Memory functioning in individuals with Williams syndrome.

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Memory Functioning in Individuals with Williams Syndrome

by

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B.Sc. Houghton College, 1987
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A dissertation
Submitted to the Faculty of Graduate Studies
Through the Department of Psychology
In Partial Fulfillment
Of the Requirements for the Degree
of Doctor of Philosophy at
the University of Windsor

Windsor, Ontario, Canada, 1997
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ABSTRACT

The aims of this study were twofold: 1. to explore theoretical links between the neuropsychological characteristics of individuals with Williams syndrome (WS) and individuals with the Nonverbal Learning Disabilities syndrome (NLD); 2. to assess the memory functioning of individuals with WS. To this end, a sample of 13 individuals with WS, ranging in age from 7 to 25 years, were administered a comprehensive neuropsychological test battery. The result of this investigation supported the hypothesis regarding similarities between the neuropsychological profiles of individuals with WS and NLD. In addition, the verbal memory functioning of the WS sample was better developed than their nonverbal memory functioning. A tendency to rely on rote learning strategies rather than on strategies that emphasize organizational skills was also evident on the California Verbal Learning Test. These findings were also expected and in support of hypotheses. Expected differences in memory functioning relative to overall verbal and nonverbal functioning (i.e., VIQ/PIQ) were not found. On the basis of similarities in neuropsychological assets and deficits, the relevance of treatment recommendations for individuals with NLD for individuals with WS is discussed. Finally, suggestions for future research were offered.
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Dr. Glenn Larrabee was kind enough to respond to a telephone call from a graduate student who wished to use his test (the CVMT) in a dissertation project and donated a copy of it for this purpose.
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Finally, I extend my gratitude to my family and friends for their support over the years. To my parents, siblings, and many aunts and uncles who knew that "I would make it," ... I MADE IT!! I hesitate to mention the names of friends and colleagues, as some may not be remembered, but I rest assured that all of you will "feel thanked and appreciated." "Toronto" Tim has been a friend who has seen me through all stages of my academic career, and has shared the drive to the Keys for several much-needed vacations. "Sarnia" Tim has always had a coffee ready for the late Sports Desk, been eager to see Geddy and the boys for another much-needed break, and been attentive during my ranting about the numerous activities that are a part of the programme. More recently, my friends and co-workers at "Fords," especially Sandy, June, and Janet, have been very supportive of my efforts. And a special word to Simone, who has proven to be the best friend I ever had! We only came together at the end of our programme, but the timing of it all couldn't have been better. I look forward to discovering, and rediscovering, many things with you!
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Chapter 1

INTRODUCTION

The purposes of the present study are two. The first is to build upon theoretical links made (Anderson & Rourke, 1995) between the neuropsychological profile of individuals with Williams Syndrome (WS) and predictions that result from the Nonverbal Learning Disabilities model (Rourke, 1989). The second is to more fully explicate the memory functioning of children and adolescents with Williams Syndrome (WS). As will be explained in the following pages, Williams Syndrome has only recently received the attention of psychology, and very recently, the attention of neuropsychology. At present, there is a limited amount of information concerning the functioning of these individuals, with that devoted to memory functioning being even less than other areas of functioning.

The available information on WS will be reviewed, followed by that part of the literature relevant to neuropsychological functioning. The theoretical relation of WS to the white-matter model developed by Rourke (1982, 1987, 1988, 1989) will also be reviewed, as it is from this relation that hypotheses concerning the memory functioning of individuals with WS will emerge. Finally, a very brief review of the memory literature relevant to the WS and NLD populations and descriptions of measures used will be covered.

History of Williams Syndrome

Williams Syndrome (WS) is a rare disorder that has been identified several times by researchers approaching it from different perspectives. For example, independent teams of endocrinologists published a number of papers in 1952 describing a disorder that was named "idiopathic infantile hypercalcaemia" (Fanconi, Girardet, Schlesinger, Butler, & Black, 1952; Lightwood, 1952; Payne,
1952). At this point, two types of the disorder were delineated. The milder, or Lightwood-type, of idiopathic infantile hypercalcaemia presented with symptoms such as anorexia, vomiting, loss of weight, constipation, and colic (Lightwood, 1952). These symptoms were thought to be caused by the hypercalcaemia. In accordance with this theory, a modification of the infant's diet was recommended, in which the intake of calcium and vitamin D were limited. These steps usually resulted in the cessation of symptoms.

The severe, or Fanconi-type, of idiopathic infantile hypercalcaemia (Fanconi et al., 1952) also presented with hypercalcaemic symptoms (i.e., those of the Lightwood-type) that were mitigated with dietary modifications. In addition to these symptoms, however, more serious symptoms were noted, including renal impairment, osteosclerosis (i.e., hardening of bones with increased heaviness), stenoses of major arteries, and moderate to severe mental retardation (Arnold, Yule, & Martin, 1985; Udwin, Yule, & Martin, 1986). The early researchers also made reference to the facial characteristics of these individuals, shortly thereafter termed "elfin facies" (Lowe, Henderson, Park, & McGreal, 1954). This combination of characteristics includes a medial eyebrow flare (i.e., eyebrow hair growth pattern toward the nose), short palpebral fissures (i.e., the opening between the eyelids), either lacy or stellate (i.e., starlike) patterns in the iris, a depressed nasal bridge with anteverted nares, a long, smooth philtrum, and thick lips with an open mouth posture. The ears have also been described as lowset, but Joseph and Parrott (1958) contended that the head posture (extended) could lead to this impression.

In 1961, a team of cardiologists from New Zealand (Williams, Barratt-Boyes, & Lowe, 1961) described a syndrome characterized by supravalvular aortic stenosis. On the basis of a number of features of one patient, including
facial characteristics and deficient mental abilities, Williams correctly predicted the presence of supravalvular aortic stenosis in three additional patients. A.G. Beuren, another cardiologist, also reported several cases of the syndrome, resulting in periodic use of the identifier "Williams-Beuren Syndrome" (Beuren, Apitz, & Harmjanz, 1962; Beuren, Schulze, Eberle, Harmjanz, & Apitz, 1964).

Black and Bonham-Carter (1963) used the phrase "elfin facies" in conjunction with WS when they emphasized the association between aortic stenosis and the facies characteristic of Fanconi-type idiopathic infantile hypercalcaemia. Garcia, Friedman, Kaback, and Rowe (1964) provided the first definitive link between the two disorders, presenting a case to "establish this association as a clinically recognizable syndrome" (p. 117). Martin, Snodgrass, and Cohen (1984) also compared the clinical features of Fanconi-type idiopathic infantile hypercalcaemia and WS. The results of this investigation indicated that there is most likely a common genetic abnormality. However, hypercalcaemia, while being linked to the underlying disorder, is seen as being "expressed with variable frequency and penetration" (Udwin, 1990) p. 129). Jones and Smith (1975) also reported that hypercalcaemia, if present, is of a transient nature.

**Epidemiology**

As mentioned, WS is a rare disorder, although perhaps not as rare as previously thought (Martin et al., 1984). Fraser, Kidd, Kooh, and Paunier (1966), based on data published by the British Paediatric Association Committee on Hypercalcaemia, found that for all forms of hypercalcaemia, there was a prevalence rate of 1 per 20,000 live births. When only Fanconi-type idiopathic infantile hypercalcaemia was considered, the prevalence rate decreased to 1 per 100,000 to 200,000 live births. These authors noted that similar statistics were not available in North America at that time. However, on the basis of available
medical records and knowledge of symptoms of the disorder (e.g., supravalvular aortic stenosis and confirmed hypercalcaemia), an estimate of 1 per 150,000 live births was reached. This is most likely an underestimate, as it is commonly accepted that hypercalcaemia, if present in WS, is transient (Jones & Smith, 1975). Therefore, if the testing for such a condition were not performed at the proper time, such a child would not be considered to have the disorder.

Some investigators have (e.g., Martin et al., 1984) reported that the incidence of idiopathic infantile hypercalcaemia in Great Britain is approximately 1 per 47,000 live births, while Greenberg (1989, 1990) estimated it to be between 1 in 20,000 and 1 in 50,000 live births. The increase in prevalence of this disorder relative to previous years was found to be due, at least in part, to the efforts of groups such as the Infantile Hypercalcaemia Foundation (UK). Beuren and colleagues (Beuren et al., 1964), discussing the increase in the early 1960s, credited the publication of more detailed studies on this disorder. Feature articles and special issues on WS in a number of journals (e.g., American Journal of Medical Genetics, Supplement 6, 1990; Burn, 1986; Shepherd & Goel, 1990) continue to play a role in the increasing awareness of WS.

Genetics

Early in the history of WS, researchers considered possible genetic causes. At the simplest level, Beuren (Beuren et al., 1964) reported that there were 46 chromosomes, and that no chromosome seemed abnormal in structure or length.

Teams of researchers have also examined the possibility of a simple genetic defect resulting in the hypercalcaemic symptoms of WS. Hitman, Garde, Daoud, Snodgrass, and Cohen (1989), for example, investigated 13 families that had one member with WS. They found no indication of a major rearrangement
of the short arm of chromosome 11 (where the calcitonin/CGRP gene is located). Russo, Chamany, Klemish, Hall, and Murray (1991) replicated this result. Russo et al., (1991) also focused on the white blood cell DNA of the WS children in an attempt to determine if a mutation in the calcitonin/CGRP gene might be responsible for the hypercalcaemic symptoms (a mutation that occurred after conception and, therefore, would not be transmitted by either parent). Similar to Hitman et al., (1989), they too did not find evidence of a large deletion or rearrangement of the gene. These investigators summarized their findings by stating that mutations may exist within the calcitonin/CGRP gene itself, or that another locus might account for the deficient levels of calcitonin in WS individuals.

Reports of monozygotic twins with WS have also fuelled research and debate about the genetic basis of WS. Murphy, Greenberg, Wilson, Hughes, and DiLiberti (1990) investigated two sets of monozygotic twins with WS and reviewed the information pertaining to two other sets previously reported. According to these investigators, concordance in monozygotic twins, and no reported cases of concordance in dizygotic twins, adds support to the likelihood of a genetic contribution to the genesis of WS. For example, Hokama and Rogers (1991) described a case of WS in one dizygotic twin, and suggested that this provides evidence for an autosomal dominant mutation. Cortada, Taysi, and Hartmann (1980) reported a mother and daughter with WS, but this was later found to be a case of Noonan syndrome (Preus, 1984). Also, Bellugi, Wang, and Jernigan (1993) reported a case of a father and son with WS, offering yet more support for a genetic basis for WS. Greenberg (1990) referred to a possible autosomal dominant pattern for the disorder, but emphasized that there is most likely a high frequency of new mutations.
Burn (1986), reviewing the literature on WS, described several genetic studies that have indicated a deletion of 15p, a balanced translocation of 9;17, and a deletion of the long arm of chromosome 4 (Jefferson, Burn, Gaunt, Hunter, & Davison, 1986). Burn also included a summary of Grimm and Wesselhoeft (1980), who attempted to determine the possible links between familial supravalvular aortic stenosis and WS. These investigators concluded that these clinical phenotypes are on a continuum, representing the variable penetrance and expression of the same gene. With reference to the earlier findings reported by Burn (1986), the possibility of each of these cases of WS being a phenocopy is left open.

There have also been a number of recently published reports in which attempts have been made to discuss the gene or genes thought to be involved in WS. A number of chromosomes have been implicated, including chromosomes 1, 2, 4, and 11 (Tupler et al., 1992), chromosome 6 (Bzduch & Lukacova, 1989), chromosome 13 and 18 (Colley, Thakker, Ward, & Donnai, 1992), and the 21st and X chromosomes (Telvi et al., 1992). It is the view of Murphy (Murphy et al., 1990) that progress is being made in this area, and that there is reason for confidence that a locus for the syndrome will soon be identified.

Most recently, preliminary results of a research project looking into the genetic basis of WS have been reported (Gene for Williams Syndrome discovered, 1994). In all cases examined in this study to date, a microdeletion on chromosome 7 has been found, this deletion involving an elastin gene. It is the thought of these investigators that this deletion may account for some of the features of the syndrome, including "the facial features, heart disease, hernias, ... and the joint problems" (p. 1).
Medical/Neurological Findings

There are a wide variety of medical complications associated with WS, including those of a cardiovascular nature. For example, Williams (Williams et al., 1961) focused on supravalvular aortic stenosis, while Daniels, Loggie, Schwartz, Strife, and Kaplan (1985) reported peripheral vascular anomalies; both of these can result in hypertension. Terhune, Buchino, and Rees (1985) related these cardiovascular anomalies to a significant increase in risk for myocardial infarcts.

Furthermore, von Arnim and Engel (1964) reported exaggerated tendon reflexes, a finding that is echoed in later publications (e.g., Bellugi, Bihrlle, Jernigan, Trauner, & Doherty, 1990; Trauner, Bellugi, & Chase, 1989). The latter authors also reported a preponderance of dolichocephaly (i.e., a skull with a long anteroposterior diameter) as well as assorted other neurologic abnormalities (e.g., gross and fine motor dysfunction, cerebellar dysfunction, and oromotor dysfunction). According to Jones and Smith (1975), mild microcephaly is common in individuals with WS (approximately 25th centile) and their voices are often described as low and hoarse, or "metallic."

Dilts, Morris, and Leonard (1990) found that a sample of WS subjects exhibited poor strength, balance, and coordination, as well as impaired motor planning. They also noted a tendency towards hypotonia in early childhood, and increasing hypertonia and joint limitations in older childhood. Furthermore, these problems appear to continue into adolescence (Kaplan, Kirschner, Watters, & Costa, 1989) and adulthood (Morris & Carey, 1990; Morris, Leonard, Dilts, & Demsey, 1990).

Dilts et al., (1990) also found that most of their subjects were described as hypersensitive, especially to auditory input. Klein, Armstrong, Greer, and Brown
(1990), tabulating results from a questionnaire survey, found that 95% of their subjects exhibited hyperacusis (i.e., super-sensitive auditory acuity). In the same study, they reported a prevalence rate of 61% for the presence of otitis media (i.e., inflammation of the middle ear) in WS patients. The otitis media reported tended to be recurrent; this typically leads to hearing loss. This fact makes the extremely high prevalence of hyperacusis in these individuals even more intriguing.

