



## **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 is the commonest type of a group of diseases called cutaneous mastocytosis, which has 3 other different types (see 'What are the symptoms of urticaria pigmentosa?' section).

Mastocytosis means increased number of mast cells. Mast cells are a type of blood cell belonging to our immune system, which secrete histamine if triggered. They can exist in the blood vessels or in any body tissue or organ. Urticaria pigmentosa is composed of pigmented brown patches, made of collections of mast cells that swell when rubbed similar to urticaria. In the majority of cases, urticaria pigmentosa is a harmless condition with excellent outcome.

More than 75% of cases of urticaria pigmentosa happen to infants and children less than 10 years old, but it can also affect older children and adults. It affects both sexes equally and occurs in all races equally.

### **What causes urticaria pigmentosa?**

The cause of urticaria pigmentosa is not known.

The release of histamine can cause a variety of symptoms ranging from an itch, hives (also known as nettle rash), wheezing, and diarrhoea, to, very rarely, life-threatening collapse. Certain factors can trigger the release of histamine and the other chemicals from mast cells, including drugs (especially aspirin and other non-steroidal anti-inflammatory drugs, codeine and

morphine, and some anaesthetics), alcohol, emotional stress, physical stimuli (heat, exercise and friction) and insect bites.

### **Is urticaria pigmentosa hereditary?**

No, it is not hereditary.

### **What does urticaria pigmentosa look like?**

Urticaria pigmentosa has a distinctive appearance consisting of brown or reddish-brown marks or swellings predominantly on the main trunk of the body and on the limbs. Underlying these marks are the small collections of mast cells. If the mark is firmly rubbed the mast cells release their chemicals and cause it to become itchy and swollen and, sometimes, blistered; this is known as Darier's sign and is very characteristic of urticaria pigmentosa.

### **What are the symptoms of urticaria pigmentosa?**

The marks that characterise urticaria pigmentosa may be considered unsightly, and they may be mistaken for changing moles. The itchy weals, i.e. hives (nettle rash), that develop if the marks are accidentally rubbed, may cause concern, and the itching is made worse by scratching. Occasionally, those with urticaria pigmentosa may develop allergic reactions, and sometimes anaphylaxis, if stung by insects, including bees and wasps.

The release of large amounts of histamine from mast cells may cause flushing or the skin becomes red, or a racing heartbeat, or diarrhoea, or wheezing, or headache, or fainting, or some of these or all of these reactions together. However, these symptoms are unusual in urticaria pigmentosa, and if they occur they might require some tests to exclude other types of mastocytosis.

Cutaneous mastocytosis consists of:

- Urticaria pigmentosa
- *Telangiectasia macularis eruptiva perstans*. This type looks different to urticaria pigmentosa and lasts longer.
- Diffuse cutaneous mastocytosis, where there are no defined areas, but the skin is diffusely affected. It is rare and more serious than urticaria pigmentosa.
- Mastocytoma of skin, where mast cells form an itchy lump that can resolve completely by time or removed surgically under local anaesthesia.

Although uncommon in young children, if urticaria pigmentosa is also associated with systemic mastocytosis (mastocytosis of internal organs and bone marrow), then this may interfere with the proper formation of blood and cause anaemia, and a decrease in bone density (osteoporosis) with some bone pain and fractures.

### **How is urticaria pigmentosa diagnosed?**

The clinical appearance of the marks and the presence of Darier's sign are usually sufficient to make the diagnosis of urticaria pigmentosa by a dermatologist. However, if there is any doubt, a skin biopsy of one of the marks may be indicated.

Other tests that are not routinely required but may be requested by your dermatologist are: full blood count, histamine level in blood and urine, tryptase enzyme in blood, bone marrow biopsy and bone scan.

### **Can urticaria pigmentosa be cured?**

No, there is no cure for urticaria pigmentosa. Childhood urticaria pigmentosa resolves by puberty in most cases. If urticaria pigmentosa started after the age of 10, the appearance and symptoms persist but tend to improve on their own with time, with a small chance to progress to systemic mastocytosis.

### **How can urticaria pigmentosa be treated?**

As urticaria pigmentosa is not serious, in the majority of cases no treatment is required. Treatments include:

- *Antihistamines as prescribed by your doctor* are usually helpful for histamine-induced symptoms such as itching, flushing, reddening, wheezing and diarrhoea.
- *Oral sodium cromoglycate* can be helpful in some cases by stabilising the mast cells and thus reducing histamine release in the gut.
- *Ultraviolet light treatment* can be helpful 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 any benefit has to be weighed against the potential for steroid preparations to damage the skin if used continuously.
- *Laser therapy* has been used to improve the cosmetic appearance of the rash of urticaria pigmentosa.

- If you or your child has reacted badly to an insect sting, your doctor may recommend that you carry a self-administered adrenaline pen to be used in the event of further stings.

### **What can I do?**

- Avoid things that you know may trigger symptoms in your own case, such as alcohol 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 cause histamine release. Over-the-counter preparations, such as cough linctus or treatments for diarrhoea (which may both contain codeine) may also 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 urticaria pigmentosa.
- Wearing a medical ID bracelet or necklace giving details of urticaria pigmentosa should be considered.

### **Where can I get more information about urticaria pigmentosa?**

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

<http://www.ukmasto.co.uk>

For details of source materials used please contact the Clinical Standards Unit ([clinicalstandards@bad.org.uk](mailto: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  
REVIEW DATE JULY 2018**



