Hydranencephaly

Hydranencephaly is a rare condition. With this condition, the brain's cerebral hemispheres (the largest and main parts of the brain) are absent. They are replaced by sacs filled with cerebrospinal fluid.

CAUSES

Hydranencephaly may be caused by infections of the brain or its blood vessels or blockage of blood vessels to the brain after the 12th week of pregnancy.

SYMPTOMS

An infant with hydranencephaly may appear normal at birth. The infant's head size and reflexes may all seem normal. These include:

- Sucking.
- Swallowing.
- Crying.
- Moving the arms and legs.

But after a few weeks the infant usually becomes irritable and has increased muscle tone. After a few months of life, certain symptoms may develop. They are:

- Convulsions (*seizures*).
- Large accumulation of cerebrospinal fluid in the brain (*hydrocephalus*).

Other symptoms may include:

- Visual impairment.
- Lack of growth.
- Deafness.
- Blindness.
- Paralysis (*spastic quadriparesis*).
- Intellectual deficits.

DIAGNOSIS

Diagnosis may be delayed for several months because some brain tissue is present and early behavior appears to be normal. Some infants may have additional abnormalities at birth. These include:

- Seizures.
- Spasm or twitching of a muscle or group of muscles (*myoclonus*).
- Respiratory problems.

TREATMENT

There is no treatment for this condition. Treatment is symptomatic and supportive.

The outlook for children with this condition is poor. Death generally happens before age 1.

RESEARCH BEING DONE

The NINDS conducts and supports a wide range of studies that explore the complex mechanisms of normal brain development. The knowledge gained from these studies provides the foundation for understanding how this process can go awry. It offers hope for new means to treat and prevent developmental brain disorders, including hydranencephaly.

Research is continually being done to learn more about this problem.

Provided by: The National Institute of Neurological Disorders and Stroke

Document Released: 12/08/2003 Document Re-Released: 10/20/2009

ExitCare® Patient Information ©2011 ExitCare, LLC.