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A Longitudinal Study of Cognitive Abilities in Williams Syndrome

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This longitudinal study assessed cognition in Williams syndrome (WS) over a 5 year period using the same test battery over the two occasions of testing. The aim was to explore whether absolute levels of ability and relative cognitive strengths and weaknesses remain consistent over time. 27 participants with WS were assessed using the Woodcock Johnson Test of Cognitive Ability – Revised (WJ-R COG, Woodcock & Johnson, 1989, 1990). Results suggested some developmental progress over time, but at a slower rate than typically developing peers. Cognitive strengths and weaknesses were consistent, at least on those abilities assessed using the WJ-R COG.

Williams syndrome is a genetic disorder with a cognitive phenotype characterized by global intellectual impairment (typically within the mild to moderate range), and peaks and valleys in more specific cognitive abilities (Bellugi, Mills, Jernigan, Hickok, & Galaburda, 1999; Mervis, Morris, Bertrand, & Robinson, 1999; Sigman, 1999; Woodcock & Johnson, 1989, 1990). Commonly reported strengths include: auditory processing, auditory short-term memory, and receptive vocabulary (Don, 1999; Jarrold, Baddeley, & Hewes, 1999; Mervis, Robinson, Bertrand, & Morris, 2000). Common cognitive impairments include: spatial processing, psychomotor skills, and processing speed (Howlin, Davies, & Udwin, 1998, Mervis et al., 2000). The majority of studies on cognitive functioning in WS have focused on a single “snapshot” in time; very few longitudinal studies of cognitive functioning in WS have been conducted. Consequently, it is unclear how various cognitive abilities change over time in this population and whether they follow the typical developmental trajectory. This study uses a longitudinal design to investigate whether cognitive ability, cognitive strengths and weaknesses, and general intellectual ability in WS are consistent over time.

Jarrold, Baddeley, Hewes, and Phillips (2001) examined whether the profile of verbal and non-verbal discrepancy in WS is consistent over time by exploring the development of vocabulary (using the British Picture vocabulary Test; Dunn, Dunn, Whetton, & Pintilie, 1982) and pattern construction (using the Differential Ability Scales, Elliot, 1990) in a longitudinal study of 15 children and adults with WS who were assessed on these tasks six times over a 40 month period. Jarrold et al. (2001) reported that mental age equivalent scores for vocabulary increased more rapidly than mental age equivalent scores for the pattern construction task over time, and concluded that vocabulary and pattern construction abilities develop at different rates in WS. They
suggested, based on their findings, that the profile of verbal and nonverbal discrepancy in WS becomes more apparent with development, with diverging verbal and nonverbal abilities in WS.

One other longitudinal study suggests that Full-Scale IQ, Verbal IQ, and Nonverbal IQ may change (or, more specifically, increase) over time in this population. In their longitudinal study, Udwin, Davies, and Howlin (1996) administered the Wechsler Intelligence Scale for Children–Revised (WISC–R) to 23 individuals with WS on an initial occasion and the Wechsler Adult Intelligence Scale–Revised (WAIS–R) at a 4 year follow-up and found a significant increase, on average, in Full-Scale IQ, Verbal IQ, and Nonverbal IQ scores over time. This suggests that the gap actually closes between WS individuals and their peers over time.

In contrast to the above studies, which suggest change over time in WS, three longitudinal case studies have been published, focused specifically on early language development in WS. Although these studies focus on only one or two select case studies, findings suggest that linguistic abilities in WS are consistently impaired over time (Capirci, Sabbadini, & Volterra, 1996; Levy, 2004; Stiles, Sabbadini, Capirci, & Volterra, 2000).

Thus, to date, very little longitudinal research on cognitive functioning in WS has been conducted, with some studies suggesting change over time (Jarrold et al., 2001; Udwin et al., 1996), consistent with cross-sectional studies that also indicate change in verbal and nonverbal abilities over time in WS (Paterson, Brown, Gsodl, Johnson, and Karmiloff-Smith, 1999; Vicari et al., 2004). In contrast, other longitudinal research suggests consistent cognitive abilities over time in WS, at least in terms of language skills (Capirci et al., 1996; Levy, 2004; Stiles et al., 2000). While the above studies have contributed greatly to our knowledge, some general methodological challenges relating to both cross-sectional and longitudinal research are worth considering, along with ways to enhance future work in the area, these include: (1) Use of the same tests and test batteries; (2) Use of standard scores to accurately assess developmental trajectories; and (3) Assessing a wide range of cognitive abilities. These methods are now discussed.

### USE OF THE SAME TESTS AND TEST BATTERIES

To date, little research has made cross sectional or longitudinal comparisons using the same cognitive tasks; an exception is Jarrold et al.’s work using the British Picture Vocabulary Test and the Pattern Construction task from the Differential Ability Scales (see above). While it is not always appropriate to use the same cognitive tasks, particularly when comparing young infants with older children and adults, where possible it is important that the same tests are used. Use of the same test at two time points, and ideally use of the same test battery where subtests are from a single test kit ensures: (a) that the same cognitive abilities are being tapped in both instances and (b) that the same normative reference group is used for each test (see also later). As Baron (2004) highlights, test scores are only comparable when a test is standardized on the same population sample, and scores “are not directly comparable when they are derived from different tests” (p. 78). This later point is also important for examining cognitive strengths and weaknesses. In order to evaluate strengths and weaknesses over time in WS, it is important that scores based on the same normative reference group are used, as otherwise scores are not comparable across tasks (Baron, 2004; Mervis & Klein-Tasman, 2004).
USE OF STANDARD SCORES TO APPROPRIATELY ASSESS DEVELOPMENTAL TRAJECTORIES

Within the literature, raw scores are most commonly reported, rather than standard scores. While these scores can be useful, a factor to take into consideration is that, from a longitudinal perspective, you would expect a greater increase in raw scores in a child that was younger at the time of first testing than for an older child who is closer to cognitive maturity. In contrast, standard scores provide a measure of how a participant compares to their age-matched peers and, in doing so, takes into account the different patterns of growth that are expected across different ages. Also, unlike raw scores and mental age equivalent scores, standard scores are relatively consistent across the lifespan, beginning from around 4 years of age (Sigelman & Rider, 2006; Weinert & Hany, 2003). For example, in their review, Sigelman and Rider assert that "even when several years have passed, IQ [a standard score] seems to be a stable attribute: the scores that children obtain at age 7 are clearly related to those they obtain 5 years later, at age 12" (p. 233). Longitudinally, a change in standard score over time, therefore, reflects a deviation from the normal developmental trajectory. Supplementary scores such as raw scores are valuable, as they assist in the interpretation of standard scores, such as whether there has been a loss in skills, a lack of progress (stagnation) or some progress or gain in skills.

Use of standard scores (as opposed to raw scores) is also important for examining cognitive strengths and weaknesses. In order to evaluate strengths and weaknesses over time in WS, it is important that standard scores (based on the same normative reference group) are used, as raw scores are not comparable across tasks or across different age groups (Baron, 2004; Mervis & Klein-Tasman, 2004).

ASSESSING A WIDE RANGE OF COGNITIVE ABILITIES

While most studies focus on verbal skills (in particular expressive vocabulary) and nonverbal skills (numeracy or spatial construction abilities), there are a wide range of skills relevant to the WS cognitive profile (Mervis et al., 2000) that are yet to be explored longitudinally, such as short-term memory, visual processing, and auditory processing. Longitudinal research should attempt to explore these areas of cognitive function.

OTHER METHODOLOGICAL CHALLENGES

Other methodological challenges include: the focus on group averages despite extensive cognitive heterogeneity in WS (Pezzini, Vicari, Volterra, Milani, & Ossella, 1999; Porter & Coltheart, 2005); the need to consider regression to the mean, the phenomenon whereby extreme scores are more likely to change over time (revert closer to the mean) than scores that are less extreme; and standard error of measurement (whether significant changes in scores are clinically meaningful or reflect statistical error).

Two of these challenges have also been acknowledged by previous researchers. For example, Jarrold et al. (2001, p. 430) note that “at the individual level, there is considerable variance in the rates at which vocabulary and pattern construction abilities develop.” And, in relation to regression to the mean, Udwin et al. (1996) acknowledged that participants in their study with
the lowest IQ scores at first assessment showed the greatest change in scores on the second assessment, suggesting a likely influence of regression to the mean. Regression to the mean may be a genuine change in some instances, but a statistical artifact in other instances.

AIMS AND HYPOTHESES

The current study aimed to address some of the methodological challenges associated with cognitive research and to systematically investigate whether cognitive abilities, cognitive strengths, and weaknesses and overall intellectual ability change over time in WS, using a longitudinal design. A single test battery that measured a wide range of abilities was conducted with thirty individuals with WS at two time points approximately 5 years apart. Standard scores were utilized to assess the developmental trajectory of various skills and the profile of skills over time. Raw scores were utilized to explore whether there was any loss, stagnation, or gain in absolute level of ability over time. Personalized z scores were utilized to explore individual cognitive profiles of strength and weakness over time. Specific hypotheses included: (1) That individuals with WS will follow a typical developmental trajectory over time, showing similar standard scores for general intellect and more specific cognitive abilities at time 1 and time 2; (2) That raw scores will indicate no loss of previously acquired skills; and (3) That personal profiles of cognitive strength and weakness will remain consistent in individuals with WS over time. Given that some research indicates a relationship between cognitive ability and chronological age in WS (e.g., Jarrold et al, 2001), we will also take chronological age into account when considering these questions. Findings are discussed in relation to practical and theoretical implications. While the discussion will focus on WS, wider implications are suggested in relation to other neurodevelopmental disorders.

METHOD

Participants

Thirty participants with WS were recruited through the Williams Syndrome Association, Australia to take part in this longitudinal study. Due to illness or misadventure, three individuals were unavailable for participation at the 5 year follow-up, leaving 27 participants in total (13 males and 14 females). The individuals who were unable to participate at time 2 were not atypical in terms of IQ or any specific cognitive ability when compared to the other WS individuals in the final cohort. Table 1 displays descriptive statistics for this cohort of 27 individuals, including chronological age, global IQ, and other more specific cognitive ability levels. There was an equal representation of children and adults in our cohort. Table 1 illustrates that our WS cohort is representative of the typical WS population, who are generally reported to display a mild to moderate intellectual disability, on average, and a relative strength in auditory processing, language, and short-term memory and weakness in spatial and psychomotor skills and processing speed. It should be noted that speed of processing tasks also assess spatial skills, a known weakness in WS (Bellugi et al., 1999; Mervis et al., 1999) (see see below).

