The Relationship Between Age and IQ in Adults With Williams Syndrome

Yvonne M. Searcy, Alan J. Lincoln, Fredric E. Rose, Edward S. Klima, and Nasim Bavar
The Salk Institute for Biological Studies, Laboratory for Cognitive Neuroscience (La Jolla, CA)

Julie R. Korenberg
Cedars-Sinai Medical Center, Laboratory for Human Molecular Genetics (Los Angeles)

Abstract

The relationship between age and IQ was evaluated in a cross-sectional sample of 80 individuals with Williams syndrome (17 to 52 years). The relationship between age and WAIS-R subtest scores was such that increases and decreases in raw scores occurred at a rate sufficient to maintain stability of age-corrected scaled scores, indicating a developmental trajectory similar to that of the WAIS-R normative sample. Despite stability of age-corrected scaled scores with age, increased age was related to higher Performance IQ. This disparity, which occurs during the conversion of sums of scaled scores to IQs, may be unique to the WAIS-R. Although Performance IQ increased with age, results imply that the overall IQ of an adult with Williams syndrome will likely remain stable.

Identifying and understanding the developmental trajectory of intellectual ability in persons with neurodevelopmental syndromes is important to the characterization of the neurocognitive phenotype of the syndrome. Researchers often employ standardized intelligence tests for this purpose. Intelligence tests are also administered to adults with developmental disabilities in order to determine competence, monitor functioning, assess employability, and assess eligibility for social services. Thus, information regarding potential developmental changes in intellectual ability in individuals with a specific neurodevelopmental syndrome can provide caregivers with valuable information useful in planning for continuing education and long-term support.

Because IQs are derived with respect to the performance of same-age peers, IQs of typically developing individuals remain relatively stable with age (Schaie, 1983). Age-related changes in IQs that vary from the normal pattern of development have been reported for several neurodevelopmental syndromes. In adults with Down syndrome, for example, longitudinal studies show declines in IQs with age in about one third of those under 45, in over 70% of those 45 to 49, and diminishing IQ into old age (Carr, 1994; Fenner, Hewitt, & Torpy, 1987). Males with fragile X syndrome show intellectual growth and stability of IQs until about 10 to 15 years of age, at which point IQs begin to decline (Dykens et al., 1989; Hagerman et al., 1989). Conversely, persons with autism and no mental retardation show evidence of verbal intellectual skills that improve through adolescence and into adulthood (Kuck, Lincoln, & Heaton, in press). The developmental trajectory of IQ in Williams syndrome has not been fully explored.

Williams syndrome is a genetic disorder associated with the deletion of one copy of the gene for elastin and several surrounding genes on chromosome 7q11.23. The phenotype of Williams syndrome includes heart defects, such as supravalvular aortic stenosis, as well as dysmorphic

body and facial features, brain abnormalities in both gross anatomy and cytoarchitecture, and visuospatial and visuomotor integrative impairments (Bellugi & St. George, 2000; Pober & Dykens, 1996). Most individuals with Williams syndrome exhibit some degree of intellectual impairment, with the majority of adults scoring in the mild range of mental retardation (55 to 69 points) on standardized intelligence tests (Howlin, Davies, & Udwin, 1998).

Much of what is known about the relationship between IQ and aging in adults with Williams syndrome comes from studies by Udwin, Davies, and Howlin (1996), whose findings suggest no decline in IQs from adolescence into early adulthood. For example, they conducted a longitudinal study of 23 individuals with Williams syndrome over an 8.5-year period. Participants ranged from 10 to 15.75 years at initial testing with the Wechsler Intelligence Scale for Children-Revised—WISC-R (Wechsler, 1974), and from 19 to 24.83 at re-testing with the Wechsler Adult Intelligence Scale-Revised—WAIS-R (Wechsler, 1981). Unlike the IQs of individuals with fragile X or Down syndrome, IQs of individuals with Williams syndrome followed the same trend as the general population in showing a slight increase from WISC-R testing to WAIS-R re-testing, indicating no apparent decline in intellectual functioning over time. Howlin et al. (1998) compared the mean WAIS-R scores of 62 individuals with Williams syndrome (19 to 39 years of age) to the mean WISC-R scores reported for 44 children in an earlier study that included some of the same individuals (Udwin & Howlin, 1987, as cited in Howlin et al., 1998). They reported that 55% of the children for whom the WISC-R was administered had an IQ below 50, with 22% of the sample scoring below the basal level of 40. In contrast, all of the adults who completed the WAIS-R scored above the basal level, and only 4.8% had an IQ below 50, suggesting "some changes in score levels over time" (Howlin et al., 1998, p. 187).

Although the two studies above provide limited evidence suggesting that IQ in people with Williams syndrome does not decline with age from adolescence to age 39, such conclusions must be tempered due to the methodological confounds imposed by the psychometric relationship between the WISC-R and WAIS-R. It is well-established that the WAIS-R yields higher IQs than does the WISC-R and that this disparity becomes more evident toward the lower end of the intelligence curve (Avery, Slate, & Chovan, 1989; Rubin, Goldman, & Rosenfeld, 1985; Sattler, 1988; Spitz, 1988). Thus, there is an increased likelihood that spurious improvement in IQs would be observed in intellectually impaired individuals tested with the WISC-R as a child and then the WAIS-R as an adult. Due to the limits on direct comparability between the WISC-R and WAIS-R, researchers who employ the WISC-R and/or WAIS-R to examine the relationship between age and IQs are restricted by the respective age-range limitations of the tests.

Although longitudinal analyses, particularly those involving cross-sequential cohort designs, may be the optimal method of examining age-related changes in IQs (see Schaie, 1983), Kaufman (1990) showed that evaluating age-related changes in the sums of the scaled scores relative to changes in standard scores earned on the WAIS-R may be useful in ascertaining whether there are true developmental changes in intellectual functioning with age in a cross-sectional sample. Using this approach in the present study, we employed the WAIS-R to evaluate age-related changes in IQ in a cross-sectional sample of 80 individuals with Williams syndrome between the ages of 17 and 52 years. We evaluated the relationship between chronological age (CA) and raw scores as well as between CA and age-corrected scaled scores. With this approach we assessed whether potential age-related changes in IQs (Verbal, Performance, and Full-Scale) were due to "losing ground" compared to the typically developing normative sample or related to true developmental change in intellectual ability (increases or decreases in raw scores) with age.

Method

Participants

Eighty individuals with Williams syndrome (35 males, 45 females; *M* age = 29.8 years, range = 17 to 52) participated. They all had a clinical diagnosis of Williams syndrome and obtained a score of at least 3 points on the Williams syndrome Diagnostic Score Sheet, indicating the presence of a minimum threshold for common medical and physical characteristics associated with Williams syndrome in clinical studies (American Academy of Pediatrics, 2001). In addition, 79 participants tested positive on a FISH test (fluorescence in situ hybridization) for the absence of one copy of the gene for elastin on chromosome 7 (Korenberg et al., 2000). The remaining partic-

ipant was not tested for the elastin deletion but was diagnosed with Williams syndrome based upon having sufficient characteristics of this syndrome according to the Diagnostic Score Sheet. All WAIS-R data were obtained as part of an ongoing research project conducted at the Salk Institute's Laboratory for Cognitive Neuroscience. Genetic evaluation of the elastin deletion by FISH was performed at the Cedars-Sinai Medical Center Laboratory for Human Molecular Genetics.

Materials and Procedures

The WAIS-R was administered to all participants. This instrument is composed of 11 subtests, including 6 Verbal Scale and 5 Performance Scale subtests. Raw scores are obtained for each subtest, from which scaled scores ($M = 10$, $SD = 3$) are derived. Verbal (VIQ), Performance (PIQ), and Full-Scale (FSIQ) deviation IQs ($M = 100$, $SD = 15$) are derived from the appropriate sums of scaled scores. The WAIS-R scaled scores are based on a reference group (age 20 to 34 years). However, the WAIS-R manual also provides age-corrected scaled score equivalents for each of the nine age groups in the standardization sample. These age-corrected scaled scores are useful in conducting profile analysis and comparing an individual's performance directly with that of same-age peers as well as for making subtest interpretations and comparisons (Sattler & Ryan, 1988). The IQs and age-corrected scaled scores were derived according to the test manual instructions.

Results

Approximately 46% of the 80 individuals tested achieved a FSIQ in the borderline range (between 85 and 70 points), 45% scored in the mild range of mental retardation (69 to 55 points), and 9% fell in the moderate range (54 to 40 points) (American Psychiatric Association, 1994). Mean IQs, subtest age-scaled scores, and raw scores are reported in Table 1. There were no differences, $p > .05$, between males and females on VIQ, PIQ, or FSIQs nor were there gender differences on any of the Verbal subtests or the Performance subtests.

The participants' WAIS-R VIQs were significantly higher than PIQs by an average of 5.5 points, $t(79) = 7.72$, $p < .001$. The significance of the VIQ–PIQ difference for each individual in our sample was tested using the age appropriate critical value of the VIQ–PIQ difference at the .05

Table 1. Mean Raw Scores and Age-Corrected Scaled Scores and Their Correlation With Age

	Sum of scaled scores			IQ		
WAIS-R	Mean	*SD*	*r*	Mean	*SD*	*r*
FSIQ	47.8	15.5	−.09	67.4	8.3	.07
VIQ	28.1	9.2	−.01	71.5	8.2	−.01
PIQ	19.8	7.3	−.18	66.0	8.0	.26*
	Raw scores			Age-corrected scaled scores		
Verbal subtests						
Information	7.24	4.2	.06	4.55	2.2	−.06
Digit Span	8.01	2.8	−.10	4.94	1.9	.04
Vocabulary	19.20	11.4	.15	5.16	2.2	−.03
Arithmetic	3.59	1.8	−.03	3.73	1.6	−.06
Comprehension	8.17	4.8	.08	4.64	2.1	.01
Similarities	11.45	4.8	−.18	6.40	1.8	−.13
Performance subtests						
Picture Completion	7.51	3.4	−.17	4.99	1.7	.08
Picture Arrangement	4.67	3.4	−.22	5.17	2.1	−.02
Block Design	6.29	5.9	−.07	3.96	1.7	.15
Object Assembly	11.51	7.3	−.03	3.55	2.3	−.02
Digit Symbol	23.50	9.2	−.33**	3.89	1.4	.01

*$p < .05$. **$p < .01$.

level of confidence as defined by Wechsler (1981). The VIQ–PIQ difference was significant for 19 participants (23.8%), and for one individual, the PIQ was significantly higher than the VIQ.

The relationship between age and WAIS-R scores was assessed using Pearson correlations (see Table 1). Increased age was related to lower raw scores on the Digit Symbol subtest, $r = -.33$, $p < .01$, with raw scores showing a slow and steady decline after age 24, as occurs in the normative sample. Although age was not related to VIQ or FSIQ, increased age was related to higher PIQs, $r = .26$, $p < .02$. Longitudinal data from 4 of the WAIS-R participants support the latter findings. The WAIS-R had previously been administered to these 4 individuals at a mean age of 32.9 years. Upon re-testing an average of 9.2 years later (*M* age = 42.1), VIQs remained stable (*M*s = 75.8 and 75.2, respectively), but small gains occurred in FSIQ (*M*s = 69.5 and 73.0, respectively), $t(3) = 12.12$, $p < .001$, due primarily to increases in PIQs (*M*s = 65.8 and 72.0, respectively), $t(3) = 5.64$, $p < .01$, with all 4 participants gaining PIQ points (4, 5, 7, and 9 points).

Figure 1 illustrates the developmental trajectory of the sums of scaled scores for our Williams syndrome sample compared to that of the WAIS-R standardization sample (Wechsler, 1981, p. 26). In the standardization sample, the sum of scaled scores of the Verbal subtests increased slightly and gradually from age 16 through 34 and began to decline after age 44, whereas the sum of scaled scores of the Performance subtests increased gradually through age 24 before beginning to decline after age 34. Note that although the developmental trajectory of the Verbal, Performance, and Full-Scale sum of scaled scores remains relatively stable with age in Williams syndrome, PIQ in this Williams syndrome sample is, nevertheless, positively correlated with age. This phenomenon will be addressed below.

