SLEEP APNEA

DESCRIPTION:

Sleep apnea is a condition that causes repetitive stops in respiratory flow during sleep. These respiratory failures, or "apneas", usually last 10 to 30 seconds, and can occur several times during the night. There are different forms of sleep apnea: obstructive apnea, caused by a blockage in the upper airways; central apnea, caused by certain drugs and pathologies (nervous and heart problems, brain trauma, etc.); and mixed apnea which combines these two causes.

CAUSES:

Certain physical characteristics, which are found in the different types of dwarfism, are conducive to the onset of sleep apnea:

- The shape of the face and neck (thick neck, narrow throat, receding chin, narrow face);
- The reduced size of the respiratory tract and the laxity (relaxation) of the laryngeal cartilage;
- Enlargement of the adenoids and/or tonsils;
- Being overweight or obese.

As a result, little people are predisposed to sleep apnea. For example, in achondroplasia and people with Morquio syndrome, narrowing of the respiratory tract and enlarged adenoids and/or tonsils may be the cause. Individuals with some form of primordial dwarfism, the small size of the face may be the cause.

In other conditions, such as metatropic dysplasia or Maroteaux-Lamy syndrome, lung damage is a predisposition for sleep apnea.

Finally, it is important to note that with achondroplasia, stenosis (narrowing) of the occipital foramen, located at the base of the skull, commonly causes central sleep apnea in infants.

SYMPTOMS:

Nocturnal symptoms involve snoring, restless sleep and night sweats.

During the day, people suffer from morning headaches, fatigue and drowsiness. Problems with concentration and memory impairment are common. Finally, there are behavioral problems, such as hyperactivity, impulsiveness and/or irritability.

Children will also have a tendency to wet the bed. Excessive daytime sleepiness is less common in children than in adults. In adulthood, libido disorders may be present.
POSSIBLE COMPLICATIONS:

The main consequence of sleep apnea is that the brain does not get enough oxygen. In children, this can cause restricted growth, and therefore compromise the extent of expected adult height.

Over time, sleep apnea can cause serious problems. There are particular risks of respiratory failure, hypertension, stroke or heart attack.

In infants with achondroplasia, sleep apnea caused by stenosis of the occipital foramen can be fatal. In 2 to 7% of babies, there is a risk of sudden death during their first year of life. As the occipital foramen gets larger, the risk subsides. It disappears around the age of 5, when this opening reaches its final size.

The risks arise from the fact that the narrowness of the occipital foramen, through which the spinal cord passes, may damage the nerve fibers involved in the control of respiratory functions and heart rate.

TESTS AND TREATMENT:

Primary care physicians (family physicians and pediatricians) who believe that there is a risk that a patient has sleep apnea may recommend a polysomnography. This test involves recording different variables (oxygen saturation, heart rate and respiratory signals) during the patient's nocturnal sleep and monitoring their brain activity to assess the stages of sleep. Polysomnography makes it possible to establish the frequency of obstructive or central pauses in breathing.

When the diagnosis of sleep apnea has been made, several treatment options are available:

- Weight loss;
- Discontinue the use of alcohol, tobacco, sleeping pills, and muscle relaxants;
- Treatment of nasal infections and allergies;
- The nocturnal fitting of an oral appliance (or mandibular advancement device);
- Uvulopalatopharyngoplasty (removal of the uvula and part of the soft palate to clear the respiratory tract);
- Tonsillectomy / adenoidectomy (removal of the tonsils and/or adenoids);
- Opening of the respiratory tract at night using continuous positive airway pressure (CPAP) or bilevel positive airway pressure (BiPAP);
- Enlargement of the occipital foramen (in infants affected by this issue);
- For more severe cases, a tracheostomy (hole in the trachea above the throat) should be considered.

Note: People who are going to have surgery should tell their anesthetist that they have sleep apnea, as anesthesia can sometimes cause the respiratory tract to narrow further.
RESOURCES:

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

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

Little People of America – Sleep Apena, Special Concern for Little People
https://www.lpaonline.org/assets/documents/Sleep%20Apnea%20Article.pdf

ENT Health – American Academy of Otolaryngol-Head and Neck Surgery
https://www.enthealth.org/conditions/pediatric-sleep-disordered-breathing/

Le manuel Merck
https://www.merckmanuals.com/fr-ca/accueil/troubles-pulmonaires-et-des-voies-a%C3%A9riennes/apn%C3%A9e-du-sommeil/apn%C3%A9e-du-sommeil

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