## **Motor neuron diseases**

A motor neuron disease (MND) is any of several neurodegenerative disorders that selectively affect motor neurons, the cells that control voluntary muscles of the body.

Three patterns of involvement:

1. mixed upper motor neurons (UMN)s & upper motor neurons (UMN)s degeneration: amyotrophic lateral sclerosis (ALS). The most common of the motor neuron diseases

2. UMN degeneration: primary lateral sclerosis. Rare, onset after age 50. No LMN signs. Slower progression than ALS (yrs to decades). Pseudobulbar palsy is common.

Usually does not shorten longevity. May present with falling due to balance problems or low back and neck pain due to axial muscle weakness

3. LMN degeneration: progressive muscular atrophy (PMA) and spinal muscular atrophy (SMA)

They include amyotrophic lateral sclerosis (ALS), hereditary spastic paraplegia (HSP), primary lateral sclerosis (PLS), progressive muscular atrophy (PMA), progressive bulbar palsy (PBP) and pseudobulbar palsy. Spinal muscular atrophies (SMA) are sometimes included in the group by some neurologists but it is a different disease with a clear genetic cause.

## **Differential diagnosis**

The immunological pathophysiologies of chronic inflammatory demyelinating polyneuropathy (CIDP) and multifocal motor neuropathy (MMN) differ considerably, but neither has been elucidated completely. Quantitative magnetic resonance imaging (MRI) techniques as diffusion tensor imaging, T2 mapping, and fat fraction analysis may indicate in vivo pathophysiological changes in nerve architecture.

A study of van Rosmalen et al. aimed to systematically study nerve architecture of the brachial plexus in patients with CIDP, MMN, motor neuron disease (MND) and healthy controls using these quantitative MRI techniques.

They enrolled patients with CIDP (n = 47), MMN (n = 29), MND (n = 40) and healthy controls (n = 10). All patients underwent MRI of the brachial plexus and obtained diffusion parameters, T2 relaxation times and fat fraction using an automated processing pipeline. They compared these parameters between groups using a univariate general linear model.

Fractional anisotropy was lower in patients with CIDP compared to healthy controls (p < 0.001), patients with MND (p = 0.010) and MMN (p < 0.001). Radial diffusivity was higher in patients with CIDP compared to healthy controls (p = 0.015) and patients with MND (p = 0.001) and MMN (p < 0.001). T2 relaxation time was elevated in patients with CIDP compared to patients with MND (p = 0.023). Fat fraction was lower in patients with CIDP and MMN compared to patients with MND (both p < 0.001).

Thee results show that quantitative MRI parameters differ between CIDP, MMN and MND, which may reflect differences in underlying pathophysiological mechanisms <sup>1)</sup>.

## 1)

van Rosmalen M, Goedee HS, Derks R, Asselman FL, Verhamme C, de Luca A, Hendrikse J, van der Pol WL, Froeling M. Quantitative MRI of the brachial plexus shows specific changes in nerve architecture in CIDP, MMN and motor neuron disease. Eur J Neurol. 2021 May 1. doi: 10.1111/ene.14896. Epub ahead of print. PMID: 33934438.

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