



PATIENT INFORMATION LEAFLET

DARIER'S DISEASE

What are the aims of this leaflet?

This leaflet has been written to help you understand more about Darier's disease. It tells you what it is, what causes it, what can be done about it, and where you can find out more about it.

What is Darier's disease?

It is a rare inherited skin condition, in which the skin in certain areas develops large numbers of small brownish warty bumps.

What causes Darier's disease?

In the outer layer (epidermis) of normal skin, the skin cells are held together like bricks cemented in a wall. In Darier's disease the sticky junctions that hold the skin cells together are not made properly, and the skin may become scaly or lumpy or even form blisters. It is not due to an allergy and it is not contagious (catching).

Is it hereditary?

Yes. It runs in certain families, being inherited in a pattern known as dominant inheritance. This means that there is a 1 in 2 (50:50) chance that each child of an affected parent will inherit the condition. It affects both men and women. Its severity may vary considerably within a single family; and if a person is badly affected it does not necessarily mean that other family members who inherit the condition will also get severe disease.

What are the symptoms of Darier's disease?

Itching is very common. The affected skin may smell unpleasant, particularly in moist areas. This is probably caused by increased numbers of ordinary skin bacteria growing in the affected skin. The appearance of the rash, and its smell, can be embarrassing.

A quarter of patients notice that the condition improves as they get older. Some people find that the sun causes their Darier's disease to flare up. Some women notice that it worsens around the time of their periods.

What does it look like?

The first signs of the condition usually appear somewhere between the ages of 6 and 20. Little brownish, rough-topped bumps develop on the skin. The severity of the condition varies a lot and is unpredictable. The rash is often on the chest, neck or upper back at the start but warty bumps may occur on any part of the body including the skin creases and skin under the breasts. It is unusual for people to have much trouble on the face except for the skin on the forehead near the hairline.

The fingernails are usually affected. They tend to be rather fragile, split easily and look as if they have been bitten or appear dirty. There may be very obvious long red or white lines running the length of the nails. Nail changes and/or flat "warts" on the backs of the hands are often present in childhood, well before there are any other skin changes. Pits or small areas of hard skin occur on the palms of the hands and less often the soles of the feet. Occasionally there may be small spots inside the mouth and these may give the roof of the mouth a rough feeling.

How will it be diagnosed?

The diagnosis can often be made on the appearance of the rash and the fact that it runs in families. To confirm it, a small sample of skin (a biopsy) can be removed under a local anaesthetic and examined under the microscope in the laboratory.

Can Darier's disease be cured?

No, there is no cure, but there are many ways of helping it (see below)

How can Darier's disease be treated?

- *The itching:* Emollient or moisturising creams may relieve some of the irritation. Sometimes corticosteroid creams are helpful if the skin is very itchy. Antibiotic creams can be used if the skin becomes infected. The steroid and antibiotic creams are sometimes prescribed together in a mix.
- *The smell.* This is part of the skin condition and does not mean that the skin is dirty. Careful washing is important but, in addition, antiseptic solutions for the bath, antiseptic or antibiotic creams may help.
- *The use of retinoid tablets.* These are derived from Vitamin A (the current one that is used is called acitretin or Neotigason) and will improve the skin in most patients, but they can have several important side effects. If women take retinoids when they are pregnant, the medicine is likely to damage the developing baby. Therefore acitretin can only be prescribed for women of childbearing age who have been sterilised or who are using an effective contraceptive e.g. the contraceptive pill. Unfortunately this particular retinoid is excreted very slowly from the body and therefore pregnancy must be avoided not only while taking the drug but also for 2 years after stopping treatment. Retinoids also cause drying of the lips, eyes and nose. Used long term, they may also have effects on bones and fat levels eg cholesterol, in the blood. Patients with mild skin disease often find they would rather live with their Dariers rather than put up with these side effects.

- *Infections.* There is an increased chance of developing skin infections if you have the condition. Infection with the same sort of bacteria that infect cuts or grazes is common and may make the condition more active - this may require treatment with antibiotics from your doctor. The "cold sore" virus (herpes simplex) sometimes causes widespread skin infections in people with Darier's disease. If the condition suddenly gets worse and is much more painful than usual this may be a sign of a herpes simplex infection. Contact your GP or dermatologist urgently if this happens.
- There are some second line medications that are sometimes used 'off-licence' to help control Darier's disease. You can discuss these with your dermatologist.

What can I do?

Most people with Darier's disease lead full, normal lives and have no other medical problems. Less than a quarter need any time off work or school because the skin is a problem. However a number of things seem to make the skin worse, particularly heat, sweating and wool or nylon clothing. It is sensible to try and avoid these triggers. Some patients find that sun causes the skin disease to flare up. Sun protection using clothing, taking advantage of the shade and sun block creams (factor 15 or above) can help.

Where can I get more information about Darier's disease?

Patient support group:

Darier's Disease Support Group
PO Box 36
Milford Haven
Dyfed
SA73 3YF

Website: www.dariers.co.nr

Web links to detailed leaflets:

www.emedicine.com/derm/topic209.htm
www.dermnetnz.org/dna.darier/darier.html

(Whilst every effort has been made to ensure that the information given in this leaflet is accurate, not every treatment will be suitable or effective for every person. Your own doctor will be able to advise in greater detail.)

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