Intellectual Assessment of Students with Williams Syndrome... Common Profiles and Testing Tools

by Carolyn Mervis, Ph.D.
University of Louisville

Different tests provide different information, and some tests are better than others for documenting the intellectual strengths and weaknesses of children with Williams syndrome (WS). Several tests that are commonly used when assessing children with WS are described below. The "right" test will vary for students with WS at different ages, and the recommended tests for students with WS may be different from the tests that are typically given by school psychologists. Therefore it is very important for families to ask what tests will be given and provide school districts with the following information to help insure that your child's team has the best possible information.

A Quick Primer on Intellectual Assessment Test Scores

Intellectual ability tests measure the abilities of an individual child relative to the abilities of children in the general population. (For well-normed tests intended for children in the United States, this means that the group of children used to norm the assessment was matched to the U.S. census proportions in terms of characteristics such as race, ethnicity, parents' level of education, rural vs. suburban vs. urban residence, and geographical region of the U.S. in which the child lived. Children with disabilities are usually represented in the norming sample in the same proportions as for children in the U.S. population.)

In order to compare an individual child's performance on a test to the test's norms (that is, relative to children in the general population), standard scores (with an average score of 100 and a standard deviation of 15), T-scores (with an average score of 50 and a standard deviation of 10), and/or scaled scores (with an average score of 10 and a standard deviation of 3) are used. All of the tests that are discussed below provide an overall IQ score that is expressed as a standard score. However, performance on the separate components of the test is often expressed as either a T-score or a scaled score.

The likelihood that a particular test will accurately capture the abilities of a child with a disability is related in part to what the lowest possible score on each subtest is. For example, a test that reports T-scores for the subtests will be better able to capture the performance of a child who has significant difficulty in the area the subtest measures if the lowest possible T-score is 10 than if the lowest possible T-score is 20. If the lowest possible T score is 20, then this score will be assigned to everyone who would have earned T-scores of 10 – 19, in addition to children who would have earned a T-score of 20. At the present time, the lowest scaled score on tests of intellectual ability that use this type of measure rather than T-scores is 1, which corresponds to a T-score of 20.

Why does it matter what the lowest possible score on a subtest is? First, it may affect the services that a child is offered. A T-score of 20 corresponds to mild-moderate disability, whereas a T-score of 10 corresponds to moderate-severe disability. Children with more severe disabilities are often provided more intensive services than children with mild disabilities. Second, if T-scores as low as 10 are available on a test, then it is more likely that an individual child's pattern of strengths and weaknesses will be made clear than if the lowest possible T score is 20.

For whom is this likely to matter? In general, this will matter most for children with WS who have a moderate or severe disability. It will also matter for many children who have a mild disability or are in the borderline range (the range between mild disability and low average ability), especially if they have a clear area of weakness (most likely visuospatial construction). It is less likely to matter for children whose intellectual abilities are in the low average or average range.

Sometimes parents are given the results of standardized assessments in the form of age equivalents (e.g., "5 years 6 months"). These scores appear to offer a lot of information, but in fact they are quite problematic, for several reasons. First, age equivalent scores do not mean what they appear to mean. That is, you should not automatically consider your child to be functioning at the age that corresponds to the age equivalent score. Second, age equivalent scores do not tell parents whether or not their child's performance is in the range expected for children his or her age. A given child's age equivalent score indicates the median (middle) age at which the children in the group used to norm the test obtained the same number of points as that child did. Depending on the particular test or subtest, a very wide range of age equivalents may be within the average range for that child's chronological age. For example, on a well-regarded academic achievement test, age equivalents between 7 years 8 months and 11 years 0 months are considered to be in the average range for a child aged 9 years 0 months on the subtest measuring ability to read English words. For a child aged 12 years 0 months given the same subtest, age equivalents between 9 years 4 months and >19 years 11 months (the highest possible age equivalent on this test) would be considered to be in the average

range. Third, age equivalent scores cannot be compared across different tests or even across subtests included in the same test. That is, if your child received a higher age equivalent score on one subtest than on another, that does not necessarily indicate that he or she actually performed better on the subtest with the higher age equivalent score than on the subtest with the lower age equivalent score. The only accurate way to compare a child's ability on one subtest or test to his or her ability on a different subtest or test is to compare standard scores, T-scores, scaled scores, or percentile scores.

Assessments for Children - Infant through Preschool

In the very early years, the *Mullen Scales of Early Learning (MSEL)* is a good choice. It is normed for children from 1 month to 5 years 8 months. For children with WS, it is best used for 18 – 48-month-olds.

Advantages for children with WS:

The MSEL yields scores for 4 separate ability areas (nonverbal reasoning – called Visual Reception on the MSEL – receptive language, expressive language, and visuospatial construction – called Fine Motor on the MSEL). For this reason, the MSEL can show a child's patterns of strength and weakness, rather than simply providing a single score averaging different types of abilities together. Children with WS often have the most

difficulty with visuospatial construction, and the MSEL Fine Motor scale highlights this need area. Goals for the OT and classroom teacher will often come to light as well as challenges that will have to be overcome.

Disadvantages: The MSEL norms do not have a large enough range to show differences for children who are having considerable difficulty in a particular ability area or overall. This is because the lowest possible T-score on each scale is 20. A very large group of children with WS earn the lowest possible scaled score (T-score), referred to as the "floor," on one or more of the four scales. For children who are having considerable difficulty in more than one ability area, relative strengths and weaknesses will be camouflaged. Note that this problem occurs with all tests that are currently available for toddlers and young preschoolers. An additional problem with the MSEL is that small differences in the age of the child at the time he or she is tested can create artificially large differences in scores.

Assessments for Preschool and School Age Children

For children aged 4 – 17 years, the Differential Ability Scales-II (DAS-II) is an excellent test. There are two versions of the DAS-II, the Early Years version for ages 2 ½ years to 8 years 11 months *(not recommended for children with WS who are younger than 4 years),* and the School Age version for children 5 years to 17 years 11 months. *The Early Years version should be used for children with WS aged 4 years – 8 years 11 months. Extended norms are also available for older children to allow for the use of the Early Years version for children aged 9 – 17 years who have moderate or severe disabilities.*

Advantages for children with WS:

The lowest T-score available for the DAS-II is 10, which means that the DAS-II is able to accurately characterized the abilities of children with WS even if they

have a severe disability. Therefore, the uneven pattern of strengths and weaknesses typical for children with WS can be clearly identified by comparing standard scores for the 3 core clusters of this test - the Verbal Reasoning, the Nonverbal Reasoning and the Spatial clusters. In addition, three diagnostic clusters that are not included in the IQ score also are available: School Readiness (for ages 5 years 0 months – 8 years 11 months), Working Memory (recommended for ages 7 years 0 months – 17 years 11 months), and Processing Speed (also recommended for ages 7 years 0 months – 17 years 11 months).

Determining an IQ score from the DAS-II

The DAS-II GCA, which is like an IQ score, is meaningful if the 3 core cluster scores do not differ significantly. However, this is the case for < 10% of children with WS. About 90% of children with WS show a clear pattern of higher scores on the Verbal cluster and/or the Nonverbal Reasoning cluster but substantially lower scores on the Spatial cluster. Only 2% showed substantially higher scores on the Spatial cluster than the Verbal cluster. When children's cluster standard scores are uneven (differ significantly), which is true for about 90% of children with WS, a single score such as an IQ or GCA is not the best estimate of their intellectual abilities. Instead, each cluster standard score should be considered separately.

Other Commonly Used Tests

School psychologists and private psychologists usually use Wechsler tests to measure children's intellectual abilities. The most commonly used versions are described below.

