MODERN prenatal imaging has improved the accuracy of the prenatal diagnosis. This has led to a greater need for interdisciplinary cooperation to provide better parent counseling and management of disorders in the fetus.

A multidisciplinary approach plays an important role in counseling expectant parents about fetal CNS malformations. Initially, the interaction of obstetricians, other specialist(s), and parents may begin in the prenatal period to plan the timing and route of delivery. Postnatal management, including surgery, may be discussed in advance. Familiarity with imaging modalities will permit a better presentation of information for family counseling and decision making during and after the pregnancy.

Although ultrasonography has been the imaging method most commonly used for evaluating a fetus, MR imaging has proved to be a valuable complementary tool; it provides good anatomical and tissue definition for further characterization of an abnormality. Magnetic resonance imaging may also be beneficial in circumstances in which ultrasonography is technically limited.1-5

The purpose of this study was to determine the value prenatal MR imaging holds as a complementary imaging modality to ultrasonography in deciding whether and what postnatal neurosurgical intervention is required.

Clinical Material and Methods

Between October 1999 and March 2003, 320 prenatal MR imaging studies were performed at our institution. These studies were performed after a suspected abnormality had been noted during obstetrical ultrasonographic examinations. Anomalies of the CNS were suspected, with or without involvement of other organs, after reviewing MR images in 249 fetuses (77.8%). Of those fetuses with CNS abnormalities, 24 were judged to be likely candidates for postnatal neurosurgical intervention, and these constitute the study group. A fetus was enrolled in this study if the abnormality demonstrated on MR images indicated a condition that would have a high likelihood of being treated by a pediatric neurosurgeon. The study therefore did not include fetuses with stable, mild ventriculomegaly or malformative brain disorders.

The mean gestational age of the study group at the time of the initial MR imaging session was 29 weeks (range 21–38 weeks, median 29 weeks). The diagnosis included spinal...
abnormalities in 12 fetuses and brain abnormalities in the other 12 fetuses (Tables 1 and 2).

Magnetic resonance imaging was performed using a 1.5-tesla imaging unit (General Electric Medical Systems, Milwaukee, WI). After a localizing gradient-echo sequence had been performed, ultra-fast T_{2}-weighted single-shot fast spin echo MR images of the fetus were obtained in the axial, coronal, and sagittal planes, using the following parameters: TR 52 msec, TE 90 msec, number of excitations 0.5, bandwidth 32 kHz, field of view 16 \times 28 \text{ cm}, matrix 256 \times 192, section thickness 3 to 5 mm, and gap 0 to 1 mm. A torso phased-array coil was usually used unless the fetus was too large to fit, in which case a body coil was substituted.

Two experienced pediatric radiologists reviewed all MR images. The diagnosis based on the MR imaging findings was confirmed in all cases by findings on postnatal images or those at operation or autopsy.

This patient group was followed up by a pediatric neurosurgery team between 6 and 36 months after birth.

Results

Fourteen fetuses (58.3%) required surgical intervention in the early postnatal period. Spinal surgery was performed in three babies with closed spinal dysraphism and in one with open spina bifida (a myelomeningocele). Of the three infants with closed spinal dysraphism who underwent surgery, one presented with a tethered cord associated with diastematomyelia, another with a spinal tail and a normal cord, and a third with a dermal sinus.

Ten of the infants underwent cranial surgery: three for shunt implantation, two for intraventricular cyst reduction via craniotomy and endoscopy, three for arachnoid cyst removal, one for anterior cranial remodeling, and one for closure of an encephalocele. Conservative follow up is still in effect for three of the patients, two with mild scoliosis and segmentation vertebral anomalies and one with ventriculomegaly and parenchymal hemorrhage.

Termination of pregnancy was selected in seven cases. The MR imaging findings in these fetuses were confirmed at autopsy (Tables 1 and 2).

Ventriculomegaly

In our series, four (33.3%) of 12 fetuses with brain anomalies had significantly enlarged (> 15 mm) ventricles. In two of them there was aqueductal stenosis and intraventricular hemorrhage (Fig. 1). Both patients displayed clinical signs in the early postnatal period and had to undergo VP shunt insertion. One patient had aqueductal stenosis only; this infant was initially treated with ETV, but later underwent VP shunt implantation to alleviate absorptive hydrocephalus. The fourth fetus had a prenatal left parenchymal hemorrhage and a porencephalic cyst. At the postnatal follow-up examination, this child demonstrated right hemiparesis and is now being treated conservatively. Interestingly, in one patient the findings of prenatal MR imaging led to a diagnosis of mild ventriculomegaly secondary to a posterior fossa retrocerebellar cyst, but after birth the patient demonstrated clinical signs of progressive hydrocephalus. This baby underwent posterior fossa decompression and opening of the large arachnoid cyst to the subarachnoid spaces, with an eventual decrease in the size of the ventricles and normalization of head circumference and development.

Intracranial Cysts

Five of the patients in our series harbored intracranial cysts (Fig. 2). The prenatal diagnoses of these cysts was made when the patients were between 30 and 38 weeks of gestation (Table 2). There were two intraventricular, two supratentorial, and one posterior fossa arachnoid cysts. The postnatal clinical signs of elevated intracranial pressure and/or enlargement of head circumference, and the finding of cyst growth on repeated ultrasonographic examinations of the brain were viewed as indications of the need for surgical intervention in all infants. As mentioned earlier, one patient underwent surgery for cyst removal because of progressive ventriculomegaly. The ages at surgery in this group

<table>
<thead>
<tr>
<th>Case No.</th>
<th>GA (wks)</th>
<th>Prenatal MRI Diagnosis</th>
<th>Treatment</th>
<th>Outcome (age)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>25</td>
<td>severe lumbar segmental formation anomaly</td>
<td>TOP</td>
<td>NA</td>
</tr>
<tr>
<td>2</td>
<td>30</td>
<td>MMC, agenesis corpus callosum</td>
<td>TOP</td>
<td>MMC confirmed</td>
</tr>
<tr>
<td>3</td>
<td>24</td>
<td>spinal tail</td>
<td>spinal tail excision</td>
<td>ND (3.3 yrs)</td>
</tr>
<tr>
<td>4</td>
<td>17</td>
<td>MMC</td>
<td>TOP</td>
<td>NA</td>
</tr>
<tr>
<td>5</td>
<td>34</td>
<td>scoliosis</td>
<td>conservative</td>
<td>ND</td>
</tr>
<tr>
<td>6</td>
<td>29</td>
<td>complex closed spinal defect</td>
<td>TOP</td>
<td>male, spina bifida &amp; diastematomyelia</td>
</tr>
<tr>
<td>7</td>
<td>25</td>
<td>DMS w/ tethered cord</td>
<td>excision of osseous spur,untethering of thoracic cord</td>
<td>ND, scoliosis (1.4 yrs)</td>
</tr>
<tr>
<td>8</td>
<td>21</td>
<td>sacrococcyeal teratoma</td>
<td>TOP</td>
<td>female, Grade II immature teratoma, sacrococcygeal</td>
</tr>
<tr>
<td>9</td>
<td>29</td>
<td>MMC</td>
<td>closure of MMC</td>
<td>no control of sphincters &amp; no walking (2 yrs)</td>
</tr>
<tr>
<td>10</td>
<td>32</td>
<td>multiple segmentation anomalies, normal cord</td>
<td>conservative</td>
<td>ND, MRI-demonstrated butterfly vertebra (5 mos)</td>
</tr>
<tr>
<td>11</td>
<td>26</td>
<td>DMS &amp; L2–3 conus</td>
<td>TOP</td>
<td>NA</td>
</tr>
<tr>
<td>12</td>
<td>21</td>
<td>tethered cord</td>
<td>untethering of cord</td>
<td>ND (7 mos)</td>
</tr>
</tbody>
</table>

* DMS = diastematomyelia; GA = gestational age; MMC = myelomeningocele; NA = not available; ND = normal development; TOP = termination of pregnancy.
were 1 day and 1 month for the two patients with intraventricular cysts, 6 weeks for the patient with a posterior fossa cyst, and 1 month and 27 months for the two patients with supratentorial cysts.

Other Intracranial Anomalies

The fetus with craniosynostosis underwent prenatal ultrasonographic evaluations at 23, 25, and 27 weeks of gestational age. These studies demonstrated a lemon-shaped head and frontal bossing. The typical signs of metopic craniosynostosis were observed.

Two fetuses in which an encephalocele was suspected following prenatal ultrasonography were included in the present patient population. In one fetus, ventriculomegaly was demonstrated and there was evidence suggestive of the presence of a midline skull abnormality; in this case the parents opted for termination of the pregnancy. The autopsy disclosed a large tongue, hypertelorism, and a probable dysplasia of the face. Partial untethering was achieved in both together with removal of the primary lesion. Neither infant had a neurological deficit before surgery, and no neurological deficit was documented during the postoperative follow-up period, before they reached the age of 2 months. Successful untethering of the cord was achieved in both together with removal of the primary lesion. Neither infant had a neurological deficit before surgery, and no neurological deficit was documented during the postoperative follow-up period (≤ 2 years). The infant with craniosynostosis had mild scoliosis at birth and continues to receive orthopedic follow up; to date there has been no deterioration in the scoliosis.

Spinal Abnormalities

Spinal abnormalities were seen in 12 of the 24 fetuses. Of the three myelomeningoceles identified, one was suspected but not diagnosed on the basis of the ultrasonographic study because the fetal position posed a difficulty to imaging of the lumbar spine. The prenatal MR images demonstrated the lumbar spinal dysraphism with a Chiari malformation Type II and raised the suspicion of partial agenesis of the corpus callosum and malformation of the lateral ventricles.

The only fetus harboring a myelomeningocele that was not aborted underwent surgery within the first 48 hours after delivery. Five (46.1%) of the 12 fetuses with spinal anomalies presented with a closed spinal defect. Three of these patients had diastematomyelia, one had a spinal tail, and one had a dural sinus with a tethered cord (Fig. 4). Two of the fetuses, one with diastematomyelia and the other with a dural sinus, underwent surgery during the early postnatal period, before they reached the age of 2 months. Successful untethering of the cord was achieved in both together with removal of the primary lesion. Neither infant had a neurological deficit before surgery, and no neurological deficit was documented during the postoperative follow-up period (≤ 2 years). The infant with diastematomyelia had mild scoliosis at birth and continues to receive orthopedic follow up; to date there has been no deterioration in the scoliosis.

Discussion

Prenatal MR imaging is an important complementary examination to routine prenatal ultrasonography. The MR images provide a better demonstration of the anatomy when a pathological condition is suspected and enable better tissue characterization. Imaging can be obtained in multiple
planes (axial, sagittal, and coronal) and no ionizing radiation is employed. Other advantages of performing prenatal MR imaging include the ability to evaluate the pattern of ventricular abnormalities (obstructive versus communicating ventriculomegaly) and to demonstrate gyral and parenchymal maldevelopment.

Fig. 1. Sagittal (upper) and coronal (lower) single-shot fast spin echo T2-weighted MR images of the brain obtained in a 35-week-old fetus with aqueductal stenosis. Severe dilation of the lateral ventricles and moderate dilation of the third ventricle are apparent. The posterior fossa and the fourth ventricle are not enlarged. No further anomalies of the brain were identified in this case. After birth the patient was treated with ETV and later a VP shunt was implanted.

Fig. 2. Upper: Axial T2-weighted MR image of the brain obtained in a 30-week-old fetus, showing a supratentorial cyst that was suspected to be a subarachnoid cyst. Lower: Postnatal coronal ultrasonographic image obtained when the patient was 1 month old demonstrating enlargement of the cyst. At surgery, shrinkage and marsupialization of the intraventricular cyst were performed.
In the spine, prenatal ultrasonography is a good screening method for the evaluation of osseous elements. Nevertheless, prenatal MR imaging is superior for the characterization of intraspinal structures, providing clear visualization of the cord and the cauda equina segment.

Prenatal Ventriculomegaly

The most commonly encountered fetal abnormality of the brain is enlarged ventricles, which may represent a variety of conditions. Efforts to determine the causes of ventriculomegaly by performing imaging studies, chromosomal analysis, or viral screening are important for the evaluation of fetal outcome.

Ventriculomegaly is defined as mild (atrial diameter 10–15 mm), moderate (atrial diameter > 15 mm, with a residual cortex > 2 mm in diameter), or severe (residual cortex < 2 mm in diameter).

Although ultrasonography is commonly used for the detection of fetal anomalies, MR imaging aids in the confirmation of ventriculomegaly and detects associated anomalies such as aqueductal stenosis, agenesis of the corpus callosum, migration anomalies, or late presentation of a Chiari malformation Type II. For example, in cases of tri-ventricular hydrocephalus in which the patient may be a potential candidate for ETV, fetal MR imaging can support the diagnosis of aqueductal stenosis. Differentiation between a good and bad candidate for ETV is based on the shape of the floor of the third ventricle together with the presence of an abnormality of the aqueduct or midbrain. These can be well demonstrated on midsagittal MR images.

Early delivery for surgical intervention may be indicated when progressive enlargement of the ventricles is noticed; however, termination of the pregnancy may be an option if a diagnosis of severe parenchymal abnormality is valid. Prenatal counseling for the expectant parents may thus include not only better prognostication by means of understanding the underlying pathological condition and its possible association with other malformations (which may not be seen on ultrasonography), but also treatment options such as the choice of an endoscopic procedure or shunt implantation surgery.

In cases of intraventricular hemorrhage, the advantage of prenatal MR imaging may be in the characterization of tissue damage. The size and presence of a blood clot are easily detected using ultrasonography, but hyperechogenicity may sometimes mask the parenchymal damage. The combination of fast spin echo T2-weighted and gradient-echo T1-weighted sequences can help in the detection and better characterization of the timing and grade of the hemorrhage.

Other Intracranial Pathological Conditions

The differentiation between arachnoid cysts and malformative brain cysts may sometimes be difficult using ultrasonography, especially if the cyst is large and causes a significant distortion of the brain. However, this differentiation is important because the prognosis for intracranial arachnoid cysts may be good, whereas the prognosis for malformative brain cysts may be associated with developmental delay, seizures, and hydrocephalus. Prenatal MR imaging can demonstrate the location of the cyst, whether intra- or extraventricular, supra- or infratentorial, and adjacent to cisterns or intraparenchymal. In the past, management of
an intracranial cyst was confined to craniotomy or shunt implantation. The option of endoscopic surgery may be mentioned, especially if the arachnoid cysts can be opened to a ventricle or cistern.9

Posterior fossa cysts are a special challenge for the obstetrician as well as for the fetal radiologist. The differential diagnosis includes severe malformations such as the Dandy–Walker malformation and the Dandy–Walker variant, as well as items associated with a very good prognosis such as arachnoid cyst. Prenatal counseling will enable decisionmaking based on detailed information made available by MR imaging.

Magnetic resonance imaging is very important in the detection of associated brain malformative disorders together with what appears to be a simple arachnoid or intraventricular cyst on ultrasonography. For example, schizencephaly or semilobar holoprosencephaly sometimes may be underdiagnosed on the basis of ultrasonographic findings alone. The combination of ultrasonography and MR imaging enables a more accurate diagnosis and assessment of the prognosis.