All participants exhibited the medical and clinical phenotype associated with WS and genetic testing (a FISH test) confirmed the characteristic WS deletion (absence of one copy of the elastin gene on chromosome 7) in our participants (Fryssira et al., 1997).
TABLE 1
Descriptive Statistics for the WS Cohort and Change in Cognitive Factor Standard Scores Over Time

<table>
<thead>
<tr>
<th></th>
<th>Time 1</th>
<th>Time 2</th>
<th>Correlations Between Time 1 and Time 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chronological Age</td>
<td>16.16 (10.20), 5.00–44.67</td>
<td>21.92 (10.31), 10.75–50.16</td>
<td>.81**</td>
</tr>
<tr>
<td>IQ</td>
<td>44 (18), 16–78</td>
<td>47 (17), 13–76</td>
<td>.80**</td>
</tr>
<tr>
<td>Short-Term Memory</td>
<td>67 (12), 47–96</td>
<td>67 (11), 50–90</td>
<td>.81**</td>
</tr>
<tr>
<td>Processing Speed</td>
<td>43 (20), 12–89</td>
<td>45 (17), 18–78</td>
<td>.80**</td>
</tr>
<tr>
<td>Auditory Processing</td>
<td>74 (17), 27–102</td>
<td>79 (17), 46–111</td>
<td>.72**</td>
</tr>
<tr>
<td>Visual Processing</td>
<td>61 (17), 15–102</td>
<td>65 (17), 17–93</td>
<td>.70**</td>
</tr>
<tr>
<td>Comprehension-Knowledge</td>
<td>57 (18), 27–90</td>
<td>56 (15), 22–78</td>
<td>.80**</td>
</tr>
<tr>
<td>Fluid Reasoning</td>
<td>64 (13), 45–95</td>
<td>68 (15), 41–99</td>
<td>.80**</td>
</tr>
<tr>
<td>Oral Language</td>
<td>59 (15), 40–87</td>
<td>58 (15), 25–78</td>
<td>.89**</td>
</tr>
</tbody>
</table>

Chronological age is represented in years; IQ and factor scores are represented as standard scores, with a mean of 100 and a standard deviation of 15; NS = no significant difference from Time 1 to Time 2; ** = significant correlation at $p < .01$. There were no significant changes in standard scores or over time at $p < .01$.

The cohort was representative of the Australian population in terms of socioeconomic status (SES). SES was obtained using the Index of Relative Socio-Economic Advantage and Disadvantage (based on residential geographic area). The national average is 1,000 with a standard deviation of 100, and our sample displayed a mean of 1,010 and a standard deviation of 80 (range 788–1,139).


Participants were administered the Woodcock-Johnson Tests of Cognitive Ability–Revised or WJ–R COG (Woodcock & Johnson, 1989, 1990). The WJ–R COG is a widely used test battery with adequate psychometric properties (e.g., reliability coefficients and validity coefficients >0.60, with many coefficients as high as 0.80 and 0.90; Woodcock & Mather, 1989, 1990). Norms exist for persons aged 2 to 95 years of age, making the battery appropriate for all participants in the current study on the basis of both chronological age and mental age (IQ). These extensive norms also allowed us to use the same test battery for the initial and follow-up assessment, and minimized the likelihood of obtaining floor effects. Development of the WJ–R COG was theoretically driven; the battery 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 (e.g., see Horn & Noll, 1997). There are 21 tests in the WJ–R COG, 7 core and 14 supplemental. Further details of core and supplemental tests are provided in the WJ–R COG Examiner’s Manual (Woodcock & Mather, 1989,1990) and in the Appendix of Porter and Coltheart (2005). Performance on the seven core tests derives a general ability score, similar to “full-scale IQ” on the Wechsler tests, and, when the full battery is administered, eight cognitive factor scores are also available. Tests 1 to 14 form seven cognitive factor scores: Long-Term Retrieval (Tests 1: Memory for Names and Test 8: Visual-Auditory Learning), Short-Term Memory (Test 2: Memory for Sentences and 9: Memory for Words), Processing Speed (Test 3: Visual Matching and 10: Cross Out), Auditory Processing (Test 4: Incomplete Words and
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Procedure

For the purpose of this study, the full battery was administered at time 1, and the majority of tests were administered at time 2, approximately 5 years after the initial assessment. For the second administration, we chose not to administer three tests: Test 8 (Visual-Auditory Learning), Test 16 (Delayed Recall Visual-Auditory Learning), and Test 17 (Numbers Reversed), as these tests seemed time consuming and stressful for our WS participants on the initial assessment, and we felt we needed to keep the participants’ best interest at heart. This meant we were unable to obtain a Long-Term Retrieval factor score, although other tests were administered that assessed Long-Term Retrieval Ability—Test 1 (Memory for Names) and Test 15 (Delayed Recall—Memory for Names, see below).

Tests were administered according to standardized instructions provided in the WJ–R COG Examiner’s Manual (Woodcock & Mather, 1989,1990). The battery was administered over two separate sessions, no longer than one week apart. On average, the battery took 4 hours in total to administer (2 hours per session with breaks). Data was scored manually using the procedure outlined in the WJ–R COG Examiner’s Manual (Woodcock & Mather, 1989,1990) and was then checked using the computerized WJ–R COG scoring system.

RESULTS

We explored findings at a group level using standard scores and raw scores and at an individual level using personalized z scores. For group analyses, we firstly used standard scores to explore the developmental trajectory of WS participants over time. Here we were interested in whether our WS individuals showed a similar relative standing to their chronological age matched peers over time. That is, whether their level of impairment on specific cognitive functions and overall intellectual functioning, relative to their peers, was consistent over time. Second, we inspected raw scores to determine whether WS individuals displayed cognitive decline, cognitive stagnation, or cognitive progression over time in terms of their absolute level of ability. We then moved on to explore the relation between chronological age and change over time at the group level, to see, for example, whether younger WS participants were more likely to show a change over time. We then addressed whether any change could be accounted for by standard error of measurement alone. Finally, we were interested in exploring cognitive profiles over time at an individual level using personalized z scores. For this comparison, we were interested in whether each individual’s personal profile of relative strength and weakness remained similar over time.

For comparisons, WJ–R COG general cognitive ability (global IQ), cognitive factor scores (e.g., short-term memory, processing speed, auditory processing, visual processing, comprehension-knowledge, fluid reasoning, and oral language) and the WJ–R COG subtest scores were utilized. For raw scores, only subtest scores were reported, as there are no raw scores
for cognitive factors. Given multiple comparisons, we set the alpha level at .01 in order to help control for type 1 error.

The WS Group and Standardized Normative Data

**General IQ and cognitive factor scores.** Table 1 displays average WJ–R COG general IQ and cognitive factor standard scores for the WS cohort at time 1 and time 2. These standardized scores have a mean of 100 and a standard deviation of 15. A score more than two standard deviations below the normative reference group (70 or below) represents an impaired performance. Correlations between test scores at time 1 and time 2 are also shown in Table 1 and indicate significant moderate to high correlations.

In terms of general ability, Table 1 illustrates a mild to moderate intellectual disability, on average, both at the initial assessment, and at the 5 year follow-up. Similarly, a repeated measures *t*-test indicated no significant change in general ability level (IQ) from time 1 to time 2. This suggests a similar relative standing to their peers over time, on average, in terms of their general IQ (the gap does not widen or close).

For specific cognitive factors, Table 1 shows that, on average, WS individuals performed best on the auditory processing factor, both at time 1 and at time 2. Moreover, on both occasions of testing, average performance on this factor fell within two standard deviations of the mean performance of the normative reference group, suggesting that, on average, our WS cohort was not significantly impaired on this cognitive domain. The worst performance on cognitive factor score was noted for the processing speed factor, where, on average, the WS cohort performed three to four standard deviations below the mean of the normative reference group; again this was evident on both testing occasions. The speed of processing factor measures not only speed of information processing, but also psychomotor speed and spatial perception, so it is not unexpected that WS individuals performed so poorly on this factor. WS individuals were impaired (that is, performed greater than two standard deviations below the mean), on average, for all other cognitive factors (at both time 1 and time 2). Table 1 illustrates similar scores for each cognitive factor over time, and repeated measures *t*-tests also failed to indicate a significant difference for any cognitive factor score from time 1 to time 2, on average.

**Subtest scores.** The left hand side of Table 2 shows average WJ–R COG standard scores for the subtests used to comprise the WJ–R COG at time 1 and time 2; correlations between these scores at time 1 and time 2 are also shown. Although subtest scores are less reliable than cognitive factor scores (Woodcock & Mather, 1989, 1990), consistent with findings on the cognitive factor scores (above), overall, there are moderate to high correlations between standard scores at time 1 and standard scores at time 2 and results on individual subtest scores suggest a similar standing relative to the normative reference group (chronological age matched peers) over time, with no significant differences in test scores from time 1 to time 2 for the majority of subtests. The exceptions were Test 9 (Memory for Words) and Test 19 (Spatial Relations), where there was, on average, a significant increase in test scores over time. The size of this discrepancy, however, was not outside the standard error of measurement for the majority of individuals, with only 11% of participants showing a significant and meaningful increase on Test 9 and only 30% of participants on Test 19. “Significant and meaningful” means outside the standard error of measurement based on the participant’s chronological age.
## TABLE 2
Change in Subtest Standard Scores and Raw Scores Over Time

<table>
<thead>
<tr>
<th>Test</th>
<th>Standard Scores</th>
<th>Correlations Between Time 1 and Time 2</th>
<th>Raw Scores</th>
<th>Correlations Between Time 1 and Time 2</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Time 1</td>
<td>Time 2</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Test 1:</strong> Memory for Names</td>
<td>77 (12), 54–105</td>
<td>75 (11), 53–97</td>
<td>.43*</td>
<td>34 (10), 16–49</td>
</tr>
<tr>
<td></td>
<td>Test 2: Memory for sentences</td>
<td>71 (12), 34–96</td>
<td>67 (13), 30–86</td>
<td>.58*</td>
</tr>
<tr>
<td></td>
<td>Test 3: Visual Matching</td>
<td>46 (18), 10–82</td>
<td>51 (14), 25–73</td>
<td>.72**</td>
</tr>
<tr>
<td></td>
<td>Test 4: Incomplete Words</td>
<td>72 (14), 45–97</td>
<td>78 (16), 47–104</td>
<td>.78**</td>
</tr>
<tr>
<td></td>
<td>Test 5: Visual Closure</td>
<td>65 (16), 20–89</td>
<td>66 (19), 20–94</td>
<td>.62**</td>
</tr>
<tr>
<td></td>
<td>Test 6: Picture Vocabulary</td>
<td>65 (18), 35–98</td>
<td>65 (17), 23–86</td>
<td>.70**</td>
</tr>
<tr>
<td></td>
<td>Test 7: Analysis–Synthesis</td>
<td>66 (14), 48–100</td>
<td>71 (14), 51–110</td>
<td>.66**</td>
</tr>
<tr>
<td></td>
<td>Test 9: Memory for Words</td>
<td>70 (11), 47–98</td>
<td>75 (11)†, 59–103</td>
<td>.74**</td>
</tr>
<tr>
<td></td>
<td>Test 10: Cross Out</td>
<td>47 (25), 20–105</td>
<td>51 (15), 28–90</td>
<td>.60**</td>
</tr>
<tr>
<td></td>
<td>Test 11: Sound Blending</td>
<td>84 (15), 59–120</td>
<td>86 (19), 55–126</td>
<td>.60**</td>
</tr>
<tr>
<td></td>
<td>Test 12: Picture Recognition</td>
<td>72 (19), 40–122</td>
<td>79 (13), 49–104</td>
<td>.56**</td>
</tr>
<tr>
<td></td>
<td>Test 14: Concept Formation</td>
<td>70 (13), 49–99</td>
<td>71 (12), 48–93</td>
<td>.74**</td>
</tr>
<tr>
<td></td>
<td>Test 15 Delayed Recall—Test 1</td>
<td>68 (18), 14–103</td>
<td>64 (16), 17–93</td>
<td>.44</td>
</tr>
<tr>
<td></td>
<td>Test 16 Sound Patterns</td>
<td>78 (25), 15–128</td>
<td>81 (24), 45–131</td>
<td>.50*</td>
</tr>
<tr>
<td></td>
<td>Test 17 Spatial Relations</td>
<td>64 (15), 43–99</td>
<td>71 (13)†, 43–100</td>
<td>.68**</td>
</tr>
<tr>
<td></td>
<td>Test 18: Listening</td>
<td>66 (13), 44–89</td>
<td>68 (15), 47–97</td>
<td>.81**</td>
</tr>
<tr>
<td></td>
<td>Test 19: Verbal Analogies</td>
<td>70 (13), 58–103</td>
<td>71 (10), 56–90</td>
<td>.60**</td>
</tr>
</tbody>
</table>

