Abstract

We investigated the relationship between age and laughing and smiling in children with Angelman syndrome. Twenty-four children with Angelman syndrome were exposed to three experimentally manipulated conditions; *proximity only*, *restricted social interaction* and *social interaction*. Children smiled the most in the social interaction condition and the least in the proximity only condition confirming the effect of social interaction on these behaviors. There was a decline in smiling and laughing in the oldest group (13.4-15.9 years) only in the social interaction condition. This trajectory of a decline in resource soliciting behaviors with age is consistent with predictions based on kinship theory.

Keywords: Angelman syndrome; genomic imprinting; kinship theory; intellectual disability; behavioral phenotype; emotion signaling

INTRODUCTION

Angelman syndrome occurs in approximately 1 in 40,000 live births (Clayton-Smith, 1993; Buckley, Dinno, & Weber, 1998) and is caused by disruption of a maternally inherited portion of chromosome *15q 11-13* (Clayton-Smith & Laan, 2003; Knoll, Nicholls & Lalande, 1989). There are four known genetic mechanisms: approximately 68-75% of individuals have a deletion on the maternally derived chromosome *15q 11-13*; 2-7% have uniparental disomy (where both copies of chromosome 15 are paternally inherited); 2-5% have an imprinting defect and 8-11% have a mutation in the UBE3A gene which lies at the *15q 11-13* locus (Jiang, Lev-Lehman, Bressler, Tsai, & Beaudet, 1999; Williams, Lossie & Driscoll, 2001). Between 5-20% (dependent upon sample and extent of molecular investigations) of individuals with the physical and behavioral features of Angelman syndrome show no identifiable abnormalities in the *15q 11-13* region (Clayton-Smith & Laan, 2003; Laan *et al.*, 1998; Lossie *et al.*, 2001; Williams *et al.*, 2001).

The physical presentation of Angelman syndrome includes movement or balance disorder, causing ataxic gait, microcephaly, and a high prevalence of epilepsy and abnormal EEGs. Other physical features that are commonly noted (in 20-80% of individuals) are hypopigmentation (i.e. light hair color compared to to family), feeding problems during infancy, wide mouth with widely spaced teeth, protruding tongue and high levels of sialorrhea (drooling). The behavioral phenotype is reviewed extensively by Horsler and Oliver (2006a). Of note are severe to profound intellectual disability, raised levels of laughing, smiling and ‘happy’ demeanor, excessive sociability, hand flapping, little or no speech, hyperactivity and sleep disturbances. Aggression is reported in 6-10% of individuals (Summers, Allison,

Lynch, & Sandler, 1995), notably pulling or grabbing people or hair (Cassidy, Dykens, & Williams, 2000).

There has been very little research into the relationship between age and the phenomenology of behavioral phenotypes generally or Angelman syndrome specifically. Whilst the identification of changes in phenotypes with age is not new (e.g. changes in physical phenotype noted in Coffin-Lowry syndrome (Hunter, 2002) and changes in behavior in older adults with Down syndrome (e.g. Adams *et al.*, 2008)), it remains a neglected area (Oliver & Hagerman, 2007). With an increase in life expectancies and in the number of genetically determined syndromes that have been identified, the effect of aging on behavioral and physical phenotypes is becoming an important issue.

There is a very limited body of literature focusing upon changes with age in children and adults with Angelman syndrome. Clayton-Smith (2001) conducted clinical interviews with carers of 28 individuals with Angelman syndrome (nineteen with deletions, seven with UBE3A mutations, one with uniparental disomy and one with an imprinting defect). The interview focused mainly upon physical and health related changes with age and gave little detail on behavioral features other than that "all adults seen continued to have characteristic, happy, sociable behavior for most of the time". Likewise, Laan, den Boer, Hennekam, Renier, and Brouwer (1996) interviewed carers of 28 adults with Angelman syndrome, reporting high levels of a "happy demeanor" but did not investigate how or if this behavior changed with age.

Buntinx *et al.*'s (1995) cross-sectional questionnaire study is at present the only study focusing upon changes in behavior with age in individuals with Angelman syndrome. Splitting their sample of 47 individuals with Angelman syndrome (with

unreported genetic etiologies), aged 0-47 years into three age groups (0-2 years, 2-16 years, 16 and over), they identified changes in the prevalence of "happy disposition" and "bursts of laughter" between age groups although statistical analyses were not conducted to substantiate these findings. Both behaviors showed an increased prevalence between the infant group (0-2 years) and the child group (2-16 years). After this, the prevalence of "bursts of laughter" continued to increase into adulthood (>16 years) whilst the prevalence of "happy disposition" decreased to below that seen in the infant group. However, there are important methodological issues to consider when interpreting these data including the collection of retrospective data for eleven of the fifteen children within the 0-2 year age group and a lack of reported reliability or validity for the questionnaire used. These methodological issues, combined with a somewhat mixed pattern of reported change in behavior, highlight the need for further research focusing upon the prevalence of these laughing and smiling behaviors in individuals with Angelman syndrome across the age span.

Initially, the laughing and smiling behaviors in Angelman syndrome were thought to be pathological due to suggestions of being context-inappropriate and random in nature (e.g. Dooley, Berg, Pakula & MacGregor, 1981; Williams & Frias, 1982). However, recent experimental literature using robust observational measures suggests that these behaviors are affected by the environment (Oliver, Demetriades & Hall, 2002), and are significantly more frequent in the presence of adult attention and eye contact (Horsler & Oliver 2006b; Oliver *et al.*, 2007). However, such environmental control was not replicated in Richman, Gernat, and Teichman's (2006) study of two young children with Angelman syndrome (18 and 42 months) which may

reflect differences between the samples, particularly with regard to age, or the methodology. Further studies are required in order to replicate early findings and extend the existing literature

There are three aims to this study. First, to extend the work of Horsler and Oliver (2006b) by evaluating whether specific behaviors seen in children with Angelman are evoked by social interaction. In line with previous studies and a functional interpretation of these behaviors, we hypothesize that laughing and smiling behaviors will be higher when social interaction, particularly eye contact, is present than when it is not, but other recorded behaviors will not differ. Secondly, we will investigate the effect of age on laughing and smiling behaviors in children with Angelman syndrome. Based upon observations made by Horsler and Oliver (2006b), we predict that lower levels of laughing and smiling will be observed in older children than in younger children with Angelman syndrome. Finally, we will investigate the relationship between adaptive behavior and laughing and smiling behaviors in children with Angelman syndrome in order to establish whether changes in laughing and smiling behavior are associated with age or an increase in adaptive skills (which occur with age). Based upon the work of Horsler and Oliver (2006b), we hypothesize that there will be no relationship between adaptive behavior and laughing and smiling behaviors in Angelman syndrome.

MATERIALS AND METHODS

Participants

Nineteen individuals with Angelman syndrome with confirmed deletion of *15q 11-13* (in order to maximize phenotypic homogeneity) were recruited by approaching the

parents of the twenty-six children with Angelman syndrome aged under sixteen years that had consented to take part in three previous studies; Horsler and Oliver (2006b), Mount, Horsler and Oliver (in review) and Strachan *et al.* (in Press). Of those who did not agree to take part, five (19%) families did not reply, one (4%) family had moved to another country and one (4%) child had been moved to full time residential care.

The data collected for the nineteen recruited in the present study were combined with the original data collected by Horsler and Oliver (2006b) from five individuals with confirmed deletions of *15q 11-13* (participants 2, 4, 6, 8 and 11) who did not reply to the invitation to take part in this study. This was done in order to increase the sample size and broaden the age range of the participants.

The combination of these two datasets resulted in a sample of 24 children (thirteen male, eleven female) with Angelman syndrome, all of whom had a confirmed deletion of *15q 11-13*. The mean age of the participants was 10.5 years (*sd*= 3.2, range 4-15.9). The mean age of the composite adaptive behavior score from the Vineland Adaptive Behavior Scales-2 (Sparrow, Chichetti, & Balla, 2005) was 32.4 (*sd*=7.7, range 11-47; for comparison, general population mean 100, standard deviation 15). This corresponds to a profound intellectual disability for one child, severe for seventeen children and moderate for six children. All of the children lived at home and attended local schools for children with intellectual disabilities.

Procedure

In order to fulfill the first aim of the study of evaluating whether specific behaviors seen in children with Angelman are evoked by the social condition, the experimental

conditions used by Horsler and Oliver (2006b) were conducted with each participant in a quiet room with minimal distractions (i.e. no other individuals present, toys covered up, televisions turned off) at home (n=22) or at school (n=2). Each participant was observed (and videotaped) while exposed to three conditions: a control condition; *proximity only* (condition A: adult sits adjacent to participant, maintaining a neutral facial expression and does not look, talk to or touch participant), a *restricted social interaction* condition (condition B: adult sits adjacent to participant while talking as per a normal social interaction, but maintains a neutral facial expression and does not look at the participant) and a *social interaction* condition (condition C: adult sits adjacent to participant while talking, giving physical contact, smiling, laughing and maintaining eye contact as per normal social interaction). Each of these three conditions lasted 30 seconds and were undertaken with a familiar adult; for seventeen children this was the mother and for seven children it was a female classroom assistant with whom they had regular contact.

The two social interaction conditions were alternated between repeated presentations of the *proximity only* condition in two series. The order of conditions in series 1 was ABACABACA and in series 2 ACABACABA. Each series lasted approximately 4.5 minutes with a five minute break between each series. The rationale for this length and number of conditions is based upon maintaining the participant's compliance with the observations whilst maximizing opportunities for data collection (see Horsler & Oliver, 2006b).

Dependent variables

Four adult behaviors and seven participant behaviors were recorded across conditions. In order to maintain consistency, five of these behaviors (*child*

laugh/smile, *adult look*, *adult talk*, *adult laugh/smile* and *adult touch*) and their definitions are taken from Horsler and Oliver (2006b) with six additional participant behaviors taken within Mount *et al.* (in review); *child look at adult*, *child reach*, *child touch adult body*, *child touch adult head*, *child pull adult body* and *child pull adult head*. .

Inter-observer agreement

Inter-observer agreement was assessed by a second observer simultaneously but independently recoding behavior for 20% of all data collected which included sessions from all conditions and all participants. Kappa indices were calculated based on a 10-second interval-by-interval comparison of observer records. The Kappa coefficients for each behavior range from .75 to .89. As all indices were greater than .6, inter-observer reliability can be considered good (Landis & Koch, 1977).

Integrity of conditions

To assess the integrity of the independent variables within and across conditions, the behaviors of the familiar adults were coded and analyzed. The percentage of time the familiar adult smiled/laughed, talked, or looked towards the face of the child was 0.7%, 2.5% and 1.1% respectively in the *proximity only* condition. In the *restricted social interaction* condition the mean percentage of these adult behaviors was 8.4%, 79.5%, and 2.7% respectively and within the *social interaction* condition 28.1%, 84.1%, and 83.3% respectively. These indices suggest that the integrity of the independent variables was maintained across conditions

Data analysis

To examine the effect of manipulations of social interaction on the child behaviors, the percentage of time the child spent showing each of the target behaviors (hereafter referred to as duration) was calculated within each of the three conditions. Kolmogorov-Smirnov tests indicated that the data were normally distributed, therefore a series of within-subjects ANOVAs was adopted to compare the duration of each of the behaviors across each of the conditions. Where significant results are identified, planned post-hoc analyses were undertaken using the Bonferroni correction. This is suggested to be the most appropriate correction for planned comparisons (Clark-Carter, 1997) and is effective in minimizing the risk of a type I error (Field, 2000). Therefore, for all post-hoc comparisons within this section, the Bonferroni-corrected α–value was set to .0167.

To explore the relationship between age and laughing and smiling behaviors the data were subjected to two stages of analyses. Again, Kolmogorov-Smirnov tests indicated that the data were normally distributed and therefore parametric statistics were used. Firstly, the association between duration of these behaviors (both overall and in each social condition) and age was explored using Pearson's correlations. A second analysis was then undertaken to identify at which point the duration of laughing and smiling differed significantly from that seen in the youngest age group. To do this, the sample was divided into four age groups; youngest (4.0-8.3 years), medium young (8.4-10.3 years), medium old (10.4-13.3 years) and oldest (13.4-15.9 years), each containing 25% (n=6) of the participants. A 4 x 3 mixed design analysis of variance was undertaken with social condition as the within-participants factor, age group as the between-participants factor and *child laugh/smile* duration as the dependent variable. Post-hoc comparisons within this analysis were corrected using

the Bonferroni correction, resulting in a corrected α–value of .0112. As the ANOVA had a relatively small number of participants within each age group, a linear regression analysis was subsequently undertaken to strengthen the conclusion drawn regarding the between age and the duration of laughing and smiling behaviors.

In order to fulfill the final aim of the study; to explore whether duration of specific behaviors are associated with ability on a standardized measure, correlations were undertaken between adaptive behavior composite scores from the Vineland Adaptive Behavior Scales and the child variables.

All analyses were undertaken on SPSS 16 where a conservative α–value of .01 was used for all non-bonferroni corrected analyses.

