

Ultrasound Imaging of Fetal Brain Abnormalities

Three Essential Anatomical Levels

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Abstract: Prenatal ultrasound evaluation of the fetal brain requires documentation of specific structures according to guidelines set by the American College of Radiology and the American Institute of Ultrasound in Medicine. Among these required structures are: cerebellum, cisterna magna, lateral cerebral ventricles, choroid plexus, midline falx, and cavum septum pellucidum. All these structures can be visualized in three crucial planes of imaging that include cisterna magna/cerebellum, cavum septum pellucidum, and ventricular atria. A systematic approach can be achieved by instituting a protocol that includes these three basic levels of imaging of the fetal head. These anatomical levels should be incorporated in daily routine for complete obstetric ultrasound evaluation.

Key Words: abnormalities, brain, fetal, imaging, ultrasound

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LEARNING OBJECTIVES

After reading this article and completing the posttest, the reader should be able to

- Identify the ultrasound prenatal appearance of the cisterna magna, cerebellum, cavum septum pellucidum, and ventricular atria
- Describe the abnormalities of the brain involving these structures
- Use the three levels of imaging of the fetal brain into their daily practice

Brain abnormalities comprise a wide spectrum of conditions ranging from developmental errors to vascular accidents. This variability results in innumerable possibilities of findings on prenatal ultrasound, which could create some diagnostic dilemmas.^{1–3} To compound this problem, the fetal brain lends

itself to an infinite number of planes of imaging, which is determined mostly by fetal position and other technical factors that can compromise visualization of fetal anatomy. In the vast majority of cases, sonographers are given a protocol for required images for documentation. Whenever possible, sonologists personally scan patients prior to terminating the study. In many instances however, sonologists are asked to review studies done in remote places or render a second opinion on studies done by another ultrasound laboratory. It is in these instances that certain standard landmarks become necessary for consistent assessment of fetal anatomy in spite of different expertise, equipment, and protocol.

The American College of Radiology (ACR) and the American Institute of Ultrasound in Medicine (AIUM) have issued guidelines for fetal brain imaging incorporated in the accreditation program of both organizations. These guidelines can be viewed on the web sites of these organizations (www.acr.org/accreditation and www.aium.org/publications). Among the required structures are: cerebellum, cisterna magna, lateral cerebral ventricles, choroid plexus, midline falx, and cavum septum pellucidum. All of these structures can be documented in three levels of imaging that include the cerebellum and cisterna magna inferiorly, cavum septum pellucidum at the middle, and ventricular atria superiorly.⁴ The demonstration of these structures will incorporate crucial areas of the brain that can serve as valuable clues to the presence or absence of anomalies. Maintaining the discipline to image these structures and defining them on every second and third trimester examination is good clinical practice that can significantly improve the accuracy of prenatal diagnosis.⁵

CEREBELLUM AND CISTERNA MAGNA LEVEL

The cerebellum is located in the posterior cranial fossa and consists of two hemispheres connected by the vermis. Two cerebellar tonsils extend caudally from the base of each hemisphere. The cisterna magna is a fluid collection posterior to the cerebellum that is persistently present in neonates. On ultrasound, the cerebellar hemispheres are normally echo-poor to moderately echogenic, bounded superiorly by the echogenic tentorium cerebelli. The bony cranium of the posterior fossa borders its dorsal and caudal aspects. The vermis is a highly echogenic structure that can be recognized as an ovoid density at the midline on axial and sagittal sections. It separates the cisterna magna from the fourth ventricle. The cisterna magna may be seen as an echo-free triangle with its point oriented towards the cerebellar vermis. The anteroposterior diameter of the cisterna magna should be 2 to 10 mm prenatally.⁶

