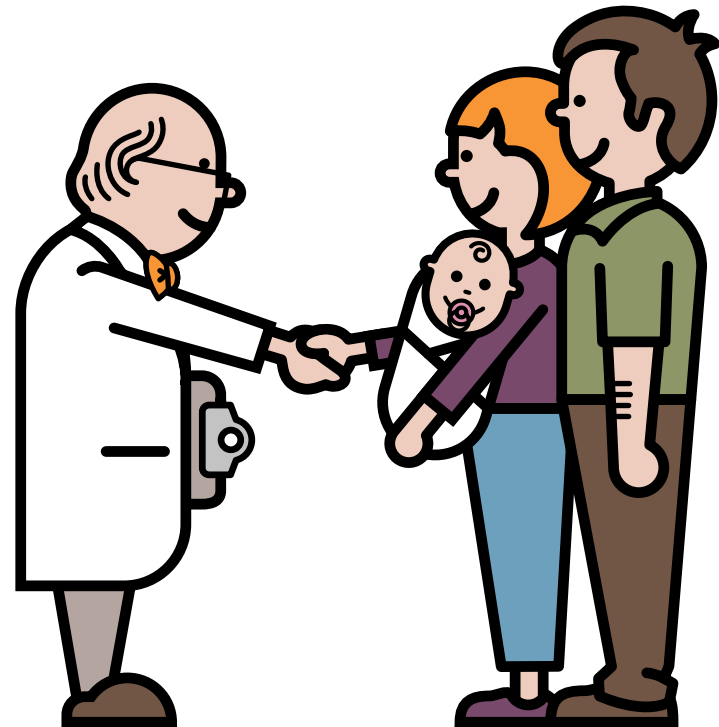


Glutaric Aciduria Type 1

GA-1

Information for families after 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 This version no longer represents clinical or dietetic practice in the UK or Ireland.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



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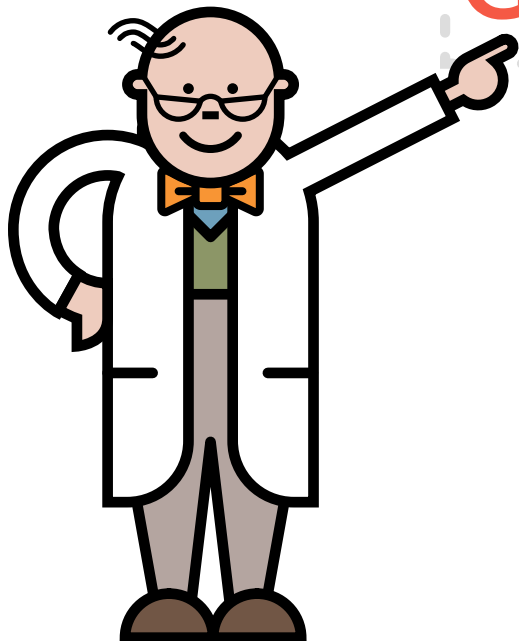
NUTRICIA

What is GA-1?

GA-1 stands for glutaric aciduria type 1.

It is pronounced glue-ta-ric-acid-ur-eea.

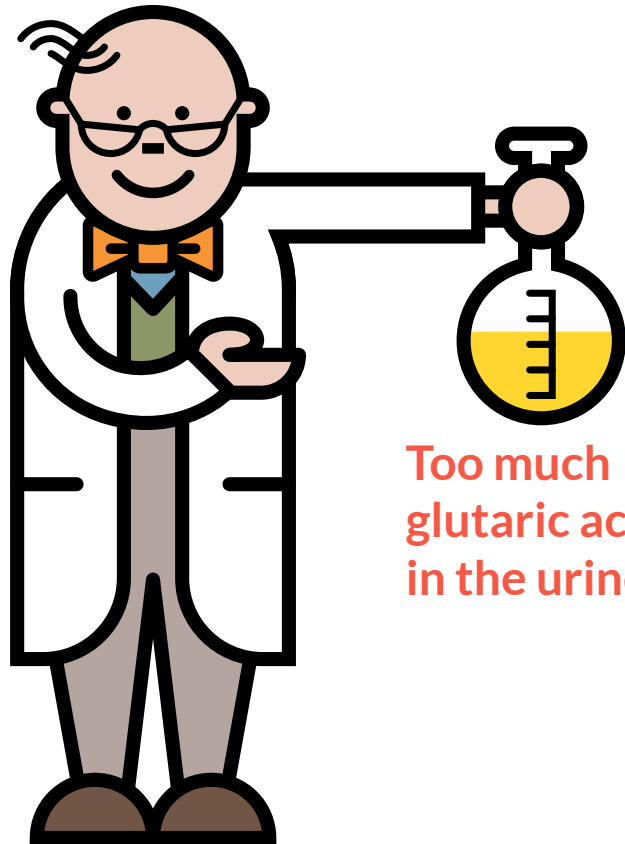
It is an inherited metabolic condition.



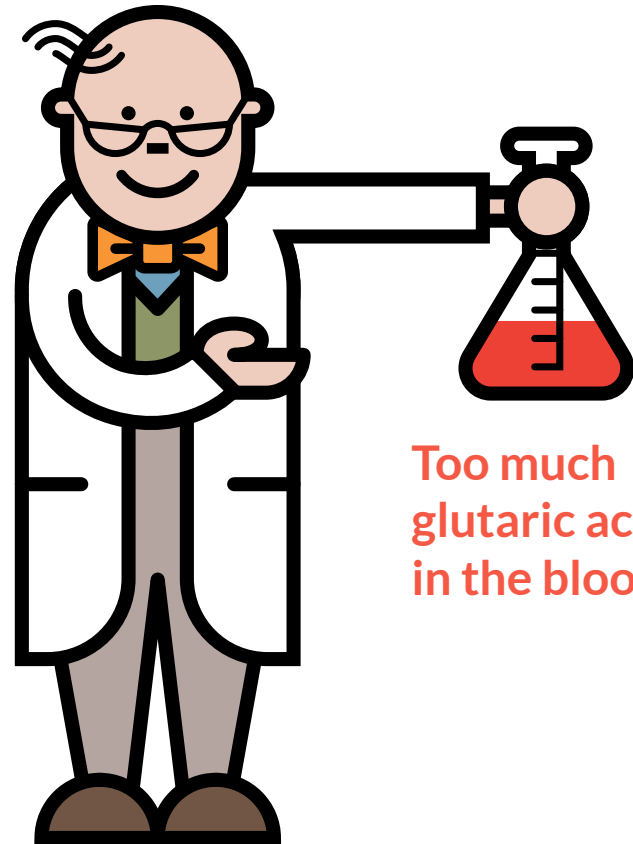
Glutaric Aciduria Type 1

GA-1

What is GA-1?



Too much
glutaric acid
in the urine

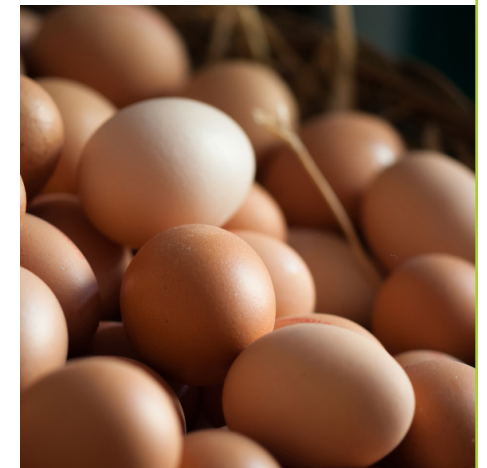


Too much
glutaric acid
in the blood

How does GA-1 affect the body?

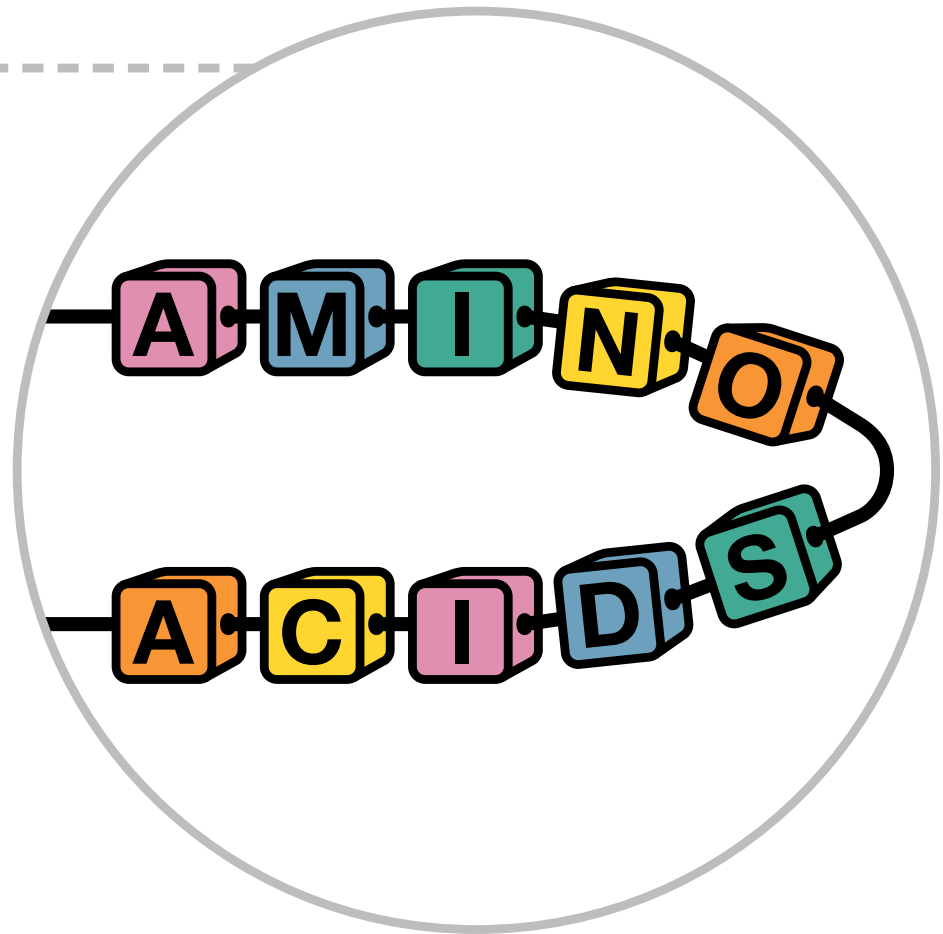
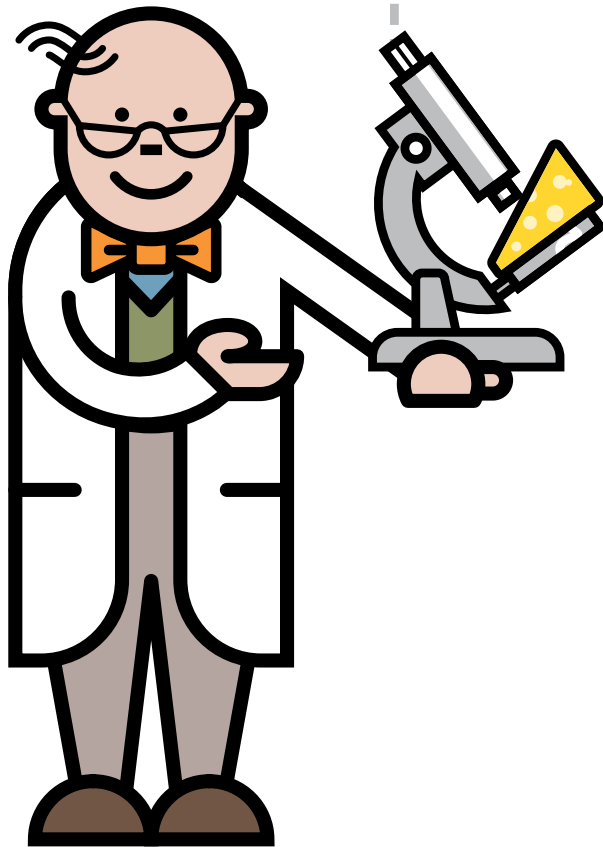
GA-1 affects the way the body breaks down protein.

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



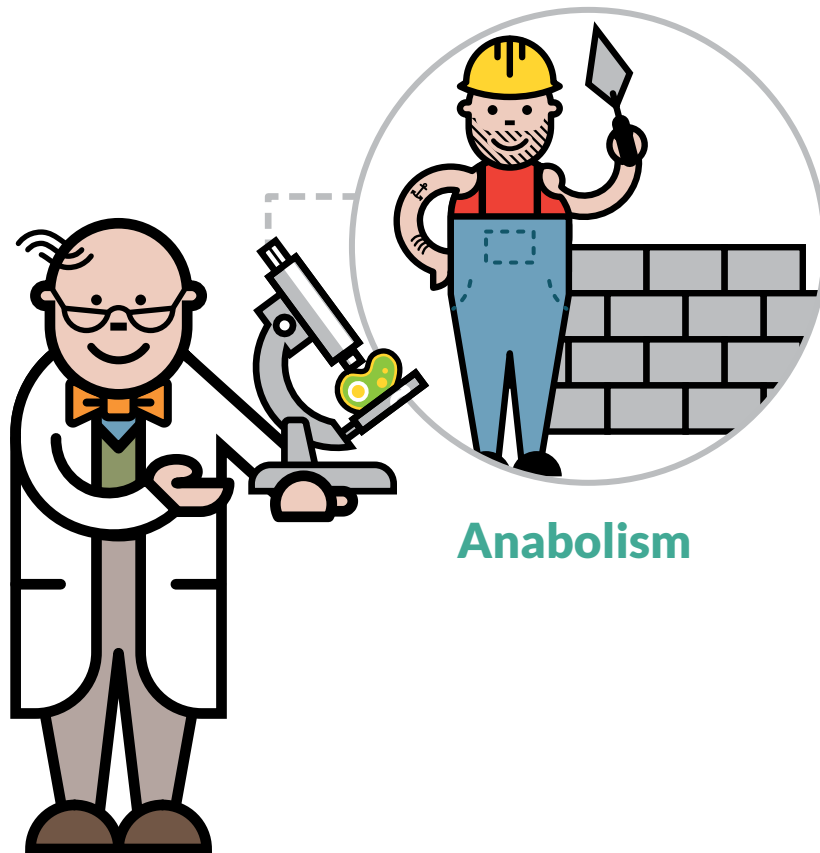
What is protein?

