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Discrimination Training Reduces High Rate Social Approach Behaviors in Angelman Syndrome: Proof of Principle

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Abstract

This proof of principle study was designed to evaluate whether excessively high rates of social approach behaviors in children with Angelman syndrome (AS) can be modified using a multiple schedule design. Four children with AS were exposed to a multiple schedule arrangement, in which social reinforcement and extinction, cued using a novel stimulus, were alternated. Twenty five to 35 discrimination training sessions were conducted and levels of approach behaviors were measured before and after the discrimination training for two children. All four participants evidenced discrimination between conditions of reinforcement and extinction after 16-20 teaching sessions as indicated by lower rates of social approach behaviors in the presence of the S^{Δ} for extinction. Reversal effects for the two children for whom this design was implemented were evident. The results demonstrate that after repeated training, the use of a novel stimulus can serve as a cue for children with AS to discriminate adult availability. This is a potentially effective component of a broader intervention strategy but highlights the need for sustained teaching procedures within this population.

Keywords: behavioral phenotype; aggression; intervention; discrimination learning; errorless learning; Angelman syndrome; extinction

1. Introduction

Angelman syndrome (AS) is a genetic disorder with a relatively well-defined behavioral phenotype (see Horsler & Oliver, 2006a). The prevalence is approximately 1 in 10,000 to 1 in 40,000 live births (Clayton-Smith, 1993; Peterson, Brøndum-Nielsen, Hansen, & Wulff, 1995). AS is caused by a de novo deletion (accounting for approximately 68-75% of individuals with AS), Uniparental Disomy (UPD; 2-7%), an imprinting defect (2-5%) or a mutation of the UBE3A gene (2-8%) (Williams, Lossie, & Driscoll, 2001), in each case affecting the 15q11-13 region. There is also a subset of individuals with AS who have no known genetic cause (Peters, Beaudet, Madduri, & Bacino, 2004). The physical and clinical characteristics include: severe intellectual disability, seizures, ataxic gait, hypopigmentation, abnormal EEG patterns (Boyd, Harden, & Patton, 1988), microcephaly, a protruding tongue and increased levels of salivation. AS is also associated with a deficit in expressive language, with the majority of individuals being non-verbal (Williams et al., 2006). The behavioral phenotype of AS includes impulsivity and a short attention span (Walz & Benson, 2002), sleep disturbances (Bruni et al., 2006), and a 'fascination' with water and reflective objects (Didden, Korzilius, Sturmey, Lancioni, & Curfs, 2008). Perhaps the most striking aspect of the behavioral phenotype of AS is the strong drive for social attention as evidenced by frequent occurrences of laughing and smiling behaviors in the presence of adult attention (Horsler & Oliver, 2006b; Oliver, Demetriades, & Hall, 2002) and the high levels of social approach behaviors towards both familiar and unfamiliar adults reported in experimental studies and case reports (Mount, Oliver, Berg, & Horsler, 2011).

In their review of the behavioral phenotype of AS, Horsler and Oliver (2006a) found that 56 case studies (88%) noted laughing or smiling. Early reports of these behaviors described them as driven by neurological atypicality and unrelated to external or environmental events (Steffenburg, Gillburg, Steffenburg, & Kyllerman, 1996; William & Frias, 1982). However, there is a growing body of literature which suggests that laughing and smiling behaviors are related to social stimuli. In one of the first studies to explore this, Oliver et al. (2002) manipulated the level of social contact received by three children across three conditions: no interaction (alone), proximity only and full interaction. Laughing and smiling occurred more frequently in conditions of adult interaction and at almost zero levels in 'alone' conditions. Higher levels of laughing and smiling behaviors in children with AS during adult interaction were subsequently reported in both experimental studies of similar design, in less structured settings and in contrast to other children with a comparable level of intellectual disability

(Adams, Horsler, & Oliver, 2011; Horsler & Oliver, 2006a; Mount et al., 2011; Oliver et al., 2007), although these findings have not been replicated with younger children (Richman, Gernat, & Teichman, 2006). In combination these observations suggest children with AS find adult contact highly rewarding, although this has yet to be explored experimentally.

More recent investigations suggest that children with AS show high rates of social approach behaviors to gain access to social attention. Mount et al. (2011) measured the rates of social approach behaviors in eleven children with AS, and found that participants showed high levels of social approach behaviors towards both familiar (mother) and unfamiliar adults. Overall, participants were found to approach their mother more than the unfamiliar adult, although interestingly this difference was only found when the mother was giving eye contact. In combination with anecdotal reports (see Horsler & Oliver, 2006a), these experimental studies show that high levels of sociability and attention seeking behaviors are characteristic of many children with AS.

In an early questionnaire study examining problem behaviors, Summers, Allison, Lynch, and Sandler (1995) found that in a sample of 11 children under 16 with AS, all parents reported aggressive behaviors. Subsequent estimates have suggested that individuals with AS are more than three times more likely to show aggression than individuals with a comparable level of intellectual disability (Arron, Oliver, Berg, Moss, & Burbidge, 2011). Typical topographies include hair pulling and skin grabbing (Cassidy, Dykens, & Williams, 2000). In the operant literature diverse functions are reported for challenging behaviour in the general intellectual disability population, including escape from demand, access to tangibles and social resources and self-stimulation (Iwata, Dorsey, Slifer, Bauman, & Richman, 1994). The heightened drive for social attention in AS has led researchers to propose that a phenotype x environment interaction might explain the high levels of aggression as a positively socially reinforced operant behavior (McGill & Langthorne, 2011; Tunnicliffe & Oliver, 2011). However, one of the first studies to explore this using experimental functional analysis found only one of twelve children presenting attention maintained behavior, and three children showing high levels of challenging behavior in the presence of adult attention (Strachan et al., 2009). This has been supported by subsequent investigations (Allen et al., 2010; Radstaak, Didden, Oliver, Allen, & Curfs, 2012), indicating that the relationship between aggression and adult attention may be more complex than initially proposed.

Interventions for attention maintained challenging behavior have been successfully implemented with children with AS. Allen et al. (2010) conducted Functional Communication Training (FCT) with five children with AS who all displayed aggression maintained by social attention. By teaching children to use an augmentative communication device to request social interaction, the authors showed significant reductions in aggression in all five participants. The use of FCT for challenging behavior in AS has subsequently been replicated with four children with AS (Radstaak et al., 2012).

