Revised Hammersmith Scale for Spinal Muscular Atrophy (RHS): Inter & Intra-Rater Reliability in a UK cohort of Neuromuscular Physiotherapists & Content Validity from a Patient Perspective

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Background:

Prior to adopting the Revised Hammersmith Scale for SMA (RHS) for use in clinical trials and wider clinical practice, further investigation is required regarding its reliability and validity properties. SMA REACH UK have undertaken preliminary validity and reliability testing of the scale within two studies.

An essential component of scale development is determining how reliable a scale is. Since the inter and intra-rater reliability properties have not previously been determined we undertook a reliability study in a cohort of UK Neuromuscular Physiotherapists.

Development of the RHS has been driven by expert clinicians validating content identified as relevant to functional activities at each re-draft. However a key aspect of scale development is to identify whether the items included reflect meaningful real life activities from a patient/parent perspective. Therefore a small scale qualitative study addressing RHS content validity from a patient/parent perspective was undertaken.

Aims:

Study 1 – Establishing the inter and intra-rater reliability of the RHS:
- To determine the inter and intra-rater reliability of the RHS in a UK cohort of physiotherapists with experience in neuromuscular diseases.
- To provide clinically meaningful interpretation of reliability measures.

Study 2 – Determining the content validity of the RHS from a patient/carer perspective:
- To identify whether each item on the RHS relates to real life activities and what meaning people with SMA/families of those with SMA give to each item.

Methods 1:

- Sample: Neuromuscular Paediatric Physiotherapists from the North Star Clinical Network, with at least one SMA patient on their current clinical caseload.
- Reliability testing was conducted via two online surveys. Raters viewed and completed two RHS assessment videos, one of a patient with SMA type 2 and one of a patient with SMA type 3, the same assessments were repeated in a second survey two weeks later to establish intra-rater reliability.
- Inter-rater reliability was analyzed using a Type 2 Intraclass Correlation Co-efficient (ICC) for Absolute agreement, and intra-rater reliability was analyzed using Bland Altman (BA) Limits of Agreement (LOA) and plots.
- The acceptable inter and intra-rater variability was set as a change of ± 2 points in the total score by the international expert panel who developed the RHS.

Methods 2:

- Sample: SMA REACH UK registered patients with SMA 2 and 3 and their parents/guardians.
- Two patient/parent focus groups were held by SMA REACH UK in September and March 2015, London.
- The SMA REACH UK Physiotherapist presented to the group each item of the RHS and facilitated discussion. For each item participants were asked to explain what each item meant to them, and what activity it could represent/be useful for. Participants fed back their answers in a variety of methods, spoken, written on post-it notes or in the form of a structured work book.
- Finally participants rated the 3 most important RHS items to them.

Results 2:

- Participants: n = 19 (SMA 2 = 6 families, SMA 3 = 3 families), 4 children with SMA contributed the patient perspective (SMA 2 = 1, SMA 3 = 3).
- Qualitative data was obtained on content validity from a patient and parent perspective for all 36 items of the RHS, please see examples of data obtained in figure 5.
- Some items were identified as not having much meaning for the patients/parents but identified as a 'good exercise' by participants to be measured by the physiotherapist.

Results 1:

Inter-rater Reliability:

- Participants: 85.7% response rate, n = 18, median 15.25 years (IQR 9.6 to 25.3) practicing as physiotherapists, median 10 years (IQR 3.5 to 14.1) assessing/treating patients with neuromuscular diseases, number of patients assessed with SMA in the last year median 10 (IQR 5 to 16.3).
- Inter-rater agreement: Type 2 ICC 0.997 with 97.1% of scores within the acceptable ± 2 limit, table and figure 1.

Table 1: Inter-rater reliability results n = 18

	Mean Total Score (95% CI)	Median Total Score	Range either side of mean	Type 2 ICC (95% CI)	Distribution of RHS Total Score around the Mean			
					± 3	± 2	± 1	± 0
SMA 2	13.22 (12.71 to 13.74)	13	-2 to +2	-	100	100	83.3	27.8
SMA 3	41.28 (40.73 to 41.82)	41	-2 to + 3	-	100	94.4	83.4	38.9
Overall	-	-	-2 to +3	0.997 (0.98 to 1.0)	100	97.2	83.3	33.3

Figure 1: Inter-rater reliability results n = 18
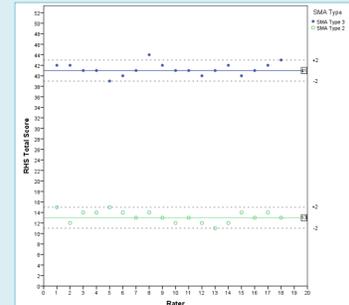
Intra-rater Reliability:

- Participants: 94.4% response rate, n = 17, experience not significantly different from inter-rater testing.
- SMA 2 intra-rater difference: mean -0.61, lower Limits of Agreement (LOA) -2.41 and upper LOA +2.29, table and figure 2.
- SMA 3 intra-rater difference: mean -0.18, lower LOA -2.79 and upper LOA +2.44, table 2 and figure 3.
- 97.1% of the actual values for SMA 2 and 3 intra-rater difference sat within ± 2 limit, figure 4.

Table 2: Intra-rater reliability results n = 17

	Mean Total Score (95% CI)	Median Total Score	Mean Intra-rater Difference (95% CI)	Range of Difference	BA Lower LOA (95% CI)	BA Upper LOA (95% CI)	Distribution of Difference around Mean (%)			
							± 3	± 2	± 1	± 0
SMA 2	13.24 (12.69 to 13.78)	14	-0.06 (-0.63 to 0.51)	-2 to +3	-2.41 (-3.39 to -1.42)	+2.29 (1.30 to 3.27)	100	94.1	82.4	47.1
SMA 3	41.53 (40.99 to 42.06)	42	-0.18 (-0.81 to 0.46)	-2 to +2	-2.79 (-3.89 to -1.69)	+2.44 (1.34 to 3.54)	100	100	70.6	17.6
Overall	-	-	-	-2 to +3			100	97.1	76.5	32.4

Figure 2: SMA2 Intra-rater difference Bland Altman Plots with set (grey) and actual (red) Limits of Agreement
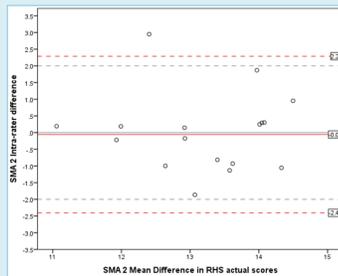

Figure 2: SMA3 Intra-rater difference Bland Altman Plots with set (grey) and actual (red) Limits of Agreement
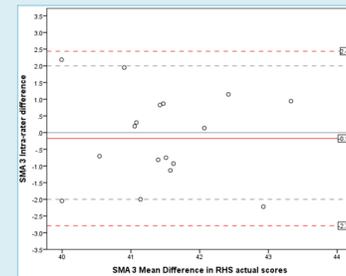

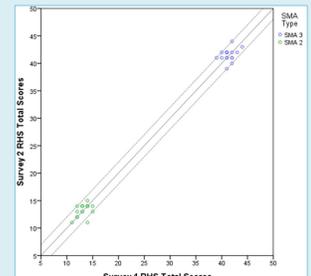Figure 4: Intra-rater difference survey 1 versus survey 2 n = 17

Figure 5: Example of qualitative content validity data from a patient/parent perspective for items on the RHS

Weaker Type 2 ——— **RHS** ——— **Stronger Type 3**

Item 2: Bringing Hands to Head

What it means to you?
'Hygiene & independence'

'Helps re-assure me that he can eat, drink, still got arm strength'

'Real basic function for quality in many activities'

'Can access head & play games'

Activities?
'Combing hair, pouring water, feeding'

'Able to eat, put food in mouth, able to wipe/scratch face'

'Brushing teeth, hair, apply make-up'

'Put glasses on and off, brush teeth'

Item 14: Lying to Sitting

What it means to you?
'Independence in daily life'

'To feel independent to bring yourself to position enabling activity, not waiting lying helplessly for someone to come'

'Emotionally it's sad and painful when she asks me to do this'

'Able to sit up when wakes up, less work/stress for us'

Activities?
'Useful for morning, for school'

'Getting up in the morning, could sit up in bed when he wakes up rather than wait for us'

Getting up/out of bed, sitting up on changing table/sofa'

'To obtain object, to reach, to drink, to engage in eye contact'

Item 16: Cruising/supported stand

What it means to you?
'I used to walk holding onto something'

'Totally change quality of life, reduces dramatically the need for equipment'

'Independence, basic freedom, mobility, dignity, avoid embarrassment'

'Independence interacting with others, making transfers easier'

Activities?
'Mobility around the house & classroom'

'Go to toilet, classrooms, friends houses'

'Ability to transfer when out and about, access to kitchen/ food, bathing activities, open doors'

'Movement at home around sofa, living room etc.'

Item 34: Climbs Box Step

What it means to you?
'Ease of living'

'Very important as this is the first step to be able to climb the stairs'

'Allows access into numerous places'

'Important'

'Useful for moving around'

Activities?
'Able to move into bath tub'

'Step to reach basin to brush teeth'

'Get into a house, shop, pavement'

'Exercise, gym'

Discussion:

- We report excellent inter-rater reliability properties of the RHS with ICC 0.997.
- The limits of agreement for intra-rater reliability as assessed with Bland Altman currently sit outside of ± 2. However as 97.1% of actual values sit within the limits, increasing sample size may address this discrepancy.
- Content validity of the RHS from a patient/parent perspective has been established for all 36 items of the RHS.

Ongoing/Future Work:

- Ethical approval has been granted to extend the reliability study in order to improve the precision of estimate for intra-rater reliability, with plans to repeat the reliability study in the Italian SMA and PNCR USA networks.
- Annual online reliability training/testing will be conducted for SMA REACH UK Network using the protocol employed in this study.
- Establish the minimally clinically important difference for the RHS in consultation with patients/parents and expert Clinicians and Physiotherapists.