

What is it?

Wolf-Hirschhorn is a genetic syndrome caused by deleted or missing genetic material on the 4th chromosome (also known as 4P syndrome). Deletion normally occurs at random. The size of the deletion predicts the amount and severity of disease characteristics. The amount of the deletion, or missing material, varies by person.

About 1 in 50,000 children are affected; with females twice as likely as males to be diagnosed. Delayed growth begins at birth and children continue to struggle with feeding and gaining weight. Poor nutritional intake leads to generalized weakness and children will often struggle to stand, walk and even sit.

WH is characterized by distinct facial features called “Greek helmet facies,” developmental delay, growth delay and seizures. Seizures affect 90% of cases and are one of the most serious health issues for these individuals. According to *Wolfhirschhorn.org*, seizures begin about 5 months of age but 50% of those affected stop having seizures between ages 3-11 years (2015).

Physical Characteristics

- Broad, flat nasal bridge
- High forehead
- Wide-set eyes
- Asymmetrical facial features
- Droopy eyelids
- Small chin
- Cleft lip/palate
- Poorly formed ears
- Short distance between nose and upper lip
- Hypotonic
- Microcephalic
- Short stature
- Abnormal spinal curvature
- Dry, mottled skin



Common Medical Concerns

- Seizures
- Heart defects
- Feeding difficulties
- Hearing/Vision impairment
- Dental problems
- Renal issues

Behavior/Developmental Concerns

- Delayed/weak motor skills
- Developmental delay
- Mental retardation
- Rocking, head shaking
- Hand flapping
- Speech impairment

Is there treatment?

There is no cure or treatment for WH. Treatments aim to alleviate symptoms specific to each individual. Recommendations to help manage symptoms include:

- Feeding therapy
- Tube feedings
- Anti-seizure medications
- Screenings for hearing loss and vision problems
- PT/OT services for mobility and skeletal abnormalities
- Speech and language therapist



Suggested school accommodations

- Considerations for hearing and vision specialist or consult
- OT/PT/Speech consult
- Plans for mobility accommodations and core strengthening
- Adaptive PE considerations
- Education about seizure management for staff
- Presenting materials in various formats for the student
- Alternative communication devices
- IEP

SHNIC school nurses information:

Specific health issues for individual health care plans

- Diagnosis information including age, symptoms
- Current medication list
- Nutrition orders, including feeding protocol
- Orders for GT/JT feeds, including replacement of tube, if applicable
- Orders for emergency medications, when to administer, dose, route
- Note if seizures are controlled
- Seizure action plan, when to call 911
- Staff education about seizures and safety
- Rest breaks or periods
- Aid or supervision

Resources & Manuals

Wolf Hirschhorn Syndrome

<http://ghr.nlm.nih.gov/condition/wolf-hirschhorn-syndrome>

National Organization for Rare Disorders

<https://rarediseases.org/rare-diseases/wolf-hirschhorn-syndrome/>

The Real Story Behind Wolf-Hirschhorn Syndrome

<http://wolfhirschhorn.org/about-wolf-hirschhorn-syndrome/>