



Feuerstein Method and Rett Syndrome: a training with the tool “Identifying Emotions” of the “Instrumental Enrichment – Basic Program”

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Abstract

Rett syndrome (RTT) is a severe neurodevelopmental disorder. The Feuerstein Method has been widely applied in patients with neurological impairment, but never in RTT girls; furthermore, few studies have been focused on emotion recognition in RTT.

The purpose of this paper is to explore Feuerstein-approach relevance for intervention in patients with Rett syndrome, using the “Instrumental Enrichment – Basic Program” to train cognitive, metacognitive and executive functions. In this study we want to assess if also girls with this important intellectual disability could be trained with a simplified version of the Feuerstein tool “Identifying Emotions” and take advantages from this dynamic neurocognitive rehabilitation approach. Furthermore, we want to assess whether this educational intervention could modify the behavioral state of the patients.

8 RTT girls (6-12 years) were enrolled and trained with an adaptation of the tool “Identifying Emotions” of Feuerstein’s “Instrumental Enrichment – Basic Program”. Assessment consisted in a preliminary phase (pre-training) and an educational intervention (training).

All the patients were able to complete both the pre-training and the training part, showing: a gradual increasing of the distance of mediation, a significant reduction of stereotypic movements and increased times of fixation and selective attention.

RTT patients, despite the neurological impairment, could perform a training on meaning of emotions. In our study we observed that through this educational tool it is possible to overcome the basic tasks of labeling emotions and work on understanding the situations that generate the emotion and the classification of their intensity.

Despite the small number of patients, these pilot data can encourage research on cognitive functioning in RTT, which probably remains underestimated, and confirm the importance of cognitive empowerment rehabilitation for these patients; they also suggest that Feuerstein-approach may offer interesting perspectives to cognitive rehabilitation in this rare condition.

Keywords: Rett syndrome; Intellectual disability; Feuerstein Method; Cognitive rehabilitation; Identifying emotion

Introduction

Rett syndrome (RTT) is an X-linked dominant neurodevelopmental disorder that mostly affects girls, with a worldwide incidence of 1:8000/10000 female births [1].

Classical RTT syndrome is associated with mutations in MECP2, a gene located on the long arm of the X chromosome [2,3], that encodes the transcriptional repressor methyl-CpG binding protein 2, which is involved in the synaptic development and function [4].

Following a short period of apparently near-normal development, affected children suffer a psychomotor regression resulting in loss of spoken language and of manual skills. Postnatal microcephaly, typical hand stereotypies, movement disorder, scoliosis and abnormal breathing are the most common features, usually associated with epilepsy [5].

The traditional view has held that children with RTT are profoundly cognitively impaired and suggested a mental age of around 8-12 months [6]. We must consider that the cognitive assessment is very challenging in these patients because of their motor and verbal deficit. However, girls with RTT show strong communication attitudes and intense eye contact is a prominent feature in these patients, that tend to develop a “eye pointing” language as a substitute for their loss of speech and fine motor skills [7,8]. Useful methods of eye communication are commonly used in rehabilitation programs for people with RTT. Using the eye-tracking technology Fabio et al., demonstrated that RTT girls pay special attention to objects and people and they can show preferences [9].

With regard to cognitive skills, understanding emotions and mental states plays a crucial role in social behaviour and interpersonal interactions [8]. Until now very few studies have been focused on emotion recognition in RTT [10], but there is evidence that RTT girls are able to understand the issues of

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subjectivity and that with the right training they could be able to detect their own and others' mental states [11].

Different approaches have been designed to improve cognitive functioning in individuals with brain impairment, and among them one of the most popular is the Feuerstein Method, a unique educational technique that combines specially designed instruments in order to identify and enhance an individual's learning potential [10]. The Feuerstein Method is based on the theory of Structural Cognitive Modifiability and Mediated Learning Experience of Reuven Feuerstein and its purpose is to teach the skill of "learning how to learn", through a metacognitive approach. This method have been widely applied in children and young adults with learning and developmental disabilities or congenital neurological impairment, but, at our knowledge, never in girls with Rett syndrome [12].

We report our experience of a sample of eight patients with RTT, trained with an adaptation of the tool "Identifying Emotions" of Feuerstein's "Instrumental Enrichment – Basic Program" (IE-B). The purpose of this paper is to explore Feuerstein-approach relevance for intervention in individuals with RTT. In this study we want to assess if also girls with this important rate of disability could be trained with a simplified version of Feuerstein's tool "Identifying Emotions" and take advantages from this dynamic neurocognitive rehabilitation approach. Furthermore, we want to assess whether this educational intervention could modify the behavioral state of the patients.

Material and Methods

Patients

Eight RTT girls with MECP2 mutation between 6 to 12 years of age (mean 9 years) were enrolled (Table 2). All patients had the clinical criteria for Classical Rett Syndrome [13].

All participants were attending an Augmentative Alternative Communication (AAC) program and they were trained to use AAC communication in everyday life both at home and at school for an extended period of time.

Instrument: Identifying Emotions

The original tool "Identifying Emotions" consists of a cover page and 21 lesson pages, three for each emotion to be identified: surprise, sadness, disgust, fear, pain, anger and happiness [14].

The task of the instrument is to identify feelings and emotions by interpreting facial expressions and understanding their appropriateness in a variety of situations.

The format of each page is similar: at the top of the page there is a photograph of a human individual expressing an emotion, while below there are four artistically rendered situations (T1,T2,T3,D) that present some degrees of relevance and appropriateness regarding the emotional expression, which can be linked to the photograph of the human above [14].

The administration procedure consists of four phases: 1) identification and labelling of the emotional expression pictured at the top of each page; 2) gathering information from the four

situational pictures that trigger the target emotion; 3) giving each situational picture the appropriate emotion and intensity, and 4) colour-code each situational picture according to the following criteria:

- red: appropriate emotion and intensity
- yellow: appropriate emotion but not intensity
- green: appropriate emotion but the condition is missing
- blue: not relevant

Adaptation of the tool "Identifying Emotions"

For this study, according to the cognitive and communicative impairment of the patients, we have modified the original tool "Identifying Emotions" of Feuerstein's "Instrumental Enrichment – Basic Program" in a simplified version. Instead of seven emotions, three basic emotions (happiness, sadness, anger) and a complex one (surprise) were analysed.

Assessment

Assessment consists in a preliminary phase (pre-training) and an educational intervention of Feuerstein's "Identifying Emotions" tool (training).

The pre-training aims to verify the ability of the girls to attend the training sessions and to recognise emotions, showing a baseline. The steps of pre-training and training phase are showed as follows.

- **Pre-training (baseline)**

Step I – Presentation of real picture of emotions.

Step II – Presentation of drawn picture of same emotion.

Step III – Reconstruction of drawn picture via the model of the real image.

Step IV – Presentation of image of real situation that may trigger the emotion.

Step V – Classification of the presented image.

Step VI – Application of colour code.

- **Training with Feuerstein's "Identifying Emotions" tool**

Step VII – Presentation of real image of emotion proposed in the original worksheet and its reconstruction.

Step VIII – Presentation of the drawn picture proposed in the original worksheet.

Step IX – Classification of the presented drawn pictures.

Step X – Application of colour code.

Step XI – Generalisation of content.

- **Observation grid**

During pre-training and training sessions an observation grid was used in order to record the phases of work and the



abilities reached by the girls. The observation grid focused on the following features:

1. Prerequisites: It is important to know what communicative channel is most easily used by the girls and if there is any residual manual skill manifested by the presence of deictic and/or indicative gestures.
2. Distance of mediation: Feuerstein's "Identifying Emotions" utilizes a style of teaching called mediation. The level (low, medium, high) was defined using a score ranging from 0 to 9 (Table 1).
3. Table of the task phases. This table allows to establish, from time to time, the level of work and whether it has been internalized or not.
4. Evaluation of answers: The scoring system was as follows: 1) right answer: +, 2) wrong answer: -. The girl's performance was assessed using parameters of eye-hand coordination, as follows [21]:

-looks exactly, touches exactly: L+, T+ = +

-looks wrong, touches exactly: L-, T+ = -

-looks exactly, touches wrong: L+, T- = +

-looks wrong, touches wrong: L-, T- = -

L+, T- is considered a positive answer because of the strong motor impairment in RTT.

5. Frequency of stereotypes: the frequency of stereotypes was assessed using three parameters as follows: 1) low: absent, 2) medium: inconstant, 3) high: always present.
6. Selective attention: selective attention was assessed using three parameters, expressed as a percentage of the session time, as follows: 1) low : <30%, 2) medium: 30-70%, 3) high: >70%.

Results

The baseline is the starting point for structuring the tool's administration of "Identify Emotions". The number of pre-training (Steps I-VI) sessions necessary varied among the participants: 2/8 patients completed pre-training in one session; 4/8 in two sessions; 1/8 in four sessions and 1/8 in five sessions for all emotions analysed.

After that, all patients have completed the training part of the Feuerstein's "Identifying Emotions" tool (consisting in 12 schedules, 3 for each emotion analysed) in an average of 11 Sessions (Table 2).

Comparing the data recorded in the observation grid during the evaluation sessions of the pre-training (referring to the first working session) and the data of the training (referring to the last working session), despite the increasing difficulty of the work, the following have been observed: 1) a gradual increasing of the distance of mediation; 2) a significant reduction of stereotypic movement; 3) increased fixation times and selective attention to the presented targets.

Table 1: scores and levels of distance of mediation.

LEVEL	SCORE	MEDIATOR	MEDIATEE
low	0	produces the response through direct action on the mediatee	passive
	1	activates a sample to copy	partially starts an action
	2	orientates with active indications and verbalizes rules	responds spontaneously to the task
	3	specifies the general characteristics of the concept	If encouraged, is willing to solve the task
medium	4	refers to elements indicated above	Acts according to the previous mediation
	5	selects and encourages strategies	chooses appropriate strategies based on previous experiences
	6	indicates strategies already used deriving rules	applies strategies and expresses awareness of the rules
high	7	focuses attention on anticipating the response	formulates rules, demonstrates self-regulation
	8	focuses attention to meta-cognitive elements	there are elements of structural change
	9	passive presence	mediation is internalized

Table 2: main features of the girls enrolled in our study and N° of sessions needed by each patient to complete the pre-training and the training part.

PATIENT	AGE (years)	MECP2 mutation	PRE-TRAINING (N° of sessions)	TRAINING (N° of sessions)
1	8,3	Exon 4 deletion	2	12
2	11,5	p. T327fs	4	8
3	11,8	Exon 4 deletion	4	12
4	6,8	p. R133C	2	12
5	11,3	p. R294X	1	10
6	6,8	p. R168X	1	12
7	8,3	p. R168X	2	12
8	12,6	p. R306C	5	12



Considering the distance of mediation, we recorded an increase in the mediation distance. In the first session of the pre-training in fact 6 girls showed a low level of the distance of mediation, while 2 girls showed a medium level. During the last session of the training part instead, 5 girls showed a medium level of the distance of mediation, while 2 girls showed a high level (Figure 1).

During the sessions patients were able to use the color-code for choosing the appropriate emotion and intensity for each situational picture.

Considering the frequency of stereotypes, we noticed that from the pre-training phase to the training one there is a significant reduction of the phenomenon. In the first working session in fact, 5 patients showed high level of frequency of stereotypes, while 3 patients showed medium level of the same parameter; in the last working session most of the girls presented a low level of frequency of stereotypes (Figure 2).

The data we present about selective attention show a positive trend. In the pre-training phase we observed a quite disomogeneous selective attention level: 3 patients show a high level, 4 patients a medium level and one patient a low level of selective attention. The data collected in the last training session show, instead, high levels of selective attention for every girl (Figure 3).

