

Myopathy

Myopathies are neuromuscular diseases affecting primarily skeletal muscles. They are usually *symmetrical* and mainly affect the limbs girdle but it can also be an *oculopharyngeal* or *fascioscapular* form.

The main clinical picture is proximal **muscle weakness**. We also observe myopathic "**duck**" **walking** due to weakened gluteal muscles. The patient does not keep the pelvis horizontal, does not stand up without help and we observe the so-called **Gower sign** - myopathic climbing. Other features are **hyperlordosis**, **atrophy**, **hypotrophy** (thigh muscles) or **hypertrophy** (rather *pseudohypertrophy* = muscle turns into ligament or fat replacement of muscle), decreased reflexes, no sensory hypoesthesia.

Etiology

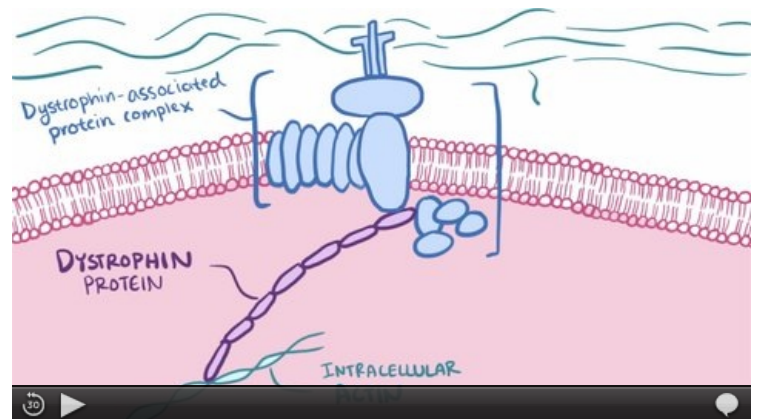
The following diseases can be included among myopathies :

Genetically determined myopathies

- Duchenne muscular dystrophy
- Becker muscular dystrophy

Acquired myopathies

- **inflammatory** - polymyositis, dermatomyositis;
- **endocrine/ metabolic conditioned** - hypo/hyperthyroidism, Cushing, hypokalaemia
- **toxic and drug-induced** myopathies - alcohol, corticoids, statins, antirheumatics/ anti-inflammatory/ immunosuppressants - colchicine, steroids, D-penicillamine, chloroquine
- **myotonia** - do not manifest as a typical myopathic syndrome, but hypertension dominates



Video in English about Duchenne muscular dystrophy and Becker muscular dystrophy

The clinical picture

The clinical picture is dominated by **muscle weakness**. **Muscle reflexes** may be **reduced** but not extinguished. *Muscle pain* may or may not be present depending on the specific cause. Muscle involvement is usually **symmetrical** and (unlike neuropathies) mainly on **the proximal girdle muscles**. This gives rise to a typical **myopathic syndrome** which includes:

- myopathic ("duck") gait - while walking, the pelvis on the side of the foot drops, which is just taking a step (normally the pelvis on this side rises to relieve the foot)
- hyperlordosis of lumbar spine with protruding abdomen
- myopathic climbing - when getting up from a squat, the patient help each other with their hands

Patients typically have difficulty walking up the stairs and getting up from chair. When their upper limbs are affected, they have trouble hanging clothes and brushing their hair.

Myasthenia and myopathy comparison

Parameter	Myasthenia	Myopathy
Name	astenia = weakness	patia = disorder
Disability	neuromuscular disc	muscle
Location	generalized	proximal limbs
Sensory impairment	no	no
Pain	no	yes
Atrophy	no	yes
Fasculation	no	no
Proprioceptive feeling	standard	standard or reduced
Tone	standard	reduced

Diagnosis

- EMG – on conventional EMG we see the activation of a large number of motor units at a given load than in normal muscle
- serum muscle enzymes – increased CK and myoglobin
- muscle biopsy – atrophy, hypotrophy, pseudohypertrophy;
- dystrophin protein deficiency
- EKG, echo;
- spirometry – respiratory muscles

References

Related Articles

- Statin myopathy
- Myasthenia gravis
- Myotonic syndrome
- Myositis

References

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- KÁBRT, Jan, et al. *Lexicon medicum*. 1. edition. 1988. ISBN 08-063-88.