

Oscar Clinical Guideline: Spevigo (spesolimab-sbzo) (CG071, Ver. 4)

Spevigo (spesolimab-sbzo)

- Spesolimab Subcutaneous 150mg/1mL Solution for injection
- Spesolimab Intravenous 450mg/7.5mL Solution for injection

Disclaimer

Clinical guidelines are developed and adopted to establish evidence-based clinical criteria for utilization management decisions. Clinical guidelines are applicable according to policy and plan type. The Plan may delegate utilization management decisions of certain services to third parties who may develop and adopt their own clinical criteria.

Coverage of services is subject to the terms, conditions, and limitations of a member's policy, as well as applicable state and federal law. Clinical guidelines are also subject to in-force criteria such as the Centers for Medicare & Medicaid Services (CMS) national coverage determination (NCD) or local coverage determination (LCD) for Medicare Advantage plans. Please refer to the member's policy documents (e.g., Certificate/Evidence of Coverage, Schedule of Benefits, Plan Formulary) or contact the Plan to confirm coverage.

Summary

Generalized pustular psoriasis (GPP) is a rare, severe form of psoriasis characterized by sudden, widespread eruptions of painful, sterile pustules accompanied by fever, malaise, and other systemic symptoms. GPP can be life-threatening due to complications such as sepsis, acute respiratory distress syndrome, and multi-organ failure. Treatment options have been limited, often relying on off-label use of systemic therapies like retinoids, cyclosporine, and methotrexate, which may have significant side effects and variable efficacy.

Spevigo (spesolimab-sbzo) is a targeted biologic therapy approved for the treatment of GPP flares in adults and adolescents. It is a monoclonal antibody that blocks the interleukin-36 receptor, a key driver

of inflammation in GPP. Spevigo (spesolimab-sbzo) is administered intravenously for acute flares and subcutaneously for maintenance therapy to prevent flares. It is supplied in two formulations:

1. For intravenous use in acute flares:
 - a. Single-dose vials containing 450 mg/7.5 mL (60 mg/mL) of spesolimab-sbzo
 - b. Each carton contains two vials.
2. For subcutaneous use in maintenance therapy:
 - a. Single-dose prefilled syringes containing 150 mg/mL of spesolimab-sbzo
 - b. Each carton contains two syringes.

Definitions

"**Flare**" refers to an acute episode of disease exacerbation in GPP, marked by the sudden appearance or worsening of pustules and associated symptoms.

"**Generalized pustular psoriasis (GPP)**" is a rare, severe, and potentially life-threatening form of psoriasis characterized by sudden, widespread eruptions of sterile pustules, often accompanied by systemic symptoms such as fever, malaise, and laboratory abnormalities.

"**Interleukin-36 receptor (IL-36R)**" is a key component of the immune system that, when activated, drives inflammation associated with GPP.

"**Psoriasis**" is a chronic inflammatory skin condition characterized by red, scaly patches (plaques) that can appear anywhere on the body, caused by rapid skin cell turnover.

Medical Necessity Criteria for Initial Authorization

The Plan considers Spevigo (spesolimab-sbzo) medically necessary when **ALL** of the following criteria are met:

1. The medication is prescribed by or in consultation with a dermatologist; **AND**
2. The member is 12 years of age or older and weighs at least 40 kg; **AND**
3. The member has a diagnosis of generalized pustular psoriasis (GPP); **AND**
4. The requested medication is being used for **EITHER** of the following indications:
 - a. Treatment of acute GPP flares **AND ALL** of the following:
 - i. The member has a flare of GPP of moderate-to-severe intensity, as defined by:
 1. A Generalized Pustular Psoriasis Physician Global Assessment (GPPPGA) total score of at least 3 (moderate); **and**

2. Presence of fresh pustules (new appearance or worsening of pustules); **and**
 3. GPPPGA pustulation sub-score of at least 2 (mild); **and**
 4. At least 5% of body surface area covered with erythema and the presence of pustules; **and**
- ii. The member has primary, sterile, macroscopically visible pustules on non-acral skin (excluding cases where pustulation is restricted to psoriatic plaques); **and**
 - iii. The dosing regimen does **NOT** exceed a maximum of 1800 mg (two 900 mg doses):
 1. Initial dose: 900 mg administered intravenously; **and**
 2. If flare symptoms persist, a second 900 mg intravenous dose may be administered 1 week after the initial dose; **or**
- b. Prevention of GPP flares (Maintenance Therapy) **AND ALL** of the following:
- i. The member has a history of at least two moderate-to-severe GPP flares in the past; **and**
 - ii. **ONE** of the following:
 1. The member has previously responded to treatment with Spexigo; **or**
 2. The member has had an inadequate response, intolerance, or contraindication to at least one systemic therapy for GPP (e.g., acitretin, cyclosporine, methotrexate); **and**
 - iii. The member is not currently experiencing an acute GPP flare; **and**
 - iv. The dosing regimen does **NOT** exceed a maximum of 600 mg (four 150 mg injections) for initial dose, then 300 mg (two 150 mg injections) every 4 weeks:
 1. Initial dose: 600 mg administered subcutaneously; **and**
 2. Maintenance: 300 mg administered subcutaneously every 4 weeks.

If the above prior authorization criteria are met, the requested product will be authorized as follows:

- **Treatment of Acute GPP Flares: A one-time treatment course of up to two doses (900 mg each) within a 1-week period.**
- **Prevention of GPP Flares (Maintenance Therapy): Initial authorization for 6 months**

NOTE: Members transitioning from acute flare treatment to maintenance therapy should initiate subcutaneous dosing 4 weeks after the last intravenous dose, starting with the 300 mg maintenance dose. A subcutaneous loading dose is not required when transitioning from IV to SC maintenance dosing.

Medical Necessity Criteria for Reauthorization

Reauthorization for 12-months will be granted if the member has recent (within the last 3 months) clinical chart documentation demonstrating **ALL** of the following criteria:

1. The member continues to meet applicable **Initial Authorization** criteria; AND
2. The member has demonstrated a positive clinical response to Spevigo (spesolimab-sbzo) therapy as evidenced by at least **ONE** of the following:
 - a. Reduction in frequency of GPP flares; **and/or**
 - b. Reduction in severity of GPP flares; **and/or**
 - c. Decreased duration of GPP flares; **and/or**
 - d. Improvement in quality of life scores.

Experimental or Investigational / Not Medically Necessary

Spevigo (spesolimab-sbzo) for any other indication or use is considered not medically necessary by the Plan, as it is deemed to be experimental, investigational, or unproven. Non-covered indications include, but are not limited to, the following:

- Treatment of hidradenitis suppurativa
- Treatment of other pustular skin conditions such as palmoplantar pustulosis
- Treatment of plaque psoriasis without GPP
- Treatment of other autoimmune or inflammatory conditions not specified in the FDA-approved labeling

Applicable Billing Codes (HCPCS/CPT Codes)

Service(s) name	
CPT/HCPCS Codes considered medically necessary if criteria are met:	
<i>Code</i>	<i>Description</i>
96365	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to 1 hour
96366	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); each additional hour (List separately in addition to code for primary procedure)

96372	Therapeutic, prophylactic, or diagnostic injection (specify substance or drug); subcutaneous or intramuscular
J1747	Injection, spesolimab-sbzo, 1 mg
ICD-10 codes considered medically necessary if criteria are met:	
<i>Code</i>	<i>Description</i>
L40.1	Generalized pustular psoriasis

References

1. Bachelez H, Choon SE, Marrakchi S, et al. Trial of spesolimab for generalized pustular psoriasis. *N Engl J Med.* 2021;385(26):2431-2440. doi:10.1056/NEJMoa2111563
2. Krueger, J., Puig, L., & Thaçi, D. (2022). Treatment options and goals for patients with generalized pustular psoriasis. *American Journal of Clinical Dermatology*, 1-14.
3. Marrakchi, S., & Puig, L. (2022). Pathophysiology of generalized pustular psoriasis. *American Journal of Clinical Dermatology*, 1-7.
4. Menter, A., Van Voorhees, A. S., & Hsu, S. (2021). Pustular psoriasis: a narrative review of recent developments in pathophysiology and therapeutic options. *Dermatology and Therapy*, 11(6), 1917-1929.
5. Morita A, Strober B, Burden AD, Choon SE, Anadkat MJ, Marrakchi S, Tsai TF, Gordon KB, Thaçi D, Zheng M, Hu N, Haeufel T, Thoma C, Lebwohl MG. Efficacy and safety of subcutaneous spesolimab for the prevention of generalised pustular psoriasis flares (Effisayil 2): an international, multicentre, randomised, placebo-controlled trial. *Lancet.* 2023 Oct 28;402(10412):1541-1551. doi: 10.1016/S0140-6736(23)01378-8. Epub 2023 Sep 19. PMID: 37738999.
6. Mrowietz, U., Burden, A. D., Pinter, A., Reich, K., Schäkel, K., Baum, P., ... & Bissonnette, R. (2021). Spesolimab, an anti-interleukin-36 receptor antibody, in patients with palmoplantar pustulosis: results of a phase IIa, multicenter, double-blind, randomized, placebo-controlled pilot study. *Dermatology and therapy*, 11(2), 571-585.
7. Spevigo (spesolimab) [prescribing information]. Ridgefield, Connecticut: Boehringer Ingelheim Pharmaceuticals Inc; September 2022.
8. Spevigo (spesolimab) [prescribing information]. Ridgefield, Connecticut: Boehringer Ingelheim Pharmaceuticals Inc; March 2024.

Clinical Guideline Revision / History Information

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