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Prenatal-postnatal correlations of brain abnormalities: how lesions and diagnoses change over time

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Abstract

A combination of prenatal ultrasound and MRI can be used to detect and characterize many primary and secondary CNS abnormalities in the developing fetus. While this information is useful in prenatal patient counseling, it is important to understand the factors that can influence change in diagnosis and prognosis over time. The etiology of the abnormality, the conspicuity of associated findings, the change in appearance over time, and the opinion of subspecialty experts all can influence the diagnosis. Additionally, technical factors of imaging acquisition may allow the detection of an abnormality in the postnatal period and not prenatally. Having an understanding of the normal fetal central nervous system anatomy at varying gestational ages will aid in the imaging detection and interpretation of CNS pathology. Understanding how these appearances and diagnoses can change over time will aid in the discussion of prognosis with expectant parents, which is crucial in fetal CNS abnormalities.

Introduction

When an abnormality is detected on fetal sonography, the question of how to counsel the pregnant patient becomes paramount. While the diagnosis of some anomalies is straightforward, and the prognosis well-described and documented, for other anomalies the outcome is less certain. Many abnormalities of the central nervous system (CNS) fall into this second category. It is not just the initial diagnosis that impacts outcome, but also associated abnormalities and the potential of other abnormalities to develop or become more apparent over time. This review illustrates the changing appearance of CNS abnormalities from prenatal to postnatal life to highlight the issues that need to be addressed when counseling patients at the time of initial diagnosis of a CNS abnormality. Many of the cases described in this review were taken from a prospective study of fetal ventriculomegaly (VM).[1–4] Studies were initially interpreted independently and in a blinded fashion by obstetric and pediatric imagers and by neuroradiologists, allowing for descriptions of how diagnoses may change not just by the timing of the examination but also by the specialist reviewing the examination and by knowledge of other imaging findings.[3]

Understanding why diagnoses may change from prenatal to postnatal life may aid in our ability to better counsel patients at the time of prenatal diagnosis. On average, we found that disagreements were common among readers of postnatal imaging studies after a fetal diagnosis of VM, and were more likely as the degree of prenatal VM increased and as the complexity of prenatally diagnosed CNS anomalies increased.[3]

One of the ways in which to categorize changes that occur over time is to assess the etiology of the change. There can be an increase in VM that leads to increases in compression of the

adjacent parenchyma or destruction due to ischemic or hemorrhagic events. There can be ischemia, hemorrhage or infection that lead to infarction and porencephaly. There can be developmental changes that become more apparent over time. In addition, diagnoses may differ due to differing opinions by subspecialists. There can be changes in diagnosis based on criteria for prenatal and postnatal imaging diagnoses. Finally, imaging techniques can be used postnatally (such as gadolinium enhanced MRI) that may allow for a diagnosis not possible with the non-contrast imaging performed in utero.

Another method to assess changing diagnosis over time uses the common prenatal appearance that can be due to a variety of factors that become evident over time, eg, the low signal lining ventricles on T2weighted imaging due to germinal matrix, blood products, heterotopias, or subependymal nodules in tuberous sclerosis.

A final method is to categorize brain abnormalities by their etiology and time/location of development, and then to describe the types of changes that can be seen with specific diagnoses.

Many of these categories overlap, and thus, in this review we use a combined approach to discuss the heterogeneous reasons for differences in prenatal and postnatal diagnoses of CNS abnormalities.

Changing degree or severity of an anomaly over time

Ventriculomegaly

A prime example of a finding that can change over time is the degree of fetal VM. VM is a common end-point for a variety of pathological processes, including obstruction, malformation and destruction. VM can be an isolated finding, but it is often associated with other neuronal, somatic, and karyotypic anomalies which have been shown to occur in conjunction with, or are the source of, enlarged fetal ventricles (rather than simply the degree of dilatation) that account for most of the morbidity and mortality associated with VM.[5–12] Therefore, identifying associated anomalies is extremely important to improve prognostic information.

However, it has also been shown that more anomalies are seen in association with larger ventricular size, and therefore assessment of ventricular diameter is important.[4,11] In our study of prenatal to postnatal imaging diagnosis, we found that the risk of postnatal diagnosis of structural abnormalities not diagnosed prenatally increases as the degree of VM increases, with new structural abnormalities seen in 0% of fetuses judged to be normal prenatally, 4% of fetuses with VM of 10–12 mm prenatally, and 36% of fetuses with VM of 12.1–15 mm prenatally.[3]

VM is an anomaly that can change appearance over time depending upon the underlying etiology of the ventricular dilatation. In some cases, prenatal VM will entirely resolve in the neonatal period and no other associated abnormalities will be detected (Fig. 1). In other cases, the ventricles can enlarge over time (Fig. 2), particularly when associated with obstruction. This can lead to increased degrees of VM and increased compression of surrounding parenchyma. If severe obstruction occurs, this can lead to hemorrhage, infarction, and ultimately porencephaly.

Similarly, cysts can change in appearance over time. It may be that the cysts newly develop or communicate with the ventricles, and thus enlarge if there is communicating hydrocephalus, or change in appearance due to other reasons (Fig. 2).

Chiari malformations

Chiari II malformations are the typical class of Chiari malformations seen in utero and are associated with abnormal primary neural tube closure pathologies such as encephaloceles, meningoceles and myelomeningocele.[13] It is well-described that Chiari malformations in association with neural tube defects (NTD) can increase in severity over time, with a recent study demonstrating reversal of hindbrain herniation in patients treated with *in utero* surgery for NTD.[14] Additionally, VM in association with a Chiari II malformation with NTD can increase in severity during pregnancy.[15] After birth, shunting will of course decrease the degree of the VM.

Chiari I malformation is less commonly diagnosed in utero. It is used to describe both congenital and acquired causes of tonsillar herniation. The true congenital Chiari I malformation is thought to arise from developmental mesodermal abnormalities,[16] which lead to severe reduction of the posterior fossa volume. Acquired Chiari I malformations may be secondary to intracranial hypertension or hypotension, pathologic malformations of the skull base, or other pre- or postnatal causes of hydrocephalus.[17] In patients with a diagnosis of Chiari I malformation, MRI of the craniocervical junction typically demonstrates tonsillar herniation >5mm, though the exact number is both age-dependent and controversial particularly when herniation ranges from 3–5 mm.[17,18] Though the distance of herniation is often used in suggesting the diagnosis, the overall volume of the posterior fossa is likely a better reflection of patients who may be symptomatic. Therefore, tonsillar herniation of <5mm does not exclude the diagnosis, and imaging and clinical symptoms should be assessed in combination to make the diagnosis.[16] Figure 3 demonstrates the interval development of a Chiari I malformation between the third trimester and one year of age.

Injuries to the fetus during the normal developmental process

Certain congenital CNS abnormalities result from injuries to the fetus during the normal developmental process. The most common example of such an acquired injury is that of intracranial hemorrhage. Other abnormalities include hydranencephaly, cerebral atrophy, porencephaly, periventricular leukomalacia, parenchymal infarcts, and calcifications. A variety of factors can result in these pathologic entities including infection, ischemia, anoxia, toxins, trauma or maternal metabolic disease.[20]

Subdural hemorrhages are very common in newborns as a result of vaginal birth, and do not typically result in a poor prognosis for the neonate.[21, 22] Obviously this type of bleed, which occurs intrapartum, will not be seen at the time of fetal imaging.

Other bleeds are common in utero. Blood products change in appearance over time, both on sonography and MRI. When hemorrhage occurs in utero, it can cause obstruction of CSF outflow which can lead to VM. Hemorrhage can also cause obstruction of terminal veins resulting in venous infarction. Thus a prenatal hemorrhage may be seen as a region of porencephaly after birth (Fig. 4). Additionally, the stress of delivery can result in intracranial hemorrhage, particularly if the structural integrity of the vasculature is weak, leading to new porencephalic cysts postnatally.

