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Active Music Therapy and Williams Syndrome: a Possible Method for the Visual-Motor and Praxis Rehabilitation?

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ABSTRACT

Introduction: Notwithstanding variation from person to person, research into Williams Syndrome identifies difficulty in the following areas of psychomotor control: co-ordinating movements, spatial orientation, physical ability and, in particular, visual-motor integration. These difficulties are magnified by physical traits, mainly low muscle tone and contraction of the joints, which present a further cause of reduced coordination. Music and sound act as specific stimuli to obtain emotive and movement responses, activating various sensory areas. We explored the efficacy of active music therapy (MT) on motor functions in patients with WS. **Aim**: We investigate the use of active music therapy, in particular the use of rhythmic components, to stimulate functional hand-eye co-ordination and visual-motor integration in patients with WS. **Method**: The study involved 10 subjects with WS, aged between 3 and 20. The patients were involved in weekly sessions of music therapy. The sessions consisted of exercises using rhythm and movement, vocal exercises and musical improvisation over a rhythmic base. Patients do not require any musical training. A music therapist who played an active part in the proceedings conducted each session. In MT sessions, Visual-motor integration and praxis was tested (VMI – Visuo-Motor Integration Test, adapted, TGM) before and after the program and every two months during the program. **Results**: The patients showed significant improvements in visual-motor ability and in praxis skills in the direct aftermath of the program. Less significant, but nevertheless important, results were observed long-term. **Conclusions**: Music therapy is demonstrated to be efficient for improving praxis skills and visual-motor integration in subjects with Williams Syndrome. We propose an idea to use music therapy as an integrated part of rehabilitation.

I. BACKGROUND

Williams Syndrome is a rare neurodevelopmental disorder characterized by deletion of genetic material from the long arm of chromosome 7. This area includes about 26 genes that probably cause the typical morphological, physiological, clinical, cognitive and behavioural features of this syndrome. Subjects with WS have a distinctive facial profile, including puffiness around the eyes, a long philtrum, and a stellate pattern in the iris. Other symptoms include failure to gain weight appropriately in infancy and low muscle tone, cardiac, hormone and gastrointestinal problems. Most individuals are highly verbal relative to their IQ, and are overly sociable, having what has been described as a “cocktail party” personality.

The focus of the research initially concerned a dissociation between the language skills that are relatively preserved and deficit of motor, visuospatial, mathematical and planning skills; moreover subsequent research made known a rather more complex neuropsychological profile and asymmetric performances. Recently there is a growing interest in finding correlations between data of genetic research, neurological and neuropsychological profiles, as evidenced by a recent book, "Journey from cognition to brain to gene".

These studies confirmed the existence of specific profiles in children with WS with more preserved abilities in some areas (verbal fluency, memory for faces), and marked deficits in other sectors (motor and praxis, visual-spatial, logic skills). The shared assumptions in the recent period are that there isn't dissociation between cognition and language, but rather a need for comparable instruments to assess all skills in similar ways and rehabilitation projects based on objective and individualized data. The specificity of each subject led us to conduct this research trying to offer individualized programs for each one, with criteria of validity and objectivity through the use of standardized tests. Another important aspect, which makes it hard for a thorough and well-defined project, is the high variability relative to IQ ranging between 40 and 90, with a wide variety of skills in tasks that require specific performance (ex. in schools). Then we explored some aspects of music as a possible intervention to integrate known rehabilitation programs. In particular, despite the music therapy is used to facilitate learning in these subjects, and although musical predisposition of these subjects is known, the literature contains few assessments of MT in medical care, and there is substantial heterogeneity across studies. Most of investigations in WS and music explored: emotional aspects, increasing of relations, attention spans. Musical strength in individuals with WS involves not so much formal analytic skill in pitch and rhythm discrimination as a strong engagement with music as a means of expression, play, and, perhaps, improvisation.

The findings show that individuals with Williams syndrome tend to be more engaged in musical activities than others, and I report a possible neuroanatomical correlate of this engagement, with increased activation in the right amygdale to music and to noise. Williams Syndrome represents a compelling model of the relationship between genes, brains, and such complex cognitive behaviours as music. As regards the object of our study, we focused on aspects of visual-spatial tasks, because the subjects with WS have the same difficulties of subjects with right hemispheric lesions (i.e.: fragmented drawings, low visuomotor integration, good reproduction of individual elements with loss of the whole complex, poor performance in tasks of copying complex geometric shapes that require integration as in VMI) without similar brain diagnostic examination. Recent investigations gave a hypothesis about the biological basis of this peculiar dissociation in visuospatial tasks: a deficit on the dorsal cortex and a relative sparing of the ventral one in the WS, that preserve the aspects of form, colour and faces, and make a loss in spatial relationships and visual-motor control. Visuospatial deficits can be defined as disorders that result in an incorrect estimation of the spatial aspects between different objects that concern the relationship between the person and the object, the same relationships between different objects and the orientation of stimuli associated with a corresponding fall in memory and spatial thought. In particular we can think that the visual-spatial deficit is a disorder characterized by: a discrepancy between verbal IQ and performance IQ, deficient ability of MLVs, erroneous estimates of the spatial relationships between objects in relation to body, disturbance praxis and visual-motor coordination, difficulty in grasping the paralinguistic aspects of communication.

