The Cavum Septi Pellucidi
Why Is It Important?

Thomas C. Winter, MD, Anne M. Kennedy, MBBCh,
Jan Byrne, MD, Paula J. Woodward, MD

Objective. The cavum septi pellucidi (CSP) is routinely imaged in the fetal brain during obstetric sonography; in fact, for well over a decade, assessment of the CSP has been considered part of the required elements of a standard examination of fetal morphology in guidelines developed by multiple specialty societies. Our objective is to present the 4 reasons why all practicing sonologists and sonographers should be familiar with this anatomic structure. Methods. Prenatal sonograms and magnetic resonance imaging examinations are used to review the following topics: terminology, embryology, and anatomy of the CSP; pitfalls in its identification; and a wide variety of abnormalities (predominantly relating to nonvisualization) associated with the CSP. Results. Embryologic development of the CSP is intimately associated with the corpus callosum (CC); thus, correct identification of the CSP essentially excludes complete agenesis of the CC. Absence of the CSP is associated with an extremely wide spectrum of neuroanatomic malformations: these range from the lethal entities of hydranencephaly and alobar holoprosencephaly; to the potentially serious but nonlethal entities of schizencephaly, porencephaly, basilar encephalocele, severe hydrocephalus, and the less severe prosencephalic cleavage disorders (including syntelencephaly); to the normal variant, the rare and somewhat controversial entity of isolated septal deficiency. The value of noting that the absent CSP allows diagnosis of very subtle and easily overlooked abnormalities such as septo-optic dysplasia is presented. Conclusions. Correct recognition of the CSP provides welcome reassurance of proper development of the central forebrain. Key words: cavum septi pellucidi; corpus callosum; prenatal diagnosis; prosencephalon; sonography.
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stenosis or the Chiari II malformation; schizencephaly; porencephaly/hydranencephaly; basilar encephalocoeles; and isolated septal deficiency.9–11

This manuscript will review prenatal sonography and magnetic resonance imaging (MRI) of the CSP, including anatomy, embryology, and scan techniques. The differential diagnosis for an absent CSP and consequences for management are discussed.

Anatomy and Embryology

Anatomy and scan techniques are well described in standard texts12,13; a superb, more detailed description of the anatomy of the CSP and corpus callosum (CC) may be found in a 2008 article by Callen et al.9

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 CC above and the fornix below.9,14

More specifically, at about 12 weeks’ gestational age, the CC 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 septa is one cavity but has two different names; anterior to the foramina of Monro, it is called the CSP, whereas posterior to this structure, it is called the cavum vergae (CV).9 Other authors use different terminology but the equivalent position anatomically: a vertical plane defined by the columns of the fornix (from Latin, a vaultlike or arched structure) as the division between the CSP and the CV.9,15 Of note, although correctly the entire space is the “CSP et vergae,”9 in common usage (and unless specified otherwise throughout this article), the entire cavity is often just called the CSP. In most individuals, the cavum (literally, a hollow space, hole, or cavity) 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.1,17 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 to 6 months of age, the CSP is closed in 85% of infants,18 although in a minority this cavity remains open until adulthood.19

Although there is no direct evidence to suggest that the CSP cannot develop independently from the CC, the CC is closely related anatomically and embryologically to the CSP; most authors state that there cannot be a CSP without a CC.13 Both the CC and the CSP develop from the commissural plate.9 Conversely, it is possible to have a normal CC without a normal CSP, eg, in isolated septal deficiency or SOD.

The CC forms late in cerebral ontogenesis, between 12 and 18 weeks13 or alternatively 8 and 20 weeks19 (depending on the citation). Most authorities state that the CC forms from front to back (ventrally to dorsally), beginning with the rostrum, then the genu, body, and splenium.13,17 although fetal MRI and other data suggest that the genu and anterior body form first, with development then proceeding both anteriorly to the rostrum and posteriorly to the splenium.19,20

Sonography

The CSP is routinely imaged on the 3 obligatory views of the fetal head obtained during obstetric sonography. Specifically, in addition to the images of the ventricles and the posterior fossa, an axial view at the level of the paired thalami yields both the biparietal diameter and the CSP.13,21 Under normal conditions, the CSP should be easily seen beyond approximately 18 to 20 weeks.13,17,22,23 A variety of sonographic and MR images of the normal CSP, both prenatal and postnatal, are provided (Figures 1–3).

Proper attention to technique is crucial in not mistakenly calling the CSP present when it is truly absent.24 The CSP must range from the medial wall of the frontal horn of one lateral ventricle to the medial wall of the frontal horn of the contralateral lateral ventricle; if it does not, one runs the risk of ignoring true absence of the CSP and missing associated neurologic abnormalities.25 Pilu et al24 have described an artifact mimicking the presence of the CSP in standard axial second-trimester sonographic images of the brain; thus, if the CSP is not seen with certainty on standard axial images, obtain a series of coronal images, and also attempt to identify the CC directly with a midsagittal view.9
Part of the problem in mistakenly calling a CSP present when it is indeed absent relates to confusing normal anatomy, the paired nerve columns of the fornix, with the CSP (Figure 4). Because the embryologic development of the fornix is not closely correlated with that of the CC, demonstration of the fornix will not exclude abnormalities of the CC and other central forebrain malformations.9 Recall that the CSP can be regarded as part of the longitudinal cerebral fissure, which is walled off by the CC above and the fornix below during embryologic development.

Figure 1. Normal CSP in utero by second-trimester sonography. The CSP appears as a fluid-filled box between the paired septa (arrows), which separate the CSP from the frontal horns of the lateral ventricles. A, Axial. Note the absence of the central echogenic line in the center of the CSP, a finding that helps prevent misinterpretation of the fornix as the CSP (see Figure 4). Anterior is to the reader's left. B, Coronal. The loss of detail on the viewer's right is due to near-field reverberation.

Figure 2. Normal CSP postnatally by MRI. Axial (A) and coronal (B) images of the normal CSP in a term neonate show the normal appearance of the CSP (arrows).
Thus, paired adjacent hypoechoic structures in the forebrain in a plane of section just caudal to the axial plane that delineates the CSP represent the fornix; if the CSP is absent, the fornix may be misinterpreted as the CSP. The normal CSP should appear as a white line (lateral margin of the CSP), dark box (fluid within the CSP), and white line (lateral margin of the opposite side of the CSP), whereas the fornix will appear as a white line (lateral margin of one of the fornices), dark area (substance of one of the fornices), white line (interface between the paired fornices), dark area (substance of the other fornix), and white line (lateral margin of the other fornix).

A squared appearance to the frontal horns on coronal views should suggest the possibility of absence of the CSP.

What is the normal size of the CSP? Jou et al studied more than 600 consecutive fetuses between 19 and 42 weeks of age and found that at 19 to 20 weeks, the mean width was 3.4 mm, and 2 SD on either side ranged from 2.08 to 4.72 mm. A smaller study also noted a difference in the size of the CSP between the second and third trimesters, with the mean width in the second trimester of 3.1 ± 1 SD of 1.5 mm.

Magnetic Resonance Imaging

Techniques for fetal MRI have been well described elsewhere. Note that fetal MRI is generally performed at a later gestational age than routine fetal survey sonography due to both technical reasons (fetal movement and size) and the fact that some brain abnormalities may not be detectable until later in pregnancy. Fetal MRI adds useful information in many patients with sonographically detected neurologic abnormalities. For example, in agenesis of the corpus callosum (ACC), prenatal MRI finds other abnormalities that were missed on sonography in more than half of cases. Many of these, such as gray matter heterotopia, are too subtle to be readily detected by sonography. In addition to its intrinsic value in the fetal brain, MRI often adds particular value in patients in whom sonography is difficult, eg, maternal obesity and oligohydramnios.
Pathology

Absence of the CSP occurs as part of a large variety of cerebral anomalies. In some instances, complex malformations are readily identified with prenatal sonography, such as the spectrum of HPE and severe ventriculomegaly. Other anomalies, particularly ACC, may be more subtle. Many of these associations are discussed below:

Figure 5. Classic complete ACC. A, Axial third-trimester sonogram, with anterior to the reader’s left, shows colpocephaly (asterisk). B, Axial in utero third-trimester MRI shows parallel lateral ventricles. C, Coronal MRI shows a “Texas longhorn” configuration to the lateral ventricles, with absence of the CSP. D, Sagittal MRI shows complete absence of the CC. Note nonvisualization of the CSP on all images.

Agenesis of the Corpus Callosum

Agenesis of the corpus callosum (Figures 5 and 6) is part of the spectrum of CC abnormalities, including hypogenesis and dysgenesis. It is one of the most common central nervous system (CNS) anomalies (0.5–70 per 10,000 live births and 4% of CNS malformations\(^\text{30}\)). Manifestations are protean, from asymptomatic individuals to associated abnormalities, including hydro-
cephalus, lissencephaly, pachygyria, and microcephaly. More than 80 associated chromosomal (including trisomy 18 and 13), genetic, and sporadic syndromes have been described. The Dandy-Walker malformation is one of the most common associations; callosal hypogenesis is present in up to one-third of patients with the Dandy-Walker malformation (Figure 7).

Because the CC largely forms from anterior to posterior, hypogenesis of the CC implies that the splenium is always abnormal. Prenatal imaging may show the classic finding of colpocephaly (focal dilatation of the trigones and occipital horns of the lateral ventricles) with the so-called teardrop-shaped ventricles. The fibers that should have crossed to the opposite hemisphere turn at the interhemispheric fissure and run parallel to it, forming the bundles of Probst. In turn the Probst bundles displace the frontal horns of the lateral ventricles, giving them a crescentic shape, producing the steer horn or Viking helmet sign in the coronal plane. Other potential sonographic findings include a high-riding third ventricle between widely separated, parallel lateral ventricles; several subtypes of interhemispheric cysts, not all of which communicate with the ventricles; lipoma in the expected location of the CC; vertical orientation of the radiating gyri (often seen best with a sagittal endovaginal approach in the vertex fetus); and an abnormal course or absence of the pericallosal artery. Despite all of these findings, diagnosis of isolated ACC may be challenging, even for expert sonologists, before 22 weeks. In particular, if the classic colpocephaly is not appreciated, recognition of the absence of the CSP prompts the sonologist to suggest the diagnosis of ACC.

Of note, infection, hemorrhage, and other injurious processes may lead to destruction of portions of the CC after normal initial formation. In this situation, the posterior portion of the callosum may be absent, but the CSP may still be present, so visualization of the CSP does not always imply a completely normal CC.

**Aicardi Syndrome**

The initial clinical description was a triad of infantile spasms, ACC, and ocular abnormalities. In a female fetus, the combination of ACC with posterior fossa malformation (posterior fossa cysts and cerebellar hypoplasia) and cortical dysplasia suggests Aicardi syndrome, especially if there are eye abnormalities (eg, microphthalmia and coloboma; Figure 8). Aicardi syndrome is an
Figure 7. (continued) B, Slightly oblique axial sonogram at the level of the lateral ventricles shows markedly dilated ventricles with colpocephaly (F indicates frontal horn; and O, occipital horn) as well as the superior aspect of the expanded fourth ventricle (DW). C, Another suboccipital bregmatic sonogram also shows the relative sparing of the mildly dilated frontal horns of the lateral ventricles of colpocephaly as well as the expanded fourth ventricle (arrow). D, Coronal in utero third-trimester MRI shows a Texas longhorn configuration to the lateral ventricles, with absence of the CSP. E, Axial in utero MRI shows parallel lateral ventricles (left) and a large fourth ventricle in the posterior fossa (right). F, Sagittal MRI shows complete absence of the CC and a large fourth ventricle in the posterior fossa. Note nonvisualization of the CSP on all images.
X-linked dominant disorder seen in females with lethal consequences for male fetuses and is rarely seen in 47,XXY karyotypes.\textsuperscript{11,31} The finding of retinal lacunae on postnatal ophthalmologic examination confirms the diagnosis.

\textbf{Holoprosencephaly}

Holoprosencephaly is a severe malformation of the brain characterized by abnormal cleavage of the prosencephalon in the fifth gestational week. Most cases are sporadic, but chromosomal, genetic, and teratogenic causes have been discussed.\textsuperscript{31} This entity has classically been divided into 3 subtypes by DeMeyer et al: alobar, semilobar, and lobar. To quote from the University of California San Francisco group: “Alobar HPE, the most severe of the three types, generally appears as a pancake-like mass of cerebral tissue and a crescentic monoventricle. Semilobar HPE is characterized by noncleaved anterior brain, with

\textbf{Figure 8.} Aicardi syndrome. A, Suboccipital bregmatic second-trimester sonogram, with anterior to the reader’s left, shows absence of the CSP with interdigitating gyri in the anterior brain but a normal appearance to the cerebellum and posterior fossa. B, Axial sonogram shows a posterior interhemispheric cyst (asterisk). C, In utero third-trimester axial MRI shows pachygyria and polymicrogyria. D, Sagittal MRI delineates a coloboma (arrow). A coloboma is a defect in which part of the eye, in this case the optic nerve, does not form correctly. The presence of a coloboma increases the likelihood of a syndromic diagnosis.\textsuperscript{33}
the posterior falx and posterior interhemispheric fissure (IHF) present. Lobar HPE is also characterized by anterior brain noncleavage that exists to a lesser degree than that of semilobar HPE, a fully formed third ventricle, and some development of frontal horns of the lateral ventricles. The alobar variety has been further subdivided by sonologists into 3 types based on the residual cerebral tissue that appears in these forms: "pancake," "cup," and "ball." As Barkovich states, though, the HPEs are a heterogeneous group of forebrain malformations, with no clear distinction between the different categories having been firmly established; other classification systems have also been suggested.

Sonographic findings in HPE depend on the severity of the forebrain cleavage malformation. In the alobar type (the easiest to recognize; absence of the CSP is overshadowed by the other striking sonographic findings; Figure 9), the falx and the interhemispheric fissure are totally absent; the thalami are fused; there is a single monoventricle, often described with a "horseshoe" configuration; and the third ventricle, neurohypophysis, olfactory bulbs and tracts, CSP, CC, and interhemispheric fissure are absent. In semilobar HPE (Figure 10), the monoventricle is still present, but there is some separation of the cerebral hemispheres posteriorly. The thalamus may be partially or fully fused but again with absence of the third ventricle. A characteristic dorsal sac is often associated with the alobar and semilobar forms. These first 2 types of HPE are generally easily recognized, but the correct findings in lobar HPE may be difficult to observe; again, absence of the CSP is particularly important in recognizing the subtle variants of lobar HPE. With lobar HPE (Figure 11), the brain is almost completely divided into two hemispheres, with the exception being a variable degree of fusion at the level of the cingulate gyrus and frontal horns of the lateral ventricles. Other findings helpful in lobar HPE are central fusion of the frontal horns with a flat, squared roof and communication with the upper portion of the third ventricle on coronal imaging and fused fornice appearing as a linear structure running within the third ventricle from the anterior to posterior commissure. Some patients with lobar HPE likely fall into the spectrum of SOD.

All types of HPE are often associated with microcephaly and less frequently with macrocephaly due to aqueductal stenosis leading to hydrocephalus. A wide variety of midline facial abnormalities are often associated with HPE: hypotelorism or cyclopia, median cleft lip and palate, a flat nose with a single naris (cebocephaly), and a proboscis superior to the level of the eyes (ethmocephaly). In 1993, Barkovich described a lesser known fourth subtype of HPE, related to semilobar HPE, known as the midline interhemispheric fusion variant or syntelencephaly (Figure 12). Syntelencephaly consists of an abnormal midline connection of the cerebral hemispheres in the posterior frontal and parietal regions, with interhemispheric separation of the basal forebrain, anterior frontal lobes, and occipital regions; prenatal MRI of this entity has been reported.

Septo-optic Dysplasia

Septo-optic dysplasia (also known as De Morsier syndrome; Figure 13) is a rare congenital disorder characterized by absence of the CSP, hypoplasia of the optic nerves and chiasm leading to alterations in visual acuity, various types of hypothalamic-pituitary dysfunction (including alterations in growth hormone, antidiuretic hormone, thyroid-stimulating hormone, luteinizing
hormone, and follicle-stimulating hormone levels) leading to growth delay, and associations with developmental delay/intellectual impairment.\textsuperscript{23,24,38} It has been reported in association with valproic acid embryopathy.\textsuperscript{39} Septo-optic dysplasia can be distinguished from HPE by the absence of fused thalami; typical lack of pronounced ventriculomegaly (although dilated ventricles may be present occasionally in SOD); and a normal appearance to the CC, columns of the fornix, anterior cerebral arteries, and falx cerebri.\textsuperscript{23} An excellent discussion of SOD, including the diagnostic utility of low levels of gravid maternal estriol, is provided by Lepinard et al\textsuperscript{23}; potential associations of SOD with cleft lip and palate, as well as cortical dysplasia, are discussed. Note that cortical dysplasias and some forebrain malformations may not be detected by sonography or MRI before 30 to 32 weeks. As the resolution of 2- and 3-dimensional sonography\textsuperscript{40} and MRI continues to improve, an effort may be made to evaluate the optic chiasm and nerves for

\textbf{Figure 10.} Semilobar HPE. \textbf{A,} Axial second-trimester sonogram, with anterior to the reader’s left, shows absence of the CSP. Note how otherwise normal this image appears. \textbf{B,} Coronal sonogram shows a fused choroid plexus (arrows) across the midline. \textbf{C} and \textbf{D,} Axial sonogram (\textbf{C}) and third-trimester MRI (\textbf{D}) denote a fused frontal lobe cortex across the midline (asterisks) as well as absence of the CSP.
Figure 11. Lobar HPE. A, Axial second-trimester sonogram, with anterior to the reader’s left, shows a round head, absence of the CSP, and ventriculomegaly. B, Coronal sonogram shows a classic fused fornix (arrow), a frequent and reportedly specific sonographic finding for lobar HPE. C, Coronal postnatal (neonatal) sonogram of the head again denotes a classic fused fornix (arrow). D, Axial postnatal MRI better delineates findings from A. E, Coronal postnatal MRI replicates findings from B and C (arrow indicates fused fornix).
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Additional information in cases of possible SOD. Counseling is difficult because SOD has such a wide range of clinical presentations; it is impossible to determine disease severity prenatally and the diagnosis must be confirmed clinically after birth. Recognition of absence of the CSP is essential to suggesting this diagnosis prenatally.

**Schizencephaly**

The cleft of schizencephaly (Figure 14) is of uncertain etiology; in the past, it has been proposed that schizencephaly is part of a spectrum of encephaloclastic disorders due to vascular infarction, although more recent work suggests a neuronal migrational anomaly as perhaps a...
more likely etiology. Two types have been described, both of which communicate with the ventricles, ie, extend from the pia to ependyma. In the open-lipped form, the edges of the cleft are splayed apart, whereas in the close-lipped form, the edges are apposed. Associations include enlarged ventricles, ACC, polymicrogyria, heterotopia, and absence of the CSP; absence of the CSP is noted in 70% of schizencephaly cases, especially when bilateral. Schizencephaly can be difficult to diagnose with sonography in the second trimester, so nonvisualization of the CSP may be a useful clue. Other terms that have been used for schizencephaly include “true porencephaly” and “agenetic porencephaly.”

**Figure 14. Schizencephaly.** A, Axial second-trimester sonogram, with anterior to the reader’s left, denotes absence of the CSP (arrow; again, note nonvisualization of the normal box between the frontal horns of the lateral ventricles). An abnormal cleft in the cortex (S) is seen rostral to the occipital horn (V). B, In utero third-trimester MRI confirms the extent of defect from the pia to ependyma (asterisk) and shows the gray matter lining.

**Porencephaly**
Also known as pseudoporencephaly or encephaloclastic porencephaly, this term is often used as collective nomenclature for a variety of large cortical defects that may communicate with the ventricles. Unlike schizencephaly, which is lined with gray matter, the lining of the porencephalic cavity often contains white matter. The cavity is almost never midline.

**Hydranencephaly**
Sonographic findings include replacement of most of the cerebral hemispheres with fluid with sparing of the brain stem and rhombencephalic structures. Proposed etiologies include overwhelming antenatal infections and occlusion of the internal carotid arteries. Obviously, nonvisualization of the CSP is an expected finding, and recognition of absence of the CSP is not crucial to making the diagnosis of this entity (Figure 15).

**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 HPE. The presence of some cortical tissue (cup, ball, or pancake form) should be seen with alobar HPE; a very thin rind of cortex indicates extreme hydrocephalus rather than hydranencephaly. Magnetic resonance imaging is helpful to confirm the presence of a cortical mantle. From a practical perspective, hydranencephaly and alobar HPE have a similar dismal prognosis, the differentiation being of concern primarily in prediction of recurrence risk. 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 (Figure 16).
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**Basilar Encephaloceles**

In an article by Barkovich and Norman on absence of the septum pellucidum, 2 of 3 patients with basilar encephaloceles (Figure 17) also had ACC and absence of the CSP.

**Isolated Septal Deficiency**

Several cases of isolated absence of the CSP have been reported. However, counseling is difficult because apparently isolated absence of the CSP may be associated with cytoarchitectural disturbances of cortical layers not currently demonstrable by MRI.

**Enlarged CSP**

After birth, an enlarged CSP (>1 cm) and persistence of a CSP beyond infancy have been described as “true subtle markers of cerebral dysgenesis,” possibly associated with neuropsychiatric disturbances (particularly schizophrenia). It is not known with certainty what percentage of fetuses with a wide CSP (Figure 18) will continue to have a wide CSP after delivery, but the largest series we located described persistence of a transvaginally diagnosed enlarged CSP in the second trimester into the third trimester in 7 of 8 fetuses. This series noted than an enlarged CSP

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**Figure 15.** Hydranencephaly. Note the characteristic appearance of the thalami and brain stem protruding within a cystic cavity without any frontal cortex present (the latter allowing differentiation from HPE and extreme hydrocephalus); as expected, there is absence of the CSP. As is often the case in hydranencephaly, the falx is present. **A**, Axial second-trimester sonogram, with anterior to the reader’s left, shows the intact falx, complete lack of anterior cerebral tissue, and abnormal posterior tissue. **B** and **C**, Coronal third-trimester MRIs confirm the intact falx, lack of cerebral rind, and absent CSP and show the characteristic appearance of hydranencephaly.
may be associated with hydrocephalus, chromosomal translocation, and growth restriction but concluded that when an enlarged CSP is an isolated finding, termination of pregnancy is not justified, although the fetal karyotype should be obtained until more is known about the importance of this entity. The differential diagnosis for an enlarged CSP has been reported to include the so-called vein of Galen aneurysm (although with knowledge of neuroanatomy and access to Doppler technology, this possibility should not be seriously entertained), arachnoid cyst, and dilated third ventricle; the latter can be distinguished by its location between the thalami, whereas the CSP is above the thalami and anterior to the third ventricle. The true clinical importance of a wide CSP prenatally is thus unknown, but it has been stated that this finding should be followed by a detailed search for associated anomalies and postnatal imaging and developmental evaluation may be indicated.

Conclusions

The CSP is an important marker for normal fetal CNS development. Although not as crucial as assessment of the lateral ventricular atrium and cisterna magna, never assume that nonvisualization of the CSP is “technical”; an absent CSP may be a marker of serious brain malformation. In their oft-quoted postnatal MRI study, Barkovich and Norman concluded that absence of the CSP was never a normal finding. More experience has been obtained with prenatal imaging since then, confirming that absence of the CSP is rarely an isolated finding.

