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.

What are urticaria and angioedema?

- Urticaria, also known as hives, is common, and affects about 20% of people (one in 5 people) at some point in their lives. Urticaria consists of pink or white raised areas of skin resembling nettle rash, known as wheals (also spelled weals), which are usually itchy. The wheals are often round or ring-shaped. Wheals can also appear as lines when the skin is firmly stroked. They can appear anywhere on the skin. Individual wheals typically disappear of their own accord within 24 hours without a trace, although the course of the condition is longer.
- Angioedema is characterised by deeper swelling in the skin, which may take over 24 hours to clear. It is not usually itchy and can affect the lips and tongue. Some patients have one or the other condition, others have both.

The most common form of urticaria is called spontaneous urticaria. In this type no cause is usually identified and often patients have hives and angioedema occurring together. Spontaneous urticaria with or without angioedema is usually divided into 'acute' and 'chronic' forms. In 'acute' urticaria, the episode lasts up to six weeks. Chronic urticaria, by definition, lasts for more than six weeks.

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Other types of urticarial rash are described later in this leaflet, including *urticarial vasculitis* (in which there is inflammation of the blood vessels and is therefore different from normal urticaria).

What causes urticaria and angioedema?

Urticaria is caused by the release of histamine from cells in the skin called mast cells.

Often a specific cause cannot be found. Sometimes an infection such as a cold can be a trigger. Other triggers include physical contact with an allergen such as an animal, sun exposure or a specific food or medicine. For young babies, in whom it is rare, cow's milk allergy is the commonest trigger. Bee and wasp stings can trigger urticaria, as can eating shellfish, nuts, apples and peaches.

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 (especially aspirin and medicines like ibuprofen), antibiotics (especially penicillins), blood products 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.

Some people with urticaria have conditions such as pernicious anaemia or thyroid disease, caused by the immune system directing an attack against the patient's own tissues.

In some patients with chronic spontaneous 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.

Urticaria is often thought to be due to allergy, but in fact, allergy is not a common cause of urticaria.

What are the symptoms of urticaria and angioedema?

The main symptom of urticaria is itch; angioedema is not usually itchy but may be painful. 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.

Is urticaria hereditary?

The vast majority of urticaria is not hereditary. A rare form of angioedema, (hereditary angioedema) and some very rare urticaria syndromes can run in families.

What does urticaria look like?

The wheals of urticaria may be white, pink or red. They can be of different shapes and sizes, and often look like nettle stings. Although the rash may persist for many weeks or months, individual lesions typically disappear within a day, and often last only a few hours. They occasionally leave bruising especially in children. New wheals may then appear in other areas. In spontaneous urticaria, wheals 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 last a few days. The skin may feel tight and painful.

How is chronic urticaria 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, and most people with spontaneous urticaria do not need any tests at all.

Occasionally, if a trigger is suspected, a specific blood test, to detect antibodies in the bloodstream, or a skin prick test may be performed by a Dermatologist or Allergist in skin or allergic disease. In chronic urticaria routine allergy tests are not necessary. In a small percentage of people, foods, food 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 urticaria be cured?

The treatments outlined below suppress the symptoms of 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 of urticaria?

- *Antihistamine tablets* 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 can be helpful to take antihistamines regularly every day. There are many different types including non-sedating and sedating antihistamines, as well as short acting and long acting types. You and 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. Even the non-sedating types can make some people sleepy, and 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 other medications you are taking.
- *A different type of antihistamine* (e.g. cimetidine and ranitidine), which is usually used to treat stomach inflammation, can be added if the standard antihistamines alone are insufficient. These can be bought over the counter but are best used in discussion with your doctor who can prescribe them.
- If antihistamine tablets are not helpful, your doctor may recommend *other medicines.*
- *Oral steroids* can occasionally be given briefly for severe flares of acute and chronic urticaria, but are generally not necessary; the 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.
- Very rarely *injections of adrenaline* (epinephrine) may be required if there are breathing problems caused by angioedema.

- A new biological injection treatment called omalizumab has recently been approved for the most severely affected chronic spontaneous urticaria patients. This is only available in specialist clinics.

Self care (What can I do?)

- If possible, do not take the medicines codeine (and other opiate drugs) or aspirin, ibuprofen and naproxen (and other non-steroidal anti-inflammatory drugs).
- It is important to 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 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, clear trigger factors for urticaria can be identified; these are called inducible urticarias. There are several types of inducible urticaria.

Physical urticarias - Urticaria may be triggered by heat, cold, friction, pressure on the skin and even by water. The wheals 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 wheals occur after friction such as rubbing or stroking the skin. Itch may be aggravated by heat. Wheals 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 wheals chilled areas. Swimming in cold water may cause severe whealing and fainting, and should be avoided. Patients should report their cold urticaria to medical personnel before operations so that, if wheals appear during the procedure, cold urticaria can be considered.

- ***Solar urticaria.*** This is rare. Redness, itching and wheals 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 wheals 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 last longer than a day. People with pressure urticaria nearly always have ordinary urticaria as well.
- **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 wheals last for less than an 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 urticaria** - Various chemicals, foods, plants, animals, and animal products, can cause wheals within minutes at the site of contact. These 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.

Many spontaneous urticarias are improved by avoiding their trigger, and by taking regular antihistamines. Delayed pressure urticaria can be more difficult to treat. Sometimes a short course of oral steroids will help if the symptoms of delayed pressure urticaria are very severe.

- **Angioedema without wheals** - Angioedema occurring without urticaria can be due to medicines (e.g. aspirin, ACE inhibitors) or food allergies. When angioedema occurs without wheals, 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 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 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 an urticarial rash develop wheals that last longer than 24 hours. 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 wheals 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.

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
www.allergyuk.org/skin-allergy/urticaria-and-angiodema
www.nhs.uk/Conditions/Nettle-rash/Pages/Introduction.apsx

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

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