



American Music Therapy Association

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Williams Syndrome (WS): Recent Research on Music and Sound

STATEMENT OF PURPOSE

Description: Music Therapy (MT) is the clinical and evidence-based use of music interventions to accomplish individualized goals within a therapeutic relationship by a credentialed professional who has completed an approved music therapy program. Although WS is a rare disease, MTs may have more contact with these clients since many people with WS have a strong affinity for music, melody and song. Parents of children with WS express a strong interest in adaptive music programs for their children. The aim of therapy is to help people with WS to optimize their talents and musical affinity in order to address multiple potential outcomes. MT sessions may include the use of active music making, singing, interactive music play, and improvisational techniques. MT may include both individual and group therapy.

STANDARDIZATION: MT sessions are documented in a treatment plan and delivered in accordance with standards of practice. Music selections and certain active music making activities are modified for client preferences and individualized needs (i.e., song selection, musical instruments, and music may vary).

REPLICATION: Yes; MT and music special education has been used with different settings, providers, and populations. Research replication in the area of music response and WS is growing.

OUTCOMES: Improved global state, enhanced learning, general functioning, improved social functioning, and enhanced leisure time.

OVERVIEW OF RESEARCH

The prevalence rate for WS is estimated at 0.01% or about 36,266 people in the United States. However, reported rates vary in the literature from 1 in 7,500 up to 1 in 25,000. It is classified as a rare disease. Given the rarity of WS, the typical music therapist may only see the occasional WS client. Therefore, local applied research tends either (a) to aggregate data of WS clients with that of individuals with other developmental and genetic disorders, or (b) focus on case studies. However, more recent efforts in the field of neurobiology and genetics have launched larger scale studies gathering data on persons with WS from multiple sites nationwide. The National Science Foundation funded the first study of WS and music within the last five years. Therefore,

findings from formal research are ongoing. In addition, it is well known and documented that persons with WS tend to have a strong affinity for music. Integrating functional academic skill development within music training has resulted in enhanced understanding of mathematics for young adults with WS. This provides preliminary evidence for the use of music as a learning cue to facilitate educational outcomes.

A leading investigator examining the relationship of music, the brain and WS is Dr. Daniel Levitin, Associate Professor at McGill University. Much of his work and that of his colleagues focuses on identifying and exploring the implications of the affinity of music among persons with WS. There exists strong evidence explaining the link between the genetics of WS and neuron-cognitive processing. This basic research is essential to understanding strategies for growth, adaptation and optimal development. Trained and qualified music therapists are recommended for the WS client assessed to respond to music as part of their therapeutic and/or educational plans. This recommendation is also endorsed by the WS Association. Furthermore, some persons with WS have tendencies for hyperacusia. The music therapist can assist the entire team on the management of this sensitivity.

Claussen, D., & Hanser, S. (2000). Music-elicited responses in individuals with Williams syndrome. Paper presented at the symposium for the Society for Research in Psychology of Music and Music Education, University of Leicester, United Kingdom.

Conclusion: Based on assessment of 20 individuals with WS, researchers found enhanced musical responsiveness in areas of independence, creativity, self-confidence, engagement, and pleasure.

Deruelle, C., Schon, D., Rondan, C., & Mancini, J. (2005). Global and local music perception in children with Williams syndrome. *Neuroreport*, 16(6), 631–634.

Abstract: Musical processing can be decomposed into the appreciation of global and local elements. This global/local dissociation was investigated with the processing of contour-violated and interval-violated melodies. Performance of a group of 16 children with Williams syndrome and a group of 16 control children were compared in a same-different task. Control participants were more accurate in detecting differences in the contour-violated than in the interval-violated condition while Williams syndrome individuals performed equally well in both conditions. This finding suggests that global precedence may occur at an early perceptual stage in normally developing children. In contrast, no such global precedence is observed in the Williams syndrome population. These data are discussed in the context of atypical cognitive profiles of individuals with Williams syndrome.

Dykens, E. M., Rosner, B. A., Ly, T., & Sagun, J. (2005). Music and anxiety in Williams syndrome: A harmonious or discordant relationship? *American Journal of Mental Retardation*, 110(5), 346–358.

Abstract: In this two-part study, we assessed musical involvements in two samples of persons with Williams syndrome compared to others with mental retardation and also related musicality to anxiety and fears in Study 2. Relative to others with mental retardation, those with Williams syndrome were more likely to take music lessons, play

an instrument, and have higher ratings of musical skills. In the Williams syndrome groups only, fewer externalizing symptoms were associated with listening to music, whereas less anxiety and fewer fears were associated with the frequency, duration, and skill in producing music as well as emotional responses to negatively toned music. Implications are discussed for future research on musical processing, musical interventions, and well-being in Williams syndrome and other groups.

Fidler, D. J., & Lawson, J. E. (2003). What do parents want?: An analysis of education-related comments made by parents of children with different genetic syndromes. *Journal of Intellectual & Developmental Disability, 28*(2), 196–204.

Abstract: This study explored whether parents of children with three different genetic syndromes, Down syndrome (n=39), Prader-Willi syndrome (n=25), and Williams syndrome (n=26), express divergent desires, for modifications in their child's current educational programming. A content analysis was performed on the parents' answers to an open-ended question about how to improve their child's current placement. The parents of children with Down syndrome spontaneously expressed a greater desire for changes or improvements in speech therapy and reading services, the parents of children with Prader-Willi syndrome expressed a desire for increases in adaptive physical education services, and the parents of children with Williams syndrome expressed a desire for increases and modifications to music services and aides in the classroom. Within-syndrome variation was also found in the specific sentiments and desires expressed. Implications for a syndrome-specific approach to special education programming are discussed.

Hopyan, T., Dennis, M., Weksberg, R., & Cytrynbaum, C. (2001). Music skills and the expressive interpretation of music in children with Williams-Beuren syndrome: pitch, rhythm, melodic imagery, phrasing, and musical affect. *Child Neuropsychology, 7*(1), 42–53.

Abstract: This paper studied music in 14 children and adolescents with Williams-Beuren syndrome (WBS), a multi-system neurodevelopmental disorder, and 14 age-matched controls. Five aspects of music were tested. There were two tests of core music domains, pitch discrimination and rhythm discrimination. There were two tests of musical expressiveness, melodic imagery and phrasing. There was one test of musical interpretation, the ability to identify the emotional resonance of a musical excerpt. Music scores were analyzed by means of logistic regressions that modeled outcome (higher or lower music scores) as a function of group membership (WBS or Control) and cognitive age. Compared to age peers, children with WBS had similar levels of musical expressiveness, but were less able to discriminate pitch and rhythm, or to attach a semantic interpretation to emotion in music. Music skill did not vary with cognitive age. Musical strength in individuals with WBS 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.

Lenhoff, H. M., et al. (2001). Absolute pitch in Williams syndrome. *Music Perception, 18*(4), 491–503.

Levitin, D. J. (2002). Rhythm, pitch, timbre and hyperacusis in Williams syndrome. In C. Morris, H. Lenhoff, & P. Wang, Eds., *Williams-Beuren syndrome: Research and clinical perspectives*. Baltimore: Johns Hopkins Univ. Press.

Levitin, D. J. (2005). Musical behavior in a neurogenetic developmental disorder: Evidence from Williams syndrome. *Annals of the New York Academy of Sciences*, 1060(27), 325–334.

Levitin, D. J., & Bellugi, U. (1998). Musical ability in individuals with Williams syndrome. *Music Perception*, 15(4), 357–389.

Levitin, D. J., & Bellugi, U. (1999). Music cognition and Williams syndrome. *Journal of the Acoustical Society of America*, 106(4, Part 2), 2225.

Levitin, D. J., Cole, K., Chiles, M., Lai, Z., Lincoln, A., & Bellugi, U. (2004). Characterizing the musical phenotype in individuals with Williams syndrome. *Child Neuropsychology*, 10(4), 223–247.

Abstract: Williams Syndrome (WS), a neurodevelopmental genetic disorder, is characterized by peaks and valleys in mental function: substantial impairments in cognitive domains such as reasoning, arithmetic ability, and spatial cognition, alongside relatively preserved skills in social domains, face processing, language, and music. We report the results of a comprehensive survey on musical behaviors and background administered to the largest sample of individuals with WS to date (n = 118, mean age = 20.4), and compare the results to those obtained from a control group of typically developing normal individuals (n = 118, mean age = 20.9) and two groups of individuals with other neurodevelopmental genetic disorders, Autism (n = 30, mean age = 18.2) and Down Syndrome (n = 40, mean age = 17.2). Individuals with WS were found to be rated higher in musical accomplishment, engagement, and interest than either of the comparison groups, and equivalent on most measures to the control group. Compared to all other groups including the controls, the WS individuals displayed greater emotional responses to music, manifested interest in music at an earlier age, and spent more hours per week listening to music. In addition, the effects of music listening (whether positive or negative) tended to last longer in the WS group. A factor analysis extracted seven principal components that characterize the musical phenotype in our sample, and discriminant function analysis of those factors was able to successfully predict group membership for the majority of cases. We discuss the neurobiological implications of these findings.

Levitin, D. J., Cole, K., Lincoln, A., & Bellugi, U. (2005). Aversion, awareness and attraction: Understanding hyperacusis in Williams syndrome. *Journal of Child Psychiatry, Psychology and Allied Disciplines*, 46(5), 514–523.

Levitin, D. J., Menon, V., Schmitt, J. E., Eliez, S., White, C., Glover, G., Korenberg, J. R., Bellugi, U., & Reiss, A. L. (2003). Neural correlates of auditory perception in Williams syndrome: An MRI Study. *NeuroImage*, 18, 74–82.

Abstract: Williams syndrome (WS), a neurogenetic developmental disorder, is characterized by a rare fractionation of higher cortical functioning: selective preservation

of certain complex faculties (language, music, face processing, and sociability) in contrast to marked and severe deficits in nearly every other cognitive domain (reasoning, spatial ability, motor coordination, arithmetic, problem solving). WS people are also known to suffer from hyperacusis and to experience heightened emotional reactions to music and certain classes of noise. We used functional magnetic resonance imaging to examine the neural basis of auditory processing of music and noise in WS patients and age matched controls and found strikingly different patterns of neural organization between the groups. Those regions supporting music and noise processing in normal subjects were found not to be consistently activated in the WS participants (e.g., superior temporal and middle temporal gyri). Instead, the WS participants showed significantly reduced activation in the temporal lobes coupled with significantly greater activation in the right amygdala. In addition, WS participants (but not controls) showed a widely distributed network of activation in cortical and subcortical structures, including the brain stem, during music processing. Taken together with previous ERP and cytoarchitectonic studies, this first published report of WS using MRI provides additional evidence of a different neurofunctional organization in WS people than normal people, which may help to explain their atypical reactions to sound. These results constitute an important first step in drawing out the links between genes, brain, cognition, and behavior in Williams syndrome.

Claussen, D., & Hanser, S. (2000). Music-elicited responses in individuals with Williams syndrome. Paper presented at the symposium for the Society for Research in Psychology of Music and Music Education, University of Leicester, United Kingdom.

Conclusion: Based on assessment of 20 individuals with WS, researchers found enhanced musical responsiveness in areas of independence, creativity, self-confidence, engagement, and pleasure.

Deruelle, C., Schon, D., Rondan, C., & Mancini, J. (2005). Global and local music perception in children with Williams syndrome. *Neuroreport*, 16(6), 631-634.

Conclusion: Provides results on how individuals with WS process music compared with typically developing children.

Don, A. (1999). Auditory pattern perception in children with Williams syndrome. Dissertation Abstracts International: Section B: the Sciences & Engineering, 59(8-B), 4459.

Conclusion: The interest and emotional responsivity toward music among children with WS combined with their relatively intact music abilities suggest that the purposeful development of musical skills could help to enrich the lives of these children.

Don, A., Schellenberg, G., & Rourke, B. (1999). Music and language skills of children with Williams syndrome. *Child Neuropsychology*, 5, 154-170.

Conclusion: 19 children with WS were compared with 19 typical children. Compared to typical children, the WS group expressed greater liking of music and a greater range of emotional responses to music.

Dykens, E.M., Rosner, B.A., Ly, T., & Sagun, J. (2005). Music and anxiety in Williams syndrome: A harmonious or discordant relationship? *American Journal of Mental Retardation*, 110(5), 346-358. (Compared to others with mental retardation, individuals with WS were more likely to take music lessons, play an instrument, and have higher ratings of musical skills.)

Hickock, G., Bellugi, U., & Jones, W. (1995). Asymmetrical ability. *Science*, 270(5234), 219-220. (preliminary data suggests that brain asymmetry may account for exceptional musical abilities)

Hopyan, T., Dennis, M., Weksberg, R., Cytrynbaum, C. (2001). Music skills and the expressive interpretation of music in children with Williams-beuren syndrome: pitch, rhythm, melodic imagery, phrasing, and musical affect. *Neuropsychology, Development, and Cognition, Section C, Child Neuropsychology*, 7(1), 42-53. (study of 14 children with WS and 14 age-matched controls; musical strength in WS is linked to strong engagement with music as a means of expression, play, and perhaps improvisation)

Lenhoff, H. (1998). Insights into the musical potential of cognitively impaired people diagnosed with Williams syndrome. *Music Therapy Perspectives*, 16, 33-36. (research professor emeritus in Biology at UC Irvine gives overview of musical responses in individuals with WS)

Lenhoff, H., Perales, O., Hickok, G. (2001). Absolute pitch in Williams syndrome. *Music Perception*, 18(3), 491-503. (it appears that the incidence of "perfect pitch" is higher among individuals with WS than in the general population)

Lenhoff, H., Wang, P., Greenberg, F., & Bellugi, U. (1997). Williams syndrome and the brain. *Scientific American*, 277(6), 68-73. (overview of Williams syndrome cognitive profile with section on musical talent and relationship to neuroanatomy)

Levitin, D. (2005). Musical behavior in a neurogenetic developmental disorder: Evidence from Williams Syndrome. *Annals of the New York Academy of Sciences*, 1060, 325-334. (individuals with WS tend to be more engaged in musical activities than others, with possible neuroanatomical correlate of this engagement being increased activation in the right amygdala to music and sound.)

Levitin, D., & Bellugi, U. (1998). Musical abilities in individuals with Williams syndrome. *Music Perception*, 15(4), 357-389. (gives evidence for relatively preserved musical rhythm processing in individuals with WS based on study of 8 WS subjects and 8 controls, supporting the theory that musical ability constitutes an independent intelligence)

Levitin, DJ., Cole, K., Chiles, M., Lai, Z., Lincoln, A., & Bellugi, U. (2004). Characterizing the musical phenotype in individuals with Williams syndrome. *Neuropsychology, Development, and Cognition. Section C, Child Neuropsychology*, 10(4), 223-247. (based on a survey of 118 individuals with WS, 1 control, and 2 non-WS special needs comparison groups, individuals with WS were found to be rated higher in musical accomplishment, engagement, and interest than comparison groups, and displayed greater emotional responses to music, musical interest at an earlier age, and spent more hours per week listening to music than all other groups)

Levitin, D., Menon, V., Schmitt, J., Eliez, S., White, C., Glover, G., Kadis, J., Korenberg, J., Bellugi, U., Reiss, A. (2003). Neural correlates of auditory perception in Williams syndrome: an fMRI study. *Neuroimage*, 18(1), 74-82. (provides evidence of a different neurofunctional organization in Williams syndrome, which may help to explain their atypical reaction to sound)

Milne, H. (2002). A comparative case study of persons with Williams syndrome and musical interests. *Dissertation Abstracts International*, 62(12-A), 4057. (speaks to musical performance skills of 18 individuals with WS and discusses how development of talents and interests may be beneficial to other areas)

Reis, S.M., Schader, R., Milne, H., & Stephens, R. (2003). Music & minds: Using a talent development approach for young adults with Williams syndrome. *Exceptional Children*, 69(3), 293-313. (16 young adults with WS participated in a music and creative arts-based curriculum aligned with functional mathematics such as fractions, time, money, and measurement. Results showed an enhancement in participant's understanding of mathematics and increased opportunity to develop music abilities.)

Williams Syndrome, The Brain and Music

Source: Salk Institute

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Note: This story has been adapted from a news release issued by Salk Institute.

<http://www.sciencedaily.com/releases/2006/10/061003191006.htm> Accessed 1/23/2007.

Science Daily — Children with Williams syndrome, a rare genetic disorder, just love music and will spend hours listening to or making music. Despite averaging an IQ score of 60, many possess a great memory for songs, an uncanny sense of rhythm, and the kind of auditory acuity, than can discern differences between different vacuum cleaner brands.

A study by a multi-institutional collaboration of scientists, published in a forthcoming issue of *NeuroImage*, identified structural abnormalities in a certain brain area of people afflicted with Williams syndrome. This might explain their heightened interest in music and, in some cases, savant-like musical skill.

Professor Ursula Bellugi, director of the Laboratory for Cognitive Neuroscience at the Salk Institute for Biological Studies --the central hub of this unique scientific alliance-- explains, "Understanding the connections between missing genes, the resulting changes in brain structure and function, and ultimately behavior may help us to reveal how the brain works."

