WILLIAMS SYNDROME

What is it?

Williams syndrome (WS) is a random genetic mutation disorder present at birth affecting both boys and girls equally. Deletion of genetic material from a specific region on chromosome 7 causes WS. Researchers believe deletion of genetic material from a specific region of chromosome 7 contribute to the characteristic features of WS. This disease is characterized by an array of medical problems that can range in severity and age of onset. Most cases are characterized by unique facial features, cardiovascular disease, and connective tissue and endocrine abnormalities. Developmental delay occurs in conjunction with striking verbal abilities, highly social personalities, and an affinity for music.

What are the characteristics or complications?

Children with WS have unique facial characteristics which include small upturned nose, wide mouth, prominent lips, small chin, and puffiness around the eyes. Individuals with WS also possess unique personality traits including excessive friendliness and lack of stranger awareness. They can be excessively empathetic, anxious, or experience specific phobias. Other characteristics or complications include:

- Heart and blood vessel problems
- Low muscle tone, joint laxity, unsteady gait, scoliosis
- Feeding difficulties in infancy, reflux, poor growth
- Dental abnormalities
- Elevated calcium levels, hypothyroidism, early puberty, diabetes in adulthood
- Developmental delay
- Speech delay in early childhood
- Hearing sensitivity
- Kidney problems
- Hernias
- Chronic ear infections, risk for hearing loss
- Constipation
- Sleep problems

What is the treatment?

Although there is no cure for WS it is important to identify and treat the different medical problems that can occur with this disorder. Treatments and interventions must be based on the unique needs of each individual. This could mean regular monitoring for potential medical problems like cardiac conditions or elevated blood calcium levels. This could also include specialized services and therapies to treat things like low muscle tone and speech delays in order to maximize the individual's potential.

Kennedy Krieger Institute

The **Specialized Health Needs Interagency Collaboration (SHNIC)** program is a collaborative partnership between the Kennedy Krieger Institute and the Maryland State Department of Education.

Suggested school accommodations

Most students with WS have some form of learning difficulties but they can significantly vary. Many students with WS appear scattered in their level of abilities across domains. As they age, you may notice the student struggling with concepts like spatial relations, numbers and abstract reasoning. Although a student with WS may be very social, remember to monitor their support systems and social interactions as they often have a difficult time understanding social cues. Supporting students with these conditions in the school require educators and parents/guardian to work as a team. Some accommodations to consider for a 504/IEP could include:

- PT/OT/SLP evaluations
- Vision services and hearing specialists
- Behavioral support
- Adapted PE
- Note anxiety and fears like noise of school bells and alarms in the school setting
- Provide visual and picture tools for learning
- Incorporate music wherever possible
- Offer direct, step-by-step instruction
- Provide frequent breaks
- Provide visual schedule to prevent perseveration on upcoming events
- Encourage safe relationships, develop social skills
- Staff education/training as appropriate
- Emergency Evacuation Plan (EEP)

Specific health issues for Individualized Healthcare Plan

- Diagnosis including all medical characteristics
- Baseline cardiac assessment including heart rate monitoring orders with reportable parameters
- Blood pressure monitoring orders with reportable parameters
- Note if student wears a Medic Alert bracelet for cardiovascular emergencies
- Orders for weight checks, if applicable
- Orders for temperature regulation
- Fever protocol including PRN medications
- Nutrition orders including feeding protocol, positioning for feeds
- Baseline skin assessment, including use of orthotics if applicable
- Orders for physical activity/ positioning restrictions
- Communicate with school staff, parents, and provider any changes or concerns about the disease
- Emergency Care Plan(s) (ECP) related to medical needs in the school setting and staff education/training as appropriate for each

Resources & Manuals

Williams Syndrome Association
www.williams-syndrome.org

Williams Syndrome Foundation
www.williams-syndrome.org.uk

Genetic and Rare Disease Information Center (GARD)
https://rarediseases.info.nih.gov/diseases/7891/williams-syndrome

Cleveland Clinic: Williams Syndrome
https://my.clevelandclinic.org/health/diseases/15174-williams-syndrome