Fetal Central Nervous System

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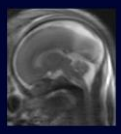

Fetal Care Center of Cincinnati
Comprehensive Diagnosis & Treatment

Cincinnati Children's Hospital Medical Center

Disclosures

- *none*

Learning Objectives

1. Standard approach to normal fetal neurosonology
2. Understand the fetal CNS pathology with regard to stages of embryologic development
3. Correlate sonography with fetal MR imaging

Lecture Outline

- Normal fetal sonography
- Limitations of sonography
- Fetal MRI as an adjunct
- Fetal CNS embryology with regard to pathology
  - Dorsal induction
  - Ventral induction
  - Cell proliferation, migration and organization
  - Destructive
  - Vascular

CNS anomalies account for 9% of isolated and 16% of multiple prenatal malformations

Many are associated with genetic/chromosomal abnormality

Ultrasound is the modality of choice in the imaging of disorders related to the fetus and pregnancy

Ultrasound

- 1st Trimester
  - Transabdominal
  - Transvaginal
- 2nd and 3rd Trimester
  - Transabdominal
  - Transvaginal
    - Cephalic presentation
    - Higher frequency probes improve anatomic detail

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Ultrasound

- Imaging planes
  - Transventricular
    - *Lateral ventricles*
    - Ventricular atrial transverse diameter at level of choroid glomus
  - Transthalamic
    - *Frontal horns and septum pellucidum*
    - BPD and head circumference
  - Transcerebellar
    - *Midline thalamus, cerebellar hemispheres, vermis and cisterna magna*
    - Transverse cerebellar and cisterna magna

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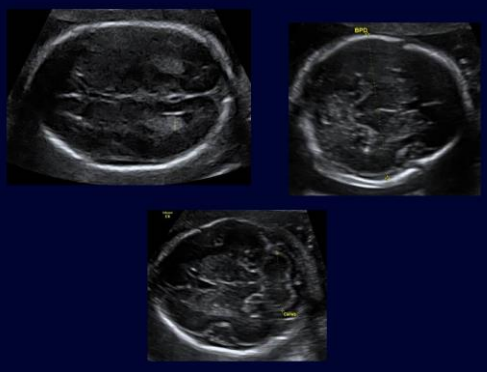

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Cerebral Pathology

- *97% of CNS anomalies can be identified on one or more of the three standard cranial views*
- *88% of CNS anomalies are identified on the tranventricular by diagnosis of enlarged ventricles*

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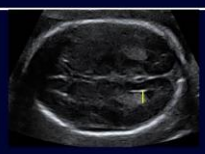
Ventricles

- Wall
  - echogenic, thin, smooth
- Widest level glomus choroid plexus
  - calipers should be inner aspect of the wall
  - Up to 10 mm normal



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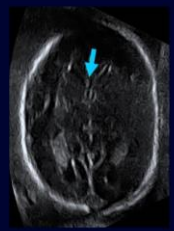
Septum Pellucidum (CSP)

- Bridge forming corpus callosum
- 18-20 wks
- Absence
  - Agenesis of the corpus callosum
  - Septo-optic dysplasia
  - Holoprosencephaly
  - Severe hydrocephalus



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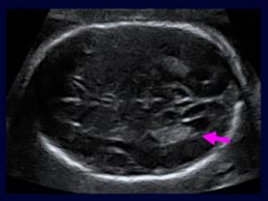
Germinal Matrix

- Not well delineated
- Walls
  - Nodularity
    - Heterotopia
    - Hemorrhage



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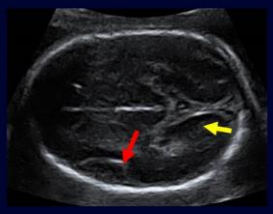
Brain Parenchyma

- Hypoechoic
- Echogenic cortex



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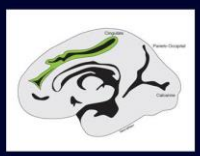
Sulcation

- Interhemispheric - late 1st/early 2nd
- Sylvian fissure -18 wks
- Parieto-occipital -20-22 wks
- Calcarine - 25 wks
- Cingulate - 26-28 wks
- Convexity sulci difficult



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Corpus Callosum

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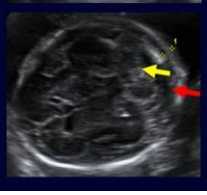
Posterior Fossa

- Cisterna magna
  - 2-10 mm
- Cerebellar hemispheres
  - hypo to moderately echogenic
- Vermis
  - Echogenic midline
  - Not cover 4th ventricle until 18 wks



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Sagittal and Coronal

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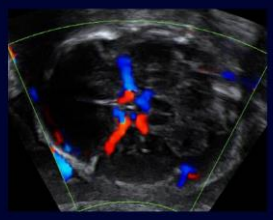
Ultrasound Doppler



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Ultrasound 3D Reconstructions

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Ultrasound 3D/4D Soft Tissue/Bone

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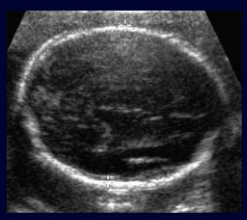

Reverberation artifact

28 WEEKS

Skull ossification

34 WEEKS

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US limitations

- Soft tissue contrast
  - Germinal matrix
  - Parenchymal detail
    - Migrating cells
- Fetal positioning
- Maternal body habitus
- Amniotic fluid
- Age- calvarial ossification

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Fetal MRI

- Large field of view (FOV)
- High soft tissue contrast
- High resolution
- Not inhibited by maternal body habitus, amniotic fluid or fetal positioning

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MRI led to a change in diagnosis in 32% of cases of US-detected fetal brain abnormalities, and changed counseling in 50%, and patient management in 19%

Levine D, Barnes PD, Robertson RR, et al. Fast MR imaging of fetal central nervous system abnormalities. *Radiology*. 2003;229:51–61.

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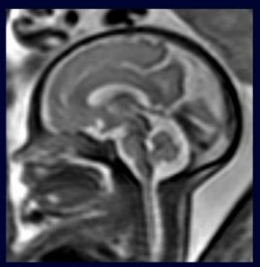
MRI Advantages in CNS Anomalies

- Intracranial soft tissue definition
  - Blood, ischemia, migrational anomaly
- Corpus callosum
- Posterior fossa anatomy
- Craniocervical anatomy
- Spinal cord depiction



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CNS Malformations

- *Ultrasound defines a CNS anomaly*
- *Fetal MR imaging to increase definition of the malformation*

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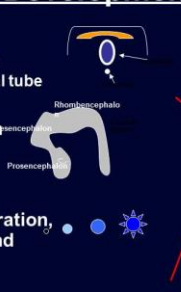
Stages of Embryonic Development

- Dorsal Induction
  - Closure of neural tube
- Ventral Induction
  - Cerebellum
  - Brainstem
- Neuronal proliferation, differentiation and migration



Driven by genes

Malformations often seen in presence of genetic disorders

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CNS Anomalies

- Dorsal Induction
  - Neural tube closure defects
- Ventral Induction
  - Midline Anomalies
  - Posterior Fossa Malformations
- Disorders of Neural Cell Proliferation and Migration
- Destructive Lesions
- Vascular

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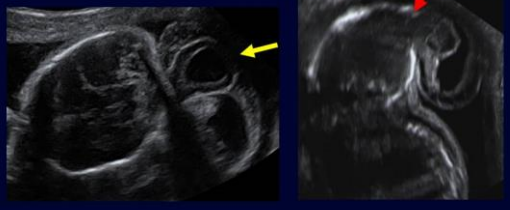
23 weeks



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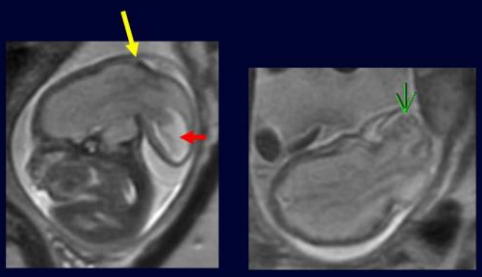

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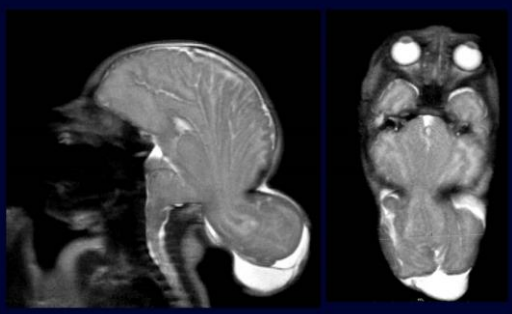

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Neural Tube Defects Dorsal Induction

- Anencephaly
  - absence cranial vault above the bony orbits
- Cephalocele
  - Protrusion of intracranial structures through a midline defect in the skull.
  - Type and frequency dependent ethnic group
    - Occipital- European
    - Frontal – Southeast Asian
  - Associated syndromes
  - Poor prognosis- microcephaly with neural elements in defect

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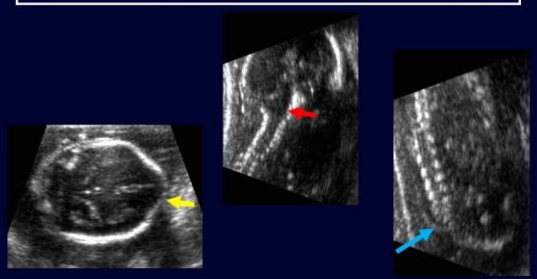
23 weeks



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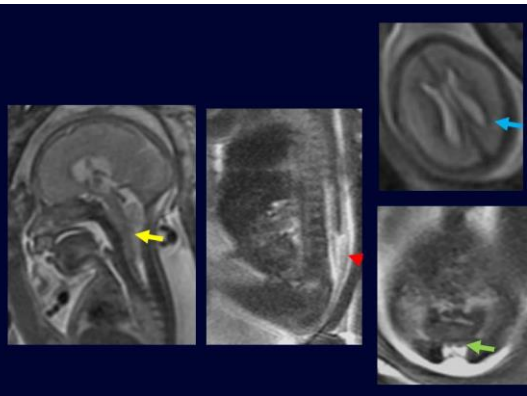

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Chiari II

- Malformation of the hindbrain
  - Small posterior fossa
    - Cerebellum and brainstem herniate through foramen magnum
    - Compressed, elongated, low-lying 4th ventricle
    - Low-lying, abnormally vertical tentorium
- Hydrocephalus
- Nearly 100% associated with neural tube defect
- Pathology
  - Folate deficiency

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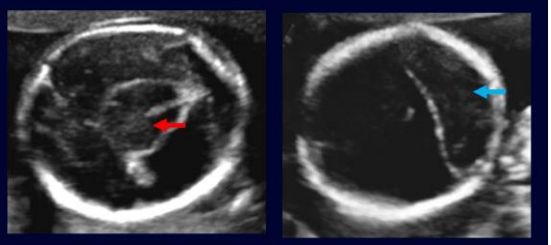
21 weeks



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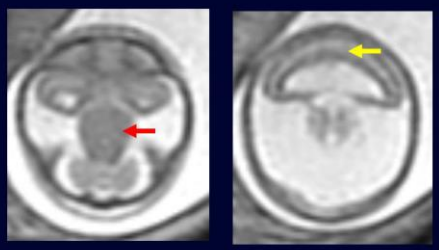

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Holoprosencephaly Ventral Induction

- Findings
  - Single primitive ventricle
  - Fused thalami
  - Facial anomalies
    - Eye/Nose/Lip
  - Pancake brain
  - Absent septum pellucidum

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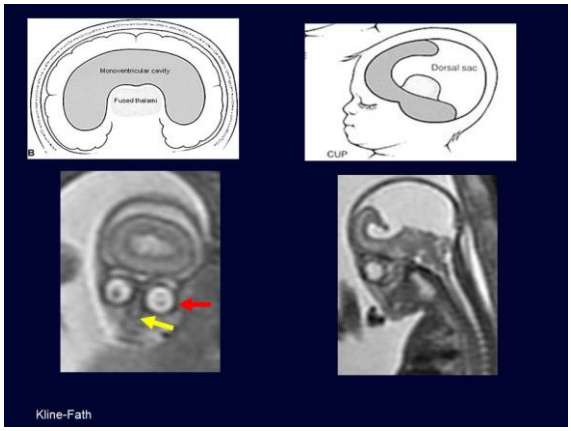

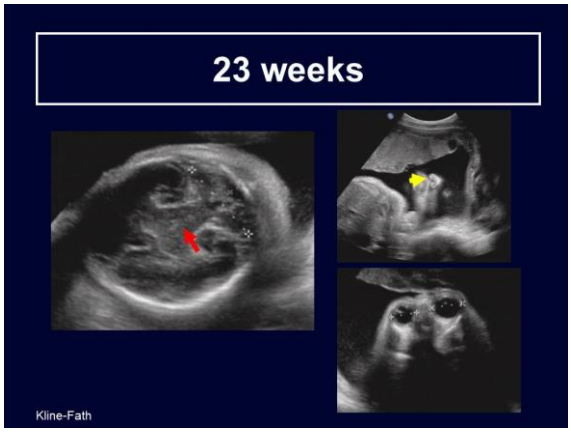

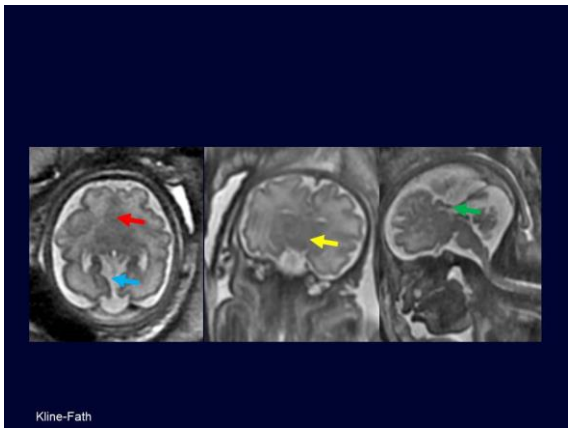

Holoprosencephaly

- Failure of cleavage of the prosencephalon (cerebral hemispheres)
  - Alobar
  - Semilobar
  - Lobar
  - Middle interhemispheric fusion
- 1 per 16,000 newborns
- High intrauterine fatality
- Genetic – Chromosome 11
  - Trisomy 13 (50-75%)

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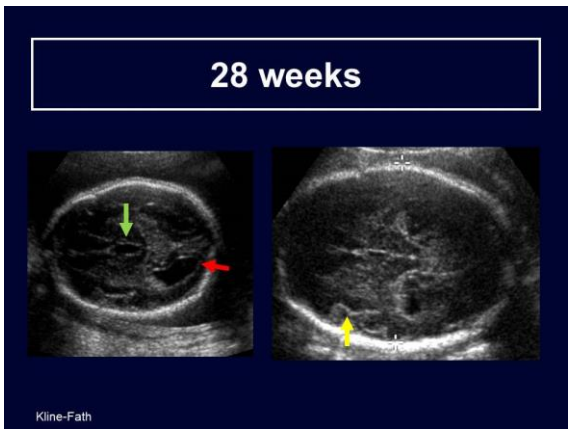

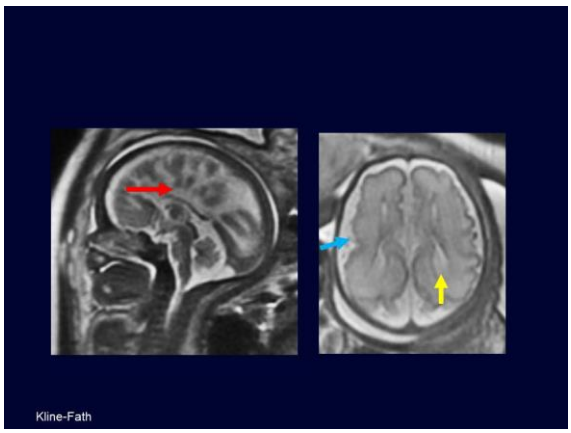

Agenesis of Corpus Callosum Ventral Induction

- Ventriculomegaly
  - Colpocephaly
  - Frontal horn deformity "moose head"
- Absence of septum pellucidum
- Increased separation of hemispheres
- Upward displaced third ventricle
- Midline cyst or other lesion
- Absence of the pericallosal artery

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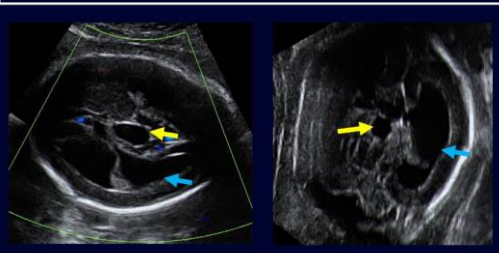
28 weeks



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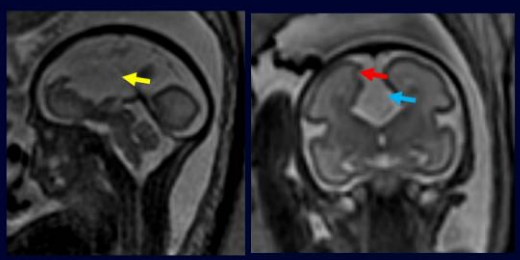

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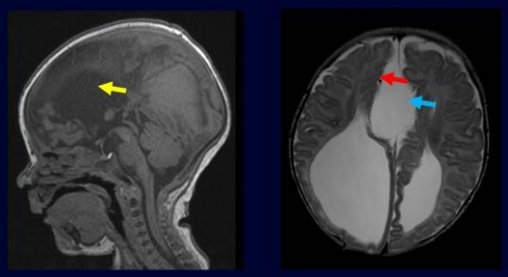

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Agenesis of Corpus Callosum

- .3 to .7% population
- Etiology
  - Genetic (Aicardi sydrome)
  - Teratogens (alcohol, valproate, cocaine, rubella and influenza virus)
- Associations
  - Lipomas
  - Interhemispheric cyst
- Increased anomalies, worse neurologic outcome

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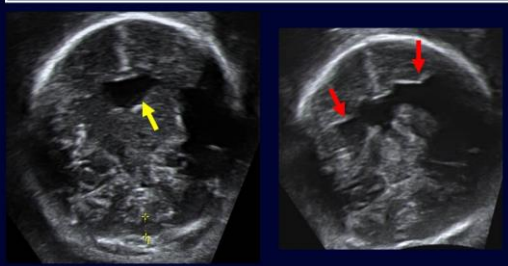
34 weeks



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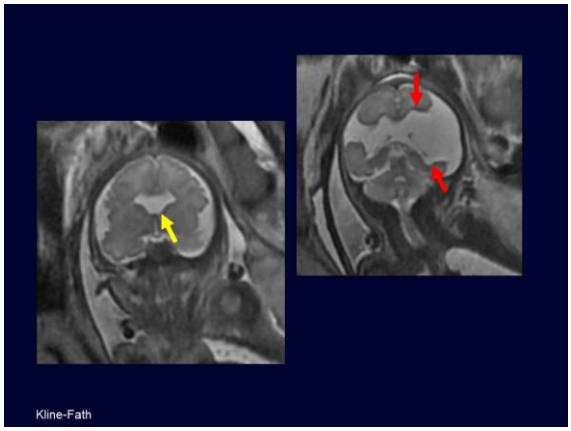

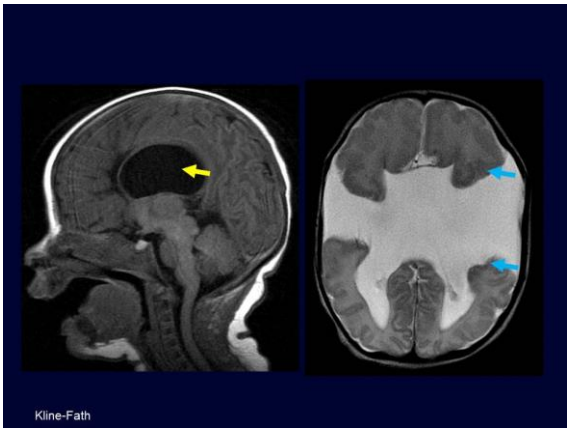

Septooptic Dysplasia Ventral Induction

- De Morsier syndrome
- Key findings
  - Hypoplasia optic nerves
  - Absent septum pellucidum
  - Hypothalamic-pituitary dysfunction
- Other cerebral anomalies
  - Schizencephaly-bilateral or unilateral full surface clefts of cortical mantle
- Prognosis
  - Sudden death with pituitary dysfunction
  - Severity of brain findings

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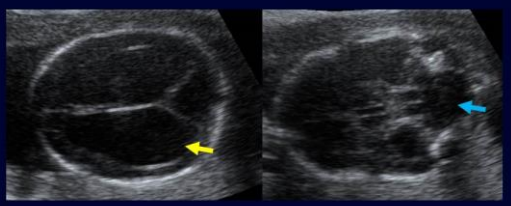
22 weeks



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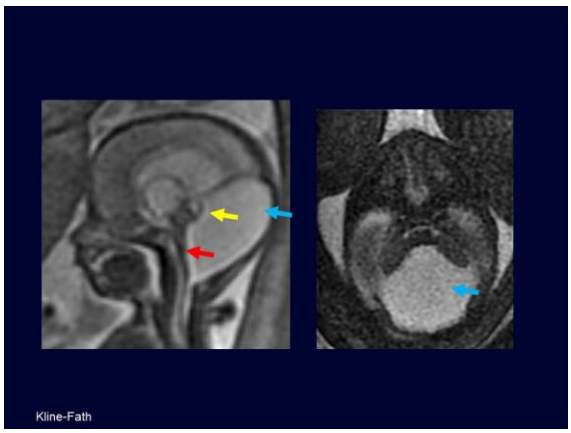

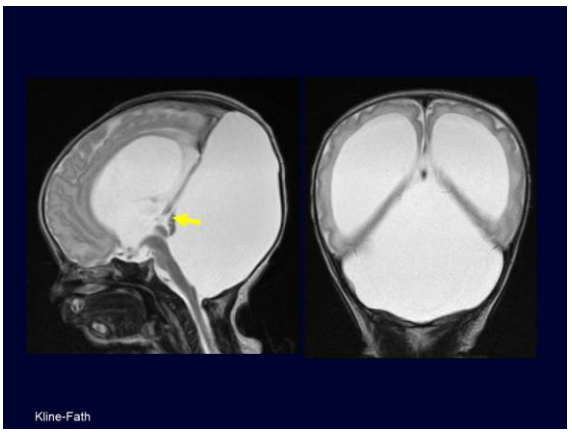

Vermian Anomalies/Posterior Fossa Ventral Induction

- Malformation (1 per 30,000 birth)
  - Cystic dilatation of the fourth ventricle
  - Dysgenesis of cerebellar vermis
  - High position of the tentorium
- Etiologies
  - Genetic
  - Teratogens (viral, alcohol, diabetes)
- 2/3 associated CNS/extracranial anomalies
  - Worse outcome
- Postnatal mortality 35%

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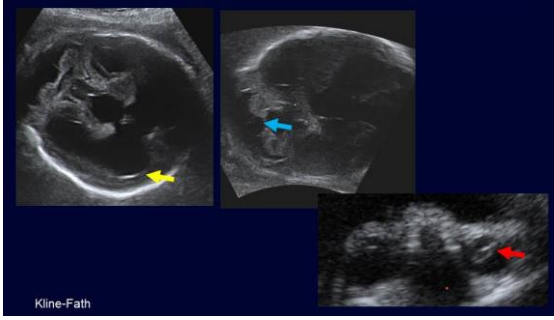
32 weeks



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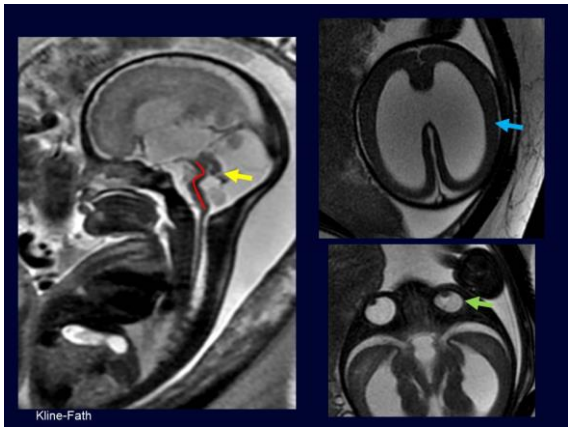

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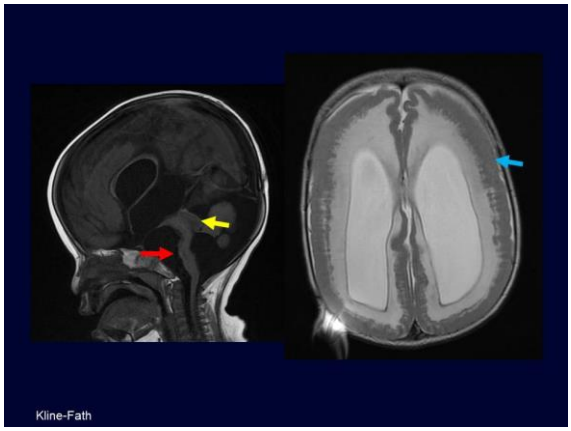

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Walker Warburg

- Congenital Muscular Dystrophy
  - Lissencephaly
  - Hydrocephalus
  - Kink at the mesencephalic-pontine junction; pontine hypogenesis
  - Cerebellar hypoplasia and dysplasia
  - Severe hypotonia
  - Eye malformation

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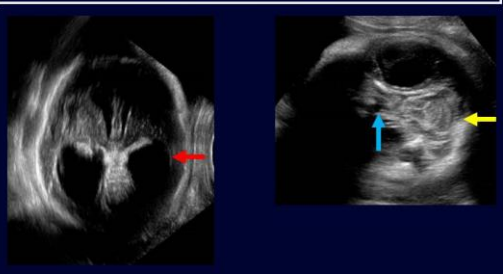
33 weeks



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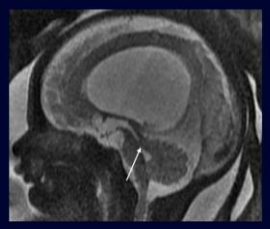

