A guide to understanding

Anaesthetic management

Society for Mucopolysaccharide Diseases
www.mpssociety.org.uk

In this guide

This guide concentrates on the procedures involved in having a general anaesthetic and special considerations for individuals with MPS or related diseases

Introduction

The first time a child has an operation or investigation under general anaesthetic is a very anxious time particularly if those involved, for example, parents or carers, have no previous experience or knowledge of the process. This anxiety is particularly heightened where a disabled child or adult is concerned and parents are often essential members of the caring team.

This booklet is produced by the Society for Mucopolysaccharide Diseases (the MPS Society) drawing on the experiences of parents and doctors with reference to medical literature. Its aim is to provide some of the background information that parents need in order to make informed decisions about anaesthesia and what care will be required after the operation or investigation. Carers and professionals working with affected individuals and their families may also find this booklet helpful.

What is anaesthesia?

Anaesthesia means the loss of feeling, particularly the sensations of pain and touch. A local anaesthetic numbs only the relevant area of the body so that the individual does not feel pain, but remains awake and aware of what is going on. During a general anaesthetic the individual will be unconscious.

Before the anaesthetic

Giving consent

The Consultant and members of the medical team will explain what is planned for the operation or investigation. Parents or guardians will be asked to sign an official form of consent. You should feel free to ask any number of questions so that you are certain you understand and agree with what you are signing. Generally, children aged sixteen or over, and who are able to understand, may choose to sign the consent form themselves. Younger children should be involved in the process of giving consent whenever possible.

Meeting the Anaesthetist

The Consultant Anaesthetist is responsible for deciding the best method of anaesthetising an individual. The Consultant or a member of the team will see the individual beforehand and will prescribe any medication needed to prepare for the anaesthetic. If an individual has had an anaesthetic on a previous occasion there may be useful information which you could give to the anaesthetist. For example, some children are afraid of injections, some hate the smell of gas. If it is possible, these views will be taken into account, but the anaesthetist must make the final decision. Particularly in the case of young children, parents or guardians may accompany their child to the anaesthetic room and remain until he or she is asleep. If you feel this would help your child you should discuss this with the anaesthetist.

Specific problems for children with an MPS diagnosis

Anything which makes it difficult for the anaesthetist to maintain an open airway will increase the risk of having a general anaesthetic. Many children and adults with MPS and related diseases have short necks, limited jaw and neck movement, thick tongues, airways obstructed by storage of mucopolysaccharides, uncontrolled secretions, unstable cervical spines (particularly those with MPS IV) and floppy tracheas (windpipes).

Children with restricted breathing or with heart or lung disease may not cope very well with the anaesthetic and its after-effects. There may be difficulties inserting a needle if the child or adult has very thick skin or poor veins.

Risks will be dependent on the age, type of condition and the general health of the individual prior to the procedure, but all risks, either minor or more serious, should be discussed with the anaesthetist prior to proceeding.

Nil by mouth

Children must think nurses are being especially cruel when they say 'no breakfast today'. However, there is a very good reason for this. There is a risk that the child might vomit during the anaesthetic and that any stomach contents could get into the lungs. You will be told when the fasting should begin but this is usually at least four to six hours for solids and milk but many centres now allow clear fluids, such as water and squash till an hour before surgery.

Pre-medication

Pre-medication is the medicine that is given while the individual is on the ward, about an hour before the anaesthetic. This will vary with the age of the child and the type of procedure. The pre-medication can be either a sedative, to make the child sleepy and relaxed, or a drug to dry up the mouth and throat, which makes the anaesthetic easier to give, or it can be a mixture of the two. Pre-medication is usually given as a drink, but in special circumstances it may need to be given as an injection into the thigh or bottom.

If your child is extremely anxious you should discuss the possibility of a pre-med with the anaesthetists. A reduced dosage of pre-med may be possible.

Anaesthetic cream

If the anaesthetic is to be given by injection (usually in the back of the hand), anaesthetic cream may be put on the chosen place an hour before the operation and taped on. This will numb the spot so that the individual will not feel the needle going in. Unfortunately, the cream only numbs the skin and does not help with the intramuscular injection sometimes given as part of the pre-medication.

Gowns

Gowns which tie down the back are often put on when the child has been given the premedication. Sometimes children can wear their own clothes and they should be allowed to keep their underwear on at least until they are unconscious.

Waiting

Pre-medication is likely to make the mouth feel dry and some individuals are rather irritable until it has had a chance to work properly. Children and adults can also become suddenly wobbly on their feet, so it is safer for them to rest on their bed or someone's lap whichever is appropriate. If a child falls asleep, it is better for him or her to lie on the bed or to be held in a horizontal position, as blood pressure can drop when the child stays upright.

If parents are not going into the anaesthetic room it would be a good idea to discuss with the nurse when would be the best time to say goodbye. This will probably be on the ward to avoid any last minute upset, particularly for children, as they go through to the anaesthetic room.

During the anaesthetic

Going into the anaesthetic room

A nurse always accompanies the individual. A trolley will be brought, but if your child is awake and small enough, you could carry him or her to the anaesthetic room. A favourite teddy, dummy or blanket could go too, but it would be an idea to bring it back with you for safe-keeping.

Before going into the theatre area you may be asked to put on a gown or overshoes. The anaesthetist will have told you how he or she will send your child to sleep. Sometimes an injection in the hand is used, sometimes the patient is asked to breathe in anaesthetic through a mask. Small children can sometimes be anaesthetised on their parent's knee by the gas tube held near their face.

You will be asked to leave when the patient is asleep. It is very important to go as soon as you are asked as the anaesthetist has a great many things to do very quickly to make sure the individual is safely anaesthetised.

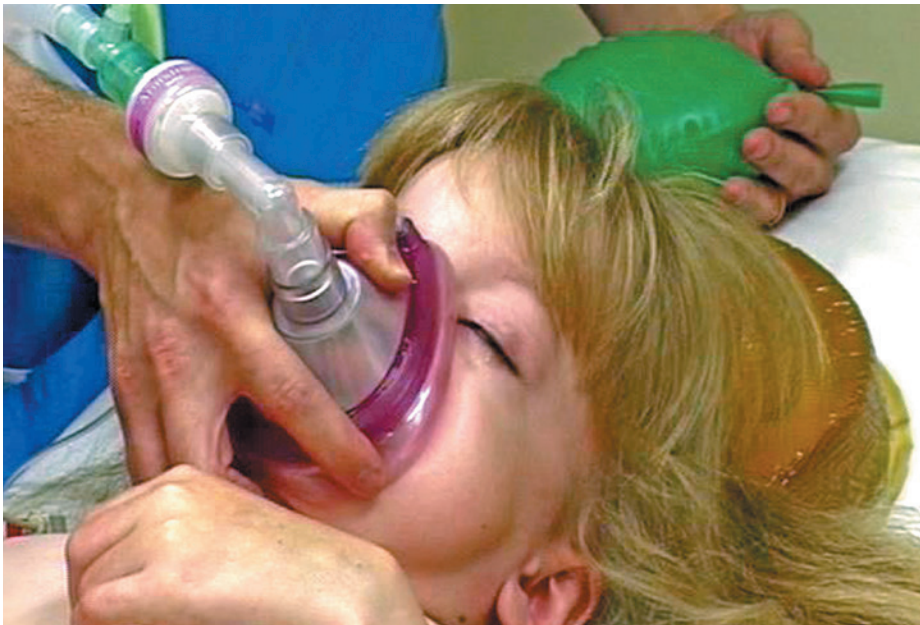
While your child is in theatre

The nurse will have told you how long the operation or investigation is likely to take.

Many parents like to go for a walk or to have a meal, but remember to make sure you let the nurse know where you are in case you need to be contacted for any reason. If the individual is to go to intensive care afterwards, parents will probably be taken to see the ward beforehand.

Many operations take longer than planned and patients usually spend a period of time in the recovery room before going back to the ward. The nurse will check how a child or adult is doing if parents are getting worried.

Anaesthesia means the loss of feeling, particularly the sensations of pain and touch



After the anaesthetic

Back on the ward

After an anaesthetic patients will need to sleep for a while, but even if he or she seems drowsy and unaware, hearing a familiar voice will help them relax and sleep more deeply. The nurse will inform parents when it is safe for sips of water to be given.

Surgeries involving general anaesthetic for individuals with MPS should not be performed without having a bed available in the intensive care unit (ICU). Some affected patients with severe forms of the disease or those requiring major surgery will require critical care management post operatively. Many patients, however, may not require this level of care.

Pain management

After the operation patients will probably need some pain killers but this depends on the type of surgery. A local anaesthetic may have been used in combination with the general anaesthesia to numb the wound. Simple pain killers such as paracetamol can be given by mouth or as suppositories (up the bottom) if the individual is feeling sick or is not able to take medicine by mouth. Stronger pain killers may be needed after major surgery, which will usually need to be given by injection, either directly into a vein or into the leg or bottom. Sometimes, if the patient has severe airway difficulties, only simple pain killers or local anaesthetics can be used safely. They may need intensive care if stronger pain killers are required. Following more major surgery pain relief can be delivered by a continuous morphine infusion or a patient controlled analgesia pump. The most appropriate form should be discussed with your anaesthetist before the procedure.

Box 1: The usual procedure for anaesthetising a patient

1. The individual is sent to sleep by the use of gas or by a drug given through a needle.
2. A muscle relaxant is introduced which paralyses all the muscles including the breathing mechanism.
3. Oxygen is given by squeezing a bag linked to a mask as a temporary measure while a tube is inserted.
4. An instrument known as a laryngoscope is passed over the tongue and down the back of the throat so that the anaesthetist can find the entrance to the larynx.
5. An endotracheal tube is introduced into the larynx and the laryngoscope is removed.
6. The tube is connected to a machine which keeps the lungs inflated with oxygen for the duration of the procedure.

Considerations for individuals with an MPS diagnosis

Planning an anaesthetic for a child with an MPS diagnosis

General anaesthetics should always be given in hospital by a skilled anaesthetist. For planned procedures there should always be a careful assessment of the child involving the anaesthetist.

In order to understand the problems it is helpful to know something of what is involved in anaesthetising an individual. The risks of anaesthesia must be weighed against the advantage to be gained from the surgery or investigation. See Box 1 for the usual anaesthetic procedure.

Specific problems in anaesthetising a child or adult with MPS

The maintenance of an open airway may be very difficult and is frequently a problem for the individual even during normal sleep. With the additional relaxation produced by general anaesthesia the obstruction increases. The usual way of dealing with this is to pass a tube through the larynx but this may be very hard to manage.

The preference of most anaesthetists will be to anaesthetise the child with a face mask, introducing oxygen and the anaesthetic gases by this method. The patient will then breathe himself or herself off to sleep and this usually takes between 1–2 minutes. Following that the anaesthetist can then assess the difficulty of the airway and assess the technique s/he will perform to secure the airway with the endotracheal tube.

What can be done?

If an individual is likely to be at risk during an anaesthetic, it may be safer for it to be carried out at a major hospital which has experience of treating individuals with MPS and related diseases, even if it necessitates travelling and inconvenience. It may be necessary for an individual to be on a ventilator for a while following surgery and an intensive care unit with good paediatric cover in the case of children will be essential. Day case surgery will not be suitable for complicated patients, even when having routine operations.

For planned procedures there should always be a careful assessment of the child involving the anaesthetist

Any worries about proposed surgery can be discussed with a GP, consultant or paediatrician who may suggest seeking a second opinion.

Make sure, as tactfully as possible, that the anaesthetist is aware of the individual's condition and possible problems. There are steps that can be taken to make the process safer, such as avoiding particular anaesthetic drugs or stabilising the neck of patients with cervical spine problems. For some procedures a local anaesthetic could be used instead.

You should inform schools and any other carers if there is an anaesthetic risk so that this information is passed on in an emergency.

Adults

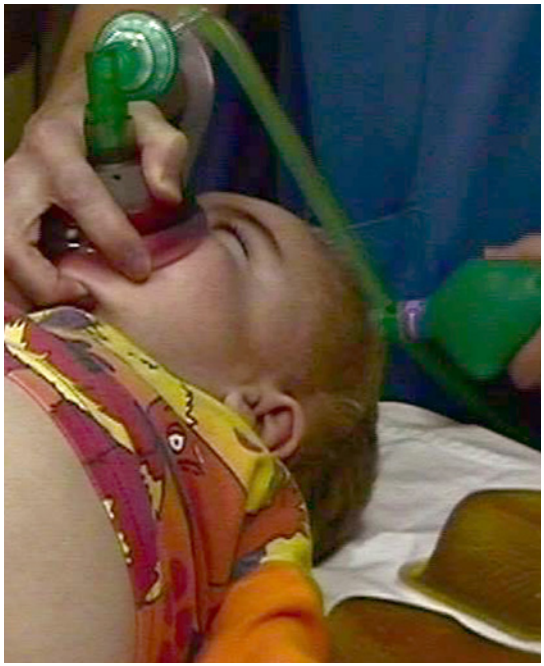
Many of the problems experienced by individuals with MPS progress with time. However, it can be advantageous that older children and adults may become more co-operative. Procedures that in childhood were necessary under general anaesthetic may instead be performed under local anaesthesia when the airway does not have to be interfered with. In these circumstances, mild sedatives may be prescribed to keep the patient as calm as possible.

Procedures that in childhood were necessary under general anaesthetic may instead be performed under local anaesthesia

It is important that adults should be treated in an adult setting but, due to the specific complications associated with MPS conditions and the subsequent high risk during general anaesthetic procedures, it is vital that all those involved have sufficient knowledge and expertise to deal with the implications. Seeking advice through consultation with experts in this particular field to ensure the best level of care for the patient is highly encouraged.

If your anaesthetist does not often deal with adults with MPS (as paediatric anaesthetists usually have more experience of patients with MPS), you should make sure that your anaesthetist understands the implications of the condition.

This booklet highlights a range of problems which can occur. Individuals with an MPS or related disease will not necessarily be affected by any of them. Many such children and adults have had successful surgery which has great improved the quality of their lives.



Checklist

Before surgery

- [ ] Make sure that hospital staff are aware of possible airway obstruction and anaesthetic issues
- [ ] Contact numbers for specialist centres have been given
- [ ] An assessment has been carried out involving the specialist team and anaesthetist
- [ ] The anaesthetist is aware of the risks and is prepared
- [ ] The surgery is taking place in the right hospital

After surgery

- [ ] A bed is available on ICU for after the procedure
- [ ] You have been informed how the surgery went and if there were any complications
- [ ] Information has been shared about any issues or complications that could affect future surgeries

Don't be afraid to ask questions or object if you feel there is risk or you need more information.

What the MPS Society does

The Society for Mucopolysaccharide Diseases is a registered charity, founded in 1982, which represents from throughout the UK children and adults suffering from MPS and related diseases, their families, carers and professionals.

The Society's advocacy team provides a unique, needs-led, individual advocacy service to individuals suffering from MPS and related diseases, their families and carers. We also raise public awareness and promote and support research into MPS and related diseases.

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