Case 7-2009: A Pregnant Woman with a Large Mass in the Fetal Oral Cavity

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A 29-year-old pregnant woman was transferred to this hospital at 38 weeks 4 days of gestation because of a large mass that had been detected in the fetal oral cavity by ultrasonography.

The patient was a primigravida and had received routine prenatal care at another facility, including ABO blood typing (type O, Rh-negative, antibody-negative), an oral glucose-tolerance test (abnormal results), and a screening test for group B streptococcus (positive); screening tests for syphilis and viral hepatitis, types B and C, were negative. Gestational diabetes was controlled by diet, and Rh\(_{(D)}\) immune globulin had been administered at 28 weeks of gestation. Fetal ultrasonography at 8 weeks 3 days of gestation reportedly showed no abnormalities, and subsequent ultrasound studies for routine monitoring purposes were reported to show no fetal abnormalities.

At 37 weeks 5 days of gestation, ultrasonography revealed a possible cystic structure, 19 mm by 17 mm, in the mouth of the fetus. The next day, a repeat ultrasound study performed at another hospital showed an intrauterine fetus in the vertex position, with an estimated weight of 3811±221 g (90th percentile) and a cystic structure in the oral cavity that was 2.3 cm in diameter and did not protrude from the mouth. No cleft lip was seen; the remainder of the facial anatomy was difficult to see. The placenta was in the anterior position.

Four days later, at 38 weeks 3 days of gestation, the mother was admitted to another hospital for elective induction of labor, and two doses of misoprostol were administered. However, a neonatologist at the other hospital expressed concern about the infant’s airway; the induction was discontinued, and the next day, the mother was transferred to this hospital.

The mother had obesity, polycystic ovaries with oligomenorrhea, a uterine leiomyoma that was 10 cm in diameter, and eczema. Papanicolaou smears had been normal, and she had had an appendectomy in the past. There was no history of sexually transmitted diseases. She had no allergies. She was married, lived with her husband, and did not smoke, drink alcohol, or use illicit drugs. There was no family history of gynecologic or gastrointestinal cancer.

On examination, the vital signs were normal and the abdomen was soft, gravid, and nontender. The fetal heart rate was 150 beats per minute, with moderate vari-
ability and no decelerations. There was no evidence of labor. Routine laboratory-test results were normal. An ultrasound examination (Fig. 1) confirmed the presence of a cystic structure, 2.0 cm by 2.3 cm, in the mouth of the fetus. The placenta was anterior, extending from the lower uterine segment to the fundus. A uterine fibroid, 9.0 cm in diameter, was seen in the anterior lower uterine segment, to the left of midline.

On the second hospital day, a T2-weighted image from a magnetic resonance imaging (MRI) scan of the fetus revealed a hyperintense, multiloculated, cystic mass (3.4 cm by 2.6 cm by 3.4 cm) in the midline of the mouth, predominantly in the sublingual space; the mass originated above the mylohyoid muscle and displaced the tongue posteriorly. The maxilla, palate, cranial cavity, and brain were normal.

A management decision was made.

**Differential Diagnosis**

Dr. Christopher J. Hartnick: I received a telephone call from a neonatologist about this pregnant woman in whom labor had just been induced, because of concern about how to ensure the safety of the newborn’s airway during and after delivery. On our advice, the induction was stopped and the mother was transferred here for discussion of a possible ex utero intrapartum treatment (EXIT) procedure.

**Obstetrical Considerations**

Dr. William H. Barth, Jr.: From the perspective of the obstetrician, the two decisions to be made were the timing and the route of delivery. In the absence of clinical indications to the contrary, our usual recommendation is to await spontaneous labor at term and anticipate a vaginal delivery. In this case, planning the timing of the delivery with the knowledge that the neonatal airway could be compromised raised the issue of how to ensure the availability of the many personnel and resources that may be needed. All involved thought that a scheduled procedure would facilitate the availability of personnel. A vaginal delivery or traditional cesarean section presented the possibility of neonatal hypoxia during any time required to secure the airway.

We determined that the optimal method of delivery would be by a planned EXIT procedure during the daylight hours. The EXIT procedure involves partial delivery of a fetus to secure an airway while maintaining the uteroplacental circulation, permitting uninterrupted fetal oxygenation and gas exchange. However, the ultrasound study revealed that the placenta covered the entire anterior uterine surface, extending well into the lower uterine segment, where a traditional cesarean hysterotomy would be performed. Therefore, it appeared that an anterior uterine incision would risk disrupting the placenta; this would still permit safe delivery of the infant but would not permit an EXIT procedure, since the placenta would be compromised by the incision. In view of the estimated fetal weight of nearly 3900 g and the maternal body habitus, we believed that a posterior or fundal hysterotomy was not possible without undue risk to the mother. Therefore, we initially recommended a traditional cesarean section of the anterior lower uterine segment and planned to hand the baby to the awaiting pediatricians for
airway management. Repeat MRI was performed on the second hospital day to better assess the relationship of the fetus and placenta to the uterus.

Dr. P. Ellen Grant: To better determine the location of the lesion in the fetus and the location of the fetus in the gravid abdomen, a fetal MRI was obtained. A multicystic lesion was again identified and confirmed to be within the sublingual space, with the tongue on the superior margin of the lesion (Fig. 1). The nasopharynx and oropharynx were almost completely obstructed by the mass. A clearly patent trachea could not be identified on these images. Cine MRI clips (Videos 1 and 2, available with the full text of this article at NEJM.org) did not provide evidence of a pedunculated mass or movement of the mass into the oropharynx. The lesion remained within the sublingual space, suggesting that it was attached to the floor of the mouth or to the tongue.

The differential diagnosis of a sublingual cyst includes dermoid, epidermoid, and thyroglossal duct cysts, congenital ranula, mucocele, vallecular cysts, and enteric duplication cysts. Dermoid and epidermoid cysts are the most common. Congenital ranulas tend to be located laterally and are therefore less likely in this case. A mucocele is possible. A vallecular cyst or thyroglossal duct cyst is also possible, but these are usually at the base of the tongue. When they are large, they can look similar to this lesion. An enteric duplication cyst in this location would be rare, but it has been reported. A specific diagnosis is not possible on the basis of the imaging findings.

Additional MRI views of the gravid abdomen determined the location of the fetus and provided the information necessary to assess the possibility of an EXIT procedure. The MRI scans showed a small window for fetal access (Fig. 2) between the pubic bone inferiorly, the placenta superiorly extending to the right, and a fibroid superiorly on the left. The overall size of the window was approximately 5 cm from the placenta to the pubic symphysis and 7 cm laterally from the placenta to the fibroid. We also noted that the uterine artery was displaced medially over the fibroid.

Dr. Barth: The detailed information from the MRI suggested that it would be feasible to incise the uterus and deliver the infant without disrupting the placenta. Thus, we decided to attempt the EXIT procedure.

Dr. Hartnick: The EXIT procedure required assembling a multidisciplinary team whose members would have specific responsibilities during the procedure, including maternal and fetal medicine and obstetrics, maternal and fetal anesthesia, pediatric anesthesia, pediatric otolaryngology, pediatric surgery, and neonatal intensive care. In this case, the team comprised 28 health care professionals, including doctors and nurses. Teamwork with such a large group requires careful choreography, and a dress rehearsal was conducted so that all participants not only knew their roles but also knew where they would be standing, where their hands would be, and what instruments would be available to them. Two adjacent operating rooms were available, one for the EXIT procedure itself and another to which the infant would be moved for stabilization, as well as for the assessment and possible resolution of the airway obstruction.

MATERNAL ANESTHESIA

Dr. Meredith A. Albrecht: In any obstetrical procedure, but particularly an EXIT procedure, there are two patients — the mother and the fetus — and different goals for each. In a typical cesarean section, our goals for the mother are to ensure adequate anesthesia (both pain control and avoidance of awareness); prevent maternal aspiration, which may occur because of decreased lower-esophageal sphincter tone in pregnancy; and avoid uterine relaxation, which can lead to uterine atony and hemorrhage. Our goal for the fetus is to ensure adequate fetal perfusion by avoiding aortocaval compression and maintaining normal maternal blood pressure. We also try to minimize fetal respiratory depression by limiting maternal anesthesia before delivery of the fetus, which is performed as quickly as possible, usually in about 3 to 5 minutes from the skin incision to delivery of the fetus.