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

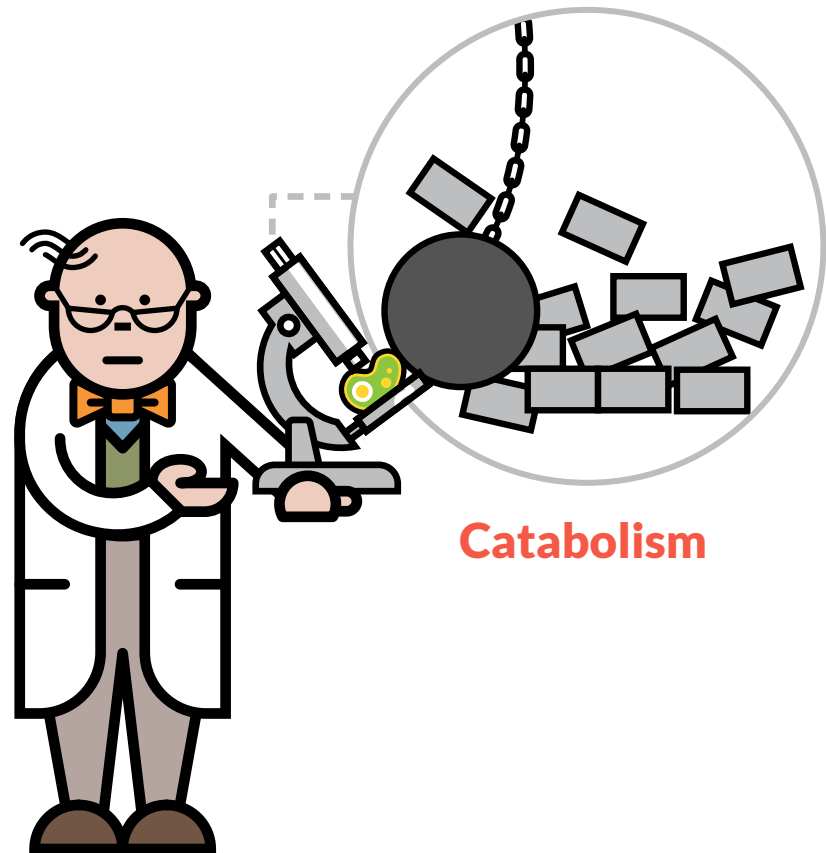


Protein metabolism

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



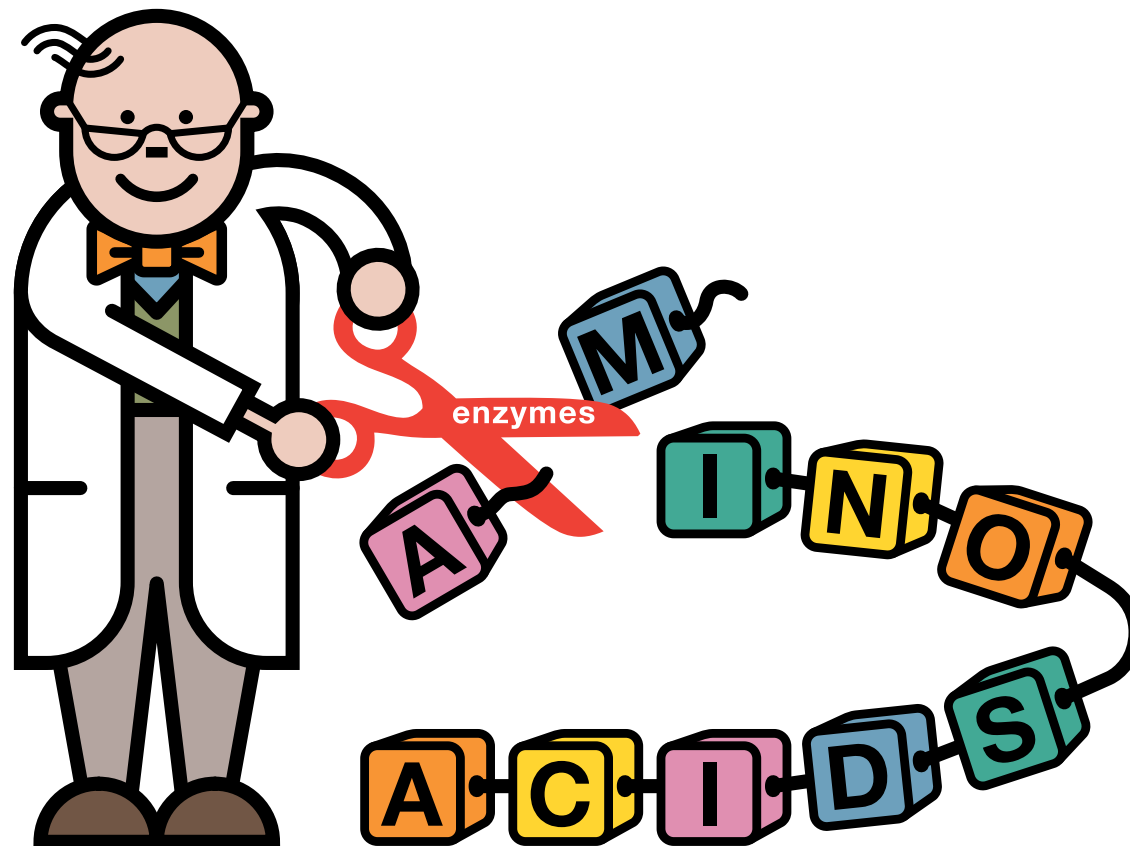
Anabolism



Catabolism

What do enzymes do?

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

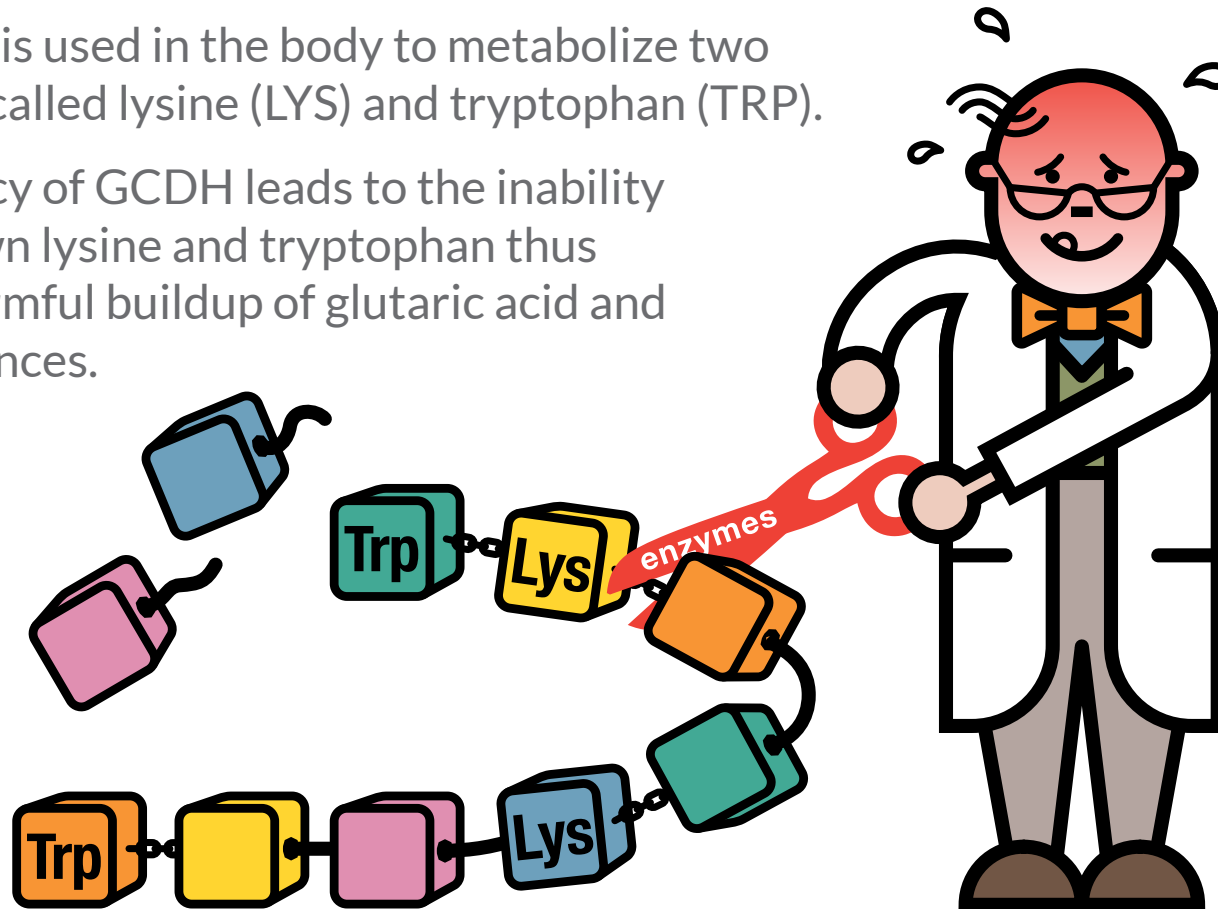


What happens in GA-1?

GA-1 is caused by a deficiency of an enzyme called **glutaryl-CoA dehydrogenase (GCDH)**.

This enzyme is used in the body to metabolize two amino acids called lysine (LYS) and tryptophan (TRP).

