Congenital Laryngeal Anomalies

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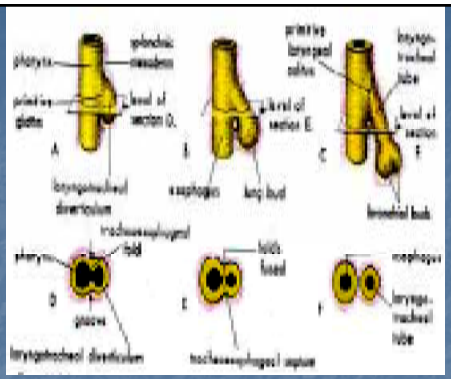
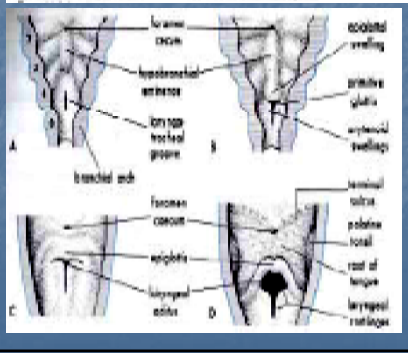
Alexandria - Egypt

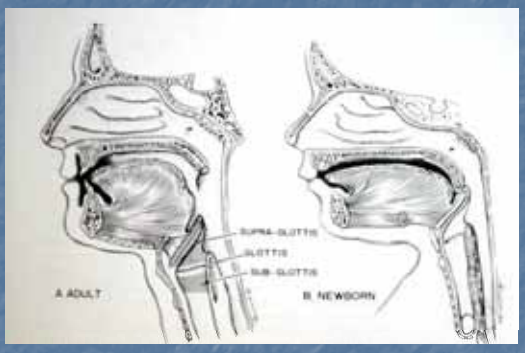
Embryology

- 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



1. Smaller air-way
2. Shorter air-way
3. Loose submucosa
4. Rich lymphatics
5. Shape of the epiglottis
6. Soft Cartilages
7. Higher in position

•Vestibule
•subglottis

In Adults >2/3
In Children >1/2
(1 mm subglottic narrowing =32% Lumen reduction)

Narrowing>>> sever air way distress

New Born subglottic = 4.5mm
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 sever
  - Endoscopy in OR is the Gold standard

Classification

Site:

- Supraglottic – Glottic – Subglottic.

Structure:

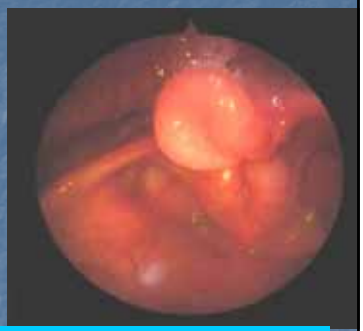
- Cartilage – nerve – joint – BV.

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**
- **Radiological**
- **Endoscopy:**
  - **Flexible endoscopy.**
  - **Rigid endoscopy**
    - **GA or NO GA !!!! muscle relaxant.**

- •Inspiratory stridor
- •Starting usually few weeks after birth
- • Most cases are mild & it is the noise which annoys the parents
- •Self limiting, disappear between 18-24 months of age
- •If the stridor is sever, other anomalies needs to excluded

Laryngomalacia

Endoscopic findings

I) Epiglottis : Omega shaped /Floppy .

II) AE fold : Short / Inwarding.

III) Arytenoid : Redundant MM/ bulky.

