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Cognitive Heterogeneity in Williams Syndrome

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This study used the Woodcock–Johnson Tests of Cognitive Ability–Revised to investigate a wide range of cognitive abilities in people with Williams syndrome (WS). It involved a comparatively large sample of 31 people with WS, but took a case-series approach. The study addressed the widespread claims of a characteristic "WS cognitive profile" by looking for heterogeneity rather than homogeneity. People with WS showed a variety of preserved (significantly above mental age [MA]), expected (at MA), and significantly impaired (significantly below MA) levels of functioning. Such results provide clear evidence for heterogeneity in cognitive functions within WS. We found the most homogeneity on a test of phonological processing and a test of phonological short-term memory, with half of the WS sample performing at MA levels on these tests. Interestingly, no WS individual showed a weakness on a test of nonverbal reasoning, and only one WS individual showed a weakness on a test of verbal comprehension. In addition, we found that strengths on analysis–synthesis and verbal analogies occurred only for WS individuals with an MA less than 5.5 years (our sample median MA); people with an MA greater than 5.5 years performed at MA level on these 2 tests. Results also provided preliminary evidence for distinct subgroups of WS people based on their cognitive strengths and weaknesses on a broad range of cognitive functions. On the basis of the findings, caution should be made in declaring a single cognitive profile that is characteristic of all individuals with WS. Just as there is heterogeneity in genetic and physical anomalies within WS, not all WS individuals share the same cognitive strengths and weaknesses. Also, not all WS individuals show the profile of a strength in verbal abilities and a weakness in spatial functions.

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Williams syndrome (WS) is a reasonably rare neurogenetic disorder with documented cognitive, behavioral and physical manifestations. The prevalence is typically reported as 1 per 20,000 to 50,000 live births, although a recent article suggests 1 per 7,500 (Greenberg, 1990; Stromme, Bjornstad, & Ramstad, 2002). Cognitive manifestations include intellectual delay (typically within the mild to moderate range) and a well-reported "syndrome-specific" profile with strengths in certain verbal abilities and poor visual construction and spatial skills, but good face processing (Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000; Galaburda, Wang, Bellugi, & Rossen, 1994; Lenhoff, Wang, Greenberg, & Bellugi, 1997; Wang, Doherty, Rourke, & Bellugi, 1995). Behaviorally, individuals with WS are said to be very sociable and often experience anxiety, phobias, obsessions, social disinhibition, attention difficulties, and poor emotion regulation (Bellugi, Adolphs, Cassady, & Chiles, 1999; Dykens, 2003; Einfeld, Tonge, & Florio, 1997; Gosch & Pankau, 1997; Greer, Brown, Pai, Choudry, & Klein, 1997). Physical features of people with WS include dysmorphic facial features, heart murmur, and often vascular anomalies, growth retardation, hoarse voice, hyperacusis, premature aging, and often infantile hypercalcemia (Williams, Barrat-Boyes, & Lowe, 1961). Physical features vary among individuals with WS (Borg, Delhanty, & Baraitserer, 1995).

WS is associated with a microdeletion of the elastin and contiguous genes on Chromosome 7q11.23. The typical deletion size is 1.5 Mb, but this varies, with reports of smaller, larger, and partial deletion (Borg et al., 1995; Fryssira et al., 1997). A percentage of WS individuals show inversion rather than deletion of the WS region (Osborne et al., 2001). Deletion of the centrally located elastin gene has been shown to be associated with the physical features of WS, especially supravalvar aortic stenosis (SVAS), but has no cognitive or behavioral manifestations (Lowery et al., 1995; Nickerson, Greenberg, Keating, McSaskill, & Shaffer, 1995). Certain genes are linked to the physical characteristics of WS, whereas others are said to have cognitive manifestations, such as LIMK1, which plays an important role in brain development. To date, 19 genes have been discovered within the WS deletion region, with ongoing discoveries.

Genetic testing for WS centers on the elastin gene, as this gene is reportedly deleted in 90% of cases (Borg et al., 1995; Fryssira et al., 1997). Fluorescent in situ hybridization (FISH) is used to look for a deletion at the elastin locus, but it will not detect inversion of the elastin gene (Fryssira et al., 1997). A person with a negative FISH result (i.e., no elastin gene deletion) may be diagnosed with WS on the basis of clinical features such as cardiac anomalies, epicanthal folds, facial dysmorphology and stellate, or "star-like," irises or both (McKusick, 1988; Morris & Sigman, 1988). Although the FISH test is used to investigate elastin gene deletion in WS, individuals who appear to have limited deletion of only the elastin gene do not manifest the cognitive phenotypes associated with WS and do not warrant a WS diagnosis. The previously mentioned information suggests the elastin gene

mutation is neither necessary nor sufficient for WS diagnosis. There are currently no routine clinical procedures to investigate the number of genes affected, which can differ across individuals with WS.

WS AND THE WS COGNITIVE PROFILE

Despite the existence of genetic heterogeneity in WS, it is frequently reported that there is a Williams Syndrome Cognitive Profile (WSCP; Mervis, Robinson, Bertrand, & Morris, 2000; Pinker, 1999), with claims of "a syndrome specific pattern of cognitive, spatial, linguistic, and academic functioning" (Howlin, Davies, & Udwin, 1998, p. 183), "Performance IQ scores … lower than Verbal IQ scores" (Howlin et al., 1998, p. 183), "a cognitive phenotype" in WS (Udwin & Yule, 1991), "a general consensus … that overall their verbal abilities are markedly superior to their visuospatial and motor skills" (Udwin & Yule, 1991, p. 233), a "unique neurobehavioral profile" (Wang, Hesselink, Jeringan, Doherty, & Bellugi, 1992, p. 1999), a "distinctive psychological profile" (Udwin, Davies, & Howlin, 1996, p. 1020), "an uneven cognitive-linguistic profile" (Rae et al., 1998, p. 33), "an uneven cognitive profile that is *a near universal feature* [italics added] of WS" (Karmiloff-Smith et al., 2003, p. 140), and a "syndrome cognitive profile" (Karmiloff-Smith, 2002; Karmiloff-Smith et al., 2003). In some instances, claims are made that "almost every person diagnosed with WS has the WSCP" (Tassabehgi et al., 1999, p. 125). For a formal definition of the WSCP, see Mervis et al. (2000); Tager-Flusberg (1999); and Bellugi, Lichtenberger, Mills, Galaburda, and Korenberg (1999). People with WS are claimed to show intact verbal skills and impaired spatial and visual construction abilities. Some authors acknowledge that not all individuals with WS display the WSCP (e.g., Mervis et al., 2000).

We now discuss research supporting a WSCP with strengths in verbal skills and a weakness in spatial and visual construction abilities. We challenge the focus on group means rather than individual scores. We also challenge the focus on task performance rather than on cognitive processes when describing the WSCP.

Research on the WSCP

Individual tests of cognitive abilities. Tests of confrontation naming and receptive vocabulary, such as the Boston Naming Test, the Peabody Picture Vocabulary Test–Revised (PPVT–R), the British Picture Vocabulary Test, and tests of verbal fluency are commonly used as measures of "language" in WS (V. Anderson, Northam, Hendy, & Wrennall, 2001; Bellugi, Bihrle, Jeringan, Trauner, & Doherty, 1990; Dunn & Dunn, 1981; Dunn, Dunn, Whetton, & Pintilie, 1982; Elliot, 1990; MacDonald & Roy, 1988; Mervis et al., 2000; Temple, Almazan, & Sherwood, 2002; Tyler et al., 1997). Tests of spatial and visual construction skills

within the WS literature typically include the Benton Facial Recognition test, the Visual Motor Integration Test, the Benton Line Orientation Test and Block Design (or block construction) from the Wechsler intelligence tests. Performance on these tasks is often used to support claims of the characteristic and uneven WSCP (Bellugi et al., 1990, 2000).

Some tests, such as block construction, measure numerous dissociable abilities. For example, block construction measures cognitive abilities, including perceptual abilities, spatial rotation and nonverbal reasoning. Nevertheless, failure on the block construction task in WS is commonly interpreted as evidence for a WSCP. Because block construction measures numerous dissociable abilities, individuals with WS may fail on the block construction task for different reasons. Therefore, it does not necessarily imply cognitive homogeneity if most individuals with WS fail this test. One WS individual may fail due to poor nonverbal reasoning, whereas another may fail due to perceptual impairments or spatial deficits.

