USING FUNCTIONAL ANALYSIS TO TEST WHETHER IDIOSYNCRATIC AND CHALLENGING BEHAVIOR IN RETT SYNDROME IS SENSITIVE TO SOCIAL REINFORCEMENT CONTINGENCIES

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Abstract

Rett syndrome (RS) is a severe neurodevelopmental disability that results in cognitive impairment, loss of functional and communication skills, and an emergence of idiosyncratic behaviors. There is little behavioral research to date using functional analysis to examine the operant properties of behaviors exhibited by individuals with RS. The purpose of this study was to test whether functional analysis technology could be applied to a broader range of idiosyncratic behaviors observed in a clinical sample of six girls and women living with RS. Brief functional analyses were conducted to evaluate target behaviors including breath holding, vocalizations, vocal upsets, hand mouthing, and self-injury. Results indicate that four out of the six analyses conducted with the participants produced differentiated results. Further research is needed to evaluate how identifying idiosyncratic behaviors that are sensitive to reinforcement contingencies can inform treatment for this population in areas of pronounced deficits such as communication.

Key words: Rett syndrome, functional analysis, idiosyncratic behavior

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Rett syndrome (RS) is a severe neurodevelopment disorder almost exclusively affecting females (Kerr & Engerstrom, 2001). It is estimated that 1 in 10,000 female are affected by RS (Amir & Zoghbi, 2000). RS is considered one of the most common specific causes of severe cognitive impairment in females (Amir & Zoghbi, 2000; Skjeldal et al., 1997). In 85-90% of cases, the syndrome is caused by mutations in the X-linked gene encoding methyl CpG–binding protein (MeCP2) (Amir, Ignatia, den Veyver, et al., 1999; Amir & Zoghbi, 2000; Shahbazian & Zoghbi, 2001). Continued research is underway on variants of this observed mutation and how it affects the clinical phenotype of the syndrome (Shahbazian & Zoghbi, 2001).

Developmentally, RS is characterized by neurodevelopmental stagnation and regression starting between 6 and 18 months of age (Hagberg, 1993). Early gains in motor and communication skills deteriorate and deceleration of head growth becomes evident. The period of regression following stagnation of development between 1 and 4 years of age is associated with profound loss of skills in the domains of hand use, social contact and communication, and language (Engerstrom, 1992). Loss of hand skills, apraxia of gait, increased spasticity and scoliosis, as well as behavioral features such as stereotypic hand movements, breath holding, and hyperventilation characterize this regression period more specifically (Rett, 1966; Hagberg, 1993). Development tends to then plateau post-regression between 2 and 10 years old, and consequently results in profound cognitive impairment (Woodyatt & Ozanne, 1997). Development of other neurological impairments such as epileptic seizures also becomes apparent within this stage for individuals with RS (Hagberg, 1993). Communication and adaptive behavior are two areas where rapid changes in skill level and functioning are most prevalent.

The large amount of genetic research to date provides valuable information in regard to the etiology of RS. Although clinical accounts of the behavioral phenotype specific to RS preceded the genetic findings, there has been very little sustained research investigating behavioral features of the syndrome in a systematic way. In particular, there is almost no data evaluating behavioral issues in RS aimed at creating practical assessment strategies that could inform behavioral interventions for communication and behavior problems. The following sections will review the behavioral research specific to communication and behavioral issues associated with RS and explicate the rationale and purpose of the present study.

Communication and Behavioral Issues in RS

Research related to the RS behavioral phenotype looks to identify emotional and behavioral features that characterize the syndrome. Phenotypic work in general aims at comparing behavior across different syndromes to assess which behaviors are unique to a specific syndrome (Dykens, 1995). The unique attributes that are identified can then be used to start to examine correlations between genes and behavior in theory. There are few studies specifically comparing individuals with RS with others with intellectual and developmental disabilities. In one study completed, a sample of women with RS was compared with a normative sample of adults with intellectual disabilities. Results indicated that the women with RS had lower levels of irritability, hyperactivity and inappropriate speech behavior in comparison (Mount, Hastings, Reilly, Case, & Charman, 2002). A similar study completed found no differences overall in terms of emotional and behavioral disturbance between girls with RS and a control group of girls with developmental disabilities based on scores from the Developmental Behavior

Checklist (Mount, Hastings, Reilly, Cass, & Charman, 2003). No definitive conclusions can be made from the findings presented but the data regarding prevalence, form, and underlying mechanisms will be used in this paper to describe issues germane to individuals with RS. The current literature regarding behavioral issues specific to communication, stereotyped hand mannerisms, and idiosyncratic behaviors will be examined in the following sections in more detail.

