NHS
Royal Papworth Hospital
NHS Foundation Trust

Cystic fibrosis transition information leaflet

A patient's guide

Compassion
Excellence
Collaboration

A big welcome from the CF dietetic team at Royal Papworth Hospital

You may have transitioned from your paediatric CF service or from another centre managing your care. This leaflet provides an overview of our team and role as well as what service options are available to you.

Contacting the dietetic team at Royal Papworth Hospital

Department telephone:
01223 639372

CF email address:
papworth.cfdietitians@nhs.net

Our department is open 08:30 – 16:00 Monday to Friday, if we cannot help you straight away, please leave a message on our answerphone or send an email and someone will get back to you as soon as possible.

Working with you

We are here to help you with nutrition and cystic fibrosis but we can only do this by working with you. As part of our role, we will be happy to explain reasons for our recommendations or help you find out more about your own health and nutrition, but this also requires taking ownership of your health and letting us know how we can help.

Accessing dietetic services at Royal Papworth Hospital

N.B. for pancreatic sufficient patients and patients who have had a transplant, the dietetic team review these patients only at annual reviews.

1. Your annual review

Every CF patient is invited to attend an 'annual review' which is your opportunity to discuss any concerns with various members of the multi-disciplinary team and an opportunity for us to monitor your progress. As part of this, you will have an opportunity to spend 30 minutes with a dietitian who will complete a full nutritional assessment and work with you to set nutritional goals for the coming year.

As part of your annual review, we also take hand grip strength measurements, more information on which is included later in this leaflet.

2. Virtual clinic

Our specialist dietetic assistant practitioner is available to speak to all pancreatic insufficient patients at their virtual clinic appointments which occur approximately every three to four months.

Prior to your clinic appointment, we send a pre-clinic questionnaire. This questionnaire enables us to appropriately triage your concerns so that you speak to the appropriate member of our dietetic team at your clinic appointment.

We also ask that you remember to have a current weight available prior to your clinic appointment.

3. Ambulatory care unit (ACU)

If you are coming into the hospital as an outpatient, you will usually be seen on the ACU. When attending a face-to-face appointment here, there will usually be the opportunity to speak to a dietitian, just let us know prior to your appointment and we can arrange for someone to come see you.

4. Inpatient admission

If you are admitted to Royal Papworth as an inpatient, you will be assessed by a dietitian and reviewed as regularly as required during your inpatient admission.

5. At other times

If you have nutritional / dietetic concerns but do not have a planned clinic appointment scheduled soon, please do not let this stop you seeking our help. Please call or email the team and we can arrange for one of the team to give you a call to see how we can help.

General diet advice

The advice below is some general guidance however, every individual is unique, and this advice is not necessarily appropriate for everyone; if you have any queries about this, please contact the team to discuss.

Much of this information is likely to be more appropriate for those eligible for modulator therapies such as Kaftrio, if you are not on a modulator therapy, we will work alongside you to provide tailored advice to your situation. You can find further resources at the end of this guide.

If you are pregnant, please let your medical team know as soon as possible so that all your care, including nutrition, can be adapted to your needs.

Hydration

Our body cannot function without water. Additionally, having adequate fluid in the body can help to thin mucus, making it easier to clear your airway and gut. It is important to drink even if you don't feel thirsty as thirst is a sign that your body is already dehydrated.

People with CF have higher requirements than the general population and fluid requirements are even higher in hot weather, when unwell or fighting infection, are exercising or, if you have diabetes, when there is a high amount of glucose in the blood.

Signs of dehydration can include thicker sputum, irritability, tiredness, headaches, dizziness and poor concentration.

Most people need at least two to three litres of water each day; this includes tea, coffee, water, milk, sugar free drinks and squash (NHS). If you would like further advice on your individual fluid requirements, please speak to the dietetic team.

Caffeine

Caffeine is found in many drinks – your morning coffee or tea, many carbonated drinks and even some foods. There is specific guidance around caffeine when you are pregnant; please speak to us if you would like to know more about this.

