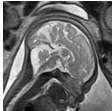
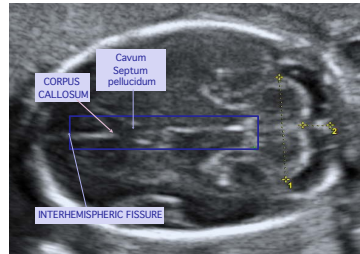


MIDLINE ANOMALIES



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Department of Obstetrics and Gynecology
Texas Children's Hospital

MIDLINE STRUCTURES



MIDLINE STRUCTURES

- Anomalies in the development of the septum pellucid
- Anomalies in the development of corpus callosum

MIDLINE STRUCTURES

- Anomalies in the development of the septum pellucid
- Anomalies in the development of corpus callosum

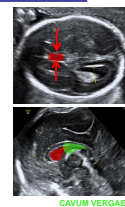
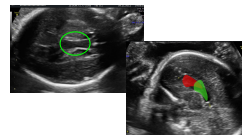
MIDLINE STRUCTURES

Cavum septum pellucidum

- Always visible
- 18 to 37w
- Average width: $5.3 \pm 1.7\text{mm}$
- Increases w GA

INCREASE

ABSENCE

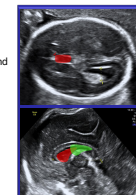
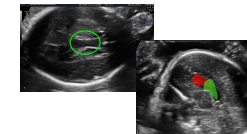


CAVUM VERGAE

MIDLINE STRUCTURES

Prominent Cavum vergae

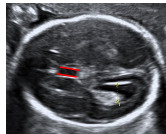
- Variant of normality
- Check for its correspondences on sagittal planes and check for the integrity of CC



MIDLINE STRUCTURES

Septum pellucidum

- borders of CSP
- WM and GM
- Its absence communicates anterior horns



ABSENCE ?

Pattio et al. UOGS, 2000

AGENESIS OF THE SEPTUM PELLUCIDUM

Anterior communication



Association to facial anomalies
ACC, alteration in the division of the cortex, BBGG

MRI: Alteration in the chiasma

Absent

Lobar
Alobar
semilobar
Holoprosencephaly

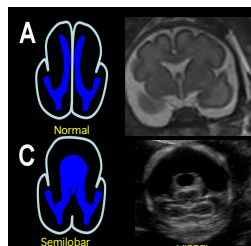
SOD

Isolated
agenesis of
the SP

AGENESIS OF THE SEPTUM PELLUCIDUM

Holoprosencephaly

Plu UOG 2003



Associated to midline anomalies
Hypotelorism-cyclopia
Arhinia, cecocephaly
cleft lip-palate

Associated to chromosomopathies
(trisomy 13)

Prognosis:
Semilobar and Alobar: MRI
neurological sequelae 100%
Lobar: Suboptimal cognitive
outcome in most cases

AGENESIS OF THE SEPTUM PELLUCIDUM

Anterior communication



Association to facial anomalies
ACC, alteration in the division of the cortex, BBGG

MRI: Alteration in the chiasma

Absent

Lobar
Alobar
semilobar
Holoprosencephaly

SOD

Isolated
agenesis of
the SP

Separation of the anterior process of the LV

Lack of division of the cortex and BBGG

MH Holoprosencephaly

Normal division

Septal Remnant

Present

Hydrocephalus

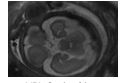
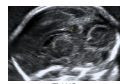
Absent

Cortical malformation
Schizencephaly

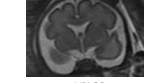
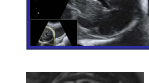


AGENESIS OF THE SEPTUM PELLUCIDUM

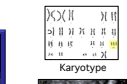
Complementary studies



MRI: Optic chiasm



MRI: CC



MRI: Cortical study

AGENESIS OF THE SEPTUM PELLUCIDUM

PROGNOSIS

85% ASSOCIATED LESIONS

- Holoprosencephaly
- Neuronal migration abnormalities
- Pituitary alterations
- ACC

15% isolated

Bad prognosis

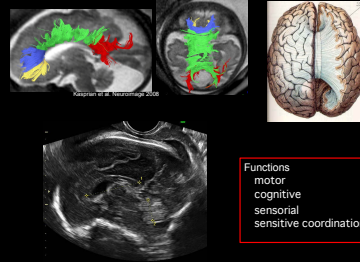
50-80% asymptomatic
Motor, cognitive, behavioral, psychological,
endocrinological alterations

Damei et al. 2010; Bellone et al. 2005; Li et al. 2011

MIDLINE STRUCTURES

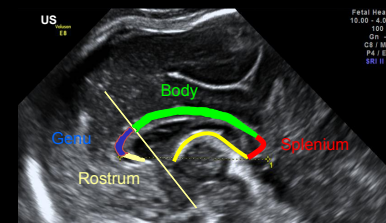
- Anomalies in the development of the septum pellucidum
- Anomalies in the development of corpus callosum

