Spinal Fusion, Spinal Muscular Atrophy, Signal Loss and Blood Loss in a Jehovah’s Witness Patient
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Disclosures: These presenters have no financial relationships with commercial interests.

Stem Case and Key Questions Content
Jacklyn is a 17-year-old wheel chair dependent female with worsening neuromuscular scoliosis secondary to spinal muscular atrophy type 2. She presents for a T2 to pelvis posterior spinal fusion. She has a history of a failed intubation 11 years ago. She is on BiPAP at baseline secondary to her neuromuscular disease but can tolerate short periods off BiPAP relatively well. She is free from any recent pneumonia or upper respiratory tract infections. The pulmonology clinic attempted a pulmonary function test but was unsuccessful. She is G-tube dependent secondary to her inability to swallow. An ECHO was obtained preoperatively and showed a bicuspid aortic valve with mild regurgitation, but was otherwise unremarkable.

1. What are the anesthetic concerns for a patient with SMA?
2. What are the options for obtaining a definitive airway in this patient?
3. How would you assess this patient’s ability to tolerate an awake intubation?
4. What instruments or plans would you want in place before attempting an airway on this patient?

You perform a physical exam on Jacklyn and note that she is calm, and able to follow commands. She has limited cervical spine motion with ankylosis of the jaw. On examination, her mouth opening is less than 1 fingerbreadth. Though she is hypotonic, she is able to tolerate 10 minutes off of her BiPAP without desaturations in the preoperative area. Her lungs sound clear, and when asked to demonstrate a cough, you note she has mild limitation in her ability to produce a forceful cough.

5. How does this change your plan for induction?

While obtaining consent for your anesthetic plan the parents of the patient disclose that the family
members are Jehovah’s Witnesses and they do not wish for their daughter to receive blood products.

6. What options are available for providing volume and red cells to a patient who is a Jehovah’s Witness?

7. What kind of preoperative and intraoperative interventions can be done to conserve blood loss?

8. In what circumstances are you legally obligated to give blood products despite the family’s wishes?

The family and patient decide that they are okay with products such as albumin and crystalloids. Cell saver will be available in the room and the patient has agreed to autologous blood transfusion.

9. What are the contraindications for intraoperative blood salvage techniques?

You elect to do a FO awake intubation using nebulized lidocaine and versed. The best view obtained on your fiberoptic is a grade III.

10. What could be a suitable airway technique or device to use next?

After starting your case, the technician monitoring evoked potentials notices a loss in SSEP and MEP signals.

11. What are the potential causes of evoked potential signal loss?

12. What interventions can be done to restore evoked potential signals?

After turning off your volatile anesthetic the technician reports the SSEPs have improved, but the MEPs still appear dampened.

13. What other techniques can help restore signals?

At the conclusion of your case you are making preparations for admission and transport to the ICU. Your patient is spontaneously breathing at a rate of 18, on a support of 15/5 with an FiO2 of 100%. EtCO2=45 She is not yet following commands.

14. Does your patient meet extubation criteria?

15. What is your plan for extubation in this patient?
Model Discussion Content

Spinal muscle atrophy (SMA) is an autosomal recessive neuromuscular disease and one of the most common recessive inherited lethal diseases in children. SMA is classically divided into three groups: SMA I, SMA II, and SMA III. Classification is based on age at onset and best achieved motor function. Early onset of symptoms correlates with greater severity and more rapid progression. In general muscles are affected proximally more than distally and upper limbs are generally more affected than lower limbs. In severe cases, hyporeflexia and sensory nerve dysfunction can be seen. In general, sensory nerve function is intact and intellectual and cognitive function is typically normal. [1]

SMA I- Symptoms manifest before 6 months of age, and patients require artificial ventilation to survive. Noninvasive ventilation is preferred over invasive ventilation as it promotes chest wall development and lung function. Both motor neurons and the nuclei of lower cranial nerves are affected leading to poor head control and dysphagia. Swallowing difficulties and regurgitation are of concern. Cardiac malformations and autonomic dysfunction are rarely associated with SMA I but can be seen with more severe genetic defects. These patients typically present for procedures such as: gastrostomy, fundoplication, tracheostomy, muscle biopsy.

SMA II- Symptoms manifest between 6-18 months of age. Patients may require artificial ventilation at night due to sleep disordered breathing. Respiratory support may be needed during acute illness and postoperatively. Bulbar symptoms can be seen in more severe forms. Progressive muscle weakness during development often results in kyphoscoliosis which can worsen lung function. Due to abdominal wall weakness, the parturient SMA patient often requires instrumented deliveries or cesarean section. These patients typically present for procedures such as: surgery for scoliosis, club foot, joint contracture release, and muscle biopsy.

SMA III- Symptoms develop after 18 months of age, they often reach all of their major motor milestones including walking before their symptoms develop. Respiratory function is usually normal; however, as patients age the risk for developing sleep disordered breathing and respiratory failure increases. If sleep disordered breathing goes undiagnosed, conduction abnormalities of the heart and cardiomyopathy may develop. These patients often complain of chronic pain from muscles and symptoms related to overuse of joints.

Anesthesia: no evidence-based advice or guidelines have been developed for the management of SMA patients as the disease is rare, and clinical expression varies. Currently anesthetic care must be based upon pathophysiologic knowledge, animal studies, case reports and common sense principles. Preoperative pulmonary evaluation consultation, and optimization, as well as airway evaluation is strongly recommended by most sources.
Airway: Tracheal intubation can be difficult due to cervical spine contractures or ankylosis of the temporomandibular joint. 30% of teens and almost 80% of 30 - 50 year olds will have difficulties in mouth opening. The physician should have a frank discussion with the patient and family regarding possible emergent need of tracheotomy or cricothyotomy perioperatively. Difficult airway carts, fiberoptic tools and intubating-supraglottic airways should be made readily available in those patients likely to be at risk for difficult intubation. In those patients requiring awake intubation due to a known difficult airway, assessing the patient’s cognitive ability to follow directions and ability to breathe while lying flat with unassisted ventilation should be performed prior to entering the operating suite. Premedication in individuals with difficult airways should be administered only after entering the operating suite as it may jeopardize the airway or ventilation. Intubation using techniques that preserve spontaneous ventilation is prudent.

Respiration: Pulmonary complications are the leading cause of anesthetic morbidity and mortality in SMA patients. Though SMA I and II patients are at high risk for pulmonary complications requiring postoperative ventilatory support, SMA III patients rarely are affected. Restrictive lung disease is common, as a consequence of chronic and repeated lung infections due to weak thoracic and abdominal muscles, dysphagia, and regurgitation-aspiration syndromes. Paradoxical breathing is typical as intercostal muscles are weak and in general the diaphragm is unaffected. The imbalance in muscle structures can lead to pectus excavatum, which can contribute to further impairment of lung development. Be cautious of the SMA patient with sleep-disordered breathing, as this is often an early marker of respiratory failure. Extreme use of oxygenation is not recommended and a goal of near baseline saturation on baseline FiO2 settings should be sought prior to extubation. Over oxygenating prior to extubation can mask hypoventilation in these patients. Point of care ABG can help guide extubation. [3]

Circulation: Cardiovascular complications are uncommon, however, the patient with restrictive respiratory failure, scoliosis or obstructive sleep apnea is at a higher risk of impaired cardiac function. Gastrointestinal: Gastrointestinal reflux is a phenomenon that can be seen in any SMA patient; the risk of aspiration must always be evaluated and precautions should be taken if in doubt. Venting and aspiration of the gastrostomy tube should be considered prior to induction of anesthesia to decompress the stomach.

Nutrition: Both hypo- and hyperglycemia can develop in SMA patients. While patients with severe muscle atrophy are at high risk for hypoglycemia, those with over production of glucagon can have hyperglycemia. Mitochondrial dysfunction and abnormal fatty acid oxidation has been described in SMA patients which can lead to problems with hypoglycemic lactacidosis perioperatively. Therefore, monitoring glucose intraoperatively is recommended, especially for longer procedures and for those patients who have a history of impairment of glucose regulation.[4]
Neuromuscular blockade: Patients with SMA often have increased sensitivity to non-depolarizing agents. Intubation without relaxation should be considered. Reports exist of patients requiring prolonged intubation, even in the setting of full reversal of neuromuscular blockade and four equal twitches on train-of-four stimulation. Succinylcholine should be avoided due to risk of hyperkalemia. [5]

Opioids: Should be titrated appropriately, and multimodal pain coverage with non-narcotics agents should be strongly considered. Short acting narcotics (remifentanil) may be preferred over long acting narcotics in the intraoperative phase. Caudal, epidural, or regional anesthesia should be strongly considered as it can potentially eliminate intraoperative narcotics. [2,6]

Postoperative Care: Patients with SMA I and II will most likely require postoperative care in an ICU setting. SMA I patients will most likely require postoperative ventilatory support, while SMA II or III patients may not, depending on progression of illness. Non-invasive ventilation can be a useful bridge from intubation to spontaneous ventilation.

Anesthetic management in the Jehovah’s Witness (JW) patient:
What is Jehovah’s witness?
The Witnesses’ refusal to accept transfusions is based on literal interpretation of the Bible and the belief that any hope of eternal life or salvation is forfeited if they do not strictly adhere to Bible directives. Gardner et al points out, that with accepting transfusion "their corporal malady is cured but the spiritual life with God, as they see it, is compromised, which leads to a life that is meaningless and perhaps worse than death itself". [7]

Legal Issues: A landmark case confirming a competent adult's right to refuse treatment occurred in 1914 in Schloendorff vs. Society of New York Hospital. In this case, a woman agreed to an examination under anesthesia but refused any operative intervention. Once under anesthesia, however, an operative procedure was performed. The presiding judge stated that, "Every human being of adult years and sound mind has a right to determine what shall be done with his own body." [8]

In the United States a citizen's right to freedom of religion allows a competent adult Jehovah’s Witness patient the right to refuse transfusion even though the result of such a refusal may be death. Therefore, the key determinations in a patient’s right to refuse life-saving treatment are "competency" and "adulthood".

Generally those under the age of 18 are considered too young to make medical decisions; however, exceptions can be made for "self-sufficient minors" and "emancipated minors". As an example,
the California Civil Code Section 34.6, a self-sufficient minor is one who is age 15 or older and:

1. lives separate and apart from his or her parents or legal guardian, whether with or without the consent or acquiescence of the parent or legal guardian, and

2. manages his or her own financial affairs, regardless of the source of income.

Similarly, under the California Civil Code Section 62, an emancipated minor is any person under the age of 18 who:

1. has entered into a valid marriage, whether or not such marriage was terminated by dissolution, or

2. is on active duty with any of the armed forces of the United States of America, or

3. has received a declaration of emancipation pursuant to California Civil Code Section 64.

Other States may have different definitions, as may other countries; practitioners should always be cognizant of the laws applicable to their practice location.

Anesthetic Management of the JW patient:
Anesthesia care begins with preoperative assessment and includes the exploration of all options to identify the patient's preferences for blood conserving interventions. Once agreement on acceptable interventions has been made, the physician is ethically and legally obligated to adhere to limitations. An exception to this legal obligation is made when the patient also happens to be a non-emancipated minor. In most states in the US, physicians are obligated to treat minors for life threatening conditions despite religious beliefs of the parents or child.

Intraoperatively, the techniques such as acute normovolemic hemodilution, cell-saver scavenging devices as a form of autotransfusion, hypotensive anesthesia, and deliberate hypothermia can all be used. The surgeon must also pay scrupulous attention to minimizing operative blood loss and securing hemostasis.

Acute normovolemic hemodilution (ANH) is a method that reduces, or may even eliminate, the need for blood transfusion during surgery. In addition, this technique has been used in surgeries in which the patient is difficult to cross match or for those patients who wish to avoid the inherent risks sometimes associated with blood transfusion such as hepatitis or AIDS. ANH should be done prior to surgical incision since surgical blood loss during hemodilution may result in acute hypovolemia. Remember, with a reduced amount of hemoglobin and subsequent decrease in oxygen availability, it is important to monitor tissue perfusion, the hematocrit (HCT), and the volume status of the patient throughout surgery.
The amount of blood to be removed during hemodilution is determined by the following formula:

\[ V = \frac{EBV \times (HCT_i - HCT_f)}{HCT_{av}} \]

where \( V \) = amount of blood removed, \( EBV \) = estimated blood volume, \( HCT_i \) = initial hematocrit, \( HCT_f \) = final hematocrit, and \( HCT_{av} \) = average of \( HCT_i \) and \( HCT_f \). [9]

Moderate hemodilution would be the instance where \( HCT_f \) is in the range of 20 to 25%. With hemodilution, the objective is that any blood lost during the procedure would contain fewer red blood cells. Although hemodilution reduces the oxygen carrying capacity of blood, oxygen transport to the tissues may be maintained if cardiac output remains adequate. [9]

Replacement fluid can be either crystalloid or a colloid. If crystalloid is used, replacement is done using a 3 to 1 ratio of crystalloid to blood. If colloid is used, a ratio of 1 to 1 is usually sufficient. A second alternative to transfusion is the use of cell-saver scavenging devices. These devices aspirate blood from the patient, filtrate or centrifuge it, and reinfuse the processed red blood cells. Depending on the patient, this may satisfy the condition of a continuous circuit of blood collection. Complications associated with the use of these devices include coagulation disturbances and hemolytic reactions. [9]

Relative contraindications to cell salvage techniques include:

1. use of clotting agents (surgical, gelfoam), methylmethacrylate, and irrigating solutions (betadine, topical antibiotics);

2. contaminants such as urine, bone, fat, bowel contents, infection, and amniotic fluid.
   a. case reports exist that suggest transfusion of cell salvage blood contaminated with bowel contents and especially amniotic fluid may be safer than previously thought. [14]

3. Malignancy
   a. It has been estimated that only 0.01%-0.000001% of circulating tumor cells have the potential to form metastatic lesions[15];

4. Sickle cell, thalassaemia;

5. Catecholamines (pheochromocytoma), oxymetazoline (Afrin), papaverine
   a. Blood pressure affects have been seen following reinfusion of blood tainted with vasoactive agents (both endogenous and exogenous) [16]
Anesthetic Considerations and evoked potentials:
During surgical procedures on the central nervous system (CNS) that have potential risk of damage to the spinal cord or brain the surgeon may elect to monitor evoked potentials. With the use of intra-operative neurophysiology monitoring (IONM), consisting of monitoring of somatosensory evoked potentials (SEPs) and motor evoked potentials (MEPs), the integrity of neural pathways can be monitored without requiring a patient's cooperation while under anesthesia.

SSEP:
SSEPs responses are very low in amplitude and therefore require prolonged averaging. It may take 3-5 min to determine a significant change depending on the ambient noise level. A decrease in amplitude of 50% or increase in latency of 10% while monitoring SSEPs can be indicative of injury to the large fiber dorsal column pathways, assuming these changes are not caused by anesthetics or temperature changes. A decrease in amplitude or increase in latency of SSEP’s can be caused by volatile anesthetics, nitrous oxide, barbiturates, propofol and opiates. If volatile anesthetic must be used then nitrous oxide should be avoided and a MAC of <1 maintained to minimize effects seen on evoked potentials. Ketamine and etomidate cause increased amplitude of SSEPs, while benzodiazepines, dexmedetomidine and neuromuscular blockers have no effect.

MEP:
Usually an increase of more than 100 V in the threshold for obtaining a muscle MEP is considered an early sign of injury. However, thresholds often increase gradually during surgery and are significantly influenced by even small changes in anesthesia. MEPs are exquisitely sensitive to most agents in use in the operating suite, with the exception of opioids. For that reason, anesthetics that avoid large boluses of medications are ideal. If a bolus is needed however, communication with the technician should occur.

In some patients, the threshold of anesthesia effects on cortical sensory and peripheral myogenic responses may make any amount of inhalational agent unacceptable during IONM. In these cases total intravenous anesthesia (TIVA) is needed. This is especially common if a patient has a compromised neural pathway from pathology or age (e.g., patients aged below 3 years). In the latter cases, an amplitude enhancing agent (like ketamine or etomidate) may be infused to increase MEPs and reduce those components of TIVA which are known depressants of MEPs.

Non pharmacologic factors affecting evoked potentials:
1. Blood pressure, spinal cord perfusion and cerebral perfusion pressures- decreased amplitude and increased latency
2. Moderate to severe hypoxemia- increased latency, decreased amplitude
3. Increased ICP or uncal herniation

4. Anemia

5. Hypocapnia (20-25mmHg) decrease latency and increases amplitude

6. Hypercapnia (>100mgHg) increase latency and decrease amplitude

7. Hypothermia (<35 °C) increased latency, unpredictable amplitude

Regional temperature changes can also alter evoked responses that would not be otherwise predicted based on unchanged core temperature. Irrigation of spinal cord, brainstem, etc., with cold saline causes alterations in evoked responses. In addition, limb cooling (from cold infusion of fluids) can change the SSEP originating a nerve in that extremity. [13]

Extubating the difficult airway patient

According to the American Society of Anesthesiologists there remains insufficient evidence for the benefits of one extubation strategy in comparison to another in the difficult airway patient. However, a pre-formulated extubation plan should be considered.

The ASA Practice Guidelines for Management of the Difficult Airway 2003 suggest: “The preformulated extubation strategy should include

1. A consideration of the relative merits of awake extubation versus extubation before the return of consciousness.

2. An evaluation for general clinical factors that may produce an adverse impact on ventilation after the patient has been extubated.

3. The formulation of an airway management plan that can be implemented if the patient is not able to maintain adequate ventilation after extubation.

4. A consideration of the short-term use of a device that can serve as a guide for expedited reintubation.” [19]

The ASA Task Force on Management of the Difficult Airway, recommends using an airway exchange catheter (AEC) to facilitate reintubation as needed. The AEC remains in place until the patient has demonstrated their ability to maintain effective ventilation and the clinician is comfortable with its removal. The AEC is a semi-rigid, hollow device inserted through the endotracheal tube and fixed at the level just above the carina. In emergent circumstances, the hollow core can serve as a means of
Complications of Reintubation

<table>
<thead>
<tr>
<th>AEC Present (n=51)</th>
<th>AEC Absent (n=36)</th>
<th>P</th>
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<tbody>
<tr>
<td>First Pass success for reintubation</td>
<td>87%</td>
<td>14%</td>
</tr>
<tr>
<td>Hypoxemia during reintubation (SPO2&lt;90%)</td>
<td>8%</td>
<td>50%</td>
</tr>
<tr>
<td>Severe Hypoxemia during reintubation (SPO2&lt;70%)</td>
<td>6%</td>
<td>19%</td>
</tr>
<tr>
<td>Bradycardia (HR&lt;40) with hypotension</td>
<td>4%</td>
<td>14%</td>
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<tr>
<td>Multiple intubation attempts (&gt;3) including placement of accessory airway device</td>
<td>10%</td>
<td>77%</td>
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<tr>
<td>Esophageal intubation</td>
<td>0%</td>
<td>18%</td>
</tr>
<tr>
<td>Rescue Airway device/technique</td>
<td>6%</td>
<td>90%</td>
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Other considerations:
1. Assess the amount of secretions. (Patients classified as producing no or mild secretions were 96% more likely to have successful extubation)
2. Assess cough strength.
3. Perform cuff leak test to help rule out upper airway edema. Consider use of steroids to prevent/treat laryngeal edema.
4. Consider head up positioning for extubation to facilitate diaphragm movement.
5. Although preoxygenation is routinely performed in the difficult airway patient, in the patient with SMA it should only be done after the patient has demonstrated the ability to maintain near baseline saturations on their typical vent settings.
6. Minimize stimulation and activation of airway reflexes during emergence (lidocaine, remifentanil, dexmedetomidine) [20]

References
[1] Anesthesia and spinal muscle atrophy. Gunilla Islander. Department of Intensive and Perioperative Care, Skane University Hospital, Lund, Sweden
[4] Patients with severe muscle wasting are prone to develop hypoglycemia during fasting. Ørngreen MC, Zacho M, Hebert A Neurology, 2003
[10] Anesthetic Challenges and Considerations Presented by the Jehovah's Witness Patient. Anna L. Harris, M.D. and Thomas P. Engel, M.D. Department of Anesthesiology, Loma Linda University School of Medicine, Loma Linda, California. http://anestit.unipa.it/gta/JehovahsWitness.html