

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



Clinical Discussion Guide: Explaining Treatment Options to Patients

Purpose of This Guide

This is a resource to facilitate point-of-care discussions with patients and families about treatment options for 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

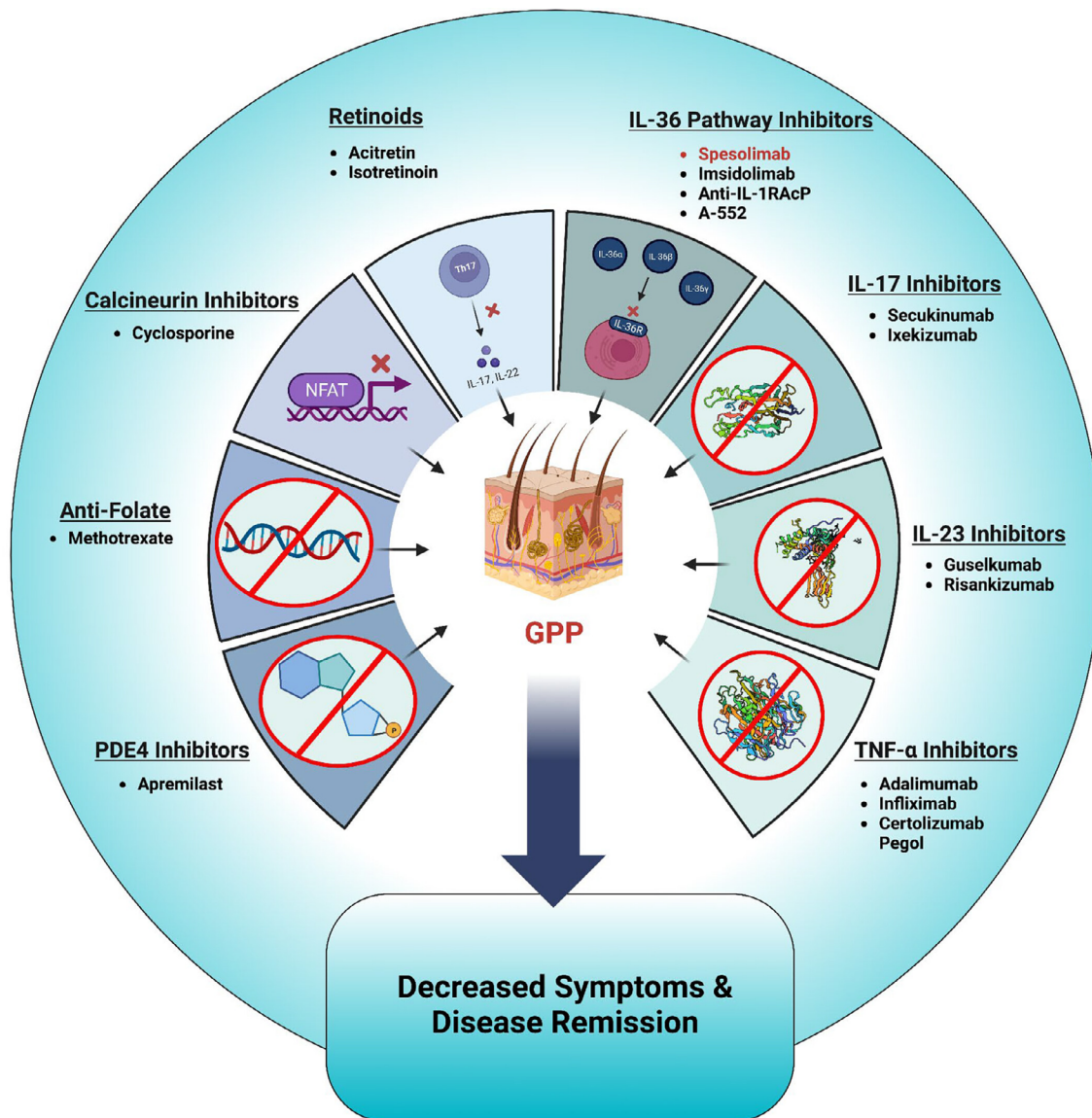
- It is critical that GPP is treated promptly to improve the patient's skin, manage symptoms, and prevent potentially serious complications¹
- Health care teams work with patients to assess their individual situation and determine an appropriate treatment plan based on the severity of symptoms, medical history, goals of therapy, and patient preferences¹
- There are a range of treatments that may be used for the treatment of GPP, including topical treatments, phototherapy, oral drugs, and biologics.² In some cases, topical emollients or mild topical steroids may be appropriate as part of a treatment plan.³ The first line of treatment for GPP typically includes oral agents or biological therapies such as retinoids, methotrexate, cyclosporine, or infliximab⁴
- Spesolimab is currently the only FDA-approved targeted therapy specifically for the treatment of GPP flares in adults⁵
- When patients experience a GPP flare, a health care provider may administer spesolimab as a 90-minute intravenous infusion. If patients still have flare symptoms a week later, the provider may administer a second 90-minute infusion
- Before receiving spesolimab, health care providers should be aware of all current medical conditions, particularly if a patient has tuberculosis or was recently in close contact with someone who has tuberculosis. Patients should also let their care team know if they have recently received any vaccines or have any scheduled. If a patient opts to receive spesolimab, they should not receive any live vaccines.⁶ Live vaccines are used to protect against diseases such as measles, mumps, and rubella (MMR combined vaccine), rotavirus, smallpox, chickenpox, and yellow fever⁷

