

# Top Tips

for  
GSD  
III

Helpful hints for living well  
with Cori Disease



Edition 1  
October 2018

The CATS team would like to say a huge thank you to Dr Elaine Murphy of the Charles Dent Metabolic Unit at The National Hospital for Neurology and Neurosurgery for her hard work, freely given, checking our Top Tips.

**Association for Glycogen Storage Disease (UK) Ltd**

Old Hambledon Racecourse  
Sheardley Lane  
Droxford SO32 3QY  
[www.agsd.org.uk](http://www.agsd.org.uk)

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# What is Cori Disease?

Cori disease is a rare disease of varying severity, affecting primarily the liver, heart and skeletal muscle. It is caused by deficient activity of glycogen debranching enzyme, which is a key enzyme in the breakdown of glycogen.

The condition has a wide clinical spectrum. Children are often diagnosed when a swollen abdomen due to a large liver is noticed. Patients may also have low blood sugar, a high level of fats in the blood and delayed growth.

Symptoms related to liver disease and progressive cardiac and skeletal muscle involvement vary in age of onset, rate of disease progression and severity.

# Who Wrote the Book?

## **The CATS – Cori Action Team Support**

CATS is a group of patients and carers who have got together to support those affected by Cori disease. As well as supporting patients and families we aim to be active in recognising and promoting the needs of Cori patients with medical staff through the work of AGSD-UK.

**If you would like to know more or ask for some  
help contact 0300 123 2792  
or email [cats@agsd.org.uk](mailto:cats@agsd.org.uk)**

### **Ailsa Arthur** (*Trustee AGSD UK*)

Ailsa has twenty years' experience of supporting her son with Cori's Disease.

She feels patient and family involvement are crucial to help improve treatment and support. She is very keen to take an active involvement in getting recognition and awareness of this little known condition out to the public.

### **Nikki Christie**

I was diagnosed with Cori's Disease 51 years ago, although my life has changed somewhat over the last 7 years I am learning to cope with my new limitations due to muscle deterioration. Despite my limitations I am still working full time and leading a different, but happy life. I am eager to try and help to find ways to help adults experiencing the same difficulties, as well as helping parents with newly diagnosed children.

**Sylvia Wilson** (*Type III co-ordinator*)

I have a granddaughter diagnosed with Cori's disease. She lives abroad and it took almost a year to get her diagnosis. She was the first case her hospital had dealt with. It was a very frightening time for the family and I took to the internet to research the disease. After much research I eventually came across AGSD-UK.

I was warmly welcomed into the GSD family. I attended my first conference not sure what to expect but it was such a positive and informative occasion.

I would urge everyone to attend if they can. GSD has been a steep learning curve. I'm interested in genetics and the view that carriers can also display aspects of the disease. I joined CATS to help other people cope with this disease.

## **Gary, Andrea and Lauren Thompson**

Lauren is 18 years old and our only child. Lauren was diagnosed with GSD type 3 at the age of 11 months. Over the last two years Lauren has been taking daily doses throughout the day of Glycosade and protein powder. This year (2018) Lauren is making the transition from the Children's to Adults Hospital.

We have had many, many concerns to deal with over the previous years, including working with CAMHS (Child, Adults Mental Health Services) and we have worked together to support and encourage Lauren in how to deal with her issues.

We have joined CATS team to help others by sharing our own knowledge with those who have been diagnosed with GSD type 3.

# The Tips in this Book

The tips in this book were collated from many people with Cori disease from all over the world and are there to help parents who have children with Cori disease, newly diagnosed patients, and adults that are learning to cope with myopathy.

Hopefully, you will find this book useful and find some tips that are new to you.

There is a glossary at the back to help you with any unfamiliar words.



# They are just tips!

First, before reading this book, make sure you have the correct diagnosis. Then, remember we are all affected in different ways by Cori. Assess each tip individually and think about whether it suits you.

## Disclaimer

Use this book as a guide, it should not be replaced for personalised medical advice from your Cori medical consultant.

Research is still ongoing to gain further knowledge of Cori disease. Please check the AGSD-UK website to read any updates to this book

# Using this book

## **Glycogen storage disease type III (GSD III, previously known as 'Cori disease') Top Tips**

These tips are intended as a guide only. Please discuss any specific queries with your medical team or dietitian. Remember that individuals with GSD III will vary in the severity of their disease and so not all these tips may be relevant to you.


### **There are four forms of GSD III:**

GSD IIIa - affects liver and muscle (low blood glucose + muscle symptoms)

GSD IIIb - affects liver only (low blood glucose)

GSD IIIc - affects liver and muscle

GSD IIId - affect liver

If you are unsure which type of GSD III you have, then ask your medical team or dietitian. The tips with a blue star  are more specific for GSD IIIa and may not be so relevant for individuals with GSD IIIb.



# Shopping

# 1

## Shopping

When doing  
your shopping  
try to think  
about including  
some protein in  
every meal.

Make your own information card for shopping, include foods you can and cannot eat and your likes, this will make shopping much easier.

# 3

Purchase an SOS  
Bracelet, or  
download a medical  
ID app onto your  
smartphone.



Use shopping  
apps that  
support  
AGSD UK.



# Emergencies





You can apply  
for a free  
message in a  
bottle in case of  
an emergency.



[www.scas.nhs.uk/news/campaigns/message-in-a-bottle](http://www.scas.nhs.uk/news/campaigns/message-in-a-bottle)

# 6

## Emergencies

### **Smart phone**

Keep medical ID in  
this section -  
paramedics will look  
for this in an  
emergency situation.

Smart phone  
use the **ICE**  
**I**n  
**C**ase of  
**E**mergency  
number.

# 8

## Emergencies

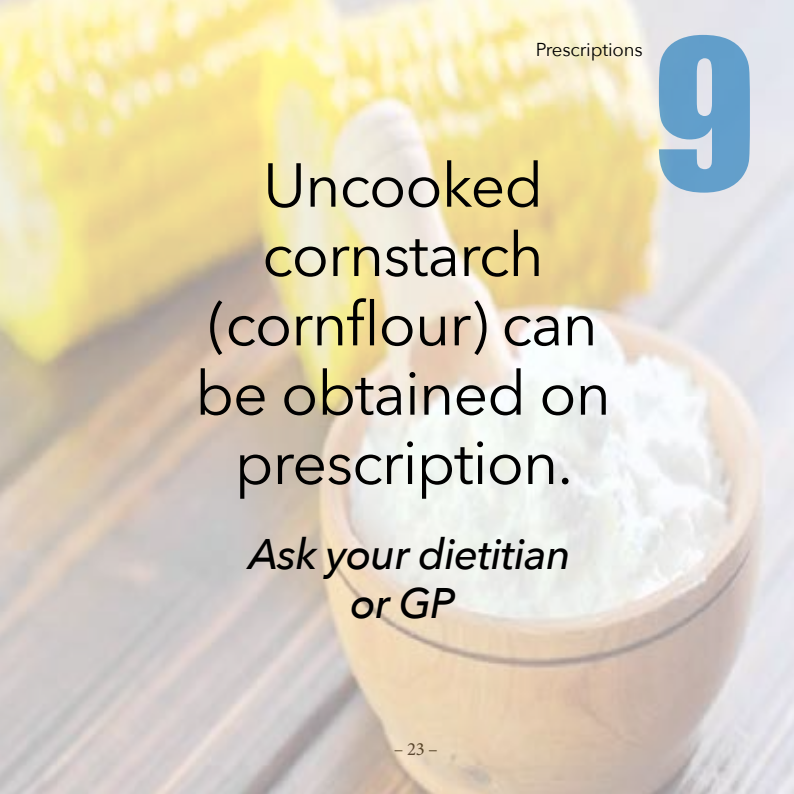
The British Inherited Metabolic Disease Group have published agreed guidelines on the emergency management of GSD III.



These are freely  
available on their  
website:

*[www.BIMDG.org.uk](http://www.BIMDG.org.uk)*

# Prescriptions



Uncooked  
cornstarch  
(cornflour) can  
be obtained on  
prescription.

*Ask your dietitian  
or GP*

# 10 Prescriptions

Protein powder  
can be  
obtained on  
prescription.

*Ask your dietitian  
or GP.*



You may be exempt  
from prescription  
costs.

See eligibility here:

*[www.nhsbsa.nhs.uk/exemption-certificates/medical-exemption-certificates](http://www.nhsbsa.nhs.uk/exemption-certificates/medical-exemption-certificates)*

*Speak to your pharmacist for  
further details.*

# 12

If you are not exempt from prescription charges then you may want to purchase a pre-payment certificate to reduce the cost

*<https://apps.nhsbsa.nhs.uk/ppcwebsales/patient.do>*

*Speak to your pharmacist  
for further details*

A large, stylized, light blue eye graphic is centered in the background of the slide. The eye is composed of concentric, curved lines that form the shape of an eye, with a central circular area representing the pupil.

# Monitoring your GSD

# 13

Monitoring your GSD

At 18 although you are an adult, if you wish, you can tell your medical team that they CAN talk to named people, e.g. parents, about your condition.

Keep a running notebook of all your medical appointments so you can remember who said what and when.

# 15

Monitoring your GSD

Keep a record in a notebook of how you are affected daily, this will help if and when you have to complete benefit claims such as ESA or PIP.

Keep any letters that consultants have copied you in on; these can also be used to help to support PIP or ESA claims.

Take videos of your worst days  
- these can also be used to  
support PIP and ESA claims.  
Make sure videos are dated  
correctly.

*Videos cannot be used for DWP  
evidence, but can be a reminder and  
help to complete the forms.*



Very rarely, blood sugar levels are difficult to control in GSD III – if this is you, you might want to consider a sensory support dog.

# Gastrostomy & PEG Tubes

Keep it clean and follow the guidance for care of your tube.

*A few people might need a gastrostomy or PEG tube. This will be discussed with your doctor.*

# 20

Gastrostomy / PEG Tubes

Speak to your local nutritional care team- community nurses – they can help a lot.

*Your dietitian or GP can refer you.*



# Dentist

# 21

Dentist

Keeping your mouth open can be painful.  
Ask for a mouth prop.





Dentist

22

If your diet includes a lot of carbohydrate or sugar then see a dental hygienist regularly.

# Exercise





In some parts of the UK you can self-refer yourself for physiotherapy, which may lead to a course of hydrotherapy, which can benefit muscle pain and weakness.

*[www.nhs.uk/conditions/physiotherapy/](http://www.nhs.uk/conditions/physiotherapy/)*

***Ask your GP or consultant.***

# 24

Exercise

Don't exercise  
on an empty  
stomach.

Be careful not to  
push to the  
maximum, take  
regular breaks and  
explain to others  
why you need this.

# 26

Exercise

## Swimming

Take regular breaks and use a clear dressing over stoma sites.

It is important to  
keep exercising.  
If you don't use  
it you will lose it.

# 28

Exercise

Get a dog or walk a friend's dog. When walking - take rests and snack on route.

Walking up hills -  
take the gentler  
route even if longer  
and 'zig-zag'...

"Take the gentler  
route through life"

# 30

Exercise

Avoid  
dehydration  
during exercise  
to reduce the  
risk of muscle  
cramps.



Consider an e-bike, if you find standard cycling too strenuous.



# Diet


If you do not have a dietitian, find one.

*Ask your GP to refer you to a dietitian who knows about GSD III.*

# 33

Diet

Don't be afraid to ask your dietitian questions, or talk to them if you are finding it hard to manage your diet.



Eggs, chicken,  
pork and lean  
beef are good  
protein source.

# 35

Diet

Fish and seafood are good sources of protein and are typically low in fat.

Salmon and other  
oily fish have a  
slightly higher fat  
content compared to  
white fish, because  
they contain  
omega-3 fats.

# 37★ Diet

Use minimal fat when cooking protein-rich foods – use a spray oil to stir fry or pan fry.



Reduced-fat dairy products (such as low-fat or fat-free yoghurt, lower-fat cheeses and semi-skimmed or skimmed milk) are all good sources of protein.

# 39

Diet

Unsalted nuts and seeds are a practical protein choice if you're on the move (but do contain fat).


50 pistachio nuts (35g) provides 6g of protein.

Use nut butters  
on toast  
instead of jam  
or honey.

# 41

Diet

Sprinkling  
seeds on  
salads will  
boost protein  
content.



