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I Passed Gas in the Operating Room. Are Volatile Agents Really Silent and Deadly to Patients With Muscular Dystrophy?

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Stem Case and Key Questions Content

It's a Sunday evening and you are on call as a general anesthesiologist at a large community based hospital. At 8 pm you are called by the pediatric general surgeon to do an exploratory laparotomy on a 10 year old male. The surgeon states that he needs to take the child to the operating room urgently secondary to his clinical condition. By the way, the surgeon says, " I think he has Duchenne Muscular Dystrophy (DMD)." *What is the pathophysiology of muscular dystrophy? Is this a case that a general anesthesiologist should do or should one call in the pediatric anesthesiologist on call?*

The patient is a 10 year old male with DMD, wheel chair bound and ill appearing. According to his parents, he was in his usual state of health until 4 days prior when he began to have a gastroenteritis illness. Several members of the family had similar symptoms, however, his symptoms have progressed to severe abdominal pain, intermittent nausea and vomiting, and fever. The patient does interact with you, but is noticeably uncomfortable. He has a 24 gauge peripheral IV (right hand) which was placed after several failed attempts. His parents state there is no family history of anesthetic problems and that the patient had a spinal fusion with hardware instrumentation which was complicated by significant blood loss and prolonged inbutation in the intensive care unit. He has seen a pulmonolgist and cardiologist in the past. *What are the specific suggestions for preoperative evaluation of patients with DMD before general anesthesia or procedural sedation? Are children with DMD at higher risk for blood loss during surgery? Should this patient have coagulation studies prior to going to the OR?*

The patient's cardiac and pulmonary records have been located. He is noted to have severe restrictive pulmonary disease with a forced vital capacity (FVC) of 26% predicted, and a forced expiratory volume (FEV1) of 25% predicted. He is not using non-invasive positive pressure ventilation assistance and has not had a recent pneumonia. He is followed by cardiology and his recent transthoracic echocardiogram was notable for low normal left ventricular systolic function. Additionally, a Holter monitor showed no atrial or ventricular ectopy.

The patient appears ill, cachetic and is wheel chair bound. His vital signs are notable for a pulse rate in 120-140's, BP 95/60, RR of 20 and O2 sat of 97% on 2 L nasal cannula. He is 45 kg, 149 cm tall with a body mass index (BMI) of 15. He has a Mallampati 3 airway on exam and has been NPO for greater than 8 hours, but has vomited some yellow bilious material in the past 3 hours. *What additional preoperative information including physical examination, laboratory and radiological studies would you request? How will you determine if the patient is optimized to go*

to the operative room?

In the emergency department he has received 1 liter of lactated ringer's solution, and a dose of antibiotics. Initial lab-work and the abdominal X ray have been completed. The preliminary reading on the abdominal X ray is “notable for dilation of stomach and large amount of free air.” Luckily you are working with an enthusiastic resident, but they have a lot of questions, including: *How will a rapid sequence induction be done; will you use succinylcholine? Are these kids at increased risk of malignant hyperthermia? What is your plan for maintenance of anesthesia? What is the role of TIVA in children with DMD? Is there a safe amount of inhalation agent to be used? What types of monitoring will you use in this child? How will you obtain optimal IV access?*

Standard ASA monitors are applied in the OR. You’ve decided to proceed with a modified rapid sequence induction in this case (propofol/rocuronium/fentanyl) and a TIVA was planned for maintenance of anesthesia with propofol and remifentanil along with a BIS monitor. Upon direct laryngoscopy you are unable to see the glottic opening. *What is the likely mechanism of a difficult airway in a patient with DMD?* The patient’s oxygen saturation is now 78% and falling, you try to mask the patient but cannot move air. You are in a cannot intubate, cannot ventilate scenario (CI/CV). You want to call for help, but nobody else is around. You do have an anesthesia tech who brings you the difficult airway cart (loaded with a fiberoptic scope). *Would you go directly to a fiberoptic scope and intubation or would you place an LMA in this CI/CV situation?*

An intubating LMA is placed a-traumatically and swiftly. You see a carbon dioxide tracing and hear normal bilateral breath sounds on auscultation. You’re able to ventilate with relative ease and the pulse oximetry is now 99% on 100% oxygen. You then are able to carefully intubate with help of the fiberoptic scope through the LMA.