Communication. Girls and women living with RS have profound deficits in the area of functional communication skills as a result of the regression period characterized by the syndrome. The dramatic degeneration in adaptive skills including expressive and receptive language, functional use of hands, and mobility results in a limited behavioral repertoire of responses exhibited by individuals with RS (Sigafoos, Woodyatt, Tucker, Roberts-Pennell, & Pittendreigh, 2000). As a result, assessment of communication used by individuals with RS is complex. There is a small set of published studies attempting to address the communication abilities of individuals with RS within the literature. Of the research that has evaluated communication and RS, profound impairments in cognitive and functional communication skills are repeatedly reported (Bartolotta, Zipp, Simpkins, & Glazewski, 2011; Sigafoos et al., 2000).

Many studies have noted the prevalence of a combination of behaviors including vocalizations, eye gaze, gestures, facial expressions, and walking to a desired object exhibited by girls and women with RS (Sigafoos et al., 2000, Hetzroni & Rubin, 2006; Bartolotta et al., 2011). The repeated observation of a combination of behaviors suggests that these behaviors could be communicative in nature and possibly meaningful attempts at communicating by individuals with RS. Bartolotta et al. (2011)

conducted a survey study evaluating the perceptions of expressive communication skills by parents and professions working with girls and women with RS. On average, respondents reported that each individual with RS used three different modalities of communication, with eye gaze the most frequent modality reported to be used. Picture or symbol augmentative communication systems were rated as the second most frequently used modality followed by body movements. Of the girls reported in this study (n=116), 70% used oral speech at some point in their lives and 86% of those girls experienced a regression in speech skills thereafter. Comparison ratings of parents and professionals' perceptions indicate that individuals with RS are perceived by familiar communication partners as able to intentional communicate with them.

In past research, it has been argued that individuals with RS do not use idiosyncratic behaviors such as stereotyped hand mannerisms, sustained eye gaze, undifferentiated vocalizations, facial expressions, body movements, and hyperventilation to intentionally communicate with others (Sigafoos et al., 2000). In a study that utilized semi structured interviews with parents and direct observations of potential communicative acts of 42 girls and women with RS found little evidence of intentional communicative behaviors (von Tetzchner, 1997). The role of the communicative partner has been highlighted as critical in terms of interpretation and responsiveness to unique behaviors exhibited by individuals with RS (Sigafoos et al., 2000; Bartolotta et al., 2011). However, no systematic research to date has investigated the role of the communicative partner with girls and women with RS. More work is needed to evaluate how potential communication acts are addressed and responded to by caregivers and professionals working with individuals with RS.

Sigafoos et al. (2000) attempted to assess potential communicative function of a range of response topographies observed in three people with RS with structured interviews and direct observations. The aim of the study was to examine if there was evidence that supported the notion that observable motor responses were conditioned as unconventional forms of communication. Results indicated that some existing behaviors exhibited reliably by participants within the study were differentially sensitive to various structured conditions. There were no conclusive findings as to the extent to which the behaviors observed were forms of intentional communication acts. The study did report however that staff interviewed attributed many behaviors exhibited by the participants as forms of communication even though agreement as to what the participant was communicating was unclear.

When adaptations are applied in the form of augmentative communication, there is some evidence of intentional communication displayed by individuals with RS apparent (Bartolotta et al., 2011). Van Acker & Grant (1995) taught three girls with RS to touch a computer screen to request food items reliably in an augmentative communication training for instance. A similar study evaluated the use of picture symbols displayed on a computer screen for classroom instruction with three girls with RS (Hetzroni, Rubin, & Konkol, 2002). Meaningful eye gaze was tracked to examine the accuracy and utilization of the picture symbols. Results from both studies cannot be generalized to all individuals with RS since skill sets are variable across girls and women living with RS, however both highlight the need for more research to be done to address communication and RS. For individuals with RS, research and identification of functional alternative modes of communication is needed to increase opportunities and

enhance skills using any residual traditional and non- traditional forms of communicative behaviors.

Stereotypic and repetitive hand mannerisms. The replacement of functional hand use with stereotypic and repetitive hand mannerisms is a core behavioral feature of RS. Hand mannerisms exhibited by individuals with RS include wringing, clasping, patting, mouthing, and releasing (Engerstrom, 1992; Hagberg, 1993; Wales, Charman, & Mount, 2004). Mount et al. (2001) reported in a literature review that in 38 case reports reviewed, hand stereotypy was present in 100% of the cases cited. Survey studies have found similar reports of highly intensive rates of hand stereotypies in girls and women with RS (Coleman, Brubaker, Hunter, & Smith, 1988; Sansom, Krishnan, Corbett, & Kerr, 1993). Stereotyped hand mannerisms can cause tissue damage due to the repetitiveness of the action and are ubiquitous within the behavioral repertoire characterized by RS. The underlying root cause of repetitive hand mannerisms specific to RS are not clearly understood at this time but are widely accepted as due to neurophysiologic abnormalities among the scientific community (Mount et al., 2001).