Experimentally it was found that listening to music in immobile position activates the same brain areas that "manage" the motor activity. We have therefore highlighted the results of this research done on brains subjected to listening music through fMRI, "which makes it possible to observe in vivo activity of different brain areas during listening to music". Researchers of McGill University discovered that, even in stillness, listening to music can activate areas that coordinate motor activities. Evidences on the rehabilitation plan were found, for example, in movement disorders seen in patients recovering from stroke or in Parkinson Disease. Music has been used as a form of therapy for many different diseases and may be experienced and appreciated by even the most severely physically or cognitively impaired subjects. MT has been widely used in the rehabilitation of handicapped children, providing one of the few ways in which these subjects can attain self-expression. The brain areas involved in music are active in processing language, auditory perception, attention, memory, executive control and motor control. Music efficiently accesses and activates these systems and can drive complex patterns of interaction among them. The idea is that brain systems underlying music are shared with other functions. Evidence suggests that music may activate these systems differently than speech or other stimuli do and might enhance the way the systems work together. The second helpful insight was the development of the auditory scaffolding hypothesis. This model proposes that the brain assigns nearly everything that deals with temporal processing, timing, and sequencing to the auditory system. This process works because sound is inherently a temporal signal, and the auditory system is specialized and highly sensitive for perceiving time information. Experiences with sound may help bootstrap—or provide a kind of scaffolding for—developing or retraining such abilities. As music may be the most complex temporal auditory language, it may offer superior auditory scaffolding for cognitive learning. The extended shared brain system theory and the auditory scaffolding theory provided a new theoretical foundation for the therapeutic use of music in motor, speech and language, and cognitive rehabilitation. Music can also activate the attention network on both sides of the brain, which can help overcome attention problems caused by motor difficulties.

II. METHOD

A. Subjects

Ten WS patients were invited to participate in the study, through the participation of the National Williams Syndrome Association, Turin. The study lasted 12 months. Because of the rarity of the conditions, the subjects age range varied between 3.8 and 18.6 years. According to our selection criteria, patients with other physical (visual or auditory sensory deficits, movement diseases) or cognitive and relational diseases were excluded from the study. During the study all subjects attended school or disability centre, but not rehabilitation activities for the specific disease. For the whole year, in accordance with the Association programs, the group attended music, dance and theatre workshops, to improve emotions, wellbeing and socialization, but the aspects of these sessions are not discussed in this study. However, we emphasize this feature, because aggregating group activities and expressive situations favoured the relationship with the therapist, reducing the situations of anxiety and difficulty in performance and testing. The subject took part in weekly sessions of traditional praxis and visuomotor rehabilitation for 16 weeks and, after a stop of two months, had weekly sessions of MT for 16 weeks. Furthermore, no significant differences in praxis and visuomotor functions, as assessed by the TGM (Test of Motor Development) and VMI (Beery-Buktenica Visual Motor Integration Test) , emerged among the subjects (Tab 1-2).

TABLE 1.

No. of patients		10
Gender		
	Male	7
	Female	3
Mean Score		
	VMI	67,2

TABLE 2.

Subject N°	VMI (SS)
1	66
2	59
3	71
4	76
5	51
6	68
7	72
8	67
9	69
10	73

During the VMI test, the subjects experienced difficulties in complying with the correct orientation of the figures and the relationships among their component parts. The patients often failed to reproduce correctly the complex figures, exhibiting a very poor performance, both in terms of quantity (number of figures reproduced correctly) and in terms of quality (types of figures reproduced correctly. The TGM test showed that none of the children and the teenagers had reached an appropriate level of motor self-possesion for their age. Finally, compared to subjects with typical development, WS individuals show a greater impairment of fine motor skills than global ones. Object Control (subtest 2) and balance tests turned out to be very complex. Moreover, the assessment leads us to the conclusion that the individual variability is considerable.

B. Study Design

The study main aim was: to evaluate the MT effects in praxis and visuomotor skills; to compare the MT results with traditional rehabilitation in the same group of subjects. The controlled, semi-experimental case study lasted 12 months.
The first part of the study lasted 4 months (16 weeks), with sessions of traditional rehabilitation activities in the absence of music. The second part of the study also lasted 4 months (16 weeks), but with individual, in pairs or in small groups sessions of active music therapy. The subjects' visuomotor and praxis ability was assessed using two tests (VMI and TGM). We preferred to value the same subjects because of the intra-individual variability in the syndrome and the interest for each subject reaction to the two treatments.
TGM examination was administered to all patients at weeks 3, 6, 9, 12 of the two period of study and at the follow-up examination, which was conducted 2 months after completion of the study. The VMI test (Standard Score 40-117) was administered at weeks 8 and 17 and a the follow-up examination. A psychologist conducted patient examinations 1 hour before the start of the sessions. At the and of both the treatments a Parents Interview Question was administred. Post-session examinations were conducted within 1 hour after conclusion of each session.

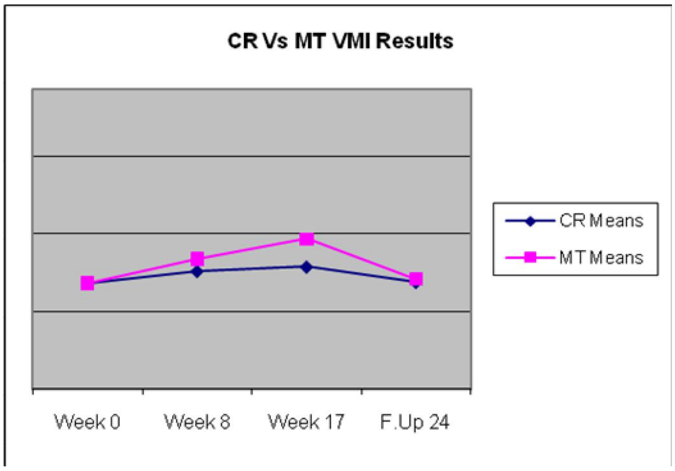
In summary, the VMI test consists in copying a developmental sequence of geometric forms: the full form has 27 items, but for children from 3 to 7 years old there is a 18-item short form. The two additional tests help us to compare an individual's test result with relatively pure visual and motor performances. The Visual Perception of Geometric Shapes (supplemental test 1) consists in identifying the exact match for as many of the 27 stimuli as possible in a three-minute period; in Motor Coordination (supplemental test 2) the subjects have to outline the stimulus forms with a pencil without going outside the double-lined paths. The point of the TGM test (Test of Gross Motor Development) is to determine the subjects' gross motor skills. Each one includes three of four behavioral components that are presented as performance criteria. In general, these behaviors represent a mature pattern of the skills. In particular, 12 skills are assessed divided into locomotor (run, gallop, hop, horizontal jump) and object control (strike, catch, kick overhand throw) skills.

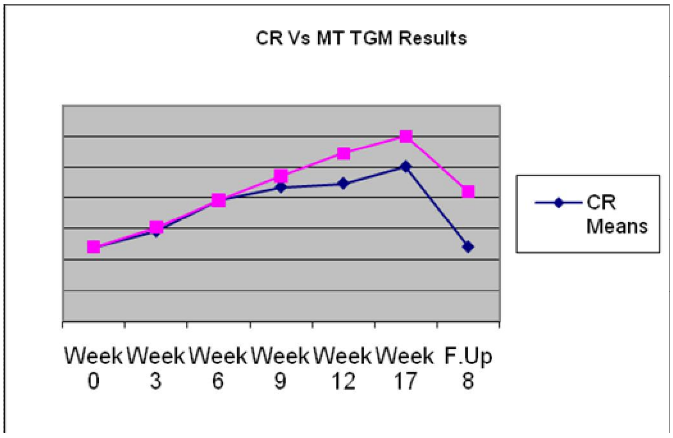
In the first part of the study (T1), the subjects were treated with Conventional Rehabilitation (CR) sessions to develop visuospatial and motor skills. These sessions are set up according to a structured program of rehabilitation, as follows: physical activities with games (walking, running, free movement and imitation, weight shifting and balance trining); works with draws (reproduce figures, freehand drawing); recognition and completion of figures; reconstruction of images and objects. During the sessions patients performed the exercises concurrently, with minimal interaction with each other.
In the second part of the study (T2), each group took part in 16 weekly sessions of active MT. Active MT involved improvisation by the therapist, who invited patients to play an active role using instruments and voice. Patients do not require any musical training. Each session was conducted by a music therapist who played an active part in the proceedings. The MT session lasted 1 hour: rhythmic movements, 20 minutes; active music involving collective invention and improvisation, 20 minutes; free body expression to melodic and rhythmic music, 20 minutes. The equipment consisted of a piano, an organ, percussion instruments (eg, metallophones, xylophones, drums, wood blocks and cymbals) and a high-fidelity system. Patients used all instruments, adopting a free technique. In MT sessions, exercises were performed individually, in pairs or in small groups, with a high level of interaction and communication within the group (eg, patients performed rhythmic or melodic improvisation using instruments and voice freely, or, in another exercise, some of the patients played the wood blocks with an alternating movement of the arms while the rest of the group marched to the rhythm). The use of instruments is structured to devolve to the sensory organs, the

rhythmic and melodic components of music may be used as specific stimuli to obtain certain motor and emotional responses, thus combining movement and stimulation of different sensory pathways (multi-sensory stimulation).

III. RESULTS

We used Friedman's test to compare paired data emerging from the evaluation of all presession scores (overall evaluation), within MT and CR sessions, to compare pre-session and post-session differences between the CR and MT groups, of the following measures: VMI, TGM.
The statistical tests were two-tailed at the .05 significance level.
The difference between MT pre-test and post-test values demonstrated a significant improvement in VMI and TGM scores. The difference between CR pre-test and post-test was less then MT but significant (Friedman's, *p*, .0001). Analysis of changes in Visuo-motor skills demonstrated that MT had a significant overall effect (Friedman's, *p* , .034). A comparison of pre-test and post-test differences in the VMI and TGM score values between the MT and CR sessions revealed a statistically significant effect of MT on these parameters (Friedman's, *p* , .0001).
The final evaluation, conducted one week after the sessions, demonstrated a maintained Gross Motor (TGM) and Visuomotor (VMI) benefit in MT and CR. A comparison of differences in score values between the MT and CR sessions revealed a statistically significant effect of MT on these parameters (Friedman's, *p* , .0001). At the follow up, 2 months after the sessions, demonstrated a maintained Gross Motor (TGM) benefit but a lack in Visuomotor benefit for both the therapies.
At the final interview (Parent Interview Question), all parents reported feelings of wellbeing and dynamism, saying that they were more active and keeping themselves busy at home, more after MT then CR. In particular, they said they appreciated the social contact and creative means of communication that MT offered them.





IV. DISCUSSION

This study assesses objectively the effect of a systematic program of active MT on standardized measures of WS. Moreover, compared the efficacy of MT and CR to highlight any eventual difference between the two methods in their effect on both visuomotor and emotional functions.
Our case study proves the effectiveness of MT as a possible method for the integrated rehabilitation of the WS and promises results in a more intensive training than the one tested. The active MT operates on motor functions through the integration of a multi-sensory stimulation and it is able to fulfill a need for a innovative rehabilitation intervention. Although the MT-related visualmotor response seemed to decline after two months, a trend of improvement was observed in the MT group in the overall evaluation. Moreover, the MT gross-motor effects seemed not to decrease as the time goes on. This probably occur because of the aptitude for listening and playing music in WS subject, that urges them into training after the MT session. CR has been found to be effective in patients with WS, but probably they don't understand and remind it. Generally, CR serves as reinforcement of the visual motor program, but this kind of intervention is usually lacking in the motivational and emotional spheres. It is well known, on the other hand, that psychosocial variables, such as emotional state or psychosocial stress, strongly influence the most of performances. In accordance with such observations, occupational and behavioral therapies based on psychological and motivational aspects can induce improvements in movement initiation and quality.

The beneficial effect on emotional variables valued with Parents Interview in MT may be explained by the different emotional impact that MT has on patients, which is related to its high level of sensory stimulation and high degree of personal interaction. In line with this view, our study suggests a connection between music, emotions and the facilitation of movement.

In accordance with the clinical literature, it may be argued that the MT-induced improvement could be due to the effect of external rhythmic cues, which may stabilize the movement formation process in patients with WS. Indeed, it has been demonstrated that the initiation and execution times in sequential button pressing tasks are positively influenced by acoustic cues, as are gait velocity, cadence, and stride length. Along with the rhythmic aspect of music, another factor possibly involved in motor improvement is the affective arousal effect of music, which can influence both motivational and emotional processing.

Current knowledge of the cerebral structures involved in the perception of music is derived from clinical studies and from fRMI. Listening to music seems to involve distinct neural processes that correspond to the basic components of music, such as rhythm, pitch, and timbre, or even to lexico-semantic access to melodic representations, functions that involve one or both hemispheres. Music has been shown to relax and reduce anxiety, modifying release of stress hormones, cardiac function,
and respiratory pattern.

A desirable future step will be investigating the possibilities of this MT "technique", extending this study to a wider group of individuals, and with homogeneus ages and IQ. Moreover, the ideal research would provide an opportunity to observe the peculiarities of this related neurological effects of MT, thus providing a more structured rehabilitation protocol. The results also offer that the musical model is a privileged tool to enable rehabilitation processes and thus easily convey to an enhancement of cognitive activities. The hypothesis is open in the use of methods for kinematic analysis of movement and provides examples about data size, strength, rhythm and speed of each single bio-dynamic parameter involved in the rehabilitation process induced by MT, and also a larger related study on this activation in neurological motor and visuospatial aspects.

Suggestions that music is a privileged instrument in patients with WS are not new in the clinical literature, even though they are rather scarce.

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