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Word reading and reading-related skills in adolescents with Williams syndrome

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Background: Individuals with Williams syndrome have good spontaneous language despite low levels of intelligence. This study explores the relationship between intelligence, word decoding and reading-related skills in 20 individuals with Williams syndrome. **Methods:** In addition to the KBIT, the participants were administered standardized measures of reading, vocabulary, rapid naming, phonological skills and an experimental measure of rhyme judgement. **Results:** There was wide variability in the reading achievement among the individuals with WS. While some participants were unable to recognize letters of the alphabet, others scored within the normal range of tests of single word reading and decoding. Reading scores were correlated with intelligence as measured on KBIT matrices but not with the vocabulary measures. Reading also correlated with phonological awareness tasks yet, surprisingly, not with rapid naming. **Conclusion:** It is suggested that in individuals with retardation, intelligence rather than language and language-related skills predict achievements in word reading. **Keywords:** Intelligence, phonological processing, reading, reading disorder, Williams syndrome.

Williams syndrome (WS; Williams, Barratt-Boyes, & Lowe, 1961) is a rare autosomal genetic disorder (1:25,000 live births) characterized by typical facial dysmorphology, renal and cardiovascular anomalies, statural deficiencies, characteristic dental malformation and hypercalcemia (McKusick, 1988). Most cases of WS are sporadic (but see recent findings, Osborne et al., 2001). A microdeletion on chromosome 7q11.23 has been identified in 98% of the individuals with WS. The missing region typically includes the ELN gene, which is hypothesized to account for the vascular and connective tissue abnormalities (Ewart, Jin, Atkinson, Morris, & Keating, 1994). The other phenotypic characteristics are presumably linked to the adjacent 16 or more genes that are part of the standard deletion in WS (Mervis, Morris, Bertrand, & Robinson, 1999; Tassabehji et al. 1999).

IQ scores in individuals with WS typically range between 50 and 70. While verbal mental age is not at age level, it is typically higher than general IQ (Mervis et al., 1999). Detailed studies of children and adolescents with WS, ages 10–20, found them to be severely deficient on spatial tasks (Bellugi, Marks, Bihrle, & Sabo, 1993; Bellugi, Poizner, & Klima, 1989; Bihrle, 1990; Mervis et al., 1999). Deficits in the spatial domain are particularly evident in construction tasks such as the tasks in the VMI (Beery & Buktenica, 1967) and the DAS (Elliot, 1990). There is evidence to suggest that this deficit is of a specific nature and cannot be reduced to low-level vision problems (Atkinson et al., 2001), nor does it affect the perception of static displays (Dilks, Landau, Hoffman, & Siegfried, 2001; Hoffman & Landau, 2000).

Long-term memory is impaired in individuals with WS (Vicari, Brizzolara, Giovanni, Pezzini, & Volterra, 1996). However, face recognition, expressive syntax and vocabulary are relatively preserved (Mervis et al., 1999; Rossen, Jones, Wang, & Klima, 1994).

There is general consensus that language is an area of relative strength, although researchers differ in their assessment of detailed aspects of linguistic skills in WS (Clahsen & Almazan, 1998; Clahsen & Temple, in press; Karmiloff-Smith et al., 1997; Patterson, Brown, Gsödl, Johnson, & Karmiloff-Smith, 1999). Oral language skills in individuals with WS are generally good. Language is rich and fluent and phonology is very good. Auditory short-term memory as measured by digit span is typically within the normal range (Mervis et al., 1999). Verbal working memory is also an area of strength. Vocabulary, as measured by the Peabody Picture Vocabulary Test (PPVT), is often within the normal range despite an overall IQ that is below normal (Grant et al., 1997; Mervis et al., 1999). Morphology and syntax are relatively preserved (Clahsen & Almazan, 1998; Clahsen & Temple, 2003; Levy & Hermon, 2000; but see Karmiloff-Smith et al., 1997 and Volterra, Capirci, Pezzini, Sabbadini, & Vicari, 1996 for a different view). Research on adults with WS confirms many of the cognitive characteristics described for children with WS. Specifically, good oral language skills as well as overall intelligence scores are preserved into adulthood in WS (Howlin, Davies, & Udwin, 1998; Udwin, 1990; Udwin & Yule, 1991).

The majority of research on word decoding views reading as a language-related skill (e.g., Algeria & Morais, 1991; Frith, 1985; Frost, 1998; Share, 1995; Stuart & Coltheart, 1988; Wimmer, Landerl,

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Linortner, & Hummer, 1991 among many others, but see also Stein, 1991). This view predicts relatively good reading and reading-related phonological skills in the WS population. However, despite good, and at times age-equivalent, verbal skills, reading achievements of individuals with WS remain poor. Understanding this discrepancy is of particular theoretical interest.

Udwin, Yule, and Martin (1987) studied reading in 44 children aged 6–16 with WS. Twenty-two children obtained scores on the reading tests and 23 children obtained scores on the spelling tests. The readers had a mean chronological age of 12 years, with a mean age for Reading Accuracy of 7 years 10 months and for Reading Comprehension of 7 years 9 months. Performance on spelling was poorer than performance on reading, which may be due to the additional perceptual–motor skills involved in writing. Mervis (unpublished data, personal communication, Sept. 1999) found a very wide range of reading competencies with a mean performance at fourth grade level. In the group studied by Howlin and her colleagues (Howlin et al., 1998), whose average age was 26 years, mean IQ of 61, reading, spelling and arithmetic were generally at the 7–8 year level.

It is instructive to compare language-related competencies in children with WS with similar skills in typically developing children with reading difficulties. Auditory speech perception is within the normal range in disabled readers of normal intelligence (Stanovich, 1991). Under masking conditions, however, there is some indication of differences between normal and disabled readers in auditory speech perception (Bentin, Deutsch, & Liberman, 1990). Short-term memory as measured on the digit span, as well as long-term memory, is within the normal range (Stanovich, 1991 and many others). In contrast, disabled readers have a pronounced difficulty in word repetition, particularly with rare words or pseudo-words (Ellis, 1991).

Despite their normal digit span, poor readers perform less well than good readers on auditory short-term memory requiring phonological recoding, suggesting that working memory rather than storage is implicated. This deficit is seen in children before they enter school so it cannot be simply the product of their reading history (Ellis & Large, 1987; Mann & Ditunno, 1990). Furthermore, short-term memory as measured through repetition of non-words links it with vocabulary development in the school years in typically developing children as well as in poor readers (Gathercole & Baddley, 1990; Gathercole, 1990; Gathercole, Willis, Baddley, & Emslie, 1991; Snowling, 1987).

Numerous studies have reported that poor readers of normal intelligence are worse than good readers on rapid automatized naming (RAN) of numbers, letters and pictured objects (e.g., Denckla & Rudel, 1976; Mattis, French, & Rapin, 1985; Wolf, 1991). Naming difficulties do not appear to be primarily attributable to factors such as preprocessing of adjacent visual information or sequential scanning. Rather, such difficulties may be related to a temporal processing deficit evident in the processing of stimuli presented in rapid succession (Lovegrove, Martin, & Slaghuis, 1986; Merzenich et al., 1996; Wolff, Michel, & Ovrut, 1990). RAN combines the ability to connect the visual to the verbal with speed of processing and as such it turned out to be a major predictor of reading competence, accounting for variance beyond that accounted for by timed tests of discrete naming (Denckla & Cutting, 1999).

Of particular relevance to an understanding of reading difficulties in the general population are tasks requiring phonological awareness (PA). PA tasks require *explicit* awareness of the phonological structure of words (e.g., Morais & Kolinsky, 1995; Bretelson & de Gelder, 1990). They involve the ability to detect, isolate, delete and manipulate sub-word units, i.e., syllables or phonemes. (Examples of these PA tasks are given in the section on Methods.) These abilities develop during the early school years in typically developing children and some are evident even in preschoolers who cannot as yet read. Training studies suggest a causal relation between PA and reading acquisition, namely that they, in turn, mutually influence one another and become correlated (Ball & Blachman, 1991; Perfetti, Beck, Bell, & Hughes, 1987; Lundberg, Frost, & Peterson, 1988). This has been shown repeatedly for English as well as for other languages (Bentin, Hammer, & Cahan, 1991; Bentin & Leshem, 1993).

Rhyme detection and rhyme production are among the few PA tasks that typically developing preschoolers do well on. Views vary with respect to the relationship between these rhyme tasks and learning to read. Morais, Algeria, and Content (1987) argued that rhyme appreciation and manipulation did not require segmental analysis and consequently did not correlate with reading skills. Numerous studies support this contention. Recently Muter, Hulme, Snowling, and Taylor (1998) failed to find a correlation between rhyming skills in pre-readers and reading in either the first or the second year at school, while phoneme segmentation did correlate with reading acquisition.

There are, however, opposing findings. Bradley and Bryant (1985) argued that rhyme and alliteration were indirectly related to reading acquisition since they were the precursors to phonemic segmentation. However, Goswami (1986, 1988) and Bryant, MacLean, and Bradley (1990) argued that rhyme made a direct contribution to reading that was independent of phoneme segmentation, although the latter was enhanced by rhyme too. The present study tests participants' ability to detect rhymes in an implicit test of rhyme detection and examines its correlation with level of word reading.

Not much is known about PA in children whose IQs are below normal (Blanton, Semmel, & Rhodes, 1987). Gottardo and Rubin (1991) documented the apparent ability of children and adolescents with mental retardation to analyze words into syllables and phonemes. Rondal (1995) described a case of an adult with Down syndrome (DS) who learned to read and write with limited phonological awareness. Cossu and Marshall (1990) presented a case of a hyperlexic child who had excellent transcoding abilities but no phonemic awareness. Morais and Kolinsky (1995), however, rejected the case and argued that the data did not warrant such a conclusion.

Cossu, Rossini, and Marshal (1993a, 1993b) studied 10 children with DS and argued that the children were able to read despite poor performance on PA tasks. These claims raised numerous objections, including suggestions for alternative interpretations of the data (Bretelson, 1993; Byrne, 1993; Morton & Frith, 1993). Recently, Cupples and Iacono (2000) argued against the conclusion reached by Cossu et al. (1993). They studied 22 children with DS and found a clear association between PA and early oral reading abilities.

A recent study by Laing et al. (2001) investigated reading, spelling, memory and PA skills in individuals with WS and compared them to normal controls. Word reading level of the individuals with WS was low but better, on average, than comprehension. The findings suggest that there is a stronger relationship between phonological abilities and real word reading in the control group than in the WS group. Furthermore, these relationships became much weaker in the WS relative to the controls once general cognitive ability was controlled for.

These findings are important and constitute the first systematic study of word reading and reading related skills within a relatively young group of individuals with WS. However, there are a number of methodological problems with this study. Of the 15 individuals with WS who participated in the study, three did not score above floor level on the word-reading test. In other words, these were individuals who did not learn to read. These participants were included in the analyses of the group as a whole. Since altogether 15 participants were studied, these non-readers may have had a significant effect on the means reported for the group.

Clearly, the existence of such a large proportion of individuals who cannot read reflects reality in this population. Similar findings were reported in other studies as well (Udwin & Dennis, 1995; Howlin et al., 1998). In fact, we too found non-readers in the group we studied, although in a somewhat lower proportion (see Table 1 below). The question is whether individuals who cannot read at all should be included in a study that focuses on correlations between reading and reading-related skills in WS. In our view, if the focus is on the nature of reading in the WS population, non-readers should not be included in the calculations for the group.

The inclusion of non-readers in the study likewise affected the choice of controls. It lowered considerably the reading level of the group, resulting in a control group that included very young children (mean age of the controls was 6;9 years; range 5–9;2 years). One can imagine that the 5-year-olds among the controls were matched to the non-readers among the WS. Given what is known about mutual effects of reading and PA skills in the normal population, an individual match based on reading level between a retarded child who failed to acquire reading and a child who does not read because he is too young to have been exposed to reading, seems rather problematic.

A further issue to bear in mind concerns the age range of the participants with WS in Laing et al.'s study – 9 to 27;7 years. In view of what is clinically known about the time it takes for children with WS to learn to read, it is quite possible that the youngest children in that group were still in the early stages of

Table 1 Average standard scores and age-equivalencies

	All participants ($n = 20$)		Participants able to read ($n = 17$)	
	Standard score (s.d.)*	Age equivalent**	Standard score (s.d.)	Age equivalent
PPVT	76.20 (15.04)	9–6	76.59 (16.24)	9–4
Rhyme Task	84.06 (23.58)	na	92.83 (8.25)	na
K-BIT Vocabulary IQ	60.75 (15.51)	na	63.47 (15.11)	na
K-BIT Matrices IQ	59.55 (13.32)	na	61.88 (12.53)	na
K-BIT Composite IQ	57.05 (12.99)	na	59.47 (12.48)	na
WJ-R Letter-Word I.D.	67.55 (26.75)	8–6	77.24 (13.50)	9–6
WJ-R Word Attack	–	–	74.88 (13.88)	8–2
Rapid Object Naming	63.75 (11.80)	6–6	65.29 (12.18)	6–6
Elision	70.50 (12.97)	7–3	73.24 (12.11)	7–6
Segmenting Words	81.50 (11.01)	7–0	84.12 (9.72)	7–0
Segmenting Nonsense Words	77.00 (11.17)	7–0	80.88 (6.43)	7–0

*Mean standard score on each test, except on the Rhyme Task, which is the average percent correct.
**Age equivalent scores derived from mean raw score for each applicable test.
na = age equivalency score not available.

reading acquisition. Note that the authors do not report whether the three non-readers were the youngest children in the WS group. The group should therefore be considered mixed in more than one way – it included non-readers and it had individuals who were in the process of reading acquisition as well as others who have achieved their highest reading level.

In the current study some of the methodological problems with the Laing et al. (2001) study were taken into consideration. Our work concerns single word decoding and PA skills in individuals with WS who are beyond the age of reading acquisition. The focus is on ability to read non-words. While reading familiar words is of interest as well, ability to read non-words is the ultimate evidence that decoding has been mastered. The findings presented below will be confined to individuals who could score above floor level on the non-word reading test. Performance on PA tasks, general IQ, vocabulary, rapid object naming and rhyming skills were likewise assessed.

Method

Participants

Twenty young individuals with a positive FISH diagnosis of Williams syndrome (12 males, 8 females) participated in the study. The participants were recruited and tested in the course of the US National Williams Syndrome Association Family Conference, in Dearborn, Michigan, in July 2000. Ages ranged from 12;8 to 20;4, with a mean age of 16;5. Thus participants have all had a rather long exposure to reading and could be considered past the acquisition stage, yet they were young enough to have benefited from modern instruction methods.