In an effort to increase functional hand use, operant reinforcement strategies have been successfully used to reduce inappropriate hand stereotypies with individuals with RS (Roane, Piazza, Sgro, Volkert, & Anderson, 2001). Piazza, Anderson, and Fisher (2001) demonstrated that self-feeding hand skills could be taught to five individuals with RS that had previously lost functional hand abilities. A method of prompting and reinforcement was used and showed improvements in self-feeding skills across all five participants. Interventions and therapies aimed at facilitating adaptive functioning in girls and women with RS are thus possible. Information regarding how

specific reinforcement contingencies affect the prevalence of stereotypic and repetitive hand mannerisms for individuals with RS have also been investigated in the past. The following section on functional assessment will review the current findings in detail.

Self-injury. Self-injurious behaviors (SIB) are problematic due to the physical damage that can be caused, the effects it can have on health, and the impact it has on quality of life (Symons, Koppekin, & Wehby, 1999). Coleman et al. (1988) surveyed 63 cases of RS between the ages of 2 -20 and reported the presence of self-injury in nearly half the cases. Sansom et al. (1993) reported similar rates of SIB in a clinical sample of 107 individuals with RS aged 2 to 28 years old surveyed in the form of hand biting and hand to head hitting most frequently. The etiology of SIB in RS is unknown but the prevalence of repetitive and stereotyped hand mannerisms that can cause tissue damage suggests an organic cause (Roane et al., 2001). As previously stated, other forms of SIB are also reported in the RS literature, which indicates that not all SIB may have an organic cause and could be maintained by operant mechanisms.

Advances have been made in the past two decades in the analysis and treatment of some forms of SIB in persons with developmental disabilities (Symons et al., 1999). It is now clear that SIB can be maintained by behavioral learning mechanisms (i.e., positive/negative reinforcement) and that treatments that effect a change in social reinforcement can be effective in persons whose SIB is socially-mediated. SIB specific to RS in the form of hand biting has been evaluated in the literature with experimental analyses (Iwata, Pace, Willis, Gamache, & Hyman, 1986; Oliver, Murphy, Crayton, & Corbett, 1993; Roane et al., 2001). Reviewed in detail below, the limited work to date examining behavioral mechanisms and SIB suggest that treatment demonstrations based

on reinforcement strategies have been effective in minimizing SIB in individuals with RS (Iwata et al., 1986; Roane et al., 2001).

Idiosyncratic behavior. There are a variety of idiosyncratic forms of behavior engaged in by girls and women with RS that are ambiguous from a functional perspective (i.e., it is not clear what their etiology is nor the maintaining mechanism). Idiosyncratic behaviors in RS include breathing irregularities (hyperventilation), body movements (rocking, wiggle, moving torso or head towards or away), stereotyped hand mannerisms, eye gaze (sustained), undifferentiated vocalizations, and facial expressions (Coleman et al., 1988; Engerstrom, 1992; Hagberg, 1993; Sigafoos et al., 2000).

Several studies have gathered information on the prevalence of idiosyncratic behavioral features with questionnaires. Questionnaires collected by Coleman et al. (1988) reported that a large proportion of individuals with RS exhibited a range of idiosyncratic behaviors including screaming fits, intermittent vocalizing (moaning/groaning), breath holding, teeth grinding, and night laughing co-occurring with sleep problems and aggression. Sansom et al. (1993) documented similar behaviors in their survey study. Mount et al. (2001) analyzed 32 case reports and found that 84% reported breathing irregularities among girls and women with RS. Comparatively, Naidu et al. (1990) conducted direct behavioral observations of 22 post-regression RS cases and reported that 87% exhibited hand mouthing, 33% screaming, and 13% aggression.

The range of idiosyncratic behaviors is a challenge because it has been difficult to identify their function and what they might ‘mean’ for anyone case or whether there are commonalities across cases. Research has focused in the past on the prevalence of

behavioral clinical features specific to RS, but little has been done to address treatment or function of the behaviors reported.

In summary, although it has been noted that individuals living with RS have limited behavioral repertoires, it should be emphasized that reliable patterns of behavior, albeit idiosyncratic at times, occur. To date, little functional information is available about the idiosyncratic behaviors observed. Furthermore, there have been no reported functional assessments investigating the extent to which idiosyncratic behaviors exhibited by individuals with RS may be sensitive to social reinforcement contingencies.

