Sarah Bagher (2009)

Central Hypotonia

Definition;

Is a condition of decreased muscle ton. Hypotonia can result from a many central or peripheral causes. Therefore, hypotonia is a phenotype of many clinical conditions with variable prognosis (1).

Central hypotonia originates from the central nervous system, while peripheral hypotonia is related to problems within the spinal cord, peripheral nerves and/or skeletal muscles.

Muscle tone;

It is the resistance to passive movement .It is important to recognize that hypotonia is not equivalent to weakness. Infants with central causes, such as Down syndrome, may have severe hypotonia and normal muscle strength (1).

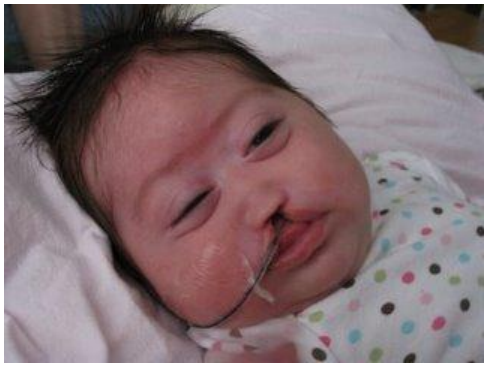
Causes;

Hypotonia are usually of unknown origin. Scientists believe that they may be caused by trauma, environmental factors, or by other genetic, muscle, or central nervous system disorders. A retrospective, cross–sectional study was done in Iran to find out the main cause of hypotonia, they found that central hypotonia was the cause in (94.4%) and peripheral in (3.7%), The most common cause of central hypotonia was idiopathic central hypotonia , followed by cerebral palsy, brain structural abnormality , inborn errors of metabolism , genetic disorders and syndrome (2).

Causes of Central Hypotonia;

- **Idiopathic central hypotonia** (persistent hypotonia without other cause)
- **Hypotonic cerebral palsy**
- **Intracranial haemorrhage**
- **Hypoxic ischemic encephalopathy**(damage to cells in the central nervous system due to inadequate oxygen)
- **Structural brain anomalies:**
  - **Holoprosencephaly** (caused by the failure of the embryonic forebrain to divide into the double lobes of the cerebral hemispheres. The result is a single-lobed brain structure and severe skull and facial defects that may affect the eyes, nose, and upper lip.
- **Metabolic disorders:**

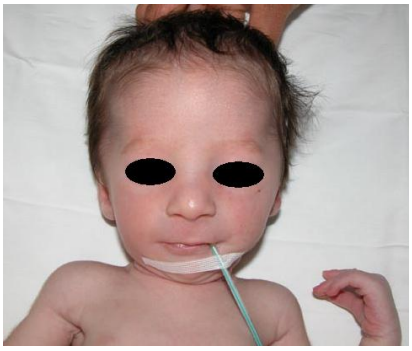
1. **Mitochondrial encephalomyopathies** (chronic progressive disorders affecting neuromuscular system. Symptoms are induced by insufficient energy supply resulting from a deficiency of oxidative phosphorylation.



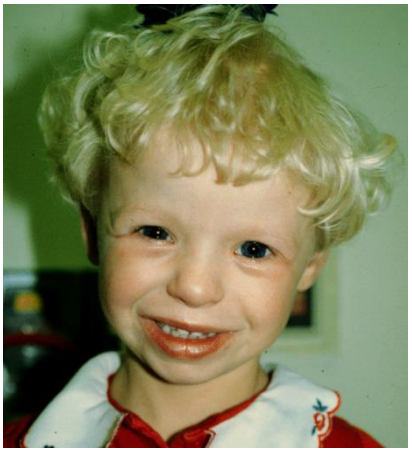
2. **Peroxisomal disorders** (group of congenital diseases characterized by the absence of normal peroxisomes in the cells of the body. Peroxisomes are special organelles within a cell that contain enzymes responsible for critical cellular processes, including oxidation of fatty acids, biosynthesis of membrane phospholipids and cholesterol.

The patient usually has developmental delay and mental retardation . Hypotonia, which in the most severe cases is generalized. Facial abnormalities, including high forehead, frontal bossing (swelling), small face, and slanted eyes.

