

Rehabilitative Solutions for Promoting Independence and Self-Determination of Children with Cornelia de Lange Syndrome

Editorial

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Cornelia de Lange syndrome (CdL) is a congenital disorder characterized by multiple disabilities. It includes growth delays, behavioral abnormalities, both hands and feet problems, intellectual difficulties, autistic features, stereotypic movements, language deficits, self-injuries [1, 2]. Although there is a substantial variance in its phenotype, the majority of CdL individuals are socially, communicatively and cognitively impaired, ranging between the moderate and the profound level of developmental disabilities [3]. Microcephaly, highly arched eyebrows, long and thick eyelashes, short neck, hirsute ears with thickened helices, small widely-spaced teeth, arched palate with clefts, downturned mouth corners are physical traits commonly described [4]. The diagnosis is usually based upon clinical grounds. Specifically, positive mutation on CdL gene testing or facial findings and meet-criteria such as development and behavior basic points (e.g., learning difficulties, attention deficits hyperactivity disorders, anxiety, withdrawal, impulsive behavior, roaming, obsessive-compulsive features) should be considered [5, 7]. Accordingly, a primary rehabilitative goal of CdL children is the independence and self-determination towards their environment because they constantly rely on parents and/or caregivers' assistance due to their pathology [8-10]. One way to pursue the latter objective, is the use of cognitive-behavioral interventions [11, 12]. Eventually, assistive technology-based program should be implemented [13, 14].

Cognitive-behavioral rehabilitative strategies are frequently based upon learning principles (i.e., causal association between behavioral responding and environmental consequences), for enabling children with multiple and/or severe to profound developmental disabilities with the awareness of their own behavior and enhancing it to be functional for getting independent access to positive stimulation [15, 16]. Within this framework, AT refers to any piece, device, equipment or tool fostering the autonomy of participants involved in the rehabilitative packages. Furthermore, augmentative alternative communication (AAC) aided-options may be useful and helpful for improving communication skills

[17]. Positive outcomes may have beneficial effects on children's quality of life [18]. Moreover, both parents and caregivers' burden might be significantly reduced [20].

Depending upon participants' levels of functioning and their general and/or clinical conditions, both economical and human resources, and the rehabilitative aims, one may envisage different typologies of solutions. For instance, early intensive behavioral interventions may be successful for both reducing aggression, and self-injuries, and re-direct participants' behavior profitably for the acquisition of new adaptive skills [19]. Eventually, a functional assessment of the challenging behavior may be meaningful [20]. Else, one may implement a cluster technology for reaching the dual goal of increasing an adaptive responding and decreasing a challenging behavior [21]. Otherwise, a microswitch-based program with contingent stimulation may be implemented for facilitating locomotion and/or its fluency [22]. AAC-based programs such as PECS and VOCA or computer-assisted options may be adopted for promoting the independent communication of both personal needs and academic performances [23-25]. Additionally, occupation purposes through AT may be achieved [26, 27]. Finally, social validation assessments by external sensitive raters and indices of happiness and/or positive participation as outcome measures of the quality of life may be carried out [28-30].

In light of the above, new research perspectives within this area should deal with the following topics: (a) extend the use of both cognitive-behavioral interventions and AT-based programs to new individuals with CdL syndrome, because the current literature on this specific framework is lacking, (b) find out new updated and individualized technological solutions for each CdL participant, (c) conduct new social validation assessments involving parents, students, physicians, teachers and caregivers as expert external raters, and (d) consider indices of happiness and/or positive participation as outcome measures of participants' quality of life as to corroborate the intervention's clinical and practical validity.

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