Functional Assessment and RS

Functional analysis (FBA/FA) is a behavioral assessment strategy designed to identify the environmental influences in the form of reinforcement mechanisms and functional characteristics of behavior (Iwata & Worsdell, 2005). A number of studies based on functional assessment have demonstrated that aberrant and problem behavior among individuals with severe intellectual and related developmental disabilities (I/DD) can be responsive to reinforcement contingencies and thus can inform effective clinical treatment (Iwata, Dorsey, Slifer, Bauman, & Richman, 1982/1994; Vollmer, Marcus, Ringdahl, & Roane, 1995; Carr & Durand, 1985)**.**

Contingent reinforcement increases the likelihood that a response will be repeated in the future and functions in three commonly accepted ways (Carr, 1977; Iwata & Worsdell, 2005)**.** Negative reinforcement is the contingent removal of aversive stimuli following a response (e.g., stopping tooth brushing after a tantrum occurs) and positive reinforcement is the contingent presentation of a stimulus after a response (e.g.,

giving attention in the form of verbal reprimand when self-injury occurs) (Carr, 1977; Iwata & Worsdell, 2005). Negative and positive reinforcement contingencies have been demonstrated repeatedly to maintain problem behavior in a range of I/DD samples (Iwata & Worsdell, 2005). Automatic reinforcement has been documented as well in the maintenance of challenging behavior, which contends that the behavioral response itself provides reinforcement for the individual (e.g., eye poking for visual stimulation) (Carr, 1977). Behaviors that are maintained by automatic reinforcement are non-socially maintained as opposed to positive and negative reinforcement contingencies that are mediated by other people (social) (Iwata & Worsdell, 2005). Overall, functional assessment of behavior is an effective behavioral technology to identify social reinforcement contingencies maintaining specific problem behaviors within social contexts.

In a recent PubMed search using the terms Rett syndrome and functional behavior assessments, only five articles were identified that directly address the relation between behaviors exhibited by individuals with RS and possible environmental contingencies maintaining them (Iwata et al.,1986; Oliver et al., 1993; Wehmeyer, Bourland, & Ingram, 1993; Roane et al., 2001; Wales et al. 2004). Of the five studies, hand stereotypies and self -injury in the form of hand biting were assessed using functional analyses. No research has examined the idiosyncratic behavioral features and responses described above using functional analyses technology.

Of the studies that have investigated the effects of reinforcement contingencies with functional analyses, the results reported are variable. Iwata et al. (1986) used functional analyses to address hand biting in two children with RS and determined that

the behavior functioned as a self-stimulatory response for both cases. Wehmeyer et al. (1993) documented elevated levels of hand stereotypies during escape from demands conditions (negative reinforcement) for one woman with RS and elevated levels of stereotypies during a non- engagement condition (automatic reinforcement) for another woman with RS. Oliver et al. (1993) evaluated two women using a similar approach and found that the self-injurious behaviors for these individuals were multiply maintained by various social and non-social reinforcers. Roane et al. (2001) found that there was no differentiation between conditions suggesting automatic maintenance of behaviors for the two individuals with RS assessed based on the conditions used to evaluate the behaviors. Finally, Wales et al. (2004) utilized analogue conditions for eight people with RS examining repetitive hand behaviors. The study found little difference between the conditions lending evidence that automatic reinforcers maintained the behaviors exhibited.

Although the findings from Roane et al. (2001) and Iwata et al. (1986) were not differentiated (i.e., the function was unclear), they both demonstrated that targeted operant based interventions decreased the frequency of aberrant behaviors for the participants with the introduction of differential reinforcement of other behaviors paired with a response interruption procedure. Therefore, it possible to impact the frequency of some behaviors exhibited by girls and women with RS using operant learning principles in the cases reported. Repetitive hand stereotypies and self-injurious behaviors have been addressed with functional analyses in the RS literature but little is known about other behavioral features of RS in this regard.

Rationale for Study and Specific Purpose

Given the limited understanding of the functional properties of the various idiosyncratic forms of behavior associated with RS, there is a need for continued investigation into the behavioral features of the RS phenotype, particularly because they may be relevant for improving our ability to target potential classes of communicative responses. As reviewed above, the limited work to date using FBA/FA technology has focused almost exclusively on hand stereotypy and self-injury. The specific purpose of this study was to test whether FA technology could be applied to a broader range of idiosyncratic behaviors observed in a clinical sample of girls and women living with RS.

Method

Participants and Setting

Six females diagnosed with Rett syndrome ranging from age 4 to 47 were recruited (Table 1). Functional skills and communication abilities varied among the participants. All but one participant lived at home with their caregivers. All were dependent upon others for their basic needs. None of the participants had functional speech, all engaged in frequent stereotypic hand mannerisms with limited use of hands, and had profound deficits in adaptive behavior. There was a range of idiosyncratic behavior reported for each of the participants based on caregiver report (Table 2). All interviews and analyses were completed in the homes of the participants.

Procedure

Brief functional analyses were conducted with each participant to systematically test whether any of the targeted idiosyncratic behaviors (including self-injurious for one

participant), exhibited were sensitive to social reinforcement contingencies (Northrup et al., 1991). Multi-element single subject experimental designs were used to evaluate the dependent relationships between positive and negative reinforcement contingencies and the target behaviors. Prior to conducting the analysis, a brief interview based on Mace and Lalli's descriptive assessment (1991) was completed to gain information from the caregivers of each participant as to whether the participant had any problem behaviors. If no challenging behaviors were evident, as was the case for the majority of the participants, information was also gathered regarding antecedents and consequences for any possible idiosyncratic behaviors, which were the focus of the study.

Response definitions and measurement

Response definitions for each participant were operationalized based on informal observation. Informal thirty- minute observations were completed along with an interview with the caregivers prior to the assessment (Table 3). Data were collected using video cameras and were later behaviorally coded using MOOSES™ (Multi-Option Observation System for Experimental Studies) software program (Tapp, Wehby, & Ellis, 1995). Frequency counts of the target behaviors were recorded on site by a trained observer in order to repeat any elevated conditions observed during the implementation of the analyses.

Experimental Conditions

The brief functional analyses conditions or analogs were based on the conditions described by Iwata et al. (1982/1994). All conditions used were conducted within the home of each participant within the course of one day. Instances of the target behaviors for each participant produced a specified consequence in the form of positive or

negative reinforcement within each analog condition. Sessions were five minutes in duration for all but one participant. Evelyn's sessions were ten minutes because of the low frequency of the target behavior (vocal upsets). Trained research assistants implemented all conditions for the participants. Descriptions of each analogue condition and reinforcement contingencies are below. Note, for two participants (Evelyn and Tami), tangible conditions were also included. Based on observation and caregiver reports, tangible conditions were not tested for the other four participants because it was not evident that a strong reinforcement history was in place.

Control (free play*).* This condition was designed as a control condition. The therapist provided non-contingent attention every 15 seconds to the participant. Access to leisure materials, such as music and preferred toys, were made available to the participants with no task demands placed upon the participants in the condition. No contingencies were implemented for target behaviors.

Social Positive Reinforcement (attention*).* This condition was designed to assess the influence of contingent attention on the target behaviors. The therapist was sitting 3 feet away from the participant with their back turned 45 degrees away. The therapist started with sitting next to each girl and then said "Ok, I have to go do some paperwork now" and got up and walked across the room. Any demonstration of the target behavior resulted in the therapist coming back to where the participant was sitting and talking with the participant for 15 seconds.

Negative reinforcement (escape*).* This condition was designed to assess the influence of task demands on target behaviors. Task demands included daily living activities in the form of stretching, hair brushing, or hand over hand prompting to

activate a toy. Contingent on each occurrence of the target behavior, the task was terminated and task materials were removed for 15 seconds.

Positive Reinforcement (tangible). The tangible condition was designed to assess the influence of access to preferred toys, music, television or food on target behaviors. A preferred item as indicated by the parent was given at the start of the condition. Contingent on an occurrence of the target behavior, the preferred item was given for a 15 second period.

The experimental conditions were all implemented at least twice if elevated rates of behavior were recorded. The control condition was used primarily as a comparison for any elevated conditions respectively. The average time to complete the experimental analysis was 60 minutes.

Inter-Observer Agreement

Two coders independently reviewed the videos and behaviorally coded the targeted behaviors with MOOSES™ (Tapp et al., 1995). Inter-observer agreement was calculated for approximately 27% of all the experimental conditions implemented across all six participants. Percent of agreement was defined as the number of agreements between observers divided by the sum of agreements and disagreements multiplied by 100 to yield a percentage. The mean inter-observer agreement for each participant for the functional analyses was 93% (range, 85% to 100%) for Katie, 87% (range, 75% to 100%) for Estella, 87% for Ann (range, 85% to 90%), 94% for Tami (range 83% to 100%), 84% for Rachel (range 82% to 94%), and 85% for Evelyn (range 82% to 86%).

Analysis of the Data

Frequency or duration counts, depending on the target behaviors, were recorded from videos of the functional analyses. Data from the analyses of each participant were then evaluated by comparing the overall average response rate occurring during each condition. Differences in trend, level, and variability across conditions for each participant were visually examined based on the graphs displayed in figure 1. The mean rate (frequency per minute) of responding for each condition was then calculated to confirm the visual inspection of the data summarized in figure 1.

Results

Figure 1 displays the summary data from all the participants across all the experimental conditions conducted. For four out of the six participants, differentiated results as indicated by visual analysis of the data were observed. The following section includes descriptions of each participant's results. Overall, differentiated results among the experimental conditions used to examine the relation between environmental reinforcement contingencies and target behaviors suggests that for some of participants, targeted idiosyncratic or challenging behaviors were differentially sensitive to various reinforcement contingencies.