** = correlation significant at p < .01; * = correlation significant at p < .05. Standard scores have a mean of 100 and a standard deviation of 15. There were no significant changes in standard scores or raw scores over time at p < .01 unless marked †.
Another consistency between cognitive factor standard scores and subtest standard scores arises when comparing Tables 1 and 2. Namely, just as the cognitive factor auditory processing is a relative strength, and, on average, is not impaired (see Table 1), so too are the subtests that comprise this domain—Test 4 (Incomplete Words) and Test 11 (Sound Blending) (see Table 2). Also, speed of processing is the weakest cognitive domain in Table 1, and so too are the subtests that comprise this domain—Test 3 (Visual Matching) and Test 10 (Cross Out), which both fall in the moderately impaired range (see Table 2). As with cognitive domains, at the subtest level, group cognitive profiles of strength and weakness are very similar at time 1 and time 2.

Overall, results suggest a similar standing relative to the normative reference group (chronological age matched peers) over time, both in terms of general intellect and in terms of more specific cognitive abilities. At a group level, cognitive profiles of strength and weakness appear to be very similar across time, both at the cognitive factor level and at the subtest level.

**Raw scores.** The right hand side of Table 2 shows average WJ–R COG raw scores for the subtests used to comprise the WJ–R COG at time 1 and time 2, correlations between these scores at time 1 and time 2 are also shown. Table 2 suggests a significant increase in raw scores for the majority of subtests (Exclusions include Tests: 1, 2, 10, 11, 15, 18, and 21 where there was no significant increase in raw scores over time; instead, scores at time 1 and time 2 were similar). There were also significant correlations between raw scores at time 1 and time 2 for the majority of subtests (Except subtests: 1, 5, 6, 7, 15, and 18, where there were low to moderate, but non-significant relationships). Table 1 suggests that, at the group level, there is progression for the majority of subtests (that is, some gain in raw scores), stagnation for certain subtests (no change in raw scores), but no decline or loss of skills, on average, for any subtest.

**The Relation Between Chronological Age and Cognitive Change**

The above analyses average across children and adults in our cohort. We were also interested to examine whether patterns of change are affected by the age of participants. For example, whether younger participants (children) with WS display more of a change in performance on the WJ–R COG over the 5 year period.

In order to explore the effect of chronological age on change in scores over time, we ran random intercept (mixed model) analyses centering chronological age at zero. Analyses were run on difference scores (score at time 2 – score at time 1) for cognitive factor standard scores, subtest standard scores, and raw scores. For each analysis, we ran the first model including both linear and quadratic relationships; quadratic relationships were evaluated along with linear relationships as it is possible that performance may plateau once a certain chronological age (or developmental level) has been reached. If the quadratic relationship was significant, no further analyses were conducted. If the quadratic relationship was not significant, we then removed the quadratic relationship and re-ran the analysis to explore linear relationships (Brown, & Prescott, 1962; Peugh & Enders, 2005).

**Standard scores and the relationship with chronological age.** Random intercept results for the cognitive factor standard scores are shown in Table 3 and indicate a significant linear relationship between chronological age and change in Comprehension/Knowledge scores and Fluid Reasoning scores over the 5 year period and a significant quadratic relationship between
chronological age and change in Oral Language scores over time (note this is perhaps marginal with an alpha level of .01).

Figure 1a illustrates the significant linear relationship between chronological age and change in Comprehension/Knowledge standard scores over time, and suggests that change in scores over time was proportional to chronological age. Younger participants were more likely to show a decrease in Comprehension/Knowledge standard scores over time and this increase was proportional to chronological age. Figure 1b shows a similar linear relationship between change in Fluid Reasoning standard scores over time and chronological age. Figure 1c illustrates the quadratic relationship between chronological age and change in Oral Language standard scores over time, similar to a linear relationship, younger participants were likely to show a decrease in Oral Language abilities over time and with age scores tended to increase over time. However, unlike the linear relationship, scores then seemed to plateau at around the age of 20 years.

At the subtest level, there was a significant linear relationships between change in standard scores and chronological age for: Test 13 (Oral Vocabulary, $F = 10.849, p = .004$, which feeds into the comprehension/knowledge cognitive factor), Test 14 (Concept Formation, $F = 9.134, p = .006$, which contributes to the fluid reasoning cognitive factor), and Test 21 (Verbal Analogies, $F = 8.648, p = .008$, which contributes to the oral language cognitive factor). All other analyses failed to reach significance. The linear trends were similar to those outlined in Figures 1a and 1b.

