IN THE NAME OF GOD

CORPUS CALLOSUM

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Overview

- The corpus callosum is the largest of the white matter interhemispheric tracts connecting the cerebral hemispheres.
- These connections are important for the functional integration of sensory, motor, visuomotor, and cognitive processes (language, abstract reasoning, integration of complex sensory information).

Gross anatomy

- The corpus callosum is approximately 10 cm in length and is C-shaped (like most supratentorial structures) in a gentle upwardly convex arch.

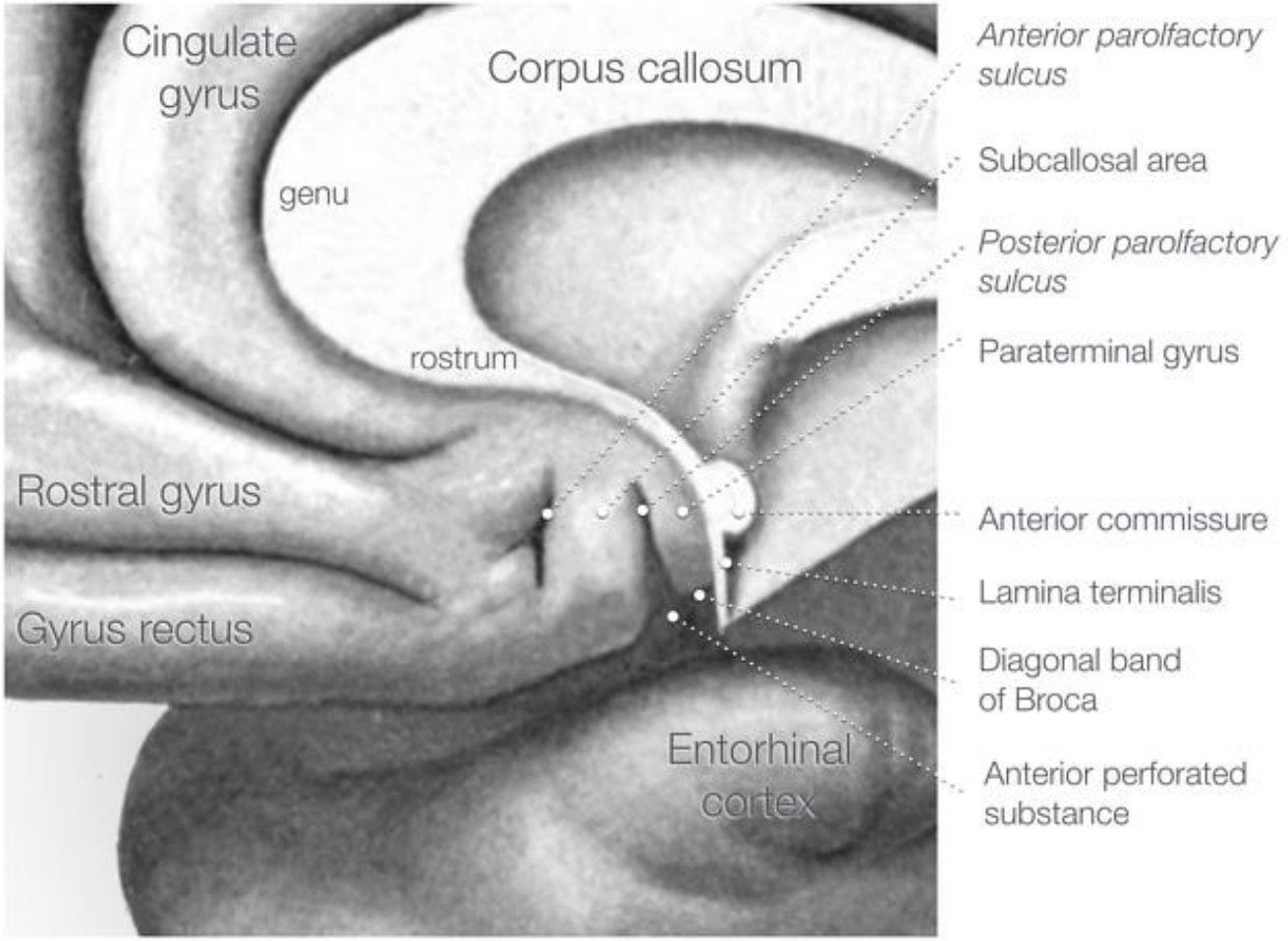
Prenatal diagnosis

Normal appearance:

- The normal appearance of the corpus callosum is a hypoechoic structure located between the cavum septi pellucidi inferiorly and the cingulate gyrus superiorly.
- The pericallosal artery is imaged superior to the corpus callosum when using color Doppler.

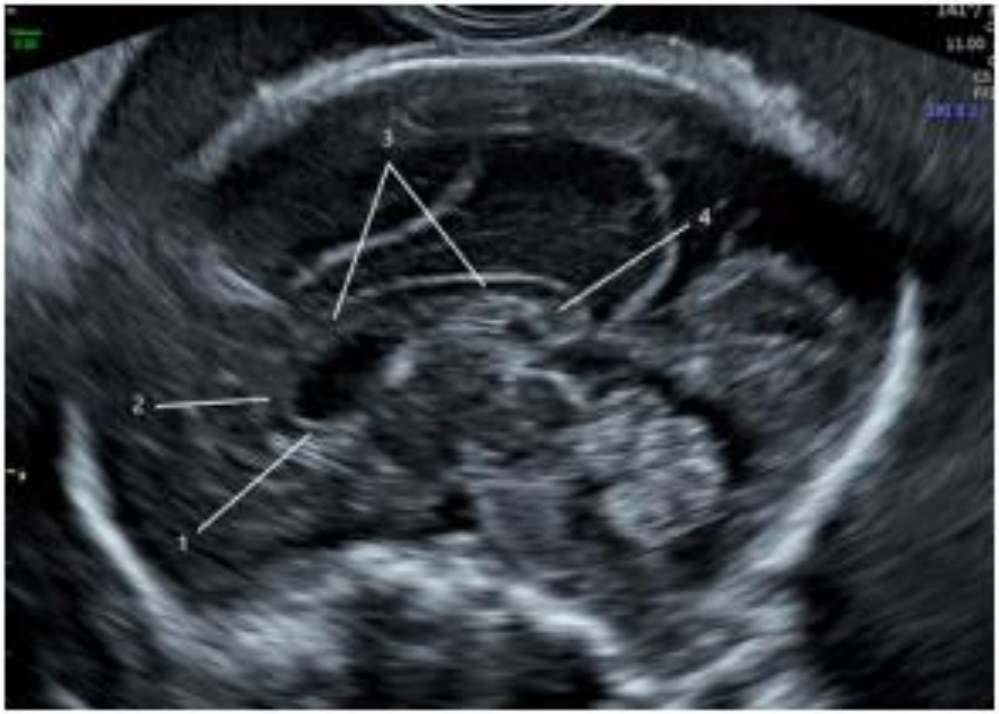
- The CC extends from the frontal lobe anteriorly to above the collicular plate posteriorly.

- Immediately above the body of the corpus callosum, lies the interhemispheric fissure in which runs the falx cerebri and branches of the anterior cerebral vessels.


Cingulate gyrus
Corpus callosum
genu
rostrum
Rostral gyrus
Gyrus rectus
Entorhinal cortex
Anterior parolfactory sulcus
Subcallosal area
Posterior parolfactory sulcus
Paraterminal gyrus
Anterior commissure
Lamina terminalis
Diagonal band of Broca
Anterior perforated substance

- The corpus callosum is composed of four main parts: the rostrum,genu, body,and splenium

Figure 1. Parts of the normal corpus callosum at 28 weeks (1—rostrum; 2—genu; 3—body; 4—splenium).



- Studies on the early development of the CC have been performed postmortem with magnetic resonance imaging(MRI) and immunostaining .
- These studies demon-strated that human pioneering axons originate fromthe cingulate sulcus and that the first wave of midlinecrossing occurs at 12–13 weeks' gestation .

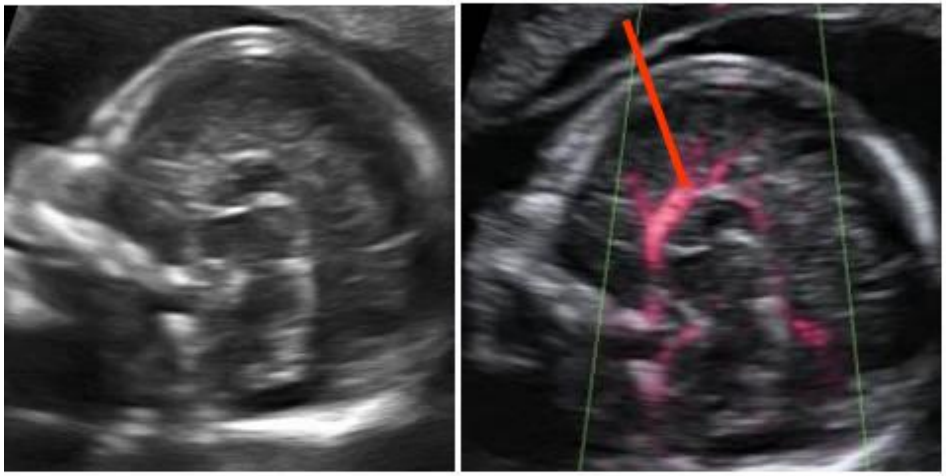
- Most(but not all) researchers agree on the fact that, after the first pioneering axons cross the midline at 12–13 weeks,the CC then forms, following an anteroposterior vector. from genu to splenium, with the significant exception of the rostrum, which is the last part to develop. All the components are visible at 20-22 weeks; then, CC thickness increases until 30 weeks to plateau thereafter.

Timing

- A normal corpus callosum can be seen sonographically by 20-22 weeks of gestation on a median section of the brain and should extend to the region of the quadrigeminal cistern.

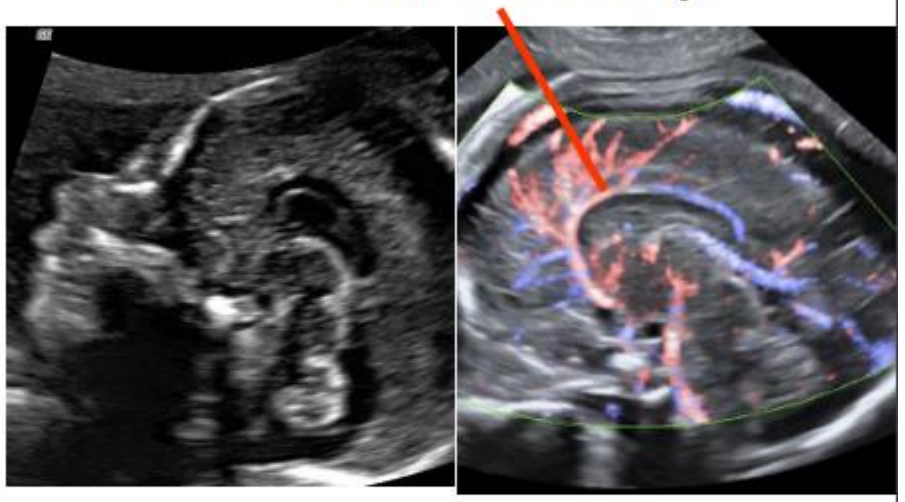
16 weeks

Pericallosal artery

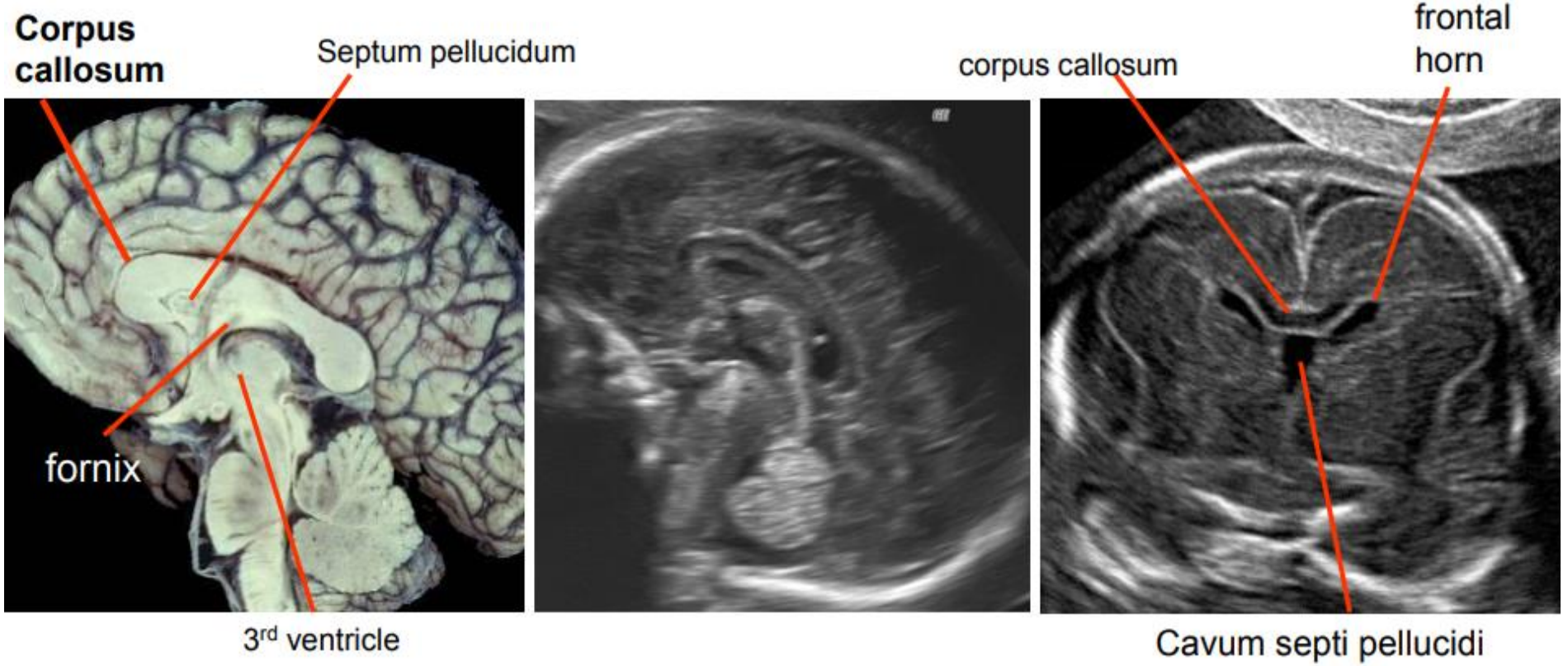


20 weeks

Pericallosal artery



Formation of the corpus callosum is a late event in cerebral ontogenesis. The most anterior part may be seen around 15 weeks' gestation, but the demonstration of the entire structure is usually possible only around 19 weeks' gestation. Visualization is enhanced by color Doppler demonstration of the pericallosal artery


Corpus callosum
Septum pellucidum
fornix
3rd ventricle
corpus callosum
frontal horn
Cavum septi pellucidi

- The formation of the CC starts with the development of the genu; the body and splenium develop at a later stage. If the normal developmental process is disturbed, the CC may be completely or partially absent (hypogenetic).
- Because the developmental process starts from the anterior part and progresses front to rear, when the CC is hypogenetic, usually the posterior portioportion is affected (the posterior body and the splenium).

Fiber tracts

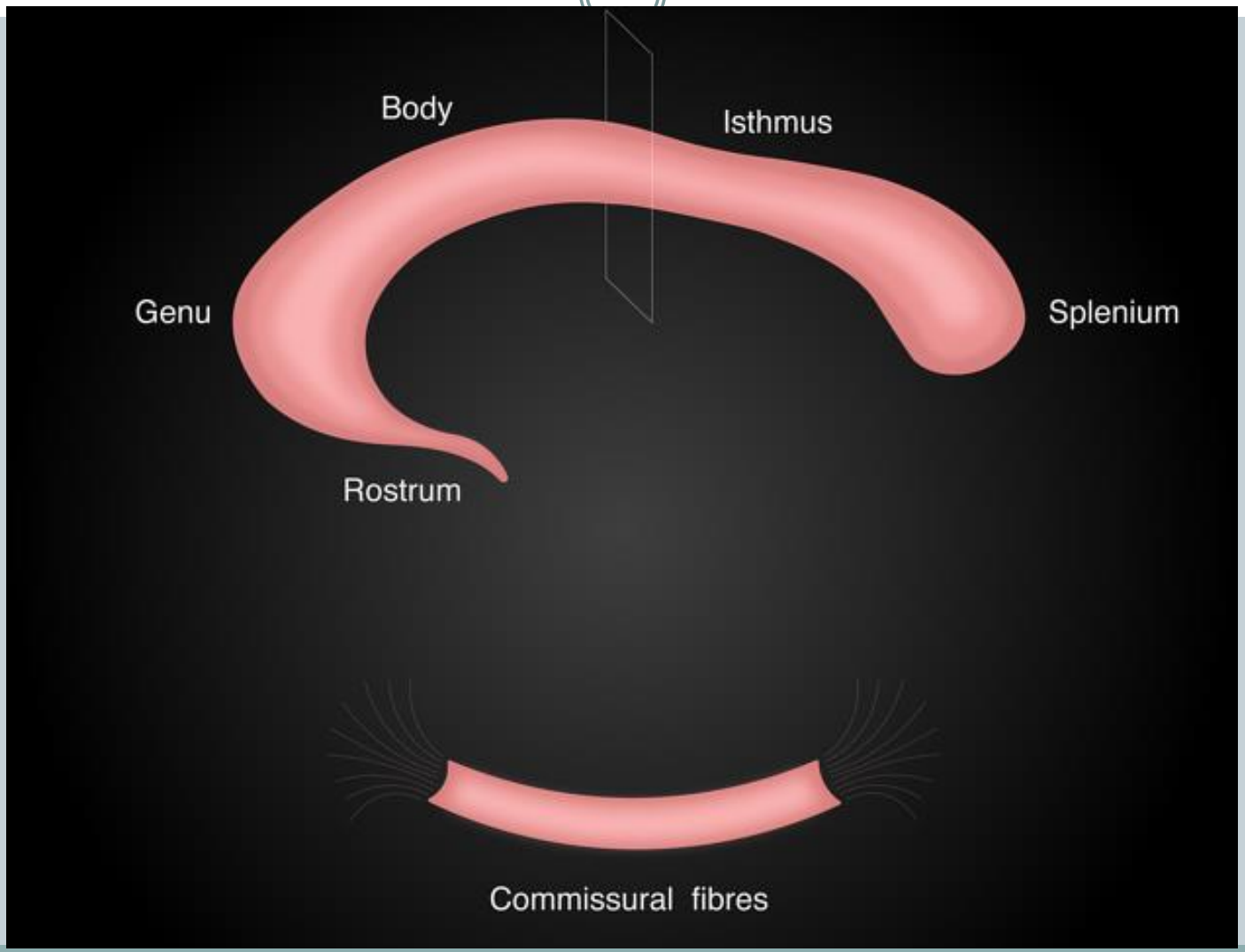
- Although the corpus callosum can be seen as a single large fiber bundle connecting the two hemispheres, a number of individual fiber tracts can be identified. These include:

genu: forceps minor connects medial and lateral surfaces of the frontal lobes.

rostrum: connecting the orbital surfaces of the frontal lobes.

body: pass through the corona radiata to the surfaces of the hemispheres.

splenium: forceps major; connect the occipital lobes.


Body
Isthmus
Genu
Splenium
Rostrum
Commissural fibres

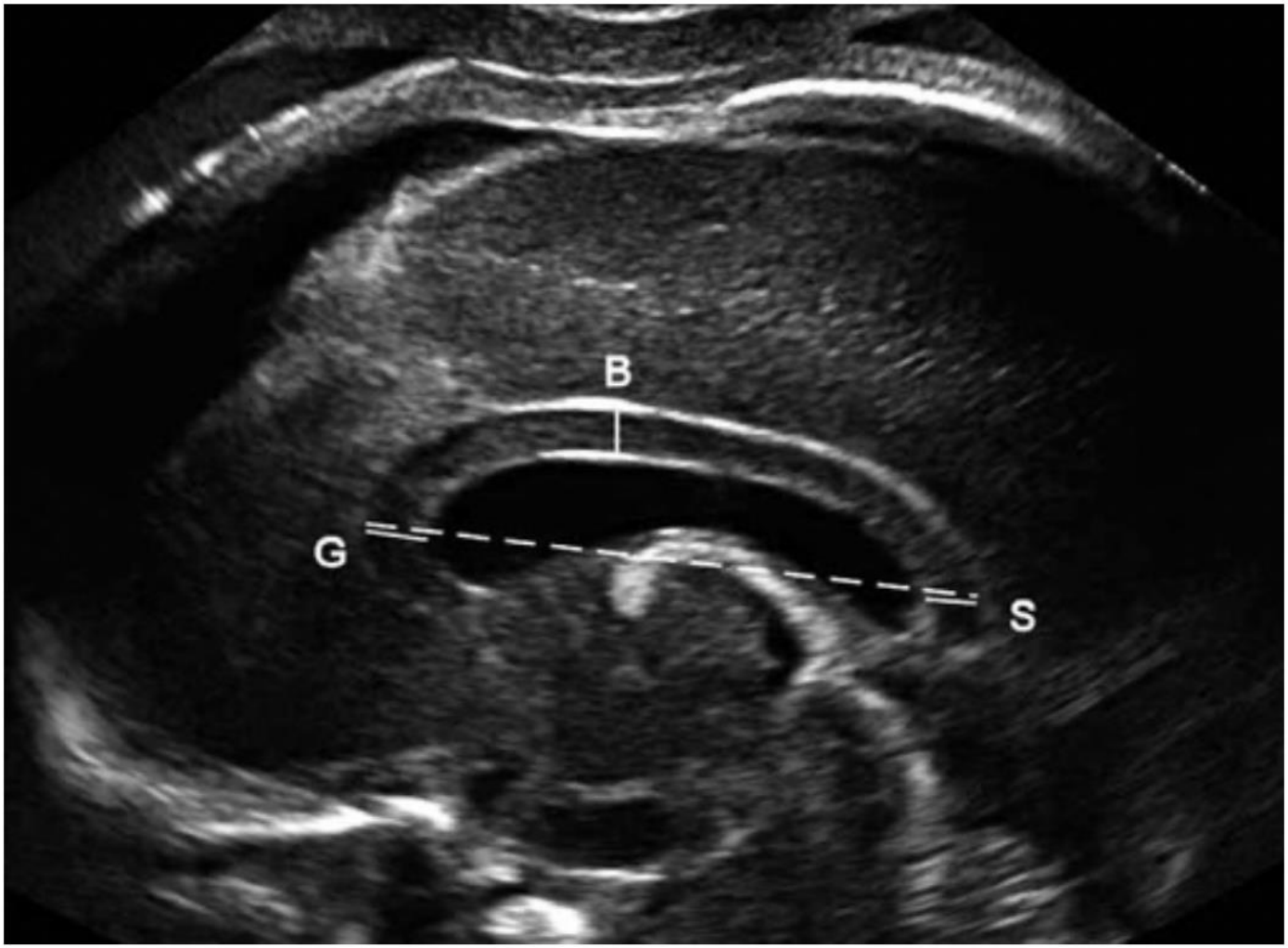
Developmental abnormalities

- Complete agenesis (absence)
- Partial agenesis (hypogenesis)
- Thinning (hypoplasia) – The corpus callosum is normal in its anterior-posterior length, but there is thinning
- Thickening (hyperplasia) – The corpus callosum is thicker than expected.

- Partial agenesis (hypogenesis) – The corpus callosum is shorter in its anterior-posterior length as a result of missing segment(s) such as the splenium and/or the rostrum.

- In primary dysgenesis parts of the corpus callosum which form before the insult will be present whereas later parts will be absent. The presence of the rostrum essentially excludes primary agenesis.

- One apparent exception to this rule is holoprosencephaly in which it is the anterior parts of the corpus callosum which are absent . This has been termed atypical callosal dysgenesis.


B
G
S

Etiology of abnormal development

- Agenesis of the corpus callosum (ACC) is a heterogeneous condition resulting from disruption of multiple developmental steps.

- The cause of the disruption may be genetic, infectious (TORCH infections, Zika virus), vascular, or toxic (fetal alcohol syndrome).

- Genetic factors are most common. Among the genetic causes, a "syndromic" diagnosis is made in 30 to 45 percent of cases and a monogenic cause can be identified in 20 to 35 percent.

Prevalence of ACC

- The reported prevalence of ACC is 1:4000 to 1:5000 live births; however, rates of 2 to 3 percent have been reported among patients with neurodevelopmental disabilities.

AGENESIS OF THE CORPUS CALLOSUM

Incidence. From 0.3%–0.7% in the general population to 2%–3% in the developmentally disabled population.

Ultrasound diagnosis. *Midsagittal view*: Complete or partial absence of the corpus callosum. *Axial views*: Colpocephaly, absence of the CSP (in complete agenesis). *Coronal views*: lateral convexity of and increased distance between the frontal horns.

Risk of chromosomal anomalies. High: 20%.

Risk of nonchromosomal syndromes. High.

Outcome. About 20%–30% rate of significant neurodevelopmental delay in isolated forms has been reported; in nonisolated form the prognosis is poor.

Associated abnormalities

- Corpus callosum anomalies are often associated with other cerebral or extra-cerebral abnormalities.
- Among the cerebral anomalies, ventriculomegaly, Dandy Walker spectrum, heterotopias, and cortical dysplasias may be present.

- Extra-cerebral anomalies include cardiac, facial, and growth abnormalities.
- Chromosomal abnormalities are seen in approximately 20 percent of ACC, and include trisomy 18, trisomy 13, and mosaic 8.

- Genetic syndromes including autosomal dominant (Apert), autosomal recessive (Joubert), and sex linked(Aicardi) have been reported.

When to suspect a corpus callosum abnormality

- During routine screening for fetal anomalies at 20 to 22 weeks of gestation, the two most important clues that the corpus callosum needs further assessment to exclude a callosal abnormality are:
- • Nonvisualization of the cavum septi pellucidi and
- • Ventriculomegaly (lateral ventricles measuring >10 mm).

- Because the CC shares a common anatomic and embryogenetic formation with the septum pellucidum, complete ACC is commonly associated with a hypoplastic or absent CSP.

Absent Cavum Septi Pellucidi

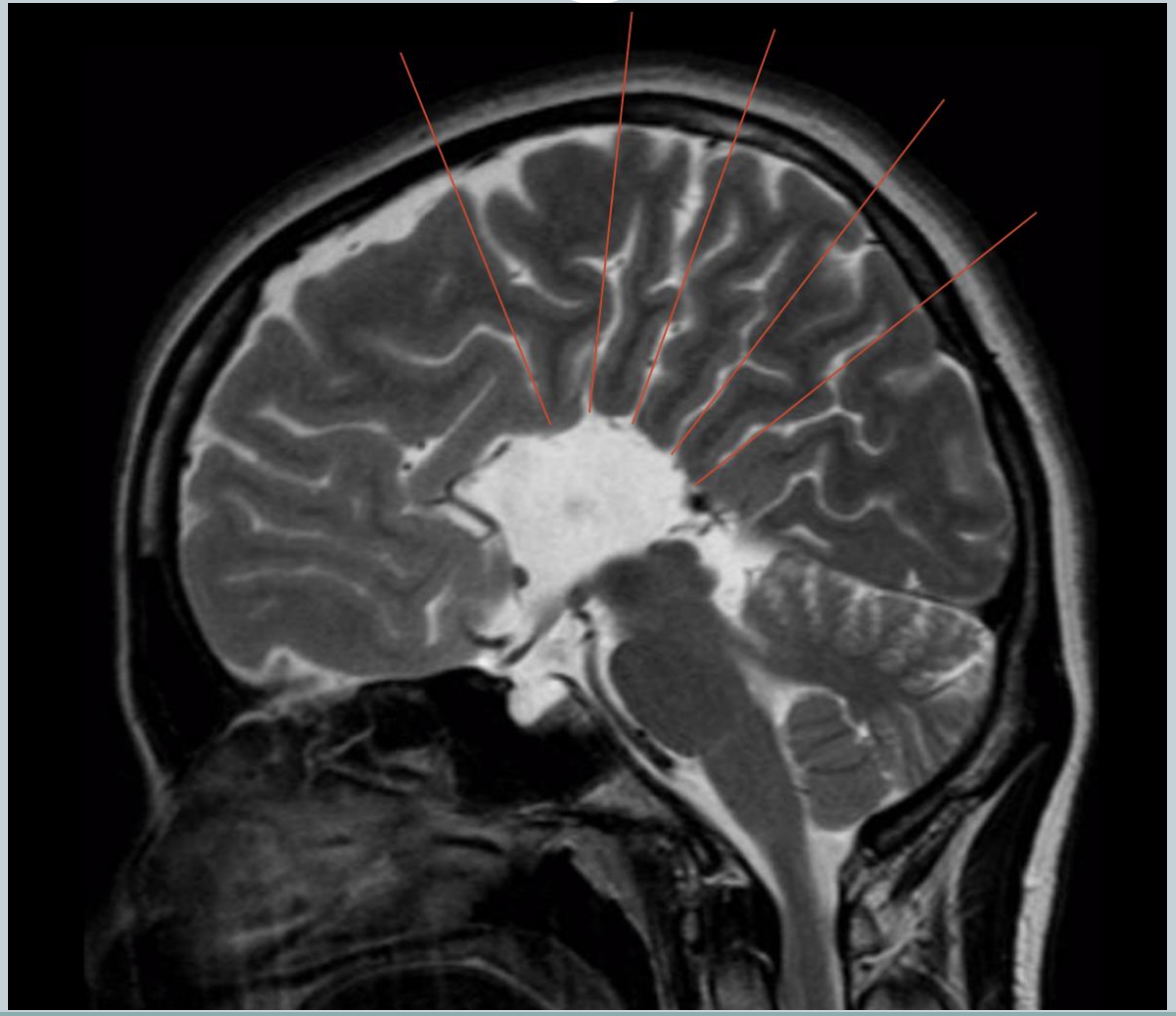
- The cavum septum pellucidum (CSP) is a critical anatomic landmark of normal midline development and should be visualized during biparietal diameter and head circumference measurement. This cerebrospinal fluidefilled cavum typically becomes apparent at 17 weeks' gestation. It gradually closes and is not visualized by the late third trimester.

Sonographic criteria for diagnosis:Agenesis

Direct sonographic features of complete ACC are:

- In a median section, complete absence of the corpus callosum and cavum septi pellucidi; and after 25 weeks, additional findings include absence of the cingulate gyrus and radial array of the sulci, which appear to radiate in a perpendicular fashion from the dilated third ventricle in a "sunburst" pattern.

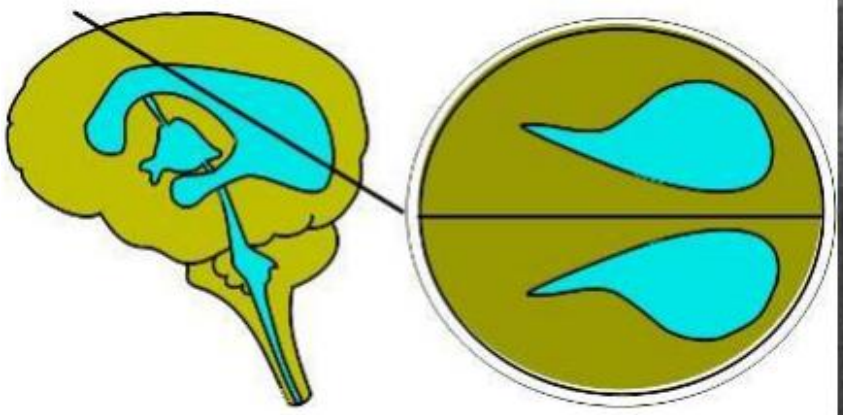
sunburst

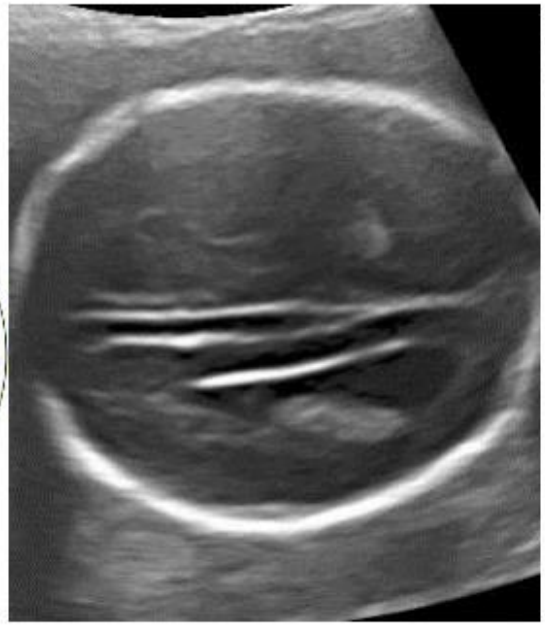


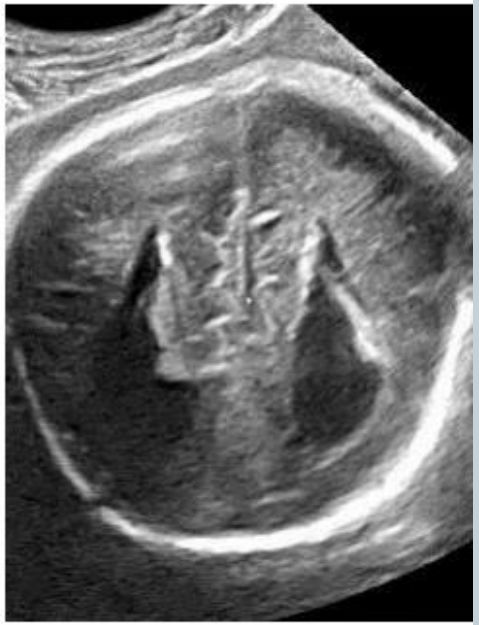
Additional indirect sonographic features can be seen on axial and coronal sections.

- In the axial section, the frontal horns appear narrow and laterally displaced, and the atria and occipital horns are slightly dilated (colpocephaly); the shape is similar to a teardrop.

The tear-shaped lateral ventricles: a typical finding of agenesis of the corpus callosum







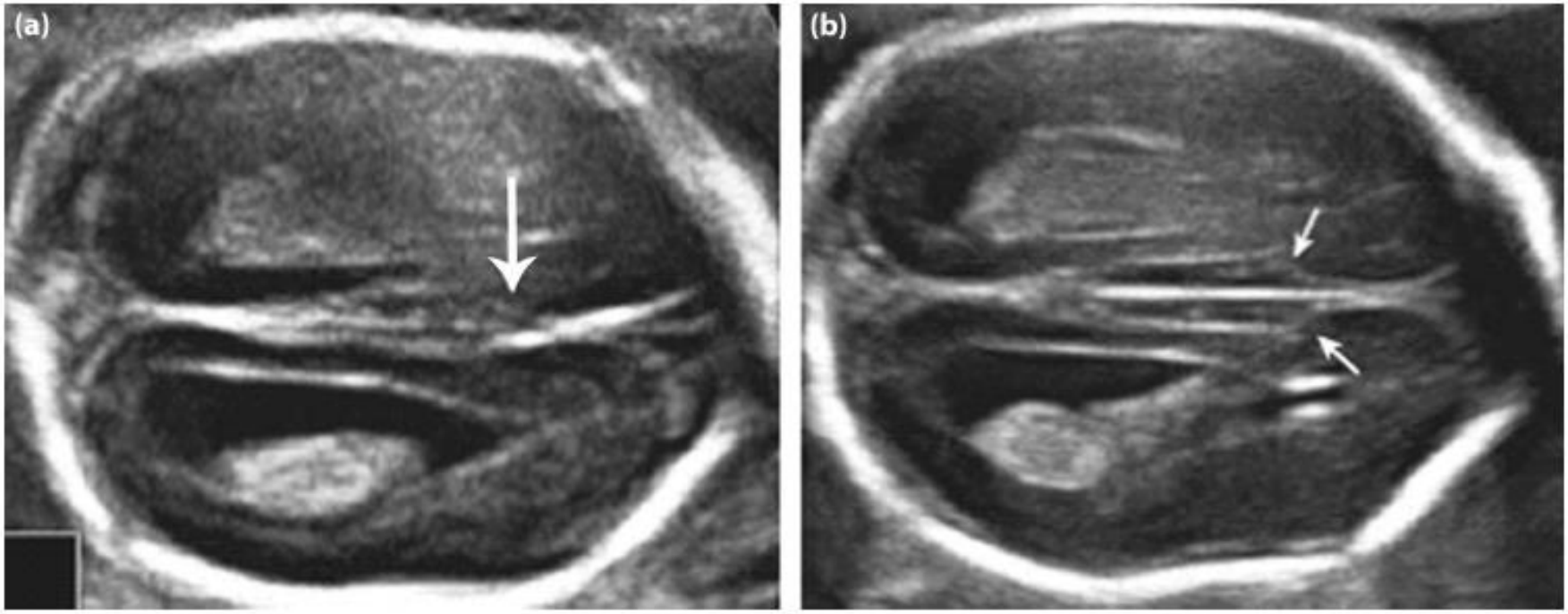
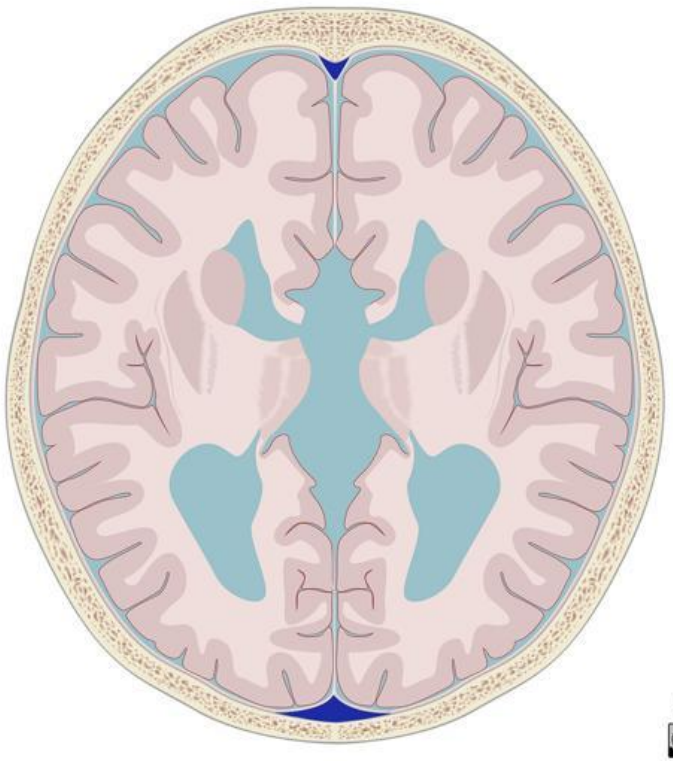


Figure 2.24 Agenesis of corpus callosum. Indirect signs: (a) axial scan of a 22-week fetal brain showing the absence of the CSP (arrow) and the teardrop shape of the lateral ventricle owing to dilatation of the atria and occipital horns (colpocephaly). (b) Arrows indicate increased separation of the hemispheres with the bodies of the lateral ventricles parallel to each other and shifted laterally. Colpocephaly is also present.

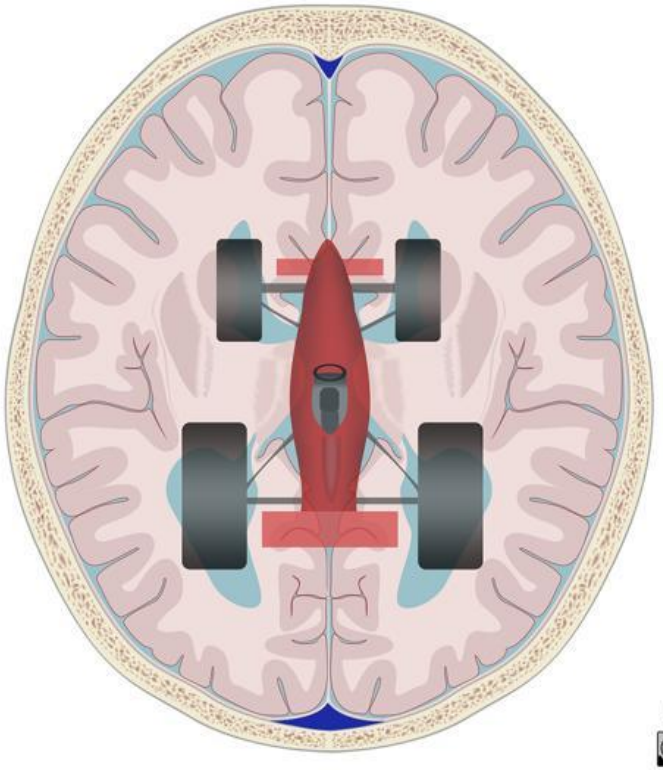
Racing car sign

of corpus callosum agenesis



Racing car sign

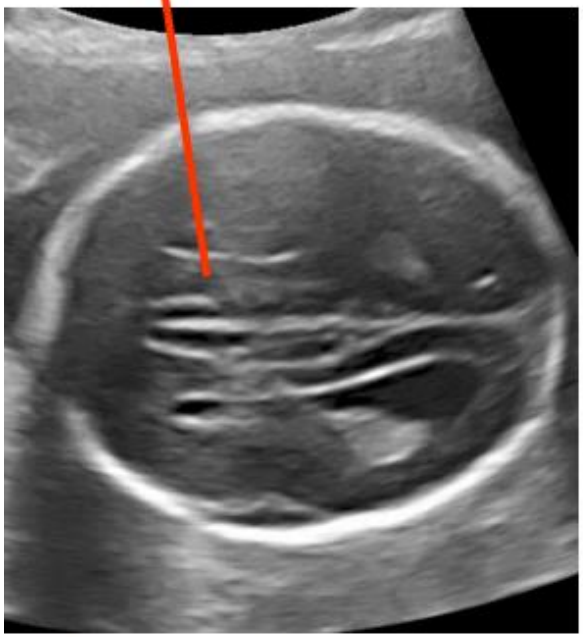
of corpus callosum agenesis

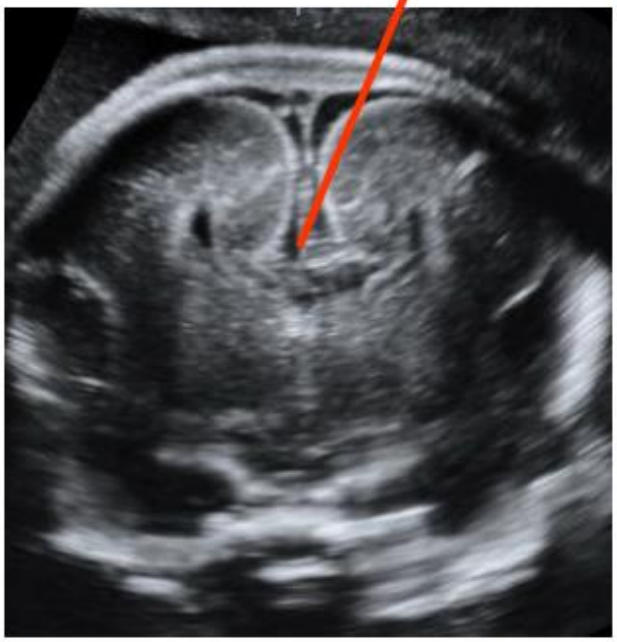


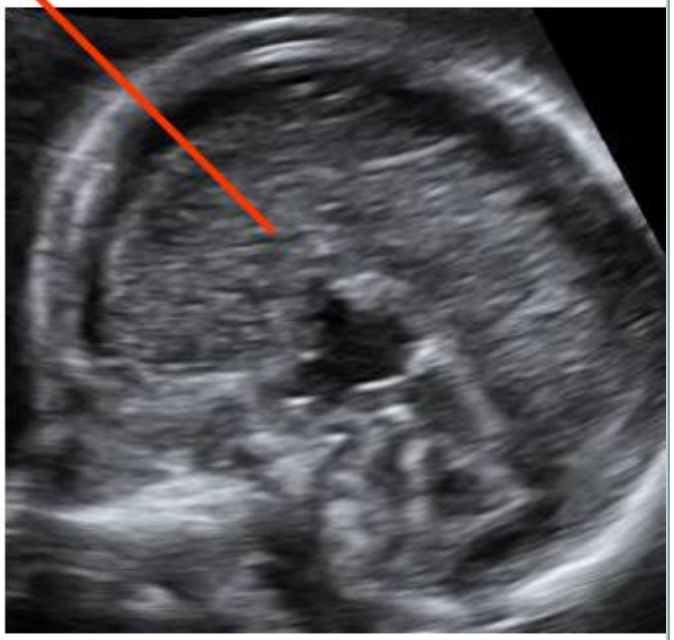
- On coronal section, the falx cerebri can be seen in a broad interhemispheric fissure which meets the third ventricle; the lateral ventricles are widely separated and vertically oriented ("Viking's helmet" sign). The thalami may be widely separated due to a dilated third ventricle.

No cavum septi pellucidi, wide interhemispheric fissure

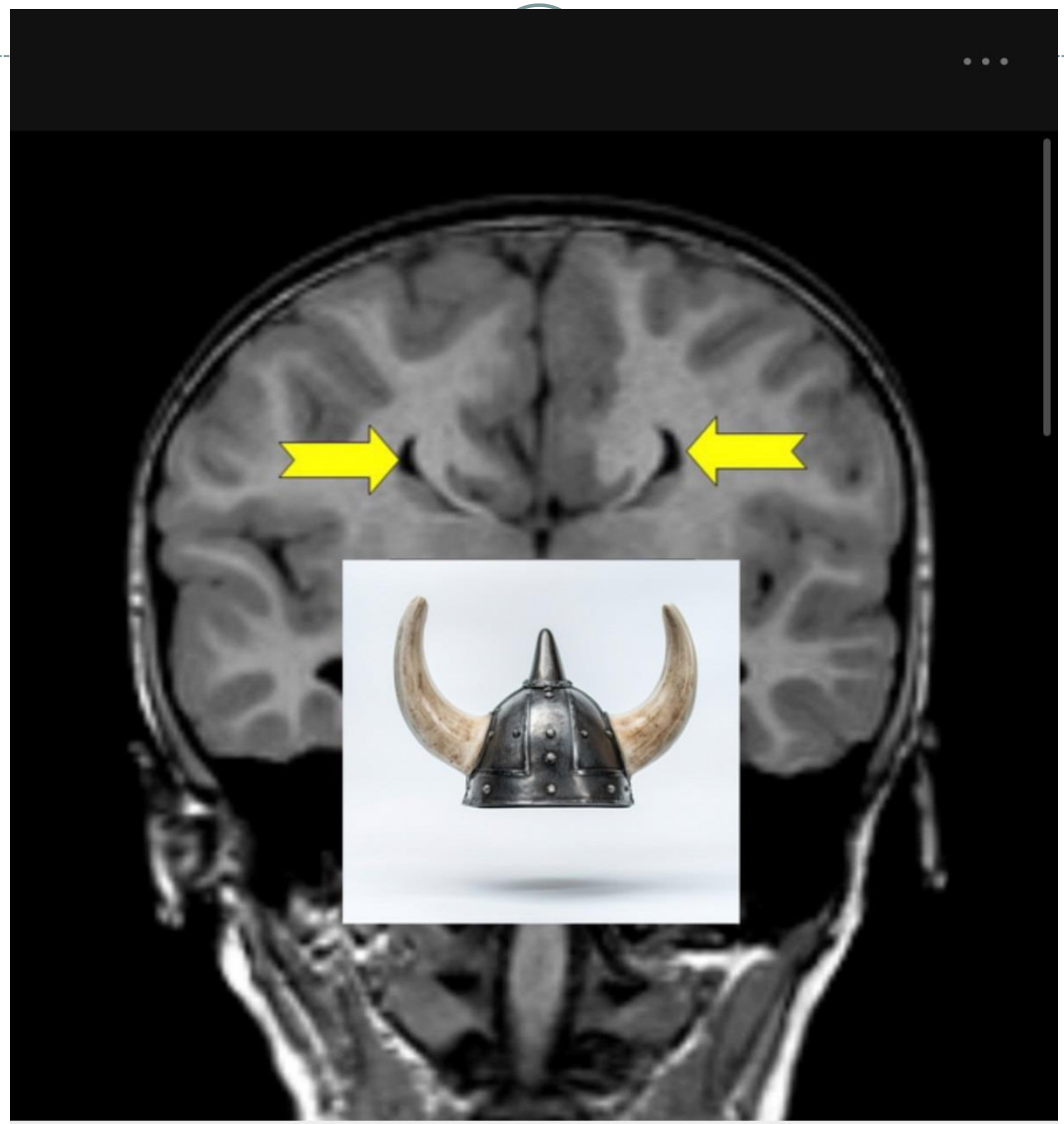
no corpus callosum and cavum septi pellucidi above the third ventricle

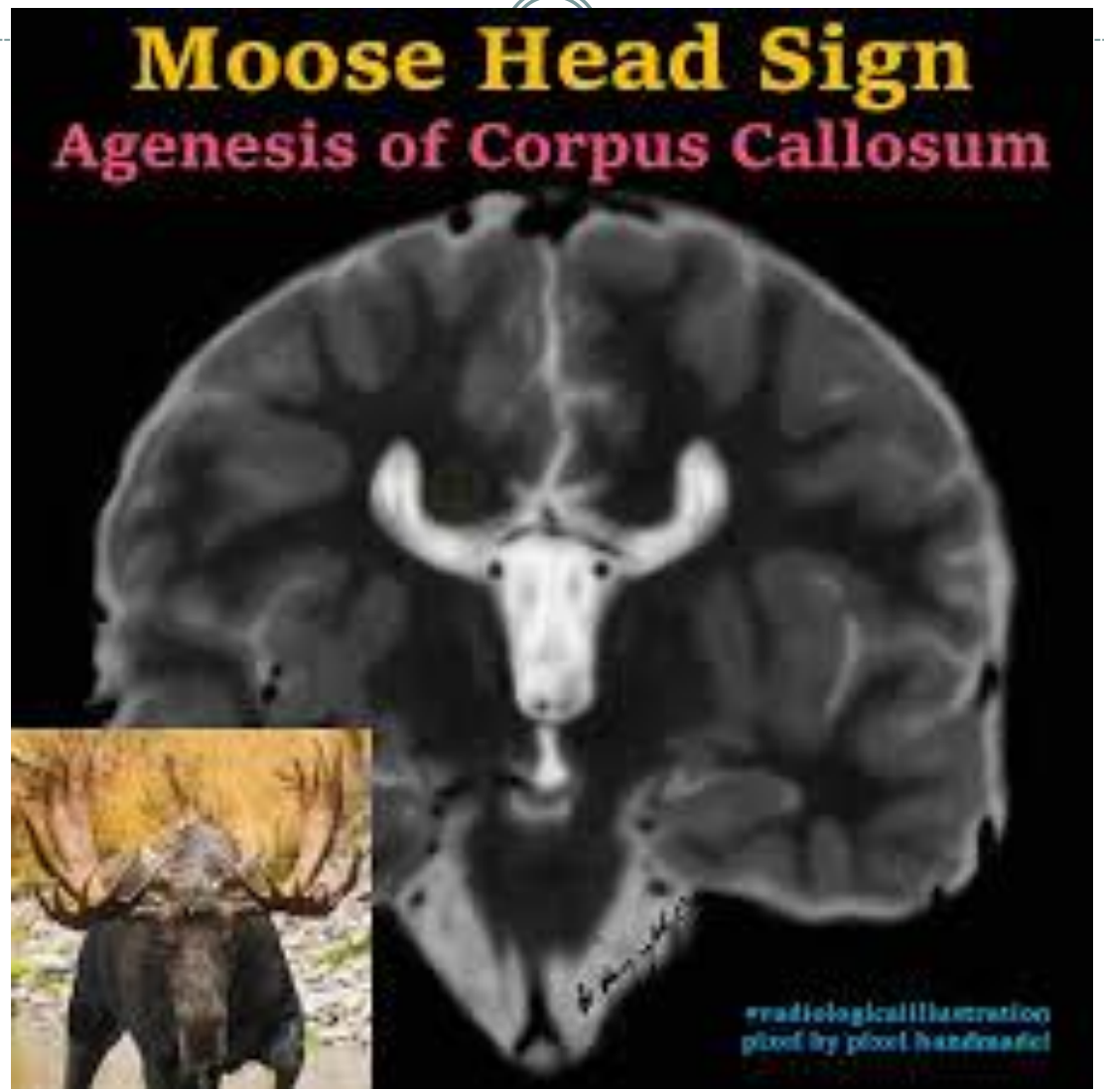


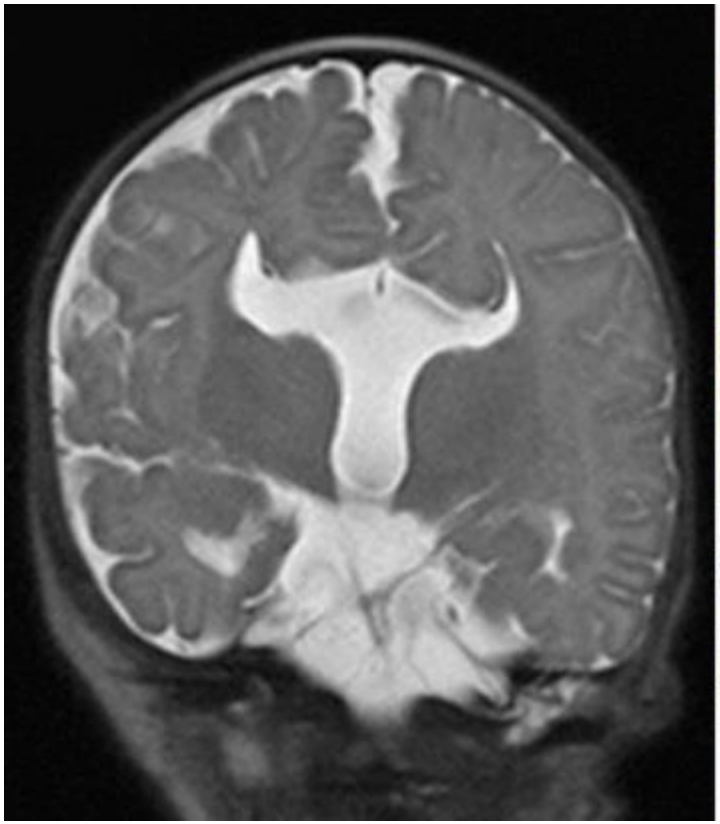


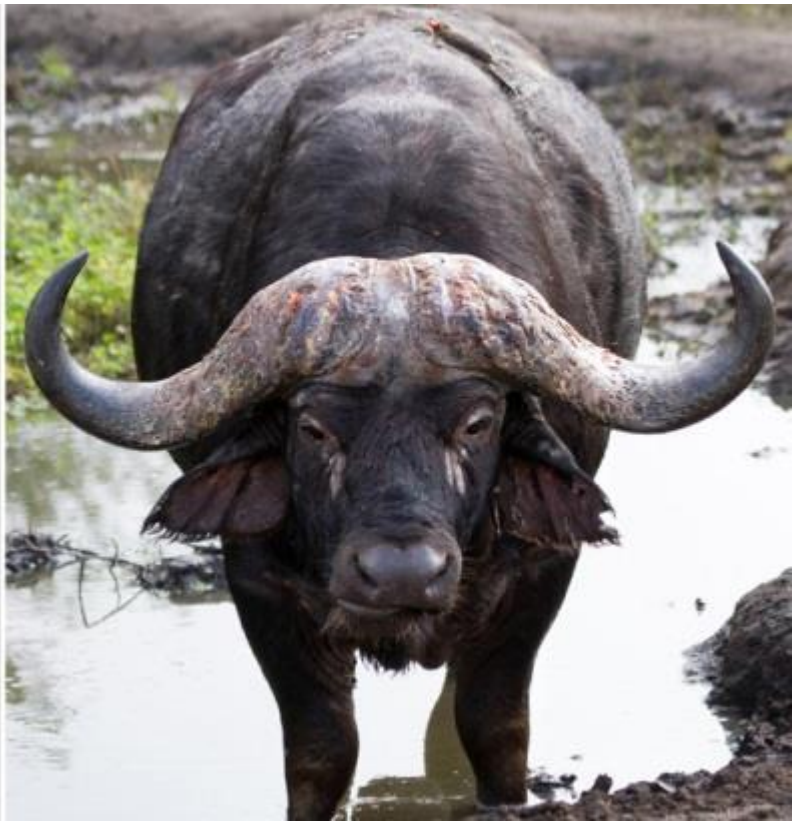


Viking's helmet" sign




Moose Head Sign
Agenesis of Corpus Callosum





KEY DIAGNOSTIC FEATURES

- • Colpocephaly on the axial section
- • Absent cavum septi pellucidi
- • Absent corpus callosum on median brain section
- • Absent or abnormal pericallosal artery on median brain section with color/power Doppler
- • Teardrop-shaped, parallel lateral ventricles on axial section
- • Third-trimester “sunburst” sign—radial gyri and sulci on the median surface seen on the median section
- • Widely spaced, upward-pointing anterior horns (“Viking helmet” sign) on coronal section

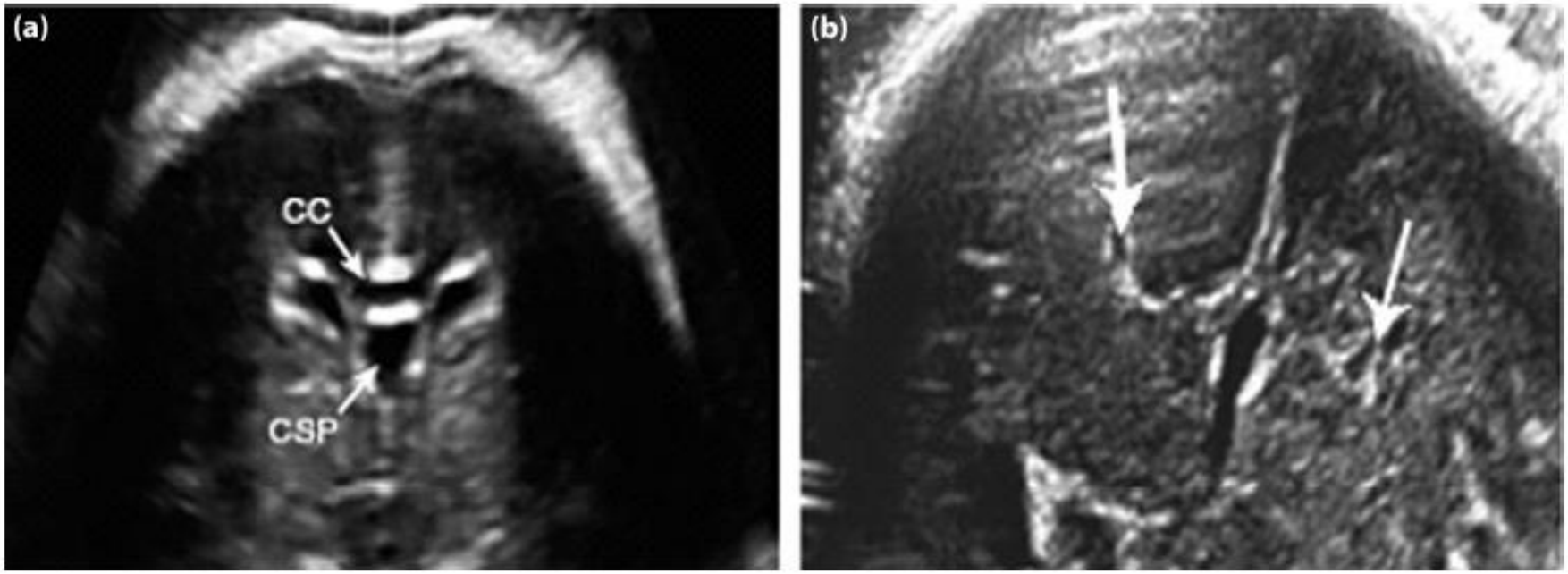
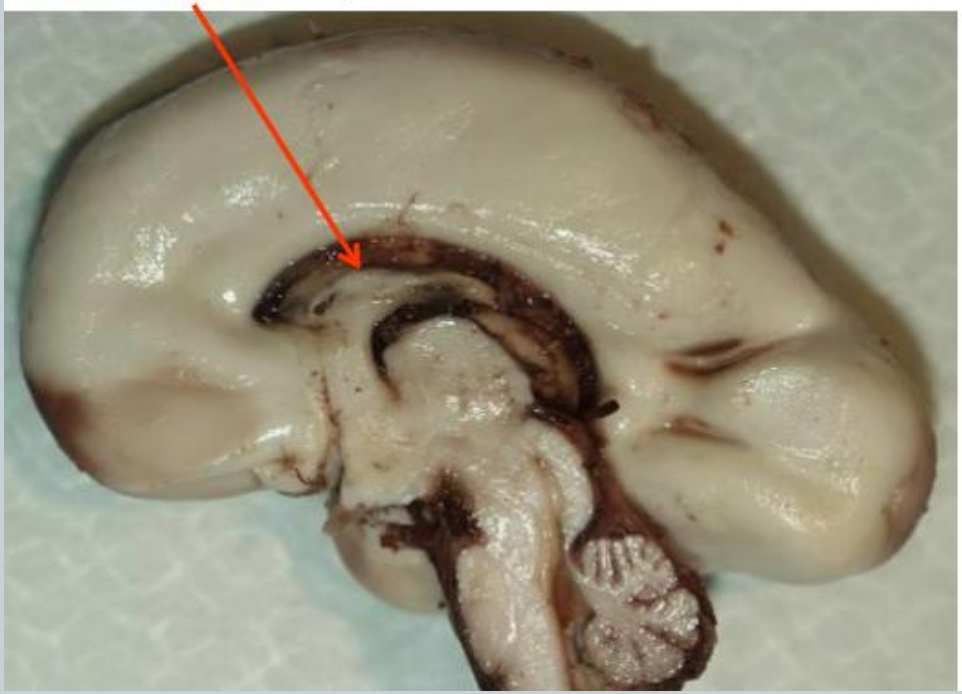


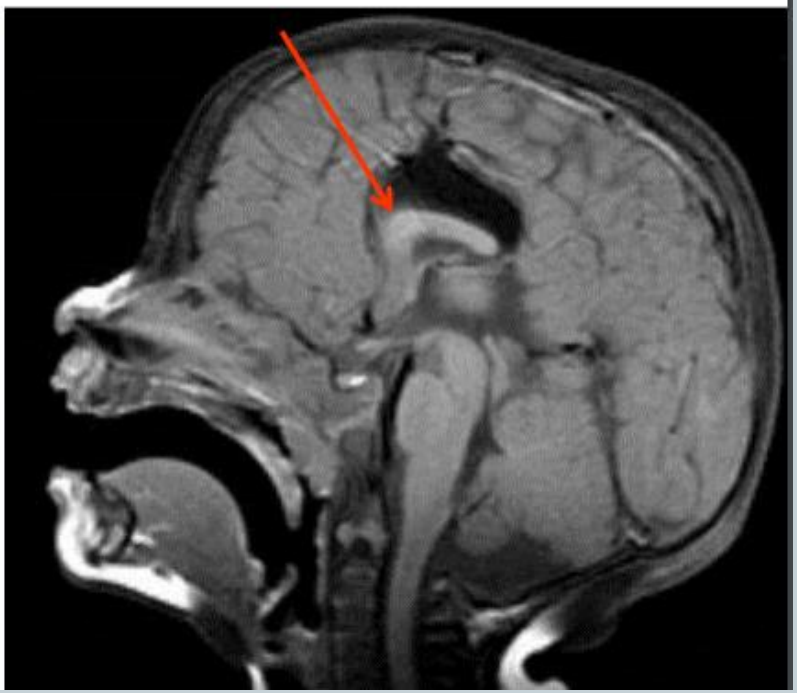
Figure 2.25 (a) Midcoronal view of the brain in a 23-week normal fetus shows the normally developed frontal horns, genu (CC), and cavum septi pellucidi (CSP). (b) Agenesis of the corpus callosum (ACC). Coronal scan of a 29-week fetal brain shows absence of the genu and increased distance between the frontal horns (arrows). The inner walls of the frontal horns are concave medially because of the medial compression exerted by the Probst bundles. Together with the lumen of the third ventricle, this makes up the typical appearance of a "bull's head."

Partial agenesis (hypogenesis)

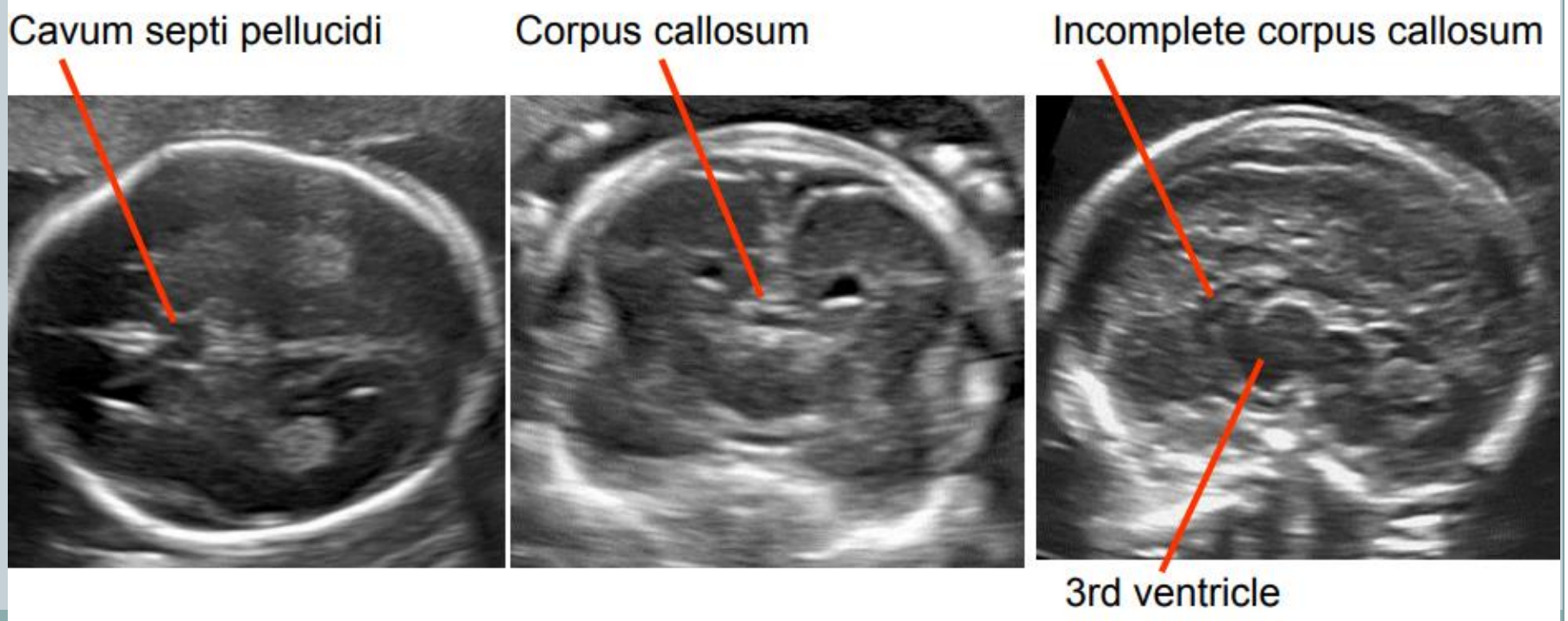
- The sonographic features of partial agenesis (hypogenesis) are more subtle, but the key feature is the shorter anterior-posterior length of the corpus callosum seen in the median section of the fetal brain. The splenium and rostrum as well as multiple segments of the corpus callosum can be absent or dysmorphic.

