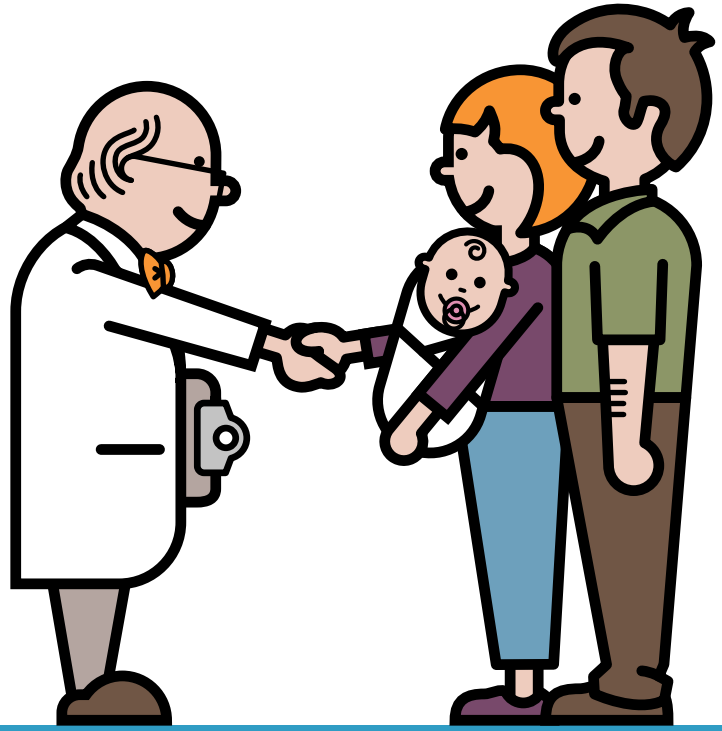
Isovaleric Acidemia

IVA

Information for families following a positive newborn screening



Adapted by the Dietitians Group BIMDG

BIMDG
British Inherited Metabolic Diseases Group

BASED ON THE ORIGINAL TEMPLE WRITTEN BY BURGARD AND WENDEL

Reviewed & revised for North America by: A. Huber

This version of the TEMPLE tool, originally adapted by the Dietitians group of the BIMDG for use within the UK and Ireland, has been further adapted by Nutricia North America for use within United States and Canada. This version no longer represents clinical or dietetic practice in the UK or Ireland.

TEMPLE
Tools Enabling Metabolic Parents LEarning

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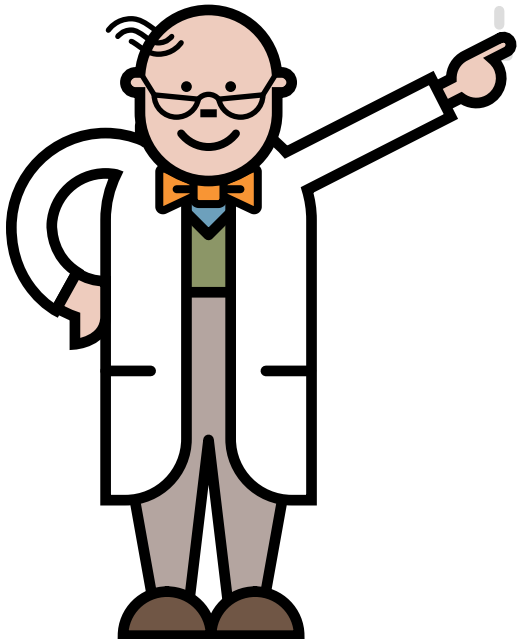
What is IVA?

IVA stands for isovaleric acidemia.

It is pronounced iso-val-air-ic acid-ee-me-a.

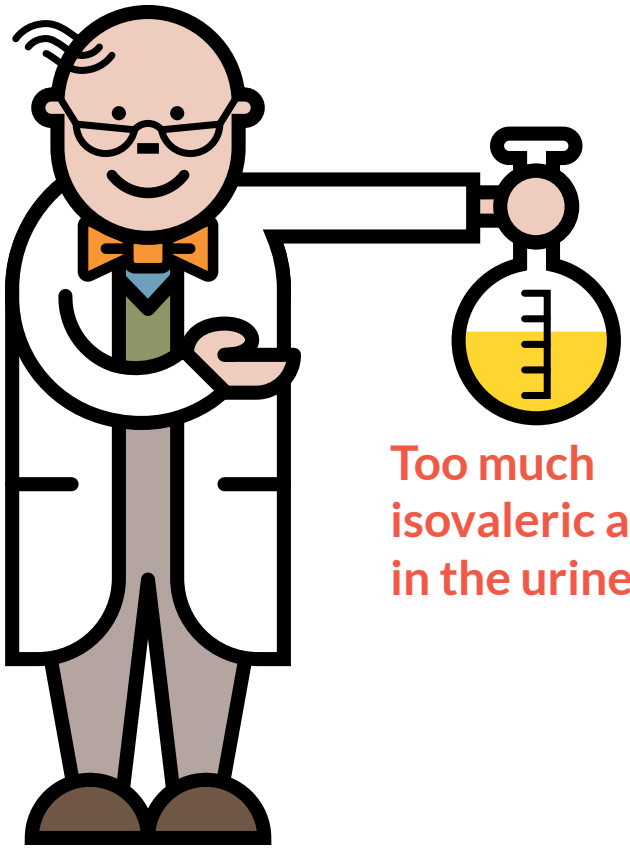
It is an inherited metabolic condition.



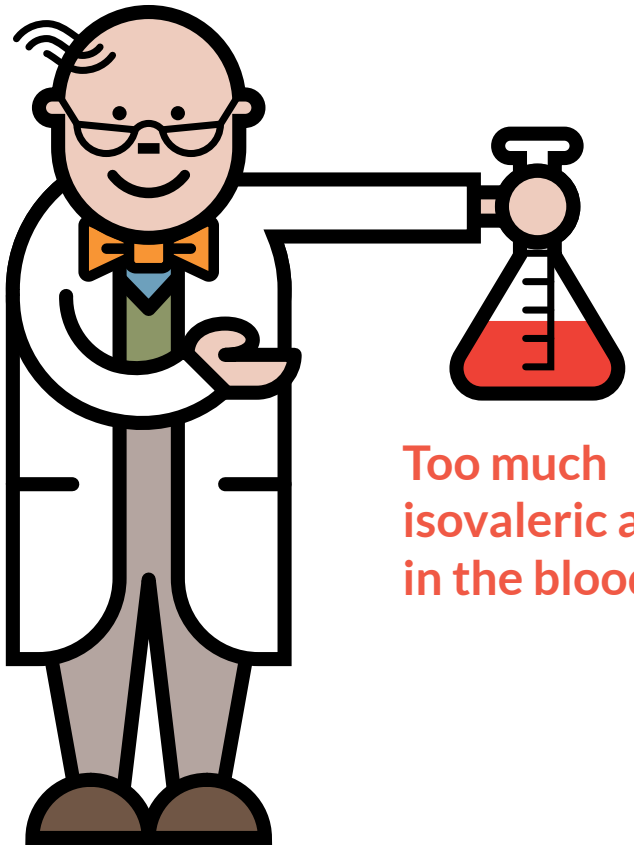
Isovaleric Acidemia

IVA

What is IVA?



Too much isovaleric acid in the urine

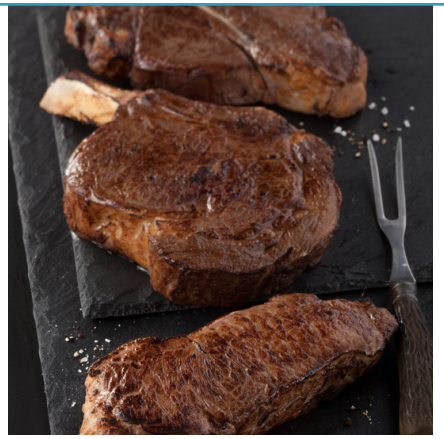


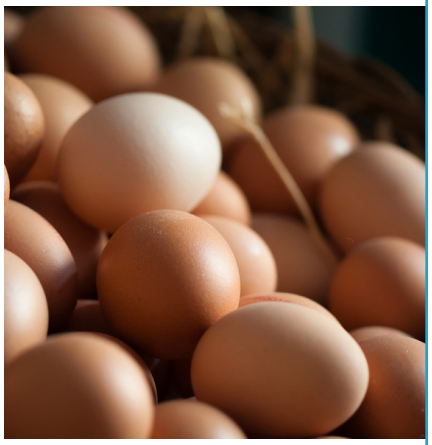
Too much isovaleric acid in the blood

How does IVA affect the body?

IVA affects the way the body breaks down protein.