You notice that the 24 G PIV is no longer flowing and upon inspection it appears to have infiltrated. Prior to induction your BIS read 90 and after induction it read 40. You proceed to try for another PIV but the patient has many bruises and blown veins, and you are unable to place a PIV. The surgeon arrives 5 minutes later and you ask them to help you place a central line. The BIS now reads 90.

With no IV access and a rising BIS you decide to turn on Sevoflurane to 1.5%, after 3 minutes the BIS reads 55. The surgeon ultimately places a triple lumen central line in the left subclavian space, however the process was prolonged secondary to the patient’s contorted anatomy and has taken 30 minutes. While the surgeon was working you are able to place a right radial A-line. Now that you have adequate central IV access you turn off the inhalation agent and begin the TIVA (propofol/remifentanil) infusion. You aim for a BIS of 50 which is achieved.

The abdomen is prepped and draped and you administer antibiotics. The OR desk informs you that two units of crossmatched packed red blood cells are available. Approximately 5 minutes later, the incision is made. The surgeon works diligently and with meticulous dissection the problem is revealed: superior mesenteric artery syndrome. A rare, but well known entity where a portion of the duodenum is compressed against the aorta by the SMA resulting in chronic, intermittent or acute obstruction. *What are the risk factors for developing SMA syndrome?* The surgeon is satisfied with the findings and is able to perform the necessary intervention of

duodenal derotation, gastrostomy, and jejunostomy. The whole operative time has been just over 90 minutes and the patient has tolerated the procedure well. The blood loss is estimated at less than 100 ml. Hemodynamically the patient has been stable, but urine output has been scant. You're glad that the case is finishing up. The resident asks you about the risk of propofol infusion syndrome (PRIS) in this patient. *What is PRIS?* You calculate that his total propofol dose has been 13.5 mg/kg, or 9 mg/kg/hr for 1.5 hours. *Are you worried about this propofol dose potentially causing propofol infusion syndrome (PRIS)?*

As the surgeon is completing the final sutures, he asks you about your plan for post op pain control and requests an epidural. *Describe your postoperative plan for analgesia. Is an epidural an appropriate means of analgesia in this patient? What about a TAP block?*

You decide to place a TAP block with ultrasound guidance and achieve excellent visual results and infiltrate the space with ropivacaine 0.5% 0.2ml/kg directly between the internal oblique and the transversus abdominis muscle.

The patient appears to be making some efforts to breathe spontaneously. You check the TOF twitch monitor and still only have 2/4 twitches. *How is the neuromuscular blockade different in DMD children than non-DMD? Will you attempt to reverse his neuromuscular blockade?*
You decide to go directly to the PICU intubated. As you move the patient from the OR table to the stretcher you notice a change in the ECG. The T waves have become peaked and the blood pressure has dropped. You listen for breath sounds, which are clear and peak airway pressures are normal. You scan the patient head to toe and do not see any evidence of a rash. His temperature is 36.2 and stable, ETCO2 is 36 and stable, however the ECG has now evolved to ventricular fibrillation. *What are the initial steps in the management of a pediatric cardiac arrest? What pharmacological and or electrical therapy would you choose and in what sequence? What are the likely etiologies of his cardiac arrest?*

Model Discussion Content

I. Introduction:
Duchenne muscular dystrophy (DMD) is a disease characterized by progressive loss of muscle strength, eventually resulting in loss of ambulation, loss of respiratory muscle strength, and death from cardiopulmonary causes (1). DMD is an X-linked recessive trait that occurs almost exclusively in boys. The incidence of DMD is approximately 1:3,500 male births and is caused by mutation of the dystrophin gene. The progression of the disease if rapid, resulting in a failure to walk by adolescence and eventual death from cardiopulmonary causes by the end of the third decade. Dilated cardiomyopathy occurs in over 50% of patients by 15 years of age (2).

