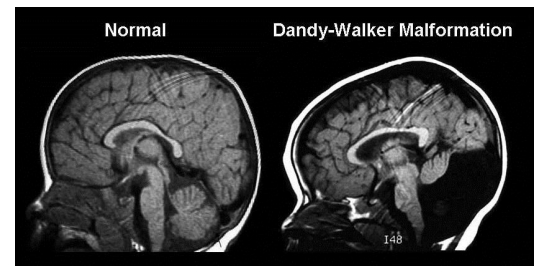


What is it?

Dandy-Walker Syndrome is a congenital birth defect that involves the cerebellum of the brain and the fluid filled spaces around it. The cerebellum, located at the back of the brain, controls movement. No gene or cause has yet to be identified but a familial link has been seen. Researchers also support that potential causes could include viral infections passed from mother to developing baby, intrauterine exposure to certain toxins or medications or diabetes. DW is often associated with other disorders of the central nervous system including absence of the corpus callosum and malformation of the heart, face and extremities. About 70% of DW patients also have hydrocephalus. Researchers have not yet determined the relationship between the degree of malformation and the severity of related effects. Key features of this include:

- The 4th ventricle, the area that allows fluid to flow between the brain and spinal cord, is enlarged
- The area between the 2 hemispheres, the cerebellar vermis, is completely absent or partial
- At the base of the skull, a cyst can form
- Increase in size of fluid filled spaces
- Increase in intracranial pressure



What are common effects?

Infants and older children can display symptoms differently. Diagnosis is normally made around 3-4 years of age, but symptoms can appear by age 1. The key is early intervention. Infants may first display signs of delayed motor development and a progressive skull enlargement. They will be behind in skills like sitting, walking and talking. Characteristics may include:

- Enlarged skull
- Increased head circumference
- Bulging at the back of the skull
- Increased intracranial pressure that includes vomiting, irritability, seizures, lack of muscle coordination
- Malformation of heart, face, limbs, fingers and toes
- Affected nerves of eyes, face, neck
- Abnormal breathing related to cerebellar dysfunction
- Lack of coordination

What is the treatment?

There is no cure for DW and treatment usually involves treating the associated symptoms. Often, a shunt may be placed when hydrocephalus is diagnosed. A shunt will help drain excess fluid in the brain to reduce swelling and intracranial pressure. A compilation of multiple affects may shorten life span. Medications are often prescribed to control seizures.

Suggested school accommodations

Effects on cognitive development is variable. Not all DW children have intellectual disability. Again, early intervention is key. Below is a list of issues and/or recommendations for school.

- PT/OT/Speech: Consult to identify needs
- Monitor visual, hearing, speech impairment
- Coordination issues of fine/gross motor skills
- Sensory issues to noise, touch, light
- Monitor for fatigue
- Monitor poor motor skills
- Access mobility devices, orthotics
- Assess seizure disorder
- Extended processing time
- Offer rest breaks as appropriate
- Offer repetition
- Visual tools/location in classroom to aid with vision
- Tools to aid in memory
- Use of assistive technology
- Offer clear, concision direction
- Allow time for response

SHNIC school nurses information:

Specific health issues for individual health care plans

- 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
- Emergency shunt protocol
- Order for types of seizures
- Child specific characteristics of seizures
- Seizure action plan
- Nutrition orders, including feeding tube replacement per county policy
- Fever protocol
- Activity and positioning restrictions
- Orders for orthotics or assistive devices
- Orders for supervision and safety

Resources & Manuals

Dandy-Walker Alliance

<http://dandy-walker.org/>

National Institute of Neurological Disorders and Stroke

<http://www.ninds.nih.gov/disorders/dandywalker/dandywalker.htm>

Genetics Home Reference: Dandy-Walker malformation

<http://ghr.nlm.nih.gov/condition/dandy-walker-malformation>