



URTICARIA AND ANGIOEDEMA

What are the aims of this leaflet?

This leaflet has been written to help you understand more about urticaria and angioedema. It tells you what they are, what causes them, what you can do about them, and where you can find out more about them. The first part of this leaflet deals mainly with ordinary urticaria and angioedema - the most common form. Other types, and the ways in which they differ from ordinary urticaria, are described in the second part of the leaflet.

What are urticaria and angioedema?

- Urticaria is common, and affects about 20% of people at some stage of their lives. It is also known as hives, nettle rash, or welts. The short-lived swellings of urticaria are known as weals (see below).
- Angioedema is a deeper form of urticaria.

An affected individual may have urticaria alone, or angioedema alone, or both together. The most common form is called 'ordinary urticaria', which is usually divided into 'acute' and 'chronic' forms. In 'acute' urticaria/angioedema, the bout lasts from a few days up to six weeks. Chronic urticaria, by definition, lasts for more than six weeks.

Other less common types – such as the physical urticarias - are described later in this leaflet. They include urticarial vasculitis (in which urticaria is due to an inflammation of the blood vessels) and contact urticaria (in which substances, such as fruit, nuts or rubber, cause urticaria when they are applied to the skin).

What causes urticaria and angioedema?

Both are caused by the release of histamine from mast cells (allergy cells) within the skin. This can be triggered in many ways, for example by exercise, by pressure on the skin, and by other physical factors as well as by foods, drugs and infections. However in the common 'ordinary' form of urticaria and angioedema, it is unusual for an external cause to be identified. In some patients with ordinary chronic urticaria, the release of histamine from skin mast cells is triggered by factors circulating in the blood, such as antibodies directed against their own mast cells - a process known as autoimmunity. Tests for this are not routinely available, and generally do not alter the treatments used.

Often no cause for acute urticaria can be found, but sometimes it may be caused by infections such as a cold, 'flu or a sore throat. Almost any medicine can cause 'acute' urticaria but painkillers (especially aspirin and medicines like ibuprofen), antibiotics (especially penicillins), and vaccinations, are most likely to be responsible. Angioedema is particularly caused by a type of drug used to treat high blood pressure (ACE inhibitors). Rarely foods are responsible, including nuts, fish, tomatoes, vegetables and berries.

What are the symptoms of urticaria and angioedema?

The main symptom of urticaria is itching: angioedema, however, may not be itchy. Although urticaria can be distressing, because of itching and its appearance, it has no direct effect on general health.

Are urticaria and angioedema hereditary?

The 'ordinary' common type of urticaria and angioedema is not hereditary.

What do ordinary urticaria and angioedema look like?

The weals of urticaria may be pale, pink or red, and may look like nettle stings. They can be of different shapes and sizes; often they are surrounded by a red flare. They are usually itchy. As the raised weals flatten, they leave red marks that usually disappear in a day. New weals may then appear in other areas. In ordinary urticaria the weals can occur anywhere on the body, at any time, and usually fade within a day.

The pale or pink, deeper swellings of angioedema occur most frequently in the eyelids, lips and sometimes in the mouth. They may not be itchy, and usually settle in a few days. If the hands are affected, they may feel tight and painful.

How will ordinary urticaria be diagnosed?

Usually its appearance, or a description of it, will be enough for your doctor to make the diagnosis. In the vast majority of people no cause can be found, though your doctor will ask you questions to try to identify one. There is no special test that can reliably identify the cause of urticaria, but some tests may be done if your answers suggest an underlying cause.

- In acute urticaria, investigation is usually unnecessary. Occasionally, if an allergic reaction is suspected, a specific blood test for allergic sensitisation (RAST), or a skin prick test may be performed. These and other tests should be carried out by a specialist in skin or allergic disease.
- In chronic urticaria, it is rare for allergy to be the cause, so routine allergy tests such as skin prick tests or RAST are not necessary. In a small percentage of people, foods, colouring agents and preservatives appear to worsen the urticaria. A food diary can then be kept: these substances can be left out of the diet to see if the condition improves, and later deliberately reintroduced. However, as urticaria is such a variable disease, the interpretation of these elimination diets is difficult.

Can ordinary urticaria and angioedema be cured?

The treatments outlined below suppress the condition rather than cure it. In half of the people with chronic ordinary urticaria, the rash lasts for 6-12 months, and then gradually disappears. It usually does not return. However in any one individual the course of urticaria is unpredictable.

What is the treatment for ordinary urticaria?

- It is important to avoid *anything that may worsen urticaria*. These are listed below in detail under the heading 'What can I do?'
- *Antihistamines* block the effect of histamine, and reduce itching and the rash in most people, but may not relieve urticaria completely. If urticaria occurs frequently, it is best to take antihistamines regularly. There are many different types. The older ones often cause drowsiness. The newer ones are much less likely to cause drowsiness, but may do so if taken with alcohol. No particular antihistamine is best for everyone, so your doctor may need to try different ones to find the one that suits you best. Antihistamine tablets may need to be taken for as long as the urticaria persists. Reports of serious side effects are rare, but occasionally a few cause weight gain, and some should not be taken at the same time as particular medicines.
- *A related type of antihistamine* (e.g. cimetidine and ranitidine), which is usually used to treat stomach ulcers, can be added to the standard antihistamines used to treat the skin.
- If antihistamine tablets are not helpful you can discuss this with your doctor who may arrange further tests, and try *other medicines*. Some of these may not be licensed for urticaria, but can be useful treatments.

- *Oral steroids* may occasionally be given briefly for severe flares of acute and chronic urticaria, but generally are not necessary.
- *New treatments that act by suppressing the immune system* (e.g. ciclosporin) are being used in a few of the most severely affected people in specialist skin and allergy centres, and may be beneficial.
- Tongue or throat swelling is an unusual but alarming sign of angioedema, which is rarely life threatening except in acute food or medicine allergies and the rare hereditary form of angioedema. *Injections of adrenaline* (epinephrine) (which can be self administered) often provide rapid relief.

What can I do?

It is important to avoid anything that may worsen urticaria - such as heat, tight clothes, alcohol, aspirin and aspirin containing compounds, and if possible other similar medicines such as Nurofen (paracetamol does not usually cause a problem). Medicines called ACE inhibitors (often used to treat high blood pressure) should be avoided, especially if angioedema is present. Foods, colouring agents and preservatives can be avoided in the rare instances where these have proved to be a problem.

Other urticarias

- ***The physical urticarias.*** Other forms of urticaria are triggered by physical factors such as heat, cold, friction, pressure on the skin and even by water. The weals usually occur within minutes, and last for less than one hour (except delayed pressure urticaria). Physical urticarias usually occur in healthy young adults, and are not uncommon. They may occur in association with ordinary urticaria, or with each other, and tend to improve with time. They include the following types:

Dermographism (“*skin writing*”). In this type, itching weals occur after friction such as rubbing or stroking the skin, which is generally very itchy especially when hot. Weals and red marks often appear as lines at the sites of scratching, and generally last for less than one hour. Usually no cause is found.

Cold urticaria. Cold, including rain, wind and cold water, causes itching and wealing in chilled areas. Swimming in cold water may cause severe wealing and fainting, and must be avoided. Patients should report their cold urticaria to medical personnel before operations so that, if weals appear during the procedure, cold urticaria can be considered. Usually no cause can be found for cold urticaria.

Solar urticaria. This is rare. Redness, itching and weals occur on the skin immediately after exposure to sunlight, and last for less than one hour.

Aquagenic urticaria. This is extremely rare. Small weals occur on the skin at the site of contact with water of any temperature, usually on the upper part of the body.

Delayed pressure urticaria. Swellings occur at skin sites to which pressure has been applied, for example from tight clothes or from gripping tools. Usually the swelling develops several hours later. It can be painful and last longer than a day. People with pressure urticaria nearly always have ordinary urticaria as well.

Many of the physical urticarias are improved by avoiding their cause, and regular treatment with antihistamines. However antihistamines do not usually help delayed pressure urticaria. Sometimes a short course of oral steroids will help if the symptoms of delayed pressure urticaria are very severe.

- ***Cholinergic urticaria.*** This occurs under conditions that cause sweating, such as exertion, heat, emotional stress and eating spicy food. Within minutes, small itchy bumps with variable redness appear, usually on the upper part of the body but they may be widespread. The weals last for less than one hour, but in severe cases may join together to form larger swellings. Antihistamines usually help, and are sometimes best taken before a triggering event (e.g. exercise).
- ***Urticarial vasculitis.*** A small percentage of people with urticaria develop weals that last longer than two days. These may be tender and occasionally bruise. People affected with this condition may feel unwell and have joint and stomach pains. This is because their blood vessels become inflamed (a process known as vasculitis). The diagnosis is confirmed by examining under the microscope a small piece of a weal that has been removed. The cause is rarely found, though blood tests are usually undertaken. Antihistamines are not very helpful but other medicines that help inflammation can be used.
- ***Contact urticaria.*** Various chemicals, foods, plants, animals, and animal products, can cause weals within minutes at the site of contact. These weals do not last long. Some of the commoner causes are eggs, nuts (e.g. peanuts), citrus fruits, rubber (latex) and contact with cats and dogs. Although often the reactions are mild, occasionally they can be severe – for example after contact with rubber and peanuts in very sensitive individuals.

- **Angioedema without weals.** Angioedema occurring without urticaria can be due to a variety of causes such as medicines (e.g. aspirin, ACE inhibitors), or food allergies. Most commonly it is a component of chronic ordinary urticaria/ angioedema, where no cause can be identified.
- **Hereditary angioedema.** This is a very rare form, which tends to run in families. Patients get swellings of the face, mouth, throat, and sometimes of the gut, leading to colic. The condition is due to an inherited deficiency of a blood protein and can be identified by a blood test. It can be treated by medicines to prevent attacks and sometimes by replacing the deficient protein in the blood in an acute attack. A severe attack of hereditary angioedema can be life threatening if left untreated; therefore patients may be advised to wear a Medic Alert bracelet (for address see below) to alert physicians in an emergency.

Where can I find out more about urticaria?

Address of the Medic Alert Foundation:

1 Bridge Wharf
156 Caledonian Road, London N1 9UU
Tel: (020) 7833 3034
Freephone 0800 581 420

Web links to detailed leaflets:

www.dermnet.org.nz/dna.urticaria/urt.html

(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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