Environmental Influences on the Behavioral Phenotype of Angelman Syndrome

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Abstract

Using observational methods, we examined the social influences on laughing and smiling behavior in children with Angelman syndrome by systematically manipulating aspects of social interaction. Seven boys and 4 girls who were between 4 and 11 years of age and who had a confirmed maternal deletion of chromosome 15q11-q13 completed the study. Each child was observed while repeatedly exposed to three conditions in which parameters of social interaction were manipulated. Laughing and smiling behavior varied across all children and was significantly heightened in a condition involving adult speech, touch, smiling, laughing, and eye contact. The findings highlight the importance of examining environmental and social influences on purported phenotypic behavior in genetic syndromes.

Angelman syndrome is a rare neurodevelopmental disorder with prevalence estimates ranging from 1 in 10,000 to 1 in 20,000 (Clayton-Smith & Pembrey, 1992; Peterson, Brondum-Neilson, Hasen, & Wulff, 1995). The genetic cause of Angelman syndrome is a maternally derived abnormality in the region of chromosome 15q11-q13 (Knoll et al., 1989). There are four main genetic mechanisms that can lead to an abnormality in this region: interstitial deletion of chromosome 15q11-q13 (70% to 75% of individuals); uniparental disomy, when both copies of chromosome 15 are inherited from the father (2% to 3%); an imprinting defect, including an imprinting center deletion (3% to 5%); and a mutation within the gene UBE3A (5% to 10%) (Clayton-Smith & Laan, 2003).

As is the case with many genetic syndromes, a raised prevalence of specific physical characteristics and behaviors and physical and behavior variability is evident in individuals with Angelman syndrome. Consensus for diagnostic criteria of this syndrome developed by the Scientific and Research Advisory Committee of the Angelman Syndrome Foundation in 1995 (Williams et al., 1995), identified four clinical characteristics that were 100% consistent: developmental delay, speech impairment, movement disorder, and behavioral uniqueness (any combination of frequent laughter/smiling, apparent happy demeanor, easily excitable personality, hypermotoric behavior, and short attention span). The consensus also identified frequent characteristics (80%): microcephaly, seizures, an abnormal EEG, and "associated" characteristics (20% to 80%), such as flat occiput, occipital groove, protruding tongue, tongue thrusting, feeding problems during infancy, prognathia, wide mouth, widely spaced teeth, frequent drooling, excessive chewing/mouthing behaviors, strabismus, hypopigmented skin, hyperactive lower limb reflexes, uplifted flexed arm position, increased sensitivity to heat, attraction to water, and sleep disturbance.

The consensus statement on Angelman syndrome includes behaviors in addition to physical characteristics and, thus, acknowledges the likelihood of a specific behavioral phenotype. However, this close association between physical and behavioral phenotypes can lead to bias toward biological models of the determinants of behavior,

with psychological and social influences being overlooked (O'Brien & Yule, 1995).

The laughing and smiling behavior that was included in the category Behavioral Uniqueness of the consensus was proposed by Summers, Allison, Lynch, and Sandler (1995) to be considered as a pathognomic sign. However, although the behavior is reported to be one of the most salient features, there is disagreement as to whether it is inappropriate given the context or environment (Angelman, 1965; Clarke, & Marston, 2000; Clayton-Smith, 1992, 1993; Fridman, Santos, Ferrari, & Koiffmann, 2000; Fung, Yu, Cheong, Smith, & Trent, 1998; Oliver, Demetriades, & Hall, 2002). It is this question of inappropriateness as opposed to the excessive nature of the behavior that highlights the comparative emphasis placed on biological events. In earlier reports, investigators predominantly conceptualized the cause of the behavior as stemming from a neurological impairment. However, although in the original paper, Angelman (1965) suggested that this behavior was "often in an almost convulsive state" (p. 685) and that laughter often proceeded and/or followed the child's seizures, he also stated that it was easily provoked, thus acknowledging the role of the environment. Dooley, Berg, Pakula, and MacGregor (1981) proposed that these children were not "happy in the traditional sense" (p. 623), and Williams and Frias (1982) suggested that the laughter seemed to be independent of happy or sad environments and that an absence of any emotional association with the laughter suggested that there was an abnormality at the brain stem level. The laughter was considered to be forced laughter and unrelated to appropriate social and environmental contexts.

Recently, researchers have reported that the laughing and smiling behavior is provoked, although the nature of the provocation is unclear (Clayton-Smith, 1992, 1993; Kuroki, Matsui, Yamamoto, & Ieshima, 1980; Willems, Dijkstra, Brouwer, & Smit, 1987; Williams & Frias, 1982). Yamada and Volpe (1990) reported smiling and laughing on face-to-face contact, and Kibel and Burness (1973) stated that the paroxysms of laughter occurred on any social contact, whether pleasant or unpleasant. Oliver et al. (2002) reported that smiling and laughing were influenced by social and environmental events. Other investigators reported that the behavior occurred when the child was brought into a new situation (Buntinx et al., 1995) or that it followed vomiting (Magenis, Brown, Lacey, Budden, & LaFrach, 1987).

To summarize, although the above reports evidence disagreement as to the underlying causes of the laughing and smiling behavior in Angelman syndrome, more recently researchers have begun to acknowledge the role of the social environment. However, to date there is only one experimental study that has investigated the laughing and smiling behavior of people with Angelman syndrome (Oliver et al., 2002); other reports are case descriptions. Oliver et al. examined the environmental influences on smiling and laughing behavior of individuals with Angelman syndrome by exposing individuals to several conditions in which social variables were manipulated. They found that laughing and smiling was minimal in the absence of social interaction but heightened during a social interaction condition. However, because these authors compared laughing and smiling in Angelman syndrome when the individuals were either alone, in proximity to an adult, and when an adult was socially interacting with them, the components of social interaction that the behavior is dependent upon could not be ascertained. In addition, the sample only consisted of three children with Angelman syndrome whose genetic subtype was not identified.

Given the debate regarding the role of social factors in provoking smiling and laughter and the paucity of empirical approaches, in the current study our aim was to further examine the influences on laughing and smiling behavior in children with Angelman syndrome. We adopted observational methods while systematically manipulating aspects of social interaction. The laughing and smiling behavior of each child was observed while he or she was repeatedly exposed to conditions in which parameters of social interaction were manipulated. Whereas Oliver et al. (2002) observed laughing and smiling behavior in one condition of social interaction compared to an alone and a proximity-only condition, here we compared laughing and smiling across two conditions of social interaction and a proximity-only condition to determine whether the laughing and smiling behavior is related to social interaction and discern the components of social interaction that elicit the highest levels of the behavior. We hypothesized that if the laughing and smiling of children with Angelman syndrome can be heightened by social interaction, then the duration of laughing and smiling will be higher in a social

interaction condition involving adult speech, smiling, laughing, and eye contact compared to restricted social interaction and a control adult proximity-only condition.

Method

Participants

We recruited 13 children through the Angelman Syndrome Support Education and Research Trust in the United Kingdom. These children were selected from a large database, and they lived closest to the research base. Once approval from the University Ethics Committee had been received, we sent parents an information sheet describing the purpose of the research and a consent form. Cytogenetic testing, from parents' report, confirmed a de novo maternal deletion of chromosome 15q11–13 in all children. Of the 13 children recruited, 11 completed the study. Table 1 shows data on age, gender, and mean age equivalent for the Adaptive Behavior Composite (calculated from the Vineland Adaptive Behavior Scale—Sparrow, Balla, & Cicchetti, 1984) for all 11 children. The Vineland was completed by each child's teacher on the day that the observations were conducted.

Participants were 7 boys and 4 girls; their mean age was 6.81 years (range = 4 to 11). The mean of mean age equivalent for the Adaptive Behavior Composite of the children was 12.82 months (range = 7 to 22). The Adaptive Behavior Composite scores for 9 children corresponded to a severe mental retardation level and for 2 children, a profound level. All of the children lived at home and attended local schools for children with mental retardation. Observations were not completed for 2 children who were initially included in the study. One child chose not to participate in the observations (age 16, male, mean age equivalent for the Adaptive Behavior Composite = 24 months), and for the other child, we were unable to get a full 30 seconds of unconcealed footage of her face in each condition because she persistently walked towards the camera operator and looked at the camera (age 13, female, mean age equivalent for the Adaptive Behavior Composite = 21 months).

Table 1. Demographics of Participants (P)

P	G[a]	CA[b]	AB[c]	Amb.[d]
1	F	4	8	Partly
2	F	4	12	Partly
3	M	4	12	Partly
4	M	6	7	Partly
5	M	6	12	Partly
6	M	6	13	Partly
7	M	8	11	Partly
8	F	8	17	Partly
9	F	9	11	Partly
10	M	9	16	Fully
11	M	11	22	Fully

[a]Gender. [b]In years. [c]Adaptive behavior. Mean age equivalent derived from the Vineland Adaptive Behavior Scales; composite in months. [d]Ambulatory. Partly = able to walk with support, Fully = able to walk unaided.

