AAC News for Newbies
Title: A Perspective on Rett Syndrome and Communication Success with this Population

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Introduction:

How do you measure communication outcomes when working with children who have Rett syndrome? Many factors contribute when trying to measure positive outcomes for specific populations that use AAC devices. Speech-language pathologists and other professionals must collaborate to achieve the best outcomes for individuals with Rett syndrome using the AAC device. Specifically, when working with individuals who have RRT it is important to work together with family members to generate meaning communication goals. Further, as RTT progresses it expectedly follows four stages of regression, so it is critical that the individual have careful prioritization of communication goals for all environments.

Understanding the Syndrome:

Rett syndrome is rare postnatal neurological and developmental disorder, which occurs approximately one in 10,000 to 23,000 female births. Though, Rett syndrome occurs primarily in females, it can occur in males, and most times the males are severely affected. Further, this disorder is the second-most common cause of severe intellectual disability in females and is characterized with impairments in cognitive ability, communication and motor function. According to Johnson, Mullaney, & Blue (2003), this disorder is regarded as an "age-related regressive disorder of neuronal development," and not considered to be degenerative, which it was once labeled. This disorder is commonly diagnosed with genetic testing as most cases of RTT are caused when the MECP2 gene mutate; the gene responsible for producing sufficient amounts of protein. Though, the identification of this gene mutation alone cannot be the basis for diagnosis of RTT, there is other criteria to consider. The classic criteria of this disorder presents with a period of typical development early in life, followed by a period of decline in development, and then a period of regression. Further, RTT has been reported to be misdiagnosed as autism and cerebral palsy, though there are other distinctions such as stereotypical hand movements, acquired microcephaly, and gait disturbances.

Remember to Explore the Evidence:

In a systematic review by Sigafoos et al. (2008), nine communication intervention studies involving people with Rett syndrome were assessed. Before assessing these nine studies Sigafoos et al. (2008) explained the progression of RTT according to the following four stages. Stage 1 also known as, "early onset deceleration," occurs between 6 and 18 months and characterized by reduced play, social and communication skills. Stage 2 also referred to as, "rapid destructive stage," can occur between 1 to 3 years and is characterized by a near total loss of speech and purposeful hand movement. Stage 3, also referred to as the "pseudostationary stage," occurs between 2 to 10 years and is characterized by consistent repetitive hand movements, seizures and scoliosis. The final stage or, "motor deterioration stage," occurs by approximately age 10, and is characterized by continual deterioration of mobility and decline of responsiveness to the environment. Further, as the onset of each stage is variable for each individual, the studies

assessed in this systematic review of communication interventions included individuals with Rett syndrome from ages 2:7 to 17:0.

Sigafoos et al. (2008), conducted a systematic review that measured the effectiveness of communication interventions used with 31 participants in nine studies diagnosed with Rett syndrome. Across the 9 studies included in this systematic review 10 participants were identified to be in the third stage, 4 participants in the final stage, and the remaining 17 participants did not have their stage indicated. Researchers measured improvements in interactions skills, eye gaze to request or initiate, imitation of sounds, and the use of graphic symbols or speech generated device. All participants in the nine studies included had RTT, however each participant had variable outcomes depending on their gross-motor ability and age.

Results across the nine studies indicated that various factors influence improved outcomes of interventions with AAC. In the one study it was reported that two of the three participants made communication gains using dynamic symbols on a computer screen to request food or drink. Though, one of the nine studies reported a small gain it was not statistically significant and the remaining studies reviewed reported that there was no significant difference in baseline after the intervention interaction. Further, it was discussed that positive outcomes with the communication intervention is seemly dependent upon the stage at which the individual is in and severity of the gene mutation.

In a recent cross-sectional, correlational study conducted by Urbanowicz et al. (2015), the retention and use of gestures and eye gaze to request attention, objects and information in females with Rett syndrome was assessed. The researchers acquired 136 participants through the Australian Rett Syndrome Database and assessed their use of eye gaze and gesture with the use of video-based filming protocol and parent-report questionnaire called the Functional Ability Checklist or FAC.

Results of the Urbanowicz et al. (2015) study indicated that gross motor ability and gene mutation type influenced communication with eye gaze and gesture. Further, the study results concluded that girls and women aged 8 to 19 years used eye gaze more frequently than girls 19 years and older. Additionally, it was concluded that girls with more motor ability used eye gaze more often than girls with decreased motor ability. When eye gaze was reported, it was used to gain attention of a caregiver, request help or an object. These communication interactions established with eye gaze relate to the functionality and linguistic complexity of intentions by the individual using eye-gaze with AAC.

Concluding, before an individual with Rett syndrome utilizes AAC with eye gaze, it is important to consider not only their cognitive abilities but also their motor abilities. If their motor abilities are more intact, a communication goal should aim to facilitate joint attention, as this targets more advanced eye gaze skills, which can be utilized for an AAC device with eye-gaze. Urbanowicz et al. (2015), underscores joint attention as important skill for woman and girls with RTT, as the study indicates eye-gaze is more often utilized as the primary mode of communication than gesture.

Works Cited:

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