Beans and  
pulses are cheap  
protein sources.

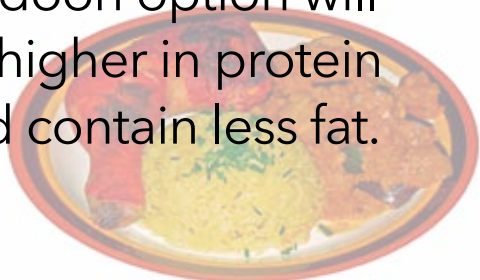
Add them to  
stews, soups  
and casseroles.

# 43

Diet

Quorn is a  
good source of  
non-animal  
protein.

If you are eating out  
in an Indian  
restaurant, the  
tandoori option will  
be higher in protein  
and contain less fat.

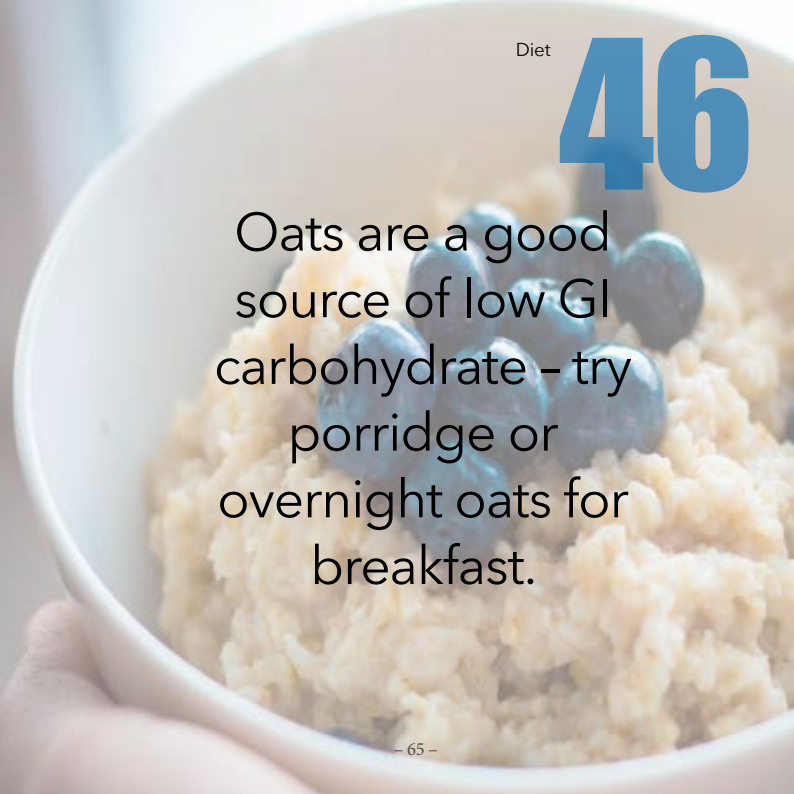


# 45

Diet

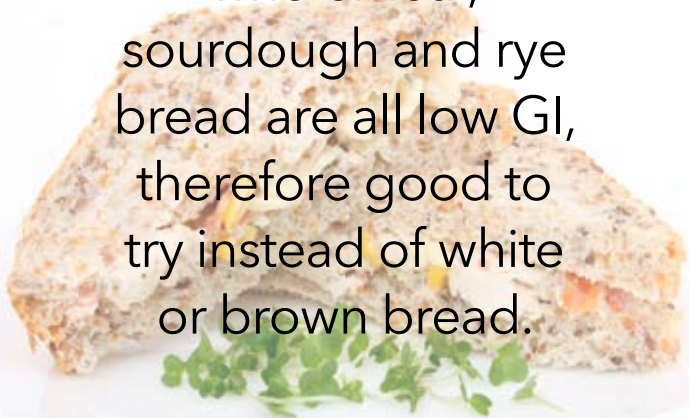
Slow release or low glycaemic index (GI) carbohydrates are broken down more slowly, so can be helpful for maintaining blood sugar levels.



A close-up photograph of a white ceramic bowl filled with cooked oatmeal. Several fresh blueberries are scattered on top of the oatmeal. A person's hand is visible at the bottom left, holding the bowl. The background is softly blurred.

Oats are a good source of low GI carbohydrate – try porridge or overnight oats for breakfast.

Granary,  
wholemeal,  
sourdough and rye  
bread are all low GI,  
therefore good to  
try instead of white  
or brown bread.



Both wholewheat  
and white pasta are  
low GI  
carbohydrates, but  
wholewheat  
contains more fibre.

# 49 Diet

Berries are a  
better low GI fruit  
option - including  
frozen.



Many fruits are  
low GI – including  
apples, oranges  
and pears.

# 51

Diet

Milk contains both protein and also low GI carbohydrate, so can be helpful for managing blood sugar levels.


Small portions of  
breakfast cereal  
can be a handy  
carbohydrate  
snack at school.

# 53

Diet

In general, try to limit sugary foods in your diet – except in situations to treat hypoglycaemia where quickly released carbohydrates (such as sugar) is important.





Natural sugar is still sugar  
– aim to limit portion sizes  
of foods that contain a lot  
of natural sugars such as  
fruit juice and honey.

Some reduced  
fat products can  
often have  
added sugar  
– check the  
label.

Always check food labels, and keep checking them because food manufacturers change their recipes.

# 57

Diet

Try sweeteners  
such as Xylitol  
or Stevia as  
alternatives to  
sugar.

Don't forget  
savoury products  
often contain high  
levels of sugar – eg.  
tinned soups or  
ready-made sauces.

59<sup>Diet</sup>

Check all  
ingredients on  
tins and  
packaging to  
avoid added  
sugars.

Be cautious with alcohol as this can lower blood sugar levels – avoid this before exercise, and be sure to have this with food.

Limit your intake to 1-2 drinks with a meal.





# At Home



## **Batch cooking**

If you make  
complicated  
measured meals,  
make a lot and  
freeze portions.

Contact your local occupational health therapist. They will help with making sure you are equipped with any aids and supports to help maintain your independence around the house.

*Ask your GP or physiotherapist for a referral.*

# Daily Activities

# 63

Daily Activities

Don't wait until  
your blood sugar is  
low - eat small  
amounts regularly.



## Education

Consider whether your child needs extra time to complete exams or assignments.

*Ask for extra breaks during exams to eat / drink if needed.*

## Education

Consider an Education Health Care Plan, speak to AGSD-UK and your medical team for supporting letters.



## 66

Wear a backpack,  
not a shoulder bag  
or handbag, this will  
be easier for you to  
manage if you have  
muscle weakness.



# Support & Benefits



Consider PIP and ESA;  
get professional  
support letters as  
evidence.

DWP might not  
understand your  
condition.

*Speak to AGSD-UK.*

## Benefits

Don't give up, understand the system and make your case within the rules.

REMEMBER: Many PIP claims are refused first time – so fill in the form carefully and with lots of detail.

Be prepared to appeal the decision if you feel it is incorrect.



Understand that  
complex feelings  
happen around  
inherited  
conditions, there is  
no shame or guilt in  
genetics.

# 70

Support and Benefits

AGSD-UK  
will help you.





Don't be afraid to reach out  
to as many support agencies  
as you can.

GSD is now officially under  
the Muscular Dystrophy  
umbrella.

You can get help from  
M.D.U.K. group now too.

# 72

Support and Benefits

Government scheme,  
Access to Work will  
help with any aids and  
support you in your  
fight to stay in work.

[www.gov.uk/access-to-work](http://www.gov.uk/access-to-work)

Keep fighting for the  
best care, the best  
doctor, the best  
consultant.

Not everyone is a  
GSD expert!



If the time comes that you need to use a wheelchair, try not to feel embarrassed; mobility aids are there to help you, not make you feel worse.



Reach out for support, try  
Facebook, there are several  
different groups you can join;

*Adults with Glycogen Storage Disease*

*Behind The Mystery Rare & Genetic Diseases*

*Glycogen Storage Disease type III*

*Glycogen Storage Disease in the UK and Ireland*

You are not the only one,  
find a friend.

Spread the word



We need to raise  
awareness of our rare  
condition.

Tell everyone about  
GSD.

You never know who  
might be listening.