Besides a strength in verbal skills and poor spatial and visual construction abilities, people with WS have been reported to show strengths in auditory processing (AUD; Don, 1999; Don, Schellenberg, & Rourke, 1999; Lenhoff, Perales, & Hickok, 2001; Levitin & Bellugi, 1998), nonverbal memory (Udwin & Yule, 1991), and verbal (or phonological) short-term memory (STM; Jarrold, Baddeley, & Hewes, 1999; Wang & Bellugi, 1994). People with WS are reported to show a weakness or abnormality in speed of processing (Farran & Jarrold, 2003; Howlin et al., 1998), semantic processing (Jarrold, Hartley, Phillips, & Baddeley, 2000; Tyler et al., 1997; Vicari, Carlesimo, Bizzolara, & Pezzini, 1996), and global form recognition (Bellugi, Sabo, & Vaid, 1988; Bihrle, 1990; Bihrle, Bellugi, Delis, & Marks, 1989).

A major problem with the previously mentioned research is that the majority of researchers have focused on group averages when describing strengths and weaknesses in WS, thus hiding individual variability. Also, one cannot describe the WS profile on the basis of individual tests, as some tests (e.g., tests of auditory processing) may be less difficult than other tests (e.g., block construction). Standardized test scores must be provided before one can directly compare performance across individual tests. Thus, it would be incorrect to generalize from the previously mentioned research and describe WS as showing a strength in auditory processing and a weakness on visual construction. Finally, failure on specific tests does not necessarily occur for similar reasons. Using individual test performance rather than cognitive processes to describe the WSCP may mask individual heterogeneity in cognitive strengths and weaknesses.

Standardized intelligence batteries. Some articles on cognitive functioning in WS report administration of an intelligence or cognitive battery where verbal and nonverbal tests are directly comparable in terms of task difficulty and normative data and where a combination of verbal and nonverbal tasks is sampled.

Unfortunately, whereas such studies overcame some of the difficulties associated with use of individual tests, many are not without faults.

Studies on WS have used standardized intelligence tests, such as versions of the Stanford–Binet Intelligence Scale, the Wechsler Intelligence Scale, the Differential Ability Scales (DAS), and the British Ability Scales (Greer et al., 1997; Howlin, Davies, & Udwin, 1996; Howlin et al., 1998; Jarrold, Baddeley, & Hewes, 1998; Mervis et al., 2000; Rae et al., 1998; Tassabehgi et al., 1999; Udwin et al., 1996; Udwin & Yule, 1991). These tests provide an overall level of functioning (such as an IQ) and various subscales, most typically a verbal and a nonverbal ability score, and some include other subscales such as reasoning (DAS, British Ability Scales, and the Stanford–Binet). More than one task is used to derive overall domain scores such as Performance IQ (PIQ) and Verbal IQ (VIQ).

Many researchers have tended to report averages (i.e., VIQ and PIQ) rather than scores on individual tests (e.g., Don et al., 1999; Udwin et al., 1996). This hides specific strengths and weaknesses within the verbal domain and specific strengths and weaknesses within the nonverbal domain. For example, Udwin et al. (1996) focused on IQ scores, yet these scores would be invalid following the presence of variability in the individual subtests used to derive performance (nonverbal) and verbal IQs. Language and spatial domains are divisible constructs, and it is possible that on certain tasks spatial abilities were above verbal abilities in WS.

Some authors have acknowledged that language and spatial domains are broad terms covering numerous dissociable abilities (Farran & Jarrold, 2003; Farran, Jarrold, & Gathercole, 2003; Karmiloff-Smith et al., 1997, 1998; Mervis & Robinson, 2000; Volterra, Capirci, Pezzini, Sabbadini, & Vicari, 1996). Such authors have documented an uneven profile, with some language abilities impaired and some nonverbal abilities intact. Nevertheless, many researchers still describe people with WS as showing a strength in verbal abilities and a weakness in nonverbal abilities.

Udwin and Yule (1991) did not investigate variability in nonverbal subtests within WS, but they did acknowledge differences in nonverbal abilities within WS at a more coarse level, with WS individuals showing poor performance (or nonverbal) IQ yet good nonverbal memory. Udwin and Yule (p. 241) asserted, "the present study goes on to suggest that WS children's nonverbal skills are not all equally depressed."

To their credit, some authors have published mean subscale scores (Arnold, Yule, & Martin, 1985; Don et al., 1999; Howlin et al., 1998; Jarrold et al., 1998; Udwin & Yule, 1991; Udwin, Yule, & Martin, 1987). Unfortunately, however, there is no mention of calculation of subtest scatter at an individual level, and interpretation is based on group averages rather than individual profiles. IQ scores should not be used for any individual where there is high variability across verbal subtest scores or nonverbal subtest scores at an individual level.

Use of intelligence batteries to provide support for a group WSCP is compromised by a further three factors: (a) Many intelligence batteries limit measures to spatial and verbal functioning when there are many other cognitive domains such as attention, perceptual abilities, memory, auditory processing, and reasoning; (b) In many instances, researchers used a verbal task, such as picture naming or receptive vocabulary, to match "verbal abilities" across WS and control groups (e.g., MacDonald & Roy, 1988). First, picture naming or receptive vocabulary is only one aspect of language; and second, verbal skills are an overestimation of general cognitive ability within this syndrome (Bellugi, Lichtenberger, et al., 1999). Accordingly, WS and control groups were perhaps not matched on verbal abilities or level of intellectual functioning after all. People with WS show a relative strength on picture naming tasks, so their overall IQ was likely to be lower, if anything, than the comparison group; (c) control groups have often consisted of individuals with mental retardation from a wide variety of etiologies, thus increasing the chance of finding more homogeneity in WS and more heterogeneity in the control group (MacDonald & Roy, 1988; Mervis et al., 2000; Tager-Flusberg, 1999; Udwin & Yule, 1991).

MacDonald and Roy's (1988) study is subject to all of the previously mentioned criticisms. MacDonald and Roy matched their WS group to a heterogeneous group of people with mental retardation on the basis of expressive vocabulary (PPVT–R). As we now know, this meant the WS group most likely had lower nonverbal skills and lower general level of intellect than the control group. MacDonald and Roy found their WS group performed most poorly on visual–motor tasks, a now reported feature of WS. However, their control group also performed most poorly on visual–motor tasks compared to other tasks (academic and simple motor tests); their mean *t* scores for visual–motor tasks was between 30 and 35. This suggests that the visual–motor tasks were the most difficult tests. Given that the WS group was likely to have lower nonverbal and general IQs than controls, it is not surprising that most people with WS performed, on average, at or below a *t* score of 10 on these tests. Scores were not discussed at an individual level.

Developmental differences in WSCP. Although the WSCP has been widely accepted, the applicability of this profile to younger WS individuals has been challenged. Again, however, this research focused on task performance rather than specific cognitive abilities and averaged scores across WS individuals.

Paterson, Brown, Gsodl, Johnson, and Karmiloff-Smith (1999) studied language abilities in young children with WS. They compared their results to findings from previous studies on language abilities in older WS children and WS adults. Because these latter studies indicated intact language abilities in WS, Paterson et al. claimed to demonstrate impaired language in young children with WS, but intact language in older WS children and WS adults (Paterson et al., 1999). They argued that language ability varies with development in WS. The difficulty here is that (a) different measures of language were compared, and (b) different people

were compared. It would be preferable to study the same WS individuals and to study language abilities within these individuals longitudinally.

Using the DAS, Jarrold et al. (1998), like Paterson et al. (1999), argued that there are different strengths and weaknesses in WS across development. They demonstrated that the discrepancy between DAS verbal and nonverbal abilities only occurred in older WS children. Difficulties with this argument arise, however, because although both the preschool and school-age DAS yield verbal and nonverbal ability scores, the preschool DAS does not measure the same skills as the school-age DAS. For example, vocabulary requires picture naming for the preschool DAS, but word definitions for the school-age DAS. Discrepancies between verbal and spatial abilities on the preschool DAS and the same discrepancies on the school-age DAS are, therefore, not directly comparable. Also, norms were not available for older WS children who, on the basis of their mental age (MA), were administered the preschool DAS (despite having a chronological age [CA] above preschool level).

Despite the methodological flaws, articles such as Jarrold et al. (1998) and Paterson et al. (1999) are significant, as they were the first to challenge the well-documented verbal–nonverbal discrepancy in WS, at least in younger individuals, but they highlight the need to understand exactly what cognitive functions a test measures and how directly comparable various tests and test scores are.

Single-case approach. A recently more popular trend in WS research has been to investigate people with WS at an individual rather than a group level (Jarrold et al., 1998; Nakamura et al., 1999; Pezzini, Vicari, Voltera, Milani, & Ossella, 1999; Tassabehgi et al., 1999; Thal, Bates, & Bellugi, 1989). When this approach is taken, suggestions of cognitive heterogeneity within the WS population emerge. For example, Pezzini et al. noted that 4 out of 18 individuals failed to demonstrate the "group" profile of good face processing, good verbal fluency, and poor spatial and constructional skills. In particular, one individual showed poor verbal fluency, whereas another showed better spatial construction than face recognition. Volterra, Longobardi, Pezzini, Vicari, and Antenore (1999) noted that their single case with WS was able to demonstrate good constructional and visuospatial abilities, the opposite to what the WSCP would predict. Similarly, Mervis et al. (2000) reported that 12% (10 out of 84) of their WS individuals did not fit the WSCP. Also Pani, Fonaryova Key, and Mervis (2000, p. 1) stated that visual constructional abilities in their WS sample were "better than predicted by previous estimates." Mervis, Morris, Bertrand, and Robinson (1999) found that 13% (12 out of 80) of their WS sample performed within normal limits on block construction.

The disadvantage of looking at task performance to define a syndrome-characteristic profile is demonstrated when we consider that the profile of good language and poor spatial and visual construction skills is said to be characteristic of other

syndrome profiles such as Turner syndrome, Nonverbal Learning Deficit, and Velocardiofacial/DiGeorge syndrome, despite chromosomal, genetic, and neurological differences across each syndrome (Bearden, 2002; Don et al., 1999; Rourke, 1988, 1989; Tager-Flusberg, 1999). Thus, the WSCP as it stands is not unique to the syndrome and may not be characteristic of all WS individuals.

This Study

As we have seen previously, the literature remains influenced by a syndrome-specific neuropsychological profile with strengths in verbal skills and a weakness in nonverbal abilities (Bellugi, Lichtenberger, et al., 1999; Howlin et al., 1998; Mervis et al., 2000). This is surprising given the genetic, physical, and clinical variability within this group (Borg et al., 1995; Fryssira et al., 1997; Pankau et al., 2001). The aim of this study was to investigate whether universal cognitive characteristics exist in WS by looking at a variety of tasks measuring long-term memory, immediate memory, processing speed, auditory processing, visual processing, comprehension, reasoning and expressive language.[1] This aim was pursued by using a single comprehensive cognitive battery where the same tests and test norms could be used across a wide range of MA and CA groups so that test scores were directly comparable.

Specifically, our aims were (a) to investigate whether there is within-syndrome variability in cognitive functions within WS and whether any cognitive abilities are homogeneous within WS and (b) to further investigate Jarrold et al's (1998) finding of different strengths and weaknesses across development by investigating whether there is a specific profile for low versus high mental age or low versus high chronological age. We aim to investigate whether there is a specific profile for low versus high MA or low versus high CA.

METHOD

Participants

Participants included 31 individuals with a formal clinical diagnosis of WS (15 males and 16 females). All participants were diagnosed independently by a minimum of two professionals (cardiologists, ophthalmologists, geneticists, pediatricians) based on a combination of unique facial, physical, and behavioral characteristics associated with the syndrome (McKusick, 1988; Morris & Sigman, 1988). Participants were recruited through the Williams Syndrome Association NSW and the Williams Syndrome Association SA. Of these individuals, 19 had had the ge-

[1]We did not wish to compare WS to other clinical populations. We were simply interested in cognitive heterogeneity and cognitive homogeneity within WS itself.

netic FISH test, with 18 out of 19 showing the elastin gene mutation. Twenty-nine percent were left-handed or ambidextrous, and 71% were right-handed. Interestingly, there was a substantial proportion of left-handed individuals with WS compared to the 5% to 10% within the normal population. Many right-handed participants also had left-handed or ambidextrous relatives. CA ranged from 5 years 4 months to 43 years 8 months. Mean CA was 16 years 11 months (*SD* = 9 years 4 months). Mental age (MA) was assessed on the Woodcock–Johnson Tests of Cognitive Ability–Revised or WJ–R COG (Woodcock & Johnson, 1989, 1990) and ranged from 3 years 3 months to 9 years 4 months. Mean MA was 5 years 7 months (*SD* = 1 year 4 months). Table 1 shows details of genetic testing, physical characteristics, development, and schooling and intervention for each participant.

One individual obtained a negative FISH test. This individual was included, as he was diagnosed by a cardiologist, a geneticist, and a pediatrician as having WS. He has SVAS (usually linked to the elastin gene), facial dysmorphology characteristic of WS, hyperacusis, and cognitive and behavioral features consistent with WS. Indeed, 10% of WS individuals do not have the elastin gene mutation (Lowery et al., 1995; Nickerson et al., 1995). It is possible that this individual has inversion rather than deletion of the elastin gene (Osborne et al., 2001), especially given his SVAS. The elastin gene has no cognitive manifestation, so absence of the elastin gene mutation should not affect this individual's cognitive profile in comparison to other WS individuals.

Materials

Participants were administered the Woodcock–Johnson Tests of Cognitive Ability–Revised or WJ–R COG (Woodcock & Johnson, 1989, 1990). A third version, Woodcock–Johnson III, has become available since the commencement of this study, although this version lacks the extensive research on reliability and validity that its predecessor has accumulated. The WJ–R COG was chosen for a variety of statistical, methodological, theoretical, and practical reasons. First, the WJ–R COG is widely acknowledged and widely used as a valid and reliable test of cognitive abilities. Also, the battery is theoretically based and is the most comprehensive measure of fluid or "innate" intelligence (Flanagan & McGrew, 1997; Woodcock & Mather, 1989, 1990). The WJ–R COG is based on the Horn–Cattell Gf–Gc theory proposing two types of intelligence called *fluid* (Gf) and *crystallized* (Gc) intelligence, or innate and learned intelligence (Horn & Noll, 1997; Woodcock, 1994). The WJ–R COG assesses nine Gf–Gc abilities.

Another reason for choosing to administer the WJ–R COG is that this battery includes tests of many abilities that are reported to be particular strengths or weaknesses in individuals with WS, such as speed of processing (Farran & Jarrold, 2003; Howlin et al., 1998), auditory processing (Don, 1999; Don et al., 1999; Lenhoff et al.,

TABLE 1
Description of Williams Syndrome Subgroups

Subgroup	*FISH*	*Hand*	*Hypercalcemia*	*SVAS*	*School*	*Intervention*	*Age at Diagnosis*	*Milestones*	*Hyperacusis*
1	na	R	N	N	IO		5 years	Delayed	Y
1	na	R	Y	Y	S		10 months	Delayed	Y
1	na	A	N	N	M	OT, ST	4 years	Delayed	Y
1	–ve	R	N	Y	IM	OT, ST	2 years	Delayed	Y
1	+ve	A	Y	N	M	ST	12 years	Delayed	Y
1	+ve	R	Y	N	M	ST, OT, PT	12 years	Delayed	Y
1	na	R	Y	Y	S	ST	7 months	Delayed	Y
1	na	R	N	Y	S		29 years	Delayed	N
1	na	R	N	N	S	ST	5 months	Delayed	N
2	+ve	L	N	Y	S	ST, PT	8 months	Delayed	Y
2	+ve	R	N	Y	S	OT	7 years	Delayed	Y
2	na	R	Y	Y	S		2 years	Delayed	Y
3	na	R	N	N	IO	OT, PT, ST	na	Delayed	Y
4	na	L	N	N	IM	PT, OT	5 years	Delayed	N
4	+ve	L	N	Y	M	ST, OT, PT	3 years	Delayed	Y
4	na	R	N	Y	M		30 years	Delayed	Y

4	+ve	R	N	Y	S	OT	18 months	Delayed	Y
4	+ve	R	Y	Y	M	OT	4 months	Delayed	Y
4	+ve	R	Y	Y	S		na	Delayed	Y
4	+ve	R	N	Y	S	ST, PT, OT	18 months	Delayed	Y
4	+ve	R	Y	Y	S		1 year	Delayed	Y
4	+ve	L	N	Y	S	ST	4 years	Delayed	Y
5	+ve	L	N	Y	M	OT, ST	4 months	Motor delayed	Y
6	+ve	R	Y	Y	M	OT, ST, PT	2 years	Delayed	Y
7	+ve	R	Y	Y	S	ST, OT	9 months	Delayed	Y
7	na	R	N	Y	M	ST, PT	6 months	Delayed	Y
8	na	R	Y	N	M	ST, OT	10 days	Delayed	Y
na	+ve	L	Y	Y	M	ST, OT, PT	18 months	Delayed	Y
na	+ve	L	N	Y	M	OT, PT	2 years	Delayed	Y
na	+ve	R	na	N	IM	ST, OT	3 years	Delayed	Y
na	+ve	R	N	N	S	ST, OT, PT	1 year	Delayed	Y

Note. FISH = fluorescent in situ hybridization; SVAS = supravalvar aortic stenosis; –ve = FISH test shows elastin gene present; +ve = FISH test shows elastin gene deletion; R = right; A = ambidextrous; L = left; N = no; Y = yes; S = special class or school; M = mainstream school; IM = integrated mainstream school; OT = occupational therapy; ST = speech therapy; PT = physiotherapy.

2001; Levitin & Bellugi, 1998), expressive and receptive vocabulary (Mervis & Robinson, 2000), semantic processing (Jarrold et al., 2000; Tyler et al., 1997; Vicari et al., 1996), global form recognition (Bellugi et al., 1988; Bihrle, 1990; Bihrle et al., 1989; Pani, Mervis, & Robinson, 1999), nonverbal memory (Udwin & Yule, 1991), and verbal STM (Jarrold et al., 1999; Wang & Bellugi, 1994). Also, the WJ–R COG includes measures of nonverbal abilities such as spatial processing, perceptual abilities, visuospatial STM, and nonverbal reasoning (independent processes required for good performance on the block construction task).

Norms exist for people aged 2 to 95 years, making the battery appropriate for all participants in this study, on the basis of both CA and MA, and enabling the possibility of longitudinal studies without requiring the use of different tests for children and adults. The battery includes 21 tests, 7 core and 14 supplemental. Tests 1 to 14 measure 7 cognitive factors and an oral language (LANG) factor (a total of 8 different cognitive factors, which are described later). The WJ–R COG is, therefore, more than a test of verbal and nonverbal abilities. The test measures short- and long-term memory, expressive and receptive language, fluid reasoning (FLUID R), visual and AUD, and speed of information processing (SIP). Mental-age equivalents are provided for both individual test performance and for cognitive factors, making WJ–R COG tests directly comparable to one another. Core and supplemental tests of the WJ–R COG are briefly described in the Appendix. Further details are provided in the WJ–R COG examiner's manual (Woodcock & Mather, 1989b, 1990b).

Tests 1 to 14 form seven cognitive factor scores: Long-Term Retrieval (LTRET, Tests 1 and 8), STM (Tests 2 and 9), SIP (Tests 3 and 10), Auditory Processing (Tests 4 and 11), Visual Processing (VIS, Tests 5 and 12), Comprehension–Knowledge (COMP, Tests 6 and 13) and FLUID R (Tests 7 and 14). Tests 15 to 21 supply additional information regarding each cluster. In addition, a LANG score is derived form Tests 2, 6, 13, 20, and 21.

Procedure

Tests were administered according to standardized instructions provided in the WJ–R COG examiner's manual (Woodcock & Mather, 1989b, 1990b). The battery was administered over two separate sessions ranging from 1 to 8 days apart. On average, the battery took 4 to 5 hr in total to administer. As recommended in the examiner's manual, the preschool core (Tests 1, 2, 4, 5, and 6) was administered to those participants whose MA fell below 4 years. These tests form an early development scale. This occurred for 3 individuals. Two participants with MAs above 4 were not administered the full test battery due to illness or misadventure.

Scoring. Tests were scored both manually and with the WJ–R COG computer scoring program. MA equivalents were obtained for each of the 21 tests, and MAs were obtained for each of the eight cognitive factors. A broad cognitive abil-

ity (BCA) score was also provided: This was the average MA based on standardized scores across all 21 tests. The computer scoring system calculated the number of standard error of estimate (SEE) units each cognitive factor score deviated from the average level of cognitive ability based on the participant's MAs for the other six cognitive factors (excluding the LANG factor). SEE was estimated from the WJ–R COG normative population.

RESULTS

Aim 1: Heterogeneity

Figure 1 shows box plots including each person's performance on all 21 tests. Box plots show the median, range, and extreme values for each group on a single cognitive test. The box represents the interquartile range that contains 50% of values; the whiskers are lines that extend above and below the box to represent the highest and lowest values, and the line across the box indicates the median. Open circles repre-

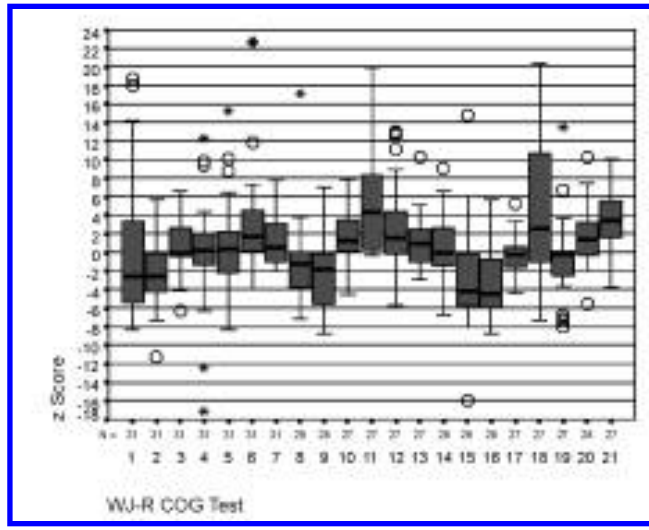

FIGURE 1 Each individual's z score for the 21 tests. *Note.* Test 1 = Memory for Names; Test 2 = Memory for Sentences; Test 3 = Visual Matching; Test 4 = Incomplete Words; Test 5 = Visual Closure; Test 6= Picture Vocabulary; Test 7 = Analysis-Synthesis; Test 8= Visual-Auditory Learning; Test 9 = Memory for Words; Test 10 = Cross Out; Test 11 = Sound Blending; Test 12 = Picture Recognition; Test 13 = Oral Vocabulary; Test 14 = Concept Formation; Test 15 = Delayed Recall- Memory for Names; Test 16 = Delayed Recall- Visual-Auditory Learning; Test 17 = Numbers Reversed; Test 18 = Sound Patterns; Test 19 = Spatial Relations; Test 20 = Listening Comprehension; Test 21 = Verbal Analogies.

sent outliers (> 1.5 and < 3 interquartile ranges from median), and asterisks represent very extreme values (> 3 interquartile ranges from median). Scores represent their MA on that test minus their BCA score, divided by their standard error of test scores. This represents the equivalent of a *z* score showing deviation from their BCA. Accordingly, any score with an absolute value of 1.96 or greater represents a significant deviation from that person's BCA. Figure 1 highlights the extreme variability in cognitive strengths and weaknesses across WS individuals.

In addition, Figure 1 shows there are only two tests with reasonable homogeneity across all WS individuals—Test 4 (Incomplete Words), a measure of phonological processing, and Test 17 (Numbers Reversed), a test of phonological STM. For Test 4 and Test 17, 50% of WS individuals performed at BCA (or average MA) levels (between –1.96 and 1.96). Even within Tests 4 and 17 there is some variability, with some WS individuals performing significantly above BCA (> 1.96), and some significantly below BCA levels (< –1.96) on these tests.