When pathological conditions are suspected in the calvaria, MR imaging may provide information about brain development as well as an assessment of cranial deformity. The multiplanar views afforded by fetal MR imaging can help the clinician evaluate the calvarial shape in relation to the sutures and secondary signs of various craniosynostoses,1 whereas an ultrasonographic evaluation of the calvaria may be difficult due to attenuation of sound waves by the skull and the cranial curvature.

For a prenatal diagnosis of metopic synostosis, MR imaging proved to be important in establishing the diagnosis of suture synostosis and in excluding associated cerebral malformations. As for neural tube defects involving the brain with suspected encephalocele, findings of ultrasonography may underestimate the pathological condition, especially with respect to a problematic location such as the craniocervical region or a multiple fetus pregnancy. In addition, encephaloceles may be mistaken for an extracranial mass with a variability of cystic or solid components.4

Spinal Dysraphism

Spinal dysraphism and myelomeningocele are usually diagnosed accurately based on findings of ultrasonography. The contribution of fetal MR imaging lies in its demonstration of associated brain anomalies such as agenesis of the corpus callosum or migration malformation. Presence of a Chiari malformation Type II and location of an open defect may be better estimated on the basis of MR imaging findings if ultrasonography is technically difficult. In centers in which fetal surgery is undertaken to treat myelomeningoceles, MR imaging must be performed to evaluate the lesion in detail, to provide the inclusion criteria for fetal surgery, and to analyze the postoperative outcome. These parameters include the level and configuration of the spinal lesion, the degree of hydrocephalus, the degree of hindbrain herniation, and the presence or absence of other intracranial anomalies.11–15

Small meningoceles or even myelomeningoceles can be mistaken for closed spina bifida with lipomyelocele. Sattar and colleagues12 reported findings of prenatal ultrasonographic images in eight fetuses with spina bifida. In five the spinal cord was tethered and in three a meningocele was thought to be present; however, in one patient a postnatal MR image revealed a lipomeningocele instead of a simple meningocele.

Closed spina bifida is detected relatively rarely when using prenatal ultrasonography because identification of the level of the conus is not routinely advocated at most centers. Scoliosis or other abnormalities of the spine, including segmentation formation anomalies, may be detected by prenatal ultrasonography, but the associated intraspinal pathological condition may be overlooked. The presence of a tethered cord together with a malformed spine has important consequences for the patient’s postnatal condition; close postnatal neurological, urological, and neurosurgical follow up is advocated in conjunction with pediatric orthopedic surgery.

Early prenatal identification of a spinal dysraphic lesion allows earlier surgical untethering before neurological signs appear. In the past, in many children who underwent surgery for diastematomyelia, an irreversible neurological motor deficit occurred. The prenatal diagnosis of diastematomyelia may allow planning and early surgery, with possible avoidance of clinical deficits. Because prenatal MR imaging allows better demonstration of intraspinal lesions, the need for postnatal imaging requiring sedation in young patients could sometimes be avoided.16,17

Conclusions

In the era of advanced antenatal diagnostic techniques that enable detection of structural abnormalities of the fetal CNS, the neurosurgeon is an active part of the team for prenatal family counseling. Together with prenatal ultrasonography, genetic counseling and fetal MR imaging are important tools for appropriate pre- and postnatal management. Magnetic resonance imaging provides a detailed anatomy of the brain, spinal cord, and ventricles with superior tissue characterization, and it may help detect complex brain malformations.

In selected cases prenatal MR imaging eliminated the need for postnatal MR imaging, which in infants requires induction of anesthesia. In many cases prenatal MR imaging allows early diagnosis and intervention by the neurosurgical team. Fetal MR imaging enables parents to receive detailed information, not only about the expected prognosis of the fetus, but also about the different options of neurosurgical treatment.

References


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Address reprint requests to: Liana Beni-Adani, M.D., Department of Pediatric Neurosurgery, Tel-Aviv Sourasky Medical Center, Weizman 6, Tel-Aviv 64239, Israel. email: lianabenia@gmail.com.