

# Lissencephaly

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## History

Child with developmental delay.

## Diagnosis

Lissencephaly

## Discussion

Lissencephaly is a rare developmental defect of the brain. Lissencephaly is a descriptive term used in reference to the flat surface of the brains of lower mammals; agyria-pachygyria complex is the preferred term for human brain migrational anomalies.

There are two types of lissencephaly. In Type I the radial migration of neurons from the periventricular germinal zone towards the cortical plate is interrupted around the 13th week of gestation. This defect is often related to abnormality of the LIS 1 gene on chromosome 17; sometimes the genetic defect is x-linked. The brain is small. The cerebral cortex is 4-layers instead of 6. Facial dysmorphism is fairly recognizable. Syndromes associated with Type I lissencephaly include Miller-Dieker, Norman-Roberts and Neu-Laxova.

Type II lissencephaly the cerebral cortex is totally disorganized. Disruption of the glia limitans leads to overmigration of glioneural tissue into the subarachnoid space. The brain surface is not always agyric but can have broad gyri or even areas of polymicrogyria. Three autosomal recessive disorders associated with type II lissencephaly are Walker-Warburg, Fukuyama congenital muscular dystrophy and cerebro-ocular muscle dystrophy.

Patients with lissencephaly have seizures and global developmental delay. The age of onset is variable and related to the degree of parenchymal involvement.

## Findings

MR-Small cerebrum with smooth surface, radially oriented dysplastic/cystic T1-hypointense and T2-hyperintense regions, and enlarged ventricles.

## Reference

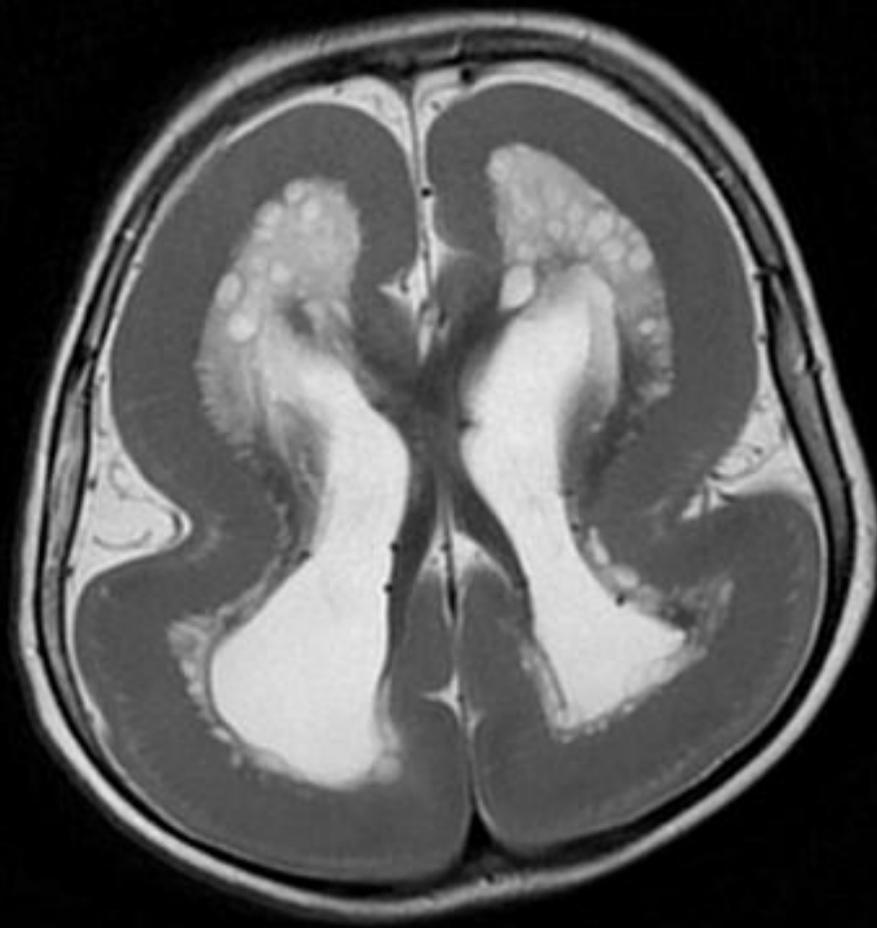
Stoltenburg-Didinger G and Steinbrecher A. Morphogenesis of Type II Lissencephaly: Neuropathology, Genetics and Pathomechanisms. *Neuroembryology* 2003; 2:32-39.

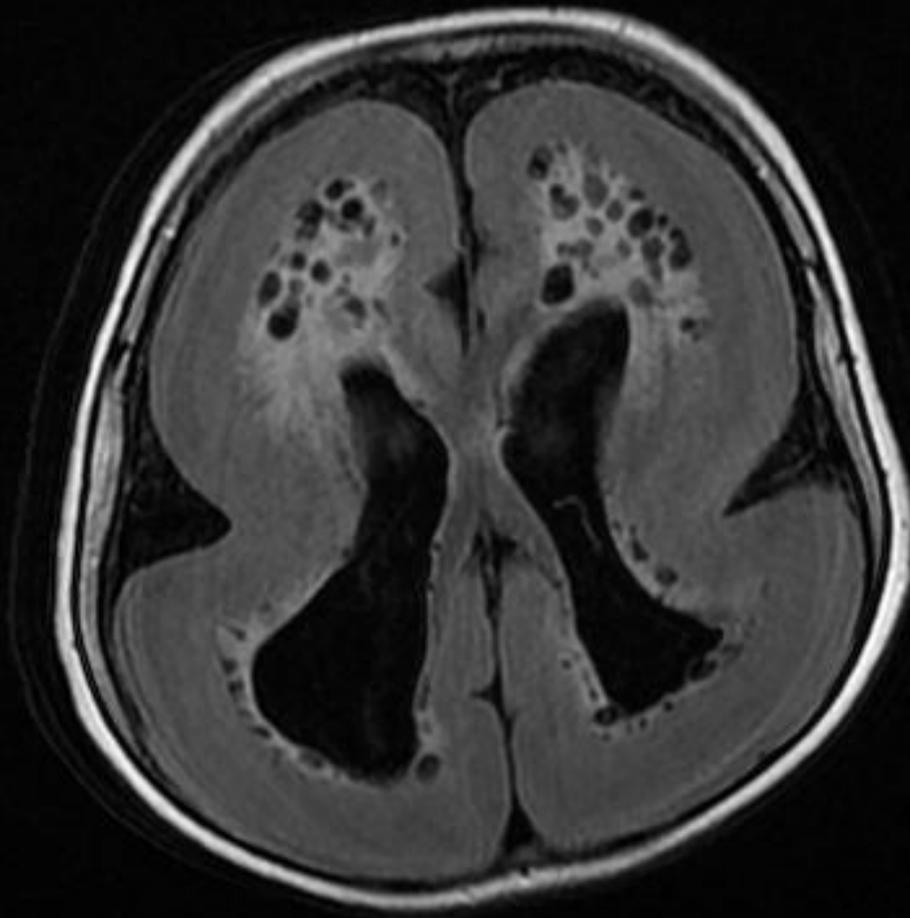
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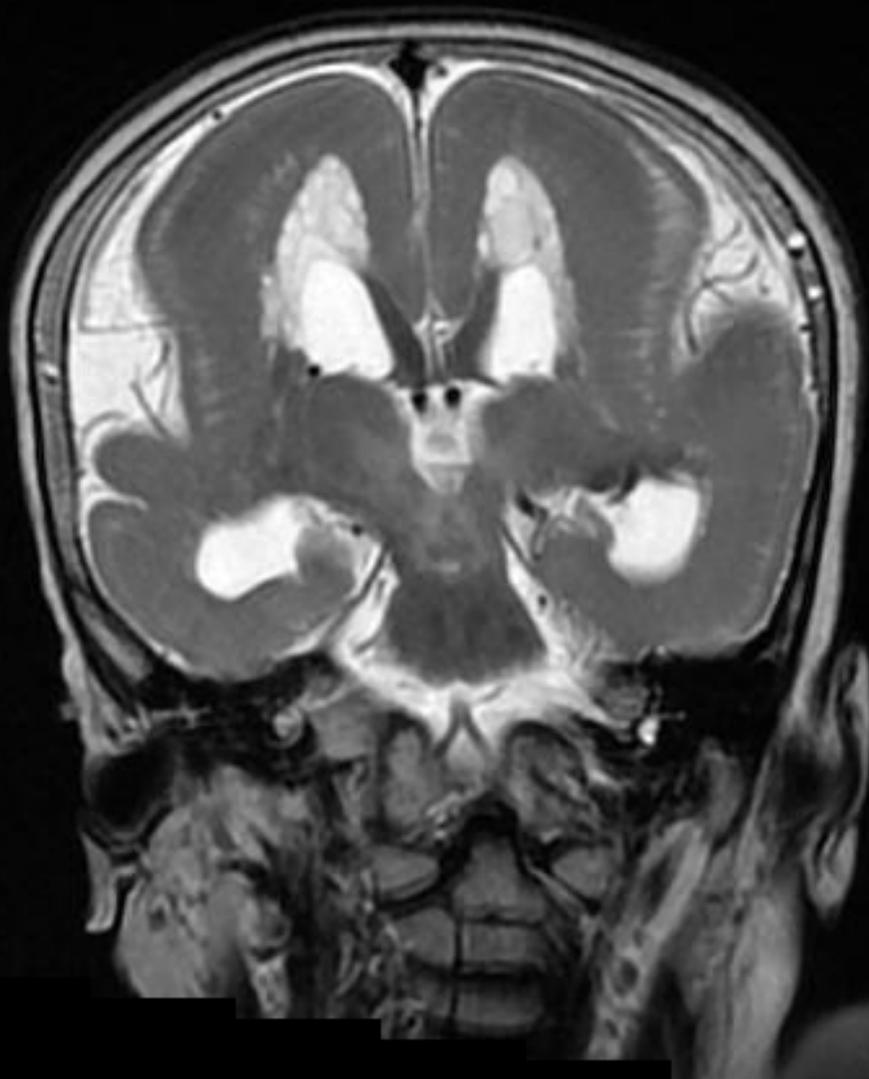
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