

# Prurigo Pigmentosa

Prurigo Pigmentosa (PP), also known as *Nagashima Disease*, is a rare inflammatory dermatosis that is commonly associated with the ketogenic diet, earning it the colloquial name "keto rash." The ketogenic diet is characterized by a low-carbohydrate, high-saturated fat regimen. While PP can affect individuals on various types of diets or with underlying conditions, its strong correlation with ketosis, particularly from the ketogenic diet, has been widely recognized. PP is characterized by the appearance of itchy pink papules, macules, or vesicles that progress into a distinctive brown reticulated pattern. This review explores the clinical features, pathogenesis, and current treatment strategies for PP.

## Clinical Features

PP predominantly affects young females, likely due to the higher prevalence of dieting behaviors among this demographic. However, individuals of all ethnic backgrounds can develop the condition, with a noted higher frequency in individuals of Asian descent. The hallmark feature of PP is the progressive development of pink, itchy papules, which may evolve into vesicles or macules that eventually form brown, net-like patterns as the lesions resolve.

Initially, the lesions present as erythematous, pruritic papules that later crust or scale. Over a period of weeks, the lesions gradually resolve and coalesce into a brown reticulated pattern due to the deposition of melanin and other pigments. Symmetry in lesion distribution is typical, with the trunk and neck being the most commonly affected areas, though lesions can also be seen on the face, scalp, and suprapubic region. Notably, PP spares the mucous membranes, hair, and nails.

PP has several overlapping features with other inflammatory dermatoses, leading to frequent misdiagnoses. Conditions such as urticaria, drug eruptions, dermatitis herpetiformis, eczema, contact dermatitis, and confluent and reticulated papillomatosis can resemble PP both clinically and histologically.

## Etiology and Triggers

The precise cause of PP remains undetermined, though various mechanical, hormonal, and metabolic factors have been implicated in its development. A particularly notable trigger is ketosis, fasting, insulin-dependent diabetes mellitus, or bariatric surgery. In the context of the ketogenic diet, the rash can appear as early as six days to as long as four months after initiating the diet, although the timing can vary widely.

Other recognized triggers include:

- Sweating, often exacerbated by the restrictive nature of the ketogenic diet.
- Physical irritation, such as rubbing from tight clothing or friction.
- Chromium exposure from acupuncture needles.
- Underlying conditions such as atopic dermatitis, diabetes mellitus, anorexia nervosa, rapid weight loss, pregnancy, Sjogren's syndrome, and *Helicobacter pylori* infection.

The association between ketosis and PP underscores the importance of considering metabolic and dietary factors during the diagnostic process. Accurate diagnosis is contingent on reviewing the patient's dietary history and identifying any triggering events that may have preceded the onset of the rash.

## Diagnosis

Diagnosis of PP is based on clinical presentation, patient history, and histopathological findings. The hallmark features—itchy papules evolving into a brown reticulated pattern—are highly suggestive of PP. If the patient is on a ketogenic diet or has other triggering factors, the diagnosis becomes more straightforward. Histopathology, while not always required, may assist in confirming the diagnosis, particularly in cases where the clinical picture overlaps with other dermatoses.

## Treatment Options

The management of PP focuses on addressing the underlying cause, particularly the cessation of ketosis. If the rash is associated with the ketogenic diet, the first line of treatment is to discontinue the diet, as resolution of ketosis is essential for the remission of PP. If ketosis persists, the rash is likely to recur.

The primary treatments for PP include:

- **Tetracyclines:** Oral minocycline or doxycycline are commonly used in the management of PP. These antibiotics inhibit neutrophil chemotaxis and function, potentially preventing the formation and worsening of lesions. They are considered first-line treatments, particularly in more severe or persistent cases.
- **Topical and Systemic Corticosteroids:** While corticosteroids are commonly used in inflammatory dermatoses, they are generally ineffective in resolving PP. This may be due to the unique pathophysiological mechanisms at play, which do not respond to standard anti-inflammatory treatments.
- **Other Treatments:** Although oral antibiotics such as tetracyclines are often effective, other potential options include phototherapy or the use of topical immunomodulators in refractory cases, though further studies are required to establish their efficacy.

## Prognosis

The prognosis of PP largely depends on its association with ketosis and the timing of dietary changes. If the rash is related to the ketogenic diet, discontinuation of the diet generally results in resolution of symptoms. Recurrence is possible if ketosis is reintroduced, but with appropriate management, the condition is self-limiting in most cases.

For individuals who experience recurrent or more severe episodes, the use of antibiotics may help to reduce symptoms and prevent further progression of the rash.

## Conclusion

Prurigo Pigmentosa (PP) is a rare but distinctive inflammatory dermatosis, most commonly associated with the ketogenic diet, but also triggered by a variety of metabolic, hormonal, and mechanical factors. The condition typically presents with pruritic papules that evolve into a brown reticulated pattern. Diagnosis is largely clinical, supported by histopathological features, and treatment involves discontinuing the ketogenic diet, alongside the use of oral tetracyclines to manage inflammation. As more is understood about the triggers and mechanisms of PP, further advances in targeted therapies may improve the management of this condition.

## References

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