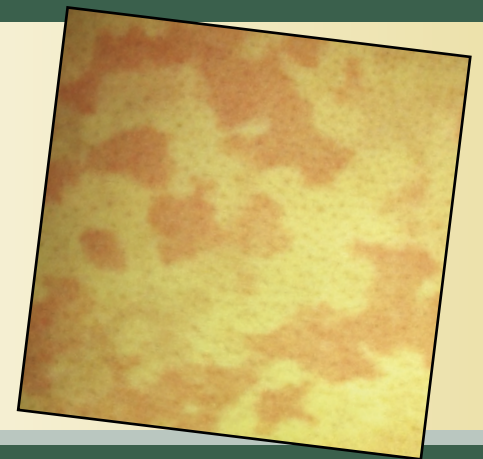


What you should know about

Urticaria and Angioedema



Published by the Department of Immunology
St James's Hospital, Dublin 8, Ireland

First publication: July 2015

This booklet may be downloaded in .pdf format from
the St James's Hospital website: please visit www.stjames.ie

Guidelines for patients

• Symptoms • Diagnosis • Treatment

What you should know about

Urticaria and Angioedema

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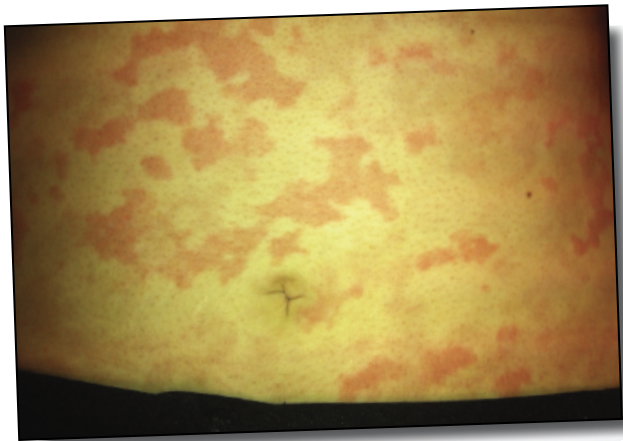
• Symptoms • Diagnosis • Treatment

Urticaria is a medical condition commonly known as hives; it is frequently associated with skin swelling, termed **angioedema**.

This patient booklet is to help you develop an understanding of chronic or recurring urticaria and angioedema.

The booklet aims:

- to describe the symptoms associated with urticaria and angioedema
- to discuss reasons why you develop urticaria and angioedema
- to describe treatments that may be used to control these conditions successfully
- to answer frequently-asked questions.



Urticaria: red, itchy blotches on the skin, sometimes said to look like “nettle rash”

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1 Urticaria

Urticaria is the medical term for **hives**, a common skin condition.

Most people will have an outbreak of hives at some point in their lives and the rash consists of red, itchy blotches, sometimes described as being like a “nettle rash”. Urticaria is often accompanied by swelling of the skin and the medical term to describe this is **angioedema**.

The urticaria experienced by most people is short-lived, lasting for a few days. The rash then clears up completely. This is termed **acute urticaria**. However, in some people, the urticaria will persist for a long time and become chronic.

Chronic urticaria is also quite common and about one in 100 people will have a prolonged bout of urticaria at some point in their lives.

Treatment of chronic urticaria is normally very successful, with the use of anti-histamine drugs.

The skin rash in urticaria

Urticaria can occur anywhere on the body. It can appear very suddenly. It may be widespread or occur in just one area. The appearance can vary considerably. Individual hives can be as small as a pencil tip, or hives can join together and involve a very large area.

Most often, hives are about the size of a one-euro coin and consist of a central, slightly raised white area known as a **wheal**, surrounded by a larger red area. They are characteristically intensely itchy.

Each hive lasts only for a short time – if you draw a circle around a single rash spot it will be gone within a few hours. However, in some patients, new hives can appear, resulting in the rash persisting over days, weeks or months.



Differences in the appearance of urticaria

Swelling of the skin – angioedema

Swelling of the skin, or angioedema, often develops with urticaria. When angioedema occurs on the face and is severe, it can be quite disfiguring and cause alarm. If the lips are swollen they may become very prominent. The loose skin around the eyes may swell and even impair vision.

If the tongue swells it may make speech and even swallowing difficult. If a patient develops angioedema like this anyway frequently, it can seriously disrupt daily living.

Occasionally, angioedema appears alone and there is no associated urticaria.

This booklet focuses on the condition **chronic urticaria** and **angioedema**. People with this condition can become frustrated because the rash and swelling keeps coming back and often become preoccupied with finding a cause.



Angioedema: swollen and prominent lips

2 Frequently-asked questions

What causes chronic urticaria?

It is quite understandable that someone with chronic urticaria will want to find a cause for their condition. The expectation is that some event – perhaps a food or a chemical – is responsible. In an attempt to find a cause patients will often modify their diet or avoid certain detergents or cosmetics.



However, this search for a trigger is rarely successful and mostly no cause is found. The reality is that no single triggering event is responsible. Because of this the condition is known as **chronic spontaneous urticaria**. This type of urticaria is the most common form found in adults.

Is chronic urticaria caused by an allergy?

No. Chronic urticaria is not caused by allergy.

However some patients who develop chronic urticaria have a background of allergic disease such as asthma or hay fever. This may make them more prone to chronic urticaria and, as we will see, allergy-type mechanisms are involved in both conditions. In contrast, a single episode of urticaria or angioedema may be caused by an allergic reaction to a drug or food and it is usually fairly obvious that these acted as triggers of the rash.

Do drugs cause chronic urticaria?

No. Drugs are very unlikely to be the cause of chronic urticaria.

However, it is very important to obtain information on any drugs that a patient may be taking. This is particularly the case when angioedema is developing alone, in the absence of urticaria – in this situation, certain drugs used to treat high blood pressure, called *ACE inhibitors*, may be responsible.

It is well known that certain drugs can trigger the development of acute urticaria and angioedema. Classic examples include the commonly-used anti-inflammatory drugs, such as *Brufen* or *Difene*, taken for pain relief or arthritis.

The involvement of these drugs is normally obvious, since the reaction usually takes place within an hour of taking the medication.

Can foods cause chronic urticaria?

No. Food allergy is not an important cause of chronic urticaria.

Many patients report that frequently they wake in the morning with a rash or swelling and, because there has been a long time lapse since they last ate, this is good evidence that food is not involved. Clearly, food allergy can result in **acute** urticaria and again the link with the food is usually obvious, since the reaction may develop within minutes, or at most two hours, of eating the food.

Alternative practitioners may advise patients that a wide range of foods are responsible for their chronic urticaria.

They may base their opinion on various tests that lack sound scientific evidence. Wide-ranging dietary restriction, as often advised by these practitioners, is rejected by all major medical and governmental organisations that have examined this matter.



Are there any pointers to the cause of chronic urticaria?

Single triggers are rarely responsible for chronic urticaria. Instead several factors may combine and contribute to the development of the disease.

This combination can include factors such as genetic inheritance, variation in hormone levels and stress: either from physical events or emotional stress.

What physical events or factors can cause urticaria?

It is now evident that a range of physical factors can cause urticaria. Some of these relate to changes in temperature. So, for example, patients may react to hot water when taking a shower or to very cold water after swimming in the sea.

Others may develop urticaria and angioedema during strenuous physical exercise, such as long-distance running. Tight clothing pressing against the skin can cause urticaria in that part of the body. Some people are exquisitely sensitive to sunlight and develop urticaria on exposure.



When a careful history is taken from a patient, it should become evident that specific physical factors, such as those above, are involved. In cases where physical triggers are involved, avoidance strategies can be very helpful.

What is the cause of the skin rash?

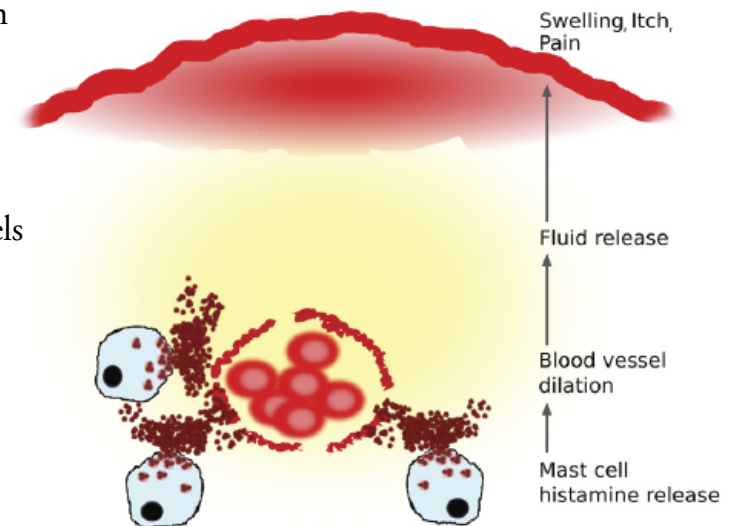
The rash in urticaria is caused by an increase in blood flow to that area of skin: this is why the rash is red in colour. The increased blood flow is caused by widening of the small blood vessels in the skin. The blood vessels can leak fluid into the skin and this causes a weal to develop, the white elevated area in the rash.

In some instances more marked leaking of fluid occurs with greater amounts of skin swelling and this is the cause of angioedema.

What causes blood vessels to behave like this?

A cell in the skin, called a **mast cell**, plays a key role in the reaction. Mast cells are abundant not only in the skin but also in tissues such as the tongue and lips. They are located close to blood vessels and contain high amounts of histamine and of various other chemicals.

When mast cells release histamine the local blood vessels widen and increase the blood flow to the area. Histamine also stimulates local nerves in the skin, causing the symptoms of itch and sometimes pain.



Why do mast cells release these chemicals?

Many different events can trigger mast cells to release their chemicals. The central role of mast cells was first identified in true allergic reactions. However, we now know that various stimuli can directly activate mast cells, causing them to release their chemicals. These stimuli include local physical stress such as pressure on the skin. Emotional stress and hormonal changes also influence the behaviour of mast cells.

What causes triggering of mast cells in chronic urticaria?

In most instances there is no single explanation for the triggering of mast cells in chronic urticaria.

As already stated, it is very unlikely that chronic urticaria is caused by a specific allergy to some unidentified substance. Your doctor will take a careful history of what has been happening and enquire in particular about any association of the rash with drugs or foods.

Information will also be sought about a possible contribution of physical or emotional stress. Emotional stress can make urticaria worse, but on its own is unlikely to cause the rash. Since mast cells are linked to nerves in the body, this may help explain how emotional and physical stress can contribute to urticaria.

Very occasionally, molecules called 'autoantibodies' are found in some patients and can trigger mast cells.



Stress may be a factor in chronic urticaria

3 More about angioedema

How frequently is skin swelling found in chronic urticaria?

Patients with urticaria often develop swelling of the skin, called angioedema. This can occur throughout the body; the face is often affected, in particular around the eyes and involving the lips. This can be disfiguring and cause distress. Swelling in the hands may cause stiffness of the joints, making them clumsy to use.

Angioedema can develop quite rapidly over the course of one hour and then resolve over 24 hours. Occasionally the swelling can persist for several days.

Is angioedema dangerous?

If marked angioedema develops it can be very alarming and upsetting for a patient. If the swelling affects the mouth or throat it may result in swallowing difficulties or a choking sensation.

However, it is important to emphasise that life-threatening breathing difficulty is extremely unlikely in the angioedema that can develop with chronic urticaria. This is in contrast to certain specific acute allergic reactions (for example, in peanut allergy) when dangerous angioedema of the throat can develop.

Your doctor will be able to determine if you are at risk.

4 Making a diagnosis

How is the diagnosis of urticaria made?

The most important step by far in making a diagnosis of chronic urticaria is to get a detailed account of the patient's history. Careful evaluation of this is very important.

If no identifiable triggers are involved, **chronic spontaneous urticaria** is the accepted medical diagnosis. The word *spontaneous* is used to acknowledge that no single external stimulus can be identified.

What tests should be performed in a patient with chronic urticaria or angioedema?

In the majority of patients with chronic urticaria, when the patient's account of their symptoms is clear-cut, there is no particular need for blood tests or other investigations. A photograph of the rash may be helpful in confirming the nature of the problem.

What tests for possible triggers might be performed?

In a small number of patients, inflammation is involved in causing the skin rash. Tests to investigate this possibility may be carried out.

Carrying out multiple allergy tests without pointers from the patient's story is not appropriate. If allergy is suspected blood or skin reaction tests can be performed; however, in the majority of patients, neither of these tests is warranted.

5 Treatment for urticaria

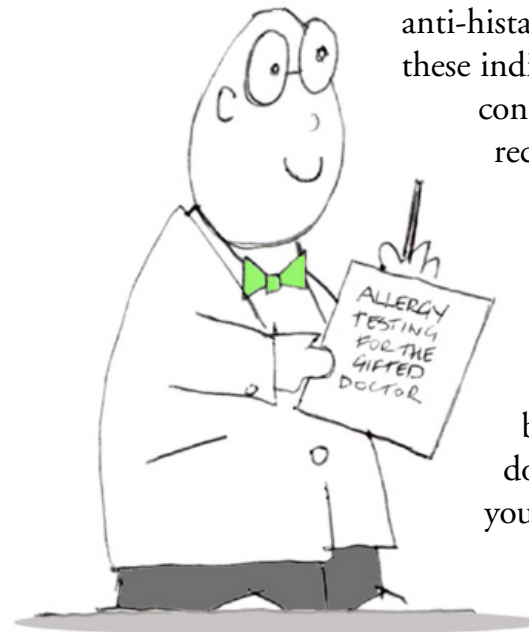
Anti-histamine tablets are the mainstay treatment for urticaria. Steroid tablets should generally be avoided. If a trigger is identified, clearly exposure to this trigger needs to be avoided. Reassurance of the patient is also very important, since some individuals become very upset by the continuing symptoms.

Which anti-histamines are recommended?

