

## CASE REPORT

# Perioperative care of an adolescent with Charcot-Marie-Tooth disease with placement of a caudal epidural catheter as an adjunct to general anesthesia

Omar Alkhatib<sup>1</sup>, Joseph D. Tobias, MD<sup>2,3</sup>

*1The Ohio State University School of Medicine, Columbus, Ohio (USA)*

*2 Department of Anesthesiology & Pain Medicine, Nationwide Children's Hospital and the Ohio State University, Columbus, Ohio (USA)*

*3 Department of Pediatrics, Nationwide Children's Hospital and the Ohio State University, Columbus, Ohio (USA)*

**Correspondence:** Joseph D. Tobias, MD, Chairman, Department of Anesthesiology & Pain Medicine, Nationwide Children's Hospital, 700 Children's Drive, Columbus, Ohio 43205 (USA); Phone: (614) 722-4200; FAX: (614) 722-4203; E-mail: Joseph.Tobias@Nationwidechildrens.org

### ABSTRACT

First described in 1886, Charcot-Marie-Tooth (CMT) disease is an inherited peripheral neuropathy which was originally termed peroneal progressive muscular atrophy. Given the invariable involvement of the neuromuscular system, anesthetic care is frequently required during surgical procedures aimed at correcting the orthopedic sequelae of the disorder. The authors present a 13-year-old boy with CMT who presented for anesthetic care during triple arthrodesis to treat pes cavus deformity of the foot. The perioperative considerations of patients with CMT are discussed with particular emphasis on the feasibility and safety of using regional anesthetic techniques.

**Key words:** Charcot-Marie-Tooth disease; Inherited peripheral neuropathy; Muscular atrophy; Regional anesthetic techniques

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### INTRODUCTION

First described in 1886 by Jean-Martin Charcot, Pierre Marie, and Howard Henry Tooth, Charcot-Marie-Tooth (CMT) disease is an inherited peripheral neuropathy which was originally termed peroneal progressive muscular atrophy.<sup>1</sup> The investigators also correctly localized the pathology to the peripheral rather than the central nervous system. CMT is caused by a genetic mutation that produces abnormal protein in the myelin sheath of axons of the peripheral nerves leading to susceptibility to degeneration. The more recent nomenclature designates CMT as Hereditary Motor and Sensory Neuropathy type I (HMSN-I). Further advances in genetic research have identified several types of CMT, which correspond with specific genetic mutations. Symptoms usually present during the first decade of life with distal limb muscle weakness, skeletal deformities including pes cavus, and decreased or absence of tendon reflexes.<sup>2</sup> Given the invariable and progressive involvement of the neuromuscular system, anesthetic care is frequently needed during surgical procedures aimed at correcting

the orthopedic sequelae of the disorder.<sup>2</sup> The authors present a 13-year-old boy with CMT who presented for anesthetic care during triple arthrodesis to treat pes cavus deformity of the foot. The perioperative considerations of such patients are discussed with particular emphasis on the feasibility and safety of using regional anesthetic techniques.

### CASE REPORT

Institutional Review Board approval is not required for single case reports at Nationwide Children's Hospital (Columbus, Ohio). This patient was cared for during a trip to San Miguel, Mexico by the Kids First Orthopedic Surgical Group. The patient was a 13-year-old, 53 kg adolescent with a diagnosis of Charcot-Marie-Tooth disease who was scheduled for triple arthrodesis to treat pes cavus deformity of the foot. He initially sought medical attention at 9 years of age when he started tripping and falling more during play. At that time, no firm diagnosis was made, but as his symptoms progressed, he also developed pes

cavus deformity. An electromyogram was performed and based on the results and the associated clinical symptoms including the pes cavus deformity of his foot, a diagnosis of CMT was made.