Procedure

To evaluate whether laughing and smiling behavior is related to specific social events, each child was observed (videotaped) while repeatedly exposed to three conditions. In the proximity-only condition, the teacher, who maintained a neutral facial expression, sat adjacent to the child and did not look at, talk to, or touch him or her. In the restricted social interaction condition, the teacher sat adjacent to the child while talking as she would when normally interacting with the child, but she did not look at the child and maintained a neutral facial expression. In the social interaction condition, the teacher sat adjacent to the child while talking, giving physical contact, smiling, laughing, and maintaining eye contact as they typically would. Each experimental session was comprised of two series of reversal designs (Series 1 and 2).

The conditions in which the parameters of social interaction were manipulated (restricted social interaction and social interaction) were alternated between repeated presentations of the control condition (proximity-only). Thus, each series consisted of five proximity-only (A) conditions, two restricted social interaction (B), and two social interaction (C) conditions. The order of conditions in Series 1 was ABACABACA and in Series 2, ACABACABA. Each series lasted approximately 4.5 minutes, with a break of approximately 5 minutes between series.

The rationale for the length and number of conditions was based on discussions with parents, caregivers, and teachers, who stated that their children would find it difficult to sit down for several minutes at a time, particularly as they would have

times when they were not engaged in an activity (control condition) or with their teacher, along with a high incidence of hyperactivity and short attention span being reported in individuals with Angelman syndrome (Summers, Allison, Slynch, & Sandler, 1995; Walz & Benson, 2002; Zori, Hendrickson, Woolven, Whidden, Gray, & Williams, 1992). We therefore decided that each condition would last 30 seconds to enable two social interaction and two restricted social interaction conditions to be presented in each series, and for the control condition to be presented between each interaction condition.

Each session was conducted with the child, his or her teacher, and the camera operator (who did not interact with the child) present in the room. The video recordings of each child were coded, and data on response duration were recorded on data-collection software Obswin (Martin, Oliver, & Hall, 1999).

To evaluate the integrity of the independent variables, we recorded the behavior of the teacher. The mean percentage of 1-s intervals for each of the teacher behaviors (smile/laugh, talk, eye contact) was 0% in the proximity-only condition. In the restricted social interaction condition, the mean percentages of 1-s intervals that the teacher smiled/laughed, talked, touched, or made eye contact were 1.22%, 96.49%, 1.14%, 0.24%, respectively, and in the social interaction condition, the mean percentages of 1-s intervals that the teacher smiled/laughed, talked, touched, or made eye contact were 40.25%, 95.27%, 48.42%, and 33.88%, respectively. Thus, the integrity of the independent variables was maintained.

Measurement and Interobserver Agreement

One child behavior, 3 adult, and 1 adult/child behavior were recorded. *Child smiling/laughing* and *adult smiling/laughing* was defined as: any horizontal stretching of the lips, upturning of the corners of the mouth followed by parting of the lips and viewing of the teeth, or any short burst of inarticulate voiced noises accompanied by an opening of the mouth, upturned corners of the lips, displayed teeth, half or completely shut eyes, and raised cheeks. *Adult touch* was defined as any physical contact made by the teacher towards the child as a result of moving a part of their body towards the child's body. *Adult talk* was defined as any verbal action and *eye contact* as any mutual visual contact directed toward the eyes between the child and the teacher.

Interobserver agreement was assessed by having a second observer simultaneously but independently collect data during 50% of each child's session. Kappa indices were calculated based on a 1-s interval-by-interval comparison of observer records. The Kappa coefficient for child smiling/laughing was .80. For adult smiling/laughing, adult touch, and adult talk, the Kappa indices were .85, .88, and .90, respectively, and for eye contact, .75. All indices were greater than .6, suggesting that interobserver reliability was good (Landis & Koch, 1977).

