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Auditory pattern perception in children with Williams syndrome.

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Auditory Pattern Perception in Children with Williams Syndrome

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ABSTRACT

Children with Williams syndrome (WS) have a unique neuropsychological profile characterized by extremely poor visuospatial skills but relatively preserved verbal skills. This pattern, in addition to anecdotal reports of relatively preserved music skills in these children, suggested that children with WS may be relatively good at processing auditory patterns in general. In the present study, language and music skills of 19 children with WS (8-13 years) were examined and compared to 19 normal children (5-12 years) equivalent for mental age (M=8 years, 1 month) based on the Peabody Picture Vocabulary Test-Revised (PPVT-R). Measures included the following: WISC-III (WS group only), PPVT-R, Auditory Closure Test, Controlled Oral Word Association Test, Digit Span, Sentence Memory Test, parent and child questionnaires and interviews, and the Tonal and Rhythm subtests of Gordon's Primary Measures of Music Audiation (1986), which requires discrimination between pairs of melodic or rhythmic fragments. As expected, results confirmed that the previously observed pattern of better verbal than visuospatial performance in children with WS was also evident in the present sample. For the WS group, the pattern of performance on linguistic tests appeared to be based on the complexity of processing required, indicating that basic auditory perception is more intact in these children than rote learning or auditory

memory. The present study provided the first empirical evidence of relatively intact musical abilities among children with WS, commensurate with their relatively strong receptive vocabulary (a relatively simple language skill). In addition, significant but moderate correlations between language and music skills were found for both groups of children, which implies that language and music skills are subserved by a common mechanism used to process auditory patterns in general. The results of the present study also raise the possibility that auditory processing among children with WS might be atypical when compared to normal children. For example, children with WS exhibit a rather intense interest in music that is accompanied by strong affective responses. Moreover, this peculiar affinity for music may be related to the hyperacusis that is observed among these children. 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. Continued research to explore and expand upon the present findings would be theoretically and clinically beneficial.

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AUDITORY PATTERN PERCEPTION IN CHILDREN WITH WILLIAMS SYNDROME

Children with Williams syndrome (WS) have "elfin" facial features and relatively small stature (Lowe, Henderson, Park, & McGreal, 1954). Although these children generally are mentally retarded, they have an unusual cognitive profile, exhibiting relatively preserved verbal skills in contrast to extremely poor nonverbal skills. For example, a 15-year-old girl with WS who required a babysitter for supervision and had a Full Scale IQ of 49 described herself in the following manner: "You're looking at a professional book writer. My books will be filled with drama, action, and excitement" (Bellugi, Bihrle, Neville, Jernigan, & Doherty, 1992, p. 205). Moreover, these children are also overly sensitive to noise; they cringe and cover their ears or cry out at noises that do not cause discomfort in normal or other mentally retarded children (e.g., Arnold, Yule, and Martin, 1985; Bellugi, Wang, & Jernigan, 1994; Klein, Armstrong, Greer, Brown, 1990; Udwin, Yule, & Martin, 1987). Thus, their characteristic physical features, hyperacusis (unusual sensitivity to noise), and unusual cognitive profile distinguish children with WS from normal children and other mentally retarded children.

There are suggestions from the literature and from parental reports that "musicality" may also be a characteristic of these children (Anonymous, 1985; Lenhoff, 1996; Levine, 1992; Udwin et al., 1987; von Arnim & Engel, 1964). Hence, the ability of children with WS to process auditory patterns may be relatively preserved in general. Indeed, in an early report delineating the psychological characteristics of the syndrome, each child was noted to be musical in addition to having relatively well developed language skills (von Arnim & Engel, 1964). In a case study of a child with WS, music was reported to be the child's "truest love" (Anonymous, 1985, p. 968). Recently, Levine (1992) reported that music was an area of strength in these children. Unfortunately, the musical abilities of children with WS have not been investigated in a systematic manner.

On one hand, the notable verbal skills and musicality of children with WS suggest that their auditory pattern processing skills are relatively intact. On the other hand, their hyperacusis suggests atypical auditory processing. Hyperacusis is characterized by the experience of discomfort or pain at levels of sound that do not cause discomfort in normal individuals. The cause of hyperacusis in individuals with normal hearing (e.g., children with WS) is unknown. The presence of relatively intact verbal skills and apparent musicality in combination with hyperacusis raises a wide range of questions about children with WS. Does their hyperacusis reflect unusually good hearing or a low tolerance of particular sounds? Are these children particularly musical? If so, in what way? Finally, are language and music skills in individuals with WS linked, perhaps by common auditory pattern processing abilities?

The purpose of the present study was to examine the auditory pattern processing abilities of children with WS in order to answer some of these questions. The results of this study could potentially have important implications for modularity hypotheses (Fodor, 1983; Jackendoff, 1987) -- which postulate discrete, impenetrable processing modules for language and music -- as well as for Rourke's (1989) developmental model of the syndrome of Nonverbal Learning Disabilities (NLD) -- which postulates intact processing of aspects common to both music and language, such as auditory perception, attention, and memory.

Williams syndrome

History of WS

A syndrome is diagnosed based on the presence of behavioral, cognitive, or medical symptoms that represent a subset of a prototypical constellation of symptoms. Originally, the prototypical constellation of symptoms for WS was defined independently by medical specialists from two specialty areas: endocrinology and cardiology. Common medical problems associated with WS are the severe (Franconi) form of idiopathic infantile hypercalcemia (i.e., a disorder of calcium metabolism in infancy) and supravalvular aortic stenosis (i.e., a narrowing of the aorta above the aortic valve). Whereas endocrinologists diagnosed and treated the hypercalcemia, cardiologists diagnosed and treated the supravalvular aortic stenosis. Because of the relative independence of these two areas of medicine, recognition that the two syndromes were actually one and the same was not widespread until recently.

Hypercalcemia. WS was first described in association with the severe (Franconi) type of idiopathic infantile hypercalcemia (Franconi, Girardet, Schlesinger, Butler, & Black, 1952; Lightwood, 1952; Payne, 1952). Infantile hypercalcemia is characterized by an excess of calcium in the blood. In both severe and mild forms of the disorder, the affected individual fails to thrive during the first year of life. Manifestations such as characteristic facies (facial features), mental retardation, and significant heart murmur distinguish the severe from the mild form of idiopathic infantile hypercalcemia (Franconi et al., 1952; Fraser, Kidd, Kooh, & Paunier, 1966).

Von Arnim and Engel (1964) described the psychological characteristics of the syndrome associated with the severe form of idiopathic infantile hypercalcemia. The affected children were noted to exhibit "an unusual command of language" (von Arnim & Engle, 1964, p. 367) that contrasted with their mental retardation. They also displayed anxiety and -- most relevant to the present report -- hyperacusis and musicality.

Supravalvular stenosis. WS was named by a group of cardiologists (Williams, Barratt-Boyes, & Lowe, 1961) who described characteristic symptoms of a group of four mentally retarded children with supravalvular aortic stenosis and "elfin" facies. In WS, cardiac abnormalities are often mild. Few affected individuals display cardiovascular symptoms although an associated hypertension has been noted in adolescents and adults (Lopez-Rangel, McGillivray, & Friedman, 1992; Morris, Leonard, Dilts, & Demsey, 1990; Pagon, Bennett, LaVeck, Stewart, & Johnson, 1987).

Beuren and his colleagues (Beuren, Apitz, & Hamjanz, 1962; Beuren, Schulze, Eberle, Hamjanz, & Apitz, 1964) reported a number of additional cases in the early sixties and Beuren's name is added to "Williams" in some references (i.e., Williams-Beuren syndrome). He noted that seven of twenty patients with supravalvular aortic stenosis had characteristic facies and the mental and physical retardation typical of WS, while the remaining patients had normal appearance and intelligence. He cautioned that supravalvular aortic stenosis could be viewed as an isolated medical condition that does not necessarily occur in conjunction with WS.

Beuren also reported that 11 children with supravalvular aortic stenosis appeared to display a late stage of infantile hypercalcemia that had not been documented in infancy or early childhood. Severe infantile hypercalcemia was also absent in the cases described by Williams et al. (1961). Nonetheless, links between severe infantile hypercalcemia, supravalvular aortic stenosis, "elfin facies", hyperacusis, and mental retardation with relatively preserved language abilities were subsequently reported by other researchers (e.g., Garcia, Friedman, Kaback, & Rowe, 1964).

Further investigations confirmed that hypercalcemia and supravalvular aortic stenosis are frequently associated with WS, although neither condition is present in all cases (e.g., Greenberg, 1989; Jones, 1990; Morris, Demsey, Leonard, Dilts, & Blackburn, 1988; Prues, 1984). The discovery of the commonalities between the syndromes identified separately by endocrinologists and cardiologists led to a search for a common underlying etiology (Black & Bonham-Carter, 1963; Garcia et al., 1964; Martin, Snodgrass, & Cohen, 1984). Evidence for disturbed calcium metabolism in the absence of hypercalcemia has been found in individuals with WS (Culler, Jones, & Deftos, 1985). Bellugi, Bihrle, Jernigan, Trauner, and Doherty (1990) suggested that a disturbance in the calcitonin-gene-related-peptide (which regulates calcium metabolism and is found in substantial quantities in the brain) is responsible for both the disordered calcium metabolism and the mental retardation.

Genetic research. Recently, a genetic marker for WS was discovered that linked the two syndromes. This genetic anomaly, a submicroscopic deletion on chromosome seven which contains the elastin gene, may explain many of the physical features of the syndrome (Ewart, Jin, Atkinson, Morris, & Keating, 1994; Morris, Thomas, & Greenberg, 1993). Further studies have revealed that deletions greater than 500kb in size are associated with WS, whereas smaller deletions are not associated with the full expression of the syndrome (Morris et al., 1996). The LIMK gene (contiguous with the elastin gene), a novel protein kinase gene expressed in the adult brain, has also been found to be deleted in individuals with WS (Tassabehji et al., 1996). As the LIMK gene is thought to be involved in neural cell development, this gene deletion may contribute to the neuropsychological profile associated with WS.

Diagnosis

Prior to 1996, a diagnosis of WS was most often made on the basis of phenotypic characteristics. Over the past few years, however, the explosion of genetic research concerning WS has led to a the development of a simple blood test (FISH) that can detect the presence of the characteristic deletion on chromosome seven. Across numerous studies, 96% to 98% of affected individuals with a diagnosis of WS tested positive for the genetic deletion with the FISH test (Mari et al., 1996; Metcalf et al., 1996; Morris et al., 1996).

At more and more hospitals, detection of this gene deletion has become an accepted biological marker for WS.

Epidemiology

Because WS is a rare disorder that has only recently been identified, accurate estimates of incidence are not yet available. Existing estimates suggest a range from 1 in 20,000 to 1 in 50,000 live births (Greenberg, 1990; Udwin & Yule, 1991).

Clinical presentation

Individuals with WS display a range of symptoms that vary in severity. For example, supravalvular aortic stenosis is found in most but not all of WS patients. Cardiac symptoms are minimal in most affected individuals, although some infants with WS have required corrective surgery (Morris et al., 1988). Such symptoms may also change over the course of development.

Preus (1984) developed a scoring system for diagnosing WS that was relatively accurate and descriptive but somewhat difficult to use (Greenberg, 1990). In making a diagnosis, Preus considered growth retardation, craniofacial features (i.e., skull and facies), behavioural and medical characteristics of infancy, hyperacusis, associated medical problems, and neuropsychological profiles. The following sections use this system to describe the physical characteristics of children with WS.

Growth. Infants and young children with WS are typically below the 5th centile for height. Although reports of adults with WS are few, those available describe "catch-up growth," with many adults in the low-normal to below-normal range for height (Lopez-Rangel et al., 1992; Morris et al., 1988; Pagon et al., 1987).

Facies. The facial features of children with WS are so distinctive that these children may appear more related to each other than to their respective families. Such features, dubbed "elfin facies" by Lowe et al. (1954), are most distinct between one and five years of age. During this period, typical facies include periorbital (around the eyes) fullness, epicanthal folds (characteristic of the eyes of Asian peoples), stellate iris (lacy iris pattern), a low nasal roof, a long philtrum (vertical groove between the nose and the mouth), narrowing at the temple region, flat mala (cheekbones), full cheeks below the cheekbone, a wide mouth, full lips, and dental malocclusions (misaligned teeth) (e.g., Franconi et al., 1952; Fraser et al., 1966; Lowe et al., 1954; Preus, 1984). These features, in addition to growth retardation, make the children look somewhat "elfin" and younger than their chronological age.

By adulthood, however, people with WS often appear older than their actual age because their features coarsen and their skin appears to age prematurely (Lopez-Rangel et al., 1992; Morris et al., 1988; Pagon et al., 1987). The deletion of the elastin gene may explain the early periorbital fullness and full cheeks as well as the premature aging of the skin in adulthood. As noted above, elastin, which provides the elasticity in tissues, is deficient in these children due to their genetic anomaly (Morris et al., 1993).

Hypercalcemia. Infants with WS are often examined by physicians during infancy because of their failure to thrive. Typical characteristics of infants with WS include feeding difficulties or vomiting, constipation, and irritability (e.g., Arnold et al., 1985; Bellugi et al., 1994; Klein et al., 1990; Pagon et al., 1987; Udwin et al., 1987). These symptoms are also noted in infantile hypercalcemia (e.g., Franconi et al., 1952; Lightwood, 1952). Most infants who are later diagnosed with WS are not tested in early infancy for serum calcium levels. In those who are evaluated, hypercalcemia is often absent or transient. Morris et al. (1990) discovered ectopic (i.e., displaced) calcium deposits and hypercalciuria (i.e., excess calcium in the urine) in some older WS patients, which raises the possibility that disordered calcium metabolism may not decrease with age.

Cardiac and vascular abnormalities. Cardiovascular abnormalities, most often supravalvular aortic stenosis, have been identified in 75-80% of children with WS (Jones & Smith, 1975; Morris et al., 1988; Preus, 1984). Morris et al. (1990) noted that for some patients, cardiac and vascular symptoms (including renal and pulmonary systems) can become progressively worse. They also discovered early onset of hypertension (appearing in the second or third decade of life) (Lopez-Rangel et al., 1992; Morris et al., 1988). The missing copy of the elastin gene would result in less elastic arteries, which, in turn, could lead to early onset of hypertension (Morris et al., 1993).

Neurologic and neurodevelopmental characteristics. Infants and young children with WS are often noted to be hypotonic (i.e., abnormally low muscle tone), hyperreflexic, hyperflexible, and poorly coordinated (e.g., Morris et al., 1988; Trauner, Bellugi, & Chase, 1989). Motor and language development are typically delayed (e.g. Morris et al., 1988; Trauner et al., 1989; Udwin & Yule, 1991). Onset of walking has been reported to range from 1.5 to 4 years; expressive language typically appears between 3 and 4 years of age (Lopez-Rangel et al., 1992; Morris et al., 1988). As the children mature, they tend to become hypertonic (i.e., abnormally increased muscle tone), and orthopedic problems can lead to joint limitations or contractures (i.e., abnormal shortening of muscle tissue). Scoliosis (i.e., curvature of the spine) and lordosis (i.e., forward curvature of the lower spine) are commonly reported. Almost all affected individuals are noted to have a low hoarse voice. Morris et al. (1993) suggested that the progression from hypotonia to hypertonia, the orthopedic problems, and the low hoarse voice may all result from deficient elastin.

Hyperacusis. An overwhelming majority of individuals with WS exhibit hyperacusis (Arnold et al., 1985; Klein et al., 1990; Marriage, Udwin, Oppe, 1992; Martin et al., 1984). Hyperacusis is characterized by aversive reactions to sounds that do not cause such reactions in normal individuals. Children with WS often react so extremely that their hyperacusis interferes with their everyday functioning (Klein et al., 1990; Marriage et al., 1992).

Although the prevalence of hyperacusis diminishes with age, it remains extremely high. Marriage (1992) reported an overall incidence for hyperacusis of 86% in 312 individuals with WS in Great Britain; in her school age sample (n=153), an incidence of 96% for hyperacusis was reported. Numerous studies (e.g., Klein et al., 1990; Marriage, 1992; Myerson & Frank, 1987; Udwin et al., 1987) have reported an 80% to 96% prevalence rate of hyperacusis in individuals with WS. By contrast, hyperacusis is evident in about 3% of normal children (Klein et al., 1990). Thus, hyperacusis is a distinguishing characteristic of individuals with WS. Nevertheless, the cause of hyperacusis in these individuals is unknown. Studies of children with WS typically report normal hearing despite a high incidence (61%) of otitis media (Anonymous, 1985; Klein et al., 1990; Morris et al., 1988). In a study of 65 children with WS, Klein et al.

(1990) found that hyperacusis was not associated with incidence of otitis media.

Associated medical problems. Additional physical difficulties include problems with visual acuity, gastrointestinal symptoms, urinary tract infections, and chronic otitis media (e.g., Greenberg, 1990; Lopez-Rangel et al., 1992; Morris et al., 1988; Pagon et al., 1987; Preus, 1984). These physical difficulties, in addition to hyperacusis and poor visual/spatial skills, make it difficult for individuals with WS to participate in the typical manual work placements open to other retarded individuals, such as those with Down syndrome.

Neuropathology

Neuroimaging studies. Brain imaging studies show mild microcephaly (abnormally small brain) and no evidence of discrete lesions (Wang, Doherty, Hesselink, & Bellugi, 1992). This reduced cerebral volume is similar to that found in individuals with Down syndrome (Jernigan & Bellugi, 1990). Individuals with Down syndrome have been used as a comparison group by some researchers because they demonstrate similar levels of global intelligence (i.e., Full Scale IQ) but display a neuropsychological profile of generalized retardation across motor and cognitive domains (Trauner et al., 1989) that contrasts with the uneven cognitive profile of individuals with WS.

Wang et al. (1992) compared Magnetic Resonance Images (MRIs) of the brains of normal children and children with WS and Down syndrome. Cerebellar volumes were increased in the WS group relative to the Down syndrome group, reaching the levels of normal controls (Jernigan & Bellugi, 1990). A tendency toward dolichocephaly (relative elongation from the frontal lobes to the occipital lobes) was reported for the WS group and brachycephaly (relative expansion of the brain from side to side) was reported for the Down syndrome group (Wang et al., 1992). In addition, the corpus callosum of children with WS was similarly shaped but smaller than those of normal controls (Wang et al., 1992). These differences may help to account for the different neuropsychological profiles of normal children, children with WS, and children with Down syndrome.

Recently, exaggerated left-side asymmetry of the planum temporale (most often associated with language processing) has been reported in professional musicians, with the greatest asymmetry noted in professional musicians with absolute pitch (Schlaug, Janke, Huang, et al., 1995). Because musicality and absolute pitch have been reported in individuals with WS, Bellugi et al. (1996) measured the planum temporale in some individuals with WS using the criteria developed by Schlaug et al. For the majority of their sample, the planum temporale asymmetry was greater than that of professional musicians but less than that of musicians with absolute pitch. This intriguing finding suggests an anatomical basis for the reported strengths in language and music in individuals with WS.

Electrophysiological studies

Electrophysiological studies provide evidence of atypical auditory processing in children with WS. Bellugi et al. (1992) compared event-related potentials (ERP) of eight individuals with WS with a normal control group matched for age. Although auditory ERPs from the brainstem were similar for both groups, atypical patterns of ERPs over the temporal cortex were observed in the WS group in response to a rapidly repeated tone and during presentation of semantically anomalous sentences. Specifically, the WS children displayed larger responses (the N100-P200 responses) to the repeated tone that were less refractory (slow to return to normal levels) than those of the normal controls, suggesting hyperexcitability.

When listening to semantically anomalous sentences, the WS children displayed responses that were abnormal (positive) within the first 200-300 milliseconds following the anomalous word. In addition, the WS children did not display the normal hemispheric asymmetry in response to auditory stimulation that is found in individuals over seven years of age (i.e., greater response in the left temporal region). Bellugi et al. (1992) suggested that the abnormal reactions to auditory stimuli may underlie the hyperacusis and unusual language characteristics of individuals with WS. These responses could also influence the apparent musicality reported in these children.

Neuropsychological profile

The basic cognitive profile of individuals with WS was described in the earliest reports delineating the syndrome (e.g., Beuren, Apitz, & Hamjanz, 1962; von Arnim & Engle, 1964). Only in the past 10 years, however, has there been systematic neuropsychological study of this population. While many results are similar, contradictory findings indicate varied manifestations of the syndrome. In the United States, Dilts, Morris, and Leonard (1990) studied the general cognitive and behavioral characteristics of a large group of children diagnosed with WS, and Crisco, Dobbs and Mulhern (1988) used more specialized measures to further define the neuropsychological profile. In Britain, Udwin, Yule, and their colleagues (Arnold et al., 1985; Udwin et al., 1987; Udwin & Yule, 1990, 1991) conducted a series of neuropsychological and behavioral investigations of children with WS who had been diagnosed with the severe (Franconi) form of hypercalcemia. Reports from smaller samples (e.g., Bellugi et al., 1992) provide corroboration of the basic profile and suggestions for additional research. The results of these neuropsychological studies can provide insight into the similarities between WS and the syndrome of nonverbal learning disabilities (NLD) described by Rourke (1989).

American studies. Dilts et al. (1990) reported on the developmental and behavioral characteristics of 69 individuals with WS (age range 1-32 years), 32 of whom were available for psychological testing (age range; 3-32 years). The mean Full Scale IQ was in the Mildly Retarded range (ranging from severely retarded to average). Although Verbal IQ and Performance IQ scores were not reported, there was evidence of relatively spared linguistic skills. Most of the participants demonstrated age-appropriate grammatical and articulation skills that contrasted with their visual-motor integration skills, which were three standard deviations below the mean. In addition, fine and gross motor skills, as measured by adaptive tasks (e.g., tying shoelaces, cutting with a knife, using a pencil), were lower than would be expected from their age or their mental age based on Full Scale IQ. The findings of this study support the general clinical impression of relatively well developed linguistic skills that contrast with poor nonverbal skills.

Crisco et al. (1988) used the Stanford-Binet Intelligence Scale (Form L-M) and the Illinois Test of Psycholinguistic Abilities (ITPA) to investigate the auditory and visual information processing skills of 22 children with WS (age range: 4 years, 2 months to 10 years, 0 months). A comparison group of developmentally delayed children was matched for age, sex, and global intelligence. Each child was administered the Stanford-Binet, which is heavily weighted with verbal materials (Sattler, 1992), and the ITPA. The researchers described the ITPA as "a measure of information processing, memory, and general rule-learning" (Crisco et al., 1988, p. 651). The mean IQ for both groups was 67.5 (mildly mentally retarded range).

On the ITPA, performance of the WS group and the control group was equivalent on all verbal subtests, including measures of digit span, word understanding, verbal analogies, and grammatical understanding. In addition, performance on a measure of praxis (ability to demonstrate imagined movements) was equivalent. The WS participants performed significantly worse than the controls on visual subtests requiring categorization of images, identification of incomplete visual images, and memory for visual sequences. On a measure of the ability to associate different visual images (e.g., putting a picture of a bone with a dog), however, the WS group performed equivalently to the control group.

The failure to find a relatively superior facility with language among the WS group may have occurred for a number of reasons. Because the Stanford Binet (Form L-M) loads heavily on verbal factors, the control group may have been more comparable on verbal functioning than on global intelligence. Thus, instead of matching groups based on global intelligence, the groups may have been matched on their verbal skills, effectively precluding the possibility of between-group differences. Moreover, because the children were quite young (mean age = 7 years) and because language development is slow and extended in children with WS, delayed language development may also have influenced the results. Finally, the results suggested substantial variation in the language skills of children with WS. Nonetheless, despite the lack of superior verbal skills in the WS group in comparison to the control group, the discrepancy between verbal and visuo-perceptual skills in the WS group was clearly evident.

British studies. In an early study, Arnold et al. (1985) assessed the intelligence of 23 children with WS aged 7-13 years using the Wechsler Intelligence Scale for Children, Revised (WISC-R). Comparison of Verbal IQs and Performance IQs failed to corroborate previous findings of significantly superior verbal abilities in comparison to non-verbal abilities. Interestingly, however, these children attained significantly higher scores on measures of expressive language in comparison to comprehension on the Reynell Developmental Language Scales. The variation in language abilities of children with WS and the fact that a significant proportion (over 30%) of the children failed to score above the floor on the Performance Scale of the WISC-R may have rendered the sample too small for meaningful statistical comparison of Verbal IQ and Performance IQ. Floor effects and low levels of psychometric intelligence may have reduced profile differences.

Additional studies by Udwin, Yule, and their colleagues supported previous findings of significantly better verbal than nonverbal skills and provided further insights into the neuropsychological profile associated with WS. Udwin et al. (1987) administered measures of intelligence, reading, and spelling in 44 children with WS aged 6 to 16 years of age. All of the children were in special schools or receiving special education assistance. Teachers described a pattern of relatively well developed verbal skills that contrasted with extremely poor nonverbal skills. The language of 48% of the children was described as fluent, articulate, and imitative of adult speech and prosody (the stresses, rhythm, and contours of speech). Rote recall of words, phrases, stories and tunes and verbal imitative skills were reported to be well developed in 41% of the group. Ninety-five percent of the children were moderately to severely retarded. Ten of the children scored below the basal level of the WISC-R. The mean IQ of the remaining group was 54.5.

For the group as a whole, Verbal IQ was significantly higher than Performance IQ. For six children, however, Performance IQ was equal to or exceeded Verbal IQ. In terms of academic achievement, only half of the children could be assessed on the reading and spelling measures because the other half was essentially illiterate. Although the literate children were on average 12 years of age, they read at levels equivalent to normal children aged 7 years, 10 months and spelled at levels equivalent to normal children aged 6 years, 10 months. Unlike the Arnold et al. (1985) study, reading accuracy and comprehension were equivalent. Poorer writing (from spelling tests) than reading was due in part to poor hand-eye coordination; several children who were unable to produce words graphically were able to spell orally. This finding highlights the discrepancy between auditory processing and visuomotor skills.

In an additional study, Udwin and Yule (1990) investigated intelligence and expressive language functioning in 43 children with WS (age range 6 to 15 years, 9 months). All participants were mentally retarded: 54% had an IQ below 50. Verbal IQ was significantly higher than Performance IQ. For each child, a loosely structured, 30 minute conversation with an adult was taped and analyzed for characteristics such as mean length of utterance, total number of utterances and words, percentage of idioms and cliches, social phrases and fillers, functional category (e.g., requests, responses, conversational devices), and dysfluencies.

A substantial minority of the children (37%) were found to exhibit speech similar to the "cocktail party speech" described in some children with hydrocephalus or Spina Bifida (e.g., see Dennis, Hendrick, Hoffman, & Humphreys, 1987). Like the speech described by Dennis et al. (1987), the speech of the WS group was characterized as fluent, abundant, and articulate, consisting of an excess of irrelevant personal experiences and an overuse of social phrases, fillers, and perseverations. Udwin and Yule (1990) reported, however, that the content of the WS children's "cocktail party" like speech was not meaningless but communicative in intent. A much smaller minority of the children (16%) did not speak fluently. The contrast between the children with "cocktail party" like speech and those without fluent language highlights the range of verbal expressive abilities found in children with WS.

Twenty children were subsequently chosen at random from the WS sample of 43 and compared to a control sample matched for age and Verbal IQ (Udwin & Yule, 1990). Few differences were reported in quantity or complexity of speech. WS children had a significantly greater percentage of total utterances that were complete and intelligible; they also used significantly more idioms, social phrases, and fillers. Compared to the control group, children with WS obtained significantly higher scores on the Vocabulary subtest of the WISC-R and on the verbal subtests of the Rivermead Behavioral Memory Test, despite the matching based on Verbal IQ. By contrast, perceptual-motor skills of children with

WS were significantly below those of the control group, but there was no difference between groups on a measure of visuo-spatial memory. In another study (Udwin & Yule, 1991), the performance of the WS group was superior to that of the control group on a test of facial memory. This finding suggests that within the domain of visual-spatial skills, some aspects of visual memory may be relatively spared in individuals with WS.

In a follow-up survey of 119 adults with WS (range: 16 years to 39 years), 77% were reported to speak fluently and to use complex sentence structures (Udwin, 1990). The speech of more than half the sample was characterized by incessant chatter (58%) and excessive use of social phrases and cliches (51%). The relatively high incidence of such characteristics compared to children with WS (Udwin & Yule, 1990) implies that linguistic skills continue to develop into adulthood. Only three adults (2.5%) were unable to speak in full sentences.

Forty-two percent of the sample could read books written for children aged 9 or older whereas 20 percent could not read at all. The opposite pattern was described for writing skills; 44 percent of the sample could not write whereas only 24 percent could write understandable letters. The author attributed this finding to continued difficulties with visuomotor skills. Forty-one percent of the sample could manage simple addition and subtraction whereas 18 percent could do no more than count to 10.

The investigations of Udwin, Yule, and their colleagues reveal that although the children with WS whom they studied exhibited a similar neuropsychological profile to that of the samples reported by Dilts et al. (1990) and Crisco et al. (1988), they also demonstrated greater overall cognitive impairment. Udwin and Yule (1990) suggested that their sample, with documented severe infantile hypercalcemia, reflected the more severe end of the spectrum of WS.

Additional studies. Bellugi and her colleagues at The Salk Institute for Biological Studies have published a number of intriguing studies linking the neuropsychological profile, neurophysiology, and neuroanatomy of children with WS. Unfortunately, only results from "carefully selected" children with WS have been reported to date (e.g., 6 children with WS from a group of 50 in Bellugi, Marks, Bihrle, & Sabo, 1988; 10 children with WS from a group of 100 in Bellugi et al., 1992). Selection criteria were not described. Children with Down syndrome provided a comparison sample, matched for age, sex, and Full Scale IQ, which served to highlight the relatively spared language skills of the WS individuals.

Bellugi et al. (1988, 1990, 1994) reported a neuropsychological profile of poor visual and spatial functions in the presence of a relatively good command of the grammatic and semantic aspects of language. Despite delayed language development, children with WS spoke significantly more than their Down syndrome counterparts, using long, grammatically complex sentences with prosodic patterns that contained many intonations and expressions of affect. In spontaneous speech, the WS children used unusual low-frequency words with comprehension. On a timed, verbal fluency task (naming animals), WS children provided unusual, non-prototypical answers as well as the names of prototypical animals. The Down syndrome children, in contrast, produced few responses, which consisted almost entirely of category exemplars and perseverations.

The deficits in visual and spatial skills of the WS group were quantitatively similar to those of the Down syndrome group but qualitatively different. Indeed, the performance of the WS group was remarkably similar to that of adults with right parietal lobe damage despite the fact that there is no evidence of right hemisphere damage in individuals with WS (MacDonald & Roy, 1988). On a task requiring global integration of component parts (e.g., reproducing a triangle made out of circles), WS participants depicted the component parts (e.g., the circles) but failed to represent the global organization. In contrast, children with Down syndrome maintained the global organization (reproducing the triangle) but omitted the component parts. On all visuospatial-organizational tasks (e.g., the Block

Design subtest of the WISC-R, drawing simple objects), children with WS demonstrated poorer visual-spatial integration, closure, and maintenance of the overall configuration. Bellugi et al. (1988) also reported that facial recognition was a relatively intact component of the visuospatial skills of individuals with WS, a finding similar to that of Udwin and Yule (1990).

In other studies, Bennett, LaVeck, and Sells (1978), MacDonald and Roy (1988), and Pagon et al. (1987) evaluated small groups (7 to 9 participants) of children with WS. On various measures of psychometric intelligence, verbal scores exceeded performance scores for most participants. Participants scored uniformly poorly on tests of fine motor coordination; performance on measures of gross motor skills was variable.

In recent studies (Finegan, Smith, & Meshino, 1996; Mervis et al., 1996; Vicari et al., 1996; Volterra et al., 1996) more detailed examinations of language skills revealed relative strengths in immediate auditory memory and phonological fluency in comparison to Verbal IQ. By contrast, in two reports, long-term auditory memory was reported to be equivalent to Verbal IQ (Finegan et al., 1996; Vicari et al., 1996). In addition, normal levels of phonological encoding with impaired access to lexical-semantic knowledge were reported (Vicari et al., 1996).

Summary. Neuropsychological studies of children with WS substantiate the early clinical reports of relatively intact linguistic skills in contrast to extremely poor visual-spatial and visual-motor abilities. In addition, most of the children were reported to score in the mildly to moderately mentally retarded range on tests of intelligence, with scores ranging from profoundly retarded to average. Individuals with diagnosed hypercalcemia in infancy may represent a severely affected subset of individuals with WS.

Verbal skills in individuals with WS may appear unusually good because of the contrast to their extremely poor visual-spatial skills. As a group, individuals with WS appear articulate and fluent in their speech and reportedly demonstrate intact prosody. When individuals with WS are compared to controls matched for Verbal IQ, many language skills appear equivalent. In the British studies, however, relatively better vocabulary and verbal memory were reported in the WS participants despite matching by Verbal IQ. In recent studies, immediate auditory memory and phonological encoding were identified as strengths within the verbal domain. By contrast, long-term memory was not a relative strength in the auditory domain. When individuals with WS were compared to controls matched for global IQ, however, overall language skills of the WS groups were generally much better than those of the control groups.

In all studies, extremely poor visuospatial skills were reported for individuals with WS, who had remarkable difficulty with spatial organization and representation of visual material. In addition, individuals with WS had poor visuomotor coordination. One relatively intact aspect of nonverbal processing was revealed by the performance of WS children on tasks requiring memory for faces. Information regarding general visual memory was equivocal; some studies reported relatively better visual memory than visual perceptual skills, while others reported no difference.

Although overall linguistic skills of children with WS appeared relatively intact, less complex linguistic skills (those requiring intact auditory pattern perception and auditory memory) were better preserved than more complex aspects (such as comprehension, reading, and writing). Based on these findings and anecdotal accounts of good musical skills in children with WS, the purpose of the present study was to investigate whether other domains of auditory functioning such as music are also relatively intact in children with WS. Music provides an ideal domain for the study of auditory functioning. Like language, music is a universal aspect of human cultures that includes simple and complex components arranged in hierarchical structures.

The discrepancy between relatively superior verbal skills and inferior visuospatial-visuomotor skills in children with WS is striking. An explanation for this unusual neuropsychological profile is needed to further our understanding of WS in particular, and of auditory pattern processing in general. The model of NLD (Rourke, 1989) provides a promising explanation by incorporating a developmental perspective.

Nonverbal Learning Disabilities

Individuals with WS have neuropsychological profiles that are remarkably similar to those of individuals with NLD (Anderson & Rourke, 1995). Individuals with NLD are thought to develop their verbal assets from a subset of intact auditory perception and memory functions that underlie many dimensions of linguistic functioning. These same intact auditory functions may very well underlie relatively intact music processing skills.

The NLD syndrome is considered to be the developmental outcome of the interaction between basic <u>assets</u> in auditory perception, simple motor skills, and an ability to learn in a rote fashion, and basic <u>deficits</u> in nonverbal processing and adaptation to novelty (Rourke, 1989). A summary of the NLD model (Rourke, 1989, 1995; Rourke & Tsatsanis, 1996) follows.

During infancy and early childhood, motor development in children with NLD is generally delayed. Primary deficits in tactile and visual perception, poor psychomotor coordination, and difficulty in adapting to novelty lead to secondary deficits in deployment of visual attention and decreased exploration of the environment. As a consequence, much of what is typically learned through hands-on experience is unavailable to children with NLD. Nonetheless, primary assets in auditory perception, rote learning, and simple motor skills lead to well developed auditory and verbal attention and memory. Once these skills develop, an individual with NLD tends to explore and connect to the world through language.

This reliance on learning about the environment though language leads to secondary assets in selective and sustained attention for simple verbal material presented auditorially. Simple auditory skills involving rote verbal memory, verbal repetition, and phonological and syntactical skills become well developed in children with NLD but their deficits in nonverbal abilities and adaptation to novelty restrict the development of more complex language skills. For example, individuals with NLD typically experience difficulty understanding complex linguistic information that requires interpretation of contextual information (e.g., the appreciation of humour and irony). Children with NLD pay little or no attention to speech prosody and their memory for complex verbal and nonverbal material is poor. Moreover, their difficulty in integrating nonverbal and verbal aspects of their experience leads to poor social skills because information communicated through nonverbal expression, such as mood and lack of interest, is poorly perceived or misunderstood. For example, individuals with NLD are often overly talkative and do not perceive when the listener becomes bored or irritated.

In terms of academic abilities, nonverbal deficits result in delayed reading and spelling. Single-word reading and spelling develop rapidly after a delayed start, but poor reading comprehension is evident as academic demands increase. Tasks that require rote learning and auditory attention and memory are relatively easy for children with NLD. By contrast, tasks demanding visual-spatial-organizational skills, reading comprehension, mechanical arithmetic skills, and cognitive flexibility become increasingly difficult for these children as they mature. In addition, higher level cognitive skills such as concept formation, problem solving, and strategy generation -- which depend on the integration of nonverbal and verbal skills -- are also poor. Thus, NLD is the developmental outcome that arises from a set of primary nonverbal deficits and primary auditory assets.

Rourke (1982, 1989) observed that the deficits associated with NLD were those primarily subserved by right hemisphere systems, whereas the assets were found in functions primarily subserved by the left hemisphere. Based on the findings of Goldberg and Costa (1981), Rourke (1982, 1989) proposed that the greater the amount of generalized white matter destruction, the more likely the NLD syndrome would be evident. Although right hemisphere white matter is thought to be important during all stages of life for the development and maintenance of intermodal integration and the acquisition of new descriptive systems, left hemisphere white matter is less important for the maintenance of automatized processes. Thus, NLD would be expected as the result of generalized white matter dysfunction in childhood. Such white matter dysfunction has been observed in children who exhibit features of the NLD syndrome (e.g, those with agenesis of the corpus callosum) (see Rourke, 1987, 1988, 1989; Rourke, Bakker, Fisk, & Strang, 1983).

Goldberg and Costa (1981) investigated the anatomical differences between the right and left hemispheres of the brain and found that the ratio of white to grey matter is larger in the right hemisphere than in the left hemisphere. Based on these findings and additional work concerning the lateralization of functions, they concluded that the right hemisphere is superior at intermodal processing, whereas the left hemisphere is better suited for automatized or single modal tasks. Although investigations of brain structure function in WS are few in number, the relatively small corpus callosum reported by Wang et al. (1992) is suggestive of white matter dysfunction in individuals with WS. Most importantly for this study, the neuropsychological profile associated with WS closely resembles the typical profile of the NLD syndrome.

A comparison of the primary and secondary assets and deficits in NLD and WS

The assets and deficits of the NLD syndrome (Rourke, 1989) are listed below and compared with findings from the WS literature.

Assets.

1. **NLD:** Auditory perceptual capacities are well developed.

WS: The relatively intact speech perception and production abilities that are generally reported in individuals with WS suggest that their auditory perception abilities are relatively well developed. Moreover, their hyperacusis may indicate heightened auditory sensitivity (e.g., Bellugi et al., 1994; Udwin et al., 1987).

2. **NLD:** Simple repetitive motor skills are generally intact.

WS: Information regarding simple motor skills in individuals with WS is limited. Gross and fine motor skills are typically reported to be poor (e.g., Morris et al., Udwin & Yule, 1991). MacDonald & Roy (1990) reported equivalent performance on a finger tapping test for children with WS and matched controls.

3. **NLD:** Ability to learn in a rote fashion is intact.

WS: Studies of long-term auditory memory suggest that these skills are less developed than immediate auditory memory (Finegan et al., 1996; Vicari et al., 1996). The relative strength or weakness of rote learning, however, has not been systematically investigated. The good memory for songs reported in individuals with WS suggests well developed rote memory (e.g., von Arnim, 1964; Udwin et al., 1987).

4. **NLD:** Selective and sustained attention for auditory and verbal material is good.

WS: On a measure of immediate auditory memory (forward digit span), the performance of children with WS was a relative strength (Finegan et al., 1996; Mervis et al., 1996; Vicari et al., 1996; Volterra et al., 1996), suggesting relatively intact auditory attention.

5. **NLD:** Auditory memory and rote memory for material that is verbally encoded are good.

WS: Anecdotal accounts suggest that individuals with WS have good memory for songs and poems (von Arnim & Engel, 1964). On tests of verbal memory, children with WS generally perform at equivalent or higher levels than controls matched for overall verbal ability (Crisco et al., 1988; Udwin & Yule, 1990).

6. **NLD:** Individuals with NLD are described as having excellent phonemic hearing, segmentation, blending, and repetition, as well as a high percentage of speech output involving rote verbal material and verbal associations.

WS: Although there is variation in verbal fluency and speech output, the majority of children with WS are reported to be talkative with well-articulated, grammatically correct speech (Bellugi et al., 1994; Crisco et al., 1988; Udwin et al., 1990). In addition, a significant proportion of individuals with WS appear to rely on social phrases and cliches in their conversation (Udwin, 1990; Udwin et al., 1990)

Deficits.

1. **NLD:** Bilateral tactile-perceptual deficits are described, usually more marked on the left side of the body.

WS: Information regarding tactile-perception has not been reported.

2. **NLD:** Bilateral psychomotor coordination deficiencies are described, often more marked on the left side of the body.

WS: Evidence for poor psychomotor coordination is overwhelming. For example, Dilts et al. (1990) reported that children with WS performed poorly on psychomotor coordination tasks such as tying their shoelaces or cutting with a knife.

3. **NLD:** Impaired visual perception (e.g., impaired discrimination and recognition of visual detail and visual relationships) and extremely poor visual-spatial-organizational abilities are common.

WS: Children with WS perform poorly on most measures of visual perception. For example, Crisco et al. (1988) reported that children with WS performed significantly worse than controls matched for Full Scale IQ on measures that required the categorization of images, the identification of incomplete visual images, or memory for visual sequences. By contrast, on a measure of ability to relate images (e.g., putting a picture of a dog with a picture of a bone), children with WS performed as well as controls (Crisco et al., 1988). Moreover, relatively intact performance on measures of facial memory and recognition indicates that some visual perception skills are relatively intact (Udwin & Yule, 1991; Bellugi et al., 1988).

4. **NLD:** Adaptation and accommodation to novelty are poor.

WS: Although specific information regarding adaptation to novelty is sparse, relevant findings suggest that individuals with WS are deficient in this area. For example, individuals with WS perform poorly on the nonverbal subtests of the Weschler Intelligence tests, which are thought to assess aspects of fluid (flexible and adaptive) intelligence (Sattler, 1992).

5. **NLD:** Visual and tactile attention are poor.

WS: Information in this area is limited, but performance on measures of visual attention and memory suggests that visual attention is poor (e.g., Crisco et al., 1988).

6. **NLD:** Exploratory behaviours are diminished.

WS: Again, information regarding exploratory behaviours has not been reported. Dilts et al. (1990) found, however, that many parents described their children as cautious when climbing stairs and encountering uneven surfaces. Such caution may indicate decreased exploratory behaviours.

7. **NLD:** Individuals with NLD have poor visual and tactile memory and poor memory for complex verbal material.

WS: Reports regarding visual memory are equivocal. Whereas Crisco et al. (1988) reported poor performance on a measure of memory for visual sequences, Udwin and Yule (1990) found no difference between a WS group and a control group (matched for VIQ) on a measure of visual-spatial memory. Moreover, facial memory appears to be a relatively spared aspect of visual memory.

8. **NLD:** Concept formation, problem solving, strategy generation, and hypothesis testing are poor.

WS: Little information regarding these areas has been reported. Poor performance on the Category Test (a measure of problem solving and the ability to benefit from informational feedback) was identified among the children with WS described by Bellugi et al. (1992).

9. **NLD:** People with NLD are described as having mild problems with oral-motor praxis and poor prosody. They exhibit verbosity of a repetitive, straightforward, rote nature.

WS: In general, good verbal articulation has been reported in children with WS (Dilts et al., 1988). In one study (Bellugi et al., 1990), mild oral-motor praxis problems were reported. Speech prosody is reportedly intact in individuals with WS. Bellugi et al. (1994) described prosody in individuals with WS as expressive and imitative of the intonations of adult speech (Udwin et al., 1987). In addition, in a survey of adults with WS, just over half were overly talkative and repetitive, with an overuse of social phrases and stereotyped utterances (Udwin, 1990). Thus, it is unclear whether the reportedly intact speech prosody represents an extension of well learned rote auditory patterns or spontaneous affective expression in speech.

10. **NLD:** Individuals with NLD demonstrate greater facility with structural aspects of language than with the semantic content and functional use of language.

WS: Unlike other retarded children, children with WS are noted for their use of complex, syntactically complete sentences (e.g., Bellugi et al., 1990; Udwin et al., 1990). On measures of comprehension, however, their performance is typically equivalent to that of other retarded children (e.g., Crisco et al., 1988).

Summary: Comparison of NLD and WS. The profile of relative strengths and weaknesses of children with WS closely parallels the profile associated with NLD syndrome. Although the level of skills in all areas is lower in children with WS than in children with NLD, the pattern of assets and deficits is strikingly similar for both groups. Children with WS display assets in auditory perception, auditory attention, verbal memory, speech articulation, and quantity of speech, but deficits in psychomotor coordination, visual perception, visual attention and memory, visual-spatial-organization, and adaptation to novelty. Areas of strength that appear unique to children with WS include facial memory and, possibly, speech prosody.

Music in children with WS

Little has been reported on the musical skills and abilities of children with WS. For three of the four children described by von Arnim and Engel (1964), good singing skills and memory for songs or poems were identified, and Udwin et al. (1987) reported that children with WS easily learn songs, stories, and names. Individuals with WS also learn complex rhythmic patterns easily and they can learn songs in foreign languages with nearly perfect word pronunciation (Lenhoff, 1996). Moreover, in the Anonymous (1985) case report of a child with WS whose "truest love" was music, the child began taking piano lessons at age 7 and was discovered to have absolute (perfect) pitch.

The vast majority of normal individuals process pitch in a relative rather than an absolute manner. Absolute pitch is most often (but still rarely) found in individuals who have had formal music training beginning at age 7 or younger (Takeuchi & Hulse, 1993). Absolute pitch processing is the norm in non-human mammals (e.g., D'Amato & Salmon, 1984) and songbirds (e.g., Hulse, Cynx, & Humpal, 1984) and has been reported in some mentally retarded children (see

Ward & Burns, 1982). Thus, although absolute pitch is often thought to be a musical gift, it may indicate a rather primitive level of pitch processing in humans.

More recently, increased interest in the music skills of individuals with WS has led to the development of a summer "music" camp for these individuals (Lenhoff, 1996) as well as to the neuroanatomical investigation of the planum temporale (Bellugi et al., 1996).

Implications of NLD for music. Assets in verbal skills and on rote tasks observed in individuals with NLD are thought to develop from primary assets in auditory perception and rote memory (Rourke, 1989). If language were a strictly modular skill, auditory strengths in language would not be expected to generalize to music. However, should some aspects of language and music processing be based upon shared auditory processing skills, then the primary assets described for the syndrome of NLD could be applied to music as well as to language.

Although the model for the syndrome of NLD does not address the issue of modularity or of musical skills, it can be used to predict a pattern of assets and deficits in music perception if common aspects of auditory processing are assumed to be shared between domains of music and language. Good auditory perception and rote memory would lead one to expect a well developed memory for simple and repetitive musical patterns. Such patterns can be simple and repetitive in terms of their rhythms or their pitch structures.

Simple melodies often consist of repeated, integrated patterns of rhythm and pitch. Speech prosody may be closely related to melody (Fernald & Mazzie, 1991), in that prosody also integrates rhythm and pitch. Given the poor perception and production of speech prosody described in children with NLD, however, it is unclear what predictions would be made about their ability to process simple melodies. In adult speech, prosodic <u>variability</u> is the norm, whereas in melodies from popular, folk, and children's songs, <u>repetition</u> with little variability is the norm. Because of the simple structure and repetitive nature of many melodies, it might be expected that children with the NLD syndrome would have average to above average skills in processing simple melodies. By contrast, their perception of linguistic prosody might be expected to be poor because it is more variable. Alternatively, because the right hemisphere is considered dominant for melodic processing (e.g., Milner, 1962; Peretz, 1990) and access to right hemisphere systems is dysfunctional in NLD, poor processing of melody <u>and</u> prosody might be expected.

<u>Implications of NLD for WS and music.</u> As noted above, children with WS exhibit a pattern of assets and deficits that closely parallels the neuropsychological profile associated with NLD. Thus, predictions about the music skills of individuals with WS would be similar to those made for individuals with NLD. Nonetheless, just as the level of performance of individuals with WS on measures of verbal and nonverbal skills is depressed in comparison to NLD, their overall level of performance on measures of music skills should also be lower.

The primary neuropsychological assets in auditory perception and rote verbal memory evident in children with WS and NLD suggest that their perception of rhythm should be relatively intact. Although it could be reasoned that the perception of melody would be poor in children with NLD, the relatively intact speech prosody reported in children with WS raises the possibility that their abilities in melodic perception and memory might also be relatively intact. In addition, their memory for simple songs and rhythmic patterns would be expected to be relatively well developed. By contrast, more complex skills requiring complex intermodal processing, such as reading music, would likely be particularly difficult for the WS population. Complex tasks such as reading and producing music are generally mastered by only a small percentage of the population, at least in Western society. Thus, predictions regarding the acquisition of these complex skills cannot be tested fairly by observing the level of skill manifested by individuals without specialized music training. Only musical skills that are generally available across members of a culture would provide fair tests of relative ability. Music processing skills that are evident in infants provide a good measure of general auditory pattern processing abilities because these skills are evident without years of exposure to music. Musical experiences that are typically available to most members of a culture provide the basis for implicitly learned music skills and would also be relevant for assessing general skills in auditory pattern processing.

Music processing

Although most of us have an implicit understanding of what music is, when we try to define it systematically our task becomes extremely complex. Trehub and Schellenberg (1995) describe a musical auditory stimulus as one that is non-random, non-referential, and humanly generated. Thus, the process of music perception and cognition is, essentially, the discovery of patterns among tones. Because musical stimuli can have a complex organization without being referential, they may be ideal for testing auditory pattern perception.

Theoretical models of music processing

Modularity refers to autonomous aspects of cognition that occur automatically and quickly (Fodor, 1983). According to Fodor, modular functions are: (1) domain specific, (2) innately specified, (3) hardwired, and (4) autonomous. Non-modular functions occur in processes that are not domain specific and require inter-modal processing (e.g., problem solving and the generation of ideas). Modular and non-modular processing roughly coincide with the notions of crystallized (automatized) and fluid intelligence. According to Fodor, however, modular functions are innate, whereas many aspects of crystallized intelligence are learned. Once a learned function is automatized, it is processed automatically and quickly in a manner similar to modular functions. Automatic auditory functions are relative assets in people with NLD, whereas functions that require intermodal processing (fluid intelligence) are deficient. Thus, the assets described in NLD could represent intact modular-like functions, whereas the deficits may represent poor non-modular processing.

Modular systems are thought to be affected by brain damage in an all-or-none fashion; if a specific module is impaired, its function as a whole is lost (Fodor, 1983). Thus, functions that are found to be dissociable in brain damaged individuals are likely candidates for modular processing. By contrast, non-modular functions are thought to be affected by brain damage in an incremental fashion; the degree of impairment depends on the degree of damage (Fodor, 1983). Functions that become relatively inefficient but remain present following brain damage are considered non-modular.

Whereas Fodor's proposed modules encompassed whole domains such as language and music, Jackendoff (1987) proposed that the need for integration and interpretation of various processes within a domain (e.g., language, music) requires smaller processing modules. For example, perception and expression may be seen as different processes within the domain of language that require smaller processing modules. If Fodor's theory of autonomous, impenetrable, domain specific modules is correct, then only one type of aphasia or amusia would occur and links between the two would not be expected. Studies of brain damaged individuals reveal a wide variety of aphasias and amusias, however, and provide support for Jackendoff's proposition that smaller processing modules exist within a domain. Nonetheless, the co-occurrence of many aphasias and amusias suggests that some aspects of music and language processing may be linked. If some aspects of music and language were found to stem from a general ability, this would constitute evidence against the domain specificity proposed by modular theorists.

Amusia

Henschen (1920, cited in Judd et al., 1983) reviewed 314 historical cases of reported amusia and found that in 309 of the 314 cases, left hemisphere damage was present; 301 (97%) of these patients were also aphasic. Receptive and expressive amusias and aphasias were described, as well as alexias (inability to read) for both music and language. Although these associations suggest a link between music and language processing, definitive evidence requires more detailed information regarding the deficits and the extent and location of the lesions.

Five cases of amusia without aphasia following unilateral right hemisphere damage were also reported (Henschen, 1920, cited in Judd et al., 1983). Interestingly, in each of those five cases, the ability to whistle or sing in tune was impaired. These findings suggest that the right hemisphere is involved in processing the tonal aspects (pitch relationships) of music. It is unclear, however, whether the right hemisphere damage was affecting the patients' music comprehension abilities, their performance abilities, or both. Milner (1962) reported acquired impairments of tonal memory and timbre discrimination in patients who had undergone right temporal lobectomies, which is consistent with the concept of right hemisphere involvement in processing the non-rhythmic aspects of music.

Additional dissociations in music processing have been reported. Peretz (1990) assessed melodic and rhythmic processing in a sample of brain damaged non-musicians and found that perception of melodic contour (the global aspects of melody involving upward and downward changes in pitch) was disturbed in patients with right hemisphere damage. In some, bilateral damage also resulted in poor processing of pitch intervals (a more local aspect of melody involving the distance in pitch between two tones). Meter in music (e.g., the 3/4 time of a waltz or the 2/4 time of a march) involves the temporal relations in music suggested by the time signature of a piece, whereas rhythm (e.g., reggae, disco or samba rhythms) involves the emphasis of some beats but not others within the metrical structure. In patients with right or left hemisphere damage, the perception of meter was relatively preserved in comparison to rhythm (Peretz, 1990). Although deficits in temporal processing were not lateralized in the group as a whole, within individuals, melodic and temporal processing of melody were dissociated, suggesting that these aspects of music may be subserved by separate systems.