In general, consuming caffeine in moderation is not a problem although drinking excessive amounts, such as in energy drinks, can lead to palpitations, dizziness and other unpleasant symptoms. Caffeine is also a stimulant and therefore may make it difficult to sleep. If you are struggling with insomnia, potentially as a result of your medication regime, it might be worth considering reviewing the amount of caffeine in your diet.

Fibre and bowels

If you are taking pancreatic enzyme replacement therapy such as Creon or Nutrizym 22, we have additional information on this.

Please request a copy of this information from the dietetic team if this applies to you. Fibre is an essential nutrient for keeping bowels regular as it is used in the body to keep the gut functioning. Keeping your gut as healthy as possible is especially important for those with cystic fibrosis.

Additionally, a diet with adequate fibre has been shown to help reduce the risk of inflammatory disorders in the gut. Fibre is broken down in the large intestine by good bacteria; it can be soluble (as in fruit, cooked vegetables or oats) or insoluble (as in wheat bran or nuts).

All adults are recommended to get 30g fibre each day but on average people eat only 18g per day and often people with CF consume even less, so try and look for ways to boost your fibre intake. Foods rich in fibre include fruits and vegetables, wholemeal bread or pasta, nuts and seeds.

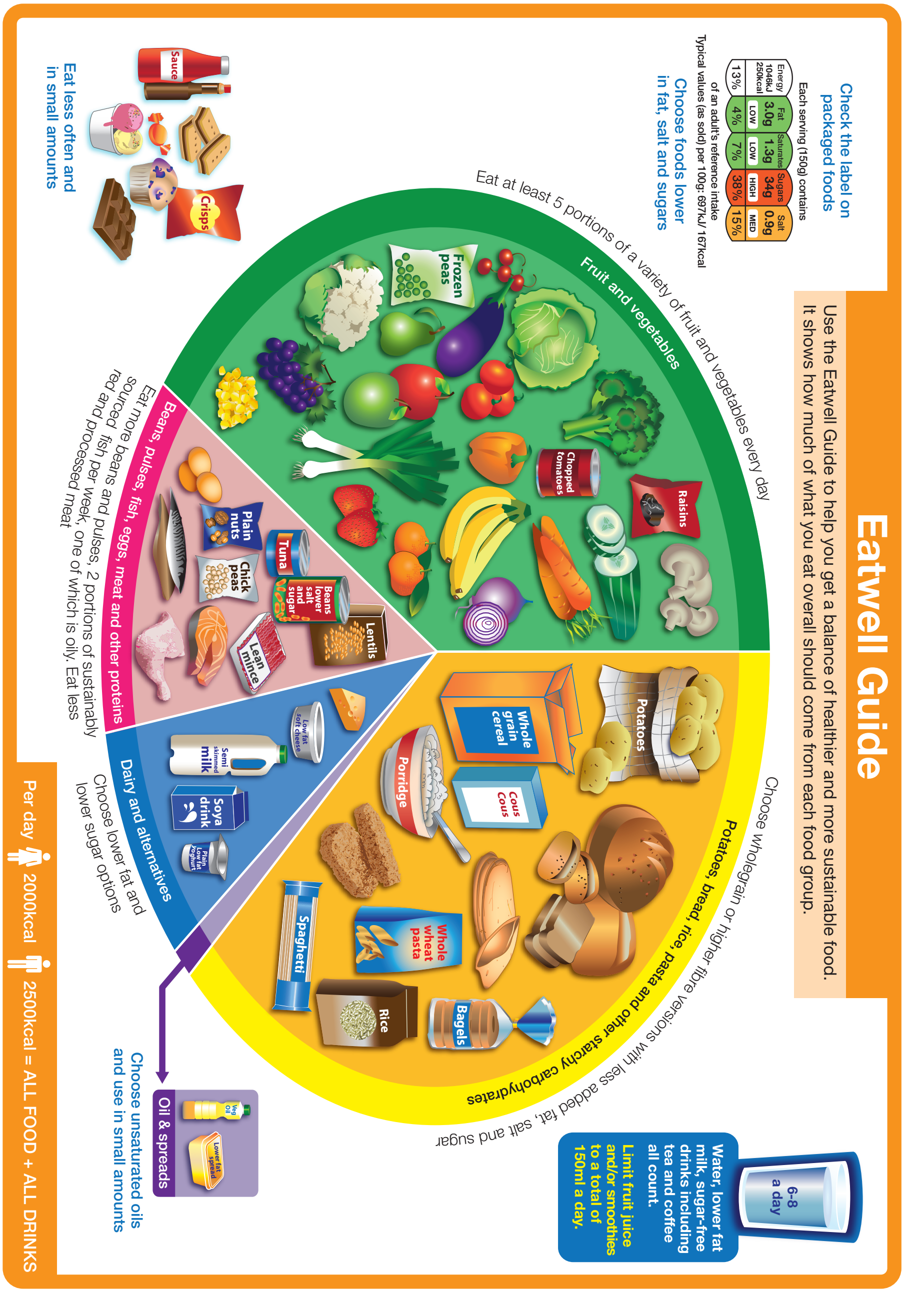
A 'healthy weight'

