CNS Pathologies

CH 60 1385-1405

Anomalies associated with
Ventriculomegaly
Craniosynostosis
Holoprosencephaly
Dandy-Walker Complex
Agenesis of Corpus Callosum
Microcephaly
Spina Bifida
Cephalocele
Vascular Malformations
Porencephaly
Schizencephaly
Intracranial Hemorrhage
Lissencephaly
Infection

Other Central Nervous System Anomalies
Anencephaly
Dandy-Walker Malformation
Hydranencephaly
Choroid Plexus Cysts
Aqueductal Stenosis
Vein of Galen Aneurysm
Porencephalic Cyst
Cephalocele

Central Nervous System

Ectodermal neural plate @ 18GD
Prosencephalon = forebrain
Mesencephalon = midbrain
Rhombencephalon = hindbrain
Most defects come from the incomplete closure of the neural tube

Normals for
CSP= 18-37 weeks
Cisterna magna = 2-10mm
Lateral Ventricle at atrium = <10mm
Cerebellum = # weeks gestation
Nuchal Fold = <6mm

Anencephaly
MC neural tube defect 1:1000
1:1000 births
Lethal – absence of brain, failure of closure of neural tube at cranial end.
Associated with other anomalies: Acrania is a precursor, trisomy 13 & 18, cleft lip/palate, hydronephrosis, polyhydramnios.
Sono: bulging orbits (frog eyes), absence of brain and calvarium.
Female > Male White > Black
Risks: amniotic band syndrome, diabetes

Holoprosencephaly : Forebrain
Alobar, Semi-Lobar, Lobar
Aneuploidy, rare hereditary component
Risks: alcohol, phenyltoin, retinoic, radiation exposure, oral contraceptives during 1st trimester
Outcome: Poor or severe mental retardation
Sono: Abnormal facial features, first trimester abnormal choroid plexus (no butterfly)
Alobar: most severe: pancake, cup, ball single monoventricle brain, fused thalamus, no CC, CSP, or optic
Semi Lobar: Single ventricle, partial occipital horns, no CC, CSP, or olfactory bulbs
Lobar: Almost complete division of ventricles with w/o CC, no CSP

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Ventriculomegaly
Dilation of the ventricles
May be caused by an obstruction in ventricles
Rarely from overproduction of CSF
Linked with aqueductal stenosis, arachnoid cysts, and VAGAs. 80% have associated anomalies like trisomy 13/18
Causes include spina bifida and encephaloceles
SONO: l
lateral vent >10mm
"dangling choroid sign"
Possible dilation of 3/4th ventricles

Hydrocephalus
Dilation of ventricles WITH enlarged fetal head = 2+ standard deviation of BPD/HC

Dandy Walker Malformation
Hindbrain with agenesis/hypoplasia of cerebellar vermis, dilation of 4th ventricle, enlarged posterior fossa
Associations: 50% aneuploidy, macrocephaly, maternal diabetes, cardiac/urinary/face/phalanges defects
Sono: splayed cerebellum, dilation of 4th ventricle, ventriculomegaly
Differentials: arachnoid cysts, cerebellar hypoplasia
Prognosis: severity of mental impairments depends on other findings.

Agenesis of Corpus Callosum
Corpus callosum develops between 12-20 weeks. Communication between hemispheres
Sono: absence of corpus callosum
dilation of third ventricle "tear drop"
Dilated occipital horns
Absence of cavum septum pellucidum
Linked with other CNS anomalies
X linked syndromes

Agenesis of Corpus Callosum (Cont)
Chromosomal anomalies (21,13,18)
Aicardi, Apert, Opitz, Joubert syndrome
Maternal disease, infection, and alcohol abuse
Prognosis: isolated = asymptomatic but may have seizures mental retardation. With other anomalies = poor prognosis

Hydranencephaly
Sono: absence of brain tissue, fluid filled brain, absent or partially absent falx
Presence of midbrain, cerebellum, possible macrocephaly.
Causes: occlusion of the internal carotid arteries. By congenital infection or ischemia.
Differentials: Hydrocephaly, Holoprosencephaly

Microcephaly
Small head, measuring more than 2 standard deviations behind
A sloping forehead may also be seen in the profile view.
Disorganized brain tissue
Linked with aneuploidy, cranial anomalies, and other syndromes.
Risk factors are alcohol abuse, heroin, mercury poisoning, radiation, and hypoxia.

Aqueductal Stenosis
From obstruction, atresia, or stenosis of the aqueduct of sylvius.
Sono: ventriculomegaly, flexion and adduction of the thumb.

Craniosynostosis

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Premature fusion of the skull that results in a “clover leaf” appearance.

Choroid Plexus Cysts

Round or ovoid anechoic structures within the choroid plexus
Unilateral or bilateral
Usually isolated finding, resolve between 22-26 weeks
Linked with aneuploidy, heart defects, abnormal hands/feet.
*** check for open hand***

Vein of Galen Aneurysm (VAGA)
Rare arteriovenous malformation
Isolated or CHD, cystic hygromas, hydrops
May cause neurologic damage
Poor prognosis
Sono: cystic space midline and posterior to third ventricle. Turbulent flow within
Male predominance
Differentials: arachnoid cysts (no blood flow)