AT society
awareness research care

Gastrostomies & Ataxia-Telangiectasia (“A-T”)

What is gastrostomy?
Deriving its name from the Greek word for belly, Gastrostomy is a surgical procedure by which a tube, having a sealable point of entry to the external wall of the abdomen, connects directly with the interior of the stomach. Once the procedure has been completed, the individual is able to receive all of his or her daily nutritional needs directly to the stomach via the tube, using pre-prepared feed packs. These commercially prepared feed packs provide a balanced diet including all the essential vitamins, minerals and dietary fibre that an individual needs. Although eating and drinking for pleasure and in social contexts may still continue as before, depending on the advice of your health care team, the individual will no longer be dependent on this as his or her sole source of nutrition.

There are several types of feeding tube: the most commonly used are the PEG (“Percutaneous Endoscopic Gastrostomy”) and the Mic-Key button. The PEG is a length of tubing with a valve; one end is attached to the stomach while the other protrudes a couple of inches outside the body. The PEG is usually inserted first and, when the site has healed well, can be replaced by a Mic-Key button, which is much smaller and fits snugly against the skin. The button looks very like the small valve that is found on a beach-ball or a child’s floatation aid. These gastrostomies are usually a permanent arrangement for someone who has A-T, although they can be easily removed if necessary.

Why might I need a gastrostomy?
The medical profession is increasingly recommending the use of gastrostomies in cases where there are chronic feeding, swallowing and nutritional concerns in people with neuromuscular conditions, such as A-T. There is, therefore, an increase in the use of this procedure in children with A-T in order to address the following problems:

- Poor appetite resulting in loss of weight.
- Difficulties with chewing and swallowing (dysphagia).
- Problems with choking and aspiration (food going down the “wrong way” and so reaching the lungs) leading to bacterial infections and possibly pneumonia.
- Poor nutritional uptake. A well-nourished body is better able to resist and to fight infections.

Physicians at the UK and US A-T Clinics are now recommending these procedures where they see a need.

Your health care team may assess your child’s need for a gastrostomy using “video fluoroscopy”, a method of filming and assessing the swallowing process using a barium meal. As you are aware, A-T is a disorder associated with increased sensitivity to radiation, however our medical advisers agree that a video fluoroscopy represents a low radiation risk.

The procedure

The procedure is considered to be a minor operation; however there is an elevated risk for people with neuromuscular conditions, such as A-T. The main constituent of this risk is in relation to the administering of anaesthetic where the respiratory system may already be weak. It is important, therefore, that these issues be fully discussed with your child’s health care professionals before proceeding. Where your health care team may not be fully aware of the elevated risks associated with administering an anaesthetic to a person with A-T, we recommend that you contact us at the A-T Society before proceeding with the operation.

Fitting the device is a surgical procedure usually requiring general anaesthetic and lasting about 30 minutes; (occasionally the operation may be performed under sedation, although this is rare). The most common method is for the surgeon to pass a tube with a light at the end (an endoscope) through the child’s mouth into the stomach. The surgeon then locates the light through the skin of the abdomen and makes a small (pen sized) incision, appropriately called a stoma from the Greek word meaning mouth. The tube is next passed through the stoma directly into the stomach and is then secured in place.

The procedure usually requires a hospital stay of between a few days and a week in order to give the child time to recover and to become accustomed to the gastrostomy. The body sometimes requires a little time to accept the tube and to tolerate the regular feeds. The whole process will be monitored and assessed by the health care team. There may be some discomfort after the operation but this can be alleviated using ordinary painkillers. Once the healing process is complete, the child will quickly become accustomed to the gastrostomy, hardly noticing it is there at all.

Living with a gastrostomy

The feeds are pre-prepared packs of liquid containing a balanced mix of nutrients. The external valve of the gastrostomy is opened and the feed is introduced directly into the stomach. There are two methods of delivering the feed: (a) in one bolus feed (a single dose given all at once, usually over about 20 minutes) using a large syringe, or (b) as a slow continuous delivery using an electric pump. Each method can be used on its own or in combination with the other, for example, bolus feeds may be given during the day and a continuous feed overnight. Most children with A-T receive their feed overnight using the continuous feed method. A dietitian will help to develop a suitable feeding regime and will recommend the type of feed to be used. Parents or carers will be shown how to connect the feed and how to respond to any problem.

Occasionally, the placement of a gastrostomy can cause gastro-oesophageal reflux – the flowing back of the contents of the stomach towards the mouth, (although for some people with A-T this may be an already existing problem). Where this happens, your health care team may recommend a procedure called a Nissen's fundoplication in order to tighten the muscles at the top of the stomach thus reducing the opportunity for reflux.

Some common questions

- ***Will I still be able to eat and drink normally?***
  Most, but not all, people with A-T who have a gastrostomy fitted can still eat and drink normally if they wish to. However your health care team may recommend that only certain types of food and drink are safe by mouth, particularly if aspiration is a problem. Liquid can also be given through the gastrostomy if needed.

- ***Will my friends know it is there?***
  Not if you do not want them to know.

- ***Can I still go swimming?***
  Yes. You can return to normal activities, like swimming. Just make sure that the external valve is closed. For girls, a one-piece swimming costume will cover the valve; for adults and older children a sticking plaster will provide a good disguise.

- ***Can it fall out?***
  This can happen (although it is rare). A nurse, doctor or even a parent can reinsert it quickly and easily. You will need to do this without delay as the stoma will begin to close naturally and reinsertion may become difficult after as little as four hours.

- ***Is it easy to keep clean?***
  Yes. To keep it clean and dry, use cotton wool and warm water to clean it and a towel to pat it dry. Patting dry is important as rubbing with a towel

will make the site sore. It is best to keep a towel just for this purpose as infection may be spread if family members share the same one.

- ***What will I feel while I am being fed?***
  You will probably not feel anything provided the feed is delivered at a rate to which your stomach has become accustomed, however if the feed is transferred too quickly you may feel sick. If this happens, the rate at which the feed is delivered should be reduced.

- ***Will I feel hungry before and full after a feed?***
  Yes. The stomach will still fill and empty in the normal way, giving the usual sensations of hunger and satisfaction.

- ***Can I still travel?***
  Of course. You can take the equipment with you and in some cases the feed can be delivered to your accommodation (as one family arranged before their holiday in Florida). You can also have a backpack fitted to the rear of your wheelchair so you can be fed during the day if required. You may, however, wish to consider taking or having delivered extra supplies in case you stay longer than planned. If you are taking a beach holiday, it is a good idea to cover the gastrostomy with a large sticking plaster to keep out any sand or sea water as these can irritate the site.

  If you are flying to your holiday destination, it is a sensible precaution to take some supplies as hand luggage in case your main suitcases go astray. If you do this, it would be wise to produce a letter from your local consultant explaining that you are carrying medical equipment because of your feeding needs. It might also be sensible to advise the airline in advance in order that all security issues can be addressed calmly.

- ***How much maintenance does it need?***
  Very little. It should be kept clean, but normal bathing or showering will be sufficient. Like most items in daily use, however, it will need replacing periodically. This is a very simple matter taking about one minute and can be done by a nurse or even a parent.

- ***Will it leak?***
  Most gastrostomies leak small amounts but not sufficiently to stain or mark clothing. This can usually be remedied with a little damp cotton wool. Because the fluid that leaks from the stomach can irritate the skin, it is important to keep the site clean and to apply a little protective cream.

- ***What prevents the food coming out?***
  The gastrostomy is fitted with a one-way valve, keeping the stomach contents in and the air out.

- ***Will I taste anything?***
  Because a person’s taste-buds are on his or her tongue you will not taste the feed because it is delivered via the gastrostomy.

- ***If I am not eating, do I still need to brush my teeth?***
  Yes. Even if you are not taking food by mouth, you should still clean your teeth twice daily and continue to have regular dental checks.

- ***How do I take medication?***
  When you are prescribed medication, remind the medical staff that you have a gastrostomy. Because tablets may block the tube, medicine in liquid form may be better. If you need to take medication in tablet form, ensure that it has been finely ground and mixed with cooled water that has been boiled. Tablet crushers can be bought from your local pharmacy. Always flush the gastrostomy well after taking medication.

