ERYTHROMELALGIA

Plan

Management
PLAN
1. Patient education-publications concerning this entity.
2. Prognosis- over time, the symptoms may worsen, stay the same, or get better. It happens about equal portions of people over time. In approx. 10%, patients may report resolution of symptoms.
3. Management-
The aim of management is that you will get back to as normal a style of life as possible. Our aim is to get your erythromelalgia under control and also to learn to live with whatever is left. My experience is that most patients can get acceptable control of their symptoms with lifestyle modifications and topical treatments; only a minority of patients need systemic medications.

There are three main strategies that we use to achieve this- * the use of topical medications for this,
* the use of systemic medications for this, and
* the employment of the Pain Rehabilitation Center in patients who are very disabled by the symptoms.

Lifestyle changes-- avoid situations as much as practically possible that set off symptoms, while keeping a balance and trying to maintain a normal lifestyle. This is a difficult balance, but ideally, she should be able to return to a normal style of life and not be inhibited by her symptoms as this is the aim of the medications that we have been prescribing.

Cooling behaviors:
Moderation is key.
* It is okay to use techniques that cool the affected areas for short periods of time (eg 5 minutes every 1-2 hours), but these should not be used for prolonged periods as they can damage the
skin, vasculature and nerves in the affected area and in the long run make the pain worse and lead to complications.

* Also moderation in the technique used is advisable- avoid extreme behaviors. Cool water indirectly applied to skin is advisable - but not ice or very cold water directly applied to the skin. Examples that patients report that help include Chillow pillows but again, these should be used for short periods. http://www.chillow.com/chillow.html

Suggested activities
Many patients report that swimming is helpful to them.

4. Patient support group- The Erythromelalgia Association

Topical treatments that may help are as follows.

Each should be tried one at a time for a period of 2-4 weeks, if medication does not work then move on to the next medication.

It would be very helpful if you could let us know which of below you have found most helpful via the patient portal after approximately 3 month

Most of these prescriptions are compounded products: it is best to have compounded creams filled at Mayo Clinic as they are familiar with how to compound these specific ones

A. A trial of amitriptyline 2% and ketamine 0.5% applied up to three times daily. Many patients report that this helps tremendously with their discomfort, but other patients say it does not work at all. I have advised to try this. If it does not work, a trial of giving a mixture with a higher concentration of ketamine such as 5% ketamine instead of 0.5% ketamine could be considered as a trial.

* a. Amitriptyline 2% combined with ketamine 0.5% in lipoderm
* b. Amitriptyline 2% combined with ketamine 5% in lipoderm
B. Lidoderm patches-- could try Lidoderm patches. These are easiest to use on the dorsal feet.
C. Topical mildodrine 0.2% in Vanicream applied TID
Other medications that could be tried in the future:
D. Once-daily topical brimonidine tartrate gel 0.5% (Mirvaso)
   (FDA approved for the redness of rosacea) could be tried—apply
   1-3 times daily
E. High-dose topical Capsaicin patch (Qutenza)—this recently
   came on the market, but we have not used it in anybody with
   erythromelalgia yet. Again, this might be a consideration in the
   future if the other treatments have not worked.
   http://www.qutenza.com/

F. Doxepin 5% cream
G. Trial of pain-relieving rub or patch (Ultra Strength BenGay
   Cream, Ultra Strength BenGay Pain Relieving Patch, others),
   voltaren 1% gel, gabapentin gel

Systemic treatments:
All systemic treatments are associated with potential side-effects,
and different people respond to different drugs. These are
summarized in the manuscript
   Drugs that have been reported to help include:
*   Aspirin—Trial of aspirin (if not contraindicated) 325 mg
daily x 1 month
*   Because a subset of patients respond to aspirin (particularly
   erythromelalgia associated with myeloproliferative disease), it is
   always worthwhile trying oral aspirin first for up to a month to
   see if that helps symptoms
*   Gabapentin
*   Pregabalin
*   Venlafaxine
*   Other drugs used for small fiber neuropathy
*   Mexiletine
*   Misoprostol

Pain Rehabilitation Center—
http://www.mayoclinic.org/pain-rehabilitation-center-rst/
For more than 30 years, Mayo Clinic's Comprehensive Pain
Rehabilitation Center (PRC) has helped people with chronic pain
return to an active lifestyle. Mayo's pain rehabilitation program was one of the first in the world. Now, it is one of the largest pain rehabilitation programs in the United States, treating approximately 600 patients annually. The programs use a behavioral therapy approach to help restore physical activities and improve the quality of life for people who have chronic pain conditions. A major emphasis of the programs is discontinuation of pain medications. The core team of health professionals who care for patients in the Pain Rehabilitation Center include physicians, psychologists, clinical nurse specialists, mental health therapists, nurses, physical therapists, occupational therapists, biofeedback therapists, pharmacists, dietitians, vocational rehabilitation psychologists, chaplains and nicotine-cessation therapists.

About Chronic Pain Rehabilitation
Chronic pain refers to pain that does not improve with time. This pain can affect a specific part of the body, as in low-back pain or headaches, or involve many regions at the same time, as in fibromyalgia or osteoarthritis. Chronic pain is more than a symptom of a disease, illness or injury; it becomes an illness unto itself. Regardless of location or cause, chronic pain causes many people to have difficulty functioning in daily activities. They may experience occupational disability, depression, drug-related complications and diminished quality of life.
In many cases, pain treatment such as medications, injections and surgery do little to relieve chronic pain or the long-term suffering and disability that can develop. Patients, their families and their doctors can become discouraged and frustrated. Mayo's Pain Rehabilitation Center offers hope and assistance in reversing the downward course that chronic pain can cause.
Please visit our

Mayo Clinic

Patient Education Center

to obtain the articles of interest from this list.

