FREQUENT INTERVENTIONS AMONGST LITTLE PEOPLE

INTRODUCTION ;

The various diagnoses which result in dwarfism bring with them their share of complications, including malformations, musculoskeletal and/or neurological problems. This factsheet lists the main surgeries associated with these complications. However, it should be understood that the complications vary according to the forms of dwarfism. Some types of dwarfism cause few problems, while others cause more. Please refer to the proper fact sheet regarding a specific type to see the main surgeries associated with the condition.

PALATAL FISSURE :

Palatal fissure (opening of the palate) is a congenital malformation characterized by the absence, more or less complete, of a palate. It is often associated with a cleft lip (lip). Surgery is available to correct it, often performed around the eighteenth month, or in severe cases, from the sixth month after birth.

JAW :

The conditions that cause dwarfism can sometimes lead to deformities of the jaw. If necessary, maxillofacial surgery can be utilized to remedy the situation. In most cases, surgery can be performed once the jaw is fully grown.

OBSTRUCTIVE APNEA :

Obstructive apnea, common in many types of dwarfism, results from an obstruction (partial or complete closure) of the respiratory tract. People who are affected by obstructive apnea have particularly loud snoring. An adenoidectomy (removal of the adenoids) and/or a tonsillectomy (removal of the tonsils) can help resolve this problem.

OTITIS :

Many children with dwarfism have a deformity of the middle ear, which increases the frequency of ear infections. If necessary, a myringotomy can be performed. This operation entails making a small incision in the eardrum to insert a tube. This procedure helps restore

hearing as well as ventilate the middle ear. Myringotomy is done frequently and is more often carried out while the individual is receiving local anesthesia.

HYDROCEPHALUS :

Hydrocephalus (excess cerebrospinal fluid in the brain), which occurs in the first few months of life, affects some infants with short stature. In most cases, it resolves on its own around 2 or 3 years of age. There are two types of hydrocephalus; communicating and non-communicating. In the non-communicating form, which is more rare, cerebrospinal fluid does not circulate. The establishment of a bypass is then necessary. This operation involves placing a tube to divert the cerebrospinal fluid to another part of the body, most often the abdomen. When a child wears a bypass, it should be monitored regularly to ensure that the device is working properly.

LEG DEFORMITY :

Depending on the type of dwarfism, legs can be bowed into varus (outward) or valgus (inward). If a surgical correction is needed, it should be delayed as long as possible because of the possibility of recurrence. The surgical procedure, called an osteotomy (surgical sections of the long bones) corrects the axis of the legs so that they are straightened. It requires hospitalization for a few days as well as rehabilitation.

HIP AND KNEES :

Dwarfism sometimes causes premature wear and tear on the joints, especially the hip and knees. When the wear is too great, an arthroplasty, a surgery to restore the shape and mobility of the joint using prostheses, will be performed. This surgery requires hospitalization for a few days, followed by a period of rehabilitation. It should be noted that the younger the person who is undergoing the operation, the greater the risks are of having to undergo another operation because of the limited lifespan of prostheses.

SCOLIOSIS OR KYPHOSCOLIOSIS :

Curvatures of the spine, such as scoliosis (a three-dimensional deformity) or kyphoscoliosis (a combination of scoliosis and a hunchbacked back) can occur. Although a corset or brace can slow down its progression, sometimes surgery will be necessary to correct it. Several surgeries can be performed. In the most severe cases, it is necessary to perform a surgical fusion which



consists of fixing rods and screws on the spine in order to straighten it. Small pieces of bone are then grafted onto the spine; these will grow along with the spine and merge with it into the correct position. Note that a recovery period of at least six months will be necessary to allow the vertebrae transplant time to take hold.

SPINAL CORD COMPRESSION :

Spinal cord compression, which is present in some little people, can be the cause of serious neurological problems. In infants with achondroplasia, compression may be caused by stenosis of the foramen magnum (the narrowing of the passage that houses the spinal cord at the base of the skull). A neurosurgeon may perform the surgery to enlarge this occipital foramen. The operation, called a laminectomy, is performed under general anesthesia. The surgeon enlarges the spinal canal (where the spinal cord passes) by removing a posterior portion of the vertebrae. They place rods along the involved vertebrae or weld them together with bone tissue grafts. After the operation, rehabilitation is necessary.

INSTABILITY OF THE CERVICAL SPINE :

Many little people, for example those with Morquio syndrome or spondyloepiphyseal dysplasia, have cervical instability that can cause compression of the spinal cord. Complications can be serious (slow and gradual myelopathy, paralysis and even sudden death).
A surgery called cervical spondylodesis corrects cervical instability by fusing the vertebrae. During this procedure, small pieces of bone are screwed into the neck. As they grow, these bones strengthen and stabilize the joint formed by the first two cervical vertebrae and the base of the skull.
After the cervical vertebrae have fused, it is essential to immobilize the neck with a halo brace for three or four months, while the bone graft grows and connects to the base of the skull. The halo brace is a metal ring attached to the surface of the skull; rods from the halo are attached to the body by a corset or brace.

LIMB LENGTHENING :

First of all, you should know that this intervention which is reserved for people with a disproportionate form of dwarfism is currently not recommended by professionals due to the controversies surrounding this procedure, as well as the pain and risk of complications (infections, etc.) that may result.
That being said, as the name suggests, limb lengthening aims to increase the size of the patient's legs and/or arms. The surgery is done at the end of the growth process, on

adolescents or young adults. It can allow a gain of up to twenty centimeters or more for the individual. However, results vary from person to person, so the gains can be much smaller.
There are currently four categories of fixation systems used to lengthen limbs: the Orthofix system, by Wagner, Ilizarov and the Albizzia nail. The surgery is performed under general anesthesia. The orthopedist installs a fixator to hold the bone to be lengthened in a stable position, then cuts the bone widthwise before attaching pins. These pins, connected to a fixator installed on the leg or to a rod that passes through the bone, will then be stretched, by the patient, as the bone is remodelled. So, each time the patient turns the pins, a new section of bone forms to fill the empty space left by the stretch. Lengthening occurs at a rate of 1 millimeter per day.

After the operation, the patient should remain hospitalized for the duration of the lengthening, approximately one month, if there are no complications. After the lengthening, the bone should then heal completely at a rate of one month per centimeter lengthened. Thus, the treatment lasts about 30 months for the lengthening of the lower limbs. If the arms must also be lengthened, the treatment time increases further. Finally, be aware that a long rehabilitation program is to be expected.

RESOURCES:

Association québécoise des personnes de petite taille
https://www.aqppt.org/

Little People of Ontario
https://littlepeopleofontario.com/

Nathalie Boëls, *Le nanisme. Se faire une place au soleil dans un monde de grands*, Montréal, éditions du CHU Sainte-Justine, 2008

Association québécoise des personnes de petite taille, *Recherche médicale sur le nanisme*, Montréal, AQPPT, 1993

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