Course: Facial and skull cystic lesions

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Facial and skull cystic lesions

Mostly cystic masses:
- Cystic hygroma
- Vallecular cyst
- Ranula
- Epidermal scalp cyst
- Cephalocele
- Cystic heterotopic brain tissue
- Dacrocyctocele
- Choanal atresia

Mixed cystic and solid lesions:
- Hemangio-lymphangioma.

Mostly solid lesions with cystic component:
- Hemangioma
- Cystic facial teratoma
- Epignathus
- Epulis

1. Mostly cystic masses:

1.1. Facial cystic hygroma

For a more complete discussion of cystic hygroma, see nuchal cyst-Part 1.

1.1.1. Case 1: cystic hygroma, anterior.

A fetus with a large facial mass. The fetus was diagnosed with severe lymphangioma and hemangioma. Surgery was completed initially to take out a portion in the chin, with hopes that the swelling would go down. A week later a tracheostomy was performed due to the weight of the mass. The third week another surgery was done to stop some of the bleeding in the left cheek. MRI confirmed a massive lymphomatous malformation consistent with cystic hygroma within the neck with airway compromise.
1.1.2. Case 2:

The following images represent an interesting case of the Cystic hygroma diagnosed at the 34 weeks of gestation. There was a large round mass protruding to the right side of the neonate’s face. Deforming the right cheek and eye.

The Magnetic Resonance Imaging performed after delivery, described a very large cystic mass, 93x84x84 mm, located at the right side of the neck and facio-maxillary region. There was a notable thinning of the greater and lesser wings of the sphenoid bone, temporal, frontal bone and maxillary and zygomatic arch with eccentric proptosis. There was no evidence of the intraorbital or intracranial expansion. The differential diagnosis according to the MRI was lymphangioma, specifically cystic hygroma.

The neonate underwent the surgery to remove the mass. The pathologist confirmed the diagnosis of the cystic hygroma.

Images 1,2: Image 1 shows an axial view of the fetal head. Image 2 shows a round mass on the side of the head.

Image 3,4: Images show both orbits and large anechoic mass protruding on the right side of the head.
Images 5,6: 3D images of the fetal face, note the right cheek with the protruding mass.

Images 7,8: MRI images showing the cystic mass located at the facio-maxillary region.

Image 9: shows a proptosis of the right eye.
Images 10,11,12,13: a neonate with a large mass deforming the right side of the face before and after surgery.

Discussion

Cystic hygromas result from the blockage of the lymphatic vessels. They are located in the regions which contain lymphatic tissue. Most common location for cystic hygroma is a neck region, head and axilla. The location of the cystic hygroma at the presented case is a very unusual because there are no lymphatic vessels in the central nervous system or meninges. Lymphatic drainage of the brain is via perivascular space along the basal membranes in the walls of capillaries and cerebral arteries. Lymphatics are present within the nasal cavity, orbits, around the jugular foramen and leptomeningeal sheaths around cranial nerves. One of the above mentioned locations maybe on origin of the cystic hygroma in the presented case.
1.2. **Vallecular cyst:**
Adapted and updated from Fabrice Cuillier, MD

1.2.1. **Synonyms:** mucous retention cyst, epiglottic cyst, base-of-the-tongue cyst, congenital cyst and ductal cyst. This later name originates from the classification of DeSanto et al, in which they grouped laryngeal cysts according to their location and surface mucosa (Thyroid cartilage foraminal cysts; Saccular cysts; Ductal cysts). This classification has become popular but has the limitation that, it was largely based on observation in adults and did not recognize different anatomical sites and variations in clinical presentation.

1.2.2. **Definition** Vallecular cyst is a unilocular cystic mass of variable size arising from the lingual surface of the epiglottis and containing clear, non-infected fluid.

1.2.3. **Prevalence:** Not precisely known

Laryngeal cysts are uncommon in children. Any epiglottic cyst is the most common laryngeal cyst in children, followed by vallecular cyst (VC), ventricular cyst and subglottic cyst. Although these cysts are benign, they may cause serious airway obstruction and even death if not treated appropriately. A mortality rate of about 40% in children with laryngeal cysts was reported. Vallecular cyst or pre-epiglottic cyst is a rare but recognized cause of respiratory distress in infancy or immediately after birth. It has been associated with sudden airway obstruction resulting in death.

1.2.4. **Etiology:** Sporadic.

1.2.5. **Pathogenesis:**

There are several theories for the pathogenesis of vallecular cyst. These lesions most likely result from obstruction of mucous gland at the base of the tongue. The vallecular cyst are lined with mucous glands and with continued mucous secretion, the lesion gradually increases in size. Other potential causes of cyst include embryological malformation (angiomatous, lymphatic malformation, ...). Vallecular cyst is considered to be more common in adults than children. But vallecular cyst with respiratory epithelium are rare.

When the embryo is approximately three weeks old, the primordium of the respiratory system appears as an endodermal outgrowth from the ventral wall of the foregut immediately caudal to the hypobranchial eminence. The ventrally placed respiratory primordium then becomes separated from the dorsal portion, the esophagus, except at the entrance to the larynx, where it maintains its communication with the foregut through the laryngeal orifice. The remaining part of the gut contributes to the developement of the stomach and duodenum, just caudal to the liver. Because of the proximity of the primitive gut and the pharyngeal

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arches, which contain the developing tongue, embryonal rests of the gut may become misplaced in the developing tongue. It is believed that these epithelial remnants may subsequently contribute to the development of cystic lesions of the tongue.4,5,6 Constantinides et al have suggested that the presence of ciliated epithelium, which is the hallmark of the primitive foregut, warrants use of the term «intralingual cyst of foregut origin»5.

1.2.6. Associated anomalies: Associated anomalies are rare.

1.2.7. Pronosis: Due to the anatomical location of the cyst, an infant with vallecular cyst is at risk of sudden airway obstruction and death1.

1.2.8. Complications:
- Polyhydramnios.
- Airway obstruction.
- Pulmonary hypoplasia.
- Trachea, cervical vessels and hypoglossal nerve compression.

1.2.9. Differential diagnosis: The differential diagnosis of lesions in the vallecular region includes epidermoid cyst, dermoid cyst, thyroglossal duct cyst, mucocele, enteric duplication cyst, lingual bronchogenic cyst, hemangioma, cystic hygroma, teratoma, hamartoma, lymphangioma and thyroid remnant cyst.1,4,5
- Hemangioma: The diagnosis of fetal neck hemangioma is made by ultrasonography as a translucent mass, in which pulsating Doppler flow signals is detected. MRI is recommended to plan perinatal treatment and particularly for obtaining precise information about tumor and adjacent organ. Nevertheless this type of mesenchymal tumor has never been reported to involve the tongue in a fetus.8
- Lingual lymphangioma: The most common vascular malformation affecting the pediatric airway is the lymphatic or lymphatic-venous malformation. These lesions can be divided into three subtypes based on the size of the predominating lymphatic spaces (capillary, cavernous) and the most common type cystic hygroma.9 This one is the most difficult differential diagnosis, due to similarity in size, gross characteristics and often location. The margins are usually less well defined and most of these tumors arise in the posterior triangle of the neck. Nevertheless ultrasound scanning mostly reports a thin-walled multicystic multiseptate hypoechoic mass whose size varies from small collections of fluid to enormous cysts10.

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• Thyroglossal duct cyst and brachial cleft cyst: have a similar echo-texture to lymphangioma, but are usually located below the mandible in the cervical region.

• Cervical meningocele and anterior cephalocele: are easily eliminated, because of the anterior localization of the cysts observed on the sagittal view.

• Congenital epulis: or congenital gingival granular cell tumor, is an uncommon benign intraoral tumor. Seven cases of antenatal detection on late second or third-trimester ultrasound have been reported. The tumor may be single or multiple and is usually found on the maxillary alveolar ridge or the mandible.

• Congenital ranula: or retention salivary, normally seen in children, results from a dilatation of the sublingual or submaxillary gland ducts in the floor of the mouth. These pseudocysts are normally located in the sublingual space between the mylohyoid muscle and the lingual mucosa. They can be classified according to their localization, simple ranulas (in the floor of the mouth), cervical ranulas (in the paracervical area) and plunging ranulas near the superior airway. They can extend into the floor of the mouth and the only way to differentiate them from other types is by histopathology. Ultrasonographically they are indistinguishable.

• Mucoceles: In the oral cavity, mucoceles refer to collections of mucus resulting from obstruction of, or leakage from a major or minor salivary gland. Mucoceles occur commonly in the oral cavity, typically on the lower tip but also in unusual locations, such as in young patients.

• Heterotopic gastrointestinal cyst: Intra-oral duplication cysts of the alimentary tract are rare. They tend to occur in the floor of the mouth or within the tongue. In large series, only 1.8 % of alimentary tract duplications were noted to be in the cervical region. They are usually diagnosed in the newborn period but may be undetected for years if asymptomatic and small. In fact the most difficult differential diagnosis is with heterotopic gastrointestinal cysts of the oral cavity, which involve the tongue and floor of the mouth in 97 %. However prenatal diagnosis of such lesions has never been reported. They tend to occur in the floor of the mouth or within the tongue.

• Cervical-facial teratoma: About only 5 % of all teratoma occur in the face and neck region, with anterior and lateral neck region being most

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common\textsuperscript{18}. When the teratoma arises from the sphenoid bone, the palate or the pharynx, it is called an epignathus, which appears as a complex solid-cystic tumor with mixed areas of hypo and hyper-echogenicity\textsuperscript{9}. It found in association with polyhydramnios. Protrusion through the mouth can distort the oro-facial anatomy and cause respiratory distress at birth\textsuperscript{19,20}.

1.2.10. Sonographic findings: In the second trimester, ultrasonography in this area may be more informative because the fetal head is not yet in a flexed position and imaging of the head can usually be carried out from several directions\textsuperscript{21}. There were no flow voids on either image or extensions of the mass into the surrounding soft tissue. The thyroid scan can be made and demonstrated no functioning thyroid tissue at the base of the tongue\textsuperscript{22}. Transabdominal sagittal imaging, which would be particularly useful for examination of the nasopharynx, and laryngeal area, is often impossible by ultrasound in late pregnancy, because the fetal head is deeply engaged in a transverse position in the maternal pelvis. A karyotype study is not absolutely necessary.

1.2.11. Magnetic resonance imaging (MRI): Even though obstetric ultrasonography currently has a high-resolution capacity, acoustic shadowing disturbs visualization of structures that are surrounded by bony material\textsuperscript{21}. Otherwise in late pregnancy in particular, in the region of the fetal pharynx, neck and posterior fossa, interpretation of ultrasonographic images is sometimes equivocal. Therefore MRI is a valuable additional imaging method both to diagnose and to exclude malformations and also to image the fetal anatomy adjacent to a malformation. Computed tomography and or MRI can be used to determine the extent of the cyst. The patient’s young age, the sharp margination of the lesion, the lack of surrounding tissues edema and a homogeneous T2 signal favors a benign diagnosis. Although greatly aided by imaging studies, visualization by direct laryngoscopy is still the major diagnosis tool in the diagnosis of vallecular cyst\textsuperscript{22}.

1.2.12. Implications for targeted examinations:

In cases of polyhydramnios, pharynx and neck should be investigated thoroughly in order to identify lesions that could cause obstructions labor and neonatal ventilations problems. Prenatally it possible to define the anatomical relationship of cysts to the adjacent tissues and thus the planning of postnatal surgery became easier. Therefore, a chance to localize obstruction or verify the

patency of the pharyngeal area prenatally by ultrasound or by MRI is important. Nevertheless, three dimensional ultrasonography and harmonic ultrasound imaging may also provide better possibilities to visualize the anatomy of the fetal pharynx, neck.

1.2.13. Prognosis:
Depends on the size of the tumor and the tracheal compression. Most infants can die immediately without perinatal management.

1.2.14. Recurrence risk: none

1.2.15. Management:
The widespread use of prenatal ultrasound can lead to earlier diagnosis of vallecular cyst and allows for appropriate counseling and preparation at delivery and for the proper preparation of personal and equipment in the management of these neonates. Prenatal ultrasound can detect the presence of a fetal neck mass and allows the obstetrician to collaborate with the neonatologist, pediatric surgeon and pediatric otolaryngologist in order to plan for perinatal management. So that the explications are made for parents and the time and place of delivery can be addressed and planning for resuscitative efforts can be organized in advance. If the airway is quickly stabilized and resection of the tumor is not delayed, the prognosis is good. If the airway is compromised, aspiration of the cyst can be done to improve the access to the oropharynx for intubation. Preparation for emergency tracheostomy should be done before delivery in case oral intubation is not possible. Once extent of the lesion is identified, complete cyst excision is the treatment of choice. Indeed, aspiration of the cyst does not resolve the problem and recurrence is likely.

During the birth, the placenta cord can be left undivided until the airway is secured. So perinatal management might require complicated procedures such as OOPS (Operation on placental support), in which the neonate undergoes surgery during an elective Cesarean section under tocolysis while its oxygen supply is still maintained through the placenta.

Marsupialization, which is the treatment for the vallecular cyst, may be performed with a carbon dioxide laser or an electrotcoagulation unit. In the literature, there were no long-term recurrences in any of the patients.

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23 Myer C.M.- Vallear ctal cyst in the newborn. Ear Nose Throat J 1988; 67: 122-4
1.2.16. **Case report:**

The margins were distinct and no echoes could be seen inside the mass. Color Doppler studies of the fetal neck mass showed no neovascularity, either inside or outside the cyst. While we were studying the buccal mass, movement of the fetal mouth was not impaired and the fetus swallows normally some amniotic fluid. This caused the mass to be pulled towards the retropharynx and demonstrated that the walls of the mass were soft and malleable. There was no dilatation of the hypopharynx. Nevertheless the presence of polyhydramnios and of a little filled stomach led us to conclude that the cystic mass was not obstructing totally the esophagus. The same conclusion could not be drawn for the trachea, however and so the possibility of upper airway obstruction had to be taken into account. Antenatal aspiration of the cyst was not attempted in utero. After discussions (about fetal risk) with the patient, we decided to perform serial ultrasound examinations at weekly intervals to follow the growth of the tumor.

