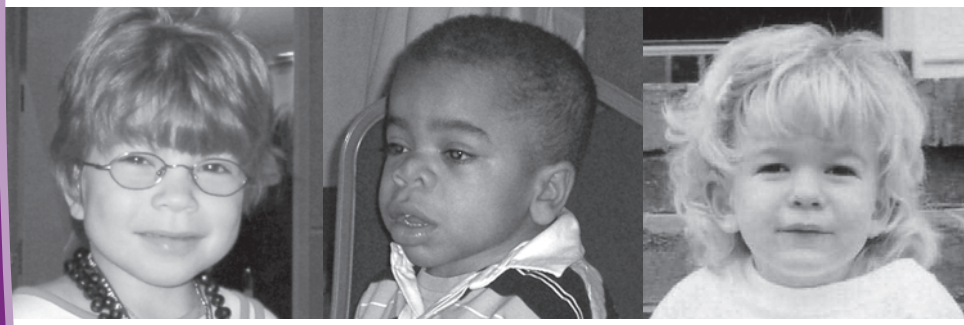
National MPS Society

Support for Families. Research for a Cure.



Understanding MPS and Related Diseases

Is Your Child Having an Anesthetic?

The National MPS Society exists to find cures for MPS and related diseases. We provide hope and support for affected individuals and their families through research, advocacy and awareness of these devastating diseases.

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Pictured on the cover: TAYLOR (MPS I), JONATHAN (MPS II), MACKENZIE (MPS III)

Introduction

An anesthetic may be required to safely perform various treatments and surgical procedures offered to manage the symptoms of MPS and related diseases. If your child is scheduled for a treatment or procedure and will be having an anesthetic, you may be wondering what happens. This booklet gives an overview of what anesthesia is, and addresses the potential problems relevant to individuals with MPS who receive anesthesia. It explains what can be done to reduce the risks of anesthesia and concludes by highlighting what happens on any routine operating day.

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Parents are essential members of the team caring for their child, particularly when that child has a disability. The information provided will help you to make informed decisions about anesthesia, and assist both you and the individual having surgery in preparing for the anesthetic experience.

Overview

What is anesthesia?

"Anesthesia" means a loss of feeling, particularly to the sensations of pain and touch. The term refers to the use of medication that temporarily suppresses these sensations during surgery. There are two main types of anesthesia:

Local anesthesia

- Only the relevant area of the body is numb and experiences no pain.
- In "regional anesthesia," a type of local anesthesia, an entire region of the body is made numb by injecting local anesthetic medication around major nerves that go to that area.
- The individual does not feel pain in this area, but remains awake and may be aware of what is going on. Sedative medications are often given to individuals receiving a local anesthetic helping to minimize awareness and create a state of relaxation and sleepiness.

- No breathing assistance is required.
- Local anesthesia is sometimes combined with general anesthesia.

General anesthesia

- Intravenous (IV) medications medications and inhaled anesthesia gases are used to "put the individual to sleep" before surgery begins.
- The entire body is numb and experiences no pain.
- The individual remains unconscious and unable to move for the duration of the medical treatment.
- Mechanical help is required to breathe. To assure safe oxygen levels while the individual is unconscious under general anesthesia, the airway leading from the mouth or nose to the lungs needs to be kept open. The typical way to manage this is to pass a breathing tube, known as an endotracheal tube, through the mouth into the larynx (voice box) and trachea (airway to the lungs).

MITCHELL (MPS VI)

- The endotracheal tube remains in place during the procedure, but it is usually removed at the end of the procedure as the individual awakens.

This booklet will focus on the procedures involved with general anesthesia. Certain aspects of local anesthesia also will be explained.

Normal procedure for general anesthesia

To understand how MPS has an impact on the delivery of general anesthesia, it is best to first review the basic steps involved in administering a general anesthetic. They are as follows:

1. Anesthesia gas delivered by a face mask or a medication given by injection is used to make the individual go to sleep.
2. A muscle relaxant is introduced which paralyzes the muscles, including the muscles used to breathe.

3. Oxygen is given by squeezing a bag linked to a face mask. This is a temporary means of providing oxygen until a breathing tube (endotracheal tube) is inserted.
4. An instrument known as a laryngoscope is passed over the tongue and down the back of the throat so the anesthesiologist can view the entrance to the larynx (voice box).
5. The endotracheal tube is put through the larynx into the trachea and the laryngoscope is removed.
6. The endotracheal tube is connected to a machine (ventilator) that breathes for the individual during the procedure.
7. The individual is continuously given anesthesia gas and oxygen through the endotracheal tube to maintain unconsciousness.
8. At the end of the surgical procedure the anesthetic is stopped. The individual awakens as the anesthetic begins wearing off.
9. The endotracheal tube is usually removed before the individual is fully awake.