RESULTS

The mean durations for the seven child dependent variables were calculated. Three child behaviors had a mean duration of more than 10% across conditions. These were *child look* (mean=14.3%, *sd*=9.3), *child laugh/smile* (mean=25.9%, *sd*=17.7), and *child touch adult body* (mean=14.8%, *sd*=20.2). Four child behaviors had a mean duration of less than 10% across all three conditions; these were *child pull adult body* (mean=7.1%, *sd*=9.8), child pull adult head (mean=2.0%, *sd*=5.4), *child reach* (mean=.41%, *sd*=.82), and *child touch adult head* (mean=1.1, *sd*=3.9). Therefore the variables *child touch adult body*, *child touch adult head*, *child pull adult body*, *child pull adult head* and *child reach* were combined as per Mount *et al.* (in

review) to create one variable; *child approach adult* which had a mean duration across all conditions of 23.2% (*sd*=22.8).

The effect of social interaction on behavior

To test the hypothesis that specific behaviors in children with Angelman syndrome are evoked by social interaction, a series of within-participants ANOVAs were undertaken comparing the duration of the three variables *child look*, *child laugh/smile,* and *child approach adult* across the three conditions; *proximity only*, *restricted social interaction* and *social interaction*.

There were no significant effects of condition on *child look* ($F(2,46)=2.5$, $p=.096$) or *child approach adult* ($F(2,46)=.72$, $p=.49$). However, the within-participants ANOVA undertaken to compare percentage of time that the child spent laughing and smiling in each of the three conditions revealed a significant difference ($F(2, 46)=37.3$, $p<.001$). Bonferroni corrected post-hoc analyses highlighted significant differences between all three conditions. As shown in Figure I, the children smiled significantly more in the *social interaction* condition than in the *restricted social interaction* ($p<.001$) and *proximity only* conditions ($p<.001$). They also smiled significantly less in the *proximity only* condition compared to the *restricted social interaction* conditions ($p=.001$).

(place Figure I about here)

In summary, the series of within-participants ANOVAs highlighted that the only behavior to differ significantly between social conditions was laughing and smiling. The duration of this behavior systematically reduced with the degree of adult contact.

The duration of behaviors across the age span

To test the hypothesis that lower levels of laughing and smiling will be observed in older than younger children with Angelman syndrome, three child behaviors were correlated with child chronological age (in months); *child look*, *child laugh/smile*, and *child approach adult*. As the duration of *child laugh/smile* differed significantly between the *proximity only*, *restricted social interaction* and *social interaction* conditions in the first analysis, the analyses was undertaken separately for each of the three conditions. The results are presented in Table I.

(place Table I about here)

Using Pearson's correlation, no significant associations were found between chronological age and overall duration of *child look*, *child approach adult*, *child laugh/smile* overall or *child laugh/smile* in the *proximity only* condition or *child laugh/smile* in the *restricted social interaction* condition. The only condition where the duration of *child laugh/smile* is significantly correlated with age is the *social interaction* condition ($R(23)=.-.65$, $p=.001$). These results show that the duration of *child laugh/smile* behaviors decrease as age increases, but only within the *social interaction* condition.

The association between the duration of laughing and smiling behaviors and age

In order to clarify the nature of the relationship between age and duration of *child laugh/smile*, the sample was split into four equal groups based upon age in months; youngest (4.0-8.3 years), medium young (8.4-10.3 years), medium old (10.4-13.3 years) and oldest (13.4-15.9 years), each containing 25% of the participants. A 4 x 3 mixed design analyses of variance was undertaken with duration of *child laugh/*smile

as the dependent variable, social condition as the within-participants factor and age group as the between-participants factor. As sphericity could not be assumed, the Greenhouse-Geisser correction was applied.

The results are plotted in Figure II. There was significant interaction between age and social condition ($F(6,40)=6.44$, $p=.001$). Post-hoc comparisons indicate a significant difference in the duration of child laughing and smiling behaviors between the youngest and the oldest groups only ($F(3,20)=5.2$, $p=.009$) in the *social interaction* condition but no differences in the *proximity only* ($F(3,20)=1.4$, $p=.26$) or the *restricted social interaction* conditions ($F(3,20)=2.1$, $p=.13$). There was a main effect of condition ($F(2,40)=63.7$, $p<.001$) that can be explained by the interaction, but there was no main effect of age group ($F(3,20)=2.1$, $p=.13$). These results indicate that age is an important factor in the duration of laughing and smiling behaviors seen in children with Angelman syndrome in response to social stimuli, and eye contact specifically.

(place Figure II about here)

As the number of participants within each group entered into the ANOVA was small, a linear regression was undertaken to confirm the results. The results of the linear regression support the finding of the ANOVA, showing that laughing and smiling behaviors systematically decrease with increasing age ($R^2 = .42$, $F(1,22)=15.7$, $p=.001$).

