INTRODUCTION:
Erythromelalgia (EM) is a rare chronic pain disorder. Little is known regarding the pathogenesis of the pain. In this case we discuss the pain management of an adolescent female with EM refractory to medical management.

CASE DESCRIPTION:
• 16 yo female with early-onset EM presented with bilateral leg and feet burning sensation and severe 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, multiple blisters and chronic super-infections.
• Her pain had been refractory to gabapentin, fluoxetine, opiates, acupuncture and routine wound care.
• A tunneled epidural catheter at L1-L2 was placed under general anesthesia.
• Within 24 hours she no longer required cold water immersion to control the pain.
• 30 days later, once the skin lesions had completely healed we placed a temporary spinal cord stimulator (SCS) under sedation.
• Given the resolution of the pain, a permanent SCS was placed 56 days later.
• Patient reported resolution of the pain but still complained of persistent burning sensation.
• We started a trial of Mexiletine (100 mg BID later increased to 300 mg BID) with significant improvements of the burning sensation.
• She is back in school, her mood and sleep pattern have improved and she can wear shoes again.

DISCUSSION:
• Invasive treatments can be considered in children with moderate to severe pain when the conditions that are life-limiting or irreversible and refractory to conservative treatment.
• A recent study of familial erythromelalgia demonstrated a mutation in the Nav1.7 channel.
• Digital cutaneous perfusion studies showed that patients with early-EM had increased regional tissue perfusion but decreased local capillary perfusion.
• This is the third case report on the use of a spinal cord stimulator (SCS) to manage pain in patients with EM but with persistent burning pain.
• The exact mechanism of action of SCS is unknown and proposed theories include: segmental antidromic recruitment and activation of large A beta afferents, blocking of transmission in the spinalthalamic tract, supraspinal inhibition, activation of central inhibitory mechanisms affecting sympathetic afferent neurons, or modification of neurotransmitters or neuromodulators.
• Both Aδ and C-fiber neurons contain thermosensitive receptors and perhaps the Aδ pain fibers were spared by the SCS because of size or location on the spinal cord.

SUMMARY:
• Erythromelalgia (EM) is a rare chronic pain disorder characterized by intense burning pain, erythema, swelling and warmth in the extremities.
• Patients with pain secondary to early-onset EM can be treated with a spinal cord stimulator but it may not cover the burning sensation.

REFERENCES: