



URTICARIA PIGMENTOSA

What are the aims of this leaflet?

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

What is urticaria pigmentosa?

Urticaria pigmentosa (UP) is the commonest presentation of cutaneous mastocytosis. The other three are called mastocytoma, diffuse cutaneous mastocytosis (DCM) and telangiectasia macularis eruptiva perstans (TMEP). UP and TMEP are now grouped together using the term **maculopapular cutaneous mastocytosis**.

Mastocytosis means too many mast cells. Mast cells are part of our immune system. They develop in the bone marrow then spread in the blood all over the body, including the skin. They release histamine if stimulated by different triggers. Urticaria pigmentosa is composed of persistent brown or red marks, made of collections of mast cells that swell and itch transiently when rubbed, similar to a hive. In the majority of cases, urticaria pigmentosa may cause skin symptoms but does not progress to more serious types of mastocytosis.

More than 75% of cases of urticaria pigmentosa start in infants and children less than 10 years old, but it can also affect older children and adults for the first time. It affects male and female patients equally.

What causes urticaria pigmentosa?

Urticaria pigmentosa is an accumulation of mast cells which have the same genetic mutation. The common mutation called D816V in the receptor called c-KIT has been found in up to 95% of adults with the condition and in some children. The reason for this mutation is still unknown.

The release of histamine can cause a variety of symptoms ranging from itching, hives (also known as nettle rash), wheezing, and diarrhoea, and uncommonly to life-threatening collapse (anaphylaxis). Certain factors may trigger the release of histamine and the other chemicals from mast cells, including drugs (codeine and morphine, some general anaesthetic agents, aspirin and other non-steroidal anti-inflammatories), alcohol, emotional stress, physical stimuli (heat, exercise and skin friction) and insect stings.

Is urticaria pigmentosa hereditary?

No, it is not hereditary but there are a few reports of more than one family member being affected. The reason for this is unknown.

What does urticaria pigmentosa look like?

Urticaria pigmentosa has a distinctive appearance consisting of brown or red marks or swellings called papules that are predominantly on the trunk and limbs. The face is not usually affected. They may be mistaken for changing moles. Your dermatologist may consider taking a biopsy of the skin lesion if there is diagnostic doubt.

What are the symptoms of urticaria pigmentosa?

If urticaria pigmentosa marks are rubbed firmly, the mast cells release histamine causing them to become itchy, swollen and, occasionally, blistered. This is known as Darier's sign and is very characteristic of urticaria pigmentosa. Up to 50% of adults with urticaria pigmentosa may develop anaphylaxis over their lifetime but this is unusual in children unless they have a large number of urticaria pigmentosa skin lesions affecting a large area of their skin. Triggers for anaphylaxis include bee or wasp stings, certain foods or medicines. These reactions may be unpredictable and due to multiple factors. Some patients may receive a recommendation to carry an adrenaline autoinjector.

The release of large amounts of histamine from mast cells may cause flushing in addition to itch, a racing heartbeat, diarrhoea, wheezing, headache, fainting, or some of these or all of these reactions together. This is more likely when UP is a feature of systemic mastocytosis.

The four classical types of cutaneous mastocytosis are:

- Urticaria pigmentosa.

- Telangiectasia macularis eruptiva perstans. They look like persistent flat red and brown patches with visible blood vessels under the surface of the skin.
- Diffuse cutaneous mastocytosis. There is widespread involvement of the skin which may appear red, brown or similar to unaffected skin. It is rare and may start in young children and persist into adult life.
- Mastocytoma of skin usually presents in very young children, usually at birth or shortly afterwards, where mast cells form an itchy red or brown lump that often resolves completely as children grow up.

Some patients with urticaria pigmentosa may have accumulation of mast cells at sites other than the skin such as the bone marrow, gastrointestinal tract and or bones, known systemic mastocytosis. This is rarely associated with a blood disorder.

How is urticaria pigmentosa diagnosed?

The clinical appearance of the marks and the presence of Darier's sign are usually sufficient for a diagnosis of urticaria pigmentosa to be made by a dermatologist in children. However, skin biopsies may be carried out to exclude other skin conditions or changing moles.

Other tests that may be requested by your dermatologist include a blood test for full blood count, liver profile, vitamin D and tryptase, a bone marrow biopsy and a bone density scan called a DEXA. Some centres may have access to peripheral blood KIT mutation analysis on blood and this can be useful as a pointer to systemic mastocytosis when a bone marrow biopsy has not been done.

Can urticaria pigmentosa be cured?

No, there is no cure for urticaria pigmentosa. Childhood urticaria pigmentosa resolves by puberty in many cases. If urticaria pigmentosa started after the age of 10, there is a greater chance of progressing to systemic mastocytosis where mast cells may accumulate at sites other than the skin. There are medicines being developed for symptom control and advanced systemic mastocytosis.

How can urticaria pigmentosa be treated?

Symptomatic treatments include:

- Non-sedating *H1 antihistamines*: these are usually helpful for histamine-induced symptoms such as itching, flushing, reddening,

wheezing and diarrhoea. An old 'classical' antihistamine, *ketotifen*, has some additional anti-inflammatory properties that may provide additional benefit for some mastocytosis patients but may be sedative. Not all patients with mastocytosis feel a need to take antihistamines. *H2 antihistamines*, such as ranitidine and famotidine, may be useful for stomach acid symptoms.

- All adults should be offered at least one *adrenaline* auto-injector because anaphylaxis may happen unpredictably. Children with UP do not usually require an autoinjector unless they already have allergies or extensive skin lesions.
- *Oral sodium cromoglycate* can be helpful in some people by stabilising mast cells in the gut and thus reducing bowel symptoms but is not usually helpful for symptoms elsewhere.
- *Ultraviolet treatment can reduce itch and improve the skin's appearance*, but the benefits tend to be short-lived.
- *Potent steroid creams* can reduce itch and improve the skin's appearance, but you should discuss the risks versus benefits of long term use of potent steroid creams.
- *Laser therapy* has been used to improve the appearance of urticaria pigmentosa on selected areas. It is not known if the cosmetic improvement is long term and this treatment is not routinely available.

What can I do?

- Avoid situations that you know may trigger symptoms in your own case, such as alcohol, stress and extreme exercise. Hot baths or showers and vigorous towelling can cause histamine release from mast cells.
- If you or your child is given a prescription for a new medication, remind your doctor of the diagnosis of urticaria pigmentosa in case the drug has the potential to make the symptoms of urticaria pigmentosa worse.
- Over-the-counter preparations that contain codeine or aspirin may make the symptoms of urticaria pigmentosa worse.
- If a surgical operation is necessary, always ensure that your surgeon and anaesthetist are aware of the diagnosis of mastocytosis.
- Wearing a medical alert bracelet or necklace giving details of urticaria pigmentosa should be considered.
- Consider requesting referral to a specialist centre where dermatologists, haematologists and allergists work together to co-ordinate care and provide long term review when this is needed.

Where can I get more information about urticaria pigmentosa?

<http://www.ukmasto.co.uk>
<http://www.dermnetnz.org/colour/urticaria-pigmentosa.html>

For details of source materials used please contact the Clinical Standards Unit (clinicalstandards@bad.org.uk).

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.

This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel

**BRITISH ASSOCIATION OF DERMATOLOGISTS
PATIENT INFORMATION LEAFLET
PRODUCED SEPTEMBER 2008
UPDATED JANUARY 2012, JULY 2015, JUNE 2020
REVIEW DATE JUNE 2023**