What is discoid lupus erythematosus?

Cutaneous lupus erythematosus is a diverse group of autoimmune connective tissue disorders **localised to the skin** that can occasionally be associated with **systemic lupus erythematosus** (involving other organs within the body) to varying degrees.

Discoid lupus erythematosus (DLE) is the most common form of cutaneous lupus erythematosus, which is **usually confined to the skin** and is not commonly associated with symptoms from other organs. DLE is characterised by persistent, localised, red-pink or dark, scaly areas of affected skin most often on the head and neck. Less often the skin changes can be more generalised. Uncontrolled, DLE causes permanent skin damage such as scarring, darker or lighter pigment changes, and sometimes hair loss.

What does discoid lupus erythematosus look and feel like?

Occasionally the affected areas of skin can be uncomfortable or itchy. In most cases, DLE is confined to the skin with no effect on general health, although some people develop Raynaud's phenomenon (pain and colour changes in the fingers and/or toes in the cold) or perniosis (chilblains).

Patients with DLE can be **divided into localised and generalised**:

- **Localised DLE** - occurs when the head and neck only are affected and is nearly always confined to the skin. The most common sites are the cheeks, nose, and ears (especially the concha, which is the hollow next to the ear canal). The eyebrows may be thin and erythematous (pink-red)
- **Generalised DLE** - occurs when other areas are affected, regardless of whether the head and neck are involved. Patients with widespread involvement often have haematological and other abnormal blood tests, are more difficult to treat, and are more likely to develop SLE, although the overall risk for all cases of cutaneous lupus is somewhere in the region of 1.3-6.5%

The **skin changes** consist of:

- One or several well-defined **pink-red** areas of affected skin that may be flat or slightly raised. In skin of colour the affected skin is **often darker**
- **Adherent scale** (i.e. scale that is not easily removed) is common, as are **horny plugs** (plugs of keratin within hair follicles)

- **Untreated, DLE leads to permanent skin damage e.g. scarring, darker or lighter pigment changes, and sometimes hair loss**

What causes discoid lupus erythematosus?

DLE is an **autoimmune disease**, in which the body's natural defence system can't tell the difference between your own cells and foreign cells, causing the body to mistakenly attack normal cells

It is thought that a **combination of environmental factors and genetics** most likely contribute to the development of DLE.

DLE is more common in **females**, and in patients from **particular ethnic groups**, being slightly more common in African Americans than in whites or Asians. Although DLE is **uncommon in children**, onset at a young age increases the risk of progression to systemic lupus erythematosus (SLE).

Some families may carry **genes** that increase the risk of developing DLE; however, it is not entirely clear how the affected genes do this, or to what degree they influence the disease.

Environmental factors that may increase the risk of DLE or make it worse include exposure to sunlight, stress, infection, smoking, and trauma.

Rarely, DLE can be caused by drugs (e.g. anti-TNF inhibitors).

Things you can do to help yourself

The two most importing things are related to smoking and UV-protection.

Smoking

If you smoke, we strongly recommend that you stop. **Smoking tends to make DLE worse** and may result in a poorer response to treatment.

UV-protection

The **most important UV protective measures** are:

- **Avoid sitting or lying directly in the sun**, and remember you also get UV exposure when walking, playing sports, gardening, and even driving
- **Protect your skin with clothing**. Ensure that you wear a hat that protects your face, neck and ears, and a pair of UV protective sunglasses

- When outside, even on a cloudy day or when under a sunshade, use a **'high protection' sunscreen of at least SPF 30 which also has high UVA protection (4 or 5 star)**. Apply sunscreen generously 15 to 30 minutes before going out in the sun, or go swimming, and make sure you reapply frequently when in the sun. No sunscreen can offer you 100% protection - they should be used to provide additional protection from the sun, not as an alternative to clothing and shade. The sunscreen **should be prescribed by your GP** as it is being used for medical reasons
- **Avoid sunbeds**
- For more detailed information on UV-protection and advice on increasing your vitamin D intake (your vitamin D levels will drop if you reduce your UV exposure significantly) refer to our ***Skin Cancer Prevention*** leaflet, which can be found in the ***Patient Information Leaflet*** section at the bottom of the homepage of www.pcds.org.uk

Referral to a specialist

In the majority of cases you will need to be referred to a specialist, who could be a dermatologist or a GPwER/GPwSI (a GP who has been trained in relevant areas of dermatology).

What will happen when you are referred?

In most cases it is necessary to take a small sample of skin (a biopsy) to be examined under a microscope in order to confirm the diagnosis. Other tests may be performed including blood and urine tests.

Treatment

- **Skin (cutaneous) treatments** - these may include strong (potent) or very strong (super-potent) steroid creams, ointments, gels, or lotions. In the case of DLE these treatments can be used safely even on the face, under the direction of your specialist. Other topical treatments which may be offered in addition or as an alternative to topical steroids, are the topical calcineurin inhibitors, pimecrolimus and tacrolimus. In some patients with localised skin involvement, injections of steroids into the affected skin may be effective
- **Oral medications (tablets) -** if your skin/scalp changes are more severe and/or do not respond to cutaneous treatments, then systemics medications may be required. The most commonly used medications are the **anti-malarial drugs**

hydroxychloroquine and mepacrine. Occasionally, some patients may need additional medications, known as immunosuppressive therapy (i.e. drugs that suppress your immune system)

Your mental health

DLE can have a significant psychological impact and may affect many areas of daily life including work and personal relationships. If you require support with your mental health, then discuss this with a healthcare professional (see additional support below in the section on ***other resources***).

Can discoid lupus erythematosus be cured?

50% of DLE patients achieve complete resolution over many years. The prognosis (outcome) is worse if associated with Raynaud's phenomenon, chilblains, or hair loss.

Other resources

There are two patient support groups that can help with DLE:

- **Lupus UK** - www.lupusuk.org.uk
- **Changing Faces** - www.changingfaces.org can assist with cosmetic camouflage and psychological support

Helping with other skin conditions

If you, a family member, or friend have an undiagnosed skin condition; or you want to learn more about how to treat skin conditions, please visit www.pcds.org.uk – if you click the purple tile near the top of the homepage that reads '**Take a Tour of the PCDS Website'** you can learn how best to use this website.