Conclusions

All the patients were able to complete both the pre-training and the training part, respecting the times and rates of administration. The girls were trained to understand the situations presented through images and to associate them with the corresponding emotional mental states. In our study we observed that through this educational tool it is possible to overcome the basic tasks of labeling emotions and work on understanding the situations that generate the emotion and the classification of their intensity.

Furthermore, we observed that the girls have implemented a self-regulation process. During the sessions of pre-training and training, in fact, we recorded a reduction in the stereotypes

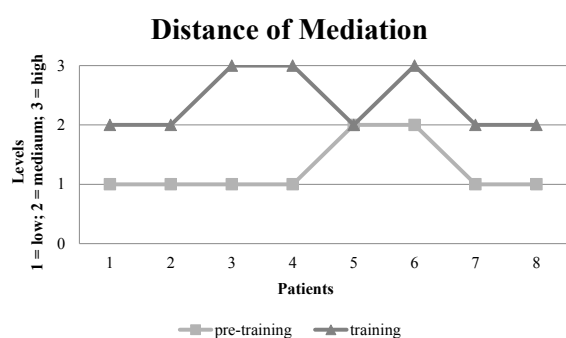


Figure 1 Comparison between data of distance of mediation recorded during the pre-training (first working session) and the training (last working session)..

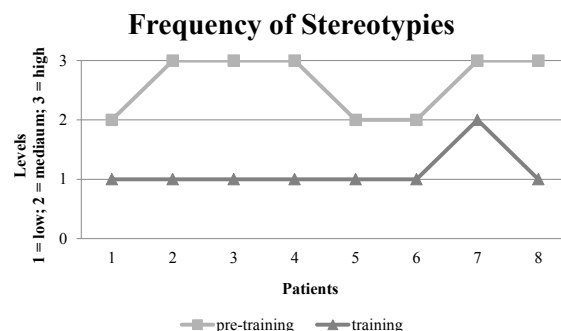


Figure 2 Comparison between data of frequency of stereotypes recorded during the pre-training (first working session) and the training (last working session).

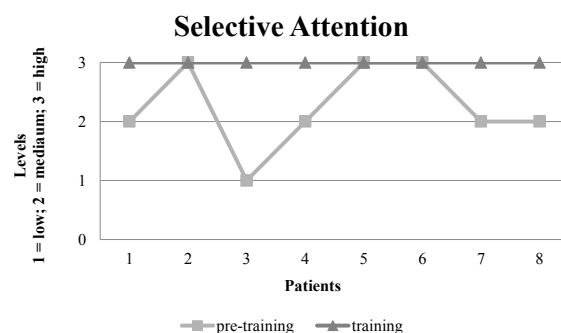


Figure 3 Comparison between data of selective attention recorded during the pre-training (first working session) and the training (last working session).

and an increase in the selective attention and in the distance of mediation. We also observed that these girls can memorize the color code used in the training.

We must consider that Feuerstein-approach provides a personalized educational technique, allowing individual differences in speed of choice. With this approach, even compromised patients like the girls of our study could perform the training and improve their cognitive skills learning the meanings of emotions.

Bearing in mind the large phenotypic variability, these results lead us to believe that girls with RTT have an undiscovered developmental potential that might deviate significantly from the manifested level of functioning. Feuerstein's approach has been a valuable support for facilitating communication between the mediator and the girls with RTT through images, enhancing learning.

Despite the small number of patients, these pilot data can encourage research on cognitive functioning in RTT, which probably remains underestimated, and confirm the importance of cognitive empowerment rehabilitation for these patients; they also suggest that Feuerstein-approach may offer interesting



perspectives to cognitive rehabilitation in this rare condition.

More extensive research is needed to provide a broader scientific evidence base and in the future it could be useful to administer this tool to a larger sample of RTT patients in order to validate these data.

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