Pediatric patient with Charcot Marie Tooth undergoing foot reconstruction, role for regional anesthesia?

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Introduction

Charcot-Marie Tooth disease (CMTd), the most common hereditary neuropathy, affects approximately 1 in 3,300 people. Patients often require repeated palliative orthopedic procedures with challenging postoperative pain management. The use of regional anesthesia (RA), mainly peripheral nerve blocks (PNB) in patients with CMT is controversial due to fear of disease exacerbation. A few cases in the adult literature using PNB in CMTd patients, however, report no complications (2-4). There is no report of RA in pediatric CMT patients. We described the use of continuous perineural anesthesia in two pediatric patients with CMT disease undergoing foot reconstruction.

Case presentation

A 12 year old, 55 Kg girl with CMTd type I, was scheduled for unilateral foot reconstruction for equinovarus deformity. She had been asymptomatic without leg pain, weakness or numbness, but her foot deformities caused frequent falls. Nerve conduction studies showed demyelinating neuropathy. After discussion of the potential risks and benefits consent for RA was obtained. After induction of general anesthesia, an ultrasound (US) guided Sciatic Nerve (SN) block via popliteal approach was performed with one needle pass. 8 ml of 0.2% Ropivacaine was given and a catheter placed. A single shot ultrasound guided adductor canal block of the saphenous nerve was performed with 8 ml of Ropivacaine 0.2%. Ultrasound imaging showed abnormal sciatic nerve morphology with the presence of vacuoles within the perineurium. (Figure 1) Prior to emergence, the catheter was bolused with 7 ml of Ropivacaine 0.2%. In recovery room she reported 0/10 pain, and a perineural infusion of Ropivacaine 0.1% was started at 4 ml/hr. She remained pain free until POD 1, the infusion was stopped and she transitioned to PO meds. The patient had no neurological exacerbation or new deficit observed at follow up at 2, 3 weeks and 2 months after surgery.

Second case: a 17-year old, 54 kg, developmentally-delayed male with CMT-1A presented for right foot triple arthrodesis. For a similar previous contralateral surgery, the patient and family reported poor pain control using hydromorphone patient-controlled-analgesia. After discussion of the potential risks and benefits consent for RA was obtained. After induction of general anesthesia, an ultrasound guided sciatic nerve block via popliteal approach was performed with one needle pass. No motor response was elicited using a nerve stimulator. 5ml of 0.2% ropivacaine was administered and a catheter was placed. An infusion of ropivacaine 0.1% was started at 5ml/hr. Patient was comfortable in the postoperative period and catheter was discontinued on postoperative day 2. No complications from RA were noted. There was no exacerbation of the patient’s neuropathy at 6 wk and 1-yr follow-up.

Background

Charcot-Marie Tooth disease was first identified in 1886 by Jean-Martin Charcot and Pierre Marie from France, and Howard Henry Tooth from England. CMT is also known as hereditary motor and sensory neuropathy (HMSN) or peroneal muscular atrophy. CMTd includes a heterogeneous group of diseases classified in five groups with different genotypes and phenotypes, patients present varying degrees of demyelination and axonal loss which may affect susceptibility to additional nerve damage. ASRA guidelines recommend a cautious approach to RA and advise minimizing potential for needle trauma and local anesthetic (LA) exposure(s). Abnormal nerve conduction velocities, however, make nerve localization with nerve stimulation unreliable and nerve trauma from repeated needle passes to unresponsive nerves is a concern. When ultrasound is used nerve localization is possible with one needle pass. The low concentrations/volumes of LA we used gave excellent analgesia. This case demonstrates the safe use of a conservative approach to RA in a child with CMTd. If RA is considered, we recommend the use of ultrasound guidance to minimize the possibility of needle trauma and facilitate nerve identification. We also recommend using the lowest possible volume and concentration of LA. Additional studies documenting the use and safety of RA in pediatric patients with CMTd are needed.

References: