

Additional information

You may have lots of questions after reading the content of this leaflet. It may be helpful to ask to see a dietitian from your own specialist unit, or speak to your consultant/nurse about how to obtain specialist dietary advice.

Remember, everyone is individual and what works for one person might not be appropriate for another. Getting personalised advice can set you on the right track towards a diet that meets your own health needs.

Additional information sources used in developing this guide

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- Dr Derralynn Hughes, Royal Free Hospital
- Association for Glycogen Storage Disease (AGSD)
- MPS Society



EATING WELL

AN OVERVIEW OF HEALTHY EATING AND SPECIAL DIETS



HEALTHY EATING AND PSYCHOLOGY

The main booklet in this pack described how a healthy balanced diet is recommended for everyone. Eating healthily involves balancing the different food groups. Your goal is to eat lots of fruit and vegetables, including a starchy, high fibre food with each meal (like potatoes, rice and wholegrain bread or pasta), and a smaller amount of protein and milk/dairy foods. Take a look at the 'Eatwell guide' in the main leaflet for guidance on balanced meals. Choosing foods lower in fat, salt and sugars is also a good idea.

For some people with a lysosomal storage disorder (LSD), adjustments to these principles can have other health benefits as we shall see in this leaflet. By making well-informed choices about your diet you may even feel more in control of other aspects of your life too.

Whatever changes you want to make to your diet, it can be helpful to first understand a bit about the psychology behind our eating habits. We often get into a rut and choose the same types of food. However, variety is the 'spice of life' and that's also true for a healthy diet.



TRY THESE TIPS TO HELP STICK TO A HEALTHY DIET:



Set realistic goals: Putting too much pressure on yourself might make you feel bad if you stray from your diet plan. Choose a few small changes and make them one at a time



Think about what matters to you: Remind yourself why you want to have a healthier diet. You could make a list and put it on the fridge door



Be kind to yourself: If you 'fall off the wagon' and eat something you're trying not to, forgive yourself and move on with a strengthened resolve to do better next time – after all, you're only human



Keep healthy snacks to hand: It's easy to make unhealthy diet choices when you're out and about. Try carrying healthy snacks with you to keep hunger at bay until your next meal



Be patient: Research suggests it can take around 66 days to turn your new behaviour into a healthy habit, so stick with it



Be prepared: Having a weekly meal plan and a shopping list can help you to avoid pitfalls like filling your shopping trolley with everything you fancy. Shopping when you're not hungry can also help

SPECIAL DIETS

"I used to find I was very tired in the mornings, but after experimenting with meal times, I found that having a small protein snack before bed meant I had more energy on waking."

Patient with Pompe disease, UK

Sometimes dietitians recommend special diets for people with a LSD. This might be to help with weight management or to reduce some common gastrointestinal (GI) symptoms such as bloating or constipation, as well as help to maintain your energy levels.

Fluid intake and urinary continence: Whatever diet you choose to follow, or is recommended for you, fluid is an essential part. It can be tempting to drink less than you should if you are worried about incontinence. Yet water is really important and dehydration can cause unpleasant feelings like dizziness and poor concentration, which can lead to falls. Dehydration can also increase the risk of getting urinary infections. The recommended amount to drink per day is 2 litres for men and 1.6 litres for women. If you are correctly hydrated, your urine will be a pale colour; if it is dark you are not drinking enough.

WATCH THE COLOUR OF YOUR URINE TO MAKE SURE YOU'RE DRINKING ENOUGH FLUID

Remember – healthy pee is 1 to 3, 4 to 8 you must hydrate



Dietary changes to manage GI symptoms: There are a number of things that can help if you have GI symptoms, such as bloating or constipation. These include eating regular and smaller meals; drinking plenty of fluid but limiting alcohol, fizzy drinks and caffeine; cutting down on rich or fatty foods; and cooking from fresh ingredients where possible. There's more information in the 'Managing GI symptoms' leaflet in this pack.

Adjusting how you eat could help you manage GI symptoms

Kidney-friendly diets: If your kidneys aren't working as well as they should, you might be advised by your doctor, nurse or dietitian to follow a diet that is low in sodium, potassium and protein. That's because these are processed by the kidneys, so following a low salt diet should put less strain on your kidneys. **You should always speak to your doctor or dietitian before you change your diet** to ensure you don't cut down on essential nutrients too much.

You can help your kidneys if they're not working as well, by limiting certain food types

Higher protein, lower carbohydrate diets: Sometimes this type of diet is recommended for people with Pompe disease. There is some evidence that adjusting the diet to contain higher amounts of protein, carbohydrates and fats can have benefits for the muscles and the balance of carbohydrate stores in your body. Those proportions are quite different from a 'normal' healthy diet, which tends to have less protein and more carbohydrate.

People with Pompe disease may benefit from a high protein, low carbohydrate diet

High energy diets: If you have an enlarged liver or spleen, you may find that you feel full more quickly. You may, therefore, need to consume food that is energy rich as your portion sizes might be smaller.

VITAMINS AND MINERALS

Your body needs vitamins and minerals in small amounts to help it to work properly.

Usually a healthy diet that has plenty of fruit and vegetables, some calcium containing foods (such as cheese, milk or soya milk), protein (such as fish, meat or a vegetarian alternative) and starchy foods (such as potatoes, bread, rice and pasta) will give people all the vitamins and minerals they need.

For some people, supplements might be helpful if they're not getting enough of a particular vitamin or mineral from their diet or if a health condition means they need more of a particular nutrient than it's possible to get through food alone.

Do ask your healthcare team for advice as although supplements are unlikely to be harmful, you probably won't want to take something you don't need – particularly if you have to pay for it! Also, some supplements don't mix well with certain prescribed medicines.

Iron: If you have anaemia, your healthcare professional may advise that you take iron tablets or increase the iron content in your diet. Foods that are high in iron include spinach, wholegrain cereals, pulses such as lentils and kidney beans and some dried fruits.

Look at the 'Iron leaflet' to learn more about your iron balance

Calcium: For people with a LSD, one of the most important minerals is calcium. This is because lysosomes act as calcium stores. Calcium helps to keep the bones and teeth healthy and it prevents osteoporosis (weak bones). Dairy foods and non-dairy foods that are fortified with calcium are good dietary sources. There is also calcium in broccoli, nuts and seeds. If you're not getting enough calcium in your diet, a supplement might be helpful.

You can get calcium from dairy foods (including the low fat options) or non-dairy foods fortified with calcium

Vitamin D: This vitamin works alongside calcium for healthy bones, muscles and teeth. Although there are foods rich in vitamin D like oily fish (salmon, sardines and kippers), eggs and fortified breakfast cereals, the main source of vitamin D is actually sunshine. You make vitamin D under your skin when you are outside in daylight. In fact, you might have heard of vitamin D being called the 'sunshine vitamin'. We make the most vitamin D from sunshine during the months of April to September. Over winter it is recommended that you take a supplement to make sure your body gets enough vitamin D.

You can get vitamin D from spending time in the sunshine or from supplements

Getting the balance right: It's a bit of a balancing act as you will need to watch calcium-containing or dairy products and it's advisable to limit your time unprotected in the sun to around 15 minutes two or three times a week.

If your healthcare team thinks you would benefit from calcium and vitamin D supplements, you might get a prescription. Make sure you discuss the options fully with your healthcare team, including what strength of tablets and daily dose you should take.

"I take a lot of vitamins and minerals which is quite costly but as I am self treating I need to make sure that I'm not lacking in anything."

Patient with Fabry disease, UK