Anesthetic Management of an Obstetric Patient With Charcot-Marie-Tooth Disease: A Case Study

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Additional documentation of regional anesthesia in patients with Charcot-Marie-Tooth disease (CMT) is needed to guide practitioners and patients in exploring appropriate options for anesthesia and analgesia management. This case report describes the successful use of a combined spinal-epidural technique for labor progressing to cesarean delivery in a patient with CMT. Previous similar case reports were reviewed and an extensive literature search was conducted to organize the limited body of research regarding use of regional anesthesia in patients with CMT.

Opinions regarding regional anesthesia in patients with neuromuscular diseases such as CMT are often contradictory and based on theory rather than documented practice. This case report confirms what seems to be the developing consensus in anesthesia that regional management is a safe alternative to general anesthesia in these patients. Considering that CMT is among the most common of hereditary neuromuscular diseases, it seems valid to establish a more research-driven recommendation for practice.

Keywords: Charcot-Marie-Tooth disease, epidural, obstetric, spinal.

Charcot-Marie-Tooth disease (CMT) is categorized as an inherited peripheral neuropathic disease leading to distal muscle atrophy, sensory disturbances, and absent reflexes in the extremities. CMT is cited to affect 1 in 2,500 people, representing a relatively large patient population. The nature of the disease process is a progressive demyelination of nerve cells, with symptoms generally manifesting in the teenage years. Of the 3 known subgroups of CMT disease, CMT type 1A (CMT1A) is the most common; similarities between subgroups far outnumber differences. Although research regarding the basis of the nerve derangement and discovery of genetic markers is being expanded, recommendations for anesthesia—regional anesthesia in particular—are limited.

Regional anesthesia in patients with neuromuscular diseases has historically been controversial and even suggested as a risk factor for nerve damage following regional anesthesia. This case report describes the anesthetic management of an obstetric patient with CMT1A that used a combined spinal-epidural technique for labor. The epidural was then successfully used to establish adequate anesthesia for cesarean delivery after fetal complications developed during labor.

Case Summary
A 35-year-old woman, American Society of Anesthesiologists (ASA) physical status 3, gravida 1, para 0, presented at approximately 37 weeks’ gestation and was admitted to the labor and delivery unit for induction of labor. The patient had been admitted 2 weeks earlier secondary to gestational hypertension and was subsequently sent home to delay induction until the 37th week. The patient was 172.7 cm tall and weighed 85.5 kg. The patient denied having any drug allergies. Sensory deficits and lack of reflexes of both lower extremities and contractures of the right hand were noted during the physical examination. Physical findings of the cardiovascular and respiratory systems were unremarkable, and laboratory values were all within normal limits. She reported having several disease processes primarily related to the diagnosis of CMT1A. The patient stated that the diagnosis of CMT1A had been made based on clinical symptoms and denied any familial history of neuromuscular symptoms. She was very knowledgeable about her diagnosis and associated comorbidities.

Comorbidities were as follows: neurogenic bladder with frequent urinary tract infections, gastric reflux attributed to pregnancy, numbness and tingling in extremities, occasional transient paralysis occurring over the last 2 to 3 years, headaches/migraines, and hypertension. The patient denied any history of back pain and was ambulatory. Home medications included labetalol, famotidine (Pepcid), and prenatal vitamins. The patient reported previous knee surgery, left claw-toe release procedure, and bilateral heel-cord releases. No anesthetic complications were recognized in conjunction with the previous surgeries.
The patient expressed explicit desire for regional pain management. Although the patient was well informed about her CMT, limited protocol was found to be readily available. It was agreed that regional anesthesia would be provided after consulting available anesthesia texts, the Internet, and discussion of available options and risks with the patient. The attending obstetrician was also consulted in the decision.

Monitors were in place as per American Association of Nurse Anesthetists (AANA) and ASA guidelines. Baseline vital signs were noted. The patient was prepared and draped in a sterile fashion in the sitting position. Local anesthetic was used at the identified L3-L4 interspace. An 18-gauge Tuohy epidural needle was used to identify the epidural space. A 27-gauge Pencan spinal needle was then used to provide a subarachnoid dose of 2.5 mg of bupivacaine with 12.5 µg of fentanyl citrate (Sublimaze) to initiate the combined spinal-epidural anesthetic. Cerebral spinal fluid (CSF) was clear, and no paresthesias were noted. The spinal needle was removed, and epidural catheter placement was achieved without difficulty.

After negative aspiration of CSF or blood from the epidural catheter, a continuous infusion of 0.125% bupivacaine with fentanyl, 2 µg/mL, was initiated at 16 mL/h. No difficulties with blood pressure or quality of pain management occurred. Three hours after the labor epidural was placed, additional anesthesia was established in preparation for cesarean delivery secondary to fetal intolerance to labor. Fifteen milliliters of 2% lidocaine (Xylocaine) in conjunction with 2 mL of sodium bicarbonate was given via epidural catheter in divided doses after negative aspiration of CSF.

The cesarean delivery was completed without incident, use of the epidural catheter was discontinued, and the patient recovered in a standard fashion. One day postoperatively the patient noted numbness of the right leg and foot. On the second postoperative day the patient described numbness reduced to the bottom of her right foot. The patient was discharged home on the third postoperative day and had no further complication.

**Discussion**

Charcot-Marie-Tooth disease is one of the most common inherited neurologic disorders. Obstetric anesthesia providers are sometimes hesitant or even emphatically opposed to offering a patient with CMT regional anesthesia secondary to the nature of the disease process. General anesthesia not only prevents maternal involvement in the birthing experience but also exposes the patient to the exaggerated inherent risks of general anesthesia in the obstetric population. The foremost source of morbidity and mortality among obstetric patients is the difficult airway/failed intubation and increased aspiration tendencies, especially associated with general anesthesia in the parturient.

This case report describes successful pain management for cesarean delivery using a regional anesthetic regimen in a patient with CMT. The only consistent sequela found unique to these patients, and confirmed by this case report, is delayed resolution of sensory block following epidural anesthesia. However, it is important to consider that neurologic complications following labor occur in the absence of anesthesia and are actually 5 times more common after childbirth itself independent of regional blockade. Also, pregnancy alone has been reported to cause exacerbations in CMT, causing some authors to consider it a risk factor for complications during delivery.

There are limited reports of this nature that suggest a regional anesthetic is an appropriate and effective option in this population. Consideration of the specific CMT subtype in anesthetic management was not found in the literature and is not considered pertinent in this case report. Only 8 specific reports of spinal and/or epidural anesthesia in CMT-affected patients were discovered in our literature review with only half used in obstetric patients. Six of the 8 cases reported no untoward effects, and none reported effects lasting beyond discharge. This case report supports the developing consensus in anesthesia that regional management is a safe alternative to general anesthesia in patients with CMT.

**REFERENCES**


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