The Relationship Between Age and IQ in Adults With Williams Syndrome

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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; Fenn, 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 visuo-spatial 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 23 individuals with Williams syndrome over an example, they conducted a longitudinal study of in IQs from adolescence into early adulthood. For Howlin (1996), whose findings suggest no decline between IQ and aging in adults with Williams syndrome, & Udwin, 1998).

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

<table>
<thead>
<tr>
<th>WAIS-R</th>
<th>Sum of scaled scores</th>
<th>IQ</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean</td>
<td>SD</td>
</tr>
<tr>
<td>FSIQ</td>
<td>47.8</td>
<td>15.5</td>
</tr>
<tr>
<td>VIQ</td>
<td>28.1</td>
<td>9.2</td>
</tr>
<tr>
<td>PIQ</td>
<td>19.8</td>
<td>7.3</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Raw scores</th>
<th>Age-corrected scaled scores</th>
</tr>
</thead>
<tbody>
<tr>
<td>Information</td>
<td>7.24</td>
</tr>
<tr>
<td>Digit Span</td>
<td>8.01</td>
</tr>
<tr>
<td>Vocabulary</td>
<td>19.20</td>
</tr>
<tr>
<td>Arithmetic</td>
<td>3.59</td>
</tr>
<tr>
<td>Comprehension</td>
<td>8.17</td>
</tr>
<tr>
<td>Similarities</td>
<td>11.45</td>
</tr>
</tbody>
</table>

Verbal subtests

Performance subtests

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 level.
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, \( r = -0.33, p < .01 \), 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, \( r = 0.26, p < .02 \). 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 (\( M = 42.1 \)), 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.

**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 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


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