Tracheal Foreign Body Aspiration in a Pediatric Patient with Cystic Fibrosis – This Should be a Quick Case, Right?
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Goals:
1. Discuss the anesthetic implications of a patient with cystic fibrosis and cerebral palsy.

2. Compare and contrast various approaches in the anesthetic management of tracheal foreign body in a pediatric patient.

3. Describe the considerations for airway management and cardiopulmonary support in patients with tracheobronchial injury.

4. Discuss the physiologic effects and general considerations in management of one lung ventilation.

5. Describe the options for postoperative analgesia following a thoracotomy.

Description: This is a case of a pediatric patient with medical history of cerebral palsy and cystic fibrosis who presents with a tracheal foreign body. During bronchoscopy and removal of the airway foreign body, the patient develops tracheobronchial perforation and requires open repair via thoracotomy. Anesthetic management of tracheobronchial injuries, including one lung ventilation and extracorporeal membrane oxygenation is discussed.

Stem: A 12 year-old, 39 kg female presents to the emergency department with excessive drooling and stridor. She suffered a hypoxic ischemic insult at birth secondary to vacuum extraction leading to cerebral palsy. She was diagnosed with cystic fibrosis at 19 months of age. Chest x-ray reveals a round, radiopaque tracheal foreign body above the carina. The child was seen playing with her mother’s change purse prior to symptom development. She is presenting to the operating room for rigid bronchoscopy and foreign body removal. The patient uses 2L nasal cannula oxygen while awake and BiPAP 10/5 while asleep.

What are the anesthetic implications of cystic fibrosis?
What are some common co-morbidities associated with cerebral palsy?
Is this an emergency? Are there any other studies or lab results you would request before proceeding?

Upon examination in the holding area, the child’s initial vital signs are SpO2 88% on 2L nasal cannula, heart rate 162, and a respiratory rate of 52. You note substernal retractions and biphasic stridor. The child is visibly agitated and continuously drooling. Lung
auscultation reveals right sided wheezing and bilateral rales. Staff in the emergency
department were unable to place a peripheral IV.

What other questions would you like answered before taking this patient to the OR?
Is pre-medication appropriate for this patient?
What are the options for airway management in a patient with a tracheal foreign
body? In this patient specifically?
What is your plan for anesthetic induction? Is it essential to place a peripheral IV before
proceeding to the OR?
What monitors would you use for this case?
What is your plan for maintenance of anesthesia in this patient?

Once anesthesia has been induced, the ENT surgeon requests the head of the bed be
turned 90 degrees and the patient remain in suspension while a rigid bronchoscope is
placed. Upon insertion of the rigid bronchoscope into the pharynx, the patient violently
coughs and develops laryngospasm; oxygen saturation acutely falls to 65% within 10
seconds.

How will you treat the laryngospasm?
Discuss the role of topical local anesthetic in this situation?
What is the maximum dose of 0.25% bupivacaine that can be used for this patient?

Once the episode of laryngospasm is treated and the airway topicalized with local
anesthetic, the ENT surgeon locates the tracheal foreign body, a penny, immediately
above the carina. During airway manipulation, the patient becomes impossible to
ventilate. SpO2 falls to 12% and HR decreases to 42.

What is your differential diagnosis for impossible ventilation?
Describe appropriate next steps.
Discuss available techniques to maintain oxygenation in a patient undergoing rigid
bronchoscopy after muscle relaxant has been given?

The child is appropriately resuscitated and the penny is removed with a rigid
bronchoscope. The ENT surgeon intubates the child due to excessive airway secretions
and SpO2 lingering at 85%. A PICU bed is secured.

Should the child be extubated or remain intubated following this procedure?
Discuss postoperative ventilation strategies in this patient?

The child is transported intubated, ventilated and sedated to the PICU. On post-operative
day 1 the child is noted to have crepitus, mediastinal emphysema as well as dyspnea.

What is the differential diagnosis for the patient’s presentation?

BP is noted to be decreasing to 60/40 and HR has increased to 172. Blood is noted to be
collecting in the endotracheal tube.
What is the diagnosis and management at this time?

After the patient is stabilized, a bedside flexible bronchoscopy and Chest CT confirm tracheobronchial perforation. This likely occurred when the patient coughed with the rigid bronchoscope in place. The endotracheal tube is advanced past the tracheal injury to minimize the air leak. Although placed on broad spectrum antibiotics, the patient develops mediastinitis and a surgical repair of tracheobronchial perforation is scheduled.

What are the anesthetic considerations for surgical repair of tracheal injury?
Is veno-venous cannulation for ECMO appropriate? What monitors are appropriate? Describe the challenges associated with one lung ventilation in a patient with cystic fibrosis?

The thoracic surgeon requests one lung ventilation for tracheal perforation repair via right sided thoracotomy. After IV induction the patient is intubated with a #32F double lumen tube (DLT). Positioning of DLT was confirmed via auscultation and use of fiberoptic bronchoscope. Patient was then positioned in the left lateral decubitus position. DLT placement was reconfirmed once in correct position for surgery. Vital signs at this time were SpO2 100%, BP 90/50, HR 60, RR 10 (on controlled ventilation), ETCO2 33, normal sinus rhythm. One lung ventilation was initiated, isolating the right lung. There was immediate desaturation and acute cardiopulmonary decompensation. VSS at this time were SpO2 79%, BP 82/55, HR 90 with manual ventilation ETCO2 30.

What is the differential diagnosis for desaturation in this patient?
What steps can be taken to improve oxygenation?
Describe respiratory mechanics in the lateral decubitus position.
Discuss ventilation strategies in this patient for optimal hemodynamics.
What are your next steps?