Chromosomal disorders;



Prader-Willi syndrome (a very rare genetic disorder, in which seven genes on chromosome 15 are missing or unexpressed .They usually have slow mental development, small hands and feet as compared to the body, almond shaped eyes, and skeletal abnormalities, such as a narrow bifrontal skull. Symptoms also include floppy infants

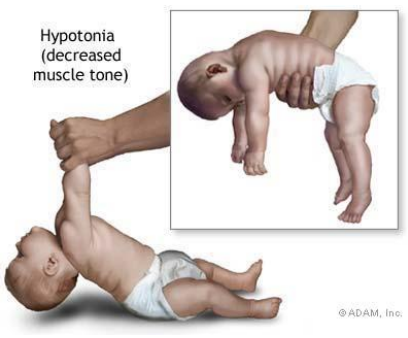


Williams syndrome (a rare neurodevelopmental disorder caused by a deletion of about 26 genes from the long arm of chromosome 7. It is characterized by a distinctive, "elfin" facial appearance, along with a low nasal bridge; mental retardation and cardiovascular problems, such as supravalvular aortic stenosis .

Trisomy21 (down syndrome)

Symptoms of hypotonia;

- Hypotonic children also have trouble feeding and are unable to suck or chew for long periods.
- weak ligaments and joints
- Poor reflexes(mouth hangs open with tongue protruding)
- Failure to acquire motor skill developmental milestones (such as holding head up without support from parent, rolling over, sitting up without support, walking)
- Since the muscles that support the bone joints are soft, there is a tendency for hip, jaw, and neck dislocations.
- breathing difficulties



Diagnosis;

- **History taking**
  History of birth trauma, asphyxia, or infection may predispose to central hypotonia.
  It is important to identify whether the hypotonia was present at birth (congenital) or later on. The distinction between a static course (e.g., due to brain insults) or progressive course (due to dystrophies) is critical. Static causes usually results in slow improvements, while progressive causes results in relentless deterioration.
- **Physical examination**
  Abnormal head size could suggest a central cause.
  Bruising could suggest an acute traumatic etiology.
  By inspection, Central hypotonia is usually associated with abnormal primitive reflex in response to handling (3).
- **Some diagnostic tests may also be used.**
  Computerized tomography scan (CT scan)
- Magnetic resonance imaging (MRI)
- Electroencephalogram (EEG): a test that measures the electrical activity in the brain.
- Karyotype: a test that performs a chromosomal analysis from a blood test, used to determine whether the hypotonia is the result of a genetic disorder
- Muscle biopsy: a sample of muscle tissue removed and examined under a microscope.
- Creatine kinase (CK) test: elevated CK level in blood indicating muscles are damaged or degenerating

Treatment;

Once the correct diagnosis is confirmed, the cause is treated first, followed by symptomatic and supportive therapy for the hypotonia.

- Physical therapy can improve fine motor control and overall body strength.
- Occupational and speech-language therapy can help breathing, speech, and swallowing difficulties.
- Therapy for infants and young children may also include sensory stimulation programs.

Prognosis;

Central hypotonia due to static usually improves with time. The prognosis is worst for hypotonia of progressive central disorders.

Children with mild hypotonia may not experience developmental delay, although some children acquire gross motor skills (sitting, walking, running, jumping) more slowly than most. Most hypotonic children eventually improve with therapy and time. Some children are more severely affected, requiring walkers and wheelchairs and other adaptive and assistive equipment.

Dental concerns;

You must take in consideration that the neuromuscular problems can affect the oral health in several ways.

- Malocclusion, hypotonia of the orofacial muscles result in forward tongue posturing, class II malocclusion, poor swallow reflex and mouth breathing (4).
- The higher prevalence of class II malocclusion with protruded incisors increases the risk of dental trauma (4).
- Sialorrhea (drooling), because of the neuromuscular problems the patient is not able to swallow the saliva .which result in infection, dehydration and perioral inflammation. They also, tend to build up extensive calculus deposits.
- The higher calculus deposit and plaque index together with mouth breathing will help to increase the caries rate in these patients (5).

During dental treatment;

- Determine the best position for your patient in the dental chair and the safest way to move his or her body, especially the head and neck. Talk with the physician about ways to protect the spinal cord. Use pillows to stabilize your patient and make him or her more comfortable.
- In a study done to compare various management techniques in patients with CP during their dental treatment they found that he use of assistive stabilization reduces the number of patient referred to general anesthesia(6).
- Maintain a clear path for movement throughout the treatment setting.

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