American Pediatric Surgical Association

Prenatal Counseling Series

Fetal Neck Masses

APSA
American Pediatric
Surgical Association
Saving Lifetimes

from the
Fetal Diagnosis and Treatment Committee

of the
American Pediatric Surgical Association

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Differential Diagnosis

The majority of these lesions will be comprised of either cervical teratoma, lymphatic malformation or other vascular malformations. Significantly more rare considerations include thymic cysts, intestinal duplication cysts or congenital neuroblastoma.

Initial evaluation

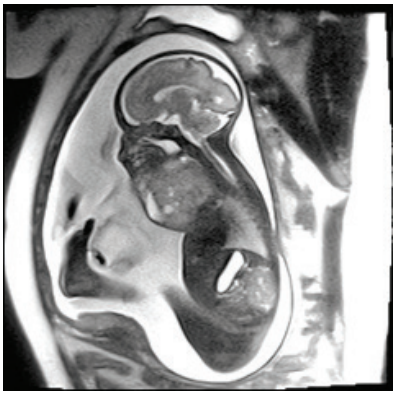
Obstetrical Ultrasound
Fetal magnetic resonance imaging

Obstetrical Ultrasound

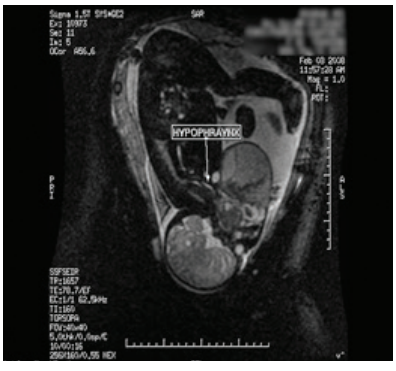
- For anatomical position, growth characteristics and presence of polyhydramnios
- Document whether the lesion is solid or cystic and presence or absence of calcifications
- Document presence of normal for gestational age stomach and lung anatomy
- Document presence of hydrops: pleural effusion, ascites, pericardial effusion, skin and scalp edema, placentamegaly

Fetal MRI

- Delineate anatomy
- For solid masses, measure the tracheoesophageal displacement index (TEDI)
- For lesions consistent with lymphatic malformations, evaluate for involvement of the tongue, aryepiglottic folds and larynx



A 29-week fetus with a rapidly enlarging neck mass consistent with a cervical teratoma. Associated polyhydramnios is noted. The neck mass is solid and causing deviation of the cervical trachea.
© Image courtesy of David Schindel, MD



A 30-week fetus with a neck mass consistent with a lymphatic malformation. Associated polyhydramnios is appreciated. The lesion has both solid and cystic components involving the upper airway
© Image courtesy of David Schindel, MD

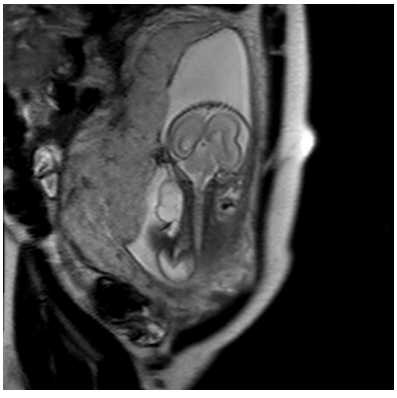
Fetal MRI Lymphatic Malformation Staging System

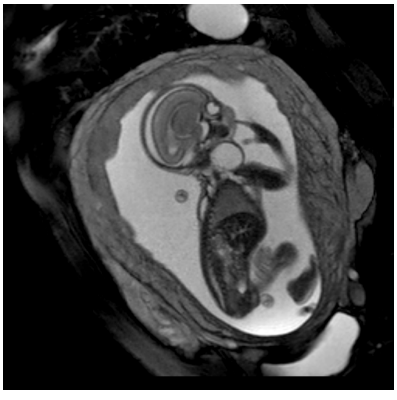
Stage I: No evidence of polyhydramnios with free egress of amniotic fluid and clear visualization of the aryepiglottic folds and larynx

Stage II: Lesions of the tongue or epiglottis present but with normal aryepiglottic folds without polyhydramnios

Stage III: Lesions of the tongue or larynx; no visualization of the aryepiglottic folds without free egress of amniotic fluid along with polyhydramnios

Stage III: Lesions are associated with lesions at risk for airway compromise at birth.





Coronal and Sagittal MRI images of a 22-week-gestation fetus that demonstrate a mildly complex cystic mass with internal septations centered within the right neck that extends centrally adjacent to the airway and inferiorly into the thorax. This is concsistent with a lymphatic malformation.