The patient is stabilized and vital signs return to baseline. The tracheal defect is repaired via an autologous pericardial graft. The case proceeds uneventfully until the surgeon begins to close the surgical wound. You notice a steady rise in end tidal carbon dioxide. Delivered tidal volume falls precipitously and the patients desaturates to 85%. An arterial blood gas reveals pH 7.19, pCO2 79, pO2 110, O2 sat 99, BE -7, HCO3 20.

What is your differential diagnosis? What are your next steps?

Upon manual ventilation with 100% FiO2, there is improvement in oxygen saturation to 92%, but you note a decrease in lung compliance. Peak airway pressures increase to 32. Upon suctioning of the endotracheal tube, a 10 French catheter is unable to be passed. Oxygen saturation declines to 72%.

Discuss management at this time.
The patient is turned supine and re-intubated with a new single lumen endotracheal tube. Multiple inspissated mucous plugs are noted in the endotracheal tube. After restoration of oxygenation and ventilation, the thoracotomy incision is closed.

**Is extubation of this child with cystic fibrosis appropriate?**

**What are your recommendations for postoperative analgesia?**

**What are the advantages to a paravertebral nerve block catheter vs. a thoracic epidural?**

**Is IV PCA appropriate?**

**Discussion:**

Cystic Fibrosis (CF) is an autosomal recessive condition that causes chronic suppurative lung disease as well as a host of multisystemic complications. Although it is the most commonly fatal inherited disease in Caucasians, median survival has improved in patients due to refinements in nutrition, antibiotic regimens, and airway clearance techniques with regular chest physiotherapy (1). CF is caused by mutations in a single gene on the long arm of chromosome 7 that encodes an amino acid protein known as CF transmembrane regulator (CFTR)- a chloride channel found in the epithelial cells lining most exocrine glands in the body (2). Progressive lung disease leads to the early mortality commonly associated with CF. Decreased mucociliary clearance leads to patchy atelectasis, airway inflammation and chronic hypoxia, while bronchiectasis and air trapping lead to airway obstruction. Some patients develop a steady decline in lung function leading to cor pulmonale, respiratory failure and eventual death (3).

The multisystemic effects of CF include malabsorption of vitamins A, D, E, and K due to obstruction of pancreatic ducts. CF-related diabetes mellitus (CFRD) occurs due to pancreatic fibrosis and gland destruction. Patients with CFRD have worse lung function, worse nutrition status, more frequent hospitalization and higher mortality than patients without diabetes (4). Significant liver disease is seen in 13-25% of children with CF. Meconium ileus is a common presentation of the disease in the neonatal period. The diagnostic sweat test is due to abnormal sodium and chloride levels in all exocrine secretions. 98% of men with CF suffer from congenital bilateral absence of the vas deferens leading to primary male infertility (2).

Respiratory function is maintained with appropriate physical therapy, including at least twice daily physiotherapy, inhaled bronchodilators and mucolytics. Physical therapy includes postural drainage, percussion, and autogenic drainage. Oscillatory devices, positive expiratory pressure devices, and high frequency chest compression devices are all useful for mucous dislodgement and clearance. Lung and liver transplantation is increasingly common in children with this disease. Bronchiolitis obliterans is the most important complication after pediatric lung transplantation and limits both the quality of life and duration of survival (2).

Anesthetic management of CF patients relies on preoperative risk classification and optimization. Patients should be optimized for elective surgical procedures with intense daily physiotherapy and targeted medication regimens. Echocardiography may be useful
in diagnosing cor pulmonale. Emergency surgery is significantly more challenging, but multidisciplinary management is paramount along with chest radiography, baseline arterial blood gas analysis, and bedside spirometry. The risk of respiratory complications is significantly increased by prolonged duration of anesthesia, upper abdominal or thoracic incisions, nasogastric tube insertion or emergency surgery (2). A preoperative evaluation will include a thorough medical history, the patient’s current therapeutic interventions, medications, and recent exacerbations. The need for patient isolation is determined by microbial infection, the volume and purulence of sputum. Tolerance of physiologic stress by the patient can be extrapolated by the patient’s functional ability. Glycemic control is important in these patients, especially those with CFRD (2,3).

Intraoperative monitoring for CF patients includes arterial line placement for frequent blood gas analysis. In the case of central venous catheter use, ultrasound guidance should be utilized to minimize the risk of pneumothorax. Consideration for a pulmonary artery catheter or cardiac output monitoring should be utilized in patients with cor pulmonale or those undergoing major surgery. Although the use of a laryngeal mask airway in a spontaneously breathing patient may prevent respiratory complications, placement of an endotracheal tube facilitates tracheal suctioning of secretions and allows for controlled ventilation. Nasal intubation should be avoided in CF patients due to nasal polyposis. Airway pressures should be kept low when utilizing positive pressure ventilation techniques, and gases should be humidified (3). Short acting anesthetic agents should be utilized to facilitate rapid emergence in CF patients. These cachectic, malnourished patients often require careful positioning and padding to avoid nerve injury. Regional anesthesia can be utilized to avoid airway manipulation and facilitate analgesia for early mobilization. Chest physiotherapy should be resumed as early as possible (5).

Cerebral palsy refers to a spectrum of neurologic disorders resulting from injury to the neonatal or infant brain from a variety of causes. Anesthetic management of these patients can be challenging due to comorbidities including gastroesophageal reflux, epilepsy, and altered respiratory function and mechanics. Patients often exhibit recurrent aspiration, respiratory distress syndrome, and musculoskeletal complications leading to restrictive lung disease and pulmonary hypertension. Polypharmacy from anti-spasticity medications as well as anticonvulsants must be taken into consideration when selecting anesthetic agents. Contractures of the extremities may make patient positioning difficult. Approximately 60% of patients will have some degree of impaired cognitive function and delayed intellectual development (6). There is no recommended anesthetic technique for the management of patients with cerebral palsy and care must be tailored to the individual patient and their comorbidities. Use of succinylcholine is not contraindicated as studies have not shown an increased release of potassium after its administration despite the potential for extrajunctional acetylcholine receptors. Because acetylcholine receptors are upregulated at the neuromuscular junction in cerebral palsy, patients may exhibit relative resistance to nondepolarizing neuromuscular blockade, particularly if they are concurrently on anticonvulsant therapy (7).

Many options exist for airway management during removal of an aspirated foreign body, and no technique has been shown to be superior to the others. Decisions must be made
regarding induction of anesthesia, mode of ventilation during rigid bronchoscopy, and maintenance of anesthesia. Communication with the surgical team is key as the airway will be shared. Choice of induction technique must take into consideration the location of the foreign body and its’ potential movement during induction. Of great concern is converting a partial airway obstruction to a complete obstruction during induction. Use of both inhalational and IV induction techniques have been reported. Despite the technique selected, patients should have intravenous access established prior to induction of anesthesia due to the potential for rapid decompensation. NPO status should be assessed but an urgent case should not be delayed in the event of a full stomach. In a meta-analysis of rigid bronchoscopy for foreign body removal, no cases of aspiration of stomach contents were reported (8). Although no strict recommendations exist regarding choice of induction technique, maintenance of spontaneous ventilation is commonly practiced amongst pediatric anesthesiologists.

Once the patient has been induced and the bronchoscope introduced, the decision must be made regarding maintenance of spontaneous ventilation versus controlled positive pressure ventilation. Oxygen can be provided by connecting to the side port of the rigid bronchoscope, use of a nasal cannula, or a combination of the two. Providing positive pressure ventilation, particularly for proximal foreign bodies, may prove challenging because of a large leak around the bronchoscope. Closing the patient’s mouth and nose may improve this situation if mechanical ventilation is desired. A third option is the use of low frequency jet ventilation. This technique may provide better ventilation to both lungs since it is a separate apparatus not related to the position of the bronchoscope. High frequency jet ventilation (HFJV) - (60–600 cycles/min) is provided by a specialized ventilator, and it diminishes airway pressures and the risk of barotrauma (9).

A well conducted intravenous technique can allow for maintenance of spontaneous ventilation while still providing a consistent and adequate depth of anesthesia. Commonly used agents include propofol, remifentanil, and dexmedetomidine. The patient’s airway should be well topicalized with local anesthetic by the proceduralist. This is helpful in desensitizing the airway and reducing the risk of patient movement and laryngospasm during the intense stimulation of bronchoscope placement and manipulation (8). The maximum dose of local anesthetic based on the patient’s weight should be calculated prior to beginning the case. Any movement of the patient during bronchoscopy may result in serious injury, but the use of a neuromuscular blocker may be dangerous in patients with partial airway obstruction. During rigid bronchoscopy, the bronchoscopist is exposed to anesthetic gases, and higher PaCO2 values are often observed with positive pressure ventilation. TIVA allows for adequate anesthesia without interruption of ventilation and prevents contamination of the operating room with anesthetic gases. Complications of rigid bronchoscopy include soft-tissue injury (mouth, lips, tongue), tooth damage, sore throat, airway perforation, hemorrhage, airway edema, and laryngospasm (10).
If the foreign body moves during induction or is dropped during the attempted extraction, a partial airway obstruction may become a complete airway obstruction with the patient becoming impossible to ventilate. Other potential causes for an inability to ventilate that must be considered are bronchospasm, laryngospasm, or obstruction caused by tissue edema and inflammation. If the object is unable to be removed immediately, it can be pushed distally to allow ventilation of unobstructed lung segments (8). If this is not possible and complete airway obstruction continues, initiation of ECMO may be the only option for oxygenation.

Pneumothorax is a known serious complication in CF patients. In a retrospective cohort study, Flume et al. (11) recognized a number of risk factors associated with an increased occurrence of pneumothorax, including the presence of P. aeruginosa, Burkholderia cepacia or Aspergillus in sputum cultures, FEV1 <30% of predicted, enteral feeding, pancreatic insufficiency, allergic bronchopulmonary aspergillosis (ABPA) and massive hemoptysis. Pneumothorax was more prevalent in patients with more severe pulmonary impairment and was responsible for increased 2-year mortality. There is poor correlation between the presence of blebs or cysts and pneumothorax in CF patients (12). Endobronchial obstruction due to accumulation of viscous secretions and macrophages can cause air trapping in alveoli, resulting in rupture of pulmonary parenchyma (13).

Management of pneumothorax in CF patients entails safe and effective resolution as well as prevention of a recurrence. Therapeutic options with various sclerosing agents have been employed, including quinacrine, silver nitrate, iodine and talc. Pleural abrasion, intercostal drainage, pleurectomy and the Heimlich flutter valve have also been utilized (14). According to the British Thoracic Society (BTS) guidelines for the management of Pneumothorax [2003], the care in patients with CF is similar to that for non-CF patients, including early, aggressive treatment and surgical intervention after the first episode (15). BTS guidelines state that a small pneumothorax without symptoms can be observed or aspirated, while large pneumothoraces require treatment with intercostal tube drainage. Lung transplantation is considered the definitive solution for refractory and/or recurrent

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pneumothorax. High-frequency chest compression, active cycle breathing and autogenic drainage can be utilized immediately after a pneumothorax for chest physiotherapy. Physiotherapy should focus on maintaining effective sputum clearance to prevent atelectasis and lobar collapse due to obstructive secretions. Effective analgesia should be administered to relieve aggravation of pain caused by chest tube placement in order to avoid bronchiolar consolidation, fibrosis and air-trapping (16). Persistent pneumothorax, associated with excessive air leakage, should increase the suspicion of intrathoracic tracheal or bronchial injury.

Tracheobronchial injuries are rare, with a reported incidence of 0.5-2%, but may be fatal. The most common tracheal injury is a tear near the carina, likely due to the role of shear forces on the airway near the relatively fixed carina. Patients present with varied symptoms including dyspnea, respiratory failure, hoarseness, dysphonia, subcutaneous emphysema, pneumothorax, and hemoptysis. Chest radiography, neck and chest CT are required for diagnosis. A fiberoptic bronchoscope (FOB) is the best method to assess the size and location of injury. These patients often require immediate intubation because of respiratory failure and hemodynamic instability (17). Surgical repair of a tracheal injury presents a litany of challenges for the anesthesiologist. Of primary concern is the mechanism of providing adequate ventilation and oxygenation to the patient while allowing for optimization of the surgical field. Other challenges include positioning difficulties if the lateral decubitus position is utilized as well as provision of a consistent anesthetic level if the use of inhaled anesthetics is desired.

In a patient with tracheal disruption, it is imperative that the endotracheal tube (ETT) be placed distal to the disruption to avoid the risk of rapid pneumothorax and death. A double lumen intubation is preferred to protect both lungs from bleeding distal to the injury. For patients with a standard ETT, providers must be vigilant for signs of hypoxia, subcutaneous emphysema of the neck and chest, and discordance between inspiratory and expiratory pressures. These signs may indicate that the ETT may have moved proximal to the injury or that the wound has extended distal to the end of the ETT. Early diagnosis and surgical repair is critical for the preservation of lung function (17).

For upper tracheal lesions, the trachea is explored through a U-shaped cervical incision, between the lower part of the cricoid cartilage and the sternal notch. For lower tracheal lesions, the surgical approach is via median sternotomy or right thoracotomy (18). In the presence of an upper tracheal lesion, the ETT is placed in the trachea above the lesion. The area of tracheal rupture or injury is exposed, and the ETT is withdrawn into the proximal trachea. A sterile ETT is placed into the distal trachea by the surgeon (19). This tube is connected to a sterile anesthetic circuit across the surgical field. Ventilation is maintained by the anesthesiologist and confirmed by evaluating chest inflation, peripheral oxygen saturation, ETCO2, and blood gas analysis (20).

Anesthetic management for distal tracheal or carinal injuries is via surgical approach through right posterolateral thoracotomy. Ventilation strategies during surgery involve one-lung ventilation (OLV), extracorporeal membrane oxygenation (ECMO), or cardiopulmonary bypass (CPB). Different approaches are used for OLV. After the ETT is
pulled above the injury during the procedure, an endobronchial blocker, double lumen
tube (DLT) or single lumen endobronchial tube may be placed into the left mainstem
bronchus (21). If feasible, a DLT is preferred because it allows physiologic separation of
the lungs if an air leak becomes severe enough to require varied ventilator modes or
settings for each lung. According to the airway device used, low-frequency jet ventilation
or low tidal volume ventilation are applied during resection. ECMO is another alternative
strategy for gas exchange, which can be performed veno-venously or veno-arterially (22).

Anesthetic maintenance and ventilation strategies including manual jet ventilation, HFJV,
OLV and CPB can be performed according to the localization and size of tracheal injury
(23). The initial approach to ventilator settings is to provide low Vt (6–7 mm Hg/kg) and
allow hypercapnia by maintaining Pco2 below 60 mm Hg and pH at 7.3 or greater (24).
PEEP should be low, ideally 5 mm Hg, to minimize expiratory air leak (25). Goal plateau
pressure (Ppl) should be less than 26 mm Hg for optimal compliance and gas exchange
(26). In patients with refractory respiratory acidosis, persistently elevated peak airway
pressure, or oxygenation difficulties, salvage modes include high-frequency ventilation
(HFV) or 2L-ILV (synchronous or asynchronous) (27, 28). An advantage of HFV is that
ventilation is achieved through an ETT with the cuff deflated which facilitates surgical
repair and healing by removing cuff ulceration as a risk to the wound. More useful for
facilitating surgical repair is a traditional long ETT cannulation of the uninjured lung,
with the inflated cuff anatomically isolating that lung from contamination (24).

During one lung ventilation (OLV), although only one lung is ventilated, both lungs are
perfused. Perfusion of the collapsed, nonventilated lung inevitably leads to
transpulmonary shunting, impaired oxygenation, and hypoxemia. In the lateral decubitus
position, gravity leads to better perfusion of the lower, ventilated lung than of the upper,
nonventilated lung. Because oxygenation increases as perfusion of the nonventilated lung
decreases, oxygenation during OLV will be better with the patient in the lateral decubitus
position as compared to the supine position. Optimizing pulmonary function before a
surgical procedure via physical therapy and drugs to dilate bronchi and loosen secretions
will not only decrease postoperative pulmonary complications but also improve
oxygenation during OLV (29). There are various causes for hypoxemia during OLV. A
distal misplacement of a left-sided DLT during right thoracotomy may lead to hypoxemia
because only the left lower or the left upper lobe will be adequately ventilated through
the bronchial limb. A proximal dislodgment of a left-sided tube may lead to ventilation
difficulties through the tracheal limb because the cuff of the bronchial limb may partially
obstruct the trachea. Ventilating through the bronchial limb of a right-sided tube may lead
to hypoxemia if the right upper lobe is not adequately ventilated, leading to an increase in
shunt fraction (30).

Ventilation strategy is important in decreasing the incidence of hypoxemia during OLV.
Expansion of the dependent lung is impeded by the pressure of the abdominal organs and
cephalad displacement of the diaphragm, and by the pressure and noncompliance of the
thoracic wall. Atelectasis readily occurs in the dependent, ventilated lung, which activates
hypoxic pulmonary vasoconstriction (HPV), increasing both perfusion to the
nonventilated lung and the shunt fraction. For these reasons, it is crucial to avoid the
occurrence of atelectasis in the dependent lung (30). In a study by Wrigge et al (31), large tidal volumes without PEEP, potentially injurious to the lung, do not translate into better oxygenation during OLV as compared with moderate or low tidal volumes combined with PEEP.

There are various strategies to treat hypoxemia during OLV. Increasing FIO2 can be effective immediately. If this strategy does not improve oxygenation, the surgeon should be informed and the operated lung expanded with pure oxygen. Although hand ventilation of the operated lung may be sufficient every 3–5 min, CPAP application to the nondependent lung in the range of 3–10 cm H2O is a more effective way of improving oxygenation. It is often easier for the surgeon to operate on an immobile but expanded lung than on a lung with frequent tidal ventilations (32). To keep the ventilated lung expanded, recruitment maneuvers and increasing PEEP up to 10 cm H2O maybe be needed. High-frequency jet ventilation with small tidal volumes and high respiratory frequency of the nondependent lung is also useful in treating hypoxemia (33). The most common treatable causes of hypoxemia during OLV are dislodgment of the DLT, atelectasis in the ventilated lung, and occlusion of major bronchi of the ventilated lung with secretions or blood. If dislodgment has occurred or if secretions have occluded major bronchi, a FOB can be used to correct the DLT position or to clear secretions. If the DLT is not dislodged and no occlusion is found, the ventilation strategy should be reassessed (30).

In addition to OLV for tracheal surgery, extracorporeal membrane oxygenation (ECMO) has become an invaluable tool in the care of patients with severe cardiac and pulmonary dysfunction refractory to conventional management (34). Indications for ECMO include a bridge to cardiac or lung transplant, and support for tracheal or lung resections in unstable patients (35). If circulatory support is not indicated, veno-venous (VV) ECMO can provide full respiratory support while avoiding in-field ventilation and optimizing surgical conditions. For patients with cardiac comorbidities that require circulatory support, veno-arterial (VA) ECMO is more appropriate. VV ECMO can be accomplished with peripheral cannulation and requires a lower level of systemic heparinization than VA ECMO or cardiopulmonary bypass. The most common complication associated with ECMO is hemorrhage, due to the need for heparinization (36). Other complications include heparin-induced thrombocytopenia, neurological complications, ECMO circuit clots, and resultant oxygenator failure (37). For decades, CPB has been used successfully in operative procedures of the trachea, either to ensure adequate gas exchange during induction of anesthesia in a patient with difficult airway or as a rescue technique in case of insufficient ventilation. ECMO may have several advantages compared to CPB, including less fluid overload due to hemodilution and reduced demands for anticoagulation, leading to lower transfusion requirements (38).

Although general anesthesia is the preferred and most common approach for tracheal resection, few patients have been managed with regional anesthesia. It has been shown to be a proven technique with regard to patient safety and efficacy. Cervical epidural anesthesia and cervical plexus blocks have been performed successfully in patients for carotid surgery; common complications include hypotension (10.9%), bradycardia
(2.8%), and respiratory muscle paralysis (0.8%) (39). Thoracic epidural anesthesia and local anesthetic block may also provide significant benefit, but their role and safety in tracheal surgery requires further research.

At the end of tracheal resection and reconstruction, the ETT is placed above the anastomosis line, and ventilation is conventionally maintained. Extubation should be planned in the operating room if the patient meets criteria and has adequate respiratory effort. Prolonged intubation with positive pressure ventilation can disrupt the anastomosis line due to tracheal irritation (18). In patients who have undergone carinal resection with prolonged operative time, postoperative mechanical ventilation may be required for respiratory support. The goals of postoperative ventilator care are to reduce strain on the operative site and prevent complications such as pneumonia and atelectasis, especially in the CF patient. Aggressive pulmonary toilet is mandatory to reduce the risk of postobstructive pneumonia. Patients with tracheal disruption are at high risk for postoperative pneumonia and should receive prophylactic broad spectrum antibiotics until they are extubated and clearing their own secretions (40). Before extubation, neck flexion is supported with a suture between the chin and the manubrium sterni to decrease anastomotic tension (41). Following extubation, preparation should be made for possible emergent airway rescue and re-intubation. Early postoperative complications involve respiratory distress and vocal cord paralysis due to recurrent laryngeal nerve injury, causing hoarseness or dyspnea. Later complications can be anastomotic, including granulation tissue formation, necrosis, dehiscence or hemoptyis (20).

Adequate control of postoperative pain after thoracic surgery can be a challenge. Strategies for postoperative analgesia include placement of a thoracic epidural, use of paravertebral nerve block techniques, and the use of intravenous opioids and nonopiates such as acetaminophen and nonsteroidal anti-inflammatory drugs (NSAIDs). Placement of a thoracic epidural was previously the gold standard for pain management after thoracotomy; however, this technique is associated with a multitude of side effects and potential risks including hypotension, urinary retention, itching, epidural hematoma formation, and damage to the spinal cord (42). Performance of a paravertebral nerve block with catheter insertion can potentially avoid these risks and side effects while providing the same level of analgesia. However, this technique is associated with inadvertent pleural puncture and formation of a pneumothorax.

