# Dandy-Walker Syndrome

Dandy-Walker Syndrome is a *congenital* (present since birth) 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 *cerebellar vermis* (the area between the two cerebellar hemispheres), 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* (problems), which often occur in early infancy, include slow *motor* (movement) 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 *corpus callosum* (the connecting area between the two cerebral hemispheres), 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. A special tube to reduce intracranial pressure may be placed inside the skull to control swelling. Parents of children with Dandy Walker Syndrome may benefit from genetic counseling if they intend to have more children.

## WHAT WILL HAPPEN

Children with Dandy-Walker Syndrome may never have normal intellectual development, even when the excessive accumulation of cerebrospinal fluid in the brain 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.

## RESEARCH BEING DONE

The NINDS conducts and supports a wide range of studies that explore the complex mechanisms of normal brain development. The knowledge gained from these fundamental studies provides the foundation for understanding abnormal brain development and offers hope for new ways to treat and prevent developmental brain disorders such as Dandy-Walker Syndrome.

**Research is continually being done to learn more about this problem.**

Provided by: The National Institute of Neurological Disorders and Stroke

Document Released: 12/08/2003 Document Re-Released: 03/16/2010

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