What is different for individuals with MPS?

The impact of the underlying disease

Anything that makes it difficult for an anesthesiologist to perform the steps required to deliver anesthesia will increase the risks associated with a medical intervention. In individuals with MPS, the effect of the storage of glycosaminoglycans (GAGs) in many soft tissues and organs, as well as the effects on bone formation, can create difficulties. For example:

In individuals with MPS, the effect of the storage of GAGs in many soft tissues and organs can create difficulties with anesthesia.

- The storage of GAGs narrows the nasal passages; enlarges the tonsils, adenoids, lips and tongue; and causes loose, extra tissue and thick secretions to form around the larynx. These problems have the collective effect of causing airway obstruction, which can present as snoring when the individual is asleep.

When severe, it may cause *obstructive sleep apnea* where the individual stops breathing for short periods while sleeping. These airway problems decrease the anesthesiologist's ability to view the larynx, and can make insertion of the endotracheal tube very difficult.

- The muscle relaxation produced by anesthetic pre-medications and/or by general anesthesia leads to further obstruction of the airway.
- A stiff cervical spine (neck region of the spine) and the possibility of spine instability may prevent an anesthesiologist from placing the head and neck of the individual in the best position to view the larynx. The limited jaw movement and short neck sometimes seen in individuals with MPS make it that much more difficult for even a very skilled anesthesiologist to see the larynx.
- Airway difficulties increase with age of the individual, most likely secondary to continuous infiltration of GAGs in the soft tissue.

Individuals with MPS have other symptoms that can contribute to the increased risks associated with general anesthesia. For example:

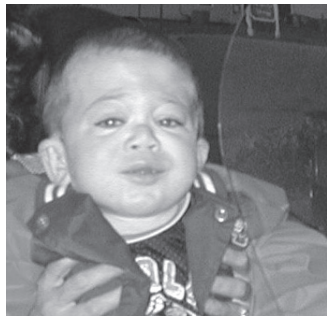
- The storage of GAGs causes joint stiffness and thick skin, which can make it difficult to establish an IV line, especially in emergency situations.
- The storage of GAGs in the heart muscle, heart valves and blood vessels of the heart can add to the overall burden to the cardiovascular system. The heart muscle may become sensitive to anesthetics and/or low levels of oxygen.
- Spine deformities seen in many individuals with MPS can restrict the volume of the lungs causing pulmonary dysfunction.

Potential risks and complications

As a result of the burden of symptoms described, individuals with MPS may experience some of the following complications while under general anesthesia:

Airway (breathing) problems

- There may be difficulty in placing the endotracheal tube into the trachea (i.e., difficult intubation).


AUSTIN (ML II)

- Breathing in of stomach contents into the lungs (aspiration) may occur during intubation attempts, causing damage to the lung tissue.
- There may be difficulty in removing the endotracheal tube (extubation) after surgery due to excessive tissue swelling.
- There may be difficulty in keeping the airway open after removing the endotracheal tube.
- The endotracheal tube may have to remain in place after surgery if the initial placement was difficult or traumatic.
- Emergency tracheotomy (making an incision in the neck and inserting a tube directly into the trachea) may be necessary if the airway becomes compromised during intubation or extubation.

Cardiac (heart) problems

- Heart failure may occur.
- The heart rhythm may become irregular.
- There may be large changes (higher or lower) in blood pressure.

Neurological (brain and nerve) problems

- Long-term effects of general anesthesia on the brain of MPS individuals remain unknown.
- Dislocation of the spine in the neck region (subluxation) during intubation can cause spinal cord injury.
- Pressure point injuries to peripheral nerves can occur.

Skeletal (bone) problems

- Stress can be placed on the muscle and bones of the arms, legs and spine during long surgical procedures.

It is important to remember that this list does not give all the possible complications, nor does it imply that all individuals with MPS will experience these problems. Depending on what MPS disease the individual has, certain symptoms, and therefore risks, may or may not be present.

What can be done to reduce the risks?

Assessing the risks prior to a procedure

There are no minor anesthetics for most individuals with MPS. *The risks of general anesthesia should be weighed against the advantage to be gained from the surgery or procedure.* If you are worried about the proposed surgery, discuss it with your primary care doctor, surgeon and/or medical geneticist. He or she may suggest seeking a second opinion.

