

## Choice behavior in Rett Syndrome: The consistency parameter

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### Abstract

*Background: The opportunity to make choices is important for improving the quality of life and increasing the sense of control experienced by people with neurodevelopmental disorder. One of the most important criteria to understand the ability to make choices is the consistency parameter. The aim of this study was to analyse the consistency of choice behaviour in girls with Rett Syndrome (RTT). Methods: Twenty-four girls (13 with RTT and 11 controls, matched on mental age and gender) were tested with stimuli at three different levels of difficulty: with actual items, pictures of the items, and written name items. Results: Consistency of choice index was calculated to compare the performances of girls with RTT and controls. The group factor shows no significant effect. Girls*

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*with RTT make choices consistently as the control group. Discussion: This study has shown that individuals with profound neurodevelopmental disorders, as the girls with RTT, are able to make choices keeping consistency in their choices.*

**Keywords:** Rett Syndrome; Choice ability; Consistency of choice; Cognitive abilities; Quality of life.

## 1. Introduction

Within the context of regular daily life, individuals make dozens of choices ranging from what to eat to what to wear (Lancioni, O'Reilly, & Emerson, 1996; Djukic & Valicenti McDermott, 2012; Stasolla, Caffò, Perilli, Boccasini, Damiani, & D'Amico, 2018). The opportunity to make choices is important both to improve the quality of life and to increase the sense of control experienced by persons with developmental disabilities (Conyers, Doole, Vause, Harapiak, & Yu, 2002; Fabio, Antonietti, Marchetti, & Castelli, 2009; Fabio, Giannatiempo, Antonietti, & Budden, 2009; Vignoli, Fabio, La Briola, Giannatiempo, Antonietti, Maggiolini *et al.*, 2010; Rose, Djukic, Jankowski, Feldman, & Rimler, 2016).

Through rigorous assessments many studies show that individuals with intellectual disabilities demonstrate clear preferences and can make choices that results in increased appropriate behavior and decreased challenging behavior (Lancioni *et al.*, 1996; Cannella, O'Reilly, & Lancioni, 2005; Fabio, Gangemi, Capri, Budden, & Falzone, 2018). However, it is often not easy to enhance the choice ability in children with severe developmental disabilities. One of the procedures used in the assessment to enhance their ability to show the preferences is the consistency of choice. The consistency of choice behavior may be very important in people with severe deficits in communication like girls with Rett syndrome.

Rett Syndrome (RTT) is a severe neurodevelopmental disorder occurring almost exclusively in females, with a frequency of 1:10.000-15.000. RTT is caused by mutations in X-linked *MECP2* (Amir, Van den Veyver, Wan, Tran, Francke, & Zogh, 1999; Fabio, Castelli, Marchetti, & Antonietti, 2013; Fabio, Colombo, Russo, Cogliati, Masciandri, Antonietti, & Tavian, 2014). Clinical presentation of classical RTT usually occurs in the first years of life with regression of early-acquired developmental skills, such as speech and motor functions, and, in particular, loss of purposeful hand use (Hagberg, 2002). The communicative functions are often impaired in these girls, but one important distinguishing feature is their intense eye communication, preserved throughout adulthood.

The knowledge of the preferences and the evaluation methodologies can be useful in girls with RTT for many reasons. First, by identifying objectively the preference for a variety of stimuli may allow for easy identification of elements that can have a high degree of effectiveness as reinforcement (Cannella *et al.*, 2005). Second, it allows people who have limited communication skills to help teachers and other operators to

determine which stimuli are used as possible reinforcement or to assist in the selection of favorite leisure activities. Moreover, the objective measures of the preference may be a more accurate method of determining stimulus preference (Green, Reid, White, Brittain, & Gardner, 1988). The purpose of the present study was to examine whether girls with RTT with different discrimination skills can make consistent choices among alternatives in the same way as normally developing girls. The second aim was to analyze if the higher level of abstraction (from actual items, to pictures of the items and to written name presentation of the items) influences their level of consistency.

## 2. Method

### 2.1. Participants

Thirteen girls with RTT (aged 5 to 12 years) participated in the experiment. According to the paperwork provided by the parents, 12 patients examined for *MECP2* mutations were positive to the molecular analysis of *MECP2*. One girl had not yet been confirmed for the mutation, but she presented many characteristics of the syndrome. Their families had been contacted by the Italian Rett Association (AIR). The Institutional Review Board of the University of Messina approved this study (prot. 34/2017). The parent sign consent forms.

A licensed psychologist carried-out a general assessment, through both the Vineland Adaptive Behavior Scale (VABS) (Sparrow, Balla, & Cicchetti, 1984) and the Portage Scale (Bluma, Shearer, Frohman, & Hilliard, 1976). Only three girls with RTT (D. D., C. F. and S. R.) were able to read and to discriminate the written word.

Eleven normally developing girls (aged 18 to 30 months) attending a public nursery school in a town of Northern Italy constituted the control group. They were matched for mental age with RTT girls. The psychologist carried out the same assessment procedure based on the VABS and the Portage Scale to match the two groups.

Table 1 A/B shows the chronological age of the participants and their mental age assessed through both the Portage Scale and the Vineland Adaptive Behavior Scales (VABS).

**Table 1A - Characteristics of the participants: Rett Syndrome group**

	D. D.	C. F.	A. D.	L. L.	S. R.	E. C.	N. C.	S. C.	S. C. C.	A. C.	C. F.	A. D.	L. L.	B.
Age (years)	12.5	26	6	17	15	6.5	5.5	14	5	26	7	15	12	
Mental age (months)	27	22	19	24	28	19	24	23	19	22	18	23	21	
VABS (total score)	99	96	77	88	96	86	122	94	95	96	75	88	87	
VABS subscales														
<i>Communication</i>	30	28	24	27	31	29	30	32	29	21	23	26	28	
<i>Daily ability</i>	22	20	19	23	26	22	22	20	23	20	19	23	22	
<i>Socialization</i>	19	17	20	24	22	24	30	27	29	17	20	24	22	
<i>Motor ability</i>	30	31	14	14	23	11	27	14	14	31	14	14	14	
Portage subscales														
<i>Language</i>	8	7	8	10	10	11	13	11	11	7	5	10	10	
<i>Socialization</i>	20	16	16	21	25	25	32	29	29	16	16	21	26	
<i>Autonomy</i>	12	9	7	9	12	8	8	10	8	9	7	9	8	
<i>Cognitive level</i>	22	20	11	17	21	12	20	20	16	20	11	17	12	
<i>Motor level</i>	25	25	12	24	29	22	29	16	18	25	12	24	18	

**Table 1B - Characteristics of the participants: Normally developing group**

	G. C.	B. S.	I. V.	C. C.	S. R.	G. R.	M. N.	N. N.	E. N.	B. S.	I. V.
Age (years)	18	27	28	20	21	27	28	27	22	26	18
Mental age (months)	21	29	29	22	22	25	27	26	24	28	21
VABS (total score)	154	202	200	172	176	186	192	190	180	202	155
VABS subscales											
<i>Communication</i>	43	56	52	45	44	50	50	48	46	56	44
<i>Daily ability</i>	35	50	48	41	45	47	46	46	46	50	37
<i>Socialization</i>	42	56	59	50	51	48	54	52	50	56	40
<i>Motor ability</i>	34	40	41	36	36	41	42	44	38	40	34
Portage subscales											
<i>Language</i>	23	40	38	25	27	32	35	33	30	40	24
<i>Socialization</i>	40	48	50	45	42	41	48	46	38	48	39
<i>Autonomy</i>	19	29	30	21	23	24	29	28	22	29	20
<i>Cognitive level</i>	22	38	36	26	25	34	34	36	29	38	23
<i>Motor level</i>	64	74	75	66	62	70	72	70	69	74	61

## 2.2. Procedure

Observations of the present study were collected at the patients' houses and in the nursery for the control group. In the first phase the examiner evaluated the mental age and the adaptive behavior on the Portage and Vineland Adaptive Behavior Scale through an interview with parents and educators. Caregivers were also asked to generate a list of girls' hobbies and known food. During the experiment they were asked to sit directly behind their girl and not to initiate interactions with the girl during the procedure. The examiner presented the test. For each girl a session of 20 min duration was conducted. The task was carried out in three phases. Table 2 A/B shows the preferred items for each girl.

**Table 2A - Items used: Girls with Rett Syndrome**

Name	Food / Drinks	Hobbies
D. D.	Banana / Peach juice	Carillon / Game 1
C. F.	Yogurt / Kiwi	Book 1 / Book 2
A. D.	Yogurt / Strawberries	Rag doll / Peluche
L. L.	Fruit juice / Milk	Carillon / Peluche
S. R.	Yogurt / Brioche	Doll / Rag doll
E. C.	Peach juice / Yogurt	Carillon / Game
N. C.	Chocolate / Yogurt	Doggie / Doll
S. C.	Pear / Yogurt	Teddy bear / Carillon
S. C. C.	Brioche / Pudding	Bear / Frog
A. C.	Banana / Peach juice	Carillon / Game 1
C. F.	Yogurt / Kiwi	Book 1 / Book 2
A. D.	Yogurt / Strawberries	Rag doll / Peluche
L. L. B.	Yogurt / Tea	Rag doll / Peluche

**Table 2B - Items used: Control group**

Name	Food / Drinks	Hobbies
G. C.	Pear / Banana	Game 1 / Game 2
B. S.	Apple / Pear	Doll / Peluche
I. V.	Peach / Banana	Rag doll / Book
C. C.	Pear / Banana	Game / Doll
S. R.	Apple / Banana	Rag doll / Doll

G. R.	Apple / Pear	Teddy bear/ Book
M. N.	Pear/ Peach	Xylophone / Peluche
N. N.	Banana / Pear	Doll / Ball
E. N.	Banana / Peach	Ball / Rag doll
B. S.	Apple / Pear	Doll / Peluche
I. V.	Peach / Banana	Rag doll / Book

In the first session, the child and experimenter sat facing one another at a small table. In the present study each interaction was videotaped so that the upper front half of the child and the profile of the experimenter were in view. Caregivers were asked to sit directly behind their child and not to initiate interactions with the child during the procedure. The examiner placed a pair of concrete objects on the small table in front of the girl, to the right and left of her. Location of the items on the right or on the left was presented in a random order. The items were placed 40 cm from each other, in this way both items were observable and within an easy reach and grasp of the girls. Each session consisted of five choices between two items (food and/or drink and hobbies). The examiner asked participants to choose an item and he was asked “Which do you prefer?” and waited for the answer for 75 seconds.

The participants could answer by grasping the item or by looking at it. The answer was considered accepted if the girl with RTT grasped at it and/or looked at it for 3 seconds. If the girl looked alternately at both stimuli, the behavior was coded as “no answer”.

Table 3 shows the choice modality and the number of choices for each item. If the participant chooses one item during the 75 seconds, this was given to her as a reinforcer. A new possibility of choice started after 10-15 seconds.

Table 3 - *Modality and number of choices of the girls with RS*

Name	Answer Mode	Food / Drinks			Hobbies		
		Item 1	Item 2	No answer	Item 1	Item 2	No answer
D. D.	Grasping	2	13	0	15	0	0
C. F.	Looking	0	10	5	0	15	0
A. D.	Looking	3	6	1	10	0	0
L. L.	Looking	10	0	0	10	0	0
S. R.	Grasping	0	10	0	15	0	0

E. C.	Looking	0	10	0	0	10	0
N. C.	Grasping	10	0	0	0	10	0
S. C.	Grasping	0	10	0	0	10	0
S. C. C.	Looking	0	10	0	10	0	0
C. F.	Looking	0	10	5	0	15	0
A. D.	Looking	3	6	1	10	0	0
L. L. B.	Looking	10	0	0	10	0	0

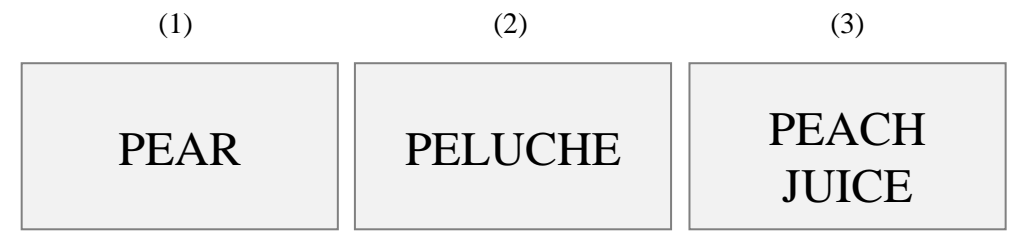
In the second session the examiner used the pictures of the items as choice items, with cards of about 15×20 cm, reporting individual objects and hobbies (Fig. 1).