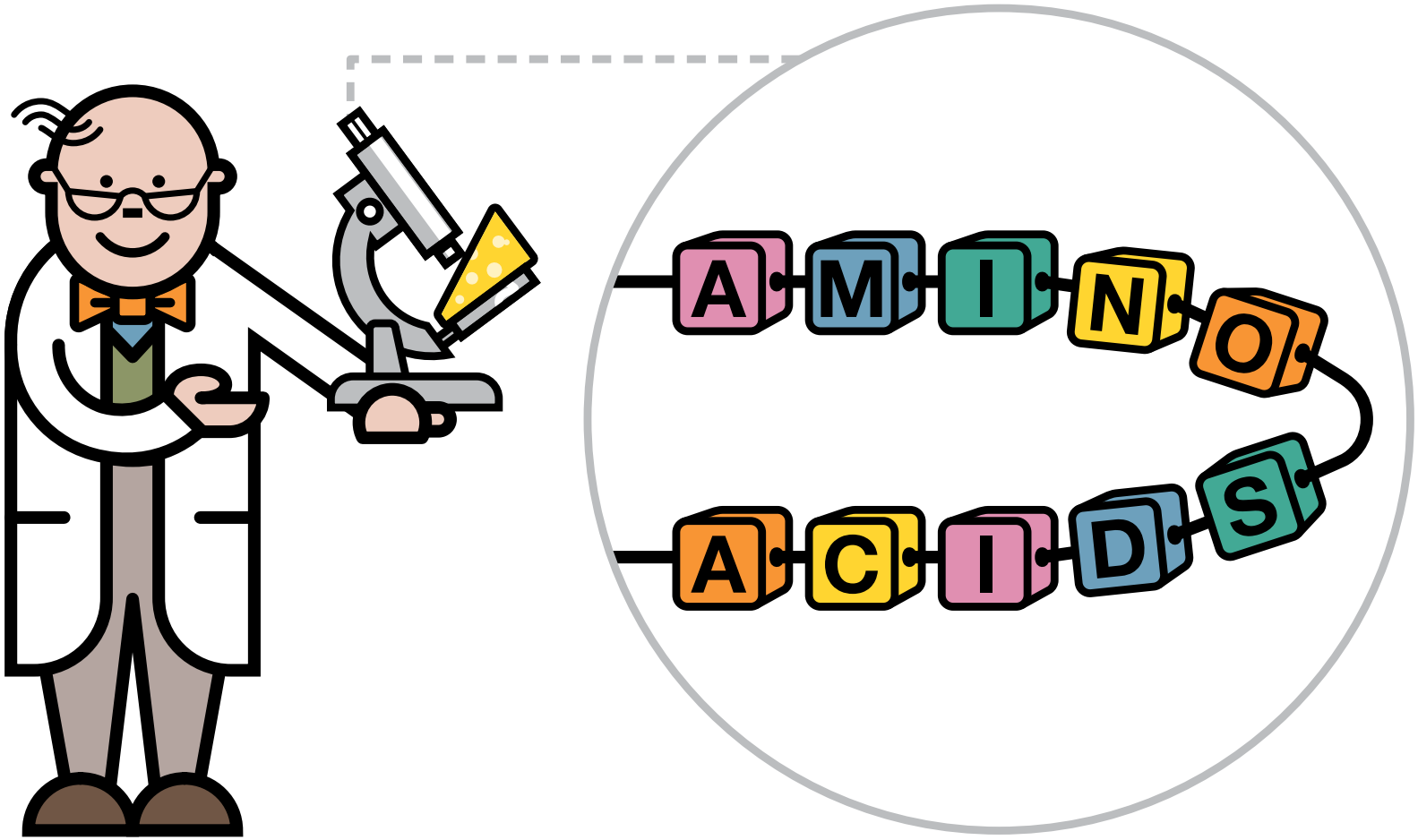
Protein is found in our bodies and in many foods. The body needs protein for growth and repair.



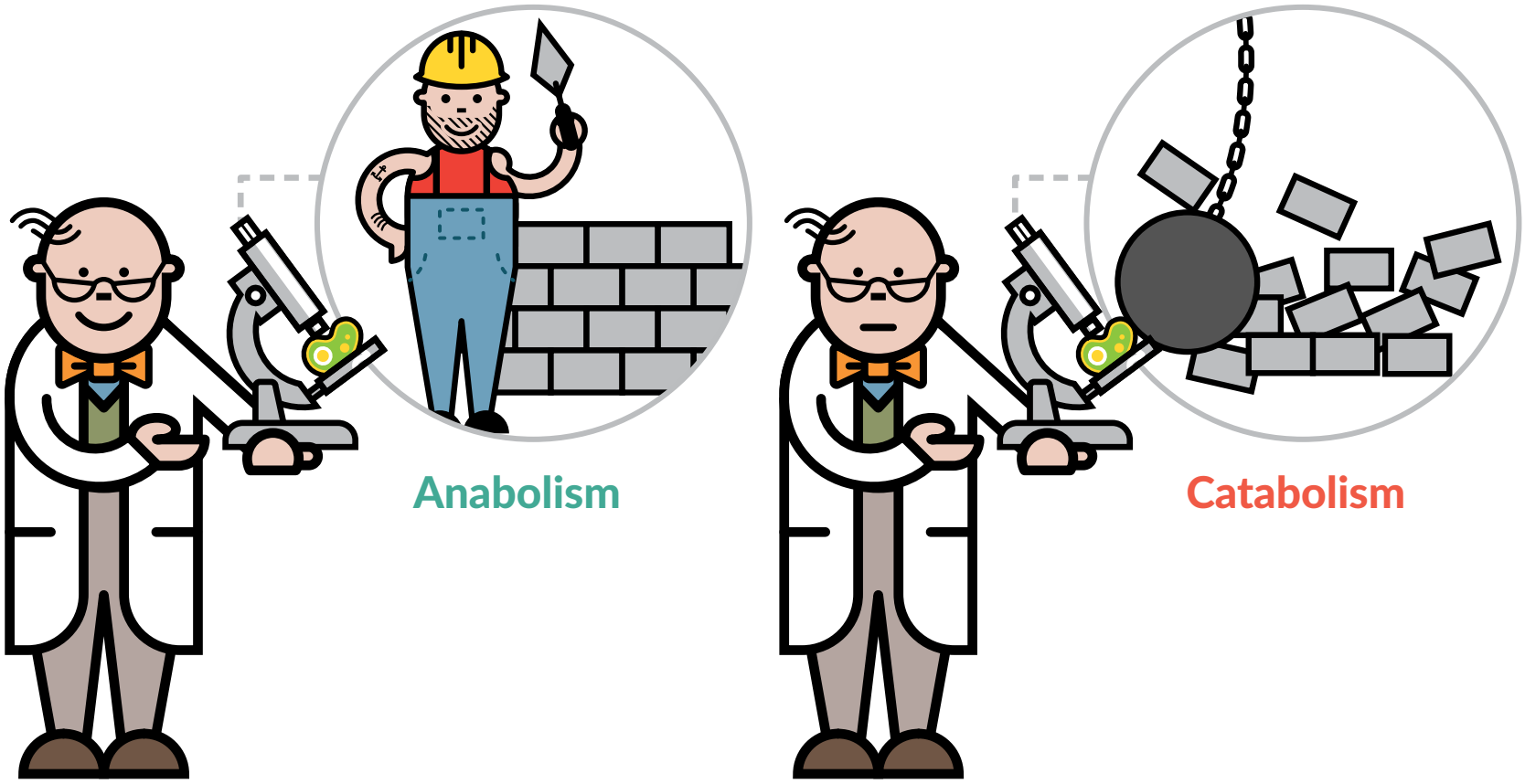
What is protein?

Protein consists of chains of many smaller units called amino acids.



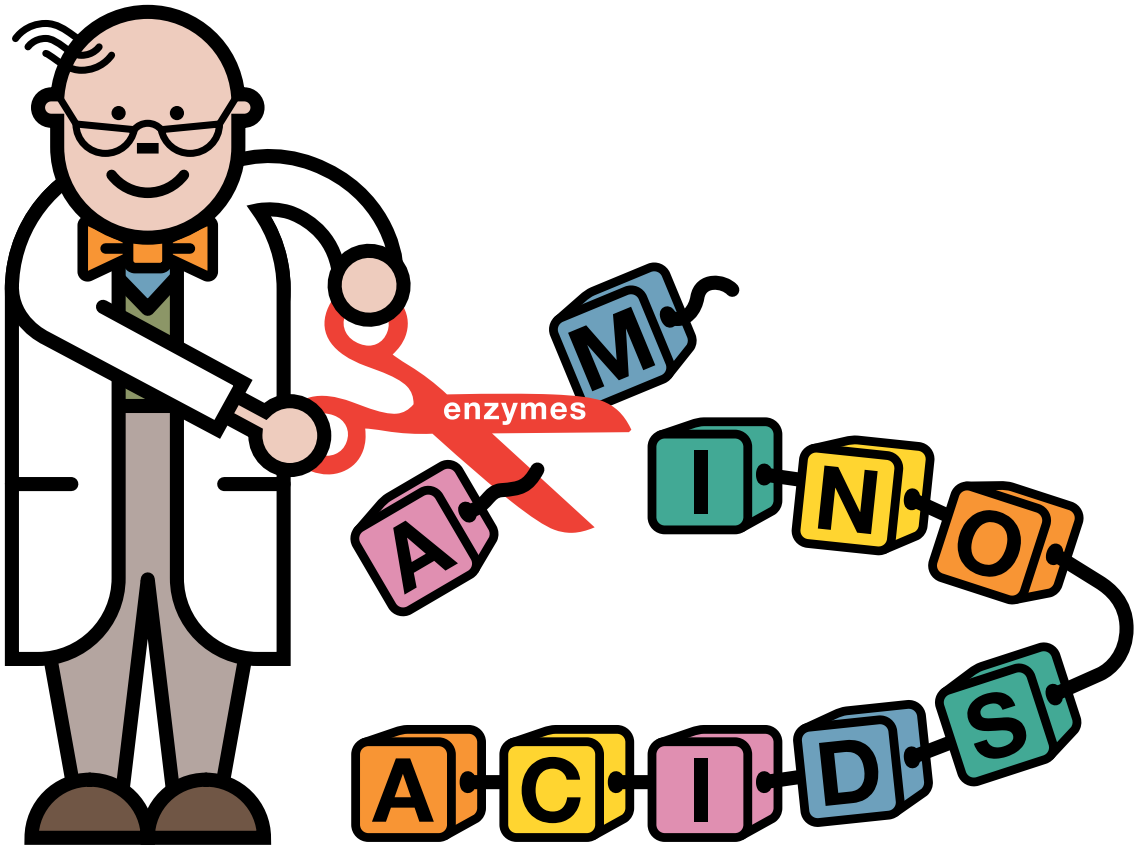
Protein metabolism

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



What do enzymes do?

