

Clinical Pediatric Neurology

CONGENITAL MALFORMATIONS OF THE BRAIN:

Malformation	Description
Lissencephaly	Absence of sulci, "smooth brain"
Pachygyri	Few gyri that are broad and thick. "Incomplete lissencephaly". (<i>pachy means thick</i>).
Polymicrogyri	Numerous small gyri
Schizencephaly	Cleft lined with grey matter connecting the ventricle to the pial surface of the brain. Either open lip or closed lip.
Porencephaly	Cleft or cyst in the cerebral hemisphere, <u>not</u> lined with grey matter; "encephalomalacia".
Hydranencephaly	Cerebral hemispheres are absent and replaced by CSF
Holoprosencephaly	Forebrain fails to divide into two hemispheres. <ul style="list-style-type: none"> Alobar : Severe form, hemispheres fail to separate resulting in cyclopia. : hemispheres are partially separated Semilobar : Incomplete separation, may lead normal life. Lobar : inter-hemispheric fissure is complete, thalami separated but fornices are fused, frontal horns of lateral ventricle are fused, septum pellucidum absent
Anencephaly	Absence of the brain, skull and scalp
Encephalocele	A sac like protrusion of the brain and meninges
Septo-optic dysplasia (SOD) (De Morsier synd)	2 of the following: absence of septum pellucidum, pituitary gland and optic nerves. Can be caused by in utero Valproate exposure.
Dandy Walker Syndrome:	
A. DW malformation	Most severe form, consists of: <ul style="list-style-type: none"> -Absence of cerebellar vermis -Large posterior fossa and highly tentorium -Cystic dilatation of 4th ventricle
B. DW Variant:	Mild form with hypoplastic vermis, mildly enlarged 4 th ventricle
Chiari Syndrome:	
A. Chiari I:	Displacement of Peg tail like cerebellar tonsils through foramen magnum > 5mm
B. Chiari II: (Arnold Chiari)	Displacement of medulla, 4 th ventricle and vermis through foramen magnum. Usually associated with peaked tectum and lumbar myelomeningocele
C. Chiari III:	Similar to Chiari II with occipital encephalocele
D. Chiari IV (Obsolete):	Cerebellar hypoplasia (now called primary cerebellar agenesis).

CONGENITAL DISORDERS OF THE SKULL: