Intellectual Assessment...

Common Profiles and Testing Tools

Assessments are conducted at regular intervals during a child’s development to qualify the child for services and to better understand his or her current abilities, strengths and weaknesses. Testing also helps us to see how well a child can generalize what he or she has learned. Different tests provide different information, and some tests are better than others for documenting the intellectual strengths and weaknesses of children with WS. Several tests that are commonly used when assessing children with Williams syndrome are described below. The “right” test will vary for students with Williams syndrome 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.

Assessments for Children - Infant through Preschool

In the very early years, the Mullen Scales of Early Learning is a good choice. It is normed for children from 1 month to 5 years, 8 months. It is best used for children aged 54 months or younger.

Advantages for children with WS: It yields scores for 4 separate ability areas so it can show patterns of strength and weakness, rather than averaging everything together. Children with WS often have the most difficulty with visuospatial construction and the Mullen Scales highlight 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 norms do not have a large enough range to show differences for children who score at the ‘floor’ of the test. Note that this problem occurs with most tests for children in this age range. Additionally, 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.

Advantages for children with WS:
The uneven pattern of strengths and weaknesses typical for children with WS can be clearly identified in 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 – 8 years 11 months), Working Memory (recommended for ages 7 years – 17 years 11 months), and Processing Speed (also recommended for ages 7 years – 17 years 11 months).

Conversely, in the more commonly used Wechsler tests (the WISC-IV and the WPPSI-III), the “Performance” scale (or the Perceptual Reasoning Index) lumps together subtests on which children with WS generally perform relatively well (e.g., Matrix Reasoning) and those they have the most difficulty with (e.g., Block Design) and hence their uneven pattern of abilities is not readily discernible at the Index level and overall scores are less meaningful. In addition, the DAS-II has norms for children across a broader range of abilities than the WISC-IV or WPPSI-III. Hence, DAS-II scores obtained by children who are performing further below their same age peers can still be meaningfully interpreted and patterns of relative strength and weakness can be more easily identified.

Determining an IQ score from the DAS-II
The “GCA”, which is like an IQ score, from the DAS-II is meaningful if the 3 core cluster scores do not differ significantly. However, this is the case for only about 10% of children with Williams syndrome. About 50% of children with WS show a clear pattern of higher scores on the Verbal cluster and the Nonverbal Reasoning cluster, with scores on these two quite similar, and substantially lower scores on the Spatial cluster. Only 2% showed
higher scores on the Spatial cluster than the Verbal cluster and the other children show a variety of patterns. When children’s cluster scores are uneven, 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

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

The WPPSI-IV was released in the fall of 2012. This test 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 appears to be more similar to the DAS-II than to the WPPSI-III. For example, the WPPSI-IV has a separate Visual Spatial Index (similar to the DAS-II Spatial cluster) and the standard scores cover a wider range of ability than the WPPSI-III making it more likely that the test will be able to accurately describe the abilities of children with significant developmental delay. As this test was very recently released, no information about the performance of children with WS on it is available.

Some schools or private psychologists may still be administering the WPPSI-III. This test is not recommended for children with WS.

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

The WISC-IV is the most commonly used assessment by school districts and many private psychologists for students ages 6 - 17yrs 11months. It is recommended that if a Wechsler test is to be used for a child with WS, the WPPSI-IV (rather than the WISC-IV) be administered to children with WS aged 6 years – 7 years 7 months. The WISC-IV includes indexes for verbal comprehension (measured entirely by verbal expression), perceptual reasoning, working memory and processing speed.

Disadvantages of WISC-IV for assessing children with WS

The perceptual processing index includes both an area of relative strength (matrix reasoning) and an area of severe weakness (block design). Therefore the WISC Index scores do not give an adequate picture of the relative strengths and weaknesses of children with WS and there often is not a significant difference between performance on the 4 indexes, which leads to the impression that the overall IQ score provides an accurate measure of the child’s intellectual ability.

Additionally, the “floor” for the subtests is only 3 standard deviations from the mean, and therefore not low enough to allow the test to accurately characterize the performance of a large portion of children with WS.
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 formal assessments.

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 either of 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 or Wechsler tests. Additionally, the test 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 generally have 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 have relative strengths in language and nonverbal reasoning, and significant weakness in visuospatial construction. This area of difficulty impacts writing, drawing, and pattern construction.

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 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’s important that goals in the child’s IEP address this area. This difficulty is often most apparent in the social skills area (see above). Another pragmatic difficulty for many children with WS is that they often do not realize that they did not understand what their conversational partner or teacher has said or they may misunderstand what was 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**

**The Beery Test of Visual Motor Integration (VMI)**

The Beery Test measures drawing ability in people ages 2 1/2 to 21 years. It is not recommended for children with Williams syndrome who are younger than 6. The test measures the child’s ability to copy single lines and simple shapes, and to copy more complex forms that require the integration of lines and shapes.

**Drawings by Students with Williams syndrome**

<table>
<thead>
<tr>
<th>5 yr. old</th>
<th>11 yr. old</th>
<th>9 yrs. 7 months</th>
<th>12 yrs. 11 months</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bicycles drawn by the same child with WS</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Advantages:** The Beery is a good test to document abilities and needs in writing related activities (drawing), especially for youngsters with WS, age 6 and older. For success in this area, OT as well as classroom activities and work accommodations are key.

**Disadvantages:** For very young children, the earliest items on this test (e.g. lines and circles) are often worked on in early OT with children and hence their performance on this test may mask real needs in this area.

Fine motor and visual-motor 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, such that their work is not held back to the level of the writing / paper work output difficulties. It is important to keep in mind that paperwork tasks across most subjects involve a great deal of fine motor and visual-motor construction (e.g., writing, drawing). *Children with WS should not spend more time than their peers doing paperwork.* Time can be dedicated for work on fine motor and writing tasks but this skill area difficulty should not 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 as early as possible 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 visual motor 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**

Predicted scores are available based on performance on the DAS-II, the WISC-IV, and the WPPSI-IV. 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. 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 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 comprehension, social skills) than are higher level math skills (e.g. geometry, multiplication and division or 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. Even if a school program is sight
word based, the student with WS should have a phonics based program in their IEP as this 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 include 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 visual-motor integration, 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” within literature as well as in assignments, oral lectures and projects
- Sticking to a task or project from beginning, middle, to end

Children with Williams syndrome 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 their day are key.