

## Characterizing the Musical Phenotype in Individuals With Williams Syndrome

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### 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.

### INTRODUCTION

Two of the most fascinating unsolved puzzles in cognitive neuroscience concern the neuroanatomical basis for music cognition and the architecture of cognitive function in the neurodevelopmentally impaired. Interest in the cognitive neuroscience of music perception, cognition, memory, and performance has become a central and compelling question in recent years (Peretz & Coltheart,

2003; Sergent, 1993; Zatorre, 2003). Part of the reason for this interest is that music is marked by its ubiquity and its antiquity – nearly all known human cultures have music, some of the earliest human-made artifacts discovered are of musical instruments (Cross, 2001; Huron, 2001), and in the present day, music plays a central role in the lives of most of us (Sloboda, 1999). Yet fundamental issues in music cognition research remain unsolved, and the results of investigations –

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especially neuropsychological ones – are contradictory (Hodges, 1996). We still cannot say, for example, whether or not a given individual has “musical talent” in a meaningful or quantifiable way, both because definitions and assessment procedures are elusive. And efforts to fractionate musical behaviors into their functional neuroanatomical components have generally not met with the success seen in other domains (Marin & Perry, 1999; Peretz & Coltheart, 2003), such as language (Patel, 2003), mathematical ability (Dehaene, 1996) or visual perception (Zeki, 1993), and this has raised still more questions about the relation between music and other cognitive processes.

The other puzzle, involving the cognitive architecture among those with neurodevelopmental impairments, is also rich with promise yet without clear findings thus far. The study of distinct, well-defined, and atypical populations is important because it offers a unique opportunity to investigate specific aspects of cognition, and to establish the degree to which various cognitive abilities are correlated with, or can be decoupled from one another (Burack, 1997). In particular, the study of populations with genotypic abnormalities (including Down’s Syndrome, Autism, and Williams Syndrome) have sparked anew debates regarding the modularity of brain function, independence of mental faculties, and theories of neural organization (Don, Schellenberg, & Rourke, 1999; Karmiloff-Smith, 1998; Tager-Flusberg & Sullivan, 2000). Indeed, one of the newest and most exciting theories in cognitive neuroscience, the neuro-constructivist view, is strongly inspired by these special populations, and in particular by Williams Syndrome (Karmiloff-Smith, 1998; see also Elman et al., 1996; Quartz & Sejnowski, 1997). Individuals with Williams Syndrome are unique among those with neurogenetic developmental disorders in presenting widespread cognitive impairment which reputedly does not show up in either language or music (Bellugi, Lichtenberger, Jones, Lai, & St. George, 2001; Levitin & Bellugi, 1998, 1999; Levitin et al., 2003).

Seemingly unrelated, the two topics of music cognition and neurogenetic developmental disorders have very recently come together in the study of individuals with WS. Researchers seek to better understand the nature of the cognitive

strengths and deficits in Williams syndrome, and how these compare to members of other populations. In particular, people with WS present a mysterious pattern of peaks and valleys in mental and motor function. The emerging evidence suggests that cognitive and motor function with respect to musical activities remain relatively preserved in WS, despite the presence of gross impairments in cognitive domains that would seem to be subserved by the same neural circuitry (Bellugi, Korenberg, & Klima, 2001; Levitin & Bellugi, 1998; Levitin et al., 2003).

This article reports on the third stage of a systematic research program to study music cognition, perception, performance, and listening in WS. The first stage, a preliminary investigation of rhythm perception, was published as Levitin & Bellugi (1998), the second stage, a neuroimaging study of music and noise perception was published as Levitin et al. (2003). The present work is an attempt to quantify and characterize the WS musical profile based on a detailed questionnaire completed by parents of WS individuals. For comparison purposes, the questionnaire was also administered to parents of typically developing normal individuals, as well as individuals with Autism and Down syndrome (two other neurogenetic developmental disorders that result in distinctive types of mental impairment). Questionnaire studies have previously been conducted with the WS population (Don et al., 1999; Klein, Armstrong, Greer, & Brown, 1990; Udwin, 1990). This questionnaire differs from previous ones in several significant respects including a larger sample, the use of three comparison groups (Down Syndrome, Autism, and normals), and its more extensive topic coverage.

### **Background on Williams Syndrome**

Over the past decade, research on WS has increased at a rapid rate as this disorder has come to be regarded as one of the most compelling genetic models of human cognition (Korenberg, Chen, Hirota, Lai, Bellugi, Burian, Roe, & Matsuoka, 2000). WS – also known as Williams-Beuren Syndrome (WBS; Beuren, Schulze, Eberle, Harmjan, & Apitz, 1964) – is a neurogenetic developmental disorder occurring in approximately 1 in 20,000 live births, and is caused by

the hemizygous deletion of approximately 17 genes on chromosome 7 (band 7q11.23) between the polymorphic markers D7S1816 and D7S489B (Francke, 1999), including the genes for elastin (ELN), LIM1kinase (LIMK1), Frizzled (FZD9, previously called FZD3), and Syntaxin1A (STXN1A), and representing 1.6–2 million missing base pairs (Francke, 1999; Frangiskakis et al., 1995; Korenberg et al., 2000). A number of physical abnormalities are believed to be secondary to the deletion of ELN at 7q11.23 including infantile hypercalcaemia (Udwin, 1990), supra-valvular aortic stenosis (SVAS; Fanconi, 1952; Williams, Barratt-Boyes, & Lowe, 1961), a particular craniofacial dysmorphology (Hovis & Butler, 1997), and scoliosis (Hagerman, 1999). The gene for DNA replication factor C2 (RFC2) and STXN1A are now believed to affect the release of neurochemicals, and FZD3 affects cell signaling during neurodevelopment (Karmiloff-Smith, 1998; Korenberg, Bellugi, Salandanan, Mills, & Reiss, 2003). In the few reported cases of offspring reproduction involving WS individuals, the heritability of WS appears to be 0.5.

WS is characterized by low IQ, ranging from 40 to 100 (mean  $\sim 61$ ,  $SD 11$ , Bellugi, Korenberg, & Klima, 2001, p. 9; for slightly different estimates see also Karmiloff-Smith, 1998; Morris & Mervis, 1999). WS individuals also present deficits in key cognitive domains including conceptual reasoning (Bellugi, Klima, & Wang, 1996; Bellugi, Lichtenberger et al., 2001), problem solving, arithmetic, and spatial cognition (Bellugi, Korenberg, & Klima, 2001; Frangiskakis et al., 1995). As noted above, the most intriguing aspect of the WS cognitive profile is the presence of relatively *strong* abilities in four specific domains: *social drive* (Gosch & Pankau, 1997; Jones et al., 2001; Losh, Bellugi, Reilly, & Anderson, 2000; Tager-Flusberg, Sullivan, Boshart, Guttman, & Levine, 1996; Udwin & Yule, 1991), *face processing* (Bellugi, Korenberg et al., 2001; Mills et al., 2001; Paul, Stiles, Passorotti, Bavar, & Bellugi, 2002; Pezzini, Vicari, Volterra, Milani, & Ossella, 1999), *language* (Bellugi, Korenberg et al., 2001; Mervis, Morris, Bertrand, & Robinson, 1999; Volterra, Capirci, Pezzini, Sabbadini, & Vicari, 1996), and *music* (Don et al., 1999; Hopyan, Dennis, Weksberg, & Cytrynbaum, 2001; Lenhoff, Wang,

Greenberg, & Bellugi, 1997; Levitin & Bellugi, 1998, 1999). Because the deletion is known in WS, and the phenotypic manifestations are relatively well-defined and stable among members of the group, WS presents a unique opportunity to uncover the neurobiological basis of complex cognitive behaviors, and in particular, to draw out the links between genes, brain, cognition, and behavior.

### Previous Studies of Williams and Music

Most of the evidence about the musical abilities of individuals with WS has come from anecdotal reports. To date, only three published papers have addressed their musical abilities. In a preliminary study, Levitin and Bellugi (1998) tested rhythmic skill in an echo clapping task, and found that WS were commensurate with mental-age-matched, typically developing controls (CTLs) in their ability to reproduce musical rhythms. One notable difference was that for those trials on which participants made errors, the WS were far more likely than typically developing normals to produce errors that were *musically compatible* with the example rhythm, what we called “creative completions.” Double-blind, professional musician raters had noticed this difference without coaching, and we interpreted this result as one index of the overall *musicality* of WS.

Don et al. (1999) compared the music and language skills in a small group of children with WS ( $n = 19$ , ages 8–13) with typically developing normal controls ( $n = 32$ , ages 5–12). The researchers employed a variety of standard instruments, such as the Peabody Picture Vocabulary Test – Revised (PPVT-R; Dunn & Dunn, 1981), the Auditory Closure Test (Kass, 1964), the *Digit Span* subtest from the Wechsler Intelligence Scale for Children (WISC-III; Wechsler, 1991), the Primary Measures of Music Audition (PMMA; Gordon, 1986), and two questionnaires they designed in order to elicit information from children and parents about the childrens’ musical interests, activities, knowledge, and environment. WS and controls were found to have comparable musical backgrounds and environments, and equivalent histories of creating music. Both groups reported that music could make them happy, whereas the WS group reported a significantly

greater propensity for music to make them feel sad. Moreover, the WS group were rated to have significantly greater *interest* in music than the CTLs. Finally, Don et al. found significant differences between groups for ratings of both *hyperacusis* (sensitivity to sound) and *hypertimbria* (unusual attraction to or liking of certain sounds), with the WS being far more likely to present both. Levitin et al. (in press) confirmed the incidence of hyperacusis in individuals with WS, and further fractionated their auditory experience into three additional components or symptoms: auditory allodynia, odynacusis, and auditory fascinations. WS were found to suffer from all four of these symptoms more often than individuals with DS, AUT, or typically developing normal controls.

On the standardized tests, Don et al. found that WS and CTLs scored similarly on the PMMA tonal tests, whereas the WS group scored significantly worse on the rhythm test. Hopyan et al. (2001) administered the same test to 14 children with WBS (mean age 12, *SD* 3) and 14 chronologically-aged matched controls (mean age 12, *SD* 3) and reported that the control group performed significantly better than the WBS group on the rhythm test. While on the surface this may seem to contradict the earlier findings of Levitin and Bellugi (1998), three explanations present themselves. First, as Don et al. point out, the rhythm subtest is always administered *after* the tonal (melody) subtest in accordance with Gordon's (1986) standardized testing procedures. WS are known to have attentional deficits, and Don et al. note that their WS participants found it increasingly difficult to pay attention on the second of these two tests (an observation we have confirmed in our own laboratories). Second, there is evidence that the PMMA contains manufacturing defects (Levitin & Bellugi, 1999) that could actually penalize a careful listener. A third and intriguing possibility has to do with the demand characteristics of the experiment and social facilitation. In a separate study, we administered a modified version of the PMMA to a group of 20 WS individuals (Levitin & Bellugi, 1999) and found that they had a great deal of difficulty paying attention to the test *when it was administered from a recording*. Our subjects seemed to lack any sense of engagement with

the recorded experimenter, yet when those same items were administered *in person* by the experimenter, the subjects' performance increased. It may simply be the case that WS – being unusually interested in social interaction (Jones et al., 2000) – perform poorly on standardized tests that are administered from a computer or recording.

Three limitations of the Don et al. and Hopyan et al. studies therefore were the relatively small sample size, the small number of questionnaire items, and the fact that they did not employ comparison groups of individuals with other forms of mental retardation. A first reasonable null hypothesis would be that the observed results are in some way related to neurodevelopmental impairment or to developmental delay. We included individuals from two neurodevelopmentally impaired groups, Autism and Down Syndrome, as well as age-matched controls. We used the Don et al. and Hopyan et al. studies as a starting point, building on them to develop a more comprehensive and detailed study.

## METHODS

### Subjects

Questionnaires were administered to the parents of individuals with WS, Autism (AUT), and Down Syndrome (DS), as well as to typically developing normal control subjects (CTLs).

Parents of WS subjects ( $n = 130$ ) were solicited from William Syndrome Association National Conferences and from the Laboratory for Cognitive Neuroscience at the Salk Institute for Biological Studies in California. All WS subjects were positively diagnosed by the florescent insitu hybridization test (for absence of one copy of the gene for elastin on chromosome 7, the FISH test), or by the WS Diagnostic Score Sheet (DSS; American Academy of Pediatrics, 2001). Of 149 people with WS that we originally recruited, 118 were positively diagnosed by the above criteria and thus retained in the present report. The mean full scale IQ of our retained sample was 66 (*SD* 11). Although this is slightly higher than the IQ obtained in some studies, it is well within the expected range of sampling variability and is not significantly different from that obtained in previous studies.

Parents of DS participants ( $n = 40$ ) were solicited from ongoing studies at the Laboratory for Cognitive Neuroscience at the Salk Institute in San Diego, California. The diagnosis of DS was positively

confirmed by genetic testing for trisomy 21. Mean full scale IQ was 56 ( $SD$  9.1).

Parents of AUT participants ( $n = 30$ ) were solicited from the Developmental Neuropsychology Laboratory at the Alliant International University in San Diego, California. All AUT participants were positively diagnosed by trained neuropsychologists prior to their participation in the study using a standardized diagnostic battery that included the Autistic Diagnostic Interview – Revised (ADIR), Autistic Diagnostic Observation Schedule (ADOS), and Childhood Autism Rating Scale (CARS). Mean full scale IQ was 74.5 ( $SD$  27.8).

CTL participants ( $n = 118$ ) were obtained from an undergraduate psychology class at San Diego State University in California. They were instructed to consult with their parents in completing this questionnaire and were reminded that the questions referred to their childhood history, not their present experience. Some younger siblings and children of students enrolled in the class completed the questionnaire and were also included in the study. All subject groups completed questionnaires on a volunteer basis and, in addition, CTL subjects received extra class credit for completing the questionnaire. The age and gender distribution of the subjects by diagnosis are shown in Table 1. An ANOVA confirmed that there were no statistically significant differences in age between groups,  $F(3, 302) = 2.1$ , n.s.

## Materials

The questionnaire contained 46 items, 33 multiple choice (including Likert scales) and 13 free response. The free-response items were mapped to discrete categories during data coding by double-blind raters. The types of questions asked clustered into six categories:

- (1) Demographic (diagnosis, age, sex, handedness) and physical profile (hearing loss, physical deficits)
- (2) Interest in music (whether the child plays an instrument, hours per week spent listening, playing, etc.)
- (3) Emotional responses to music

- (4) Musical creativity and reproduction (how often the child reproduces music, or makes up music, and in the parents' view, the quality of these)
- (5) Musical training
- (6) Age of onset (when the child first began to manifest certain musical behaviors)

A more detailed description of the questions completes this section, and the full questionnaire is included here as Appendix A.

The *Interest in Music* section first assesses the subject's overall level of musical involvement by employing an item from Grison's (1972) Levels of Musical Culture, which were originally designed to rate premorbid levels of musicality in patients with hemispheric brain disease. Subsequent questions summarize the degree of interest the subject has in comparison to their peers, a description of the types of musical activities the subject is involved in, and an estimate of the hours per week spent on all music related activities. The section on *Emotional Responsiveness* characterizes the subject's general emotional responsiveness towards music, and probes specific emotional qualities within music to determine whether any specific emotional patterns exist. This section also probes the length of time emotional reactions last in subjects. The series of questions on *Musical Creativity and Reproduction* address complexity, frequency, accuracy, number of songs or pieces of music reproduced, and specific patterns of recall. These items are specifically vague in order to include the many forms of musical productions a subject might make. The only criteria for musical reproductions are that they must be of a previously heard song or piece of music, and they must incorporate tonal qualities. Subsequent sections concerning rhythmic production and original musical creations consist of items rating the frequency and complexity of original productions, in addition to a description of typical musical creations. The series of questions on *Music Training* address whether an instrument has ever been played, the frequency of playing instruments, and the amount of formal training in music theory received.

Table 1. Age and Gender Distribution of Subjects in this Study.

|                                           | AUT         | Control    | DS         | WS          |
|-------------------------------------------|-------------|------------|------------|-------------|
| <i>n</i>                                  | 30          | 118        | 40         | 118         |
| Chronological age: <i>M</i> ( <i>SD</i> ) | 18.2 (7.7)  | 20.9 (7.4) | 17.2 (9.2) | 20.4 (10.4) |
| Range                                     | 9–39        | 5–44       | 5–51       | 5–50        |
| Mental age: <i>M</i> ( <i>SD</i> )        | 74.5 (27.8) | 110 (10)*  | 56 (9.1)   | 66 (11)     |
| Male                                      | 24          | 28         | 20         | 61          |
| Female                                    | 6           | 90         | 20         | 57          |

Note. \*Not measured. We assume the mean of a sample of San Diego State University students is probably 110 and *SD* is likely no higher than 10.

*Age of onset* comprised questions addressing when the child first took an interest in playing an instrument or listening to music.

### Analyses

The quantitative data obtained from the questionnaire (e.g., Likert scale scores and multiple-choice scores) were analyzed using either Chi-square tests (for nominal data such as handedness) or analysis of variance (for ordinal data, such as hours spent listening to music per week), with planned orthogonal contrasts to test differences between group means. All tests were adjusted for multiple comparisons. The free response data were transformed into nominal categories by three double-blind raters who were trained to score the presence or absence of characteristics in specific domains of interest. We used Kappa as a measure of interrater agreement setting a threshold of 0.75 (as recommended by Landis & Koch, 1977). The Kappa values obtained for interrater reliability in this study were all well above this level ( $\kappa = 0.80$  to  $1.00 \pm SE$  ( $\kappa = 0.09$ ) for all items measured.

## RESULTS

### Reliability

First, to assess the reliability of the questionnaire, we performed a split-subjects reliability analysis (by randomly assigning subjects to one of two arbitrary groups) and found no significant difference between the halves,  $F(1, 305) = .16$ ,  $p \sim .69$ . We performed additional split-subjects reliability tests on the seven broad content areas extracted by factor analysis (the extraction procedure and results are described below in the section entitled "Factor Analysis") and again found no significant between the halves (For the seven factors, all with  $df = (1, 283)$ :  $F_1 = .342$ ,  $p \sim .56$ ;  $F_2 = .812$ ,  $p \sim .37$ ,  $F_3 = .11$ ,  $p \sim .75$ ;  $F_4 = 2.1$ ,  $p \sim .15$ ,  $F_5 = .09$ ,  $p \sim .76$ ,  $F_6 = 1.8$ ,  $p \sim .18$ ,  $F_7 = 1.3$ ,  $p \sim .26$ ).

### Group Differences

An ANOVA showed significant intergroup differences for 10 of the items: the Grison profile,  $F(3, 301) = 5.5$ ,  $p < .001$ , Musical Interest Amount,  $F(3, 298) = 9.9$ ,  $p < .001$ , Musical Interest Age of Onset,  $F(3, 271) = 13$ ,  $p < .001$ , Emotional Response to Music,  $F(3, 299) = 12.2$ ,

$p < .001$ , Reproduction Accuracy,  $F(3, 294) = 9.1$ ,  $p < .001$ , Frequency of Spontaneous Rhythmic Productions,  $F(2, 283) = 8.3$ ,  $p < .001$ , Frequency of Original Music Productions,  $F(3, 277) = 5.1$ ,  $p < .002$ , Playing a Musical Instrument,  $F(3, 297) = 8$ ,  $p < .001$ , Hours Per Week Spent Playing a Musical Instrument,  $F(3, 284) = 5.7$ ,  $p < .001$  and Exposure to Music Theory,  $F(3, 279) = 9.6$ ,  $p < .001$ . No other tests were significant, so we report the planned tests of intergroup differences only on these 10 items. [The degrees of freedom are not identical for each comparison due to missing data. For example, if a parent felt that their child had shown no particular interest in music, they would have left the "age of onset of musical interest" item blank.]

### Planned Comparisons

WS and DS individuals were reported to show significantly earlier evidence of first musical interest than controls ( $p < .001$  adjusted;<sup>1</sup> Table 2). WS individuals listened to music an average of 3 hr more per week than controls ( $p < .001$ ), and 2–3 hr a week more than DS and AUT, respectively, although these latter differences did not reach significance ( $p \sim .2$ ; but see note below under the next section about binomial comparisons).

The overall level of musical involvement, as assessed by the Grison scale (which combines music listening, instrument playing, and music theory) found WS involvement to be significantly greater than that of the DS and AUT groups ( $p < .05$ ) and similar to CTL ( $p \sim .9$ ). Also significantly higher than controls in musical involvement were the AUT ( $p < .02$ ) and DS ( $p < .01$ ).

Parents of WS reported a higher level of interest in music-related activities than the CTL ( $p < .001$ ) and AUT groups ( $p < .02$ ) and an equivalent level of interest to DS ( $p = 1$ ). Individuals with DS showed somewhat more interest in music than those with AUT ( $p \sim .08$ ), and significantly more interest than controls ( $p < .05$ ). WS also were reported to experience significantly more emotion when listening to music than CTLs

<sup>1</sup>All  $p$  values reported for the contrasts are the adjusted  $p$  values for multiple comparisons.

Table 2. Planned Orthogonal Contrasts of Intergroup Differences for 10 Items on the Questionnaire. Values in the Cells are the Means (and Standard Deviations) for Each Item. Significance Level is  $p < .05$  for all Pairs, Adjusted for Multiple Comparisons.

| Item                                          | CTL                      | AUT                      | DS                       | WS                       |
|-----------------------------------------------|--------------------------|--------------------------|--------------------------|--------------------------|
| Grison Profile                                | 3.5 <sup>a,d</sup> (1.1) | 2.7 <sup>c</sup> (1.1)   | 2.9 <sup>c,w</sup> (.74) | 3.4 <sup>d</sup> (1.1)   |
| Musical interest amount                       | 4.9 <sup>d,w</sup> (1.3) | 4.6 <sup>w</sup> (1.9)   | 5.7 <sup>c</sup> (1.5)   | 5.8 <sup>a,c</sup> (1.4) |
| Musical interest – age of first onset         | 5.0 <sup>d,w</sup> (3.6) | 4.4 (4.0)                | 2.3 <sup>c</sup> (2.2)   | 2.7 <sup>c</sup> (2.6)   |
| Emotional response to music                   | 4.8 <sup>w</sup> (1.1)   | 4.0 <sup>w</sup> (2.0)   | 5.1 (1.3)                | 5.7 <sup>a,c</sup> (1.4) |
| Reproduction accuracy                         | 4.4 <sup>a,d</sup> (1.5) | 3.2 <sup>c</sup> (2.2)   | 3.2 <sup>c,w</sup> (1.5) | 4.4 <sup>d</sup> (1.8)   |
| Frequency of spontaneous rhythmic productions | 4.0 <sup>a,d</sup> (1.8) | 2.1 <sup>c,w</sup> (1.8) | 2.9 <sup>c</sup> (2.1)   | 3.6 <sup>a</sup> (2.2)   |
| Frequency of original music productions       | 3.1 <sup>a</sup> (1.5)   | 1.8 <sup>c,w</sup> (1.7) | 2.6 (1.7)                | 3.2 <sup>a</sup> (2.0)   |
| Musical instrument                            | .83 <sup>a,d</sup> (.38) | .50 <sup>c,w</sup> (.51) | .56 <sup>c</sup> (.50)   | .80 <sup>a</sup> (.40)   |
| Hours playing instrument (per week)           | 1.5 <sup>w</sup> (2.4)   | 1.2 (2.6)                | .82 <sup>w</sup> (1.6)   | 1.9 <sup>c,d</sup> (3.1) |
| Music theory                                  | .51 <sup>a,d</sup> (.50) | .12 <sup>c,w</sup> (.33) | .12 <sup>c,w</sup> (.33) | .35 <sup>a,d</sup> (.48) |

Note. <sup>a</sup>Significantly different from autistic group.

<sup>c</sup>Significantly different from control group.

<sup>d</sup>Significantly different from DS group.

<sup>w</sup>Significantly different from WS group.

or AUT participants ( $p < .001$ ) and slightly more than DS individuals ( $p \sim .08$ ). In spontaneous reproductions of familiar music, WS and CTLs both reproduced music similarly well, and more accurately than the DS individuals ( $p < .001$ ). CTLs were significantly better than AUT ( $p < .05$ ) and WS were better than AUT ( $p < .06$ ).

WS and CTLs tended to play rhythmic patterns as often as one another, and more often than individuals with AUT ( $p < .005$ ). In addition, the CTLs played rhythms more often than the DS ( $p < .04$ ), and although the mean rating for WS versus DS was higher, it did not reach statistical significance ( $p \sim .4$ ). The frequency with which individuals spontaneously invented their own music (melodies and rhythms together) was significantly greater for WS and CTLs versus the individuals with AUT ( $p < .01$ ), and the CTL group had greater frequency than the DS group ( $p < .03$ ). As before, the mean for the WS was higher than that for the DS, but not statistically so by the ANOVA (see note below,  $p \sim .3$ ).

Both the CTLs and the WS group were significantly more likely to play musical instruments than the individuals with AUT ( $p < .02$  and  $p < .04$ , respectively). In addition, CTLs were more significantly more likely and WS more slightly more likely to play an instrument than

DS ( $p < .02$  and  $p > .06$ , respectively). The WS played their musical instruments more hours per week than the CTL ( $p < .03$ ) or DS groups ( $p < .001$ ) and marginally more than the AUT group ( $p < .13$ ) although, it is important to note that this latter comparison is driven by a relatively small number of AUT individuals in our sample who played an instrument at all (14), and those who did tended to play it a lot. Finally, both the WS and CTL groups were more likely to have had music theory than the AUT or DS groups. (WS vs. AUT,  $p < .03$ , WS vs. DS,  $p < .02$ ; CTL vs. AUT and CTL vs. DS,  $p < .001$ ). The CTL individuals were slightly more likely to have had music theory than the WS ( $p < .08$ ).