Despite the effectiveness of FCT for lowering rates of challenging behavior both with children with AS (Allen et al., 2010) and children with idiopathic intellectual disability (see Tiger, Hanley, & Bruzek, 2008), FCT and other interventions for challenging behavior (e.g. differential reinforcement) do not address the potential problem of high levels of approach behaviors when the reinforcer is not available. This is especially pertinent in AS, where high rate social approach behaviors are observed during periods of both low and high attention (Mount et al., 2011; Strachan et al., 2009). This, taken together with the emerging literature on heightened stress in parents of children with AS (Griffith et al., 2011), provides a strong rationale for the development of an intervention to address the high rates of social approach behaviours observed in times of adult unavailability.

In the applied behavior analytic literature, alternative interventions have been designed to address social approach behaviors which are 'ill-timed'. One such intervention, a 'multiple schedule approach', has successfully manipulated children's requests (mands) for social attention by teaching them to discriminate between times of reinforcer availability and unavailability using an environmental cue (Tiger & Hanley, 2004, 2006). Tiger and Hanley (2004) developed this procedure in order to address the ill-timed requests of typically developing children in a classroom environment. The authors exposed typically developing children to alternating conditions of access and denial (extinction) to social reinforcement. Conditions were signaled using novel environmental cues and verbal instructions. After discrimination training, participants approached for social attention less frequently when extinction conditions were signaled using a cue. The 'multiple schedule' approach has successfully decreased ill-timed manding for social attention with both typically developing children (Tiger & Hanley, 2004, 2006) and individuals with an intellectual disability (Kuhn, Chirighin, & Zelenka, 2010; Leon, Hausman, Kahng, & Becr, 2010), using both novel stimuli

(Tiger & Hanley, 2004) and naturally occurring activities (Kuhn et al., 2010) as environmental cues.

The successful implementation of the 'multiple schedule approach' provides a rationale for the use of this intervention with individuals who consistently request a reinforcer when it is not available. Consequently, this would suggest a potentially effective intervention for the high rate social approach behaviors observed at times of adult unavailability in children with AS (Mount et al., 2011). However, as yet, the intervention has only been conducted with individuals who show 'ill-timed' requests, not individuals who have both an unusually strong drive for social interaction and show high levels of impulsivity, such as children with AS (Walz & Benson, 2002). The combination of both these behaviors suggests that individuals with AS may have difficulties inhibiting social approach behaviors, potentially making this type of intervention more problematic. This provides a strong rationale for conducting a 'proof of principle' investigation to assess the merits of this approach within an AS population.

The aim of the current study was to evaluate whether a multiple schedule approach can be used to teach children with AS to discriminate between times of adult availability and unavailability (for delivering social attention) using an environmental cue. The study used methodology based on Tiger and Hanley (2004) and more recent investigations using a more naturalistic methodology (Kuhn et al., 2010). Extensive pilot work with four participants with AS shaped the protocol to make it more practical for individuals with a severe intellectual disability. Discrimination training was comprised of alternating conditions of access and denial of social attention, with the denial condition paired with a novel environmental cue.

2. Method

2.1 Participants

Four children with a genetic diagnosis of Angelman syndrome (AS) aged 5-10 years participated (see Table 1 for participant characteristics). Child A, Child C and Child D had a genetic diagnosis of AS caused by a deletion on the maternal chromosome 15q11-13; Child B a UBE3A mutation. All participants were non-verbal. Participants were recruited from a large database of families held at the Cerebra Centre for Neurodevelopmental Disorders. Participants were recruited for the study if they were at least 24 months old, had a genetic confirmation of AS, and parents/carers reported high levels of drive and motivation to gain access to social attention. This information was obtained via telephone interview with the parent/caregiver.

+++Insert Table 1 about here+++

2.2 Procedure

Participants were visited at their school for the experimental observations. The experimental visit was conducted in an empty classroom at the participant's school, free from peers and distractions. Where possible, objects were removed from the room or from the participants' reach. Two adults were present in the room with the participant: Researcher 1 conducting the experimental observations and Researcher 2 who video recorded the sessions.

2.2.1 Baseline

The baseline comprised alternating conditions of reinforcement and extinction in an ABABABAB design. Each condition contained three trials. Trials were used rather than the minute long conditions usually employed in multiple schedule arrangements (see Tiger & Hanley, 2004) as previous research has suggested that trials are a more effective way to teach children with intellectual disabilities new discriminations (Lovaas & Schreibman, 1971; Smith, 2001; Taubman et al., 2001). Trials ended either when the participant approached for social attention, displayed any challenging behavior or when the trial had lasted 10s. In condition A, participants were given 10s of social reinforcement if they displayed social approach or challenging behavior. In condition B, participants received no reinforcement for social approach behaviors. If the participant approached the researcher, the trial was terminated and after a 10s period the researcher proceeded to the next trial.

After one baseline session was conducted, levels of approach behavior in each of the conditions were calculated in order to assess whether participants met criteria to proceed to discrimination training. Participants had to meet two criteria to proceed to the next phase:

1) Approach behavior in over 2/3 of trials: A cut off for the number of approaches was included to account for participants who did not approach consistently and may not be motivated to gain access to social attention. As the protocol was designed as an intervention for children who show high rate social approach behavior, if participants approached in less than two thirds of trials, the baseline was repeated. All participants displayed sufficiently frequent social approach behaviors to exceed the minimum cut-off.
2) No discrimination during baseline: In addition to measuring baseline levels of social approach behavior, the baseline was designed to assess whether children discriminated between experimental conditions before the discriminative stimulus was introduced. None of the participants showed discriminated manding at baseline.

2.2.2 Discrimination training

The training phase comprised alternating conditions of reinforcement and extinction in a CDCDCDCD design. Each condition contained a specific number of trials. Trials ended either when the participant approached for social attention, displayed any challenging behavior or when the trial had lasted 10s. In condition C, participants were given 10s of social interaction if they displayed social approach or challenging behaviors. In condition D if participants approached the researcher, they were given a 10s denial of social attention before proceeding on to the next trial. Two changes to the procedure made this phase distinct from the baseline:

1) The addition of the environmental cue: In discrimination training, condition D was paired with an environmental cue – a brightly colored jacket worn by Researcher 1. This stimulus was selected in the pilot study, as it was identified as being salient but with few sensory properties, as previous research has suggested that children with AS may have a preference for sensory items (e.g. Walz & Baranek, 2006; Peters, Horowitz, Barbieri-Welge, Taylor, & Hundley, 2012). The stimulus was also chosen based on its likely low occurrence in participants' day-to-day life. In order to control for any effects of the jacket alone (i.e. regardless of the paired condition), the environmental cue was displayed randomly across conditions during baseline sessions.