Differentiated results were observed for Katie, Estella, Evelyn, and Tami. Katie's results show that vocal upsets occurred most frequently and consistently in the escape condition (negative reinforcement). A mean rate per minute of 6.53 was calculated for the escape conditions. In comparison, the control condition and attention condition yielded lower rates of vocal upset. In the control condition, a mean rate per minute of 0.76 was observed. The attention condition (positive reinforcement) resulted

in the lowest level of responding with a mean rate per minute of 0.50. An escalating trend is apparent in the escape condition with the highest rate of responses observed in the last session. The control and attention condition data remained low and stable through the analyses. The data suggest that Katie's vocal upsets are influenced by negative reinforcement in the form of escape from demands.

Estella's results from the functional analysis show that vocal upsets occurred most frequently and consistently in the escape condition (negative reinforcement). A mean rate per minute of 1.87 was calculated for the escape conditions. In comparison, the control condition and attention condition yielded lower rates of vocal upset. The attention condition (positive reinforcement) yielded the next highest rates of vocal upsets with a mean rate per minute of 0.60. An escalating trend is apparent in the attention condition with increased rates of responses observed in the last session. Vocal upsets occurred at a mean rate per minute of 0.50 in the control condition and was the lowest overall rate recorded in the analysis. An escalation in level was observed in session 7 of the control condition. Within session analysis indicate that responding occurred within the first 30 seconds of the control condition. It is hypothesized that a carry over effect from the escape condition tested prior to the control condition is the reason vocal upsets increased within this session. The data suggest that Estella's vocal upsets are influenced by negative reinforcement in the form of escape from demands.

Evelyn's functional analysis data suggests that her vocal upsets may be maintained by positive reinforcement in the form of tangibles. Vocal upsets occurred most frequently and consistently in the tangible condition (positive reinforcement). A mean rate per minute of 0.45 was calculated for the tangible conditions. The escape

condition showed the next highest rates of vocal upsets with a mean rate per minute of 0.15. The attention and control condition yielded the lowest rates of vocal upset with a mean rate per minute of .05 and 0 respectively. An escalating trend in the data is apparent in the tangible conditions. A decelerating trend was observed for all the other conditions tested with minimal occurrences of vocal upsets.

Tami's data suggest that her self-injury is maintained by positive reinforcement in the form of attention. The highest rates of self-injury were recorded for the attention condition with a mean rate per minute of 2.7 across all three sessions. Self- injury was variable across the three sessions conducted with the most instances of self-injury occurring in the last session of the attention conditions. No instances of self-injury were observed for any of the other condition tested including escape, attention, and tangible.

It is less clear as to whether Ann and Rachel's results yielded differentiated results. Ann's functional analysis demonstrated elevated levels of breath holding and vocalizations in the escape conditions with a mean response rate per minute of 2.4. The control conditions resulted in a mean response rate of 1.56 per minute. A decelerating trend and plateau in the data in the control conditions is apparent. The lowest rates of responses were recorded in the attention conditions where the mean rate per minute was 1 respectively. Since Ann's response level never reached the zero point the data suggests that her behavior does not have a true operant function. However, Ann's functional analysis did show that the conditions tested did influence the frequency at which the behavior was observed.

Finally, Rachel's data shows relatively low rates of hand mouthing across test conditions. The condition with the highest recorded rates of hand mouthing was for the

escape conditions (negative reinforcement) with a mean rate per minute of 0.50. Data from the attention conditions were the next highest rate of hand mouthing observed with a mean of 0.30. No instances of hand mouthing were recorded for the control conditions. The data suggest that Rachel's hand mouthing was differentially sensitive to both negative and positive reinforcement in the form of escape from demands and attention, however it was difficult to discern if this behavior had one operant function because the rates of responding in the escape conditions were only slightly higher than the rates observed in the attention conditions.

Discussion

The present study investigated if idiosyncratic behaviors exhibited by a small clinical sample of individuals with Rett syndrome were sensitive to operant contingencies. Brief functional analyses were used to test whether systematically implemented reinforcement contingencies influenced the frequency of the identified target behaviors. The results indicated that for four out of the six analyses completed, differentiated results were apparent as indicated by visual analysis of data and comparisons of the mean rates of responses recorded across experimental conditions conducted. The main findings of this study are, 1) a range of idiosyncratic behaviors was found to be differentially sensitive to reinforcement contingencies tested in a clinical sample of individuals with RS, and 2) functional analyses technology provided a viable tool for identification of environmental social reinforcement contingencies that influence target behaviors for the group of individuals with RS examined.

Results of this study extend the literature on RS in several ways. First, in terms of the application of functional assessment (FA) technology, this study was successful

at using FA to address a range of idiosyncratic behaviors associated with RS. As examined in the introduction, previous research demonstrated that self-injurious behavior in the form of hand biting and stereotyped repetitive hand mannerisms in individuals with RS were not differentially sensitive in all but one case report to negative and positive reinforcement suggesting a automatic and self-stimulatory reinforcement maintaining the majority of the behaviors examined (Iwata et al.,1986; Oliver et al., 1993; Wehmeyer et al., 1993; Roane et al., 2001; Wales et al. 2004). Differentiated results from this study add valuable information for the RS behavioral phenotype in that it demonstrates that contingent reinforcement can influence the rate at which the targeted behaviors are exhibited. Differentiated results for target behaviors such as vocalizations in particular have implications for potential facilitation of communicative responses. Targeted interventions utilizing reinforcement or replacement strategies may be effective for the class of behavioral responses examined in this study.

Second, research on Interventions that are based on functional analyses are shown to be more effective than non-function based treatments typically for people with intellectual and developmental disabilities (Smith, Iwata, Vollmer, & Zarcone, 1993; Roane et al., 2001). Due to the deleterious effects that RS has on adaptive skills and communication abilities, effective treatments are needed to facilitate skill acquisition and maintenance to improve the quality of life of individuals with RS. The results of this study can be used to inform function- based treatments such as functional communication training. Functional communication training (FCT) is a strategy that utilizes differential reinforcement to increase appropriate communicative behaviors by

replaces alternative problem behavior with a functionally equivalent response (Carr & Durand, 1985). Specific to the present study, FCT could be implemented based on the functional analyses data to replace upset vocalizations for Katie, Estella, and Evelyn with a more appropriate response in the form of activating a speech-generating switch. FCT could also be implemented to replace Tami's self-injurious behavior, which was maintained by positive reinforcement in the form of social- attention.

One limitation of the current study is that treatment demonstrations were not utilized to confirm functional information derived from the assessment. FCT demonstrations would have emphasized the differentiated results based on the functional analyses. Another limitation of this study is that it is unclear as to if some of the analyses have a true operant function. Ann's results in particular did show differential sensitivity to reinforcement contingencies systematically implemented, however the target behavioral responses (vocalizations and breath holding) never reached the zero point. A third limitation of this study is that there is little clinical information regarding the participants in this study, such as if they have seizures, scoliosis and other health related information that is important for examining the RS phenotype. A better overall picture of each individual participant can be made with more clinical data since individuals with RS vary in terms their skills and abilities.

Future directions for research in behavior and RS should focus on adaptive behavior and potential classes of communicative responses. For example, Piazza et al. (2001) increased functional hand skills and reduced inappropriate hand stereotypies through implementation of reinforcement strategies. Iwata et al. (1986) and Roane et al. (2001) also successfully implemented treatment demonstrations using differential

reinforcement of other behaviors paired with response interruption to minimize the occurrence of hand biting and hand stereotypies. Thus behavioral interventions have been shown to improve skills yet more research is needed to elucidate effective treatments for other areas of deficits such as communication. More regarding how communication is addressed and responded to specifically in girls and women with RS is critical. The extent to which caregivers and professional respond to a range of idiosyncratic behavior has not been systematically examined in the literature to date.

In closing, the purpose of this study was to investigate whether idiosyncratic and challenging behaviors exhibited by several individuals with RS were differentially sensitive to reinforcement contingencies tested. More systematic research is warranted regarding the functional properties of a range of behavioral features characterized by RS to better inform clinical treatment and interventions in the areas of adaptive skills and communication in particular. Function-based interventions have the possibility of enhancing the quality of life for individuals with RS and more attention in the literature needs to be focused on this population of girls and women.

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Appendix

Table 1

Participant Information

Participants	Age in Years	Mobility	Form of Hand Stereotypy
Ann	28	Ambulatory	Clasping
Evelyn	5	Ambulatory	Finger Rubbing
Katie	7	Nonambulatory	Hand Mouthing
Estella	4	Nonambulatory	Finger Rubbing
Rachel	47	Ambulatory	Hand Wringing
Tami	26	Nonambulatory	Finger Rubbing and Patting

Table 2

Caregiver Reported Behaviors

Participant	Hyperventilation	Screaming Spells	Laughing for no reason at times	Bites Hands	Hits Herself	Hits or Kicks Others	Communicates Most Often With
Ann	No	No	No	No	No	No	Eyes & Gestures
Evelyn	No	Sometimes	Sometimes	No	Sometimes	Sometimes	Eyes
Katie	No	No	Sometimes	Often	No	No	Eyes
Estella	Sometimes	Sometimes	Sometimes	Sometimes	No	No	Eyes
Rachel	No	No	Sometimes	Sometimes	Sometimes	Sometimes	Eyes & Gestures
Tami	Sometimes	Sometimes	Often	Sometimes	Often	Sometimes	Gestures

Note. The rating scale included 'No, Sometimes, and Often' as the three choices to select from.