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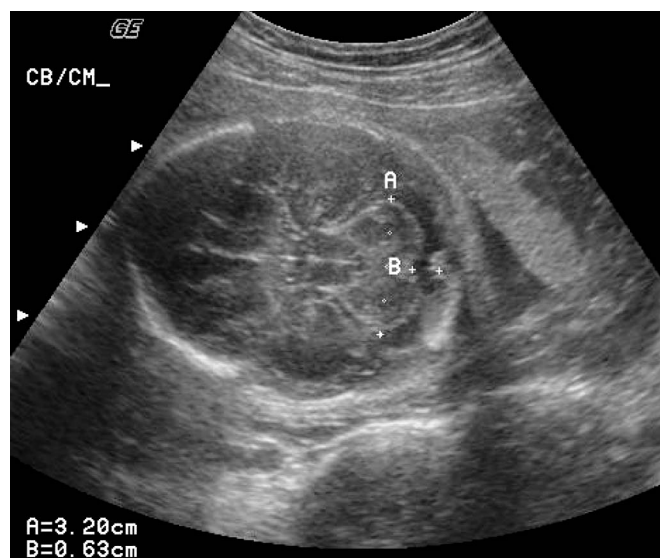


FIGURE 1. Level of cerebellum and cisterna magna. This is taken about 15 to 30 degrees above the canthomeatal line. The transcerebellar diameter (A) and the anteroposterior dimension of the cisterna magna (B) are measured.

In prenatal ultrasound, an axial plane 15 to 30 degrees from the canthomeatal line visualizes both the cerebellum and cisterna magna (Fig. 1). This plane is usually reached by starting with the level where the standard biparietal diameter (BPD) is obtained, then exaggerating the posterior tilt of the transducer to include the cerebellum. Even if the focus is on the cerebellum and cisterna magna, it is also at this level that measurement of the nuchal skin can be performed in the early second trimester (Fig. 2). Care must be taken when performing an examination for this purpose. To obtain an accurate measurement, the posterior fossa structures should be lined up with the third ventricle and cavum septum pellucidum. Any measurement exceeding 5 mm is considered abnormal and should be considered a marker for Down's syndrome.⁶ In the third trimester, this measurement is not performed since fetuses at this stage of gestation tend to have increased subcutaneous fat and easily exceed 5 mm.⁷

Among the brain abnormalities that may be recognized at this level are enlargement of the cisterna magna due to

overall dysmorphic brain development or Dandy Walker malformation and its variants^{8,9} (Fig. 3). The "banana sign" in fetuses with Chiari II malformation is also seen at this level. This is due to the small posterior fossa resulting in crowding of the cerebellum. The cerebellar hemispheres become oriented anteriorly and appear to wrap around the cerebral peduncles giving rise to the elongated crescentic "banana" (Fig. 4). Cephaloceles tend to occur in this region as one of the most common locations for brain or meningeal herniation¹⁰ (Fig. 5). To make this diagnosis, one must be able to demonstrate an opening in the calvarium at the site of herniation. When this is not possible, the differential diagnosis of a cystic hygroma should be included. In addition to abnormalities that can arise from structures that are normally in the posterior fossa, arachnoid cysts can occur in this location as in any other location in the brain. A large arachnoid cyst that occurs in the midline of the posterior fossa may be difficult to differentiate from the enlarged fourth ventricle in a Dandy Walker malformation due to the associated mass effect on the cerebellum. In this instance, the definition of a normal vermis will favor the diagnosis of arachnoid cyst since the vermis is dysgenetic in Dandy Walker malformations. Arachnoid cysts that manifest in the posterior fossa cause mass effect that has more significant sequelae due to the smaller posterior fossa and the vital areas present in this region.¹¹⁻¹³

CAVUM SEPTUM PELLUCIDUM LEVEL

The septum pellucidum is a thin plate consisting of two laminae located between the fornix and corpus callosum. At birth, these laminae are not yet fused, giving rise to a fluid-filled space called the cavum septum pellucidum (CSP), which is usually obliterated by the age of 3 to 6 months. Its parallel walls are normally no further apart than 1 cm, although the cavum may be relatively large in preterm infants. During prenatal ultrasound the cavum septum pellucidum can be usually encountered during the routine measurement of biparietal diameter where it is seen anterior and superior to the third ventricle (Fig. 6). It could vary in shape from a triangular to a trapezoidal fluid collection, which does not have a single designated normal measurement. However, the CSP has been demonstrated to increase in size relative to the increasing head circumference and advancing gestational age.¹⁴ It can also be

FIGURE 2. Nuchal skin thickening. The normal thickness of the nuchal skin is less than 5 mm at this level (A). Abnormal measurements are highly suggestive of Down's syndrome (B).

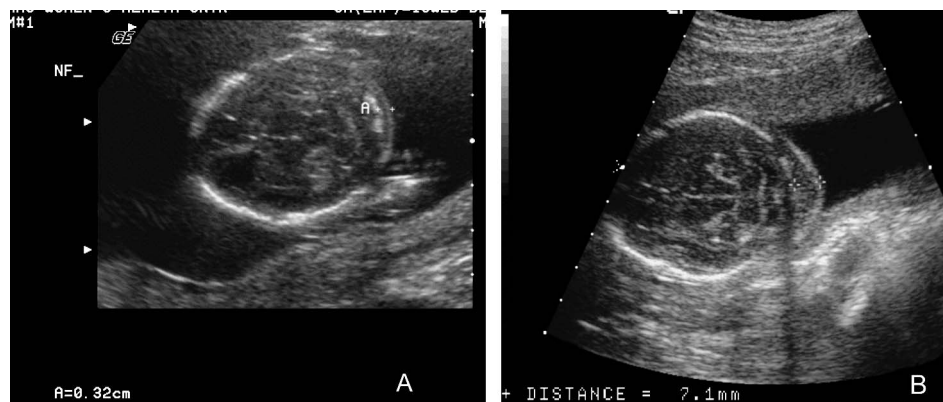


FIGURE 3. Dandy Walker malformation. A, There is marked enlargement of the cisterna magna (arrow). The vermis anterior to the cisterna magna is hypoplastic. B, There is severe ventriculomegaly with enlargement of the third ventricle (arrow).

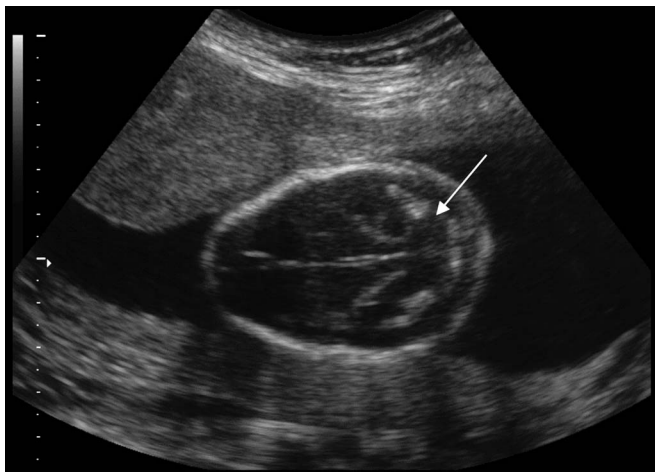
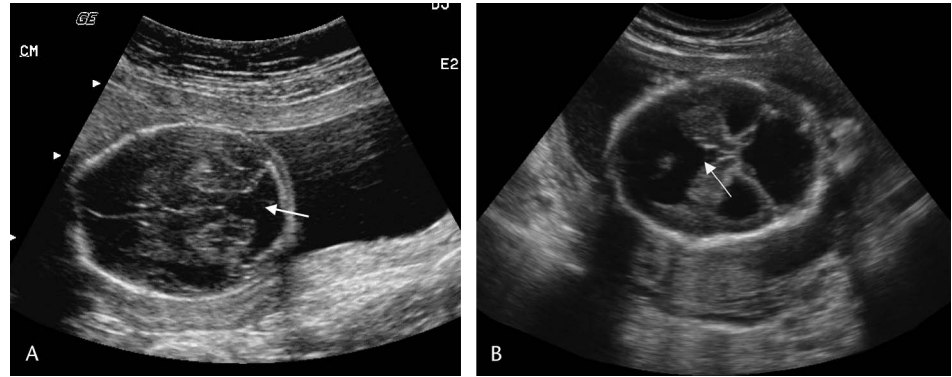


FIGURE 4. Banana sign. There is obliteration of the cisterna magna in a Chiari II malformation. The cerebellum (arrow) is elongated and wraps around the cerebral peduncle.



FIGURE 6. Cavum septum pellucidum. At the BPD level, the cavum septum pellucidum (arrow) is seen anterior to the third ventricle and posterior to the interrupted falx.

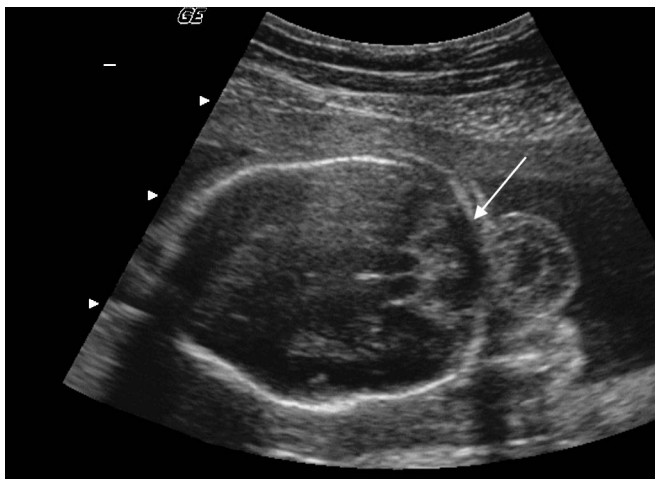


FIGURE 5. Occipital cephalocele. There is a defect in the calvarium at the occiput. Brain is seen herniating through the defect (arrow).

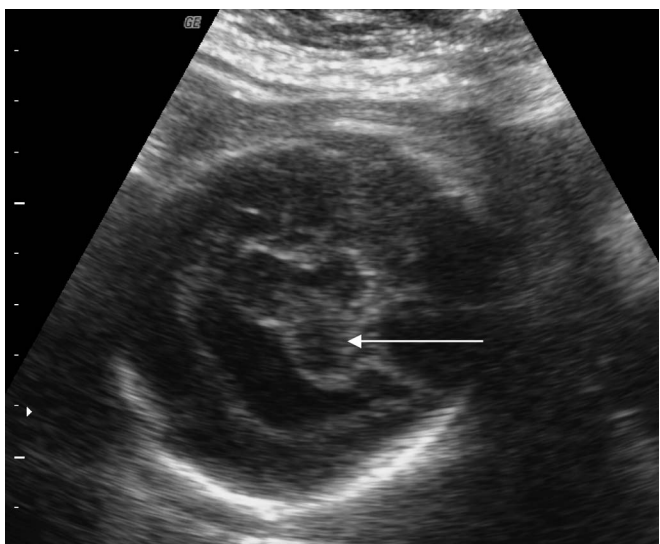


FIGURE 7. Alobar Holoprosencephaly. The thalamus is fused (arrow) and there is a monoventricle that is not interrupted by falx at midline. The cavum septum pellucidum is not visualized.

FIGURE 8. Agenesis of the corpus callosum. Axial images of the brain show absence of the cavum septum pellucidum, colpocephaly (short arrows), and parallel orientation of the frontal horns (long arrows).

