Agenesis of Corpus Callosum in a Fetus at 22 Weeks: Role of MRI Based Scoring and MR Tractography in Clinical Decision Making

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Abstract

Keywords

- agenesis
- corpus callosum
- fetal brain
- fetal MRI
- MR tractography

A 32 year old woman at 22 weeks gestation underwent fetal magnetic resonance imaging (MRI) after an ultrasound detected corpus callosum agenesis. MRI confirmed complete agenesis and showed an enlarged left temporal lobe, moderate ventriculomegaly, and abnormal hippocampal orientation. The fetal MRI scoring system indicated a poor prognosis. MR tractography revealed abnormal neuronal connections. Parents were counseled and the pregnancy was terminated due to poor prognosis. This case highlights the value of fetal MRI and tractography in assessing agenesis of the corpus callosum and guiding clinical decisions.

Case

Magnetic resonance imaging (MRI) of the fetal brain was performed in a 32 year female at 22 weeks gestation, following the ultrasound diagnosis of corpus callosum agenesis. There was complete visualization of corpus callosum and cavum septum pellucidum (►Fig. 1A–E). Transverse T2-turbo spin echo (TSE) image of the fetal brain showed asymmetry in the dimensions of the temporal lobes, which was enlarged on the left side (►Fig. 1A). Both Sylvian fissures showed optimal operculization with normal signal intensity and the appearance of bilateral basal ganglia (►Fig. 1B). There was moderate ventriculomegaly with colpocephaly and parallel nonconverging lateral ventricles (►Fig. 1C, black arrows). A dorsal interhemispheric cyst was noted (►Fig. 1D). No signs of abnormal sulcation–gyration were identified (►Fig. 1B–E).

Coronal T2-TSE image of the fetal brain showed altered vertical orientation of the hippocampal cortex on both sides (►Fig. 2A, white arrows). Transverse color-coded diffusion weighted MRI images of the fetal brain showed a 5 layered cerebral mantle with preserved telencephalic lamination (►Fig. 2B). Based on the above mentioned morphological parameters, the fetal MRI based scoring system1 revealed a score of 5/11, which annotates poor neurodevelopmental prognosis in this case.

MR tractography transverse image showed prominent Probst bundles in bilateral frontal lobes with anteroposterior orientation (►Fig. 3A, vertical arrows). Though not seen crossing the interhemispheric fissure, the altered curvilinear course of fibers suggests an aberrant neuronal connection between the right frontal and left occipital lobe, possibly the sigmoid bundle (►Fig. 3B, horizontal arrow). These findings further suggested poor neurodevelopmental outcomes and the parents were counseled accordingly. The pregnancy was promptly discontinued, as the upper gestational age limit for termination of pregnancy in India is 24 weeks.

 Fetuses with agenesis of the corpus callosum (ACC) may have a wide spectrum of neurodevelopmental outcomes ranging from completely normal cognitive and motor functions to severe neurodevelopmental delay. Fetal MRI allows for a detailed assessment of the brain in fetuses with an ultrasound diagnosis of isolated ACC. A fetal MRI based
scoring system helps further categorize these fetuses in terms of neurodevelopmental prognosis and, therefore, helps in better parental counseling and clinical decision making.

MR tractography allows visualization of the neuronal connections in fetuses with ACC. Prenatal delineation of white matter connectivity provides greater insight into clinical outcomes and may improve neurological counseling. MR tractography results correspond well with histology based tractography of fetal brain sections and show a positive correlation with neurologic outcomes.

As illustrated in this case, a detailed anatomical assessment of the fetal brain using the MRI based scoring system along with MRI tractography offers greater insights into fetal brain anatomy and the brain neuronal networks. Appropriate fetal MRI and MR tractography use in fetuses with ultrasound diagnosis of isolated ACC provides an additional dimension in anatomical assessment and neurodevelopmental prognostication as early as 22 weeks of gestation. This will likely help in more effective parental counseling and may allow appropriate and timely clinical decisions, as illustrated in this case.
Conflict of Interest
None declared.

References