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Experimental functional analysis of aggression in children with Angelman Syndrome

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ABSTRACT

Background: Kinship theory suggests that genomic imprinting could account for phenotypic behaviors that increase (in the case of Angelman syndrome) or decrease (for Prader-Willi syndrome) the drive to access social resources (adult contact) depending on the imprinting parent-of-origin. Difficult to manage behaviors, such as aggression that are common in Angelman syndrome, could serve the function of increasing social interaction. We hypothesise that the commonly reported aggressive behavior in children with Angelman syndrome will be attention maintained.

Methods: Experimental functional analysis was carried out with twelve children with Angelman Syndrome caused by either a deletion (n=10) or uniparental disomy (n=2).
The relative increase and decrease of aggressive behaviors was observed in response to experimentally manipulated levels of adult attention and demand. Laughing and smiling, crying and frowning, and physical initiation with adult were also measured.

Results: Aggression was seen in ten of the twelve children. One child evidenced a pattern of aggression across conditions consistent with maintenance by attention, three children showed higher levels of aggression during social interaction and two children showed escape motivated aggression.

Discussion: With the exception of one child the results did not confirm the hypothesis. However, the pattern of increased aggression in the high social contact condition combined with evidence of positive affect during this condition suggests aggression may serve to both maintain and initiate social contact and this interpretation is consistent with previous research. The negative results may also have been influenced by the age of the children and the low levels of observed aggression.

INTRODUCTION

Angelman Syndrome (AS) is caused by loss of genetic information at 15q11-13 on the maternally inherited chromosome, whilst loss of genetic information from the same site on the paternally inherited chromosome causes Prader-Willi Syndrome (Kaplan, Wharton, Elias, Mandell, Donion & Latt, 1987; Magenis, Brown, Lacy, Budden, & LaFrach, 1987; Knoll, Nicholls, Magenis, Graham, Lalande & Latt, 1989; Knoll et al., 1989; Cassidy, Dykens, & Williams, 2000). The observation that the same genetic cause could give rise to different phenotypes led to Angelman and Prader-Willi Syndromes being the first syndromes to be identified as associated with genomic imprinting (Knoll et al, 1989). Genomic imprinting refers to a parent-of-origin-specific process of gene inheritance whereby either the paternally or maternally inherited allele is rendered inactive (Reik & Walter, 2001). Thus, the same gene is expressed differently depending on parent of origin (Clayton-Smith, 1992).

Prevalence estimates of Angelman Syndrome range from approximately 1 in 12,000 to 1 in 25,000 live births, depending on the nature of the clinical diagnosis (Dykens, Hodapp & Finucane, 2000). Approximately 70% of cases of Angelman syndrome are due to a deletion on chromosome 15 at q11-13; 2-7% are caused by paternal unipaternal disomy (UPD); 2-7% due to an imprinting mutation; and 5-15% have a mutation in the UBE3A gene (Jiang, Lev-Lehman, Bressler, Tsai & Beaudet, 1999). Between 5-10% of people who show the clinical features of Angelman syndrome have no demonstrable cytogenetic or molecular abnormality of chromosome 15q11-13 (Clayton-Smith & Laan, 2003; Laan, Halley, den Boer, Hennekam, Renier & Brouwer, 1998; Lossie, Whitney, Amidon, Dong, Chen, Theriaque, Hutson, Nicholls, Zori, Williams & Driscoll, 2001). The syndrome is associated with specific physical characteristics, developmental delay and distinctive cognitive and behavioral phenotypes (Williams, Beaudet, Clayton-Smith, Knoll, Kyllerman, Laan, Magenis, Moncla, Schinzel, Summers & Wagstaff, 2006; Horsler & Oliver, 2006a; Moss, Oliver, Arron, Burbidge & Berg, 2009). One of the most salient behavioral features of Angelman syndrome is the presence of pro-social behaviors, such as excessive laughing and smiling, noted in a detailed review of 64 studies documenting 842 cases in the literature (Horsler and Oliver, 2006a).