During an EXIT procedure, some of these goals are altered, as in this case. First, there are two critical time periods: before and after complete delivery of the fetus. The goals of avoiding aspiration and ensuring anesthesia in the mother are the same as those in a typical cesarean section. However, while the fetus is only partially delivered, uteroplacental circulation must not be compromised; this requires deep uterine relaxation and preservation of the maternal blood pressure, which should be monitored with an arterial catheter.
Unfortunately, most agents that relax the uterus, such as deep inhalational anesthetic agents or nitroglycerin, cause a decrease in the maternal blood pressure; this can be treated with vasopressors such as phenylephrine and ephedrine. Since profound uterine relaxation increases the risk of uterine atony, we had to be prepared for hemorrhage, with adequate blood available and large-bore, intravenous catheters in place. Several uterotonic agents were available for use after complete delivery of the fetus.1,4-6

ANESTHESIA OF THE INFANT
Dr. Charles J. Coté: The day before delivery, after review of the MRI scans and discussions with our colleagues in otolaryngology, obstetrics, and neonatology, multiple options for airway management were proposed: establishing an airway by nasal or oral intubation, direct laryngoscopy and intubation, aspiration of the cyst, insertion of a laryngeal mask airway (LMA) or fiberoptic intubation through the LMA, rigid bronchoscopy, the sliding of an endotracheal tube off the bronchoscope, cricothyrotomy, and tracheostomy. We also had a detailed discussion with the family. Sterilized airway equipment, an anesthesia breathing circuit, and a pulse-oximeter probe and cable were made available.

The setup for the morning of delivery included suction equipment; oxygen; airway equipment consisting of a separate, sterile table with the necessary ear, nose, and throat and anesthesia equipment (a nitric oxide delivery system, blood [type O, Rh-negative], tuberculin syringes with preestimulated drug doses for possible intramuscular administration to the fetus,7 although maternal inhalation anesthetic agents should be adequate,8,9 and a pulse-oximeter probe and cable10-12); and a cricothyrotomy catheter. We prepared a separate, warmed operating room for neonatal anesthesia that included intravenous infusion (10% dextrose in water) with piggybacked and primed remifentanil (0.1 μg per kilogram of body weight per minute) and resuscitation drugs.

THE EXIT PROCEDURE
Dr. Barth: As with any cesarean section, we administered prophylactic antibiotics and used pneumatic compression boots to prevent deep-vein thrombosis. Cross-matched blood was available in the operating room. During a routine cesarean delivery, the patient is usually placed in the supine position with slight left tilt and with her legs together on a flat operating table. In order for the pediatric surgeon to stand between the patient’s legs and have access to the operative field, we placed the patient’s legs in Allen stirrups with her knees slightly abducted. After general anesthesia was achieved, we began with a traditional Pfannenstiel incision, but we made it larger than...
normal to avoid compromising the procedure. Before incising the uterus, we placed an ultrasound probe covered by a sterile sleeve directly on the myometrium to define the lower margin of the placenta, so that we could avoid it. We found that we had more room than imaging had suggested. Possibly, the lower segment had thinned slightly because of the misoprostol that the mother had received for the induction of labor. We moved the bladder away from the lower uterine segment and made a traditional low transverse incision in the uterus, but not through the amniotic membranes. We encountered the uterine artery, which we controlled with large ring clamps; we were thus able to maintain hemostasis and proceed with the EXIT procedure. At that point, we delivered the baby’s head to the point of the shoulders and put the ultrasound transducer probe back on the wall of the uterus, which allowed us to monitor the fetal heart rate during the EXIT procedure.

Dr. Barth: In contrast to the typical cesarean section, in which the elapsed time from the uterine incision to delivery averages less than 1 minute, the duration of the EXIT procedure was 10 minutes. Once the airway was secured and the infant was moved to the warmer, our technical role was not very different from the role we would have had in a typical cesarean section, except that the mother had a very flaccid uterus. We depended on the obstetrical anesthesiologist to reverse the maneuvers that had intentionally been done to relax the uterus. The concentration of general anesthetic was decreased; relatively high-dose oxytocin was administered, and methylergonovine, a uterotonic agent, was given intramuscularly to cause the uterus to contract. We manually removed the placenta and then closed the uterus, incorporating and controlling the uterine artery, as with a standard cesarean section. The estimated blood loss was 1200 ml, which is slightly more than the amount lost in a normal cesarean section. The mother was transferred to the recovery room, and she recovered uneventfully.

