

Appendix 1. Papers on intervention

Authors	Participants	Topic	Intervention/Method	Outcome and Results
Bartolotta, T.E Remshifski, P.A (2012)	4	Coaching communication partners: investigation of communication intervention during mealtime	A program for coaching and increase communicative interactions was used	Coaching teachers and helpers can be used to enhance communication in persons with Rett syndrome
Bergström-Isacsson, M (2011)	35/11	Music and vibroacoustic stimulation in people with Rett syndrome	Physiological data from neurophysiological brainstem assessment was used. Facial expressions from video were also studied using the Facial Action Coding System (FACS). Recommendations for intervention are included	Musical stimuli had measurable effects on autonomic functions in 35 people with Rett syndrome and 11 normally developed controls. Responses to different kinds of music could not be predicted. Facial movements are related to the syndrome, expressions occurring spontaneously and not seeming to imply specific emotions
Bumin, G Uyanik, M Kayihan, H Duger, T Topcu, M (2002)	4	The effect of hand splints on stereotypic hand behaviour in Rett syndrome	Video tape recordings were used to settle baseline, intervention, and withdrawal	Using splints decreased hand movements in Rett syndrome but the effect could be other unwanted movements as well
Bumin, G Uyanik, M Yilmaz, I Kayihan, H Topcu, M (2003)	1	Hydrotherapy for Rett syndrome	Eight weeks of hydrotherapy.	Anxiety decreased as well as hand stereotypies. Functional hand use and feeding improved as did balance in walking
Elefant, C Wigram, T (2005)	7	Learning ability in children with Rett syndrome	Songs in music therapy was used in trials 30-min long, three times per week	There was ability to learn and to sustain what they had learnt over time
Elefant, C Lotan, M (2004)	1	Rett syndrome: dual intervention–music and physical therapy	Physical- and musical intervention was used jointly	It was possible to carry out the physical therapy with the girl accepting it. The child's communication skills improved during intervention period

Appendix 1. Papers on intervention

Authors	Participants	Topic	Intervention/Method	Outcome and Results
Elefant, C (2002)	7	Enhancing communication in girls with Rett syndrome through songs in music therapy	Music therapy was used as a way to promote communication	Certain songs were preferred and others rejected. There was intentional choice making and ability to learn and sustain what they had learnt over time. Intentional communication was shown with consistent behaviour and emotional responses
Evans, I. M Meyer, L. H (1999)	1	Modifying adult interactional style as positive behavioural intervention	Behaviour was studied and a program for a three year intervention was made for teaching voluntary, manual as well as communicative response	The first intervention program failed. A revised intervention was later used using more playful interactions resulting in positive social interactions and play and reduction of hand stereotypies and other excessive behaviours
Fabio, R. A Giannatiempo, S Oliva, P Murdaca, A. M (2011)	12	Increase of attention in Rett Syndrome	Well structured procedures were used in order to increase high attention abilities	There was improvement in selective attention and a decrease in the amount of help needed
Hetzroni, O Rubin, C Konkol, O (2002)	3	Using assistive technology for symbol identification by children with Rett syndrome	An individualized multimedia program was made in order to facilitate matching spoken words to symbols.	There was a steady learning curve across symbol sets as well as partial retention of knowledge
Jacobsen, K Viken, A von Tetzchner, S (2001)	1	Rett syndrome and ageing	Intensive physiotherapy intervention was performed	Walking without support was regained after 21 years without walking. There was also improved hand use
Kjoerholt, K Salthammer, E (1990)	6	Teeth-grinding project.	A 6 week intervention with massage/relaxation therapy and 2 weeks follow up	During the intervention there was reduced muscular tension, but at follow up there was no remaining effect

Appendix 1. Papers on intervention

Authors	Participants	Topic	Intervention/Method	Outcome and Results
Lotan, M Isakov, E Merrick, J (2004)	4	Improving functional skills and physical fitness in children with Rett syndrome	Intervention using a treadmill	Improved physical fitness as well as improved functions like walking and walking stairs was found
Lotan, M Merrick, J Carmeli, E, (2005)	1	Managing scoliosis in a young child with Rett syndrome	Intensive asymmetrical activation of trunk muscles through equilibrium reactions was performed for 18 months. Follow up after 1 year	There was reversing of scoliosis during the period of intervention. At follow up after one year with no intervention, scoliosis had progressed
Lotan M (2006)	3	Management of Rett syndrome in the controlled multisensory (Snoezelen) environment	Using a Snoezelen facility to improve physical, sensorial, and functional abilities	Positive effects of interventions were found in agitation, falling and range of motion
Lotan, M Schenker, R Wine, J Downs, J (2012)	3	Improving gross motor function using an educational approach	Conductive education program was used to enhance gross motor function	Improved gross motor function was found, though skills declined slightly in all participants during summer holidays. After resuming school, improvement was seen again after a few months
Pizzamiglio, M. R et al (2008)	1	Investigating the possibility to rehabilitate purposeful use of the hands and hand-eye coordination in a person with Rett syndrome	Computerized visual-motor coordination training as well as a sensory-motor rehabilitative program specifically designed for her was used	During a 3 year therapy she partially regained the possibility to use her hands for handling objects and for communication
Qvarfordt, I Witt Engerstrom, I Eliasson, A. C (2008)	3	Guided eating or feeding: three girls with Rett syndrome	A comparison between being fed and having meals with guided eating was performed	There was indication that guided eating made participants more involved in and participated more in eating. There was improved coordination between opening of the mouth and spoon, if guided eating was used

Appendix 1. Papers on intervention

Authors	Participants	Topic	Intervention/Method	Outcome and Results
Ryan, D McGregor, F Akermanis, M Southwell, K Ramke, M Woodyatt, G (2004)	3	Facilitating communication in girls with Rett syndrome	The interaction of student-teacher in three different activity types was examined	If increased opportunities for communication was provided by caregivers this did not result in increased responses from the girls
Sigafoos, J Laurie, S Pennell, D (1996)	2 in study 1 2 in study 2	Teaching children with Rett syndrome to request preferred objects	Study 1: Communication board and a symbol for "WANT" was used. Intervention was made of verbal support, gesture, and physical prompting. Study 2: two other participants who failed to show progress during the initial training had modified response requirements	Study 1: One participant increased in independent requests. The other participant became more consistent during intervention. Study 2: With some modifications the children's responses increased
Wales, L Charman, T Mount, R. H (2004)	8	Assessment of repetitive hand behaviours in girls and young women with Rett syndrome	Repetitive hand behaviour was studied as well as how environmental conditions affected the frequency of these	Variation in environment had limited effect on repetitive hand behaviours
Wigram, T Lawrence, M (2005)	1	Using music therapy as a tool for assessing hand use and communicativeness in children with Rett Syndrome	Improvisational music therapy was used to study performance of hand use, eye-referencing and intentional communication	Responsiveness and potential for development in the music therapy assessment as well as functional hand use, eye-referencing and intentional communication was found. Hand clasping/plucking was reduced or discontinued and there was turn-taking and vocalisation with appropriate emotional expression