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WHAT IS ERYTHROMELALGIA?

Dr. Cohen developed severe erythromelalgia (EM) in 1995, when little was known about treating EM. He was disabled for several years but is pain free and highly active today. Dr. Cohen is an adjunct (voluntary) professor at the University of California, San Diego. He has published several medical journal articles on EM.

What is erythromelalgia (EM)? Why do our feet or hands turn red and hot? Why do they burn and swell? Why does the reaction last for many hours, and why is EM usually most intense in the late afternoon, into the evening and through the night?

These symptoms of EM are caused by excessive vasodilation, that is, an excessive opening of blood vessels. The symptoms occur in the skin of specific areas of the body: feet and legs and/or hands and arms, and less often ears or nose. EM symptoms occur in these areas because they are the heat exchange areas of the body. The blood vessels of these areas are meant to open a little to give off heat when the internal temperature of the body warms up.

In EM, this vasodilation is excessive. Why? Because injuries called neuropathies to the underlying nerve fibers in the skin. There are many types of neuropathies. Best known are the neuropathies that occur with diabetes. Other causes are illnesses, medications or surgery. Unlike other types of neuropathies, the neuropathies in EM set off a unique cascade of symptoms, a distinctive triad of redness, warmth and burning pain. Even in mild cases, EM causes people to limit their activities, and severe EM is disabling.

Instant Sunburn

EM has many similarities with migraine disorder, and both conditions can be reactive to a wide range of substances or conditions. Changes in temperature or barometric pressure can trigger an attack in some migraineurs. In EM, heat is the key trigger. But why? So far, no one has explained why mild ambient temperatures of 65 or 70 degrees Fahrenheit trigger EM symptoms so readily.

Equally baffling, why do EM symptoms stop almost immediately when ice or cool air is applied? I call EM "instant sunburn," yet a real sunburn does not disappear quickly. It takes days for a sunburn to go

away, and when it does, the skin is fully healed and the condition is gone. Why does EM respond so quickly to cooling, yet not heal or go away for good?

A Dysfunction, Not a Disease

A decade ago, researchers in Norway suggested that EM is not a distinct disease. Instead they suggested that EM is a dysfunction that causes abnormal activity of the blood vessels. The dysfunction in EM retreats when the skin is cooled, but it persists and returns quickly when triggered again by warmth.

If EM is a dysfunction rather than a disease, this explains why, in people who have had EM for decades, an effective treatment can cause EM to disappear overnight with no trace of damage to the tissues. Diseases tend to destroy, yet dysfunctions may not. This strongly suggests that the researchers in Norway are right, that EM is a dysfunction rather than a disease. This is helpful information, but it does not explain where or why the dysfunction in EM occurs.

Is EM An Impairment of the Body's Hyperthermia Response?

It is telling that EM occurs in the specific heat exchange areas of the body -- feet, hands, ears, nose - which the body uses to blow off excess heat. Blood vessels in these areas open, heat is dissipated, and the vessels narrow again. This usually occurs imperceptibly.

The human system functions optimally within a narrow range of internal temperatures around 98.6F (37C). What if the temperature rises much higher, as can occur when a person runs a marathon or hikes in the desert in summer? The body can overheat, and if severe, hyperthermia develops. Hyperthermia is a dangerous, life-threatening state. The body must cast off heat and quickly. To do so it initiates emergency actions that include a massive, maximum opening of the arteries in the heat exchange areas of the body. In seconds, the body can increase blood flow in these areas of the skin up to 32 times normal. Blood flow can skyrocket from one-quarter to eight liters per minute, an increase of 3200%. This mechanism is built into our systems to save our lives during hyperthermia. The point is that in hyperthermia, massive vasodilation in the heat exchange areas is normal!

But it is not normal under other circumstances. The massive vasodilation, causing the heat, redness, and burning pain of EM is not normal. At 70F, the skin of the feet and hands should remain cool. At 80 or 90F, the skin might become a bit warm and moist but otherwise normal. Yet in EM, the body reacts at 70F as if hyperthermia were occurring. Why is this? Clearly, the mechanism in the nervous system that regulates blood flow to these heat exchange areas is dysfunctional. It unleashes the skin reaction as if the temperature was 120F and hyperthermia was occurring. The regulator is off. The reaction is an aberration of a normal response.

Is there any proof of this hypothesis? There are two intriguing pieces of evidence. I have always wondered why the reaction responds so quickly to cooling. The ease with which EM's symptoms can be reversed is astonishing. And why does cooling work so well? Because it sends a specific signal to the hyperthermia regulator that all is well. Heat is the trigger. Cold is the antidote, temporarily, but not a cure.

Another piece of evidence is that the primary chemical mediator of the massive vasodilation in the skin in hyperthermia is nitric oxide. And in EM, what is the main mediator of increased blood flow? Nitric oxide.

This explanation is just a hypothesis, but it seems to have merit. The concept of a dysfunctional

hyperthermia trigger makes sense and offers a medically reasonable explanation for the development of EM. In addition, it offers patients and doctors a logical a way to understand this baffling and often disabling disorder, erythromelalgia. Hopefully it will lead to new avenues of research and treatment for those of us with EM, which is a dysfunction, not a disease.

NOTE TO READERS: Few studies have been done on EM, so there is a lack of established scientific fact about EM and its treatment. This article reflects my knowledge and personal experience with EM, and is meant to provide information for use by you and your doctor. This information should not be considered as a substitute for the medical advice of your doctor, nor is it meant to encourage the diagnosis or treatment of any illness, disease, or other medical problem without your doctor's direction. Readers should not make any changes in drugs, doses, or any other aspects of their treatment unless directed by their doctor. Finally, after many years of disability from EM in the 1990s, Dr. Cohen is now highly active with no pain, but because people with EM vary greatly in what helps them, he makes no claim that his methods and suggestions will benefit anyone else.

Dr. Cohen is an Associate (Voluntary) Professor of Preventive Medicine and Psychiatry at the University of California, San Diego, one of the top 20 universities in America. His work in the area of preventing medication side effects has been widely published and is recognized nationally. If you would like Dr. Cohen's input on your EM, he is available for office or telephone consultations. He charges a fee for his time, just as he charges people with other medical conditions who come to his office or consult with him from around the world. For information, contact Leslie at 858-345-1760 or schle@att.net.

Dr. Cohen's Publications on Erythromelalgia:

Cohen JS. The Medical Treatment of Erythromelalgia. MedicationSense.com. January 3, 2012.

Cohen JS. Supplements and Herbs in the Treatment of Erythromelalgia. MedicationSense.com. January 3, 2012.

Cohen JS. What Is Erythromelalgia? MedicationSense.com. January 3, 2012.

Cohen, JS. Erythromelalgia: New Theories and New Therapies. Journal of the American Academy of Dermatology, November 2000; 43:841-7.

Cohen, JS. Magnesium and erythromelalgia: a clinically important vasoactive mineral and a rare disorder. Italian Journal of Pediatrics 2004;30:69-72.

Also consider joining The Erythromelalgia Association (TEA), an excellent resource for information, published articles, and support for people with EM as well as for their families, friends and health care professionals. Readers can obtain information about membership and resources at www.erythromelalgia.org.

If you find this article informative, please tell your friends, family members, colleagues, and doctors about **www.MedicationSense.com** and the free MedicationSense E-Newsletter.