Seven types of porphyria are recognized. The light sensitivity in the six types with cutaneous lesions is caused by the formation of porphyrins, which absorb light at wavelengths 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 skin may occur. In an autosomal recessive pattern, hypertrichosis and brown-stained teeth that fluoresce are additional features.

In erythropoietic protoporphyria, the usual reaction to light is erythema and edema followed by thickening and superficial scarring of the skin. 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.
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. There are also cutaneous manifestations indistinguishable from those of porphyria cutanea tarda and porphyria variegata.
Histopathology

The histologic changes in the skin lesions are the same in all six types of porphyria. Differences are noted in the severity of the lesions. In mild cases, homogeneous, pale, eosinophilic deposits are limited to the immediate vicinity of the blood vessels in the papillary dermis. These deposits are best visualized with a PAS stain, being PAS positive and diastase resistant.
In severely involved areas, which are most common in erythropoietic protoporphyria, the perivascular mantle may contain iron deposits, which can be identified by staining with Sudan IV or Sudan black B. In addition, the PAS-positive dermo-epidermal basement membrane zone may be thickened.
In areas of sclerosis, which occur especially in porphyria cutanea tarda, the collagen bundles are thickened. In contrast to scleroderma, PAS-positive, diastase-resistant material is often present in the dermis in perivascular locations.

The bullae, which are most common in porphyria cutanea tarda and least common in erythropoietic protoporphyrin.
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, amorphous material that is absent in large amounts in the dermis of lipoid proteinosis and produced by fibroblasts as amorphous material. On electron microscopic examination, concentric duplications of the basement membrane around the dermal blood vessels are seen. Intermingled filamentous and amorphous material is seen throughout the upper dermis and even in the mid dermis.
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 IgG, in the skin of patients with porphyria cutanea tarda. Although the immunoglobulins appear to be directly involved in the skin lesions, they are not the primary cause of the disease. 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 deficiencies result in a failure of the normal pathway of porphyrin metabolism and lead to accumulation of the corresponding porphyrin in the erythrocytes, urine, and feces.
Liver damage is generally mild and chronic in porphyria cutanea tarda. In erythropoietic protoporphyria, liver damage may be more severe and chronic.
Porphyria Cutanea Tarda = اﻠآﺠﻠﺔ اﻠﺠﻠدﻴﺔ اﻠﺒورﻔﻴرﻴﺔ
Porphyria Cutanea Tarda = اﻠآﺠﻠﺔ اﻠﺠﻠدﻴﺔ اﻠﺒورﻔﻴرﻴﺔ
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, because in drug-induced cases, withdrawal of the drug is curative.

**Histopathology**

In patients with pseudoporphyria, the histologic picture is indistinguishable from that seen in mild cases of porphyria. However, additional features are seen in pseudo-porphyria cutanea tarda. The histologic picture reveals festooned dermal papillae. The blisters usually are situated above the PAS-positive basement membrane zone.
immunoglobulins are often observed in vessel walls and at the dermal-epidermal junction. Complement is