

# Dandy Walker Malformation

BIRTH DEFECT RESEARCH FOR CHILDREN



## What is Dandy Walker Malformation?

Dandy Walker Malformation is a congenital (present at birth) malformation of the brain. It involves the cerebellum, which is the lower, hind portion of the brain. Cyst formation and an associated enlargement of the fourth ventricle (cavity within the brain) that frequently results in hydrocephalus (excess fluid in the brain) characterize Dandy Walker Malformation. Hydrocephalus results from the cyst's interference with the function of draining cerebrospinal fluid from the brain. Dandy Walker Malformation is sometimes seen with other abnormalities such as cleft palate, enlarged kidneys with cysts, spinal cord defects, and other brain defects.



# Dandy Walker Malformation



How many children have Dandy Walker Malformation?

Dandy Walker Malformation occurs in one out of 25,000 babies, affecting girls more often than boys.

How do you know if your child has Dandy Walker Malformation?

Dandy Walker Malformation is usually discovered by ultrasound before the baby is born. If not diagnosed before birth, then it is usually diagnosed within the first year of life. Symptoms in infants tend to be related to hydrocephalus and pressure within the head. Hydrocephalus can cause enlargement of the skull, especially the forehead. Other symptoms in newborns may include a lump on the back of the head, a patch of long hair above this lump, irritability, poor feeding, and recurrent vomiting. Diagnosis is usually made by x-ray or a procedure called transillumination. The diagnosis is often confirmed by a CT (computer tomography) scan. This is a special x-ray to determine the size of the ventricle, whether there is an obstruction, and the size and location of the obstruction.

What Causes Dandy Walker Malformation?

In most cases, the cause of Dandy Walker Malformation is unknown. A few cases have resulted from autosomal recessive genes, meaning both parents carried the gene. Dandy Walker cysts form during early embryonic development when the brain forms. Exposure to isotretinoin (a synthetic vitamin A derivative used to treat acne) increases the risk of Dandy Walker Malformation.

How can you help a child with Dandy Walker Malformation?

The primary malformation of the brain cannot be corrected. The hydrocephalus usually requires a surgical by-pass (shunt) to allow the continuous drainage of the blocked fluid. A team of specialists is helpful for children with Dandy Walker Malformation and usually should include a physical therapist, pediatric neurologist, pediatric neurosurgeon, and educational specialist.

What's in the future for a child with Dandy Walker Malformation?

The prognosis can be moderately favorable for a child with Dandy Walker Malformation, but it depends on whether there are any associated abnormalities and their severity. Generally, the more abnormalities, the poorer the prognosis. Early intervention is important because untreated hydrocephalus can cause mental retardation. Possible affects of Dandy Walker Malformation include developmental delay, learning disabilities, subnormal intelligence, motor skill deficits, and seizures. Fifty percent of children with Dandy Walker Malformation are in the mainstream at school.

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