Figure 1 - *Examples of pictures*



The third session takes place only if the participants were able to read. In this case the items were written stimuli representing the concrete objects used in the first session (Fig. 2).

Figure 2 - *Examples of written stimuli*





### 3. Analysis of data

To calculate the interobserver agreement two independent observers coded each videotaped interaction. The interobserver agreement was used to estimate the grasping and the looking behaviors. Interobserver agreement was calculated by dividing the number of agreements by the number of agreements plus disagreements and converting this number to a percentage. Agreement was defined as both observers scoring the same selection for the trial. Agreement data was higher than 89%.

With reference to statistical analysis, as the performance of the control group was not normally distributed, non-parametric statistics were used. To compare performances on the consistency indexes, a non-parametric ANOVA Kruskal-Wallis (1952) and Wilcoxon (1945) was used both for main effect and for multiple comparisons. Participants were tested three times with three types of stimuli (with actual items, pictures of the items, and written name presentation) to determine whether the choices were consistent across trials.

To calculate the consistency index (CI) the formula here used is:

$$CI = \frac{n}{\max n} \times \frac{a}{\max a} \times 100$$

Where  $n$  is the number of items selected in a consistent way,  $\max n$  is the maximum number of possible consistent items,  $a$  is the number of acceptance behavior and  $\max a$  is the maximum number of possible acceptances. For example, selecting the same item from the same-paired items throughout the three sessions and accepting it always resulted in a consistency of the 100%.

### 4. Results

Table 4 A/B shows the number of choices for each item and the number of acceptances and no replies of the related stimuli.

Table 4A - *Number of choices for each item and number of acceptances (Acc.) and no replies (N. R.) of the related stimuli: Girls with Rett Syndrome*

Name	Food/drink choice											
	Concrete stimuli				Pictures				Written stimuli			
	Item 1	Item 2	N. R.	Acc.	Item1	Item2	N. R.	Acc.	Item1	Item2	N. R.	Acc.
D. D.	1	4	0	5	0	5	0	5	1	4	0	5
C. F.	0	5	0	5	0	5	0	5	0	0	5	0
A. D.	1	4	0	5	2	2	1	4				
L. L.	5	0	0	5	5	0	0	5				
S. R.	0	5	0	5	0	5	0	5	0	4	1	4
E. C.	0	5	0	5	0	5	0	5				
N. C.	5	0	0	5	5	0	5	0				
S. C.	0	5	0	5	0	5	0	5				
S. C. C.	0	5	0	5	0	5	0	5				
A. C.	5	0	0	5	5	0	0	5				
C. F.	0	5	0	5	0	5	0	5				
A. D.	1	4	0	5	2	2	1	4				
L. L. B.	5	0	0	5	5	0	0	5				

Name	Hobbies choice											
	Concrete stimuli				Pictures				Written stimuli			
	Item 1	Item 2	N. R.	Acc.	Item1	Item2	N. R.	Acc.	Item1	Item2	N. R.	Acc.
D. D.	5	0	0	5	5	0	0	5	5	0	0	5
C. F.	0	5	0	5	0	5	0	5	0	5	0	5
A. D.	5	0	0	5	0	5	0	5				
L. L.	5	0	0	5	5	0	0	5				
S. R.	5	0	0	5	5	0	0	5	5	0	0	5
E. C.	0	5	0	5	0	5	0	5				
N. C.	0	5	0	5	0	5	0	5				
S. C.	0	5	0	5	0	5	0	5				
S. C. C.	5	0	0	5	5	0	0	5				
L. L.	0	5	0	5	0	5	0	5				
A. C.	0	5	0	5	0	5	0	5				
C. F.	5	0	0	5	0	5	0	5				
L. L.	5	0	0	5	5	0	0	5				
A. D. G.												

Table 4B - *Number of choices for each item and number of acceptances (Acc.) and no replies (N. R.) of the related stimuli: Control group*

Name	Food/drink choice							
	Concrete stimuli				Pictures			
	Item 1	Item 2	N. R.	Acc.	Item1	Item2	N. R.	Acc.
G. C.	5	0	0	5	5	0	0	5
B. S.	0	5	0	5	0	5	0	5
I. V.	5	0	0	5	5	0	0	5
C. C.	0	5	0	5	0	5	0	5
S. R.	5	0	0	5	5	0	0	5
G. R.	5	0	0	5	5	0	0	5
M. N.	5	0	0	5	5	0	0	5
N. N.	0	5	0	5	0	5	0	5
E. N.	5	0	0	5	5	0	0	5
A. L.	5	0	0	5	5	0	0	5
I. V.								