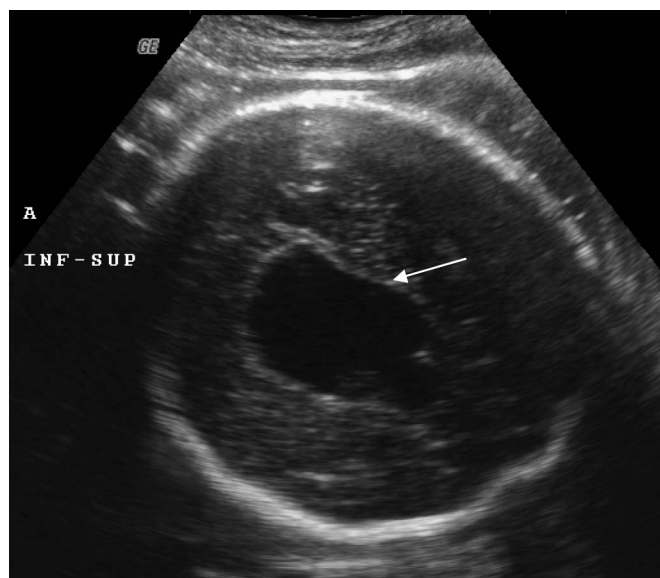
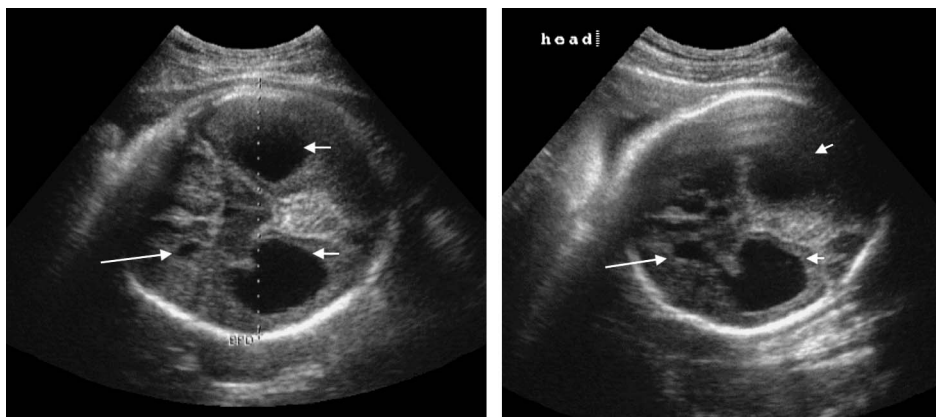


FIGURE 9. Suprasellar arachnoid cyst. A well-circumscribed cystic mass (arrow) is seen just above the thalami, which was associated with agenesis of the corpus callosum.

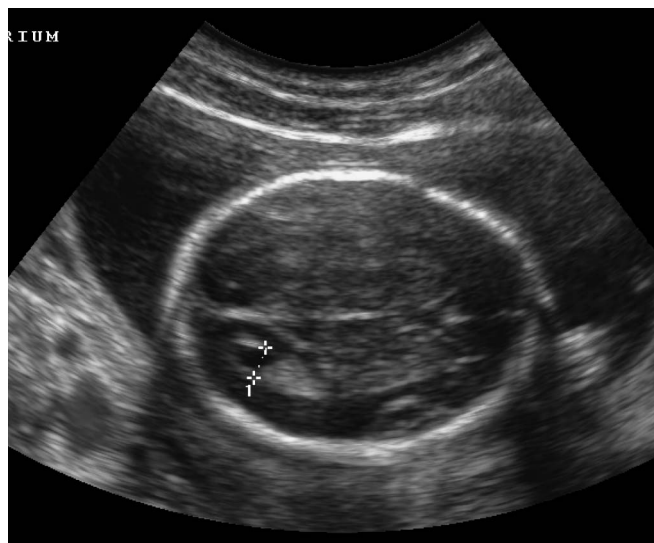


FIGURE 10. Ventricular atrial level. The ventricular atrium is measured in transverse diameter (cursors) just posterior to the glomus of the choroid plexus. This measurement is not more than 10 mm in a normal fetus.

FIGURE 11. Mild ventriculomegaly. In a normal ventricle (A), the choroid plexus fills the transverse diameter of the body of the ventricle. In mild ventriculomegaly, there is separation between the medial wall of the ventricle and the choroid (B).

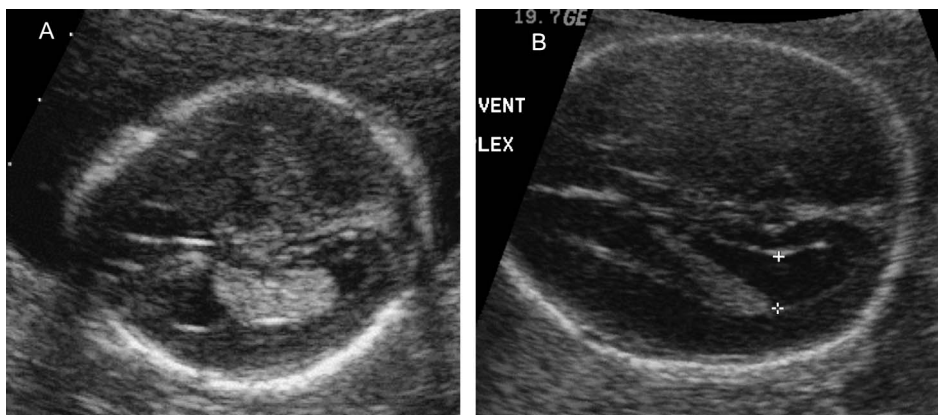
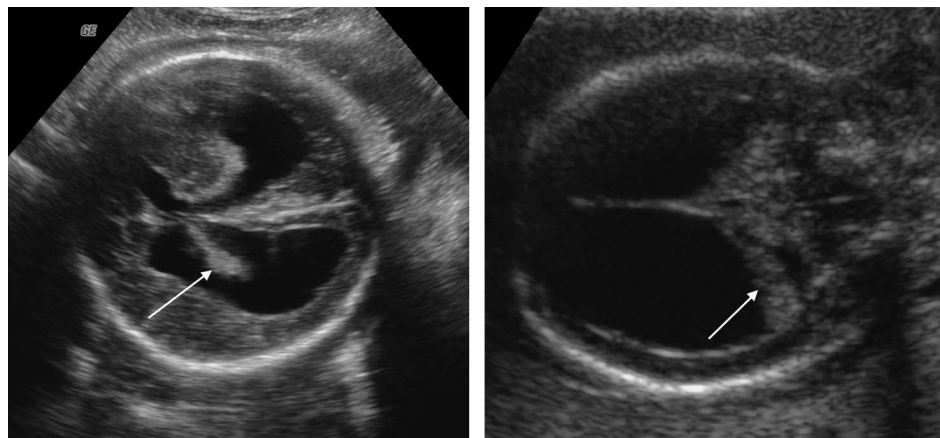


FIGURE 12. Moderate and severe hydrocephalus. The dependent choroid plexus (arrow) dangles within the markedly dilated ventricle. No matter how dilated the ventricle becomes, there is always a rim of brain that surrounds the periphery of the ventricle.



seen on sagittal or coronal views as a constant fluid-filled landmark that should be documented in all fetuses.¹⁵

The presence of the CSP is a reassuring sign that midline structures in the brain have developed and differentiation has occurred to a level where the right and left sides of the brain are distinguishable. In cases of holoprosencephaly,¹⁶ the CSP cannot be found in the alobar and semilobar variety (Fig. 7). In its most differentiated form called lobar holoprosencephaly, the CSP may be seen. Absence of the CSP is also another hallmark of agenesis of the corpus callosum.^{17,18} In this case however, an associated dilated, high-riding third ventricle can mimic the cavum septum pellucidum and other views may be needed to make this differentiation. When this is suspected, other findings such as colpocephaly, parallel orientation of the frontal horns, and associated suprasellar cysts complete the diagnosis^{19–21} (Figs. 8 and 9). A pitfall in the diagnosis of absent CSP occurs in severe hydrocephalus where markedly enlarged frontal horns can compress the CSP. In these cases, a coronal image through the frontal horn may increase the chances of finding the CSP or defining the corpus callosum. Other abnormalities where CSP is not seen are schizencephaly^{22,23} and hydranencephaly where the absence of brain tissue includes the midline.

VENTRICULAR ATRIA LEVEL

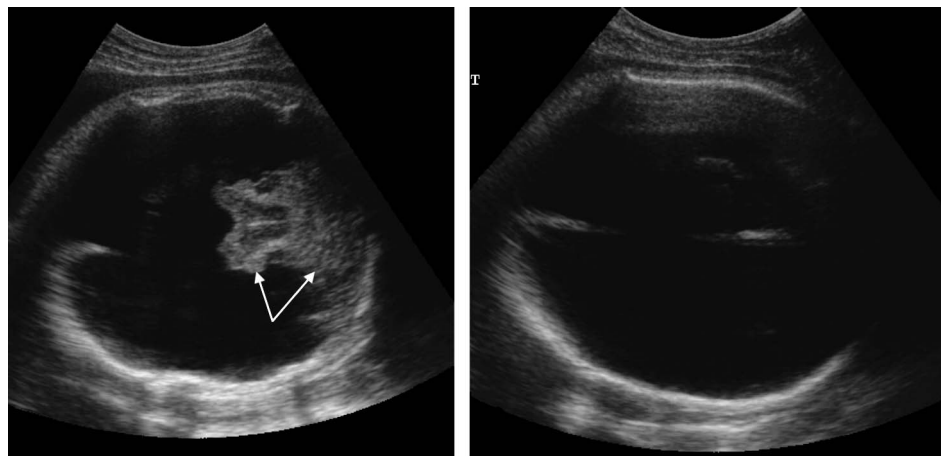
The atrium of the lateral ventricle is a triangular space that curves around the pulvinar of the thalamus. It is located posterior to the temporal horn, anterior to the occipital horn and inferoposterior to the body of the lateral ventricle. The choroid plexus extends from the floor of the body of the lateral ventricle, through the atrium to end on the roof of the temporal horn. As the choroid plexus bends through the atrium, it forms the prominent glomus of the choroid. In the prenatal period, the ventricular atrium is routinely imaged and measured as a gauge of ventricular size (Fig. 10). The transverse ventricular diameter should be no larger than 10 mm at any time during pregnancy.²⁴ When this measurement is borderline, comparison of several serial examinations becomes important. Because of this, it becomes crucial to consistently reproduce this view on every study. From the BPD plane, the transducer is tilted superiorly until the choroid plexuses can be seen as

a pair of echogenic linear structures that point toward each other in an “arrowhead” configuration. The angle of the transducer can then be adjusted until the occipital horn is seen as a fluid collection posterior to the choroid plexus. It is important to maintain symmetry when measuring the atria so as not to take a falsely enlarged measurement due to an oblique plane. Also, it is important to visualize both occipital horns to make sure they are symmetric. Because of the reverberations from the near skull interface, it may be difficult to see the non-dependent ventricle on a direct axial plane. Other planes of imaging (coronal or sagittal) may be necessary to visualize the atria in the near field. In most cases, the choroid plexus fits snugly within the body of the lateral ventricle along its axial plane. In early or mild ventriculomegaly, the borders of the choroid plexus separate from the walls of the lateral ventricle^{25,26} (Fig. 11). In the later stages, the change in the orientation of the dependent choroid gives rise to the “dangling choroid plexus sign” (Fig. 12). It is important to remember however that no matter how dilated the lateral ventricle becomes in cases of hydrocephalus, a rim of cerebral mantle



FIGURE 13. Brain infarction. At the level of the ventricular atrium (cursors), severe brain infarction has destroyed the majority of the surrounding brain mantle anteriorly.

FIGURE 14. Hydranencephaly. The absence of brain high in the convexities distinguishes hydranencephaly from severe hydrocephaly. The midbrain and posterior fossa (arrows) are normal.



should always be visible around its periphery.²⁷ In most cases, the thickness of this mantle is taken into consideration when making prognostic decisions about fetal outcome.^{28–31}

A specific type of ventriculomegaly occurs in complete agenesis of the corpus callosum (ACC). This is called “colpocephaly” and refers to the disproportionate enlargement of the occipital horn relative to the milder enlargement of the frontal horn. This has become characteristic of ACC and should lead to other findings such as parallel orientation of the ventricles, splaying of the frontal horns, dilation of a high-riding third ventricle, and absence of the CSP.^{15,18}

The plane of imaging that passes through the ventricular atria also includes the parietal convexities and part of the frontal, temporal, and occipital lobes depending upon the obliquity of angulation. Therefore, any vascular insult to this part of the brain resulting in hemorrhage, porencephaly, or encephalomalacia will be detected at this level^{32–36} (Fig. 13). Also, in the differentiation of hydranencephaly from other causes of severe ventriculomegaly, the absence of the parietal cortex at this level helps in making the distinction (Fig. 14). Since only branches of the middle cerebral artery supply the parietal convexities, collateral circulation from the anterior or

posterior cerebral arteries will not affect the finding of deficient cortical brain mantle, which clinches the diagnosis of hydranencephaly.³⁷ The surrounding calvarium can also be important in the diagnosis. The “lemon sign” is an indirect sign of Chiari II malformation defined by the concavity of the frontal bones of the skull³⁸ (Fig. 15). This is a result of decreased intracranial pressure brought about by decompression of the cord from an opening in the spine. It is important to look for this finding cephalad to the temporal lobe because a “pseudolemon sign” can result from imaging at the level of the temporal bone. Finally, it is at this level that choroid plexus cysts can be detected. Although they can be isolated findings in an otherwise normal fetus, there is an increased association with Trisomy 18. Choroid plexus cysts have become synonymous with this diagnosis but have also been described as a marker in other karyotypic abnormalities as well.^{39–41} Most often, the detection of choroid plexus cysts is incidental to the initial purpose of measuring the ventricular atria (Fig. 16).



FIGURE 15. Lemon sign in Chiari II malformation. There is indentation of the frontal bones of the skull (arrows). There are also associated ventriculomegaly present.

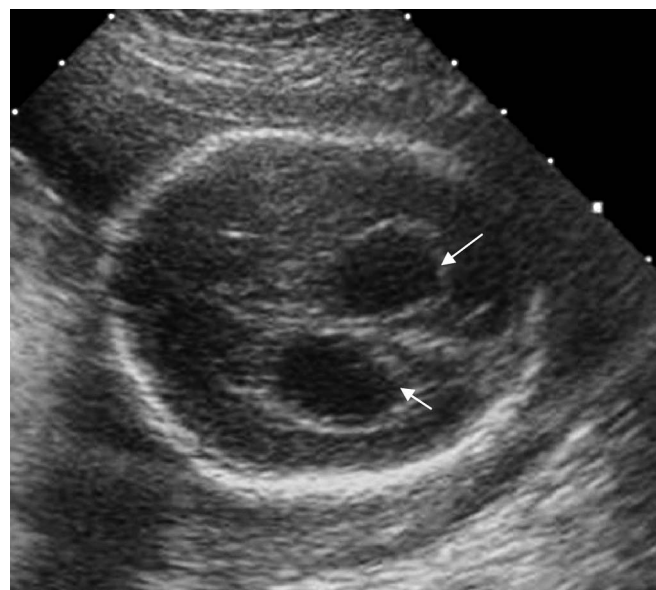


FIGURE 16. Choroid plexus cysts. Large cysts (arrows) fill the atria of both ventricles in this fetus with Trisomy 18.

This is especially true in a population of pregnancies that are not at high risk for chromosomal anomalies. Because of this known association however, further search for other anomalies is usually initiated and the screening ultrasound may be converted to a targeted examination. This is the next level of diagnosis that follows once suspicions for anomalies are raised. This entails a more dedicated exam that requires a higher level of expertise that is focused on a specific organ or diagnosis.

CONCLUSION

Imaging the fetal brain is limited primarily by the position of the fetal head and technical factors such as maternal body habitus and lack of adequate amniotic fluid.⁴² The diagnosis of brain anomalies in the fetus therefore must depend upon constant landmarks that could be documented in normal fetuses.⁴³ These landmarks are part of the requirements for a comprehensive OB ultrasound examination as outlined by guidelines of the ACR and AIUM. Three levels of imaging comprise the minimum planes that will satisfy these guidelines. These are: the cerebellum/cisterna magna level, cavum septum pellucidum level, and ventricular atria level. When these normal structures are documented, the vast majority of congenital brain anomalies can be reliably excluded. However, any subtle changes that could suggest the presence of anatomical dysmorphism warrant further investigation. They could be valuable clues to a more complex problem, which could prove invaluable in the prognosis of the fetus.^{44,45} Once an abnormality is documented at any of these levels, neonatal correlation of the abnormality becomes possible because these same landmarks can be reliably reproduced in the neonate.⁴⁶ The incorporation of these structures in the daily protocol for documentation of fetal anatomy can enhance the diagnostic accuracy of any comprehensive obstetric examination.

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