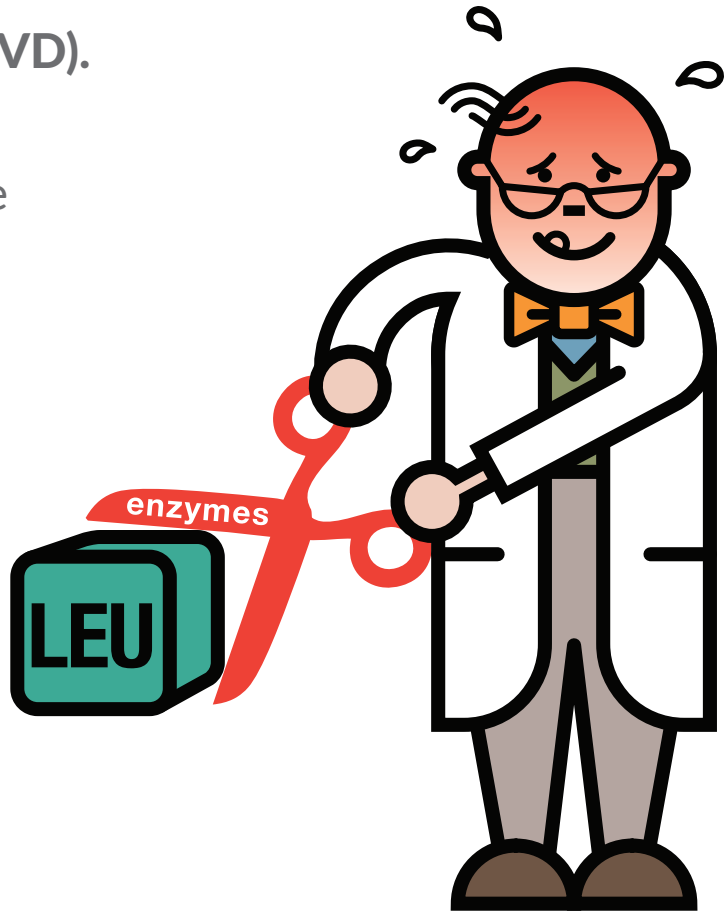
Enzymes help with metabolism by functioning like scissors.
They break down proteins into smaller parts, including amino acids.



What happens in IVA?

IVA is due to a deficiency of an enzyme called **isovaleryl-CoA dehydrogenase (IVD).**

This results in the body being unable to break down an amino acid called leucine (LEU). This leads to a harmful buildup of metabolites, specifically isovaleric acid.



What can go wrong in IVA?

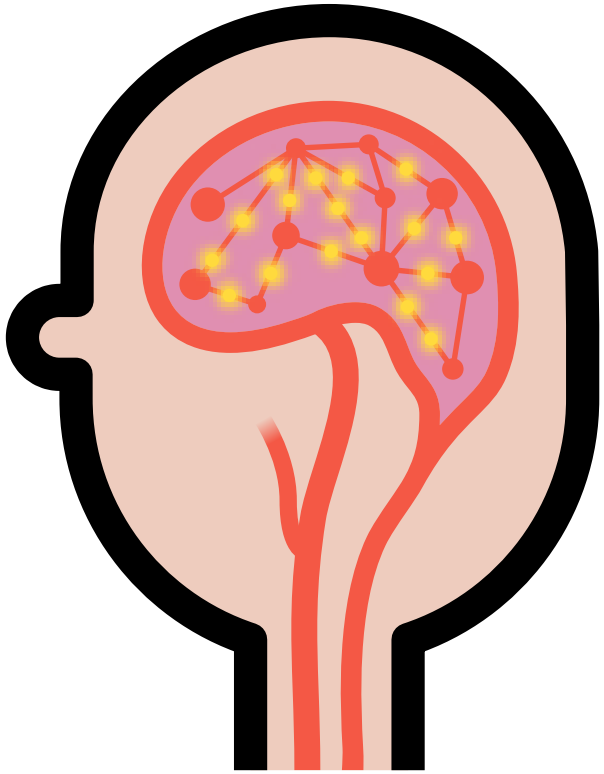
The buildup of isovaleric acid is harmful, particularly to the brain.

Before management, early symptoms include poor feeding, irritability, sleepiness, vomiting, rapid breathing and coldness.

Left unmanaged, some babies may have a metabolic crisis, seizures and become unconscious.

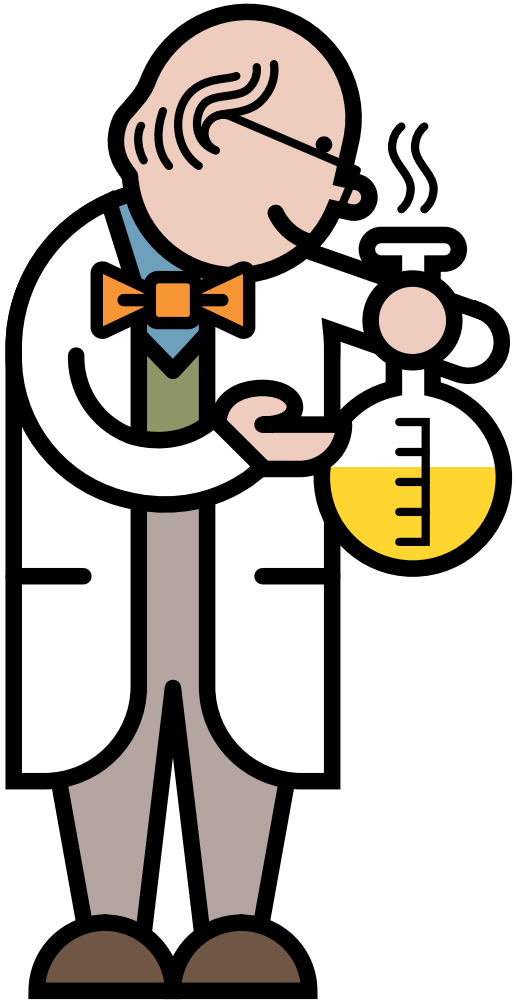
Some children may not develop early symptoms but present when they are a little older with learning difficulties.

Early management can prevent brain damage and learning difficulties.



What about other symptoms?

Isovaleric acid smells like sweaty feet. Therefore, people with IVA can smell of sweaty feet before they start management.



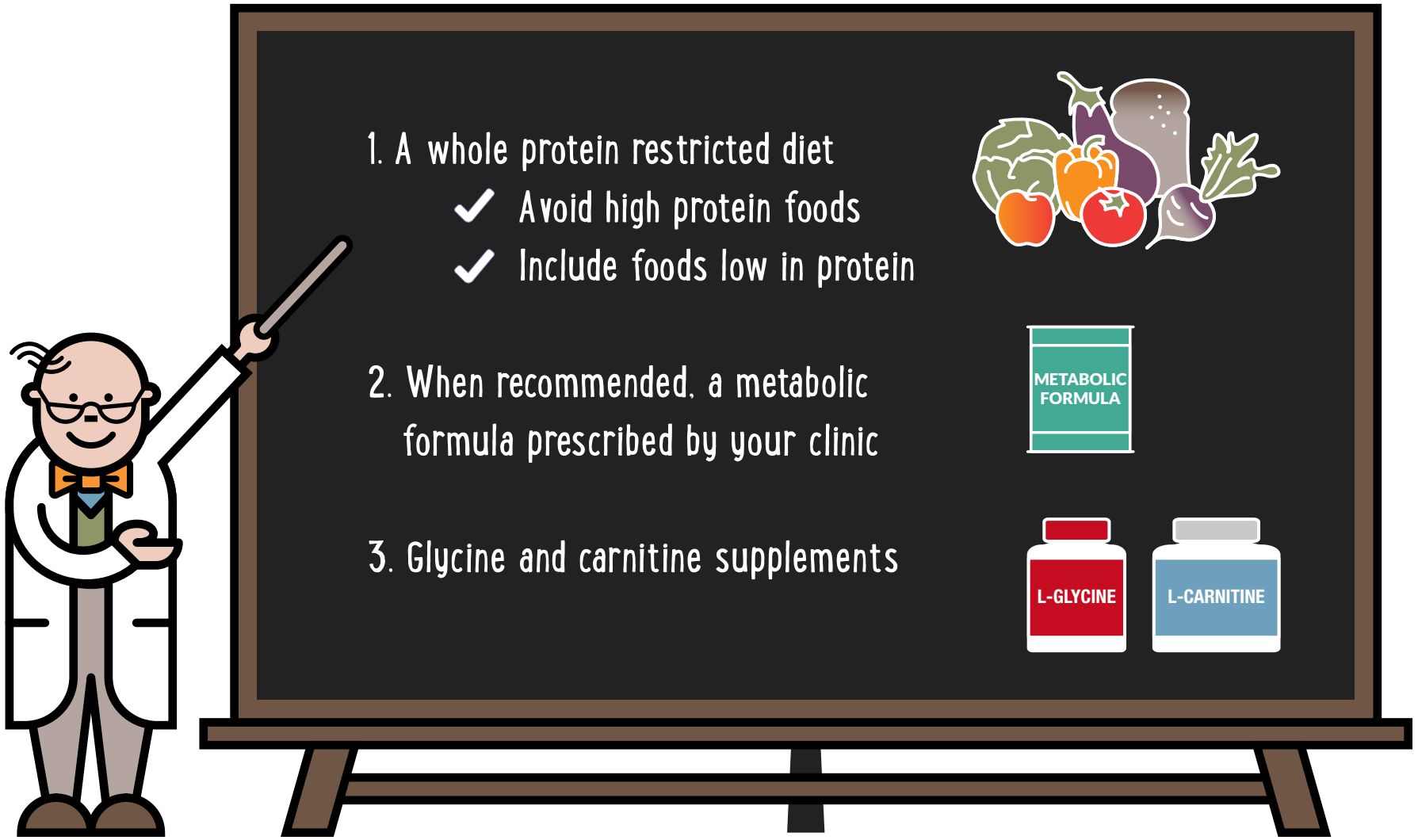
How is IVA diagnosed?

As part of newborn screening, a few drops of blood are collected.

The blood sample is then analyzed.

High levels of isovalerylglycine, 3-hydroxyisovaleric acid and isovalerylcarnitine could mean your child has IVA which will prompt your clinician to do further testing to confirm the diagnosis.

How is IVA managed day-to-day?



Avoid high protein foods

Foods rich in protein are also rich in leucine, and therefore, should be avoided. This includes **meat, fish, eggs, cheese, milk, bread, pasta, nuts, soy and tofu**.

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Dietary management of the condition should only be done under medical supervision.

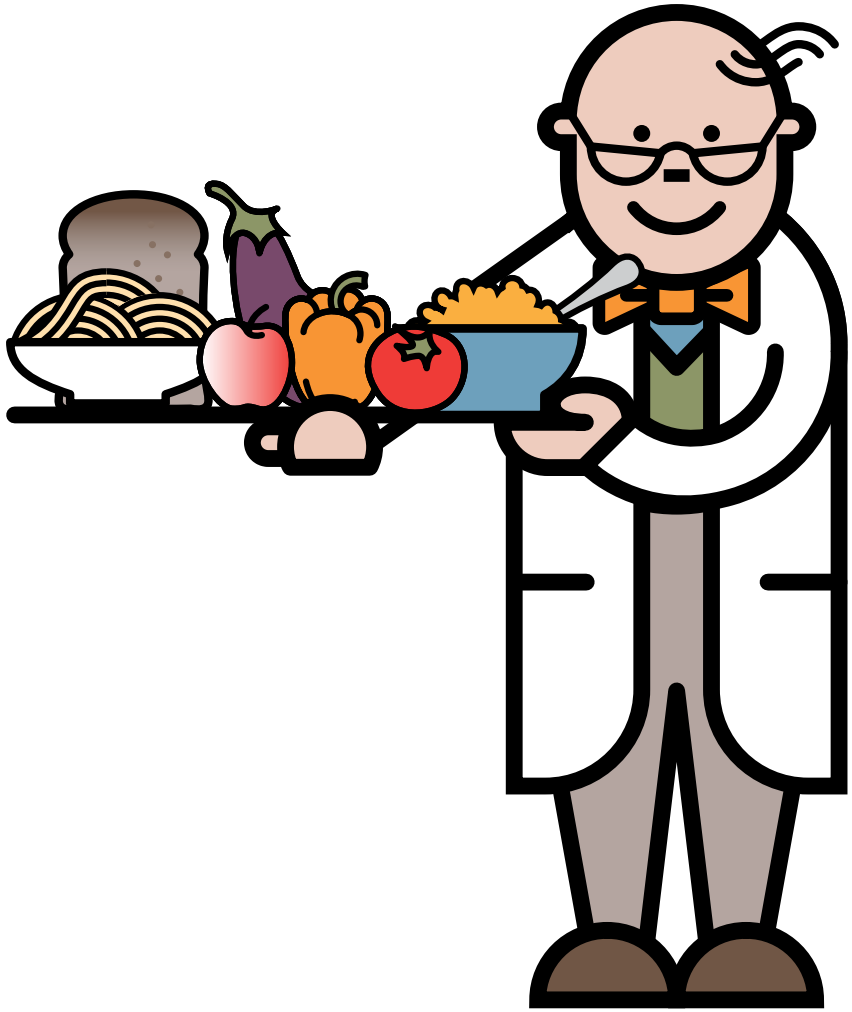
Include foods low in protein

These are foods that contain small amounts of leucine which can be used in typical quantities.

They include many fruits and vegetables, and specially formulated low protein foods

They provide:

- An important source of energy
- Variety in the diet



Low protein cooking

Cooking low-protein meals for your child can still be appealing to the eye and taste good.

There are many low-protein cookbooks to choose from. Your dietitian may be able to recommend a few favorites.

Feeding your baby with metabolic formula

Leucine is essential for normal development and therefore a limited and controlled amount must be taken daily.

Breast milk or standard infant formula will provide the leucine required by your baby prior to the introduction of solids, generally around 4-6 months of age.

Your baby will also need a special metabolic formula to provide protein without leucine.

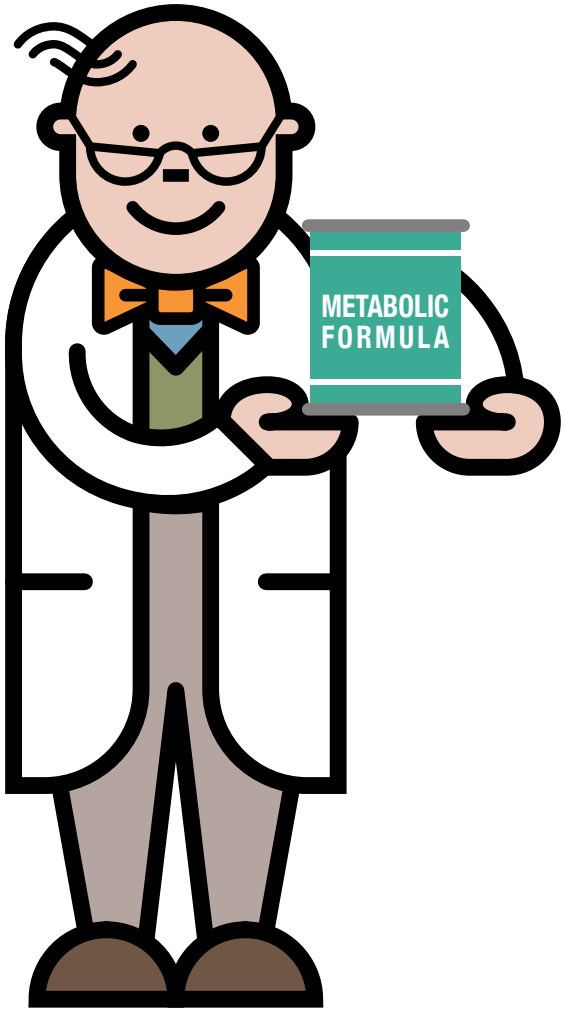
Your dietitian will determine how much breast milk or standard infant formula and metabolic formula to offer.

Leucine-free metabolic formula

Leucine-free metabolic formula is an essential part of meeting your baby's nutritional requirements.

Like breast milk or standard infant formula, metabolic formula has carbohydrate, fat, vitamins, minerals and protein in the form of amino acids without leucine.

Metabolic formula, plus the prescribed amount of leucine, allows your baby to get all the nutrients he or she needs to grow.



Tracking leucine

As your baby starts to eat solids your clinic will work with you to track leucine.

Foods must be weighed or measured using household measures (1 cup, 1 tablespoon, etc.) to determine leucine content.

Your clinic can help you find the best tools to help determine the leucine content of foods.

Metabolic crisis

A '**metabolic crisis**' causes a buildup of isovaleric acid, isovalerylglycine, 3-hydroxyisovaleric acid and isovalerylcarnitine in the urine and blood.

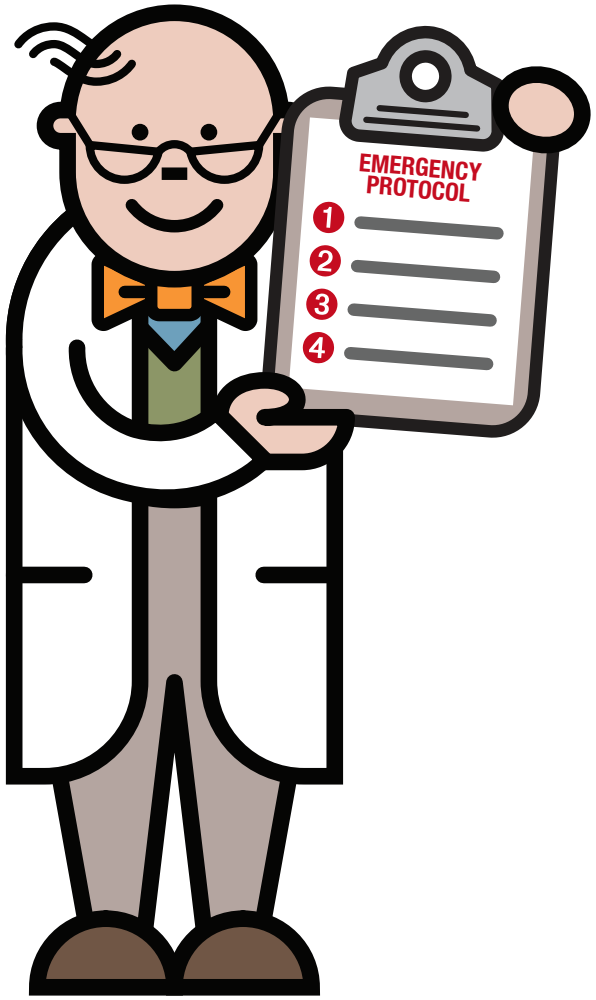
It is usually triggered by childhood infections or viruses causing high temperatures, vomiting, and diarrhea.

It is important to manage a metabolic crisis quickly and properly.

How is IVA managed during illness?

During any illness, our bodies need extra energy. The body will start breaking down cell protein, causing blood leucine levels to increase. This process is also referred to as catabolism. This will lead to a rapid buildup of the harmful metabolite, isovaleric acid, thus causing a metabolic crisis.

It is extremely important to start the emergency protocol your metabolic team has developed for you and contact them right away.



How is IVA managed during illness?

Always follow your medical team's guidance.

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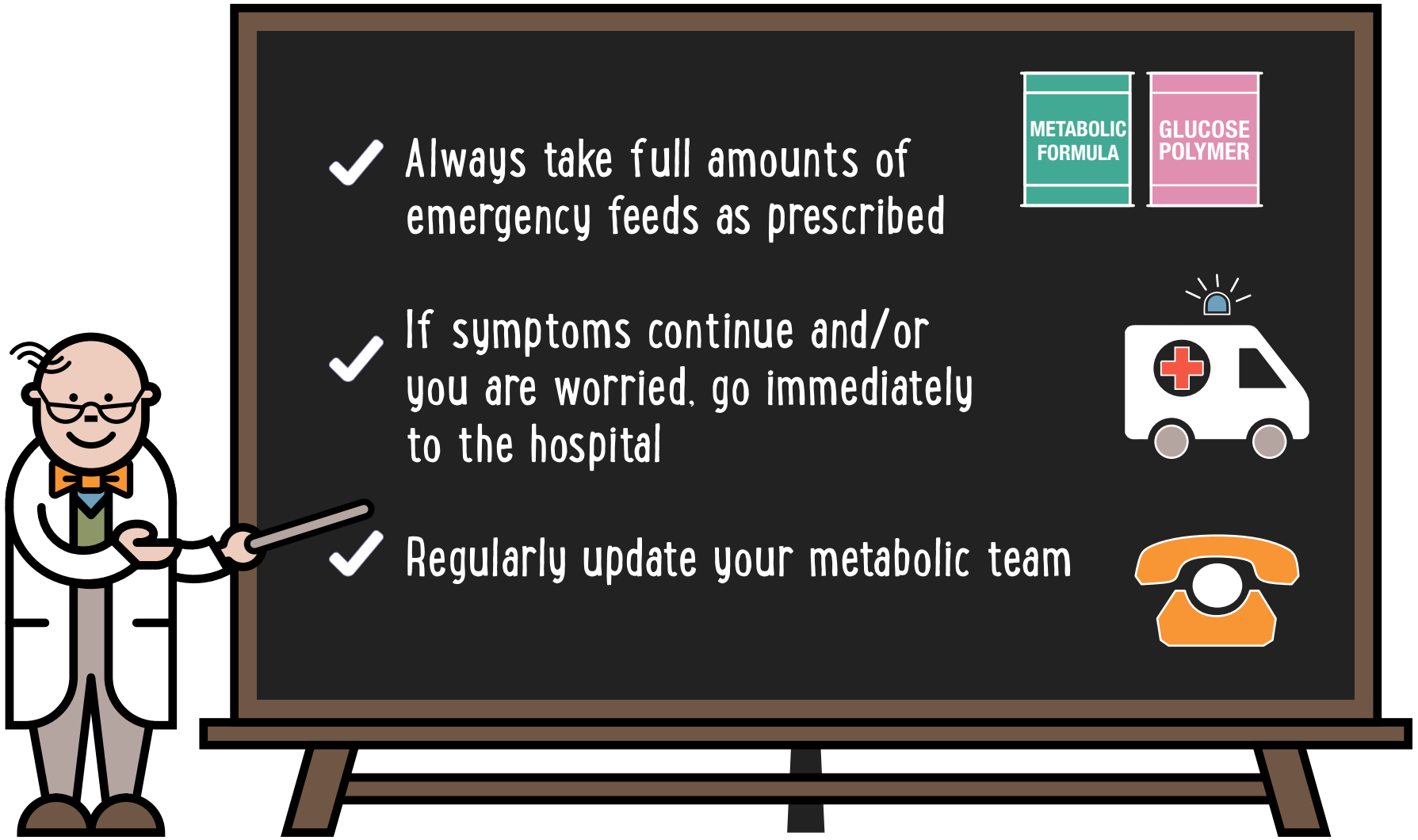
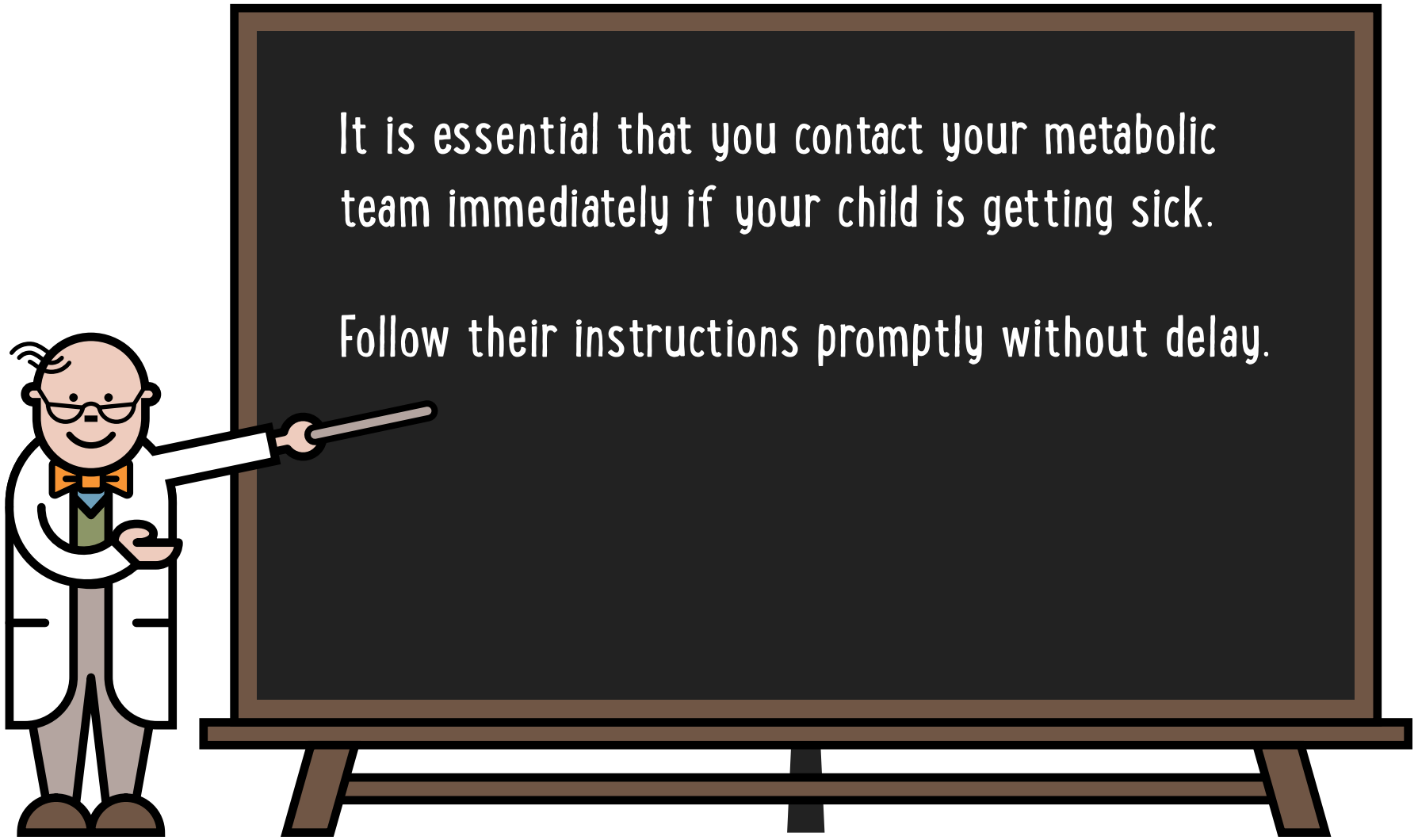
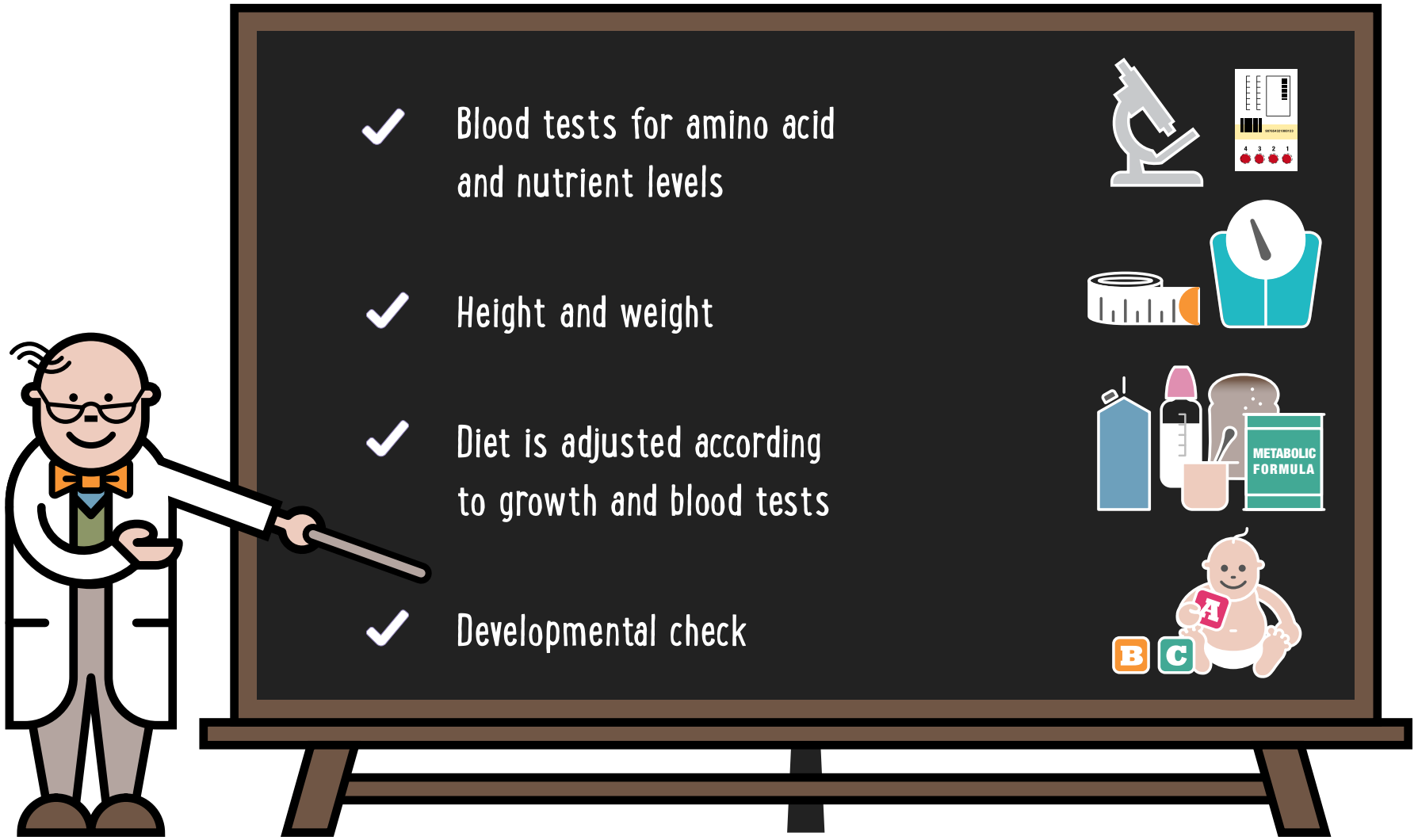


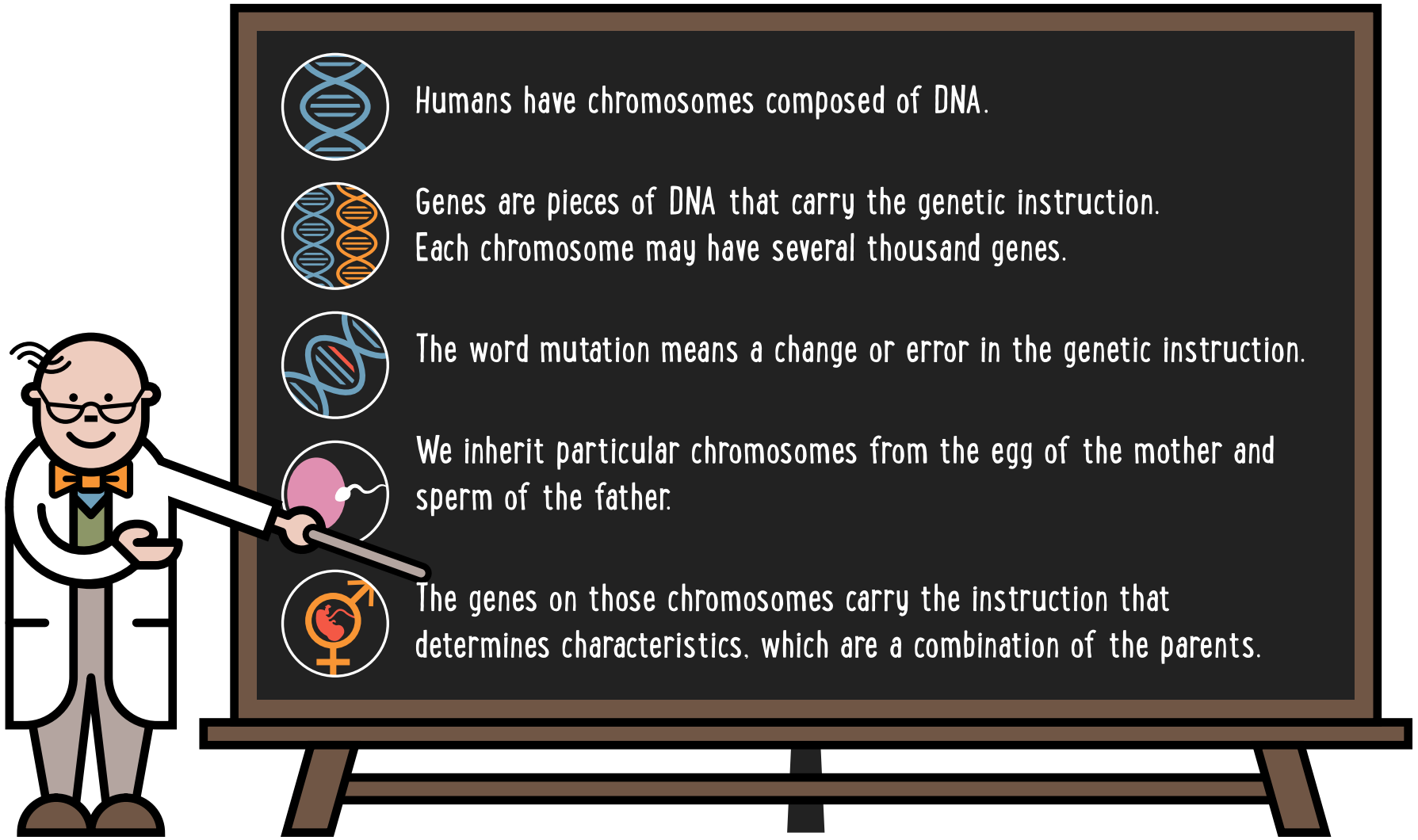
Most importantly



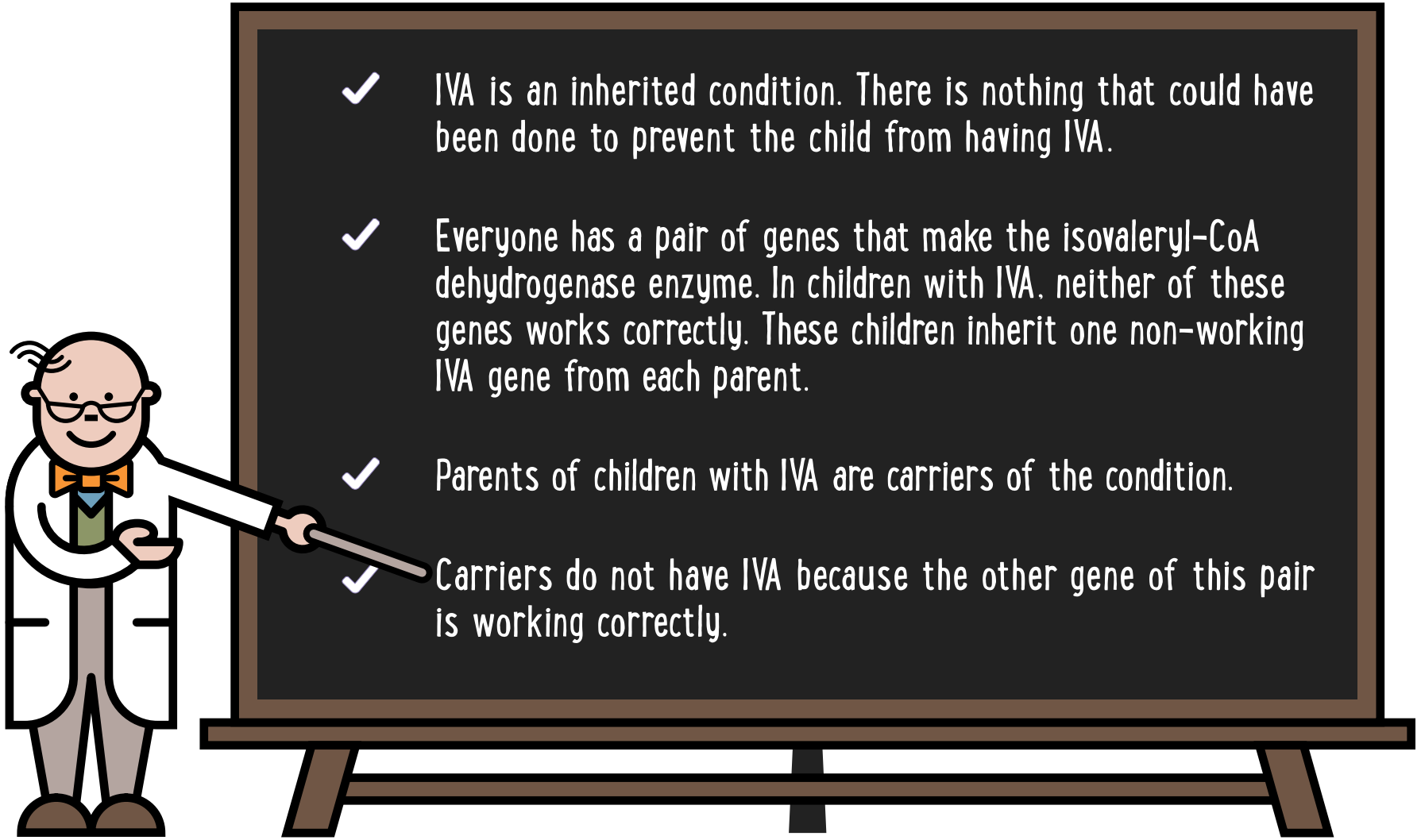
What else is monitored in IVA?



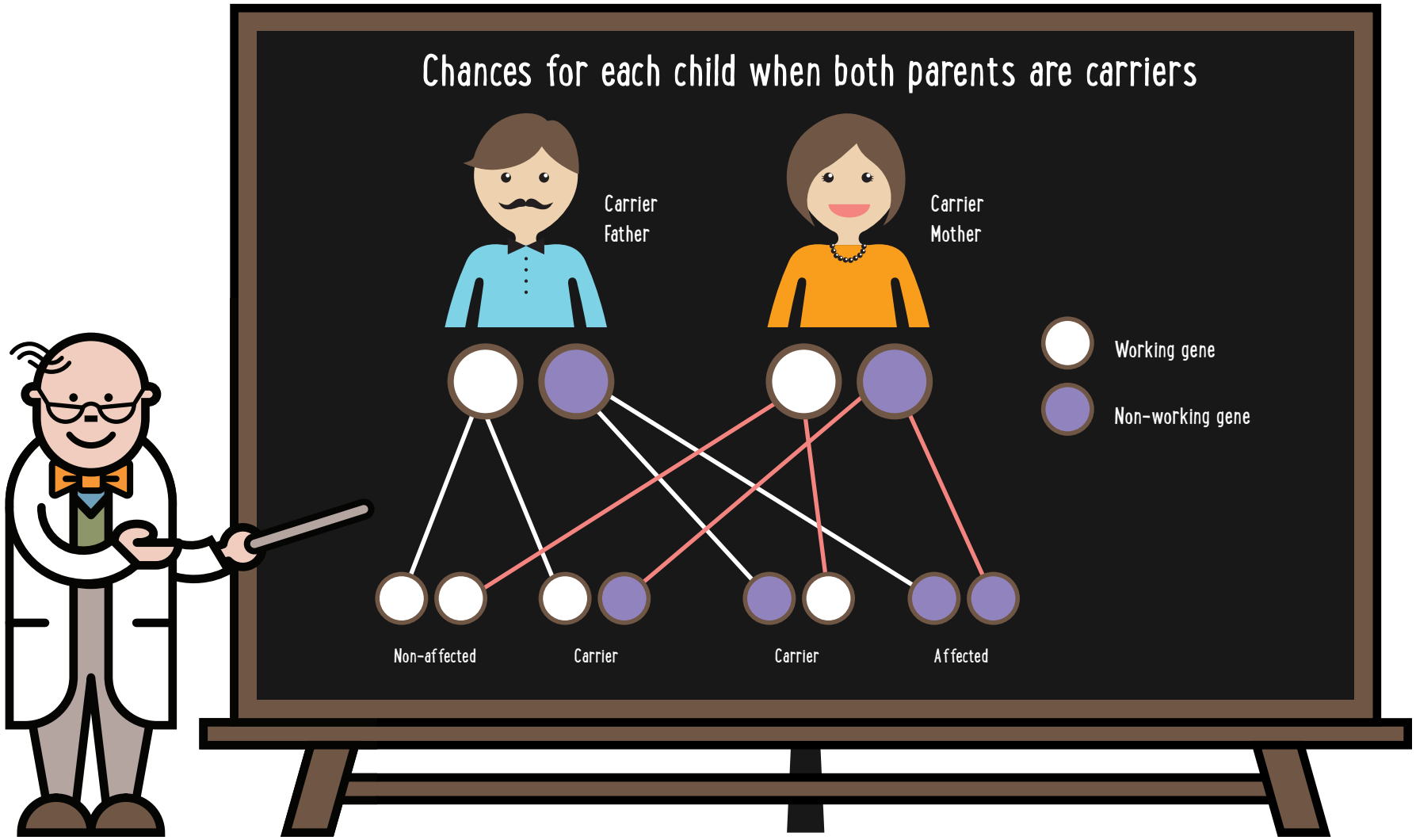
What happens in human genetics?



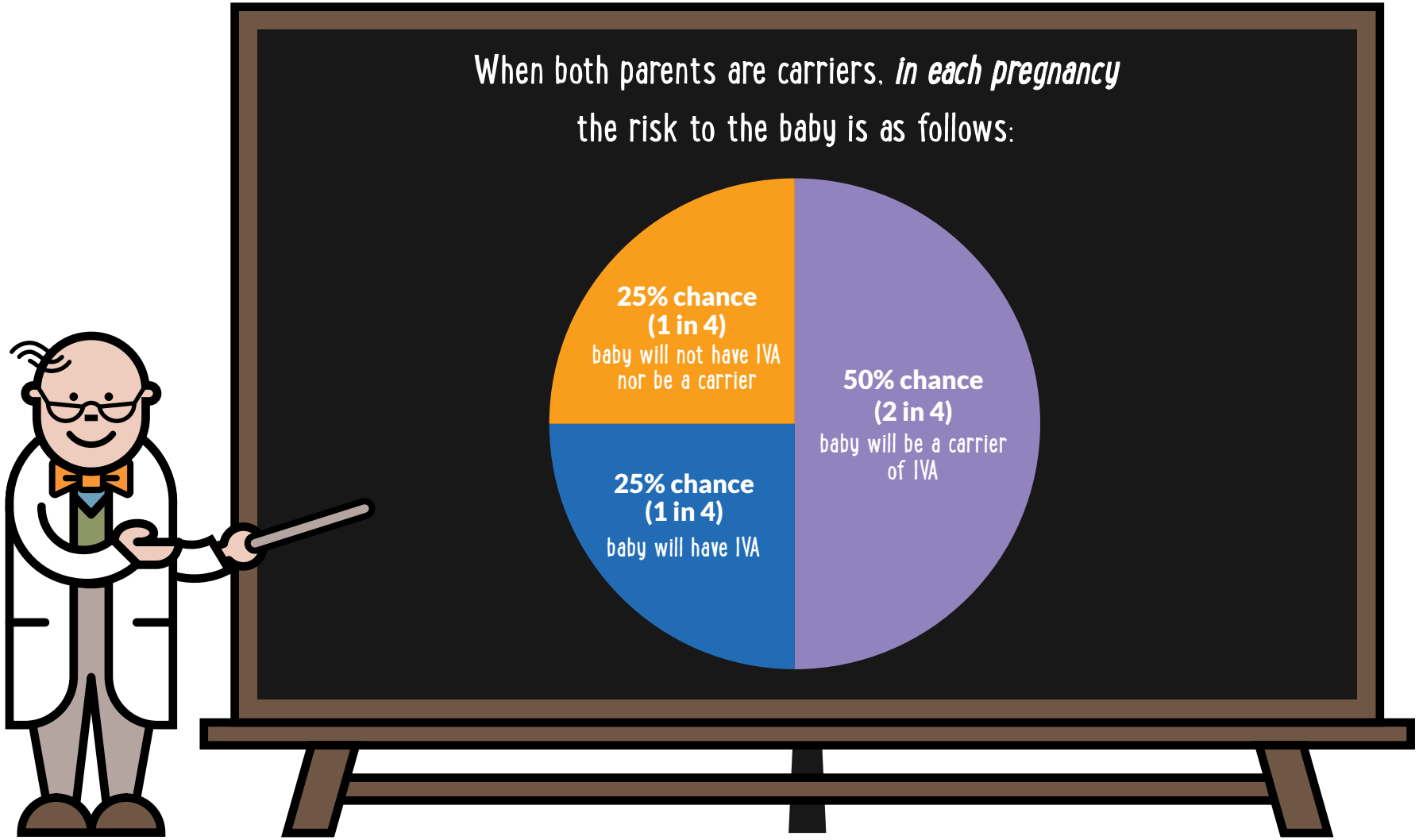
How does one inherit IVA?