Figure 1 suggests that overall our WS sample showed strengths on the following tests: Picture Vocabulary (Test 6); Sound Blending (Test 11); Picture Recognition (Test 12); Sound Patterns (Test 18), Listening Comprehension (Test 20), and Verbal Analogies (Test 21). All these medians were above 1.96. This is consistent with previous research that suggests strengths in auditory processing and verbal abilities (at least expressive vocabulary and language comprehension) in WS when group means are used (Don, 1999; Don et al., 1999; Lenhoff et al., 2001; Levitin & Bellugi, 1998; Levitin et al., 2003; Mervis & Robinson, 2000). Results also show that visual perception is a median strength in WS, consistent with claims of good face processing in WS (Rossen, Jones, & Bellugi, 1995). Figure 1 shows that overall weaknesses are found on Memory for Names (Test 1), two tests of verbal STM (Tests 2 and 9) and two tests of long-term memory (Tests 15 and 16). Although there are consistencies between our findings and previous research looking at mean cognitive abilities within a sample of WS individuals, Figure 1 suggests extreme variability in test scores across our sample, thus making average scores meaningless. For example, Figure 1 shows that whereas the median score for Test 18 (a test of auditory processing) is above BCA (or average MA) level, some individuals perform at their BCA level, and some significantly below their BCA level on this test. Not all individuals with WS show a strength in auditory processing. Figure 1 also shows extreme heterogeneity in verbal abilities (see Tests 2, 6, 13, 20, and 21) and in nonverbal abilities (see Tests 3, 5, 7, 10, 12, 14, and 19).

Thus, there is neither a homogeneous strength in verbal abilities nor a homogeneous weakness in nonverbal abilities within our WS sample. We found substantial within-syndrome variability in cognitive functions within WS and no convincing evidence of homogeneous abilities in any test within WS. The most homogeneous tests were a measure of phonological processing (Test 4) and a measure of phonological STM (Test 17). The data suggest scores should be investigated at an individual level rather than looking at group means and medians.

It is interesting to note that no individual performed significantly below BCA levels on Test 7 (Analysis–Synthesis, a measure of nonverbal reasoning) and only one individual performed below BCA levels on Test 20 (Listening Comprehension). This suggests it is unlikely that individuals with WS will show a weakness on Test 7 or Test 20 of the WJ–R COG.

Cognitive domains versus individual test scores. Figure 1 shows variability in tests used to derive cognitive factor scores. For example, Tests 1 (Memory for Names) and 8 (Visual–Auditory Learning) measure LTRET (memory). Although the median for Test 1 is significantly below BCA level, the median for Test 8 is at BCA level. Similarly, whereas both Tests 4 (Incomplete Words) and Test 11 (Sound Blending) measure auditory processing, the median for Test 4 is at BCA, yet the median for Test 11 is significantly above BCA. If both tests assess common auditory processes, the medians should be similar.

Aim 2: Do Cognitive Strengths and Weaknesses Differ for Low Versus High MA and CA?

Apart from investigating heterogeneity, a second aim was to determine whether cognitive strengths and weaknesses differ in WS depending on whether an individual shows high versus low MA or high versus low CA. To investigate whether cognitive strengths and weaknesses relate to MA, we split the WS group into two—those at or below the sample median MA of 5.5 years and those with an MA above the sample median of 5.5 years. A multivariate analysis of variance with group as the between-subject factor (two levels: Group 1—those at or below our sample median MA and Group 2—those above our sample median MA) and the 21 WJ–R COG test scores as dependent variables indicated significant differences across groups for Test 7 or Analysis–Synthesis $F(1, 25) = 12.34$, $p < .01$, and Test 21 or Verbal Analogies, $F(1, 25) = 7.49$, $p = .01$. All other comparisons failed to reach significance ($p > .1$ for all other comparisons). These results remain significant with a strict alpha level of .01 to control for Type 1 error. The previously mentioned analyses suggest that Test 7 (a test of nonverbal reasoning) and Test 21 (a test of verbal analogies) are strengths for the lower MA group, but they are at BCA or MA levels for the higher MA group.

To investigate whether cognitive strengths and weaknesses in WS change with CA, we split the WS group into three: Group 1—those with a CA less than 7 years, Group 2—those with a CA between 7 and 10 years, and Group 3—those with a CA above 10 years. With a strict alpha level of .01 to control for Type 1 error, a multiple analysis of variance with group as the between-subject factor (3 levels) and the 21 test scores as dependent variables showed no significant differences across groups on any of the 21 tests.

One might suggest that the variability in cognitive strengths and weaknesses within our WS sample could reflect the fact that some individuals have a positive FISH result, whereas others have not had the FISH test, and one individual shows a negative FISH result. There is, however, the same variability when only those individuals with a positive FISH result are examined.

Investigation of Possible WS Subgroups

Principal component analyses (PCAs), multidimensional scaling (MDS), and hierarchical cluster analyses (HCAs) were used in combination as a preliminary investigation into whether subgroups of WS exist based on their profile across the 21 tests from the WJ–R COG. Analyses were undertaken using SPSS. These analyses were used to group individuals according to their similarity across the 21 tests. PCA and MDS are graphical analyses, whereas cluster analysis uses proximity data but is not a graphical technique. Whereas PCA and MDS used the first two dimensions to visualize all the interrelations between the data, the cluster analysis used all 20 dimensions to find subgroups within the data. It is often useful to superimpose the cluster solution onto the graphical solution to identify subgroups, particularly because cluster analyses inevitably produce clusters. PCA and MSA, therefore, provide further support for clusters. PCA, MSA, and HCA methods were used, as the data consisted of a set of 21 numerical test scores for each of the participants. Brief details of the three techniques are given in the following sections.

PCA—Principal component extraction with no rotation. Principal component scores were produced for the first two components. Limiting the solution to two components allows an ordination graph to be produced (Manly, 1994). These two components accounted for 37% of the variation. A plot (ordination) of the first two components was produced for the 27 subjects who completed each of the 21 tests.

MDS—A nonmetric MDS using Euclidean distances. A nonmetric MDS using Euclidean distances was conducted on the 27 subjects. Both three-dimensional and two-dimensional solutions were produced. The two-dimensional solution was interpreted provided the stress value was below .15 (Kruskal & Wish, 1978). For each MDS solution, the two-dimensional output was appropriate to interpret.

HCA—Hierarchical cluster analysis. HCA was employed starting with each participant in a cluster of their own. Clustering continued until all subjects were in one cluster. A dendrogram was produced for interpretation (Lorr, 1983). The cluster analysis method used was average linkage with squared Euclidean dis-

tance measures. The dendrogram was inspected for clear groupings that agreed with the visual clusters of the ordinations.

Subgroups were identified using these three previously mentioned techniques. Only the 27 participants with scores for each of the 21 tests were analyzed. Table 2 shows the sample size, BCA range, and CA range for each subgroup. To be in a subgroup, evidence from all three techniques was employed. In addition, individual profile plots showing the profile of scores across the 21 tests for each participant was inspected to verify the subgroups. Individual subjects who did not cluster with any other subjects were put into subgroups of $n = 1$. Although these individuals are not really subgroups, we used this term for simplicity to describe the eight groups. There are possibly three, but at least two, plausible subgroups. Table 1 shows details for each individual within a subgroup, including dominant hand, details of schooling and early intervention, details of FISH analyses, age at diagnosis, and presence of physical symptoms, including hypercalcemia, SVAS, and hyperacusis.

Varying Strengths and Weaknesses Across Subgroups

MDS, PCA, and HCA tell us how to form groups, but do not show how the various subgroups differ in terms of their strengths and weaknesses, either across tests or across cognitive factors.

Test scores across subgroups. Permutation tests (M. J. Anderson, 2001; Edgington, 1995), analogous to one-way analyses of variance, were performed to compare test scores across subgroups. At the individual test level, permutation tests showed significant differences across subgroups for Visual Matching (Test 3, $p = .000$), Cross Out (Test 10, $p = .05$), Picture Recognition (Test 12, $p = .05$), Oral

TABLE 2
Sample Size, Broad Cognitive Ability Range, and Chronological Age Range for Each Williams Syndrome Subgroup

Subgroup	*n*	*BCA Range (Years:Months)*	*CA Range (Years:Months)*
1	9	5:3–8:1	9:4–34:2
2	3	5:3–5:4	18:7–20:4
3	1	4:1	17:1
4	9	4:0–6:7	6:0–43:8
5	1	5:11	6:8
6	1	6:4	10:8
7	2	4:0	7:1–8:4
8	1	9:4	16:1

Note. Only individuals administered the full battery are represented in Table 2. BCA = broad cognitive ability; CA = chronological age.

