Seven types of porphyria are recognized. The light sensitivity in the six types with cutaneous lesions is caused by protoporphyrin IX, a compound that occurs in the skin. These wavelengths lie in the 400-nm range, representing long-wave ultraviolet light (UVA) and visible light.
In *erythropoietic porphyria*, a very rare disease that typically develops during infancy or childhood, recurrent vesiculobullous eruptions in the skin are common. This condition is characterized by an autosomal recessive pattern. Hypertrichosis and brown-stained teeth that fluoresce are additional features.

In *erythropoietic protoporphyria*, the usual reaction to light (erythema and edema followed by thickening and superficial scarring of the skin) is common. However, in rare instances, fatal liver disease develops quite suddenly, usually in persons of middle age but occasionally in patients only in the second decade of life.
In *porphyria variegata*, different members of the same family may have either cutaneous manifestations identical to those of *porphyria cutanea tarda*. Also, a sharp fluorescence emission peak at 626 nm is specific for the plasma of *porphyria variegata*.
Three forms of *porphyria cutanea tarda* can be distinguished: sporadic, familial, and hepatoerythropoietic. In the sporadic form, only the hepatic activity of uroporphyrinogen decarboxylase is decreased. Almost all patients are adults, and no clinical ...
In hereditary coproporphyria, a very rare disorder, there are episodic attacks of abdominal pain and a variety of neurologic and psychiatric symptoms.
Histopathology.

The histologic changes in the skin lesions are the same in all six types of porphyria.

In mild cases, homogeneous, pale, eosinophilic deposits are limited to the immediate vicinity of the blood vessels.
In severely involved areas, which are most common in erythropoietic protoporphyria, the perivascular mantles may take on a distinctive appearance. These areas can be stained with Sudan IV or Sudan black B. In addition, the PAS-positive dermal-epidermal basement membrane zone may be thickened.
In areas of sclerosis, which occur especially in porphyria cutanea tarda, the collagen bundles are thickened.

The bullae, which are most common in porphyria cutanea tarda and least common in erythropoietic protoporphyria...
dermal papillae often extend irregularly from the floor of the bulla into the bulla cavity (104,122). This phenomenon, referred to as festooning, is explained by the rigidity of the upper dermis induced by the presence of eosinophilic material within and around the capillary walls in the papillae and the papillary dermis.

The epidermis forming the roof of the blister often contains eosinophilic bodies that are elongate and sometimes intracellularly or extracellularly; and (c) electron-dense material thought to be of basement membrane origin (124).
Pathogenesis

The substance around dermal vessels has the appearance of hyalin because it consists of homogeneous, eosinophilic ... in large amounts in the dermis of lipoid proteinosis and produced by fibroblasts as amorphous material, is absent.

On electron microscopic examination, concentric duplications of the basement membrane around the dermal vesse
Proof that the perivascular material in porphyria represents excessively synthesized basement membrane material.
In the majority of patients, direct immunofluorescence testing has revealed the presence of immunoglobulins, particularly in the sun-exposed areas of the skin. Rather than indicating an immune phenomenon; rather, they are the result of "trapping" of immunoglobulins and complement in the filamentous material.

The enzymatic defect that causes each form of porphyria is known. Enzyme determination...
Liver damage is generally mild and chronic in porphyria cutanea tarda. In erythropoietic protoporphyria, liver involvement can be serious with jaundice and even acute liver failure.
Porphyria Cutanea Tarda = اﻠآﺠﻠﺔ اﻠﺠﻠدﻴﺔ اﻠﺒورﻔﻴرﻴﺔ
Porphyria Cutanea Tarda

Pseudoporphyria Cutanea Tarda
In patients with chronic renal failure who are receiving maintenance hemodialysis, an eruption indistinguishable from porphyria cutanea tarda may not be representative of the porphyria metabolism, and the plasma and fecal porphyrins should always be measured.
Pseudoporphyria cutanea tarda may also occur following the ingestion of certain drugs, such as furosemide.

**Histopathology**

In patients with pseudoporphyria, the histologic picture is indistinguishable from that seen in mild cases of porphyria.
immunoglobulins are often observed in vessel walls and at the dermal-epidermal junction. Complement is
Porphyria Cutanea Tarda = اﻠآﺠﻠﺔ اﻠﺠﻠدﻴﺔ اﻠﺒورﻔﻴرﻴﺔ