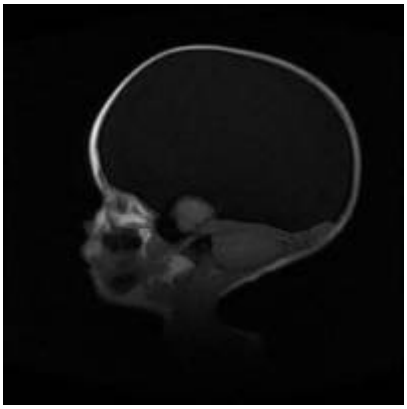
# Hydranencephaly



Hydranencephaly is a condition in which the brain's cerebral hemispheres are absent to a great degree.

Absence of the septum pellucidum.

The remaining cranial cavity is filled with cerebrospinal fluid. Hydranencephaly is a type of cephalic disorder.

Causes failure of normal skull growth. There is usually progressive macrocrania, but head size may be normal (especially at birth), and, occasionally, microcephaly may occur. Facial dysmorphism is rare.

Very severe forms of schizencephaly may mimic hydranencephaly.

## Differential diagnosis

Differentiation from hydrocephalus:

Progressive enlargement of CSF spaces may occur which can mimic severe (“maximal”) hydrocephalus (HCP). It is critical to differentiate the two since true HCP may be treated by shunting which may produce some re-expansion of the cortical mantle. Many means to distinguish hydranencephaly and HCP have been described, including:

1. EEG: shows no cortical activity in hydranencephaly (maximal HCP typically produces an abnormal EEG, but background activity will be present throughout the brain) and is one of the best ways to differentiate the two. [1)].

2. CT [2) 3)] , MRI or ultrasound: majority of intracranial space is occupied by CSF. Usually do not see frontal lobes or frontal horns of lateral ventricles (there may be remnants of temporal, occipital or subfrontal cortex). A structure consisting of brainstem nodule (rounded thalamic masses, hypothalamus) and medial occipital lobes sitting on the tentorium occupies a midline position surrounded by CSF. Posterior fossa structures are grossly intact. The falx is usually intact (unlike alobar holoprosencephaly), and is not thickened, but may be displaced laterally. In HCP, some cortical mantle is usually identifiable

3. transillumination of the skull: in a darkened room, a bright light is placed against the surface of the

skull. To transilluminate, the patient must be < 9 mos old and the cortical mantle under the light source must be <1 cm thick [4] , can also occur if fluid displaces the cortex inward (e.g. subdural effusions). Too insensitive to be very helpful.

4. angiography: in “classic” cases resulting from bilateral ICA occlusion, no flow through supraclinoid carotids and a normal posterior circulation is expected.

# Treatment

Shunting may be performed to control head size, but unlike the case with maximal hydrocephalus, there is no restitution of the cerebral mantle.

[1] , [2]
Sutton LN, Bruce DA, Schut L. Hydranencephaly ver- sus Maximal Hydrocephalus: An Important Clinical Distinction. Neurosurgery. 1980; 6:35–38
[3]
Dublin AB, French BN. Diagnostic Image Evaluation of Hydranencephaly and Pictorially Similar Entities with Emphasis on Computed Tomography. Radiology. 1980; 137:81–91
[4]
Matson DD. Neurosurgery of Infancy and Childhood. 2nd ed.Springfield: Charles CThomas; 1969