Vocabulary (Test 13, $p = .03$), and Delayed Recall Visual–Auditory Learning (Test 16, $p = .001$). Marginally significant differences were found for Visual–Auditory Learning (Test 8, $p = .08$), Memory for Words (Test 9, $p = .09$), and Sound Blending (Test 11, $p = .08$). For all other tests, $p > .1$.

Table 3 shows how each subgroup differs significantly in its strengths and weaknesses across the WJ–R COG tests. Table 3 illustrates that no two subgroups show the same pattern of strengths and weaknesses across tests.

Cognitive factor scores across subgroups. Cognitive factor scores represent SEE deviation scores. Permutation tests (M. J. Anderson, 2001; Edgington, 1995) were performed to compare cognitive factor scores across subgroups. These tests indicated significant differences across subgroups for LTRET ($p = .003$), STM ($p = .02$), SPI ($p = .0001$), AUD ($p = .04$), COMP ($p = .00006$), FLUID R ($p = .00008$), and LANG ($p = .003$). No significant difference was found for VIS across subgroups ($p = .17$). Table 4 shows how no two subgroups show the same pattern of strengths and weaknesses across cognitive factor scores.

As indicated previously, however, Figure 1 demonstrates that cognitive factor scores should not be used to describe various cognitive strengths and weaknesses in WS; instead, individual test profiles are more appropriate. Figure 1 shows that the tests used to make up a cognitive factor score are measuring different processes.

Accordingly, Table 3 is the appropriate summary of each subgroup's profile. Subgroup 1 shows strengths on Visual Matching (Test 3), Cross Out (Test 10), and Picture Recognition (Test 12), and a weakness on Delayed Recall–Visual–Auditory Learning (Test 16). Subgroup 2 shows strengths on Visual Matching (Test 3) and Cross Out (Test 10) and a weakness on Delayed Recall–Visual–Auditory

TABLE 3
Strengths and Weaknesses for Williams Syndrome Subgroups: Individual Tests

Subgroup	*n*	*Test 3*	*Test 10*	*Test 12*	*Test 13*	*Test 16*
1	9	S	S	S	—	W
2	3	S	S	—	—	W
3	1	—	S	S	S	—
4	9	—	—	—	S	W
5	1	W	—	S	—	W
6	1	—	—	—	—	—
7	2	—	—	S	S	—
8	1	—	S	—	W	S

Note. An em dash represents broad cognitive ability (mental age) level. S = strength; W = weakness; Test 3 = Visual Matching; Test 10 = Cross Out; Test 12 = Picture Recognition; Test 13 = Oral Vocabulary; Test 16 = Delayed Recall: Visual–Auditory Learning.

TABLE 4
Strengths and Weaknesses for Williams Syndrome Subgroups: Cognitive Factors

Subgroup	*n*	*LTRET*	*STM*	*SIP*	*AUD*	*VIS*	*COMP*	*LANG*	*FLUID R*
1	9	S	—	W	S	—	—	W	S
2	3	S	S	W	S	—	—	W	S
3	1	S	S	W	W	—	—	W	S
4	9	S	—	W	S	—	S	—	S
5	1	—	—	—	S	—	S	—	—
6	1	—	—	W	S	—	S	—	—
7	2	—	—	W	S	—	S	—	S
8	1	S	S	—	S	—	W	W	—

Note. An em dash represents broad cognitive ability (mental age) level. LTRET = long-term retrieval; STM = short-term memory; SIP = processing speed; AUD = auditory processing; VIS = visual processing; COMP = comprehension–knowledge; LANG = oral language; FLUID R = fluid reasoning; S = strength; W = weakness.

Learning (Test 16). Subgroup 8 (one individual) demonstrates strengths in Cross Out (Test 10) and Delayed Recall–Visual–Auditory Learning (Test 16) and a weakness in Oral Vocabulary (Test 13).

In summary, there is preliminary evidence to suggest the possibility of subgrouping WS individuals on the basis of cognitive strengths and weaknesses. To ensure subgroups are meaningful, however, subsequent research should demonstrate differences across these subgroups on measures that were not entered as variables within the cluster analyses. PCA and MSA were used in addition to cluster analyses to determine subgroups, which adds more weight to the possibility that these subgroups are in some way meaningful.

DISCUSSION

The goal of this study was to assess whether individuals with WS show variability in cognitive strengths and weaknesses and to investigate whether any cognitive strengths or weaknesses appear homogeneous in WS. Past research tended to focus on specific task performance and on group averages, thus hiding individual variability in cognitive processes. A second aim was to determine whether cognitive strengths and weaknesses differ for individuals with high versus low MA or individuals with high versus low CA. First, results indicated heterogeneity in cognitive strengths and weaknesses within WS, thus highlighting the danger of looking at group averages when studying people with WS. We also undertook exploratory analyses (HCA, PCA, and MSA) to investigate the possibility of subgrouping individuals with WS on the basis of cognitive strengths

and weaknesses. Second, the most homogeneous skills in WS appeared to be phonological processing and phonological STM, with 50% of our sample showing scores at BCA level on these tests. Also, it was unlikely for individuals within our sample to show a weakness on Test 7 (Analysis–Synthesis) or Test 20 (Verbal Comprehension). Third, strengths on Analysis–Synthesis (Test 7) and Verbal Analogies (Test 21) were found for WS individuals with an MA at or below the sample median of 5.5 years; individuals with a high MA (above the sample median of 5.5 years) did not demonstrate strengths on these tests, but rather performed at BCA (or average MA) levels on Tests 7 and 21. This suggests a relation between strengths on Tests 7 and 21 and MA.

Within Syndrome: Variability in Cognitive Functions

Our results evidenced great variability in the cognitive strengths and weaknesses across WS individuals, even when only those with a positive FISH result were investigated. All WS individuals within our sample displayed the characteristic "uneven" pattern of abilities associated with the syndrome, but their strengths and weaknesses differed considerably.

This finding of cognitive heterogeneity in WS is consistent with the wide range of day-to-day abilities, personalities, and medical characteristics apparent when interacting with these individuals and their parents. Results are also commensurate with reports of varying genetic patterns, varying physical features, and clinical variability within the syndrome (Borg et al., 1995; Fryssira et al., 1997; Pankau et al., 2001).

No evidence of homogeneous strengths and weaknesses in WS. Although as we discussed earlier there is widespread support in the literature for a single WSCP, our results are inconsistent with such a claim. First, people with WS did not consistently show strengths in verbal abilities when language was broken down into expressive vocabulary, receptive vocabulary, phonological STM, language comprehension, and verbal analogies. Second, WS individuals showed a range of strengths and weaknesses on nonverbal abilities, including those cognitive processes required when undertaking the block construction task. This suggests individuals with WS may fail the block construction task for different reasons. Although many individuals with WS find the block construction test difficult, it is an oversimplification to suggest homogeneous cognitive weakness in WS on the basis of poor performance on this task. Spatial abilities were measured independently from perceptual abilities, eye–hand coordination, visual construction, and nonverbal reasoning. In certain instances, tests of verbal abilities were impaired, and nonverbal skills were found to be a strength—the exact opposite to the WSCP. In other cases, performance on tests of language and nonverbal (e.g., spatial) functions were both commensurate with BCA (or average MA) expecta-

tions, a result also inconsistent with the claimed WSCP or higher verbal than performance (nonverbal) IQs.

Although a lot of research supporting a WSCP involves performance on the block construction task, our study did not include a measure of block construction. This is because we believe block construction measures numerous dissociable cognitive abilities and that failure on this task can occur for numerous reasons. Our research confirmed heterogeneity in our WS sample on those cognitive processes required when completing the block construction task. Although failure on the block construction task itself may be common among individuals with WS, this does not necessarily imply homogeneous cognitive weaknesses.

