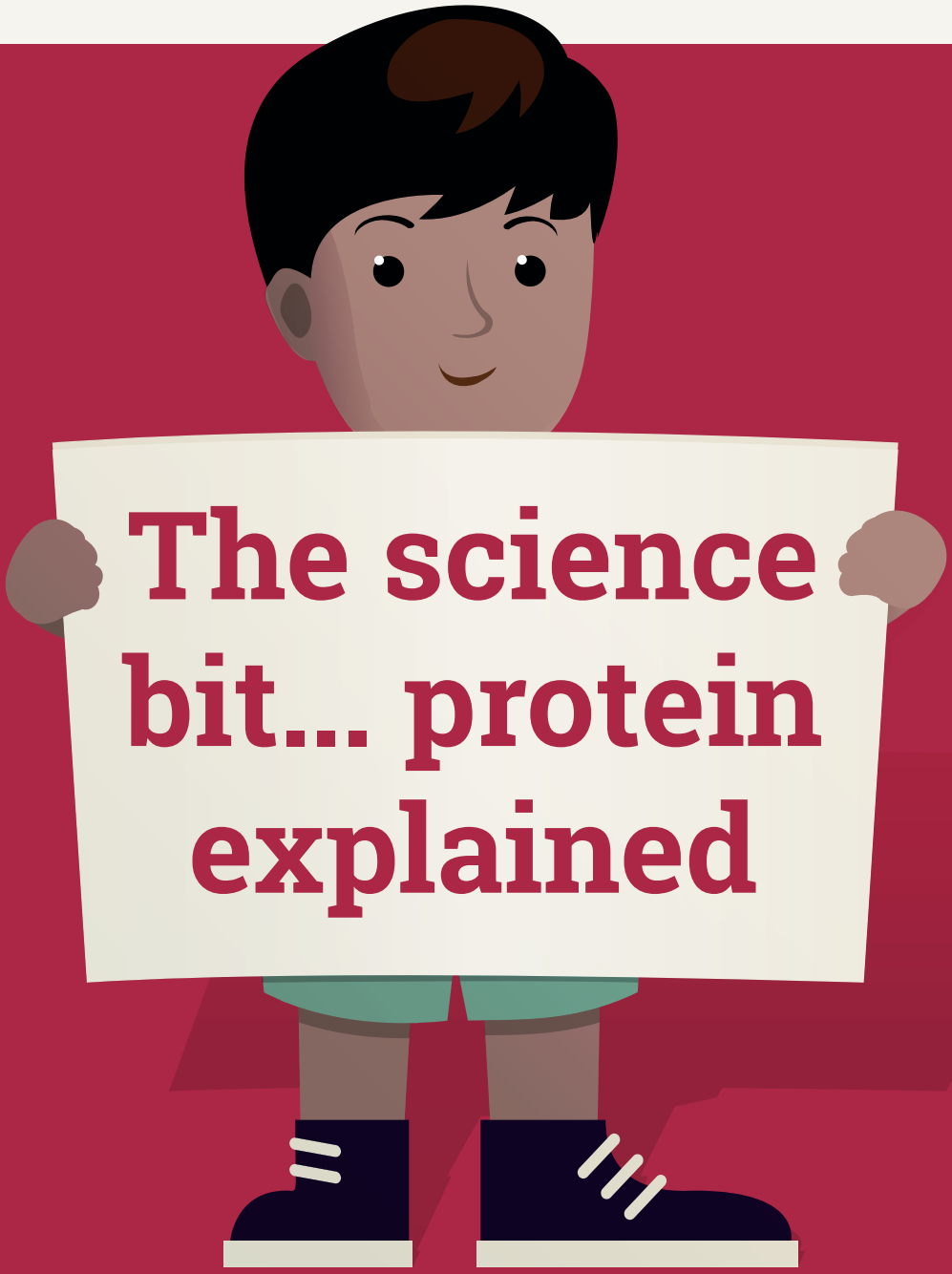
Your child has inherited 2 TYR genes, one from mum and one from dad.

# There are a small number of people who are carriers of the TYR gene

People who are carriers for TYR do not have TYR themselves and the faulty gene does not cause a problem to them.




When 2 people who are carriers of the TYR gene conceive a child there is a 1 in 4 chance of that child having TYR.


The science bit... protein explained

# Why do we need protein?

Protein is a nutrient needed by the body.

It helps to build, repair and maintain body cells and tissues, like your skin, muscles, organs, blood and even bones.

When protein is eaten, it is broken down in the body (during digestion) into smaller pieces (like building blocks). These smaller pieces are called amino acids.

