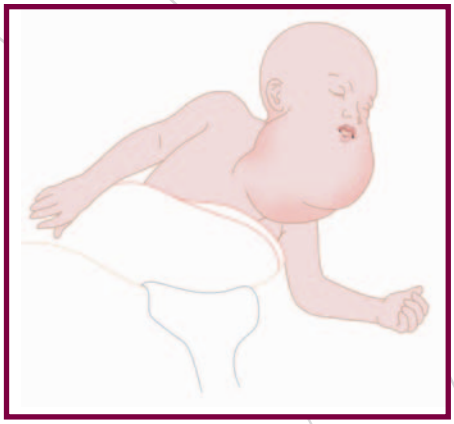
Cervical Teratoma (CT)

What is cervical teratoma (CT)?

CT is a rare tumor of the neck made up of a variety of tissues (nerves, cartilage, skin and thyroid among others) that come from more than one embryonic layer.



Above is an illustration of CT.

What causes cervical teratoma (CT)?

The cause of CT is unknown. An old theory holds that it is caused by an inability of cells to develop into a complete body, or an abnormal development of a conjoined twin. More likely, CT arises from stem cells within the thyroid gland that grow abnormally into a tumor.

What is the incidence of cervical teratoma (CT)?

Over 150 cases of congenital CT have been reported. There is no apparent relationship to the mother's age, no greater odds of it occurring in males versus females and no racial or ethnic preference.

How is cervical teratoma (CT) diagnosed?

An ultrasound is usually the best way to make a diagnosis. CTs are asymmetric, well-defined masses usually off to one side of the baby's neck. As many as 50 percent of CTs have calcifications. The CT is typically large and bulky, measuring five to 12 cm in diameter.

These tumors may grow larger than the fetal head. Polyhydramnios (excessive accumulation of amniotic fluid) can complicate 20 to 40 percent of prenatally diagnosed cases. Other fetal abnormalities have been reported in association with CT. They are generally benign but have malignant potential.

How is pregnancy managed when cervical teratoma (CT) is diagnosed?

Frequent ultrasound exams are recommended to monitor amniotic fluid volume, tumor size, growth and the general health of the fetus. Premature labor and delivery in cases of CT are common. The increase in uterine size due to polyhydramnios can precipitate preterm labor and/or delivery. This may necessitate an emergency EXIT (ex utero intrapartum treatment) procedure to secure the newborn's airway at delivery. Airway obstruction can be life threatening and accounts for up to 45 percent of the mortality associated with CT.

What are the fetal interventions for cervical teratoma (CT)?

There have been several anecdotal reports of intrapartum laryngoscopy or bronchoscopy in cases of fetal neck masses in which the fetus is delivered and the cord is not clamped. However, this approach offers no advantage over standard cesarean section because there were no attempts to prevent uterine contractions. In most cases, the fetus was removed from the uterus, resulting in the loss of uterine volume, uterine tone, placental separation and cessation of uteroplacental gas exchange.

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The procedure was originally developed for the delivery of fetuses with diaphragmatic hernias who had undergone in utero tracheal clip application to induce prenatal lung growth. The EXIT procedure was specifically designed to preserve uteroplacental gas exchange to provide time to secure the airway as an adjunct to performing the fetal tracheal clip procedure. For this indication, the EXIT procedure provides time for:

- Neck dissection
- Clip removal
- Bronchoscopy
- Endotracheal intubation
- Surfactant administration
- Placement of umbilical arterial and venous catheters

Unlike the EXIT, in conventional cesarean section, there is no attempt to prevent bleeding from the hysterotomy (surgical incision of the uterus). Hemostasis is achieved by return of the uterine tone following delivery of the fetus, clamping of the cord and placental separation.

In the EXIT procedure, deep inhalational anesthesia insures uterine relaxation, which is crucial to preserving uteroplacental gas exchange. Because of the significant hemorrhage from the hysterotomy, the uterus is opened using a uterine stapling device.

Skarsgard et al were the first to apply these principles of the EXIT procedure to the management of the fetus with prenatally diagnosed tracheal obstruction. Liechty et al reported a series of five giant fetal neck masses managed by EXIT procedure, describing cord blood gas data for procedures lasting eight to 54 minutes while on placental support.

The ability of inhalational anesthetic agents to maintain uterine relaxation and uteroplacental gas exchange is apparent by the relatively normal cord blood gas values seen up to 54 minutes while on uteroplacental support.

There are a number of potential risks of the EXIT procedure to the mother, including uterine atony (lack of tone), which may result in increased hemorrhage. The risk of hemorrhage from uterine atony can be minimized through coordination between surgeon and anesthesiologist to decrease the concentration of inhalational anesthetic and to administer oxytocin before umbilical cord ligation.

This, in combination with the uterine stapling device, keeps the average intraoperative maternal blood loss at 930 ml, well within the accepted range for traditional cesarean section. The involvement of the placenta in hysterotomy can occur resulting in increased blood loss.

Polyhydramnios can compress the edge of the placenta, obscuring its edge as assessed by ultrasonography. Amnioreductions are performed in cases with severe polyhydramnios before performing an EXIT procedure to allow better delineation of the placental edge.

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Uterine rupture during labor in future pregnancies is an additional risk if a classic cesarean incision is required. The low-anterior transverse uterine incision is preferred because it allows the possibility of vaginal births. However, vaginal birth after cesarean (VBAC) may be contraindicated with a low anterior placental position. Before the EXIT procedure, the mother should be counseled that cesarean delivery may be required for all future pregnancies.

