**Case Report: CT-guided Lumbar Sympathetic Block on a 17-year-old Female with Erythromelalgia**

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**Background**

Erythromelalgia, a rare disorder, is described by affected individuals as "being on fire". Feet are most often affected, but may also involve hands and face. The primary form, seen in the pediatric population, is due to a genetic mutation at SCN9A. The amino-acid alteration within the alpha subunit of the 1.7 sodium channel isofrom dominant within C-Face (1, 2, 3, 2). Consequently, C-Face becomes hypereexcitable and small stimuli such as warmth, results in a disproportionate large response causing the individual to feel like they are being burned (4). Vasoconstrictor changes such as erythema and edema, believed to be secondary to autonomic dysfunction leading to vasocostriction and ischemia followed by reactive hyperemia, usually occur (5).

- Treatments include trigger avoidance, cooling the affected area, elevation, aspirin, phosphonates, mebendazole, clonidine, nortriptyline, anticoagulants, selective serotonin reuptake inhibitors, serotonin norepinephrine reuptake inhibitors, topical (capsaicin, local anesthetics, NSAIDS), antihistamines, acupuncture and cognitive behavioral therapy.
- There are several case reports of therapeutic response with epidural and sympathetic plexus blocks in individuals resistant to less invasive modalities (6, 7).
- Effective treatment is a challenge and response varies with each individual.

**Case Report**

- A 17-year-old SCN9A positive female, symptomatic from her first year of life, and was diagnosed with Erythromelalgia at five years of age. At the time of presentation, the lower 1/3 of her calves and face were warm, tender, and discolored, her hands and face were affected, and she was hypertensive (HTN).
- Starting at five years of age, her treated therapies include in ovo bullets, aspirin, cortisone, antihistamines, and hydroxyzine. She was then effectively treated with imuran, antihistamines and labetolol until August of 2001. In August 2001, she required hospitalization for treatment of lower extremity ulcers. Treatment consisted of intravenous antibiotics and sodium nitroprusside for vasodilation. Post hospitalization, her pain and hyperesthesia remained well controlled on phenytoin and nortriptyline until 2010. In 2010 (14 YO) there was a change in anti-hypertension therapy from phenytoin/nortriptyline to labetolol, and her EM became symptomatic.
- She was referred to Seattle Children’s Pain Medicine Clinic (PMC) in February of 2010. Additional therapies included T-Valcures, mexiletine, phenytoin, nortriptyline, clonidine, and gabapentin, though these failed to follow up.
- In February of 2012 she returned to the PMC after gabapentin and clonidine but more symptomatic. Her calves and feet were red-purple, edematous, the skin was exsanguinated, and her burning pain was rated at 9 to 10 on a scale of 10. She was unable to tolerate shoes, and kept her feet elevated with an electric fan. Moving air on them.
- Based on previous response to phenytoin/nortriptyline and clonidine, a case report documenting therapeutic success from a lumbar sympathetic block for erythromelalgia, lumbar sympathetic block (LSB) was performed for her.

- From March of 2012 to November of 2012, she has undergone a series of four, computer tomography (CT)-guided, LSBs using Triamcinolone (40mg) and Indomethacin (0.25% ID) bilateral side. Clinodrome (40 mg per side), was also added to the fourth block. Recently, gabapentin was changed to pregabalin secondary to the side effects of sedation and thus poor compliance with.
- Each LSB has been effective in providing pain relief for approximately three months with reduction in pain severity from 5 to 10 to 0 to 3 or 4 to 5 on a scale of 10, and decreased frequency of pain flare-ups. She has improved her quality of life as indicated by improved functional status, school attendance, ability to wear compression stockings and closed shoes; and participation in family activities including zip lining in Costa Rica.

**Therapeutic Target**

<table>
<thead>
<tr>
<th>Agent</th>
<th>Efficacy</th>
<th>Limitations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Central sensitization</td>
<td>Gabapentin</td>
<td>Effective</td>
</tr>
<tr>
<td>Sodium Channel Blocker</td>
<td>Phenoxybenzamine</td>
<td>Effective for 6 weeks</td>
</tr>
<tr>
<td>Sympathetic block, alpha 1</td>
<td>Clonidine</td>
<td>Effective</td>
</tr>
<tr>
<td>Nitrergic receptors</td>
<td>Neuraxial steroids</td>
<td>Effective</td>
</tr>
<tr>
<td>Information</td>
<td>Tramadol</td>
<td>Not effective</td>
</tr>
<tr>
<td>Antidepressants</td>
<td>Aripiprazole</td>
<td>Not effective</td>
</tr>
<tr>
<td>Antihypertensives</td>
<td>Nebivolol</td>
<td>Not effective</td>
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</table>

**Erythromelalgia**

Erythromelalgia is a life-long disease, therefore the lumbar sympathetic block serves both a therapeutic and diagnostic role. As her therapy moves beyond the limits of neuraxial steroids, our goal to produce durable vasodilation and blockade of the sympathetic chain remain. Possible future therapies include sodium channel blockers (IV lidocaine infusions and mexiletine have been ineffective for our patient) and highlighting the challenges in treatment of erythromelalgia despite a recognizable etiological factor (SCN9A positive).

**Discussion**

Treatment of Erythromelalgia poses significant challenges. The primary form is due to sodium channel dysfunction resulting in excessive firing of c-fibers. The life-long burning pain is exacerbated by every day activities such as walking, exposure to heat, and emotional stress. The disease is also characterized by decreased vasomotor control in response to sympathetic stimulation with increased sensitivity to circulating catecholamines. Erythema, calorimetry and temperature increase are thought to be a result of vasomotor control in response to sympathetic circulations with ischemia, followed by reactive hyperemia.

With the existing progress in understanding primary erythromelalgia, there is optimism that isoform 1.7 specific sodium channel blockers may be possible. Until then, our therapies remain focused on symptomatic management with therapies targeting central sensitization, sodium-channel blockers, alpha 1 and alpha 2 blockers, vasodilation, inflammation and humoral release. Unfortunately, for many erythromelalgia patients those therapies are not always effective in reducing the pain and symptoms enough to improve function and quality of life highlighting the multifactorial nature of the disease.

**References**


**Conclusions**

Our case report highlights that lumbar sympathetic block can be an effective therapeutic modality for erythromelalgia combined with other therapies improving patients quality of life. CT guidance allows for precision in technique while minimizing side effects and potential for injury to major blood vessels and internal organs.

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EM” by Andrea Dospert

The Erythromelalgia Association

*Image of CT-guided Lumbar Sympathetic Block on a 17-year-old Female with Erythromelalgia*