Although many of these patients undergo anesthesia and surgery without complication, perioperative adverse events are not uncommon (3,4). Acute rhabdomyolysis is one such event, and is thought to be triggered by the administration of succinylcholine, a depolarizing muscle relaxant. Potent inhalational agents have also been implicated as a cause of rhabdomyolysis and other perioperative metabolic reactions that resemble malignant hyperthermia (MH) (5,6). However, there are case reports of intraoperative cardiac arrest in DMD children during spinal

surgery under propofol-sufentanil anesthesia without succinylcholine or volatile agents (7,8). Suffice it to say, patients with DMD are especially vulnerable to the adverse physiologic effects of general anesthesia and procedural sedation, prompting the need for expert care and recommendations on the topic (9,10).

II. Preoperative evaluation

Numerous problems face the anesthesiologist called upon to manage anesthesia in DMD children, especially older ones who are wheelchair dependent. There is no way of identifying with any degree of accuracy, the boys who are likely to develop anesthesia related complications (3). Obesity is common in the older boys and they frequently have hypertrophy of the tongue making intubation and mask ventilation more difficult. Many will have associated deformities and contractures of the limb joints that will hinder vascular access and also positioning on the operating table will need extra care to prevent pressure necrosis (1). Children with Duchenne muscular dystrophy tend to bleed more during surgery than do children with other conditions. It is postulated that the lack of dystrophin in smooth muscle leads to an ineffective vaso-constrictive response leading to increased blood loss. Type and cross matched blood is prudent to have available prior to surgery where there is potential for significant blood loss (11).

Respiratory function is compromised both by the progression of the disease and collapse of the spine, due to muscular weakness, resulting in scoliosis that adds to the pulmonary restrictive deficit (1). At minimum one should obtain anesthesiology and pulmonolgy consults before procedures involving general anesthesia or procedural sedation.

The pulmonary evaluation should include measurement of forced vital capacity (FVC), maximum inspiratory pressure (MIP),maximum expiratory pressure (MEP), peak cough flow (PCF), and oxyhemoglobin saturation measured by pulse oximetry in room air. Measurement of the blood and/or end tidal carbon dioxide level if SpO2 is <95% in room air is suggested. The FVC is a helpful predictor of which patients are at greater risk for respiratory complications. For an FVC of <50% of predicted patients are at high risk for complications, consider preoperative training in the use of non-invasive positive pressure ventilation (NPPV). For patients at high risk of ineffective cough, defined by PCF <270 L/min or MEP <60 cm H20 consider preoperative training in manual and mechanically assisted cough, emphasizing use of mechanical insufflation-exsufflation (MI-E) with a bronchial secretion clearing device (9,10).

Cardiac involvement is universal in individuals with DMD. Cardiac disease is the second most common cause of death in persons with DMD, with 10–20% of individuals dying of cardiac failure (1,2,9,10). Dilated cardiomyopathy primarily involves the left ventricle, and can lead to dyspnea and other symptoms of congestive heart failure (1,2,10). Conversely, right ventricular failure can result from respiratory failure and pulmonary hypertension. Individuals with DMD are

also at risk for ventricular arrhythmias (2,9,10). Whereas some studies have suggested that the respiratory and peripheral muscle weakness tend to be inversely related to the risk of cardiac failure, other studies suggest that left heart and respiratory failure tend to occur in parallel (1,2,9,10). There are retrospective data suggesting that cardiac involvement is less frequent in children treated with deflazacort (2). The current recommendation is for all individuals with DMD to have regular cardiac evaluations with annual ECG's and echocardiograms, starting at least by school age (9,10).

Good nutritional support is integral to the proper care of patients with DMD. Preoperative nutritional status should be evaluated and optimized because poor nutrition can increase postoperative morbidity. Consider measuring serum albumin and pre-albumin to identify patients at risk for poor wound healing. Also, one should consider strategies to manage dysphagia, as inability to eat postoperatively can lead to malnutrition (12) .

