Management of urticaria

Malcolm Greaves

Urticaria is a very common and disabling disorder. Recent advances in understanding of pathomechanisms has led to important advances in diagnosis and treatment. However, the cause in many patients remains obscure and the treatment poorly effective.

Urticaria can be classified as acute, chronic or intermittent. Chronic urticaria conventionally includes the physical urticarias and urticarial vasculitis. Intermittent urticaria can be arbitrarily defined as the occurrence of repeated bouts of acute urticaria lasting up to 6 weeks, the patient being completely or almost completely symptom-free in between. The types of urticaria to be considered in this account are included in *Table 1*. This account principally deals with management of urticaria in adults. The reader is referred to a recent review by the author (Greaves, 2000) for details of specific problems of management of urticaria in childhood.

ACUTE URTICARIA

Arbitrarily defined as daily or almost daily urticaria lasting up to 6 weeks, acute urticaria may be associated with angio-oedema, oropharyngeal swellings and systemic symptoms, which range from mild (headache, nausea, flushing and sweating) to, less commonly, severe (bronchospasm, vomiting, diarrhoea, collapse).

Aetiology

Acute allergic urticaria (e.g. as a result of peanut or tree-nut allergy) is a type I (Gell and Coombs classification) immunoglobulin E (IgE)-mediated reaction. However, acute urticaria can also be non-allergic (e.g. as a result of histamine as in scombrotoxic fish poisoning, or of aspirin in an aspirin-sensitive individual).

Upper respiratory infections are also an important cause (Zuberbier et al, 1996). However, no cause can be identified in about half of patients with acute urticaria.

Management

Mildly affected patients can usually be managed by oral histamine-1 receptor (H_1) antagonists alone.

Acute urticaria with angio-oedema requires emergency measures. Administration of parenteral adrenaline 0.5–1.0 mg can be repeated every 15 minutes, depending on pulse rate and blood pressure, which should be regularly monitored. Concurrently an H_1 antihistamine such as chlorpheniramine 4 mg or diphenhydramine 25 mg can be given intramuscularly. A short tapering course of oral steroids (e.g. prednisolone 30 mg daily, reducing by 5 mg every other day) can be commenced as soon as the patient is stabilized. This should be given concurrently with a low sedation H_1 antihistamine in the morning and a sedative H_1 antihistamine at night.

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TABLE 1.
Classification of urticaria

Acute		
Chronic	Physical	Symptomatic dermographism
		Delayed pressure
		Cholinergic
		Cold
		Solar
	'Idiopathic'	
	Autoimmune	
Intermittent*		
Urticarial vasculitis		
Papular (insect bite reactions)†		
Contact†		
Drug-induced†		

* causation and management as for autoimmune/'idiopathic'; † listed for completeness, will not be discussed in detail

In patients in whom an allergen can be identified (e.g. penicillin, peanuts) the patient should be educated regarding avoidance, wear a disc identifying the wearer as a sufferer from specified allergies and, in the case of food allergies, receive dietary advice. Most patients can be taught to self-administer adrenaline and commercial kits are available for this purpose. At the present time desensitization to food and other oral allergens is not a practical proposition. Menthol 1% in aqueous cream is a useful mild topical antipruritic. The drug treatment of severe acute urticaria is summarized in *Table 2*.

PHYSICAL URTICARIAS

The physical urticarias listed in *Table 1* will be considered here. Each of these presents as an urticarial response to a physical stimulus. For symptomatic dermographism, delayed pressure urticaria, cold and solar urticaria, the itching and wealing occur directly at the site of application of the appropriate physical stimulus. For cholinergic urticaria the physical stimulus is a rise in body temperature or emotional stress.

Symptomatic dermographism

Wealing and itching occur locally and immediately in response to firm stroking of the skin. There is no angio-oedema and there are no systemic symptoms. The weals fade within a few minutes without residual marking of the skin. The skin looks clinically normal before and after occurrence of wealing, and histologically there is no increase in mast cell numbers. The cause is unknown, but dermal perfusion studies indicate that histamine is the principal, if not the only, mediator (Greaves and Sondegaard, 1970). Once the diagnosis has been confirmed by challenge testing (elicitation of an immediate weal and itch following firm stroking of the skin) no further investigations are indicated.

As with nearly all physical urticarias, there is no evidence for food intolerance in symptomatic dermographism. The natural history is for spontaneous remission in 1–2 years in most cases, although rarely it can be persistent.

Management: As with other physical urticarias, attention needs to be paid to lifestyle and environment. A cool ambient temperature reduces itching and helps break the itch–scratch–weal–itch cycle. Wool and coarse garments should be avoided. Loose-fitting light clothes are ideal. Regular sleep and eating patterns also help, and patients with high levels of personal and occupational stress suffer more severe symptoms.

Patients are routinely managed with H_1 antihistamines. A good combination is to use a low sedation antihistamine such as fexofenadine 180 mg or loratadine 10 mg each morning. Nocturnal pruritus is best dealt with by a tepid shower, coupled with a sedative antihistamine such as hydroxyzine 25 mg. Controlled studies of combined H_1 and H_2 antihistamines have shown significant statistical evidence of better suppression, but it is doubtful if the difference is clinically useful (Breathnach et al, 1983).

TABLE 2.
Drug treatment of severe acute urticaria

Emergency treatment*	Adrenaline (epinephrine) intramuscularly 0.5–1.0 mg, repeated every 15 minutes until patient is stabilized
Maintenance	Prednisolone 30 mg daily (tapering regimen)*
	Loratadine 10 mg, fexofenadine 180 mg or cetirizine 10 mg daily plus hydroxyzine 25 mg at night

* severely affected patients only

Delayed pressure urticaria

This physical urticaria exhibits major clinical differences to other physical urticarias. Wealing develops at the site of pressure perpendicularly applied to the skin after a latent period of several hours. The painful and/or itchy swelling lasts 24 hours or more before clearing. Areas characteristically affected include the palms and soles, waistband and shoulders, causing considerable disability, especially in manual workers. In the author's experience, delayed pressure urticaria never occurs alone; it is invariably accompanied by chronic 'idiopathic' urticaria, although its severity may overshadow symptoms caused by the latter. Histologically there is no vasculitis and the appearances of a skin biopsy are those of a late-phase reaction.

Angio-oedema does not occur but systemic symptoms in the form of arthralgia and fatigue are common. The aetiology is unknown, but analysis of interstitial dermal fluid from involved skin indicates the involvement of leukotrienes and interleukin-6. The natural history is one of persistence, and histories of several years' duration are common. Once the diagnosis is made by challenge testing (application of vertical pressure to the skin with a weighted rod) to elicit the characteristic delayed-onset persistent swelling, no further investigations are indicated.

Management: General measures including wearing of loose-fitting garments and soft footwear are valuable. Although not yet subjected to controlled trial, the author has found that topical application of a non-steroid anti-inflammatory cream (benzydamine 3%) causes useful relief of symptoms.

Unfortunately delayed pressure urticaria is poorly responsive to oral H_1 antihistamines and

early suggestions of superiority of cetirizine have not been confirmed. Oral corticosteroids in high dosage may be effective but their toxicity precludes long-term usage. Because of its persistence and poor response to antihistamines and other measures, delayed pressure urticaria is an unsatisfactory condition to manage (Lawlor et al, 1989).

Cholinergic urticaria

This physical urticaria generally affects older children and young or middle-aged adults and is common in patients with an atopic background. It is very common, affecting up to 20% of the population at least to a mild degree. Characteristically itchy monomorphic symmetrical maculo-papules or small weals occur, especially in flexural areas of the limbs and the neck, in response to heat, emotion or exercise. If the patient 'chills down' the rash usually subsides rapidly. Angio-oedema may occur and systemic symptoms are common in more severely affected patients. These consist of headache, flushing, wheezing and occasionally syncope.

Diagnosis is by challenge testing (hot bath or shower, or exercise) to elicit the typical rash. The condition is mediated by activation of sympathetic cholinergic nerve fibres supplying the eccrine sweat glands, but sweating itself is not necessary — the condition occurs in individuals with congenital absence of sweat glands. It seems that activation of sympathetic cholinergic pathways releases acetylcholine in the skin which, in turn, degranulates dermal mast cells leading to local histamine release (Herxheimer, 1956). Atropinization of the skin by topical application of 6% hyoscine solution will block the rash, but anticholinergic therapy is not a practical treatment. No investigations are necessary once the diagnosis is confirmed — the condition usually improves spontaneously in 2–3 years but can be disabling in severely affected patients. A rare form of this physical urticaria involves initial ingestion of food followed promptly by exercise which in turn leads to development of the rash.

Management: Severity and frequency of attacks can be minimized by a regular and well-ordered lifestyle, in which physical and mental stress, and a hot environment are avoided as far as is possible. For those in whom emotional stress and anxiety are major factors an oral anxiolytic β-blocker such as propanolol can be administered in low dosage. This drug should not be given to patients in whom bronchospasm accompanies attacks of the rash. However, the mainstay of treatment for the majority of patients is once daily administration of a low-sedation antihistamine such as loratadine 10 mg or fexofenadine 180 mg.

For patients with severely disabling cholinergic urticaria, unresponsive to H_1 antihistamines, treatment with low-dose anabolic steroids can be considered. This treatment, which is based upon an early finding of significantly lowered levels of plasma protease inhibitors in cholinergic urticaria (Eftekhari et al, 1980), was subsequently shown to cause statistically significant amelioration of symptoms in a double-blind controlled trial (Wong et al, 1987). Stanazolol or danazol should be used cautiously in low dosage and are unlicensed for this indication. Patients receiving long-term treatment with these drugs should be regularly monitored by liver function tests and liver ultrasound scans. Because of the weak androgenic action of anabolic steroids they are better tolerated by males than females who may notice reversible voice changes and menorrhagia. Unfortunately, tolerance treatment (see below) is ineffective in cholinergic urticaria.

Cold urticaria

This physical urticaria is equally common in children and adults, and in highly cold-sensitive patients can be very disabling. Exposure to cold air or water or contact with cold surfaces causes immediate wealing and itching which subsides within about 30 minutes on rewarming. Angio-oedema (especially of the lips and tongue after a cold drink) and systemic symptoms (after extensive exposure, and ranging from mild headache to anaphylactoid symptoms in more severe cases) are common. The diagnosis is confirmed by an immediate weal and flare response to placing an ice cube on the skin for 15 minutes. Five minutes should be allowed for rewarming after removal.

Although patients with cold urticaria may occasionally be demonstrated to have circulating cryoglobulins or cold agglutinins, usually none can be found. However, a circulating passively transferable factor can be demonstrated in most patients (Misch et al, 1983). An autoimmune mechanism has been proposed by Gruber et al (1988). Histamine is probably the main mediator of cold urticaria but analysis of venous effluent from cold-challenged forearm skin raises the possibility of involvement of other mediators including leucocyte chemotactic factors and eicosanoids. The prognosis is good, spontaneous improvement occurring in 2–3 years in most patients. However, the disease can be intractable and severe in a small minority. Although circulating cold agglutinins and cryoglobulins should be sought, no other laboratory investigations are indicated. If cryoglobulins are found, a search for evidence of hepatitis B or C infection and paraproteinaemia should be initiated.

Lymphoreticular malignancy and glandular fever should also be excluded.

Management: Fatalities have occurred in highly cold-sensitive individuals during bathing in cold pool or sea water. Patients or their parents should be advised that even in heated pools, bathing should only be carried out under close and responsible supervision. Low sedation antihistamines should be effective in most cases. As with all forms of urticaria, regular daily antihistamine dosage achieves better results than medication on an occasional 'as required' basis.

There is no convincing evidence that any one antihistamine is more effective than others, although for most purposes one of the low sedation antihistamines should be preferred. The author has had a number of mainly adult patients with extremely intractable antihistamine-unresponsive cold urticaria, necessitating change of occupation in some instances. In these cases, cold tolerance treatment (sometimes wrongly termed 'cold desensitization) can be offered (Bentley-Phillips et al, 1976). This treatment involves repeated cold challenge of progressively extending areas of skin, until the required regions of the body are rendered unresponsive to cold. It requires a highly motivated patient; the more so as daily maintenance cold challenges are necessary. It should, in any case, only be initiated on a hospital inpatient basis, under close supervision. Patients with severe antihistamine-resistant cold urticaria number among the author's most spectacular therapeutic failures in his urticaria clinic.

Solar urticaria

Solar urticaria is a rare physical urticaria which is nevertheless important to identify correctly. Patients complain of immediate itching and wealing in response to sunlight exposure, which rapidly subsides if the patient seeks the shade. Angio-oedema and systemic symptoms are usually absent. The diagnosis is confirmed by challenge testing using natural sunlight or artificial light, e.g. from a slide projection lamp, which should evoke an immediate urticarial reaction. Most patients referred to the author with this suggested diagnosis turn out to have polymorphic light eruption — a common photodermatosis in which a pruritic eruption occurs hours after sun exposure and persists for several days. In children it is important to exclude the hereditary inborn error of porphyria metabolism, erythropoietic protoporphyria, which presents with symptoms superficially resembling solar urticaria.