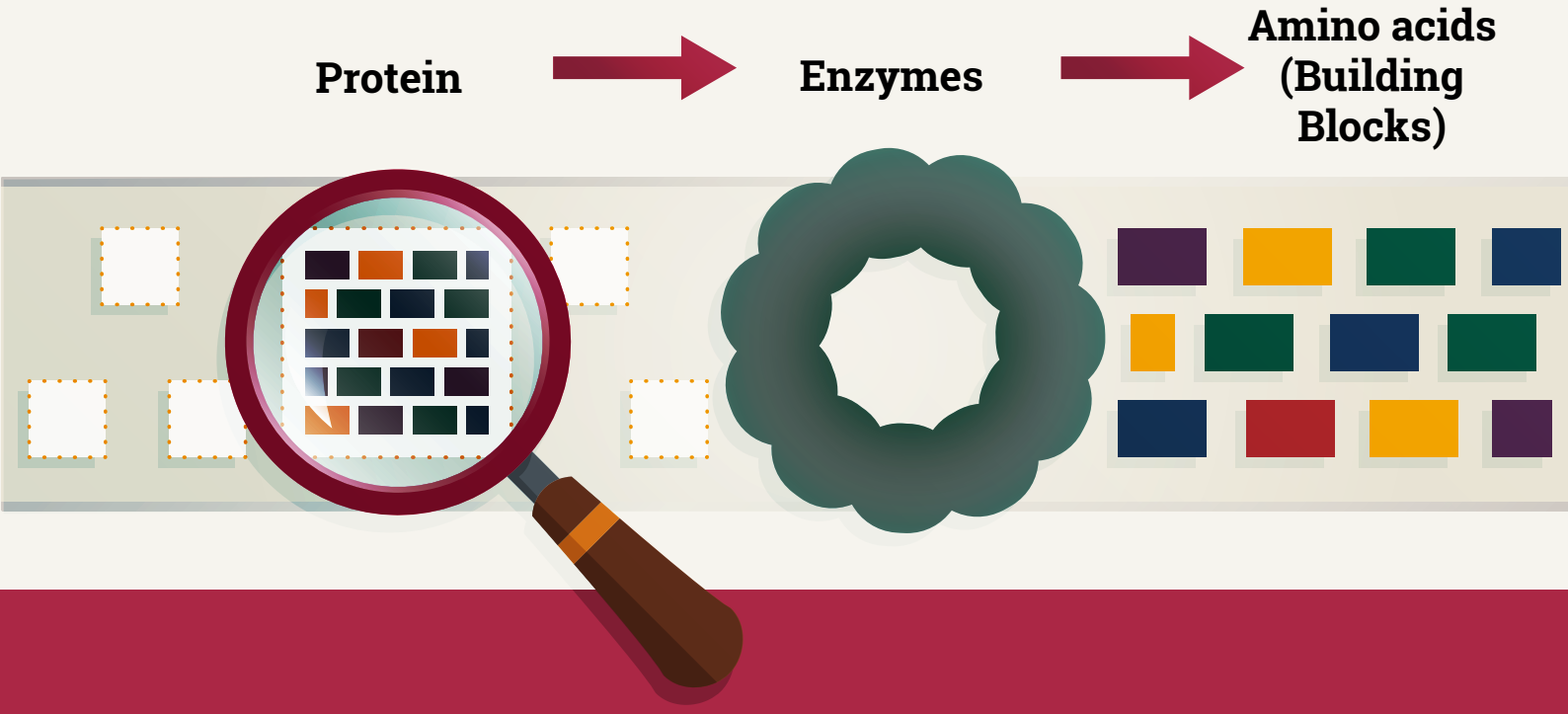
# Protein is made up of many building blocks called amino acids


There are 20 amino acids (building blocks) that make up protein. Some of these are ESSENTIAL and cannot be made in the body — so they must come from the food we eat.

Tyrosine and Phenylalanine (Phe) are two of these 20 amino acids. It is these particular amino acids that a person with TYR cannot process correctly.

# What normally happens

## The conversion of protein into amino acids

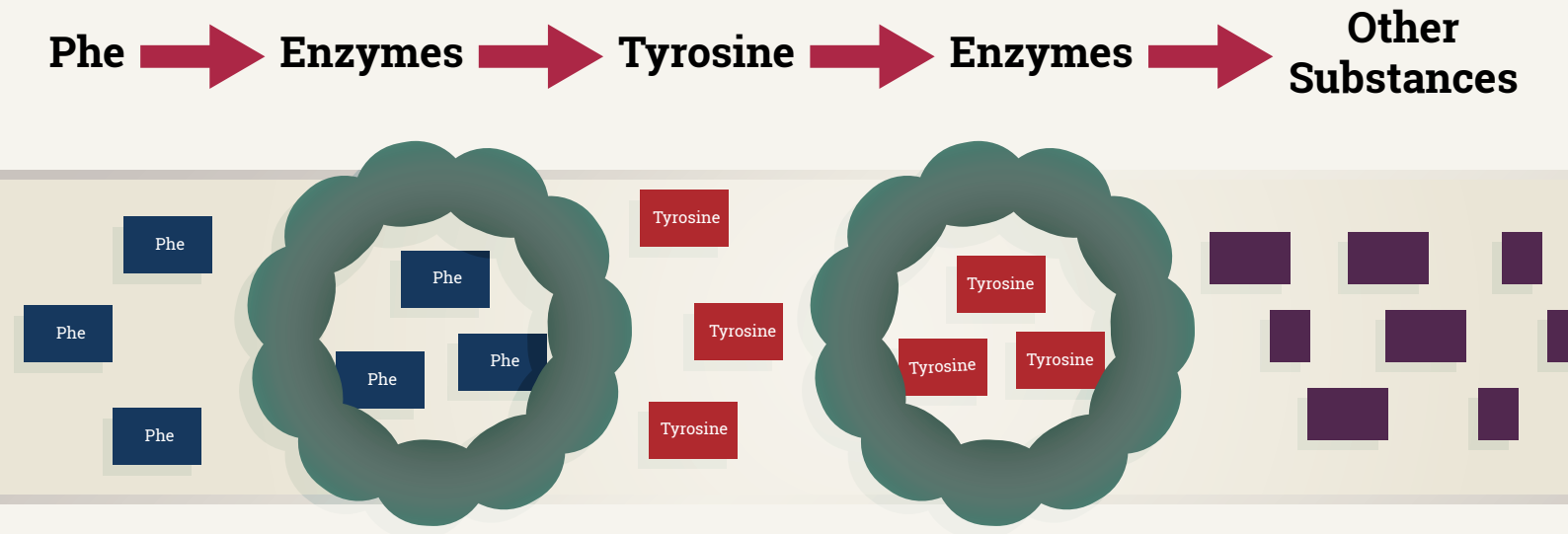



Enzymes are like chemical scissors needed for the breakdown of protein into amino acids.

The body uses these amino acids for growing, building up muscle and helping the body stay healthy.

