Congenital Laryngeal Anomalies

Prof. Hesham Abd Al-Fattah
Alexandria - Egypt

Embryology

• Resp primordium 3rd wk
• Resp primordium separated by tracheoesophageal folds
• Fuse to form septum (4-5 wks)
• Larynx from 4th & 6th arches
• Primitive larynx altered by hypobranchial eminence, epiglottis, arytenoids
• Laryngeal lumen obliterated and recanalized
ANATOMICAL DIFFERENCES

- Smaller airway
- Shorter airway
- Loose submucosa
- Rich lymphatics
- Shape of the epiglottis
- Soft cartilages
- Higher in position

In Adults
- >2/3
- >1/2
- (1 mm subglottic narrowing = 32% Lumen reduction)
- Narrowing >>> severe airway distress

In Children
- New Born subglottic = 4.5 mm
- Non-pass of 3 mm bronchoscope = Subglottic Stenosis

Diagnosis

- History
- Clinical Picture
- Investigation:
  - Radiography
  - Neck films, chest films
  - Barium swallow
  - Multi-slice CT with virtual endoscopy / rarely MRI (when there is swelling to determine its nature)
  - Flexible Endoscopy, if the stridor is not severe
  - Endoscopy in OR is the Gold standard
**Classification**

**Site:**
- Supraglottic – Glottic – Subglottic.

**Structure:**
- Cartilage – nerve – joint – BV.

**Incidence of occurrence:**
- Laryngomalacia.
- Vocal Fold paralysis.
- Subglottic stenosis.
- Subglottic Haemangioma.
- Other.

---

**Laryngomalacia**

- Most common (60%)
- M:F = 2 : 1

**Etiology:**
- Immaturity of cartilage
- Immature neuromuscular c.
- GERD.

- No difference between the quality of the cartilage structures in infants with laryngomalacia and those who have normal development.
- An increase in the incidence of Laryngomalacia does not occur in premature infants who have classic hypotonicity.
- Inflammation of variable intensity beneath the epithelium with edema deep to it.
**Laryngomalacia**

**Diagnosis**
- **Clinical**
  - Inspiratory stridor
  - Starting usually few weeks after birth
  - Most cases are mild & it is the noise which annoys the parents
- **Radiological**
- **Endoscopy:**
  - Flexible endoscopy.
  - Rigid endoscopy
    - GA or NO GA !!! muscle relaxant.

**Endoscopic findings**

I) Epiglottis: Omega shaped / Floppy.

II) AE fold: Short / Inwarding.

III) Arytenoid: Redundant MM/ bulky.

IV) Discoordinate pharyngolaryngomalacia:
  - Complete collapse of supraglottic structures.
  - No anatomical abnormalities.

V) Posteriorly displaced Lat Glossoepi Fold
Laryngomalacia

Grades

I

II

III

Laryngomalacia

- Assurance /observation
- Treat GERD.
- Surgical treatment: (Indications)
  - Severe stridor with failure to thrive.
  - Weight loss, chest deformity.
  - Cyanotic attacks, Cor pulmonale.
  - Hypoxemia and Hypercapnia.
Laryngomalacia

Surgical treatment:
- Tracheostomy.
- Supraglottoplasty:
  - Division of the AE fold.
  - Removal of the redundant supra-arytenoid mucosa and lateral borders of epiglottis.
- Suprahyoid epiglottectomy.

Laryngomalacia

Surgical treatment:
- Tracheostomy.
- Supraglottoplasty:
  - Division of the AE fold.
  - Removal of the redundant supra-arytenoid mucosa and lateral borders of epiglottis.
- Suprahyoid epiglottectomy.
Laryngomalacia

**Surgical treatment:**
- Tracheostomy.
- Supraglottoplasty:
  - Division of the AE fold.
  - Removal of the redundant supra-arytenoid mucosa and lateral borders of epiglottis.
- **Suprahyoid epiglottectomy.**
**Cong. VC Paralysis**

- The second most common anomaly (15-20%)
- M = F

**Bilateral:**
- Neuromuscular immaturity.
- CNS anomalies.
- Birth trauma.
- Idiopathic.

**Unilateral:**
- Birth trauma.
- Mediastinal lesions.
- Iatrogenic.

---

**Clinical presentation:**

**Bilateral:**
- Insp. Stridor:
  - after birth immediately
  - Severe → airway support.
- Voice → near-normal.
- Aspiration.

**Unilateral:**
- Few weeks after birth or pass unnoticed.
- Hoarse breathy cry.
- Aspiration (rare).
Cong. VC Paralysis

**Diagnosis:**
- **Endoscopy:**
  - Flexible (mild airway distress)
  - Rigid (severe airway distress)
  - Radiological (CNS, chest)
**Diagnosis:**
- Endoscopy:
  - Flexible (mild airway distress)
  - Rigid (severe airway distress)
- Radiological (CNS, chest)

**Management:**
1) Unilateral:
- Observation.
- Upright position.
- Tracheostomy (severe aspiration)
Cong. VC Paralysis

Management:

II) Bilateral:

- Treatment of cause e.g. shunt op.
- Tracheostomy and F-U (1-2 years):
  - Spont. Recovery (>50%) \(\rightarrow\) Decannulation.
  - No recovery \(\rightarrow\) Lateralization procedure.

Cong. VC Paralysis

VC Lateralization:

- **Arytenoidectomy**
  - Open.
  - Endoscopic.
- Laser cordotomy.
Cong. VC Paralysis

VC Lateralization:
- Arytenoidectomy
  - Open.
  - Endoscopic.
- Laser cordotomy.

Cong. SG Stenosis
- SG lumen < 4 mm in full term & < 3 mm in premature. (N = 4.5 mm)
- 3rd common anomaly.
- Cartilaginous X Soft tissue.
- Four grades: (Cotton grading)
  - I = < 50% Obst.
  - II = 51 - 70% Obst.
  - III = 71 - 99% Obst.
  - IV = No lumen.
Cong. SG Stenosis

Clinical presentation:

- **Mild:**
  - Persistent / Rec. croup.
  - Difficulty intubation during general anesthesia.
  - Difficult Decannulation after tracheostomy

- **Severe:**
  - Upper airway obstruction after birth.

Diagnosis:

**Endoscopy:**

"SG stenosis is an endoscopic diagnosis" (Cotton RT. Otol. Clinic N Am. 2000)

- Passing ET or bronchoscope of known diameter.
- Exclude ass. anomalies.
Cong. SG Stenosis

Diagnosis:
- Plain X-ray
- CT scan
- 3D CT scan
- Virtue endoscopy

Management:
- Follow-up (Child outgrow the problem)
- Tracheostomy & FU.
- Endoscopic (laser) dilation:
  - limited role.
  - Only for soft stenosis.
- Surgical treatment.
Cong. SG Stenosis

Surgical treatment:
- Anterior Cricoid Split:

![Image of surgical procedure]

Cong. SG Stenosis

Surgical treatment:
- Laryngotracheoplasty:

![Image of surgical procedure]
Cong. SG Stenosis

Surgical treatment:
- Laryngotracheoplasty:

![Image of surgical procedure]
SG Haemangioma

- Congenital vascular malformation of mesodermal rests.
- 1.5% of congenital laryngeal anomalies.
- M : F = 1 : 2
- 30% at birth, usually starts to grow 6-18 mon → UAO
- Stridor (Mixed long insp & short exp) feeding problems later
- Usually involutes by the age 4 - 5 ys.
- Associated with other haemangiomas of H&N (50%)

Stridor (Mixed long insp & short exp) feeding problems later

Diagnosis:

- Radiological.
- Rigid endoscopy
SG Haemangioma

Management:
1. Tracheotomy and follow-up for spontaneous regression (2-4 ys.).
2. Corticosteroid (systemic or intralesional)
3. Laser endoscopic surgery (KTP/CO2).
4. Interferon.
5. Cryosurgery.
7. Surgical excision.

Repeated Laser debulking
(Laser → Fibrosis → SG stenosis)

Tracheostomy & FU 4-5 ys.

Non involuting
Involution

Surgical excision
Decannulation
**S G Haemangioma**

- **Present Policy**
  - Tracheostomy
  - Laser Debulking Once Only
  - Mitomycin-C
  - Decannulation
  - 4 Cases > 2 ys
Laryngeal Web

- Failure of re-canalization.
- Site:
  - Glottic.
  - Supraglottic
  - Subglottic.
- Size:
  - Thin.
  - Thick (associated SG stenosis)

Open Excision of Subglottic Haemangiomas to Avoid Tracheostomy
Laryngeal Web

**Diagnosis:**
- Endoscopy.
- Radiological.

**Management:**
- Excision:
  - Thin → MLS - Laser.
  - Thick → Laryngofissure
- Prevent recurrence:
  - Keel.
  - Mitomycin.

---

Laryngeal Web

1. Endoscopic keel insertion:

---
Laryngeal Web

1. Endoscopic keel insertion:
Laryngeal Web

2. Mitomycin application:
   - Local application for 5 minutes.
   - Inhalation by nebulizer:
     1/2 cc of diluted MMC every 8 hrs. for 3 weeks

---

Laryngeal papillomatosis

- Human Papilloma Virus (6 & 11)
- Bimodal age distribution (<2 & 20 yrs.)

**Risk factors:**
- Teenage mother.
- First born child.
- Vaginal delivery.

**Clinical presentation:**
- Change of voice (early)
- Airway obstruction.
Laryngeal papillomatosis

**Diagnosis:**
- Endoscopic.

**Management:**
- Tracheostomy.
- Laser debulking.
- Antiviral drugs.
- Mitomycin.

Histologic characterization of human papilloma virus in respiratory papillomas after Mitomycin-C application

Hesham Abd Al-Fattah, Ashraf Hamza, Manal Nasr, University of Alexandria, Egypt.

Histopathology and PCR data failed to identify any HPV pathology or DNA in all remission specimens.
Laryngeal Cyst

Types:
- Saccular cyst
- Ductal cyst
- Duplication cyst
- Thyroglossal cyst
- Cyst in ectopic thyroid gland

Management:
- Endoscopic excision
- Surgical excision

Cystic Hygroma

- Dilated lymphatic spaces
- Invade the larynx → UAO 1st year

Diagnosis:
- Endoscopy
- Radiological

Management:
- Tracheostomy
- Laser ablation
- Surgical excision
Cri Du Chat Syndrome

Chromosomal abnormality, 1/50,000 births

Criteria:
- Growth retardation, microcephaly.
- CVS defects, skeletal abnormalities.
- Mowing cry & high pitched stridor (paralysis of interarytenoid muscle).
- Triangular epiglottis.

Management of a Child with Congenital Laryngeal Anomaly
1. History:
   I) Respiratory distress + cyanotic attacks:
      - Relieved by crying = Supralaryngeal obstruction (e.g. choanal atresia)
      - Onset:
        1. Immediately after birth:
           - SG stenosis, Cong. VC paralysis
        2. Delayed:
           - Laryngomalacia (1st week)
           - Laryngeal hemangioma (1st 6 months)

   II) Change of voice:
      - Muffled voice = Supraglottic cyst
      - Weak or absent voice (cry):
        - Laryngeal web.
        - Severe subglottic stenosis

   III) Aspiration:
      - Laryngeal cleft.
      - VC paralysis.
2. Examination:
VC Dysfunction

**Def.:**
Paradoxical mov. of VC → abnormal adduction of the anterior part during inspiration.

**Etiology:**
lesion affects the Vagus nerve → alter laryngeal tone → dec. laryngeal threshold for stimuli. e.g. *Bulbar Myesthenia Gravis.*

**Clinically:**
- Change of voice.
- Stridor.
- Cough
- Attacks of laryngeal spasm.
- Wheezy chest (miss diagnosis as B.A)
VC Dysfunction

Diagnosis:
Endoscopic ex. of the larynx

Post. Glottic chink while ant. Parts of VC are adducted.

2. Examination:
Use of stents in Paediatric Age Gp

Cases of congenital laryngeal anomalies presented to ORL & Pediatric Departments – Alexandria University in the period from January 2002 – January 2005. (endoscopically + Radiologically)

- 56 cases:
  - 32 boys : 57%
  - 24 girls : 43%
Laryngomalacia

**Surgical treatment:**
- Tracheostomy.
- **Supraglottoplasty:**
  - Division of the AE fold.
  - Removal of the redundant supra-arytenoid mucosa and lateral borders of epiglottis.
- Suprahypoid epiglottectomy.

**Results**
- Laryngomalacia: 17 + 1 (*VC paralysis*) 32%
- Cong. VC paralysis: 7 13%
- Cong. SG stenosis: 8 14%
- Laryngeal web: 7 + 1 (*SG stenosis*) 14%
- SG hemangioma: 4 7%
- Papilloma: 6 10.5%
- Laryngeal cyst: 2 3.5%
- VC dysfunction: 1 2%
- Cri du chat: 1 2%
- Cystic hygroma: 1 2%
Conclusion

- Laryngomalacia, VC paralysis and SG stenosis are the most common congenital laryngeal anomalies.
- Endoscopy is the gold standard investigation in cases of congenital laryngeal anomalies.
- Management depends on the severity and the degree of the disease.

Thank You
Congenital Laryngeal Anomalies: Incidence & Management.

Prof. Dr. H. Abdel-Fattah, Prof. Dr. N. Faseyh, Dr. A. Gaafar, Dr. Y. Nour
Alexandria - Egypt