

FETAL CENTRAL NERVOUS SYSTEM ABNORMALITIES

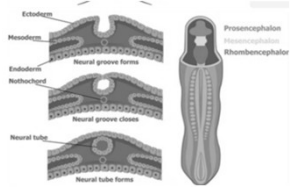
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DISCLOSURES

I have no actual or potential conflict of interest in relation to this presentation.

2

NEURULATION



The diagram illustrates the process of neurulation in four stages: 1. Ectoderm differentiates into the neural plate (neural groove forms) and surface ectoderm (neural groove closes). 2. The neural plate folds inward to form the neural tube. 3. The neural tube closes cranially and caudally. 4. The neural tube forms the neural tube, which will become the central nervous system. Labels include: Ectoderm, Neural groove forms, Neural groove closes, Neural tube, Neural tube forms, Prosencephalon, Rhombencephalon, and Endoderm.

- Dorsal ectoderm forms neural plate
- Neural plate folds inward forming neural folds
- Neural folds fuse to form neural tube
- Neural tube closes cranial and caudal
 - Disruption= Neural tube defect

3

OBJECTIVES

- Recognize 1st trimester indicators of CNS malformations
- Image optimization
- Updates in fetal surgery for Spina Bifida

4

RISK FACTORS

- Lack of folate in maternal diet
- Medication (Epilepsy)

5

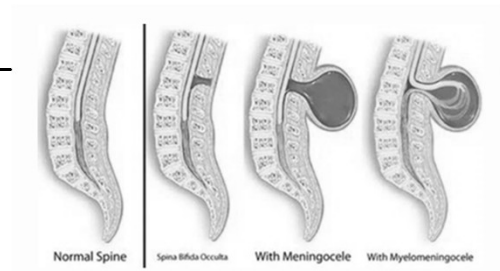
NEURAL TUBE DEFECTS- CRANIALY		
Name	Area Involved	Presentation
Anencephaly	Cranium & brain	Lack of cranial vault No cerebral tissue
Acrania	Cranium	Lack of cranial vault
Encephalocele	Herniation of brain & meninges	Occipital or frontal opening Cystic/ solid mass
Iniencephaly	Occipital skull & cervical spine	Fusion of occipital skull to cervical spine Hyperextension of head

6

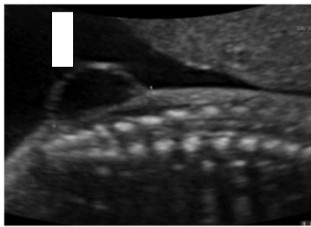
NEURAL TUBE DEFECTS CAUDALLY- SPINA BIFIDA

Name	Area Involved	Presentation
Spina bifida occulta	Gap between vertebrae	Dimple (post natively)
Meningocele	Meninges	Sac/cyst
Myelomeningocele	Meninges, spinal cord, nerves	Sac containing cord Chiari malformation Hydrocephalus

7



8



MENINGOCELE

9



MYELOMENINGOCELE

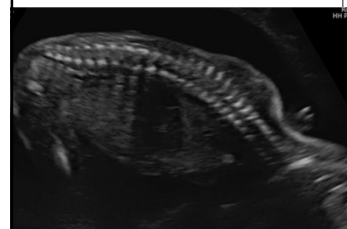
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Results	Report
Gest age.	16.4
Gest age based on	Ultrasound
Maternal age at edd	33.8
Race	
Weight	
Multiple gestation	no
AFP Value	84.7
AFP MoM	2.60
OSBR risk 1 in	238

SCREENING TESTS FOR ONTD

- MS AFP
 - Blood test
 - 16-18 weeks
- Ultrasound
 - 1st trimester
 - 2nd trimester

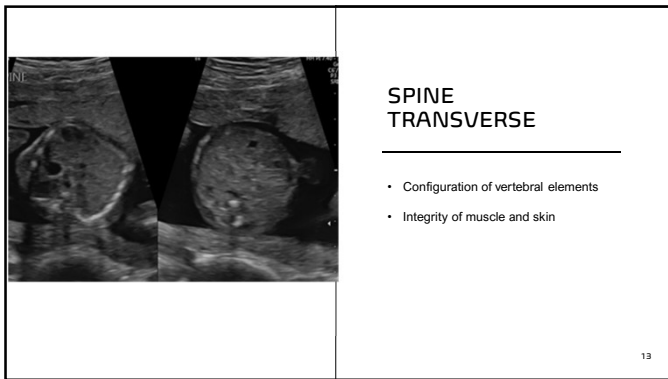
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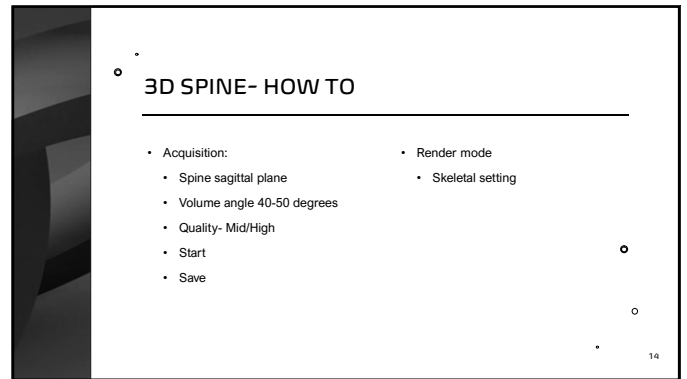
SPINE SAGITTAL

- Parallel arrangement anterior/posterior vertebral elements
- Curvature cervical and lumbosacral regions
- Sacral tapering
- Integrity of muscle and skin

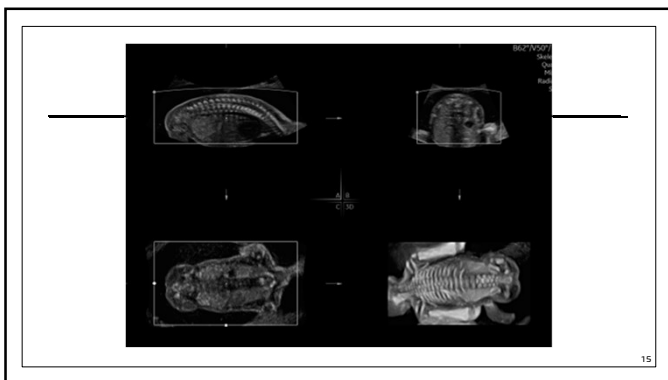
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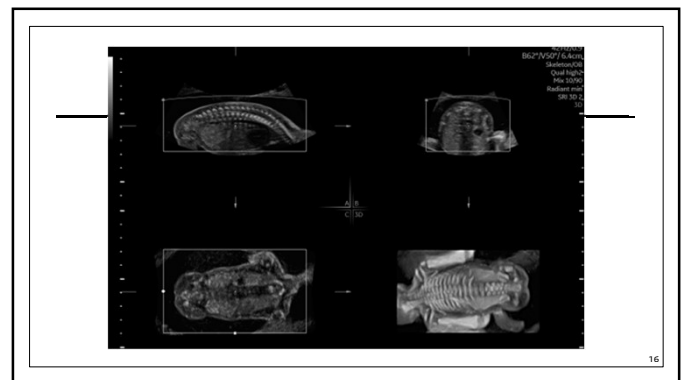
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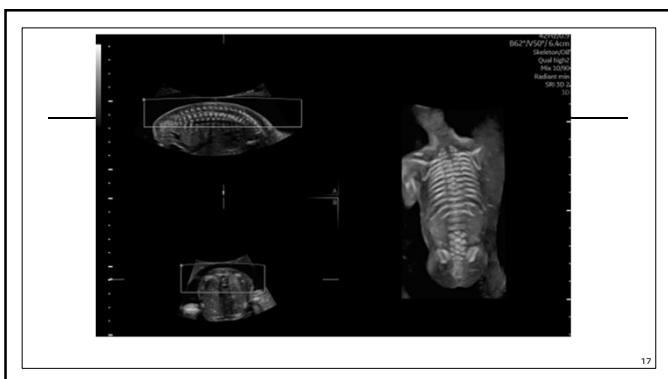
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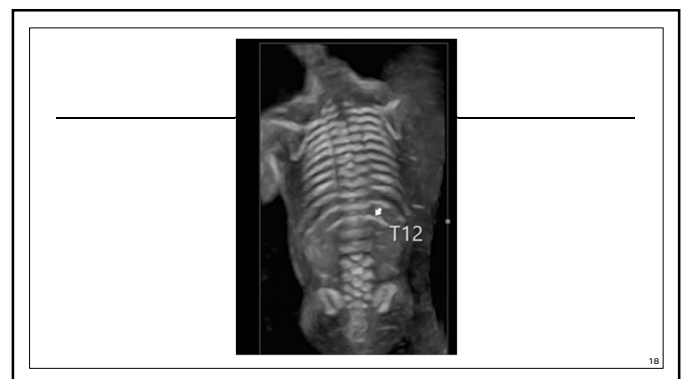
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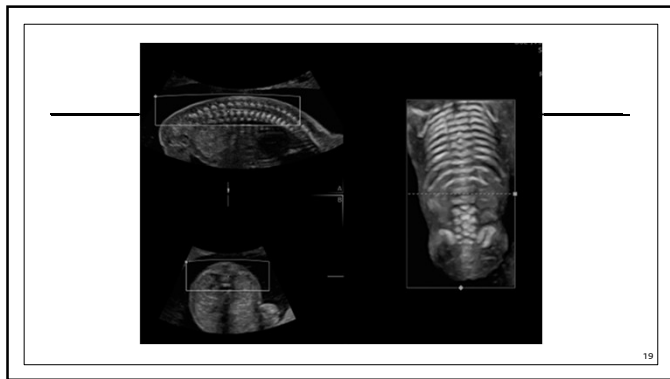
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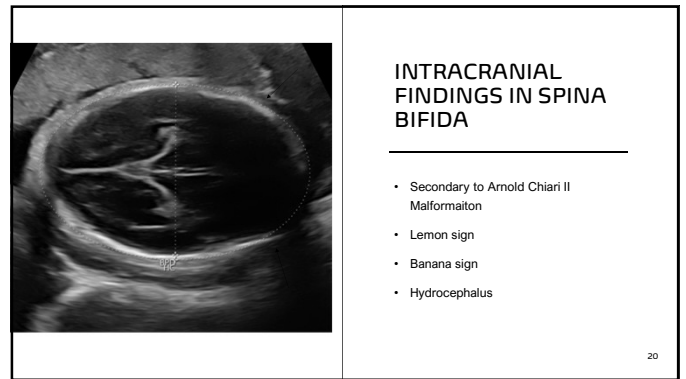
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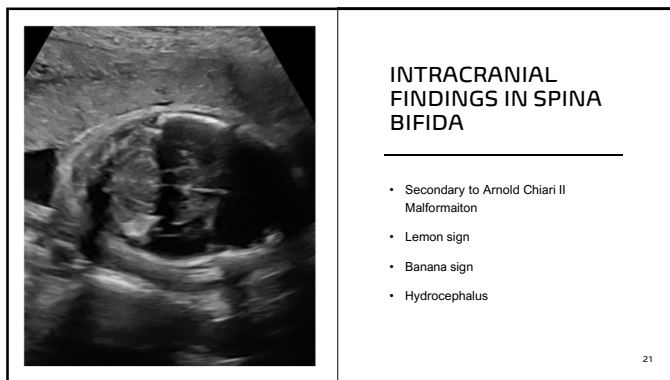
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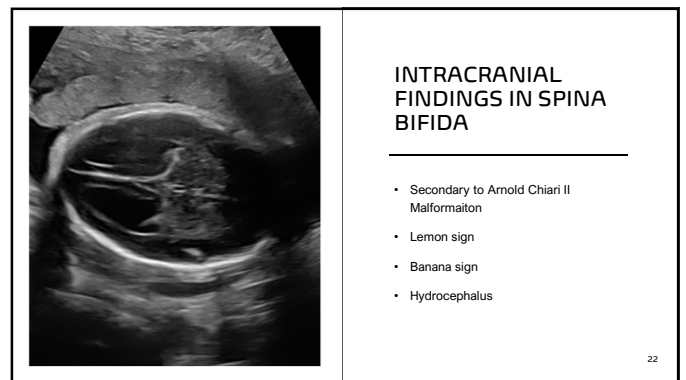
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20



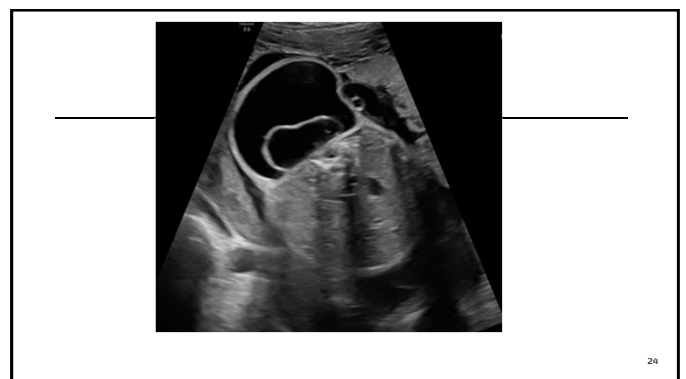
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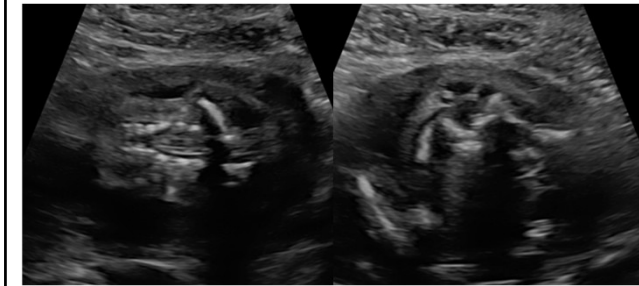
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23



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25

1ST TRIMESTER EVALUATION OF BRAIN 11-13 WEEKS

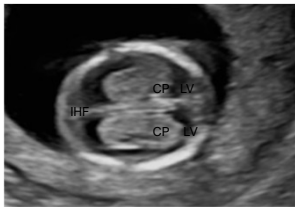
- Cranial bones
- Midline falx
- Choroid plexus filled ventricles

Abnormalities detected:

- Acrania
- Holoprosencephaly
- Cephalocele

26

26

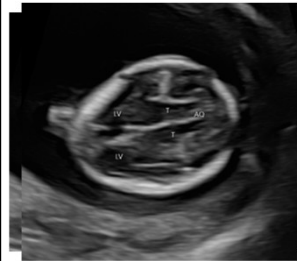


AXIAL VIEWS

- Suprathalamic
 - Interhemispheric fissure (IHF)
 - Lateral ventricles (LV)
 - Choroid plexus (CP)
 - Calvarium

27

27



AXIAL VIEWS

- Transthalamic
 - Third Ventricle (3V)
 - Aqueduct of Sylvius (Aq)
 - Thalamus (T)
 - 3rd ventricle (arrow)

28

28



AXIAL VIEWS

- Transthalamic
 - Third Ventricle (3V)
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 - 3rd ventricle (arrow)

29

29



SAGITTAL VIEWS

• Midsagittal brain- Nuchal Translucency view

- Diencephalon (D)
- 4th Ventricle (4v)
- Intracranial translucency
- Brain stem
- Mesencephalon (M)
- Pons (P)
- Medulla (Md)

30

30

SAGITTAL VIEWS

- Midsagittal brain- Nuchal Translucency view
 - Diencephalon (D)
 - 4th Ventricle (4v)
 - Intracranial translucency
 - Brain stem
 - Mesencephalon (M)
 - Pons (P)
 - Medulla (Md)

31

1ST TRIMESTER EVALUATION OF BRAIN 11-13 WEEKS

- Cranial bones
- Midline falx
- Choroid plexus filled ventricles

Abnormalities detected:

- Acrania
- Holoprosencephaly
- Cephalocele

32

1ST TRIMESTER DETAILED EVALUATION OF BRAIN 11-13 WEEKS

- Cranial bones
- Midline falx
- Choroid plexus filled ventricles
- Supratthalmic section
- Transtthalmic section
- Midsagittal (NT/IT) view

Abnormalities detected:

- Acrania
- Holoprosencephaly
- Cephalocele
- Ventriculomegaly
- Open spina bifida
- Dandy Walker Malformation
- Agenesis of Corpus Callosum

33

VENTRICULOMEGALY

- Reduction of Choroid Plexus size
- Too small to reach roof of lateral ventricle

34

OPEN SPINA BIFIDA

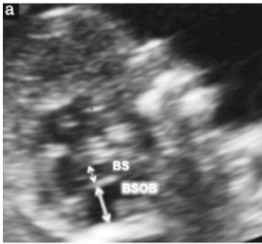
- Reduced width of cisterna magna
 - Brainstem thicker, displaced posteriorly
 - Compressed 4th ventricle