Haig and Wharton (2003) developed kinship theory (or maternal investment theory) as an explanation for the phenomenon of genomic imprinting. They propose that maternal and paternal alleles can be expressed differently in order to favour continuation of maternal or

paternal chromosomes. Haig & Westoby (1989) proposed that imprinted genes might influence the level of parental investment for offspring. The paternal allele would increase the probability of maternal resources being allocated to an offspring, thus increasing the probability of its survival and ensuring the perpetuation of the paternal line. In contrast, the maternal allele would decrease the probability of maternal resources being allocated, thereby increasing the likelihood of further births and survival of the maternal line. This theory has received support from murine studies of placental function and neonate suckling (Isles, Davis & Wilkinson, 2006). Brown and Consedine (2004) suggest that the function of some of the behavioral characteristics of Angelman Syndrome is to increase the level of maternal investment. More specifically, they draw on emotion signalling theory to explain how pro-social behaviors such as smiling in Angelman syndrome would have the effect of evoking high levels of social contact, an indicator of parental investment.

Early opinion on the pro-social behaviors seen in Angelman syndrome suggested that the behaviors were contextually inappropriate and not influenced by the environment (e.g. Dooley, Berg, Pakula, & MacGregor, 1981; Williams and Frias, 1982). However, contemporary research has strengthened the argument for an environmental influence on these behaviors, with recent experimental studies demonstrating that smiling and laughing are influenced by the level and type of social interaction (Oliver, Demetriades & Hall, 2002; Horsler and Oliver, 2006b). However, Richman, Gernat & Teichman, (2006) did not replicate these findings, although they had a very small sample (n=2) and the children were considerably younger than those in the other studies (18 months and 42 months). More recently, Oliver, Horsler, Berg, Bellamy, Dick & Griffiths (2007) found that in comparison with a matched contrast group, children with Angelman syndrome laughed and smiled more frequently, showed a greater increase in laughing and smiling in response to social interaction, actively sought social interaction with adults prior to laughing and smiling and were more successful than the contrast group in maintaining adult attention. In combination, these studies suggest that social interaction with adults plays a significant role in influencing laughing and smiling in Angelman syndrome and that the laughing and smiling in turn influences adult behavior. In summary, it appears that children with Angelman syndrome have a propensity to seek out social interaction and, by implication, find interaction extremely pleasurable (see Oliver et al., 2007). This indicates that children with this syndrome have an unusually strong motivation to interact with adults, which provides some support to the maternal investment theory.

Anecdotal reports of aggressive behavior in individuals with AS date back to the 1970's (e.g. Moore and Jeavons (1973), Hersh, Bloom, Zimmerman, Dinno, Greenstein, Weisskopf & Reiss (1981), and Williams & Frias (1982)). Summers, Allison, Lynch & Sandler (1995) found that 10% of case reports reviewed (n=108) made reference to aggressive behaviors, consisting largely of biting, pinching or slapping, whereas a questionnaire survey found 100% of parents (n = 11) reported that their child engaged in low level aggressive behaviors (grabbing, hair pulling). Horsler & Oliver (2006a) also found that 15% of the 64 studies they reviewed made reference to aggressive behaviors, and Arron, Oliver, Hall, Sloneem, Forman & McClintock (2006) showed that aggression, but not self-injury, is significantly more common in Angelman syndrome than a contrast group. However, the causes of aggressive behavior in children with Angelman syndrome have yet to be studied empirically.

There is substantial empirical evidence within the operant learning literature to suggest that social attention has a strong reinforcing effect on self-injurious and aggressive behaviors (e.g. Iwata et al., 1982; Lovaas, Freitag, Gold & Kassorla, 1965; Oliver, Hall & Murphy, 2005). It has been suggested that aggression could be sensitive to positive reinforcement in the form of access to adult attention, peer attention and tangible items (Hawkins, Peterson, Schweid & Bijou, 1966; Patterson, Littman & Bricker, 1967) and Marcus, Vollmer, Swanson, Roane & Ringdahl (2001) provide empirical evidence that aggression in children with intellectual disabilities can be positively or negatively reinforced. Hanley, Iwata & McCord (2003) carried out a meta-analysis of the functional analysis of problem behavior, and found that an operant reinforcement process could be identified for 50 out of 52 studies that reported aggression in individuals with an intellectual disability . This behavior was attention-maintained in nine of the individuals reported across the studies. Given the possibility of operant reinforcement and the empirical evidence for the drive to seek adult attention in children with Angelman syndrome, it may be predicted that aggressive behavior in these children will be socially positively reinforced, thus demonstrating an interaction between one aspect of the behavioral phenotype of Angelman syndrome and operant learning. Models incorporating interaction between genetic disorders and environmental factors offer a plausible account of the development of behaviors such as aggression and self-injury in Angelman, Prader-Willi, Lesch-Nyhan, Rett, Down, Cornelia de Lange, and Smith-Magenis syndromes (see respectively: Oliver *et al.*, 2007; Woodcock, Oliver & Humphreys, 2009; Hall, Oliver & Murphy, 2001; Oliver, Murphy, Crayton & Corbett, 1993; Millichap, Oliver,

McQuillan, Kalsy, Hall & Lloyd, 2003; Arron et al, 2006; Taylor and Oliver, 2008). Social attention may act as a particularly effective reinforcer for aggressive behavior in children with Angelman syndrome due to their unusually strong motivation to interact with adults.

The empirical assessment of operant reinforcement of behaviors has its roots in the procedure first described Iwata, Dorsey, Slifer, Bauman, & Richman (1982). This methodology has since been refined and alternative techniques developed (Carr & Durand, 1985; Iwata, Dorsey, Slifer, Bauman, & Richman, 1994; Iwata, Duncan, Zarcone, Lerman, & Shore, 1994; Vollmer, Iwata, Duncan, & Lerman, 1993; Vollmer, Iwata, Zarcone, Smith, & Mazaleski, 1993; Neef & Iwata, 1994). Experimental functional analysis is now widely accepted as one of the best procedures for ascertaining if operant social reinforcement influences a given behavior.

There are two aims to this study. First, we employ experimental functional analysis to ascertain the frequency of aggressive behavior in children with Angelman syndrome, whether the behavior is influenced by environmental events and, if so, the mode of operant social reinforcement. We predict that children with Angelman syndrome will engage in aggressive behaviors and that these behaviors will occur at a higher rate when social contact is withheld, thus demonstrating reinforcement by social positive reinforcement. Secondly, we aim to extend previous research by examining the influence of social contact on smiling, laughing and social approach behaviors in children with Angelman Syndrome. In line with previous studies, we predict that the percentage of time spent engaging in pro-social behaviors will vary according to the degree of social contact available from an adult.

METHOD

Participants

Thirteen children diagnosed with Angelman syndrome aged between 4 and 16 years were recruited via families that had indicated they would like to be contacted, and were originally recruited through the Angelman Syndrome Support Education and Research Trust (ASSERT). Thirteen families demonstrated their interest in taking part by returning the consent form and questionnaire. Of the thirteen children recruited, twelve completed the study. Table 1 shows participant data for age, gender, mean age equivalent for the Adaptive Behavior Composite

(calculated from the Vineland Adaptive Behavior Scale; Sparrow, Balla & Cicchetti, 1984) and genetic cause.

++++++ Insert Table 1 here +++++++++

Seven participants were boys and five were girls, with the mean age of the sample being 8 years 11.5mths (range: 5yrs 0mths – 11yrs 9mths). The mean mental age equivalent for the Adaptive Behavior Composite of the children was 29 months (range = 21 – 48 months). The Adaptive Behavior Composite scores for all the children with deletions corresponded to a severe intellectual disability. The Adaptive Behavior Composite score for participant U1 corresponded to a moderate intellectual disability, and participant U2 was moderate-severe. All the children lived with their parents or full time carers at home and attended local schools for children with intellectual disabilities. For those children who lived with carers, their placement in the family had commenced from birth.

Setting

Participants were observed in an informal 'playroom' setting. The aim was to make the setting informal and comfortable. Two wall-mounted video cameras were positioned in the room, and efforts were made to make these as discrete as possible. A one-way mirror separated the playroom from the video room next door.

On arrival each family was briefed about the study and parents and carers were interviewed in order to collect the data required for The Vineland Adaptive Behavior Scale (Sparrow, Balla & Cicchetti, 1984). During this time, each child was given the opportunity to meet the researchers, explore the playroom and play with the toys provided. Following the completion of the Vineland, the parents and carers were invited into the video room next door, where they were able to watch their child during the experimental procedure through the one-way mirror. They were encouraged to request that the procedure be stopped at any time if they felt uncomfortable or were concerned. Four permanent wall-mounted toys remained throughout the procedure.

Procedure

Each child was observed (videotaped) whilst exposed to three conditions. The protocol was carried out by one researcher (RS), but a second researcher was present (although did not interact with the child) throughout the procedure.

The experimental functional analytic methodology used was similar to that devised by Carr & Durand (1985) for classroom-based assessment of disruptive behavior. The ***High Attention*** condition (A) acted as the control condition. In this condition, the researcher maintained a high level of verbal and physical attention (such as talking, singing, and clapping) and remained in close proximity to the child (within arms' reach). No demands were placed on the child in this condition. If the child displayed any aggressive behavior during this condition, the researcher ignored it and continued to maintain a high level of attention. In the ***Low Attention*** condition (B) the researcher did not speak to the child and maintained conversation with the second researcher. The first researcher began the condition in close proximity (within arms' reach) of the child and remained in this position even if the child chose (and was able) to move away. If the child displayed any aggressive behaviors during this time, the researcher gave them brief (approximately 5s) verbal and physical attention, and then continued to interact with the second researcher. For the ***High Demand*** condition (C), the child was requested to take part in a task that was challenging for them. This task was chosen based on parental report of ability and observations of the child during completion of the Vineland (Sparrow, Balla & Cicchetti, 1984). Initially the child was given a verbal prompt to complete the task, followed by the researcher modelling the task, and then a physical prompt (guided through task hand-over-hand), until the child was able to complete the task. This process was repeated until the end of the condition. If the child displayed any aggressive behavior during this condition, the researcher gave them verbal attention ("ok, you don't have to do that any more"), then the task was removed and the researcher turned away from the child. The researcher then remained turned away from the child for two consecutive 5s periods. If the child displayed aggressive behaviors during the second 5s interval, the researcher turned away for a further 5s from the time at which the behavior occurred.

Each condition lasted for 5 minutes, The *Low Attention* and *High Demand* conditions were alternated between repeated presentations of the control condition (*High Attention*). A series of conditions consisted of four *High Attention* (A) conditions, two *Low Attention* (B) and two *High Demand* (C) conditions (such as ABACABAC). The two experimental conditions (B

and C) were counterbalanced for each participant to control for order effects (for example, ACABABAC and ABABACAC).

The video recordings of each child were coded, and duration data on several behaviors were recorded using data collection software Obswin (Martin, Oliver & Hall, 1999).

Measurement and interobserver agreement

To evaluate the integrity of the independent variables the behavior of the researcher was recorded. Table 2 illustrates the percentage of time for the researcher behaviors (verbal attention, physical attention, verbal prompt, model prompt, and physical prompt). As can be seen from the data, the integrity of the independent variables was maintained.

+++++++++ Insert Table 2 here ++++++++++

25% of all the observations were independently coded by a second observer. Inter-observer agreement between the index and second observer were compared on a 10s interval-by-interval basis, with agreements and disagreements scored on occurrence and non-occurrence for each response category. In order to control for 'chance' levels of agreement, Cohen's Kappa was employed. The Kappa coefficients for child codes were as follows: smiling (.97); laughing (.96); frowning (.77); crying (.77); initiating physical contact with researcher 1 (.85); and aggression towards researcher 1 (.95). For researcher codes, verbal attention, physical attention, verbal prompt, model prompt and physical prompt, the Kappa indices were .98, .93, .88, .85, and .94 respectively. All indices were greater than .60 suggesting that interobserver reliability was good (Landis & Koch, 1977).

RESULTS

In order to consider the function of aggression, individual plots were produced. These are shown in Figure 1. The data in figure 1 shows that ten out of twelve participants demonstrated some form of aggressive behavior (hair pulling, spitting, biting, smacking, grabbing, pinching), although only four of these exhibited this behavior for more than 5% of the time during any one condition. Eight out of the ten participants who displayed aggressive behavior did so in the *high attention* condition (80%), three out of ten (30%) in *low attention*, and four

out of ten (40%) in *demand* condition. Participants D4, D8 and D9 exhibited aggressive behavior in more than one condition, with D4 being the only one to do so across all conditions. D4 was also the only participant whose pattern of aggressive behavior was in agreement with the hypothesis that children with Angelman syndrome engage in aggressive behavior in order to access social attention. D7 and U2, and, to a lesser extent, D8 evidenced a pattern of responding that is consistent with demand escape function to aggression. It is notable that three participants (D2, D8 and U2) showed aggression in both trials of the high attention condition. Participants D1 and U1 were the only participants who did not show any aggressive behavior.

++++++++++++++ Insert Figure 1 here ++++++++++++

To examine the occurrence of laughing/smiling and crying/frowning and physical initiation to researcher, individual plots were created to show the percentage of time spent engaged in these behaviors for each condition. These plots are shown in Figure 2.

