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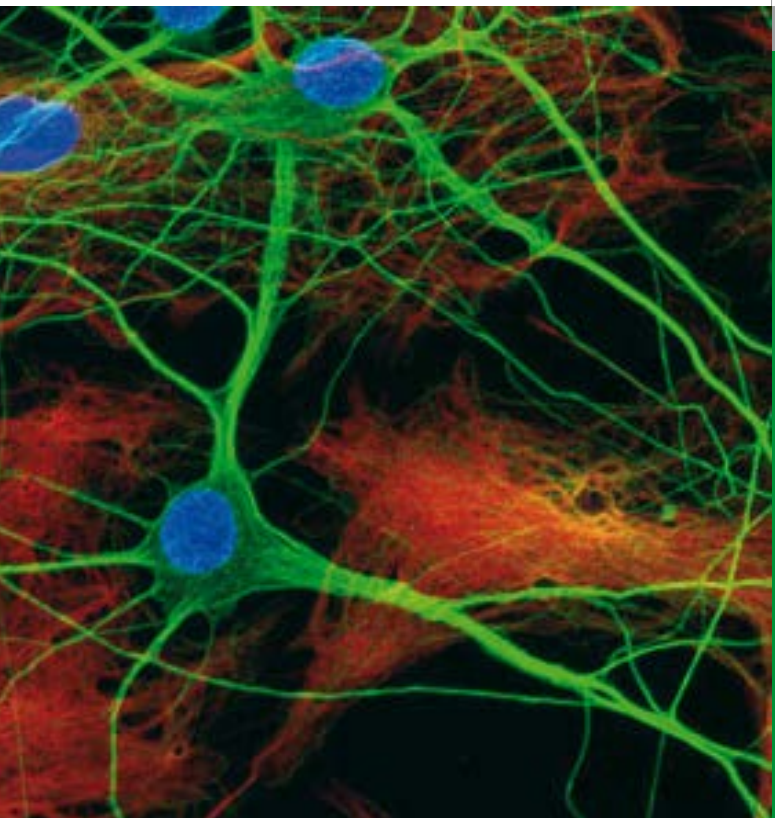
# Fetal brain imaging



April 2008

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# INTRODUCTION

Ultrasound imaging (US) is the screening modality of choice for the detection of fetal abnormalities. It has many advantages over other imaging modalities in that images have high anatomical resolution and are performed in "real time", allowing dynamic assessment of the fetus. In addition, ultrasound is widely available at relatively a low cost compared with other imaging modalities. Its accuracy is however operator dependent. Technical advances in the field of magnetic resonance imaging (MRI) throughout the past 2 decades have influenced its use in fetal imaging. The introduction of fast imaging techniques has ameliorated the problem of image artefacts caused by fetal movement. The use of MRI for diagnosing central nervous system (CNS) abnormalities has been widely reported with many authors agreeing that fetal MRI in the second and third trimesters can demonstrate additional findings, which may influence the diagnosis and case management and could thus be useful in counselling patients (1-5).

We present a case where fetal MRI added further information and changed the clinical management of the patient.

## CASE REPORT

A 19-year-old primigravida woman was referred at 21 weeks gestation to the fetal medicine unit (FMU) at University College London Hospital. Booking bloods were all normal. The 20-week fetal anomaly scan in the referral hospital suggested microcephaly and oligohydramnios; the head circumference was on the 3rd centile while all other body measurements were within normal limits. Doppler flow measurements in both uterine and umbilical arteries were normal. Suboptimal views of the brain were obtained due to fetal lie and oligohydramnios on subsequent serial scanning, however fetal growth was good. At 26 weeks gestation, good views of the fetal brain were acquired and an echolucent area near the anterior horn of the right lateral ventricle was seen. The patient was then referred to the FMU neuroclinic at UCLH for more detailed brain scans.

The fetal neuroclinic at UCLH consists of a multidisciplinary team of neonatologists with special interest in brain development, a geneticist and fetal medicine specialists. Both transabdominal and transvaginal ultrasound scans showed microcephaly and a large cleft in the right hemisphere (Fig. 1). This cleft was thought to be either a porencephalic cyst, arachnoid cyst or unilateral schizencephaly. Maternal TORCH and antiplatelet antibody screen were negative. It was decided to perform a fetal MRI to elucidate the brain abnormality since prognosis of those abnormalities varied in severity.

A fetal MRI was performed at 27 weeks gestation using a 1.5T Siemens system using a phased-array wrap-around body coil. T2 weighted, single shot, fast-spin echo MRI images were acquired in the coronal, axial and sagittal planes with respect to the fetal brain. The acquisition time is approximately 10 to 20 minutes depending on the fetal movements, and no sedation is required.

On MRI bilateral schizencephaly was clearly seen (Fig. 2-5). MRI diagnosis of a bilateral schizencephaly influenced the parental counselling in that a poor prognosis was given. The parents chose to terminate the pregnancy. Parents declined any postmortem examination.

Schizencephaly is a malformation due to abnormal cortical organisation and late migration. There is a complete agenesis of a portion of germinal matrix, leaving seams or clefts spanning the cerebral hemisphere from pial surface to the lateral ventricles lined by cortical gray matter. The lips of the clefts can become widely separated and they tend to be in the regions of the Rolandic and Sylvian fissures, and involve predominantly frontal areas.

The etiology of schizencephaly remains unclear. The main question is whether schizencephaly results from abnormal brain development or from destruction of already formed brain structures. Familial cases of

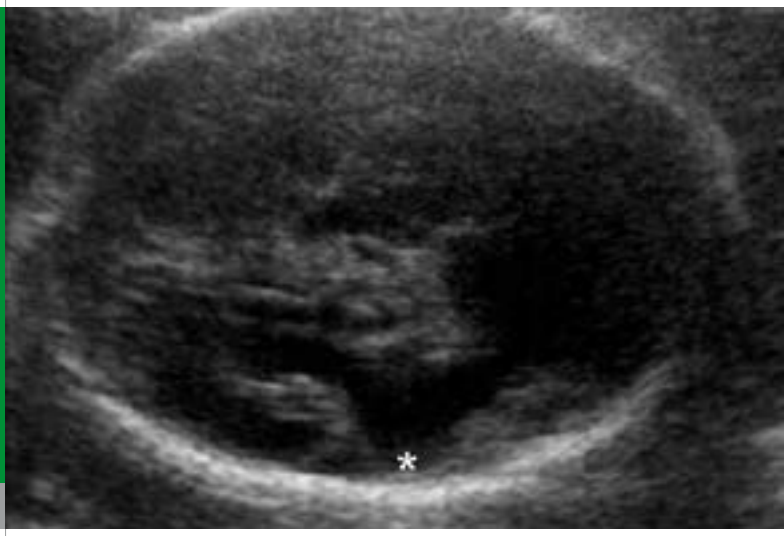
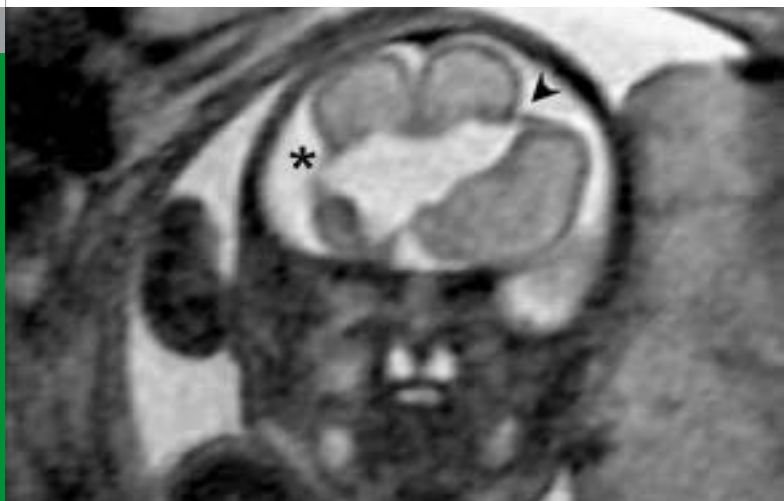


Fig. 1

Ultrasound image showing unilateral cleft in the right hemisphere at 26 weeks.

Coronal MR image at 27 weeks of gestation showing bilateral schizencephaly, open-lipped on the right side (asterisk) and close-lipped on the left side (arrow head).

Fig. 2



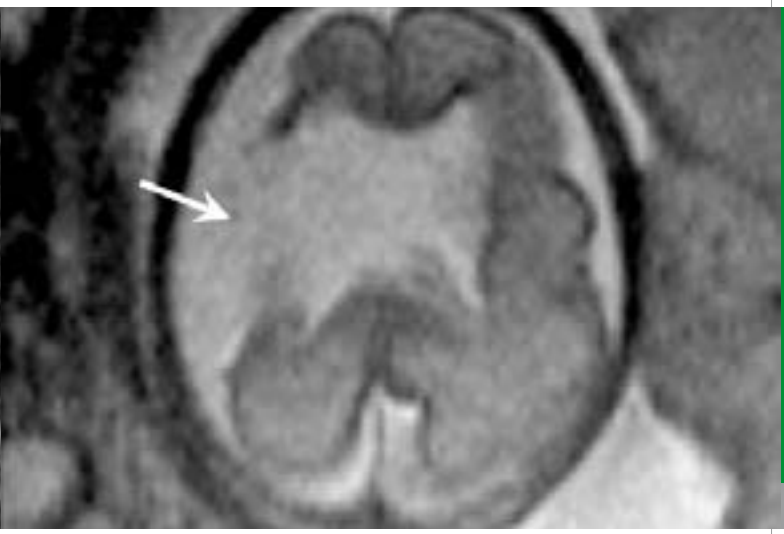


Fig. 3

Axial MR image the open schizencephalic cleft on the right side.

Coronal MR image the open schizencephalic cleft on the right side.

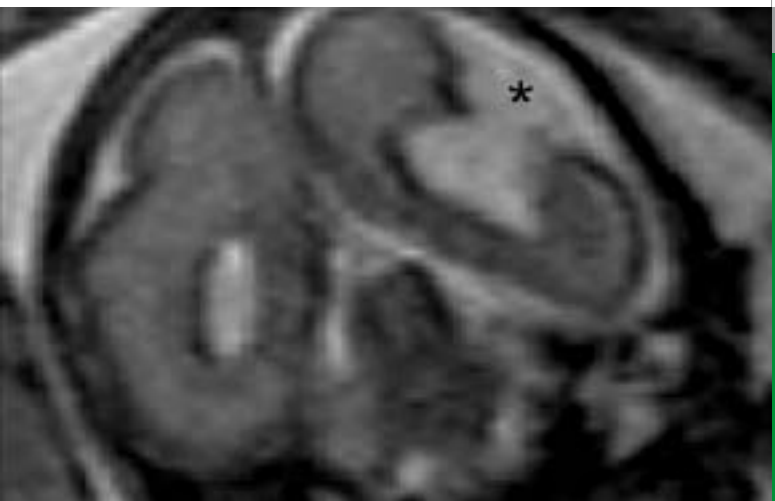


Fig. 4

schizencephaly have been reported, suggesting a possible genetic origin within the group of migrational disorders. Brunelli et al have reported heterozygous mutations of the homeobox gene, EMX2, associated with schizencephaly (6). However, a multifactorial origin of schizencephaly is supported by the association with cytomegalovirus infection (7, 8).

The most frequently presenting neonatal sign is asymmetrical muscle tone in the unilateral form, or childhood developmental delay when both hemispheres are involved (9). Seizures can be the first sign of the abnormality. Epilepsy is noted in 50-80% of cases of schizencephaly, with onset before the age of 3 years in 81% of cases (10, 11). A neurological deficit was detected in almost half of the cases within the first year of life (9). The clinical (motor and cognitive outcome) features of schizencephaly are extremely variable and their severity is closely related to the size and bilaterality of the cleft. Children with unilateral schizencephaly often present with hemiparesis and mild mental delay whereas children with bilateral cleft are usually tetraplegic with severe mental deficits (9, 11). The major prognostic factor is the bilaterality of the cleft and the location of the cleft; hence, optimal antenatal imaging must be performed.

In this case due to fetal lie and oligohydramnios it was very difficult to obtain satisfactory views of the brain. Fetal MRI clearly showed the abnormality and this

additional information changed the management of this patient. Although MRI can be performed in the early stage of the second half of gestation, identifying abnormalities of cortical development is not easy, especially in fetuses at 18 to 25 weeks as the normal development of the cortex and neuronal migration is occurring at this time. Indeed, Fogliarini et al have suggested that as the pattern of cortical abnormalities seen on MRI may be different at different gestational ages, it may be necessary to repeat an MRI examination at a later stage to ascertain the diagnosis (12).

Fetal MRI is also very useful in diagnosing brain abnormalities that are associated with schizencephaly such as agenesis or dysgenesis of the corpus callosum, absence of the septum pellucidum and arachnoid cysts. Furthermore, fetal MR can help to exclude other differential diagnoses of CSF-containing brain abnormalities such as porencephalic cysts, hydrancephaly, severe ventriculomegaly, arachnoid cysts and agenesis of the corpus callosum with large interhemispheric cysts.

In summary, prenatal imaging allows detection and characterisation of migrational disorders such as schizencephaly. Detection of bilaterality of this lesions and associated abnormalities is important for counselling the parents. MRI can aid in distinguishing schizencephaly from other differential diagnoses and has the additional benefit of delineating other associated brain abnormalities.

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