The Cavum Septi Pellucidi in Utero

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The Cavum Septi Pellucidi

• What is it?
  ■ Embryology
  ■ Anatomy
  ■ Etymology
• Why Does it Matter?
  ■ Four reasons illustrated by a variety of in utero images
  ■ Imaging Pitfalls

The CSP: What is it?

“You see what you look for, and you look for what you know”

Apocryphal, Sir Arthur Conan Doyle

The Cavum Septi Pellucidi

• The CSP is conceptually regarded as a part of the longitudinal cerebral fissure which becomes walled off by the union of the hemispheres forming the corpus callosum above and the fornix below

The CSP: What is it?

Etymology

• Cavum: Latin- cavus, a hollow space, hole, or cavity
  ■ Septum: Latin- septus = wall or enclosure
  ■ Septum = neuter singular
  ■ Septa = neuter plural
  ■ Septi = genitive singular
  ■ Shows possession, like the apostrophe- s
  ■ “of the”
• Pellucidum: Latin- translucent or clear
  ■ Thus, cavum septi pellucidi translates as “the cave (hollow) of the clear walls (septa)"

Winter et al, AJR Dec 2011
The Cavum Septi Pellucidi

- AKA septum lucidum or "fifth ventricle"
- At ~12 weeks gestational age the corpus callosum starts to develop from the lamina terminalis as a bundle of fibers that connects the two hemispheres
- This is associated with the development of the septa pellucida, two paired clear membranes

The space between the two septa pellucida is one cavity, but has two different names:

- Anterior = cavum septi pellucidi
- Posterior = cavum vergae

Dividing Point (equivalent)

- Foramina of Monro
- Vertical plane defined by the columns of the fornix

Vergae: Verga’s ventricle. The cavity was first described by the Italian anatomist, Andrea Verga, in 1851. AKA the "sixth ventricle".

In most individuals the cavum is closed early in childhood and the two septa are fused.

- The fused membranes, now using the Latin singular, are properly referred to as the septum pellucidum
- This closure of the CSP et vergae begins at approximately 6 months gestational age, and progresses from back to front.
- By term closure has occurred posteriorly in 97%, so that there is generally only a true CSP at birth.
- By 3-6 months of age, the CSP is closed in 85% of infants, although in a minority this cavity remains open until adulthood.

Although correctly the entire space is the "CSP et vergae", in common usage the entire cavity is often just called the CSP.

In Utero

Normal CSP

Axial  Coronal

In Utero

Cavum septi pellucidi

Normal

Axial  Coronal
Normal

Axial  In Utero  Coronal

Normal

Axial  Postnatal  Coronal

Normal (29 week premature)

Axial  Postnatal  Coronal

Normal (term)

Axial  Postnatal  Coronal

Normal Variant CSP et Vergae

In Utero  Axial

Is this the CSP???

In Utero  Axial
Columns of the Fornix: Don’t Mistake for the Cavum Septi Pellucidi!

Callen et al, JUM 2008; 27:25–31

Columns of the Fornix: Don’t Mistake for the Cavum Septi Pellucidi!

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Etymology: Fornix

- **Fornix**
  - Vaultlike or arched structure
  - Archlike anatomical structure or fold
  - From Latin: arch, vault, vaulted chamber
- Derivation of “fornication”
  - In ancient Rome, prostitutes frequented the vaulted arches surrounding the Colosseum
  - The act of carrying on an illicit sexual relationship consequently came to be called "going under the arches" or fornication

Columns of the Fornix: Two More Examples

Callen et al, JUM 2008; 27:25–31

Columns of the Fornix: Don’t Mistake for the Cavum Septi Pellucidi!

Callen et al, JUM 2008; 27:25–31

Normal

In Utero

Axial
The CSP: Why Does it Matter?

• Required by "Guidelines" (AIUM, ACR, ACOG, ...)

The Cavum Septi Pellucidi

Head and neck
1. Cerebellum
2. Choroid plexus
3. Cisterna magna
4. Lateral ventricles
5. Midline falx
6. Cavum septi pellucidi

Fetus #1

In Utero
Axial

Fetuses #2 and #3

What's Your Diagnosis?
Agenesis of the Corpus Callosum

- One of the most common CNS anomalies (0.5 – 70 per 10,000 live births, and 4% of CNS malformations)
- Prevalence varies in different studies, ranging between 0.3 and 0.7% in the general population and between 2 and 3% in the developmentally disabled population
- Associated with major cerebral and/or extracerebral malformations, including 50 different human congenital syndromes and metabolic diseases
- Up to 80% risk of associated brain anomalies, such as Dandy–Walker complex, gyral anomalies, and heterotopia
- Up to 60% risk of associated extra-CNS abnormalities, including CHD, MSK, and GU

Prenatal Diagnosis of ACC

- Suspect when the CSP is not visualized on the axial transthalamic view
- Additional indirect signs
  - Colpocephaly
  - Increased separation of the hemispheres with the bodies of the lateral ventricles parallel to each other and shifted laterally
  - Abnormal third ventricle, which extends upward between the lateral ventricles in 1/2 of cases
  - Abnormal course of the pericallosal artery


Agenesis of the Corpus Callosum

- Axial
- In Utero
- Axial

Agenesis of the Corpus Callosum

- Coronal
- In Utero
- Sagittal

Hypogenesis of the Corpus Callosum

- Axial
- In Utero
- Coronal

Hypogenesis of the Corpus Callosum

- In Utero
- Sagittal
Pitfall: High Riding Third Ventricle

ACC & DWM

Fetus #1
What's Your Diagnosis?

