Nursing Care Resource for Generalized Pustular Psoriasis (GPP)

This resource aims to:

- assist healthcare professionals
- inform patients with GPP and their families about the condition and treatment
- foster health literacy by providing clarity on pathways to care

What is GPP?

It is a severe and rare variant of psoriatic disease characterized by a widespread eruption of painful sterile pustules, sometimes accompanied by systemic symptoms such as fever.

GPP is NOT an infection and is NOT contagious

Because of its rarity, GPP remains unknown to many healthcare professionals globally.

How does GPP present?

It tends to appear suddenly with erythema (redness) of the skin together with the appearance of pustules. The sensitive skin brings pain, itching and burning sensations. Apart from the pustules, there can be mucosal changes, such as on the tongue, lips and eyes.

The pustules can join together then burst, breaking open the skin barrier, leaving the skin raw. New pustules can develop creating a harmful cycle with possible sepsis.

GPP, like other forms of psoriatic disease, is characterized by unpredictable flares. The uncertain course of the disease can result in remission, relapses or can be persistent. Possible complications include acute respiratory distress syndrome, renal failure and congestive heart failure, hence the seriousness of this disease.

(Fujita, Gooderham & Romiti, 2021)

The main types of pustular psoriasis are:

- Generalized Pustular Psoriasis, which refers to pustular psoriasis across the body
- Localized Pustular Psoriasis such as Palmoplantar Pustulosis, which erupts on the palms of the hands and soles of the feet

From the patient's viewpoint: Self-examination

Falling ill felt like my body decided to hit the brakes, forcing me to slow down when all I wanted was to keep moving. Every ache and shiver reminded me that sometimes, rest isn't just needed—it's demanded. (GPP Patient, 2024)

Physical discomfort and burden include:

- Painful sores with blisters on large areas of skin
- Sudden unexpected flare
- Fever, shivers
- Itching and burning sensation

Emotional burden

GPP usually has a severe impact on people's lives though anxiety, difficulty sleeping, worries about employment, tension around relationships, concerns about future flares

The unpredictability of the disease adds to the burden of anxiety

Depression has been identified as a key factor in psoriatic disease. Unlike previously when depression was considered a consequence of the chronic disease, recent research has shifted this thinking. Now, the inflammatory aspect of depression has been identified to exhibit a pattern of inflammation described as being very similar to that in psoriatic disease. Furthermore, obesity adds to the risk of depression due to the additional inflammation produced by the fatty tissue.

(Mrowietz, Sümbül, & Gerdes, 2023).

Diagnosis

Diagnosis can be difficult. It requires a "careful assessment of the patient's skin symptoms, potential disease triggers, medical history, histopathologic features, laboratory tests, and clinical disease course" (Fujita, Gooderham & Romiti, 2021:531).

About 3% of people living with psoriatic disease develop pustular psoriasis. It is a heterogeneous disease which affects more women than men, is more common in adults and those with other chronic diseases (known as comorbidities).

(Frysz et al., 2024)