Data Analysis

Our first approach to data analysis consisted of examining the frequency, burst duration (duration of discrete episodes of laughter), and total duration of child behaviors across all trials of all conditions to examine between participant variability. In the second approach, we examined the frequency per minute, mean burst duration, and total duration of child smiling/laughing in each condition: proximity-only, restricted social interaction, and social interaction to evaluate the hypothesis. A within-subject ANOVA was adopted because the assumptions about the nature of the data required were fulfilled. The data were normally distributed as indicated by the results of the Kolmogorov-Smirnov test. Mauchly's test of sphericity indicated that the variances of differences were not significantly different for total duration of smiling/laughing. Because the variances of differences were significantly different for frequency and burst duration, we employed the Greenhouse-Geisser correction. We used a Bonferroni post hoc test to identify the conditions in which child smiling/laughing differed significantly; Clark-Carter (1997) suggested that this test is most appropriate when comparisons have been planned, and Field (2000) stated that it is the most robust in terms of power and control of type 1 error. To avoid the problem of inflated error rates because of repeated tests, we report only results significant at the Bonferroni-corrected .0167 adjusted significant level. In the third approach we used Pearson's product moment correlation to examine the relationship between child smiling/laughing, adult smiling/laughing, adult touch, adult talk, eye contact, age, and Vineland scores; the assumptions were fulfilled.

Results

Participant variability; the mean, *SD*, and range for the total percentage of time; mean burst

duration; and frequency per minute of laughing and smiling was calculated for each participant across trials of conditions (see Table 2).

Table 2 shows that the percentage of time spent laughing and smiling varied considerably between participants. Participant 11 did not smile or laugh in any of the conditions, and the mean percentage of time Participant 7 laughed and smiled across all conditions was 38.34. Mean burst duration varied from 0 to 9.71 s and frequency per min varied from 0 to 2.78.

To examine each child's data individually, we calculated the percentage of time each participant engaged in laughing and smiling in each condition (see Figure 1). This figure illustrates that the percentage of time each participant engaged in laughing and smiling appears to vary systematically across conditions. To evaluate the effect of social interaction at group level, we derived the mean (± one standard error) percentage of time, mean burst duration, and frequency per minute of laughing and smiling in each of the three conditions for all participants (see Figure 2).

The mean percentage of time, mean frequency per minute, and mean of mean burst durations of laughing and smiling for all participants were 8.16, 1.14, and 1.88, respectively (*SD*s = 7.47, 0.89, and 1.43, respectively) in the proximity-only condition, 20.61, 1.98, and 4.16, respectively (*SD*s = 18.27, 1.33, and 3.54, respectively) in the restricted social interaction condition, and 45.22, 2.80, and 9.66, respectively (*SD*s = 27.87, 1.57, and 7.83, respectively) in the social interaction condition.

To test the hypothesis that the laughing and smiling of children with Angelman syndrome is related to social interaction, we used a within-participants ANOVA to compare the percentage of time spent laughing and smiling across the proximity-only, restricted social interaction, and social interaction conditions. There was a significant difference between the three conditions, $F(2, 20) = 18.13$, $p < .001$. A Bonferroni-corrected post hoc analysis indicated a significant difference in percentage of time spent laughing and smiling between the proximity-only and social interaction condition, between the restricted social interaction and social interaction condition, but no difference between the proximity-only and restricted social interaction condition. Thus, the percentage of time engaged in laughing and smiling was higher in a condition involving adult speech, touch, smiling, laughing, and eye contact compared to a condition involving adult speech only and a control condition.

To examine the potential effect of social interaction on burst duration and frequency, as opposed to just percentage of the time, secondary analyses were undertaken. A within-participants ANOVA was used to compare the mean duration of each burst of laughing and smiling across the proximity-only, restricted social interaction, and social interaction conditions. There was a significant difference between the three conditions, *F*(2,

Table 2. Mean, *SD*s, and Range for Total Percentage of Time, Mean Burst Duration, and Frequency per Minute of Laughing and Smiling by Participant (P) Across all Conditions

	Total % of time			Mean burst duration[a]			
P	Mean	*SD*	Range	Mean	*SD*	Range	Frequency[b]
1	16.25	21.74	0–66.67	2.70	3.16	3–10	1.77
2	8.71	12.76	0–42.42	1.45	1.78	2–4.67	1.41
3	25.21	27.51	0–87.1	5.18	6.46	2–27	2.41
4	2.34	5.71	0–22.58	0.72	1.78	1–7	.42
5	18.90	18.95	0–65.63	4.10	4.36	2–13	1.89
6	20.10	30.83	0–84.85	4.67	7.37	1–28	1.50
7	38.34	29.60	0–76.67	5.62	4.91	2–23	2.78
8	33.91	37.22	0–93.94	9.71	12.22	2–31	2.25
9	20.29	23.73	0–58.05	5.87	5.71	4–18	1.98
10	25.24	31.57	0–93.33	5.24	7.80	1–19	2.02
11	0	0	0	N/A[c]	N/A	N/A	0

[a]In seconds. [b]Per minute. [c]Not available.

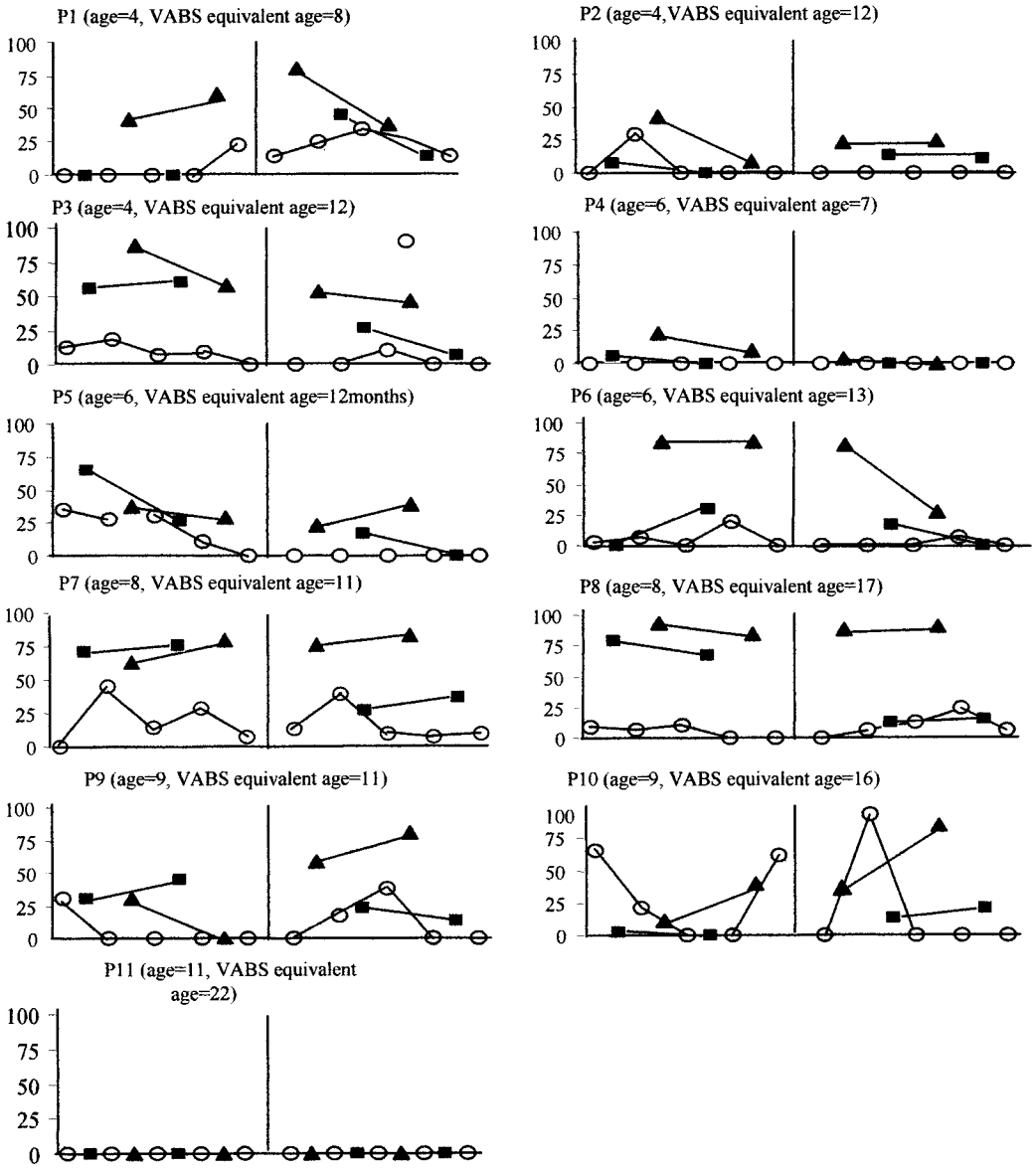

Figure 1. Percentage of time engaged in laughing and smiling by participants across conditions. Age in years, mean age equivalent for the Adaptive Behavior Composite, in months. Left plot is Series 1; right plot, Series 2. Triangle = social interaction, square = restricted social interaction, circle = proximity-only.

20) = 10.31, $p < .01$. A Bonferroni-corrected post hoc analysis indicated a significant difference between the mean burst duration in the proximity-only and social interaction condition, between the restricted social interaction and social interaction condition, but no difference between the proximity-only and restricted social interaction condition. Thus, the mean burst duration of laughing and smiling is longer in the social interaction condition compared to both the restricted social interaction and the control adult proximity condition.

A within-participants ANOVA was used to compare the frequency per minute of laughing and smiling behavior across the proximity-only, restricted social interaction, and social interaction conditions. There was a significant difference between the three conditions, $F(2, 20) = 9.75$, $p < .01$. A Bonferroni corrected post hoc analysis indicated a significant difference between the prox-

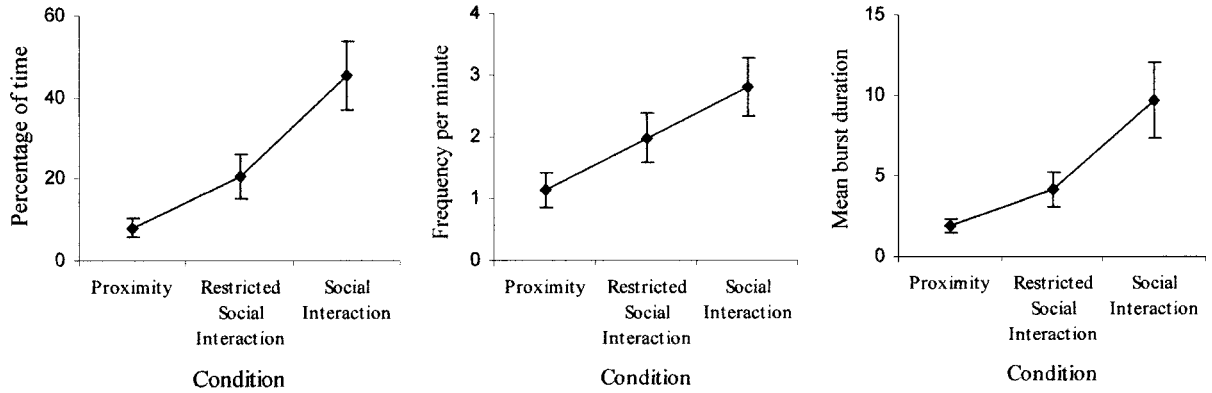

Figure 2. Mean (± one SE) percentage of time (left panel), frequency per minute (middle panel), and mean burst duration (right panel) of laughing and smiling by conditions.

imity-only and social interaction condition and between the social interaction and restricted social interaction condition but no difference between the restricted social interaction and proximity-only condition. Thus, the frequency of laughing and smiling was higher in the social interaction condition compared to both the restricted social interaction and control adult proximity conditions.

We used Pearson's product moment correlation coefficients to examine the association among age, Vineland Behavior Scale scores, adult laughing/smiling, talking, touching, eye contact, and child laughing/smiling within each of the social interaction and restricted social interaction conditions. In the social interaction condition, there was a significant correlation between adult laugh/smile and eye contact, $r(10) = .77$, $p < .01$, child laugh/smile and eye contact, $r(10) = .62$, $p < .05$, adult laugh/smile and child laugh/smile, $r(10) = .71$, $p < .05$, and adult laugh/smile and adult touch, $r(10) = .83$, $p < 01$. Thus, within the social interaction condition, the more the children laughed/smiled, the more the adults laughed/smiled and the more eye contact was maintained.

In addition, the more the adults laughed/smiled and the more the adults touched the children, the more eye contact was maintained.

Discussion

In this study the laughing and smiling behaviors of 11 children with Angelman syndrome were shown to vary when parameters of social interaction were systematically manipulated. The sample was comprised of only children who lived at home, had a documented deletion of chromosome 15q11-q13, and were observed at school with their usual teacher. The integrity of the manipulation of the independent variables was maintained as evidenced by observation of teacher behaviors in each condition, and interobserver reliability for child and adult behaviors was robust. The laughing and smiling behavior was found to be heightened in a condition involving adult speech, touch, smiling, laughing, and eye contact compared to a condition involving adult speech-only and a control proximity-only condition.

Our first hypothesis was that the duration of laughing and smiling would be higher in a social interaction condition. This was confirmed by comparing the percentage of time spent laughing and smiling across the three conditions (social interaction, restricted social interaction, and proximity-only). There was a significant difference in percentage of time spent laughing and smiling between the social interaction condition and the other two conditions, confirming that laughing and smiling behavior in children with Angelman syndrome is heightened by features of social interaction. This finding is consistent with results found by previous investigators who described and acknowledged the role of the social environment (Clayton-Smith, 1992; Kuroki et al., 1980; Willems et al., 1987; Williams & Frias, 1990) and Oliver et al.'s experimental demonstration (2002) that laughing and smiling was heightened in the presence of social interaction. However, unlike Oliver et al. (2002), we carefully controlled the parameters of social interaction, and all 11 children had a documented deletion of chromosome 15q11-q13. Homogeneity of genetic cause is important in behavioral phenotype research because

researchers have demonstrated that difference in within-syndrome phenotype may be dependent on genetic variability (e.g., Boer et al., 2002). Future researchers might explore this possibility in Angelman syndrome.

The results of the secondary analyses reveal the nature of the parameters of laughing and smiling behavior in Angelman syndrome when it is heightened by social interaction. The frequency and burst duration of laughing and smiling were compared across the three conditions, and a significant difference was shown between the social interaction condition and the restricted social interaction and proximity-only conditions.

The experimental control of the present study might have been improved if each parameter of social interaction was manipulated separately as opposed to several parameters being manipulated together, but this proved too difficult for the teachers who participated in the interactions. Although the integrity of the adult behaviors across the three conditions was maintained, it was difficult to control adult behaviors within each condition. The frequency and manner in which adults engaged in speaking, touching, laughing, smiling, and maintaining eye contact varied across conditions and across adults. However, experimental control of these behaviors is problematic because some behaviors become aversive if maintained for too long (e.g., eye contact: Arnold, Semple, Beale, & Flinn, 2000). To further examine the impact of the variability of adult behaviors on child behaviors, we conducted secondary analyses. A significant association was found between child laughing and smiling and adult laughing and smiling, child laughing and smiling and eye contact, adult laughing and smiling and adult touch, and adult laughing and smiling and eye contact. One interpretation of these findings is that adult behaviors elicited child laughing and smiling but given the problem of experimental control, it is possible that child behaviors elicited adult behaviors. These strong correlations indicate the need for tight experimental control and may also suggest that research is warranted in which the effect of child smiling and laughing on adult behavior is examined.

Although in the present study laughing and smiling behavior in children with Angelman syndrome was shown to be heightened by features of social interaction, we did not address methodological issues related to the brief length of each condition and the brief number of conditions. Because each condition only lasted for 30 seconds, one could argue that individuals laughed and smiled when a change in context occurred (when a new condition began) rather than or as well as being evoked by social contact. In addition, as the conditions were brief, laughing and smiling may have carried over from one condition to the next. However, if laughing and smiling occurred as a result of a transition, we would predict that the behavior would have occurred each time a session ended and the next one began. Similarly, if laughing and smiling carried over from one session to the next, the differences in the behavior between the social interaction conditions and the control conditions would have been reduced, because the social interaction conditions were alternated between repeated presentations of the control conditions. Therefore, although the above methodological issues need to be acknowledged, they do not account for the significant difference found between percentage of time spent laughing and smiling between the social interaction condition and the other two conditions.

Although the findings support the notion that the laughing and smiling behavior in Angelman syndrome is heightened by social interaction, it cannot be claimed that the behavior is associated with positive emotion because the topographical forms of the laughter and smiles were not examined. Further, there was no independent or self-report appraisal of emotion. Ekman, Friesen, and Ancoli (1980) reported the smile to be the Duchenne smile that occurs during a positive emotive display, with other smiles occurring during fear, disgust, or in deception. Researchers should explore whether the smiles and laughter observed in individuals with Angelman syndrome are Duchenne smiles and, therefore, potentially associated with positive emotion.

Similarly one cannot claim that that the laughing and smiling behavior in Angelman syndrome is only evoked by social interaction. The behavior may also be brought about by other environmental events, such as venipuncture (Clayton-Smith, 1992; Dooley, 1981; Kibel, Burness, 1973), which may explain the continued disparity in the descriptions of the behavior. Researchers should explore whether the laughing and smiling behavior observed is evoked by other environmental events through discussions with the individual's family and teachers.

Although the laughing and smiling behavior varied systematically across conditions, the behav-

ior also varied considerably across children. One child did not laugh and smile in any of the conditions, whereas another child spent nearly 40% of the time smiling and laughing. This individual variability may threaten the validity of the conclusions made. However, due to the inferential statistics, we argue that the individual variability does not undermine the differences found between conditions. Conversely, when considered alongside the small sample and brief conditions, we could argue that these issues could strengthen the findings.

