WOLF HIRSCHHORN SYNDROME

What is it?

Wolf-Hirschhorn syndrome (WHS) is a genetic syndrome caused by deleted or missing genetic material on the 4th chromosome. It is also known as 4P syndrome. Deletion normally occurs at random, varying by person. The size of the deletion predicts the amount and severity of disease characteristics. WHS is characterized by distinct facial features called "Greek helmet facies," developmental delay, growth delay and seizures. Seizures affect almost all individuals with WHS and are one of the most serious health issues for these individuals. Seizures typically begin at about 5 months of age but will stop between ages 3-11 in about half of the cases. Delayed growth begins at birth and children continue to struggle with feeding and gaining weight. Poor nutritional intake leads to generalized weakness. About 1 in 50,000 children are affected with females twice as likely as males to be diagnosed.

What are the characteristics or complications?

The most distinctive feature of WHS is the typical asymmetrical facial appearance including a widening and prominence of the area located at the top of the nose between the eyebrows. This is associated with prominent and wide-set eyes, arched eyebrows, a small chin and jaw, and a short distance between nose and upper lip. While intellectual disability ranges from mild to severe in individuals with WHS, their socialization skills are strong. Verbal communication and language tend to be weaker. Other additional features of WHS could include:

- Microcephaly
- Short statue
- Delayed growth and development
- Poor muscle tone, scoliosis
- Seizures
- Renal issues
- Hearing/vision/speech impairment
- Skin changes like mottling or dry skin
- Dental problems, missing teeth,
- Cleft palate and/or lip
- Rocking, head shaking
- Hand flapping

What is the treatment?

Although there is no cure for WHS, it is important to identify and treat the different medical problems that can occur with this disorder. A team of medical and developmental specialists are key to the best possible outcome. Those diagnosed with WHS will often be followed by numerous specialists which might include cardiologists, neurologists, nephrologists, gastroenterologists and clinical geneticists. The pediatrician will often manage the multidisciplinary approach. Some treatments might include: feeding therapy, anti-seizure medications, PT/OT/SLP therapists to strengthen mobility and skeletal abnormalities.

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

Supporting students with WH in the school setting require the educators and parents/guardians to work as a team. A well coordinated plan promotes success for the student in the educational setting. Some accommodations to consider for an 504/IEP could include:

- PT/OT/SLP evaluations
- Vision/Hearing consultations
- Behavioral supports
- Plans for mobility accommodations and core strengthening
- Adapted PE to accommodate motor development
- Assistive technology
- Extra adult support
- Offer preferential seating
- Organizational strategies and aids
- Presenting materials in various formats for the student
- Offer designated rest area
- Emotional support
- Staff education/training as appropriate
- Emergency Evacuation Plan (EEP)

Specific health issues for Individualized Healthcare Plan

- Diagnosis including all affected systems and symptoms
- Current medication list for school and home
- Nutrition orders, including feeding protocol
- Orders for GT/JT feeds, including flush and replacement per county policy, if applicable
- Communicate with school staff, parents, and provider any changes or concerns about the disease
- Emergency Care Plan (ECP) related to seizures and other medical needs in the school setting and staff education/training as appropriate for each

Resources & Manuals

NIH Genetics Home Reference
https://ghr.nlm.nih.gov/condition/wolf-hirschhorn-syndrome

National Organization for Rare Disorders
https://rarediseases.org/rare-diseases/wolf-hirschhorn-syndrome/

The Real Story About Wolf-Hirschhorn Syndrome
http://wolfhirschhorn.org/