2) The number of trials in conditions: The number of extinction trials was gradually increased across sessions according to errorless learning techniques. Errorless learning techniques have previously been found to be a more successful method of teaching children with intellectual disabilities (see Jones & Eayrs, 1992). Initially the ratio of C:D trials was 5:2, which then progressed to 4:2, 4:3 and finally 3:3. Initially it was decided that participants would only move on to the next ratio when they had reached criteria for discrimination. For this reason, Child A only completed sessions at the 5:2 ratio. Due to the slow rate of progression for Child A, for Child B, Child C and Child D the ratio was changed every five sessions regardless of any behaviors observed.

2.2.3 Return to Baseline

After discrimination training had been conducted, participants were again exposed to baseline sessions. These were a repeat of the initial baseline protocol. The return to baseline was only conducted with participants Child C and Child D due to time constraints[1].

2.3 Data analysis

The experimental visits were video recorded in order to allow for coding of participants' behavior. In order to assess the effectiveness and broader effects of discrimination training, two methods of analysis were employed: visual inspection, and comparison of rating scales measuring the frequency and nature of interactions (Child Sociability Rating Scales; Moss et al., in review).

2.3.1 Visual inspection

The percentage of trials with approach behaviors was graphed and inspected. Visual inspection is the most widely used method for assessing the effectiveness of a behavioral intervention, and allows for the comparison of the levels of approach behaviors across reinforcement and extinction conditions. As there are only one or two baseline and return to baseline conditions, only visual inspection allows for the comparison of behavior across baseline, return to baseline and discrimination training sessions. This allows for a broad assessment of the effectiveness of the cue. Visual inspection has previously been the

[1] The initial protocol indicated that the return to baseline should be completed when participants consistently discriminated between extinction and reinforcement conditions. As this was not consistent over sessions, Child A and Child B did not complete the return to baseline. Due to the inconsistent nature of the behavior, the protocol was amended in order to allow for the inclusion of the return to baseline regardless of behaviors observed.

predominant method used to assess the effectiveness of a multiple schedule approach (e.g. Tiger & Hanley, 2004; Kuhn et al., 2010).

2.3.2 Child Sociability Rating Scales

In addition to visual inspection, the footage was coded using the Child Sociability Rating Scales (CSRS; Moss et al., in review). These scales assess the quality and nature of a range of child and adult interactions. The scales comprise fourteen variables rated on a five point Likert scale. For the purposes of the current study, only three variables were coded – 'spontaneous initiation of interaction', 'frequency of eye contact' and 'focus of attention' (see Table 2 for definitions). 'Frequency of eye contact' and 'focus of attention' were combined in order to create an 'intention' variable, with higher scores indicating a greater focus towards the researcher. The variables were combined following the procedure outlined by Moss et al. where variable scores are multiplied and then rescaled (0=0; 1-4 =1; 5-8=2; 9-12=3; 13-16=4) to create a single score of 0-4.

+++Insert Table 2 about here+++

Other variables from the CSRS were not included as they measured the reaction to social attention; responsiveness or avoidance. Laughing and smiling behaviors were not coded as in some cases because the participant's face was turned away from the camera[2]. For each variable, trials were combined for coding, producing condition scores. Condition scores within a session were summed to create separate session scores for reinforcement and extinction conditions. A third of the footage from all four children was randomly selected for inter-rater reliability analyses. The Spearman co-efficients for the inter-rater reliability calculations were above .60 for each variable across reinforcement and extinction conditions for each participant, indicating good inter-rater reliability (Landis & Koch, 1977).

In order to analyze the data using statistical tests, first the data were analyzed in order to assess whether the data were autocorrelated – i.e. one datum point predicted the next. This is important in order to be able to analyze single case studies and use non-parametric statistics. Autocorrelation was calculated using the Durbin-Watson statistic, and cut-offs for autocorrelation were taken from Durbin-Watson tables. The data from each variable for Child

[2] This did not affect the 'eye contact' variable, as participants had to turn their head in order to make eye contact with the researcher, and then their faces could be seen.

A, Child B and Child D were not autocorrelated, therefore statistical analyses could be conducted. The data for Child C was found to be autocorrelated, therefore statistics reported for the data from Child C are displayed for completeness and should be interpreted cautiously.

As the data were not normally distributed, non-parametric tests were conducted. Wilcoxon signed ranks tests were conducted to assess whether there were any significant differences in the frequency of the behaviors coded across reinforcement and extinction conditions. The analyses were conducted for discrimination training sessions only as only one to three baseline sessions were conducted and return to baseline sessions were only conducted for two participants.

3. Results

3.1 Visual inspection

Figure 1 depicts the percentage of trials in which social approach behaviors were observed for Child A, Child B, Child C and Child D respectively. As the levels of approach behaviors varied considerably between sessions, percentages were calculated across blocks of five sessions to enable more clear visual comparisons of behavior patterns across reinforcement and extinction conditions.

+++++Figure 1 about here+++++

3.1.1 Baseline

During baseline, all four participants manded for social attention in over two thirds of trials, meeting the specified minimum cut-off to proceed to discrimination training. Child D exceeded the cut-off in baseline 1, and Child A, Child B and Child C in baseline 2. During baseline, levels of approach behaviors across reinforcement and extinction conditions were similar for Child A (approaches in 83% of reinforcement trials and 75% of extinction trials), Child B (71% of reinforcement trials and 67% of extinction trials), Child C (53% of reinforcement trials and 56% of extinction trials) and Child D (75% of reinforcement trials and 75% of extinction trials). Due to the consistent levels of approach behaviors across conditions, none of the participants showed discriminated manding at this stage. As the levels of approach behaviors exceeded the cut-off and none of the participants met criteria for discriminated manding, all four participants proceeded to the discrimination training phase.

3.1.2 Discrimination training

Figure 1 shows the percentage of trials in which participants displayed approach behaviors in discrimination training sessions. 25-35 training sessions were conducted with each participant. During the first fifteen sessions of discrimination training, levels of social approach behaviors were similar across reinforcement and extinction conditions. None of the participants showed over 10% difference in the percentage of trials with approach behaviors across conditions. From sessions 15-20 onwards, all children showed initial signs of discriminated manding, with higher percentages of trials with approach behaviors in reinforcement conditions. Child D in particular displayed the most noticeable difference, manding in 50-60% of trials in reinforcement conditions compared to 20-30% in extinction conditions. Child C also showed a marked difference in the levels of approach behaviors across conditions from session 21 onwards, manding in over 48% of trials in reinforcement

conditions, compared to 20-30% in extinction conditions. Child A and Child B showed a slight difference in the levels of approach behaviors across conditions in the last five sessions with a more modest 20% difference in behavior, however, both participants took part in fewer sessions than Child C and Child D due to time constraints.

