

Anesthetic Management of Familial Hypokalemic Periodic Paralysis During Parturition

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Familial hypokalemic periodic paralysis (FHPP) is a rare inherited disease characterized by attacks of severe muscle weakness (1,2) and flaccid muscle paralysis (2,3). Menstruation (4), pregnancy (2), and anesthesia (3,5–7) have been reported to exacerbate FHPP. Anesthetic management during parturition has not been previously described.

Case Report

An 18-yr-old nulliparous woman with a previous diagnosis of FHPP presented for routine obstetric care at 12 wk gestation. She had a positive insulin-glucose provocative test with subsequent genetic analysis revealing the classic FHPP mutation. She reported that several relatives had FHPP, including a history of ventricular arrhythmias coincident with paralytic episodes. Our patient reported onset of symptoms at the age of 12 yr, with attacks occurring one to three times per month. She described large carbohydrate-rich meals, emotional stress, and exercise as precipitating factors. Weakness would typically last 1–3 days. At the first sign of an attack, she would begin potassium supplementation (approximately 120 mEq/d). She did not have a history of respiratory failure or involvement of oral-pharyngeal muscles. She also noted mild asthma and that bronchodilators worsened her FHPP.

On consultation at 34 wk gestation, her cardiopulmonary and neurologic examinations were normal, with the exception of a slight unilateral ptosis. She reported few paralytic attacks during her first trimester, with a return to one to three attacks per month since that time. She voiced her concern that stress during parturition, labor pain, and expulsive efforts during the second stage of labor might combine to result in a paralytic episode.

An interdisciplinary management plan was developed and included continuous IV KCl supplementation during labor and postpartum, avoidance of IV glucose, insertion of an arterial catheter for frequent assessment of maternal pH and potassium concentration, early regional anesthesia, and outlet forceps delivery after passive descent of the fetus during the second stage of labor.

The patient presented in labor at 40 wk gestation. The cervix was 4 cm dilated and completely effaced. Fetal heart rate assessment was reassuring. Continuous pulse oximetry and electrocardiogram monitoring were initiated. IV and radial arterial catheters were inserted. Lactated Ringer's solution with an additional 40 mEq KCl/L was infused at 200 mL/h. Four hours after admission the patient that reported her contractions were becoming painful. Her cervical examination was 5 cm dilation, 100% effacement, 0 station. An 18-gauge Tuohy epidural needle was inserted in the L3-4 interspace without difficulty. After identification of the epidural space, a 26-gauge Gertie Marx needle was inserted via the epidural needle, and 10 µg of sufentanil and 2.5 mg of bupivacaine were injected intrathecally. After removal of the spinal needle, a 20-gauge epidural catheter was inserted. An epidural infusion of 0.0625% bupivacaine with 2 µg/mL fentanyl was initiated at 12 mL/h. The patient reported immediate profound analgesia.

Six hours after admission, the patient reported onset of perineal pressure. Cervical examination revealed complete dilation and +3 fetal station. An additional bolus of epidural local anesthetic (bupivacaine 0.25%, 10 mL in divided doses) was administered. Outlet forceps were applied, and a vigorous male fetus weighing 3780 g was delivered with Apgar scores of 9 and 9 at 1 and 5 min after delivery. Potassium chloride supplementation was continued for 8 h after delivery.

The patient experienced no overt episodes of paralysis. She briefly reported mild weakness when she was noted to have complete cervical dilation. Serum potassium concentrations varied between 3.6 and 4.1 mEq/dL during her labor through the first 6 h postpartum. The patient displayed a mild respiratory alkalosis during labor, with arterial pH ranging from 7.41 to 7.44 and Pco₂ ranges from 26 to 30 mm Hg.

The patient and her baby were discharged home 30 h after delivery. At a follow-up clinic visit 2 wk postpartum, the patient reported two episodes of mild weakness since discharge, both of which responded to oral potassium supplementation.

Discussion

This report is the first to describe the anesthetic management of a parturient with FHPP. The familial periodic paralyses are a group of disorders involving

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muscle weakness and skeletal muscle ion channel mutations(8). Although they share some phenotypic characteristics, hypokalemic and hyperkalemic periodic paralysis differ in the electrolyte alteration associated with weakness, the ion channel mutation responsible for the condition, and the treatment (8-13).

FHPP is an autosomal dominant inherited condition characterized by intermittent attacks of proximal muscle weakness and flaccid paralysis. Severe perioperative attacks have necessitated urgent intubation and controlled ventilation (3). Rarely have complete respiratory arrest and death resulted (11).

The clinical management of FHPP (8) includes avoiding known triggers of attacks, such as psychological stress, surgery, and anesthesia. Medications known to cause intracellular shift of potassium (e.g., β -agonists) may also provoke paralysis.

Because of the rarity of the condition, perinatal experience with FHPP is limited (14,15). General anesthesia, postoperative stress, glucose-containing IV solutions, and long-acting neuromuscular blockers are associated with postoperative paralytic episodes.

The major goals of our management plan were avoidance of carbohydrate loads, administration of IV potassium supplementation, and avoidance of maternal hyperventilation during labor. Epidural analgesia minimizes pain-induced hyperventilation and lowers maternal serum catecholamines, both of which may prevent decreases in serum potassium levels (16-18). The epidural analgesia technique chosen was designed to minimize muscle weakness associated with larger doses of local anesthetics. We chose to avoid an epinephrine-containing local anesthetics because of the known risk of precipitating hypokalemia with sympathomimetics(19). Had uterine hyperstimulation occurred after intrathecal analgesia, we planned on administering IV nitroglycerin (instead of terbutaline) for uterine relaxation. Passive descent of the fetus during the second stage with elective outlet forceps delivery was chosen to avoid the need for active maternal expulsive efforts.

In summary, we report the management of a parturient with FHPP. Avoidance of IV glucose, potassium supplementation, early epidural analgesia, and a passive second stage of labor may have aided in the prevention of paralytic episodes.

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