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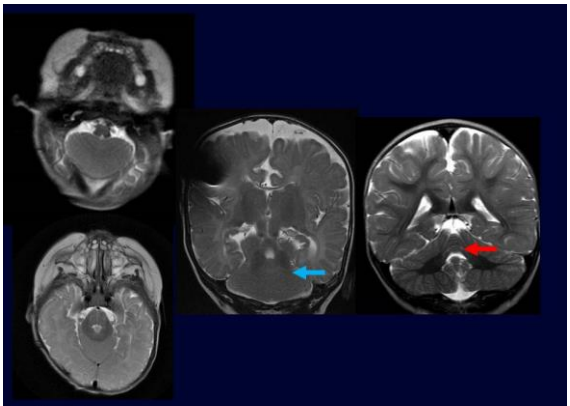

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Aqueductal Stenosis Ventral Induction

- Severe >15mm
- 1 in 2000
- Sporadic
  - Infection
  - Hemorrhage
  - Tumors
- Rhomboencephalosynapsis
  - Fusion of cerebellar hemispheres & vermian agenesis
- Genetic (X-linked 5%)
- Prognosis poor (10% normal development)

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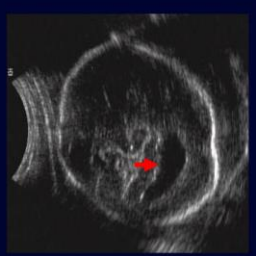
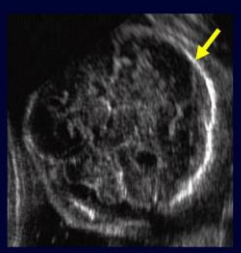
Neuronal Proliferation/Migration

- Proliferation
  - Hemimegalencephaly- hamartomatous overgrowth defect in neuronal proliferation and migration.
- Migration
  - Lissencephaly – smooth brain
  - Schizencephaly – full thickness gray matter line clefts of the cerebral mantle

25 weeks





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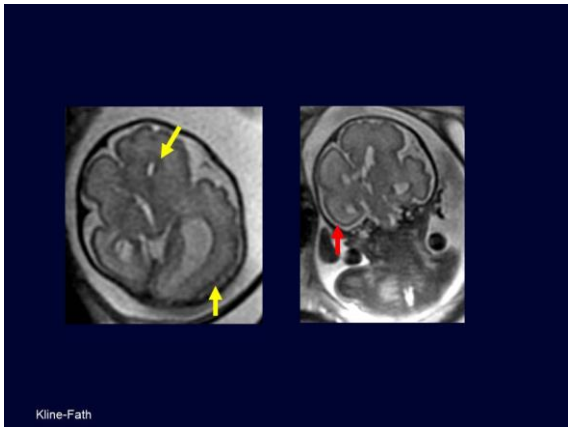
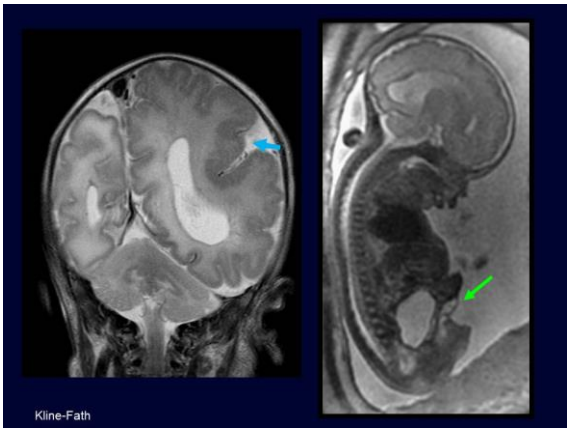
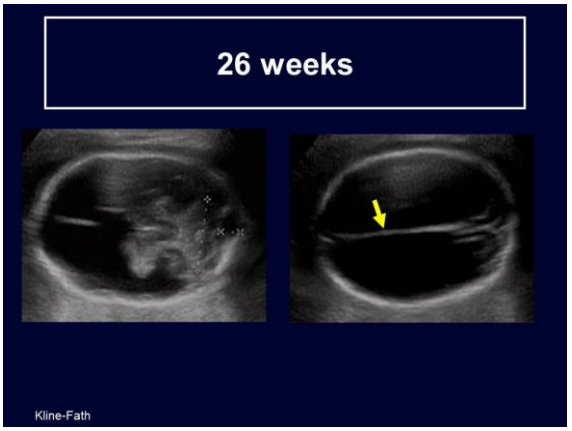


