# Lissencephaly

Lissencephaly, which literally means "smooth brain," is a rare brain formation disorder characterized by the lack of normal convolutions (folds) in the brain, and an abnormally small head (microcephaly). It is caused by defective neuronal migration, the process in which nerve cells move from their place of origin to their permanent location. Symptoms of the disorder may include unusual facial appearance, difficulty swallowing, failure to thrive, and severe psychomotor retardation. Anomalies of the hands, fingers, or toes, muscle spasms, and seizures (convulsions) may also occur. Lissencephaly may be associated with other diseases including isolated lissencephaly sequence, Miller-Dieker syndrome, and Walker-Warburg syndrome.

## TREATMENT

The severe malformations of the brain in lissencephaly most likely will not respond to treatment. Normal supportive care may be needed to help with comfort and nursing needs. Seizures may be controlled with medication. Progressive hydrocephalus (an excessive accumulation of cerebrospinal fluid in the brain) may require shunting. If feeding becomes difficult, a gastrostomy tube may be considered.

## WHAT WILL HAPPEN

The prognosis (likely outcome) for children with lissencephaly is variable depending on the degree of brain malformation. Many individuals show no significant development beyond a 3- to 5-month-old level. Some may have near-normal development and intelligence. Many will die before the age of 2. Aspiration and respiratory disease are the most common causes of death.

## RESEARCH BEING DONE

The NINDS conducts and supports a wide range of studies that explore the complex systems of normal brain development, including neuronal migration. The knowledge gained from these studies provides the foundation for understanding how this process can go awry and, thus, offers hope for new means to treat and prevent neuronal migration disorders such as lissencephaly.

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

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