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Current Treatment Options and Common Questions



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QUESTION	RESPONSE
How is apremilast administered?	Apremilast is a pill you take by mouth. The dosing schedule is as follows: Day 1: one 10 mg tablet in the morning Day 2: one 10 mg tablet in the morning and again in the evening Day 3: one 10 mg tablet in the morning and a 20 mg tablet in the evening Day 4: one 20 mg tablet in the morning and a 20 mg tablet in the evening Day 5: one 20 mg tablet in the morning and a 30 mg tablet in the evening Day 6 and thereafter: one 30 mg tablet in the morning and a second one in the evening
What are the side effects of apremilast ?	As with all medications, apremilast has potential side effects. Some patients taking apremilast have experienced weight loss, nausea, vomiting, and diarrhea. There have also been reports of increased depression in patients taking apremilast. Apremilast may cause birth defects and, therefore, the risks and benefits should be discussed before use if you are pregnant or are planning to become pregnant.
How is methotrexate administered?	Methotrexate is an oral medication taken once a week. Your dose will be determined by your age and other health concerns. You will need to have bloodwork done while taking methotrexate. This is usually done weekly at first, and then advanced to every other month. Your provider will be tracking your blood count, kidney, and liver function.
What are the side effects of methotrexate ?	Methotrexate can lower folate stores, which are needed for red blood cell formation. As a result, you may feel more tired, experience hair loss, nausea, and vomiting. Therefore, it is important to take a folic acid 1-mg supplement every day. Methotrexate should not be used by anyone who is pregnant, has liver or kidney disease, is an alcoholic, has anemia, or has other blood disorders.

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How is cyclosporine administered?	Cyclosporin is an oral medication taken daily. The dose will be determined by weight and other health conditions. Cyclosporin requires blood work monitoring before you start, and then every 2 weeks for the first 3 months. There will be ongoing monitoring of blood count, electrolytes, cholesterol, kidney, and liver function. Your blood pressure will also be checked every 2 weeks for the first 3 months.
What are the side effects of cyclosporine ?	Cyclosporine should not be used by patients with infections or poorly controlled diabetes, uncontrolled hypertension, cancer, or abnormal kidney function. Some patients complain of nausea, diarrhea, headaches, restlessness, swollen/inflamed gums, and numbness/tingling in their hands and feet. Cyclosporin should not be used by anyone who is pregnant.
How is acitretin administered?	Acitretin is a daily oral medication. The usual dose is between 25 mg every other day and up to 50 mg daily. Acitretin does require blood work monitoring before you start therapy and while you are on therapy. There will be ongoing monitoring of triglycerides and liver function.
What are the side effects of acitretin ?	Common side effects of acitretin are dry skin and lips, sun sensitivity, and even hair loss. Acitretin should not be used by anyone who is pregnant, and pregnancy should be avoided for 3 years after discontinuing the medication.
How is secukinumab administered?	Secukinumab is a self-injection, which can be administered at home. The injection schedule is as follows: 300 mg weekly for the first 5 weeks, then 300 mg once every 4 weeks. Secukinumab does require blood work monitoring before you begin treatment and every year thereafter. Commonly, patients are checked for HIV, hepatitis, TB, blood count, kidney/liver function, and cholesterol.

