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2. Reading of the Source Article
3. Achievement of a 70% or higher on the online Case-based Post Test
4. Completion of the Journal CME Evaluation

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**Learning Objectives**
After completing this learning activity, participants should be able to recognize the clinical characteristics of immunologically mediated photodermatoses, drug- and chemical-induced photodermatoses, and cutaneous porphyrias in children; and delineate the appropriate diagnostic and management steps for each of these disorders.

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Photosensitivity disorders in children encompass a diverse group of diseases. Compared to adult patients, underlying systemic disorders, including genetic or metabolic defects, are common causes in pediatric photosensitivity disorders. Photosensitivity in a child should be suspected if the child develops a sunburn reaction in sun-exposed sites after limited sun exposure. Diagnosis of a photodermatosis is made based on careful history taking and a physical examination. Early recognition and prompt diagnosis are essential to minimize long-term complications associated with inadequate photoprotection. In part I of this continuing medical education article, immunologically mediated photodermatoses, photodermatoses caused by exogenous photosensitizers, and the cutaneous porphyrias will be covered. (J Am Acad Dermatol 2012;67:1093.e1-18.)

Key words: children; photodermatoses; photosensitivity; phototesting; polymorphous light eruption; porphyrias.

Ultraviolet (UV) radiation and visible light are portions of the spectrum of electromagnetic radiation, which is classified by wavelength (Table I). Ultraviolet C (UVC) light or germicidal radiation (200-290 nm) is absorbed completely by the ozone layer and does not reach the earth’s surface at sea level. Ultraviolet A (UVA) light has been divided into UVA I (340-400 nm) and UVA II (320-340 nm). Because of the proximity of the wavelength range of UVA II to ultraviolet B (UVB) light, as compared to UVA I, the biologic properties of UVA II are closer to that of UVB.

The depth of penetration of UV radiation into the skin depends on the wavelength. The longer wavelength UVA easily reaches the reticular dermis; shorter wavelength UVB light is absorbed in the epidermis and little reaches the papillary dermis. Even though only 2% to 3% of UV radiation from the sun reaches the earth’s surface, there are well described physiologic and pathologic cutaneous effects of UV exposure. UVB is the major spectrum responsible for cutaneous erythema or sunburn response, while UVA elicits a tanning response. Recently, visible light has been also shown to induce a tanning response that is more persistent than that induced by UV. Both UVA and UVB exposures are known to generate metalloproteinases and to induce immunosuppression, and they therefore play a role in photoaging and photocarcinogenesis.

Photodermatoses have their action spectrum in the UVA and/or UVB and/or visible light range. UVA light is involved in the majority of drug-induced photosensitivity reactions.

Photosensitivity can be defined as an abnormal or adverse reaction of the skin to UV or visible radiation. It usually follows exposure to sunlight, but rarely artificial light sources may be also responsible. Similar to adults, photosensitivity in children encompasses a diverse group of diseases. Many of them are the result of genetic or metabolic defects, and others may indicate an underlying systemic disorder. Photosensitivity in a child should be suspected if the child develops a sunburn reaction, swelling, or intense pruritus after limited sun exposure or develops an eruption, skin fragility, or scarring predominantly in sun-exposed areas (face, the V of the neck, and the dorsal surfaces of the hands and arms).

Photodermatoses disorders in children can be classified into 4 main categories: (1) immunologically mediated photodermatoses (IMPs; previously called idiopathic photodermatoses); (2) drug- or chemical-induced photosensitivity; (3) hereditary photodermatoses; and (4) photoaggravated dermatoses (Table II).

The incidence of photodermatoses in the pediatric population is much lower than in adults. From the Department of Pediatrics, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand, and the Department of Dermatology, Henry Ford Hospital, Detroit, Michigan.

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The distribution of photosensitivity disorders in children as reported in the literature is shown in Table III. Jansen reported 95 (26%) cases of a total 370 patients with photosensitivity disorders started before 15 years of age. Eighty-two percent of these cases were diagnosed as polymorphous light eruptions (PMLEs), and the remaining patients in this study had xeroderma pigmentosum (XP), erythropoietic protoporphyria (EPP), systemic lupus erythematosus (SLE), and pellagra. A study by Horkay et al collected 83 childhood-onset photodermatoses evaluated between 1967 and 2006 in Debrecen, Hungary, and found that the vast majority of photosensitivity cases in children were PMLEs, similar to the result of the previous report. However, Horkay et al found that the second most common cause was EPP. The latest literature in pediatric photodermatoses, published by Ten Berge et al, also reported that PMLE was the most common diagnosis (39% of patients). Interestingly, 24% of the patients had photosensitivity associated with atopic dermatitis, and an equal percentage had EPP. Photosensitivity induced by systemic or topical agents is relatively rare in children, but it has become increasingly common in recent years because of the wide use of photosensitizers in the environment.

**DIAGNOSTIC STEPS IN CHILDREN SUSPECTED PHOTOSENSITIVITY DISORDERS**

**Key points**
- History taking should include age of onset, seasonality, timing of development of lesions or symptoms after sun exposure, duration of the eruption, exposure to photosensitizers, and family history
- The physical examination should include careful observation on the distribution and morphology of lesions
- As appropriate, plasma porphyrins, autoimmune profile, a skin biopsy specimen, and phototesting may be useful

As in adults, the diagnosis of photodermatoses in children is made based on careful history taking and physical examination (Fig 1). Questions should include the age of onset of lesions, season of the eruption, timing of development of lesions or symptoms after sun exposure, the nature of the lesions, duration of the eruption, exposure to potential photosensitizers (both topical or systemic), systemic reviews particularly those suggestive of autoimmune disorder, and family history of photosensitivity and consanguinity. Because UVB light is filtered out by window glass, the development of an eruption after exposure to window glass-filtered sunlight would indicate action spectra in the UVA and/or visible light range. Patients with suspected photosensitivity should undergo a thorough physical examination, with a focus on the distribution of lesions, including the areas of sparing. In a photosensitivity disorder, relatively photoprotected sites, such as the upper eyelids, the postauricular area (Wilkinson’s triangle), and submental area, nasolabial folds and neck folds, the volar aspect of the wrist, and the antebrachial fossae tend to be spared. The morphology of the lesions may be helpful, such as urticarial lesions suggesting acute lesion of EPP or solar urticaria (SU), and atrophic scarring which could be associated with EPP and hydroa vacciniforme (HV).

Initial investigations should include a complete blood cell count and autoimmune profile, if appropriate, particularly in patients with systemic manifestation, such as arthritis or myalgia. Screening plasma porphyrin levels and, if available, spectrofluorimetric scanning of plasma porphyrins are recommended if one of the cutaneous porphyrias (CPs) is suspected. If screening plasma porphyrins are elevated, a quantitative porphyrin assay to determine biochemical defects in the erythrocytes, plasma, urine, and/or stool should be performed. Skin biopsy specimens for routine histopathology may be helpful in some diseases, such as PMLE, HV, and actinic prurigo. Phototesting is necessary in a minority of cases. It can be helpful in determining the cause of an acquired photodermatosis. Evaluation of growth parameters, developmental milestones, systemic involvement, and neurologic abnormalities are also helpful for identifying patients with genodermatoses associated with photosensitivity. Chromosomal analysis and molecular
genetic studies can confirm the diagnosis in many cases of hereditary photodermatoses. Phototesting, when practical, is helpful in many cases to determine the action spectrum.

In this review, we will discuss the prevalence, clinical manifestations, pathogenesis, investigations, and therapies of all pediatric photodermatoses. We divided our review into 2 parts. Part I focuses on immunologically mediated photodermatoses and drug- and chemical-induced photosensitivity. Part II covers hereditary photodermatoses and photoaggravated dermatoses.

IMMUNOLOGICALLY MEDIATED PHOTODERMATOSES

Key point

- Immunologically mediated photodermatoses include polymorphous light eruption, juvenile spring eruption, solar urticaria, hydroa vacciniforme, and actinic prurigo.

In children, IMPs, also previously known as idiopathic or primary photodermatoses, consist of PMLE, juvenile spring eruption (JSE), SU, HV, and AP. The exact pathophysiology of these IMPs has not been clarified. Immunologic mechanisms, autoimmunity, and genetic predisposition all seem to play a role.² UV-induced endogenous photoantigens likely represent the etiologic background of IMP; however, these antigens have not been identified.

Polymorphous light eruption

Key points

- Polymorphous light eruption is the most common pediatric photodermatosis
- Patients are less likely to be photoimmunosuppressed after sun exposure; this results in an enhanced immunologic response to cutaneous neoantigens generated by sun exposure

Table I. Electromagnetic radiation spectrum

<table>
<thead>
<tr>
<th>Waveband</th>
<th>Wavelength range (nm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>X-ray</td>
<td>0.1-1</td>
</tr>
<tr>
<td>Vacuum ultraviolet</td>
<td>10-200</td>
</tr>
<tr>
<td>Ultraviolet C</td>
<td>200-290</td>
</tr>
<tr>
<td>Ultraviolet B</td>
<td>290-320</td>
</tr>
<tr>
<td>Ultraviolet A</td>
<td>320-400</td>
</tr>
<tr>
<td>UVAll</td>
<td>320-340</td>
</tr>
<tr>
<td>UVAI</td>
<td>340-400</td>
</tr>
<tr>
<td>Visible light</td>
<td>400-760</td>
</tr>
<tr>
<td>Near infrared</td>
<td>760-1,000</td>
</tr>
<tr>
<td>Far infrared</td>
<td>1,000-100,000</td>
</tr>
<tr>
<td>Microwaves and radiowaves</td>
<td>&gt;10⁶</td>
</tr>
</tbody>
</table>

Table II. Classification of pediatric photodermatoses

<table>
<thead>
<tr>
<th>Immunologically mediated photodermatoses</th>
</tr>
</thead>
<tbody>
<tr>
<td>Polymorphous light eruption</td>
</tr>
<tr>
<td>Juvenile spring eruption</td>
</tr>
<tr>
<td>Actinic prurigo</td>
</tr>
<tr>
<td>Hydroa vacciniforme</td>
</tr>
<tr>
<td>Solar urticaria</td>
</tr>
<tr>
<td>Drug- and chemical-induced photosensitivity</td>
</tr>
<tr>
<td>Exogenous</td>
</tr>
<tr>
<td>Phototoxicity: systemic and topical</td>
</tr>
<tr>
<td>Photoallergy: systemic and topical</td>
</tr>
<tr>
<td>Endogenous: cutaneous porphyrias</td>
</tr>
<tr>
<td>Hereditary photodermatoses</td>
</tr>
<tr>
<td>Caused by defects in nucleotide excision repair</td>
</tr>
<tr>
<td>Xeroderma pigmentosum</td>
</tr>
<tr>
<td>Cockayne syndrome, including cerebro-oculo-facio-skeletal syndrome</td>
</tr>
<tr>
<td>Trichothiodystrophy</td>
</tr>
<tr>
<td>Ultraviolet light sensitive syndrome</td>
</tr>
<tr>
<td>Caused by double strand break repair defects</td>
</tr>
<tr>
<td>Rothmund–Thomson syndrome</td>
</tr>
<tr>
<td>Bloom syndrome</td>
</tr>
<tr>
<td>Caused by abnormal chemical substances</td>
</tr>
<tr>
<td>Smith-Lemli-Opitz syndrome</td>
</tr>
<tr>
<td>Hartnup disease</td>
</tr>
<tr>
<td>Others</td>
</tr>
<tr>
<td>Kindler syndrome</td>
</tr>
<tr>
<td>Photoaggravated dermatoses</td>
</tr>
<tr>
<td>Atopic dermatitis</td>
</tr>
<tr>
<td>Darier–White disease</td>
</tr>
<tr>
<td>Dermatitis herpetiformis</td>
</tr>
<tr>
<td>Herpes simplex infection</td>
</tr>
<tr>
<td>Lupus erythematosus and neonatal lupus erythematosus</td>
</tr>
<tr>
<td>Juvenile dermatomyositis</td>
</tr>
<tr>
<td>Pellagra</td>
</tr>
<tr>
<td>Psoriasis</td>
</tr>
<tr>
<td>Seborrheic dermatitis</td>
</tr>
</tbody>
</table>

Adapted from Lim and Hawk.²

- Management includes photoprotection and hardening with narrowband UVB treatment

Prevalence. PMLE affects both adults and children. A positive family history is reported in 3% to 56% of patients with PMLE.⁶ It has an inverse relation to latitude, with a prevalence in the adult population of approximately 21% in Scandinavian countries, 10% to 15% in the United States, 5% in Australia, and 1% in Singapore⁷; the low prevalence in Australia and Singapore may represent “hardening” occurring in patients living in sunny climates. It predominantly affects young females during the second and third decades of life; however, the age of onset in PMLE may range from childhood to late adult life. Horkay et al⁴ reported that 4% of 398 PMLE cases manifested
before 5 years of age and 10% between 6 and 14 years of age. In a study from Scotland, 20% of the patients presented before 10 years of age, whereas in Finland, approximately 25% of the cases occurred before 15 years of age.

**Clinical manifestation.** Eruptions in PMLE typically develop within a few hours after sun exposure, although less commonly they may appear within 20 to 30 minutes or 1 to 2 days after exposure. PMLE tends to be most severe at the beginning of the sunny season and becomes less severe as the sunny season progresses (a phenomenon known as “hardening”). Patients present with a nonscarring pruritic eruption on a sun-exposed area (Fig 2, A and B). There is a wide spectrum of clinical presentation, ranging from the common papular type to the relatively rare large papules, vesicular, plaque-like, urticarial, hemorrhagic, insect bite–like, and even erythema multiforme–type variants. In darker skinned patients, pinhead papular eruption is the most common morphology. In an individual patient, a single morphologic feature tends to present with each occurrence. Mild pruritus or a burning sensation may be experienced by affected subjects. The lesions are commonly symmetric, and in the absence of additional sun exposure resolve completely over several days. Associated systemic symptoms are quite rare; chills, headache, fever, and nausea have been reported, but may have been the consequence of accompanying sunburn.

**Investigations**

**Histology.** The basic features of the papular form of PMLE include edema, focal spongiosis, and occasionally small vesicles in the epidermis. Acanthosis, spongiosis, focal parakeratosis, and basal vacuolization can be present. Moderate to dense perivascular lymphocytic infiltrate is seen in the papillary and middle dermis, comprised predominantly of lymphocytes and, to a lesser degree, neutrophils and eosinophils. Direct immunofluorescence studies are negative.

**Phototesting.** Most PMLE patients have normal minimal erythemal doses (MEDs) to UVA and UVB and pigmentary responses to UVA. However, there are several reports showing decreased MED values either to UVA or UVB. Diagnosis can be confirmed by evaluating the patient after exposure to sunlight or by performing provocative phototesting in the clinic. The latter can be performed by exposing the skin either to multiples of MEDs or to suberythemogenic UV doses for 3 to 4 days followed by evaluation 1 to 2 days later for the development of characteristic PMLE lesions.

**Pathogenesis.** A delayed-type hypersensitivity (DTH) response to an endogenous, cutaneous UV-induced antigen has been considered in the pathogenesis of PMLE. It has been reported that patients with PMLE are less likely to be photoimmunosuppressed, resulting in an enhanced response to the cutaneous neoantigens generated after sun exposure. With repeated UV exposure, photoimmunosuppression occurs in these patients; this explains why patients tend to improve as the sunny season progresses or with UV desensitization therapy.

**Differential diagnosis.** The differential diagnosis of PMLE in children includes AD, SLE, EPP, SU, HV, and photosensitivity induced by exogenous agents. Careful history taking, physical examination, and appropriate laboratory evaluations should help in the diagnosis.

**Management.** Many PMLE patients are mildly affected. They can be satisfactorily controlled by proper photoprotection measures. Symptomatic treatment, including the use of topical corticosteroids, is helpful once the lesions have developed. In young, otherwise healthy adults who go on short winter holidays to a sunny region, a short course (5-7 days) of systemic corticosteroids (usually prednisone, 0.6-1 mg/kg/d) has been shown to be helpful (level of evidence, IB). Although this regimen has not been studied in the pediatric age group, it should

### Table III. Summary of the frequencies of childhood onset photosensitivity disorders

<table>
<thead>
<tr>
<th>Disease</th>
<th>Finland, 1981</th>
<th>Hungary, 2008</th>
<th>Netherlands, 2010</th>
</tr>
</thead>
<tbody>
<tr>
<td>Polymorphous light eruption</td>
<td>78 (82.1)</td>
<td>38 (46)</td>
<td>22 (39)</td>
</tr>
<tr>
<td>Erythropoietic protoporphyria</td>
<td>2 (2.1)</td>
<td>23 (28)</td>
<td>13 (23)</td>
</tr>
<tr>
<td>Xeroderma pigmentosum</td>
<td>13 (13.7)</td>
<td>3 (4)</td>
<td>1 (2)</td>
</tr>
<tr>
<td>Photosensitivity in atopic dermatitis</td>
<td>—</td>
<td>—</td>
<td>13 (23)</td>
</tr>
<tr>
<td>Photosensitive atopic dermatitis</td>
<td>—</td>
<td>—</td>
<td>8 (14)</td>
</tr>
<tr>
<td>AD with coexistence PMLE</td>
<td>—</td>
<td>—</td>
<td>5 (9)</td>
</tr>
<tr>
<td>Juvenile spring eruption</td>
<td>7 (8)</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>Systemic lupus erythematosus</td>
<td>1 (1)</td>
<td>1 (1)</td>
<td>1 (2)</td>
</tr>
<tr>
<td>Porphyria cutanea tarda</td>
<td>4 (5)</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>Solar urticaria</td>
<td>1 (1)</td>
<td>3 (5)</td>
<td>—</td>
</tr>
<tr>
<td>Hydroa vacciniforme</td>
<td>2 (2)</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>Phototoxic contact dermatitis</td>
<td>3 (4)</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>Pellagra</td>
<td>1(1)</td>
<td>1 (1)</td>
<td>—</td>
</tr>
</tbody>
</table>

AD, Atopic dermatitis; PMLE, polymorphous light eruption.
be considered in severely affected patients. Individuals with recurrent attacks may require a course of prophylactic narrowband UVB (NB-UVB) phototherapy or psoralen plus UVA light photother-apy (PUVA; 3 times per week for 5 weeks; level of evidence, IB), done in the early spring.7 Because of its known photocarcinogenicity, the use of PUVA in children should be done with caution. Other thera-pies, such as antimalarial agents, beta-carotene, and nicotinamide are of uncertain efficacy.

**Juvenile spring eruption**

**Key points**
- Juvenile spring eruption is considered a variant of polymorphous light eruption
- Juvenile spring eruption is most commonly seen on the top of the ears of young boys

**Prevalence.** In a prevalence study of children in New Zealand,13 the overall prevalence of juvenile spring eruption (JSE) was estimated to be 6.7%. The most commonly affected site is the top of an ear that is not covered by hair.

**Clinical manifestation.** JSE is considered a localized form of PMLE. This photodermatosis occurs more commonly in boys than in girls, and particularly between the ages of 5 and 12 years. The eruption typically occurs in the spring and consists of pruritic erythematous papules that are usually confined to the helix of the ears and that evolve into vesicles and crusts and heal with minimal or no scarring.

**Investigations**

**Histology and phototesting.** Stratigos et al14 reported the results of histologic findings and
Phototesting in 4 individuals with JSE. Histopathology revealed apoptotic cells and mild spongiosis in the epidermis, intraepidermal and subepidermal vesicles, and dermal mononuclear cell infiltrate. All patients had normal phototest results.

**Pathogenesis.** The etiology of JSE remains unknown, although it is probably similar to PMLE.

**Management.** Photoprotection and symptomatic treatment are usually sufficient for these patients.

### Solar Urticaria

#### Key points
- **Solar urticaria is a rare pediatric photodermatosis**
- **Solar urticaria represents a type I hypersensitivity response**
- **The management of solar urticaria includes photoprotection, antihistamines, and the careful use of narrowband ultraviolet B light hardening**

**Prevalence.** SU is an uncommon form of physical urticaria and is also a rare subtype of IMP. It has been described throughout the world. The age of onset of SU is variable. New cases have been described in ages ranging from 1 week old to the eighth decade of life. Most patients develop symptoms in young adulthood.

**Clinical manifestation.** SU is characterized by the development of wheal and flare, associated with pruritus and burning sensation, within 5 to 10 minutes after exposure to radiation. Individual lesions resolve within 24 hours. Headache, nausea, wheezing, dizziness, syncope, and, rarely, anaphylactic shock have been reported. The hardening effect has been observed in SU patients, but the mechanism of SU remains elusive.

**Investigations**

**Phototesting.** Phototesting serves as a provocative measure and a means to determine the eliciting action spectrum and the minimal urticarial dose, all of which can be of help in management. Caution should be exercised when performing phototesting in exposing only small portions of the skin; in patients who are highly sensitive to UV or visible light, unintended exposure of skin surface during phototesting may provoke an anaphylactic reaction. The majority of SU patients have broad-spectrum sensitivity, particularly UVA and visible wavebands. Less commonly, UVB and, rarely, infrared are responsible; the latter is probably more appropriately classified as heat urticaria.

**Pathogenesis.** SU represents an immediate type I or immunoglobulin E (IgE)-mediated hypersensitivity reaction to electromagnetic radiation. It is thought that a photoallergen is produced in the skin after exposure to the action spectrum. This is then recognized by the specific IgE to this allergen on the surface of mast cells, causing degranulation and the release of histamines and other mediators.

**Differential diagnosis.** This includes PMLE, EPP, and drug- or chemical-induced photosensitivity.

**Management.** Photoprotection is an important part of the management of SU. Relief of the symptoms can be achieved with topical corticosteroids or oral antihistamines. Hardening by exposure to UVA has been well described. While PUVA has been used in adults, its use in children should be reserved as a last resort because of its potential ocular side effects and long-term photocarcinogenicity. NB-UVB phototherapy is another treatment option (level of evidence, IIB); however, similar to phototesting,
care should be taken that a low starting dose and very graduated increases in the dose are done because treatment may trigger a generalized eruption resulting in anaphylaxis. To further minimize the side effects, only sites that are normally exposed to sunlight should be treated. Beta-carotene, antimalarial drugs, plasmapheresis (removal of a circulating serum factor/photoallergen), systemic immunosuppressive agents, and intravenous immunoglobulin (IVIG) have been reported to be effective in limited studies in adults. Similar to PUVA, plasmapheresis, IVIG, and systemic immunosuppressive agents should be used with caution in children.

Hydroa vacciniforme

Key points

- Hydroa vacciniforme is a rare photodermatosis that occurs almost exclusively in children
- After sun exposure, macules evolve into vesicles, which heal with varioliform scars
- Association with latent Epstein–Barr virus infection has been reported
- Management consists primarily of photoprotection

Clinical manifestation. HV is a rare photosensitivity disorder occurring almost exclusively in children. It usually begins in childhood and resolves spontaneously by early adulthood. The mean age of onset is 8 years. Boys are more often affected than girls. The cutaneous lesions in HV follow a distinct clinical pattern. Recurrent crops of discrete 2- to 3-mm erythematous macules evolve into blisters hours to a day or two after sun exposure. Healing occurs within days, with umbilication followed by crusting and pitted, varioliform scarring (Fig 3, A and B). This condition tends to appear each summer in children on uncovered parts of the body after exposure to sunlight. The face and dorsal surfaces of the hands are most frequently affected. Itching and burning and mild constitutional symptoms may occur a few hours before the outbreak of the cutaneous lesions. Uncommon presentations of HV include ocular involvement (keratoconjunctivitis and uveitis) and blistering of the lips. As in other photodermatoses, significant adverse effect on quality of life in children has been recently reported, with patients citing the inability to play outdoors as a major factor.

Investigations

Histology. Skin biopsy specimens of the early lesions reveal multilocular vesicles within the epidermis. The underlying dermis may present with an inflammatory infiltrate and also show hemorrhage and thrombosis of the vessels. In fully developed lesions, histopathologic findings reveal necrosis of the epidermis and dermis with mononuclear cell infiltrate.

Phototesting. Most patients show sensitivity to UVA radiation in monochromatic phototesting, which may induce papulovesicular lesions.

Pathogenesis. The pathophysiology of this disorder has not been clarified. An association between latent Epstein–Barr virus (EBV) infection and HV has been reported. EBV infection has also been well described to be associated with HV-like lesions in adults; these cases—some fatal—have been mostly reported from Mexico and Asian countries. It probably represents a different disease than that seen in children.

Differential diagnosis. This includes other blistering photosensitive disorders, such as EPP, vesicular PMLE, bullous SLE, and porphyria cutanea tarda (PCT).
Management. Photoprotection is the important component of the management of HV. No treatment has been universally successful. Chloroquine (level of evidence, IV), beta-carotene (level of evidence, IV), and PUVA (level of evidence, IV) have been effective in some cases, while thalidomide and cyclosporine are of unknown efficacy. In severe cases, systemic corticosteroid can also be used (level of evidence, IV).

Actinic prurigo

Key points

- Actinic prurigo is mostly seen in the Indian and Mestizo populations in Central and South America living at high altitudes, although it has also been reported in whites and Asians
- Actinic prurigo has a strong association with human leukocyte antigen DRB1*0407
- Cheilitis and conjunctivitis are common
- Thalidomide is the most consistently effective form of therapy

Prevalence. AP is most commonly seen in the Indian and Mestizo (mixed ancestry) populations of Mexico and Central and South America, especially individuals living at high altitudes (>1000 m above sea level); however, it has also been described in white and Asian populations.25 There is a strong association with human leukocyte antigen (HLA) subtypes. HLA DRB1*0407 is most prevalent, and is found in at least 60% to 70% of patients, but only a small percentage of controls (4-8% of DR4-positive patients).25,26 HLADRB1*0401 is the second most prevalent subtype, present in up to 20% of patients.27

Clinical manifestation. AP is characterized by the presence of intensely itchy papules, plaques, and nodules, along with excoriations and scars on the sun-exposed area. The onset is usually in childhood, but it ranges from 2 to 43 years.25 The mean age of onset is <10 years of age in a study from the United Kingdom,26 and at 4 to 5 years of age in the native Amerindian population.28 Clinically, AP causes a variety of lesions papules, plaques, and nodules (Fig 4). Some patients show secondary eczematization and lichenification. Very shallow linear, flat, or punctate scars may occur on the face. The eruptions usually manifest on the face, neck, extensor forearms, dorsal surfaces of the hands, and the upper aspect of the chest. However, in patients described in the United Kingdom, covered sites, including the back and buttocks, have been reported to be affected. Patients frequently complain about pruritus throughout the year with an exacerbation of the symptoms during spring or summer. Involvement of the lips and the conjunctiva is common, and it causes cheilitis, conjunctivitis, and pseudopterygium. Both the upper and lower lips can be affected, although it is found mainly on the lower lip.28 In addition, alopecia of the eyebrows has also been reported in association with AP. Spontaneous remission may occur in adolescence, particularly in patients with onset in childhood, but persistence is common.29 In Latin American patients living at high altitudes, relocation to a lower altitude area may result in resolution of the disease.

Investigations

Histology. The histology of skin lesions reveals a nonspecific subacute or chronic dermatitis, thickening of basement membrane, and dense dermal lymphocytic infiltrate. However, a biopsy specimen of the lip or conjunctiva usually reveals germinal centers, which is diagnostic.30

Phototesting. An Australian study by Crouch et al25 found lowered MEDs in approximately 60% of cases, with abnormal results in the UVA spectrum in all cases. In contrast, Hojyo-Tomoka et al28 from Mexico reported that their patients had abnormal response to photoprovocative test to both UVA and UVB, and had normal MEDs to UVA and UVB.

Differential diagnosis. The diagnosis is generally based on the clinical appearance and, if available, histologic findings; induction of lesions by repeated exposure to UVA or UVB can be performed and may achieve a positive result. The presence of mucosal and conjunctival involvement, persistence beyond 4 weeks, and residual scarring of skin distinguish actinic prurigo from PMLE. Other photodermatoses and photoaggravated dermatoses,
including HV, EPP, SLE, and atopic dermatitis with photosensitivity need to be considered. History, physical examination, and appropriate blood tests can usually differentiate AP from these other conditions.

Management. Photoprotection, including the use of sunglasses and photoprotective lip balms, is the most important factor in the treatment of AP. Topical corticosteroids, emollients, and oral antihistamines may be helpful for pruritus. Other treatments, such as antimalarial agents, beta-carotene, vitamin E, and pentoxyphillin are of uncertain efficacy. UV-based therapy, both NB-UVB (level of evidence, III) and PUVA (level of evidence, III) can be efficacious in clearing and preventing new lesions in some patients. Currently, thalidomide (level of evidence, III) is the most effective treatment in the majority of patients with AP. The dose of thalidomide in children is 50 to 100 mg per day, with maintenance achievable with doses as low as 50 mg per week. Some patients are able to stop the drug without a need for ongoing therapy. The main side effects of thalidomide are teratogenic effects and peripheral neuropathy; the former is not an issue for prepubescent children, and the latter rarely occurs in children.

**DRUG- AND CHEMICAL-INDUCED PHOTOSENSITIVITY**

**Photosensitivity by exogenous sensitizers**

**Key points**

- **Photosensitivity by exogenous sensitizers are rarely seen in the pediatric population**

- **Phytophotodermatitis is the most common type seen in children**

- **Identification and avoidance of the photosensitizer is treatment of choice**

In contrast to adults, photosensitive disorders caused by exogenous sensitizers are relatively rare in pediatric populations. Exogenous photosensitizers may reach the skin by topical or systemic routes. Pathophysiologically, it can be divided into phototoxicity and photoallergy, although clinically, it may not always be possible to differentiate between the two. Comparison between phototoxicity and photoallergy is shown in Table IV.

Almost all exogenous photochemical reactions are induced by UVA radiation. This is important because such radiation penetrates window glass. UVA is also present in lamps used in tanning booths, lamps used in nail salons for acrylic lacquer hardening (though only in relatively low doses), in dentist offices for bonding, and in small amounts in light emitted by photocopy machines and uncovered fluorescent bulbs. Patients may therefore have reactions while driving in a car with closed windows or while using a tanning salon. Another important clinical implication is that NB-UVB phototherapy is relative safe for most patients who are taking photosensitizing medications; this is because the light source used emits practically no UVA.

Plant-induced photosensitivity (phytophotodermatitis) is the most common phototoxic reaction of children. Citrus fruit, especially lemons and limes, along with parsnips, carrots, dill, parsley, figs, meadow grass, giant hogweed, wheat, clover,
cocklebur, buttercups, Shepherd’s purse, pigweed, and celery contain furocoumarin, a phototoxic agent. The UV-induced eruption usually begins the day after exposure to the furocoumarin and sunlight, ranges in severity from mild erythema to severe blistering, and eventuates in a characteristic hyperpigmentation. Linear streaks of hyperpigmentation, particularly on the face, chest, hands, and lower legs of children, are characteristic (Fig 5). Hyperpigmentation occurs over 1 to 2 weeks and can persist for 6 to 12 months.33

The incidence of phototoxic reaction and the frequency of reactions to drugs used in childhood are unknown. In all likelihood, the most common drugs responsible for this type of reaction in pediatric patients are antibiotics, including sulfonamides, quinolones, and tetracyclines. Systemic phototoxic agents include systemic antibacterial agents (predominantly doxycycline, tetracyclines, sulfonamides, nalidixic acid, and fluoroquinolones), antifungal preparations (griseofulvin), sulfonylurea hypoglycemic agents, furosemide, nonsteroidal antiinflammatory agents, amiodarone, quinine, isoniazid, and thiazide diuretics. As a personal gestalt on the incidence of photo–drug interactions in childhood, in 2 of our practices (Drs Shwayder and Lim) of >30 years each, we have seen 1 case of tetracycline-induced phototoxicity and no others. With regard to photoallergic reactions, organic UV filters used in sunscreens are currently the leading cause. Some of the commonly reported UV filters causing photoallergic reaction are octocrylene, benzophenone-3, and butyl methoxydibenzoylmethane.34 It should be noted that considering the extensive use of sunscreens worldwide, the frequency of photoallergy secondary to UV filters is uncommon.

The first line of treatment is removal of the photosensitizer; restriction of UV radiation exposure is needed until this is done. Good broad spectrum sunscreens need to be used because the action spectrum is in the UVA range.

**Endogenous chemical-induced photosensitivity: The porphyrias**

**Key points**
- While all cutaneous porphyrias have been reported in the pediatric population, the most common ones are erythropoietic protoporphyria and congenital erythropoietic porphyria
- Cutaneous manifestations range from a burning and stinging sensation, edema, skin fragility, and blisters to scarring
- Because the action spectrum is in the visible light range, only physical photoprotection is an effective form of photoprotection

The porphyrias are a group of inherited and acquired disorders that are caused by a specific enzymatic defect in the heme biosynthesis pathway (Fig 6).35 Heme is assembled from simple precursors (glycine and succinyl coenzyme A synthetase) in an 8-step pathway. Each step is catalyzed by a different enzyme. In CPs, enzyme deficiency results in an accumulation of tetrapyrrole heme precursors, porphyrinogens, which are spontaneously oxidized to the corresponding porphyrins. The toxic profile of the accumulate intermediate determines the variety of clinical features, ranging from acute neurovisceral attacks, skin lesions, or liver disease. Most enzyme defects represent partial deficiency, because a complete enzyme deficiency along the heme pathway is not compatible with life.36

Heme synthesis starts in the mitochondria; the subsequent steps, from deamination, tetrapyrrole ring formation, to successive decarboxylation to 4-carboxyl porphyrinogen (coproporphyrinogen) take place in the cytoplasm. The final stages occur in the mitochondria (Fig 6). Heme is mostly produced in the erythrocytes for hemoglobin synthesis and in the hepatocytes for synthesis of cytochromes and hemoproteins.

Porphyrias are commonly classified as either hepatic or erythropoietic, depending on the principal organ in which heme precursors accumulate. Alternatively, they can be classified as acute porphyrias, CPs, and rare recessive porphyrias according to the type of clinical presentations.35 However, some have both acute neurovisceral and cutaneous manifestations.

Porphyrins, the oxidized products of heme precursor-porphyrinogens, are potent photosensitizers responsible for photosensitivity in patients.
with CPs. The major action spectrum of the porphyrins ranges from 400 to 405 nm (the Sorbet band), with a minor action spectrum range at 600 to 650 nm (visible red); this is the reason commercially available sunscreens, which only protect against UVB and UVA, are not helpful for patients with CPs. Dihydroxyacetone, an agent responsible for sunless tanning, has a proven benefit in blocking longer UVA and visible light. It can provide excellent protection in cases of CPs. Cutaneous presentations in porphyrias range from acute burning pain and edema within a few minutes after sun exposure in EPP to chronic skin fragility and blistering of the sun-exposed area without obvious relation to sun exposure, such as in PCT. These varied clinical manifestations result from the difference in water solubility of the porphyrinogens. Sequential decarboxylations—from uroporphyrinogen (8-carboxyl porphyrinogen) to coproporphyrinogen (4-carboxyl porphyrinogen) and to protoporphyrinogen (2-carboxyl porphyrinogen)—are associated with a progressive decrease in the water solubility of the porphyrinogens. In PCT, water-soluble uroporphyrin diffuses throughout the skin up to the dermoepidermal junction, so the light-induced reaction occurs in the papillary dermis, resulting in skin fragility and blisters. In contrast, in EPP, protoporphyrin accumulates in erythrocytes, and, being lipid soluble, does not diffuse further than the endothelial lining of the blood vessels. Therefore, the phototoxic reaction causes endothelial necrosis in the papillary dermis, resulting in the typical pain and edema of EPP.

In the subsequent paragraphs, we will focus on porphyrias with cutaneous involvements that can be present in childhood: congenital erythropoietic porphyria (CEP), EPP, PCT, hepatoerythropoietic porphyria (HEP), hereditary coproporphyria (HC), and variegate porphyria (VP).

### Congenital erythropoietic porphyria

**Key points**

- **Congenital erythropoietic porphyria** is the most frequent of the rare autosomal recessive porphyrias, and the most disfiguring
- It presents with dark colored urine, severe photosensitivity and scarring, and may lead to the development of hepatosplenomegaly and hemolytic anemia
- **Bone marrow or stem cell transplantation is curative**

CEP, also known as Günther disease, is a rare autosomal recessive disease; however, it is the most frequent of the rare recessive porphyrias. It is the most severe disfiguring form of porphyrias; it is
caused by the deficiency of uroporphyrinogen III synthase (UROS). This deficiency results in a shift from production of III to production of I isomers of uroporphyrinogen and coproporphyrinogen (Fig 6). This leads to overproduction and excretion of the nonphysiologic and pathogenic isomer I of uroporphyrinogen and coproporphyrinogen. The human UROS gene has been assigned to the chromosome region 10q25.3 to q26.3. Mutations in affected individuals are heterogenous. These include missense and nonsense mutations, large and small deletions and insertions, splicing defect, and intronic branch mutations. However, a common missense mutation, C73R (Cys → Arg at position 73), is identified in up to 40% of white patients; this mutation is associated with <1% of UROS activity. In addition, CEP cases associated with UROS deficiency secondary to a GATA-1 erythroid–specific transcription factor gene mutation have also been reported.

The disorder is characterized by the appearance of dark colored urine during infancy, which fluoresces red upon exposure to a Wood’s lamp (Fig 7, A and B), severe photosensitivity that occurs during the first 2 or 3 years of life, splenomegaly, and hemolytic anemia. Affected cases become symptomatic in infancy. Patients show signs of discomfort after sun exposure, although they cannot verbally communicate. Phototherapy during the neonatal period for hyperbilirubinemia may lead to generalized blistering. When teeth erupt, they are dark colored, and fluoresce under examination with a Wood’s lamp because porphyrins bind to dental calcium phosphate (erythrodontia; Fig 8, A and B). Most patients have severe photosensitivity, leading to the recurrent eruption of vesicles and bullae filled with fluid that fluoresces pink and eventually resulting in mutilating ulceration, scarring, and loss of acral tissues (Fig 9, A and B). Other common clinical features include hypertrichosis of the face and extremities and hyperpigmentation (Fig 9, C). Ocular involvement includes photophobia, corneal scarring, ulceration, ulcerative keratoconjunctivitis, and cataracts. The majority of patients have hemolytic anemia with associated hypersplenism. Bony involvements include osteodystrophia, combining osteolysis and osteoporosis; hypercellular bone marrow is present in most patients. These changes may lead to bone fragility and fracture.

Biochemical studies show the marked elevation of uroporphyrin I and coproporphyrin I in circulating erythrocytes, bone marrow cells, plasma, urine, and feces. Exposure to a Wood’s lamp may reveal reddish orange porphyrin fluorescence in urine or aqueous suspensions of feces; thin smears of peripheral erythrocytes glow red under a fluorescent microscope. Patients have normochromic anemia with increased reticulocyte count. A skin biopsy specimen may be helpful, but may only differ in severity from other forms of porphyria. Identification of responsible mutations is helpful for genetic counseling, and genotype—phenotype correlation can now be achieved. Prenatal diagnosis is now available.

Treatment consists of rigorous sun avoidance including avoidance of light through window glass, wearing protective clothing, and the use of opaque sunscreens containing nonmicronized titanium dioxide or zinc oxide that block in the visible spectrum. Beta-carotene has been used and shows some photoprotective effect. Hypertransfusion and splenectomy may be necessary in cases with severe hemolytic anemia. Bone marrow and allogeneic stem cell transplantation (level of evidence, IV) may correct all disease manifestations except erythrodontia. The prognosis of the severe form of CEP is poor. Death, when it occurs, is frequently associated with hemolytic anemia.

**Erythropoietic protoporphyria**

**Key points**
- Erythropoietic protoporphyria is the most common form of childhood porphyria
- Patients complain of a burning and stinging sensation upon sun exposure
Thickening of the skin on the knuckles and subtle scarring on the bridge of the nose can be seen.

Inheritance is complex, with 2 molecular defects in the ferrochelatase gene.

A much less common autosomal recessive form has been described, with a higher risk of severe liver disease.

EPP is the most common form of porphyria in childhood. It is caused by partial deficiency of the ferrochelatase (FECH) enzyme; clinical presentations appear only when FECH activity is <50% of normal function. EPP is an inherited disorder, however, the mode of inheritance is complex. The FECH gene has been cloned and mapped to the long arm of chromosome 18q21.3. EPP is almost always associated with 2 molecular defects. In about 94% of patients with overt disease, clinical expression usually requires coinheritance of a private FECH mutation and the low expression FECH*IVS3-48C allele. The effect of this coinheritance is to lower mitochondrial ferrochelatase activity below a crucial threshold of about 35%. In a UK study, autosomal recessive EPP without an IVS3-48C allele has been identified, with a prevalence of 3%. Two of the 7 families with autosomal recessive EPP had liver disease, suggesting that this form of EPP carries a higher risk of severe liver disease than other forms of EPP.

Accumulation of free protoporphyrin, mainly in erythrocytes and secondarily in other tissues (skin and liver) or biologic specimens (serum, bile, and feces), leads to painful photosensitivity and potential liver complications. EPP usually becomes symptomatic between 1 and 6 years of age because of the photosensitivity. The average age of presentation is 4 years. Cutaneous photosensitivity in EPP usually presents with immediate painful reaction within minutes of sun exposure. There is severe burning pain, especially on the dorsal surfaces of the hands and feet and the face, which is usually unresponsive.

Fig 8. Congenital erythropoietic porphyria. A, Teeth showing pink to brown color. B, The teeth fluoresce pink under examination with a Wood’s lamp because porphyrins bind to dental calcium phosphate (erythroodontia). (Photographs courtesy of Dr Wisuthsarewong, Bangkok, Thailand.)

Fig 9. Congenital erythropoietic porphyria. A, A 10-month-old white female with mutilating ulceration and scarring secondary to recurrent eruption of vesicles and bullae. B, Hypertrichosis on the face when the same patient was 28 months old (after bone marrow transplantation).

• Thickening of the skin on the knuckles and subtle scarring on the bridge of the nose can be seen.
to any analgesic drugs except cold air and cold water. Crying upon exposure to sunlight frequently occurs in young children. The burning pain is associated with erythema and edema in the exposed area, which could be followed by ecchymosis. Photoonycholysis may occur. Chronic lesions are common, such as thickening of skin on the knuckles and subtle scarring on the bridge of the nose (Fig 10). Increased rugosity of the upper cutaneous lip can also be seen. The severity of the photosensitivity may vary over the years. Hypochromic and microcytic anemia have been reported. Liver dysfunction can be identified in 10% to 20% of EPP patients. An increase incidence of cholelithiasis is also present. In about 2%, a rapidly progressing and irreversible cholestatic liver failure develops. 

Laboratory findings include elevated free protoporphyrin levels in circulating erythrocytes, plasma, bone marrow cells, and feces. Because protoporphyrin is highly lipophilic, urinary porphyrin levels are normal, unless the patient is in hepatic failure. Examination of the peripheral blood smear under a fluorescence microscope reveals fluorescence of the erythrocytes. Fluorescence of teeth and nails is not present. Plasma fluorescence emission spectroscopy shows a characteristic peak at 634 nm. 

Based on a systematic review, the available data are insufficient to prove efficacy of any treatments studied so far in EPP. The management of EPP is dependent on photoprotection and the use of opaque sunscreens. The administration of betacarotene (30-150 mg in children) may be considered, although its efficacy has been questioned. Other therapeutic approaches include cholestyramine (4 g/day), which helps prevent reabsorption of protoporphyrin excreted into the intestinal lumen, and chenodeoxycholic acid (15 mg/kg/day), which is a primary bile acid and help to inhibit the production of protoporphyrin in the liver. Recently, subcutaneous implants of alfa-melanocyte stimulating hormone analog, which induces a tanning response, have been successfully used to increase sun tolerance in a pilot study. For patients with progressively deteriorating liver function, liver transplantation can be lifesaving. To prevent potentially severe phototoxic reaction in the exposed organs in the operating room, all surgeries need to be performed under a light source that emits minimally in the blue spectrum of the visible light, which is the major action spectrum of porphyrin-induced phototoxicity; shielding of organs and skin should be done if possible.

Porphyria cutanea tarda and hepatoerythropoietic porphyria

Key points
- Porphyria cutanea tarda is the second most common porphyria in children; hepatoerythropoietic porphyria is a homozygous form of porphyria cutanea tarda
- Cutaneous manifestations include skin fragility, blisters, and hypertrichosis
- In children, the treatment of choice is low dose hydroxychloroquine

PCT is the most common form of porphyria in adults and the second most common form in children. Heterozygous deficiency in uroporphyrinogen decarboxylase causes PCT, whereas homozygous deficiency in this enzyme results in HEP. Estimated to have an incidence of 1 in 25,000, PCT usually presents during the third and fourth decades of life. Most patients have sporadic disease (type I, sporadic PCT), and the enzyme deficiency is limited to the liver. About 20% to 30% of the patients have mutations in the gene for uroporphyrinogen decarboxylase enzyme (type II, familial form), resulting in approximately 50% reduction of the enzyme activity in all tissue. Another variant of PCT (type III) is characterized by a family history of the disease although it is biochemically indistinguishable from sporadic PCT, with enzyme deficiency limited to the liver. The familial subtype has an earlier onset and arises equally in both sexes; it is inherited in an autosomal dominant manner with low penetrance.

PCT always presents with cutaneous symptoms only. Skin fragility is perhaps the most common presentation. Skin lesions in PCT are erosions, vesicles, or bullae on sun-exposed cutaneous surfaces, especially the dorsal aspects of the hands and forearms. The blisters vary markedly in size from 1 mm to 3 cm. Secondary infection may occur. The lesions heal with hyperpigmentation; milia frequently occur. Periorbital hypertrichosis and mottled hypopigmentation are frequently present. Uncommonly, sclerodermoid skin changes may be
observed; this is the only cutaneous manifestation of PCT that can be seen in the sun-exposed and also in photoprotected sites. Rare ocular complications have been reported, such as ocular pain, photophobia, and perforation of sclera. HEP has similar but usually more severe presentations; in some cases, it can result in early onset sclerodermoid changes on fingers and hands. Variable degrees of liver dysfunction are frequent in patients with this disorder. In adults, PCT can be precipitated by, or associated with, iron overload, hepatitis C, alcoholic cirrhosis, hemodialysis, administration of estrogens, hydantoins, or griseofulvin, HIV infections, and hemochromatosis.

Confirmation of the clinical diagnosis of PCT can be made by increased levels of plasma, urinary and fecal porphyrins, with a predominance of uroporphyrin (8-carboxyl porphyrin) and 7 carboxyl porphyrin in plasma and urine, and isocoproporphyrin in feces. Erythrocyte porphyrin levels are normal. Fluorescence emission spectroscopy of plasma shows a characteristic peak at 620 nm. A biopsy specimen of vesicular lesions reveals subepidermal bullae with dermal papillae arising irregularly from the floor of the bulla into its cavity (“festooning”). Periodic acid–Schiff positive material is deposited around blood vessels and sometimes at the dermoeidermal junction.

As with other porphyrias, sun avoidance, the use of protective clothing, and the use of opaque sunscreens are crucial to lessen skin symptoms. Predisposing factors should be eliminated. In childhood PCT, low-dose hydroxychloroquine treatment (3 mg/kg twice a week; level of evidence, IV) is widely used. Clinical improvement, which precedes biochemical improvement, is observed in 4 to 6 months. Urinary and plasma concentration of porphyrin should be monitored every 3 months and usually returns to normal within 6 months. Liver function test should be monitored regularly (every few months or annually, depending on the degree of abnormality) in these patients. Phlebotomy is the treatment of choice in adult cases, but it is difficult to perform in children. Therefore, it should be reserved for pediatric patients who do not respond to antimalarial agents. Spontaneous remission with advancing age may occur. In adults, 5% to 16% of patients with long-standing untreated disease and associated chronic active hepatitis develop hepatocellular carcinoma.

Hereditary coproporphyria

Key points

- Patients with hereditary coproporphyria present with neurovisceral complaints; 20% to 30% have skin lesions resembling those of porphyria cutanea tarda

Childhood onset of hereditary coproporphyria has only rarely been reported

HC is an autosomal dominant porphyria with primary neurovisceral manifestations caused by the deficiency of coproporphyrinogen oxidase (CPO). Rare homozygous variants have been described. HC has almost always been described in adults. The few cases that presented during childhood were initially mistaken for HV.

The acute neurologic and abdominal attacks in HC are generally identical to acute intermittent porphyria but are usually less severe. Twenty to thirty percent of patients are photosensitive; these patients have skin lesions resembling those of PCT. The diagnosis is made after finding elevated levels of coproporphyrin III in plasma, urine, and feces.

Treatment consists of the avoidance of agents known to induce acute neurovisceral attacks, especially drugs, including estrogen and progesterone. The avoidance of exposure to sunlight, sun-protective clothing, and the regular application of opaque sunscreens are mandatory. During an acute attack, precipitating factors must be identified; their avoidance is a must. Neurologic symptoms like abdominal pain, nausea, and vomiting should be treated symptomatically (eg, by the administration of opioid analgesic drugs or chlorpromazine). The most important therapeutic step is the early intravenous administration of hemin preparations.

Variegate porphyria

Key points

- Patients with variegate porphyria have clinical manifestations similar to those seen in HC
- Variegate porphyria is rarely seen in pediatric patients

VP is an autosomal dominant disorder caused by the decreased activity of protoporphyrinogen oxidase (PPO). This disease has its onset after puberty and generally appears in the fourth to fifth decades of life; only a few cases in childhood have been described.

Acute VP resembles acute intermittent porphyria in terms of neurovisceral clinical features. Sixty percent of adult patients manifest with cutaneous lesions. The cutaneous features of VP are similar to those of PCT, but tend to be milder and less easily provoked. Porphyrin profiles show the predominance of 4-carboxyl and 5-carboxyl porphyrins in the urine, which contrast with the predominance of 8-carboxyl porphyrin (uroporphyrin) and 7-carboxyl porphyrin seen in PCT. Plasma fluorescence...
emission spectroscopy reveals a characteristic peak at 624 to 627 nm. Treatment is similar to that of patients with HC, consisting of the avoidance of sunlight and precipitating drugs.

**CONCLUSION**

Photosensitivity in a child should be suspected if the child develops a sunburn reaction, swelling, intense pruritus, skin fragility, or scarring after limited sun exposure predominantly in sun-exposed areas. The incidence of immunologically mediated photodermatoses in children is lower than in adults, and systemic disorders, such as genetic or metabolic defects, should be considered, especially in patients who develop the symptoms early in life. PMLE is the most common pediatric photodermatosis, while photosensitivity induced by systemic or topical agents is relatively uncommon. The diagnosis of photodermatoses in children is usually established by careful history and physical examination. Skin biopsy specimens for routine histopathology and phototesting may be helpful in some diseases.

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