Giant fetal neck masses may complicate perinatal management due to polyhydramnios, pre-term labor and the challenge of airway compromise. Airway compromise is a result of the location of the mass which distorts the airway, and does not necessarily represent the absolute size of the neck mass.

Intensive maternal-fetal monitoring, hemostatic hysterotomy with maximal uterine relaxation and maintenance of intact utero-placental circulation provide a controlled environment for securing the airway.

Conventional cesarean delivery and routine newborn airway management are very difficult, if not impossible, which may result in delays in securing the airway and causing possible anoxic brain injury or death.

Airway obstruction at birth is life threatening and associated with a high mortality. In giant fetal neck masses, this mortality is usually associated with a delay in obtaining an airway and the inability to effectively ventilate the infant.

This delay can result in hypoxia (oxygen deficiency) and acidosis (result of excess acid production) and, if the delay is greater than five minutes, anoxic brain injury may occur. This complication is all the more tragic as most of these children have an isolated benign tumor and do well after postnatal resection.

Delaying surgery can result in retention of secretions, atelectasis, and/or pneumonia due to interference with swallowing. In addition, precipitous airway obstruction may occur due to hemorrhage into the tumor, even in minimally symptomatic newborns. For this reason, orotracheal intubation is indicated in all patients regardless of the presence or absence of symptoms.

Mortality decreases to between nine and 17 percent in infants treated surgically. It should be noted, however, that these tumors tend to be large, disfiguring masses that envelope vital structures in the neck.

Extensive neck dissection and multiple procedures may be necessary to achieve the goals of complete extirpation of the tumor with acceptable functional and cosmetic results.

In a review of 18 cases of cervical and oral facial teratomas, Azizkhan and colleagues reported that life threatening airway obstruction occurred in seven cases (39 percent), two of whom died without ever having a secure airway.

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Two neonates with prenatally diagnosed tumors survived because tracheostomies were performed in the delivery room by an attending pediatric surgeon. The overall survival was observed in 15 of the 18 cases (83 percent). One neonate required multiple surgeries to achieve complete tumor extirpation. Morbidity included:

- Two cases with recurrent laryngeal nerve injury
- Two cases with hypothyroidism
- Two cases with developmental delay and mental retardation secondary to asphyxia at birth due to airway obstruction

These infants are at risk for transient or permanent hypoparathyroidism and hypothyroidism. CT may completely replace the thyroid gland, and tumor resection may result in permanent hypothyroidism. More commonly, thyroid tissue may be preserved but may not be adequately functioning. An interval of thyroxine supplementation may be necessary.

Due to the massive nature of these tumors and the difficulty of identifying parathyroid glands in a newborn, transient or permanent hypoparathyroidism may be observed. Calcium and Vitamin D supplementation may be needed postoperatively. A pediatric endocrinologist should be consulted should these complications be encountered.

CTs are functional tumors in approximately two-thirds of the cases, producing markedly elevated levels of alpha-fetoprotein (AFP). However, an elevated AFP level obtained immediately postoperatively must be interpreted with caution. AFP levels in normal newborns have an enormous range of values, with some as high as 100,000 units in the first month of life.

The AFP values progressively fall during infancy until levels of less than four units are obtained at one year of age. While an elevated AFP may not necessarily be abnormal, the levels should decrease during infancy. A rising AFP should alert the clinician to the possibility of teratoma recurrence.

While CT is generally a benign tumor, there is the possibility of malignant transformation. Close surveillance for tumor recurrence is essential.

It is recommended that AFP levels be obtained and followed at three-month intervals in infancy and yearly thereafter, with CT or MRI scanning twice a year for the first three years of life.

What are treatment options for the newborn?
Airway obstruction at birth is the primary reason for mortality in cases of CT. Mortality can be as high as 80 to 100 percent in untreated infants. Mortality decreases to 9 to 17 percent in infants treated surgically.

What is the long-term outcome with cervical teratoma (CT)?
Infants with CT are at risk for serious thyroid conditions, including:

- Hypoparathyroidisim (deficiency of parathyroid hormone, causing abnormal calcium metabolism)
- Hypothyroidism (deficient activity of the thyroid gland, characterized by a lowered metabolic rate and general loss of vigor)

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CT may completely replace the thyroid gland. Surgical removal of the tumor may result in permanent hypothyroidism. More commonly, a portion of the thyroid is preserved but may not function properly, requiring supplemental thyroid hormones as well as vitamin and calcium supplements.

CTs may grow so large that the parathyroid glands are enveloped. If all four glands are removed with the CT, profound hypocalcemia (low blood calcium levels) results. This causes muscle spasms and must be treated with supplemental calcium and Vitamin D.

While CT is generally a benign tumor, there is the possibility it can transform into a malignant tumor. Close observation of the patient is essential to detect tumor recurrence. CT has been shown to metastasize to reginal lymph nodes even though the CT is a benign tumor.

Serial examinations and CT (computed tomography) scans of the neck are necessary to exclude the possibility that a benign focus of CT undergoes malignant transformation.

There has been only one report of congenital CT occurring among siblings.

Contact the Fetal Care Center of Cincinnati

For more information, please call 1-888-FETAL59 or email us at info@fetalcarecenter.org.

References

For a detailed reference list, visit our web site at www.fetalcarecenter.org.