On the day of surgery, the patient was held nil per os for 6 hours and transported to the operating room. Routine American Society of Anesthesiologists' monitors were placed and a 20 gauge intravenous cannula was placed in his left forearm following local anesthesia with subcutaneous lidocaine. Anesthesia was induced with propofol (4 mg/kg) and a laryngeal mask airway was placed. Anesthesia was maintained with sevoflurane (inspired concentration of 2-4%). Following anesthetic induction, the patient was turned into the lateral position and the caudal area was prepped with chlorhexidine. The caudal epidural space was accessed with an 18 gauge, 2" Crawford needle and a 20 gauge catheter was threaded 10 centimeters into the caudal epidural space. The Crawford needle was removed and the catheter secured with a transparent, bio-occlusive dressing. Twenty mL of 0.375% bupivacaine with epinephrine (1:200,000) was administered in increments through the epidural catheter. There was no response to surgical incision. Sevoflurane requirements during the case varied from 1.5-2% (inspired concentration). Spontaneous ventilation was maintained throughout the procedure. No opioids were administered intraoperatively. The surgical procedure lasted 2 hours and 30 minutes. Estimated blood loss was 100 mL and total intravenous fluids included 1500 mL of lactated Ringers. At the completion of the case, the epidural catheter was redosed with 10 mL of 0.25% bupivacaine with epinephrine plus 50 µg of clonidine. The LMA was removed without difficulty and the patient was transferred to the post-anesthesia care unit (PACU). He denied pain in the PACU. The caudal epidural catheter was dosed the evening of surgery and again every 8 hours with 10 mL of 0.25% bupivacaine with epinephrine. The patient received only oral ibuprofen for postoperative pain management. The caudal epidural catheter was removed on postoperative day 2 and he was discharged home. His postoperative course was uncomplicated and he was doing well on his post-surgery follow-up visit at 6 weeks.

## DISCUSSION

Charcot-Marie-Tooth (CMT) disease is the most common inherited form of peripheral inherited neuropathy with an estimated incidence of 1/2500 to 10,000. The average age at onset is 16 years, but presentation in the early infancy and as late as 80 years of age has been reported. Presenting symptoms include both motor and sensory involvement usually of the distal lower limbs with weakness, muscle atrophy, and disturbances of gait with frequent falls as noted in our patient. Although motor involvement predominates, the majority of patients also report mild

to moderate sensory deficits involving the hands and feet. Life expectancy is generally not shortened.<sup>3</sup> CMT can be classified into various subtypes based on the nerve pathology, gene involvement, and inheritance pattern.<sup>4,5</sup> In general, there is the production of unstable myelin that accelerates programmed cell death of Schwann cells, resulting in demyelination with peripheral nerve involvement.

Given the invariable neurologic involvement resulting in muscular atrophy and weakness, one of the primary perioperative concerns regarding anesthetic management involves decisions regarding the use and choice of neuromuscular blocking agents (NMBAs). In our patient, given that the surgical procedure involved the extremity, neuromuscular blockade was not required especially in the setting of successful epidural blockade (see below). The airway was easily managed with an LMA which was placed following the induction of general anesthesia with sevoflurane and propofol. However, for more involved procedures, NMBAs may be required. Although there are limited data on which to draw precise conclusions, we would suggest that succinylcholine be avoided because of the potential risks of rhabdomyolysis and hyperkalemia in the setting of neuro-myopathic conditions.<sup>6-8</sup> However, in a review involving 86 cases, succinylcholine was administered during 56 operations in 41 patients without problems.<sup>9</sup> A defasciculating dose of a non-depolarizing NMBA was administered prior to 32 of the 56 uses of succinylcholine. The author indicated that although no clinical sequelae were noted, serum potassium levels were not measured and therefore the safety of succinylcholine cannot be verified. Likewise, Greenberg and Parker reported no sequelae following the administration of succinylcholine to 3 of 7 children with CMT during 10 surgical procedures.<sup>10</sup>

The response to non-depolarizing NMBAs may be variable. Although prolonged recovery has been anecdotally reported even following an intermediate acting agent (vecuronium), larger series reported no issues with prolonged neuromuscular following intubating doses of NMBAs including rocuronium, vecuronium, and atracurium.<sup>9-14</sup> However, concerns regarding the potential for a prolonged duration following a non-depolarizing NMBA have led to the use of mivacurium in this group of patients.<sup>15,16</sup> When compared to patients without neuromuscular diseases, the onset and recovery characteristics following mivacurium have been shown to be similar. However, this agent is no longer commercially available in the United States and many other countries.