Incomplete corpus callosum





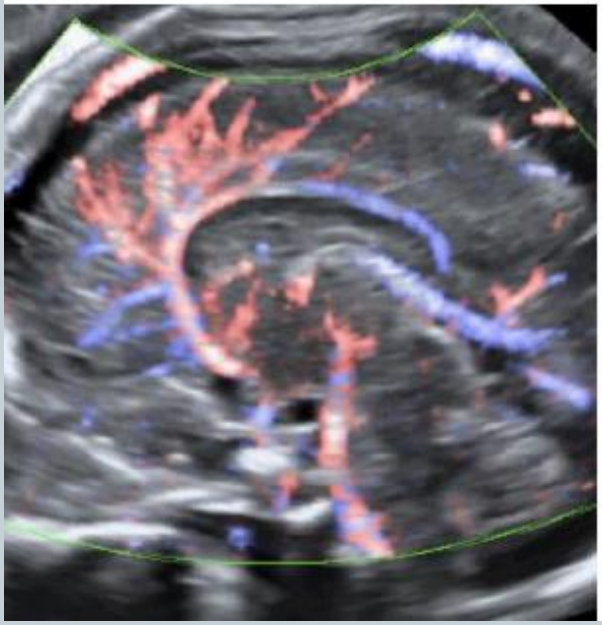
Sonography of partial agenesis



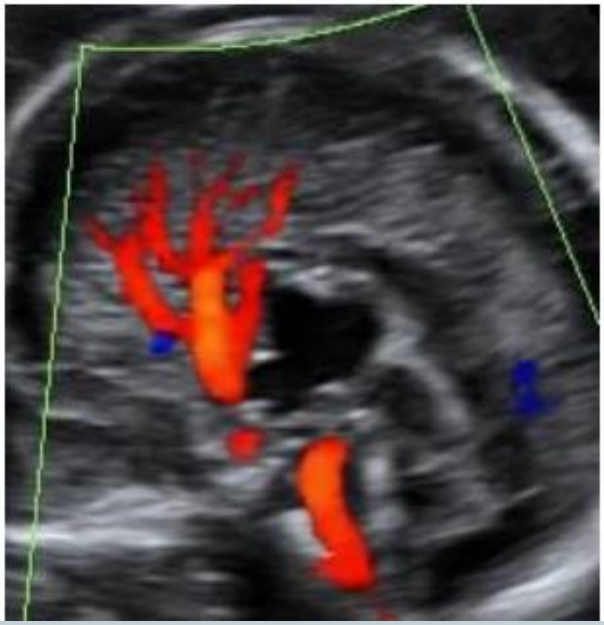
- Diagnosis of partial agenesis is challenging since the cavum septi pellucidi is almost always present. The cavum septi pellucidi can be present and normal in appearance, dysmorphic, or unusually short and wide.

Color Doppler of the cerebral circulation

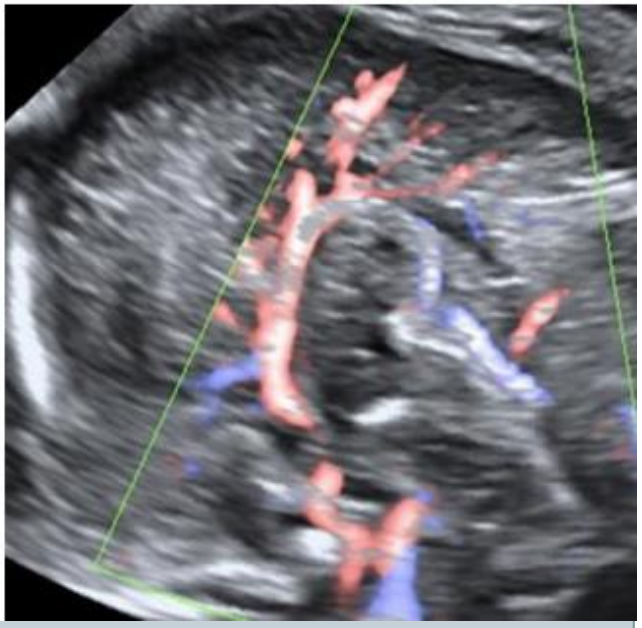
Normal



Complete agenesis



Partial agenesis



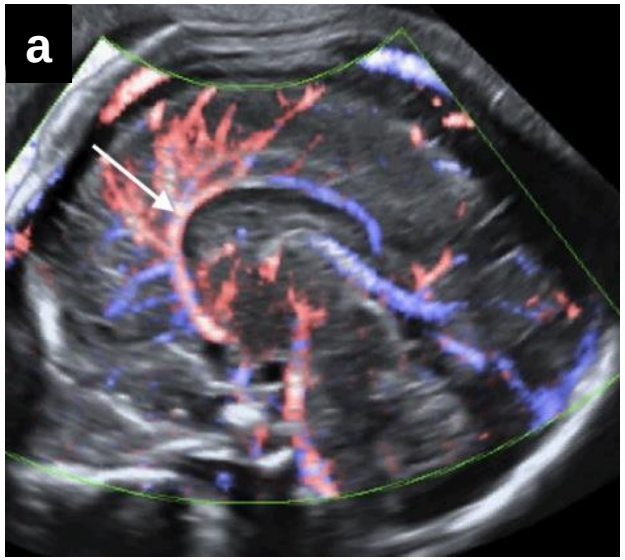
Pericallosal artery

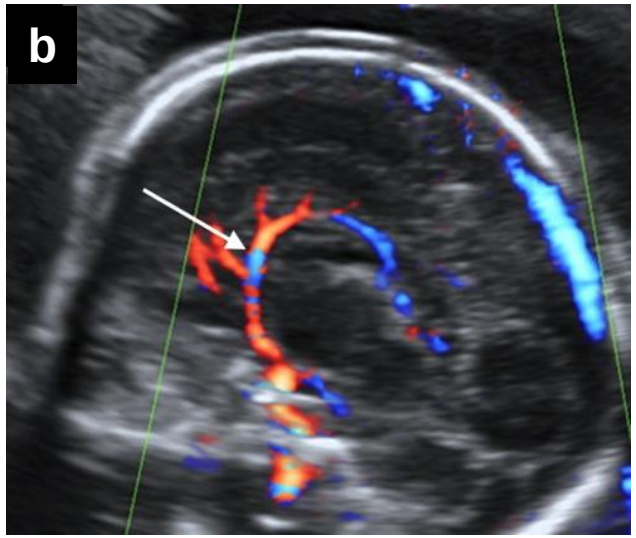
• The pericallosal artery (arrows) is demonstrated by color Doppler in the mid-sagittal view

• Identification of the pericallosal artery highlights the presence of the corpus callosum

• Appearance of the pericallosal artery in a normal fetus at 20 weeks' gestation (a)

• Partial agenesis of the corpus callosum inferred by a partial Doppler signal from the pericallosal artery where the presence of the corpus callosum was difficult to assess in b-mode (b)





Thin corpus callosum (hypoplastic)

- Sonographic diagnosis of a thin corpus callosum (hypoplastic) is made when the anterior-posterior length of the corpus callosum is normal, but the body appears thin.

Thick corpus callosum

- The prenatal diagnosis of a thick corpus callosum is a rare finding, and its significance prenatally is uncertain.
- Among 59 fetuses with suspected callosal anomalies, 9 were diagnosed with isolated thick corpus callosum at 21 to 29 weeks. All 9 had a normal karyotype.

Role of magnetic resonance imaging

- Magnetic resonance imaging (MRI) is most helpful after the 20 week of gestation, since approximately 20 percent of apparently isolated cases diagnosed by ultrasound have associated CNS anomalies on MRI .

Associated abnormalities:

- Chromosomal abnormalities (trisomies 8, 13 or 18, deletions and duplications) are found in 20% of cases.
- In about 50% of cases there is an association with any one of 200 genetic syndromes, defects in the central nervous system (mainly abnormal gyration, midline arachnoid cysts, Dandy-Walker complex and encephalocele) or defects in other systems (mainly cardiac, skeletal and genitourinary).

Investigations:

- Detailed ultrasound examination, including neurosonography.
- Invasive testing for karyotyping and array.
- TORCH test for fetal infections.
- Fetal brain MRI at ≥32 weeks for diagnosis of abnormalities that are not detectable by ultrasound, such as grey matter heterotopias, late sulcation and migration anomalies.

Follow up:

- Follow-up should be standard.

Delivery:

- Standard obstetric care and delivery.

Prognosis:

- Isolated: neurodevelopmental delay in 30% of cases.
- Other defects: prognosis could be poor depending on the type of defect.

Outcome

- Outcome of individuals with disorders of the corpus callosum is dependent on the presence or absence of associated anomalies and genetic syndromes. Identification of a cause and/or associated abnormalities can result in more appropriate counseling to the pregnant patient regarding the long-term outcome of the child. When ACC is confirmed as isolated, two-thirds of children appear to have a normal outcome.


Thank
you