VIDEO CORNER

Stridor After Tracheoesophageal Fistula Repair: Where is the Lesion?

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A male infant with a birthweight of 2,500 g (between the 3rd and 10th percentile) is born at 37 weeks' gestation to a 27-year-old primigravida woman with severe polyhydramnios (amniotic fluid index of 37 cm). After delivery, the neonate is vigorous, with an Apgar score of 8 and 9 at 1 and 5 minutes of age, respectively. He develops excessive frothy oral secretions, and the neonatology team is unable to pass a nasogastric tube. Chest radiography shows the coiling of a nasogastric tube with air in the stomach suggestive of esophageal atresia (EA) with tracheoesophageal fistula (TEF).

On day 4 of age, a primary surgical repair of the TEF with eso-esophageal anastomosis is performed under mild tension. Initially, the neonate receives minimal ventilator settings (mean airway pressure 8 cm H_2O, positive end-expiratory pressure of 5 cm H_2O, and fraction of inspired oxygen [FiO_2] 0.21) for 4 days. Three days after surgery, the neonate undergoes extubation to noninvasive positive pressure ventilation and is then weaned to high-flow nasal cannula on postoperative day 4. Soon after extubation, the infant develops intermittent stridor but is weaned to high-flow nasal cannula by postoperative day 4. The stridor progresses over the next 48 hours, and a video of the infant at that time is shown (Video 1).

QUESTION 1

The stridor in this infant occurs in which of the following phases of respiration?

A. Inspiratory phase
B. Expiratory phase
C. Biphasic

The infant's stridor was present at rest but worsened with crying. There was no significant decrease in stridor with neck extension and prone position. An intravenous dexamethasone trial was given after extubation for 3 days with no clinical improvement. The neonate's baseline oxygen saturation in room air was between 90% and 92%; however, with high-flow nasal cannula (flow of 5 L, FiO_2 0.21), the oxygen saturations increased to 94% to 98%. To determine the

AUTHOR DISCLOSURES Drs Ankur, Prasad, Parakh, Chetry, Jain, and Prasad have disclosed no financial relationships relevant to this article. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

Video 1. Infant at 8 days of age.

cause of the stridor, flexible bronchoscopy was done transnasally using a 2.8-mm video bronchoscope (Video 2); this procedure was performed with minimal sedation to ensure spontaneous respirations.

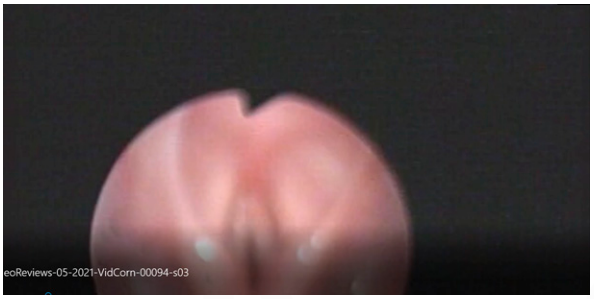
QUESTION 2

Based on this infant's clinical course and findings in the flexible bronchoscopy, the most likely diagnosis in this infant is:

A. Double aortic arch
B. Laryngomalacia
C. Subglottic edema or stenosis
D. Tracheomalacia
E. Vocal cord palsy

DISCUSSION

In infants with stridor, a careful clinical assessment of the stridor can provide information about the type, severity, and level of obstruction. In Video 1, the infant has inspiratory stridor during agitation that is associated with suprasternal and sternal retractions and oxygen desaturations. In addition, the infant's hoarse cry suggests vocal cord swelling or injury. Of note, bronchoscopy (Video 2) reveals that the infant has reduced and paradoxical vocal cord movements (Figs 1 and 2), suggestive of bilateral vocal



Video 2. Flexible bronchoscopy.

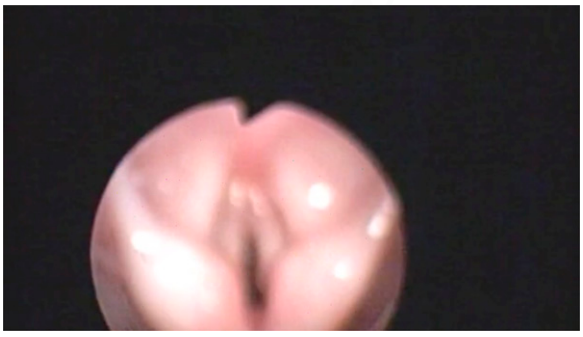


Figure 1. Vocal cords during expiration. Note the limited abduction and a small glottis opening.

cord paralysis (VCP). The video also demonstrates the presence of a repaired TEF in the posterior wall of the midtrachea (Figs 3 and 4) with nonpulsatile narrowing of the trachea. There was no variability with respirations, thus ruling out tracheomalacia. The lower trachea appeared normal. The infant's improvement in oxygen saturations with high-flow cannula was likely because of the airway stents. The infant was discharged with orogastric feedings at 3 weeks; he did not have any stridor and was feeding well on follow-up at 3 months of age.

VCP is the absence of movements of the vocal folds following dysfunction of the recurrent laryngeal nerve. This nerve is a branch of the vagus nerve that runs along the lateral surfaces of the trachea near the tracheoesophageal groove and is vulnerable to injury during surgeries of the neck and thorax. (1) Similar to the patient in this case, bilateral VCP presents with inspiratory stridor at rest that worsens upon agitation. VCP has been reported to occur in 3% to 11.2% of patients following EA/TEF repair. (1)(2) According to a retrospective review, 48% of the postoperatively diagnosed cases were bilateral, and the etiology was mostly (43%) iatrogenic. (3)(4)

A double aortic arch is a rare cause of stridor in infants and is known to be associated with EA/TEF. (5) The mean

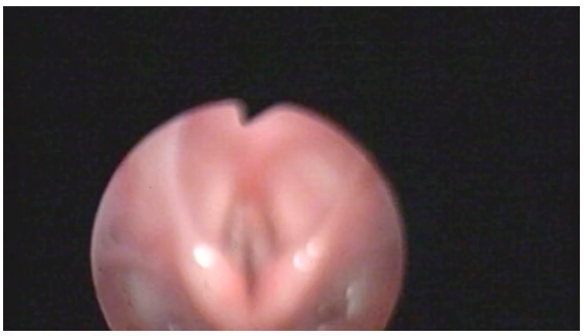


Figure 2. Vocal cords during inspiration. Note the position of cords during inspiration is in adduction, suggesting a paradoxical cord movement.

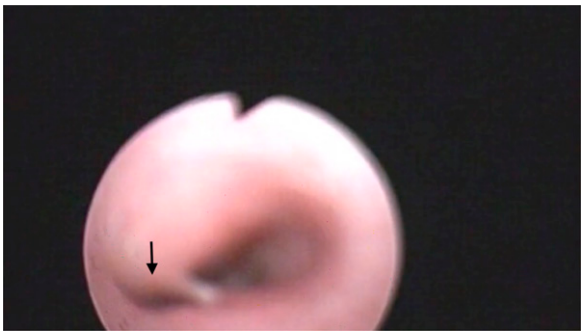


Figure 3. Midtracheal image showing the tracheal end of the repaired fistula (arrow).

age at diagnosis for this vascular ring is 6 months, but it can present earlier. The clinical features are related to compression of the esophagus and trachea, such as dysphagia or difficulty in feeding and stridor in the expiratory phase. Computed tomography angiography is considered the gold standard for diagnosis. In children with a double aortic arch, flexible bronchoscopy may show pulsatile compression in the midtrachea.

Laryngomalacia typically presents with inspiratory stridor and is the most common cause of congenital stridor in neonates. (6) Affected patients typically have inspiratory stridor that increases with feeding, crying, agitation, supine positioning, and upper respiratory tract infections. In patients with laryngomalacia, the inspiratory stridor decreases with neck extension and in the prone position. In this case, the infant had stridor mainly when crying, with no significant decrease in stridor with neck extension and prone position, making laryngomalacia less likely. (7) In a retrospective study, laryngeal anomalies were associated with approximately 30% of cases of TEF, and of these, 4.3% were found to be laryngomalacia. (8) Movements of laryngeal structures should be analyzed in a spontaneously breathing infant to diagnose laryngomalacia. During bronchoscopy, an omega-shaped epiglottis with inspiratory supraglottic tissue collapse is the hallmark of laryngomalacia. (6) However, in our case, flexible bronchoscopy did not demonstrate these findings, thus ruling out laryngomalacia.

Subglottic stenosis can be congenital or acquired. Acquired subglottic stenosis has been recognized as a postoperative complication in 12.9% of TEF patients. (8) It has also been seen as a result of subglottic injury caused by endotracheal intubation, especially if prolonged. The presence of stridor during both phases of respiration remains the most common symptom of a subglottic lesion, which was not noted in our patient. Flexible bronchoscopy ruled out the possibility of stenosis or edema.

The incidence of congenital tracheomalacia has been reported to be at least 1 in 2,100 and has been found to be associated with EA. (4) Indeed, a retrospective study found that tracheomalacia is the most common airway finding in 37.4% of patients with TEF. (8) Localized secondary tracheomalacia may occur due to compression from a vascular malformation (such as a double aortic arch). (5) Acquired forms of tracheomalacia may develop as a result of prolonged ventilation, tracheostomy, or severe tracheobronchitis. Intrathoracic tracheomalacia causes excessive trachea collapse during expiration, whereas extrathoracic tracheomalacia causes tracheal collapse during the inspiratory phase. This leads to the presence of monophonic (expiratory) wheezing in patients with intrathoracic tracheomalacia and inspiratory or biphasic stridor in patients with extrathoracic tracheomalacia. In addition to these symptoms, a 'barking cough' and increased work of breathing is often present. In a patient with tracheomalacia, bronchoscopy typically shows dynamic tracheal compression with respiration, which was not found in our patient. Preoperative bronchoscopy is not a routine practice; instead, it is need-based.

Correct Responses:

Question 1: A. Inspiratory phase

Question 2: E. Vocal cord palsy

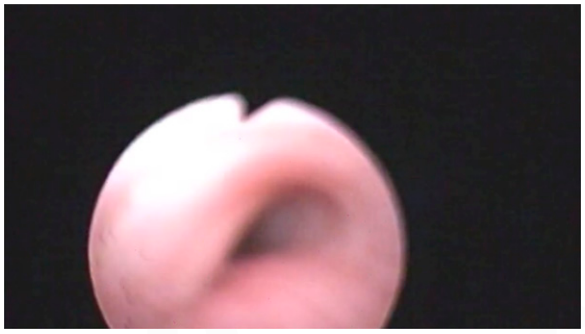


Figure 4. Significant midtracheal narrowing.

American Board of Pediatrics Neonatal-Perinatal Content Specifications

- Know the clinical features of an infant with airway obstruction.
- Plan appropriate diagnostic evaluation and management for an infant with airway obstruction.

Acknowledgment

We wish to thank Dr Ramji Bhardwaj and Dr Manazir Hasan Rahmani for producing and editing the video.

References

1. Fung SW, Lapidus-Krol E, Chiang M, et al. Vocal cord dysfunction following esophageal atresia and tracheoesophageal fistula (EA/TEF) repair. *J Pediatr Surg*. 2019;54(8):1551–1556
2. Mortellaro VE, Pettiford JN, St Peter SD, Fraser JD, Ho B, Wei J. Incidence, diagnosis, and outcomes of vocal fold immobility after esophageal atresia (EA) and/or tracheoesophageal fistula (TEF) repair. *Eur J Pediatr Surg*. 2011;21(6):386–388
3. Kovesi T, Porcaro F, Petreschi F, Trozzi M, Bottero S, Cutrera R. Vocal cord paralysis appears to be an acquired lesion in children with repaired esophageal atresia/tracheoesophageal fistula. *Int J Pediatr Otorhinolaryngol*. 2018;112:45–47
4. Pfleger A, Eber E. Assessment and causes of stridor. *Paediatr Respir Rev*. 2016;18:64–72
5. Hartenberg MA, Salzberg AM, Krummel TM, Bush JJ. Double aortic arch associated with esophageal atresia and tracheoesophageal fistula. *J Pediatr Surg*. 1989;24(5):488–490
6. Landry AM, Thompson DM. Laryngomalacia: disease presentation, spectrum, and management. *Int J Pediatr*. 2012;2012: 753526
7. Bluher AE, Darrow DH. Stridor in the Newborn. *Pediatr Clin North Am*. 2019;66(2):475–488
8. Hseu A, Recko T, Jennings R, Nuss R. Upper airway anomalies in congenital tracheoesophageal fistula and esophageal atresia patients. *Ann Otol Rhinol Laryngol*. 2015;124(10):808–813

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