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

D. Ramsey¹, M. Scoot¹, A. Mayhew², G. Ramdharry³, A. Wallace³, F. Muntoni¹
¹Dubowitz Neuromuscular Centre, UCL & Great Ormond Street Hospital, London, UK, ²Institute of Human Genetics, Newcastle University, UK, ³Faculty of Health Social Care and Education, Kingston University and St George’s University of London, ⁴UCL Institute of Child Health, London

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. UCL 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:

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 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.

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) -2.42 and upper LOA +2.29, table and figure 2.
• SMA 3 intra rater difference: mean -0.18, lower LOA -2.79 and upper LOA +4.24, 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.

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, the please see examples data of 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.

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.

Figure 1: Example of qualitative content validity data from a patient/parent perspective for items on the RHS

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