

Cockayne syndrome

Getting the right nutrition and fluids

Growth in Cockayne syndrome

Children and young people with Cockayne syndrome (CS) have a different growth pattern due to the nature of their underlying condition.

They do not need the same number of calories as other children of the same age, the amount of protein they need may also be different.

Each child is treated individually and their height and weight plotted on the specially developed CS growth charts. This will enable the dietetic team to assess your child and formulate a feeding plan based on their individual needs.

Use this QR code to link to growth charts from the patient support group [Amy and Friends](#)



It is important to look at your child and their weight rather than their age when assessing the calories they need. The size of your child's stomach is about the size of their fist so this may help when thinking about how much they can manage for a meal or drink.

Giving more calories does not necessarily mean an increase in height or weight. Instead it can cause your child to be sick (vomiting) due to the small stomach volume.

Calories and nutrition

Research shows that giving the right number of calories is essential for the health and well-being of children and young people with CS. Giving too many or too few can be harmful. However, it is not just about calories, giving the right amount of other nutrients is equally important.

Type and texture

The type and texture of food that children and young people are able to eat can vary.

Sometimes dry foods can break off and scatter in the mouth causing coughing or difficulty chewing. Let your clinical team know if your child is coughing when eating. Also, if there is any change in the type of food or fluid your child is able to take. We can advise on how to continue to give food and drink by mouth safely.

We may recommend having soft, moist food or making sure food is cut up into small pieces. These suggestions may make it easier for your child to eat.

Feeding support

Many children and young people with CS will need support with feeding. Sometimes it is not safe for them to eat food or take fluids by mouth. This is often because their swallow may not be strong enough. There is a risk that food or fluid may go into the lungs.

This may mean that enteral feeding (using a feeding tube) is necessary to give your child the calories and nutrition they need.

Feeding tubes

In this information we use the term 'feed' when we are referring to the medicinal product which patients receive through their tube. By using this term we are consistent with widely used clinical terminology. This avoids potential confusion between the terms 'food' (usually taken by mouth) and 'feed' (taken via a tube), which could put patients at risk of harm if interchanged. If you have any questions, please speak to one of our team.

Nasogastric (NG) tube

This is a tube that passes from the nose into the stomach. We may recommend this as a short-term solution to ensure safe feeding. It should not be used to administer feeds or fluid overnight or unsupervised.

Gastrostomy tube

This is a tube that is placed directly into the stomach through the tummy (abdomen). We may recommend this as a long-term solution. A gastrostomy tube needs to be inserted under general anaesthetic, so your child is asleep when this is done.

It is important that the amount of specialist feed and/or fluid given is carefully calculated to prevent giving too much. This could cause your child to vomit. The calculations take in to account any food or fluid taken by mouth.

Feeds may be given slowly by a pump, or a little quicker by a bolus. This is done by using a syringe and allowing gravity to dictate the speed, and not by using the plunger. Wherever possible feeding overnight is avoided. This can disrupt the natural sleep cycle, interfere with how the body uses energy and may affect repair that occurs during this time.

It is important to remember that your child's weight is key to getting the feed and fluid intake right, not their age. The amount your child can tolerate may be different to that of another child with CS, so each regimen needs to be personalised.

It is advisable to start with a small volume and gradually increase to make sure your child can tolerate it. We recommend to set a baseline of about 10-20% higher than your child tolerated before the tube was inserted. This can be slowly increased until your child is receiving their optimum amount.

If healthcare professionals are not familiar with CS they may feel that a significant increase in fluid is needed. This may not be appropriate for your child. If further advice is needed please contact the CS team.

Poor tolerance of enteral feeds

Some children may not be able to tolerate the specialist feed/regimen that is initially prescribed. In this case there are some alternatives that can be trialled:

- A feed which has a different composition, for example the protein and fat is already broken down.

- If your child is having their feed via a bolus, using a pump that drips the feed in slowly might be beneficial.
- If the feed is still not tolerated a tube that goes beyond the stomach into the small intestine (a jejunostomy), may be required.

Additives

It is important to remember that it is not just about getting the amount of calories right. A lower calorie intake may mean other nutrients such as protein, minerals and vitamins are not present in the amounts needed to maintain health.

A multivitamin and mineral supplement or other supplements may be advised to ensure your child gets the right nutrition.

This leaflet was created by the multi-disciplinary team (MDT), in partnership with parents and Amy and Friends.

Contact us

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For more information on conditions, procedures, treatments and services offered at our hospitals, please visit [web](http://www.evelinalondon.nhs.uk/leaflets) www.evelinalondon.nhs.uk/leaflets

Evelina London Medicines Helpline

If you have any questions or concerns about your child's medicines, please speak to the staff caring for them or contact our helpline, [phone](tel:02071883003) 020 7188 3003, Monday to Friday, 10am to 5pm [email](mailto:letstalkmedicines@gstt.nhs.uk) letstalkmedicines@gstt.nhs.uk

Your comments and concerns

For advice, support or to raise a concern, contact our Patient Advice and Liaison Service (PALS), [phone](tel:02071888801) 020 7188 8801 [email](mailto:pals@gstt.nhs.uk) pals@gstt.nhs.uk. To make a complaint contact the resolution department [phone](tel:02071883514) 020 7188 3514 [email](mailto:complaints2@gstt.nhs.uk) complaints2@gstt.nhs.uk

Language and accessible support services

If you need an interpreter or information about your care in a different language or format, please contact the department your appointment is with.

NHS 111

This service offers medical help and advice from fully trained advisers supported by experienced nurses and paramedics. Available over the phone 24 hours a day,

phone 111

web www.111.nhs.uk

NHS website

This website gives information and guidance on all aspects of health and healthcare, to help you take control of your health and wellbeing, **web** www.nhs.uk

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Leaflet number: 5476/VER1

Date published: February 2024

Review date: February 2027

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A list of sources is available on request