For many individuals with MPS, it is possible to determine, before they undergo anesthesia, whether they have a significantly increased risk associated with such a procedure. To avoid potential anesthetic complications, the individual having surgery will require a preoperative examination and evaluation.

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An evaluation by a pulmonologist (lung doctor) and cardiologist (heart doctor) can be helpful in determining preoperative risks. The pulmonologist may recommend chest X-rays, cervical spine (neck) X-rays, pulmonary (lung) function tests and possibly a sleep study. Sleep studies can help define the amount of airway obstruction an individual might have. The pulmonologist also may want to look at the airway using a flexible bronchoscope, which is a small fiber-optic tube used to view the airway. The cardiologist may recommend an electrocardiogram (ECG) and/or echocardiogram in order to better evaluate the individual's heart rhythm, muscle and valves. Medical clearance for surgery from one or both of these specialists is frequently advised, especially for individuals with advanced disease.

The importance of the facility

For both children and adults with MPS, it is safer to have medical interventions at a major hospital with staff experienced in treating individuals with complicated diseases such as MPS. This usually entails traveling to a regional medical center, children's hospital or university hospital. These facilities have the medical specialists, including pediatric anesthesiologists and medical intensivists, capable of handling the challenging aspects of providing care for individuals with MPS. In addition, these facilities have intensive

care units, including pediatric intensive care, required to take care of individuals needing prolonged ventilatory (breathing machine) support after surgery.

Although outpatient surgery (where the person returns home on the same day of the surgery) has become more common in the general population, it may **not be suitable** for individuals with MPS, even when having a routine procedure or operation. This is especially pertinent for severely affected individuals.

The importance of the anesthesiologist

As mentioned earlier in this booklet, there are no minor anesthetics for most people with MPS. Unfortunately, children with MPS have died as a result of anesthesiologists who did not expect, or were not prepared to deal with, the severely compromised airway seen in individuals with MPS. Planned procedures should always involve careful assessment by a pediatric or general anesthesiologist with appropriate skills and training. In fact, for many surgical procedures, it may be important to select an anesthesiologist even before choosing a surgeon. Therefore, anesthesiologists should be consulted during the planning process for a surgical intervention.

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The anesthesiologist should be aware of the affected individual's disease and current health condition. A list of the individual's medications and allergies, as well as any notes or recommendations from specialists involved in the preoperative assessment of the individual, should be provided to the anesthesiologist. All complications or problems from previous anesthetic experiences should be described. This includes problems as simple as fear of injections and dislike of the smell of anesthetic gas, to more serious issues such as postoperative breathing problems and prolonged intubation. In addition, the medical team should be informed of all of the individual's disabilities, including ambulation, mobility, feeding, hearing and vision problems.

The anesthesiologist is responsible for deciding the best method of anesthetizing the person who is undergoing medical intervention. There are several aspects to this:

- The anesthesiologist will assess the individual and determine whether or not he or she is in the best possible condition to proceed with surgery and the required anesthetic.
- He or she will determine what kind of anesthetic (general vs. local) is best for the given situation.
- The anesthesiologist may prescribe a pre-medication, which is given to sedate the individual prior to entering the operating room.
- The anesthesiologist will determine the degree of monitoring necessary for the particular procedure and health status of the individual. Specialized equipment is used to monitor the individual's heart rate and rhythm, blood pressure and oxygenation. Invasive monitors are occasionally required, where small catheters are placed in an artery or large vein to monitor arterial blood pressure and central blood pressures around the heart, respectively.
- As described earlier in this booklet, individuals with MPS present the anesthesiologist with difficult airways, and therefore difficult intubations using conventional methods. There are several alternative methods that can be used to establish an airway. As an alternative to intubating an individual with an endotracheal (breathing) tube, the anesthesiologist may opt to place a soft intra-oral device known as a ***laryngeal mask airway (LMA)***. The LMA creates an open airway from the mouth to the larynx, is relatively easy to insert, and can be used during general anesthetics for procedures that do not absolutely require securing the airway with an endotracheal tube. If the individual requires an endotracheal tube, a flexible fiber-optic bronchoscope may be used to pass the endotracheal tube into the trachea. The anesthesiologist may use the previously described LMA device to create a conduit for passing the bronchoscope and, in turn, the endotracheal tube. While such techniques have been found to be beneficial for individuals with MPS, they require an anesthesiologist who is skilled with these methods and decision making.

