

Williams Syndrome

Background

Williams syndrome (WS) is a genetic developmental disorder characterized by mild to moderate intellectual disability, distinct facial features, and unique social characteristics. An array of medical problems range in severity and age of onset, but the majority affect the cardiovascular and gastrointestinal systems. Low muscle tone and connective tissue abnormalities are also common.

Children with WS have distinct facial features including a small, upturned nose, wide mouth, prominent lips, small chin, and puffiness around the eyes. Teeth are often small and widely spaced. These features tend to become more noticeable with age. Other characteristics or complications include:

- Developmental delay
- Heart and blood vessel problems (e.g., cardiovascular disease, hypertension)
- Bowel motility changes, reflux, stomach pain
- Low muscle tone
- Dysphagia
- Joint laxity, unsteady gait
- Bone malformations
- Scoliosis
- Hypothyroidism
- Early puberty
- Hearing sensitivity
- Hernias
- Chronic ear infections, risk for hearing loss
- Sleep problems

Delays in language development are common at a young age, but strong verbal abilities are typically a strength as the child matures. A strong relationship exists between individuals with WS and music. Their affinity for music contributes to their auditory strengths.

Children with WS are excessively empathetic and outgoing.

Their personalities are pleasant and sociable by nature.

Children with WS tend to engage more easily with adults than peers their age. They can also experience anxiety or specific phobias contributing to outbursts of aggression or challenging behavior.



Top Takeaways for School

WS is a rare genetic disorder characterized by distinctive facial features, cardiovascular abnormalities, and unique cognitive and behavioral traits, including strong social personality and learning difficulties.

Expressive language and social skills may be a relative strength.

A student with WS may be very social and engage more easily with adults versus peers. Developing a safety support system should be considered.

Low muscle tone and poor visuo-spatial awareness can make tasks like handwriting difficult.

Affinity for music can be useful to help engage children with WS in the classroom.

Individuals with WS may lack the understanding to sense what others may be feeling or thinking. Use of play scenarios and social stories may help foster social skills and friendships with peers.

Considerations for the Individualized Healthcare Plan (IHP)

- Nursing diagnoses: Risk for unstable blood pressure, risk for injury and impaired thought process
- Nutrition interventions and equipment
- Student-specific triggers, avoidance, or intervention strategies
- Use of specialized equipment, adaptive equipment, orthotics
- Activity and positioning precautions and/or restrictions
- Consider emergency action plans (EAPs) and emergency evacuation plans (EEPs) related to special health care needs, including staff education/training

Discussion Starters for the Educational Team

1. Would the student benefit from evaluations or assessments in any of the following areas: physical therapy, occupational therapy, speech and language therapy, assistive technology, adapted physical education, functional behavior, psychology, hearing and vision?
2. Would the student benefit from additional academic support and/or modified education (e.g., copies of notes, extra time, reduced workload, simplified instructions, alternative formats for presentation of material, 504/IEP)?
3. Does the student need additional adult support to access the academic curriculum in the least restrictive environment?
4. Can rest breaks, safe spaces or reduced stimulation times be built into the student's schedule?
5. Will staff receive education/training to implement the student-specific emergency plan?

Resources

Kennedy Krieger Institute: Neurology and Neurogenetics Clinics
[kennedykrieger.org/patient-care/centers-and-programs/neurology-and-neurogenetics-clinics](https://www.kennedykrieger.org/patient-care/centers-and-programs/neurology-and-neurogenetics-clinics)

Williams Syndrome Association
[williams-syndrome.org/](https://www.williams-syndrome.org/)

Williams Syndrome Foundation
[williams-syndrome.org.uk/](https://www.williams-syndrome.org.uk/)



For more information, please scan the QR code or visit: [KennedyKrieger.org/SHNIC](https://www.KennedyKrieger.org/SHNIC)