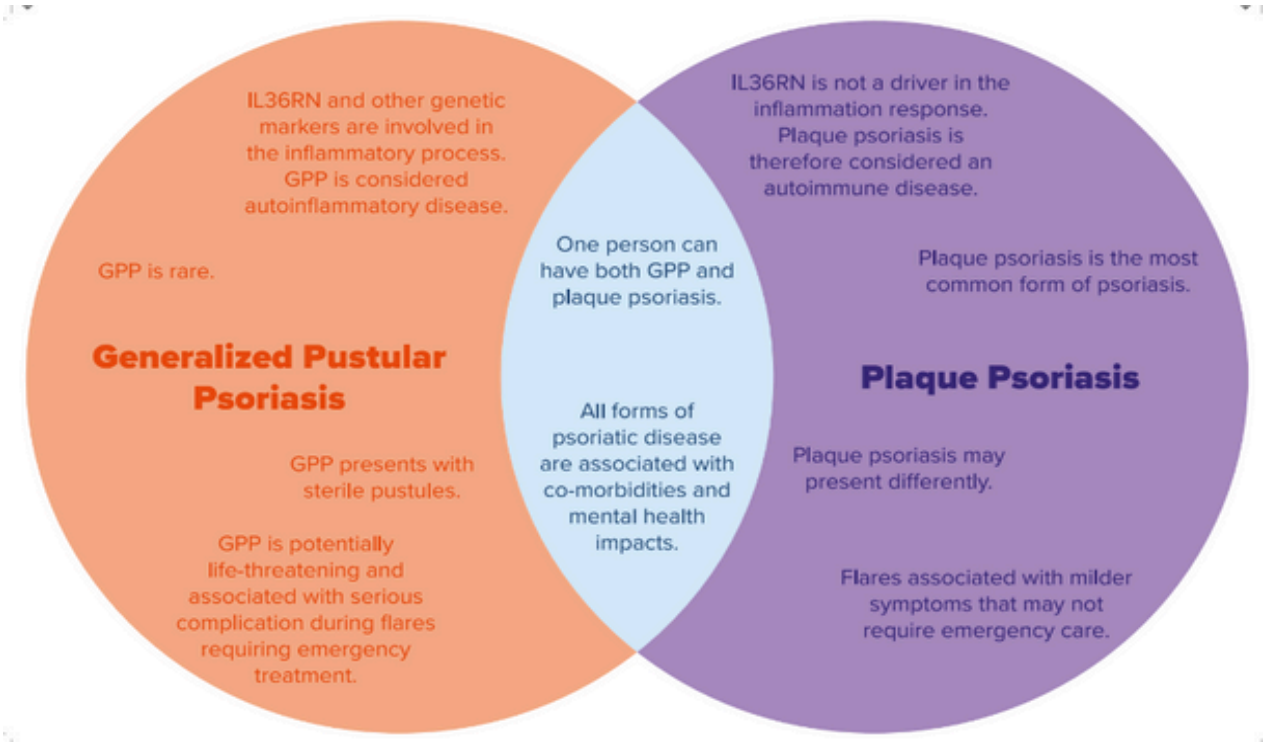


Image from IFPA website here

Triggers that have been identified include:

- Sudden corticosteroid withdrawal
- Infections e.g. streptococcal
- Certain vaccines e.g. for severe acute respiratory syndrome
- Certain medical conditions e.g. hypocalcemia
- Pregnancy, especially in the early stage of the 3rd trimester. Close monitoring is needed as there can be serious consequences for both the mother and foetus.
- Emotional stress

(Fujita, Gooderham & Romiti, 2021)

There are numerous measurements to assess the severity of GPP such as

- Skin signs
- Systemic inflammation score
- Blood findings eg serum C-reactive protein and albumin levels

(Fujita, Gooderham & Romiti, 2021)

GPP needs to be differentiated from:

- Acute generalized exanthematous pustulosis - caused by the body's reaction to an antibiotic
- Impetigo with septicaemia
- Uncommon autoimmune diseases such as immunoglobulin A (IgA) pemphigus

Standardized guidelines for diagnosis and management of GPP are still in the early stages of development. There is an urgent need for research on care and treatment development for diseases like GPP which are classified as Orphan diseases which means that they have not been taken up by the pharmaceutical industry due to the belief that there is limited financial gain in developing and marketing new medications for treatment or prevention of these rare diseases. There have also been few clinical trials because of the rarity of GPP.

Medical Treatment

An informal survey conducted by the South African Psoriasis Association (SAPSA) in 2024 found that among 127 Dermatologists in Southern Africa a wide range of medication options were prescribed for GPP patients that included:

· Topical steroids such as clobetasol (Dovate)

· Systemic medications, such as acitretin, methotrexate, prednisone, cyclosporin

· Vit D analogues

· Biologics, when available and needed, such as infliximab, adalimumab & secukinumab

Nursing during the acute severe stage

An analgesic like Tramadol can help to reduce the pain and enable drowsiness. Caution needs to be taken due to its addictive quality and side effects such as constipation.

A sedating antihistamine may be useful to help the patient sleep.

Applications used in the treatment of GPP include

- Topical steroids - twice daily, weaning to daily
- An antibacterial body wash - preferably in a bath
- Twice daily wet/dry wraps
- Oral agents such as methotrexate, prednisone, acitretin, cyclosporine and biologics (oral or injectable), like secukinumab, which needs special attention for the injection i.e. a twirling motion. A GPP specific biologic treatment like spesolimab can be applied subcutaneously or intravenously.

Currently there are new GPP treatment options being developed that will focus on the IL-36 pathway. In time, biosimilars will become available to offer a more affordable alternative to the originator biologic drugs.

Practical application tips

Give attention to possible comorbidities throughout the treatment programme.

Listen and be attentive to the patient's personal story. Dissect what you hear to determine the hidden aspects of their healthcare.

Be gentle as the condition can be distressing. Any touching of the skin is extremely painful.

Apply ointments by dabbing onto the skin rather than rubbing, to avoid any skin damage.

Advise a warm bath. It can be comforting and relaxing for the patient. Use povidone-iodine as an antiseptic and a mild soap. Dry the body very carefully through a patting motion rather than rubbing.

Be alert to a spiking temperature, or sudden drop in blood pressure and tachycardia due to the inflamed blood vessels. Septic shock can occur.

Treatment during the stage of moderate severity

Wean topical treatment to clobetasol daily

On the face use 1% hydrocortisone (Mylocort)

Develop a caring relationship with the patient to consider:

- Is the patient able to access a health facility once home?

- Will that facility be able to manage the needs of someone with a rare disease?
- Will the prescribed treatment be available to the patient, once home?
- What is the social economic status of the patient and the implications for home care?

Treatment for on-going care

Continue weaning off ultrapotent topical steroids.

Wean to fluocinolone (Cortoderm) daily when the skin has settled

Clobetasol ought to be at hand in case of a repeat flare

For the scalp

- use an anti-inflammatory shampoo such as tar shampoo or Selsun shampoo
- give Synalar gel or Topivate cream daily

If the skin is dry, use a moisturizer like an emulsifying ointment. Pot hygiene is important.

Lotion hygiene
A few tips for applying lotions

Caution – don't contaminate your product through poor hygiene

1. Always first wash your hands
2. Scoop the ointment with the back of a spoon or spatula – never put your fingers directly into the pot
3. Take the lotion from spoon/spatula to a dish
4. Apply to affected body areas from the dish
5. Discard leftover lotion – do not put leftovers back in the pot

In preparation for the next treatment, wash the spoon/spatula and tightly close the jar.

Heating and air conditioning can contribute to drying out the skin.

Discharge alerts

Quotes from patients with GPP

"I'm so scared to use the medication…I'm supposed to take meds on Thursdays but don't take it all the time"

"I don't want to go out of the house as everything is red"

"It is like, you are in the dark"

Recommend referrals regarding comorbidities such as a dietician if the patient is diabetic.

Useful question to ask the patient:

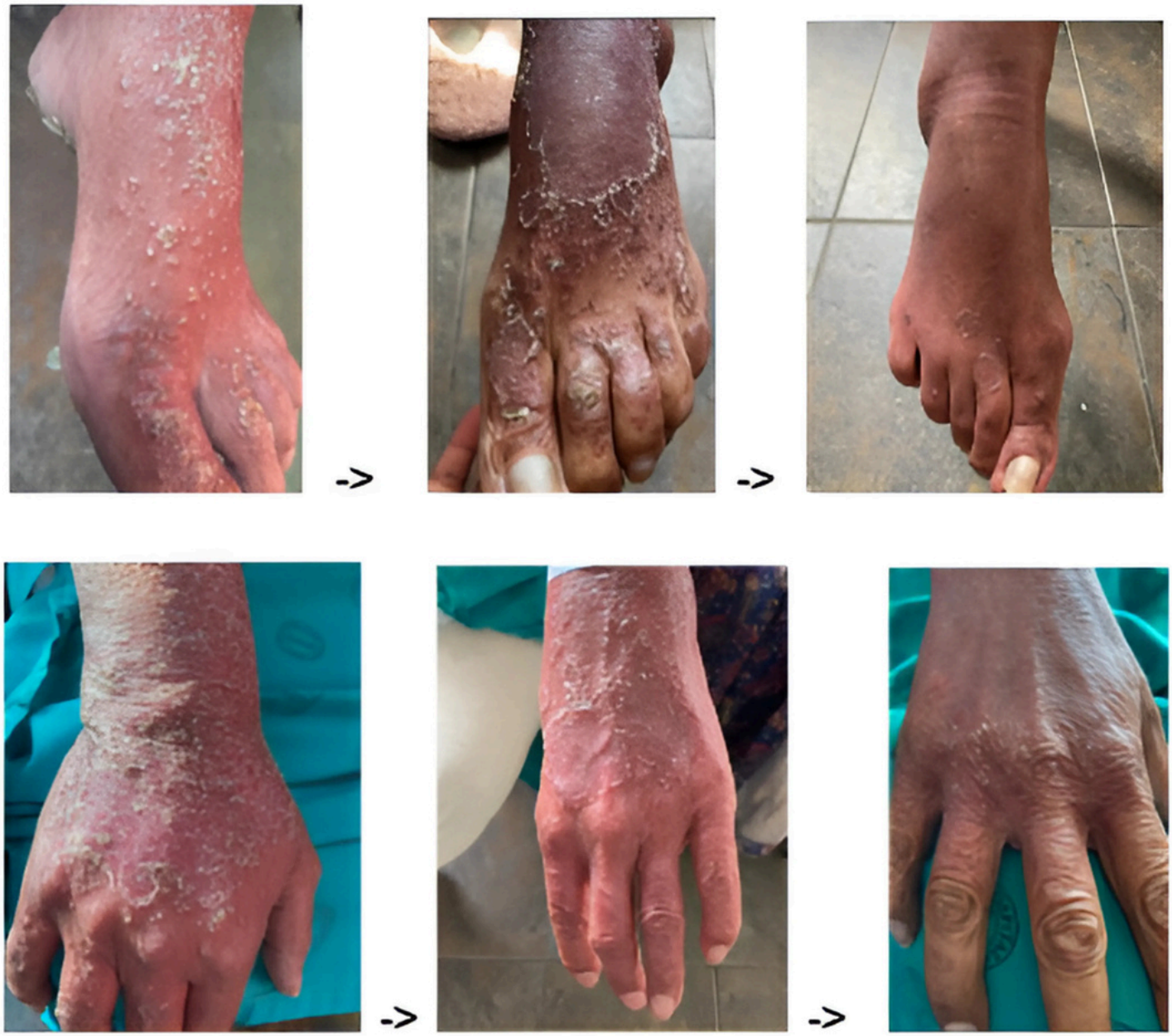
- Do you understand the importance of the regime to wean off steroids?
- Is your local healthcare facility able to cater for your special needs?
- Will you be able to get your medications on time?
- Will your socio-economic status enable you to get your medications?
- Do you understand how to deal with small flare-ups to avoid them spreading to become serious?

Advice to patients

- Keep using the topicals as prescribed

- Don’t suddenly stop your treatment if you are feeling better
- If on methotrexate, remember to check on liver function regularly
- Keep follow-up appointments
- Avoid excessive sun exposure by wearing protective clothing when outside and sunscreen protection
- Do not scratch your skin

Example of patient progress



Initial Presentation → 4 days later → On discharge

Thanks to:

- Groote Schuur Hospital Dermatology Nursing Team, Cape Town, South Africa
- Dr Sue Jessop MBChB UCT, FF Derm SA

References

Frysz, M., Patel, S., Oy Yee Li, M.,Griffiths, C.E. M., Warren, R. B., & Ashcroft, D. M. (2024). Prevalence, incidence, mortality and healthcare resource use for generalized pustular psoriasis, palmoplantar pustulosis and plaque psoriasis in England: a population-based cohort study, *British Journal of Dermatology*, 191(4),529–538. https://doi.org/10.1093/bjd/ljae217

Fujita, H., Gooderham, M., & Romiti, R. (2022). Diagnosis of generalized pustular psoriasis. *American Journal of Clinical Dermatology*, *23*(Suppl 1), 31-38. https://link.springer.com/article/10.1007/s40257-021-00652-1

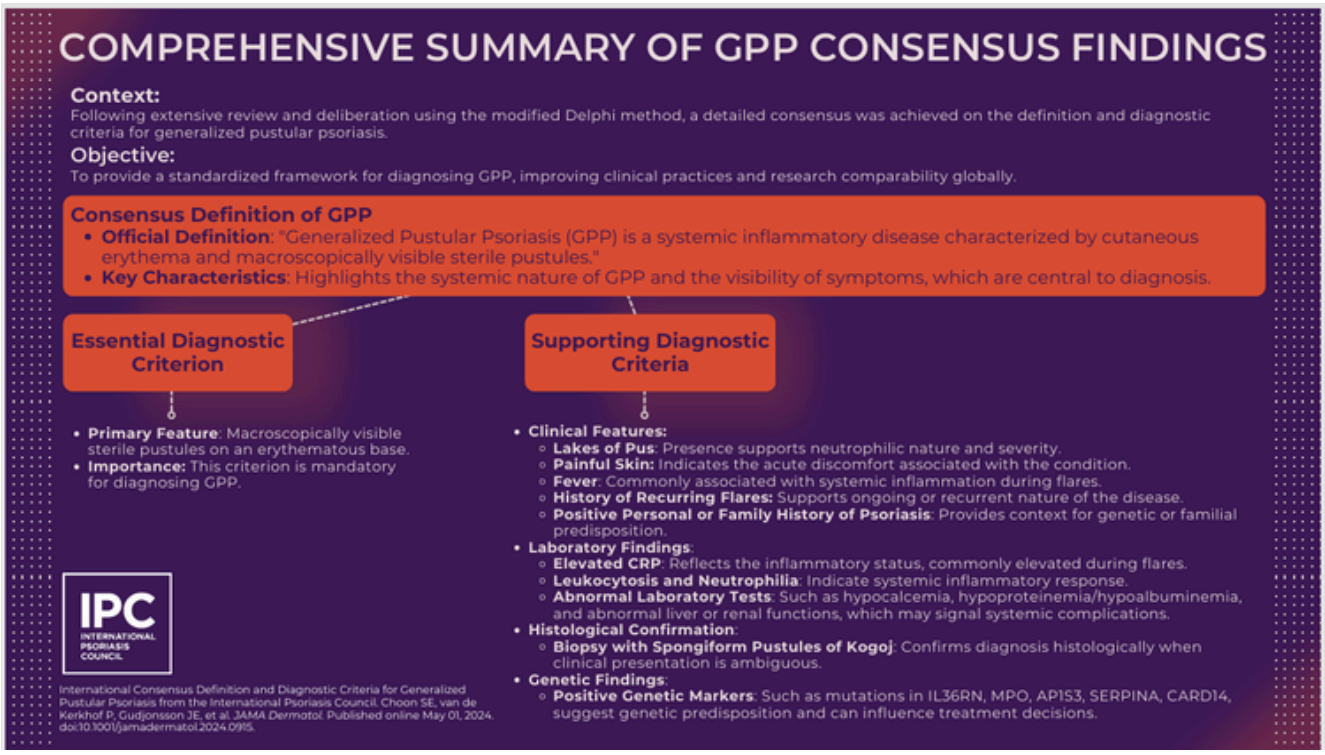
Genovese, G., Moltrasio, C., Cassano, N., Maronese, C.A., Vena, G.A., & Marzano, A.V. (2021), Pustular Psoriasis: From Pathophysiology to Treatment. *Biomedicines* 9, 1746. https://www.mdpi.com/1371080

Mrowietz, U., Sümbül, M., & Gerdes, S. (2023). Depression, a major comorbidity of psoriatic disease, is caused by metabolic inflammation. *Journal of the European Academy of Dermatology and Venereology*, *37*(9), 1731-1738.

Links further resources

US National Psoriasis Foundation: https://www.psoriasis.org/advance/understanding-pustular-psoriasis/

UK Psoriasis Association: https://www.psoriasis-association.org.uk/pustular-psoriasis



https://ifpa-pso.com/search?search=generalized%20pustular%20psoriasis