The Prevalence and Disease Characteristics of Generalized Pustular Psoriasis (GPP)

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GPP is a rare and potentially life-threatening disease. While it shares some signs and symptoms with less serious forms of psoriasis, GPP is a separate condition and requires an accurate diagnosis

What are the characteristics of GPP?

1 **Recurrent or intermittent disease flares**

Large areas of painful, non-infectious pustules

May co-present with

Fever · Chills · Loss of appetite

2 **Can require emergency treatment**

3 **Can present at any age, including in children**

~50 years

Median age at diagnosis

4 **IL-36 pathway implicated**

Some patients have mutations in genes associated with the IL-36 pathway

→ Overexpression of IL-36 cytokines

→ GPP skin lesions

5 **Regional differences in frequency of IL-36 receptor antagonist gene (*IL36RN*) variants**

34.7% Europe

28.8% Malaysia

46.8–60.5% China

How common is GPP?

Estimating the number of individuals who are affected by GPP is challenging

? Misdiagnosis
? Different data sources
? Lack of awareness
? Non-standardized methodologies
? Low patient numbers

Published GPP data are highly variable…

1.76 France

7.46 Japan

88–124 Republic of Korea

Cases per million persons

…and may be indicative estimates only

How to differentiate GPP from plaque psoriasis?

- GPP can manifest with or without concomitant plaque psoriasis
- GPP is a distinct clinical entity from plaque psoriasis with different genetic markers, and a differential diagnosis is critical

Several key factors exist to differentiate GPP from plaque psoriasis

IL36RN variants and genetic markers

Flare recurrence

Fever

Leukocytosis with neutrophilia and elevated erythrocyte sedimentation rate

Inflammatory cell infiltrate

Edema

Respiratory, cardiac, or renal failure

No transient primary pustule formation at plaque periphery

Neutrophilic cholangitis

Increased alkaline phosphatase, transaminase, and bilirubin levels