3.1.3 Return to baseline

Figure 1 shows the percentage of trials in which participants displayed social approach behaviors in the return to baseline. Due to time constraints, the return to baseline was only conducted with Child C and Child D. Levels of social approach behaviors across conditions of reinforcement and extinction when the cue was removed were similar for Child C (51% of C trials and 36% of D trials) and Child D (25% of C trials and 33% of D trials).

3.2 Child Sociability Rating Scales (CSRS)

3.2.1 Initiation of interaction

The frequency of social approach behaviors displayed during discrimination training was coded using the variable 'spontaneous initiation of interaction'. Higher scores on this variable indicate a greater frequency of initiating behaviors displayed by participants (see Table 2 for a full description). Figure 2 shows the median session rating scores across reinforcement and extinction conditions during discrimination training for Child A, Child, B, Child C and Child D.

+++Insert Figure 2 about here+++

A Wilcoxon signed ranks test was conducted to evaluate whether there was a significant difference in the frequency of initiating behaviors across reinforcement and extinction conditions. The analysis indicated significantly higher levels of initiating behaviors displayed in reinforcement conditions by Child A ($Z = -3.811$, $p<0.01$), Child B ($Z = -3.49$, $p<0.01$) and Child D ($Z = -2.80$, $p<0.01$). Child C also displayed significantly higher levels of initiating behaviors in reinforcement conditions ($Z=-3.20$, $p<0.01$), as suggested by Figure 2. However, as the data from KS are autocorrelated, these statistics should be treated cautiously.

3.2.2 Intention

Variables 'focus of attention' and 'frequency of eye contact' were coded from one to five on a Likert Scale, with higher scores indicating a greater focus of attention towards the researcher. The variables were combined to make an 'intention' variable, indicating where

the focus of participants' attention remained. Figure 2 shows the median session rating score across reinforcement and extinction conditions for Child A, Child B, Child C and Child D.

Wilcoxon signed ranks tests showed no significant difference in intent across reinforcement and extinction conditions for Child A ($Z=-0.8$, $p=0.38$), Child B ($Z=-1.56$, $p=0.12$), and Child D ($Z=-1.58$, $p=0.11$). Child C showed significantly higher levels of focus towards the researcher in extinction conditions ($Z=-2.91$, $p<0.01$), although as the data are autocorrelated, these data should be treated cautiously.

4. Discussion

The current study was designed to examine the use of a multiple schedule approach to manipulate levels of social approach behaviors in four children with Angelman syndrome (AS). This is the first study which directly addresses the high levels of social approach behaviors observed in children with AS; a 'proof of principle' investigation designed to teach children to discriminate between times of adult availability using an environmental cue. The social approach behaviors of participants were recorded during alternating conditions of social reinforcement and extinction paired with an environmental cue. The results show discrimination learning in all participants. One participant in particular showed consistently lower levels of social approach behaviors in extinction conditions in the presence of the cue. The results from this study are a promising indication that this may be a useful component of intervention for children with AS in order to manage the high levels of approach behaviors described in the literature (e.g. Mount et al., 2011).

The main hypothesis was that after discrimination training, participants would evidence association of the presence/absence of the environmental cue with the availability of adult attention. Visual inspection of graphed approach behaviors in reinforcement and cued extinction conditions revealed that this hypothesis was at least partially confirmed in all four participants. During the first 20-25 discrimination training sessions all participants showed similar levels of approach behaviors towards the researcher across reinforcement and extinction conditions. However, during the final 10-15 discrimination training sessions, all participants showed comparatively lower levels of social approach behaviors in extinction conditions suggesting that participants were beginning to discriminate between times of adult availability using the environmental cue. Child D in particular showed a consistent and sustained difference in the frequency of social approach behaviors across conditions during discrimination training, with 13 sessions with lower responding in extinction conditions and a 44% difference in the frequency of approach behaviors across conditions in the final six sessions. Although Child A, Child B and Child C showed a more modest difference in behavior across conditions, this difference in behavior was maintained for at least ten sessions.

Evidence of discrimination learning was also supported by the ratings of the frequency and quality of participants' behaviors using the Child Sociability Rating Scales (CSRS; Moss et

al., in review). Statistical analyses revealed that the frequency of initiating behaviors was significantly lower in extinction conditions for Child A, Child B and Child D. The data for Child C also indicate fewer initiating behaviors in extinction conditions, however as the data are autocorrelated this could not be supported by statistical analyses. The results support the findings from the visual inspection, and suggest that participants used the environmental cue to discriminate between times of adult availability. Once again, this supports the original hypothesis that after discrimination training, children would begin to discriminate between times of adult availability using the novel environmental cue. This is consistent with previous investigations which have tested the use of a multiple schedule approach with individuals with and without an intellectual disability (e.g. Kuhn et al., 2010; Tiger & Hanley, 2004).

In contrast to social approach behaviors, participants' focus of attention did not appear to differ across conditions. Participants' 'intention' – a combination of the 'focus of attention' and 'frequency of eye contact' CSRS variables – was not significantly different across conditions for Child A, Child B and Child D. This suggests that for these participants that although their social approach behaviors were manipulated by the multiple schedule approach, the focus of attention remains consistent across conditions, regardless of the environmental cue. This is in contrast to the striking and consistent difference in social approach behaviors across conditions in discrimination training. This may provide an initial indication that although the multiple schedule approach influenced behavior, it did not change the *motivation* for social attention for these participants, as evidenced by vigilance for adult contact, and merits further investigation in subsequent investigations.

Although there is initial indication that participants were beginning to discriminate between times of adult availability using the environmental cue, the difference in social approach behavior across reinforcement and extinction conditions is neither as consistent nor prolonged as previous studies implementing a multiple schedule approach (e.g. Kuhn et al., 2010; Leon et al., 2010; Tiger & Hanley, 2004, 2006). Across all participants, lowered manding for attention in the presence of the cue was only observed from session 20-25 onwards, and was not consistently lower as in other studies using this protocol. Although this may potentially indicate a weakness in the design of the intervention, it is more likely that the absence of a consistent and prolonged difference in the frequency of approach behaviors across conditions is due to the time constraints on the intervention. There are indications in both the data from the current study and the literature on Angelman syndrome which suggest

that the time constraints on training may have impacted on the rate of acquisition. Child D, who showed greater a difference in behaviors across reinforcement and extinction conditions was exposed to at least five extra training sessions than other participants, and was therefore given a greater number of opportunities both for learning, and displaying discriminated manding. This is supported by the literature on implementing a multiple schedule approach with individuals with an intellectual disability. Although typically developing children have been shown to display discriminate manding after relatively few training sessions, discrimination learning with individuals with an intellectual disability appears to comparatively slower, with changes in behavior only observed after over 100 training sessions in some cases (Leon et al., 2010). This provides a strong rationale for the investigation of the intervention over a longer time period.

