

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 2, SEPTEMBER 2020

VLCADD

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).

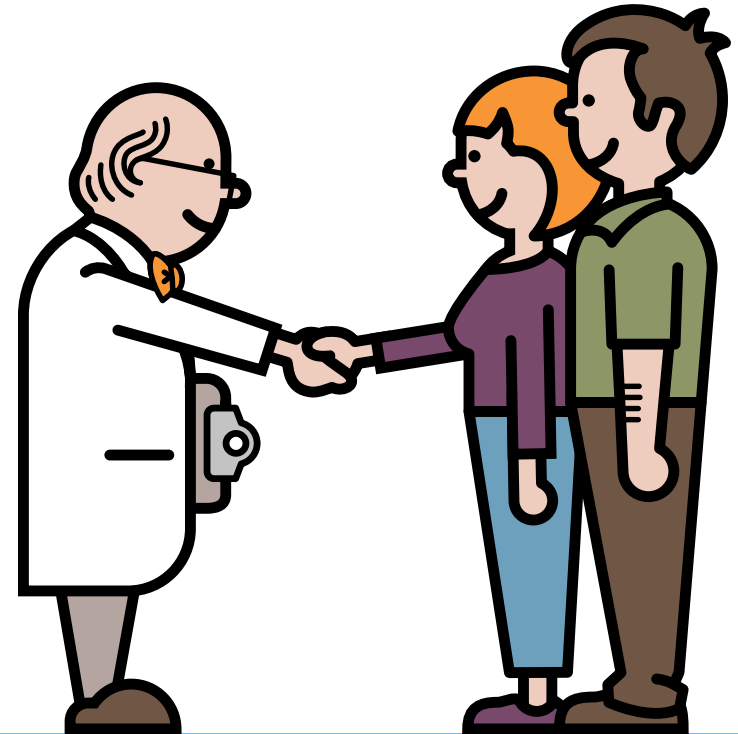
The team are Rachel Skeath, Karen van Wyk, Pat Portnoi and Anita MacDonald. The group is facilitated by Heidi Chan from Nutricia.

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.

VLCAD Deficiency

Information for families
following a new diagnosis



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Tools Enabling Metabolic Parents LEarning

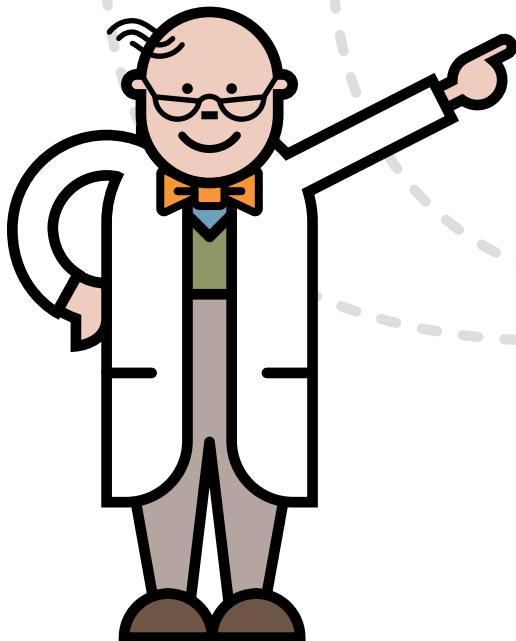
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What is VLCADD?

VLCADD stands for very long chain acyl-CoA dehydrogenase deficiency

It is an inherited metabolic condition

Very Long Chain Acyl-CoA Dehydrogenase

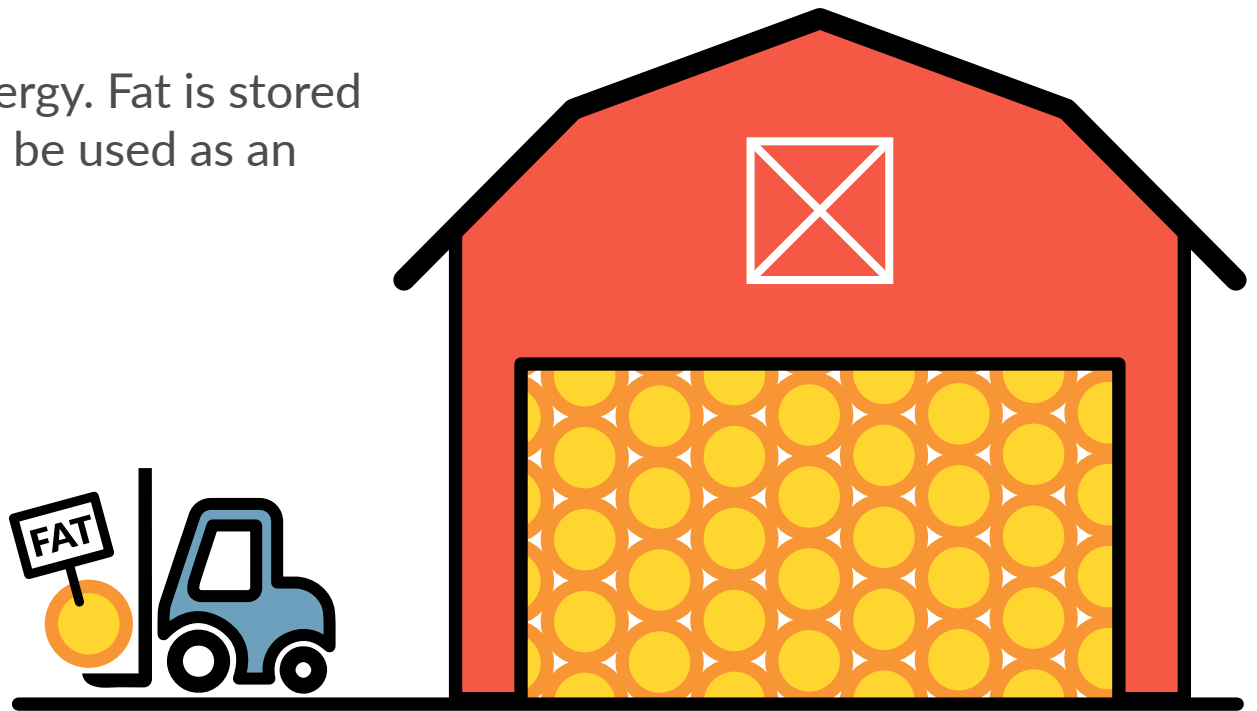


VLCAD
Deficiency

Which foods supply the body with energy?

There are two main food groups that supply the body with energy:

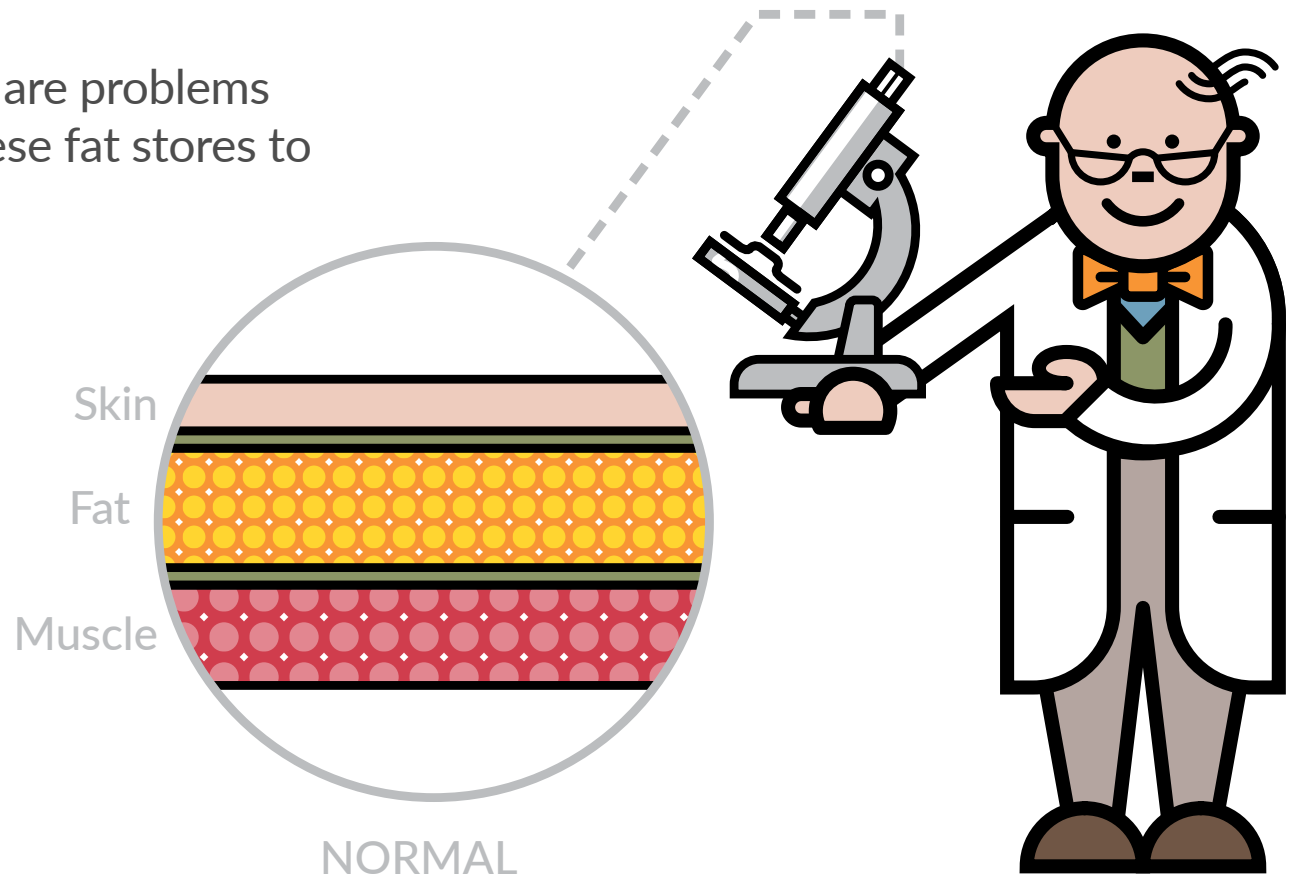
- Carbohydrates (starches and sugars) provide a readily available energy source
- Fats also provide energy. Fat is stored in the body so it can be used as an energy reserve



