



URTICARIA AND ANGIO-OEDEMA

What are the aims of this leaflet?

This leaflet has been written to help you understand more about urticaria and angio-oedema. It tells you what they are, what causes them, what you can do about them and where you can find out more about them.

What is urticaria?

- Urticaria is a common illness affecting up to 20% of people (one in 5 people) at some point in their lives. Urticaria presents with highly itchy raised skin reactions known as weals (also known as hives) that may be round or ring-shaped, and may join together. They come and go within hours. Weals can also appear as raised lines after scratching. They can appear anywhere on the skin. Individual weals typically disappear of their own accord within 24 hours without a trace but the condition lasts longer. Angio-oedema, swelling deep to the skin, often occurs in urticaria. Angio-oedema usually affects soft areas of skin, such as the eyelids, lips or inside the mouth but may occur anywhere. These swellings often take longer to clear and tend to be painful rather than itchy. Urticaria may present with weals alone, angio-oedema or both together.
- If angio-oedema occurs without weals it may be an inherited illness called hereditary angio-oedema. This is a different problem to urticaria. It can be diagnosed with blood tests and needs different treatment.

The most common form of urticaria is called spontaneous urticaria. Spontaneous urticaria is usually divided into 'acute' and 'chronic' forms. In 'acute' urticaria, the episode lasts up to six weeks. Chronic or long-lasting urticaria lasts for six weeks or more.

What causes urticaria?

Urticaria is caused by the release of histamine and other chemicals from cells in the skin called mast cells. Urticaria is often thought of as an allergy but, in fact, it usually results from histamine release from mast cells due to other reasons.

Allergens in food, or medicine may sometimes cause **acute** urticaria. For young babies, in whom urticaria is rare, cow's milk allergy is the commonest cause. As children grow up, they may react to different foods, including nuts, fruits or shellfish if they become allergic. Bee and wasp stings can cause acute urticaria.

However, a specific reason for urticaria is often not found. Chronic spontaneous urticaria may be autoimmune, the patient's own antibodies that release histamine from mast cells. Tests for autoimmune urticaria are not routinely available and generally do not alter treatment. When a cause cannot be found, it is called 'idiopathic'. Weals may be set off by a physical trigger, such as cold, pressure or friction. This type of urticaria is called inducible because hives are induced by a specific stimulus and are not spontaneous.

Some people with urticaria have other conditions, such as thyroid disease or other autoimmune disorders.

If you suspect that a medicine may have caused urticaria, you should inform your doctor. You or your doctor can complete a 'yellow card' to inform the Medicines and Healthcare Regulatory Authority (MHRA) (<https://yellowcard.mhra.gov.uk/>). Almost any medicine can cause urticaria, but painkillers such as aspirin, ibuprofen and codeine, some antibiotics, blood products such as blood transfusions and vaccinations are most likely to be responsible. Angio-oedema without weals may be caused by a group of drugs called ACE inhibitors used to treat high blood pressure.

What are the symptoms of urticaria and angio-oedema?

The main symptom of urticaria is itch; angio-oedema is not usually itchy but may be painful. Although urticaria may be very distressing because of the itching and its appearance, it generally has no direct effect on general health. However, it can cause a high impact on quality of life, including mood, and lead to problems with work and relationships. Rarely, angio-oedema may affect the tongue or throat. This can be alarming but is only life-threatening when angio-oedema is due to hereditary angio-oedema or ACE inhibitor medication.

Is urticaria hereditary?

The vast majority of urticarias are not hereditary. Hereditary angio-oedema runs in families.

How is chronic urticaria diagnosed?

Usually its appearance, or a description of it, will be enough for your doctor to make the diagnosis. There is no special test that can reliably identify the cause of urticaria, and most people with spontaneous urticaria who respond well to antihistamines do not need any tests at all. Routine allergy tests are not necessary.

In a small percentage of people, foods, food colouring agents and preservatives appear to worsen urticaria, and it is occasionally possible to identify these by keeping a food diary. These substances can be left out of the diet to see if the condition improves, and later reintroduced to confirm whether they are the cause of the urticaria or not. However, as urticaria is such a fluctuating disease, this is not always accurate and will not always show what is causing the problem.

Can urticaria be cured?

The aim of treatment is to suppress the symptoms of the condition until it becomes better naturally rather than cure it. In about half of the people affected by chronic spontaneous urticaria, the rash lasts for 6-12 months, and then gradually disappears. It can however last considerably longer. In any one individual the course of urticaria is unpredictable.

What is the treatment of urticaria?

- *Antihistamine tablets* block the effect of histamine by reducing itch and the rash in most people but may not relieve urticaria completely. If urticaria occurs frequently, antihistamines should be taken regularly every day. There are many different types, including non-sedating and sedating antihistamines, as well as short acting and long acting types. They are known as H1 antihistamines. You and your doctor may need to try different ones to find a regime that suits you best. It has become common practice to increase the daily dose of non-sedating H1 antihistamines up to fourfold if necessary to control symptoms. They can be taken for as long as the urticaria persists. Even the non-sedating types can

- occasionally make people sleepy, especially with higher doses. As with all medications there can be side effects; the balance of risk and benefit needs to be considered when taking these and all treatments. Some can be bought over the counter; discuss with the pharmacist any potential side effects and possible interactions with other medications you are taking.
- *A different type of antihistamine called H2 antihistamines* (e.g. cimetidine, famotidine and ranitidine) that are usually used to treat stomach acid symptoms, can be added if standard H1 antihistamines are insufficient on their own.
 - If antihistamine tablets are not helpful, your doctor may recommend *other medicines*, including montelukast, that is usually used as an add-on treatment of asthma.
 - *Oral steroids* can occasionally be given as rescue treatment for severe flares of acute and chronic urticaria but are generally not necessary; their potential side effects usually outweigh the benefits in this condition.
 - *Treatments that act by suppressing the immune system* (e.g. ciclosporin) may be beneficial for the most severely affected people not responding to the treatments outlined previously, especially those with autoimmune urticaria.
 - Very rarely *injections of adrenaline* (epinephrine) may be required if there are breathing problems caused by angio-oedema.
 - A biological injection called omalizumab has been approved for severely affected chronic spontaneous urticaria patients in whom other treatments have been ineffective. This is only available in specialist clinics.

Self-care (what can I do?)

- Do not take aspirin, ibuprofen (and other non-steroidal anti-inflammatory drugs) or opiates, such as codeine unless it is essential, since these medicines may aggravate urticaria.
- Avoid anything that may worsen urticaria, such as heat, tight clothes, and alcohol. Triggers vary between individuals.
- Avoidance of specific foods, colouring agents and preservatives may be helpful.
- Seek medical advice urgently if you are having problems with breathing or swallowing.

Other urticarias

In some patients, clear trigger factors for urticaria can be identified although the cause remains unknown; these are called inducible urticarias. There are several types of inducible urticaria.

Inducible urticarias: weals may be triggered by 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, which is delayed in onset and lasts for hours at least). Physical urticarias often occur in otherwise healthy young adults but can affect any age. Some patients suffer from more than one type of urticaria; they include the following types:

- **Symptomatic dermographism (“skin writing”).** In this type, itchy weals occur after friction, such as rubbing or stroking the skin. Weals and red marks often appear as lines at the sites of scratching and generally last for less than an hour.
- **Cold urticaria.** This is triggered by exposure to cold, including rain, wind and cold water, causing itchy weals in chilled areas. Swimming in cold water may cause extensive wealing with a risk of fainting and should be avoided. Patients should report cold urticaria to medical personnel before operations so they can be kept warm during the procedure.
- **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 after avoidance of exposure.
- **Aquagenic urticaria.** This is extremely rare. Small weals occur on the skin at the site of contact with water, usually on the upper part of the body.
- **Delayed pressure urticaria.** Urticaria develops where pressure has been applied to the skin, for example from tight clothes or from gripping tools. Usually the swelling develops several hours later. It can be painful and lasts longer than a day. People with delayed pressure urticaria nearly always have spontaneous urticaria as well.
- **Cholinergic urticaria** - This occurs under conditions that cause sweating, such as exertion, heat, emotional stress or eating spicy food. Within minutes, small itchy bumps with redness appear, usually on the upper part of the body but they may be widespread. The weals last for less than an hour, but in severe cases may join together to form larger swellings. Antihistamines usually help. They may be taken an hour or two before a triggering event (e.g. exercise) if these are infrequent.
- **Contact urticaria** - Various chemicals, foods, plants, animals, and animal products, can cause weals within minutes at the site of contact. These do not last long. Some of the more common causes are eggs, nuts (e.g.

peanuts), citrus fruits, rubber (latex) and contact with cat and dog saliva. Reactions are occasionally severe, for example after contact with rubber and peanuts in very sensitive individuals.

Other illness presenting with urticarial rash or swelling:

- **Angio-oedema without weals** - angioedema occurring without weals can be due to medicines (e.g. aspirin, ACE inhibitors). When angio-oedema occurs without weals, hereditary angio-oedema should be excluded by testing.
- **Hereditary angio-oedema** - this is a very rare form of angio-oedema, which usually runs in families because it is due to a genetic mutation. Patients have intermittent swelling 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 or a non-functional version of C1 inhibitor 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 during an acute attack. A severe attack of hereditary angio-oedema affecting the throat can be life threatening if left untreated; therefore, patients may be advised to wear Medic Alert bracelet or an alert on their mobile phone for an emergency.
- **Urticarial vasculitis** - a very small percentage of people with an urticarial rash develop weals that last longer than 24 hours. These may be tender and occasionally bruise. People affected with this condition may feel unwell with joint and stomach pains. This is because their blood vessels become inflamed (a process known as vasculitis). The diagnosis is confirmed by examining a skin biopsy from one of the weals under the microscope. The cause is rarely found, though extensive blood tests should be undertaken. Antihistamines are not very helpful but other medicines that help inflammation can be used.

Where can I find out more about urticaria?

Links to patient support groups:

Allergy UK
Planwell House
LEFA Business Park
Edgington Way
Sidcup, Kent

DA14 5BH

www.allergyuk.org

Tel: 01322 619898

Web links to detailed leaflets:

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

<https://www.allergyuk.org/information-and-advice/conditions-and-symptoms/416-urticaria-hives-and-other-skin-allergy>

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

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