

My

GSD and Me!

My GSD is Liver Type 1a



By Dr Gabriella Clark



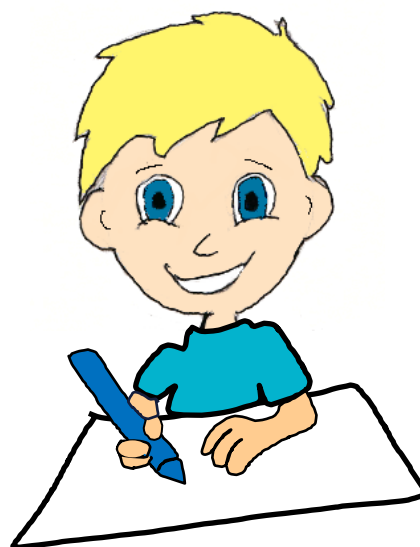
Hello Everyone!

My name is George and I am 9 years old. I live with my mum, dad and sister Charlotte and I love to play football.





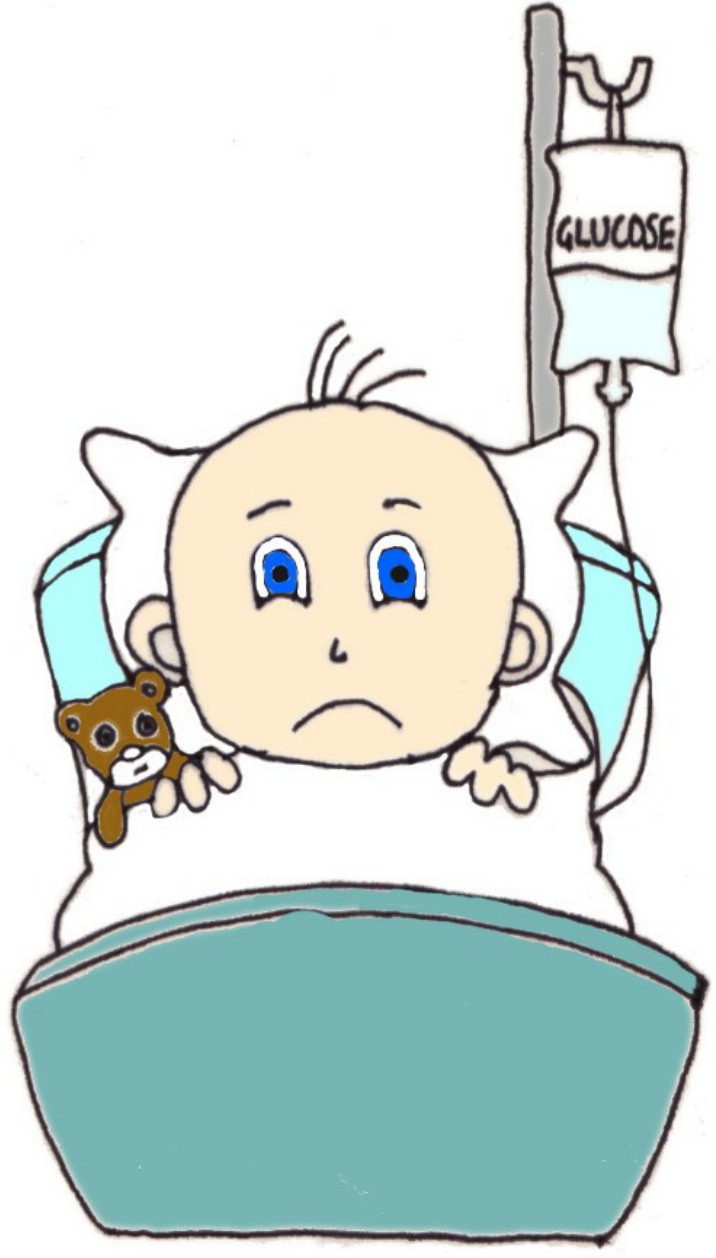
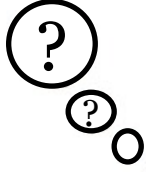
I have something called Glycogen (Gly-ku-jun) Storage Disease, there are lots of different GSDs for the liver and for muscles. My GSD is a liver type called Type 1a. It is a long and confusing name so I just call it my GSD. Not many people in the world have GSD 1a so people don't know much about it. I have had my GSD forever, so I thought it would be cool if I could explain what it is to other kids with it or anyone else who wants to know more about it. I hope you enjoy my story!





When I was born I was a strong and healthy baby but I soon started getting very poorly. I was being sick all the time and my tummy got really big! The doctors and nurses got really worried because they didn't know what was wrong with me. One day I started to shake a lot and an ambulance took me to a big hospital. There were lots of machines and doctors but I don't remember any of it. That is when they found out I had GSD 1a.







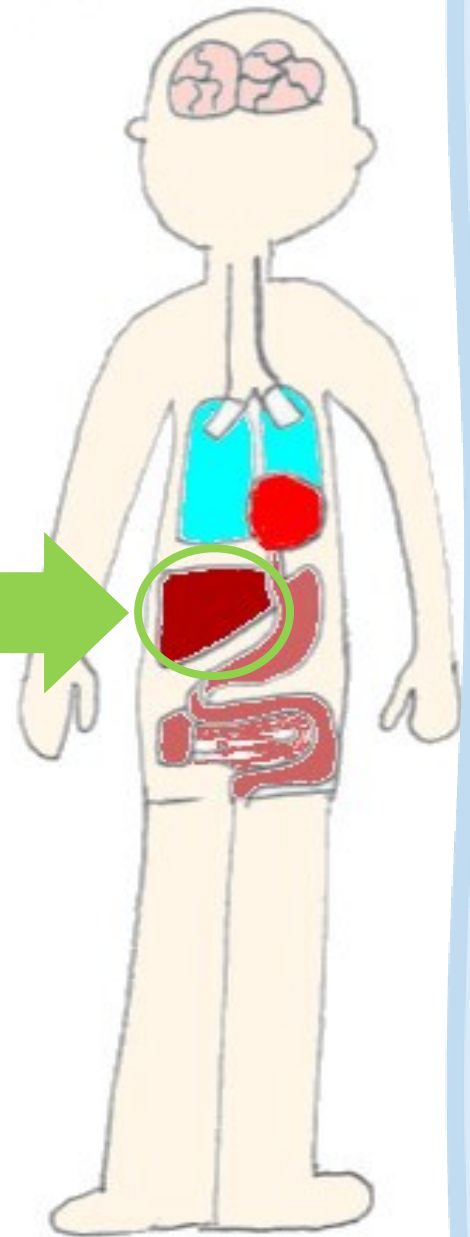
Lots of people ask me what my GSD 1a actually is, and this is what it means to me...

So that we can run around and do stuff, our body needs **energy** every single second of the day. We get this **energy** when we eat food. But we can't eat little bits every second of the day or we wouldn't have time to go to school or play!

So, when we have a meal, our body puts a bit of **energy** in a safe place to use when we are not eating. This safe place is called the **liver**.



This is where the **liver** is, circled in green. The **liver** does loads of different things actually. But I'm just going to talk about how it takes care of our energy.





I think of my **liver** as a kitchen cupboard. We store food in our kitchen cupboards and take it out to give us **energy** when we need it. This is just what our **liver** does too, we just don't feel it!

But, did you know that food gives us different types of **energy**? The important type of **energy** in food to remember is a special sugar called **glucose**.





**I am Glucose! I am
the energy you get
straight away from
most foods you eat! If
you are hungry- it's
because your body
wants more glucose!**





When there is spare **glucose** from a meal, it goes to the **liver** and the muscles. This means we can have **energy** ready for when we are not eating, such as between breakfast and lunch!

My GSD is about the liver, not the muscles.

But now it gets a little bit confusing. When **glucose** goes to the **liver**, it changes its name to **Glycogen**. Why does it do this?

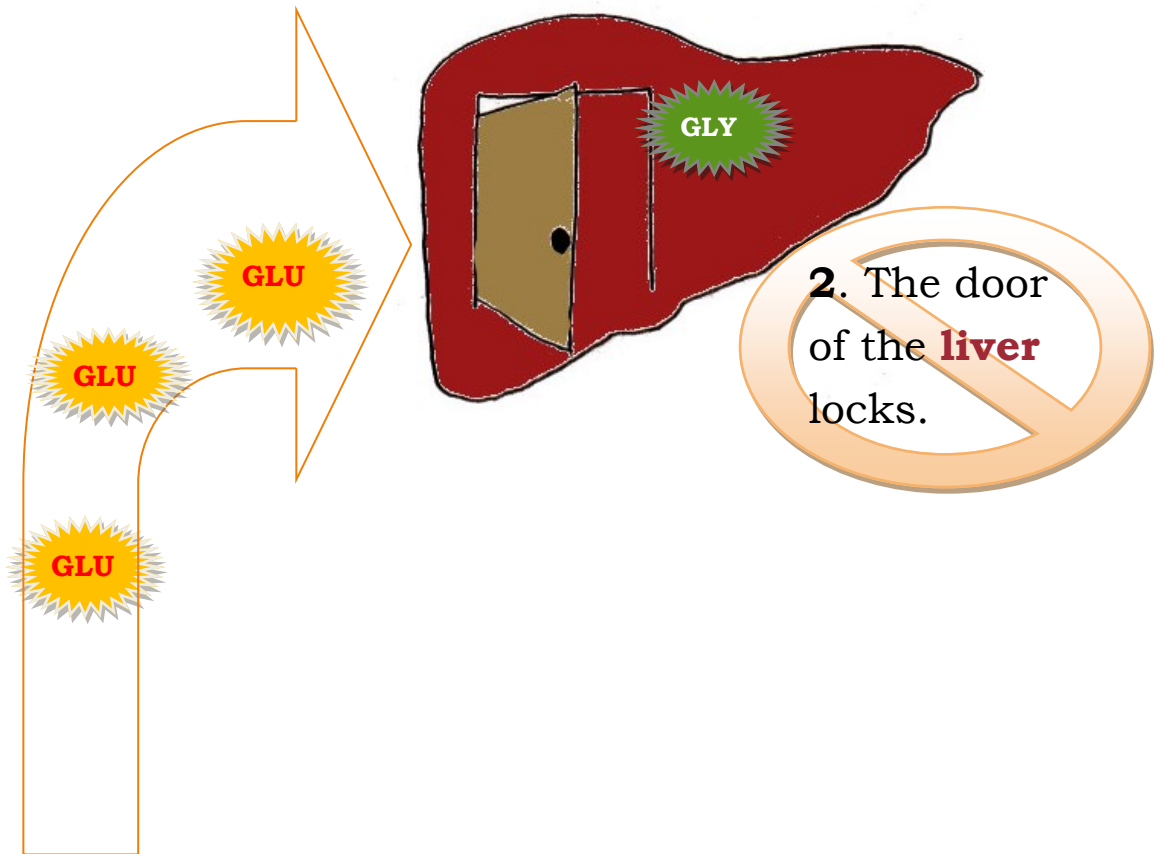




I am Glycogen! I am **glucose** that rests in the **liver**. I have a different name because your body needs to know I am locked away for safe keeping. If you need to use this **energy**, you need a key.

**Turn over, and see how this happens...
you just need to start at GO!**





1. You have spare **glucose energy** in your tummy from a big lunch! So off it goes to your **liver** to be **glycogen**...

6. Now you have some **energy**, you carry on playing until its time for lunch! Then it all starts again!



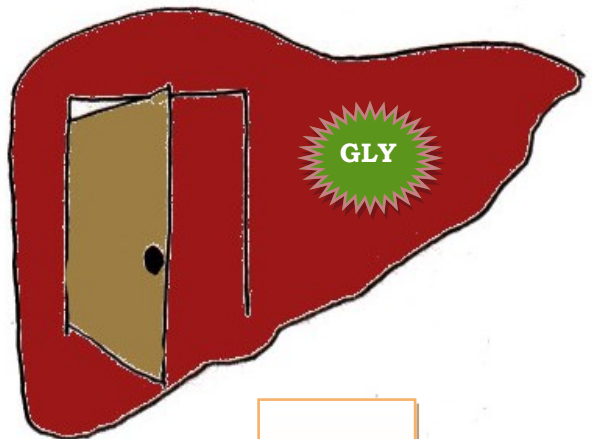


3. You have been running round loads, and start to feel hungry... But you can't eat lunch until later....

4. So, you get your key out to unlock the door of the **liver!**



5. The door opens...



To let out...

Glucose!
You use me
as energy!

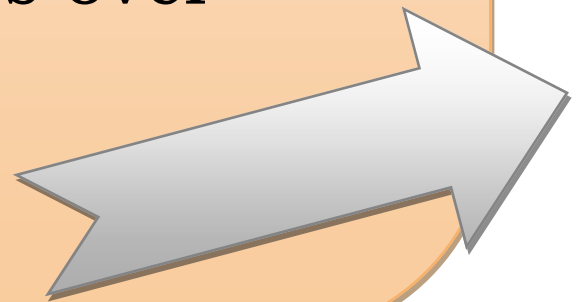




Because I have my GSD 1a, my body can't use **energy** like other people. Something is missing in my body's kitchen cupboard. This is not my fault, I was just born like that.

So, if I have spare **glucose**, it goes into my **liver** just fine. The door is always open for me to store spare **energy**.

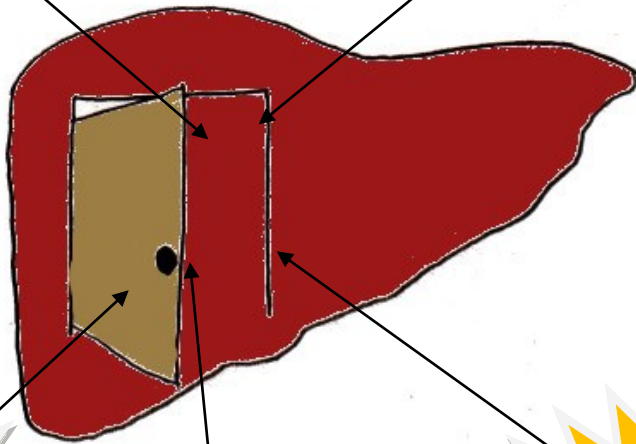
You can see this over
here





**I am
Glucose!**

**I am
Glucose!**



**I am
Glucose!**

**I am
Glucose!**

**I am
Glucose!**



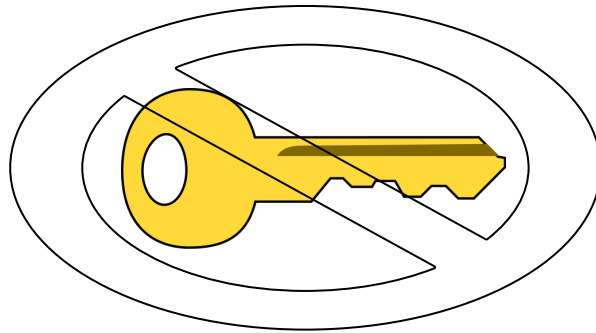


BUT, once the **glucose** has gone into my **liver**, it gets stuck. This is because I don't have that key I showed you earlier that is needed to let any **glucose** out. It's just like someone has put all their food shopping in their kitchen cupboard, locked it for safe keeping and lost the key.