VLCADD and fat

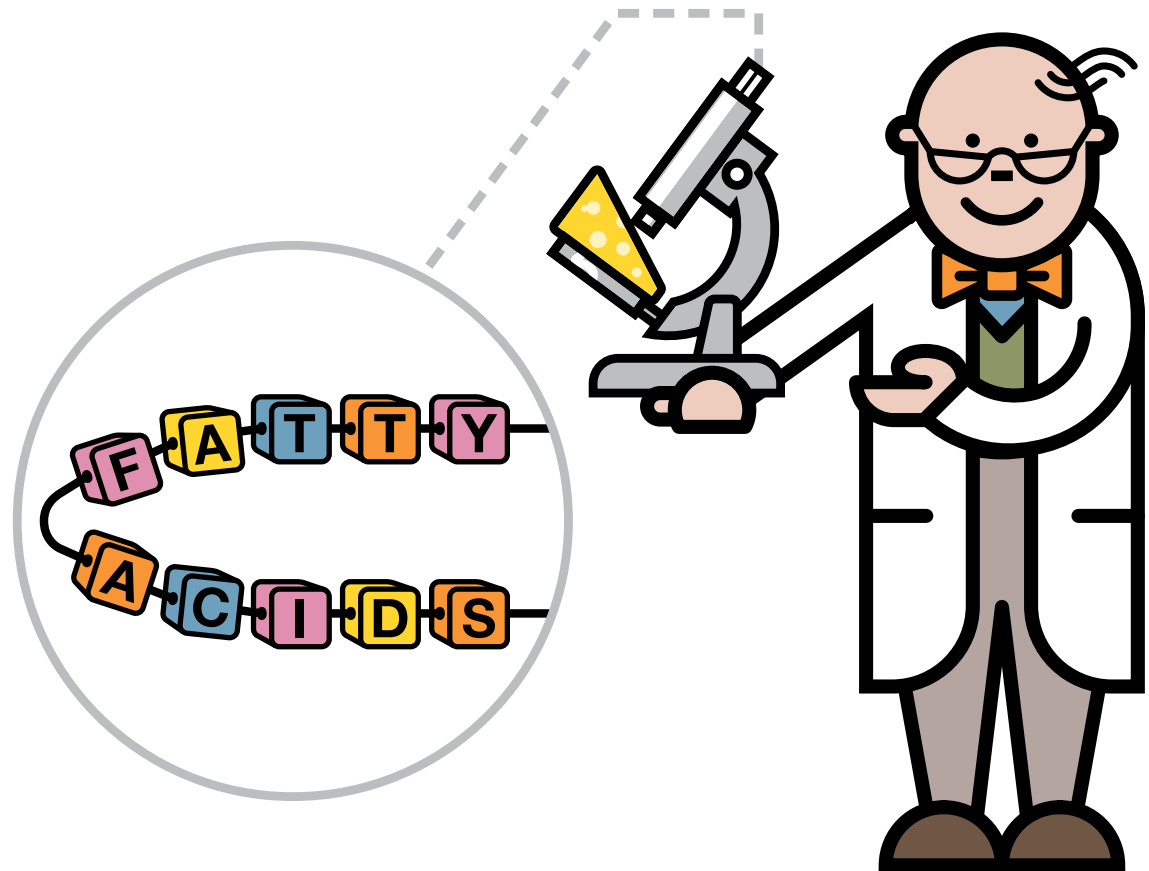
The body uses its own fat stores to provide energy when carbohydrate is depleted.

In VLCADD, there are problems breaking down these fat stores to release energy.



Breaking down fat stores for energy

Body fat stores are broken down into fatty acids.



What are fatty acids?

Fatty acids are made up of carbon atoms joined together to form chains of many different lengths.



Short chain



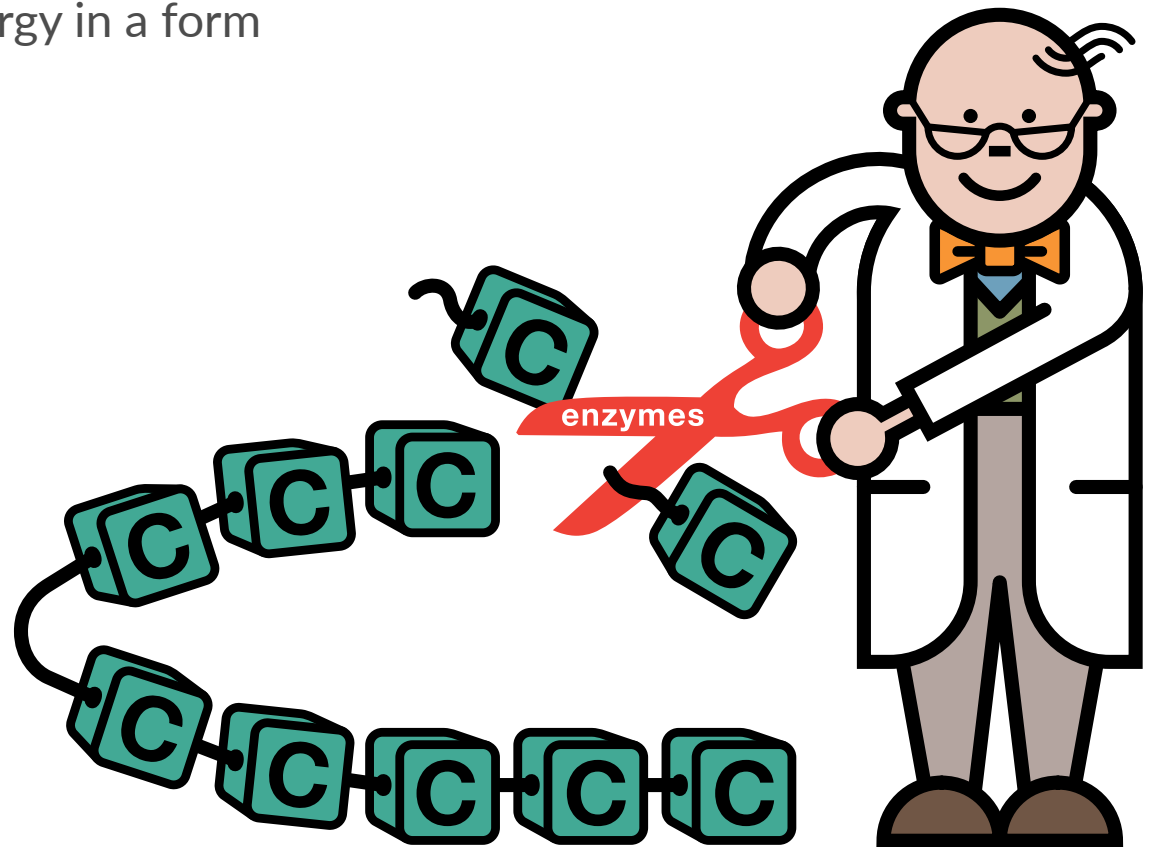
Medium chain



Long chain

Fatty acids and enzymes

Fatty acid chains are then broken down into smaller units by enzymes (which are like chemical scissors). This enables the body to produce energy in a form which it can use.



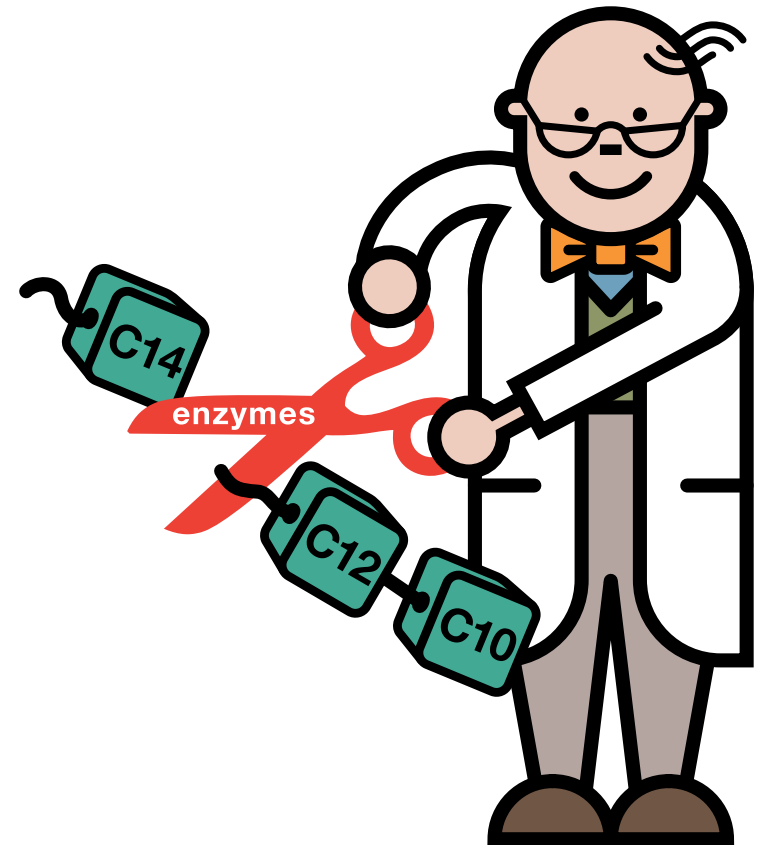
What happens in VLCADD?

In VLCADD, the body lacks an enzyme that helps convert fat stores into energy.

This **stops** the very long chain fats from being used for energy.

There is a problem when it is necessary to break down fats quickly.

This occurs when there is a shortage of energy supply.



What can go wrong in VLCADD?

There can be a shortage of energy supply and a build up of harmful chemicals with illness or lack of food.

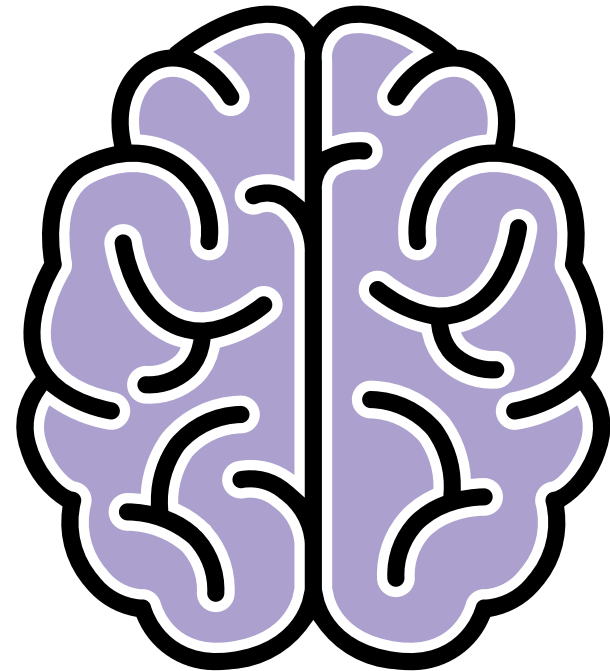
Symptoms include:

- poor feeding
- excessively sleepy
- rapid breathing
- seizures
- low blood sugar
- floppiness
- enlarged heart, irregular heartbeat

What can go wrong in VLCADD?

