Assessing cognitive functioning in females with Rett syndrome by eye-tracking methodology

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ABSTRACT

Background: While many individuals with severe developmental impairments learn to communicate with augmentative and alternative communication (AAC) devices, a significant number of individuals show major difficulties in the effective use of AAC. Recent technological innovations, i.e., eye-tracking technology (ETT), aim to improve the transparency of communication and may also enable a more valid cognitive assessment.

Objectives: To investigate whether ETT in forced-choice tasks can enable children with very severe motor and speech impairments to respond consistently, allowing a more reliable evaluation of their language comprehension.

Methods: Participants were 17 girls with Rett syndrome (M = 6.06 years). Their ability to respond by eye gaze was first practiced with computer games using ETT. Afterwards, their receptive vocabulary was assessed using the Peabody Picture Vocabulary Test-4 (PPVT-4). Target words were orally presented and participants responded by focusing their eyes on the preferred picture.

Results: Remarkable differences between the participants in receptive vocabulary were demonstrated using ETT. The verbal comprehension abilities of 32% of the participants ranged from low-average to mild cognitive impairment, and the other 68% of the participants showed moderate to severe impairment. Young age at the time of assessment was positively correlated with higher receptive vocabulary.

Conclusions: The use of ETT seems to make the communicational signals of children with severe motor and communication impairments more easily understood. Early practice of ETT may improve the quality of communication and enable more reliable conclusions in learning and assessment sessions.

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1. Introduction

Rett syndrome (RS) is a genetic neurodevelopmental disorder associated with dominant MECP2 gene mutations on the X chromosome.\(^1\) The syndrome is characterized by apparently normal early growth and development followed by slowing of development, distinctive hand movements, acquired microcephaly, loss of purposeful hand use and loss of acquired spoken language.\(^4\) The first signs of deterioration are commonly detected between 6 and 18 months of age.\(^5\) The occurrence of RS is estimated to vary between 1/10,000 and 1/15,000 of newborn females.\(^6\) Females with RS are commonly considered severely intellectually impaired, an impression supported by the characteristic deceleration of head growth and low brain weight.\(^7,8\) Seizures and abundant epileptiform activity are common features of the disorder,\(^9\) but the relationship between epilepsy and cognitive level has not been addressed in the literature concerning patients with RS, although more common forms of epilepsy have frequently been associated with cognitive decline.\(^10\)

Assessment of the cognitive skills of females with RS, as well as other individuals with severe motor and communication limitations, is extremely challenging.\(^11\) Neuropsychological and cognitive assessments are generally developed for and standardized with typically developing children who do not have physical impairments.\(^12\) When standard assessment procedures requiring manual motor functioning for responding have been used to evaluate their cognitive functioning, females with RS generally achieve age-equivalent performances close to young infants.\(^13\) Some studies have aimed to recognize any communicative acts (vocalizations, stereotyped hand movements, body movements, facial expressions, or eye gaze) that would represent a consistent and unequivocal response used by participants with RS. However, even though individual variation in communicative gestures has been taken into account, low inter-rater agreement has been reported in assessing the communicative intent of commonly performed behavioral gestures.\(^16\) Therefore, it has been questioned whether these gestures in fact have a communicative intent among females with RS and can be useful as reliable responses in cognitive assessment.\(^16\)

Together with improved feasibility of technological innovations, the use of eye gaze as a means of access to communication has become increasingly appealing. One of the supportive criteria for clinical RS diagnosis is intense eye gaze, which is typical in females with RS both classical and preserved speech variant.\(^17\) Females with RS have displayed preferential eye fixation on social stimuli,\(^18\) and especially on people’s eyes.\(^21\) Their pattern of preferential looking has commonly been considered an indication of interest, as well as a way to make requests and communication.\(^22,23\) However, few studies have tried to investigate the utility of preferential eye gaze in communicating responses in cognitive tasks. In studies using eye gaze as an indication of a response, cognitive performance among females with RS has generally been very low.\(^16,23\) Visual processing and visual memory among females with RS was demonstrated to be significantly lower than controls, though significant individual variation was found.\(^23\) One study demonstrated that most participants with RS were able to follow simple verbal instructions and to recognize, match, and categorize pictures.\(^24\)

As cognitive functioning among females with RS has rarely been assessed by validated cognitive tools that do not require motor functioning, the aim of this study was to assess their receptive language abilities using eye-tracking technology (ETT). Receptive language was assessed by Peabody Picture Vocabulary Test-4th edition (PPVT-4).\(^26\) Our hypothesis was that most of the girls would respond correctly to the easiest level of stimuli. Additionally, we hypothesized that girls with a lower severity score of Rett syndrome symptoms as measured by Percy scales\(^27\) would achieve higher scores on the PPVT-4 test as an ETT-based task, compared to girls with higher RS severity scores. We also assumed that a lower level of epileptiform activity on sleep EEG, as well as minimal or no use of antiepileptic drug (AED) therapy, would be correlated with higher scores on the PPVT task.

2. Method

2.1. Participants

Twenty-two girls diagnosed with Rett syndrome were enrolled in the study (see Table 1). Three participants had to discontinue the assessment due to medical reasons (orthopedic operations), and two participants were excluded due to lack of cooperation and lack of eye gaze on the screen, even in the game activities preceding the cognitive assessment. Seventeen participants completed the assessment. Age range varied between 3 years and 4 months to 12 years and 2 months; the mean age was 6 years and 6 months. Collected data regarding the patients included, in addition to their age, the genetic mutation type, level of epileptiform activity during 45 min sleep recording (0 – no epileptiform activity, 1 – low epileptiform activity (0–25%), 2 – moderate epileptiform activity (25–50%), 3 – high epileptiform activity (more than 50%), type of antiepileptic drug therapy (AED), severity level of Rett syndrome as measured by Percy scales, and type of AAC.

Percy scales\(^27\) is a comprehensive, commonly used clinical scale assessing the course and the current functioning of girls with Rett syndrome in most of the relevant areas affected (i.e., growth, motor and verbal skills, stereotypical movements). The type of AAC refers to the most common method of communication used in the participant’s daily life. While AAC may be used with a communication book or a computer, the method of choice for most participants (12 of the 17) was eye gaze. The remaining five participants chose either by gestures and sounds or by pointing to the chosen stimuli. Only three participants had prior experience in using the Tobii eye gaze system. All of them had begun using it 3–4 months prior to the study and were in the practice phase.

Vision examinations were conducted for all the participants as part of a routine medical follow-up. Participants who regularly use eye glasses did so during this study.

All procedures were approved by the Hospital Institutional Review Board and were performed in compliance with ethical
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