Regardless of the NMBA that is used, monitoring of neuromuscular function using train-of-four stimulation has been recommended. Given the typical early involvement of the lower extremity, it has been suggested that monitoring should use either the facial nerve or the

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adductor pollicis muscle.<sup>14</sup> More importantly, it may be optimal to identify a suitable site for monitoring and assess the baseline response prior to the administration of a NMBA. Unlike other neuromuscular disorders, there is no evidence to suggest abnormal responses to reversal of neuromuscular blockade with acetylcholinesterase inhibitors such as neostigmine. Anecdotally, suggamadex has been suggested as an alternative to ensure complete reversal of neuromuscular blockade.<sup>17</sup>

There are limited data to demonstrate the benefit of one intraoperative anesthetic technique over another with efficacy and safety demonstrated with both total intravenous anesthesia including propofol as well as volatile anesthetic based techniques. Although one report raised the theoretical concern of the potential for MH, the remaining literature demonstrates no such associations. Of note, it has been demonstrated that the thiopental dosing requirements for anesthetic induction are decreased in patients with CMT.<sup>18</sup> In a prospective study, comparing the anesthetic induction dose of thiopental in 20 adults with CMT and 50 control patients, there was a decrease in the minimum induction dose (MID) of thiopental, defined as the dose required to produce loss of the eyelash reflex.<sup>18</sup> The 95% confidence interval of the MID in control patients was 2.5-4.9 mg/kg. The MID in 11 patients with CMT was less than 2.5 mg/kg and overall the MID in the patients with CMT was significantly less than control patients. The authors reported a strong relationship between the MID and the severity of both motor and sensory disturbances. Given the impact that this can have on recovery, careful titration of anesthetic agents is suggested with consideration to depth of anesthesia monitoring for prolonged cases. Another issue surrounding maintenance anesthesia is the safety of nitrous oxide. Given its interaction with vitamin B<sub>12</sub> and the theoretical impact on neurological function, several CMT associations caution against the use of nitrous oxide. However, in a retrospective review of 11 published reports involving 41 exposures to nitrous oxide, no adverse effects were noted.<sup>19</sup> Based on this, the authors concluded that nitrous oxide should be considered as a safe agent for use in children with CMT.

The potential for perioperative respiratory insufficiency may be the result of poor airway control with obstructive sleep apnea or diaphragmatic/phrenic nerve involvement.<sup>20-24</sup> Diaphragmatic involvement generally occurs later in the disease process in the adult years. There tends to be a correlation of upper extremity motor involvement with respiratory involvement.<sup>20</sup> Such involvement can significantly impact the perioperative course as demonstrated by a report of one patient being ventilator dependent for one month following a Cesarean section.<sup>25</sup> Anecdotal involvement of the recurrent laryngeal nerve resulting in vocal cord paralysis has also been reported.<sup>26</sup>

Any combination of these issues may place patients with CMT at risk for perioperative respiratory insufficiency or failure. Involvement of respiratory function may occur in the absence of clinical symptoms with a restrictive pattern noted on pulmonary function testing. For prolonged procedures or those involving thoracic or upper abdominal procedures which may impact postoperative respiratory function, preoperative assessment with formal pulmonary function testing may be indicated with a perioperative approach similar to what is recommended for patients with muscular dystrophy.<sup>27</sup> General precautions include postoperative monitoring of respiratory function in an ICU setting, tracheal extubation when the patient is fully awake following the dissipation of residual effects of intraoperative anesthetic agents, reversal of residual neuromuscular blockade, and the judicious use of opioid analgesia. Patients with impairment of respiratory function may benefit from extubation to non-invasive ventilation with a slow transition back to unassisted breathing.<sup>28</sup> The postoperative use of incentive spirometry or assisted cough devices may also be useful in preventing postoperative respiratory complications. As feasible, regional anesthesia, as was used in our patient, can provide postoperative analgesia while limiting the risk of respiratory depression from systemic opioids. When opioid analgesia is necessary, the use of adjunctive agents is suggested to decrease perioperative opioid requirements.<sup>29</sup>

Although relatively uncommon, myocardial involvement associated with CMT has been noted including the presence of arrhythmias and alterations in myocardial function. Cardiac conduction problems including heart block and arrhythmias (atrial flutter, premature ventricular contractions) have been previously reported including one report in a 9-year-old boy.<sup>30-32</sup> Likewise, cardiac involvement with cardiomyopathy has rarely been reported with motor neuron diseases, although only in the adult population.<sup>33,34</sup>

Over the years, concern has been expressed regarding the use of regional anesthesia in patients with pre-existing neurological disorders.<sup>35</sup> These relate to the theoretical concern that the regional anesthetic technique might somehow accelerate the progression of the disease process. Additionally, there may be acute exacerbations of the disease process in many of these disorders which may be inappropriately attributed to the regional anesthetic procedure. Despite this, the literature fully supports the safety and efficacy of regional anesthesia in the patient with CMT. Anecdotal experience has demonstrated the safety and efficacy of neuraxial blockade including spinal and epidural anesthesia.<sup>36,37</sup> Likewise, there have been anecdotal reports outlining the utility of peripheral nerve blockade of both the upper and lower extremity.<sup>38-41</sup> Ultrasound-guide techniques have demonstrated that the dimensions of peripheral nerves in patients with CMT are

similar to the normal population. Given its widespread use in clinical practice, the use of ultrasound is suggested given its utility in facilitating successful block placement while avoiding direct trauma to the nerve. Clinically palpable nerve enlargement occurs in 25% of patients with CMT type 1. It has been postulated that these patients may be at greater risk of positional nerve injuries due to pressure during prolonged procedures or the use of regional blockade in areas such as the elbow.<sup>11</sup>

In summary, we present the perioperative management of a 13-year-old boy with CMT for peripheral orthopedic surgery. Given the associated nerve and muscle involvement, one of the main perioperative concerns

is the use of NMBAs. Although there are no reports regarding complications following its administration, the use of succinylcholine is not recommended. Although anecdotal reports document prolonged duration following intermediate acting, non-depolarizing NMDAs, the majority of the literature reports no exaggerated response. Given the potential for diaphragmatic involvement with postoperative respiratory insufficiency, short-acting anesthetic agents and NMBAs are recommended with full awakening and recovery of neuromuscular blockade prior to tracheal extubation. Regional anesthesia may be used instead of general anesthesia or as an adjunct to provide postoperative analgesia.

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### **My most unforgettable patient**

### **History, history, history**

**Sarvjeet Kaur, MD**

*Associate Professor, Department of Anesthesiology, Guru Gobind Singh Medical College, Faridkot, Punjab, (India); Cell: +919915680508; E-mail: drsarvjeetk@gmail.com*

I want to share an incidence of a missed diagnosis. On a particular day, on call duty, I received a call from the emergency operating room, regarding a young 28 years female patient with severe abdominal pain in need of urgent exploratory laparotomy. I rushed to the emergency theatre. On examination, patient was conscious, anxious with pulse rate of 150/min, respiratory rate of 30/min, 100/60 mmHg of blood pressure and SpO<sub>2</sub> 97%. Since pregnancy test was positive and previous ultrasound report revealed ectopic tubal pregnancy, an immediate diagnosis of ruptured ectopic was made by the senior gynecologist. In view of the clinical condition of the patient and the diagnostic report, it was decided to proceed with exploratory laparotomy. Since major blood loss was anticipated, two wide bore cannulas were secured before anesthetizing the patient. Intravenous fluid were administered to stabilize the vitals of the patient. Requisition for blood and blood products was also sent to the blood transfusion department. The patient was administered general anesthesia and was intubated. On opening the abdomen, the ectopic pregnancy was found to be intact which added to the diagnostic dilemma. The ectopic pregnancy was managed appropriately. However, the patient continued to bleed and her vital sign worsened. Intravenous fluids and blood was transfused. The patient continued to bleed although temporary control was achieved by packing the surface of liver. Blood, platelets and FFP's were administered in the ratio of 1:1:1. In spite of the ongoing resuscitation, the patient continued to bleed and deteriorate. Vasopressors and ionotropes were started to maintain the hemodynamics. The liver surface was packed with reexploration scheduled after 48 hours and patient was transferred to intensive care unit (ICU).

While the patient was in ICU, the relatives of the patient revealed an old ultrasound report of the patient done about six months prior to this episode that clearly demonstrated multiple liver hemangiomas. Although the patient did not have any prior history related to the same, but the cause and site of bleeding was obvious now. It was certain that rupture of either single or multiple hemangiomas had lead to massive hemorrhage. Unfortunately, despite all efforts to maintain the patient hemodynamically stable, the patient died before she could be taken up for reexploration. Had we not missed the previous scan of the patient, we could have kept the possibility of the rupture haemangioma and could have sent the patient to a higher centre for embolization by an interventional radiologist in time since the same was not available at our institute.

Sure, it was a case of misdiagnosis. Taking proper history from patient and relatives and seeking any previous investigations should not be missed. Had a proper diagnosis been made, a precious life may have been saved.