Prenatal Diagnosis of Polymicrogyria by Fetal Magnetic Resonance Imaging in Monochorionic Cotwin Death

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Fetal magnetic resonance imaging (MRI) is being used increasingly for confirmation and further characterization of brain abnormalities detected on routine prenatal sonography. It offers improved contrast resolution and direct visualization of both sides of the developing brain and is not susceptible to the limitations of sonography such as fetal position and amniotic fluid volume. With fetal MRI, more sensitive imaging of the cortical and sulcal development of the fetus can be obtained. Although most brain abnormalities occurring in cases of complicated monochorionic twin pregnancies are characterized by localized or diffuse areas of parenchymal destruction, cortical abnormalities have also been described postnatally or on fetal autopsy.8–21 We report a case in which fetal MRI showed not only encephalomalacia but also associated polymicrogyria in a survivor of a monochorionic intrauterine cotwin death.

Case Report

A 32-year-old woman, gravida 4, para 3, was referred for fetal MRI because of the fetal death of 1 twin of a monochorionic, diamniotic pregnancy. An initial ultrasound examination performed at 15 weeks 1 day showed living twins of a monochorionic, diamniotic pregnancy. The fetal size and amniotic fluid volume were concordant, and there was no hydrops in either twin. A routine diagnostic ultrasound examination performed at 18 weeks 5 days, showed the death of 1 twin whose gestational age was calculated to be 15 weeks 5 days and who had severe oligohydramnios. The fetal size and amniotic fluid volume were concordant, and there was no hydrops in either twin. A routine diagnostic ultrasound examination performed at 18 weeks 5 days, showed the death of 1 twin whose gestational age was calculated to be 15 weeks 5 days and who had severe oligohydramnios. The gestational age of the surviving twin was calculated to be 18 weeks 2 days; this twin had mild polyhydramnios. The diagnosis of twin-twin transfusion syndrome was not made previously. The surviving twin had a heart rate of 151 beats per minute, normal rhythm, normal heart size, and a normal umbilical cord Doppler waveform. The patient returned for follow-up sonography at 20 weeks 4 days, and appropriate interval

Abbreviations

MRI, magnetic resonance imaging
growth as well as resolution of the mild polyhy-
dramnios for the surviving twin were document-
ed. Another ultrasound examination performed at 22 weeks 6 days showed a slight interval
decrease in head growth (head circumference
calculated to be 21 weeks 1 day); however, the
brain anatomy, including the visualized “down-
side” left cerebral ventricle, was normal (Figure
1), and amniotic fluid volume was normal.
Amniocentesis was performed and showed a nor-
mal karyotype, 46,XY. Because of the death of the
cotwin, the concern for potential brain damage to
the survivor of a monochorionic twin death, and
the slight decrease in head growth, the patient
was referred to us for further evaluation with MRI.
Magnetic resonance imaging performed at 23
weeks 3 days, showed a large area of encephalo-
malacia involving the left parietal and frontal
lobes (Figure 2). There were multiple abnormal
infoldings of the overlying developing cortex in
the left frontal and parietal lobes consistent with
polymicrogyria. The ipsilateral thalamus was
also slightly small, possibly because of decreased
connections between the left frontal and parietal
lobes and the thalamus. Postnatal MRI per-
formed at 5 months showed extensive polymicr-
ogyria involving the left frontal and parietal lobes
(Figure 3). Clinically, the child, who was 2 years
old at the time of this writing, has a congenital
right hemiparesis as well as a gross motor delay.
He sat at 11 to 12 months, crawled at 15 months,
and began walking with holding on at 18 months.
Seizures developed at 20 months of age.

Discussion

Improved prenatal detection of malformations
of cortical development is now possible with
prenatal MRI. Although sonography can show
abnormalities in ventricular size and morpho-
logic features, as well as abnormalities of the
corpus callosum and posterior fossa, it is limited
in its ability to visualize the developing cortex
and therefore to show cortical malformations.22
In contrast, fetal MRI has been shown to depict
malformations of cortical development that are
sonographically occult, including polymicrogy-
ria.23–27 To our knowledge, our case is the first
report of prenatally diagnosed polymicrogyria in
a survivor of monochorionic cotwin death.
Multiple brain abnormalities have been
described in complicated monochorionic twin
pregnancies.8–21 Most of the brain abnormalities
occurring in such cases involve localized or diffuse areas of parenchymal destruction, such as porencephaly, multicystic encephalomalacia, and periventricular white matter injury,\textsuperscript{11,17,21} and are most likely the result of hypoperfusion and resultant hypoxia-ischemia.\textsuperscript{11} In cotwin death, it has been postulated that the intrauterine death of 1 twin leads to acute hemodynamic changes in the surviving fetus, resulting in hypoxic-ischemic lesions.\textsuperscript{11,12,14,15,28}

**Figure 2.** Images from fetal MRI performed at 23 weeks 3 days. \textbf{A}, Axial single-shot fast spin echo T2-weighted image above the level of the lateral ventricles shows a large area of encephalomalacia involving the left parietal and frontal lobes (arrow). \textbf{B}, Coronal single-shot fast spin echo T2-weighted image through the frontal lobes shows a diminished volume of the left frontal lobe with several abnormal cortical infoldings (arrow) consistent with polymicrogyria.

**Figure 3.** Images from postnatal MRI performed at 5 months of age. \textbf{A}, Axial dual-echo T2-weighted image shows a diminished volume of the left frontal and parietal lobes with an abnormal sulcal pattern. An abnormally deep sulcus is also identified (arrow). \textbf{B}, Coronal fast spin echo T2-weighted image shows abnormal thickening of the perisylvian cortex consistent with polymicrogyria.
Polymicrogyria has also been described in monochorionic twin pregnancies, usually associated with intrauterine cotwin death and twin-twin transfusion syndrome. Polymicrogyria is a malformation of cortical development and is characterized by abnormal cortical organization and numerous small gyri. Barth and van der Harten reported bilateral posterior polymicrogyria on autopsy in a term neonate in the setting of twin-twin transfusion syndrome with intrauterine cotwin death at 13 to 16 weeks. Larroche et al described 3 fetuses with twin-twin transfusion syndrome, 2 of whom also had cotwin death, with polymicrogyria on autopsy. Baker et al reported a case of bilateral parietal polymicrogyria and arthrogryposis multiplex congenita in a monochorionic twin pregnancy complicated by intrauterine cotwin death during the first trimester. Congenital bilateral perisylvian polymicrogyria was reported in a monochorionic twin pregnancy complicated by intrauterine cotwin death.

Polymicrogyria can result from perfusion abnormalities, intrauterine infection, toxins, and genetic causes. There are several reports of polymicrogyria occurring in singleton pregnancies complicated by maternal hypotension, severe maternal trauma, and maternal ingestion of ergotamine, all presumably resulting in impaired fetal perfusion. Changes in fetal perfusion are also the most likely cause of polymicrogyria in complications of monochorionic twin pregnancies, including this case. Indeed, in their study of 5 pairs of fetuses with twin-twin transfusion syndrome, Larroche et al found evidence of hypoxic-ischemic lesions in all 3 fetuses in which they observed layered polymicrogyria; 2 of the fetuses were also survivors of intrauterine cotwin death. Bordarier and Robain also reported necrotic lesions of the white matter and cortex in 2 fetuses with polymicrogyria. Similarly, the large area of encephalomalacia associated with the development of the polymicrogyria in this case is also indicative of parenchymal injury.

Polymicrogyria can be associated with developmental delay, cognitive impairment, epilepsy, and focal neurologic deficits. The clinical symptoms depend, in part, on the location and extent of the polymicrogyria. Patients with unilateral polymicrogyria involving the perioral cortex (as in our patient) tend to have congenital hemiparesis as well as epilepsy.

Survivors of intrauterine cotwin death have a 20% risk of cerebral impairment, and this risk is higher in monochorionic twin pregnancies. In a review of the literature, neurologic abnormalities were reported in 72% of survivors of monochorionic cotwin death. The risk of cerebral palsy is also greater in survivors of cotwin death compared with other twin and singleton pregnancies. Our patient had congenital hemiparesis, which is considered a type of cerebral palsy.

In summary, we report a case of monochorionic twin pregnancy with cotwin death in which fetal MRI showed polymicrogyria in the surviving twin that was presumably due to hemodynamic impairment related to the death of the cotwin. The only prenatal sonographic abnormality was mild delay in head growth compared with a prior sonogram, which prompted the fetal MRI at 23 weeks 3 days. Because fetal MRI can show sonographically occult cortical malformations such as polymicrogyria, it should be strongly considered in complicated monochorionic twin pregnancies.

References