Infection in utero is another common cause of acquired fetal CNS abnormalities. Traditional TORCH (toxoplasmosis, rubella, cytomegalovirus (CMV), herpes simplex) infections can all lead to CNS abnormalities, though toxoplasmosis and CMV are particularly neurotropic (Fig. 5). Toxoplasmosis can cause meningoencephalitis which can result in hydrocephalus, microcephaly, calcifications, porencephaly and hydranencephaly. CMV is considered the most serious infection because it can lead to some of the same CNS abnormalities as

toxoplasmosis; but, in addition, periventricular leukomalacia, migrational abnormalities (lissencephaly and polymicrogyria), and cerebella hypoplasia have been described.[42]

Developmental changes that become more apparent over time

Cortical migrational abnormalities

One of the more straight-forward reasons for changing appearance in the CNS over time is that of cortical development. Our ability to detect and characterize sulcation of the fetal brain begins at 20–23 weeks of gestation and increases through the third trimester.[23] If fetuses are scanned in the first half of pregnancy, cortical migrational abnormalities are typically missed since the sulci which eventually will appear abnormal have not yet completely formed. Even later in gestation, the slightly thickened cortical ribbon, which is a key to the diagnosis of polymicrogyria, can be a subtle finding.

In our study comparing prenatal to postnatal diagnosis, we found that of 119 fetuses initially referred for VM who had imaging postnatally, diagnoses pre- and postnatal disagreed 84/119 (74%) times. In fetuses who went on to have neonatal MRI, 40/53 (75%) had disagreements between prenatal and postnatal MRI diagnoses, the most common clinically important new diagnoses were dysgenesis of the corpus callosum (N=15) and polymicrogyria (N=9).

Lissencephaly, or smooth brain, cannot be confidently diagnosed prior to 24 weeks since the lack of sulci, which are diagnostic of this condition, is a normal finding early in gestation. Similarly, when heterotopias are present these may not be visible early in gestation. Heterotopias are regions of abnormal migration of normal neurons to unexpected locations, are often inapparent in utero and not recognized until either later in pregnancy or postnatally.[3] Patients can have a variety of symptoms depending upon the number of heterotopias and the location. Heterotopias may be subependymal or subcortical. Subependymal heterotopias on MRI are typically visualized as round nodules that are isointense to normal gray matter on all pulse sequences. Though often bilateral, there is a predilection for the right hemisphere due to the later migration of right-sided neuroblasts.[32] Subcortical heterotopias can have a nodular, curvilinear or mixed morphology. They typically extend from the ventricular surface to the deep white matter. The overlying cortex is often decreased in size with shallow sulci due to the decrease in neurons.[32] Figure 6 is an example of a irregular dark signal in lining the ventricle, which prenatally was felt to represent subependymal hemorrhage, which postnatally seen to be secondary to heterotopias.

The CNS lesions of tuberous sclerosis also may not be apparent early in gestation. The most common underlying brain lesions of tuberous sclerosis are subependymal nodules and cortical tubers, which often are not apparent early in fetal development. They have been reported as early as 21 weeks.[24] The tubers appear dark on T2WI in fetuses either due to magnetic susceptibility artifact (from calcification) and/or low mobile water concentration. The cerebral abnormalities of tuberous sclerosis change appearance on MRI from early neonatal MRI to childhood and adulthood,[30] due to changes in the underlying myelination of the brain. Therefore, it is common that additional cerebral abnormalities will be seen in neonatal life and childhood that were not visualized on fetal imaging (Fig. 7).

Hemimegalencephaly is a hamartomatous process involving an asymmetric overgrowth of one or a part of one cerebral cortex. Hemimegalencephaly is sometimes associated with neurocutaneous syndromes such as neurofibromatosis type I, epidermal nevus syndrome, proteus syndrome, unilateral hypomelanosis of Ito, Klippel-Trenaunay syndrome, and tuberous sclerosis.[29] Hemimegalencephaly also may be difficult to diagnosis early in

pregnancy (Fig. 8) and prenatal diagnosis of this condition has only been recently reported. [25–28] In the few cases where it was diagnosed prenatally, the diagnosis was suggested by routine second trimester ultrasound but only confirmed with fetal MR or 3D ultrasound. Ultrasound features indicating the diagnosis in a recent case series of two patients included asymmetry of the cerebral hemispheres and/or lateral ventricles, macrocephaly, displacement of the occipital lobe across the midline and asymmetric dilation of the occipital horn of the contralateral ventricle.[25] MR imaging typically reveals an overall increased size of the cerebral hemisphere, abnormal gyration, and VM.[31]

Abnormalities in myelination

MRI of preterm infants <30 weeks of gestational age demonstrates the earliest signs of myelination at 25 weeks gestation in the cerebellum, brain stem, and thalamus with later myelination at 36 weeks in the cortical spinal tracts of the pre- and post- central gyri, corona radiata, posterior limb of the internal capsule, and lateral geniculate bodies.[33] Most myelination is not complete until postnatal age 2. It makes sense, therefore, that disorders of myelination will not be seen early in gestation, may infrequently be seen in the third trimester, and typically will not be diagnosed until postnatal life.

