LARINGOMALACIA

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ABSTRACT:

Laryngomalacia is existing in the manifestation of stridor, a high-pitched, musical, vibrating, multiphase inspiratory noise occurring within the first 10 days of life. Recognizing symptoms that determine disease severity helps anticipate end results. Infants with stridor who do not have remarkable feeding-related symptoms can be managed confidently, without intervention. Infants, who have stridor and feeding-related symptoms, are managed from acid suppression therapy. Those with added symptoms of aspiration, failure to thrive, and consequences of airway obstruction and hypoxia require surgical intervention. Most with laryngomalacia will have mild-to-moderate symptoms and not require surgery.

***Key words**: laringomalacia, stridor, gastroesophageal reflux.*

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INTRODUCTION:

Congenital anomalies are the effect of misconception in embryogenesis (malformations) or the outcome of intrauterine events that influence embryonic and fetal growth (deformations and disruptions).

Defects in the formation and growth of the larynx lead to laringomalacia. The causes, types, embryology, pathophysiology, clinical features, diagnostic investigation, management and nursing care plan for laringomalacia are reviewed here.

Laryngomalacia refers to collapse of the supraglottic structures during inspiration. It is important to identify the laringomalacia in early infancy as it may affect the growth and development. Clinical manifestations include stridor that worsens in the supine position and during crying, feeding, and upper respiratory infections. Laryngomalacia is suspected based upon the history and physical examination, and confirmed with flexible fiberoptic laryngoscopy. Infants who have stridor with feeding difficulty, dyspnea, tachypnea, cyanosis, or apnea should be referred to an otolaryngologist for further evaluation and possible intervention.

DEFINITION:

It is a congenital condition in the form of weak laryngeal tissue above the vocal cords. Noisy breathing is commonly caused by a floppy voice box which a condition is called laryngomalacia.

Incidence: More than 50 % infants with laringomalacia have noisy breathing during the first week of life, and remaining infants have developed by 2-4 weeks of age. Infrequently, it occurs in older children. It is evaluated to be around one in 2100-2600 children.

Causes:

- Unknown
- It may be inherited. It is an autosomal dominant.
  - An important gene associated with Laryngomalacia is WFS1 (Wolframin ER Transmembrane Glycoprotein) and MECP2.
  - WFS1 provides instructions for producing a protein called wolframin that is thought to regulate the amount of calcium in cells.
  - MECP2 gene is essential for normal function of nerve cell.
- Lack of muscle tone in the upper airway
- Acid reflux
- Vitamin D deficiency may be a factor

Types:

- Mild laryngomalacia – baby's breathing sounds do not interfere and do not cause health problems. It's usually outgrown within 18 months.
  moderate- vomiting, blockage of the airway, some problems with feeding (difficult to suck), regurgitation, and mild or moderate chest retractions. 99% of infants have mild or moderate type of laringomalacia.
- Severe: Apnea, cyanosis, failure to thrive.

Holinger's classification:

- Type 1: anterior prolapse of the arytenoid and corniculate cartilages;
- Type 2: tubular epiglottis which curls on itself, often associated with type 1;
- Type 3: anteromedial collapse of the arytenoids;
- Type 4: posterior prolapse of the epiglottis;
- Type 5: short aryepiglottic folds.

Congenital or acquired laringomalacia:

Acquired laryngomalacia is occur in both older children and adults. It has usually been described in the context of neurological events or coma and can be reversible in some cases, reinforcing the neuromuscular hypothesis of congenital laryngomalacia, although these acquired cases usually exclusively concern the epiglottis that becomes flaccid and able to invaginate between the vocal cords.

Embryology:

- The larynx develops from the **endodermal lining** and the adjacent mesenchyme of the foregut
- The endodermal tissues of the cranial end of the laryngotracheal tube made up the epitheal lining of the larynx.
- The larynx has cartilages surrounding it and it is develop from the mesenchyme of the fourth and the sixth pairs of pharyngeal arches. It is from the neural crest cells.

- At 20 days' gestation, the foregut is first identifiable with a ventral laryngotracheal groove. The laryngotracheal groove continues to deepen until its lateral edges fuse
- Arytenoid swelling is formed at the cranial end of the laryngotracheal tube by the proliferation of the mesenchymal tissue (derived from neural crest cells).
- Transverse fold at the cephalic rim of the opening. This fold is the anlage of the epiglottis.
- Laterally the epiglottic fold becomes continuous with the arytenoid ridges, forming the ary epiglottic ridges.
- Arytenoids swelling will grow towards the tongue and forms the primordial glottis. As it grows further, it changes the primordial glottis into a T-shaped laryngeal inlet**.**
- The T-shaped laryngeal slit (aditus) is formed anteriorly by the growth of the primordium of the epiglottis (from the hypobranchial eminence, arches III and IV) and laterally by the precursors of the arytenoid cartilages (ventral ends of arch VI)
- The fifth and sixth weeks of gestation, the tracheoesophageal septum extends to the first tracheal ring.
- By the seventh week of gestation, the cricoid ring is complete, and the cartilaginous hyoid is visible below the epiglottis.
- Temporary occlusion of the laryngeal inlet occurs due to rapid proliferation of the epithelial lining of the larynx.
- By the 10th week of gestation, recanalization occurs and consequently, pair of laryngeal ventricles are formed.
- The laryngeal ventricles are bound by tissue that progress into false and true vocal cords.
- Proliferation of the mesenchyme in the ventral part of the 3rd and 4th pharyngeal arches will formed hypopharyngeal eminence with later form the epiglottis.
- The superior laryngeal nerve will supplies the part of larynx that develop from the 4th pharyngeal arches. The recurrent laryngeal nerve will supplies the part of larynx that derives from the 6th pharyngeal arches. The 2 nerves are branches of the vagus nerve.

Pathophysiology:

- The folds of tissue between the front and back of the voice box (aryepiglottic folds) are shortened. (Infolding of the aryepiglottic folds)
- It leads to epiglottis to curl inward and become omega-shaped and causes the tissue over the cartilage in the back of the voice box (arytenoids) to suck into the airway.
- supraglottic structures that can invaginate during inspiration
- poor control of the tone of supraglottic structures (either pathological neurological mechanisms or simple physiological variations related to changes in tone during sleep), mucosal oedema, and increased airflow.
- Upper larynx collapses inward during inhalation.

Clinical manifestations:

- Dyspnea
- Feeding difficulties, failure to thrive, cyanosis.
- Airway obstruction
- stridor (noisy breathing) - at the level of the voice box obstruction occur
- sleep apnea.

Diagnosis:

- History –complete birth history, any surgical procedure or intubation, history of breathing difficulty
- Physical examination – focus on head, neck and oral cavity
- Neck and chest X-ray
- Flexible laryngoscopy and Nasopharyngoscopy.- Fully evaluate the dynamics of the voice box
- Flexible fiberotic laryngoscopy –to see the folds of the vocal cords
- Airway fluoroscopy
- Direct laryngoscopy
- Bronchoscopy
- Functional endoscopic evaluation of swallow (FEES)
- Polysomnogram - to identify the level of obstructive sleep apnea

- Endoscopy - visualization of more or less complete collapse of the supraglottis, anterior prolapse of the arytenoid cartilages and posterior prolapse of the epiglottis which can be curled up to form a tubular structure.

Management:

- Mild laryngomalacia: baby can usually recover from stridor at the age of 12 months to 18 months
- Intermediate: omeprazole - to treat gastroesophageal reflux disease (GERD)
- Severe – need extra oxygen for breathing
- CPAP to treat sleep apnea.

Management to reduce GERD:

- Elevate the head end while breast feeding,
- Stays upright for atleast 30 minutes after breast feeding
- After feeding burp the child
- Avoid giving juices or foods with sour taste such as orange juice, lemon juice and grape juice
- Don't keep the baby on his back with the bottle in his mouth.
- Surgery for GERD: Nissen fundoplication (a procedure to tighten the upper valve of the stomach.

Surgery:

- **Tracheostomy:**
- The tracheostomy tube varies in size and type depending on the child's airway size and health and the length of time the child will require the tracheostomy.
- Silastic tracheostomy tubes are soft and flexible; they are available with a single lumen or may have an outer and inner lumen.
- Both types have an obturator (the guide used during tube changes). Typically, the tubes with inner cannulas are used with older children and in children with increased mucus production.
- Cuffed tracheostomy tubes are generally used in older children also. The cuff is used to prevent air from leaking around the tube.
- The funnel-shaped airway in younger children acts a physiological cuff and prevents air leak.
- Complications immediately postoperatively include hemorrhage, air entry, pulmonary edema, anatomic damage, and respiratory arrest.
- At any point in time the tracheostomy tube may become occluded and ventilation compromised.
- Complications of chronic tracheostomy include infection, cellulitis, and formation of granulation tissue around the insertion site.
- Tracheostomies require frequent suctioning to maintain patency.
- **Supraglottoplasty :**
- It can be performed bilaterally –(dividing the cartilage of the larynx and the epiglottis).
- Multiple techniques are used in this procedure, including the use of cold steel, a laser laryngeal microdebrider, or coblator.
- supraglottoplasty procedure should be individualized to the particular patient's anatomy but may consist of dividing shortened aryepiglottic folds, removing redundant arytenoid mucosa, performing an epiglottopexy, or a combination of these.
- Care should be taken to avoid the interarytenoid mucosa, as glottic stenosis can occur due to scarring of this area.

