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Original article

Learning ability in children with Rett syndrome

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Abstract

The purpose of this article is to present results of a research study examining learning ability in individuals with Rett syndrome. The material for this article was drawn from a more extensive doctoral study, designed to investigate intentional communication in this population, through the use of songs in music therapy. Rett syndrome is a neurological disorder resulting from an X-linked mutation, affecting mainly females, and found across racial and ethnic groups worldwide. One of the main areas affecting functioning in individuals with Rett syndrome is a severe impairment of receptive and expressive communication. This creates difficulties when attempting to reveal their potential learning abilities. This population has been observed as very responsive to music hence music therapy intervention has been advocated in promoting and motivating them to communicate and to learn. Seven girls with Rett syndrome, between ages 4 and 10 participated in the study. A single subject, multiple probe design was applied during 30-min trials, three times per week and lasted 8 months. During the trials the participants were asked to choose from a selection of 18 familiar and unfamiliar songs, while their ability to learn was observed and measured. Findings revealed that all seven girls demonstrated an ability to learn and to sustain learning over time. This intervention demonstrated that individuals with Rett syndrome could be promoted and motivated to communicate and learn when therapeutically employed by a trained music therapists.

Keywords: Rett syndrome; Learning ability; Songs; Music therapy; Communication

1. Introduction

Individuals with Rett syndrome experience severe learning disability and as a result have special educational needs [1]. Despite the fact that they are characterized as severely developmental disabled, it is difficult to assess their intellectual potentials, thus leaving teachers and therapists working in special education teams with doubts about their educational potential.

Formal ways of learning are not always appropriate for the general population with developmental disabilities, including Rett syndrome. Their ability to learn is frequently underestimated and they are seldom expected to make achievements. Not only can they participate in their family's activities, attend school, enjoy the company of friends, and leisure time activities, they can become an integral and a contributing part of their community (Puescel, Bernior, and Weidenman, 1988 in [1]).

There is a wealth of literature reporting research into the development of communication in normal and developmentally disabled children some of which addresses issues related to learning ability and intentional (or lack of intentional) communication in individuals with Rett syndrome [2–6].

In a case study by Sullivan et al., a girl with Rett syndrome gained control when using switch-activated toys [7]. Initially, she was taught to use the toys and as she gained experience, she developed independence and initiation in activating these toys in her classroom. Sigafoos et al. found in their study assessing behaviors in three girls with Rett syndrome, that some of the girls' motor movements and other idiosyncratic behaviors might have been conditioned as unconventional forms of communication [8]. They call these behaviors 'potential communicative acts'. Although the girls in their study had limited behavioral repertoires,

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all had some types of behaviors that could be interpreted as a form of communication [8].

As Rett syndrome is considered a neuro-developmental disorder, it is believed that if individuals with Rett syndrome are given the right opportunity, they may demonstrate the capacity to learn new skills [9–11] even after reaching adult life [9,12]. These capabilities have been illuminated by girls who have been described as having learnt to make choices [13,14] and girls who identify symbols when taught in a motivating form [15]. The latter was demonstrated in a study which examined the effectiveness of computer-based interactive language development system [15].

Educational/social development in individuals with Rett syndrome might be achieved if their intellectual capacity and learning ability would be exposed. The lack of clear accepted forms of teaching them, and the ambiguous knowledge regarding learning ability in this population, urges us to look for alternative ways of learning [9,16].

In order to facilitate and to expose all those hidden skills mentioned randomly in this introduction, a strong motivational factor is needed. One such motivational factor is music therapy.

Music therapy was recommended by Dr Andreas Rett as early as 1982 as a means of treating individuals with Rett syndrome based on his observations that music evokes positive response in them [17].

Reports have shown how music making and music therapy promotes and motivate their desire to interact and communicate with their surroundings as well as develop their cognitive, affective, sensori-motor and physical skills [12,18–32].

Singing familiar songs has been an excellent motivator for encouraging individuals with Rett syndrome to attempt a task such as selecting a picture of a familiar song by touching it [21,22] or as an accompaniment for promoting hand use with musical instruments or objects [31,33]. Hetzroni et al. performed a study in which familiar songs were successfully applied as one of the domains in a computer-assisted instruction study which investigated girls with Rett syndrome's ability to identify picture symbols [15].

Due to the personal experience and to the important role music plays in the lives of individuals with developmental disabilities as a group, and Rett syndrome in particular, it was of our interest to see whether the clinical opinion of different therapists and teachers could be investigated in an experimental study. Although some reports suggest that individuals with Rett syndrome can only reach a pre-intentional level of development [4,5] our experience has made us think differently.

The hypothesis of the research was that music (songs in particular) as a highly motivating therapeutic tool, could ascertain the individual with Rett syndrome to achieve optimal educational potential [20].

2. Subjects and methods

2.1. The research question

This article will address the following research question: *Are girls with Rett syndrome able to learn and sustain learning over time?*

2.2. Participants

Seven girls, ranging in age from 4 to 10 participated in this study. All of the participants had been diagnosed with Rett syndrome according to guidelines established by the 'Rett Syndrome Diagnostic Criteria Work Group' [34]. Six girls were in stage III, the 'plateau stage' of whom two are not ambulant. One girl was in stage IV, the 'late motor deterioration stage'.

2.3. Design

A single subject, multiple probe design was used in this study to evaluate individual choice of, and response to, familiar and unfamiliar songs. This method of research enabled several comparisons of behaviors, responses, and musical elements within each case. In this design, each participant was viewed as a single experimental investigation [35–39]. The independent variable or treatment variable in this study comprised of 18 selected familiar and unfamiliar songs.

2.4. Procedure

The intervention trials incorporated choice-making of 18 familiar and unfamiliar children's songs. The 18 songs were divided into three 'sets', with a total of six songs in each set (four familiar and two unfamiliar songs in a set). Once the participants had made their choice during the trials (according to the pre-structured intervention procedure), the songs were sung by the investigator (a music therapist) accompanied by a guitar. The sessions were held individually three mornings per week and each lasted between 20 and 30 min. The duration of the study was 5 months and included baseline, intervention and maintenance trials followed by an additional 3 months with an additional 3 maintenance trials.

The pre-structured intervention procedure was set as follows. The investigator asked the participant to indicate a choice of a song (represented by a picture symbol or orthography cards placed on a communication board). The selection was made between two or four possible choices (depending on each participant's individual ability, as determined prior to the investigation). Each participant indicated a choice through her eye gaze, nose pointing or with her hand, according to her preference and ability. Once the song was chosen, the order of the symbols was randomly changed out of the participant's sight. The communication