Procedure

Each participant was given tests of intelligence, vocabulary, word reading and phonological awareness skills. Intelligence was tested using the Kaufman Brief Intelligence Test (KBIT; Kaufman & Kaufman, 1990), the PPVT-III Form A (Dunn & Dunn, 1997), two subtests from the Woodcock-Johnson Psycho-educational Battery – Revised (WJ; Woodcock & Johnson, 1990), and four subtests from the Comprehensive Test of Phonological Processing (CTOPP; Wagner, Torgesen, & Rashotte, 1999). An experimental measure of rhyme judgement was also administered. Participants completed this battery in a single testing session, lasting about 90 minutes. Three participants completed the KBIT and/or the PPVT with another research group at the conference, and their scores were obtained from them.

IQ test. The KBIT assesses general cognitive abilities and generates verbal, non-verbal, and composite domain scores. The verbal component consists of two subtests that require spoken responses. The first is a picture-naming task (KBIT Expr). The second is the Definitions subtest (KBIT Defs), in which the participant must determine a word given two clues: a brief definition of the word and a partial spelling of the word. Thus this subtest involves familiarity with the written form of words. KBIT non-verbal matrices (KBIT Mats) is composed of a set of visual stimuli, which vary from matching category and functionality of concrete objects, to completing dot patterns and solving abstract visual matrices (Kaufman & Kaufman, 1990). The KBIT correlates highly with the K-ABC full intelligence test as well as with the WISC-R and WAIS-R. KBIT is commonly used in the US. The choice of KBIT to assess intelligence is discussed further in the Discussion.

Language test. The PPVT is a test of receptive vocabulary (Dunn & Dunn, 1997), in which the subject must select the correct picture that matches the single word spoken aloud by the administrator, from a tableau of four pictures.

Reading tests. To assess basic reading skills, the Letter-Word Identification and Word Attack subtests of the WJ were administered. The Letter-Word Identification subtest (LWID) evaluates the participant's ability to identify isolated letters and single words. Words are presented in sequence with later words reflecting a decrease in relative frequency in printed English. The Word Attack (WordAttk) subtest evaluates the participant's ability to read non-words that obey the rules of English orthography and phonology (Woodcock & Johnson, 1990). Recall that our interest is in the participants' word decoding ability, hence WordAttk is the crucial test.

Rapid Object Naming (RON). This subtest, taken from the CTOPP, measures how quickly the participant can name a set of six object drawings arranged in random order. There are 36 items on each of two pages. Each item is repeated 6 times on each page, and order among items varies. Items are arranged in four rows of nine columns, and participants name the items in a standard left-to-right manner. Each page is timed separately, with the raw score equal to the sum of the times from the two trials (Wagner et al., 1999). Note that a lower score on this test indicates better performance. Reported correlations will therefore have a negative sign.

Phonological skills tests. To assess phonological processing abilities, we administered three subtests from the CTOPP: Elision, Segmenting Words (WordSeg), and Segmenting Non Words (NonSeg). The Elision subtest assesses the ability to imitate words and then repeat them with certain sound components removed. Initially, the sound components removed include one word of a compound (e.g., "*Say toothbrush without tooth*"). Later, individual phonemes are removed (e.g., "*Say bold without/b/*"). In all cases the resulting sound pattern is a well-formed frequent English word. The Segmenting Words and Segmenting Non-words subtests assess the ability to isolate and pronounce the separate phonemes of a given word or non-word, which the participant must first repeat.

Implicit rhyme judgment task. On this task, the experimenter spoke a CVC word and then played a tape recording of a voice speaking another CVC word. The experimenter awarded the 'man on the tape' a token when the word he spoke rhymed with the first word, yet without mentioning to the child that a rhyme was required in order to receive a token. After several demonstrations, there began a training phase (6 items) during which the participant determined whether 'the man on the tape' earned a token, and the experimenter agreed or disagreed with that judgment. At no time was the fact that 'the man on the tape' was rewarded for saying rhymes explained or discussed with the participant. When the participant spontaneously inferred this, the experimenter neither confirmed nor denied it. The notion of rhyme was meant to remain implicit throughout the test. After training, the participants were given 32 trials on which they were asked to determine whether 'the man on the tape' should be given a token. There was no feedback from the experimenter apart from general encouragement. The rhymes and foils were varied to cover the full vowel space as well as a range of final consonants.

Results

Table 1 presents the means (and standard deviations) of all standardized test scores, as well as the age equivalents that correspond to the mean raw score on each test. Note that one cannot derive separate standard scores for each of the subtests that make up the verbal component, although this would have been desirable given the fact that the topic of the current investigation is word reading. However, since raw scores are used to calculate correlations this problem is alleviated. Raw scores in each of the subtests comprising the KBIT are given in Appendix 1. Percent correct scores on the Rhyme Judgment task are included in Table 1.

Three of the participants were unable to identify correctly the letters of the alphabet on the LWID subtest. They also failed to achieve the basal on WordAttk, defined as being able to read the first nonword on the list. The middle columns in Table 1 present the data for the entire sample, while the right-hand columns present the data excluding the three participants who could not read. The correlational analyses presented in the tables below relate to the 17 readers among the participants.

Correlational analyses

The first set of analyses, presented in Table 2, explored relationships between the various measures in the 17 participants who could read. Raw scores on the standardized tests and on the Rhyme Judgment task were used in the correlational analyses. Pearson correlations coefficients were calculated and level of significance $p <. 01$ was used throughout.

Significant correlations were found between KBIT Defs and LWID. Since performance on the former benefited from familiarity with written words, this finding did not come as a surprise. Note, however, that Defs did not correlate significantly with WordAttk. In fact, there were no other significant correlations between the verbal subtests in the KBIT and any of the other measures. PPVT did not correlate with any of the reading measures either. The nonverbal KBIT Mats correlated with WordAttk and both LWID and WordAttk correlated with Elision. WordAttk also correlated with WordSeg. Neither RON nor Rhyme correlated with any of the other measures. In fact, all but one participant in the group of readers performed close to ceiling on the Rhyme test, correctly judging 27 or more of the 32 rhyme trials. In contrast, the three non-readers all performed at chance on this task.

Tables 3 and 4 present raw scores correlations when KBIT Expr and KBIT Defs are controlled for, respectively. While the correlations change in some of the details, the trends remain the same – LWID correlates with WordAttk, WordAttk correlates with at least one of the PA tasks, as does LWID. RON, Rhyme and PPVT do not correlate with any of the reading measures.

Table 5 gives raw scores correlations when KBIT Mats is controlled for. LWID and WordAttk are now correlated with each other as well as with Elision.

Participant profiles

To determine how well participants' reading scores compared to their general intelligence level, we explored the profiles of their standard score performance on the standardized tests. Readers as well as non-readers were included in this analysis since our focus now was on the profiles that characterize the participants' group as a whole, especially the way intelligence combines with other skills. Groups were created based on composite IQ: those below 50 (group A; $n = 8$), those between 50 and 70 (group B; $n = 9$), and those at or above 70 (group C, $n = 3$). While each of the administered tests allows for the computation of a standard score with a mean of 100 and standard deviation of 15, each differs in terms of the detail available at the extremes of the standard score range. For instance, the WJ-R has a floor standard score of zero, while the PPVT and KBIT each have floors of 40. To compensate for these variations, the mean standard scores were transformed to equivalent percentile rankings. Figure 1 shows the percentile ranking corresponding to the mean standard score for each group on the tests administered.

Unlike the classical WS profile (Mervis et al., 1999; Levy, in press b), the mean percentile of the eight participants with IQ below 50 on KBIT Verbal was not higher than their scores on KBIT Mats, nor was it higher than KBIT Composite. This is probably due to the fact that the non-readers were part of this group. Recalling that the verbal score on the KBIT includes Defs, which uses written words as cues, it is

Table 2 Full raw score correlations

	K-BIT Expr	K-BIT Defs	K-BIT Mats	PPVT	LWID	Word Attk	RON	Elision	Word Seg	Non Seg	Rhyme
Age	–.364	–.005	.065	.000	.145	.208	.038	.433	.077	.228	.119
K–BIT Expr	–	.078	.444	.497	.344	.429	–.492	.262	.404	.128	–.062
K–BIT Defs		–	.136	.307	**.663***	.503	–.223	.460	.055	.081	.133
K–BIT Mats			–	**.706***	.218	**.610***	–.574	.594	.390	.343	–.109
PPVT				–	.499	.508	–.520	.531	.203	.306	.012
LWID					–	**.694***	–.384	**.636***	.372	.201	.000
Word Attk						–	–.548	**.780***	**.677***	.509	.043
RON							–	–.291	–.110	.132	.100
Elision								–	**.674***	.594	–.144
Word Seg									–	**.706***	–.123
Non Seg										–	–.580

*$p < .01$.

expected that individuals who can't read will have lower scores on this measure. Notice that this group's scores on the PPVT were slightly elevated than Verbal Composite. The participants in this group were able to do some word segmentation, at least at the syllable level, and some could perform syllable-level elision. All but one of these participants performed at floor level on Rapid Object Naming (RON).[1]

The second group of participants, those with IQ scores between 50 and 70, exhibit reading abilities that were at their IQ level. Recall that we are concerned with WordAttk rather than with LWID. Performance on the RON subtest is still at floor level. There is an overall slight upward shift in performance on the segmentation tasks for this group compared to the lower IQ group.

In the third group (IQ scores 70 and higher), we found decoding abilities considerably higher than IQ level. In fact, this group's performance on LWID and WordAttk spans the low-average to average range of the WJ-R norms for these tests, as does their performance on all the PA tasks, most clearly on segmentation. Particularly interesting is the high score achieved in WordAttk. Contrary to the second group, the participants with higher IQs are better in the latter than in reading familiar words. RON, however, is still at floor level. Thus, RON, which was expected to correlate with reading, was as low in the high IQ, good reading group, as it was in the lower IQ groups.

Discussion

This study investigated single word reading skills in a group of adolescents with WS, whose IQ and vocabulary indicate that they are a typical group of individuals with WS, comparable to those included in other published work (e.g., Laing et al., 2001; Mervis et al., 1999; Udwin et al., 1987). The participants were all beyond the stage of reading acquisition. The focus was on decoding and thus the main concern was with ability to read non-words.

IQ was assessed on the KBIT, which has two subtests – Expressive vocabulary and Definitions – estimating verbal IQ, and one subtest – Matrices – estimating performance IQ. KBIT has been shown to correlate highly with full-scale IQ derived from the K-ABC and the Wechsler scales. The Definitions subtest of the KBIT offers as cues partial spelling of the word that the subject is asked about. One may therefore question the use of this subtest in a study that investigates reading. However, since the focus of the current study is on non-word reading we estimated that there would be no confounding between the two measures. The lack of correlation between

[1] Floor level for 12-year-olds (our youngest subject) on the RON is 96 seconds. Floor for 20-year-olds (our oldest subject) is 77 seconds.

Table 3 Raw score partial correlations controlling for KBIT expressive raw score

	K-BIT Mats	PPVT	LWID	Word Attk	RON	Elision	Word Seg	Non Seg	Rhyme
K–BIT Defs	.114	.310	**.679**	.521	−.213	.457	.026	.072	.138
K–BIT Mats	–	**.625**	.078	.518	−.456	.553	.257	.322	−.091
PPVT		–	.403	.376	−.365	.478	.003	.281	.050
LWID			–	**.644**	−.263	.603	.272	.169	.232
Word Attk				–	−.428	**.765**	.610	.507	.077
RON					–	−.193	.111	.226	.080
Elision						–	**.644**	.585	−.133
Word Seg							–	**.721**	−.107
Non Seg								–	−.050

*p <. 01.

Table 4 Partial raw score correlations controlling for KBIT definitions raw score

	K-BIT Mats	PPVT	LWID	Word Attk	RON	Elision	Word Seg	Non Seg	Rhyme
K–BIT Expr	.439	.498	.392	.452	−.489	.255	.402	.123	−.073
K–BIT Mats	–	**.705**	.173	**.632**	−.563	.605	.387	.337	−.130
PPVT		–	.415	.429	−.487	.461	.196	.296	−.030
LWID			–	.557	−.324	.498	.449	.198	−.118
Word Attk				–	−.517	**.714**	**.752**	.544	−.028
RON					–	−.218	−.101	.154	.134
Elision						–	**.732**	**.629**	−.233
Word Seg							–	**.705**	−.132
Non Seg								–	−.070

*p < .01.

Table 5 Partial raw score correlations controlling for KBIT matrices raw score

	K-BIT Defs	PPVT	LWID	Word Attk	RON	Elision	Word Seg	Non Seg	Rhyme
K-BIT Expr	.019	.288	.282	.221	−.323	−.004	.279	−.029	−.015
K-BIT Defs	–	.301	**.655**	.534	−.178	.476	.002	.037	.150
PPVT		–	.500	.137	−.198	.194	−.112	.095	.127
LWID			–	**.725**	−.324	**.645**	.320	.138	.025
Word Attk				–	−.305	**.654**	.602	.403	.139
RON					–	.076	.151	.428	.046
Elision						–	.597	.516	−.099
Word Seg							–	**.662**	−.088
Non Seg								–	−.022

KBIT Defs and WordAttk, seen in Tables 2 and 3, suggests that this assumption was correct.

Similar to previous work (Udwin et al., 1987; Laing et al. 2001), our findings revealed a wide range of reading achievements in the group. Among the 20 participants, three were not able to name some letters of the alphabet, nor were they able to read familiar or non-words. These individuals were excluded from the calculations of the group means since our interest was in the nature of reading skills in WS, rather than in the general profile of the entire group. At the other end, there were six participants whose scores on the LWID test were within one standard deviation of the normal mean, and three participants who achieved the age-appropriate level of performance on the non-word reading test (WordAttk).

As for visuo-spatial deficits in WS and their potential effects on reading, recent research reports of relative strength in visual object identification in WS (Hoffman & Landau, 2000) and in perceptual matching tasks (Milner & Goodale, 1995; Dilks et al. 2001), with major difficulties in pattern construction and reproduction of global elements, while reproduction of local elements is relatively preserved (Bellugi et al. 1994). Thus, it is not perception per se that is the core of the deficit in WS spatial difficulties and neither can it be the basis for their reading problems.

Furthermore, results in the current work are reported separately for the non-reading participants and for those who could identify all the letters in the alphabet and were able to read real words.

One of the main goals of this investigation was to explore whether PA and RON, which have been found to correlate with reading skills in normally developing children, would be related to reading achievement in individuals with WS. Our findings confirmed that PA skills are significantly correlated with reading achievement in individuals with WS. Elision and Word segmentation were significantly correlated with WordAttk. Elision correlated with LWID as well.

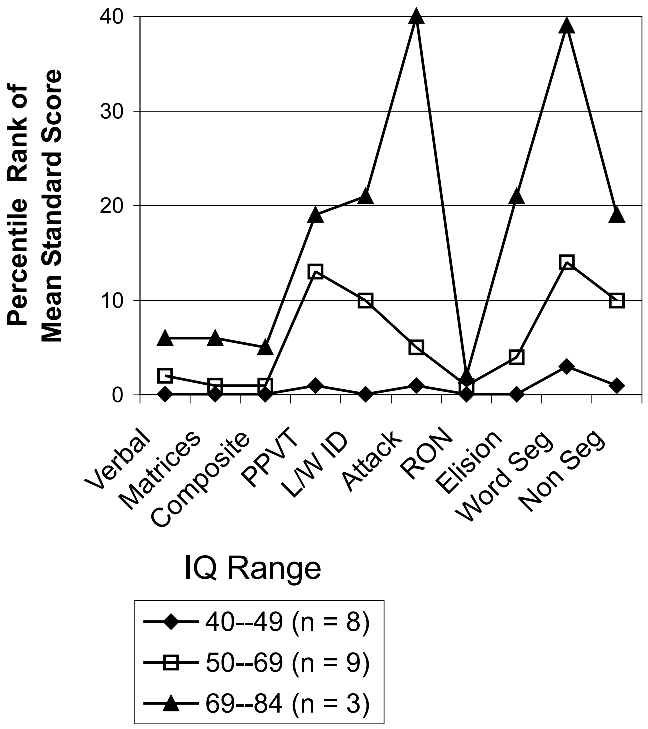

Figure 1 Profiles by IQ groups for participants with WS, readers and non-readers

There were no participants with WS who achieved good reading scores but had poor scores on the PA measures.

These findings suggest that individuals with WS show similar links between phonological skills and reading to normally developing children. It parallels results reported by Laing et al. (2001) for WS, as well as results reported for Down syndrome in the recent study by Cupples and Iacono (2000), yet not the findings reported by Rondal (1995) and Cossu and his colleagues (Cossu & Marshall, 1990; Cossu et al., 1993a, b).

In contrast to the findings on tasks measuring PA skills, we did not find a correlation between reading and RON in WS. Furthermore, only eight of the participants in our study (representing 40% of the sample) were able to perform slightly above floor level on the rapid naming task. Note that object naming is the least automatized of the naming skills – digits, letters and colors present a lesser problem to dyslexics, although on these tasks too they are not as good as good readers (Denckla & Rudel, 1976). It seems that a number of factors may be responsible for the participants' failure in the picture naming task. Attentional drift, slow verbal retrieval and subtle visual-processing delays may all be involved in poor performance in naming (Denckla & Rudel, 1976). However, none of these factors was critical enough to impede word reading in our group. The fact that Laing et al. (2001) did find correlation between naming speed and word reading may be due to the fact that their naming task included digits as well as pictures. Furthermore, in Laing et al. (2001) study correlations were calculated with real word reading rather than with non-word reading. The majority of individuals with WS in our study, i.e., all those whose IQs were lower than 70, received better scores on the former and thus the difference in performance between this measure and RON was smaller. Our subjects' poor performance on RON merits further investigation that will dissociate specific factors which could be causal in bringing about this unexpected result.

Since most of the participants in the study were close to ceiling on the implicit rhyme judgment task, no significant correlations were found between reading and performance on this measure. Our original attempt to get implicit rhyme judgments was guided by the belief that implicit phonological processing may be preserved in poor readers as well as in participants who failed to acquire reading. While all the readers, good as well as poor, were close to ceiling on the rhyme test, the second half of this hypothesis was not supported – the three non-readers in the sample were at chance on this task. It is not clear to us that failure to find the expected results should dismiss the hypothesis altogether. One may question whether the task was in fact implicit or whether the fact that judgment was based on rhymes was immediately clear to the participants, even if not stated aloud. Further research is needed to answer this question. These findings provide support, however, for the view that rhyming may be a prerequisite skill for reading acquisition, as has been suggested by Goswami (1988) and Bryant et al. (1990), among others.

Note that Laing et al. (2001) report moderate to strong correlations between reading and rhyme in their WS group. However, those correlations were not independent of age and general IQ. In other words, it may be that if Laing et al. (2001) had removed their three non-readers from the group the correlation between reading and rhyme would no longer be significant.

The finding that is of particular interest is the significant correlation in our WS participants between word reading and general IQ, more specifically with Mats, which is a measure of non-verbal IQ. This is evident in Tables 2–4 as well as in Figure 1. When Defs, which engages recognition of written words, is controlled for, Mats remains correlated with non-word reading. Notice that neither PPVT nor KBIT expressive vocabulary correlates with non-word reading. Receptive and expressive vocabulary is not correlated with real word reading either. Similar correlations between performance IQ and word reading are reported in Laing et al. (2001) for their WS group.

Further support for the effects of IQ on word decoding can be gleaned from Figure 1. The findings should be considered suggestive since group C, in which IQ is 70 or higher, has only three subjects. Still, the graphs are striking – individuals with an IQ of 70 and above show *much* better decoding and PA skills than individuals with IQ that is below 70. This advantage is much less clear on LWID. While group C shows non-word reading in the 40 percentile, their real word reading is in the 20 percentile. In the 50 to 70 IQ group, group B, the findings are reversed. There is better reading of real words than of non-words. Note that unlike non-word reading, real word reading may be achieved through the 'whole word' reading method, i.e., words may be recognized as wholes without there being phoneme-to-grapheme decoding.

It seems that a certain level of general intelligence is required to master decoding. This is further strengthened by the fact that the three non-readers were in group A, i.e., their IQ was below 50. Whether PA skills enable this achievement or rather PA skills are the result of reading, we cannot tell. But those abilities are certainly correlated in this population. However, vocabulary, as measured on the PPVT, is not. The achievements of groups B and C on the PPVT are fairly similar and probably reflect WS good lexical performance. Yet, despite similar vocabularies, group B remains poor in their reading, while group C shows impressive skills.

Let us further consider the issue of correlation between reading and IQ with respect to children with reading disabilities in the general population. Despite disparities in general IQ, research has failed to uncover significant differences in performance on language-related tasks of dyslexics, whose IQ is normal and above, and that of 'garden-variety' poor readers, whose IQ is lower than average, yet within the normal range (Ellis & Large, 1987; Stanovich, 1991). Both populations have deficits in PA tasks as well as in auditory verbal short-term memory for words, non-word repetition, rapid naming of numbers, letters and pictured objects and syntactic awareness. On the other hand, differences that were found between the groups on non-verbal tasks were all predicted on the basis of the differences in IQ and did *not* correlate with reading achievement (Shaywitz, Fletcher, Holahan, & Shaywitz, 1992). Similar findings were reported for Spanish-speaking children (Lopez & Gonzalez, 2000).

As for the heritability of reading difficulties, recent studies suggest that heritable deficits in word recognition are greater for individuals with specific reading deficits (relative to intelligence) compared to subjects with more general learning problems, where the discrepancy between IQ and achievement scores is not great (Rack & Olson, 1993; Meyer, 2000). The conclusion from most of the research on these definitional issues is that, contrary to the original hypotheses, reading in the general population is not predicted on the basis of IQ. Reading disability is distributed along a continuum that blends imperceptibly with normal reading ability and cannot be predicted on the basis of IQ scores.[2]

The well-researched cognitive profile of individuals with WS (Grant et al., 1997; Levy, in press b; Mervis et al., 1999; Vicari et al., 1996; Volterra et al., 1996), coupled with the current findings, suggests that performance on reading-related language skills does not predict reading level in WS. Good vocabulary, excellent auditory short-term memory, good phonology, or good non-word repetition skill did not predict good decoding skills. Poor performance on rapid object naming did not prevent acquisition of decoding skills either. Word reading did correlate, however, with general level of achievement, specifically with KBIT Mats. As noted earlier, unlike memory and oral language tasks, correlation between performance on PA tasks and non-word reading characterizes WS individuals in the same way that it does readers of normal intelligence.

Based on this profile of achievements, we offer the following hypothesis for future research. When IQ is within the retarded range, it becomes a major factor in predicting reading acquisition. The reason for this relates to the nature of the task. PA tasks as well as word decoding involve explicit, meta-cognitive procedures and as such they relate directly to IQ. Good oral language and related implicit phonological and memory skills, such as are seen in individuals with WS, are not likely to compensate for the level of awareness that is needed in learning to read. The causal direction between IQ and explicit meta-cognitive abilities is not clear. The prediction, however, is that there will be other tasks, not necessarily in the verbal modality, in which there will be good performance when knowledge is probed implicitly and poor performance when explicit meta-procedures are called for. Similar to reading, performance on such explicit tasks will be correlated with general IQ.[3]

Contrary to the retarded population, in dyslexic children in whom IQ is average and above, ability to handle meta-cognitive tasks may not be at the heart of their lower-than-average reading achievements. In these children there might be a deficit that is indeed unique to reading acquisition and that affects both reading and PA. In some children this deficit may be of a specifically linguistic nature as is probably the case in dyslexic children who evidence language problems in their preschool years. In such children explicit meta-cognitive tasks that do not relate to reading should not be impaired.

Finally, we note that the findings reported here have important implications for remediation programs and educational planning for teaching reading

[2] A recent twin study, however, separated between twin pairs with full-scale IQ below 100 or 100 and above and found that estimates of group deficits in reading performance were .43 and .72 respectively. The results provided evidence that the genetic etiology of reading disability differs as a linear function of IQ (Wadsworth, Olson, Pennington, & DeFries, 2000).

[3] This hypothesis may be tested with respect to tasks that involve implicit and explicit face recognition as well as Theory of Mind tasks. This is currently work in progress in our lab.

skills to children with WS. The emphasis on the phonics approach to teaching reading, which has regained wide popularity, should also be useful for individuals with WS in whom we have found strong links between reading level and performance on measures of phonological awareness.

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Appendix 1 Raw scores on the KBIT subtests for participants with WS

	All participants (n = 20)	Participants able to read (n = 17)
KBIT Expr. [max = 45]	32.7 (5.70)	32.59 (6.11)
KBIT Defs. [max = 37]	8.20 (8.63)	9.35 (8.89)
KBIT Mats. [max = 48]	19.55 (5.34)	20.35 (5.26)