Located in the

Siebens Building, Subway Level

507-284-8140

patientec@mayo.edu
Sex differences in the incidence of skin and skin-related diseases in Olmsted County, Minnesota, United States, and a comparison with other rates published worldwide.

Andersen LK, Davis MD.

PMID: 27009931

Topically Applied Midodrine, 0.2%, an o1-Agonist, for the Treatment of Erythromelalgia.

Davis MD, Morr CS, Warmdahl RA, Sandroni P.

PMID: 25946117

Immersion foot associated with the overuse of ice, cold water, and fans: a distinctive clinical presentation complicating the syndrome of erythromelalgia.

Davis MD.

PMID: 23700236

Topical amitriptyline combined with ketamine for the treatment of erythromelalgia: a retrospective study of 36 patients at Mayo Clinic.

Poterucha TJ, Weiss WT, Warmdahl RA, Rho RH, Sandroni P, Davis MD, Murphy SL.

PMID: 23545913

Infrequent SCN9A mutations in congenital insensitivity to pain and erythromelalgia.

Klein CJ, Wu Y, Killfoyle DH, Sandroni P, Davis MD, Gavrilova RH, Low PA, Dyck PJ.

PMID: 23129781 Free PMC Article

Results of computer-assisted sensory evaluation in 41 patients with erythromelalgia.

Genebrieria J, Michaels JD, Sandroni P, Davis MD.

PMID: 22396570

Severe case and literature review of primary erythromelalgia: novel SCN9A gene mutation.

Skeik N, Rooke TW, Davis MD, Davis DM, Kalsi H, Kurth I, Richardson RC.

PMID: 22033523

1. Pediatric erythromelalgia: a retrospective review of 32 cases evaluated at Mayo Clinic over a 37-year period.
Gook-Norris RH, Tollefson MM, Cruz-Inigo AE, Sandroni P, Davis MD, Davis DM.
PUBMED: 21786623

2. Clinical pearls in dermatology.
Davis MD, Bundrick JB, Litin SC.
PUBMED: 20810755 Free PMC Article

3. Intervention for erythromelalgia, a chronic pain syndrome: comprehensive pain rehabilitation center.
Mayo Clinic.
Durosaro O, Davis MD, Hooten WM, Kerkvliet JL.
PUBMED: 19075140

4. Incidence of erythromelalgia: a population-based study in Olmsted County, Minnesota.
Reed KB, Davis MD.
PUBMED: 17132297 Free PMC Article

5. Thermoregulatory sweat testing in patients with erythromelalgia.
Davis MD, Genebrier J, Sandroni P, Fealey RD.
PUBMED: 17178864

Davis MD, Wilkins F, Rooke TW.
PUBMED: 15924077

7. Histopathologic findings in primary erythromelalgia are nonspecific: special studies show a decrease in small nerve fiber density.
Davis MD, Weening RH, Genebrier J, Wendelschafer-Crabb G, Kennedy WR, Sandroni P.
PUBMED: 16908308

Kumar N, Davis MD.
PUBMED: 16901020

9. Combination gel of 1% amitriptyline and 0.5% ketamine to treat refractory erythromelalgia pain: a new treatment option?
Sandroni P, Davis MD.
17. **Erythromelalgia.**
   Davis MD, Rook T.
   PMID: 16533490

18. **Lidocaine patch for pain of erythromelalgia: follow-up of 34 patients.**
   Davis MD, Sandroni P.
   PMID: 16220576

19. **Erythromelalgia.**
   Davis MD.
   PMID: 15008600

20. **Erythromelalgia: vasculopathy, neuropathy, or both? A prospective study of vascular and neurophysiologic studies in erythromelalgia.**
   Davis MD, Sandroni P, Rook T W, Low PA.
   PMID: 14568838

21. **Erroneous erythromelalgia.**
   Gastineau DA, Davis MD.
   Transfusion. 2003 Sep;43(9):1337; author reply 1337. No abstract available.
   PMID: 12919439

22. **Erythromelalgia.**
   Davis MD, Rook T.
   PMID: 12003720

23. **Lidocaine patch for pain of erythromelalgia.**
   Davis MD, Sandroni P.
   PMID: 11700162

24. **Mechanisms other than shunting are likely contributing to the pathophysiology of erythromelalgia.**
   Davis MD, Rook T W, Sandroni P.
   PMID: 11211183 Free Article

25. **Natural history of erythromelalgia: presentation and outcome in 168 patients.**
   Davis MD, O’Fallon WM, Rogers RS 3rd, Rook T W.
   PMID: 10724194


7/15/2016
26. Lidocaine and mexiletine therapy for erythromelalgia.
   Kuhnert SM, Phillips WJ, Davis MD.
   PMID: 10606048

   TV, Low PA.
   PMID: 19076553

28. Erythromelalgia as a form of neuropathy.
   Staub DB, Munger BL, Uno H, Dent C, Davis JS 4th.
   PMID: 1456799