Ultrasound Evaluation of the Central Nervous System

Mani Montazemi, RDMS
Director of Ultrasound Education & Quality Assurance
Baylor College of Medicine
Division of Maternal-Fetal Medicine
Department of Obstetrics and Gynecology
Texas Children's Hospital, Pavilion for Women
Houston Texas
&
Clinical Instructor
Thomas Jefferson University Hospital
Radiology Department
Philadelphia, Pennsylvania

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Ultrasound Evaluation of the Central Nervous System

- CNS malformations are the second most frequent category of congenital anomaly, after congenital heart disease
- Poor timing of the examination, rather than poor sensitivity, can be an important factor in failing to detect a CNS abnormality

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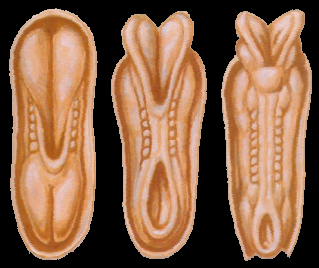
Central Nervous System

5th Menstrual Week

Arises from the posterior surface of the embryonic ectoderm

A small groove is found along the midline of the embryo and the edges of this groove fold over to form a neuro tube that gives rise to the fetal spinal cord and brain



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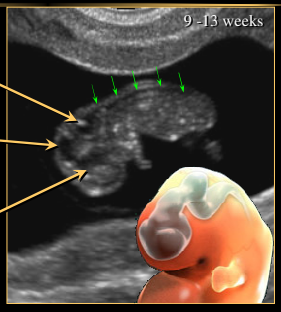
Brain Development

Rhombencephalon
- Gives rise to hindbrain
- 4th ventricle

Mesencephalon
- Gives rise to midbrain
- Aqueduct

Prosencephalon
- Gives rise to forebrain
- Lateral & 3rd ventricles



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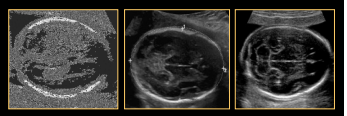
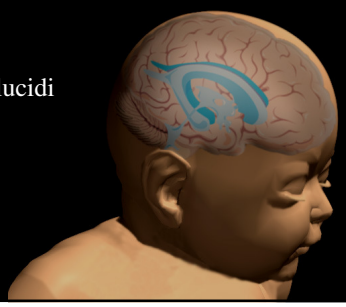
Ventricular view
- Lateral ventricles
- Choroid plexus



Thalamic view
- Midline falx
- Cavum septi pellucidi
- Thalami

Cerebellar view
- Cerebellum
- Cisterna magna



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Neural Tube Defects

Group of malformations:
- Anencephaly
- Cephaloceles
- Spina bifida

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Anencephaly

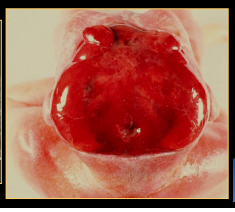
- Lethal abnormality
- Absence of the brain & lack of the cranial vault
  - No soft tissue above orbits
- The prevalence of anencephaly in the United States in 2001 was 9.40 per 100,000 live births

National Center for Health Statistics

Fetal Head

Anencephaly

- Defect is covered by a membrane known as cerebrovasculosa
- Often contiguous with cervical spine defect



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Anencephaly

- Fetal face from the orbits to the chin is usually normal
- Frontal bone is defective above the orbits
- Parietal bones & occipital bone are absent

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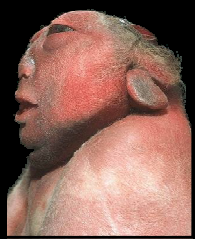
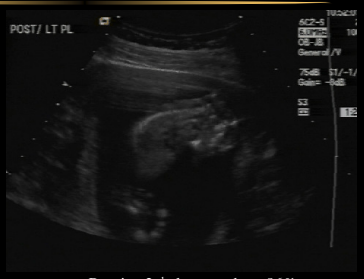
Anencephaly

- Bulging eyes
- Large tongue
- Very short neck



Fetal Head

Anencephaly



Routine 2nd ultrasound	96%
MSAFP	78%

Kooper et al, Prenat Diagn. 2007 Jan;27(1):29-33

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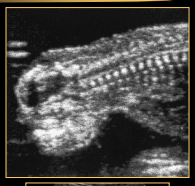
Anencephaly

- Elevated MSAFP
  - Leakage thru open neural tube
- Polyhydramnios after 25 wks
  - Failure to swallow
  - Excessive micturition
  - Failure of reabsorption of CSF

Preterm labor and delivery



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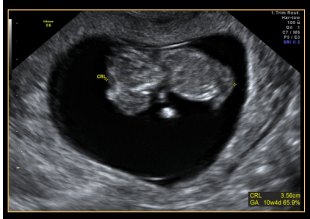
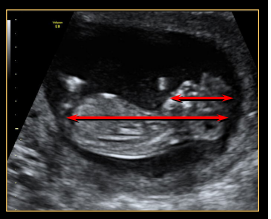
Early Recognition of Anencephaly





"crown-chin to crown-rump length ratio"

Sepulveda W, Sebire NJ, Fung TY, Pipi E, Nicolaides KH Am J Obstet Gynecol. 1997, 176(4):852

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Anencephaly

- More common
  - in girls than boys
  - in whites than blacks
  - in Irish compared to other ethnic groups
  - in mothers at the younger and older extremes of age
- Associated malformations
  - Spina bifida
  - Cleft lip or palate
  - Clubfoot

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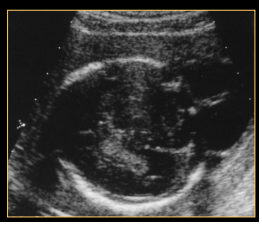
Cephalocele

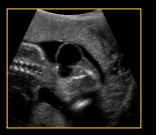
- Protrusion of intracranial contents through a bony defect of the skull
- Size - variable



Fetal Head

Cephalocele

- Herniated
  - Brain tissue & CSF – 85%
    - Encephalocele
  - Meninges & CSF – 15%
    - Cranial meningocele



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Cephalocele

- 75% Occipital
  - Western hemisphere
- 13% Frontal
  - Asian population
- 12% Parietal

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Cephalocele

- Other CNS anomalies common
  - Absent cavum septi pellucidi
  - Anomalous corpus callosum
  - Dorsal interhemispheric cysts
  - Chiari malformations
  - Dandy Walker malformation
  - Cerebellar cortical dysplasia

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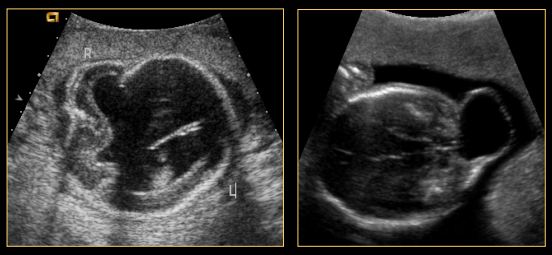
Cephalocele



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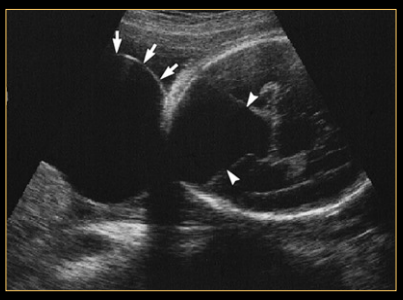
Occipital meningocele with an associated Dandy-Walker cyst



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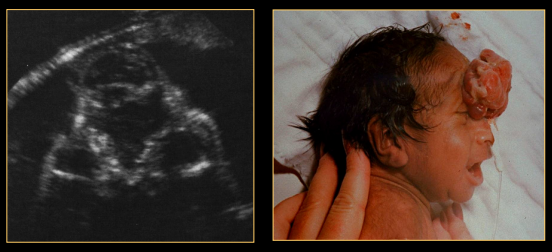
Cephalocele



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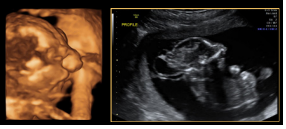
Cephalocele

Usually occurs as an isolated lesion, but may be a part of a syndrome

- Meckel-Gruber syndrome
  - Cepahalocele + B/L polycystic Kid + post-axial polydactyly
- Walker-Warburg syndrome
  - Lissencephaly + cerebellar hypoplasia + Dandy-Walker cyst + occular abnormalities
- Amniotic band Syndrome
  - Can involve any part of skull



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Structural Midline Defects

- High incidence of associated anomalies
  - Structural & chromosomal
- Karyotyping should be offered

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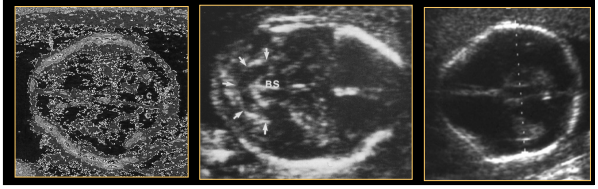
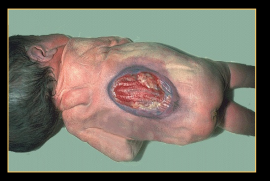
NTD – Cranial Signs

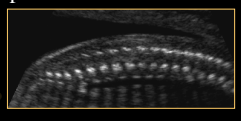
- Lemon sign: head deformity
- Banana sign: cerebellar deformity
- Ventriculomegaly

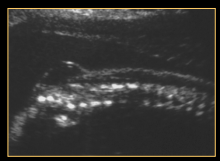


Spina Bifida or Spinal Dysraphism

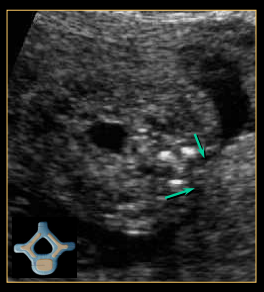
- A defect that can occur anywhere along the spinal axis
- Lower spine more common than higher
- Spinal cord and nerve roots exposed

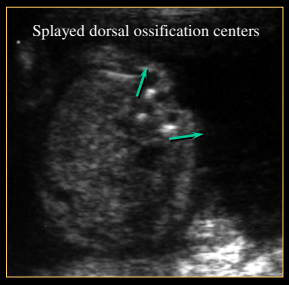






Fetal Head





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Remember

- Ossification occurs from cervical to coccygeal
  - Lower levels ossify later
  - Lumbosacral area appears open & wider

Higher level

Lower level

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Spina Bifida

There are several forms of spina bifida

- Ventral defects (rare)
  - Splitting of the vertebral body
  - Occurrence of a cyst that is neuroenteric in origin
- Dorsal defects (most common)
  - Occulta (closed spina bifida)
  - Aperta (open spina bifida)

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Closed Spina Bifida

- Spina bifida occulta (15%)
  - Small defect completely covered by skin
  - Asymptomatic/incidental findings

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Skin-Covered NTD

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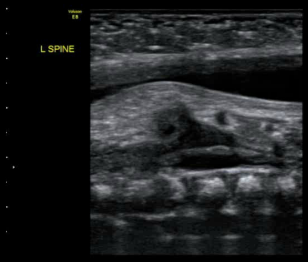
Skin-Covered NTD



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NTD

- Lesion level is most predictive of ambulatory function
  - Higher lesion levels associated with disphagia
  - The absence of covering membrane associated with scoliosis

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Open Spina Bifida

- Spina bifida aperta (85%)
  - Neural canal may be exposed
  - Defect may be covered by a thin meningeal membrane

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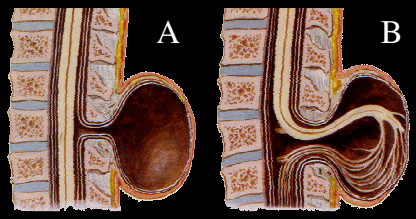
Classifications of Spina Bifida



A) Meningocele – If the tumor contains purely meninges & CSF

B) Myelomeningocele – protrusion of a sac containing CSF & neural elements

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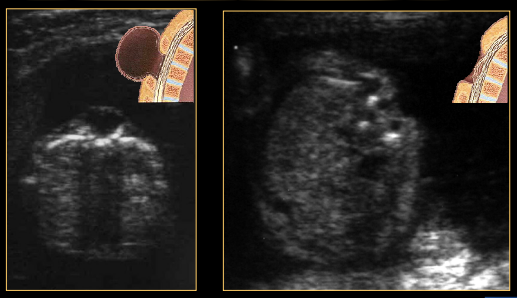
Meningocele Vs. Myeloschisis



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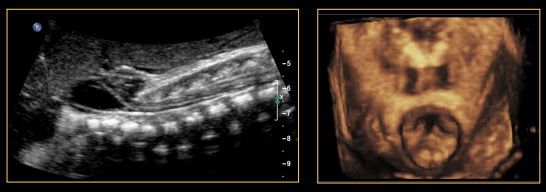
Myelomeningocele



It contains solid component due to presence of neural elements within the herniated sac

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Reversal of Chiari Malformation

"Healed" back at birth

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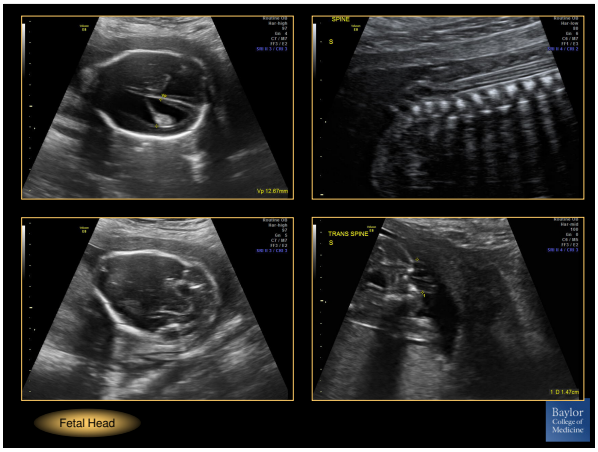

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Cerebellar View - Cisterna Magna

- Size: 2-10 mm
  - < 2 mm spina bifida (ACII)
  - > 10 mm Dandy-Walker

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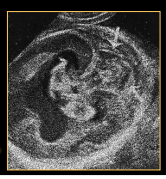
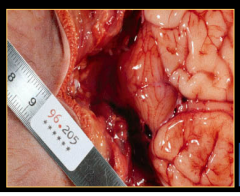
Arnold-Chiari II

- Herniation of the cerebellar vermis through foramen magnum
- Fourth ventricle is displaced downward inside the neural canal





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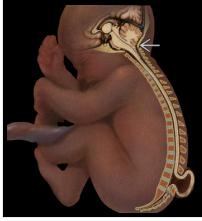
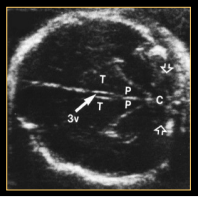
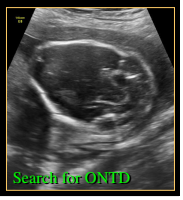
Spina Bifida - Arnold-Chiari

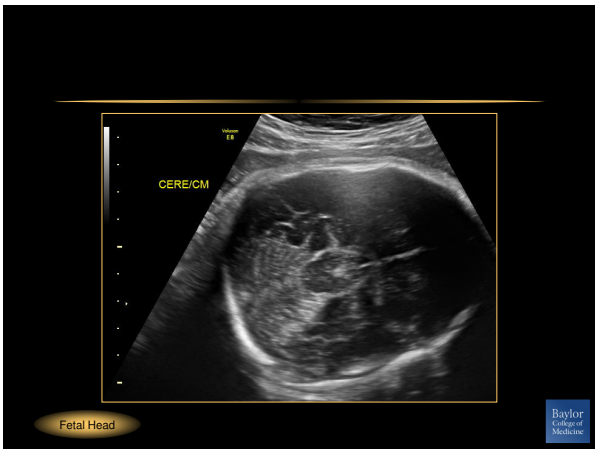
- Shallow posterior fossa
- Small cerebellar diameter
- "Banana" sign if severe
- Malformation is present in almost every case of thoracolumbar, lumbar, and lumbosacral myelomeningocele



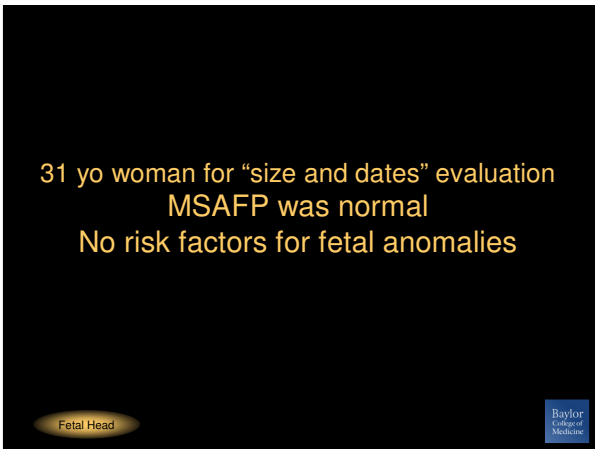




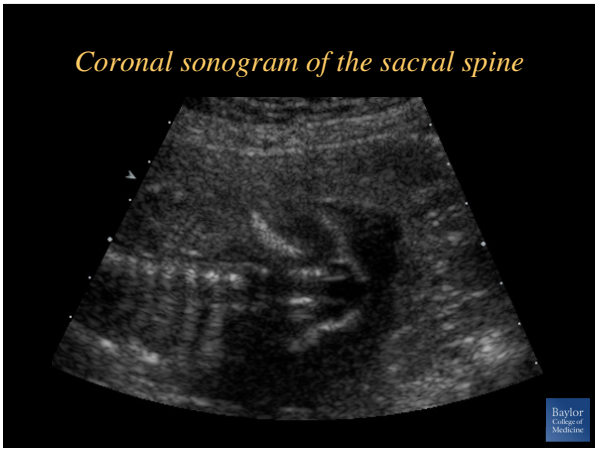
Search for ONTD



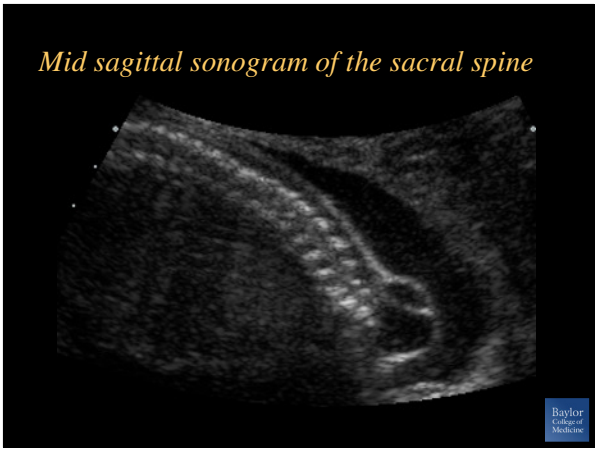
31 yo woman for “size and dates” evaluation
MSAFP was normal
No risk factors for fetal anomalies



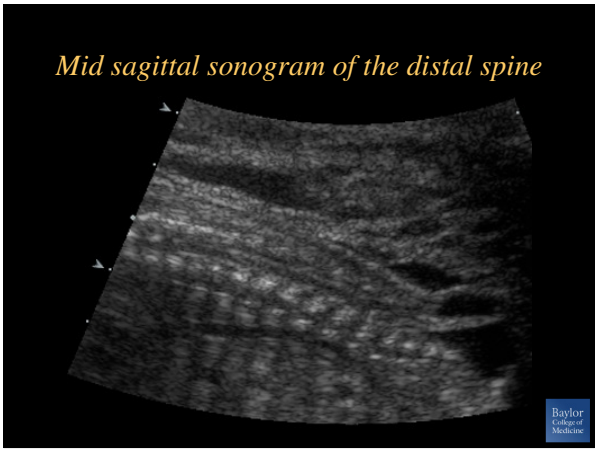
Coronal sonogram of the sacral spine



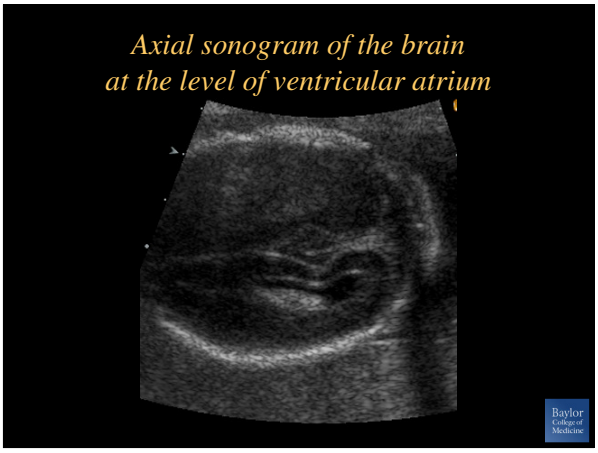
Mid sagittal sonogram of the sacral spine



Mid sagittal sonogram of the distal spine



Axial sonogram of the brain at the level of ventricular atrium



Axial sonogram of the brain at the level of posterior fossa

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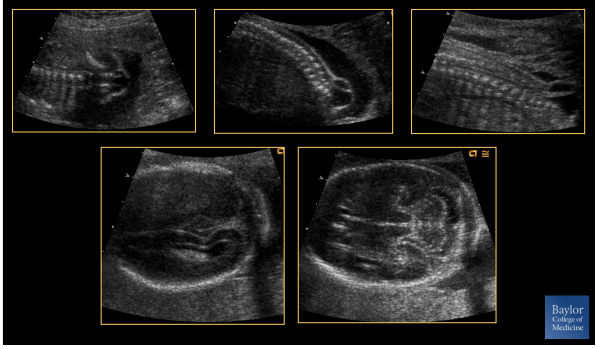
Skin Covered Spina Bifida



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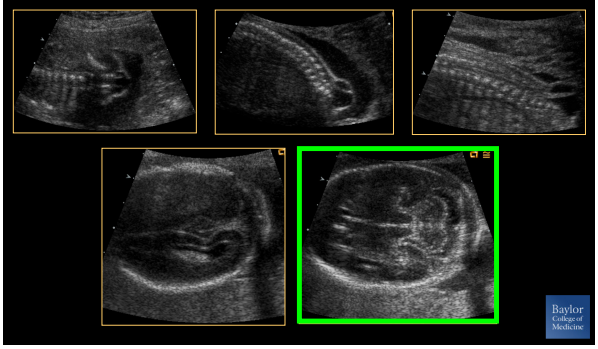
Open neural tube defect is associated with Arnold-Chiari II malformation



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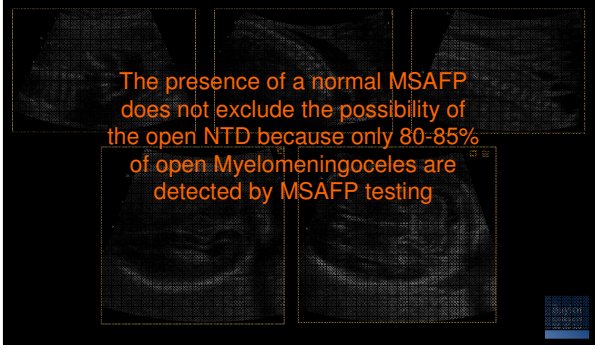
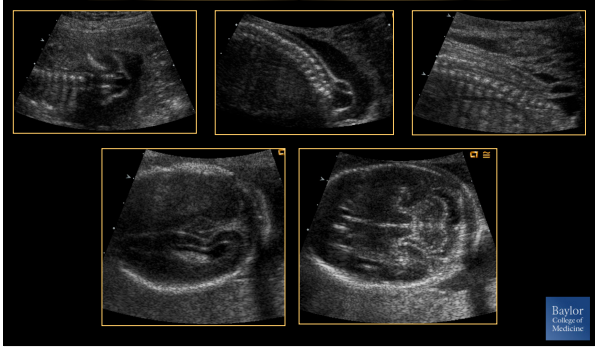
With a closed lesion one would anticipate that the mother would demonstrate a normal MSAFP
MSAFP test is only useful for detection of the open NTD



The presence of a normal MSAFP does not exclude the possibility of the open NTD because only 80-85% of open Myelomeningoceles are detected by MSAFP testing

The AFAFP is positive in virtually ALL open NTDs



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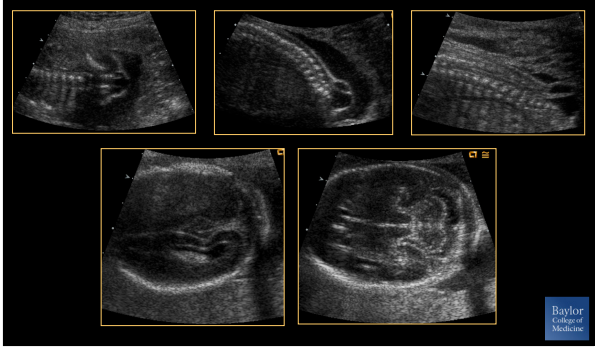
The closed lesions typically have a better prognosis
In part this is due to the lack of the Chiari II malformation and a lack of development of hydrocephalus



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Dandy-Walker Complex

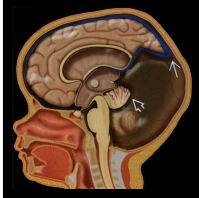
- Dandy-Walker malformation
  - Complete or partial agenesis of the cerebellar vermis
  - Enlarged posterior fossa
- Dandy-Walker variant
  - Partial agenesis of the cerebellar vermis
  - Without enlargement of the posterior fossa
- Mega cisterna magna
  - Normal vermis & 4th ventricle

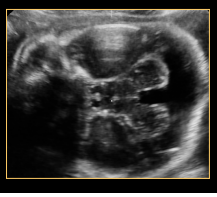
Fetal Head

Baylor College of Medicine

Dandy-Walker Malformation

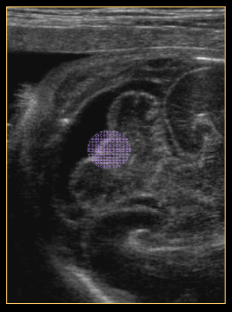
- Defect in the cerebellar vermis through which the cyst communicates with the 4th ventricle
- Cerebellar hemispheres are separated





Fetal Head

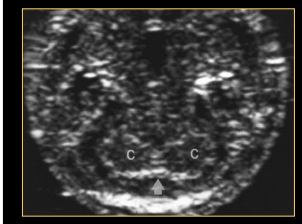
Cisterna Magna – Caution

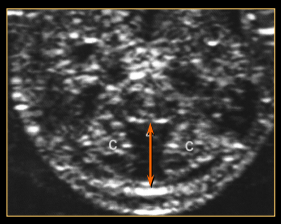


Fetal Head

Baylor College of Medicine

Cisterna Magna – Caution





Inferior vermis < 18 wks

Reevaluation at 20 to 22 postmenstrual weeks is crucial to confirm presence or absence of a normal vermis

Fetal Head

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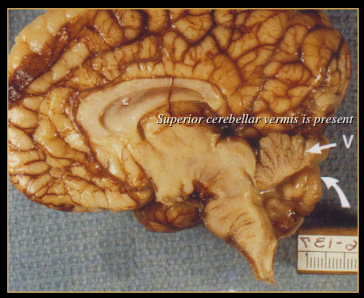
Dandy-Walker Varient

Absence of inferior cerebellar vermis



Fetal Head

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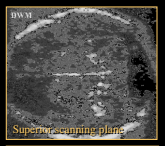
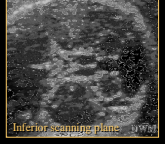
Dandy-Walker Variant

- Less severe malformation
- Smaller 4th ventricle cyst
- Ventriculomegaly usually absent
- Harder to see
- More associated with chromosomal abnormalities





Fetal Head

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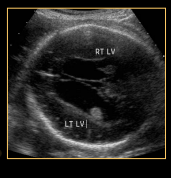
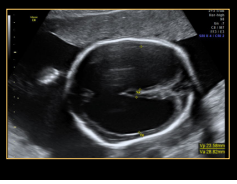
Ventriculomegaly

- Enlarged cerebral ventricles
- "Hydrocephalus" implies obstruction
- High association with
  - CNS & non CNS anomalies
  - Chromosomal anomalies





Fetal Head

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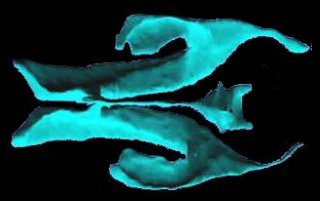
Lateral Ventricle





Stable in size from 16-40 wks – age independent

CHOROID PLEXUS
Usually fills 90% of atrium
Always fills 60% of atrium

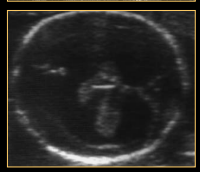
Mami Moriyama, RDMS
Fetal Head
Fetal Survey

Baylor College of Medicine

Ventriculomegaly

- Enlarged atrial measurement
  - < 25 weeks: > 8mm
  - > 25 weeks: >10mm

  Male fetuses have wider measurements
- Choroid fills < 50% atrial diameter
- Dangling choroid sign

Fetal Head

Ventriculomegaly

- Three major pathologic processes can result in ventricular enlargement
  1. Obstructive hydrocephalus
     - Communicating
     - Non-communicating
  2. Maldevelopment of the ventricle or surrounding brain tissue
  3. Destruction of surrounding brain tissue
     - Congenital infection - bacterial meningitis or viral infections
     - Vascular mechanism
     - Tumors
  4. Over production of CSF 2º to the CP tumor – Rare

Fetal Head

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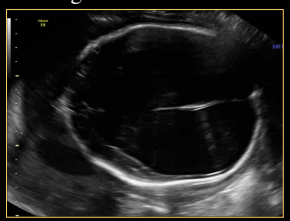
Hydrocephalus

- Abnormal accumulation of CSF results in enlargement of the ventricular system




Fetal Head

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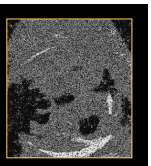
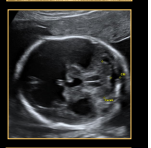
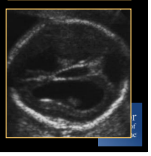
Hydrocephalus

- Enlarged lateral ventricles

  Plus at least one of the following
  - 3rd & 4th ventricular dilatation
  - Disrupted falx midline echo
  - Posterior fossa abnormalities
  - Head enlargement
  - Increasing ventricular size
    - Interval study

Fetal Head

Hydrocephalus

- Results from an imbalance between the production and absorption of CSF

3 major forms:

- Aqueductal stenosis, 43%
- Communicating hydrocephalus, 38%
- Dandy-Walker syndrome, 13%

Fetal Head

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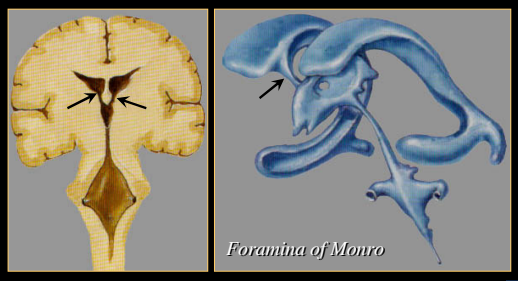
Cerebrospinal Fluid Circulation



Fetal Head

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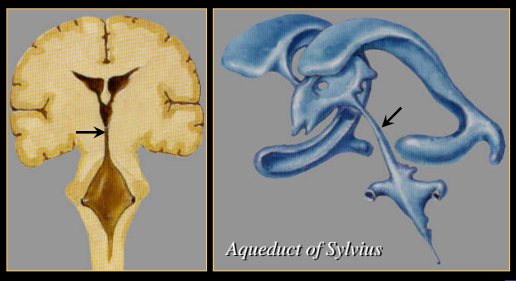
Cerebrospinal Fluid Circulation



Fetal Head

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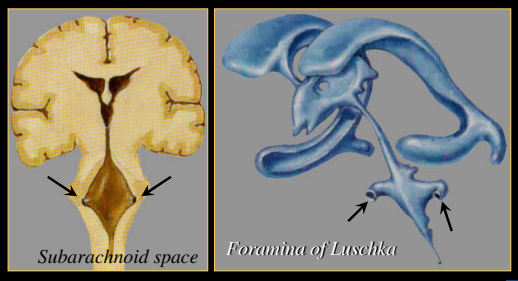
Cerebrospinal Fluid Circulation



Fetal Head

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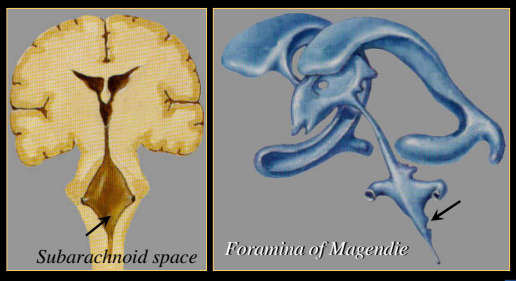
Cerebrospinal Fluid Circulation



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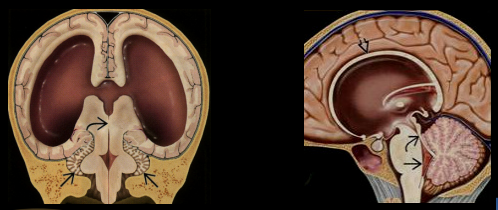
Aqueductal Stenosis

- A form of obstructive hydrocephalus caused by narrowing of the aqueduct of sylvius
- Narrowed connection between 3rd & 4th ventricle



Fetal Head

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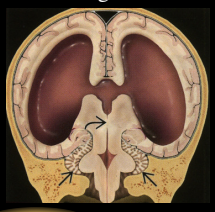
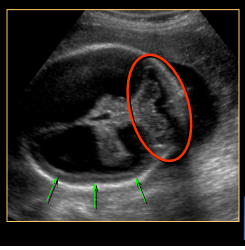
Aqueductal Stenosis

- Enlarged lateral & 3rd ventricles
- Normal posterior fossa (4th ventricle)
- Thinning of cortical mantle





Fetal Head

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Communicating Hydrocephalus

- A form of enlargement of the ventricles and subarachnoid system caused by an obstruction to CSF flow outside the ventricular system

Fetal Head

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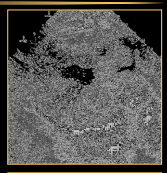
Communicating Hydrocephalus

- Ventriculomegaly
  - Dilatation of the lateral, 3rd, & 4th ventricles
- Brain has shrunk
- Surface has fallen away from the skull
- CSF filled subarachnoid space



Fetal Head

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Midline Anomalies of the Brain

- Holoprosencephaly
- Septo-optic dysplasia
- Agenesis of corpus callosum

Fetal Head

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Holoprosencephaly

- Heterogeneous entity of central nervous system anomalies

Categorization introduced by DeMyer & Zeman in 1963
Recognition by ultrasound by Kurtz in 1980

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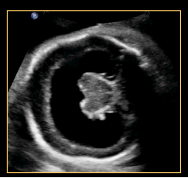
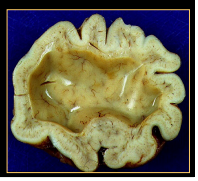
Holoprosencephaly

- Incomplete cleavage or diverticulation of the premitive forebrain into two cerebral hemispheres
- Single ventricle with no separation of the frontal lobes





Fetal Head

Holoprosencephaly

- Classifications:
  - Alobar *(lethal)*
  - Semilobar *(lethal)*
  - Lobar *(mental retardation)*

Fetal Head

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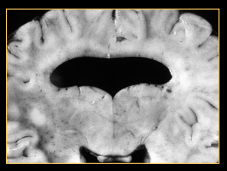
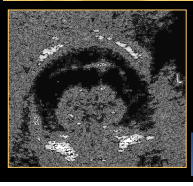
Alobar Holoprosencephaly

- No cerebral separation into two hemispheres
- Single ventricle
- Fused thalami
- No interhemispheric fissure
- Absence of the 3rd ventricle
- Absent corpus callosum





Fetal Head

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Alobar Holoprosencephaly

- Facial abnormalities common
  - proboscis, cyclopia, cebocephaly, B/L cleft lip & palate with premaxillary agenesis, hypotelorism

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Alobar Holoprosencephaly

- "Face Predicts the Brain" ~ 70% of time
- Converse is NOT true, 20% of alobar HPE have only minor facial dysmorphism

Fetal Head

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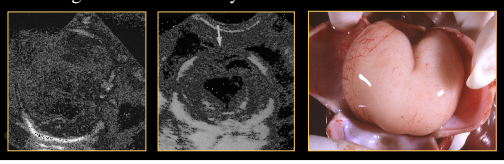
Semilobar Holoprosencephaly

- Lethal
- Partially separated two hemispheres
- Absence of falx
- Partial fusion of the thalami
- Partial Agenesis of the CC
- Single ventricular cavity



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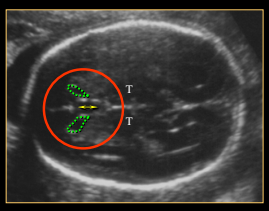
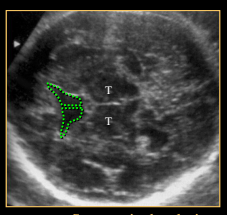
Absence of the CSP & fused frontal horns





Septooptic dysplasia

Fetal Head

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Agenesis of the Corpus Callosum

- Can be complete or partial, depending upon the stage of development at which growth was arrested
- Outcome is heavily dependent on the presence or absence of associated anomalies

Fetal Head

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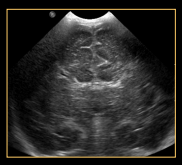
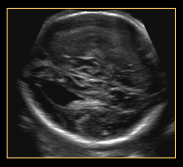
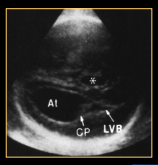
Agenesis of the Corpus Callosum

- No cavum septum pellucidi
- Teardrop configuration of the lateral ventricle
- Upward displacement of the 3rd ventricle

Colpocephaly

Fetal Head

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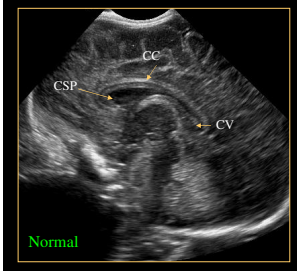
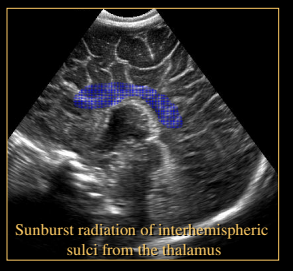
Agenesis of the Corpus Callosum



Normal



Sunburst radiation of interhemispheric sulci from the thalamus

Fetal Head

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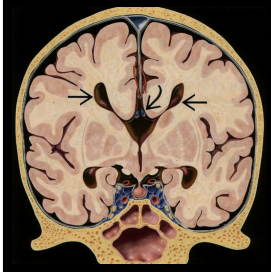
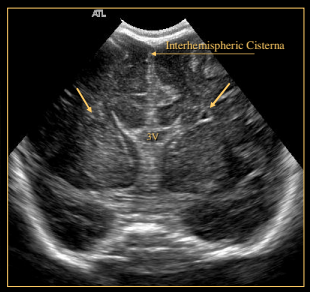
Agenesis of the Corpus Callosum