++++++++++++ Insert Figure 2 here ++++++++++++++

Participants D4, D8 and D9 showed the highest levels of all social behaviors. D8 and D9 are striking in the similarity of their pattern of laughing/smiling and crying/frowning. Neither showed any distress during the procedure, but both engaged in laughing/smiling behaviors over 80% of the time during the *demand* condition, and over 25% of the time in *high attention*. Neither demonstrated any laughing/smiling behavior during *low attention*. Participant D4 followed a similar pattern, although with a lower percentage of time spent laughing/smiling during *demand*, and a higher percentage in both *high attention* and *low attention*. D8 and D9 engaged in physical initiation during both *high* and *low attention*, with D8 additionally showing minimal levels of this behavior during *demand*. D4 also engaged in physical initiation across all conditions, with the highest level occurring in *high attention*. The pattern of behavior demonstrated by participant D2 was comparable to that of D4, although with a lower percentage of time spent laughing/smiling across all conditions. D2 showed minimal levels of physical initiation, with this behavior only occurring during *high attention*.

Two participants (D1 and D7) did not engage in any smiling/laughing behavior at all. Crying/frowning was observed for both participants, with the vast majority of these behaviors occurring during the *demand* condition for D1, whereas for D7 they occurred across conditions. The minimal levels of physical contact displayed by these participants followed the same pattern as described for crying/frowning.

Out of the ten participants that did engage in laughing/smiling behaviors during the procedure, five spent the highest percentage of time laughing/smiling in the *high attention* condition and four in *demand*. Participant D10 engaged predominantly in laughing/smiling behaviors during *low attention*. Six participants exhibited some degree of crying/frowning behaviors.

From the results presented above it can clearly be seen that participants D2, D4, D8 and D9 are similar in their pattern of behaviors, with higher levels of smiling during conditions with higher levels of social contact. Participants D1 and D7 appear quite different from the other participants with AS caused by a deletion due to their low levels of social behaviors. It is also notable that the two participants with AS resulting from UPD present differently from those with deletions. For these participants, laughing/smiling occurred solely in *high attention* and only for a small percentage of time. In contrast, crying/frowning was observed across all conditions and for a greater percentage of time than laughing/smiling. Their levels of physical initiation were negligible.

DISCUSSION

The first aim of this study was to consider the occurrence and function of aggressive behaviors in children with Angelman syndrome. The second aim was to replicate previous findings that laughing and smiling behaviors in children with AS occur as a social response, and to widen the range of social behaviors studied to include physical initiation and crying and frowning. Experimental functional analysis was carried out under controlled conditions, based on the methodology devised by Carr & Durand (1985), and observational data were then coded reliably. The integrity of both the conditions and coding procedure was robust. This was the first empirical study into aggressive behavior in children with Angelman syndrome, and the methodology extends previous research into gene-environment interactions in genetic syndromes by employing more rigorous experimental techniques (c.f. Taylor &

Oliver, 2008; Hall et al., 2001; Moss, Oliver, Hall, Arron, Sloneem & Petty, 2005; Arron et al, 2006). The use of analogue methodology in this study enables the experimental manipulation of antecedents and consequences for behaviors, which increases the validity of results.

As predicted, the majority of participants did engage in aggressive behavior (10 out of 12), although the frequency of the behavior was low for some children. These rates of aggressive behavior are more comparable with those suggested by the questionnaire survey conducted by Summers et al. (1995) where 100% of parents reported aggressive behavior in their children rather than those suggested by the case reports they reviewed (10%). However, the small number and short observation period of this study mean reliable conclusions regarding the prevalence of these behaviors cannot be drawn.

Generally, aggressive behavior did show variability across conditions but the results were inconsistent. The model of functional analysis (e.g. Iwata et al., 1982/1994; Carr & Durand, 1985) would suggest that for one participant (D4) aggressive behavior was clearly attention maintained, and two participants (D7 and U2) were rewarded by the withdrawal of task demands. For three participants (D2, D8 and U2) there were higher levels of aggression when levels of social interaction were high. This pattern could suggest a social escape function but this seems unlikely given evidence of positive affect during this condition and well documented reports of enhanced sociability. It is possible that this pattern reflects motivation to maintain, as opposed to initiate, a social interaction. This interpretation would be consistent with the sequential data of Oliver et al., (2007) that shows the function of smiling is more likely to be maintenance of a social interaction as opposed to initiation. Anecdotally it is notable that some participants appeared to experience elevated mood when engaging in aggressive behaviors during *high attention*. Another possible explanation for the low levels of aggressive behavior seen in *low attention* is that the results are confounded by limitations of the experimental design. As the *low attention* condition always follows *high attention*, participants may satiate on attention and so the drive to seek attention in low attention is lessened (Murphy, McSweeney, Smith & McComas, 2003). This is interesting in itself because, in contrast with what we might expect, it suggests that children with AS can satiate on attention.

