Chapter 7
Assistive Technology to Promote Adaptive Skills in Children and Adolescents With Rett Syndrome: A Selective Review

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ABSTRACT

Rett syndrome is classified within the rare genetic syndromes, characterised by intellectual delays, extensive motor impairments, lack of speech and communication difficulties, sensorial deficits, and problems in adaptive responding. That clinical conditions may be deleterious on their social image, status, and quality of life. A practice for addressing this problem is technology-based interventions. The use of assistive technologies, in particular microswitches, with children with RTT has been shown to effectively change the impact on their quality of life, facilitating access to recreational activities and improving their performance. Through the use of technology-aided programs, a child with RTT and multiple disabilities will be ensured with an independent access to positive stimulation. In this chapter, a selective literature review was carried out considering Rett Syndrome, assistive technologies, quality of life, and rare genetic syndromes. Empirical data demonstrated the effectiveness and suitability of interventions with AT, allowing participants to increase their level of independence.

INTRODUCTION

Rett syndrome (RTT) is a progressive neurodevelopmental disorder, X linked, due to mutations in the methyl-cytosine phosphate guanine binding protein 2 (MECP2) gene, located on Xq28, largely affecting females, first described by Andreas Rett (1966). It is associated with severe to profound intellectual, communicative and motor impairments, beginning between 6 and 18 months old, preceded by a normal

DOI: 10.4018/978-1-7998-7430-0.ch007

birth and an initial regular development (Didden et al., 2010). RTT traditionally follows four stages, namely: (a) stagnation, (b) regression, (c) stationary, and (d) motor deterioration (Stasolla & Caffò, 2013).

Subsequently to an apparent regular birth, the stagnation commonly includes a global arrest of child's development between 5 and 6 months. The regression usually has an onset between 1 and 4 years old. A general loss of adaptive functions occurs. Thus, either a progressive speech or a social interactions' impairments are frequently observed. Nevertheless, eye contact for communicative goals is preserved. During the stationary phase stereotypical hand-related behaviors emerge (e.g., clapping and washing). In addition, respiratory abnormalities (i. e., hyperventilation) and behavioral problems (e. g., night crying and laughing attacks) may be highlighted. Seizures, ataxia, apraxia, motor scoliosis are broadly embedded, between 2 and 8 years. The motor deterioration is described by motor decline, reduced physical activity, decreased stereotyped behaviors, and withdrawal, after 10 years old. Beside 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., 2015)

Individuals with RTT usually present a vast array of orthopedic and neurological difficulties. Typical challenges, which may need to be addressed, when treating this population are functional limitations, low cardiovascular capacity, hypotonia, ataxia, apraxia, loss of transitional movements, spasticity, scoliosis and/or kyphosis, loss of ambulation, loss of hand function, foot deformities, and spatial disorientation (Lotan, 2006). Moreover, individuals with RTT are frequently describes as isolated, with minimal opportunities to interact positively with their environment and surrounding objects. Additionally, reduced sensory stimulation, deprivation, and negative consequences on their quality of life may be evidenced (Felce & Perry, 1995). Finally, the latter conditions (i.e., deprivation, isolation, passivity, and withdrawal) may seriously hamper their social image and status (Stasolla et al., 2014).

Interventions for people with RTT may be aimed at compensating and reducing physical impairment and include therapeutic interventions, adaptive techniques, environmental changes and assistive technology-based interventions (AT) (Lancioni, Sigafoos, O'Reilly, & Singh, 2012). That is, by using technological aids such as sensors and personal computers, one may envisage to improve self-determination, with the opportunity of enhancing occupation, constructive engagement, and choice options, almost lacking among children with RTT. The Australian RTT study (Leonard, 2002) has gathered information regarding the use of assistive technology among individuals with RTT. Their findings suggest that the majority of individuals with RTT are using such devices. As detailed in this review, the most commonly used AT for people with RTT are: (a) mobility aids, (b) AT for physiological needs, (c) transport aids, (d) foot supports, (e) communication aids, (f) arm supports, and (g) lifting aids. (see Figure 1).

For instance, 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.

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