



URTICARIA PIGMENTOSUM

<http://www.aocd.org>

Urticaria pigmentosum is an eruptive skin disease, caused by a sudden increase in mast cells. Mast cells are released by the body, in response to an increase in inflammation, or during an allergic reaction. Urticaria pigmentosum is the most common form of cutaneous **mastocytosis**.

In response to excessive heat, or rubbing the skin, the mast cell releases a substance called histamine. The release of histamine produces the characteristic red-to-brown lesions, or hives on the skin. When lesions develop on areas of the skin directly caused by the irritation, it is referred to as the Darier's sign. Environmental and psychosocial factors can trigger an allergic reaction. Certain drugs (aspirin, codeine), excessive heat or friction, alcohol, stress, and infection, have all been considered as irritants that can cause a reaction.

Urticaria Pigmentosum is mostly caused by a mutation in the proto-oncogene, C-kit. C-kit is a transmembrane, that when bound, signals the Mast Cell Growth Factor (MCGF) to divide. A point mutation in the C-kit gene results in excessive mast cell production.

Diagnosis is usually done in infancy. It is a clinical diagnosis that can be confirmed by the physician, through rubbing on the patient's skin. When the disease is diagnosed in adulthood, the disease may be more severe in presentation, including anaphylaxis and death. A **biopsy** of a skin lesion can be performed if there is doubt about the diagnosis.

Treatments for urticaria pigmentosum are the same as those used for cutaneous mastocytosis, and are aimed at mast cell stabilization. **Antihistamines** and mast cell stabilizers (cromolyn sodium) are used to prevent mast cell degranulation. Researchers suggest that nifedipine, a calcium channel blocker, may have mast cell stabilizing effects. **Topical steroids** under occlusion and phototherapy (**PUVA**) are common forms of treatment for adults.

This information has been provided to you compliments of the American Osteopathic College of Dermatology and your physician.

The medical information provided in this article is for educational purposes only and is the property of the American Osteopathic College of Dermatology. It is not intended nor implied to be a substitute for professional medical advice and shall not create a physician - patient relationship. If you have a specific question or concern about a skin lesion or disease, please consult a dermatologist. Any use, re-creation, dissemination, forwarding or copying of this information is strictly prohibited unless expressed written permission is given by the American Osteopathic College of Dermatology. For detailed information including links to related topics on this and many other skin conditions with photos, visit: <https://www.aocd.org/page/DiseaseDatabaseHome>