Assessment and Treatment of Stereotypy in an Individual with Cornelia de Lange Syndrome and Deafblindness

Marc J. Lanovaz

Université de Montréal

John T. Rapp

Auburn University

Isabella Maciw

Université de Montréal

Author Note

Our study was conducted in accordance with the "Tri-Council Policy Statement: Ethical Conduct for Research Involving Humans" governing federally funded research in Canada. The study was supported in part by an experimentation grant from the Office des personnes handicapées du Québec (#2361-09-51). We have no conflict of interest to report.

Correspondence concerning this article should be addressed to Marc J. Lanovaz, École de Psychoéducation, Université de Montréal, C.P. 6128, succ. Centre-Ville, Montreal, QC, Canada, H3C 3J7.

Email: marc.lanovaz@umontreal.ca, tel.: 1 514 343 6111 ext. 81774

Keywords*:* behavioural intervention, blindness, Cornelia de Lange syndrome, deafness, intellectual disability, stereotypy

This is an Accepted Manuscript of an article published by Taylor & Francis in the Journal of Intellectual & Developmental Disability on August 28, 2014, available online: https://doi.org/10.3109/13668250.2014.953464

Abstract

Background: Several researchers have reduced engagement in stereotypy in individuals with intellectual disability and deafblindness using interventions containing a punishment component. The purpose of our study was to examine whether we could produce reductions in stereotypy in an individual with Cornelia de Lange syndrome and deafblindness by using noncontingent and differential reinforcement only.

Method: We used single-case experimental designs to examine the effects of noncontingent reinforcement alone and in combination with differential reinforcement of sitting on mouthing, tapping, and appropriate behaviour.

Results: Noncontingent access to edible items reduced mouthing whereas access to tactile stimuli did not. Combining noncontingent access to tactile items with differential reinforcement reduced mouthing and tapping while strengthening appropriate behaviour.

Conclusions: Antecedent- and reinforcement-based interventions were effective at reducing engagement in stereotypy in an individual with Cornelia de Lange syndrome and deafblindness without relying on punishment. However, more research is necessary to replicate our findings.

Assessment and Treatment of Stereotypy in an Individual with Cornelia de Lange Syndrome and Deafblindness

Cornelia de Lange syndrome is a congenital disorder characterized by physical abnormalities and dysmorphology, which has been associated with varying levels of intellectual disability (Barisic et al., 2008; Oliver, Arron, Sloneem, & Hall, 2008). Individuals with Cornelia de Lange syndrome often engage in repetitive and invariant behaviours that serve no apparent social function, which are generally referred to as stereotypy in the research literature (e.g., body rocking, hand flapping, mouthing, tapping; Oliver, Sloneem, Hall, & Arron, 2009; Moss, Oliver, Arron, Burbidge, & Berg, 2009). Although stereotypy is rarely physically harmful, engaging in the behaviour has been shown to interfere with social integration, adaptive functioning, and learning, which underscores the importance of targeting it for reduction (Lanovaz, Robertson, Soerono, & Watkins, 2013).

From a behaviour analytic standpoint, stereotypy is conceptualised as an operant behaviour that is maintained by the sensory stimulation that it generates (Lovaas, Newson, & Hickman, 1987). To reduce stereotypy, practitioners typically attempt to replace stereotypy by other behaviours that will produce more functional and socially acceptable forms of stimulation. However, these alternative sources of stimulation may be more challenging to identify when the individual is also affected by deafblindness. Researchers have reduced engagement in stereotypy in individuals with deafblindness, but the interventions being assessed generally contained a punishment component (e.g., Hanley, Iwata, Thompson, & Lindberg, 2000; Myrbakk, 1991; Sisson, Van Hasselt, & Hersen, 1993). In a notable exception, Lindberg, Iwata, and Kahng (1999) found that noncontingent and differential reinforcement were ineffective at reducing stereotypic self-injury in one participant with deafblindness. However, the study was limited insofar as the items provided as part of each intervention did not produce stimulation similar to the form of stereotypy emitted by the

participant (Rapp et al., 2013). From an ethical standpoint, some caregivers may not approve of using punishment-based procedures and these interventions may produce several undesirable side-effects (Lerman & Vordran, 2002). Alternatives to punishment are antecedent- and reinforcement-based interventions, which have been shown effective at reducing stereotypy in other populations and are more likely to strengthen appropriate behaviour facilitating social integration (Lanovaz et al., 2013). Therefore, the purpose of our study was to examine whether noncontingent and differential reinforcement could reduce engagement in stereotypy in an individual with Cornelia de Lange syndrome and deafblindness without relying on punishment.

