

# Your quick guide to: Chronic Spontaneous Urticaria(CSU)

Chronic Spontaneous Urticaria (CSU) is a common and distressing skin condition that causes red, raised, itchy and sometimes painful wheals (often referred to as hives, bumps or nettle rash) on the skin with no known obvious trigger. To be considered chronic the wheals (urticaria) must be present daily for at least six weeks and is referred to as 'spontaneous' when the trigger is not known.

## Who is affected by Chronic Spontaneous Urticaria?

CSU is thought to affect 0.5-1% of the population in the UK, with women more than twice as likely than men to be diagnosed with it. Most people develop symptoms between the ages of 20 to 40 and although CSU can affect children, it is more common in older children and adolescents than infants.

## Triggers of Chronic Spontaneous Urticaria

Urticaria, the rash that occurs on the skin, is caused by an immune response when a mast cell is activated in the immune system. Mast cells are cells which circulate in the blood and are found in most tissues in the body, including the skin and lungs.

Mast cells help the immune system to reduce inflammation, fight infection and are involved in wound healing and repair. In urticaria, mast cells are activated due to a signal (trigger) and release chemicals, including histamine, into the skin tissue and this causes the red itchy raised rash often referred to as hives.

Research has shown that around 50% of cases are linked to autoimmune disease, especially those who do not respond to antihistamine therapy. It is thought that in these cases the immune system mistakes its own cells as harmful and activates cells, including mast cells, to begin attacking itself and causing the symptoms of urticaria.

## Could allergy be a trigger for CSU?

CSU is not caused by an allergic reaction to a trigger allergen, although it is an immune

response that causes the symptoms. Allergy testing, or elimination diets are not usually helpful in the management of CSU. Some people can identify things that can make their CSU worse, such as stress, infection and medication. Common medications which can trigger urticaria include NSAIDs (non steroidal anti inflammatory drugs) and if angioedema (deep swelling) is present, it is advisable to avoid ACE (Angiotensin-converting enzyme) inhibitors where possible.

Often there is no obvious external trigger or aggravating factor that causes these symptoms. This is because CSU differs from other urticarial reactions in that there is often no known cause. CSU is linked with autoimmune disorders in 50% of individuals with the condition, so tests for thyroid function and autoimmune disorders may be performed.

## Symptoms of Chronic Spontaneous Urticaria

- Raised rash or patches surrounded by red, raised, inflamed skin often called hives or nettle rash and can affect any skin type or tone. There may be tiny bumps or large raised patches of variable sizes which can be white or red in colour with a red flare.
- On darker skin tones the raised patches often match the surrounding skin tone with no red flaring of the skin visible.
- Can affect any part of the body and is usually itchy but can also be painful or have a burning sensation.
- Wheals often change shape before resolving, typically within 24 hours, but as one wheal resolves others can develop and the rash can present for long periods.
- The rash does not cause any lasting damage to the skin, but in darker skin tones post inflammatory hyperpigmentation (darkening of the skin), can occur to the affected area and may take months to settle.
- May also be accompanied by deep

## Key facts:

Chronic Spontaneous Urticaria is an **unpredictable** and **debilitating** condition which can affect daily life in many ways including sleep deprivation, **anxiety** and **social isolation**.

CSU is a **common** and distressing **skin condition**. To be considered chronic the wheals (urticaria) must be present daily for at least **six weeks** and referred to as 'spontaneous' when symptoms appear with **no obvious trigger**.

If you feel your symptoms are not being managed adequately, it is important that you seek advice from a healthcare professional.

## Allergy UK Helpline

Mon-Fri, 9am-5pm:

Call: 01322 619 898

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swelling (angioedema) of the face, neck, hands or feet.

Some people also state they experience generalised symptoms with CSU, including headache, fatigue, joint pain, swelling, stomach problems, flushing, palpitations and occasionally wheeze.

## Diagnosing Chronic Spontaneous Urticaria

CSU can sometimes be difficult to diagnose, as urticaria can be a symptom of other medical conditions. It may take several visits to your GP or healthcare professional to get a diagnosis or to find a treatment that helps to control your symptoms. Your healthcare professional may carry out diagnostic testing to rule out other medical conditions. They may also ask you to keep a symptoms diary or complete a CSU severity scoring system to help assess the severity of your CSU.

It is important to seek help if...

- Your symptoms are not well controlled, and/or are affecting your day-to-day activities, and/or affecting your mental health
- Antihistamines are needed continuously (daily) to control symptoms for more than six weeks
- Symptoms are painful and persistent

CSU symptoms are rarely life threatening, but you should seek urgent medical attention if you are concerned that your tongue or throat is swelling or that your swallow or airways and breathing are affected.

It is important that if you feel your symptoms are not being managed adequately that you consult your healthcare professional who, if necessary, can refer you to specialist treatment.

## Treating Chronic Spontaneous Urticaria

Unfortunately there is no cure, but there are various treatment options available for CSU, with therapies designed to

relieve symptoms. There are guidelines and treatment pathways to support the recommend treatments of CSU, and you can discuss with your healthcare professional the most effective regime to find a treatment that will enable patients to enjoy a good quality of life and manage symptoms.

## Standard treatment pathways in managing CSU

### Step 1: Antihistamines

Antihistamines are the first-line medication offered for CSU. This simple treatment is effective in reducing symptoms for up to 40% of people with CSU. Current guidelines recommend using non-sedating antihistamines in the first instance, but occasionally sedating antihistamines may be prescribed, particularly at night to help with night-time itch and sleep disturbance. However, it is worth noting that sedating antihistamines can cause additional issues, such as problems dealing with fatigue and restriction on daily activities. It is not advisable to drive or operate machinery when taking sedating antihistamines, due to the sedating action and their effect on the central nervous system.

If a once-a-day dose of the antihistamine doesn't reduce symptoms, then medical guidelines recommend that doctors can increase antihistamines up to four times the licensed dose depending on personal circumstances such as other medical conditions, age and general health. However, despite this recommendation, a proportion of patients (40%) will still experience breakthrough symptoms of CSU.

It is advised that once symptoms are completely controlled patients should remain on the medication for at least 3-6 months. After this time, under the management of a healthcare professional, patients can step down and gradually reduce the non-sedating antihistamines. If at any stage symptoms recur, patients should go back to the previous step that provided complete control and re-attempt stepping down.

### Short course corticosteroids:

Corticosteroids can be used as a short course to help cope with an acute flare up, but should not be used on a long-term basis due to the risk of serious side effects.

### Step 2: Biologic therapies

Where standard treatments are not effective in symptom relief or reducing the burden of CSU, biologic therapies can be introduced alongside other medications such as antihistamine therapy.

Biologic therapies are designed to target a specific part of the immune system, and modify or suppress the immune response which can provide relief from the condition. These treatments are only available under supervision from specialist hospital departments.

There is currently only one biologic medication approved and licensed to treat CSU, which is referred to as anti-IgE monoclonal antibody (Mab) It has been shown to have great success in reducing symptoms for individuals with CSU. This treatment works by blocking mast cells, in the blood from producing chemical mediators, such as IgE and histamine, which are thought to cause the inflammatory process that produces the symptoms of urticaria. As this treatment targets a specific part of the immune system, this reduces the risk of serious side effects.

There are other products targeting specific areas of the immune system that have been developed with some going through clinical trials that look very promising and could certainly provide other treatment options and improve symptom control for CSU in the future. These include Bruton Tyrosine Kinase (BTK) inhibitor and Tumor Necrosis Factor Alpha (TNF $\alpha$ ) inhibitor. All will need to go through the appropriate approvals process before they will become available on the NHS.

### Step 3: Immunosuppressive treatments

Third-line therapy involves immunosuppressive treatments, designed

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to suppress the immune system and reduce the inflammatory process and include medications such as ciclosporin and methotrexate. While immunosuppressive treatments are not specifically licensed or designed to treat CSU, they are useful in controlling symptoms in individuals where there is a possible autoimmune aspect to their condition, or where other treatments such as antihistamine therapy have not been effective.

These medications work by having a suppressive effect on the whole immune system, and do not only target the parts of the immune system that are causing the inflammatory process. This can lead to some serious side effects. To prevent the risk of unwanted side effects, individuals on these treatments usually require regular monitoring and are therefore usually started in a hospital setting.

## Living with Chronic Spontaneous Urticaria

CSU is an unpredictable and debilitating condition. It can affect the daily life of suffers in many ways including the ability to carry out simple daily tasks such as walking, sitting, dressing and bathing. It can also have a detrimental impact on sleep, social and intimate relationships and sexual function. Having symptoms of CSU is known to be a risk factor for mental health issues, including anxiety and depression.

## How long does CSU last?

CSU can continue for long periods of time; between one to two years in 80%

of individuals and then often resolves spontaneously. However in around 20% of individuals, CSU has been known to continue for longer than 10 years.

## Pregnancy and breast feeding

Sedating antihistamines (such as Chlorphenamine) should be avoided during pregnancy and breastfeeding. Studies have shown that antihistamines can cross into breast milk in low quantities.

Although no medication can be advised to be risk free during pregnancy or breastfeeding, non-sedating antihistamines, such as cetirizine or loratadine, have been shown to have the best safety profile and are the preferred medication of choice if symptoms remain troublesome.

It is advised that if medication is required during pregnancy and breastfeeding, then it is to be taken at the lowest possible dose that offers symptom control.

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*This factsheet was prepared by Allergy UK's Clinical Team*

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