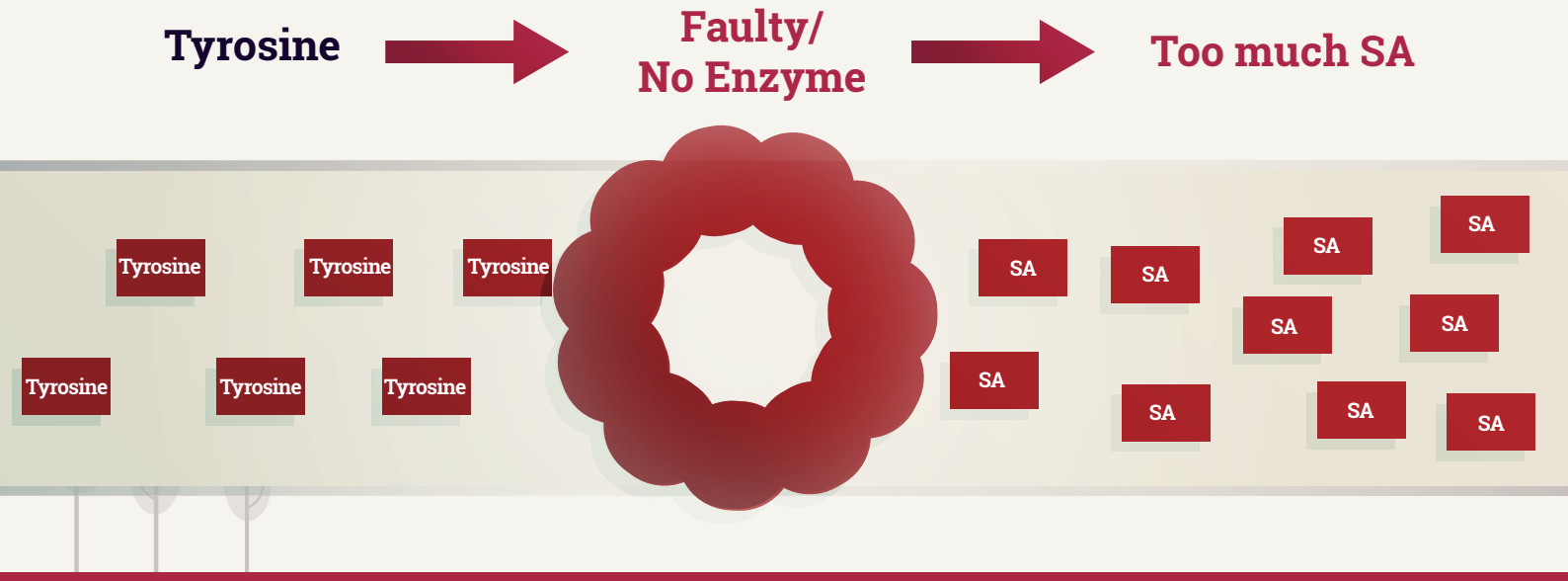

Why can someone with TYR not process Phe & Tyrosine correctly?

# In a person without TYR, Phe is broken down to Tyrosine and then Tyrosine is converted to other substances

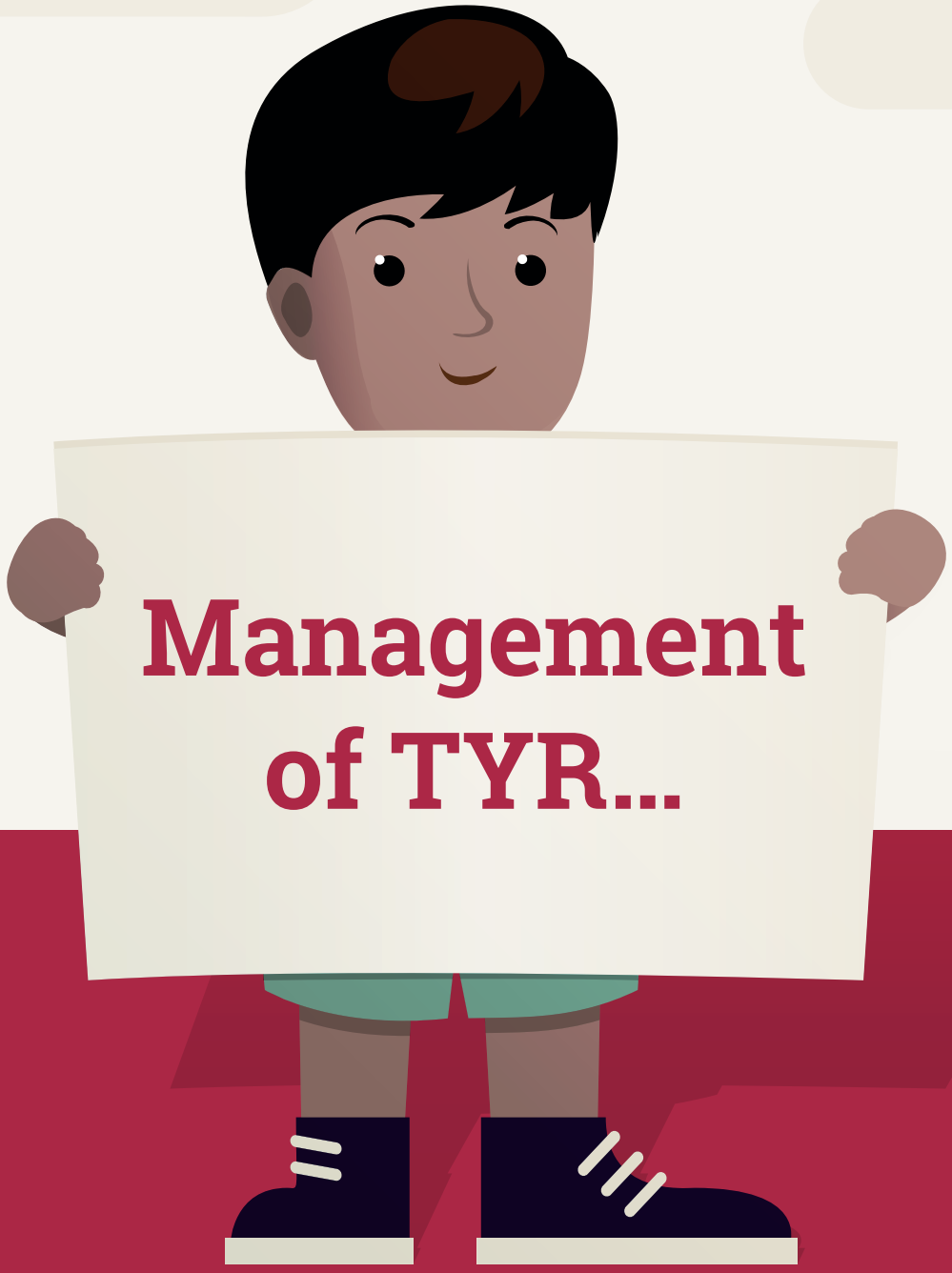



Normally, the liver produces a number of enzymes which break down Tyrosine into other substances that have important functions in the body.

# In a person with TYR, the Tyrosine can not be broken down as it should be.




In TYR, the enzyme fumarylacetoacetate hydrolase (FAH) is not made by the liver or does not work properly. This means that Tyrosine is not broken down properly and a substance called Succinylacetone (SA) builds up to toxic levels in the blood and cause the symptoms of TYR. This is the reason a special diet low in protein (Phe and Tyrosine) needs to be followed.



# Management of TYR...

There are two important parts to the management:

1. Medication called Nitisinone, which prevents the accumulation of SA.
2. Low protein diet.

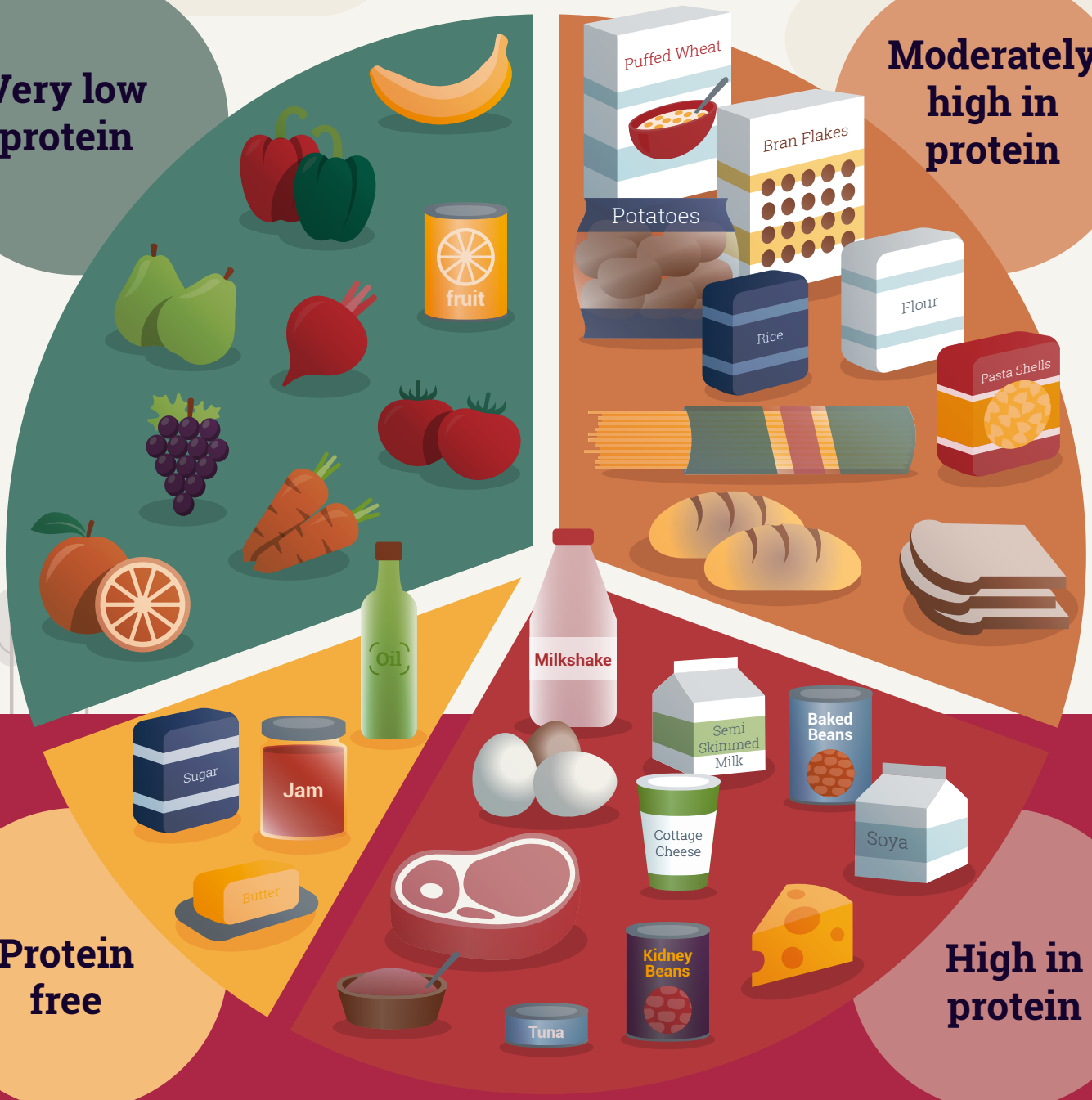
# A Regular Diet

**Fruit and Vegetables**

**Very low protein**

**Carbohydrate**

**Moderately high in protein**




**Protein free**

**High in protein**

**Fat and sugar**

**Protein**

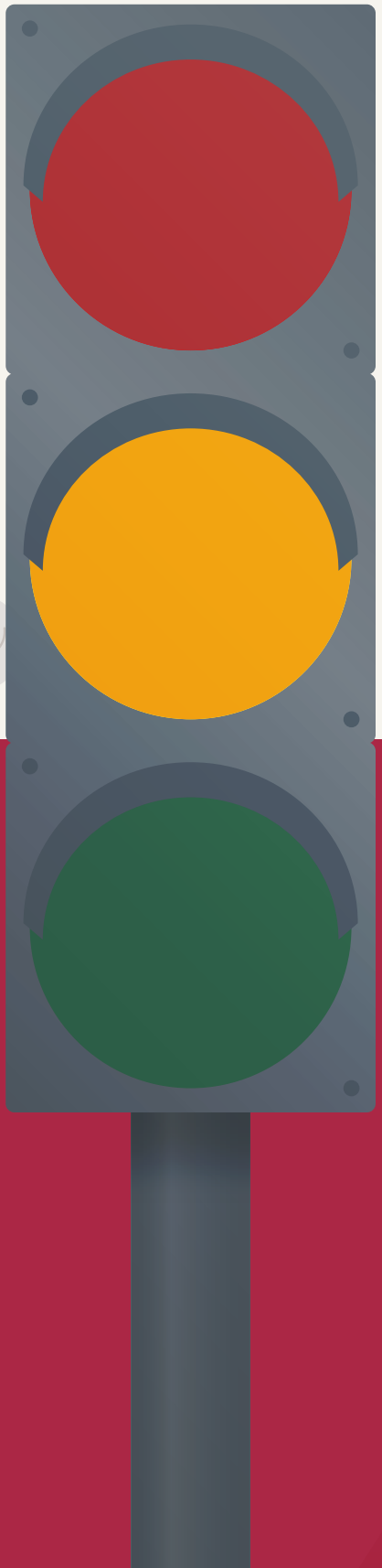
A regular diet has too much protein for a person with TYR. Therefore they must follow a special diet.

# TYR is managed by following a strict low protein diet

- Tyrosine and Phe are found in all protein containing foods, e.g. meat, fish, eggs, cheese, quorn, nuts, soya, pulses and in foods such as bread, biscuits and chocolate.
- In order to manage TYR, all protein foods **must be restricted** and a low protein diet must be followed.
    - All amino acids have a role or function in the body. Tyrosine and Phe help us with our growth, development and tissue repair.
    - It is important that someone with TYR gets some Tyrosine and Phe, but not so much that it becomes harmful.

# Traffic Light System

The low protein diet that a person with TYR must follow is best described by using the traffic light system:



## RED STOP!

Do not eat these foods

**Too high in protein / Tyrosine and Phe**

## AMBER Go Cautiously!

These foods can be eaten in measured amounts (exchanges)

**Contains some protein / Tyrosine and Phe**

## GREEN

These foods can be eaten freely

**Very low in / free from protein / Tyrosine and Phe**

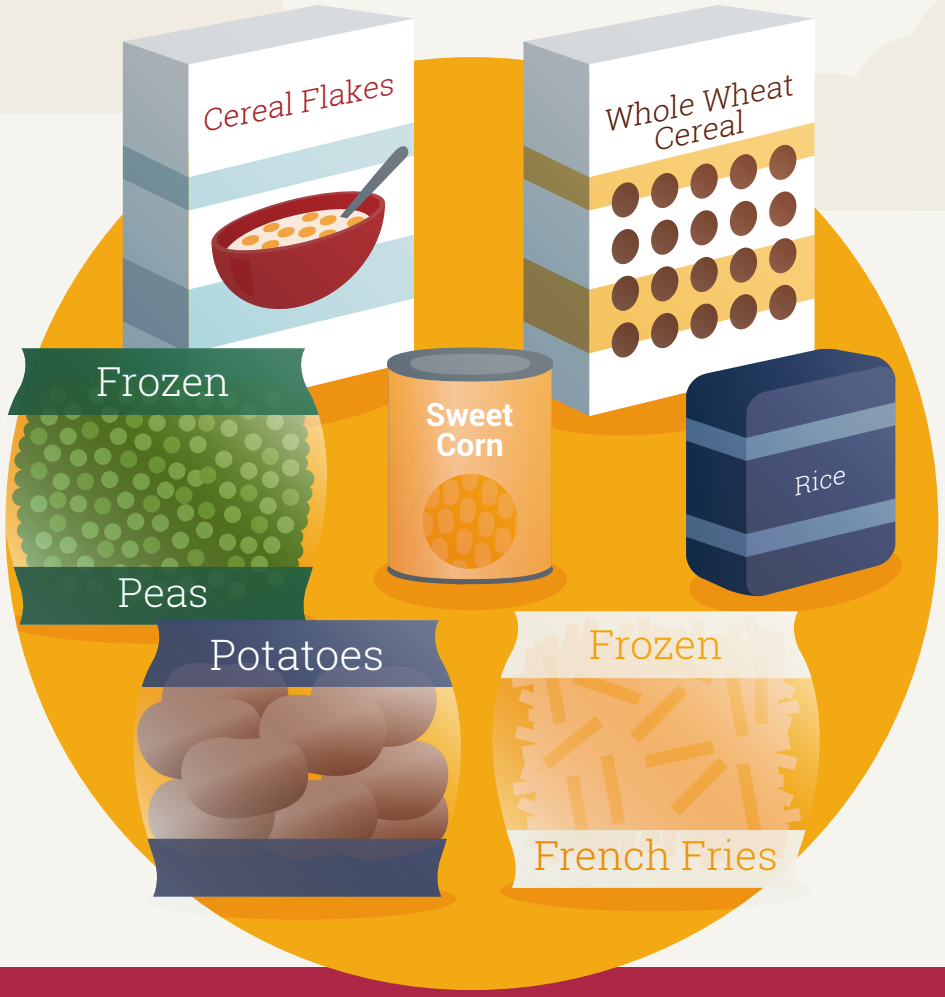
# RED STOP!

## Do not eat these foods

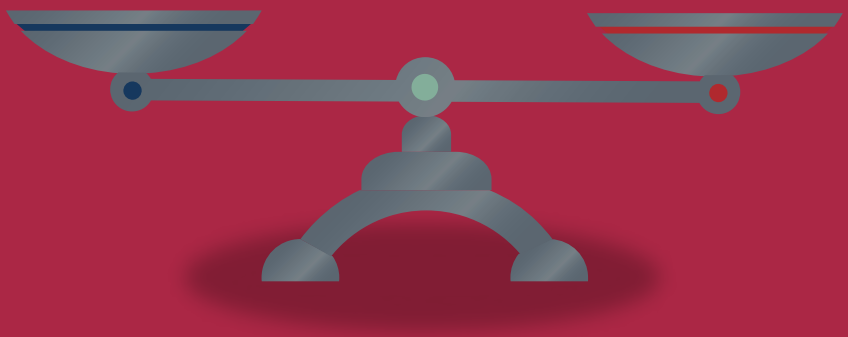

These foods are all rich in protein and therefore high in Tyrosine and Phe, so they are not allowed.