#### *Binomial Comparisons Between WS and Other Neurodevelopmentally Impaired Individuals*

Of the 10 items in the previous section for which intergroup differences were identified by ANOVA, post hoc tests revealed that the WS individuals were statistically higher on 4 of the measures than DS individuals. However, a careful examination of Table 2 reveals that in fact the means for the WS group were in the direction of increased musicality for 9 of the 10 items compared to the DS group (note that with the exception of “age of

onset of first musical interest,” higher means represent more musicality for all of these measures, and DS in fact were reported to show a lower age of onset for musical interest). We can consider this as a binomial experiment (formally equivalent in this case to a sign test) and ask what is the probability that the WS means would all show an effect in the same direction compared to the DS means on at least 9 out of 10 items? This is exactly  $[(10!)(.5^9)(.5^1)/9!1!] + (10!)(.5^{10})(.5^0)/10!0!]$ , or  $p \sim .01$ . Thus we can conclude that for these 10 measures, WS were in fact significantly different (higher) than the DS group. The same holds true for a direct comparison of the WS and AUT group: by binomial test, the WS group was significantly different (in the direction of “more musical”) than the AUT group at  $p \sim .01$ . By this same test, there were no significant differences found between DNS and AUT.

#### Physical Characteristics

We found differences in handedness with the parents reporting that DS and WS were both more likely ( $p < .02$ ) to be left handed or ambidextrous than the control group (Table 3). DS were more likely than other groups to have suffered from hearing loss (Table 4), with WS more likely than AUT or CTLs to have also suffered hearing loss

Table 3. Reports of Handedness by Diagnosis (DX) and Statistically Significant Differences at  $p < .02$ .

| Sig  | DX  | Left (%) | Ambi. (%) | Right (%) |
|------|-----|----------|-----------|-----------|
| D, W | CTL | 6.8      | 0.8       | 92.4      |
|      | AUT | 10.0     | 0.0       | 90.0      |
| C    | DS  | 17.1     | 1.7       | 81.2      |
| B    | WS  | 20.0     | 7.5       | 72.5      |

Table 4. Reports of Hearing Loss by Diagnosis (DX) and Statistically Significant Differences at  $p < .01$ .

| Sig     | DX  | Hearing loss (%) |
|---------|-----|------------------|
| D, W    | CTL | 4.2              |
| D, W    | AUT | 0.0              |
| C, A, W | DS  | 43.6             |
| C, A, D | WS  | 17.8             |

Table 5. Reports of Physical Deficits by Diagnosis (DX) and Statistically Significant Differences at  $p < .01$ .

| Sig                    | DX  | Physical deficits |
|------------------------|-----|-------------------|
| A, D, W<br>C<br>C<br>C | CTL | 1.7 (%)           |
|                        | AUT | 13.8              |
|                        | DS  | 15.0              |
|                        | WS  | 15.9              |

( $p < .01$ ). The three neurodevelopmentally impaired groups had similar levels of reported physical deficits to one another, and more than the CTL group ( $p < .01$ ; Table 5).

#### Factor Analysis

By design, our questionnaire contained a number of items that attempted to index similar latent qualities of musical behavior and therefore one would expect some items to be significantly correlated with others. Factor analysis can reduce the 33 multiple choice items to a smaller, more manageable number, while at the same time bringing together items that in fact index the same underlying attributes. We performed an exploratory factor analysis and settled on seven orthogonal components which were rotated by the varimax algorithm, and which together account for 67% of the variance (and use only Eigenvalues greater than 1, see Fig. 1). The seven factors were truly orthogonal, and intercorrelations among the

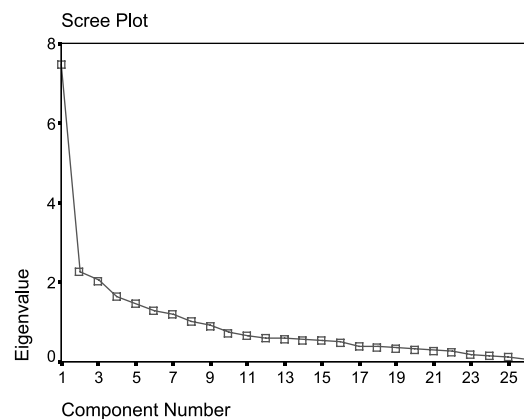


Fig. 1. Eigenvalues for the 33 questionnaire scale items.

Table 6. Factor Analysis Solution of the 26 Multiple Choice Items on the Questionnaire. Seven Principal Components were Extracted Accounting for 67% of the Variance.

| Rotated component matrix |                                               | Component |      |      |      |      |      |      |                       |
|--------------------------|-----------------------------------------------|-----------|------|------|------|------|------|------|-----------------------|
|                          |                                               | 1         | 2    | 3    | 4    | 5    | 6    | 7    |                       |
| I                        | Frequency of original music production        | .81       | .15  | .15  | .08  | .00  | .03  | .06  | Spontaneity           |
|                          | Frequency of spontaneous rhythmic productions | .81       | .12  | .18  | .13  | .03  | .14  | -.08 |                       |
|                          | Rhythmic complexity                           | .78       | .11  | .32  | .06  | -.08 | .02  | .09  |                       |
|                          | Original music complexity                     | .74       | .09  | .34  | -.03 | -.12 | .03  | .20  |                       |
|                          | Original music description                    | .71       | .11  | .05  | .25  | .13  | .08  | .04  |                       |
|                          | Hours/week playing instrument                 | .49       | .36  | .08  | .35  | .11  | -.13 | -.05 |                       |
| II                       | Reproduction length                           | .16       | .77  | .09  | .05  | .01  | .06  | .09  | Reproduction of Music |
|                          | Reproduction complexity                       | .25       | .77  | .16  | .11  | -.11 | .12  | .12  |                       |
|                          | Reproduction number                           | .20       | .67  | .04  | .11  | .31  | .14  | .14  |                       |
|                          | Reproduction frequency                        | .31       | .65  | .33  | .16  | -.08 | -.04 | .08  |                       |
|                          | Reproduction accuracy                         | .35       | .63  | .11  | .29  | .22  | -.02 | -.08 |                       |
| III                      | Emotions and listening                        | .21       | .11  | .77  | .12  | .02  | .10  | .16  | Listening Habits      |
|                          | Musical interest amount                       | .15       | .22  | .77  | .14  | .02  | .09  | -.04 |                       |
|                          | Positive reactions to happy music             | .18       | .08  | .64  | -.04 | .03  | .38  | .13  |                       |
|                          | Music Listening Hours/week                    | .18       | .33  | .59  | .10  | -.03 | .14  | -.05 |                       |
| IV                       | Exposure to music theory                      | .01       | .18  | -.05 | .80  | .03  | .10  | .12  | Theory, Achievement   |
|                          | Playing a musical instrument                  | .25       | .13  | .04  | .68  | .00  | .00  | .00  |                       |
|                          | Grison Profile                                | .27       | .28  | .27  | .59  | -.18 | .01  | -.11 |                       |
| V                        | Reproduction age of onset                     | -.03      | .12  | -.02 | .06  | .79  | .06  | -.04 | Age of Onset          |
|                          | Interest age of onset                         | -.02      | .13  | .37  | -.19 | .69  | -.16 | -.07 |                       |
| VI                       | Negative reactions to sad music               | .22       | .08  | .27  | -.02 | .13  | .82  | .13  | Negative Reactions    |
|                          | Negative reactions to happy music             | -.03      | .19  | -.07 | .04  | -.34 | .82  | -.11 |                       |
| VII                      | Happy music carryover                         | .13       | .05  | .23  | -.1  | -.06 | -.10 | .82  | Sensitivity           |
|                          | Sensitivity to sound                          | -.04      | -.06 | .38  | -.02 | .19  | .07  | -.58 |                       |
|                          | Sad music carryover                           | .02       | .15  | -.01 | .22  | .03  | .44  | .59  |                       |
|                          | Positive reactions to sad music               | -.12      | .33  | .25  | -.03 | -.49 | .11  | .49  |                       |

factors were all zero. We explored as alternatives a six factor solution (accounting for 62% of the variance) and an eight factor solution (accounting for 71% of the variance) but the rotated component matrix made more sense theoretically with seven factors, and in fact corresponded quite well to our initial *a priori* categories (as outlined in the *Methods* section). The difference between the factor analytic solution and our *a priori* categories is that the factor analysis split off two meaningful categories or factors from our initial conception, one for sensitivity to music and another for negative reactions to music. The six factor solution placed, what seemed to us to be conceptually separate items, in the same component for factor number 5 (negative reactions to happy music, reproduction age of onset, interest age of onset, and positive reactions to sad music). The seven factor solution separated these out. The final rotated components matrix with factor interpretations are presented in Table 6, and the hierarchical structure of the factors is presented in Figure 2.

Figure 2 displays the hierarchical structure of the seven factors extracted, beginning with the first unrotated principal component (FUPC). Note that Factors I (Spontaneity) and III (Listening Habits) split off relatively early, and stay virtually the same all the way to the end of the hierarchy. Factor II (Reproduction) breaks away at Level 5 and remains virtually unchanged through the next two iterations.

We performed an ANOVA on the seven components with diagnosis as a factor in the ANOVA model, and the first six components were significant at  $p < .01$  or less. Planned orthogonal contrasts revealed the following intergroup differences (Table 7): For Spontaneity, WS and controls were similar.

The individuals in the AUT and DS groups showed significantly lower scores on this factor than controls ( $p < .02$ ). For Listening Habits, individuals with WS were higher than CTL ( $p < .001$ ) and AUT ( $p < .005$ ), and CTL were higher than DS ( $p < .001$ ) who in turn were higher than AUT ( $p < .03$ ). For Reproduction, WS differed significantly from AUT ( $p > .02$ ) and AUT differed from DS ( $p < .04$ ); WS and CTL and WS and DS were statistically similar on this factor.

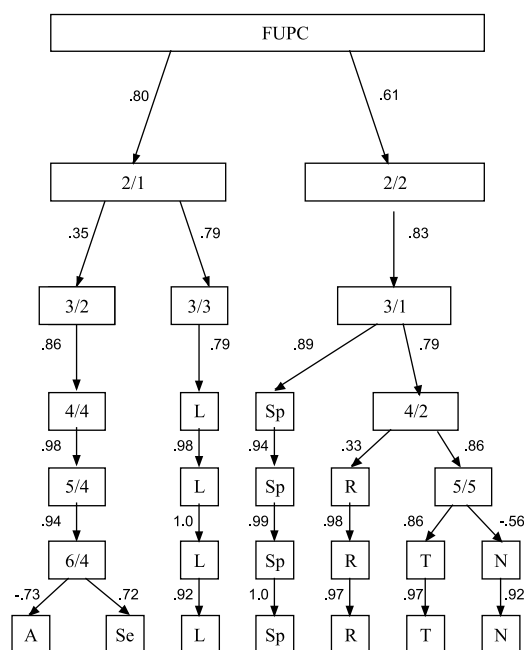


Fig. 2. Hierarchical structure of the seven principal components extracted through factor analysis and their intercorrelations. Letters refer to the factor names as designated in Table 7. Note that Factors I (Spontaneity) and III (Listening Habits) break away from the rest relatively early and remain virtually unchanged until the end of the analysis.

For Theory/Achievement, the WS and CTLs were again similar, with WS showing higher scores than AUT ( $p < .005$ ) and DS ( $p < .001$ ), and significant differences between AUT and CTL ( $p < .001$ ) and DS and CTL ( $p < .001$ ). The Age of Onset at which various musical behaviors was first noticed showed differences between WS and CTL ( $p < .03$ ) and WS and AUT ( $p > .03$ ). Finally, the Sensitivity factor showed differences between the CTLs and all other groups ( $p < .001$ ). *Sensitivity* is an especially interesting factor – it comprises items that concern how long the individual remains happy after hearing happy music, or sad after hearing sad music. It was designed as an index of the affective response to music in terms of how long music's effects can last. The WS individuals scored higher on this factor than any other group.

Table 7. ANOVA on the Seven Factors. Values in the Cells are the Means (and Standard Deviations) for Each Factor. Superscript Letters Indicate Significant Differences Between a Cell and Other Groups (C = Differences to Controls, A = Differences to Autism, D = Differences to Down Syndrome, W = Differences to Williams Syndrome). Significance Level is  $p < .05$  for all Pairs, Adjusted for Multiple Comparisons.

| Factor                 | CTL                       | AUT                      | DS                       | WS                      |
|------------------------|---------------------------|--------------------------|--------------------------|-------------------------|
| I. Spontaneity         | 4.2 <sup>a,d</sup> (.8)   | 3.6 <sup>c,w</sup> (.9)  | 3.7 <sup>c</sup> (.7)    | 4.0 <sup>a</sup> (1.2)  |
| II. Reproduction       | 4.1 (.8)                  | 4.4 <sup>w</sup> (.7)    | 3.8 <sup>a</sup> (.8)    | 3.8 <sup>a</sup> (1.2)  |
| III. Listening habits  | 3.6 <sup>d,w</sup> (.8)   | 3.5 <sup>d,w</sup> (1.2) | 4.3 <sup>a,c</sup> (1.0) | 4.4 <sup>a,c</sup> (.9) |
| IV. Theory/Achievement | 4.3 <sup>a,d</sup> (1.1)  | 3.4 <sup>c,w</sup> (.9)  | 3.6 <sup>c,w</sup> (.6)  | 4.0 <sup>a,d</sup> (.9) |
| V. Age of onset        | 4.1 <sup>w</sup> (.9)     | 4.3 <sup>w</sup> (.9)    | 4.0 (.8)                 | 3.8 <sup>a,c</sup> (.1) |
| VI. Negative reactions | 4.0 (.8)                  | 4.4 (.9)                 | 3.8 (.6)                 | 3.9 (1.3)               |
| VII. Sensitivity       | 3.4 <sup>a,d,w</sup> (.7) | 4.2 <sup>c</sup> (1.0)   | 4.1 <sup>c</sup> (.9)    | 4.5 <sup>c</sup> (.9)   |

### Discriminant Function Analysis

Because the seven derived factors showed some intergroup differences, we performed a Discriminant Function Analysis to see how well group membership (or diagnosis) could be predicted from those seven factors. The regression scores for the first six factors were all significant (Wilks' Lambda for Factors 1–6, respectively, was .96, .84, .97, .88, .96, and .78;  $p < .01$  for all). Seventy percent of the cases were correctly classified using the Discriminant Functions derived in this analysis. Figure 3 shows the location of each individual subject within the dimensional space provided by the first two discriminant functions, with each of the four types of subjects differ-

entiated by different symbols. Note that the group centroids are easily distinguishable, and that within this two-dimensional representation, the CTL group is easily distinguished, spatially, from the three developmentally impaired groups.

### Controlling for Age and Sex

Because age of onset of various musical behaviors was found above to be a significant variable, we sought to explore and clarify the relation between age and the seven principal components extracted – in particular, in order to address concerns that age might be a significant confound in interpreting the principal components. We performed a stepwise linear regression of age against the seven factors, and found that age resulted in a poor fit to the model, accounting for only 10% of the variance (n.s.). We tried again to control for the possible confounding effects of age by adding age as a variable to the Discriminant Function Analysis to predict group membership alongside Dx, and age was again found to be insignificant (Wilks' Lambda = .98,  $F(3, 302) = 2.1$ ,  $p \sim .10$ ).

Similarly, because our sample included four times as many Autistic males as females, and three times as many females as males in the control group, we controlled for this variable by performing a binary logistic regression with sex against the seven principal components, and again found no significance,  $\chi^2(7) = 12.4$ ,  $p \sim .09$ .

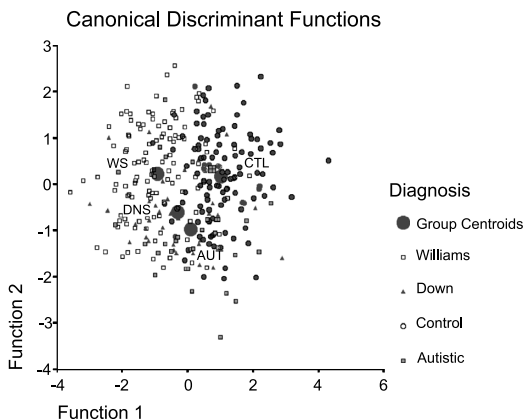


Fig. 3. Discriminant function plot showing differentiability of the subject populations by the seven factors derived from the questionnaire.

### Trends

Several trends that did not reach statistical significance are worth noting. The WS individuals

seem to start playing musical instruments at an earlier age than the CTL group do,  $F(3, 131) = 2.463$ ,  $p < .06$ . Parental reports we tallied from the questionnaires noted that, as infants, many more of the WS subjects made “instruments” out of household items (such as a cooking pot and wooden spoon) compared to the non-WS children in the same home.

While CTL subjects are reported to begin playing a musical instrument at the same time they begin musical training (mean age of 8.12 years and 8.87 years, respectively), the WS subjects begin playing a musical instrument on average 5 years *before* they begin musical training (mean age of 6.49 years and 11.38 years, respectively). Finally, WS were more likely to reproduce music (i.e., sing or play familiar music) than DS individuals ( $p < .06$ ).

## DISCUSSION

We found a number of differences between the individuals with WS and the comparison groups. To begin with, WS were reported to be as accomplished at reproducing music as chronologically age-matched controls, showed similar levels of achievement as indexed by the Grison profile, and spontaneously produced rhythmic sequences as often as controls. They were just as likely to play a musical instrument and to have taken music theory. When the questionnaire items are compiled into higher order factors, WS individuals are indistinguishable from controls on Factors I, III, IV, and VII: the dimensions of Spontaneity, Reproduction Profile, Theory/Achievement, and Negative Reactions to Music. There exist few dimensions on which people with WS test comparable in ability to age-matched controls (Bellugi, Lichtenberger, Jones, et al., 2001) but musical involvement certainly appears to be one of them.

For more than a century, it has been stated that people with Down Syndrome (Shuttleworth, 1900; Stratford & Ching, 1989; Tredgold, 1908) and with Autism (Applebaum, Egel, Koegel, & Imhoff, 1979; Edgerton, 1994; Sherwin, 1953) show high levels of interest and particular ability in music but up to this point no one has compared individuals with DS or AUT and WS directly for

music-oriented behaviors. It is important to establish what effect developmental delay might have on an individual's interest in music in order to completely understand what determines the salience of musical activities for humans. Although it might be tempting to suggest that developmentally delayed populations in general are drawn to music, this is too simplistic an approach. On every one of the 10 items that showed significant inter-group differences by an initial ANOVA, the WS individuals were reported to be more musical than the DS and AUT individuals, and this is highly significant statistically. Specifically, WS spent more time listening to music per week than DS or AUT, demonstrated more interest in (and spent more time on) musically-related activities, expressed more emotions as a response to music listening, were more accurate in their reproductions of songs, played original music and rhythms more frequently, were more likely to play a musical instrument, spent more hours per week playing an instrument, showed higher achievement on the Grison Profile, and were more likely to have been exposed to music theory. A discriminant function analysis confirmed that the four groups are clearly differentiable on the basis of their responses to this questionnaire, with 70% correct classifications.

As Udwin (1990) noted, several caveats with his questionnaire study of WS that apply in the present study as well. The data presented here are subject to the limitations of parental recall and subjectivity, and they are based on a somewhat self-selected sample of WS families, all of whom are members of either the Williams Syndrome Foundation or the Williams Syndrome Association. At present, these two organizations are the only registers of affected individuals in North America, and hence the only way we had to contact families with WS.

One important limitation in the present study is that our method may have underestimated the amount and strength of musical reactions by people with Autism and Down Syndrome. People with WS tend to be more verbally and emotionally expressive, more loquacious, than the comparison groups and this could have biased our findings – it may be that the parents of children with WS are more aware of their musical

behaviors precisely because their children are more likely to communicate them through facial expression and language (two areas of preserved skill) than are children with Autism or Down Syndrome. Indeed, the reports we received from parents of people with WS were far more rich in detail than the reports we received from any other group.

It is difficult to say if the children in the DS and AUT groups had different emotional experiences in relation to music or if rather, they had different ways of expressing or showing their emotional reactions to music. Children with DS tend to use less internal state language in general, compared to typically developing children (Beeghly & Cicchetti, 1997), although when they do refer to internal states it is far more often a referenced to emotional, rather than cognitive or volitional, states. Thus, emotional language should be more salient to the parents of DS children even though its occurrence might be less frequent. We note also that the parents who completed the surveys were all typically developed adults themselves, and so they would presumably be most sensitive to the signs of emotion that are common in typically developing people. However, the adults' familiarity with their atypically developing children presumably made those parents more sensitive to the emotional states of their children, even if those states were idiosyncratically displayed.

We chose individuals with Autism and Down Syndrome as comparison groups because of several salient similarities to individuals with Williams Syndrome. All three syndromes are of presumed genetic origin, and are characterized by neurological impairment, cognitive impairment, and cerebral dysmorphologies. The mean IQ for members of these groups is somewhat similar. However, each syndrome is characterized by distinct differences in cognitive function, overall phenotype, and developmental trajectory (e.g., Kasari, Freeman, & Hughes, 2001). Autism, for example, experience greater anxiety in early childhood, and this may delay the development of musical skills. Research with autistic individuals has found deficits in the ability to recognize other people's emotions and mental states (Baron-Cohen, Wheelwright, Hill, & Plumb, 2001; Bormann-Kischkel, Vilsmeier, & Baude, 1995;

Moore, Hobson, & Lee, 1997), a necessary precursor to understanding music (Meyer, 1956). It is thought that these impairments arise from fundamental problems with Theory of Mind, which involves the meta-representation of mental states in other people (Baron-Cohen, Leslie, & Frith, 1985; Tager-Flusberg & Sullivan, 2000).

In spite of their difficulties with emotion perception and meta-representation, people with autism exhibit relatively intact musical processing capabilities including the musical emotion (Heaton, Happé, Williams, & Cummins, 2003; Heaton, Hermelin, & Pring, 1999), tonal structure (Frissell, 2001) and melodic contour (Heaton, Pring, & Hermelin, 1999; Mottron, Peretz, & Ménard, 2000), and individuals with autism actually perform above average on tasks that involve memory for absolute pitch information (Heaton, 2003; Heaton, Hermelin, & Pring, 1998).

What is characteristic or unusual about WS? Anecdotal reports have cited a passion for music, increased emotional responsiveness to music, and greater time spent in musical activities. These anecdotal claims were borne out by the results of the present study. Based on both anecdotal evidence and findings from the Music Questionnaire, it seems that WS exhibit a higher degree of emotionality than the AUT, DS or CTL groups when listening to music (Levitin & Kreiter, 2002).

Individuals with WS (Bellugi, Adolphs, Cassady, & Chiles, 1999; Doyle et al., 2004; Jones et al., 2001; Losh et al., 2001) and DS (Kasari & Hodapp, 1996) have a stronger than typical inner drive to socialize, whereas individuals with AUT have a less than usual drive toward social stimuli. In previous studies (Levitin & Bellugi, 1999), testing of WS and DS subjects was made more difficult by their preference for interacting with the experimenters as opposed to actually completing the tasks before them.

In the current study, we note that our WS respondents were significantly more involved in music than our AUT respondents, and on six out of eight measures (ref. Table 2), the DS respondents were as well – this latter difference, although not significant ( $p \sim .14$ ) is suggestive. Is music a vehicle for social connection? Huron (2001) suggests that there may exist a common genetic underpinning to musical and social

behaviors, and that the two co-evolved (see also Hagen & Bryant, 2003). Our own observations of hundreds of individuals with WS in naturalistic, informal settings confirm that music serves a social function – they use it to bond, to open up conversations, and as a way to get to know one another; music is a frequent topic of spontaneous conversation when two people with WS first meet. The co-occurrence and relative sparing in WS of social and musical drive suggests the possibility that the gene deletion in WS is somehow contributing to both of these markers of the WS phenotype. Because WS have a gene deletion (unlike DS, e.g., who have extra genetic material), the mechanism by which social and musical behaviors are facilitated must be through the removal of an inhibition of some sort. But music is serving more than the role of a social bonding agent among WS. WS individuals make music alone, listen alone, and engage in musical behaviors without an immediate social purpose.

One might argue that the intense engagement one sees with social interactions and with music in WS is the result of a general lack of inhibition. If this were true, we would expect to see emotional behavior during many other activities, but this is not documented. The domains of language, sociability and music seem to be domains of remarkable strength and of intense interest to individuals with WS, to the exclusion of other non-social domains.

WS individuals were rated significantly higher than CTL individuals on questions such as “How much interest does your child show in music-related activities?” and “How many hours a week does your child spend on music related activities?” The reported age that interest was first observed in WS is over 2 years younger than the CTL group. Based on these findings, WS show more and earlier interest in music than Normal Control groups.

Although we have provided support for differences between groups on measures of interest and emotionality, the specific patterns of musical abilities across these four groups will reveal the most about their music *processing* abilities. It is clear that the WS individuals are performing much more similarly to the CTL group than to

the DS and AUT groups on measures such as accuracy of musical reproductions, and this is supported by experimental work as well (Levitin & Bellugi, 1999). In those studies, individuals with WS, DS, AUT and typically developing controls completed a battery of musical tests, including rhythmic and melodic completion tasks. The WS reproductions tended to be faithful to the originals, as were those of the control group. A high proportion of the DS individuals were unable to complete the tests at all, and the individuals with AUT performed at a level significantly below the others.

Our findings are relevant to several important questions. These include the WS musical profile and its position in understanding the broader WS phenotype; the differences between the musical behaviors of people with WS and people with DS or AUT (and other neurogenetic developmental impairments); and the possible neural and genetic bases of these behaviors. As Hopyan et al. (2001) noted, WS individuals show “a strong engagement with music as a means of expression, play, and, perhaps, improvisation.” Our data provide even stronger support to this claim.

## Neurobiological Considerations in WS

### *Cytoarchitectonics*

Cytoarchitectonic studies of individuals with WS have found that auditory cell packing density and neuronal size (in area 41) were abnormal (Galaburda & Bellugi, 2000; Holinger, McMennamin, Sherman, & Galaburda, 2001). Individuals with WS had an excess of mid and large cells in layers II in both cerebral hemispheres, and in layer VI in the left hemisphere. There was a hemisphere-by-diagnosis interaction between WS and control brains in cell-packing density in layer IV, and in neuronal size in layer III. Larger than normal pyramidal neurons were found bilaterally in layer II, in left layer III and VI and were interpreted as being consistent with a hypothesis of increased connectivity in WS auditory cortex. This hyperconnectivity may be related to the relative sparing of language, music, and other auditory function, and could account for some of the unusual reactions to auditory stimuli documented in the present study.

