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- CONGENITAL DISEASES OF THE LARYNX –

1- Laryngomalacia (congenital laryngeal stridor)
2- Congenital vocal fold paralysis
3- Congenital subglottic stenosis
4- Congenital web
5- Laryngeal atresia
6- Congenital subglottic haemangioma
7- Congenital laryngocoele

(1) Laryngomalacia -

- **Definition:** Congenital laryngeal stridor due to flaccidity of supraglottic structures. It is a misnomer because there is no softening of the laryngeal cartilages.
- **Incidence:**
  - The commonest congenital anomaly of the larynx
  - The commonest cause of congenital stridor
- **Etiology:**
  - **Structural defect:** Flaccidity of supraglottic structures → the supraglottis collapses during inspiration.
  - **Anatomic abnormality:** (fig 17)
    - The epiglottis is tall, tubular, and folded upon itself (omega-shaped epiglottis)
    - The aryepiglottic folds are short
    - The arytenoids are bulky

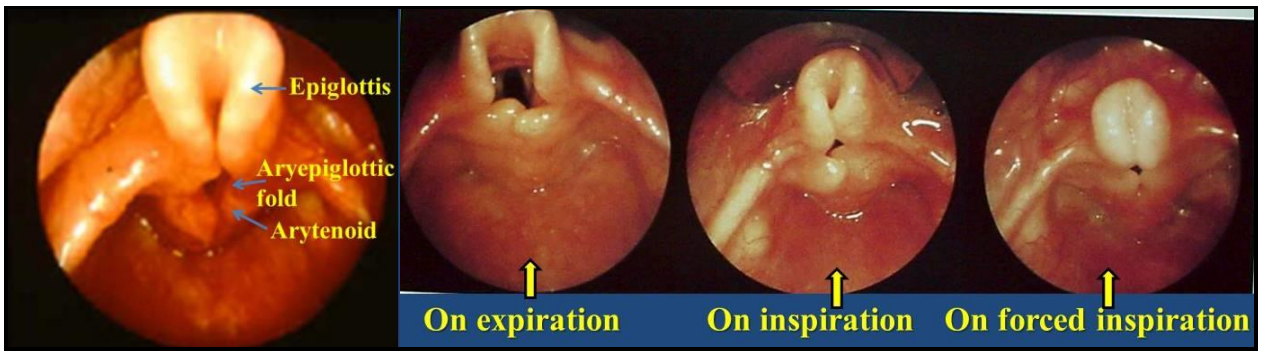

Fig (17) Omega-shaped epiglottis and supraglottis in respiration

- **Clinical picture:**
  - **Symptoms:**
    - Inspiratory stridor starts soon after birth, increases with respiratory effort (e.g. crying, feeding) and in the supine position and improves in the prone position.
    - Normal cry with no hoarseness
  - **Signs:** on endoscopy: (fig 17)

- Tall tubular epiglottis, short aryepiglottic folds, bulky arytenoids
- The supraglottis collapses during inspiration and becomes normal during expiration.
- No signs of respiratory distress.

- **Investigations:** Fiberoptic or rigid laryngoscopy
- **Treatment:**
  - **Reassurance:** as it improves and disappears spontaneously by the age of 18 months-2years in most cases
  - **In severe cases (rare):** with severe airway obstruction and feeding problems → surgical treatment:
    - Tracheostomy to relieve stridor
    - Supraglottoplasty or epiglottopexy

(2) Congenital vocal fold paralysis -

- **Incidence:**
  - 2nd most common cause of congenital stridor
  - Bilateral paralysis is commoner than unilateral paralysis
  - In unilateral paralysis, the left side is commoner than the right side
- **Etiology:**
  - **Bilateral paralysis:**
    - CNS lesions: - meningocoele, MEC - hydrocephalus
      - Arnold Chiari malformation
    - Congenital syphilis
    - Congenital myasthenia gravis
  - **Unilateral paralysis:**
    - Congenital CV anomalies: - VSD - PDA - Fallot's tetralogy
    - Unknown
- **Clinical picture:**
  - **Symptoms:**
    - Unilateral paralysis: → weak, breathy cry or aspiration
    - Bilateral paralysis: → stridor and normal cry or aspiration with recurrent chest infections.
  - **Signs:**
    - In bilateral paralysis: There is a significant respiratory distress (nasal flaring, supraclavicular or intercostal indrawing, cyanosis).
    - Laryngoscopy shows the position of the paralyzed vocal fold
- **Treatment:**

- **Severe stridor:** → tracheostomy
- **Treatment of underlying cause:** to relieve the nerve compression

(3) Congenital subglottic stenosis -

- **Definition:** subglottic lumen < 3.5 mm in full term newborn (fig 18)
- **Incidence:** 3rd common cause of congenital stridor
- **Classification:**
  - - Cartilaginous - Membranous or - Combined stenosis