# AMBER Go Cautiously!

## These foods can be eaten in measured amounts and are known as exchanges




**1 Exchange** **1g Protein**



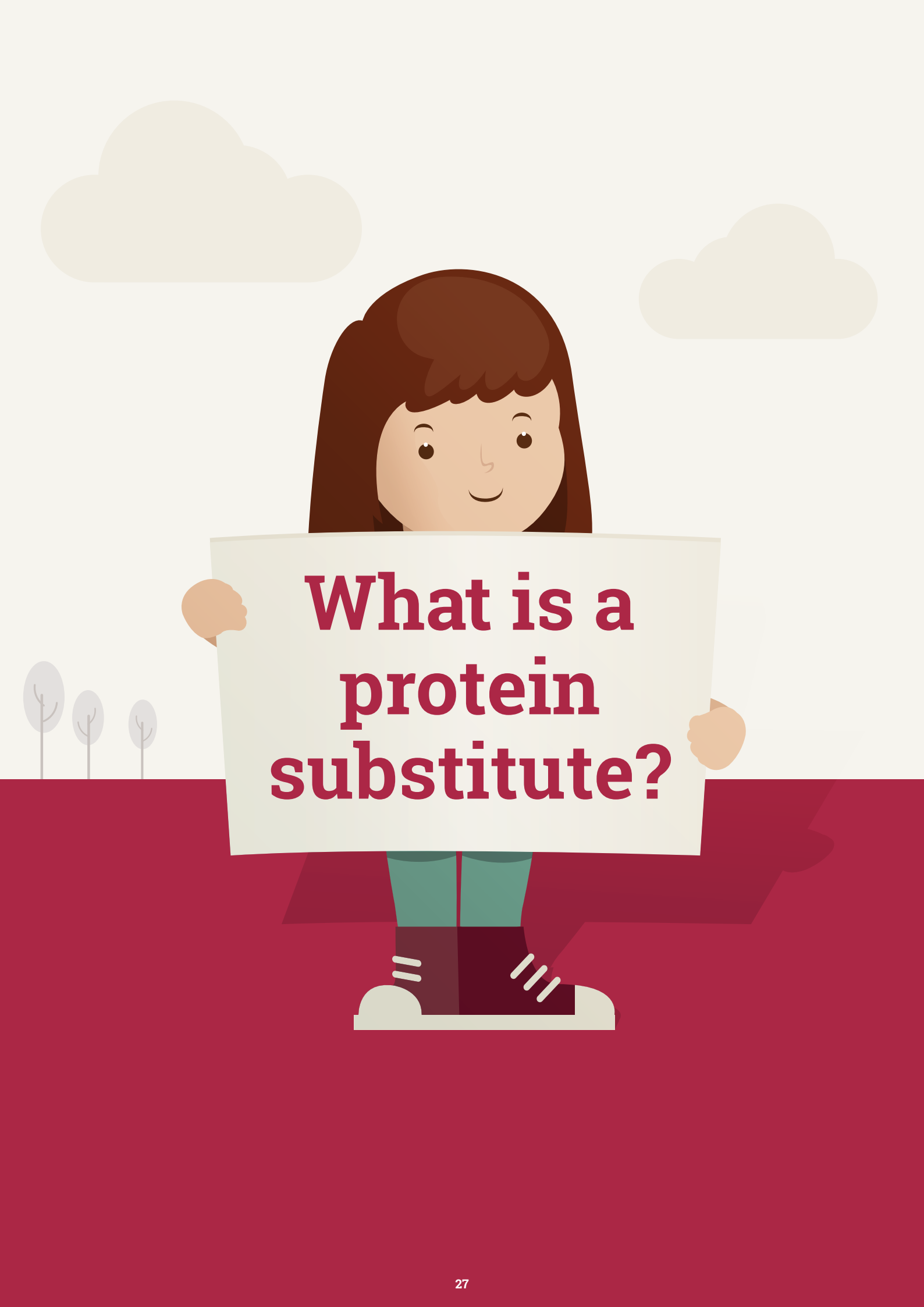
Other foods like these contain small amounts of protein. These foods can be eaten in measured amounts. The measure is called an exchange. These foods are spread out between the day's meals to provide small amounts of essential Tyrosine and Phe. The quantities allowed will vary from person to person and from time to time in the same person.

# GREEN Go!

## These foods can be eaten freely


These foods are either naturally low in Tyrosine and Phe or have been specially made to be low in protein and so are “FREE from exchanges” or do not have to be measured.

Please note certain low protein manufactured foods will need to be counted as part of exchanges. Please check with your dietitian.


What is a protein substitute?