A general growth deficiency is widely reported (Jones & Smith, 1975; Morris, Demsey, Leonard, Dilts, & Blackburn, 1988; Pankau, Pertsch, Gosch, Opperman, & Wessel, 1992). Pankau et al. (1992), presenting data on 165 patients, described an average birth length, followed by growth rates at the 3rd centile for males and females. This was followed by a growth spurt between the ages of 11 and 13 years of age for males (resulting in average height at the 10th centile), and a growth spurt between the ages of 10 and 12 years of age for females (resulting in average height at the 25th centile). After the growth spurt, both male and female growth rates slowed once again, with both sexes reaching an average final height at the 3rd centile. Morris and colleagues (Morris et al., 1988) also reported a low-normal height (3rd centile) relative to the combined mid-parent height.

Morris et al., (1988) have referred to a catch-up pattern in relation to the head size of individuals with WS. Their sample is described as having had microcephaly (approximately 2nd centile) during the first four years. After this point, there appeared to be a "catch-up period" to approximately the 25th centile. According to Morris et al., (1988) this pattern is not known in other microcephalic conditions, and thus could be of diagnostic significance.
In adults, lordosis and limitation of joint movement are common (Kaplan et al., 1989; Morris & Carey, 1990; Morris et al., 1988). Morris et al. (1988) also noted a tendency toward obesity in adults. Facial characteristics are thought to endure, with the most noticeable changes being the addition of a broad brow with a prominent supraorbital ridge and bitemporal narrowness (Morris et al., 1990).

Overall, the literature on WS is quite circumscribed at this point in time. Moreover, researchers in the past have tended to focus on the difficulties with, as well as the possible etiological roles played by, the endocrine and cardiac systems of these individuals. As a result, the current knowledge about the effects of development or maturation on the expression of this disorder is somewhat limited. Also, with the exception of the recent past, very little emphasis seems to have been placed on the neuropsychological profile of assets and weaknesses commonly associated with WS.

Morris and colleagues (1988) have attempted to provide a natural history of the disorder and, in doing so, suggested that the facies (typically the group of characteristics most noticeable, and thus most linked to WS) is not readily identifiable at birth. Nevertheless, some parents retrospectively recalled that their affected child was "queer-looking" at birth (Joseph & Parrott, 1958). Morris et al., (1988) found that the mean age of diagnosis was 6.4 years, although for individuals under the age of 16, the mean age at diagnosis was 4.0 years. A series of infant photographs reviewed by the investigators showed that most children "had discernible facial features" by age 4 months, and "obvious Williams syndrome facies by 18 months" (Morris et al., 1988). On the basis of these findings, these authors concluded that the disorder could be diagnosed by the age of 2 years.
These investigators also reported that most parents found the first 12 months to be "miserable". More specifically, many of these parents reported that the child with WS had a number of problems relating to constipation, feeding, vomiting, and colic, all of which may be associated with hypercalcaemia. In addition, these parents indicated that much of their time was occupied with frequent visits to the paediatrician and other specialists (mean of 9.6 visits to the paediatrician in the first 12 months, excluding well-child care).

Udwin and colleagues (Udwin et al., 1986) examined the age at diagnosis and the psychological profile of children with idiopathic infantile hypercalcaemia. Their results were not statistically significant, but they did note a trend towards poorer performances on tasks involving visuospatial and motor abilities for children diagnosed at a later chronological age. Significant effects were found in relation to total behavioral deviance on the Rutter teacher scale (Rutter, 1967; Rutter, Tizard, & Whitmore, 1970) and with hyperactivity subscale scores on the Rutter teacher and parent scales (Rutter, 1967; Rutter et al., 1970). This suggests that earlier identification of the disorder might be beneficial in terms of the identification and attempted remediation of (nonverbal) cognitive and behavioral symptoms.

Dilts (Dilts et al., 1990) reported delays in weight gain and normal growth, as well as delays in reaching developmental milestones (walking independently on average at 21.0 months, talking on average at 21.6 months). These findings were supported by other reports (Lopez-Rangel, Maurice, McGillivray, & Friedman, 1992; Thal, Bates, & Bellugi, 1989). Dilts (Dilts et al., 1990) also suggested that these delays could contribute to delays in the diagnosis of WS (i.e., these children look much younger than their actual chronological age and may not appear to be delayed).
Morris (Morris et al., 1988) reported academic difficulties by the first grade in virtually all of their sample of children with WS, as well as difficulties in adaptive skills and visual-motor integration, noting tentativeness for walking on slight grades or stairs. Joint limitations were also seen to begin in childhood.

Morris et al., (1990) noted that the deficits evident in childhood continue into adolescence and adulthood and, in most cases, increase with the increasing task demands, although Lopez-Rangel (Lopez-Rangel et al., 1992) reported that problems vary greatly from patient to patient. The cardiovascular and hypertensive symptoms also remained problematic, as did a host of other medical concerns discussed above (see Greenberg, 1990; Morris et al., 1990). Joint limitations became more problematic, and kyphoscoliosis and lordosis were common. As a result of their deficits, the majority of these individuals were found to live with their parents or in an institutional setting into adulthood. These findings, especially those related to the social and behavioral limitations, are echoed by Udwin (1990).

**Neuroimaging and Neurophysiological Findings**

Concerning findings from neuroimaging studies, the Salk Institute for Biological Studies has recently published a number of articles and chapters delineating morphological differences between the brains of WS individuals, normal controls and, in some cases, individuals with Down Syndrome. The following reasons were given for using age and IQ matched Down Syndrome controls in these studies: Down Syndrome is relatively well-characterized in the literature as being a syndrome with generalized "psychomotor retardation with language, motor, and cognitive skills all appropriate for their developmental levels" (Trauner et al., 1989, p. 166). This stands in contrast to individuals with
WS, who exhibit similar levels of gross intelligence (i.e., FSIQ of approximately 50), but a radically different overall neurocognitive profile.

An example of such a study is that conducted by Wang, Doherty, Hesselink, and Bellugi (1992), who examined MRI scans of groups of normal controls, WS, and Down Syndrome individuals to search for discernible differences in the morphology of the corpus callosum. A thorough description of the procedure is provided by these authors (Wang, Doherty, et al., 1992). The results of this procedure demonstrated that the corpus callosa of WS subjects, while smaller than those of control subjects, was similar in shape. In the case of Down Syndrome subjects, however, the corpus callosa was found to be shorter, and therefore, more circular than the corpus callosa of both WS and control subjects.

Wang, Doherty, et al. (1992) also reported that, commensurate with the strengths of the WS individuals in linguistic variables relative to the performance of Down Syndrome individuals (WS subjects typically demonstrating better language skills), the width of the rostral fifth of the corpus callosum was more preserved in subjects with WS.

Moreover, these investigators described scans of these same subjects, focusing on cerebral and cerebellar size differences. Using a method developed by Jernigan, Press, and Hesselink (1990), they found that the cerebral volumes of the WS and Down Syndrome groups were significantly smaller than those of the normal controls (approximately 80% for WS and 77% for Down Syndrome; Bellugi et al., 1990). A trend toward brachycephaly was noted for the Down Syndrome subjects, and toward dolichocephaly for the WS subjects. In both cases, the smaller cerebral volume corroborates reports of microcephaly.
When the cerebellar volume of these groups of subjects was examined, a similar picture was noted for the Down Syndrome subjects, with the volume being 69% of the volume of the normal control subjects. The WS cerebellar volume was significantly larger than that of the Down Syndrome subjects', being approximately 99% of the cerebellar volume of the control subjects. This disparity resulted in a significant difference in the cerebellum-to-cerebrum ratio between the WS subjects and the Down Syndrome subjects and controls.

Bellugi then compared measurements of the cerebellar vermal area of the controls and WS subjects (Bellugi et al., 1990). Regarding the scans of the WS subjects, they found that the vermal areas I through V were low-normal relative to the controls, but that the vermal areas VI and VII were significantly larger than those of the controls. The vermal ratio (I through V to VI and VII) was also larger in the WS subjects relative to normal controls.

These results were then related to findings concerning the vermal areas of autistic subjects (where in the opposite pattern is characteristic of individuals with autism: hypotrophy of VI and VII). Hypotheses were proffered concerning the significance of these findings with respect to the different neurobehavioral patterns (e.g., strength of language for WS subjects relative to subjects with autism; see also Wang, Hesselink, Jernigan, Doherty, & Bellugi, 1992).

As a base for these hypotheses, Jernigan and Bellugi (1990) drew on a theoretical paper by Leiner, Leiner, and Dow (1986), in which the latter authors argued that the role of the cerebellum may well extend beyond that which is typically depicted in neuroanatomy textbooks (i.e., control of motor function). More specifically, according to Leiner et al. (1986), phylogenetic and ontogenetic evidence suggests that the increased size of the neocerebellar vermi (VI and VII) in humans, relative to speechless primates, can be linked to the development of
speech. Evidence from positron emission tomography scans during verbal processing tasks has further supported the involvement of area VI and VII in linguistic tasks (Leiner, Leiner, & Dow, 1989). The results of other studies also suggest that it is likely that the cerebellum plays a contributory role in mental processes (Bracke-Tolkmitt et al., 1989).

Jernigan, Bellugi, Sowell, Doherty, and Hesselink (1993) further examined the scans of these subjects and reported that some frontal and temporal limbic structures (i.e., uncus, amygdala, hippocampus, and parahippocampal gyrus) appeared to be spared relative to other structures. These findings are interesting given the (over)sociability of individuals with WS.

Further information concerning the central nervous system of individuals with WS is forthcoming, as Bellugi and colleagues (Bellugi, personal communication, August 29, 1993; Bellugi et al., 1993) recently reported the acquisition of a brain specimen for analysis.

Finally, Neville, Mills, and Bellugi (1993) presented various findings utilizing an event-related potential (ERP) technique. The WS subjects and controls in this study were divided by age. They first reported that the brainstem auditory evoked potentials were similar for WS and control subjects. Stimuli were then presented in the auditory and visual modalities. While the ERPs of WS subjects produced by the visual stimuli were similar to those of the controls, the potentials produced by the auditory stimuli were less refractory (i.e., they demonstrated increased amplitude at fast repetition rates). These results were interpreted to suggest that the visual system of individuals with WS is similar to that of controls, but that the auditory system is different. The authors, based on the ERPs of adults with WS, suggested that the initial stages of auditory language processing are conducted by systems other than those used by normals. They
also hypothesized that the unusually high auditory sensitivity of WS individuals may be responsible for the "island of sparing" (i.e., their linguistic skills).

It is important to note that Neville (Neville et al., 1993) suggested that one should exercise caution when interpreting these findings, due in part to the limited number of WS subjects at each age. Nevertheless, these authors believe that these findings could be utilized in an attempt to develop further working hypotheses.

**Neuropsychological Findings**

Typical neuropsychological findings, when available, will be presented in a manner similar to that of Rourke (1989). With respect to the first category, there does not appear to be any pertinent information relating to *Tactile Perceptual Skills*.

Findings presented relating to the second category, *Motor and Psychomotor Skills*, seem to indicate that, relative to controls matched for age, sex, and various measures of global intelligence or linguistic abilities, individuals with WS perform significantly more poorly on tests demanding more complex skills, but not on those requiring more basic skills. For example, matching children with WS with a clinical control group matched for age, sex, and Peabody Picture Vocabulary Test (PPVT-R; Dunn & Dunn, 1981) Standard Score, MacDonald & Roy (1988) reported similar levels of performance on the Finger-Tapping Test (Reitan & Davison, 1974), but significantly poorer performances for the children with WS on the Grooved Pegboard Test (Klove, 1963). Bennett, LaVeck, and Sells (1978), using the McCarthy Scales of Children's Abilities (McCarthy, 1972), noted that there were no differences between children with WS and controls selected on the basis of referral for "developmental problems" on several of the subscales (Verbal, Memory, and
Quantitative). However, these authors reported that the performance of the WS subjects was significantly poorer than that of the controls on the Motor subscale, consisting of gross and fine motor tasks. Likewise, Pagon, Bennett, LaVeck, Stewart, and Johnson (1987), using the Bruininks-Oseretsky Test of Motor Proficiency (Bruininks, 1978; a measure of both gross and fine motor abilities), reported that WS individuals performed at or below the 3rd centile.

There has been a considerable amount of research concerning the Visual-Spatial-Organizational Abilities of individuals with WS. For example, poor performances were noted on various measures of visual-spatial abilities by MacDonald and Roy (1988) (Target Test; Reitan & Davison, 1974), and Bennett et al., (1978; Perceptual Performance subscale of the McCarthy measure). Pagon et al., (1987) utilized the Developmental Test of Visual-Motor Integration (Beery, 1982), a task requiring the subject to copy designs that follow a "developmental gradient of difficulty" (Spren & Strauss, 1991) and noted that each child with WS performed in a manner below his/her academic achievement age. Likewise, Crisco, Dobbs, and Mulhern (1988) found that the WS subjects performed in a deficient manner on several subtests (e.g., processing of visual information, visual perception, visual closure) of the Illinois Test of Psycholinguistic Abilities (Kirk, McCarthy, & Kirk, 1968).

Inferences can also be drawn concerning other results. For example, examining differences between Verbal and Performance scores on the Wechsler Intelligence Scale for Children - Revised (WISC-R; Wechsler, 1974) can be indicative of the relative levels of the verbal and visual-spatial-organizational abilities of individuals with WS. The majority of studies of WS have reported the Full Scale Intelligence Quotient (FSIQ) of WS subjects to range from mildly to severely mentally retarded; thus, it would be expected that both Verbal and
Performance scales could be rather depressed. However, the Verbal abilities of WS children are often found to be significantly better developed than their Performance abilities (e.g., Udwin et al., 1986, Udwin, Yule, & Martin, 1987), suggesting more poorly developed visual-spatial-organizational abilities.

Bellugi and colleagues have investigated the visual-spatial abilities of individuals with WS utilizing a variety of instruments. They have noted that the pattern of visual-spatial abilities of children with WS is somewhat fragmented. For example, Bellugi, Sabo, and Vaid (1988) reported that WS subjects performed poorly on the Block Design subtest of the WISC-R, on the Developmental Test of Visual Motor Integration, and on the Judgement of Line Orientation Test (Benton, Hannay, & Varney, 1975). These findings are in keeping with those outlined earlier (i.e., weak visual-spatial ability in individuals with WS).

However, further investigation revealed several "islands of sparing". For example, average performances were noted on several tests of facial recognition (Mooney Faces Test; Mooney, 1957); Benton Test of Facial Recognition; Benton, Hamsher, Varney, & Spreen, 1983). The sparing of facial recognition is echoed by Udwin and Yule (1991).

