



European Dermatology Forum

Evidence-Based Guidelines for the Classification and Management of the
Photodermatoses,

*namely of skin disorders induced by ultraviolet or visible irradiation from sunlight or artificial
sources*

for the use of

*Consultant Dermatologists, Dermatologists in Training, Consultant Physicians, General
Practitioners, Dermatological Nurse Specialists*

and for the advice of

Patients

Section I

The Auto-Immune Photodermatoses

(formerly

The Idiopathic Photodermatoses)

*as based on an assessment of all the accessible published literature on the disorders in question
obtained through a Reference Manager-enabled online PubMed literature search, with irrelevant
or evidentially unacceptable references discarded*

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Overall Classification of the Photodermatoses

Increasing evidence suggests that the wide-ranging group of abnormal human skin responses to ultraviolet radiation (UVR) exposure comprise four categories, as follows, with any former names in bold in brackets after the new names:

1. The auto-immune photodermatoses (**the idiopathic photodermatoses**)
2. The DNA repair-defective photodermatoses
3. Drug- or chemical-induced photosensitivity disorders
 - i) exogenous
 - ii) endogenous (the porphyrias)
 - a) the hepatic porphyrias
 - b) the erythropoietic porphyrias

4) The photoaggravated dermatoses

The Auto-Immune Photodermatoses

This contribution now discusses the evidence base for the classification of the first, auto-immune group of these disorders and their management.

Polymorphic (Polymorphous) Light Eruption

Polymorphic light eruption (PLE) is a common acquired sunlight-induced disorder, particularly at temperate latitudes, where it affects some 10-20% of the population. It is normally characterized clinically by the occurrence of an itchy, erythematous, symmetrically distributed, papulovesicular rash of usually just some exposed areas within hours of UVR exposure, with full resolution in days to a week or two. There is epidermal spongiosis with a dermal, perivascular, mainly mononuclear cell infiltrate and oedema.

Pathogenesis

A delayed-type hypersensitivity (DTH) response to sunlight-induced cutaneous photo-antigen was suggested as the possible cause of PLE in 1942, based on the delay between UVR exposure and onset of the eruption, the occurrence of typical rash on previously affected but recently non-exposed sites, and the lesional histology (1). Modern research has now proved this contention almost certainly to be the case,

although the very strong evidence in favour still remains ultimately circumstantial because of the lack of irrefutable isolation of a responsible antigen.

In detail, characterization of the inflammatory infiltrate in naturally occurring PLE of uncertain and varying age remained inconclusive until serial biopsies from lesions artificially induced by low-dose solar-simulated irradiation demonstrated the consistent appearance of a T cell-dominated perivascular infiltrate within several hours, peaking at three days. CD4+ T cells were most numerous early on, whereas by three days, CD8+ T cells predominated, very probably helping obliterate the response (2, **A1**). Increased dermal and epidermal Langerhans cell and dermal macrophage numbers were also noted. This pattern of cellular infiltration was the same as previously documented in known DTH responses against confirmed antigens, particularly the allergic contact dermatitis and tuberculin reactions, indicating PLE to be almost certainly of the same nature.

Further work then demonstrated that intercellular adhesion molecule (ICAM)-1 was expressed in PLE in the keratinocyte layer, but just at sites overlying the perivascular infiltrate (3, **A1**). Similar findings had recently been reported to occur in and almost certainly to be specific for DTH, not occurring in irritant contact dermatitis or UVB-induced sunburn in normal skin (4,5), further supporting the same basis for PLE.

PLE patient epidermal cells irradiated with high-dose UVB and UVA were then noted to attract autologous peripheral blood monocytes but not unirradiated control cells (**B. 6**),

suggesting likely endogenous photo-antigen production by the irradiation. Such photo-altered and notionally foreign antigen could therefore induce a DTH reaction following sun exposure in PLE.

Hence several lines of evidence strongly suggest that PLE is an auto-immune response, with further work suggesting that only irradiated skin cells attract autologous monocytes, thus presumably reacting against photo-altered endogenous antigen,.

If PLE is indeed an auto-immune response to photo-altered endogenous antigen, a difference in immunological behaviour between irradiated PLE and normal skin should be demonstrable and this has now been shown to be the case. Thus, the induction of allergic contact sensitization, a DTH response, to dinitrochlorobenzene (DNCB) following solar-simulated irradiation of the sensitization site has been shown to occur significantly more easily in PLE patients than normals (7,8), indicating that PLE patient sensitisation to putative UVR-induced cutaneous antigen remains possible during their exposure because of continuing skin immunocompetence, whereas normal subjects immunosuppress during such exposure so as lose the ability to recognise antigen. The elicitation of the allergic contact response to DNCB in previously sensitized PLE patients and normals is however gradually and equally suppressed by irradiation (9, **A1**), presumably explaining the frequent development of immunological tolerance in PLE as summer progresses, as well as the efficacy of prophylactic phototherapy in the condition.

The precise radiation-absorbing molecules initiating the PLE rash have not been definitively identified, such that the very strong evidence in favour of a photo-induced immune response remains ultimately circumstantial, but this is almost certainly because a wide variety of molecules is likely to be involved in the same and different patients, and because these altered molecules very probably revert to normal within minutes to hours of their presumed photo-induced deformation.

Once such molecular absorption occurs, it would seem likely that either the altered molecule may remain deformed long enough to become antigenic, or else that it may re-emit the radiation to produce secondary antigenic molecular change, or instead free radical elements which interact with nearby molecules to produce antigen.

The radiation wavelengths required to initiate this putative antigen-forming process clearly vary between patients but remain uncertain, mostly because reliable artificial PLE induction is usually difficult, even in patients who react to very low doses of natural sunlight. This is probably because the relatively small areas of skin often irradiated during artificial exposure, especially if with inappropriate irradiation spectra, doses or dose rates, or on skin not normally affected by sunlight, are unlikely to develop sufficient presumed antigen to initiate a reaction. Finally, any cutaneous immunologic tolerance induced by recent sunlight exposure will again inhibit the presumed DTH response. Nevertheless, the eruption can certainly be artificially induced on frequent occasions without external elements other than irradiation being necessary, and UVA (315 to 400nm) has usually been more effective than UVB (280 to 315nm) (10,11),

being successful in over 50 percent of patients exposed to UVA or UVB daily for four to eight days in one study (12), UVB in 17 percent, and both in 27 percent. Another report (13), however, suggests that UVB may perhaps be effective in over 50 percent of patients. In very broad terms, therefore, taking all studies into account, it is clear that the eruption can indeed be induced artificially by irradiation, with about 50% of PLE patients seeming sensitive to UVB and 75% to UVA, including in each case approximately 25% sensitive to both, while visible light may also be responsible on very rare occasions (14). As a result, paradoxically, again supporting an immunological reaction, the use by PLE patients of sunscreens, which tend preferentially to remove the more immunosuppressive UVB while simultaneously permitting the passage of UVA, may have a significant PLE-enhancing effect, as often reported by patients.

The major predisposing factor to PLE appears to be genetic, one study suggesting perhaps 70% of all subjects have a tendency to the condition (15, **A2**). Its actual expression, however, appears likely to depend on both gene penetrance and sufficient initial UVR exposure to induce enough putative antigen for PLE sensitisation.

Treatment

Mild PLE is clearly controlled by the moderation of sun exposure, patients not exposed to appropriate light not apparently developing PLE (**D**), by wearing of appropriately protective clothing (**D**) and the regular application of broad-spectrum high-protection sunscreens, particularly against UVA, mostly UVB sunscreens often being ineffective, as shown in at least one controlled trial (15, **A1**).

Patients who develop their disorder only infrequently such as on vacations usually respond well to short courses of oral steroids prescribed to be taken with them in case their eruption develops (16, **A1**); if it does, around 25mg prednis(ol)one at the very first sign of itch (or more if necessary) and then each morning until clear is usually effective after at most several days, following which recurrence is relatively rare on the same vacation. Rare adverse effects of nausea and depression only occasionally necessitate stopping the drug. This treatment if well tolerated may be safely repeated every few months if required.

More severely affected subjects suffering repeated PLE attacks throughout the summer may require prophylactic low-dose photo(chemo)therapy (PUVA) courses in spring. This appears more effective than broadband UVB, controlling symptoms in up to 90% as compared with 60% of cases (17, **A1**). Narrow-band 312nm UVB phototherapy (so-called TL-01) is now probably the usual treatment of choice, however, being simpler to administer and safer than PUVA, although doses are low anyway, with at most a slightly reduced efficacy (18, **A1**). Prophylactic PUVA or UVB may sometimes trigger the eruption, particularly in severely affected subjects, but brief oral steroid therapy usually deals with this.

A small proportion of patients are unsuitable for, unable to tolerate or not helped by any of these measures, and for these, if severely affected, oral immunosuppressive

therapy, usually intermittent, with azathioprine or ciclosporin is generally helpful if the patient is a suitable candidate for such treatment (19, **C**, 20, **C**).

A series of other therapies have also been tried but are largely ineffective and should almost certainly be discarded except as last resort trials in patients not helped by any other therapy. These include the traditional hydroxychloroquine (21, **A1**), perhaps occasionally useful, beta-carotene (22, **A1**), probably never effective, nicotinamide (23, **D**), probably also never effective, and omega-3 polyunsaturated fatty acids, perhaps of moderate assistance in a few patients (24, **A2**).

Actinic Prurigo

Actinic prurigo (AP) is a rare, persistent, pruritic, excoriated, papular or nodular eruption of sun-exposed skin, with fading towards the non-exposed sites. It is generally worse in summer before improving, sometimes fully, in winter. Onset of the condition is usually in childhood, often with remission at puberty,

Pathogenesis

AP is UVR-induced, in that it is more severe in spring and summer, more pruritic after sun exposure and reasonably often demonstrates abnormal skin phototest responses to UVB or UVA irradiation, or both. In addition, sunlight exposure or solar simulated irradiation may sometimes induce a rash resembling PLE in AP patients, PLE is frequent in their families (15, **A1**) and a dermal, perivascular mononuclear cell infiltrate similar to that of PLE may occur in early AP lesions.

AP therefore appears to be a slowly evolving, excoriated form of PLE, and thus also a DTH reaction, again supported by the fact that many AP patients have close relatives with PLE (15, **A1**). In addition, human leukocyte antigen (HLA) DR4B1*0401 (DR4), present in some 30% of normal subjects, occurs in around 80-90% of those with AP, while HLA DRB1*0407, present in some 6% of normal subjects and not infrequently native Americans, occurs in around 60% (25, **A2**,26, **A2**,27 **A2**), such that this inherited feature may well be responsible for converting PLE into AP. In addition, some patients with the AP tissue type demonstrate clinical PLE but also have persistent lesions, while some with clinical AP convert to clinical PLE and some with clinical PLE change to clinical AP (26, **A2**), all further suggesting a relationship between the two disorders. The cutaneous molecular UVR absorbers responsible for initiating the eruption are not known but may well be diverse as suggested for PLE.

Treatment

The restriction of sun exposure and use of broad-spectrum, high-protection factor sunscreens may help milder cases, assisted by intermittent topical and perhaps rarely oral steroids, while oral thalidomide, generally in low doses (50 to 200mg at night) and preferably intermittently, is almost always effective for more resistant disease in all age groups within weeks. The published evidence for this is not strong (28, **B**) but it is clear to long-term experienced senior users of the drug that it is virtually always effective in adequate doses within about a month for this and other prurigo disorders. Adverse effects are generally mild and may include drowsiness, headache, constipation or weight gain, while careful nerve conduction studies every few months are important to

avoid a moderate, probably dose-related risk of slowly progressive peripheral neuropathy. Pregnancy must also be rigorously avoided because of the high risk of teratogenicity. If thalidomide is unavailable or unsuitable, phototherapy with narrowband UVB or PUVA may occasionally help (29, **B**), perhaps more reliably if the skin has been cleared first with oral steroids or thalidomide. Anecdotally, the topical calcineurin inhibitors, tacrolimus or pimecrolimus may also perhaps help on occasion if the skin is again cleared first (**C**), while oral immunosuppressive therapy with azathioprine or ciclosporin may well also be useful if the other therapies are ineffective, unsuitable or not tolerated (**C**).

Hydroa Vacciniforme

Hydroa vacciniforme (HV) is a very rare, chronic, scarring photodermatosis characterized by the occurrence of recurrent crops of papulovesicles and vesicles within hours of sun exposure on exposed sites, most commonly the face and dorsa of the hands, with fading to leave pock scars over weeks. It usually begins in childhood, may often remit at puberty and appears likely be a scarring variant of PLE.

Pathogenesis

The precise aetiology of HV is unknown. No chromophores have been identified and while the UVB MED reaction is normal in most patients, some have reduced UVA values (30, **B**). Blood, urine and stool porphyrin concentrations are normal, as are all other laboratory parameters, including circulating lupus titres. Nevertheless, the relationship of the eruption to sunlight exposure, its distribution and its early clinical

appearances are all very similar to those of PLE, strongly suggesting a possible relationship with that disorder. On the other hand, the fully developed HV eruption is much more severe than that of PLE, always being associated with permanent pock scarring and unresponsive to treatments effective for PLE, apart perhaps from sunscreens and occasionally prophylactic phototherapy. In spite of this, HV appears likely to be a scarring variant of PLE, and therefore possibly also a DTH reaction, with a tendency to scarring because the presumed endogenous antigen is located at a strategically important site such as the basement membrane, or the reaction is very severe, or a toxic photoproduct is released. Central American and Asian reports that the condition is frequently associated with Epstein-Barr virus infection probably refer to a similar but not identical condition (31,32,33).

Treatment

The treatment of HV consists of the restriction of sun exposure and use of High-protection broad-spectrum sunscreens. Occasionally, antimalarials and beta-carotene have been said to help, but this appears unlikely to be the case. As in PLE, prophylactic phototherapy with narrowband UVB or PUVA may sometimes be helpful, particularly the latter, but must be administered with care to avoid disease exacerbation (34). If conservative treatment is ineffective, however, as is often the case, topical or intermittent oral steroids, topical calcineurin inhibitors and perhaps oral immunosuppressive medication might perhaps be tried if clinically appropriate (C)..

Chronic Actinic Dermatitis

Chronic actinic dermatitis (CAD) is a rare, acquired, apparently not genetically based, sunlight-induced, persistent, usually eczematous, sometimes pseudolymphomatous eruption of the exposed skin, with corresponding histological features. It mostly affects older men with outdoor interests, although young atopics and rarely patients with human immunodeficiency virus infection are also susceptible. The disorder precisely resembles allergic contact dermatitis, a DTH reaction, and CAD therefore appears to be the same reaction, but on this occasion against endogenous, photo-induced, epidermal antigen.

Pathogenesis

The likely pathogenesis of CAD has steadily become clarified. Thus, detailed studies of its clinical, histological and immunohistochemical features all show it precisely to resemble the DTH reaction, allergic contact dermatitis (35, **A1**,36, **A1**,37, **A1**), even in its severe pseudolymphomatous form, formerly known as actinic reticuloid, in which the clinical and histological features are those of long-standing allergic contact dermatitis (38, **B**). It is therefore highly probable that CAD is a similar reaction, but in that it occurs with just irradiation and no contact allergen in place, it is presumably in this instance a reaction against photo-induced endogenous skin antigen.

If CAD is indeed such a response, it must follow either direct absorptive or secondary oxidative skin molecular distortion to form antigen, a process for the occurrence of which important support comes from the fact that albumin can become antigenic *in vitro* through photo-oxidation of its contained histidine (39). There is no evidence for a

genetic susceptibility to CAD, but a stimulus for development of its abnormal skin reactivity may conceivably be the frequent presence also of true allergic contact dermatitis, often airborne, to ubiquitous exogenous sensitizers or photosensitisers (40, **A1**,41, **A1**), which may conceivably predispose to CAD through sufficiently altering skin immune activity so as also to permit endogenous photo-antigen recognition. Long-standing, prior endogenous eczema (42, **A3**,43,**A2**), drug-induced photosensitivity (44, **B**), human immunodeficiency virus infection (45, **A2**) or possibly PLE (**C**) may also perhaps have the same effect. On the other hand, in addition or instead, chronic photo-damage in constantly sun-exposed elderly outdoor enthusiasts, who most often develop CAD, may arguably impair normal UVR-induced skin immunosuppression sufficiently for endogenous UVR-induced photo-antigen to be recognized, as apparently also occurs for genetic reasons in PLE (15, **A2**).

Assessment of the inducing action spectrum for CAD should theoretically help identify the postulated antigen, and this has been shown to resemble in shape that for sunburn in many patients (46, **A2**). However, the eruption in CAD is eczema, while much lower exposure doses are also often needed to evoke the response. Nevertheless, photobiological theory suggests that the UVR-absorber in such cases is the same as, or else a substance associated with, that causing sunburn, namely DNA, but in this instance acting as an antigen. In other CAD, however, it must be different, a few patients apparently reacting to just UVA (47, **B**), and perhaps a very few others to just 600nm visible light (48,**C**).

In spite of the foregoing, however, preliminary work in a single study using the autologous mixed epidermal cell leukocyte culture reaction did not detect antigen in CAD dermal or epidermal skin (49, **B**), although this may well have been because of imprecise experimental conditions. Nevertheless, final proof that CAD is indeed an UVR-induced endogenous antigenic process is still lacking.

In summary, CAD appears to be an allergic contact dermatitis-like reaction against UVR-altered DNA or a similar or associated molecule, or more rarely other molecules, perhaps as a result of airborne contact dermatitis-enhanced immune reactivity, or photo-damaged immunosuppressive activity, or both, in mainly long-standing sunlight- and airborne allergen-exposed subjects.

Treatment

The treatment of CAD is often difficult and frequently not fully effective. Rigorous avoidance of UVR and exacerbating contact allergen exposure is essential, along with the regular application of high protection factor broad-spectrum topical sunscreens of low irritancy and allergenic potential (**B**). Strong topical steroids such as clobetasol propionate are also often needed, and frequently produce marked symptomatic relief without adverse effects even if continued, provided their use is confined to affected skin; occasional oral steroid use is often helpful too for disease flares (**C**). In more resistant disease, the topical calcineurin inhibitors, tacrolimus and pimecrolimus, can produce good results if tolerated (50, **B**,51, **C**), but for refractory CAD, oral immunosuppressive therapy is almost always necessary and generally helpful if

tolerated. Thus, azathioprine 1.5-2.5 mg/kg/day often achieves remission in months (52, **A1**), when it may be reduced in dose or discontinued, often for the winter, while ciclosporin 3.5-5 mg/kg/day too is usually effective (53, **C**), but more likely to produce adverse effects, often renal, sometimes necessitating withdrawal. Mycophenolate mofetil is less often useful, while thioguanine has also been used with good effect (**C**). Finally, long-term, low-dose phototherapy with PUVA, usually several times weekly initially followed by maintenance exposures every three weeks or so may help (54, **B**), generally under initial oral and topical steroid cover to avoid disease flare.

Solar Urticaria

Solar urticaria (SU) is an uncommon, sunlight-induced, wealing disorder generally occurring spontaneously at any age and slightly more commonly in females. Very rarely it may be secondary to phototoxic drug use or cutaneous porphyria. It is an immediate type 1 hypersensitivity response against cutaneous or circulating photoallergen

Pathogenesis

Primary solar urticaria is an immediate type 1 hypersensitivity response against cutaneous or circulating photoallergen generated from a precursor following UVR or visible light absorption. Both circulating photoallergen and reaginic antibodies have been demonstrated. Very rarely, secondary SU may occur in association with drug photosensitivity, cutaneous porphyria or lupus. There appears to be no genetic basis for the condition.

Two types of primary solar urticaria have been proposed. Type 1 is an IgE-mediated hypersensitivity against specific photoallergens generated only in solar urticaria patients. Type 2 is an IgE-mediated hypersensitivity against non-specific photoallergen generated in both SU patients and normal subjects (55, **A1**). Therefore, in type 1 SU, passive transfer tests may be positive or negative, while in type 2 they are always positive. The wide range of responsible inducing wavelengths reported is presumably attributable to different photoallergens. Patients with type 1 appear to have photoallergens of molecular mass 25 to 34 kDa and an action spectrum in the visible region, while those with type 2 have photoallergens of molecular mass 25 to 1000 kDa and a variable action spectrum (56, **A2**). The range of eliciting wavelengths can narrow or broaden over months or years, presumably relating to decreasing or increasing photosensitivity respectively.

Exposure to long wavelength visible or UVA irradiation before, during or after the urticaria-inducing irradiation inhibits wealing in some patients, possibly by inactivation of the initial photoproduct or the inhibition of subsequent reactions (57, **A2**).

Conversely, Horio and Fujigaki (58, **A2**) reported a patient with SU induced by 320 to 420 nm light in whom pre-irradiation with 450 to 500 nm visible light augmented wealing. Post-irradiation with the same spectrum failed to increase the response, suggesting absorption of the long-wavelength radiation by a precursor to the photosensitizer altered it to become more reactive to the urticaria-eliciting radiation.

Although mast cell degranulation and histamine release are important in solar urticaria, antihistamine therapy (H_1 or H_2) is not always effective. This suggests other mediators such as neutrophil and eosinophil chemotactic factors accompanying histamine in the blood from irradiated skin can be important. Thus, the mast cell degranulation is accompanied by neutrophil and eosinophil recruitment and eosinophil major basic protein release, which may all amplify the wealing response (59, **A1**)

Treatment

Restriction of sun exposure, high-protection broad-spectrum sunscreen use and appropriate clothing cover may be helpful for UVA-sensitive but generally not visible light-induced SU, in which dark clothing is however better than light (**C**). Non-sedating, often higher-than-normal dose H_1 antagonists, best taken an hour or so before expected exposure and probably not helped by concurrent H_2 use, are very effective in about a third of patients (60, **A1**), and partially in a further third. In patients who develop SU tolerance as summer advances, prophylactic phototherapy may be helpful, and also sometimes in persistent disease, though the therapy then generally needs to be continued to maintain efficacy, with a consequent risk of long-term adverse effects. Such phototherapy should be undertaken with extreme care early on to avoid any risk of anaphylaxis, particularly in severely affected subjects (61, **B**). Multiple UVA exposures with increasing doses during the same day (so-called “rush hardening”) have helped some patients (62, **B**), while others respond to plasma exchange, or plasmapheresis, particularly if shown to have a circulating SU-associated serum factor by its intradermal injection after irradiation beforehand, remissions in some cases

being long-lived (63, **B**,64, **B**). Intravenous immunoglobulin has also been helpful on occasion (65, **C**), as rarely has oral ciclosporin (**C**).

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Summary Page

Evidence-Based Guidelines for the Classification and Management of the Photodermatoses

Section I

The Auto-Immune Photodermatoses (formerly the Idiopathic Photodermatoses)

These disorders, formerly called the idiopathic photodermatoses, now all appear to different levels of evidence to be autoimmune photodermatoses occurring as reactions against ultraviolet radiation-induced cutaneous photo-antigen. They are polymorphic (polymorphous) light eruption (PLE), actinic prurigo (AP), hydroa vacciniforme (HV), chronic actinic dermatitis (CAD) (formerly variously known as actinic reticuloid, photosensitive eczema, photosensitivity dermatitis or persistent light eruption) and solar urticaria (SU). Most now also have fairly to very good preventive or remedial therapies available.

The first three appear to be delayed-type hypersensitivity reactions (PLE level of evidence **B**, AP **B**, HV **D**) against perhaps predominantly dermal antigen, being largely dermal with important epidermal components, the fourth eczematous (**B**) against perhaps predominantly epidermal antigen, and the last an immediate hypersensitivity reaction (**B**) against probable dermal antigen. All except HV have strong levels of evidence in their favour, but have not been eligible for **A** levels because in no case has definite antigen been isolated, probably because many different potential antigens exist, although in CAD, there is strong circumstantial evidence for the nature of one antigen. For HV, however, the only evidence for an autoimmune pathogenesis is its very close clinical resemblance to PLE, but careful studies as for the other conditions have not been possible because of its extreme rarity and the fact that it almost always affects children.

The treatment for PLE is potentially effective, sunscreens with high UVB- and UVA-protective efficacy giving good if not always total protection (level of evidence **B**), while prophylactic courses of low-dose psoralen photochemotherapy (PUVA) or narrowband UVB (TL-01) prior to sun exposure give a high chance of minimal PLE for months thereafter (**A**). If rash does develop, short oral prednisolone courses (25-30mg daily) rapidly and safely ablate the rash (**A**), while azathioprine or cyclosporin immunosuppression for severe cases is also helpful if needed (**D GPP**). For AP, thalidomide is reliably useful (**GPP D**) as is other oral immunosuppression (**GPP D**), but HV responds at best minimally to therapy, except on occasion to UVB- and UVA-protective sunscreens (**C**). CAD responds well to oral immunosuppression with azathioprine (**A**) or cyclosporin (**GPP D**). SU responds in about half of cases to high-dose antihistamines (**B**) or failing that sometimes to long-term phototherapy (**B**), plasmapheresis (**C**) or intravenous immunoglobulin (**D**).

Patient Advice Sheet

On the Sunlight-Induced Skin Disorders and their Treatment

Section I

The Auto-Immune Photodermatoses (formerly called the Idiopathic Photodermatoses)

Polymorphic Light Eruption (PLE) (colloquially known as prickly heat), Actinic Prurigo (AP), Hydroa Vacciniforme (HV), Chronic Actinic Dermatitis (CAD) and Solar Urticaria (SU)

These disorders, formerly called the idiopathic photodermatoses, now all appear to be allergic, or so-called autoimmune, disorders, apparently occurring as reactions by the body's immune system against skin molecules altered by ultraviolet radiation (superficially penetrating UVB, deeply penetrating UVA, or both) from sunlight or sunbeds. The molecules presumably alter in all exposed people but the immune, or allergic, system in only some mistakenly recognises the changed molecules as foreign. Most of the conditions now have good treatments available.

The first three disorders (PLE, AP, HV) appear to be allergic reactions lasting days against fairly deep skin molecules, the fourth (CAD) an allergic reaction lasting weeks to months against superficial molecules, leading to a so-called eczema, and the last a short, only hour-long-lasting allergic hive- or weal-like reaction against probable deepish molecules. All except HV definitely appear to be allergic reactions, but in no case has a definite allergy-causing molecule been isolated, leaving some slight doubt, but this is probably because there are many different possible causative substances. For HV, however, the only

evidence for autoimmunity so far is its very close behavioural similarity to PLE, because it is too rare to study easily.

The treatment for PLE is usually very effective, modern sunscreens with high UVB- and UVA-efficacy, also now very cosmetically acceptable, often being helpful. If not, medically administered courses of low-dose ultraviolet treatment over several weeks before summer or holidays give a high chance of reducing or preventing the eruption for months. If rash does develop, occasional short oral steroid courses (25-30mg prednisolone daily for a few days) rapidly and safely clear the rash, while stronger carefully supervised treatment with azathioprine or cyclosporin for severe cases is also helpful when rarely needed. For AP, carefully administered thalidomide is reliably useful, as also probably is azathioprine or cyclosporin, but HV responds minimally to any therapy, except on occasion to highly protective sunscreens. CAD responds well to azathioprine or cyclosporin, while SU responds in about half of cases to high-dose antihistamines, or failing that, sometimes to long-term phototherapy, plasma exchange or intravenous immunoglobulin.