

Introduction

Primary Erythromelalgia is a rare hereditary disorder with:

- Extremity burning pain - triggered by warmth and relieved by cool temperatures and elevation.
- Erythema and increased temperature of the affected areas.
- Mutations in the SCN9A gene, leading to hyperexcitability of peripheral nociceptive neurons.
- Refractory pain with no established guidelines.

Here, we present the first documented case of using regional sciatic nerve block and peripheral nerve stimulator (PNS) for the treatment of erythromelalgia pain.

Case Presentation

17 year-old male with SCN9A-related inherited primary erythromelalgia presented with chronic, intractable burning pain in bilateral legs that had progressively worsened since age 7, resulting in disability. His condition was complicated by a recent hospitalization for foot/ankle ulcers and fungal osteomyelitis caused by prolonged direct exposure to air conditioning and ice water.

- Previously tried topical analgesics, NSAIDs (aspirin, meloxicam, nabumetone), duloxetine, gabapentin, carbamazepine, sodium-channel blocker (mexiletine), and opioids (morphine, methadone, oxycodone, tapentadol, lumbar sympathetic blocks and epidural infusions.



Image 1 and 2. Patient's bilateral leg wounds complicated by osteomyelitis caused by prolonged cold exposure and insensitivity.

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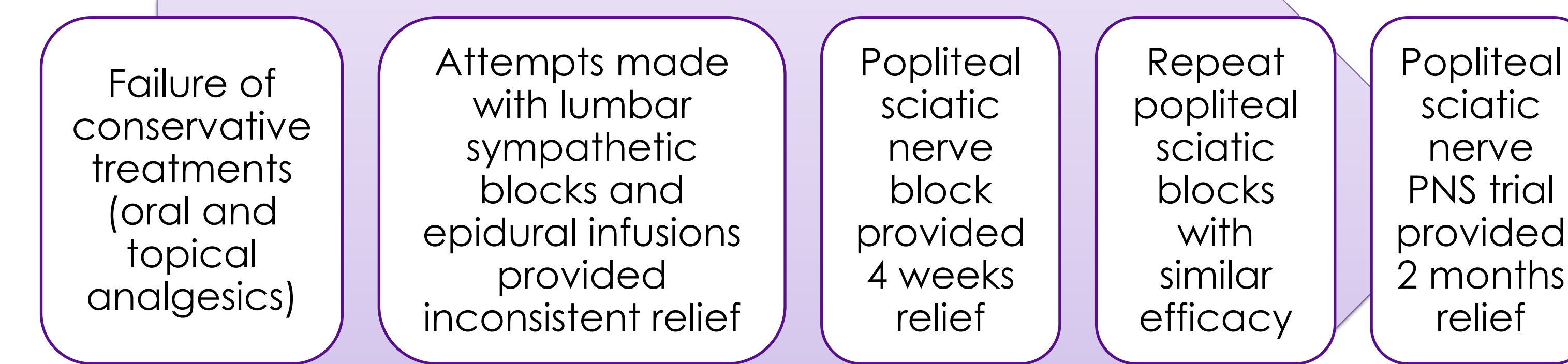
Successful Novel Treatment of Primary Erythromelalgia with Popliteal Sciatic Nerve Block and Peripheral Nerve Stimulator

Chun Maung, DO¹,
Christopher Chiodo Ortiz, MD¹,
Navi Plaha, DO¹,
Jinpu Li, MD¹

¹NYU Langone Health Rusk Rehabilitation

NYU Langone Health

Results



- Popliteal sciatic nerve block – 5cc of 1% lidocaine mixed with 20mg of triamcinolone under ultrasound guidance.
- Popliteal sciatic nerve PNS – 2 months of relief and improved functionality and wound healing. Able to return to school + activity.

Discussion

- Primary erythromelalgia is difficult to treat and often complicated by ulcerations, gangrene, and disability.
- Patient often immerse their limbs in ice water or cold exposure.
- Unclear exact pathophysiology on nociceptive pathway.
- Current literature consists of reviews on topical analgesics and oral medications with inconsistent efficacy, along with scattered cases of epidural infusions, sympathetic blocks, sympathectomy, DRG stimulation, and SCS as alternatives.
- **Regional nerve blocks and PNS may be viable treatment options for refractory erythromelalgia.**

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