Figure 16. Fenestrated CSP. Coronal second-trimester sonogram shows absence of the normal CSP. This fetus has dilated third (3) and lateral (L) ventricles; other images showed a normal fourth ventricle. The obstructive hydrocephalus resulting from aqueductal stenosis has torn the CSP so that only a remnant (arrow) is visualized.

Figure 17. Encephalocele. A, Suboccipital bregmatic sonogram, with anterior to the reader’s left, at 26 weeks shows encephalocele (asterisk) but no CSP. The faint hypoechoic structure seen anteriorly in the midline (arrow) represents the fornix, a normal structure caudal to the CSP that is commonly mistaken for the CSP. The normal CSP must be a boxlike fluid-filled structure that is bordered on either side by the medial walls of the frontal horns of the lateral ventricles. B, Axial follow-up sonogram, with anterior to the reader’s left, at 32 weeks shows colpocephaly and absence of the CSP; note how easy it is to miss absence of the CSP unless one is specifically looking for it.
Conversely, presence of the CSP virtually ensures normal development of the central forebrain, thereby excluding many complex malformations. In the most recent American Institute of Ultrasound in Medicine guideline, intracranial structures represent the minimum elements of a standard examination of fetal neuroanatomy: cerebellum, choroid plexus, cisterna magna, lateral cerebral ventricles, midline falx, and CSP. In their seminal 1989 article “Detection of Fetal Central Nervous System Anomalies: A Practical Level of Effort for a Routine Sonogram,” Filly et al. state that “identification of this easily demonstrated structure (the CSP) provides an added measure of certainty that the fetal CNS has developed normally.”

Equally as important as proper identification of a normal CSP is not mistakenly calling other structures the CSP in the setting of abnormal midline development. In other words, “don’t make it up!” A classic pitfall already discussed is incorrectly attributing the paired columns of the fornix as the CSP; remember that the CSP is located superior to the fornix and lacks the central linear echo seen in the fornix (Figure 4). Another potential error is simply noting fluid between the lateral margins of the frontal horns and calling that a normal CSP. The CSP must range from the medial wall of the frontal horn of one lateral ventricle to the medial wall of the frontal horn of the contralateral lateral ventricle; note that in SOD (Figure 13), the normal “box” between the medial wall of the frontal horns is not appreciated, a subtle abnormality that can easily be mistaken as a normal CSP. In the example of schizencephaly (Figure 14) again note the absence of the normal box between the medial wall of the frontal horns. Also, take care not to confuse the high-riding third ventricle or prominent interhemispheric fissure for the CSP (Figure 6); as mentioned before, the CSP is located anteriorly and should be directly between the frontal horns of the lateral ventricles. As a final potential pitfall, remember that the CSP et vergae is normal. Many sonologists struggle with a “big CSP” and mistakenly call it some type of midline cyst, when in fact it is just a long and normal CSP et vergae (Figure 3).

There are 4 main reasons why awareness regarding the CSP is important. First, it is required by guidelines promulgated by multiple societies. Second, it is a marker for ACC, one of the most common CNS anomalies. Although ACC can be diagnosed after 18 to 20 weeks, especially in expert hands, it can be a difficult diagnosis. Failure to visualize the CSP and colpocephaly are crucial clues in suggesting ACC. Third, nonvisualization of the CSP is often one of...
the only clues to the diagnosis of certain entities, classically SOD. Fourth, knowledge of the CSP provides an impetus to review and study many other more obvious entities affecting the central forebrain (eg, HPE, porencephaly, hydranencephaly, and severe hydrocephalus).

In summary, we have presented information regarding the anatomy and embryology of the CSP; scan techniques for prenatal sonography and MRI of this structure; prenatal examples of a wide variety of entities, both common and uncommon, associated with absence of the CSP; differential diagnosis of various entities associated with an absent CSP; and the potential clinical importance of many of these entities. Visualization of a normal CSP is a strong indicator of proper development of the central forebrain.

References

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