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Case Based Reviews in Pediatric Pulmonology









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SECTION

Congenital Disorders of Respiratory Tract

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CHAPTER

10

Laryngomalacia and Tracheomalacia

Paramesh D H, Salim A Khatib

LARYNGOMALACIA

CASE 1

- First full-term normal delivery (FTND) male, presented with inspiratory stridor soon after birth, associated with chest wall retractions
- Had normal air entry with resting SpO_2 of 96% in room air, required supportive care initially. After 24 hours, the baby started accepting breastfeeds. At times, the stridor was high pitched by agitation and crying, and diminished during sleep. Followed up to 6 weeks baby is thriving well.

Q. What are the causes of purely inspiratory stridor in a well baby?

Common causes for stridor in otherwise normal newborn are as follows:

- Laryngomalacia
- Vocal cord palsy
- Supraglottic cyst, laryngeal cyst
- Subglottic stenosis
- Airway wall hemangioma
- Laryngeal cleft, laryngeal web
- Lingual thyroid
- Hypocalcemia and hypomagnesemia

Q. Possible approach to evaluate baby with inspiratory stridor?

The most common cause is laryngomalacia (60–70%), where it is mild and purely inspiratory stridor. It is with normal cry and feeding well with good thriving starts soon after birth in an otherwise well baby. It is a self-limiting condition and resolves without any intervention by 12–18 months.

If there is any doubt about the cause of the stridor, associated with poor feeding, hoarse cry, extended neck, and poor growth, the following investigations can be considered:

- Chest X-ray: To assess the mediastinum, cardiac size, and position of aorta
- Laryngobronchoscopy: To visualize the airway structures
- Blood tests: Serum electrolytes, metabolic disorders such as hypocalcemia, and hypomagnesemia.

Q. What are clinical manifestations of laryngomalacia?

Laryngomalacia is the term used to describe a condition in which there is partial or complete airway closure caused by an inward folding of the aryepiglottic folds and epiglottis during inspiration.

Laryngomalacia is characterized by following clinical manifestations:

- Purely an inspiratory stridor, if there is any expiratory component, one should seek an alternative diagnosis
- The stridor may be present from birth, but generally becomes apparent by several weeks of age (4–8 months)

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- Child has normal cry and thriving well
- The stridor is staccato and crescendo, increasing in loudness and pitch toward the end of inspiration. It is variable and may be absent during sleep
- When laryngomalacia is severe, there may be failure to thrive and poor feeding
- In approximately 5–20% respiratory concerns such as obstructive sleep apnea, tachypnea, dyspnea, respiratory distress, or hypoxia can occur.

Q. What is etiology of laryngomalacia?

Precise etiopathogenesis of laryngomalacia is not known.

- Theory of immature and abnormally collapsible cartilage was further discredited by histological examinations demonstrating normal fibroelastic cartilage tissue in children with symptomatic disease
- Presently, attention has focused on a neuromuscular etiology, consisting of immaturity or abnormal integration of the peripheral nerves, brainstem nuclei, and pathways responsible for swallowing and maintenance of airway patency
- Additional corroboration is provided by histological studies showing significant size difference in the superior laryngeal nerve branches of patients with severe laryngomalacia, compared to age-matched controls
- Subsequent neurological and central nervous system maturation would provide a reasonable explanation for the spontaneous resolution generally seen in the disease.

Q. How to make a diagnosis of laryngomalacia?

- A presumptive clinical diagnosis of laryngomalacia can be made based on the classic symptoms of inspiratory stridor worsened by feeding, agitation, supine positioning, or crying
- A definitive diagnosis can be made accurately by flexible fiber optic laryngoscopy alone in vast majority (88%) (Fig. 1)
- Several anatomical staging systems for laryngomalacia have been proposed which focus on defining the site of collapse:
  - Posterior collapse (from redundant arytenoids mucosa or cuneiform cartilage)
  - Lateral collapse (from shortened aryepiglottic folds)
  - Anterior collapse (from retroflexed epiglottis)
  - Combined collapse (involving multiple areas of anatomic collapse).

