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# ANAESTHETIC MANAGEMENT IN A PATIENT WITH CHARCOT-MARIE-TOOTH DISEASE UNDERGOING TRIPLE ARTHRODESIS OF THE FOOT

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**Abstract**

**Background:** Charcot-Marie-Tooth (CMT) disease is an inherited peripheral neuropathy that causes progressive muscle weakness and atrophy, particularly in the distal limbs. Anesthetic management for patients with CMT is challenging due to risks of neurotoxicity, altered responses to anesthetic agents, and potential for exacerbating muscle weakness. This report details the anesthetic care for a pediatric patient with CMT undergoing corrective foot surgery.

**Case Presentation:** A 19-year-old male with a confirmed diagnosis of CMT was scheduled for a triple arthrodesis of the foot to address progressive deformities and instability. Preoperative assessment revealed significant distal muscle wasting and sensory loss in the lower limbs, but no significant respiratory or cardiac compromise. The anesthetic plan prioritized regional anesthesia to minimize systemic drug exposure. The patient received a low-dose subarachnoid block with hyperbaric bupivacaine and a popliteal nerve block with ropivacaine for postoperative analgesia. The intraoperative course was stable, and neuromuscular function was closely monitored. The postoperative recovery was uneventful, with effective pain control achieved through multimodal analgesia, primarily using NSAIDs and paracetamol.

**Discussion:** The anesthetic management of patients with CMT requires careful consideration of their heightened sensitivity to both general and local anesthetics, as well as neuromuscular blocking agents. While regional anesthesia is beneficial for postoperative pain, it must be administered cautiously to avoid worsening the existing neuropathy. In this case, a balanced approach combining a carefully titrated subarachnoid block with diligent neuromuscular monitoring proved to be a safe and effective strategy, resulting in an uncomplicated postoperative course.

**Conclusion:** This case highlights the critical importance of a tailored anesthetic plan for patients with Charcot-Marie-Tooth disease. An approach that utilizes titrated regional anesthesia combined with close intraoperative monitoring can provide safe and effective management for patients undergoing orthopedic procedures such as triple arthrodesis.

**Keywords:** Charcot-Marie-Tooth Disease, Anesthesia, Triple Arthrodesis, Regional Anesthesia, Neuromuscular Monitoring, Peripheral Neuropathy.

## INTRODUCTION

Charcot-Marie-Tooth (CMT) disease, also known as hereditary motor and sensory neuropathy (HMSN), represents a group of inherited disorders that specifically damage the peripheral nerves. These nerves, located outside the brain and spinal cord, are responsible for transmitting commands from the brain to the muscles and relaying sensory information back to the brain. Named after the three physicians who first described it in

1886—Jean-Martin Charcot, Pierre Marie, and Howard Henry Tooth—CMT is one of the most common inherited neurological disorders, affecting an estimated 1 in 2,500 people worldwide.

The core of CMT's pathology lies in genetic mutations that affect the proteins involved in the structure and function of the peripheral nerves. These mutations can disrupt either the **myelin sheath**, the protective, insulating layer that surrounds the nerve axon, or the **axon** itself, which is the nerve fiber that conducts electrical impulses.

- **Demyelinating CMT (e.g., CMT1):** When the myelin sheath is damaged, the nerve signals are slowed down, leading to a progressive loss of function. This is the most common form of the disease.
- **Axonal CMT (e.g., CMT2):** When the axon is directly affected, the strength of the nerve signal is diminished.
- **Intermediate CMT:** Some forms of the disease share features of both demyelinating and axonal damage.

Clinically, CMT is characterized by a slow, progressive deterioration of the peripheral nerves, which primarily manifests as muscle weakness and atrophy, as well as sensory loss. The symptoms typically begin in the feet and lower legs and gradually advance to affect the hands and arms. This distal pattern of weakness is a hallmark of the disease.

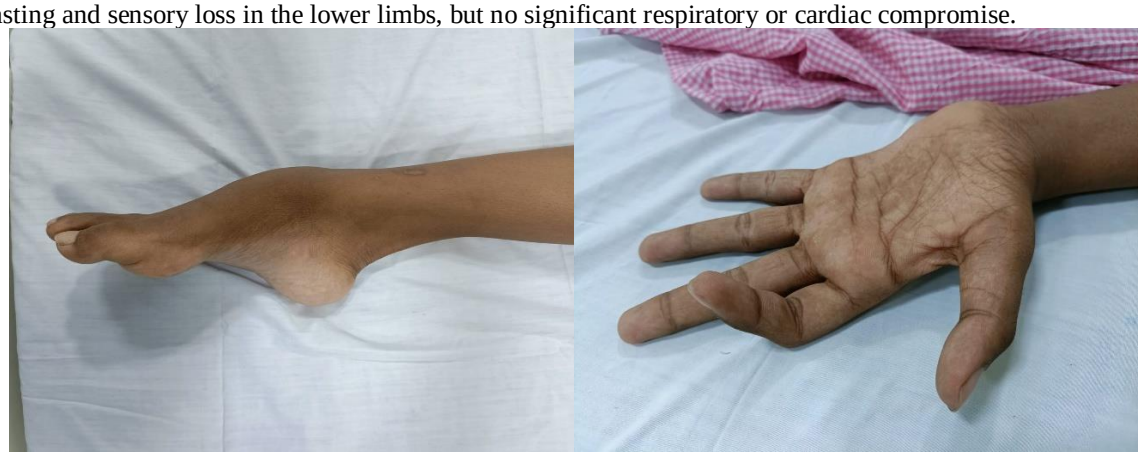
Common physical signs and symptoms include:

- **Foot deformities**, such as high arches (pes cavus) and hammertoes, which are often among the earliest signs.
- **"Foot drop,"** an inability to lift the front part of the foot, leading to a characteristic high-stepping gait and frequent tripping or falls.
- **Muscle wasting** in the lower legs, often described as having an "inverted champagne bottle" or "stork leg" appearance.
- **Loss of sensation** (touch, pain, and temperature) in the feet and hands.
- **Balance problems** and an unsteady gait.
- **Weakness and atrophy in the hands**, leading to difficulty with fine motor skills like writing or fastening buttons.

While CMT is a progressive condition, its severity and rate of progression can vary significantly, even among individuals within the same family. Most people with CMT have a normal life expectancy. However, the disease can lead to significant disability and a reduced quality of life due to chronic pain, mobility challenges, and the need for assistive devices like braces or wheelchairs. As there is currently no cure for CMT, management focuses on supportive care, including physical and occupational therapy, orthopedic surgery to correct deformities, and pain management to help individuals maintain function and independence for as long as possible.

**CASE PRESENTATION**

A 19-year-old male with a confirmed diagnosis of CMT was scheduled for a triple arthrodesis of the foot to address progressive deformities and instability. Preoperative assessment revealed significant distal muscle wasting and sensory loss in the lower limbs, but no significant respiratory or cardiac compromise.



**Preoperative Evaluation**

-Neurological Assessment: The patient had muscle wasting, especially in the distal lower limbs, with marked foot drop and claw toes. Sensory loss was observed in the lower limbs, predominantly in the feet. Gene sequencing studies were done it was confirmed to be CMT.

-Respiratory Assessment: No significant respiratory compromisewere observed in spirometry, maximal expiratory and maximal inspiratory pressures (MEP, MIP) were observed to within normal Limits

- Preoperative Tests: Routine bloodwork was unremarkable. Nerve conduction studies and EMG had absent Compound muscle action potential and absent sensory nerve action potential, consistent with CMT.
- Cardiac Assessment: ECG and Echocardiogram revealed no significant cardiac involvement

**Anaesthetic Challenges**

*1. Airway Management*

- Patients with Charcot-Marie-Tooth disease presents significant complications for anaesthetic administration, owing to the dangers of neurotoxicity, muscular weakness, and altered pharmacological reactions.
- Plan 1: Low Dose Spinal anaesthesia to minimize the involvement of abdominal and respiratory muscle and to limit the sub arachnoid block to Level of T10 or Lower.
- Plan 2: General anaesthesia with intubation, using short-acting agents to minimize post-operative respiratory depression.

*2.Peripheral Neuropathy and Sensitivity to Anaesthetics*

- Patients with CMT may be more sensitive to both local anaesthetics and neuromuscular blockers. The use of muscle relaxants increases the risk of persistent paralysis and may exacerbate muscle weakness.
- Plan:Avoid long-acting neuromuscular blockers. Short-acting medicines such as rocuronium, with close monitoring via a nerve stimulator, were preferable. Succinylcholine was not recommended because of the danger of hyperkalaemia.
-Monitoring: Routine Multiparametric monitoring HR, ECG (Lead II and Lead $V_5$) Neuromuscular function was carefully monitored intraoperatively to avoid prolonged blockade.

*3. Regional Anaesthesia Concerns*

- While regional anaesthesia reduces general anaesthetic doses and improves postoperative pain control, it is not without dangers for CMT patients. The use of local anaesthetics in neuropathic limbs may theoretically worsen nerve damage
- Plan:Anaesthesia administered with a carefully titrated Sub arachnoidblock was chosen for postoperative pain control. Low concentrations of local anaesthetics (hyperbaric bupivacaine) were used to minimize potential neurotoxicity.

*4. Postoperative Pain Management*

- Multimodal pain management was implemented, taking into account the patient's sensitivity to opioids. To limit opioid use, NSAIDs, paracetamol, and regional anaesthesia were prioritised.
- Plan:Regular acetaminophen, with rescue doses of a weak opioid (tramadol) if warranted.

**Intraoperative Course**

For maintenance, the patient had regional anaesthesia with a subarachnoid block. Following anaesthesia induction, ropivacaine was used to do a popliteal nerve block. The intraoperative monitoring was usual, with no major haemodynamic changes. Neuromuscular monitoring was used throughout the procedure.The procedure was done satisfactorily in three hours, with minimal blood loss and no intraoperative problems.

**Postoperative Course**

The patient was safely extubated and brought to the recovery room. The popliteal block provided great analgesia, with the patient reporting minor pain in the immediate postoperative period. He was watched overnight in the hospital and showed no signs of respiratory distress or muscle weakness. His discomfort was successfully managed with NSAIDs and paracetamol.

## DISCUSSION

The anaesthetic management of CMT patients presents many complications because to the possibility of increased sensitivity to both general anaestheticsand neuromuscular blocking medications. While regional anaesthesia can help with postoperative pain, it should be used with caution to avoid aggravating peripheral neuropathy.

In this situation, a balanced approach with attentive neuromuscular monitoring and a low-concentration regional block resulted in a safe and effective anaesthesia regimen. The postoperative recovery was uncomplicated, emphasising the necessity of individualisedanaesthetic management in individuals with neuromuscular disorders like CMT.

## CONCLUSION

This case highlights the importance of understanding the specific anesthetic risks associated with Charcot-Marie-Tooth disease. A tailored anesthetic approach, titrated regional anesthesia with close intraoperative monitoring, can provide safe and effective management for pediatric patients undergoing orthopedic surgeries like triple arthrodesis.

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