35

OPEN SPINA BIFIDA

- 3rd Ventricle & Aqueduct of Sylvius not seen

36



DANDY WALKER MALFORMATION

- Ratio brainstem thickness (BS) and brainstem to occipital bone (BSOB) reduced.
- Only 2 parallel anechoic spaces seen

Volpe, N., Dall'Asta, A., Di Pasquo, E., Frusca, T. and Ghi, T. (2021), First-trimester fetal neurosonography: technique and diagnostic potential. *Ultrasound Obstet Gynecol*, 57: 204-214

37

37



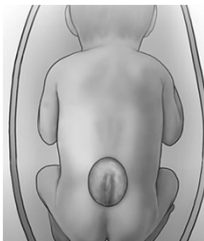
AGENESIS OF THE CORPUS CALLOSUM

- Increased ratio between diencephalon (D) and falx (F)

Volpe, N., Dall'Asta, A., Di Pasquo, E., Frusca, T. and Ghi, T. (2021), First-trimester fetal neurosonography: technique and diagnostic potential. *Ultrasound Obstet Gynecol*, 57: 204-214

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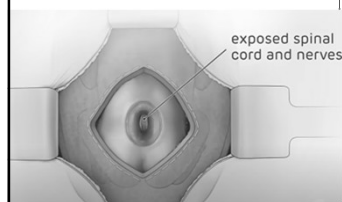
MYELOMENINGOCELE REPAIR

- Goal- protect spinal cord and nerves from damage from amniotic fluid
- Prevent hydrocephalus

CH Children's Hospital of Philadelphia

40

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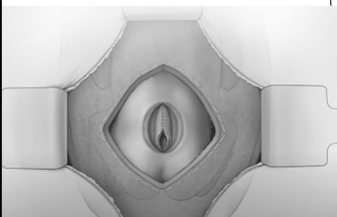


MYELOMENINGOCELE REPAIR

- Close dura
- Close muscle
- Close skin

41

41

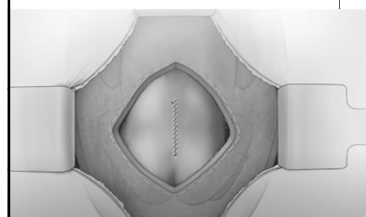


MYELOMENINGOCELE REPAIR

- Close dura
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42

42



MYELOMENINGOCELE REPAIR

- Close dura
- Close muscle
- Close skin

43

43

CRITERIA FOR ELIGIBILITY (SURGERY)

Eligible:

- Myelomeningocele Level T1- S1 with hindbrain herniation
- Gestational age: < 25w6d
- Maternal age: >18
- Singleton
- Labs: Elevated MSAFP, positive acetylcholinesterase
- Normal Karyotype

Contraindications:

- Insulin dependent diabetes mellitus
- Associated anomalies: cardiac, intracranial hemorrhage
- Fetal kyphosis
- Cervical insufficiency or cerclage
- Placenta previa
- Placental abruption
- BMI >40
- Other

44

SURGERY

Close to protect further damage

- Dura
- Muscle
- Skin

Contraindications:

- Insulin dependent diabetes mellitus
- Associated anomalies: cardiac, intracranial hemorrhage
- Fetal kyphosis
- Cervical insufficiency or cerclage
- Placenta previa
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- BMI >40
- Other

45

POST SURGERY OUTCOMES- MOMS TRIAL

- Reduced need for ventricular shunting
- Reduced incidence/severity of neurological effects
 - Impaired motor function
 - Sensory function of legs
- Improved mobility and chances child able to walk independently
- Reversed hindbrain herniation component of Arnold Chiari II malformation

MOMS2

- Most able to walk by school age
- Better fine/gross motor skills
- Better control of bladder and bowels

46

RESOURCES

- Volpe, N., Dall'Asta, A., Di Pasquo, E., Frusca, T. and Ghi, T. (2021), First-trimester fetal neurosonography: technique and diagnostic potential. *Ultrasound Obstet Gynecol*, 57: 204-214
- Children's Hospital of Philadelphia
 - <https://gps.chop.edu/condition/spina-bifida-myelomeningocele>

47

THANK YOU

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