

## Williams Syndrome *At a Glance*

Williams syndrome (WS) is a genetic condition characterized by heart, facial and personality differences and often accompanied by mild to moderate intellectual disability. It is caused by missing material, called a microdeletion, on chromosome 7. This missing piece disrupts the gene that helps make elastin. Elastin is a protein that gives elasticity to our tissues and organs. It is found in the walls of arteries, and in the lungs, intestines, and skin. Reduced or abnormal elastin may explain the physical, medical, and facial differences seen in Williams syndrome.

About one in every 7,500 babies is born with Williams syndrome.

*There is wide variability within individuals who have this condition.* Symptoms that may be associated with WS include:

### Heart disease

Heart issues:

- Narrowing of the aorta just above the valve is common.
- Other findings may include:
  - Mitral valve insufficiency
  - Narrowing of the renal artery

### Unique facial appearance

These facial features include:

- Prominent lips with an open mouth
- A long philtrum (distance from the midline of upper lip margin to base of nose)
- Low nasal bridge (top of the nose between eyebrows is flattened) with an upturned tip
- Epicanthal folds (extra fold of skin over inner corner of eyes)
- Partial absence of teeth or defective tooth enamel possible
- Irregular star like pattern of the iris may be present

### Connective tissue problems

These include:

- Joint laxity and limitation
- Soft, lax skin
- Low muscle tone and floppy joints can result in delayed motor milestones.
  - This can lead to compensatory postures to get more stability.
  - With age, may have tighter tone, stronger reflexes, tightening of heel cords and hamstrings.
  - This can result in a stiff and awkward gait and curving of the spine by adolescence.
  - Consider range of motion exercises to prevent or improve joint tightness.
- Inguinal/umbilical hernia
- Bowel/bladder diverticula
- Rectal prolapse

### **Hoarse or low pitched voice**

- Vocal cord changes due to lack of elastin

### **Progressive sensorineural hearing loss**

- Mild to moderate high frequency hearing loss is common in adults

### **Vision/Eyes**

- Farsightedness in 50% of individuals with WS
- Crossed eyes (strabismus) in 50% of individuals with WS

### **Intellectual disability (usually mild) occurs in 75% of individuals**

- Specific cognitive profile: strengths in verbal short term memory and language
- Extreme challenges with visuospatial activities

### **Unique personality characteristics**

- Overfriendly
- Empathetic
- Generalized anxiety
- Attention deficit disorder

### **Growth abnormalities**

- Prenatal growth deficiency
- Failure to thrive in infancy
- Poor weight gain and linear growth in first four years
- Rate of linear growth 75% of normal in childhood

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- Adults usually below 3rd % in height

### Endocrine abnormalities

- Hypercalcemia- too much calcium in the blood
- Hypercalciuria- too much urinary calcium is excreted

## ***Things to Think About***

### 1. Medical / Dietary Needs

#### **What you need to know**

##### Medical

- High calcium levels can be found in individuals with Williams syndrome, usually in infancy. In some individuals this can recur in childhood or even adulthood.
- High calcium levels can also occur in the urine and cause kidney stones.
- Hypothyroidism, or low thyroid hormone levels, occurs at an increased frequency in Williams syndrome.
- Diabetes occurs more frequently in Williams syndrome.
- Ongoing evaluations by a cardiologist may be necessary in some children for monitoring.

##### Diet

- Most children with Williams syndrome do not require any special diet, but may be on a reduced calcium diet.
- A well-balanced diet and exercise is important to reduce the risk of diabetes.
- Gastroesophageal reflux may be present.
- Chronic constipation is common.
- Sensitivity to textures and tastes as well as poor motor control may make mealtimes challenging.

#### **What you can do**

- Talk with the parents about their child's individual medical needs
- Alert the parents about any changes in activity level
- Alert the parents about any changes in bathroom or eating habits
- Screen vision and hearing regularly

## 2. Education Supports

It is important to have **HIGH EXPECTATIONS** for learning for children who have Williams syndrome.

### What you need to know

People who have WS have a unique cognitive profile with specific strengths and challenges. They can be challenging to teach due to good verbal and communication skills. These challenges may mask problems in other areas: understanding language, hyperactivity, difficulty with attention and visuospatial tasks.

Children who have WS can be included in their neighborhood schools. They may do well when extra supports are provided. Being with their peers is good for developing social and communication skills. Their team can make decisions about the need for specific instruction and pace.

Individuals who have Williams syndrome have a cognitive profile that is independent of IQ. Their verbal subtest scores are usually higher than visuospatial construction scores. Intellectual Disability (usually mild) occurs in 75% of individuals.

If the child has vision and/or hearing problems, good classroom seating is important.

### General Considerations

Strengths:

- Concrete language
- Concrete nonverbal reasoning
- Verbal short term memory
- Auditory rote memory
- Social interactions/interpersonal skills
- Expressive vocal language
- Short and long term auditory memory

Challenges:

- Concentration
- Distractible
- Impulsive
- Visuospatial development
- Visual memory
- Abstract reasoning
- Perseveration

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- Reading nonverbal cues

## Communication

Studies show that speech development is typical but delayed.

- Individuals have good spoken language but poor comprehension.
- They may have trouble learning grammar (plurals, past tense, etc.). Once they learn the concepts they can quickly catch up to peers.
- Verbal expression is better than understanding of speech.
  - May recite instructions or stories word for word but may have trouble with basic concepts
  - May echo or repeat with little understanding of what's being said
  - Answering a question oddly may mean they do not understand the conversation.
  - May use long words or expressions which may not fit the context. This occurs when talking about topics of interest.
  - May be difficult to take turns and maintain a conversation
  - May chatter quite a bit, often at a superficial level
  - May have conversations with adult-like style, and great vocabulary, phrases, and clichés
  - May initiate conversation well
  - Can maintain conversation flow

## Visuospatial, perceptual, and motor challenges

- Individuals may have visuospatial problems. They may also have gross and fine motor coordination issues.
- Difficult visual processing tasks include: discrimination, sequencing, and visual memory.
- Visuospatial problems may lead to the following:
  - Slower in learning to sit and walk
  - Poor posture
  - Limitations of joint movements
  - Fine motor challenges leading to difficulty with tool writing and handwriting at all ages
  - Difficulty sorting, matching objects/shapes, tracing over lines
- They may have difficulty with:
  - Riding a bike
  - Buttoning buttons
  - Cutting with a scissors
  - Holding a pencil
- Motor and perceptual problems may include challenges with:
  - Seeing things in, or as if in, 3 dimensions
  - Eye hand coordination

- Orienting body and objects in space
- Judging distances and directions. This may result in:
  - Fear of heights and climbing
  - Hard time throwing
  - Difficulty on stairs and uneven surfaces (grass, gravel, sand)
  - Difficulty with jigsaw puzzles
  - Difficulty copying a “t” or “x” with blocks

## Reading

Most people with WS learn to read at a basic or, in some cases, more advanced levels.

With reading and literacy, individuals who have WS generally have:

- Good verbal skills
- Good memory for sounds and words
- Good auditory sequencing skills
- Good short and long term auditory memory
- Reading skills that match cognitive ability rather than language-related skills
- Difficulty generalizing rules to new materials
- Strength with concrete vocabulary
- Difficulty with relational and conceptual vocabulary

## Writing and spelling

Writing and spelling tend to be more difficult than reading. This is due to the visual and fine motor skill required for these tasks.

With writing and spelling, people who have WS may have:

- Problems with forming letters, getting distracted (“spacing out”), and aligning words
  - May have difficulty keeping his/her place or skip sections
  - Progress may be slow
  - Need for practice and repetition
- Fine motor delays make it challenging to hold a pencil/pen
  - Be sure to address seating, posture, hand position, pencil grip
  - Teach keyboarding and word processing skills

## Math

Math can be challenging for individuals with WS.

- Perceptual, visuospatial, and motor problems may make math and comprehension hard

## Music

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Individuals who have WS may have musical interests.

- Many individuals with WS have an affinity for music.
- They are often very moved by music.
- Many have absolute and relative pitch.

## What you can do

### INTERVENTIONS to Support Academic Differences:

Each child should be looked at individually to find ideas that work for them. You may want to consider the following strategies and decide if any of these suggestions will work for the child.

The Williams Syndrome Association ([www.williams-syndrome.org](http://www.williams-syndrome.org)) features educational strategies, information for teachers, and testing and evaluation strategies.

### Help with communication

- Direct instruction
- Encourage oral expression so they become an active part of instruction
- Simplify information
- Use pictures and hands-on activities
- Use verbal strengths to assist in learning spatial tasks
- To help with vocabulary development:
  - Make concrete associations for unknown words
  - Encourage students to say and re-state their understanding
  - Teach multiple meanings of words
  - Build semantic webs
  - Teach rules and patterns
- Use verbal skills to start and maintain social contact.
  - They may exaggerate and be dramatic.
  - Reciprocal conversation may be hard
- Speech therapy is important to promote early development of speech and language.
- Even though they may speak clearly, difficulty with conceptual and relational language, and pragmatics can occur. They may also have difficulty with grammatical comprehension, gender assessment, forming patterns of words and sentences, and oral fluency.

### Help with visuospatial development

- Extra help and practice
- Reassurance

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- Occupational therapy
- Minimize tracing and pencil work
- If writing name is hard, use a stamp
- Use of computer

## Help with reading

Success in **reading comprehension** requires comprehending words and grammatical structures in the text.

- Can address these issues in speech and reading therapy
- Need help in comprehension strategies, and in summarization, question generating, graphic organization
- Teach reading comprehension skills directly
- Self question techniques to monitor comprehension

When teaching reading, rely on **auditory skills and phonetic approach**.

- Phonics should be integrated into reading and writing.
- Learn phonics rules by reading and writing vs. memorizing
- Teach a few skills at a time
- Use mnemonic devices

Think about how to **present information**.

- Can be easily distracted by extraneous visual stimuli.
- Use books with simple pictures or outline drawings, which are less likely to distract.
- Lots of color and pictures may be too over stimulating.
- Little information on each page
- Need explicit instruction and extensive practice
- Introduce materials related to interests

Encourage verbalization.

Teach organization and structure of paragraphs.

Teach signal words indicating transition.

## Help with writing and spelling

- Relate work to interests
  - Trace cars, trains, or anything of high interest
- Use computers
- Dictate stories and/or homework

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- Brief daily practice to improve handwriting rate
- Teach use of verbal self directions
- Focus on only one aspect of writing at a time
- Teach traditional words
- Teach patterns for writing paragraphs
- Provide purpose and structure
- Trace over and copy letters
- Address seating, posture, hand position, pencil grip
- Teach keyboarding and word processing skills

### Help with math

- Work at their pace
- Use repetition
- Use a digital watch for telling time
- Use real coins
  - Take child to cafeteria or book sale to buy items
- Use manipulatives
- Use concrete examples
- Separate the writing of numbers from teaching of concepts
- Use lined paper or squared paper to help align numbers in addition
- Use music to teach math (i.e. when counting)

## 3. Behavior & Sensory Support

### What you need to know

Children who have Williams syndrome tend to have unique personalities. They may be overfriendly, empathetic, or have generalized anxiety, or anticipation anxiety vs. social anxiety. They may worry and have anxiety about themselves and others. This may lead to problems with toileting, eating, and sleeping.

School age children who have WS are often outgoing, sociable, and articulate. Many love talking with adults and can be very helpful. Being overly friendly to adults may make it hard to make and keep friends.

Children who have WS are usually cooperative and eager to please. They can be highly sensitive to emotions of others and may cry tears of empathy. They may be sensitive to mood changes in adults and sensitive to criticism. They may be very sensitive to their own feelings of frustration and have temper outbursts. They may have trouble interpreting words or facial expressions.

### Some of the personality and behavioral issues may include:

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## Attention deficit hyperactivity disorder

- 50-90% of adolescents and adults meet Diagnostic and Statistical Manual of Mental Disorders, fourth edition (known as DSM-4) criteria for anxiety disorder, phobic disorder, Attention Deficit Hyperactivity Disorder (ADHD), or a combination.
- 65% of adolescents meet criteria for ADHD and about 1/2 of that group meet criteria for specific phobia.
- Can be challenging to sit still and concentrate on tasks for length of time

## Sensory defensiveness

### Auditory system challenges

- Very sensitive to sounds such as loud bangs, clapping, and laughter
- May become tense and fearful when hearing or anticipating sounds

### Tactile system challenges

- May be sensitive to different textures
- Sensitivity can include textures in foods and may cause gagging.

## Perseveration

- Preoccupation and fascination with objects and topics
- Unusual or restricted interests

## Sleep difficulties

Individuals with Williams syndrome often have significant sleep disturbance, including:

- Sleep apnea
- Bedtime resistance
- Increased sleep latency
- Frequent night walking

## Specific phobias

Phobias are common, especially those involving loud or sudden noises.

Other phobias may include:

- Thunderstorms
- High places

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- Animals
- Escalators

## What you can do

### Supports for children who have WS in dealing with their anxiety and social challenges

- Occupational therapy, cognitive, and behavioral approaches can address anxiety.
- Individuals with strong verbal skills may benefit from counseling (relaxation, rehearsal)
  - Medication may be considered

### Supporting children who have behavioral challenges

- Use Positive Behavioral Interventions and Supports including Applied Behavior Analysis (ABA), functional behavioral assessments, and person-centered planning.

### Working on social skills development

Improve conversation skills

- How to make appropriate physical contact
- How and when to interrupt

Practice cooperative play skills

- Taking turns

Support development of friendships

- Inclusion in typical classrooms
- Respecting personal boundaries
- Peer supports
- Small group work to help support friendships
- Social coaching from adults

Support development of emotional skills

- Recognize feelings of self and others
- Recognize nonverbal cues

Teach safety with strangers

### Improving adaptive behavior

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Strengths in tasks with verbal skills

- Social interaction
- Communication

#### Challenges in skills depending on visual motor or spatial skills

- Self help
- Community living skills

## 4. Physical Activity, Trips, Events

### What you need to know

- Occasionally, there can be significant narrowing in the major blood vessels around the heart that can lead to hypertension or an enlarged heart.
- Any change in routine may produce anxiety, fears and/or worry.
- Children with Williams syndrome are often overly friendly and may be too familiar with strangers.
- Problems with depth perception may cause the child to be fearful of stairs or high places.
- Low muscle tone may continue into school age and may cause easy fatigue.
- Children often have difficulty balancing on one foot or with a typical alternating gait.

### What you can do

- Consider any medical factors, such as heart issues, and discuss adaptations needed with parents
- Offer anticipatory guidance and preparation
  - Create a picture story about the upcoming event. The child can rehearse it alone or with others.
  - Crowds and loud noises can be overwhelming to children who have WS. Try:
    - Earplugs or headphone
    - IPod with headphone
  - Consider a one-on-one aide
  - Assign an adult or student for them to be “partner” with during trip or event
- Arrange for preferred seating for any hearing or vision issues
- Work with parents to develop an adaptive physical education program for balance and fatigue issues, if present

- Be aware of new situations that might cause increased anxiety in the child – such as preparing them for fire alarms, etc.

## 5. School Absences and Fatigue

### What you need to know

- Most children with Williams syndrome will not have increased absences due to medical issues, but this possibility should be discussed with the parents.
- Sleep problems are common and may lead to fatigue during the day.
- Low muscle tone and problems with balance and body position sense may lead to increased fatigue.

### What you can do

- Ask the parents about medical issues that might affect the child's attendance
- Alert the parents of changes in the child's energy level
- Use a communication book between school and home to keep track and learn about issues at home that may affect fatigue

## 6. Emergency Planning

### What you need to know

Emergency plan should be on an as needed basis, based on each individual child. Know if the child has any medical issues (i.e., cardiovascular).

## 7. Resources

### Williams Syndrome Association newsletter focusing on education

<https://issuu.com/williamssyndrome/docs/educationissue2013> 2013 Heart to Heart Special Issue

### Williams Syndrome Association (including education strategies)

<http://www.williams-syndrome.org> See their educational strategies for teachers.

*This is a resource for: EDUCATORS; parents; grandparents; individuals; doctors; researchers; volunteers.*

### GeneReviews

<http://www.ncbi.nlm.nih.gov/books/NBK1249/>

*Learn more about the genetics of Williams syndrome.*

### American Academy of Pediatrics (AAP) – Health Supervision Guidelines

[https://williams-syndrome.org/sites/williams-syndrome.org/files/Child\\_Health\\_Care\\_Supervision\\_0.pdf](https://williams-syndrome.org/sites/williams-syndrome.org/files/Child_Health_Care_Supervision_0.pdf)

The AAP endorses these Health Supervision Guidelines. Families may find these helpful when talking to their pediatricians or family physicians.