Limited pediatric studies comparing use of a thoracic epidural versus paravertebral blockade have been performed but results from studies in the adult population have shown similar analgesia with a reduction in minor side effects with the paravertebral technique. However, the paravertebral technique may be technically more challenging and has a steep learning curve for successful block placement. In general, 1-2 dermatomes will be covered by a paravertebral catheter depending on the spread of local anesthetic. Multiple catheters may need to be placed for adequate post-operative analgesia depending on the size and location of the surgical incision and residual drains that remain in place postoperatively (42). Epidural or paravertebral techniques allow optimization of respiratory function and reduced need for opioid administration.
When neuraxial analgesia is not feasible, an intercostal nerve block coupled with systemic parenteral analgesia is an option. Unfortunately, the block’s limited duration of action, approximately 6 hours, necessitates repeating the block at multiple levels or starting an infusion. This can increase the risk of systemic toxicity from the highly vascular intercostal space. Incomplete analgesia is also a problem since the dorsal rami supplying the back are not blocked, which is relevant in posterolateral thoracotomies, and the lateral cutaneous branch may also be missed if the block is performed too anteriorly (43). Epidural analgesia has been shown to be superior to narcotics via patient-controlled analgesia (PCA) devices. Opioids are primarily used as adjuncts to regional techniques due to significant respiratory depression when used as sole agents. The role of NSAIDs in thoracotomy is two-fold, to reduce opioid requirements and to treat ipsilateral shoulder pain resistant to thoracic epidural analgesia. Chronic post thoracotomy pain can be controlled with agents including NSAIDs, amitriptyline, gabapentin, opioids, and ketamine, useful in reducing peripheral and central sensitization. Non-pharmacological treatments used have shown varying success and include transcutaneous electrical nerve stimulation, cryoanalgesia, radiofrequency ablation, and spinal cord stimulation (43).
References:


44. https://medicine.uiowa.edu/iowaprotocols/maximum-recommended-doses-and-duration-local-anesthetics
A Heartbreaking Event: Catastrophic Complication of a Nuss Procedure

Moderators: Stephanie Grant, MD – Assistant Professor of Anesthesiology and Pediatrics
Julie Williamson, DO – Associate Professor of Anesthesiology and Pediatrics

Institution: Emory University and Children’s Healthcare of Atlanta at Egleston

Goals:
At the conclusion of this discussion, participants will be able to:
1. Identify the surgical and anesthetic risks of the Nuss procedure to create a perioperative management plan that anticipates rare but catastrophic complications.
2. Develop an intraoperative plan for resuscitation from surgical hemorrhage requiring massive transfusion.
3. Manage debriefing after an unexpected adverse event in the OR including conflict negotiation during a vulnerable time.

Case history – Preoperative Clinic:
A 14 year old boy with pectus excavatum presents for preoperative evaluation for a Nuss procedure. He complains of constant low grade chest pain that worsens with exercise. A chest CT scan demonstrates a severe pectus excavatum deformity with a Haller index of 4.8. His vital signs are weight 44 kg, SpO$_2$ 98%, HR 84, and BP 109/68.

Questions:
1. What is the natural history and incidence of pectus excavatum?
2. What are indications for pectus excavatum repair?
3. Compare the Nuss and Ravitch procedures.
4. What is the Haller index?
5. What preoperative diagnostic evaluations will you review or request for this patient?
6. Do you routinely order a type and screen for patients undergoing the Nuss procedure?
7. What are the anesthetic complications associated with pectus excavatum repair?
8. What are the surgical complications associated with pectus excavatum repair?

Case history – Case Progression:
On the day of the procedure, the pre-op vital signs are SpO$_2$ 99%, BP 115/68, HR 95. The patient is given premedication with midazolam.

1. What is your anesthetic plan for this patient?
2. How will you manage his pain intraoperatively? Post-operatively?
3. What monitoring and equipment should you have ready for this patient?
4. Does this patient need invasive monitoring?
5. How will you maintain anesthesia for this patient?
6. What are the hemodynamic goals for this patient?

Case History – Intraoperative Management:
The patient tolerates the induction of anesthesia and intubation. He remains stable with your plan for maintenance of anesthesia. A thoracic epidural is placed for postoperative pain management.

**Case History – Case Progression:**
Thoracoscopic access is obtained in the left chest and the thorax is insufflated for visualization of the left hemithorax. During placement of the Nuss bar across the chest, a burst of clear fluid is seen. The patient becomes acutely hypotensive and the heart rate decreases to 54 bpm. On EKG, T waves become acutely elevated. The SpO2 remains 100%.

**Questions:**
1. What is your differential diagnosis?
2. What is the significance of the T wave elevation and bradycardia in the absence of hypoxemia?
3. How do you respond to the hypotension? What do you do next?

**Case History – Case Progression:**
You call for help and several anesthetists and anesthesiologists report to the OR. The surgeon extends the left thoracic port site to an anterolateral incision. A large amount of bright red blood is present in the chest. BP is now 60/30 with a palpable carotid pulse. An injury to the right ventricle is confirmed.

**Questions:**
1. Do you have adequate vascular access? Do you need additional monitoring devices?
2. How do you manage massive hemorrhage? Is there an optimal ratio of which blood products to use?
3. What are the morbidities associated with massive transfusion? What electrolyte anomalies do you anticipate and how do you correct them? Describe TACO and TRALI and the time course expected for each to manifest.
4. Compare classic coagulation tests to viscoelastic hemostatic assays. Is there an advantage to either? What are the goals in “goal directed therapy” regarding resuscitation of massive hemorrhage?
5. Is there a role for antifibrinolytics in acute surgical hemorrhage?
6. How do you judge that your resuscitation efforts are effective?

**Case History – Case Progression:**
The Nuss bar has lacerated the pericardium and perforated the anterior and lateral walls of the right ventricle. While you and your anesthesia team resuscitate the patient, a cardiac surgeon assists with repair of the ventricle.

A third large bore IV and a radial arterial line are inserted. Epinephrine and albumin are administered to maintain perfusion until emergency release blood is obtained. The massive transfusion protocol is initiated. Over the course of the resuscitation, 4 units of PRBC, 800 mL of cell saver blood, 2 units of FFP, 1 unit of platelets and 1 unit of cryoprecipitate are administered. Calcium chloride and sodium bicarbonate are also administered. Blood loss is
estimated at 3 liters. Hemostasis is achieved. The patient is well perfused with a normal blood pressure and normal sinus rhythm on the EKG.

**Question:**
1. What is the goal of intraoperative debriefing versus root cause analysis or formal review?
2. When should the family be notified of the event? Do you accompany the surgeon to talk to the family about the intraoperative events?
3. What is the second victim syndrome? How do you support your OR staff and surgeons after an adverse event?

**Case History – Progression:**
After the patient is stable, the team takes a moment to debrief the event. The attending surgeon suggests that now would be a good time to place the Nuss bar. The surgeon also asks if you plan to extubate the patient since he is well resuscitated with a normal ABG.

**Question:**
1. Do you agree to proceed with the Nuss bar? If so, should the family be made aware of the cardiac injury first and consent? If you do not agree, how do you negotiate with the surgeon?
2. What are some negotiating tactics to deescalate conflict?
3. What are the risks versus benefits of extubating the patient?
4. Would you extubate the patient at the end of the case? Should the patient go to the PACU or ICU at the end of the case?

**Case History – Progression:**
You review the risks and benefits of proceeding with the repair with the surgeon. You advise the surgeon that the presence of a Nuss bar may make chest compressions or further surgical repair of the heart more difficult if needed. You and the surgeon agree that the best course is to abort the procedure. Bilateral chest tubes and a pericardial drain are placed and the thoracic incisions are closed.

The patient is taken to the cardiac intensive care unit intubated for postoperative care. His postoperative course includes brief hypertension that is treated with nicardipine. He has several short runs of pulsatile ventricular tachycardia treated with lidocaine. He does not suffer from fluid overload or respiratory complications and is extubated POD#1. There are no neurologic sequelae.

**Discussion:**
- The Nuss procedure for pectus excavatum is a common procedure in adolescents. Thoracic injuries are rare but may be catastrophic. Early recognition and immediate surgical exposure and repair are crucial for survival.
- Massive transfusion recommendations in children are evolving. Transfusing PRBC:FFP:Platelets in equal ratios and using viscoelastic hemostatic assays are
reasonable approaches. Antifibrinolytics may be indicated in massive intraoperative hemorrhage.

- Avoidance of hypothermia and acidosis during resuscitation is obligatory. Hypocalcemia and hyperkalemia are to be expected. Transfusion related circulatory overload or transfusion related acute lung injury may occur.
- Adverse intraoperative events may be debriefed in the OR when the patient is stable.
- Conflict management is an important skill for peak team performance.

References:

“I Don’t Want to be Intubated…” Managing Laparoscopic Surgery on a Duchenne Muscular Dystrophy Patient with Severe Restrictive Pulmonary Disease

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Objectives:

1. Review pathology and physiology pertinent to the care of patients with Duchenne muscular dystrophy.
2. Discuss induction and airway management strategies for a patient with severe restrictive pulmonary disease that wants to avoid intubation.
3. Discuss challenges that patients with Duchenne muscular dystrophy present including age-related changes that occur.
4. Review the pediatric anesthesiologist role in the perioperative surgical home for patients with Duchenne muscular dystrophy.

Case Description:

This is a case of a 17-year-old, 40kg male with Duchenne muscular dystrophy (DMD), neuromuscular weakness, severe restrictive respiratory failure requiring nasal BiPap with a FVC of 10% presenting for laparoscopic-assisted gastrostomy tube placement for malnourishment.

- What is Duchenne muscular dystrophy (DMD)? How does it differ from Becker muscular dystrophy?
- What is the pathophysiology of DMD?
- What are the anesthetic implications associated with DMD?

Patient refuses to have this procedure done unless it can be performed without intubation due to fear of needing postoperative ventilation and possible tracheostomy as this occurred with other family members with similar disease.

- What questions are important in preop consult for this patient and his family? Any concerns with consent?
- What anesthetic techniques could be employed to facilitate the surgical procedure?
- Any further studies/consults needed preoperatively?

TTE performed earlier was technically difficult due to ultrasound window limitations. No intracardiac structural abnormality seen in the images able to be obtained. Low normal LV systolic function. Biplane left ventricular ejection fraction 60%. Acoustic windows cause LV wall measurements and shortening fraction of 26% to be of questionable accuracy. Follows with cardiology and currently on carvedilol.

The patient and family ask if an epidural is an option despite his spinal fusion 5 years prior.
• Can an epidural be safely placed after a spinal fusion?
• What about a spinal anesthesia? If yes, is that a safe option in this patient?

The patient has decided he would like to proceed with the operation under a combination of sedation and regional technique. He is moved to the operating room and full ASA monitors are applied. The patient states he is quite anxious.

• What are the options to maintain respiratory mechanics?
• What sedation goals and medications should be used?
• Is nitrous oxide a good option?

