

# The National MPS Society

## Families Joining Together

Common bonds unite the lives of those affected by mucopolysaccharidoses (MPS) and mucopolipidoses (ML) disorders – the need for support and the hope for a treatment.

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

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

Join the National MPS Society and enjoy a variety of benefits, including:

- Courage, our quarterly newsletter that shares stories and information about people with MPS or ML.
- News about various National MPS Society-sponsored conferences and gatherings, where families and leading MPS and ML scientists, physicians and researchers are brought together.
- Information on local events like regional picnics and fundraisers. Opportunities for families to meet each other and help raise community awareness of these rare genetic diseases.
- A listing in our annual directory of members, which assists families in locating each other.

For more information or to join the National MPS Society, please visit:

[www.mpssociety.org](http://www.mpssociety.org)  
or contact us by e-mail at  
[info@mpssociety.org](mailto:info@mpssociety.org)



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## Understanding MPS and ML Is Your Child Having an Anesthetic?



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## Introduction

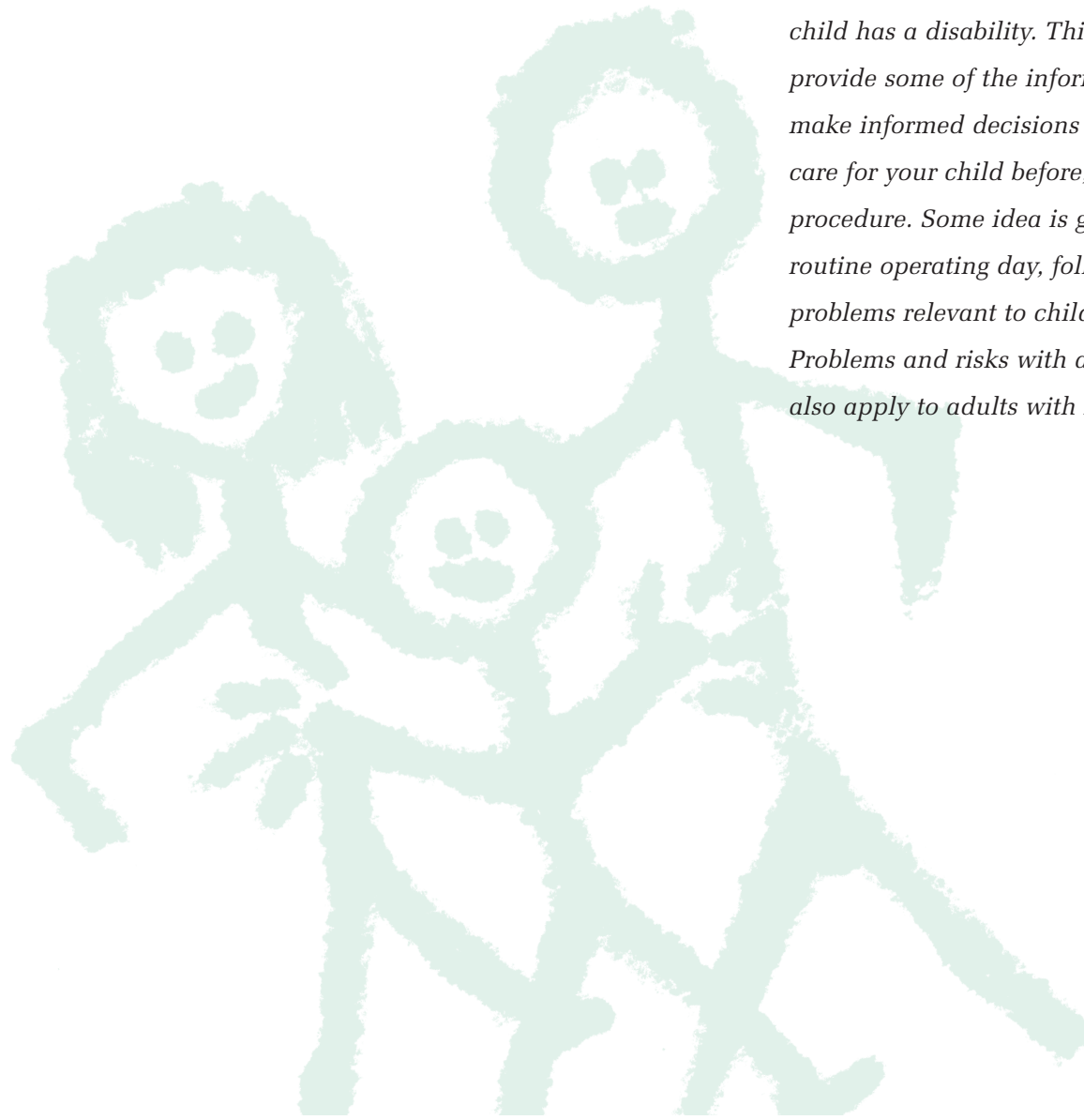
*If your child is scheduled for an operation or procedure and will be having an anesthetic, you may be wondering what happens. Parents are essential members of the team caring for their child, particularly if that child has a disability. This booklet has been written to provide some of the information you will need in order to make informed decisions about anesthesia, and to help care for your child before, during and after the procedure. Some idea is given of what happens on any routine operating day, followed by the particular problems relevant to children with MPS and ML. Problems and risks with anesthesia in children with MPS also apply to adults with MPS.*

*front cover*

*top: Ryan, age 6, MPS IV*

*middle: Cameron, age 7, MPS II*

*bottom: Gilberto, age 9, MPS II*



## What is anesthesia?

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Anesthesia means a loss of feeling, particularly the sensations of pain and touch.

A local anesthetic numbs only the relevant area of the body. The individual does not feel pain in this area, but remains awake and aware of what is going on.

During a general anesthetic your child will be unconscious, unable to move or breathe and will not feel pain. To assure safe oxygen levels while unconscious, an adequate airway needs to be maintained. The typical way to managing this problem is to pass a tube through the larynx into the trachea. The tube remains in the airway (trachea) during the procedure and is removed prior to fully waking up at the end of the procedure.

This booklet will focus on procedures involved while under a general anesthetic.

## Giving consent

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The surgeon or members of the medical team will explain what is planned for the operation or procedure. You will be asked to sign an official form of consent. You should continue to ask questions until you feel you completely understand what you are agreeing to.

If your child is able to understand, he or she could also be asked to sign the consent form.

## Meeting the anesthesiologist

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Most children with MPS need to be anesthetized in a regional medical center or university hospital by a trained PEDIATRIC ANESTHESIOLOGIST. For many surgical procedures, it is important to first identify the



Daniel, age 22  
MPS II

anesthesiologist prior to choosing the surgeon. There is no substitute for an examination of the child by the anesthesiologist PRIOR to the day of surgery. MPS children have died as a result of anesthesiologists who did not expect or were not prepared to deal with the severely compromised airway seen in MPS individuals.

To avoid potential anesthetic complications in children with MPS, you and your child's doctor(s) will need to consider preoperative examination and evaluation, anesthetic planning, planning for the unusual or unexpected occurrence and anesthesia training and experience. Your child's cardiologist (heart doctor) and pulmonologist (lung doctor) should be consulted about risk for potential problems during anesthesia. If your child has had an operation before, you may already have useful information to provide to the anesthesiologist. Some children are afraid of injections, some hate the smell of gas. The anesthesiologist is responsible for deciding the best method of anesthetizing your child.

The anesthesiologist or a member of the team will visit your child before the procedure and prescribe the medication needed to prepare for the anesthetic.

Sometimes parents accompany their child to the operating room and remain there until he or she is asleep. If you feel this will help your child, you should discuss this with the anesthesiologist.

## Nothing by mouth (NPO)

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You will be told that your child should not eat or drink anything (usually at least four to six hours) prior to receiving the anesthetic. Children undergoing general anesthesia are at risk to

vomit. To diminish the chance of the stomach's content, such as food, entering the lungs (aspiration) if your child vomits, it is important to have the stomach empty prior to the procedure.