Changes in diagnosis based on criteria for prenatal and postnatal imaging

VM in prenatal imaging is clearly defined as a measurement of 10 mm at the level of the atria. From our prospective study of fetuses with prenatal VM, we discovered that disagreements in the diagnosis of VM from the prenatal to the postnatal period often arose because of the lack of a standard definition for VM in neonates,[3] as no clear established standard exists for postnatal ventricular size. Measurement technique differs: on prenatal ultrasound we use a slightly oblique axial view in order to depict the atrium of the lateral ventricle. On postnatal head ultrasound, ventricles, when measured, typically are imaged in an angled coronal plane, from the anterior fontanelle, angled back to the ventricular atrium. In our study on prenatal-postnatal consensus diagnosis, we found a significant difference of opinion ($P < 0.01$) between radiologists interpreting postnatal cranial ultrasound regarding when VM should be diagnosed postnatally. Even on postnatal MRI with axial imaging planes, ventricles can measure larger than 10 mm and still be classified as not enlarged. Therefore, without an interval change in the size of the ventricles, a fetus could be given an in-utero diagnosis of VM, while after delivery (even on the same day) be considered normal because of the lack of a standardized definition.[3] Figure 9 is a case of prenatally diagnosed VM with decreasing size of the ventricles later in gestation. Postnatal MRI showed smaller ventricles which still qualified as VM by prenatal size criteria; however VM was not diagnosed by all readers.[3]

Similar appearances of anomalies with more accurate diagnosis dependent on time of imaging

Etiology of dark T2 areas lining the ventricles

Low signal on T2 weighted images may be visualized in focal regions lining the ventricles, and most frequently is due to normal germinal matrix early in gestation and to imaging artifacts at the interface of the fluid filled ventricle and the parenchyma. However, low signal in this region can also represent areas of hemorrhage or heterotopia. To confirm that an area of dark signal reflects true pathology we recommend that a lesion: 1) has to be seen in at least two scanning sequences in orthogonal planes; and 2) has to cause a contour deformity of the ventricular wall.[24] This is particularly useful in assessing for heterotopias (Fig. 6) and subependymal nodules in tuberous sclerosis (Fig.7).

Appearance of enlarged ventricles in septo-optic dysplasia and holoprosencephaly

When VM is severe, the differential diagnosis of the etiology includes obstruction, dysgenesis and destruction (or a combination of these factors). Thus a fetus may have hydrocephalus, septo-optic dysplasia, holoprosencephaly, or hydranencephaly. Typically, assessment of the surrounding parenchyma, visualization of the falx and other midline structures aid in the diagnosis. However, at times the distinction can be difficult, particularly when midline structures are obscured by the severe degree of VM (Fig. 6, 10 and 11).

Prenatal sensitivity for detection of alobar and semilobar holoprosencephaly is much greater than the prenatal detection of lobar variant holoprosencephaly and septo-optic dysplasia.[34] In a study of children diagnosed with holoprosencephaly from 1998 through 2002 (by definition, these were less severe forms of holoprosencephaly since the children are alive and were referred for assessment), only 22% (21/95 with prenatal ultrasound) were diagnosed as having holoprosencephaly prenatally.[35] 50% of children with alobar holoprosencephaly and 83% with semilobar holoprosencephaly were not diagnosed until after birth. Lobar and middle interhemispheric forms of holoprosencephaly can be more difficult to detect by prenatal ultrasound, since in these forms the interhemispheric non-separation can be quite subtle and the characteristic dorsal cyst of holoprosencephaly is often absent.[35]

Etiology of “holes in the brain:” schizencephaly vs. porencephaly

While postnatal diagnosis is typically more accurate than prenatal diagnosis, (given limitations of fetal position, fetal motion, and fetal size) on rare occasions, the fetal images may give information about the etiology of a lesion that is not apparent postnatally. An example of this is shown in Figure 11 where porencephaly is suggested prenatally due to the difference in signal intensity in the fluid in the cyst compared with the cerebrospinal fluid in the ventricles of the brain, whereas postnatal diagnosis (initially interpreted without the benefit of knowledge of prenatal imaging) was that of schizencephaly.

Schizencephaly is an example of a malformation that results from abnormalities during proliferation/apoptosis, migration, and cortical organization. Schizencephaly is a full-thickness cerebral cleft lined by gray matter; it is thought to be a disorder of cortical maturation because the cells lining the cleft demonstrate varying degrees of abnormality. The exact etiology of schizencephaly is uncertain and can be genetic or secondary to an insult in pregnancy. Clinical manifestations of schizencephaly depend upon the location, laterality, and whether it is an open or closed lip variety. Some cases may require the review of both prenatal and postnatal imaging to make a final accurate diagnosis.

Distinguishing schizencephaly from porencephaly is important because they are often caused by different pathologies. Although both can be acquired in utero, the predisposition for developing one or the other is very different. For example, porencephaly may result from vascular injury particularly in patients with acquired vasculopathies such as factor V Leiden disease. Additionally, patients with porencephalic cysts due to weakened vessels, may be prone to strokes in the future.[37]

Different imaging techniques

Ultrasound and MRI

The standard prenatal evaluation of the fetus is by second trimester ultrasound. If a CNS abnormality is suspected, then the patient may go on to fetal MRI. It is important to know the limitations of each modality in counseling prospective parents in the prenatal period. The postnatal diagnosis may differ depending on the modality with which the infant with CNS

abnormality is imaged after birth. In our study on why diagnosis changes from prenatal to postnatal life, we had 31 subjects in whom both a postnatal cranial ultrasound examination and postnatal MRI were performed after a referral for suspected fetal VM. 27/31 had disagreements in postnatal diagnosis between ultrasound MRI. After excluding decisions to diagnose VM and those with no clinical consequence, there were 23 studies with 21 major new findings. The most common findings seen on postnatal MRI which were not recognized on postnatal cranial ultrasound were migrational abnormalities (n = 8), hemorrhage (n = 7) and infarction (n = 4).[3]

Callosal abnormalities are another example of CNS pathology often detected on prenatal ultrasound. By sonography, portions of the corpus callosum can be visualized by 16 weeks gestation [40] and the entirety by 20 weeks gestation using transvaginal sonography.[41] Agenesis of the corpus callosum can be an isolated finding, though is often associated with additional CNS findings, karyotypic and syndromic abnormalities.[38] These additional findings which are crucial for prenatal patient counseling are often not seen on prenatal ultrasound. In a study of 10 fetuses with callosal dysgenesis on ultrasound, 63% had additional findings on follow-up fetal MRI.[39] The additional associated findings are important to be aware of because children with isolated callosal dysgenesis often have only mild developmental delay that resolves over time whereas children with additional findings frequently have persistent developmental delay.[39] Figure 12 shows an irregular appearance to the corpus callosum with areas of thinning on postnatal ultrasound. However, on follow-up postnatal MRI, the corpus callosum was described as normal. Therefore it is important to recognize the limitations of the imaging modality being used when diagnoses from different imaging modalities differ.

Postnatal MRI with additional sequences and with gadolinium

Certain CNS abnormalities may be present in gestation, though not apparent on fetal MRI due to limitations from rapid image acquisition, limited sequences, and lack of use of intravenous contrast agent. Examples include the ability to complete susceptibility imaging in neonatal MRI (i.e., for the detection of hemorrhage, Fig. 4f) that is not easily acquired in prenatal imaging due to the rapid imaging speed needed to image the moving fetus. Gadolinium contrast may highlight an abnormality in neonatal imaging (i.e., brainstem tumor) that was less obvious on non-contrast fetal imaging (Fig. 13).