Was there a reliable change in standard scores over time? Because we knew that test/re-test reliability was lower for children than for adults (Woodcock & Mather, 1989, 1990), we needed to consider whether the general decrease in standard scores for the Comprehension/Knowledge, Fluid Reasoning, and Oral Language cognitive factors for the younger participants reflected real change or statistical error. To explore this, we referred to standard error of measurement scores in the WJ–R COG technical manual (these are based on an individual’s chronological age). We then calculated confidence intervals using these standard errors.

### TABLE 3
Change in Cognitive Factor Standard Scores Over Time With Chronological Age

<table>
<thead>
<tr>
<th>Cognitive Factors</th>
<th>Linear with Quadratic</th>
<th>Quadratic</th>
<th>Linear+</th>
<th>Significant Relationship With Chronological Age</th>
</tr>
</thead>
<tbody>
<tr>
<td>Short-Term Memory</td>
<td>$F = 0.055, p = .945$</td>
<td>$F = 0.488, p = .493$</td>
<td>$F = 0.500, p = .487$</td>
<td>NS</td>
</tr>
<tr>
<td>Processing Speed</td>
<td>$F = 3.548, p = .079$</td>
<td>$F = 1.053, p = .321$</td>
<td>$F = 3.064, p = .099$</td>
<td>NS</td>
</tr>
<tr>
<td>Auditory Processing</td>
<td>$F = 1.555, p = .228$</td>
<td>$F = 2.251, p = .150$</td>
<td>$F = 0.014, p = .905$</td>
<td>NS</td>
</tr>
<tr>
<td>Visual Processing</td>
<td>$F = 0.002, p = .967$</td>
<td>$F = 0.008, p = .931$</td>
<td>$F = 0.032, p = .859$</td>
<td>NS</td>
</tr>
<tr>
<td>Comprehension-Knowledge</td>
<td>$F = 16.547, p = .001$</td>
<td>$F = 5.312, p = .033$</td>
<td>$F = 10.809, p = .004$</td>
<td>Linear</td>
</tr>
<tr>
<td>Fluid Reasoning</td>
<td>$F = 12.792, p = .002$</td>
<td>$F = 2.189, p = .155$</td>
<td>$F = 14.245, p = .001$</td>
<td>Linear</td>
</tr>
<tr>
<td>Oral Language</td>
<td>$F = 24.298, p = .000$</td>
<td>$F = 7.792, p = .012$</td>
<td>—</td>
<td>Quadratic</td>
</tr>
</tbody>
</table>

+ The linear mixed model analysis was only performed if the quadratic relationship was not significant when the model included both the linear and the quadratic relationship. NS = not significant.
error of measurement scores, with each individual’s true score represented in error bands at time 1 and time 2. We found a true decrease in language capability (Comprehension/Knowledge and Oral Language) relative to peers for 54% of participants aged 17 years or younger, suggesting that half of the children displayed a true decrease in language abilities, relative to their peers, over time. None of the children showed a reliable increase over time. In contrast, only 10% of adults (participants aged 18 years and older) showed a reliable decrease in Comprehension/Knowledge and Oral Language test scores over time and 33% of adults showed a reliable increase in Comprehension/Knowledge and Oral Language scores over time. For Fluid Reasoning (and all other WJ–R COG abilities), 36%, or less, of children and adults showed a reliable change (increase or decrease in test scores) from time 1 to time 2. Of those children who did show a reliable change, particularly a decrease in Comprehension/Knowledge or Oral Language, it
is worth noting that there was a large age range and a wide ability range (from a moderate impairment to a low average performance at time 1), suggesting that patterns are not occurring only in very young children or only in children whose scores are at the higher extreme end (that is, patterns are not indicative of regression to the mean).

**Raw scores and the relationship with chronological age.** Random intercept analyses for raw subtest scores suggested significant linear relationships for: Tests 3 (Visual Matching, $F = 14.241, p = .002$); Test 12 (Picture Recognition, $F = 8.855, p = .007$); and Test 20 (Listening comprehension, $F = 27.395, p = .000$) and significant quadratic relationships were obtained for: Test 4 (Incomplete Words, $F = 10.196, p = .004$); Test 11 (Sound Blending, $F = 9.306, p = .007$); and Test 14 (Concept Formation, $F = 10.115, p = .005$). Linear relationships indicated a significant increase in raw scores over time with age. Quadratic relationships indicated a significant increase in raw scores over time with age and scores then seemed to plateau, with similar scores over time for older participants. These relationships are likely to, at least partially, reflect more change in younger participants, whose brains and cognitive capabilities are developing rapidly, compared to adult participants, whose brains and cognitive capabilities are more mature, and should, therefore, be more static.

**Was there a reliable change in raw scores over time?** Examination of children’s change in raw scores over time for the subtests that comprise Comprehension/Knowledge and Oral Language (Test 2, Test 6, Test 13, Test 20, and Test 21) tended to suggest an increase over time for the majority of participants. Moreover, 65% of children, on average, showed a significant and meaningful increase outside the range of standard error of measurement on these tests. This indicates that children are progressing on language subtests, but that they are progressing at a significantly slower rate compared to their peers.

