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Technological supports to promote choice opportunities by two children with fragile X syndrome and severe to profound developmental disabilities

Fabrizio Stasolla [a,*], Rita Damiani [b], Viviana Perilli [c], Antonia Di Leone [b], Vincenza Albano [b], Anna Stella [b], Concetta Damato [b]

[a] Lega del Filo d'Oro Research Center, Molfetta, Italy
[b] Department of Educational Sciences, Psychology, Communication, University of Bari, Italy
[c] Lega del Filo d'Oro, Research Center, Lesmo, Italy

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ABSTRACT

This study was aimed at assessing whether technological supports (i.e. optic sensors such as photocells) were successful enabling two boys with fragile X syndrome and severe to profound developmental disabilities to perform occupation and choice opportunities. A second goal of the study was to reduce stereotyped behaviours (i.e. hand mouthing and eye poking) exhibited by the participants. Finally, the third purpose of the study was to verify the rehabilitative effects of the intervention program on the indices of happiness of the participants. The study has been conducted according to a non-concurrent multiple baseline design across participants followed by intervention and cross over phases, where the associations between behavioural responses and environmental consequences were systematically inverted. Moreover, a maintenance phase was assessed. The results demonstrated that the technology is useful to facilitate employment and opportunities of choice, showing a growth of the indices of happiness and a decrease of stereotyped behaviours, from both participants involved. Clinical, practical and psychological implications of the findings are discussed.

1. Introduction

Fragile X syndrome (FXS) is a single gene disorder and represents one of the most common cause of developmental disabilities, usually associated with autism spectrum disorders. It is the result of an excessive length concerning a repetitive sequence of trinucleotides (CGG) in a specific gene (FMR1), located on the long arm of X chromosome (Glaser et al., 2003; Murphy & Abbeduto, 2007; Roberts, Weisenfeld, Hatton, Heath, & Kufmann, 2007). The FMR1 gene is linked to the production of FMR1 protein, that seems to be crucial for a regular brain functioning. FXS, primarily occurring in males, is generally described with physical, behavioural and cognitive characteristics, due to its full mutation. That is, physical features include a long narrow face, prominent ears, hyper-extensible joints, and macroorchidism. Furthermore, the syndrome causes moderate to severe intellectual disabilities, inattention, impulsivity, anxiety, gaze avoidance, self-injury,

* Corresponding author. Tel.: +39 3496635397; fax: +39 069384564.
E-mail addresses: f.stasolla@psico.uniba.it, f.stasolla@libero.it (F. Stasolla).

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and stereotypies (Arron, Oliver, Moss, Berg, & Burbidge, 2011; McCary & Roberts, 2013; Roberts et al., 2011). Moreover, they pose serious problems regarding learning disabilities. Thus, individuals with FXS may be considered within the range of multiple disabilities (Lancioni et al., 2010; Lancioni, O'Reilly, et al., 2007, Lancioni, Singh, et al., 2007).

Children with multiple disabilities may present a certain number of challenge problems to rehabilitation staff (Belva & Matson, 2013). For example, they can exhibit lack of positive interaction with surrounding objects, stereotyped behaviours, and passivity. A critical rehabilitative goal is to point out an effective strategy that help them to acquire constructive engagement with environmental stimulation, such as object manipulation leading to contingent preferred stimuli (i.e. environmental events enabling and motivating the performance by increasing and consolidating behavioural responses over the time) (Chantry & Dunford, 2010). In order to emphasize such perspective, one may envisage the use of assistive technology, aimed at monitoring the aforementioned behaviours and consequently providing the contingent stimulation, independent of caregivers, based on learning principles (Lancioni & Singh, 2014; Lancioni, Sigafoos, O'Reilly, & Singh, 2012).

Research efforts to pursue the aforementioned objectives are basically sparse, although encouraging. For example, Lancioni, O'Reilly, and Campodonico (2001) carried out a study with two men who presented severe visual impairments and profound intellectual disabilities, and limited interaction with environmental objects. Both participants were successfully engaged in a positive object manipulation (insert daily object in a container). The technology ensured participants with brief periods of positive stimulation contingently with object responses. Recently, Lancioni et al. (2014) conducted a study with two participants (an adolescent and an adult), involving them in constructive object-manipulation behaviours. The technology monitored their responses allowing them the independent access to preferred stimuli during intervention phases.

The present study was aimed at replicate and extend the use of such technology, providing a new setup for two boys with FXS and severe to profound intellectual disabilities, pursuing the following objectives: (a) promote occupation and choice opportunities for both participants, (b) decrease stereotyped behaviours exhibited by the participants involved, (c) monitor the intervention effects on the indices of happiness, and (d) assess the learning consolidation over the time with a maintenance phase for each participant involved.

2. Method

2.1. Participants

The participants (Bernard and Vincent) were 8.7 and 9.7 years old at the beginning of the study (mean age 9.2 years) respectively. They were diagnosed with FXS full mutation based on DNA results. Moreover, they were both mosaic (i.e. they presented the full mutation and a pre-mutation), consistent with its prevalence among male population with FXS (Nolin, Glicksman, Houck, Brown, & Dobkin, 1994). Although no formal scores of IQ were available since no test was feasible, due to their general conditions, both were estimated within the range of severe to profound intellectual disabilities from clinical observations. They were described with lack of speech and communication abilities, sphincter control incapacities, failing locomotion independently, quite passive and isolated across the days. Furthermore, they exhibited stereotypic behaviours such as hand mouthing (Bernard) and eye poking (Vincent). They attended regular class with special training. Bernard received speech sessions and Vincent stimulation sessions twice a week in a rehabilitative medical centre that they attended in the afternoon. They were recruited and reported to the research team from their neurologist. Both participants lived at home with their parents, who considered the assistive technology-based program high desirable by promoting occupation and choice opportunities for their children. Consequently, their parents signed a formal consent for the participation of Bernard and Vincent to the rehabilitative intervention, which was approved by a local scientific and ethic committee.

2.2. Selection of stimuli

An informal parents interview preceded a formal preference screening (Crawford & Schuster, 1993). Thus, within a 10 min session (see below Section 2.3) 30–40 non-consecutive presentations of preferred stimuli occurred, with 15–20 s rest interval across them. Stimuli were retained whether they caused participants' alerting, orientation and/or smiling reactions, at least for the 70% of their presentations. For both participants, music and familiar voices on one side and vibrations and coloured light on the other were selected, in order to proceed to choice opportunities (see below Section 2.4).

2.3. Setting, responses, sessions and data collection

The study was carried out in a quiet room at participants' home. Both participants were sitted in front of a table (i.e. 120 cm × 60 cm) with two containers available (i.e. one on their left and one on their right) during the study. The response consisted of placing an object inside one of the two containers. Sessions lasted 10 min. Typically, 2–4 sessions per day, 4 days per week were conducted. Research assistants provided a physical and a verbal prompt every 30 s periods of non-responding. At the end of each session, prompted responses were subtracted from the total responses recorded by