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Attention and identification of the same and the similar visual stimuli in Rett Syndrome

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Abstract

Rett Syndrome (RTT) is a developmental disorder, predominantly affecting girls, which causes Intellectual Disability and neuro-behavioral disability. Individuals with RTT present with apraxia and movement disorders and most of them are unable to speak, walk and use their hands. For these reasons, eye tracker technology is being increasingly used to their assess cognitive processes.

The aim of this study was to investigate three cognitive processes in girls with RTT compared with typical developing girls (TD): the ability to attend to visual stimuli, the ability to identify the same stimuli and the ability to identify the similar stimuli. With the help of Eye Tracker technology, three tasks were administered (1. Attention; 2. Identification of the same stimuli; 3. Identification of the similar stimuli) to 21 girls with RTT, compared with TD girls. Results show that girls with RTT performed worse than girls with TD in all conditions. The absence of

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Compliance with Ethical Standards: The authors declare no conflict of interest and no source of funding for the present research. All procedures performed in this study involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. Informed consent was obtained from all participants' parents recruited for the study.

significant interactions between group and type of task and between groups and contents indicates that the attention behavior in girls with RTT can reflect a general delay in cognitive maturation.

Keywords: Rett Syndrome; Attention; Visual stimuli; Identification of visual stimuli.

1. Introduction

Rett Syndrome (RTT) is characterized by cognitive and neuromotor impairments, including loss of communicative skills. It is very difficult to complete a valid assessment of girls with RTT; severe physical, language, and social impairments impose a wide range of limitations (Demeter, 2000; Kaufmann, Tierney, Rohde, Suarez-Pedraza, Clarke, Salorio *et al.*, 2012; Bergström-Isacsson, Lagerkvist, Holck, & Gold, 2014; Fabio, Colombo, Russo, Cogliati, Masciadri, Foglia *et al.*, 2014). Most studies on cognitive functions and behavioral features in RTT were based on information provided by parents and professional carers (Reid & Green, 2002). Currently, several studies have attempted to assess RTT cognitive processes through new technologies (Djukic, Valicenti McDermott, Mavrommatis, & Martins, 2012; Castelli, Antonietti, Fabio, Lucchini, & Marchetti, 2013; Gangemi, Fabio, Caprì, & Falzone, 2017; Fabio, Gangemi, Caprì, Budden, & Falzone, 2018; Fabio, Magaudda, Caprì, Towey, & Martino, 2018; Fabio, Martino, Caprì, Giacchero, Giannatiempo, La Briola *et al.*, 2018; Gangemi, Caprì, Fabio, Puggioni, Falzone, & Martino, 2018).

Fabio, Antonietti, Castelli and Marchetti (2009) carried out a study with 20 children (10 girls with RTT and 10 controls, matched on mental age), who were tested on both attention and non-verbal communication abilities. As far as attention procedures were concerned, children were asked to carefully look at a "target" toy placed in the middle of a table and then sort it among other distractor stimuli. Results showed that girls with RTT focused their attention on the target toy for a shorter interval and tended to look at the caregiver's face for a longer interval than controls. Baptista, Mercadante, Macedo and Schwartzman (2006), through eye tracking technology, presented two stimuli at a time, a target and a distractor, and asked the child to choose the target stimulus during different cognitive tasks. Girls with RTT reported high rates of correct answers, thus suggesting that their intentional gaze is measurable and can be used to explore their cognitive performances. In another study (Velloso, de Araujo, & Schwartzan, 2009), 10 girls (aged 4-12 years) were assessed with a computer system for visual tracking for their ability of indicating the recognition of color (red, yellow and blue), shape (circle, square and triangle), size (big and small) and spatial position (over and under) with their eyes. Results comparing the time of eye fixation on required and not required concepts did not differ significantly. Children did not seem to recognize the required concepts when assessed with eye tracking system. Authors recommended a reconsideration of the

method, since the small group of 10 girls was not able to recognize the required concepts within a fixation time interval of 4 seconds, although the authors argued that the young age of the sample and the brief fixation span might have interfered with results. Results from other studies, however, confirmed that eye gaze technology can be used to explore RTT cognitive performance (Vignoli, Fabio, La Briola, Giannatiempo, Antonietti, Maggiolini *et al.*, 2010; Stasolla, Perilli, Di Leone, Damiani, Albano, Stella *et al.*, 2015; Fabio, Billeci, Crifaci, Troise, Tortorella, & Pioggia, 2016).