### *Brain Morphometry*

Structural MRI (Reiss, Eliez, Schmitt, Straus, Lai, Jones, & Bellugi, 2000) revealed an overall reduction in cerebral volume in WS, with the relative size of the temporal lobe and amygdala preserved bilaterally and posterior cerebral volumes selectively decreased. Gray matter density was found to be reduced in some regions (caudate nucleus and intraparietal sulcus) and increased in others (insular cortex, cingulate gyrus, and cerebellum). Double dissociations were also observed between brain morphometry in WS and AUT, particularly in regions of the cerebellum that are believed to mediate emotion.

### *Electrophysiology*

People with WS showed morphology, distribution, sequence, and latency of evoked response potential (ERP) components similar to typically developing normal controls (Bellugi, Bihle, Doherty, Neville, & Damasio, 1989; Bellugi, Bihle, Neville, Jernigan, & Doherty, 1992; Hickok et al., 1995). However, in tests of the auditory recovery cycle, people with WS showed a marked increase in the amplitude of the N100 and P200 responses at faster repetition rates, suggesting that the refractory period for neurons responding to sound is shorter in WS, and indicating cortical-level hyperexcitability.

### *Neuroanatomy*

A functional neuroimaging study of individuals with WS and normal age-matched controls (Levitin et al., 2003) found that the overall pattern of activation in the whole brain was markedly different between the two groups as they listened to music, noise, or silence. The WS participants showed substantially decreased activation in temporal lobe regions normally associated with auditory processing (perhaps a consequence of hyperconnectivity), accompanied by more variable and diffuse patterns of activation throughout the cortex and neocortex, and higher activation levels than controls in the paleocortical amygdaloid complex. Compared to the controls, the WS group showed significantly higher activation levels in the right amygdala, cerebellum (particularly the vermis), pons and brain stem, which may be related to affective processing. These activations

are presumably mediated by major connections linking the prefrontal cortex with the basal ganglia and the cerebellar vermis, and are consistent with the notion that a region in the right cerebellum may be functionally related to those in the left inferior frontal cortex for semantic processing (Levitin & Menon, 2003), thus serving to link the cognitive and emotional aspects of sound. The role of the cerebellum in WS thus deserves further study.

Taken together, cytoarchitectonic, structural MRI, and ERP findings suggest the possibility that the brains of WS individuals are organized differently than normals, at both a micro- and a macro-level. These findings thus provide neuroscientific confirmation of self- and parental-reports that sounds hold special emotional meaning to people with WS. The pattern and levels of activations observed point to the anatomical underpinnings of the emotional responses to sound, and in particular, to the recruitment of emotional centers of the brain in response to auditory stimuli among individuals with WS.

### *Handedness*

A number of studies put the incidence of right handedness at about 90% and left handedness at 8–10% in the general population (Kagan, 1997). We found intergroup differences in handedness of our participants with parents reporting that DS and WS individuals were more likely to be left-handed or ambidextrous than controls. Higher rates of left-handedness have been reported in musicians (Aggleton, Kentridge, & Good, 1994; Hassler & Gupta, 1993). The neurological basis for this is presumed to follow from the right hemisphere dominance of left-handers who would putatively possess superior spatial/motor skills (Porac & Coren, 1981). A possible confound in our study therefore is that because our DS and WS samples contained a larger than normal proportion of left-handers, by extension the sample contained a larger than normal proportion of musicians. But higher incidence of left-handedness has been shown to occur in individuals with AUT, DS and WS (Batheja & McManus, 1985; Previc, 1996), and we believe our samples simply reflected the natural demographics of these groups. The reasons why individuals with AUT, DS, and WS might show higher incidence of

left-handedness is not clear and requires further investigation.

### Phenomenology

A flavor of what it is like to have or live with WS emerged from the questionnaires. Many WS individuals were reported to sit for hours enchanted by certain sounds, or to learn to name cars and vacuum cleaners by their make and model numbers, based solely on the acoustic information. There were no similar reports from the comparison groups. The reports of Williams parents were clearly more vivid, and described higher levels of emotional engagement with sound. A potential confound exists here, in that WS individuals are known to be particularly verbal (especially compared to DS and AUT individuals) and so it may be precisely because WS individuals are so verbal that their parents became aware of their experiences. But such an explanation does not account for why WS parents reported so much more engagement than did CTL parents, nor does it account for the many anecdotes that described the parents' direct observation of their WS child, rather than the child's report to the parent about their enthusiastic relationships with music.

Three examples taken from the "free response" portion of the questionnaires will illustrate this.

The parent of a 12 year-old female with WMS reported, "When she listens (to music) she is completely absorbed and can't do anything else."

The parent of a 28 year-old male with WMS reported that he "gets so into music that he loses touch with reality."

A teacher reported that when two teenagers with WS first meet, they immediately begin to talk about music, they "get a certain look in their eyes" and spontaneously start to sing with each other, becoming completely "taken over" by the experience.

### CONCLUSIONS

We administered a questionnaire to individuals with Williams Syndrome, Down Syndrome, Autism, and typically developing normal controls

in an effort to characterize their musical background, behaviors and penchants. We found significant intergroup differences with the individuals with WS rated higher in musical accomplishment, engagement, and interest than either of the comparison groups (with DS and AUT), and equivalent on most measures to the control group of typically developing individuals. Moreover, 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 than members of the other three groups. In addition, the effects of music listening tended to last longer in the WS group. A factor analysis extracted seven principal components that characterize the musical phenotype in this sample: Spontaneity, Reproduction, Listening Habits, Exposure to Music Theory, Age of Onset of Musical Behaviors, Negative Reactions to Music and Sensitivity to Music. A discriminant function analysis of those components successfully predicted group membership for 70% of the cases. We have thus verified empirically the anecdotal accounts of differences in musical phenotype between individuals with WS, DS, AUT and normal controls.

Phenomenologically, people with WS seem to truly experience music more fully than most people, and to be consumed by their affective reactions to music. As the parent of a WS child reported, her daughter began weeping after a couple of notes were played at a Mozart concert. The girl's reaction was so strong that she left the concert and after returning, once again burst into tears. After hearing a more uplifting Mozart song some months later, she explained to her mother "there are two kinds of Mozart: the kind that hurts and the kind that does not hurt."

### ACKNOWLEDGEMENTS

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## APPENDIX A

**The Salk/McGill Music Inventory (SAMMI)**  
**Questionnaire of Music Ability and Interest**

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**GENERAL INFORMATION**

Name \_\_\_\_\_ Today's Date: \_\_\_\_\_

1. Sex (M/F) \_\_\_\_\_ 2. Date of Birth \_\_\_\_\_ Age Today \_\_\_\_\_

Name of Parent/Guardian \_\_\_\_\_

Relationship to Subject \_\_\_\_\_

Street Address \_\_\_\_\_

City, State, Zip \_\_\_\_\_ Phone ( ) \_\_\_\_\_

3. Does your child have Williams syndrome, Down syndrome, Asperger, Autism, or another neurodevelopmental disorder?

Yes No If yes, Specify: Williams Down Asperger Autism 11Q

Other(\_\_\_\_\_)

4. Is your child right or left handed? Right Left Ambidextrous

5. Has your child ever had problems with hearing loss? Yes No

6. If so, what were those problems? \_\_\_\_\_

7. Does your child have physical deficits that may affect his/her participation in music activities?

Yes No

8. If so, what are those deficits? \_\_\_\_\_

9. Has your child ever had symptoms of unusual fright or sensitivity to certain sounds? Yes No

*If yes, please answer questions 10–13, otherwise, please skip to question 14:*

10. At what age were these symptoms first observed? Age \_\_\_\_\_

11. Does your child now have the same symptoms of sound sensitivity that were first observed?

Yes No

12. If not, describe how your child's symptoms of sound sensitivity have changed with age:

\_\_\_\_\_  
 \_\_\_\_\_  
 \_\_\_\_\_

13. Give 4 (or more) examples of situations or sounds that have caused a sensitive response in your child:

\_\_\_\_\_  
 \_\_\_\_\_  
 \_\_\_\_\_

**Musical Ability and Interest**

14. Please check the highest category that describes your child:

( ) Is an excellent musician in both performing ability and theoretical knowledge.

( ) Plays an instrument, and has fair sight reading abilities as well as a general musical knowledge.

( ) Plays an instrument, but has not been trained in theory and cannot sight read (reproduce written music on an instrument).

- ( ) Enjoys singing and has a set of songs which are commonly sung or is a critical listener of songs on the radio or buys CD's, tapes, or records and listens to them often.
- ( ) Occasionally sings familiar melodies, such as nursery rhymes and popular songs or sometimes listens to music on the radio.
- ( ) Shows no interest; does not care to listen to or sing music.

