In-utero imaging, Decision making and surgical planning
Fetal Imaging

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Goals and Objectives

- Goal:
  - Understand the utility of fetal MRI
- Objectives:
  - Identify the main imaging features of fetal neck and chest pathologies affecting the airway
  - Demonstrate how fetal MRI aids in the diagnosis of different neck pathologies

Do we really need fetal MRI?

- US is still the first screening/diagnostic tool. MRI does not replace it
- Helps predict outcome, mortality and morbidity in fetuses with multiple pathologies, such as cervical teratoma or CDH
- Helps plan surgery and delivery: neck masses, shunt placements, EXIT procedures
- Helps identify additional abnormalities not seen on US

Fetal airway compromise

Lesions causing airway obstruction
1. Facial lesions
2. Neck lesions
3. Thoracic lesions

Facial lesions

- Oral lesions
- Cleft lip/palate
- Micrognathia
Epulis

- Benign. Arise from gingival mucosa of alveolar ridge
- Congenital gingival granular cell tumors (adults)

Pedunculated/ well defined
- Protrudes through the mouth
- Vascular pedicle
- Usually doesn’t affect swallowing/ airway
- Hypoechoic in US
- Hypointense T2 MRI- slightly hyper T1
- Ddx: epignathus, hemangioma, dermoid, teratoma
Giant epignathus
### Teratoma
- Congenital: 1- sacrococcygeal; 2- Head/Neck
- MC-immature; embryonal thyroid and neuroectoderm
- Anterolateral large solid /cystic mass; +/- calcifications
- Calcifications: specific for teratoma but in <20%
- In the vast majority of cases, the cervical airway is compressed and obstructed
- US: polyhydramnios, lesion vascularity, Doppler changes, hydrops
- MRI: lung volumes, intracranial and mediastinal invasion, and airway delineation

Rubio, et al, SPR poster 2011

### Amniotic AFP
- Amniotic AFP could help in the ddx of a cervical mass BUT only 30% produce AFP
- Maternal and fetal AFP levels can be elevated

### Facial anomalies
- Oral lesions
- Cleft lip/palate
- Micrognathia

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### Cleft lip/palate
- Lip closes 7-8 wks GA
- Palate closes by 12 wks
- Nonfusion: cleft lip/palate (CL/P)
- Predisposing factors for CL/P: Rubella, thalidomide, isotretinoin, retinoic acid, valproic acid, phenyoin, Alcohol, drug abuse, cigarette
- Preventing factor: folic acid
CL/P classification

Type 1: Cleft lip
Type 2: Unilateral cleft lip and palate
Type 3: Bilateral cleft lip and palate (premaxillary protrusion)
Type 4: Midline cleft lip and palate. Gaping midline with hypoplastic midface, absence primary palate
Type 5: Amniotic bands and slash-type defects

Bilateral CL/P permits forward migration of the premaxillary segment

Normal respiratory activity
Communication of the nasopharynx and oral cavity

Type 5 Slashed - Associated with amniotic bands
TABLE 2. ASSOCIATIONS WITH FACIAL CL/P

<table>
<thead>
<tr>
<th>Condition</th>
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</thead>
<tbody>
<tr>
<td>Anencephaly</td>
</tr>
<tr>
<td>Cleft lip/palate</td>
</tr>
<tr>
<td>Craniofacial dysmorphism</td>
</tr>
<tr>
<td>Ear defects</td>
</tr>
<tr>
<td>Facial dysmorphism</td>
</tr>
<tr>
<td>Hypogonadism</td>
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<tr>
<td>Mental retardiation</td>
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<tr>
<td>Microophthalmia</td>
</tr>
<tr>
<td>Nystagmus</td>
</tr>
<tr>
<td>Obstructive sleep apnea</td>
</tr>
<tr>
<td>Partial anomalous facial cleft</td>
</tr>
<tr>
<td>Renal anomalies</td>
</tr>
<tr>
<td>Septo-optic dysplasia</td>
</tr>
<tr>
<td>Skeletal anomalies</td>
</tr>
<tr>
<td>Turner syndrome</td>
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</tbody>
</table>

CL/P is associated with multiple syndromes.

Bilateral
Facial anomalies

- Oral lesions
- Cleft lip/palate
- Micrognathia

No nasopharynx/oropharynx

Narrow maxilla

Piriform sinuses

Oropharynx level

Broad nose

Left ear

Right ear
Differential diagnosis

- Treacher- Collins Syndrome
- Goldenhar syndrome
- Roberts syndrome
- Pierre Robin syndrome
- Nager syndrome
- Chromosomal abnormalities: 13/18
- Crouzon’s syndrome
- Cornelia de Lange
- Smith Lemli-Opitz syndrome

Micrognathia index by US

- Linear relationship between mandibular growth and gestational age or BPD
- Cutoff level of less than 23, the jaw index had a 100% sensitivity and 98.1% specificity in diagnosing micrognathia
Mandibular index = AP mandibular diameter / BPD x 100

Profile

Mandibular index 28.3%

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Neck pathology
1. Congenital lesions of the neck: BCC, CTC, ranula, TDC, Dermoid/epidermoid,
2. Inflammatory lesions: goiter
3. Vascular malformations:
   - Low flow: LM/ mixed lesions
   - High flow: AVF, AVM
4. Tumors: hemangioma, teratoma, myofibromatosis, neuroblastoma, rhabdosarcoma, etc.

Table 4. The jaw index statistics and percentiles of the normal population of 262 fetuses.

<table>
<thead>
<tr>
<th>Statistic</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean</td>
<td>15.5 (9.8)</td>
</tr>
<tr>
<td>Standard deviation</td>
<td>3.9</td>
</tr>
<tr>
<td>Median</td>
<td>14.2</td>
</tr>
<tr>
<td>Range</td>
<td>25-51</td>
</tr>
</tbody>
</table>

Figure 4: Boxplot of the jaw index in normal fetuses and fetuses with micrognathia. Boxes indicate quartiles (25th and 75th percentiles), and vertical bars indicate ranges.

Cervical thymic cyst
- Persistence of the thymopharyngeal tract
- Lateral superior neck. Midline inferior
- Left + common
- Unilocular vs multilocular
- Close to the carotid space: splay CA and JV
- Anechoic/ debris. Hyperintense T2
- Ddx: LM, TDC, BCC
- Good prognosis

Branchial cleft cyst
- Defect in embryogenesis of the branchial apparatus
- Several types: Type II most common*
- Lateral neck although can extend to midline
- Anechoic/ hypoechoic. Debris. Thin walled
- Hypo T1- Hyper T2
- Isolated or associated to syndrome: brachio-oto-renal syndrome
- Ddx: LM, TDC
- Can cause airway compression. EXIT

Hyperthyroid goiter:
- Tachycardia
- Advanced bone age
- Hydrops
- IUGR
- Hepatosplenomegaly

Thyroid Goiter
- Causes: Mom with thyroid problems (Hashimoto’s, Graves, iodine ingestion, antithyroid medications)
- Best views-coronal; sagittal
- US-homogeneous, bilobed, echogenic, hypervascular; possibly poly, high output cardiac failure
- MRI-hypointense on T2 (nl-iso). Bright T1
- Complications: hyperextension and head-airway obstruction
- Check fetus thyroid status by cord blood (0.5-1.4 % risk fetal loss)
- TTx: correct Mom’s issues; Intra-amniotic/cervical T4* 
- Good prognosis

References:
Cervical teratoma

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Teratoma

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Sometimes the mass is resected while in utero – placental bypass

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Vascular malformations/tumors

- **Vascular malformations**
  - Low flow: Lymphatic/Venolymphatic
  - High flow: AVM-AVF

- **Vascular tumors:**
  - Hemangioma


Lymphatic malformations

- **Nuchal:** macrocystic lesions (cystic hygroma) 1st trimester
- **Non-nuchal:** microcystic lesions (lymphangioma)

  - Failure in communication of the jugular lymphatic sac and jugular veins at 40 days GA
  - Nuchal LM: 51% associated with chromosomal anomalies (T 13-18-21, Turner’s), cardiac and skeletal malformations
  - Non-nuchal LM: isolated or Gorham-Stout, Klippel-Trenaunay


Nose

Chin

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Congenital High Airway Obstruction Syndrome (CHAOS)
- Absence of all or part of trachea
  - Trachea atretic, stenotic or thick web
  - Aberrant pulmonary budding off the foregut
  - Most have connection with esophagus
  - Associated anomalies and syndromes, Fraser or DiGeorge
- Prognosis poor
- Rare

CHAOS
- Prenatal
  - Intrauterine demise
  - Hydrops
- Postnatal
  - EXIT to ECMO
  - Tracheal intubation

CHAOS
- US
  - Symmetric enlarged echogenic lungs
  - Heart compressed and diaphragm flattened
  - Dilated bronchi
  - Hydrops
  - Decreased or increased amniotic fluid
- MRI
  - Lungs hyperintense and large with inverted diaphragms
  - Heart compressed
  - Dilated high signal fluid filled trachea and bronchi

25 weeks
Differential: Bilateral CCAM

Don’t Forget to Look At The.....

CDH

Mediastinum
Esophageal atresia

Thank you!