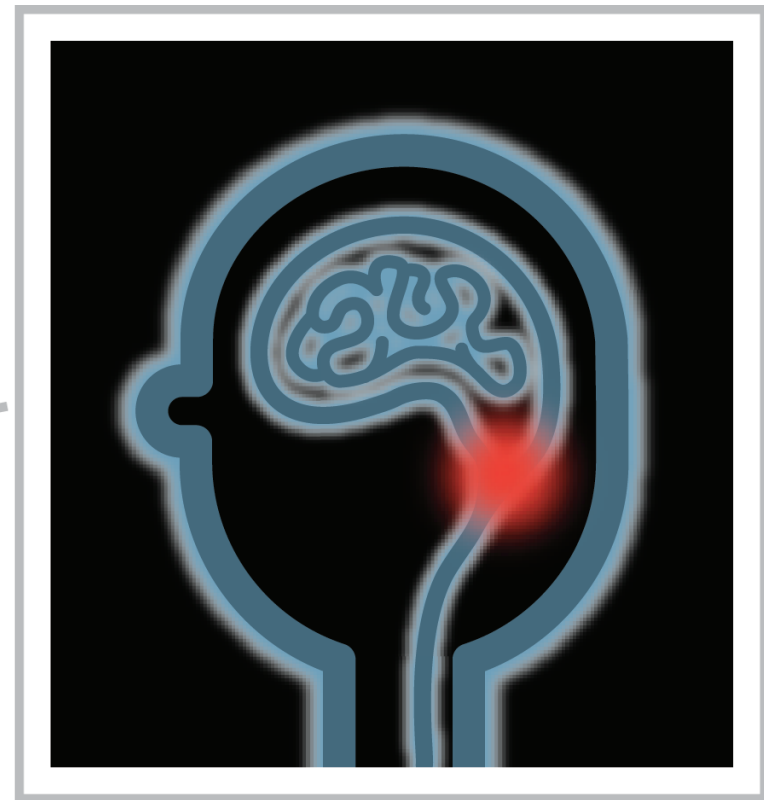
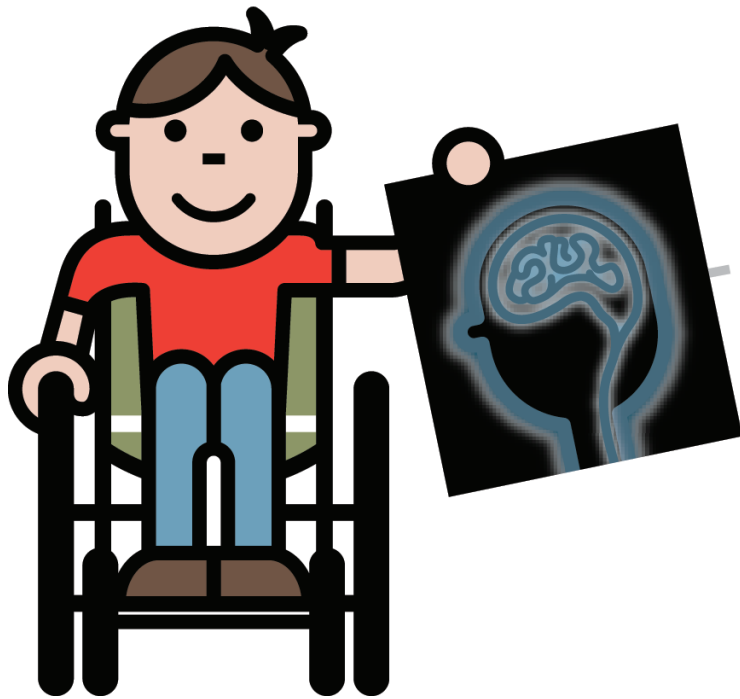
The deficiency of GCDH leads to the inability to break down lysine and tryptophan thus causing a harmful buildup of glutaric acid and other substances.



What can go wrong in GA-1?

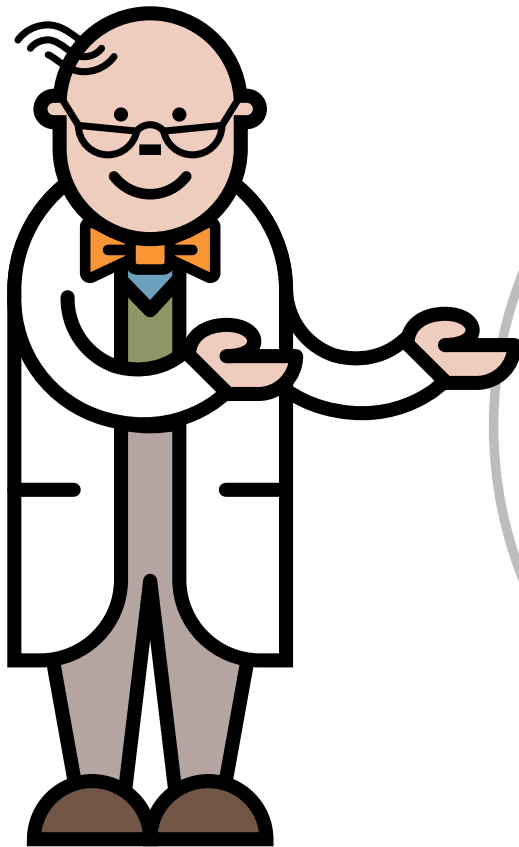
The basal ganglia in the brain controls movement.

The buildup of glutaric acid and other substances damage the basal ganglia and causes movement problems. Intellectual disability may also occur.



What about other symptoms?

Babies with GA-1 are usually healthy at birth although many are born with a larger than average head size.



How is GA-1 diagnosed?

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

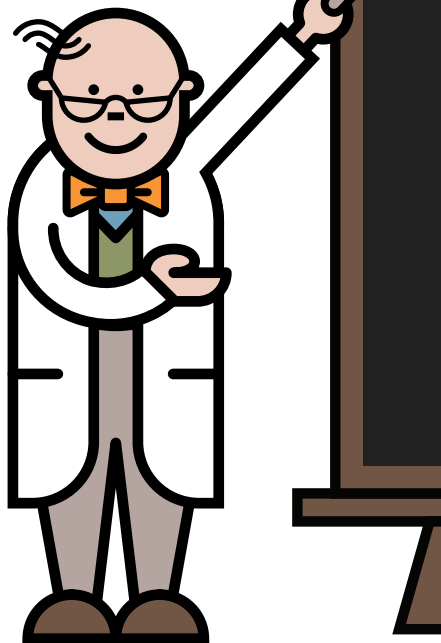
The blood sample is then analyzed.

Abnormal results could mean your child has GA-1 which will prompt your clinician to do further testing to confirm the diagnosis.



How is GA-1 managed day-to-day?

1. A whole protein restricted diet
2. A metabolic formula, prescribed by your clinic
3. When recommended, carnitine supplement



