



Dysphagia

Dysphagia is the medical term for difficulty in swallowing

What may happen

Niemann-Pick type C disease can weaken or damage the muscles and nerves used for swallowing. Difficulty in swallowing can be distressing for the person and those caring for them. Eating and drinking can become a challenge, as can taking in enough fluids and calories to nourish the body.

Signs that a person is having difficulty swallowing can include weight loss, dribbling and holding food in the mouth without chewing. Other symptoms include spitting food out, refusing food or drinks, and coughing or choking when eating or drinking.

You may also notice the person having difficulty in chewing or controlling saliva, liquids or food in the mouth. They may be unable to fully swallow, which can lead to coughing, choking or aspiration (when food 'goes down the wrong way'). Sometimes you may hear gurgling or wet noises once the person has swallowed.

At first you may notice that clear fluids can cause choking or coughing and this can be a sign that the swallowing reflex is not coping. Taking a drink through a straw may help to regulate the amount of fluid taken in. If coughing or choking starts to occur on a regular basis, seek advice from your speech and language therapist as soon as possible.



What you can do

As soon as you notice there may be a problem, speak to your speech and language therapist, or ask your GP or consultant to refer you to one. Speech and language therapists are trained to test for and to treat swallowing disorders. They will arrange to test the person's ability to eat and drink, and will look at alternative ways of swallowing that may help. You will be given advice about food preparation; for instance, those who cannot swallow liquids may need to add special thickeners to their drinks, others may have to avoid hot or cold foods or drinks.

If you have problems with swallowing, saliva and mucus can build up in the mouth and throat. This can be embarrassing if it results in dribbling and naturally many people feel upset about this.





Here are some suggestions that may help with this:

- think about your posture. A well-supported head position such as a reclining chair, chin support or headband can ease or assist with dribbling and/or coughing/choking
- adopting different sleeping positions can help. Try sleeping on your side or reclined rather than flat, as this can help to prevent saliva 'pooling' in the throat
- there are medications that can help by drying the mouth and reducing the amount of saliva
- clothing can be adapted to provide a discrete waterproof-backed insert and specialist 'scarves' are also available
- there may come a time when you need to consider using a suction machine rather like the one used by a dentist. A small tube attached to the machine is placed in the mouth and sucks out excess mucus and saliva. It is essential that the person operating the machine receives appropriate training
- using a barrier cream to protect the skin can also help

Some ways to ensure safe eating and drinking practices are:

- try to keep mealtimes calm and relaxed
- sit upright and keep the chin down
- consider different consistencies of food, such as puree or mash
- consider use of a thickener for liquids. These are available from your GP and your speech and language therapist will advise you
- take small sips of liquid or small spoonfuls of food
- ensure the mouth is clear before taking in more food or liquid
- you may find that mealtimes take longer – be patient





When to consider a feeding tube

For some people, consuming foods and liquids by mouth may no longer be possible. At this time, a feeding tube or system might be considered.

Although this is not an easy decision to make, most families find that a feeding tube can bring benefits and improvements to the affected person's quality of life. You could speak to the NPDG (UK) clinical nurse specialist for more information, or to a family in a similar situation for their thoughts and advice. The final decision, however difficult, is personal to you and your family and should be whatever you feel is right for you or your loved one at the time.

Once swallowing problems start, it may be an idea to think about having a feeding tube, even if the person is still able to eat. Generally, your healthcare team will recommend getting a tube earlier, to avoid choking, aspiration, dehydration or malnourishment. If meal times are taking more than an hour, or if the person has lost more than 10 per cent of their weight, the time to consider a tube is now.





Once a tube is in use, the person may still be able to enjoy food by mouth. You may want to give them small amounts of their favourite food, such as yoghurt or ice cream.

There are two methods of tube feeding to consider – a nasogastric tube (NG tube) and a gastrostomy tube, otherwise known as a PEG or G-tube.

For those who do not wish to consider surgical interventions, a nasogastric tube (or NG tube) may be the right option. The NG tube is inserted through the nose and threaded into the stomach. It is easily changed, usually once a month or sooner if the need occurs. The disadvantage is that the tube needs to be taped into place on the face, to avoid movement. This can be uncomfortable and can cause irritation. It is also important to check that the tube is correctly placed before feeding begins. NG tubes are usually used as a temporary measure. Most parents will learn how to handle this tube and to change it themselves after proper training.

A gastrostomy feeding tube is placed during a surgical procedure, while the patient is under general anaesthetic. A flexible, narrow tube is inserted through the skin and the stomach wall, directly into the stomach, during a short operation lasting about 30 minutes.





There are two main types of gastrostomy tube in use. These are known as a G-tube and a PEG (percutaneous endoscopic gastrostomy). Both of these will have a long piece of tube permanently attached. They can be held in place by a soft disc or a balloon just inside the stomach wall. These tubes are often used until the stoma – the surgical hole made in the stomach that the tube passes through – has fully formed, which usually takes about six months.

Once the stoma is fully formed the tube may be replaced with a ‘button’ which lies flat against the stomach, and requires an attachment to be added before feeding takes place.

Your community nurse will be able to show you examples of each option and explain in detail how they are inserted and how they work.

Once a tube is in place, it is important to keep the area clean and to follow guidelines for keeping the tube in good working order. Your local healthcare team will provide you with this information as well as providing detailed training. The NPDG (UK) clinical nurse specialist can also offer advice and support in this area.





You will also be offered advice from a dietitian or nutritionist, who will help you to create an individual feeding plan, ensuring the person receives the correct amount of calories and hydration.

The advantages of a feeding tube are:

- swallowing problems can be distressing for all and a feeding tube can remove the anxiety which can accompany mealtimes
- the person will be able to receive the correct nutrition/ medication without danger of choking or aspiration
- this method of feeding can reduce the risk of chest infections caused by aspiration
- comfort will be increased due to proper hydration and nutrition, which can also reduce bowel problems
- the person may still take food by mouth, if they are able
- energy levels can be increased due to proper nutrition and hydration

The disadvantages can be:

- anaesthesia can pose a risk in the later stages of the disease
- the tube must be cared for properly to prevent infection or trauma





How we managed ... one family's story

Very gradually we realised that our daughter, who had previously been a very good eater, was starting to cough or choke when given her favourite foods – spaghetti bolognese and yoghurt. For a while we were able to puree her food and add thickeners to her drinks. However, the time came when meals were taking well over an hour and becoming a difficult and challenging part of our day. After seeking advice, we decided it was time for a feeding tube. This was a difficult decision but worth it to see our daughter looking and feeling so much better with proper hydration and nutrition. We were also able to give her the medication she needed without causing any stress.

Who can help?

Your speech and language therapist is an expert in the function of the mouth and throat, and this includes eating and drinking as well as speech and communication. He or she can help to assess swallowing and offer advice and practical solutions. The NPDG (UK) clinical nurse specialist will be able to provide information and ideas that have worked for other families and will be able to discuss all of the options in detail.

Your dietitian will advise you about the best foods to help you stay nourished, and on consistency of foods, working closely with your speech and language therapist. Your GP will give medical advice and can prescribe treatments if needed. A pharmacist will be able to advise you as to which medication can be dispensed in liquid form and your occupational therapist will advise about posture and the equipment that can help.

This leaflet forms part of a resource pack published by the Niemann-Pick Disease Group (UK). It is intended to be read in conjunction with the other parts of the pack. If you do not have the other documents in the pack or you would like further information, please contact us at the email address below.

niemann-pick@zetnet.co.uk

June 2011