At 28 week and three days, the amniotic fluid was on excess. An amniocentesis was necessary (2.6 l). The genetic studies reported a normal female chromosome study (46, XX) and normal amniotic fluid acetylcholinesterase.

By 30 weeks, the mass has the same dimensions, but there was a further increased amniotic fluid and suggestions of obstruction of the hypopharynx. There was a significant uterine activity and maternal betamethasone therapy was begun at 32 weeks due to the high risk for premature delivery. A repeat amniocentesis was done to relieve the patient (2.4 l).
Fig 6, 7: Magnetic resonance imaging (MRI) demonstrated the mass to be a 3 x 3 x 3 cm sublingual cystic lesion with no attachment to the hyoid bone and salivary glands.

At 33 weeks a conference including obstetricians, perinatologists, pediatrics otolaryngologists, and pediatric surgeons was convened and a cesarean section was programmed to relieve the patient. The otolaryngologist specialized in airway management, were to be ready with all the equipment necessary to perform immediate tracheostomy if the airway was compromised. Two neonatologists were present in the delivery room, to assist in resuscitation of the baby and provision of vascular access. The mass occupied the entire oral cavity without enlargement of the bony structures. The baby could not cry. So after cesarean section, the cyst was partly drained with a syringe and decompressed. The baby was orally intubated in less than three min from delivery. The airway was stable with intubation. The cord pH was 7.38. Birth weight was 1850 g. Nevertheless, the baby received surfactant. Her lungs were very noncompliant and required high-pressure settings on the ventilator during 14 days. This was thought to be secondary to the mass preventing alveolar expansion in utero. When attempting to wean her off of mechanical ventilation, the airway would collapse. Postoperative complications arrived at days 6 with respiratory distress. A multicystic leukomalacia periventricular hemorrhage was noticed at six weeks. The aspirated fluid for amylase, LDH and cell count were normal. The fluid contained ciliated epithelium. Surgical excision was performed on day ten of life (marsupialization), using nasotracheal intubation. The baby was kept intubated for 30 days postoperatively and subsequently extubated with difficulties. The patient, oxygeno-dependant, was discharged from hospital after 38 days. Neurologic forecast is reserved.

1.3. **Ranula:**

Adapted and updated from Chaitali Shah, MD.

1.3.1. **Synonyms:** Retention cyst, mucocele, oral pseudocyst.

1.3.2. **Definition:**

A congenital ranula is a cystic malformation seen in the oral cavity. A ranula usually results from the obstruction of the sublingual or minor salivary glands. Clinically a fluid-filled structure fills the mouth and is seen to elevate the tongue. These
pseudocysts are normally located in the sub-lingual space between the mylohyoid muscle and the lingual mucosa.

1.3.3. Prevalence: The incidence of a congenital ranula is estimated to be 0.74%.

1.3.4. Etiopathogenesis:
A ranula is a fluid collection that occurs either due to:
- Disruption of minor salivary ducts leading to extravasation of mucous structures into adjacent structure and resulting in a mucous extravasation cyst. These are more common in children and young adults and rarely occur in neonates. The ranula is not lined by an epithelium in this case.
- A blocked duct causing proximal expansion and resulting in a mucous retention cyst seen in neonates. The fluid collection is lined by salivary duct epithelium.

1.3.5. Types: Ranulas can be classified according to their site of location. They can be
- A simple ranula – located in the floor of the mouth
- A cervical ranula – located in the paracervical region
- A plunging ranula – located near the upper airway and extending into the floor of the mouth. [plunging ranulas exhibit a so called ‘tail sign’ on MRI]

1.3.6. Sonographic findings:
A hypoechoic cystic mass in the floor of the mouth, with no solid components. If very large, the mass may displace the tongue upwards. No vascularity can be seen within this cystic structure.

1.3.7. Implications for targeted examinations:
Once an oral tumor is identified, it is necessary to rule out other associated anomalies. If no other abnormalities are noted, then adequate monitoring of the growth of the intra-oral mass must be done. If the mass becomes too large, it may interfere with the swallowing mechanism in the fetus, resulting in polyhydramnios. If it is a cystic mass, decompression may be carried out via percutaneous cyst aspiration.

1.3.8. Differential diagnosis:
The large mass arising from the mouth could represent
- Epignathus – This is an oropharyngeal teratoma mostly arising from the palate. It would be more solid and the roof of the mouth would be abnormal. It appears as solid-cystic tumor with mixed areas of hypo and hyperechogenicity,

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may have calcifications and is found in association with polyhydramnios. It can cause significant morbidity and mortality.

- An epulis (a solid granular cell tumor usually from the gingiva): this is a rare, benign granular cell tumor which is solid. It arises from the alveolar ridge. Color and power Doppler usually demonstrates marked blood flow in the tumor. It is a self-limiting lesion and responds to conservative excision.
- An oral cephalocele (but the intracranial anatomy would be abnormal)

Other oral tumors arising in a newborn are very rare. The following tumors must be considered:

- Foregut duplication cyst: this rare enteric duplication cyst may occur in the floor of the mouth. It is cystic in nature and may closely mimic a ranula.
- Median palatal mucosal cyst [Epstein's pearl]: this is benign and self-resolving.
- Gingival cyst of the newborn [dental lamina cyst]: this is benign and self-resolving.
- Vascular hamartomas: these may be hemangiomas or lymphangiomas [cystic hygromas] and are usually located on the tongue. These tumors may appear as solid-cystic masses on ultrasound and require surgical excision.

Other tumors of the tongue include:

- Thyroglossal duct cyst
- Lingual thyroid
- Dermoid cyst
- Granular cell myoblastoma and
- Heterotopic gastric mucosal cyst.

1.3.9. Complications: A ranula or any oral tumor if large enough can cause obstruction of the airway or feeding problems.

1.3.10. Prognosis: Excellent prognosis. Even if the tumor is large, an airway can be established via the EXIT procedure; and the tumor can be excised at a later stage.

1.3.11. Recurrence risk: If properly excised, a ranula does not recur.

1.3.12. Management:

Ex-utero intrapartum treatment [EXIT procedure] can be used to obtain a fetal airway in cases of large obstructing masses that may potentially obstruct the airway and cause respiratory distress. This procedure involves establishing an airway before the feto-maternal circulation is interrupted.

The definitive treatment of a ranula is surgical,\textsuperscript{32} and according to a recent study by Haberal, et al either marsupialization or excision of the ranula may be performed with equally successful results. The commonest complication of surgery that they found was rupture of the ranula during the operation, which if occurred, appeared to cause no increased risk of recurrence.

