



# Dandy-Walker Syndrome

## Symptoms or Behaviors

Enlargement of the fourth ventricle

Absence (partial or complete) of the cerebellar vermis

Cyst formation in the posterior fossa (internal base of the skull)

Slow motor development in early infancy

Progressive macrocrania (abnormally enlarged skull)

Hydrocephalus

Seizures

Intracranial pressure in older children, causing irritability, vomiting, and convulsions

Cerebellar dysfunction causing ataxia and ataxia and nystagmus

Bulging occiput (back of head)

Cranial nerve dysfunction

Abnormal breathing patterns

Agenesis of the corpus callosum

Malformations of the face, limbs, digits, and heart

Cleft lip and palate

Urinary structural abnormalities

(Revised 2012)

## About the Disorder

Dandy-Walker Syndrome is a congenital brain malformation involving the cerebellum (an area at the back of the brain that controls movement) and the fluid-filled spaces around it. The key features of this syndrome are an enlargement of the fourth ventricle (a small channel that allows fluid to flow freely between the upper and lower areas of the brain and spinal cord), a partial or complete absence of the area of the brain between the two cerebellar hemispheres (cerebellar vermis), and cyst formation near the internal base of the skull. An increase in the size of the fluid spaces surrounding the brain as well as an increase in pressure may also be present. The syndrome can appear dramatically or develop unnoticed. Symptoms, which often occur in early infancy, include slow motor development and progressive enlargement of the skull. In older children, symptoms of increased intracranial pressure such as irritability, vomiting, and convulsions, and signs of cerebellar dysfunction such as unsteadiness, lack of muscle coordination, or jerky movements of the eyes may occur. Other symptoms include increased head circumference, bulging at the back of the skull, problems with the nerves that control the eyes, face and neck, and abnormal breathing patterns.

Dandy-Walker Syndrome is frequently associated with disorders of other areas of the central nervous system, including absence of the area made up of nerve fibers connecting the two cerebral hemispheres (corpus callosum) and malformations of the heart, face, limbs, fingers and toes.

## Treatment

Treatment for individuals with Dandy-Walker Syndrome generally consists of treating the associated problems, if needed. A shunt to drain off excess fluid may be placed inside the skull. This will reduce intracranial pressure and help control swelling. Parents of children with Dandy-Walker Syndrome may benefit from genetic counseling if they intend to have more children.

## Educational Implications

The effect of Dandy-Walker Syndrome on intellectual development is variable, with some children having normal cognition and others never achieving normal intellectual development even when the excess fluid buildup is treated early and correctly. Longevity depends on the severity of the syndrome and associated malformations. The presence of multiple congenital defects may shorten life span.



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## Instructional Strategies & Classroom Accommodations

- Provide a work environment that is quiet, well organized and clearly structured
- Focus on single activities and ensure competing distractions are minimized
- Ensure high demand working periods are brief and interspersed with more relaxing activities
- Instructions should be clear and step by step and repeated when required
- Adult assistance is available to redirect student after lapses in concentration
- Assist student in maintaining shared topic of conversation, and redirect as needed
- Routinely check student's understanding by asking them to paraphrase what is said to them, i.e., instructions
- Reduce amount of information presented at one time and allow extra opportunities for rehearsal
- Emphasize key points in a logical sequence
- Extraneous information should be minimized
- Reinforce conceptual learning through practical activities related to the student's interests and life experience
- Break down complex tasks such as assignments and projects into smaller steps, helping the student generate a plan of approach before they begin. Review progress after each step has been completed and provide visual cues to guide the student's progress
- Encourage organized work habits, such as consistent homework times, assignment log, checklists, use of color coding
- Encourage student to check and proofread their work
- Watch for symptoms of shunt malfunction

Note: Document all accommodations on the student's IEP.

## Resources

### National Institute of Neurological Disorders and Stroke

NIH Neurological Institute  
P.O. Box 5801, Bethesda, MD 20824  
[www.ninds.nih.gov](http://www.ninds.nih.gov)  
(800) 352-9424

### Dandy-Walker Alliance, Inc.

4422 Clearbrook Lane  
Kensington, MD 20895  
[comments@dandy-walker.org](mailto:comments@dandy-walker.org)  
<http://www.dandy-walker.org>  
877-Dandy-Walker

### Guardians of Hydrocephalus Research Foundation

2640 East 28th Street  
Brooklyn, NY 11235  
[GHRF2618@aol.com](mailto:GHRF2618@aol.com)  
<http://ghrf.homestead.com/ghrf.html>, 718-743-GHRF (4473)

### Hydrocephalus Association

870 Market Street, Suite 705  
San Francisco, CA 94102  
[info@hydroassoc.org](mailto:info@hydroassoc.org)  
<http://www.hydroassoc.org>  
415-732-7040 888-598-3789

### March of Dimes Foundation

1275 Mamaroneck Avenue  
White Plains, NY 10605  
[askus@marchofdimes.com](mailto:askus@marchofdimes.com)  
<http://www.marchofdimes.com>  
914-428-7100 888-MODIMES  
(663-4637)

### National Organization for Rare Disorders (NORD)

P.O. Box 1968, 55 Kenosia Ave  
Danbury, CT 06813-1968  
[orphan@rarediseases.org](mailto:orphan@rarediseases.org)  
<http://www.rarediseases.org>  
203-744-0100  
Voice Mail 800-999-NORD (6673)



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