

# Improving the Understanding, Diagnosis, and Management of Generalized Pustular Psoriasis (GPP)



## Clinical Discussion Guide: Explaining GPP to Patients

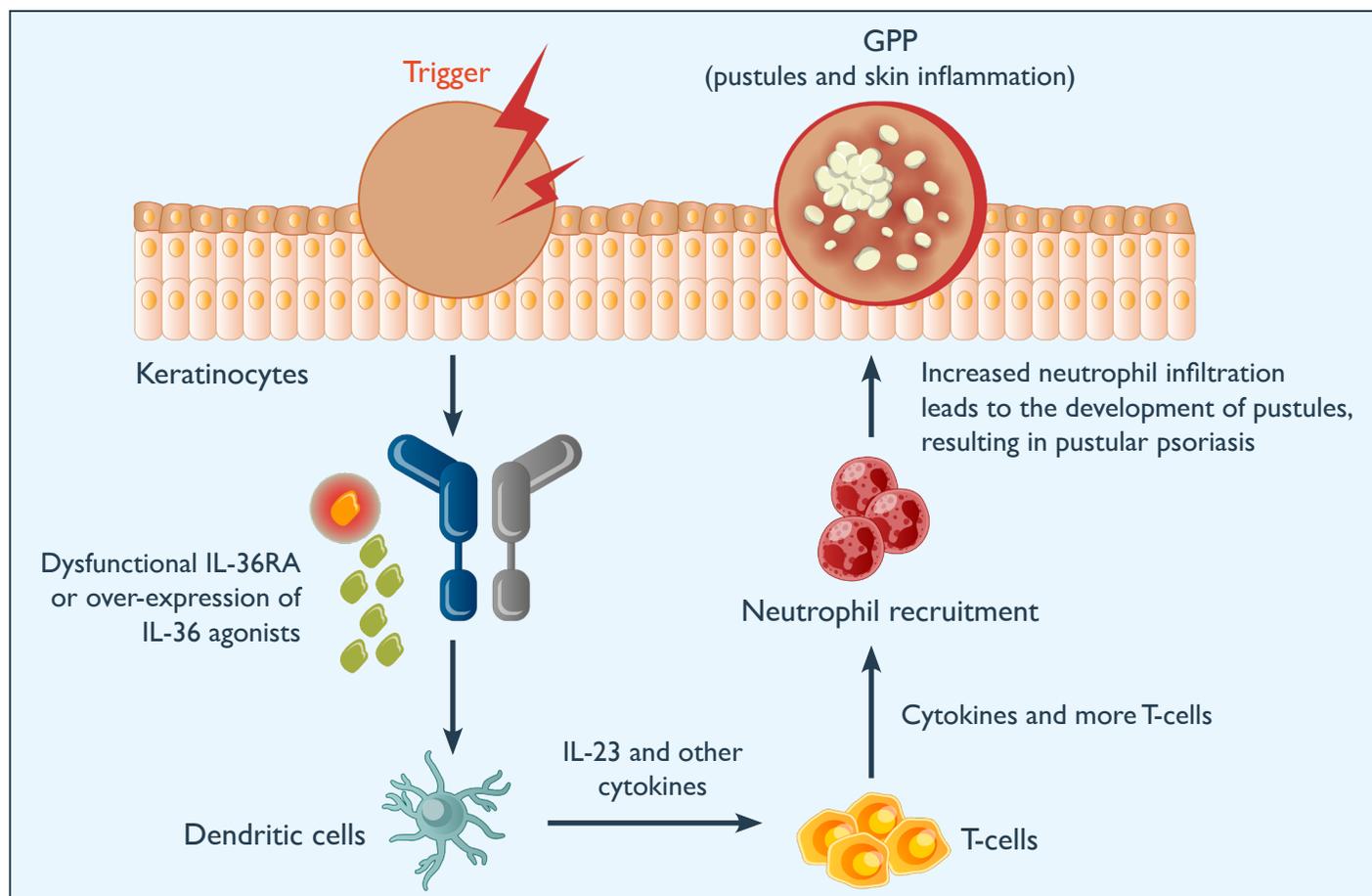
### Purpose of This Guide

This is a resource to facilitate point-of-care discussions with patients and families about generalized pustular psoriasis (GPP). It may be used with children, adults, or caregivers—either when a person is first diagnosed or later.

### Information to Convey to Patients and Families

#### Mechanism of action

- The pathogenesis of GPP can be influenced by genetic mutations in genes that mediate inflammation; cytokine signaling, including overactivation of the inflammatory IL-36 cascade, and environmental factors. These factors include viral infections, medications, and corticosteroid discontinuation.<sup>1</sup>



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### Epidemiology and Clinical Features

- Pustular psoriasis is a rare and unique form of psoriasis, accounting for only ~1% of all cases seen in clinical practice<sup>2</sup>
- Pustular psoriasis can affect children or adults of any age and is commonly observed in infancy or among adults between the ages of 40 and 50 years<sup>2,3</sup>
- Pustular psoriasis is more common in women than men, and is more commonly observed in individuals of Asian descent compared to other ethnicities<sup>2,3</sup>
- People with pustular psoriasis have red, tender skin that is covered with small, yellowish, and neutrophil-rich blisters called pustules that can appear anywhere in the body<sup>3,4</sup>
- GPP is a serious form of pustular psoriasis. It is characterized by pustules that cover large areas of the arms, legs, or body. GPP can occur suddenly and cause potentially life-threatening complications that require emergency medical treatment<sup>3,4</sup>

There are 4 subtypes of GPP seen in clinical practice<sup>2</sup>:

Subtype	Name	Characteristics
1	Acute GPP, von Zumbusch	The most common type in adults (~66%); characterized by sudden onset of widespread, erythematous patches/plaques accompanied by multiple pinhead-sized sterile pustules that can cover the whole body, causing erythroderma. Systemic symptoms include fever, leukocytosis, and general malaise
2	Annular GPP (erythema annulare centrifugum-like psoriasis cum pustulatione)	A subacute, low-grade disease characterized by gyrate lesions and annular pustules and usually with only minor systemic involvement
3	GPP of pregnancy (impetigo herpetiformis)	A rare gestational dermatosis most commonly seen in the third trimester of pregnancy with rapid resolution following birth
4	Juvenile and infantile GPP	Mostly benign, with a higher remission rate and spontaneous resolution rate than annular GPP

### Impact of comorbidities

The most common comorbidities observed in patients with GPP include diabetes, obesity, hypertension, and hyperlipidemia. As a result of its multiple complications, GPP can be potentially life-threatening during an acute flare. GPP complications can include cardiovascular failure, secondary infections such as pneumonia and sepsis, aseptic acute respiratory distress syndrome, and neutrophilic cholangitis.<sup>2</sup>

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### Questions That Patients May Have and Potential Responses

QUESTION/CONCERN	RESPONSE
Can this be cured?	GPP cannot be cured. However, like most chronic conditions, it can be controlled with appropriate medication and the management of triggers
Can I continue to play basketball/sports?	Yes, you can continue to do all the activities you love to do. When you have a flare of pustular psoriasis, you may not feel up to being too active. The goal is to reduce the frequency and length of your flares, so you can enjoy your favorite activities
How can I reduce the frequency of flares?	Patients with GPP tend to have flares that include fever, pain, and even mood changes. Everybody is different in what they experience, and it is important to have a treatment plan in place that works specifically for you. We can work together to create a plan that works for you and helps control your flares
No one else in my family has this problem; why do I have it/how did I get it?	GPP is a very rare disease. It is estimated that 1 out of every 10,000 people in the US have GPP. While we don't exactly know what causes it, we know some people are genetically prone. Others tend to have flares driven by certain medications, stress, and even pregnancy
How long should I expect to have these symptoms?	The hope is you can start a treatment that allows for optimal disease control
Can I spread this to other people?	No, GPP is not contagious. You cannot catch it from someone else, nor can it be spread to anyone
Are there resources to help me stop smoking?	Yes, there are several resources that can help you quit smoking. We can discuss a plan that works for you and even discuss medications, like nicotine patches and Chantix, for example. The CDC and The American Lung Association can also provide guidance: <a href="https://www.cdc.gov/tobacco/campaign/tips/quit-smoking/index.html">https://www.cdc.gov/tobacco/campaign/tips/quit-smoking/index.html</a> <a href="https://www.lung.org/quit-smoking">https://www.lung.org/quit-smoking</a> <a href="https://smokefree.gov/">https://smokefree.gov/</a>

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### Strategies and Tips for Speaking With Patients and Families

**Use plain language and diagrams.** The AHRQ Health Literacy Universal Precautions Toolkit is a useful resource: <https://www.ahrq.gov/health-literacy/improve/precautions/toolkit.html>

**Verify patients' understanding** of information using teach-back <https://www.ahrq.gov/health-literacy/improve/precautions/tool5.html>

#### Smoking Cessation:

- **I-800-QUIT-NOW:** (1-800-784-8669) (in English)
- **I-855-DÉJALO-YA:** (1-855-335-3569) (in Spanish, en Español)
- **Asian Language Quitline:** 1-800-838-8917 (in Chinese, 中文) 1-800-556-5564 (in Korean, 한국어) 1-800-778-8440 (in Vietnamese, Tiếng Việt)

### Resources for Patients and Families

- **National Psoriasis Foundation:** <https://www.psoriasis.org/>
- **CDC Tips on Smoking Cessation:** <https://www.cdc.gov/tobacco/campaign/tips/quit-smoking/index.html>
- **American Lung Association Tips on Smoking Cessation:** <https://www.lung.org/quit-smoking>

### References and Resources for Clinicians

1. Marrakchi S, Puig L. [Pathophysiology of Generalized Pustular Psoriasis](#). *Am J Clin Dermatol*. 2022;23(Suppl 1):13-19. doi:10.1007/s40257-021-00655-y
2. Boehner A, Navarini AA, Eyerich K. [Generalized pustular psoriasis - a model disease for specific targeted immunotherapy, systematic review](#). *Exp Dermatol*. 2018;27(10):1067-1077.
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4. Mirza HA, Badri T, Kwan E. [Generalized Pustular Psoriasis](#). [Updated 2022 Sep 12]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan.
5. Blair HA. Spesolimab: [First Approval](#) [published correction appears in *Drugs*. 2023 Jan;83(1):103]. *Drugs*. 2022;82(17):1681-1686.

**Disclaimer:** This Clinical Discussion tool is for educational purposes only and does not constitute medical advice.