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Children with Williams Syndrome: Developmental Trajectories for Intellectual Abilities, Vocabulary Abilities, and Adaptive Behavior

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Abstract

To examine longitudinal trajectories of intellectual abilities, single-word vocabulary abilities, and adaptive behavior for 76 children with Williams syndrome (WS) aged 4 – 15 years, we compared their standard scores (SSs) at two time points approximately 3 years apart on the same standardized measures. At the group level, mean SS declined significantly for 8 of the 12 measures and showed a slight (nonsignificant) increase or decrease for 4 measures. However, for most measures significant changes in SS were found for only a small proportion of the children, with some children evidencing significant declines and a smaller proportion evidencing significant increases. Significant SS changes were most common for adaptive behavior. For all measures, the mean magnitude of SS change was smaller for older children (> 7.5 years at Time 1) than for younger children (< 7.5 years at Time 1). Furthermore, correlations between Time 1 and Time 2 SSs were larger for the older cohort than for the younger cohort, indicating that SS stability was greater for older children than for younger children. Although mean SSs declined for most measures, indicating that children with WS as a group were not making the expected amount of progress relative to their general population peers who earned the same SS at Time 1, there was little evidence either of regression (loss of skills) or stagnation (failure to increase raw scores). The relations of these results to those of previous smaller-sample longitudinal studies of children with WS and the implications of the findings are considered.

Keywords

Williams syndrome; intellectual ability; language; visuospatial construction; nonverbal reasoning; vocabulary; adaptive behavior; longitudinal

INTRODUCTION

Williams syndrome (WS) is a neurodevelopmental disorder caused by a hemizygous microdeletion of 26 – 28 genes on chromosome 7q11.23 [Hillier et al., 2003]. The prevalence of this syndrome is estimated at 1 in 7500 live births [Strømme et al., 2002]. WS is associated with specific physical and medical features including a characteristic facies, cardiovascular disease (especially supravalvar aortic stenosis), connective tissue

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abnormalities, infantile hypercalcemia, and failure to thrive or growth deficiency [Morris, 2010]. The WS phenotype also includes specific behavioral characteristics, especially hypersociability (social disinhibition), empathy, worry, and anxiety (most commonly non-social specific phobia) [Klein-Tasman & Mervis, 2003; Leyfer et al., 2006].

WS also is associated with lowered intellectual abilities and more limited adaptive skills. Mean overall IQ is typically at the mild intellectual disability level, with a range from severe intellectual disability to average [Martens et al., 2008; Mervis and John, 2010]. This overall IQ masks a characteristic pattern of relative strengths and weaknesses which is best measured by the Differential Ability Scales-II [DAS-II; Elliott, 2007]: Mean standard scores (SS) are in the borderline range for Verbal abilities and Nonverbal Reasoning abilities but in the moderate intellectual disability range (~ 20 points lower) for Spatial abilities [Mervis and John, 2010]. This pattern of relative strengths and weaknesses also is found on measures of adaptive behavior, with performance on average significantly stronger for adaptive domains that are verbally-based than for domains that depend on spatial skills. For example, on the Vineland Adaptive Behavior Scales [VABS; Sparrow et al., 1984], children with WS on average earned higher SSs on the Socialization Skills and Communication Skills domains than on the Personal Living Skills and Motor Skills domains [Mervis et al., 2001] and on the Scales of Adaptive Behavior-Revised [SIB-R; Bruininks et al., 1996], children with WS on average earned higher SSs on the Social Interaction and Communication Skills cluster than on the Motor Skills, Personal Living Skills, and Community Living Skills clusters [Mervis and John, 2010].

Almost all of the studies delineating the intellectual and adaptive behavior components of the WS phenotype have been cross-sectional. For example, Martens et al. [2008] reported that 47 peer-reviewed journal articles (46 of which were cross-sectional) had been published that reported full-scale IQ results for individuals with WS, and Pitts and Mervis [in press] reported that 24 peer-reviewed journal articles (23 of which were cross-sectional) had provided IQ results for the Kaufman Brief Intelligence Test-2 [KBIT-2; Kaufman and Kaufman, 2004]. While cross-sectional studies offer an efficient approach to initial delineation of the phenotype associated with a particular syndrome, these studies provide a snapshot of different individuals' abilities at a single time point. In order to understand the developmental trajectory of phenotypic characteristics, longitudinal studies are crucial. As described in the next section, to date there have been very few longitudinal studies of the intellectual or adaptive behavior abilities of individuals with WS, and no longitudinal studies of single-word vocabulary abilities, that were based on analyses of SSs and that used the same assessment throughout the study. (For a discussion of the methodological problems with the use of age-equivalent scores rather than SSs, see Mervis and Klein-Tasman [2004] and Mervis and Robinson [2005].) Furthermore, the sample size for most of the extant longitudinal studies was small. The purpose of the present study was to consider the trajectories of intellectual, vocabulary, and adaptive behavior development of a relatively large sample of children with WS over a 3-year period.

In the remainder of the Introduction, we briefly describe the findings of the prior longitudinal studies. We then provide an introduction to the present study.

Longitudinal Studies

Five SS-based longitudinal studies of the intellectual abilities of individuals with WS have been published for which the same test was used at each time point. The results of these studies were mixed. Crisco [1990] used the Stanford-Binet Intelligence Scale Form L-M [Terman & Merrill, 1972] to compare the IQs of 14 children with WS at a mean chronological age (CA) of 4.2 years to their IQs five years later. Mean IQ was almost identical at the two time points. Gosch and Pankau [1996] compared the IQs of 18 children with WS at a mean CA of 6.5 years to their IQs two years later, using the German editions of the Columbia Mental Maturity Scale [CMMS; Bondy et al., 1969] and the Draw-a-Person Test [DAP; Zeiler, 1971]. On the DAP, mean IQ was 1 point higher at Time 2 than Time 1. However, on the CMMS, mean IQ decreased significantly, by an average of 9 points.

Mervis et al. [2012] used multilevel modeling to study possible changes in IQ Composite, Verbal SS, and Nonverbal SS (assessing nonverbal reasoning ability) as measured by the KBIT-2 [Kaufman and Kaufman, 2004] for 40 children with WS who were tested at least four times at intervals of at least 11.75 months. Mean CA was 7.44 years at the first assessment and 12.50 years at the last assessment. At the group level, no significant change was found for any of the IQ measures, although considerable variability was evidenced by individual children with some showing relatively little change and others showing large decreases or increases. No significant sex differences were found.

Fisch et al. [2010, 2012] used the Stanford-Binet IV [S-B IV; Thorndike et al., 1986] to compare the IQs of 17 children with WS at a mean CA of 8.85 years to their IQs two years later. Mean IQ declined by about 2 points over the full sample, with boys but not girls evidencing a significant decline. As shown in Fisch et al. [2012] Figure 2, all six of the participants who were < 7 years old at Time 1 had lower IQs at Time 2 than at Time 1; among the older participants results were mixed, with some showing increases in IQ and others decreases. For the eight children who were > 9 years at Time 1, changes in IQ at Time 2 were considerably smaller than for those who were younger at Time 1.

In contrast to the preceding studies which included only child participants, the participants in the remaining study spanned a very broad age range. Porter and Dodd [2011] used the Woodcock-Johnson Tests of Cognitive Ability-Revised [WJ-R; Woodcock and Johnson, 1989] to assess the overall IQ and Cognitive Factor SSs of 27 individuals with WS at two time points about five years apart. The participants' CA at Time 1 ranged from 5.00 – 44.67 years with a mean of 16.16 years. Paired t-tests indicated that there were no significant differences between Time 1 and Time 2 performance for overall IQ or any of the Cognitive Factor SSs. However, analyses taking into account the participant's CA at Time 1 indicated a significant linear effect for Comprehension-Knowledge SS and Fluid Reasoning SS and a significant quadratic effect for Oral Language SS. As shown in Porter and Dodd [2011] Figure 1, the SSs for all 11 participants who were children at both time points declined from Time 1 to Time 2 on both Comprehension-Knowledge and Oral Language; SSs for 15 of the 16 adults increased. On Fluid Reasoning, SS declined for six of the 11 child participants including the four youngest; of the remaining five, SS was identical or increased by 1 point for three. In contrast, the SSs for 14 of the 16 adults increased, in many cases by 10 or more points. At the individual level, 54% of the child participants showed a reliable decrease in

Comprehension-Knowledge SS and in Oral Language SS over time (no overlap in the confidence intervals for Time 1 and Time 2 SSs) and none showed a reliable increase. Of the adult participants, 10% showed a reliable decrease and 33% showed a reliable increase. The authors noted that Time 1 SSs for the children who showed a reliable decline varied from the low average range for the general population to moderate intellectual disability, indicating that the decline was not due simply to regression to the mean. For the remaining factors, ≤ 36% of participants showed a reliable SS change; no information about the age of the participants showing a reliable change or the direction of the change was provided.

One SS-based longitudinal study of the adaptive behavior of children with WS has been reported. Fisch et al. [2010, 2012] measured adaptive behavior, using the VABS, at the same time points as IQ was assessed. Mean VABS Composite SS was almost identical at the two time points, although as indicated in Fisch et al. [2012] Figure 5, many of the children evidenced substantial changes. Among the children < 7 years old, more than half evidenced declines, with the smallest decline larger in magnitude (absolute value) than the largest increase. The pattern among older children was more variable with slightly more than half showing an increase in SS at Time 2. No significant sex difference was found. Among the VABS domains, Socialization SS was significantly higher than either Communication SS or Daily Living SS [Fisch et al., 2012].

The Present Study

The findings of prior longitudinal studies of the intellectual abilities of children with WS have been mixed. In general, Time 1 and Time 2 IQ scores have not differed significantly, although Gosch and Pankau [1996] found a significant decline in CMMS SS and Fisch et al. [2010, 2012] found a significant decline in S-B IV overall IQ for boys. However, the picture changes somewhat when the performance of the children who were younger at Time 1 is compared to the performance of the children who were older. In the two studies for which figures were provided showing the difference in SS between Time 1 and Time 2 as a function of initial CA [Fisch et al., 2012; Porter and Dodd, 2011], the younger participants tended to show larger changes than the older participants and were more likely to evidence declines. Fisch et al. found this pattern for S-B IV Full Scale IQ; analyses for standard area scores were not reported. Porter and Dodd reported this pattern for the WJ-R Knowledge-Comprehension, Oral Language, and Fluid Reasoning cognitive factors but not for overall IQ or the other four WJ-R cognitive factors. In all of these studies, the sample size was relatively small leading to reduced power to detect significant differences.

In the present study, we considered the intellectual abilities, vocabulary abilities, and adaptive behavior of 76 children with WS at two time points approximately three years apart. We began by addressing whether there were significant group-level differences in SS as a function of assessment point (Time 1 vs. Time 2), sex, and/or type of ability. We also determined the proportion of children for whom differences between Time 1 and Time 2 SSs were reliable (no overlap between the confidence intervals for Time 1 and Time 2 SSs) and the direction of these differences. Correlations between CA at Time 1 and change in SSs were evaluated. To determine if the SSs for children who were older at the start of the study were more stable over the 3-year time period than those for children who were younger at

the start of the study, we conducted a second set of analyses comparing the magnitude of SS change from Time 1 to Time 2 for the children who were < 7.5 years old at Time 1 (n = 39) to the magnitude for the children who were > 7.5 years old (n = 37) for each of the assessments administered. We also compared the correlations between Time 1 and Time 2 SSs for the two groups of children.

MATERIALS AND METHODS

Participants

The final sample of participants was composed of 76 children (42 girls, 34 boys) with WS ranging in age from 4.02 – 15.24 years (M = 8.25, Mdn = 7.36, SD = 3.47) at Time 1 and from 7.05 – 17.98 years (M = 11.41, Mdn = 10.65, SD = 3.48) at Time 2. Children were included in the study if they had completed two assessments that were between 2.50 and 3.99 years apart and that included the four tests considered in this project, had a genetically-confirmed classic-length deletion of the WS region, had English as a native language or had been in an English-speaking school for at least 3 years, and did not have any additional diagnoses associated with intellectual disability. English was the native language of 97% of the children (74); the native language of the remaining children was Chinese (1) or one of the languages of India (1). Many of the children had completed more than the two assessments included in the present study. For these participants, the data from the most recent pair of assessments that was more than 2.5 but less than 4.0 years apart were used for this project. Based on the exclusionary criteria, four additional children were not included in the sample. Two children (1 boy, 1 girl) were excluded because their deletions were shorter than the classic WS deletion. Two boys with classic WS deletions were excluded because they had a comorbid diagnosis of fetal alcohol syndrome (1) or a gold-standard (based on ADOS-2, ADI-R, and clinical judgment) co-morbid diagnosis of an autism spectrum disorder (1). Finally, two additional children were excluded from the final sample because examination of standardized residuals identified one influential case (boy) for the measure of intellectual ability and one influential case (girl) for the measure of adaptive behavior. These children were excluded so that the analyses for the four assessments would be based on the same participants.

Measures

For each of the standardized assessments used in this study, the mean SS for the general population for both overall performance and performance on the clusters contributing to overall performance is 100, with a standard deviation (SD) of 15. All assessments were administered according to the standardized procedures.

The Differential Ability Scales 2nd edition [DAS-II; Elliott, 2007] was used to measure children's intellectual abilities. The Early Years form was administered to children aged 4 – 8 years and the School Age form to children aged 9 – 17 years. Both forms include six core subtests divided into three clusters of two subtests each: Verbal, Nonverbal Reasoning, and Spatial. The General Conceptual Ability (GCA; similar to Full-Scale IQ) is derived from performance on the six core subtests, and the Special Nonverbal Composite (SNC; similar to

Performance IQ) is based on performance on the four core subtests included in the Nonverbal Reasoning and Spatial clusters.

Single-word receptive vocabulary was measured by the Peabody Picture Vocabulary Test 4th edition [PPVT-4; Dunn & Dunn, 2007], and single-word expressive vocabulary was measured by the Expressive Vocabulary Test 2nd edition [EVT-2; Williams, 2007]. The PPVT-4 and EVT-2 were normed on the same standardization sample so their SSs can be directly compared.

The parent interview form of the Scales of Independent Behavior-Revised [SIB-R; Bruininks et al., 1996] was used to measure adaptive behavior. The SIB-R includes four clusters: Motor Skills, Social Interaction and Communication Skills, Personal Living Skills, and Community Living Skills. The Broad Independence SS is based on performance on all four clusters.

RESULTS

Group Level Differences

Descriptive statistics for DAS-II, PPVT-4, EVT-2, and SIB-R SSs at Time 1 and Time 2 are reported in Table I. To evaluate if SSs changed significantly over time, separate repeated measures (RM) ANOVAs were conducted for DAS-II GCA, DAS-II SNC, DAS-II Clusters, PPVT-4 and EVT-2, SIB-R Broad Independence, and SIB-R Clusters, with SSs at Time 1 and Time 2 as the dependent variables. Sex was included as a between-subjects factor. The assumption of normality of the residuals was met for all measures (Kolmogorov-Smirnov Test, Ps ≥ .30). Homogeneity of variance (Levene's Test, Ps ≥ .09) was met for SSs for the DAS-II, PPVT-4, EVT-2, SIB-R Broad Independence, and all of the SIB-R clusters except Time 1 Community Living Skills SS (P = .013). To correct for the violation of sphericity on the DAS-II Cluster analysis, Greenhouse-Geisser estimates were reported [Field, 2013]. The assumption of sphericity was met for all other analyses (Mauchly's Test, Ps > .10).

Intellectual Abilities—Descriptive statistics for SSs for all DAS-II components are reported in Table I. RM ANOVAs indicated there were no significant main effects or Time × Sex interaction for either DAS-II GCA (Ps ≥ .28) or DAS-II SNC (Ps ≥ .61). Thus, neither DAS-II GCA nor DAS-II SNC changed significantly over time, and SSs were similar for girls and boys.

In contrast, a RM ANOVA evaluating changes in DAS-II Cluster SSs revealed a significant Time × DAS-II Cluster interaction (F(1.87, 138.23) = 12.77, P < .001, η_p^2 = .15). The main effects of Time (F(1, 74) = 4.81, P = .031, η_p^2 = .06) and DAS-II Cluster (F(1.78, 131.75) = 230.31, P < .001, η_p^2 = .76) also were significant. There was no significant effect of Sex (P = .73), and no other interactions were significant (Ps ≥ .16). Pairwise comparisons revealed that, overall, Nonverbal Reasoning SS was significantly higher than both Verbal SS (P = .003) and Spatial SS (P < .001), and Verbal SS was significantly higher than Spatial SS (P < .001). Post hoc paired t-tests addressing the significant Time × DAS-II Cluster interaction indicated that Verbal SS was significantly higher at Time 1 than at Time 2 (t(75) = 2.56, P = .012, r = .28). Similarly, Nonverbal Reasoning SS was significantly higher at

Time 1 than at Time 2 ($t(75) = 4.10$, $P < .001$, $r = .43$). While Spatial SS increased from Time 1 to Time 2, this increase did not survive correction ($t(75) = -2.11$, $P = .038$, $r = .24$; Bonferroni corrected $\alpha = .017$).

Vocabulary Abilities—The PPVT-4 and EVT-2 are co-normed, allowing for a direct comparison of performance. Descriptive statistics for SSs for these measures are provided in Table I. RM ANOVA revealed a significant main effect of Assessment ($F(1, 74) = 35.90$, $P < .001$, $\eta_p^2 = .33$), with overall mean PPVT-4 SS ($M = 87.86$) significantly higher than overall mean EVT-2 SS ($M = 83.83$). There also was a significant main effect of Time ($F(1, 74) = 11.80$, $P = .001$, $\eta_p^2 = .14$), with overall mean SS significantly higher at Time 1 ($M = 87.11$) than at Time 2 ($M = 84.58$). Neither the main effect of Sex ($P = .73$) nor any of the interactions (Ps ≥ .16) was significant.

Adaptive Skills—Descriptive statistics for SSs for all components of the SIB-R are provided in Table I. To evaluate change in overall adaptive skills, a RM ANOVA was conducted for SIB-R Broad Independence SS. There was a significant main effect of Time ($F(1, 74) = 27.24$, $P < .001$, $\eta_p^2 = .27$), with mean SS significantly higher at Time 1 than at Time 2. Neither the main effect of Sex ($P = .07$) nor the Time × Sex interaction ($P = .91$) was significant.

Next, change in SIB-R Cluster SSs was evaluated. As indicated in Table II, RM ANOVA revealed significant main effects of Sex, SIB-R Cluster, and Time as well as several significant interactions: Time × Sex, SIB-R Cluster × Time, and SIB-R Cluster × Time × Sex. The three-way interaction is illustrated in Figure 1.

Descriptive statistics for SIB-R Cluster SSs at Time 1 and Time 2 are reported in Table III, separately for boys and girls. Separate RM ANOVAs were conducted for girls and boys in order to better evaluate the effect of SIB-R Cluster as a function of Sex. A similar pattern of results was found for the girls and boys. For girls, RM ANOVA revealed a significant Time × Cluster interaction ($F(2.59, 106.21) = 6.63$, $P = .001$, $\eta_p^2 = .14$). The main effects of Time ($F(1, 41) = 14.74$, P < .001, $\eta_p^2 = .26$) and Cluster ($F(2.45\ 100.46) = 70.66$, $P < .001$, $\eta_p^2 = .63$) also were significant. For boys, RM ANOVA revealed a significant Time × Cluster interaction ($F(2.65, 87.45) = 16.68$, $P < .001$, $\eta_p^2 = .34$). The main effects of Time ($F(1, 33) = 6.53$, $P < .001$, $\eta_p^2 = .17$) and Cluster ($F(2,55, 84.18) = 44.50$, $P < .001$, $\eta_p^2 = .57$) also were significant. Pairwise comparisons indicated that for both boys and girls, mean Social Interaction & Communication Skills SS was significantly higher than mean SS for each of the other clusters (Ps < .001). For boys, SSs for the other clusters did not differ significantly (P ≥ .70). For girls, mean Motor Skills SS and Personal Living Skills SS did not differ significantly ($P = .14$). Mean Community Living Skills SS was significantly lower than both mean Motor Skills SS and mean Personal Living Skills SS (Ps ≤ .001).

In order to compare SSs across time for each cluster, post-hoc analyses were conducted separately for girls and boys. For girls, paired t-tests indicated that mean Motor Skills SS was significantly higher at Time 1 than at Time 2 ($t(41) = 4.03$, $P < .001$, $r = .53$). Mean Community Living Skills SS also was significantly higher at Time 1 than at Time 2 ($t(41) = 3.83$, $P < .001$, $r = .51$). No difference in mean SS at Time 1 and mean SS at Time 2 was

detected for Social Interaction & Communication Skills ($P = .28$) or Personal Living Skills ($P = .51$). Paired t-tests revealed a similar pattern for boys. Mean Motor Skills SS was significantly higher at Time 1 than at Time 2 ($t(33) = 2.65$, $P = .012$, $r = .42$). Mean Community Living Skills SS also was significantly higher at Time 1 than at Time 2 ($t(33) = 4.77$, $P < .001$, $r = .64$). No significant difference in mean SS at Time 1 and mean SS at Time 2 was found for Social Interaction & Communication Skills ($P = .25$). While mean Personal Living Skills SS increased from Time 1 to Time 2, this increase did not survive correction ($t(33) = -2.60$, $P = .014$, $r = .41$; Bonferroni corrected $\alpha = .013$).

Next, post-hoc analyses were conducted comparing boys' and girls' mean SSs at Time 1 and Time 2 for each SIB-R Cluster. Independent sample t-tests revealed that mean Personal Living Skills SS at Time 1 was significantly higher for girls than boys ($t(74) = -4.01$, $P < .001$, $r = .42$). While Personal Living Skills SS at Time 2 ($t(74) = -2.20$, $P = .031$, $r = .25$) and Motor Skills SS at both Time 1 ($t(74) = -2.11$, $P = .039$, $r = .24$) and Time 2 ($t(74) = -2.26$, $P = .027$, $r = .25$) were higher for girls than boys, these differences did not survive correction (Bonferroni corrected $\alpha = .013$). Boys' and girls' mean SSs at Time 1 and Time 2 did not differ significantly for Social Interaction & Communication Skills (Ps $\geq .066$) or Community Living Skills (Ps $\geq .092$).

Individual Change

For each assessment, the pattern of SS change from Time 1 to Time 2 also was evaluated for each child. First, whether a child's SS increased, was identical, or decreased from Time 1 to Time 2 was determined. Then, the standard error of measurement (SEM, which varied as a function of the assessment and the child's age) was used to calculate the 95% confidence interval for SS at Time 1 and at Time 2. If the 95% confidence intervals for SS at Time 1 and at Time 2 did not overlap, the SS change was considered significant.

Intellectual Abilities—Patterns of change at the individual level for the DAS-II are reported in Table IV. For GCA, the pattern of change was evenly split between increases and decreases, with only a small proportion of children exhibiting a significant change (≥ 11 points) from Time 1 to Time 2. For SNC, scores decreased for slightly more than half of the children. However, for the small proportion of children whose SNC changed significantly (≥ 11 points), more showed an increase than a decrease. Figure 2 shows the individual SS trajectories for the Verbal, Nonverbal Reasoning, and Spatial Clusters from Time 1 to Time 2 (~3 years later). For the majority of children, SSs decreased from Time 1 to Time 2 for Verbal SS and Nonverbal Reasoning SS and increased for Spatial SS, although most of these SS changes were not significant. For the small proportion of children who did exhibit a significant change in SS from Time 1 to Time 2 (≥ 19 points for Verbal SS, ≥ 15 points for Nonverbal Reasoning SS, ≥ 12 points for Spatial SS), Verbal SS and Nonverbal Reasoning SS typically decreased, whereas Spatial SS typically increased.

Vocabulary Abilities—Pattern of change at the individual level for PPVT-4 and EVT-2 SSs is reported in Table IV. For about two-thirds of the children, SSs decreased from Time 1 to Time 2, but most of the changes were not significant. For the small proportion of children

who did exhibit a significant change in SS from Time 1 to Time 2 (≥ 12 points for PPVT-4, ≥ 14 points for EVT-2), about half evidenced an increase and half evidenced a decrease.

Adaptive Skills—Patterns of SS change at the individual level for the SIB-R are reported as a function of sex in Table V. For Broad Independence, the SSs of three-fourths of the girls and almost two-thirds of the boys decreased across time. The change in SIB-R Broad Independence SS was significant (≥ 8 points) for about one-third of the girls, with almost all showing a significant decrease. Almost two-thirds of the boys evidenced a significant change, most commonly a significant decrease.

Figure 3 shows the individual trajectories of SSs for the SIB-R Motor Skills, Social Interaction & Communication Skills, Personal Living Skills, and Community Living Skills clusters from Time 1 to Time 2 (~3 years later), as a function of sex. For the majority of both boys and girls, SSs decreased from Time 1 to Time 2 for Motor Skills, Social Interaction & Communication Skills, and Community Living Skills, although a small proportion of children evidenced increases. For both girls and boys, significant decreases (≥ 12 points) were most likely to occur for Community Living Skills. Only a small proportion of children exhibited a significant change in Motor Skills SS (≥ 19 points) or Social Interaction & Communication Skills SS (≥ 19 points); almost all of these children evidenced a significant decrease. Personal Living Skills SSs increased from Time 1 to Time 2 for about half of the girls and half of the boys. Although this increase in Personal Living Skills SS was not significant for any of the girls, it was significant (≥ 12 points) for about 18% of the boys.

Initial Age and Change in SSs

To determine if there was a linear relation between CA at Time 1 and change in SS from Time 1 to Time 2, difference scores (Time 2 SS – Time 1 SS) were computed for each measure. Descriptive statistics are reported in Table I for the full sample for all assessments and in Table III for boys and girls separately for the SIB-R clusters. For each measure, correlations between CA at Time 1 and change in SS from Time 1 to Time 2 (difference score) were computed. Mean CA at Time 1 did not differ significantly between girls (M = 8.66, SD = 3.82, range: 4.05 – 15.24 years) and boys (M = 7.74, SD = 2.97; range: 4.02 – 14.22 years) (independent t-test, $P = .25$).

No significant relation was detected between CA at Time 1 and the change in SS from Time 1 to Time 2 for DAS-II GCA ($r = -.08$, $P = .47$), DAS-II SNC ($r = -.20$, $P = .087$), DAS-II Verbal SS ($r = .13$, $P = .27$), DAS-II Nonverbal Reasoning SS ($r = -.005$, $P = .96$), PPVT-4 SS ($r = .04$, $P = .72$), or EVT-2 SS ($r = .12$, $P = .32$). Correlations between CA at Time 1 and change in SS from Time 1 to Time 2 for DAS-II Spatial SS ($r = -.24$, $P = .039$) and for SIB-R Broad Independence SS ($r = .29$, $P = .012$) did not survive correction (Bonferroni $\alpha = .004$).

As significant sex differences had been found for SIB-R Cluster SSs, correlations between CA at Time 1 and change in SS from Time 1 to Time 2 were evaluated separately for girls and boys. For the girls, no significant relation was detected for Social Interaction & Communication Skills ($r = .-068$, $P = .67$) or Personal Living Skills ($r = .-054$, $P = .73$). Significant relations between CA at Time 1 and change in SS from Time 1 to Time 2 were

found for Motor Skills ($r = .44$, $P = .004$) and Community Living Skills ($r = .48$, $P = .001$). For the boys, no significant relation was detected for Motor Skills ($r = .27$, $P = .12$) or Social Interaction & Communication Skills SS ($r = .16$, $P = .36$). The correlations for Personal Living Skills ($r = -.39$, $P = .023$) and Community Living Skills ($r = .43$, $P = .011$) did not survive correction (Bonferroni $\alpha = .004$).

Magnitude of Individual Change in Standard Scores

In order to evaluate the magnitude of change in SSs, the absolute value of the change in SS from Time 1 to Time 2 was computed (|SS at Time 2 – SS at Time 1|) for each assessment for each child. Descriptive statistics are reported in Table I for the full sample. To determine if SSs were more stable over the 3-year period for the group of children who were older at Time 1 than for the younger group of children (as suggested by the individual difference data reported in Fisch et al. [2012] and Porter and Dodd [2011]), the full sample was divided into approximately equal-sized cohorts of younger children (< 7.5 years at Time 1, $N = 39$) and older children (> 7.5 years at Time 1, $N = 37$). |Difference scores| for the two cohorts are reported in Table VI. Independent t-tests (also reported in Table VI) were used to compare the |difference scores| for the two cohorts for each measure. Variances were assumed to be unequal across the cohorts.

As indicated in Table VI, mean magnitude of change was larger for the younger cohort than for the older cohort for each of the 12 measures (binomial test, $P < .0005$). This difference in mean magnitude of change was significant for DAS-II GCA, DAS-II SNC, DAS-II Nonverbal Reasoning SS, DAS-II Spatial SS, SIB-R Broad Independence SS, SIB-R Motor Skills SS, and SIB-R Community Living Skills SS. Although there was a trend in this direction for SIB-R Personal Living Skills SS, it did not survive correction (Bonferroni corrected $\alpha = .013$). No significant difference between cohorts in magnitude of change was detected for any of the language-based measures (DAS-II Verbal SS, PPVT-4 SS, EVT-2 SS, SIB-R Social Interaction & Communication Skills SS).

Relations between Individual Participants' Standard Scores at Time 1 and Time 2

To determine the relations between individual children's SSs at Time 1 and Time 2, we computed correlations between Time 1 and Time 2 SSs for each of the assessments administered, first for the full sample and then separately for each cohort. As indicated in Table VII, all correlations were significant (Ps ≤ .001). However, all 12 correlations were stronger for the older cohort than the younger cohort (binomial test, $P < .0005$). Time 1 SSs accounted for 27% – 62% of the variance in Time 2 SSs for the younger cohort and 69% – 85% of the variance in Time 2 SSs for the older cohort.

DISCUSSION

The present study considered changes over time in intellectual, vocabulary, and adaptive behavior SSs for the largest longitudinal sample that has been described in the literature on WS. In the Discussion, we address our major findings in relation to the prior literature focusing first on intellectual and vocabulary abilities and then on adaptive behavior. We then

consider the pattern of differences in findings for younger children and older children and the implications of these patterns.

Intellectual and Vocabulary Abilities

In the present study, overall intellectual ability was measured by DAS-II GCA, which is similar to full-scale IQ. Mean GCA was almost identical at Time 1 and at Time 2 three years later, for both girls and boys. At the individual level, 11% of children showed a significant change, with GCA decreasing significantly for 7% and increasing significantly for 4%. It is important to note that the almost identical mean GCAs at Times 1 and 2 masked very different patterns of change for mean Spatial cluster SS on the one hand and mean SSs for the Nonverbal Reasoning and Verbal clusters on the other, as described below.

Our finding that mean full-scale IQ was consistent over 3 years for children with WS fits with the findings reported by Crisco [1990] for mean S-B L-M full-scale IQ over a 5-year period and by Porter and Dodd [2011] for mean WJ-R full-scale IQ, also over a 5-year interval. In these two studies, possible sex differences were not addressed. Fisch et al. [2010, 2012] who did consider possible sex differences, found that S-B IV full-scale IQ was consistent for girls over a 2-year period but declined significantly for boys. None of these authors addressed the question of whether changes in full-scale IQ were significant for individual children.

Performance on the DAS-II Spatial cluster was used to measure spatial ability in the present study. Mean DAS-II Spatial SS increased slightly from Time 1 to Time 2 for both boys and girls but the increase was not significant after Bonferroni correction. DAS-II Spatial SS changed significantly over the 3-year period for 20% of the children, with 17% evidencing increases and 3% decreases. Almost all of the children whose Spatial SS increased significantly were in the younger cohort. Gosch and Pankau [1997], who used a spatial measure, the Draw-a-Person Test, as a measure of IQ, reported that mean SS increased slightly but not significantly over the 2-year period of study. Results for individual participants were not reported. Porter and Dodd [2011], whose participants included both children and adults, found that mean WJ-R Spatial Relations SS increased significantly over the 5-year period of study, with 30% of participants evidencing a significant increase. Separate results for children and adults were not reported.

The DAS-II Nonverbal Reasoning cluster provided a measure of nonverbal reasoning ability in the present study. Mean DAS-II Nonverbal Reasoning SS declined significantly over the 3-year period of study with 8% of individual children showing a significant decrease and no child showing a significant increase. Once again, this finding was consistent with that reported by Gosch and Pankau [1997], who found that SS on the CMMS (a nonverbal-reasoning assessment used by the authors to measure IQ) declined significantly over the 2-year period of study. The findings of Porter and Dodd [2011] for the child participants in their sample on the WJ-R Fluid Reasoning cognitive factor also were consistent with a decline in nonverbal reasoning SSs.

In the present study, Verbal IQ was measured by the DAS-II Verbal cluster. In addition, single-word vocabulary abilities were measured by the PPVT-4 and EVT-2. Mean SS on all

three measures decreased significantly over the 3-year period for both boys and girls, with 8% of the children demonstrating a significant decline for DAS-II Verbal SS and PPVT-4 SS and 3% for EVT-2 SS. Significant SS increases were shown by 7% of children on the PPVT-4 and 3% on the EVT-2; no child showed a significant increase on the DAS-II Verbal cluster. In the only previous longitudinal study that considered verbal abilities, Porter and Dodd [2011] found that SSs on the WJ-R Knowledge-Comprehension and Oral Language cognitive factors declined for all of the child participants over the 5-year period of study, with 54% evidencing a significant SS decline on one or both of these cognitive factors. Given the pattern of declines in verbal abilities over time for multiple measures and across studies, increased focus on building vocabulary skills, especially by continuing to read to the child at his/her listening comprehension level (as a separate activity from working with the child to learn to read independently), explaining to the child words that he/she does not seem to understand, and incorporating those words into the family's and classroom's daily vocabulary is crucial. Similarly, a focus on the verbal reasoning skills and executive functioning skills needed for both listening and reading comprehension is important [Mervis, 2009].

Adaptive Behavior

Overall adaptive behavior was measured by SIB-R Broad Independence SS, which declined significantly over the 3-year period of study with no significant difference between girls and boys. At the individual level, 42% of participants evidenced a significant decrease in SS and 7% evidenced a significant increase. Given the large proportion of children who showed a significant decline in SS over a 3-year period, increased focus on developing adaptive skills both at home and in the local community is crucial.

This set of findings differs from the results reported in the only other longitudinal study of the adaptive behavior abilities of children with WS. Fisch et al. [2010, 2012], who used the VABS to measure adaptive behavior over a 2-year period, reported that overall SS was almost identical at the beginning and end of the study. Fisch et al. also did not find significant differences between girls and boys. It is likely that the contrasting patterns in overall SS are due in part to differences in the manner in which adaptive skills are divided into clusters and the weight given to each type of skill in computing the overall SS. In particular, although the SIB-R includes Motor Skills (an area of considerable weakness for individuals with WS) in the computation of Broad Independence SS for all ages, the VABS only measures Motor Skills for children < 7 years old, so includes Motor Skills only in the VABS Composite for children < 7 years old (4 of the 18 participants in Fisch et al.). In addition, the SIB-R Social Interaction & Communication Skills cluster (an area of relative strength for individuals with WS) is divided into two domains on the VABS, and the SIB-R Personal Living Skills and Community Living Skills clusters (areas of considerable weakness) are collapsed into one domain on the VABS. Thus, the types of adaptive skills emphasized by the two assessments differ considerably, with the emphasis on the VABS corresponding to areas of relative strength for individuals with WS and the emphasis on the SIB-R corresponding to areas of relative weakness. Furthermore, as described below, the pattern of change over time as a function of SIB-R cluster is consistent with Fisch et al.'s

finding that overall adaptive skill SS as measured by the VABS did not change significantly over the 2-year period of study.

The findings for changes in SIB-R cluster SSs over the 3-year period of the present study were complex, evidencing differing patterns as a function of interactions among time point, adaptive behavior cluster, and sex. Consistent with Fisch et al.'s finding of no significant change in VABS Composite SS, mean Social Interaction & Communication Skills SS did not change significantly from Time 1 to Time 2 for either girls or boys. Motor Skills SS (not included in VABS Composite SS for most of the participants in Fisch et al.'s study) and Community Living Skills SS declined significantly for both boys and girls, with 29% of children evidencing a significant decline in Community Living Skills SS. For Personal Living Skills SS there was no significant change for girls. For boys, Personal Living Skills SS increased from Time 1 to Time 2 but not enough to survive Bonferroni correction. Importantly, however, 18% of the boys showed a significant increase.

Patterns of Change for Younger Children vs. Older Children

As indicated above, there is a consistent pattern of longitudinal decreases in nonverbal reasoning and language SSs for children with WS over the 2 to 5-year period studied by different researchers. There also was a pattern of longitudinal decreases in the present study for two types of adaptive ability SSs. These decreases, although often significant at the group level, usually were relatively small and well within the SS confidence interval for the assessment. Furthermore, for most of the measures only a small proportion of individual children showed a significant decrease in SS. However, if the mean SS decreases continued at the same average rate over the 14-year period from age 4 – 17 years, later SSs eventually would be outside of the confidence interval and a large proportion of individual children would show a significant decline in SS. For example, the decline found for DAS-II Nonverbal Reasoning mean SS (4.45 points over 3 years) would correspond to a 17.8 point decline over 12 years, well outside the 95% confidence interval. Individual-difference data presented graphically in Porter and Dodd [2011] and Fisch et al. [2012] suggested that large changes (typically declines) in SS were common among young children but less common for older children or adults on most measures. To test this possibility statistically, we compared the magnitude of change in SS for children who were < 7.5 years old at Time 1 to those for children who were > 7.5 years old at Time 1. The magnitude of SS change was smaller for the older cohort than for the younger cohort for every measure considered in this study and the difference was significant for all of the non-language measures except for Personal Living Skills. Furthermore, the correlations between Time 1 and Time 2 SSs were stronger for the older cohort that the younger cohort for each of the 12 measures considered, with the weakest correlation for the older cohort larger than the strongest correlation for the younger cohort. Time 1 SS accounted for 27% – 62% (median: 50%) of the variance in Time 2 SS for the younger cohort but 69% – 85% (median: 75%) of the variance for the older cohort. The pattern of magnitude and correlation findings indicates that SSs earned by children older than 7.5 years are on average considerably more stable over time than are SSs earned by younger children. In a longitudinal study of 220 typically developing children, Weinert and Hany [2003] also found stronger correlations over time for full-scale IQ for older children

than for younger children (e.g., correlation from age 4 years to age 7 years: .47; correlation from age 9 years to age 12 years: .80).

In the context of the significant decline in SS for most of the measures over the 3-year period, it also is important to remember that a SS decrease indicates a slower rate of development than expected relative to same-CA general-population peers who earned the same original SS. It does not mean that the child has lost skills. There was little evidence of loss of skills for any of the measures included in the present study over the 3-year period. There also was little evidence of stagnation (lack of increase in raw scores) on any of the measures over the 3-year period. Thus, the children in the study continued to progress in all of the areas assessed over the 3-year period of study.

Limitations and Future Directions

Although the sample size for the present study was almost three times as large as the largest previous longitudinal study of individuals with WS that included children, it still is relatively small for the 14-year age range that was included, especially given the presence of significant differences between girls and boys for some components of adaptive behavior. In addition, the time period covered by the study for each child (3 years) is relatively short. Finally, as was true for all of the prior longitudinal studies of full-scale IQ for individuals with WS, only two data points were included per participant. Studies spanning a longer time period and including multiple data points for each participant are needed to allow for the use of growth curve modeling to more accurately delineate the developmental course of intellectual, vocabulary, and adaptive abilities for individuals with WS relative both to their general-population peers and their peers with WS.

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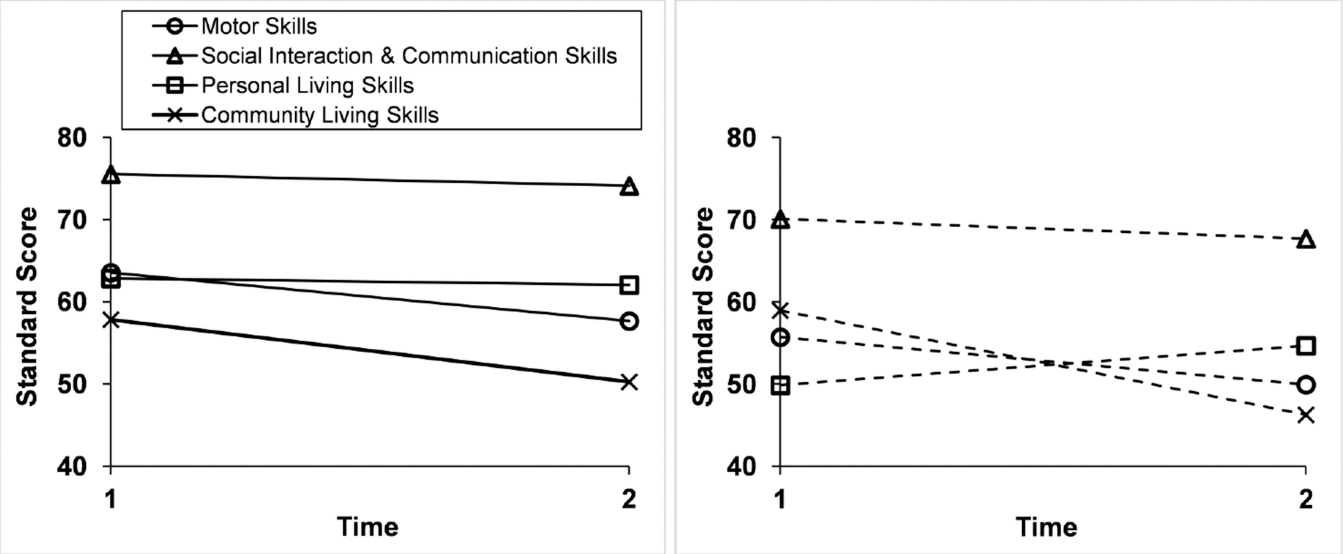
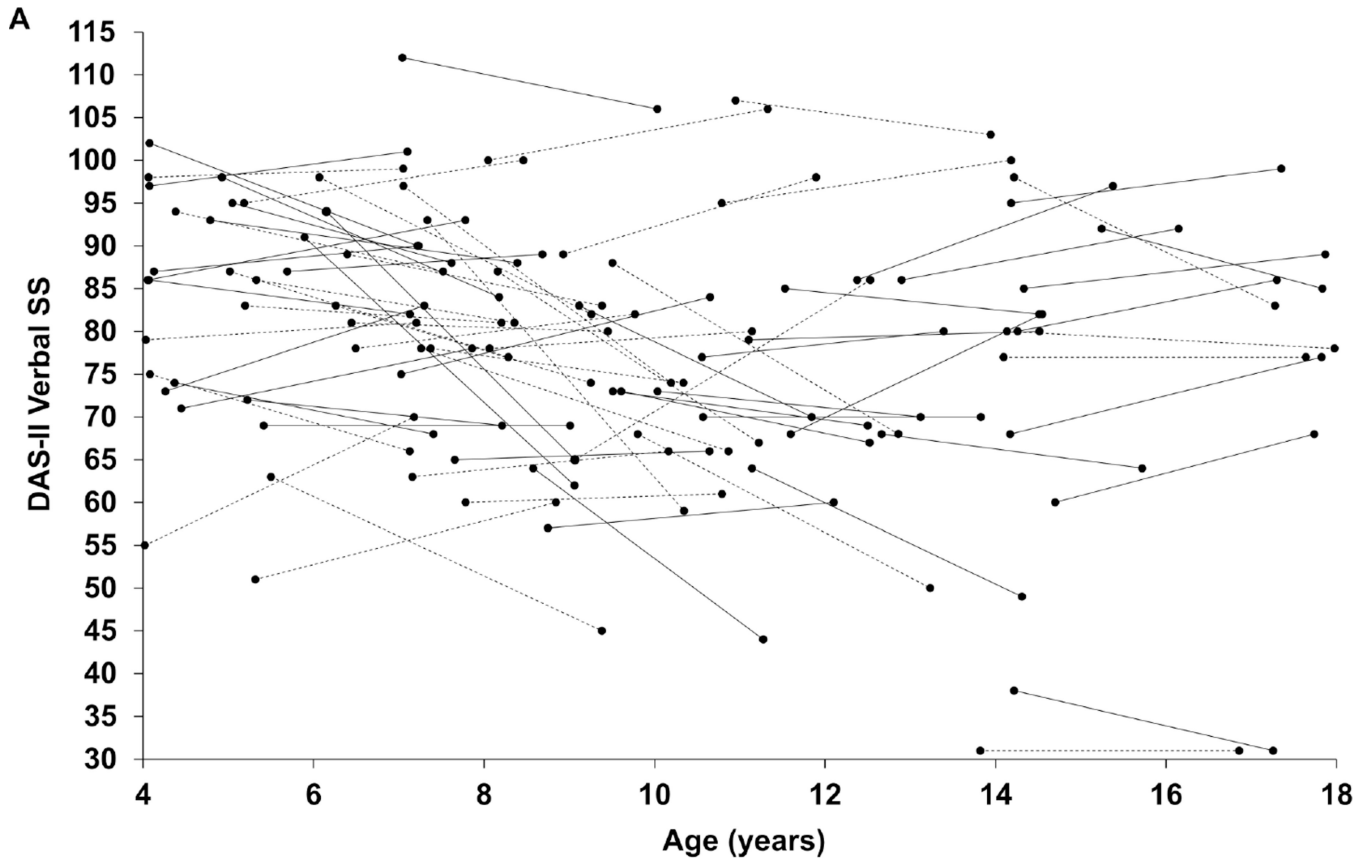
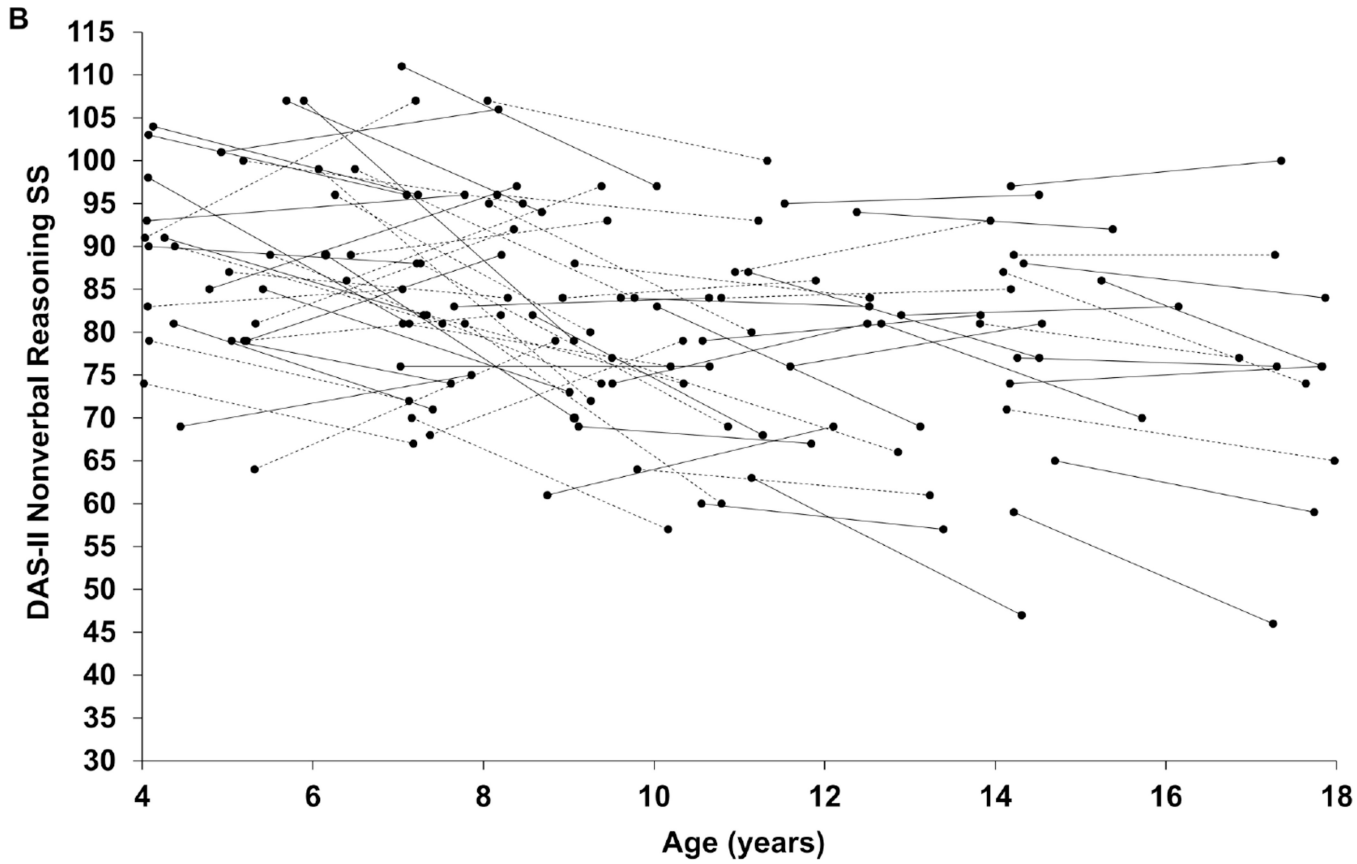


Figure 1.

Mean Scales of Independent Behavior-Revised (SIB-R) Cluster Standard Scores (SS) as a Function of Time for Girls (solid lines) and Boys (dashed lines)





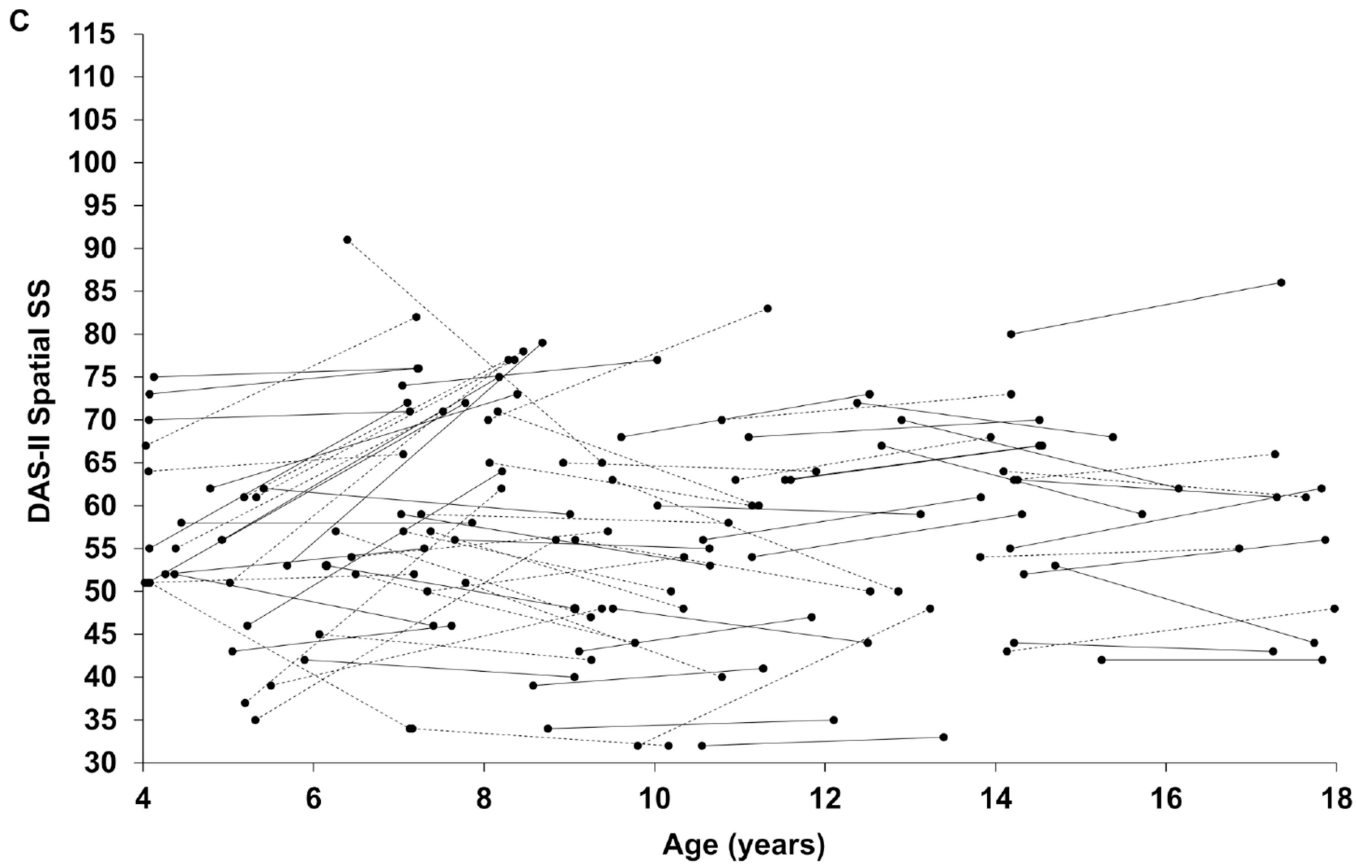
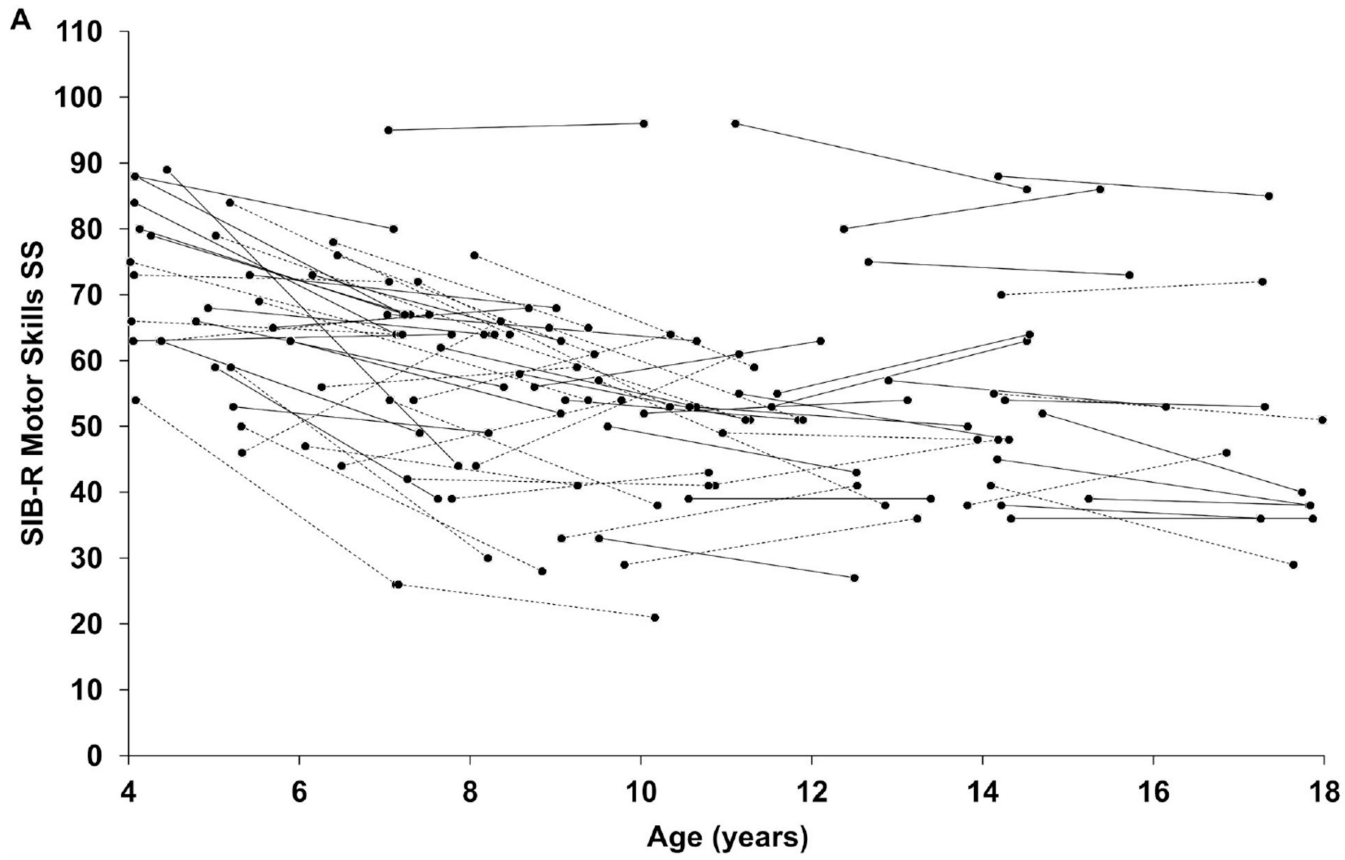
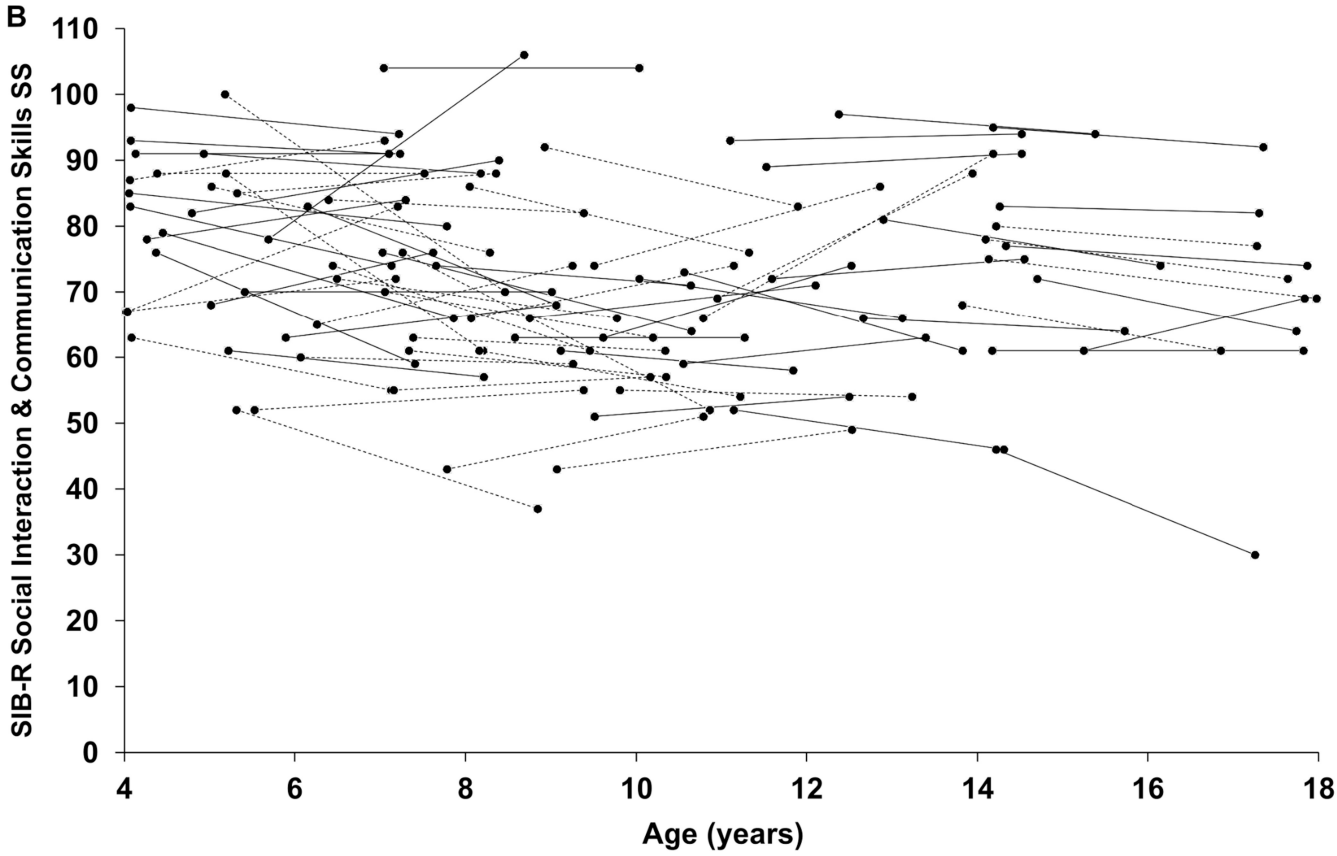
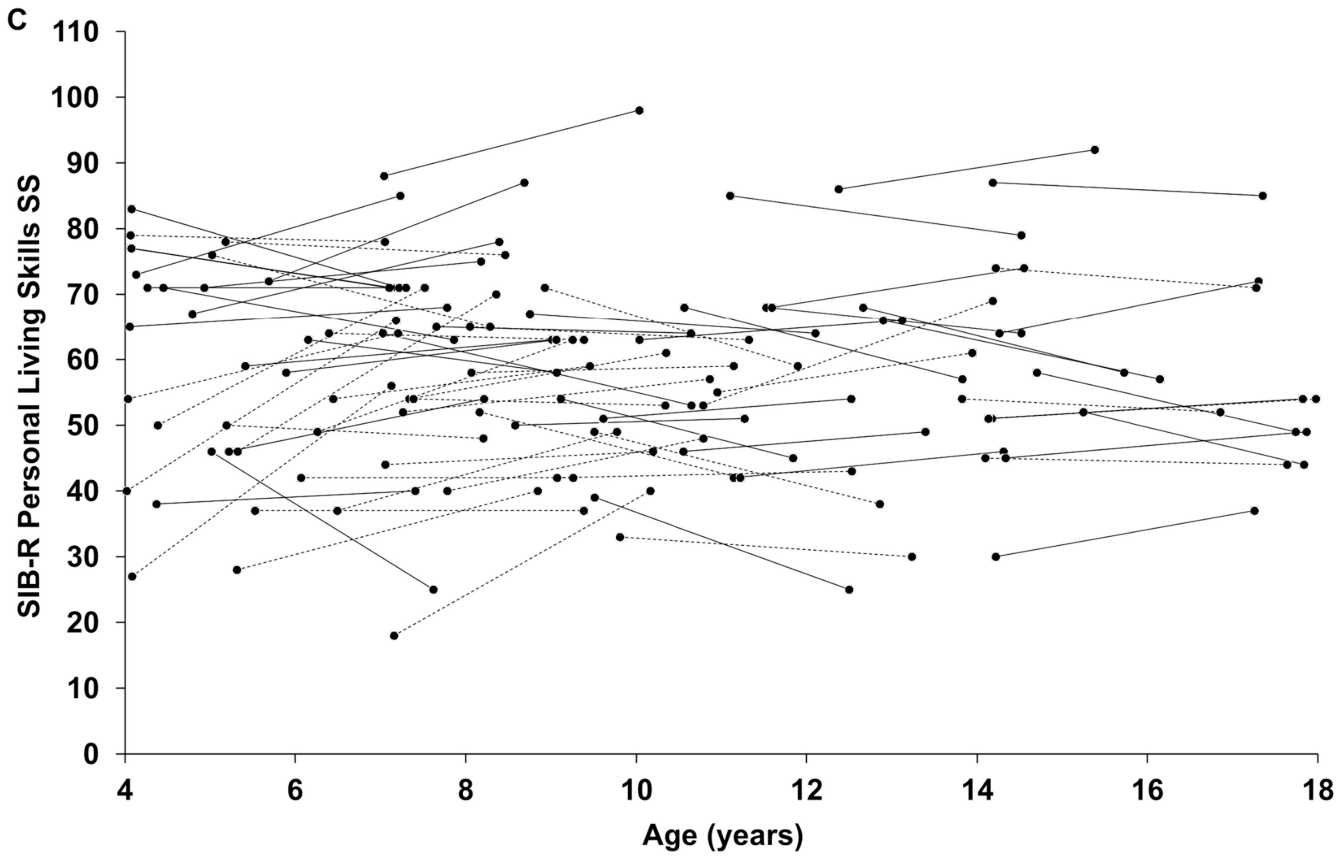


Figure 2.

Individual Differential Ability Scales-II (DAS-II) Cluster Standard Score (SS) Trajectories as a Function of Age for Girls (solid lines) and Boys (dashed lines). Panel A. DAS-II Verbal Cluster SSs. Panel B. DAS-II Nonverbal Reasoning Cluster SSs. C. DAS-II Spatial Cluster SSs.







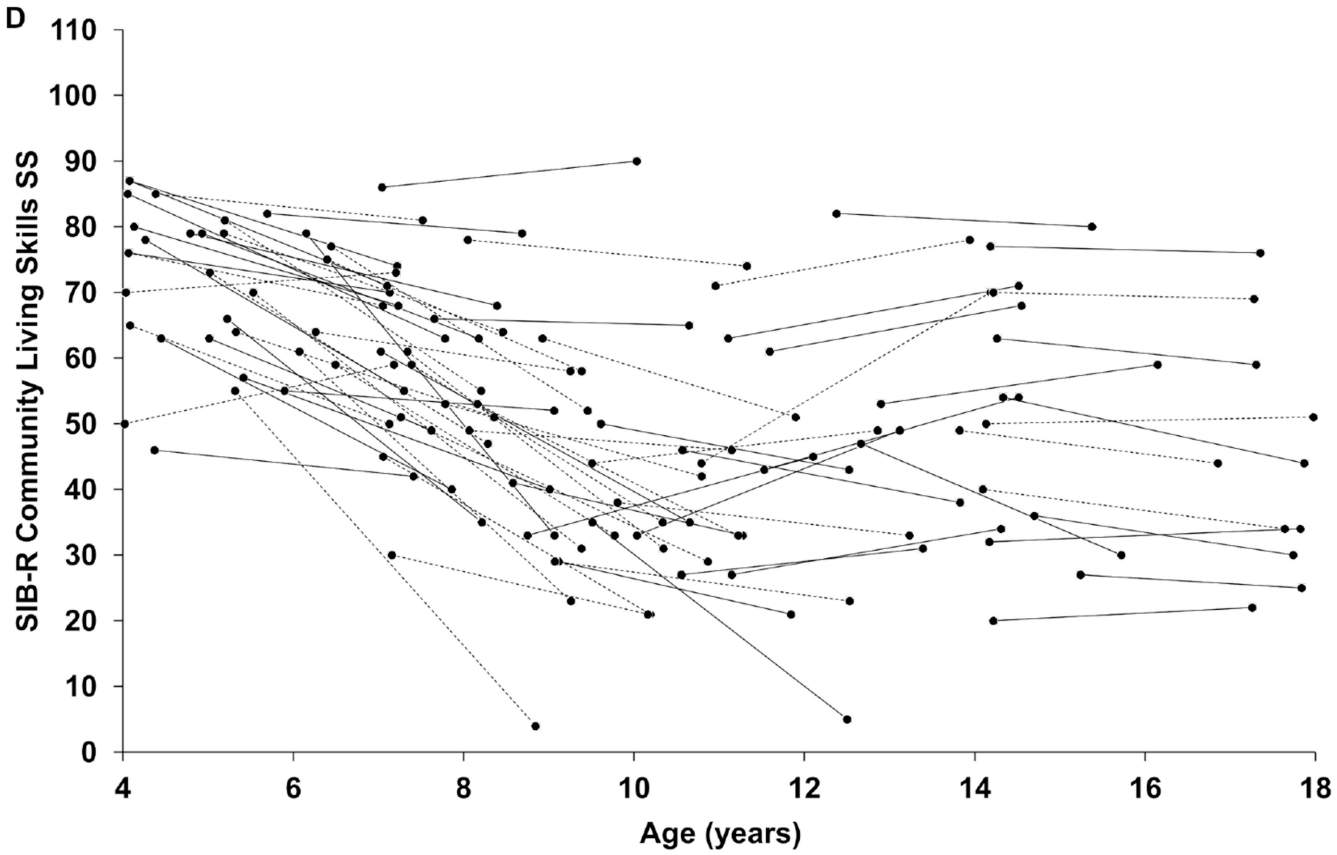


Figure 3.

Individual Scales of Independent Behavior-Revised (SIB-R) Cluster Standard Score (SS) Trajectories as a Function of Age for Girls (solid lines) and Boys (dashed lines). Panel A. Motor Skills SSs. Panel B. Social Interaction & Communication Skills SSs. Panel C. Personal Living Skills SSs. Panel D. Community Living Skills SSs.

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Table I

Descriptive Statistics for Measures of Intellectual Abilities, Vocabulary Abilities, and Adaptive Behavior

Measure	Time 1			Time 2			Difference			|Difference|		
	Mean	SD	Range	Mean	SD	Range	Mean	SD	Range	Mean	SD	Range
DAS-II GCA	68.88	11.01	44 – 98	68.03	12.22	38 – 96	−0.86	6.98	−19 – 14	5.67	4.11	0 – 19
DAS-II Verbal SS	79.97	14.99	31 – 112	76.86	15.83	31–106	−3.12	10.61	−34 – 21	8.25	7.31	0 – 34
DAS-II Nonverbal Reasoning SS	84.38	11.90	59 – 111	79.93	12.77	46 –107	−4.45	9.47	−28 – 16	8.37	6.22	0 – 28
DAS-II Spatial SS	56.26	11.78	32 – 91	58.68	13.20	32 – 86	2.42	10.02	−26 – 26	7.50	7.02	0 – 26
DAS-II SNC	66.30	11.17	39 – 90	65.82	13.09	39 – 93	−0.49	8.41	−16 – 21	6.83	4.87	0 – 21
PPVT-4 SS	89.18	14.02	47 – 120	86.59	14.62	41 – 115	−2.59	8.31	−19 – 17	6.72	5.49	0 – 19
EVT-2 SS	85.08	13.71	46 – 112	82.74	12.48	49 – 113	−2.34	7.53	−22 – 22	5.92	5.17	0 – 22
SIB-R Broad Independence	55.51	17.13	16 – 92	49.37	18.11	0 – 95	−6.14	10.12	−40 – 20	8.99	7.67	0 – 40
SIB-R Motor Skills	60.05	16.45	26 – 96	54.21	15.24	21 – 96	−5.84	10.97	−45 – 20	9.50	7.97	0 – 45
SIB-R Social Interaction & Communication Skills	73.12	13.96	43 – 104	71.26	15.19	30 – 106	−1.86	10.01	−30 – 28	7.49	6.85	0 – 30
SIB-R Personal Living Skills	57.07	15.39	18 – 88	58.76	14.93	25 – 98	1.70	9.67	−21 – 29	7.41	6.39	0 – 29
SIB-R Community Living Skills	58.36	17.89	20 – 87	48.47	19.04	4 – 90	−9.88	14.25	−51 – 26	13.30	11.08	1 – 51

Note. Difference = Time 2 SS – Time 1 SS. |Difference| = Absolute value of the difference between Time 2 and Time 1 SSs.

Abbreviations: DAS-II (Differential Ability Scales-II), General Conceptual Ability (GCA; similar to IQ), SS (standard score), SNC (Special Nonverbal Composite; similar to Performance IQ), PPVT-4 (Peabody Picture Vocabulary Test-4), EVT-2 (Expressive Vocabulary Test-2), SIB-R (Scales of Independent Behavior-Revised)

Table II

Repeated Measures ANOVA for Standard Scores (SS) as a Function of Scales of Independent Behavior-Revised (SIB-R) Cluster, Time, and Sex

Effect	*df*	*Error*	*F*	*P-value*	η_p^2
Sex	1.00	74.00	4.16	.045	.05
SIB-R Cluster	2.31	170.80	64.30	< .001	.46
Time	1.00	74.00	36.41	< .001	.33
SIB-R Cluster * Sex	2.31	170.80	0.20	.85	.00
Time * Sex	1.00	74.00	8.26	.005	.10
SIB-R Cluster * Time	2.28	169.09	89.58	< .001	.55
SIB-R Cluster * Time * Sex	2.28	169.09	5.27	.004	.07

Note. Greenhouse-Geisser estimates were reported

Table III

Descriptive Statistics for the Scales of Independent Behavior-Revised (SIB-R) Adaptive Skills Clusters as a Function of Sex

Measure	Time 1						Time 2						Difference					
	Girls			Boys			Girls			Boys			Girls			Boys		
	Mean	SD	Range	Mean	SD	Range	Mean	SD	Range	Mean	SD	Range	Mean	SD	Range	Mean	SD	Range
Motor Skills	63.55	16.59	33 – 96	55.74	15.44	26 – 84	57.67	15.68	27 – 96	49.94	13.73	21 – 72	−5.88	9.46	−45 – 10	−5.79	12.73	−29 – 20
Social Interaction & Communication Skills	75.55	13.83	46 – 104	70.12	13.73	43 – 100	74.14	15.90	30 – 106	67.71	13.66	37 – 91	−1.40	8.25	−17 – 28	−2.41	11.95	−30 – 25
Personal Living Skills	62.88	14.34	30 – 88	49.88	13.66	18 – 78	62.07	16.52	25 – 98	54.68	11.69	30 – 76	−0.81	7.98	−21 – 15	4.79	10.75	−12 – 29
Community Living Skills	57.86	20.33	20 – 87	58.97	14.58	29 – 85	50.26	19.40	5 – 90	46.26	18.64	4 – 81	−7.60	12.87	−46 – 16	−12.71	15.53	−51 – 26

Note. Difference = Time 2 SS – Time 1 SS.

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Table IV

Pattern of Individual Standard Score Change from Time 1 to Time 2 for Measures of Intellectual and Vocabulary Abilities and Overall Adaptive Behavior Skills

Measure	Pattern of SS Change from Time 1 to Time 2[a]							Significance of Change[a]		
	Any Decrease			Identical Score	Any Increase			Significant Decrease	Not Significant	Significant Increase
	%	Mean	SD	%	%	Mean	SD	%	%	%
DAS-II GCA	50.00	−6.53	4.43	3.95	46.05	5.23	3.48	6.58	89.47	3.95
DAS-II Verbal SS	52.63	−10.80	8.28	5.26	42.11	6.09	4.60	7.89	90.79	1.32
DAS-II Nonverbal Reasoning SS	64.47	−9.94	6.44	2.63	32.89	5.96	4.58	7.89	92.11	0.00
DAS-II Spatial SS	39.47	−6.43	5.48	2.63	57.89	8.57	7.82	2.63	80.26	17.11
DAS-II SNC	53.95	−6.78	4.08	3.95	42.11	7.53	5.55	7.89	80.26	11.84
PPVT-4 SS	64.47	−7.22	5.34	7.89	27.63	7.48	5.45	7.89	85.53	6.58
EVT-2 SS	67.11	−6.16	4.95	7.89	25.00	7.16	5.45	2.63	94.74	2.63
SIB-R Broad Independence SS	71.05	−10.65	8.05	3.95	25.00	5.68	4.53	42.11	51.32	6.58

Abbreviations: DAS-II (Differential Ability Scales-II), General Conceptual Ability (GCA; similar to IQ), SS (standard score), SNC (Special Nonverbal Composite; similar to Performance IQ), PPVT-4 (Peabody Picture Vocabulary Test-4), EVT-2 (Expressive Vocabulary Test-2)

[a]Standard error of measurement (dependent on the child's age and the assessment) was used to calculate the 95% confidence interval for SS at Time 1 and at Time 2. The change was considered significant if the 95% confidence intervals for Time 1 SS and Time 2 SS did not overlap.

Table V

Pattern of Individual Change for Adaptive Skills as Measured by the Scales of Independent Behavior-Revised (SIB-R) Clusters from Time 1 to Time 2 as a Function of Sex

Measure		Pattern of SS Change from Time 1 to Time 2							Significance of Change[a]		
		Any Decrease			Identical Score	Any Increase			Significant Decrease	Not Significant	Significant Increase
		%	Mean	SD	%	%	Mean	SD	%	%	%
Girls (N = 42)	Motor Skills	76.19	−8.94	8.59	4.76	19.05	4.88	3.60	9.52	90.48	0.00
	Social Interaction & Communication Skills	54.76	−6.83	4.96	11.90	33.33	7.00	6.63	0.00	97.62	2.38
	Personal Living Skills	47.62	−8.97	6.99	2.38	50.00	5.81	3.67	2.38	97.62	0.00
	Community Living Skills	73.81	−12.84	10.55	0.00	26.19	7.18	4.38	21.43	76.19	2.38
Boys (N = 34)	Motor Skills	64.71	−13.50	7.94	0.00	35.29	8.33	5.45	8.82	91.18	0.00
	Social Interaction & Communication Skills	61.76	−9.43	8.27	2.94	35.29	9.67	7.11	8.82	88.24	2.94
	Personal Living Skills	38.24	−4.69	4.44	5.88	55.88	11.79	8.92	0.00	82.35	17.65
	Community Living Skills	82.35	−17.25	12.59	0.00	17.65	8.50	9.03	38.24	58.82	2.94

[a]Standard error of measurement (dependent on the child's age and the assessment) was used to calculate the 95% confidence interval for SS at Time 1 and at Time 2. The change was considered significant if the 95% confidence intervals for Time 1 SS and Time 2 SS did not overlap.

Table VI

Descriptive Statistics for |Difference Scores| as a Function of Age Cohort

Measure	Younger Cohort (*N* = 39)			Older Cohort (*N* = 37)			Difference			Independent t-test		
	Mean	SD	Range	Mean	SD	Range	Mean	SE	95% CI	t (*df*)	*P*-value	*r* (Effect Size)
DAS-II GCA	7.05	4.45	0 – 19	4.22	3.18	0 – 14	2.84	.88	1.07 – 4.60	3.21 *(68.88)*	.002	.36
DAS-II Verbal SS	9.03	8.09	0 – 34	7.43	6.41	0 – 21	1.59	1.67	−1.73 – 4.92	0.95 *(71.75)*	.343	.11
DAS-II Nonverbal Reasoning SS	10.31	6.44	0 – 28	6.32	5.33	0 – 21	3.98	1.35	1.29 – 6.68	2.94 *(72.70)*	.004	.33
DAS-II Spatial SS	9.90	8.42	0 – 26	4.97	3.88	0 – 16	4.92	1.49	1.93 –7.91	3.30 *(54.10)*	.002	.41
DAS-II SNC	8.59	5.16	0 – 21	4.97	3.78	0 – 16	3.62	1.03	1.55 –5.68	3.50 *(69.62)*	.001	.39
PPVT-4 SS	7.72	5.51	0 – 17	5.68	5.34	0 – 19	2.04	1.24	−0.44 –4.52	1.64 *(73.96)*	.105	.19
EVT-2 SS	6.77	5.65	0 – 22	5.03	4.51	0 – 19	1.74	1.17	−0.59 –4.07	1.49 *(71.92)*	.141	.17
SIB-R Broad Independence	11.92	8.60	0 – 40	5.89	5.02	0 – 20	6.03	1.61	2.82 –9.24	3.76 *(61.80)*	< .001	.43
SIB-R Motor Skills	12.08	9.37	1 – 45	6.78	4.97	0 – 19	5.29	1.71	1.87 –8.71	3.10 *(58.42)*	.003	.38
SIB-R Social Interaction & Communication Skills	8.51	7.98	0 – 30	6.41	5.33	0 – 25	2.11	1.55	−0.98 –5.20	1.36 *(66.58)*	.178	.16
SIB-R Personal Living Skills	8.97	7.76	0 – 29	5.76	4.00	1 –16	3.22	1.41	0.40 – 6.03	2.29 *(57.54)*	.026	.29
SIB-R Community Living Skills	18.46	12.00	3 – 51	7.86	6.69	1 – 30	10.60	2.21	6.17 – 15.02	4.79 *(60.16)*	< .001	.53

Note. Variances were assumed to be unequal for the younger cohort (< 7.5 years old at Time 1) and the older cohort (> 7.5 years at Time 1).

Abbreviations: CI (confidence interval), DAS-II (Differential Ability Scales-II), General Conceptual Ability (GCA; similar to IQ), SS (standard score), SNC (Special Nonverbal Composite; similar to Performance IQ), PPVT-4 (Peabody Picture Vocabulary Test-4), EVT-2 (Expressive Vocabulary Test-2), SIB-R = Scales of Independent Behavior-Revised)

Table VII

Correlations between Standard Scores (SS) at Time 1 and Time 2

Measure	Overall (*N* = 76)	Younger Cohort (*N* = 39)	Older Cohort (*N* = 37)
DAS-II GCA	.82	.71	.92
DAS-II Verbal SS	.76	.63	.85
DAS-II Nonverbal Reasoning SS	.71	.52[a]	.84
DAS-II Spatial SS	.68	.55	.87
DAS-II SNC	.77	.65	.89
PPVT-4 SS	.83	.70	.89
EVT-2 SS	.84	.79	.88
SIB-R Broad Independence	.84	.79	.91
SIB-R Motor Skills	.76	.68	.86
SIB-R Social Interaction & Communication Skills	.77	.71	.83
SIB-R Personal Living Skills	.80	.77	.88
SIB-R Community Living Skills	.70	.73	.84

Note. Younger Cohort = < 7.5 years old at Time 1. Older Cohort = > 7.5 years old at Time 1.

[a] $P = .001$; all other Ps < .001.

Abbreviations: DAS-II (Differential Ability Scales-II), General Conceptual Ability (GCA; similar to IQ), SS (standard score), SNC (Special Nonverbal Composite; similar to Performance IQ), PPVT-4 (Peabody Picture Vocabulary Test-4), EVT-2 (Expressive Vocabulary Test-2), SIB-R (Scales of Independent Behavior-Revised)