CASE 2

- A 5-day-old girl child presented with a history of respiratory noises and at times feeding difficulty
- Examination revealed inspiratory stridor with accessory muscles in action. Clinical diagnosis of laryngomalacia was made and reassured along with positional support for breastfeeding
- Two days later, the baby presented with severe respiratory distress, with all the accessory muscles in action, and increased intensity of stridor with SpO_2 of 88% in room air
- After evaluation, diagnosed to have severe form of laryngomalacia near complete closure during inspiration and underwent surgical intervention—supraglottoplasty and did very well.

Q. Is there any laryngomalacia severity scale?

Based on respiratory symptoms and feeding symptoms, laryngomalacia can be assessed as mild, moderate, or severe (Table 1).

Q. Any additional diagnostic evaluation is necessary?

In mild to moderate severity of laryngomalacia, no additional evaluation is necessary.

- Gastroesophageal reflux disease has been reported in 65–100% of infants with severe laryngomalacia
- Reflux events could then irritate the laryngeal mucosa, causing edema, and thereby worsening the airway collapse. The incidence of synchronous airway lesions is 12–64%.

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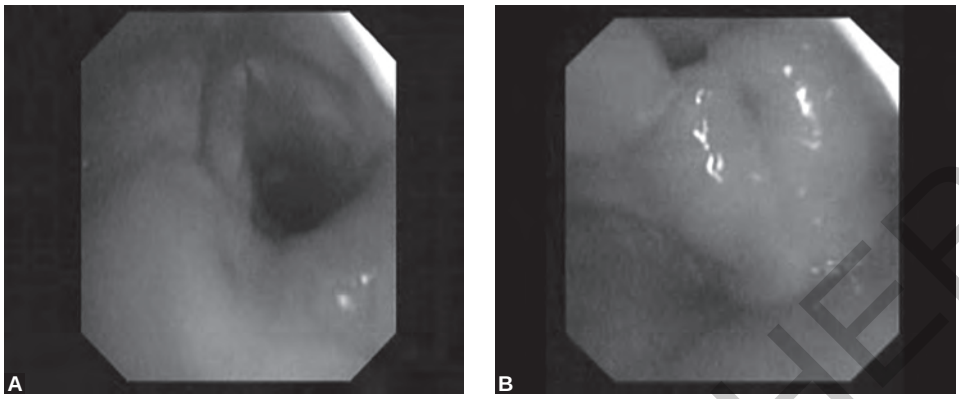


Figure 1: Fiber optic laryngoscopic view of child with laryngomalacia. **A,** during expiration and **B,** during inspiration *(For color version, see Plate 2)*

TABLE 1: Assessment of severity of laryngomalacia

Severity level	Respiratory symptoms	Feeding symptoms
Mild	Inspiratory stridor (average resting SpO_2: 98–100%)	Inspiratory stridor with cyanosis or apnea (average resting SpO_2: 86%)
Moderate	Inspiratory stridor (average resting SpO_2: 96%)	Frequent regurgitation or other feeding issues
Severe	Inspiratory stridor with cyanosis or apnea (average resting SpO_2: 86%)	Failure to thrive or aspiration

Q. What are clinical indications for consideration of surgical intervention?

Symptoms of mild to moderate laryngomalacia improve with age. Children with severe disease having respiratory and feeding problems may need surgical intervention (Table 2).

TABLE 2: Indications for surgery in an infant with severe laryngomalacia

Respiratory*	Feeding*
• Stridor with respiratory distress • Dyspnea with retractions • Pulmonary hypertension • Severe obstructive sleep apnea	• Episodic cyanosis with feeding • Recurrent aspiration pneumonia • Failure to thrive

*Presence of single manifestation is indication for surgery.

TRACHEOMALACIA

CASE 3

- Baby S, 27-month-old girl presented with history of fever, cold, and cough of 3 days, which rapidly progressed to severe respiratory distress. Her clinical findings, radiological picture, was with acute respiratory distress syndrome. She was treated with oseltamivir and antibiotics. Her ventilation requirement was high and required 4 weeks of ventilation
- She was extubated onto noninvasive ventilation. However, she continued to have inspiratory stridor, and marked recessions which were variable during the course of the day. Bronchoscopy confirmed significant tracheomalacia (portion below cricoid bone)
- Tracheostomy was undertaken and the tracheostomy tube bypassed the malacia part and her distress was reduced.

Q. What is tracheomalacia?

Malacia refers to "softness" of a tissue, typically a bone or cartilage. Collapsibility of the central airways is an important cause of obstruction in childhood respiratory conditions such as tracheomalacia, bronchomalacia, and tracheobronchomalacia (TBM).

Q. What is pathogenesis of tracheomalacia?

To understand pediatric tracheomalacia, it is first necessary to understand the development and structure of the airway as well as airway mechanics.

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Airway development

By the third week of gestation the endoderm develops into foregut. The trachea develops from the proximal ventral foregut endoderm while the esophagus arises from the dorsal foregut endoderm. The separation of the two structures is completed by the end of fourth week of gestation. Improper foregut separation of the trachea from the esophagus results in congenital abnormalities such as laryngo-tracheoesophageal clefts, tracheoesophageal fistulae (TOF), and esophageal atresia (EA) that are associated with tracheomalacia.

The posterior membrane or pars membranacea contains the trachealis muscle, which also changes throughout development. There is an age-related increase in the passive tone, contractility, size and length of the trachealis.

The net effect of the age-related changes to the tracheal rings and posterior membrane is an increase in radius, cross-sectional area (CSA), and circumference of the trachea and a decrease in tracheal compliance. Weakness of either the cartilage or posterior membrane results in increased tracheal compliance and possible compromise of the tracheal lumen during the respiratory cycle.

Q. Can we clinically differentiate between extrathoracic and intrathoracic tracheomalacia?

Extrathoracic dynamic airway collapse causes inspiratory symptoms including stridor, a prolonged inspiratory phase, and low lung volumes.

Intrathoracic airway narrowing whether generalized or localized causes expiratory problems including wheezing, barking cough, a prolonged expiratory phase, and impaired secretion clearance. Children with intrathoracic tracheomalacia can also experience recurrent bacterial bronchitis, cyanotic spells, apnea, and difficulty weaning from respiratory support (Fig. 2).

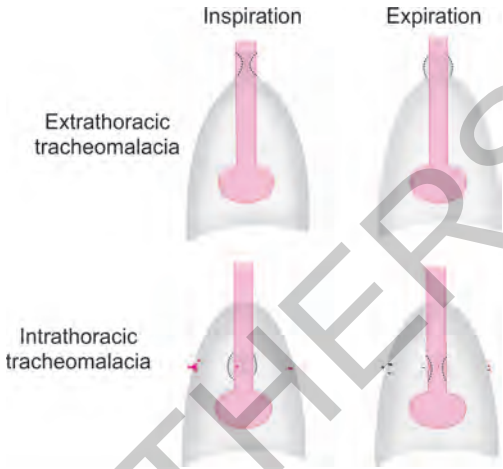


Figure 2: Airway mechanics in intrathoracic and extrathoracic tracheomalacia

Q. How to confirm diagnosis of tracheomalacia?

Tracheomalacia should be suspected in infants at risk when their disease is unusually severe or persistent, where there is wheeze that does not respond to bronchodilators, or where there are episodes of unexplained respiratory distress and cyanosis.

On physical examination, one can appreciate inspiratory stridor with retractions in extrathoracic tracheomalacia and wheezing with retractions in intrathoracic tracheomalacia.

Most authors agree that 50% narrowing of the airways is diagnostic of tracheomalacia by the bronchoscopist during visual inspection (Fig. 3).

More recently, efforts have been made to quantify tracheal collapse accurately using the color histogram method. This technique objectively identifies the airway lumen and determines CSA by accounting for the magnification and distortion of the bronchoscopic image.

Another challenge in the diagnosis of tracheomalacia is that current methods only evaluate the change in CSA without taking the applied transmural pressure into account.

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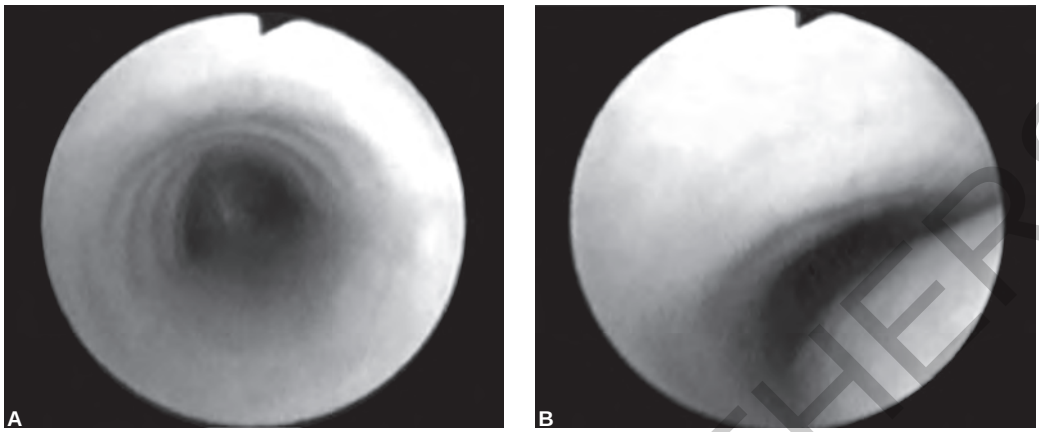


Figure 3: Bronchoscopic findings. **A,** in normal trachea and **B,** in tracheomalacia ***(For color version, see Plate 2)***

CASE 4

- A 1-year-old baby boy, who was diagnosed with large patent ductus arteriosus (PDA) and pulmonary arterial hypertension was referred for evaluation with significant respiratory distress
- His presenting features were shortness of breath, feeding difficulty, and failure to thrive
- On examination, he had hyperinflated chest, recession, and diminished breath sounds in left hemithorax. X-ray confirmed hyperinflation of the left lung
- Bronchoscopy showed narrowing of the left main bronchus in fish-mouth pattern. The computed tomography (CT) scan showed compression of trachea at carina and left main bronchus (Fig. 4). He underwent PDA ligation. He continued to have noisy breathing and check bronchoscopy showed persistent malacia
- A diagnosis of tracheobronchomalacia secondary to compression of airways due vessels was made and symptomatic treatment was given. He slowly showed improvement and his symptoms resolved after 2 years.

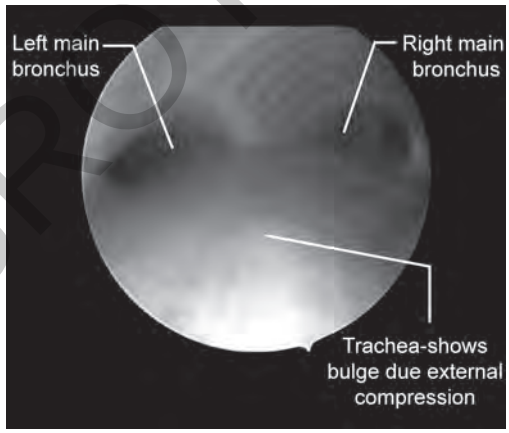


Figure 4: Bronchoscopic findings. Compression of left main bronchus ***(For color version, see Plate 2)***

Courtesy: Anilkumar Sapare, NH Hospital, Bengaluru.

Q. What are the probable causes of tracheobrochomalacia?

Tracheomalacia, or TBM can be primary (congenital) or secondary (acquired). Primary disease is rare.

Primary tracheomalacia was first described by Holinger et al. in 1952, and later defined as weakening of the tracheal wall due to abnormal formation or maturation of the airway.

Secondary or acquired tracheomalacia occurs in the normally developed trachea after some insult such as trauma, external compression, and positive pressure ventilation (PPV), infection or inflammation.

Tracheotomy or prolonged intubation is perhaps the most common cause of acquired extrathoracic tracheomalacia.

The earliest reports of secondary tracheomalacia were described in the 1930s and 1940s and were associated with vascular compression

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of the central airways, i.e., vascular ring. Later, Gross and Neuhauser noted that this type of deformity of the airways persisted even after the vascular compression was alleviated.

External compression can also occur by axial skeleton as seen in severe scoliosis and pectus excavatum or by space occupying lesions such as goiter, tumors, and cysts.

This baby has congenital heart disease—PDA, pulmonary hypertension with cardiomegaly. The mechanism for the TBM in such children is not well-understood.

Compression of the bronchus (left) by a large heart seems to be the main culprit.

The effects of the TBM can be worsened by increased airway resistance caused by heart failure as well.

The picture below shows narrowing of left main bronchus, which is commonly involved with dilated pulmonary arteries, e.g., pulmonary arterial hypertension, absent pulmonary valve syndrome, and truncus arteriosus.

Treatment

- Tracheomalacia is often self-limited and will resolve or become asymptomatic by the second year of life without intervention
- For patients who are symptomatic of central airways collapse, available treatments include pharmacotherapy, positive pressure application, and surgery
- Pharmacotherapy is directed at increasing the tone of the trachealis muscle, as its contraction has been shown to decrease tracheal compliance. Type 2 and 3 muscarinic receptors play important roles in governing airway smooth muscle tone. Muscarinic agonists—bethanechol, and methacholine directly stimulate airway smooth muscle and improve airway mechanics in children with tracheomalacia
- Patients with tracheomalacia can also be treated with continuous positive airway pressure (CPAP) which provides distending intraluminal pressure which stents open the airway during expiratory maneuvers
- Continuous positive airway pressure can be provided via noninvasive ventilation and via invasive ventilation with either endotracheal intubation or tracheostomy placement. Invasive ventilation can have the added benefit of supporting the malacic tracheal segment with the artificial airway
- Surgical intervention is occasionally necessary to alleviate symptoms and sequelae of tracheomalacia. Aortopexy is a surgical procedure in which the aortic arch is fixated to the sternum. It results in the tracheal lumen being pulled open. It is used to treat severe tracheomalacia or tracheal compression. It is the mainstay of surgical interventions in pediatric intrathoracic tracheomalacia and can provide symptomatic relief
- While patients demonstrate clinical improvement, the collapsible segment of the airway remains. More recently, tracheomalacia associated with TOF has been treated using slide tracheoplasty. This technique removes the affected portion of the airway and can be an attractive method for treatment of short segments of severe tracheomalacia
- In adult patients, silicone stents have been used to stabilize the airway with improvement in respiratory symptoms; however, airway stents are infrequently used in pediatric patients due to the tendency for stents to migrate. While surgical interventions can improve symptomatic tracheomalacia, these methods should be reserved for the most severe cases.

LEARNING POINTS

- Laryngomalacia is the most common cause of inspiratory stridor in infants
- Definitive diagnosis can be made by flexible laryngoscopy without sedation
- Most children can be managed conservatively with close monitoring
- Additional diagnostic evaluations should be considered in severe form of laryngomalacia
- Surgical intervention—supraglottoplasty is now considered the mainstay of treatment for severe form of laryngomalacia
- Tracheomalacia results in excessive dynamic collapse of the central airway that results in clinical symptoms
- Extrathoracic tracheomalacia causes retractions with inspiratory stridor

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- ☞ Intrathoracic tracheomalacia causes retractions with expiratory wheeze and impaired airway secretion clearance
- ☞ Acquired tracheomalacia occurs in the normally developed trachea due to trauma, external compression, or airway inflammation
- ☞ Although tracheomalacia can be suspected by history, physical examination, and supportive radiographic findings, flexible fiberoptic bronchoscopy remains the "gold standard" for diagnosis
- ☞ Current treatment strategies involve pharmacotherapy with cholinergic agents, positive pressure ventilation, and surgical repair.

SUGGESTED READINGS

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Case Based Reviews in Pediatric Pulmonology

Key Features

- Comprehensive coverage of all aspects of pediatric pulmonology in a crisp question and answer format
- The book focuses on the assessment and treatment of pulmonary disorders commonly encountered in clinical practice
- The book has been divided into various sections covering disorders of upper respiratory tract, congenital disorders of respiratory tract, wheezing disorders in children, respiratory tract infections, interstitial lung diseases, pleural diseases, and miscellaneous respiratory disorders
- Each section has various chapters that begin with a case and take the how-to-do-it right approach for managing the patient
- Each chapter is supported by key points that summarize the discussion
- Written by experts with extensive research and clinical experience
- An ideal reference book for all pediatricians, pulmonologists, physicians, and postgraduate students involved in the diagnosis and treatment of respiratory diseases.

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