Name	Hobbies choice							
	Concrete stimuli				Pictures			
	Item 1	Item 2	N. R.	Acc.	Item1	Item2	N. R.	Acc.
G. C.	0	5	0	5	0	5	0	5
B. S.	0	5	0	5	0	5	0	5
I. V.	5	0	0	5	5	0	0	5
C. C.	0	5	0	5	0	5	0	5
S. R.	5	0	0	5	5	0	0	5
G. R.	0	5	0	5	0	5	0	5
M. N.	0	5	0	5	0	5	0	5
N. N.	5	0	0	5	5	0	0	5
E. N.	5	0	0	5	5	0	0	5
A. L.	0	5	0	5	0	5	0	5
I. V.								

The girls of the control group always chose the same item in all conditions, that is, in all cases they obtained a “ceiling effect”. Because only three girls with RTT can discriminate written stimuli, only for these girls their data are based on 15 and not 10 stimuli.

The data of consistency indices were analyzed according to ANOVA Kruskal-Wallis (1952) and Wilcoxon (1945), with one between subject

factor and two within subject factors: 2 (Group: girls with RTT vs. control girls)  $\times$  2 (Type of discrimination: actual vs. picture objects)  $\times$  2 (Contents: food or drinks vs. hobbies). The Group factor showed no significant effect ( $H = 1.62, p = .44$ ). The Type of discrimination factor also showed no significant effect ( $H = .99, p = .63$ ). The Content showed also no significant effect ( $H = .76, p = .54$ ). No group interaction was also significant.

## 5. Discussion

The results of the present study show that the girls with RTT can make consistent choices among alternatives in the same way of the normally developing girls. They continue to have a high level of choice consistency with the actual items, with the pictures of the items and three girls with written-name presentation. The consistency may be used in girls with Rett syndrome as an assessment methodology to enhance their ability of choice. The application of preference assessment methodologies is a practice that is supported by ample research and a number of empirical literature reviews (Lancioni *et al.*, 1996; Lohrmann-O'Rourke & Browder, 1998; Cannella *et al.*, 2005; Fabio, Billeci, Crifaci, Troise, Tortorella, & Pioggia, 2016). Assessment methodologies preference have the potential to improve educational experiences of individuals with moderate to intense intellectual disability and development. Based on available research, the identification of preferred stimuli among individuals with disabilities is increasing, thus potentially increasing the overall quality of life and education (Cannella *et al.*, 2005; Castelli, Antonietti, Fabio, Lucchini, & Marchetti, 2013). Between 2002 and 2010, about 50 studies examined the effects of choice-making and preference assessments for individuals with severe to profound disabilities. Matching the appropriate choice-presentation method to the person's discrimination skill is important for several reasons. First, there is an increasing emphasis on providing choice opportunities to people with developmental disabilities and measuring stability of preferences over time (Zhou, Iwata, Goff, & Shore, 2001; Fabio, Giannatiempo, Oliva, & Murdaca, 2011). The choice opportunities, if they are to be meaningful, must be provided in a way that is matched to the client's discrimination skills. Second, choice-presentation methods that are beyond the client's discrimination skills may lead to two erroneous conclusions: that the person is unable to choose or that the person does not have a preference among the options presented. The effectiveness of any assessment methodology can be

influenced by individual differences as behavioral repertoire and physical capacity.

The current literature has provided information on reinforce potency (Milo, Mace, & Nevin, 2010) and preference stability (Zhou *et al.*, 2001). Based on the results of the present study, we can conclude that the girls with RTT are capable of making choices, keeping consistency and they do make self-determined responses, but to a lesser degree compared to normally developing girls. From this we can conclude that this is an important area of future investigation by analyzing, for example, a longer period of time and how their preferences tend to change. Finally, this research supports some of the assumptions that preferences for stimuli with people with moderate to intense intellectual and development disabilities remain stable (Zhou *et al.*, 2001; Ciccone, Graff, & Ahearn, 2007).

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