Cervical Neck Masses, Including Lymphatic Malformation (LM) and Teratoma, are Commonly Diagnosed by Prenatal Imaging. Although often benign, the size and location of these masses may encroach on the fetal airway and cause respiratory distress and asphyxiation from airway compression at birth. The mass may distort the neck structures to such an extent that it becomes extremely difficult to either intubate or surgically establish a safe airway.

Advances in prenatal ultrasonography and magnetic resonance imaging (MRI) have been effective in diagnosing fetal neck masses, which has had a direct effect on prenatal management and fetal outcome. Prenatal imaging allows radiologists to identify a potentially fatal airway obstruction before labor and delivery. Obstetricians, neonatologists, radiologists, anesthesiologists, and surgeons can then develop and discuss with families a comprehensive plan of action for the safe delivery of the fetus.

Increasingly, fetal surgery and the ex utero intrapartum treatment (EXIT) procedure are used to manage these complex cases. These procedures were designed to provide time to secure an airway while uteroplacental gas exchange is preserved. In this study, we describe our institution’s experience with the diagnosis and management of fetal cervical neck masses and discuss the efficacy of prenatal imaging and the safety of the EXIT procedure in these patients.

METHODS

We reviewed our consecutive experience of all fetal patients with cervical teratoma and LM between January 2001 and December 2003 at Children’s Hospital. Data collected retrospectively from medical records included maternal and fetal demographics (maternal age, antepartum complications, and gestational age at diagnosis), indications for referral, results of prenatal imaging studies (fetal ultrasonography and MRI), need for surgical intervention, and complications.
RESULTS

The database search generated 8 patients with prenatally diagnosed cervical neck masses. Four of the patients were diagnosed as having LM, and 4 were diagnosed as having teratoma. Maternal age ranged from 20 to 39 years (average, 28.4 years). Mothers of fetuses diagnosed as having cervical LM were slightly older (average age, 30.3 years) than those of fetuses diagnosed as having cervical teratoma (average age, 26.5 years). All of the patients were initially diagnosed as having a fetal cervical mass at outside institutions by ultrasonography.

Gestational age at initial diagnosis ranged from 16.6 to 38.7 weeks (average, 27.2 weeks). The average gestational age of patients with LM was 26.6 weeks, which was similar to the patients with teratoma (average, 27.4 weeks). Overall gestational age at delivery was 35.3 weeks (range, 30-38 weeks). Fetuses diagnosed as having LM were slightly older at delivery (average gestational age, 37.8 weeks) than those diagnosed as having teratoma (average gestational age, 33.4 weeks). Only 1 fetus was diagnosed as having a medial comorbidity (Down syndrome).

All 4 patients with LM underwent normal vaginal delivery without complications. None of the newborns required intubation in the delivery room. All 4 mothers with fetal teratomas underwent a successful EXIT procedure without complications. Three of the patients required tracheotomy, and 1 was successfully intubated during the EXIT procedure. Each of the patients with suspected teratoma underwent resection, with a confirmed histopathologic diagnosis of teratoma in all.

IMAGING RESULTS

All 8 patients underwent ultrasonographic and MRI studies at a tertiary care center after a neck mass had been discovered by ultrasonography in an outside institution. Ultrasonography and MRI accurately characterized the mass as either an LM (n=4) or a teratoma (n=4). Lymphatic malformations were characterized as simple fluid collections with internal septations, centered in the subcutaneous fatty layer on ultrasonography and MRI. The involved skin included the region of the anterior neck (n=1), parotid neck (n=1), and lateral neck (n=2). The mean LM was 4.6 x 4.4 x 3.4 cm. None of the LMs seemed to impinge on or deviate the fetal trachea.

Teratomas were characterized as complex, primarily solid masses, containing visible blood flow on ultrasonography and MRI. The masses that proved to be teratomas were positioned in the fetal neck (n=1), oropharynx (n=1), or both (n=2). The mean teratoma was 8.3 x 7.3 x 6.7 cm. All of the teratomas obstructed the oropharynx (n=1), caused a deviation in the fetal trachea (n=1), or both (n=2).