The pathogenesis is unclear. Patients may respond to solar emission wavelengths ranging from medium wavelength ultraviolet B (290–310 nm) up to and including visible light. A passively transferable light-activated serum factor has been identified. Histamine appears to be the main, if not the only, mediator. Investigations should, in children, include ultraviolet microscopy of the patient's red blood cells to eliminate erythropoietic protoporphyria. It is also useful to determine the action spectrum for elicitation of wealing in each individual case, so as to enable focused prescription of a suitable sunscreen.

Management: Antihistamines are usually poorly effective in this distressing physical urticaria. Sunscreens, tailored to the patient's action spectrum (see above), can be useful. Tolerance treatment, based upon the same underlying principle as for cold urticaria (see above) (Ramsay, 1977), may also be tried. PUVA (8-methoxypsoralen- ultraviolet A) photochemotherapy has been successful with or without accompanying plasmapheresis (Hudson-Peacock et al, 1993) in some patients.

The main strategies in therapy of the physical urticarias are summarized in *Table 3*.

TABLE 3.
Treatment of physical urticarias

Physical urticaria	Antihistamines	Other measures
Symptomatic dermographism	Effective	None
Delayed pressure	Poorly effective	None
Cholinergic	Effective in most patients	Anabolic steroids in antihistamine-resistant severely disabled patients
Cold	Effective in most patients	Cold tolerance treatment in highly motivated patients
Solar	Not very effective	Tolerance treatment; PUVA with or without plasmapheresis

PUVA = 8-methoxypsoralen-ultraviolet A

CHRONIC IDIOPATHIC URTICARIA

Chronic idiopathic urticaria (CIU) is a common aetiologically enigmatic and intractable disease. Often regarded by dermatologists and allergists alike as a 'heart-sink' condition, it defies explanation in most cases, and is at best only partially responsive to treatment. However, recently progress has been made.

Fifty per cent of patients with CIU also have angio-oedema and almost 50% also have delayed pressure urticaria. At the outset it is important to decide whether or not any associated delayed pressure urticaria is the dominant cause of disability; if it is, there is no point in further investigating the patient because the aetiological factors which might be involved in CIU have no bearing on delayed pressure urticaria which, as noted above, is of unknown aetiology.

CIU in which individual weals have a time course of less than 24 hours must also, at the out-

set, be distinguished from urticarial vasculitis (see below). In the latter, individual lesions last longer than 24 hours, may stain the skin as a result of extravasation of red blood cells, and are poorly responsive to antihistamine treatment. Systemic symptoms are rare in CIU but common in urticarial vasculitis. Ultimately, a skin biopsy is the only sure way of distinguishing the two. Weals of CIU do not show evidence of vascular damage.

Most patients with CIU believe, or have been told by friends or relatives, that the cause of their urticaria is a food 'allergy'. This is only rarely the case. CIU pursues an intermittent course in most patients, and temporary remissions are often wrongly attributed to dietary factors. True food 'allergy' (i.e. IgE-mediated type I allergy) is an important cause of acute, but not chronic, urticaria. Non-allergic idiosyncratic reaction to food additives is unusual but can occasionally be established by placebo-controlled challenge testing, with confirmation by demonstrating reproducibility. Using this gold-standard procedure, up to 5% of CIU patients can be shown to have a specific reactivity to a food additive such as tartrazine, sodium benzoate, antioxidants and stabilizers (Black et al, 1991).

Chronic infections including *Helicobacter pylori* infection and infestations are, contrary to popular notions, not causative in CIU and any apparent association is coincidental. Similarly 'stress' and intercurrent acute infections may make any chronic urticaria worse, but are also not causative. However, up to 40% of cases of CIU are now known to have an autoimmune basis. This important subset of CIU will be discussed separately below.

Management of CIU

Irrespective of success in identification of a cause the strategy for treatment is the same. General measures are important. These include a cool ambient environment and avoidance (as far as is possible) of stress, intercurrent infections, aspirin, tight-fitting garments, alcohol consumption and exertion.

