Hypotonia (pediatrics)

Hypotonia is pathologically reduced muscle tone, i.e. lower resistance during passive muscle stretching. It is necessary to distinguish hypotonia from reduced muscle strength, which corresponds to active muscle involvement (classification 0-5 according to the Medical Research Council).

Sometimes the terms hypotonia and muscle weakness are used interchangeably to emphasize the opposite of spasticity. At other times, on the other hand, the term spasticity is included in the group of diseases manifested by "muscle weakness", since the spastic patient is also unable to use the muscles properly, and at the same time, diseases manifested by spasticity usually begin with hypotonia, they manifest by spasticity over time.

Symptoms and examination

- scarf sign;
- *floppy baby syndrome*;
- „f*rog-leg posture*“.

(On the contrary, spasticity is manifested by a flexed position: on the upper limbs, flexion in the elbow, and shortening of the tendons of the forearm, on the lower limbs, by shortening of the Achilles tendons, which brings the instep in line with the tibia.)

Classification

The character of the present hypotonia

Symptom localization

Hypotonia can be expressed in all muscle groups, or only some muscle groups can be affected: **proximal, distal, ophthalmoplegia, facial weakness, and bulbar weakness.**

Location of the lesion

- disorder at the central level, at the spinal or peripheral level, at the neuromuscular disc, and at the muscle level
- iatrogenic

Muscle weakness

- with or without muscle weakness

Floppy baby syndrome

Diseases in which an infant lying on its back moves normally, but when lifted there is significant hypotonia, are referred to as a floppy baby syndrome. When lying down, they raise their limbs against gravity without difficulty, they have a normal facial expression. After being raised, their head falls, they fall with their hands and when they turn onto their stomach they take the **Landau position** (an inverted U position). When placed on the mat on their stomachs, they can have their limbs loosely placed on the mat instead of tucked under them.

It can be a benign syndrome that will go away, but it can also be a serious brain disability. In terms of differential diagnosis, it is important whether it is hypotonia with muscle weakness or without muscle weakness and whether consciousness is affected.

Hypotonia with muscle weakness

- with preserved consciousness
  - neuromuscular disease: spinal muscular atrophy, myasthenic syndromes, congenital neuropathy or myopathy
  - spinal cord involvement: tumor, spinal cord infarction, malformation, spina bifida, syringomyelia
- with impaired consciousness:
  - severe brain damage
  - hydrocephalus
  - infection
  - metabolic disorders (anoxia, hypoglycemia, hyperbilirubinemia with kernicterus)
  - intoxication (transfer from the mother, e.g. anesthesia during childbirth, benzodiazepines, narcotics, magnesium sulfate)

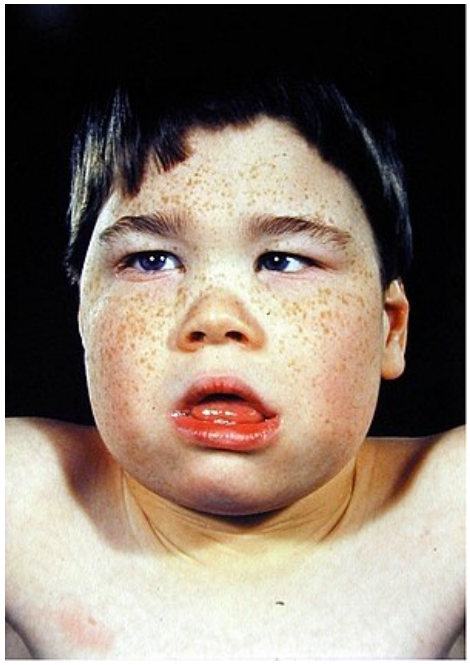
Hypotonia without muscle weakness

- acute systemic disease
- Down's syndrome
- mental retardation
- Prader-Willi syndrome
- Ehlers-Danlos syndrome
- Marfan syndrome
- rickets
- renal tubular acidosis
- celiac disease
- biliary atresia
- congenital heart defect
- benign congenital hypotonia

Other diseases manifested by hypotonia

Impairment of the central motor neuron

- hypotonic cerebral palsy
- intracranial bleeding
- tumors
- infection
- demyelination processes
- metabolic disorders
- degenerative processes



The face of a child with hypotonia with ATR-X syndrome

Impairment of the peripheral motor neuron

- transverse spinal cord lesion
- transverse myelitis – proximal and distal motor defect, sensory defect, urinary and stool incontinence, and severe local pain
- poliomyelitis
- **spinal muscular atrophy - the Werdnig-Hoffmann type** – initially proximal weakness, floppy baby, reduction of spontaneous movement, marked atrophy, progression to weak quadriplegia, the disappearance of facial expressions, respiratory difficulties; lateral fasciculation of the tongue especially noticeable during sleep; mental, social and language development is normal; causal treatment does not exist, prevention of aspiration, infections, treatment of contractures, scoliosis, social and symptomatic therapy; intrauterine development is possible already or in the first weeks of age, but typically between 6 months and 6 years of age; may be slow or rapid progression; AR disease caused by a mutation in the SMN1 gene ("spinal motor neuron 1"), the frequency of the mutated allele in the population is 1:50; an almost identical SMN2 gene at another location in the genome modulates the severity of phenotypic manifestations
- **spinal muscular atrophy - Kugelberg-Welander type** – proximal muscle weakness; a milder form of spinal muscular atrophy appearing in later childhood or adolescence

Impairment of the neuromuscular disc

- myasthenia gravis – distal involvement, facial involvement, bulbar syndrome, ophthalmoplegia
- botulism – distal involvement, facial involvement, bulbar syndrome, ophthalmoplegia

Neuropathy

- polyneuropathy of the Guillain-Barré type - distal involvement
- Miller-Fisher variant of Guillain-Barré syndrome - facial involvement and bulbar syndrome
- Charcot-Marie-Tooth
- tick paralysis
- secondary peripheral neuropathy – more typical for the adult population (diabetes mellitus, alcoholism, chronic kidney disease, toxins, vasculitis, paraneoplastic)

Muscle diseases

- Duchenne muscular dystrophy - proximal muscle weakness
- limb-girdle muscular dystrophy - proximal muscle weakness
- myotonic dystrophy – distal muscle weakness, facial involvement, bulbar syndrome
- distal myopathy – distal muscle weakness, ophthalmoplegia
- congenital myopathy - facial involvement, bulbar syndrome
- facioscapulohumeral dystrophy – facial involvement, bulbar syndrome
- dermatomyositis – proximal muscle weakness
- polymyositis – proximal muscle weakness

Links

References

- MARCDANTE, Karen J, Robert M KLIEGMAN a Richard E JENSON, et al. *Nelson essentials of pediatrics.* 6. vydání. Philadelphia : Saunders/Elsevier, 2011. 831 s. s. 683–691. ISBN 978-1-4377-0643-7.