

## **Darier's Disease**

### **What is Darier's disease?**

Darier's disease is a rare, genetic disorder which causes a decreased and abnormal sticking together of skin cells and results in a variety of changes including irritating greasy crusted lumps on the skin and nail changes.

### **What causes Darier's disease?**

Darier's disease is caused by a gene mutation. Each child of an affected adult will have a 50% chance of inheriting the disorder. The disorder may also be caused by a new mutation without any previous history of family members being affected.

### **What does Darier's disease look like?**

Visible skin changes of Darier's disease vary from person to person but are usually first noticed between the ages of 6 and 20 years. Yellow-brown or skin-coloured, small greasy lumps develop in the "greasy" areas of the body such as the scalp, face, neck, chest, armpits, groin and under the breasts. These can become joined together to form a raised, flat-topped or warty mass which can become smelly, itchy, irritated and uncomfortable.

Abnormal changes to the nails may include a 'v' shaped nick at the end of the nail and red and white stripes.

Other changes may include small depressions in the skin of the palms of hands and soles of feet, small lumps on the back of the hands and white lumps inside the mouth and vulva.

Darier's disease often gets worse in summer due to increased heat, sweating and exposure to sunlight. Bacterial, viral (cold sore virus) and yeast (thrush) infections are common. Infections are usually painful and blisters may develop. Some women with Darier's disease may have premenstrual flare-ups. Stress and friction may also trigger flare-ups.

There are more unusual forms of Darier's disease which have features such as acne (pimples) and others where either one side or a particular part of the body is affected.

Most people affected with Darier's disease do not have other medical problems.

### **How is Darier's disease diagnosed?**

A skin biopsy is necessary to confirm the diagnosis.

### **How is Darier's disease treated?**

There is no known cure for Darier's disease. Treatment is given to relieve symptoms, treat complications such as infection and improve quality of life.

The following general measures may be helpful in preventing flare-ups. These include: avoiding triggers, using adequate sun protection, avoiding excessive heat and wearing loose cool cotton clothing.

Soap substitutes, moisturisers and creams containing urea or lactic acid may be used in addition to topical vitamin A creams, although these may also cause irritation. Topical steroid creams may be helpful in reducing irritation and itch. Regular use of antiseptics in the bath may be recommended to reduce the

chance of developing an infection.

Sudden onset of pain or weeping or blisters requires a visit to the local doctor for a swab and then treatment with antibiotics or antiviral medication.

Oral medication (acitretin) may be used to treat widespread Darier's disease especially in the summer months. However, this form of treatment is not suitable for all affected individuals. Acitretin is not safe in pregnancy and must be prescribed by a dermatologist.

Surgical or laser removal may be appropriate for smaller severely affected areas such as under the armpits.

Referral to a geneticist or genetic counselling may be appropriate.