1.3.13. Case report:

1.3.13.1. Case 1: This is a ranula, a sublingual cyst that results from an imperforate submandibular duct. Treatments include marsupialization or total excision.

1.3.13.2. Case 2: In second trimester

The large mass arising from the mouth could represent

- an epignathus (but it would be more solid and the roof of the mouth would be abnormal)
- a epulis (a granular cell tumor usually from the gingiva)
- a oral cephalocele (but the intracranial anatomy was too normal)
- a Ranula which is a obstructed salivary duct

This was indeed a ranula.

1.3.13.3. **Case 3**

A case of a 30-year-old woman, G6 P5, who referred to our antenatal unit at 35 weeks of gestation due to a lingual cyst.

Ultrasound follow-up in 1 week did not show any growth of the mass. No intracervical extension was noted. The fetal MRI detected the same findings. The patient delivered in 39 weeks of gestation and the neonate suffered from respiratory distress after delivery.

Images 1, 2: 35 weeks, Image 1 shows an opened mouth filled with mostly anechoic cystic mass. Image shows the cystic mass measuring 33 x 39 mm.

![Images 1, 2](image1.png)

Images 3, 4: Color Doppler demonstrates the flow of the amniotic fluid through the nasal cavity excluding the obstruction of the nasal cavity.

![Images 3, 4](image2.png)
Images 5-6: 36 weeks, 3D-images showing the opened mouth and oral cavity filled with cystic mass.

Images 7-10: MRI images, cyst (white arrow) fills the entire oral cavity.

1.4. Epidermal scalp cyst:
   Also see nuchal cyst – part 1

1.4.1. Case report:

A 19-year-old G2P1 presented for routine anomaly scan. The patient had a normal tri-screen result. She had one previous child with isolated choroid plexus cysts and no other clinical history of note. A cystic extracranial mass was noted over the anterior fontanelle measuring 10 mm. A detailed scan of the rest of the fetus was unremarkable. The diagnosis of an epidermal cyst was therefore suspected. The fetus was scanned again at 18, 22 and 37 weeks
with no change in appearance of the mass. A spontaneous vaginal delivery of a normal female infant was performed at 39 weeks.

Several views of the cyst. Note the lack of anomalies in the brain and the lack of connection to the brain or ventricles. A thin echogenic membrane was seen at the base of the mass thought to represent the periosteum or soft tissue under the mass. There was no intracranial extension of the mass. The mass was mobile and not attached to underlying structures. No defect was seen in the calvarium, and the intracranial anatomy was unremarkable.

Images 1-8: at 17 weeks gestation
Postnatal clinical examination showed the cystic lesion of the scalp. Ultrasonic assessment at 1 month of age confirmed the cystic lesion.

**Image 9-10:** Transverse views (note the interhemispheric fissure). Also in the energy and velocity Doppler image note flow in the superior sagittal sinus.

**Image 11-12:** The cute little girl:

1.5. **Cranial meningocele:**
Discussion and characteristic: see nuchal cyst – Part 1

1.5.1. **Case report:**
The meningocele originating from the skull with no brain matter inside, imitated septations in amniotic fluid. Caesarean section was done and postnatal examination confirmed the diagnosis. Here are some of the images we obtained.
Images 1, 2. 36 week of gestation - a 2D transverse scan through the skull showing a big cystic structure dorsally to the fetal head - meningocele (left); and a 2D transverse plane through the meningocele with a thin septum inside (right).

Images 3, 4. 36 week of gestation - a 2D sagittal scan through the fetal spine showing a meningocele dorsally to the spine, with the septation inside, imitating septums inside the amniotic cavity (left); and a 3D image showing the sac of meningocele next to the fetal spine (right).

Images 5, 6. Postnatal appearance of the baby with the big dorsal cranial meningocele.
1.6. Cystic Heterotopic Brain Tissue:
Adapted and updated from Laurie Briare, RDMS

1.6.1. Synonyms: Brain heterotopia, cystic temporofacial brain heterotopia.

1.6.2. Definition: Heterotopic tissue consists of histologically normal tissue that is situated in an abnormal location.

1.6.3. Prevalence: Fewer than 20 cases of large heterotopic brain tissue in the neck region have been reported.33,34 Brain heterotopia is a rare developmental anomaly that has been reported to occur in the pharynx34, orbit35, lung38, palate36, nasal cavity and maxillofacial region37,38.

1.6.4. Etiology and pathogenesis: The etiology of cystic masses of heterotopic brain tissue is thought to be attributable to the early displacement of pluripotential cells and cyst formation may result from cerebrospinal fluid production by contained choroid plexus39. Other theories exist regarding the origin of heterotopia. One such theory suggests that it is derived from an encephalocele that has lost its connection to the subarachnoid space. Another theory hypothesizes that neuroectodermal tissue develops from multipotent cells during embryogenesis40.

1.6.5. Differential diagnosis: Cystic hygromas34, encephaloceles, meningoceles, teratomas and brachial cleft cysts,39 lymphangioma.41

1.6.6. Sonographic findings: Fluid-accumulated cyst with irregular inner contour, no color Doppler revealed in side the mass.

1.6.7. Prognosis: This is a benign mass, however, they can compress and deform surrounding structures and cause airway obstruction in the newborn.41,42 Prognosis is good with antenatal intervention and postnatal surgical excision for extracranial lesions.

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1.6.8. **Treatment:** excision/resection is indicated. Drainage of the cyst results in reaccumulation of fluid. 40

1.6.9. **Follow up:** Plastic surgery, gastroenterology, neurology, ophthalmology and cardiorespiratory. Complete surgical follow up arranged after discharge from the neonatal unit.

1.6.10. **Case report**

At 21 weeks of pregnancy, a large cystic structure measuring 79 x 87 mm was revealed to arise from the left mandibular/auricular area with thick walls, no septation, echogenic foci at the margins, which most likely represented calcifications. Color flow Doppler did not reveal any blood flow within the mass. At the level of the cervical spine, the midline soft tissue structures were displaced toward the right side. The mass appeared separate from the thyroid or esophagus. No dilatation of the esophagus was present. Polyhydramnios was identified with an amniotic fluid index of 243 mm.

Two weeks later, the mass increased to 100 x 95 mm and the amniotic fluid index increased to 316 mm. An amniocentesis, amniotic fluid drainage and fetal neck mass aspiration were then performed. The karyotype revealed 46XY, male chromosome complement, 650 cc’s of amniotic fluid was drained and 350 cc’s of thick brown fluid was aspirated from the neck mass with a sample sent for cytology and chemistry. The mass measured 133 mm after the drainage. The post-procedure amniotic fluid index was 170 mm. Pathology results reported the fluid aspirated from the cyst to contain fetal red blood cells and mononuclear cells-both histocytes and lymphocytes. Cystic hygroma was suggested as a differential diagnosis.
Within one week the mass had increased to its pre-drainage size and the amniotic fluid index had increased to 259 mm. The patient was placed on Indomethacin for treatment of polyhydramnios.

At 25 weeks, a second amniotic fluid drainage was performed with 795 cc’s. A shunt was placed into the neck mass and 445 cc’s of fluid was aspirated from the cyst. A pigtail catheter was inserted into the cyst to allow continuous drainage.

Weekly ultrasound monitoring continued and by 29 weeks gestation a third amniotic fluid drainage of 840 cc’s was performed. The mass had again enlarged and 300 cc’s of fluid was drained. At 30 weeks 4 days gestation, a second shunt was placed in the mass since it had again enlarged, and the first shunt was assumed not to drain. The mass measured 110 x 100 x 90 mm before the drainage and shunt placement, and 83 x 100 x 105 after the procedure. A fourth amniotic fluid drainage of 1200 cc’s was also performed. Weekly ultrasound monitoring of amniotic fluid, mass size and Doppler of ductus arteriosus continued. After one week the amniotic fluid index remained normal and the mass was unchanged in size. One week later, the amniotic fluid index was normal, but the shunt was seen completely within the mass.

At 33 weeks 5 days the patient was admitted to the hospital with preterm labor and spontaneous rupture of membranes. Fetal lung maturity amniocentesis was negative. Patient was placed on tocolytics for another week. At 34 weeks 5 days a repeat lung maturity amniocentesis was performed and 600 cc’s of fluid was drained from the mass. Because the foam stability index was 48 (mature) and the fetus developed recurrent...
late heart rate decelerations, a cesarean section was performed and a 2.77 kg baby boy was delivered.

Before and after operation

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Management: Prenatally, percutaneous drainage of the mass was performed five times to facilitate fetal swallowing. Shunt placements were attempted on two separate occasions but were not successful. Amniotic fluid drainage was performed on four occasions to treat polyhydramnios, which was causing preterm labor. Indomethacin was also used in an attempt to treat the polyhydramnios. Cystic hygroma was suggested as a differential.

Postnatally: Two days after delivery the mass had to be drained to relieve airway obstruction. Computerized tomography and magnetic resonance imaging scans showed a complex cystic mass of the left face and neck with some compression of the oropharyngeal airway. On day 9 of life, the child went to surgery and the mass was excised along with the left parotid gland and submandibular gland. The pathology report defined the mass as heterotopic brain tissue, and was confirmed by Children’s Hospital of Philadelphia and UCLA.

Postoperative condition: Paresis of the left facial nerve distribution, both upper and lower components, resultant in inability to nipple feed and lagophthalmos. Gastrostomy tube was placed on day 37. The baby had difficulty tolerating extubation and bronchoscopy was performed on day 37, which showed an abnormally shaped and deficient cartilage formation of the epiglottis. Because of the tenuous airway, a tracheostomy was performed. The left ear was somewhat deformed externally with partial occlusion of the external auditory canal. The brain appeared normal on computerized tomography scan and other than the facial nerve paresis, the baby had a fairly normal neurologic exam upon discharge.

Many differential diagnosis have been suggested in this case including:

- myoblastoma, sarcoma
- neck teratoma
- cephalocele
- brachial cyst
- epulis, ranula
- dacyrocystocele
- gingival tumors
1.7. Dacryocystocele:
Adapted and updated from Andrea Tardiff, RDMS.

1.7.1. Synonyms: Dacryoma, amniotocele, amniocele, lacrimal sac mucocele.

1.7.2. Prevalence: approximately 50 case reports literaturely.

1.7.3. Definition: Cystic dilatation of the lacrimal sac at the nasocanthal angle.

1.7.4. Etiology: Unknown.
This is a rare clinical condition.

1.7.5. Embryology
In the 7 mm embryo, a thickened ridge followed by an infolding of the ectoderm occurs at the naso-optic fissure (fig. 1). The ectoderm sinks into the mesoderm until it forms an ectodermal epithelial cord in the 12 mm embryo. This column is oriented from the medial canthus to the anterior third of the inferior turbinate. As the column extends, it bifurcates in its cranial portion (fig. 2 left). The two outpouchings develop and become the superior and inferior canaliculi (fig. 2 right).

Fig. 1: In the 7 mm embryo, a thickened ridge followed by an infolding of the ectoderm occurs at the naso-optic fissure.

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Fig. 2: The ectodermal epithelial cord is oriented from the medial canthus to the anterior third of the inferior turbinate. As the column extends, it bifurcates in its cranial portion to become the superior and inferior canaliculi and canalize from the most cephalic part downwards.

Fig. 3: The normal anatomy compared to the dacrocyctocele.

The lacrimal duct anlage vacuolize from the most cephalic part downwards towards the nasal end at the inferior turbinate. The canalization process creates the lacrimal duct during the third month. The junction between the lacrimal duct and the nasal epithelium may exist after six months, but is not always established by birth.

The intervening membrane (which will become the valve of Hasner after perforation) is responsible for the obstruction of the lacrimal duct, which is manifested in the newborn by epiphora. Since tears are not usually produced at birth, the obstruction is rarely symptomatic early, and by the time it could become symptomatic, the patency of the valve of Hasner is usually established.

The canaliculi perforate the eyelids on the ocular surface, and the opening is called the punctum. More distally, the canaliculi usually join before entering the lacrimal sac. The duct that results from the fusion of the canaliculi is called the sinus of Maier or common canaliculus, and it connects to the lacrimal sac at the valve of Rosenmüller.

1.7.6. Pathogenesis:

Complete obstruction of the lacrimal duct, inferiorly by non-permeability of the valve of Hasner, and superiorly by a valve mechanism at the valve of Rosenmüller.

When the sinus of Maier is absent, the superior and inferior canaliculi enter the lacrimal sac independently and at a hyperacute angle. This occurs in approximately 10% of normal infants and may predispose to kinking, creating a valve effect.44

An non-patent valve of Hasner (distal lacrimal drainage system) is a prerequisite for the formation of a dacrocyctocele, and may exist in up to 73% of normal term infants.45

When this valve mechanism coexists with a non-patent valve of Hasner, the stage is set for the development of a dacryocystocele: fluid that is squeezed by the blinking motion enters the lacrimal sac, but cannot exit\textsuperscript{44}. The lacrimal sac can easily expand in the medial canthal region since there is no bony or tendinous structure to directly limit its expansion.

The origin and composition of the sac contents have been discussed for years in the ophthalmic literature. This case supports the theory that the fluid in the dacryocystocele is amniotic fluid since the tear secretion normally is not significant until 3 to 4 weeks postpartum\textsuperscript{46}. However, because of the viscous nature of the sac contents, some authors have proposed that it could represent mucus produced by the intraluminal goblet cells\textsuperscript{47}.

\textbf{1.7.7. Associated anomalies:} None. The cyst may produce deformities of the inferior turbinate and even of the septum.

