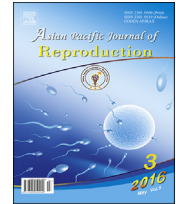


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Corpus callosum agenesis: Role of fetal magnetic resonance imaging

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ABSTRACT

Corpus callosum agenesis (CCA) was evaluated by ultrasound examination and magnetic resonance imaging (MRI) with many studies. Ultrasonography was able to suspect CCA by indirect signs but a definitive diagnosis of CCA was achieved in rare cases. MRI was able to diagnose complete CCA in majority of cases. Additional neurological abnormalities including heterotopia, gyration anomaly, asymmetry of the cerebral hemispheres, and Dandy-Walker variant were documented, as well as an ocular anomaly which was described, by MRI examination. Prenatal counseling for fetal agenesis of the corpus callosum is difficult as the prognosis is uncertain. The association with other cerebral abnormalities increases the likelihood of a poor outcome and ultrasonographic assessment of the fetal brain is limited. We found MRI to be a safe and useful additional procedure to complement ultrasonographic diagnosis or suspicion of CCA.

1. Introduction

The corpus callosum is the most important cerebral commissure allowing axonal fibers to cross the midline. Corpus callosum agenesis (CCA) is an important condition in man that can reveal numerous genetic syndromes. The corpus callosum develops from the commissural plate, a dorsal region of the telencephalon. Then, axons growing from pyramidal neurons of cortical layer III extend and cross the midline.

Predicting CCA remains dubious and debate continues on the issue. The only prognostic indicator which currently can be confirmed with acceptable reliability is whether CCA is isolated or not. Most studies have found that prognosis is generally unfavorable in the event CCA with retarded psychomotor development or epilepsy. The character partial or complete nature of the agenesis does not seem to influence prognosis significantly. Antenatal diagnosis is based on fetal ultrasound, however MRI enables a finer search for associated brain, malformations, in particular neuronal migration anomalies.

2. Case report

We report a case of diagnosis of isolated fetal CCA. No consanguineous parents. We released a fetal MRI who is a specific imaging modality, always performed after a reference ultrasound examination. MR improve prognosis evaluation,

since it enables depiction of no associated abnormalities, notably gyral abnormalities, posterior fossa malformations, and intracranial cysts. In our case MRI was confirmed isolated fetal corpus callosum agenesis (Figure 1). Caesarean section was performed at 38 weeks gestation. A healthy male baby weighing 3400 g was safely delivered.

3. Discussion

The corpus callosum, a forebrain commissure originates from the primitive lamina terminalis. The first callosal fibers form by day 74 of gestation and formation of corpus callosum is complete by 115 d; however the process of myelination continues after birth. The extent of malformation varies from partial to complete agenesis. In partial agenesis, the posterior part is not developed because the corpus callosum develops in an antero posterior direction. However cases of anterior agenesis have also been reported [1]. In CCA, the lateral ventricles are shifted laterally and the 3rd ventricle is enlarged and displaced superiorly with its roof extending dorsally. CCA could be an isolated anomaly or associated with abnormalities in other parts of the brain like heterotopias, microgyria, abnormal cerebral fissures, porencephalic cysts and hydrocephalus.

Callosal agenesis has been reported by several authors. Grogono reported 0.7% of CCA in a large series [2] while Jeret *et al.* came across 33 cases of CCA in a series of 1447 CT scans of brain [3]. Epidemiological surveys have approximately estimated the prevalence of CCA as 1 in 20000 individuals [4].

The etiology is multifactorial but in most of the cases, the cause is unknown. Genetic factors, toxic effects of alcohol on the fetus, endogenous toxins leading to lactic acidosis, vascular

