



# Analgesia and Anaesthesia Management of Labour and Caesarean Delivery for a Parturient with Paramyotonia Congenita

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## Abstract

Anaesthetic management in paramyotonia congenita (PC) or 'paradoxical myotonia' poses perioperative challenges to the anaesthesiologists both in obstetric and non-obstetric surgical patients. There are only a few case reports on the anaesthesia management particularly in the obstetric population. Therefore, we aimed to present the management of analgesia of labour and emergency caesarean delivery for a 26-year-old parturient with PC.

**Keywords:** Caesarean delivery, labour analgesia, myotonia, paramyotonia congenita, spinal anaesthesia

## Introduction

Paramyotonia congenita (PC) is classified as a non-dystrophic muscle disease with a prevalence of approximately 0.17/100000 (1). This abnormality leads to extreme muscle stiffness due to cold exposure. Eulenburg was the first to describe PC in 1886 when he investigated a German family with a syndrome showing episodic muscle cramps and paralysis profoundly exacerbated by cold and exercise (2). Patients with paramyotonia develop muscle stiffness with exercise, whereas typical patients with myotonia suffer symptoms during both rest or after exercise. Myotonia may also be induced by hyper or hypokalaemia, starvation, hypothyroidism and pregnancy (3). Because there is a case report of cold-induced abortion in a woman with PC, the need to avoid cold exposure seems to be paramount importance (4). In this case report, we aimed to present the analgesia and anaesthesia management of a term parturient with PC.

## Case Presentation

A 26-year-old primigravida with PC was admitted at 38 weeks of gestation due to spontaneous rupture of membranes. According to her history, she had muscle cramps triggered by exposure to cold air or water, consuming cold food and physical or emotional exertion since the age of 3 years. Consultation with a neurologist confirmed the diagnosis of PC via genetic studies. She denied a history of miscarriages. She was on regular follow-up by neurologists and was still receiving carbamazepine. Pre-delivery laboratory results showed only mild anaemia with a haemoglobin level of 11 g dL<sup>-1</sup> without remarkable changes in serum electrolytes. After obtaining consent of the parturient who requested systemic analgesia for labour initially, intravenous (IV) patient-controlled analgesia (PCA) using 20 µg of fentanyl bolus with a 5-minute (min) lock-out time was initiated. Soon after augmentation of labour, due to the insufficient pain relief, IV-PCA was converted to epidural analgesia. Maternal heart rate, blood pressure and temperature were monitored which were stable both prior and after induction of epidural labour analgesia. Foetal heart rate (FHR) was monitored continuously. IV fluid warmers, pre-warmed drapes and disinfection solutions

were prepared beforehand. Warm isotonic 0.9% saline infusion was initiated. Epidural analgesia was induced with 15 mL bolus of 0.05% ropivacaine including 2 µg mL<sup>-1</sup> fentanyl and maintained with patient-controlled epidural analgesia set to deliver 10 mL h<sup>-1</sup> infusion and 10 mL bolus with a 10-min lock-out time. FHR tracings showed poor beat-to-beat variability 1 hour later, necessitating discontinuation of oxytocin infusion and subsequent emergency caesarean delivery. After aspiration prophylaxis (IV 50 mg ranitidine, 10 mg metoclopramide and 30 mL sodium citrate), epidural analgesia was extended to surgical anaesthesia by top-up of 10 mL of 2% lidocaine including 1:200,000 adrenaline. Operating room temperature was pre-warmed to 29.4°C. IV fluids were infused via fluid warmers. Sensory block level reached at T4 which was confirmed with pin-prick test. A healthy infant was born followed by oxytocin infusion at 10 U h<sup>-1</sup> 4 minutes after skin incision. The surgery lasted 30 min, and total blood loss was approximately 400 mL. Her body temperature ranged from 37°C to 37.3°C throughout the surgery. Epidural catheter was removed after administering 3 mg epidural morphine for postoperative analgesia in the operating room at the end of the surgery. Additional analgesia was provided with oral diclofenac and paracetamol. She recovered uneventfully and was discharged on postoperative day 3.

## Discussion

In this case report, we presented our analgesia and anaesthesia management for a parturient with PC. PC has been shown to be caused by a mutation in the SCN4A gene on chromosome 17q23 transmitted in an autosomal dominant manner. Exposure to cold leads to spontaneous sarcolemma depolarization and myofibril contraction with slowing in the relaxation phase (5, 6). Avoiding exposure to cold air, adequate relief of labour pain and avoidance of electrolyte imbalance, particularly potassium, ensure safe labour and delivery for patients with PC (5). There are several anticipated complications and/or triggers for myotonia crisis. Obstetric complications include uterine atony and prolonged labour requiring caesarean delivery, whereas anaesthetic complications may result from medications used for general anaesthesia. Intubation using suxamethonium for rapid sequence induction may induce myotonic contracture and masseter spasm leading to failed intubation. Subsequently, chest stiffness may disrupt mechanical ventilation. For possible triggers, there might be an unpredictable sensitivity towards non-depolarizing muscle relaxants leading to postoperative prolonged block which may occur after reversal with neostigmine (7). For that reason, Buzello et al. (8) advocated the use of atracurium or cisatracurium which have spontaneous metabolism and do not require reversal. In general, rocuronium has been commonly used under neuromuscular block monitoring, and sugammadex has been the choice of reversal agent instead of neostigmine

(7). Another trigger hyperkalaemia which is either induced by suxamethonium or its presence can aggravate myotonia. Although the risk of malignant hyperthermia is very low for PC, volatile agents may lead to postoperative shivering and subsequently myotonia (9). Spinal and epidural anaesthesia has been safely used both for caesarean and vaginal deliveries (5, 7). Frossard et al. (10) described the successful management of neuraxial labour analgesia using 0.02% ropivacaine with sufentanil via epidural catheter. Similarly, we preferred regional technique because we had a functioning epidural catheter to extend for surgical anaesthesia. Because both general and regional anaesthesia techniques may pose the risk of disturbing the thermoregulation, it is wise to avoid testing sensory block level with ice and to keep the ambient temperature warm in the labour suite and operating room similar to what we did in the present case. Additionally, providing thermal comfort with hot air blanket should be considered during and after surgery.

## Conclusion

Good pain relief for labour with placement of epidural analgesia followed by successful anaesthesia for emergency caesarean delivery was provided without occurrence of myotonic crisis by avoiding precipitating factors and providing a warm environment aid safe operative delivery.

**Informed Consent:** Written informed consent was obtained from patient who participated in this case.

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**Conflict of Interest:** The authors have no conflicts of interest to declare.

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