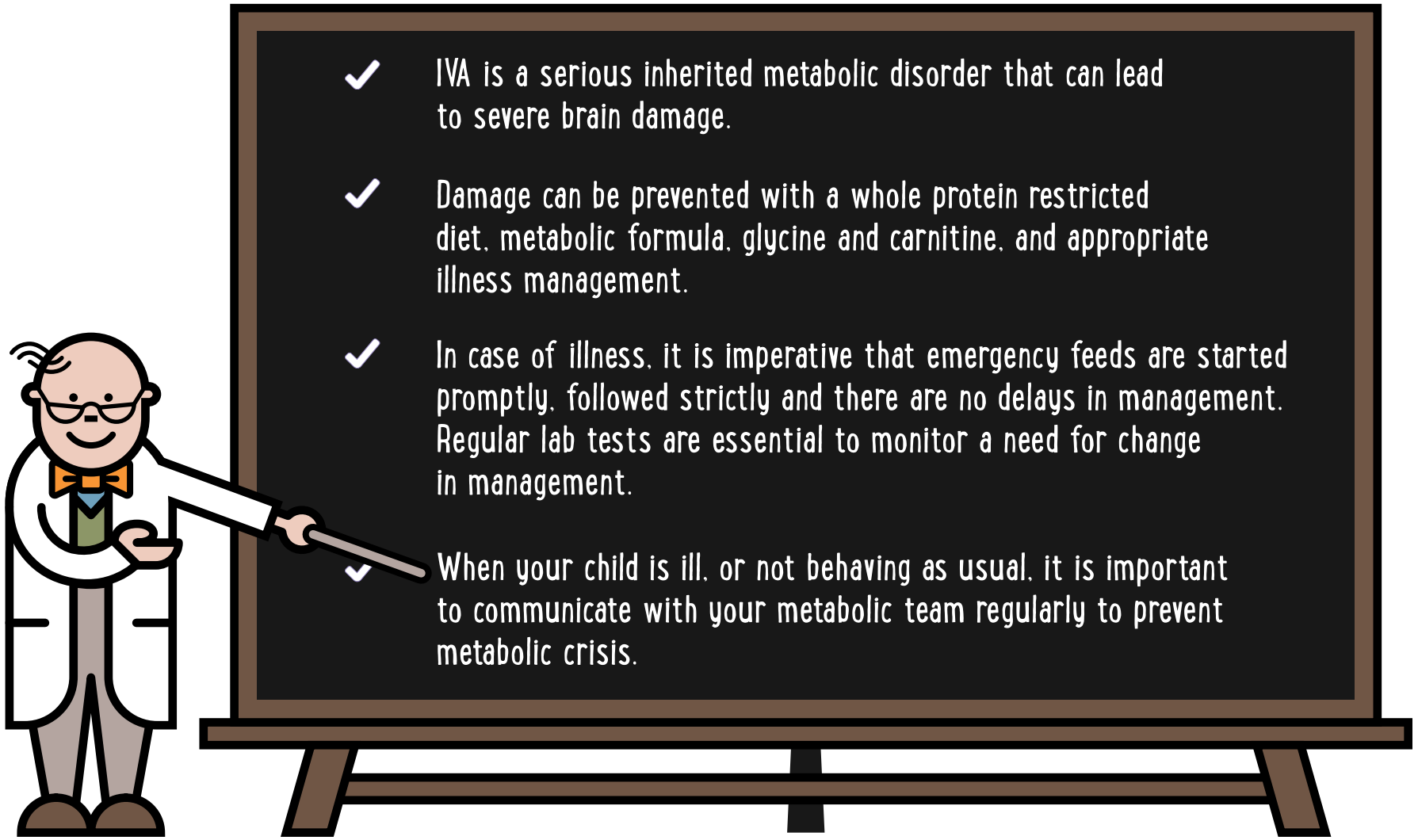
Inheritance – Autosomal recessive – possible combinations



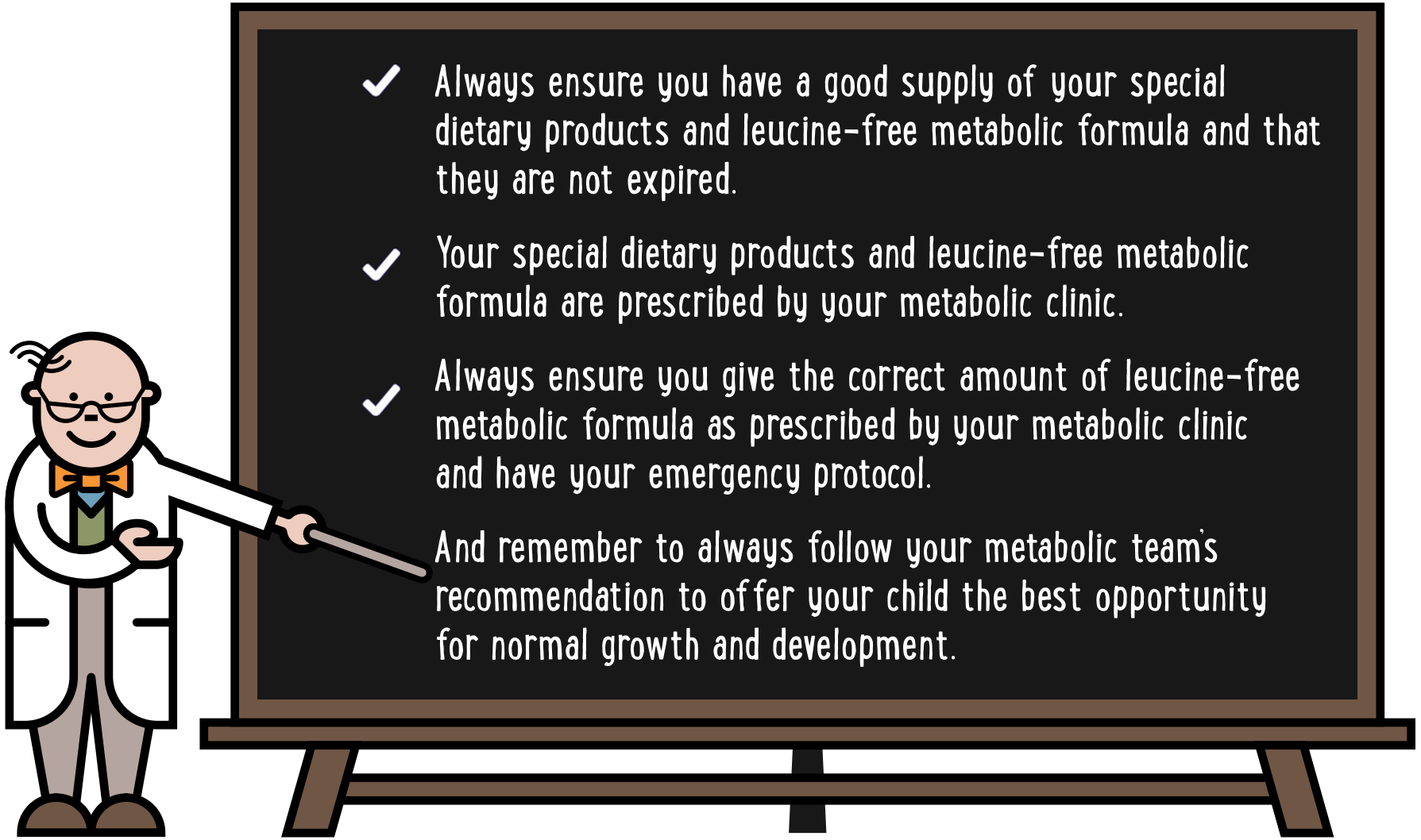
Future pregnancies



Take home messages



Helpful hints



Who's who (contact details)

My dietitian

Name: .....

Phone #: .....

Email: .....

My nurse

Name: .....

Phone #: .....

Email: .....

My doctor

Name: .....

Phone #: .....

Email: .....

Notes

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