Fellow Column: Neonatal and Infant Upper Airway Malformations

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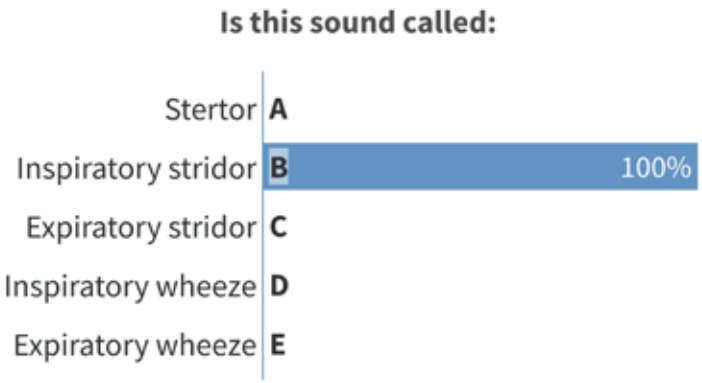
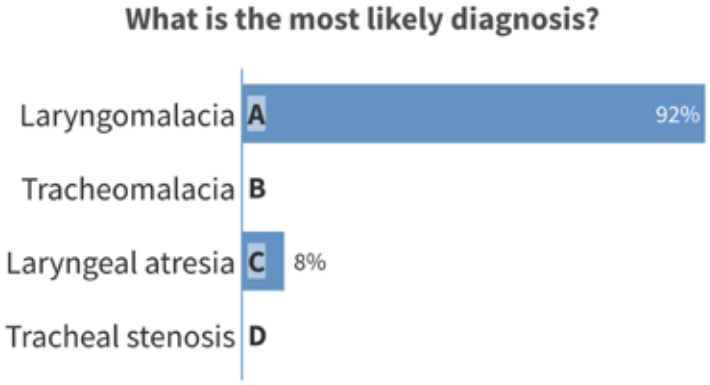
Editors Note: This Fellow's column was adapted from an academic presentation referred to as Super Tuesday, held weekly at Loma Linda University Children's Hospital by residents for their colleagues. Answers follow the audience response survey.

Q.
An infant presents with noisy breathing since birth. On exam, you hear a high pitched continuous sound when she inhales. On auscultation, the sound is best heard over her neck. What is this sound called?



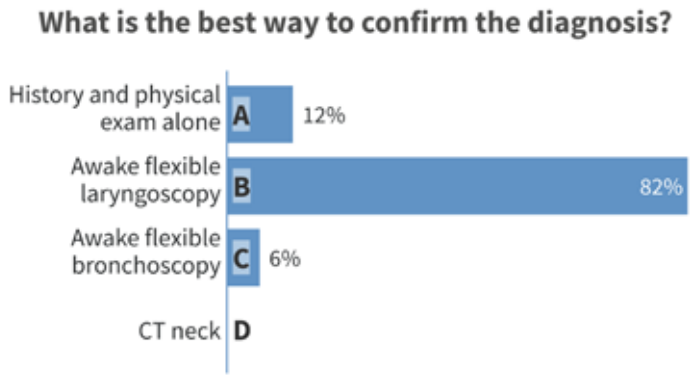
A.
The infant is presenting with inspiratory stridor. Stridor is a continuous musical sound produced in the larynx, best heard over the neck. Inspiratory stridor is produced by an upper airway lesion, such as laryngomalacia or vocal cord paralysis. It is heard during inspiration when the pressure inside the large airways falls, allowing the collapse of the airway. On the other hand, expiratory stridor is typically caused by a lower airway lesion in the thorax when intrathoracic pressure increases during expiration. Examples of lesions that cause expiratory stridor are tracheal stenosis and tracheomalacia. Another sound that may be confused with stridor is stertor, which is a snoring-like, low pitched sound typically due to a lesion in the nasopharynx. On the other hand, wheezing is multiple low or high pitched musical notes most often produced by the smaller airways and is best heard with lung auscultation.

Q.
A 6-month-old female is brought to your clinic by her mother because of worsening noisy breathing since birth. The sound is louder when she is lying down on her back. Her mother has tried nasal bulb suctioning and humidifiers without improvement. Throughout the exam, you appreciate inspiratory stridor. The sound worsens when she cries. On auscultation, it is best heard over her neck. What is the most likely diagnosis?



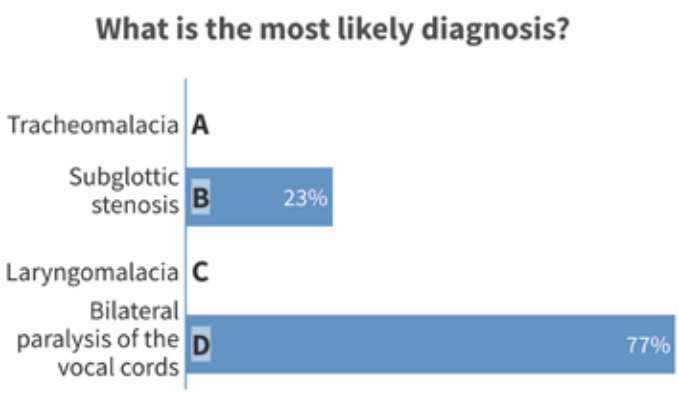
A.
Laryngomalacia is caused by delayed maturation of the cartilaginous structures of the larynx, resulting in the collapse of supraglottic structures during inspiration. This produces inspiratory stridor, which worsens with feeding, sleeping, and agitation. Although inspiratory stridor can be heard at birth, it is more commonly heard at 4-6 weeks of life after inspiratory flow rates have increased high enough to generate sounds. Symptoms usually peak around 6-8 months and remit by two years old. Treatment for laryngomalacia depends on the severity. For mild disease, usually, no treatment is necessary as most infants will outgrow it by 12-24 months. However, frequent monitoring of adequate weight gain is recommended. Treatment of the moderate disease is mainly medical management with acid suppression to decrease airway inflammation, speech and swallow therapy, and high caloric formula. Severe disease requires referral to Pediatric ENT for surgical intervention.

Q.
What is the best way to confirm a diagnosis of laryngomalacia?



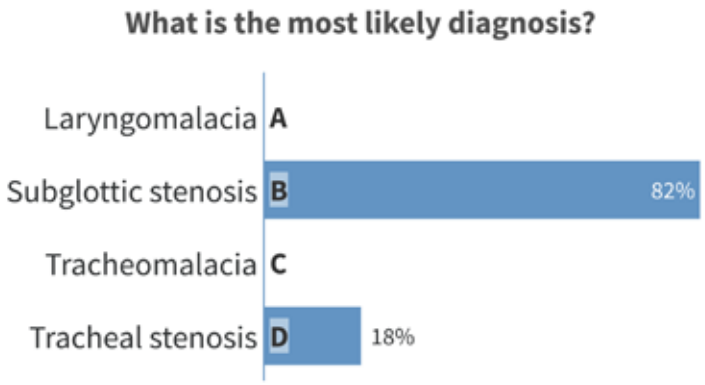
A.
Laryngomalacia can be diagnosed with a history and physical exam alone. However, the gold standard for diagnosis is awake flexible laryngoscopy to directly visualize the airway, which will reveal omega-shaped epiglottis that prolapse over the larynx during inspiration.

Q.
A 1-month-old boy presents to the clinic. He was born full-term via spontaneous vaginal delivery with shoulder dystocia. He has had inspiratory stridor and feeding difficulty since birth. The stridor is associated with retractions when the infant becomes agitated. Physical examination reveals a weight of 2.8kg (3rd percentile), no expiratory stridor, and a weak cry. What is the most likely diagnosis?



A.
Vocal cord paralysis can be an idiopathic congenital anomaly but is also associated with birth trauma, intubation, or surgery involving the neck and chest surgery. It can also be secondary to neurological disorders such as Arnold Chiari malformations and intracranial structural anomalies such as hydrocephalus, intracranial tumor, or corpus callosum agenesis. Patients present with inspiratory stridor and a weak cry. They can also have feeding difficulties and aspiration due to their inability to protect their airway adequately. Vocal cord paralysis is diagnosed with awake fiberoptic nasopharyngoscopy to visualize the movement of vocal cord during respiration directly. In mild cases, it can be treated with dietary modifications or NG feeds, but in severe cases, surgical intervention may be necessary, which include vocal cord injections, nerve grafting procedures, or tracheostomy.

Q.
A 3-month-old boy presents to the clinic. He was born via spontaneous vaginal delivery with meconium-stained fluid and was subsequently intubated for two weeks due to meconium aspiration syndrome. He has had biphasic stridor since hospital discharge. The stridor is associated with retractions when the infant becomes agitated. He has had frequent illnesses and has been to the emergency room twice since birth for croup. What is the likely diagnosis?



A.
Subglottic stenosis is due to the narrowing of the lumen of the cricoid region. It is diagnosed when the diameter is less than 4mm in term infants and less than 3 mm in preterm infants. 5% of the cases are congenital, and 95% of the cases are acquired. It is often secondary to prolonged intubation or associated with Down syndrome patients. Patients usually present with recurrent croup and have biphasic (both inspiratory and expiratory) stridor on physical exam. Subglottic stenosis is diagnosed with direct laryngoscopy or bronchoscopy and is graded into four classifications based on the severity of the stenosis. Patients typically outgrow the stenosis if it is mild. It may only need frequent monitoring or brief periods of supplemental oxygen. Treatment for severe disease is surgical intervention.

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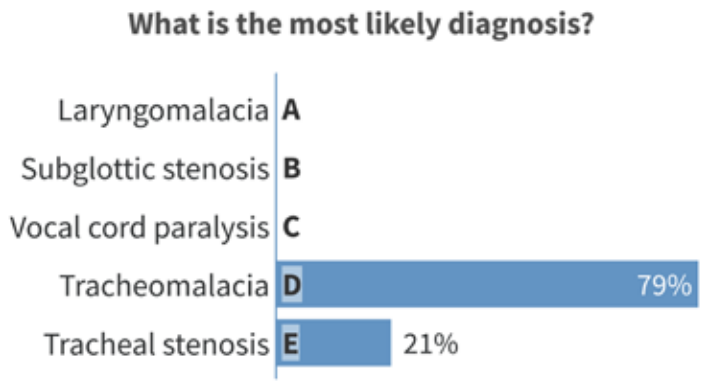
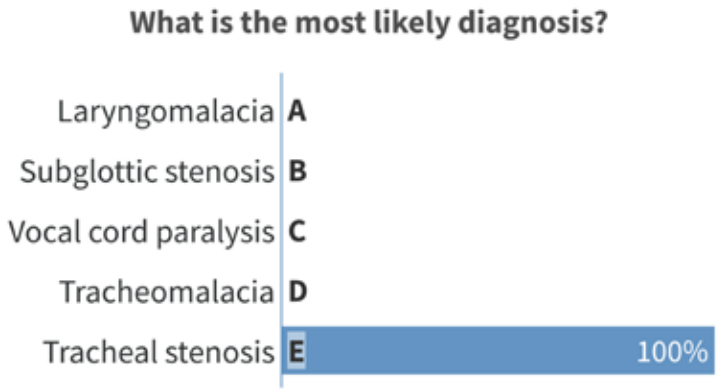
Q. A 6-month-old male infant is brought to you for hospital discharge follow up after he was admitted for croup. Mother is concerned because this is his third upper respiratory tract infection since birth, and he has been making choking sounds while feeding. Further history reveals he has had noisy breathing since he was born that worsens when he is awake and active. On physical exam, you hear expiratory stridor. What is the most likely diagnosis?





A:
Tracheomalacia is defined as the dynamic collapse of the trachea and is classified into congenital (primary) or acquired (secondary tracheomalacia). Congenital tracheomalacia is caused by an intrinsic defect in the cartilaginous portion of the trachea resulting in an increased proportion of the membranous trachea. Acquired tracheomalacia can be caused by prolonged barotrauma from positive pressure ventilation, infection, inflammation, and surgical repair of esophageal atresia and tracheoesophageal fistula. Patients commonly present with expiratory stridor, frequent and prolonged respiratory infections, feeding intolerance, and, if severe, can have cyanotic and apneic spells. The diagnosis is confirmed with flexible bronchoscopy. Treatments vary based on the severity of the tracheomalacia. If mild, close monitoring until patients outgrow symptoms around 6-12 months is sufficient. If severe, surgical intervention may be necessary.

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Q.
A patient presents with the same symptoms and signs as the patient in the previous question; however, now he has other known congenital defects, namely hemivertebrae and corrected H-type tracheoesophageal fistula. What is the most likely diagnosis this time?

A.
Tracheal stenosis is defined as the narrowing of the trachea, either complete stenosis or segmental stenosis. It can occur anywhere along the trachea and vary in length involvement. Its presentation is very similar to tracheomalacia with expiratory stridor, frequent infections, and prolonged respiratory infections. In severe cases, it may present with apnea and cyanosis. However, tracheal stenosis, unlike tracheomalacia, is more commonly associated with respiratory, esophageal, and skeletal anomalies. The diagnosis is confirmed by direct visualization of the trachea with bronchoscopy. Treatments include close monitoring until patients outgrow if symptoms are mild and surgical intervention with balloon dilation, stent placement, or resection and reconstruction if symptoms are severe.

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	Stridor	History	Associated Symptoms	Diagnosis
Laryngomalacia	Inspiratory	Worsens with cry, feeding, supine position		Awake flexible laryngoscopy
Vocal cord paralysis	Inspiratory	Birth trauma, intubation, CT surgery	Recurrent aspiration pneumonia, absent or weak cry	Awake fiberoptic nasopharyngoscopy
Subglottic stenosis	Biphasic	Intubation, Down syndrome	Recurrent croup	Airway fluoroscopy, Laryngoscopy, Bronchoscopy
Tracheomalacia	Expiratory	Extrinsic tracheal compression, prolonged positive ventilation, surgical repair of TEF	Frequent and prolonged respiratory infections	Dynamic CT, Airway fluoroscopy, Flexible bronchoscopy
Tracheal stenosis	Expiratory	Respiratory, esophageal, skeletal anomalies	Recurrent or prolonged croup, recurrent pneumonia	Bronchoscopy

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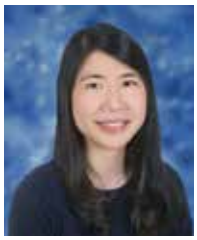

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