Motor Unit Number Index (MUNIX) detects motor neuron loss in pre-symptomatic muscles in Amyotrophic Lateral Sclerosis

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OBJECTIVE
Motor Unit Number Index (MUNIX) is a quantitative neurophysiological measure that provides an index of the number of lower motor neurons supplying a muscle. It reflects the loss of motor neurons in patients with Amyotrophic Lateral Sclerosis (ALS). However, it is unclear whether MUNIX also detects motor unit loss in strong, non-wasted muscles.

METHODS
Three centres measured MUNIX in 49 ALS patients every three months in six different muscles (abductor pollicis brevis, abductor digiti minimi, biceps brachii, tibialis anterior, extensor digitorum brevis, abductor hallucis) on the less affected side. The decline of MUNIX in initially non-wasted, clinically strong muscles (manual muscle testing, MMT grade 5) was analysed before and after onset of weakness.

RESULTS
In 49 subjects, 151 clinically strong muscles developed weakness and were included for analysis. The average monthly relative loss of MUNIX was 5.0% before and 5.6% after onset of weakness. This rate of change was significantly higher compared to ALS functional rating scale (ALSFRS-R) and compound muscle action potential (CMAP) change over 12 months prior to the onset of muscle weakness (p=0.024).

CONCLUSION
MUNIX is an electrophysiological marker that detects lower motor neuron loss in ALS, before clinical weakness becomes apparent by manual muscle testing.

SIGNIFICANCE
This makes MUNIX a good biomarker candidate for disease progression and possibly pharmacodynamics responds.
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