Treatment of refractory primary erythromelalgia in a child using a continuous epidural infusion.

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A 4-y/o male was referred to the Dermatology Clinic complaining of continuous burning pain in his hands and feet for the previous two years. He also had a poor appetite and disturbed sleep patterns. On exam, there was striking redness, warmth, and swelling of his hands and tenderness of his feet. Pain was relieved only by constant immersion of his feet in cold water. Laboratory studies revealed an iron deficiency anemia with a normal platelet count and elevated aspartate transaminase levels. The boy was of Asian origin, had two normal siblings, and was reported as floppy at birth without any other abnormalities noted. At 20 months of age he presented with a history of fainting. He was diagnosed as having seizures, which responded to anticonvulsants. Seventeen months later, he presented with evelopmental delay, hypotonia, micropenis, an undescended testicle, and eczema. All investigations at that time were normal.

A clinical diagnosis of primary erythromelalgia was made, but a number of topical and oral therapies failed to alleviate his discomfort and pain. Three months after presentation, he developed two ulcers on his left ankle, which were successfully treated with occlusive dressings. Seven months after presentation, a lumbar epidural catheter was inserted at L3-4 under general anesthesia. A solution of 0.1% plain bupivacaine and 0.25 mcg/ml clonidine was infused at 5 ml/hr following a 3 ml bolus. The rate was reduced to 3 ml/hr. He had immediate relief of pain in the left lower limb and was able to take it out of the cold water. There was no relief of pain in the right lower limb, and this leg remained in the water.

Erythromelalgia is a rare syndrome characterized by burning pain, erythema, and elevated skin temperature of the extremities. The lower limbs are more frequently affected than the upper limbs. The name is derived from three Greek words: erythros (red), melos (extremities) and algo (pain). Three types of erythromelalgia are described. Erythromelalgia associated with thrombocytopenia is the most common form. Primary erythromelalgia begins in childhood or adolescence as bilateral burning pain in the feet, ankles, and legs. The following six criteria are required for the diagnosis: 1) hands and

feet are erythematous, warm, swollen, and painful during attacks; 2) attacks are bilateral or symmetrical in the hands and feet; 3) attacks are brought on or exacerbated by exercise, standing, exposure, or warmth; 4) elevation and exposure to cold give relief; 5) attacks are refractory to medical treatment; and 6) there is no associated disease state or precipitating medication. Secondary erythromelalgia has been described in association with gout, systemic lupus erythematosus, rheumatoid arthritis, cryoglobulinemia, endarteritis obliterans, thromboangitis obliterans, polyarteritis nodosa, arteriosclerosis, diabetes mellitus, neurological conditions, vascular disease, and vasoactive drugs. Treatment is aimed at the underlying disorder.

The pathogenesis of primary and secondary erythromelalgia remains to be elucidated. A neurogenic etiology or vasomotor dysregulation has been suggested, although substance P has not been shown to be involved. Blood flow is increased in the extremities, although not at the tissue level, resulting in ischemia. Sympathetic over-activity may be the cause, which would explain why epidural infusions provide some benefit. Carbamazepine, beta-blockers, and tricyclic antidepressants have been reported to provide
relief, as well as gabapentin.