H_1 antihistamines remain the mainstay of drug treatment. They are more effective in alleviating the itch than the wealing. The timing of administration is influenced by the clinical periodicity of itching. In CIU itching predominantly occurs in the evening and at night (Sabroe et al, 1999a). While in some patients a single morning dose of a low sedation antihistamine such as loratadine 10 mg or fexofenadine 180 mg may be sufficient to bring about control throughout 24 hours, a supplementary dose of a sedative antihistamine such as hydroxyzine 25 mg at night is often necessary. Doxepin 25–50 mg is also useful as a night-time treatment. Doxepin is a tricyclic antidepressant with potent H_1 and H_2 antihistamine activity. However, because it is metabolized via the cytochrome P450 enzyme pathway it should not be concurrently administered with cimetidine. Addition of an H_2 antihistamine is rarely necessary. Carefully controlled clinical trials have shown a small statistically significant benefit by combining an H_1 and an H_2 antihistamine but the clinical benefit is dubious (Thomas et al, 1987).

Systemic corticosteroids should not be prescribed for long-term treatment of CIU since they almost invariably lead to poor control, escalating dosage and systemic toxicity. However, occasionally the author prescribes short tapering courses to meet the needs of emerging situations where rapid control is mandatory. A typical regimen would be prednisolone 30 mg daily for 3 days tapering by 5 mg every third day to zero. Ideally oral steroids should be administered as a singly daily dose early in the morning to minimize systemic toxicity.

What can be done for the severely affected patient who responds poorly to antihistamines? This is a problem for the specialist dermatologist or allergist. Oral cyclosporin is an option best addressed in a hospital setting, but is, in the author's experience, preferable as a long-term treatment to oral steroids. CIU is not a licensed indication for cyclosporin. Other manoeuvres such as increasing the dosage of fexofenadine well above the licensed maximum is best left to the specialist. Leukotriene antagonists are currently being advocated, particularly in the North American literature, but have not yet been exposed to the cold light of controlled clinical trial. *Table 4* outlines the routine management of CIU.

AUTOIMMUNE CHRONIC URTICARIA

Up to 40% of cases of CIU have an autoimmune basis, the weals being caused by a reaction between circulating IgG autoantibodies and specific epitopes expressed on the alpha-chain of the high affinity IgE receptor (FcεRI) of dermal mast cells and blood basophils, or, less commonly, with IgE itself. This observation, first reported by the author's laboratory (Hide et al, 1993; Niimi et al, 1996) and subsequently confirmed by a large number of investigators world-wide, has revolutionized the way CIU is regarded.

Clinically and histologically autoimmune CIU is indistinguishable from non-autoimmune CIU, although it tends to pursue a more severe and unremittant course (Sabroe et al, 1999b). However, autoimmune, but not non-autoimmune CIU has been shown to be associated with

marked basopenia (Sabroe et al, 1998), which could lead to a laboratory screening test for autoimmune CIU. Currently, the autologous serum (or plasma) skin test is the standard screening test for autoimmune CIU. Briefly serum is obtained at a time when the CIU is active and is injected intradermally into clinically uninvolved skin during remission.

A wealing reaction of diameter at least 1.5 mm greater than a control saline weal denotes a positive test suggestive of the presence of autoantibodies against FcεRI or less commonly IgE (Sabroe et al, 1999b). Confirmation is obtained by demonstrating that the patient's serum releases histamine from low and/or high IgE basophils of healthy volunteer donors. Some laboratories use Western blotting in preference to the basophil histamine release assay but the former may yield false positive results in certain other autoimmune diseases. Until a more convenient sensitive and specific assay is developed, the basophil histamine release assay remains the gold standard. However, the investigation of patients with CIU suspected of an autoimmune basis must remain at present within the province of the specialized laboratory.

The investigation is worth doing since severely affected patients are responsive to immunotherapy including cyclosporin, intravenous immunoglobulin and plasmapheresis. This subject has been reviewed (Greaves, 1995). However, it is important to appreciate that patients with autoimmune urticaria should routinely receive precisely the same general treatment, including the same antihistamines, as do patients with non-autoimmune CIU. It is only those severely affected and unresponsive to routine treatment who are considered for the above more drastic measures.

There remain at least 50% of all CIU patients in whom no cause whatever can be found. It is the author's belief, based upon circumstantial evidence, that many of these will turn out to have an autoimmune basis but direct evidence for this has yet to emerge, and must await improvement in the sensitivity of currently available laboratory assays. The clinical and laboratory features of autoimmune chronic urticaria are summarized in *Table 5*.

