

TEMPLE



Tools **E**nabling **M**etabolic **P**arents **L**Earning

ADAPTED BY THE DIETITIANS GROUP

BIMDG

British Inherited Metabolic Diseases Group



BASED ON THE ORIGINAL TEMPLE WRITTEN BY
BURGARD AND WENDEL

VERSION 1, DECEMBER 2022

GSD III

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).

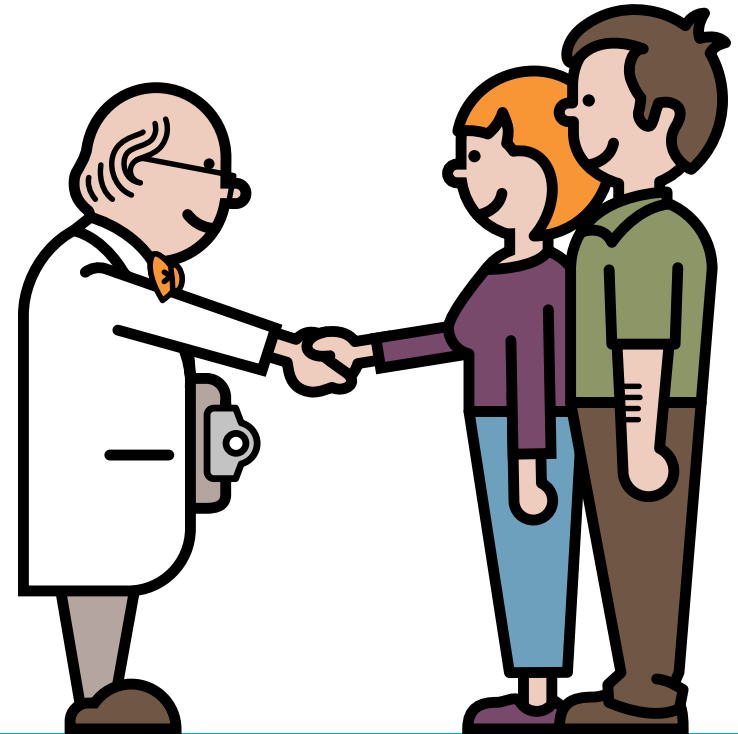
The team are Rachel Skeath, Karen van Wyk, Pat Portnoi and Anita MacDonald. The group is facilitated by Heidi Chan from Nutricia.

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.

GSD III

Information for families following
a new diagnosis



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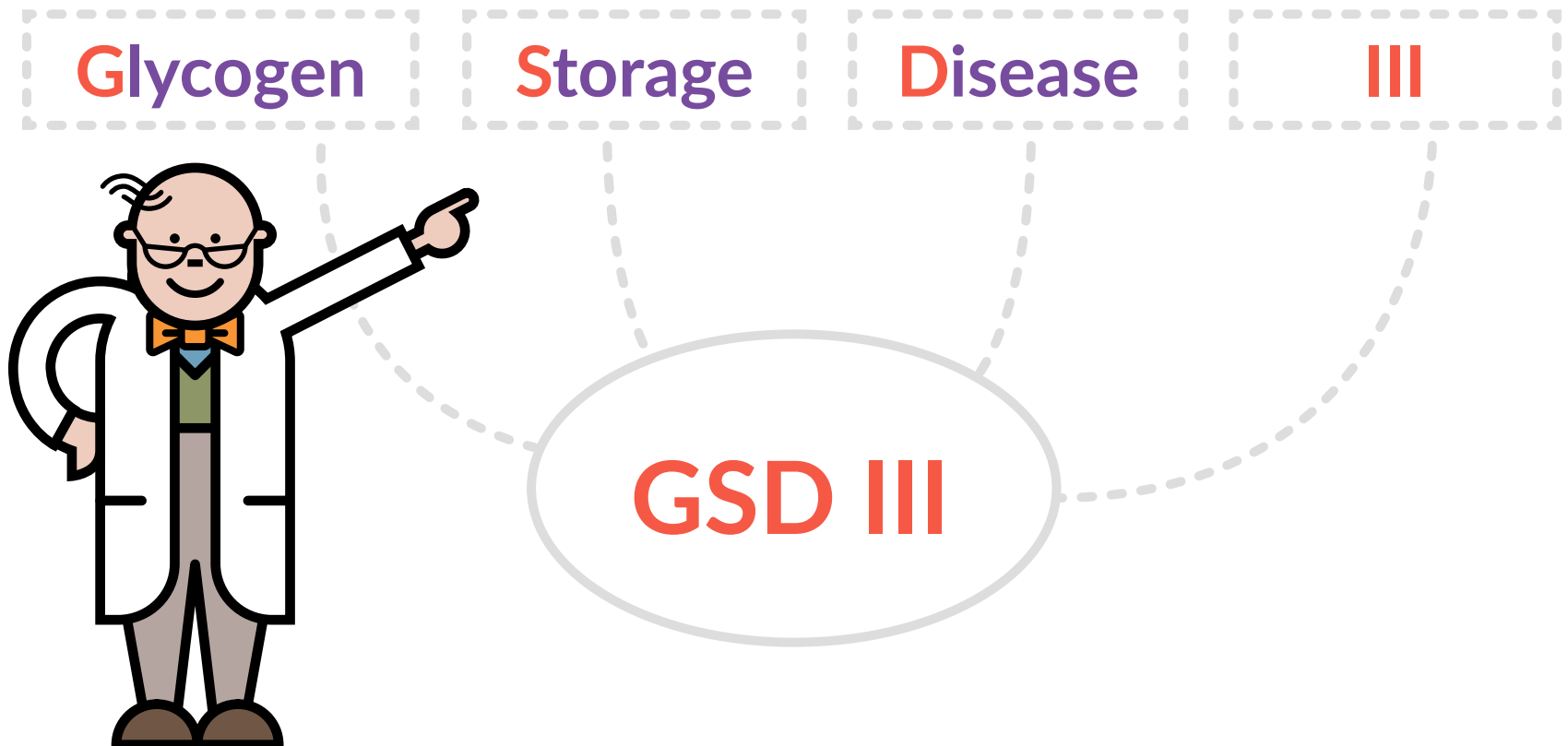


Tools Enabling Metabolic Parents LEarning

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What is GSD III?

GSD III stands for Glycogen Storage Disease III
It is an inherited metabolic condition



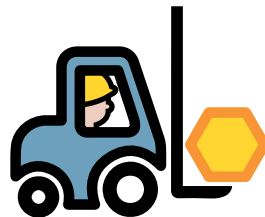
Glucose and glycogen for energy

We eat food to supply our body with energy. Our food contains carbohydrate, protein and fat.

Carbohydrate is an important supply of energy. It is broken down into a sugar called glucose.

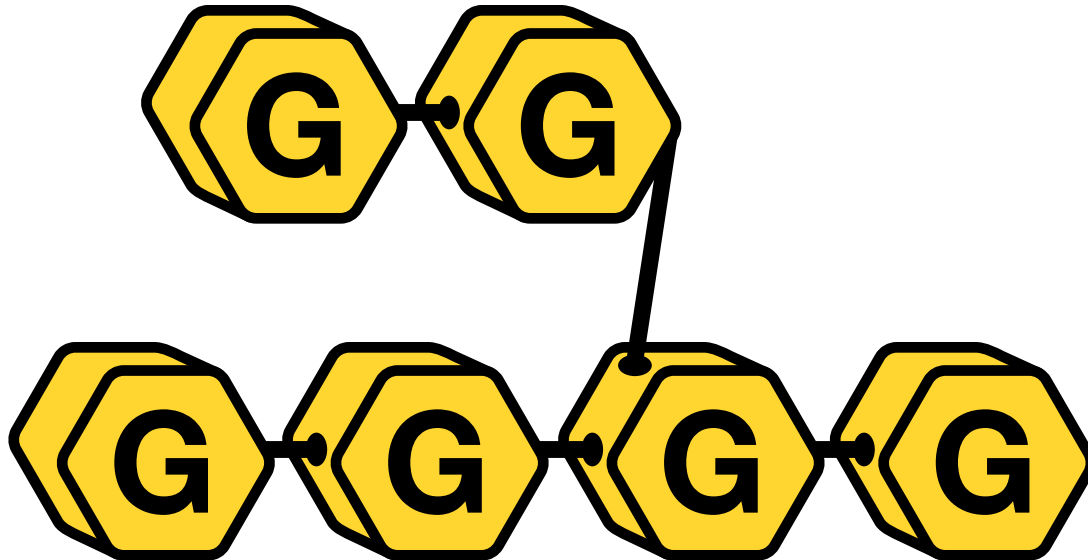
Some of this glucose from food goes into our blood stream and will give a *readily available* supply of energy.

We also put some glucose into storage as an energy reserve. **This is converted into glycogen in the liver and muscle.**



What is glycogen?