Laughing and smiling behaviors were found to vary across conditions, being more likely to occur during times of high attention. However, it was not found to covary with condition to the same extent found by previous research (Horsler & Oliver, 2006b). Crying and frowning was observed in six of the twelve participants. No laughing/smiling was reported for two children (D1 and D7). D1 was the youngest in the sample (5 years), although he had the highest VABS equivalent age of 41 months. D7 was one of the oldest participants at 10 years (VABS equivalent age was 24 months). Horsler & Oliver (2006b) also reported no laughing and smiling for one child, who was the oldest in their sample (chronological age = 11 years, VABS equivalent age = 22 months). Research has suggested that the clinical characteristics of Angelman syndrome are most pronounced between 2-16 years (Buntinx, Hennekam, Broumer, Stroink, Beuten, Mangelschots, & Fryns, 1995; Clarke & Marston, 2000). It is therefore possible that an interaction of age with behavior is being seen for these participants, with a reduction seen in laughing and smiling as children age. However, this does not provide an explanation for the lack of smiling and laughing behavior seen in participant D1.

Although there is only partial support for the predicted gene environment interaction it is notable that for a number of participants aggression was reliably evoked under a given environmental condition. This is important for two reasons. First, although the prevalence of aggression is raised in Angelman syndrome there is clearly the potential for an environmental influence on the behavior. Second, the influence of the environment on the aggressive behavior alludes to the potential for interventions based on operant reinforcement. Therefore, the results of this study extend the number of demonstrations of environmental influence in genetic syndromes in which problem behaviors are a part of the behavioral phenotype. Taylor & Oliver (2008) found that aggressive, disruptive and self-injurous behavior in children with Smith-Magenis syndrome clearly occurred as a reaction to environmental events, and more specifically to a reduction in adult attention. Hall et al. (2001) and Moss et al., (2005) also found an environmental effect for self-injurous behavior in children with Lesch-Nyhan and Cornelia de Lange syndromes respectively when using descriptive analysis of observational data. Oliver, Murphy, Crayton & Corbett (1993) and Arron et al. (2003) used analogue methodology to investigate self-injurious behavior in children with Rett and Cornelia de Lange syndromes respectively and identified clear operant functions of the behavior.

There are some limitations to this study. Participants were not selected because they were aggressive and the low level of aggression might have placed a constraint on the potential to

identify behavioral function. However, the study provided a useful insight into the frequency that aggressive behaviors occur in a general sample of children with Angelman syndrome. The procedure also would have benefited from further repetitions of each condition over a longer period of time in order to make it more robust. However, this was felt to place unnecessary demands on the participants. Finally, recent research into the effect of familiar versus unfamiliar adults when conducting analogue experiments with children with intellectual disabilities suggests that using the child's caregiver as the therapists rather than an unknown researcher can produce higher rates of problem behavior (English & Anderson, 2004). For the purposes of this preliminary study a comparison of the effect of familiar and unfamiliar therapists on functional analysis did not seem necessary, although future research in this area may benefit from comparisons.

Overall, the results of this study suggest that there is a need for further functional analytic studies to be carried out focussing on various different aspects of the behavioral phenotype of Angelman syndrome. The levels of and environmental influence over social behaviors seen in this study and that of Richman et al. (2006) in comparison with findings from other studies (e.g. Horsler & Oliver, 2006b) suggests that there may be variability in these behaviors relating to both chronological and developmental age. This would be particularly important in relation to the development of difficult behaviors as the identification of an age at which these behaviors are more likely to occur would have significant implications for early intervention and prevention. Further research into the function of aggressive behaviors in children with Angelman syndrome would be useful, given parental report of distress caused by these types of behavior. Future studies should focus on the difference in frequency and function of aggressive behaviors depending on the level of familiarity of the adult.

