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 hives or wheals 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.

What does urticaria look like
• Raised rash or patches surrounded by red, raised, inflamed skin often called hives or nettle rash and can affect any skin type or tone.
• They 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 changing shape before resolving 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 to the area that has been affected, can occur and may take months to settle.
• May also be accompanied by deep swelling (angioedema) of the face, neck, hands or feet.

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

Who is affected by CSU?
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.

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. But in around 20% of individuals, CSU has been known to continue for longer than 10 years.

What causes CSU?
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 skins tissue and it is this that causes the red itchy raised rash often referred to as hives.

Research into the causes of CSU has linked autoimmune disease with about 50% of individuals with CSU, especially in individuals not responding to antihistamine therapy. It is thought that when CSU is linked to an autoimmune disorder, 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.

Key facts:
CSU is an unpredictable and debilitating condition which can affect daily life in many ways including sleep deprivation, anxiety and social isolation.

To be considered ‘chronic’ the wheals must be present daily for at least six weeks and ‘spontaneous’ when symptoms appear with no obvious trigger.

It is important to seek advice from a healthcare professional if your symptoms are not being managed adequately.

Allergy UK Helpline
Mon-Fri, 9am-5pm:
Call: 01322 619 898
Email: info@allergyuk.org

Visit us at:
Allergyuk.org
Could allergy be a trigger for CSU?

CSU is not caused by an allergic reaction to a trigger allergen; it is an immune response which causes the symptoms. Therefore 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 medication, stress and infection, but 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.

How can I control my symptoms - what are my treatment options?

There are various treatment options available for CSU, with treatment designed to relieve the symptoms but unfortunately there is no cure. There are guidelines and treatment pathways to support the recommend treatments of CSU, and you can discuss with your healthcare professional the most effective treatment pathway to find a treatment that will enable you to enjoy a good quality of life and be symptom free.

Standard treatment pathways offer a four step approach to 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 to use 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 effect on the central nervous system and the sedating action of the treatment. If a once a day dose of the antihistamine doesn’t reduce your symptoms then medical guidelines recommend that doctors can increase antihistamines up to four times the licensed dose to achieve symptoms relief. This will be dependent on whether you have other medical conditions, your age and general health. However, despite this recommendation, a proportion of patients (40%) will still experience breakthrough symptoms of CSU.

Step 2: Medications

When antihistamines are not effective, your healthcare professional may consider a second-line therapy with medications such as Leukotriene receptor agonists. These are often used in the treatment of asthma, this type of medication is not a steroid treatment, but works to reduce inflammation by blocking the effects of leukotrienes (a chemical released by mast cells during a reaction).

Short course corticosteroids: Corticosteroids should only be used as a short course to help cope with a flare up and not be used on a long term basis due to the risk of serious side effects.

Step 3: Immunosuppressive treatments

Third-line therapy involves immunosuppressive treatments, designed to suppress the immune system and reduce the inflammatory process. This includes medications such as ciclosporin and methotrexate, immunosuppressive treatments are not specifically licensed or designed to treat CSU. However 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 has not been effective. The action of these medications means that they are not selective i.e. are not designed to only target the parts of the immune system that are causing the inflammatory process, but have a suppressive effect on the whole immune system. 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 therefore these treatments are usually started in a hospital setting.

Step 4: Biologic therapies

Newer treatments, called biologic therapies, are designed to target a specific part of the immune system and modify or suppress the immune response to gain control of symptoms and provide relief from the condition. These treatments are only available from specialist hospital departments.

There is currently only one biologic medication approved and licensed to treat CSU. This is referred to as anti IgE monoclonal antibody (Mab). This treatment works by blocking specific cells, called mast cells, in the blood from mounting an immune response. This prevents the mast cells producing chemical mediators, such as IgE and histamine. It is thought that these chemical mediators released from the mast cells, cause the inflammatory process that produces the symptoms of urticaria. It is because this treatment targets a specific part of the immune system that this reduces the risk of serious side effects.

Anti IgE monoclonal antbodies (MAbs) have been shown to have be very successful in reducing in symptoms for individuals with CSU. 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.
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Getting a diagnosis and appropriate therapy
CSU can be difficult to diagnose and 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 swallowing 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 on for specialist treatment.

Key messages
• CSU 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.
• It is important that if you feel your symptoms are not being managed adequately, that you seek advice from a healthcare professional.

References
• Allergy UK. Wheals of Despair. Chronic spontaneous urticaria: breaking free from the cycle of despair 2014.
• Powell, R, Leech, S , Till, S , Huber,P , Nasser, S, Clark, A. BSACI guideline for the management of chronic urticaria and angioedema Clinical & Experimental Allergy, 2015: 45, 547–565

Further information
For more information on Chronic Spontaneous Urticaria (CSU), contact:
• Allergy UK www.allergyuk.org
• Skin Support www.skinsupport.org.uk

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