Wechsler Preschool & Primary Scale of Intelligence (WPPSI-IV)

The WPPSI-IV is normed for ages 2 years 6 months – 7 years 7 months (not recommended for children less than 4 years old who have developmental delay). It is completely redesigned and is more similar to the DAS-II than the WPPSI-III was. In particular, instead of a single Index that includes both spatial and nonverbal reasoning subtests, the WPPSI-IV has a Visual Spatial Index (similar to the DAS-II Spatial cluster) and a Fluid Reasoning Index (similar to the DAS-II Nonverbal Reasoning Index).

However, the WPPSI-IV is not normed as low as the DAS-II is. In particular, the lowest possible scaled score on the WPPSI-IV subtests is 1, which corresponds to a T-score of 20. This means that the WPPSI-IV is not normed low enough to accurately capture the abilities of children

who have moderate to severe disability. It also is likely not normed low enough to accurately capture significant differences between performance on the Visual Spatial Index and performance on the Fluid Reasoning Index or the Verbal Comprehension Index for children with mild disability.

Wechsler Intelligence Scale for Children - IV (WISC-IV)

The WISC-V, which is normed for children aged 6 years 0 months – 16 years 11 months, was released in 2016. For children aged 6 years 0 months – 7 years 7 months, either the WPPSI-IV or the WISC-V may be administered. It is recommended that the WPPSI-IV rather than the WISC-V be administered to children with WS in this age interval.

The structure of the WISC-V is the same as that of the WPPSI-IV. Thus, unlike the WISC-IV, the WISC-V includes a Visual Spatial Index (similar to the DAS-II Spatial cluster) and a Fluid Reasoning Index (similar to the DAS-II Nonverbal Reasoning cluster). For children with WS, this is a considerable improvement over the WISC-IV Perceptual Reasoning Index, which included both spatial and nonverbal reasoning subtests.

The lowest possible scaled score on the WISC-V is 1, corresponding to a T-score of 20. Thus, the limitations described for the WPPSI-IV with regard to testing children with WS also apply to the WISC-V.

Some schools or private psychologists may still be administering the WISC-IV. If the WISC is to be administered to a child with WS, it is important that the WISC-V rather than the WISC-IV be used.

There has been no research published on the performance of children with WS on either the WPPSI-IV or the WISC-V.

Kaufman Brief Intelligence Test - 2 (KBIT-2)

The KBIT-2 is the most commonly used assessment in American research on Williams syndrome. The test is normed for people ages 4 - 90 years. It includes both Verbal and Nonverbal (Matrices) scales. It is not normally used by school psychologists and is not recommended for assessments for educational purposes.

Advantages: The KBIT-2 provides an IQ estimate that does not include visuospatial construction (the area of greatest weakness for most people with WS). Additionally it takes much less time to complete than either the DAS-II or the Wechsler tests.

Disadvantages: The KBIT-2 is a "brief" assessment and therefore does not provide the same depth of assessment of verbal and nonverbal reasoning that is derived from the DAS-II, the WPPSI-IV, or the WISC-V. Additionally, the KBIT-2 does not provide estimates of spatial ability, working memory, or processing speed.

"Typical" Cognitive Characteristics of Children with WS

While there is substantial variation, children with WS most commonly have overall intellectual abilities which fall in the Borderline to Mild Intellectual Disability range. On the DAS-II, children with WS usually present with an uneven profile. Typically children with WS have relative strengths in language and nonverbal reasoning and significant weakness in visuospatial construction. This area of difficulty impacts writing, drawing, pattern construction, relational language, and mathematics.

Relational language concepts are generally very difficult for children with WS. These include spatial terms (e.g., behind, between), temporal terms (e.g., before, after), quantitative terms (e.g., most, least), and comparative adjectives (e.g. shortest, tallest), as well as more complex relational terms (e.g., neither, nor, unless). This difficulty can be confusing to a conversational partner, if the child with WS otherwise understands and uses relatively complex grammatical constructions and has a large concrete vocabulary (labels for objects, actions, and descriptors).

Relational language can be addressed in the IEP and included as an SLP goal and should be practiced during the day by the teacher/assistant. While work on these goals is important and children do make progress in this area, awareness of the difficulty and provisions for extra clarification (e.g. visual supports; additional verbal cues) is also helpful in working with students with WS.

Language pragmatics is another area of need for students with WS and it is important that goals in the child's IEP address this area. This difficulty is often most apparent in the area of social skills. Another pragmatic difficulty for most children with WS is that they often do not realize that they did not understand (or misunderstood) what their conversational partner or teacher has said. Even if they do realize that they did not understand what was said, they may not ask for clarification. Pragmatics is often addressed in Speech therapy but it is also key to address this component of language with peers in social skills groups and throughout the child's day including during unstructured social times (e.g., recess, lunch).

Fine Motor Skills

Fine motor and visuospatial construction tend to be areas of significant struggle. These areas should be addressed in the student's IEP as well as with accommodations in classwork. In particular, when the purpose of an assignment is conceptual, a child with WS should be allowed to complete the assignment orally (or, for older children who type well, using the computer). This accommodation will allow the child to perform at his or her level of conceptual ability rather than being constrained by his or her difficulties with handwriting or typing. Almost all children with WS will eventually learn to print legibly and it is important that they be given regular opportunities to work on their printing. However, it is every bit as important that this skill area difficulty not be allowed to interfere with other aspects of the child's learning.

It is important that children with WS be taught to use technology such as Smart Boards, iPads, and laptops early in their education and that they be allowed to complete assignments using these tools. Assistive Technology assessment and supports are extremely beneficial for accessing curriculum, especially working around visuospatial challenges and difficulties with executive functioning (see below).

Academic Achievement

The Wechsler Individual Achievement Test-III (WIAT-III) is generally a good test to use for those with WS. Most children with WS perform much better on the Reading than the Math scales. Math is generally well below what would be expected based on IQ.

Advantages of the WIAT-III for testing students with WS

For most children with WS, the differences in performance on the composites clearly separate the typical relative strength in Reading and relative weakness in Math. Within Reading, the composites typically separate the pattern of relative strength in single word reading but relative weakness in reading comprehension and fluency shown by many children with WS who have good decoding skills. Finally, the floor for the Basic Reading and Oral Language scales is low enough to accurately characterize performance of most children with WS.

Disadvantages

The Oral Reading Fluency standard score is difficult to interpret for many children with WS because they tend to skip over words that they do not recognize immediately, which shortens their reading times, artificially inflating their standard scores. Additionally, the floor for the Reading Comprehension subtests and the Math and Written Language scales is not low enough.

Support in math is important to have on most children's IEPs. It is important to note that there are a few children with WS who have relatively good math skills. However for almost all children with WS, math has multiple challenges and becomes both frustrating and not meaningful over the grades. Working on basic math concepts that are useful in everyday life (e.g. more and less, basic addition and subtraction, concepts of time

and money) can be very worthwhile. However extensive time spent on advanced math concepts for a child who is not making gains is often not a productive use of the child's time. Other skills are generally much more important across multiple domains (e.g. reading decoding and reading comprehension, social skills) than are higher level math skills (e.g. geometry, algebra). Teaching children to use functional math, including schedules, planners, digital clocks and calendars, often with the support of technology tends to be the most useful.

Teaching reading through phonics rather than through sight words or whole language approaches generally leads to more advanced reading ability for all children. This is especially true for children with WS. Even if a school typically uses a whole-word or whole language approach to teaching reading, a child with WS should instead be taught with a systematic phonics-based program plus additional work focused on reading comprehension as this approach is most likely to lead to better and earlier reading ability.

Reading comprehension is often an area of weakness and is important to work on. Difficulties with reading comprehension have multiple causes, including problems with working memory, problems with relational or nonliteral language, problems with the verbal and nonverbal reasoning abilities needed to make inferences, and problems with executive functioning (see below). Difficulties with picking out the main idea versus tangential content, making inferences, figuring out complicated motivations of characters, and abstract reasoning all impact reading comprehension as well as social skills and comprehension of complex social situations.

Executive Functioning Difficulties

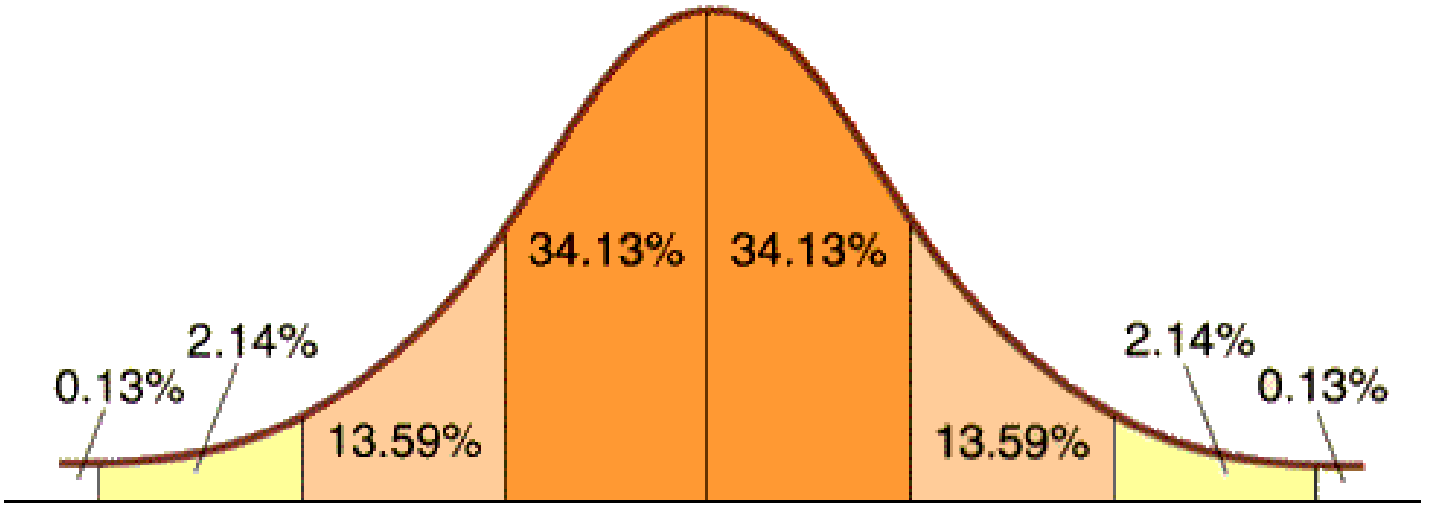
The term "Executive Functioning" refers to a set of skills that includes both behavioral regulation (e.g., inhibiting one's first response when it is inappropriate, controlling one's emotions) and metacognition (e.g., determining the steps needed to complete a task, organizing the materials needed, keeping the relevant steps in mind while carrying out the task, monitoring how well one is completing the task and making needed changes to successfully complete the task). Children in the general population who have ADHD generally have difficulties in these areas, and almost all children with WS have considerable difficulties with metacognition. Many children with WS also have significant difficulties with behavioral regulation. Difficulties with visuospatial abilities, relational concepts of space and time, and abstraction also impact these areas. Executive functioning difficulties can make the following sorts of school tasks very difficult:

- Organizing and keeping track of the "things" of school (e.g., paper, books, pens, food, etc.)
- Planning for what is needed and making sure the needed materials are available
- Time management around homework or classroom work
- Remembering to turn in homework

- Determining the main idea of a story, oral presentation by a teacher or another student, assignment, or projects
- Sticking to a task or project from the beginning of the task through to the end

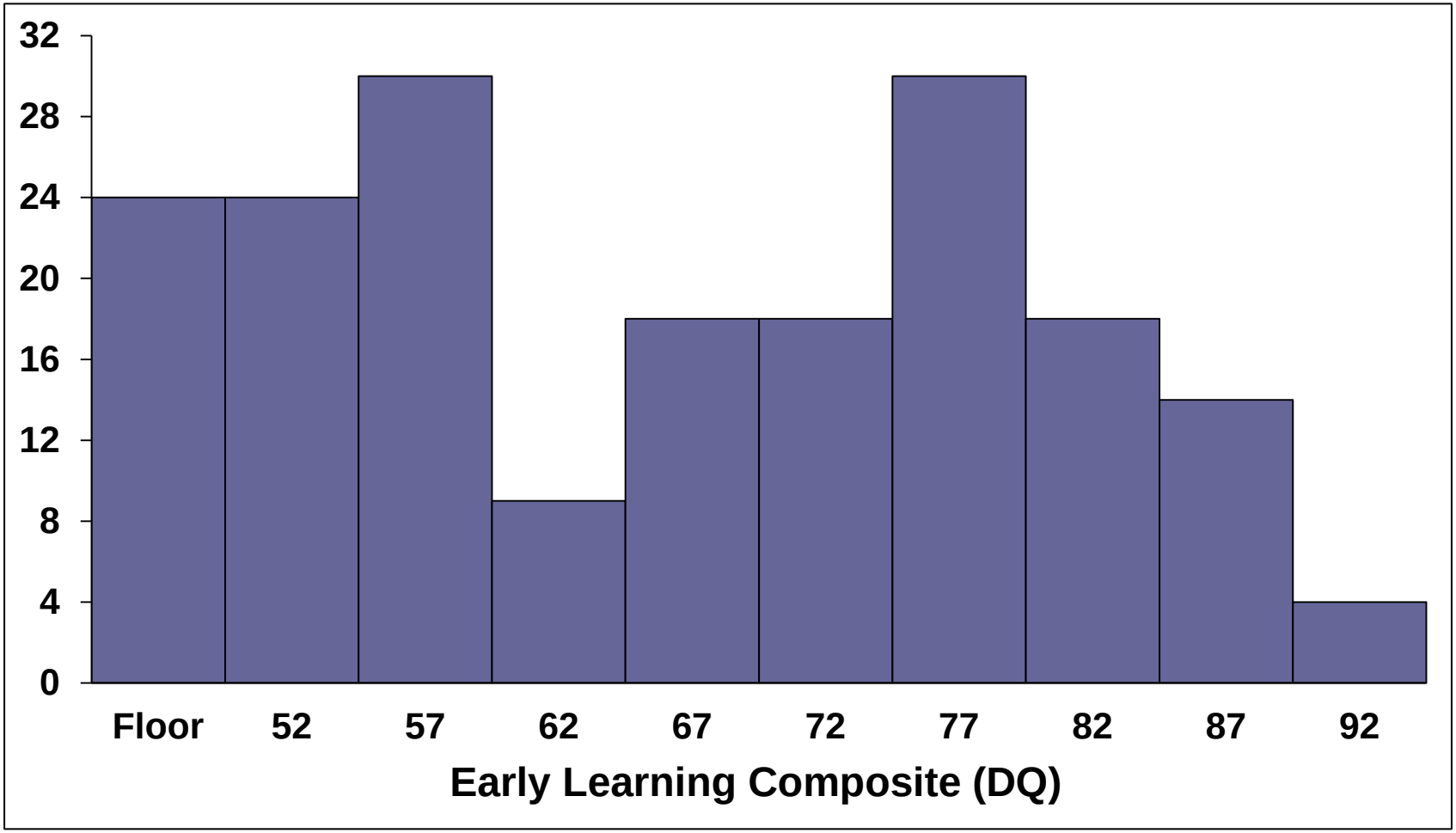
Children with WS generally make progress in these areas as they get older, but direct teaching and supervision is critical. Including work in these areas on the child's IEP and providing support and accommodations throughout the day are crucial.

Scores on Intellectual Assessments



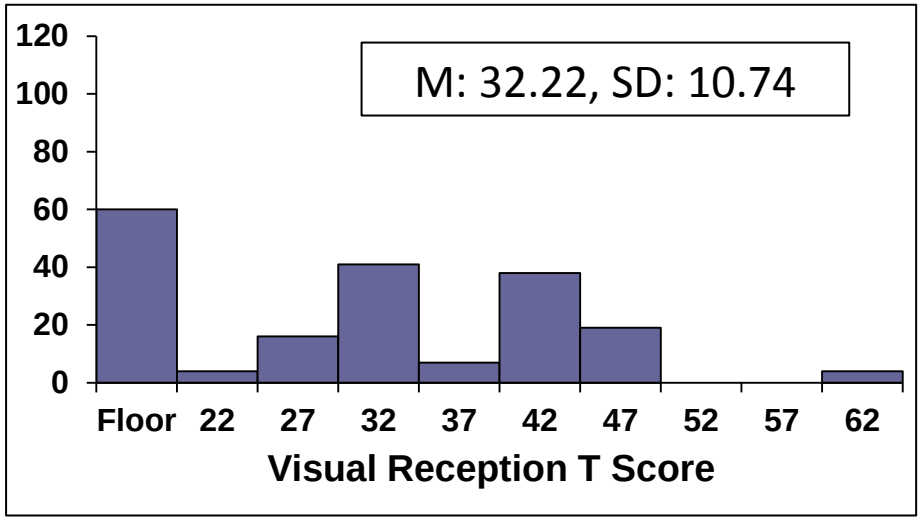
IQ	**40**	55	70	85	100	115	130	145
T	**10**	**20**	**30**	**40**	**50**	**60**	**70**	**80**
Scaled Score	**1**		**4**	**7**	**10**	**13**	**16**	**19**
Percentile								
	< 0. 1	**0.1**	**2**	**16**	**50**	**84**	**98**	**99.9**

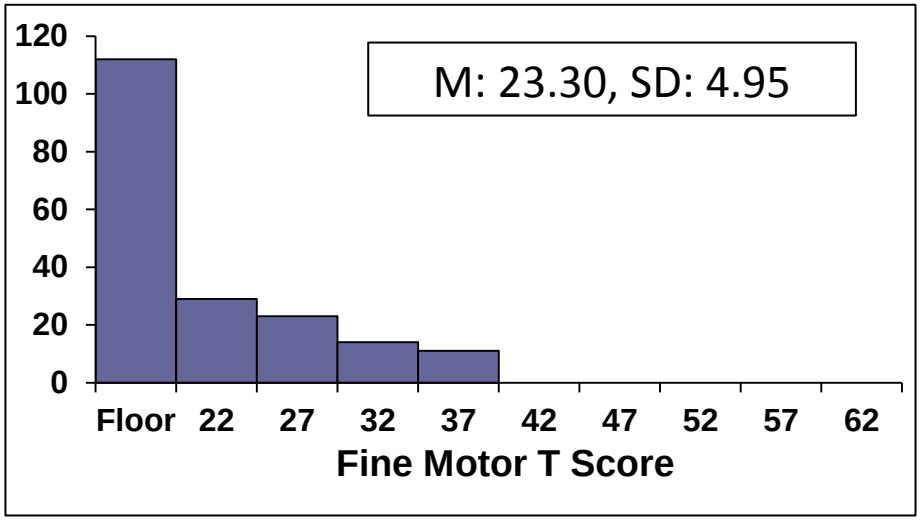
Mullen Scales of Early Learning: Early Learning Composite (DQ) (WS, Ages 18 – 48 months, N = 189)

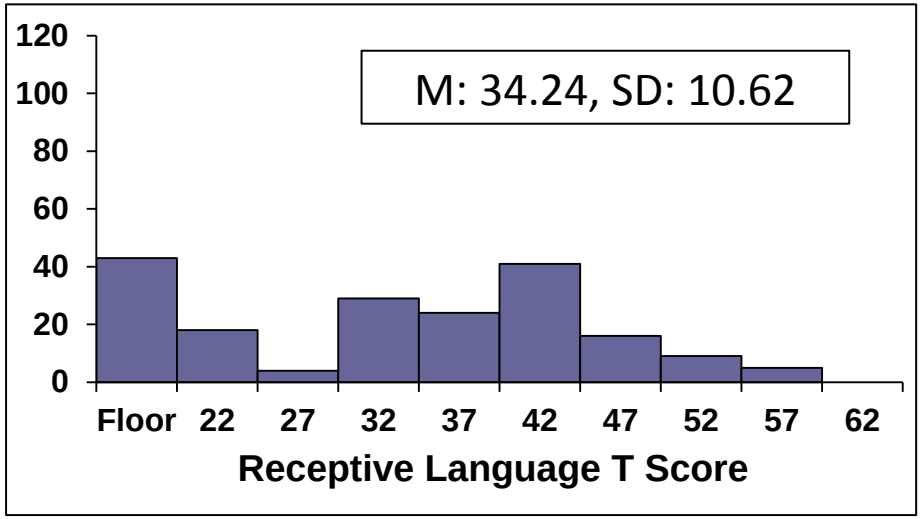


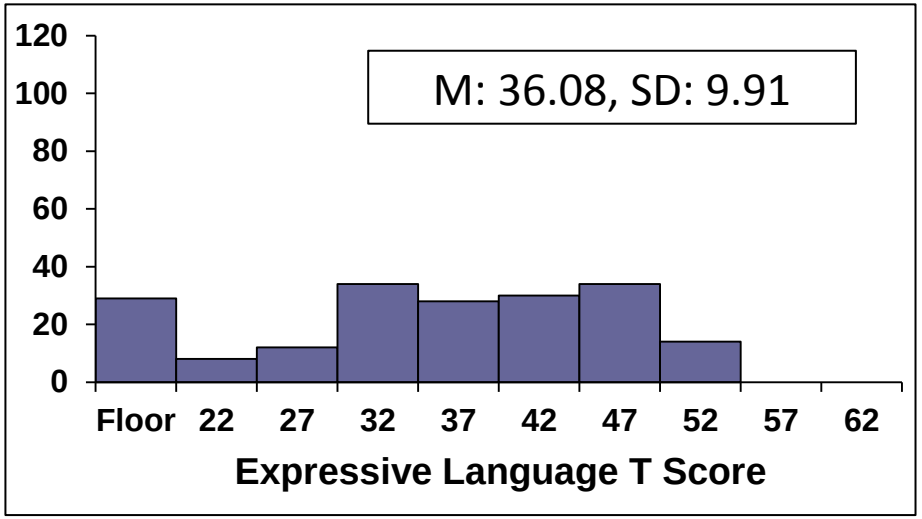
Mean: 66.58, SD: 12.91, Range: 49 (lowest possible) – 90

Mullen Scales of Early Learning: Scale *T* Scores (WS, Ages 18 – 48 months, N = 189)

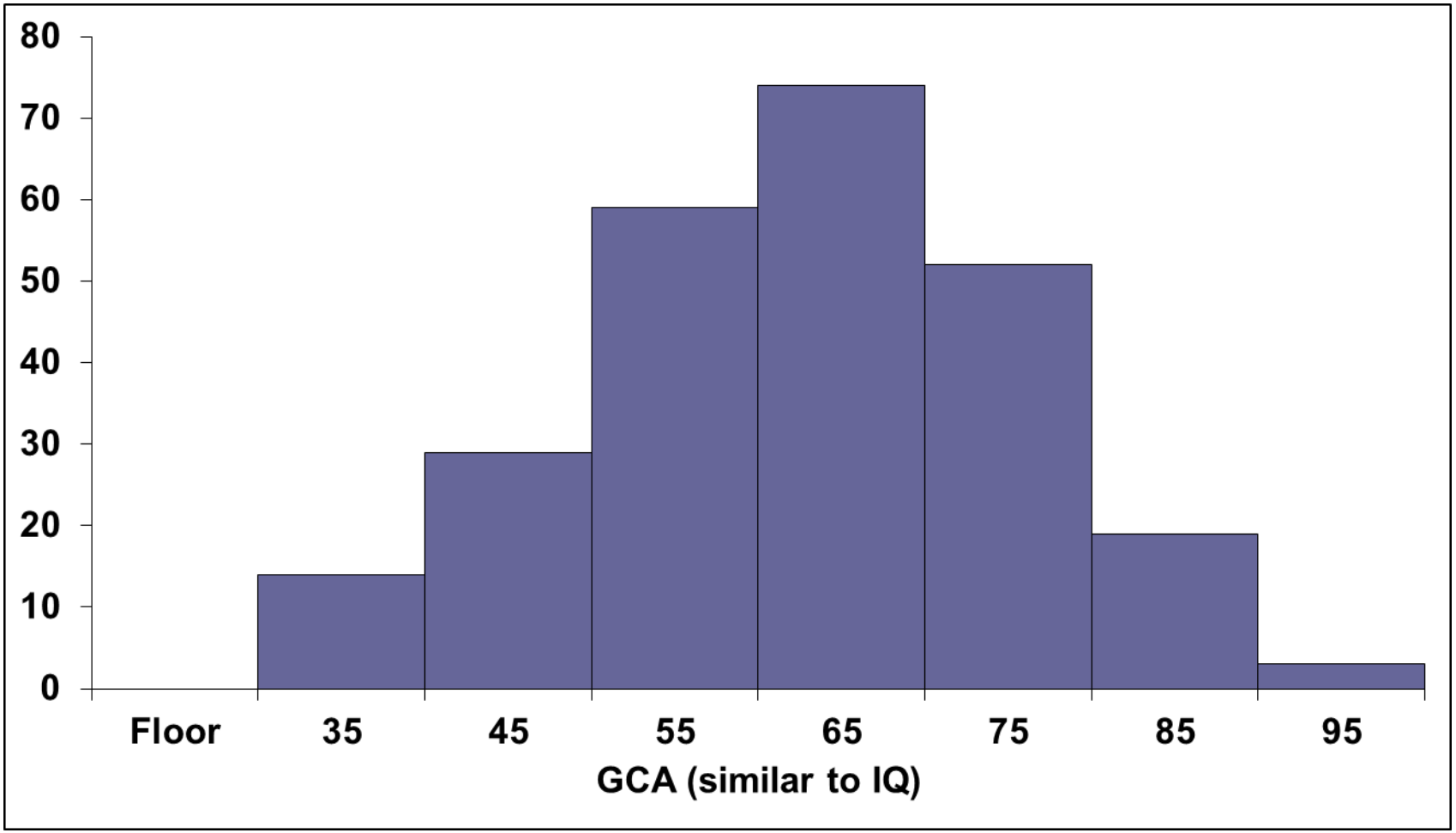






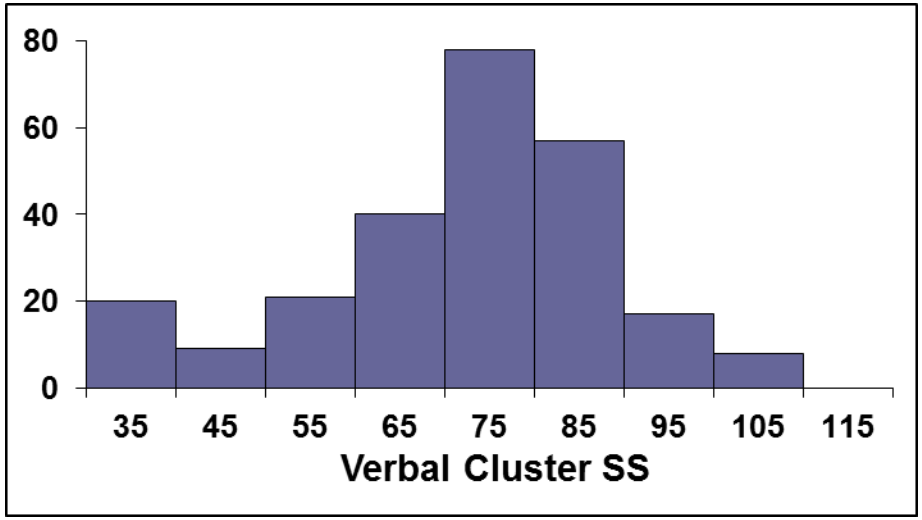


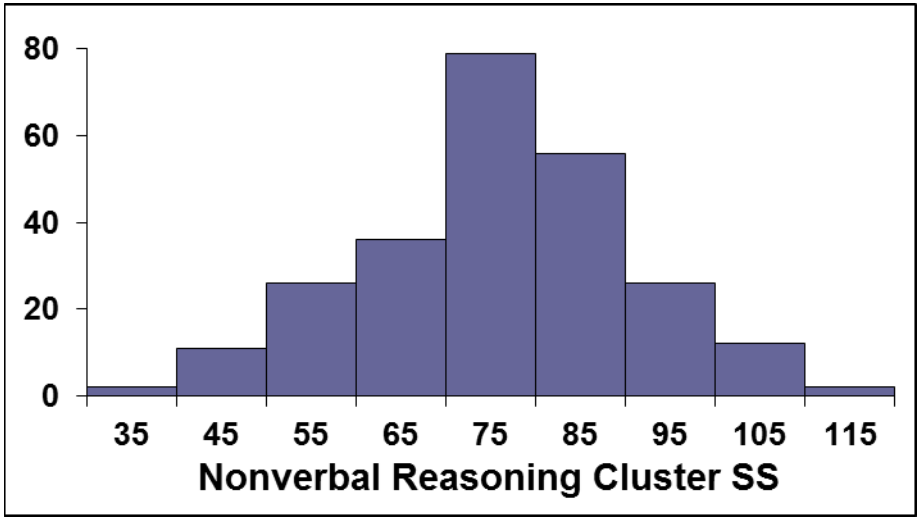
Differential Ability Scales-II GCA ("IQ")
(Ages 4 – 17 Years, *N* = 250)



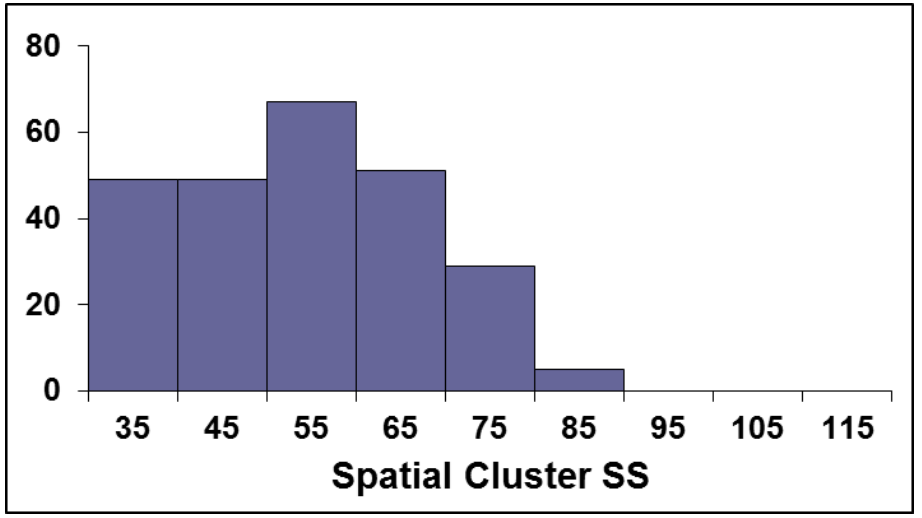
Mean: 62.31, SD: 12.97, Range: 31 – 96

DAS-II Core Cluster Standard Scores (Ages 4 – 17 Years, *N* = 250)

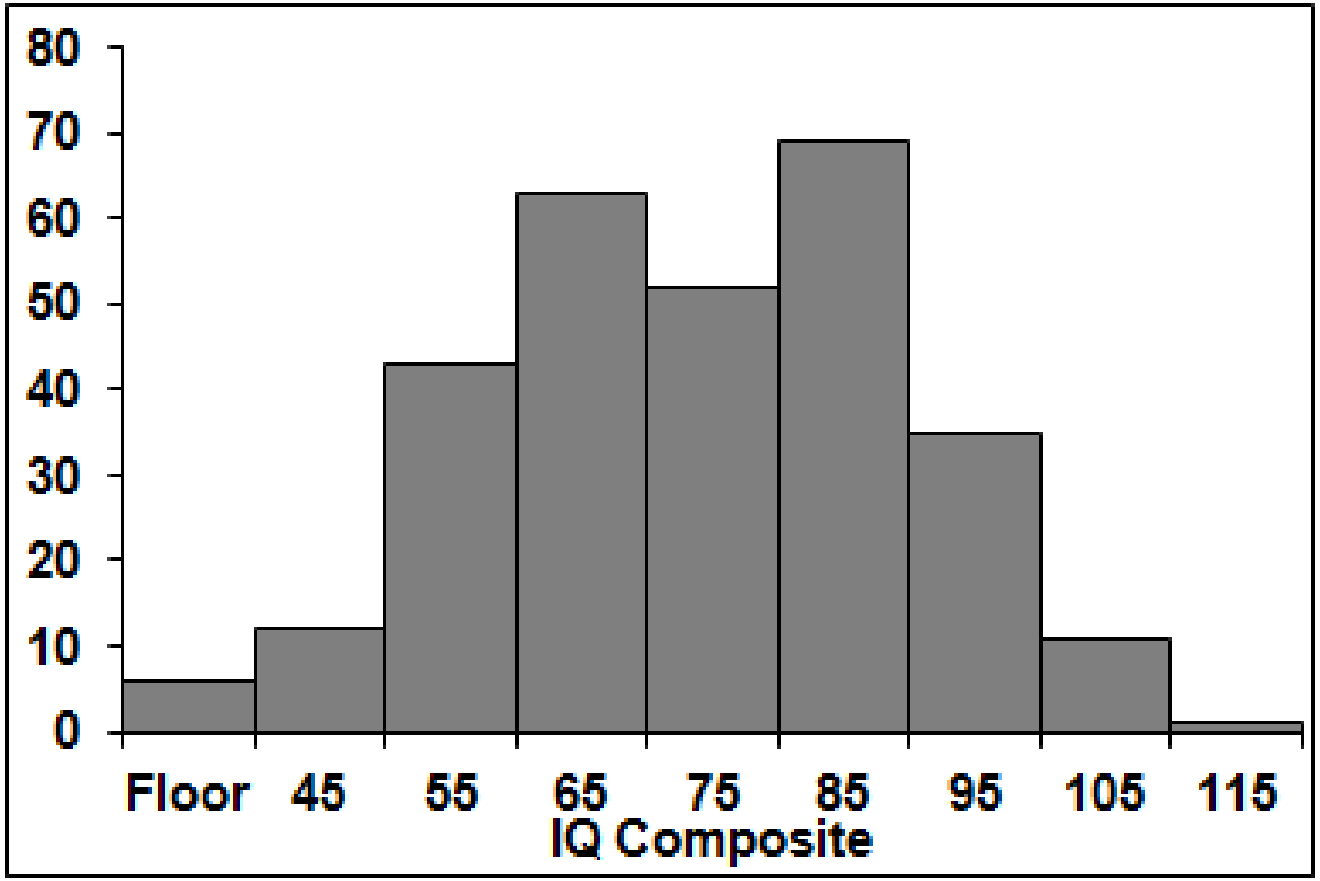




Cluster Standard Score	Mean (SD, Range)
Verbal	71.40 (16.68, 30 – 106)
Nonverbal Reasoning	75.64 (14.98, 37 – 118)
Spatial	53.35 (13.32, 32 – 86)

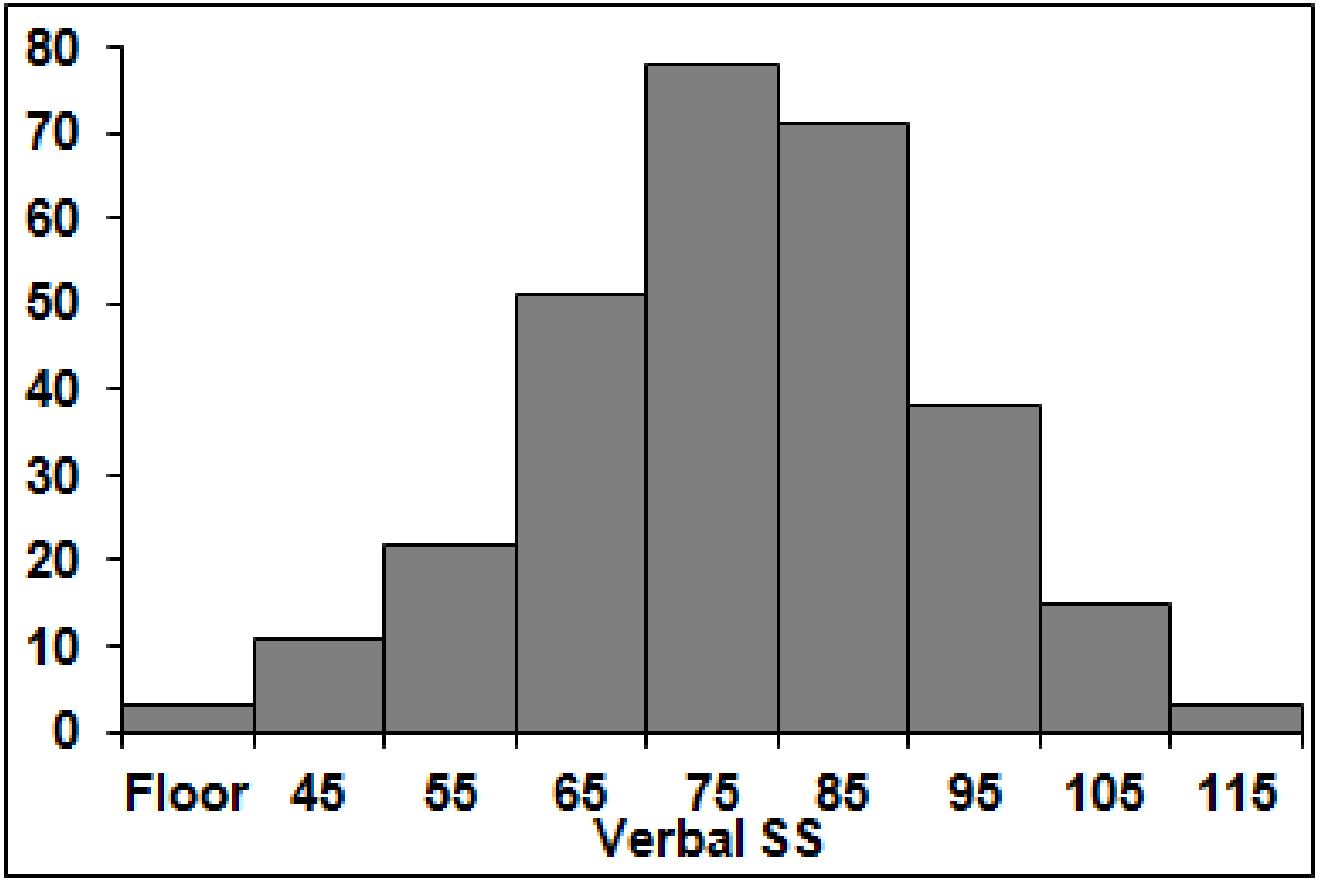


Kaufman Brief Intelligence Test – 2
IQ Composite (Ages 4 – 17 years, N = 292)



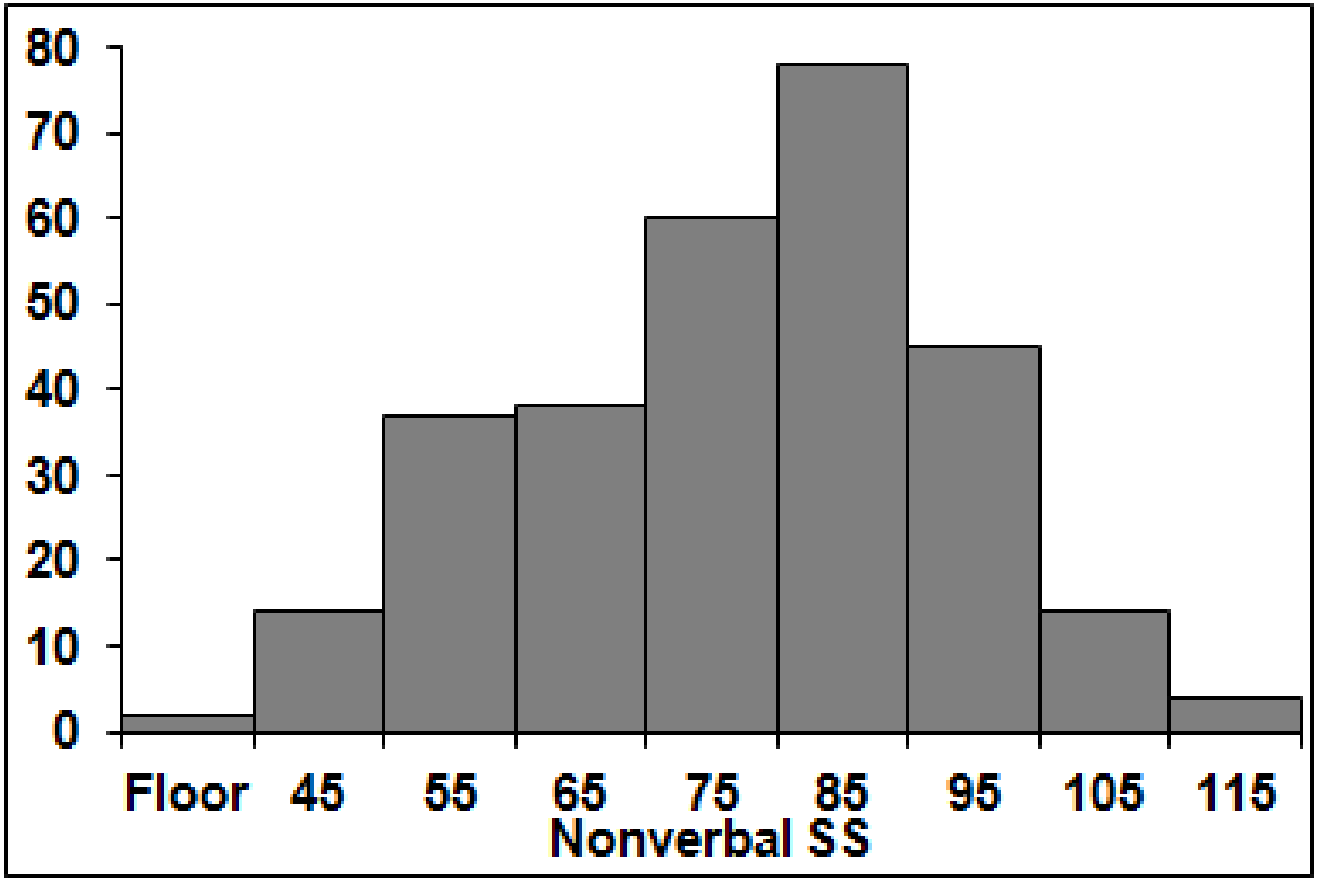
Mean: 73.50, SD: 15.44, Range: 40 – 111

Kaufman Brief Intelligence Test – 2
Verbal Standard Score (Ages 4 – 17 years, N = 292)



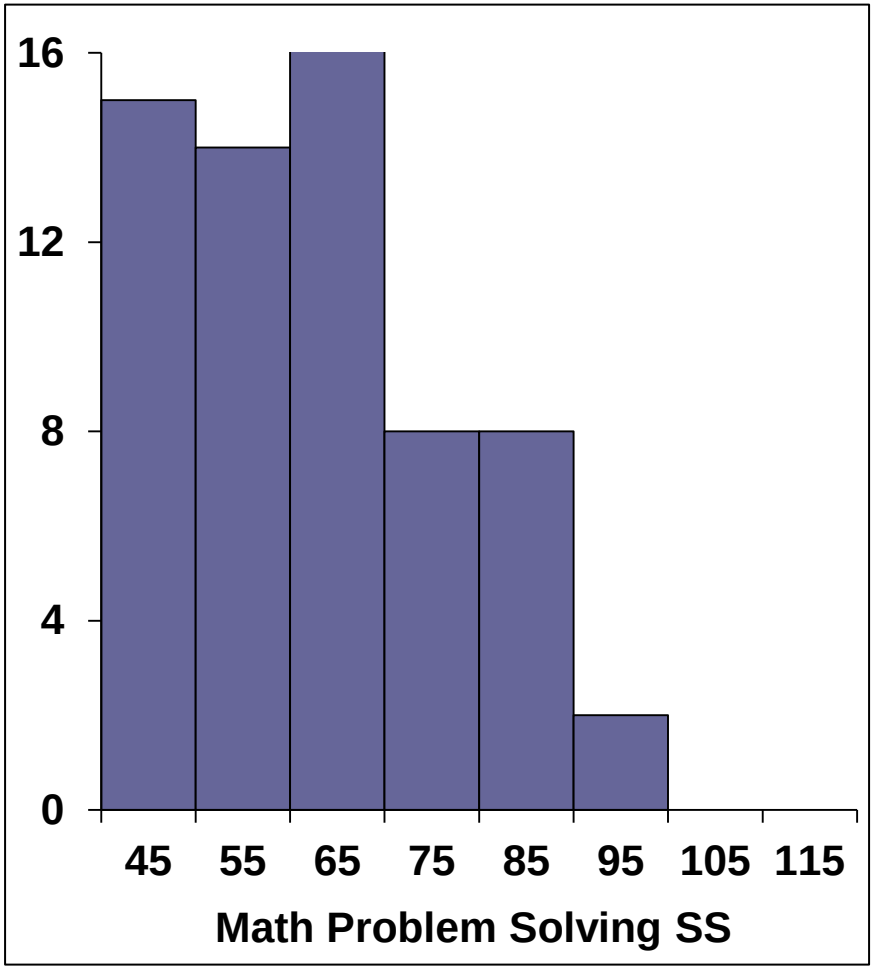
Mean: 76.57, SD: 14.81, 40 – 112

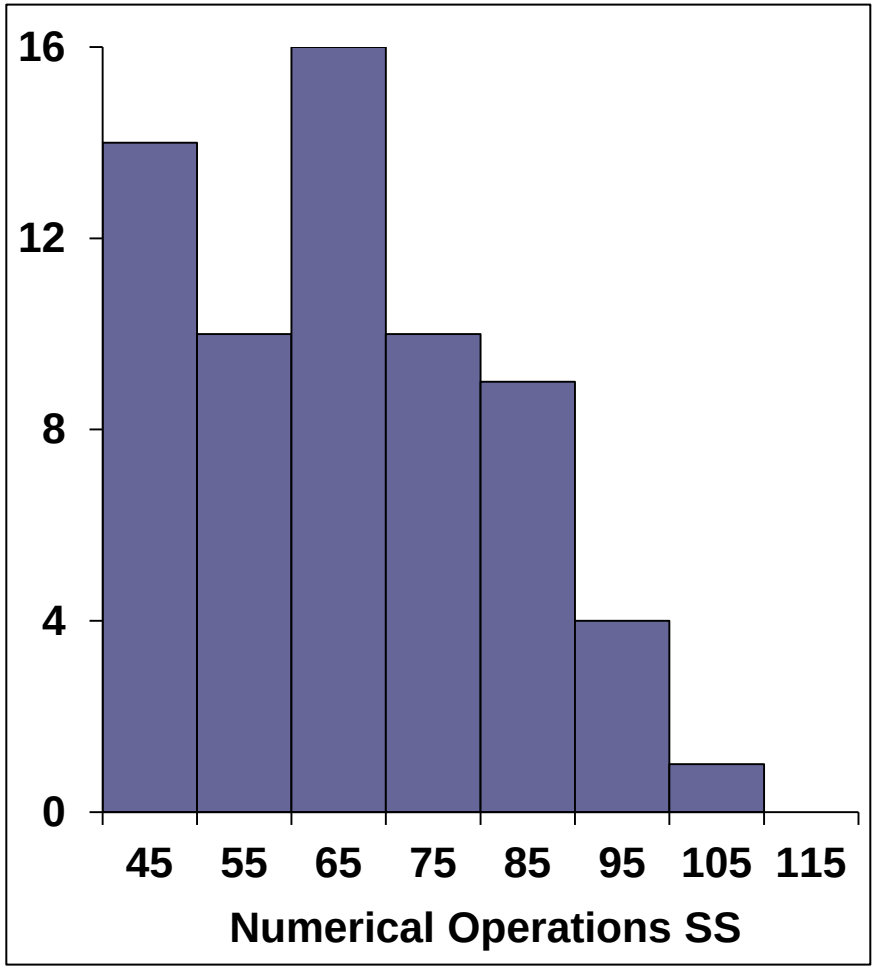
Kaufman Brief Intelligence Test – 2
Nonverbal Standard Score (Ages 4 – 17 years, N = 292)



Mean: 76.78, SD: 15.82, Range: 40 – 112

WIAT-III Math Standard Scores (Ages 9 – 17 Years, N = 74)

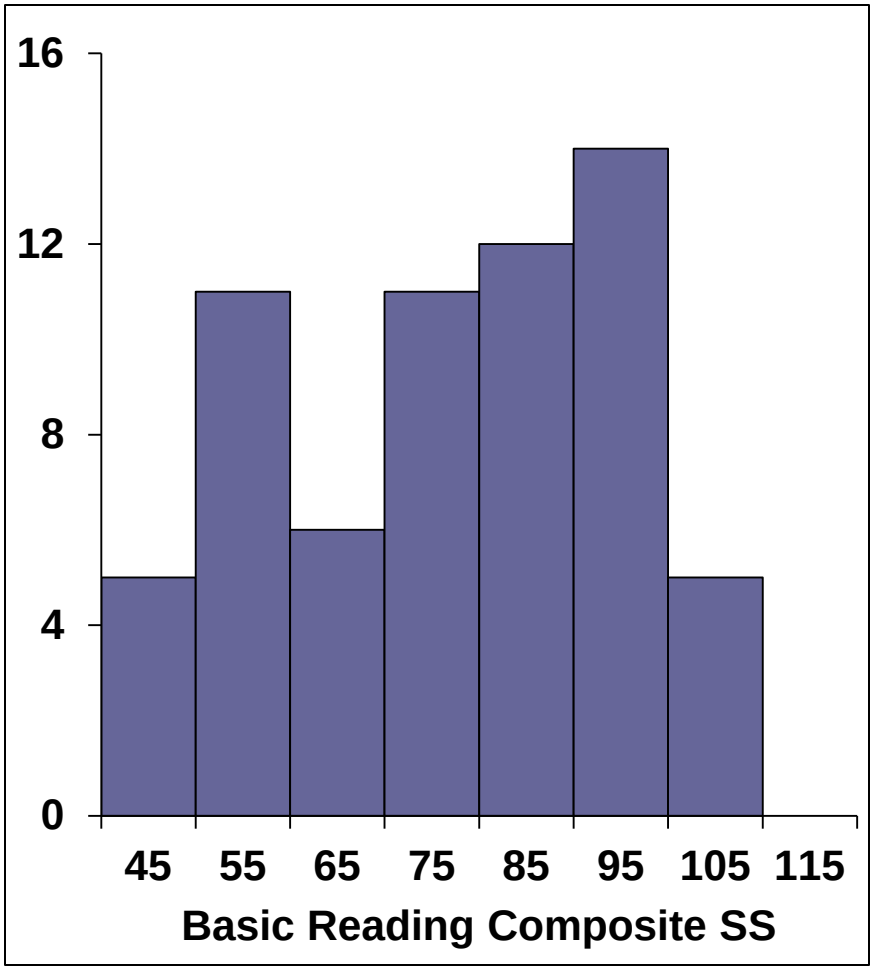




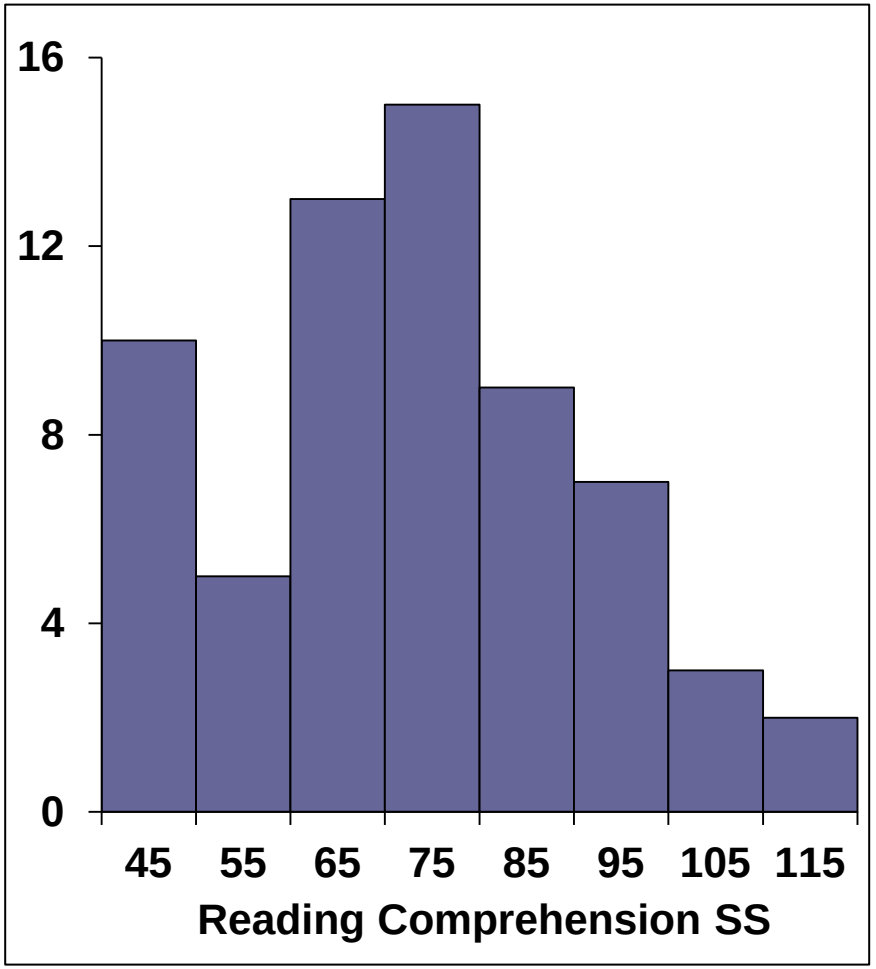
Mean: 60.15, SD: 14.92, Range: 40 – 95

M: 63.61, SD: 15.29, 40 – 100

WIAT-III Reading Standard Scores (Ages 9 – 17 Years, N = 74)



Mean: 75.89, SD: 17.52, Range: 40 – 109



Mean: 70.64, SD: 18.89, Range: 40 – 110

WIAT-III Basic Reading Composite & Reading Comprehension Standard Scores