Also, although previous research suggests a strength in auditory processing within WS, and although the overall WS group means on Tests 11 (Sound Blending) and 18 (Sound Patterns) indicated an overall strength in auditory processing within WS, some individuals performed at or below BCA expectations on these tests. This indicates no strength in auditory processing for some individuals with WS and that auditory processing is a specific weakness for other WS individuals.

Similarly, although people with WS are reported to perform poorly on Coding, a measure of SIP) from the Wechsler intelligence tests (Farran & Jarrold, 2003; Howlin et al., 1998; Wechsler, 1991), we did not find that all individuals within our WS sample performed poorly on measures of SIP (Tests 3 and 10). Moreover, median scores were at BCA level for both Tests 3 and 10, suggesting no median weakness in SIP for our WS sample. Tests 3 and 10, unlike Coding, do not require complex visual–motor skills. Although Coding is a measure of speed of processing, Coding also requires participants to draw complex symbols and thus measures visual–motor skills in addition to SIP. Because people with WS are reported to have difficulty with drawing and fine-motor control, it is likely that their poor performance on Coding actually reflects impairment in drawing rather than slowed speed of processing. For this reason the Coding test is not a valid measure of SIP for individuals with WS. Our results suggest there is variability in SIP within WS, and SIP is not a specific weakness in all WS individuals.

Similarly, there was variability on Test 19, a measure of spatial processing (without a visual constructional component), suggesting not all individuals with WS show a weakness in spatial skills as some suggest (Bellugi et al., 1988; Levitin et al., 2003; Paul, Stiles, Passarotti, Bavar, & Bellugi, 2002; Wang et al., 1995). Moreover, the median score did not suggest a weakness. The median score was at BCA (or average MA) levels. There was also variability on Test 5 (a measure of global form recognition). Although some past researchers suggest poor global processing in WS (Bihrle et al., 1989), our research suggests good global processing within WS in line with other studies (Pani et al., 1999; Tager-Flusberg, Plesa-Skwerer, Faja, & Joseph, 2003). Tager-Flusberg, Plesa-Skwerer, Faja, and Joseph recently found intact global processing of faces within WS. Many individuals performed at BCA level on Test 5, and some per-

formed significantly above BCA levels, indicating a strength in global form recognition in some WS individuals.

Scores on Test 6 (Picture Vocabulary) indicated considerable variability in expressive vocabulary within WS. Nevertheless, people with WS are often reported to perform well on tests of expressive vocabulary (see earlier mention). The median score for Test 6 approached 1.96, suggesting a significant overall strength. This explains previous reports of a strength in expressive vocabulary in WS, because past research often focused on WS group averages.

Homogeneity in Some Cognitive Abilities

Interestingly, no individual performed significantly below BCA levels on Test 7 (Analysis–Synthesis, a measure of nonverbal reasoning), and only one individual performed below BCA levels on Test 20 (Listening Comprehension). This suggests it is unlikely that individuals with WS will show a weakness in Analysis–Synthesis or Language Comprehension. These may or may not be universal and specific features of WS. Past research has shown strengths in reasoning skills (Greer et al., 1997) and verbal comprehension (Bellugi, Wang, & Jeringan, 1994) within WS.

Although there was variability across all 21 tests of the WJ–R COG, the least variability was found for Tests 4 (Incomplete Words) and 17 (Numbers Reversed). For Tests 4 and 17, 50% of WS individuals performed at the level of their BCA (or average MA), suggesting neither a significant strength nor a significant weakness on the processes assessed by these tests (i.e., phonological processing and phonological STM or verbal working memory). In line with our findings, Menghini, Verucci, and Vicari (2004) found that their sample of 16 individuals with WS showed mean reading abilities consistent with normal MA matched controls. Similarly, Majerus, Barisnikov, Vuillemin, Poncelet, and van der Linden (2003) found no strength in phonological processing abilities within their sample of four WS individuals.

Is There a Specific Profile for High Versus Low MA or High Versus Low CA?

Results indicated an association between low MA and strengths on Test 7 (Analysis–Synthesis) and Test 21 (Verbal Analogies). More specifically, Test 7 and Test 21 scores were a significant strength compared to BCA for WS individuals, with BCAs (or average MAs) at or below the sample median (5.5 years or below). Test 7 and Test 21 scores were at BCA levels for WS individuals, with MAs above the sample median (i.e., above 5.5 years). This suggests that nonverbal reasoning and verbal analogies may reach a maximum level of around the equivalent of skills seen in normal healthy 5.5- to 9-year-olds. No WS individual

showed adult levels of reasoning ability or verbal analogies. Thus, for individuals with high overall MAs, such as 8 years, their scores on tests of nonverbal reasoning and verbal analogies may be at ceiling for the WS population. The finding that certain cognitive strengths and weaknesses vary with MA is somewhat consistent with Jarrold et al (1998) who found evidence to suggest verbal abilities were a weakness early in development, but a strength in the later stages of development in WS.

We found no differences in cognitive strengths and weaknesses across young children, older children, or adults, suggesting that cognitive strengths and weaknesses assessed by the WJ–R COG do not vary with CA in WS.

Processes Versus Cognitive Domains

Our results support the argument outlined in the introduction that cognitive domains (such as WJ–R COG factor scores) are less valid measures than individual test scores when investigating cognitive functioning and cognitive profiles in people with WS. This is because cognitive domain scores are often averages of a number of tests, on which each individual with WS is likely to show extreme variability. In the past, the WSCP has been described in terms of spared and impaired cognitive domains—that is, spared language and face processing and impaired nonverbal abilities. In the introduction, we discussed the flaws associated with taking this approach, and our research findings also highlighted the danger in averaging across tests of specific cognitive domains for people with WS.

Although some researchers acknowledge that cognitive domain scores are invalid for people with developmental syndromes such as WS, many authors remain persistent in describing the WS profile in terms of strengths and weaknesses across cognitive domains. In line with some researchers (Farran & Jarrold, 2003; Farran et al., 2003; Karmiloff-Smith et al., 1997, 1998; Mervis & Robinson, 2000; Volterra et al., 1996), one should only speak of "peaks and valleys" across individual processes, and not across cognitive domains. Moreover, in the case of WS, one should only speak of peaks and valleys when referring to an individual's profile, as there are no universal strengths and weaknesses in cognitive functioning within WS.

Preliminary Suggestion of WS Subgroups

Using MDS, HCAs, and PCAs, we found preliminary evidence for subgroups of WS individuals. WS subgroups showed considerable homogeneity in cognitive strengths and weaknesses. Subgroup 1, for example, showed strengths on Tests 3 (Visual Matching), 10 (Cross Out), and 12 (Picture Recognition), and a weakness on Test 16 (Delayed Recall – Visual–Auditory Learning), suggesting good visual processing, fine motor skills, eye–hand coordination, spatial processing, attention, and nonverbal STM, and poor long-term memory. Subgroup 4 showed a strength

on Test 13 (Oral Vocabulary), suggesting good general knowledge, verbal comprehension, and semantic processing, but it showed a weakness on Test 16 suggesting poor long-term memory.

More important, members of each subgroup showed variability in both CA and BCA (or average MA). For example, the BCA (MA) of individuals in Subgroup 1 varied considerably from 5 years 3 months to 8 years 1 month. Similarly, BCA (or MA) ranged form 4 years to 6 years, 7 months for individuals in Subgroup 4. All individuals with a BCA (MA) less than 5.5 did not form one subgroup, or children did not form one subgroup and adults another. This suggests that specific profiles do not vary according to whether MA or CA is high or low.

WS subgroups and the WSCP. Subgroup 4 shows the pattern of strengths and weaknesses most similar to the WSCP. That is, Subgroup 4 shows a strength in verbal comprehension (Bellugi et al., 1994) and no strength on tests assessing spatial processing (Bellugi et al., 1988; Bihrle, 1990) or SIP (Farran & Jarrold, 2003; Howlin et al., 1998). Further investigation is required before we can say Subgroup 4 fits the description of typical strengths and weaknesses described within the WS literature. The WSCP, although perhaps sensitive in identifying some individuals with WS, is not specific to WS or characteristic of WS and may also lack the detail required to identify WS subgroups (should they exist).

It is interesting to note that of the tests that distinguish WS subgroups, many assess cognitive abilities described as characteristic strengths or weaknesses in WS. For example, Test 3 (Visual Matching) and Test 10 (Cross Out) measure SIP, a reported weakness in WS (Farran & Jarrold, 2003; Howlin et al., 1998). Test 10 also measures spatial processing, reported to be a weakness in WS (Bellugi et al., 1988). Test 12 (Picture Recognition) assesses visual perception and nonverbal working memory, reported to be a weakness in WS (Edgin, 2003), although Vicari, Bellucci, and Carlesimo (2003) reported that verbal working memory is at MA levels in WS, whereas spatial working memory is impaired. Test 13 (Oral Vocabulary) measures comprehension thought to be a strength in WS (Bellugi et al., 1994; Jarrold et al., 1999) and semantic processing, which is thought to be abnormal in WS (Jarrold et al., 2000). Test 16 (Delayed Recall–Visual–Auditory Learning) assesses long-term memory, a reported weakness in WS (Edgin, 2003).

We are currently undertaking research to investigate further differences between suggested subgroups within this study, particularly Subgroups 1 and 4 (the two largest subgroups). Measures incorporate tests of cognitive and socioemotion processing abilities, including local–global form perception, visual–motor integration, local bias in attention, emotion recognition, social approach, and theory of mind (understanding mental states such as beliefs and false beliefs). We are contin-

uing to find cognitive and socioemotion processing differences between Subgroups 1 and 4, even when WJ–R COG scores, MA and CA are covaried.

Genetic analyses of the WS individuals within this investigation may be able to determine exactly which genes are abnormal on the long arm of Chromosome 7 and whether there are deletions or inversions associated with the WS region. This will provide an opportunity to study relations between genes and cognition, especially if WS subgroups identified in this study prove to be meaningful. For example, it will be interesting to determine how many individuals in Subgroup 1 show a positive FISH result and whether there are genetic similarities across individuals within Subgroup 1 and genetic similarities among individuals within Subgroup 4. The most important analyses will be on genes apart from the elastin gene, since the elastin gene is said to show no cognitive or behavioral manifestations (Fryssira et al., 1997). It is known that genetic anomalies can be somewhat heterogeneous in WS, just as we have documented cognitive heterogeneity. Future research aiming to investigate gene–cognition relations must take a cognitive neuropsychological approach and focus on individual strengths and weaknesses and individually affected genes with sufficiently sized cohorts.

In conclusion, we have documented heterogeneity in cognitive strengths and weaknesses within a sample of WS people, along with preliminary evidence of subgroups of WS based on cognitive strengths and weaknesses. People with WS do not constitute a homogeneous population, and the claim that all exhibit intact language functioning with impaired nonverbal–spatial skills is not supported by the evidence. There is also variability in auditory processing within WS, despite claims that this is a strength in WS. Variability also arose in tests of global form perception, SIP, and semantic processing, all previously reported as a weakness in WS. The most homogeneity was found for Tests 4 and 17, measures of phonological processing and phonological STM. People with WS were unlikely to perform poorly on Test 7 (Analysis–Synthesis) or Test 20 (Verbal Comprehension). Cognitive profiles did not differ with CA or MA (BCA), but strengths on Tests 7 (Analysis–Synthesis) and 21 (Verbal Analogies) were related to MA. Strengths on Tests 7 and 21 occurred for WS individuals with MAs at or below 5.5 years, but not for individuals with an MA above 5.5 years. This suggests a limit to the maximum MA equivalent in nonverbal reasoning and verbal analogies. Future research aims to explore the possibility of subgrouping WS individuals by looking not only at cognitive abilities, but also at personality characteristics, physical anomalies, genetic abnormalities, socioemotion processing, and psychiatric disorders such as anxiety, obsessions, and phobias.

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APPENDIX
Woodcock–Johnson–Revised Test Descriptions

Test	*Description*
1: Memory for Names	Requires the participant to learn associations between unfamiliar auditory and visual stimuli. Measures learning and memory.
2: Memory for Sentences	Requires the participant to hold and repeat verbatim single words, phrases, and sentences presented on a tape recorder. Measures immediate or working memory, comprehension, and expressive language ability.
3: Visual Matching	Requires the participant to locate and circle two identical numbers within rows of six numbers for 3 min. Numbers begin as single digits and move to double, then triple digits toward the end. Measures speed of information processing and requires visual processing, intact fine motor skills, and eye–hand coordination.
4: Incomplete Words	Requires the participant to listen to words presented on a tape recorder. Each word is presented twice and has one or two phonemes missing. The participant is asked to produce the word without phonemes missing. Measures phonological awareness and auditory processing.
5: Visual Closure	Requires the participant to identify distorted pictures. Pictures may have areas missing, may be silhouettes, or may have a superimposed pattern. Measures perceptual processing and global form recognition.
6: Picture Vocabulary	This is a confrontation naming test and measures expressive language, general knowledge, and perceptual processing.
7: Analysis–Synthesis	Requires the participant to complete a logic puzzle by using the key provided. Measures nonverbal reasoning, the ability to monitor performance in response to feedback, working memory (or online problem solving), and learning.
8: Visual–Auditory Learning	Requires the participant to associate familiar words with an unfamiliar symbol, and then provide the appropriate word for a string of these symbols, which combine to make a sentence. Measures learning and memory, perceptual processing, and sustained attention. Test 8 is the first supplementary test.
9: Memory for Words	Requires the participant to repeat strings of unrelated words (from 1–8 words) verbatim. Measures attention and verbal working memory (ability to hold information in mind and then repeat the information verbatim).
10: Cross Out	Participants are given 3 min to visually scan and compare rows of 20 symbols and circle the five identical drawings within each row. Measures speed of processing and requires visual processing, spatial processing (as parts of a symbol may be included but rotated in distracters), and eye–hand coordination.

(*continued*)

APPENDIX *(Continued)*

Test	*Description*
11: Sound Blending	Requires the participant to process spoken words with syllables and/or phonemes missing and to repeat the word without any syllable or phoneme missing. Measures auditory processing and syllabic and phonological awareness.
12: Picture Recognition	Requires the participant to process pictures then recognize these pictures on a subsequent page when they are among similar distracters. Verbal mediation cannot be used because the stimuli and distracters have the same name. Measures visual perception, nonverbal working memory, attention, and speed of information processing (because stimulus presentation is limited to 5 sec).
13: Oral Vocabulary	There are two sections to this test. First, a word is spoken by the examiner and the participant must provide a synonym. Second, different words are provided and the participant must provide an antonym. Measures comprehension–knowledge and semantic processing.
14: Concept Formation	Involves learning concepts by viewing examples of the concept and noninstances of the concept. Feedback is provided for each response. Participants are not required to remember what happened over a series of items. Measures nonverbal reasoning and working memory.
15: Delayed Recall: Memory for Names	Requires the participant to recall the names learned 1 to 8 days prior in Test 1. It measures long-term verbal memory and retrieval and requires adequate learning during Test 1.
16: Delayed Recall: Visual–Auditory Learning	Requires the participant to recall the verbal association for pictorial symbols learned 1 to 8 days previously. It measures long-term retrieval and requires adequate learning during Test 8.
17: Numbers Reversed	The participant listens to a string of random numbers and must repeat the numbers in the reverse order. Items become more difficult as sequences become longer. Measures the ability to hold and manipulate information in mind or working memory.
18: Sound Patterns	Involves listening to a pair of complex sound patterns and judging whether the sounds are identical in pitch, rhythm, and sound content. Measures auditory processing.
19: Spatial Relations	Requires the participant to select which component parts are needed to make up a particular shape. The shapes are initially geometrical but become more organic or abstract as item difficulty increases. Measures spatial processing, integration of parts into a whole, nonverbal working memory, and nonverbal reasoning.
20: Listening Comprehension	Participants listen to tape recorded passages and supply the single missing word at the end of the passage. Passages increase in length. Measures comprehension, expressive vocabulary, semantic processing, and attention.
21: Verbal Analogies	Participants are asked to complete phrases using a single word that indicates an appropriate analogy. The relation among words becomes increasingly complex. Measures reasoning skills as well as comprehension and semantic processing.