Image courtesy of Jill Stein, MD – Colorado Fetal Care Center – Children's Hospital Colorado

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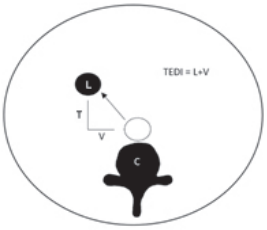
The Tracheoesophageal Displacement Index

- The severity of the fetal neck mass is defined by the measurement of the tracheoesophageal displacement index (TEDI). TEDI is defined as the sum of the lateral (L) and ventral (V) displacements of the trachea and esophagus (T) from the ventral aspect of the cervical spine (C) on fetal magnetic resonance imaging.
- TEDI = L+V
- A TEDI score >12mm indicates a complicated airway



Source: Lazar DA; Cassady, CI, Olutoye OO, et al: Tracheoesophageal displacement index and predictors of airway obstruction for fetuses with neck masses. J Pediatr Surg 2012. Jan.47 (1):46-50

Prenatal Considerations

Often diagnosed on a prenatal screening ultrasound

Prenatal natural history characterized by progressive growth that may be rapid in the third trimester

Prenatal Counseling

- **Low-risk lesions** (TEDI <12; MRI stage I; no evidence of polyhydramnios)
  - Ultrasounds weekly to assess for rapid enlargement or development of polyhydramnios
  - Expectant management in low-risk cases
  - Delivery at a tertiary center with pediatric surgical expertise having EXIT capability and specialized neonatal care is recommended

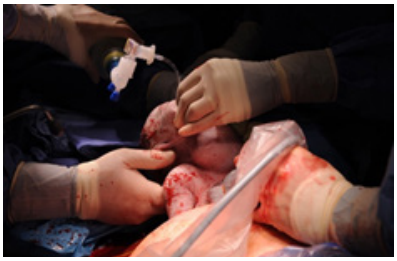
- **High-risk lesions** (TEDI >12mm, MRI stage II, teratoma pathology, and polyhydramnios all correlated with a complicated airway at birth)
  - Referral to a fetal center is recommended If hydrops is present, fetus should be delivered via cesarean section when sufficiently mature (>28 weeks' gestation) otherwise if less than 28 weeks, fetal surgery and resection should be considered.
  - EXIT-to-airway or EXIT-to-resection procedure – offered to viable fetuses with complicated airways

The EXIT (ex utero intrapartum treatment) procedure

Controlled uterine hypotonia preserving uteroplacental gas exchange thereby facilitating fetal airway intervention via a hysterotomy. The goal of these escalating interventions is to obtain an appropriate airway prior to separation from the uteroplacental circulation and subsequent delivery.

EXIT-to-airway procedure

Direct laryngoscopy and endotracheal intubation is the first option for securing a fetal airway during an EXIT procedure. This may be facilitated by flexible endoscopy. In instances where the airway is not visualized, a tracheotomy is utilized to either allow retrograde intubation or placement of a tracheostomy. Once an appropriate airway is secure, the baby is delivered and uterine atony is reversed.



A 34-week-old fetus with an airway obstructing solid neck mass successfully orally intubated during an EXIT procedure.

Source: Marwan A, Crombleholme TM: The EXIT procedure: principles, pitfalls, and progress. Semin Pediatr Surg. 2006 May; 15(2):107-15

EXIT-to-resection procedure

In instances where the fetal airway cannot be visualized and access to the trachea is impeded by a mass or lesion, the EXIT procedure allows opportunity for extensive operative interventions. These interventions include reflection of a mass away from the airway or resection of an airway obstructing mass thereby allowing securing of an appropriate airway prior to delivery.

Postnatal Considerations

Even in high risk lesions, ability to achieve an appropriate airway at the time of an EXIT procedure is excellent.

Specialized NICU with appropriate pediatric surgical and neonatal/pediatric care is necessary to manage potential related airway issues and establish treatment plans.

Need for subsequent gastrostomy is common in this population.

The most common complications post-resection are hypoparathyroidism and hypothyroidism, therefore an endocrinology work up should be initiated and consultation if indicated.

If the neck mass is cervical teratoma, there is a small malignant potential. The patient should be screened for recurrence through post-operative surveillance imaging and alpha-fetoprotein levels.

A mother's ability to conceive and carry a subsequent pregnancy to full-term following an EXIT procedure has been documented to be excellent.