TABLE 4.
Routine management of chronic idiopathic urticaria (CIU)

General measures	Avoid aspirin, alcohol, stress, warm environment, tight-fitting clothes
Antihistamines	Low sedation in the morning +/– sedative at night
Recalcitrant CIU	Use doxepin 25–50 mg at night
	Add H_2 antagonist (ranitidine, not cimetidine)
Oral corticosteroids	Short tapering courses only

TABLE 5.
Clinical and laboratory features of autoimmune chronic urticaria

More protracted and severe course
Poorly responsive to routine treatment
Frequently associated with autoimmune thyroid disease
Positive autologous serum/plasma skin test
IgG autoantibodies with specific reactivity against FcεRI or IgE
Basopenia
Low histamine content of blood

URTICARIAL VASCULITIS

Urticarial vasculitis proves to be the correct diagnosis in at least 5% of patients referred with an initial diagnosis of CIU. It is extremely important to make this diagnosis for three reasons.

First, urticarial vasculitis can be an early manifestation of autoimmune connective tissue diseases including systemic lupus, Sjögren's syndrome and juvenile rheumatoid arthritis. It is also a recognized complication of hepatitis B and C infection and of paraproteinaemia. Second, the skin biopsy and confirmed finding of urticarial vasculitis should prompt a search for evidence of vasculitis in other organs and tissues of the body including the kidneys — although fortunately urticarial vasculitis is usually confined to the skin. Third, urticarial vasculitis requires a specific treatment strategy. Antihistamines are rarely effective alone and even systemic steroid therapy is usually disappointing. Recourse usually needs to be made to non-steroid anti-inflammatory drugs (see below).

The clinical features, more often than not, closely resemble those of CIU. However, there are some useful clinical pointers. Individual weals, unlike those of CIU, last longer than 24 hours, and may leave a stain in the skin as a result of red blood cell extravasation. The weals may be tender or even painful as well as itching and they often occur at sites of local pressure, mimicking delayed pressure urticaria.

Angio-oedema is common, and systemic symptoms, especially arthralgia and fatigue, are frequent. The diagnosis must be confirmed by skin biopsy which shows post-capillary venular endothelial cell damage, granulocyte disintegration releasing 'nuclear dust', red blood cell extravasation and fibrin deposition.

Management

The priority must be to seek underlying causes as listed above. General measures are as for CIU. H_1 antihistamines are almost always inadequate alone. No drug treatment is of proven efficacy in urticarial vasculitis, but anecdotally, and in the author's own experience, the following are worth trying.

Colchicine 0.5 mg should be administered three times daily with food. In some patients gastrointestinal side-effects limit its use, but apart from this, it is well tolerated. Alternatively dapsone is favoured by many clinicians. It is prescribed in an initial dose of 100 mg daily, although this can be later reduced. Since this drug causes haemolysis, especially in patients with genetic deficiency of glucose-6-phosphate dehydrogenase, the patients' phenotype for this enzyme needs to be checked and the dosage adjusted accordingly. It also causes methaemoglobinaemia and is unsuitable for patients with emphysema and ischaemic heart disease. Finally hydroxychloroquine is enthusiastically advocated by some, although the author has little personal experience of the drug in this context. The dose of hydroxychloroquine is 200–400 mg daily depending on lean body weight, and an initial baseline ophthalmological check for retinal function is important and should be repeated regularly.

Overall, urticarial vasculitis is a difficult condition to treat, and the author has accumulated several patients whose long-term management has proved less than successful. The main features of management of urticarial vasculitis are listed in *Table 6*. The subject of urticarial vasculitis has been well reviewed (O'Donnell and Black, 1995). HM

Conflict of interest: Professor Greaves has received occasional modest financial support from Aventis (formerly Hoescht Marion Rousell).

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TABLE 6.
Routine management of urticarial vasculitis

General measures as for chronic idiopathic urticaria
Search for causes (autoimmune connective tissue disease, hepatitis B/C infection, paraproteinaemia)
Try H_1 antihistamines first, if necessary in high dosage
In event of poor response, try colchicine 0.5 mg 3 times daily or dapsone 100 mg daily* or hydroxychloroquine 200–400 mg daily†

*check glucose-6-phosphate dehydrogenase phenotype first, and carry out regular full blood counts during treatment; † obtain ophthalmological checkup before and at regular intervals during treatment

KEY POINTS

- The term 'urticaria' encompasses a variety of different disorders including physical urticaria, autoimmune chronic urticaria, urticarial vasculitis and 'idiopathic' chronic urticaria
- Distinguishing these at the outset is very important from an investigational and therapeutic standpoint.
- Antihistamines remain, however,the mainstay of management of urticaria.
- 'Difficult' diagnostic and/or management problems require referral to a specialist dermatological or allergy unit.