Learning objectives

MRI is being increasingly used to evaluate the fetal brain, particularly when a fetus is at increased risk for neurodevelopmental disabilities or when an abnormality has been detected on prenatal. The aims of this study were:

To demonstrate the role of MRI in exploration of fetal brain

To illustrate the most frequent brain abnormalities
Fetal MR is a valuable complement to prenatal sonography and is a powerful technique used to evaluate the fetal brain. Fetal MR imaging has higher contrast resolution than prenatal sonography and allows better differentiation of normal from abnormal tissue. Structural abnormalities such as cerebral malformations and destructive lesions can be sonographically occult on prenatal sonography yet detectable by fetal MR imaging. The continued advances in MR techniques, such as diffusion weighted and parallel imaging, fetal MR imaging offers the promise of contributing to our understanding of normal as well as abnormal brain development.
Utility and Limitations of Fetal brain MRI

1/ Limitation of fetal brain sonography:
Fetal MR imaging has improved contrast resolution compared with prenatal sonography. Moreover, fetal MR imaging also allows direct visualization of both sides of the fetal brain. This is an advantage over sonography, where the more anterior cerebral hemisphere is often shadowed by the reverberations from the overlying structures, resulting in visualization of only the more posterior cerebral hemisphere. Additional limitations of sonography, resulting from decreased amniotic fluid volume, fetal positioning, and acoustic shadowing from the ossifying calvaria, can also be overcome by fetal MR imaging.

2/ Limitation of fetal brain MRI:
One of the limitations of fetal MR imaging is that of fetal motion. Because sedation for fetal MR imaging is not used, it was only after the advent of rapid T2-weighted pulse sequences (where a single image can be acquired in less than 1 second) that fetal MR imaging became embraced as a clinically important imaging technique. Even with rapid image acquisition, however, fetal motion can still affect the quality of the study.

Clinical indications
Fetal MRI is most performed to evaluate a suspected abnormality detected by prenatal sonography. Fetal MRI can provide information that can assist in prenatal counseling of the current pregnancy as well as counseling of the recurrence risk in future pregnancies. The most common indications for imaging the fetal brain will be briefly discussed below and include mild ventriculomegaly, suspected callosal agenesis, complications of monochorionic twinning, and posterior fossa abnormalities.

1/ Ventriculomegaly (Fig.1, 2, 3)

Fetal ventriculomegaly refers to the presence of dilated cerebral ventricles in utero. It can result from a number of underlying pathological mechanisms which include: CNS structural anomalies (Dandy-Walker continuum, Chiari II malformation, aqueductal stenosis, agenesis of the corpus callosum or in utero fetal infections (toxoplasmosis infection, CMV infection). Radiographic assessment: Fetal ventriculomegaly is defined as an axial diameter >10 mm across the atria of the posterior or anterior horn of lateral ventricles at any gestation or alternatively a separation of more than 3 mm of the choroid plexus from the medial wall of the lateral ventricle.

The severity of ventriculomegaly can be further classified as:

- Mild fetal ventriculomegaly lateral ventricular diameter between 10 and 15 mm.

- Severe fetal ventriculomegaly (also sometimes classified as fetal hydrocephalus): lateral ventricular diameter >15 mm.

2/ Dysgenesis of the corpus callosum (Fig.4,5,6)
Dysgenesis of the corpus callosum may be complete (agenesis) or partial and represents an in utero developmental anomaly.

It can be divided into:

- Primary agenesis: the corpus callosum never forms
- Secondary dysgenesis: the corpus callosum forms normally and is subsequently destroyed

Isolated partial dysgenesis of the corpus callosum is often asymptomatic. The clinical picture in other cases is dictated by the associated abnormalities that are frequently found, especially in agenesis. Children with agenesis may have dysmorphic facies, most commonly demonstrating hypertelorism. MRI is the modality of choice in evaluating both the corpus callosum and the frequently associated anomalies. Features include:

1/ Ventricles:
- Ventricles run parallel rather than the normal "bow-tie" configuration giving a racing car appearance on axial imaging.
- Colpocephaly (dilatation of the trigones and occipital horns) gives a characteristic "longhorn"/moose head/viking helmet appearance on coronal imaging.
- Dilated high-riding 3rd ventricle communicating with the interhemispheric cistern or projecting superiorly as a dorsal cyst.

2/ Cortex:
- Bundles of Probst
- Radial gyri (absent cingulate gyrus)
- Everted cingulate gyrus

3/ Limbic system:
- Hypoplastic fornices
- Hypoplastic hippocampi

Dandy-Walker malformation (Fig 7, 8, 9)

A classic Dandy-Walker malformation was the traditionally described form of the Dandy-Walker continuum and comprises of the triad of: 1. Hypoplasia of the vermis and cephalad rotation of the vermian remnant 2. Cystic dilatation of the fourth ventricle extending posteriorly 3. Enlarged posterior fossa with torcular-lambdoid inversion (torcular lying above the level of the lambdoid due to abnormally high tentorium)

Holoprosencephaly (HPE) is a rare congenital brain malformation, resulting from incomplete separation of the two hemispheres.

Classically three sub types have been recognised, however additional entities are now included in the spectrum of the disease. The three main sub types, in order of decreasing severity are: alobar holoprosencephaly, semilobar holoprosencephaly and lobar holoprosencephaly.

4-1: Alobar holoprosencephaly:

As will most cerebral structural congenital abnormalities, lobar holoprosencephaly is visible on all modalities, but in general is identified on antenatal ultrasound (if performed), and best characterised by MRI.
The basic structure of the cerebral hemispheres is lost, with variable amounts of residual cortex. Features include:

- Single midline monoventricle (or holosphere) - Lateral and third ventricles are absent - Absent midline structures: absent septum pellucidum, agenesis of the corpus callosum, absent inter hemispheric fissure and falx cerebri dorsal cyst of holoprosencephaly. Associated craniofacial features may also be present which include:

4-2 : Semi-lobar holoprosencephaly:

The basic structure of the cerebral lobes are present, but are fused most commonly anteriorly and at the thalami and there is partial diverticulation of brain (dorsal cyst). Features include:

- Absence of septum pellucidum - Monoventricle with partially developed occipital and temporal horns - Rudimentary falx cerebri: absent anteriorly - Incompletely formed interhemispheric fissure - Partial or complete fusion of the thalami - Absent olfactory tracts and bulbs - Agenesis or hypoplasia of the corpus callosum - Incomplete hippocampal formation

In addition, it is associated with mild degree of facial abnormality such as hypotelorism and cleft lip.

4-3: Lobar holoprosencephaly:

Unlike the more severe forms, in lobar holoprosencephaly the cerebral hemispheres are present. Features include:

- Fusion of the frontal horns of the lateral ventricles - Wide communication of this fused segment with the third ventricle - Fusion of the fornices - Absence of septum pellucidum - Agenesis or hypoplasia of the corpus callosum

Unlike semilobar holoprosencephaly, the falx is present, the interhemispheric fissure is fully formed and the thalami are not fused.

5/ posterior fossa abnormalities:

5-1/ mega cisterna magna : (Fig.11)

A mega cisterna magna refers to adult patients with significantly enlarged CSF retrocerebellar cisterns in the posterior fossa with normal cerebellar morphology. Controversy remains around whether mega cisterna magna is a normal anatomical variant or due to volume loss of the cerebellum. Mega cisterna magna has also been referred to as Blake’s pouch or retrocerebellar arachnoid pouch.

Typically seen as prominent retrocerebellar cerebrospinal fluid (CSF) appearing space with a normal vermis and normal cerebellar hemispheres
Differential diagnosis

Mega cisterna magna needs to be distinguished from other causes of an (Differential diagnosis):

- arachnoid cyst - epidermoid cyst - cerebellar atrophy/cerebellar hypoplasia - Dandy-Walker malformation

**5-2/ Chiari malformation:**

Chiari malformations are a group of defects associated with congenital caudal 'displacement' of the cerebellum and brainstem. Three types were described, with a fourth added later. Types II and III are likely to be related to each other.

**Chiari I malformation:** the most common Peg-like cerebellar tonsils displaced into the upper cervical canal through the foramen magnum. MRI is the imaging modality of choice. On sagittal imaging, the best plane for assessing for the presence of Chiari I malformations, the tonsils are pointed, rather than rounded and referred to as peg-like. The sulci are vertically oriented, forming so-called sergeant stripes. Axial images through the the foramen show crowding of the medulla by the tonsils.

**Chiari II malformation:** displacement of the medulla, fourth ventricle and cerebellum through the foramen magnum. It usually associated with a lumbosacral spinal myelomeningocele

MRI

- Small posterior fossa with a low attachment of the tentorium and low torcula
- The brainstem appears 'pulled' down with an elongated and low lying fourth ventricle
- The tectal plate appears beaked: inferior colliculus is elongated and points posteriorly, with resulting angulation of the aqueduct which results in aqueductal stenosis and hydrocephalus
- Cerebellar tonsils and vermis are displaced inferiorly through foramen magnum which appears crowded

**Chiari III malformation:** Features similar to Chiari II but with an occipital and/or high cervical encephalocele

**Chiari IV malformation:** Chiari IV malformation was a term some authors gave to describe a form of extreme cerebellar hypoplasia. This can be associated with hypoplasia of pons as well as a small funnel shaped posterior fossa. It is now considered to be an obsolete term.
Fig. 1: Axial SSFSE T2-W image in a 24 gestational week fetus demonstrates several ventriculomegaly

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Fig. 2: Axial SSFSE T2-W image in a 24 gestational week fetus: ventriculomegaly

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**Fig. 3:** Coronal SSFSE T2-W image in a 24 gestational week fetus: several ventriculomegaly

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Fig. 4: axial SSFSE T2-W image in a 22 gestational week fetus: agenesis of the corpus collusum

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Fig. 5: Sagittal SSFSE T2-W image in a 22 gestational week fetus: agenisis of the corpus collusum

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**Fig. 6:** Coronal SSFSE T2-W image in a 22 gestational week fetus: agenesis of the corpus collusum

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Fig. 7: Axial SSFSE T2-W image in a 26 gestational week fetus: Dandy-walker malformation with absence of the vermis

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**Fig. 8:** coronal SSFSE T2-W image in a 26 gestational week fetus: Dandy-walker malformation with absence of the vermis

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Fig. 9: Sagittal SSFSE T2-W image in a 25 gestational week fetus: Dandy-walker malformation with inferior vermian hypoplasia

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**Fig. 10:** coronal SSFSE T2-W image in a 18 gestational week fetus: alobar holoprosencephaly

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**Fig. 11:** Sagittal SSFSE T2-W image in a 25 gestational week fetus: Mega cisterna magna

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Conclusion

Fetal MRI is being increasingly used to assess for fetal brain abnormalities. It is now being used increasingly successfully to clarify abnormal ultrasound findings, often resulting in a change of diagnosis or treatment plan. Because fetal MR involves many disciplines, the promise of fetal MR will be best achieved through continued multidisciplinary collaborative efforts.
References