CORPUS CALLOSUM

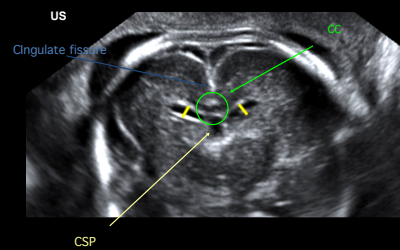


Functions
motor
cognitive
sensorial
sensitive coordination

CORPUS CALLOSUM

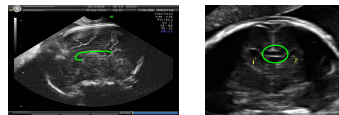


CORPUS CALLOSUM



AGENESIS OF THE CORPUS CALLOSUM

DIAGNOSIS



INDIRECT SIGNS

DIRECT SIGNS

AGENESIS OF THE CORPUS CALLOSUM

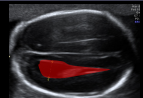
INDIRECT SIGNS



Colpocephaly

AGENESIS OF THE CORPUS CALLOSUM

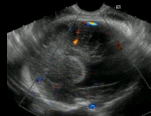
INDIRECT SIGNS



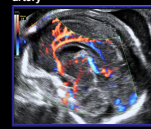
Colpocephaly



Separation of the anterior horns



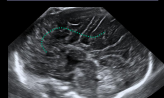
Absence of pericallosal artery



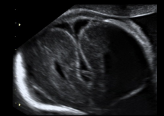
Normality

AGENESIS OF THE CORPUS CALLOSUM

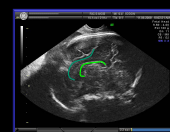
DIRECT SIGNS



Absence of CC
Vertical sulci
Absence of cingulate fissure



Normality



Pho 1993, D' Souza 1998, Barlow 2000

AGENESIS OF THE CORPUS CALLOSUM

Etiology

- Unknown 68%
- Unfrequent: External factors: Maternal alcohol, PKU
- Genetic factors: Trisomies, 39% chromosomal rearrangements
- Multiple malformations

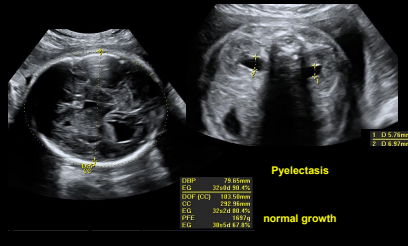
- A. Dominant
- Mowat-Wilson
 - Miller-Dieker
- A. Recessive
- Acrocallosal
 - Andermann
 - Joubert
 - Meckel-Gruber
 - Shapiro
- Chromosome X associated
- Aicardi

Karyotype

Echocardiogram
NSG
Detailed anatomy study

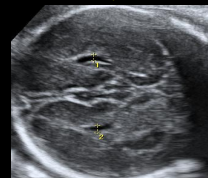
Barlow 2001, Gilhall-Aparisi 2010

32 y.o. G1
1st trimester screening: increased CVS: Normal karyotype
30weeks. Breech. NSG for VMG.

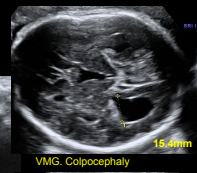


30w. Breech.

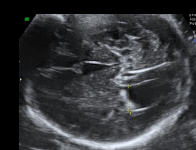
Anterior horn separation



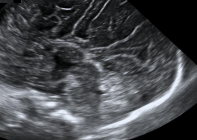
Normal PF

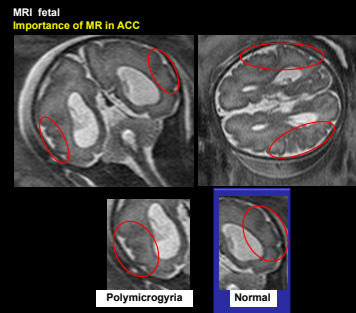
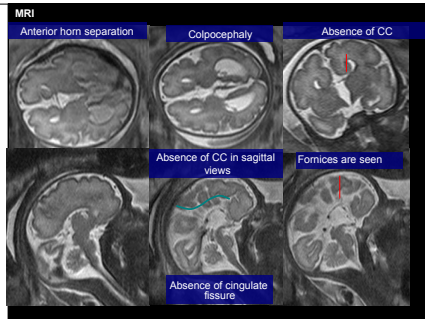


33.1w. Vertex



Atria 13 y 14mm
3rd ventricle dilated





AGENESIS OF THE CORPUS CALLOSUM

Complementary studies : Fetal MRI

- Cortical dysplasia associated
 - Polymicrogyria 29%
 - Heterotopia 14%
 - Pachgyria / lissencephaly 25%
- Cortical maturation delay
- 50% associated with intracranial abnormalities

Fetal MRI

Deholl-Aspach et al 2010; Warren et al 2010; Coakley et al 2008

Thank you for your attention

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