The association between the duration of laughing and smiling behaviors and adaptive behavior

In order to fulfill the final aim of the study; to explore whether duration of specific behaviors are associated with ability on a standardized measure, the duration of the three child variables, *child look*, *child laugh/smile*, and *child approach adult* were correlated with the adaptive behavior composite from the Vineland Adaptive Behavior Scales. As the duration of *child laugh/smile* differed significantly between the *proximity only*, *restricted social interaction* and *social interaction* conditions, the duration of *child laugh/smile* in each of the three conditions were also entered into the correlation. The results are presented in Table I.

Using Pearson's correlation, no significant associations were found between the adaptive behavior composite and overall duration of *child*, *child laugh/smile* (overall and for each condition) and *child approach adult*. This therefore suggests that there is no association between adaptive behavior and the behaviors observed within this study.

DISCUSSION

In this study, the laughing and smiling behaviors of children with Angelman syndrome (with confirmed deletions in *15q 11-13*) were differentially evoked when parameters of social interaction were manipulated. Differences between social conditions were not found for *child look* or *child approach adult* behaviors. The use of tightly controlled experimental paradigms allowed for the control of specific social variables. In combination with operational definitions of behaviors and robust inter-rater reliability, this minimizes threats to internal and construct validity.

The laughing and smiling behaviors were significantly higher in the *social interaction* condition (involving adult speech, touch, smiling, laughing and eye

contact) than the *proximity only* condition. The pattern of results suggest a relationship between the components of social interaction and the duration of laughing and smiling behaviors, with post-hoc comparisons revealing that the duration of laughing and smiling is significantly lower in the *proximity only* (no touching, talking or eye contact) than in the *restricted social interaction* condition (touching and talking without eye contact) and both had significantly lower durations of laughing and smiling than the *social interaction* condition (touching and talking with eye contact).

The systematic manipulation of the environment within this study identified higher levels of laughing and smiling behaviors within the *social interaction* condition than the *restricted social interaction* condition. The one variable that was systematically manipulated between these two conditions was the presence of eye contact. The higher duration of laughing and smiling behaviors within the condition where eye contact was present (*social interaction* condition) suggests that children with Angelman syndrome may find this reinforcing. This could be further investigated by examining response acquisition in a reinforcement program with and without eye contact. If eye contact is reinforcing for children with Angelman syndrome (as suggested by an increase in smiling and positive affect during eye contact conditions as compared to those without eye contact), this could have significant implications for therapeutic strategies used by parents and carers for managing difficult behavior.

Although the integrity of the conditions was reported and deemed acceptable by the authors, there was slight variability in adult laughing between the *restricted social interaction* and *social interaction* conditions. It is unlikely that this would be solely

responsible for the significant difference in the child laughing and smiling behaviors between the conditions, as Oliver *et al.* (2007) demonstrated that child laughing and smiling reliably precedes adult laughing and smiling within this population, a pattern that was not noted within their matched control group. This suggests that adult laughing in this study was a consequence rather than a cause of child laughing and smiling.

The duration of laughing and smiling behaviors was significantly negatively correlated with age within the *social interaction* condition only. This suggests that it is not simply the duration of laughing and smiling that decreases as the children get older but the capacity for social interaction and eye contact to evoke laughing and smiling behaviors. This is consistent with descriptions by Horsler and Oliver (2006b) who describe low levels of laughing and smiling, and difficulties in collecting full sets of data, from the oldest children within their sample. There were no significant associations between an informant-based estimate of adaptive behavior, the Vineland Adaptive Behavior Scales, and any of the child behaviors investigated. This suggests that the associations found with age are related to chronological age and not simply increasing skills or abilities. However, little data was collected on other factors, including medical conditions and factors (e.g. epilepsy, medication, sleep disorders) which could potentially effect behavior and therefore confound the results.

The 4 x 3 mixed-design ANOVA highlighted that the oldest age group (13.4-15.9 years) smiled significantly less than the youngest group (4-8.3 years) only within the *social interaction* condition. Although equal, the sample size of each of the age groups (n=6) is small for an analysis of variance and this cannot be ignored.

However, it could be argued that achieving a difference of this significance with potentially low power and with the application of the Greenhouse Geisser correction (which is considered an over-conservative adjustment with small sample sizes; Maxwell & Arvey, 1982) highlights the strong effect of age on these behaviors. Due to the small sample sizes within the age groups, a linear regression was undertaken to confirm the results. However, this regression analyses alone would not have identified the position of significant decline in laughing and smiling behaviors in the oldest age group (13.4-15.9), a trajectory that warrants further investigation.