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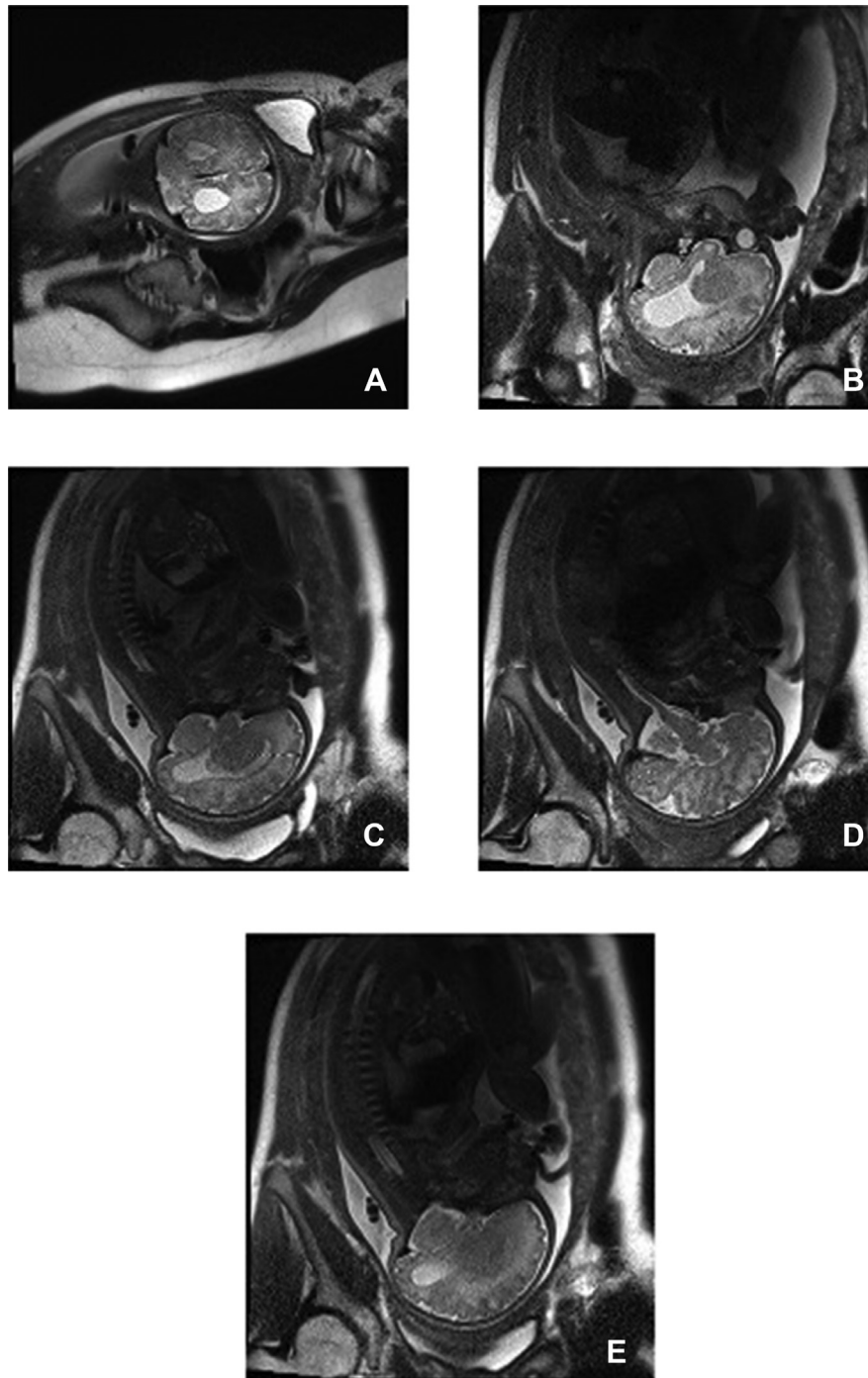


Figure 1. Isolated fetal corpus callosum agenesis.

and metabolic defects and various congenital infections have been found to be closely associated with the etiology of CCA [5].

Various chromosomal defects including trisomies 18, 13 and 8 have been reported in medical literature with CCA. Serur *et al.* [6] reviewed 81 cases of CCA from medical literature, of which 21 had trisomy 8, 14 had trisomy 13 and 18 had aberrations of chromosomes 17 and 18.

Antenatal diagnosis is possible from 20 weeks of gestation. A decision to terminate is difficult to take without reservation until more is known about the incidence of asymptomatic cases. Blum *et al.* [7] reported that six of twelve infants with antenatally diagnosed callosal agenesis had a normal development at 2–8 years of age. Infants with associated manifestations or

chromosomal anomalies had a poor outcome. Fetal karyotyping, looking for associated anomalies and thorough antenatal sonographic examination of the fetus may be of immense help in deciding to terminate pregnancy.

The diagnosis of CCA rests on neuroimaging; cranial ultrasonography in infancy and MRI of the brain in later years. The latter technique is far superior for the diagnosis of partial agenesis, while cranial ultrasonography serves as an easy alternative for the diagnosis of complete agenesis [8]. In our experience, the radiological diagnosis of CCA has been mostly coincidental, hence neuroradiological imaging remains an absolutely essential investigation of developmental delay [5].

We found MRI to be a safe and useful additional procedure to complement ultrasonographic diagnosis or suspicion of CCA.

Prenatal MRI is a valuable complementary technique for either diagnosis of corpus callosum agenesis and depiction of associated neurologic abnormalities. In light of results and data reported in the literature, MRI appears to be an essential and reliable complement to fetal ultrasound in the event of suspected complete or partial agenesis of the corpus callosum and to search for associated malformations. Antenatal counseling in the event of diagnosis of agenesis of the corpus callosum is difficult because the individual prognosis remains poor.

Conflict of interest statement

The authors declare no conflict of interests.

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