

GP Guide to Chronic Spontaneous Urticaria and Angioedema

This guide is intended to provide advice on the recognition and non-specialist management of Chronic Spontaneous Urticaria and Angioedema (CSU&A). Severe cases not responding to first line therapy or cases where the diagnosis is in doubt may benefit from a referral to the Peninsula Specialist Immunology and Allergy Service.

CSU&A

Diagnosis: CSU&A, also known as chronic idiopathic urticaria and angioedema, is a relatively common and often under-recognised condition of frequent, recurrent urticarial rashes with or without angioedema. Urticaria and angioedema can occur simultaneously or separately. A minority of patients can also suffer anaphylaxis; if there has been suggestion of airway swelling in the past the patient should be prescribed 2 self-injectable adrenaline pens and educated in their use, under these circumstances referral to the Peninsula Specialist Immunology and Allergy service at Derriford would be appropriate. Symptoms are caused by spontaneous histamine release from mast cells and as such will be at least partially responsive to antihistamines. It is important to differentiate this condition from Type 1 hypersensitivity reactions with the release of histamine and other mast cell mediators triggered by specific, reproducible and reliable triggers, eg food, drugs, stings. Such differentiation is usually clear from the history; the following are features suggestive of CSU&A;

- Frequent symptoms (multiple times per week)
- Protracted symptoms (lasting >24hours)
- No discernible pattern to symptom onset, eg, not always within 1 hour of food, patients wake up in the morning with symptoms
- No specific triggers linked to every episode of symptoms
- Exacerbated during intercurrent illness, fever, heat, with NSAIDS, alcohol, stress, oestrogens

Investigation: CSU&A can be associated with undiagnosed or poorly controlled systemic illness, the likelihood of which is related to the standard risk factors for each associated condition, eg, autoimmune thyroid disease, diabetes, H.Pylori, malignancy. Screening tests can be organised in primary care accordingly. If the patient has had symptoms suggestive of anaphylaxis measure a baseline Mast Cell Tryptase (MCT).

Other medication; ACE inhibitors should be avoided in patients with a history of angioedema, NSAIDS are not absolutely contraindicated but can cause exacerbation of CSU&A in some patients and should be used with caution

Treatment: The mainstay of treatment is regular long acting non-sedating antihistamines used at high dose. These medications are licenced for once daily use however there is good

safety data for more frequent dosing and their use up to QDS is recommended in this condition by the BSACI.

The following treatment can be initiated in primary care, each step should be tried for at least 1 month;

1. Single agent long acting non-sedating antihistamine eg Cetirizine 10mg up to QDS to achieve complete symptoms control
2. If ineffective, change agent, eg to fexofenadine 180mg up to QDS
3. An emergency pack of prednisolone 15-20mg OD for up to 3 days can be considered for slowly resolving episodes. If the patient requires this emergency pack more than once a month then the background medication should be escalated and referral considered.
4. Add ranitidine 150mg BD or Montelukast 10mg ON

Referral: Refer to the Peninsula Specialist Immunology and Allergy Service if;

- Step 4 of the treatment plan is attempted and condition not under control.
- If the patient has had symptoms suggestive of anaphylaxis in the past.
- There is a raised baseline MCT

There are a number of specialist treatments available on a case by case basis including immune suppression and immunomodulation. Such treatments are not without risk and are not suitable for all patients.

If there is any coexistent eczema, management should be optimised. If there is isolated eyelid erythema and swelling with or without rash, especially if that rash is not typically urticarial, a referral to dermatology may be more appropriate.

Angioedema without Urticaria

Angioedema without urticaria can be histamine mediated in which case it should respond at least partially to antihistamines and/or steroids. If this is the case the same advice as for CSU&A applies.

Angioedema without urticaria which does not respond to antihistamines is suggestive of bradykinin-mediation. The following steps should be taken;

- Stop ACE-I (if patient on one) – if symptoms persist 3-4 months after the ACE-I has been stopped refer to the Peninsula Specialist Immunology and Allergy Service
- Blood tests for Complement C4 level. If low, further testing for Complement C1Inhibitor level and function may be required, however specific sampling requirements mean that these tests are best undertaken in secondary care.
- Assess clinically for autoimmune disease and haematological malignancy
- Identify any first degree family history of angioedema without urticaria.

Referral: Once ACE-I related angioedema has been excluded, most of the above patients will benefit from a specialist referral to the Peninsula Specialist Immunology and Allergy Service.