If there is a shortage of energy and this is not corrected, it can lead to coma and brain damage and it may be life threatening.

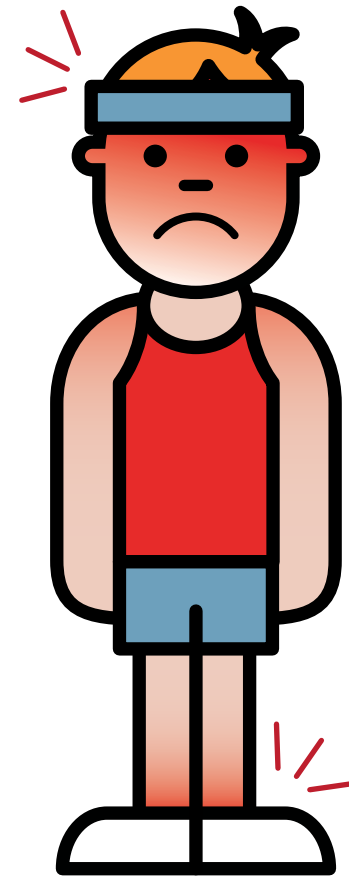
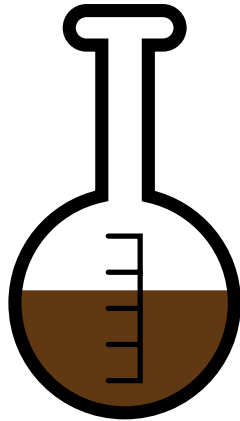
However, please remember, this can all be prevented with timely and correct management.



Some children may be diagnosed at a later stage

Signs and symptoms:

- muscle pain especially with exercise
- reddish-brown coloured urine



Metabolic crisis

- A **metabolic crisis** triggers the VLCADD symptoms
- This leads to a lack of energy and build up of toxic chemicals
- It is usually triggered by childhood infections causing high temperatures, vomiting, and diarrhoea
- It can also be caused by going a very long time without food
- Avoidance of a metabolic crisis is essential



How is VLCADD diagnosed?

VLCADD is suspected because of the pattern of chemicals (acylcarnitines and organic acids) found in the blood and urine.

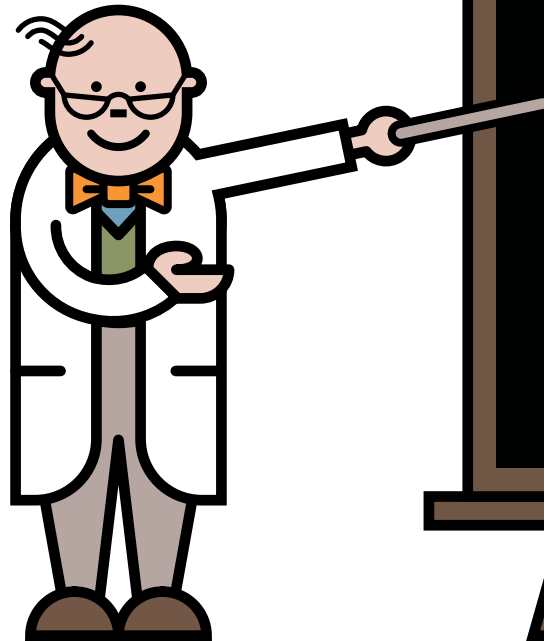
The diagnosis is confirmed by finding mutations in the VLCAD gene.



How is VLCADD managed?

In babies, it is managed day to day by **avoiding** long periods without feeding, even when well.

The length of time babies can go without feeds is known as **the safe fasting time**.



SAFE FASTING TIMES

- The safe fasting time varies for each baby with VLCADD. Your metabolic team will advise.
- It is important they receive regular feeds during the day and at night.
- They should not miss scheduled feeds.

How is VLCADD managed day to day?

Special infant feed containing a fat called medium chain triglycerides (MCT)



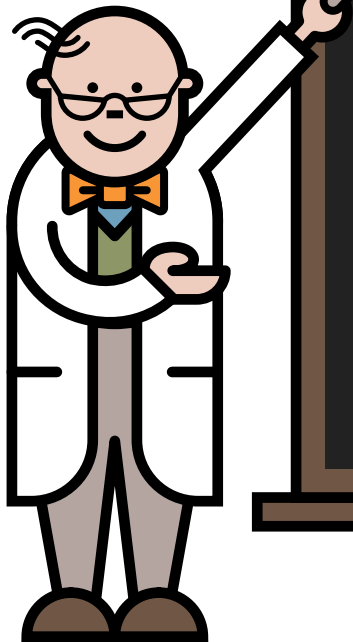
Very low fat diet



MCT oil for cooking or MCT as a supplement



Regular mealtimes as recommended by your metabolic team



How is VLCADD managed day to day?

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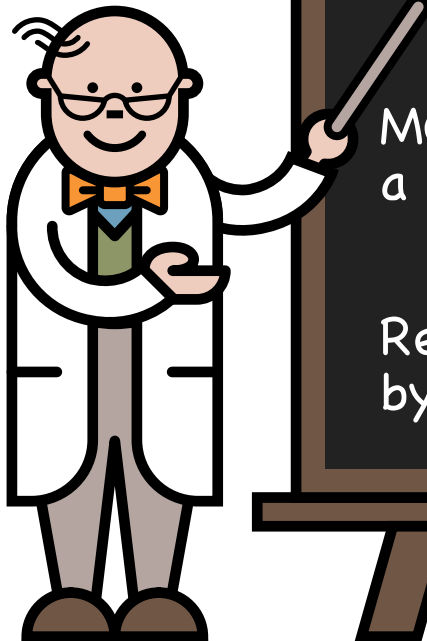
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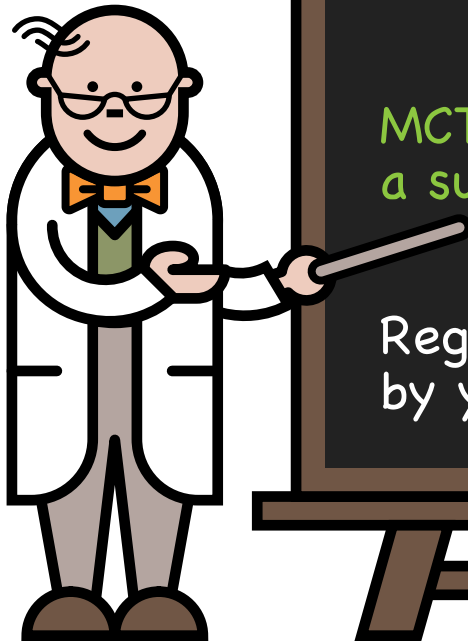
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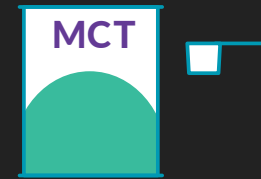


Regular mealtimes as recommended by your metabolic team



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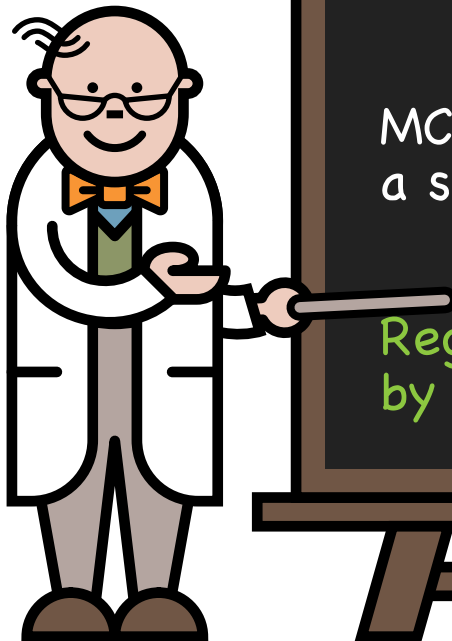
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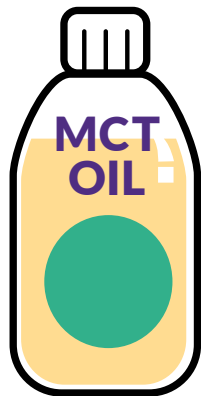


VLCADD and fat

The diet needs to be very low in fat.

Foods high in fat are avoided.

Many foods are high in fat e.g. full fat milk, full fat cheese, fatty meat, eggs, ice cream, chips, crisps and chocolate.



Very low fat foods

The diet needs to be very low in fat.

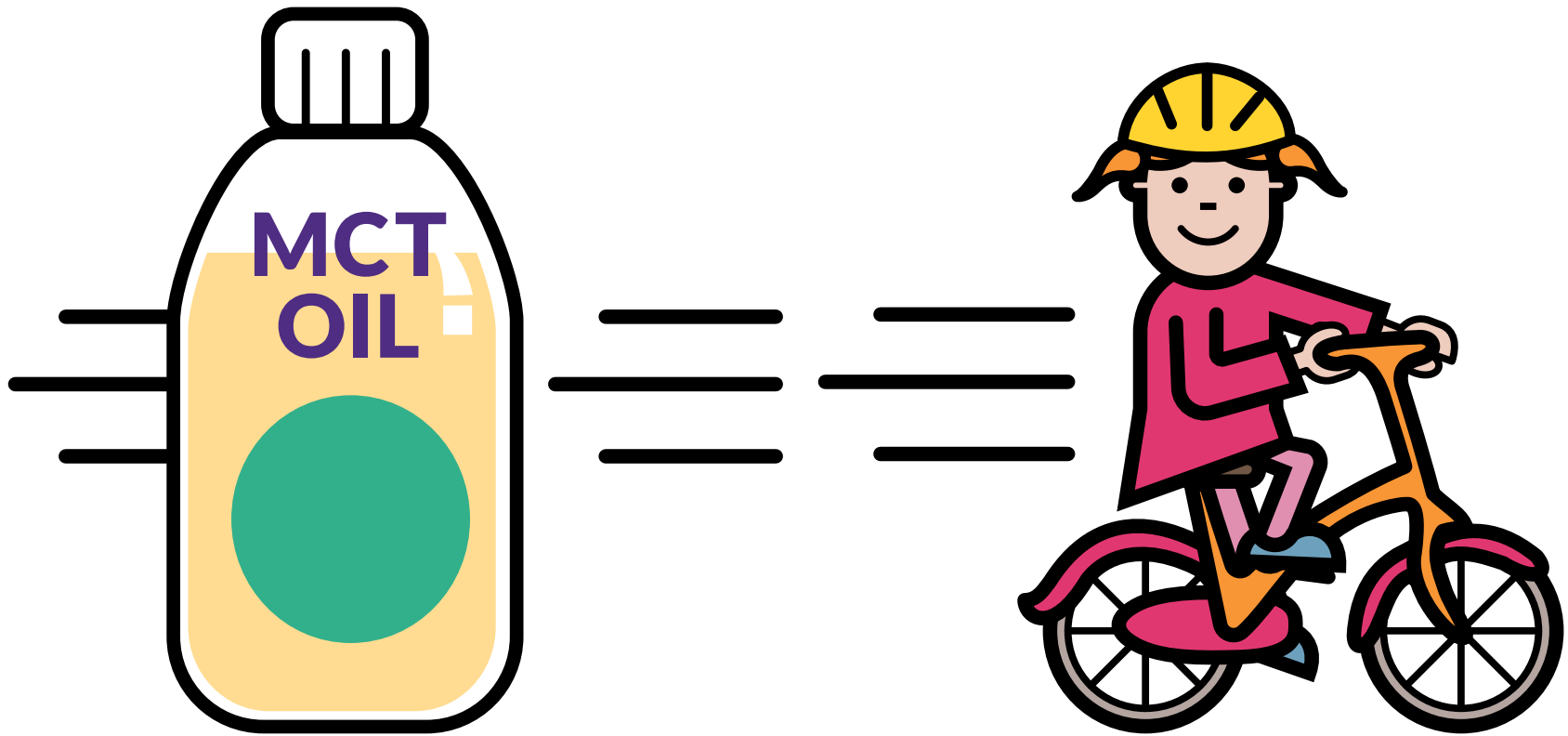
Very low fat foods are permitted.

There is a wide range of very low fat foods available e.g. skimmed milk, very low fat yoghurt/cheese, white fish, fruit and vegetables.



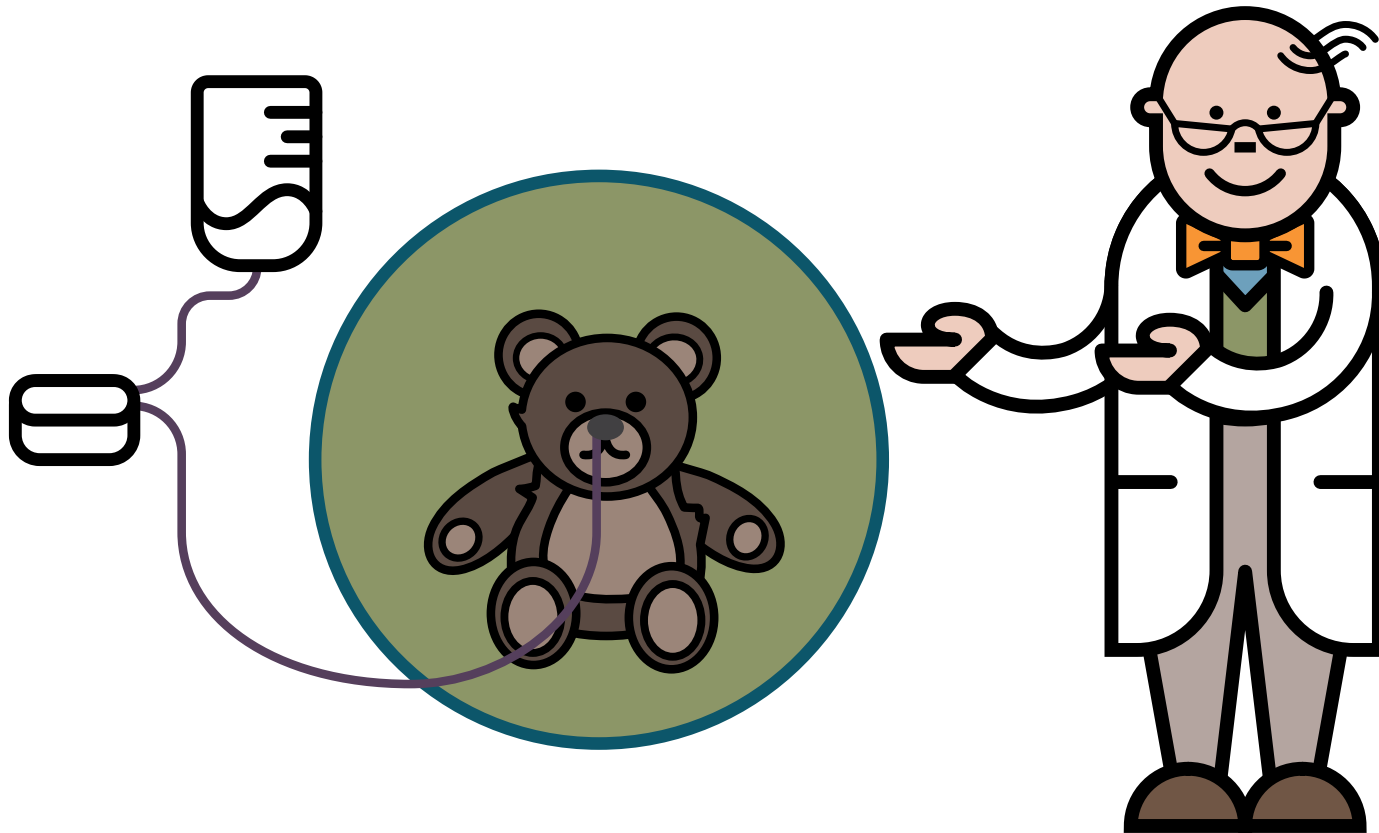
VLCADD and exercise

It is advisable to take a source of MCT, or a high sugar snack or drink pre exercise. This will supply an extra source of energy.



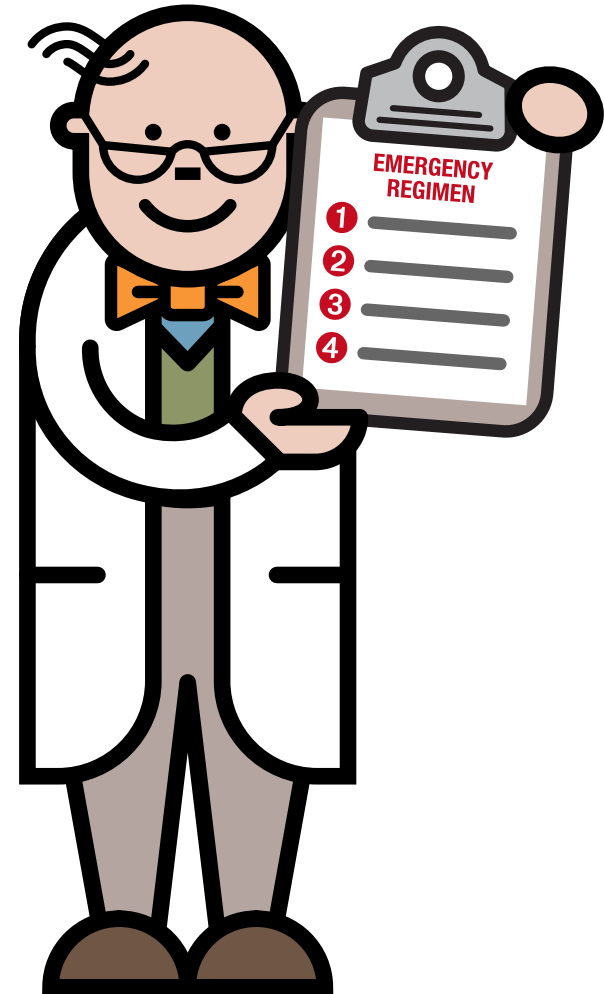
Is tube feeding needed?

In babies with the most severe forms of VLCADD, tube feeding may be necessary. This will ensure energy, nutrient and fluid needs are met.



How is VLCADD managed during illness?

- During any childhood illness, an emergency regimen is given
- This provides energy and prevents build up of harmful chemicals that cause a metabolic crisis



How is VLCADD managed during illness?

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



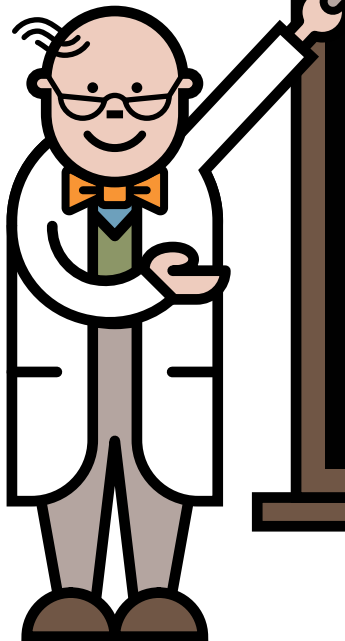
This must be given regularly
day and night



Phone your metabolic team
for help if your child is unwell



Do not use sugar-free drinks during illness



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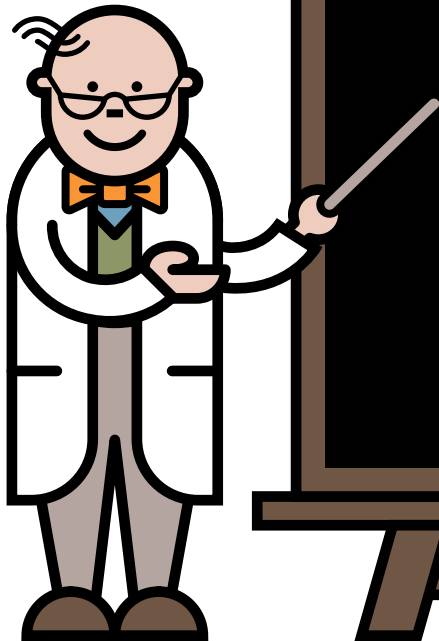
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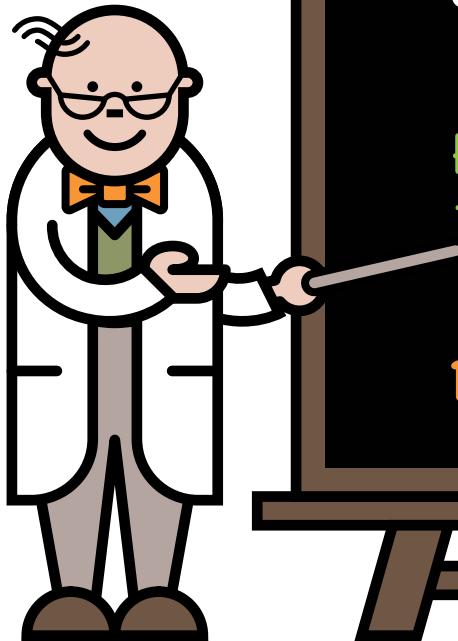
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Checklist for illness

