

Erythromelalgia: vasculopathy, neuropathy, or both? A prospective study of vascular and neurophysiologic studies in erythromelalgia.

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Davis MD; Sandroni P; Rooke TW; Low PA.
Department of Dermatology, Mayo Clinic, Rochester, MN 55905, USA.
davis.mark2@mayo.edu.

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OBJECTIVE: To assess the frequency and type of vascular changes and neurologic abnormalities in patients with erythromelalgia. **DESIGN:** Prospective study of patients with no spontaneous symptoms at the time of their visit and with provoked symptoms.

SETTING: Tertiary referral center.

PATIENTS: Sixty-seven patients presenting with erythromelalgia at Mayo Clinic, Rochester, Minn, from 1999 through 2001.

INTERVENTIONS: Testing nerve and vascular function in patients without symptoms present; testing vascular function after provoking symptoms with exercise or by increasing ambient temperature.

MAIN OUTCOME MEASURES: In patients in whom symptoms could be elicited, vascular function with and without symptoms was assessed by measurement of local skin temperature, laser Doppler flow, and transcutaneous oximetry. Neurologic assessment included electromyography, nerve conduction studies, and autonomic reflex screening (using the quantitative sudomotor axon reflex test, adrenergic function testing, heart rate response to deep breathing, and the Valsalva ratio).

RESULTS: Autonomic reflex screening was performed on 57 (85%) of the 67 patients. Of these 57 patients, 46 (81%) had abnormal quantitative sudomotor axon reflex test results; 14 (25%) had abnormal adrenergic function; and 15 (26%) had abnormal cardiovagal function. Put in another way, results were abnormal for 49 (86%) of the 57 patients who had autonomic reflex screening. Severe sudomotor abnormalities (ie, absent

or markedly reduced sweat production) were present in 46 (94%) of these 49 patients; 14 (29%) had abnormal adrenergic function, and 15 (31%) had a cardiovagal abnormality. Electromyography and nerve conduction studies were performed in 24 (36%) of the 67 patients. Of these 24 patients, 14 (58%) had abnormal electromyographic results and 10 (42%) had abnormal nerve conduction study results. Vascular function studies, with and without symptoms present, were performed in 13 of the 67 patients. During symptoms, the mean temperature of the toe skin increased by 7.8 degrees C, and blood flow increased 10.2-fold. Paradoxically, mean transcutaneous oximetry measurements did not change.

CONCLUSION: This prospective study extends and confirms our previous observation that, in addition to other forms of neuropathy, most patients with erythromelalgia have small-fiber neuropathy.