We advise to maintain a body mass index (BMI) between 20 and 25kg/m2. This is a general target that is not suitable as a goal for everyone, for example, those who are very muscular will likely have a BMI higher than 25kg/m2 and this does not mean they should look to lose weight.

You can calculate your BMI using many online tools including the NHS BMI online calculator or by using the following equation.

$$\frac{\text{Weight (kg)}}{\text{Height (m2)}}$$

If you are not sure what weight you should be aiming for or have concerns about your weight, discuss this with the dietetic team at your next appointment.



Source: Public Health England in association with the Welsh Government, Food Standards Scotland and the Food Standards Agency in Northern Ireland

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A ‘healthy diet’

Much like with weight, what constitutes a healthy diet for each individual is different. At different points of your life, you will have different nutritional needs which your Dietetic team can advise you on. For example, when you are unwell or fighting infection, it is often hard to maintain your weight and you might need to focus on energy dense foods – we have separate information on this if you require it.

There are some general principles of a balanced diet that are good practice to aim for in our diets.

One of the main recommendations is to follow the principles laid out in the Eatwell Guide. With day-to-day meals, we should aim to have about one third of our plate compromising of carbohydrates such as bread, pasta, rice and potatoes and one third consisting of fruits and vegetables. The other third should include protein such as meat, fishes, pulses and meat substitutes as well as dairy and dairy substitutes. Additionally, we should incorporate a small amount of fats, choosing unsaturated fats where possible (fats that are liquid at room temperature, such as oils).

A large part of a healthy diet is finding out what works for you for maintaining a suitable weight and ensuring you provide your body with the nutrients that it needs whilst enjoying food, including having some occasional treats. The Eatwell Guide lays out general principles, not an expectation of what a meal would consist of.

Since the introduction of Kaftrio, some are noticing an increase in weight unintentionally. This could be for many reasons such as an increased appetite, less frequent chest infections, longer periods of feeling ‘well’ or less issues within the gut resulting in improved absorption of food and drink. In order to remain at a healthy BMI, consideration of snacks should be taken into account e.g. opting for fruit and vegetables and if, suitable reducing the amount of high fat high sugar snacks. If you have any concerns regarding your weight, please contact us and we can provide you with some support.

Sodium

For those with cystic fibrosis who are not on modulator therapies, the advice is to follow recommendations set out by the UK Government regarding salt intake. Excessive and prolonged salt intake could lead to raised blood pressure and an increased risk of circulatory problems. However, some salt in the diet is important for nerve and muscle function.

For adults, we are recommended not to exceed 6g, or approximately 1tsp of salt per day. This includes salt already in foods, as well as salt added at the dinner table. Please check food packaging labels (the traffic light system red, amber and green) to see how much salt has been added before consuming.

Red meat and colon cancer risk

Studies have shown that patients with cystic fibrosis are five to ten times more at risk of colorectal cancer than the general population. You will be screened for this regularly as part of your cystic fibrosis care.