### Classroom Accommodations for Students with Visual Issues

Classroom Accommodations for Students with Visual Issues is a resource from Boulder Valley Vision Therapy, P.C. (<https://www.bouldervt.com/>), in Boulder, Colorado. It describes a variety of variety of issues and offers strategies. <https://www.bouldervt.com/wp-content/uploads/sites/478/2015/12/227-Course-Handout-CCIRA-2016.pdf>

### Genetics Home Reference

<https://ghr.nlm.nih.gov/condition/williams-syndrome>

Consumer-friendly information about human genetics from the U.S. National Library of Medicine

**Note:** *This printable version does not include the information found under the green button marked “Transitions” on the website. Those general pages may be printed separately.*

## Meet a Child with Williams Syndrome – *Musical Zhala*



At age 7, Zhala (pronounced Jah-la) can hear a song once or twice and then sing it, even in another language. “She is a 100% musical child,” says her mother Vanessa. At family gatherings where many family members play guitars, Zhala will try to play their instruments. She has listened to her Kurdish grandfather’s music on YouTube which is sung in his native language and she can sing it well after listening only twice!

Zhala is an extremely friendly child and usually quite happy. She is much loved at school and is surely missed if she misses a day of school. In fact, her teacher described her as the “glue of the classroom” because she knows the name of every child, calls them by name, and greets each one.

Her school experience has been phenomenal. She is in typical classes and her therapists come into her class to do therapy. “Math is her strongest subject, which is contrary to what children with Williams syndrome typically experience,” says Vanessa. Reading comprehension is a top priority, as is strengthening her social skills so that she has good solid friendships. “She is not isolated in any way,” says Vanessa who is very pleased at how her school team has supported her in the classroom.

Vanessa tells how the team has earned her trust. When Zhala’s eating didn’t go well on the first day of school, her case manager called home that very afternoon after receiving an email from Vanessa. They developed a communication notebook and started using it the very next day, and continued all the way through the end of the school year.

Her team also problem solved how to get Zhala into the gymnasium, which was so noisy that she couldn’t step foot inside. They slowly desensitized her by having her watch through the window, then go inside for 5 minutes, etc., progressing until Zhala could go in for full gym classes.

Zhala’s parents became concerned with her development when she was about 6-8 months of age. Her pediatrician suggested waiting for testing until she was 12 months old. At that time, Zhala was referred to Early Supports and Services in New Hampshire (early intervention). Around that time, she saw a developmental pediatrician who saw developmental delays. After the visit with the developmentalist, the family took some time to adjust to a possible disorder. Within a month or two, the family decided to move forward to see a neurologist and geneticist. During her first year, from 9-12 months of age, she failed to gain weight and they found out she

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had a gastro-intestinal infection. She was then treated by her GI specialist. GI issues continue for Zhala, but she is highly monitored for feeding/swallowing issues as well as to assure weight gain and other GI concerns.

Zhala was diagnosed with Williams Syndrome at 22 months. But like all children, Zhala has brought much light and joy into her family's life, and she can make just about anybody smile. Zhala needs to see several specialists every few months for monitoring of conditions which are common to Williams. However, Zhala is doing well, and at every visit, the staff knows her by name and greets her with gloves that she asks for every time. At this time, Zhala is in good health, and her various health issues are within normal levels.

Vanessa, who is an educator, shared some great tips for Parents, Teachers, Nurses and Therapists:

- An invaluable tool is a daily communication log which highlights any issues or news in her school or home routines such as eating, behavior, or toileting.
- Seeing her teachers daily is a wonderful luxury because she drives Zhala to school and picks her up. If that is not possible, she suggests emailing or writing frequently.
- Go into school with a good game plan, be open about significant concerns, and make sure concerns are addressed in a reasonable amount of time.
  - Vanessa brought in information from the GEMSS website as a place to start the discussion
  - Then she told them about features and issues that were unique to Zhala and listed their priorities for her education.
  - An example: the school was not completely fenced in so they discussed how Zhala, being so friendly, might walk off with anyone.
- Discuss issues that could be big problems, like sensitivity to sound. For example, they had to have a plan for fire drills because Zhala is so highly sensitive to noises that a fire drill would scare her tremendously.
- If something doesn't feel right, be an advocate and don't worry about being a complainer.
- Music can be so beneficial to help with cognitive skills. For example, Zhala's teacher has had great success teaching math facts by singing them with Zhala. "Music is a great motivator and cognitive tool for lots of kids," says Vanessa who has done a lot of reading on the topic.
- Music therapy can also be a great addition to the IEP and help students make greater achievements in their education.