\textbf{1.7.8. Differential diagnosis:}
Frontonasal meningocele, potentially also hemangioma, dermoid cyst, mucocele, although these are either unlikely in the age group, or have a different echotexture.\textsuperscript{48,49,50,51}

\textbf{1.7.9. Prognosis:} Excellent.

\textbf{1.7.10. Recurrence risk:}
The large majority of infants (71\%) treated will never reaccumulate material in their lacrimal sac\textsuperscript{52}. The recurrence risk in subsequent pregnancies is unknown.

\textbf{1.7.11. Clinical presentation}
The infant encounters no clinical signs of epiphora (tearing) or dacryocystitis (infection). The only clinical finding is that of a bluish mass below the medial canthal tendon. Thus, the condition typically is not brought to the attention of a pediatrician or ophthalmologist. The dacryocystocele is typically sterile since colonization of the lacrimal drainage system is usually not established until the first several weeks of life. When infected, the most common bacteria isolated are Staphylococcus organisms\textsuperscript{48}. Resolution—as in our case—by spontaneous opening of the distal lacrimal drainage system may occur during the first few weeks of life before bacterial colonization and significant tear production commence. This may explain why this condition is not reported more frequently.

\textbf{1.7.12. Management:}
External digital pressure on the cyst may enable the contents to decompress through the nose. If this is unsuccessful, probing of the nasolacrimal duct (from the eye towards the nose) with inferior turbinate outfracturing may establish patency of the valve of Hasner\textsuperscript{49}. In

rare cases this is still inadequate, and marsupialization (dacryocystorhinostomy) of the nasal component with silicone stenting is performed.

1.7.13. Case report

1.7.13.1. Case 1:

Images 1 and 2: 2D gray scale (image 1) and color Doppler (image 2) images showing cystic structure medially to the right ocular bulb representing dacrystocele.

Images 3 and 4: 3D images showing dacrystocele medially to the right eye of the fetus.
1.7.13.2. Case 2:

Images 1 and 2: 2D gray scale images showing cystic structure medially to the left eye representing the dacryocystocele.

![Images 1 and 2](image1.png)

Images 3 and 4: 3D images showing cystic structure medially to the left eye representing the dacryocystocele.

![Images 3 and 4](image2.png)

1.7.13.3. Case 3:

This is a 3D-scan of a bilateral dacryocystocele seen at 37 weeks of gestation. The examination revealed two cystic masses located inferior-medial to the eyes. Inside the cysts, there was echogenic material inside the dacryocystocele. No other associated anomalies were seen. After birth, the face of the baby was completely normal. A clinical examination did not reveal any mass. At day 5, the cysts appeared bilateral and drained spontaneously after a few days.

Note the bilateral dacryocystocele with a echogenic material inside.
1.7.13.4. Case 4:

These are some 2D and 3D ultrasound images of dacryocystocele.

1.8. Choanal atresia:

1.8.1. Definition:

Choanal atresia is a rare congenital anomaly characterized by the narrowing one or both sides of the posterior nasal airway.
1.8.2. **Incidence:** 1.3:10,000\(^{53}\) births, or 1.2-2:10,000\(^{54}\), predominantly in female, rate is equal to 2:1\(^{54}\).

1.8.3. **Associated anomalies:** more than half of affected infants also have other congenital problems such as: Charge syndrome, Treacher Collins syndrome and Tessier syndrome.\(^{55}\), and other craniofacial syndrome-otocephaly, 9p-syndrome.

1.8.4. **Pathology:** there are 4 hypotheses: (1) the persistence of the nasobuccal membrane of Hochstetter, (2) persistence of the foregut buccopharyngeal membrane, (3) abnormal mesodermal adhesions forming in the nasal choanae, and (4) the misdirection of mesodermal flow due to local factors. Others believe that misdirection of the neural crest cells are induced by genetic mutations or environmental factors that cause defects in the palate and nose.\(^{55,53}\).

1.8.5. **Sonographic findings:** anechoic cyst in the posterior nasal region

1.8.6. **Prognosis:** respiration restriction and complication of postoperative stenosis.

1.8.7. **Management:** Intubation immediately after delivery, then surgery for correction of the stenotic area as soon as possible.

1.8.8. **Different diagnosis:**

Single nostril, severe nasal septal deviation.

1.8.9. **Case report:**

1.8.9.1. **Case 1:**

Images 1,2: Sagittal view of the fetal face, note cystic choana with atresia (arrow) and inferior nasal concha (*).

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\(^{55}\) Jodi D. Zuckerman, MD; Syboney Zapata, MD; Steven E. Sobol, MD, MSc Single-Stage Choanal Atresia Repair in the Neonate Otolaryngol Head Neck Surg. 2008;134(10):1090-1093.
Images 3,4: The axial view of the nasal cavity, arrow indicates the unilateral choanal atresia.

Images 5,6: The oropharynx, note the choanal atresia (arrow).

Images 7,8: 3D reconstruction of the CT images of the nasal cavity, choanal atresia is nicely visible (arrow).
Images 9-10: 3D CT of the fetal face, note the atresia of the right choana.

Images 11, 12: Injection of the dye into the nostrils. Image 12 shows the dye collected in oropharynx (white arrow) after passing through the patent left choana. Image 13 shows a collection of the dye around the right eye (yellow arrow) after injecting it into the right nostril. The passage via the right choana to the oropharynx (white arrow) was blocked due to the choanal atresia.
1.8.9.2. Case 2:

This is the ultrasound scan of a fetus at 15 and 20 weeks of gestation. Actually at first glance, the primary diagnosis was the differential between a single nostril and severe nasal septal deviation. The explanation to "the disappearance of the single nostril" was a mucous plug which deviated the septum laterally (usually there is no stream of fluid in the obstructed nasal cavity) and for some reasons, the plug was pumped out or had liquefied towards week 20.

Week 15: Sagittal and coronal section shows a single ballooned nostril
Week 20: normal 2 nostrils. The outcome was unilateral choanal atresia.
2. Mixed cyst:

2.1. Hemangiolympangioma:

For a more complete discussion and characteristics, also see Nuchal cyst-Part 1

2.1.1. Case report:

These are images from a mid trimester fetus with something behind the head. The images demonstrated a soft-tissue mass on the postero-lateral aspect of the head.

A cesarean section was performed near term. Pathological diagnosis confirmed of hemangioma with part composed of cavernous lymphangioma.

3. Mostly solid cyst:

3.1. Hemangioma:

Discussion and characteristics, see nuchal cyst-part 1.

Case report:

Ultrasonography revealed a 45×41×50 mm tumor originating on the left side of the skull, spreading towards the same side of the neck, behind the ear. It appeared as an echogenic soft tissue mass with the hypoechoic areas within it. Color Doppler revealed vascularization in the tumor, mainly on its border and the 4D examination showed the ear was intact. No other abnormalities were detected. Cord blood sampling was done and karyotyping was normal (46,XY).
Images 1, 2: At 23 weeks: the fetal head with the prominent mass of hemangioma.

Images 3: 3D image showing the mass of hemangioma of the fetal head.