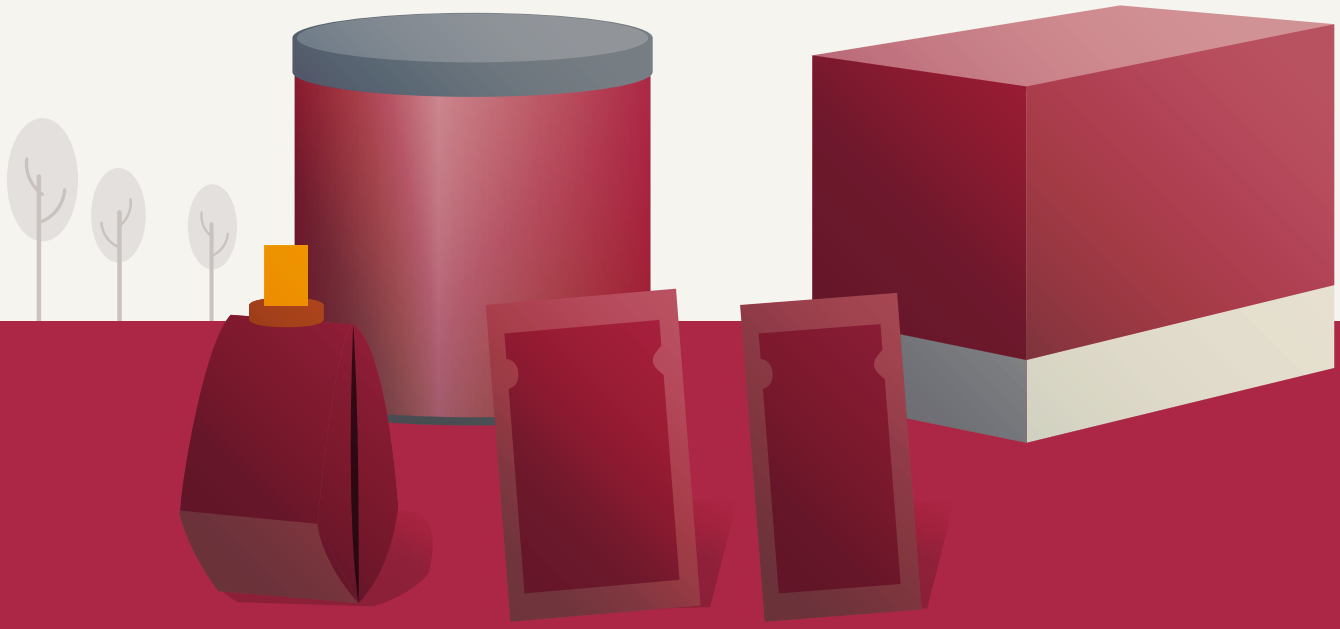
# Why does a person with TYR need to take a protein substitute?

- A protein substitute is a specially made medical supplement, which can come in various formats like liquids or powders that can be made up to various consistencies.
- A protein substitute typically contains NO Tyrosine or Phe, but it contains all of the other amino acids in protein that your child needs to grow. Most protein substitutes also contain vitamins, minerals and other important nutrients to ensure your child is getting what they need.
- The protein substitute is an extremely important part of the diet for a person with TYR and it must be taken regularly and evenly spread over the day. This helps to keep the Tyrosine and Phe levels steady throughout the day.

# Protein Substitutes

Many protein substitutes are designed to fit easily into everyday life.

Your dietitian will advise you on which protein substitute is best for your child at each stage of their development.




How will I feed my new born baby?

# Infants: breastfeeding


**Step 1:** Once your child is diagnosed, you will be seen by a dietitian.Your baby will be given a special Tyrosine and Phe-free formula until the levels of Tyrosine fall back to normal (usually within a few days).

**Step 2:** Once the levels are under control Tyrosine and Phe-free formula is given before a breastfeed to restrict the amount of breastmilk taken by the baby. Breastmilk provides limited but essential Tyrosine and Phe for normal growth.

Breastfeeding offers many benefits for baby and mother, it is recommended you discuss this with your health care professional.

# Infants: non-breastfeeding (bottle feeding)


**Step 1:** Once your child is diagnosed, you will be seen by a dietitian. Your baby will be given a special Tyrosine and Phe-free formula until the levels of Tyrosine fall back to normal (usually within a few days).

**Step 2:** Once the levels of Tyrosine are under control a small restricted amount of standard baby formula will be given along with the Tyrosine and Phe-free formula to provide the limited Tyrosine and Phe that are essential for normal growth.


Weaning

# When to start weaning*

Your dietitian will advise you when to start solid foods*, but it will be at the same time as babies without TYR. These will initially be protein-free foods or foods that have a very low protein (low Tyrosine and Phe) content.

This will help your child to develop a healthy eating pattern while eating a variety of fresh foods.

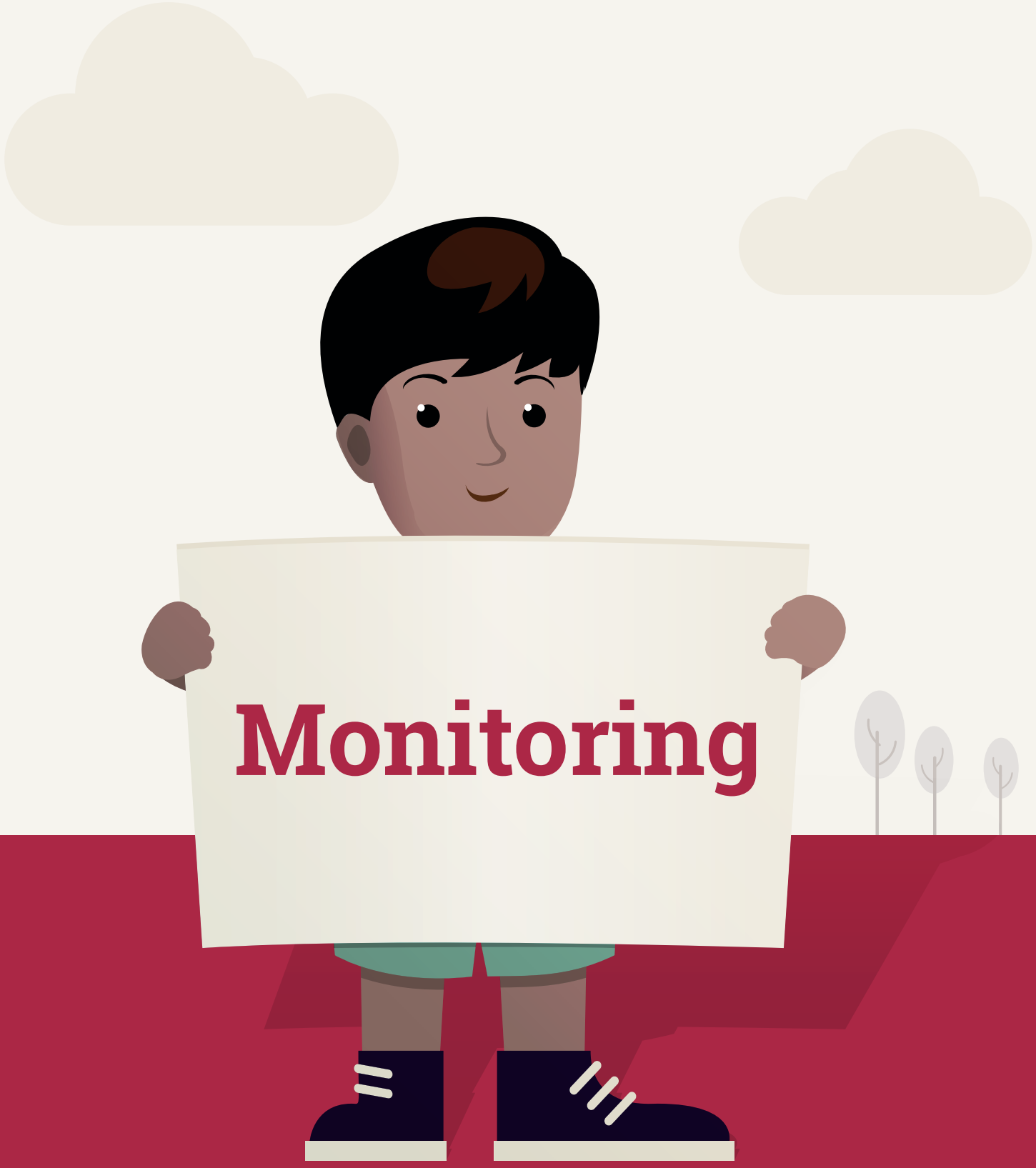
This is also the time that a second stage protein substitute can be introduced, your dietitian will advise you about this this*.

* Weaning may occur a little earlier than 6 months, depending on your baby, but it should never be started before 17 weeks.

# 7 to 12 months

Your child will then be encouraged to move to more textured food and finger foods. Over time, they will get more of their protein exchanges from food (e.g. potato or permitted cereal). Less exchanges will come from breast milk or formula milk.

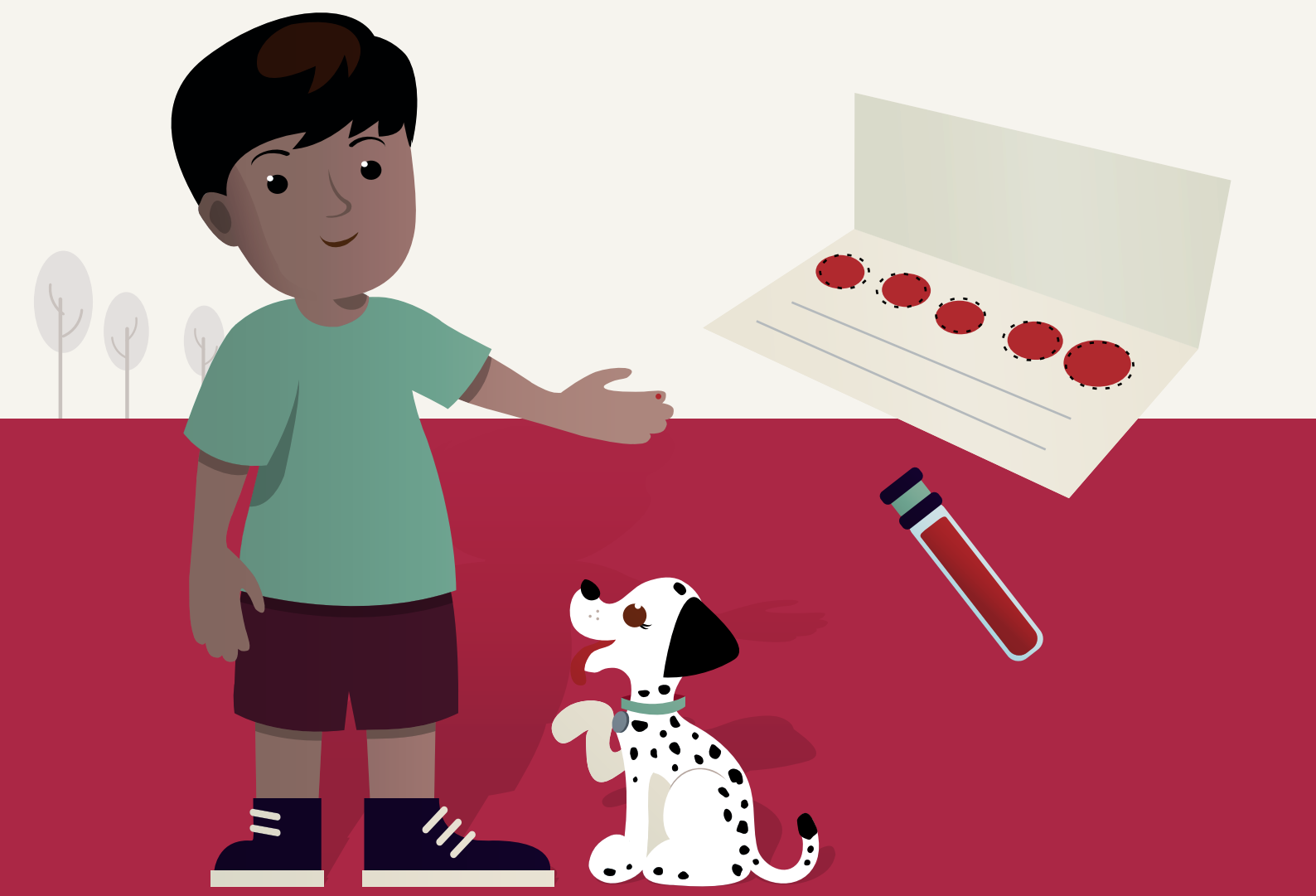
Protein free foods should still be included in the diet as well. Your child will also continue their gradual switch from their Tyrosine and Phe-free formula to second stage protein substitute.


Monitoring

Your child will have regular blood tests and monitoring to make sure their Tyrosine and Phe levels are under control.

This is very important to ensure that they do not have too little or too much Tyrosine and Phe in their diet.

Additional Phe supplementation may be required.



Your dietitian will contact you once the results are processed and discuss any changes that may be needed to your child's diet.


As your child grows up

# Children / Teenagers / Adults:

Your child will continue to follow a low protein diet as they grow up into adulthood.

You, your child and dietitian will help choose the protein substitute that best suits them.