The current study is just the latest chapter in a story that's been unfolding for quite some time -- gaining increasing momentum in recent years. It all started when Bellugi reached out across disciplines and assembled a team of experts under the umbrella of a Program Project from the

National Institutes of Child Health and Human Development to help her trace the influence of individual genes on the development and functioning of the brain.

Along with co-author Albert Galaburda, a professor at the Harvard Medical School's Department of Neurology, Professor Allan L. Reiss, Director of the Center for Interdisciplinary Brain Sciences Research at Stanford University and senior author of the current study, focuses on the overall morphology of the brain, zooming in on the cellular architecture of the brain. Molecular geneticist Julie R. Korenberg, a professor in the Department of Pediatrics at UCLA, digs even deeper and studies the genes missing in people with Williams syndrome, whereas Debra Mills, an associate professor in the Department of Psychology at Emory University, concentrates on the neurophysiology, the electrical activity of behaving neural networks. Says Bellugi, who studies the cognitive aspects of the disorder: "Things are really starting to come together now."

Identified more than 40 years ago, Williams syndrome arises from a faulty recombination event during the development of sperm or egg cells. As a result, almost invariably the same set of about 20 genes is deleted from one copy of chromosome seven, catapulting the carrier of the deletion into a world where people make much more sense than objects do.

"Williams syndrome is a perfect example where a genetic predisposition interacts with the environment to sculpt the brain in unique ways," says Reiss. "It provides a unique window of understanding on how the brain develops under typical and atypical conditions," he adds.

People with Williams syndrome are irresistibly drawn to strangers, remember names and faces with ease, show strong empathy and have fluent and exceptionally expressive language. Yet, they are confounded by the visual world around them: While they can't scribble more than a few rudimentary lines to illustrate an elephant, they can verbally describe one in almost poetic detail.

"The discrepancy between their engaging social use of language and their poor visual-spatial skills is startling," says Bellugi. "I am confident that once all the evidence is in, we will have identified genes and pathways in the Williams syndrome deletion that underlie these drastic differences in modalities," she adds.

Despite whole brain volumes that are about 15 percent smaller than normal, the temporal lobe, which lies above the ear canal and, among other things, is involved in processing sounds and interpreting music and language, is of approximately normal volume in people with Williams syndrome. In their study, the researchers tried to answer the question of whether an atypical development of the planum temporale, which is part of the temporal lobe and thought to be involved in many auditory tasks, including perfect pitch, may underlie the unusual musical and language skills.

First author Mark Eckert, formerly at Stanford and now an assistant professor at the Medical University of South Carolina, and his colleagues used data from brain scans of 42 individuals with Williams syndrome and 40 control participants to compare the surface folds of the planum temporale. In most people, the structure, a slender inch-long piece of tissue, is larger on the left side of the brain than the right.

In people with Williams syndrome, however, both sides tended toward symmetry. "There are different possible explanations: Either the left side didn't grow enough or the right side grew larger than usual," says Galaburda. The folding pattern, in particular one groove called the Sylvian fissure, pointed to an increase in size of the right planum temporale.

But size alone might not explain the unusual auditory strengths of people with Williams syndrome. A more general explanation includes variations in the connectivity of certain brain regions that might contribute to the specific strengths and weaknesses in Williams syndrome."

In recent studies, Galaburda had found that cells in the primary visual cortex of carriers of the Williams deletion are smaller and more densely packed -- allowing for fewer connections between cells. Neurons in the primary auditory cortex, on the other hand, were larger and loosely packed, denoting increased "connectedness."

"These differences in cell size and density may underlie the strengths in auditory phonology, language and possibly music, and the difficulties in visual spatial construction for primary visual areas," says Bellugi, adding, "This is really just part of the overall effect of the genes' deletion on brain development."

"Relatively subtle developmental defects can have a significant impact on neurological function," says Dennis O'Leary, a Salk neurobiologist who studies the development of the visual system. "This work opens the door to explaining how genes work through the brain and make us who we are," he adds.

The Salk Institute for Biological Studies in La Jolla, California, is an independent nonprofit organization dedicated to fundamental discoveries in the life sciences, the improvement of human health and the training of future generations of researchers. Jonas Salk, M.D., whose polio vaccine all but eradicated the crippling disease poliomyelitis in 1955, opened the Institute in 1965 with a gift of land from the City of San Diego and the financial support of the March of Dimes.