The conductive environment enhances gross motor function of girls with Rett syndrome. A pilot study

Short title: Conductive education for Rett syndrome

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Keywords: Rett Syndrome, Conductive Education, PetÖ, functional abilities

Abstract

Introduction: Rett syndrome (RTT) is a rare neurological disorder usually associated with a mutation in the *MECP2* gene. Conductive Education (CE) is an educational approach that has not yet been explored with regard to children with RTT.

Objective: Assessing functional abilities of individuals with RTT due to CE intervention

Design: A single subject, AB design.

Method: This study assessed the functional skills of three girls with RTT aged 3, 4 and 5 years before and during participation in a Conductive Education program.

Results: Gross motor function improved whilst overall functional skills and hand function were unchanged at the end of the intervention period. Gross motor skills declined slightly in all participants over the summer holidays but improved again a few months after recommencement of the Conductive Education program.

Conclusion: Replication of this study with more subjects is justified as is comparison with other educational methods. A home intervention program should be constructed to prevent decline of functional skills over the long summer vacation.

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Introduction

Rett syndrome (RTT) is a severe neurological disorder affecting mainly females [1] and is usually associated with a mutation in the *MECP2* gene [2]. Core features include progress in early development followed by a period of regression with loss of hand and communication function, the onset of stereotypical hand movements and impairment of gait [3]. These difficulties are further complicated by neurological signs including altered muscle tone and dyspraxia, and the development of co-morbidities such as epilepsy, autonomic dysfunction and scoliosis.

The disability is usually severe and those affected are usually dependent on carers for assistance with most activities of daily living [4], but there is nevertheless potential for skills to improve. In two small samples of girls with RTT, treadmill training improved aerobic fitness and gross motor function [5] and walking and transfer skills improved with practice [6]. Active therapy and educational programs for girls with RTT likely contribute to improved functional abilities.

Conductive Education (CE) is an educational approach originally designed for children and adults with motor disorders such as cerebral palsy that aims to improve psycho-social development, co-ordination and motor control, and participation in activities of daily living. Conceptualized by Prof. Andras Peto in Hungary more than 60 years ago, its underlying concepts are that the biological damage and physical difficulties experienced by the child leads to a development of learned non-use at the physical level and learned helplessness at the psycho-social level, that many of the disabilities presented by the child relate to lack of motivation which require a pedagogic rather than a medical approach, and are based on the belief that individuals have a capacity for change [7,8]. Feuerstein's model of "the Active Modifying Environment" [9] and Lebeer's "Stimulating Ecology" model [10, 11] may serve as theoretical explanations for the general conditions for learning. Potentially

useful strategies include providing adequate learning opportunities, assisting motivation to develop new functions, facilitating learning processes by active stimulation and mediation, taking the child out of equilibrium and passivity, and practising goal directed activities. Implementation of a CE program is closely guided by conductor teachers [7, 8]. CE is conceived as a partnership between educator and learners to create a favourable environment for activity and learning and encouraging optimal levels of active participation [12].

Girls with RTT present with limitations in functional domains including mobility, hand function and social skills. Not previously explored, we hypothesised that a CE program based on the general conditions for learning described above, would improve functional skills of girls with RTT.

Methods

A single subject, AB study design was conducted with three girls diagnosed with RTT. Baseline assessment was conducted on two occasions separated by one month prior to commencement of the CE program. This was followed by an intervention period of approximately 18 months during which gross motor and hand function were assessed five times and general functional abilities assessed three times. Although not blind as to the purpose of this study, the assessors (1st, 2nd and 3rd authors of this paper) were not involved with the educational and therapeutic team.

Measures

Gross motor function

Gross motor function was measured with two scales. The *Rett Functional Evaluation Scale* [5] is a 30 item scale that measures upright abilities ranging from 2-point kneeling to standing and walking in terms of time over which a posture or movement can be sustained and the

distance of locomotion. It has been found to have high inter-rater reliability (ICC = 0.98) and to be sensitive to changes of functional abilities in four girls with Rett syndrome who participated in a two month treadmill program [5]. Based on the Gross Motor Function Measure [13], the *Rett Syndrome Gross Motor Scale* used was developed for RTT and comprises 15 items describing the level of assistance required to carry out various gross motor skills. Previous factor analysis of the scores of 99 girls and women with Rett syndrome identified two factors - a general gross motor score of 10 items (sitting on the floor, chair and stool; standing for 3, 10 and 20 seconds; transferring from sitting to standing; walking, side-stepping, and turning 180 degrees) and a complex gross motor skills score of 5 items (transferring from floor to standing, bending to the floor and returning to standing, walking up a slope, stepping over an obstacle, and running) [14].

Self-care and social function

Functional performance in the domains of self-care and social function were assessed using the *Pediatric Evaluation of Disability Inventory* (*PEDI*), designed for children aged six months to seven years with severe disability [15]. The PEDI has shown good inter-rater reliability and concurrent validity with the Bartelle Developmental Disability Inventory Screening Test [16]. For this study, part I of the inventory was used to assess 197 items of discrete functional skills. Skills are rated as 0 (unable, or limited in capability to perform the skill) or 1 (able to perform the skill, or beyond the level), summed for each section and scaled scores then determined. Scaled scores range from 0 to 100 with lower scores representing poorer function.

Hand function

Hand function was measured with a scale developed for RTT [17] and based on the Hand Apraxia Scale [18]. The subject is presented with several large (cup, fork, ball and toy) and small objects (usually a small piece of finger food), and abilities to grasp, pick up and hold each object are observed. The scale comprises eight levels of grasping function ranging from no purposeful grasping to the ability to pick up large and small objects with accurate pre-shaping of the hand as well as transfer objects [17].

The educational program

A daily conductive educational program was implemented in the pre-school centre of Tsad Kadima, Israel which is staffed by a trans-disciplinary team led by conductors and allied health professionals. Procedural integrity is built in the conductive education framework. All activities within the daily routine were recorded in detail including personal aims, facilitations in use, task series and periodical updates, which were in constant use by every team member.

The peer group, the conductor, and the components of rhythmical intention and the active daily routine, are some of the essential elements of the conductive program. The peer group is considered a powerful environmental stimulus for development, comprising a community of children whose activities are led by a conductor. The conductor plans a comprehensive program, guides the development and reinforcement of new activities and skills, and facilitates the development of interpersonal relationships and social behaviours. The conductive daily routine designed for each of the girls was a structured scheduled activity program, starting early morning with the arrival of the girls to the kindergarten until departing in the late afternoon. Activities of play, transitions, educational activities, meals, and toilet training were all viewed as learning opportunities.

Rhythmical intention is one of the methods of facilitation used in CE and uses two elements: rhythm and intention. Combining speech and activity into a single circle of feedback provides conscious and verbally directed attainment of goals [7, 8]. The combination of several sensory modalities to suggest a direction for action could be helpful in the presence of dyspraxia as is characteristic of individuals with RTT [19]. The activities in the daily routine were designed to challenge each child and simultaneously address different aspects of development. Routines appear important for creating a safe atmosphere which is a key component of learning [20]. Children with RTT need regular mediation to maintain activity levels and alertness. Therefore, within the group context, each girl had a helper during most of the daily activities.

Tsaad Kadima, The Association for Conductive Education in Israel, approved this study and families gave informed consent to participate.

Analysis

Serial assessment scores were viewed and plotted graphically to determine the trends, levels and slopes.

Results

Three girls aged 3 (p.Thr158Met), 4 (p.Arg255X) and 5 (p.Asp90FS) years participated in the CE program between December 2008 and July 2010. Functional gains were made in the gross motor domain whereas self-care and social skills and abilities to grasp objects remained mostly stable.

Gross motor function

Rett Functional Evaluation Scale: All subjects demonstrated a stable baseline period with much lower gross motor function for subjects 1 and 2 compared with subject 3. The intervention period was characterised by a gentle upward slope for each subject although this was steeper for subject 3. Assessment 5 was conducted after the summer holidays and scores decreased slightly for all three subjects before increasing again over the course of the subsequent academic year (Figure 1).

Figure 1 about here

Rett Syndrome Gross Motor Scale: Similar to the previous gross motor findings, all subjects demonstrated a stable baseline period. For the general gross motor skills subscale, subjects 1 and 2 had similar scores around the middle of the scale and then showed steady improvement throughout the intervention period. Subject 3 had high baseline scores and maintained this level throughout the intervention, illustrating a ceiling effect. For each subject, some decrease in skills after holiday periods (assessments 4 and 5) was observed (Figure 2).

The complex gross motor skills scores were stable during the baseline period remaining near this level for the first half of the intervention period after which scores then steadily increased. Each subject showed improvement in the more complex gross motor tasks with longer duration of intervention (Figure 3).

Figures 2 and 3 about here

Self-care and social function

Scores from the two domains of the PEDI scale were low with generally no or small changes across the five assessments. The self-care score of one subject decreased slightly

over the intervention period, and the social domain score of one subject showed a large increase at the final assessment after being very poor at all previous assessments (Table 1).

Hand function

Hand function was variable over time for each subject, in particular for subjects 2 and 3. Subject 1 demonstrated good grasping skills at baseline and generally maintained those skills during the intervention period. Hand function appeared poorer at the end of the intervention period although this was not a clear trend because of the fluctuating assessment findings (Table 1). Additional assessments could have assisted in the understanding of trends in this data.

Table 1 about here

Discussion

We conducted a single subject, AB study design with three young girls with RTT to assess functional change over a two year period whilst within a CE environment. After a stable baseline period, gross motor skills improved and social skills and the level of hand function was generally stable. Some functional gains appear to have been made by each of the girls.

The three girls demonstrated a range of gross motor skills at baseline with one able to sit, stand, transfer and walk with ease whilst the others needed assistance for these skills. We assessed gross motor function with two assessment scales which were developed for girls and women with RTT [5, 14] and which appeared to have sensitivity to change. Changes in different aspects of the gross motor domain were illustrated. The two girls with the poorest

gross motor function improved in their basic skills including the duration they were able to sit independently on the floor and on a stool. Over the course of the intervention, each also made gains in the more complex gross motor skills such as transferring from floor to standing and walking on a slope as well as demonstrating greater endurance in the maintenance of upright postures.

In a recent study of 70 girls and women with RTT in Australia, approximately 60% showed a small decrease in gross motor skills over a 3 to 4 year period, often whilst they were younger than eight years of age [21]. Each of the girls in this study were young and had recently regressed but made gains in the gross motor domain, in part developing further the basic skills of maintaining postures, standing and walking but also developing more challenging skills that enable better negotiation of the complexities of the environment. Other studies have also demonstrated the responsiveness of girls with RTT to physical training opportunities. For example, increased fitness was highly correlated with increased functional abilities (r=0.76) following a daily exercise regimens on a treadmill over a two month period [5] and improved transition and walking skills developed after practice [6]. The CE program included many physical activity opportunities and repetition that were directed towards improving participation in activities of daily living. This proactive approach appeared to have been particularly supportive of the development of gross motor skills.

Small setbacks in motor development were observed in both motor assessments at the beginning of each academic year. Similar findings have been reported in regards to autistic children [22]. In the Israeli special education system, there is a reduction in therapeutic availability over the summer vacation and the early part of the academic year because of a series of Jewish holidays. The implementation of summer and/or home programs and the continuation of proactive lifestyles could be important in the maintenance of abilities over these time periods.

General functional abilities relating to self-care and social skills as measured by the PEDI were relatively low at baseline and changed little over the intervention period, although the social skills of one girl improved at the final assessment. This is consistent with the severity of the disability and high dependence on caregiver assistance for most activities of daily living [4]. However, parents and staff observed functional improvements in these domains over the course of the intervention period, especially in the area social function even though these observations were not reflected in the PEDI scores. This perhaps related to the binary scoring for each of the items which could not measure smaller increments of change, suggesting that the PEDI was not sensitive for the identification of smaller increments of change in a population with severe disability such as RTT.

Poor hand function in association with dyspraxia is a cardinal sign of RTT [1] although variability of hand function has been observed in a population-based sample of 144 girls and women with RTT [17] and was reflected in the range of baseline skills seen in this small sample. Over the course of the study, performance of grasping skills was variable and we acknowledge that additional data points could have been required. One subject maintained her strong grasping skills illustrating a ceiling effect and the other two subjects with less well established grasping skills appeared to be less stable over the study period. These observations were consistent with a longitudinal study of 72 girls and women with RTT in which those with more advanced hand function were more likely to retain skills compared to those with poorer hand function over a three to four year period [23]. The literature has described improvement of hand function in RTT in small numbers of cases [23] and sometimes following intensive and structured supported feeding activities [24]. The CE program included intensive intervention in different motor areas including the promotion of hand use at meal times, but in contrast to effects on hand function seen in children with cerebral palsy [25], the program did not appear able to improve abilities to grasp objects in

the small sample in this study. The subjects in this study were young and regression of hand function sometimes observed for individuals with RTT at these ages could have been continuing. Further investigation of a larger number of subjects is needed to clarify effects on hand function.

The CE program tested in this study used many strategies to form a rich framework of teaching and rehabilitation activities as has been described in the literature [7, 8]. At the system's level, strategies related to the conductor-teachers team (ensuring continuously updated knowledge, supervision and quality control); the grouping of children (for motivation, social facilitation, and integration); the educational program (comprehensive and age-related curriculum); and the daily routine (continuity, coherency in activities, regulatory role). At the level of the program, strategies included task analysis (a number of underlying abilities which are functional prerequisites were embedded in any task; and the task series (from simple to most complex, alternative ways, algorythmical order). At the session's level, strategies related to each lesson's plan (age, curriculum); rhythmical intention (using speech to help to create intention, to regulate skill performance and bring tasks to the level of consciousness); facilitation and feedback (educational, visual, auditory, environmental, emotional, manual achieve movements or hold postures); and the organisation of the environment (usage of plinths, chairs, splints, other devices). Overall the CE environment was demanding and represented one way to provide an enriched environment. When housed in an enriched environment, *MECP2* null mice have demonstrated improved motor abilities which may be related to increased levels of BDNF and reinforcement of neural synapses [26, 27]. The benefit of each individual strategy is not known but the composite appeared to nurture the development of gross motor functional abilities in these three subjects.

We established a stable baseline period prior to implementing the intervention, and then observed the effect of the intervention with a series of assessments for each subject. The control period and multiple assessments over a length of time under standardised conditions give some confidence in the association between the intervention and subsequent changes in the outcome variables [28], and we are not aware of other events in the lives of the three subjects that could provide alternative explanation for the improved gross motor skills. Rett syndrome is a rare disorder [29] and we took the opportunity to try to understand the effect of the intervention with a single subject design. The enrolment of the three subjects into the CE program afforded the opportunity to introduce control into the study design to provide a higher level of evidence than simple description when a randomised controlled trial was not feasible. The range of assessment findings in the hand and gross motor domains at baseline indicated variability in the phenotype which is characteristic of RTT suggesting some external validity of our findings. On the other hand, the current study is still limited by the small subject number and we recommend replication of this study to confirm our findings, further explore the development of social as well as communication skills in response to CE, and compare CE with other educative and rehabilitation strategies.

This single case design study found improved gross motor abilities in association with participation in a CE program over a two year period. A comprehensive educational program with physical activity components, the reinforcement of repetition and social enjoyment of group work is consistent with an enriched environment and has the potential to provide advantage to girls with RTT.

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Declaration of interest

The authors hereby declare that there are no financial or other relationships that might lead to a conflict of interest.

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Table 1. Scaled scores for the self-care and social skills domains of the Pediatric Evaluation of Disability Inventory and levels of hand function at the two baseline assessments and during the intervention period for each subject.

Domain	Subject	Baseline		Intervention period (months)				
		Initial	1 month	6	9/10	12	16/17	20
Self-care skills[a]	1	28	26.2	17.4	17.4		17.4	
	2	29.4	29.4	29.4	29.4		29.4	
	3	24.1	24.1	24.1	24.1		24.1	
Social skills[a]	1	35.1	33	35.1	35.1		33	
	2	35.1	35.1	35.1	35.1		35	
	3	3.1	3.1	0	0		37	
Hand function[b]	1	8	8	8	7	8	7	8
	2	5	5	2	4	4	5	1
	3	3	1	2	1	3	4	1

[a] Scaled scores range from 0 to 100 with higher scores representing higher function; [b] Eight levels of hand function with level 8 representing better hand function.

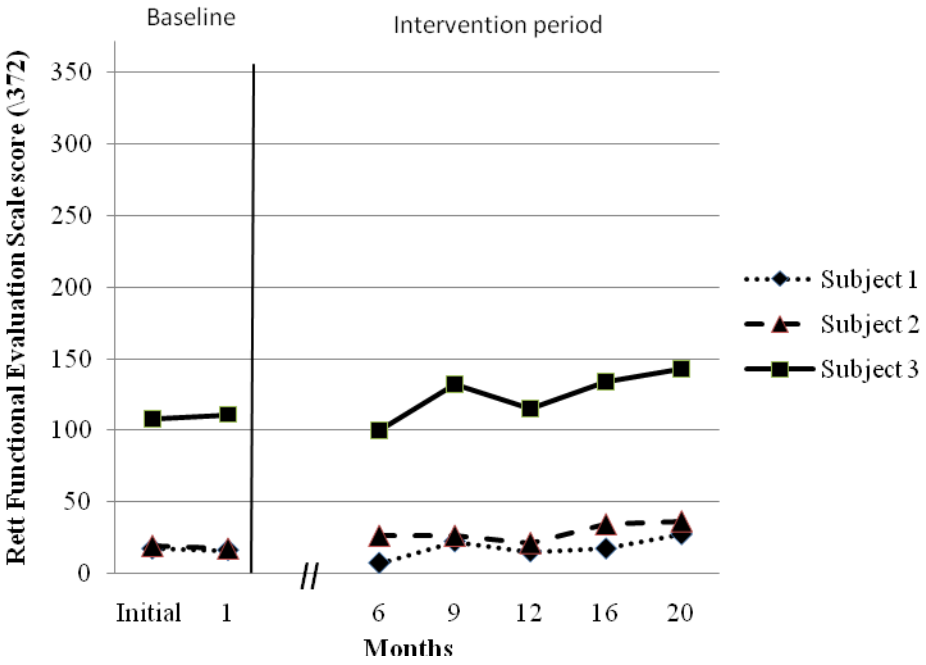

Figure 1: Level of gross motor function of the three subjects at each assessment measured with the Rett Functional Evaluation Scale

