

Lupus and the Skin

A Patient's Guide to Skin Involvement in Lupus



LUPUS and the Skin

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LUPUS UK is the national charity caring for those with systemic lupus erythematosus (SLE) and discoid lupus erythematosus (DLE), supporting people as they develop the symptoms prior to diagnosis and those already diagnosed.

You can help by taking up membership
For more information contact:

LUPUS UK, St James House, Eastern Road,
Romford, Essex RM1 3NH
Tel: 01708 731251

www.lupusuk.org.uk

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1. Introduction

Many people with lupus may have skin problems, and a rash may be the first sign of the condition. The rashes seen in lupus can be specific in shape and pattern.

Sometimes lupus only affects the skin as in Chronic Cutaneous or Discoid Lupus.

All forms of skin rashes in lupus are made worse by exposure to light (particularly sunlight). Other conditions such as rosacea can also be made worse by the light. Examination and testing the skin may help with the diagnosis.

Some people experience severe rashes, others relatively mild ones that respond to medications. Remember that the signs and symptoms of lupus can differ from person to person and the severity of signs such as a rash can be varied.

WHAT IS LUPUS ERYTHEMATOSUS (LE)?

This confusing name (literally “red wolf”) covers a group of related diseases, all of which can affect the skin. It includes Systemic Lupus Erythematosus (SLE), which can affect any system of the body. However, there are forms of LE (lupus erythematosus), which chiefly affect the skin. These include:

- **DISCOID LUPUS (DLE)** This typically affects sites exposed to sunlight, such as the face, backs of hands, scalp and ears. It presents as a red, scaly rash, which, if untreated, can cause scarring of the skin. Systemic symptoms are very rare.
- **SUBACUTE CUTANEOUS LUPUS (SCLE)** This form presents as a red, sometimes scaly, rash, also in sun-exposed areas, notably the V of the neck, arms and upper back. It is more common in women. Mild systemic symptoms are not uncommon, including joint aches and feeling ‘under the weather’. Severe illness is rare.



2. Types of rashes in lupus

Chronic Cutaneous (Discoid) Lupus

Is usually confined to light exposed areas like the head, neck, and the backs of the hands. The affected patches may be coin-shaped (discoid); they are usually scaly, red and sometimes slightly raised leading to scarring. If the scalp is involved, patches of permanent hair loss may result. The Latin word “lupus” means wolf; the scarred areas were likened to wolf bites in the past!



Note the rash on the side of the face and nose



Note the rash on eyebrows and nose

In time the areas may become paler or darker than the rest of the skin. Most patients with Discoid Lupus have no problems with their general health; only a few people with more widespread discoid lesions may develop Systemic Lupus Erythematosus in the future.



Note scaly rash on the side of the face



Note hair loss on scalp

In rare cases, the rash can cause loss of deeper tissue – where it can be localised such as in Lupus Profundus (a form of lupus that affects the fat underlying the skin, this can also be called panniculitis, which can result in firm deep nodules).

Subacute Cutaneous LE (SCLE) is a form of lupus that often starts as a widespread rash in areas exposed to sunlight. It has a red, ring like pattern, sometimes with a little scaling in sun exposed areas, and may be confused with psoriasis or ringworm. With treatment, it heals without scarring.

Sometimes SCLE can be sparked off by a reaction to a drug. People with SCLE usually have auto-antibodies (Ro and La), which are also found in patients with Sjögren's syndrome (dryness of the eyes and mouth).



Subacute lupus involving the neck and back



Subacute lupus involving the lips



Lupus Erythematosus involving the eyelid



Chilblain Lupus Erythematosus



Lupus Erythematosus involving the cheek

Newborn Infants

Very occasionally, a newborn baby develops a lupus rash. This happens during the first few weeks of life. The rash, which may resemble SCLE, mainly affects the head and face and can be brought out by sun exposure. It disappears, usually by the age of six months, leaving no scarring. The rash is due to the passage of

anti Ro antibodies across the placenta from the mother's circulation. Sometimes the mother is known to have lupus or a related disorder, but quite often she is symptom free.

Women who are known to be Ro positive are advised to seek specialist advice before planning a pregnancy in order to ensure the best outcome for mum and baby. Pregnancy is best planned while the disease is inactive and medication at a minimum. It may be best to continue with some medications to control active lupus (especially in those with SLE) and this should be discussed with your local medical and specialist nursing team preferably prior to planning conception.

SLE

The skin is involved in about two-thirds of people with lupus. Only about a third develop the classic 'butterfly' rash over the bridge of the nose and cheeks. Other skin changes include a widespread measles like rash, patches of discoid lupus or small areas of bleeding into the skin. Thinning of the hair can be seen at times of flare of the disease.

SLE can have an impact on your quality of life, especially when multiple symptoms such as joint pain, fatigue, mouth ulcers, headaches and many other symptoms affect your daily life. It is important to talk about your symptoms with your specialist nurse and review how these are managed on a regular basis. You can find more information on SLE from LUPUS UK.



Other causes of a 'Butterfly' rash

A butterfly rash is not always due to lupus and doctors need to be aware that certain common skin conditions can mimic lupus. Rashes commonly mistaken for lupus include rosacea, seborrhoeic dermatitis and occasionally, allergic reactions of the skin due, for example, to cosmetics.

Some people can develop a form of Vasculitis secondary to SLE, which is an inflammation within the blood vessels, as seen in the picture of a lower leg.



3. Mechanism of Photosensitivity

How sunlight affects lupus

Everybody will develop reddening of the skin after spending too long in the sun. The chief culprits are UVB, or the 'sunburn' wavelengths of ultraviolet (UV) light. Most people with lupus, both SLE and Chronic Cutaneous Lupus, are abnormally sensitive to UV light.

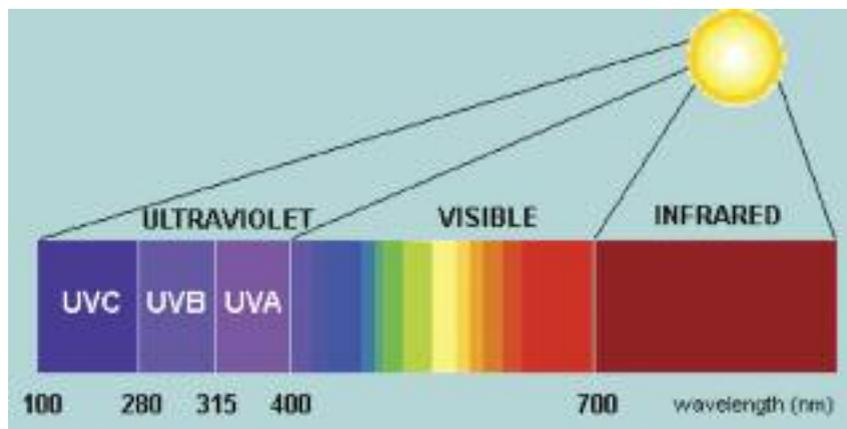
(Some light sensitive patients may also be harmed by long wave ultraviolet light (UVA) or even visible light, most commonly blue/violet spectrum).

UV light damages cells in the skin (Keratinocytes) causing them to die. In healthy people without lupus, these dead cells are cleared away quickly and any inflammation caused by the sun-induced skin damage is short lived (sunburn). However in lupus patients, the skin cells may be more sensitive to sun-induced damage and there is increasing evidence that the dying (apoptotic) cells are not cleared away efficiently. As a result the dying cells may be released and cause inflammation. Also, cell contents such as double-stranded DNA (the genetic material) and other molecules including the auto-antibody Ro, which are never normally exposed to cells of the immune system, are available to start an immune response.

Immune responses and inflammation are the normal reactions of the body to infection, but here, they are being generated inappropriately by the dying cells and the body mounts an immune response against its own cell constituents (autoimmunity). The end result of this process in susceptible people with certain types of lupus is the development of characteristic photosensitive rashes.

Auto-antibodies to Ro, in particular, are often found in people with these rashes. Why the rashes affect only some parts of the body at any one time and are not always sun-sensitive is still not understood.

Table A: The ultraviolet (UV) and visible part of the electromagnetic spectrum



[It is possible that some forms of household light can also cause a lupus flare. You may want to discuss this with your doctor or specialist nurse.]

Fluorescent lights

UVB is blocked out by window glass but many lupus patients report worsening of the rash even when sitting behind a window – for example, in the car. This suggests that UVA, which is not blocked out by glass, is also involved. Sometimes, even visible light aggravates lupus. Some patients are affected by fluorescent tubes in the home or at work, or even by photocopiers, which predominately radiate visible light.

4. Treatment of the skin in lupus

Treatment of the skin in lupus often needs a multi-disciplinary approach. Involvement of all members of the multi-disciplinary team, Dermatologist, Rheumatologist and Specialist Nurse, provide you with the support you need, with both medical and supportive nursing approaches. The emphasis is on self management so that you can understand how best to manage your symptoms in order to remain independent and improve your quality of life.

The great majority of cases, even with severe skin involvement, can be controlled with a combination of effective protection against UV light, the use of topical corticosteroids, and anti-malarial agents.

Drugs

Some drugs may cause or worsen SCLÉ or SLE. A large number of drugs such as thiazide diuretics and sulfasalazine have been implicated, but the risk of this is very low. Although stopping the offending drug can sometimes improve or resolve lupus, you must always discuss this with your doctor.

Topical treatment

Table B (page 9) lists some of the topical treatments for skin involvement. Locally applied corticosteroids can be very effective if the area of affected skin is not too large. These preparations vary in their strength from mild to very strong. Strong steroid preparations are often needed to begin with, but these should always be used under medical supervision because of the risk of skin damage.

Treatment by mouth

Only very rarely is it necessary to resort to steroids or immunosuppressive agents (drugs that suppress the immune system) by mouth for skin disease on its own. However if topical treatment (usually creams or ointments) fails, or if skin involvement is too extensive, tablet treatment may be needed.

Anti-malarial drugs are usually very effective, as they help to reduce inflammation and the severity of the rash. As the name suggests, these were used to treat malaria, but they are also effective in controlling skin lupus. We prefer to use hydroxychloroquine, as it is well tolerated and usually does not cause any major side effects. It is advised to have yearly eye checks with an optician as hydroxychloroquine may rarely affect visual acuity (reading the written word).

Table B lists some other drugs that can be useful, such as methotrexate.

Thalidomide, for example, can be effective in difficult cases, but pregnancy must



be avoided and a watch should be kept for side effects such as nerve damage, which may give rise to tingling in the fingers.

All drugs have the potential to cause side effects and the prescriber's responsibility is to balance the potential risks of the drug, versus the potential benefits and the risks of leaving the condition untreated which may be more harmful in the longer term. Always discuss with your doctor or nurse any concerns and worries you may have about medication so that you can make an informed decision about what you are being prescribed. It is important to continue with these medications sometimes over lengthy periods of time (months and years rather than days and weeks), in order to gain control of the symptoms and prevent any complications wherever possible.

Table B: Some of the drugs used for treating skin lupus

a) Topical creams or ointments - applied on the skin			
Agent	Comment	Precaution	Potential Side Effects
Corticosteroids	Often very effective. High strength ones usually needed. Prolonged use of high strength preparations can damage the skin, so should be used under medical supervision		Risks with skin thinning
Tacrolimus	A topical version of an immunosuppressive agent	Avoid smoking	Rash, irritation, pain, tingling or prickly sensation in the skin
b) Tablet Treatment – used when sun screens & topical agents alone have not worked. NB Monitoring of blood tests is required with most forms of tablet treatment			
Agent	Comment	Precaution	Side Effects
Anti-Malarial agents, e.g. hydroxychloroquine and/or mepracrine	Less effective if you smoke. Slow acting. Generally very safe	Eye screening and avoid alcohol above normal recommended limits	Gastro-intestinal upset, headache, very rarely visual changes and tinnitus
Dapsone	Can be effective for certain types of skin involvement, especially inflammation of blood vessels (vasculitis)	Regular blood tests	Usually related to higher doses, sometimes can cause changes in red blood cells but regular blood monitoring should detect this
Thalidomide	Can be very effective for resistant skin disease but only occasionally needed	Avoid pregnancy	Dry mouth, gastro-intestinal upset and sometimes nerve damage
Oral Steroids	Only required when in a severe flare		High blood pressure, diabetes, thin skin, thin bones although all of these are more of a risk when taken for longer periods of time
Acitretin	Sometimes effective for resistant discoid lupus	Avoid pregnancy	Dry mouth, dry skin, dry eyes
Other longer acting drugs that suppress the immune system such as Methotrexate, Azathioprine, Mycophenolate	Occasionally needed for skin lupus when rashes are severe and not responding to creams or antimalarials etc.	Avoid pregnancy when taking mycophenolate	Sometimes can cause reduction in white blood cells and abnormal liver function tests but regular blood monitoring should detect this

5. Sun Protection

Remember that sun is most intense at midday, and can be quite powerful on a hazy day. Lying under a sunshade does not protect you totally as sun can be reflected from water, etc.

There is no such thing as a 'total sunblock'. The protection factor given on the packet is accurate when the sunscreen is applied carefully under laboratory conditions, and only measures protection against short-wave ultraviolet light (UVB). In practice, sunscreens are never as powerful as they say, so you will need to re-apply your sun block regularly, particularly after swimming or if you are perspiring excessively, remembering the temples, ears, lips (specific lip blocks are available) and the back and sides of the neck, and the top of the scalp if there is thinning hair.

People who are sensitive to UV light should avoid lengthy exposure to direct sunlight, and wear a hat and long sleeves to provide a barrier. Sun screens are also needed for exposed parts of the body. Sun screening agents need to give broad protection against both UVB and UVA. The Sun Protection Factor (SPF) relates to the ability of the sun screen to absorb UVB. Most preparations also use a star-rating system (****UVA) for protection against UVA. You should use a sun screen that has an SPF of at least 50/60 and a four-star UVA rating. Your doctor can prescribe some 'total' sun screens. Most failures occur because the sun screen is not adequately applied. Sunscreen technology is changing all the time, but some good brands include Sun E45 factor 50 (white), or tinted sunblocks made by firms such as Clinique and Roc or the Sunsense range (www.sunsense.co.uk).

We recommend that sun screens are used from early spring and throughout the summer months, even if the weather is dull. For individuals who are very sensitive to UVA, a sun screen may be needed throughout the year.

Many outdoor pursuit shops sell clothes that can feel cool in hot weather but which block out the sun (some are even given a protection factor). If you hold up a garment to the light, and it lets any through, it will let sun through as well. Specialist suppliers of UV protective clothing include Sunsibility (**0208 224 2299** or www.sunsibility.co.uk) and (in the US) Sun Precautions (www.sunprecautions.com).

Laminated car windscreens block out UV, but side windows usually don't. DermaGard is a useful clear film that can be applied to windows in the car or house (Bonwyke Ltd, Unit 3, Salterns Lane Industrial Estate, Salterns Lane, Fareham, Hampshire, PO16 0SU. Telephone **01329 289621** or

www.bonwyke.co.uk). Protection against UV light is essential. Sometimes this is the only treatment needed. More information about such protective films can be found on the 'Eclipse' section of the LUPUS UK website www.lupusuk.org.uk/eclipse.

The sun is our main source of vitamin D which is necessary to help us to absorb calcium and form healthy bones. As many people with lupus are advised to avoid the sun due to its capacity to flare your symptoms and lead to rashes, aches and pains and fatigue, it is recommended that you ask your doctor if you can have a blood test to check that your vitamin D levels are within the normal range. If the results show low vitamin D, this can be replaced by eating foods high in vitamin D or taking supplements if required.

6. Quality of Life

Discoid Lupus

Discoid lupus has the capacity to scar the skin which can be distressing although the scars tend to heal well once the condition has burnt out. It is important to take advice and support when needed as this is something that can be improved with appropriate professional help. Sometimes plastic surgery is appropriate in certain situations; your hospital consultant dermatologist will be able to advise you.

There are some practical things that can be done to help such as a make-up service called cosmetic camouflage. This service previously run by the British Red Cross has recently been transferred to Changing Faces who support people that have conditions which affect their appearance. You can self-refer to this service or be referred by your GP/hospital team. More details can be found on www.changingfaces.org.uk The NHS has access to camouflage product suppliers such as Veil, Keromask, Dermacolor, Dermablend and Covermark.

7. Fatigue

Fatigue can be a major problem in lupus and can affect all aspects of daily activity. People with both discoid and systemic lupus can experience fatigue. This is different from normal tiredness and often described as exhaustion or an inability to re-charge our batteries. There is considerable help available to you if you suffer fatigue and certainly managing your daily activities with an awareness of fatigue is a good place to start.

The fatigue that you experience in lupus is different from the everyday tiredness that can be caused by the activities of daily life. When it is severe, it is a tiredness that is not always improved with rest, and can last for some considerable time. It could be triggered by certain activities or stress, but sometimes, the fatigue of lupus can catch you out. This can lead to frustration and sometimes periods of depression when you may feel that the fatigue of lupus is significantly interfering with your life plans.

Your local specialist hospital team can spend time guiding you through the principles of pacing and planning, managing exercise and rest and setting realistic goals. Often nurses and occupational therapists work together to help in this.

8. Self Help

Remember, you become the expert as you are living with lupus every day; your rheumatology team are there to help and advise you whenever needed. Everyone's experience of lupus is different and we all respond to symptoms in different ways, so don't worry about how you react as there are no specific rules! Here are a number of ways that you can help yourself to manage your lupus symptoms better:

- **Have a healthy respect for lupus**

Learning to recognise some of the things that can flare your lupus symptoms can help you to manage and limit the frequency and severity of problems. Listening to your body is important, recognising that some things you do in your everyday life may be worsening symptoms and acting on that knowledge can only help.

- **Holiday plans**

Sensible options in terms of holidays are always important and avoiding destinations of extreme heat when you suffer with photo reactive symptoms is always recommended. This does not mean you cannot go on holiday, just that you choose somewhere that is cooler and less likely to flare your symptoms. Always wear sun block of SPF 50/60 – and a four star UVA rating, and apply regularly, avoiding the midday sun and wearing clothing to protect your skin especially the arms, legs and V or neck. (If you are on medication to suppress the immune system, you would have to avoid certain 'live' vaccinations).

- **Understanding symptoms**

Keeping a log of symptoms and possible triggers can be useful to help you to understand your lupus; it could also be helpful to your doctors and nurses who could step in before major problems occur. You may for example notice more symptoms in the summer than in the winter due to the effects of sunlight, when you are stressed or when you are due your monthly menstrual period or during the menopause.

- **Living a healthy lifestyle**

By looking after your body, you can help limit complications. Eating a healthy balanced diet, one that is low in sugar, fat and protein and high in fibre is good for everyone. Controlling weight and blood pressure is particularly important in lupus as there is an increased risk of heart disease. Making sure exercise is part of your daily regime will also help. Smoking should be avoided and reduced wherever possible as this contributes to the risks of heart disease and you will improve your overall health if you stop smoking.

- **Work with your rheumatology and dermatology team**

It is important to talk to your doctors and nurses if you genuinely do not feel able to take certain drugs, as working together on an agreed treatment plan has to be the goal. You need to be informed of your health choices in managing your lupus and preventing complication. Always ask questions about your medication if you are worried about side effects. Prescribed drugs are an essential part of managing and controlling your lupus and most importantly, in preventing complications wherever possible.

More information is available from both LUPUS UK on www.lupusuk.org.uk and also Arthritis Research UK available on www.arthritisresearchuk.org.

9. Research

Research in the past decade has greatly increased our understanding of how skin inflammation occurs and about the causes of lupus in general. It has helped to explain, for example why the presence of some auto-antibodies are accompanied by a risk of skin inflammation and photosensitivity. This knowledge is leading to more targeted approaches to therapy which, hopefully, will transform the treatment of lupus and its skin complications in the not too distant future.

10. Further Reading

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A number of publications are available from LUPUS UK either online or by requesting a Sales Brochure from the charity on 01708 731251

Publicity materials, leaflets, posters, a dvd for the newly diagnosed, media releases and more are always available from the charity's National Office for better awareness about lupus in clinics, hospitals and public places.