DMD is a progressive and fatal disease. Thus, advance directives (including resuscitation parameters) and attitudes toward prolonged dependency on mechanical ventilation and tracheostomy should be discussed with patients and their guardians preoperatively (10). Respiratory failure in DMD can occur either suddenly, in association with a respiratory tract infection, or gradually; education about ventilatory and palliative options should be provided before either of these scenarios occurs. The patient's and family's views on quality of life should be sought. These types of decisions are most difficult in the rare case of the young child with severe respiratory muscle weakness, who is too immature to participate in the discussion. End-of-life directives established by the patient, family, and health care team must be clearly documented and available for use in the case of an emergency (10).

The ASA provides a statement of practice recommendations for the care of pediatric patients. "Optimal perioperative care of infants and children requires proximate availability of qualified medical personnel and contemporary equipment designed specifically for this purpose. Local and regional circumstances may differ with respect to the immediate availability of specialized personnel and access to facilities" (13). At a large west coast university children's hospital, the university service has a team of pediatric anesthesiologists with specialized training who attend to each pediatric anesthetic. When private or university attendings reapply for medical staff privileges every 2 years, they are required to tally the number of children they have anesthetized in the following age groups: (a) newborn to 6 months, and (b) 6 months to 6 years. A minimum number of cases is needed to maintain privileges.

Things are different at a community hospital, where a smaller team of anesthesiologists shares night call. Unless the hospital is very large, it is uncommon to have multiple specialist anesthesiologists on call each day, e.g. one for pediatrics, one for cardiac cases, one for trauma, one for obstetrics, and one for the general OR. It is common for general anesthesia

practitioners to cover many or all specialties when they are on call. If they are not comfortable with an individual case, they can seek out a better trained anesthesiologist, if one is available. The trend for having a specialist anesthesiologist for every type of case, at all hours of the night and weekend, is a difficult one to staff. The decision to care for a patient at a community hospital is a judgment as to whether standards of care can be met with the physicians who are available.

It is generally the consensus that neonates and young infants should be cared for by anesthesiologists with specialized pediatric training. Whether specialized training should be mandated for older children is debatable. Policies to define a minimum age limit for patients of general anesthesiologists may be a hot topic for the future (13).

III. Intraoperative management

Patients with DMD are at increased risk for rhabdomyolysis when they are exposed to certain anesthetics, especially inhaled agents such as halothane, isoflurane, sevoflurane, and desflurane (1,4,5). Such episodes can cause hyperkalemia and sudden cardiac arrest. These events mimic malignant hyperthermia, but DMD and MH are genetically distinct diseases (1,4,14).

Succinylcholine, a depolarizing muscle relaxant, has been linked to acute rhabdomyolysis, hyperkalemia and cardiac arrest in patients with DMD. While succinylcholine is widely recognized to be contraindicated in patients with DMD, more recently it had been suggested that inhaled anesthetic agents should be considered contraindicated for patients with DMD (9,10,14).

Although only a small proportion of DMD patients develop anesthesia induced rhabdomyolysis (AIR) after exposure to inhalational agents, the question is: should we continue to use inhalational agents when TIVA is a safe and readily available alternative. The opinion in the literature seems to be shifting towards the TIVA approach (4,9,10,14,15). In fact some authors even propose a “trigger free” anesthetic and a “clean” anesthesia machine be used, similar to that for MH susceptible patients (14). This recommendation is based on the fact that the minimum triggering concentrations of inhalational agent remains unknown.

Anesthesia induced rhabdomyolysis has occurred after anesthetics of 10 minutes duration to longer uneventful operations and then cardiac arrest in the recovery room upon spontaneous movement by the patient (1,4,14). Some authors recognize that certain clinical situations, such as a DMD patient with the potential for difficult airway and inability to secure an intravenous line, as an option for a short exposure to an inhalational agent. However, once the airway is secured and intravenous access has been successful immediate conversion to a TIVA and a clean anesthesia machine is recommended. The child should be carefully monitored for signs of rhabdomyolysis (14).

