Sustained Attention in the Face of Distractors: A Study of Children with Rett Syndrome

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Abstract

Objective. The object of the present study is to advance our understanding of the cognitive profile of Rett Syndrome (RTT), an x-linked neurodevelopmental disorder caused by mutations in the MECP2 gene. We focus on sustained attention, which plays a critical role in driving cognitive growth, and use an innovative, gaze-based task that minimizes demands on the limited verbal and motor abilities associated with RTT.

Method. The task required the ability to sustain attention on a visual target (a butterfly) whilst inhibiting a prepotent response to look to moving distractors (trees and clouds) presented in the peripheral visual field. The sample included children with RTT (N = 32) and their typically developing (TD) counterparts (N = 32), aged 2-12 years.

Results. Our findings revealed that children with RTT had more difficulty sustaining attention (with the TD group averaging 60% looking at the butterfly vs only 25% for the RTT group). Furthermore, they showed that RTT was associated with difficulties in three fundamental factors influencing sustained attention: engagement, distractibility, and re-engagement. The RTT group was *slower* to engage, *more* distractible, and *slower* to re-engage.

Conclusion. Our findings suggest there may be a fundamental disruption to sustained attention in RTT, identifies factors related to this impairment, and points to cognitive areas that could be assessed in evaluating the usefulness of interventions.

Public Statement. Rett syndrome is an x-linked neurodevelopmental disorder characterized by a developmental regression that typically begins between 6 and 18 months, robs the child of purposeful hand use and expressive language, and results in the development of numerous medical problems. These deficits make standard neuropsychological testing becomes all but impossible, and thus little is known about their cognitive abilities. We were able to by-pass the motoric and language problems using eye-tracking technology. Here we used an innovative gaze-based task to assess sustain attention, a core driver of cognitive growth. We found that children with Rett Syndrome showed impairments in sustained attention, and on three

fundamental factors influencing sustained attention: engagement, distractibility, and re-engagement. Children with Rett Syndrome were *slower* to engage, *more* distractible, and *slower* to re-engage than their age-matched peers. This work not only begins to elucidate the nature of the cognitive problems associated with Rett syndrome, but is essential for designing markers to assess the effects of pharmacological interventions.

Key Words: Rett syndrome; sustained attention; gaze-based task; eye-tracking; cognition

Sustained Attention in the Face of Distractors: A Study of Children with Rett Syndrome

Rett syndrome (Rett, 1966) is a severely disabling, x-linked neurodevelopmental disorder characterized by apparently normal early development followed by developmental regression between 6 and 18 months in which purposeful hand use and expressive language are lost and impaired gait and hand stereotypies appear (Chahrour & Zoghbi, 2007). Other symptoms include the development of seizures, apraxia, spasticity and scoliosis, breathing irregularities (hyperventilation, breath holding, apnea), and a slowing of brain and head growth (Neul et al., 2010).

This disorder, which affects about 1 in 10,000 females, is caused by spontaneous mutations in the *MECP2* gene, located on the long arm of the X chromosome – Xq28 (Amir et al., 1999). The *MECP2* gene encodes methyl-CpG-binding protein 2 (MeCP2), which is involved in regulating the transcription of other genes, synaptic development and maintenance (Guy, Gan, Selfridge, Cobb, & Bird, 2007), and is required for learning and memory (Moretti et al., 2006.). Mutations lead to a significant reduction in long-term potentiation after symptom onset in *MECP2*+/– females, with the magnitude of the defect similar to that reported in MeCP2-null mice (Guy et al., 2007).

The severe limitations in language and purposeful hand use associated with Rett syndrome (RTT) have precluded most neuropsychological testing of these children, with the result that little is known about the cognitive phenotype of the disorder. However, recent studies using eye tracking technology have shown progress in characterizing the behavioral and cognitive profile of RTT. These studies found that children with RTT showed a preference for socially weighted stimuli, as well as selective attention to salient areas and novel elements

(Djukic & Valicenti McDermott, 2012; Djukic, Valicenti McDermott, Mavrommatis, & Martins, 2012). While they were able to recognize simple patterns, faces and some emotional expressions, their performance was significantly poorer than that of typically developing (TD) children, and appeared to be related to attentional difficulties (Djukic, Rose, Jankowski, & Feldman, 2014; Rose et al., 2013; Rose, Djukic, Jankowski, Feldman, & Rimler, 2016). These difficulties included less looking at the targets and frequent failure to look at critical aspects.

These problems in attention are of particular concern because attention is a core dimension of cognitive growth that has a cascading effect on subsequent learning and development. Recent studies have shown that attention plays a pivotal role in gating the development of working memory (Astle & Scerif, 2009) as well as in driving the development of more complex outcomes, including IQ (Rose, Feldman, Jankowski, & Van Rossem, 2005, 2008), language (Rose, Feldman, & Jankowski, 2009; Whedon, Perry, Calkins, & Bell, 2016), executive functions (Rose, Feldman, & Jankowski, 2012), academic achievement (Bornstein, Hahn, & Wolke, 2013), and eventual employment status (Kalechstein, Newton, & van Gorp, 2003). In our own lab, we identified a developmental cascade in which elementary abilities evidenced in infancy (attention and speed) influenced more complex abilities (memory and representational competence) that, in turn, influenced general cognition in toddlerhood and early adolescence (Cornish, Cole, Longhi, Karmiloff-Smith, & Scerif, 2012; Rose, Feldman, Jankowski, & Van Rossem, 2012; Rose et al., 2005, 2008; Rose, Feldman, Jankowski, & Van Rossem, 2011; Scerif, Longhi, Cole, Karmiloff-Smith, & Cornish, 2012).

To understand the role of attention, we need to recognize that it is a multi-dimensional construct that includes a number of different processes, with different attentional functions subserved by distinct, but overlapping neural systems (Fan, McCandliss, Fossella, Flombaum, &

Posner, 2005; Posner & Petersen, 1990). Posner distinguished three specialized brain networks underlying attention – alerting, orienting, and executive attention (Petersen & Posner, 2012). Alerting, which involves the thalamus, as well as right frontal and parietal cortical sites, and is mediated primarily by the neuromodulator norepinephrine, achieves and maintains high sensitivity to stimuli (Aston-Jones & Cohen, 2005; Petersen & Posner, 2012). Orienting, which involves a dorsal network (including the frontal eye fields and superior parietal lobe), as well as a more ventral network (including the parietal-temporal junction), and is thought to be subserved primarily by cholinergic networks (Davidson & Marrocco, 2000), is important for the selection of stimuli from sensory input. Although it was previously thought that the dorsal network was endogenously driven, and the ventral network exogenously driven (Corbetta & Shulman, 2002), more recent evidence indicates that both networks are involved in re-orienting, showing that this process is endogenously as well as exogenously driven (Corbetta, Patel, & Shulman, 2008). Executive attention, which involves the anterior cingulate cortex and prefrontal areas, is important for situations involving conflict, where inhibition is necessary.

The tasks used in our earlier work involved several aspects of attention in combination. One that figured prominently was sustained attention -- the ability to focus or concentrate attention on a task or maintain vigilance in the face of distractors. The present study attempts to better understand the difficulty Rett children have with this aspect of attention and identify factors influencing it. Sustained attention, which is thought to involve top-down connectivity extending from the anterior attention system, particularly prefrontal and parietal regions in the right hemisphere, right down into V1 (Grahn & Manly, 2012; Sarter, Givens, & Bruno, 2001; Silver, Ress, & Heeger, 2007), has repeatedly been found to be compromised across a wide range of neurological and psychiatric disorders, e.g., ADHD, autism, bipolar disorder and Fragile

X (Cornish, Scerif, & Karmiloff-Smith, 2007; Cornish, Turk, & Levitas, 2007; Fortenbaugh et al., 2015; O'Connell, Bellgrove, Dockree, & Robertson, 2004)

While sustained attention is often tested in adults with the continuous performance test, the verbal instructions and motoric requirements preclude using this task in children with RTT. To overcome these limitations, sustained attention was assessed here by building on tasks that have assessed how well children can visually concentrate on a target while ignoring distractors (Oakes, Kannass, & Shaddy, 2002; Richards, 1987; Ruff & Rothbart, 1996). We used an innovative, gaze-based task modeled after Wass and colleagues (Wass, Porayska-Pomsta, & Johnson, 2011). A target (a butterfly) was presented on the screen. When the child fixated on the target it moved from left to right and distractors (trees and clouds) scrolled in the opposite direction. When the child looked to any of the distractors, the display froze. The task has three key features. First, the movement of the butterfly is gaze-contingent (it moves only when fixated) and thus there is a reward component for sustaining attention. Second, the necessity for motoric and verbal abilities is minimized. Third, the task allows us to assess not only sustained attention, but also factors that impact it, including time to engage the target, distractibility, and re-engagement. This new task thus targets executive attention and the orienting network. Executive attention is involved in inhibiting attention to the distractors, and the orienting network when the child initially directs attention to the target at the outset of a trial or re-directs attention from the distractor to the butterfly during a trial.

We hypothesize that the Rett children will show less sustained attention and more distractibility (time off task) than typically developing children, particularly as the number of distractors increases. This hypothesis is based on brain imaging studies of children with Rett showing global decreases in brain volume (Carter et al., 2008), selective reductions in frontal white matter (Mahmood et al., 2010), and selective vulnerability of the frontal lobes (Naidu et al., 2001), all areas involved in inhibiting attention to distractors. We also hypothesized that group differences in orienting and re-orienting might be less marked, given data showing selective preservation of the occipital cortex, although selective reductions in dorsal parietal grey matter, an area involved in re-orienting, makes this hypothesis more tenuous (Carter et al., 2008).

Method

Participants

This study was conducted on 32 females with clinically diagnosed classical Rett syndrome (Neul et al., 2010), consecutively recruited from the Rett Center at the Children's Hospital of Montefiore (M=7.92 years; SD=2.89, range=2-12) and a comparison group of 32 typically developing (TD) females (M=7.66 years; SD=2.83, range=2-12), $t(62)=.35$. The TD group, recruited from Outpatient Clinics of the same hospital, was drawn from children who were family members of patients with appointments at pediatric specialty clinics. The TD group was screened to exclude any children with significant neurological disorders (e.g., epilepsy, brain tumor), sensory impairment, neurodevelopmental disorders (e.g., autism, ADHD) or first-degree relatives with neurodevelopmental disorders.

RTT was genetically confirmed in all Rett participants. Testing was attempted, but terminated, for an additional 3 Rett patients who could not successfully complete the calibration procedure (described below) and 5 who were too overactive/restless to complete the testing procedure; these eight did not differ in clinical/background factors from the rest of the RTT group (falling in the moderate range of the RSSS scale described below).

Clinical characteristics of the Rett sample. Table 1 shows the genetic mutation, age at test, age at regression, scores on the Rett Syndrome Severity Scale (RSSS) (Kaufmann et al.,

2012) and notes their status on two subscales of the RSSS – walking and seizures. Composite scores on the RSSS averaged 8.25 (*SD* =2.43), with 14 patients (43.8) scoring in the *mild range* (0–7), and the remainder (56.2%) in *the moderate range* (8–14). Many (46.9%) were ambulatory (able to walk unaided or with support); 43.8% of the group had a history of seizures.

The protocol was approved by the institutional review board and written consent was obtained for all participants.

Table 1 goes about here.

Apparatus

Stimuli were presented on a 23-inch flat panel monitor (resolution, 1024 × 768 pixels) and integrated with a Tobii X2-60 eye-tracker, using Matlab and Psychtoolbox. Talk2Tobii software was used to allow for a live gaze-contingent interface via Matlab during stimulus presentation. Manufacturer-supplied algorithms for pupil, corneal reflection, and face identification were used during eye-tracking; gaze data were sampled at 60 Hz.

Calibration

At the beginning of the session each participant's point-of-gaze- was calibrated using a 5-point calibration procedure. The calibration stimuli, five pulsing colored blocks (1° to 1.5°) were presented sequentially, at different locations on the screen, accompanied by a sound ('Whee'). Point-of-gaze was calibrated by comparing each look to the known coordinates of the target, and results were inspected graphically. The quality of the calibration data was determined by the closeness of the fixation points to the calibration points. If the points did not cluster, or any targets were missed, the calibration was repeated until a satisfactory calibration was achieved. Each calibration attempt took less than a minute.

Stimuli and Procedure

Testing was conducted in a quiet room, with participants seated approximately 45 cm from the monitor. Ambient light levels were reduced to diminish distraction. Verbal instructions, limited to 'Look at the TV,' were used at the beginning of the session. To minimize body and head movement, all participants with RTT (and all TD participants < 5 years) were seated on their parent's lap. Parents kept their eyes closed during testing.

Trials started with a target, a butterfly (subtending 6°), presented on the screen (Wass et al., 2011). When the child fixated the target, it moved, fluttering its wings and 'flying" horizontally from left to right across the screen. Distractors, consisting of a house, a tree, and clouds (subtending 5-15°), scrolled in the opposite direction. The butterfly travelled at a rate of 2.5 cm/s, while the distractors moved in the opposite direction at the same rate. When the child looked at any of the distractors they disappeared, with only the butterfly target remaining. On re-fixating the target, it recommenced moving across the screen and fluttering its wings, and the distractors re-appeared and continued scrolling. Trials lasted 15 s and an engaging sound track (the melody, Zip-a-Dee-Doo-Dah) played throughout each trial. There were two blocks of 9 trials; each block contained three trials each with 1, 2, and 3 distractors, presented in pseudo-random order. The two blocks of trials, each lasting less than 2.5 minutes, were interleaved with two other attention tasks. The entire testing session took about 10 min.

Data Analyses

All measures were examined for normality and outliers and analyzed using a mixed model 2 (Group: RTT vs TD) x 2 (Age: younger vs older) x 3 (number of distractors: 1, 2, or 3)

ANOVA, with repeated measures on the last factor. Age was dichotomized for these analyses using a median split (< 8 years vs ≥ 8 years, for both groups). Where necessary, measures were transformed to achieve normality, using a log or square root transform (see below). All effects were evaluated at a .05 level of significance; SPSS (version 24) was used in all analyses; Bonferroni-adjusted significance tests were used for all pairwise comparisons.

Results

Latency of first looks to the target. The latencies of the child's first look to the butterfly are shown in Table 2. (Data values were $\log_{10}$ transformed for analysis to correct for positive skew in the distributions.)

As can be seen in Table 2, latencies were longer for the RTT group, $F(1,60)=10.31$, $p < .05$, $\eta_p^2=.15$, who took over 2.5 s to engage with the target (compared with slightly more than 1 s for the TD group). There was also a significant Group x Age interaction, $F(1,60)=8.34$, $p < .01$, $\eta_p^2=.12$, reflecting the finding that latencies got shorter with age for the TD children but not for the RTT children.

Table 2 goes about here.

Percentage of time looking at the target. As can be seen in Table 3, the RTT group looked at the butterfly for only about half as long as the TD group, $F(1,60)=132.39$, $p < .001$, $\eta_p^2 =.69$. A significant Age effect, $F(1, 60)=12.50$, $p = .001$, $\eta_p^2 =.17$, coupled with a Group x Age interaction, $F(1, 60)= 4.84$, $p=.03$, $\eta_p^2 =.08$, indicated that older children showed more sustained attention than younger children, although such age-related improvement was largely restricted to the TD group. There was also a significant Distractor effect, $F(2,120)=3.12$ $p=.04$, $\eta_p^2 =.05$, due to looking at the target decreasing as the number of distractors increased; a marginally significant Group x Distractor interaction, $F(2,120)=2.72$, $p=.07$, $\eta_p^2=.04$, indicated that the fall-off in performance was most pronounced for the RTT children.

Table 3 goes about here.

Time off target (Number of looks to the distractors). Given that the TD group spent more time looking at the target than the RTT group, they had more opportunities to look away. To adjust for this factor, the number of looks away was divided by looking time to the target. These values, number of looks away from the target (per s), were created for each trial and then averaged over trials for each of the three distractor conditions. (Scores were $\log_{10}$ transformed for analysis to correct for positive skew.)

As can be seen in Table 4, the RTT group showed more looks away from the target per second than the TD group, $F(1,60)=43.28$, $p < .001$, $\eta_p^2=.42$. There was also a Group x Age interaction, $F(1, 60)= 4.27$, $p=.03$, $\eta_p^2=.07$, indicating that distractibility decreased with age for the TD children, but not for the RTT group. The Distractor effect indicated that, as expected, the number of looks away per second increased for both groups as the number of distractors increased, $F(2,120) = 9.59$, $p<.001$, $\eta_p^2= .14$.

Table 4 goes about here.

Time to re-focus on the target after looking at distractors. The average time to re-engage the target after each off-target look is shown in Table 5. (A square root transformation was used to normalize these distributions for analysis.) The difference between groups is marked, with the Rett group taking about three times as long to re-engage as the TD group, $F(1,60)=157.01$, $p<.001$, $\eta_p^2= .72$. There was also an Age effect, $F(1,60)=11.10$, $p=.001$, $\eta_p^2= .16$, with older children re-engaging faster than their younger counterparts. Although this effect was particularly marked in the TD group, the interaction was not statistically significant. Additionally, there was a Distractor effect, $F(2,120)=6.16$, $p<.01$, $\eta_p^2= .09$, and a Group x Distractor interaction, $F(2,120)=4.48$, $p=.01$, $\eta_p^2= .07$ reflecting longer re-engagement times with two distractors in the RTT group. These last two effects were unexpected, given that the distractors disappeared as soon as they were attended to.

Table 5 goes about here.

Clinical characteristics of the Rett children and performance. None of the clinical characteristics of the Rett sample listed in Table 1 correlated significantly with any measure of performance.

Discussion

In this study, we examined the degree to which sustained attention is affected in children with Rett syndrome (aged 2-12 years), and the role of factors that impact sustained attention, including time to engage, distractibility, and re-engagement. We used an innovative task which required the child to maintain their gaze on a moving target while ignoring distractors that moved in the opposite direction (Wass et al., 2011). The movement of the target was gaze-contingent – that is, the butterfly moved across the screen only when the child looked at it, thus rewarding the child for sustaining attention to it. The verbal and motor requirements of this task are minimal, making it uniquely suited for use with the RTT population. We varied the number of distractors, to make the task more or less taxing, and examined age-related effects by using a median split on age.

While the RTT children were able to sustain attention on the butterfly, they did so for only 25% of the time, while the TD group did so for more than 60%. One factor that appears to underlie their difficulty is distractibility, with RTT children being drawn to the distractors nearly twice as often as TD children. Moreover, while performance for both groups tended to fall off as the number of distractors increased, this effect was accentuated in the RTT group. That is, as the number of distractors increased, they showed a more marked downturn in the time spent looking at the butterfly and looked away more often to the distractors. A second factor that had an impact on the ability of the RTT children to sustain attention was the latency to re-engage the butterfly after having their attention pulled away from it. Indeed, once they looked to the distractors, the RTT children took nearly three times as long to re-engage with the butterfly as did the TD group.

Why is the RTT group so much more distractible than the TD group? The most likely possibility, often discussed in regard to distractibility in ADHD, is impairment in inhibitory systems (Barkley, 1997; Nigg, 2001). That is, children with ADHD are thought to be unable to resist the pull of irrelevant stimuli when completing a task. The same problem may be operating here as well. That is, the RTT children may not have the inhibitory control ability needed to ignore the distractors, even though the distractors disappear as soon as the child turns to them, and thus there is little pay off in continuing to turn to them. The RTT children clearly found the moving distractors compelling, and were less able than the TD group to resist their draw, especially as their number increased.

Why are the RTT children so much slower than the TD group to re-engage the butterfly after being distracted? This finding is more difficult to explain. After all, the distractors have disappeared. Oculomotor factors cannot fully account for this difference given that, despite similar oculomotor demands, the re-engagement latencies in the RTT group were substantially longer than their initial latencies to engage the target, $t(31) = 3.88$, $p = .001$, $d = .69$. One possibility is that the RTT children have more difficulty making the complex set of adjustments involved in the interaction of the dorsal and ventral frontoparietal systems involved in re-orienting (Corbetta & Shulman, 2008). This possibility receives some support from a recent study which showed that states of high global integration of neural networks are associated with better performance (Shine, 2016). Any differences that exist between groups in arousal level may also have affected re-orienting, since the integration of networks tracks with fluctuations in arousal (Shine, 2016), and attention has been shown to be modulated by arousal (Aston-Jones & Cohen, 2005; de Barbaro, Clackson, & Wass, in press). Another possibility is that the RTT children are slower to re-calibrate, and in appraising the situation realize that there is no pay-off in continuing to look to locations where the distractors had been. This possibility is consistent with earlier findings where children with RTT had difficulty learning the rule underlying event sequences (Rose et al., 2016).

The effects of age were examined using a median split for both groups. Children in the TD group showed improvement over age for all measures, significantly so for latency to first look and sustained attention (time spent looking at the target) and marginally so for re-engagement. There were no age effects for the RTT group, a finding consistent with previous work (Rose et al., 2013). It is probable that any tendency to improve over age is counteracted by the progressive nature of the disorder.

The gaze-based task would appear to be a useful way for testing sustained attention in other populations where verbal and motoric impairments preclude using other tasks, such as the continuous performance test. In the latter, often treated as the 'gold standard' for assessing sustained attention, a button is to be pressed as quickly as possible each time a target appears, while distractors are to be ignored; the critical measure is errors of omission (failures to press when the target appears). Two of the typical effects found with this task -- a strong negative effect of distractors and age-related improvement in sustained attention (Conners, Epstein, Angold, & Klaric, 2003) -- also prominent effects for the TD children on the gaze-based task used in the present study. This agreement in findings supports the usefulness of the gaze-based task for assessing sustained attention.

