What is Dandy-Walker Syndrome?

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 *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, 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 *corpus callosum* (the connecting area between the two cerebral hemispheres, and malformations of the heart, face, limbs, fingers and toes.

Is there any treatment?

Treatment for individuals with Dandy-Walker Syndrome generally consists of treating the associated problems, if needed. A special tube 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.

What is the prognosis?

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 hydrocephalus 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.

What research is being done?

The National Institute of Neurological Disorders and Stroke 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.

Who is the Dandy-Walker Alliance and what is the purpose?

We are an all-inclusive organization comprised of individuals directly and indirectly affected by Dandy-Walker sharing a collective interest in educational, informational activities and supporting non-partisan research to increase public awareness of the congenital birth defect Dandy-Walker. We also support all efforts to determine the cause(s), find the cure and to ameliorate the effects of Dandy-Walker. We believe that by making findings available to families affected in an organized and accessible way and by disseminating the direct and indirect outcomes of translational research we can more swiftly move the results from the bench to the bedside in an expeditious manner.

Source: National Institute of Neurological Disorders and Stroke, 2008.