Dandy-Walker Syndrome

Dandy-Walker Syndrome is a rare 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.

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.

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.

Resources

NOTE: This page contains links to other World Wide Web sites with information about this disorder. The Department of Health and Senior Services (DHSS) hopes you find these sites helpful. Remember the DHSS does not control nor does it necessarily endorse the information presented on these web sites.

For a complete list of resources related to birth defects, including state programs and resources, support groups and not-for-profit organizations click on the following link. http://www.health.mo.gov/living/families/genetics/birthdefects/resources.php

- National Institute of Neurological Disorders and Stroke
- National Organization for Rare Disorders (NORD)

Genetic Tertiary Centers

How to Obtain Genetic Services
Your family physician can usually provide information regarding genetic services in your area. Genetic clinics are periodically held in a location near you. For information, contact one of the centers listed below.

Cardinal Glennon Children's Medical Center
St. Louis, Missouri 314-577-5639

Children's Hospital at University Hospital and Clinics
Columbia, Missouri 573-882-6991

Children's Mercy Hospital
Kansas City, Missouri 816-234-3290

St. Louis Children's Hospital
St. Louis, Missouri 314-454-6093