Lee Clinic Dermatology Information Leaflet

Urticaria & Angioedema

What is urticaria and angioedema?

- Urticaria is common, and affects about 20% of people at some point in their lives. It is also known as hives or nettle rash. The short-lived swellings of urticaria are known as weals (see below) and typically any individual spot will clear within 24 hours although the overall rash may last for longer.
- Angioedema is a form of urticaria in which there is deeper swelling in the skin, and the swelling may take longer than 24 hours to clear.

An affected individual may have urticaria alone, angioedema alone, or both together. Both are caused by the release of histamine from cells in the skin called mast cells. When angioedema occurs in association with urticaria, the two conditions can be considered part of the same process. When angioedema occurs on its own, different causes need to be considered.

,p>There are different types of urticaria of which the most common form is called ‘ordinary or idiopathic urticaria’. In this type no cause is usually identified and often patients have hives and angioedema occurring together. Ordinary urticaria with or without angioedema is usually divided into ‘acute’ and ‘chronic’ forms. In ‘acute’ urticaria/angioedema, the episode lasts from a few days up to six weeks. Chronic urticaria, by definition, lasts for more than six weeks. Ongoing ordinary urticaria should be differentiated from an isolated individual episode of acute urticaria which occurs for example with ingestion of or contact with an allergen.

Other less common types of urticaria are described later in this leaflet. Also included is urticarial vasculitis (in which inflammation of the blood vessels causes an urticarial-like rash, and is therefore different from normal urticaria).

What causes urticaria and angioedema?

In the common ‘ordinary’ form of urticaria and angioedema, it is unusual for an external cause to be identified. Intercurrent infections such as a cold, influenza or a sore throat may act as a trigger.

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, in particular, can be caused by a type of drug (ACE inhibitors) used to treat high blood pressure.

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.

What are the symptoms of urticaria and angioedema?

The main symptom of urticaria is itch but angioedema is not usually itchy. Although urticaria can be distressing, because of the itching and its appearance, it has no direct effect on general health. Rarely, the swelling of angioedema may affect the tongue or throat, causing difficulty with breathing or swallowing. This can be alarming but is rarely life-threatening.

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 flesh-coloured, pink or red. They can be of different shapes and sizes, but usually look like nettle stings. An important feature of urticaria is that although the rash may persist for weeks, individual lesions usually disappear within a day, and often last only a matter of hours. However, they sometimes leave bruising especially in children. New weals may then appear in other areas. In ordinary urticaria, the weals can occur anywhere on the body, at any time.

The deeper swellings of angioedema occur most frequently on the eyelids, lips and sometimes in the mouth, but they may occur anywhere. They are not usually itchy, and tend to settle within a few days. If the hands and feet 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 ordinary (idiopathic) urticaria, but some tests may be done if your answers suggest an underlying cause, which may be suspected from a detailed history.

Occasionally, if a trigger is suspected, blood tests or a skin prick test may be performed by a specialist in skin or allergic disease. In chronic urticaria, no specific trigger is usually identified so routine allergy tests are not necessary. In a small percentage of people, foods, colouring agents and preservatives appear to worsen urticaria, and it might be helpful 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. However, as urticaria is such a fluctuating disease, this is not always accurate and will not always show you definitely what is causing the problem.

Can ordinary urticaria and angioedema be cured?

The treatments outlined below suppress the condition rather than cure it. In about half of the people affected by chronic ordinary 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 for ordinary urticaria?

- 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 every day. There are many different types including non sedating and sedating

antihistamines, in addition to short acting and long acting types Your doctor may need to try different ones to find a regime that suits you best. The antihistamine tablets can to be taken for as long as the urticaria persists.
- 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, your doctor may recommend other medicines. These may include other types of histamine blockers, or drugs such as montelukast, which is a treatment for asthma and although they may not be licensed for urticaria, they can be useful treatments
- Oral steroids can occasionally be given briefly for severe flares of acute and chronic urticaria, but generally are not necessary.
- 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.
- Very rarely injections of adrenaline (epinephrine) may be required if there are breathing problems.

Self care (What can I do?)

- It is important to avoid anything that may worsen urticaria, such as heat, tight clothes, and alcohol.
- Avoidance of foods, colouring agents and preservatives may be helpful in the rare instances where these have proved to be a problem.
- Seek medical advice urgently if you are having problems with breathing or swallowing.

Other urticarias In some patients, when a detailed history is taken, clear trigger factors for urticaria/angioedema can be identified. These may include physical factors (called physical urticaria) or foods, drugs and infections.

The physical urticarias - Urticaria may be 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. Some patients suffer from more than one type of urticaria; they include the following types:

Dermographism ("skin writing"). In this type, itchy weals occur after friction such as rubbing or stroking the skin. Itch may be aggravated by heat. Weals and red marks often appear as lines at the sites of scratching, and generally last for less than one hour.

Cold urticaria. This type is precipitated by exposure to 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 should 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.

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. 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 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 trigger, and by taking regular treatment with antihistamines. Delayed pressure urticaria can be more difficult to treat.

Cholinergic urticarial. 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).

Contact urticarial. 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. When angioedema occurs without associated urticaria, a hereditary form of angioedema should be considered.

Hereditary angioedema - This is a very rare form of angioedema which tends to run in families. Patients get 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 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 to alert physicians in an emergency.

Urticarial vasculitis - A small percentage of people with urticaria develop weals that last longer than one day. 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 a skin biopsy from one of the weals under the microscope. 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.

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