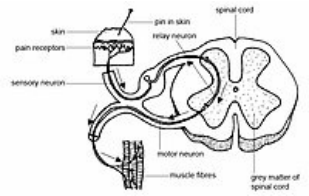
Motorneuron Diseases

Motorneuron diseases (MND) are progressive neurodegenerations, which affect the movement ability (walking, chewing, moving your limbs,breathing). According the place of damage we recognize the upper and lower motorneurons diseases. Generally we can say that these diseases lead to muscle weakness, instability, fasciculations or spasticity, which annoy the patients. Typical patient is an adult **over 40 years**, twice as many men than women. However, we can give them just a supportive treatment, because motorneurons diseases are **incurable**.

Etiology

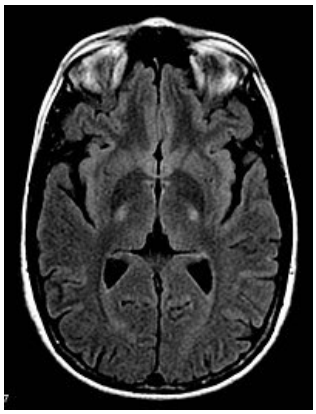


Motor neurons in a spinal cord

We still don´t know exactly what is the etiology of **non-inherited MND**. As usual, we assume the influence of genetic predisposition, viruses, enviromental or toxic. A prolonged used of some toxic drugs or cancers could be also very important trigger points of MND. It is hard to find out something about the etiology when we don´t know much about a patogenesis. The only certain thing is a presence of the neurodegeneration and the death of neuron cells.

The **inherited MND** are divided into three groups: *autosomal dominant*, *autosomal recessive* and *X-linked forms*.

Classification


MRI-Amyotrophic lateral sclerosis

1. **Upper motorneurons diseases**: primary lateral sclerosis
2. **Lower motorneurons diseases**: progressive bulbar palsy, progressive muscular atrophy
3. **Combinate motorneurons diseases**: amyotrophic lateral sclerosis

Symptoms

The symptoms of MND are so variable and different for each of disease, that it is hard to describe them generally, so please read also the seperate articles of MND. Here is just a list of the most common symptoms:

- muscle weakness and atrophy
- spasticity
- muscle cramps and fasciculations
- swallowing and speaking problems
- weakness in facial muscles (*hypomimia*)

Diagnosis

There is **no specific test** for MND and their symptoms are usually not as specific as they should be, so to diagnose them well is pretty hard work. The common neurological examination can give us some basic information of motorneurons damage, but after that we have to use specific methods.

Electromyography (EMG) can inform us about the speed of nerve impulses transfer, so we can find damage of spinal cord or nerve and muscle dysfunction. If there is any affection of the upper motorneurons we should find more diffuse damage as compared with lower motoneurons.

MRI can help us to exclude or confirm brain or spinal cord tumors, multiple sclerosis, brain injuries and trama or some inflammation, which can be also detect from **the blood tests** (CRP, antibodies). Tests of the blood, urine or cerebrospinal fluid can help us in differential diagnosis. Especially some enzymes - as *creatine kinase* - are typical for muscle diseases (muscular dystrophy).

The first special method for the motor neurons examination was **transcranial magnetic stimulation** and later **magnetic resonance spectroscopy** (for upper motorneurons).

The last options is **muscle or nerve biopsy** (needle biopsy), which we should choose just in case that there is no better choice, because it is pretty invasive and unpleasant for the patients.

Therapy

The is no specific treatment, so we can offer just the symptomatic and supportive one, which can help the patients live with their disabilities. Regular **physical therapy** and rehabilitation can prevent some contractures or muscle atrophy.

People with MND very often suffer from swallowing problems, so we have to give them **balanced diet** and right **nutrition** (feeding tube or PEG). Speech therapy is also one from the symptomatic treatment method.

There are also few **medications**, which we can prescribe: muscle relaxants (*baclofen*), benzodiazepines (against spasticity), phenytoin (against cramps), nonsteroidal anti-inflammatory drugs and in terminal stages also opiates (against pain).

Each of MND has its own special symptoms that we should cure, but generally we have to say that these diseases are still **incurable**. Some of them develop slowly and don´t annoy their carrier so much, but the others can be very progressive and lead to death within few months or years. It depends on the time of onset and on the severity of the disease.

Links

Related articles

- Neurodegeneration
- Motor Neurons
- Hypertonia
- Amyotrophic Lateral Sclerosis
- Primary Lateral Sclerosis
- Progressive Bulbar Palsy
- Progressive Muscular Atrophy
- Electromyography
- MRI
- Spinal Cord

External links

- National Institute of Neurological Diseases and Stroke (http://www.ninds.nih.gov/disorders/motor_neuron_diseases/detail_motor_neuron_diseases.htm)

Bibliography

- JEDLIČKA, KELLER,. *Speciální neurologie.* 1st edition. 2005. ISBN 80-7262-312-5.
- AMBLER,. *Základy neurologie.* 7th edition. 2011. ISBN 978-80-7262-707-3.