**Personal Profiles of Strengths and Weaknesses: z Scores**

We now explore whether personal profiles of cognitive strength and weakness change over time in individuals with WS, first using cognitive factor scores and then using subtest scores. We calculated WJ–R COG $z$ scores for each individual, with a $z$ score $\geq 2$ representing a significant personal strength compared to overall cognitive ability (or IQ) and a $z$ score $\leq -2$ a significant personal weakness compared to overall ability (or IQ). The cutoff of 2 and –2 were chosen as they correspond to an alpha level of .05 and represent an unusual strength or weakness, which is only observed in approximately 2 to 5% of the population (Howell, 2007).

**Cognitive factors.** Figures 2a and 2b show box plots of $z$ scores for each of the cognitive factors at time 1 and time 2, respectively. $Z$ scores were calculated by subtracting the score on a specific cognitive domain from the individual’s general ability level and dividing this figure by the standard deviation (or variation) across the remaining cognitive factor scores. Comparison of Figures 2a and 2b show a similar performance at time 1 and time 2, with considerable overlap in the median score and range of cognitive factor scores over time.

Exploring cognitive factor $z$ scores further at the individual case level, we calculated the percentage of individuals who displayed a distinct difference in cognitive strengths and weaknesses on the WJ–R COG from time 1 to time 2. A “distinct” difference is defined as follows. A significant increase is defined as a score changing from the range $\leq -2$ (significant weakness relative
A LONGITUDINAL STUDY OF WS

FIGURE 2 Boxplots of z scores for each cognitive factor at time 1 and time 2.

Note: STM = Short-Term Memory; SIP = Processing Speed; AUD = Auditory Processing; COMP = Comprehension-Knowledge; VIS = Visual Processing; REAS = Fluid Reasoning; LANG = Oral Language.

Box plots show the median, range, and extreme values for each group on a single cognitive test. The box represents the inter-quartile range, which 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 represent outliers (>1.5 inter-quartile ranges from median).

A significant decrease is defined as a score changing from the range ≥2 (significantly above general IQ level) to <2 [at or significantly below (≤−2) general IQ level]. Very few individuals displayed a change in relative strengths and weaknesses over time, with no individual showing a change for Short Term Memory or Fluid Reasoning; only 4% showing a change for Processing Speed, Visual Processing, or Comprehension/Knowledge and 12% showing a change for Auditory Processing.
Of those few individuals that did show a change, it was not the case that individuals displaying a significant difference were those whose scores were extreme, and there was a wide range in SES, chronological age, and IQ across these individuals who did show change.

We should acknowledge that this process of determining a change in strength and weakness over time meant that some individuals may show a small quantitative change in relation to a significant qualitative change (e.g., a $z$ score change from 1.80—no personal strength or weakness at time 1, to 2.02—now a personal strength at time 2) and, similarly, some individuals may display a large quantitative change (e.g., from a $z$ score of 1.70 at time 1 to 0.69 at time 2) but no qualitative change. There were, however, only a few individuals who showed a small quantitative change (a change of less than 1 $z$ score) in the context of their significant qualitative change. Similarly, only a minority of individuals showed a large quantitative change (a change of more than 1 $z$ score) in the context of no qualitative change.

**Subtest scores.** Exploring subtest $z$ scores in the same way, Figures 2c and 2d show box plots including $z$ scores for each of the subtests at time 1 and time 2, respectively, and indicate similar profiles at time 1 and 2. Also, as with the cognitive factor scores, very few individuals displayed a change in relative strengths and weaknesses over time; the most change was observed for Test 20 (Listening Comprehension), where 20% of individuals showed a change (8% showing a significant increase (4% adults and 4% children) and 12% showing a significant decrease (all children); other change ranged from 15% for Test 18 (Sound Patterns) to 0% for Tests 3 (visual Matching), Test 10 (Cross Out), Test 13 (Oral vocabulary), and Test 14 (Concept Formation). Aside from Test 20 (Listening Comprehension), where it appeared that children were more likely to show a significant drop in relative scores compared to adults, there did not appear to be a relationship with chronological age for other changes, and for all tests, of those few individuals that did show a change, it was not the case that individuals displaying a significant difference were those whose scores were extreme, and there was a wide range in SES and IQ across these individuals who did show change.

Thus, examination of personal $z$ scores for both cognitive factor scores and subtest scores indicated that, overall, very few individuals showed a change in their personal profile of strengths and weaknesses over time; significant strengths at time 1 typically remained strengths at time 2 and significant weaknesses at time 1 typically remained weaknesses at time 2.

**DISCUSSION**

This detailed longitudinal study allowed us to track intellectual abilities and a wide range of cognitive skills over time in individuals with WS, and to explore patterns of change in these abilities, both in terms of absolute levels of ability and personal profiles of strength and weakness. Raw scores and standard scores were utilized, as well as subtest and cognitive factor scores and there was consistency across results.
There were three major findings from this study with regard to our research hypotheses. First, we found that, overall, intellectual and more specific cognitive abilities in WS followed the typical developmental trajectory. That is, the relative standing of WS individuals to their typically developing peers remained consistent in these areas. Second, raw score patterns over time suggested developmental progress, overall, with some subtest scores indicating stagnation, but no loss of previously acquired skills. Third, group profiles (measured using standard scores) and personal profiles of strength and weakness (measured using personalized $z$ scores in comparison to global IQ) remained consistent over time, both at the cognitive factor and the subtest level. One finding that partially failed to support our first hypothesis was the finding that language functions decreased over time (relative to peers) in a subset of children with WS, a finding that is also inconsistent with previous research suggesting consistent language skills over time in children with WS (Capirci et al., 1996; Levy, 2002; Stiles et al., 2000).

What Role Does Chronological Age Play in Language Ability Over Time?

Children were more likely to show a significant and reliable decrease in language skills over time compared to adults. Because standard scores take into consideration the fact that there is rapid development of vocabulary and general knowledge throughout childhood, whereas in adults, development of these skills tends to plateau, this developmental explanation cannot account for our findings (e.g., see Woodcock & Johnson, 1989, 1990). Also, it was not the case that individuals with extremely high language abilities at time 1 were those who were more likely to display a decrease in language abilities relative to peers, so our findings cannot be explained by regression to the mean.

Other possible explanations for this decrease over time in language skills for a subset of children in our cohort include: (1) characteristics of the language tasks themselves, compared to other tasks on the WJ–R COG and (2) development of executive functions. In terms of the former explanation, one possible reason for the decrease in oral language skills over time in children with WS is that Oral Language and Comprehension/Knowledge tasks on the WJ–R COG measure crystallized intelligence rather than fluid or innate intelligence, and are, therefore, more reliant on environmental factors than other WJ–R COG abilities (Horn & Noll, 1997; Woodcock & Johnson, 1989, 1990). For example, Oral Language and Comprehension/Knowledge measure vocabulary acquisition and general knowledge, and these skills develop rapidly throughout childhood with cultural and environmental exposure, such as reading text. It could be argued, for example, that WS children have less exposure to materials such as text (because many children with WS have impaired reading skills), so they do not develop vocabulary or general knowledge at the same rate as their peers. Therefore, the gap between WS children and their peers widens over time.

A second possible explanation is that the decrease in language abilities relative to peers reflects rapid maturation of the frontal lobes (and executive functions) in typically developing children during this period (Anderson, Northam, Hendy, & Wrennall, 2001) and slowed or abnormal frontal lobe development in WS. The subtests that comprise the Comprehension/Knowledge and Oral Language subtests rely heavily on higher-level frontal lobe functions such as verbal abstract reasoning, concept formation and semantics (Woodcock & Johnson, 1989, 1990), all...
abilities known to relate to frontal lobe or executive functions (Anderson et al., 2001). This may also account for our finding of a significant decrease in Fluid Reasoning scores for a subset of children, although in the majority of cases, this decrease in Fluid Reasoning represented statistical error rather than reliable change.

Practical Implications

First, findings have practical implications for families and professionals working with individuals with WS. Contrary to previous claims that one cannot reliably predict end states of cognition in WS based on early cognitive assessments (Vicari et al., 2004), the findings from the current study seem to suggest that one can make somewhat accurate predictions regarding long-term cognitive capabilities of persons with WS (at least from age 5 years and onwards, the youngest age of children in the current study). This is a question of obvious importance to families and also professionals interacting with these individuals.

Furthermore, findings may also lead to thoughts about possible ways to minimize environmental influences on cognitive impairment, for example, through methods such as environmental modifications or early intervention that is targeted for specific deficits. For example, in the case of WS, the gap between WS individuals and their peers seems to widen over time in terms of vocabulary and general knowledge skills. As mentioned earlier, this may be due to a lack of exposure to text as a result of reading difficulties. One way to assist here would be to have WS children listen to books on tape to try and enhance their vocabulary and their general knowledge acquisition. Of course, this environmental modification must be trialed and empirically tested to determine its efficacy, but it serves as a good example of how environmental factors and early environmental modification of these factors might change a child’s developmental trajectory within the context of a developmental disorder.

Theoretical Implications

The findings of the current study also assist our understanding of theoretical issues, including whether or not individuals with WS follow the typical trajectory in terms of brain and cognitive development. To date, it remains debatable as to whether cognitive functioning is simply delayed in WS, or whether cognitive development is distinctly different in WS compared to the typical population, reflecting aberrant brain development. Some researchers suggest that cognitive development in WS follows the typical trajectory, but that it is delayed (Capirci et al., 1996; Levy, 2002; Stiles et al., 2000). In contrast, other researchers argue that people with WS follow an atypical developmental trajectory and that their cognitive development is “fundamentally distinct” to that of the typical population (Karmiloff-Smith, 1992; Paterson et al., 1999).

While cognitive development is clearly atypical in WS evident by the uneven profile of strengths and weaknesses, longitudinal examination of each cognitive domain on the WJ–R COG in isolation suggests some support that cognitive development may be delayed rather than atypical, at least for some cognitive abilities within this population. It is possible of course, that some cognitive domains follow a typical trajectory, while other cognitive skills follow an aberrant path of development. There is some suggestion that language development may, for example, follow an aberrant developmental pathway at least for a subset of children, based on our findings in the current study.
Limitations

Limitations of the current research include: a small sample size; a restricted age range (with no children younger than 5 years of age); and the use of a single cognitive battery. Future research should aim for a larger sample size, which would allow for the possibility of subgrouping participants into more restricted age ranges. Furthermore, while in our study we did not find evidence to suggest greater differences between younger versus older children over time, this may be due to our restricted age range or small sample size. The longitudinal assessment of younger children is, thus, important. Finally, it would be useful to replicate the present findings using a different cognitive battery and over a third time point. Other areas of interest for future research include whether social abilities in WS are consistent over time, in particular, face processing, Theory of Mind, and emotion recognition skills.

Conclusions

Although additional research is required, and these results need to be replicated, the current study generally indicates consistent neuropsychological profiles in WS over time from childhood onwards, at least on those abilities measured using the WJ–R COG. This is with the exception of a widening gap in language development between a subset of WS children and their peers. Thus, findings indicate that change over time in cognitive abilities is not seen in all children with WS and depends on the domain in question, raising intriguing questions as to why this might be the case. The article extends previous longitudinal research on WS by examining a wide range of cognitive abilities longitudinally in a relatively large sample of individuals.

REFERENCES