Please answer the following questions to the best of your ability. Observations should include behavior at school as well as at home. When a rating is requested, please circle **one** number only. Most questions are directed towards your child's **current** musical ability and interest. Please feel free to write in the space provided when elaboration is needed.

15. How much interest does your child show in music-related activities?

|                      |                  |   |   |   |                  |   |
|----------------------|------------------|---|---|---|------------------|---|
| 1                    | 2                | 3 | 4 | 5 | 6                | 7 |
| very little interest | average interest |   |   |   | extreme interest |   |

16. At what age did your child first show interest in music? Age \_\_\_\_\_

17. Please describe all of the music-related activities that your child engages in:

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18. How many hours a week does your child spend on music-related activities?

|       |       |     |     |     |      |       |       |       |
|-------|-------|-----|-----|-----|------|-------|-------|-------|
| 0     | 1-2   | 3-4 | 5-6 | 7-8 | 9-10 | 11-12 | 13-14 | 15-16 |
| 17-18 | 19-20 | 20+ |     |     |      |       |       |       |

### Responsivity to Music

19. When your child actively listens to music (i.e., pays attention to it), how would you compare his or her level of expressed emotion with a typical child of the same age?

|                   |   |                 |   |   |                   |   |
|-------------------|---|-----------------|---|---|-------------------|---|
| 1                 | 2 | 3               | 4 | 5 | 6                 | 7 |
| much less emotion |   | similar emotion |   |   | much more emotion |   |

20. When your child listens to "upbeat" music how would you describe his or her mood? (Upbeat music makes most people feel positive, such as "Good Vibrations" by the Beach Boys or "Old McDonald Had a Farm")

**If your child reacts positively, use this scale:**

|                   |   |                     |   |   |                    |   |
|-------------------|---|---------------------|---|---|--------------------|---|
| 1                 | 2 | 3                   | 4 | 5 | 6                  | 7 |
| a little positive |   | moderately positive |   |   | extremely positive |   |

**If your child reacts negatively, use this scale:**

|                   |   |                     |   |   |                    |   |
|-------------------|---|---------------------|---|---|--------------------|---|
| 1                 | 2 | 3                   | 4 | 5 | 6                  | 7 |
| a little negative |   | moderately negative |   |   | extremely negative |   |

21. How long does your child's reaction to upbeat music last?

**Please check one of the following:**

|                     |                                    |
|---------------------|------------------------------------|
| A couple of minutes | About 1 hour                       |
| About 15 minutes    | A couple of hours                  |
| About 30 minutes    | Other ____ (please fill in a time) |

22. When your child listens to "sad" music how would you describe his or her mood? (Sad music makes most people feel sad, such as "Sounds of Silence" by Simon and Garfunkel or "Where have all the flowers gone?" by Bob Dylan)

**If your child reacts positively, use this scale:**

|                   |   |                     |   |   |                    |   |
|-------------------|---|---------------------|---|---|--------------------|---|
| 1                 | 2 | 3                   | 4 | 5 | 6                  | 7 |
| a little positive |   | moderately positive |   |   | extremely positive |   |

**If your child reacts negatively, use this scale:**

- |                   |   |   |                     |   |   |                    |
|-------------------|---|---|---------------------|---|---|--------------------|
| 1                 | 2 | 3 | 4                   | 5 | 6 | 7                  |
| a little negative |   |   | moderately negative |   |   | extremely negative |
23. How long does your child's reaction to sad music last?

**Please check one of the following:**

- |                     |                                    |
|---------------------|------------------------------------|
| A couple of minutes | About 1 hour                       |
| About 15 minutes    | A couple of hours                  |
| About 30 minutes    | Other ____ (please fill in a time) |

24. If your child demonstrates a reaction to music that was not addressed above please describe it here:

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### **Musical Memory, Reproduction and Creativity**

25. Does your child have any ability to reproduce music or songs with his/her voice **or** an instrument? (Include humming or whistling, but **not** rhythmic reproduction, of musical tones/notes) **Yes No**  
*If you answered YES to Question 25, please answer Questions 26–33 below, otherwise, skip to Question 34:*

26. At what age did your child reproduce music (any attempts to mimic music, with voice or musical toys, etc.)?

Age \_\_\_\_\_

27. Describe the type of music that was first reproduced and under what circumstances.

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28. Currently, when your child reproduces music how accurate is it?

|             |   |   |                   |   |   |         |
|-------------|---|---|-------------------|---|---|---------|
| 1           | 2 | 3 | 4                 | 5 | 6 | 7       |
| no accuracy |   |   | somewhat accurate |   |   | perfect |

29. How much of a particular song is your child able to reproduce?

**Please mark one of the following:**

- Reproduces only the beginning
- Reproduces a small portion
- Reproduces many small portions spread across the entire piece
- Reproduces most of the piece
- Reproduces the entire piece

30. How frequently does your child reproduce music in comparison to a typical child of his or her age?

|                      |   |   |                   |   |   |                      |
|----------------------|---|---|-------------------|---|---|----------------------|
| 1                    | 2 | 3 | 4                 | 5 | 6 | 7                    |
| much less frequently |   |   | similar frequency |   |   | much more frequently |

31. How many songs or pieces of music has your child ever reproduced?

**Make your best estimate:**

- |           |                                    |
|-----------|------------------------------------|
| 1–2 songs | 50 songs                           |
| 10 songs  | 75 songs                           |
| 25 songs  | Other ____ (please fill in amount) |

32. How would you rate the complexity of songs or pieces of music that your child reproduces?

|                        |   |   |                       |   |   |                       |
|------------------------|---|---|-----------------------|---|---|-----------------------|
| 1                      | 2 | 3 | 4                     | 5 | 6 | 7                     |
| complex as a TV jingle |   |   | complex as a pop song |   |   | complex as a symphony |

33. Describe any aspects of your child's ability to reproduce music that were not addressed.

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34. How often does your child play rhythmic patterns that have a steady, repeated beat (includes finger tapping, drumming with household objects)? Do **not** include rhythmic tapping due to nervous reactions (i.e., fear of new situations, performance anxiety about tests, social situations, etc.)

|                     |   |                         |   |   |                          |   |
|---------------------|---|-------------------------|---|---|--------------------------|---|
| 1                   | 2 | 3                       | 4 | 5 | 6                        | 7 |
| never plays rhythms |   | sometimes plays rhythms |   |   | plays rhythms very often |   |

35. How would you describe the complexity of rhythmic patterns that your child creates in non-anxiety provoking situations?

|                  |   |                    |   |   |                   |   |
|------------------|---|--------------------|---|---|-------------------|---|
| 1                | 2 | 3                  | 4 | 5 | 6                 | 7 |
| extremely simple |   | average complexity |   |   | extremely complex |   |

36. How often does your child create music or songs of their own?

|               |   |                   |   |   |                         |   |
|---------------|---|-------------------|---|---|-------------------------|---|
| 1             | 2 | 3                 | 4 | 5 | 6                       | 7 |
| never creates |   | sometimes creates |   |   | creates extremely often |   |

37. How would you describe the complexity of music or songs that your child creates?

|                        |   |                       |   |   |                       |   |
|------------------------|---|-----------------------|---|---|-----------------------|---|
| 1                      | 2 | 3                     | 4 | 5 | 6                     | 7 |
| complex as a TV jingle |   | complex as a pop song |   |   | complex as a symphony |   |

38. Which of the following describes the music that your child creates?

**Please mark all that apply:**

- ☐ It has a repeating, definable beat.
- ☐ It sounds “pleasing” (like music and not random tones or sounds).
- ☐ It has more than one section (not just one repeated part).
- ☐ It uses a wide range of tones (both low and high tones).
- ☐ It seems based on other pieces of music that your child has heard.

39. Please describe any aspects of your child’s ability to create music that were not addressed above:

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### **Musical Training**

40. Has your child ever played an instrument (including informal play at home)? Yes No

41. List any instruments your child has played and at what ages? (Include all instances of playing: during professional instruction, at school, or uninstructed playing at home)

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42. How many hours a week does your child currently spend playing instruments?

|   |    |     |   |   |   |   |   |   |
|---|----|-----|---|---|---|---|---|---|
| 0 | 1  | 2   | 3 | 4 | 5 | 6 | 7 | 8 |
| 9 | 10 | 10+ |   |   |   |   |   |   |

43. Has your child had formal training in music theory (includes instruction to read, analyze, and compose music)? **Yes No**

44. Please list and describe all instances of training in music theory and at what ages they occurred. If possible include exactly what was studied (if your child was trained on an instrument, list the topics which were covered such as reading music, fingering, chord analysis, etc.):

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45. Does your child have perfect pitch (ability to correctly identify a note when it is played, or correctly sing a specific note when requested without being given a note from which to cue)? **Yes** **No**

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46. **Please add any additional comments or examples about overall or special music abilities that were not already addressed in this questionnaire (continue on back if necessary):**

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