Images 4, 5: MRI images showing hemangioma of the head.
3.2. Facial teratomas:
Adapted and updated from Marcos Antonio Velasco Sanchez, MD;

In 1940 Ewing classified these nasopharyngeal tumors as:

Dermoids - consisting of epidermal and mesodermal germ layers, attached to soft / hard palate and / or pharynx near midline. Minimal or extensive intracranial extension may be present with skin cover.

Teratomas - consisting of all 3 germ cell layers with an indifferent degree of organization. It differs from type 1 by their greater structural complexities, earlier development and larger size.

Epignathus - consisting of teratomas with high degree of organizational and recognizable structures. Formed organs are present.

Case report: cystic facial teratoma

The following images show a case of mature cystic facial teratoma (arising from the right lateral moiety of the fetal face, jaw and neck) diagnosed at 20 weeks a pregnancy.
3.3. **Epignathus:**

Adapted and updated from Elke Sleurs, MD and Luc de Catte, MD, Boris M. Petrikovsky, MD, PhD, Marcos Antonio Velasco Sanchez, MD;

3.3.1. **Synonyms:**

Oral teratoma, nasopharyngeal teratoma, extra-gonadal teratoma, facial teratoma, congenital oropharyngeal teratoma.

3.3.2. **Definition:**

A teratoma which arises from the oral cavity and/or pharynx.

3.3.3. **Incidence:**

Teratomas, being the most common neoplasms in the newborn, occur at a rate of 0.3:10,000 live births. About 2% of all pediatric teratomas occur in the nasopharyngeal area.

3.3.4. **Recurrence:**

There is no known genetic or recurrence risk or predisposing factors.

3.3.5. **Etiology:**

The midline location of most congenital teratomas has served to support the hypothesis that primordial germ cells migrate along the dorsal midline from the hindgut-yolk sac region into the embryonic genital ridge. Some cells continue their cephalad migration to eventually settle in the mediastinum, neck, nasopharynx, and brain (pineal and hypothalamic regions).

3.3.6. **Pathology:**

Most teratomas have at least one tissue type from each of the three embryonic layers. Regardless of the primary site, teratomas have several pathologic features in common. The cystic or multicystic components are usually accompanied by solid areas that often have a brain-like tissue quality. Hemorrhage is inconspicuous, with

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the exception of those tumors subjected to trauma during delivery. Tumor-related necrosis is usually minimal because these neoplasms are rarely malignant.

3.3.7. Associated anomalies:

Epignathus is mostly an isolated anomaly. Less than 10% of newborns with epignathus will have associated congenital malformations including cleft palate, hypertelorism, congenital heart anomalies, umbilical hernia and facial hemangiomas as well as Aicardi syndrome (agenesis of the corpus callosum, infantile spasms, and ocular anomalies) and Pierre Robin sequence. Facial defects have been attributed to the mechanical effects of the teratoma on developing structures. Occasionally chromosomal malformations have been reported. (Yapar et al. 1995) (trisomy 13) and Abendstein et al. (1999) (inverted duplication of chromosome, conventional karyotyping was normal).

3.3.8. Sonographic findings:

Like the other teratoma, complex heterogeneous mass arising from the fetal mouth and/or nose.

3.3.9. Differential diagnosis:

- Cystic hygromas: which are predominantly in the lateral and posterior neck regions. Unilocular or multilocular lymphatic hamartomas ranging in size from several millimeters to 80 mm, containing a clear or cloudy fluid like lymph. They are usually located at the nuchal area but they can also be seen in the axilla. They are sometimes associated with polyhydramnios.
- Congenital epulis (congenital granular cell tumors of the gingiva or congenital myoblastoma)
- Ranula
- Hemangioma: hemangioma consists of huge vascular beds, and color Doppler flow imaging could depict marked blood flow in the solid part of the mass.
- Lymphangioma:
- Thyroglossal duct cyst
- Cervical meningomyelocele,
- Neuroectodermal tumor or
- Brachial cleft cyst.

57 Isaacs H, jr: Potter’s pathology of the fetus and the infant. Chapter 28: Tumors.
3.3.10.  Prognosis

The neonatal prognosis depends on the size of the tumor and the degree of involvement of other structures. Nasopharyngeal teratomas are often associated with polyhydramnios, nonimmune fetal hydrops, and exophthalmos. Although the majority of these tumors are benign in nature, fetal and neonatal death is very common due to the local mass effect, which produces life-threatening dysfunction (basocranial teratomas) or cessation of function (respiratory compromise from a nasopharyngeal or cervicothyroidal lesions). Most cases reported in the literature revealed dismal outcome\textsuperscript{62,63,64}.

3.3.11. Management:

When the prenatal ultrasonographic diagnosis of a giant mass protruding from the mouth of the fetus is done, genetic counseling and planned obstetric management with subsequent surgery in a specialized unit must be arranged. Post delivery the umbilical cord and fetoplacental circulation must be left intact to allow oxygenation of the fetus until a rapid examination and a tracheostomy is done, if it is necessary.

After airway is secured, the umbilical cord can be clamped and the baby can be transported to the neonatal intensive care unit and subsequent surgery can be done after baby's stabilization. Before surgery, plain X-ray and computed tomography (CT) scan must be performed to rule out intracranial extension of the tumor or primary lesions of central nervous system. Radical disfiguring surgery is contraindicated in the neonate as it may result in impairment of speech and deglutition.

Even with optimal management, survival rates are not higher than 30-40%. If diagnosis is made before 24 weeks gestation termination of pregnancy should be offered.

3.3.12.  Case report:

3.3.12.1.  Case 1:

Images 1 and 2: 2D sonography at 32nd week. Images show irregular predominantly solid mass protruding out of fetal mouth.

Images 3 and 4: 2D sonography at 32nd week; Image 3 - gray scale image showing irregular solid mass with small cystic components protruding out of fetal mouth; and image 4 - color Doppler image showing vessels supplying the tumorous mass.

Images 5 and 6: 3D sonography at 32nd week; Images show tumorous mass protruding out of the fetal mouth.
Images 7 and 8: Perioperative images during the EXIT surgical resection of the epignathus protruding from the oral cavity attached to the tongue of the baby. The umbilical cord was left intact during the surgical procedure.

Images 9 and 10: Final suture of the baby’s tongue after the EXIT surgical resection of the epignathus and postnatal appearance of the baby after surgical resection of the epignathus.

Images 11: Pathological specimens showing the resected epignathus.
3.3.12.2. **Case 2:**
This fetus was scanned in the late second trimester

![Ultrasound images of Case 2](image1.png)  
©2000 Luc de Catte

3.3.12.3. **Case 3:**
The ultrasound examination revealed a singleton fetus in oblique lie. Polyhydramnios and placentomegaly were noted. The measurements of BPD, abdominal perimeter and femur length were compatible with 22 weeks of pregnancy.

**Images:** A complex mass, originating from the fetal mouth and measuring 56 x 49 x 28mm: Epignathus with cystic and solid components (between arrows).

