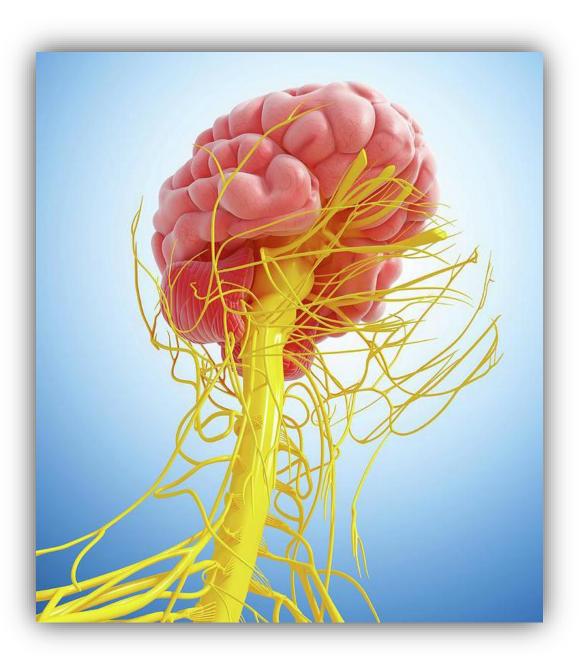
OB GYN SONOGRAPHY REVIEW

The Fetal Central Nervous System



Course Outline

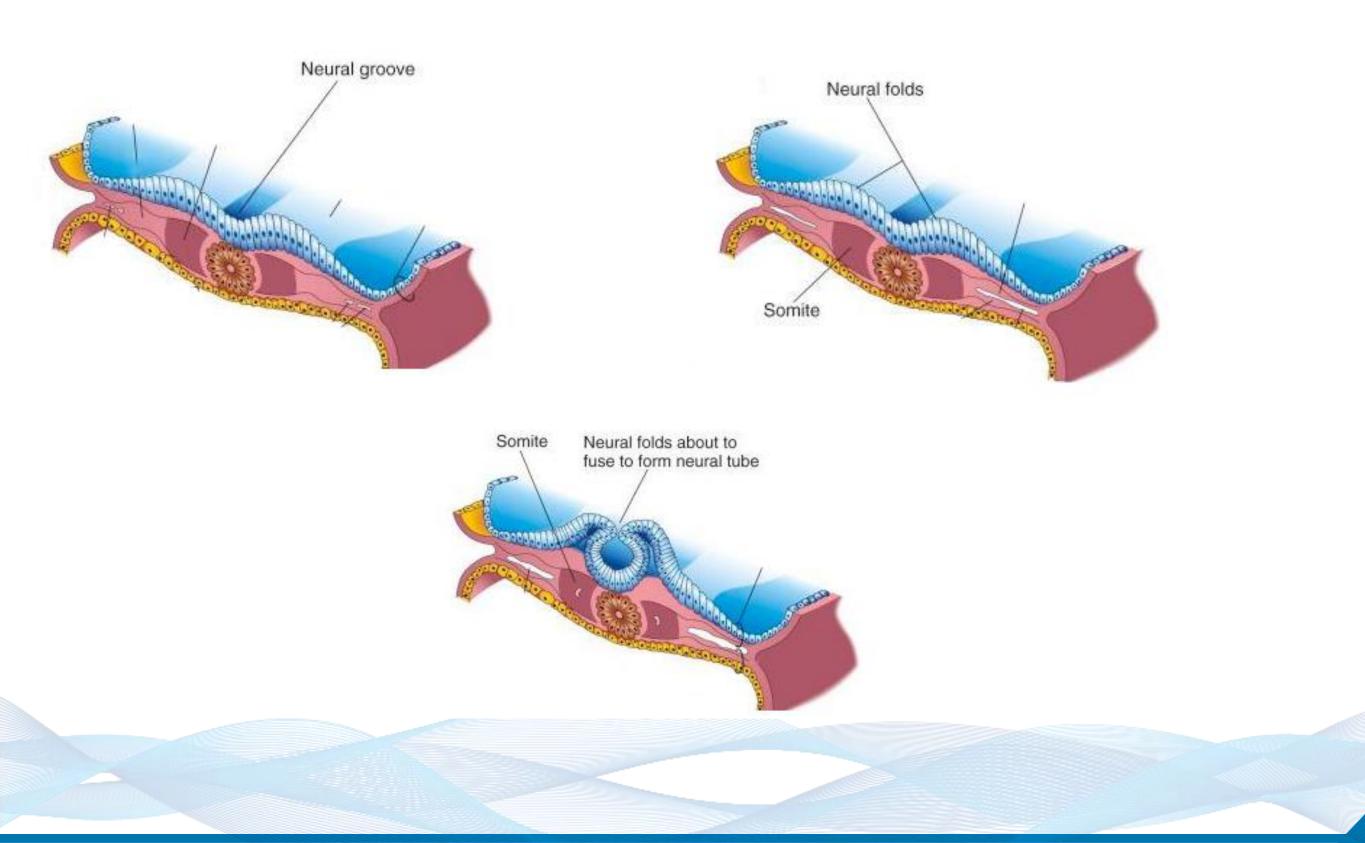
- Embryology
- Normal Sonographic Anatomy
 - Brain
 - Spinal column
- CNS Abnormalities



Embryology

Embryological Development

- Neurulation: the process by which the neural tube ss formed:
 - Begins 21 23 days post conception (35-37 menstrual days)
 - Neural plate invaginates along central axis to form neural groove
 - Neural folds rise up and grow toward the midline
 - Come together and fuse to form neural tube

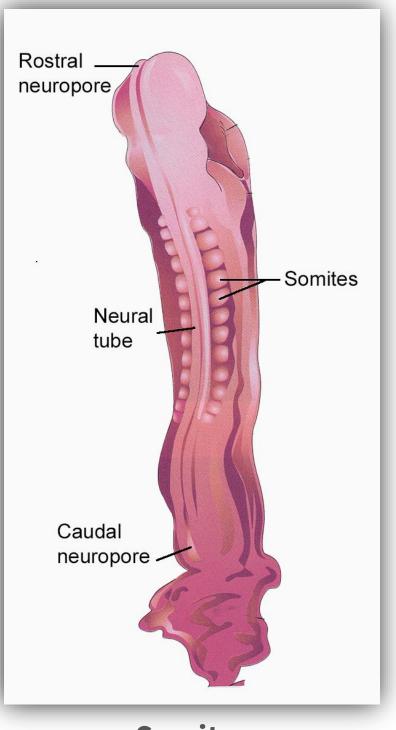


Neural Tube

- Fusion of the neural tube begins in dorsal aspect of embryo and proceeds in both cranial and caudal directions
- Neural canal communicated freely with amniotic cavity via *neuropores*
 - Rostral neuropore (head end) closes at 25 days (conceptual)
 - Caudal neuropore (tail end) closes at 27 days (conceptual)
- Failure of neuropore closure results in *neural tube defect*

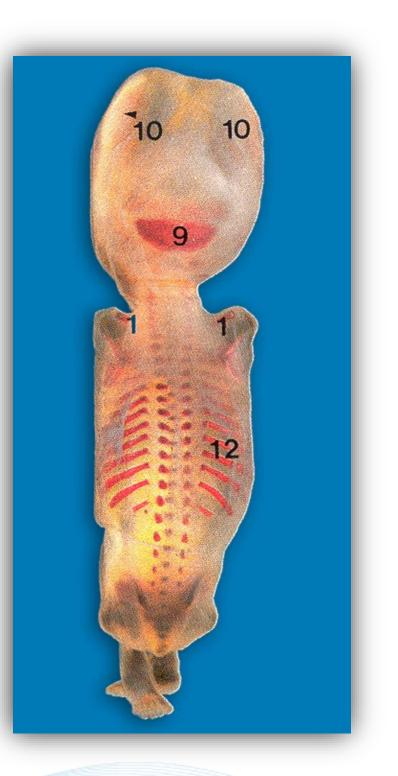
Somites

- Blocks of mesodermal tissue located on either side of neural tube
- Embryonic precursors to vertebrae
- Ossification centers begin to develop around 8 weeks (conceptual)
 - Will serve as landmarks in sonographic assessment of fetal spine



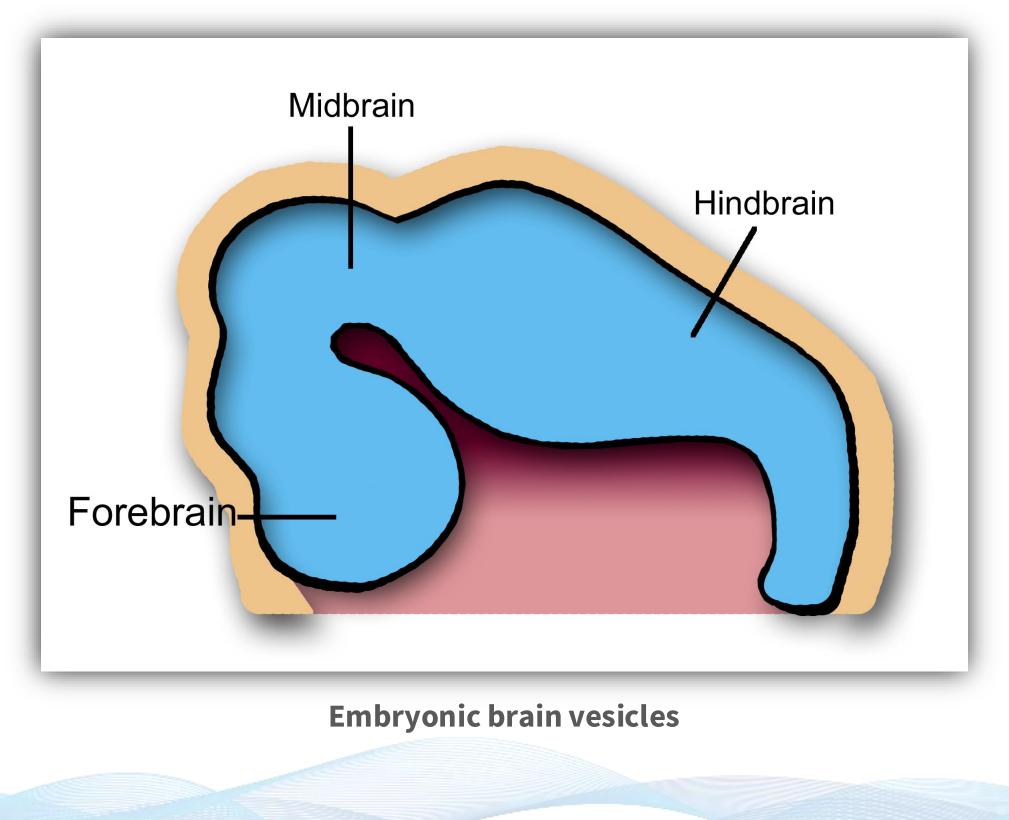
Somites





Intracranial Vesicles and Ventricles

- After closure of the rostral neuropore, three primary vesicles arise:
 - Prosencephalon: gives rise to forebrain structures
 - Mesencephalon: gives rise to midbrain structures
 - Rhombencephalon: gives rise to posterior fossa structures

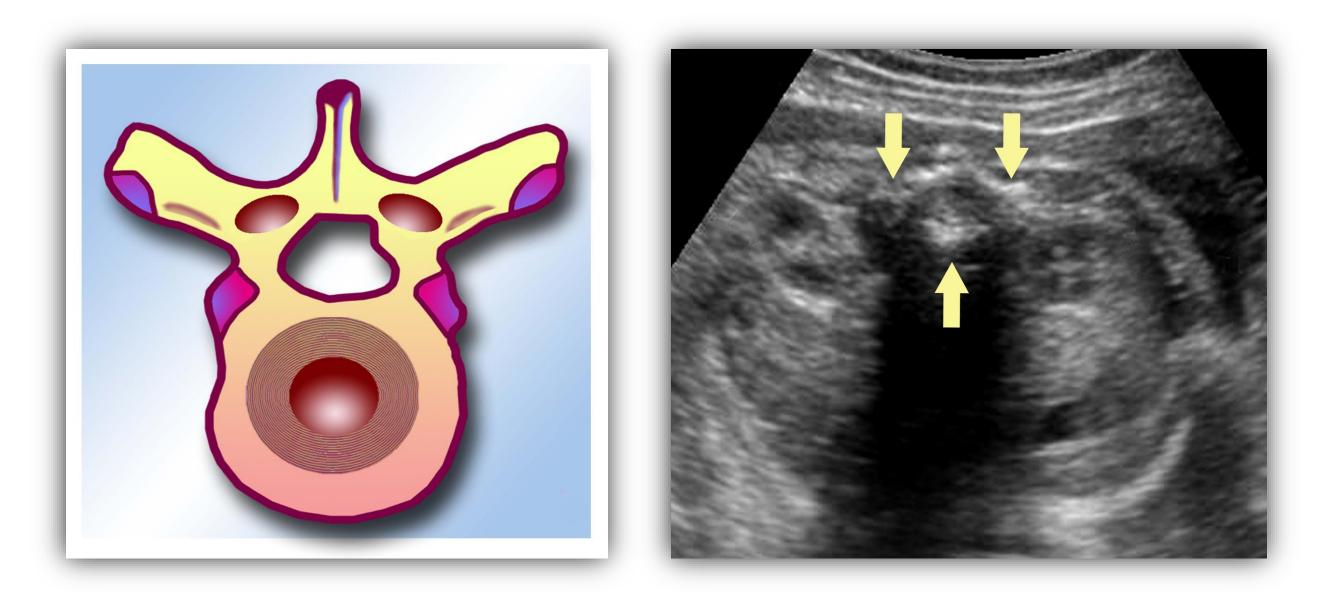


Normal Sonographic Anatomy

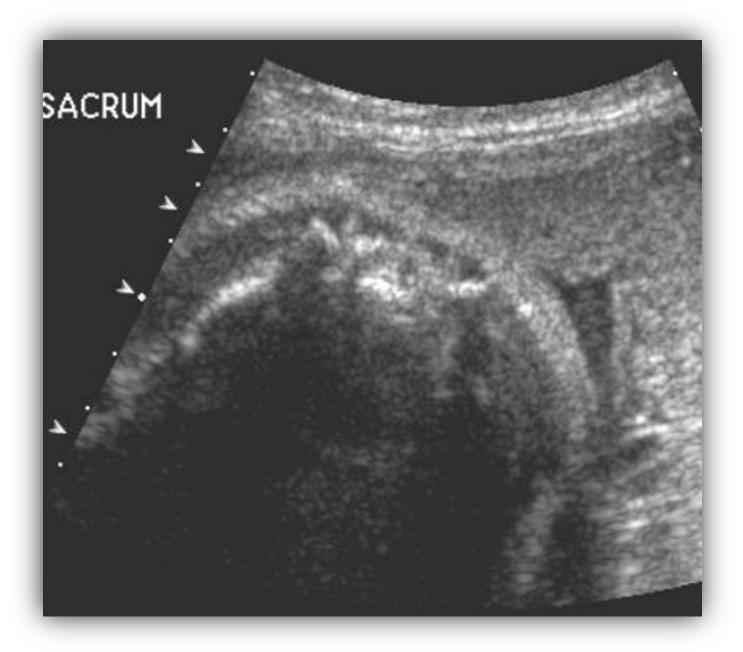
NORMAL SONOGRAPHIC ANATOMY

Fetal Spine

- The fetal spine is imaged in two orthogonal planes of sections
- *Transverse*, or *axial*, section to assess:
 - Location, configuration, and number of ossification centers in each vertebra
 - Integrity of musculature and soft tissues of the back
 - Integrity of the posterior skin line



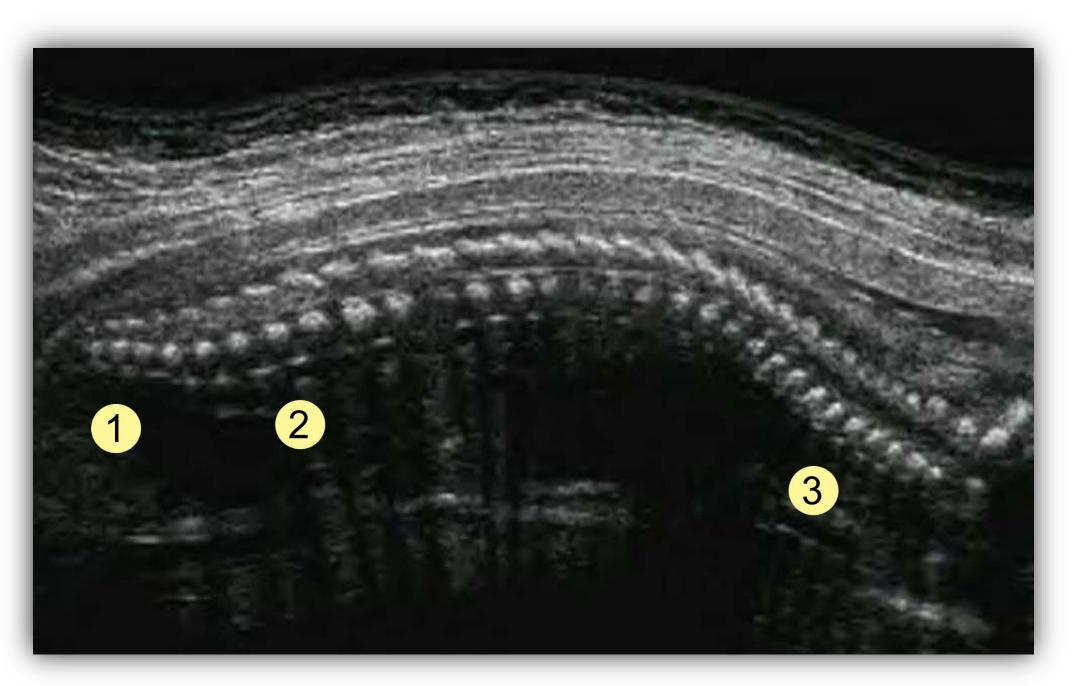
Location, configuration, number of ossification centers in each vertebra



Integrity of musculature, soft tissues and skin line

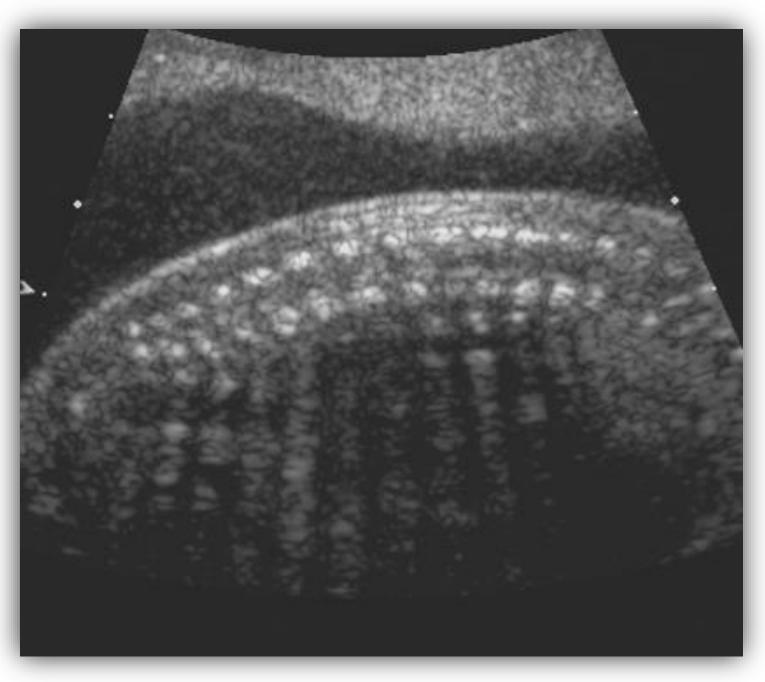
Fetal Spine

- The fetal spine is imaged in two orthogonal planes of sections
- Sagittally, or coronally, section to assess:
 - Cervical and lumbar curvatures
 - Sacral-caudal tapering
 - Configuration of vertebral ossification centers

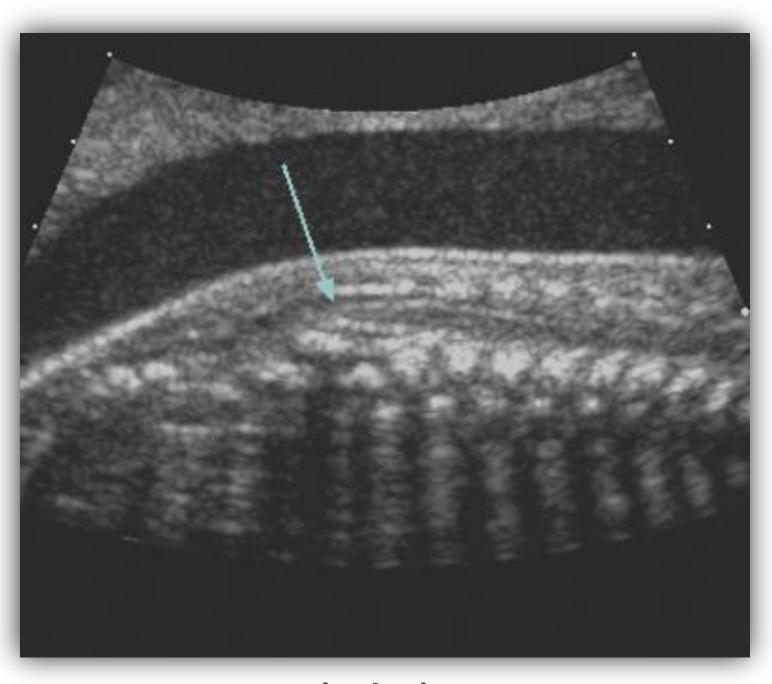


Sagittal spine

1 = sacral-caudal tapering
2 = lumbosacral curvature
3 = cervical curvature



Sagittal spine Configuration of ossification centers



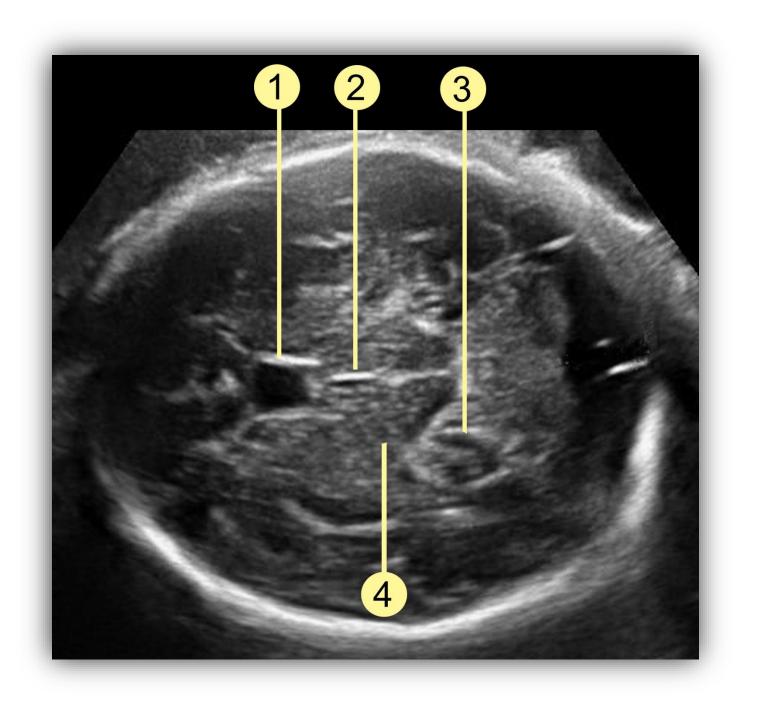
Sagittal spine Abnormal - hemivertebrae



Sagittal spine Fetal demise

Fetal Brain

- The fetal brain is imaged in *axial* planes of sections at multiple levels to assess:
 - Cavum septi pellucidi
 - Both lateral ventricles
 - Thalami
 - Choroid plexuses



2 = third ventricle

1 = cavum septi pellucidi **3** = atrium of lateral ventricle 4 = thalamus

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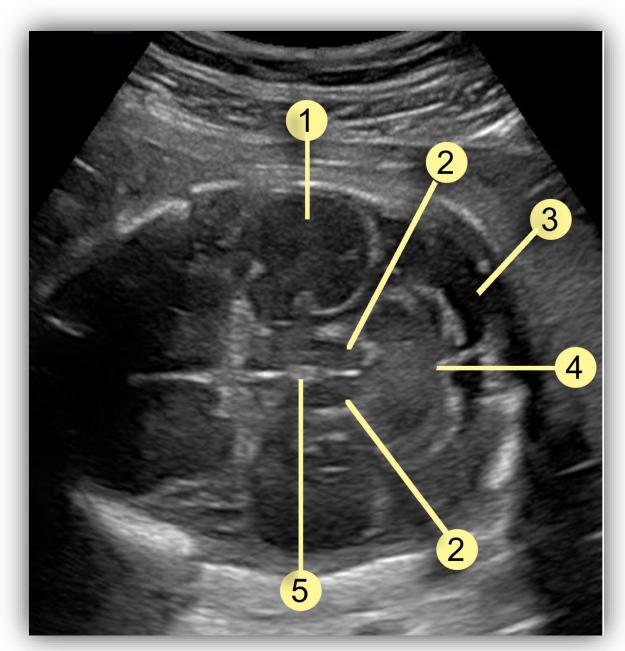
Axial section

Fetal Brain

- The fetal brain is imaged in *axial* plane of sections with measurements taken of:
 - Atrium of the lateral ventricle
 - Biparietal diameter
 - Head circumference

Fetal Brain

- The fetal brain is imaged in an *axial oblique* plane of sections through the posterior fossa to assess:
 - Location, configuration, and number of ossification centers in each vertebra
 - Integrity of musculature and soft tissues of the back
 - Integrity of the posterior skin line



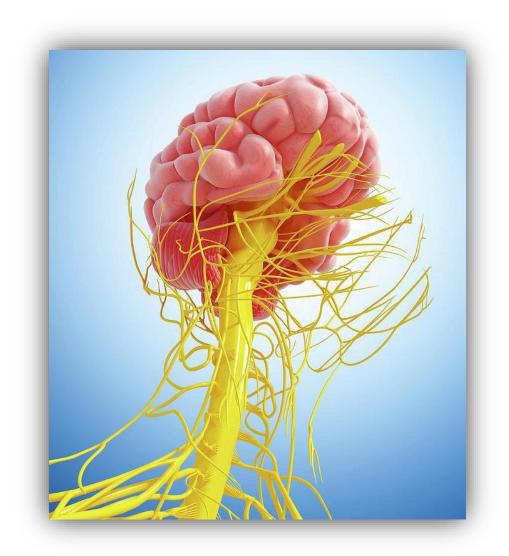
Axial oblique section

1 = temporal lobe 2 = cerebral peduncles 3 = cisterna magna 4 = cerebellum 5 = aqueduct of Sylvius

CNS Abnormalities

Categories of CNS Abnormalities

- Intracranial cystic abnormalities
- Intracranial solid abnormalities
- Neural tube defects



CNS ABNORMALITIES

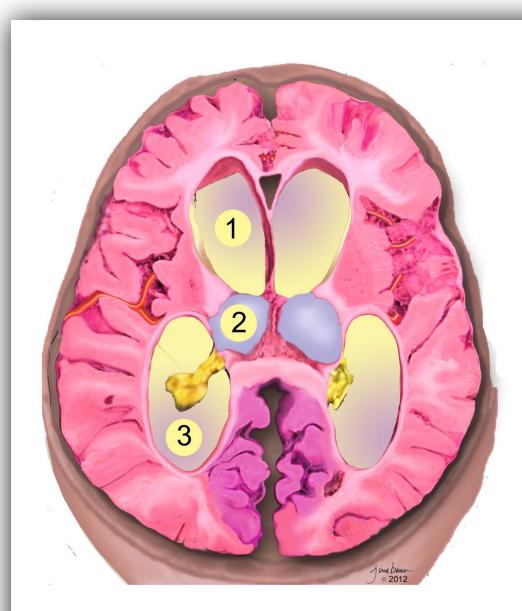
Intracranial Cystic Abnormalities

- Hydrocephalus
- Hydranencephaly
- Holoprosencephaly
- Dandy-Walker malformation
- Vein of Galen aneurysm
- Choroid plexus cysts

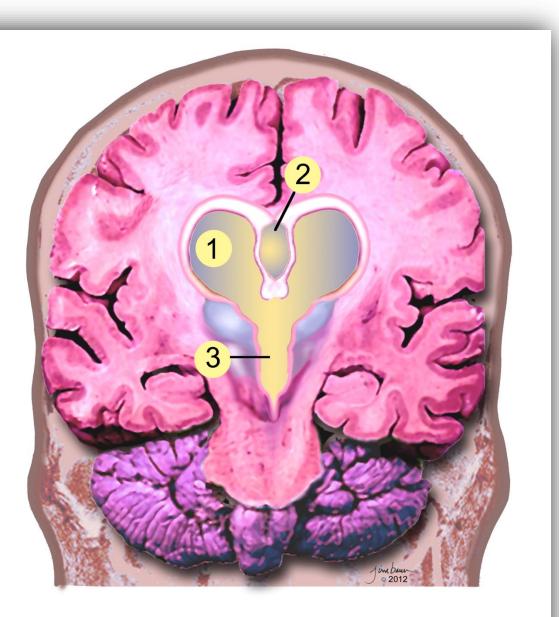
INTRACRANIAL CYSTIC ABNORMALITIES

Hydrocephalus

- An increase in the amount of cerebrospinal fluid resulting in dilatation of the ventricular system
- Four classifications:
 - Aqueductal stenosis (43%)
 - Communicating hydrocephalus (38%)
 - Dandy-Walker malformation (12%)
 - Idiopathic (7%)



- 1 = frontal horn, lateral ventricle
- 2 = thalamus
- 3 = atrium, lateral ventricle



1 = lateral ventricle2 = cavum septum pellucidum3 = 3rd ventricle

INTRACRANIAL CYSTIC ABNORMALITIES

Hydrocephalus

- Associated abnormalities:
 - CNS anomalies
 - Craniofacial anomalies
 - Syndromes (Meckel-Gruber, Miller-Dieker)
 - Chromosomal anomalies (trisomy 21, triploidy)

INTRACRANIAL CYSTIC ABNORMALITIES

Hydrocephalus

- Sonographic findings include:
 - Normal ventricular configuration just dilated



- Dangling choroid plexus
- Ventriculomegaly (>10mm axial measurement)
- Brain echogenicity may be dense
- Associated findings concomitant with other anomalies present
 - Polyhydramnios
 - Abnormal fetal lie
 - Other intracranial anomalies



If the atrium of the lateral ventricle and the cisterna magna both measure ≤ 10mm, there is a 95% negative predictive value for the presence of ANY central nervous system anomaly.



Ventriculomegaly



Dangling choroid plexus

INTRACRANIAL CYSTIC ABNORMALITIES

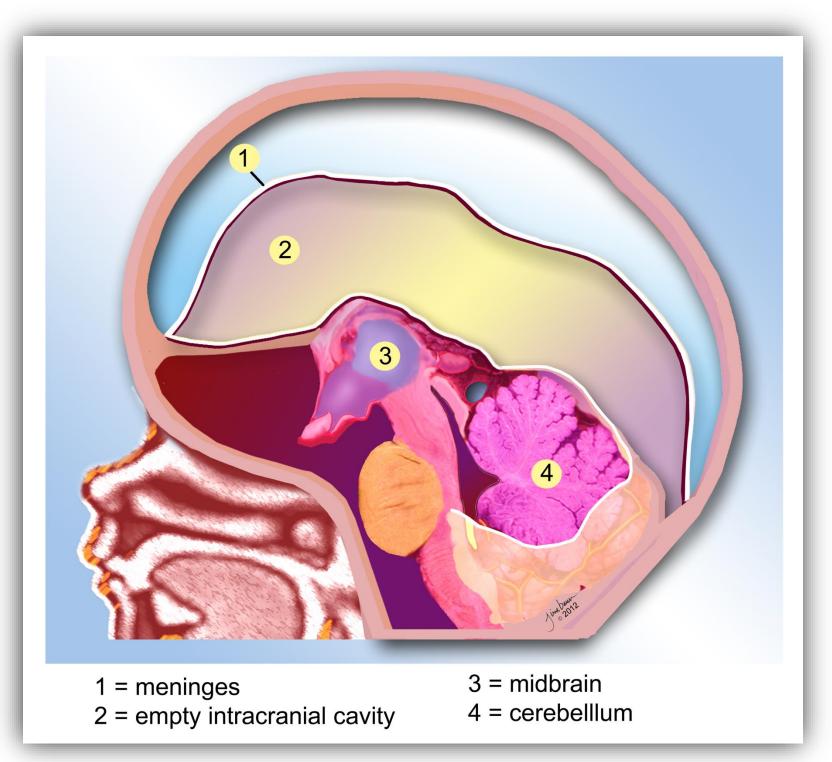
Hydranencephaly

• "Water, no brain"



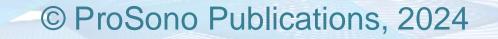
- Total or near total absence of cerebral hemispheres
- Meninges and skull are normal
- Unknown etiology
- May be result of infarction secondary to bilateral ICA occlusions or primary agenesis of neural wall

HYDRANENCEPHALY



Hydranencephaly

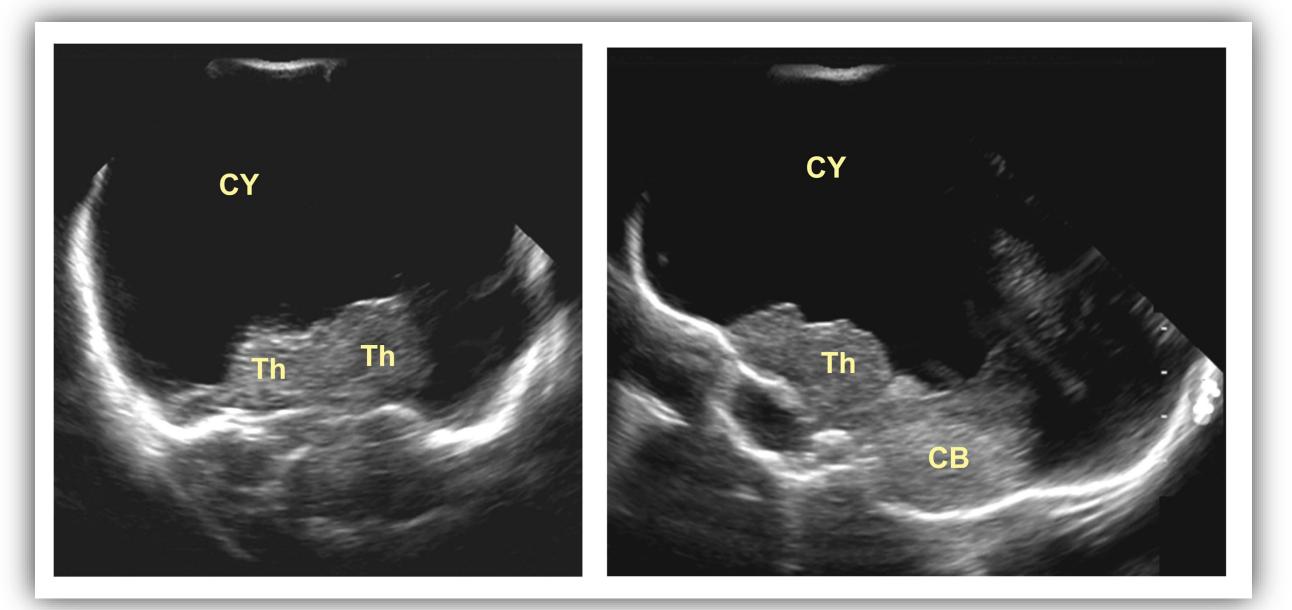
- Associated abnormalities:
 - Consequential arthrogryposis
 - Renal aplastic aplasia
 - Polyvalvular heart disease
 - Fowler syndrome
 - Trisomy 13
 - Polyhydramnios



Hydranencephaly

- Sonographic findings include:
 - Macrocephaly
 - Large anechoic area in cranial vault surrounding midbrain and basal ganglia (*water*)
 - Absent cortical mantle (no brain)
 - Tentorium separating normal posterior fossa from anterior and middle cranial fossae
 - Polyhydramnios
 - Occasionally small portions of occipital lobe present

HYDRANENCEPHALY



Coronal section

Sagittal section

CY = cystic filling Th = thalamus CB = cerebellum

Holoprosencephaly

- Abnormality of forebrain diverticulation resulting in the presence of a single midline ventricle
- Varying degrees of severity:
 - Alobar: most severe form with minimal amount of brain tissue present
 - Semilobar: less severe form with some normal brain structures present
 - Lobar: least severe form with mostly cerebral hemispheres

Holoprosencephaly

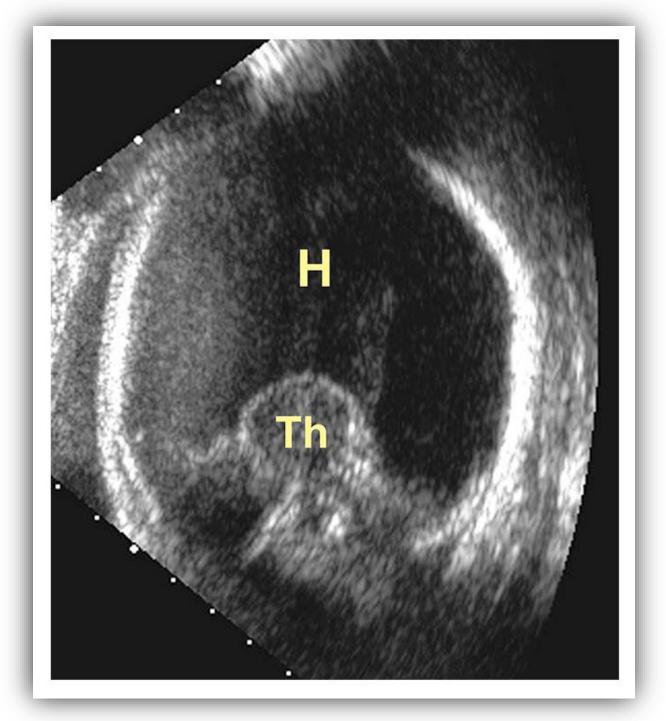
- Associated abnormalities include:
 - Trisomy 13, 18
 - Congenital renal anomalies
 - Congenital cardiac anomalies
- Associate facial anomalies include:
 - Proboscis
 - Cyclopia
 - Ethmocephaly
 - Cebocephaly

Holoprosencephaly

- Sonographic findings include:
 - Presence of a single, midline ventricle (*monoventricle*)

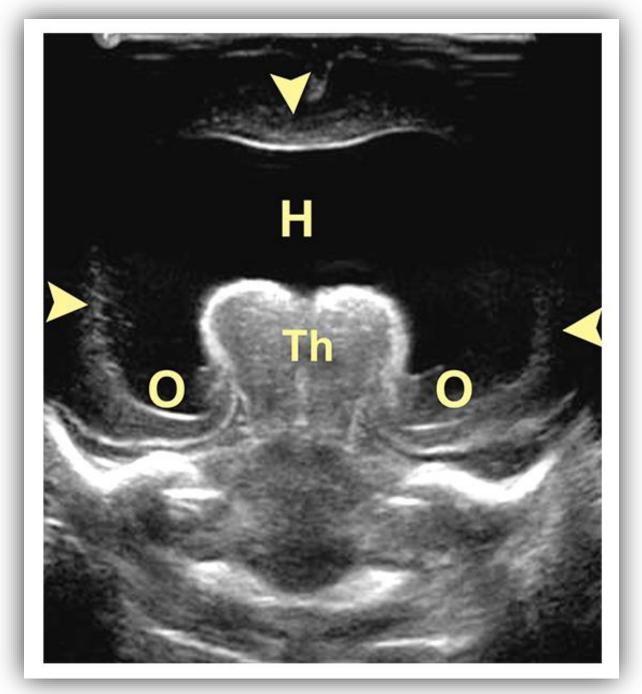


- Mantle of cerebral tissue present around monoventricle
- Varied appearance depending on degree of severity
- Associated craniofacial anomalies



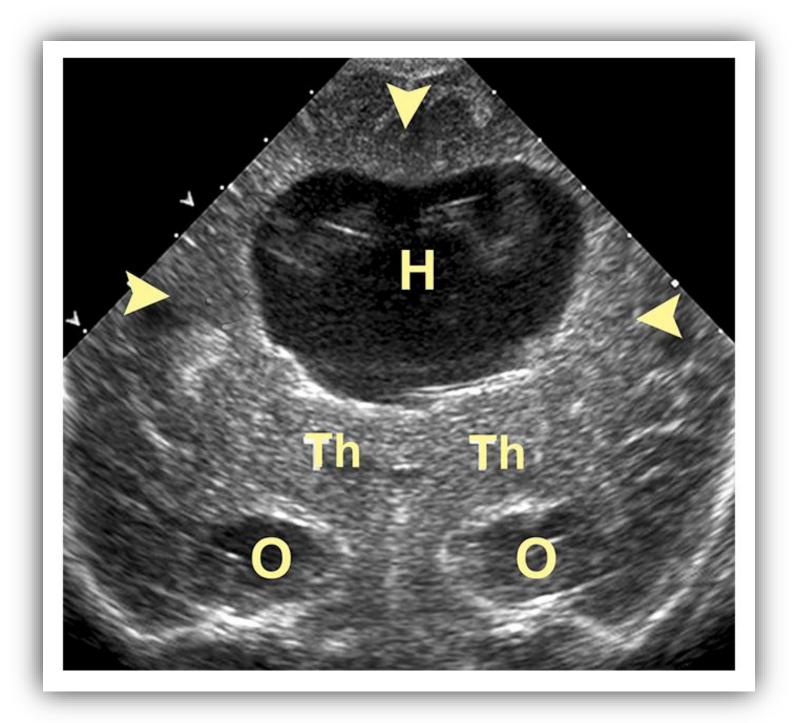
Alobar

H = holoventricle Th = thalamus



Semilobar

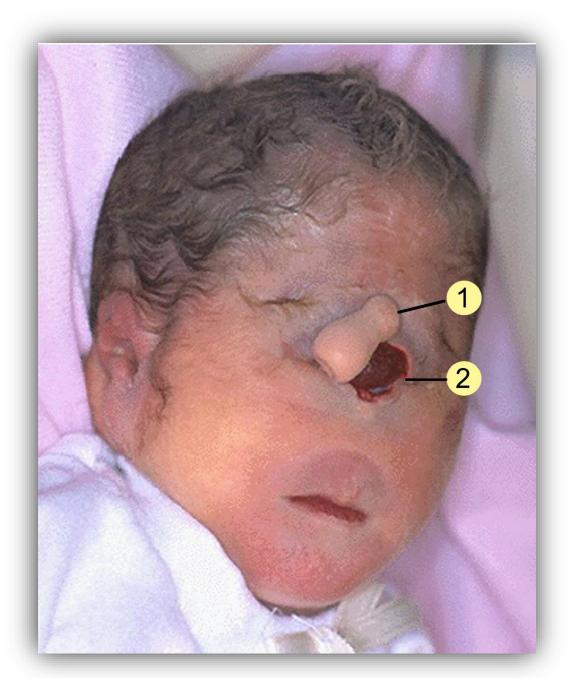
- H = holoventricle Th = thalamus
- O = occipital horns



Lobar

H = holoventricle Th = thalamus

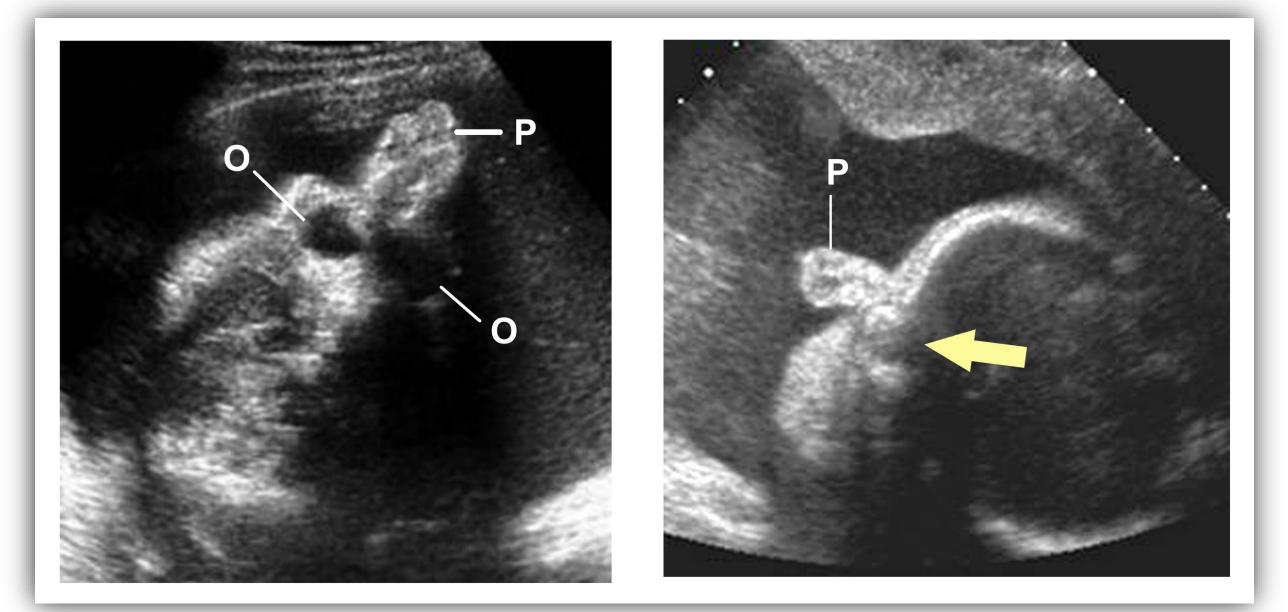
cle O = occipital horns arrowheads = cerebral cortex



1 = proboscis2 = cyclopia

Facial anomalies

Facial anomalies

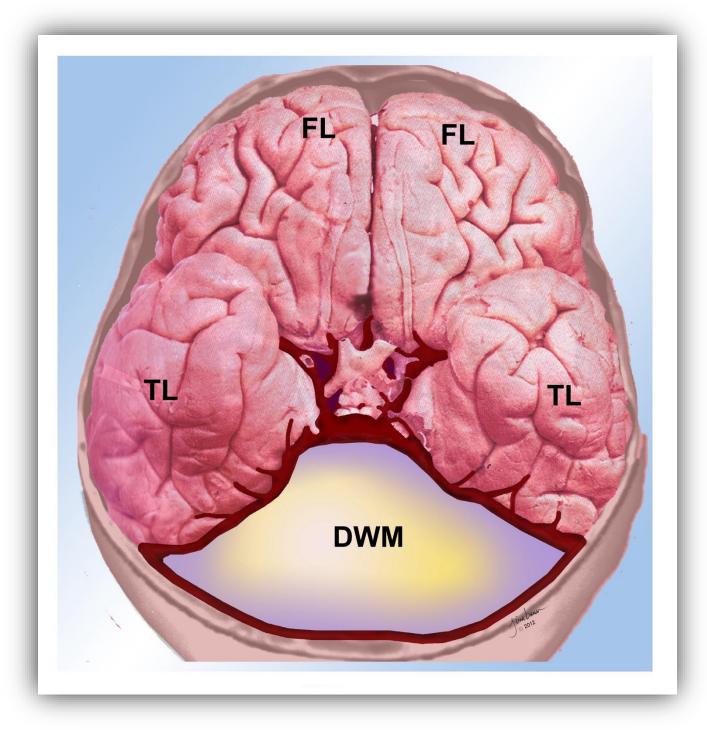


P = proboscis O = orbits ethmocephaly Arrow = cyclopia

Dandy-Walker Malformation

- Cystic abnormality of posterior fossa resulting from 4th ventricle outflow obstruction
- Characterized by:
 - Posterior fossa continuous with 4th ventricle
 - Posterior fossa enlargement
 - Cerebellar vermian dysgenesis

DANDY-WALKER MALFORMATION



FL = frontal lobe

TL = temporal lobe

DWM = Dandy-Walker malformation

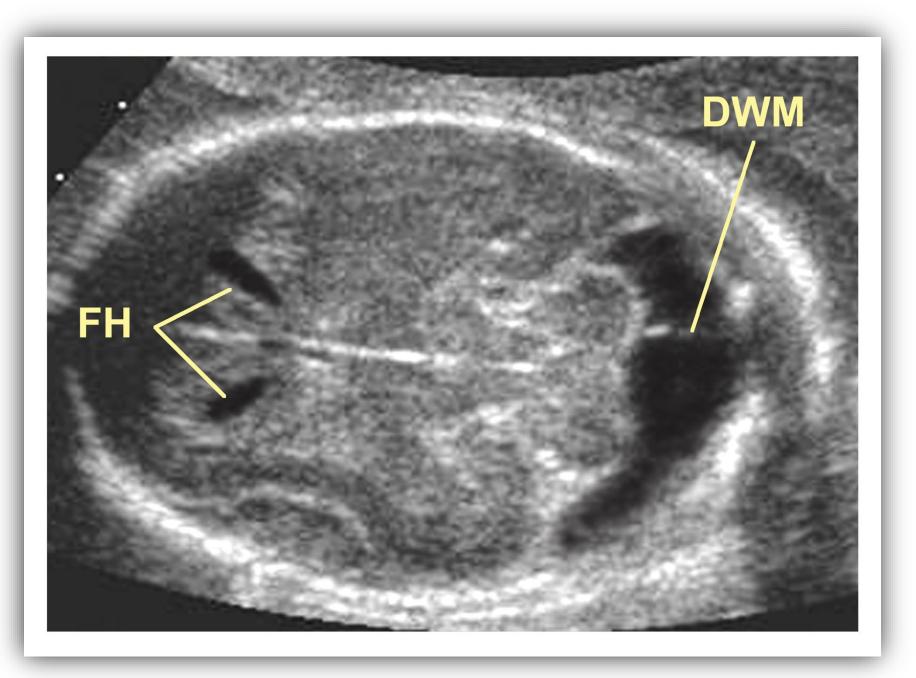
Dandy-Walker Malformation

- Associated abnormalities include:
 - Hydrocephalus (25%)
 - Other CNS abnormalities
 - Cardiac anomalies
 - Cleft palate
 - Meckel-Gruber syndrome

Dandy-Walker Malformation

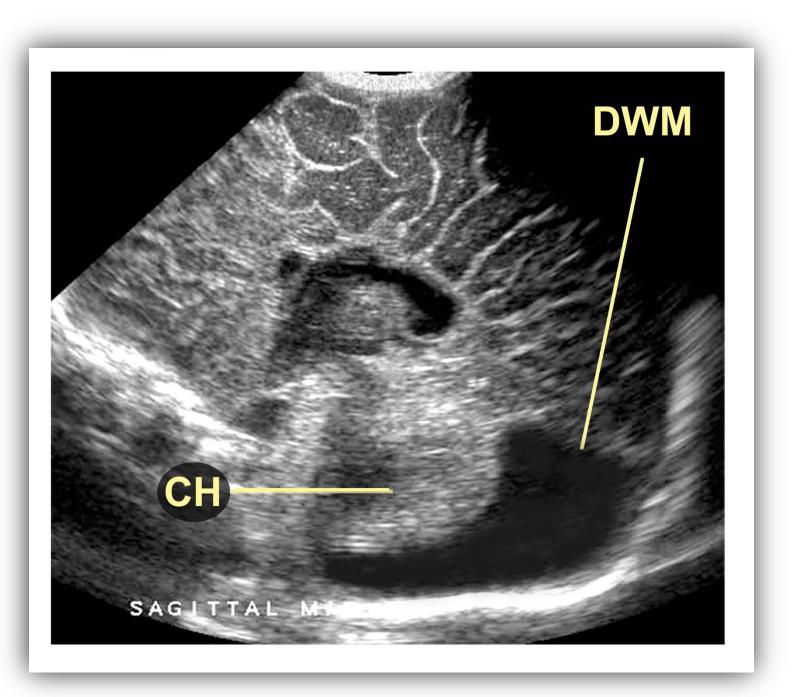
- Sonographic findings include:
 - Cystic enlargement of posterior fossa
 - Cerebellar hemispheres may be separated and flattened
 - Concomitant hydrocephalus (25%)
 - Contiguity with 4th ventricle (differentiates from arachnoid cyst)

DANDY-WALKER MALFORMATION



FH = frontal horns DWM = Dandy-Walker malformation

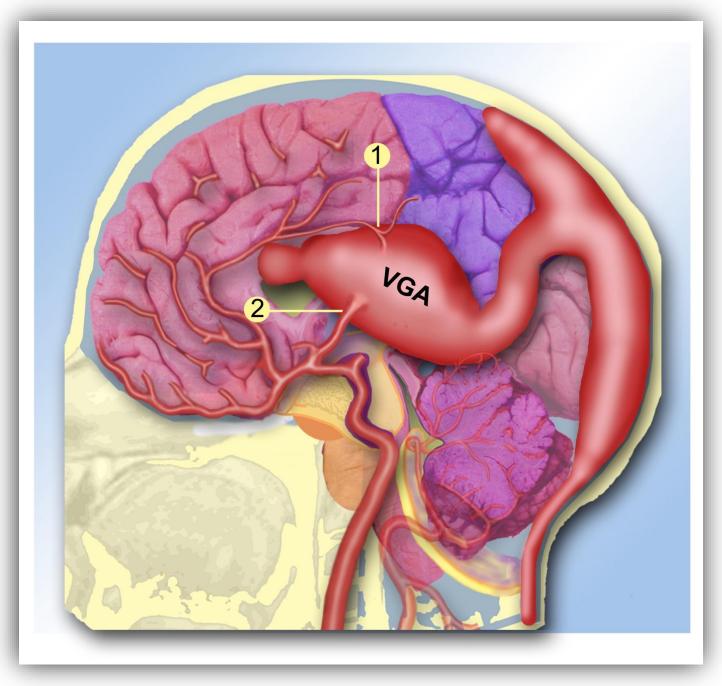
DANDY-WALKER MALFORMATION



CH = cerebellar hemisphere DWM = Dandy-Walker malformation

Vein of Galen Aneurysm

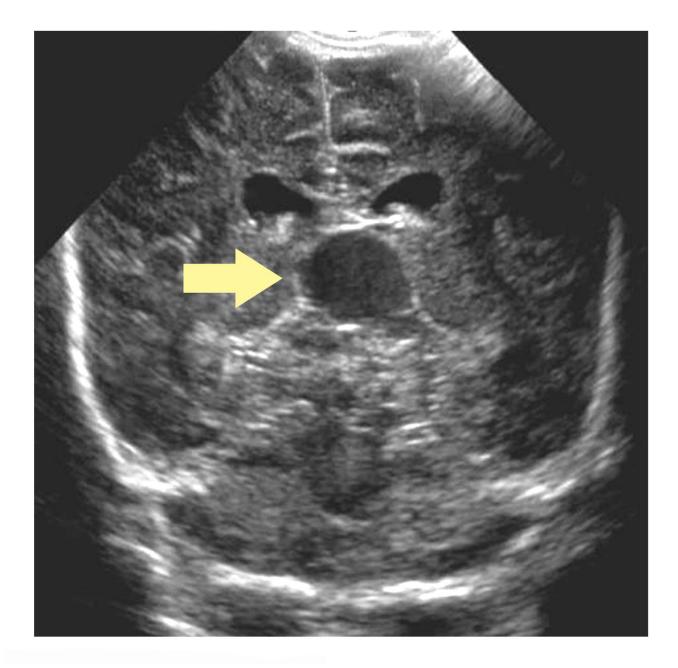
- Anomalous anastomoses between high-pressure branches of cerebral arteries and low-pressure prosencephalic vein
- Concomitant outflow obstruction of vein creates a dilated, midline vascular mass
- Associated abnormalities include:
 - High-output cardiac failure
 - Hydrocephalus



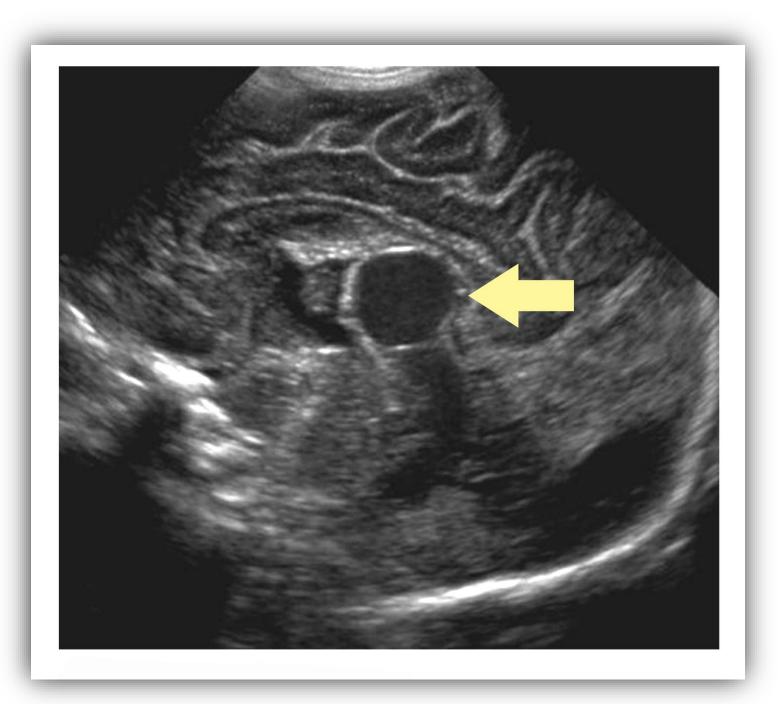
- **1.** = superior cerebral artery
- 2 = middle cerebral artery
- VGA = vein of Galen aneurysm

Vein of Galen Aneurysm

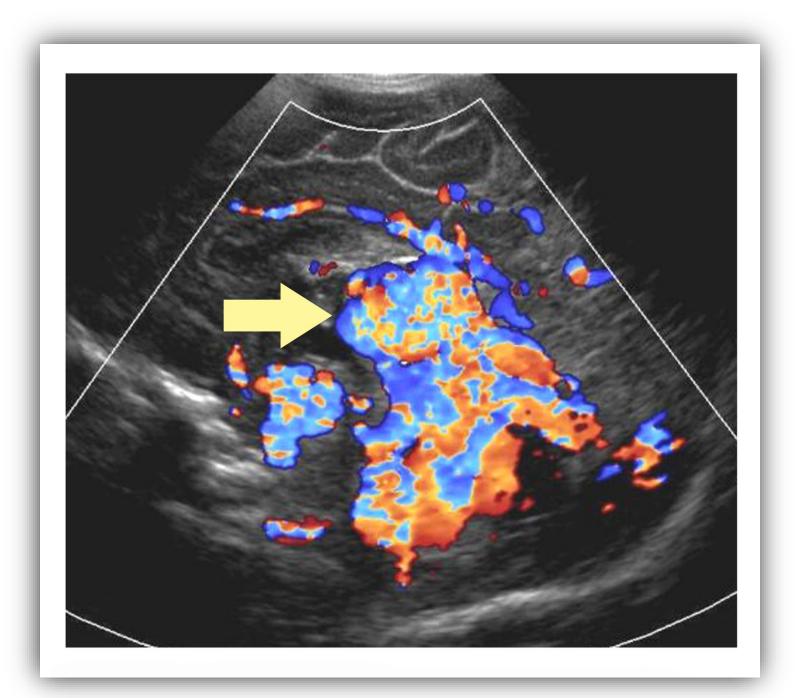
- Sonographic findings include:
 - Dilated cystic structure in the posterior midline
 - High-amplitude, arteriovenous hemodynamic patterns with color Doppler imaging



Coronal section Arrow = vein of Galen aneurysm



Sagittal section Arrow = vein of Galen aneurysm



Color Doppler imaging Arrow = vein of Galen aneurysm

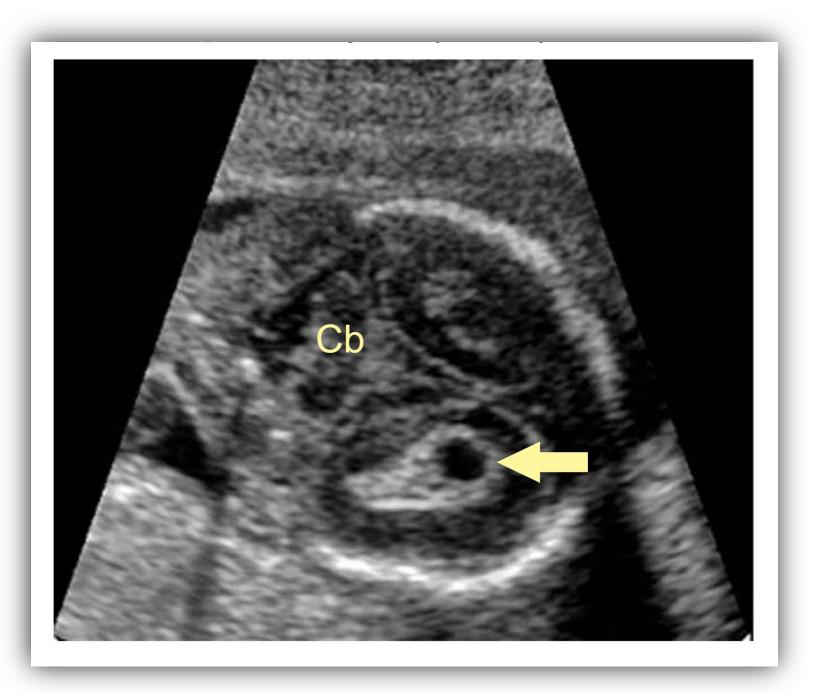
Choroid Plexus Cysts

- Benign fluid collection caused by an abnormal folding in the choroid epithelium
- CSF, produced by the choroid, becomes entrapped in this pouch
- Observed in 1–2% of mid-trimester fetuses
- Carry virtually NO clinical significance. They do not predict fetal anomalies

Choroid Plexus Cysts

- Sonographic findings include:
 - Small, well-circumscribed, anechoic structure within the echogenic choroid plexus
 - Maybe be unilateral or bilateral

CHOROID PLEXUS CYSTS



Cb. = cerebellum Arrow = choroid plexus cyst

CNS ABNORMALITIES

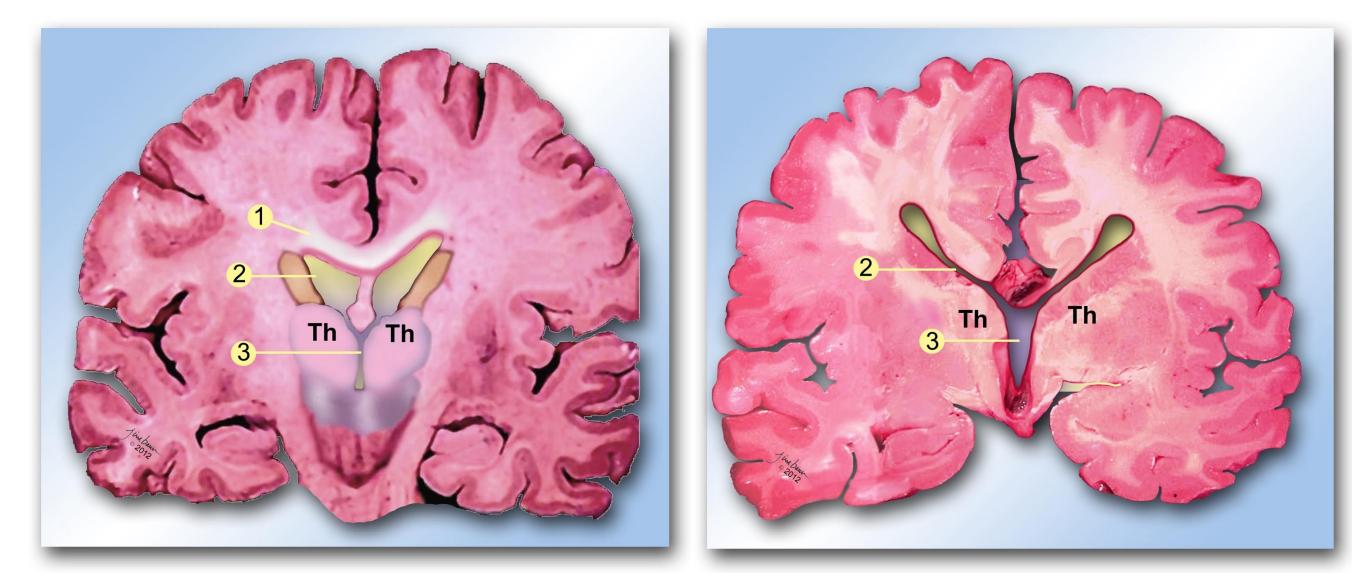
Intracranial Solid Abnormalities

- Agenesis of the corpus callosum
- Intracranial tumors
- Intracranial hemorrhage *in utero*
- Porencephaly (can also be cystic)
- Schizencephaly
- Intracranial calcifications
- Microencephaly
- Megaloencephaly

Agenesis of the Corpus Callosum

- Corpus callosum is midline anatomical structure connecting cerebral hemispheres
- Serves as roof of lateral ventricles
- Absence of corpus callosum:
 - Male predilection 2:1
 - Fetal alcohol syndrome has been implicated as etiological factor
 - Numerous associated CNS abnormalities

AGENESIS OF THE CORPUS CALLPOSUM



Normal

Absent corpus callosum

1 = corpus callosum 2 = lateral ventricles

3 = third ventricle Th = thalamus

Agenesis of the Corpus Callosum

- Associated abnormalities include:
 - Aneuploidy: trisomies 13, 18
 - Other syndromes are many including: fetal alcohol syndrome, Alpert syndrome, Zellweger syndrome
 - CNS anomalies include:
 - Chiari II malformation
 - Dandy-Walker spectrum
 - Holoprosencephaly
 - Hydrocephalus
 - Porencephaly

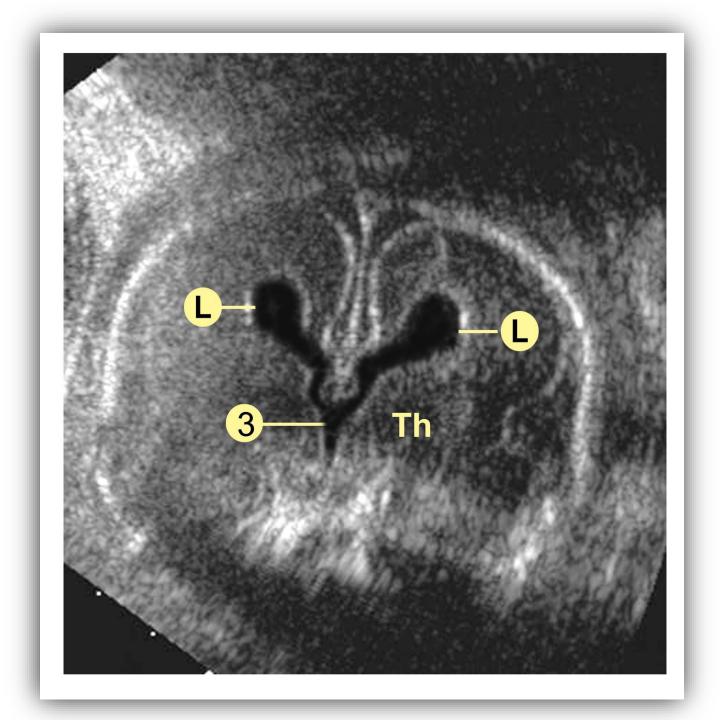
Agenesis of the Corpus Callosum

- Sonographic findings vary based on severity but may include:
 - Absent cavum septi pellucidi (in complete)
 - Lateral ventricles displace upward and outward "bat wing sign"



- Third ventricle dilated and displaced superiorly
- Posterior horn hydrocephalus

AGENESIS OF THE CORPUS CALLOSUM



Coronal section

- L = lateral ventricle
- Th = thalamus
- 3 = third ventricle

Intracranial Tumors

- Rare occurrence.
- May be cystic, solids, or complex
- Most are supratentorial (above posterior fossa)
- Most common type is teratoma
- Other types include:
 - Glioblastomas
 - Craniopharyngiomas
 - Sarcomas

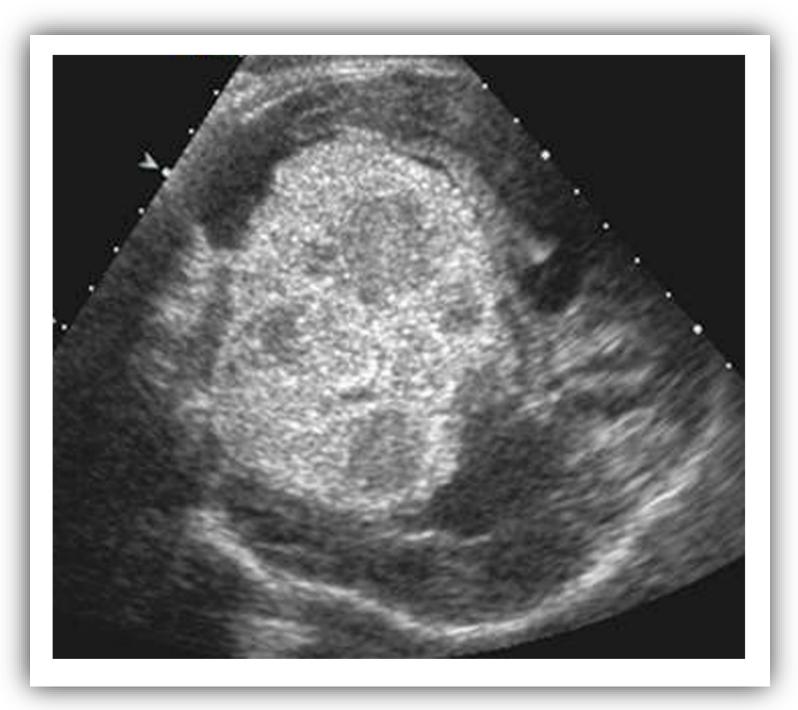
Intracranial Tumors

- Associated abnormalities include:
 - Large, complex, echogenic mas in fetal brain
 - Gross distortion of normal intracranial architecture
 - Hydrocephalus
 - Macrocephaly
 - Polyhydramnios

Intracranial Tumors

- Sonographic findings include:
 - High-output cardiac failure
 - Hydrops fetalis
 - Abnormal cerebral Doppler flow velocimetry

INTRACRANIAL TUMORS



Teratoma

INTRACRANIAL TUMORS



Glioblastoma

Intracranial Hemorrhage in utero

- Rare *in utero* event.
- Results from cerebral hypoxic events
- Etiologies include:
 - Severe fetal or maternal hypoxia
 - Maternal abdominal trauma
 - Fetal or maternal thrombocytopenia
 - Twin-to-twin transfusion with demise of co-twin

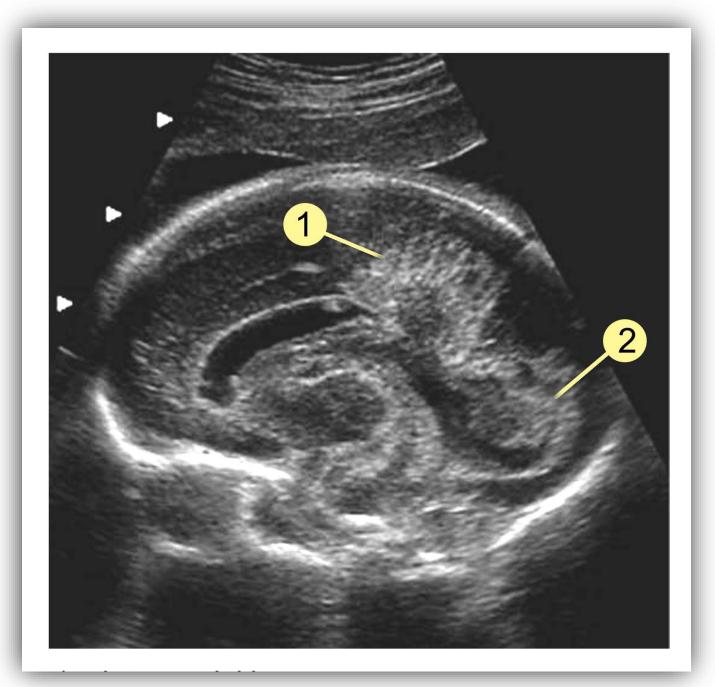
Intracranial Hemorrhage in utero

- Sonographic findings include:
 - Irregularly marginated, heterogeneous echogenic structures within the cerebral parenchyma

Porencephaly

- Focal cystic area of encephalomalacia (softening of brain tissue) that communicates with a ventricle, subarachnoid space, or both
- When found in association with intracranial hemorrhage, refers to cystic lesions created by lysis of ischemic areas of tissue

INTRACRANIAL HEMORRHAGE *IN UTERO*



1 = intracranial hemorrhage
2 = porencephaly

Schizencephaly

- Rare condition characterized by large gray matter clefts extending through cerebral parenchyma from meninges to lateral ventricles
- Etiologies include:
 - Abnormal neuronal migration during embryological development
 - In utero vascular insults resulting in encephalomalacia
- Associated abnormalities include:
 - Absent cavum septi pellucidi
 - Hydrocephalus

Schizencephaly

- Sonographic findings include:
 - Unilateral or bilateral cystic defect extending from brain surface to ventricular wall
 - Absence of the cavum septi pellucidi
 - Hydrocephalus

SCHIZENCEPHALY



Coronal section

Intracranial Calcifications

- May be indicator of several types of pathology
- Possibilities include:
 - Congenital infection (CMV, toxoplasmosis) MOST COMMON



- Calcifications within a tumor
- Sturge-Weber syndrome

Intracranial Calcifications

- Sonographic findings include:
 - Brightly echogenic foci within the parenchyma of the fetal brain

INTRACRANIAL CALCIFICATIONS



Sagittal section

Microencephaly

- Condition characterized by abnormally small and underdeveloped brain and skull
- Defined as head measure > 3 standard deviations below mean for gestation age
- Etiologies include:
 - Craniosynostisis
 - Holoprosencephaly
 - Multiple genetic syndromes and malformations
 - In utero exposure to drugs, infections and environmental teratogens

Microencephaly

- Sonographic findings include:
 - Small biometric head measurements relative to femur length and abdominal circumference
 - Head circumference > 3 standard deviations below mean for gestational age
 - Abnormal intracranial architecture
 - Severe cases differentiated from anencephaly by presence of cerebral parenchyma

MICROENCEPHALY



Sonographic appearance

Gross pathology

Megaloencephaly

- Condition characterized by abnormally large and heavy brain
- Exceed mean volume by > 2 standard deviations expected for gestation age
- Associated with a number of syndromes
- Postnatal complications include: autism, mental retardation, epilepsy, motor impairment

Megaloencephaly

- Sonographic findings include:
 - Abnormally large head circumference for dates
 - HC/AC ratio above 99th percentile

Neural Tube Defects

- A spectrum of CNS anatomic abnormalities resulting from failure of closure of the neural tube (*neurulation*)
- Location of defect along neural axis determines type of anomaly that will manifest
 - Cranial portion: head anomalies
 - Caudal portion: spine anomalies

CNS ABNORMALITIES

Neural Tube Defects

- Anenecephaly
- Exencephaly (acrania)
- Cephalocele/encephalocele
- Iniencephaly
- Spina bifida

Anencephaly

- Most common NTD results from failure of closure of caudal neuropore
- Lethal anomaly (not compatible with life)
- Occurs 1 : 1,000 live births and carries a 4% chance of recurrence
- More common in female fetuses

Anencephaly

- Pathologically characterized by:
 - Complete absence of cranial vault and cerebral hemispheres
 - Portions of midbrain and brain stem remain present
 - Typical facies include:
 - Macrophthalmia (bulging eyes)
 - Macroglossia (thick tongue)
 - Dramatically shortened neck

Anencephaly

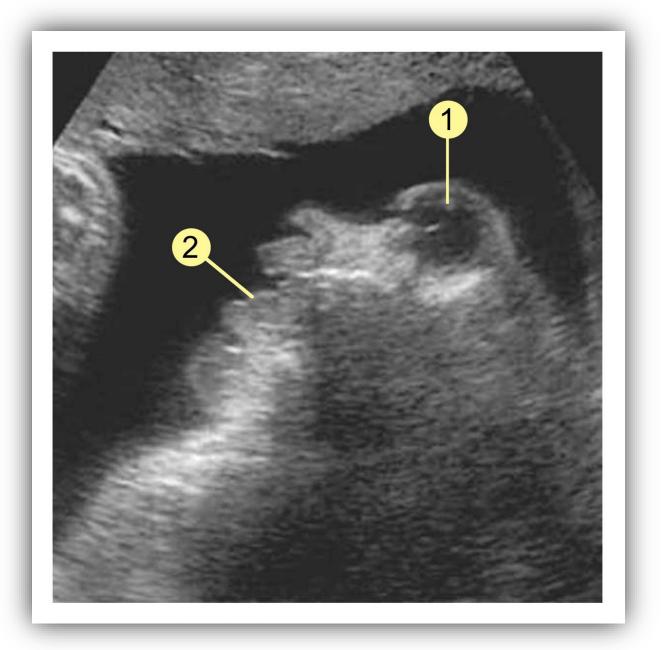
- Sonographic findings include:
 - Unidentifiable fetal head by 15 weeks (definitive)
 - Major portions of cranium and intracranial structures are absent
 - Parts of midbrain and brain stem may be present
 - Typical anencephalic facies (as described above)
 - Polyhydramnios (40 50% of cases)

ANENCEPHALY



Coronal section – 14 weeks

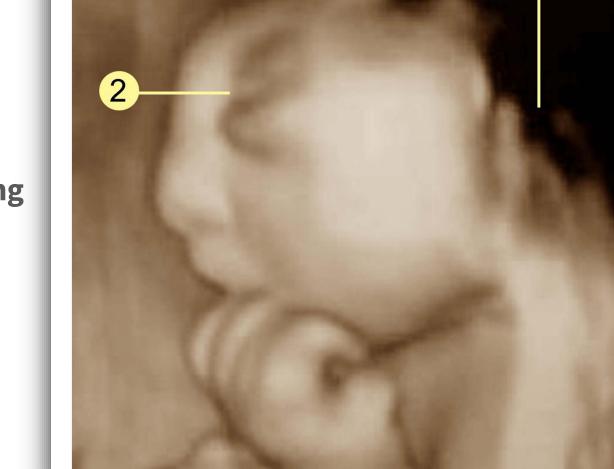
ANENCEPHALY



1 = macrophthalmia 2 = macroglossia

Sagittal section typical facies

ANENCEPHALY



1

1 = absent cranium 2 = macrophthalmia

3D surface rendering

Anencephaly

- Associated anomalies include:
 - Spina bifida (cervical)
 - Cleft lip and palate
 - Clubfoot (talipes equinovarus)
 - Congenital heart defects
 - Hydronephrosis
 - Polyhydramnios

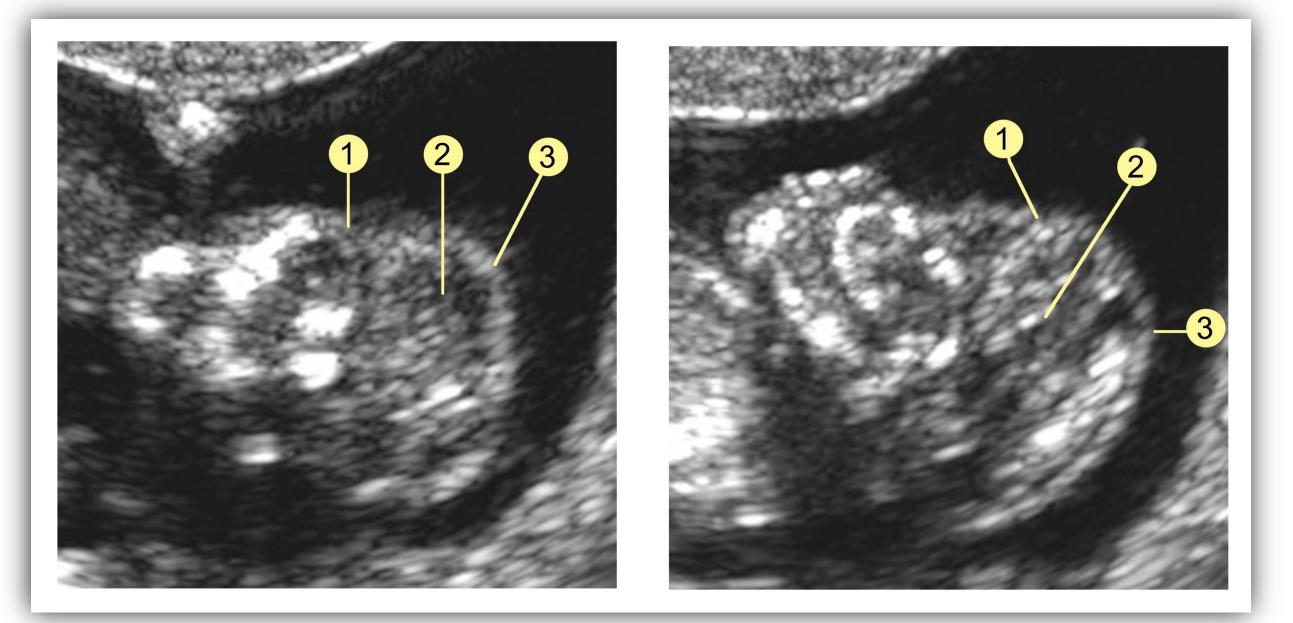
Exencephaly (Acrania)

- Rare congenital anomaly characterized by absence of calvaria (skull bones) with complete but abnormal development of brain tissue
- Cerebellum, brain stem, and cranial nerves are present and normal
- Associated abnormalities include:
 - Cleft lip and palate
 - Club foot (talipes equinovarus)

Exencephaly (Acrania)

- Sonographic findings include:
 - Absence of cranial vault
 - Presence of cerebral hemispheres
 - Thin membrane covering cerebral hemispheres

EXENCEPHALY (ACRANIA)



Coronal section

Sagittal section

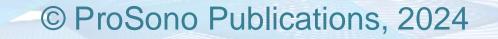
- 1 = absent cranial vault
- 2 = cerebral hemispheres
- 3 = covering membrane

Cephalocele/Encephalocele

- Focal defect in bony calvarium through which meninges and/or brain herniate
- Terminology:
 - Cephalocele herniation of meninges only
 - Encephalocele herniation of brain and meninges
- Most commonly located in midline occipital region above the tentorium
- Vary in size and severity

Cephalocele/Encephalocele

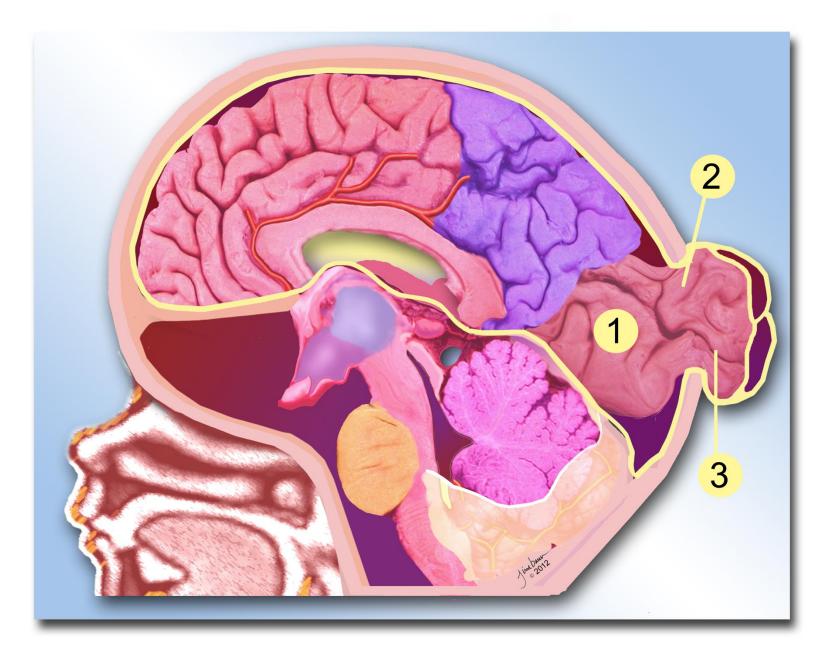
- Associated abnormalities include:
 - Agenesis of corpus callosum
 - Orofacial clefting
 - Dandy-Walker malformation
 - Arnold-Chiari malformation
 - Hydrocephalus
 - Spina bifida
 - Many others



Cephalocele/Encephalocele

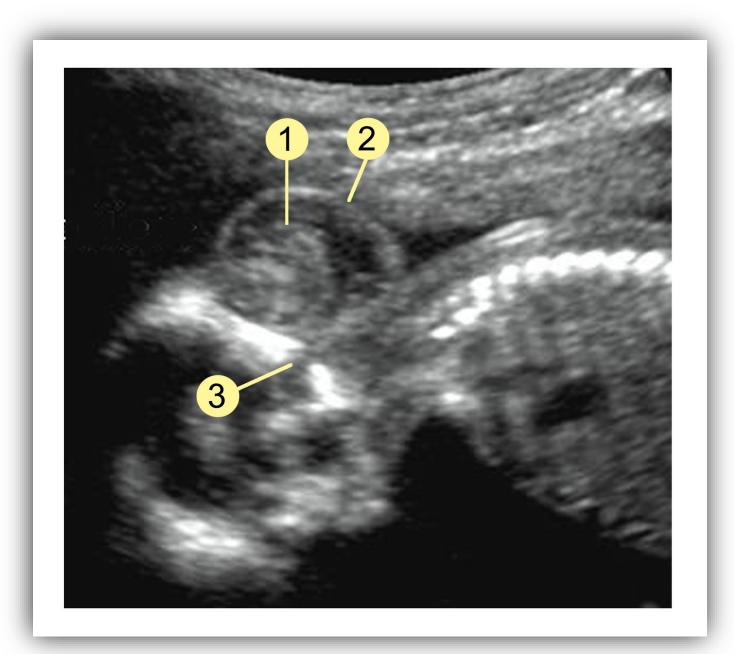
- Sonographic findings include:
 - Mass extending from the calvaria
 - May be totally cystic (cephalocele)
 - May be cystic with septations (meningocele)
 - May contain brain tissue (encephalomeningomyelocele)
 - Cranial defect (occasionally seen)
 - Cranial cavity appears small if significant portion of brain is herniated
 - Associated hydrocephalus and/or polyhydramnios

CEPHALOCELE/ENCEPHALOCELE



1 = occipital lobe
2 = calvarial defect
3 = meningeal sac

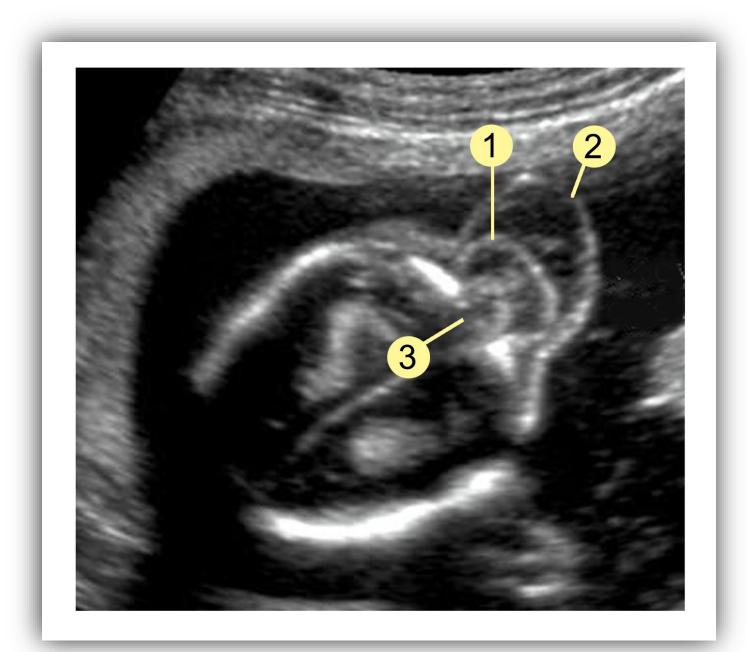
CEPHALOCELE/ENCEPHALOCELE



Sagittal section

1 = occipital lobe 2 = meningeal sac 3 = calvarial defect

CEPHALOCELE/ENCEPHALOCELE



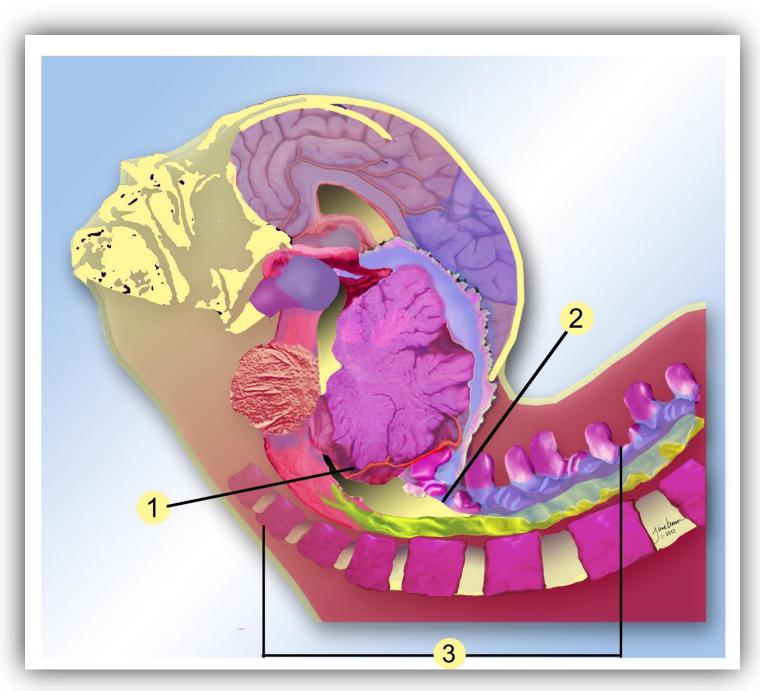
Axial section

1 = occipital lobe 2 = meningeal sac 3 = calvarial defect

Iniencephaly

- Rare, uniformly lethal, NTD of the cervical and upper dorsal spine characterized by:
 - Cervical rachischisis
 - Occipital encephalocele
 - Exaggerated spinal lordosis
- Anomalous features include:
 - Retroflexion of the upper spine
 - Short neck and trunk
 - Defects of the thoracic cage
 - Anterior spina bifida