This means that:

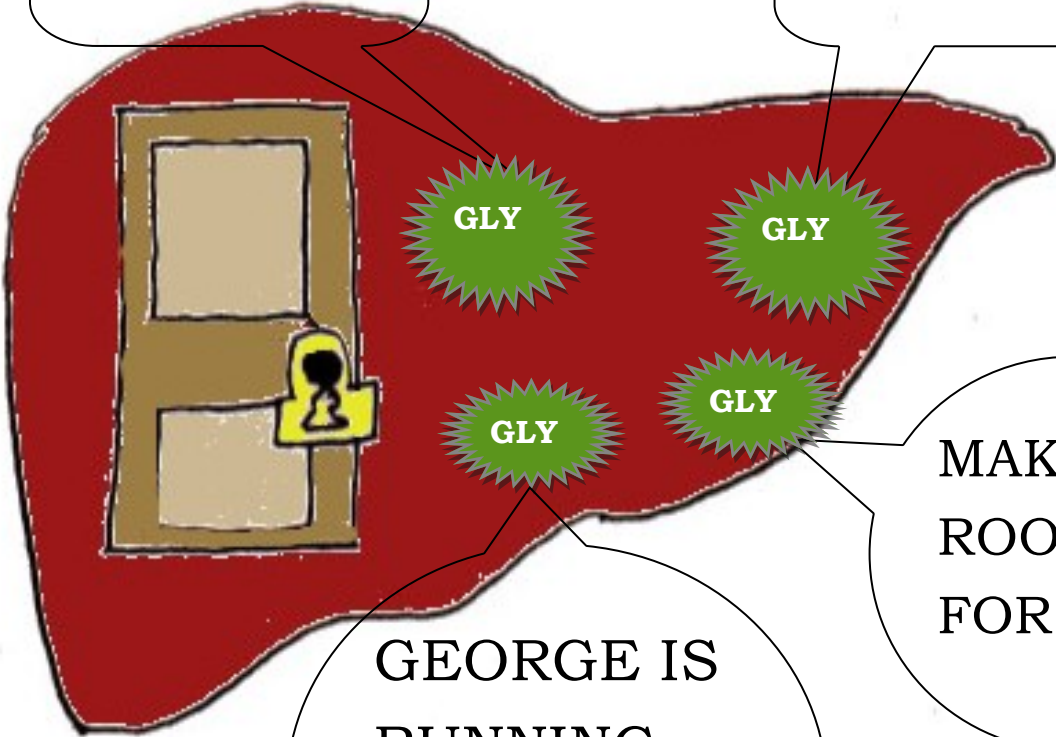
1. Once **glucose** energy is in my **liver**, I can't use it, so, I quickly run out of **energy**
2. If I kept putting **glucose** in, my **liver** would keep getting bigger and bigger with **glycogen** and start to break.





THERE IS NO
KEY!!!

WE ARE
STUCK!!!



GLY

GLY


GLY

GLY


GEORGE IS
RUNNING
OUT OF
ENERGY!!!

MAKE
ROOM
FOR ME!!!





This means I have to eat special food that doesn't need to be stored away in my **liver**. Also, because I don't have spare **energy** in my body, I need to eat more regularly. If I don't do these things, I start to feel 'low.' When I am low I feel weak and wobbly and I need to eat something quickly or else I would get poorly.





So you are probably thinking there isn't much I can eat...



WRONG!!! I can still eat **LOADS** of different things!!





The food I have the most, about five times a day, is called uncooked cornflour. You mix it with water and give it a really good shake before drinking it. It looks just like milk and I don't think it tastes bad.



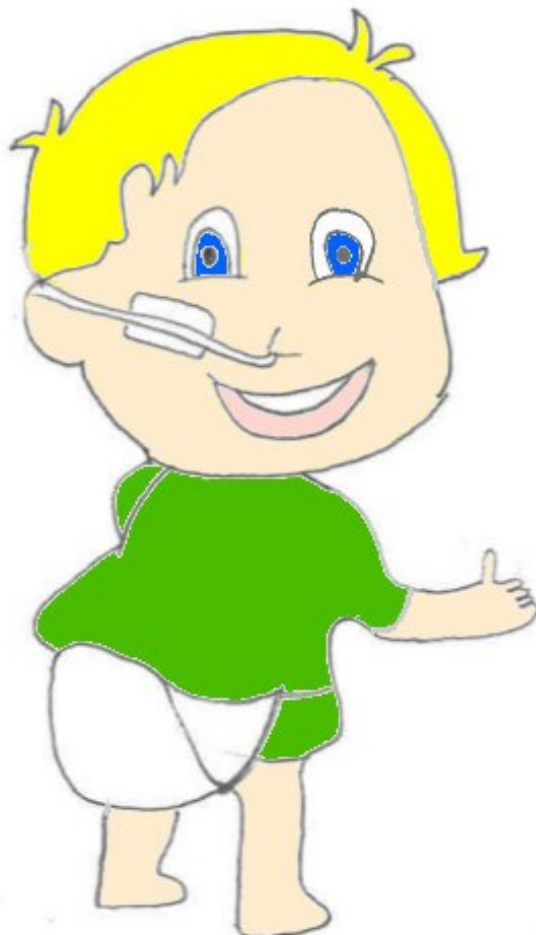
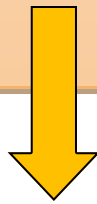
Cornflour is special because the spare **energy** it gives me doesn't go into my **liver** like other foods. Instead it stays in my tummy and is let out slowly for me to use. It doesn't make my GSD go away, but it helps keep me going until my next meal.



After I have my cornflour I tell everyone I have corn powers because it gives me loads of **energy!**



Some people with GSD don't like drinking cornflour at all. They don't like the taste or they don't like to swallow it. They can have a tube in their nose that goes to their tummy called a naso-gastric tube. You then put the cornflour down this tube instead of drinking it. I had one when I was a toddler! Look!





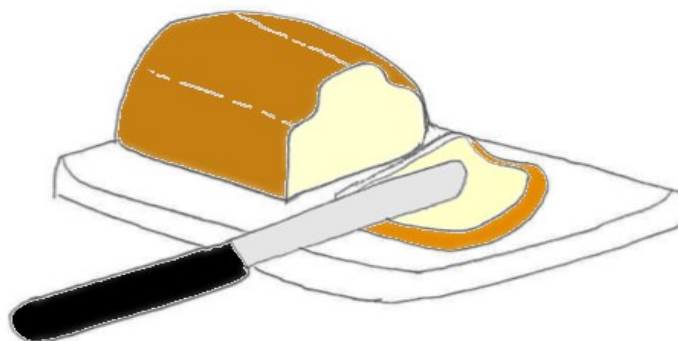
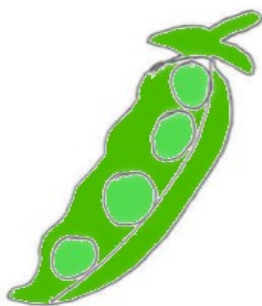
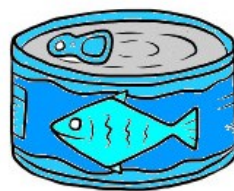
Some foods are bad because they have too much **glucose** that loves hiding in my **liver**.

I usually know a food is bad for me if it is sweet, so stuff like cake, fizzy drinks and fruit. Milk and yoghurt isn't good either because it has lots of a sugar like **glucose** that goes straight to the **liver**.

If I eat foods like this my body can panic and make me go really low.

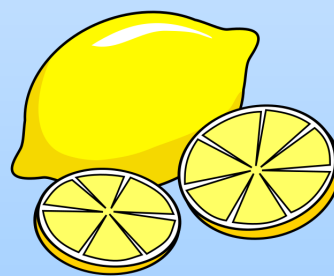


Foods that are good for me are things like bread, meat, fish, vegetables and my cornflour meals. I quite like salty foods like olives too. Because I can't really eat certain things, everyday I take a multivitamin to keep me fit and strong.

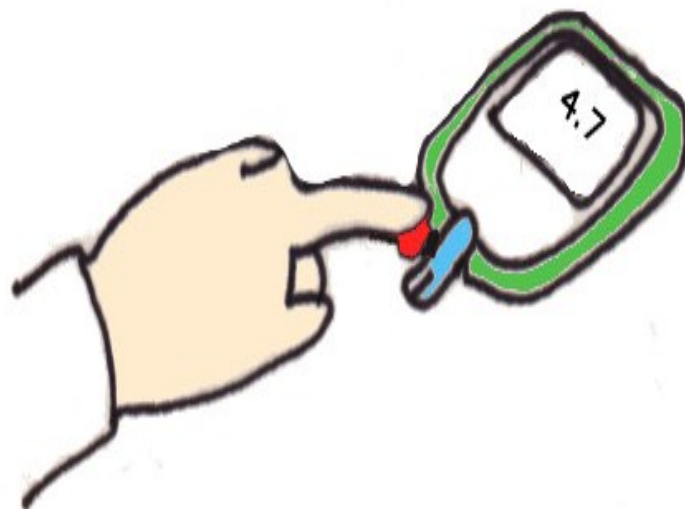




Sometimes it is difficult having my GSD. It makes me feel sad or a bit angry when I can't eat birthday cake or have a fizzy drink at the cinema. But, when I really think about it, it is horrible feeling poorly and being ill can stop me doing other fun stuff. If I do want something sweet, my mum adds some lemon to my food which my **liver** doesn't mind.



