POSTERIOR FOSSA

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Objectives

Part I – The cerebellum

Part II – The cisterna magna

Part I
The cerebellum

Distinguish the normal and abnormal embryology of the cerebellum, fourth ventricle, and cisterna magna

Identify the presence or absence of the key vermian landmarks and be able to apply biometric criteria, to determine if the vermis is morphologically normal

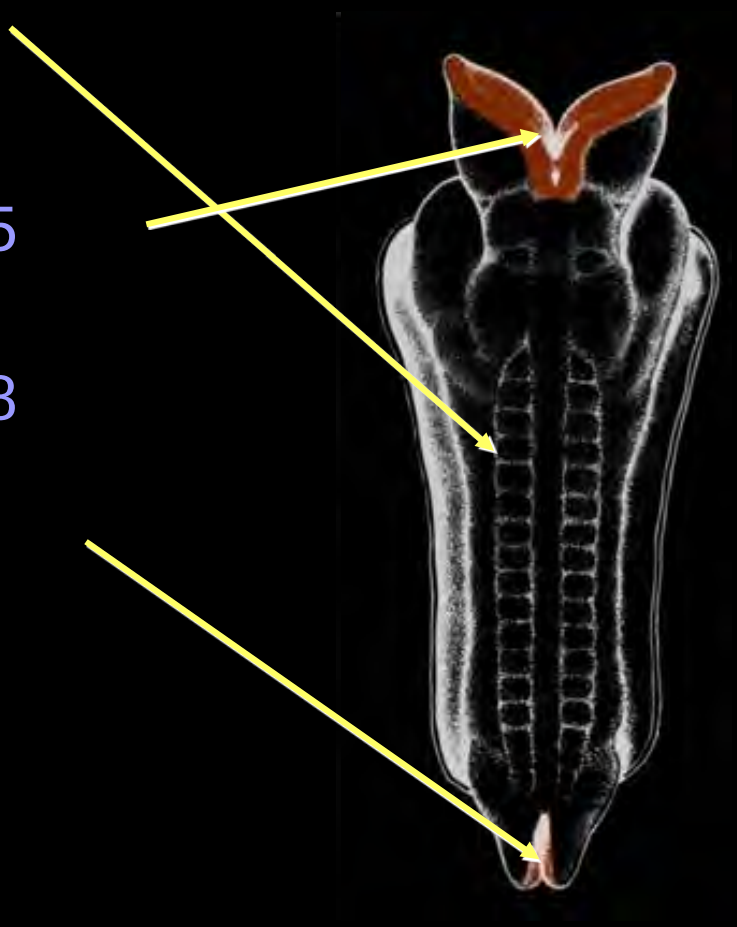
Identify the serious vermian pathologies and establish how to differentiate them from those with a potentially good prognosis

The neural tube: Embryology

- Neural tube develops in 3rd & 4th weeks of embryogenesis
- Longitudinal groove along dorsum of trilaminar germ cell disc
- Groove deepens until edges meet
- Edges fuse over the top

The neural tube: Embryology

- Fusion starts in the middle and works out
- This forms a hollow tube
- Cranial neuropore closes at 25 days
- Caudal neuropore closes at 28 days



The neural tube: Embryology

At its cranial end the neural tube forms three brain vesicles

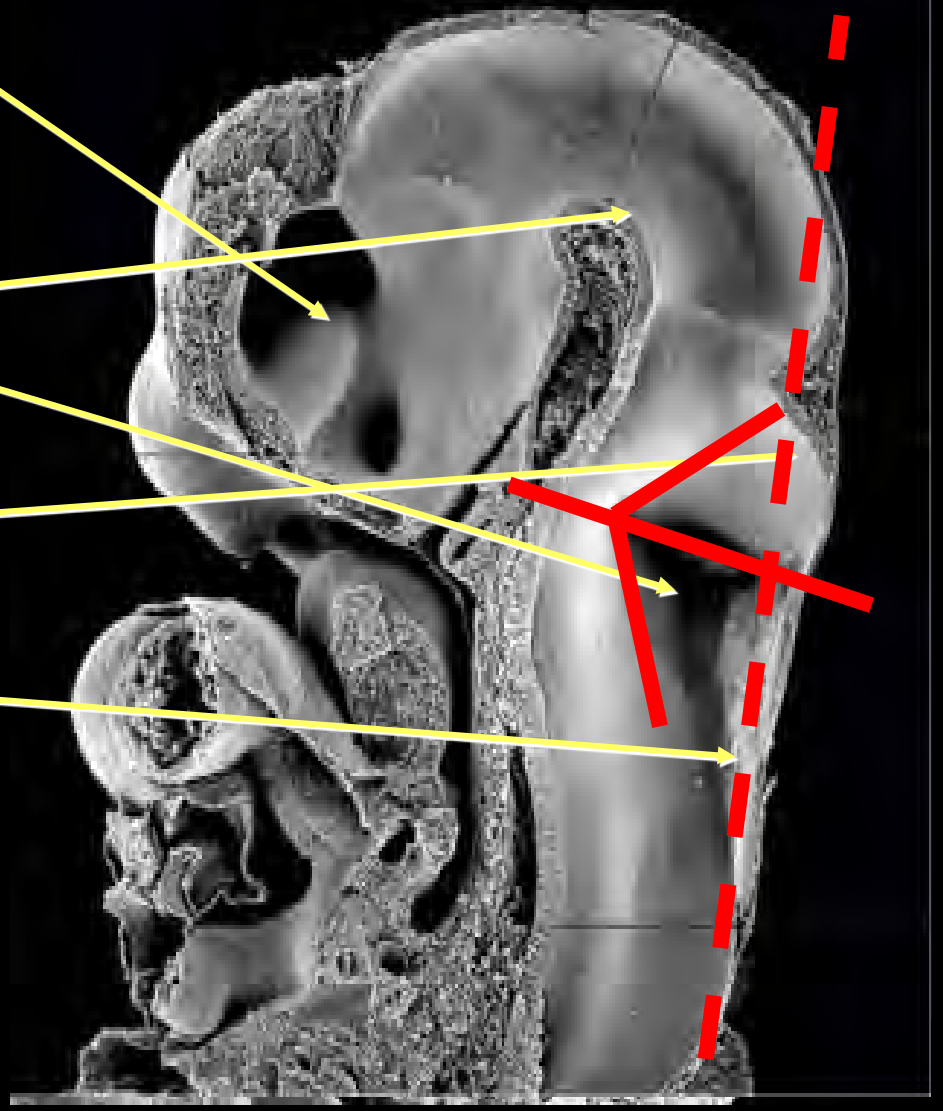
Procencephalon

- Telencephalon
- Diencephalon

Mesencephalon

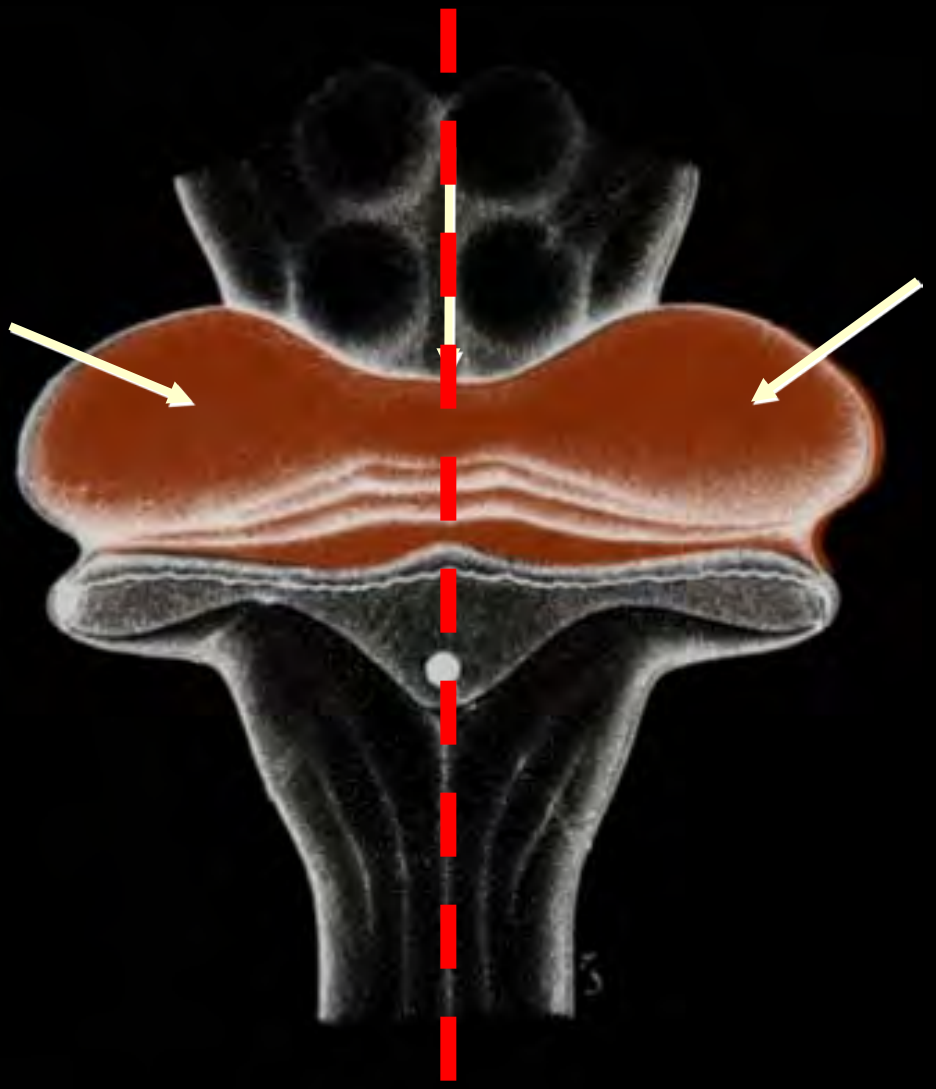
Rhombencephalon

- Metencephalon
  - Cerebellum
- Myelencephalon
  - Medulla Oblongata



The cerebellum: Early development

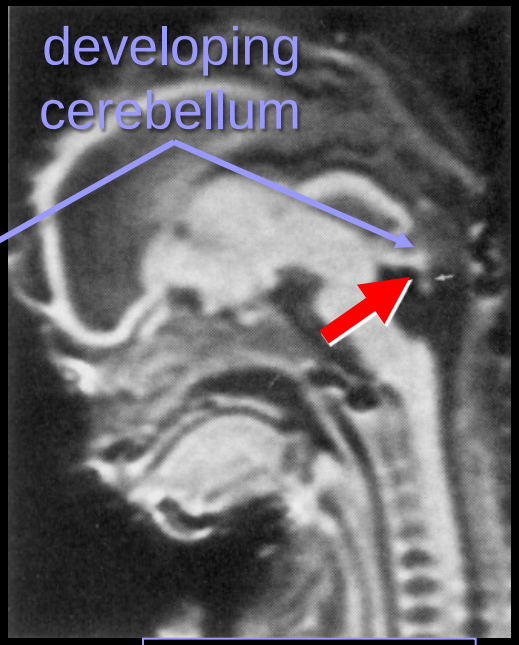
- Rhombic lip forms across midline under influence of isthmic organiser and various genetic factors
- Cerebellar hemispheres grow laterally
- Vermis grows through proliferation of midline tissue



The cerebellum: Progression of vermian development

11-12 weeks

Cerebellar plate enlarges



13-14 weeks

Fastigial crease develops along ventral surface of cerebellar plate

16 weeks

Cerebellar plate grows caudally and covers roof of 4th ventricle

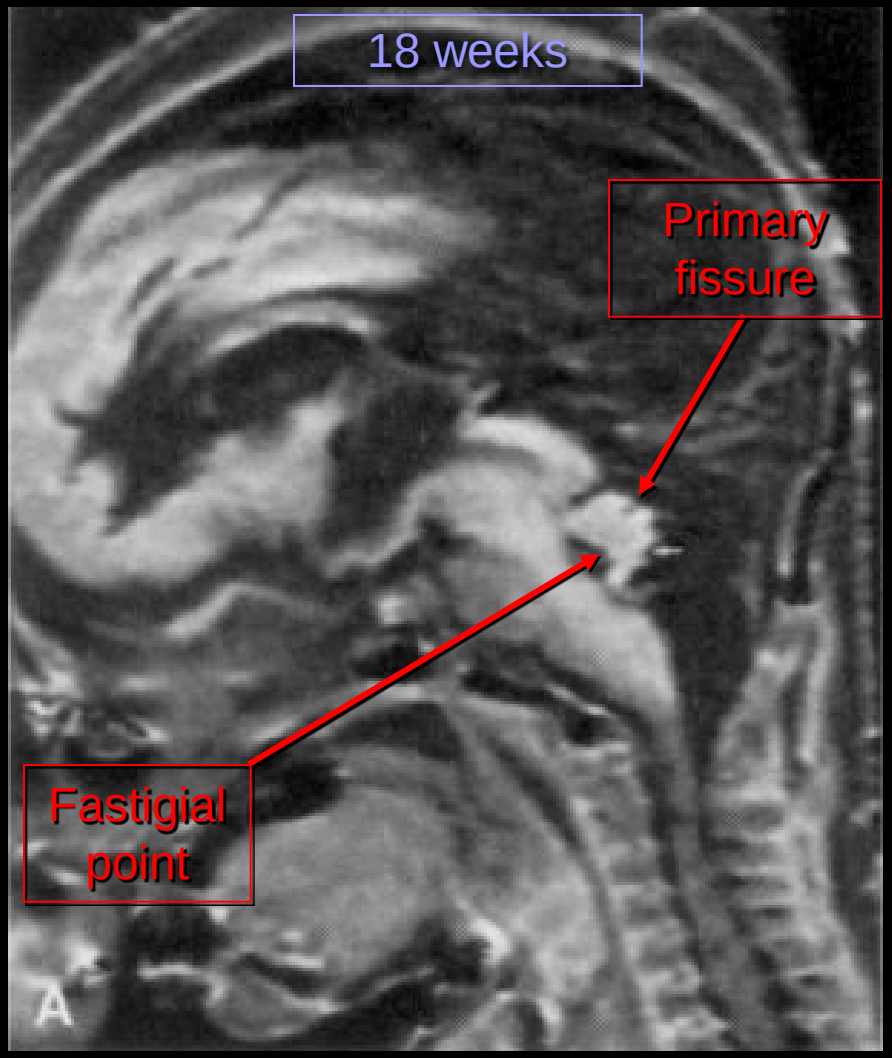
Chong et al. MR on fetal specimens

The cerebellum:

By 18 weeks there should usually be:

- •fastigial point
- •primary fissure
- •covered 4th ventricle roof

–may be physiologic delay as late as 24 weeks



Analysis

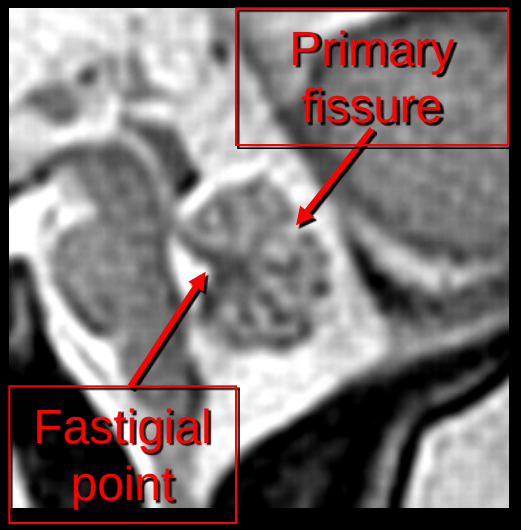
- Analysis includes:

Assessment for presence of a vermis

- identification of fastigial point / crease
- Identification of primary fissure

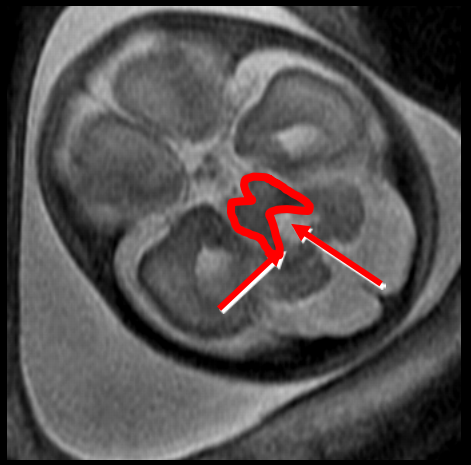
Assessment for maturity of the vermis

- cranio-caudal diameter
- ratio ‘above:below’ fastigium-declive line
- tegmento-vermian angle (“closure” of 4th V)
- vermian lobulation



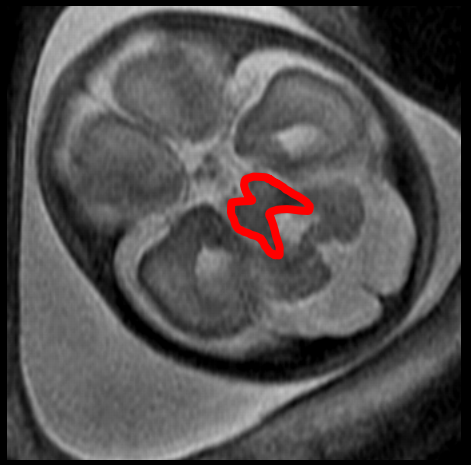
Joubert syndrome

- No fastigial point
- No primary fissure
- 4th ventricle wide & “bat wing” shape
- Tegmental cleft
- “Molar tooth” configuration of brainstem



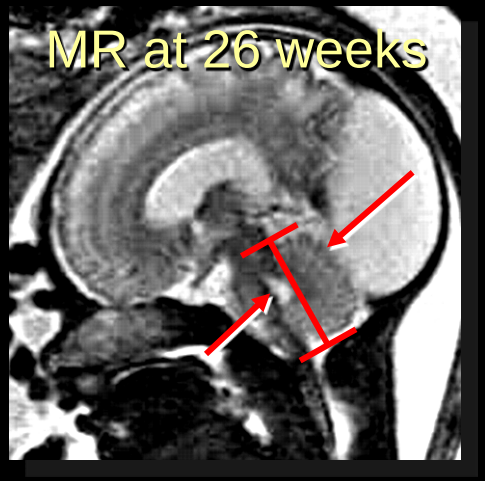
Joubert syndrome

- Anatomically
  - –Near total aplasia of vermis
  - –Hemispheres appose each other in midline
  - –Dysplasia and heterotopia of cerebellar nuclei
- Clinically
  - –Episodic hyperpnea
  - –Abnormal eye movements
  - –Ataxia
  - –Mental retardation



Rhombencephalosynapsis

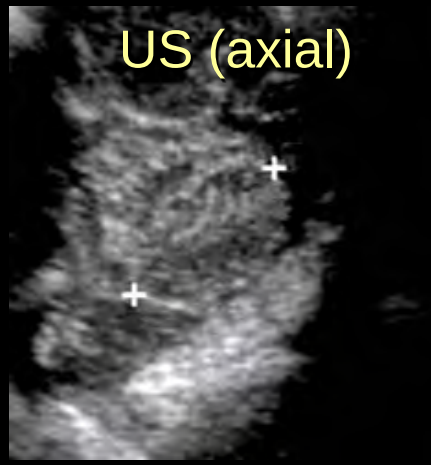
- Fastigial point rounded
- No primary fissure
- Craniocaudal diameter too big
  - –not measuring true vermis
- Folia continuous across midline



- Multiple other anomalies

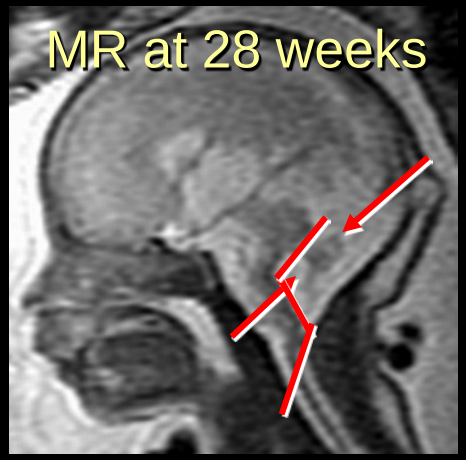
Rhombencephalosynapsis

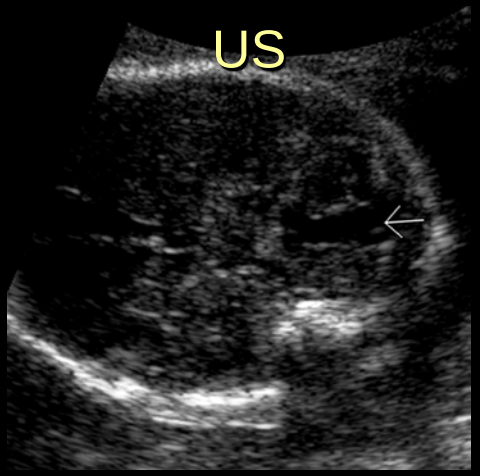
- Anatomically
  - –Fastigial nuclei apposed to midline
  - –Associated with other supratentorial abnormalities most frequently in holoprosencephaly spectrum
- Severity of these other abnormalities determines prognosis



Congenital Muscular Dystrophy

- No fastigial point
- No primary fissure
- Z-shaped brainstem





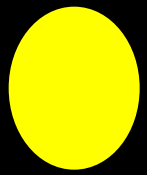
Congenital Muscular Dystrophy (Walker-Warburg)

- Anatomically
  - –Z-shaped brainstem
  - –Cobblestone lissencephaly
  - –Occipital cephalocele
  - –ocular asymmetry
- Clinically
  - –dismal prognosis

11-12 weeks

Microlissencephaly

- No fastigial point
- No primary fissure
- Z-shaped brainstem
- “Shell-like” hemispheres



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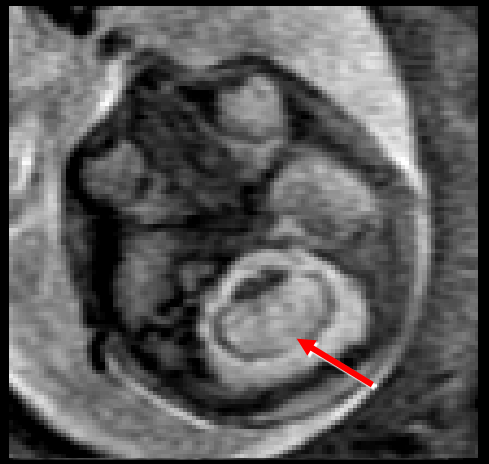
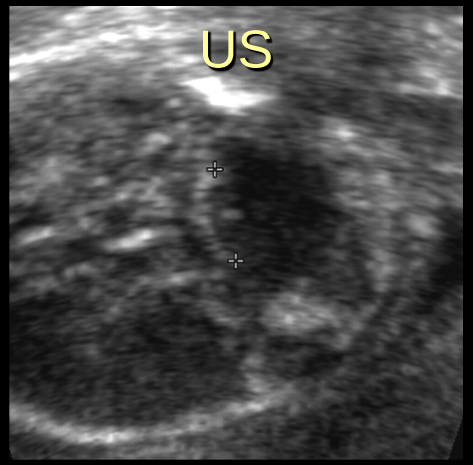


- Fetal demise

Microlissencephaly

- Anatomically
  - –Microcephaly + lissencephaly = microlissencephaly
  - –Z-shaped brainstem
- Clinically
  - –dismal prognosis

13-14 weeks



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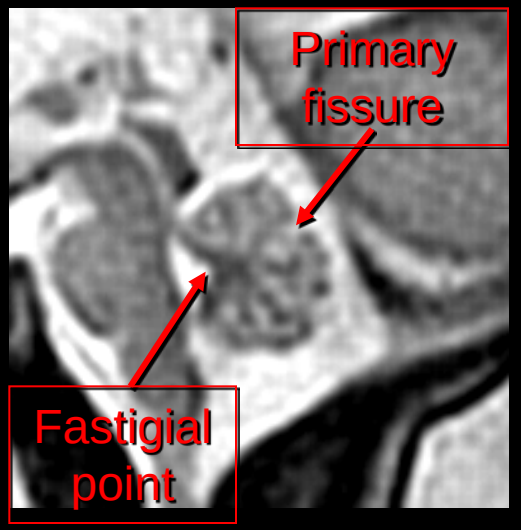
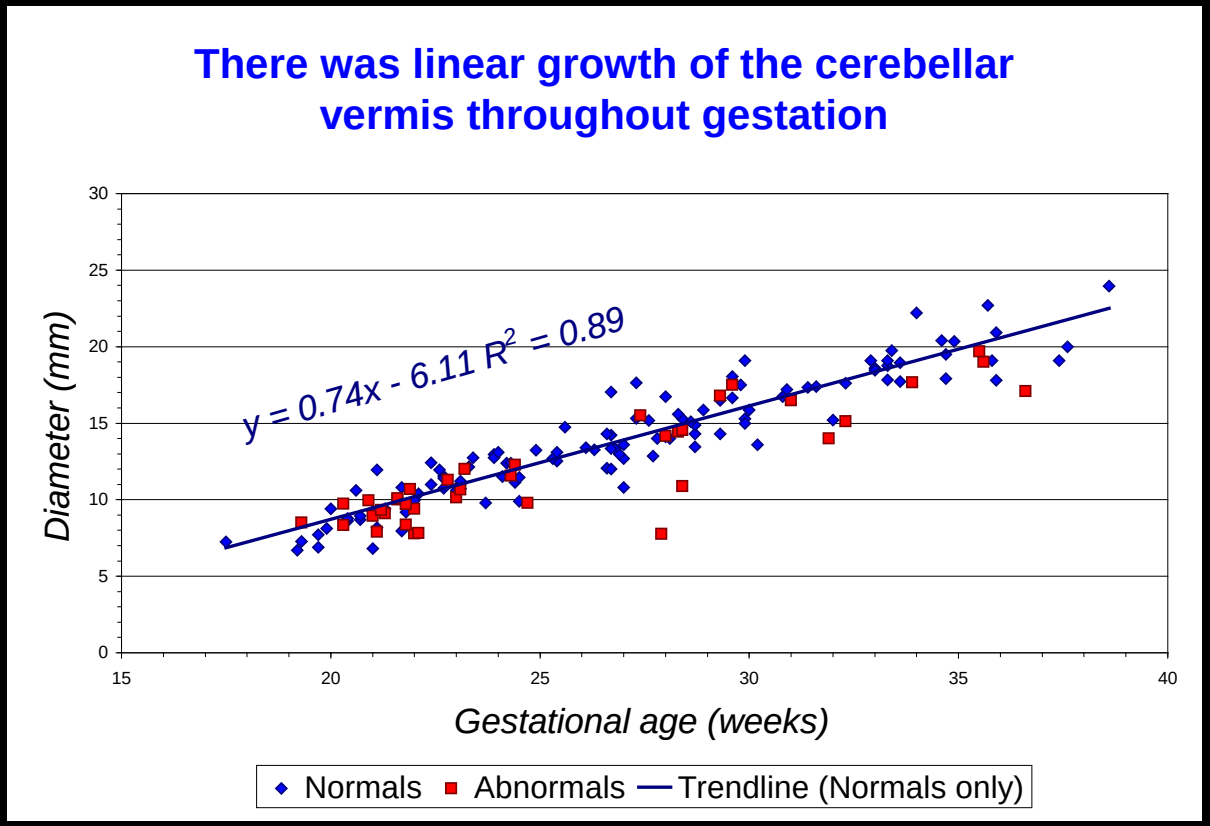
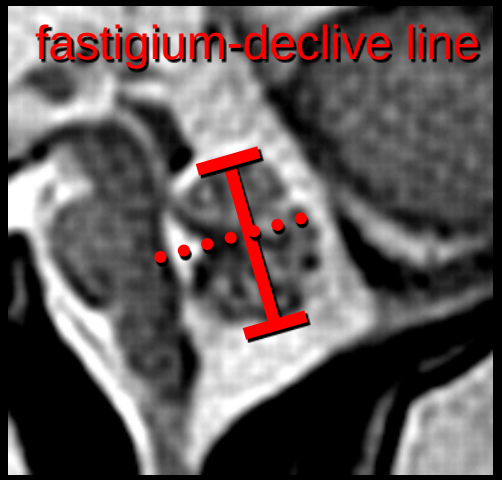


Assess: Growth

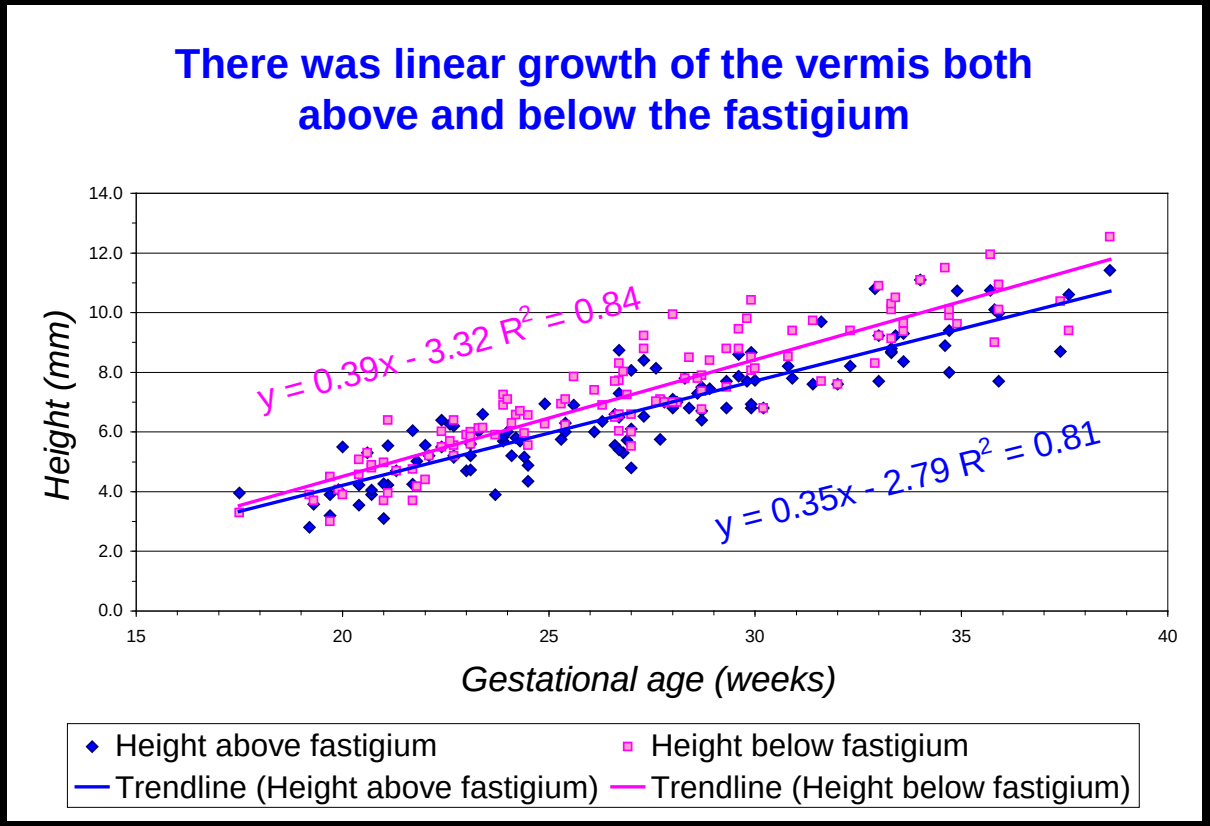
- Absolute growth – craniocaudal
- allows measurement independent of abnormal angulation

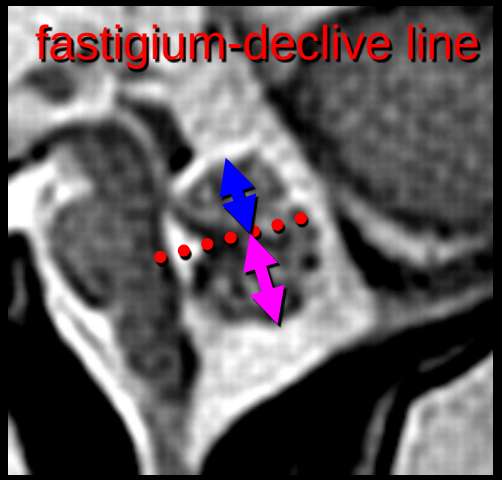




Assess: Growth

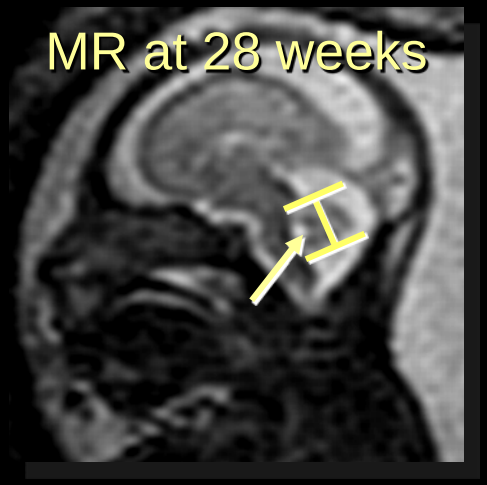
- Relative growth – ‘above:below’ fastigium

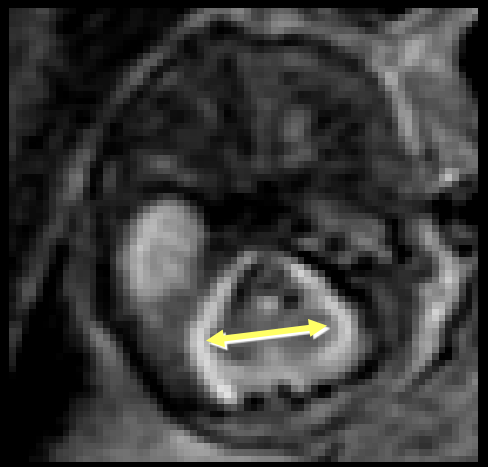




•Severe micrencephaly: Cerebellar hypoplasia

- Fastigial point present
- Small craniocaudal diameter
- Small transcerebellar diameter





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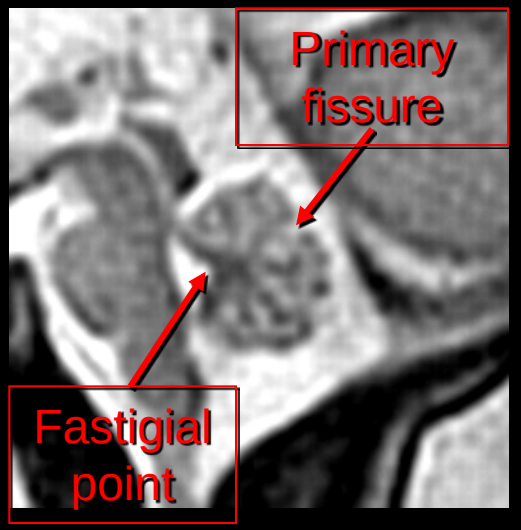
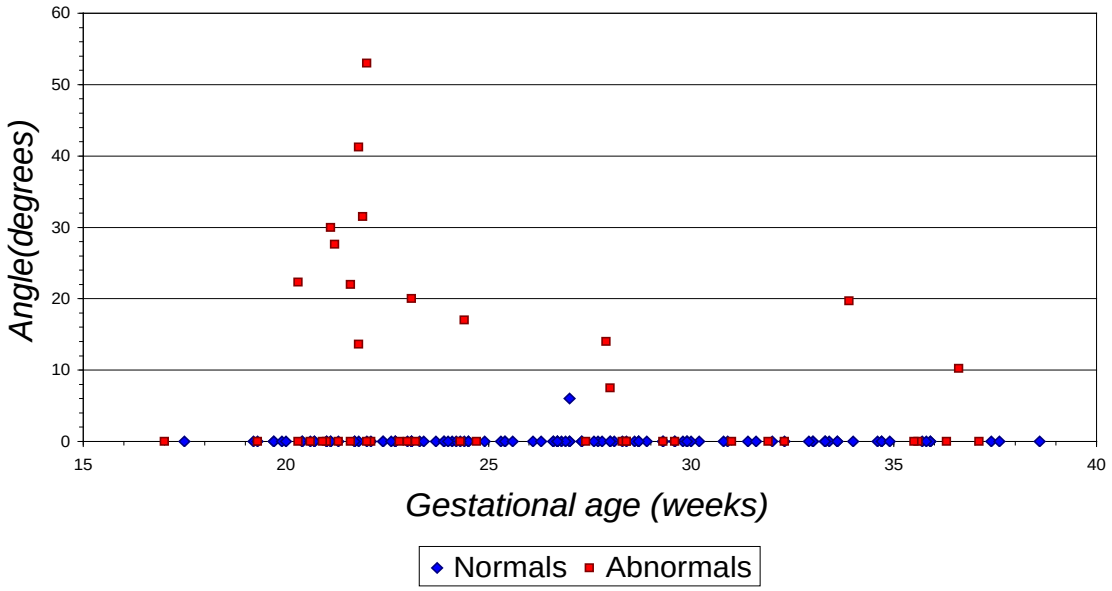
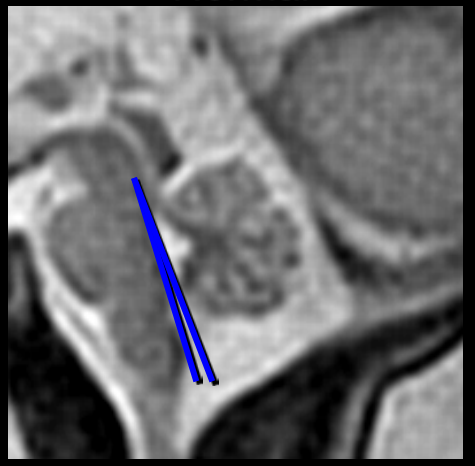


Assess:
Tegmento-vermian angle

A slightly increased tegmento-vermian angle may be a normal variant

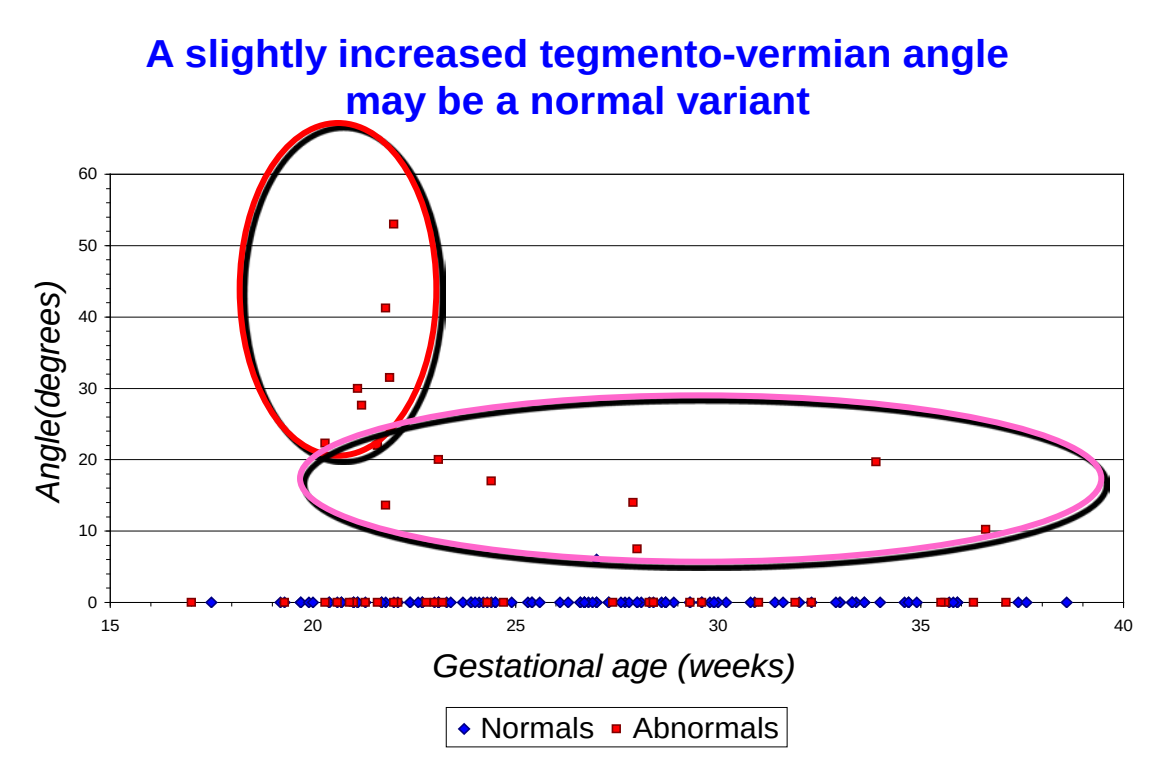


Normal



Abnormal

Assess:
Tegmento-vermian angle



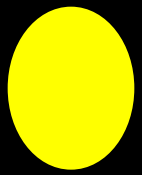
Large angles always associated with vermian hypoplasia

Intermediate angles not necessarily pathologic

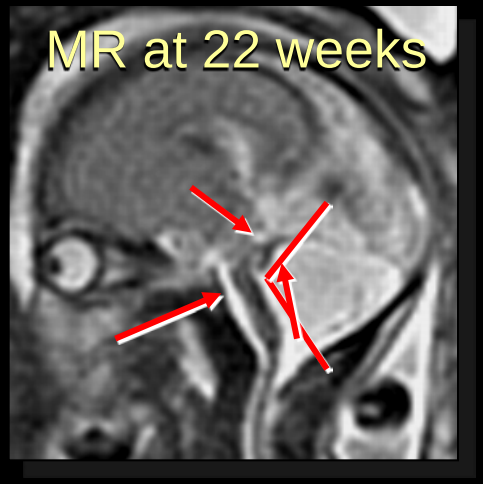
Recently shown that angulated but morphologically normal vermis can have normal outcome

Dandy-Walker Continuum "Plus"

- Tegmentovermian angle markedly increased
- Vermis very small
- No primary fissure
- No fastigial point
- Pontine hypoplasia
- Small hemispheres



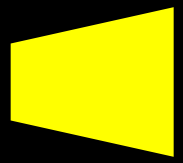
ovoid



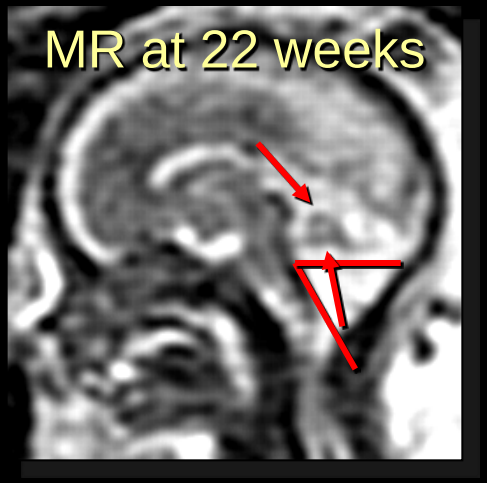
- Death from respiratory failure

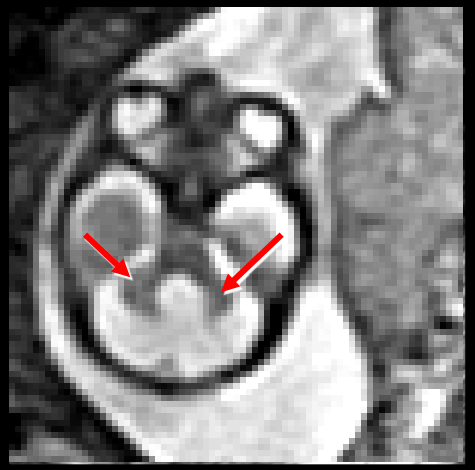
Dandy-Walker Continuum
"Classic"

- Tegmentovermian angle increased
- Vermis small
- No primary fissure
- Fastigial point flattened
- Larger hemispheres



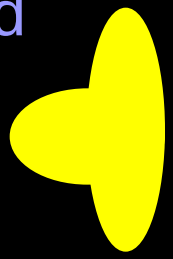
trapezoid



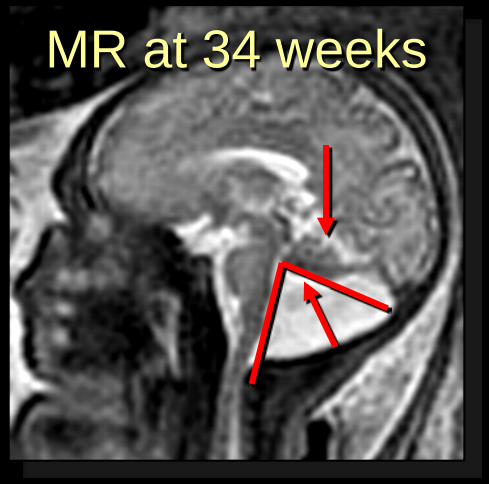


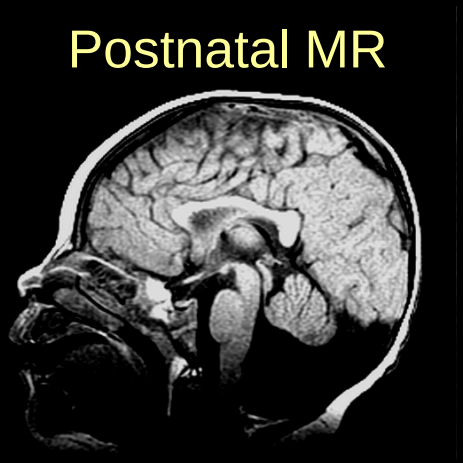
Dandy-Walker Continuum "minus"

- Tegmentovermian angle moderately increased
- Vermis less dysplastic
- Primary fissure seen
- Fastigial point less flattened
- Even larger hemispheres
- Good vermian lobulation



trefoil

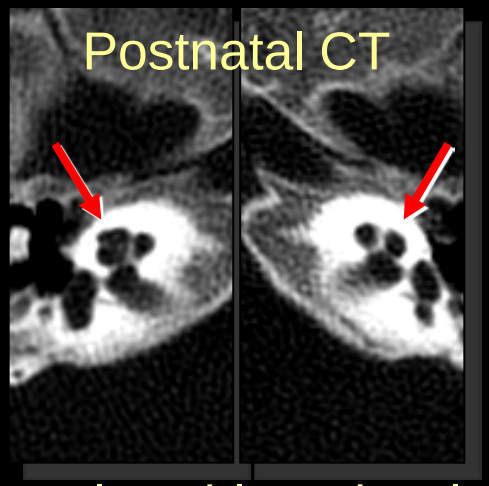




- Deaf, but otherwise normal development

Cerebellar & labyrinthine anomalies often coexist

- *Rhombic lip forms cerebellum and cochlear nucleus*
- *Rhombencephalon induces otocyst*

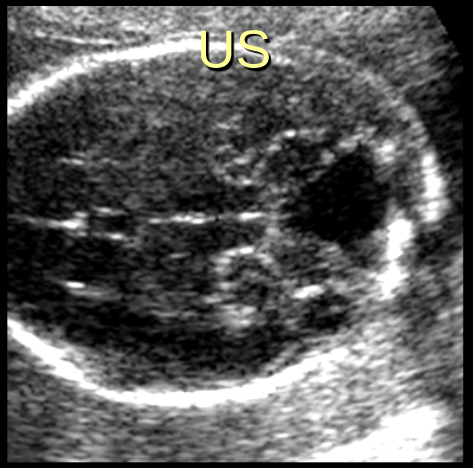


Bilateral cochlear dysplasia

- Deaf, but otherwise normal development

Dandy-Walker Continuum

- Anatomically:
  - –High tentorium
  - –Cerebellar hypoplasia
  - –Enlarged posterior fossa
  - –(hydrocephalus)
- Clinically can have normal intelligence
- depends on:
  - –degree of vermian hypoplasia/dysplasia
  - –plus associated abnormalities
    - Structural (both brain and body)
    - Genetic – Walker-Warburg, Meckel-Gruber
    - Chromosomal - trisomies
- *Counselling extremely difficult*



Analysis

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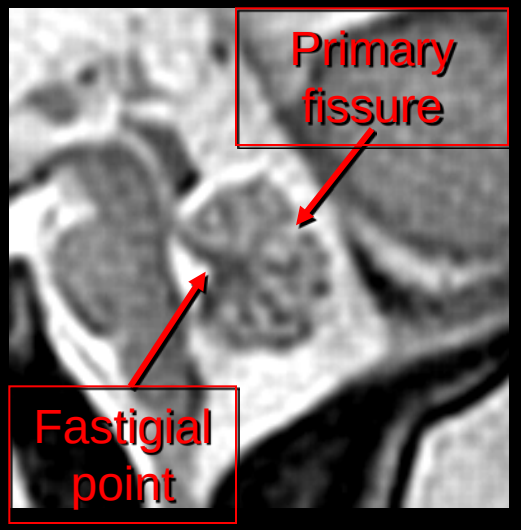
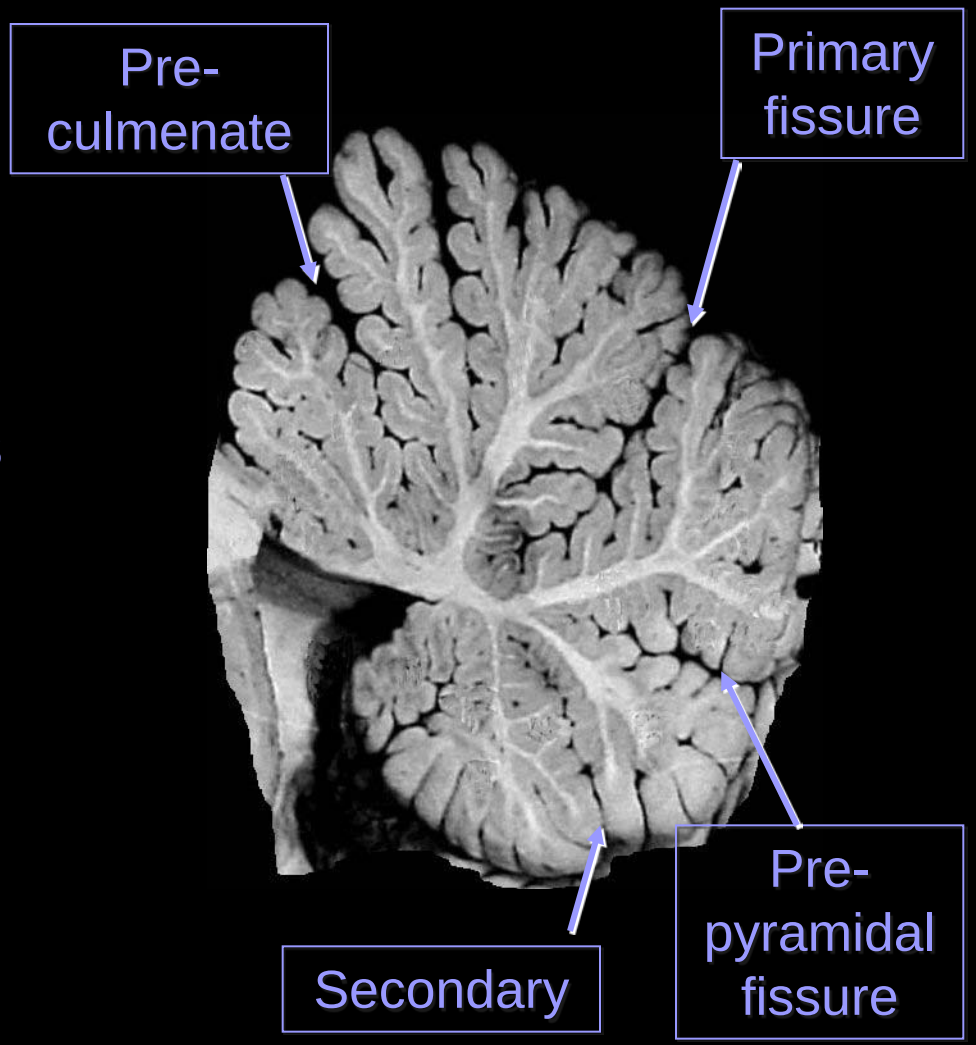
- tegmento-vermian angle (“closure” of 4th V)
- vermian lobulation



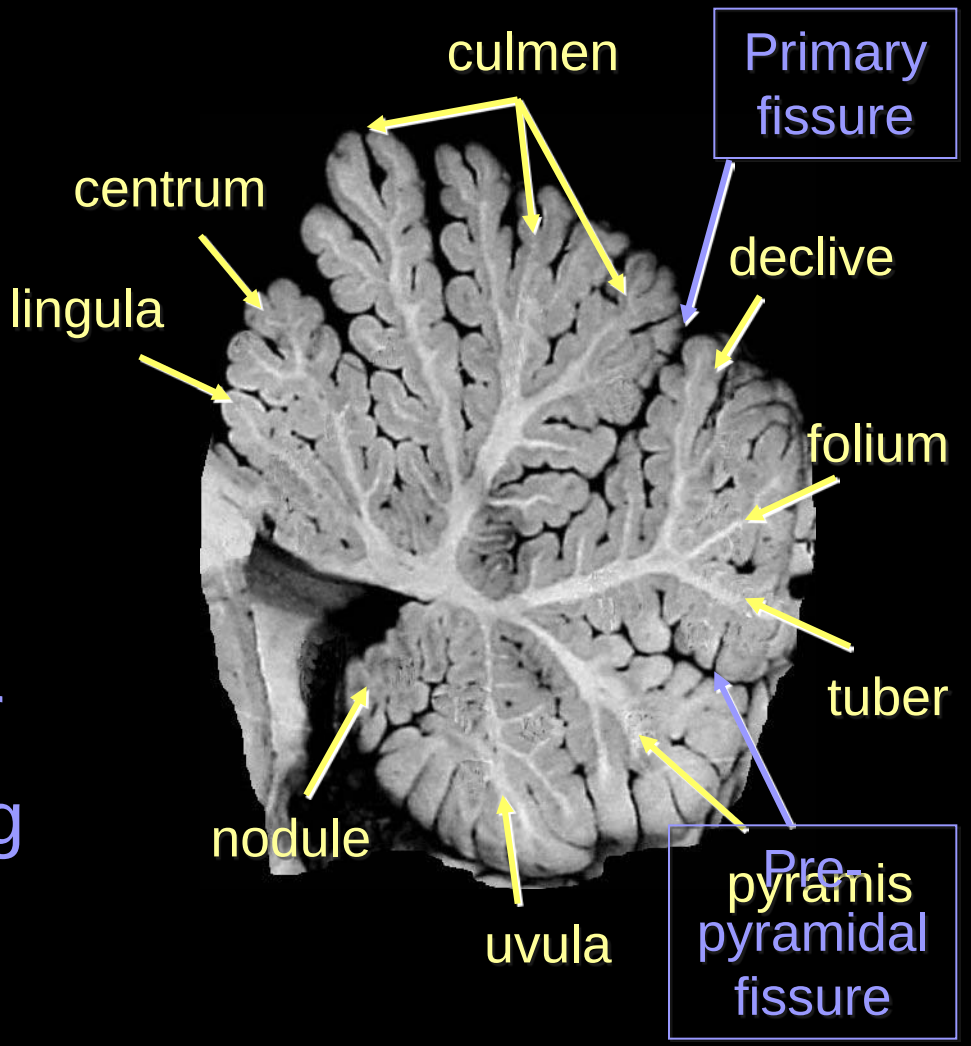
Assess: lobulation

- Fissures appear as follows:
  - –Primary – 17.5 weeks
  - –Secondary – 20 weeks
  - –Pre-pyramidal – 21 weeks
  - –Pre-culmenate – 22 weeks



Assess: lobulation

- 9 lobules present on sagittal section
- Earliest all seen 27 weeks
- 3 lobules between primary and pre-pyramidal fissure
- These are last to appear
- May lead to mis-counting of vermian lobules on early imaging



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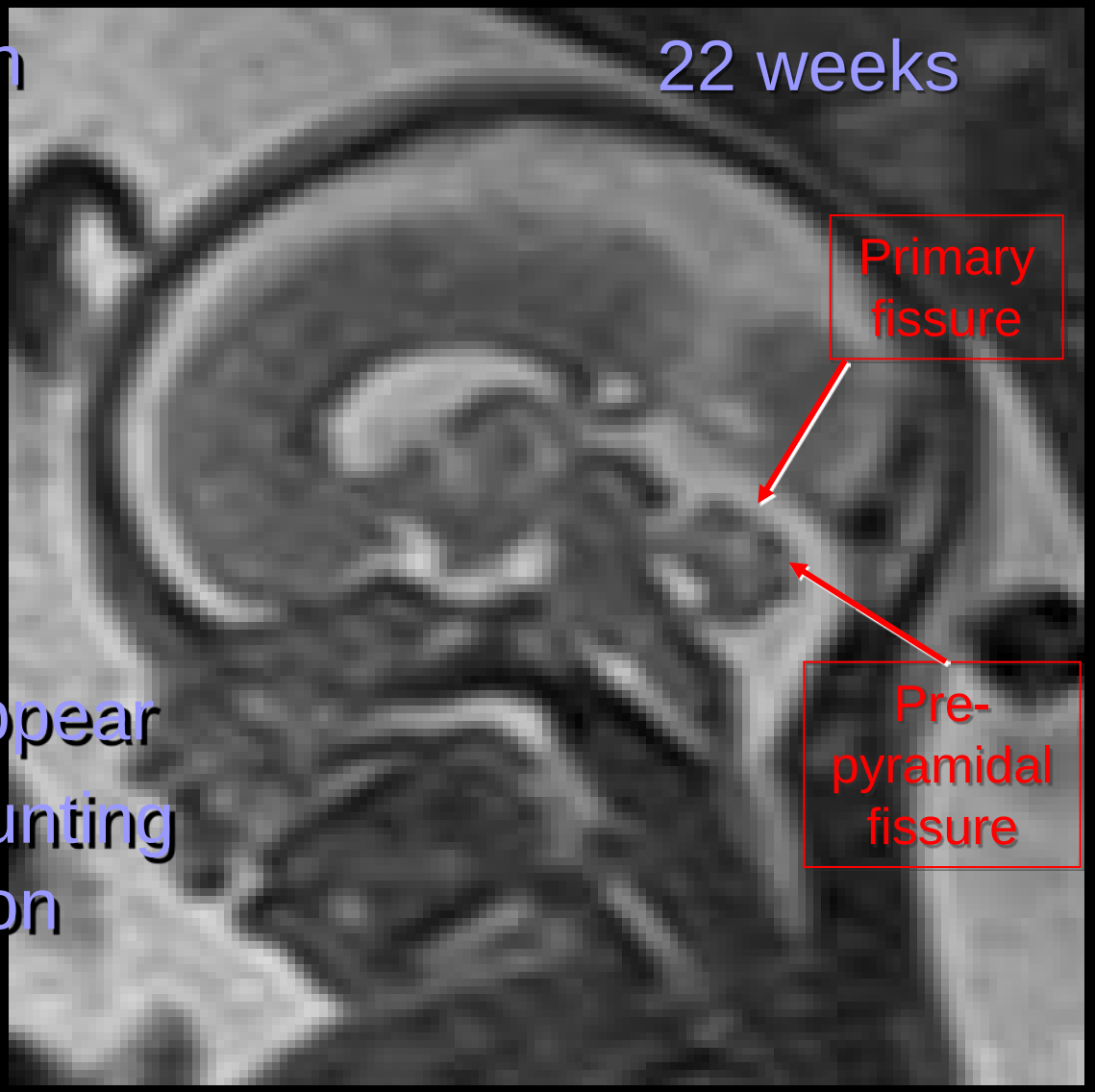

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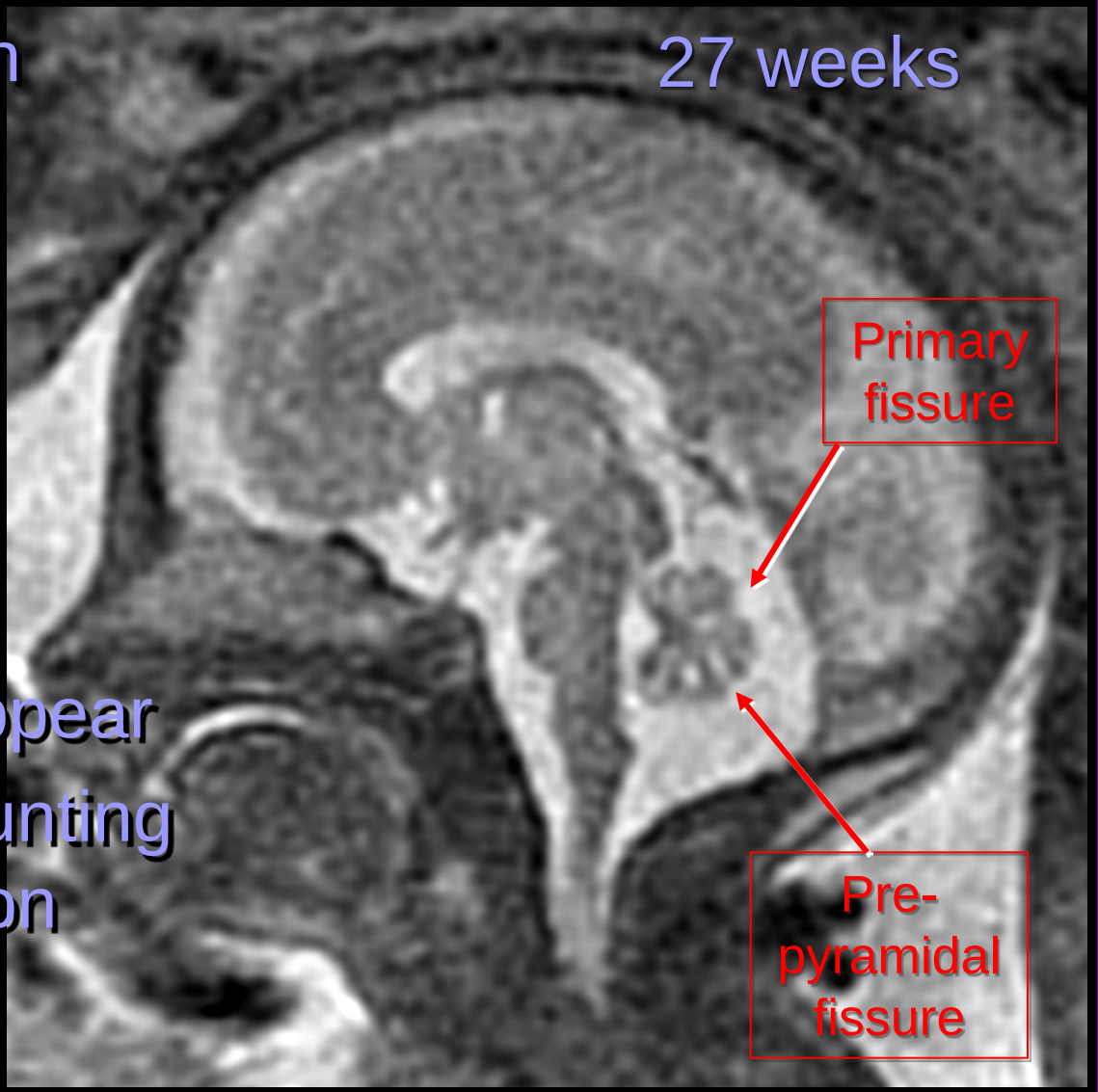


Summary

- Assessment of vermian morphology is key
  - –Including presence of fastigial point and primary fissure
  - –Absolute and relative cranio-caudal growth
- Vermis should “close” over the 4th ventricle by 18 weeks but can be as late as 24 weeks
- A large tegmentovermian angle is always associated with vermian hypoplasia
- Vermis grows through proliferation of mesial primordium not through midline “fusion” of the cerebellar hemispheres
- All lobules not visible until 27 weeks at the earliest
- Better vermian lobulation correlates with a better prognosis

Part II
The cisterna magna

- Current theories suggest that
  - Dandy-Walker malformation
  - Mega cisterna magna
  - Blake's pouch cyst
- Are variants within the Dandy-Walker spectrum

Objectives

- To support and advance this theory by demonstrating:
  - –normal and abnormal anatomy at various stages of development
  - –by pre- and post- natal ultrasound
  - –with fetal MRI and neonatal CT correlation where available

Sonoembryology

20 weeks

- By mid-trimester, linear echoes are almost always seen in the fetal and neonatal cisterna magna
- variously described as:
  - –the straight sinus
    - Mahony BS, Callen PW, Filly RA, et al. The fetal cisterna magna. Radiology. 1984 Dec;153(3): 773-6
  - –the Torcula Herophili
    - Pilu G, Romero R, De Palma L, et al. Ultrasound investigation of the posterior fossa in the fetus. Am J Perinatol. 1987 Apr;4(2): 155-9
  - –dural folds - inferior attachments of falx cerebelli
    - Pretorius DH, Kallman CE, Grafe MR et al. Linear echoes in the fetal cisterna magna. J Ultrasound Med. 1992 Apr;11(4): 125-8
  - –bridging arachnoid septations
    - Knutzon RK, McGahan JP, Salamat MS, et al. Fetal cisterna magna septa: a normal anatomic finding. Radiology. 1991 Sep;180(3): 799-801
- Clearly there is disagreement about what they represent, even on pathologic studies

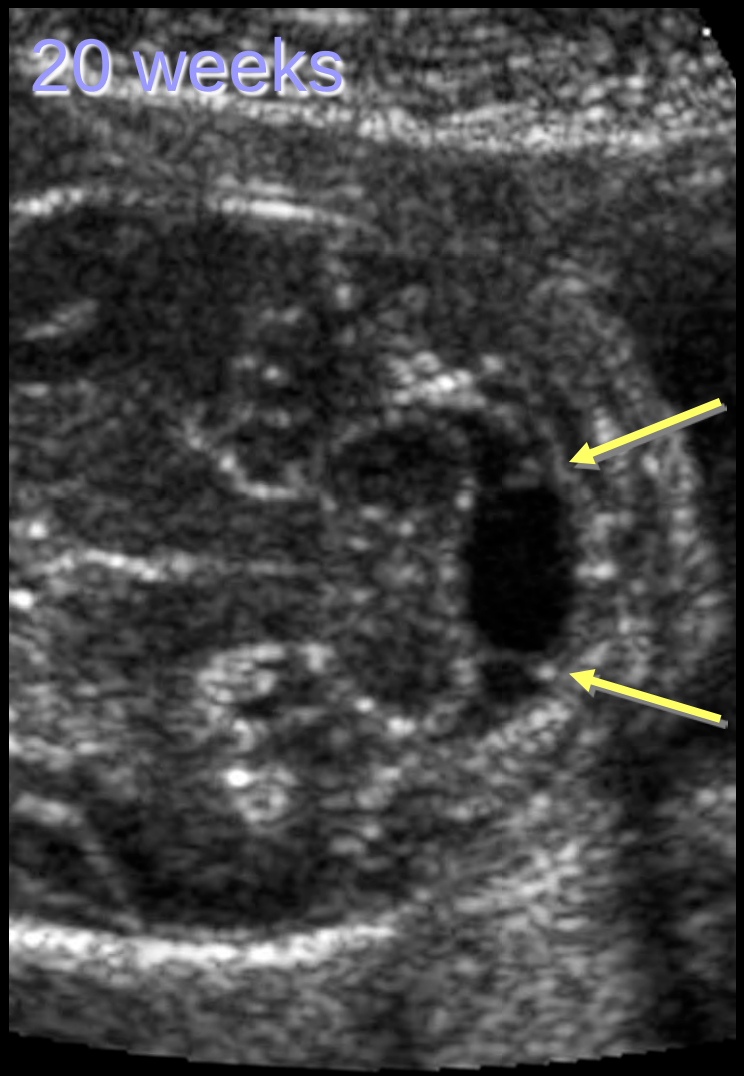
Sonoembryology

- Previous studies have shown:
  - –cisterna magna septa are seen
    - •in 84% to 92% of fetuses in second and third trimester
- Most often observed when the cisterna magna is over 3mm in antero-posterior diameter
- If not seen:
  - –technical or positional issue
  - –skull ossification

Pretorius DH, Kallman CE, Grafe MR et al. Linear echoes in the fetal cisterna magna. J Ultrasound Med. 1992 Apr;11(4): 125-8

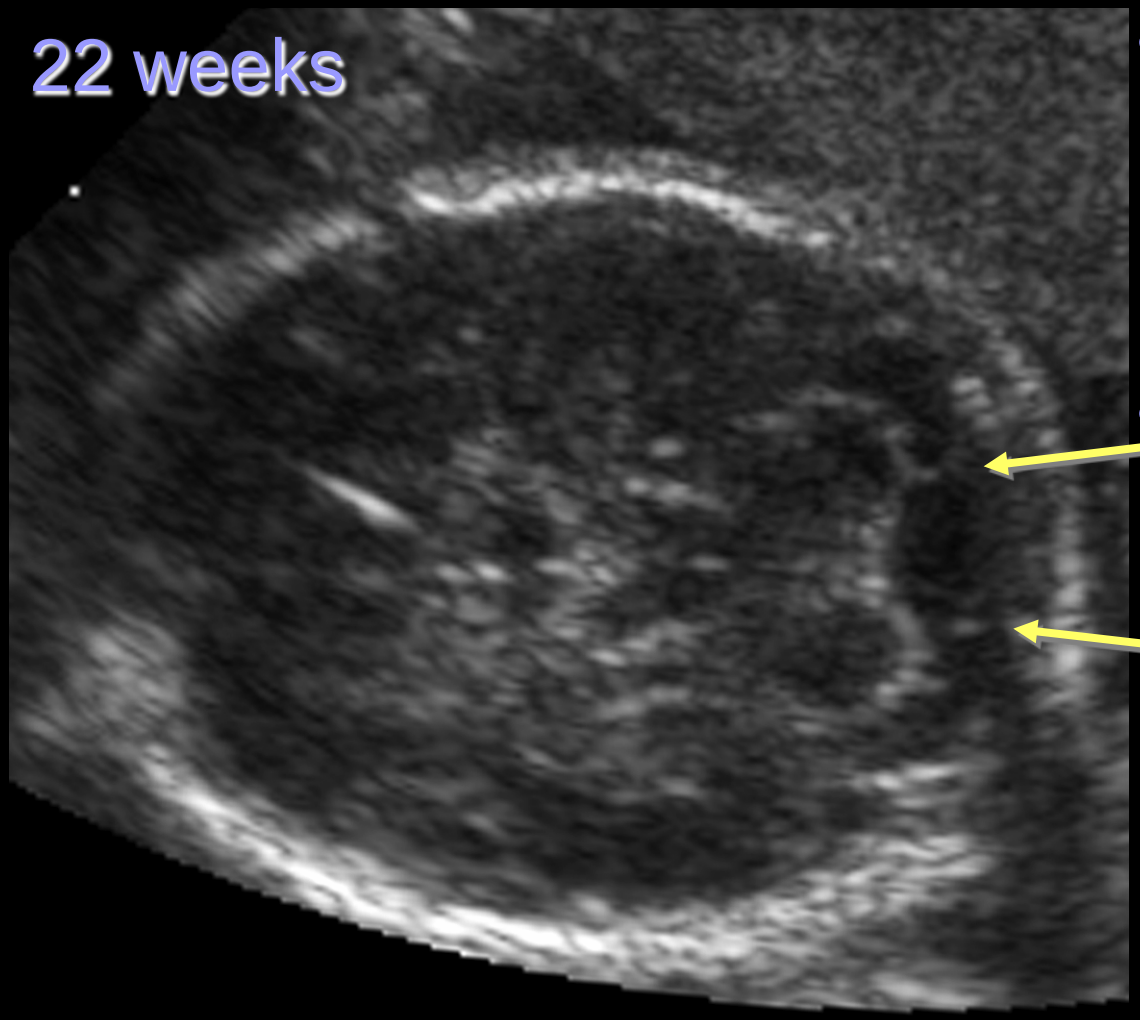
Knutzon RK, McGahan JP, Salamat MS, et al. Fetal cisterna magna septa: a normal anatomic finding. Radiology. 1991 Sep;180(3): 799-801

Sonoembryology



- The cisterna magna septa:
  - –infero-posterior to the cerebellar vermis
  - –usually straight, arising at the cerebellovermian junction
  - –course directly to the occipital bone

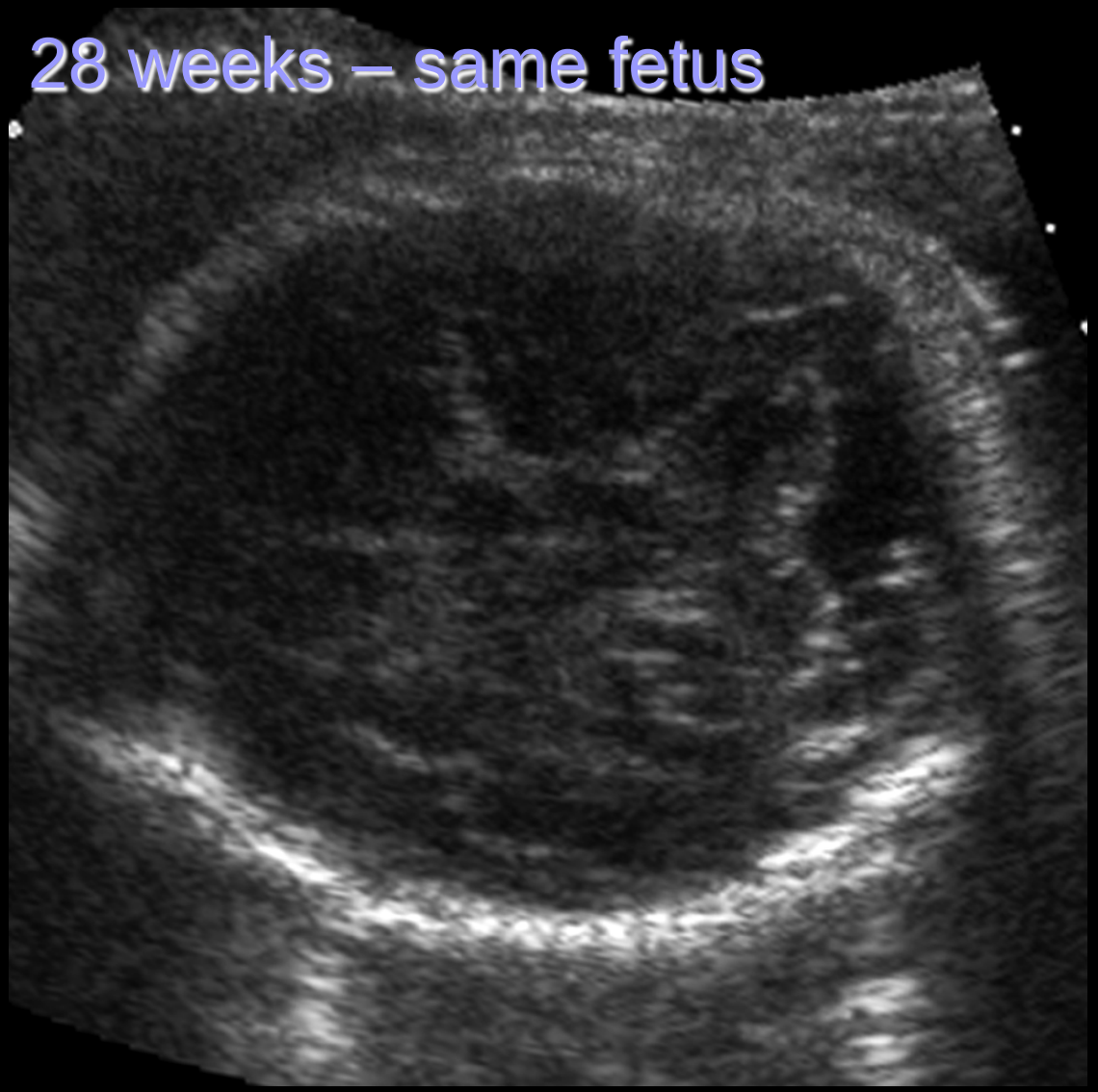
Sonoembryology



- Septa become more difficult to discern later in gestation
  - –even in the same fetus
- but the posterior fossa structures develop normally

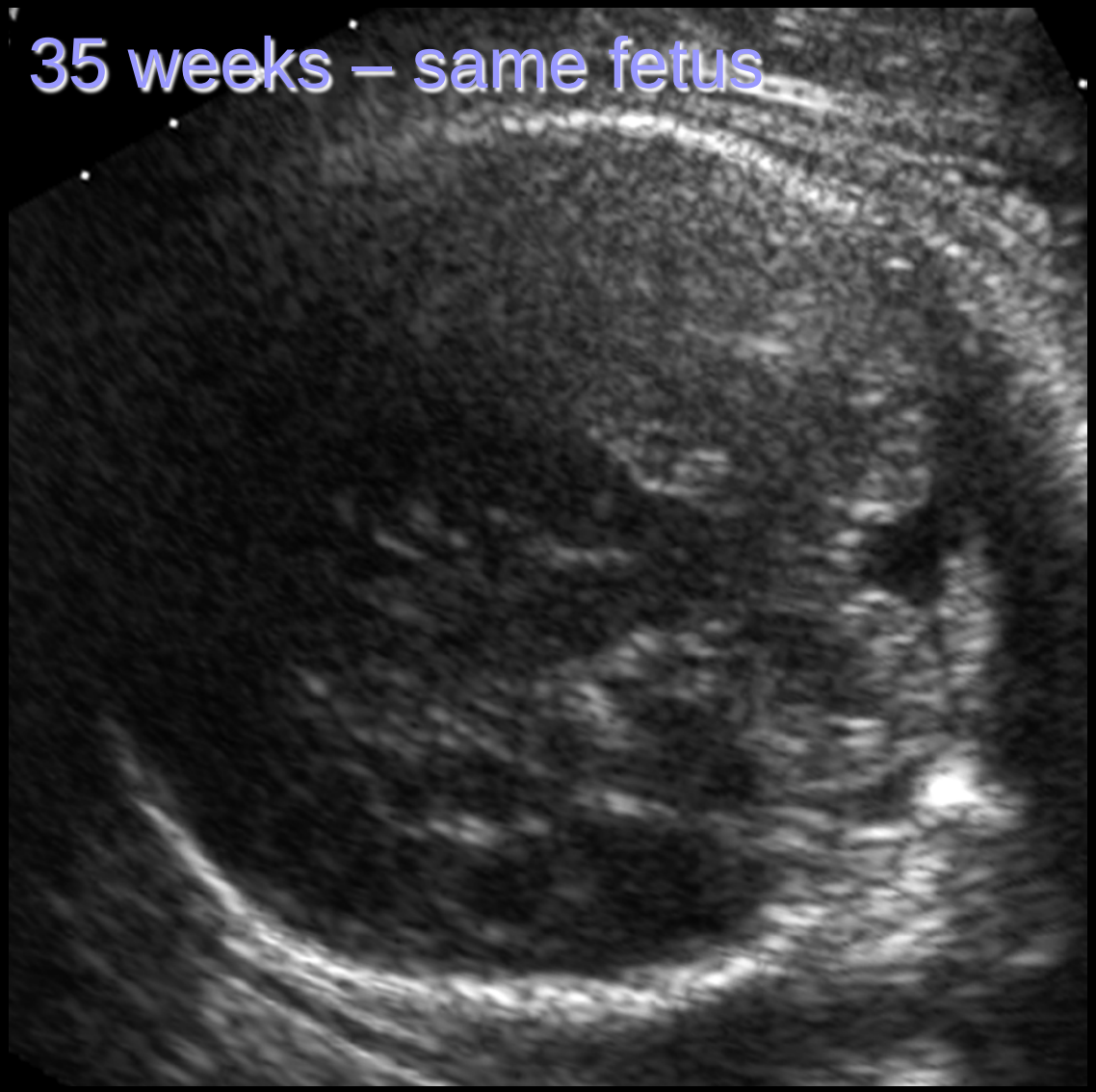
Sonoembryology

28 weeks – same fetus



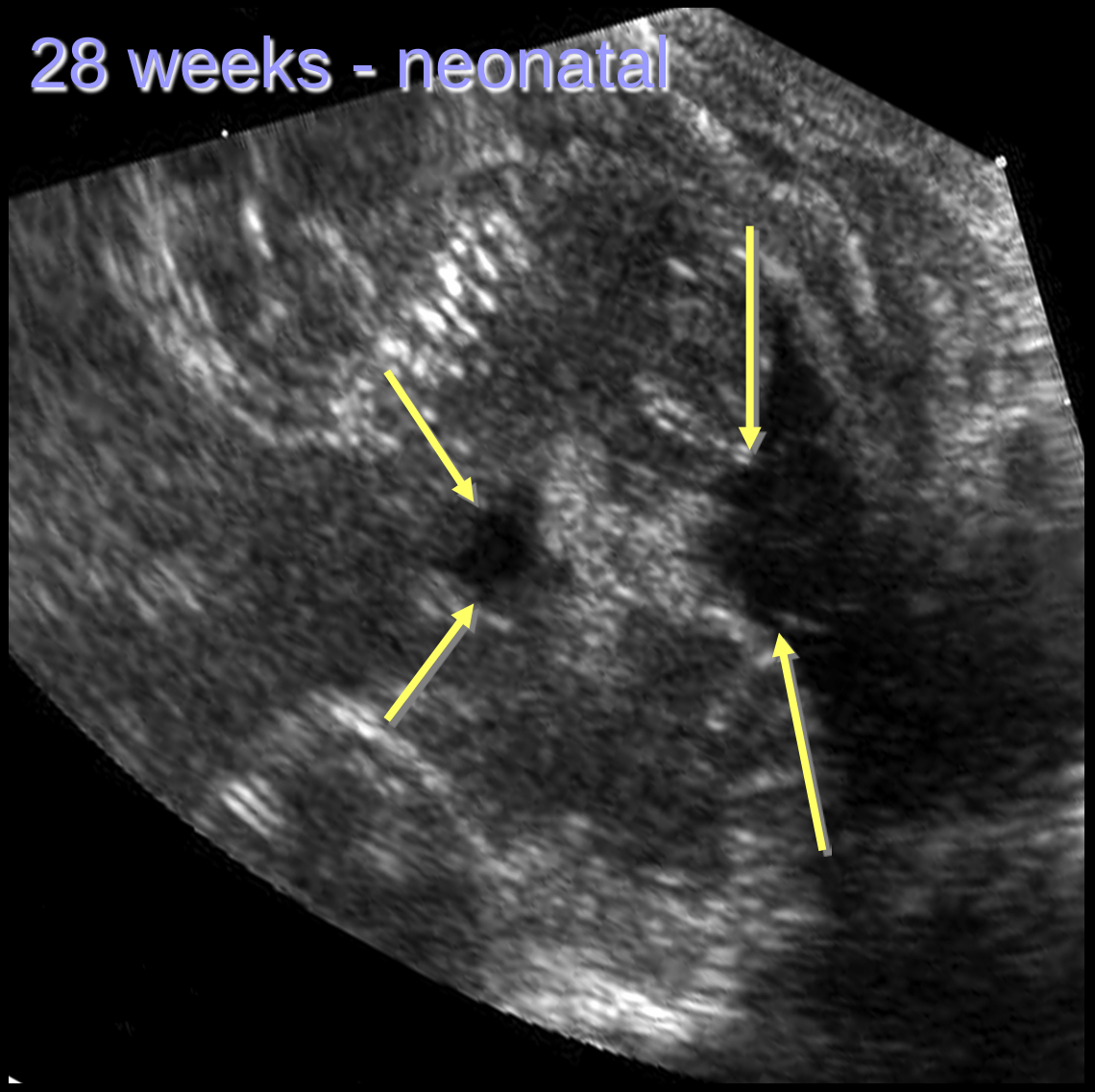
Septa less visible

Sonoembryology



- Septa not visible
- Visible structures appear normal

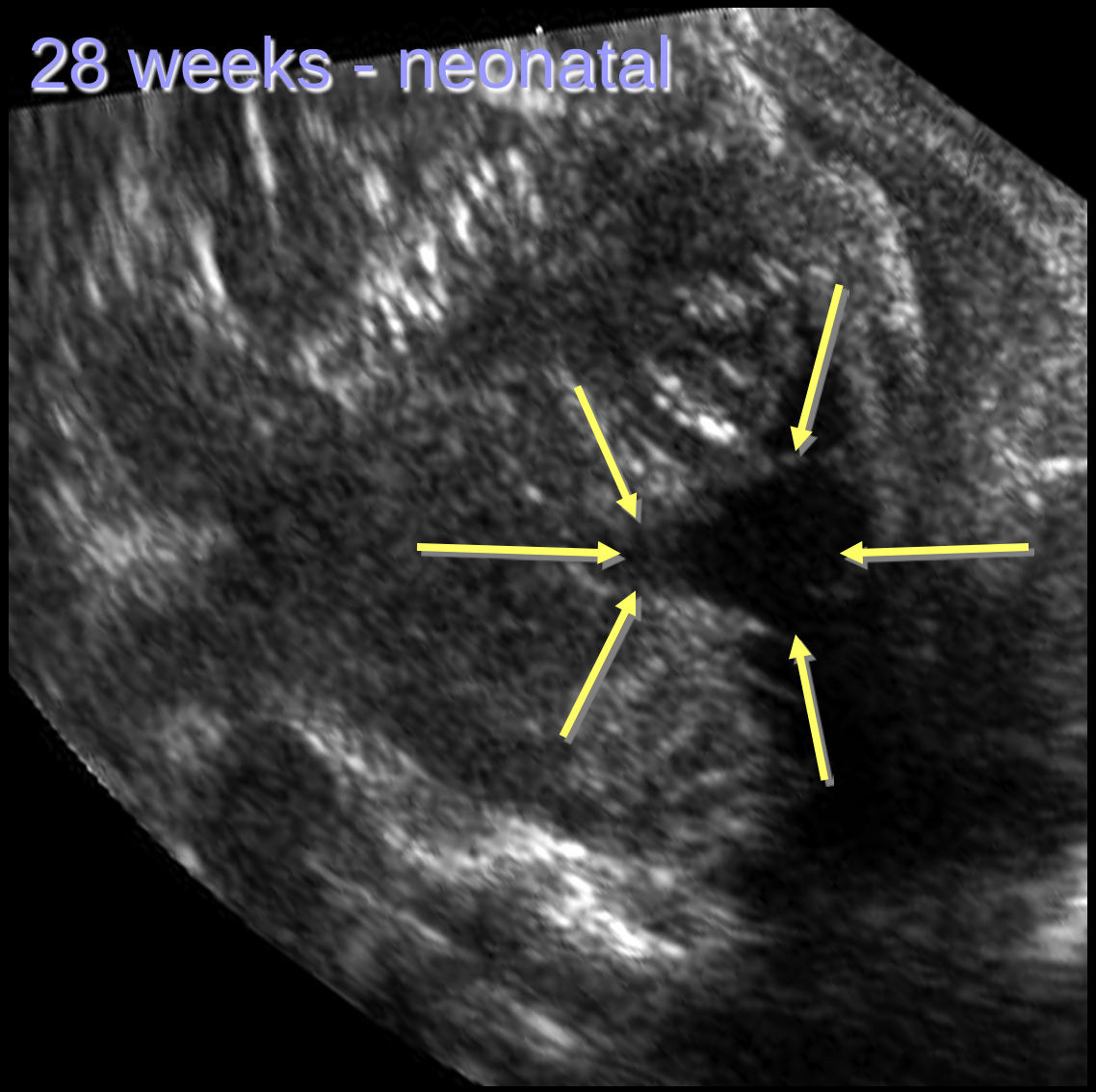
Sonoembryology



Above vallecula

4th ventricle walls separate from septa

Sonoembryology



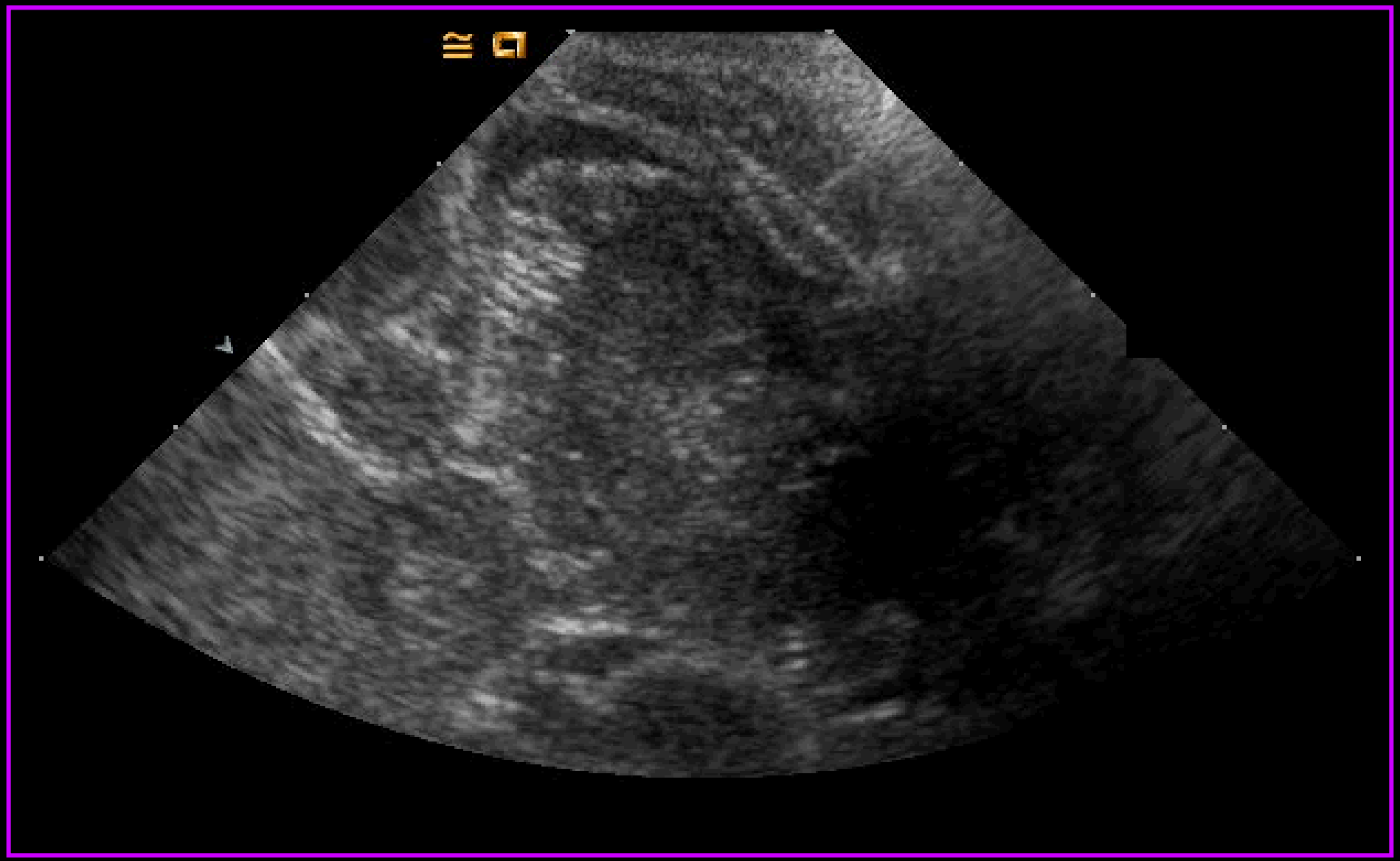
At vallecula

4th ventricle walls continuous with septa via vallecula

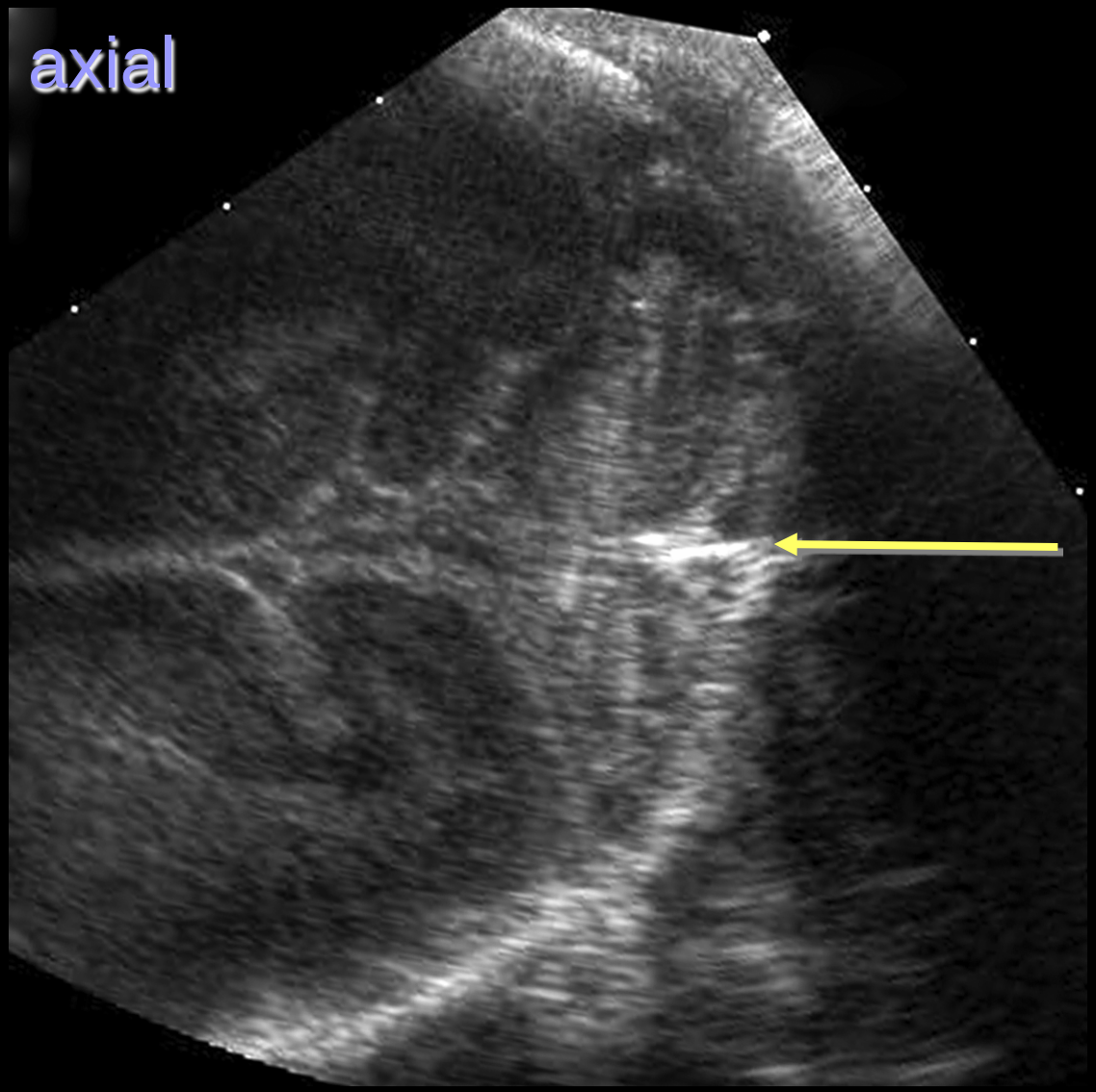
CSF space enclosed between septa

is in direct continuity with CSF space of the fourth ventricle

Sonoembryology

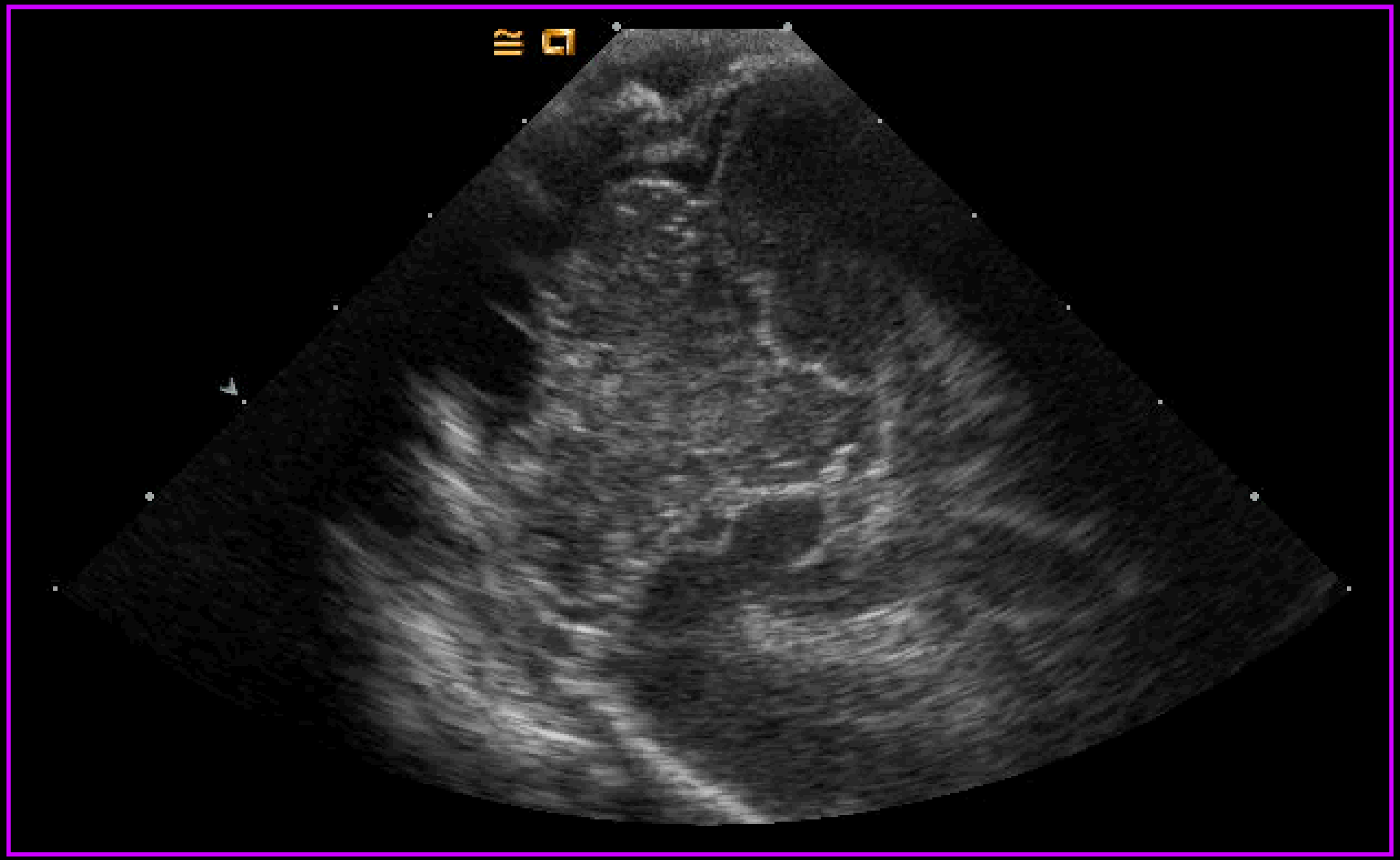


Variable appearances
28 weeks - neonatal

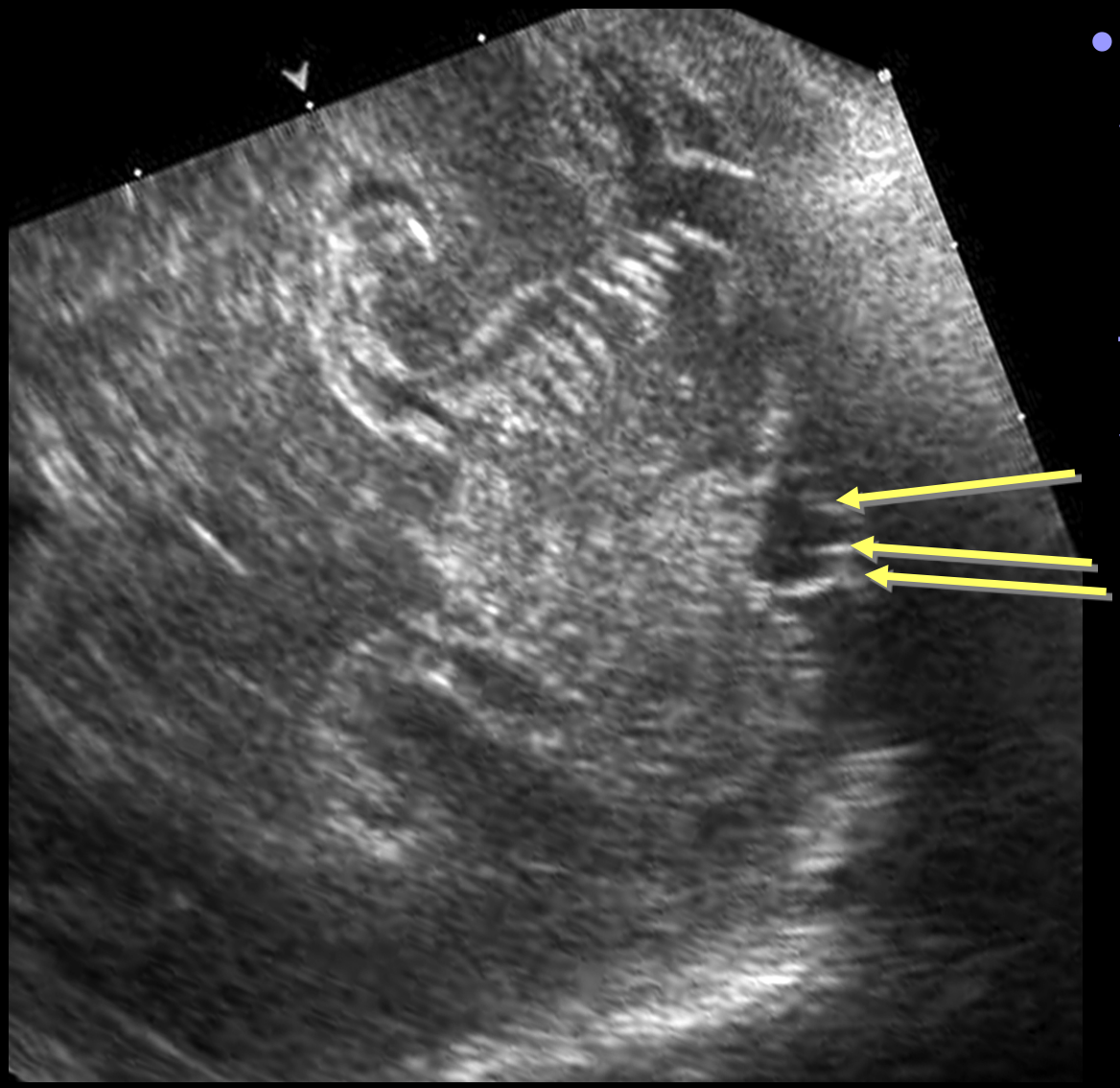


- The falx cerebelli is visible as a midline septum superiorly in the posterior fossa

Variable appearances

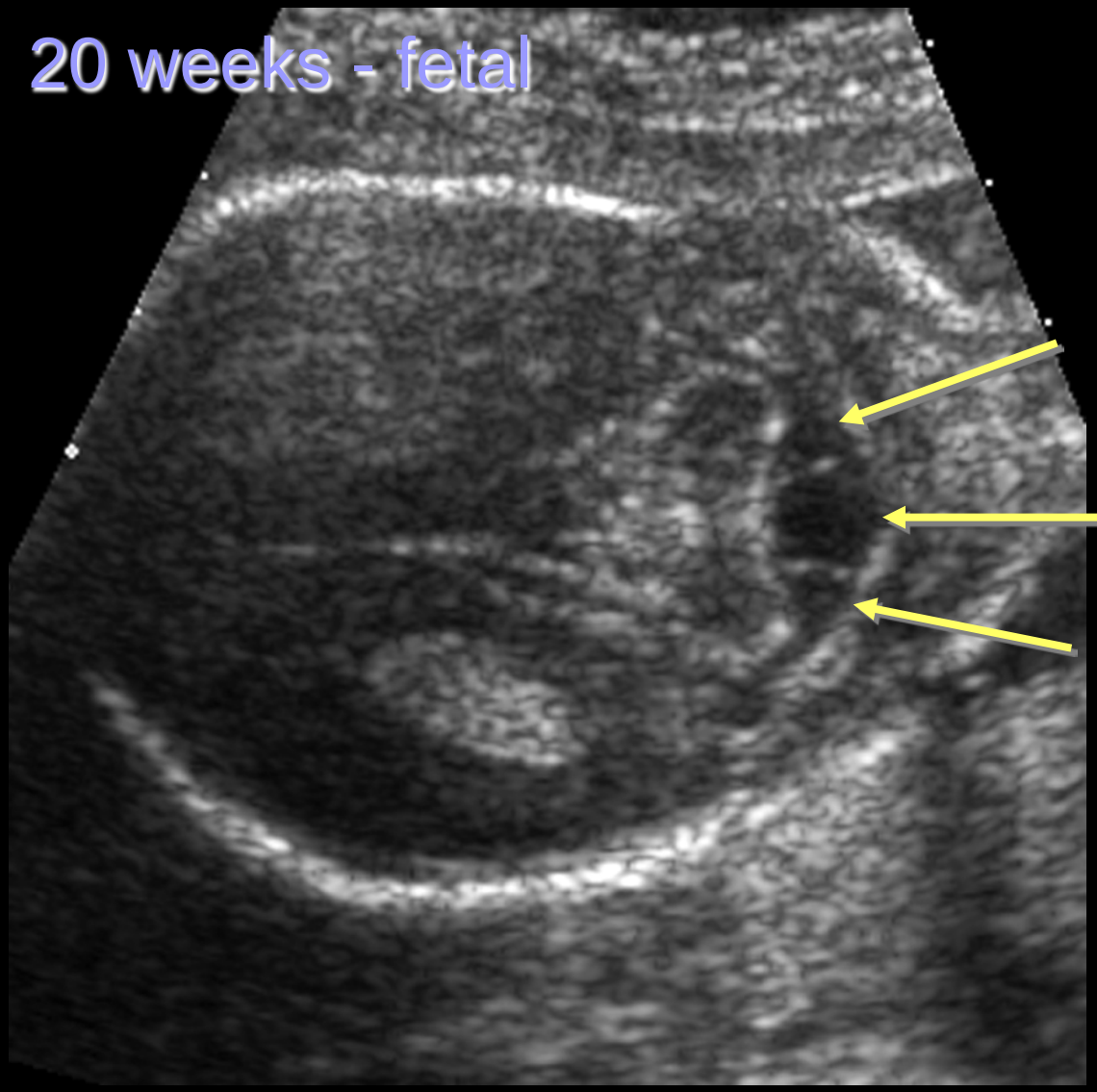


Variable appearances
27 weeks - neonatal



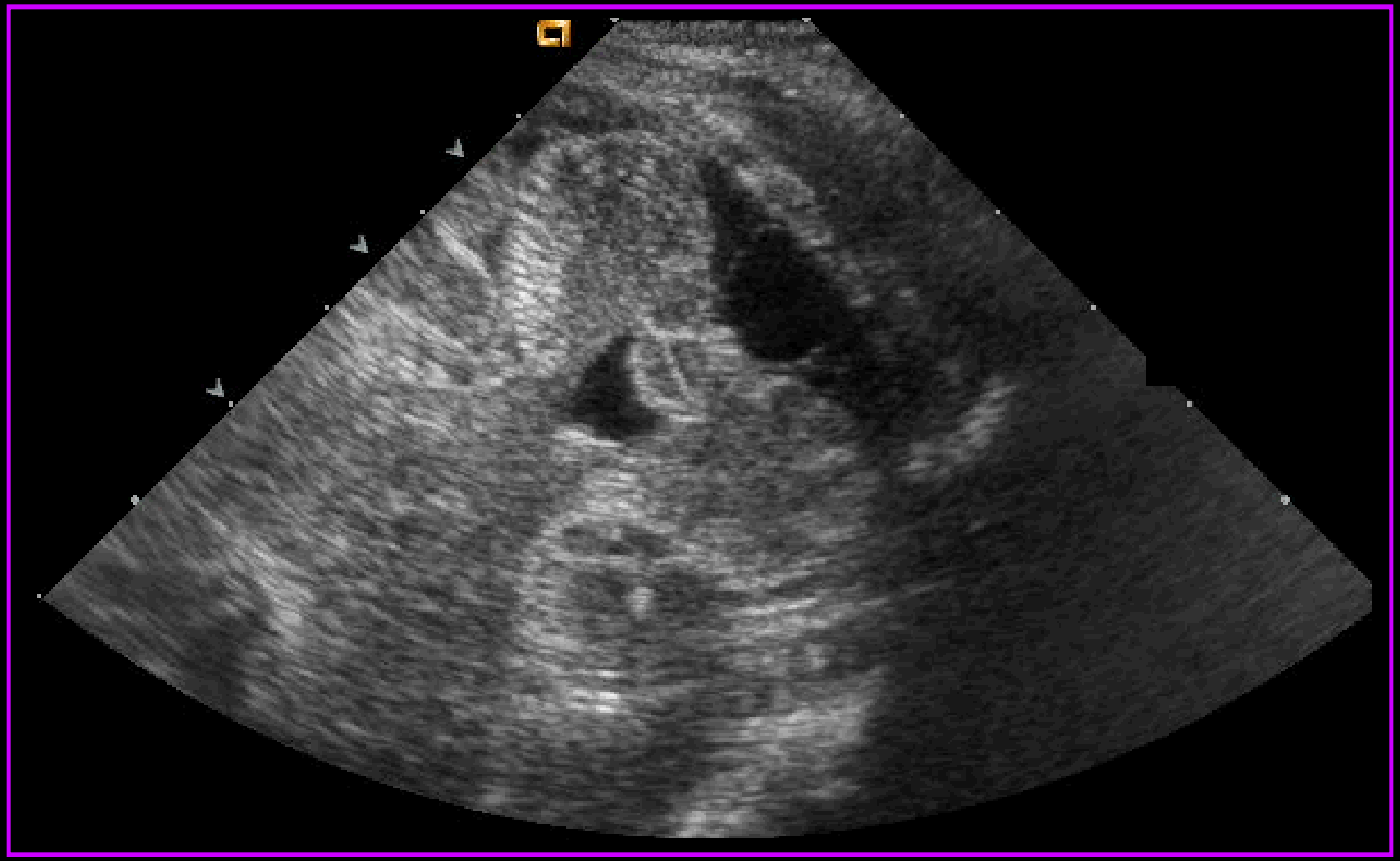
- Sometimes the septa
  and falx cerebelli
  can be seen as three septa on the same scan plane

Sonoembryology



- space between the septa is always completely anechoic
- space outside of the septa is slightly echogenic
  - Especially at earlier gestations
- Reasons for this will be explained

Sonoembryology



The cisterna magna septa
Discussion

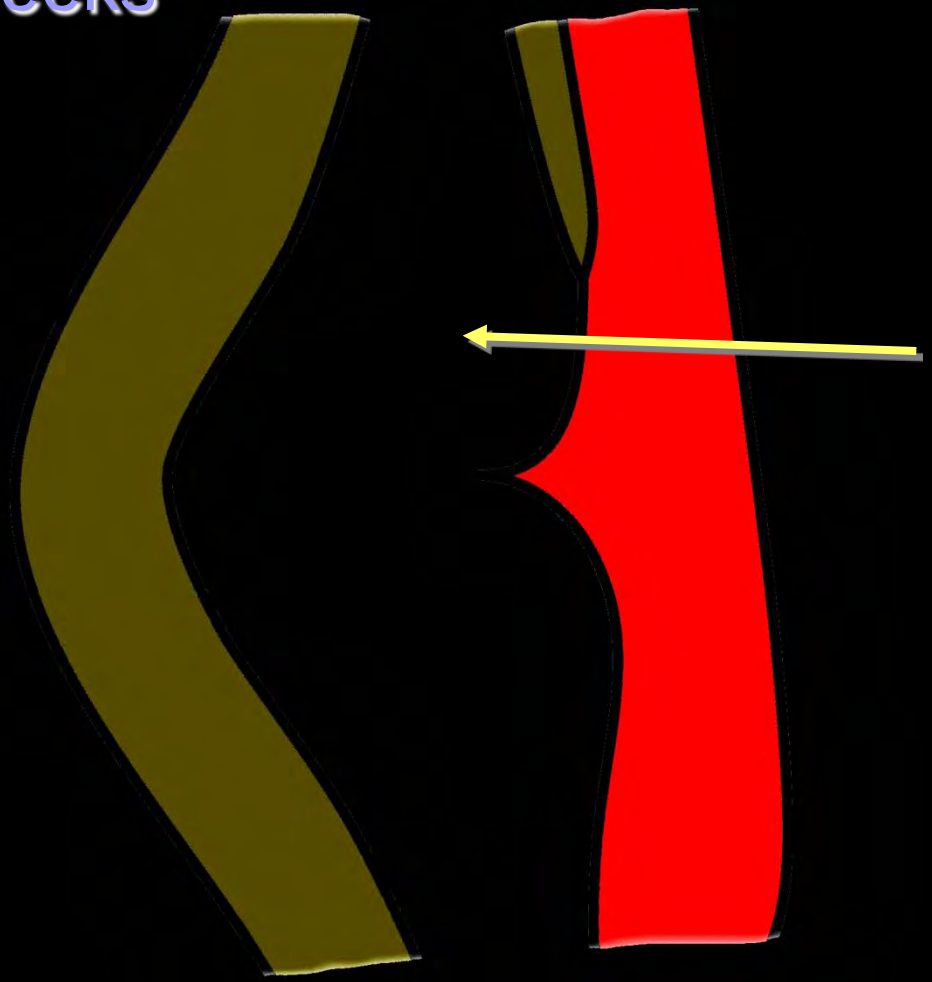
- In view of these observations it seems unlikely that the septa represent:
  - –dural folds
  - –the Torcula Herophili
  - –the straight sinus
  - –bridging arachnoid septations

Sonoembryology

- The cisterna magna septa most likely represent the walls of Blake's pouch
  - Blake JA. The roof and lateral recesses of the fourth ventricle considered morphologically and embyrologically. J Comp Neurol 1900;10: 79-108

Normal embryology rhombencephalon

7 weeks

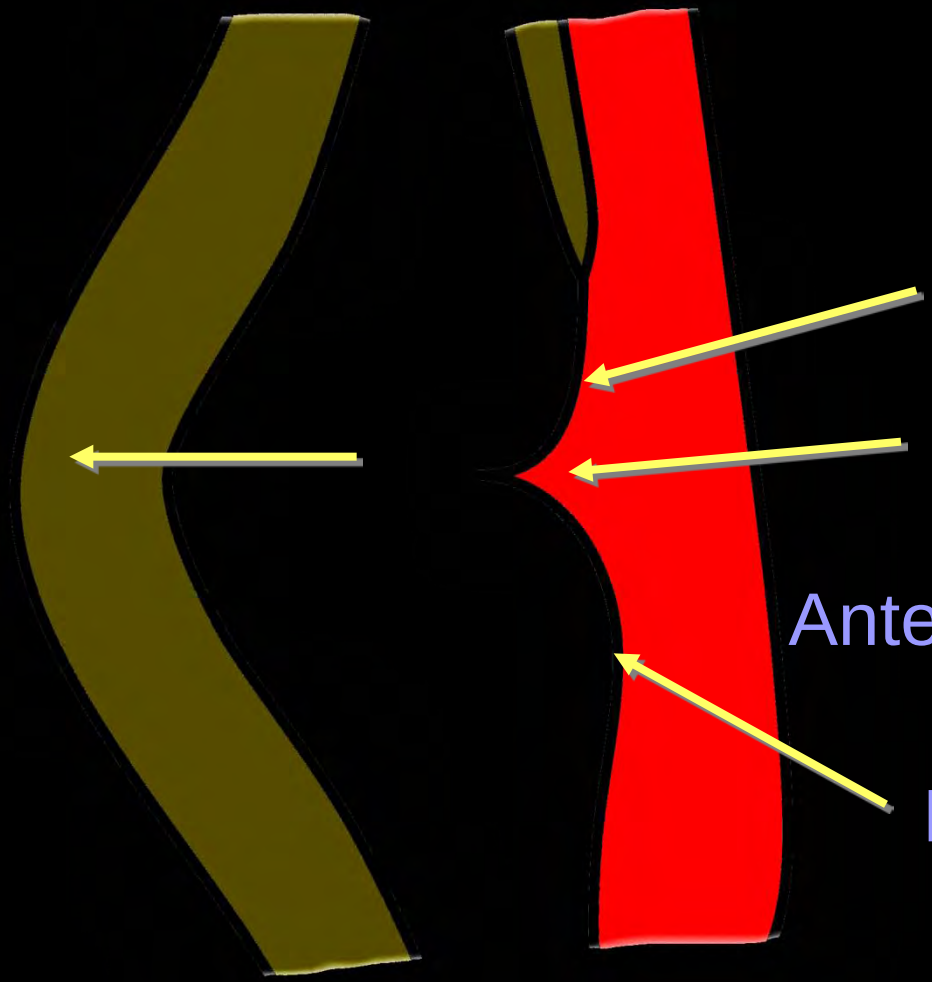


sagittal

focal dilatation of the central canal of the neural tube

rhombencephalic vesicle

Normal embryology rhombencephalon



sagittal

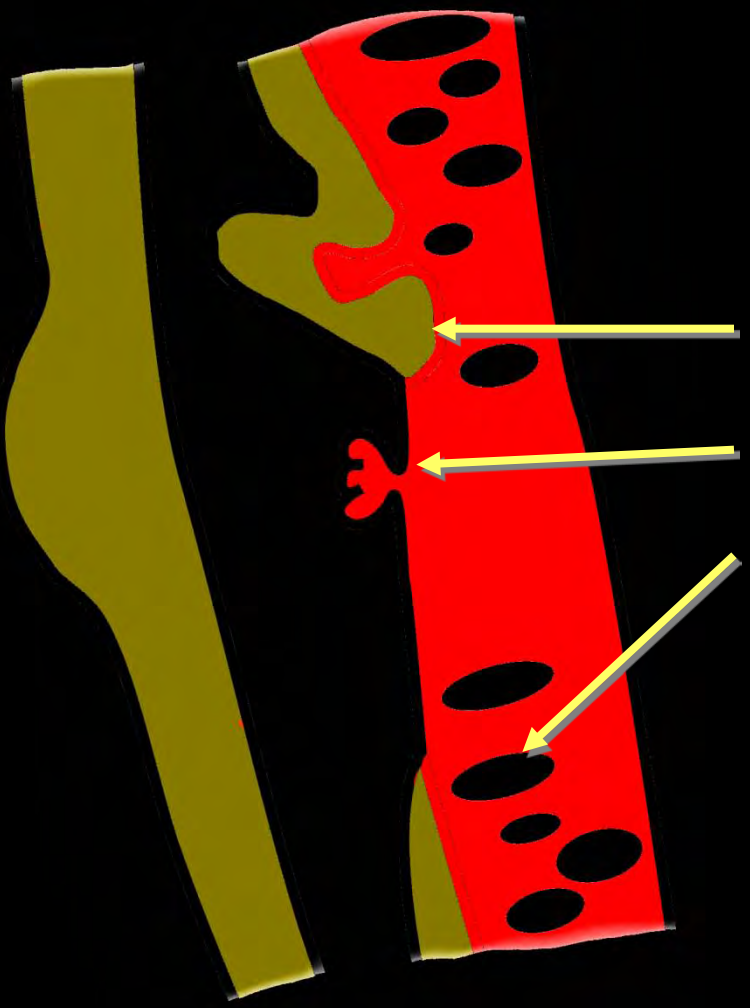
Dorsal pontine flexure

Transverse crease

Anterior membranous area (AMA)

Posterior membranous area (PMA)

Normal embryology rhombencephalon



sagittal

AMA
develops into vermis

Choroid plexus
forms in crease

Cavitation starts in
overlying meninx
primitiva

Normal embryology rhombencephalon

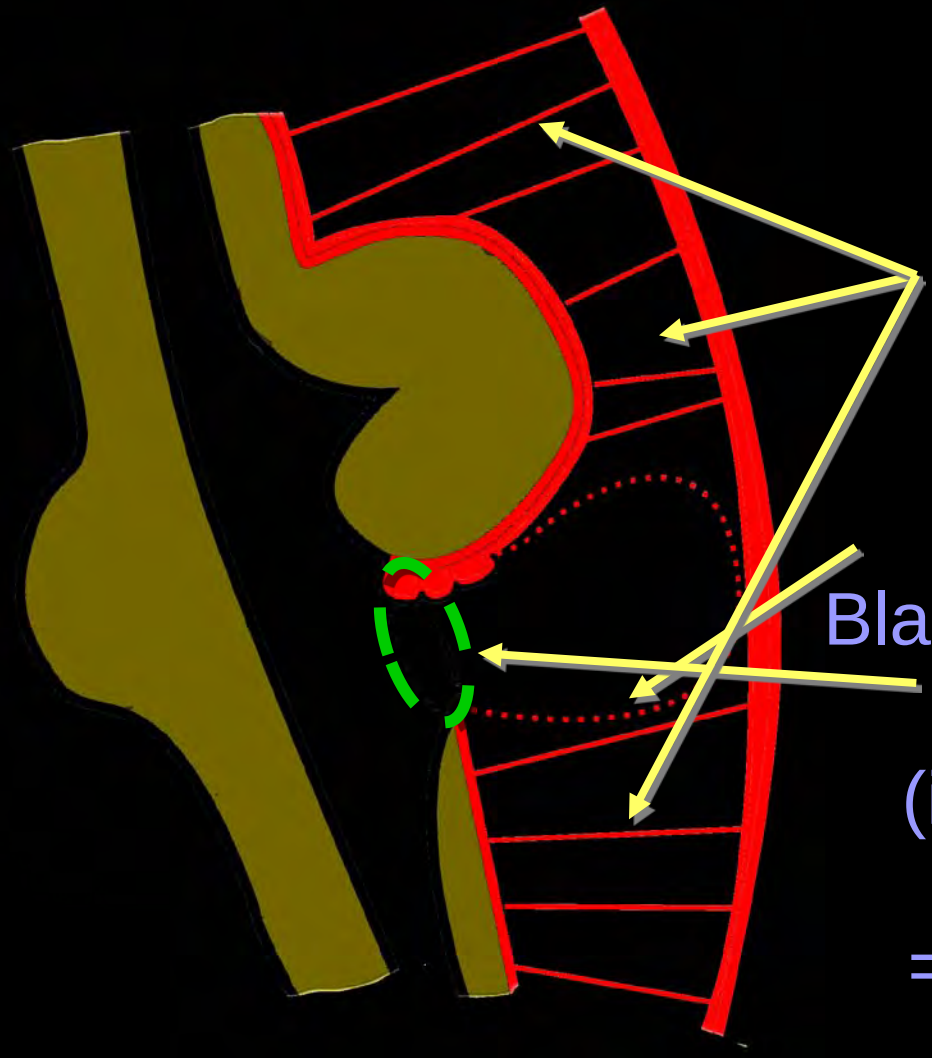
sagittal

PMA evaginates = ependyma-lined diverticulum into the meninx primitiva

Blake's pouch

Further cavitation in meninx primitiva

Normal embryology rhombencephalon

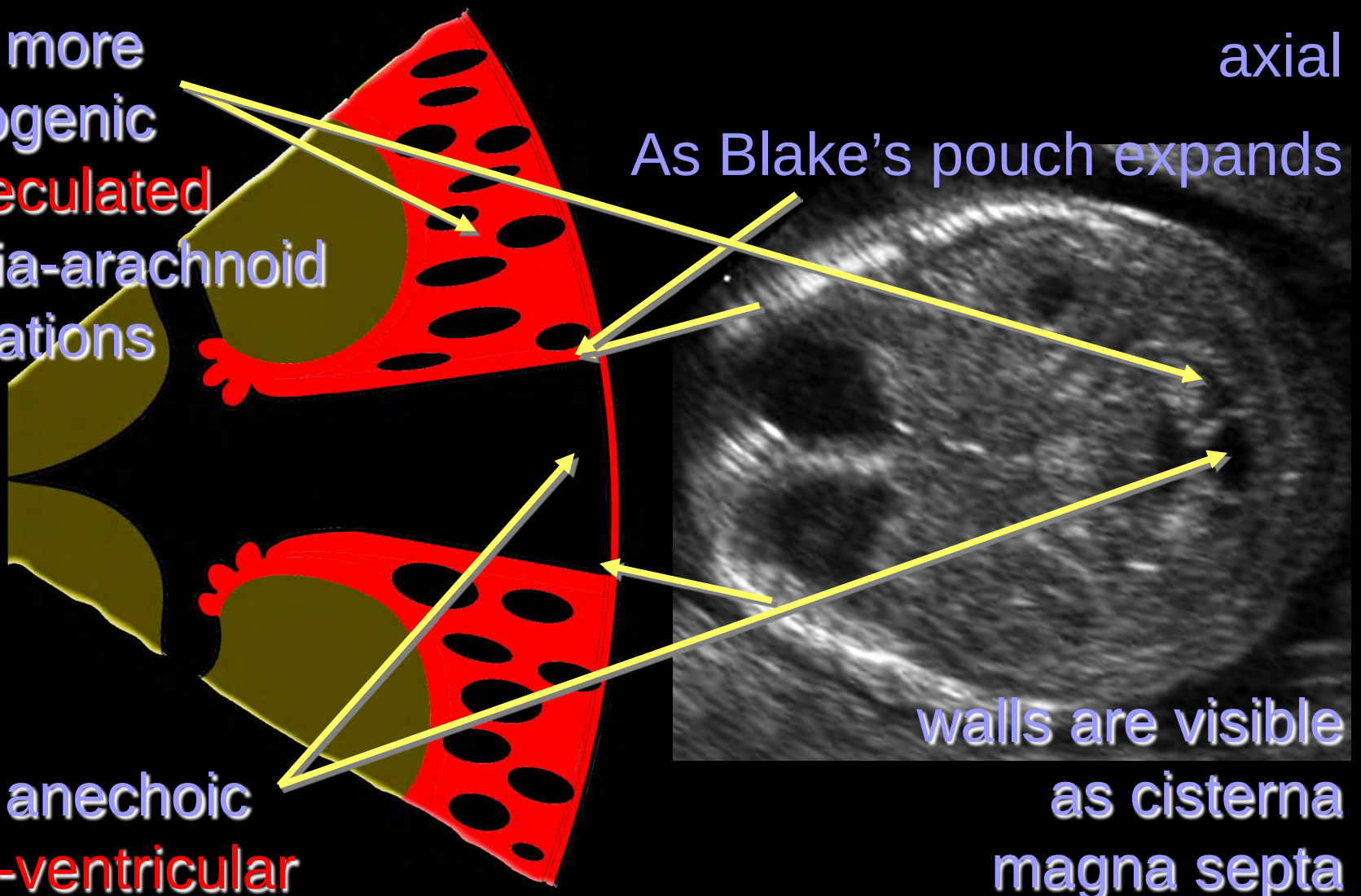


sagittal

Multiple pia-arachnoid trabeculations in subarachnoid space

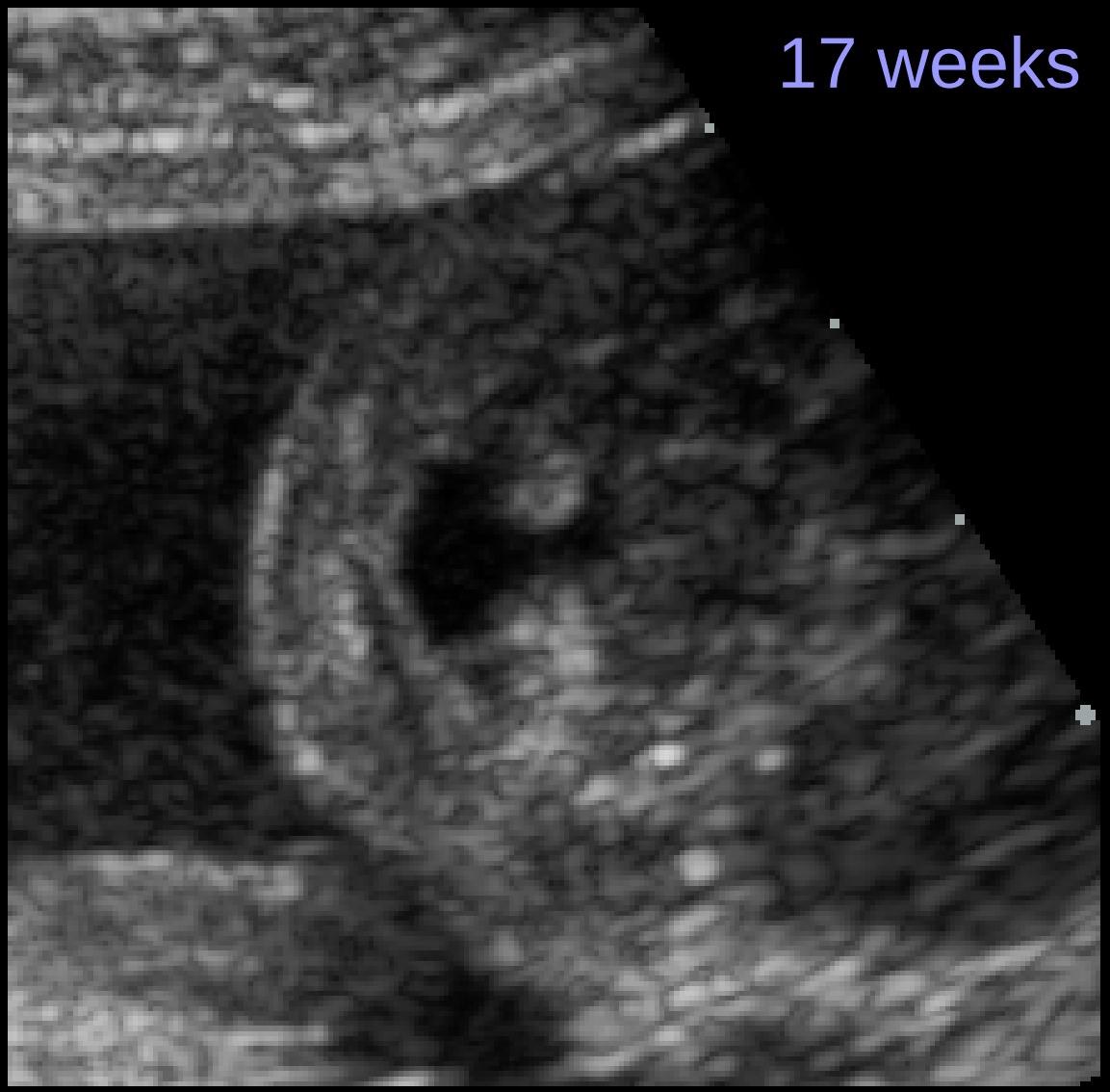
Blake's pouch fenestrates variably down to obex (inferior recess of 4th V)
neck of Blake's pouch = foramen of Magendie

Normal embryology rhombencephalon



Normal embryology rhombencephalon

Vermis incomplete
Blake's pouch freely communicating

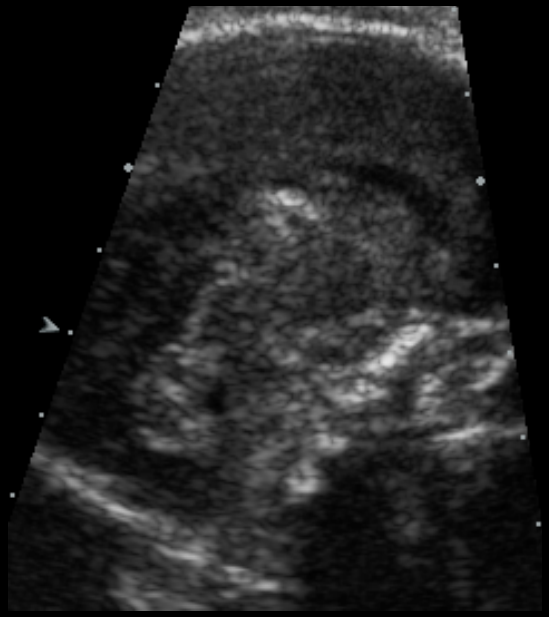


17 weeks

Normal embryology rhombencephalon

22 weeks – same fetus
Vermis complete
Communication not easily visible

Sagittal

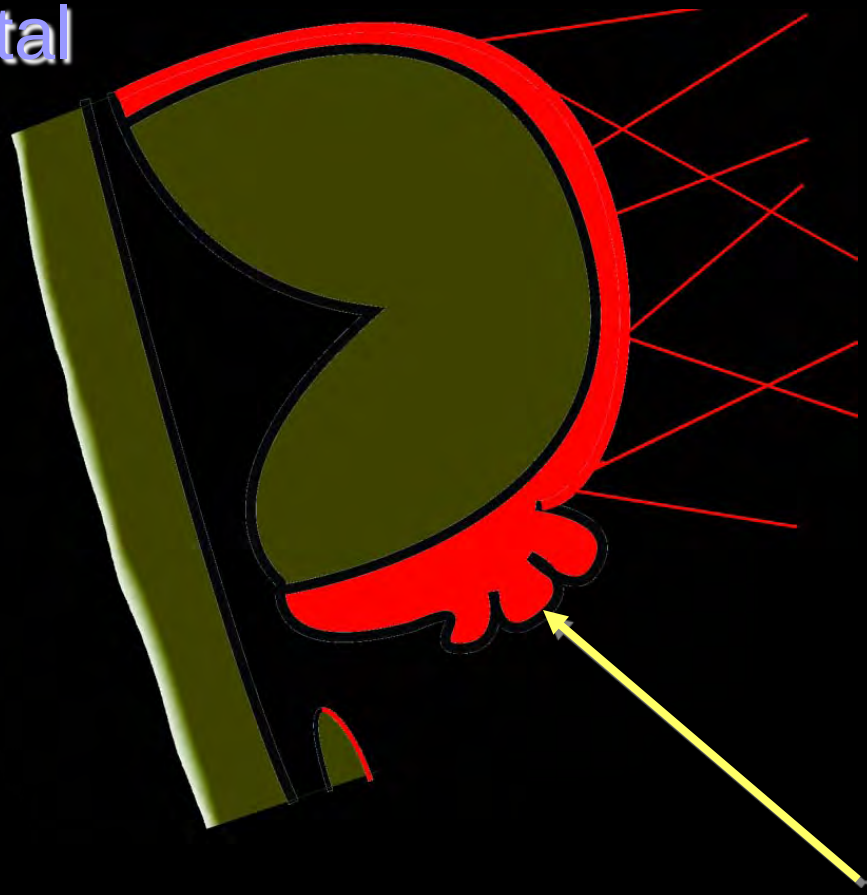


Coronal

Axial

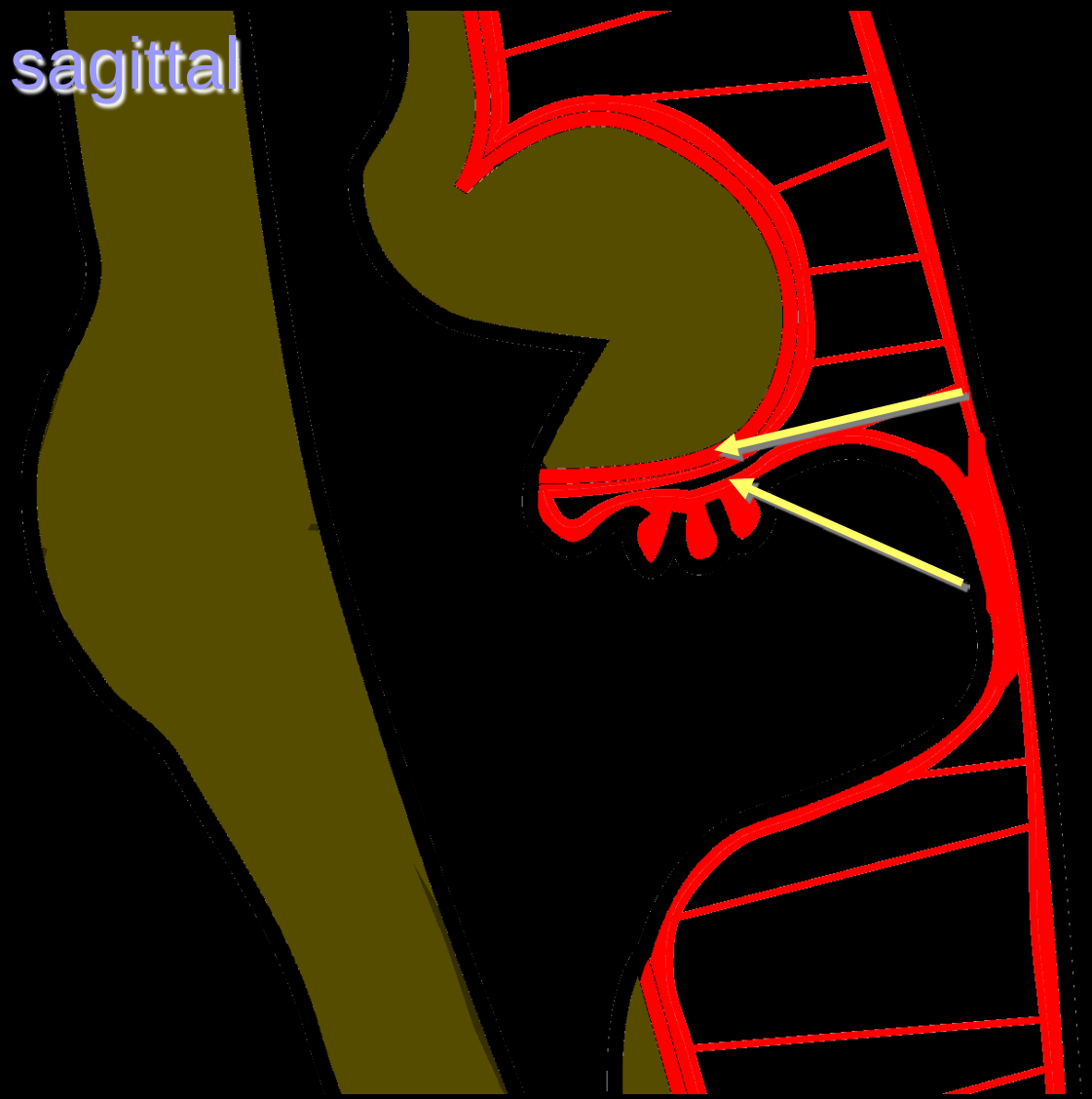
Question

sagittal



Why does choroid often extend outwards beneath the vermis into the cisterna magna?

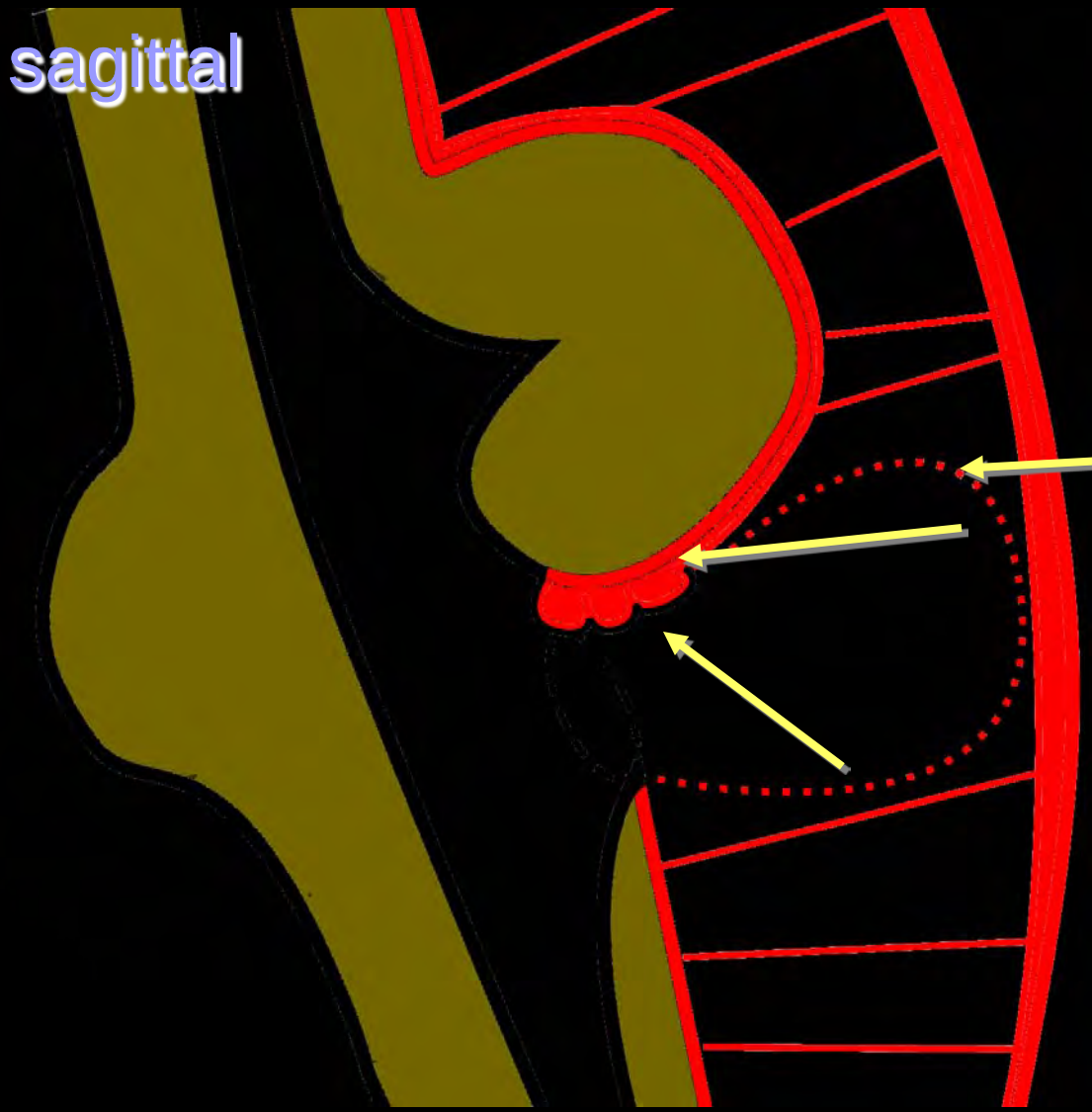
Answer



Because it is carried in the roof and walls of Blake's pouch as it expands dorsally

as it expands, two layers of pia mater appose and fuse

Answer



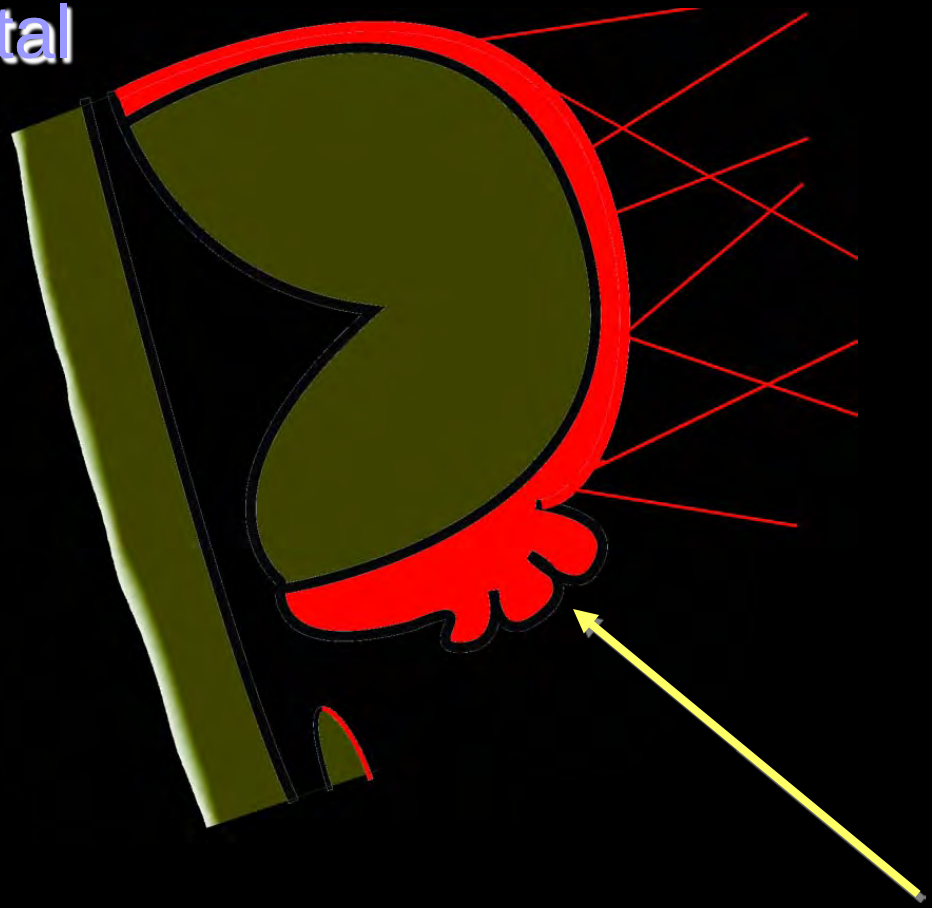
Choroid becomes continuous with pia mater over cerebellum / brainstem

Blake's pouch fenestrates

Choroid appears to be in cisterna magna

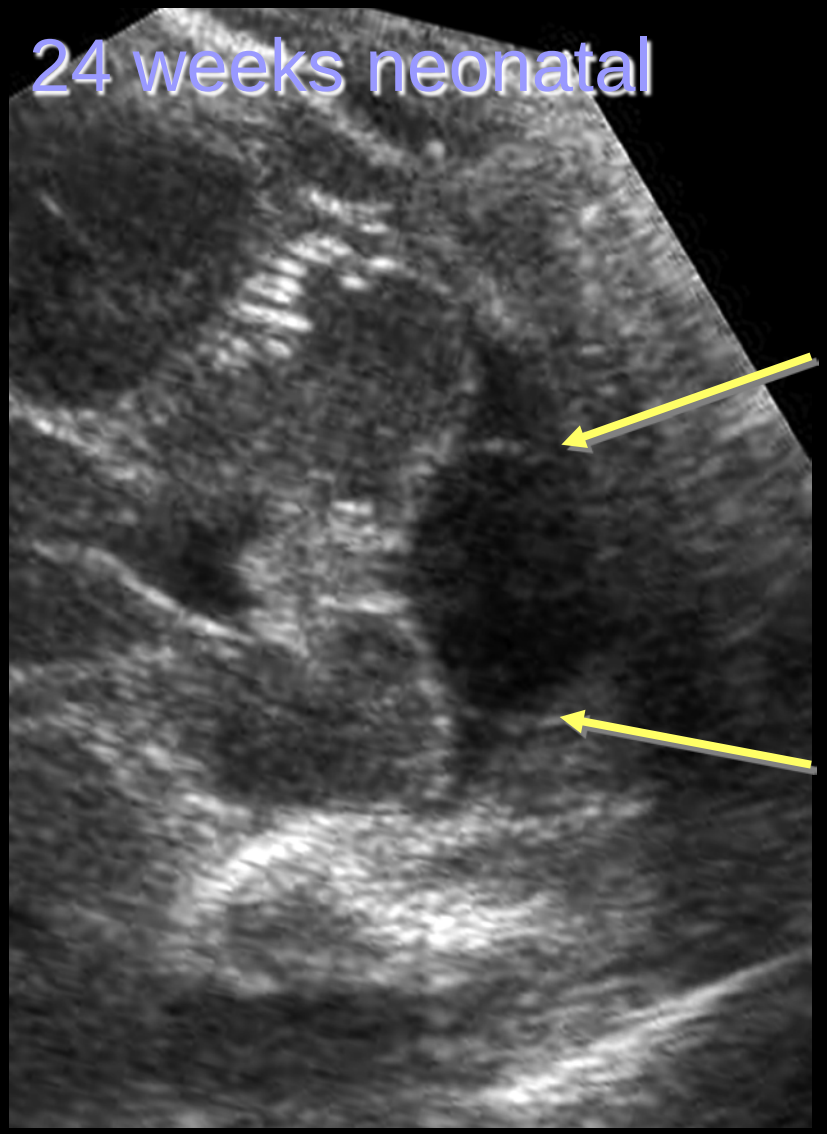
Answer

sagittal



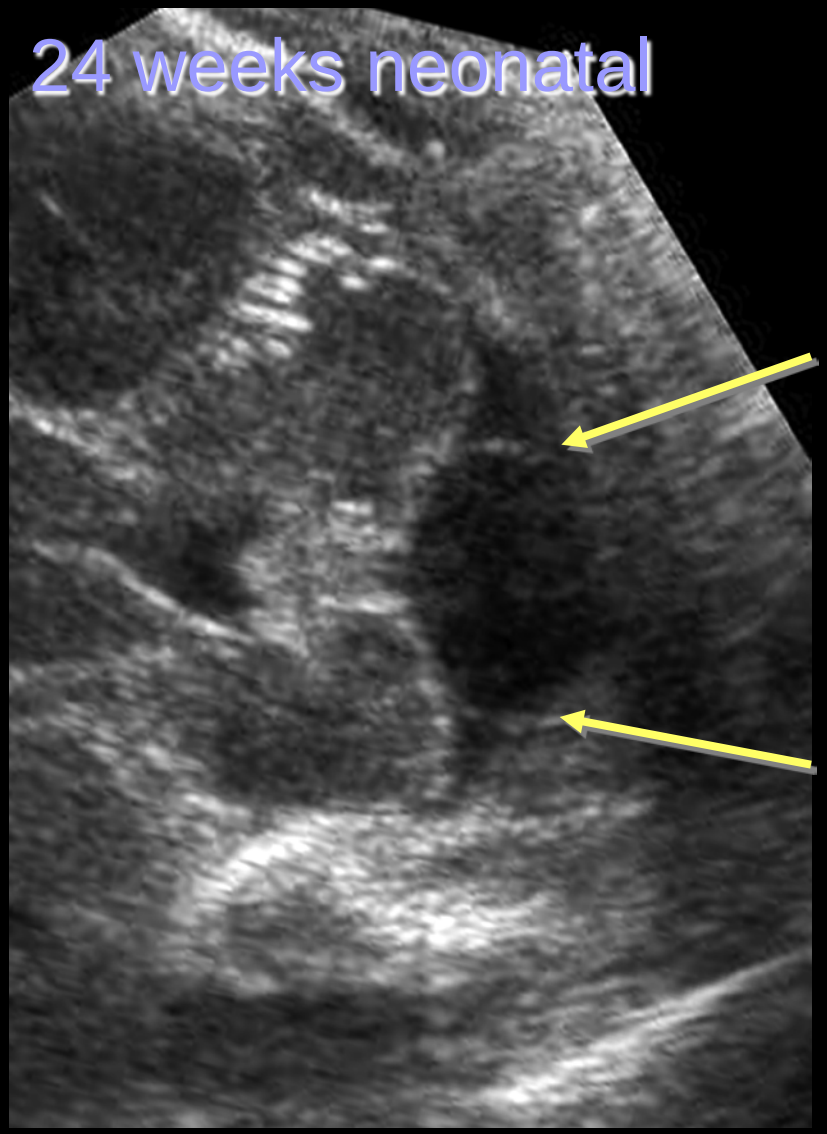
Choroid appears to be in cisterna magna

Question



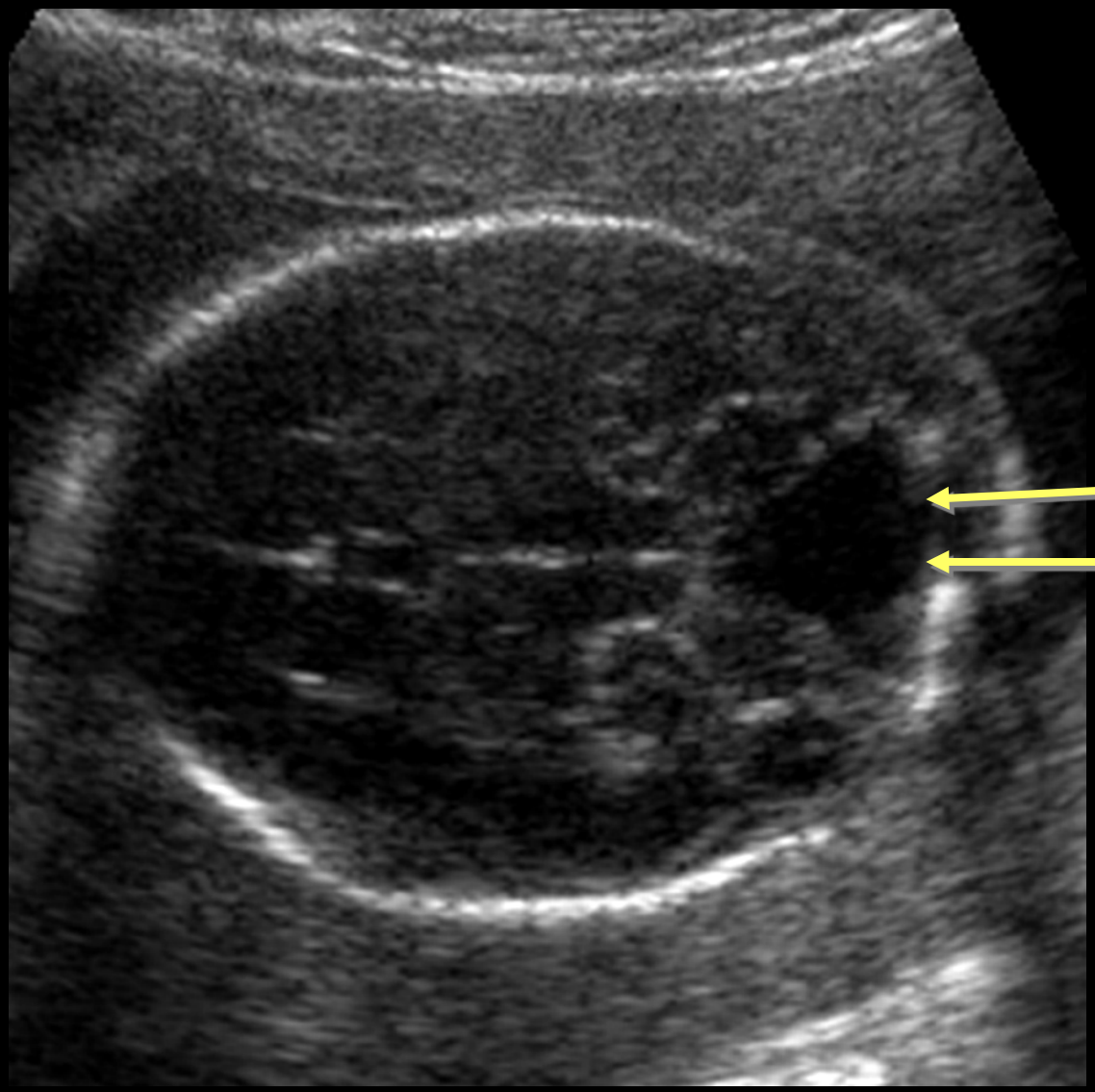
- Why are the septa sometimes deviated giving impression of a cyst?
  - Knutzon RK, McGahan JP, Salamat MS, et al. Fetal cisterna magna septa: a normal anatomic finding. Radiology. 1991 Sep;180(3): 799-801
- Why do we see transient enlargement of the ventricles at 14-16 weeks, resolving by 22-23 weeks gestation?
  - Bronshtein M, Zimmer EZ, Blazer S. Isolated large fourth possible ventricle in early pregnancy--a benign transient phenomenon. Prenat Diagn. 1998 Oct;18(10): 997-1000

Answer



- fenestration of Blakes' pouch variable in timing
- outward bowing of the cisterna magna septa may be due to a delay or non-fenestration of Blake's pouch
- In a small percentage of individuals it is the normal later opening of the foramena of Luschka which leads to equilibrium of CSF between ventricles and subarachnoid cisterns

Question



Why are the septa not seen in fetuses with Dandy-Walker malformation?

–Pretorius DH, Kallman CE, Grafe MR et al. Linear echoes in the fetal cisterna magna. J Ultrasound Med. 1992 Apr;11(4): 125-8

Because they are displaced laterally

Answer

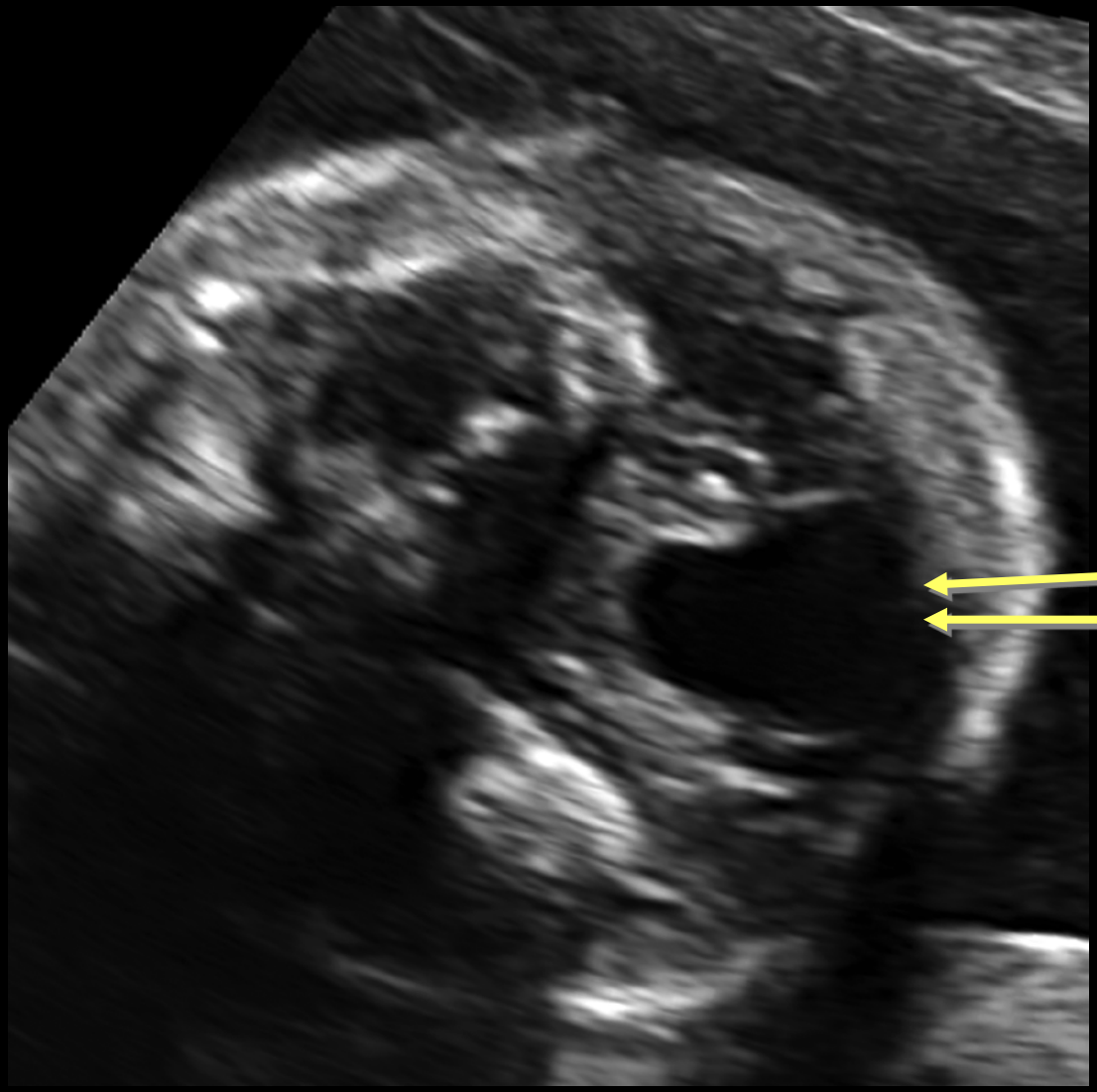
cisterna magna septa / walls Blake's pouch

=

walls of Dandy-Walker "cyst"

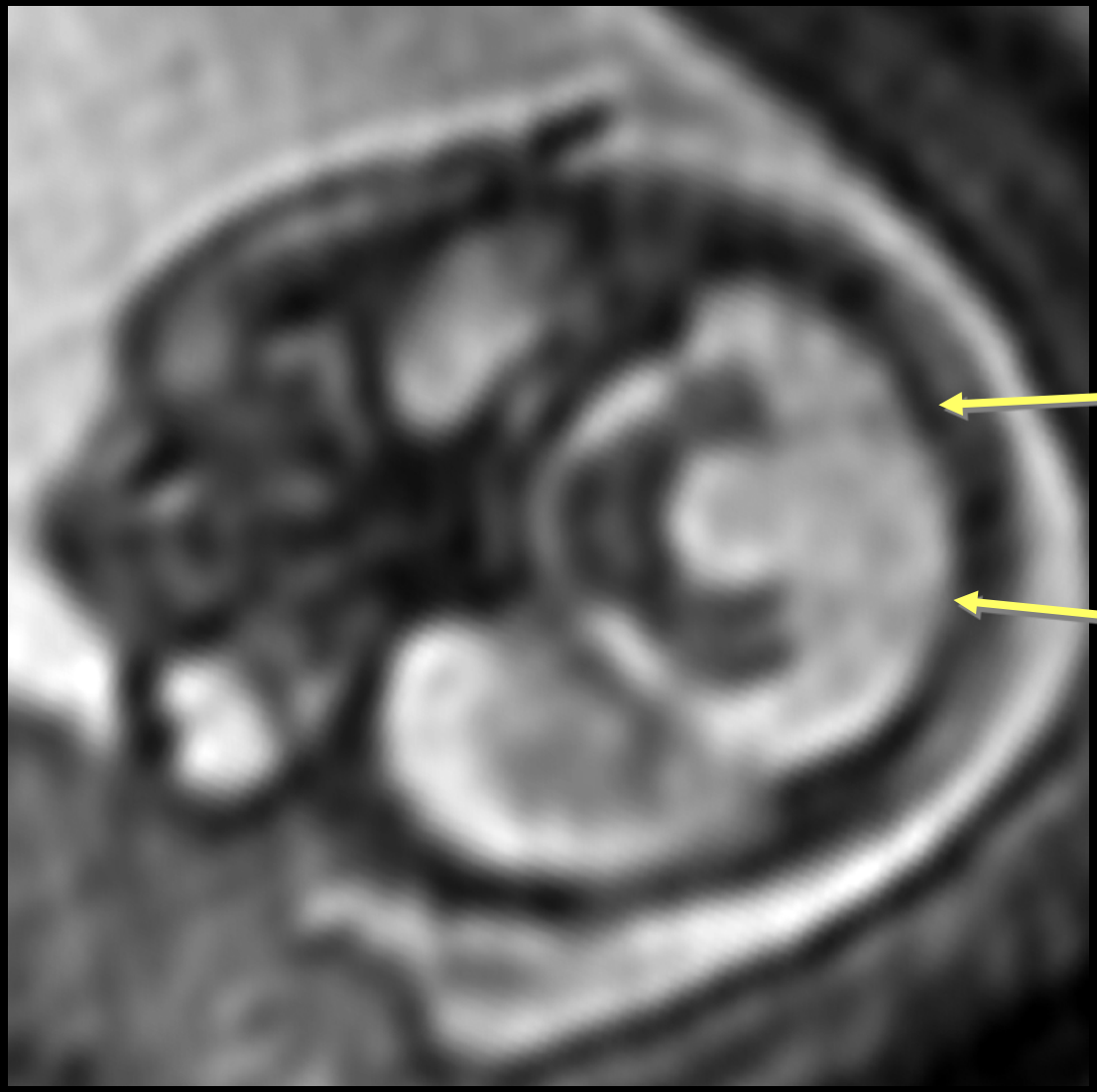
- the same anatomical structure
- during expansion of Blake's pouch it's walls become apposed to and indistinguishable from the sides of the posterior fossa

Dandy-Walker continuum



Septa displaced laterally

Dandy-Walker continuum



Septa occasionally visible by MRI

Dandy-Walker continuum

vermis elevated & hypoplastic

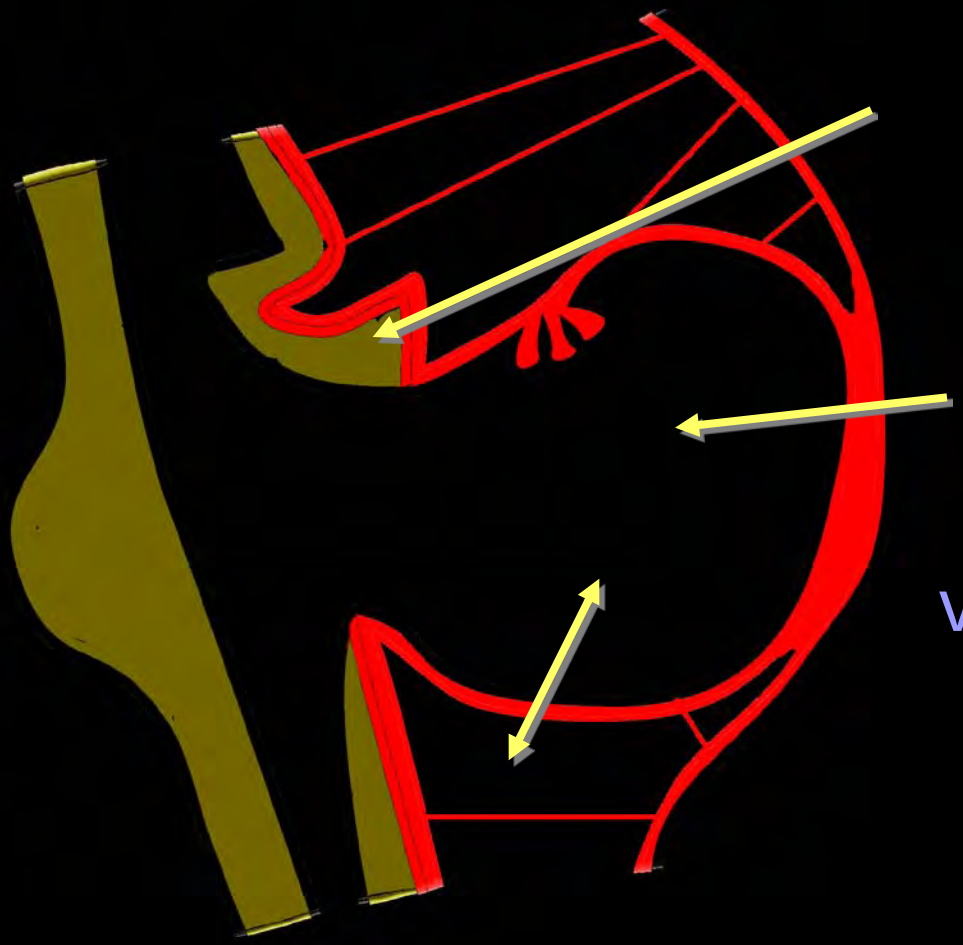
Wide communication between 4th ventricle & pouch

Dandy-Walker continuum

vermis elevated & lobulation abnormal

Blake’s pouch dilated

Dandy-Walker continuum

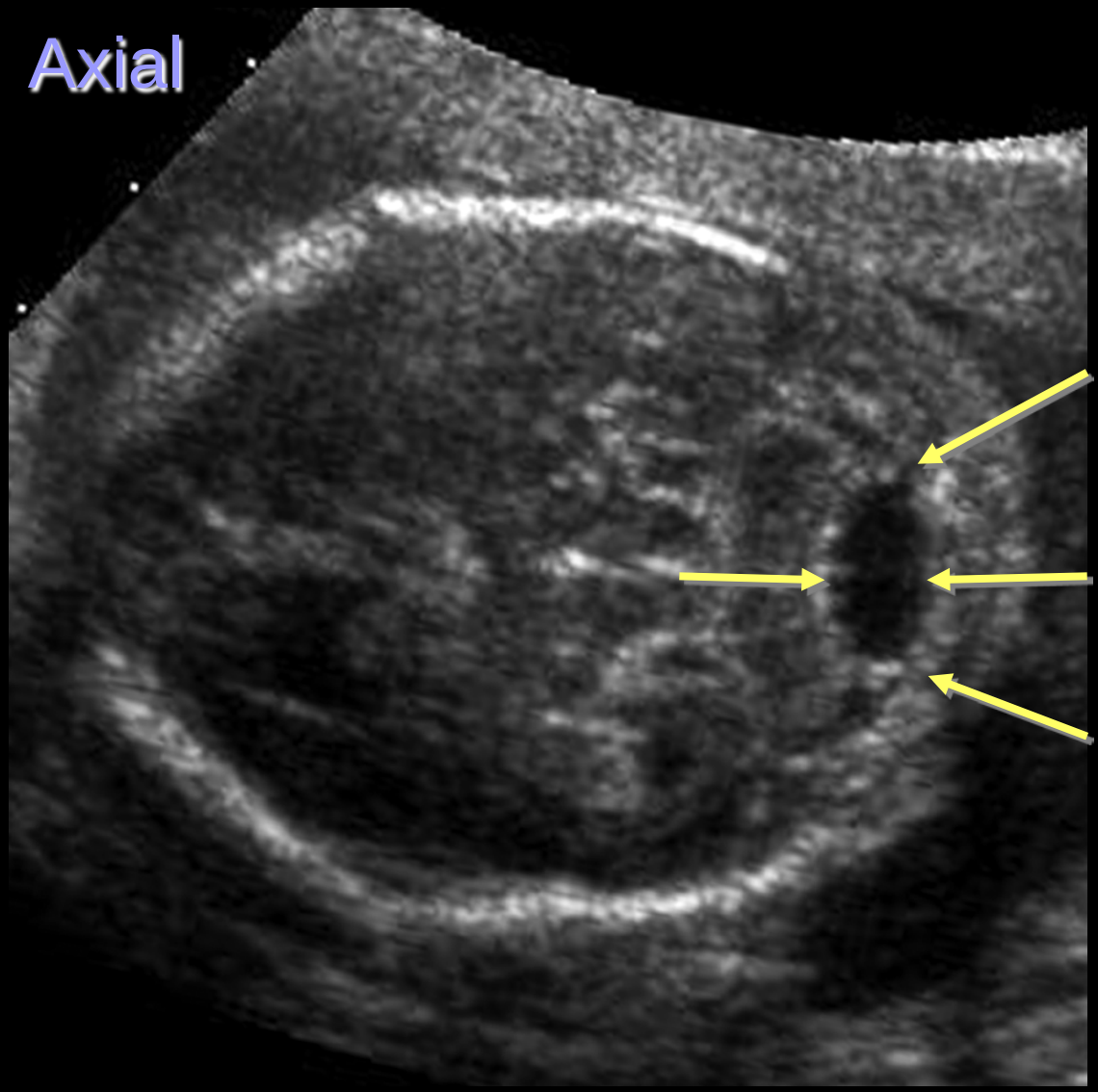


vermis elevated & hypoplastic

Blake’s pouch dilated

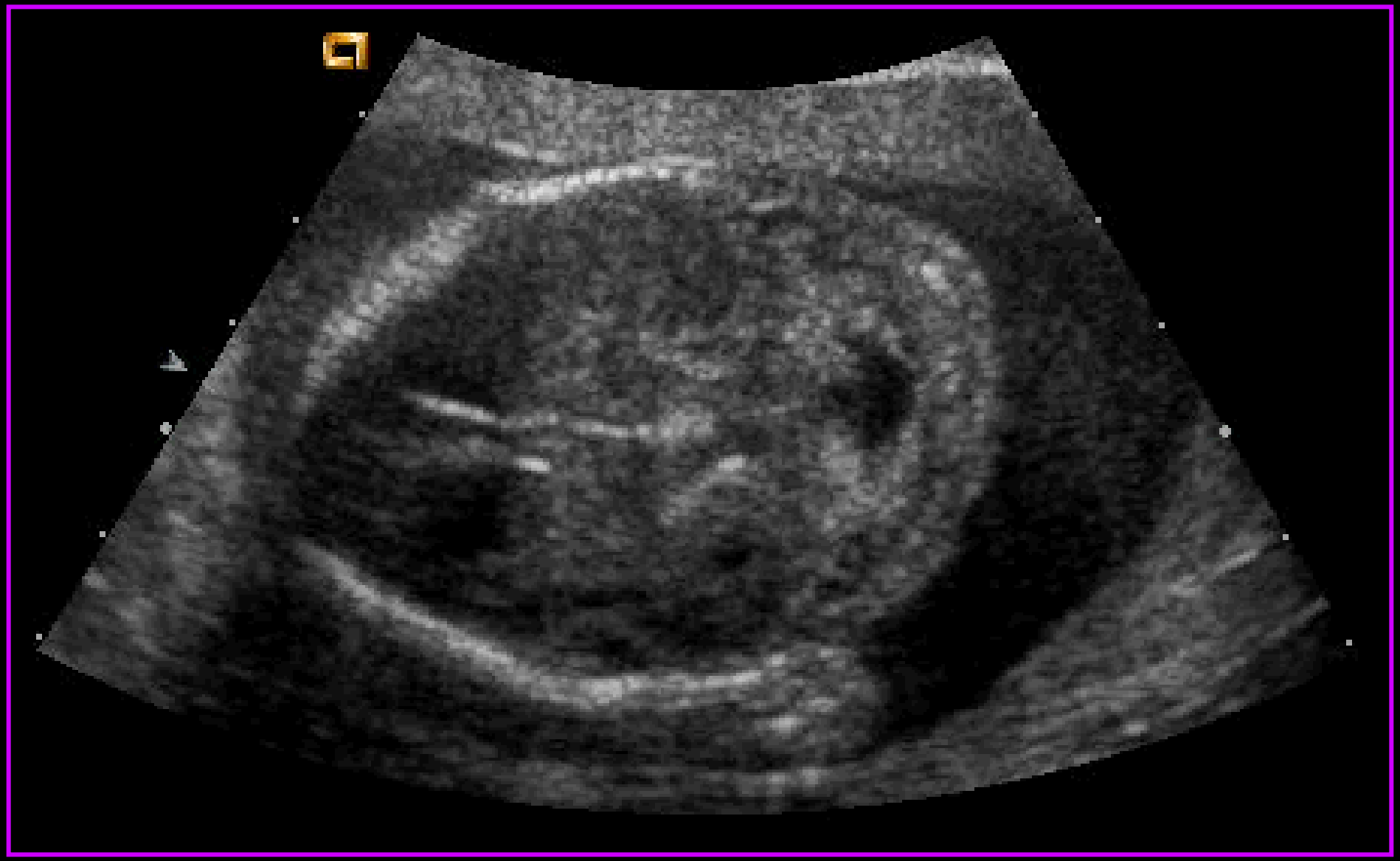
variable communication on cisternography

Question

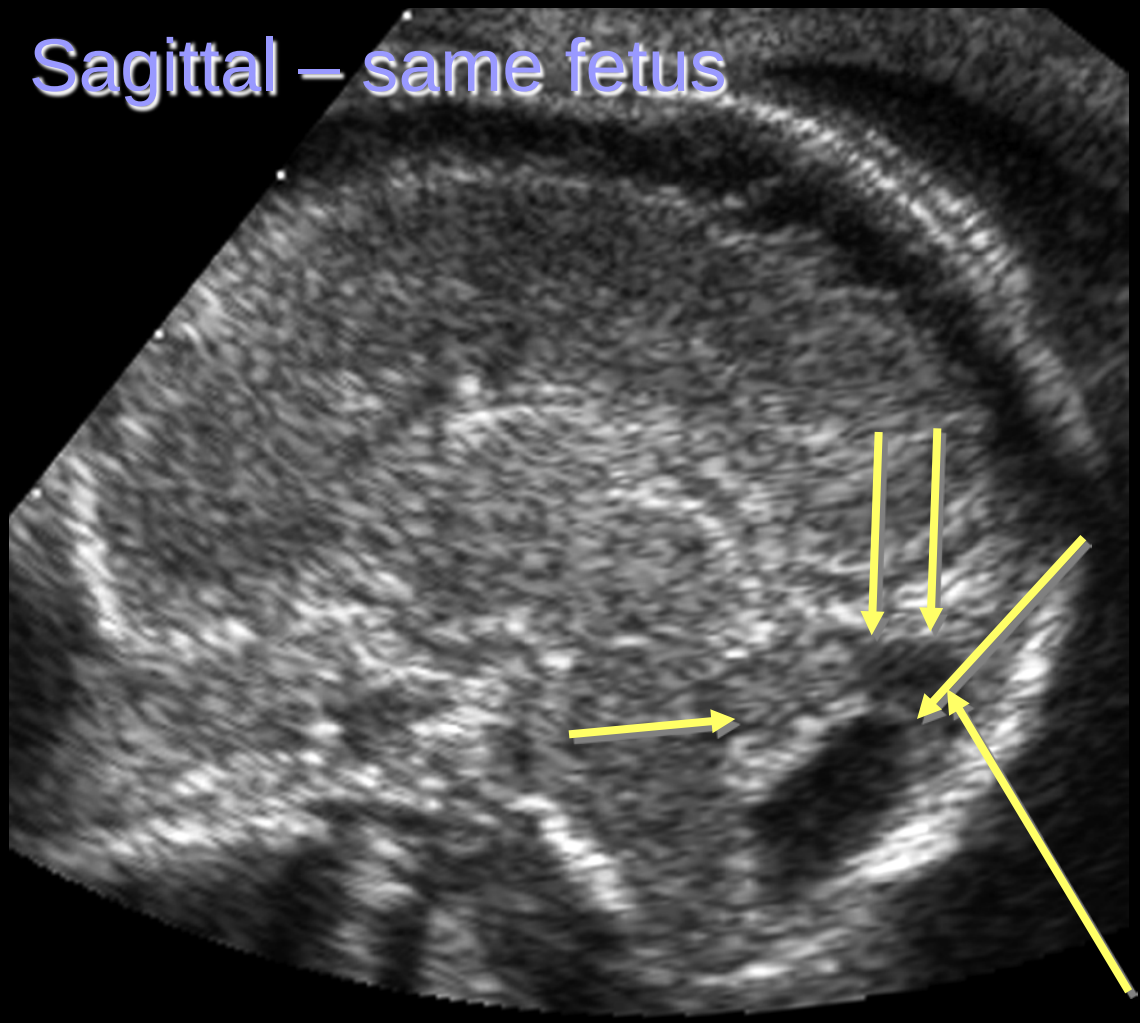


How can we explain the sonographic diagnosis of mega cisterna magna?
Blake's pouch expanded with walls deviated laterally

Question



Answer



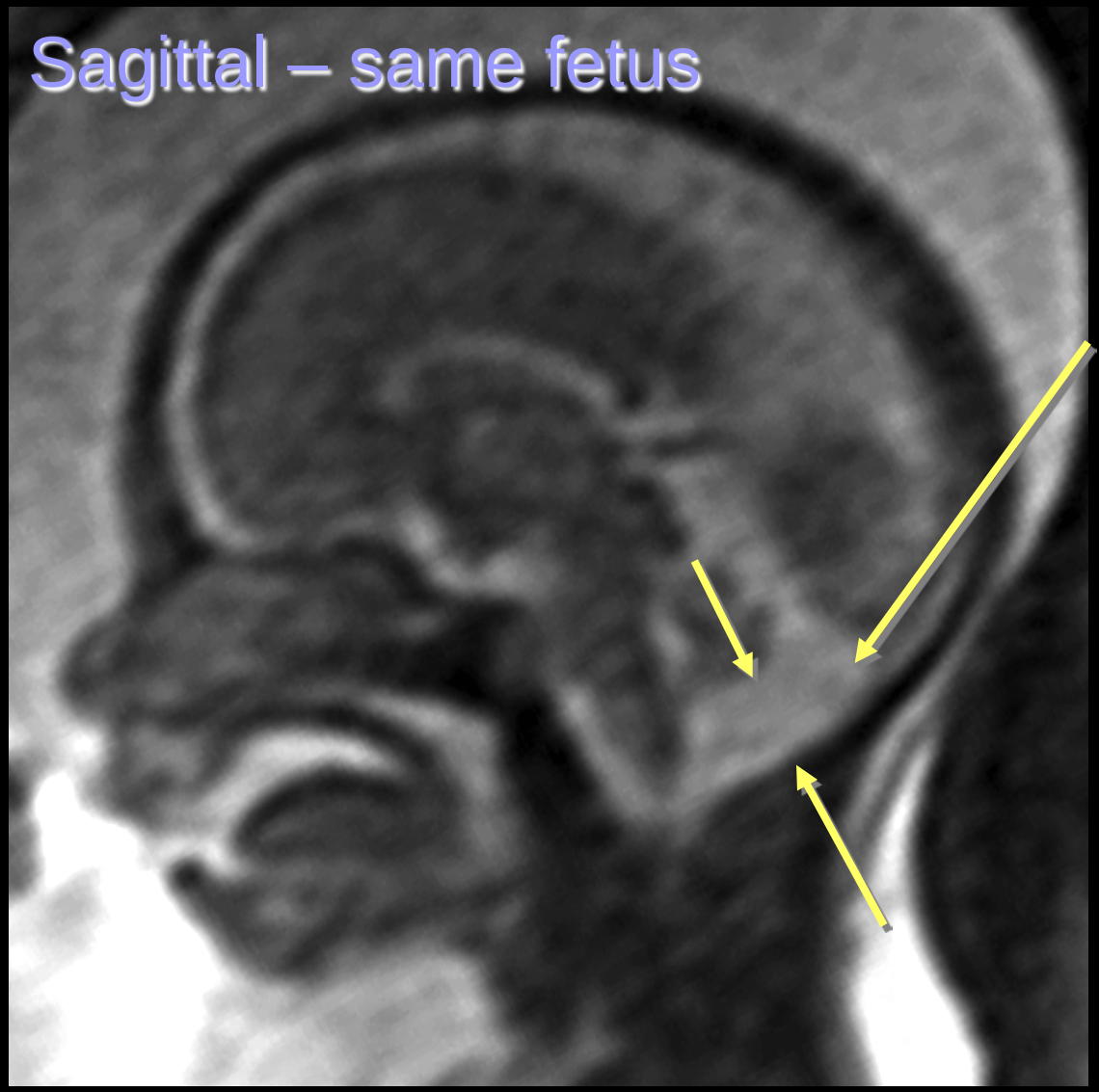
vermis

Blake's pouch in cisterna magna

falx cerebelli

echogenic subarachnoid space

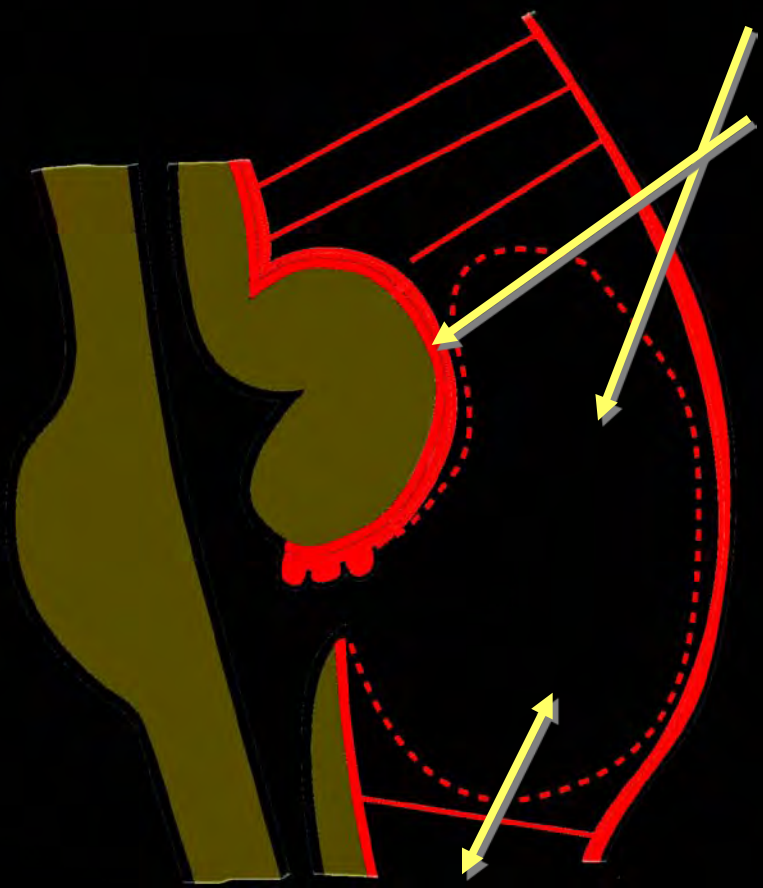
Answer



Blake's pouch not resoluble by MR

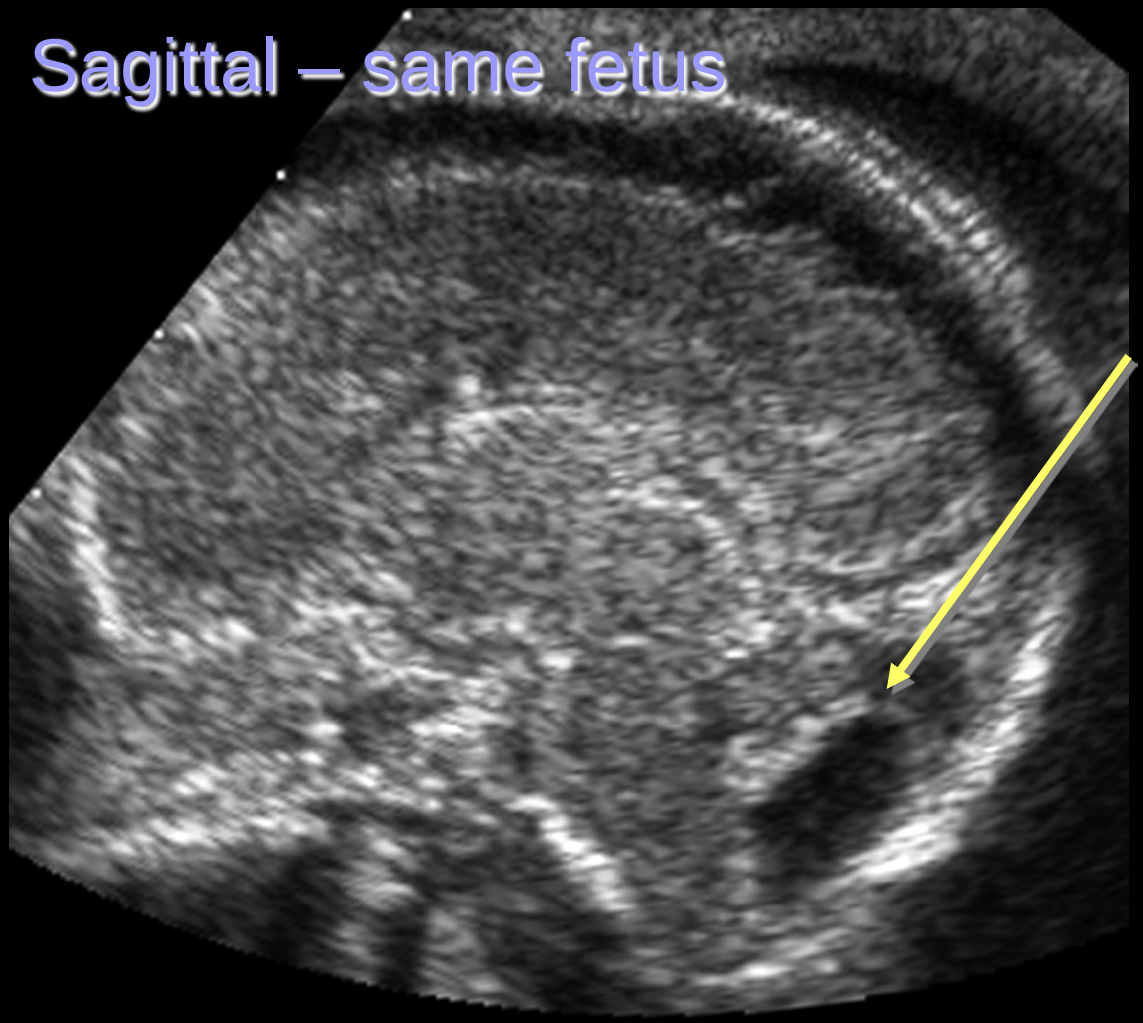
MR diagnosis: mega cisterna magna

Mega cisterna magna



- Blake’s pouch persists and expands
- posterior to a normally formed cerebellum
- it eventually fenestrates leaving behind an expanded cisterna magna
- explaining the free communication between the fourth ventricle and subarachnoid space that is seen in this condition

Mega cisterna magna

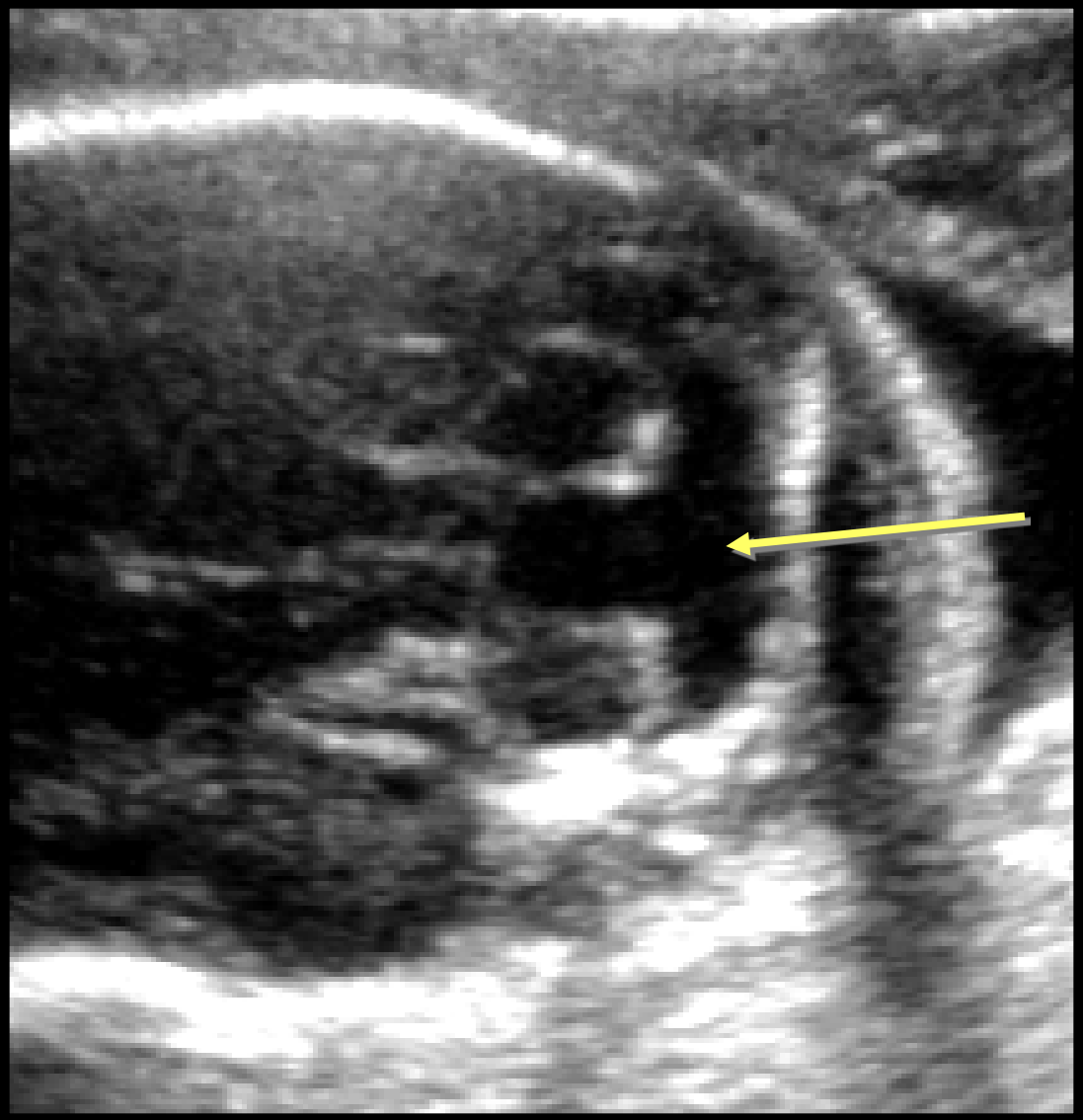


Mega cisterna magna

=

mega Blake's pouch

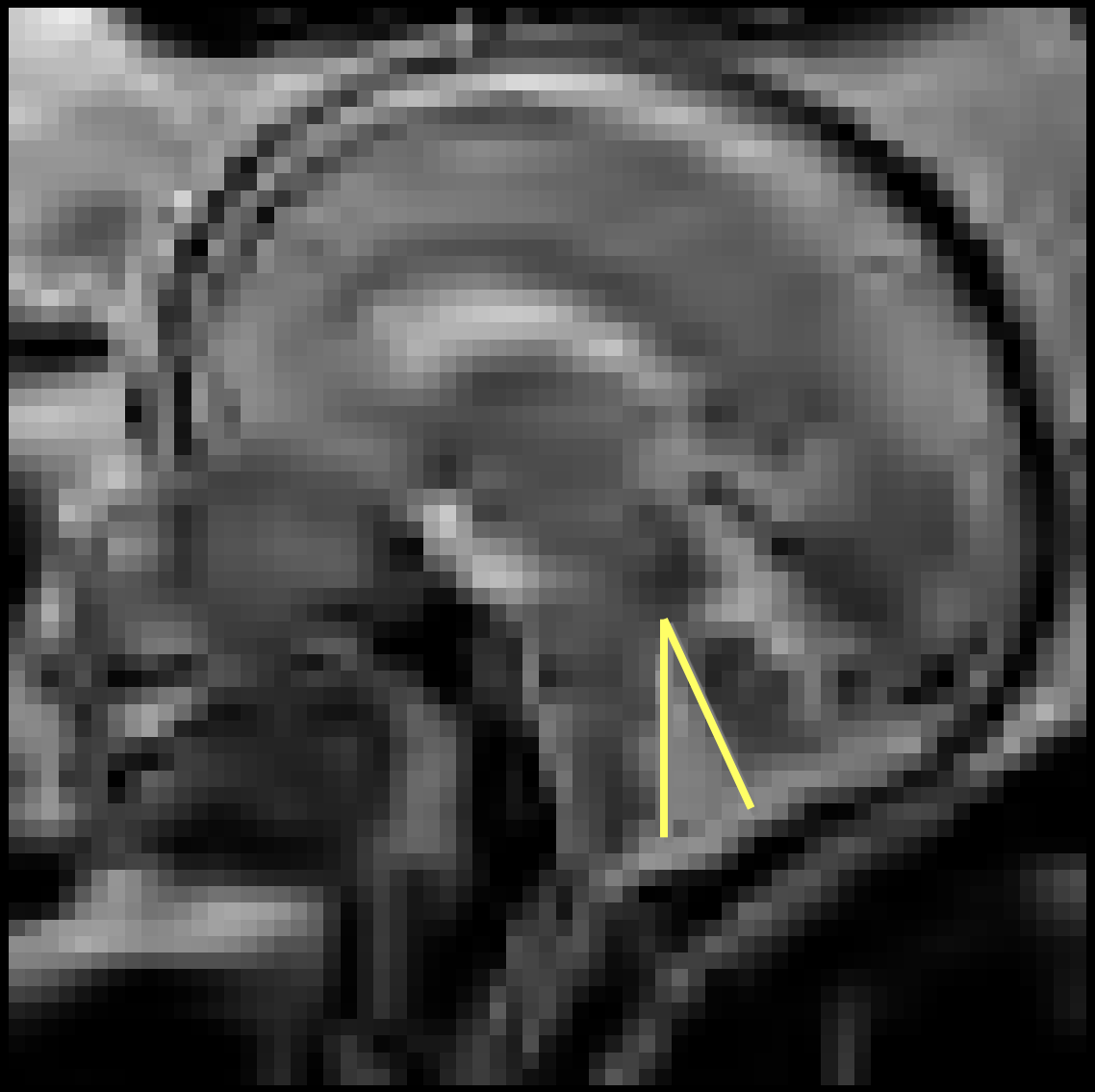
Question



- Why is there such a poor correlation of ultrasound and autopsy findings in apparent cystic malformations of the posterior fossa?

- Carroll SG, Porter H, Abdel-Fattah S, Kyle PM, Soothill PW. Correlation of prenatal ultrasound diagnosis and pathologic findins in fetal brain abnormalities. Ultrasound Obset Gynecol 2000; 16: 149-53

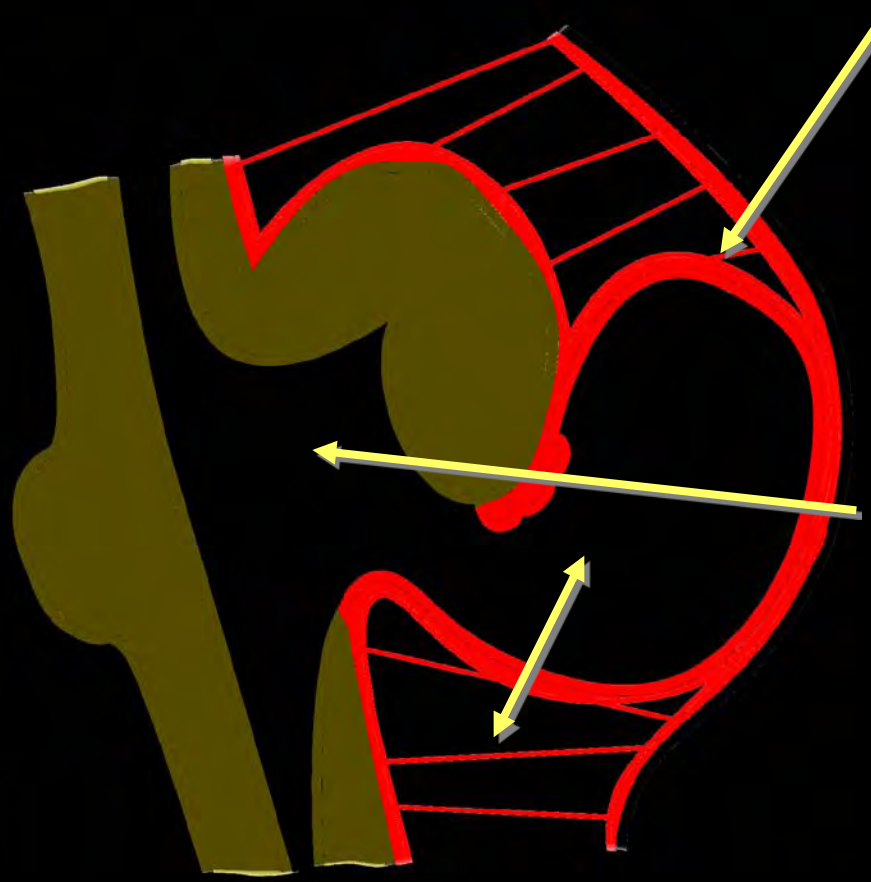
Question



- Why does angulation of a morphologically normal vermis away from the brainstem have a good prognosis?

- Zalel Y, Gilboa Y, Gabis L et al. Rotation of the vermis as a cause of enlarged cisterna magna on prenatal imaging. Ultrasound Obstet Gynecol 2006; 27:490-3

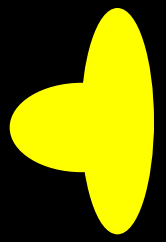
Answer



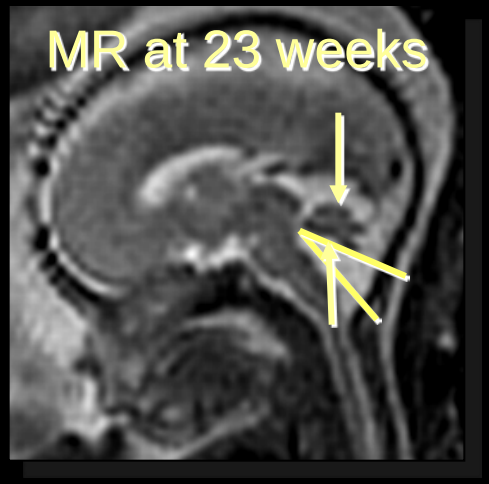
- Inadequate fenestration of Blake's pouch
- Inadequate fenestration of the foramena of Luschka
- Lead to a persistent Blake's pouch
- dilatation of the 4th ventricle
- Inadequate communication with the subarachnoid space

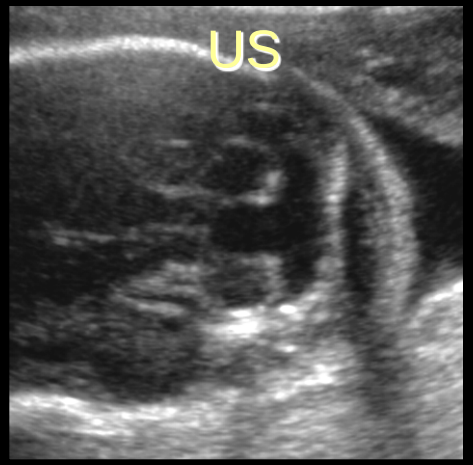
Persistent Blake's pouch
Blake's Pouch Cyst

- Vermis well-lobulated
- Tegmentovermian slightly increased
- Fastigial point normal
- Biometry normal

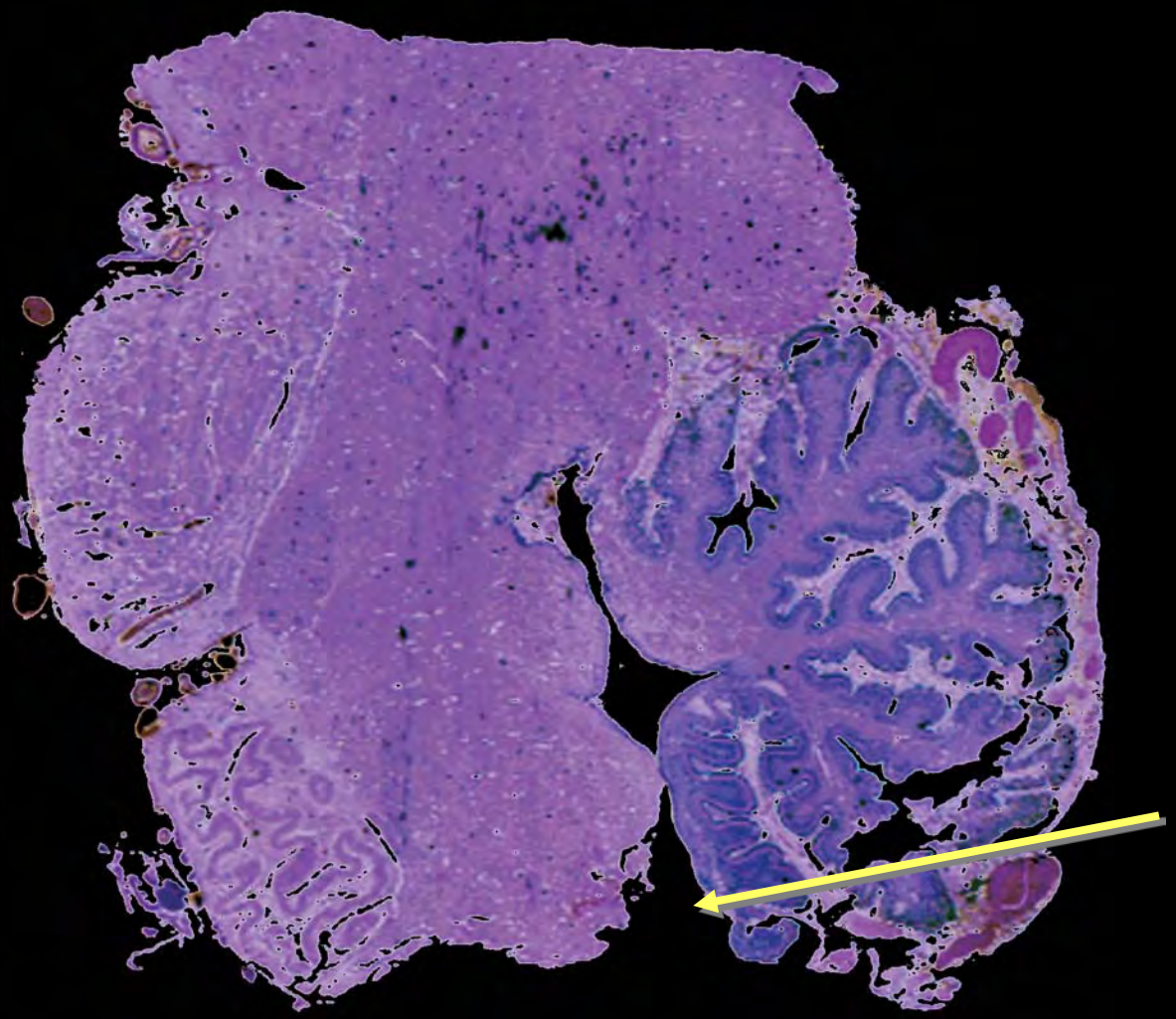


trefoil





Answer



In this fetus that died due to maternal sickle cell disease

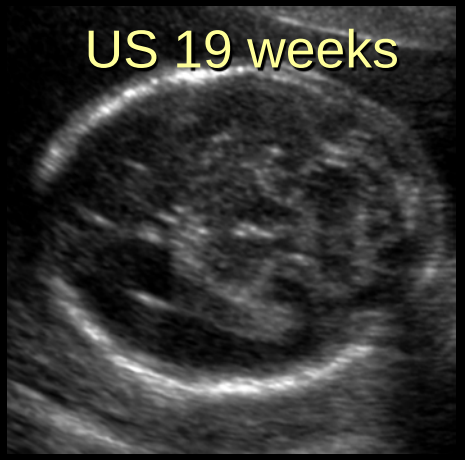
vermis normal histologically

Blake’s pouch collapsed

Normal Blake's metapore = foramen of Magendie

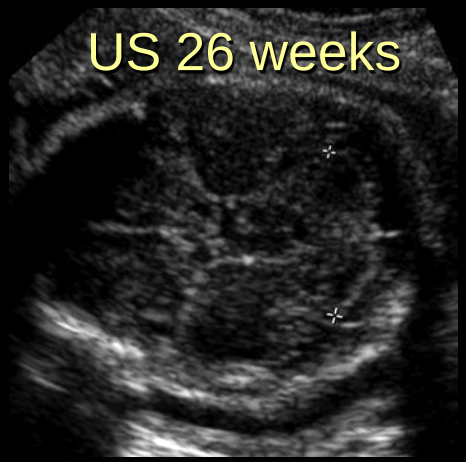
- Vermis well-lobulated
- Tegmentovermian minimally increased
- Fastigial point normal
- Biometry normal

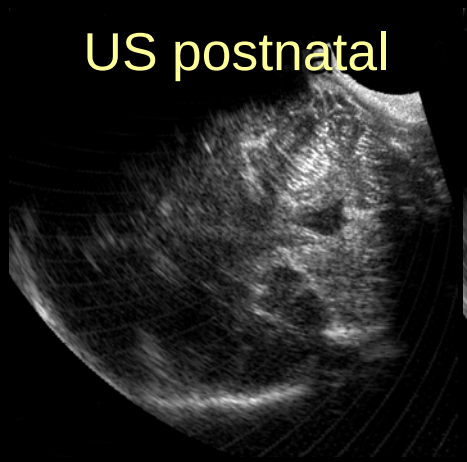
omega

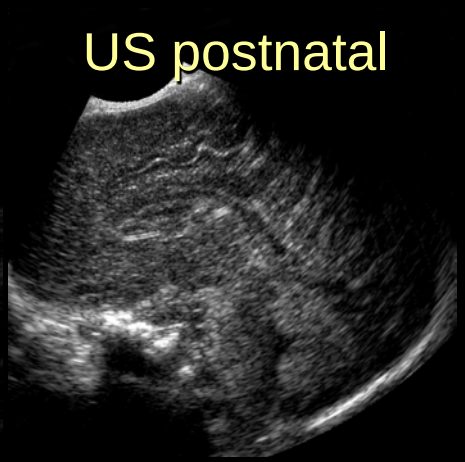


Normal Blake's metapore = foramen of Magendie

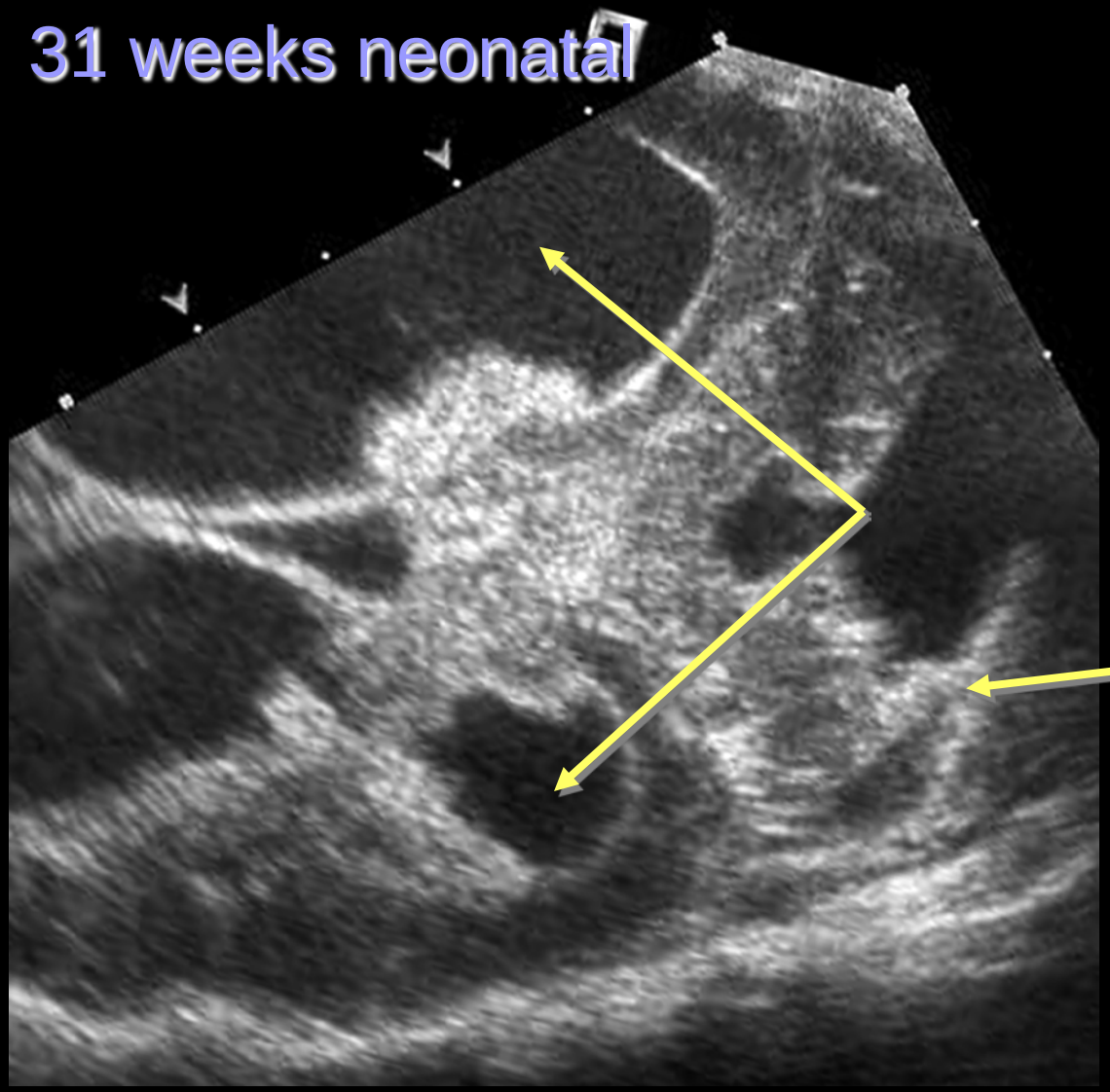
- Vermis well-lobulated
- Tegmentovermian minimally increased
- Fastigial point normal
- Biometry normal







Question



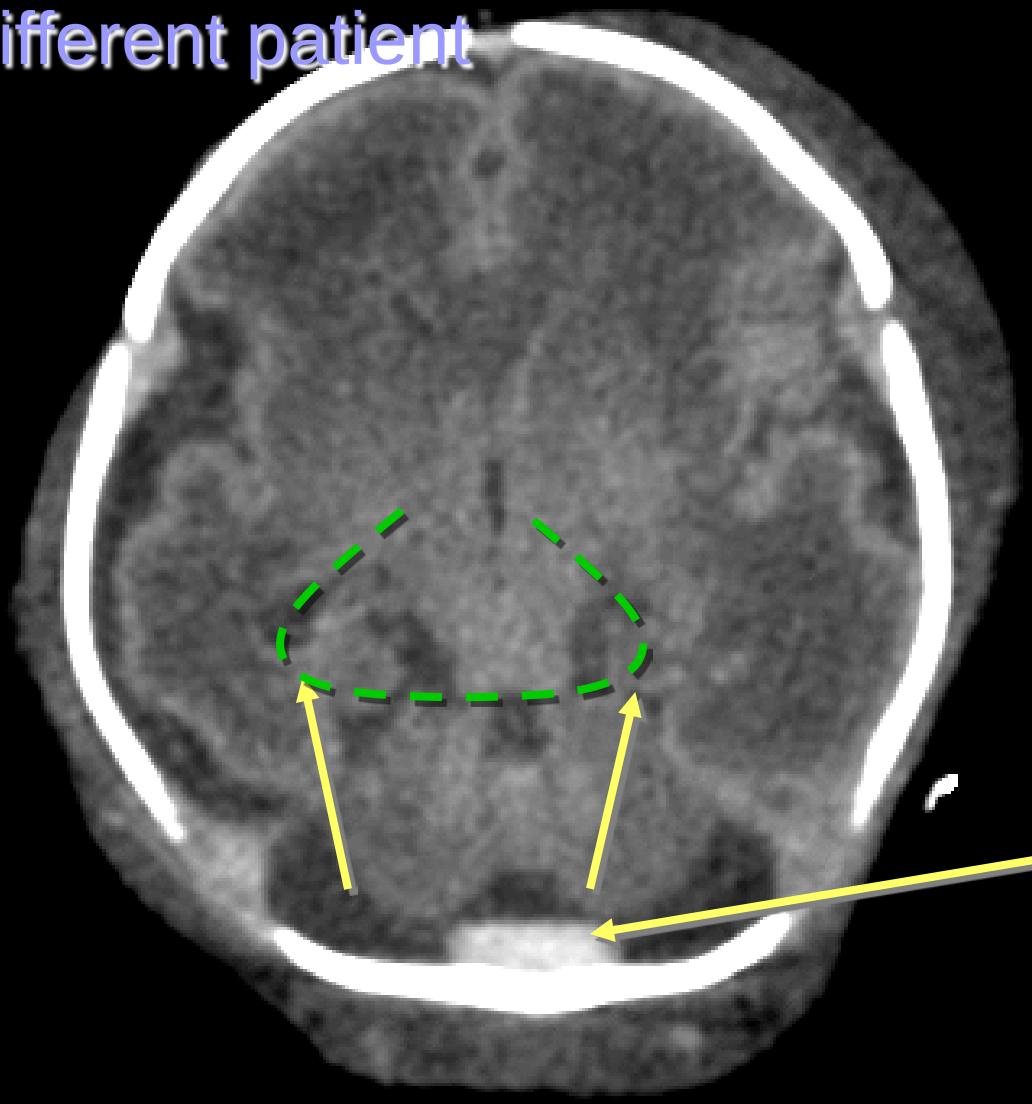
Why does blood in the cisterna magna have a high PPV for development of post-hemorrhagic hydrocephalus?

–Cramer BC, Walsh EA. Cisterna magna clot and subsequent post-hemorrhagic hydrocephalus. Pediatr Radiol. 2001 Mar;31(3):153-9

Blood is not in cisterna magna

Answer

Different patient



Blood “in cisterna magna”

Blood contained laterally

Blood is not in cisterna magna

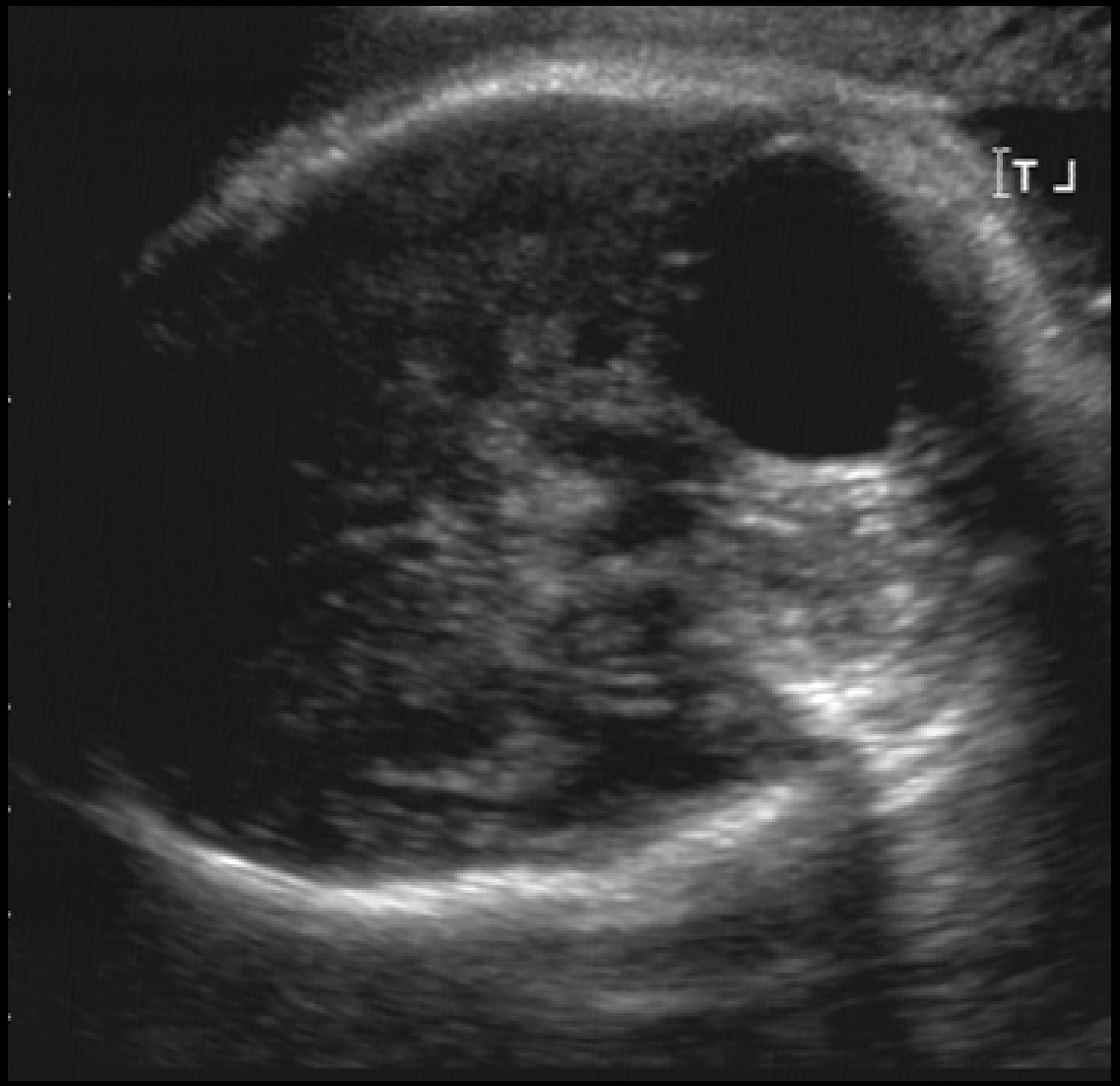
Blood is intra-ventricular

in Blake’s pouch

not resoluble by CT

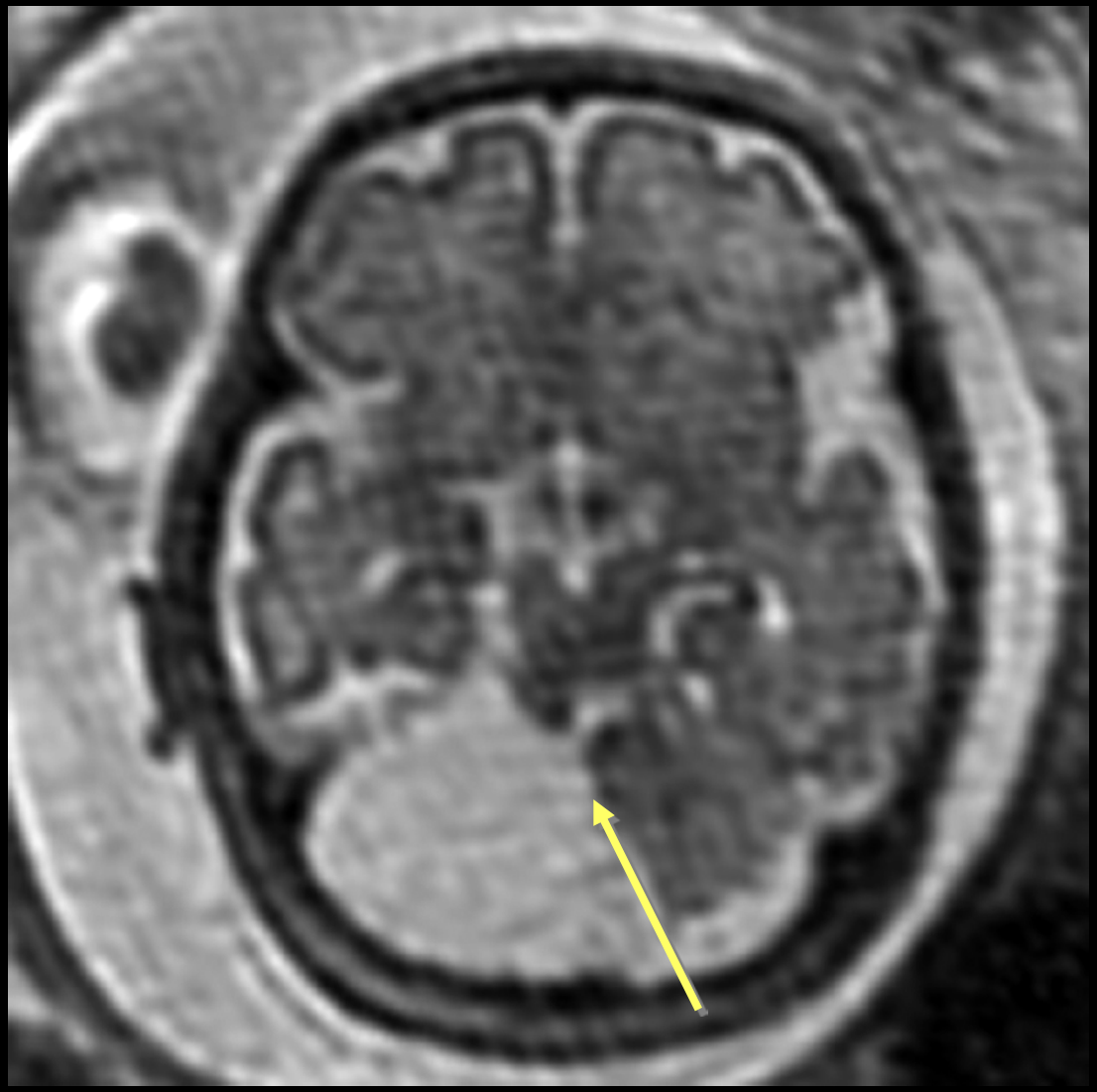
blocks CSF egress

How can we differentiate a Blake's pouch cyst from an arachnoid cyst?



- An arachnoid cyst results from non-coalescence of the cystic areas within the developing subarachnoid space

How can we differentiate a Blake's pouch cyst from an arachnoid cyst?



- An arachnoid cyst results from non-coalescence of the cystic areas within the developing subarachnoid space

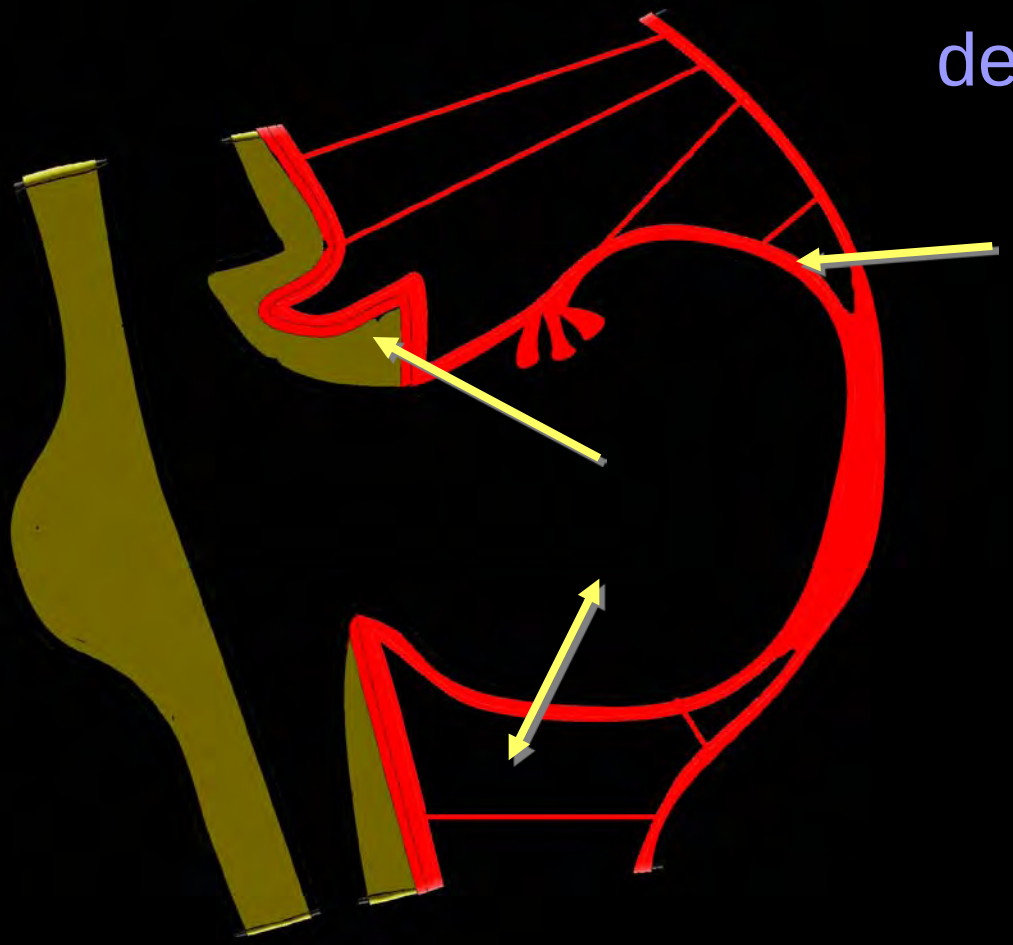
How can we differentiate a Blake's pouch cyst from an arachnoid cyst?

- Arachnoid cyst does not communicate with the 4th ventricle
  - –Thin septum separates them

Conclusions

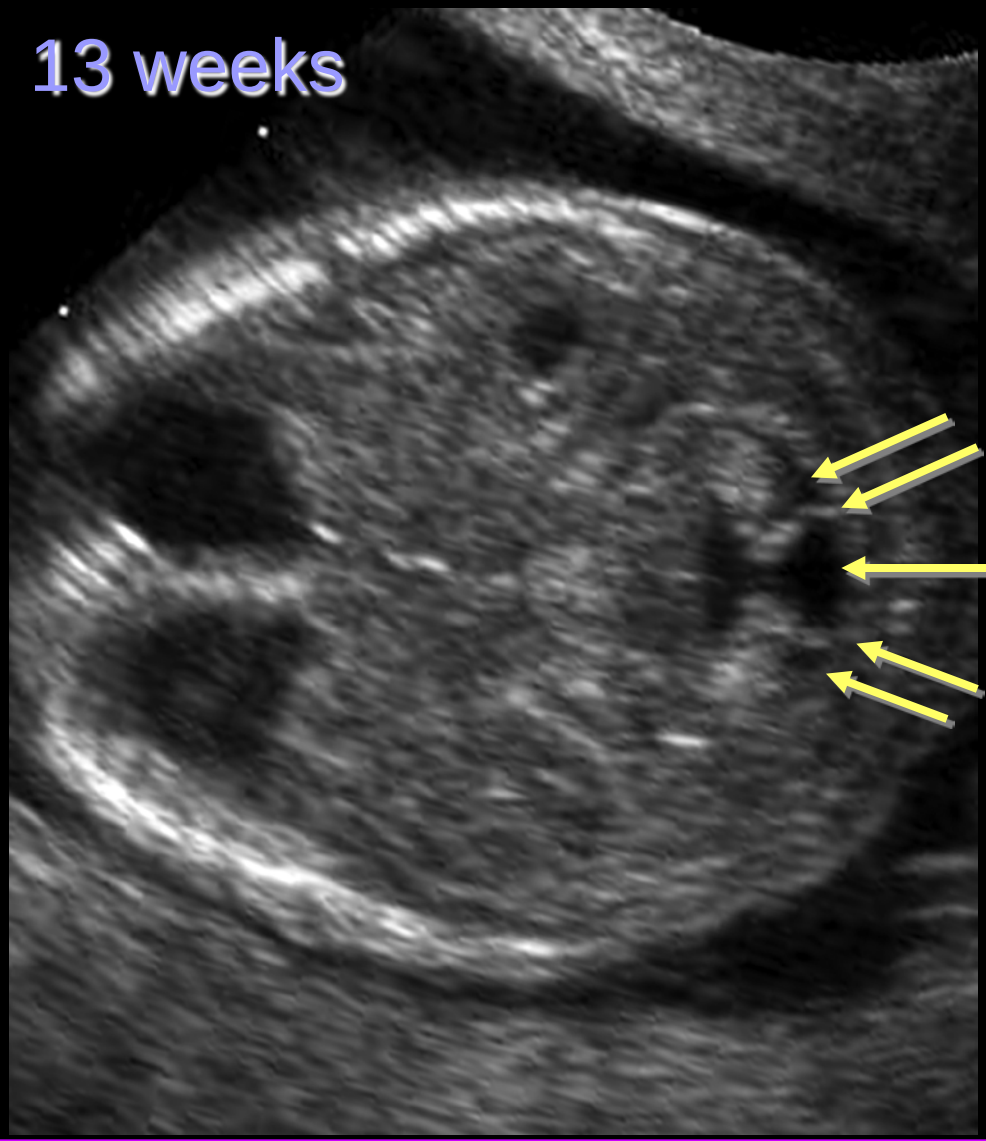
- These findings support and advance current theories that
  - –Blake's pouch cyst
  - –Dandy-Walker continuum
  - –Mega cisterna magna
- are a single spectrum of developmental abnormalities of roof of the rhombencephalic vesicle

Conclusions



- differences probably depend on:
  - degree of dilatation of Blake's pouch
  - degree & timing of fenestration of Blake's pouch and the foramena of Luschka
  - degree of vermian hypoplasia

Conclusions



- Blake's pouch is a normal and persistent structure within the cisterna magna
- The cisterna magna septa represent the walls of the pouch, and mark the boundary between:
- median ventricular-derived compartment
- lateral sub-arachnoid compartments of the cisterna magna
- These septa are a potential new marker for normal development of roof of the rhombencephalon
- Deviation from their normal position should prompt additional assessment in orthogonal planes

The cisterna magna septa
Introduction

- The phrase:

"Posterior fossa cyst communicating with fourth ventricle"

~~Dandy-Walker malformation~~

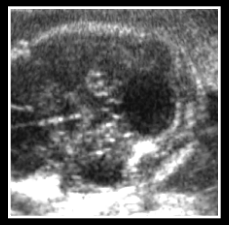
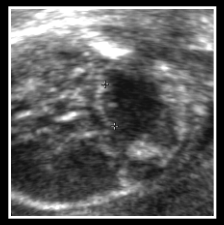
is actually a description of normal developmental anatomy

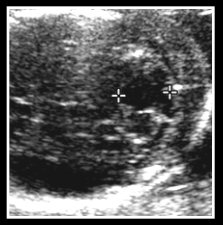
New perspectives

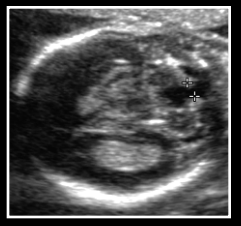
- Armed with our new knowledge of:
  - –Vermian anatomy and development
  - –Cisterna magna anatomy and development
- New perspective on “cystic malformations” of the posterior fossa
- Sonographic morphology of the derivatives of the rhombencephalic vesicle:
  - –Fourth ventricle
  - –Cerebellar vallecula
  - –Blake’s pouch
- “rhombencephalic vesicular derivatives”

Sonomorphology of the rhomebencephalic vesicular derivatives

Prenatal ultrasound

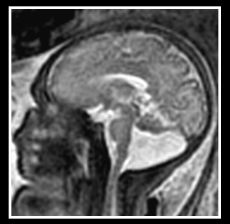


Prenatal magnetic resonance imaging







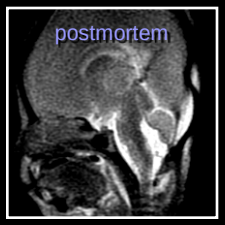
Findings and/or postnatal imaging

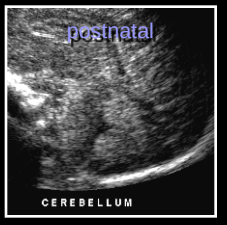
Dandy-Walker continuum - severe with pontine hypoplasia

Microlissencephaly - primitive Z-shaped brainstem shell-like hemispheres smooth brain dismal prognosis

Dandy-Walker continuum - more classic appearance

Wolf-Hirshhorn syndrome - Dandy-Walker continuum with additional anomalies poor prognosis





Outcome

Death from respiratory failure

TOP

TOP

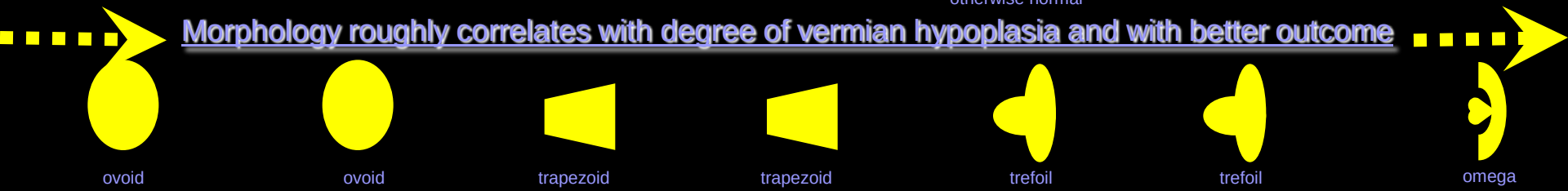
TOP

Dandy-Walker continuum Deaf but otherwise normal

Probably persistent Blake's pouch

Normal including follow-up

Morphology roughly correlates with degree of vermian hypoplasia and with better outcome



Thank you for your attention

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