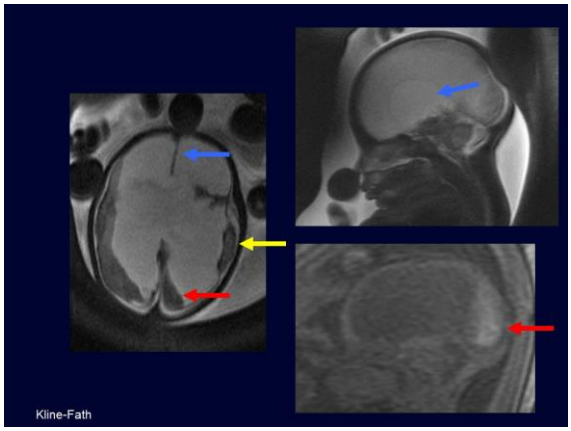


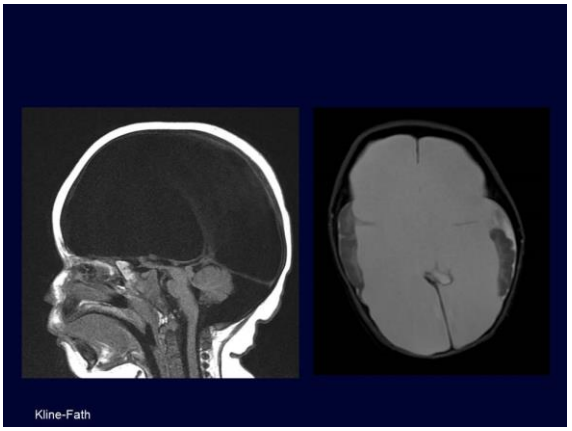
Hemimegalencephaly Neuronal Proliferation/Migration

- Hamartomatous overgrowth of part/all of a hemisphere
- Defect of cell organization/neuronal migration
- Imaging
  - Enlarged dysplastic hemisphere
  - Large lateral ventricle
- Associated syndromes
  - Hemiovergrowth
- Poor outcome intractable seizure/hemiparesis

Kline-Fath







Destructive Lesions

- Intracranial hemorrhage
- Porencephaly
  - Large defect that communicates with ventricular system
- Hydranencephaly
  - Absence of cerebral hemispheres, replaced by sac like CSF structures
  - 1 to 2.5 per 10,000
  - Occlusion of carotid vessels/toxic exposure
  - Lethal

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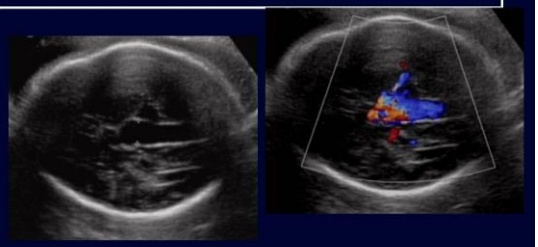
32 weeks



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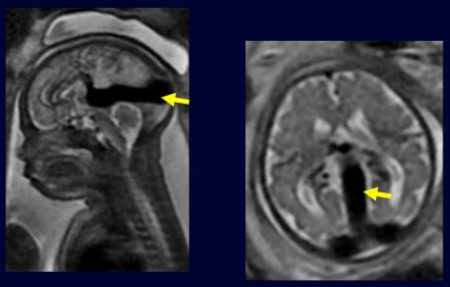

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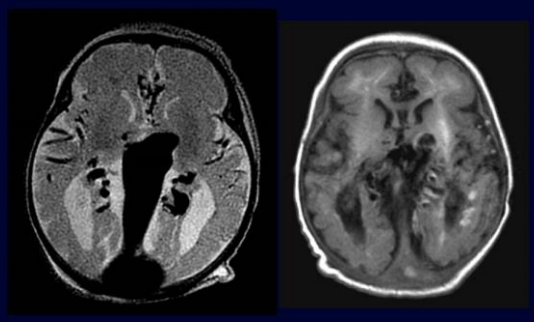

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Vascular

- Vein of Galen Aneurysm
  - Spectrum of arteriovenous malformations
  - Persistence of a fetal vein, the median prosencephalic vein
  - Findings
    - Anechoic color Doppler structure in posterior third of brain
    - Hydrops
    - Cardiomegaly
    - Brain injury/ischemia
  - Prognosis worse-hydrops and cardiomegaly

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Conclusion

- Goal
  - Understand normal neurosonology
  - Understand neural embryology
  - Define pathology

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