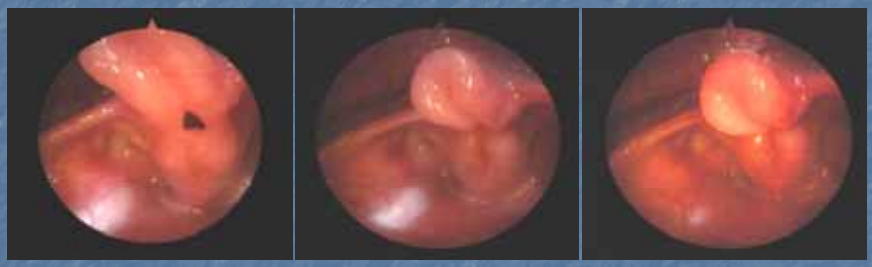
IV) Discoordinate pharyngolaryngomalacia:

- 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

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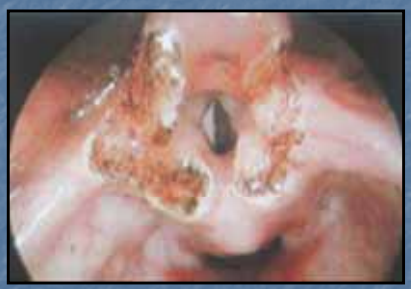

Laryngomalacia

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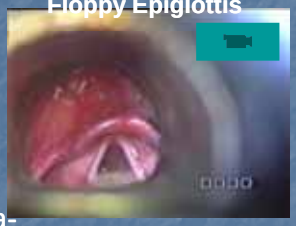
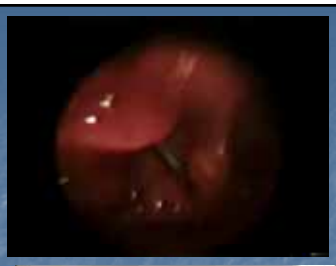
Floppy Epiglottis



1
Long Tubular epi
Redudent arytenoid



2..\video & pictures\cong anomalies larynx\MOV01269.AVI
Supraglottoplasty

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.

Cong. VC Paralysis

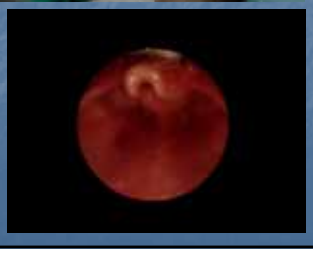
Clinical presentation:

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)

Cong. VC Paralysis

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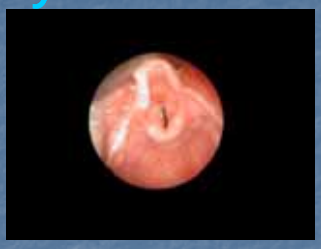
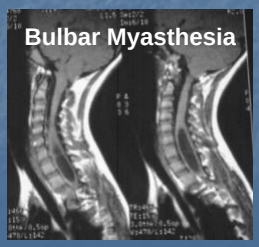




Cong. VC Paralysis

Management:

I) Unilateral:

- Observation.
- Upright position.
- Tracheostomy (severe aspiration)

Cong. VC Paralysis

Management:

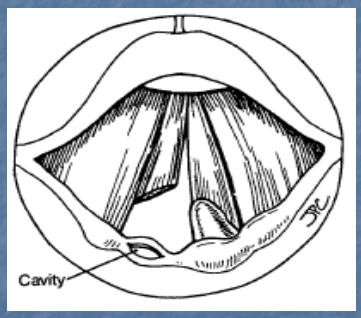
II) Bilateral:

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

Cong. VC Paralysis

VC Lateralization:

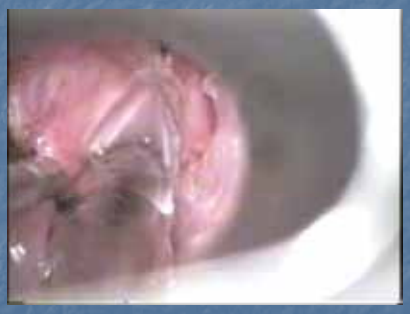
- **Arytenoidectomy**
  - Open.
  - Endoscopic.
- Laser cordotomy.



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:

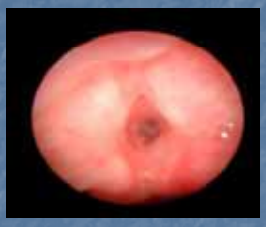
- Mild:
  - Persistent / Rec. croup.
  - Difficulty intubation during general anesthesia.
  - Difficult Decannulation after tracheostomy
- Severe:
  - Upper airway obstruction after birth.

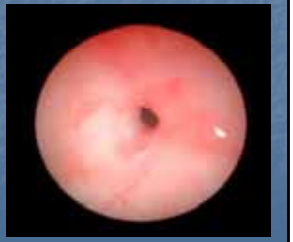
Cong. SG Stenosis

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.

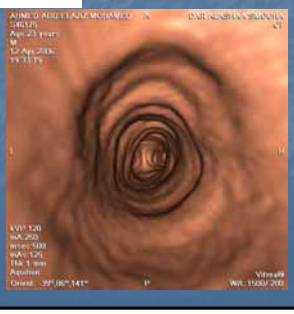




Cong. SG Stenosis

Diagnosis:

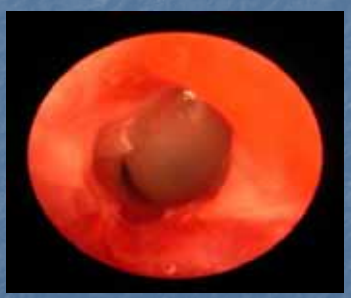
- Plain X-ray
- CT scan
- 3D CT scan
- Virtue endoscopy



Cong. SG Stenosis

Management:

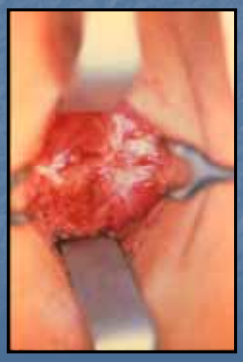
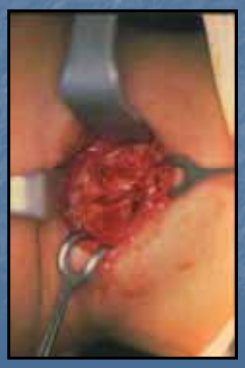
- Follow-up (Child outgrow the problem)
- Tracheostomy & FU.
- Endoscopic (laser) dilation:
  - limited role.
  - Only for soft stenosis.
- Surgical treatment.



Cong. SG Stenosis

Surgical treatment:

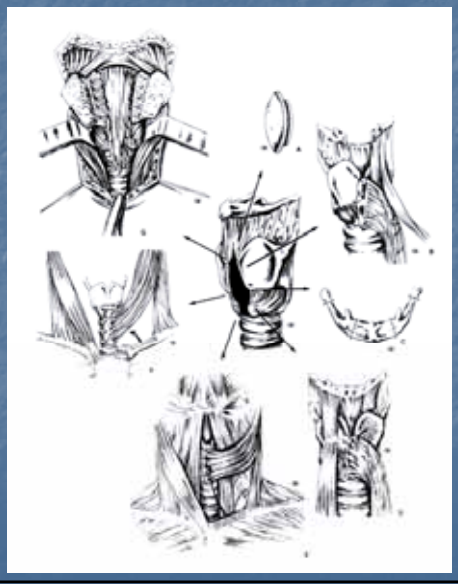
- Anterior Cricoid Split:


Cong. SG Stenosis

Surgical treatment:

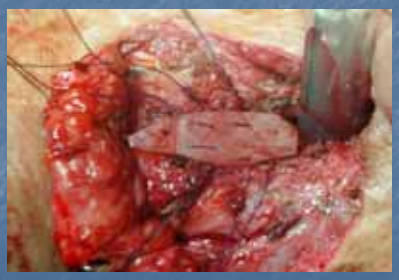
- Laryngotracheoplasty:



Cong. SG Stenosis

Surgical treatment:

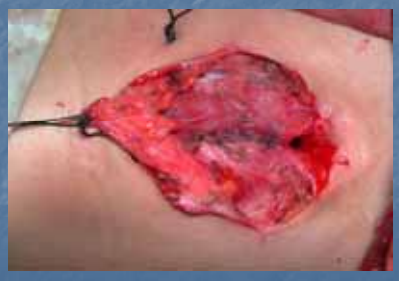
- Laryngotracheoplasty:



Cong. SG Stenosis

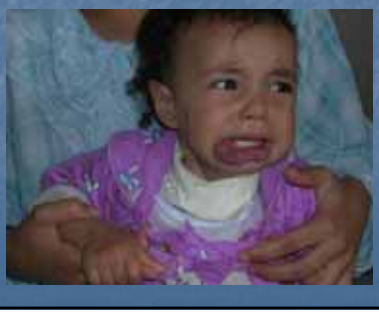
Surgical treatment:

- Laryngotracheoplasty:



SG Haemangioma

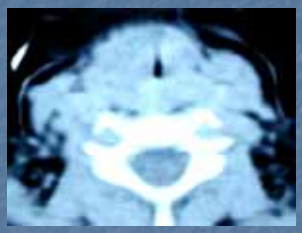
- Congenital vascular malformation of mesodermal resets.
- 1.5% of cong. laryngeal anomalies.
- M : F = 1 : 2
- 30% at birth, usually starts to grow 6-18 mon → UAO
- Stridor (Mixed long insp & short exp) feeding problems later
- Usually involutes by the age 4 – 5 ys.
- Associated With other haemangiomata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

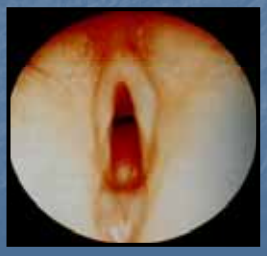


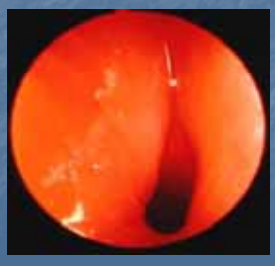
SG Haemangioma

Diagnosis:

- Radiological.
- Rigid endoscopy







SG Haemangioma

Management:

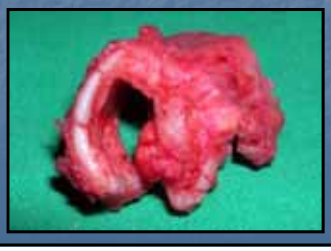
1. Tracheotomy and follow-up for spontaneous regression (2-4 ys.).
2. Corticosteroid (systemic or intralesional)
3. Laser endoscopic surgery (KTP/CO2).
4. Interferon.
5. Cryosurgery.
6. External irradiation.!!!!!
7. Surgical excision.

SG Haemangioma

Management

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

↓

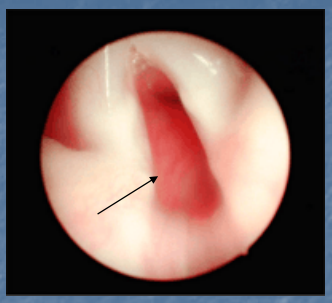
Mitomycin-C

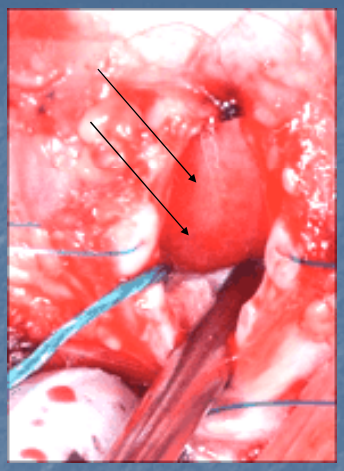
↓

Decannulation

↓

4 Cases > 2 ys

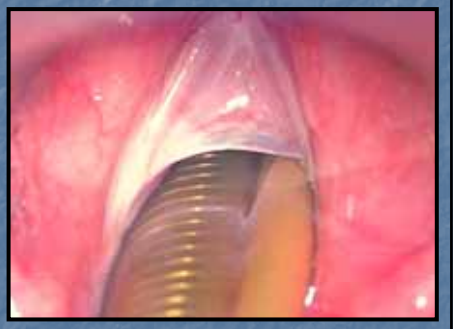




Vijayasekaran S,. White D R, Hartley B EJ, Rutter M J, Elluru RG, Cotton RT
Open Excision of Subglottic Haemangiomas to Avoid Tracheostomy
Arch Otolaryngol Head Neck Surg. 2006;132:159-163.

Laryngeal Web

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



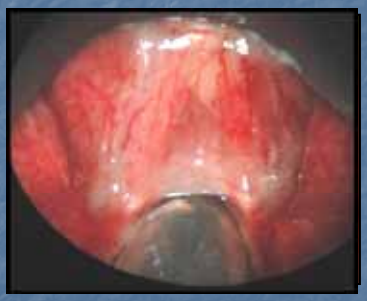
Laryngeal Web

Diagnosis:

- Endoscopy.
- Radiological.

Management:

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



Laryngeal Web

1. Endoscopic keel insertion:

Laryngeal Web

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Laryngeal Web

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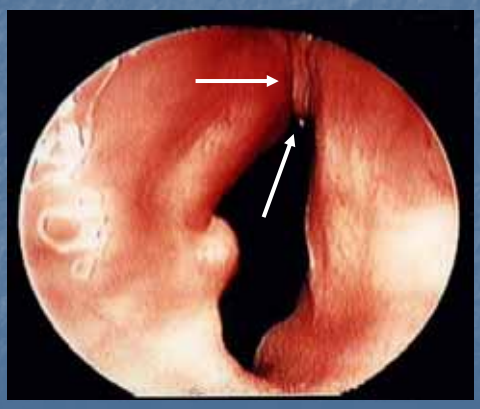


Laryngeal Web

2. Mitomycin application:
   - Local application for 5 minutes.
   - Inhalation by nebulizer:

     1/2 cc of diluted MMC every 8 hrs. for 3 weeks

Inhalation mitomycin-C in the management of laryngeal fibrosis: rationale, benefits, and pitfalls.
Hesham A. Fattah*, Ashraf Hamza*, Alaa Gaafar*, Mervat Hamza**, Zinab Mourad***
International Congress Series 2426 (2003)

Laryngeal papillomatosis

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

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

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

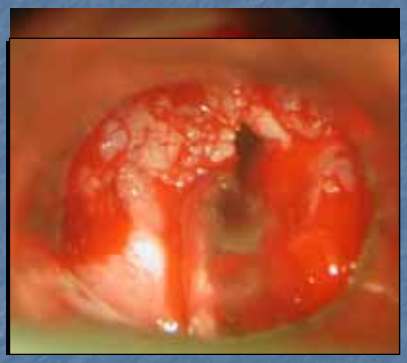
Laryngeal papillomatosis

Diagnosis:

- Endoscopic.

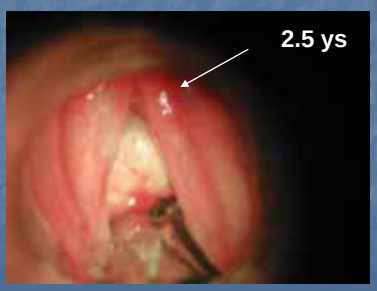
Management:

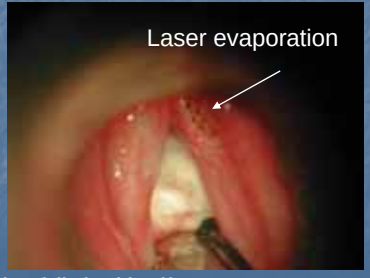
- Tracheostomy.
- Laser debulking.
- Antiviral drugs.
- Mitomycin.



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.

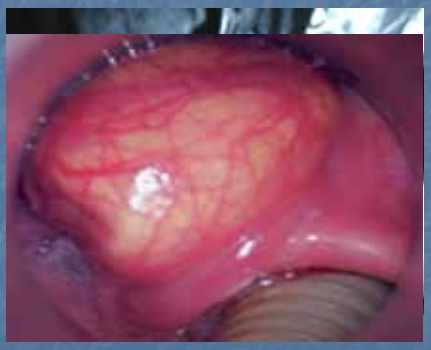
Laryngeal Cyst

Types:

- Saccular cyst
- Ductal cyst.
- Duplication cyst.
- Thyroglossal cyst.
- Cyst in ectopic thyroid gland.

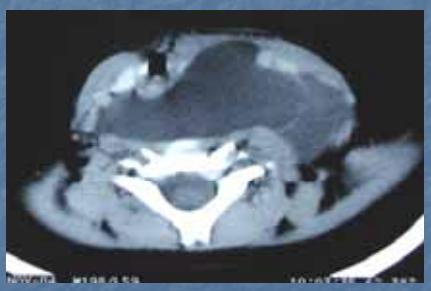
Management:

- Endoscopic excision.
- Surgical excision.



Cystic Hygroma

- Dilated lymphatic spaces.
- Invade the larynx → UAO 1st year.
- Diagnosis:
  - Endoscopy.
  - Radiological.
- Management:
  - Tracheostomy.
  - Laser ablation.
  - Surgical excision.

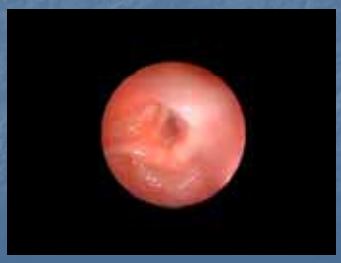


Cri Du Chat Syndrome

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)

1. History:

II) Change of voice:

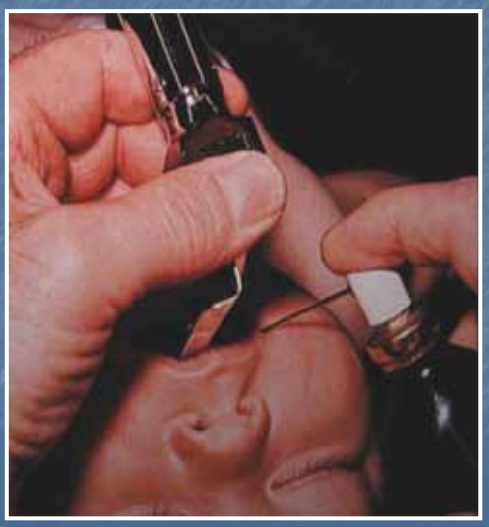
- Muffled voice = Supraglottic cyst
- Weak or absent voice (cry):
  - Laryngeal web.
  - Severe subglottic stenosis

III) Aspiration:

- Laryngeal cleft.
- VC paralysis.

2. Examination:

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:

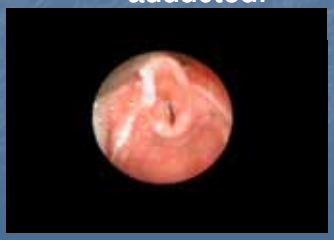
- Change of voice.
- Stridor.
- Cough
- Attacks of laryngeal spasm.
- Wheezy chest (miss diagnosis as B.A)

VC Dysfunction

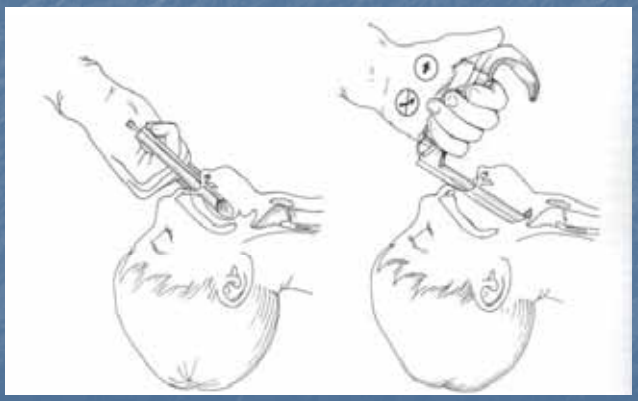
Diagnosis:

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.
- Suprahyoid epiglottectomy.

Results

Laryngomalacia:	17 + 1 *(VC paralysis)*	32%
Cong. VC parlaysis	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.

Congenital Laryngeal Anomalies: Incidence & Management.

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