Widely spaced lateral ventricles



3rd ventricle is elevated and is contiguous dorsally with the interhemispheric fissure

Fetal Head

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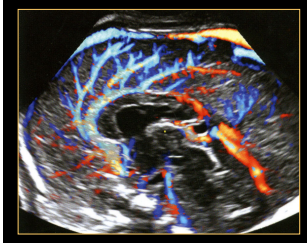
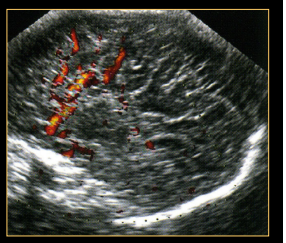
Agenesis of the Corpus Callosum

Pericallosal Artery

Fetal Head

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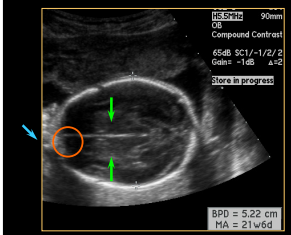
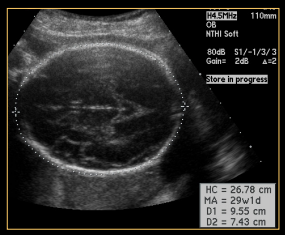
Head Measurements





Fetal Head

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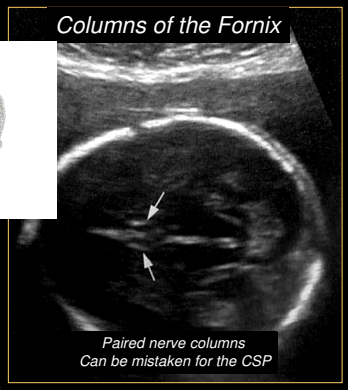
Columns of the Fornix



Paired nerve columns
Can be mistaken for the CSP

Fetal Head

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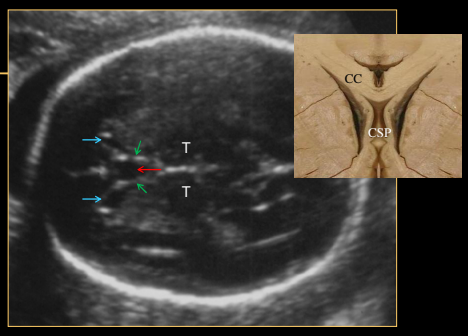

CSP appears as a fluid-filled box between its septi, which separate this space from the frontal horns of the LVs

Fetal Head

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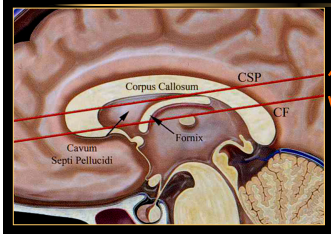

Fetal Head

Peter W. Callen et al JUM January 1, 2008 vol. 27 no. 1 25-31
Thomas C Winter et al *JUM March 1, 2010 vol. 29 no. 3 427-444*

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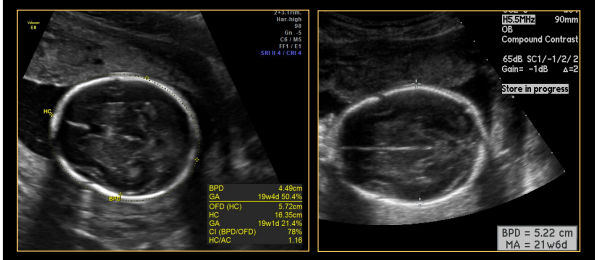
Thalamic View



Fetal Head

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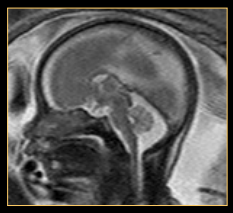
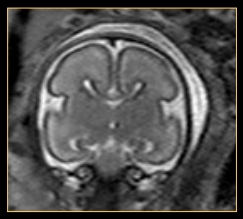
Agenesis of the Corpus Callosum

- Failure of axons to cross midline and form CC





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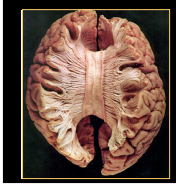
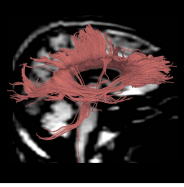
Diffusion Tensor Imaging

- The motion of protons within different tissue types, and, by measuring the amount and directionality of diffusion, it provides information on 3D tissue properties





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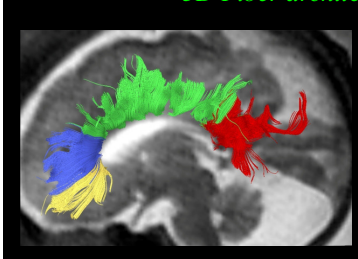
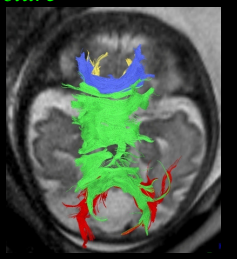
Normal development

3D Fiber architecture





Rostrum Genu Truncus Splenium

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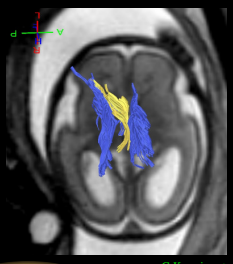
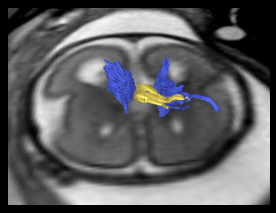
Callosal abnormality?

- MRI - Tractography





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