



# Generalized Pustular Psoriasis Caregiver Booklet





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As a caregiver, I know accompanying and supporting a family member or friend with **Generalized Pustular Psoriasis (GPP)** is a challenging journey. GPP is a rare and complex disease, which affects physical health and places a heavy strain on a person's mental and emotional well-being. My role isn't just to help manage the disease, but also to provide the emotional support they need so they never feel alone.

This guide is designed to help caregivers like us. It provides essential information about GPP, its disease course, treatment options, and daily care tips, helping us face each day with more confidence. With the right information and support, we can improve our loved one's quality of life and feel more empowered and at ease in our role.

Whether during flares, stable periods, or long-term care, this guide offers practical advice to help you provide the most appropriate care. We hope it helps you feel less helpless and more hopeful for the future.



## ✦ Understanding Generalized Pustular Psoriasis (GPP)

### What is Generalized Pustular Psoriasis (GPP)?

GPP is a rare and potentially life-threatening inflammatory skin disease, characterized by intermittent acute flares and widespread, sterile (non-infectious) yellow pustules on the body. They are not contagious but can significantly affect overall health and quality of life. GPP can occur alongside plaque psoriasis; under current international definitions, pustules must not be limited to pre-existing plaque psoriasis lesions.

### What are the key features of Generalized Pustular Psoriasis (GPP)?

Symptoms may include: joint pain, muscle weakness, widespread pustules, dry peeling/ cracked skin, burning or painful red/ discolored skin, severe itching, headache, touch-sensitive or tender skin, fever, chills, nausea, extreme fatigue, severe pain.

#### Pustule features

Yellow pustules can appear anywhere on the body, not just on the limbs, and are not contagious

#### Acute flares

Last for several days to weeks and can recur



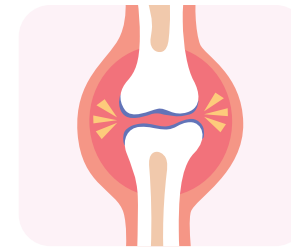
#### Impact on daily activities

During flares, systemic symptoms like fever, inflammation, extreme fatigue, and pain can significantly impair daily functioning

### Possible complications

Generalized Pustular Psoriasis (GPP) is not only a skin condition; it may also be associated with:

#### Psoriatic arthritis



May cause joint swelling and pain

#### Hypertension



Requires close monitoring of blood pressure

#### Gastrointestinal ulcers



Systemic inflammation may affect digestive tract health

#### Obesity



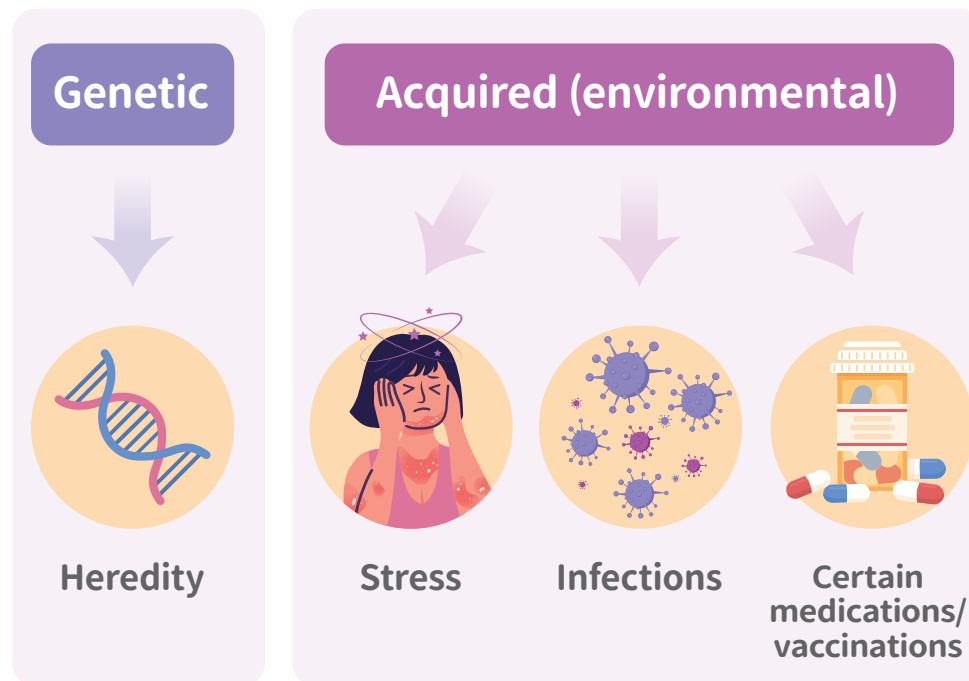
Linked to chronic inflammation and may impact overall health

**Others:** Liver disturbances, myocardial infarction, acute respiratory distress syndrome, renal dysfunction, osteoporosis, anemia, infection, neutrophilic cholangitis (jaundice)

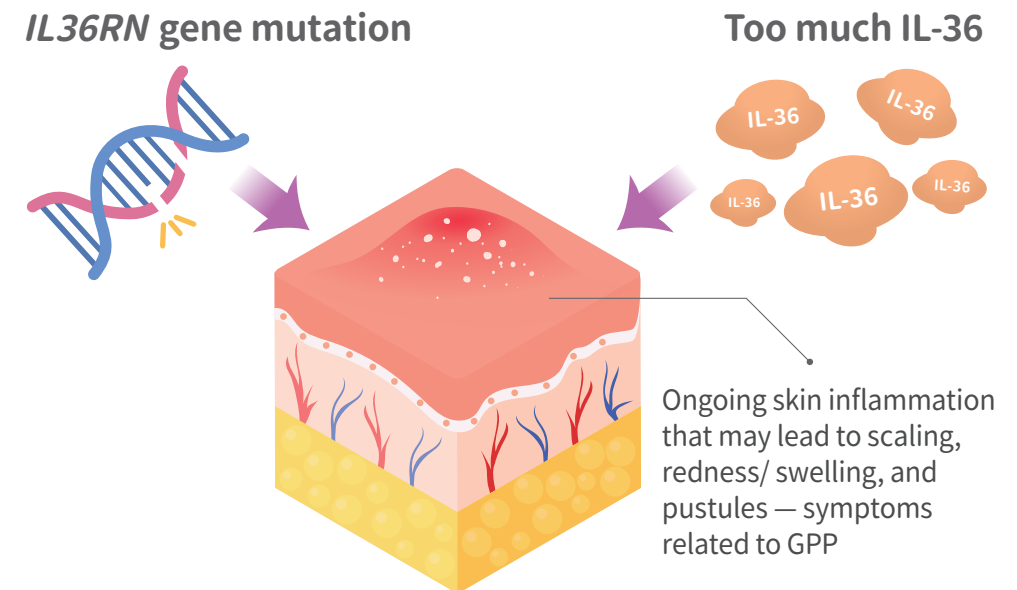
## Causes of Generalized Pustular Psoriasis (GPP)

GPP is a rare but serious autoinflammatory disease. Flares are closely related to a combination of genetic and environmental factors. For caregivers, understanding what may trigger GPP is essential, as it allows you to support the patient more effectively and lower the risk of flares.

### What causes Generalized Pustular Psoriasis (GPP)?



**Genetic factors** are mainly related to heredity. Studies show that GPP is closely linked to abnormal regulation of interleukin-36 (IL-36) in the immune system. Especially when the *IL36RN* gene is mutated, or IL-36 is overexpressed, the inflammatory response may stay “switched on,” leading to severe skin inflammation and pustules. These genetic changes impair the body's ability to control inflammation, preventing the immune system from turning off, eventually triggering acute flares. A family history of GPP or other forms of psoriasis also increases an individual's risk of developing the condition.



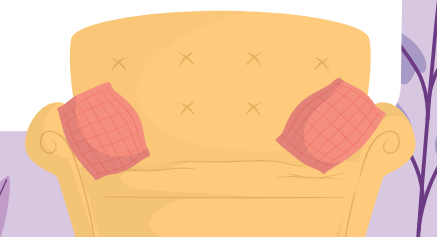
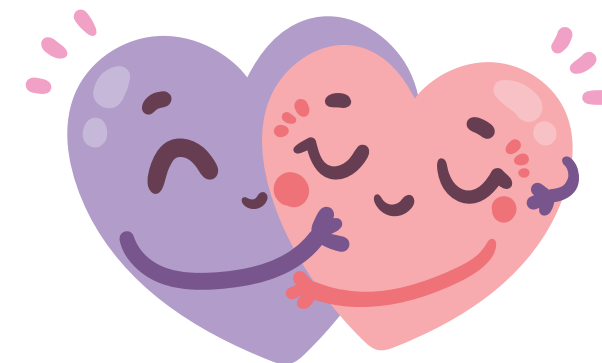
Because genetics play a role, *IL-36* gene testing, personal medical history and family history review can be a key part of the diagnostic process. In Taiwan, the process for *IL-36* gene testing generally includes the following steps:

- 1. Clinical assessment:** If GPP-like symptoms (e.g., recurrent, widespread pustular rashes) appear, the doctor makes a preliminary clinical diagnosis and decides whether *IL-36* testing is needed.
- 2. Test request:** If deemed necessary, the doctor will order an *IL36RN* test. This service is available at some medical centers and genetic testing labs. However, to receive NHI coverage for the medication Spevigo, the sample must be collected in person at a certified medical facility.
- 3. Sample collection and testing:** Usually, a blood sample is required to detect *IL36RN* variants, though some labs accept a cheek (buccal) swab.
- 4. Result Analysis:** The presence of an *IL36RN* mutation may indicate an increased GPP risk; the doctor will use the result to create a tailored treatment plan.

Through *IL-36* testing, doctors can confirm a GPP diagnosis more accurately and develop a personalized treatment plan to lower flare risk and improve the patient's quality of life.

Beyond genetics, environmental changes can also trigger GPP flares. For many patients, worsening symptoms are closely related to stress, infections, and medication changes. Other common triggers include infections (bacterial or viral), hormonal changes (e.g., menopause or pregnancy), certain medicines (e.g., antibiotics and immuno-oncology drugs), and vaccinations. Although vaccines are beneficial overall, they may trigger inflammation in a small number of GPP patients; please fully discuss with your doctor before vaccination.

As a caregiver, understanding these potential causes is crucial. During stable periods, help the patient monitor stress, infection risks, and medication changes to lower the likelihood of flares. When hormonal changes (such as pregnancy, menstruation), vaccinations, or long-term stress are anticipated, discuss a plan with the healthcare team in advance to maintain better control over the condition. With adequate knowledge and appropriate care, caregivers can help GPP patients manage the condition better, improve the patient's quality of life, and reduce its impact.



## ✦ Diagnosis and Treatment Options

### How is Generalized Pustular Psoriasis (GPP) diagnosed?

#### (1) Medication history and medication review

When diagnosing GPP, doctors usually start by asking for a detailed medication and family history. These can provide key clues:

- **Use and discontinuation of systemic corticosteroids** (as tapering or abruptly stopping them can trigger flares)
- **Recent infections** (e.g., bacterial or viral, which may disrupt the immune system)
- **Hormonal changes** (e.g., menopause, pregnancy or menstruation)
- **Excessive stress** (chronic psychological stress can affect the immune system)
- **Vaccinations** (may trigger immune reactions in some patients)

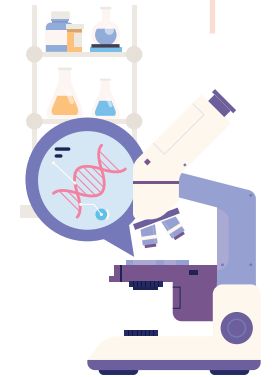
In addition, if family members have been diagnosed with GPP or other types of psoriasis, this will also be considered.



#### (2) Laboratory tests

During a GPP flare, blood tests may show changes in immune and inflammatory markers that help doctors assess severity. Possible abnormalities include:

- **Elevated neutrophils** — indicates ongoing inflammation
- **Elevated C-reactive protein (CRP)** — a common inflammation marker
- **Hypocalcemia and hypoalbuminemia** — may be linked to severe inflammatory states
- **Increased levels of liver enzymes (AST, ALT, bilirubin, gamma GT, Alk-p)** — which may indicate that systemic inflammation is impacting liver function

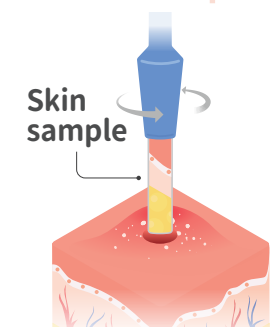


#### (3) Skin biopsy

A skin biopsy is one of the most specific diagnostic methods. Under local anesthesia, the doctor takes a small piece of skin from an affected area—especially where pustules are present—for pathological analysis. This test can help:

- **Confirm the typical pathological features of GPP.**
- **Rule out other conditions that can cause similar skin symptoms, such as infectious dermatoses, blistering diseases, or other types of psoriasis.**

This is typically a quick outpatient procedure (about 15 minutes) that provides crucial diagnostic information with minimal burden on the patient.



## What are the available treatment options of Generalized Pustular Psoriasis (GPP)?

GPP is a serious inflammatory skin disease that can affect the whole body. The main goals of GPP treatment are to ease discomfort, control inflammation, and lower the risk of relapse. Treatment options usually include oral systemic medicines, topical therapies, phototherapy, and biologics. Based on disease severity, the doctor will choose the most suitable plan.

### (1) Oral systemic therapy

The first goal of GPP therapy is to reduce the patient's discomfort. Oral medications are often used to reduce the body's abnormal inflammatory response. Common oral medicines include:

	Acitretin (Oral retinoic acid)	Methotrexate (MTX)	Cyclosporin
Potential side effects	<ul style="list-style-type: none"> <li>• Teratogenicity (Risk of birth defects)</li> <li>• Hepatotoxicity (liver damage)</li> <li>• Potential for abnormal bone development with long-term use in children</li> </ul>	<ul style="list-style-type: none"> <li>• Respiratory symptoms</li> <li>• Bone marrow suppression</li> <li>• Hepatotoxicity</li> </ul>	<ul style="list-style-type: none"> <li>• Hypertension</li> <li>• Nephrotoxicity (kidney damage)</li> </ul>
Monitoring items/precautions	<ul style="list-style-type: none"> <li>• Liver enzyme levels, blood lipids, white blood cell (WBC) count, renal function</li> <li>• Pregnancy testing</li> </ul>	<ul style="list-style-type: none"> <li>• Hepatitis B/C screening</li> <li>• WBC count</li> <li>• Liver enzyme levels</li> <li>• Use with caution in patients with renal impairment</li> </ul>	<ul style="list-style-type: none"> <li>• Blood pressure</li> <li>• Renal function</li> <li>• Liver enzyme levels</li> <li>• WBC count</li> </ul>

### (2) Topical therapies and phototherapy

In addition to systemic treatment, doctors may prescribe therapies applied directly to the skin to control skin lesions. These include:

- **Topical corticosteroids** — mainly for short-term control of inflammation, reducing redness and itch
- **Vitamin D analogs (e.g., calcipotriol)** — help regulate skin cell growth and reduce scaling
- **Phototherapy (photochemotherapy, narrowband UVB)** — uses ultraviolet light to modulate immune responses and improve skin condition, suitable for mild to moderate chronic cases

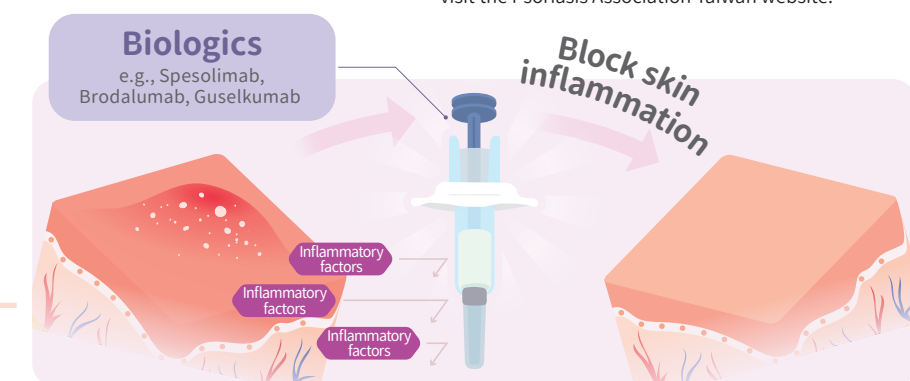
These are typically used as supplementary treatments alongside systemic medications for better overall control.

### (3) A new frontier in treatment — biologics

Biologics are advanced protein-based medications, created through genetic engineering, that precisely target and block specific factors in the immune system that cause inflammation. Their advantages include faster onset and greater treatment precision, and they may have fewer side effects than traditional immunosuppressants.

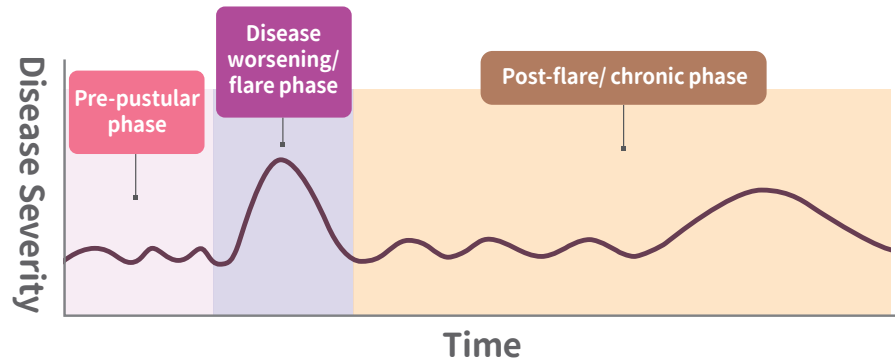
Several biologics are used to treat GPP. In Taiwan, NHI-covered options include: IL-36 receptor inhibitor (Spesolimab), IL-17 receptor inhibitor (Brodalumab), and IL-23 inhibitor (Guselkumab). These newer biologics have shown good effectiveness. Many clinical studies indicate they can significantly improve GPP symptoms in a short time, providing meaningful new treatment options.

※ For the latest information on GPP medications, please visit the Psoriasis Association Taiwan website.



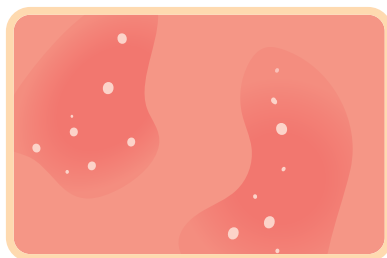
## ✨ The Caregiver's Key Roles and Tasks Across the Course of Generalized Pustular Psoriasis (GPP)

GPP is a highly variable and unpredictable chronic inflammatory skin disease, and caregivers play an important role at every stage. Through careful observation, support, and care, you can help reduce your loved one's discomfort, improve their quality of life, and lower the risk of relapse.



### 1. Pre-pustular phase

When a patient shows symptoms suggestive of GPP—such as recurrent, widespread pustular rashes—the doctor will make a preliminary diagnosis based on clinical signs and decides whether *IL-36* gene testing is needed.



During non-flare periods, numbers of pustules and areas of redness are generally smaller.

### Caregiver's roles and key tasks

- Monitor skin changes, including redness, dryness, and scaling, and record their progression.
- Understand the course of GPP and possible triggers so you can provide timely support and companionship.
- Encourage regular clinic visits so the healthcare team can detect early signs of worsening.



### Everyday care tips



#### Adults

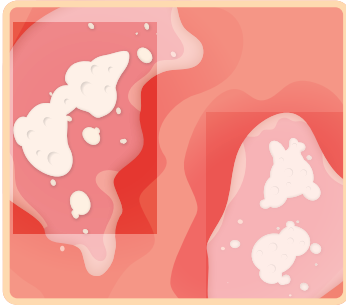
- Use gentle, non-irritating moisturizers to prevent dryness.
- Avoid scratching or irritating the skin; choose breathable, soft clothing.
- Manage stress and practice relaxation, such as meditation or deep breathing.



#### Children

- Use mild bath products and warm water and gently pat the skin dry.
- Avoid dressing the child too warmly; keep the skin well-ventilated.
- Help children understand their condition to ease anxiety and feelings of low self-esteem related to their skin.