Peretz and Morais (1989) reported double dissociations between rhythmic and melodic processing. Rhythmic but not melodic processing was disturbed in a patient suffering left hemisphere damage, whereas melodic but not rhythmic processing was affected in an individual suffering from right hemisphere damage. Peretz and Morais (1988) also reported a left ear (presumed right hemisphere) advantage for the processing of melodic contour in both normal and brain damaged individuals. In these studies and others (e.g., McKinnon & Schellenberg, 1997), evidence for right hemisphere involvement in processing the pitch contour of melody is fairly consistent (see Marin, 1982 for a review).

By contrast, evidence for lateralization of rhythmic features or metrical structure is inconclusive.

In a unique study, Patel and Peretz (1994) examined melodic and prosodic processing in two patients with amusia without aphasia. Both patients had undergone bilateral temporal-lobe surgery over seven years previously. In one patient, the perception of prosodic and melodic elements was intact, whereas in the second patient, both melodic and prosodic processing was impaired. Although the results differed between patients, they suggest a link between the processing of melodic and prosodic elements.

Dissociations in more complex musical skills such as composition, music reading, and instrumental performance have also been reported (e.g., Brust, 1980; Judd et al., 1983). These dissociations suggest that music processing in musicians involves many types of skills. Although such cases of amusia provide insights into music processing in trained musicians, they do not necessarily further our understanding of music processing in children or in the general (musically untrained) population. Nonetheless, the findings from the studies of amusia provide interesting suggestions about how music is processed in the brain. Both musicians and non-musicians appear to process many aspects of music in a modular fashion (Peretz & Morais, 1989).

Unfortunately, it is difficult to generalize findings from brain damaged adults to children. The outcome of brain damage in childhood may be quite different from the outcome of brain damage in adulthood (Hecaen & Albert, 1986), which indicates that brain organization and function is somewhat different for children than for adults. For example, a profound expressive aphasia would be one of the expected outcomes of a left hemispherectomy in an adult. When such damage occurs early in childhood, however, some aspects of expressive language typically approximate normal functioning (Kolb & Whishaw, 1990).

Music development in normal individuals

Studies with infants provide evidence that some aspects of adult music processing abilities are present in infancy (see Trehub & Trainor, 1993, for a review). Skills that are evident in infancy are unlikely to result from exposure or training. Hence, they are more likely to be candidates for musical universals and more likely to stem from auditory processing predispositions. Such skills might also be likely candidates for modular (or automatized) processing, and could be expected to be relatively well developed in individuals with WS. By contrast, skills that appear later in ontogeny are likely to require more complex processing and would be expected to be less developed in individuals with WS.

Infants are able to detect changes in simple repeated rhythmic patterns (Chang & Trehub, 1977; Demany, McKenzie, & Vurpillot, 1977). They can also recognize changes in metric organization and tempo (e.g., Chang & Trehub, 1977; Demany et al.,1977; Trehub & Thorpe, 1989). Unfortunately, research concerning infants' rhythmic perception is limited (Dowling & Harwood, 1986). Moreover, results from studies of arhythmia in brain damaged individuals are equivocal (e.g., Judd et al, 1983; Peretz & Morais, 1989; Shuter-Dyson, 1982).

By contrast, our knowledge of infants' perception of melody is relatively well developed (see Trehub & Trainor, 1993, for a review). Infants can readily detect changes in the global contour of melody (e.g., Chang & Trehub, 1977; Trehub, Bull, & Thorpe, 1984). They find it more difficult, however, to detect transpositions (when the beginning pitch of the melody is changed but the relations between consecutive tones remain constant) or changes in the interval structure that maintain the melodic contour (see Trehub & Trainor, 1993). Both infants and adults show a left ear advantage (a presumed right hemisphere advantage) in detecting changes that alter the contour of a melody (Anderson, 1994; McKinnon & Schellenberg, 1997). Thus, results from studies of infants, amusic patients, and normal adults converge in implicating involvement of the right hemisphere in processing melodic contours (changes in pitch direction) (e.g., Dowling & Harwood, 1986; Judd, 1988; Marin, 1982; Peretz, 1990), just as the left hemisphere appears innately organized to process language (e.g., Hecaen & Albert, 1986).

"Motherese" or infant directed speech (baby talk) appears to capitalize on the auditory pattern processing skills of infants. Motherese is characterized by exaggerated prosody in comparison to normal speech. It is also higher in pitch with simple pitch contours that span a greater range. Motherese typically involves relatively slow and regular rhythmic patterns, shorter phrases, and greater repetition than speech directed to adults (Papousek & Papousek, 1981). Lullabies (infant directed songs) appear to be similar to infant-directed speech in terms of their melodic contour and rhythmic characteristics (Trehub & Schellenberg, 1995). Such similarities are consistent with the notion of shared processing for speech and music. As noted, basic skills involving the perception of pitch and rhythm that are present in infancy are likely to be innate and to represent the most simple aspects of auditory pattern processing. Hence, they may also be well developed in individuals with WS.

Other aspects of music perception clearly develop over time and appear more specific to music. For example, systems of tonality (the organization of tones around a reference tone, or tonic) differ across cultures and sensitivity to tonality develops with greater exposure to music (Krumhansl & Keil, 1982). This does not preclude the possibility that some aspects of tonality may stem from general auditory processing abilities. Indeed, even infants are better at detecting changes in successive melodies when they are presented in keys considered to be closely related in Western music (Trainor & Trehub, 1992). The tonics of closely related keys stand in a frequency ratio of 3:2. This relatively simple (small integer) ratio appears to be innately easy to process (Schellenberg & Trehub, 1996b). Hence, the structure of Western scales, which is based on simple frequency ratios between tones, could be partly determined by processing predispositions for such ratios (Schellenberg & Trehub, 1994a, 1994b, 1996a, 1996b). Although, processing predispositions may form the basis for tonality, it is clear that much of the tonal system of one's musical culture is learned, perhaps in a manner similar to the acquisition of one's native language.

Moog (1976) and Dowling and Harwood (1986) studied musical development in children and found a typical progression in the acquisition of musical skills. Between 1 year and 18 months children begin to sing spontaneously although their pitch and rhythmic patterns are indeterminate (Dowling & Harwood, 1986; Moog, 1976). By 3 to 4 years of age, children are able to sing entire songs and their pitch and rhythmic patterns approximate those of their culture (Moog, 1976). At age 5, children are sensitive to key membership but not to implied harmony (Trainor & Trehub, 1994). By age 7 or 8, implicit understanding of implied harmony becomes evident (Dowling & Harwood, 1986; Trainor & Trehub, 1994).

The ages at which different musical skills are acquired provides a measure of their difficulty and the degree to which they are likely to be the result of culturally determined learning. Skills that appear later in childhood, but do not require specific training (e.g., the understanding of implicit harmony and the increased ability to match rhythmic patterns) may reflect implicit learning based on general intact auditory pattern processing. Skills that depend on general auditory pattern processing abilities would be expected to be relatively well developed in individuals with WS. By contrast, skills that require specialized training and intermodal integration should be poor. Unfortunately, music skills (unlike language) are not typically developed through standardized school programs. Thus, only skills that are present in infancy or acquired implicitly through exposure to music provide a basis for the evaluation of general auditory pattern processing abilities.

Summary

Like all abilities, music processing reflects contributions of nature (innate abilities) and nurture (learned abilities). The templates for the processing of melodic contour and its probable right hemisphere dominance appear to be well established in infancy. Infants also show evidence of the ability to detect changes in temporal groupings (e.g., rhythmic patterns, meter, and tempo).

The exaggerated prosody of "motherese" and its similarity to the melodic contour of lullabies sung to infants also raises the possibility of innate, shared aspects of prosodic and melodic processing in infancy. Prosodic and melodic processing may also be linked in adulthood (Patel & Peretz, 1994).

Processing of specific pitch intervals, perception of within-key relationships, and perception of implied harmony appear to develop during early childhood even without formal music lessons. In addition, children's skill in the perception of the rhythmic aspects of music improves with age. Studies of normal musical development and amusia and theoretical models of music and language processing provide a basis for evaluating the music skills of children with WS.

Conclusions

An exploration of the language and musical abilities of children with WS could further our understanding of their unique neuropsychological profile and its relationship to the syndrome of NLD. Specifically, investigations of melodic and rhythmic processing and musical development in children with WS would complement our knowledge of their linguistic abilities. Moreover, the integration of these results with the findings from neuropsychological studies should broaden our understanding of auditory pattern processing in general. Such an investigation could provide evidence that language and music functioning are based on general principles of auditory pattern processing. The implications of such a finding would challenge the proposal of autonomous processing for language and music suggested by modularity theorists. In addition, the results would further inform the developmental model of NLD and suggest fruitful avenues for future research.

Rationale for the study

Music, like language, is a universal aspect of human cultures. Children with WS are limited in many ways, but appear least limited in language and, perhaps, music. If relatively well developed music perception abilities exist in these children, further development of their abilities in this area could enrich their lives by providing skills for leisure and social activities. Systematic investigation of these abilities has yet to be conducted.

Relatively preserved verbal skills and musical skills in this population would imply that intact auditory pattern processing abilities are the source of these skills. The study of auditory pattern processing in children with WS would help to evaluate theories that propose dissociations of processing for music and language. For example, if general auditory pattern processing abilities seem to determine linguistic and music abilities, this would present a challenge to theories that isolate language from other aspects of auditory functioning (Fodor, 1983; Jackendoff, 1987). A positive association between linguistic and musical abilities could be predicted from other theoretical perspectives, however, such as Rourke's (1989) developmental model of the NLD syndrome, or Trehub and Trainor's (1993) developmental perspective on the processing of prosody and melody. Thus, this study could provide evidence relevant to the ongoing debate between modularity theorists -- who postulate discrete processing of language and music -- and developmental theorists -- who suggest the possibility of shared processing at basic levels.

Hypotheses

1. Children with WS (like those with NLD) will display significantly higher scores on the Verbal-Comprehension factor than on the Visual-Perception factor of the WISC-III.

2. The level of performance of children with WS on a normative measure of music processing, Gordon's (1980) Primary Measures of Music Audiation (PMMA), will be comparable to their level of performance on a measure of verbal ability (Peabody Picture Vocabulary Test-Revised). In addition, their level of performance on the PMMA will be higher than their level of performance on a measure of nonverbal skills (Visual-perception factor of the WISC-III).

3. Levels of performance on measures of language and music will be correlated, such that individuals with better linguistic skills will also have better music skills.

4. Results from the Child Music Interest Interview will be consistent with the notion of relatively preserved music perception in this population. Specifically, children with WS will exhibit an interest in music that is similar to that of normal children of equivalent mental age.

Chapter II

METHOD

Participants

The study group was comprised of 20 children with Williams Syndrome (WS) who were between 8 and 13 years of age. Data from one participant could not be used because he performed below floor levels on the measure used to establish mental age. Thus, 19 children were included in the final sample (10 boys and 9 girls). The comparison group consisted of 19 children (11 boys and 8 girls) who were selected from a larger group of 32 children between 5 and 12 years of age without mental or physical handicaps. Because 'average' children were sought for the comparison group, only children with standard scores on a measure of receptive vocabulary (Peabody Picture Test-Revised) between 85 and 115 were selected.

Mean chronological age of the WS group was 10 years, 6 months (SD = 1 year, 10 months), mean mental age was 8 years one month (SD =2 years, 2 months). Mean chronological age of the comparison group was 7 years, 11 months (SD = 2 years 4 months) and mean mental age was 8 years 1 month (SD = 2 years 5 months). Twelve children in the WS group and 10 children in the comparison group had a past history of otitis media. Of those, two children in the WS group and one child in the comparison group had an ear infection in the past year. Three children in the WS group and one child in the comparison group had a past history of tubes in their ears for treatment of chronic ear infections. One child in each group was reported to have a past history of hearing loss, but no child had a reported hearing impairment or ear infection at the time of testing. Two children in the WS group and three children in the comparison group were taking private music lessons at the time of testing. In sum, groups did not differ on sex, mental age, history of otitis media and its complications, or musical background.

Procedure

Data collection took place in two stages. Stage 1 involved collecting data in the province of Ontario and extended from May to August, 1995. The second stage of data collection took place in New England from September, 1995 to September, 1996. Similar recruitment procedures were followed in each location.

Participants for the WS group were recruited from the Ontario division of the Canadian Association for Williams Syndrome and from the National Williams Syndrome Association of the United States. These associations are comprised of parents or guardians of children with WS and concerned professionals. Parents or guardians of participants with WS were invited to participate in the study through association newsletters. Parents or guardians and their children were also invited to participate during meetings of the Canadian Association for Williams Syndrome held in Ontario. Children for the comparison group were recruited through fliers posted at local churches and at the University of Windsor.

All parents were informed that the purpose of the study was to explore the association between music and language in children with WS through a sampling of affected children and a comparison group. Parents of children with WS were informed that their child would be asked to participate in two sessions (both about 2 hours) to be carried out at the University of Windsor, at the child's home, or at another location convenient to all parties. A brief description of the testing procedure was provided. In addition, parents were informed that confirmation of each participating child's diagnosis would be sought. Parents of children for the comparison group were informed that their child would participate in one session at the University of Windsor. Parents were asked to contact the primary researcher if interested in participating in the study.

The WS group was comprised of the first 19 children who met the age criteria (8-13 years) and whose mental age could be established based on Peabody Picture Vocabulary Test-Revised scores. Potential data for the comparison group were collected from all participants meeting comparison group age criteria (5-12 years) who were tested over a three month period. The actual sample was selected from the larger sample to create an equivalent comparison group to the WS group based on mental age.

For both groups, the first session was used to collect data on music and language skills. At the beginning of the session, consents and assents were reviewed and signed. The proposed procedure was described to both parent and child and the voluntary nature of the study was emphasized. The parent(s) or guardian(s) was given a questionnaire to complete. Although a few parents chose to stay in the room during the testing session, most did not.

The assessment began with a structured interview concerning the child's subjective and objective experiences of music. Following the interview, tests measuring music and verbal skills were administered in a standardized order (Tonal subtest, Sentence Memory, Auditory Closure, Verbal Fluency, Digit Span, Rhythm Subtest, Peabody Picture Vocabulary Test). When necessary, however, test order was adapted slightly to maintain the child's interest. In addition, tests were introduced in a manner calculated to engage the child's interest. For example, the Tonal and Rhythm subtests were introduced as "Mr. Gordon's tests" and the child was asked, "When is Mr. Gordon going to start?" before the music portion of the taped tests began. The Tonal subtest was always given before the Rhythm subtest and at least three verbal tasks were presented between the Tonal and Rhythm subtests.

Four of the children with WS and two of the children in the comparison group did not understand the same/different discrimination required for completing the Tonal and Rhythm subtests. For these children, the concept of 'right' for equivalent patterns and 'wrong' for different patterns was used. For each child, the practice trials (included with the Tonal and Rhythm subtests) were repeated up to five times to ensure understanding. The test was administered after perfect completion of one trial or after five repetitions. Two children in the WS and comparison groups did not achieve one perfect practice trial for the Tonal subtest. Two children in the WS group and three children in the comparison group did not achieve a perfect practice trial for the rhythm subtest.

Children who scored below chance on either music subtest were given a score of 50% correct (at chance level) for statistical analysis. No children scored below chance on the Tonal subtest. On the Rhythm subtest, three children in the WS group and one child in the comparison group scored below chance. These children scored below chance because of numerous "I don't know" or multiple responses per item.

For children with WS, standardized administration of an intelligence test occurred during the second testing session. For one child with WS, results from a recent psychological assessment (one week prior to testing) were used. One child declined WISC-III testing. During each testing session, breaks were given as required. Because of difficulties with attention, children with WS received more breaks and cues to attend than children in the comparison group. The need for breaks and cuing was most noticeable for the younger children in the WS group. Children received a gift of a small toy upon completion of each session. All participants received a letter of thanks for their efforts.

Measures

1. Wechsler Intelligence Scale For children-III (WISC-III) (Wechsler,1991).

The WISC-III is a standardized test of intelligence for children 6 to 16 years of age. Scores for Full Scale IQ, Verbal IQ (VIQ), and Performance IQ (VIQ) scores are computed based on the results of 10 standard subtests. Factor analytic studies reveal, however, that a three factor model best characterizes the subtests of the WISC-III (Sattler, 1992). Scores on these Factors are computed as standardized scores of Verbal Comprehension, Perceptual Organization, and Processing Speed. The Verbal Comprehension Factor is computed based on scores of 4 out of 6 verbal subtests (Information, Similarities, Vocabulary, and Comprehension) because the other subtests (Digit Span & Arithmetic) appear to assess additional components such as attention and concentration in addition to verbal comprehension. The Perceptual Organization Factor is computed based on scores of 4 out of 5 performance subtests (Picture Completion, Picture Arrangement, Block Design, and Object Assembly) because the 5th subtest (Coding) has a much higher loading on the Processing Speed Factor. For this study, Verbal Comprehension and Perceptual Organization scores as well as VIQ and PIQ scores were used as measures of verbal and nonverbal processing.

Psychometric properties of the test are excellent and well established (Sattler, 1992). When the WISC-III is used for mentally retarded children, reliability and validity may be limited by floor effects that are most apparent in younger children (Sattler, 1992). By 8 years of age, however, floor effects should be minimized.

WISC-III Verbal subtests

Information. The child is asked to answer questions about objects, dates, literary figures, history, and geography.

Similarities. The child is asked how objects or concepts are alike.

Arithmetic. The child is asked to verbally solve simple and complex arithmetic problems presented verbally and visually.

Vocabulary. The child is asked to define words.

Comprehension. The child is asked to explain situations or appropriate actions that relate to events familiar to most children.

Digit Span. The child is asked to repeat a series of orally presented digits ranging from 2 to 8 digits in length. During the first portion of the subtest, the child is asked to repeat the digits in the same order as the examiner. During the second portion of the subtest, the child is asked to repeat the digits in the reverse order.

WISC-III Performance subtests

Picture Completion. The child is asked to name or point to important missing details in pictures of common objects.

Coding. The child is asked to copy simple symbols paired with other symbols.

Picture Arrangement. The child is asked to arrange a series of pictures in a logical order to tell a story.

Block Design. The child is asked to reproduce geometric designs presented in three and two dimensions using coloured blocks.

Object Assembly. The child is asked to put puzzle pieces together to form common objects.

The Verbal Comprehension and Perceptual Organization Factors were used to assess verbal and nonverbal processing characteristics of the sample. The Vocabulary, Similarities, Digit Span, and Comprehension subtests (in addition to the language measures listed below) were used as verbal measures for correlation with music measures.

Other Language measures

Auditory Closure: The child is asked to blend segmented sounds of a word and produce the corresponding word. The measure has good consistency (.60) and stability (.59) (Brown, Rourke, & Cicchetti, 1989).

Sentence Memory: This test provides a measure of rote verbal memory. The child is asked to provide immediate repetition of taped sentences of increasing length. Spreen and Strauss (1991) reported developmental norms for children 6 to 13 years of age, which were merged from 4 sources. Consistency and stability (.71) are excellent (Brown et al., 1989).

Controlled Oral Word Association to category (animals): This measure of verbal fluency assesses word-finding ability and verbal output. The child is asked to name as many animals as possible within one minute. Normative data from Halperin, Healy, Zeitchik, Ludman, and Weinstein (1989) (as cited in Spreen & Strauss, 1991) will be used.

Controlled Oral Word Association to Category (musical instruments) experimental version: This unstandardized experimental measure of verbal fluency is similar to the previous mentioned test, but no time limit is given.

Music Measures

Primary Measures of Music Audiation (PMMA) (Gordon, 1980). The PMMA is a standardized test of music audiation for children in kindergarten through grade three. Audiation is defined by Gordon (1980) as that which "takes place when one hears music through recall or creativity, the sound not being physically present" (p. 107). The test consists of tonal and rhythm subtests. Composite, tonal, and rhythm percentile scores are computed based on the rhythm and tonal subtests.

Psychometric properties of the test are good. Standardization was conducted in 1978 on a sample of 873 children in kindergarten through grade three in a suburban city in New York State. Test-retest reliability ranged from .73 to .76 while split-half reliabilities ranged from .72 to .86. The test also has been used to assess music audiation in mentally retarded adults, with test-retest reliabilities at .81 or higher (Hoskins, Kvet, & Oubre, 1988). Intercorrelations between the tonal and rhythmic subtests range from .45 to .51, which indicates that the tests have less than 25 percent of variance in common and measure different constructs.

PMMA subtests

Tonal Test. The tonal test has 40 taped items. Each item consists of a pair of melodic phrases that are either the same or different. All phrases have two to five pure tones of equal length that are presented in either major or minor tonality. Two sample items and two practice items are provided. The child is asked to circle a pair of identical smiling faces if the phrases are the same or a pair of smiling and frowning faces if the items are different.

Rhythm Test. The rhythm test has 40 taped items, each consisting of two short rhythmic phrases that are either the same or different. A single pure tone is used to present the rhythmic phrases. Macro beats or tempo beats are included and are heard at a relatively low dynamic level and a different timbre than the rhythmic phrase. Two practice items are provided to familiarize participants with the task. Answer sheets are in the same format as those for the Tonal Test. Centile scores from each subtest will be converted to Z-scores to assess levels of performance.

Scale Discrimination (Peretz, 1995). This test has 30 taped items, each consisting of a pair of melodic phrases that are either the same or different. In the different condition, changes are either within the key or outside of the key of the melody. Adults and children over seven typically detect out-of-key changes better than in-key changes (Trainor & Trehub, 1992, 1994). The child is asked to circle paired smiling faces if the phrases are the same or paired smiling and frowning faces if the items are different.

Child Music Interest Interview: The interview was constructed for this study (see Appendix 1). It contains questions pertaining to the children's musical interests and knowledge, their subjective enjoyment of music, their musical activities (e.g., time spent listening to or producing music), and environmental musical factors. The interest and knowledge section contains untimed fluency measures. The child is asked to name as many songs, musical groups, and musical instruments as possible. The total number of different instruments named is used as a musical fluency score.

Parent Language and Music Questionnaire: The questionnaire was constructed for this study (see Appendix 2). It contains questions pertaining to their child's musical interests and knowledge, musical activities (e.g., time spent listening to or producing music), auditory characteristics, and environmental musical factors.

Overview of the analyses

To test Hypothesis 1 (i.e., to substantiate previously reported patterns of better verbal than visuospatial skills), standardized scores on the Verbal Comprehension and Perceptual Organization Factors of the WISC-III and the more commonly used VIQ and PIQ were compared using paired t-tests. Additional analyses examined patterns of performance across verbal tasks differing in levels of complexity.

To test the hypothesis that music skills of the WS group would be at the level of verbal skills and better than visuospatial skills (Hypothesis 2), mean age-equivalent scores on the Peabody Picture Vocabulary Test-Revised were used to provide a level of verbal functioning. These levels were used to establish mental age from which performance on the Primary Measures of Music Audiation Tonal and Rhythm subtests was evaluated. Independent samples t-tests were used to compare performance between groups. To test whether performance on music measures was better than performance on visuospatial tasks, Z-scores for the Perceptual Organization Factor and PIQ of the WISC-III were compared to estimated Z-scores (see Chapter III) on the Rhythm and Tonal subtests using paired t-tests. In addition, performance on music measures was compared to overall verbal performance (as measured by the Verbal Comprehension Factor and VIQ) using paired t-tests.

To test the hypothesis that language and music skills would be correlated (Hypothesis 3), Pearson correlations were used to measure the association between performance on music and language tasks. Although three measures of music skills were proposed originally, only two (the Tonal and Rhythm subtests of the Primary Measures of Music Audiation) were used in the final analysis. The Scale Discrimination test proved too difficult for both the comparison and WS groups and was excluded from the test battery after 20 children had been tested. None of these children (15 in the comparison group, 5 in the WS group) scored above chance on this measure.

Age, musical training, and history of otitis media and its complications were examined for associations with measures of language and music. After these potential covariates were identified, correlations between music and language performance were assessed both with and without the covariates held constant. In addition, correlations between music and language performance were assessed separately and compared across the two groups of children. For the WS group, additional analyses compared associations between music and overall verbal performance with associations between music and overall visuospatial performance.

Data from the questionnaires and interviews were used to assess the hypothesis that children with WS would exhibit an interest in music that is comparable to normal children, consistent with the notion of relatively preserved music in this population (Hypothesis 4). Data were analyzed descriptively and examined for differences between the WS and comparison groups.

CHAPTER III

RESULTS

Overall pattern of results

The first set of analyses confirmed that the previously reported pattern of better verbal than visuospatial performance in children with WS was also evident in the present sample. Means, standard deviations, and ranges of the WS group for WISC-III Factors and IQ scores and for Peabody Picture Vocabulary Test-Revised standard score equivalents are presented in Table 1. Scores are directly comparable as all have a mean of 100 and a standard deviation of 15. As predicted, a paired t-test revealed a superiority for verbal over visuospatial skills, as measured by the Verbal Comprehension and Perceptual Organization Factors of the WISC-III, $t(17)=7.422$, $p<.001$.

The Verbal Comprehension and Perceptual Organization Factors of the WISC-III were chosen as the primary measures of overall verbal and visuospatial functioning because they eliminate extraneous factors (e.g., mental arithmetic, graphomotor speed) present in the more commonly used VIQ and PIQ. Nonetheless, a comparison of the more standard VIQ and PIQ measures also confirmed that the verbal skills of the children with WS were superior to their visuospatial skills, $t(17)=6.03$, $p<.001$. Indeed, the pattern of higher verbal than nonverbal performance was so pervasive that not one child scored higher on the Perceptual Organization Factor

Table 1. Means, standard deviations, and ranges for Peabody Picture Vocabulary Test-Revised (PPVT-R), WISC-III VIQ and PIQ, and Verbal Comprehension (VC) and Perceptual Organization (PO) Factors for the WS group

Variable	N	M (X = 100) (SD = 15)	SD	Range
PPVT-R	19	77.53	18.33	48-109
VC	18	65.44	9.65	50-84
VIQ	18	61.83	10.27	46-81
PO	18	52.56	3.94	50-62
PIQ	18	50.61	4.84	45-62
FSIQ	18	52.72	7.60	40-69

than on the Verbal Comprehension Factor, or higher on PIQ than on VIQ (see Table 2). Predictably, this difference was most noticeable in children with higher Full Scale IQs. In general, differences between verbal and nonverbal functioning are less likely to be found in lower functioning children (i.e., FSIQ $\leq$ 50) because floor effects on subtests tend to obscure differences that may be present.

Because performance on the Peabody Picture Vocabulary Test-Revised (a measure of receptive vocabulary) was used to establish mental age, additional tests were conducted to confirm the superiority of the WS group on this measure of verbal skill relative to visuospatial measures. Paired *t*-tests revealed that scores on the Peabody Picture Vocabulary Test-Revised were higher than both Perceptual Organization and PIQ standard scores, $t(17)=6.43$, $p<.001$, and $t(17)=7.35$, $p<.001$, respectively.

Because the functioning of children with WS closely parallels that of children with nonverbal learning disabilities (Rourke, 1989), performance on the Peabody Picture Vocabulary Test-Revised (a measure of relatively simple verbal skills) was expected to be superior to performance on measures of more comprehensive and complex verbal skills. These hypotheses were confirmed by significant differences between Peabody Picture Vocabulary Test-Revised and both Verbal Comprehension and VIQ standard

Table 2. Individual standard scores and differences between VIQ and PIQ and Verbal Comprehension (VC) and Perceptual Organization (PO) for the WS group

Subject	VIQ	PIQ	VIQ-PIQ=	VC	PO	VC-PO=
1	81	62	19	84	62	22
2	79	47	32	77	50	27
3	75	55	20	76	54	22
4	69	57	12	75	59	16
5	69	55	14	75	57	18
6	67	52	15	73	53	20
7	66	57	9	69	59	10
8	66	46	20	69	50	19
9	60	53	7	63	51	12
10	60	49	11	63	50	13
11	58	48	10	63	50	13
12	57	46	11	61	50	11
13	54	49	5	57	51	6
14	54	49	5	57	50	7
15	52	46	6	56	50	6
16	50	48	2	54	50	4
17	50	46	4	56	50	6
18	46	46	0	50	50	0

Note. One participant declined WISC-III testing.

scores, $t(17)=6.26$, $p<.001$, and $t(17)=5.53$, $p<.001$, respectively.

Additional analyses examined further the verbal functioning of the WS group by comparing performance levels across the different verbal tasks. Such analyses revealed a hierarchy of performance that appeared to be based on the complexity of processing required (See Table 3). This pattern was generally consistent with predictions based on the previously noted parallels between the model of nonverbal learning disabilities and the neuropsychological profile associated with WS. Performance on verbal tasks appeared to cluster at four levels. At the first level, within one standard deviation from the norm, were two tests: Auditory Closure and the Controlled Oral Word Association Test. A single test, a measure of receptive vocabulary (Peabody Picture Vocabulary Test-Revised), was at the second level, one and one half standard deviations from the norm. Performance on the third cluster of tests was approximately two standard deviations below the norm and included the Digit Span, Similarities, Vocabulary, and Information subtests of the WISC-III, and the Sentence Memory Test. At the fourth level, almost three standard deviations below the norm, was a single measure, the Comprehension subtest of the WISC-III.

A repeated-measures analysis of variance confirmed that performance differed across the verbal measures for the WS

Table 3. Performance on linguistic measures: Means and standard deviations for the WS group

Measure	N	M (Z-scores)	SD
Auditory Closure	19	-0.27	1.13
Controlled Oral Word Association	18	-0.78	1.15
Peabody Picture Vocabulary Test-Revised	19	-1.50	1.22
Similarities*	18	-1.87	0.88
Digit Span*	19	-1.88	0.75
Vocabulary*	18	-1.98	0.84
Information*	18	-1.98	0.85
Sentence Memory	19	-2.25	0.98
Comprehension*	18	-2.74	0.47

* WISC-III subtest

group, $F(8,128)=20.23$, $p<.001$. Post-hoc contrasts compared performance levels between clusters (corrected for multiple tests). Significant performance decrements were revealed between all pairs of consecutive clusters, such that performance was better on Cluster 1 than on Cluster 2, better on Cluster 2 than on Cluster 3, and better on cluster 3 than on cluster 4 (see Table 4).

The next set of analyses compared performance on individual verbal measures that were administered to both groups of children (PPVT-R, Digit Span, and the three non-WISC-III verbal measures). Mean raw scores were compared to examine differences in performance at equivalent receptive vocabulary levels. Mean Z-scores were used to test for differences between groups and to compare performance levels to normative data. It should be noted, however, that mean Z-scores for the comparison group were based on only 14 children because norms were not available for the entire age range of the control group.

Based on a comparison of raw scores, performance on least complex tasks (i.e., those in Cluster 1 and 2) did not differ between groups. At higher levels of difficulty (Digit Span and Sentence Memory Test), the comparison group's performance was superior to that of the WS group (see Table 5). Performance on the Digit Span subtest was further analyzed to examine group differences on the less complex forward condition and the more complex backward condition. On both tasks, the comparison group performed significantly better than the WS group.

Table 4. Comparisons of Z-scores of the WS group on verbal tasks across 4 levels of difficulty

	Cluster 1	Cluster 2	Cluster 3
Cluster 2	$F(1,17)=13.1$		
Cluster 3	$F(1,17)=77.5$	$F(1,18)=7.4$	
Cluster 4	$F(1,17)=98.1$	$F(1,17)=27.2$	$F(1,17)=22.3$

Note. All p values (corrected for multiple comparisons) were $<.05$.

Table 5. Comparisons of performance on verbal measures between groups

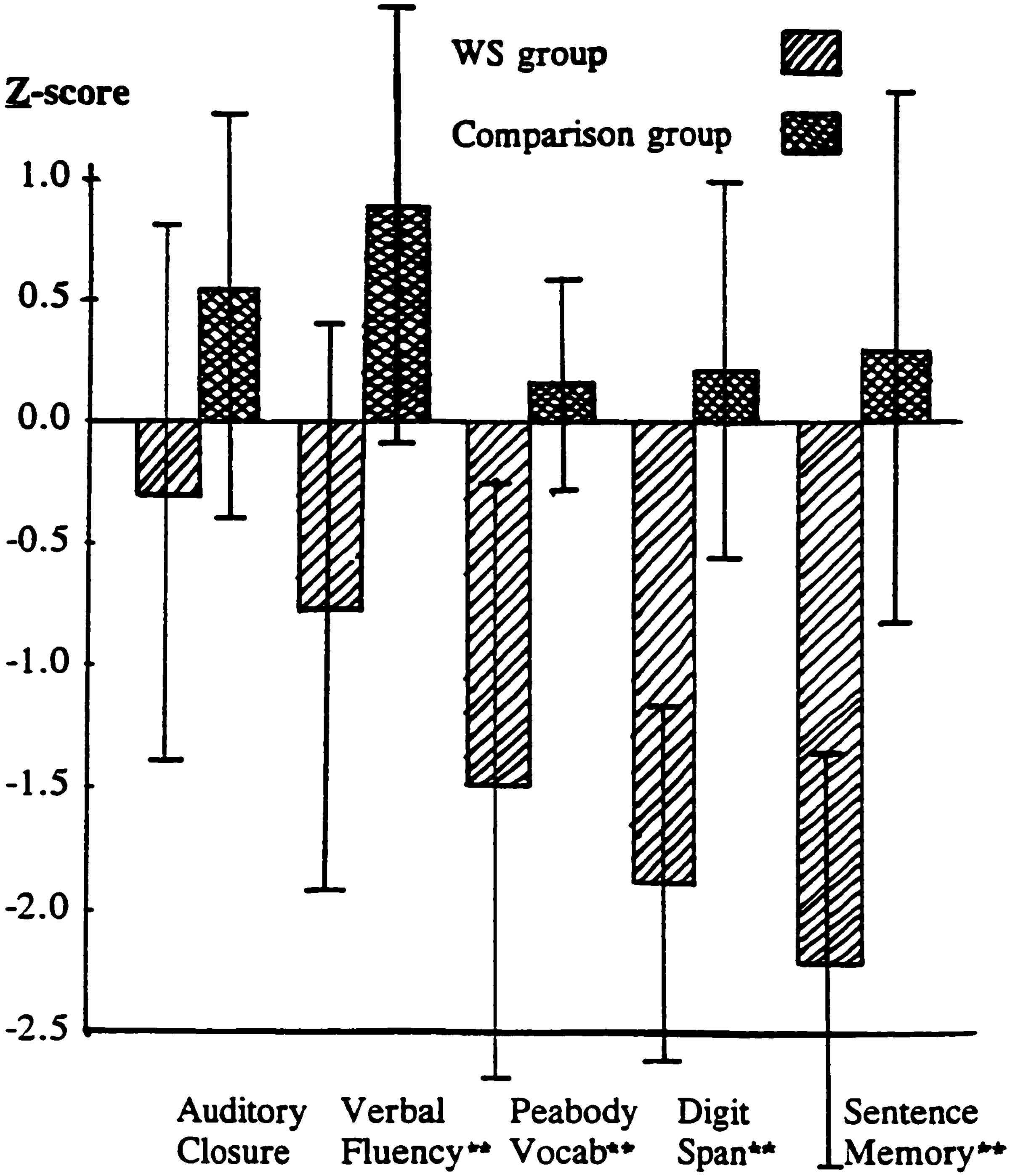
Measure	WS		Comparison		t-test
	M	SD	M	SD	
Auditory Closure	12.90	5.03	10.68	6.16	t(36)=1.21, p=.233
Controlled Oral Word Association	11.79	4.24	13.90	5.22	t(35)=1.35, p=.187
Sentence Memory	10.48	2.44	12.84	3.50	t(36)=2.42, p=.021
Digit Span	7.53	2.50	10.79	3.19	t(36)=3.51, p=.001
Forward	5.42	1.35	7.0	1.73	t(36)=3.14 p=.003
Backward	2.10	1.37	3.79	1.93	t(36)=3.10 p=.004

When normative performance was compared (i.e., using Z-scores), independent samples *t*-tests revealed significant differences between groups on all but the simplest test, Auditory Closure (see Figure 1). Although the pattern of performance for the comparison group demonstrated a trend toward better performance on the less difficult tasks, performance on all tests was within one standard deviation of the norm. Thus, the pattern of significantly decreasing performance across verbal tasks of increasing complexity was evident only for the WS group. Indeed, a repeated-measures ANOVA with one within-subject factor (verbal tasks: 4 levels) and one between-subjects factor (WS versus comparison group) revealed a significant interaction effect, $F(3,93)=7.24$, $p<.001$.

Music skills

The second set of analyses examined the WS group's performance on music measures. Scores on the Tonal and Rhythm subtests of the Primary Measures of Music Audiation were evaluated based on each individual's mental age (derived from the Peabody Picture Vocabulary Test-Revised). Mental age based Z-scores for the Tonal and Rhythm subtest scores were calculated for the 14 children in the WS group whose mental age equivalents were within the limited range of the Primary Measures of Music Audiation norms (5.5 - 9.5 years). For these 14 children, mean mental age based on raw scores on the Peabody Picture Vocabulary Test-Revised was 7 years, 5 months (1.74 standard deviations below the norm for chronological age). Mean Tonal scores

Figure 1. Z-scores for WS and comparison groups on verbal measures.



** p<.01 with Bonferroni correction for multiple comparisons.

(mental age Z-score equivalent=0.36) and Rhythm scores (mental age Z-score equivalent=-0.1) were typical for mental age based on receptive vocabulary, as predicted in Hypothesis 2. Although performance on both music subtests was consistent with mental age, a paired t-test revealed that performance on the Tonal subtest was superior to performance on the Rhythm subtest, $t(13)=2.55$, $p=.024$. Given the hypothetical prediction of better rhythmic than melodic skills in the syndrome of NLD, this finding suggests a discrepancy between the neuropsychological profiles of WS and NLD.

To further test the hypothesis that music performance would be appropriate for mental age, raw scores of the WS and control group children were compared with independent-samples t-tests. The groups did not differ on the Tonal subtest, $t(35)=.54$, $p=.596$, but the performance of children with WS was inferior to that of the comparison group on the Rhythm subtest, $t(35)=2.19$, $p=.036$. Mean scores and standard deviations for the Tonal and Rhythm subtests of the Primary Measures of Music Audiation subtests are presented in Table 6.

Hypothesis 2 stated that levels of performance on music tests for the WS group should be equivalent to levels of performance on language measures and better than performance on visuospatial measures. Explicit testing of this hypothesis proved to be complicated because age norms for the music tests did not extend high enough to include the chronological age range of the WS group. Test difficulty

Table 6. Performance on Tonal and Rhythm subtests: Group means and standard deviations

	WS	Comparison
Tonal subtest	N=18	N=19
M (raw score)	31.94	33.00
SD	5.80	6.17
Rhythm subtest	N=19	N=18
M (raw score)	25.95	30.11
SD	4.38	6.99

was an additional complicating factor; performance for the WS group could not be compared directly to a 'normal' control group (equivalent for chronological age) because such a group would perform at ceiling levels. Thus, a conceptual measure of the WS group's level of performance (e.g., an estimated Z-score) on the music subtests was derived. The following example is provided to illustrate how this was done. A hypothetical child with WS had a mental age of 8 years 4 months and a chronological age of 11 years 9 months. This child's performance on the Tonal subtest was 0.5 standard deviations above the norm for mental age, whereas performance on the Rhythm subtest was 0.2 standard deviations below the norm for mental age. This child's raw score on the Peabody Picture Vocabulary Test-Revised (from which mental age was derived) was, however, 1.67 standard deviations below the norm for chronological age. For chronological age, then, Tonal subtest performance would be relatively higher than performance on the Peabody Picture Vocabulary Test-Revised, whereas Rhythm subtest performance would be relatively lower. To create the estimated Z-scores, Z-scores for the Peabody Picture Test-Revised were added to the mental age equivalent Z-scores for each music subtest. Thus, the hypothetical child would have an estimated Z-score of -1.17 (-1.67 + .5) for chronological age on the Tonal subtest and an estimated Z-score of -1.87 (-1.67 + -.2) for chronological age on the Rhythm subtest.

Thirteen children in the WS group had both mental age equivalent Z-scores for the Tonal and Rhythm subtests and complete testing on non-verbal measures. Estimated Z-scores for level of performance on the Tonal and Rhythm subtests were compared to Z-scores for visuospatial measures (Perceptual Organization Factor and PIQ). Mean estimated Z-scores for the Tonal and Rhythm subtests were -1.17 (SD = 1.19) and -1.75 (SD = 1.08), respectively, whereas mean PIQ and Perceptual Organization Z-scores were -3.29 (SD = 0.32) and -3.16 (SD = 0.26), respectively. Paired t-tests revealed that estimated Z-scores for the Tonal subtest were significantly higher than PIQ, $t(12)=-6.7$, $p<.001$, and Perceptual Organization scores, $t(12)=6.02$, $p<.001$, and that estimated Z-scores for the Rhythm subtest were higher than PIQ, $t(12)=5.37$, $p<.001$, and Perceptual Organization scores $t(12)=4.71$, $p=.001$.

To test the parallels between relatively simple language and music skills, performance on music measures was compared to performance on complex measures of language (WISC-III VIQ and Verbal Comprehension Factor). Paired t-tests revealed that estimated Z-scores for the Tonal subtest were significantly higher than VIQ, $t(12)=5.24$, $p<.001$, and Verbal Comprehension scores, $t(12)=4.19$, $p=.001$. Estimated Z-scores for the Rhythm subtest were also significantly higher than VIQ $t(12)=4.22$, $p=.001$, and Verbal Comprehension scores $t(12)=2.83$, $p=.015$.

In sum, as predicted in Hypothesis 2, children with WS demonstrated levels of music skills that were commensurate with their level of relatively simple verbal skills. When levels of performance were compared to the control group (equivalent for Peabody Picture Vocabulary Test-Revised scores), only performance on the Tonal subtest was at expected levels. Performance on the Rhythm subtest was significantly below that of the comparison group. Nonetheless, for the WS group, overall levels of music skills were significantly better than levels of performance on visuospatial tasks and on tasks measuring complex verbal skills.

Correlations between language and music measures

The third group of analyses compared performance on language and music measures to evaluate the hypothesis that language and music skills would be correlated (Hypothesis 3). Initial analyses examined whether such correlations differed between groups; no differences were found. Thus, associations between language and music did not differ for the WS and comparison groups. To maximize statistical power, correlations were re-calculated for the groups combined (see Table 7).

All measures of language, except perseverations on the Controlled Oral Word Association Test, demonstrated moderate but significant positive simple associations with the Tonal and Rhythm subtests. A significant negative correlation between perseverations and performance on both music subtests was noted. Because increased perseverations are scored positively and are considered to be pathological, the negative correlation actually implies a positive association between language and music performance.

Associations between potential mediating variables and

Table 7. Correlations between language and music measures for groups combined [Coefficient/(Cases)/1-tailed significance]

Measure	Tonal	Rhythm
Auditory Closure	.520**	.373*
	(37)	(37)
Digit Span	.501**	.551***
	(37)	(37)
Controlled Oral Word	.466**	.489**
Association Test	(36)	(37)
Perseverations	-.606***	-.349*
	(36)	(37)
Peabody Picture		
Vocabulary Test-	.579***	.436**
Revised	(37)	(37)
Sentence Memory	.572***	.611***
	(37)	(37)

* p<.05, ** p<.01, *** p<.001

language or music skills were assessed (see Table 8). A set of multiple regression analyses confirmed that the previously reported associations between language and music were still evident when differences in age, past history of otitis media, history of ear infection in the past year, past history of hearing loss, current or past history of music lessons, months of music study, or hours per week spent listening to music were held constant.

To assess whether these correlations reflected a general association between level of performance on all tests or a more specific association, correlations between music and visuospatial skills (as measured by the Perceptual Organization Factor) were also examined for the WS group. Both Tonal and Rhythm subtest scores exhibited virtually no correlation with visuospatial abilities, rs=.040 and .044, respectively (ps were non-significant).

In sum, a moderate positive correlation was exhibited between music and language skills as predicted in Hypothesis 3. Such correlations between music and language skills were equivalent for both groups. Additionally, for the WS group, music and visuospatial skills appeared to be independent.

Musical experience and interest

The last group of analyses used responses on the Child Music Interest Interviews and the Parent Language and Music Questionnaires to evaluate the hypothesis that children with WS would exhibit an interest in music that was comparable to that of normal children. Overall, results from the Child Music Interest Interview and Parent Language and Music

Table 8. Correlations between potential covariates and language or music measures [(coefficient/(cases)/1-tailed significance]

Potential Covariates	Music and Language Measures							
	Tonal subtest	Rhythm subtest	Auditory Closure	Digit Span	Controlled Oral Word Association	Persev	Peabody Picture Vocabulary	Sentence Memory
Age	.306*	.226	.633***	.101	.431**	.079	.645***	.289*
	(37)	(37)	(38)	(38)	(37)	(37)	(38)	(38)
Current lessons	.305*	.252	.224	.294*	.300*	-.243	.228	.190
	(37)	(37)	(38)	(38)	(37)	(37)	(38)	(38)
Past lessons	-.033	-.120	.270	-.178	.146	.008	.123	-.055
	(36)	(36)	(37)	(37)	(36)	(36)	(37)	(37)
Months lessons	.293*	.393**	.187	.405**	.302*	-.153	.162	.272*
	(37)	(37)	(38)	(38)	(37)	(37)	(38)	(38)
Hours/week listening to music	-.116	.098	-.092	.005	.058	.166	-.123	-.075
	(37)	(37)	(38)	(38)	(37)	(37)	(38)	(38)
Any history or Otitis Media	-.100	-.112	.269	.035	.043	.038	.186	-.146
	(36)	(36)	(37)	(37)	(36)	(36)	(37)	(37)
Otitis Media in past year	.060	-.048	.168	.227	.117	.150	.075	.124
	(37)	(37)	(38)	(38)	(37)	(37)	(38)	(38)
History of past hearing loss	.001	.041	.030	-.012	.414**	.120	.094	-.049
	(37)	(37)	(38)	(38)	(37)	(37)	(38)	(38)
History of tubes in ears	.045	.176	.182	-.096	.180	.121	.102	-.071
	(37)	(37)	(38)	(38)	(37)	(37)	(38)	(38)

* p<.05, ** p<.01, *** p<.001

Questionnaires were similar for both groups, although the WS group demonstrated slightly more involvement and affinity for music. See Table 9 for a summary of results from questionnaire and interview data.

The musical environment was similar for both groups of children. On average, parents estimated that children spent 10 to 12 hours per week listening to music, although exposure to music varied widely. For children in the WS group, the time spent listening to music ranged from 2 to 28 hours per week; for children in the comparison group, the range was from 1 to 34 hours per week.

Although few of the participating children were involved in private music lessons at the time of testing ($N = 5$; 13%), almost all children had at least one musical instrument in their home. Indeed, only one (5%) child in the WS group and five (26%) children in the comparison group lived in homes without musical instruments.

Interestingly, the majority of children in both groups (74% of the WS group and 79% of the comparison group) had a history of creating songs. A child was considered to have created songs if either the child or the parent responded positively when asked. Although most songs created by children (both groups) were word variations to familiar tunes, many different types of song creations were reported. In the WS group, two children were reported to make up "opera" songs, two created stories in song (one through use of rhyming phrases), one created rap songs, another rock and roll, and another made up songs about feelings. In the

Table 9. <u>Group means and standard deviations for interview and questionnaire data concerning music</u>

Item	WS *M*	WS *SD*	Comparison *M*	Comparison *SD*	Test
	n		*n*		
Instrument fluency	8.84	4.78	6.26	3.30	$t(36)=1.94$
Hours per week listening	9.93	6.56	12.16	9.27	$t(36)=0.85$
Number of instruments in home	2.05	1.27	1.63	1.30	$t(36)=1.01$
Total number:					
keyboard type	19		13		
drums	4		0		
guitar	5		4		
other	17		13		
Total number homes without instruments	1		5		$X^2(1,n=38)=3.167$
Child creates songs	14		15		$X^2(1,n=38)=0.146$
Type of song:					
new words to melody	7		6		
silly	4		5		
other	8		11		
Capacity for music to make the child:					
happy	19		16		$X^2(1,n=36)=1.139$
sad	15		8		$X^2(1,n=36)=3.955$*

* $p<.05$

comparison group, two children were reported to create rhyming songs, and one child each was reported to create season songs, forest songs, relationship songs, dreaming songs, and feeling songs.

In both groups, almost all children reported that music could make them feel happy (100% of the WS group and 84% of the comparison group). One child in the WS group responded negatively when asked if music could make him happy, but his response was scored affirmatively after he explained that music could not make him happy because it made him more than happy. Children in the WS and comparison group differed on their responses when asked if music could make them sad (p=.047). Whereas 15 children (79%) in the WS group reported that music could make them sad, only 8 children (42%) in the comparison group made the same claim.

Just as children with WS were moved to a greater range of emotions by music than children in the comparison group, they were also more extravagant in rating their overall feelings for music. When asked to describe their interest in music on a scale from 1 (no interest) to 5 (love it), children in the WS group responded more enthusiastically (M = 4.63, SD=.76) than those in the comparison group (M = 3.95, SD=1.08), $t(36)$=2.26, p=.030.

When asked to name as many instruments as possible without a time limit (Controlled Oral Word Association Test -- musical instruments), children in the WS were able to name more than children in the comparison group. This finding contrasted with their lower scores on the timed

Controlled Oral Word Association Test -- animals. Although the differences between groups on both the instrument fluency measure and the Controlled Oral Word Association Test were not significant, a MANOVA revealed a significant difference between groups for performance considered jointly on both fluency measures (Wilk's Lambda=.703, $\underline{F}(2,34)=.002$).

Auditory characteristics that are thought to be related to WS or musical interest were also compared. Findings corroborated previous reports of significant hyperacusis and sound sensitivity in children with WS (see Table 10). Although unusual fearfulness toward sound was expected and found for the WS group ($\underline{N}$ = 19), a substantial number of children in the comparison group ($\underline{N}$ = 9) were also reported to have or have had an unusual fear for certain sounds. Interestingly, unusual liking for sound also distinguished the groups. Over half (63%) of the children with WS but only one child (5%) in the comparison group were reported to exhibit an unusual liking for specific sounds.

In summary, groups did not differ on hours/week spent listening to music, number of children currently taking music lessons, number of musical instruments in the home, a history of creating songs, or capacity for music to make them happy. Nonetheless, children with WS were able to name more musical instruments on an untimed fluency test. Moreover, children with WS expressed a greater interest in music and were more likely to be saddened by music. In addition, children with WS exhibited the expected distinguishing auditory characteristics of hyperacusis.

Table 10. Percentage of children in each group with a history of unusual reactions to sound or otitis media and its complications

Item	WS N	WS %	Comparison N	Comparison %	Test
History of hyperacusis	19	100%	2	10%	$X^2(1, N=38)=30.76$*
History of unusual fear of sound	19	100%	9	47%	$X^2(1, N=38)=13.57$*
History of unusual liking for sound	12	63%	1	5%	$X^2(1, N=38)=14.14$*
History of otitis media	12	63%	10	53%	$X^2(1, N=38)=0.43$
Otitis media in past year	2	10%	1	5%	$X^2(1, N=38)=0.36$
History of tubes in ears	3	16%	1	5%	$X^2(1, N=38)=1.11$
History of hearing loss	1	5%	1	5%	$X^2(1, N=38)=0.0$

* $p<.05$

Interestingly, some characteristics thought distinctive to children with WS were also noted in a significant proportion of children in the comparison group (i.e., unusual fearfulness to sound and past history of otitis media).

CHAPTER IV

DISCUSSION

Synopsis

The presumption of distinctive strength in music, together with known strength in simple language skills, for children with WS, suggests that intact auditory pattern perception is a likely basis for these skills. This hypothesis led to two of the major goals of the present study. The first was to determine the level of music skills in children with WS; there has been no prior systematic research addressing these skills. The second was to evaluate the association between music and language skills in these children.

Since the functioning of children with WS closely parallels that of children with NLD, the developmental model of NLD was used as an initial basis for predicting the music skills of children with WS. Specifically, the NLD model was used to describe the performance of children whose overall cognitive functioning was in the mentally retarded range. In addition, primary assets in basic auditory perception and rote learning (especially within the auditory domain) were assumed to encompass the domains of both music and language.

These suppositions led to four hypotheses: (1) children with WS (like those with NLD) would perform better on language than on visuospatial-visuomotor tasks; (2) music skills of children with WS would be at levels comparable to their receptive vocabulary and better than nonverbal skills; (3) music and language skills would be positively correlated; and (4) children with WS would exhibit an interest in music similar to normal children of equivalent mental age. Overall, each hypothesis was supported by the results of this study, although detailed examination of the results suggested some modifications of the hypotheses and revealed evidence of atypical auditory processing.

