



## 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 clinical form of a group of conditions known collectively as *mastocytosis*, in which mast cells proliferate and infiltrate, to a varying degree, certain tissues of the body. Mast cells are a type of cell characterised by containing a number of inflammation-provoking chemicals, which include histamine, and the release of these chemicals can cause a variety of symptoms ranging from itch and hives (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), bacterial toxins and insect venom.

In urticaria pigmentosa, small collections of mast cells occur primarily in the skin, but other organs (such as the gut, lungs and bone) may rarely be involved. It most commonly affects young children, but may arise in adulthood.

### What causes urticaria pigmentosa?

The cause of urticaria pigmentosa is not known for certain. However, there is recent evidence to suggest that a genetic change in a protein (called c-kit) on the surface of mast cells may result in the abnormal proliferation of these cells.

## **Is urticaria pigmentosa hereditary?**

Despite a possible underlying genetic cause, urticaria pigmentosa does not appear to be inherited from one generation to the next, and brothers and sisters are not usually affected.

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

Urticaria pigmentosa has a distinctive appearance consisting of brown or reddish-brown freckles, predominantly on the trunk and limbs. Underlying these freckles are the small collections of mast cells, and if the freckle is firmly rubbed the mast cells release their chemicals and cause the site of the freckle to become itchy and swollen and, sometimes, blistered: this is known as Darier's sign and is very characteristic of urticaria pigmentosa.

Some adults have a prominent degree of dilated small blood vessels associated with the pigmented spots, a variant of urticaria pigmentosa called *telangiectasia macularis eruptiva perstans*.

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

The freckles that characterise urticaria pigmentosa may be considered unsightly, and they may be mistaken for changing moles. The itchy weals (hives or nettle rash) that develop if the freckles are accidentally rubbed may cause concern, and the itching is worsened by scratching.

The release of large amounts of histamine from mast cells may cause flushing, a racing heartbeat, diarrhoea, wheezing, headache and fainting, although these symptoms are unusual in urticaria pigmentosa, tending to occur in the more severe forms of mastocytosis.

Occasionally, urticaria pigmentosa sufferers may develop severe allergic reactions, and sometimes anaphylaxis, if stung by certain insects, such as bees and wasps.

Very rarely, involvement of the bone marrow in adults with urticaria pigmentosa may interfere with the proper formation of blood and cause anaemia, and a decrease in bone density (osteoporosis) may result in bone pain and fractures.

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

The clinical appearance of the freckles and the presence of Darier's sign are usually sufficient to make the diagnosis of urticaria pigmentosa. However, if there is any doubt, your dermatologist may suggest a skin biopsy of one of the freckles to look for an increased number of mast cells.

Normally, no further tests are required, but if there are any symptoms to suggest mast cell accumulation in organs other than the skin your doctor may suggest blood or urine tests and, perhaps, X-rays.

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

No, there is no cure for urticaria pigmentosa. Childhood urticaria pigmentosa usually, but not always, resolves by adolescence. In adults, the appearance and symptoms tend to improve with time, although rarely it may progress to a more severe form of mastocytosis.

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

Generally, urticaria pigmentosa is not serious, and it is often the case that no treatment is required.

*Antihistamines* are usually helpful for histamine-induced symptoms such as itching, flushing, wheezing and diarrhoea.

*Oral disodium cromoglycate* can be helpful in some cases by stabilising the mast cells and thus reducing histamine release.

*Ultraviolet light treatment* can be helpful and improve the appearance, but the benefits tend to be short-lived.

*Potent steroid creams* can reduce itch and improve appearance, but any benefit has to be weighed against the potential for steroid preparations to damage the skin if used in the long-term.

*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 kit such as an EpiPen® to be used in the event of further stings. Desensitisation (reducing sensitiveness) to the venom of the insect may be possible.

### **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, as may immersion in very cold water.
- 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 MedicAlert® bracelet or necklace giving details of urticaria pigmentosa should be considered.

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

[http://dermnetnz.org/colour/urticaria\\_pigmentosum.html](http://dermnetnz.org/colour/urticaria_pigmentosum.html)

[http://en.wikipedia.org/wiki/Urticaria\\_pigmentosa](http://en.wikipedia.org/wiki/Urticaria_pigmentosa)

<http://www.mastokids.org>

[www.ukmasto.co.uk](http://www.ukmasto.co.uk)

**(Whilst every effort has been made to ensure that the information given in this leaflet is accurate, not every treatment will be suitable or effective for every person. Your own doctor will be able to advise in greater detail.)**

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