- ***Can I still be sick?***
  In most cases a gastrostomy will not impair a person's normal reaction when the body needs to reject something in the stomach. However, where a person has had a Nissen's fundoplication this may not be possible. In these cases the contents of the stomach can be emptied through the gastrostomy.

Benefits of a gastrostomy

Those families whose children with A-T have had this operation are very positive about the benefits it brings, not just to the child but to the whole family.

- The child generally will have more energy (fatigue is a common problem in A-T) so he or she will be able to participate more fully in school and the whole family will be able to get out more often.

- Mealtime pressure on parents to have their child "eat up" will cease.

- Mealtimes will be shorter and will become social occasions rather than times of stress.

- As the child approaches a normal weight for his or her size and build as a result of improved nutrition, he or she will be able to fight infection more effectively.

- The opportunities to aspirate food and the consequential exposure to pulmonary infections will be considerably reduced.

Some Parental Perspectives

Family in Wales with 13 year old son:
"*The plus points of having a peg are that our son has lots more energy, has put on weight, looks healthier and we no longer have fights at dinner time to get food into him! He is also able to concentrate more on schoolwork. The negatives are that he gets upset at having feeds when out of the house (he*

thinks people are looking at him). He also wants to eat, but can't and gets upset seeing other people eating. Being sick is also a problem due to reflux and he has had thrush around the peg site due to antibiotics going through. I know there seem to be more bad points than good but over all I'm glad Alex had the peg fitted, as he is now a much healthier boy. I don't think we would still have Alex with us if he hadn't had the peg as he was literally wasting away from lack of nutrition".

Family in Scotland with 14 year old daughter:
"My daughter was always very thin and continually at the doctor's because I was worried about her weight, she also suffered from reflux. Finally at 7 years old she had her gastrostomy tube fitted, this was a turning point and her weight gain has been great. All the family have seen a big difference in her energy and she looks healthier. We see lots of good points to having this done and it has been a lifeline for her. The only problem is that sometimes the area around the skin gets red, but cream helps this. I would say to other parents if your child is underweight please think seriously about having a peg tube. As a first step we tried having a tube through her nose to the stomach to see if any weight was gained first, this is at least worth a try".

Daughter's point of view: *"When I had my tube fitted I only weighed 12kg, I was unable to eat much because I had a reflux as well. The tube has been a great help as I now weigh 34kg. I feel more able to do things all day and don't get as tired; I also eat what I want to and drink as well. The good thing is that I can get fed all night and wake up with energy to enjoy my day. I haven't come across any bad points".*

Family in West Midlands with 14 year old son:
"*Obviously this is a big decision for parents to make on their child's behalf. In our case we would not have continued without our child's consent. It really has made a vast improvement on his health. He is not as tired as before, he eats meals with the family (without the pressure of having to 'eat it all up') and without it taking 1-2 hours. No it's not all plain sailing – you have to be prepared to change feeds in the middle of the night and change the bed when the tube comes off. The results of our case far out-weigh these minor inconveniences. We have recently been on holiday to France and during the journey Joe was having his feed. When we arrived we were all starving but Joe had already eaten!"*

Family in Kent with 10 year old son:
"I was very nervous before and found the thought of tube feeding George very daunting. At first he was shy about his tube but after about six weeks he was going outside in the hot weather with no top on! The training we got from the nurse who visited us from the company that delivers the food was excellent and we all took to it straight away, even Francesca who is 6 can hook the feed up to George. It is important to get the whole family involved and then it becomes part of your 'normal' everyday life. At first George was only sick occasionally, but after about seven weeks the sickness was terrible, everyday in the mornings up to lunch times 3 or 4 times. We were really worried but

spoke to another family who knew exactly what I was going through and told me it was a probably a build up of saliva that was causing the sickness and suggested using 'hyocin' plasters behind the ears. They have been fabulous, the difference is unbelievable. The sickness has stopped; he has put on weight and has so much more energy. We also found out from other parents that toothpaste can start the sickness off so I have put George back on baby toothpaste which seems to be working. I must say although the last four months have been hard we are so glad George had the Gastrostomy fitted, it has changed his life totally".

If you would like to have further information on Gastrostomy or if you would like to discuss any of the issues raised in this leaflet, please contact us at the A-T office.

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