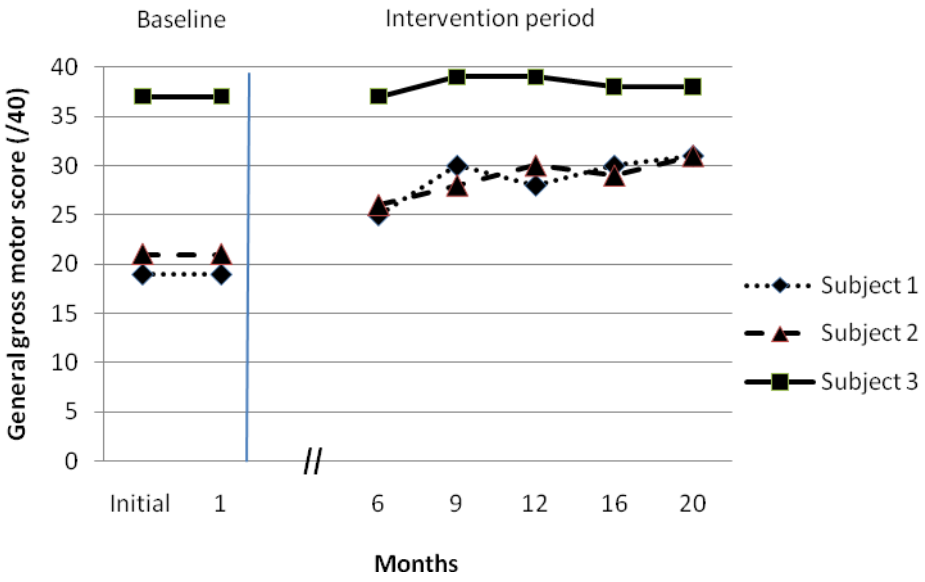

Figure 2: Gross motor function of the three subjects at each assessment measured with the general gross motor subscale of the Rett Syndrome Gross Motor Scale

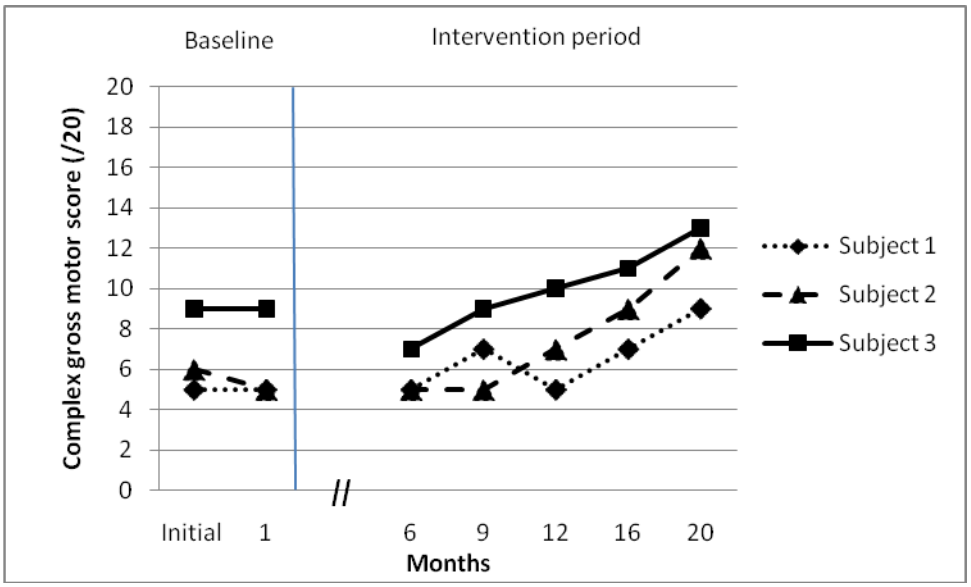

Figure 3: Gross motor function of the three subjects at each assessment measured with the complex gross motor subscale of the Rett Syndrome Gross Motor Scale