When Bellugi, Marks, Bihrlle, and Sabo (1988) requested the WS subjects to produce freehand drawings of a number of items (e.g., elephant, bicycle; Boston Diagnostic Aphasia Examination; Goodglass & Kaplan, 1972), another interesting discrepancy was noted. The verbal descriptions produced by the subjects as they "talked their way through" the task were quite rich and accurate. The actual drawings, however, were quite poor and particularly deficient in the integration of the parts.
The Delis Hierarchical Processing Task (Delis, Robertson, & Efron, 1986) was used to provide another dimension of the visual-spatial abilities of individual with WS (Bihrlle, Bellugi, Delis, & Marks, 1989). This task requires the individual, after a delay, to reproduce a number of large stimuli, such as a letter of the alphabet or a simple shape (e.g., a square or triangle), that are comprised of small local forms (e.g., a letter of the alphabet or a simple shape). Examples of such stimuli would be a capital "M" made up of "z"s or a large triangle made up of small squares. Using children with Down Syndrome as a control group, Bellugi and colleagues found striking differences between the performances of these two groups. Specifically, the WS subjects tended to produce the local forms (e.g., the "z"s) to the exclusion of the global form (e.g., the "M"), while the Down Syndrome subjects produced the global form (e.g., the "M") to the exclusion of the local forms (e.g., the "z"s). The interesting patterns of visual-spatial abilities, with both deficits and islands of sparing, is uncommon enough that this is thought to be a hallmark of WS and perhaps, could be of use diagnostically.

*Nonverbal problem-solving or concept formation* is an important aspect of adequate neuropsychological functioning, but a part that has not been intensively examined to date with respect to the abilities of individuals with WS. The findings of research by Bellugi, Bihrlle, Neville, Jernigan, and Doherty (1992) indicated that subjects with WS performed very poorly on the Category Test (Reitan & Davison, 1974). Also, as mentioned earlier, these individuals performed poorly on the Block Design subtest of the WISC-R, a subtest that is thought to require some form of nonverbal problem-solving ability for successful completion.
Rote Memory skills represent another area of neuropsychological functioning that is not widely discussed in the WS literature. When performances on tests are presented in a fashion that allows one to determine the overall level of performance, subjects with WS have tended to perform in the moderately to severely impaired range of functioning (e.g., Bennett et al., 1978; Crisco et al., 1988; MacDonald & Roy, 1988).

As with many domains of functioning, the area of Adaptation to novel, complex situations has not been studied extensively in a formal sense. Some evidence would seem to suggest that this would not be an area of strength for individuals with WS. For example, the information obtained from parent-completed measures indicated that their children with WS are prone to overfriendliness with others, especially in the context of language and topics of conversation (e.g., Udwin & Yule, 1990, 1991). It would seem that, in some cases, where a “normal” person learns to tailor his/her conversation to situations, treating each individual as a new situation, WS subjects do not appear to do so. That is, individuals with WS do not appear to modulate or adapt their behavior to a new person or situation. Moreover, in some cases, it is reported that subjects with WS fail to demonstrate adaptation to a situation that is identical to one with which they are familiar (Reilly, Klima & Bellugi, 1991). Bellugi and colleagues (1993) related the experience of requesting a subject with WS to tell and retell a story to the same individual. They noted that there was very little change in the method of recounting the story, despite the listener having previously witnessed the telling of the tale.

As Reading, Spelling, and Arithmetic skills are sometimes associated with supposed precocious linguistic abilities in this population, these areas have been more carefully explored. Generally speaking, these skills tend to be poorly
developed in WS subjects, as assessed by a variety of measures of academic functioning (e.g., MacDonald & Roy, 1988; Pagon et al., 1987). For example, MacDonald and Roy (1988) reported that the achievement scores (Reading, Spelling, and Arithmetic) for the children with WS were significantly lower than those of their controls (e.g., between 2 and 3 SDs below the mean). Pagon et al. (1987), also reported extremely low scores on an achievement test (using the norms as a reference group) and furthermore, that eight of the nine children with WS recorded their lowest or next-to-lowest score on the Mathematics subtest. Udwin et al. (1987) reported that approximately one-half of their sample was unable to register a basal level of performance on tests of reading and spelling (the Neale Analysis of Reading Ability; Neale, 1966; the Vernon Graded Word Spelling Test: Vernon, 1977). Of those subjects able to record at least a basal score (mean age of approximately 12 years), reading and spelling achievement levels were more than 4 years lower than one would expect.

*Speech and language characteristics* are significant dimensions of the unusual neuropsychological profile of WS; hence, there is a plethora of relevant information in this area. A number of researchers, for example, refer to the receptive language skills of individuals with WS. MacDonald & Roy (1988) reported a moderately impaired performance on the PPVT-R (approximately 2 SD's below the mean of 100). Bellugi and colleagues, as part of a large, ongoing study of WS reported receptive language scores as lower than the chronological age of their subjects, but higher than the Mental Age of each subject. Arnold et al. (1985), comparing expressive and receptive language skills of individuals with WS, found the former to be better developed. They also found, however, that the receptive language skills of their subjects were several years behind their chronological age.
Bellugi and her colleagues have undertaken a comprehensive exploration of the linguistic capabilities of individuals with WS, going beyond gross measures such as VIQ (e.g., Bellugi, Marks, et al., 1988). Following the Piagetian theory that cognitive concepts need to be mastered before complex language is possible, these researchers administered a number of tasks that, if completed successfully, demonstrate mastery of the concrete operational stage of cognitive development. Bellugi, Marks, et al. (1988) found that WS adolescents are routinely unable to succeed with tasks of seriation/insertion and conservation/reversibility. Therefore, it was with some surprise that they discovered areas of strength with certain linguistic devices. For example, the WS adolescents demonstrated a capacity to understand complex sentences (e.g., a reversible passive sentence - "The horse is chased by the man"), as well as to detect and correct grammatically incorrect sentences. Again, these linguistic tasks were performed at a level much higher than their performances on nonverbal cognitive tasks would lead one to expect.

Finally, Udwin and Yule (1990), attempted to determine the aptness of the "cocktail party speech" descriptor, and discovered that approximately 37% of individuals with WS have this characteristic. Language samples from this group of children were then compared with WS children who did not display cocktail party speech. The former (i.e., WS children with cocktail party speech) produced a significantly greater number of utterances, intelligible utterances, and utterances containing cliches, idioms, social (filler) phrases, and irrelevant personal experiences. However, Udwin and Yule (1990) concluded that the "content of their speech is by no means meaningless, repetitive, or superficial, since a significantly greater proportion of their speech also serves complex communicative purposes" (p. 111).
Udwin and Yule (1990) went on to compare the language of a group of 20 WS children and controls matched for age, sex, social class, and VIQ. The WS group again produced significantly more utterances containing idioms and social phrases. Udwin and Yule also noted that 70% of the WS group displayed an "over-familiar" manner with the experimenter, relative to only 15% of the control group.

These authors concluded that hyperverbal speech is not characteristic of all WS individuals, but is characteristic of some. When WS children were compared to matched controls, few differences were noted, the main ones involving social aspects of language (e.g., over-familiarity, greater use of adult vocabulary, and social phrases) that contributed to an overall sense of a "glib" or facile use of language. Udwin and Yule (1990) posited that these vocabulary and stylistic differences, in conjunction with severe deficits in other areas of functioning, combine to produce the illusion of an "unusual command of language" (von Arnim & Engel, 1964).

The final area of functioning described by Rourke (1989) is Socio-emotional functioning. This is an area in which individuals with WS tend to experience difficulties, as shown by parent and teacher responses to questionnaires and by anecdotal evidence. Pagon et al. (1987) asked the parents of nine children with WS (age 10-20) to complete the Achenbach Child Behavior Checklist (Achenbach & Edelbrock, 1986). Common behavioral problems (i.e., those endorsed by 7 or more of 9 parents) included immaturity, inattentiveness, clumsiness, talkativeness, and a tendency to be argumentative.

Udwin and colleagues have routinely assessed the socioemotional functioning of the WS children they have studied (Arnold et al., 1985; Udwin & Yule, 1991; Udwin et al., 1986, 1987). Results from the Rutter Parent and
Teacher Scales (Rutter, 1967; Rutter et al., 1970) include 83% of the children exceeding the cutoff score for disturbed behaviour, either at school, at home, or in both venues (Arnold et al., 1985). These authors also reported a prevalence rate of 87% for hyperactivity at home and/or school. In addition, both parents and teachers described the WS children as solitary, and parents were of the opinion that their child was not liked by other children.

Udwin and Yule (1991), with controls matched on age, sex, social class, and VIQ, found that children with WS scored significantly higher on the Deviant score and the total raw score (Teacher Scale; Rutter, 1967; Rutter et al., 1970). Udwin et al. (1987) examined the items comprising the Neurotic score of both scales, and found a significant difference in that a greater number of WS children were described as being worried, fussy, tearful, and complaining of aches and pains.

Udwin (1990) described the findings of a survey of the caretakers of 119 adults with WS, and suggested that difficulties noticed in childhood remained and, indeed, became exacerbated with increasing age. With respect to social interactions and social judgement, 67% of these caretakers reported that these areas remained problematic for the adults with WS. For example, these individuals were found to have difficulties making and keeping friends of their own age. Indeed, it is reported that, even as adults, WS individuals preferred the company of those older than themselves. Udwin reported that 14% of these adults were currently in a relationship, and that 1 (0.8%) was married, while only 8.5% were capable of managing their own leisure time. In excess of 60% of the care-givers reported continued restlessness and inattention, as well as worrying, fearfulness, irritability, and attention-seeking behaviour. In addition, more than 70% reported that a major concern was the safety of the individual with WS, as
there was a tendency towards overfriendliness and a lack of restraint in communications with others (e.g., strangers).

Relationship to White Matter Model

In comparing the neuropsychological profile of WS with that of the NLD syndrome/white matter model, it is of paramount importance to focus on the pattern of assets and deficits, rather than simply the level of functioning in each skill/ability domain. However, as the absolute level of functioning tends to be quite low in individuals with WS, the similarity of patterns tends to be washed out. With this caveat in mind, we turn to a discussion of the findings in infancy and childhood.

Rourke (1987, 1988, 1989) provided a list of characteristics or abilities in which children with the NLD syndrome are expected to be deficient, several of which are applicable to WS children. For example, the expected delayed achievement of developmental milestones predicted by Rourke is supported by the findings of Morris (Morris et al., 1988; average age of walking independently, 21.0 months; average age of talking, 21.6 months) and Thal et al. (1989; two children, age 23 and 66 months, at the one-word stage of language). Dilts and colleagues (Dilts et al., 1990), in a paper defining a behavioral phenotype for WS, reported that many parents described their WS children as very cautious while traversing slopes, stairs, or uneven surfaces, as well as an inability to use common tools (e.g., spreading or cutting with a kitchen knife), corroborating Rourke's prediction of noticeable psychomotor clumsiness in NLD children (Pagon et al., 1987; see Strang & Rourke, 1985, p. 314, and Anonymous, 1985, for delay of specific psychomotor skills such as tying shoes). Others have also noted the similarity of this phenotype to the nonverbal-perceptual-organizational-output disability described by Strang and Rourke (1985) (Dilts et al., 1990).
Rourke (1987, 1989) also postulated the delayed acquisition of self-help skills, an early and consistent finding in WS children. Arnold and colleagues (Arnold et al., 1985) described their sample of 42 children (mean age of 10 years 4 months) as deficient in independent skills and, thus, overly dependent on adults for assistance. In addition to these difficulties, the NLD model would predict significant complications with peer interactions (Rourke & Strang, 1983); this is strongly and consistently supported in the work of Udwin and colleagues (Arnold et al., 1985; Udwin & Yule, 1991; Udwin et al., 1986, 1987).

Udwin and Yule (1991) supported previous anecdotal and parental reports by finding that the language of WS children tends to be of the "cocktail party" variety (37% of their sample), making up in verbosity for what it seems to lack in content. Bellugi et al. (1992) also mentioned the tendency of WS subjects to provide extremely lengthy, somewhat anecdotal responses to items on the Vocabulary subtest of the WISC-R. Moreover, according to Udwin and Yule (1991), these children tended to speak rather formally, with noticeable adult intonations. This observation, in conjunction with the oft-noted tendency to be overfriendly with others, especially adults (e.g., Udwin & Yule, 1991) is also similar to manifestations of the NLD syndrome (e.g., see Sparrow, 1991).

There is evidence to suggest that the pattern of abilities and weaknesses observed in individuals with NLD persists into adolescence and adulthood (Bieliauskas, 1991; DeLuca, 1991; Rourke & Fisk, 1992; Rourke, Young, Strang, & Russell, 1986). This is consistent with reports in the WS literature, and, indeed, it is argued that the deficits are thought to become more apparent and more debilitating with age, especially in relation to situations where judgement and reasoning are required (Morris et al., 1990; Udwin, 1990). Also reported by Udwin (1990), is the finding that older persons with WS are typically found
living with parents or in an institutional setting, and are remarkably deficient in adaptive living skills.

Rourke's (1989) observation of the paucity of formal operational thought with respect to the NLD syndrome is certainly supported by the work of Bellugi (Bellugi, Marks, et al., 1988). That is, these authors demonstrated that three WS adolescents had yet to master basic tasks associated with the concrete operational stage of cognitive development.

Rourke (1989) provided a complete discussion of the neuropsychological assets and deficits of an individual exhibiting the NLD syndrome, many of which are similar to the assets and deficits of an individual with WS. For example, the verbal skills of WS children generally tend to be somewhat more developed than their nonverbal or performance skills (Udwin et al., 1987). Their relative facility with rote material (Udwin et al., 1987) and verbal memory has also been noted (Bennett et al., 1978; Udwin & Yule, 1991). More specific language abilities have been demonstrated to be well-developed, at least in relation to the overall pattern of abilities and deficits (e.g., verbal reception - Bellugi et al., 1990; verbal associations - Bellugi et al., 1990; Udwin et al., 1987; verbal output - Udwin & Yule, 1990). Pagon (Pagon et al., 1987) also reported that the performance of WS subjects on reading subtests of academic achievement tests was better than their performance on tests of arithmetic achievement, a major prediction in the NLD model. Udwin and colleagues (Udwin et al., 1987) also reported early graphomotor difficulties in WS children, consistent with NLD expectations.

Other deficits that appear to be present in NLD and WS children include visual perception (Bihrlé et al., 1989; MacDonald & Roy, 1988), as well as visual attention (Crisco et al., 1988). Visual or visual-spatial memory is also an area of weakness for WS children (Crisco et al., 1988; Udwin & Yule, 1991), although
the latter authors found that tasks that are easily coded in a verbal manner are better handled than those that do not lend themselves to this, another prediction of Rourke (1989). Bellugi (Bellugi et al., 1992) noted the tendency of WS subjects to verbalize visual-spatial tasks spontaneously (e.g., drawing tasks), although the end performance remained remarkably poor. Complex psychomotor skills (Bennett et al., 1978; MacDonald & Roy, 1988) and concept formation are also deficient (Bellugi et al., 1992) in individuals with WS.

Bellugi et al., (1990) described WS subjects as having difficulty with oral-motor praxis, while others have reported that these individuals often demonstrate a good understanding of phonology (Udwin et al., 1987). Both of these findings are in keeping with the predictions of the NLD model. In contrast to the profile of the NLD child, WS children seem to possess adequate abilities in the area of prosody (Reilly, Klima, & Bellugi, 1991). These investigators also reported the use of many other story-telling techniques employed by WS subjects, including voice modulation, character speech (i.e., providing speech for characters in a wordless story), and describing the internal states of characters. However, it is important to bear in mind that, while skilled in this area, the tendency (or need?) to use such story-telling techniques as voice modulation remained abnormally high, suggesting a lack of adaptation on the part of the child. Also, the child with WS may simply be mimicking the prosody of others, rather than using prosody to convey meaning.

Finally, in relation to socioemotional or adaptational abilities, it is well documented that children with WS have deficiencies in social competence (Arnold et al., 1985; Udwin et al., 1986, 1987), in emotional stability (Arnold et al., 1985), and in making and keeping friends (Dilts et al., 1990). These children have also been described by both teachers and parents as hyperactive, immature,

Another link to the NLD/white matter model relates directly to the findings of Wang and colleagues concerning the corpus callosum of WS subjects (Wang, Doherty et al., 1992). It will be recalled that there was a pattern of irregularity, relative to normal controls, with the calloso of WS subjects being smaller than those of the controls but similarly shaped. The relative smallness suggests some compromising of this crucial bridge between the hemispheres.

Finally, Rourke posits a link between these manifestations and deficient functioning of right-hemisphere systems (Rourke, 1982, 1987, 1988, 1989). A good deal of information exists concerning differential deficits of left-hemisphere damaged and right-hemisphere damaged patients, some of which suggests that the visual-spatial deficits of individuals with WS are similar to those of right-hemisphere damaged patients. For example, the performance of children with WS on the Delis Hierarchical Processing Task is very similar to that of patients with damaged right hemispheres (Delis, Kiefner, & Fridlund, 1988; Delis, Kramer, & Kiefner, 1988; Delis et al., 1986). Both groups (children with WS and right-hemisphere damaged patients) exhibited a tendency to focus on the local forms (e.g., the small "x"s making up the global form of "M") to the exclusion of the global form.

Evidence presented by Stiles-Davis (1988) and Stiles and Nass (1989) concerning the pattern of errors on visual-spatial tasks (e.g., Block Design subtest of the WISC-R) again demonstrated the similarities between the performance of children with WS and individuals with right hemisphere damage. Both groups of patients in this case demonstrated an inability to maintain the gestalt of the design (e.g., the two-by-two matrix), similar to their performances on the Delis
Hierarchical Processing Task. Stiles-Davis (1988) also found that the drawings of children with a right hemisphere injury "lack(s) cohesion, [and] the parts ... are scattered around the page. They are not configured into a coherent whole" (p. 262). This is very similar to descriptions of WS children's drawings reported by Bellugi and her colleagues (e.g., Bellugi et al., 1990).

**Memory Functioning in Children with the NLD Syndrome**

There are several studies that have targeted the memory functions of children with the NLD Syndrome (or arithmetic-disabled children). For example, (Fletcher, 1985) utilized the selective reminding technique of Buschke (1974) with verbal and nonverbal tasks to assess the memory functioning of a group of children (aged 92-144 months). These children were first assigned to one of five groups, dependent on their performance on the Wide Range Achievement Test. These groups were arithmetic-disabled, arithmetic- and spelling-disabled, reading- and spelling-disabled, reading-, spelling-, and arithmetic-disabled, and normal (i.e., not learning-disabled). These children were then presented with a list of animal names (verbal task) in a selective reminding task, and an assortment of random dot patterns (nonverbal task). The result that bears most significantly on the current work related to the performance of the arithmetic-disabled children. Fletcher found that these children were not distinguished from the controls on the verbal tasks, but were on the nonverbal task. Thus, (Fletcher, 1985) found that the verbal memory performance of the arithmetic-disabled children was superior to the nonverbal memory performance.

Another study compared the memory functioning of normally achieving, reading- and spelling-disabled, and arithmetic-disabled children (Siegel & Linder, 1984). These investigators used auditorially and visually presented verbal memory tasks. They found that the reading- and spelling-disabled group
of children were deficient on both segments of the task, while the arithmetic-disabled group was performed poorly on the visually presented material only. This suggests an impairment in visual-organizational ability, an impairment that could have an impact on nonverbal memory.

Brandys and Rourke (1991) provide another example of a study that targeted the memory functioning of children exhibiting arithmetic disabilities. Again, a subset of participants were classified as controls, while two groups of learning-disabled children were classified as reading-disabled and arithmetic-disabled. These investigators used a series of experimental verbal memory tasks in an attempt to determine not only the memory functioning of reading-disabled and arithmetic-disabled children, but to point the way to potentially productive avenues for remediation. Briefly, there were four verbal tasks, two presented visually, and two presented through the auditory modality. The material presented in each modality was tested in each of two formats, a free-recall format and an forced-choice recognition format. To assess the nonverbal memory abilities of the groups of children, the Rey-Osterrieth Complex Figure task (Osterrieth, 1944) was utilized.

The results pertinent to the current study, focusing on the findings related to the arithmetic-disabled children, indicated that this group was "particularly poor in the encoding and recall of nonverbal material" (Brandys & Rourke, 1991). An interesting note concerning the recall of the complex figure was reported. More specifically, when the effect of the encoding phase of the task was factored out (i.e., when the measure was percentage of material retained), the three groups performed in a similar manner. This finding suggests that when information (even nonverbal information) is well-encoded by arithmetic-disabled children, it is available for recall.
Another investigation (Fisher & DeLuca, 1997) utilized the California Verbal Learning Test (CVLT; Delis, Kramer, Kaplan, & Ober, 1987) in an effort to examine the learning strategies employed by adolescents and adults with NLD. Briefly, these investigators found that, relative to a matched group of individuals with a verbal learning disability, the NLD subjects preferred a strategy that emphasized rote memory skills (i.e., a serial learning strategy) to one that emphasized organizational skills (i.e., a strategy that relied upon clustering based on semantic similarities). These findings were explained in terms of the neuropsychological strengths and weaknesses detailed in the NLD model and, as such, were consistent with it.

Finally, a recent study examined the memory functioning of groups of WS and Down Syndrome groups (Wang & Bellugi, 1994). As mentioned, WS subjects in previous studies have demonstrated superior verbal skills and inferior visual-spatial skills relative to DS subjects (e.g., Bellugi et al., 1992). In this study, a subtest of the WISC-R, Digit Span was used as the measure of verbal short-term memory, and the Corsi Blocks, as the measure of visual-spatial short-term memory. In support of hypotheses, the WS group exhibited superior performance on the verbal memory task relative to the performance of the DS group, and the DS group performed in a superior manner on the visual-spatial task relative to the performance of the WS group.

**Rationale for Study**

The rationale for this study was alluded to in the introductory remarks. Despite the increasing participation of psychology and neuropsychology in delineating the abilities and deficits of individuals with WS, there is a paucity of information available concerning their memory functioning. In an attempt to contribute to the existing literature concerning mnemonic functioning in children
and adolescents with WS, these functions will be examined in the context of a complete neuropsychological battery. It is expected that the results of this study will, in addition to the above contribution, assist those involved with these individuals (e.g., parents, teachers) in better understanding, and therefore, better assisting these individuals in many areas of day-to-day functioning.

Statement of Hypotheses

The following hypotheses are introduced on the basis of the above-presented theoretical relation between the WS and the NLD Syndrome.

Hypothesis 1

It is expected that the pattern of assets and deficits of children and adolescents with WS will approximate the pattern exhibited by children or adolescents with NLD Syndrome (see Table 1 for an itemization of predictions).

Hypothesis 2

The WS group will perform in a deficient manner, relative to the standardization sample and expectations based on VIQ, on tests of nonverbal memory. This group will perform in a manner consistent with expectations based on PIQ on tests of nonverbal memory.

Hypothesis 3(a)

The WS group will perform in a manner superior to that which would be expected based on PIQ on tests of verbal memory that do not involve a novel component. This group will perform in a manner consistent with expectations based on VIQ on these tests of verbal memory.

Hypothesis 3(b)

The WS group will perform in an impaired manner, relative to the standardization sample and expectations based on VIQ, on tests of verbal memory that involve a novel component.
Hypothesis 4

The performances of the WS group on measures of verbal memory (with the exception of those with a novel component) will exceed their performances on measures of nonverbal memory.
Chapter 2

METHOD

Subjects

Fifteen individuals with WS were assessed. The parents of these children were contacted through the bi-monthly newsletter of the Ontario Chapter of the Canadian Association for Williams Syndrome. In this communication, members of the Association were informed of the general intent of the study and invited to contact the Chairperson of the Ontario Chapter or the author of the study. This resulted in two initial contacts. In addition to this, the author, at the invitation of the Chairperson of the Ontario Chapter, attended the Annual Picnic of the Association. At this time, a handout outlining the study, the requirements for participation, and the benefits of participation were disseminated to interested parents. These parents were also encouraged to indicate their interest on a sign-up sheet. On this sheet, each family indicated the age of the family member with WS, their geographic location, and when they would be interested in participating in the study (i.e., month, day of week). From this sheet, families were contacted on the basis of the age of their child and their geographic location (i.e., there was an attempt by the author to limit travel to those cities between Windsor and Toronto).

Procedure

After the project was cleared by the Ethics Committee of the Department of Psychology, University of Windsor, the author began meeting with families to complete the assessments. To obtain a measure of the optimal functioning of these participants, the assessment, normally a full-day session, consisted of two half-day sessions. For those families in the Windsor area, the Clinical Neuropsychology Research and Resource Centre was used. When this was not
feasible, the author traveled to the location of the family and arrangements made for a suitable testing arrangement.

At the time of meeting the family of the individual with WS, the parent(s) and individual with WS were given a verbal overview of the study, highlighting the purpose of the study and the procedure involved. At this time, they were also asked to read and sign a consent form (see Appendix A), indicating their willingness to participate in the study. They were also informed that the information that they give will be strictly confidential, and furthermore, that any reports that are published as a result of this study would ensure their anonymity. A copy of this consent form remained with the family. Another copy was stored with the file containing the test protocols.

The parent(s) were then given a number of questionnaires to complete while the assessment of the child was being conducted. This packet included a Background History Questionnaire (covering such areas as education history of all family members, birth history of all family members, school history of all family members, medical history of all family members), an account of the medical history of the individual with WS, including the measures utilized to diagnose WS, the Personality Inventory for Children - Revised, and the Behavioural Assessment Scale for Children - Parent's Version (BASC).

The author accompanied the individual with WS to the appropriate testing room and began the assessment. The format of this procedure followed generally accepted assessment principles (see Rourke et al., 1983; Rourke et al., 1986 for a description of these principles) and was individually administered to each participant. This battery included measures of various areas of functioning, such as sensory-perceptual functioning (Sensory-Perceptual Test), motor and psychomotor skills (Grip Strength, Finger Tapping Speed, Grooved Pegboard
Test), psycholinguistic abilities (Speech-Sounds Perception Test, Seashore Rhythm Test, Auditory Closure Test, Verbal Fluency, PPVT-R, Aphasia Screening Test, Verbal subtests from WISC-III, WRAT-R), visual-spatial functioning (Performance subtests from WISC-III, Target Test, Developmental Test of Visual-Motor Integration), concept-formation and problem-solving abilities (Category Test, Trail Making Test), personality functioning (Personality Inventory for Children - Revised, Behavior Assessment System for Children), in addition to measures of memory functioning (Sentence Memory Test, California Verbal Learning Test, Continuous Visual Memory Test). A brief description of the last two memory measures follows.


The CVLT-C (Delis, Kramer, Kaplan, & Ober, 1994) is a list learning task that is based on the technique originally developed by Rey (the Rey Auditory Verbal Learning Test; Rey, 1964), with significant input from cognitive psychology. This measures involves the use of two shopping lists of 15 items each (i.e., List A and List B). List A is composed of 5 different types of fruit, toys, and articles of clothing, while List B is composed of 5 different types of fruit, desserts, and household items. Items from the same semantic category are not presented consecutively on either list. Subjects are first told that they will be read a list of items that they are to buy on a pretend Monday shopping trip, and are instructed to listen carefully and try to remember as many items from the list as possible, in any order. List A is then read at a rate of one word per second. Following this presentation of List A, the responses of the subjects are recorded verbatim. List A is then re-read (the words are in the same order with each presentation), and the subjects are instructed to recall as many items from the list
as they can in any order, including those reported on the first trial. The responses are again recorded verbatim. This procedure is repeated for three more learning trials.

After the five List A learning trials, subjects are informed that they will now be read a new list of items for a Tuesday shopping trip (i.e., List B). Again, the subjects are instructed to listen carefully while the list is read (at a rate of one word per second), and to try to remember as many items of the list as possible, in any order. After the list is presented, the responses are recorded verbatim.

Immediately following completion of the List B free recall trial, the subjects are asked to tell the examiner as many words from the Monday list (i.e., List A) as possible. List A is NOT presented at this time. After this recall, a List A category cued recall trial is given, in which subjects are asked to list all of the items on the Monday list that were “things to wear,” then “things to play with,” and “fruit.”

After this cued recall trial, other measures that will not present direct confounds (i.e., other verbal tasks) are administered for a 20 minute period. Following this delay, the subjects are again asked to list as many items from the Monday shopping list as possible (List A is NOT presented). Next, a long delay cued recall trial is administered, using the same category cues used in the short delay cued recall trial. Finally, a lengthy list of words from List A, List B, and new (phonemically or semantically similar) distractor items are read, with the subjects required to say “yes” if the word was on the Monday list and “no” if it was not on the Monday list.

The adult version of this test is analogous to the children’s version, except that there are four semantic categories, each with four items, rather than three categories of five items (see Delis et al., 1987, for a complete description).
Continuous Visual Memory Test (CVMT)

The CVMT (Trahan & Larrabee, 1988) is a visual recognition memory test that is comprised of 112 complex, ambiguous figures that were designed with the intent of being difficult to verbalize, and thus dependent on nonverbal memory. A subset of the stimuli (i.e., seven figures) are repeated seven times. All other figures are variations of these seven, and therefore, unique. These figures, each depicted on a white background, are presented to the subjects one at a time, with an exposure time of two seconds. The subjects are asked to identify the figure as one that they have seen previously (i.e., respond "old"), or as a figure that they have not seen previously (i.e., respond "new"). Scores that can be derived from this measure of nonverbal memory include the Total Score, the number of designs correctly identified, and a d prime (d') score. This latter score represents a signal detection index of memory sensitivity in the course of the 112 items (using the correct number of "hits" and "false alarms").

