20 y/o male with history of Emery-Dreifuss muscular dystrophy presenting for bilateral heel cord lengthening and right mid-foot osteotomy with short leg cast application.

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Objectives:
- Discuss perioperative evaluation, preparation and anesthetic implications of Emery-Dreifuss muscular dystrophy.
- Describe risks and benefits of different modalities of postoperative pain control for patients undergoing orthopedic surgery involving extremities.
- Recognize the pathophysiology and manifestations of tourniquet mediated morbidity.
- Discuss the evidence informing current guidelines for intraoperative tourniquet use.

Case history:
A 20 year old, 50 kg male with complex medical history including Emery-Dreifuss muscular dystrophy, cardiomyopathy, atrial fibrillation/flutter, complete heart block and restrictive lung disease presenting for bilateral heel cord lengthening, right mid-foot osteotomy and short leg cast application.

Questions:
What is Emery-Dreifuss muscular dystrophy (EDMD)? What is the inheritance pattern of EDMD? What is the natural progression of EDMD? What are the anesthetic implications of EDMD?

Case history continued:
The patient's other comorbidities included a history of cardiomyopathy, atrial fibrillation for which the patient was anticoagulated with warfarin, sick sinus syndrome (pacemaker placement October 2006), restrictive lung disease and scoliosis s/p posterior fusion. The patient had developed bilateral Achilles tendon contractures and a right rigid foot, rendering him wheelchair dependent and the hope with the surgery was to help him become once again ambulatory. The patient also had a history of adverse reactions to various opioids: morphine (rash), hydromorphone (severe nausea) and methadone (respiratory depression).

Questions:
What preoperative information would you like to know? Do you require consultation with any specialists prior to proceeding with the case?

Preoperative studies:
A preoperative echocardiogram revealed mild left ventricular dilation and dysfunction.

What is your plan for anesthetic induction, maintenance and postoperative recovery? Is a clean technique required for this case? Beyond standard American Society of Anesthesia recommended monitors, are additional monitoring modalities warranted? What are the different

options for postoperative analgesia? How would you manage his pacemaker and anticoagulation perioperatively?

Intraoperative course:
- IV induction with propofol/rocuronium/fentanyl
- Uneventful endotracheal intubation
- Adequate IV access
- Arterial blood pressure as well as Bispectral Index (BIS) monit oring
- Anesthetic maintenance with sevoflurane/isoflurane, minimum alveolar concentration (MAC) 0.5-0.9 and ropivacaine 0.1% at 10ml/hour via a right popliteal catheter
- dopamine 2-5 mcg/kg/min along with multiple doses of phenylephrine to maintain mean arterial pressure (MAP) > 60 mmHg

After an initial tourniquet time of 127 minutes, the tourniquet was deflated with the intent of reinflating it shortly after.

Questions:
What are the maximum recommended limits of tourniquet time? What are your concerns related to prolonged tourniquet use? If additional tourniquet time is needed, what actions can be taken to mitigate morbidity?

Intraoperative course continued:
-After a deflation time of 14 minutes, the tourniquet was reinflated for an additional 175 minutes, for a total tourniquet time of 302 minutes. The total anesthesia time was 9 hours and 26 minutes.

Postoperative course:
Following an uneventful PACU recovery, the patient was admitted to the cardiac medical service for management of his anti-coagulation post-operatively. The patient was clinically stable on the floor on postoperative day 1 (POD 1), and the popliteal catheter was continued, with pain scores of 0-2/10. However during the evening of POD 1, the patient reported 12/10 pain that resolved with removal of the upper portion of his bivalved cast and limb elevation.

Questions:
How would you evaluate and treat his pain?

Postoperative course continued:
Postoperative day 2
- 9.00 AM removal of popliteal catheter.
- Throughout remainder of the day, patient received oxycodone 10 mg and diazepam 2.5 mg iv.
- In the late evening, toes of the right foot appeared pale and cool, with a diminished dorsalis pedis artery pulse (DPA).
- The lower portion of the cast was removed with the patient subsequently developing significant pain.
- The foot appeared bruised, slight blotchy pink color and mottled though capillary refill and sensory/motor examination all remained normal.
- Dopper ultrasound showed diminished blood flow through DPA but intact flow through posterior tibial artery (PTA).

Questions:
How should this patient be managed at this point? What additional information related to this patient's postoperative would you like to know? Are additional studies and consultations needed? What is the role of regional anesthesia in patients at risk for acute compartment syndrome?

Postoperative course continued:
A computed tomography scan of the arteries (CTA) was performed did not reveal any identifiable thrombus and the PTA was found to be extremely narrow in caliber from the mid-calf with short segments of extremely poor perfusion with brief reconstitution at the medial malleolus and no flow seen distally in the plantar foot. The patient was evaluated by vascular surgery upon return to the floor, and physical exam was concerning for late compartment syndrome. The patient was transferred to the adjacent adult hospital emergently for surgical intervention. Findings during the surgery were not consistent with compartment syndrome; the patient's calf was not really swollen and the patient's calf muscle, tissues were all viable. Additionally, no arterial lacerations could be identified. A popliteal arteriogram was performed that showed "vessels petered out mid-calf." A balloon embolectomy was attempted without success; no clot was identified and there was no improvement in vascular flow. A right foot fasciotomy was also performed without significant findings. Following the fasciotomy, there was no subsequent improvement in forefoot perfusion following the procedure and the patient eventually required a below the knee amputation.

Questions:
What is the pathophysiology of reperfusion injury? Did the patient have comorbidities that predisposed him to complications from tourniquet use? How does compartment syndrome present?

Discussion:
Emery-Dreifuss muscular dystrophy is an x-linked (EDMD1) or autosomal dominant (EDMD2) disorder as a result of mutations in the emerin, lamin A/C or LINC complexes. The disease pattern is generally that of early muscle contractures, progressive muscle weakness, cardiomyopathy and issues with cardiac conduction. Patients commonly present initially with Achilles, elbow and postcervical muscle contractures with later development of proximal muscle weakness. During the second or third decade of life, patients may go on to develop cardiomyopathy and cardiac abnormalities such as ECG PR interval prolongation and complete heart block. Though EDMD patients have not been reported to have an increased risk of malignant hyperthermia, anesthetic considerations include difficult tracheal intubation and neuraxial anesthesia due to limitations in joint mobility due to contractures, issues with cardiac conduction and management of associated pacemakers and defibrillators, dilated cardiomyopathy, delayed gastric emtpying, gastric reflux and risk of anesthetic induced rhabdomyolysis. Succinylcholine is contraindicated due to concerns for hyperkalemia following administration. Due to the possible cardiac conduction issues and cardiomyopathy, strong consideration should be given to invasive hemodynamic monitoring such as an arterial line, which would have the additional advantage of facilitating ready assessment of metabolic status.

The anesthesia care team elected to place a popliteal catheter for intraoperative analgesia with the goal of lessoning hemodynamic compromise from high dose volatile anesthetics given the patient's history of complete heart block and cardiomyopathy. This patient also had existing restrictive lung disease and a history of adverse side effects from opioids and postoperatively, the popliteal catheter was to be utilized to minimize the potential for respiratory compromise from systemic opioids.

The use of regional anesthesia in the setting of surgeries with increased risk of acute compartment syndrome (ACS) remains controversial. There are several adult case reports inferring that central or peripheral nerve block may have delayed the appropriate diagnosis of ACS. In all of these case reports, the regional block did result in dense sensory and motor block that masked symptoms of ACS. However, in the pediatric literature, 12 cases of ACS in the setting of regional anesthesia have been reported without clear evidence that the regional anesthetic contributed to a delay in diagnosis. There are multiple case reports detailing the timely diagnosis of ACS in the setting of continuous peripheral nerve block leading to the conclusion that regional anesthesia can be used safely used without risk of masking ACS if the following precautions are adhered to: use of ultrasound-guided regional anesthesia, low concentration of local anesthetic avoiding dense sensory and motor blockade and prompt evaluation of patients for ACS if there is new onset pain despite previously adequate pain control not controlled by PO adjuncts, especially when accompanied by recurring motor block.

While the maximum recommended tourniquet time is 2 hours, this patient had a total tourniquet time of over 5 hours. Additionally, patients who have poor cardiopulmonary or renal reserve, such as this patient, are at increased risk of tourniquet mediated tissue injury. Because of its vasodilating properties, regional anesthesia has been recommended to mitigate the risk of tourniquet induced injury. Even in healthy subjects without known neuromuscular disease, 4 hours of ischemic time has been found to be the limits of tissue viability for skeletal muscle. Though tissue ischemia from tourniquet compression causes an initial insult to the affected tissue, it is the reperfusion of the tissue that causes the most significant damage and it is likely that the resultant injury was the major contributor to morbidity in this patient. Reperfusion injury is mediated by free radicals, activated polymorphonuclear leukocytes, nitric oxide, cytokins and arachidonic acid metabolites. Clinically, reperfusion injury can lead to metabolic acidosis, hyperkalemia, myoglobinemia, myoglobinuria and possibly renal failure.

In summary, EDMD patients have a myriad of anesthetic issues that require consideration in order to optimize their perioperative outcome. Patients who have poor cardiopulmonary or renal reserve are especially prone to complications from prolonged tourniquet use. While tourniquet ischemia causes the initial tissue insult, it is the reperfusion injury that causes the most damage to tissue. Regional anesthesia can potentially mitigate tourniquet complications and under the correct clinical circumstances should not delay diagnosis and treatment of ACS.

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