



DARIER DISEASE

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Darier disease (also known as keratosis follicularis or Darier-White disease) is a rare genetically inherited skin disorder that can be characterized by multiple dark scaly patches of itchy skin most commonly affecting the chest, back, ears, forehead, scalp, neck, and groin areas. These wart-like lesions can be foul smelling and disfiguring and has been known to affect nails and mucous membranes of the individual.

The occurrence of Darier disease is rare with the age of onset usually in the first or second decade of life. It is frequently worse in the summer with heat and humidity as major precipitating factors and can be exacerbated by sun exposure, trauma, or bacterial infections.

The cause of this disease is oftentimes due to a mutation in a specific gene known as ATP2A2 which can alter the function and development of the skin. The affected parent with this gene has at least a fifty percent chance of passing it down to their children. Patients with this disease may be associated with behavioral disorders and rarely with decreased intelligence. Most patients with Darier disease have a family history of either one or both parents being affected. However, it can also present itself without any family history as well. Even though the severity fluctuates over time, Darier disease is a chronic condition that persists throughout life and is not associated with any skin cancers.

Depending on the area affected, patients with Darier disease often presents with multiple crusty and itchy patches of skin. When the scaly crusts are removed, a slit-like opening becomes visible. In areas such as nails, they are described as a sandwich of red and white bands running along the length of the nail that is thin with its characteristic V-shaped scalloping. Mucous membranes may present as white, cobblestone appearing lesions of the cheeks, palate, and gums. With the discovery of the ATP2A2 gene, **skin biopsy** is helpful in diagnosis through gene sequencing when suspected.

Patients diagnosed with Darier disease are recommended to wear **sunscreen**, cool cotton clothing, and avoidance of warm environments to prevent flares, especially during the summer season. Moisturizers with urea or lactic acid can help reduce scaling and thickening of the lesions. Low to medium strength **topical steroids** are sometimes useful for reducing inflammation and when bacterial growth is suspected, application of antiseptics can be helpful. **Topical retinoids** such as tazarotene and adapalene have been shown to be effective in reducing the localized symptoms of this disease but the most effective medical treatment for severe cases by far has been the use oral retinoids such as **acitretin** and **isotretinoin**. The prolonged use of oral retinoids, however, is often limited by its side effects which includes, but are not limited to, mucosal dryness, photosensitivity, abnormal liver functions, and bone deformities. Oral retinoids are highly teratogenic and should not be given to female patients without proper counseling and contraception. Surgical treatment of Darier disease includes dermabrasion, **electrosurgery**, and **MOHS micrographic surgery**, however, recent advances in the use of **lasers** have been reported with positive outcomes of remission up to two years.

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