The CVMT also involves a Delayed Recognition Task, in which the subjects are asked to identify the recurring figure (i.e., the figure that occurred a total of seven times) from an array consisting of that figure and its' six unique variations.

Finally, there is a Visual Discrimination Task. The subjects are shown the display from the Delayed Recognition Task on one card, along with the recurring item on another card. They are then asked to identify the target item from the card with the single item in the display of seven on the other card. The purpose of this task is to identify those subjects whose poor performance is due to visual discrimination deficits rather than visual memory deficits.
As the norms provided in the CVMT manual are for adults aged 18 and greater, supplemental norms were used for those subjects under the age of 18 (Miller, Murphy, Paniak, LaBonte & Spackman, 1996; Ullman, McKee, Campbell, Larrabee, & Trahan, unpublished manuscript).

**Statistical Tests**

For the purpose of assisting with statistical analyses, some of the scores were converted to standard T-scores ($M = 50$, $SD = 10$) based on scores obtained by the standardization sample. The purpose of converting scores from these measures to standard units was to allow direct inter-task comparison between tasks involving different units of measurement (e.g., those on the CVLT-C and the CVMT). This procedure also facilitated the graphic representation of the data. It should also be noted that the type of statistics that was used was influenced by the limited number of subjects. Finally, preliminary analyses were conducted to examine the data descriptively.

To test Hypothesis 1 concerning the general suitability of applying the NLD model to WS, the following procedure was followed. First, the performance of each individual with WS was assessed to determine if the predictions outlined in Table 2 were met. Each prediction received a yes/no rating (1 if the prediction did obtain; 0 if the prediction did not obtain). These ratings were then summed to determine the total number of predictions obtained for each individual, and therefore, the appropriateness of using the NLD model as a template for each individual. The mean of all of these sums represented the appropriateness of applying the NLD model to the WS group.

To test Hypothesis 2 concerning the performance of the WS group on a measure of nonverbal memory, the CVMT, a t-test was run, comparing the mean performance of the children and adolescents with WS to the performance of the
standardization sample. To compare the mean performance of the WS group on the CVMT to their mean performance on a measure of psychometric verbal intelligence (VIQ), the T-scores from the former were compared to the standardized score from the latter. A similar procedure was used to compare the mean performance of the WS group on the CVMT to their mean performance on a measure of psychometric nonverbal intelligence (PIQ).

To test Hypothesis 3(a), the prediction that the WS group would perform in a manner superior to that which one would expect on the basis of PIQ on verbal tests that do not involve a novel component (e.g., some components of the CVLT-C, Digit Span Forward), the T-scores of the verbal tests that do not involve a novel component were compared to the standardized score of the PIQ. A similar procedure was used to compare mean performances of the WS group on these verbal tests with their mean performance on a measure of psychometric verbal intelligence (VIQ).

To test Hypothesis 3(b), the prediction that the WS group would perform in an impaired manner, relative to the standardization sample, on verbal tests that do involve a novel component (e.g., Sentence Memory, Digit Span Backward), a t-test was performed, comparing the mean performance of this group to the performance of the standardization sample. To compare the mean performance of the WS group on verbal tests that do involve a novel component to their mean performance on a measure of psychometric verbal intelligence (VIQ), the T-scores of these verbal tests were compared to the standardized score of the VIQ.

To test Hypothesis 4, the prediction that the WS group would perform significantly better on measures of verbal memory (with the exception of those with a novel component) than on measures of nonverbal memory, a t-test was performed to compare scores on verbal and nonverbal measures.
Chapter III

RESULTS

The first step in the analysis of the data was to determine if the
performances of the sample on the various measures were normally distributed.
Examination of the skewness and kurtosis values of the measures revealed that
all but the PIQ were normally distributed. As the majority of scores were
normally distributed, all performances on all measures were converted into
standard T scores based on performances reported in the standardization sample
for the various tests. The T scores were calculated so that larger values
represented better performance on the measure in question. The use of this
transformation allowed many different variables to be expressed in comparable
units. This, in turn, facilitated comparisons of performance among tasks.

Concerning the question of the level of significance to be accepted during
the analysis of the data, the following issues were taken into consideration. First,
the standard in psychological research is to use either a .05 or a .01 alpha level to
determine if a finding is significant. The choice of alpha level is determined, in
part, by the purpose of the study.

The purpose of this investigation was clearly exploratory in nature. More
specifically, it was the author's intent to explore (1) the association between the
neuropsychological characteristics of individuals with Ws and the NLD model
and (2) potential discrepancies between verbal and nonverbal memory in the Ws
participants. The long-range goal of this project was to initiate a course of study
into the neuropsychological characteristics of Ws. To meet both short- and long-
term goals, a large number of measures were administered to the participants. 
This not only allowed an examination of the current hypotheses, but also the 
generation of questions and ideas for future investigations.

Given the exploratory nature of the current study, a decision was made to 
maximize the power of the design and use the more liberal (.05) level of 
significance. This tack is often recommended in exploratory research (Tabachnik 
& Fidell, 1989), as the increase in power will decrease the possibility of 
committing a Type II error, and increase the probability of significant findings. 
These findings can then be examined in a more stringent manner in future 
investigations.

Another aspect of the decision to maximize the power of the current 
investigation was to use t-tests (a parametric test) to explore the data and test the 
hypotheses, rather than nonparametric tests. Nonparametric analyses allow one to 
avoid many, if not all, of the assumptions about the normality of the data that are 
made with the use of parametric analyses (Hayes, 1973). However, a 
nonparametric test is less sensitive to smaller differences in the data and, 
therefore, the use of these tests also increases the possibility of a Type II error.

To address the hypotheses advanced in this study, the data were analyzed 
using the Statistical Package for the Social Sciences (SPSS for Windows, Release 
6.0) developed by SPSS, Inc. (1993).
Demographic Data

The participants in the study were 13 children with Ws. One of the parents (12 mothers, 1 father) completed several questionnaires. The families were contacted through their participation in the Ontario Chapter of the Canadian Association for Williams Syndrome (one was also recruited by a parent of a participant). A total of 16 families were contacted. A total of 13 parents gave their informed consent to have their children participate (81.3%) and 3 (18.7%) declined. The data from all 13 participants were used in the investigation. It should be noted that not all measures were administered to all participants. In some cases, completion of the measures was refused (e.g., one child refused to complete the Grooved Pegboard Test), while in other cases, the age limitations of the measures came into effect (e.g., the parents of the 25-year-old participant did not complete the PIC). Finally, in several instances, completed forms were not returned to the investigator.

The age of the participants ranged from 7 to 25 years (M = 154.69 months, SD = 62.69 months). Eight of the participants were male and five were female. One of the participants was left-handed. All others were right-handed. All of the participants were Caucasian. Discussion of the level of education is not as meaningful for this population as for other populations. The severe limitations in overall intelligence for the sample restricted their academic potential, such that some type of “special education” was the norm. However, it
should be noted that the oldest participant in the investigation did graduate from high school and lives fairly independently.

**General Findings**

The average performance of the entire sample on all psychological tests is presented in Table 1. As mentioned, all scores have been converted to linear T scores, based on the means and standard deviations of the norms for each test. Summary scores from the measures of psychometric intelligence are presented in the standard manner (i.e., IQ scores; they were, however, converted to T scores for the purpose of the analyses).

A perusal of the data presented in Table 1 leads to a general observation. Specifically, relative to the normative sample, this clinical sample was quite impaired in their level of functioning. This is reflected in the summary scores of the appropriate Wechsler measure (i.e., VIQ - $M = 55.00$, $SD = 8.16$; PIQ - $M = 50.69$, $SD = 8.15$; FSIQ - $M = 49.38$, $SD = 9.47$). These WISC-III/WAIS-R scores are several standard deviations below the mean of the standardization sample. This overall level of ability had a great deal of impact on the performance of the participants on the other measures used in the study.

**PIC**

As the data from the neuropsychological tests will be discussed in the section following this one, the information gleaned from the Personality Inventory for Children (PIC; $n = 11$) will comprise the remainder of this section. Parental report on this measure revealed a valid and interpretable mean profile
Table 1

Performance on psychological test measures (T scores except for IQ)

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<th>Neuropsychological assets</th>
<th>n</th>
<th>M</th>
<th>SD</th>
<th>Neuropsychological deficits</th>
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### Table 1, cont

Performance on psychological test measures (T scores except for IQ)

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<th>SD</th>
<th>n</th>
<th>M</th>
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<td>16.07</td>
<td>11</td>
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that was suggestive of several areas of concern. Of note was the similarity of the mean PIC profile of these parents to that of parents of arithmetic-disabled children (see Figure 1). Chief among these concerns were issues of related to intellectual functioning ($M = 126.18; SD = 22.39$). Given the level of psychometric intelligence of these participants, this concern would seem to be well warranted.

Following this, the next highest mean was obtained on the Psychosis scale ($M = 103.91; SD = 14.24$). Parents who respond the these items in a similar manner are usually concerned with behavior that would be judged as "strange or peculiar." Scores in this range are also common if parents are concerned about the impaired development and immaturity of their child or adolescent.

The level of response to items corresponding to the Developmental Concerns scale was the next highest ($M = 87.82; SD = 9.20$). This level of parental concern is consistent with responses to the Children's History Form, and is reflective of developmental delays in physical maturation and verbal expression.

The T score for the F scale was at a similar level (87.09). A score of this magnitude is thought to be reflective of a somewhat atypical, but still interpretable response set.

Finally, the parents of the current sample was quite concerned with the level of academic achievement of their offspring ($M = 77.73; SD = 6.93$). This is
consistent with concerns with intellectual development, as well as with the pattern of responses to the open-ended Children's History Form.

A. Hypothesis 1 - Applicability of NLD Model to Ws

To assess the applicability of the NLD model to the current findings, a profile analysis would be ideal. However, as is apparent from the above discussion, the profile of the current sample is, for the most part, flat. Therefore, a simpler approach was taken in response to this first hypothesis.

Table 2 presents a summary of predictions made regarding the performance of the Ws participants on a number of measures included in the battery. The basis for determining the accuracy of these predictions is found in Table 3. In this table, each prediction is itemized for each participant, with a “0” recorded if the prediction was not met. If the prediction was met, a “1” was recorded. In those instances where a test was not administered to a subject, “NA” (not administered) was recorded. The sum of each of the predictions was recorded across the subjects to assess the consistency of the average Ws profile with that of the NLD model. Finally, the sum of each prediction was recorded for each individual subject to assess each individual’s adherence to the NLD model.

The framework developed by Rourke (1989) to discuss the neuropsychological strengths and weaknesses of individuals with the NLD syndrome is utilized in the discussion of the present findings. That is, the neuropsychological assets and deficits of the participants are discussed in terms of their primary, secondary, and tertiary importance (see Table 4). As Rourke has
Table 2

Predictions of NLD Model

<table>
<thead>
<tr>
<th>Prediction</th>
<th>Measures</th>
<th>Prediction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Simple psychomotor &gt;</td>
<td>Finger Tapping; Grooved Pegboard</td>
<td>Finger Tapping &gt; Grooved Pegboard</td>
</tr>
<tr>
<td>Complex psychomotor</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Auditory Perception &gt;</td>
<td>Sensory Perceptual Test</td>
<td>Auditory &gt; Visual/Tactile</td>
</tr>
<tr>
<td>Visual/Tactile Perception</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Auditory Attention &gt;</td>
<td>Digit Span; Target Test</td>
<td>Digit Span &gt; Target Test</td>
</tr>
<tr>
<td>Visual/Tactile Attention</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Auditory Memory &gt;</td>
<td>CVLT/Digit Span; CVMT/Target Test</td>
<td>CVLT/Digit Span &gt; CVMT/Target Test</td>
</tr>
<tr>
<td>Visual/Tactile Memory</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rote &gt; Novel</td>
<td>Digit Span (For); Digit Span (Back); Sentence Memory</td>
<td>Digit Span (For) &gt; Digit Span (Back); Sentence Memory</td>
</tr>
<tr>
<td>Reading &gt; Arithmetic</td>
<td>WRAT-R</td>
<td>Reading &gt; Arithmetic</td>
</tr>
<tr>
<td>Spelling &gt; Arithmetic</td>
<td>WRAT-R</td>
<td>Spelling &gt; Arithmetic</td>
</tr>
<tr>
<td>VIQ &gt; PIQ</td>
<td>WISC-III/WAIS-R</td>
<td>VIQ &gt; PIQ</td>
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<tr>
<td>Internal Dysfunction &gt; External Dysfunction</td>
<td>PIC</td>
<td>Internal &gt; External</td>
</tr>
</tbody>
</table>
Table 3

Adherence to Predictions of NLD Model

<table>
<thead>
<tr>
<th>SS # (age)</th>
<th>S&gt;C &gt;D</th>
<th>Rote &gt;Aud per</th>
<th>Aud per &gt; Aud mem</th>
<th>Aud mem &gt; VIQ</th>
<th>VIQ &gt; PIQ</th>
<th>Read &gt; Arith</th>
<th>Spell &gt; Arith</th>
<th>Int &gt; Ext</th>
<th>Total</th>
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<tr>
<td>10-89</td>
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<td>0</td>
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<td>0</td>
<td>0</td>
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<td>0</td>
<td>5/9</td>
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<tr>
<td>6-101</td>
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<td>0</td>
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<td>0</td>
<td>0</td>
<td>3/5</td>
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<tr>
<td>2-105</td>
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<td>4/8</td>
</tr>
<tr>
<td>4-110</td>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>6/9</td>
</tr>
<tr>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>7/9</td>
</tr>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>6/8</td>
</tr>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>6/9</td>
</tr>
<tr>
<td>12-146</td>
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<td>0</td>
<td>0</td>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>3/4</td>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>8/9</td>
</tr>
<tr>
<td>1-200</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
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<td>0</td>
<td>0</td>
<td>6/9</td>
</tr>
<tr>
<td>9-210</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>8/9</td>
</tr>
<tr>
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<td>0</td>
<td>0</td>
<td>6/8</td>
</tr>
</tbody>
</table>

Note: 0 was recorded if a prediction was not met; 1 was recorded if the prediction was met; NA was recorded if the necessary measures were not administered. Age is presented in months.
<table>
<thead>
<tr>
<th>Neuropsychological</th>
<th>Assets</th>
<th>Deficits</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary</td>
<td>Auditory perception</td>
<td>Tactile perception</td>
</tr>
<tr>
<td></td>
<td>Simple motor</td>
<td>Visual perception</td>
</tr>
<tr>
<td></td>
<td>Rote material</td>
<td>Complex psychomotor</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Novel material</td>
</tr>
<tr>
<td>Secondary</td>
<td>Attention (auditory; verbal)</td>
<td>Attention (tactile; visual)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Exploratory behavior</td>
</tr>
<tr>
<td>Tertiary</td>
<td>Memory (auditory; verbal)</td>
<td>Memory (tactile; visual)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Concept formation</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Problem solving</td>
</tr>
<tr>
<td>Verbal</td>
<td>Phonology</td>
<td>Oral-motor praxis</td>
</tr>
<tr>
<td></td>
<td>Verbal reception</td>
<td>Prosody</td>
</tr>
<tr>
<td></td>
<td>Verbal repetition</td>
<td>Phonology&gt;semantics</td>
</tr>
<tr>
<td></td>
<td>Verbal storage</td>
<td>Content</td>
</tr>
<tr>
<td></td>
<td>Verbal association</td>
<td>Pragmatics</td>
</tr>
<tr>
<td></td>
<td>Verbal output (volume)</td>
<td>Function</td>
</tr>
<tr>
<td>Academic</td>
<td>Graphomotor (late)</td>
<td>Graphomotor (early)</td>
</tr>
<tr>
<td></td>
<td>Word decoding</td>
<td>Reading comprehension</td>
</tr>
<tr>
<td></td>
<td>Spelling</td>
<td>Mechanical arithmetic</td>
</tr>
<tr>
<td></td>
<td>Verbatim memory</td>
<td>Mathematics</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Science</td>
</tr>
<tr>
<td>Socioemotional/adaptational</td>
<td>???</td>
<td>Adaptation to novelty</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Social competence</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Emotional stability</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Activity Level</td>
</tr>
</tbody>
</table>
been careful to point out, these assets and deficits are to be viewed in the context of a cause-and-effect model, with the primary characteristics being causative with respect to the secondary characteristics, and so on.

The first set of neuropsychological assets and deficits to be discussed are the most basic in the hierarchy and are considered by Rourke (1989) to be primary (see Table 4). For example, the prediction that auditory perception (represented by performance on the auditory perception portion of the Sensory Perceptual Test) would be better developed than visual and tactile perception (represented by performance on the visual and tactile portions of the Sensory Perceptual Test) was met by approximately half of the sample (i.e., 44%, 4 of 9 met the prediction). The prediction that memory for rote material (represented by performance on the Digit Span [forward] of the appropriate Wechsler measure) would be better developed than memory for novel information (represented by performance on the Digit Span [backward] of the appropriate Weschler measure and the Sentence Memory Test) was met at a higher level (i.e., 85%, 11 of 13 met the prediction). Finally, the prediction that simple motor skills (represented by performance on the Finger Tapping Test) would be better developed than complex motor skills (represented by performance on the Grooved Pegboard Test) was met by each participant (i.e., 100%, 10 of 10 met the prediction). The mean for these three predictions was 76%.

When determining the adherence to the predictions for the next set of characteristics, several measures were used in the computation of both secondary
and tertiary assets and deficits (e.g., Digit Span [this measure was used in the evaluation of several hypotheses], Target Test). With respect to the prediction that auditory attention (represented by performance on the Digit Span subtest of the appropriate Wechsler measure and the initial trial of the CVLT) would be better developed than attention for visual stimuli (represented by performance on the Target Test), 12 of 13 participants performed in the expected direction (i.e., 92%).

The prediction that auditory memory (represented by performance on the Digit Span subtest of the appropriate Wechsler measure and the initial trial of the CVLT) would be better developed than visual memory (represented by performance on the Target Test and the CVMT) was met by 12 of 13 participants (i.e., 92%).

The next characteristics are related to the verbal functioning of the participants. The prediction that overall verbal psychometric intelligence (represented by the VIQ) would be greater than the overall nonverbal psychometric intelligence (represented by the PIQ) was met by 10 of the 13 participants (i.e., 77%).

The final set of characteristics was related to the academic and socioemotional functioning of the participants. Two predictions were based on performance on the WRAT-R and were met at an identical level. A total of 6 of 11 participants performed at a higher level on the Reading and Spelling subtests than they did on the Arithmetic subtest (i.e., 55%). Finally, the prediction that
parent-completed measures of personality functioning would result in greater internalizing than externalizing tendencies was met 64% of the time (i.e., a total of 7 of 11 participants were described by their parents as being in the expected direction). The mean for these three predictions was 58%.

The strength of the predictions within the participants (i.e., the degree to which each individual met each of the predictions of the NLD model) was quite variable (see Table 3). In a manner similar to that of the above section, the number of predictions applicable to each subject varied, relative to the number of measures that were completed. Taking this into consideration, the strength of the predictions ranged from 50% (i.e., 4 of 8 predictions were in the expected direction) to 100% (i.e., 8 of 8 predictions were in the expected direction). The mean for all individuals in the sample was 75.2%.

The participants in the current study can also be divided into two groups based on age (i.e., young - $M = 8.8$ years, $SD = 0.8$ years; range = 7.4 years - 9.7 years; old - $M = 16.4$ years, $SD = 4.7$ years; range = 11.8 years - 25.4 years). When the extent to which each individual met the predictions was considered, strong evidence was found for an age trend. More specifically, 64% of the predictions across the group of young participants were in the expected direction, whereas 85% of the predictions across the group of old participants were in the expected direction.
B. Hypothesis 2 - Nonverbal Memory in Ws

The purpose of the second hypothesis was to explore the nonverbal memory functioning of individuals with Ws. Accordingly, the performance of these individuals on a measure of nonverbal memory, the CVMT, was compared to three different values. The first of these comparisons was to the standardization sample of the CVMT, the second to their overall level of verbal intelligence (VIQ), and the third to their overall level of nonverbal intelligence (PIQ).

A number of scores are generated by individuals completing the CVMT (Trahan & Larrabee, 1988). The developers of the measure recommend that the "Total" score be used as a summary score. The use of this score takes the magnitude of many other factors into consideration, such as the number of hits (items correctly identified as being seen before) and the number of false alarms (items incorrectly identified as being seen before).

The mean Total T score of participants on this measure was 23.85 (SD = 16.07). The difference between the participants in this study and those of the standardization sample (M = 50) is highly significant (t(24) = -5.87, p = .000; see Table 5) in the direction of the Ws sample performing much more poorly.

The performance of the subjects on the Total score of the CVMT was then compared to the overall level of verbal intelligence (VIQ; M = 55.0; SD = 8.16). The difference between these two means was not significant (t(12) = -.72, p = .485). In a similar fashion, the Total score of the CVMT did not differ from the
Table 5

Nonverbal Memory Relative to Verbal and Nonverbal Intelligence

<table>
<thead>
<tr>
<th>Variables</th>
<th>t-value</th>
<th>df</th>
<th>level of significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total vs standardization</td>
<td>-5.87</td>
<td>12</td>
<td>.000*</td>
</tr>
<tr>
<td>VIQ and Total (CVMT)</td>
<td>-.72</td>
<td>12</td>
<td>.485</td>
</tr>
<tr>
<td>PIQ and Total (CVMT)</td>
<td>-1.20</td>
<td>12</td>
<td>.252</td>
</tr>
</tbody>
</table>

Note. * denotes a significant difference between performance on measure and standardization sample
overall level of nonverbal intelligence (PIQ; \( M = 50.69; \ SD = 8.15 \)) (\( t(12) = -1.20, p = .252 \)).

**C. Hypothesis 3a - Simple Verbal Memory in Ws**

The performance of the participants on measures used to measure simple verbal memory (i.e., Digit Span, indices from the CVLT) was compared to their overall level of nonverbal psychometric intelligence (PIQ) as well as to their overall level of verbal intelligence (VIQ); see Table 6).

A number of these measures of verbal memory were significantly different from the performance on both measures of psychometric intelligence (i.e., PIQ and VIQ; see Table 6). In general, the participants performed at a similar level on both subscales of the WISC-III/WAIS-R, which resulted in the summary scores (i.e., PIQ and VIQ) being very similar. Thus, all measures that were significantly different from the mean PIQ were also significantly different from the mean VIQ. For example, the participants performed in excess of expectations on the Digit Span subtest of the Wechsler tests of intelligence (for PIQ, \( t(12) = -6.16, p = .000 \); for VIQ, \( t(12) = -5.36, p = .000 \); see Table 6).

With some exceptions, similar results were noted with respect to the measures obtained from the CVLT. For example, the immediate recall after the first trial (A1) was in excess of expectations based on level of psychometric intelligence (for PIQ, \( t(12) = -4.67, p = .001 \); for VIQ, \( t(12) = -3.98, p = .002 \); see Table 6).
Table 6

Verbal Memory Relative to Verbal and Nonverbal Intelligence

<table>
<thead>
<tr>
<th>Variables</th>
<th>t-value</th>
<th>df</th>
<th>level of significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>PIQ and Digit Span</td>
<td>- 6.16</td>
<td>12</td>
<td>.000*</td>
</tr>
<tr>
<td>PIQ and A1 (immediate recall)</td>
<td>- 4.67</td>
<td>12</td>
<td>.001*</td>
</tr>
<tr>
<td>PIQ and A15 (total recall across trials)</td>
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<td>12</td>
<td>.193</td>
</tr>
<tr>
<td>PIQ and SDELVA5 (savings)</td>
<td>-25.64</td>
<td>12</td>
<td>.000*</td>
</tr>
<tr>
<td>PIQ and SLOPE (rate of learning)</td>
<td>- 7.76</td>
<td>12</td>
<td>.000*</td>
</tr>
<tr>
<td>PIQ and DISCRIM (recognition)</td>
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<td>12</td>
<td>.575</td>
</tr>
<tr>
<td>VIQ and Digit Span</td>
<td>- 5.36</td>
<td>12</td>
<td>.000*</td>
</tr>
<tr>
<td>VIQ and A1 (immediate recall)</td>
<td>- 3.98</td>
<td>12</td>
<td>.002*</td>
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<tr>
<td>VIQ and A15 (total recall across trials)</td>
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<td>12</td>
<td>.471</td>
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<tr>
<td>VIQ and SDELVA5 (savings)</td>
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<td>.000*</td>
</tr>
<tr>
<td>VIQ and SLOPE (rate of learning)</td>
<td>- 6.88</td>
<td>12</td>
<td>.000*</td>
</tr>
<tr>
<td>VIQ and DISCRIM (recognition)</td>
<td>- .06</td>
<td>12</td>
<td>.949</td>
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</tbody>
</table>

Note. * denotes a significant difference between performance on measure and PIQ/VIQ
The total number of words recalled over five trials (A15), however, was consistent with expectations based on the level of nonverbal and verbal psychometric intelligence (for PIQ, $t(12) = -1.38, p = .193$, ns; for VIQ, $t(12) = -.74, p = .471$, ns).

The level of the next two measures reported from the CVLT were in excess of expectations based on the level of psychometric intelligence. The mean of the first score, SDELVA5 (a measure of savings), was found to be significantly different from the means of nonverbal and verbal intelligence (for PIQ, $t(12) = -25.64, p = .000$; for VIQ, $t(12) = -19.41, p = .000$). The second measure, SLOPE (a measure of the rate of learning), was also found to be significantly higher than expected (for PIQ, $t(12) = -7.76, p = .000$; for VIQ, $t(12) = -6.88, p = .000$).

The next measure, DISCRIM, as noted, reflects the ability to discriminate target words from distracters in a recognition format. The mean score on this measure was consistent with expectations relative to level of psychometric intelligence (for PIQ, $t(12) = -.58, p = .575$; for VIQ, $t(12) = -.06, p = .949$).

The learning characteristics of the group are notable in that they allow the analysis of self-initiated learning strategies. For example, consistent with expectations, there was a trend toward the utilization of rote learning skills (i.e., a serial strategy; SERCLUS; $M = 48.85$, $SD = 12.94$) rather than the utilization of organizational skills (i.e., recalling words by clustering those from semantic categories; SEMCLUS; $M = 40.38$, $SD = 16.13$; $t(12) = -1.21, p = .248$).
Also in keeping with expectations was the consistency with which the participants drew on words from the latter part of the List A relative to those from the initial part of the shopping list (RECEN - $M = 58.85, SD = 19.81$; PRIM; $M = 33.46, SD = 14.77, t(12) = -3.22, p = .007$). The participants also drew more significantly from the middle of List A relative to the initial part of the list (MIDDLE - $M = 56.92, SD = 19.53, t(12) = -3.07, p = .010$).

Another way to examine the use of these learning strategies is in relation to the levels of nonverbal and verbal functioning. Without exception, each of these indices significantly exceeds that predicted from the PIQ and VIQ of the sample (see Table 7).

