Objectives—The purpose of this study was to describe the characteristics and outcomes of umbilical cord hernias diagnosed prenatally.

Methods—We conducted a retrospective study of all pregnancies with the diagnosis of a fetal umbilical cord hernia during a 5-year period. All women received care from a multidisciplinary team and underwent complete meticulous sonography for structural malformations as well as fetal echocardiography and amniocentesis.

Results—Between 2004 and 2009, isolated fetal umbilical cord hernias were diagnosed in 8 pregnant women. The gestational ages at the time of referral ranged from 16 to 28 weeks (median, 20 weeks). In 1 case, intrauterine fetal death occurred at 35 weeks due to rupture of the umbilical cord. All remaining cases were delivered at 36 to 40 weeks, and the neonates underwent corrective surgery with good outcomes.

Conclusions—Despite 1 case complicated by intrauterine fetal death in this study, the outcome of an isolated fetal umbilical hernia seems favorable.

Key Words—prenatal diagnosis; sonography; umbilical cord hernia
As opposed to omphalocles, which are well-described anomalies, the literature on umbilical cord hernias is limited. The aim of this study was to describe the sonographic characteristics, natural history, and outcomes of fetuses with the diagnosis of an umbilical cord hernia in the first or second trimester.

Materials and Methods

Eight cases of umbilical cord hernias were referred for evaluation at the Chaim Sheba Medical Center over a 5-year period between 2004 and 2009. All women underwent multidisciplinary assessment and care by a team consisting of fetal medicine specialists, geneticists, and pediatric surgeons. Prenatal evaluation included a detailed anomaly scan to rule out associated malformations, fetal echocardiography, and amniocentesis. In cases evaluated during the early second trimester, vaginal sonography was used. All cases were followed by sonography every 2 to 4 weeks to assess the gastrointestinal tract and fetal well-being. Cases in which other major malformations were found were excluded from the study. The antenatal data including the sonograms and neonatal charts were reviewed.

The study protocol was approved by the Institutional Ethics Committee. MEDLINE was searched for articles published in English from 1960 using the key words umbilical cord hernia, fetus, and prenatal diagnosis.

Results

Eight fetuses with prenatal sonographic documentation of isolated umbilical cord hernias were included in this study. During the study period, there were 3 additional cases of umbilical cord hernias associated with other anomalies, which were excluded, and there were no cases that were missed prenatally and diagnosed only after delivery. The median gestational age at diagnosis was 20 weeks (range, 16–28 weeks).

In all 8 cases, a normal insertion into the umbilical cord was observed, and in 3 cases, umbilical cord cysts were identified (Table 1). Seven patients had amniocentesis showing a normal karyotype in all of them. The sizes of the hernias at the time of diagnosis ranged from 12 to 30 mm, and in half of the cases, the sizes of the umbilical cord masses increased during follow-up. In 1 case, sonography showed a hyperechoic bowel at the first examination at 18 weeks’ gestation, which remained hyperechoic until birth (Figure 1).

Apart from 1 case (case 6) no intestinal dilatation or perforation was observed. In this case, intestinal dilatation and fetal ascites were noticed at 28 weeks’ gestation, with loops of bowel floating in the amniotic fluid (Figure 2). At 28 to 30 weeks’ gestation, the size of the lesion increased to a maximal length of 90 mm. This pregnancy was complicated by intrauterine fetal death at 35 weeks’ gestation, and after the birth, rupture of the umbilical cord and umbilical vessels was observed, which might have been the underlying reason for the fetal death. The other 7 patients gave birth to live neonates between 36 and 40 weeks’ gestation (Table 2). In 2 cases, induction of labor was performed, and the other 5 delivered spontaneously. On delivery, the prenatal findings were confirmed. Clamping and dividing of the umbilical cord was performed distally to avoid possible intestinal injury. Surgery was performed on the first day of life and included immediate exploration of the cord and repositioning of the intestines into the abdomen.

Of the 7 neonates, 6 had uneventful postoperative courses, whereas 1, who had a hyperechoic bowel during antenatal follow-up, underwent a second operation 26 days after the primary surgery because of obstruction of the short bowel, and a 10-cm segment of the short bowel was resected. The following postoperative course was without additional complications, and the infant was discharged 41 days after birth. Long-term follow-up at 1 year of age was completed in 4 of the 7 live-born infants. One case was lost.
to follow-up at the age of 1 month, and 2 other cases were followed for several months with no apparent morbidity. Three of the 4 infants who completed the long-term medical follow-up had no medical complications. The fourth patient returned to the hospital at the age of 6 months with unilateral ptosis and juvenile rheumatoid arthritis. Brain magnetic resonance imaging was performed, which showed no abnormalities. The infant was successfully treated with steroid injection and apart from the ptosis and juvenile rheumatoid arthritis was healthy and showed no cognitive abnormalities.

Discussion

Omphaloceles and umbilical cord hernias have similar morphologic features but differ in the embryonic development. An omphalocele results from primary failure of the 4 body folds to form the primary umbilical ring, thus creating an abdominal wall defect. In an umbilical cord hernia, the body folds develop normally and form the umbilical ring. Moreover, in an omphalocele, the rectus muscles have a broad insertion laterally on the costal margins instead of meeting in the midline at the xiphoid, whereas the abdominal wall layers in an umbilical cord hernia are all in their correct anatomic locations. In an omphalocele, the umbilical cord has a characteristic abnormal insertion into the upper region of the herniated sac, unlike an umbilical cord hernia, in which the bowel usually herniates into the base of a normally inserted umbilical cord. Therefore, the major difference between an umbilical cord hernia and an omphalocele is the morphologic characteristics of the umbilical cord insertion.

Embryologically, an umbilical cord hernia is thought to result later than an omphalocele.2,5 Normally, the intestines elongate into the umbilical coelom at about 5 weeks, and between 10 and 12 weeks’ gestation, the intestines return to the peritoneal cavity.3 When this process is interrupted, the intestines are left to some degree in the umbilical cord coelom, resulting in an umbilical cord hernia.

In this study, we showed that the prognosis of an umbilical cord hernia is utterly better than that of an omphalocele, which is associated with an abnormal karyotype in 30% to 40% of cases and other major anomalies in 50% to 70% of cases.3,4 Therefore, it is crucial to differentiate prenatally between these two entities. Only 1 of 8 cases had a fatal complication, and of the 7 live neonates, only 1 had a complicated postoperative course that resolved after a second operation and resection of a 10-cm small-bowel segment.

There are limited data in the literature regarding umbilical cord hernias. Achiron et al2 reported 4 cases that were diagnosed antenatally, 2 of which were terminated in the second trimester. Pal et al6 published a series of 4
neonates with umbilical cord hernias that were diagnosed only postnatally. Three of the 4 neonates were born prematurely at 33 to 35 weeks’ gestation, whereas in our study, all of the neonates were born between 36 and 40 weeks. El-Messidi et al. recently reported on an umbilical cord hernia mimicking a cord teratoma, in which an echogenic mass was noted 10 mm from the abdominal wall but within the umbilical cord substance. Magnetic resonance imaging showed hyperintense structures, suggesting adipose tissue and supporting the diagnosis of a teratoma, but an umbilical cord hernia was found postnatally. Seven of our patients had amniocentesis showing a normal karyotype, but more case series are required before we can reach a conclusion about the need for amniocentesis in cases of isolated umbilical cord hernias.

Once an umbilical cord hernia is diagnosed, the possibility of bowel injury exists and should be discussed with the patient. Sonographic findings such as an echogenic bowel and bowel dilatation may implicate such an injury. In our study, 1 case was complicated by intrauterine fetal death, which occurred several weeks after bowel dilatation and ascites were identified. Therefore, when an umbilical cord hernia is associated with sonographic findings suggesting bowel complications, close fetal monitoring is probably indicated, and early induction of labor at 34 to 36 weeks’ gestation might be considered. However, our suggestion to induce labor earlier once bowel complications are observed is based on this single case of intrauterine fetal death, and more case series are required before this conclusion can be established.

To our knowledge, our study included the largest number of fetuses with the diagnosis of an umbilical cord hernia reported to date. Our findings suggest that in the absence of associated anomalies, the outcomes of fetuses with umbilical cord hernias are favorable; however, these findings need to be confirmed in larger prospective series.

Table 2. Cases of Umbilical Cord Hernias: Neonatal Outcomes

<table>
<thead>
<tr>
<th>Case</th>
<th>Mode of Delivery</th>
<th>Live-Born</th>
<th>Weight, g</th>
<th>Gestational Age at Delivery, wk + d</th>
<th>Onset of Labor</th>
<th>Hospitalization, d</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Vaginal</td>
<td>Yes</td>
<td>3035</td>
<td>36 + 2</td>
<td>Spontaneous</td>
<td>17</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>Cesarean</td>
<td>Yes</td>
<td>2102</td>
<td>36 + 4</td>
<td>Spontaneous</td>
<td>9</td>
<td>None</td>
</tr>
<tr>
<td>3</td>
<td>Vaginal</td>
<td>Yes</td>
<td>2900</td>
<td>40 + 2</td>
<td>Induction</td>
<td>9</td>
<td>None</td>
</tr>
<tr>
<td>4</td>
<td>Cesarean</td>
<td>Yes</td>
<td>3040</td>
<td>37 + 1</td>
<td>Spontaneous</td>
<td>9</td>
<td>None</td>
</tr>
<tr>
<td>5</td>
<td>Vaginal</td>
<td>Yes</td>
<td>2702</td>
<td>40 + 1</td>
<td>Spontaneous</td>
<td>41</td>
<td>Obstruction of short bowel</td>
</tr>
<tr>
<td>6</td>
<td>Vaginal</td>
<td>No</td>
<td>NA</td>
<td>35 + 1</td>
<td>Induction</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>7</td>
<td>Vaginal</td>
<td>Yes</td>
<td>3954</td>
<td>38 + 0</td>
<td>Induction</td>
<td>11</td>
<td>None</td>
</tr>
<tr>
<td>8</td>
<td>Vaginal</td>
<td>Yes</td>
<td>3366</td>
<td>39 + 2</td>
<td>Spontaneous</td>
<td>11</td>
<td>None</td>
</tr>
</tbody>
</table>

NA indicates not applicable.

References