Indeed, not only individuals with RTT demonstrated to be capable of attending to stimuli, but they succeeded in learning new skills as well (Leonard, Fyfe, Leonard, & Msall, 2001). Similar capabilities have been described in girls who had been taught how to make choices (Baptista *et al.*, 2006; Fabio, Castelli, Antonietti, & Marchetti, 2013; Fabio, Gangemi *et al.*, 2018; Fabio, Magaudda *et al.*, 2018) as well as in girls who succeeded in identifying abstract concepts when taught in a motivating context with the help of a computer-based interactive language development system (Hetzroni, Rubin, & Konkol, 2002; Antonietti, Fabio, Boari, & Bonanomi, 2005). A work by Fabio, Giannatiempo, Antonietti and Budden (2009) showed that patients with RTT were able to learn how to discriminate complex stimuli. Girls executed an attention task in which a complex stimulus was shown, followed by individual stimuli presented together with distractors. Results showed that girls could learn to discriminate complex stimuli. Recently, Rose, Djukic, Jankowski, Feldman and Rimler (2016) also examined different aspects of attention in children with RTT, using eye tracking. The authors evaluated the shifting and sustaining attention in two groups (RTT and typically developing as the comparison group), using a task in which an attractive central stimulus was followed, after a short gap, by a dynamic target presented in one side. Time to shift to the target location (reactive and anticipatory saccades) and time of eye fixation of the target were assessed. Results indicated that children with RTT were consistently slower in shifting attention and their reactive saccades were also slower than those of typically developing children. However, results didn't reach statistical significance. Also, it was found that RTT group spent considerable time looking at the target, although this time was less significant than that of the typically developing group. The study by Rose and colleagues (2016) demonstrated that children with RTT could maintain attention on a stimulus and orient relatively quickly to it, although they showed some difficulties in anticipating predictable events and in endogenous attention.

The aim of this study was to investigate more in depth the attention processes and the abilities of girls with RTT to identify the same and the similar visual stimuli. The cognitive skills were evaluated through three types of tasks: an attention task; a task for the identification of the same stimulus; and a task for the identification of the similar stimulus.

With regard to the attention task, girls were requested to look at visual stimuli of fruits, animals, people and emotions. In the second and third tasks, that is identification of the same and the similar stimuli respectively, girls had to comprehend two levels of abstract concepts ("sameseness" and "similarity"). In the first one, each girls were presented with only one stimulus and they had to look at it (for example, "look at the grapes"); in the second task, girls were presented with 3 stimuli (one target stimulus and two additional stimuli) and were requested to match one of the two additional stimuli with the target stimulus ("which one of these two is the same as the grapes"); in the third task, girls were once again presented with three stimuli but this time they had to choose the stimulus that fell into the same category as the target one ("which one of these two is similar to the target stimulus").

It was expected that girls with RTT would easily perform the attention task, while both tasks for the identification of the same and the similar stimuli might be harder, since the identification of similar stimuli is a more abstract concept and a more demanding cognitive task.

2. Method

2.1. Participants

Twenty-one girls, aged between 6 and 21 years (M = 12.7; SD = 4.1), with a clinical diagnosis of classical RTT (Fabio, Martinazzoli, & Antonietti, 2005; Neul, Kaufmann, Glaze, Christodoulou, Clarke, BahBuisson *et al.*, 2010) were classified into two clinical conditions: III and IV. RTT was genetically confirmed in all patients. Analyses of MCP2 mutation were conducted in the laboratories of the Istituto Auxologico Italiano. MCP2 specific mutations, cognitive and demographic information are shown in Table 1.

