DANDY-WALKER SYNDROME

What is it?

Dandy-Walker (DW) Syndrome is a rare congenital birth defect of the brain. DW involves the cerebellum, the back part of the brain, and the fluid filled spaces that surround it. The cerebellum controls movement, behavior, and cognition. Many people experience difficulties with balance, coordination, voluntary muscle movements, mood and intellectual disability. DW is often associated with other disorders of the central nervous system including absence of the corpus callosum and malformation of the heart, face, limbs, fingers, and toes. DW can disrupt the normal flow of cerebrospinal fluid (CSF) leading to a condition called hydrocephalus. A DW diagnosis may be more common in females.

Although a small percentage of cases occur in a family, most diagnoses occur sporadically with no familial pattern of inheritance. In most cases, the cause is a combination of genetic and environmental factors that could include viral infection or intrauterine exposure to certain toxins or medications. A diabetic mother is also more likely to have a child with DW.

Prognosis largely depends on severity of DW and its associated congenital defects. Malformations causing key features associated with DW include:

- Enlargement of the 4th ventricle, the area that allows fluid to flow between the brain and spinal cord
- Absence or partial development of the cerebellar vermis, the area between the 2 hemispheres
- Development of a cyst at the base of the skull
- Increase in size of fluid filled spaces of brain

What are common effects?

Some symptoms develop slowly and seemingly unnoticeable over time while others appear dramatically. Symptoms often beginning in early infancy include slow motor development and progressive skull enlargement. Children could demonstrate delays in sitting, walking, and talking while intellectual development varies. Diagnosis is normally made around 3-4 years of age. Other advancing symptoms include:

- Malformation of heart, face, limbs, fingers and toes
- Affected nerves of eyes, face, neck
- Jerky, uncoordinated movements, stiffness
- Vomiting, irritability
- Seizures
- Increased head circumference, increased intracranial pressure
- Bulging at the back of the skull
- Abnormal breathing related to cerebellar dysfunction

What is the treatment?

There is no cure for DW and treatment focuses on relieving pressure on the brain and managing the associated symptoms. A ventriculoperitoneal shunt will be surgically placed to drain excess fluid and reduce swelling and intracranial pressure. Medications are often prescribed to control seizures.

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

The effect of Dandy-Walker syndrome on cognitive development is variable. About half of those diagnosed will have intellectual disability. Early intervention is key. Supporting students with this condition in the school may require educators and parents/guardian to work as a team. Some accommodations to consider for a 504/IEP could include:

- PT/OT/SLP evaluation
- Monitor visual, hearing, speech impairment
- Monitor coordination/motor skills
- Monitor fine/gross motor skills
- Assess sensory issues to noise, touch, light
- Monitor for fatigue
- Educate on mobility devices, orthotic
- Extended processing time
- Offer rest breaks as appropriate
- Offer repetition
- Use of visual tools to aid with vision
- Student location in classroom/preferential seating
- Tools to aid in memory
- Use of assistive technology
- Offer clear, concise direction
- Staff education/training as appropriate
- Emergency Evacuation Plan (EEP)

Specific health issues for Individualized Healthcare Plan

- Complete diagnosis documentation
- Current medication orders for school and home
- Orders and documentation for hidden device like shunt, VNS
- Child specific signs and symptoms of increased intracranial pressure
- Order for types of seizures
- Child specific characteristics of seizures
- Nutrition orders, including feeding tube replacement per county policy
- Fever protocol
- Activity and positioning restrictions
- Orders for orthotics or assistive devices
- Communicate with school staff, parents/guardian, and provider any changes or concerns about the disease
- Emergency Care Plan(s) (ECP) related to medical needs in the school setting and staff eduction/training as appropriate for each

Resources & Manuals

Dandy-Walker Alliance
http://dandy-walker.org/

National Institute of Neurological Disorders and Stroke
https://www.ninds.nih.gov/Disorders/All-Disorders/Dandy-Walker-Syndrome-Information-Page

Dandy Walker Syndrome
https://www.brainfacts.org/