Table 3

Response Definitions

Target Behavior	Definition	Participants
Breath holding	Breath holding as indicated by audible breath exhalations	Ann
Vocalizations	Audible vocal behavior	Ann, Rachel
Upset vocalizations	Audible vocal behavior accompanied by crying	Evelyn, Katie, Estella
Aggression	Audible contact between the participant's hand and another person	Evelyn
Hand mouthing	The insertion of one or more fingers in the mouth	Rachel
Head hitting	Any forceful hand contact with any part of the head or face	Tami
Stomach hitting	Any forceful hand contact with any part of the participant's abdomen	Tami

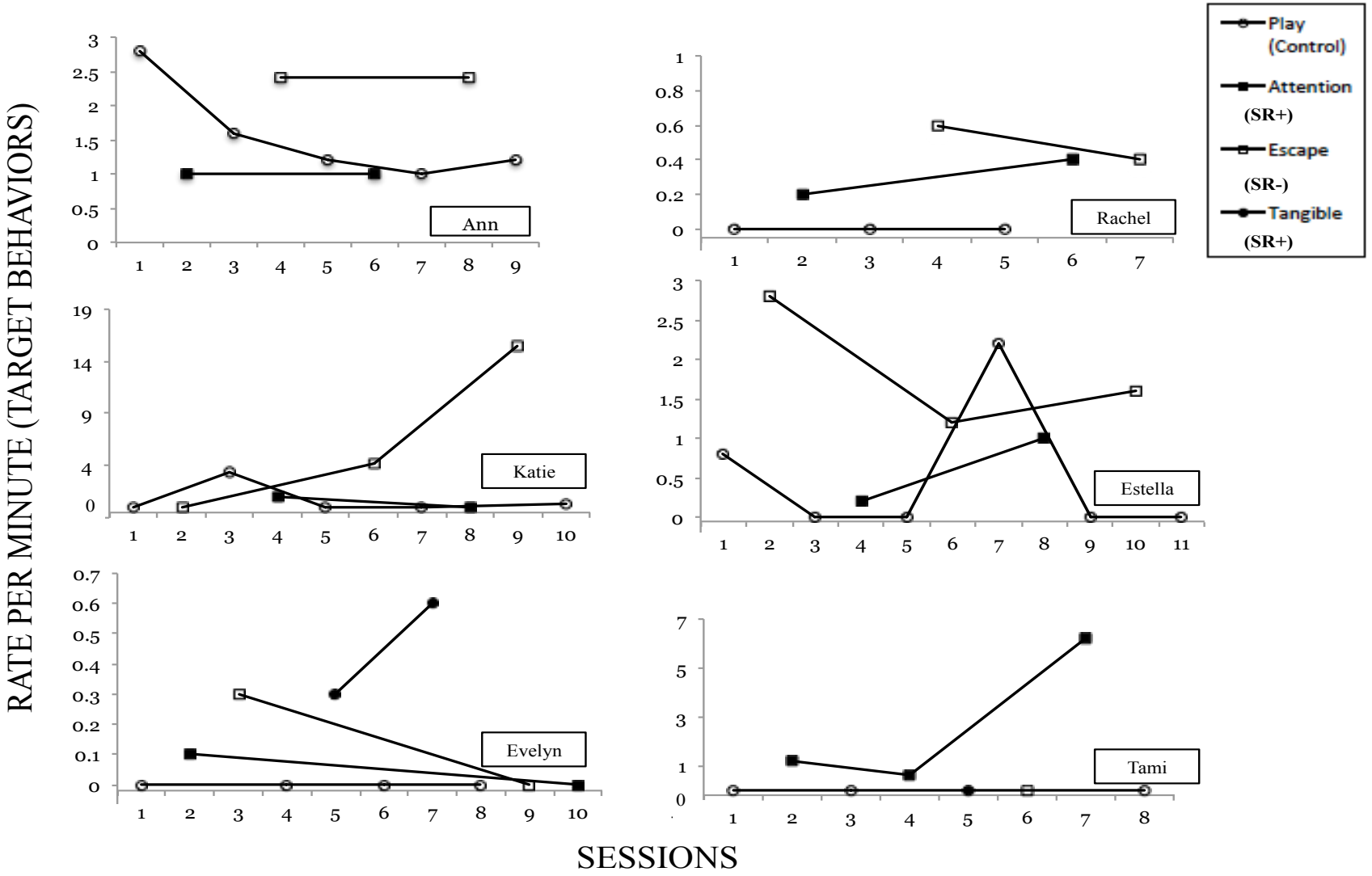

Figure 1. Summary data for all participants across experimental conditions.