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QUESTION	RESPONSE
What are the side effects of secukinumab ?	Secukinumab should not be used by patients who are pregnant. You should not receive a live vaccine (MMR, chickenpox, yellow fever, rotavirus, smallpox) while on secukinumab. Secukinumab should not be used in those with a history of Crohn's disease or ulcerative colitis. Common side effects include injection site reaction, upper respiratory tract infection, sore throat, and diarrhea.
How is ixekizumab administered?	Ixekizumab is a self-injection administered at home. The injection schedule is as follows: Week 0: 160 mg Week 2-12: 80 mg every 2 weeks Weeks 12+: 80 mg every 4 weeks Ixekizumab does require blood work monitoring before you begin treatment and every year thereafter. Commonly, patients are checked for HIV, hepatitis, TB, blood count, kidney/liver function, and cholesterol.
What are the side effects of ixekizumab ?	Ixekizumab should not be used in patients who are pregnant. You should not receive a live vaccine (MMR, chickenpox, yellow fever, rotavirus, smallpox) while on ixekizumab. Common side effects include injection site reaction, upper respiratory tract infection, nausea, and fungal infections.
How is guselkumab administered?	Guselkumab is a self-injection administered at home. The injection schedule is as follows: Week 1: 100 mg Week 4: 100 mg Every 8 weeks: 100 mg Guselkumab requires blood work monitoring before you begin treatment and every year thereafter. Commonly, patients are checked for HIV, hepatitis, TB, blood count, kidney/liver function, and cholesterol.

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What are the side effects of guselkumab ?	Guselkumab should not be used in patients who are pregnant. You should not receive a live vaccine (MMR, chickenpox, yellow fever, rotavirus, smallpox) while on guselkumab. Common side effects include injection site reaction, upper respiratory tract infections, joint pain, headaches, and fungal and viral skin infections.
How is risankizumab administered?	Risankizumab is a self-injection administered at home. The injection schedule is as follows: Week 0: 150 mg Week 4: 150 mg Every 12 weeks: 150 mg Risankizumab does require blood work monitoring before you begin treatment, and every year thereafter. Commonly, patients are checked for HIV, hepatitis, TB, blood count, kidney/liver function, and cholesterol.
What are the side effects of risankizumab ?	Risankizumab should not be used by patients who are pregnant. You should not receive a live vaccine (MMR, chickenpox, yellow fever, rotavirus, smallpox) while on risankizumab. Common side effects include injection site reaction, upper respiratory tract infections, headache, fatigue, and fungal infections.
How is adalimumab administered?	Adalimumab is a self-injection administered at home. The injection schedule is as follows: Day 1: 80 mg Day 8: 40 mg Every other week: 40 mg Adalimumab does require blood work monitoring before you begin treatment and every year thereafter. Commonly, patients are checked for HIV, hepatitis, TB, blood count, kidney/liver function, and cholesterol.

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QUESTION	RESPONSE
What are the side effects of adalimumab ?	Adalimumab should not be used by patients who are pregnant. You should not receive a live vaccine (MMR, chickenpox, yellow fever, rotavirus, smallpox) while on adalimumab. Common side effects include injection site reaction, upper respiratory tract infections, sinusitis, headache, and rash.
How is infliximab administered?	Infliximab is administered as an infusion at weeks 0, 2, and 6 in your provider's office or by an infusion center. A maintenance dose is given every 8 weeks. Infliximab does not require blood work monitoring before you begin treatment and every year thereafter. Commonly, patients are checked for HIV, hepatitis, TB, blood count, kidney/liver function, and cholesterol.
What are the side effects of infliximab ?	It is unclear if infliximab is safe during pregnancy, therefore caution is advised. Infliximab should only be used if the potential risks are clearly understood, and no other option remains. You should not receive a live vaccine (MMR, chickenpox, yellow fever, rotavirus, smallpox) while on infliximab. Common side effects include upper respiratory infections, sinusitis, sore throat, headache, abdominal pain, and infusion-related reactions.
How is certolizumab pegol administered?	Certolizumab pegol is a self-injection administered at home. The injection schedule is as follows: Week 0: 400 mg Week 2: 400 mg Week 4: 400 mg Every other week: 200 mg Certolizumab pegol does require blood work monitoring before you begin treatment and every year thereafter. Commonly, patients are checked for HIV, hepatitis, TB, blood count, kidney/liver function, and cholesterol.

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QUESTION	RESPONSE
What are the side effects of certolizumab pegol ?	Certolizumab pegol can be used in individuals who are pregnant. You should not receive a live vaccine (MMR, chickenpox, yellow fever, rotavirus, smallpox) while on certolizumab pegol. Common side effects include injections site reactions, upper respiratory tract infections, rash, and urinary tract infections.
How is spesolimab administered?	Spesolimab, the only FDA-approved treatment for GPP, is administered at an infusion center over a 90-minute period. Most patients are clear after one treatment. However, some need a second infusion 1 week later. Some people feel a little different after having an infusion and you may want to rest.
What are the side effects of spesolimab ?	The most common side effects of spesolimab are infusion site reactions with pain and bruising, feeling tired, nausea and vomiting, headaches, and itching. Spesolimab does increase your risk of infections, so you will want to discuss this side effect with your HCP to ensure all steps are taken to keep you healthy.

Tailoring Treatment to the Individual Patient

Long-term goals should be to delay or prevent future GPP flares.¹ First, it is important that patients adhere to the treatment plan, which may include general skin care recommendations, medications, and other interventions. When it comes to building a treatment plan, it is important that patients work with their care team throughout the decision-making process. By devising a treatment plan that matches their preferences and goals, it is more likely that patients will adhere to the regimen, increasing the chances of treatment success.²

Additionally, there are various triggers that may cause a GPP flare. Some known triggers include emotional stress, starting or stopping certain medications, and infections, including COVID-19. Although some of these triggers are unavoidable, minimizing patient exposure is crucial. For example, stress can trigger a disease flare in some patients, so a patient may find yoga or meditation to be helpful.³ A flare could also be triggered by pregnancy or other medical conditions such as a low level of calcium in the blood in people with a hormonal disorder.⁴

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Questions That Patients May Have and Potential Responses

QUESTION/CONCERN	RESPONSE
Will I be able to do _____ again after receiving treatment?	You and your care team will assess your outcomes, improvement, treatment response, and any side effects at follow-up visits. Most therapeutic options have assigned dosing schedules based upon clinical studies.
I heard about clinical trials for _____ therapy. Will that work for my symptoms?	We are living in such an exciting time for medicine. There are so many medications being investigated to help with GPP. The hope is that in the future, we will have more options with which to treat GPP. However, right now, the only FDA-approved treatment is spesolimab.
Will this be covered by insurance/Medicaid?	Most pharmaceutical companies offer robust patient-assistance programs allowing patients who are commercially insured to receive the therapy at a low cost. When you attempt to get this approved by insurance, ensure the first two doses are approved.
Do I need to see a specialist?	I may refer you to a dermatology provider to help in managing your GPP. However, you do not need to see a specialist to receive prescribed treatment.
When will my skin/joints/hands/feet stop hurting?	In studies with spesolimab, most patients felt relief within 1 week of their infusion.

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>

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Resources for Patients and Families

National Psoriasis Foundation: <https://www.psoriasis.org/>

References and Resources for Clinicians

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