



## **ERYTHROMELALGIA**

### **What are the aims of this leaflet?**

This leaflet has been written to help you understand more about erythromelalgia. It explains what this condition is, what causes it, what can be done about it, and where more information can be found.

### **What is erythromelalgia?**

Erythromelalgia is a rare condition. In ancient Greek, 'Erythros' means 'red' and 'melalgia' means pain in a limb. It can cause episodes of intense burning pain, painful swelling and [redness](#) of the feet or hands. Sometimes the legs and arms can be affected, or, less often, other areas such as the ears or face. It usually affects both sides of the body. Females are more commonly affected than males. Erythromelalgia can occur in all ethnic groups.

### **What causes erythromelalgia?**

Erythromelalgia is thought to be due to problems with the nerves that control the body's blood vessels. As a result, there is an increase in blood flow to the affected area which leads to pain, swelling and redness or skin darkening.

Erythromelalgia can be either 'primary' or 'secondary' and both have similar features. Primary erythromelalgia is caused by a change in your pain nerves due to a change in one of your genes. Primary erythromelalgia can occur at a younger age. In some cases symptoms may start at puberty.

The rarer 'secondary' form comes on later in life usually between the age of 40 – 60 years and is usually linked to another illness. Tests might be done to rule out other rare causes, such as medications, diabetes, lupus, HIV, bone marrow disorders, and pregnancy. In very rare cases, there may be other cause, including leukaemia.'

### **Is erythromelalgia hereditary?**

Primary erythromelalgia can run in families, so there is a one in two chance of an affected person passing this onto each child they have. However, sometimes a family history of erythromelalgia is not found (this is called "sporadic mutation").

### **What are the symptoms of erythromelalgia?**

The symptoms of erythromelalgia are redness or increased darkening of the skin, increased skin temperature and a burning sensation at the affected area. The condition may produce symptoms intermittently but occasionally they may be continuous. It can last for a few minutes to several days. Symptoms can begin gradually or suddenly. Sometimes people get itching before a burning pain, which can be mild or severe. In some people ulcers can develop at the affected sites.

A flare-up of erythromelalgia can be triggered by exposure to warmth, for example coming into a warm room, or by tight-fitting clothing, or by exercise. Some patients described symptoms worsening at night due to the increase in temperature from bedding.

Some people may experience significant psychological distress due to the pain and swelling that can be associated with this condition.

### **What does erythromelalgia look like?**

During a flare-up, the affected area may look red, blue, darker or mottled and swollen, and feels warm to the touch. In between episodes the skin can look normal, feel mildly cool or occasionally paler.

### **How is erythromelalgia diagnosed?**

Erythromelalgia may be diagnosed by the signs and symptoms without tests. Genetic testing may be considered if primary erythromelalgia is suspected. The genetic test looks for mutations or changes in your DNA called the sodium channel gene (SCN9A). If you have secondary erythromelalgia, blood tests may be requested to look for other diseases which could be causing the problem.

### **Can erythromelalgia be cured?**

There is no cure for erythromelalgia. The underlying cause for secondary erythromelalgia should be treated where possible.

### **How can Erythromelalgia be treated?**

Unfortunately, treatment can be difficult and is mainly aimed at trying to prevent flare-ups. Responses to treatment vary. If an underlying medical condition has been found, treating this may improve the erythromelalgia.

Self care:

- Loose clothing, cooling measures and elevating the affected body part may relieve the burning pain.
- Prolonged submersion in cold water is not recommended as this can lead to damage to the skin.

Primary erythromelalgia

- Treatments such as mexiletine that block sodium channels may be effective for some people.

Secondary erythromelalgia:

- It is important to treat any underlying cause.
- Aspirin can help relieve symptoms if the secondary erythromelalgia is related to myeloproliferative disorders

Medical treatments for erythromelalgia:

There are several different medications that can be used to try and relieve symptoms. Sometimes people may need to try several different medications or combinations of medications to find what helps them.

Treatments that your doctor may use include:

- Capsaicin cream or doxepin cream on the affected area.

- Medications which may help reduce pain caused by increased sensitivity of the nervous system
  - *Anti-convulsant medications:* e.g. Gabapentin, Pregabalin, carbamazepine.
  - *Newer antidepressants:* e.g. venlafaxine, sertraline, fluoxetine, paroxetine.
  - *Tricyclic antidepressants:* e.g. amitriptyline.
- Calcium channel blockers (e.g. nifedipine, diltiazem) which can alter blood flow.
- Intravenous infusions at the hospital (medicine through a drip) of nitroprusside, prostaglandin (iloprost) or lignocaine (lidocaine). Mexilitine tablets work similarly to lignocaine, but can be difficult for hospitals to obtain. A lidocaine patch applied to the skin may also help.
- Nerve blocks or surgical sympathectomy (a procedure in which sympathetic nerve fibres are cut) have helped some people, but not others. Sympathectomy can cause permanent severe side effects in some people.

If you experience any psychological distress associated with the condition, you should talk about this with your doctor. Cognitive behavioural therapy and other talking therapies can help some people cope with the distress the condition may cause.

### **Where can I get more information about erythromelalgia?**

*Web links to detailed leaflets:*

<http://dermnetnz.org/vascular/erythromelalgia.html>

*Support group for patients with erythromelalgia:*

<http://www.erythromelalgia.org>

For details of source materials used please contact the Clinical Standards Unit ([clinicalstandards@bad.org.uk](mailto:clinicalstandards@bad.org.uk)).

**This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: individual patient circumstances may**

**differ, which might alter both the advice and course of therapy given to you by your doctor.**

*This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel*

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