2) Congenital Disorders of the Larynx - Dr. Hiwa

Introduction

- The incidence: 1: 10,000 - 1:50,000 births.
- Some of these children will have more than one anomaly in the airway.

Supraglottis

Laryngomalacia

- Partial or complete collapse of the supraglottic structures on inspiration.
- Most common congenital cause of stridor.
- Pathophysiology-------obscure.
- Anatomical abnormalities responsible for the airway obstruction
  1. The epiglottis is long and curled (omega-shaped).
  2. The aryepiglottic folds are tall & bulky, short anteroposteriorly & tightly tethered to the epiglottis.
     - The result is a tall, narrow supraglottis with a deep interarytenoid cleft.
- The epiglottis is soft and may curl and collapse.
- The mucosa and submucosa of the aryepiglottic folds may prolapse into the airway.
- There may be an element of neuromuscular immaturity and consequent incoordination of arytenoid movements.

Clinical features:

- High-pitched, fluttering inspiratory stridor at, or shortly after, birth.
- Most noticeable when the infant is active or upset, and may disappear when the child is asleep.
- The severity of the stridor increases as the child becomes more active during the first nine months of life, and then gradually diminishes until by the age of two years it has generally disappeared.
- Very rarely, stridor may persist into late childhood.

Examination:

1. Flexible fibreoptic laryngoscopy.
   - The supraglottic collapse on inspiration (typical of laryngomalacia), is easily seen but may obscure the vocal cords, and the examination provides no view below the glottis; therefore a second, coexisting airway pathology cannot be excluded.
2. Microlaryngoscopy and bronchoscopy under general anaesthesia:
   - If the stridor is severe,
   - If there is failure to thrive or
   - If there are any atypical features
   - If an adequate view cannot be obtained with the fibrescope
   - 20 percent will have another airway abnormality.
   - 5 percent this will be clinically significant.
Treatment:

- 90 percent of reported cases the condition is mild, no intervention is needed and the parents can be reassured accordingly.
- In severe laryngomalacia, there is serious respiratory obstruction and feeding difficulties, and consequent failure to thrive.
- So restoration of an adequate airway becomes necessary.
  1. Endoscopic aryepiglottoplasty (supraglottoplasty): Using Microlaryngeal surgery or alternatively a CO$_2$ laser.
  2. Tracheostomy in severe cases: If stridor and feeding difficulties persist, an underlying hypotonic neurological disorder is likely.

Saccular cysts

- An unusual lesion.
- Present with respiratory obstruction in infants and young children.
- Represents an abnormal dilatation or herniation of the saccule of the ventricle of the larynx.
- It differs from a laryngocoele in that there is no opening into the larynx and it is filled with mucus instead of air.
- Develops as the result of a developmental failure to maintain patency of the orifice between the saccule and the ventricle.
- Anterior or lateral type.
  - The anterior saccular cyst: extends medially and posteriorly from the saccule and so protrudes into the laryngeal airway between the true and false vocal cords.
  - The lateral saccular cyst: is most common in infants and expands posterosuperiorly into the false cord and Ary-epiglottic folds.

Diagnosis:

- Endoscopy
- Imaging

Treatment:

- Endoscopic marsupialization.
- If the cyst recurs----- Then by a lateral cervical approach the cyst can be completely excised.

Lymphangioma

- Cystic malformations (cystic hygromas) that result from abnormal development of the lymphatic vessels.
- Types:
  - macro cystic (usually infrahyoid),
  - micro cystic (usually suprahyoid)
  - combination of the two.
- Occasionally, a micro cystic lymphangioma may extend into the tongue base, valleculae and supraglottis, and airway obstruction may result.

Treatment:

- If very extensive……. tracheostomy.
- If less severe supraglottic involvement…debulk the lesion by endoscopic vaporization using a CO2 laser.
Glottis

Laryngeal Webs

- Failure of complete canalization of the larynx during embryogenesis results in a glottic or a supraglottic web.
- The majority involve the anterior glottis, fusing the vocal cords along a variable part of their length, and producing a variable degree of respiratory obstruction and dysphonia.

Clinical features:

- Inspiratory stridor
- Weak, high-pitched voice.
- The combination of a weak cry from birth and recurrent croup in infancy should always raise suspicion of a laryngeal web.
- Occasionally, a congenital posterior, interarytenoid web occurs, and may be associated with crico-arytenoid joint fixation.

Treatment:

- If small and causing little symptoms it is usually best to leave.
- A longer web divided endoscopically with a knife or CO\textsubscript{2} laser.
- Longer, thicker webs with an inadequate airway managed with a tracheostomy, and to be corrected at the age of three to four years via a laryngofissure technique.
- In cases where the airway is very small and there is subglottic stenosis a laryngo-tracheal reconstruction (LTR) with anterior cartilage grafting is required.

Vocal Cord Paralysis

- Second most common congenital anomaly of the larynx after laryngomalacia.
- 45 percent of patients may have other, coexisting airway pathology.
- Out patient flexible fibreoptic laryngoscopy may indicate the diagnosis.
- Microlaryngoscopy and bronchoscopy under G/A is essential.

Laryngeal ultrasound:

- An accurate method of assessing vocal cord movement.
- Useful in monitoring a child with known vocal cord palsy.
- Useful in the diagnosis of the very sick child who may be unfit for endoscopy under general anaesthesia.
- Approximately half of cases are unilateral and half bilateral.

Unilateral vocal cord paralysis:

- Usually not congenital.
- Most cases being acquired as a result of surgical injury to the left recurrent laryngeal nerve, often following correction of a congenital cardiac anomaly.
- The vocal cord lies in an intermediate position.
- Patients present with mild stridor, dysphonia and sometimes aspiration.
- Surgical intervention is not usually necessary, and the voice can be expected to improve as time passes and either recovery occurs or the other vocal cord compensates.
Bilateral vocal cord palsy:

- Usually a congenital abductor paralysis.
- The vocal cords lie in the paramedian position with consequent inspiratory stridor.
- Tracheostomy is necessary in approximately half of cases.
- Causes:
  1. A classical cause of congenital bilateral vocal cord palsy is hydrocephalus. Correction of the raised intracranial pressure with a shunt often improves vocal cord movement and a tracheostomy may thus be avoided.
  2. Most cases of congenital bilateral vocal cord paralysis are idiopathic and the approach to management is greatly influenced by the fact that up to 58 percent will eventually recover, with 10 percent taking more than five years to do so. The problem is often delayed maturation in the vagal nuclei and conservative management is favourable. The infant with an inadequate airway and failure to thrive will require a tracheostomy. If vocal cord movement does not develop and the airway does not become adequate as a result of laryngeal growth, then:
     - An endoscopic laser cordotomy or arytenoidectomy should be considered at the age of eleven or over.
     - If it fails then an external arytenoidectomy via a laryngofissure may be carried out.

Subglottis

Congenital Subglottiic Stenosis

- Due to defective canalization of the cricoid cartilage and/or conus elasticus; resulting in a small, elliptical, thickened cricoid and/or excessive submucosal soft tissue.
- Third most common congenital anomaly of the Larynx.
- Typically, there is gross thickening of the anterior lamina of the abnormal cricoid.

Mild degrees present as:

- inspiratory or biphasic stridor as the child becomes older and more active.
- or as recurrent‘croup’ owing to superimposed oedema from upper respiratory tract infections.

Diagnosis: microlaryngoscopy and bronchoscopy.

The degree of stenosis is measured by passing bronchoscopes, endotracheal tubes, dilators or bougies of different sizes through the stricture. Classification of severity:

- The Myer-Cotton grading system:
  - Grade I represents 0-50 percent obstruction.
  - Grade II 51-70 percent obstruction.
  - Grade III 71-99 percent obstruction.
  - Grade IV 100 percent obstruction.
- This helps to predict the outcome of surgical reconstruction.

If the airway is not severely compromised then surgery may not be required, as a congenital stenosis can be expected to enlarge with growth.

- Congenital cartilaginous stenosis is a contraindication to dilatation or laser resection: any type of endoscopic treatment is liable to worsen the initial condition, and attempted dilatation is inevitably ineffective as the thickened ring of cricoid cartilage cannot be expanded.
If the airway is severely compromised then a tracheostomy is needed.

- This can be avoided in specialist centres where there are facilities for single stage airway reconstruction in which an endotracheal tube is used as a stent, usually for a period of five to seven days.

**The surgical options:**

- Laryngotraceoaplasty to achieve laryngeal framework expansion.
- Laryngo-tracheal reconstruction, The LTR involves augmentation of laryngotracheal complex by anterior and/or posterior midline incision of cricoid with insertion of costal cartilage grafts to expand the airway.
- Complete resection of stenotic segment with end-to-end anastomosis of the trachea to the thyroid cartilage.

**Subglottic Haemangioma**

*Def:* A capillary hamartoma which enlarges rapidly up to the age of about one year and then involutes slowly. It is life-threatening because of its situation in the narrowest part of the airway and if untreated carries a mortality rate of 50 percent.

*C.F:* 85 percent present within first six months of life.

- By gradually increasing stridor, which peaks at the age of six weeks.
- Endoscopy --- appearance of a compressible, pear-shaped red swelling in the subglottis on one side (left > right).
- Larger haemangiomas may be circumferential and, may extend down into the trachea or through its wall into the surrounding soft tissues of the neck or mediastinum. Where MRI becomes indicated.
- Biopsy is unnecessary.

**Treatment:**

1. Systemic steroids --- dramatic regression of subglottic haemangiomas.
2. Intralesional steroid injection followed by intubation for seven days.
4. Radiotherapy delivered by low-dose external beam or gold-grain implant.
5. CO2 laser vaporization can be effective but is probably only suitable for small lesions.
6. Submucosal excision of the haemangioma combined with a cricoid split to decompress and expand the subglottis.
7. Tracheostomy will maintain the airway until involution occurs.

**Conclusion:**

- Very small haemangiomas may not require treatment, or may be amenable to CO2 laser vaporization.
- Medium-sized lesions seem suitable for intralesional steroids and intubation.
- Large ones are probably best managed by primary submucous resection.
- Very large haemangiomas, more safely dealt with by performing a tracheostomy and awaiting spontaneous involution.