General anesthesia should be performed with an anesthesiologist in attendance and with full monitors and safety measures, according to the guidelines of the ASA. Additional monitors to be considered in this case would be a Bispectral index (BIS) monitor to help evaluate the depth of anesthesia during a TIVA, a central venous pressure (CVP) line to help guide volume replacement, an invasive arterial line for continuous measurement of blood pressure in a patient who is critically ill and showing signs of poor organ perfusion, and a foley catheter to help determine volume status as well as a potential sign of ensuing rhabdomyolysis (dark urine).
As stated earlier, these children can have joint abnormalities and contractures making intravenous access very difficult. Having an anesthesiologist experienced in the management of DMD can be helpful (9,10). Use of ultrasound technology can greatly aid in the placement of PIV's and CV catheters too. In fact, studies among emergency physicians have shown increased success in cannulation of vessels with a shorter time to execute the procedure in difficult patient populations (16).

IV. Miscellaneous and Analgesic Plan

This child had an acute abdominal disease process that could have been a myriad of disease entities, but was diagnosed as Superior Mesenteric Artery (SMA) Syndrome. This is a rare but well recognized entity characterized by compression of the transverse portion of the duodenum against the aorta by the SMA, resulting in chronic, intermittent or acute complete obstruction. Any process that sharply narrows the aorta-mesenteric angle can cause entrapment and compression of the third part of the duodenum as it passes between the SMA and the aorta. In addition to the vague abdominal symptoms, there is usually a history of scoliosis corrective surgery. The cause after spinal surgery is thought to be due to the spinal elongation, which decreases the superior mesenteric/aortic angle. Additional risk factors are thin body build, depletion of mesenteric fat and malabsorption (17).

Propofol infusion syndrome (PRIS) is a clinical entity of acute refractory bradycardia leading to asystole in the presence of one or more of the following: metabolic acidosis, rhabdomyolysis, hyperlipidemia, and enlarged or fatty liver. There is an association between PRIS and propofol infusions at doses higher than 4 mg/kg/hr for greater than 48hr duration. However, some patients receiving propofol in typical doses for short surgical procedures presented with symptoms typical for PRIS but without cardiac involvement and the reporting authors argued that these symptoms may have been precursors of PRIS. Predisposing factors include young age, severe critical illness of the central nervous system or respiratory origin, exogenous catecholamine or glucocorticoid administration and subclinical mitochondrial disease (18).
Achieving optimal analgesia in children with DMD is very challenging because of their co-morbid pulmonary and multi-systemic disease states. However, adequate pain control should not be compromised because of concern about suppression of respiratory drive. When patients are sedated after administration of opioid analgesics, adequate ventilation can be achieved by using NPPV continuously or by delaying endotracheal extubation for 24-48 hours (9,10). While pain control is essential, the chance of successful extubation is optimized in an awake, cooperative

patient.

In patients undergoing spinal fusion surgery, neuraxial techniques have been used to achieve analgesia through single dose or continuous infusion of opioids and local anesthetics via epidural catheters with minimal respiratory side effects (19).

This child's anatomy of a surgically fused spine with hardware poses a technical challenge for placement of an epidural. Additionally, their current clinical situation of a febrile state with hypotension may be a sign of sepsis. An indwelling catheter may be considered too high a risk in a potentially bacteremic patient (20). An alternative method that is gaining popularity in abdominal procedures for post operative analgesia is a transversus abdominis plane (TAP) block. A recent meta analysis of the TAP block showed that it is safe, reduces postoperative morphine requirements, nausea and vomiting and possibly the severity of pain after abdominal surgery (21). It should be considered as part of a multimodal approach to anesthesia and enhanced recovery in patients undergoing abdominal surgery.

In patients with DMD, the onset of blockade by non de-polarizing neuromuscular blockers may be significantly delayed, which should be kept in mind in situations when a rapid airway protection is necessary. Furthermore, the prolonged recovery from rocuronium induced block emphasizes the need for neuromuscular monitoring in DMD patients (22).

V. Intraoperative cardiac arrest

Initial steps during a cardiac arrest should include calling for help and the code cart/defibrillator. Verification of a secure airway and ventilation should follow. Chest compressions should be started without delay. The FiO2 should be 100% with the vaporizers turned off. Emergency drugs can be delivered by the endotracheal or intraosseous routes if loss of intravenous access should occur (23).