One explanation for this variability is that although the integrity of the adult behaviors across the three conditions was maintained, the manner and frequency in which adults engaged in behaviors varied across conditions and adults. Alternatively, laughing and smiling may vary across individuals with Angelman syndrome; however, there is a lack of experimental studies that have examined such variability. The majority of investigators were concerned with either the excessive nature or the inappropriateness of the behavior in people with Angelman syndrome compared to individuals without Angelman syndrome. However, Buntinx et al. (1995) concluded from case descriptions that older individuals with Angelman syndrome appear to have less bursts of laughter. Although in the present study a relationship was not found between age and laughing and smiling, further examination showed that the child who did not laugh or smile in any of the conditions, and the 2 children for whom observations were not completed (due to 1 child deciding not to participate and the other child being distracted by the camera operator and camera), were the 3 oldest children in our sample. Research is needed to determine the variability in laughing and smiling across a wider age range of individuals with Angelman syndrome because findings may lead to further insight into the contribution of psychological, social, and developmental factors towards behavior.

These findings between participant variability and the effect of the environment on a phenotypic behavior confirm the importance of examining the interaction between genetic and environmental influences on behavior. The concept that a behavioral phenotype can be affected by environmental situations is not innovative. For example, the behavioral phenotype for Prader-Willi syndrome consists of overeating and food-seeking behaviors. Although these behaviors are argued to stem from an impaired satiety response or an abnormality of the brain in sending out signals of being full (Dykens, Hodapp, & Finucane, 2000), individuals with this syndrome have been shown to benefit from behavioral approaches (Dykens & Cassidy, 1996). The theoretical implication is that purely biological models of purported phenotypic behavior may be inadequate, highlighting the need for more sophisticated models that incorporate psychological, social, and biological determinants of behavior. In addition, there is clearly a need for a developmental perspective on phenotypic behaviors.

These findings also raise the issue of whether a behavior that is observed in a genetic syndrome is better considered as secondary to other factors. For example, the genetic disorder that causes Angelman syndrome may not directly increase an individual's likelihood of laughing and smiling but, instead, may lower the threshold at which individuals find social interaction reinforcing, which then leads to positive affect that elicits laughing and smiling behavior. Research is needed to examine the laughing and smiling behaviors in a control group of individuals with mixed or heterogeneous mental retardation matched to participants with Angelman syndrome in age, gender, and intellectual disability, as recommended by Hodapp and Dykens (2001). Because our purpose in the present study was solely limited to establishing whether the laughing and smiling behavior of children with Angelman syndrome is elicited by features of social interaction, we did not use a control group.

Overall, the results of this study provide evidence that the laughing and smiling behavior of individuals with Angelman syndrome can be heightened by social interaction. The implications of these findings are that professionals and families should be provided with more accurate information regarding this behavior in people with Angelman syndrome. Our results also suggest that there is a need for additional research to examine the relationship between social interaction and laughing and smiling behavior in Angelman syndrome. For example, if the laughing and smiling behavior is shown to be associated with positive emotion, this may suggest that social interaction is a naturally occurring highly potent reinforcer for individuals with Angelman syndrome. More effective interventions could then be developed based on the reinforcing properties of social interaction or specific facets of social interaction.

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Errata

In the May 2006 issue, errors were inadvertently made in "Collaboration in Referential Communication: Comparison of Youth With Down Syndrome or Fragile X Syndrome" by L. Abbeduto et al.

On p. 172, in the second paragraph of the *Participants* section, the final sentence should read "The Down syndrome and fragile X syndrome groups did not differ in nonverbal IQ according to the Stanford-Binet subtests, $t(27.95) = .61$, $p = .55$, or chronological age (CA), $t(41) = .59$, $p = .56$." A comma was used in the original $t(27,95)$, which is incorrect.

Also on p. 172, in the last full paragraph in the left column, the N for the chi-square should be 43, not 68. The sentence should read "Across the Down and fragile X syndrome groups, 60% of participants had mothers (or females guardians) with a college degree or higher, with no difference across syndromes in this regard, $\chi^2(2, N = 43) = .15$, $p = .70$."

On p. 176, in the last line of the right column, the symbol is incorrect. It should be an eta. The sentence should read. "The effect of group was significant, Wilks' Lambda, $F(6, 110) = 6.20$, $p \leq .005$, and a partial η of .25."