ISABELE (MPS III)

- The anesthesiologist will be responsible for monitoring the depth of the individual's anesthesia, for administering IV fluids during surgery, and for determining whether there is a need for prolonged intubation.

- The anesthesiologist might decide that the individual may benefit from a type of local anesthetic known as a ***regional anesthetic***. There are a variety of regional anesthesia options, including spinal anesthesia, epidural anesthesia and peripheral nerve blocks. Regional anesthetics can be used alone to numb a region of the body, or in combination with a general anesthetic. When combined with a general anesthetic, regional anesthesia is usually given to provide post-procedural pain relief.

ELIOT (MPS I)

- In regional anesthesia, the anesthesiologist injects a local anesthetic medication through a needle or small catheter around various nerves of the body depending on where numbness is desired. For example, when ***epidural anesthesia*** is performed, a needle is inserted into the back of the spine through which a small catheter is threaded into the epidural space near the spinal cord. Local anesthetic medication is delivered through this catheter numbing major nerves that go to the chest, abdomen or legs.

- Many steps are taken to make the overall process safer. Certain anesthetic drugs might be utilized or avoided. The anesthesiologist will ensure the safe positioning of the individual while they are anesthetized, including stabilization of the cervical spine (neck), padding of pressure points and protection of the eyes.

What does the overall process involve?

Preparation

Below are some of the steps involved in preparing the individual for anesthesia.

Consent

The surgeon or members of the medical team will explain what is planned for the operation or procedure. Risks and benefits of the procedure and anesthesia will be described, as well as an explanation of possible outcomes. The parent or power of attorney is required to sign an official form of consent giving permission for the medical intervention to take place. This consent form is legal documentation showing the consenter's understanding of the proposed surgery and its risks. The person signing the consent form should continue to ask questions until he or she is comfortable with what is being agreed to.

"Nothing by mouth (NPO)"

The individual undergoing surgical intervention will be asked not to eat or drink anything for usually four to eight hours, depending on age, before receiving an anesthetic. The abbreviation "NPO" comes from the Latin saying for "nothing by mouth." Individuals undergoing general anesthesia are at risk of vomiting, which can in turn lead to breathing in of stomach contents, such as food and drink. This complication, known as aspiration, can cause significant damage to the lungs. Its chance of happening can be reduced by having the stomach empty prior to anesthesia, therefore the "NPO" rule.

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Checking in

On the day of surgery, the individual will be admitted to the pre-surgical area where he or she will change into a hospital gown. All of the essential medical documentation is gathered in the individual's medical chart. Admission vital signs are obtained including body temperature, blood pressure, heart rate and breathing rate. A

painless piece of equipment known as a pulse oximeter is placed on the individual's finger showing the degree of oxygen saturation in the blood. The individual should remember to bring his or her hearing aids and glasses to assist the medical team in communicating.

Pre-medication

Pre-medication is the medicine that is given prior to a general anesthetic. This may or not be given, and can vary with the age of the individual and the type of procedure to be performed. Medication is usually given to help people relax, dry up moisture in the mouth and throat, and to facilitate the onset of anesthesia. Pre-medication may be consumed by mouth (for example, via a drink), or be given by an IV injection or intramuscular injection (for example, in the thigh muscles or buttocks). After the pre-medication has been given, the individual should rest on the bed or sit with a caregiver, as the individual will become sedated and may even fall asleep. The individual may become restless or irritable prior to the medication taking its full effect and, rarely, the medicine may have a reverse effect and make the individual more energetic rather than drowsy. If this has happened previously, this should be communicated to the anesthesiologist.

Starting the IV

It is safer to have an IV line in place prior to commencing a general anesthetic. An IV allows for easy administration of a variety medications required for the safe administration of general anesthesia. The IV may be the most difficult part for individuals, as it requires the individual to remain still for a needle stick. Oral or intramuscular sedative pre-medications can help. Injecting local anesthetic through a small needle can numb the site where the IV is inserted. This is quick and usually well tolerated. Certain facilities might offer a topical anesthetic cream which is applied to the IV site about an hour before its insertion. Unfortunately, this topical cream numbs only the skin and is not beneficial for intramuscular injections.

CARLY (MPS IV)

The anesthesiologist may decide to start the IV in the operating room after the individual is put to sleep by breathing anesthesia gas

from a face mask. This decision is made only if starting the IV is too difficult for the awake individual, and only if the individual can be safely placed under general anesthesia using a face mask. Discuss with the anesthesiologist the risks and benefits of using this method of starting the IV.

In the operating room

A nurse always accompanies the individual undergoing surgery into the operating room. The child's favorite stuffed animal, doll or blanket can be taken into the operating room.

Depending on the rules of the facility, a parent may be offered the opportunity to accompany their child to the operating room and remain there until he or she is asleep. This will be discussed ahead of time and mutually agreed upon by the anesthesiologist and parent. The parent will be asked to put on a gown and shoe coverings before going into the sterile operating room.

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The anesthesiologist will explain how the procedure will move forward and how the parent can be of assistance. Occasionally, small children can be anesthetized on their parent's lap while the face mask is held on the child's face. Once the child is asleep, the parent will be escorted out of the operating room. It is important to leave quickly, as the anesthesiologist has many things to do to ensure the safety of the child being anesthetized.

The nurse will estimate how long the individual is likely to be in the operating room. Many parents or caregivers like to go for a walk or have a meal. If the individual is going to the intensive care unit after the procedure, the parent or caregiver may be taken to this ward beforehand. Many operations take longer than planned, and individuals usually spend a period of time in the recovery room before going back to their room or being discharged. Parents can obtain updates on the surgical progress by asking personnel at the information desk in the family waiting area.

Back in the recovery room

After the medical procedure, individuals are transported to the recovery room to wake up and be monitored. Individuals emerging

from anesthesia may be drowsy, disoriented and unaware. In most children's hospitals and in recovery rooms that deal with pediatric individuals, parents will be able to join their child once he or she has been taken to the recovery room. For a child emerging from anesthesia, hearing a familiar voice will help him or her relax and rest peacefully. The nurse will indicate when it is safe for the individual to drink and/or eat something.

The intensive care unit

If the individual requires intensive postoperative observation and monitoring, he or she may be admitted to the intensive care unit (ICU). Admission to the ICU is required if the individual requires prolonged breathing support from a ventilator (breathing machine). ICU rooms have sophisticated monitors, and usually have one nurse for each individual. Once the intensive care of the ICU is not needed, the individual is transferred to a regular hospital room, and occasionally discharged directly to home.

Discharge

When the individual with MPS has recovered from anesthesia and has reached the health status allowing for recovery at home, he or she will be discharged. Due to the multiple medical problems individuals with MPS have, discharge to home may take longer than normal. As explained earlier in this booklet, outpatient surgery (where the person returns home on the same day of surgery) may not be suitable for severely affected individuals, even when having routine operations. The surgeon will send home orders that instruct individuals and caregivers about eating, activity and the administration of medicines.

Conclusion

Familiarity with and education of the many problems associated with MPS allows for optimal care of people with these diseases. This booklet describes what anesthesia is and the impact MPS has on the provision of anesthesia. It lists many of the risks and complications associated with general anesthesia, as well as steps to potentially minimize them. It must be remembered that individuals with MPS have a higher risk of complications from anesthesia and that there are no "minor" anesthetics for them. The information provided here is intended to help individuals, caregivers and doctors better prepare for anesthesia, with the hope that individuals with MPS can safely receive medical interventions intended to improve their long-term health.

Familiarity with and education of the many problems associated with MPS allows for optimal care of people with these diseases.

Common bonds unite the lives of those affected by MPS and related diseases—the need for support and the hope for a cure.

The National MPS Society is committed to making a difference in the lives of MPS families through support, research, education and advocacy. Families from around the world gain a better understanding of these rare genetically determined diseases through the Society's assistance in linking them with healthcare professionals, researchers and, perhaps most importantly, each other.

Individuals affected with an MPS or related disease and their families have a resource. One that stands ready to help—a resource that takes an active role in fostering the courage necessary to confront these diseases every day.

Benefits of membership in the National MPS Society:

- *Courage*, our quarterly newsletter containing stories and information about individuals with MPS and related diseases;
- Educational materials such as fact sheets and an MPS glossary;
- Conference and education scholarships;
- The Family Assistance Program, which provides financial support for durable medical goods;
- News about various Society sponsored conferences and gatherings, where families and leading MPS scientists, physicians and researchers join together for a common cause;
- Information on local events, such as regional social events and fundraisers. These events create opportunities for families to meet each other and help raise community awareness of these rare genetic diseases; and
- A listing in our annual directory of members that assists families to connect with one another.

National MPS Society

For more information or to join the National MPS Society:

Visit **www.mpssociety.org**
Contact us at **877.MPS.1001**
Or email us at **info@mpssociety.org**