

Validity and Reliability of a New Clinical Myotonia Rating Scale for Non-Dystrophic Myotonia

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Introduction

- The severity of myotonia is difficult to assess without standardized and validated tools.
- This study assessed the validity and reliability of the Clinical Myotonia Rating Scale (CMRS), a novel measure for the impact of myotonia.

Methods

CMRS components

Myotonia severity scale

| Body area | Severity (S) | Frequency (F) | Formula (score range) |
|------------------------|--|---------------------------|---------------------------------------|
| Eyelids | 0 (none) to 4 (severe, permanent) | 0 (none) to 4 (every day) | $S \times F \times 0.5$ (0 to 8) |
| Eyes | 0 (normal) to 4 (severe, diplopia) | 0 (none) to 4 (every day) | $S \times F \times 0.5$ (0 to 8) |
| Chewing and swallowing | 0 (normal) to 4 (unable to chew, choking) | 0 (none) to 4 (every day) | $S \times F \times 0.5$ (0 to 8) |
| Upper limbs | 0 (none) to 4 (severe, permanent) | 0 (none) to 4 (every day) | $S \times F$ [left + right] (0 to 32) |
| Lower limbs | 0 (none) to 4 (severe, permanent) | 0 (none) to 4 (every day) | $S \times F$ [left + right] (0 to 32) |
| Respiratory muscles | 0 (no difficulty) to 4 (permanent dyspnea) | 0 (none) to 4 (every day) | $S \times F$ (0 to 16) |
| | | Total | 104 |

Disability scale

| Disability scale | Scoring | Score range |
|------------------|--|-------------|
| Talking | 0 (normal) to 4 (incomprehensible) | 0 to 4 |
| Writing | 0 (normal) to 4 (unable to handle pen) | 0 to 4 |
| Eating | 0 (normal) to 3 (dependent on others) | 0 to 3 |
| Hygiene | 0 (normal) to 4 (requires 100% help) | 0 to 4 |
| Dressing | 0 (normal) to 4 (requires 100% help) | 0 to 4 |
| Walking | 0 (normal) to 4 (wheelchair) | 0 to 4 |
| Stair climbing | 0 (normal) to 4 (impossible) | 0 to 4 |
| | Total | 27 |

Validity and reliability testing in the MYOMEX trial

- The CMRS was evaluated in patients with myotonia congenita (MC) and paramyotonia congenita (PMC) during the randomized, crossover, double-blind MYOMEX trial of mexiletine versus placebo.¹
 - Two investigators used the CMRS to assess myotonia severity at baseline; this was repeated by one of them at the end of each treatment period in MYOMEX.
- Interrater reliability was estimated by weighted Kappa coefficients.
- Intraclass correlation coefficients (ICC) were calculated for global scores (GS). Bland-Altman methods were also used (data not shown).
- Spearman correlation coefficients were estimated for correlations with the stiffness score using a visual analogue scale (VAS) and the Individualized Neuromuscular Quality of Life (INQoL) self-questionnaire.

Results

- Patients with MC (n=13) and PMC (n=12) were evaluated at six centres in France.
- Kappa** ranged from -0.02 to 0.82, with **most showing fair/good or excellent** interrater reliability.²
 - Severity:** Highest interrater agreement for frequency of eyelid blinking (0.73) and severity of respiratory muscle intensity (0.72).
 - Disability:** Highest interrater agreement for hygiene (0.82) and getting dressed (0.73).
- Intra-class correlation coefficient** severity (0.54) and disability scores (0.65) indicated moderate interrater reliability.³

Weighted kappa coefficient (95% CI)

Severity score

Frequency

| | |
|---------------------|--------------------|
| Eyelids | 0.73 (0.54, 0.91) |
| Eyes | 0.62 (0.40, 0.84) |
| Chewing/swallowing | 0.67 (0.44, 0.90) |
| Right + left UL | 0.40 (-0.08, 0.88) |
| Right + left LL | 0.58 (0.19, 0.97) |
| Respiratory muscles | 0.61 (0.33, 0.89) |

Severity

| | |
|---------------------|---------------------|
| Eyelids | 0.65 (0.46, 0.84) |
| Eyes | 0.44 (0.18, 0.71) |
| Chewing/swallowing | 0.60 (0.38, 0.81) |
| Right UL | -0.02 (-0.07, 0.03) |
| Left UL | 0.35 (-0.04, 0.73) |
| Right + left LL | 0.23 (-0.02, 0.48) |
| Respiratory muscles | 0.72 (0.45, 0.98) |

Disability score

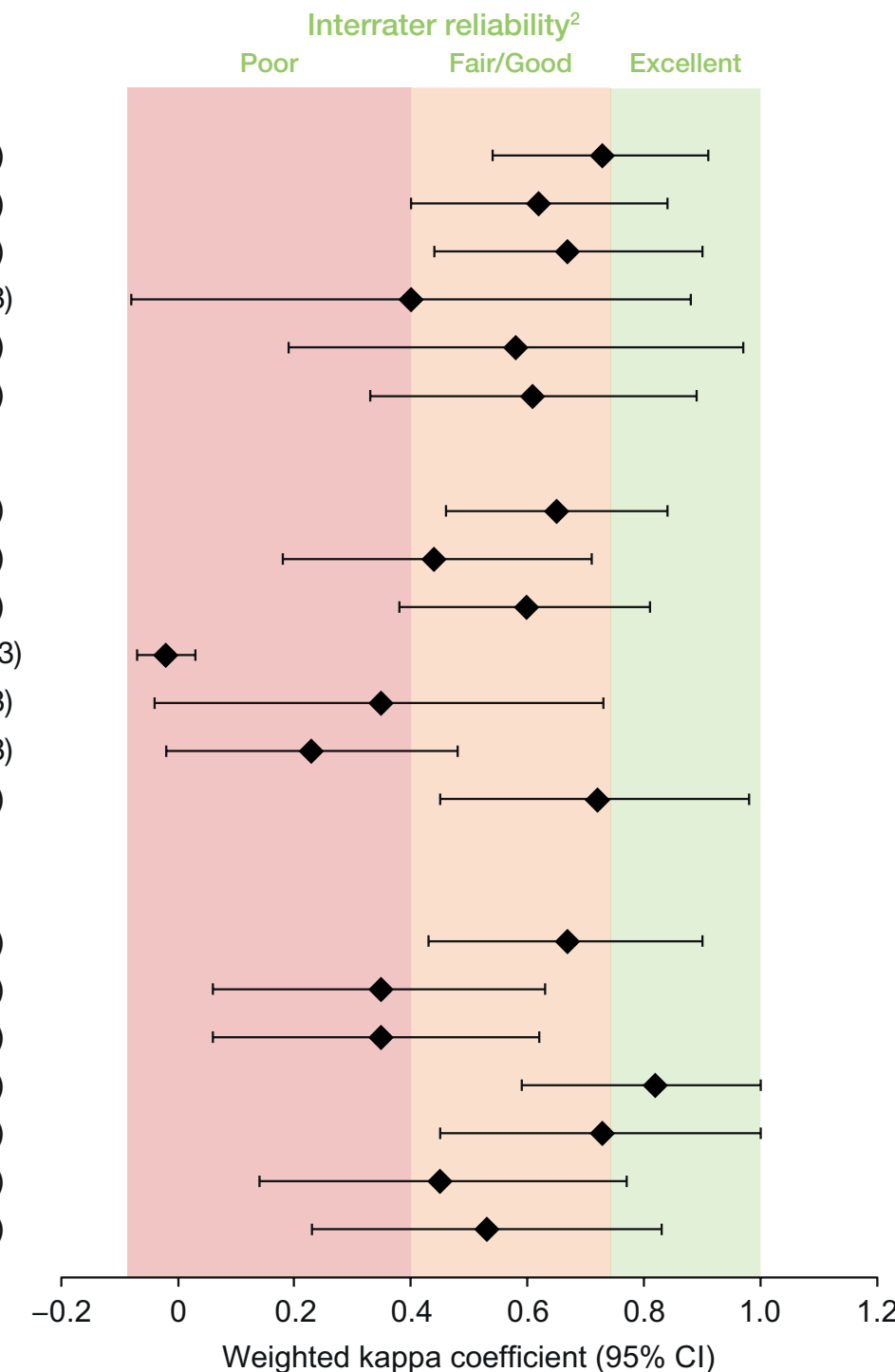
| | |
|----------------|-------------------|
| Speech | 0.67 (0.43, 0.90) |
| Writing | 0.35 (0.06, 0.63) |
| Eating | 0.35 (0.06, 0.62) |
| Hygiene | 0.82 (0.59, 1.00) |
| Dressing | 0.73 (0.45, 1.00) |
| Walking | 0.45 (0.14, 0.77) |
| Stair climbing | 0.53 (0.23, 0.83) |

Severity score

0.54

Disability score

0.65



CI, confidence interval; ICC, intra-class correlation coefficient; LL, lower limb; UL, upper limb

- Severity global score** strongly correlated with both **VAS** (0.70; $p \leq 0.001$) and **INQoL** (0.67; $p \leq 0.001$) scores.

| | CMRS | INQoL | VAS |
|-------------------------|-------------------------|-------------------------|-------------------------|
| | Severity global score | Disability global score | Quality of life |
| Severity global score | 1 | 0.73 ($p \leq 0.001$) | 0.67 ($p \leq 0.001$) |
| Disability global score | 0.73 ($p \leq 0.001$) | 1 | 0.47 ($p \leq 0.001$) |
| | | | Stiffness score |
| | | | 0.70 ($p \leq 0.001$) |
| | | | 0.69 ($p \leq 0.001$) |

CMRS, Clinical Myotonia Rating Scale; INQoL, Individualized Neuromuscular Quality of Life; VAS, visual analogue scale

Conclusions

- The CMRS is a promising scale for assessing the severity and impact of myotonia in patients with NDM.
- In this small exploratory analysis, the CMRS scoring system demonstrated moderate interrater reliability.
- The CMRS will undergo further validation in study populations with myotonic disorder.

References

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