



Assistive Technology for Promoting Adaptive Behaviors by Children with Rett Syndrome

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Rett syndrome (RS) is a progressive neuro-developmental disorder due to mutations of methyl CpG binding protein 2 gene (MECP2), located on Xq28 chromosome, primarily occurring in females and first described by Andreas Rett [1]. It is characterized by four main stages, namely: (a) stagnation, (b) regression, (c) stationary, and (d) deterioration. During the first stage, occurring with an early onset and beginning between 6 and 18 months of age, the child development is overall arrested. Notably, the infant is commonly described with a loss of eye contact, interest and attention, easily irritated, stop of head growth. The second stage, oncoming between 1 and 4 years old presents, hand-related stereotypic movements and a loss of speech. Furthermore, breathing abnormalities such as apnea and/or iper-ventilation may occur as well as seizures, failing of locomotion and decreased social interactions capacities. During the third stage, starting between 2 and 10 years old, the child may exhibit apraxia and ataxia disorders, autistic features such as withdrawal, crying and irritability. Finally, the fourth stage can last four years and the child usually has dystonic movements, spasticity, scoliosis [2]. Therefore, a child with RS is totally dependent of caregivers and needs both educational and rehabilitative arrangements. To overcome this issue, ensuring RS children with self-determination and independence towards the environment, one may resort on assistive technology-based programs (AT) [3]. Within this framework, different strategies may be adopted [4]. For example, one may envisage the use of micro switches (i.e. electronic devices enabling individuals with multiple disabilities with the independent access to positive stimulation) [5].

Stasolla and Caffò [6] exposed two girls with RS to a micro switch-based program with the dual objective of: (a) providing the independent access to positive stimulation, and (b) promoting their locomotion fluency. Stasolla et al. [7] compared two different alternative and augmentative communication opportunities such as PECS and VOCA, involving three girls with RS and assessing their choice preferences. Stasolla et al. [8] pointed out the choice making process among different categories of stimuli, exposing three girls with RS to an AT-based intervention. All the aforementioned empirical evidences outlined the effects of such programs on: (a) the decreased stereotyped behaviors (e.g. body rocking, hand washing), and (b) the increased intervals with indices of happiness as outcome measure of the quality of life. Finally, external raters involved in a social validation assessment favorably endorsed the use of the AT interventions.

In light of the above, one may argue on the effectiveness, the suitability and the reliability of AT-based programs for individuals with RS, since all the participants involved learned the positive use of such technologies for enhancing adaptive responding, reducing stereotyped behaviors and improving their quality of life [9]. Accordingly, one may envisage, at least, four main domains to be fostered as consequence of the implementation of an AT-based strategy, namely: (a) independent access to preferred stimuli, (b) communication of personal needs and/or recreational/occupational/academic purposes, (c) locomotion, and (d) choice making. All these solutions should always be rigorously individualized as to ensure individuals with RS with the most preferable and suitable opportunity, considering their clinical and general conditions on one hand and the families/caregivers/context(s) expectations on the other [10].

Future research perspectives within this area should consequently deal with the following topics: (a) further extension of the previous findings to other participants with RS, (b) further integration of AT-based solutions in daily contexts considering both economical and human resources, (c) enlarging social validation assessment to different groups of raters (e.g. caregivers, parents, students and teachers), (d) assessing constantly the quality of life of participants involved through indices of happiness and/or preference checks as outcome measures of their quality of life [10].

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