## 2. Disease worsening/ flare phase



This is the acute phase of GPP, marked by widespread pustule eruptions, red and swollen skin, fever, pain, fatigue, and even systemic inflammation (e.g., fever, organ dysfunction). A flare can progress rapidly and may become life-threatening, requiring urgent medical attention.

### Caregiver's roles and key tasks

- Respond quickly by seeking emergency medical care; hospitalization may be necessary.
- Assist in managing pain and inflammation by ensuring medications are taken as prescribed.
- Provide emotional and psychological support, as severe skin symptoms can cause significant anxiety and distress.

### Treatment and management options

	Adults	Children
First-line	<ul style="list-style-type: none"> <li>• Conventional therapy: oral retinoic acid</li> <li>• Biologics: IL-36 receptor inhibitor (IV Spesolimab)</li> </ul>	<ul style="list-style-type: none"> <li>• Conventional therapy: oral retinoic acid</li> </ul>
Second-line	<ul style="list-style-type: none"> <li>• Conventional therapy: cyclosporine, methotrexate (MTX), oral corticosteroids</li> <li>• Biologics: TNFi, IL-17i, IL-23i, IL-12/23i</li> </ul>	<ul style="list-style-type: none"> <li>• Conventional therapy: cyclosporine</li> <li>• Biologics: TNFi</li> </ul>

### Everyday care tips



#### Adults

- Follow the doctor's prescription for anti-inflammatory and immunomodulatory drugs. If oral or injectable corticosteroids are needed, be aware of infections and long-term use, and taper carefully to prevent relapse (e.g., with oral steroids or biologics).
- Keep the skin clean to prevent secondary infections and avoid scratching.
- Monitor for systemic symptoms like body temperature, chills, or weakness.



#### Children

- Ensure children drink enough fluids and proper nutrition to prevent dehydration.
- Reduce activity and allow plenty of rest for children.
- Cover affected areas with soft gauze to minimize friction and pain.

### 3. Post-flare/ chronic phase

After a flare, symptoms may ease, but many patients continue to experience skin damage (dryness, scaling, redness) and systemic fatigue. Studies show 83–86% remain in a chronic phase with a risk of future flares.

#### Caregiver's roles and key tasks

- Support patients in following their treatment plan and help avoid known triggers. Encourage regular follow-up appointments to monitor changes and adjust treatment.
- Provide skin care and anti-itch strategies for long-term symptom control and comfort.
- Encourage participation in support groups or counseling to improve quality of life.

#### Treatment and management options

	Adults	Children
First-line	<ul style="list-style-type: none"> <li>• Conventional therapy: oral retinoic acid</li> <li>• Biologics: IL-36 receptor inhibitor (IV Spesolimab)</li> </ul>	<ul style="list-style-type: none"> <li>• Conventional therapy: oral retinoic acid</li> </ul>
Second-line	<ul style="list-style-type: none"> <li>• Conventional therapy: cyclosporine, methotrexate (MTX), oral corticosteroids</li> <li>• Biologics: TNFi, IL-17i, IL-23i, IL-12/23i</li> </ul>	<ul style="list-style-type: none"> <li>• Conventional therapy: cyclosporine</li> <li>• Biologics: IL-36 receptor inhibitor (subcutaneous Spesolimab, for patients at least 12 years old and weighing <math>\geq 40</math> kg), TNFi</li> </ul>

#### Everyday care tips



##### Adults

- Keep using moisturizers and skincare products to prevent dryness.
- Monitor skin changes for early signs of a potential relapse. Consider taking regular photos to record any abnormal changes and share with doctors to assess disease progress and treatment response.
- Exercise regularly and eat a healthy diet to improve overall health.



##### Children

- Change bed linens and clothing regularly to maintain hygiene and reduce infection risk.
- Provide a gentle play environment to avoid injuries and skin irritation.
- Help children build a positive self-image and reduce anxiety or distress related to their skin.

- Navarini, A. A., et al. (2017). *J Eur Acad Dermatol Venereol*, 31(11), 1792-1799.
- Choon, S. E., et al. (2015). *Int J Dermatol*, 54(7), 737-752.
- Lyk, L., et al. (2019). *Psoriasis (Auckl)*, 9, 27-41.
- Lebwohl, M. G., et al. (2022). *J Psor Psor Arth*, 7(1), 71-78.

## ✦ Stress Management for Caregivers of Patients with Generalized Pustular Psoriasis (GPP)

Caring for GPP patient requires constant adaptation and can be challenging both physically and emotionally. Managing your own stress and staying healthy are just as important. Below are effective strategies:

### Focus on yourself — emotional self-care

When a patient shows symptoms suggestive of GPP—such as recurrent, widespread pustular rashes—the doctor will make a preliminary diagnosis based on clinical signs and decide whether *IL-36* gene testing is needed.



Keep a regular daily routine



Do moderate exercise and relaxation



Maintain good nutrition and hydration



Take breaks



Find positive emotions through hobbies



Have regular health checkups

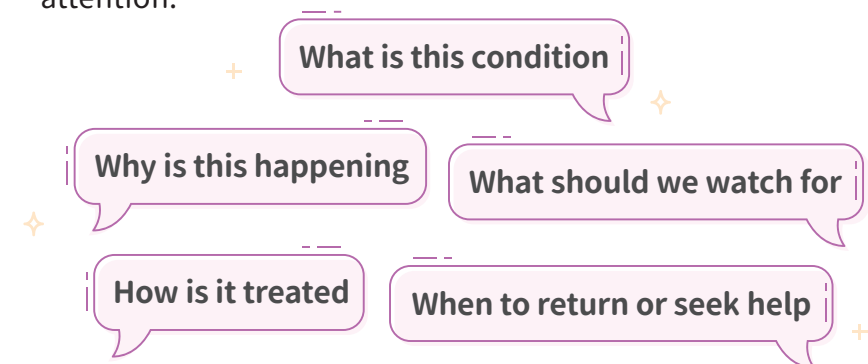
### Key reminder

Effectively managing your stress not only protects your own health but also improves the quality of care you can provide. Whatever self-care you choose, make sure you have people who support you.

### Improve communication— skills for effective clinical visits

When accompanying a patient with GPP to appointments, clear and effective communication is vital to ensure your loved one receives the best care. Here are practical tips to build good collaboration with the healthcare team:

- **What is this condition:** Describe changes in symptoms and bring records to help the doctor understand the course and make an accurate diagnosis.
- **Why is this happening:** Ask about possible triggers—such as stress, infections, or medications—to adjust care for patients.
- **How is it treated:** Understand different treatment options, side effects, and coping strategies to secure the most suitable plan.
- **What should we watch for:** Clarify instructions for daily care, including diet, skincare, and infection prevention.
- **When to return or seek help:** Confirm follow-up appointment and know the warning signs that require urgent medical attention.



### Key reminder

Good communication keeps information aligned between patients and clinicians, improves the care experience, and makes caregiving smoother.

## Build your support system — creating and maintaining a support network

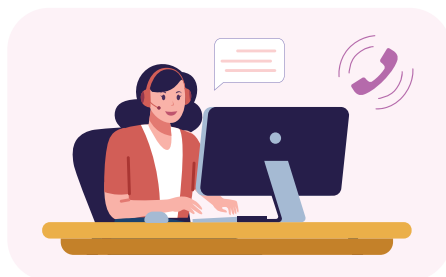
You are not alone in this journey. A strong support network can ease stress and make caregiving smoother. Here are ways to build effective support:



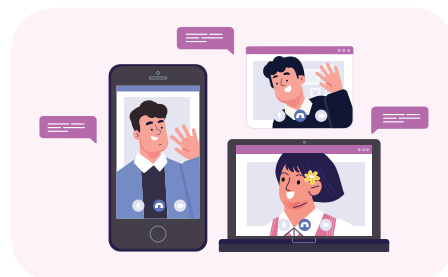
Join support groups



Encourage the patient to engage with peer communities



Use community and social resources



Stay connected with family and friends

## Caregiver stress management and counseling resources

In Taiwan, several services are available to help caregivers manage the physical and emotional burden of care:

- **Family Caregiver Support Hotline (0800-507-272):** This Ministry of Health and Welfare hotline, with the slogan "Having You Makes a Difference," offers social welfare counseling, referral services, and psychological support. It is available Monday–Friday, 9:00 AM to 5:00 PM.
- **Mental health counseling:** If you are feeling overwhelmed, you can request individual counseling via the Long-Term Care hotline 1966 or the Family Caregiver Support Hotline 0800-507-272 to process emotions and receive professional support.
- **Health education:** The Taiwanese Dermatological Association and the Psoriasis Association Taiwan provide reliable professional resources on Generalized Pustular Psoriasis (GPP), including causes, symptoms, diagnosis, and treatments, to raise public awareness of this rare disease.

“

**Building a strong support network not only reduces caregiver stress but also improves the quality of life for GPP patients, making care warmer and more resilient.**

”



## ✦ Frequently Asked Questions (FAQ)

### Q1: How is Generalized Pustular Psoriasis (GPP) different from common psoriasis?

Both GPP and psoriasis (especially moderate to severe) are considered systemic inflammatory diseases, but GPP is typically more acute and progresses rapidly; fever and fatigue are common symptoms. Skin lesions in common psoriasis are relatively stable and rarely causes systemic symptoms unless it is very severe (e.g., erythroderma).

### Q2: Do Generalized Pustular Psoriasis (GPP) patients need hospitalization?

During acute flares, hospitalization may be necessary for systemic treatment and close monitoring.

### Q3: Are biologics covered by National Health Insurance (NHI) for Generalized Pustular Psoriasis (GPP)?

The NHI may cover biologics for GPP patients who meet specific criteria. This requires a physician's evaluation and application.

### Q4: Are there any dietary recommendations for Generalized Pustular Psoriasis (GPP) patients?

Choosing anti-inflammatory foods (e.g., omega-3 fatty acids) may be beneficial. It is often recommended to limit high-sugar and high-fat foods.

### Q5: Can Generalized Pustular Psoriasis (GPP) patients get vaccinated?

Yes, but it is essential to consult your doctor first to ensure it does not interfere with any ongoing immune-based treatments.

### Q6: Is Generalized Pustular Psoriasis (GPP) contagious?

No. GPP is an immune system disorder and is not contagious.

### Q7: How can I help a patient cope with the emotional stress of GPP?

Offer emotional support, help find peer support groups, and seek counseling when needed.

### Q8: Can Generalized Pustular Psoriasis (GPP) patients exercise?

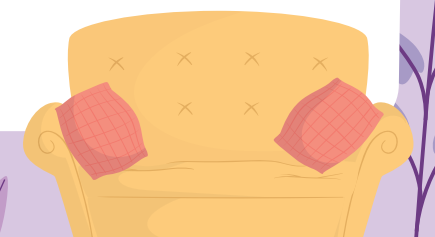
Yes, choose low-intensity activities like yoga or walking, and avoid exercises that irritate the skin.

### Q9: How long does the treatment of Generalized Pustular Psoriasis (GPP) last?

This is highly individual. Patients who experience recurrent flares or have persistent symptoms between flares often require long-term treatment to prevent relapse.

### Q10: How can relapse risk of Generalized Pustular Psoriasis (GPP) be reduced?

Discussing a suitable long-term treatment plan with the doctor, managing stress effectively, reducing respiratory tract infection and maintaining a healthy lifestyle are key strategies.



## ✦ Generalized Pustular Psoriasis (GPP) Symptom Tracker

As a caregiver, please use this sheet to note your loved one's symptoms but over time you'll become more familiar with the patient's patterns, helping

and any potential triggers you observe. Not every trigger will cause a flare, you spot early warning signs and provide more effective care.

		First week	Second week
Skin symptoms	Blisters or pustules		
	Red or discolored skin		
	Skin pain (burning or stinging)		
	Severe itching		
Systemic symptoms	Fever or chills		
	Headache		
	Joint pain		
	Muscle weakness		
	Extreme fatigue		
	Other symptoms		

Third week	Fourth week	Notes (include possible triggers and how long the symptoms last)

# Generalized Pustular Psoriasis Caregiver Booklet

We gratefully acknowledge

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This material provides medical and health education information about the related condition only. For questions about your personal health or the treatment, please consult healthcare professionals for individualized advice.

