Motor Unit Number Index (MUNIX) of hand muscles is a disease biomarker for adult spinal muscular atrophy

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OBJECTIVE
There is still insufficient knowledge about natural history in adult spinal muscular atrophy, thus valid markers for treatment and disease monitoring are urgently needed.

METHODS
We studied hand muscle innervation pattern of 38 adult genetically confirmed 5q spinal muscular atrophy (SMA) patients by the motor unit number index (MUNIX) method. Data were compared to healthy controls and amyotrophic lateral sclerosis (ALS) patients and systematically correlated to typical disease-relevant scores and other clinical as well as demographic characteristics.

RESULTS
Denervation of hand muscles in adult SMA was not evenly distributed. By calculation of the MUNIX ratios, we identified a specific hand muscle wasting pattern for SMA which is different to the split hand in ALS. Furthermore, MUNIX parameters strongly correlated with established disease course parameters independent of disease stages.

CONCLUSION
We found a pathophysiological remarkable denervation pattern of hand muscles, a 'reversed split hand'. MUNIX of single hand muscles correlated well with disease severity and thus represents an easily available biomarker for adult SMA.

SIGNIFICANCE
Our data show the power of the MUNIX method as a biomarker for upcoming questions in adult SMA.

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