

# Lobar Holoprosencephaly

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## History

2 year old with microcephaly and multiple anomalies.

## Diagnosis

Lobar holoprosencephaly

## Discussion

Holoprosencephaly is the result embryologic failure of cleavage of the prosencephalon. Disruption of cleavage affects the olfactory and optic bulbs and as a result holoprosencephaly is associated with dysmorphic facies (including cyclopia, ethmocephaly, cebocephaly, and premaxillary agenesis). Diverticulization of the prosencephalon into right and left telencephalons is also variably affected. Maternal diabetes mellitus, fetal alcohol syndrome, antenatal infections and chromosomal abnormalities have been implicated in the development of holoprosencephaly.

Holoprosencephaly represents a spectrum of disease. The most severe form is alobar holoprosencephaly. This form is associated with a single crescentic monoventricle which communicates with a dorsal cyst, fused basal ganglia and thalami, poorly developed intracranial arterial supply and rudimentary or absent sagittal sinus. Semilobar holoprosencephaly represents an intermediate form where the posterior cerebral hemispheres are partially separated, the posterior corpus callosum is present posteriorly, the temporal horns are small and the thalami are partially fused. Lobar holoprosencephaly is the least severe form with better cerebral differentiation.

## Findings

MR-Frontal lobes are continuous; the anterior interhemispheric fissure is absent. The frontal horns of the lateral ventricles are very small and dysplastic and the septum pellucidum and anterior corpus callosum are absent.

## Reference

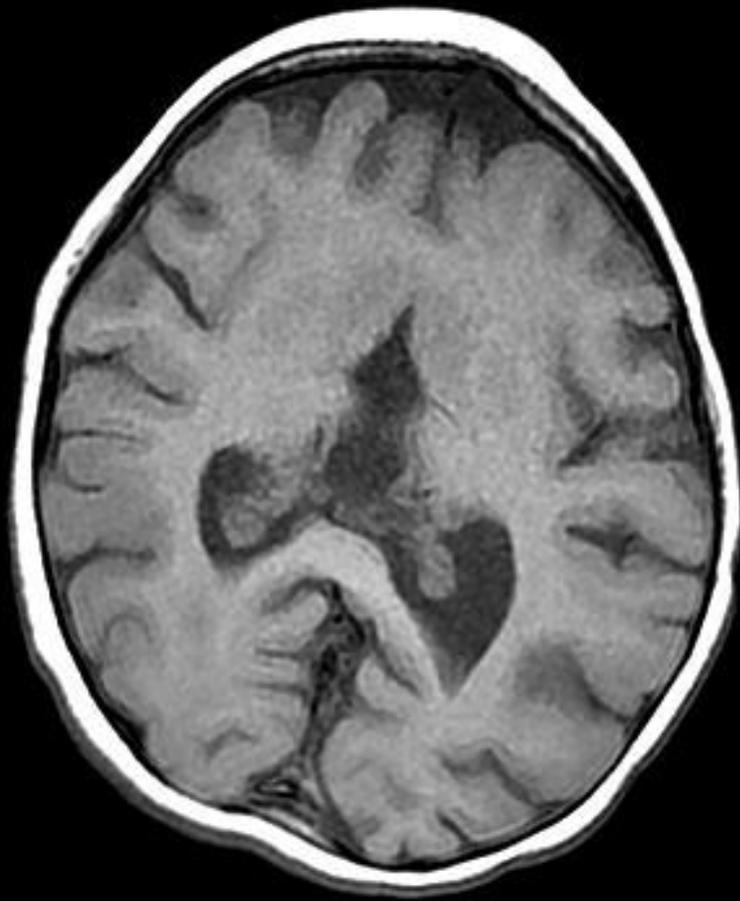
Barkovich AJ. Pediatric Neuroimaging, 4th Ed. Lippincott, Williams and Wilkins (2005).

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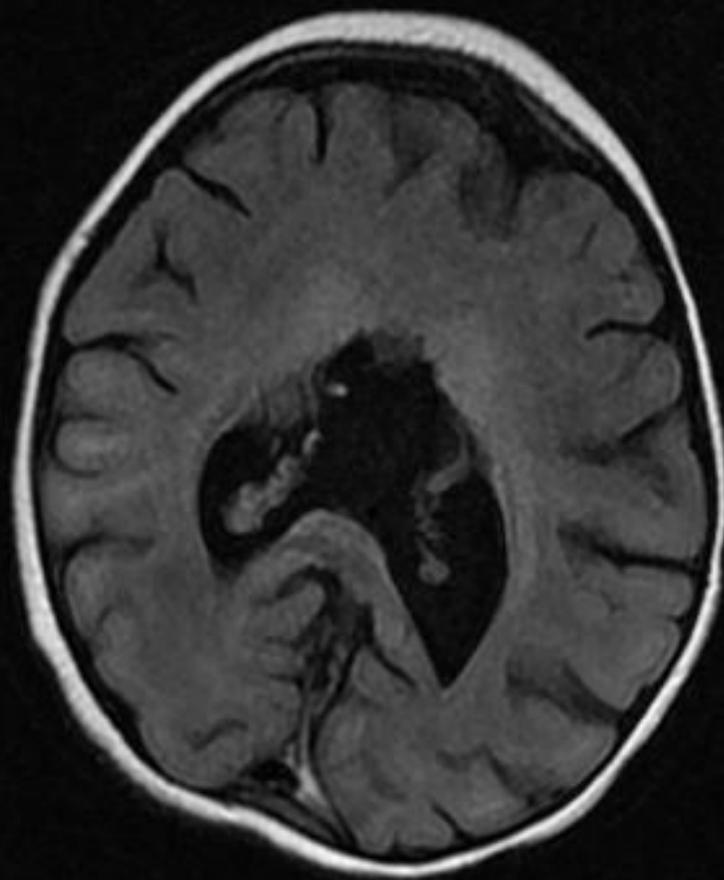
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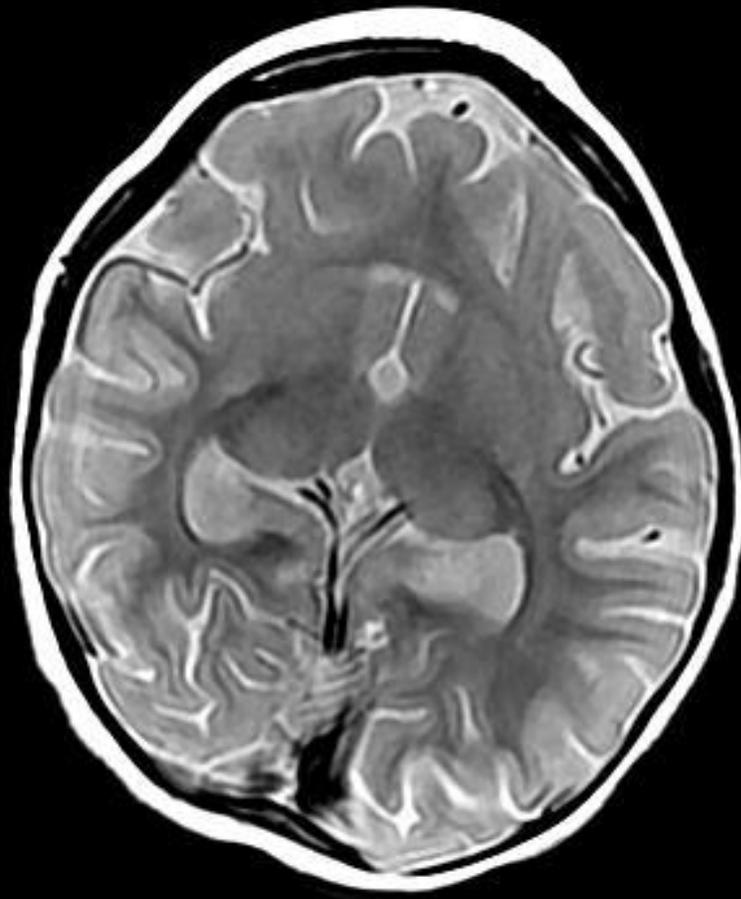
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