Aicardi Syndrome

- Initial clinical description was the triad of infantile spasm, ACC and ocular abnormalities
- In a female fetus, the combination of ACC with posterior fossa malformation (posterior fossa cysts, cerebellar hypoplasia) and cortical dysplasia suggests Aicardi syndrome, especially if there are eye abnormalities (e.g. microphthamia, coloboma)
- X-linked dominant disorder seen in females, with lethal consequences for the male fetus
- The finding of retinal lacunae on postnatal ophthalmologic exam confirms the diagnosis

Aicardi Syndrome

- Axial
- In Utero
- Axial

Aicardi Syndrome

- Axial
- In Utero
- Axial

Aicardi Syndrome

- Axial
- In Utero
- Axial

Aicardi Syndrome
The CSP: Why Does it Matter?

- Required by "Guidelines" (AIUM, ACR, ACOG, ...)

The Common and Difficult to Diagnose: Agenesis of the Corpus Callosum (including Aicardi)

Fetus 1

Axial

In Utero

Coronal

What's Your Diagnosis?

Septo-optic Dysplasia

- Absence of the CSP, hypoplasia of the optic nerves and chiasm, various types of hypothalamic-pituitary dysfunction leading to growth delay, and associations with developmental delay/intellectual impairment
- Distinguish from HPE by the absence of fused thalami; typical lack of pronounced ventriculomegaly; and normal appearance to the CC, columns of the fornix, anterior cerebral arteries and falx cerebri
- Recognition of absence of CSP is essential to suggesting this diagnosis prenatally

Fetus 1

Axial

Postnatal

Axial

Septo-optic Dysplasia

Axial

In Utero

Coronal
The CSP: Why Does it Matter?

- Required by “Guidelines” (AIUM, ACR, ACOG, ...)
- The Common and Difficult to Diagnose:
  - Agenesis of the Corpus Callosum (including Aicardi)
- The Uncommon and Very Difficult to Diagnose:
  - Septo-optic Dysplasia
- An excuse to review a whole host of midline abnormalities

Basic Embryology for Sonographers and Sonologists

- Ventral induction takes place during weeks 5 to 10 of neurulation
- Three primary vesicles and four secondary vesicles
  - Forebrain = prosencephalon
    - Telencephalon: cerebrum
    - Diencephalon: thalamus and hypothalamus
  - Midbrain = mesencephalon
    - Tectum and cerebral peduncles
  - Hindbrain = rhombencephalon
    - Metencephalon = cerebellum and pons
    - Myelencephalon = medulla oblongata
Holoprosencephaly


Alobar Holoprosencephaly

Axial

In Utero

Coronal

Semi-lobar Holoprosencephaly

Axial

In Utero

Axial

Lobar Holoprosencephaly

Axial

In Utero

Coronal

Lobar Holoprosencephaly

Postnatal

Axial
**Syntelencephaly**

- Neuronal migrational anomaly rather than vascular insult
- Two types have been described, both of which communicate with the ventricles: open lipped and close lipped
- Difficult to diagnose with U/S in the 2nd tri, so nonvisualization of the CSP may be a useful clue
- Absent CSP is noted in 70% of schizencephaly, especially when bilateral
- Associations include enlarged ventricles, ACC, polymicrogyria, and heterotopia

**Schizencephaly**

- Replacement of most of the cerebral hemispheres with fluid with sparing of the rhombencephalic structures
- Proposed etiologies include overwhelming antenatal infections and occlusion of the internal carotid arteries
- Obviously, non-visualization of the CSP is an expected finding; recognition of absence of the CSP is not crucial to making the diagnosis

**Hydranencephaly**
Basilar Encephalocele

- In Barkovich’s paper on absence of the septum pellucidum, two of three patients with basilar encephaloceles also had ACC and absence of the CSP

Chronic Severe Hydrocephalus

- Etiologies of severe hydrocephalus associated with nonvisualization of the CSP include aqueductal stenosis and the Chiari II malformation
- Severe hydrocephalus may be difficult to distinguish from hydranencephaly and alobar holoprosencephaly
- Severe hydrocephalus of any type with fenestration of the CSP is suggested as one of the more common causes of CSP nonvisualization on prenatal sonograms

The CSP: Why Does it Matter?

- Required by "Guidelines" (AIUM, ACR, ACOG, ...)
- The Common and Difficult to Diagnose: Agenesis of the Corpus Callosum (including Aicardi)
- The Uncommon and Very Difficult to Diagnose: Septo-optic Dysplasia
- An excuse to review a whole host of midline abnormalities:
  - Holoprosencephaly
  - Schizencephaly
  - Hydranencephaly
  - Basilar Encephalocele
  - Chronic Severe Hydrocephalus
Pitfalls

1. Incorrectly attributing the paired columns of the fornix as the CSP
2. Confusing the high riding third ventricle or a prominent interhemispheric fissure for the CSP
3. Normal variant CSP et vergae
4. Noting fluid between the lateral margins of the frontal horns and calling that the normal CSP

Pitfall #1
- Incorrectly attributing the paired columns of the fornix as the CSP

Pitfall #2
- Do not confuse the high riding third ventricle or a prominent interhemispheric fissure for the CSP

Pitfall #3
- Normal variant CSP et vergae

Pitfall #4
- Simply noting fluid between the lateral margins of the frontal horns and calling that the normal CSP
- To quote one of the gurus, “don’t make it up!”

Septo-optic Dysplasia

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Thank you!

- Professor Margaret Toscano for help with Latin (all mistakes mine!)
- NASA Astronomy Picture of the Day (http://apod.nasa.gov/apod/)