

Abstract

Background: Rett syndrome (RTT) is a neurodevelopmental disorder which affects basic body functions including the central control of the autonomic nervous system in the brainstem. Music is used by parents and carers in different situations, e.g. to calm down, to activate, to motivate and in communication. The aim of the study was to examine what effect musical stimuli had on the control functions of the autonomic nervous system, and on cortical emotional reactions, in participants with RTT.

Methods: The study included 35 participants with RTT who were referred to the Swedish Rett Center for routine brainstem assessment during the period 2006-2007, and 11 children with a normal development. A repeated measures design was used, and physiological data were collected from a neurophysiological brainstem assessment. To identify facial expressions elicited by possible pathological brainstem activities, data were also collected from video analyses of facial expressions using the Facial Action Coding System (FACS). The control situation was the physiological baseline of the participant's own autonomic function at rest. After establishing a baseline the participants were exposed to six musical stimuli. *Horn* was chosen to elicit an arousal response and *Activating* (parents' choice) a sympathetic response. *Calming* (parents' choice), *VT* (Vibroacoustic stimulation), *VT+Mu* (VT combined with calming music) and *Mu* (that same music without vibrations) were expected to elicit a parasympathetic response. The continuous dependent variables measured were: Cardiac Vagal Tone (CVT), Cardiac Sensitivity to Baroreflex (CSB), Mean Arterial blood Pressure (MAP) and the Coefficient of Variation of Mean Arterial blood Pressure (MAP-CV). These parameters were used to categorise brainstem responses as parasympathetic (calming) response, sympathetic (activating) response, arousal (alerting) response and unclear response. The analyses were supplemented by case vignettes, where additional physiological parameters were also taken into account. Facial expressions were coded and categorised into positive emotions, negative emotions and ambiguous responses. These expressions were then related to results from brainstem assessment and the music used.

Results: Continuous responses showed that *Calming* and *VT* increased CVT significantly in the RTT group. *Horn* elicited a similar response in both groups (decrease in CSB, indicating an arousal). In the RTT group, the expected categorical responses related to the hypotheses were observed in 7% for *Horn*, 36% for *Activating*, 39% for *Calming*, 52% for *VT*, 32% for *VT+Mu* and 28% for *Mu*. The FACS analyses indicated that a majority of the RTT

participants had specific disorder-related movement patterns in their facial expressions. The findings from analysing the case vignettes also disclosed the impact of blood gases and breathing patterns on RTT participants' physiological responses to the music and on their facial expressions.

Conclusion: Musical stimuli have measurable effects on brainstem autonomic functions in RTT and non-clinical individuals, but it is not possible to foresee responses to different kinds of music. The disorder-related movement patterns in facial expressions found in RTT individuals occurred spontaneously and may not directly indicate emotion. Brainstem assessment is a new method to observe and analyse autonomic responses to music. In combination with brainstem assessment, FACS can be used for identifying and separating pure brainstem triggered facial responses from facial expressions of emotions elicited from the cortex. However, FACS is a new method related to both RTT and music therapy, and presents a new area for further research. The findings from this present study might help caregivers, teachers and therapists to be more observant of specific details, which would ultimately benefit people with RTT.