



## **ERYTHROMELALGIA**

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

This leaflet has been written to help you understand more about erythromelalgia. It tells you what this condition is, what it is caused by, what can be done about it, and where you can find out more about it.

### **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 is seen in skin types.

### **What causes erythromelalgia?**

Erythromelalgia is thought to be due to problems with nerves running from the brain to blood vessels in the affected area. As a result, there is an increase in blood flow to the affected area which leads to pain, swelling and redness.

Erythromelalgia can be either the more common 'primary' form of unknown or hereditary cause, or a rarer 'secondary' form, when it is linked to another illness. Secondary erythromelalgia usually presents later in life between the age of 40 – 60 years. Primary erythromelalgia can occur at a younger age. In some cases, puberty may precipitate symptoms suggesting a hormonal influence.

Causes of secondary erythromelalgia may include disorders of the bone marrow (myeloproliferative disorders) such as polycythaemia vera or essential thrombocytopenia, diabetes mellitus, rheumatoid arthritis, gout, human immunodeficiency virus, pregnancy, medications, postural tachycardia

syndrome, systemic lupus erythematosus, multiple sclerosis and some cancers including leukaemia.

### **Is erythromelalgia hereditary?**

Primary erythromelalgia can run in families and may be traced to a particular gene. However, in some cases a family history of erythromelalgia is not found

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

The symptoms of erythromelalgia are redness, increased skin temperature and a burning sensation at the affected site. Episodes may be intermittent or occasionally continuous and may last for a few minutes to several days. The onset of symptoms can be gradual or sudden. An itching sensation, which can range from mild to severe and disabling, may precede the burning pain. 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 worsening symptoms at night due to an increase in temperature from bedding.

Some patients 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, the affected area looks red or blue, 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 without tests by its signs and symptoms. Blood tests may be requested to look for an underlying medical condition.

### **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 unsatisfactory and mainly aimed at trying to prevent flares. Responses to treatment vary. If an underlying medical condition has been found, treatment of 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.

### 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. In some cases, patients may need to try several different medications or combinations of medications to find what helps them. Treatments that your clinician may use include:

- Capsaicin cream or doxepin cream to the affected area.
- Medications to 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 help regulate blood flow.
- Intravenous infusions at the hospital (medicine through a drip) of nitroprusside, prostaglandin (iloprost) or lignocaine (lidocaine). Mexilitine tables work similarly to lignocaine, but can be difficult 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, but were not beneficial in others. Sympathectomy can cause permanent severe side effects in some people.

Psychological distress associated with the condition should also be addressed by health care providers. Cognitive behavioural therapy and other talking therapies are of benefit to some patients to help patient cope with the distress the condition can 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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