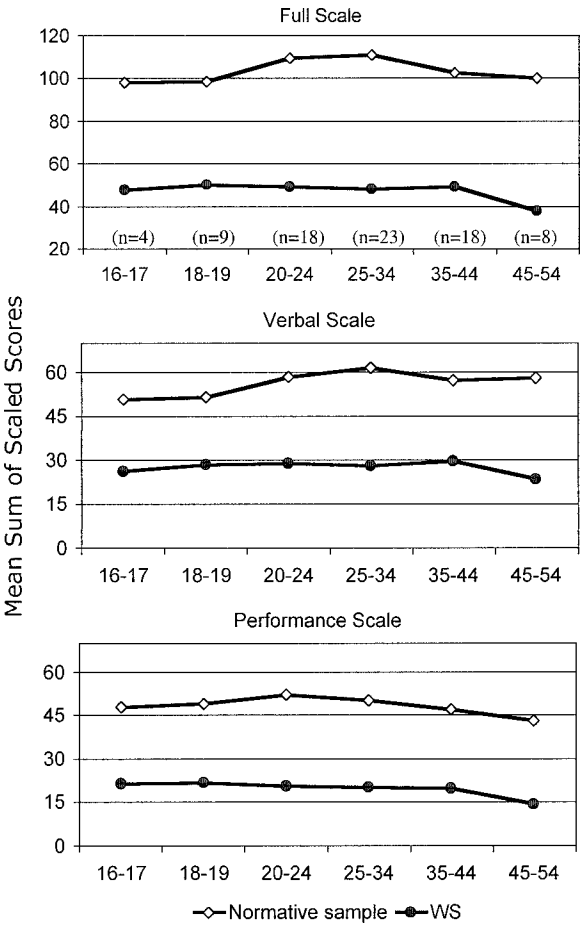

Figure 1. The developmental trajectory of the mean sums of scaled scores earned by each age group of our Williams syndrome sample compared to the mean sums of scaled scores earned by each relevant age group of the WAIS-R normative standardization sample (Wechsler, 1981, p. 26).

Discussion

Although decline in IQs with aging has been reported for other neurodevelopmental disorders, our results indicate no such decline in adults with Williams syndrome from age 17 to 52 years. Increases and decreases with age in WAIS-R subtest raw scores of individuals with Williams syndrome occurred at rates comparable to the WAIS-R normative sample, resulting in stability of age-corrected scaled scores in Williams syndrome with age. Thus, although IQs of individuals with Williams syndrome tend to be lower than average at any age, changes in the abilities measured by the various subtests of the WAIS-R occurred at a rate similar to typically developing same-age peers. The stability of age-corrected scaled scores with age further indicates that the pattern of strengths and weaknesses that are part of the phenotypic variability in Williams syndrome is maintained through adulthood. Similarly, Howlin et al. (1998) reported stability of the characteristic Williams syndrome profile of cognitive strengths and weaknesses from childhood (age 6 to 14) into early adulthood (age 18 to 39).

Although the stability of sums of scaled scores with age in the present study resulted in stability

of VIQ and FSIQ with age, PIQ increased significantly with age. The positive relationship between age and PIQ without concurrent improvement in raw scores on any of the Performance subtests, or increases in the scaled scores used to calculate IQs, indicates that the increase in PIQ in this sample is likely an artifact of how the PIQ is derived by the WAIS-R. For example, the mean PIQ of our Williams syndrome sample was 66. In the WAIS-R standardization sample, from age 16 to 54, an individual's Performance Scale sum of scaled scores can decline by up to 8 points while maintaining a PIQ of 66. Because the Performance Scale sum of scaled scores for our Williams syndrome sample remained stable with age, the mean PIQ of our sample actually increased with age. In contrast, in order to maintain a VIQ of 72 over the same age range, one would need to achieve an *increase* of up to 4 points in the sum of the Verbal scaled scores.

Evaluation of IQs of individuals in a special population for the purpose of assessing the stability of intellectual functioning with aging and comparison of their IQs to those of a normative sample is informative only in determining whether the *rate* of improvement or decline in the special population is comparable to that of the normative sample. Our findings highlight the need to consider changes in raw scores when evaluating meaningful change in intellectual functioning with age. By considering both raw and age-scaled scores, we differentiated age-related changes in IQs that actually reflected the ability of the individuals taking the test from changes in IQs resulting from the psychometric properties of the test.

Because our sample was cross-sectional and included individuals from families who volunteered to participate in an extended research protocol, care should be taken in generalizing the findings. Nonetheless, our results are consistent with those of Mervis, Morris, Bertrand, and Robinson (1999), who also found that Block Design and Digit Span ability scores (raw scores) on the Differential Ability Scales (Elliot, 1990) remained stable with age in a cross-sectional sample of adults with Williams syndrome (18 to 46 years). Further, our results are consistent with those from longitudinal studies indicating no evidence of decline in IQs in persons with Williams syndrome from childhood into early adulthood (e.g., Howlin et al., 1998), and with our concurrent finding for 4 individuals for whom we obtained longitudinal WAIS-R scores. Upon re-testing with the WAIS-R after an average delay of 9.2 years, these 4 individuals showed small gains in FSIQ, due primarily to gains of 4 to 9 points in PIQs, with no consistent increase in Performance subtest raw scores.

In sum, our results indicate that although IQs in adults with Williams syndrome were generally below average, subtest raw scores, and, thus, the sums of scaled scores used to calculate IQs, neither increased nor decreased differently than normal with age. This stability of sums of scaled scores resulted in VIQs and FSIQs that remained stable with age. Because PIQ increased with age despite a slight normal decline in Performance subtest raw scores, we suggest that this increase in PIQ with age *may* be a phenomenon unique to the WAIS-R. Therefore, one might expect that the IQs obtained by an individual with Williams syndrome as a young adult will generally remain relatively stable through their early 50s, as they do in the typically developing population. The finding of stability of intellectual functioning with age in Williams syndrome provides impetus for advocating continued enrichment and/or educational opportunities throughout the life-span of individuals with Williams syndrome.

References

American Psychiatric Association. (1994). *Diagnostic and statistical manual of mental disorders* (4th ed.). Washington, DC: American Psychiatric Association.

American Academy of Pediatrics. (2001). Health care supervision for children with Williams syndrome. *Pediatrics, 107,* 1192–1204.

Avery, R. O., Slate J. R., & Chovan, W. (1989). A longitudinal study of WISC-R and WAIS-R scores with students who are educable mentally handicapped. *Education and Training in Mental Retardation, 24,* 28–31.

Bellugi, U., & St. George, M. (Eds.). (2000). Linking cognitive neuroscience and molecular genetics: New perspectives from Williams syndrome [Special issue]. *Journal of Cognitive Neuroscience, 12*(1).

Carr, J. (1994). Long-term outcome for people with Down's syndrome. *Journal of Child Psychology and Psychiatry, 35,* 425–439.

Dykens, E. M., Hodapp, R. M., Ort, S., Finucane, B., Shapiro, L. R., & Leckman, J. F. (1989). The trajectory of cognitive development in

males with fragile X syndrome. *Journal of the American Academy of Child and Adolescent Psychiatry, 28,* 422–426.

Elliot, C. D. (1990). *Differential Ability Scales.* San Diego: Harcourt, Brace, Jovanovich.

Fenner, M. E., Hewitt, K. E., & Torpy, D. M. (1987). Down's syndrome: Intellectual and behavioural functioning during adulthood. *Journal of Mental Deficiency Research, 31,* 241–246.

Hagerman, R. J., Schreiner, R. A., Kemper, M. B., Wittenberger, M. D., Zahn, B., & Habicht, K. (1989). Longitudinal IQ changes in fragile X males. *American Journal of Medical Genetics, 33,* 513–518.

Howlin, P., Davies, M., & Udwin, O. (1998). Cognitive functioning in adults with Williams syndrome. *Journal of Child Psychology and Psychiatry and Allied Disciplines, 39,* 183–189.

Kaufman, A. (1990). *Assessing adolescent and adult intelligence.* Boston: Allyn & Bacon.

Korenberg, J. K., Chen, X., Hirota, H., Lai, Z., Bellugi, U., Burian, R., & Matsuoka, R. (2000). Genome structure and cognitive map of Williams syndrome. In U. Bellugi & M. St. George (Guest Eds.), Linking cognitive neuroscience and molecular genetics: New perspectives from Williams syndrome [Special Issue]. *Journal of Cognitive Neuroscience, 12*(1), 89–107.

Kuck, J., Lincoln, A. J., & Heaton, R. (in press). Age related changes in intellectual abilities among individuals with autistic disorder. *Journal of Autism and Developmental Disorders.*

Mervis, C. B., Morris, C. A., Bertrand, J., & Robinson, B. F. (1999). Williams syndrome: Findings from an integrated program of research. In H. Tager-Flusberg (Ed.), *Neurodevelopmental disorders* (pp. 65–110). Cambridge: MIT Press.

Pober, B. R., & Dykens, E. M. (1996). Williams syndrome: An overview of medical, cognitive, and behavioral features. *Child and Adolescent Psychiatric Clinics of North America, 5,* 929–943.

Rubin, H. H., Goldman J. J., & Rosenfeld, J. G. (1985). A comparison of WISC-R and WAIS-R IQs in a mentally retarded residential population. *Psychology in the Schools, 22,* 392–397.

Sattler, J. M. (1988). *Assessment of children* (3rd ed.). San Diego: Sattler Publisher.

Sattler, J. M., & Ryan, J. J. (1988). Wechsler Adult Intelligence Scale-Revised (WAIS-R). In J. M. Sattler, *Assessment of children* (3rd ed.). San Diego: Sattler Publisher.

Schaie, K. W. (1983). The Seattle Longitudinal Study: A twenty-one year exploration of psychometric intelligence in adulthood. In K. W. Schaie (Ed.), *Longitudinal studies of adult psychological development* (pp. 64–135). New York: Guilford Press.

Spitz, H. H. (1988). Inverse relationship between the WISC-R/WAIS-R score disparity and IQ level in the lower range of intelligence. *American Journal on Mental Retardation, 94,* 376–378.

Udwin, O., Davies, M., & Howlin, P. (1996). A longitudinal study of cognitive abilities and educational attainment in Williams syndrome. *Developmental Medicine and Child Neurology, 38,* 1020–1029.

Wechsler, D. (1974). *Wechsler Intelligence Scale for Children-Revised.* New York: Psychological Corp., Harcourt Brace Jovanovich.

Wechsler, D. (1981). *Wechsler Adult Intelligence Scale-Revised.* New York: Psychological Corp., Harcourt Brace Jovanovich.

Received 5/8/02, accepted 9/26/03.
Editor-in-charge: Elisabeth M. Dykens

The second author is also affiliated with Alliant International University, San Diego. *Williams Syndrome: Bridging Cognition and Gene* is an ongoing research project conducted at The Salk Institute for Biological Studies Laboratory for Cognitive Neuroscience in collaboration with Cedars-Sinai Medical Center Laboratory for Human Molecular Genetics. The project is supported by grants awarded to Ursula Bellugi from the National Institutes of Health (PO1 HD33113) as well as the Oak Tree Foundation and the James S. McDonnell Foundation (P50 NS22343). We thank Teresa Doyle for helpful comments made on earlier drafts of this paper. Special thanks go to the Williams Syndrome Association and to the individuals with Williams syndrome and their families who participated in this study. Some of these data were previously presented in Technical Report 2110, Institute for Neural Computation, University of California, San Diego. Requests for reprints should be sent to Yvonne M. Searcy, The Salk Institute for Biological Studies, Laboratory for Cognitive Neuroscience, 10010 N. Torrey Pines Rd., La Jolla, CA 92037. E-mail: searcy@lcn.salk.edu.