INIENCEPHALY



- **1** = occipital encephalocele
- 2 = cervical rachischisis
- 3 = exaggerated cervicothoracic lordosis

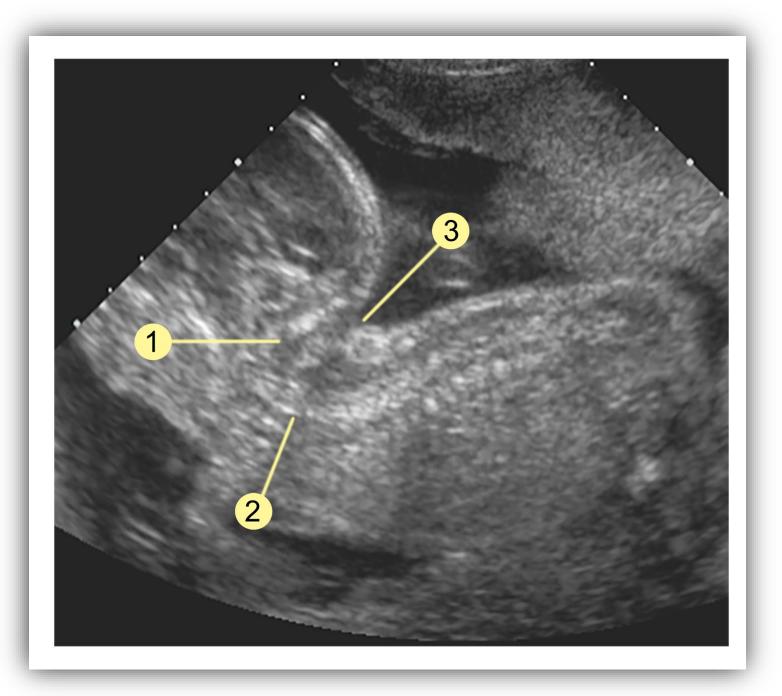
Iniencephaly

- Associated abnormalities include:
 - Anencephaly and variants
 - Myelomeningoceles
 - Diaphragmatic hernia
 - Pulmonary hypoplasia
 - Cardiac defects

Iniencephaly

- Sonographic findings include:
 - Cervicothoracic lordosis ("stargazer" posture)
- Occipital bone defects including encephalocele
- Short, fused cervical vertebrae
- Polyhydramnios
- Cervical rachischisis

INIENCEPHALY

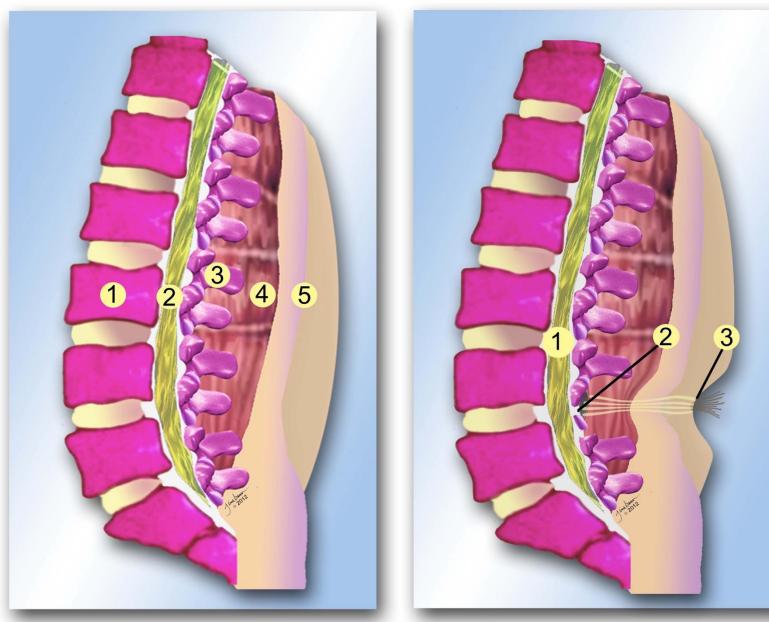


- 1 = occipital encephalocele
- 2 = cervical rachischisis
- 3 = exaggerated cervicothoracic lordosis

Spina Bifida

- NTD resulting from failure of rostral neural tube to close
- Also called *spinal dysraphism*
- Can occur anywhere along spinal column but most commonly located in the lumbosacral region
- Two types:
 - Spina bifida occulta: (*hidden*) defect in bony spinal column with intact overlying tissue and skin
 - Spina bifida aperta: (*visible*) defect in spinal column and overlying tissue and skin

SPINA BIFIDA OCCULTA



Normal spine.

- 1 = vertebral body
- 2 = spinal cord & meninges
- 3 = posterior vertebral elements
- 4 = musculature
- 5 = posterior fascia & skin

Spina bifida occulta.

- 1 = intact spinal cord
- 2 = localized defect
- 3 = skin dimple/tuft

- Gross pathological characteristics:
 - *Myelocele:* herniated sac of meninges alone
 - *Meningomyelocele*: herniated meninges and neural tissue
 - *Rachischisis*: severe form where entire spinal column is splayed open posteriorly from neck to sacrum

- Sonographic findings (transverse) include:
 - Posterior ossification centers splayed into a U ov V shape
 - Cystic or complex structure extending from tom back (when sac is intact)
 - Sac may appear as simple cystic structure, cystic with septations, or a complex/solid mass

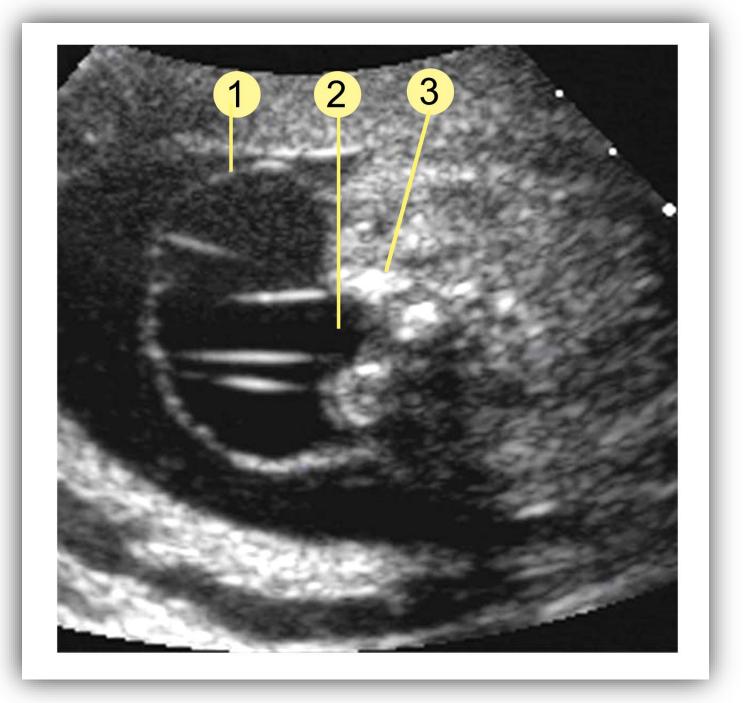
- Associated abnormalities include:
 - Trisomy 18 (Edwards syndrome)
 - Trisomy 13 (Patau syndrome)
 - Chiari II malformation
 - Hydrocephalus
 - Clubfoot (talipes equinovarus)
 - Rockerbottom foot (congenital vertical talus)





TRANSVERSE

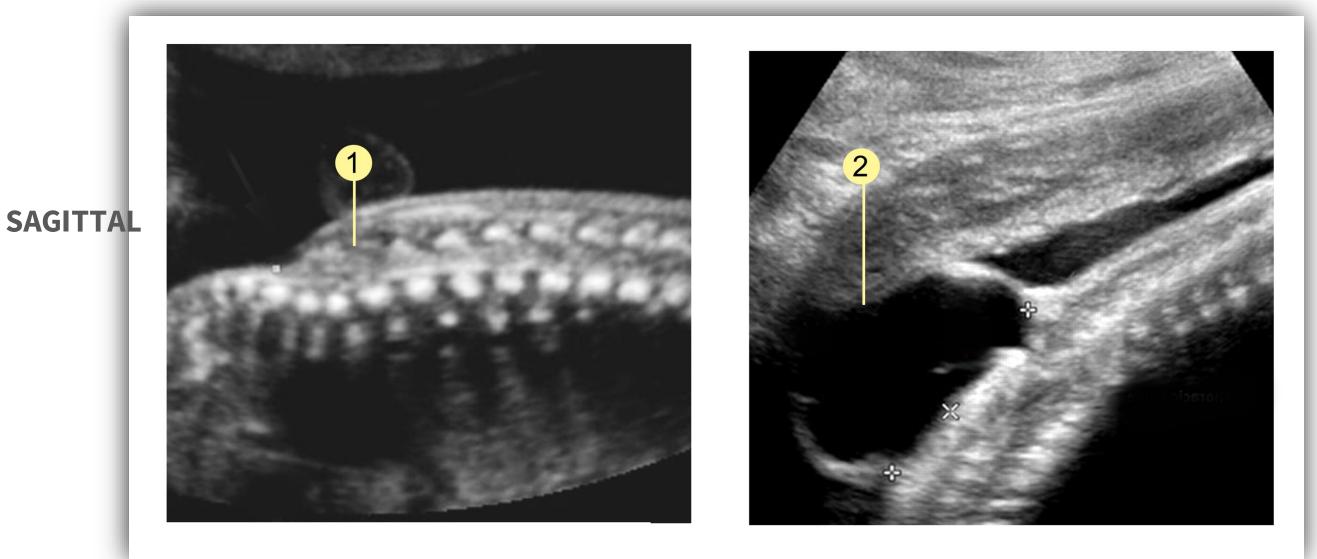
Arrows = splayed ossification centers



TRANSVERSE

- 1 = herniated sac
- 2 = open defect
- 3 = splayed ossification centers

- Sonographic findings (sagittal) include:
 - Discontinuity of parallel ossification centers
 - Herniated sac protruding posteriorly



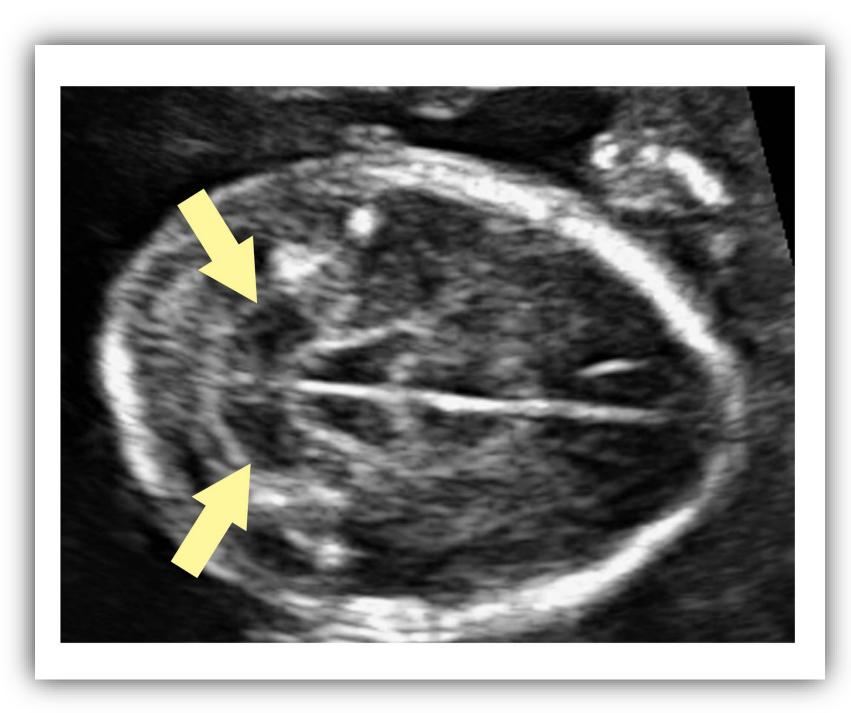
1 = discontinuity of ossification centers 2 = herniated sac

- Sonographic findings (intracranial) include:
 - Lemon sign: overlapping of frontal bones creating a lemonshaped fetal head
 - Banana sign: effacement of the cisterna magna due to downward displacement of the cerebellum
 - *Chiari II malformation*: bulging of brain stem and part of cerebellum though foramen magnum into upper cervical spine



Intracranial

Lemon sign



Intracranial

Banana sign



Intracranial

Chiari II malformation

OB GYN SONOGRAPHY REVIEW

The Fetal Central Nervous System