Opinion of experts

Inter-observer agreement is an issue whenever the accuracy of imaging diagnoses is being investigated. The same finding can be noted, but there can be differences of opinion as to the etiology and nomenclature used to describe the abnormality. In addition, human error can occur, as there can be errors of observation. In fetal imaging differences of opinion can occur that stem from the primary specialty of the individual interpreting the study. Knowledge of the perinatologist or radiologist performing an ultrasound, expertise of the radiologist be it obstetric imaging, pediatric imaging, or neuroradiology or pediatric neuroradiology can all influence the manner in which diagnoses are made. Additionally, our prospective studies of both prenatal imaging and pre- and postnatal imaging in fetuses with VM demonstrate that disagreements are much more common when the fetus has a larger number of abnormalities.[1, 3]

Conclusion

A combination of prenatal ultrasound and MRI are used to detect and characterize many primary and secondary CNS abnormalities in the developing fetus. This information is

useful in prenatal patient counseling, but understanding the factors that can influence change in diagnosis and prognosis over time is crucial.

The etiology of the abnormality, the conspicuity of associated findings, the change in appearance over time, and the opinion of subspecialty experts all can influence the diagnosis. Additionally, technical factors of imaging acquisition may allow the detection of an abnormality in the postnatal period and not prenatally. Having an understanding of the normal fetal central nervous system anatomy at varying gestational ages will aid in the imaging detection and interpretation of CNS pathology.

Understanding how these appearances and diagnoses can change over time will aid in the discussion of prognosis with expectant parents, which is critically important in fetal CNS abnormalities.

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Fig. 1. Interval resolution of ventriculomegaly

(a) Axial T2 weighted MR at 25 weeks gestational age demonstrates borderline ventriculomegaly with the atria measuring 10 mm. Neonatal head ultrasound (b) and (c) on day of life 50 (with a 159 day interval from the fetal MR) depicts resolution of ventriculomegaly and normal intracranial anatomy.

Fig. 2. Progression of ventriculomegaly and interval development of a para-midline cyst
Axial T2 weighted MR image of the fetal brain at 27 weeks gestation (**a**) demonstrates agenesis of the corpus callosum and no frontal cyst. Axial T1-weighted neonatal brain MRI taken on day of life 1 (**b**) demonstrates increasing size of the frontal and occipital horns of the lateral ventricles and a new finding of a frontal para-midline cyst (C). Reproduced with permission from reference. [43]

Fig. 3. Interval development of a Chiari I malformation

(a) Sagittal T2 weighted MR at 29 weeks gestational age shows a normal cisterna magna (arrowhead). (b) Sagittal T1 weighted brain MRI at 13 months (388 days) of age (with a 455 day interval from the fetal MR) shows a new Chiari malformation (arrow). Mild ventriculomegaly seen prenatally did not change in the interval (not shown).

Fig. 4. Fetus with VM and porencephaly at 35 weeks gestational age with new areas of hemorrhage on followup

(a) Axial ultrasound at 35 weeks gestational age shows ventriculomegaly and disruption of the ventricular margin with fluid extending into the parenchyma (white arrow) consistent with porencephaly. Axial (b) and Sagittal (c) T2 weighted MRI at 35 weeks gestational age shows the ventriculomegaly and porencephaly (white arrows). The small low signal intensity area in the temporal horn is due to intraventricular blood (black arrow). Coronal (d) head sonogram on day of life 10 (19 days after prenatal US) shows persistence of VM and porencephaly (white arrow) with interval development of a new porencephalic region (arrowhead). (e) Axial susceptibility sequence on day of life 1 (with a 9 day interval from fetal MRI) shows extensive regions of hemorrhage.

Fig. 5. CMV infection

Oblique axial US (**a**) at 29 weeks gestational age shows periventricular calcifications (arrowhead) and abnormal echogenicity in the periventricular parenchyma (thin arrow) suggesting congenital infection. Axial T2 weighted fetal MR (**b**) demonstrates irregularity of the cortex with a sawtooth appearance (thick arrow) consistent with polymicrogyria which was more evident postnatally rather than prenatal. Note that the calcifications are more evident on the ultrasound. Sagittal T1 weighted neonatal MRI (**c**) on day of life 2 (with a 79 day interval from the fetal MRI) demonstrates VM and cerebral calcifications (arrowhead).

Fig. 6. Prenatal ventriculomegaly with suspected subependymal hemorrhage changed to holoprosencephaly with heterotopias postnatally
Axial (a) T2 weighted MRI at 22 weeks gestational age demonstrates ventriculomegaly and irregularity of the right ventricular wall (arrows) thought at the time to be due to hemorrhage. Axial T2 (b) weighted MRI on day of life 1 (with a 49 day interval from fetal MRI) demonstrates multiple abnormalities including holoprosencephaly with bilateral nodules lining the frontal horns (arrows) diagnosed as neuronal heterotopias.