![Ultrasound images of Case 3](image2.png)  
©1993 Boris M. Petrikovsky

The patient elected pregnancy termination. A 700g stillborn was delivered with a length of 20cm. The autopsy showed a spherical, exophytic palatine mass which protruded from the mouth and was attached to the hard palate in the midline.
3.3.12.4. **Case 4:**

Images 1, 2, 3, and 4: The images show sagittal (Images 1, 2) and coronal (Images 3, 4) scans of the fetal head with the epignathus. The images on the right represent a fusion of the images on the left with the postnatal appearance of the fetus.

Images 5, 6: 19 weeks of pregnancy; the images shows sagittal (Images 5) and oblique coronal (Image 6) scans of the fetal head with a tumorous mixed solid and cystic mass protruding out of the fetal mouth (arrows) the epignathus.
Images 7, 8, 9: The images show pathological specimens of the fetus after the termination of the pregnancy at 21st week. Massive epignathus sticks out of the fetal mouth.

3.4. Granular cell tumor (Epulis):

3.4.1. Definition:

An epulis is a gingival granular cell tumor also known as granular cell myoblastoma. It is considered a degenerative lesion of mesenchymal cells and not a true neoplasm. This is a rare benign intraoral tumor.

These are intraoral masses arising from the gingival ridge. They are well circumscribed, homogeneous and have smooth borders. Calcifications and cystic components may also be present. They increase in size as gestation progresses.

3.4.2. Sonographic findings

Ultrasound revealed a smooth mass protruding from the mouth at the level of the upper gum.

Color Doppler does not reveal any peculiar features, but is useful to differentiate them from hemangiomas. A single feeding vessel may also be identified.

The images below represent the same fetus in 2D and on MRI. Note the solid homogeneous texture of the epulis and its clear attachment to the upper gum.

An intriguing possibility is the assessment of nasal patency by observing color flow Doppler at the level of the mouth and nostril while the baby is breathing in utero. This may be academic, since the tumors are rarely obstructive and the newborn will be delivered with neonatologists/surgeons available for immediate assistance. Should that not be the case, an EXIT procedure may be planned.

3D helps define the relationships of the mass to surrounding structures. As for all facial lesions, it helps the parents conceptualize the extent of the lesion.

The lesion characteristically appearing as a “chewing gum bubble” on 3D rendering is epulis. Note the very smooth outline; the “cystic” appearance on the image is an artifact of rendering. The lesion is solid. If it were cystic, think more of a “ranula,” which we will review later. On 2D sections, the solid nature of the epulis is clear.
Patency of the upper airways is better demonstrated on MRI than on ultrasound. Typically, epulis is an isolated anomaly without associated abnormalities.

This is the same fetus in 2D and on MRI. Note the solid homogeneous texture of the epulis and its clear attachment to the upper gum. Patency of the upper airways is better demonstrated on MRI than on ultrasound. Typically, epulis is an isolated anomaly without associated abnormalities.

3.4.3. Prognosis:
Complications result from obstruction to the mouth and upper airways causing polyhydramnios from impaired deglutition, and neonatal respiratory compromise. If intracranial extension is seen, consider an epignathus instead.

3.4.4. Management:
Delivery by C-section helps reduce disruption of the mass and can be coupled with ex utero intrapartum treatment (EXIT) when there is suspicion of airway obstruction. Surgical resection of the epulis is performed right after delivery.

3.4.5. Histology:
Histologically, epulis consists of large cells with eosinophilic cytoplasm, within a network of vascular channels and dense fibrous connective tissue.

3.4.6. Case report:
Scan performed at 31 weeks
Scan at 32 weeks

Postnatal images
Histology:
<table>
<thead>
<tr>
<th>Anomaly</th>
<th>Vascularization</th>
<th>Single/multi cystic</th>
<th>Solid</th>
<th>Calcification</th>
<th>Special features</th>
<th>Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemangioma</td>
<td>++</td>
<td>+</td>
<td>+++</td>
<td>+/-</td>
<td>Solid mass with echotexture similar to placenta, huge vascular beds</td>
<td>Head, limbs, skin, abdomen</td>
</tr>
<tr>
<td>Cystic hygroma</td>
<td>-</td>
<td>+++</td>
<td>-</td>
<td>-</td>
<td>Often septate and multilocular</td>
<td>Posterior neck commonly, others: anterior neck, cheek, axilla, mediastinum, chest wall, mesentery…</td>
</tr>
<tr>
<td>Hemangiolympangiomas</td>
<td>+</td>
<td>++</td>
<td>++</td>
<td>-</td>
<td>a rapid growth tumor confirmed after birth</td>
<td>Axilla, abdomen cavity, limb, bladder</td>
</tr>
<tr>
<td>Epulis</td>
<td>+/-</td>
<td>++</td>
<td>-</td>
<td>-</td>
<td>Smooth, well-derlineated, homogenous solid mass</td>
<td>Anterior face (usually from the gingiva)</td>
</tr>
<tr>
<td>Cystic facial teratoma</td>
<td>+/-</td>
<td>+</td>
<td>++</td>
<td>+</td>
<td>Typically, Shadowing with calcification</td>
<td>Anterolateral face, neck or jaw</td>
</tr>
<tr>
<td>Meningocele</td>
<td>-</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>Related to neural tube defect</td>
<td>Spine and skull</td>
</tr>
<tr>
<td>Epignathus</td>
<td>+/-</td>
<td>++</td>
<td>+</td>
<td>+</td>
<td>Usually associated with polyhydramnios.</td>
<td>Anterior, emanating from fetal mouth</td>
</tr>
<tr>
<td>Epidermal scalp cyst</td>
<td>-</td>
<td>Unilocular</td>
<td>-</td>
<td>-</td>
<td>Extracranial cystic mass with no intracranial extension and no calvarial defect</td>
<td>Face surface and scalp</td>
</tr>
<tr>
<td>Ranula</td>
<td>-</td>
<td>unilocular</td>
<td>-</td>
<td>-</td>
<td>Purely cystic</td>
<td>Sub-lingual space</td>
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<tr>
<td>Vallecular cyst</td>
<td>-</td>
<td>unilocular</td>
<td>-</td>
<td>-</td>
<td>Moves with swallowing, polyhydramnios</td>
<td>Oral cavity, from the lingual surface of the epiglottis</td>
</tr>
<tr>
<td>Dacrycystocele</td>
<td>-</td>
<td>unilocular</td>
<td>-</td>
<td>-</td>
<td>Self-resolved +/- bilateral or unilateral</td>
<td>Lacrimal duct-related position</td>
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<tr>
<td>Cystic heterotopic brain tissue</td>
<td>-</td>
<td>unilocular</td>
<td>-</td>
<td>+/-</td>
<td>Fluid-accumulated cyst, irregular inner contour</td>
<td>Head and neck</td>
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<td>Choanal atresia</td>
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<td>-</td>
<td>-</td>
<td>Polyhydramnios</td>
<td>Posterior nasal region</td>
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