What is Dandy-Walker Syndrome?

Dandy-Walker is a brain developmental condition in which the cerebellum, the part of the brain responsible for the coordination of movement, is malformed. Typically, the central part of the cerebellum called the vermis is absent or has not developed fully.

Hydrocephaly is a related complication in individuals with Dandy-Walker due to cysts that form in the brain’s fourth ventricle and block the flow of cerebrospinal fluid (CSF). This blockage allows excessive CSF to accumulate, swelling the ventricle and putting pressure on the brain. This swelling can increase the child’s head size resulting in a condition known as macrocephaly (“macro” meaning big and “cephaly” meaning head).

Prevalence and Symptoms

The prevalence rate of Dandy Walker is 1 in every 25,000 newborns, and signs and symptoms of the syndrome typically occur within the first year of a child’s life. While some children with Dandy-Walker have normal intelligence (rare), the associated brain abnormalities caused by Dandy-Walker usually result in intellectual impairments that can range from mild to profound depending on the abnormalities of the child’s brain. Symptoms of Dandy-Walker vary with age and can include the following:

Symptoms in Infancy:
- slow motor development
- progressive skull enlargement

Symptoms in Older Children:
- signs of intracranial pressure, including irritability and vomiting
- unsteadiness
- lack of muscle coordination
- jerky eye movements
- increased head circumference
- bulging at the back of the skull
- abnormal breathing patterns

Other symptoms and effects associated with Dandy-Walker may include hypertonia; leg paralysis; seizures; malformations of the face, arms, legs, fingers, and toes; and vision and hearing problems, including deafblindness.

While rare, death can result in children with Dandy-Walker syndrome, but is usually due to complications related to hydrocephaly.

Causes of Dandy-Walker

The etiology of Dandy-Walker is suspected to be genetic in nature. Dandy-Walker syndrome has been found to occur most often in children who have an extra copy of chromosome 18 (trisomy 18). However, Dandy-Walker is not exclusive to children with trisomy 18, nor does it occur in every child with trisomy 18. Dandy-Walker has also been associated with trisomy 13, 21, & 9.

While children and brothers and sisters of people with Dandy-Walker have been shown to be at an increased risk of acquiring the syndrome, there is not an established pattern of inheritability of the syndrome, and Dandy-Walker can occur in children whose families have no history of the disorder.

Treatment

There is no treatment for the physical brain development abnormalities that are present with Dandy-Walker syndrome, however, a Ventriculoperitoneal shunt can be surgically implanted to allow excessive cerebrospinal fluid to be drained away from the fourth ventricle of the brain, reducing complications from hydrocephalus.

For motor complications and neurological and cognitive disabilities that are present, a team approach that utilizes various therapies and educational adaptations designed to meet each child’s specific mobility and learning needs will be necessary.

References:


Other symptoms and effects associated with Dandy-Walker may include:
- increased head circumference
- bulging at the back of the skull
- abnormal breathing patterns

Taken from http://www.ninds.nih.gov/disorders/dandywalker/dandywalker.htm