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Participant No	Gender	Age[1]	Adaptive Behavior Composite in months[2]	Genetic Cause
D1	Male	5yrs 0mths	41	Deletion
D2	Female	7yrs 9mths	27	Deletion
D3	Male	7yrs 9mths	29	Deletion
D4	Male	7yrs 11mths	29	Deletion
D5	Female	8yrs 6mths	31	Deletion
D6	Female	8yrs 9mths	27	Deletion
D7	Male	9yrs 7mths	24	Deletion
D8	Female	10yrs 11mths	22	Deletion
D9	Male	11yrs 3mths	21	Deletion
D10	Male	11yrs 9mths	23	Deletion
U1	Male	8yrs 7mths	48	UPD
U2	Female	9yrs 9mths	30	UPD

[1]Chronological age in years and months
[2]Mean age equivalent in months derived from the VABS

Table 1. Demographics of the participants

	High Attention	Low Attention	Demand
Verbal Attention	97.51[1]	3.55	11.39
Physical Attention	46.30	3.08	6.98
Verbal Prompt	0.00	0.00	2.67
Model Prompt	0.00	0.00	7.47
Physical Prompt	0.00	0.00	8.15

[1]Total percentage of time for each condition can total over 100% as behaviors were not mutually exclusive

Table 2. Percentage of time the researcher engaged in each behavior across conditions.

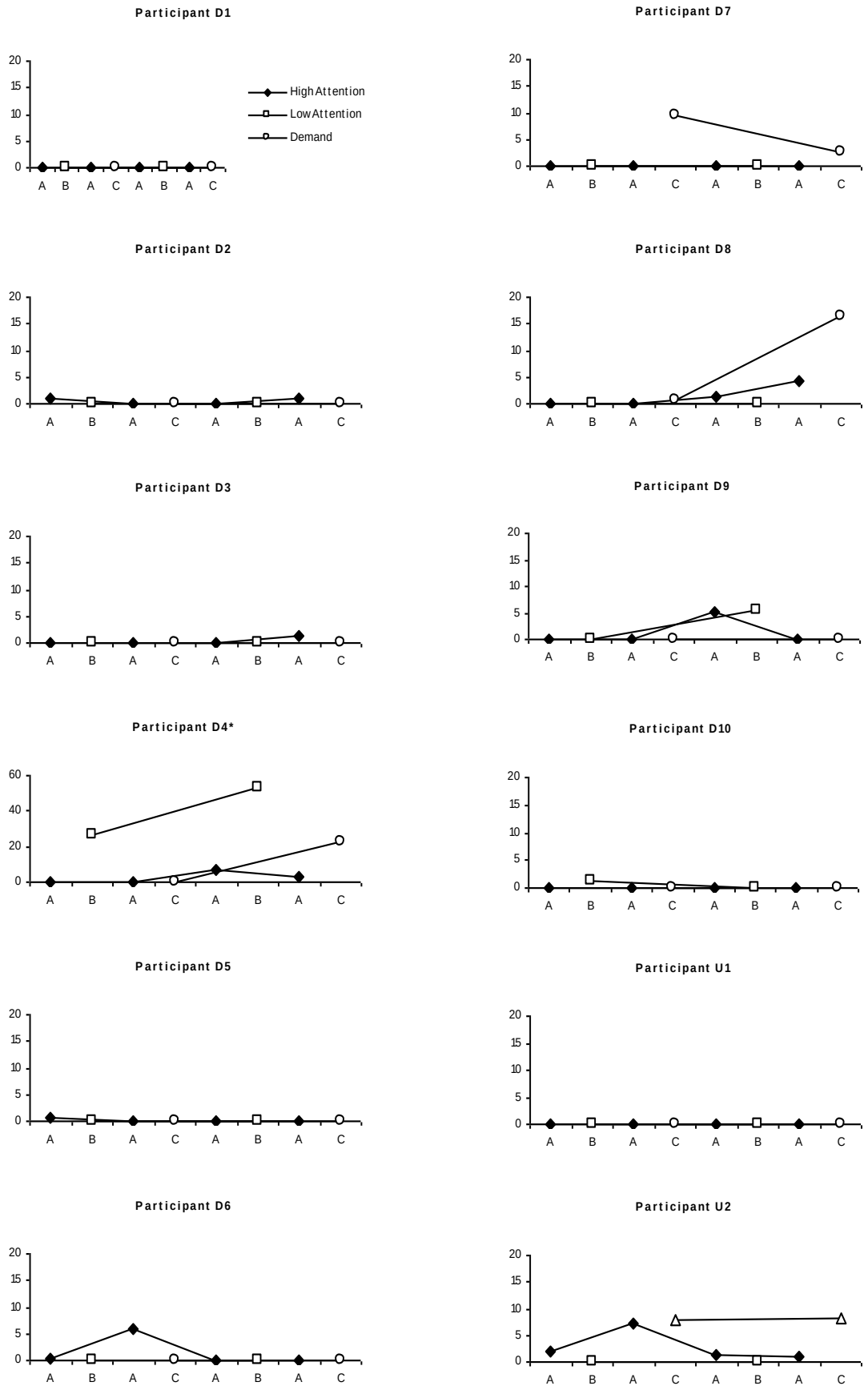

Figure 1: Percentage of time participants engaged in aggression across conditions

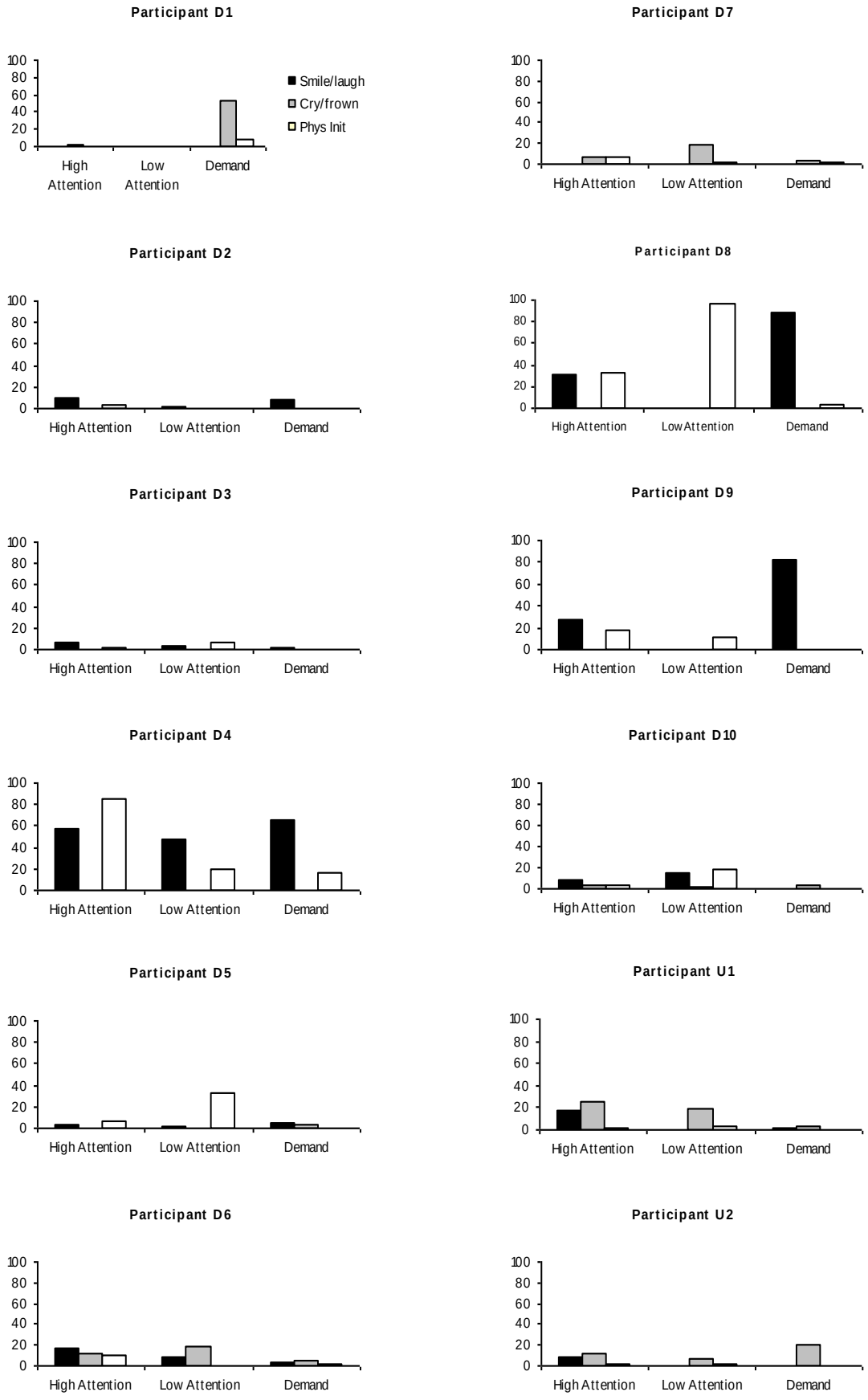

Figure 2: Percentage of time engaged in smiling/laughing, crying/frowning and physical initiation across conditions