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**What are basic differences between anencephaly and acrania?**

With anencephaly one would expect to see no brain, skull or scalp present above the eyes in a fetus. If brain is present but exposed this is termed acrania meaning absence of the cranium. With acrania the cerebral hemispheres are largely present but often still abnormal and covered only with a thin membrane. Acrania alone is often lethal and acrania can progress to anencephaly.

**What are basic differences between aqueductal stenosis with massive hydrocephalus, hydranencephaly and holoprosencephaly?**

With aqueductal stenosis with massive hydrocephalus one should be able to see a thin rim of peripheral brain tissue and presence of the falx.

With holoprosencephaly one would expect to see a monoventricle with no falx. Holoprosencephaly results from incomplete separation of the two hemispheres. Remember the association with Patau syndrome/trisomy 13. Holoprosencephaly will often have other associated anomalies to include facial abnormalities, renal abnormalities, and things like polydactyly.

With hydranencephaly some sort of insult such as ischemia (often bilateral MCA thrombosis) or infection (to include congenital HSV/CMV/toxoplasmosis) causes supratentorial cerebral parenchymal destruction resulting in loss of brain tissue and a residual CSF-filled membranous sac from ex vacuo expansion of the ventricles. Presence of the falx is expected. Hydranencephaly is the sequela of supratentorial destruction of a normally-formed brain so is not associated with things like facial abnormalities. Be cautious that with hydranencephaly the brainstem can bulge and mimic fused thalami, causing potential confusion with holoprosencephaly.

**What is porencephaly (sometimes termed a porencephalic cyst)?**

Porencephaly is a mild form of hydranencephaly in which only a small portion of brain tissue has been destroyed with a residual CSF-filled space occupying the area of insult. Remember that a porencephalic cyst does not act as a space occupying lesion and does not enlarge over time. It is simply the fluid filled remnant of what was once brain parenchyma.

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**What are some key features of Patau syndrome?**

Patau syndrome has an association with holoprosenchephaly, polydactyly, hitchhiker thumb and echogenic bowel on ultrasound. Remember this is a chromosomal 13 trisomy anomaly. Alobar holoprosencephaly is often considered a characteristic association of trisomy 13. About 75% of neonates with Patau syndrome do not survive past 6 months. Don't confuse this with Edwards syndrome that has a chromosomal trisomy 18 association and is associated with cardiac disease, rocker bottom foot, clenched 3$^{rd}$ and 4$^{th}$ fingers, intrauterine growth retardation and perinatal demise.

**What are some key differences between a Chiari II malformation and Dandy Walker?**

Chiari II: Posterior fossa is too small. Other features include the banana cerebellum and lemon head configuration on prenatal ultrasound, absent cisterna magna, ventriculomegaly, spinal dysraphism, a beaklike tectal plate, tonsillar herniation, thalamic pseudofusion, and a hypoplastic falx with cerebral interdigitations.

With Dandy Walker the posterior fossa is too big with cystic dilatation of the 4$^{th}$ ventricle. Has varying degrees of vermian hypoplasia. More than half have hydrocephalus and may have other anomalies such as polydactyly, corpus callosum dysgenesis, cardiac anomalies, etc.

**What are key features of Chiari I malformation?**

Chiari I malformation has tonsillar ectopia, typically at least 5 mm below the foramen magnum. Chiari I has risk of hydrocephalus secondary to a ball and valve type obstruction at the foramen magnum and risk of syrinx. Chiari I is usually asymptomatic. Treatment can include shunting and posterior fossa decompression surgery.

**What is more common: congenital cytomegalovirus (CMV) or congenital toxoplasmosis?**

Congenital CMV is something like 10 times more common than congenital toxoplasmosis. One key feature that can help separate these entities on imaging is that congenital CMV is more associated with periventricular calcifications whereas congenital toxoplasmosis can show random, scattered calcifications.

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**What are key features of Meckel-Gruber syndrome?**

Meckel-Gruber syndrome is considered a triad of:

1. Renal cystic dysplasia/multiple renal cysts

2. Holoprosencephaly/occipital encephalocele

3. Polydactyly

This is an autosomal recessive disease similar but different than trisomy 13. This is often fatal at birth due to renal failure and/or pulmonary hypoplasia.

**What is rhombencephalosynapsis?**

Rhombencephalosynapsis is basically an apparently fused cerebellum with vermian agenesis.

**What is Joubert syndrome?**

Joubert syndrome is sometimes termed cerebello-oculo-renal syndrome. Cerebellum anomalies include congenital vermian hypoplasia or aplasia and abnormal shaped 4$^{th}$ ventricle. Renal anomalies include small dysplastic kidneys. Ocular anomalies include abnormal eye movements.

**What are key differences between alobar, semilobar and lobar holoprosencephaly?**

Alobar holoprosencephaly has features that include fused thalami, a monoventricle, no interhemispheric fissure, no falx or corpus callosum.

Semilobar holoprosencephaly has partially fused thalami, a partially formed falx and presence interhemispheric fissure posteriorly with fusion of the anterior brain.

Lobar holoprosencephaly has hypoplastic frontal lobes, a falx that extends frontally, and presence of the temporal horns of the lateral ventricles. Septooptic dysplasia may be considered the least severe form of lobar holoprosencephaly and has absent septum pellucidum, hypoplastic optic nerves and olfactory bulbs and pituitary dysfunction.

**What are differences between schizencephaly and lissencephaly?**

Lissencephaly: brain is smooth with subcortical band heterotopia. Schizencephaly: brain has abnormal clefts lined by gray matter that, if extending to ventricle, can be considered an open lip schizencephaly.

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### What is pachygyria-polymicrogryria?

Pachygyria-polymicrogyria is a cortical malformation with features of too thick and too numerous gyri. Association with CMV and metabolic derangements including Zellweger's syndrome (cerebrohepatorenal syndrome).