A group of five stylized figures, possibly representing children or people of different backgrounds, are gathered in a circle. They are wearing colorful clothing and headgear. The background is a light, textured surface.

Talk to schools and groups (as appropriate) about GSD.

Most people are interested and will support you. If they don't know they can't adapt their provision or behaviour.



# Travel

# 78

Travel



Preparation is key.

Make sure you  
have enough  
cornstarch and / or  
protein powder for  
your entire trip,  
including possible  
delays.

# 80

Travel

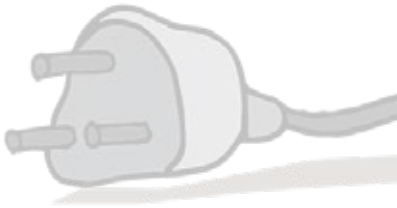
Make sure you  
have appropriate  
travel insurance.

See *AGSD-UK website or  
phone for advice*



Even if you are  
making a car  
journey make  
sure you always  
have a snack  
with you.

Research, research  
research! Make  
sure your holiday  
destination has  
wheelchair access  
if needed.



If travelling abroad, your power chair charger may not be compatible with foreign voltage.

It may be easier to hire a wheelchair when you arrive instead of taking yours with you.

Power chair joysticks are very fragile and may get damaged in transit.



A really useful travel site  
for wheelchair users:



[www.curbfreewithcorylee.com](http://www.curbfreewithcorylee.com)

# 86

Travel

Ask your dietitian or doctor for a customs letter before flights – this will allow you to take medications, cornstarch or protein powder on board the plane as hand-luggage if needed.

# Other Tips

# 87

## Other Tips

Do not underestimate how much hypoglycaemia can affect intellectual and emotional states as well as physical ability.





Everyone is different  
including everyone with  
GSD III - pay heed to your  
own body and  
reactions, and don't  
uncritically accept what  
others do.

# 89

Other Tips

Be adaptable and attentive -  
symptoms and  
tolerances change  
with time and age.



# Useful Links

**Access to Work**

[www.gov.uk/access-to-work](http://www.gov.uk/access-to-work)  
[atwosu.london@dwp.gsi.gov.uk](mailto:atwosu.london@dwp.gsi.gov.uk)

**Association for Glycogen Storage Disease UK**

[www.agsd.org.uk](http://www.agsd.org.uk)

**Citizens Advice Bureaux**

[www.citizensadvice.org.uk](http://www.citizensadvice.org.uk)

**Carer's Federation**

[www.carersfederation.co.uk/services/counselling](http://www.carersfederation.co.uk/services/counselling)

**Carer's Trust**

[www.carers.org](http://www.carers.org)

**CEA Card (Cinema Exhibitors Association)**

[www.ceacard.co.uk](http://www.ceacard.co.uk)

**Disability rights**

[www.gov.uk/rights-disabled-person/overview](http://www.gov.uk/rights-disabled-person/overview)

## **Disabled Railcard**

[www.disabledpersons-railcard.co.uk](http://www.disabledpersons-railcard.co.uk)

## **Raise Funds for Charity with your everyday shopping**

[www.giveasyoulive.com](http://www.giveasyoulive.com)

## **Motability**

[www.motability.co.uk](http://www.motability.co.uk)

## **Aids for daily living**

[www.dlf.org.uk](http://www.dlf.org.uk)

## **Reasonable Adjustments in the workplace**

[www.equalityhumanrights.com/your-rights/  
employment/work-place-adjustments](http://www.equalityhumanrights.com/your-rights/employment/work-place-adjustments)

[www.gov.uk/reasonable-adjustments-for-disabled-  
workers](http://www.gov.uk/reasonable-adjustments-for-disabled-workers)

## **Disabled students allowance**

[www.gov.uk/disabled-students-allowances-dsas/overview](http://www.gov.uk/disabled-students-allowances-dsas/overview)

## **Self referral Physiotherapy/Hydrotherapy**

[www.nhs.uk/conditions/pysiotherapy](http://www.nhs.uk/conditions/pysiotherapy)

## **Joining Groups**

[www.facebook.com](http://www.facebook.com)

## **Travel Site for wheelchair users**

[www.curbfreewithcorylee.com](http://www.curbfreewithcorylee.com)

## **Better Mobility**

[www.bettermobility.co.uk/charity\\_funding\\_options.php](http://www.bettermobility.co.uk/charity_funding_options.php)

## **Online Self-Help for Depression & Anxiety**

[www.moodgym.anu.edu.au](http://www.moodgym.anu.edu.au)

## **Online Courses to manage Anxiety/Stress**

[www.lltff.com](http://www.lltff.com)

## **Free Legal Advice for Disabled/Carers**

[www.dls.org.uk](http://www.dls.org.uk)

## **National Database of Volunteering Opportunities**

[www.do-it.org](http://www.do-it.org)

## **Organised Walks Group for people with Mental/ Physical Health problems.**

[www.walkingforhealth.org.uk](http://www.walkingforhealth.org.uk)

# Glossary



## **Blood Glucose**

The amount of glucose that is in your blood.

## **Protein (Nutrient)**

Essential nutrients for the human body. They are one of the building blocks of body tissue, and also serve as a fuel source. As a fuel, proteins contain 4 calories (kcal) per gram, just like carbohydrates, but unlike fats, which contain 9 kcal per gram. The defining characteristic of protein from a nutrition standpoint is its amino acid composition.

## **SOS Bracelet**

An bracelet available to buy containing your personalised medical information and emergency contact numbers.

## **Personal Independent Payment. (PIP)**

A UK government benefit that helps with some of the extra costs caused by long term illness, ill health or a disability if you're aged 16 to 64. The rate depends on how your

condition effects you, not the condition itself. You'll need an assessment to work out the level of help you get.

## **ESA**

Employment and Support Allowance (ESA) is for people of working age who cannot work because of illness or disability.

Entitlement usually depends upon your National Insurance record (except for some young adults), and may be subject to a medical assessment.

## **Gastrostomy / PEG tubes**

PEG stands for percutaneous endoscopic gastrostomy, a procedure in which a flexible feeding tube is placed through the abdominal wall and into the stomach.

PEG allows nutrition, fluids and/or medications to be put directly into the stomach, bypassing the mouth, often used for children with eating problems.

## **Physiotherapy**

A therapy to help restore movement and function when someone is effected by injury, illness or disability.

A neuromuscular physiotherapist would be needed to teat a person with Cori disease.

## **Cori Disease**

Another name for GSD III in honour of the 1947 Nobel laureates Carl Cori and Gerty Cori. Cori disease is a rare disease of variable severity affecting primarily the liver, heart and skeletal muscle. It is caused by deficient activity of glycogen debranching enzyme, which is a key enzyme in the breakdown of glycogen.

## **Forbes Disease**

Another name for GSD III in honour of American clinician Gilbert Burnett Forbes.

## **Dehydration**

Dehydration happens when your body doesn't have as much water as it needs. Without enough, your body

can't function properly. You can have mild, moderate, or severe dehydration depending on how much fluid is missing from your body.

## **Dietician**

A dietician is a degree-qualified health professional who helps to promote nutritional well-being, treat disease and prevent nutrition related problems. They provide practical, safe advice, based on current scientific evidence.

## **GI**

GI stands for glycaemic index. It is a rating system for foods containing carbohydrates. It shows how quickly each food affects your blood sugar level when that food is eaten on its own.

## **Glycemic Load**

The glycemic load (GL) of food is a number that estimates how much the food will raise a person's blood glucose level after eating it. One unit of glycemic load approximates the effect of consuming one gram of glucose. Foods can be referred to as having high or low GL.

## **Occupational Health Therapist**

Occupational therapists provide practical support to help children and adults of all ages, with mental, physical, social or learning disabilities, to independently carry out everyday tasks or occupations with more confidence and independence.

## **Department for Work and Pensions**

The Department for Work and Pensions (DWP) is responsible for welfare, pensions and child maintenance policy.

As the UK's biggest public service department it administers the State Pension and a range of working age, disability and ill health benefits.

## **Hypoglycemia**

Hypoglycemia refers to an abnormally low level of sugar, or glucose, in the blood.

Signs of low blood sugar include hunger, trembling, heart racing, nausea, and sweating, becoming easily irritated, tearful, stropky or moody.

**Notes**