The 2010 Pediatric Advanced Life Support (PALS) guidelines should be followed. The first decision to be made is if the rhythm is shockable (VF/VT) or not shockable (Asystole/PEA). For shockable rhythms such as VF or VT, defibrillate one time at 2 J/kg and resume CPR immediately. Five cycles of CPR follow. If the rhythm is still shockable, defibrillate once at 4 J/kg and resume CPR. After defibrillation, epinephrine is given every 3-5 minutes. The dose for epinephrine is 0.01 mg/kg IV or IO and 0.1 mg/kg if given via the endotracheal tube. Five cycles of CPR then occur followed by evaluation of the rhythm. If the rhythm is shockable, defibrillate at 4 J/kg followed by resuming CPR. One then will administer antiarrhythmics. Antiarrhythmics include amiodarone 5 mg/kg IV push or lidocaine 1 mg/kg. This cycle of 1 shock followed by 5 cycles of CPR is repeated until a decision is made to terminate efforts or the rhythm is not shockable (23). One should attempt to identify and treat possible contributing factors such as hypoxia, hypovolemia, acidosis, hyperkalemia, pneumothorax, trauma, and cardiac tamponade.

Based on this case scenario, hyperkalemia should be strongly considered.
Several etiologies of this cardiac arrest can be entertained: allergic reaction and anaphylaxis, malignant arrhythmia, propofol infusion syndrome, anesthesia induced rhabdomyolysis, malignant hyperthermia, and local anesthetic cardiotoxicity to name a few. Sorting these out in the heat of the moment can be challenging and one must maintain a broad differential diagnosis, but look for clues to a causative event.

In this case PRIS would be unlikely given the dose and duration of propofol. A malignant arrhythmia certainly could have caused a cardiac arrest especially in a child with a cardiomyopathy, again, this would likely be supported by deviations in electrolytes. Anaphylaxis with cardiovascular collapse would be unlikely without signs of cutaneous flushing or erythema, wheezing or increased peak pulmonary pressures, however, antibiotics and neuromuscular blocking agents were used during the case and thus one must have a high index of suspicion. If contemplating an anaphylactic/anaphylactoid reaction, then a serum tryptase level can be obtained and help with the diagnosis.

Local anesthetic toxicity could certainly lead to cardiac arrest in the setting of an inadvertent intravascular injection. Abiding by safety measures of limiting the total dose of local anesthetic, frequent aspiration and injection of small volumes over 30 seconds while continuous ECG monitoring is recorded can decrease the incidence of toxicity, however, there is no fail safe method. The onset of cardiotoxicity is usually quite fast, but it can be delayed up to one hour. Although bradycardia and asystole are the most common presenting arrhythmias; tachyarrhythmias can occur as well. The American Society of Regional Anesthesia (ASRA) strongly advises anesthesiology departments to establish a plan for managing systemic local anesthetic toxicity at their facility. This should include stocking 20 percent lipid emulsion and the means for its rapid delivery close to every site where local anesthetics are used. Having a Local Anesthetic Toxicity Kit is encouraged (24).

However, the known exposure of inhalational agent in a susceptible patient, along with clinical evidence of acute rhabdomyolysis (peaked T waves, hyperkalemia, cardiac arrest) leads one to consider AIR vs. MH in this case. MH would be unlikely given the lack of rapidly rising ETCO2, muscle rigidity, and hyperthermia.

However, in the heat of the moment who would with-hold the treatment of dantrolene? Currently, the role of dantrolene in the management of AIR is unknown. The mechanism of action of dantrolene for the treatment of MH is likely inhibition of excessive release of calcium from the sarcoplasmic reticulum by binding to the ryanodine receptor isoform 1. Dantrolene may be of no use in AIR as the proposed mechanism involves the breakdown of muscle cell membranes and subsequent leakage of cell contents. The Malignant Hyperthermia Clinical

Grading Scale is a useful resource for retrospective analysis of the event if uncertain about the diagnosis (25).

Children with DMD represent a complex patient population that will continue to need routine elective and emergency operations. Although, it is unclear what the safest anesthetic technique is for these children; what is clear is that they are prone to develop serious complications during the perioperative period and can benefit from a multi-disciplinary care team skilled in the unique needs of these individuals.

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