



The Development of a Cognitive Assessment for Individuals with Rett Syndrome

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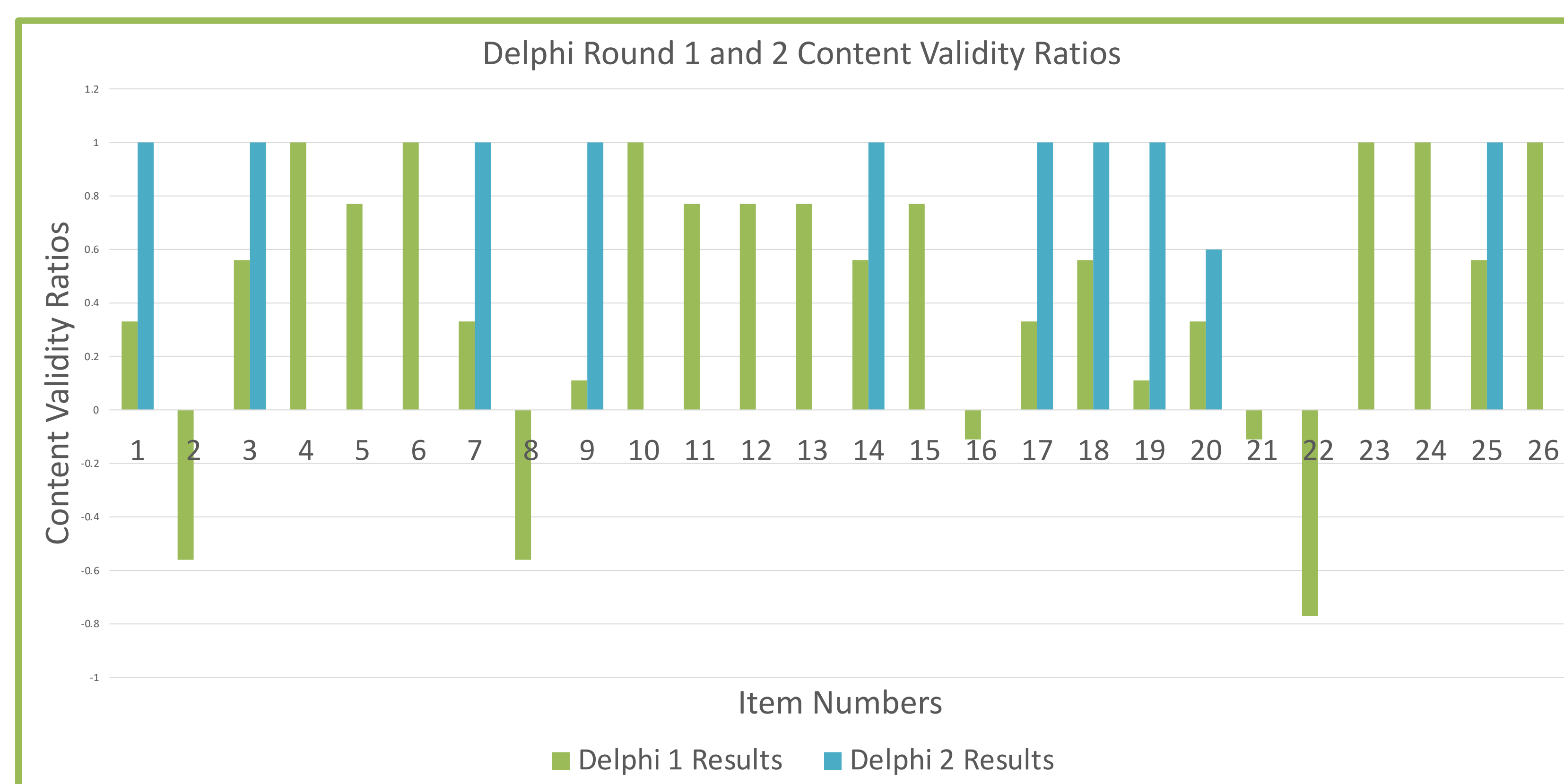
Introduction

Many cognitive assessment tools are available to assess individuals; however, families of children with Rett Syndrome express a need for more appropriate tools for the population. Girls diagnosed with Rett Syndrome display characteristics that make common assessment tools inadequate, such as repetitive hand motions and a lack of verbal communication (Consentino et al., 2019). Current tools utilize components inappropriate for this population, leading to possible underestimation of cognitive abilities (Loffler & Gordon, 2018). Researchers have expressed a need for a cognitive assessment designed for the population (Byiers & Symons, 2013). The Occupational Therapy Practice Framework emphasizes the importance of an accurate occupational profile to provide the best interventions and outcomes (American Occupational Therapy Association [AOTA], 2020). This led to the research question, "For girls with Rett Syndrome, would the design of a tailored cognitive assessment be more effective in evaluating cognitive function?" This study aims to design and validate a cognitive assessment tailored to the skills of girls with Rett Syndrome.

Methods

This study was a mixed-method design. Participants were identified using both convenience sampling and snowball sampling. Inclusion criteria consisted of proficiency in English and self-identifying as an expert. An expert was defined as having personal or professional experience with an individual with Rett Syndrome over the course of at least one year. Research began by interviewing caregivers of children with Rett Syndrome. Questions from pre-existing caregiver report cognitive assessments were then gathered through a literature review and adapted to suit the target population. The Delphi Method was used for initial validation. Experts ranked each question on a three-point Likert scale, and all scores were used to calculate a Content Validity Ratio (CVR) for each question. The Content Validity method, as described by Veneziano and Hooper (1997), was used to assess content validity. Two rounds of Delphi were performed to reach statistical significance.

Results



Changes Made to Measure After Review 1

Item #	Item	Change	Justification for Change
1	They are able to make decisions or solve problems on their own.	Added "if given an appropriate communication avenue"	Made the language more inclusive
3	They are able to learn how to use a tool, toy, or gadget.	Added "activate a cause and effect device" to exemplar	Expanded the definition of tool, toy, or gadget
7	They remember where they have placed objects.	Changed to "where a familiar object was last left."	Removed the word placed to reflect potentially limited upper extremity use
9	They are aware of who they are and the environment they are in.	Added "as expressed by correct statement or response using any communication avenue" to exemplar	Cleared up confusion regarding how this could be demonstrated
17	They know at least three of their own body parts.	Changed to "They can identify at least three body parts using a communication device."	Changed to reflect feedback that this was a more realistic task
18	They are able to indicate the location of at least three objects shown in a picture when you ask.	Added "or indicate it using a communication device" to exemplar	Made the language more inclusive
20	They are able to let someone know when they need to use the bathroom, or if their diaper or pants are wet or soiled.	Added "ambulating towards the bathroom" to exemplar	Expanded on the definition of "letting someone know"

Discussion

Implications for OT Practice	Future Research	Study Limitations
<ul style="list-style-type: none"> use of the assessment to look for discrepancies in testing Better advocacy and more accurate intervention planning 	<ul style="list-style-type: none"> Cognitive interviews to further validate Pilot testing Cross validation with other assessments for the population 	<ul style="list-style-type: none"> Multiple rounds of review led to participant dropout Confusion regarding instructions

Conclusion

Next steps include cognitive interviews with caregivers regarding assessment content, regression analysis, and cross-validation with other accepted assessments or assessments currently being developed for this population. One set of caregivers interviewed for this study said the lack of reliable assessment options for their daughter led to a long and challenging battle with the school system to allow their child to spend a portion of the day with typical peers. Suppose assessment is an essential element of the occupational therapy process, and the cognitive abilities of individuals with Rett Syndrome cannot be assessed accurately with current tools. In that case, occupational therapy practitioners must develop an assessment tailored to them. The development of this assessment will allow for better treatment, care coordination, and advocacy for this population.

References

Loffler, G. & Gordon, G.E. (2018). Cognitive function in Rett Syndrome: Profoundly impaired or near normal? *European Journal of Paediatric Neurology*, 22(1), 2-3. <https://doi.org/10.1016/j.ejpn.2017.12.006>

Cosentino, L., Vigli, D., Franchi, F., Laviola, G., & De Filippis, B. (2019). Rett syndrome before regression: A time window of overlooked opportunities for diagnosis and intervention. *Neuroscience and Biobehavioral Reviews*, 107, 115-135. <https://doi.org/10.1016/j.neubiorev.2019.05.013>

Byiers, B., & Symons, F. (2013). The need for unbiased cognitive assessment in Rett Syndrome: Is eye tracking the answer? *Developmental Medicine & Child Neurology*, 55(4), 301-302. <https://doi.org/10.1111/dmcn.12088>

Venziano, L. & Hooper, J. (1997) A method for quantifying content validity of health-related questionnaires. *American Journal of Health Behavior*, 21(1), 67-70.

American Occupational Therapy Association. (2020). Occupational therapy practice framework: Domain and process (4th ed.). *American Journal of Occupational Therapy*, 74(Suppl. 2), 7412410010. <https://doi.org/10.5014/ajot.2020.74S2001>

Acknowledgement & Contact Information

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