There are some things that you can do to help reduce your risk of developing colorectal cancer, including being mindful of your consumption of red meat which, when consumed to excess, is shown to be correlated to an increased prevalence of colorectal cancers. Red meat includes beef, lamb, and pork as well as products made with them such as burgers or sausages.

NHS advice is to reduce regular red meat intake to 70g or less each day. You don't need to exclude red meat from your diet as it is an excellent source of protein, vitamins, and minerals, but it is a good idea to be aware of how much you are consuming.

Hand grip strength monitoring

As part of your annual review, we ask to take a measurement of grip strength in each hand. This compromises squeezing a device called a dynamometer with your full force, repeated three times with each hand so that we can take an average to get as accurate a result as possible.

We use this reading alongside your weight to help contextualise weight change through muscle strength. An increase in grip strength alongside weight gain would indicate potential muscle gain whereas a stable or decreased grip strength alongside weight gain indicates likely gain of fat rather than muscle.

There is also research suggesting a link between a reduced grip strength and reduced respiratory function and bone density – this doesn’t mean there is necessarily cause for concern if your result decreases, but it is one part of forming an overall picture of your health that can help ensure the entire CF team provide you with the best treatment.

There are lots of factors that might increase these results. For example, grip strength rises and falls based on age; increasing exercise, especially strength training, will help develop muscle function, as will a diet that has an increased quantity of protein in. Equipment, mood and technique can also all have an impact.

When your gut does not feel right?

In people with CF, there is an increased complexity to, and likelihood of, gut issues.

Gut-related issues can include nausea, bloating, diarrhoea, stomach cramps, changes in colour or consistency of stools, and loss of appetite. There can be numerous causes of these and if your gut does not feel right for more than a few days, it might be worth contacting the team at Royal Papworth to try and identify the cause.

Some common causes of gut issues are:

- Pancreatic insufficiency and mis-dosing pancreatic enzyme replacement therapy
- Are you prescribed Creon, Nutrizym 22 or another pancreatic enzyme; if so, are you taking the correct dose at the correct time and in the correct way? Are they stored correctly?
- Constipation
- If you have not opened your bowels regularly or are experiencing stools that are very hard to pass,

your medical team might recommend some laxatives.

- Poor appetite and dietary changes
- Gut upset can be caused by not eating as much as usual or eating differently to usual. Even if you are feeling unwell and have lost your appetite, it is important to continue to eat, both for your gut health, recovery from illness, and overall health.
- Pain
- When we are in pain, this can often cause stomach and gut upset and changes.

If you are in any doubt about your symptoms, please speak to the medical team.

Further resources

cysticfibrosis.org.uk/sites/default/files/2020-12/Staying%20hydrated%20and%20cystic%20fibrosis%20July%202019.pdf

bda.uk.com/resource/fibre.html

nhs.uk/live-well/eat-well/food-guidelines-and-food-labels/the eatwell-guide/

bhf.org.uk/informationsupport/support/healthy-living/healthy-eating/salt

Sources

pubmed.ncbi.nlm.nih.gov/9059158/

sciencedirect.com/science/article/abs/pii/0012369215347231

ncbi.nlm.nih.gov/pmc/articles/PMC8622157/

gov.uk/government/publications/the-eatwell-guide

bhf.org.uk/information support/support/healthy-living/healthy-eating/salt

cysticfibrosis.org.uk/sites/default/files/2020-12/Salt%20in%20the%20CF%20diet%20A4%20June%202019.pdf

nhs.uk/live-well/eat-well/food-guidelines-and-food-labels/water-drinks-nutrition/

bda.uk.com/resource/coffee-and-health-it-s-not-just-about-the-caffeine.html

cff.org/managing-cf/about-colorectal-cancer

ncbi.nlm.nih.gov/pmc/articles/PMC4698595/

nhs.uk/live-well/eat-well/food-guidelines-and-food-labels/red-meat-and-the-risk-of-bowel-cancer/

nhs.uk/live-well/eat-well/food-guidelines-and-food-labels/water-drinks-nutrition/

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Large print copies and alternative language versions of this leaflet can be made available on request.

View a digital version of this leaflet by scanning the QR code.

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