## Premedication

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"Premedication" is the medicine that is given before the general anesthetic. This can vary with the age of the child and the type of procedure. Something is usually given to help the child relax, to dry up moisture in the mouth and throat, and to make it easier for the anesthetic to be given.

Your child may be given some medicine to drink, or it may be given intravenously (IV) or by an intramuscular (IM) injection in the thigh or buttocks.

## Anesthetic cream

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If the anesthetic is to be given by IV injection, an anesthetic cream may be applied to the site of application an hour before the operation. This will numb the area so that the child will not feel the needle going in. Unfortunately, the cream numbs only the skin and does not help with an IM injection given as part of the premedication.

## Waiting

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Inform the medical team of your child's developmental ability and any hearing or vision problems. Bring hearing aids and glasses to the waiting area to assist the team in communicating with your child.



*From top left, clockwise:  
Donna, age 38, MPS IV  
Noel, MPS IV  
Ryan, age 6, MPS IV  
Ian, age 12, MPS IV  
David, age 4, MPS IV*

Premedication is likely to make the child's mouth feel dry. Some children are rather irritable until the medication has had a chance to work properly. Children can also become suddenly wobbly on their feet. It is safer for them to rest on their bed or sit with a caregiver. If your child falls asleep, it would be better for him or her to lie on the bed or be held in a horizontal position, as blood pressure may drop if the child stays upright.

Occasionally, some of the premedication given to children with MPS will have a reverse effect and make them energetic rather than drowsy. If this has happened previously, be sure to tell the anesthesiologist about it, as well as the type of medication that produced the effect.

If you are not going to the operating room, you should discuss with the nurse the best time to say good-bye. This will avoid any last minute distress as the child goes to the operating room.

## Going to the operating room with your child

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A nurse always accompanies the child into the operating room. A favorite teddy, doll or blanket could go too, but it would be a good idea to bring it back with you for safekeeping. These procedures may vary from hospital to hospital.

You may be asked to put on a gown and shoe coverings before going into the operating area. The anesthesiologist will explain the procedure being used for your child. As mentioned earlier, sometimes an IV injection is used to initially let your child fall asleep or the

child is asked to breathe in an anesthetic through a mask. Small children can occasionally be anesthetized on their parent's knee while the gas tube is held near their face.

Once your child is asleep, you will be asked to leave. It is very important to go as soon as you are asked. The anesthesiologist has many things to do very quickly to ensure your child is safely anesthetized.

### **While your child is in the operating room**

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The nurse will estimate how long your child is likely to be in the operating room. Many parents like to go for a walk or have a meal. If your child is going to intensive care afterwards, you could be taken to see the ward beforehand.

Many operations take longer than planned and children usually spend a period of time in the recovery room before going back to their room or being discharged. If you are worried, you can ask the nurse to check how your child is doing. In most children's hospitals, you will be able to join your child once he or she has been taken to the recovery room.

### **Back in the room**

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After having an anesthetic, your child may seem drowsy and unaware, but hearing your voice will help him or her relax and sleep more deeply. The nurse will tell you when it is safe for your child to drink something.

### **What are the special considerations when planning an anesthetic for a child with MPS/ML or a similar condition?**

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General anesthetics should always be given in a hospital by a skilled anesthesiologist. Planned procedures should always involve careful assessment of the child by the anesthesiologist. There are no MINOR anesthetics for most children with MPS.

In order to understand the possible problems, it is helpful to know something of what is involved in anesthetizing a child.

### **Anesthetizing a patient - the usual procedure**

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1. Gas or a medication given by injection is used to make the child go to sleep.
2. A muscle relaxant is introduced, which paralyzes all the muscles including the breathing mechanism.
3. Supplemental oxygen is given by squeezing a bag linked to a face mask. This is a temporary supply until a breathing 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 anesthesiologist can view the entrance to the larynx.
5. A tube (endotracheal) is introduced through the larynx into the trachea and the laryngoscope is removed.
6. The endotracheal tube is connected to a machine that breathes for your child during the procedure.
7. At the end of the surgical procedure, the anesthetic agent is withdrawn and your child should start to wake up.
8. The endotracheal tube is usually removed before the individual is fully awake.

## Problems for MPS/ML children

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Anything that makes it difficult for the anesthesiologist to perform the steps required to anesthetize your child will increase the risks of having general anesthesia. The storage of glycosaminoglycans in all tissues (e.g. nose, tonsils, adenoids, throat, trachea, and heart) and its effect on bone formation contribute to the difficulty in safely anesthetizing MPS children.

The accumulation of the glycosaminoglycans narrows the nasal passages; enlarges the tonsils, adenoids and tongue; and causes loose redundant tissues to form around the larynx. These problems severely limit the anesthesiologist's ability to visualize the larynx with the laryngoscope. With the muscle relaxation produced by premedication and/or general anesthesia, obstruction of the airway increases. In addition, a stiff cervical spine and the possibility of an unstable spine prevents the anesthesiologist from placing the head and neck in the best position to view the larynx. The limited jaw movement, short neck and thick secretions compound the difficulty in visualizing the larynx by even a very skilled anesthesiologist. Children with MPS have other characteristics that contribute to the increased risk of anesthesia, such as thick skin and joint contractures which make IV access difficult, especially in an emergency situation.

During general anesthesia of MPS patients, the storage of glycosaminoglycans in the heart and blood vessels of the heart can contribute to increased risk. Individuals with MPS should be cleared for anesthesia by their cardiologist prior to the day of the procedure. The risk of heart-related problems is much less compared to airway problems in the MPS individual during anesthesia.

## Potential problems while MPS/ML individuals are under anesthesia

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1. Airway problems
  - a. Difficulty in placing (intubation) the breathing tube into the trachea.
  - b. Difficulty in maintaining an adequate airway after the breathing tube has been removed (extubation).
  - c. The breathing tube might have to remain in place after the surgery, if the initial placement was very difficult or traumatic.
  - d. Emergency tracheostomy (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.
  - e. Even minor procedures may require a stay in the intensive care unit so that breathing may be monitored.
2. Cardiac problems
  - a. Heart failure
  - b. Heart muscle may be very sensitive to anesthetics
  - c. Heart rhythm may be irregular
  - d. Heart muscle may be sensitive to oxygen needs
  - e. Prone to wide swings in blood pressure

## What can be done?

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Your MPS child experiences increased risk during an anesthetic. Therefore, it is safer to have the procedure at a major hospital experienced with treating MPS children, even if extensive travel is required. It may be necessary for your child to remain intubated and on a ventilator (breathing machine) for a period of time following surgery. So, a hospital with a

pediatric intensive care unit is essential. Outpatient surgery may not be suitable for severely affected patients, even when having routine operations.

If you are worried about the proposed surgery, discuss it with your primary care physician or medical geneticist. He or she may suggest seeking a second opinion. The risks of anesthesia should be weighed against the advantage to be gained from the surgery or procedure.

Make sure that the anesthesiologist is aware of your child's condition and possible problems. There are steps that can be taken to make the process safer, such as avoiding particular anesthetic drugs or stabilizing the neck of patients with cervical spine problems. For some procedures a local anesthesia may be an option.

For many MPS individuals, it is possible to determine prior to anesthesia if they have a significantly increased risk with anesthesia. An evaluation by a lung doctor (pulmonologist) and heart doctor (cardiologist) can be helpful in determining the risk of anesthesia. The pulmonologist may recommend a sleep study and a pulmonary function test. The pulmonologist may want to evaluate the anatomy of the airway using a flexible bronchoscope, which is a small tube that can be used to view the airway.