The error bars in Figure II highlight the variability of the data within each of the age groups. It is interesting to note that the error bars are largest within the 10.4-13.3 year age group; the group just before the decline becomes prominent. They are at their smallest within the youngest and oldest age groups. This suggests that the duration of laughing and smiling behaviors evoked by social conditions is more homogenous within the youngest (4-8.3 years) and oldest participants (13.4-15.9 years). However, during the “transition” period (from high levels of smiling in response to social interaction to low levels of smiling in response to social interaction) there appears to be greater variability in the duration of the behavior. This suggests that this design would benefit from being replicated on a larger scale using age groups with a smaller range (but larger number of participants) or, ideally, using longitudinal designs to begin to map these changes in behavior more accurately.

Although this sample has a broad age range, further studies using a broader age range could provide valuable insight into the association between age and laughing and smiling. Richman *et al.* (2006) did not identify any association between laughing

and smiling behaviors in their two young children with Angelman syndrome, although their environmental conditions differed somewhat from the three (*proximity only, restricted social interaction, social interaction*) used by Horsler and Oliver (2006b) and within this study. For example, participants had access to toys during the *social interaction* condition and there was no manipulation (or measurement) of eye contact by the adult. There have been no studies using experimental manipulation of the environment with individuals with Angelman aged sixteen or over. Further research should encompass both the younger and older individuals with Angelman syndrome to help to further understand the association between age and these characteristic laughing and smiling behaviors.

It could be suggested that the decline in laughing and smiling behaviors with age is in line with that predicted by the genomic conflict and kinship theory (Haig & Wharton, 2003; Brown & Consedine, 2004), although it is important to bear in mind that these theories are in their infancy with regards to empirical support. It states that although most genes have the same effect regardless of whether they were inherited from the mother or father, for a small group of genes, the parent or origin does matter. This phenomenon is referred to as genomic imprinting. Summarized by Haig and Wharton (2003) and Brown and Consedine (2004), the kinship theory (or genomic conflict hypothesis) of genomic imprinting proposes that paternal and maternal alleles favor different behavioral expressions. Paternal alleles ensure survival of the offspring by increasing the cost to the offspring's mother whilst the maternal alleles promote maternal survival by reducing these costs.

Regardless of genetic subtype (e.g. uniparental disomy or deletion), the maternal genes on chromosome *15q 11-13* are not expressed in individuals with Angelman

syndrome. As a consequence of this, kinship theory suggests that individuals with Angelman syndrome may be genetically predisposed to show behaviors that increase access to social resources in a competitive setting, for example, laughing and smiling. Oliver *et al.*'s (2007) observational study supports this, suggesting that the laughing and smiling behaviors seen in children with Angelman syndrome have a powerful social function. They result in the allocation of more social resources for children with the syndrome in terms of adult attention, smiling and eye contact than the same behaviors shown by a matched comparison group. Therefore, it might be hypothesized that as a child ages and requires less maternal resources, resource soliciting behaviors will decrease. This would be consistent with a decrease in other behaviors that solicit maternal resources, such as hyperactivity (Clayton-Smith, 2001).

A control group was not recruited for this study as the presence of elevated levels of laughing and smiling behaviors in children with Angelman syndrome in comparison to other children with intellectual disabilities is already well established (e.g. Oliver *et al.*, 2007). However, it is acknowledged that a control group could have considerably strengthened this study by allowing comparisons of changes in the frequency of such behaviours at specific ages in relation to aspects of the environment. In order to further understand the changes in the laughing and smiling behaviors in relation to developmental trajectory, future research could use developmental and chronologically age matched control groups. Not only would these protect against threats to internal validity within the study, but they would allow for the use of methods proposed by Thomas *et al.* (2009) and Annaz, Karmiloff-

Smith, and Thomas (2008) whose analysis allow for comparisons to both mental and chronologically age matched control groups.

Whilst this study shows that laughing and smiling behaviors are heightened within the *social interaction* condition, it cannot be inferred that this behavior reflects an increase in positive emotion or 'happiness'. Oliver *et al.*'s (2007) sequential lag analysis led them to suggest that the children with Angelman syndrome smile as a result of the positive affect that they experience upon initiating social contact. Social reinforcement may also account for some of the crude aggressive behaviors described within the literature (e.g. hair pulling, grabbing; Summers *et al.*, 1995; c.f. Taylor & Oliver, 2008) if they provide a method of gaining or sustaining social interaction (see Oliver *et al.* 2007, for further discussion). This hypothesis warrants further investigation using experimental designs with adequate sample sizes.

Overall, the results of this study provide further evidence that laughing and smiling in Angelman syndrome is not simply sporadic and can be evoked by social contact. It has also provided evidence for a decrease in the duration of laughing and smiling with age, and then only when eye contact is present. The implications of these findings are that both professionals and families may benefit from more accurate descriptions and information about these behaviors. The potential for social interaction, and particularly eye contact, to be reinforcing has obvious clinical implications within behavioral management programmes as social contact is an easy to administer reinforcer that may be highly resistant to satiation in this group (Oliver *et al.*, 2007). However, the decline in laughing and smiling in the *social interaction* condition with age may reflect a decreased potency in eye contact and social attention as a reinforcer as the children reach adolescence and therefore early

intervention allow us to maximise the potential reinforcing properties of social interaction within this population.

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TABLES

Table I: Correlation matrix for age, adaptive behaviour and child behaviours

	Child laugh smile				**Child look at adult**	**Child approach adult**
	All conditions	*Proximity only*	*Restricted social interaction*	*Social interaction*	All conditions	All conditions
Age (years)	.04	.14	.07	-.65**	-.005	.26
Adaptive behaviour composite	.03	.04	.05	.14	.01	.02

** indicates significant at the .001 level

FIGURES

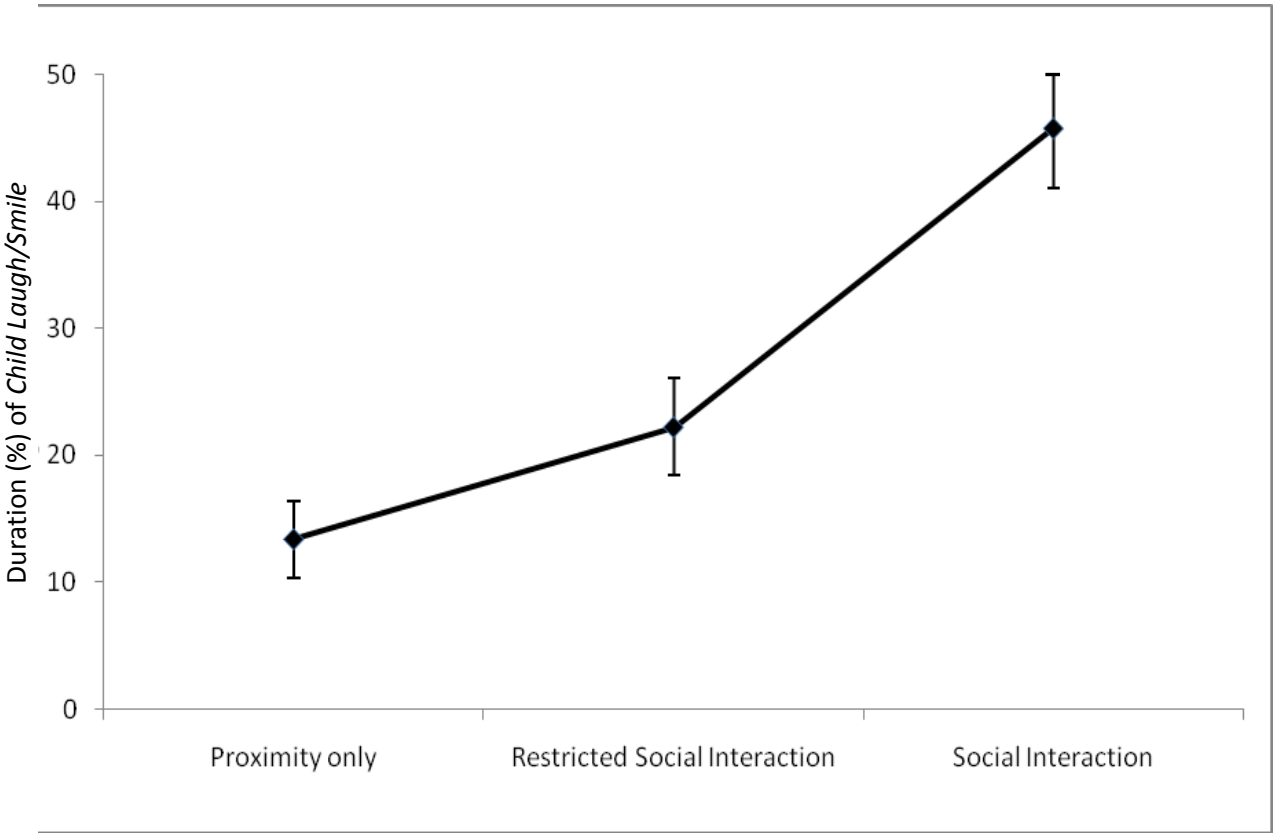

Figure I: Mean (± one SE) percentage of time of *child laugh/smile* by condition

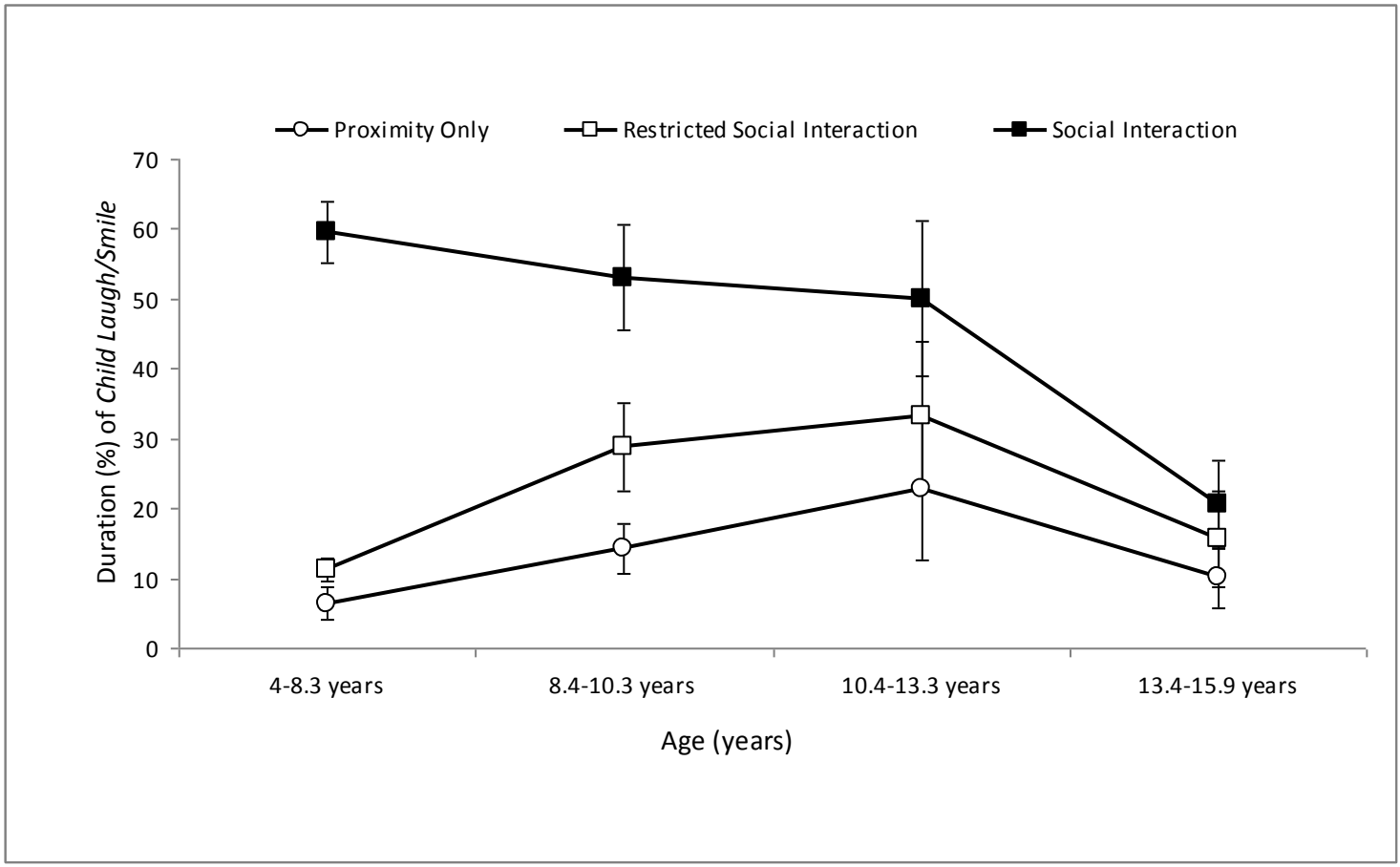

Figure II: Mean (± one SE) percentage of time of *child laugh/smile social interaction* condition by age group