WOLF-HIRSCHHORN SYNDROME

Background

Wolf-Hirschhorn syndrome (WHS) is a rare genetic condition caused by deleted or missing genetic material on the 4th chromosome, also known as 4P syndrome. WHS is characterized by distinct facial features, delayed growth and development, and seizures.

Distinct facial features called "Greek helmet facies" include a small head size (microcephaly), prominent eyes spaced widely apart, a downturned mouth, small chin and jaw, and a short distance between nose and upper lip. Cleft lip and palate are also common.

Delayed growth and development begin at birth. Children struggle with feeding and gaining weight and can be diagnosed as failure to thrive. Poor nutritional intake leads to generalized weakness and poor muscle tone. Children struggle to meet early developmental milestones like sitting and walking.

Approximately 90% of children with WHS will be diagnosed with seizures at an early age. Seizures are most often triggered by fever and tend to lessen or go away as the child ages. Intellectual disability is generally moderate to severe. Verbal communication and language tend to be weak but social sills strong. Speech is often limited to sounds although some individuals will achieve use of simple sentences.

Other body systems are affected by WHS and anomalies can include:

- Congenital heart defects
- Vision problems
- Hearing impairment
- Dental anomalies
- Recurrent respiratory tract infections
- Frequent ear infections
- Urinary tract malformations
- Scoliosis

There is no cure for WHS. Treatment is focused on managing specific health issues and the related symptoms which may include feeding therapy, anti-seizure medications, and surgery or therapy to correct or strengthen skeletal abnormalities and improve mobility.

Top Takeaways for School Considerations

WHS is characterized unique facial features, delayed growth and development, intellectual disability, and seizures.

Intellectual disability and language disorders are common, with a difficulty specifically in the expressive type. Communication may improve over time.

Children with WHS often have strong social skills with engaging personalities.

Side effects of anti-seizure medication can cause fatigue, inattention, and restlessness that further interrupt school performance.

Considerations for the Individualized Healthcare Plan (IHP)

- Nursing diagnosis of social isolation, impaired physical mobility, impaired swallowing, low situational self-esteem, and ineffective breathing patterns
- Current diagnosed health condition including date of diagnosis, progress of disease process and other chronic health conditions
- Current medication and treatment orders (consider schedule, equipment needs and side effects)
- Allergies or food restrictions
- Nutrition interventions and equipment needs
- Student-specific triggers, avoidance, or intervention strategies
- Use of specialized equipment, adaptive equipment, orthotics
- Consider emergency care plan(s) (ECP) and emergency evacuation plan(s) (EEP) as related to medical needs in the school setting, and staff education/training, as appropriate

Discussion Starters for Educational Team

1. Has the school staff been trained to implement the student-specific emergency plan?
2. 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?
3. 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)?
4. Does the student need additional adult support to access the academic curriculum in the least restrictive environment?
5. Is the physical school environment safely accessible for the student's mobility needs (e.g., entry and exit, ramps, location of classes, access to elevator, doorways)?
6. Does the classroom environment support the student's needs and/or equipment (e.g., desk/seating options, maneuverability space, electrical outlets, flash pass for bathroom or nurse)?

Resources

Kennedy Krieger Institute: Neurology and Neurogenetics Clinics
kennedykrieger.org

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

Scan QR code or visit **KennedyKrieger.org/Redirect** for more information.

Kennedy Krieger Institute