My mum and dad also have to prick my finger a couple of times a day to check my **energy** levels in my blood. This is called my blood **glucose** level. Sometimes I do it myself! If the number is below 4, I need a snack or some cornflour quickly. Not everyone with my GSD needs to check their blood **glucose** levels, it just depends on you and what your doctor says.





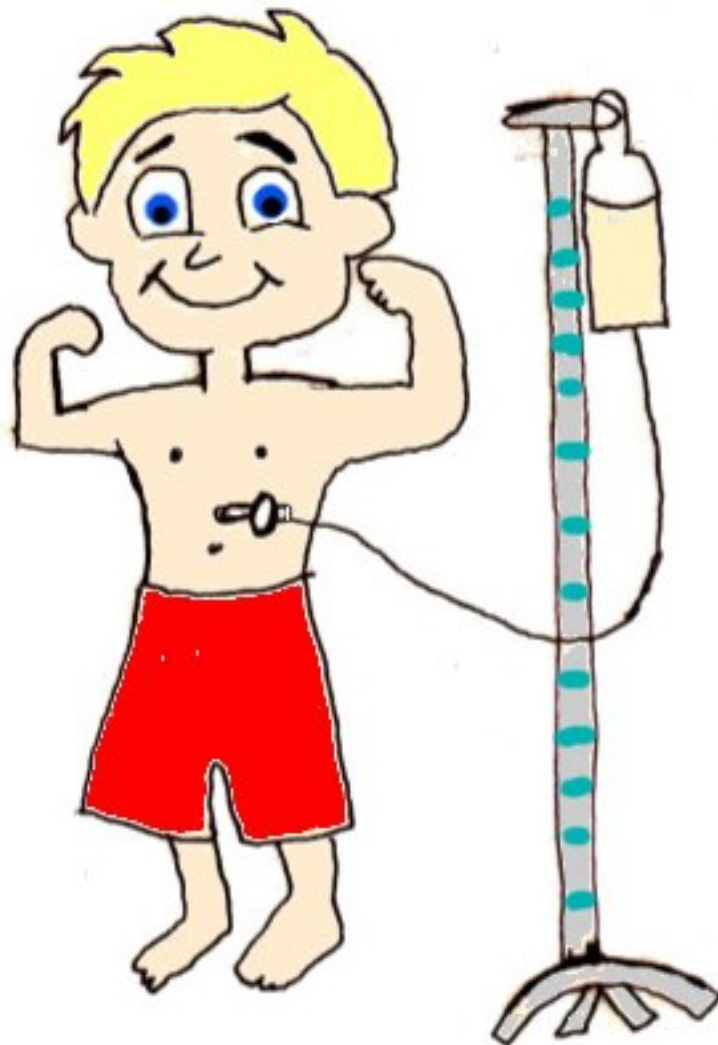
At night time, my GSD doesn't go to sleep like me! Even though you are asleep, your body is still working away, making sure it is ready for the next day. But, because you don't eat when you are asleep, your spare **energy** from your **liver** is VERY important.



This means I need to have my extra **energy** like I do in the day time. It would be really annoying if I had to get up lots in the night to drink cornflour. So, my doctor gave me a little button on my tummy called a Mickey...



When I go to bed my dad puts a small tube on the Mickey button which connects to a bag of special night time **glucose energy** that I hang on a stand next to my bed.

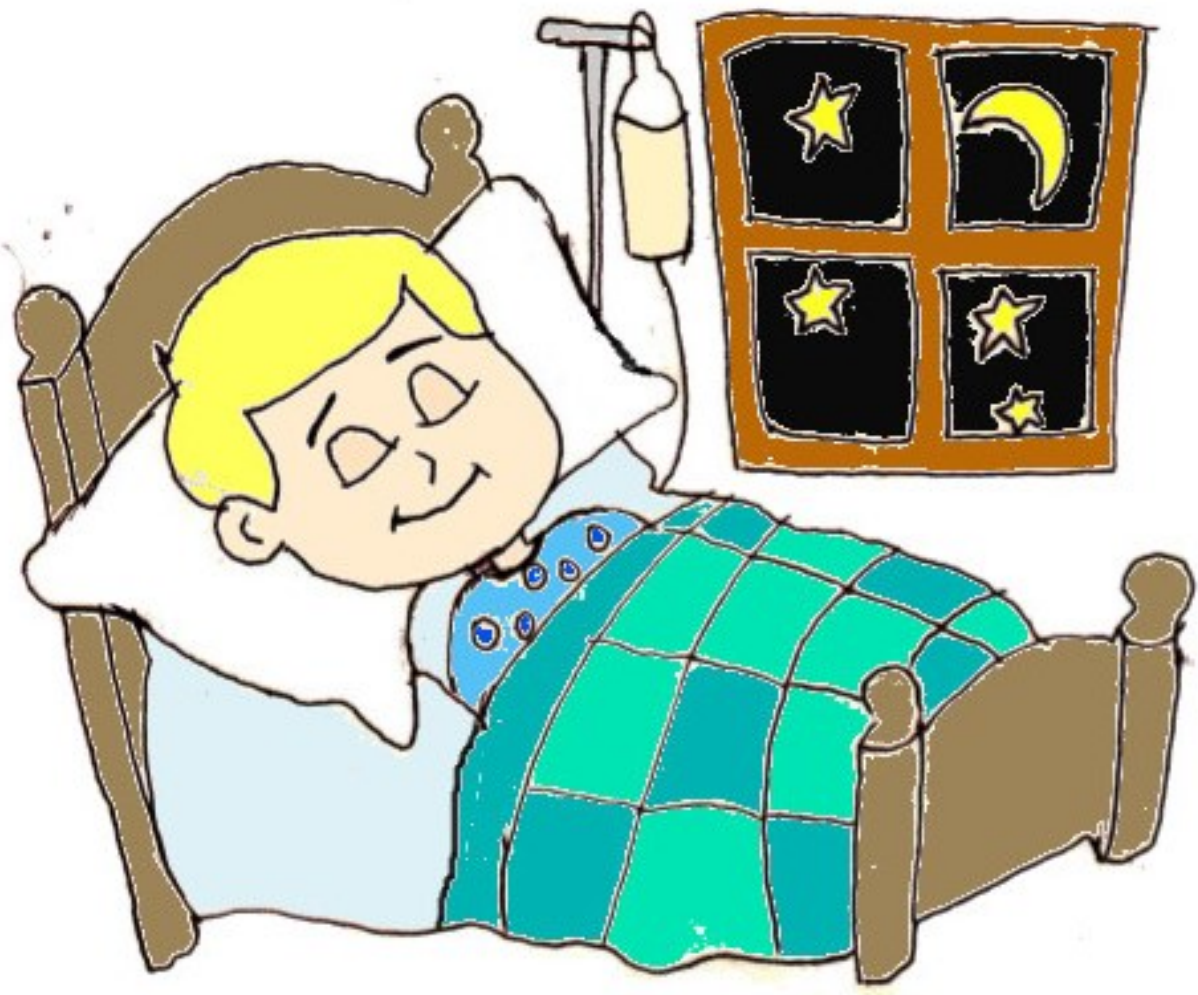




When the bag of **glucose energy** is attached to my Mickey button safely, I am ready to go to bed! The special **glucose** then slowly drips through the tube and into my tummy through the Mickey. It goes all night so I don't go low on **energy**. I don't feel it going in, but I know it is, because I wake up feeling just fine!

The reason I don't have cornflour running through is because it is quite thick and might get stuck!







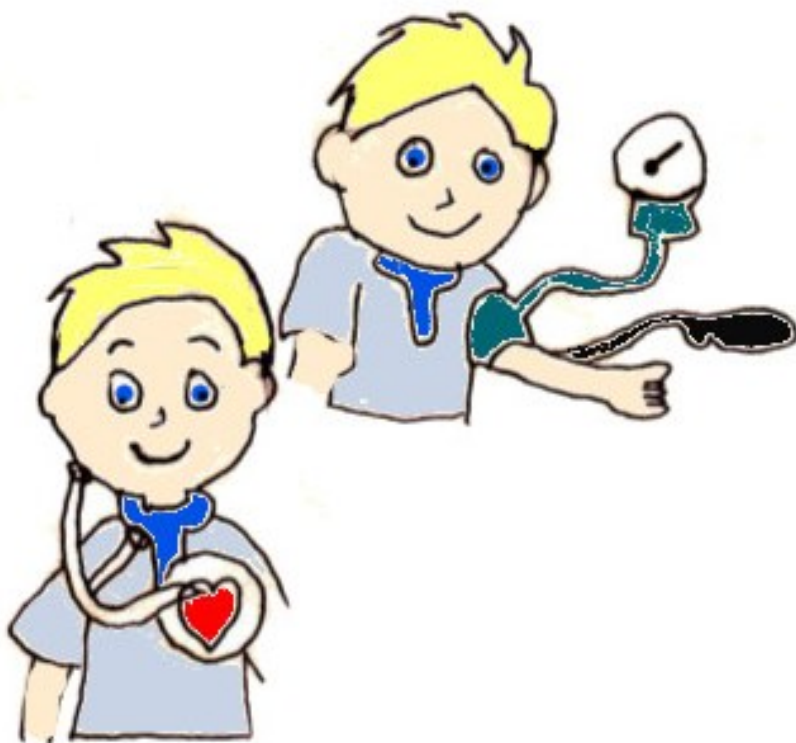
When I first had the Mickey put in, I went to hospital. They gave me medicines that made me sleep, and when I woke up it was in!


But now my mum or dad can change it at home every 6 months. I always get scared but they make sure it doesn't hurt by putting some magic cream on.

They also have to clean it every day and cover it with a plaster to make sure no bugs get in.




I sometimes have to go to the hospital to meet my doctor or a nurse who tells me what I need to eat to stay healthy. I have blood tests to make sure my **liver** is happy and that everything else is healthy. I get weighed and measured to make sure I'm growing well and sometimes I even have a tummy scan.






Not many people have GSD1a, but I know a few! Since I was young my mum and dad have been taking me to a fun weekend away in different places each year. They call it a conference. Here I get to meet lots of other children and adults with GSD.





Some of my friends have Mickey buttons just like me, and some also have a naso-gastric tube. It is great seeing lots of people around me drinking cornflour because it makes me realise I am not alone.

There are also lots of doctors there from around the world who want to make people with GSD better forever.



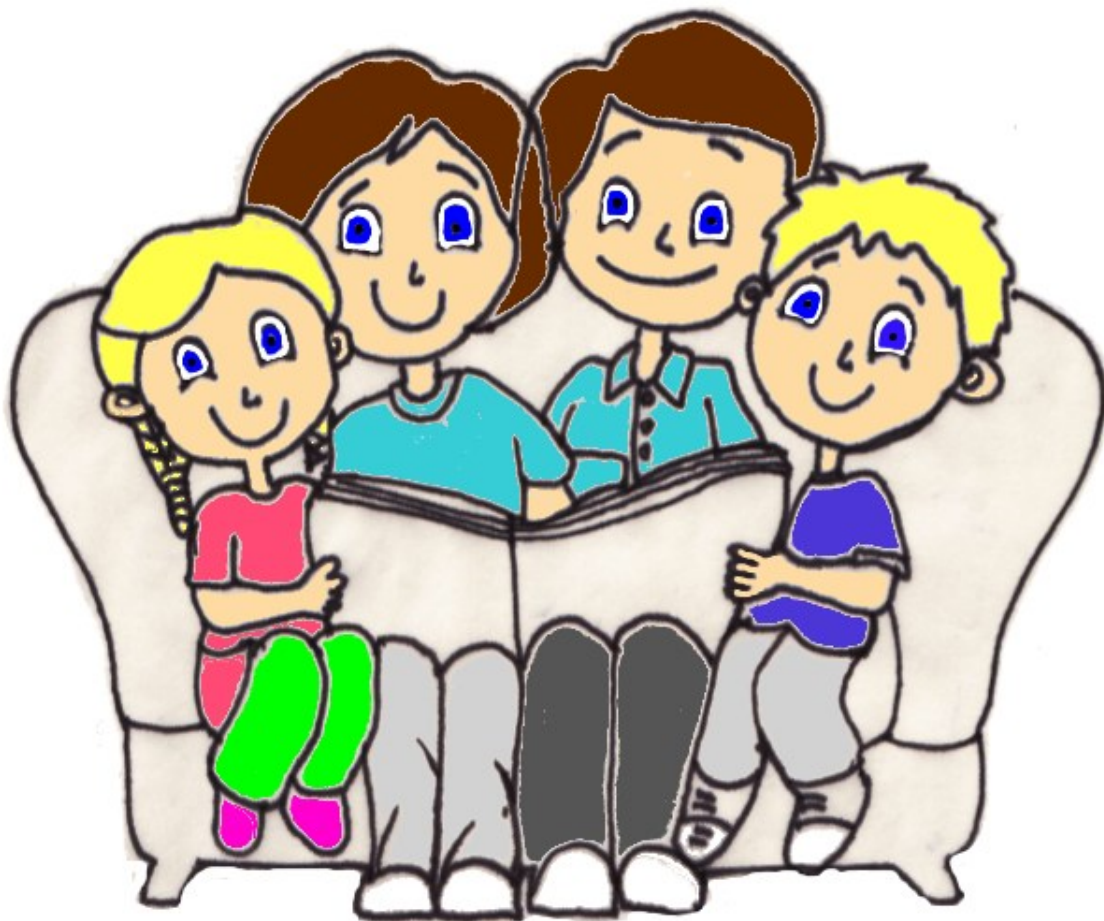



Living with my GSD can be hard sometimes. Because I don't look poorly to other people it is sometimes difficult to explain how important it is for me to eat the right foods at the right time. My doctor has given me a sheet of information to give to people like my teachers, which explains what it means to have GSD and what I need to do every day to stay well.






This is really helpful, but it is more for adults and my friends get confused if I show it to them. So instead I will show them this story. I read it with my mum, dad and Charlotte!





It is important to remember that not everyone with GSD 1a is the same. We all have the same condition, but our bodies deal with it differently. This means we all need to have slightly different treatments which are unique to us.

For me, I find that if I eat the right things at the right time, and go to see my doctors, I can do everything that my friends do. I play football everyday at school, and when I grow up I want to be a doctor to help other children just like me.






**I hope my story has
helped you to
understand a little bit
more about my GSD
and Me!**



'The author, Dr Gabriella Clark, is a junior doctor at the Royal Cornwall hospital. She has had a long standing interest in paediatric endocrinology since being diagnosed with type 1 diabetes. She was also greatly inspired by a friend who had GSD 1a, and who's courage never let the disease hold her back.'





There are many different types of Glycogen Storage Disease. To find out more about GSD, you can always contact us or visit the charity website...



Phone: 0300 123 2790

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Website: www.agsd.org.uk



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