All participants presented with cognitive and physical disabilities. They all had severe intellectual disability; one girl (the only variant) was speech-preserved, while the other girls had not verbal speech. All the girls showed little or no purposeful hand use and striking pervasive hand stereotypies. When properly supported, some girls succeeded in walking, although with

difficulties. The sample girls, aged 7-14 years, were attending nursery, primary or secondary schools; older girls (14-21 years) attended day care centers. Families of each of the girls were contacted by the Italian Rett Syndrome Association and were asked to participate in the study. Written consent was obtained for all participants.

A comparison group with 20 typically developing (TD) girls was recruited among healthy children accompanying patients referred to pediatric specialty clinics. Their chronological age ranged from 2 to 7 years (M = 3.7; SD = 4.2). Mental age differences between groups, as evaluated through the Vineland Scales (Sparrow, Balla, & Cicchetti, 1984), were not significant, $t(38) = .75$, $p = .54$. The TD group was further screened to exclude any girls with significant neurological disorders, sensory impairments, neurodevelopmental disorders and disabilities.

Table 1 - Sample characteristics

	Name	Age	Mutation MeCP2	RARS[1] score	Level of severity	VABS[2] score
1	D. D.	18	R306C	55.8	Moderate	82
2	R. E.	18	/	55.0	Moderate	58
3	D. S.	21	R306C	61.5	Moderate	110
4	T. M.	10	/	70.5	Moderate	89
5	G. A.	7	1156 del 44	43.5	Mild	291
6	E. M.	14	R306C	64.5	Moderate	116
7	S. V.	7	1156 del 44	47.0	Mild	118
8	G. A.	10	/	56.5	Moderate	169
9	A. V.	11	/	64.5	Moderate	123
10	I. E.	12	R255X	66.5	Moderate	107
11	G. S.	19	R306C	63.5	Moderate	142
12	V. A.	8	R306C	49.0	Mild	121
13	D. C. N.	19	R294X	66.0	Moderate	83
14	N. G.	11	R168X	85.0	Severe	34
15	P. E.	19	/	65.0	Moderate	126
16	V. S.	12	R270X	74.0	Moderate	84
17	R. A.	11	/	61.5	Moderate	131
18	F. M.	8	R133C	64.0	Moderate	110
19	Z. F.	16	R255X	64.0	Moderate	113
20	A. I.	16	P322A	60.0	Moderate	134
21	B. M. R.	14	R106W	77.0	Moderate	92

[1] Composite Rett Assessment Rating Scale (RARS) score (total score from communication, daily living skills, socialization, and motor skills domains).
[2] Composite adaptive behavioral scores (Vineland total score from communication, daily living skills, socialization, and motor skills domains).

2.2. Materials

2.2.1. Eyegaze device

An eyegaze device was used to record participants' visual scanning responses to visual computer screen stimulation. This device records ocular movements, such as location and duration of eye fixations (pause of eye movements on an object of interest) and saccadic movements (rapid movements between fixations (Salvucci & Goldberg, 2000). Eyegaze device was installed in a 15-inch LCD-monitor computer with a Matrix-like video plate, which captures signals sent from a video camera equipped with lenses sensitive to high-speed infrared light. Moreover, the camera has a LED that emits low-intensity infrared light directly onto the retina of the person sitting in front of the computer. The direction of the gaze was determined according to the Pupil Center/Corneal Reflection Method. The Passive Gaze Tracing software (LC Technologies, Inc., Sao Paulo, Brazil) was used to generate eye found flag, gaze point, pupil diameter during visual scanning, and eyeball position.

2.2.2. Attention and identification paradigm

A special setting was arranged for participants' cognitive activities, and all distracting stimuli were removed so that they could only focus on the task. Each girl was invited to seat in front of the eye tracker, at a distance of about 30 cm, and the calibration procedure was started, in order to establish the exact point that she was fixing on the computer screen. During the calibration session, participants are requested to fixate 5 points on the screen, while the eye tracker monitors their eyes' movements. In our study, the first task was to pay attention to animals (dog, horse, cat); fruits (banana, apple, grapes); emotions (happy, sad, angry).

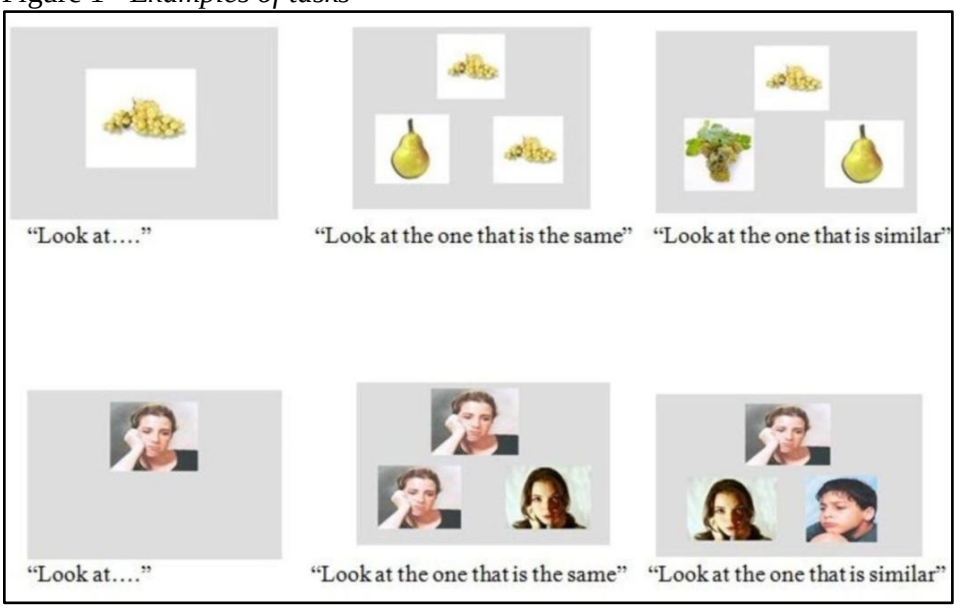
Nine stimuli were presented, 3 figures of the animal category (dog, horse, cat); 3 figures of the fruit category (banana, apple, grapes); 3 figures of the emotion category (happy, sad, angry).

In the first task, examiner asked each girl to look at the figure of a cat on the screen of the Eye tracker: "Where is the cat? Please, look at the cat". The stimulus appeared alone, without other stimuli; the girl had to look at it. Each image remained on the screen for 10 seconds. In the second task (identify the same), three figures were presented on the screen: two identical figures of the cat and one distractor (bird): the examiner asked the girl to

look at the figures that were identical "Please, look at the cat that is the same as this one!".

In the third task, three figures were presented: the target (cat), the figure of another type of cat and the distractor, "Please, look at the cat that is similar to this one!" (Fig. 1)

Figure 1 - *Examples of tasks*



After the animal visual stimuli, participants continued with fruits and, finally, emotions visual stimuli. Each stimulus was shown for 5 times, with a time interval between figures of 10 seconds. The overall duration of the experimental session was about 30 minutes.

3. Results

Results were analyzed based on the two neuropsychological parameters: fixation length (FL) and fixation count (FC). With regard to FL, a repeated measure analysis of variance subject 2 X 3 X 3 was applied, with the first "group" factor being "between subjects", while the second and the third being "within subjects": groups (RTT vs TD); contents (figures of fruits, animals and emotions) and type of task (attention, identification of the same, identification of the similar; see Tab. 2).

Table 2 - *Means and standard deviations of target fixation length for the three tasks*

Group	Type of task	Fruits		Animals		Emotions	
		M	*(SD)*	*M*	*(SD)*	*M*	*(SD)*
RTT	Attention	4.47	(3.26)	5.77	(4.95)	4.48	(3.16)
	Identification of the same	2.64	(2.76)	3.70	(4.41)	1.40	(1.63)
	Identification of the similar	1.99	(2.25)	2.95	(3.10)	2.08	(2.17)
TD	Attention	6.93	(1.28)	7.01	(3.20)	6.98	(1.28)
	Identification of the same	5.83	(1.29)	6.04	(3.10)	5.32	(1.23)
	Identification of the similar	5.01	(1.25)	5.60	(3.10)	5.71	(1.17)

As far as the FL is concerned, the "group" factor showed a significant effect, $F_{(1, 38)} = 3.22$, $p < .01$, $d = .75$. TD group performed better than RTT group. The "type of task" factor showed significant statistical effects $F_{(2, 38)} = 10.91$, $p < .001$, $d = .88$. For all participants, the attention task was easy, while the identification of the same and the identification of the similar were more complex tasks.

As for FC, the same repeated measure analysis of variance was applied. Again, the "group" factor showed a significant effect $F_{(1, 38)} = 2.99$, $p < .01$. The "type of task" factor showed also significant statistical effects $F_{(2, 38)} = 8.91$, $p < .001$ (see Tab. 3). Contents, on the contrary, had no significant effect.

Table 3 - *Means and standard deviations of target fixation count in different types of tasks*

Group	Type of task	Fruits		Animals		Emotions	
		M	*(SD)*	*M*	*(SD)*	*M*	*(SD)*
RTT	Attention	12.06	(8.80)	14.06	(10.94)	13.88	(7.53)
	Identification of the same	10.18	(11.66)	11.73	(11.30)	7.29	(9.28)
	Identification of the similar	8.00	(8.31)	8.75	(8.73)	6.81	(6.51)
TD	Attention	15.07	(6.40)	17.32	(5.49)	15.98	(4.52)
	Identification of the same	12.40	(5.60)	15.21	(5.32)	9.98	(5.48)
	Identification of the similar	9.91	(6.21)	10.30	(6.87)	8.97	(4.35)

4. Discussion

The aim of this study was to analyze the attention processes and the abilities of girls with RTT implied in the identification of the same and the similar stimuli. Using an eye-tracking paradigm, girls with RTT and girls with TD – as the comparison group – were tested on three tasks: (1) attention task; (2) identification of the same stimuli; (3) identification of the similar stimuli.

Results showed that RTT group performed worse than the TD group in all conditions. The absence of significant interactions between group and type of task and between groups and contents indicates that the attention behavior of the RTT individuals is similar to that of age-matched TD group, thus showing a general delay in cognitive maturation.

Our results are in line with previous studies demonstrating that the performance of patients with RTT is significantly poorer than that of the TD comparison group (Djukic *et al.*, 2012; Rose, Djukic, Jankowski, Feldman, Fishman, & Valicenti-Mcdermott, 2013; Djukic, Rose, Jankowski, & Feldman, 2014; Rose *et al.*, 2016).

This study has generalized findings to the identification of the same and similar stimuli. It turned out that girls with RTT can attend, just like normally developing children, to visual stimuli, although their visual scanning is poorer and delayed. Indeed, RTT group turned out to look at the stimulus for a shorter interval than TD group, although the way they attend to it is was the same.

This study has some treatment implications. Since vision and gaze are the most important ways in which people with RTT relate to the world (Hagberg, 2002; Velloso *et al.*, 2009; Didden, Korzilius, Smeets, Green, Lang, Lancioni *et al.*, 2010; Berger-Sweeney, 2011; Djukic & Valicenti McDermott, 2012; Stasolla *et al.*, 2015; Fabio, Magaudda *et al.*, 2018; Fabio, Caprì, Nucita, Iannizzotto, & Mohammadhasani, 2019), attention training is necessary to help individuals with RTT to identify the similar visual stimuli. Given that girls with RTT are usually nonverbal and show a very limited ability to use their hands, and because they can use vision and gaze, we can conclude that the characteristics of their eye gaze can be used as basic skills to develop complex abilities, for example the identification of the same and similar stimuli.

In conclusion, this study provides additional information relating to attention processes of girls with RTT, that appear to be important predictors of cognitive complex performance.

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