

# ANESTHESIA

## INTRODUCTION:

General anesthesia refers to putting a patient to sleep before an operation. Most of the time, in order for the person under anesthesia to get enough oxygen during the surgery, they are ventilated with a laryngeal mask. In some cases, the use of the mask is not possible. The patient must then be intubated by inserting into their throat a tube connected to a machine that helps them breathe.

Due to their unique anatomy, and certain complications inherent with their diagnosis, general anesthesia can be more challenging in people with dwarfism.

This factsheet identifies the main problems related to anesthesia in little people. The goal is not to cause any additional anxiety for the patient needing surgery, but to prepare the medical team (family doctor, surgeon, anesthetist, etc.) for possible complications, so that they can be avoided or properly addressed.

## GENERALITIES RELATED TO THE CONDITION:

It is important to note that the dosage of anesthesia and other drugs must be administered according to body weight. Therefore, adults with dwarfism do not necessarily require the same dosage as adults of average height. A lower dosage is therefore adequate for the needed result.

Also, little people often have smaller diameter airways and windpipes than people of average height. The insertion of the breathing tube can also be difficult and require the use of a flexible bronchoscope. A fiber optic intubation may also be necessary.

In patients with osteogenesis imperfecta, the size of the breathing tube should be determined by the size of the head of the patient, not the size of the body. In some cases, however, alternative intubation procedures will have to be carried out (e.g., Nasal Tube Insertion and Passage). Finally, we should keep in mind that people with osteogenesis imperfecta can easily suffer from bone fractures if not handled carefully during certain medical procedures.

The medical team should therefore be aware of the susceptibility of the bones to fractures associated with various movements, impacts or stress.

## PALATE MALFORMATION:

Many types of dwarfism are associated with cleft palates or submucosal membranes. Cleft palate, an absence of the palate, is visible from birth. On the other hand, the submucosal cleft often goes unnoticed, since it occurs below the surface of the mucosa. It is important to be



aware of these anatomical differences, as they can cause difficulties when inserting breathing tubes.

### **SPINAL CORD COMPRESSION:**

The spinal vertebrae of many little people can have abnormalities that can cause crushing or pinching of the nerves of the spinal cord. For example, achondroplasia can be associated with the narrowing of the spinal canal, the canal through which the spinal cord passes. Spinal anesthesia can therefore be complicated in people with achondroplasia. This can possibly be a problem if a pregnant woman with achondroplasia has to deliver by caesarean section, as it is the usual route for anesthesia to be delivered in this case. General anesthesia should then be considered, even if an epidural is still possible.

In other types of dwarfism, the cervical spine may be unstable. In people with Morquio syndrome, for example, this instability is associated with the shape of the vertebrae and the build-up of glycosaminoglycans. This problem is also present in other lysosomal overload diseases (such as Maroteaux-Lamy Syndrome).

When cervical instability is present, it is imperative to support the patient's neck during intubation as well as all other manipulations to avoid spinal cord compression. Because of these risks, it is also recommended that people with cervical instability always carry a letter or bracelet advising of the risks associated with intubation.

Spinal cord compression is potentially very serious and can cause paralysis and even death. This complication is therefore important to bear in mind while protecting the spine throughout the duration of the anesthesia.

### **HYPERTHERMIA:**

The body temperature of people with osteogenesis imperfecta may rise during and after surgery. This increase in temperature is usually not associated with malignant hyperthermia. Precautions such as not using electric blankets or heavy curtains are often effective and sufficient. However, under some circumstances ice packs or other cooling measures are necessary. Some anesthetic agents such as atropine should be avoided due to their tendency to exacerbate hyperthermia.

### **RESPIRATORY AND HEART PROBLEMS:**

Some types of dwarfism can cause heart and respiratory complications which present risks in the event of anesthesia. For example, with osteogenesis imperfecta, deformities of the chest and ribs and/or scoliosis can interfere with breathing. The presence of heart valve disease is



also likely to be a problem.

In Morquio syndrome, heart complications can lead to heart failure. Likewise, the respiratory problems associated with this diagnosis can make it difficult to remove the endotracheal tube after surgery. Finally, the medical team should be aware that they may need to perform an emergency tracheostomy during the operation.

In people with achondroplasia, there are risks of obstructive apnea and concomitant risks of post-operative obstruction or post-extubation pulmonary edema.

### **TO VERIFY BEFORE ANESTHESIA:**

Due to the possible issues and risks described above, precautions are required before considering anesthesia for certain little people.

People whose diagnosis presents a risk for neurological, respiratory and/or cardiac complications (achondroplasia, osteogenesis imperfecta, Morquio syndrome, etc.) should therefore be carefully evaluated before surgery. The anatomy and health of the spine (especially the cervical spine), lungs and heart should be assessed, and an evaluation performed by an anesthesiologist.

### **CONCLUSIONS:**

In all cases, it is preferable to undergo the operation in a hospital which has a solid knowledge of the diagnoses associated with the different forms of dwarfism and to call in an experienced anesthesiologist.

In addition, to minimize complications, it is recommended that several operations/procedures be performed at once if possible, in people for whom anesthesia may be risky.

Likewise, due to the possibility of complications, one can choose to have the operation at night, even for surgery that is usually done during the day.

### **RESOURCES:**

Association québécoise des personnes de petite taille

<https://www.aqppt.org/>

Little People of Ontario

<https://littlepeopleofontario.com/>

Regroupement québécois des maladies orphelines - Centre iRARE

<https://rqmo.org/centre-dinformation-et-de-ressources-en-maladies-rares/>

Little People of America - Judith G. Hall, Special Problems Of Anesthesia For Little People



<https://www.lpaonline.org/assets/documents/Special%20Problems%20Of%20Anesthesia%20For%20Little%20People.pdf>

Little People of America - Ivor D. Berkowitz, Srinivasa N. Raja, Karen S. Bender, Steven E. Kopitz, "Dwarfs: Pathophysiology and Anesthetic Implications", *Anesthesiology*, Vol. 73, No. 4, october 1990.

<https://www.lpaonline.org/assets/documents/Dwarfs%20Pathophysiology%20and%20Anesthetic%20Implications.pdf>

Orphan Anesthesia: Anesthesia recommendations for patients suffering from Achondroplasia, 2011

[https://www.orpha.net/data/patho/Pro/en/Achondroplasia\\_EN.pdf](https://www.orpha.net/data/patho/Pro/en/Achondroplasia_EN.pdf)

Osteogenesis Imperfecta Foundation - Anesthesia Information for People who have Osteogenesis Imperfecta

<http://www.oif.org/site/DocServer/Surgery.pdf?docID=11101>

*Guide pour mieux comprendre la mucopolysaccharidose IV*, Canadian MPS Society for mucopolysaccharidose & related diseases, 2013

<http://www.mpsociety.ca/wp-content/uploads/2018/07/MPSIVBookletFrench.pdf>

*Vivre avec la MPS IV : guide à l'intention des patients et des familles*, Canadian MPS Society for mucopolysaccharidose & related diseases Inc., 2016

[http://www.mpsociety.ca/wp-content/uploads/2018/07/MPSIV\\_binder\\_20160902\\_final\\_FRCA.pdf](http://www.mpsociety.ca/wp-content/uploads/2018/07/MPSIV_binder_20160902_final_FRCA.pdf)



**Please contact us for more information**

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