Fig (18) Subglottic stenosis

- **Clinical picture:**
  - **Symptoms:**
    - Severe cases: Biphasic stridor, normal cry
    - Mild cases:
      - Atypical croup: stridor lasting for a longer time than usual following laryngitis
      - Difficult intubation, extubation, and decannulation
  - **Signs:** on direct laryngoscopy → Narrowing of the subglottic lumen < 3.5 mm (fig 18)
- **Treatment:**
  - **Mild cases:** → no treatment
  - **Severe cases:**
    - Tracheostomy with expiratory valve and wait up to the age of 2-5yr to allow growth of the larynx
    - If failed decannulation: surgical treatment of the stenosis

(4) Congenital web -

- **Definition:** Fibrous band extending between the anterior parts of vocal folds
- **Etiology:** Failure of resorption of the epithelial lamina
- **Classification:**
  - **Site:**
    - Supraglottic web and subglottic web → rare

- Glottic web → commonest, it may be:
  - Anterior glottic web: commonest or
  - Posterior glottic (interarytenoid) web: rare

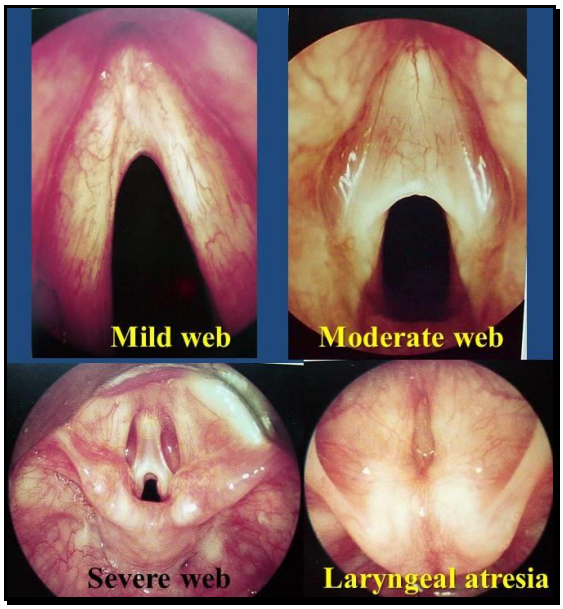

Fig (19) Glottic web severity

- **Thickness:** thin web and thick web
- **Severity:** mild, moderate and severe web (fig 19)

- **Clinical picture:**
  - **Symptoms:** according to the severity of the web:
    - Asymptomatic
    - Abnormal cry (weak, hoarsy)
    - Inspiratory stridor with severe web
  - **Signs:** on direct larygoscopy:
    - A white triangular band in the anterior part of the glottis with thin, regular, concave posterior edge

- **Treatment:**
  - **Asymptomatic web:** → no treatment
  - **Symptomatic web:**
    - Thin web: Microlaryngosurgical excision with insertion of a keel to prevent recurrence.
    - Thick web: tracheostomy to relieve airway obstruction and excision of the web through laryngofissure with insertion of a keel.

(5) Laryngeal atresia -

- **Etiology:** Complete failure of resorption of the epithelial lamina

- **Clinical picture:** At delivery the newborn makes a strong respiratory effort but with no air movement, no cry or stridor with deep cyanosis and death
- **Treatment:** Urgent tracheostomy

(6) Congenital subglottic hemangioma -

- **Clinical picture:**
  - Subglottic swelling, smooth, usually unilateral
  - Associated with cutaneous haemangiomas in 50% of cases
  - Can cause severe biphasic stridor
  - Spontaneous regression with age may occur
- **Treatment:**
  - Tracheostomy: if there is severe airway obstruction
  - Conservative treatment till the age of 5years as spontaneous regression may occur

(7) Congenital laryngocoele -

- **Definition:** air filled dilatation of the saccule
- **Etiology:** due to abnormally large saccule
- **Clinical picture:**
  - **Symptoms:**
    - Asymptomatic or
    - Intermittent hoarseness or stridor that increases with straining
  - **Signs:** may be:
    - External laryngocoele: passes through a hole in the thyrohyoid membrane into the neck → smooth swelling over the thyrohyoid membrane, fluctuant, soft, increases on straining and disappear on pressure
    - Internal laryngocoele: A smooth swelling distending the ventricular band and aryepiglottic fold
    - Combined external and internal laryngocoele
- **Investigations:**
  - Plain antero-posterior radiographs of the neck without Valsalva and with Valsalva's
  - Direct laryngoscopy: diagnostic
- **Treatment:**
  - Asymptomatic: no treatment
  - Symptomatic: surgical excision

Pearls in bullets:

- Laryngomalacia is the commonest congenital anomaly of the larynx.
- Laryngomalacia is presented with inspiratory stridor that starts soon after birth with normal cry. The epiglottis in laryngomalacia is omega-shaped.