In summary, the present work identified difficulties in sustained attention associated with RTT and determined at least two factors implicated in these difficulties – distractibility and slowness to re-engage after distraction. This work helps to elucidate the nature of the cognitive problems associated with RTT, is essential for the design of intervention, and begins to indicate functions and tasks that could serve as markers for the effects of pharmacological interventions.

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Table 1

Clinical and Genetic Characteristics of the Children with Rett Syndrome

Patient	Genetics	Age (years)	Age at Regression (months)	RSSS Total Score[a]	Ambulatory[b]	Seizures[c]
1	R133C	7	15	8	0	0
2	R306C	11	18	5	1*	1
3	R133C	7	15	7	0	0
4	deletion	6	12	9	0	1
5	R270X	5	30	5	0	0
6	deletion	11	27	8	1*	0
7	deletion	9	18	6	0	1
8	R168X	7	15	9	1*	1
9	R255X	10	2	14	0	1
10	C916T	4	24	7	0	0
11	R168X	4	15	7	1	0
12	deletion	11	18	6	0	3
13	T158M	12	12	11	1*	1
14	T158M	4	13	4	1*	0
15	R168X	9	36	10	0	0
16	R294X	5	NA	7	0	0
17	R168X	4	32	11	1*	0
18	R168X	2	6	9	1	0
19	T158M	9	18	12	1*	1
20	deletion	9	18	9	1*	1
21	R270X	7	12	10	0	0
22	R133C	8	18	13	0	1
23	deletion	8	30	7	1*	1
24	P450L	6	18	5	0	0
25	deletion	11	33	8	0	0
26	T158M	3	22	9	0	0
27	R168X	5	10	7	1*	1
28	P322S	12	15	5	1*	0
29	R168X	10	36	10	0	1
30	R294X	4	14	8	0	0
31	P152A	6	12	6	1*	0
32	P152R	11	12	11	0	1

[a]RSSS, the summary score of the expanded Rett Syndrome Severity Scale,[15] comprises clinical ratings on seven parameters (seizure frequency/manageability, respiratory irregularities, scoliosis, ability to walk, hand use, speech, and sleep problems). Each parameter is rated on a 4-point Likert scale from 0 (absent/normal) to 3 (severe).

[b]Walking: 0, no walking; 1, unsupported walking; 1*, walking with support.

[c]Seizures (subscale of RSSS): 0, absent; 1, mild; 2, moderate; 3, severe

Table 2

Latency of First Look to the Target (ms)

		Number of Distractors					
		One		Two		Three	
Group	N	M	SD	M	SD	M	SD
Rett Syndrome							
Younger (2-7 yrs)	16	2628	1648	2377	1561	3113	2626
Older (8-12 yrs)	16	2868	1620	2473	1895	2937	2329
Typically Developing							
Younger (2-7 yrs)	16	1738	891	1568	1127	1793	1037
Older (8-12 yrs)	16	744	410	688	412	729	473

Table 3

Time Spent Looking at the Target (%)

		Number of Distractors					
		One		Two		Three	
Group	N	M	SD	M	SD	M	SD
Rett Syndrome							
Younger (2-7 yrs)	16	26.84	10.12	18.98	11.21	21.16	12.88
Older (8-12 yrs)	16	29.23	16.53	26.62	15.91	24.26	11.34
Typically Developing							
Younger (2-7 yrs)	16	52.12	16.91	55.40	18.19	51.00	16.30
Older (8-12 yrs)	16	72.63	11.72	72.40	16.58	69.93	16.11

Table 4

Number of Looks Away from the Target to the Distractors (per s)

		Number of Distractors					
		One		Two		Three	
Group	N	M	SD	M	SD	M	SD
Rett Syndrome							
Younger (2-7 yrs)	16	1.44	1.30	1.33	0.73	2.19	1.60
Older (8-12 yrs)	16	1.80	1.50	1.94	1.93	2.22	1.75
Typically Developing							
Younger (2-7 yrs)	16	0.85	0.56	0.74	0.45	0.98	0.50
Older (8-12 yrs)	16	0.51	0.23	0.54	0.39	0.67	0.41

Table 5

Time to Re-engage Attention to the Target (msec)

		Number of Distractors					
		One		Two		Three	
Group	N	M	SD	M	SD	M	SD
Rett Syndrome							
Younger (2-7 yrs)	16	3680	1776	5691	2142	3908	1786
Older (8-12 yrs)	16	3650	1879	4610	2624	3558	1822
Typically Developing							
Younger (2-7 yrs)	16	1473	807	1494	1092	1352	818
Older (8-12 yrs)	16	647	304	863	701	689	353