Avoid high protein foods

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



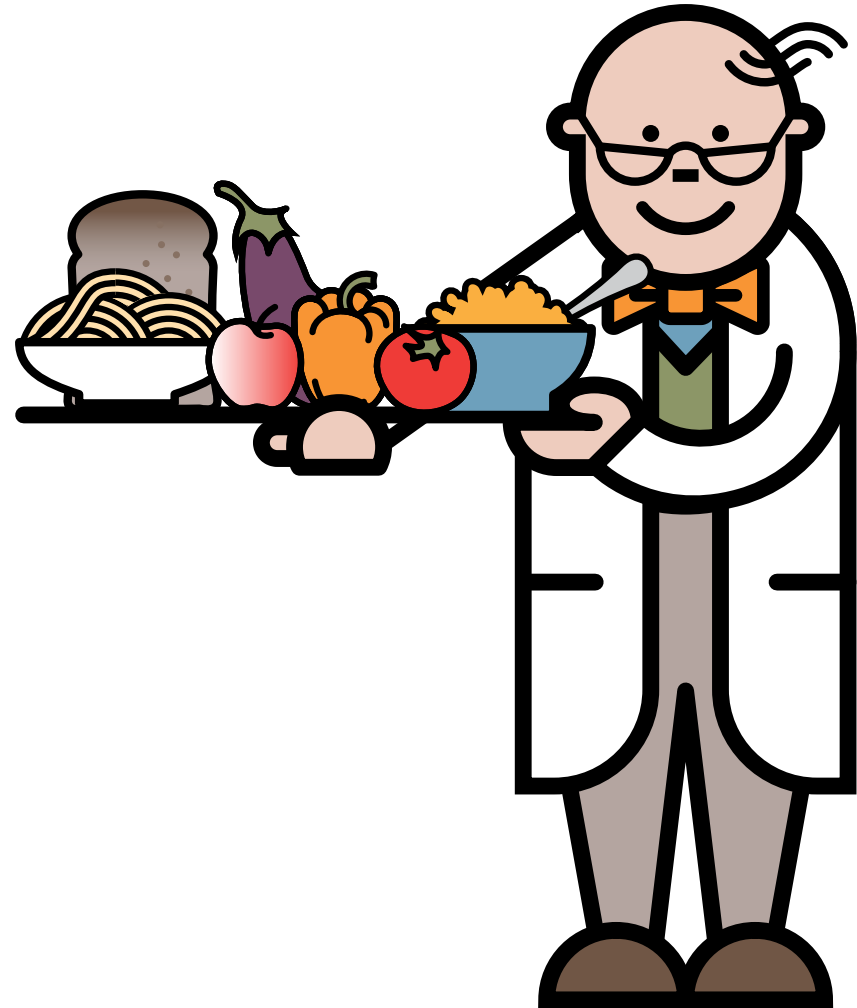
Include foods low in protein

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

They include many fruits and vegetables, and special 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

Lysine and tryptophan are essential for normal development and therefore a limited and controlled amount must be taken daily.

Breast milk or standard infant formula will provide the lysine and tryptophan 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 lysine and a small amount of tryptophan.

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

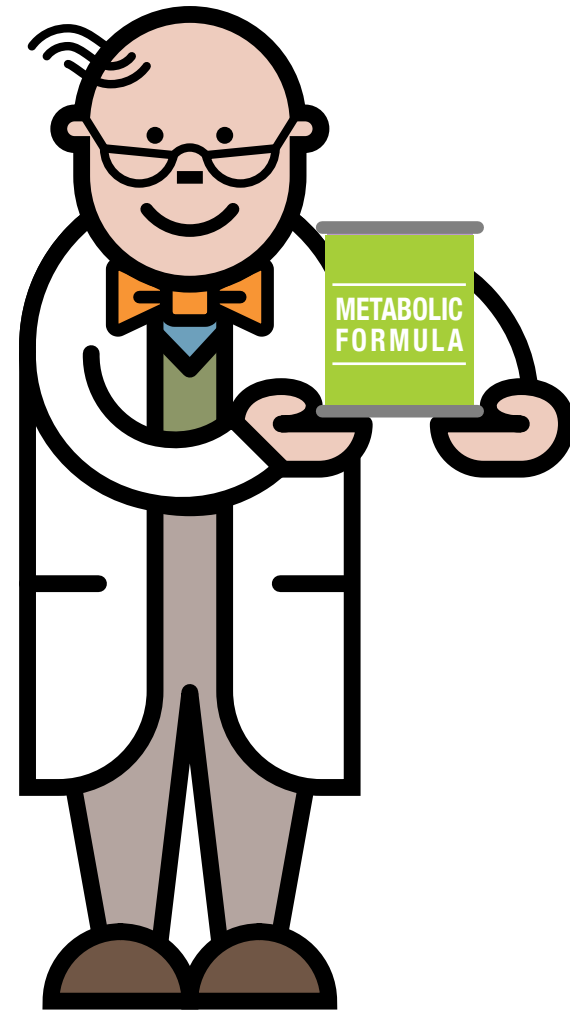


Lysine free and low tryptophan metabolic formula

Lysine-free, low tryptophan 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 and minerals; while the protein comes in the form of amino acids without lysine and low levels of tryptophan.

Metabolic formula, plus the prescribed amounts of lysine and tryptophan, allows your baby to get all the nutrients he or she needs to grow.



Tracking lysine and tryptophan

As your baby starts to eat solids your clinic will work with you to track lysine and tryptophan.

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

Your clinic can help you find the best tools to help determine the lysine and tryptophan content of foods.



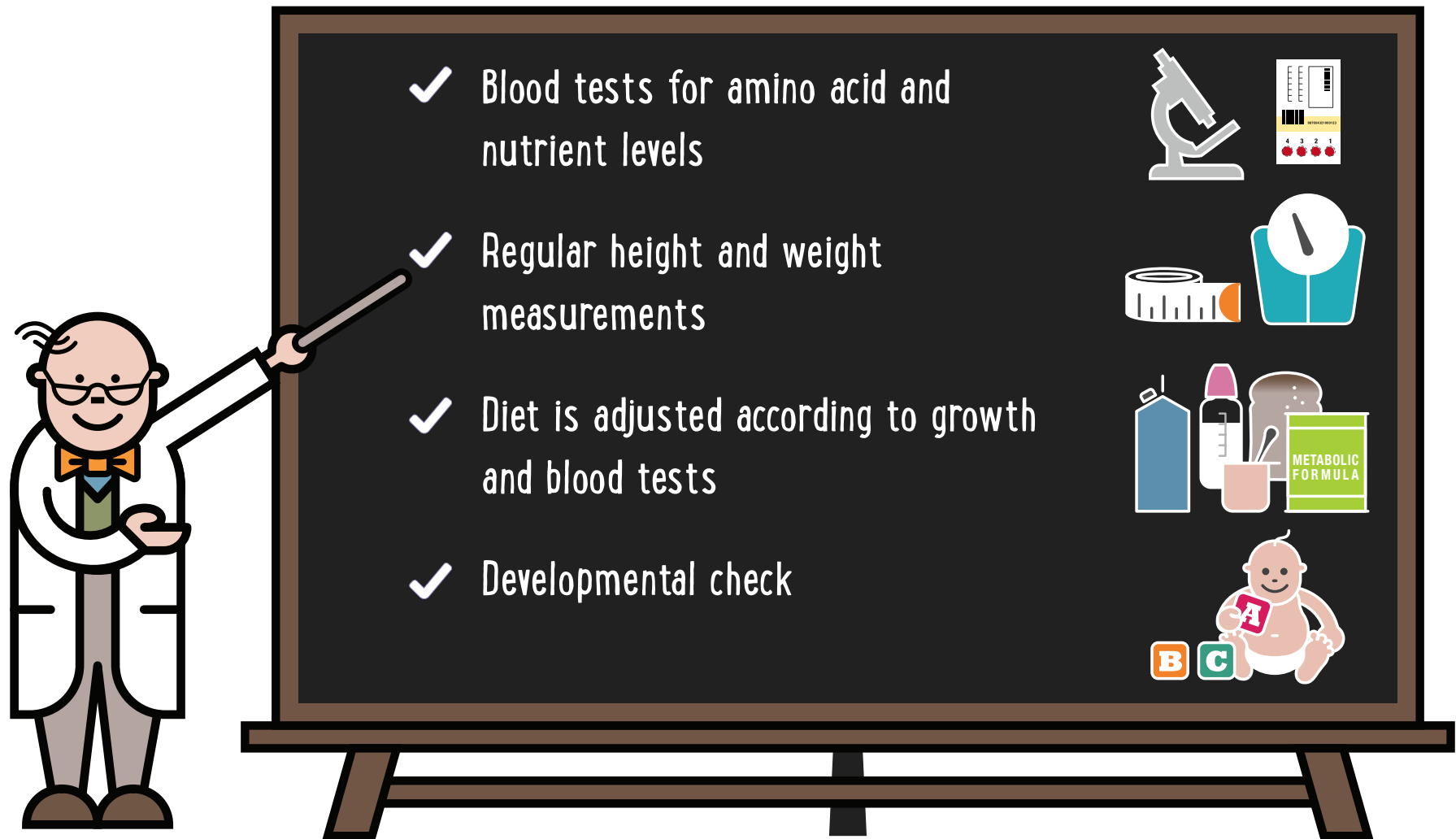
How is GA-1 monitored?

Regular blood and urine tests are reviewed by the clinic.