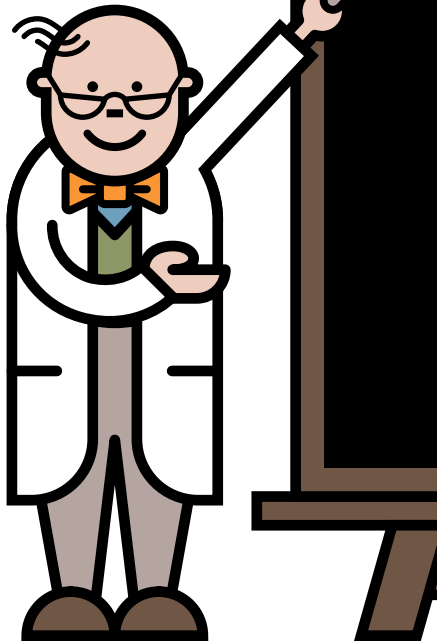
Always take full amounts of emergency feeds as prescribed



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



Phone your metabolic team



Checklist for illness

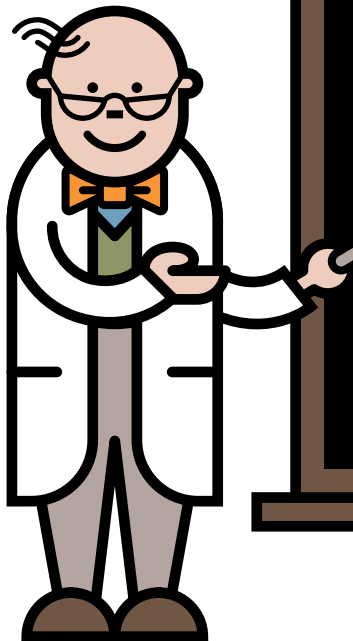
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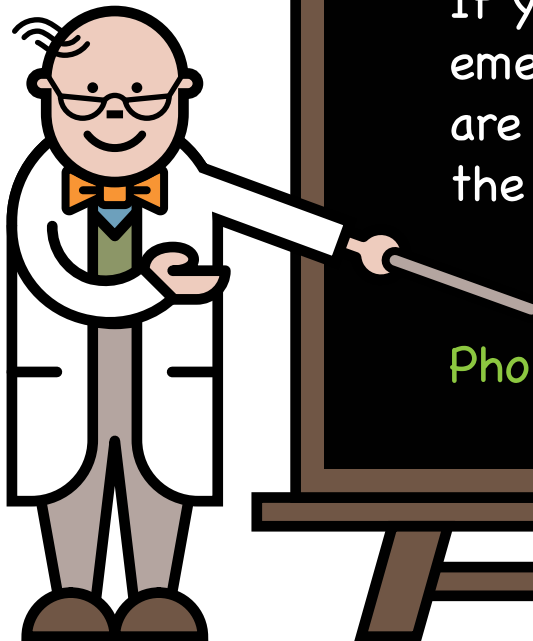
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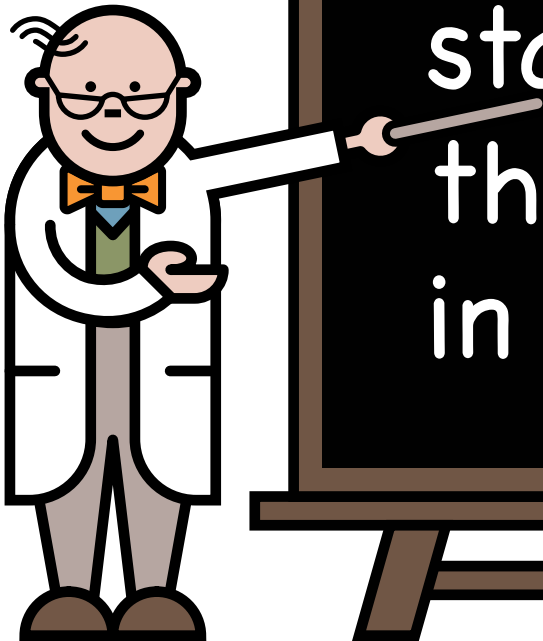


Phone your metabolic team

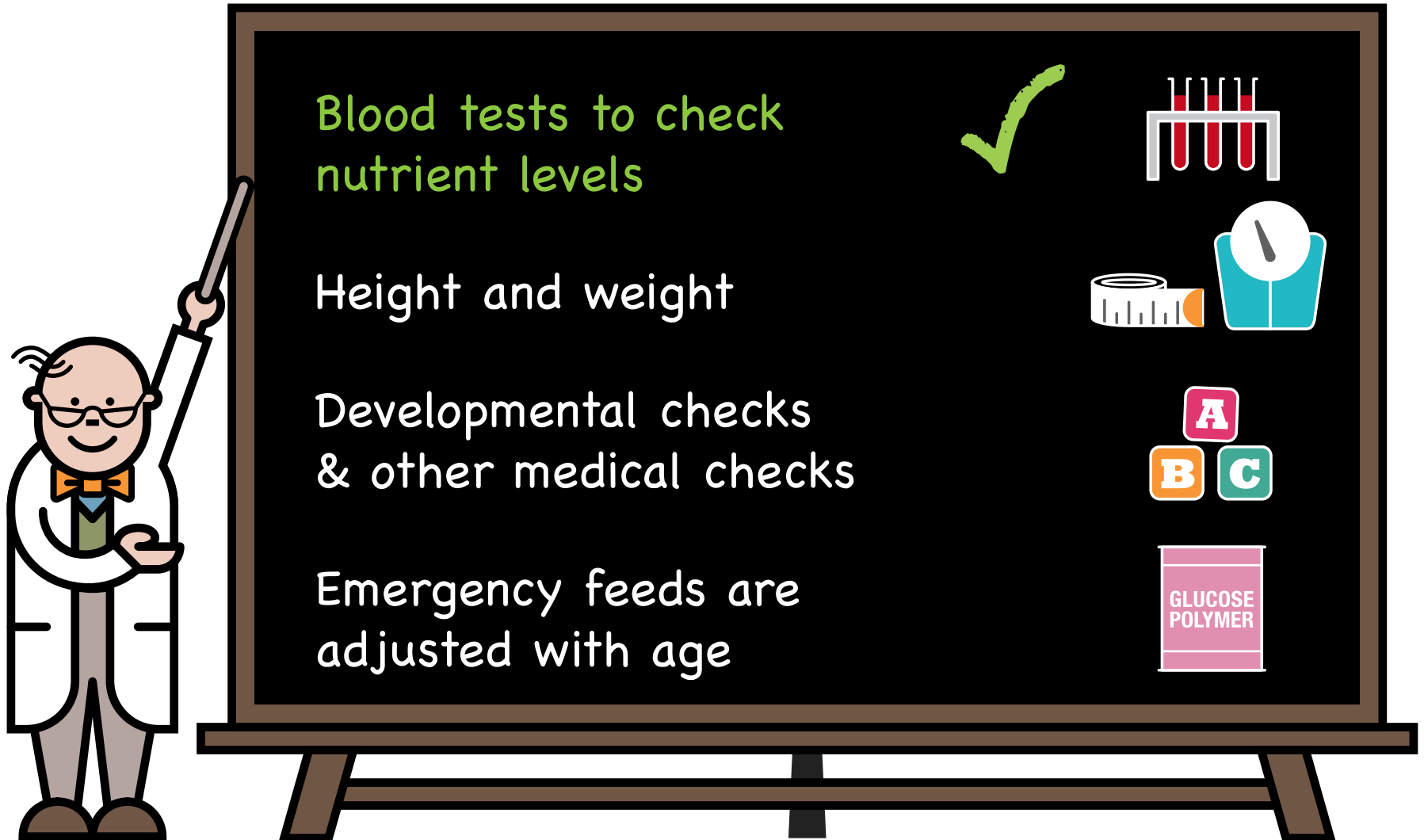


Key message

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



How is VLCADD monitored?



Blood tests to check nutrient levels

Height and weight

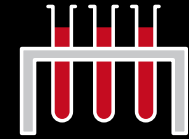
Developmental checks & other medical checks

Emergency feeds are adjusted with age

GLUCOSE POLYMER

How is VLCADD monitored?

Blood tests to check
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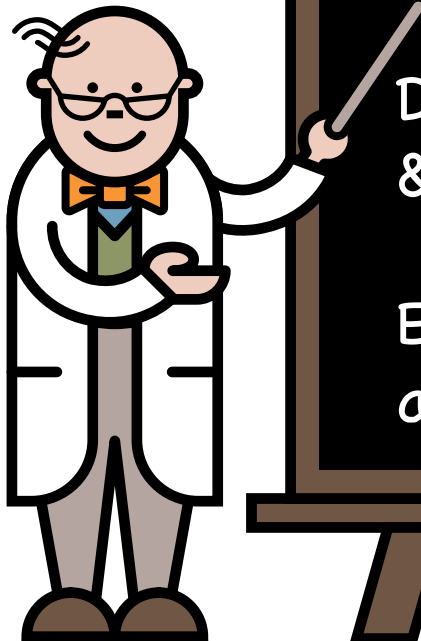
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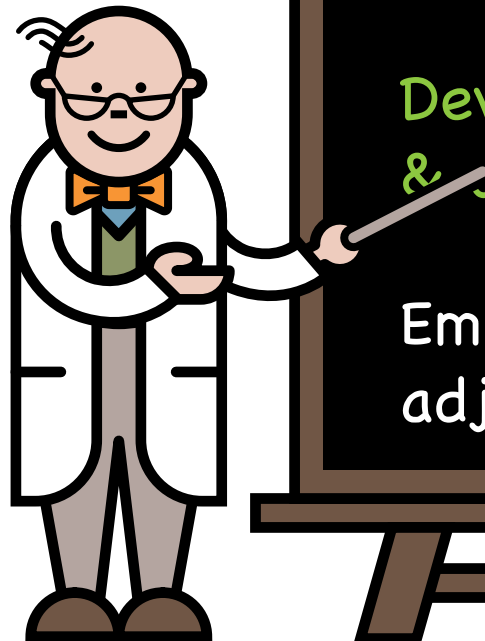
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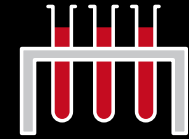
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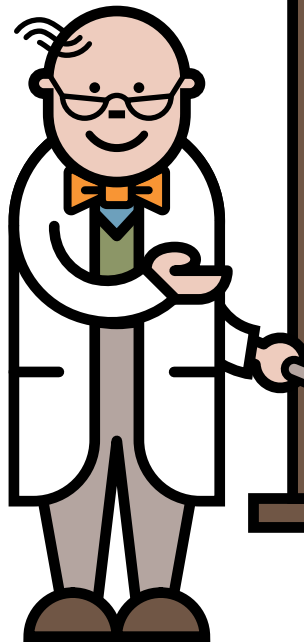
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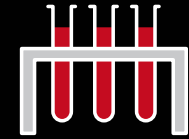
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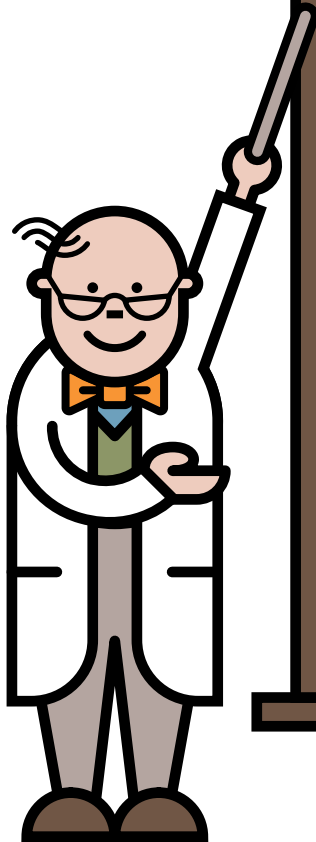
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Emergency feeds are
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Chromosomes, genes, mutations



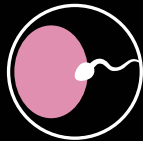
Humans have chromosomes composed of DNA



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

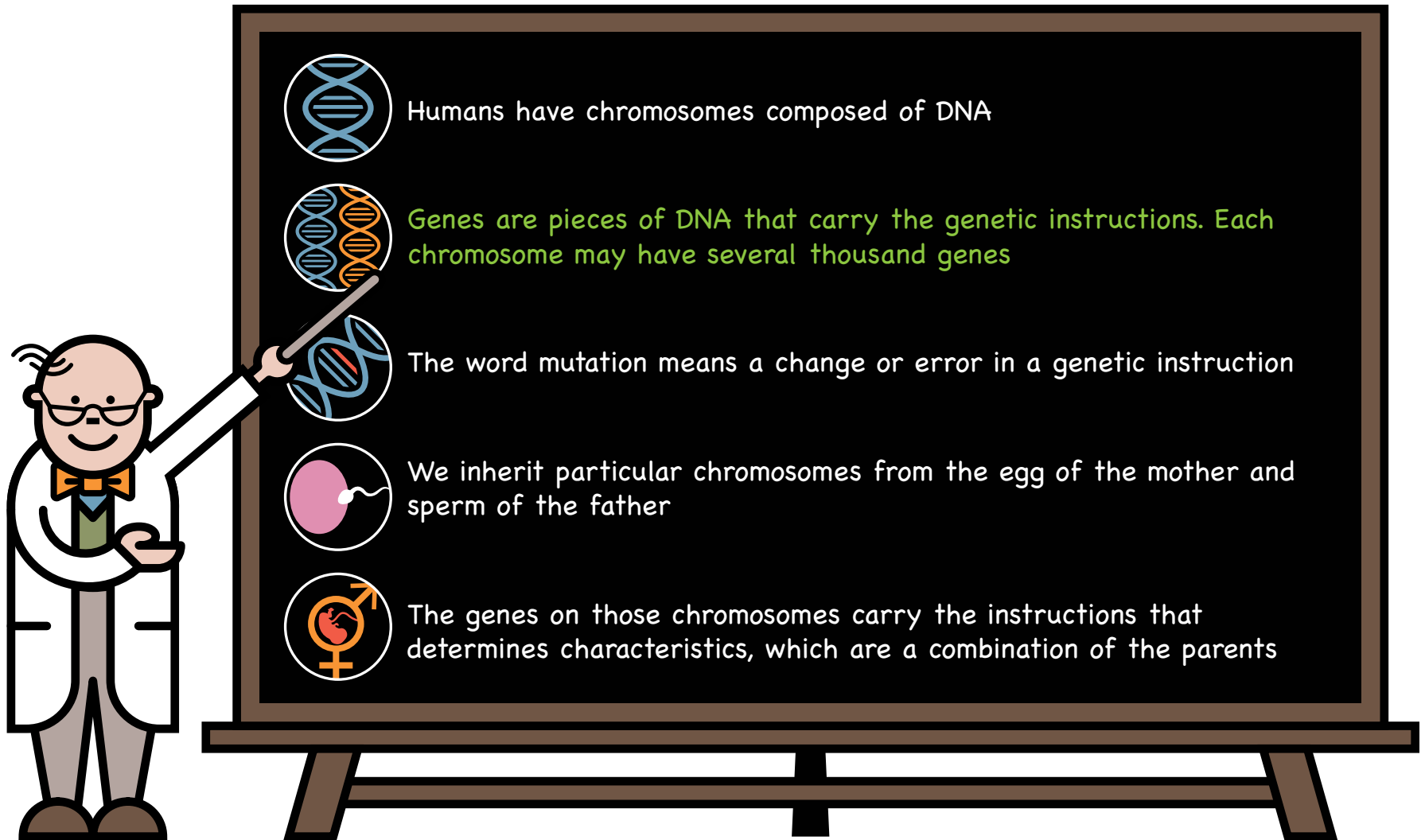



We inherit particular chromosomes from the egg of the mother and sperm of the father





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


Chromosomes, genes, mutations




 Humans have chromosomes composed of DNA

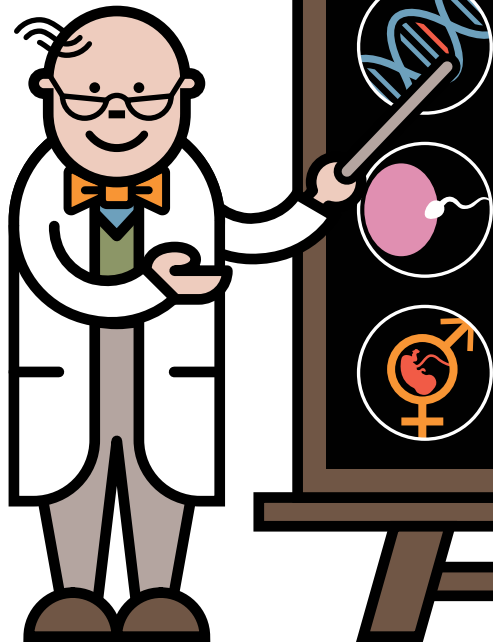
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Chromosomes, genes, mutations



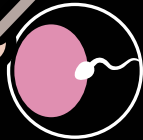
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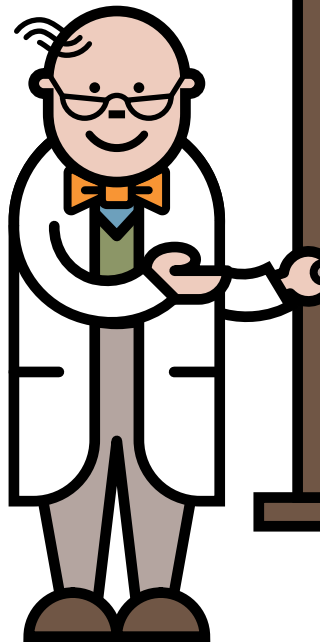


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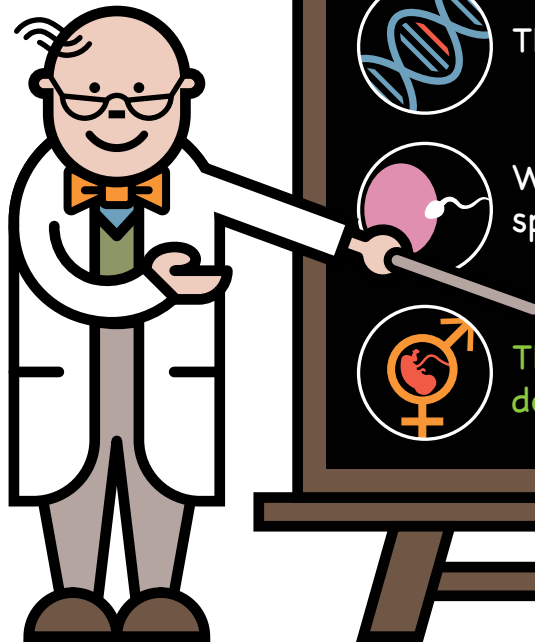


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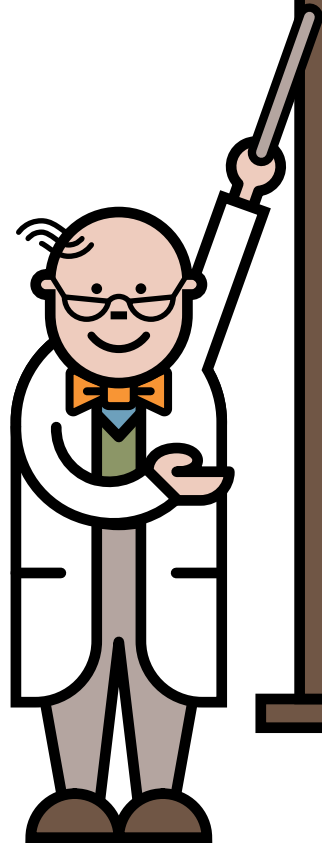


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Inheritance



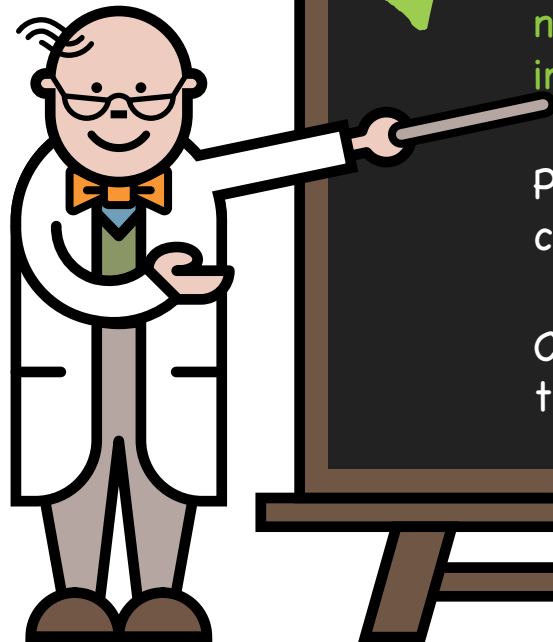
VLCADD is an inherited condition. There is nothing that could have been done to prevent your baby from having VLCADD

Everyone has a pair of genes that make the very long chain acyl-CoA dehydrogenase enzyme. In children with VLCADD, neither of these genes works correctly. These children inherit one non-working VLCAD gene from each parent

Parents of children with VLCADD are carriers of the condition

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

Inheritance



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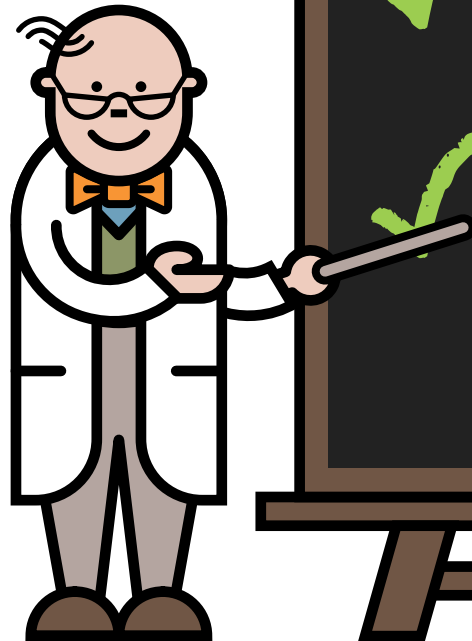


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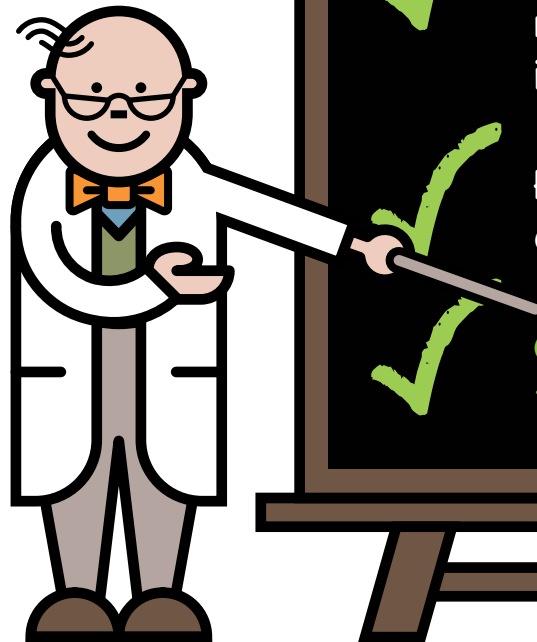
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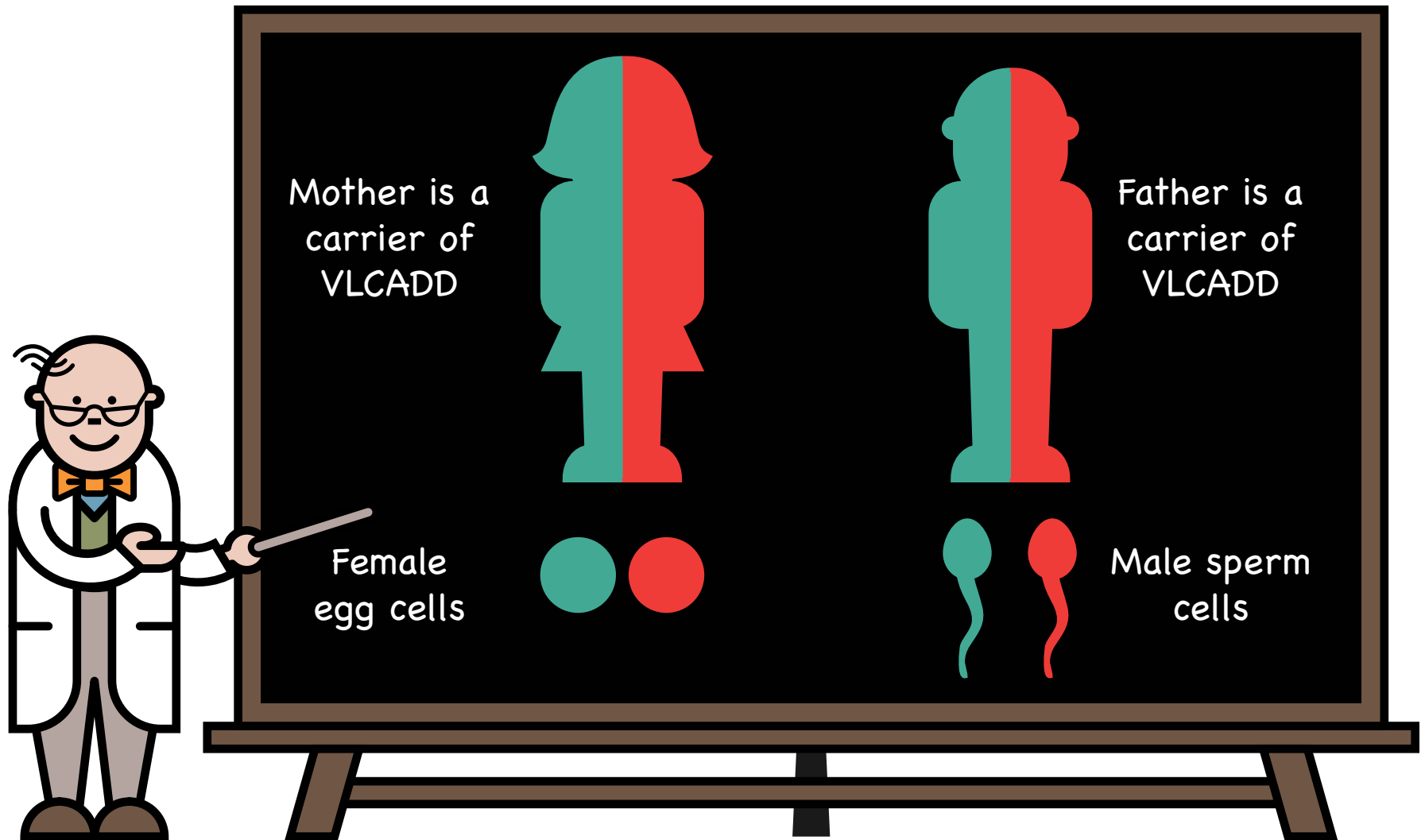
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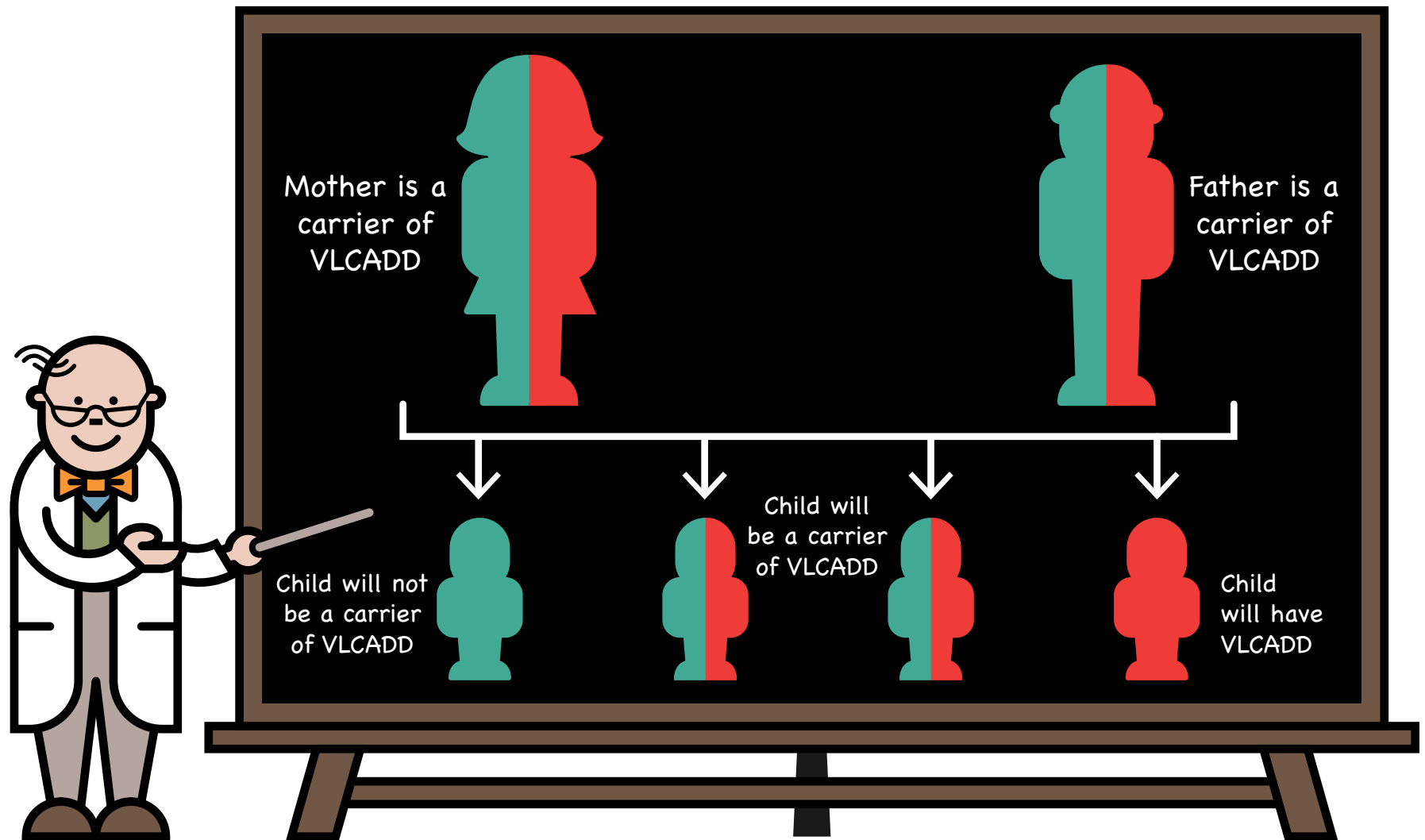
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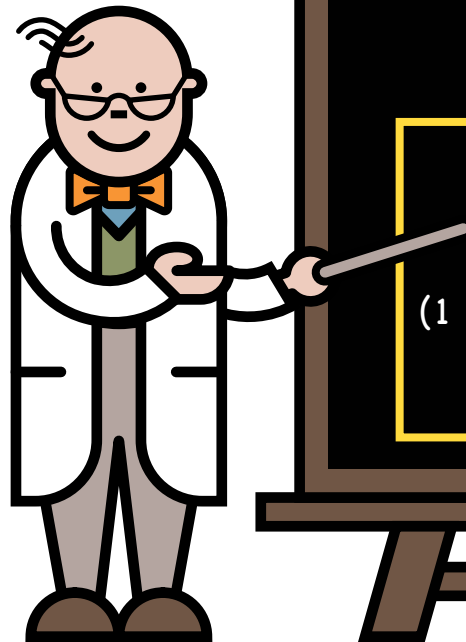
Inheritance – Autosomal recessive (carriers of VLCADD)



Inheritance – Autosomal recessive – possible combinations



Future pregnancies



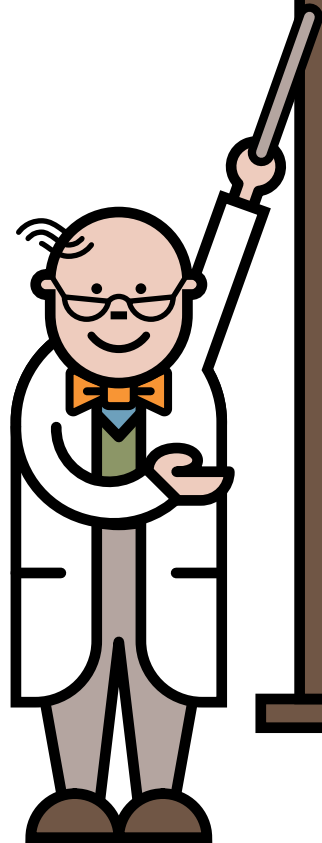
When both parents are carriers, in each pregnancy the risk to the baby is as follows:

25% chance
(1 in 4) of VLCADD

50% chance
(1 in 2) for the baby to be a carrier of VLCADD

25% chance (1 in 4) for the baby to have two working genes and neither have VLCADD or be a carrier

Take home messages



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

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

Children with severe VLCADD are managed with a low fat diet, MCT oil and regular meals.

People with VLCADD should lead normal, healthy and active lives

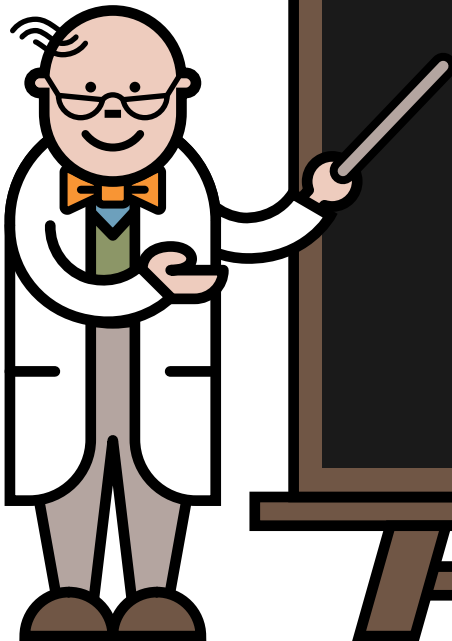
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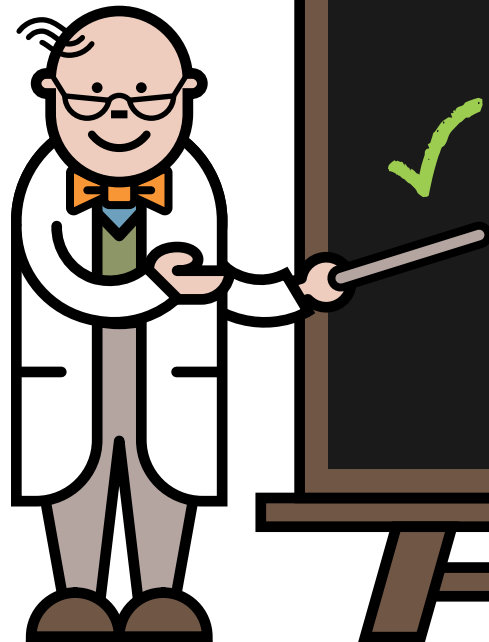
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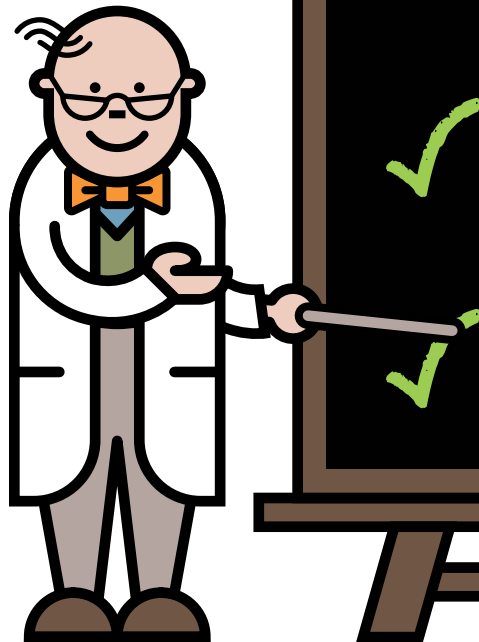
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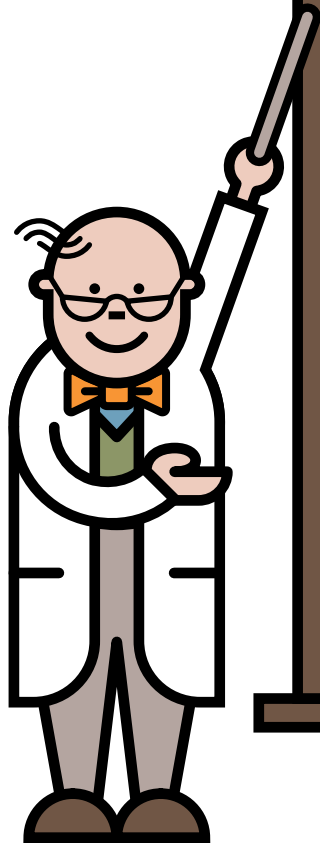


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Helpful hints



Always ensure you have a good supply of your emergency 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

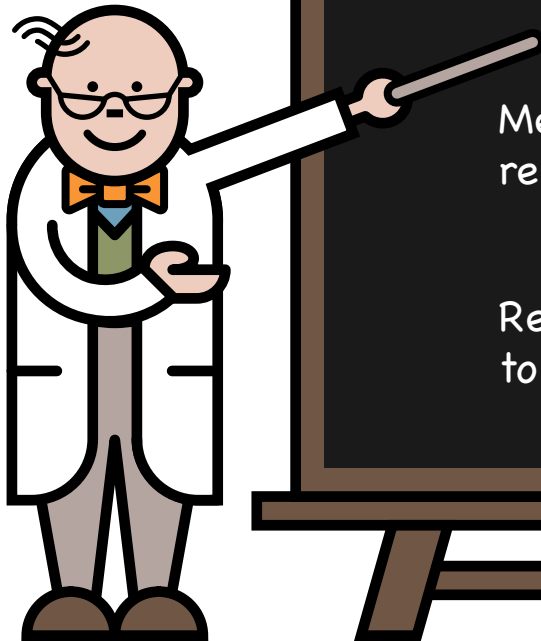
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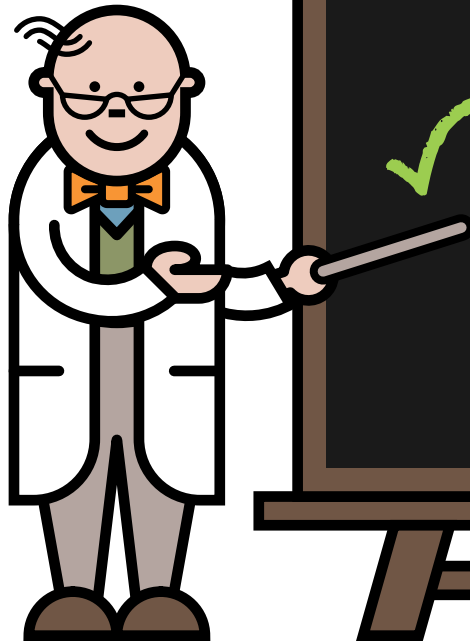
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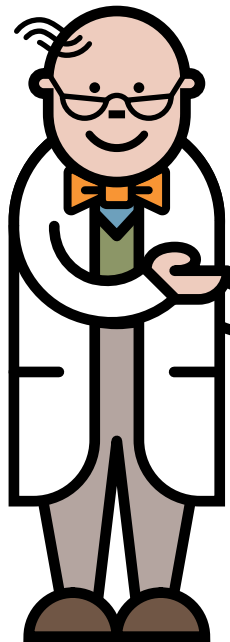
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Who's who

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

BIMDG

British Inherited Metabolic Diseases Group



NUTRICIA
LIFE-TRANSFORMING NUTRITION

**METABOLIC
SUPPORT UK**

Your **rare** condition.
Our common fight.

www.bimdg.org.uk

www.nutricia.co.uk

www.metabolicsupportuk.org