

Quantification of mobility impairment and self-assessment of stiffness in patients with Myotonia Congenita

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In order to develop reliable and valid functional measurement instruments to quantify myotonic stiffness and mobility impairment in the clinical assessment of patients with myotonia congenita, this study was performed during 2004.

Test-retest reliability and responsiveness in two functional measuring instruments, Timed Up&Go (TUG) and Timed-Stands Test (TST), and three self-assessment scales, Visual Analogue Scale (VAS), Borg's Category-Ratio Scale (BorgCR10) and Myotonia Behaviour Scale (MBS) were evaluated together with treatment efficacy of an antimyotonic drug.

Six male patients (age 31-61, median 40 years) followed a standardised protocol with time scoring, following warming-up and rests, on two separate occasions, with and without drugs. Time scoring of TUG and TST and self-assessments of stiffness were performed at the clinic. A fourteen-day diary was used at home, with self-assessment of stiffness in the morning and stiffness in general.

The functional measurement instruments TUG and TST showed very good test-retest agreement ($ICC=0.87-0.95$) and significant sensibility to change ($p=0.005$ and 0.001 respectively). One reason to prefer the TUG to the TST in MC is that the TUG consists of a sequence of different movements. It is thus likely that the TUG could better reflect the myotonia-induced muscle stiffness in MC, as different muscle groups are enrolled during the test. All self-assessment scales revealed excellent responsiveness and good test-retest reliability, according to the rank-invariant method.

When considering objective results, clinical factors and experiences given by the study, three instruments are favoured; Timed Up&Go and BorgCR10 for short, and MBS for long-term evaluations of treatment effects in Myotonia Congenita.

Keywords: Functional measuring instrument, self-assessment, reliability, sensibility to change, Timed Up & Go, Timed-Stands Test, VAS, BorgCR10, Myotonia Behaviour Scale.