D. Hypothesis 3b - Verbal Memory with a Novel Component in WS

An additional hypothesis in this vein was related to the performance of WS participants on verbal memory tasks that are considered to be complex in nature. For example, the Sentence Memory test (Benton, 1983) is a measure that is not reliant on "pure" auditory verbal memory or "rote" memory. Instead, an individual must also make use of contextual cues to perform competently. Therefore, it was hypothesized that individuals with WS would perform in a deficient manner on this task. Indeed, the participants in the current study performed in an impaired manner on this test ($M = 19.39; SD = 9.17$) and differed significantly from the mean of the standardization sample (Knights & Norwood, 1980; ($t(24) = 12.04, p = .000$; see Table 8). This level of performance was, however, in keeping with expectations based on the performance of the
Table 7

Verbal Memory Learning Characteristics Relative to Verbal and Nonverbal Intelligence

<table>
<thead>
<tr>
<th>Variables</th>
<th>t-value</th>
<th>df</th>
<th>level of significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>PIQ and SEMCLUS</td>
<td>-4.60</td>
<td>12</td>
<td>.001*</td>
</tr>
<tr>
<td>PIQ and SERCLUS</td>
<td>-8.96</td>
<td>12</td>
<td>.000*</td>
</tr>
<tr>
<td>PIQ and PRIM</td>
<td>-3.88</td>
<td>12</td>
<td>.002*</td>
</tr>
<tr>
<td>PIQ and MIDDLE</td>
<td>-7.25</td>
<td>12</td>
<td>.000*</td>
</tr>
<tr>
<td>PIQ and RECEN</td>
<td>-7.28</td>
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<td>.000*</td>
</tr>
<tr>
<td>VIQ and SEMCLUS</td>
<td>-3.88</td>
<td>12</td>
<td>.002*</td>
</tr>
<tr>
<td>VIQ and SERCLUS</td>
<td>-9.18</td>
<td>12</td>
<td>.000*</td>
</tr>
<tr>
<td>VIQ and PRIM</td>
<td>-3.27</td>
<td>12</td>
<td>.007*</td>
</tr>
<tr>
<td>VIQ and MIDDLE</td>
<td>-6.26</td>
<td>12</td>
<td>.000*</td>
</tr>
<tr>
<td>VIQ and RECEN</td>
<td>-7.15</td>
<td>12</td>
<td>.000*</td>
</tr>
</tbody>
</table>

Note. * denotes a significant difference between performance on measure and PIQ/VIQ
Table 8

**Verbal Memory with a Meaningful Component Relative to PIQ/VIQ**

<table>
<thead>
<tr>
<th>Variables</th>
<th>t-value</th>
<th>df</th>
<th>level of significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sentence Memory vs standardization</td>
<td>12.04</td>
<td>24</td>
<td>.000*</td>
</tr>
<tr>
<td>VIQ and Sentence Memory</td>
<td>.26</td>
<td>12</td>
<td>.800</td>
</tr>
<tr>
<td>PIQ and Sentence Memory</td>
<td>- .86</td>
<td>12</td>
<td>.407</td>
</tr>
</tbody>
</table>

*Note.* * denotes a significant difference between performance on measure and standardization sample
group on both nonverbal and verbal measures of psychometric intelligence (for PIQ, t(12) = -.86, p = .407; for VIQ, t(12) = .26, p = .800).

E. Hypothesis 4 - Verbal Memory Relative to Nonverbal Memory in Ws

The final hypothesis involved a direct comparison of the nonverbal and verbal memory abilities of the participants. More specifically, it was hypothesized that these individuals would perform significantly better on measures of verbal memory than on measures of nonverbal memory.

Accordingly, the Total score from the CVMT was compared to the previously described scores derived from the CVLT (with the exception of the “learning characteristics”) to determine if the means were significantly different. The mean performance of these subjects on the initial trial of the verbal learning task (A1) was significantly higher than their mean score on the nonverbal learning task (t(12) = 3.26, p = .007; see Table 9). The mean of the total number of words recalled over the first five trials, however, did not differ from the mean Total score of the nonverbal task (t(12) = -.13, p = .900; see Table 9).

The mean savings score from the verbal learning task (SDELVA5) was significantly higher than the mean Total score of the nonverbal task (t(12) = 5.37, p = .000). Likewise, the score related to the rate of learning (SLOPE) on this verbal learning task was significantly higher than the summary score of the nonverbal memory task (t(12) = 3.19, p = .008). Finally, the mean performance on the recognition memory portion of the verbal task (DISCRIM) did not differ from the summary score of the nonverbal task (t(12) = -.45, p = .663).
Table 9

Nonverbal Memory Relative to Verbal Memory

<table>
<thead>
<tr>
<th>Variables</th>
<th>t-value</th>
<th>df</th>
<th>level of significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total and A1 (immediate recall)</td>
<td>3.33</td>
<td>12</td>
<td>.006*</td>
</tr>
<tr>
<td>Total and A15 (total recall across trials)</td>
<td>.88</td>
<td>12</td>
<td>.396</td>
</tr>
<tr>
<td>Total and SDELVA5 (savings)</td>
<td>4.47</td>
<td>12</td>
<td>.001*</td>
</tr>
<tr>
<td>Total and SLOPE (rate of learning)</td>
<td>2.70</td>
<td>12</td>
<td>.019</td>
</tr>
<tr>
<td>Total and DISCRIM (recognition)</td>
<td>.06</td>
<td>12</td>
<td>.956</td>
</tr>
</tbody>
</table>

Note. * denotes a significant difference between performance on nonverbal memory measure and verbal memory measures
Summary

There are several notable characteristics of the data generated by the performance of these participants on the measures used in the study. First, these participants are rather uniformly impaired across the majority of tests, relative to the respective standardization samples. As mentioned above, the mean performance on the Wechsler measures was several standard deviations below the mean of the standardization sample. This level of impairment, no doubt, had a great impact on their ability to complete the remainder of the tasks.

The evidence regarding the memory abilities of these individuals was also indicative of some degree of impairment. This was true for both verbal and nonverbal memory (i.e., Digit Span, Target Test, CVLT, CVMT). The level of impairment was especially notable relative to the standardization samples of the respective measures. When performance on these various measures of verbal and nonverbal memory were compared to levels of verbal intelligence (VIQ) and nonverbal intelligence (PIQ), a different perspective emerged. More specifically, their performance on a number of measures of the CVLT (e.g., immediate recall, rate of learning) were far in excess of those that would be consonant with these levels of psychometric intelligence.

The CVLT, in particular, allowed a more fine-grained analysis of the learning strategies employed by the participants and supported the link between WS and the NLD syndrome. More specifically, a strength in rote memory skills and a weakness in organizational skills was reflected in a trend toward the
adoption of passive learning strategies (e.g., clustering on the basis of serial/rote cues rather than on semantic category).
Chapter IV

Discussion

A. Comparison of WS with NLD

A proper discussion of the results of this study requires mentioning the work conducted by Rourke and colleagues relating to the NLD syndrome. The findings, culminating from some 20 years of clinical work and theorizing, resulted in the forwarding of a developmental neuropsychological model designed to discuss central processing difficulties in children in a cause-and-effect manner (Rourke, 1982). This model, since revised (Rourke, 1987, 1988), is the basis for the current work. It was painstakingly developed to examine and explain more aspects of a particular subtype of learning disabled child. The result of this was the publication of a treatise on the syndrome and model of NLD (Rourke, 1989).

Numerous accounts in the literature have commented on the similarities between various pediatric diseases and disorders and characteristics of the NLD syndrome (e.g., hydrocephalus - Fletcher et al., 1992; Williams syndrome - MacDonald & Roy, 1988). An edited volume (Rourke, 1995a) takes up where these comparisons left off. One of the chapters in this work (Anderson & Rourke, 1995) examined the literature on Ws and reported a strong theoretical link between the two disorders. In an effort to further develop this link, as well as to further demonstrate the "real life" significance of the NLD syndrome, the current project was undertaken.
In addition to the theoretical value of the current work, a practical consideration is noteworthy. As mentioned previously, WS has only recently come under the neuropsychological microscope. To date, a majority of the studies are still seeking to explicate the neuropsychological characteristics of the disorder. This exploration is often somewhat piecemeal in nature, examining an area of neuropsychological functioning in isolation, but not in relation to other areas of functioning in the same sample. The current study provided an opportunity to do this.

The application of a framework, such as the NLD model, can also serve as a theoretical guide for future investigations of WS. What is often lacking in the exploration of a disorder is a coherent model that will assist in the understanding and explanation of current results as well as in the direction of future investigations. By demonstrating the applicability of the NLD model to WS, we have provided such a framework for the study of WS.

In addition, the similarity in neuropsychological assets and deficits in these two disorders can be very beneficial with respect to treatment considerations. Many recommendations, based on these neuropsychological assets and deficits, have been suggested for children with NLD (e.g., Rourke, 1995b; Rourke, Del Dotto, Rourke, & Casey, 1990; Strang & Rourke, 1985). Mapping the characteristics of individuals with WS onto those with the NLD syndrome opens this literature to parents, school personnel, and healthcare professionals. The efficacy of these methods can also be examined empirically,
as they may require some degree of modification from their current state.

However, as the principles were initially developed on the basis of the asset and
deficit stream of the NLD model, the application of these treatment principles to
WS should proceed fairly smoothly.

A discussion of the results and their meaning with reference to the
hypotheses follows.

Applicability of the NLD model to WS

The support for Hypothesis 1, relating to the consistency of the data with
the tenets of the NLD syndrome was quite strong. This support was present both
in the group and individual data. That is, the majority of predictions were met for
the group as a whole, which supports previously made theoretical links (e.g.,
Anderson & Rourke, 1995). In addition, when the data for each of the
predictions was examined for each individual, there was a strong adherence to the
predictions of the NLD model.

The relative strength of these predictions is also of interest. Rourke
(1989) has proposed that the NLD model is a “stream” of primary, secondary,
tertiary, verbal assets and deficits that are posited to interact in a cause-and-effect
relationship and to have predictable effects on academic and socioemotional
functioning. That is, the primary neuropsychological assets and deficits are
thought to result in (i.e., be a cause of) the secondary neuropsychological assets
and deficits. If the characteristics of the individuals with WS are truly
representative of the NLD syndrome, the same cause-and-effect relationships
would be expected to obtain. Continuing in this vein, an individual (or group) should be expected to match more strongly on those characteristics considered more primary (i.e., "causative") in the NLD hierarchy (see Harnadek & Rourke, 1994) than on those "further down the stream". For the most part, this is consistent with the data.

For example, Harnadek and Rourke (1994) reported that a poor performance on tasks tapping psychomotor functioning (i.e., the Grooved Pegboard Test) was the characteristic that most consistently identified their sample of NLD children. This prediction, that simple motor skills would be better developed than complex psychomotor skills was met in each of the participants who completed the necessary measures (10 of 10 met the prediction - 100%). Results of a similar magnitude were found with respect to the secondary and tertiary characteristics of attention and memory (i.e., that verbal attention and memory would be better developed than visual or tactile attention and memory; 12 of 13 met the prediction - 92%). (It should be noted that the measures used to test the predictions regarding attention were the same as those used to test the predictions regarding memory; thus the identical value for each prediction.)

In accordance with the tenets of the NLD model, less consistent findings were reported with respect to those characteristics farther removed from the most basic (i.e., primary) neuropsychological characteristics. For example, those predictions concerned with the verbal or academic functioning of the participants were met relatively less frequently (e.g., VIQ > PIQ - 77%; WRAT-R Reading >
than those predictions concerned with the aforementioned primary characteristics.

In the current investigation, an explanation for this finding is related to the
developmental delays that are so prominent in this population. More specifically,
it takes time for the characteristics “farther down the stream” to develop for
individuals with WS. Despite clear strengths and weaknesses in the primary
assets and deficits (e.g., motor and psychomotor functioning) of the participants,
the ability to read, spell, or do arithmetic only emerges in the “older” individual
(e.g., those 12 years of age or older). It should also be mentioned that the
academic abilities of these “older” individuals are somewhat poorly developed
and in keeping with levels of nonverbal and verbal psychometric intelligence.

An examination of the personality functioning of these individuals is also
quite enlightening with respect to the similarities between these two syndromes.
More specifically, the mean PIC profile for these participants is identical (albeit
somewhat more pronounced) to that of an arithmetic-disabled (NLD) group of
children (Group 3) that was described by Strang and Rourke (1985, p. 309). That
is, the primary concerns endorsed by parents were related to the intellectual
functioning, “strange or peculiar” behaviors, level of developmental maturation,
and academic achievement of their children. This is reflective of the similarity in
the manner in which the parents of these groups of individuals described their
children. Thus, it would appear that the parents of these WS participants have
cconcerns very similar to those of parent of children with the NLD syndrome.
An aspect of the current study that is in keeping with previous investigations into the development of individuals with the NLD is the tendency for the characteristics of the syndrome to become more evident over time (Casey et al., 1991). When these investigators divided their sample into two age groups, this was, indeed, the case. This trend was also evident in the current study. More specifically, predictions based on the characteristics of the NLD syndrome were supported in the order of 64% for the young group, whereas 85% of the predictions were in the expected direction for the old group.

This trend was particularly strong with regard to those predictions that deal with some aspect of academic functioning (i.e., those involving performances on the WRAT-R). These predictions were met very infrequently in the young participants (i.e., in 1 of 10 possible instances). In the older participants, however, these same predictions were met in 11 of 12 possible instances. If the results are considered in this manner, it would seem that individuals with WS require time to “grow into” their deficits. This is the very pattern that was evident in the above (i.e., Casey et al. 1991) study.

There is one final point to consider in the comparison of the neuropsychological profile of the WS participants with predictions based on the research into the NLD syndrome. Rourke (1989) has emphasized that the NLD syndrome is not defined by a score on any particular measure, but instead by the pattern of scores across a number of measures (see Table 2). As noted previously, the participants in this study were, in many respects, quite “low
functioning" in terms of level of psychometric intelligence (e.g., FSIQ of 49.38). The impact of this level of functioning is reflected in poor performances on the majority of other measures, with the result being a "flat" profile. When this fact is taken into consideration, the emergence of any discernible pattern becomes even more meaningful.

**Memory Functioning**

The remaining hypotheses relating specifically to memory functioning in WS also stem from the theoretical link between WS and the NLD syndrome. A confounding factor in testing these remaining hypotheses is the overall level of functioning of the sample, which, as mentioned previously, was quite low (i.e., FSIQ - M = 49.38; VIQ - M = 55.00; PIQ - M = 50.69). Nevertheless, the hypotheses comparing memory functioning to the overall level of functioning were supported. On the other hand, hypotheses comparing the level of memory functioning to differences expected in terms of a VIQ/PIQ split were not supported. This lack of support is thought to be a direct result of the similarity between levels of nonverbal and verbal psychometric intelligence.

In keeping with expectations, however, and consistent with previously published reports (Wang & Bellugi, 1994), verbal memory, as measured by the CVLT, was better developed than nonverbal memory, as measured by the CVMT (see Table 9). Moreover, the use of the CVLT and its various scales permitted some degree of insight into the manner in which memory functions in these individuals.
The performance of the sample on the initial trial of this measure (CVLT - A1) was in excess of expectations based on level of functioning (i.e., PIQ/VIQ), as well as in excess of their overall score on this measure (CVLT - A15). This is suggestive of relatively good deployment of attentional resources. Performance on the initial trial is also thought to tap into relatively more “pure” mnemonic abilities, whereas the use of Trials 2-5 allows the investigator to tap into patterns of learning.

A measure of general memory functioning/verbal learning (CVLT - A15) was several standard deviations below that of the normative sample, and consistent with overall functioning (i.e., FSIQ). However, such was not the case for all indices of this task. For example, the participants demonstrated a learning curve (SLOPE) in excess of expectations based on (a) the measure of general memory functioning and (b) their overall level of functioning.