Prevention:

Cannot prevent laringomalacia, but prevent medical emergencies related to this condition. Know the signs and symptoms of laringomalacia, carefully watching the babies feeding pattern and breathing. If note any deviation in feeding and breathing immediately consult the paediatrician.

Nursing care plan:

1. Ineffective air way clearance related to air way obstruction

Interventions: maintaining a patent airway

• Position with airway open (sniffing position if supine): open airway allows adequate ventilation.

• Humidify oxygen or room air and ensure adequate fluid intake (intravenous or oral) to help liquefy secretions for ease in clearance.

• Suction with bulb syringe or via nasopharyngeal catheter as needed, particularly prior to bottle-feeding to promote clearance of secretions.
• If tachypneic, maintain NPO status to avoid risk of aspiration.
• Perform chest physiotherapy if ordered to mobilize secretions.
• Ensure emergency equipment is readily available to avoid delay should airway become unmaintainable

2. Ineffective breathing pattern related to anatomical defect

Interventions: promoting effective breathing patterns
• Assess respiratory rate, breath sounds, and work of breathing frequently to ensure progress with treatment and so that deterioration can be noted early.
• Use pulse oximetry to monitor oxygen saturation in the least invasive manner to note adequacy of oxygenation and ensure early detection of hypoxemia.
• Position for comfort with open airway and room for lung expansion and use pillows or padding if necessary to maintain position to ensure optimal ventilation via maximum lung expansion.
• Administer supplemental oxygen and/or humidity as ordered to improve oxygenation.
• Allow for adequate sleep and rest periods to conserve energy.

3. Imbalance nutrition less than body requirement related to difficulty feeding as evidenced by poor oral intake, tiring with feeding secondary to dyspnea
   Interventions: promoting adequate nutritional intake
   • Weigh on same scale at same time daily: weight gain or maintenance can indicate adequate nutritional intake.
   • Calorie counts over a 3-day period are helpful in determining if caloric intake is sufficient.
   • Assist family and child to choose higher-calorie, protein-rich foods to optimize growth potential.
   Advised the mother to give breastfeeding after feeding burp the child to prevent aspiration

4. Parental fear and anxiety related to difficulty in breathing, unfamiliar procedures

Interventions: relieving fear
• Establish trusting relationship with child and family to decrease anxiety and fear.
• Explain procedures to child at developmentally appropriate level to decrease fear of unknown.
• Provide comfort measures preferred by client such as rocking or music.
• Involve parents in care to give child reassurance and decrease fear

5. Impaired Family processes related to child's illness or hospitalization as evidenced by family's presence in hospital, missed work, demonstration of inadequate coping
   . Interventions: promoting adequate family processes
   • Encourage parents' verbalization of concerns related to child's illness: allows for identification of concerns and demonstrates to the family that the nurse also cares about them, not just the child.
   • Explain therapy, procedures, and child's behavior to parents; developing an understanding of the child's current status helps decrease anxiety.
   • Encourage parental involvement in care so that parents may continue to feel needed
6. Parental Knowledge deficit related to unknown condition
   - Focus teaching sessions on a single concept or idea.
   - Pace the instruction and keep sessions short.
   - Use the teach-back technique to determine the patient's understanding of what was taught:
   - The nurse gives information in a caring manner, using plain language.
   - Ask the patient to explain in his or her own words.
   - Rephrase the information if unable to repeat it accurately.
   - Again ask the parent to teach-back the information using his or her own words until the nurse is comfortable that is understood.
   - If the parent still does not understand, consider other strategies.

CONCLUSION

Laryngomalacia mild disease can be managed easily. Continued monitoring of the symptoms are necessary, it helps to determine which child will need otolaryngology consultation. Identify the disease severity and outcomes are an important aspect for care givers according to that give care to the child with laryngomalacia.

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