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Attention and communication in Rett Syndrome

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ABSTRACT

The study of selective attention and its influence on communication in patients with Rett Syndrome (RS), in which communication abilities are impaired is particularly relevant. The aim of this study was to analyse attention and communication abilities in RS. A sample of 20 children (10 girls with RS and 10 control girls, matched on mental age) were tested on both attention and non-verbal communication abilities. Results showed that girls with RS have specific deficits in the ability to attend selectively to the relevant sources of information, and that they pay attention to irrelevant stimuli. Results related to non-verbal communication partially show specific impairment in girls with RS. Educational implications are discussed.

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1. Introduction

The ability to attend selectively to meaningful sources of information while ignoring irrelevant ones is essential to competent and adaptive functioning (Burack, 1994; Lane & Pearson, 1982). Conversely, impaired selective attention leads to increased distraction and diminished cognitive functioning, since responses to irrelevant stimuli interfere with the processing of targeted information (Fabio, 2001; Kerr, 2002; Lane & Pearson, 1982). The importance of selective attention and of individual differences in learning and memory has been studied for many years (Fabio, Perini, & Stefanoni, 1992; Fabio, Giannatiempo, & Antonietti, 2008; Fisher & Zeaman, 1973; Zeaman & House, 1979). An important development in the study of the relations between cognitive and social activities concerns the mechanisms of selective attention and ability to communicate. During input processing,

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persons comprehend relational messages through the interaction between externally derived information and information stored in social-knowledge structures. This results in interpretations and inferences derived from the integration process of cognition and communication (Hewes & Planalp, 1987). To study selective attention and communication in patients with Rett Syndrome (RS), in which communication abilities are impaired, is of particular relevance.

RS is a genetic disorder with a specific biological marker for its diagnosis: its aetiology is due to the genetic mutation of gene MECP2 on the x-chromosome (Amir et al., 1999). This developmental disorder which occurs largely in females results in severe mental retardation and neurological disability. In late infancy, after a period of superficially normal but subtly flawed development (Nomura & Segawa, 1992) children with RS undergo striking developmental regression (Hagberg, 1993; Moeschler, Charman, Berg, & Graham 1998). There is a characteristic loss of pre-existing hand use such as object reach, grasp, and manipulation, and the appearance of distinctive hand stereotypies (hand wringing, tapping, and mouthing). Post-regression, though severely intellectually disabled, patients often regain social interest and are relatively stable for an extended period before some of them occur in progressive motor deterioration in the form of weakness, wasting, and dystonia (Hagberg, 1993). During this period, crude self-feeding capabilities may be retained, but voluntary hand use is generally limited and hand stereotypies are pervasive (Fontanesi & Haas, 1988).

Some studies analysed attention and communication abilities in girls with RS. Witt Engerström (1990) shows that patients often remain visually attentive to objects and people, tracking their movements and even showing preferences by means of “eye pointing”. Also Demeter (2000) studied attention in RS. The author conducted some experiments on the relationship between cognitive and emotional-social abilities in girls with RS. In that study caregivers systematically presented an object, picture or action prior to engaging in an activity known and liked by the patient. After some weeks, a change in patients’ reaction was observed, demonstrating not only that they recognized the object, picture or action, but also that they displayed emotional reactions: they began to laugh, to be excited or to cry when the stimuli were presented. Demeter concluded that this form of learning, based upon attention to objects, is not to be considered as a mechanical process, but as a part of a rich human interaction, because the stimuli elicited not only cognitive replies but also emotional replies.

On the other hand Budford and Trevarthen (1997) suggested that girls with RS present higher communication attitudes, and that their positive orientation to human face and eyes may facilitate learning. With respect to communication abilities, Woodyatt and Ozanne (1992) reported that the literature indicates that the majority of RS patients are at a pre-intentional level of development with only a few girls able to signal intentions. Patients also display fluctuating attention to objects and a lack of motivation for handling them (Woodyatt & Ozanne, 1994). Olsson (1987) observed a greater interest for social stimuli and Watson, Umansky, Marcy, and Repacholi (1996) confirmed results by Budford and Trevarthen (1997) demonstrating that in the girls with RS the capability for showing intention and preference concerning a social stimulus is common. Also Budden, Myer, Butler (1990) assessed a group of 20 girls with RS using the Sequenced Inventory of Communication Development (Hedrick, Prather, & Tobin, 1975), and compared the communication abilities of their participants to the stage of their disorder. By stages II and III of the disorder communication skills had regressed to the 8- to 12-month level with no further regression noted in stage IV (Bates, Benigni, Bretherton, Camaioni, & Volterra, 1977). Bates et al. (1977) proposed that cognitive skills of the Piagetian Sensorimotor Stage V of means-end behaviour were necessary for intentional communication in normal developing subjects. Woodyatt and Ozanne (1992) concluded that the cognitive impairment reported in RS children, especially the failure to exhibit Piagetian Sensorimotor Stage V means-end behaviour, may account for the lack of the development of intentional communication reported in the literature. These authors studied six girls with RS and concluded that their communication skills regressed to the pre-intentional level of development. Communicative functions were inferred from a restricted number of non-verbal behaviour including primitive vocalizations, crying, facial expressions, gaze behaviours, breathing pattern, and touching. All these behaviours were interpreted by parents and caregivers for a variety of functions including request for social interactions and objects, refusal of food, greeting, pleasure, and pain.

Recent investigations (Antonietti, Castelli, Fabio, & Marchetti, 2001, 2002a, 2002b, 2003) in which RS received intensive cognitive rehabilitation using the assessment of non-verbal communication and

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