ANESTHESIA MANAGEMENT

The anesthetic management was standardized in the 4 patients who required the EXIT procedure. All women received a rapid-sequence induction with thiopental sodium, succinylcholine chloride, and fentanyl citrate, followed by endotracheal intubation. Anesthesia was then maintained with 2 to 3 minimal anesthetic concentration isoflurane or desflurane in 100% oxygen. After a low transverse hysterotomy and exposure of the fetus, a fetal upper extremity was delivered to apply a pulse oximeter probe and to obtain intravenous access. Although the fetus was anesthetized via placental transfer of maternally administered inhaled anesthesia, additional analgesia and paralysis agents were administered (ie, fentanyl, atropine, and vecuronium bromide). Coordination between the surgery, obstetric, and anesthesiology teams was crucial to prevent excessive maternal hemorrhage from a completely relaxed uterus. No anesthetic complications were noted.

COMMENT

Congenital anomalies of the head and neck frequently compromise the airway and may cause asphyxia after delivery of the newborn. Cervical teratoma and LM are 2 of the most common congenital lesions that may present with airway obstruction at delivery. In addition to airway obstruction, these lesions may cause compression of the esophagus, resulting in polyhydramnios, uterine irritability, and preterm labor. Depending on the size and location of the mass, these anomalies may have a mortality of 80% to 100%.

In the past decade, with advances in fetal ultrasonography and MRI, a host of fetal anomalies, including cervical neck masses, can be identified in the first trimester. Lymphatic malformations and teratomas have specific ultrasonographic and MRI characteristics that permit their accurate prenatal diagnosis. Lymphatic malformations are often characterized as simple fluid collection with internal septations, centered in the subcutaneous fatty layer on ultrasonography and MRI. Teratomas are characterized as complex primarily solid masses, containing visible blood flow on ultrasonography and MRI. Because of the advances in prenatal imaging, the nature of these lesions and their...
proximity to surrounding vital neck structures, such as the trachea, can be evaluated to determine airway patency or compression. This information provides the opportunity for proper intervention to prevent a fatal outcome.

FETAL SURGERY

In 1963, Sir William Liley performed the first successful fetal procedure, consisting of an intraperitoneal blood transfusion to a fetus affected with erythroblastosis fetalis. In the past 2 decades, fetal surgery has become popularized in the management of congenital anomalies that may lead to complications at delivery.7-10

The indications for fetal surgery are evolving (Table). In general, fetal therapy or surgery is indicated when the risk of death or severe disability to the fetus is greater than no intervention and the risk to the mother remains low.2 Conditions such as maternal polyhydramnios or oligohydramnios should raise the suspicion for fetal disease, which leads to further investigation. Contraindications to perform fetal surgery include a disabling or lethal structural and/or genetic abnormality in the fetus or a serious medical disease in the mother.2

Fetal airway obstruction is a common reason for fetal surgery. While generally caused by compression of a giant neck mass, such as a cervical teratoma or an LM, fetal airway obstruction can also be due to intrinsic defects in the larynx or trachea. These latter abnormalities fall under the clinical syndrome of congenital high airway obstruction syndrome. Some causes of congenital high airway obstruction syndrome include laryngeal web, laryngeal cyst, laryngeal atresia, and tracheal atresia.11,12

When airway obstruction is suspected, an EXIT procedure at delivery can provide the needed time to secure the fetal airway. The EXIT procedure was designed initially for reversal of mechanical tracheal occlusion, performed in fetuses with severe congenital diaphragmatic hernias.13 It provided an optimal controlled environment to reverse the tracheal occlusion while receiving uteroplacental circulation. Because of the success of the EXIT procedure and the prolonged stable fetal hemodynamic condition it provided, it has been used in the management of fetal neck masses causing airway obstruction.14-16

The EXIT procedure entails delivering the fetal head through a controlled hysterotomy and managing the airway while fetal gas exchange is maintained via the placenta. Deep inhalational anesthesia is used to maintain uterine relaxation and preserve uteroplacental gas exchange. This is in contrast to a cesarean delivery performed under general anesthesia, in which the major goals are to minimize the time from anesthesia to cord clamping and to decrease the exposure of the fetus to inhalational anesthetics.4