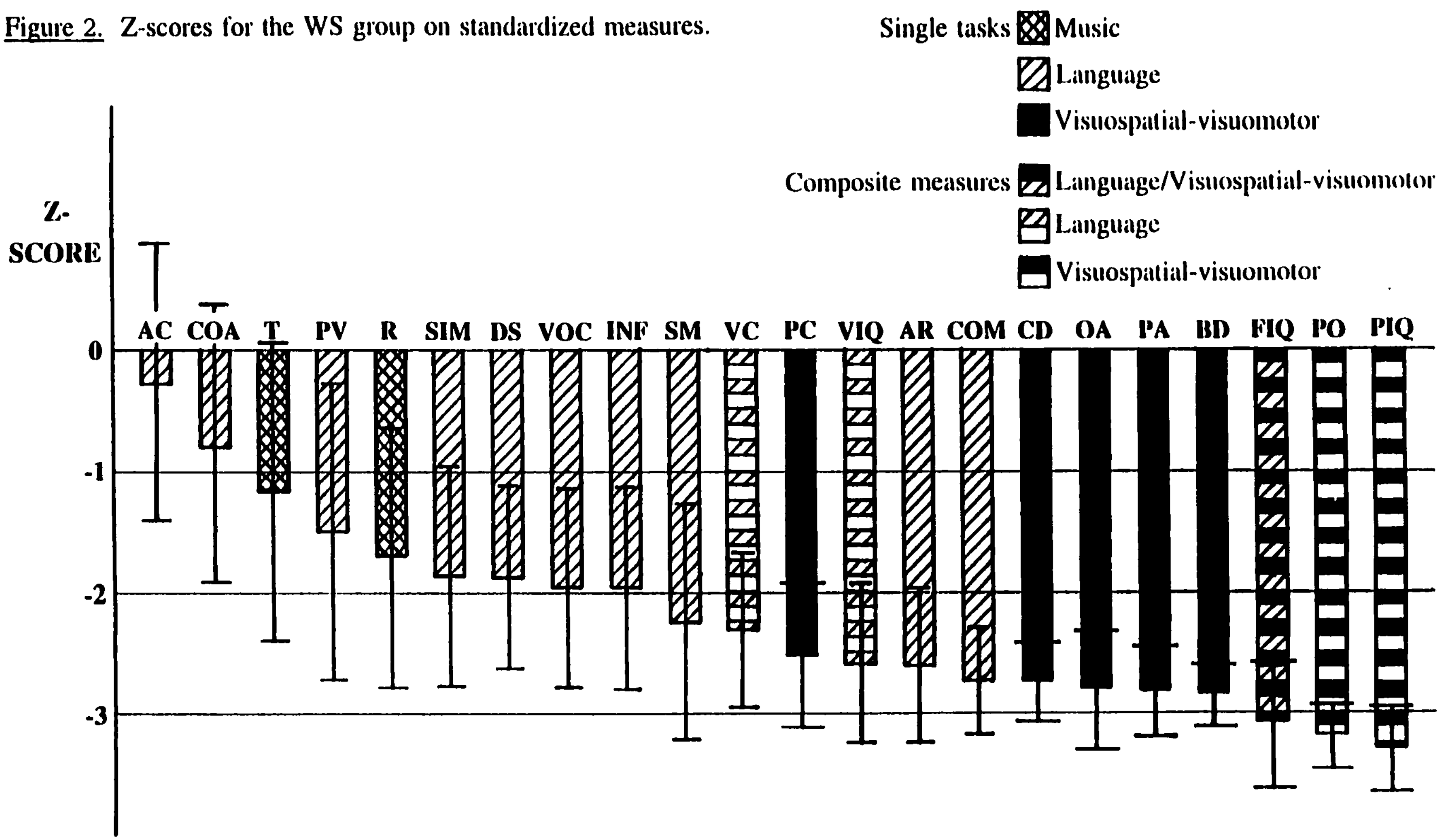
These findings are discussed in the following sections of this chapter. In the first section, the extension of the NLD model to the WS population and support for the first three hypotheses is examined through the perspective of this extended NLD model. In addition, it is suggested that auditory pattern perception is more intact than rote learning or auditory memory among children with WS. In the second section, an alternative perspective is explored; namely, the possibility that atypical auditory processing may underlie some of the music and language skills evident in children with WS. In the third section, implications of the present research for the development of music in the lives of children with WS are explored. Limitations of the present study are addressed in the fourth section. Finally, a summary of the results and possible directions for future research are presented in the last section.

Section 1: Through the NLD lens

For the WS group, the overall pattern of performance across verbal and visuospatial-visuomotor measures was consistent with expectations for children with NLD. In addition, as expected for children with NLD, performance across language measures was related to task complexity. This pattern was clearly different from the comparison group despite equivalent receptive vocabulary (see figure 1, p. 81). For the reader's reference, a summary of the findings for the WS group is presented in Figure 2. Whereas it could be argued that the contrast between groups is simply a reflection of the difference in overall intellectual functioning (mentally retarded children, in general, are presumed to have difficulty with complexity), the pattern of performance for the WS group was also different from that reported for the majority of mentally retarded children.

Mentally retarded children typically perform better on nonverbal tasks than on verbal tasks (Sattler, 1992). This distinction was clearly illustrated by a study of performance on WISC-R subtests (Muller, Dash, Matheson, & Short, 1984). From easiest to hardest, the observed pattern for the mentally retarded children was as follows: Picture Completion, Object Assembly, Comprehension, Block Design, Coding, Picture Arrangement, Arithmetic, Vocabulary, Similarities, and Information. Unfortunately, performance on equivalent WISC-III subtests has not been reported. Nonetheless, because 73% of the WISC-R items were retained

Figure 2. Z-scores for the WS group on standardized measures.



Note. **Music measures:** T=Tonal, R=Rhythm; **Language measures:** AC=Auditory Closure, COA=Controlled Oral Word Association, PV=Peabody Picture Vocabulary, SIM=Similarities, DS=Digit Span, VOC=Vocabulary, INF=Information, SM=Sentence Memory, AR=Arithmetic, COM=Comprehension; **Visuospatial-visuomotor measures:** PC=Picture Completion, CD=Coding, OA=Object Assembly, PA=Picture Arrangement, BD=Block Design; **Composite measures:** VC=Verbal Comprehension Factor, VIQ=Verbal IQ, FIQ=Full Scale IQ, PO=Perceptual Organization Factor, PIQ=Performance IQ.

in the WISC-III (Sattler, 1992), a similar pattern of findings would be expected.

Interestingly, subtests that are considered relatively simple for children with NLD were also relatively easy for children with WS, yet these were the most difficult for the mentally retarded group discussed above. Indeed, the primary assets and deficits postulated for NLD cannot explain the WISC-R performance of most mentally retarded individuals but they can explain the pattern of performance for children with WS. Thus, it appears that the NLD model can describe the performance of specific subsets of individuals from the mentally retarded population.

Although the NLD model may prove useful across a broader range of mental retardation, the pragmatics of exploring the model are complicated by the limited scope of many standardized tests. In the present study, floor effects on the WISC-III nonverbal measures precluded detailed evaluation of visuospatial-visuomotor performance. Fortunately, this finding did not mask the overall difference between performance on verbal and nonverbal measures.

Limited variability and low overall scores on the WISC-III visuospatial-visuomotor subtests indicated that individuals in the WS group performed consistently poorly on those tasks. The limited variability could also stem from the floor effects observed on these tests. Whereas 50%

(_n_=9) of the children with WS scored at or below floor levels on at least four of the five WISC-III visuospatial-visuomotor tasks, only 2 of the 18 children (11%) scored at or below floor levels on at least four of the verbal subtests. These two children also scored at floor levels on all of the visuospatial-visuomotor subtests.

For children with WS, Full Scale IQ, Performance IQ, and the Perceptual Organization Factor (WISC-III composite measures) were more than three standard deviations below the norm (in the moderately mentally retarded range), and lower than their scores on any single task. These composite scores reflect the statistical rarity of individuals who perform so poorly overall across most of the WISC-III subtests. Although scores on the WISC-III Verbal IQ and Verbal Comprehension Factor were low (in the middle to upper end of the mildly mentally retarded range), they were significantly better than Full Scale IQ and nonverbal composite scores. Moreover, there was greater variability on the composite verbal measures and on individual verbal tasks than on nonverbal measures, possibly because of the absence of floor effects on the verbal measures. Alternatively, there could be a greater range of ability in areas where children with WS perform relatively well.

These results also illustrate the danger of relying solely on IQ scores when evaluating a child's intellectual potential. Although the difference between verbal and nonverbal performance was apparent from WISC-III scores, the presence and the extent of relative strengths and weaknesses in various areas of language for the children with WS would have been missed without additional measures.

The overall pattern of results also supported the assumption that the primary auditory assets of the NLD model encompass both the domains of music and language for children with WS.

Music and language. The present study is the first to provide empirical evidence that music skills and simple language skills are areas of relative strength for these children. Although anecdotal reports sometimes suggest that music represents a greater than average talent in children with WS, the present results did not support such a claim. Instead, music skills were at a level typical for mental age based on receptive vocabulary (also a relative strength), but below that expected of normal children of equivalent chronological age.

Although performances of children with WS on the Tonal and Rhythm subtests were consistent with their mental age, scores on the Rhythm subtest were significantly lower than scores on the Tonal subtest. Moreover, only performance on the Tonal subtest was equivalent across the WS and comparison groups. On the Rhythm subtest, the WS group's performance was inferior to that of the comparison group. There are several possible explanations for these results.

The most likely concerns test order. In compliance with a standardized testing procedure, the Tonal subtest always preceded the Rhythm subtest. Unfortunately, children with WS exhibited notably increased fatigue and attention difficulties on the Rhythm subtest in comparison to the Tonal subtest. Similar problems were not noted in the comparison group. Thus, the decrease in Rhythm subtest scores observed in the WS but not the comparison group may have been caused by test order.

Another possible explanation for the difference in performance concerns the length and complexity of the Tonal and Rhythm stimuli. The Tonal stimuli were typically shorter and contained less information than the Rhythm stimuli. Whereas all but 2 Tonal stimuli contained 2 to 3 tones and lasted 1 to 2 seconds, Rhythm stimuli contained 1 to 9 tones and lasted 2 to 3 seconds. Thus, further testing is required to reconcile the observed difference between performance on Rhythm and Tonal tests or to attribute it to a single source.

Despite the difference in performance between Tonal and Rhythm subtests, both types of music skills were relative strengths among children with WS and at levels comparable to their receptive vocabulary. This finding, in addition to significant but moderate correlations between music and language tests, provides support for the hypothesis that intact auditory pattern perception underlies <u>both</u> music and language skills in children with WS. For the WS group, this hypothesis was further supported by the finding that music and visuospatial-visuomotor skills appeared to be independent.

It is also important to note that the significant, moderate correlations between language and music skills were equivalent for both groups. This finding implies that shared auditory processing may underlie music and language skills in the general population as well as in the WS population. Thus, these results present a significant challenge to modularity theories that isolate language processing from other aspects of auditory functioning.

Auditory pattern perception. As expected for children with NLD, performance across language measures was related to task complexity. For the WS group, performance on the Auditory Closure test -- theoretically the least complex language measure -- was closest to the norm. Basic auditory pattern perception is required for this task because it requires children to identify words that are presented in phonemic segments or clusters. Nonetheless, familiarity with the lexicon also affects performance. For example, when the segmented version of the word "caterpillar" was presented, some children in both the WS and comparison groups identified the word before the last two sounds, "l" and "ar" were presented. Because "caterpillar" is the only English word beginning with the sound pattern "caterpi", familiarity with the lexicon allowed the children to identify the word early. Between-group comparisons on this task were not confounded by differing vocabulary levels because both groups were equivalent for receptive vocabulary.

As hypothesized, auditory pattern perception appeared to be relatively intact in children with WS. The WS group's level of performance was both within the average range for chronological age and equivalent to the comparison group, although overall scores for the WS group were slightly higher. The equivalent performance of both groups does not result from ceiling effects because the test is adequately normed for children through age 16. As items become more difficult, words of greater length and higher vocabulary levels are introduced.

Some children in the WS group, but none in the comparison group, could accurately synthesize sound patterns even though they were unfamiliar with the word. For example, one child, after correctly responding, "tractor", asked, "what is a tractor?" Another child, after responding correctly, asked, "is that a word?" Children with WS may attend to basic phonemic patterns more than normal children, who may rely more on word familiarity. If so, this would suggest that children with WS rely on basic auditory perception skills more than normal children.

Recent reports of relative strength in phonological fluency among children with WS (Finegan et al., 1996; Mervis et al., 1996; Vicari et al., 1996; Volterra et al., 1996) lend support to the hypothesis that basic auditory pattern perception is intact in these children. Relatively good phonological processing would lead to relatively good performance on the Auditory Closure Test and phonological fluency. Good phonological processing could also help to explain some parents' amazement at how fast their children with WS "learn" to speak or sing in foreign languages. It is likely, however, that understanding of a foreign language would be very limited for these children.

The Auditory Closure Test could be modified to contain both words and non-words, which would allow for a quantitative examination of the hypothesized difference in language processing between WS and normal children. In addition, comparative testing with music measures would help to determine the distinction between general auditory pattern processing and processing that is specific to language or music. Unfortunately, tests equivalent to the Auditory Closure Test for music are not currently available. Perception of music might be tested in a similar fashion by using segmented patterns of tones or rhythms that make up familiar melodic or rhythmic patterns, such as the NBC three note melodic logo or the rhythmic pattern from "hi ho Silver (William Tell Overture)." Uncommon melodic and rhythmic patterns might be considered the equivalent of non-word patterns.

The music measures used in the present study were more complex than the Auditory Closure Test and the modifications proposed above. Because of this greater complexity, it is not surprising that performance on the music tasks was not as strong as performance on the Auditory Closure Test. Good performance on the Tonal and Rhythm subtests requires adequate short-term memory, discrimination skills, and freedom from interference, as well as basic auditory perception skills. The melodic and rhythmic patterns must be accurately perceived and held in memory so that they can be compared to a second melodic or rhythmic fragment.

Auditory memory. Auditory perception and rote learning are considered primary assets in the NLD model. For the WS group, performance levels on the Controlled Oral Word Association Test (Animal) were similar to performance levels on the Auditory Closure Test (i.e., within 1 SD of the norm for chronological age). This measure of semantic fluency is thought to rely on rote learning, or rote associations stored in long-term auditory memory. Because rote learning and basic auditory perception are considered primary assets within the NLD model, relatively good performance on this measure was expected.

Although semantic fluency on the word association task was within the average range, performance of the WS group was inferior to that of the comparison group. This finding suggests that semantic fluency and rote learning are less developed in children with WS than the auditory perception skills required for the Auditory Closure Test. Indeed, performance on measures that place even greater demands on auditory memory (i.e., Digit Span, Sentence Memory, Vocabulary, Information) provided further support for the suggestion that auditory memory is not as strong as simple auditory perception among children with WS. Nonetheless, the wide range of performance exhibited on these measures in the WS group leaves this speculation open to further investigation. Moreover, it is difficult to define and rank the cognitive demands of the various tests without resorting to circular reasoning (i.e., poorer performance = greater difficulty).

The difficulty in determining test complexity can be highlighted by referring to the Digit Span subtest of the WISC-III. The digits backward component of the Digit Span subtest is undeniably complex; the child is asked to repeat a sequence of digits reversing the original order of presentation. On the digits forward component, however, the child is simply asked to repeat what he or she hears.

In the case of digits forward, the child hears numbers at one-second intervals. This component of the Digit Span subtest may appear comparable to the Auditory Closure Test; for both tests, the child is asked to repeat what he or she has heard in sequence (although separation between digits is slightly longer than separation between phonemes). Despite this similarity, the performance of the WS group was inferior to that of the comparison group on the forward component of the Digit Span subtest. By contrast, performance of both groups was equivalent on the Auditory Closure Test.

There are numerous possible reasons for the WS group's poor performance. For example, children with WS may be able to "chunk" phonemic patterns into larger, familiar units (i.e., words) but unable to "chunk" numbers into larger, unfamiliar units (i.e., strings of numbers). Alternatively, children with WS may actually process information on both tests similarly but normal children may be more able to "chunk" numbers.

The Digit Span subtest was discussed in detail to illustrate some of the difficulties associated with understanding the pattern of performance across measures that was observed in children with WS. Although the overall pattern of results suggests that performance decreases as auditory tasks become more complex, a comprehensive understanding of this pattern requires further research.

Section 2: Atypical auditory processing

Although the present results support the conceptual extension of the NLD model, they do not eliminate the possibility of competing or coinciding alternative hypotheses. In the previous section, the possibility was raised that children with WS rely on basic phonemic processing more than normal children. As such, children with WS may process auditory patterns in an atypical manner. Additional evidence for atypical auditory processing in individuals with WS was found in an earlier study that reported atypical evoked auditory event-related potentials from the temporal cortex and a lack of normal hemispheric asymmetry in response to auditory stimulation (Bellugi et al., 1992). Recently, exaggerated left-side asymmetry of the planum temporale (associated with language and music processing) has also been reported in some individuals with WS (Bellugi et al., 1996).

The most conspicuous evidence for atypical auditory processing in children with WS comes from the overwhelming prevalence of hyperacusis. In the present study, auditory characteristics thought to be related to WS and musical interest were surveyed. The findings corroborated previous reports of significant hyperacusis and sensitivity to sound in children with WS. Based on parental reports, <u>all</u> children in the WS group exhibited evidence of hyperacusis throughout their lives, whereas only two children in the comparison group exhibited evidence of hyperacusis at any time during their lives. Moreover, only one of the comparison children exhibited over-sensitivity to sound at the time of testing. Interestingly, unusual liking for specific sounds also distinguished the groups. Almost two thirds of the WS group (63%, n=12) but only 5% (n=1) of the comparison group were reported to exhibit unusual liking for specific sounds.

Although not questioned specifically, many children in the WS group (n=7) were described by their parents as having a love/hate relationship with certain sounds. Intense emotional reactions to music were also reported for children in the WS group. For two of the children with WS, a specific love/hate relationship with music was described. For seven children, an unusual love and fascination with music were reported. For four children, unusual negative reactions to music were described. As babies, these children were reported to scream or cry uncontrollably when they heard lullabies or slow "relaxing" music. Although these reactions diminished with time for three of these children, one child continued to react in the same negative manner at the time of testing.

Based on self-report, children with WS demonstrated greater emotional responsiveness to music than did the control children. A large majority of children with WS could be moved to both happiness and sadness by music (79%), whereas less than half of the children (42%) in the comparison group reported similar emotional responses. Moreover, children with WS were able to report the specific characteristics of songs or music that made them happy or sad. Interestingly, descriptions of music that could induce sadness were split between lyrical and musical properties. For example, one child reported that she became sad when she heard songs with word such as "don't leave me." Another child reported that he became sad when the music "sound sad, like violins crying sound."

When the interest and emotional responsiveness to music demonstrated by children with WS are considered in conjunction with their hyperacusis, this suggests that some components of their emotional responsivity are linked to atypical auditory processing. Such a link could help to explain the comparatively high level of interest and emotional responsivity to music discovered in the WS group. If so, musical skills may develop along a pathway that is motivated by fascination and interest and assisted by intact auditory perception. These proposed links between hyperacusis, emotional responses, and musical development have implications for musical training, which are considered in the next section.

Section 3: Music in children with WS

The WS group demonstrated relatively intact abilities on music tests and high levels of interest and emotional response to music. Performance on the two fluency measures also suggested relatively high levels of interest in music. Whereas children with WS scored higher on musical instrument fluency than on animal fluency, children in the comparison group exhibited the opposite pattern.

Interest and emotional response toward music were also evident in spontaneous play during testing sessions. A small keyboard was available for entertainment during breaks. All children in the WS group were enthusiastic about using the keyboard during breaks and spontaneously explored sound and rhythm patterns. In addition, some children created songs while accompanying themselves on the keyboard. Spontaneity of song-making was inferred from the lyrics (e.g., "and I can be crazy ... until my mom gets back."). Such creativity was unexpected because of the difficulty with novelty reported for children with NLD. Based on the extended NLD model, it was predicted that children with WS would be musically less creative than normal children. Children with WS, however, appeared to be equivalent to the comparison group when reports of their history of making up songs were compared. Thus, if we define one facet of novelty as making up songs, difficulty with this aspect of novelty was not apparent for the WS group.

The enthusiasm and emotional responsivity to music demonstrated by children in the WS group combined with their relatively intact music abilities suggests that purposeful development of musical skills could help to enrich the lives of these children. Nonetheless, physical and cognitive limitations of children with WS would impede skill development in many areas.

For example, difficulties with fine motor coordination would limit the choices of instrument. Because articulation skills are intact, singing might be an excellent route for the development of musical skill. Stringed instruments generally require high levels of fine motor control to produce even the simplest recognizable song and would be particularly poor choices for instrumental instruction. Guitars, however, require fewer fine motor skills (fretting aids finger placement), and have been a successful choice of instrument for some children with WS when alternative tunings and chord bars were used (National Williams Syndrome Association, 1997). Additional instruments reported to be played successfully by children with WS include keyboards, pianos, drums, harmonicas, and trombones.

Because most children with WS are mentally retarded and because their cognitive functioning parallels that of children with NLD, music instruction would be most useful if it involved simple tasks, imitation, and an abundance of repetition (within the auditory modality). Visuospatial-visuomotor impairments and difficulties with complexity are likely to make reading music an unrealistic goal. The fatigue and attentional difficulties noted during testing suggest that teaching sessions should be kept short with breaks provided as necessary. Minimizing distractions and establishing a predictable sequence of events during the music lesson should help to focus and maintain the child's attention.

Findings of creativity and emotional responsivity toward music in children with WS provide additional resources for musical development. Challenging a child to create a song using a specific sound, feeling, or technique may help to motivate musical growth. In addition, attention to sound quality and the emotional aspects of music may help the child to develop musical expression. Because sound preferences, aversions, and emotional responses to music were quite personalized, these suggestions for teaching should be individualized to help each child gain the most from his or her musical experiences.

Section 4. Limitations

Because of the primary goal to evaluate the level of music skills among children with WS, it was necessary to use standardized measures for music skills. Only a very restricted range of standardized measures, however, is appropriate for assessing musically untrained children. This drawback, therefore, imposed boundaries on this study, and only basic melodic and rhythmic discrimination skills were assessed. More complex melodic, rhythmic, and musical memory skills could not be assessed through currently available measures. Thus, an overall pattern of performance for a wide range of musical skills remains to be determined.

Two limitations involving the administration of the music tests were also noted. The first concerned the standardized testing order for the subtests of the Primary Measures of Music Audiation (i.e., administration of the Tonal subtest before administration of the Rhythm subtest). For the WS group, this order of administration consistently appeared to result in attentional lapses and fatigue during the Rhythm subtest, which may have lowered scores. Similar order effects were not observed in the comparison group. In the future, consideration should be given to administering test in counterbalanced order, or in separate sessions.

In addition, all children received additional (non-standardized) practice trials when necessary to ensure understanding of the task. Repeated practice trials may have influenced results by inducing practice or fatigue effects. Children with WS and the youngest children in the comparison group, however, appeared to gain basic understanding of the task necessary for completing the test through this adaptation of test administration. Although between-group comparisons were not affected by this adaptation, comparison with normative data may be less valid.

In a small clinical study such as this, sample size and subject selection are always basic limitations. Both factors limit generalization of the results. A replication of the present study with a larger sample that extends the age range of the individuals assessed would be required before the findings can be generalized across the life-span. Nonetheless, limiting the age range of the WS group helped to maximize generalizability of results within that range.

Self-selection of subjects was potentially a more serious limitation because the decision to participate in the study might have depended upon the child's interest in music. The possibility that this was an especially musical subgroup from the WS population could not be assessed. The WS and comparison groups, however, were equivalent in musical background and environment. Since few children in either group had standard music training, "formalized" musicality does not appear to have determined interest in participating. Indeed, the possibility of self-selection based on musical interest would apply to both groups and could not be eliminated without random sampling, which is virtually impossible.

Section 5. Summary and implications for future research

The present study provides the first empirical evidence of relatively intact musical abilities among children with WS. Within the limited range of music skills tested, the performance of children with WS was substantially better than one would predict based on their Full Scale, Verbal, or Performance IQs. The music skills of children with WS were commensurate with their relatively strong receptive vocabulary (a relatively simple language skill). In addition, significant but moderate correlations between language and music skills were found for both groups. This finding suggests that simpler aspects of language and music skills are subserved by a common mechanism that is used to process auditory patterns in the general populations as well as in the WS population. Nonetheless, continued research is required to replicate the results and to explore and expand upon the present findings.

The major finding of the present study was that a quantitative measure of music skills revealed that children with WS are relatively "musical." This finding was based on the evaluation of a restricted range of musical skills within a limited age range. Sampling a greater breadth of musical skills in addition to evaluating music skills across the life span would help to substantiate and broaden the initial findings of this study. In addition, because music appears to be an area that holds promise for enriching the lives of these children, studies of natural musical development and effective music-teaching methods would provide welcome, practical information for parents and teachers.

Moreover, further exploration of predictions for music and language based on the extended NLD model would be helpful to determine both the extent of the "fit" of the model and possible modifications for the WS population. The results of the present study suggest that basic auditory processing is more intact among children with WS than rote learning or auditory memory skills. Further investigation of auditory pattern perception, rote learning, and memory would help to provide a more comprehensive understanding of auditory processing in children with WS. In fact, more complete testing of the language and music skills of this population could help to further our understanding of auditory processing in general.

The results of the present study also raise the possibility that auditory processing among children with WS might be atypical when compared to the general population. Children with WS may exhibit a greater than normal reliance on phonemic processing, which, in turn, could lead to better or equivalent phonological fluency compared to semantic fluency. Although phonological and semantic fluency are tested in a similar fashion, the requirement for sound associations rather than semantic associations makes phonological fluency a more novel and difficult task for normal children. The comparative strengths of phonological fluency and semantic fluency in children with WS have yet to be compared.

The results of the present study also suggest that some aspects of music perception among children with WS appear atypical when compared to the general population. Children with WS, for example, exhibit a rather intense interest in music that is accompanied by strong affective responses.

Spontaneous parental comments suggest that this peculiar affinity for music may be related to the hyperacusis that is observed among these children. Systematic investigation of this potential association should be one goal of future research. Because the love/hate relationship with sound or music was reported to be evident even during infancy, investigation during infancy and early childhood would be particularly informative. In addition, it would be of value to investigate the relation between emotional responses to music and level of music skills among children with WS.

Case reports of absolute pitch among children with WS, exaggerated leftward asymmetry of the planum temporale, and overall evidence of atypical auditory processing in this population suggest that systematic study of the prevalence of absolute pitch is also warranted. Because these children are generally unfamiliar with note names, alternatives to current methods of testing absolute pitch would be required. In addition, because so many disparate pieces of evidence for atypical auditory processing have been reported, it would be beneficial to investigate the possibility of a general pattern of atypical auditory processing within this population.

In conclusion, the present study represents an investigation of both a relatively well known and a new area of auditory processing (i.e., language and music, respectively) in children with WS. The findings provide evidence of relative strength in music, as well as support for the hypothesis that intact, general auditory processing abilities may underlie the music and language skills observed among these children. Although the majority of the findings of this study were consistent with predictions based on the extended NLD model, some findings, especially those associated with auditory processing of music and atypical auditory processing suggest exciting new avenues for future research of WS.

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APPENDIX A

Music Interest Interview

Music Interest Interview

Child's name:________________ **Date of Birth:__________**

Sex:______ **Age:________** **Participant #:__________**

Parent or Guardian's name(s)____________________________

Date:______________

1. Do you like music?

On a scale from 1 to 5
(1=no interest, 2=a little interest, 3=moderate liking, 4=like it a lot, 5=love it), how would you rate your feelings about music?

1___ 2___ 3___ 4___ 5___

2. What is it that you like the most about music?

3. What music do you like to listen to?

4. When you listen to music, is it mostly music that you chose?

Yes______ No______

If you don't listen to music that you choose then do you listen to music that your brother or sister or parents chose?

Yes______ No______

Or do you choose music for yourself to listen to just as often as your brother or sister or parents choose music for you to listen to? Yes______ No______

5. Do you listen to music every day? Every week? Prompt: When do you listen to music? In the morning? After school? After dinner? Before bed? (Try to get estimate of hours/day or week.)

6. Tell me some songs that you like.

Now, tell me, do you know of any other songs? Yes_____ No_____
What are they? (Prompt: Could you tell me the names of these songs?)

Do you know of any more songs? Yes_____ No_____
What are they? (Prompt: Do you know any holiday song? Do you know any school songs? Could you tell me the names of those songs?)

Total # of songs_______

7. Tell me some music groups that you like.

Now, tell me, do you know of any other music groups? Yes_____ No_____
What are they? (Prompt: Could you tell me their names?)

Do you know any more music groups? Yes_____ No_____
What are they? (Prompt: Can you tell me their names?)

Total # of music groups_______

8. Tell me some instruments that you like.

Now, tell me, do you know of any other instruments? Yes_____ No_____
What are they? (Prompt: Could you tell me their names?)

Do you know of any more instruments? Yes_____ No_____
What are they? (Prompt: Could you tell me their names?)

Total # of instruments______

9. Do you like to sing? Yes_____ No_____

When do you sing?

What do you sing?

10. How did you learn the songs you sing?

11. Is it easier for you to learn the words or the melody of a song?

Or is it just as easy for you to learn the words of a song as to learn the melody?

12. Did you ever make up songs? Yes_____ No_____

What kind of songs do you make up?

13. Do you or did you ever sing yourself to sleep? Yes_____ No_____

Now? Yes_____ No_____

14. Now for a fun part. Will you sing a song for me? Yes_____ No_____

I'd really like it if you could. Can I tape record it? Yes_____ If yes, begin taping. If a no, then skip to the next question.

First, I'd like you to sing Happy Birthday for me. You can sing it to anyone you want. (Promt, your mom, etc.)

When finished, say: That was great. Now would you sing me another song? You can sing any song you like.

When finished, say: Thank you. That was great. Now, would you sing Happy Birthday for me without any words? (If the child does not understand, demonstrate by singing La La on a single repeated tone.)

When the taping is finished, thank and compliment the child for his or her efforts.

Now I have a few more questions to ask you about music.

15. Have you ever had music lessons? Yes_____ No_____

When did you have music lessons?

Do you have them now?

How long did you have (have you had) music lessons for? (Months? Years?)

What type of lessons were (are) they?

If they were instrumental lessons, what type(s) of instrument did you learn to play? Did you learn to play any other instruments? What instrument(s)?

How much did (do) you practice? Hours/day or week?

Did (do) you like to practice?
On a scale from 1 to 5, how much did you like to practice?
(1=not at all, 2=a little interest, 3=moderate liking, 4=like it a lot, 5=love it)

1___ 2___ 3___ 4___ 5___

Did (or do) you take lessons in a group or by yourself?

Group_____ Individual_____

16. How does music make you feel? ________________________________

What type of music makes you feel ______________________________?

Can music ever make you feel happy? Yes_____ No_____
What type of music makes you feel happy?

Can music ever make you feel sad? Yes_____ No_____
What type of music makes you feel sad?

17. Do you want to tell me anything special about music that I have not asked you about?

APPENDIX B

Parent Language and Music Questionnaire

Parent Language and Music Questionnaire

Child's name:____________________________ **Date of Birth:__________**

Sex:______ **Age:________** **Participant #:__________**

Parent or Guardian's name(s)____________________________________

Date:______________

The following questions concern your child's interests in music. Although you may not be able to answer all of these questions, your answers will help us to understand your child's auditory functioning. Please feel free to add any comments you may have at any place in this questionnaire. Thank you very much for your time and your effort.

1. Does your child like music?
   On a scale from 1 to 5 (1=no interest, 2=a little interest, 3=moderate liking, 4=like it a lot, 5=love it), how would you rate your child's interest and feeling for music?

   1___ 2___ 3___ 4___ 5___

2. What does your child like most about music?

3. What type of music does your child listèn to? Please list.

4. Does your child chose most of the music he or she listens to?

   Yes______ No______

   If not do you, or someone else in your family chose most of the music your child listens to? Yes______ No______

   Or would you say, it is a 50/50 split? Yes______ No______

5. When does your child listen to music? (In the morning? After school? After dinner? Before bed?)

6. How many hours would you estimate your child listens to music each day or each week?

Hours/day________________ Hours/week________________

7. What music groups does your child know? Please name them.

8. What instruments does your child know by sound or by sight? Please name them.

9. What songs does your child know (but does not necessarily sing)? Please name them. (Holiday and School songs too.)

10. Does your child like to sing? Yes_____ No_____
On a scale from 1 to 5 (1=no interest, 2=a little interest, 3=moderate liking, 4=like it a lot, 5=love it), how much does your child like to sing?

When does he or she sing?

What songs does he or she sing? Please list them.

11. How did your child learn the songs he or she sings?

12. Does your child find it easier to learn the words or the melody of a song?

The words______ The melody______

Or does he or she learn both together?______

Or, I do not know.__________

13. Does your child ever make up songs? Yes_____ No_____

What sort of songs does he or she make up?

14. Does your child sing himself or herself to sleep? Yes_____ No_____

Did your child ever sing himself or herself to sleep?

Yes_____ If yes, at what age? ____________ No_____

Does your child listen to music to go to sleep? Yes_____ No_____

What type of music?

Did your child ever listen to music to go to sleep?

Yes_____ If yes, at what age? ____________ No_____

What type of music?

Please continue on the next page.

15. Does someone in your home currently take music lessons?

Yes______ No______

If yes, for how long? _________ years or months (please indicate)

Type of lesson.

Piano_________ Other instrument (please specify type of instrument)_____________

Voice ___________

Who? participating child__________ other child (children)__________

self_________ spouse_________ other_________

16. Did someone in the home take music lessons in the past?

Yes______ No______

If yes, when? __________ and for long? _________ years or months (please indicate)

Type of lesson.

Piano_________ Other instrument (please specify type of instrument) ________________

Voice ___________

Who? Son_________ daughter_________ participating child__________

self_________ spouse_________ other_________

17. Does anyone (or has anyone) in your home sing in a choir?

Yes______ No______

If yes, when? __________ and for long? _________ years or months (please indicate)

Who? participating child__________ other child (children)

self_________ spouse_________ other_________

18. If your participating child has ever taken music lessons, how long did your child practice every day or week? (Please indicate in hours/day or week)

On a scale of 1 to 5 how much did or does your child like to practice? (1=not at all, 2=a little interest, 3=moderate liking, 4=like it a lot, 5=love it)

1__ 2__ 3__ 4__ 5__

Did (or does) your participating child take lessons in a group or individually?

Group______ Individual______

19. Is there an instrument in your home? Yes______ No______

Piano_______ Drums_______ Guitar_______

Other (please list)__________________

20. Can you tell us anything special about your child and music that we have not asked about, but should have.

<u>Unusual auditory perception and misc.</u>

21. Is your child, or has your child ever been unusually sensitive to sound? Describe. (e.g. what, when, how did you know?....)

22. Does your child, or has your child ever become frightened or upset by certain sounds? Describe. (e.g. what sounds, when, what does your child do?...)

23. Does your child, or has your child ever expressed an unusual liking for any sounds? Describe. (e.g. what sounds, when)

24. Does your child have, or has your child had otitis media (inner ear infections)? When? How often? How was it treated?

25. Has your child ever had tubes put in his or her ears?

When?

Does he or she currently have tubes in his or her ears?

26. Has your child experienced any permanent hearing loss that you are aware of?

Thank you very much for your help.

VITA AUCTORIS

Audrey Jean Don

1954 -	Born in Akron, Ohio, U.S.A.
1971 -	Graduated from Los Altos High School, Los Altos, California, U.S.A.
1990 -	Granted degree of Bachelor of Fine Arts from the Columbus College of Arts and Design, Columbus, Ohio, U.S.A.
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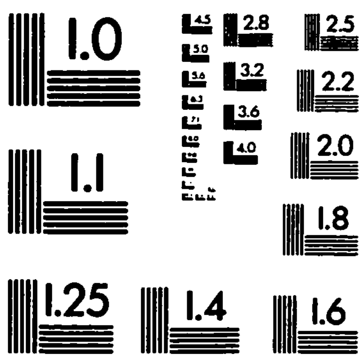
IMAGE EVALUATION TEST TARGET (QA-3)



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