The metabolic dietitian will contact you with the result and discuss any changes in management.



What happens during a clinic check-up?



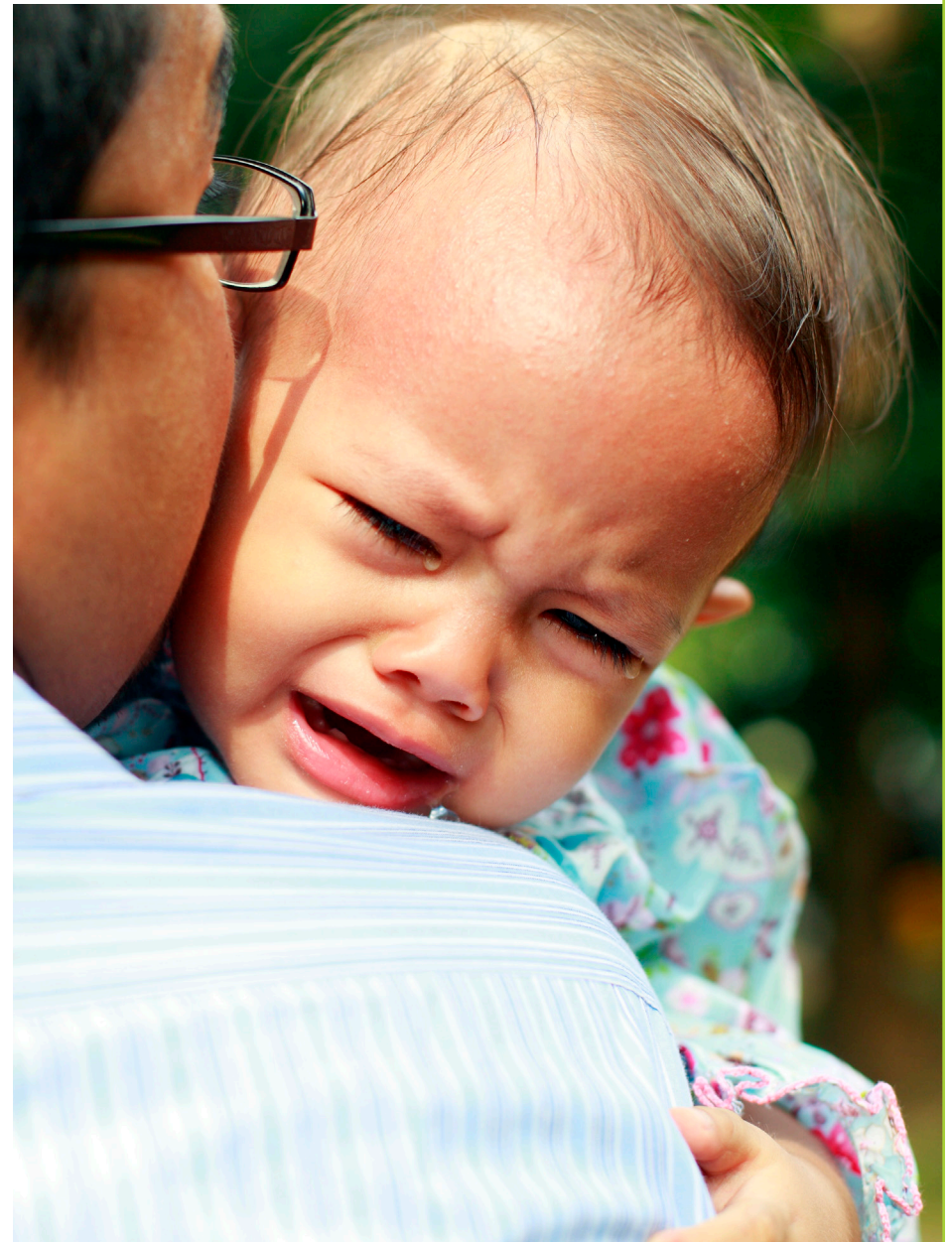
- ✓ Blood tests for amino acid and nutrient levels
- ✓ Regular height and weight measurements
- ✓ Diet is adjusted according to growth and blood tests
- ✓ Developmental check

Metabolic crisis

A '**metabolic crisis**' triggers the 'movement problems'. This is because there is a buildup of glutaric acid and other toxic chemicals.

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

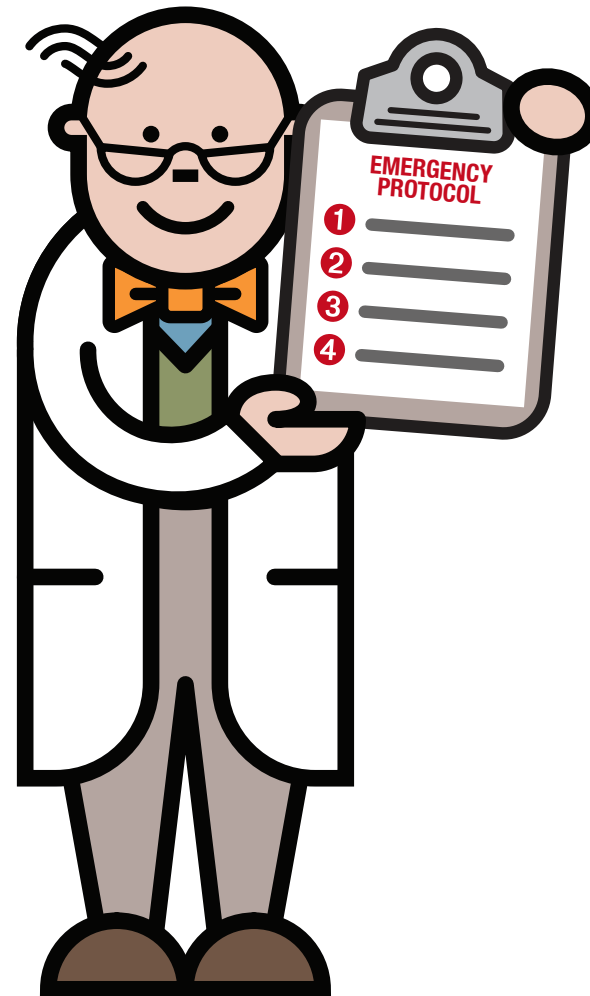
It is important to manage a metabolic crisis quickly and properly.



How is GA-1 managed during illness?

During any illness, our bodies need extra energy. The body will start breaking down cell protein, a process called catabolism. This will lead to a buildup of glutaric acid and other harmful substances, 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.

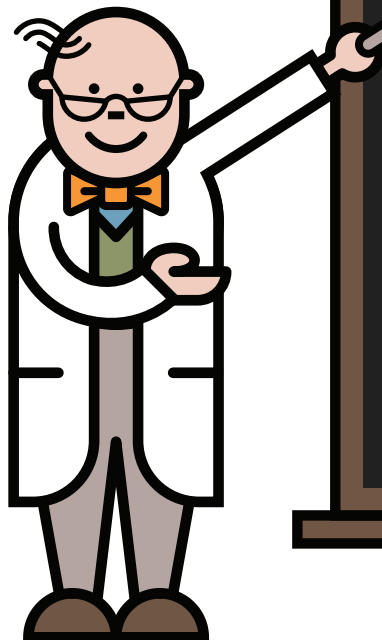
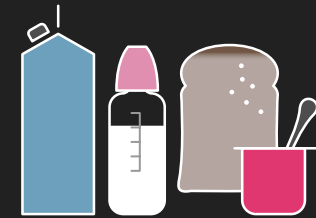


How is GA-1 managed during illness?

Always follow your medical team's guidance.

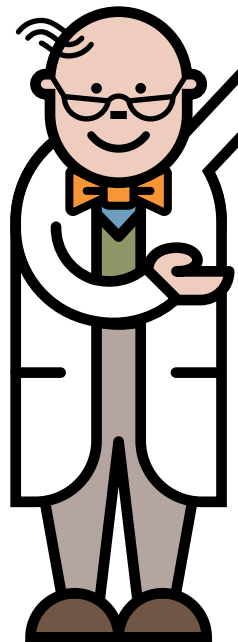
Contact your medical team at first signs of illness. They may have you:

- ✓ Stop all protein in food & drink
- ✓ Start the emergency protocol. This is made up of metabolic formula and glucose
- ✓ Continue carnitine supplement, if prescribed



How is GA-1 managed during illness?

Always follow your medical team's guidance.



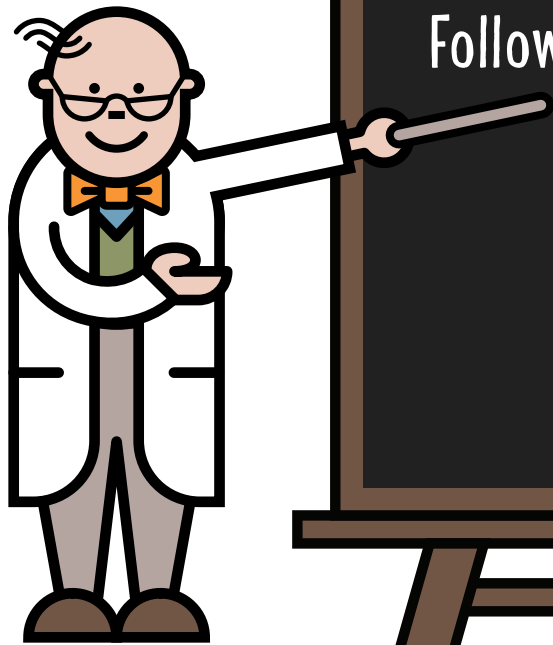
- ✓ Always take full amounts of emergency feeds as prescribed
- ✓ If symptoms continue and/or you are worried, go immediately to the hospital
- ✓ Regularly update your metabolic team



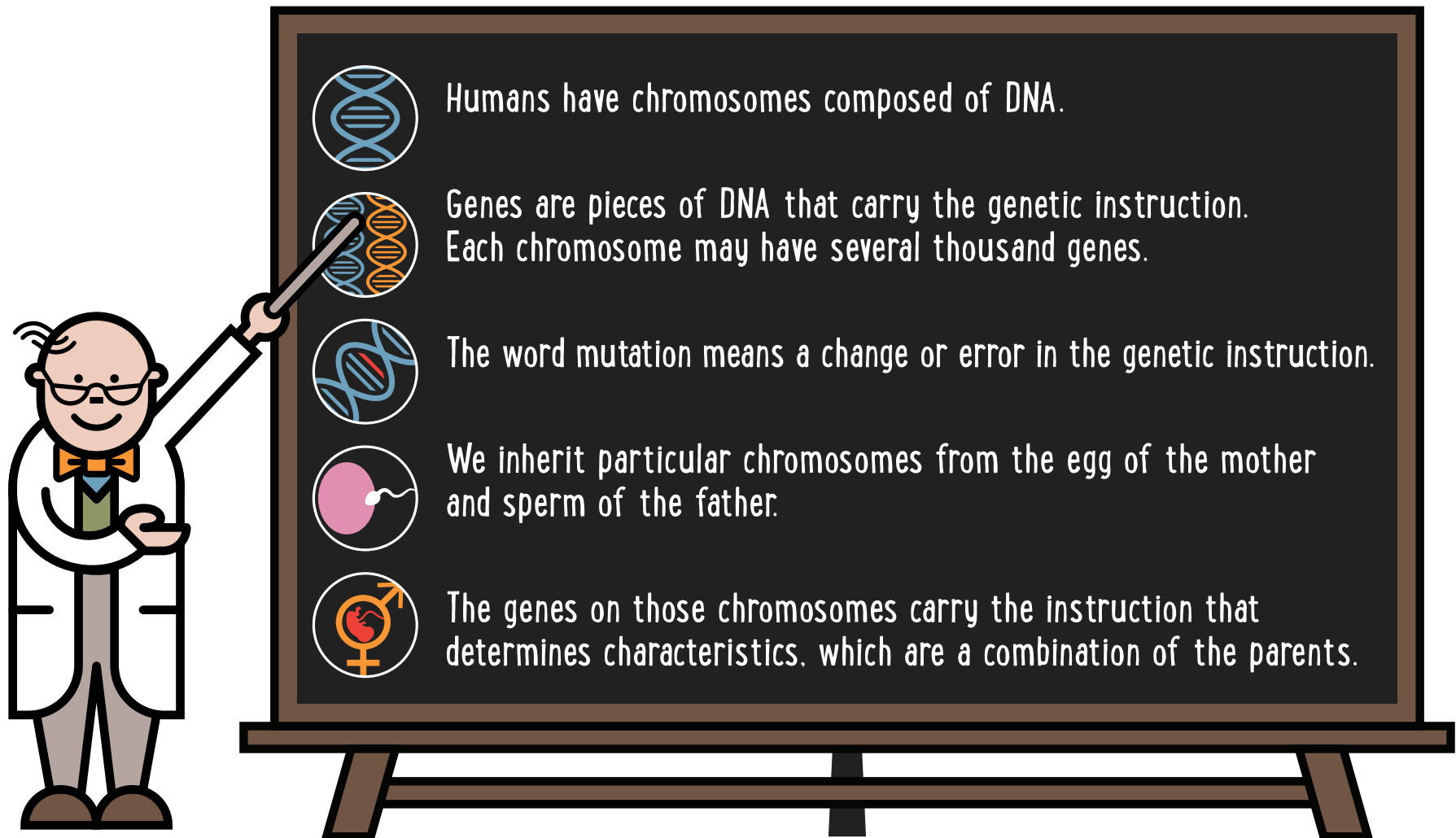
Most importantly


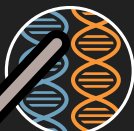



It is essential that you contact your metabolic team immediately if your child is getting sick.

Follow their instructions promptly without delay.

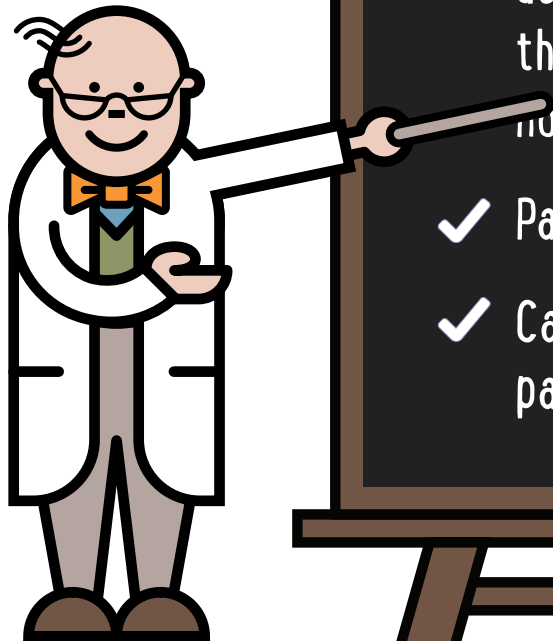


What happens in human genetics?



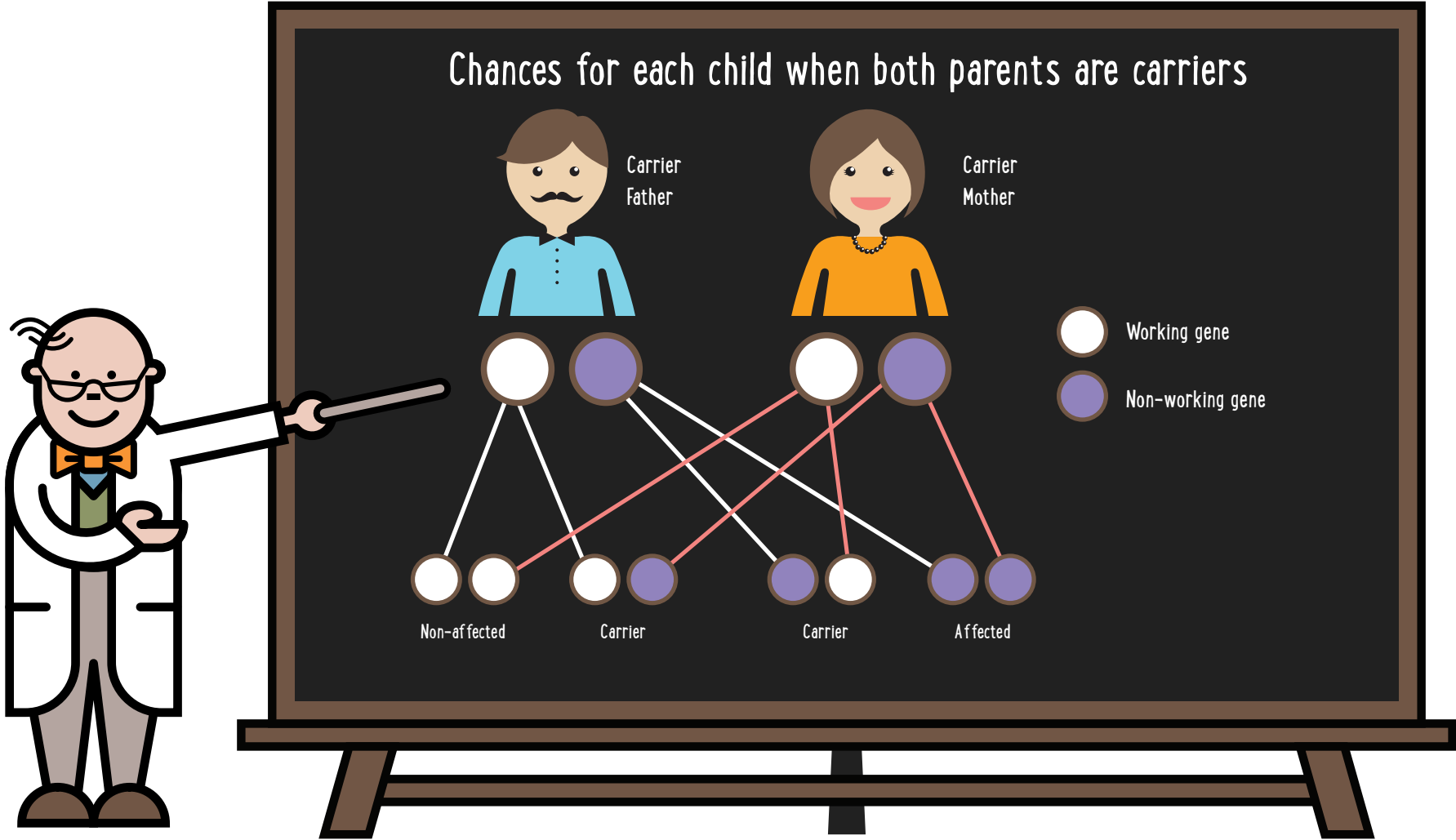
-  Humans have chromosomes composed of DNA.
-  Genes are pieces of DNA that carry the genetic instruction. Each chromosome may have several thousand genes.
-  The word mutation means a change or error in the genetic instruction.
-  We inherit particular chromosomes from the egg of the mother and sperm of the father.
-  The genes on those chromosomes carry the instruction that determines characteristics, which are a combination of the parents.

How does one inherit GA-1?



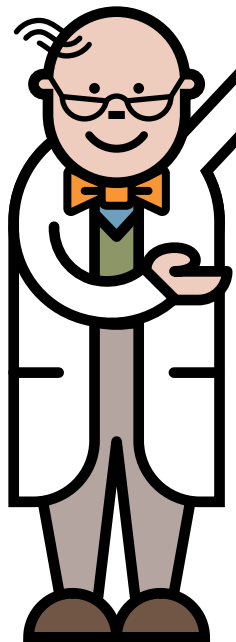
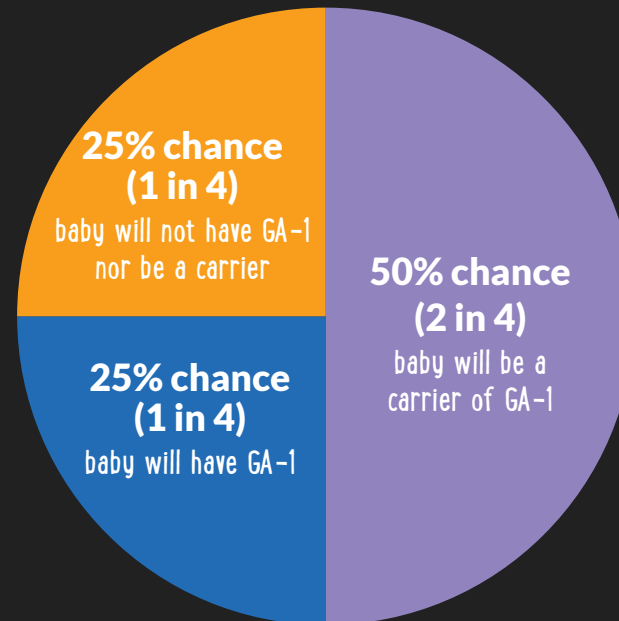
- ✓ GA-1 is an inherited condition. There is nothing that could have been done to prevent the child from having GA-1.
- ✓ Everyone has a pair of genes that make the glutaryl-CoA dehydrogenase enzyme. In children with GA-1, neither of these genes works correctly. These children inherit one non-working GA-1 gene from each parent.
- ✓ Parents of children with GA-1 are carriers of the condition.
- ✓ Carriers do not have GA-1 because the other gene of this pair is working correctly.

Inheritance – Autosomal-recessive – possible combinations

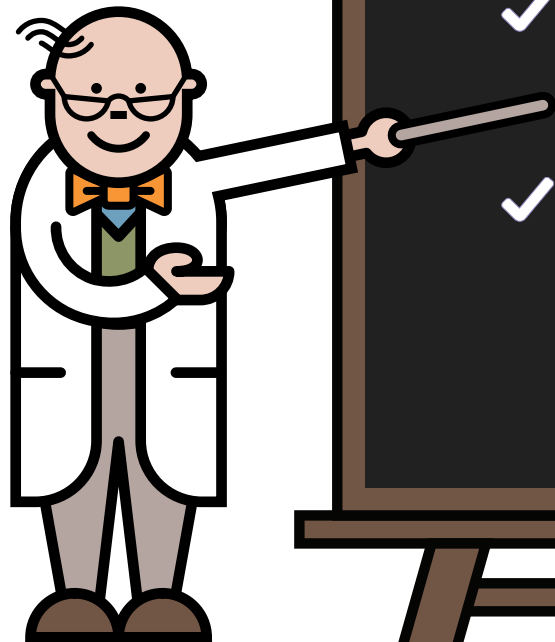


Future pregnancies

When both parents are carriers, *in each pregnancy* the risk to the baby is as follows:

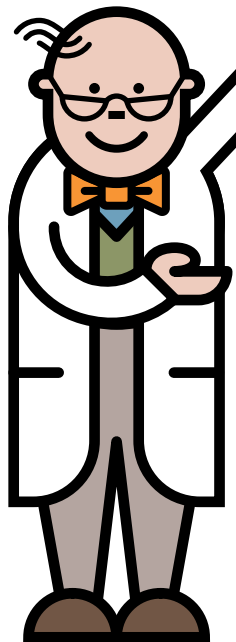


Take home messages



- ✓ GA-1 is a serious inherited metabolic disorder that can lead to severe movement problems and intellectual disability.
- ✓ Damage can be prevented with a protein restricted diet, a special metabolic formula and carnitine.
- ✓ Remember, in case of illness, it is imperative that you contact your metabolic team immediately. They will determine the best course of action.

Helpful hints



- ✓ Always ensure you have a good supply of your low protein foods, metabolic formula and that they are not expired.
- ✓ Your special dietary products and metabolic formula are prescribed by your metabolic clinic.
- ✓ Always ensure you have your emergency regimen products and a written emergency plan.
- ✓ When your child is ill, or not behaving normally, it is important to communicate with your metabolic team immediately to prevent metabolic crisis.

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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ZGA1TBEN 11/19

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