Uncommon Comorbid Congenital Airway and Pulmonary Anomalies

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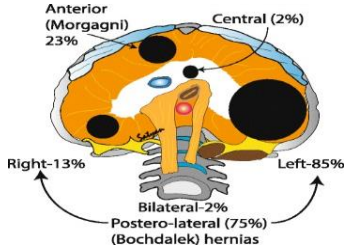
Congenital Diaphragmatic Hernia

Defect in diaphragm allowing abdominal organs to occupy space in the chest cavity, disrupting proper lung development

- Classified based on location (1)



- Most commonly diagnosed by prenatal ultrasonography with an incidence of <5:10,000 births annually
- Often associated with cardiac, renal, or bowel congenital anomalies, which can increase motality to 50% (2)
- Surgical repair is often delayed until cardio-respiratory functions are stable as pulmonary hypertension and hypoplasia may require immediate endotracheal intubation, mechanical ventilation, and extracorporeal membrane oxygenation

1. Chandrasekharan, Praveen Kumar et al. "Congenital Diaphragmatic Hernia – a Review." *Maternal Health, Neonatology and Perinatology* 3 (2017): 6.
2. McHoney M. Congenital diaphragmatic hernia, management in the newborn. Pediatr Surg Int. 2015; 31:1005-13.
3. Dwarakanath S, et al. Unexpected presentation of a type IV laryngotrachoesophageal cleft: anesthetic implications of a rare case. Indian J Anaesth 2014;58:746-8.

Case Discussion

1.8 kg female with known left sided congenital diaphragmatic hernia at 32 6/7 weeks gestation was apneic and bradycardic upon delivery. Immediate intubation and ventilation attempts were unsuccessful. Bag-mask ventilation improved oxyhemoglobin saturations to 90% and heart rate to >100 bpm. Patient was transferred to the operating room for direct laryngoscopy and rigid bronchoscopy. A Miller 0 laryngoscope used by otolaryngology revealed widely splayed arytenoids and complete opening of the larynx posteriorly into the esophagus. A 3.0 mm rigid bronchoscope advanced into the trachea confirmed contiguous communication between trachea and esophagus distal to the carina. While chest compressions were initiated for bradycardia, the scope was exchanged for a 2.5 mm rigid bronchoscope and advanced into the right bronchus. An attempt to ventilate the right lung returned minimal CO2. Given the extent of multiple severe disease processes in a premature neonate, the decision was made to stop resuscitation. The patient was pronounced dead approximately three hours after delivery. Autopsy revealed a type IV laryngotracheoesophageal cleft, left-sided congenital diaphragmatic hernia and severe pulmonary hypoplasia.

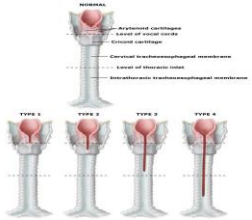
Unique Solutions for Unique Challenges

- Early diagnosis, establishing ventilation, and limiting aspiration are initial goals
  - Low grade clefts can be managed conservatively: head up, thickened feeds, antireflux Rx
  - High grade clefts require early repair to limit pulmonary damage from repeated aspiration
- Methods developed to aide in securing an airway when faced with high grade clefts (5,6)
  - Bifurcated endotracheal tubes; using two endotracheal tubes extended into each bronchus
  - Placing a large cuffed endotracheal tube to minimize leak, allow suctioning, and better control of oral secretions
  - Sealing gastroesophageal junction with Foley catheter balloon secured under traction, improves pulmonary ventilation and inhibits gastric inflation upon common tracheo-esophageal lumen ventilated
  - Proximal gastric division with placement of gastrostomy tube
  - Use of extracorporeal membrane oxygenation perioperatively

Laryngeotracheoesophageal Cleft

Defect of the posterior larynx and trachea resulting from failed fusion of the cricoid lamina leading to communication between the larynx and pharynx (3)

- Classified based on extension (4)



- Annual incidence of 1/10,000 to 1/20,000 live births
- Presenting symptoms depend on length of cleft:
  - Types I-II present with cough, stridor, hoarse cry, and feeding difficulties
  - Types III-IV present with frequent/severe aspirations, infections, and respiratory failure (Fernandez)
- Challenges establishing/maintaining an airway, protecting from aspiration
  - Existence/extent unknown at birth
  - Endotracheal tube can be dislodged into esophagus, limiting ventilation
  - May require one lung ventilation
  - Increased aspiration risk

4. Benjamin, B, Inglis, A. Minor congenital laryngeal clefts: diagnosis and classification. Ann Otol Rhinol Laryngol 1989; 98:417.
5. Ryan A, et al. Congenital diaphragmatic hernia: associated malformations – extralobular sequestration and laryngotracheoesophageal cleft: two case reports.
6. Arai, L.R., et al, (2007), Anesthesia management of a patient with laryngotracheo-esophageal cleft. Pediatric Anesthesia, 17: 171–175.