Chapter 9
Assistive Technology–Based Programs and Cognitive–Behavioral Interventions for Helping Adaptive Responding of Children and Adolescents With Rett Syndrome:
A Selective Overview

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ABSTRACT

This chapter provides the reader with a concise overview of the newest empirical evidences available on the use of assistive technology and cognitive behavioral interventions to promote adaptive skills and decrease challenging behaviors of children and adolescents with Rett syndrome and severe to profound developmental disabilities. Four main categories of studies were identified, namely (1) communication skills, (2) adaptive skills, (3) challenging behavior, and (4) on-task behavior. Twenty-six contributions were reviewed, and 936 participants were involved. Results were largely positive, although rare failures occurred. Educational, clinical, rehabilitative, and psychological implications of the findings were critically discussed. Some useful suggestions for future research and practice were emphasized.

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INTRODUCTION

Rett syndrome (RTT) is a rare genetic disease, caused by X-linked mutations of the methyl CpG binding protein 2 (MECP2) gene, located on the Xq28 chromosome, primarily occurring in females, and first described by Rett (1966). It includes severe to profound developmental disabilities, and is characterized by four basic stages, namely (a) stagnation, (b) regression, (c), stationary, and (d) motor deterioration. Subsequently to an apparent regular birth, the stagnation usually encompasses a general arrest of child's development between 5 and 6 months. The regression commonly has an onset between 1 and 4 years old. A general loss of adaptive functions is described. Thus, either a progressive speech or a social interactions' impairment are frequently observed. Nevertheless, eye contact for communicative purposes is preserved. During the stationary stage, hand-related stereotypic behaviors (e.g., clapping and washing) emerge. Additionally, breath abnormalities (i.e., hyperventilation) and behavioral problems (i.e., nocturne crying and laughing attacks) may be emphasized. Seizures, ataxia, apraxia, motor scoliosis are broadly embedded, between 2 and 8 years. The motor deterioration is reported with motor decline, reduced physical activity, decreased stereotyped behaviors, and withdrawal, after 10 years old. Next to the aforementioned clinical conditions, failing locomotion is one of the most common negative functional consequences of RTT (Neul et al., 2010; Rett, 1966; Stasolla et al., 2018). To overcome the isolation and passivity of individuals with RTT, one may resort on the use of assistive technology-based programs (AT) and/or cognitive-behavioral interventions for rehabilitative goals (Lancioni & Singh, 2014).

For example, Hornof Whitman, Sutherland, Gerendasy, and McGrenere (2017) exposed a 20-month-old girl who was diagnosed with RTT to a *Motivating Music* system. The girl presented severe to profound developmental delays, and no specific interactions with her environment. An easy-to-use and engaging music-playing system was designed for motivating her constructive engagement and positive occupation. During the intervention phases, the in-home girl's therapists (i.e., speech, physical, and occupational caregivers) noticed that the girl was highly motivated to use the technological system and incorporated it into their in-home working sessions. The girl consolidated the use of the system along three years and remained strongly engaged when using it.

Lotan (2007) reviewed the literature available on the use of AT-based treatments for reversing both intellectual and physical impairments, and fostering a comprehensive network of support, which included different types of exercise and various modalities of intervention. Supplementary adaptive techniques, environmental modifications, and technological aids were considered. This specific strategy was planned to be performed by non-professionals with the supervision of a qualified therapist. Such approach was conceived as a further support for pursuing the therapeutic goals of the child with RTT, once direct intervention was not supplied.

Recently, Sigafoos et al. (2019) reviewed 38 studies and identified promising approaches that sought to improve adaptive functioning in cases of regression. Thus, 132 participants with RTT were involved in cases where developmental regression was either already occurred or was likely to occur. Interventions targeted a range of variables (e.g., challenging behaviors, communication, motor, and social skills). Multi-component interventions based on learning principles were the norm, suggesting the need for clinical expertise in the application of behavior analytic principles. Nevertheless, only 12 retained studies with 44 participants enrolled were rated as corroborating positive conclusive evidence of a beneficial outcome. Future and more specific research on the mechanisms underlying developmental regression was warranted.