Magnetic Resonance Imaging of Developmental Brain Malformations in Utero

Essay
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LIST OF ABBREVIATIONS

2D-two dimensional
3D-three dimensional
3v-third ventricle
4v-fourth ventricle
ACC-agenesis of corpus callosum
ASP-agenesis of septum pellucidum
BPC-Blake's Pouch cyst
CC-corpus callosum
CM II-chiari II malformation
CM-cisterna magna
cm-centimeter
CNS-central nervous system
CSF-cerebrospinal fluid
CSP-cavum septum pellucidum
DWM-dandy walker malformation
fig.-figure
FMPSPGR-fast multiplanar spoiled gradient recalled acquisition in steady state
GW-gestational week
GWs-gestational weeks
HASTE-half fourier single shot turbo spin echo.
HME-Hemimegalencephaly
HPE-holoprosencephaly
IHF-interhemispheric fissure
Khz-kilohertz
MIH-middle interhemispheric
MIHV-middle interhemispheric variant
mm-millimeters
MRI-magnetic resonance imaging
MR-magnetic resonance
msec-milliseconds.
NTDs-neural tube defects
NPO-nothing per oral
PMG-polymicrogyria
PVH-periventricular heterotopia
SOD-septo optic dysplasia
ssFSE-single shot fast spin echo
T1-longitudinal relaxation time
T2-transverse relaxation time
TE-echo time
TE_{eff} – echo time effective
TR-repetition time
US-ultrasound
wtd-weighted
WWS-walker warburg syndrome

CONTENTS

CHAPTER 1

Introduction

INTRODUCTION

Magnetic Resonance (MR) is now routinely and widely used in fetal neuroimaging and has proven to be valuable in the detection of many cerebral lesions ***(Garel, 2006).*** Fetal magnetic resonance imaging is a well-established second line imaging modality in identifying complex anomalies of the central nervous system, especially when ultrasound findings are equivocal. It may enable an early and precise diagnosis, which is essential in terms of management of pregnancy and pre-, peri- and postnatal care ***(Dill et al., 2009).***

Cerebral anomalies account for approximately 9% of all isolated anomalies and manifest in 15.9% of multiple malformations, constituting a major drive for prenatal diagnosis. The high resolution of imaging in MRI can identify the critical but extremely small and subtle changes in CNS landmarks, especially at early gestational ages. These changes can manifest in gross MRI morphology, the brain microarchitecture, or the activity of the fetus when assessed in a dynamic sequence ***(Chung et al., 2009).***

Fetal MR imaging is a complement to ultrasound (US) and has several advantages, including visualization of the entire brain (as opposed to ultrasound where the upside cerebral hemisphere is often shadowed because of reverberations from overlying cranium). MR imaging is also capable of assessing the sulcation pattern and developing cortex, which is difficult to visualize on ultrasound. In addition, when an anomaly is detected, fetal MR imaging may provide better definition of the lesion because of improved contrast resolution and identify other lesions not visible on ultrasound. Fetal MR imaging has been demonstrated to accurately detect anomalies within the

second and third trimesters, providing additional information for prenatal counseling and delivery planning ***(Smith and Glenn, 2008).***

In addition, MRI has high spatial resolution, multiplanar capabilities, large field of view, strong image quality, and an ability to characterize chemical tissue properties which favor its use as a secondary diagnostic modality in the assessment of fetal pathologies ***(Chung et al., 2009).***

Fetal MRI is clinically performed to evaluate the brain in cases where an abnormality is detected by prenatal sonography. The most common indications include ventriculomegaly, abnormalities of the corpus callosum, and abnormalities of the posterior fossa. Fetal MRI is also increasingly performed to evaluate fetuses who have normal brain findings on prenatal sonogram but who are at increased risk for neurodevelopmental abnormalities ***(Glenn, 2010).***

Half-Fourier acquired single-shot turbo spin echo (HASTE) and single-shot fast spin echo (ssFSE) T2-weighted images are the mainstay of fetal MRI. Real time MRI allows almost continuous imaging of the moving fetus, using the freshly acquired set of images for orientation, obviating the need for additional scouts and thus improving the efficiency of imaging a moving fetus ***(Tang et al., 2009)***. In addition, T1-weighted images are used to visualize fat and hemorrhage, and gradient echo T2 images are used to visualize hemorrhage. Diffusion-weighted MR imaging can also be performed and is helpful in cases of suspected parenchymal injury ***(Smith and Glenn, 2008)***.

The limitations of fetal MR imaging include fetal motion, the small size of the structure being evaluated and the large distance of the

structure from the receiver coil, which limits the image resolution. Improvements in coil technology, such as parallel imaging with increased number of channels, are resulting in the reduction in these limitations. Maternal claustrophobia and discomfort from lying still for the study duration are also problems, because the MR imaging examination typically lasts at least 45 minutes ***(Smith and Glenn, 2008)***.

In summary, limitations of current radiologic practice necessitate that decisions about care of a pregnant patient and fetus must sometimes be made with inconclusive data with regard to the specific cause or severity of a fetal central nervous system anomaly. Therapeutic choices often depend on a sonographic diagnosis. Alternative imaging is important when a questionable sonographic abnormality is visualized or when an abnormality is definite but the exact diagnosis is uncertain. Use of alternative imaging will decrease any ambiguity in counseling expectant parents. US and MR imaging are complementary, noninvasive imaging methods in the evaluation of high-risk pregnancy. When a central nervous system anomaly is detected at US, MR imaging may then demonstrate additional findings that alter patient counseling and care ***(Levine el al., 1997)***.

The aim of this study is to review the role of MR imaging for Developmental Brain Malformations in utero.

CHAPTER 2

MRI Anatomy Of Fetal Brain

MR IMAGING ANATOMY OF FETAL BRAIN

Embryologic development:

The first neural structure that forms between embryonic days 20 to 27 is the neural tube. The neural tube forms as a result of neural plate edges rising and then folding inwards with fusion to form a hollow neural tube. The anterior neuropore at the most rostral end of the neural tube and the posterior neuropore at the caudal end, are the last segments to close, on embryonic day 25 and 27, respectively. The anterior end of the neural tube begins to expand and by the end of the embryonic period (Gestational week 8) five secondary brain vesicles have formed that establish the primary organization of the brain ***(Stiles and Jernigan, 2010).***

The fetal period extends from the 9th gestational week to the end of gestation. The brain begins as a smooth structure and then develops the gyral and sulcal folding pattern. Neuron formation begins on embryonic day 42 and extends through mid-gestation. Regions of brain that contain cell bodies of neurons are gray matter. Neurons are produced from the germinal matrix regions of ventricular zone. Subsequently these neurons migrate in an orderly fashion forming a six-layered cortex. Once positioned in cortex neurons begin to extend dendritic and axonal processes that form the fiber pathways of the brain. The major fiber pathways make up the brain white matter. Thus brain development in fetal period consists of neuron production,

neuron migration and neuron differentiation ***(Stiles and Jernigan, 2010).***

Cerebral parenchymal layering pattern on MRI:

MR imaging of the fetal cerebrum is characterized initially by the presence of multiple layers that disappear as the brain matures and the sulci form. Knowledge of the timing of appearance of these layers and sulci are important for the proper interpretation of fetal brain MR imaging studies ***(Glenn and Barkovich, 2006a).***

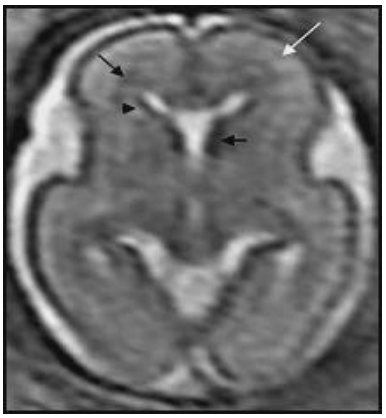

Figure 1: Axial T2-wtd image through lateral ventricles of a 24 GW fetus. From deep to superficial, these layers include the germinal matrix (dark on T2-wtd, short black arrow); periventricular zone (bright on T2-wtd, black arrowhead); then the subventricular and intermediate zones (intermediate-hypointense on T2-wtd, long black arrow); the subplate (bright on T2-wtd, white arrow); and finally the cortex (dark outer brain layer on T2-wtd imaging, no arrow), ***(Chapman et al., 2010).***

This layered pattern of cerebral parenchyma reflects migratory waves of neurons and is apparent between 20 and 28 weeks. Imaging of layering patterns by MRI are demonstrated in **figure 1** in a 24 GW fetus. In addition, **(Table 1)** summarizes these cerebral layering patterns as it appears on MRI at different gestational ages ***(Chapman et al., 2010).***

Gestational age	Number of layers	T2-wtd intensity	Cellular components
16 weeks	3	Low	Germinal matrix
		High	Intermediate zone- few neuroglial cells
		Low	Immature cortex
19-22 weeks	4	Low	Germinal matrix
		Intermediate	Neuroglial migrating cells
		High	Intermediate zone- few cells
		Low	Cortex
27 weeks	3	Low	Germinal matrix
		High	White matter
		Low	Cortex
34 weeks	2	High	White matter
		Low	Cortex

Table 1: MRI T2-wtd signal intensity of brain parenchymal layers with increasing fetal age ***(Chapman et al., 2010).***

MRI of White Matter:

The white matter originates from the intermediate zone, and increases in size by a factor of 3.8 between the 13^{th} to 20^{th} GW. On MR images, the intermediate zone and subventricular zone cannot be separated from each other. They show intermediate signals on T1- and T2-wtd images ***(Prayer et al., 2006).***

Corpus Callosum (CC):

The CC is the largest commissural connection of the cerebral hemispheres. The CC develops between 8 and 20 gestational weeks by guidance of axons from the cortex layers (2, 3 and 5) through the midline to homologous regions in the contra-lateral hemisphere. The

CC is best assessed on fetal MRI using thin (3 mm) midline sagittal images. It appears as a curvilinear T2-wtd hypointense structure at the superior margin of the lateral ventricles **(white arrow in fig. 2),** ***(Glenn, 2009)***.

Deep Grey Nuclei

The caudate, putamen and globus pallidus develop from cells of the germinal matrix areas of the ventral forebrain. At about 27 weeks gestation, they appear more hypointense on T2-wtd images relative to the developing surrounding white matter (**fig. 3),** ***(Glenn, 2009)***.

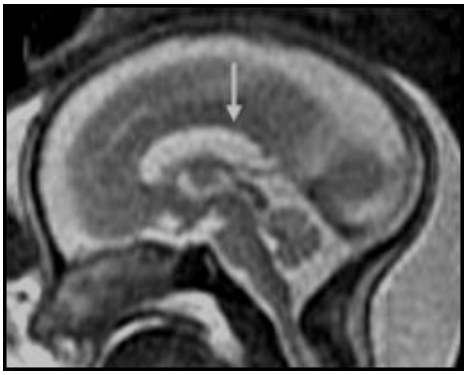

Figure 2

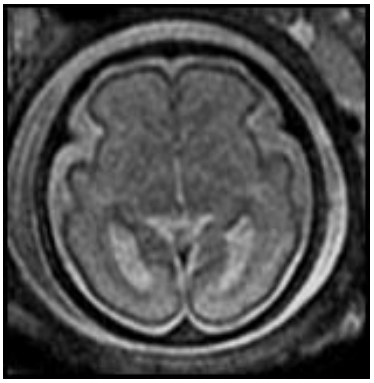

Figure 3

Figure 2: Midline sagittal T2-wtd image of a 26 GW fetus; arrow indicating corpus callosum. **Figure 3:** Axial T2-wtd image of a 27 GW fetus ***(Glenn, 2009).***

MRI of normal sulcation and gyration

During gestation, the surface of the fetal brain gradually changes from a smooth appearance to one with multiple cortical sulci. The appearance of sulci in fetal brain follows an organized spatial and temporal pattern. Primary sulci form first, followed by increasingly complex secondary and tertiary sulci. The appearance of sulci on fetal

MRI has been well described, and has been observed to lag behind that observed on fetal autopsy specimens by an average of 2 weeks. This, in part, is due to limitations of resolution of fetal MRI. There is also a 2-week lag between when a sulcus is first identified on fetal MRI to when it is present in 75% of the fetuses ***(Glenn, 2009)***.

In general, a sulcus initially appears as a smooth, shallow, and wide indentation on the surface of the brain, which progressively deepens and narrows with eventual formation of secondary and tertiary sulci. By 34 gestational weeks, all primary sulci, and some secondary sulci, are visible on fetal MRI. Because sulci appear at a specific time of development, knowledge of the correct gestational age at the time of MRI is critical in interpreting the sulcation pattern. Sulcation is best assessed after 28 gestational weeks were most primary sulci are seen by fetal MRI ***(Glenn, 2009)***.

Appearance of Sulci on MRI:

1)- Parieto-occipital Fissure:

A major part of this fissure is seen on the medial surface of brain hemisphere, although a small part is situated on the lateral surface. On MR images, it is well depicted on a mid-sagittal plane **(white arrow, fig. 4),** ***(Ghai et al., 2006).***

2)- Calcarine Fissure:

This fissure is seen on the medial surface of the occipital lobe on sagittal MR images **(fig. 4, arrowhead)**. On MR images it is best

depicted in a coronal plane through the occipital lobes, where it is seen immediately superior to the tentorium **(fig. 5, white arrow),** ***(Ghai et al., 2006).***

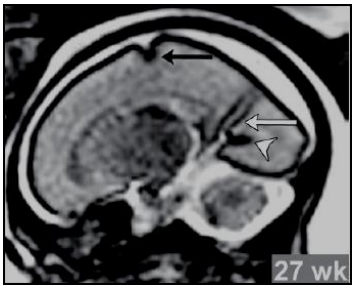

Figure 4

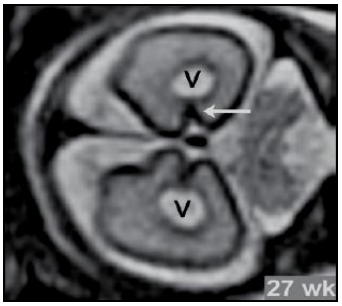

Figure 5

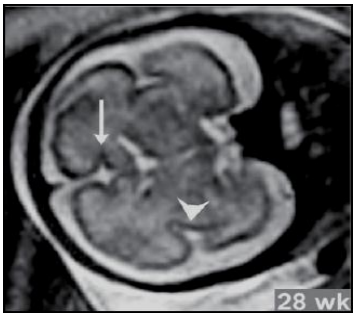

Figure 6

Figure 4 = Sagittal, and **Figure 5 and 6** = Coronal, T2-wtd fetal brain images in a 27 & 28 gestational week (GW) fetuses ***(Ghai et al., 2006).***

3)- Cingulate Sulcus:

The middle part of this fissure is best seen on the medial surface of the brain **(fig. 6, white arrow)** above the cingulate gyrus on coronal MR image ***(Ghai et al., 2006).***

4)- Central Sulcus:

The central sulcus can be identifiable at 26-27 weeks gestation **(black arrow in fig. 4),** ***(Ghai et al., 2006).***

5)- Sylvian Fissure and Insula:

After 17 weeks of gestation, the appearance of the sylvian fossa indentation (insula) takes on a plateau like appearance, with angulation at the margins (the circular sulcus), where it meets the frontal, parietal, and temporal opercula. On MR images, the insular-opercular angles are initially obtuse, but the angulation becomes acute as gestation