

THIS LEAFLET IS NOT INTENDED TO REPLACE THE  
ADVICE OF YOUR SPECIALIST HEALTHCARE TEAM

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# MANAGING GASTROINTESTINAL (GI) SYMPTOMS

WHY GI SYMPTOMS HAPPEN AND  
IDEAS TO HELP DEAL WITH THEM

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## GETTING TO KNOW YOUR GASTROINTESTINAL (GI) SYSTEM



Your GI system, or digestive system, is an amazing part of your body. Did you know that it actually has its own set of thousands of nerves (called the enteric nervous system) that automatically control how the GI system works? It's almost as if the GI system has its own 'little brain'. So perhaps it's not surprising that the health of your GI system can affect how you feel emotionally as well as physically. It works the other way round too; it's well known that stress can make GI symptoms worse.

People with lysosomal storage disorders (LSDs) are more likely to have GI symptoms than other people. In fact, GI symptoms can be amongst the earliest symptoms of Fabry disease and often start in childhood.

It's not completely understood how LSDs cause GI symptoms, but it is thought that Fabry disease may disturb the nerves of the enteric nervous system. Individuals with Fabry disease may also experience discomfort and pain after eating due to the build-up of waste products within the cells of the intestine or the blood vessels and nerves supplying the intestine.

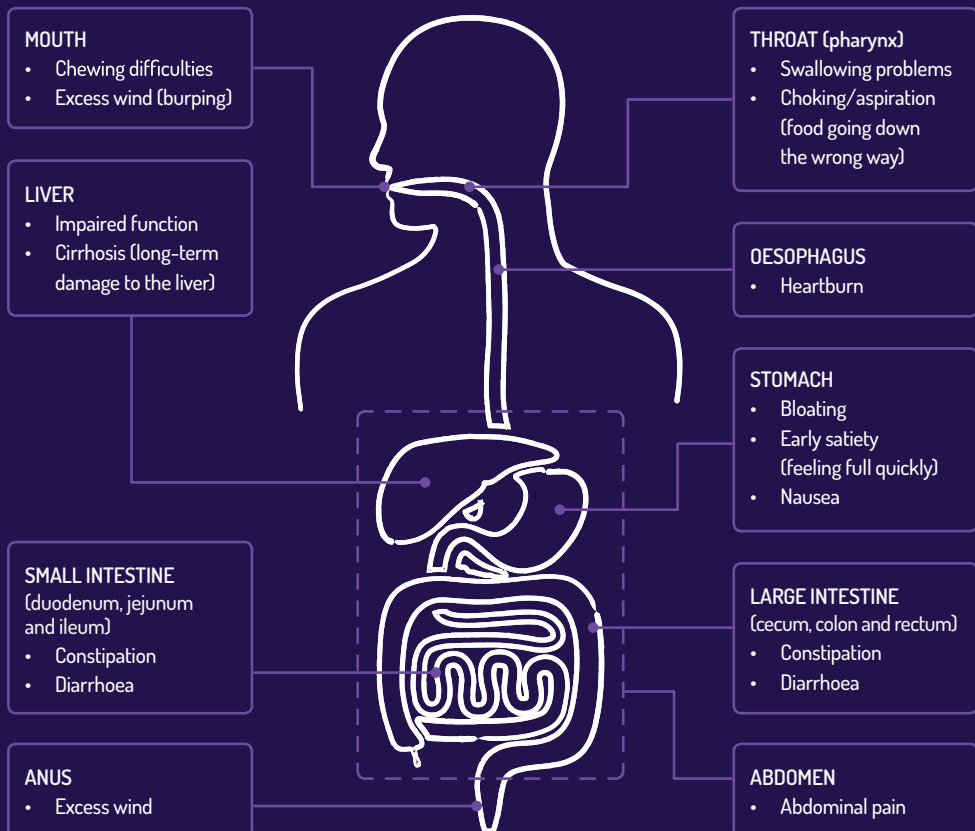
GI symptoms like constipation, diarrhoea and bloating are not only uncomfortable, or even painful, they can also be quite embarrassing to talk about. Having to rush to the loo when you're out shopping or visiting a friend's house, or spending what seems like an age sitting on the toilet waiting for something to happen are situations we could all do without.

The good news is that there are some things that can help to reduce these GI symptoms as you will see in the following pages. With the right strategies and advice, you can help control the impact of GI symptoms on your life.

# THE GI SYSTEM

Firstly, it can be helpful to understand a bit more about the GI system and what can go wrong with it. That might make it easier to talk to friends and family about what you're experiencing and to explain why you may be changing some of your food choices and eating habits. Having support from your friends and family will help to reduce any anxiety or embarrassment you may feel.

The diagram below shows the main parts of the GI system and common problems that can happen.



# COMMON GI SYMPTOMS

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Although not everyone with a LSD gets GI symptoms, some of the fairly common ones are outlined on this page. There are ways to reduce their impact on your daily life and your specialist medical team will be able to advise on the options that may be suitable for you.

## Incontinence and hydration

Both urinary and bowel incontinence are more common in people with Pompe and diarrhoea is also more common in Fabry and Gaucher diseases compared to other people, particularly when on a treatment for their condition. These can obviously have a major day-to-day impact.

Remember, even if you are concerned about urinary incontinence, our bodies are made up of two-thirds water so it is important to drink enough fluid to remain hydrated. Staying hydrated is important as being dehydrated can make you feel tired, give you a headache, affect your concentration and even make you feel dizzy or light-headed.

Bowel incontinence can be linked to either constipation or diarrhoea. You can be confident that your healthcare professionals will want to know if this is affecting you and they will want to help you find healthy ways to deal with it either through dietary changes or medication. You shouldn't be embarrassed to speak to them, it's common and GPs are used to seeing people that have difficulty controlling their bowels. So, if you experience any of these symptoms, you should speak to your doctor:

- You have sudden urges to poo that you can't control
- You soil yourself without realising it
- You sometimes leak poo, for example, when you pass wind
- It's affecting your daily life and you don't feel like you can socialise

There are also special exercises that can strengthen the muscles that open and close your bowel and there are ways to 'retrain' your bowel by following regular daily routines. The suggestions about constipation and diarrhoea on the next page may also help.

## Chewing and swallowing difficulties

People with Pompe and Gaucher disease can find their jaw muscles get tired easily and this can make swallowing and chewing difficult. These problems can lead to food 'going down the wrong way', which causes choking if they are big bits of food, and a situation called 'aspiration' if they are small bits that get breathed in. If you experience these regularly, it can be tempting to limit the amount of food you eat. However, that might mean you're not getting enough energy and nutrients from your diet.

Chewing and swallowing can be made easier by choosing foods with a slippery texture, or making chewy foods into a puree. Smaller, more regular meals may also help, as well as speaking to your speech and language therapist who can advise on how best to manage any problems you might have with eating.

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**"Constipation can be a problem but I don't take any pills. With bloating and diarrhoea I look to what I have eaten as that is usually the cause."**

**Patient with Fabry disease, UK**

# IDEAS TO HELP MANAGE GI SYMPTOMS

There are several things that people have found helpful in managing their GI symptoms. Remember, what works for one person may not work for the next, so try not to get disheartened if it takes a while to find the right solutions for you.

If you have access to a dietitian through your specialist healthcare team, they will be able to give you more details and personalised advice.

## IDEAS



### THINGS YOU CAN DO



Eating small and regular meals so that your body doesn't have to deal with too much food in one go



Try gradually increasing your fibre intake if you're constipated and eat less fibre if you have diarrhoea



Soluble fibre (that's fibre which becomes sticky when wet) is better than insoluble fibre; sources include oats, bran, barley, carrots and peeled potatoes. Insoluble fibre includes wholegrains and nuts



Sitting upright while eating and taking small mouthfuls may help reduce the risk of choking



Make one change at a time so that you can see what has helped



### THINGS YOU SHOULD AVOID



Drinking any more than three cups of coffee/tea a day



Drinking too much alcohol, the recommended daily allowance for men and women is two units of alcohol



Acidic citrus fruits like lemons, oranges and grapefruits as these can aggravate GI symptoms such as acid reflux and indigestion



If diet changes don't make enough of a difference to your symptoms, talk to your doctor to see if medication might be able to help

## Low FODMAP diets

This type of diet can help with those symptoms of Fabry or Pompe disease that are similar to irritable bowel syndrome (IBS). Basically, it involves avoiding foods that contain certain carbohydrates for a short period of time; usually six to eight weeks. Foods to avoid include wheat, some beans, some dairy products and some types of fruit.

If you follow this diet, it's important to have the advice of a dietitian to ensure you still have a healthy balanced diet with all the calories and nutrients you need. Most people will be able to see an NHS dietitian after being referred by a doctor. You might also find that an app will help you choose the right ingredients, track your progress and share information with

your dietitian. One example is *FoodMaestro*, which was developed in collaboration King's College London and Guy's and St Thomas' NHS Foundation Trust.

In case you're wondering, FODMAP stands for 'Fermentable Oligosaccharides, Disaccharides, Monosaccharides And Polyols', which are types of carbohydrate.

There are a number of FODMAP-trained dietitians in the UK; ask your healthcare team if they can refer you to one

# GETTING THE BALANCE RIGHT

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Whatever dietary changes you decide to make, the goal is always to have a healthy balanced diet with all the calories and nutrients that your body needs. You'll need to keep an eye on your body weight and nutritional intake to make sure you stay healthy overall.

## Being underweight

Body weight isn't just about what you look like. Being a healthy weight reduces the risk of a whole host of medical problems. For example, being underweight can contribute to a weakened immune system, so you would be more likely to catch a cold, the flu or other infections. Women who are underweight can also find that their periods stop.

If you lose weight quickly for no apparent reason, do speak to your doctor as soon as possible so they can help find out what's going on

Even taking actions to reduce GI symptoms, people with Fabry disease can experience weight loss and become quite thin. To make sure that your diet provides the right amount of calories for your age, height and how active you are, it may help to speak to a qualified dietitian.

## Being overweight

A body mass index (BMI) above the healthy weight range, or too much fat around your waist, can increase your risk of serious health problems like heart disease, type 2 diabetes, stroke and certain cancers. The best way to lose weight is usually through a combination of diet and exercise.

## Essential nutrients

When it comes to food, it's tempting to focus on things like taste and the effects on your GI symptoms and then forget about the little, but important, things like vitamins and minerals.

It's only through a healthy, balanced diet, and supplements when recommended, that you can ensure you have all the essential nutrients that your body needs to work properly.

Calcium, for example, is important for the maintenance of strong and healthy bones and iron is needed to prevent anaemia, which can leave you feeling drained and tired.





### **Additional information**

There are ways that you can make some of the small changes in your diet suggested in this leaflet while still looking after your weight and nutritional requirements.

Ask your healthcare team to put you in touch with a qualified dietitian to help you make the right choices about dietary adjustments.



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