Anesthetic Management for Extensive Spine Surgery in a Child with Mitochondrial Disease

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Objectives:

1- Understand the elements of the anesthetic plan for a child scheduled for extensive spine surgery
2- Discuss preoperative evaluation, preparation and anesthetic implications associated with mitochondrial disease
3- Be familiar with the effects of different anesthetic agents on different modalities of intraoperative neurophysiologic monitoring
4- Devise a strategy for anesthetic management when the neurophysiologic signals are lost intraoperatively
5- Describe appropriate postoperative consultation and care for the patient with injuries related to the procedure

Case History: A 13 year-old boy with idiopathic scoliosis and mitochondrial disease presents for anterior thoracic vertebrectomy and posterior spine decompression/fusion.

Questions:

Preoperative evaluation: Cobb angle is 70 degrees, neurologic exam is without deficits. EKG shows a prolonged QT interval of 490.

Questions: What are the risks and possible complications of a prolonged QT interval? How will you address this with the parents? What further workup would you request? Will PFTs help? Should he have an echo? What labs are appropriate preoperatively? How will you minimize the potential for intraoperative acidosis and metabolic decompensation?

Anesthetic Plan:

Questions: How will you isolate the lung for the anterior portion of the case? What impact do mitochondrial disease and neuromonitoring have on your options for intraoperative management? What monitors will you use intraoperatively? Would you place a central line and/or arterial line? How do various anesthetic agents affect SSEPs and MEPs? What agents should be avoided in mitochondrial disease? What options exist for post-operative pain control for this patient?

Intraoperative Care: --

Questions: What is your response to a significant decrease in the MEPs after decompression maneuvers? What is your transfusion threshold? What means (equipment, medications) will you use to minimize the need for transfusion? When is controlled
hypotension an acceptable technique? What could cause difficulty with hemostasis in the prone position? What is the differential diagnosis for sudden severe hypotension and tachyarrhythmia? Can you resuscitate in the prone position?

Postoperative Care: --

Questions: The tongue was noted to be lacerated post-operatively; how do you approach this complication? When do you call risk management? How will you explain this to the parents?

Discussion:

Severe scoliosis (Cobb angle greater than 45 degrees) is generally treated surgically. Many aspects of perioperative care need to be addressed ahead of time, including positioning, blood loss, pain control, and monitoring of hemodynamics and neurologic pathways. Techniques to decrease and treat blood loss include autologous pre-donation, cell salvage, isovolemic hemodilution, controlled hypotension, use of antifibrinolytic drugs and red cell transfusion. Severe scoliosis often causes restrictive lung disease, and preoperative assessment of respiratory function is necessary especially if lung isolation for an anterior procedure is planned. Risk factors for severe pulmonary impairment are a structural cephalad thoracic curve, a major thoracic curve spanning eight or more vertebral levels, or thoracic hypokyphosis (6). Intraoperative monitoring of somatosensory and motor-evoked potentials (SSEP and MEP) is becoming widespread as a means to prevent permanent postoperative neurologic deficit as a result of surgical correction of scoliosis. Neuromonitoring requires the avoidance (or limited use) of several anesthetic agents, most importantly the volatile agents as they cause a dose-dependent decrease in amplitude and increase in latency of the signals; neuromuscular blocking agents should be avoided if possible or at least monitored carefully during monitoring of MEPs.

A specific plan should be in place in the event that neuromonitoring signals are lost during spinal manipulation. This generally includes ruling out artifact (equipment malfunction or bolus dose of medication), evaluation of recent maneuvers by the surgeon, and increasing mean arterial pressure to greater than 90mmHg. If these fail to resolve the lost signal, and injury protocol should be in place which includes IV methylprednisolone and preparation for a wake-up test.

Mitochondrial disease encompasses many different diseases, all of which can have varying clinical presentations and severity depending on gene expression. Mitochondria are responsible for cellular respiration and making energy available for metabolic processes; thus mitochondrial diseases generally manifest in organs with the highest energy needs (brain, muscle, heart, liver and kidney). Patients with mitochondrial disease are more sensitive to the effects of stress, illness and preoperative fasting, and can develop a lactic acidosis and metabolic encephalopathy in the perioperative period. Barbiturates, etomidate, succinylcholine and propofol infusion are not recommended, and Lactated Ringers solution is generally avoided as well. Avoidance of situations that increase oxygen
demand and thus stress the mitochondria (i.e., hypoxia, hypercarbia, hypothermia, tachycardia) seems prudent.

A patient who sustains injury related to the surgical procedure requires careful consideration. Appropriate services to assess and manage the complication should be consulted, and risk management should be notified early in the process. Open communication with the patient and family is recommended. The incidence of neurologic impairment as a result of spine surgery is approximately 0.3% (all partial cord lesions). Risk factors for neurologic impairment are: curve more than 100 degrees, congenital scoliosis, kyphosis, and post-radiation deformity.

References: