

Facts about Encephalocele



Encephalocele is a rare type of neural tube defect (NTD) present at birth that affects the brain. The neural tube is a narrow channel that folds and closes during the third and fourth weeks of pregnancy to form the brain and spinal cord. Encephalocele is described as a sac-like protrusion or projection of the brain and the membranes that cover it through an opening in the skull. Encephalocele happens when the neural tube does not close completely during pregnancy. The result is an opening in the midline of the upper part of the skull, the area between the forehead and nose, or the back of the skull.

Usually encephaloceles are found right after birth, but sometimes a small encephalocele in the nose and forehead region can go undetected.

What We Know About Encephalocele

How often does encephalocele occur?

CDC estimates that each year about 375 babies in the United States are born with encephalocele (1). In other words, about 1 out of every 10,000 babies born in the United States each year will have encephalocele.

What problems do children with encephalocele have?

When located in the back of the skull, encephalocele often is linked to nervous system problems. Encephalocele usually is seen with other brain and face defects.

Signs of encephalocele can include:

- Buildup of too much fluid in the brain
- Complete loss of strength in the arms and legs
- An unusually small head
- Uncoordinated movement of the voluntary muscles, such as those involved in walking and reaching
- Developmental delay
- Vision problems
- Mental and growth retardation
- Seizures

What We Still Do Not Know About Encephalocele

What causes encephalocele?

Although the exact cause of encephalocele is unknown, scientists believe that many factors are involved.

There is a genetic component to the condition, meaning it often occurs among families with a history of spina bifida and anencephaly. Some researchers also believe that certain environmental exposures before or during pregnancy might be causes, but more research is needed. We at CDC work with many other researchers to study risk factors that can increase the chance of having a baby with encephalocele, as well as outcomes of babies with encephalocele. Following is an example of what our research has found:

- Several factors appear to lead to lower survival rates for infants with encephalocele, including preterm birth, low birth weight, having multiple birth defects, and being Black or African American.²(<http://www.cdc.gov/ncbddd/birthdefects/encephalocele.html#ref>)

Can encephalocele be prevented?

Currently, there is no known way to prevent encephalocele, although steps can be taken to lower the risk. Recent studies have shown that the addition of a B vitamin called folic acid to the diet of women who might become pregnant can greatly reduce the number of babies born with NTDs. CDC has recommended that all women of childbearing age consume 400 micrograms of folic acid daily. A single daily serving of most multivitamins and fortified cereals contain 400 micrograms of folic acid.

In addition, mothers can take steps before and during pregnancy to be healthy, including not smoking and not drinking alcohol during pregnancy.

Resources for Families and Individuals Affected by Encephalocele

[National Institute of Neurological Disorders and Stroke](#)

[National Organization for Rare Disorders](#)

References

1. Canfield MA, Honein MA, Yuskiv N, Xing J, Mai CT, Collins JS, et al. National estimates and race/ethnic-specific variation of selected birth defects in the United States, 1999-2001. *Birth Defects Res Part A Clin Mol Teratol.* 2006;76(11):747–56.
2. Siffel C, Wong LC, Olney RS, Correa A. Survival of infants diagnosed with encephalocele in Atlanta, 1978-98. *Paediatr Perinat Epidemiol.* 2003; 17:40-8.