Introduction: Erythromelalgia (EM) is a rare chronic pain disorder characterized by intense burning pain, erythema, swelling and warmth in the extremities. Pain is somehow relieved with immersion in cold water and aggravated by activity or heat. Early-onset EM is typically refractory to medical treatment.

Background: A 16 yo 58 kg female with early-onset EM presented to our institution for treatment. Her symptoms consist of bilateral leg and feet burning pain associated with erythema, pruritus and swelling. The pain was constant and best alleviated with prolonged cold water immersion. Patient was treated for skin breakdown, blisters and chronic fungal/bacterial superinfections. Her pain had been refractory to gabapentin, fluoxetine, opiates, acupuncture and routine wound care. She had issues with ambulation, school attendance and sleep hygiene, all contributed to a non-functional state.

Methods: After treatment of the lower extremity infection, she was offered a 1 month trial of tunneled L1-L2 lumbar epidural with 0.083% bupivacaine and 20 mcg/ml duramorph at 8 ml/hr. Within 24 hours she no longer required cold water immersion to reduce the pain which helped wound healing. Because of the successful epidural trial we placed a temporary spinal cord stimulator (SCS) under sedation. Electrodes were placed at T12-L1 level for 1 week. Because of the significant improvement in the patient’s symptoms and function a permanent SCS was placed by a neurosurgeon under general anesthesia. It was set at 2V, 10Hz and 300 microsecond pulse width. During the follow up period the patient reported resolution of the pain but still complained of persistent burning which was not managed by changing the SCS frequency (up to 100 Hz). She was then started on mexiletine 150 mg BID with some improvement of the burning sensation. The patient is back in school, mood and sleep pattern have improved and she can wear shoes again.

Discussion: To date this is the third case report on the use of SCS to manage pain in patients with EM (1 adult, 1 adolescent)(1,3). The exact mechanism of action of SCS is unknown and there are 4 proposed theories: the gate control theory, modulation of neurotransmitters release at the spinal cord level, modulation of peripheral nerve activity or supraspinal mechanisms(4). It is unclear why the SCS seems to selectively affect C-fibers transmission and spare Aδ fibers. It should be noted that a similar situation has been described in an adult patient with EM. She was treated with a SCS but the burning sensation was not affected by the SCS(3). More experimental studies are required to better delineate the interaction amongst pain, SCS, Aδ fibers and the transient receptor potential vanilloid receptor (TRPV1) to better understand our clinical observation.

References