Method

Participant

Tania (pseudonym) was a 43-year-old woman with Cornelia de Lange syndrome associated with profound intellectual disability and deafblindness who engaged in stereotypic mouthing and tapping. Her mouthing was potentially unsafe given that she did not see what she put in her mouth and regularly attempted to mouth items from trash cans. We also targeted tapping as prior research has shown that reducing one form of stereotypy may increase other untargeted forms, which would be counterproductive (Rapp et al., 2013). Tania spent her weekdays in a day centre where we conducted all the sessions. The centre provided recreational and vocational training activities to individuals with intellectual disability who could not be integrated in workshop settings. However, Tania was not involved in the scheduled activities because staff reported that her stereotypy and wandering interfered with her participation. We obtained informed consent from Tania's legal representative prior to her participation and conducted the study in accordance with the "Tri-Council Policy Statement: Ethical Conduct for Research Involving Humans" governing federally funded research in Canada.

Data Collection

A trained graduate research assistant measured the duration of mouthing and tapping on video recordings of the sessions. In addition to stereotypy, the research assistant also measured the duration of item engagement and sitting during the treatment assessment. We defined mouthing as non-edible objects or body parts touching any part of her mouth (including her lips), tapping as two or more consecutive brief contacts between the individual's fingers and another body part or object, item engagement as manipulating an object with her hands without tapping it or putting it in her mouth, and sitting as contact between her buttocks and a chair. Each measure was converted to a percentage by dividing the duration of the behaviour by the total duration of the session and multiplying the result by 100%. We used the block-by-block method with 10-s intervals to calculate interobserver agreement (IOA) scores for 34% of sessions. That is, we divided the smaller duration by the larger duration in each 10-s interval and then computed the mean for each session by dividing the sum of all intervals by the total number of intervals. Mean IOA scores were 98% (range: 85%-100%) for mouthing, 97% (range: 84%-100%) for tapping, 94% (range: 80%-100%) for item engagement, and 98% (range: 92%-100%) for sitting.

Experimental Design and Procedures

Prior to her inclusion in the study, Tania participated in a functional assessment for her stereotypy, which involved a series of observation sessions during which we provided no social consequences (Querim et al., 2013). The results showed that her stereotypy persisted in the absence of social consequences, suggesting that the function was non-social reinforcement. Then, we conducted a noncontingent stimulation assessment to identify stimuli associated with low levels of stereotypy and subsequently used these stimuli during the intervention. As part of the current study, Tania participated in 114 sessions over a 12-week period.

Noncontingent stimulation assessment. To identify stimuli associated with low levels of stereotypy, we alternated four conditions within a multielement design and conducted four 10-min sessions (each condition once) per day, approximately two days per week. During the baseline condition, Tania did not have access to any forms of stimulation other than that provided by the furniture and objects already present in the room. During the tactile condition, she had continuous access to various stimuli that were selected by the experimenter to ensure that each item provided a different type of tactile stimulation; the items were a tender ball, a brush, a beaded necklace, a feather scarf, and a microfiber wash mitt. During the edible condition, we provided a preferred edible item (a small piece of cookie identified via paired-choice preference stimulus assessment; Fisher et al., 1992) every 30 s. Finally, Tania had access to the tactile stimuli on a continuous basis and the edible item every 30 s during the tactile plus edible condition.

Treatment assessment. We evaluated the effects of providing stimuli associated with low levels of mouthing (i.e., edible items) contingent on sitting during access to the tactile items. We used sitting as a target for increase because (a) it was incompatible with wandering, (b) was less intrusive and easier to teach than on-task behaviour, which would have required more physical prompting, and (c) could eventually be used to facilitate the teaching of novel, more functional activities to Tania. The baseline and tactile conditions were similar to the initial assessment with two exceptions: we conducted the treatment assessment sessions in a new room (the same for all conditions) and we prompted Tania to sit in a chair at the beginning of each session. The tactile plus differential reinforcement of alternative behaviour (DRA) condition was similar to the tactile condition, but the research assistant additionally provided edible reinforcers on a variable-interval 15-s schedule contingent on Tania sitting in a chair. To signal the beginning of the conditions, the research assistant prompted Tania to touch one of the tactile stimuli at the start of the tactile condition and additionally provided

the edible item at the start of the DRA condition. To signal the end of the conditions, the research assistant manually prompted Tania to sign "finish" with her hands. We conducted five to eight consecutive 5-min sessions per day, two to three days per week. The tactile and tactile plus DRA conditions were alternated within a multielement design and compared to baseline within a BAB reversal design.

Results

Figure 1 displays the results of the noncontingent stimulation assessment. Levels of mouthing were lower in the edible (*M* = 1%) and tactile plus edible (*M* = 2%) conditions than in the baseline (*M* = 18%) and tactile (*M* = 29%) conditions. Her mouthing increased during tactile conditions because she put the tactile items in her mouth. Engagement in tapping was lower in the edible (*M* = 2%), tactile (*M* = 4%), and tactile plus edible (*M* = 2%) conditions than in the baseline condition (*M* = 11%). The results of the initial assessment suggested that providing edible items produced the most desirable effects when taking into account both forms of stereotypy, which supported their use as reinforcers during the subsequent DRA intervention. The persistence of mouthing and tapping across numerous sessions without social consequences further supported the results of the functional assessment by showing that each form of stereotypy was at least partly maintained by non-social reinforcement.

<Please insert Figure 1 about here>

Figure 2 compares the effects of noncontingent tactile stimulation, tactile stimulation plus DRA, and no intervention (i.e., baseline) on mouthing, tapping, item engagement, and sitting. During the initial phase, engagement in mouthing was generally lower in the tactile plus DRA condition (*M* = 1%) than in the tactile condition (*M* = 17%). Levels remained low during the baseline phase (*M* = 4%), which may be explained by the removal of the tactile stimuli that Tania typically mouthed. The second comparison phase replicated the initial results by showing that engagement in mouthing remained lower in the tactile plus DRA

condition (*M* = 6%) than in the tactile condition (*M* = 28%). Initially, levels of tapping were comparable during the tactile plus DRA (*M* = 1%) and tactile (*M* = 3%) conditions. Following an increase observed during the baseline phase (*M* = 7%), engagement in tapping remained nearly the same across the two treatment conditions (*M* = 1%). During both comparison phases, item engagement remained typically consistent across the tactile plus DRA (*M* = 15% and 21%) and tactile (*M* = 13% and 19%) conditions. We could not observe item engagement during the baseline phase because the tactile stimuli were unavailable. Finally, engagement in sitting gradually increased and leveled off above 90% during the tactile plus DRA condition (*M* = 80%) of the first comparison phase in contrast with levels observed in the tactile condition (*M* = 52%), which remained variable. Following the withdrawal of both interventions, levels of sitting considerably decreased (*M* = 19%), but the reintroduction of the tactile plus DRA (*M* = 95%) and tactile (*M* = 63%) conditions replicated previously observed patterns of responding.

<Please insert Figure 2 about here>

Discussion

Overall, our results indicate that combining noncontingent and differential reinforcement reduced engagement in mouthing and tapping in an individual with Cornelia de Lange syndrome and deafblindness while increasing sitting. In addition, the combined intervention did not interfere with item engagement. From a clinical standpoint, the lack of interference with item engagement may be important given that some researchers have shown that using edible items as reinforcers may reduce engagement in appropriate behaviour (e.g., Frank-Crawford et al., 2012). By reducing engagement in mouthing in an individual with deafblindness using edible items, our results replicated those of previous studies conducted with individuals who did not have visual and auditory impairments (e.g., Simmons, Kliethermes, & Smith, 2003). Our study also extends research on individuals with

deafblindness by showing that interventions that do not involve a punishment component may reduce engagement in stereotypy in this population. The noncontingent stimulation assessment may be used to identify potential reinforcers that may increase an appropriate behaviour while reducing engagement in one or more forms of stereotypy. Our results also suggest that combining differential reinforcement with noncontingent reinforcement may reduce some side-effects of the latter (i.e., increase in mouthing) while strengthening appropriate behaviour.

The study has some limitations, which should be addressed in future research. First, the trainer did not conduct a preference assessment for the tactile stimuli because we provided many different items simultaneously to reduce the probability of satiation (e.g., Lindberg, Iwata, Roscoe, Worsdell, & Hanley, 2003). Second, the intervention was relatively intensive to implement and required a one-to-one staff ratio, which limits its applicability in some settings. Following the treatment assessment, we gradually tripled the duration of the interval while still maintaining near-zero levels of mouthing and high levels of sitting. However, implementing the intervention in most settings would have required thinning the schedule to approximately once every 5 min. We were unable to meet this target because the participant developed a mouth ulcer beforehand, which reduced the reinforcing value of edible items. One potential solution could have been using non-edible items associated with low levels of mouthing as reinforcers. Future research should investigate this alternative and to what extent the schedule can be faded. Furthermore, researchers should include social validity measures to examine the broader impact of reducing stereotypy in the future. Finally, patterns during the comparison phases were considerably variable across the tactile plus DRA and tactile conditions for sitting, suggesting that Tania was not discriminating between the two intervention conditions. Because she forcefully removed any items that we had her wear (e.g., textured bracelet), we could only provide brief prompts at the beginning and end of sessions.

Researchers should use more salient stimuli in the future in order to facilitate discrimination and possibly increase the effectiveness of interventions being implemented with individuals with deafblindness.

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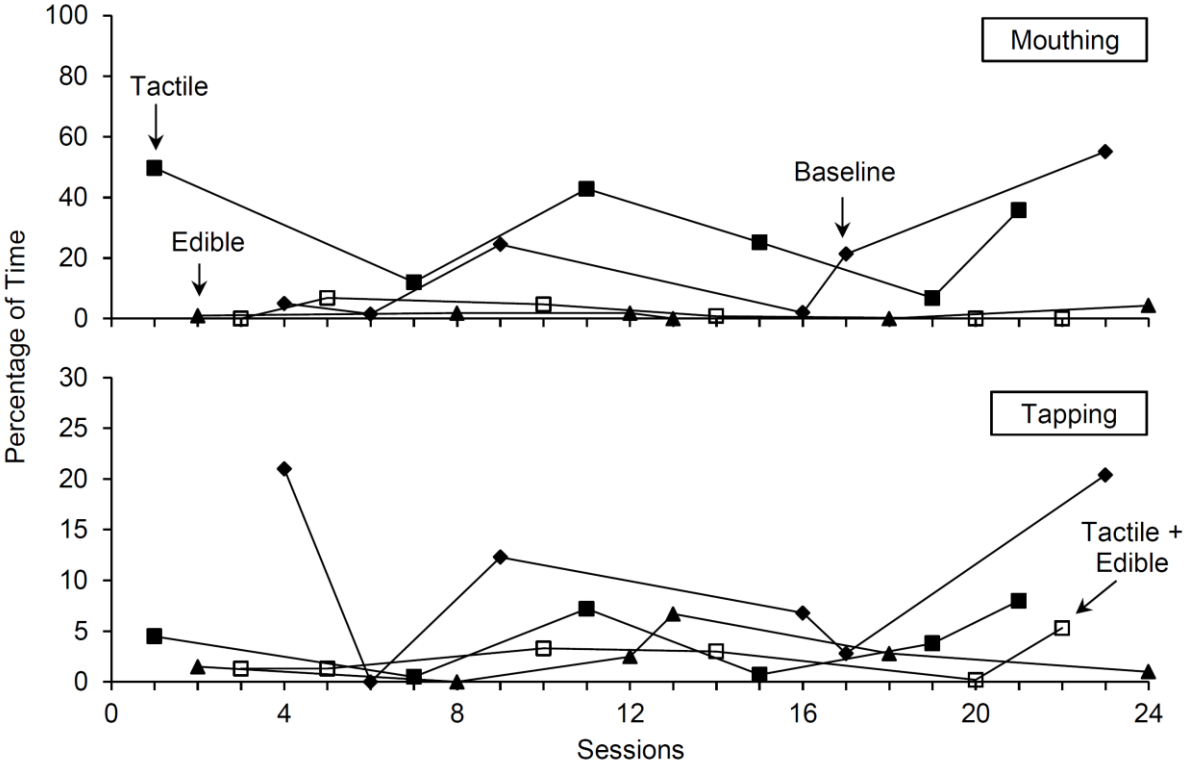

Figure 1. Percentage of time Tania engaged in mouthing and tapping during the noncontingent stimulation and baseline conditions

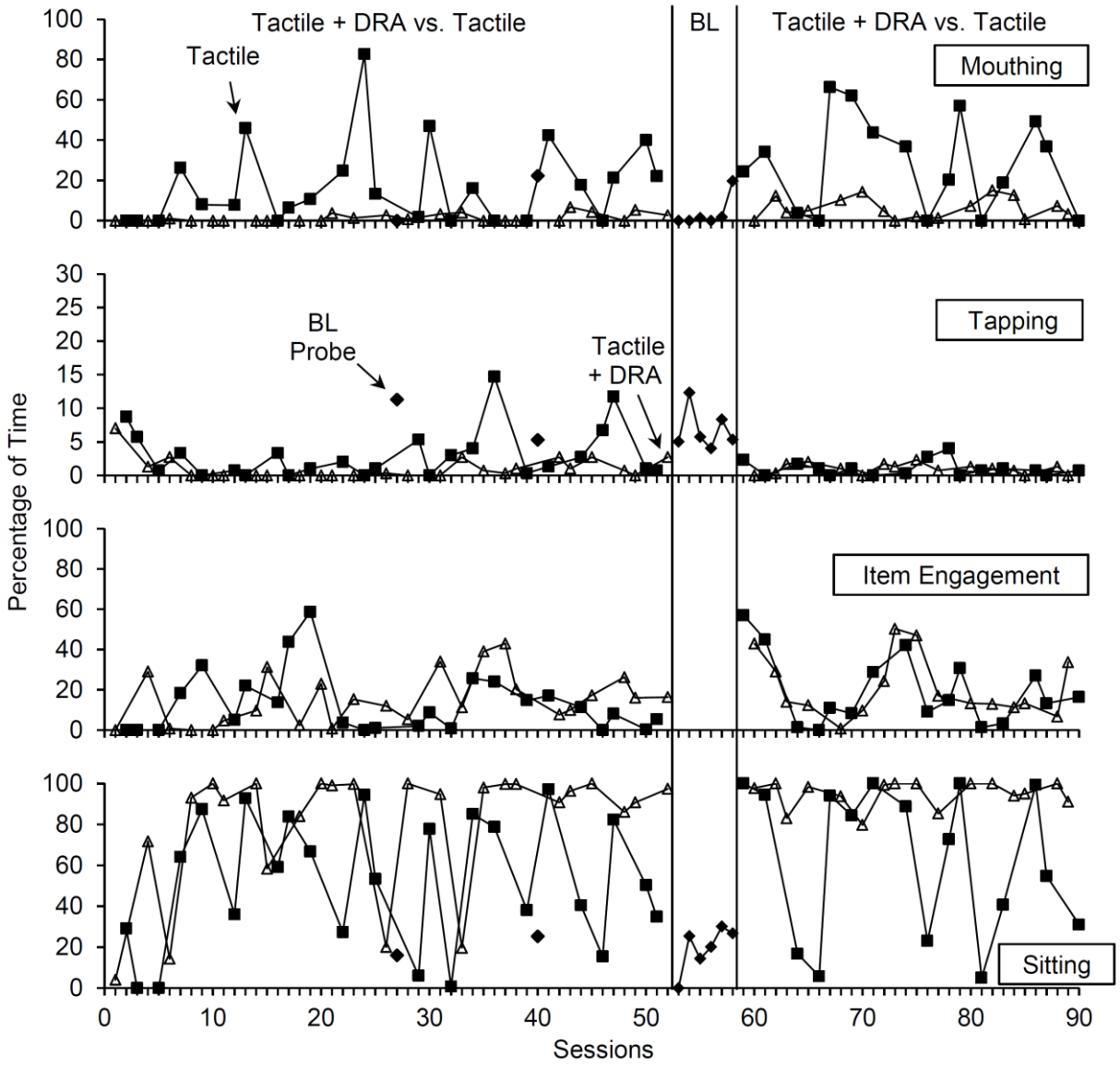

Figure 2. Percentage of time Tania engaged in mouthing, tapping, item engagement, and sitting during the treatment assessment across tactile, tactile plus differential reinforcement of alternative behaviour (DRA), and baseline (BL) conditions.