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Dandy-Walker Syndrome

National Institute of Neurological Disorders and Stroke (NINDS)

Source

National Institute of Neurological Disorders and Stroke (NINDS). [Dandy-Walker Syndrome](#).

Dandy-Walker Syndrome is a congenital brain malformation involving the cerebellum (an area of the back of the brain that coordinates 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 lowest part of the skull. An increase in the size and pressure of the fluid spaces surrounding the brain (hydrocephalus) 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 (pressure within the skull) such as irritability and vomiting, 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, abnormal breathing problems, and problems with the nerves that control the eyes, face and neck. Dandy-Walker Syndrome is sometimes 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.