**Excision of the Cyst**

Dr. Hartnick: Ten days after the EXIT procedure, the child was returned to the operating room. The tracheal stoma was examined, and the tract was noted to be stable and to have matured. Bronchoscopy revealed a small residual membrane at the level of the left false vocal fold that neither prolapsed into nor obscured the airway.

We found two adjoining cysts in the floor of the mouth (the larger one was 3.5 cm by 2.0 cm by 1.0 cm), which adhered to the ventral surface of the tongue (Fig. 4A). The two cysts were swept off the mylohyoid musculature and out of the floor of mouth; bipolar cautery was used to remove them from the tongue and to provide hemostasis. The two cysts were removed en bloc from the floor of mouth by exposing, identifying, and preserving both submandibular ducts and both lingual nerves (Fig. 4B). The wound was closed primarily.

**Clinical Diagnosis**

Floor-of-mouth cyst; possible ranula.
Figure 3. Ex Utero Intrapartum Treatment (EXIT) Procedure.
The patient’s legs were placed in Allen stirrups, slightly abducted (Panel A), to allow the pediatric surgeons access to the operative field. A Pfannenstiel incision was made (Panel B), slightly larger than normal, avoiding the lower margin of the placenta. The baby’s head and shoulders were delivered through the hysterotomy incision (Panel C), leaving the rest of the body in the uterus and the placental circulation intact. The head was held by the obstetrician while direct laryngoscopy and intubation were performed (Panel D, intraoperative photograph). A bronchoscopic image (Panel E) shows the supraglottic web, which has been pierced with the bronchoscope; there is a residual band attached to the left false vocal fold, seen on the left of the image.
**Pathological Discussion**

**Dr. Julia T. Geyer:** The specimen consisted of two adjacent cysts, 3.5 cm and 0.8 cm in greatest dimension. On opening the cysts, we found that they had a smooth gray-white to pink inner surface and contained abundant mucoid material. Histologically, both cysts had a well-defined smooth-muscle wall (Fig. 5A). The epithelial lining contained squamous mucosa, as expected in the oral cavity, but also different subtypes of ectopic epithelia, such as ciliated respiratory-type mucosa (Fig. 5B), foveolar gastric mucosa, and small-bowel mucosa (Fig. 5C). These findings are consistent with a diagnosis of a foregut-duplication cyst.\(^ {13,14}\)

Enteric duplications are uncommon congenital abnormalities found along the alimentary tract from the tongue to the anus. The foregut gives rise to the tongue, pharynx, lower respiratory tract, esophagus, and stomach and the first two parts of the duodenum and hepatobiliary tract. Foregut-duplication cysts are the least common of the enteric duplications,\(^ {15}\) and most are found in the thorax and abdomen. Of those that arise in the...
head and neck region, most originate in the anterior or posterior tongue, as this one did; rarely, they have been reported in the epiglottis, larynx, pharynx, and floor of the mouth. The pathogenesis of foregut duplications is still unclear; it is postulated that they result from failure of the notochord and the foregut to separate during early embryonic development. This theory could explain why approximately half of patients with foregut-duplication cysts have associated skeletal malformations, including cervical and thoracic hemivertebrae, scoliosis, and spina bifida.

**FOLLOW-UP**

**Dr. Hartnick:** No skeletal malformations were evident in this newborn. One week after the resection of the cyst, she was feeding well orally. Decannulation was initially unsuccessful because of the presence of moderate tracheomalacia, so the tracheostomy tube was left in place. The child was discharged at 1 month of age, with the tracheostomy tube. Repeated bronchoscopy at the ages of 3 months and 6 months disclosed persistent tracheomalacia; by the age of 9 months, the tracheomalacia had resolved, and tracheal decannulation was successfully performed when the child was 10 months old. The child is developing normally.

**ANATOMICAL DIAGNOSIS**

Foregut-duplication cyst, arising from the ventral surface of the tongue.

**REFERENCES**

17. Kim JH, Park KK. Foregut duplication cyst of the hypopharynx: a rare cause of


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