For optimal outcome of the EXIT procedure, the anesthesiologist must ensure adequate fetal anesthesia, adequate fetal oxygenation, and uterine relaxation. In general, the fetal gas exchange can be supported by ex utero placental circulation for 60 minutes. This strategy will provide enough time to attempt laryngoscopy, bronchoscopy, or possibly resection of a neck mass to secure the airway by intubation or tracheotomy. After the newborn’s airway is secured, the umbilical cord is clamped and delivery is completed. The EXIT procedure is scheduled as close to term gestation as possible to avoid the problems associated with prematurity.5 There are several potential risks to the mother that can be minimized through proper coordination between the surgeon, obstetrician, and anesthesiologist. These include uterine atony and placental abruption, both of which may result in maternal hemorrhage.14

FETAL IMAGING

Fetal imaging allows accurate assessment of the nature and position of neck masses in fetuses. Ultrasonography is readily available and relatively inexpensive. Its real-time assessment of fetal breathing efforts and tissue characterization is often diagnostic in itself.17 In addition, it is sometimes possible to diagnose tracheal position, particularly with the aid of color Doppler ultrasonography.18 Doppler ultrasonography allows distinction between flowing blood vessels and the fluid-filled trachea. The more mature the fetus, however, the more difficult tracheal visualization becomes with ultrasonography, because the growing mass and fetal skull and mandibular ossification may obscure tracheal visualization.

Fetal MRI, on the other hand, reveals more detailed airway structures as the pregnancy progresses (Figure 1). The mainstay of fetal MRI is the fast spin-echo T2-weighted sequence.19-21 With this technique, the high-signal fluid that fills the fetal trachea makes airway structures conspicuous and can even allow visualization of the palate and piriform sinuses. If the airway seems encased or deviated by a neck mass, the position and contour of the trachea can be carefully and precisely defined by fetal MRI (Figure 2). With predelivery mapping of tracheal structures in a controlled environment, airway control can be planned and likely sites for a successful tracheotomy, if necessary, can be determined ahead of time (Figure 3).

<table>
<thead>
<tr>
<th>Disease</th>
<th>Indication</th>
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<tbody>
<tr>
<td>Thoracic</td>
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<td>Congenital diaphragmatic hernia</td>
<td>Liver in chest</td>
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<td>Congenital cystic adenomatoid malfor-</td>
<td>Hydrops</td>
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<tr>
<td>Pulmonary sequestration</td>
<td>Hydrops</td>
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<td>Airway obstruction</td>
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<td>Giant neck mass</td>
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<td>Congenital high airway obstruction</td>
<td>Hydrops</td>
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<tr>
<td>Cardiac</td>
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<td>Third-degree heart block</td>
<td>Ventricular response &lt;45 beats/min</td>
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<tr>
<td>Sacrococcygeal teratoma</td>
<td>High-output heart failure</td>
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Anesthesia for fetal surgery involves the fetus and the mother. This differs from the anesthesia that is required for maternal surgery or fetal therapy. The mother is the only active recipient during the maternal surgery (cesarean delivery), and the fetus is the only active recipient during the fetal therapy (blood transfusion and amni-
otic fluid reduction). Therefore, successful fetal surgery is dependent on safe and adequate anesthesia for the mother and the fetus, who are both the active recipients during the surgery.2

The anesthesia for fetal surgery is based on maternal, fetal, and uteroplastic factors. It is of primary importance to ensure complete uterine relaxation throughout the duration of uteroplastic support to the fetus. It is also essential to preserve the maternal-fetal gas exchange at the placental interface to ensure fetal oxygenation and avoid potentially life-threatening hypoxemia. All inhaled anesthetics cross the placenta. Fetal uptake depends on uterine blood flow, the solubility of the drug in fetal blood, and the fetal distribution of the drug. Of all factors ensuring the overall success of the fetal surgery, minimal uterine vascular resistance is the most important because decreases in uterine blood flow would cause fetal hypoxemia, acidosis, and potential fetal death.

In conclusion, congenital abnormalities in the head and neck may cause airway obstruction in the newborn, requiring immediate intervention. In the past, many newborns succumbed to airway obstruction because of the inability to establish a safe airway at delivery. Technological advances in prenatal ultrasonography and MRI have improved the ability to diagnose congenital abnormalities in utero. This allows for proper assessment of the airway to prevent any unexpected problems at delivery. We believe that many of the airway emergencies can be avoided by prenatal imaging and initiation of airway management in the prenatal period.

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