A wide range of anti-histamine drugs are available. Some of these may be purchased "over the counter" and so do not require a doctor's prescription. It is best to use the newer, long-acting, non-sedating type of anti-histamine.

If urticaria occurs only intermittently, the anti-histamine tablet may be used for these individual episodes. However, if the condition is present on a chronic, recurrent basis it is best to take the drug daily, with eventual weaning when the urticaria becomes less troublesome.

In the majority of cases this common medical condition is best managed by your family doctor who will be able to advise you on an appropriate anti-histamine tablet. A referral to a hospital specialist is required on only rare occasions.



What happens if the anti-histamine does not control the rash or swelling?

In some patients a single anti-histamine tablet is insufficient to control their episodes of rash or swelling. Your doctor may then recommend that you double, or even triple, the dose of the drug. This is regarded as a safe practice and is recommended in international guidelines for the treatment of the condition.

Are other treatments likely to help?

Since emotional stress can exacerbate chronic urticaria, a useful option is to consider stress management. A variety of relaxation techniques may prove to be helpful. These techniques include yoga and cognitive behavioural therapy. The importance of stress as a factor contributing to the severity of this condition should not be underestimated.



Can steroids be used to treat chronic urticaria?

Steroid drugs, such as *Prednisolone*, are best avoided in the treatment of urticaria or angioedema. If a patient's experience is that only steroids are able to control their condition then further evaluation by a hospital specialist is advisable.

In a small number of patients the urticaria may be due to an abnormal immune response, causing inflammation. Further blood tests may reveal this to be the case. In this situation, potent drugs that suppress the immune system can be used and their role is being investigated.

Should adrenaline be used to treat chronic urticaria and angioedema?

Adrenaline is not required in the standard treatment of chronic urticaria and angioedema. This is in contrast to the need for adrenaline in acute allergic reactions which can cause severe breathing difficulties or a marked fall in blood pressure (given the medical term **anaphylaxis**). In anaphylaxis an injection of adrenaline is a life-saving treatment.

However, in chronic spontaneous urticaria with or without angioedema these life-threatening events do not develop. Accordingly there should be no need for adrenaline treatment and certainly no need for the patient to keep self-injectable adrenaline.

The unnecessary prescription of adrenaline is likely to cause a patient additional anxiety about their condition.

Have newer therapies been developed?

Yes, new drugs are under investigation.

One such drug, called *Omalizumab*, looks promising and is now recommended in the treatment of patients with very resistant urticaria or angioedema. This treatment is given by an injection every month. Other important therapies are likely to be discovered in the future.

What is the outlook for patients with chronic urticaria?

In about 50% of patients the condition resolves within one year.

About 10% of patients will still have some features of the condition after ten years.

However, it is important to stress that the majority of people with chronic urticaria, even if affected for a prolonged period of time, can have their symptoms satisfactorily controlled with anti-histamines. These medications are effective and, if needed, are safe to take for lengthy periods.

Summary

Chronic spontaneous urticaria is a common skin rash disorder that can cause significant patient discomfort and even alarm, especially if marked skin swelling also develops.

The cause of the condition is often unclear and a combination of factors may be involved.

It is usually satisfactorily controlled by taking anti-histamine tablets and normally clears up completely over a period of time.

In a small number of patients, more potent medication is needed to prevent the condition continuing.

Information available on the Internet

The following websites and locations offer patient information on urticaria and angioedema:

<http://www.bad.org.uk/shared/get-file.ashx?id=184&itemtype=document>

Entering the URL above into your web browser will link you to a direct download of a short document for patients, produced by the British Association of Dermatologists, explaining the nature of chronic urticaria.

<http://www.patient.co.uk/doctor/urticaria-pro>

This is a British patient website which describes the nature and the features of chronic urticaria.

Glossary

- **Adrenaline:** a drug given by injection which can reverse the process of anaphylaxis
- **Allergy:** an acute medical reaction, frequently to a common substance such as grass pollen
- **Anaphylaxis:** an acute allergic reaction causing severe breathing difficulties or serious reduction in blood pressure
- **Angioedema:** swelling of the skin, lips, tongue or other parts of the body, caused by a collection of fluid
- **Anti-histamine:** a drug which inhibits the action of histamine
- **Anti-inflammatory drugs:** commonly prescribed drugs used to relieve pain and inflammation
- **Mast cell:** a type of cell found in the skin or tissues such as the lips; these cells contain large amounts of chemicals such as histamine, responsible for urticaria
- **Steroids:** powerful drugs that suppress the immune system
- **Urticaria:** the medical term for hives in which a red, itchy skin rash develops rapidly and fades within hours
- **Wheal:** a small collection of fluid that is at the centre of a patch of urticaria.



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Original illustrations: Michael Conlon
Design, layout and pre-press: Ferret on the Dartboard
Printed in Dublin