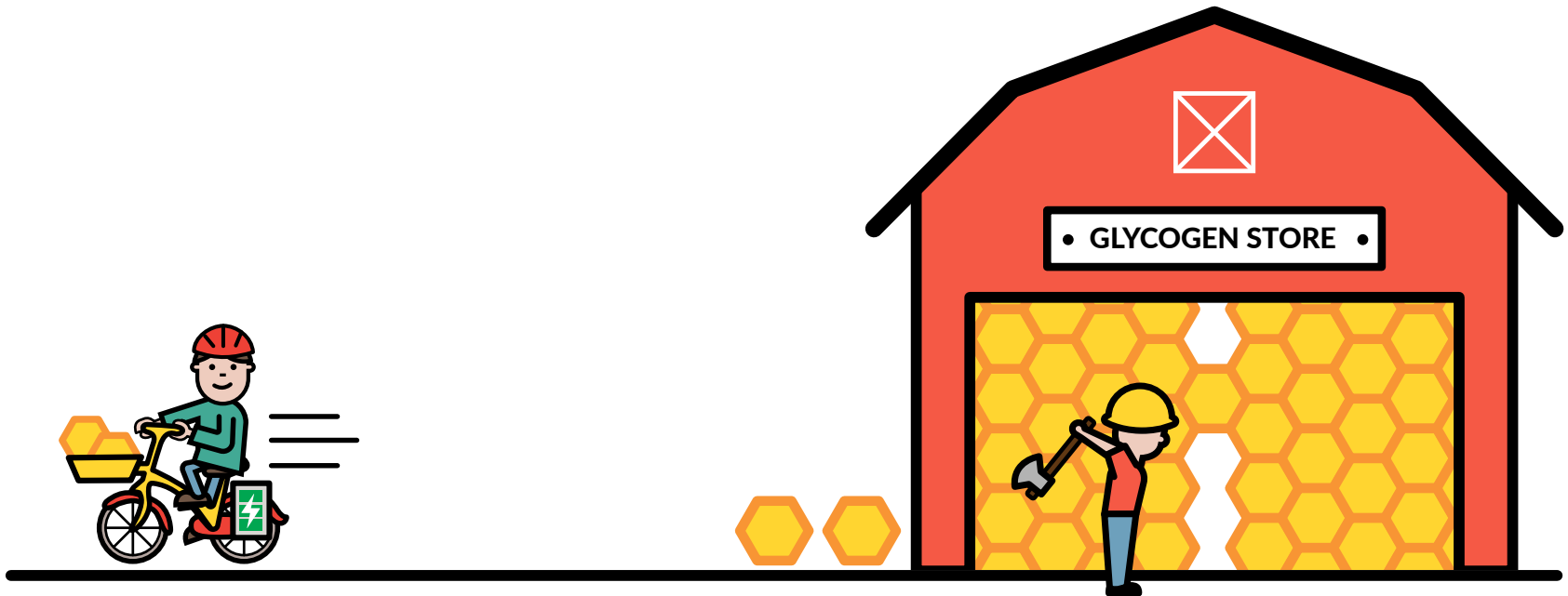
Glycogen is made up of units of glucose joined together.



Glucose and glycogen for energy

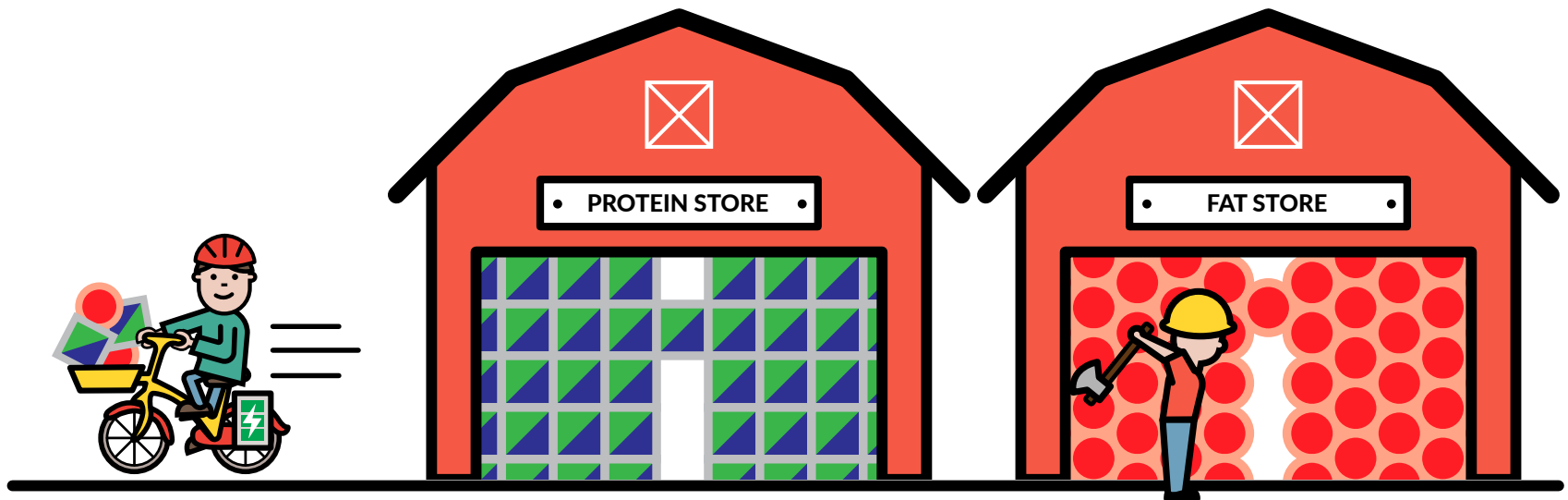
When energy is needed, the body uses the *readily available* glucose as its energy supply.

Once glucose is used up, the body then breaks down the stored glycogen to make more glucose.



Other sources of energy

When the body has used up the *readily available* glucose it can also break down protein and fat to provide energy.



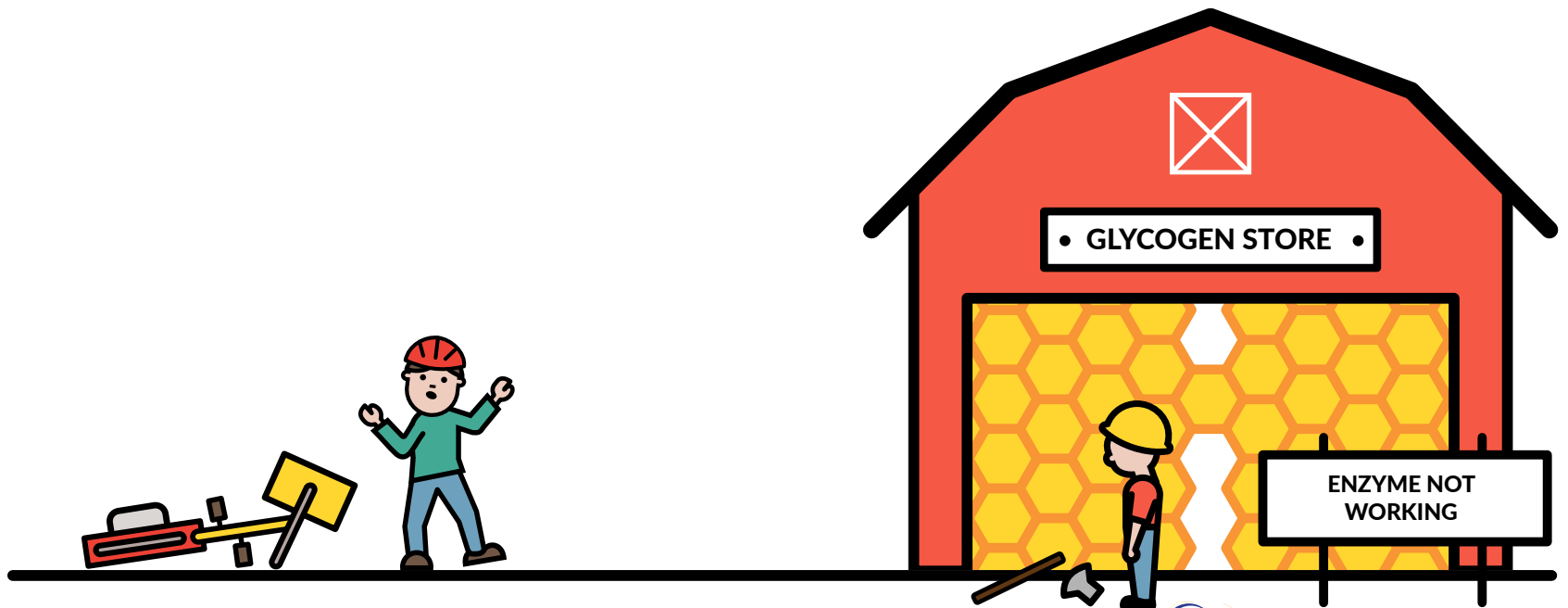
What happens in GSD III?

In GSD III there are problems breaking down the glycogen stores to release glucose.

The body lacks an enzyme called the debrancher enzyme that helps break down glycogen into glucose.

This stops the glycogen being used for energy. Instead it builds up in the muscle and the heart.

It causes a shortage of energy supply and a low blood glucose.

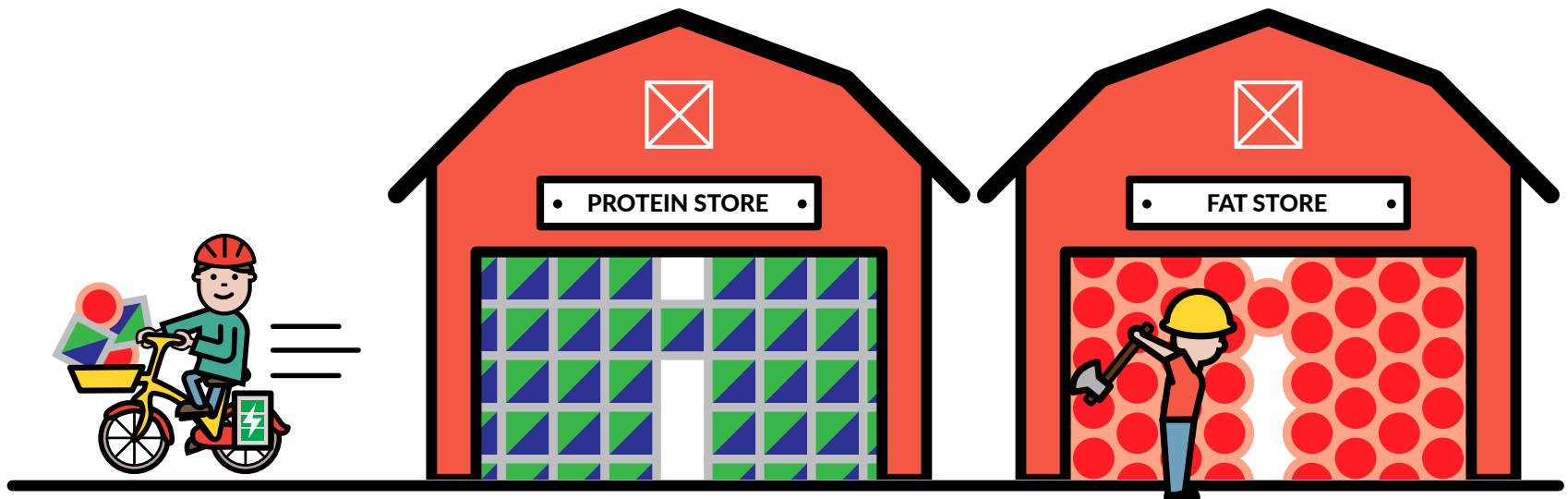


What happens in GSD III continued

Fortunately people with GSD III can still make energy by breaking down their protein and fat stores.

Therefore food containing protein and fat can be used to supply energy in GSD III.

Protein is particularly important to support muscles.



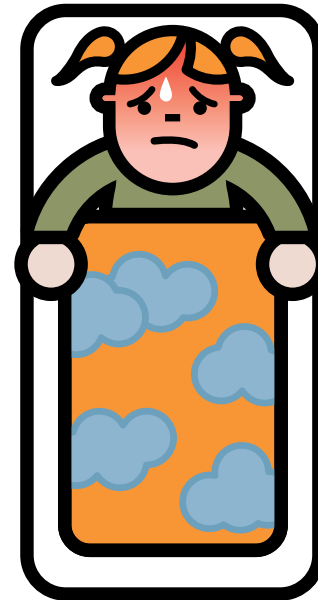
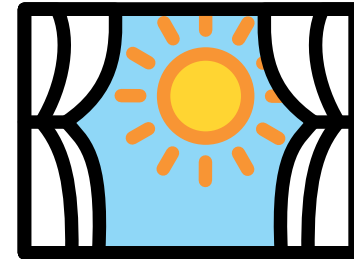
What can go wrong with GSD III?

Some babies become ill in the first few months of life.

Symptoms include:

- low blood glucose
- floppiness
- seizures
- large liver
- high levels of chemicals from the liver and fats in the blood
- poor growth

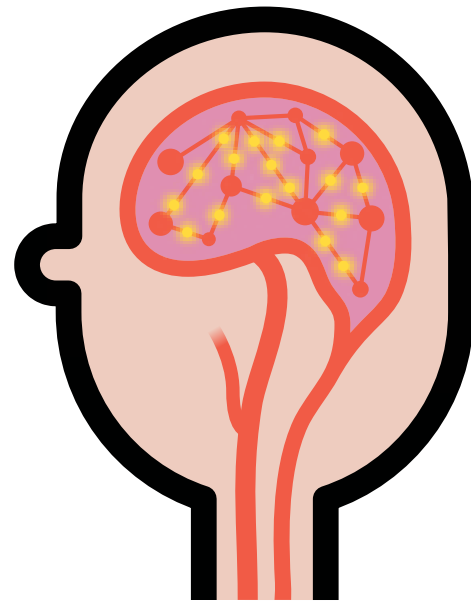
The effects of low blood glucose is very harmful if unmanaged.



What can go wrong with GSD III?

If there is a low blood glucose and this is not corrected, it can lead to coma and brain damage. It may be life threatening.

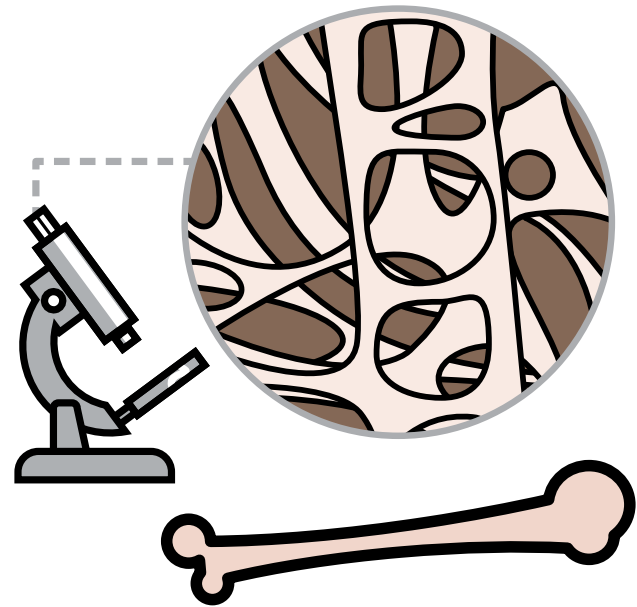
However, please remember, this can be prevented with timely and correct management.



What else can go wrong in GSD III?

Older children can develop:

- heart problems
- muscle pain and weakness which may affect ability to exercise
- thin bones
- short height



How is GSD III diagnosed?

GSD III is suspected because of the pattern of chemicals found in the blood.

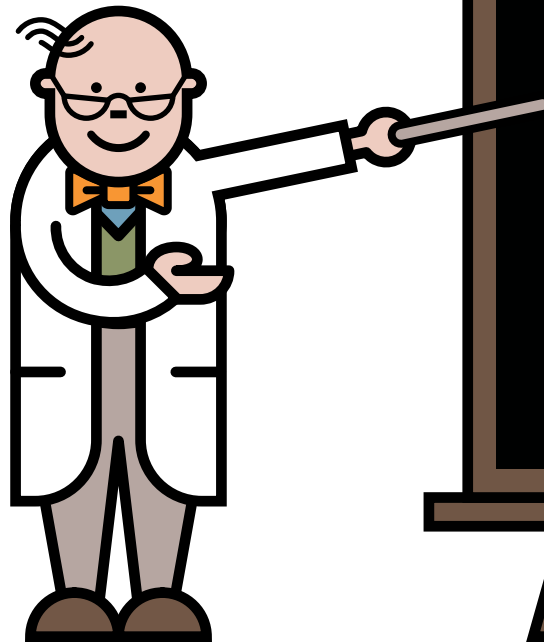
The diagnosis is confirmed by looking for mistakes in the gene for the debrancher enzyme.



How is GSD III managed?

It is managed by **avoiding** long periods without feeding.

The length of time that someone can go without feeds or eating is known as the **safe fasting time**.



SAFE FASTING TIMES

- The safe fasting time varies for each person with GSD III. Your metabolic team will advise
- It is important that regular feeds are given during the day and at night
- Scheduled feeds or meals should not be missed or delayed

How is GSD III managed day to day?

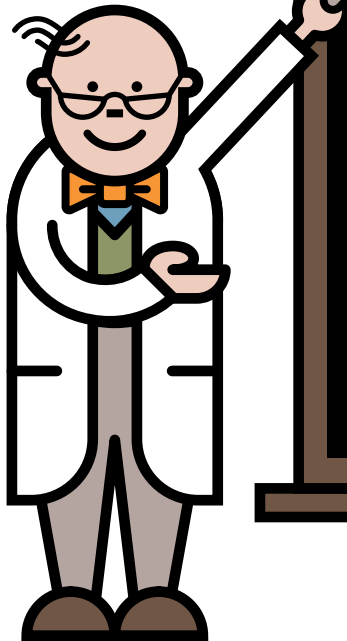
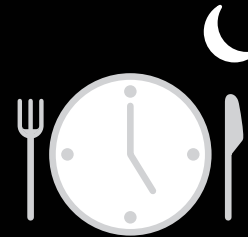
Regular meals and snacks based on carbohydrate and protein



High protein intake

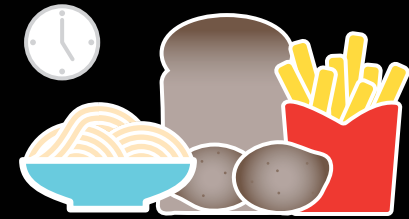


Overnight feeds or regular feeds/
drinks overnight



How is GSD III managed day to day?

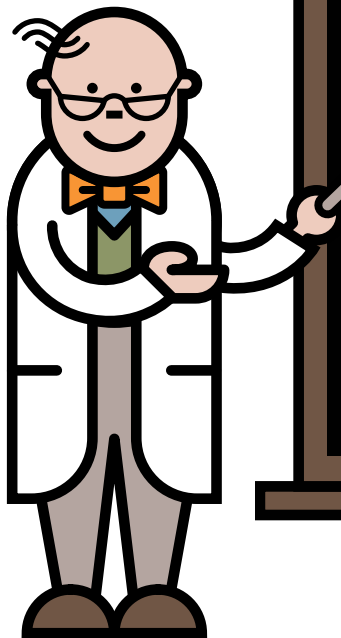
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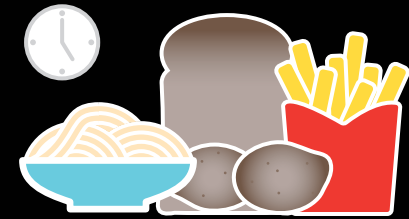


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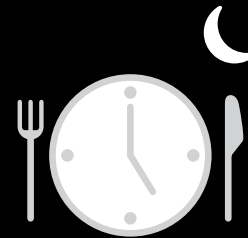
Regular meals and snacks based on carbohydrate and protein



High protein intake



Overnight feeds or regular feeds/
drinks overnight



Which foods contain carbohydrate?

Many foods contain carbohydrate.

Good sources of carbohydrate include breast milk, bread, rice, potato, pasta and cereals.



Which foods contain protein?

Good sources of protein include meat, fish, beans, pulses, eggs and dairy foods.

Sometimes extra protein supplements are used.



Uncooked cornstarch

- Uncooked cornstarch is a source of carbohydrate that is slowly broken down into glucose
- Sometimes a modified cornstarch is used instead
- These are commonly used in GSD III to extend safe fasting times
- Your dietitian will talk to you about this

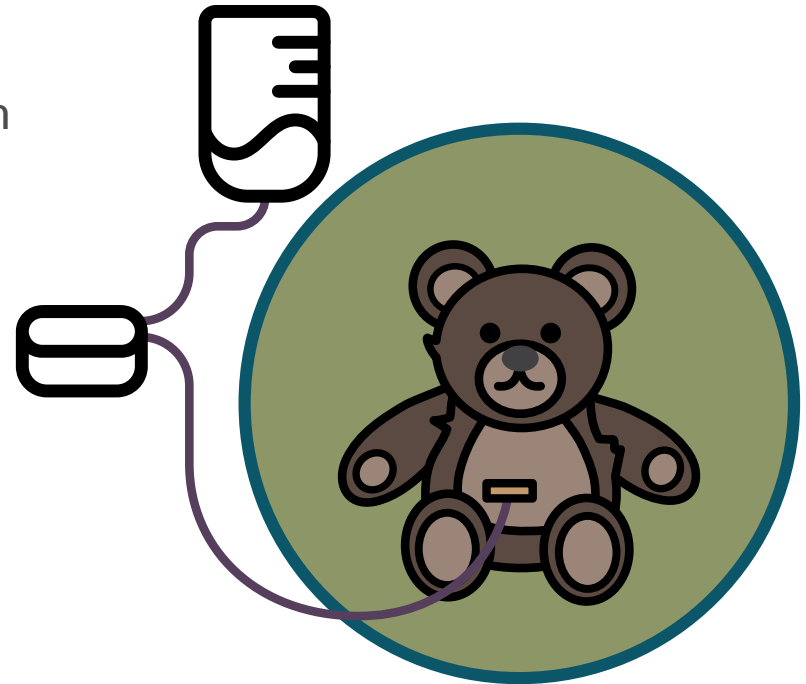


Overnight feeding

Babies and children with GSD III may not safely fast overnight without their blood glucose dropping too low.

They will need to have regular drinks or feeds during the night.

Many babies and children are fed through a tube overnight so that they can sleep without having to wake up.

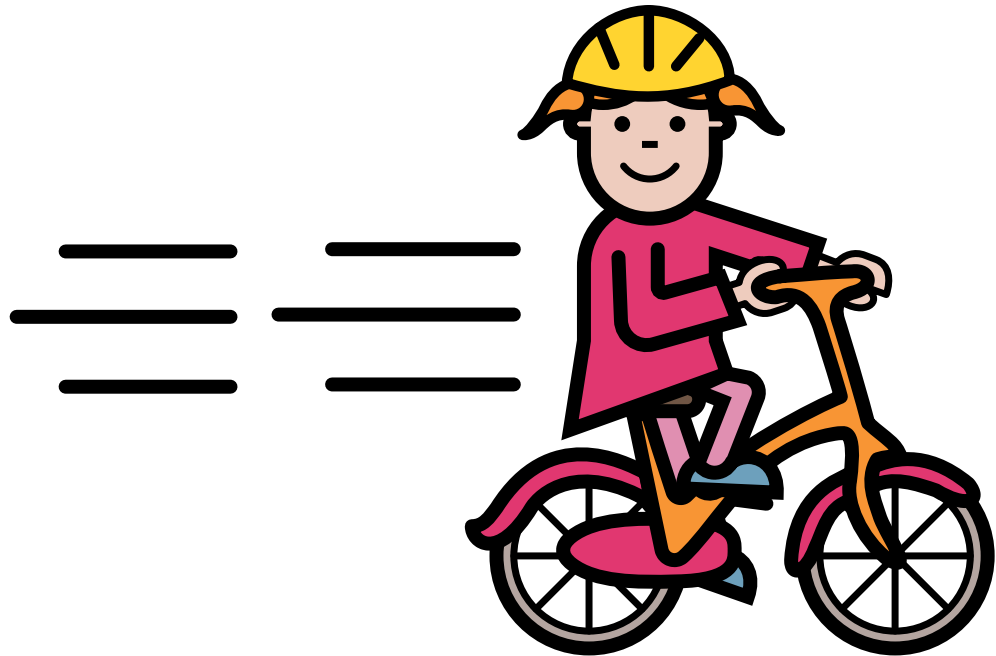


GSD III and exercise

Exercise and activity helps keep children with GSD III healthy.

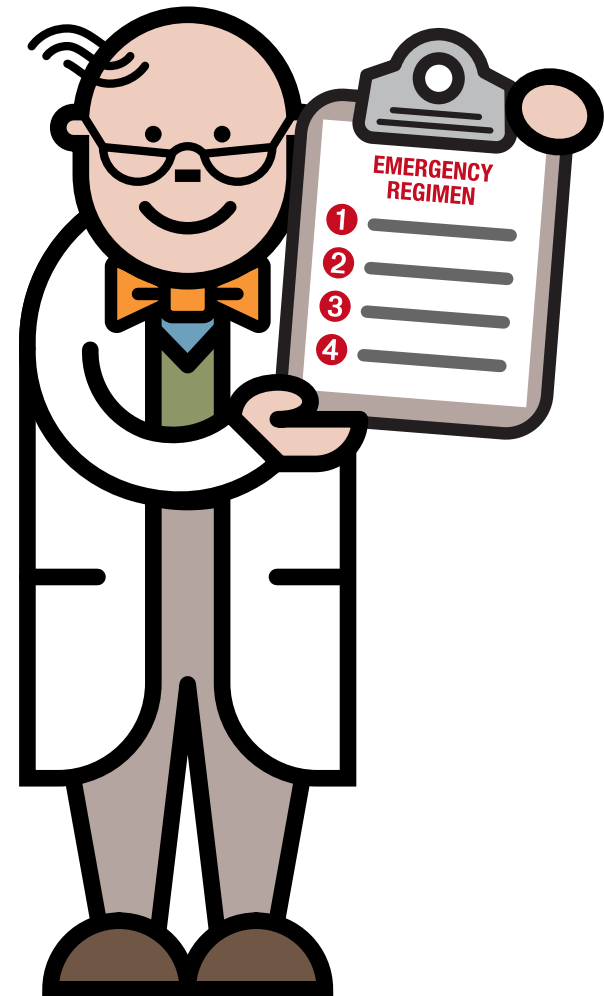
Some children find it difficult to exercise for long periods of time due to muscle weakness.

It might be advised that a snack or drink is taken before exercise. This will supply an extra source of energy.



How is GSD III managed during illness?

- During illness your child may not be able to follow their usual feeding plan
- You will be given an emergency regimen to use during this time
- This provides energy and prevent low blood glucose



How is GSD III managed during illness?

Start the emergency regimen.
This is made up of glucose polymer



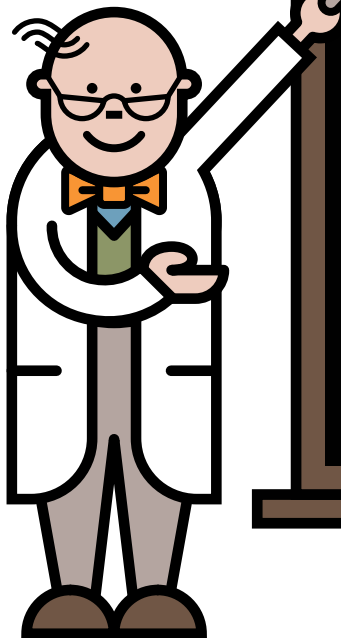
It must be given day and night



Phone your metabolic team
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Do not use sugar-free drinks during illness



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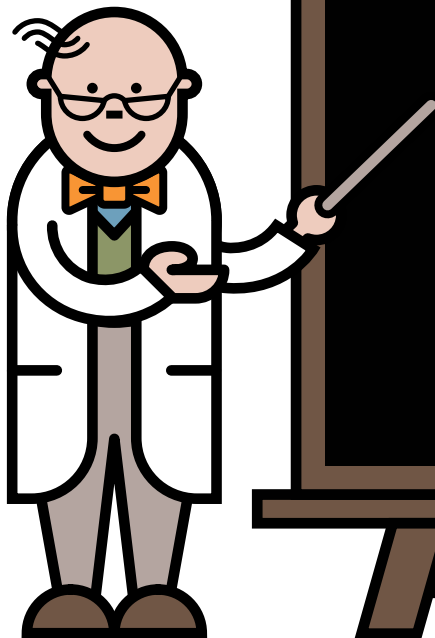
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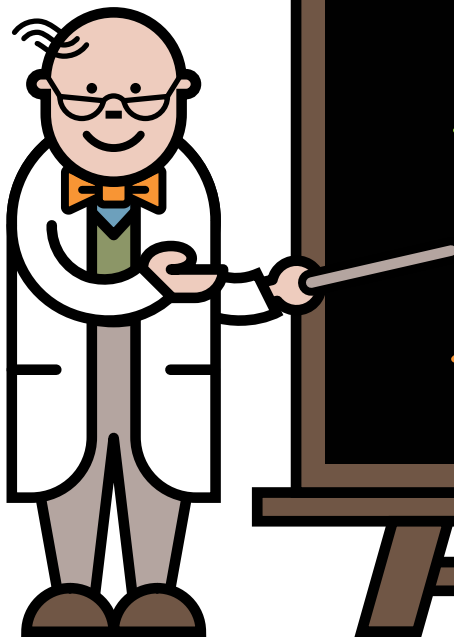
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Checklist for illness

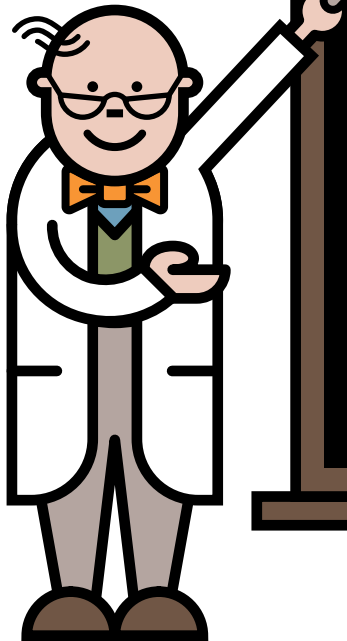
Always take full amounts of emergency feeds as prescribed



If your child vomits or refuses the emergency feeds and/or you are worried, go immediately to the hospital



Phone your metabolic team



Checklist for illness

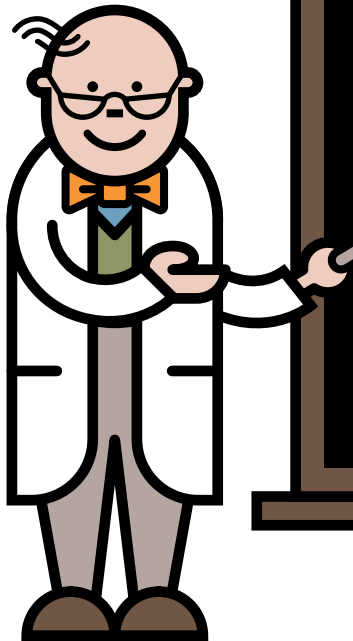
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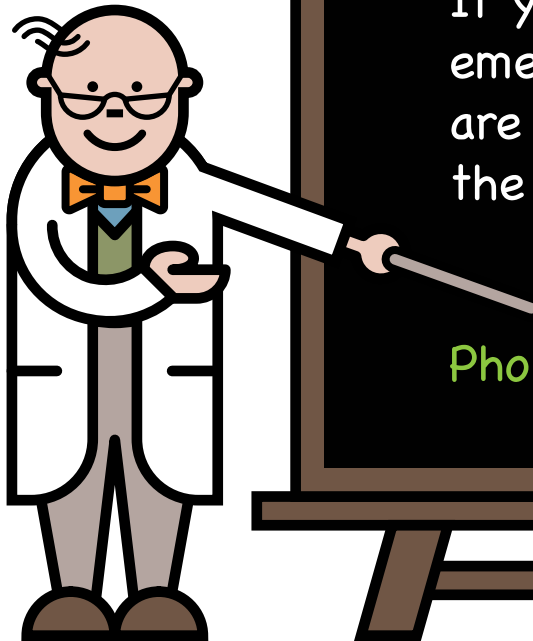
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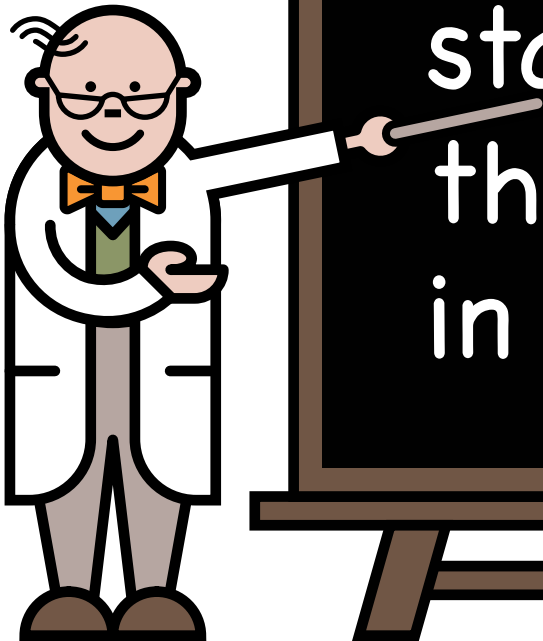


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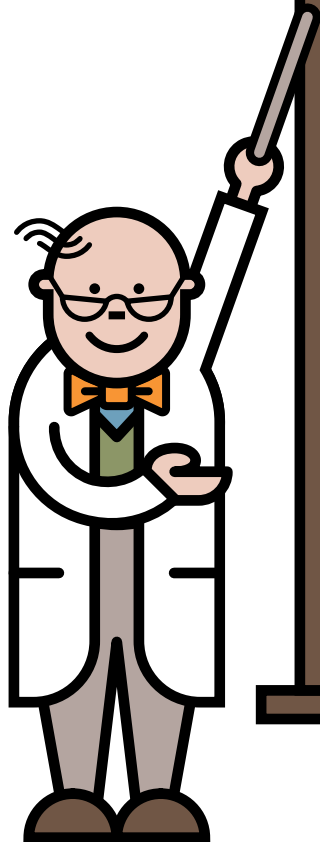


Key message

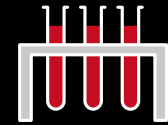
It is imperative that emergency feeds are started **promptly** and there are **no delays** in management.



How is GSD III monitored?



Blood tests to check metabolic control



Safe fasting times are tested by monitoring blood glucose levels and other chemicals in the blood



Height and weight



Medical and development checks

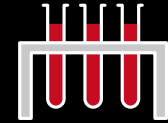


Emergency feeds are adjusted with age and weight



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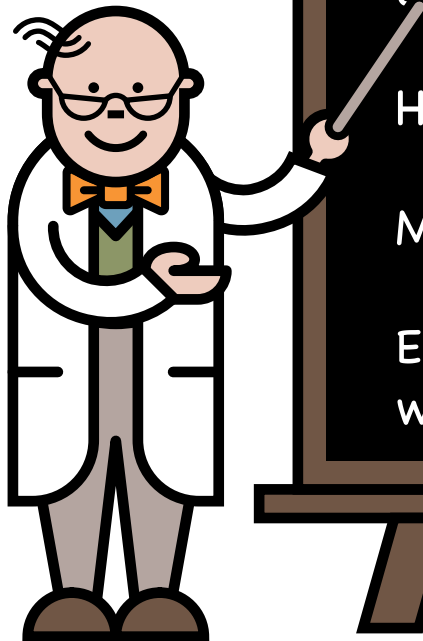
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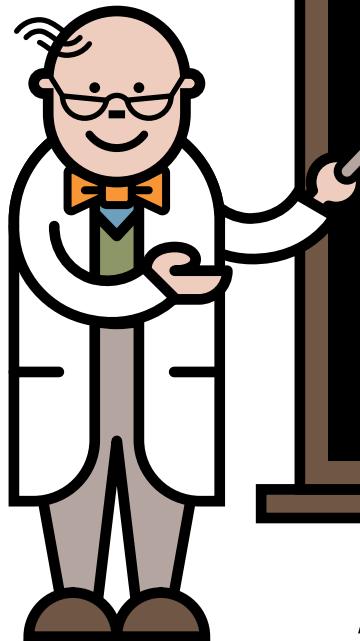
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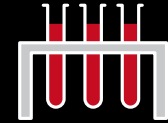
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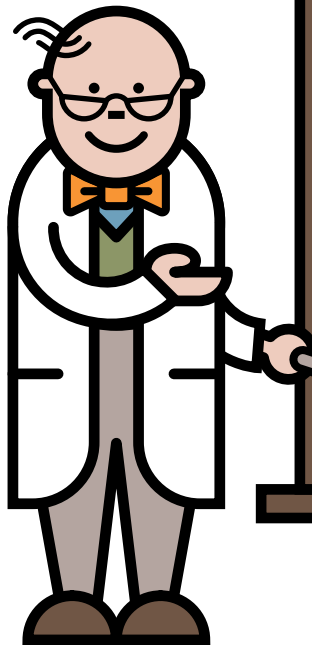
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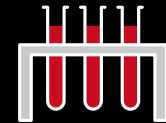
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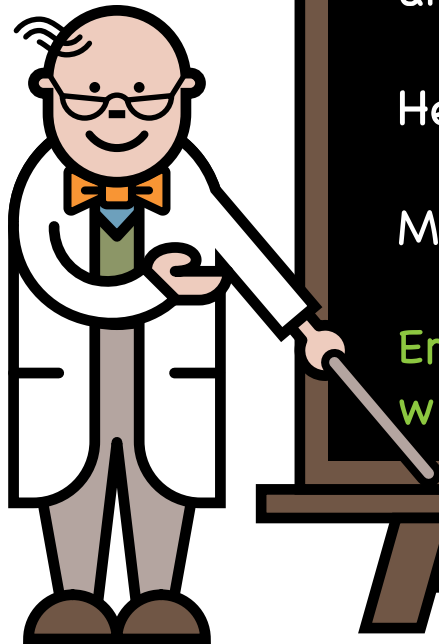
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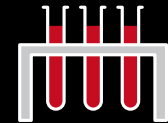
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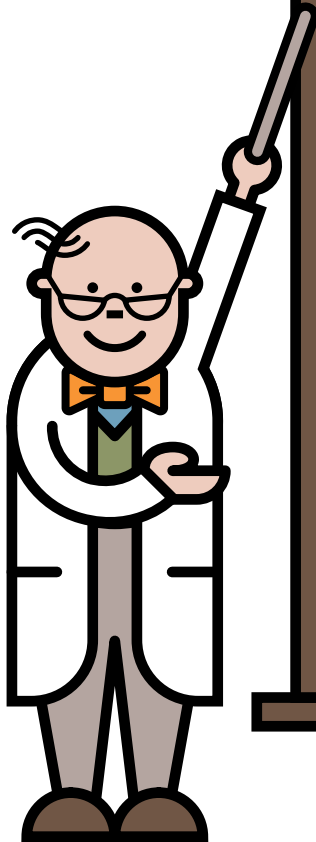
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Emergency feeds are adjusted with age and weight



Chromosomes, genes, mutations



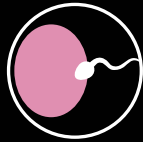
Humans have chromosomes composed of DNA



Genes are pieces of DNA that carry the genetic instructions. Each chromosome may have several thousand genes



The word mutation means a change or error in a genetic instruction

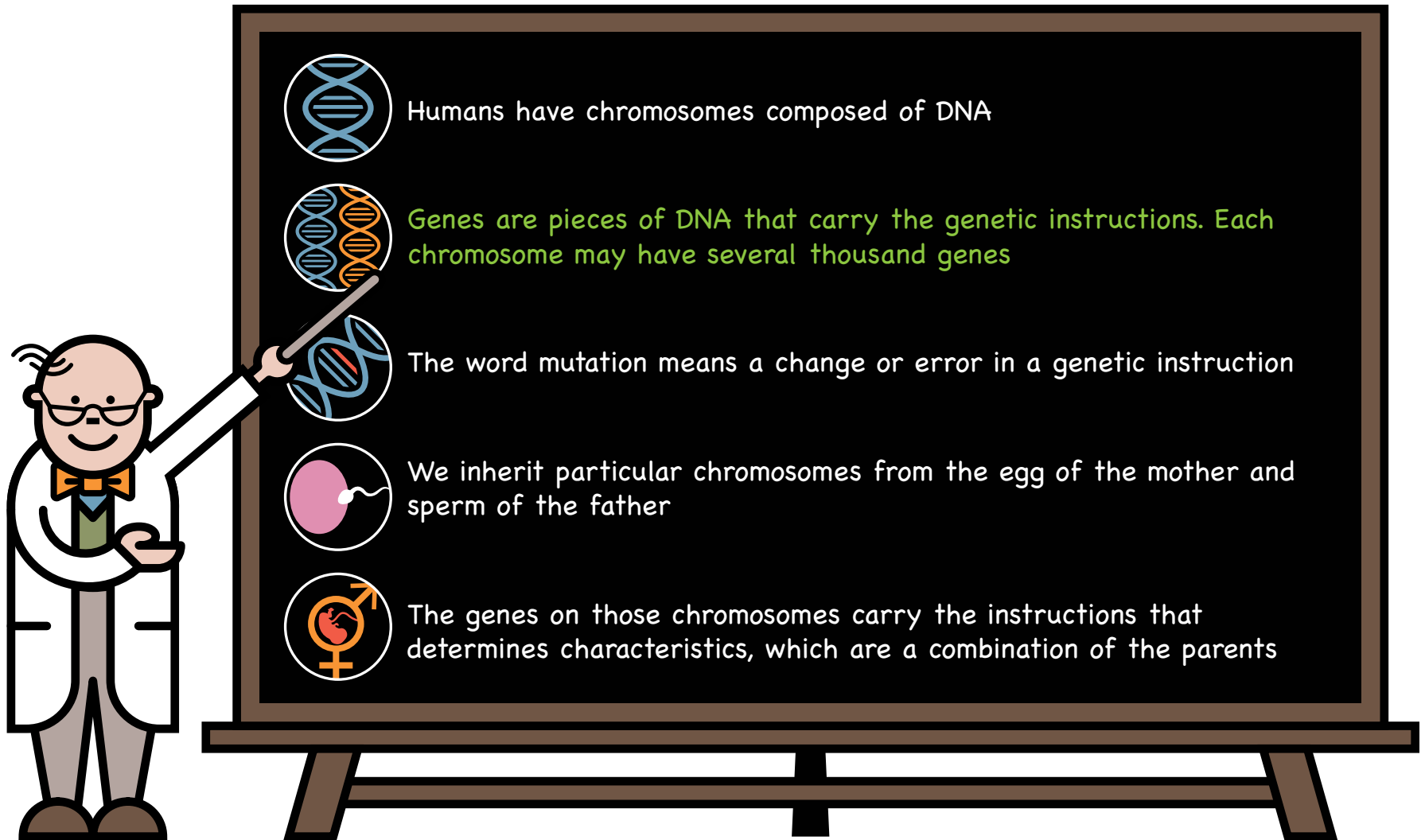



We inherit particular chromosomes from the egg of the mother and sperm of the father





The genes on those chromosomes carry the instructions that determines characteristics, which are a combination of the parents


Chromosomes, genes, mutations




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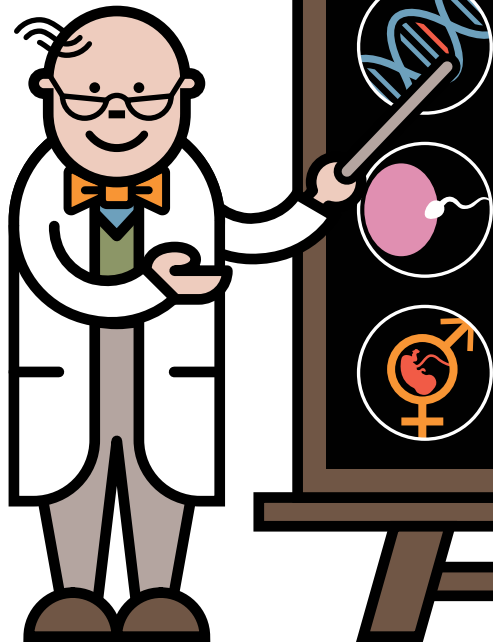
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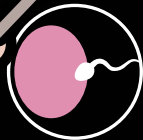
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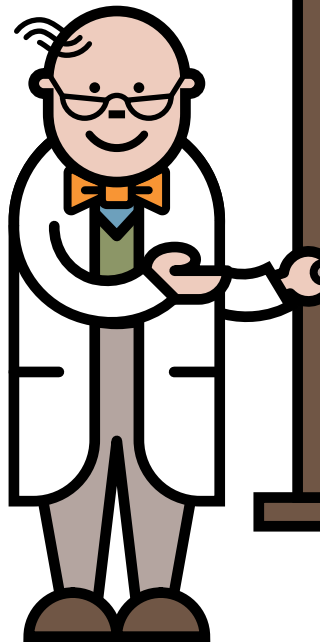


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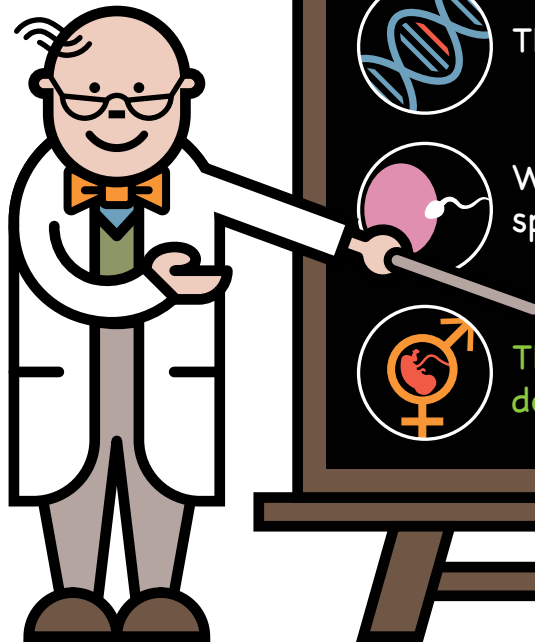


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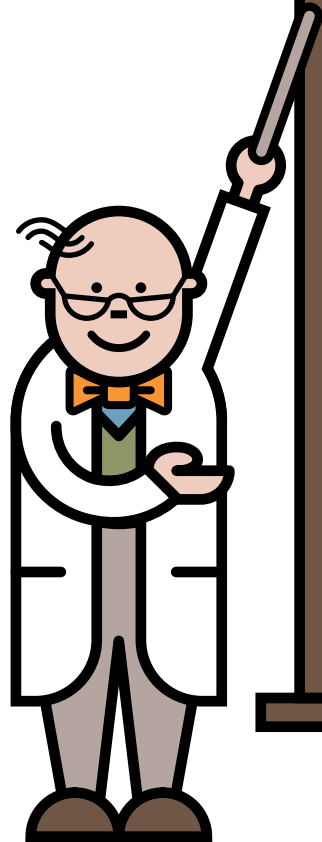


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Inheritance



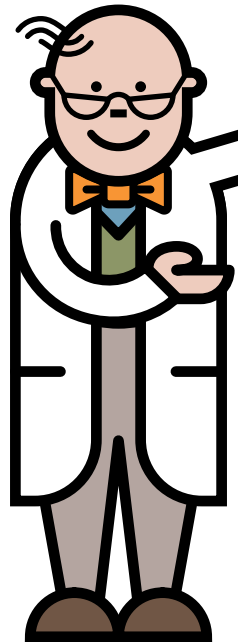
GSD III is an inherited condition. There is nothing that could have been done to prevent your baby from having GSD III

Everyone has a pair of genes that makes the debrancher enzyme. In children with GSD III, neither of these genes work correctly. These children inherit one non-working debrancher enzyme gene from each parent

Parents of children with GSD III are carriers of the condition

Carriers do not have GSD III because the other gene of this pair is working correctly

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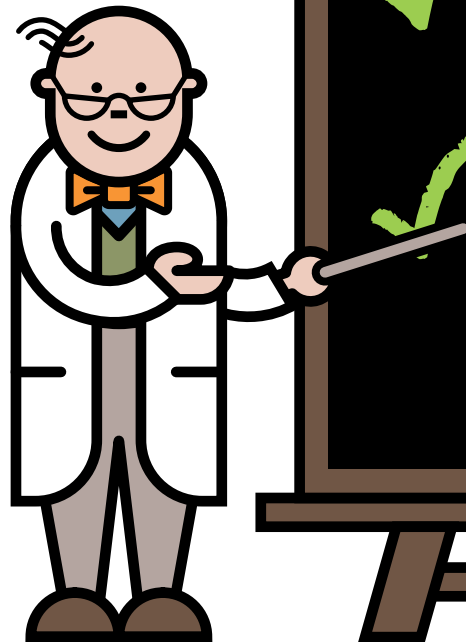


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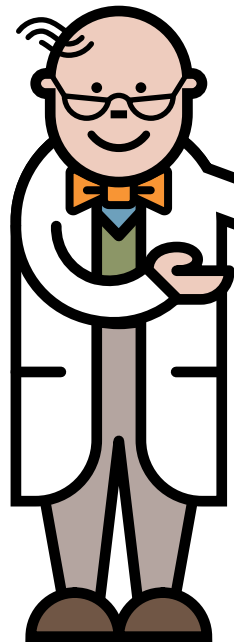
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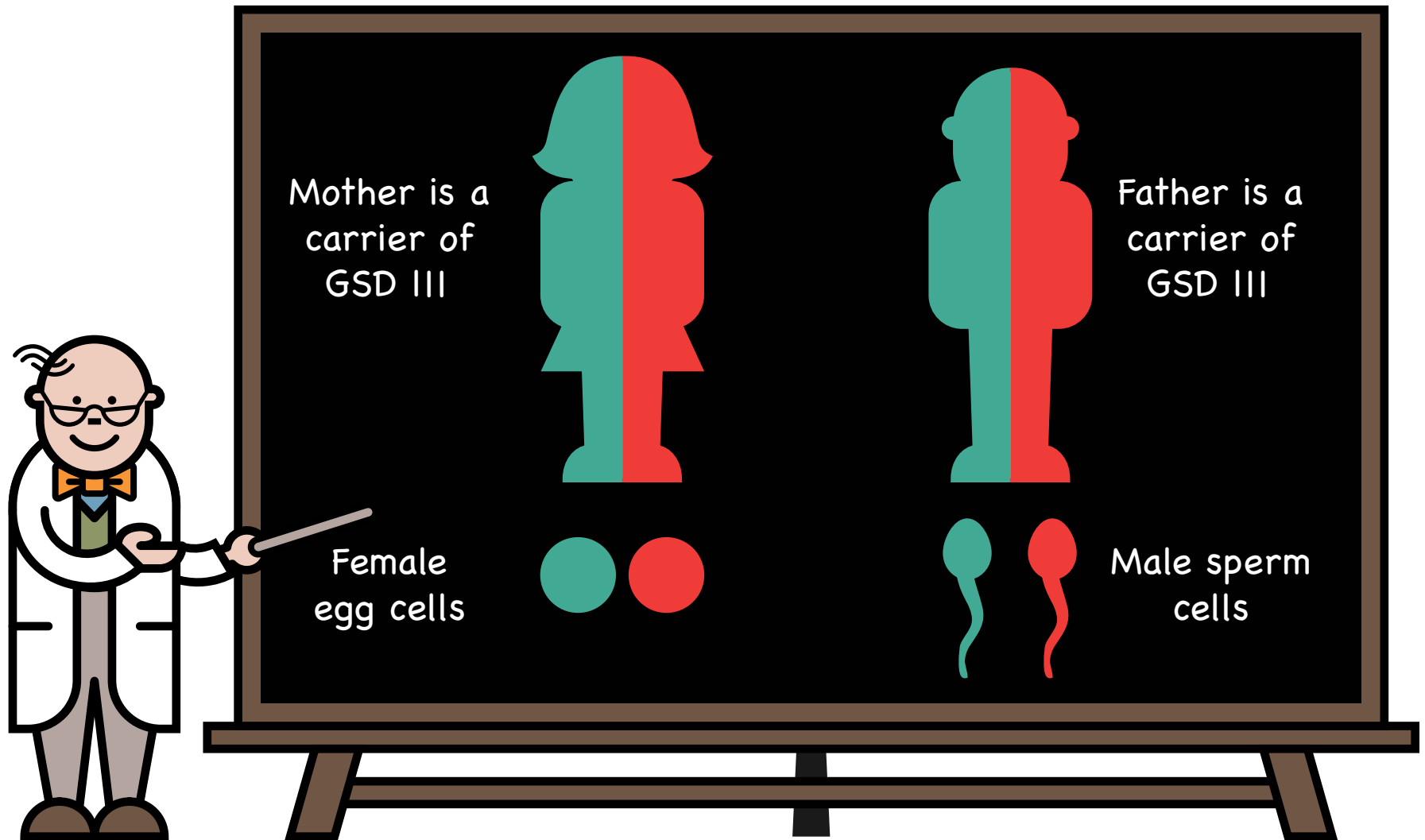


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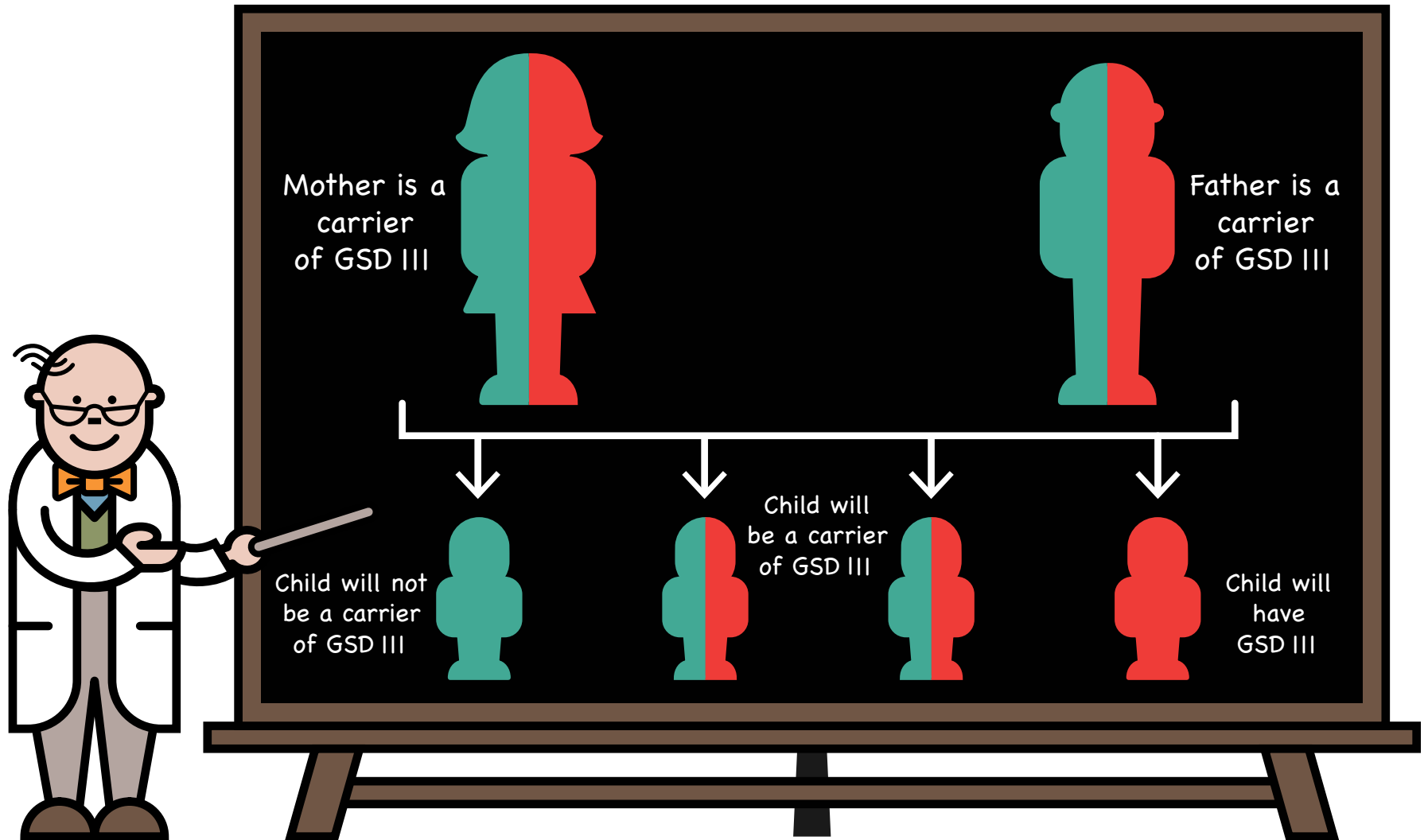


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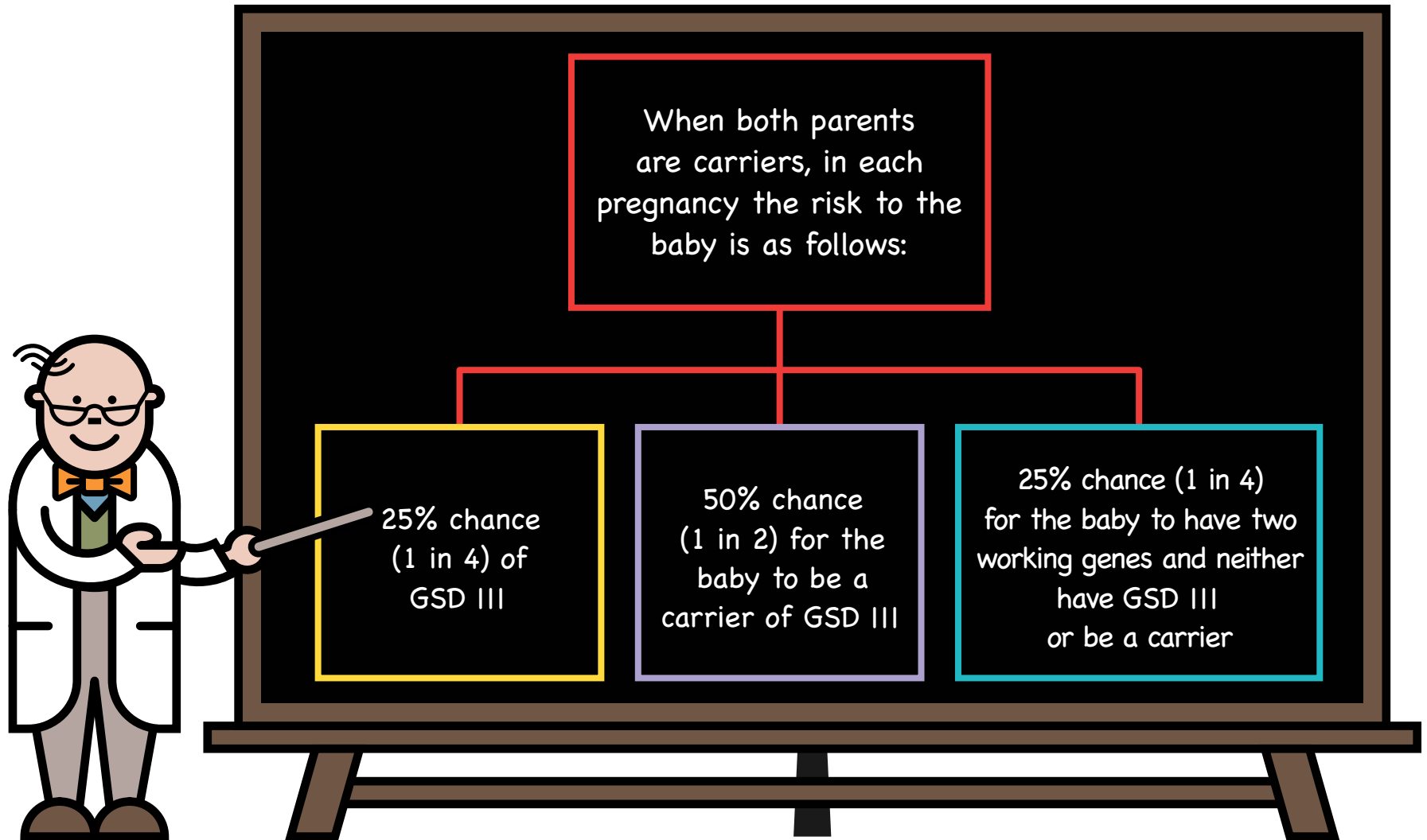
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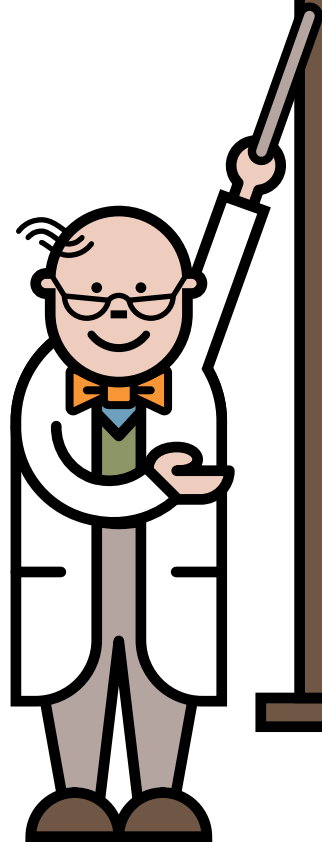
Inheritance



Future pregnancies



Take home messages



GSD III is a serious inherited metabolic condition that causes life threatening symptoms if left unmanaged

Children with GSD III are managed with regular meals and snacks based on carbohydrate and protein, a healthy balanced diet and overnight feeding or regular feeds/drinks overnight

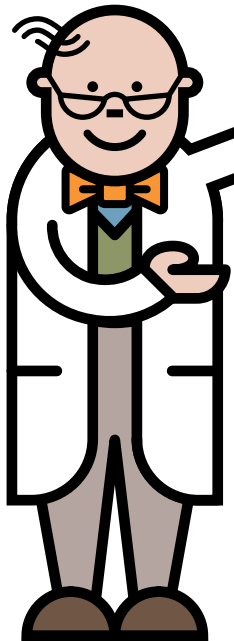
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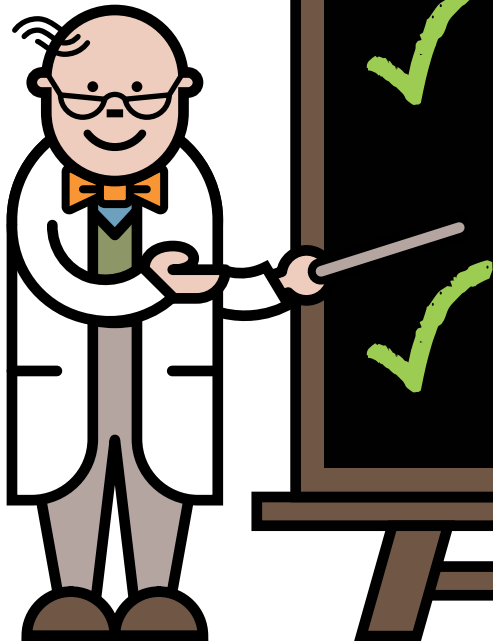


Take home messages

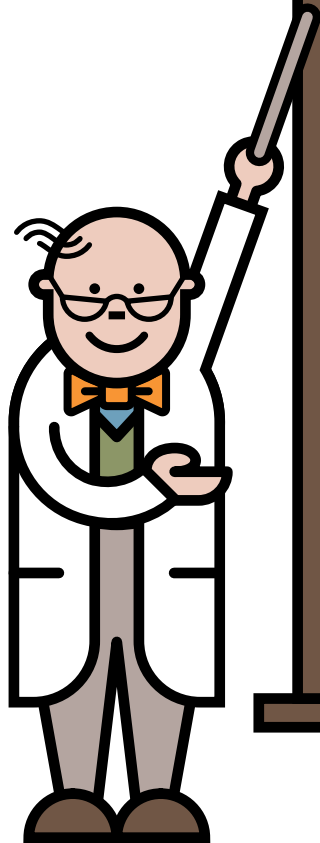
✓ GSD III is a serious inherited metabolic condition that causes life threatening symptoms if left unmanaged

✓ Children with GSD III are managed with regular meals and snacks based on carbohydrate and protein, a healthy balanced diet and overnight feeding or regular feeds/drinks overnight

✓ Remember, during illness, it is imperative that emergency feeds are started promptly, followed strictly and there are no delays



Helpful hints



Always ensure you have a good supply of your dietary products and glucose polymer powder and it is in date

Special feeds and dietary products are prescribed by your GP and you obtain them from your pharmacy or home delivery

Medications to control fever should be given as normally recommended – always keep supplies available

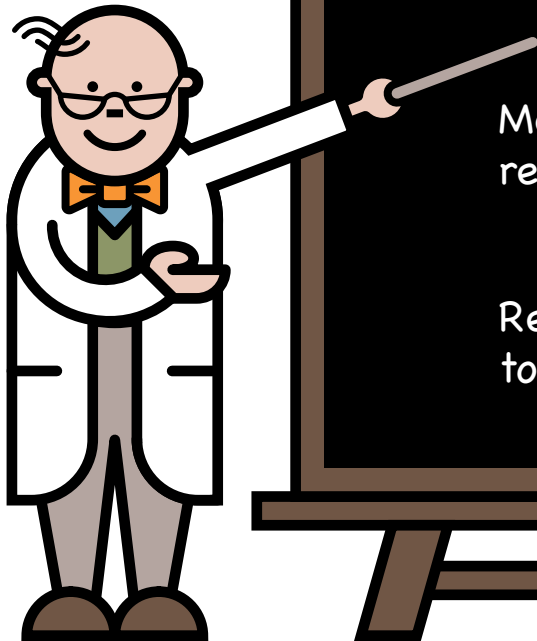
Remember to keep contact numbers of your metabolic team to hand

Helpful hints

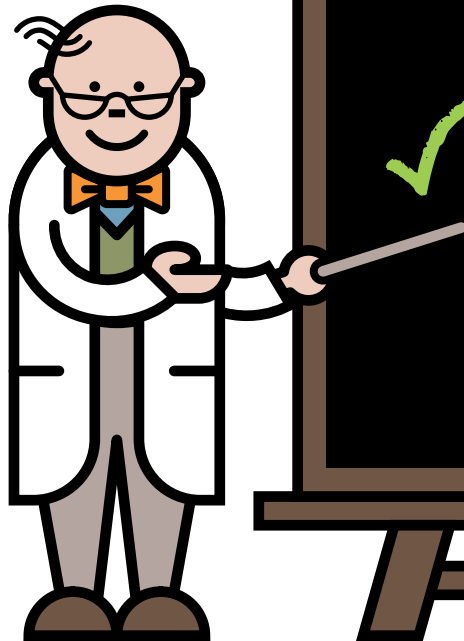
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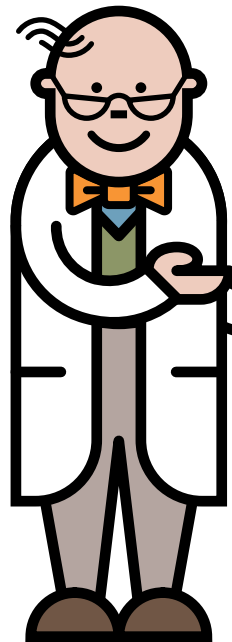
Helpful hints



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- ✓ Medications to control fever should be given as normally recommended – always keep supplies available
- ✓ Remember to keep contact numbers of your metabolic team to hand

Who's who

- My dietitians
- My nurses
- My doctors
 - Contact details, address, photos

BIMDG

British Inherited Metabolic Diseases Group



www.bimdg.org.uk

NUTRICIA

www.nutricia.co.uk

**METABOLIC
SUPPORT UK**

Your rare condition.
Our common fight.

www.metabolicsupportuk.org



www.agsd.org.uk