Fig. 7. Subependymal nodules of tuberous sclerosis

Sagittal (a) and axial (b) T2 weighted images at 34 weeks' gestational age show low intensity periventricular nodules (arrows). Axial T1 weighted postnatal image (c) confirms those nodules and shows additional lesions as well. Reproduced with permission from [44].

Fig. 8. Interval recognition of hemimegalencephaly

Coronal (a) T2 weighted MRI at 22 weeks gestational age in a fetus with ventriculomegaly demonstrates an area of low signal in the subependymal region (arrowhead) that prenatally was felt to represent hemorrhage. Note the mild irregularity of the cortex on the right (arrows) that was not appreciated prenatally. Axial T2 weighted MRI (b) on day of life 1 (with a 122 day interval from fetal MRI) shows resolution of VM and asymmetry of the cerebral hemispheres and increased conspicuity of the right frontal gyral abnormality consistent with hemimegalencephaly (arrows). The low signal in the subependymal region seen prenatally corresponded to an area of migrational abnormality (not shown) and not hemorrhage as suspected previously.

Fig. 9. Prenatal to postnatal decrease in ventricular size with not all readers diagnosing ventriculomegaly postnatally

Axial (a,b) prenatal ultrasounds at 30 weeks gestational age demonstrate severe ventriculomegaly with echogenic blood lining the ventricular walls. Axial T2 MR image (c) shows similar findings. Postnatal coronal (d) ultrasound on day of life 47 (with a 100 day interval from prenatal US) show ventricles measuring 12 mm; not all readers postnatally diagnosed these ventricles as being enlarged. Axial (e) T1 postnatal MR image shows similar findings. Figure 9c reproduced with permission from reference [3].

Fig. 10. Interval diagnosis of septal leaflet dysgenesis

(a) Axial T2 weighted MRI at 22 weeks gestational age demonstrates ventriculomegaly. The septal leaflet abnormality was not recognized. Coronal (b) head ultrasound on day of life 1 (92 day interval from prenatal US) shows a new diagnosis of dysgenesis of the septum pellucidum. Coronal (c) T2 weighted MRI at 4 days of age (with a 96 day interval from the fetal MRI) confirms the absence of septal leaflets.

Fig. 11. Example of error in postnatal diagnosis with prenatal imaging correctly diagnosing porencephaly

Sagittal (a), coronal (b), and axial (c) T2 weighted MRI at 33 weeks gestational age shows a large head with ventriculomegaly and discontinuity of the ventricular margin extending into a fluid collection (white arrows) felt to represent porencephaly. On the coronal image the fluid collection is of slightly different signal intensity than the interventricular cerebrospinal fluid. (d) Axial T2 weighted MRI on day of life 27 (with a 62 day interval from fetal MRI) read without knowledge of prenatal findings, described the region of prior porencephaly as schizencephaly. On consensus review, porencephaly was felt to be the appropriate etiology. Other findings include holoprosencephaly (which prenatally was diagnosed as

ventriculomegaly), postnatal development of extraaxial fluid collection on the right (arrowheads), and new visualization of polymicrogyria (black arrows). Figure 11b reproduced with permission from reference [3].

Fig. 12. Disagreement of callosal dysgenesis on postnatal ultrasound and MRI
Sagittal postnatal ultrasound (**a**) on day of life 1 shows incomplete visualization of the corpus callosum. Sagittal T2 weighted MRI (**b**) at 4 days of age demonstrates an intact corpus callosum and absent septal leaflets (same patient as figure 10).

Fig. 13. Interval recognition of congenital CNS tumor

(a) Sagittal T2 weighted MRI at 36 weeks gestational age in fetus referred for ventriculomegaly. Note the bulge (arrow) above the tectum that was not recognized prospectively. (b) Sagittal T1 weighted contrast enhanced MRI on day of life 22 (23 day interval from fetal MRI) a mildly hyperintense mass above the tectum (arrow). This was felt to be a hamartoma or low grade glioma causing hydrocephalus due to obstruction at the level of the aqueduct. (a) reproduced with permission from [3].