You administer midazolam 2 mg IV and initiate a dexmedetomidine infusion; the patient is visibly less anxious. His nasal BiPap is continued from preop. Vital signs are stable and his respiratory mechanics are unchanged.

• What peripheral nerve block would be effective for this procedure? Will they provide coverage during insufflation?
• What local anesthetic and concentration would you use in this 40 kg patient?

You proceed with a bilateral rectus sheath and unilateral left transversus abdominis plane block under ultrasound guidance. The surgeon is eager to begin and the peripheral nerve block seems to be working. The surgeon makes incision and the patient states he senses sharp pain.

• What are analgesic options for this patient?
• Should narcotics be avoided in DMD patients?

The surgeon infiltrates lidocaine into the wound and the patient denies any further pain. You can see the insufflation equipment being readied.

• What are the hemodynamic and respiratory changes you expect with abdominal insufflation?
• Are there any additional medications or alterations to the current anesthetic management that you would do?

You initiate a remifentanil infusion and inject a ketamine 10mg IV bolus. The abdomen is insufflated and the patient asks “have we started the surgery yet?” His hemodynamics remain stable and his hypopnea improves when you titrate his remifentanil down. The remainder of the operation proceeds without issue.

• What would you do if his hypopnea did not improve?

The sedation was stopped and he was taken to the recovery room. In the recovery room he complains of an 8/10 abdominal pain over the gastrostomy site.

• Why does a continuous infusion of remifentanil cause hyperalgesia?
• What postoperative pain management strategy should be deployed for this patient?
Discussion:

Concepts pertinent to the anesthetic management of patients with DMD and the challenges that these patients present is evident from the discussion above. Optimization of care in DMD patients has led to increased challenges for the pediatric anesthesiologist for even routinely basic procedures including gastrostomy tubes and sedation for imaging. It is important to maximize these patients in the perioperative period with comorbidities including limited cardiac function and chronic respiratory depression. Specific focus will highlight the physiological changes expected as these patients age and how to adjust anesthetic planning. We will also focus attention on various types of induction methods and anesthetic maintenance strategies that can be used when trying to avoid endotracheal intubation as applicable to this case.

The muscular dystrophies are a group of hereditary degenerative disorders with varying phenotypes associated with progressive muscle weakness. With greater than 30 different genotypes, a characteristic shared by all is muscle weakness; however, the distribution of weakness distinguishes the type. While de novo mutations can occur, most follow a Mendelian pattern including autosomal dominant, autosomal recessive or X-linked transmission. For the purpose of this discussion, we will focus on the X-linked recessive disorders: Duchenne & Becker muscular dystrophy.

Duchenne muscular dystrophy (DMD) & Becker muscular dystrophy (BMD) are rare dystrophinopathies caused by recessive mutations in the dystrophin gene on the short arm of chromosome X (Xp21). DMD is the most common form of inherited muscle disease in childhood with an approximate incidence of 1 in 3500 males, whereas BMD has an estimated incidence of 1 in 18,500 males. Dystrophin is a large protein molecule on the surface of skeletal muscle cells and regulates the integrity of the sarcolemma, a membrane enclosing striated muscle cells. Mutations in the dystrophin protein leads to dysfunction of the sarcolemma leading to myofibril atrophy, necrosis and finally fibrosis.

BMD patients have a mutation resulting in a partially functional dystrophin molecule producing a milder and more variable form of disease. DMD mutations result in total absence of dystrophin. These mutations result in progressive muscle weakness and cardiomyopathy. Symptoms become apparent around 2-3 years of age and by age 5, nearly all patients are symptomatic evidenced with loss of independent ambulation. As they continue to age, respiratory insufficiency occurs leading to recurrent pneumonias, and cardiomyopathy ensues affecting 90% by age 30; death typically occurs in the 3rd decade of life for those with DMD. BMD patients are often afforded longer lifespans.

In 2009 the American College of Chest Physicians published a consensus statement on management of patients with DMD undergoing GA or sedation. They advised that all DMD patients should have preoperative respiratory assessment including room air SpO2 monitoring to assess gas exchange and forced vital capacity (FVC) to assess lung volume. If the SpO2 reading is less than 95%, then further invasive monitoring or EtCO2 levels should be investigated as gas exchange is impaired. Next, if FVC < 50% is found, the patient is at risk for postoperative complications, but FVC < 30% shows they are at high risk for postop complications. The consensus statement advocated for use of non-invasive positive pressure ventilation (NPPV) immediately in the postoperative phase, extubating to NPPV whenever possible; especially if utilized pre-operatively. The group also encouraged avoiding general anesthesia in favor of
neuraxial or regional techniques whenever possible for these patients with low pulmonary function.

Aside from the pulmonary issues, patients with DMD can present other challenges. Nearly all older children have bulbar muscular weakness resulting in dysphagia that increases risk of aspiration along with glossomegaly and limited mandible mobility posing airway management issues. Quite commonly, DMD patients have dilated cardiomyopathy resulting in increased risk of arrhythmias and potential cardiovascular instability intraoperatively. Therefore, a preoperative electrocardiogram and echocardiogram assessment are essential for strategy management; a cardiology consultation should heavily be considered. Another intraoperative consideration is dystrophinopathies frequently have hyper-metabolic responses with inhalational volatile anesthetics that have been implicated in causing rhabdomyolysis. This concern has led some anesthesiologists to recommend avoiding volatile inhalational agents if possible. Importantly though, succinylcholine administration has absolutely been shown to cause rhabdomyolysis leading to hyperkalemic induced cardiac arrests and is considered an absolute contra-indication in patients with dystrophinopathies.

References:


