



Fact Sheet

Dandy Walker Syndrome

By Marty Elquist and MaryAnn Demchak

What is Dandy-Walker Syndrome? Dandy-Walker syndrome is a brain malformation involving the cerebellum (an area at the back of the brain that controls movement) and the fluid filled spaces around it. Common 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, which is important for the coordination of voluntary muscle movements), 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 (National Institute of Neurological Disorders and Stroke, 2005). The syndrome has an extreme range of severity. Some people with a Dandy Walker variant live their entire lives without any symptoms. Others have it in association with other symptoms, resulting in severe disabilities or even death (Incesu & Khosla, 2003).

What are the Symptoms? Diagnosis generally takes place between 3-4 years of age, but ranges anywhere from 9 months of age to 12 years of age (NORD, 2003). Symptoms that often occur in early infancy include slow motor development and progressive enlargement of the skull. An estimated 80% of patients have normal ventricles at birth, and by age one, they have an enlargement of the lateral ventricle to greater than or equal to 10 millimeters (Incesu & Khosla, 2003). This enlargement is known as ventriculomegaly. Ventriculomegaly is often secondary to hydrocephalus, which is caused by increased cerebrospinal fluid in the brain. Hydrocephalus is present in approximately 90% of patients at the time of diagnosis (see *Focus on Hydrocephalus*: www.unr.edu/educ/ndsip/newsletters/sept2002.pdf).

Symptoms that occur in older children can be related to increased brain pressure such as irritability, vomiting, and convulsions, as well as signs of cerebellar dysfunction (problems with the portion of the brain that coordinates the movement of voluntary muscles). Signs of cerebellar dysfunction include unsteadiness, lack of muscle coordination, or jerky movements of the eyes. 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 other disorders 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.

What Causes Dandy Walker Syndrome? The cause of Dandy Walker is largely unknown (Incesu & Khosla, 2003). Although family (generic) occurrence has been reported, it is believed to be the result of multiple factors (NORD, 2003). Incesu & Khosla report that some predisposing factors might include

exposure to rubella, cytomegalovirus (see *Focus on Cytomegalovirus: www.unr.edu/educ/ndsip/newsletters/april2003.pdf*), toxoplasmosis, warfarin (Cumadin), alcohol, and/or isotretinoin during the first trimester.

What are the Implications of Dandy Walker for Vision? Dandy-Walker syndrome can affect vision in a variety of ways. Common vision problems may include: nystagmus (involuntary back and forth movement of eyes); cataracts; retinal dysgenesis – abnormal formation of the retina (back part of the eye that is sensitive to light and is connected to the brain via the optic nerve); and/or choroid coloboma – abnormal formation of the eye between the sclera (white part of the eye) and the retina; vision problems due to hydrocephalus (can include visual field defects, faulty depth perception, trouble with shape recognition, and cortical visual impairment); and/or occipital encephalocele – defect in the closure of the neural tube near the base of the skull (Nelson, 2002).

What are the Implications of Dandy Walker for Hearing? Hearing loss is not a primary characteristic of Dandy Walker syndrome; however, there may be sensorineural loss, conductive hearing loss (especially in children who also have a cleft palate), or central loss due to agenesis of the corpus callosum (Nelson, 2002).

What is the frequency of Dandy Walker Syndrome in the U.S? The incidence of Dandy-Walker malformation is 1 case per 25,000-35,000 live births, Dandy-Walker malformation accounts for approximately 1-4% of hydrocephalus cases. The sex incidence of those who have Dandy Walker syndrome is approximately 40% female to 60% male (NORD, 2003).

Is There a Treatment for Dandy Walker Syndrome? Treatment for individuals with Dandy-Walker syndrome generally consists of treating the associated problems, if needed. A special tube (shunt) may be placed inside the skull to reduce pressure in the brain and control swelling. Additionally, it will be determined on an individual basis whether or not special education and related services (e.g., physical therapy, occupational therapy, and speech therapy) are needed. These educational services are determined by the individuals' needs and are intended to assist the child in receiving an appropriate education.

References

- Incesu, L., & Khosla, A. (2003, October). *Dandy Walker malformation*. Retrieved July 14, 2005 from <http://www.emedicine.com/radio/topic206.htm>
- National Institute of Neurological Disorders and Stroke. (2005, February). *Dandy Walker syndrome information page*. Retrieved July 14, 2005 from <http://www.ninds.gov/disorders/dandywalker>
- National Organization for Rare Disorders (NORD). (2003). *NORD guide to rare disorders*. Philadelphia: Lippincott Williams & Wilkins.
- Nelson, C. (2002, September). Dandy Walker malformation. Presentation at the meeting of the National Technical Assistance Consortium Area 1, Portland, OR.

For more information

Tanni Anthony	Phone: 303-866-6681	Email: anthony_t@cde.state.co.us
Gina Quintana	Phone: 303-866-6605	Email: quintana_g@cde.state.co.us

Fact Sheets from the Colorado Services to Children and Youth with Combined Vision and Hearing Loss Project are to be used by both families and professionals serving individuals with deafblindness. The information applies to children, birth through 21 years of age. The purpose of the Fact Sheet is to give general information on a specific topic. More specific information for an individual student can be provided through personalized technical assistance available from the project. Reviewed and updated: 11/09

Information for this Fact Sheet supplied with permission from the Nevada Dual Sensory Impairment Project. The article was published in their Fall 2005 Newsletter, Volume 15, Number 4.