Utilizing measures derived from the cognitive psychology literature, the developers of the CVLT allow for an examination of the strategies employed to learn the lists (knowingly or unknowingly) by those taking the test. For example, a more active approach to learning is demonstrated by a reliance on clustering the words based on their semantic similarities (e.g., to name members of the “fruit” or “clothing” category together). A more passive approach to learning would be demonstrated by a reliance on clustering the words based on their serial order (e.g., to recall the words by way of a fixed or mechanical procedure).
The WS participants demonstrated a tendency to rely more heavily on the serial clustering strategy (average - consistent with the normative sample) than on a semantic clustering strategy (one standard deviation below that of the normative sample). This suggests that these participants are inclined to adopt a passive learning strategy on this task. This, as mentioned, is consistent with expectations based on relative strengths (i.e., rote skills) and weaknesses (i.e., organizational skills), as well as with previous research (Fisher & DeLuca, 1997).

Again drawing on the cognitive psychology literature, the developers of the CVLT refer to a respondent’s tendency to recall words from certain portions of the list. For these purposes, recall of the first four words of the list (CVLT - PRIM) and the next seven (eight on the adult version of the CVLT) words (CVLT - MIDDLE) is thought to be reflective of strong learning skills. A tendency to rely on the latter part of the list (i.e., the last four words - CVLT - RECEN) is thought to be consistent with a passive approach to learning, a “parroting” of the last words presented.

The WS participants recalled words from the later portion of the list (CVLT - RECEN) more often than they recalled words from the initial part of the list (CVLT - PRIM). Indeed, their tendency to utilize the latter part of the list was in excess of the normative sample, and their tendency to utilize the initial part of the list was far below that of the normative sample (greater than 2 standard deviations below). Such a finding is also suggestive of a passive approach to learning. However, the participants also demonstrated a greater than
average tendency to draw on the middle portion of the list. This is typically thought to be suggestive of “strong learning skills” (Delis et al., 1987). In this case, it is most likely a result of the overall poor performance on this measure, as these indices are computed independent of the overall level of performance (i.e., the performance across all five learning trials).

Another aspect of learning that is derived from the cognitive psychology literature refers to the retention of words recalled following a delay (the “savings” score). The ability of the participants to recall words that were previously recalled (SDELVA5) is in excess of that expected given their overall performance on this test. Indeed, their savings score was in keeping with levels attained by the normative sample.

The final score to be discussed is related to the ability of the participants to distinguish between words from the first list and those not from this list (CVLT - DISCRIM). A measure of this ability is provided in the context of a recognition trial. The ratio of “distracter” items (those not from the first list) to “target” items (those from the first list) is two-to-one. The resulting score is typically compared to performance on other scales and interpreted in terms of relative difficulty with encoding or retrieval. The participants’ level of performance on this scale was particularly poor. On the whole, there was a tendency to over-identify words as being from the first list. This tendency could be reflective of confabulatory tendencies, perseverative tendencies, or a “yes” response bias.
Anecdotal Evidence in support of this link

In the course of this investigation, a number of anecdotal incidents were noteworthy with respect to the characteristics of WS and its' relation to the NLD syndrome (Anderson & Rourke, 1995; Rourke, 1989). Descriptions of several of these incidents follow.

Inspection of parent completed history forms revealed a number of consistencies with previously published reports (e.g., Dilts et al., 1990) and descriptions of children with the NLD syndrome. For example, without exception, the parents of the participants reported delays in the achievement of motor and language milestones. Those delays associated with motor functioning (fine motor in particular) appeared to be more pervasive and long-lasting. Only two of the participants were reported as able to master the tying of their shoes, a task that is quite demanding in terms of fine motor functioning and eye-hand coordination. The majority of the participants, in particular the younger ones, were also noted to frequently request assistance in the completion of activities of daily living (e.g., buttoning clothes).

Finally, most of the participants were reported as being comfortable in their interactions with other children and adults. However, when a preference was indicated, it was invariably in the direction of adults. There are a number of possible explanations for this finding. One is related to the language functioning of individuals with WS. The utilization of a more extensive vocabulary, as well
as the use of adult intonations (e.g., Udwin & Yule, 1990) and other "adult-like" speech characteristics could endear them to adults, moreso than to other children.

Likewise, the manner in which these individuals are treated by adults could also play a role in this preference. More specifically, adults are typically quite caring in their attitude to children, especially children who are very responsive to the talk and attention paid to them and who respond in like manner. In other words, adults are likely to encourage a more passive form of interaction, one that allows a child with WS to both avoid weaknesses and emphasize strengths. Other children, however, are more likely to insist on physical "playing", which draws on the deficit stream of children with WS (e.g., visual and tactile perception, psychomotor skills, exploratory behavior). Strengths in verbal functioning are not likely to be valued by other children.

**Hyperacusis**

As mentioned in the introduction, there are numerous accounts of hyperacusis in the WS literature (e.g., Klein et al., 1990). Several parents also discussed this when speaking with the investigator. A most striking instance of hyperacusis took place while the investigator was testing a participant. In this case, the participant, a male, was completing an untimed task (i.e., the Category Test). He interrupted his work to voice his dislike of arguments. The examiner responded that he too was not particularly fond of arguments and strained his ears in an attempt to determine whether there was an environmental stimulus for this comment. When such a stimulus was not be discerned by the examiner, the
participant was asked if there was any reason for this comment at this time. With a gesture, he thumbed over his shoulder (i.e., behind him). In the next yard, a neighbor was arguing with his son, although the examiner was only able to determine this by the look on their faces and the gestures that each was using. This individual with WS apparently heard the argument through the windows of an enclosed room (with windows on one side) and the adjoining porch (again, with windows on the outer wall). This boy's father recounted numerous instances of his son's sensitive hearing (e.g., hearing fire sirens several minutes before other family members were able to hear them).

A Similarity to Asperger Syndrome

One of the commonly described clinical features of Asperger syndrome is an "intense absorption in circumscribed topics, such as the weather, facts about TV stations, railway tables, or maps, which are learned in rote fashion ... " (Klin, Sparrow, Volkmar, Cicchetti, & Rourke, 1995). Several of the participants with WS evidenced such an absorption in various topics. The individual described above (hyperacusis), for example, possessed a burgeoning collection of used vacuum cleaners, numbering greater than 20 at the time of the evaluation. He had business cards, advertising himself as a "collector" of vacuums. His father reported that this boy had a near encyclopedic knowledge of vacuum cleaners and that he enjoyed nothing more than engaging in lengthy discussions with sales personnel in department stores. It was not uncommon for sales personnel to
spend a good bit of time conversing with this individual, convinced that they were about to make "the sale of the century."

Another male participant had developed an intense fascination with large trucks, especially garbage trucks. Whenever a truck would be sweeping through his subdivision, this boy would insist on going outside to witness the truck and to watch it work. This took place reliably, despite an initial aversion to the sounds emanating from the truck. There were also a number of household toys in this boy's possession that related to his interest. For example, his favorite toy, that he had just received for Christmas, was an "operational" garbage truck (i.e., the back opened to accept refuse, lights flashed when it was running). This youngster was also fascinated with lawn mowers, and would often sit on summer days and watch neighbors doing their yard work.

A final instance of this type and level of fascination is that of a young man who maintained an intense focus on airplanes. In the course of the two-day assessment, he regaled the examiner with many stories of the air shows that he had attended and of the airplanes that he had seen. He went into quite a degree of specifics in explaining the differences between the types of cargo planes that he had seen. In addition, he was eagerly anticipating an upcoming local air show, which was going to feature a certain plane that he had not had an opportunity to see before.

Limitations, Implications and Suggestions for Future Research
As with any investigation, there are weaknesses and limitations with the current study. The first is related to the size of the sample. To a very large extent, the small sample size (n = 13) is a by-product of the syndrome under study. More specifically, this disorder is exceedingly rare, in the order of 1 case in 25,000 live births. The rarity of the disorder had an effect in another way as well. As a result of the low prevalence, individuals with WS are considerably less than centralized than other samples. Three of the 13 subjects were assessed over two days in Windsor. All others were assessed over two days in their home (e.g., London, Toronto).

The age-range of the participants is quite varied, ranging from 7-25 years of age. This was mostly a result of the rarity of the disorder. While this fact decreased the number of individuals in any one age range (e.g., 5-8 year olds), it did allow an opportunity to follow trends over time in a cross-sectional manner. Keeping in mind the exploratory intent of this investigation, the examination of age-related trends in this population is more enlightening than using such a strictly defined age-group. The information gleaned from this approach is also beneficial with respect to the generation of further questions to be examined. For example, the younger participants were, because of their limited level of functioning, often unable to complete tests (e.g., without having acquired sound-symbol matching or graphomotor control, the younger children were all but unable to complete the spelling subtest of the WRAT-R). However, when one adopts a cross-sectional perspective, it would appear that these abilities do
develop over time. The developmental nature of this trend was clearly observed in the tendency for the predicted WRAT-R Reading and Spelling subtest scores to be greater than WRAT-R Arithmetic scores as the age of the participant increased (1 out of 10 predictions met in the younger participants, 11 of 12 predictions met in the older participants).

A final criticism of the study could be related to the low "level of functioning," with respect to psychometric intelligence, of the participants and represents a conundrum frequently faced by clinical investigators. The "floor" of the majority of measures utilized in this study was too high for the participants, which resulted in a clustering of scores in the impaired range of functioning. This clustering limited the applicability of several of the rules for classifying individuals (e.g., VIQ > PIQ at least 10 points, WRAT Standard Score for Reading > Arithmetic at least 8 points). It also resulted in a limitation in one's ability to examine the patterns and relationships among test results that are a major component of the interpretation of the test data (Reitan, 1986; Rourke, Bakker, Fisk & Strang, 1983).

Memory functioning is thought to play a large role in the overall level of cognitive functioning and has recently been reviewed with respect to individuals with mental retardation (Bray, Fletcher, & Turner, 1997). This review and the cognitive strategies recommended mesh very well with the findings of the current study. More specifically, these authors report that "strategy competency in children with and without mental retardation is similar, but the cognitive
potential of children with mental retardation requires more situational support before strategies are adopted..." (Bray et al., 1997, p. 212). In many cases, however, it is necessary to explain tasks much more extensively for individuals with mental retardation as well as to assist with the development of these strategies (e.g., organizing items on the basis of semantic cues rather than serial cues). It may also be necessary to train such individuals in recognizing appropriate times to use a strategy. These authors also report that the use of "external representations" has proved to be very helpful, especially with respect to mathematical functioning (e.g., counting numbers, months on one's fingers).

If one takes into consideration the natural strengths and weaknesses of individuals with WS and the similarities to individuals with the NLD syndrome, the above recommendations can be fine-tuned. For example, utilizing natural strengths in verbal functioning in developing compensatory strategies will be much more productive than a strategy based on visual-spatial weaknesses (see Rourke, 1995b).

Directives for future research in the area of WS should be in the vein of continuing to more clearly delineate the strengths and weaknesses of these individuals. This would have the added benefit of allowing those in the healthcare field to further modify treatment regimens. Building on this theme, empirical study of the applicability of current treatment considerations for children with the NLD syndrome (e.g., Rourke, 1995b) to the WS population should be undertaken. In addition, further research into the manner in which
these individuals process information in a variety of contexts would also prove beneficial (Baumeister, 1997), especially with respect to the development of compensatory strategies.

The utilization of control groups would be very helpful in determining patterns of neuropsychological strengths and weaknesses. Other investigators, for example, have focused on the overall level of functioning (e.g., Bellugi) and the level of verbal functioning (e.g., Udwin).

Finally, the value of adopting a developmental approach to the study of skills and abilities in this population cannot be overemphasized. For example, the differentiation of word recognition/spelling and mechanical arithmetic in the current study was only evident as the age of the participants increased. A similar approach focusing on other aspects of neuropsychological functioning, it is hoped, would uncover other such developmental tendencies. In a related vein, the use of measures that have a lower floor (i.e., that allow for greater variability in performance) would prove very useful in the examination of the aforementioned patterns in test results.
References


Appendix A

CONSENT FORM
Study of Memory Functioning in Individuals with Williams Syndrome

Purpose: This research project is being carried out by the primary investigator, Peter Anderson, as a partial requirement for the doctoral degree in Clinical Neuropsychology. This study is concerned with the memory functioning of children and adolescents with Williams Syndrome, and how this aspect of neuropsychological functioning relates to other areas of functioning.

What Participants Do: If you agree to participate in the study, you will be asked to complete a packet of questionnaires relating to information concerning the members of your family, and in particular, the individual with Williams Syndrome. This will require approximately 6 hours of your time. The individual with Williams Syndrome will be asked to complete a full day of neuropsychological tests (split up into two half-days for those between the ages of 5 and 8).

Participant's Rights: Your participation in this study is entirely voluntary. If for any reason you do not wish to continue your participation in this study, you will be free to drop out at any time. The information obtained from you will remain entirely confidential. If the results of this study are published, the information will remain anonymous and disguised so that no identification of participant's can be made.

Feedback: Once the study has been completed, you may receive a copy of the results. The results will include a summarization of the findings of the study, rather than information pertaining to any participant's individual scores. Please indicate your desire to receive a copy of the results by writing your name and mailing address on the back of this form.

I understand that this research has been cleared by the Ethics Committee of the Department of Psychology at the University of Windsor, and that any concerns about the procedure of this study may be reported to that Committee (Chair: Dr. Ron Frisch, 519-253-4232, Ext. 7012).

If you have any questions about participating, please feel free to contact me at any time (Peter Anderson, 519-253-4232, Ext. 2217) or the research supervisor (Dr. Byron P. Rourke, 519-253-4232, Ext. 2217).

I, __________________________ (please print name), have read this consent form and agree to participate, and have my child participate in this study. I also give permission for my child's teachers to complete a questionnaire about my child's behaviour in the school.

Signed ________________________ Date ____________________
VITA AUCTORIS

Peter Edward Anderson was born on January 13, 1965 in the Near North of Ontario. From that time forward, Powassan was not to be the same. With his guidance and, certainly his input, his parents, William and Virginia began to develop a close “working relationship” with numerous people in the field of education (i.e., Peter’s teachers and principles). After taking great pains (and creating some too) to make sure that he was “appreciated” in the junior and senior public schools in Powassan, Peter moved onto high school, where a somewhat more studious attitude was adopted. It was in this setting that he developed an interest in psychology and decided to pursue it “South of the Border.” A stop in Houghton, New York for 3 years was followed by another stop in West Chester, Pennsylvania for another 3 years. Despite watching the majority of his friends settle into comfortable lives, with spouses, “real” jobs, and big screen televisions, and despite having to perpetually answer the question “Are you STILL in school???” he decided to earn one more degree. This degree was in Clinical Neuropsychology at the University of Windsor. The time has come for a new response to the age-old question. “No, I’m not STILL in school!”
IMAGE EVALUATION
TEST TARGET (QA-3)