Introduction

Urticaria is conventionally classified as acute, intermittent or chronic (Greaves, 2000a). The cause of acute urticaria is often recognized by the patient if it is due to an IgE-mediated hypersensitive reaction, and will not be considered further. Intermittent urticaria – frequent bouts of unexplained urticaria at intervals of weeks or months – will be discussed here on the same basis as the ordinary presentation of chronic urticaria but may have a definable external trigger, such as food or drug intolerance. The latter is conventionally defined as the occurrence of daily or almost daily whealing for at least six weeks with or without angioedema. The etiology of chronic urticaria has conventionally been regarded as obscure and hence the term idiopathic is often applied. However, there is increasing evidence that up to 50% of patients with ‘idiopathic’ chronic urticaria have autoimmune chronic urticaria. The diagnosis is important since it carries conceptual, prognostic and therapeutic implications. Contact urticaria and angioedema without wheals will not be dealt with in this account.

Classification of Chronic Urticaria

Chronic urticaria is defined in this chapter as any pattern of urticaria showing continuous activity for 6 weeks or more. In the latest European consensus classification of urticaria chronic urticaria the inducible urticarias (physical and cholinergic) are excluded from the definition of chronic disease (Zuberbier, 2009). The clinical subtypes of chronic urticaria are illustrated in the pie chart of Fig. 1. The frequency of these subtypes is based upon the authors’ experience at the St John’s Institute of Dermatology in UK. Whilst there may well be minor differences, it is likely that the frequency distribution of these subtypes will be essentially similar in most centres in Europe and North America (Greaves, 1995, 2000b). However, our experience suggests that the incidence of angioedema, especially that complicated by ordinary chronic urticaria is substantially lower in Japan and south Asian countries (Tanaka et al., 2006 and MWG unpublished observation).
Physical Urticarias

These comprise about one third to one fourth of all urticaria patients seen in the authors’ services. The diagnosis is mainly made by careful history taking, appropriate clinical examination and is confirmed by positive physical challenge testing. Updated guidelines for physical challenge protocols have been published recently (Magerl, 2009). It is important to identify patients in whom a physical urticaria is the main, if not the only, cause of the patient's symptoms. In this case, further investigation is not indicated, with rare exceptions (see below). Almost invariably, patients with this diagnosis are over-investigated by the time they are referred to the urticaria clinic. Skin prick testing, ImmunoCAP™ tests (previously known as RAST: radioallergosorbent tests), food exclusion diets, etc, are not normally indicated in patients with a physical urticaria with the exception of food-dependent exercise-induced urticaria, a rare variety of urticaria, which develops following exercise after food ingestion.

It is also important to appreciate that different physical urticarias can occur concurrently in the same patient; cold and cholinergic urticarias represent a well recognised example. Furthermore, chronic spontaneous urticaria is often associated with dermographism or delayed pressure urticaria. The investigation and management of chronic urticaria should be influenced by establishing the relative contributions of coexisting forms of chronic urticaria to the patient’s overall disability.

Symptomatic Dermographism

This common physical urticaria presents mainly in teenagers or adults of either sex. Like most physical urticarias, individual itchy wheals, produced by gentle stroking or rubbing of the skin, last less than 30 minutes before fading (Fig. 2). Angioedema and mucosal whealing do not occur but pruritus is troublesome and there are no systemic symptoms. The eti-