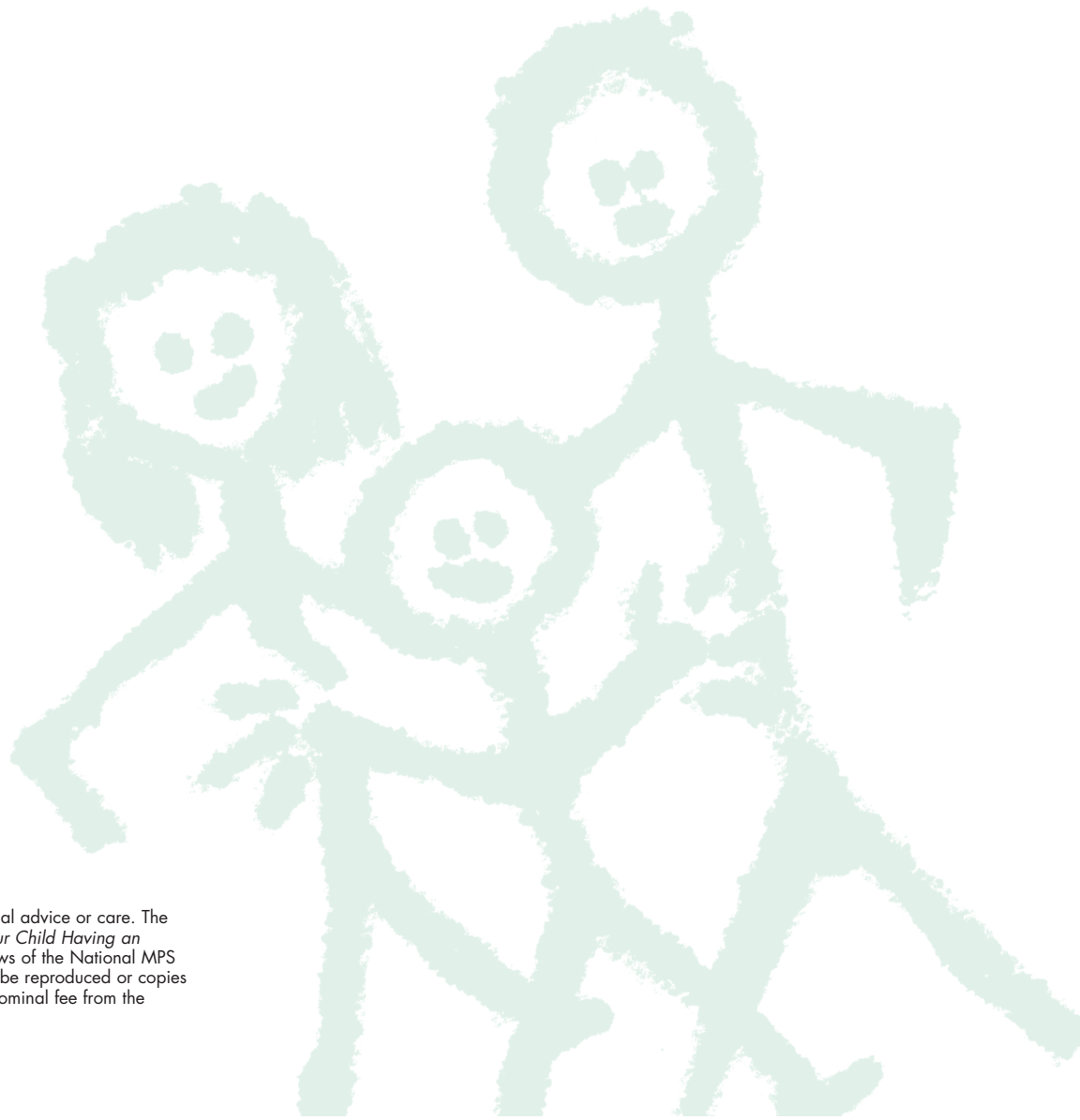
Because individuals with MPS present the anesthesiologist with both difficult airways and often an impossible intubation using conventional methods, other methods are being used. The flexible bronchoscope can be used to pass the breathing tube into the trachea at the start of the procedure. Another option for some anesthesiologists is to use a laryngeal mask airway in combination with a flexible bronchoscope. Using

a flexible bronchoscope and/or a laryngeal mask airway are techniques which have been found to be beneficial for MPS children, but require the anesthesiologist to have skill with these newer methods of establishing an airway.

## Conclusion

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We have listed a range of problems that can occur during anesthesia and ways to minimize these problems. This does not mean that your child, particularly when young, will have any of them. In general, children with MPS III are at less risk compared to individuals with other types of MPS who have more severe physical problems. Many children with MPS have had successful surgery resulting in improved quality of life.



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