The literature on Angelman syndrome provides further indication as to why there may not have been a consistent and prolonged change in behavior during discrimination training. There is an increasing body of literature which suggests that a disruption in information in the UBE3A gene affected in AS is thought to cause particular difficulties with learning (Jiang et al., 1998). This is supported by research which has focused on teaching children with AS new skills and behaviors using ABA techniques. For example, a longitudinal study using discrete trial learning to teach adaptive behaviors to three children with AS showed very slow rates of learning, with only one child only mastering five target responses after extended teaching over 12 months (Summers & Szatmari, 2009). In addition to slow rates of learning, the authors also found no transference in skill learning i.e. when one behavior had been 'mastered', participants were no quicker to learn the next behavior.

In addition to deficits in learning associated with a disruption in the UBE3A gene, a long learning period could also be a consequence of children's strong drive for social attention and high levels of impulsivity. To date, no studies have evaluated the use of a multiple schedule approach with children with an 'excessive' drive for social attention as described in the literature on AS (see Horsler & Oliver, 2006a); most focus either on typically developing children in the classroom (e.g. Tiger & Hanley, 2004) or individuals with an intellectual disability of heterogeneous cause (e.g. Leon et al., 2010). The multiple schedule approach has also not yet been implemented with individuals who have high levels of impulsivity. There is an increasing body of literature which suggests a relationship between impulsivity and deficits in inhibition skills, most notably in Attention Deficit Hyperactivity Disorder (ADHD;

Barkley, 1997). This suggests that the high levels of impulsivity reported in AS (Walz & Benson, 2002) may make the inhibition of pre-potent responses more challenging. This, taken together with the strong drive for social attention, may go some way to account for the long learning period during discrimination training. This provides further support for extended repeats of training sessions in future interventions with children with AS.

One striking observation when examining the data is the inconsistent frequency of social approach behaviors across discrimination training session. Each participant displayed great variability in approach behaviors across sessions, with the prevalence of behavior ranging from 0 to 100% of trials. This is inconsistent with previous literature which describes a strong drive for social attention as a key part of the behavioral phenotype of AS (Oliver et al., 2002; Mount et al., 2011), and that children of this age should be showing high levels of sociability (Adams et al, 2011). This leads to the question of whether the drive for social attention is either as strong or consistent as previously described in the literature (see Horsler & Oliver, 2006a). To date, although the literature often reports a heightened sociability in AS, the majority of experimental research focuses on laughing and smiling behaviors. There is limited research which directly explores children's *drive* for adult attention, and how consistent this is under different social and environmental conditions. One important factor which could influence levels of social approach behaviors could be the familiarity of the researcher. It could be that the reported high levels of social approach behaviors are only directed towards parents or caregivers, or for individuals with whom they have previous reinforcement history. This is supported by experimental observations which suggest that children with AS show higher levels of social approach behaviors towards their caregiver rather than the unfamiliar adult under certain circumstances (Mount et al., 2011).

The social environment may have also influenced the variability in social approach behaviors across sessions. There is an increasing body of literature suggesting that children with AS are as likely to try and *maintain* social interaction rather than initiate it. This may partially explain why participants had inconsistent levels of approach behavior in times of responsive rather than continuous interaction. Although research suggests that children with AS laugh and smile more in the presence of social interaction (e.g. Oliver et al., 2002), this research was conducted using conditions of *continuous* social interaction and does not explore motivation for adult attention at times of low attention. This is supported by research which suggests that children are more likely to maintain social interaction (Mount et al., 2011), and

by the research on challenging behavior in AS suggesting higher levels of aggression in the presence of adult attention (Strachan et al., 2009). The inconsistencies between previous literature and this study merit further investigations to explore the conditions for children's sociability including: the strength of the drive for social attention, satiation, and the drive for social attention from familiar/unfamiliar adults and across different settings.

There are some limitations to this study which may affect the validity of the research. The first limitation is a potential sampling bias. The nature of the study meant that participants were only selected if they were identified as being motivated for social attention. Thus, some participants were ruled out as parents reported a low or inconsistent drive for social attention. Although this was essential for the intervention i.e. there had to be a 'problem' with social motivation to justify the intervention, it is important to note that not all children of parents who expressed an interest in the study had a strong motivation for social attention. This is inconsistent with previous literature which suggests a consistently strong motivation for adult attention in AS (Oliver et al., 2002; Horsler & Oliver, 2006a; Oliver et al., 2007; Mount et al., 2011; Adams et al., 2011). As a result of the inclusion criteria, the rates of social approach behavior observed in the current study do not represent the population of children with AS and future interventions using discrimination training may only be successful with individuals with a strong motivation for adult attention.

A second limitation to this study is the absence of data concerning the maintenance of intervention effects and programmed generalization to other people and settings, due to time constraints. Many studies using the multiple schedule approach generalize the intervention to other people (e.g. parents/caregivers), other settings (e.g. home/school) and using alternative environmental cues (see Kuhn et al. 2010; Leon et al., 2010). Although this does not directly affect the results of the study, it does affect the clinical utility of the intervention across all settings. Future investigations should be conducted to assess the effectiveness of this intervention across different situations, testing a range of environmental cues. Despite the importance of these limitations, these issues do not directly affect the results of the study - they only have implications for the use of the intervention. As this was a 'proof of principle' study to explore whether discrimination training would be an effective intervention which merits further investigation, this study is only the first step to designing and implementing effective interventions with children with AS in a more naturalistic setting.

The results of this study indicate that discrimination training is an appropriate component of intervention to address high levels of approach behaviour observed in some children with AS. Despite the initial indications of an effective intervention, the data also merit further investigations exploring the effectiveness of the intervention when implemented over a longer time period and the conditions for heightened sociability in AS. Once these elements of behaviour have been examined in more detail and modifications have been made to the procedure, this intervention has the potential to decrease levels of social approach behaviour at times when a parent/carer is unavailable. Not only does the intervention have the potential to affect levels of parental stress associated with inappropriately timed social requests, the multiple schedule approach may also provide a consistent and effective method to make the environment more predictable for children.

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References

Adams, D., Horsler, K. & Oliver, C. (2012). Age related change in social behaviour in children with Angelman syndrome. *American Journal on Medical Genetics (Part A), 155*, 1290-1297.

Allen, D., Oliver, C., Petty, J., Webster, P., Reid, D., Villa, D., Beaumont, S., Hyland, S., Arowolo, I., Jones, A., Gorniak, S., Radstaake, M. (2010). Behavioural intervention for challenging behaviour in children with Angelman syndrome. *Journal of Intellectual Disability Research, 54*, 885-885.

Arron, K., Oliver, C., Berg, K., Moss, J., and Burbidge, C. (2011). Prevalence and phenomenology of self-injurious behaviour in genetic syndromes. *Journal of Intellectual Disability Research, 55*, 109-120.

Barkley, R. A. Behavioral inhibition, sustained attention, and executive functions: Constructing a unifying theory of ADHD. *Psychological Bulletin, 121*, 65-94.

Boyd, S. G., Harden, A., & Patton, M. A. (1988). The EEG in early diagnosis of the Angelman (happy puppet) syndrome. *European Journal of Paediatrics, 147*, 508-513.

Bruni, O., Ferri, R., D'Agostino, G., Miano, S., Roccell, M., & Elia, M. (2004). Sleep disturbances in Angelman syndrome: A questionnaire study. *Brain and Development, 26*, 233–240.

Cassidy, S. B., Dykens, E., & Williams, C. A. (2000). Prader-Willi and Angelman syndromes: sister imprinted disorders. *American Journal of Medical Genetics, 97*, 136-146.

Clayton-Smith, J. (1993). Clinical research on Angelman syndrome in the United Kingdom: observations on 82 affected individuals. *American Journal of Medical Genetics, 46*, 12-15.

Didden, R., Korzilius, H., Sturmey, P., Lancioni, G., & Curfs, L. (2008) Preference for water-related items in Angelman syndrome, Down syndrome and non-specific intellectual disability. *Journal of Intellectual and Developmental Disability, 33,* 59–64.

Doyle, T. F., Bellugi, U., Korenberg, J. R. & Graham, J. (2004). 'Everybody in the world is my friend' hypersociability in young children with Williams syndrome. *American Journal of Medical Genetics, 124,* 263-273.

Dykens, E. (1995) Measuring behavioral phenotypes: provocations from the 'New Genetics'. *American Journal of Medical Genetics, 99,* 522-32.

Griffith, G., Hastings, R.P., Oliver, C., Howling, P., Moss, J., Petty, J. et al. (2011). Psychological well-being in parents of children with Angelman syndrome, Cornelia de Lange syndrome and Cri du Chat syndromes. *Journal of Intellectual Disability Research, 55,* 397-410.

Holm, V. A., Cassidy S. B., Butler M. G., Hanchett, J. M., Greenswag, L. R., Whitman, B.Y., & Greenberg, F. (1993) Prader-Willi syndrome: consensus diagnostic criteria. *Pediatrics, 91,* 398-402.

Horsler, K., & Oliver, C. (2006a). The behavioural phenotype of Angelman syndrome. *Journal of Intellectual Disability Research, 50,* 33-53.

Horsler, K. & Oliver, C. (2006b). Environmental influences on the behavioural phenotype of Angelman syndrome. *American Journal on Mental Retardation, 111,* 311-321.

Iwata, B. A., Dorsey, M. F., Slifer, K. J., Bauman, K. E., & Richman, G. S. (1994). Toward a functional analysis of self-injury. *Journal of Applied Behavior Analysis,* 27, 197-209.

Jiang, Y. H., Armstrong, D., Albrecht, U., Atkins, C. M., Noebels, J. L., Eichele, G., Sweatt, J. D., & Beaudet, A. L. (1998). Mutation of the Angelman ubiquitin ligase in mice causes increased cytoplasmic p53 and deficits of contextual learning and long-term potentiation. *Neuron, 21,* 799-811.

Jones, R. S. P., & Eayrs, C. B. (1992). The use of errorless learning procedures in teaching people with a learning disability: A critical review. *Journal of Applied Researh in Intellectual Disabilities, 5*, 204-212.

Kuhn, D. E., Vhirighin, A. E., & Zelenka, K. (2010). Discriminated functional communication: A procedural extention of functional communication training. *Journal of Applied Behavior Analysis, 43,* 249-264.

Landis, J. R., & Koch, G.G. (1977). The measurement of observer agreement for categorical data. *Biometrics, 33*, 159–174.

Leon, Y., Hausman, N. L., Kahng, S., & Becraft, J. L. (2010). Further examination of discriminated functional communication. *Journal of Applied Behaviour Analysis, 43,* 525-530.

Lovaas, O. I., & Schreibman, L. (1971). Stimulus overselectivity of autistic children in a two-stimulus situation. *Behaviour Research and Therapy, 9*, 305-310.

McGill, P., & Langthorne, P. (2011) Gene-environment interactions and the functional analysis of challenging behaviour in children with intellectual and developmental disabilities. *Behavioral Development Bulletin, 11*, 20-25.

Moss, J., Howlin, P., Beaumont, S., Tunnicliffe, P., Petty, J., Hastings, R., Griffith, G, Villa, D., Yates, R. and Oliver, C. (in review). Social behavior and characteristics of Autism Spectrum Disorder in Angelman, Cornelia de Lange and Cri du Chat syndromes.

Mount, R., Oliver, C., Berg, K., & Horsler, K. (2011). Effects of adult familiarity on social approach behaviours in Angelman syndrome. *Journal of Intellectual Disability Research, 55,* 339-350.

Oliver, C., Demetriades, L., & Hall, S. (2002). The effect of environmental events on smiling and laughing behavior in Angelman syndrome. *American Journal on Mental Retardation, 107,* 194-200.

Oliver, C., Horsler, K., Berg, K., Bellamy, G., Dick, K., & Griffiths, E. (2007). Genomic imprinting and the expression of affect in Angelman syndrome: What's in the smile? *Journal of Child Psychology and Psychiatry, 48,* 571-579.

Oliver, C., Woodcock., K. A. & Adams, D. (2010). The Importance of aetiology of intellectual disability. Learning Disability A life Cycle Approach, Second Edition. Chapter 10, pages 135-146. McGraw Hill, Open University Press, Berkshire, UK.

Peters, S. U., Beaudet, A. L., Madduri, N., & Bacino, C. A. (2004). Autism in Angelman syndrome: implications for autism research. *Clinical Genetics, 66,* 530-536.

Peters, S. U., Horowitz, L., Barbieri-Welge, R., Taylor, J. L., & Hundley, R. J. (2012). Longitudinal follow-up of autism spectrum features and sensory behaviours in Angelman syndrome by deletion class. *Journal of Child Psychology and Psychiatry and Allied Disciplines, 53,* 152-9.

Petersen, M. B., Brøndum-Nielsen, K., Hansen, L. K., & Wulff, K. (1995). Clinical, cytogenetic, and molecular diagnosis of Angelman syndrome: estimated prevalence rate in a Danish county. *American Journal of Medical Genetics, 60,* 261-262.

Radstaake, M., Didden, R., Oliver, C., Allen, D., & Curfs, L. M. (2012). Functional analysis and functional communication training in individuals with Angelman syndrome. *Developmental Neurorehabilitation, 15,* 91-104.

Richman, D. M., Gernat, E. & Teichman, H. (2006). Effects of social stimuli on laughing and smiling in young children with Angelman syndrome. *American Journal on Mental Retardation, 111,* 442-446.

Smith, T. (2001). Discrete Trial Training in the Treatment of Autism. *Focus on Autism and Other Developmental Disabilities, 16,* 86-92.

Steffenburg, S., Gillberg, C., Steffenburg, U, & Kyllerman, M. (1996). Autism in Angelman syndrome: a population based study. *Pediatric Neurology, 14,* 131-136.

Strachan, R., Shaw, R., Burrow, C., Horsler, K., Allen, D., & Oliver, C. (2009). Experimetnal functional analysis of aggression in children with Angelman syndrome. *Research in Developmental Disabilities, 30,* 1095-1106.

Summers, J. A., Allison, D. B., Lynch, P. S., & Sandler, L. (1995). Behaviour problems in Angelman syndrome. *Journal of Intellectual Disability Research, 39,* 97–106.

Summers, J. A., & Szatmari, P. (2009). Using discrete trial instruction to teach children with Angelman syndrome. *Focus on Autism and Other Developmental Disabilities, 4,* 216-226.

Taubman, M., Brierley, S., Wishner, J., Baker, D., McEachin, J., Leaf, R.B. (2001). The Effectiveness of a group discrete trial instructional approach for preschoolers with Developmental disabilities. *Research in Developmental Disabilities, 22,* 205-219.

Tiger, J. H., Hanley, G. P. (2004). Developing stimulus control of preschooler mands: An analysis of schedule-correlated and contingency-specifying stimuli. *Journal of Applied Behavior Analysis, 37,* 517–521.

Tiger, J. H., Hanley, G. P., & Bruzek, J. (2008). Functional communication training: A review and practical guide. *Behavior Analysis in Practice, 1,* 16–23.

Tiger, J. H., Hanley, G. P., Heal, N. A. (2006). The effectiveness of and preschoolers' preferences for variations of multiple-schedule arrangements. *Journal of Applied Behavior Analysis, 39,* 475–488.

Tunnicliffe, P., & Oliver, C. (2011). Phenotype-environment interactions in genetic syndromes associated with severe or profound intellectual disability. *Research in Developmental Disabilities, 32,* 404-418.

Turk, J., Sales, J. (1996). Behavioural Phenotypes and their Relevance to Child Mental Health Professionals. *Child Psychol Psychiatry Rev, 1,* 4–11.

Walz, N.C., & Baranek, G.T. (2006). Sensory processing patterns in persons with Angelman syndrome. *American Journal of Occupational Therapy, 60,* 472–479.

Walz, N. C. & Benson B. A. (2002). Behavioural phenotypes in children with Down syndrome, Prader-Willi syndrome, or Angelman syndrome. *Journal of Developmental and Physical Disabilities, 14,* 307-21.

Williams, C.A., Beaudet, A.L., Clayton-Smith, J., Knoll, J.H., Kyllerman, M., Laan, L.A. et al. (2006). Angelman syndrome 2005: Updated consensus for diagnostic criteria. *American Journal of Medical Genetics. Part A, 140*, 413–418.

Williams, C. A., & Frias, J. L. (1982). The Angelman ("happy puppet") syndrome. *American Journal of Medical Genetics, 11,* 453-460.

Williams, C. A., Lossie, A., & Driscoll, D. (2001). Angelman syndrome: mimicking conditions and phenotypes. *American Journal of Medical Genetics, 101,* 59-64.

Table 1

Participant characteristics

	Age	Gender	Genetic subtype	VABS adaptive composite	Challenging behaviors identified (CBI)
Child A	10	Female	Deletion	40 (L)	Yes
Child B	5	Female	UBE3A mutation	49 (L)	Yes
Child C	8	Female	Deletion	47 (L)	Yes
Child D	8	Female	Deletion	45 (L)	Yes

Table 2

Verbal description of the three variables coded using the CSRS: 'initiation of interaction', 'focus of attention' and 'frequency of eye contact'.

Variable	Definition
Spontaneous initiation of interaction	Initiation of interaction may be verbal or non-verbal (e.g. approaching the examiner, offering or requesting objects, speaking or signing, touching the examiner to attempt to gain their attention (aggressively or otherwise), gesturing or pointing to an object while looking at examiner.
Focus of attention	Objects focus vs. people focus
Frequency of eye contact	Eye contact defined as the participant looking up/at the examiner, fixating on their eyes or face.

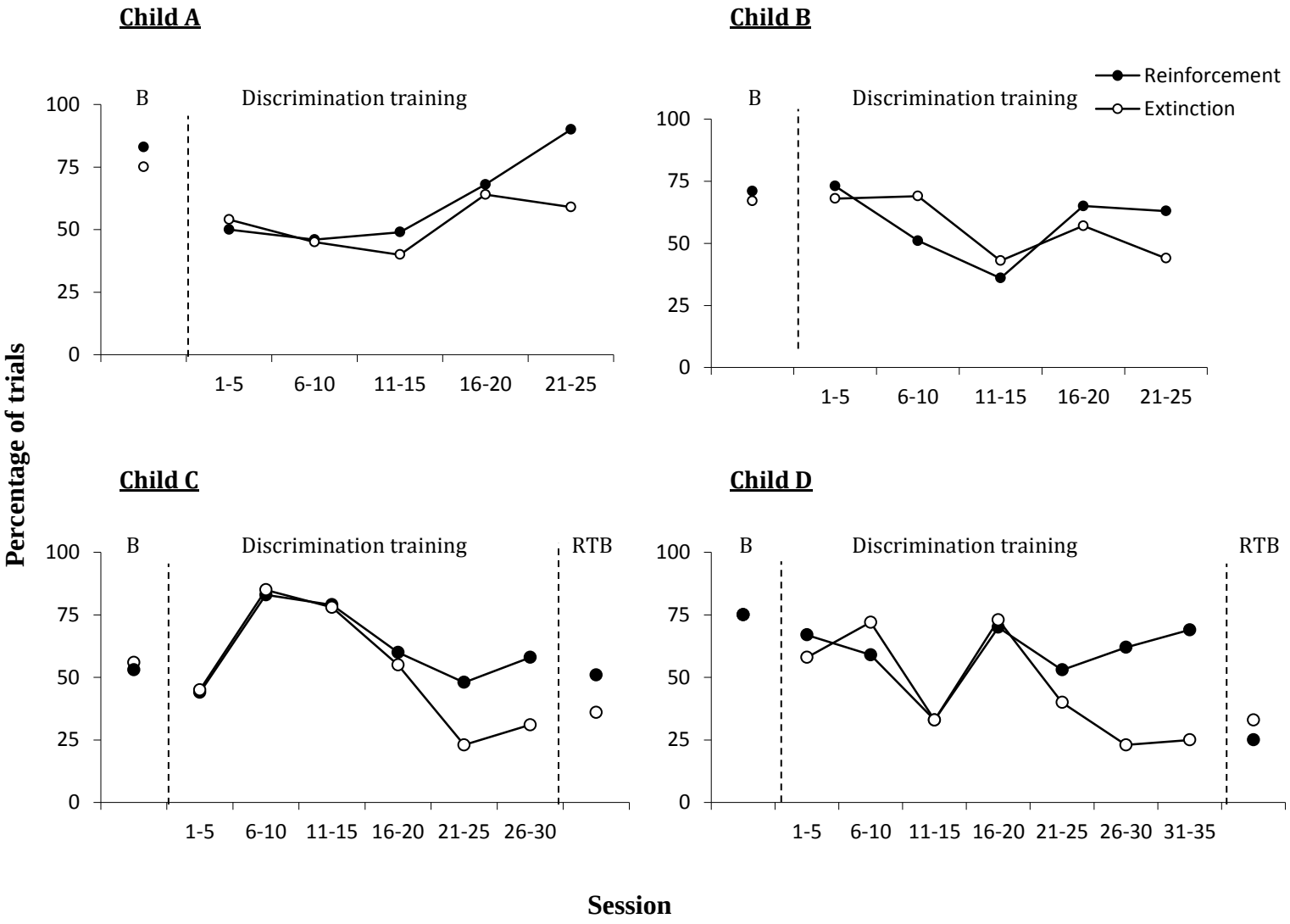

Fig. 1. Percentage of trials with social approach behaviors across Baseline (B), Discrimination Training and Return to Baseline (RTB) sessions for Child A, Child B, Child C and Child D.

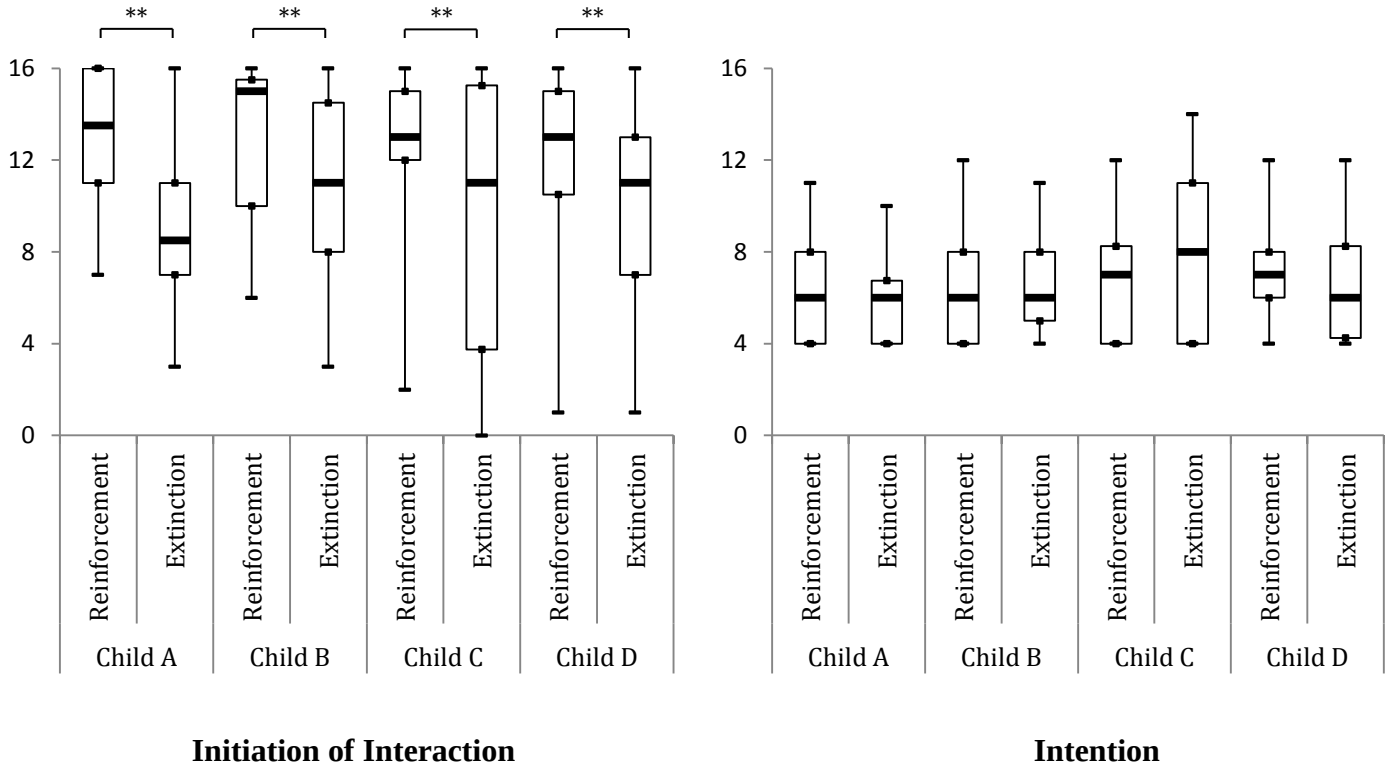

Fig. 2. Median Child Sociability Rating Scale scores for the variables ‘Initiation of Interaction’ and ‘Intention’.