Cesarean Section Delivery with Long QT Syndrome
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Keywords: Cesarean section, long QT syndrome, complex cesarean section, QT prolongation

Birth by cesarean section is performed over 1.5 million times annually in the United States and accounts for 30% of all deliveries. The volume and complexity of patients whom anesthesia providers encounter have continued to grow. It is essential for anesthesia providers to understand and integrate changes related to physiology and anatomy in patients is essential to creating a safe and effective anesthetic plan.

Case Report

A 35-year old female Gravida 2 Para 1001, 39 weeks gestation, 167 cm tall and 83 kg, presented for a cesarean section secondary to prior complex cesarean delivery. Her medical history was significant for long QT syndrome, ventricular arrhythmia, family history of sudden cardiac death, esophageal reflux, irritable bowel syndrome, duodenitis, renal calculi, and pelvic hematoma. Her current medications consisted of nadolol 20mg, ranitidine 150mg and prenatal vitamins. Her surgical history included a colonoscopy, sigmoidoscopy, esophagogastrroduodenoscopy, cytourethroscopy with ureteroscopy and pyeloscopy and prior cesarean section, none of which were associated with anesthetic complications.

Pre-operative evaluation testing consisted of a standard 12-lead electrocardiogram and serum electrolyte values. Documented electrolyte findings included a serum sodium 135mEq/L, potassium 3.7mg/dl and magnesium 1.7mg/dl.

A preoperative airway evaluation revealed a Mallampati classification of II, a thyromental distance of greater than 6 cm, an estimated oral opening of 4 cm, intact dentition, and full range of motion in her neck.

In the operating room standard monitors per the American Society of Anesthetiologist’s were applied. Nasal cannula was initiated utilizing an oxygen flow of 4L/min. Continuous ECG was monitored in leads II, V. Neuraxial anesthesia was provided via an existing epidural in which placement was confirmed. Prior to the administration of 45-mg Lidocaine 1.5% with epinephrine 1:200,000, epidural lidocaine 2% methylparaben free 200 mg with the addition of 8.4% Sodium Bicarbonate 2 mL was given upon the patient arriving to the operating room. The patient was placed supine with left uterine displacement using a 20-cm-high wedge.

The anesthesia provider evaluated the patient’s analgesia level and degree of motor blockade by temperature differentiation. Surgical anesthesia and analgesia were then
confirmed with an Allis test performed by the proceduralist prior to abdominal incision. Cefazolin 2g intravenously was administered after the surgical timeout and prior to incision. Continual evaluation of analgesia and motor blockade of the epidural resulted in a subsequent bolus in the epidural of methylparaben free lidocaine 2% 100 mg.

Upon delivery of the fetus and progression to the third stage of labor, intravenous oxytocin 5units in Lactated Ringers 1,000mL bolus was administered concurrently with misoprostol 800mg buccally. The surgical procedure concluded without event and preservative free epidural morphine 5mg was administered. The patient was transferred to post-partum where telemetry and electrolyte monitoring continued.

Discussion

Anesthetic considerations for birth by cesarean section are multifactorial and must take into consideration the maternal status, the urgency of the delivery, the health of the fetus, and the patient desire for a safe and effective anesthetic. Obstetrical anesthesia provides a continuum of options throughout the stages of labor for integration of pharmacologic, physiologic and anatomical knowledge. The complex cesarean delivery of a parturient with long QT syndrome allows for a thoughtful progression of anesthesia throughout labor.

Long QT syndrome (LQTS) is defined as a prolongation of the QT interval observed on an electrocardiogram. LQTS may manifest as arrhythmias with episodes of fainting, seizures, cardiac arrest and even death. Although there are detailed collections of medications that may lead to QT prolongation, the syndrome is most frequently related to a genetic mutation of cardiac-expressed ion channels. Mutations in cardiac-expressed voltage gated potassium channel genes (KCNQ1 or KCNH2) account for approximately 90 percent of the mutation-based long QT syndromes, with the remainder of mutations identified as voltage-gated sodium channel gene SCN5A and/or calcium channel gene CACNA1C. Genetic testing is available for long QT syndrome and is used in conjunction with echocardiography for evaluation of the structure and function of the heart.

Patients identified with long QT syndrome should be educated to avoid medications that may prolong their QT interval and/or reduce their serum levels of potassium or magnesium. Electrocardiogram and electrolyte monitoring, along with beta-blockers have been established as the initial treatment methodology for reduction of cardiac arrhythmia risk in all patients. In relation to the obstetric patient best practice recommendations include: documentation and evaluation of the electrocardiogram corrected for changes in heart rate (QTc), baseline QTc <500ms and electrolyte monitoring for a magnesium level >2mg/dl and potassium >4mg/dl.

Regarding analgesia and anesthesia, neuraxial anesthesia via epidural catheter placement can be utilized with great success in this patient population. Using Bupivacaine, an amino-amide local anesthetic with a relatively long duration of action and the ability to produce a differential block, has been used without adverse consequences in pregnant
patients with long QT syndrome. In the event that general anesthesia is required, the combination of intravenous propofol, oxytocin and alfentanil for analgesia has been shown to block the slow component of the potassium rectifier channel and therefore remains a safer choice than the use of sevoflurane.

Upon delivery of the fetus and placenta, oxytocin is routinely administered intravenously to mimic normal pre-partum contractions. The oxytocin receptors on the myometrium contract the uterine smooth muscle by increasing intracellular calcium concentrations. Oxytocin poses a risk of prolonging the QT interval, although data suggests the average prolongation may not be more than 40ms (+/- 20ms), in patients receiving 10Unit IV boluses. A reduced dose of oxytocin, 5Units per 1,000mL lactated ringers has been shown to not prolong the QT interval, while still effectively decreasing post partum hemorrhage.

Misoprostol is a prostaglandin E1 analogue that is rapidly absorbed into an active metabolite, misoprostol acid, which has been used to treat and possibly prevent postpartum hemorrhage. Although currently the Food and Drug Administration does not endorse the use of Misoprostol in the obstetrical and gynecological population, numerous studies have been conducted to evaluate the uterotonic efficacy of misoprostol. Administration of buccal misoprostol 600-1000mg followed by 200mg every six hours for the first 24 hours post-operatively has been suggested as both an adjunct to oxytocin and methylergonovine, as well as an effective agent in instances of hemorrhage refractory to the previous medications.

Recent findings for post-partum women with long QT syndrome, suggest the 9-month postpartum interval is associated with a 2.7-fold increased risk of experiencing a cardiac event and a 4.1-fold increased risk of experiencing a life-threatening event when compared to the preconception time period. Estrogen and progesterone levels are high during pregnancy and decrease well below normal levels when the mother breastfeeds. It is hypothesized that this hormonal fluctuation could influence the adrenergic response of the mutant ion channels in long QT syndrome. The possibility of deprivation of estrogen, such as with post-partum breastfeeding, may increase the adrenergic activity and cardiac myocyte excitability in the post partum period, contributing a higher risk of post-partum cardiac events. It is advisable to seek consultation with cardiology services should the patient experience any palpitations in the days to weeks during the post-partum period.

It is necessary for anesthesia providers to review a baseline echocardiogram and electrolyte levels, at a minimum, before proceeding with an anesthetic plan. After completion of the operative course with telemetry, the continuation of any prior beta-blockade regimen should be initiated. Limited case reports and studies evaluating the risk of pregnancy in patients with long QT syndrome led to a cautious and measured approach to this uneventful and successful delivery.

References


Mentor: Kelly Wiltse Nicely CRNA, PhD